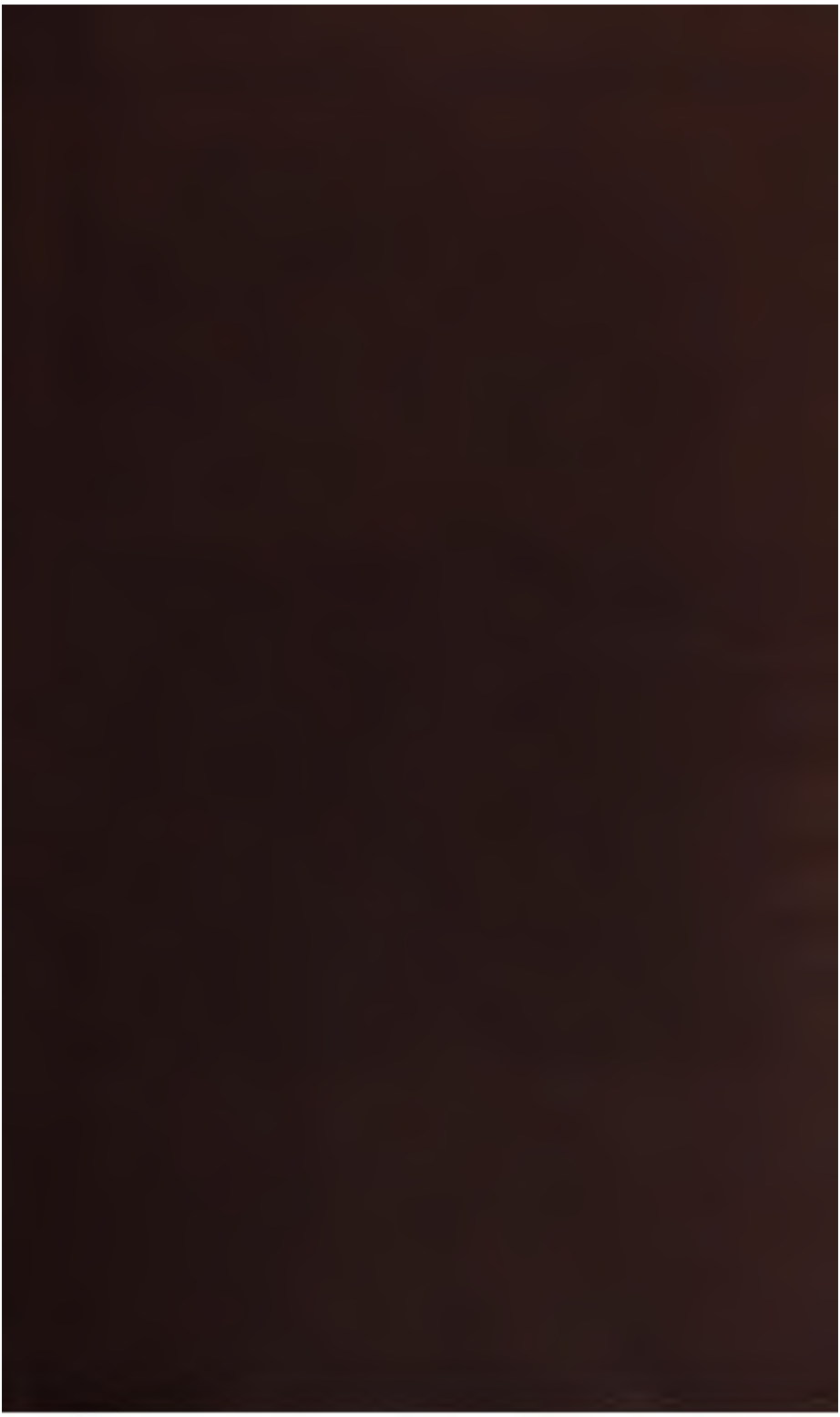

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THE NEW SYDENHAM SOCIETY.

VOLUME CLII.

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LECTURES

ON

DISEASES OF THE SPINAL CORD

BY

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CONTAINING TWO HUNDRED AND FORTY-FOUR WOODCUTS.

TRANSLATED BY

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AUTHOR'S PREFACE.

THE lectures published in this book were taken from those delivered at the Faculty of Medicine in the summer of 1891. Although but little altered, some works which appeared during the course of their publication were of such great importance that it seemed unwise to neglect them, and they were consequently utilized by me.

I have advisedly given the name of "Lectures" to this publication, more freedom of speech being thus allowed, while it may constantly be recognized during its perusal that the book is specially written for the purpose of "teaching."

Since some diseases only of the spinal cord are described in this book, I have endeavoured to discuss these as completely as possible. Numerous works have been consulted upon this branch of nervous pathology, care being taken to name their authors, although it did not accord with the plan of this publication to indicate the name of every book to which allusion is made. Those mentioned have been somewhat arbitrarily chosen, their importance, their novelty, and the difficulty in obtaining them being specially considered.

Little will be found in this work about the pathology of any special symptom. Thus I have purposely omitted to discuss the mechanism of the inco-ordination which occurs in tabes. Our knowledge with respect to this and other similar conditions is so rudimentary, and the opinions expressed about them are so contradictory that, as I believe, it is better to await a more propitious epoch before doing this.

The study of Ætiology on the other hand owing to the immense progress made in general pathology is steadily advancing; to this portion of the subject I have devoted special attention, since it is from a knowledge of it that Therapeutics worthy of the name must be derived. The symptoms have also been carefully studied.

Pathological anatomy is still a somewhat uncertain guide as regards numerous points. It should be chiefly based upon a knowledge of the anatomy of the part in a normal condition, and on that account I have thought it right to describe in these lectures the principal facts connected with the anatomy of the spinal cord. These are discussed at some length, and I trust that I have made them more or less clear by the use of numerous figures, either diagrammatic or from nature. Many of these were obtained from collections in the Hospital of the Salpêtrière, and used in these lectures with the full permission of my master, Professor Charcot, to whom I offer once more my sincere thanks.

Paris, 1892. PIERRE MARIE.

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LECTURES

ON

DISEASES OF THE SPINAL CORD.

LECTURE I.

ANATOMY OF THE PYRAMIDAL TRACT.

Reason for this denomination. Path of the pyramidal tract; its origin in the motor convolutions; its course in the centrum ovale; its seat in the internal capsule, where it occupies the anterior two-thirds of the posterior limb; its path in the crus cerebri, pons varolii, and medulla oblongata; its decussation, and path in the spinal cord.—Division into crossed (FPyC) and direct (FPyD) pyramidal tract.—Path of the crossed tract (FPyC) which is separated from the surface of the cord by the fibres of the direct cerebellar tract; its form in the different regions of the cord.—Path of the direct tract (FPyD), which does not exist in the lower part of the cord; variations in its size; these variations correspond with those in the decussation of the pyramidal tract.—The pyramidal tract in certain animals.—Development of the pyramidal tract.—Collateral branches described by Golgi, Ramon y Cajal, Kölliker, v. Lenhossek.—Termination of the pyramidal tract.

GENTLEMEN,—Before commencing to study the diseases of the Spinal Cord the principal facts connected with its anatomy will be considered, a knowledge of which is indispensable.

Our object being to combine Pathology and Anatomy, the different parts of the cord will only be described anatomically in so far as this consideration relates to the diseases from which they suffer. Morbid and healthy conditions will be contrasted, normal and pathological anatomy being found to mutually enlighten each other.

Thus the study of secondary degeneration will enable us to become rapidly acquainted with the principal tracts of the cord. In the first lectures such degeneration will be specially considered, the anatomy and origin of the numerous tracts of fibres

which compose the spinal cord showing it to be a cable of transmission rather than a central organ.

The PYRAMIDAL TRACT (FPy) holds an important place in many different alterations of the spinal cord. The knowledge which we possess about this tract, and the secondary degeneration to which it is liable will therefore be first considered.

The name *pyramidal tract* was derived from the fact that this column, which is seated in the anterior portion of the bulb, is comprised within the elongated bundles of white matter in that region which are termed *pyramids*.

The path of this tract is well known and has been long understood. In this, as in many other points, connected with neurology, pathological anatomy preceded pure anatomy. The works of Türck, of Charcot and Bouchard, of which the importance is well known to you, have specially utilized the secondary degeneration which occurs in the spinal cord after hemiplegia in their description of the pyramidal tract. At a later date Flechsig by a new method of investigation reopened this apparently exhausted question, and the description which he has given is undoubtedly correct, and will be often utilized during the following lectures.

Considered throughout its whole length the pyramidal tract extends from the cerebral convolutions through the hemispheres, crura cerebri, pons varolii, and medulla oblongata, to the lower part of the spinal cord.

The fibres of the pyramidal tract originate in the so-called *motor convolutions*, viz., the *ascending frontal and parietal convolutions*, and the *paracentral lobule*, or medial aspect of these two convolutions. Whether fibres from other parts of the brain also join this tract is uncertain, but its fibres undoubtedly come for the most part from these convolutions, in all probability from the cells of the cortex, and partly or entirely from the special cells which are termed the *large pyramidal cells** of the motor convolutions.

From the grey layer of the cortex the fibres of the pyramidal tract pass into the white substance of the motor convolutions, where they assume the form of a fan, as they converge towards the more internal parts of the brain. It is thus that they pass

*The name of *pyramidal* cells is simply due to their form, and in no way to a real or supposed connection with the *pyramidal* tract.

through the *centrum ovale* subjacent to the motor convolutions.

The fibres follow this convergent direction, and before going

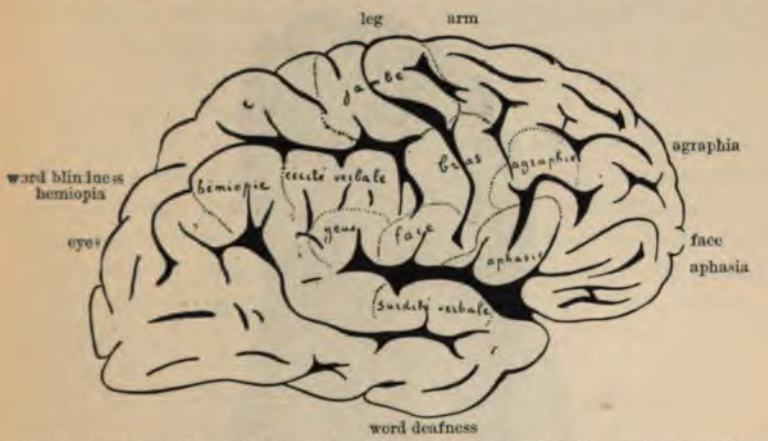

Fig. 1.—Outer surface of the cerebral hemisphere showing the motor convolutions (leg, arm, face). (This figure was extracted from the Atlas of Brissaud* upon the Anatomy of the Brain.)

Fig. 2.—Median aspect of the cerebral hemisphere showing the motor convolutions. (This figure was extracted from the Atlas of Brissaud upon the Anatomy of the Brain.)

far form a compact mass, the pyramidal tract, the fibres of which can be localized in a more exact manner. This localization is

* I have to thank my friend and colleague, Brissaud, for permission to use these figures before the publication of his Atlas upon the Anatomy of the Brain, from which they are taken.

specially interesting in the *internal capsule* on account of the differentiation which can be made of the numerous tracts which constitute this cross-road (*carrefour*, Charcot).

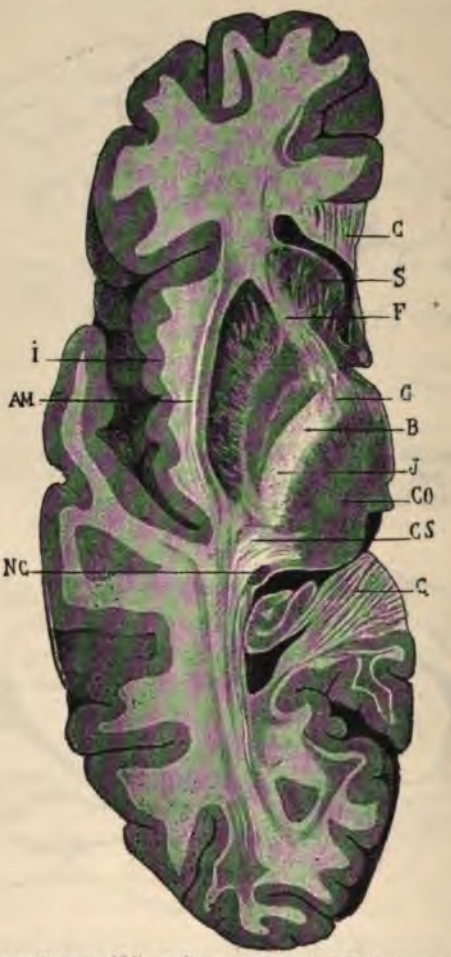

Fig. 3.—S, corpus striatum; NC, nucleus caudatus; CO, thalamus opticus; F, anterior limb of the internal capsule; G, knee of the internal capsule; B, fibres of the internal capsule connected with the movements of the upper extremity. J, fibres of the internal capsule connected with the movements of the lower extremity; CS, sensory cross-road. (This figure was extracted from the Atlas of Brissaud upon the Anatomy of the Brain.)

The position of the different tracts is most easily studied, as you know, by the horizontal division of the brain termed *the*

section of Flechsig, and, according to the recommendation of Brissaud, the following is the best direction for the section to have: the knife should pass through the inner surface of the hemisphere in a direction which is slightly oblique downwards and backwards so as to divide the corpus striatum in the centre of its head, and the thalamus opticus at the junction of its upper third with its lower two-thirds.

The anterior and posterior limbs of the internal capsule and the *knee* which separates them can be recognized in this section, the pyramidal tract being found, as Charcot expresses it, in the posterior limb of which it occupies the anterior two-thirds. It may itself be divided into secondary tracts, which from before backwards are connected respectively with the face, tongue, and limbs.

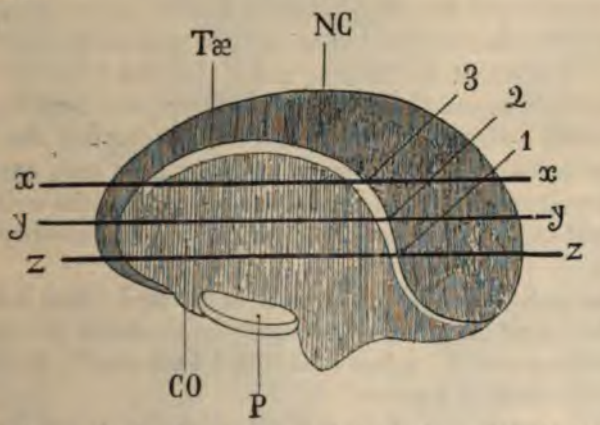

Fig. 4.—Diagram showing that the knee of the internal capsule varies in position according to the height at which the horizontal section of the hemisphere is made. (After the Atlas of Brissaud upon the Anatomy of the Brain.)

NC, nucleus caudatus; CO, thalamus opticus; P, crus cerebri; Tæ, tænia semicircularis; XYZ, horizontal sections through the corpus striatum and thalamus opticus involving the tænia semicircularis at the points 1, 2, and 3. The points 1, 2, and 3 correspond to three points in the knee of the internal capsule, which are seated at different levels.

Upon reaching the crus cerebri the tract occupies the middle part of the lower portion, or crusta, having on its inner side a band of fibres which come from the anterior part of the brain and pass downwards into the pons, beyond which they cannot be followed. On the outer side of the tract (FPy) is another band of fibres which seemingly come from the temporal and occipital

lobes, and which are, at times, though very rarely (Brissaud), found to degenerate (Bechterew), the degeneration ceasing when the pons is reached. The tract itself (FPy) may be divided into secondary tracts, which from within outwards are destined respectively for the face, tongue, and limbs.

In the *pons varolii* the fibres of the tract (FPy) again occupy the crustal portion, being divided into flattened bundles, which are separated from each other by some of the transverse fibres of the pons. The size of the tract (FPy) is here much larger than in the upper part of the pyramids in the medulla oblongata, and it seems probable that, as some authors believe, many of its fibres terminate in the pons.

In the *medulla oblongata*, on the other hand, the tracts (FPy), as already stated, are completely isolated from the other fasciculi in that part, and form the prominent bundles known as pyramids.

You will observe, gentlemen, that the summit of these "pyramids" is directed downwards, and that at the lower part of the medulla oblongata the tract (FPy) disappears from its fibres, remaining no longer at the surface, but passing into the interior of the bulbo-medullary axis, and, for the most part, crossing to the lateral column of the opposite side. Such is the well-known *decussation of the pyramids*, of which it is unnecessary to say more.

The path of the pyramidal tract in the *cord*, which it has now reached, must be studied most carefully, since it is in connection with diseases of the spinal cord that I have rapidly summarized the path which it pursues.

In the lower part of the medulla oblongata, as you know, the tract (FPy) divides into two bands, one of which does not change its direction, being called on that account the *direct pyramidal tract* (FPyD), while the other passes into the lateral column upon the opposite side of the cord, being therefore called the *crossed pyramidal tract* (FPyC). Each of these tracts will now be separately studied.

THE CROSSED PYRAMIDAL TRACT (FPyC), by far the most important as regards its anatomy and pathology, extends almost to the termination of the spinal cord. Some authors (Vulpian, Löwenthal) believe it to end at the level of the second lumbar pair of nerves, while others have traced it farther, notably Tooth, who found it below the fourth lumbar pair. Very possibly it

varies in length, individual differences existing, which are analogous to those of which I shall have to speak as regards its variations in breadth.

The seat of the tract is as follows: throughout almost its whole course it is in relation internally and posteriorly with the posterior horn until almost the extremity of its path. The direct cerebellar tract separates it from the surface of the cord (except, according to Gowers, for a short distance at the level of the third cervical pair of nerves); the consequence is that in those parts where the direct cerebellar tract does not yet exist,

Fig. 5.

Fig. 6.

Fig. 7.

Fig. 8.

Fig. 9.

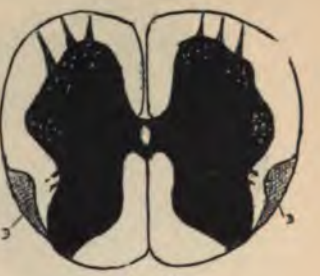

Fig. 10.

Sections of the spinal cord in which the two direct pyramidal tracts A, and the crossed pyramidal tracts B, present respectively the same symmetrical relations (after M. Flechsig). Fig. 5 at the level of the 2nd cervical pair. Fig. 6 at that of the 7th cervical. Fig. 7 at that of the 3rd dorsal. Fig. 8 at that of the 6th dorsal. Fig. 9 at that of the 1st lumbar. Fig. 10 at that of the 5th lumbar pair of nerves.

that is to say, at some point below one of the last pairs of dorsal nerves, according to the subject, the crossed pyramidal tract (FPyC) extends outwards as far as the surface of the cord. Anteriorly the tract, even where most developed, does not extend beyond an imaginary line, passing transversely through

the posterior commissure. At the same time it will be seen, when the secondary degeneration consequent upon transverse lesions of the spinal cord, and the pathological anatomy of amyotrophic lateral sclerosis are considered, that the limits of this tract are scarcely as well defined, as has been stated, anteriorly, and the presence of stray fibres belonging to this tract in other parts of the antero-lateral column, though not certain, is at any rate quite possible, the fact that individual differences may exist being always remembered.

The *form* of the crossed pyramidal tract (FPyC) varies at different heights. It is more or less oval in the cervical region, being somewhat triangular, with the apex seated antero-internally in the dorsal, and antero-externally in the lumbar region.

Its *size* decreases regularly from above downwards, on account of the way in which fibres continually leave the tract in order to pass into the grey matter.

THE DIRECT PYRAMIDAL TRACT (FPyD) differs from the crossed pyramidal tract both by its different seat in the spinal cord, and

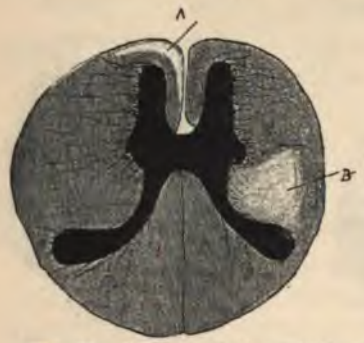

Fig. 11.—Section of the spinal cord (upper part of cervical region) in a case of hemiplegia due to a cerebral lesion. A, direct pyramidal tract, which has here a flattened form, and extends in an unusual way to the outer side of the anterior horn. B, crossed pyramidal tract. The white portion is affected by sclerosis (Damaschino collection).

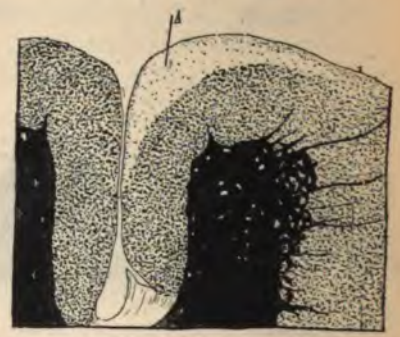

Fig. 12.—Anterior horn and direct pyramidal tract of the same section as in the preceding figure more highly magnified. (By an error on the part of the designer the patch of sclerosis A is upon the right instead of the left side, as in fig. 11.) A, the direct pyramidal tract with its singular flattened form (Damaschino collection).

its shorter course. According to Bouchard, this tract (FPyD) does not usually extend beyond the mid-dorsal region. Tooth,

however, has traced it as far as the second pair of lumbar nerves. It should be observed, however, that its length seems subject to numerous variations. Thus Bechterew states that he has known it disappear below the cervical region. Though exceptions may exist, it should be remembered, however, that as a rule the direct pyramidal tract does not extend beyond the middle portion of the dorsal region.

When a section is made its situation and form are found liable to similar variations. In some cases this tract (FPyD) extends along the whole length of the anterior median fissure, beyond which it sometimes passes externally (*vide* figs. 11 and 12), while in others it seems to occupy a very small space, and to be, as it were, surrounded by the adjoining fibres.

Fig. 13.—Anterior surface of the medulla oblongata, the shaded portion A representing the pyramids, which terminate far more abruptly than usual. In this case there was decussation of the whole pyramid, and not of part only, as is usually the case. There was, therefore, no direct pyramidal tract. (After Flechsig.)

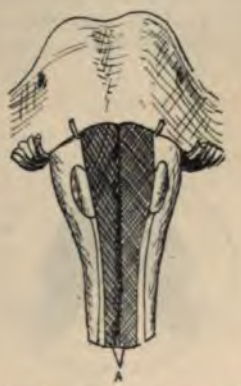

Fig. 14.—Anterior surface of the medulla oblongata, the shaded portion A representing the pyramids. These extend far lower than is normally the case. The decussation of the pyramids in this patient was very incomplete, the greater number of fibres remaining in the direct pyramidal tract. (After Flechsig.)

It must not be supposed that the relation between the sizes of the direct (FPyD) and crossed pyramidal tract (FPyC) is by any means invariable; the individual differences which exist, as Flechsig has shown, are by no means small.

In some cases, in fact, symmetrical semi-decussation occurs,

both pyramids furnishing a direct and crossed pyramidal tract, which more or less correspond with each other. This, according to Flechsig, is what usually happens (in 75 per cent. of the cases).

At other times the whole pyramid decussates, and the direct

Fig. 15.—Section of the spinal cord in which the direct (A) is far larger than the crossed pyramidal tract (B). From the same patient as fig. 14. (After Flechsig.)

Fig. 16.—Section of the same cord (upper part of dorsal region) as figs. 14 and 15. Same comparative size of the direct (A) and crossed pyramidal tracts (B). (After Flechsig.)

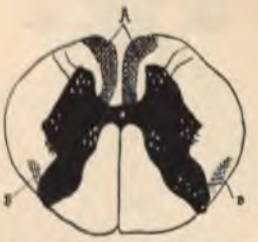

Fig. 17.—Section of the same cord (lower part of the dorsal region) as in figs. 14, 15, and 16. Same comparative size of the direct (A) and crossed pyramidal tracts (B). (After Flechsig.)

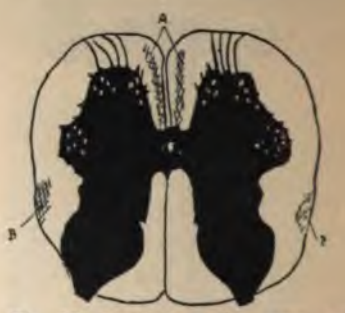

Fig. 18.—Section of the same cord (level of 4th lumbar pair of nerves) as in figs. 14, 15, 16, and 17. In this cord, in which the pyramidal fibres are mostly contained in the direct pyramidal tract, traces of this tract (A) are seen in the lowest part of the lumbar cord, instead of its disappearing, as usually happens, in the lower part of the dorsal region. (After Flechsig.)

pyramidal tracts are completely absent. Inversely the direct pyramidal tracts (FPyD) are in some cases of larger size than those which cross to the other side (FPyC). (*Vide* figs. 14, 15, 16, 17, 18.)

In some cases the right and left pyramidal tracts act differently, and the decussation is then "asymmetrical" (Charcot).

Thus one of the pyramids, usually the left, may be larger than the other by as much as about a third of its volume. It may happen, for example, that the direct tract (FPyD) on the left is larger than that on the opposite side. In some cases again the direct pyramidal tract (FPyD) only exists upon one side (figs. 19 and 20).

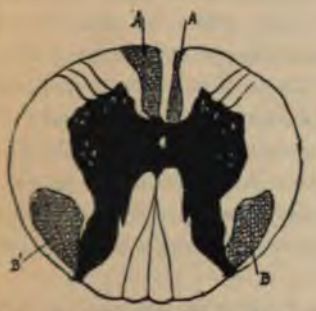

Fig. 19.—Section of a cord (cervical region) in which the direct and crossed pyramidal tracts are symmetrically unequal in size. The crossed pyramidal tract B, which is smaller than B', corresponds to a direct pyramidal tract A, which is larger than A'.

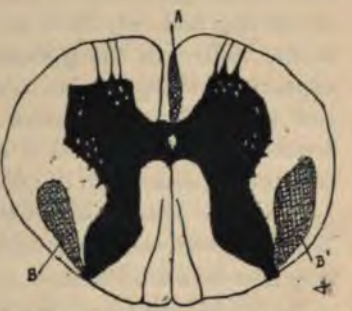

Fig. 20.—Section of a cord (cervical region) in which the direct pyramidal tract (A) only exists upon one side. The crossed pyramidal tract (B') upon the same side far exceeds that of the opposite side in size, as if it contained, in addition to its own fibres, those which should have formed the direct tract which is absent. (After Flechsig.)

Similar variations in the pyramidal tract occur in *animals*, and several observations of Bechterew, who has studied this subject, and which are contained in his recent work, will be mentioned.

The time of its development differs widely, and while in man the pyramidal tract is not completely developed until after birth, in some animals, such as the guinea-pig, the tract is fully developed when it is born, and the same is the case with other animals which can run easily from the first. Thus there is a curious correspondence between the state of powerlessness in which the young of the higher mammalia are placed at birth, and that which exists in man, in whom the pyramidal tract is not yet completely developed.

The size of the pyramidal tract also depends upon the movements which the animal is able to perform. In those animals of which the movements are but slightly differentiated, such as the whale and the elephant, the pyramidal tract is almost entirely absent, while in those living beings by which varied and delicate movements are performed, such as men and monkeys, the pyramidal tract attains its full development.

Differences again exist as regards the division into a direct and crossed pyramidal tract; some animals, for instance, such as the dog and cat, have no direct tract, while in others, such as the white rat, the pyramidal tract passes into the anterior segment of the posterior column. These facts are of interest, and show that the knowledge obtained from the study of degeneration in the cord of some animals cannot be applied in an absolute manner to the degeneration which occurs in man.

The position and path of the pyramidal tract and its divisions being understood, the general facts which relate to its study may now be considered.

Its *development* is of special interest, since it occurs at quite a late date. Thus in the human embryo 12 centimetres (4¾ in.) in length the pyramidal tract is completely absent although the other tracts of the cord can be distinctly perceived.

Those who have investigated this question believe that the pyramidal tracts are in all probability formed towards the middle or end of the 5th month of fœtal life. At the same time the sheath of myelin is not found until the end of the 9th month. This fact is of importance, and enabled Flechsig to study the precise path of this tract, which was previously unknown. Since osmic acid gives a black colour to myelin * the parts which remain of a grey or white hue when this reagent is applied to the cord, are those in which no myelin as yet exists, and Flechsig has thus been able to follow the development of the different tracts of the cord in their various stages.

The pyramidal tracts are essentially composed of fibres parallel to the axis of the cord, which are of considerable length, some of them extending from the cortex of the brain to the lower part of the lumbar region of the cord. From these, as also from the posterior columns, many of the *collaterals* emerge (Golgi, Ramon y Cajal, Kölliker, &c.). These collaterals,

* *Vide* Medical Microscopy, by F. J. Wethered, p. 90 (Translator).

according to Lenhossek, do not pass as one would expect, and as most authors believe, into the anterior, but into the posterior cornua. There they form arborescent divisions in such a way as only to be connected with the cells of the anterior cornua by the medium of other intervening cells. I must remind you that the pyramidal tracts do not form an absolutely compact mass, but between the fibres composing them others of different origin exist which are not affected by secondary descending degeneration, a lesion which indicates that the pyramidal tract is involved.

As regards the *connections* of these fibres I have already had occasion, gentlemen, to mention their origin in the cortex of the brain; I still have to speak of their *termination* in the spinal cord.

This part of their path is certainly one of the least known.

According to the generally accepted opinions the connections of these tracts are as follows:—

As regards the *crossed pyramidal tract* it is connected with the cells of the anterior cornua, the lateral parts of the grey matter, and the crossed pyramidal tract (FPyC) of the opposite side.

The *direct pyramidal tract* is connected with the cells of the anterior cornua, the internal portion of the grey substance of those horns, and the crossed pyramidal tract (FPyC) of the opposite side.

It must be acknowledged, however, that these connections, specially as you have already seen, the direct connection of the fibres of the pyramidal tract with the cells of the anterior cornua, have by no means been objectively shown to exist.

The intrinsic mode of action of the pyramidal tract is little better understood. We certainly know that it is pre-eminently the tract by means of which *voluntary movements are made*, but in what way we are completely ignorant.

It may be supposed that the nervous influence which passes through the pyramidal fibres has either an excitomotor or inhibitory effect upon the cells of the cord, a point which will be again considered when the contracture associated with secondary degeneration is mentioned. Before ending this chapter I would ask you to remember an assertion of Hómen, according to which an isolated lesion in the direct pyramidal

tract (FPyD) produces more pronounced paralysis than one in the crossed tract (FPyC), the paralysis occurring on the side opposite to that of the lesion. According to Gowers, on the other hand, the function of the direct tract (FPyD) is specially connected with the movement of the upper limbs. Thus, gentlemen, no definite agreement can by any means be said to exist as yet in connection with the action of the two parts of the pyramidal tract.

LECTURE II.

SECONDARY DEGENERATION OF THE PYRAMIDAL TRACT.

HISTORY: Cruveilhier, Türck, Charcot, Vulpian, Leyden, Cornil, &c.; Memoir of Bouchard (1866); Thesis of Brissaud (1880).

Conditions in which secondary degeneration occurs in this tract. Clinical indications of the degeneration: spastic symptoms, excess of tendon reflexes, contracture, mode of production of these symptoms; theories on this subject.

Bilateral degeneration of the pyramidal tracts consecutive to a unilateral cerebral lesion. Works of Pitres and Dignat. Varying seat of the degenerated part. Theories proposed in explanation of these facts.

Co-existence of secondary degeneration of the pyramidal tract with *amyotrophy*. Statement of such facts by numerous authors whose observations have given rise to different theories.

GENTLEMEN,—In the preceding lecture the principal facts connected with the anatomy of the pyramidal tract were discussed in detail. The *degenerations of that tract consecutive to cerebral lesions* will now be studied, the portion of the pyramidal tract contained in the spinal cord being alone considered.

The existence of such degeneration has been known for many years. Cruveilhier had already observed atrophy of the pyramid upon one side in some cases of hemiplegia, but had not perceived that the degeneration extended beyond this point. The honour of discovering, and in a remarkable way, that the pyramidal tract degenerates also in the spinal cord must be given to Türck, who described such degeneration in 1851—55. A few years later these observations were confirmed and extended by Charcot, Vulpian, Leyden, Cornil, &c., and in 1866 the important memoir of Ch. Bouchard appeared, which marks, as you know, an important epoch in the history of secondary degeneration. The fact is that after this work nothing was observed upon the subject during several years. In 1880 the inaugural thesis of Brissaud was written, this being the most complete and suggestive account of the subject in which we are now interested which has hitherto been given.

Although different varieties of cerebral lesion may produce secondary degeneration in the pyramidal tract, the lesion must

be connected with *some point in the path of the pyramidal fibres*, and be also *destructive in nature*. You know in what part of the brain the lesion must be seated in order to fulfil the first of these conditions, namely, at the very origin of the pyramidal fibres in the so-called *central convolutions near the fissure of Rolando*, or in the path of these fibres in the *centrum ovale*, or lastly in the anterior two-thirds of the posterior limb of the *internal capsule* where these fibres are seated. The second condition that the lesion must be destructive in nature should be explained. Mere compression of the pyramidal fibres, as by cerebral tumours, or meningeal lesions which are superficial would not probably produce secondary degeneration in the spinal cord. Opinions differ as to whether removal of the grey substance of the convolutions, without any lesion of the white matter, could be followed by secondary degeneration. It seems quite possible that this may be the case if the lesion is not too superficial, since it is in the deeper portion of the grey matter of the cortex that the large cells are seated, from which the fibres of the pyramidal tract seem to take origin.

When the two conditions which have just been mentioned exist, the trophic centre of the pyramidal fibres being seated in the cortex, the peripheral termination of these fibres in the spinal cord degenerates according to the law of Waller. The axis cylinder perishes, the medullary sheath of myelin undergoes the changes which usually occur in degeneration, and during the somewhat long period of its absorption granular bodies are observed in greater or less number at the part where degeneration is taking place. At the same time or shortly afterwards there is proliferation of the neuroglia. Is this due, as some authors have stated, to inflammation of the neuroglia by extension of the inflammatory condition which exists in the fibres of the pyramidal tract, or as Bouchard maintains, to simple proliferation of the neuroglia which fills the empty spaces resulting from disappearance of these fibres? For my part I am much inclined to agree with the latter opinion.

How soon after production of the cerebral lesion do the first indications of secondary degeneration appear? It is to Bouchard that we must again refer for an answer to this question. According to that author the first traces of descending degeneration can be recognized anatomically upon

about the sixth day. Clinical examination enables it to be traced some time before this. Thus Pitres has noticed the existence of the foot phenomenon as soon as 20, or even 10 hours after the occurrence of hemiplegia. At the same time, in most cases the existence of secondary degeneration cannot be recognized with certainty until the end of a week after the occurrence of the cerebral lesion. I shall also have occasion to mention that the foot phenomenon, or increase of the tendon reflexes, so soon after the occurrence of hemiplegia is not always a sign of descending degeneration.

I need not again describe in detail the parts in which degeneration occurs. I should merely have to repeat word for word what I have already said about the anatomy of the pyramidal tract. It will suffice to place before you a few different sections of the cord after hemiplegia due to a lesion in one of the cerebral hemispheres (figs. 21, 22, 23, 24, 25).

We have already seen, gentlemen, in their collective character, what, from an anatomical point of view, were the characters of secondary degeneration due to a cerebral lesion. The clinical features which characterize it deserve equal attention.

Paralysis (monoplegia or hemiplegia) is merely the direct effect of the cerebral lesion, and in no way indicates secondary degeneration. The true indication of this, which must not be forgotten, gentlemen, is solely the spasmodic condition which occurs.

Fig. 21.—Section of the medulla oblongata at its lower part. A, degeneration of the pyramid on the left side from a case of hemiplegia due to a cerebral lesion. The white zone was affected by sclerosis.

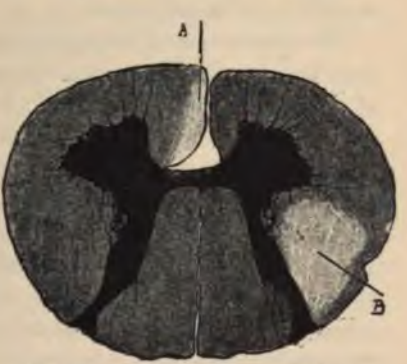

Fig. 22.—Secondary degeneration of the direct pyramidal tract A, and the crossed pyramidal tract B, in a case of hemiplegia from a cerebral lesion (cervical cord).

This spasm does not always present the same characters; in some cases it is specially marked, a contracture existing which attains a most pronounced degree, while in others there is no

Fig. 23.—Secondary degeneration of the crossed pyramidal tract in a case of hemiplegia due to a cerebral lesion. (Section of the cord in the dorsal region where the direct pyramidal tract no longer exists.)

Fig. 24.—Secondary degeneration of the crossed pyramidal tract A from a case of hemiplegia due to a cerebral lesion. (Section of the cord in the lumbar region.)

external indication of this state. The patients in whom the latter is the case have been truly said by Brissaud to be "*on the point of suffering from it*," being really, it may be said, in a condition of latent spasm.

The *appearance* of hemiplegic patients suffering from spasm is well known. In the upper extremity the position is usually one of *flexion with pronation*, in the lower extremity of *extension with adduction*. The attitude of these patients is so characteristic that they could be recognized by any observer even at a distance when walking in the street, while in the case of those who suffer from latent spasm the foot phenomenon or other forms of reflex action alone reveal the condition which exists. In these, not only is there an exaggeration of the movements, but as Brissaud has shown, both stronger muscular contraction and a diminution in *the length of time which passes between the blow being given and the movement produced* (38 instead of 45 thousandths of a second), and at times the increased irritability causes recurring contractions although but one blow has been given to the tendon.

crease in the tendon reflexes very frequently indicates

Fig. 25.—Longitudinal section of the right side of the cord from a case of hemiplegia due to a cerebral lesion. The section passes through the cord at about the union of the anterior two-thirds with the posterior third of the posterior cornu. A, healthy zone, representing the fibres of the *direct cerebellar tract*; B, zone of degeneration, formed by the *crossed pyramidal tract*; C, healthy zone, formed by the longitudinal fibres which ascend along the external margin of the posterior cornu and are connected with the system of the *lateral limiting layer*; D, healthy posterior cornu; E, healthy posterior column.

a more or less pronounced tendency to contracture. This is certain, but it must not be thought to be always the signification of this symptom. Those in whom apoplexy has occurred may present the same increase of reflex movements for some hours after an attack upon one or both sides without the occurrence of any paralysis or, for a better reason, of contracture. Thus too much importance must not be attached to such increase when it occurs during the first few days after an attack of hemiplegia, and only indicates secondary degeneration, when a certain length of time has passed since the attack occurred.

Why the spasm occurs in cases of secondary degeneration of the pyramidal tract will now be considered, though but little can be said upon this subject.

Many different explanations have been given of its occurrence. Follin believed that the deformities which exist in hemiplegia of long standing are due to retraction of the muscles and soft parts. It is certainly true that such retraction does exist, but this is merely an associated symptom, and in no way the cause of the deformities. Hitzig maintained that the contracture was due to the fact that the motor impulses extended from the healthy to the paralysed side, and produced, as it were, an uninterrupted series of movements associated with muscular spasm in the limbs upon that side. This explanation, which is ingenious, and to

some extent probable, seems to be too exclusive. The more generally believed opinion is that already adopted by Straus in his Thesis,* written in 1875, according to which the tonic spasm is merely an *increase of the muscular tone.*

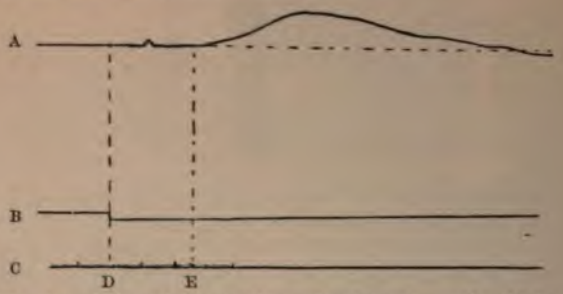

Fig. 26.—Tracing of the patellar tendon reflex taken from the thigh of the non-paralysed side in a case of hemiplegia with secondary degeneration. A, myographic line; B, line upon which the time of the blow being given is indicated; C, line upon which the duration of the reflex is indicated; DE, interval of time between the time of the blow being given and the commencement of muscular contraction = *time of reflex.* In this case the time occupied by the reflex was 40 thousandths of a second (with the correction 32 thousandths), that is to say, somewhat less than the normal time; this shows that in hemiplegia the limb upon the "sound" side has also a tendency to spasm. (After Brissaud.)

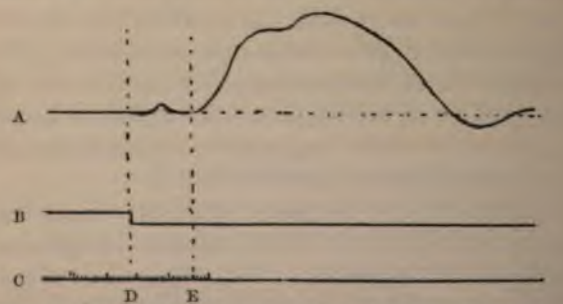

Fig. 27.—Tracing of the patellar tendon reflex taken from the thigh of the side upon which paralysis and contracture existed, in the same case of hemiplegia with secondary degeneration. Same letters as in fig. 26. It will be observed that upon this side the time of reflex DE is much shorter; it was only 36 thousandths of a second (with the correction 28 thousandths). (After Brissaud.)

It is certain in fact that in hemiplegia the spasm is due to permanent contraction of the muscles alone, as Brissaud showed in his Thesis, by a most elegant demonstration. An Esmarch's

* These d'agrégation, 1875.

bandage being applied to a limb affected by contracture, so as to render the muscles and soft parts exsanguine, and on that account unable to contract, at the end of from five to twenty minutes the spasm ceased, and any attitude whatever could be given to the limb. According to Brissaud the spasm was merely due to *permanent muscular action;* the spinal cord affected by secondary degeneration of the pyramidal tract being, as Charcot expresses it, "in a condition of spontaneous strychnism."

According to Vulpian the contracture is due, not only to increased irritability of the spinal cord on account of the disappearance of the fibres belonging to the pyramidal tract, but also and specially to the irritation which the sclerosis consecutive to degeneration of that tract produces in the centres of the cord during a number of years.

It is certain, gentlemen, that we cannot completely understand the rôle played by degeneration of the pyramidal tract in the production of contracture, until we are better acquainted with the precise functions of that tract.

As stated in the last lecture there is much difference of opinion as to the action of the fibres in the pyramidal tract. Are they *directly motor* fibres, the irritation of which has for its consequence the immediate contraction of one or more muscular fibres? This is but little probable. We know in fact that these fibres are not directly continuous with the peripheral nerves, which on the other hand are separated from them by cells belonging to the grey matter of the cord. It seems therefore most probable that *the pyramidal tract merely acts upon the grey matter of the cord.*

Most authors believe this action to be *excito-motor*, and such an opinion will be easily understood if the ingenious comparison suggested by Charcot in his lectures be mentioned to you. The mechanism of musical boxes is so far understood that it is known that by pressing a certain button, which corresponds to one or other of the airs named upon the lid, the cylinder is made to work, and the instrument immediately plays the air indicated. In the human organism the spinal cord may be looked upon as a musical box such as the one just mentioned. On account of education, on account of the local and special memory with which it is endowed, each part of the cord holds itself in readiness to execute such or such a movement, it might

be said, to play such or such an air. A wish exists to walk: the pyramidal tract produces whatever change in the cord is necessary, in order that the centres of walking may act; the lower limbs then execute the required movements, under the control, be it understood, of the higher centres which watch over the direction—the maintenance of equilibrium, &c.

In a word, it is the *motor impulse* excited in the centres of the grey matter by the fibres in the pyramidal tract which determined the movement, but these are not the fibres by which it was executed. If this very seductive theory is admitted, the contracture which occurs after hemiplegia must be looked upon as the result of a permanent impulse which sclerosis of the pyramidal tract, after degeneration of that part, excites in the grey matter of the cord.

This opinion has been most favourably received, and, as already observed, Vulpian and others support it. I myself find it very difficult to agree with them, since I can scarcely admit that a morbid process, such as that which produces sclerosis, of which the active period is limited, and the inflammatory effect can therefore be but temporary, would produce permanent spasm lasting until the death of the patient. Nor do I understand how such a contracture could exist in spastic paraplegia, when there is not degeneration but non-development of the pyramidal tract, and no morbid process therefore exists which resembles the sclerosis due to secondary degeneration.

Might it not be supposed that the function of the pyramidal tract is one of *inhibition*, analogous in a proportional degree to that of the vagus nerve upon the heart? If its action is considered to be of this nature the grey substance of the cord would still be the true motor power, but a power which is always under pressure, though at the same time ever ready to perform its functions (and in support of this suggestion, gentlemen, might not the rapidity, the suddenness with which the different reflex movements are made, be invoked?). The function of the pyramidal tract would be to exercise a restraining influence upon the cord and to prevent its ill-timed and unceasing action. If the will interferes to suspend temporarily this inhibiting effect upon the fibres of the tract which correspond to the centres of the grey substance, to the influence of which the contraction of a definite muscle is due, this muscle at once contracts in order

to produce the desired movement. If, on the other hand, this inhibiting action is permanently suspended owing to destruction of the fibres of the pyramidal tract, the cord being no longer under control, acts indefinitely. The muscular contraction due to its influence is similarly uninterrupted, and persistent spasm occurs. I would remind you that this, gentlemen, is but a hypothesis, but one which, as I think, accords with clinical observation, and it is on this account that I lay it before you.

Thus far, gentlemen, we have considered facts in their greatest simplicity; not rarely, however, they are slightly more complicated.

Fig. 28.—Hemiplegia of the right side of a female, accompanied by contracture, not only in the flexor muscles of the right leg, but also in those upon the "sound" side. (After Brissaud.)

I allude especially to the cases in which (A) secondary degeneration after a cerebral lesion occurs in the pyramidal tract on both sides of the cord, and (B) secondary degeneration is accompanied by more or less pronounced muscular atrophy.

A. *Bilateral degeneration of the pyramidal tracts consecutive to a unilateral cerebral lesion.*—Westphal was, it appears, the first whose attention was drawn to the existence of such degeneration, since, in 1875, he observed that in some cases of

hemiplegia there was foot clonus in the leg of the healthy side. In 1878 Dejerine made the same observation, and in the following year Brissaud called especial attention to contracture in the two lower extremities, which is sometimes found to occur in hemiplegia. At the same time it is only fair to say that this subject was specially studied by Pitres, both from an anatomical point of view, by himself personally (1882—1883), and from a clinical point of view by the researches which he inspired in his pupil Dignat (1883—1884).

I shall borrow many statements from these two authors upon this interesting question. Let us first consider what we learn from clinical observation, and let us suppose a hemiplegic patient suffering from contracture, due to secondary degeneration of the pyramidal tract. The attitude of the limbs on the side corresponding to that of the degeneration, gentlemen, is known to you, the upper limb being fixed in a position of flexion and pronation, the lower limb in one of extension and adduction, whilst reflex action is very excessive in both limbs. The "sound" side is by no means unaffected, as one might *à priori* suppose. Dignat, in fact, has on the contrary shown that if the condition of the limb upon the "sound" side is examined with care the muscular force in the lower limbs is found to be diminished by about 50 per cent., and proportionately more as the time of examination is nearer to that of the occurrence of hemiplegia. In addition to this, the side presents evidence of functional loss of power, as if the memory necessary for the co-ordination of movement on the "sound" side was lost. Thus the movements required in walking are often difficult or impossible upon this side, although the muscular power needed for their execution is amply sufficient. Decided increase of the knee-jerk upon the "sound" side (already noted by Brissaud), and at times foot clonus also occur. Lastly, when the tendency to spasm is still more pronounced, as Brissaud has remarked, hemiplegia may be accompanied by *contracture in the two lower extremities.*

The *upper extremity* on the "sound" side is somewhat similarly affected. Though it is not in a condition of rigidity, and hand clonus never occurs, the tendon reflexes are more or less excessive. The muscular force is diminished by about 38 per cent. (Dignat), this diminution being greater in proportion as the time of the observation is nearer to the occurrence of the

hemiplegia. As regards this point Friedlander made an interesting remark in comparing the upper extremities with each other. According to his observations the diminution in the muscular power of the left upper extremity in right hemiplegia is greater than that of the right upper extremity in left hemiplegia.

Such, from a clinical point of view, is the participation of the limbs upon the "sound" side. The anatomical cause of the symptom was shown to us by the investigations of Pitres. This author collected 10 cases of degeneration of the two crossed pyramidal tracts, due to a unilateral cerebral lesion. Minute examination of these cases led him to conclusions which I shall ask your permission to mention. In *six* cases the lesion of the two crossed pyramidal tracts (FPyC) was equally developed, that is to say, the degeneration of the crossed pyramidal tract (FPyC) in the cord upon the same side as the cerebral lesion, was as pronounced as that of the corresponding tract (FPyC) upon the opposite side. In these cases the sclerotic area was slightly more extensive than when one side is alone affected, while on the other hand the sclerosis was more moderate in degree, as if a smaller number of fibres was affected than when the sclerosis is unilateral.

In the *four* other cords the sclerosis was decidedly more pro-

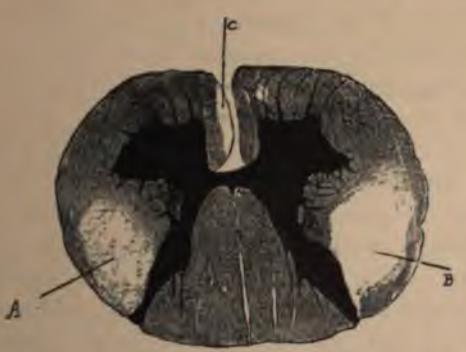

Fig. 29.—Section of the cord (cervical enlargement) in a case of hemiplegia from a lesion in the left hemisphere. The secondary degeneration involves the direct pyramidal tract (C), and the crossed pyramidal tract (B), besides the crossed pyramidal tract (A) upon the healthy side.

nounced in the crossed pyramidal tract (FPyC) upon the side

opposite to the cerebral lesion, than in the FPyC upon the same side.

The direct pyramidal tract (FPyD) in the first *six* of the cases just mentioned was affected as follows:—In *two* cases the tract (FPyD) was quite unaffected upon both sides; in the *fifth* case it was unequally affected upon the two sides, while in the *sixth* case the tract of degeneration only occurred upon one side.

As regards the *four* cords in which the crossed pyramidal tract (FPyC) on the same side as the cerebral lesion was less affected than that (FPyC) upon the opposite side, in *two* the direct pyramidal tract (FPyD) was in no way involved. In the *third* bilateral but unequal degeneration existed in the two direct pyramidal tracts (FPyD). Lastly, in the *fourth*, degeneration only occurred in the direct pyramidal tract (FPyD) upon one side alone, namely, upon that in which the crossed pyramidal tract (FPyC) was most affected.

I have quoted the results of these ten autopsies at length, since they are the most interesting illustrations of the anomalies which occur in the distribution of the fibres of the pyramidal tract.*

More than one explanation has been given of bilateral degeneration of the crossed pyramidal tract (FPyC) after a lesion in one cerebral hemisphere alone.

Charcot has expressed the opinion that the fibres of one crossed pyramidal tract (FPyC) passed to the same tract (FPyC) upon the opposite side by the anterior commissure, and the fibres might thus degenerate equally well upon either side of the cord.

According to Hallopeau the mechanism of the lesions is different. In his opinion, where the decussation of the pyramids occurs and the fibres of these tracts are closely interwoven, the inflammation of the degenerating crossed pyramidal tract (FPyC) would extend by contiguity to the corresponding tract on the opposite side. Thus the fibres of this tract would be equally involved in the degeneration. This theory, otherwise ingenious, is difficult to accept, since as the knowledge of the pathological anatomy of the nervous system becomes greater, the more con-

* Sherrington has observed in the dog and reproduced in the figures of his memoir: "On Secondary and Tertiary Degenerations in the spinal cord of the Dog" (Journ. of Physiology, Vol. VI., No. 4), a degeneration of the two crossed pyramidal tracts, after a lesion in one hemisphere alone.

vinced one becomes that secondary degeneration, properly so called, has little tendency to produce diffused inflammation around it such as that which Hallopeau believes to exist.

The opinion of Pitres seems to me the true one. These facts are considered by him as fresh proof of the extreme irregularity in the distribution of the pyramidal tracts and of the close connections which exist between the tracts on the two sides. These connections might *à priori* be believed to exist on account of the synchronism and association of the movements which occur in health.

The existence of bilateral degeneration of the pyramidal tract completely explains the troubles which occur on the "sound" side in hemiplegia; namely, decrease of muscular force, loss of power to co-ordinate certain movements, excess of the tendon reflexes, foot clonus, &c.

I would make one observation before closing these remarks upon degeneration of the pyramidal tract. It has been stated by me that the upper extremity on the "sound" side never suffers from contracture, which on the other hand is frequently the case with the lower extremity. There seems to be an anatomical reason for this fact. The upper extremities are accustomed to act in an isolated way and separately from each other, whereas in standing or walking, which are the real functions of the lower extremities, the latter always move and work simultaneously. It is thus explained why the bilateral distribution of the pyramidal tracts would not produce the same effects upon the upper and lower limbs, and why the symptoms due to degeneration of these tracts would be different in the two cases.

B. *Association of secondary degeneration of the pyramidal tracts with amyotrophy.*—This subject, gentlemen, will be considered, although the connection of amyotrophy with secondary degeneration of the pyramidal tract is not equally well recognized in the different hypotheses suggested to explain its existence.

One thing is certain, that in some cases hemiplegia is followed by pronounced muscular atrophy in the paralysed limbs. This amyotrophy is specially marked in the small muscles of the hand, the thenar and hypothenar eminences being flattened, the interosseous spaces hollowed, and although the muscular atrophy is less pronounced than in the hand called that of Duchenne-Aran the appearance of the extremity is quite characteristic. The

muscles of the fore arm, and even of the arm and lower extremity, may be similarly affected. It should be noted that this is true amyotrophy, affecting some muscles in preference to others, and not the simple condition of general muscular wasting, which is so frequently observed in the paralysed limbs when the hemiplegia is of long duration.

Different opinions have been held, gentlemen, as regards the anatomical cause of this muscular atrophy.

Charcot, who was one of the first to consider this condition, recognized in one case, which was soon followed by several others, that a characteristic lesion existed in the cells of the anterior cornua, and expressed the opinion that this lesion was consecutive to degeneration of the pyramidal fibres and due to

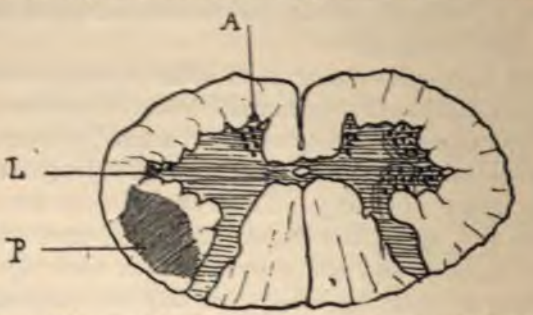

Fig. 30.—Section of the cord (cervical region) in a case of hemiplegia on the left side associated with amyotrophy. (After Pitres.) In the left anterior cornu (the side of the degeneration in the crossed pyramidal tract (P)) some cells alone of the anterior group (A) and the lateral group (L) are unaffected, the others being all destroyed by atrophy.

the fact that inflammation of those fibres extended to the grey matter. This explanation was generally accepted, and confirmatory facts had been published by Carrière, Hallopeau, Pierret, Pitres, Brissaud, &c.,* when within the last few years Babinski, in publishing the account of an autopsy in a case of this kind, stated that no lesion was found, either in the cornua of the spinal cord or the peripheral nerves. Shortly afterwards Quincke observed a case of the same kind, as did other authors, viz., Roth, Muratow, Darkschewitsch, &c. On the other hand Dejerine found in four cases of atrophy, after hemiplegia,

* During the publication of these lectures Joffroi and Achard published a work containing new facts and an interesting theory as regards the occurrence of this condition. (Arch. de méd. experim., Nov. 1, 1891.)

peripheral neuritis without any lesion of the cornua or anterior roots. On account of the frequency of peripheral neuritis these cases may, in all probability, be classed with the preceding.

Owing to the above facts one thing may be considered certain, viz., that degeneration of the pyramids of the cord almost constantly produces, at any rate after a sufficient length of time, marked diminution in the size of the anterior cornua upon the same side. Fürstner and Knoblauch called attention to this fact, and showed that it is in the lateral horn that this diminution in size is specially pronounced. It can be easily conceived that in some conditions this alteration in the grey matter may extend to the cells of the anterior cornua and produce the lesions described by Charcot. The so-called "tertiary" degeneration mentioned by Sherrington and Langley may be justly regarded as confirming this idea. On the other hand there seems no reason to doubt the results obtained by the authors previously named, by whom no lesion was found in the grey substance of the anterior horns. Perhaps these facts, as Borgherini believes, are due to the trophic effects of the brain upon the muscular system. This explanation is also confirmed by the cases in which muscular atrophy occurs rapidly after the production of a cerebral lesion. This, it is true, is a mere supposition, but it is one which is by no means inadmissible when it is remembered, gentlemen, that the most pronounced muscular atrophy is at times observed in certain hysterical patients, of which the cause is in all probability some trophic disturbance of cerebral origin.

LECTURE III.

DESCENDING DEGENERATION CONSECUTIVE TO TRANSVERSE LESIONS OF THE CORD.

A.—In the antero-lateral column:—α. *Pyramidal tract*, its degeneration in a case of transverse lesion of the cord is far more complete than when it is due to a cerebral lesion. Theories proposed in explanation of this fact. β. *Portion of the lateral column external to the pyramidal tract:* fibres of the intermediate tract of the lateral column. γ. *Anterior column:* fibres of the marginal tract. Nature, path, and origin of these fibres: commissural fibres, commissural cells. Tract of descending degeneration of cerebellar origin (Marchi).

B.—In the posterior column:—Comma-shaped degeneration of Schultze: small number of confirming facts. Suppositions as to the nature of the fibres affected by this degeneration.

Gentlemen,—In the preceding lecture the degeneration of the pyramidal tract which follows cerebral lesions was studied. The secondary degeneration which follows a lesion seated in the spinal cord will now be considered.

In order that the subject may be more clear it will be supposed that the cord has been almost completely divided by a transverse lesion.

As you know, gentlemen, in such a lesion a general, and more or less total destruction of the anatomical elements of the cord exists to the extent of four, five, six millimetres ($\frac{1}{6}$, $\frac{1}{5}$, $\frac{1}{4}$ inch) or more in height, according to the mode in which the lesion has been produced. In this zone of total destruction, "the zone of traumatic degeneration" of Schiefferdecker, the way in which each tract is affected cannot naturally be studied. It is only in a lower portion of the cord, when the lesions are sufficiently localized, that their distribution can be advantageously considered.

A.—Antero-lateral Column.

This column being composed of afferent and efferent fibres, the degeneration of many distinct tracts within it, both in the ascending and descending direction, must be described.

a. Pyramidal tract.—The fibres of this tract have (in all probability) their trophic centre in the cerebral cortex. It is therefore most probable that if, owing to some cause, they are divided in any part of their path in the cord, the portion seated below the point of division will be undoubtedly affected by secondary degeneration. This, in fact, has been already shown to occur when the continuance of these fibres is interrupted by a lesion in their path through the brain. At the same time, degeneration of the pyramidal tract has a special character according as a lesion in the brain or cord is the cause of its existence. Bouchard first remarked that when the initial lesion

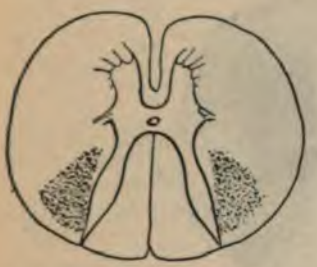

Fig. 31.—Degeneration of the crossed pyramidal tract following a lesion of the cerebral hemispheres (diagrammatic).

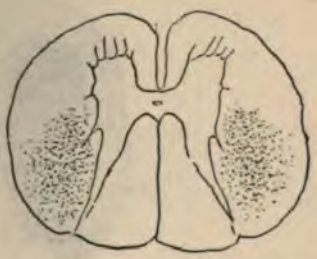

Fig. 32.—Degeneration of the lateral column following a transverse lesion of the cord a few centimetres above the region which is here represented (diagrammatic). It will be observed that the extent of the degeneration is notably greater than in the preceding figure.

is in the cord the secondary degeneration of the pyramidal tract is notably more extensive than when it is seated in the brain. This statement is absolutely true, having been confirmed by all observers, although the explanations given of this difference vary. These explanations may be connected with one of the two following opinions:—

a. A more considerable portion of the cord is affected by degeneration, because not only the pyramidal fibres but also those belonging to other tracts are involved (the commissural fibres of Bouchard connecting points of the grey substance which are at a more or less different height in the cord); these latter fibres being intimately mixed with those of the pyramidal tract, and degenerating at the same time necessarily render the diseased part more extensive.

b. The affected part is of greater size because the pyramidal fibres being united in a much more confined tract in the cord than in the brain, in the latter they are much less often injured throughout than in the former, and consequently the degeneration in the first case is less extensive than in the second.

Though disinclined, as a rule, to eclectic solutions, I must acknowledge, gentlemen, that these two opinions seem to me equally probable; the fibres added to those of the pyramidal tract will be again discussed at a future time.

Fig. 33.—Section of the cord (lumbar region) from a case of transverse lesion seated on the dorsal region. Degeneration of the two crossed pyramidal tracts (semi-diagrammatic).

So far, the whole pyramidal tract has been considered; it need scarcely be added that degeneration may not only affect the fibres of the *crossed*, but also those of the *direct pyramidal tract*, the condition being that the transverse lesion is at such a height in the cord that the fibres of the latter tract have not as yet ceased to exist.

β. Portion of the antero-lateral column external to the pyramidal tract.—As already observed, gentlemen, this portion of the cord contains nerve fibres belonging to other tracts. Most of these are affected by ascending, but some by descending degeneration. The latter are, for the most part, anterior to the pyramidal tract in the middle portion of the antero-lateral column, and Löwenthal describes descending degeneration of these fibres which he terms the *intermediate tract of the lateral column.*

These specially occupy the middle part of the lateral column on the inner side of the tract of Gowers, and the direct cerebellar

tract, a certain number of fibres seeming to join these tracts internally. It is very probable that some fibres also join

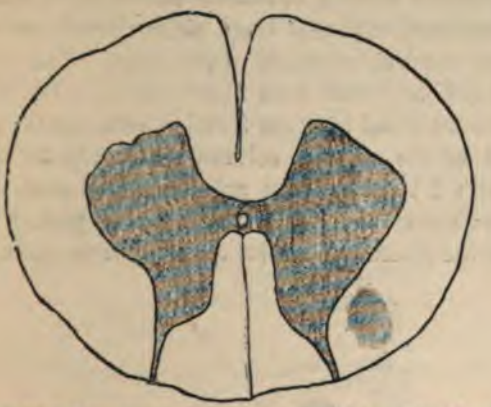

Fig. 34.—Descending degeneration of the pyramidal tract in the cord of a dog after removal of one of the cerebral hemispheres. (After Singer and Münzer.) It will be remarked how much the extent of the degeneration due to the cerebral lesion differs from that which occurs (fig. 33) after a transverse lesion of the cord.

those of the crossed pyramidal tract. At this point, in fact, nerve fibres are found which differ much in size.

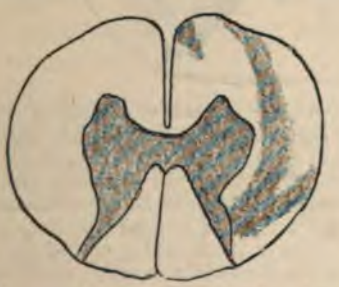

Fig. 35.—Descending degeneration in the cord of a dog after division of the antero-lateral column upon the same side, in the upper cervical region. (After Singer and Münzer.) Two regions are seen to be affected by degeneration (indicated by dots) in the right antero-lateral column; the smallest at the anterior angle of the cord is the *descending sulco-marginal tract*,* while the larger is in the *intermediate tract of the lateral column*.† The pyramidal fibres are included in this degenerated part.

γ. Anterior column.—In this column also certain fibres are affected by descending degeneration after transverse lesions of

* *Vide* p. 34 (Translator). † *Vide* p. 32 (Translator).

the cord in opposition to the opinion of Schiefferdecker, who thought that these were stray fibres of the pyramidal tract, whereas it seems certain (Flechsig and Singer) that they are quite unconnected with this tract, since they do not degenerate after lesions which are confined to the brain. This is the degeneration of the *marginal tract* (Löwenthal). The degenerated fibres of this marginal tract are found in man and in the monkey in the part of the anterior column which adjoins the anterior fissure, which I shall call the *sulco-marginal zone*. To distinguish these fibres from others in the same part, I propose to name them the *descending system of the sulco-marginal zone*.

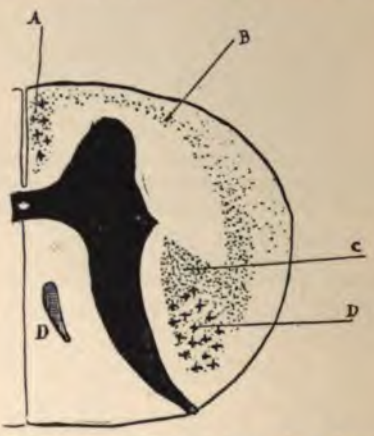

Fig. 36.—Diagram of the principal seats of descending degeneration in cases of transverse lesion of the cord in the dorsal region. A, Sulco-marginal region, in which two kinds of fibres are found: firstly, those of the *direct pyramidal tract* (+ + + +); secondly, those of the *descending system of the sulco-marginal zone* (. . . .). B, C, *Intermediate tract of the lateral column*; some fibres of this tract are found at the peripheral part of the cord intimately mixed with those of the tract of Gowers. D, Zone of the crossed pyramidal tract. This zone contains two sorts of fibres: firstly, those of the *crossed pyramidal tract* (+ + +); secondly, those of the *intermediate tract of the lateral column* (. . . .), which are intimately mixed with the preceding fibres but in smaller number.

The path and origin of these fibres of the anterior and lateral columns, which are independent of the pyramidal tract, will now be considered.

With regard to the former, it may be asserted that they almost undoubtedly pass into the grey substance of the cord, and very probably into its anterior and middle part. Some of these fibres seem to have an exceedingly long course, since traces

of their degeneration are found in the lower part of the cord, even where the fibres of the direct pyramidal tract have ceased to exist. It is, on the other hand, very difficult to trace their origin with precision. It seems very probable that some of these fibres which are liable to descending degeneration belong to the system of *longitudinal commissural fibres*, of which I have already spoken,* and which, taking origin from a cell in the grey substance, return to the grey matter seated in a lower part of the cord, after passing for a longer or shorter distance through the antero-lateral column. Since the time when the existence of such fibres was suggested by Bouchard they have been definitely proved to exist. Thus, for example, Ramon y Cajal, whose beautiful works upon the minute anatomy of the central nervous system are of scientific value, describes *commissural cells* seated, as Golgi had already shown, in many points of the grey matter.

From these, which are smaller than the motor cells, an axis cylinder passes through the anterior commissure into the anterior column of the opposite side. In this it divides into two branches, one ascending, the other descending, from which numerous branches proceed in the direction of the grey matter, dividing there into numerous fibrils, which form an extensive network round the cells, which are seated therein.

During their path through the white substance collateral branches form. After this description it is useless, I think, to add that the descending branch is involved in descending degeneration, whenever the transverse lesion is seated below the commissural cell from which it took origin; the ascending branch, on the other hand, is affected when ascending degeneration occurs.

It is doubtful, however, whether such a statement can be made with regard to all the fibres, and an opinion recently expressed by Marchi† should at any rate be mentioned. This author, having removed the whole or part of the cerebellum, studied the subsequent degeneration in the cerebellar peduncles. He was able to trace such degeneration in the inferior cerebellar peduncle, and found that at about the height of the olivary

* p. 31.

† Marchi, *Origine e decorso dei peduncoli cerebellari* (*Rivista sperimentale di Freniatria e Med. leg.*, Vol. XVII., p. 367).

body, the posterior longitudinal tract joined the band of Reil,* the conjoint mass of fibres passing into the antero-lateral columns. In these a somewhat large number of degenerated fibres existed, forming a mass of some size in front of the crossed

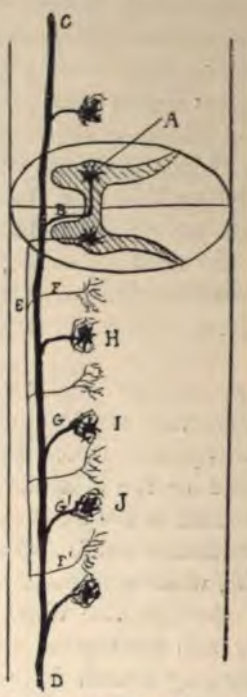

Fig. 37.—Diagram representing the longitudinal aspect of the cord as seen through a transparent medium. A, Commissural cell; B, Prolongation of that cell, passing through the anterior commissure, and dividing into an ascending (BC) and descending branch (BD); GG', divisions of the descending branch, forming branchlets round the nerve-cells of the grey matter. E, Prolongation of the same kind as B, but not passing to the other side through the anterior commissure; from this prolongation branchlets (FF') are given off, analogous to those (GG') which are connected with the descending branch (BD), and which are also divided into branchlets surrounding the nerve cells of the grey matter. These cells are not represented, that the figure may not be unnecessarily complicated. (Diagram after a figure of Ramon y Cajal.)

pyramidal tract, and extending also into other parts of the antero-lateral and anterior columns, with a pronounced tendency to occupy the peripheral parts of the cord. According to Marchi, therefore, fibres exist in the antero-lateral columns, of

* *Vide* p. 45 (Translator).

which the trophic centre is seated in the cerebellum, so that when a transverse lesion of the cord occurs these fibres would be affected by descending degeneration. Their seat in the cord exactly corresponds to the regions in which degeneration has just been studied by us, and which are termed the *intermediate tract of the lateral column* and the *descending sulco-marginal tract.*

Such, gentlemen, may be said with regard to the anterior and antero-lateral columns, but it must not be supposed that descending degeneration after transverse lesions of the cord is

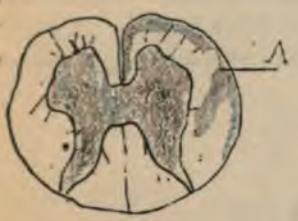

Fig. 38.

Fig. 39.

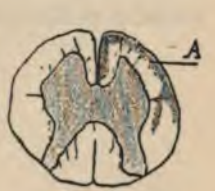

Fig. 40.

Section of the cord of a monkey (?), from which the right half of the cerebellum was removed (after Marchi). Fig. 38, Lumbar region; Fig. 39, Dorsal region; Fig. 40, Cervical region. It will be remarked that the zone of degeneration (indicated by points), which occupies in the three figures the periphery of the antero-lateral column, just extends within the region of the pyramidal tract, and encroaches slightly upon the anterior part of the direct cerebellar tract. In the original figure some points of degeneration also exist in the anterior roots, which are here omitted.

limited to these columns, as it may occur also in the *posterior columns.*

B.—The Posterior Column.

It is to Schultze that we specially owe the knowledge that secondary degeneration occurs in this part, although in his work (*Archiv für Psychiatrie*, 1883) that author points out that similar facts had been previously observed by Westphal, Kahler and Pick, and Strumpell. It is none the less true, however, that attention was specially drawn by his observations to this form of degeneration. The case to which allusion was made in these initiatory remarks was one of compression with complete division of the cord in the middle part of the cervical enlargement. At a distance of 2 or 3 centimetres ($\frac{3}{4}$ inch to 1 inch)

below the point where the compression existed, degeneration was found to exist in the region of the columns of Burdach, in the form of a line parallel in direction to the posterior horns, commencing at a short distance from the posterior commissure but not reaching the surface of the cord, from which, on the contrary, it remained at some distance. Owing to its form this degeneration may very justly be termed the *comma-shaped degeneration of the posterior columns.*

It should be added, gentlemen, that the cases in which this comma-shaped form of degeneration has been recognized are by no means numerous, although Schultze states that he has observed it three or four times. On the other hand, in a memoir recently published, Barbacci* states that he found degeneration

Fig. 41.—Section of the cord (upper dorsal region) 2 centimetres below a focus of compression, seated in the middle part of the cervical enlargement. (After Schultze.) A, Degenerated pyramidal tract; B, Comma-shaped degeneration in the posterior columns. The original figure indicated a small islet of degeneration in the antero-lateral columns which is omitted here.

in the posterior column after a transverse lesion of the cord, but that this degeneration was by no means comma shaped. Fibres were affected throughout the whole breadth of the posterior columns; lower in the cord these fibres were grouped near the posterior median fissure, and lastly in the conus medullaris occupied the posterior part of the cone. This form of degeneration could therefore be traced to a much lower point than that indicated by Schultze.

These facts require, not to be confirmed, as there is no doubt of their existence, but to be explained, and the best explanation would be that exactly indicating which tract is involved in the degeneration. This is as yet unknown, so that the uncertainty

* O. Barbacci, *Contributo anatomico e sperimentale allo studio delle degenerazioni secondarie, &c.* (*Lo Sperimentale*, 1891, parts III. and IV., pp. 335 and 406).

still exists. According to Schultze the fibres whose alteration produces comma-shaped degeneration are the descending branches of the posterior roots which enter the cord at the level of the transverse lesion. In opposition to this idea Tooth, not unreasonably, suggests that this comma-shaped degeneration is not observed to occur after division of the posterior roots, in addition to which, this degeneration, after a transverse lesion of the cord, descends lower than the descending fibres of the posterior roots are anatomically shown to do. Thus, according to Tooth, the degeneration would rather be due to destruction

Fig. 42.

Fig. 43.

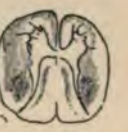

Fig. 44.

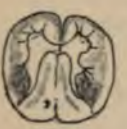

Fig. 45.

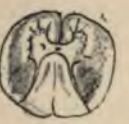

Fig. 46.

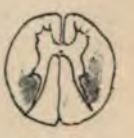

Fig. 47.

Sections of the cord from a case of fracture of the spine with destruction of the cord between the 8th cervical and 1st dorsal pair. (After Tooth.)

Fig. 42.—Transverse lesion; the destruction is almost complete.

Fig. 43.—1st dorsal pair; the greater part of the white columns is affected. The zone of traumatic degeneration exists here also.

Fig. 44.—2nd dorsal pair; in the antero-lateral column the region of the direct cerebellar tract and the tract of Gowers is already almost free from degeneration. In the posterior column the "comma-shaped degeneration" of Schultze is most clearly seen.

Fig. 45.—3rd dorsal pair; the same observations may be made.

Fig 46.—6th dorsal pair; the degeneration is confined to the crossed pyramidal tract, the intermediate tract of the lateral column, the descending sulco-marginal tract, and the direct pyramidal tract.

Fig. 47.—7th dorsal pair; the degeneration of the pyramidal tract is still most clear, while that of the intermediate and even more that of the sulco-marginal tract are far less apparent.

of the commissural fibres. Possibly, and this is a simple suggestion on my part, the *condition of the grey matter in the cord* should be considered, and in future it will be observed whether this comma-shaped degeneration only occurs when the transverse lesion involves to a greater or less extent the grey substance of the cord. If so, this would be a fresh argument in

favour of the theory that the commissural fibres are affected in this form of degeneration.

With regard to the lesions which exist in tabes I shall show you that in this disease the fibres which correspond to the "comma-shaped" tract remain unaffected for a long time after the other fibres in the posterior column are more or less involved.

As regards secondary descending degeneration within the GREY MATTER OF THE CORD nothing certain is known, although it is probable that amongst the tracts of fibres which compose it some are as liable to degenerate in the descending as in the ascending direction. Everything, however, points to the fact that such degeneration extends but a very short distance along the cord.

LECTURE IV.

ASCENDING DEGENERATION CONSECUTIVE TO LESIONS OF THE NERVE-ROOTS.

Anatomical study of the *afferent system of the cord proceeding from the fibres of the posterior roots.* Development of this system; origin in the external layer of the blastoderm; division of the neural plate into a central segment which will form the motor portions of the cerebro-spinal axis, and two lateral segments from which the *sympathetic nerve* and *ganglia of the spinal cord* will be formed.

Fibres of the posterior roots. Researches of Singer and Münzer. These fibres may be classed in three groups, according to the portion of the spinal cord in which they end. Their terminal nuclei.

Degeneration of the posterior columns after lesions of the posterior roots; researches of Singer, Tooth and Hartley; clinical observations of Kahler, Schultze, &c.

Gentlemen,—Before commencing to study ascending degeneration in the cord it will be useful to review the anatomical disposition of the *intramedullary afferent system proceeding from the posterior roots.* That this description may be more easily understood, the posterior roots will be themselves considered, which constitute, so to speak, the affluent branches of that system.

In this exposition the names of numerous distinguished neurologists or anatomists must be mentioned, who first acquired the knowledge which we possess upon this subject. Of these I would specially mention Singer, His, Ramon y Cajal, Edinger, Lenhossek, Kahler, Lissauer, &c., as those whose works are of special interest in connection with this subject.

Without entering into very complex details in reference to the development of the cerebro-spinal axis, I would merely remind you of the principal facts connected with its development which are specially applicable to the subject which now occupies our attention.

As His has shown, the nervous system derives its origin from the *external layer* of the blastoderm of which it forms at first a longitudinal thickening. This *neural plate* (*Neuralplatte*)

becomes divided longitudinally into three segments, from one of which, the *median*, the central nervous system is developed, and notably the *motor portions* (the motor cells of the cord with their prolongations which subsequently form the anterior roots and motor nerves).

On each side a *lateral segment* is found, which separates from the median segment representing the cord and gives origin to groups of cells which afterwards constitute the *sympathetic system* and *ganglia* connected with the spinal nerves.

At a later period the groups of cells which form the spinal ganglia become spindle shaped, and a nerve fibre is connected with each of their extremities. One of these fibres constitutes a *posterior root fibre* and approaches the cord into which it penetrates, and to which it therefore only belongs in a secondary way; while the other passing outward in an opposite direction constitutes a *peripheral sensory nerve fibre*. After this brief description, gentlemen, one fact will be easily understood, namely, that the posterior roots in no way belong to the spinal cord, being merely extensions from the spinal ganglia, while these ganglia constitute in connection with the system of conducting sensory fibres (peripheral nerves and posterior roots) a true trophic centre.

The development of the cord shows that the posterior roots enter the cord from without inwards, and the study of their path in the cord is in reality merely that of the composition of the posterior columns of the cord.

In each posterior root three varieties of fibres can be distinguished, according to the length of their path before entering the grey matter. These varieties have been recognized to exist by different authors, and were very accurately described by Singer and Münzer.

The fibres of the *first group* (*short fibres*) radiate at once into the grey substance of the posterior horn, some penetrating directly into the peripheral extremity of that horn, with which they appear to join, while the others approach the internal portion of that horn after having made a slight curve within the posterior column.

The fibres of the *second group* (*fibres of medium length*) ascend to a certain height in the posterior column, and as they pass onwards are seated more and more internally, so that their

general direction is oblique upwards and inwards. As, however, this group ascends, each of the fibres composing it curves outwards and passes into the grey substance of the posterior cornu, which it joins, not at its point, but more anteriorly, and to a special extent where the columns of Clarke are seated. This group thus becomes smaller and smaller as

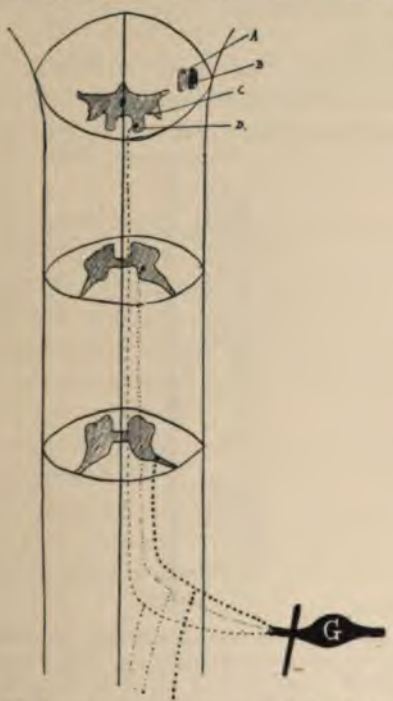

Fig. 48.—Diagram of the path and termination of the posterior root fibres; the black fusiform patch G, placed below and upon the right side, represents a spinal ganglion, the line which crosses it indicating the point of section of the posterior root which issues from it. This posterior root divides into three branches; each of these gives descending branches: + + + + short fibres passing to the apex of the posterior cornu; .... fibres of medium length entering the base of the posterior horn after having ascended within the column of Burdach; ---- long fibres passing into the nucleus of the column of Goll (nucleus gracilis) D, after having ascended within the interior of that column. The nucleus of Burdach (nucleus cuneatus) is at C receiving the long fibres and those of medium length issuing from the roots in the cervical region.

higher portions of the cord are reached, and finally, when all the fibres composing it have entered the grey substance of the posterior cornu, totally disappears.

The fibres of the *third group* (*long fibres*) though least numerous are by far the longest, since instead of being lost in the grey matter of the cord they are prolonged as far as the medulla oblongata. When this part is reached they join the grey substance in a similar manner, and the points at which they do so are known by the name of the *nucleus of the column of Goll* (nucleus gracilis) and *nucleus of the column of Burdach* (nucleus cuneatus).

It should be observed that on account of the tendency which all the root fibres of the posterior columns have to pass inwards being thrust back as it were on the outer side by fibres from the

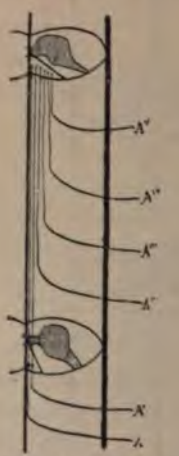

Fig. 49.—Diagram showing the constitution of the column of Goll. The fibres which constitute this column come from the posterior roots A, A', A'', A''', A^{IV}, A^{V}. In quite the lowest part of the cord the column of Goll therefore contains a smaller number of fibres than in the parts which are at a higher level. This increased number of fibres in the columns of Goll is not found to exist throughout the whole length of the cord, since, according to most authors, the roots in the lumbar region alone enter into the formation of this column.

new roots penetrating into the cord, those of the root fibres which have joined the cord at a lower level occupy the portion of the posterior columns which is closest to the posterior median fissure, constituting what are termed the *columns of Goll*. The long fibres of the third group coming from the roots which pass into the upper portion of the cord are external to these, and join the nucleus of the column of Burdach (nucleus cuneatus). At what height in the cord the root fibres cease to join the nucleus

of Goll, and are directed towards the nucleus of Burdach, even in the opinion of Singer and Münzer, is quite uncertain.

It should be observed that the columns of Goll are not universally believed to be entirely composed of long fibres coming from the lower segments of the cord alone, especially from the roots of the cauda equina. Thus, in the opinion of Barbacci, the division of the roots seated in the upper segments of the cord, especially of those in the cervical region, is followed by degeneration in some fibres of the column of Goll. However this is, it may be considered certain that by far the greatest number, if not all of the fibres which compose this column come from roots connected with the lower part of the cord.

Such, then, considered briefly, is the path of the fibres in the posterior roots. It may be said in addition that some of these fibres pass through the anterior commissure, join the antero-lateral column of the opposite side, and eventually reach the corresponding band of Reil* (Auerbach). Some fibres also pass through the posterior commissure into the posterior horn of the opposite side.

In a general way it may be said that all the fibres of the posterior roots pass eventually into the grey substance of the cord or medulla oblongata. According to recent observations (Golgi, Ramon y Cajal, Lenhossek, Kölliker) these fibres having reached the grey substance divide into numerous ramifications, which surround the nerve cells, but do not penetrate within them. The cellular groups in the grey substance are not the nuclei from which they originate (which, as we have seen, are constituted by the cells of the spinal ganglia) but *those in which they terminate* (*Endkerne*, His).

Both in the cord and medulla oblongata fresh fibres are given off from these terminal nuclei, which cross each other in both structures. Joining each other in the medulla oblongata, they constitute the plexus of fibres which is termed the *band of Reil** (*Schleife*, Germ.). As to the final termination of these fibres in the brain our knowledge is more limited. Some fibres of the band of Reil, according to Meynert, join the *corpora quadrigemina*, while some, according to other authors, pass into the *expansion of the lenticular nucleus*. More recently Flechsig

* *Vide* p. 36 (Translator).

and Hösel* have shown that a large number of fibres, specially of the middle portion of the band of Reil, pass into the *motor convolutions adjoining the fissure of Rolando.*

This summary description having been given, you will be able, gentlemen, to understand the path of that portion of the afferent system which proceeds from the ganglia of the spinal cord. The cells of these ganglia are *centres of origin*, while those in the grey substance of the cord or medulla oblongata are *centres of termination*, with the modification that no fibres penetrate into their interior, expansions being formed by the fibres which produce ramifications around the margins of the ganglia. On the other hand, these centres of termination become "centres of origin," and new fibres issue from them which cross each other and, passing into the band of Reil, reach the centres of the brain.

The course of these fibres is therefore by no means continuous, being interrupted in the grey substance of the cord and medulla oblongata, and in the terminal centres, so that it should be rather considered as a path divided into stages.

The subject which specially interests us at the present time, viz., the *secondary degeneration of the posterior roots in the spinal cord*, may now be considered, and it would be unfair to ignore the service which has been rendered by experiments upon animals.

In reality it is to them that we owe most of our knowledge about this form of degeneration. Singer, Tooth, and Horsley have obtained most interesting results which seem of the necessary precision, and their value is greater because they are entirely in accordance with the observations of human pathology, specially with those of Kahler, Schultze, &c.

If, for instance, as was done by Tooth, one of the posterior roots is divided in the cord of an animal such as the *monkey*, and a sufficient length of time (3, 4, or more weeks) is allowed to elapse before the animal is killed, a small tract of degeneration is found to exist in the posterior column of the cord above the lesion, the exact seat of which varies much according to its height in the cord. Immediately above the lesion this tract is in immediate contact with the inner margin of the posterior cornu against which its external border rests. Then when a fresh pair of

* Flechsig and Hosel, *Die Centralwindungen, ein Centralorgan der Hinterstränge* (*Neurologisches Centralblat*, 1890, p. 417).

nerve roots enters the cord, the degenerating tract being pressed inwards by the fibres of the newly arrived posterior root, becomes more distant from the cornu, and is entirely contained in the postero-lateral column or tract of Burdach. As a higher level is reached the tendency to pass inwards increases, and should the divided root belong to the lower part of the cord (the lumbar

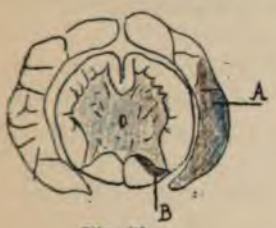

Fig. 50.

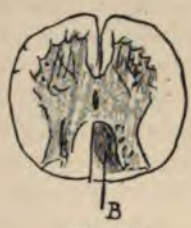

Fig. 51.

Fig. 52.

Fig. 53.

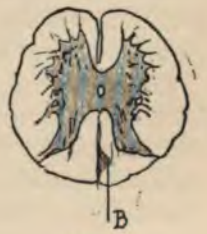

Fig. 54.

Sections of the cord from a monkey in which Horsley had divided all the posterior roots of the cauda equina at the distance of about 1 centimetre (⅖ in.) above the inferior extremity of the conus medullaris. (After Tooth.) Fig. 50: 5th lumbar; A, Section of roots which have undergone ascending degeneration, and which after being placed round the cord ascend along its surface until one after the other penetrates into its substance; B, Degeneration of the posterior column where it is in contact with the posterior cornu (entrance of the posterior roots into the cord, true external bandlets). Fig. 51: 3rd lumbar; the posterior column has undergone more degeneration (B) than at the level of the 5th lumbar, because at the latter point it had not yet received all the fibres of the posterior roots which had undergone ascending degeneration. The degenerated zone (B) commences at the level of the 3rd lumbar nerve to approach the middle line. Fig. 52: 11th dorsal; the degeneration (B) has completely left the posterior cornu in order to approach the posterior median fissure. Fig. 53: 8th dorsal. Fig. 54: 4th cervical; the degeneration (B) is seated in the posterior part of the column of Goll.

or sacral region) the degeneration occupying a more and more internal position, finally affects the postero-median column or that of Goll, and may then be followed as far as the medulla oblongata, where it is seated near the nucleus of the column of

Goll (nucleus gracilis). This statement is certainly more or less theoretical; it would be difficult to follow for such a distance the degeneration of a single root, since as a higher and higher level is reached the number of fibres lost in the grey substance is so great that at last the sclerosis almost entirely disappears. When, however, many of the posterior roots have been divided or injured the long fibres which degenerate are so numerous that they can be followed throughout their entire length.

It is unnecessary to describe at greater length the seat of secondary degeneration in these cases, the figures certainly indicating more with regard to this point than a long explanation would do. The one point upon which alone I would dwell is the following conclusion. When degeneration follows a lesion in the posterior roots the degeneration of the white substance is in the posterior columns, in which its seat is nearer to the posterior median fissure in proportion as the diseased root occupies a lower position.

The ascending branches of the posterior roots have as yet been alone considered, but it must be remembered that as they enter the cord a Y-shaped division of the posterior roots occurs, an *ascending* (which has alone been considered) and a *descending branch* being thus formed, both of which may undergo secondary degeneration. In the posterior columns not only *ascending*, but a certain degree of *descending degeneration* may occur after destruction of the descending branches which result from bifurcation of the divided posterior roots. These branches, however, being shorter, their degeneration is slighter in degree. Tooth, as already mentioned, in his observations upon division of the roots, states that he has never observed such degeneration to occur, while Schultze attributes the "comma-shaped" degeneration, described by him as occurring in the posterior columns, to the change in these fibres having a downward direction.

Tooth also observed a slight degree of degeneration in the posterior column *of the opposite side*, which may be attributed to destruction of those root fibres in which decussation had occurred.

The changes in the *grey matter* must also be considered while the secondary ascending forms of degeneration are being studied. When sufficient time has passed after division of the roots, the posterior, and even the anterior horn at its base, may be

found to be much diminished in size. The effect produced by division of the posterior roots in the cauda equina upon *Clarke's column* should also be specially noticed. It is most clearly seen in fact that after such division the fine *reticulum* of nerve fibres disappears throughout the whole section of Clarke's columns; whilst, however pronounced the alteration of these fibres may be, the *cells* of Clarke's columns remain in a perfectly healthy condition. This also shows, what has been already stated, that the fibres of the posterior roots terminate close to the cells of the grey matter, but without penetrating within them; thus the former may disappear without degeneration of the latter. These facts are well known, and have been mentioned by different authors, especially by Schultze, Lissauer, Mott, and others.

If I have thus dwelt upon the degeneration which follows lesions of the roots it is not, gentlemen, on account of its being frequently observed in cases of injury or compression of the cord, or cauda equina, which but rarely occur, but because in my opinion a very important part of the pathology of the spinal cord is intimately connected with lesions of the posterior roots, and I have therefore felt it my duty to consider them at some length.

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LECTURE V.

ASCENDING DEGENERATION AFTER TRANSVERSE LESIONS OF THE SPINAL CORD.

A. In the POSTERIOR COLUMN: α. In the *column of Burdach*. β. In the *column of Goll*. Difference in seat and extent of degeneration according to the height of the transverse lesion.

B. In the ANTERO-LATERAL COLUMN: α. *Direct cerebellar tract*, its description by Flechsig, its seat, form, origin, path, termination, and degeneration. β. *Tract of Gowers* (antero-lateral ascending tract of English authors), its seat, form, origin, path, termination, and degeneration.

C. In the GREY SUBSTANCE OF THE CORD. Degenerations in the grey matter are but little known, lesions found by Barbacci.

Sound condition of *lateral limiting layer* (*seitliche Grenzschicht* of German authors).

GENTLEMEN,—In the preceding lecture ascending degeneration of the cord after lesions of the posterior roots was considered, and the path of those roots in the cord and composition of the posterior column was carefully explained, although in a somewhat summary manner. The characters of ascending degeneration when it is secondary, not to a lesion of the roots, but to one of the cord itself, will now be discussed.

A. In the POSTERIOR COLUMN.—The fibres of this column having been just considered, it is unnecessary to enter into minute details. Whichever part of the cord is the seat of the transverse lesion, all the ascending fibres of the posterior columns in this part would evidently be totally destroyed to a distance of some millimetres (1 mm.=0·039 in.) above the lesion, this constituting the area of *traumatic degeneration* of Schiefferdecker.

But as this area is left, the effect of the lesion upon the long fibres, and those of medium length, which constitute the tracts of Goll and of Burdach, may be studied with advantage.

a. Column of Burdach.—This column, of which almost the whole at first undergoes degeneration, receives from each fresh posterior root as it enters the cord a contingent of sound fibres, which are placed at first along the inner side of the point of the posterior

horn, pressing the degenerated fibres inwards; thus the posterior and external portion of the column of Burdach is that from which the indications of ascending degeneration most quickly disappear. The anterior part of the same column, which adjoins the posterior commissure, remains altered over a somewhat longer portion of its path (usually over a distance corresponding to the entry of from two to four pairs of nerves into the cord); this is because the fibres which constitute the anterior part do not belong to the system of the posterior roots, but to that of the commissural fibres which bind together the grey substance at different heights; and the addition of such fibres appears to be less rapid and less extensive than that of the posterior root fibres. In short, the degeneration of the column of Burdach diminishes little by little, and disappears from without inwards, and from behind forwards, until completely ceasing in this tract, though still existing near the posterior median fissure in the region which has been termed the column of Goll (*vide* figs. 64, 65, 66).

β. Column of Goll.—The long fibres of the posterior roots which join together to form this tract pass to a certain point in the grey matter of the medulla oblongata, which is termed, as you know, gentlemen, the nucleus of the column of Goll (nucleus gracilis).

The greater part of, or all the fibres of this tract are derived, as you also know, from the posterior roots of the cauda equina and of the pairs of nerves connected with the lower part of the cord. Thus every transverse lesion of the cord which affects these fibres will be followed by their degeneration, which can be traced almost as far as the floor of the 4th ventricle.

At the same time the degeneration of the column of Goll must not be supposed to be always identically the same, gentlemen, whichever portion of the cord is divided. Thus, for instance, if this occurs quite at the level of the *conus terminalis* so as only to affect the inferior roots of the cauda equina the degeneration of the column of Goll which follows only affects its *posterior* and *median* part. This is the same as stating that the nerve fibres of the posterior columns are placed so much the nearer to the middle line and surface of the cord as they have the longer distance to go.

If, on the other hand, the cord is divided in its upper part, as,

for instance, in the lower part of the cervical region, the degeneration of the columns of Goll extends over a much larger area than in the preceding case, which is to be expected since a much larger number of long fibres are divided (*vide* fig. 49).

The difference which exists in the area of degeneration in this tract according to the height at which the cord is divided clearly indicates that the column of Goll is not of immutable size, as the inexperienced are inclined to think, but composed of united fibres each of which has its own individuality. Thus the column of Goll, as Schultze also believes, cannot be absolutely distinguished from that of Burdach, since during part of its path

Fig. 55.—Division of the cord (cervical region) in a case of ascending degeneration in the columns of Goll (A) due to compression in the dorsal region.

the fibres of the former tract pass within the territory of the latter.

In this respect the terms employed by some English authors may represent better what really occurs. They merely divide the posterior column into a *postero-lateral* and *postero-median* tract, so that the difference between these two columns is less decided than when they are termed the "column of Goll" and the "column of Burdach."

It is a mere question of terms, and if there is general agreement as to the facts of the case, it is unnecessary to dwell upon this point.

Such, gentlemen, is what I wished to say about ascending degeneration of the posterior column, but when transverse lesions of the cord exist, ascending degeneration is by no means

limited to the posterior columns, but occupies also, and over a considerable area, the antero-lateral columns.

B. In the ANTERO-LATERAL COLUMN two areas of degeneration may be distinguished, which are confused at certain points, but clearly separated in other portions of their path. One is that which corresponds to the direct cerebellar tract (tract of Flechsig), the other to the antero-lateral ascending tract (tract of Gowers).

a. Direct cerebellar tract.—The degeneration of this tract was observed by Türck, who traced it as far as the restiform body, but it is to Flechsig that we are really indebted for knowledge of the origin, path, and termination of the fibres which compose it. Schultze, Kahler and Pick have also studied the degeneration

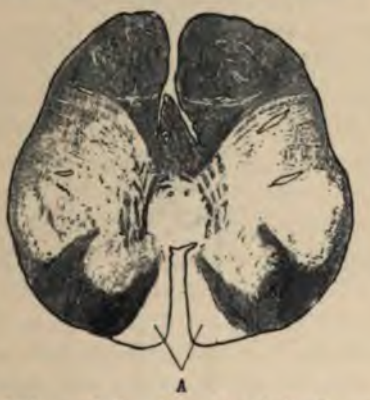

Fig. 56.—Division of the medulla oblongata at its lower part, in a case of ascending degeneration of the columns of Goll (A) due to compression of the dorsal region.

of this tract, and their works upon this subject are most interesting.

The direct cerebellar tract occupies the posterior half of the surface of the lateral column. It has the form of the segment of a ring representing about the sixth part of the circumference of the cord. Its posterior extremity (which is in relation with the posterior horn) is large and, as it were, puffed out, whilst the anterior extremity is more fine, so that for a certain distance it has the appearance of being drawn out. The anterior extremity of this tract is neither very clear nor certain, and it seems to fuse insensibly with the adjoining tracts.

Opinions differ as to the point in the cord at which this tract

can be first recognized. Some (Kahler and Pick) state that it is at the level of the *9th dorsal pair;* others (Schultze) believe it to be at the height of the *10th pair;* Tooth concluding from *his* observations that it is at the level of the 8th pair. Quite recently Barbacci states that he has observed degeneration of this tract when the lesion of the cord was seated at the level of the 11th and even of the 12th pair. I shall soon have occasion to consider this latter point, and when the origin of the fibres of this tract is better known to you the interpretation

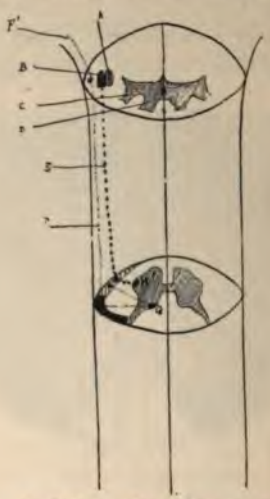

Fig. 57.—Longitudinal aspect of the cord and lower part of the medulla oblongata seen, as it were, through a transparent medium. This schematic figure shows the origin of the fibres of the direct cerebellar tract (F) and tract of Gowers (E). (The tract should have been prolonged beyond the ascending root of the fifth nerve (B), in which by an error of the designer it seems to end.) The fibres of the direct cerebellar tract issue from the cells of the columns of Clarke (G). The fibres of the tract of Gowers would issue on the one hand from the cells of the lateral horn (H), on the other from those of the columns of Clarke (G).

which may be given to this statement on the part of the Italian author will be considered.

The study of the direct cerebellar tract will now be continued by examining its path from beginning to end.

The size of this tract varies according to the height at which it is considered. It is thus smaller in the lower dorsal than in the cervical region. The fibres which constitute it ascend the whole length of the dorsal and cervical cord to the medulla

oblongata, being always seated behind an imaginary line dividing the cord and medulla oblongata transversely in its middle part. Gradually the posterior horn of grey matter leaves the surface of the cord, while the direct cerebellar tract remains there, but pressed backwards to a slight extent by the ascending root of the 5th nerve, which is placed in front of it; the tract is then contained in the restiform body, within which it passes into the cerebellum, having reached which it terminates, according to Flechsig, in the *superior worm*.

It must not be thought, gentlemen, that the fibres of the cord seated in the direct cerebellar tract all belong, on this account, to that column. It is, in fact, recognized that even when pronounced degeneration of the direct cerebellar tract occurs a small number of fibres remain unaffected in the midst of the degenerated area. It is certainly true that some of the unaffected fibres do belong to the direct cerebellar tract, and having joined it above the point at which the cord was divided, are, owing to that fact, in a sound condition; others, however, apparently belong to a different system, in which degeneration occurs in a descending direction. These are fibres which seem to form in part that tract of Marchi of which I have already had occasion to speak. You remember, gentlemen, that in the opinion of Marchi the origin of this tract is in the cerebellum, that it passes by the inferior cerebellar peduncle, and descending into the cord is dispersed through the antero-lateral column, and to a large extent near its surface.

What is the *origin* of the fibres of the direct cerebellar tract? According to what has been already remarked about the anatomy of the cord, it is probably to be found in a group of nerve cells, none other in fact than that termed the *cells of the columns of Clarke*.

The columns of Clarke will again be considered with regard to the pathological anatomy of tabes. I will merely observe that by this name are designated two important groups of cells, each of which is seated at the base of the corresponding posterior horn, and which are much developed in the dorsal region. The pyramidal prolongation of each cell passes obliquely across the lateral columns to the surface of the cord, where it seems to curve upwards and constitute one of the fibres of the direct cerebellar tract. According to some authors the cells of the

columns of Clarke are not the only origin of the fibres of the direct cerebellar tract, some of which (especially in the cervical region) seem to be directly connected with the posterior roots. This, however, is quite uncertain.

Whatever the truth may be, let us suppose, if you will, gentlemen, that the cells of the columns of Clarke constitute the sole origin of the direct cerebellar tract, and consider what the consequences will be as regards ascending secondary degeneration.

When the transverse lesion of the cord is seated at a position (dorsal region) in which the columns of Clarke exist, and the direct cerebellar tract is already formed, the degeneration which follows division of the fibres in that tract, and that due to

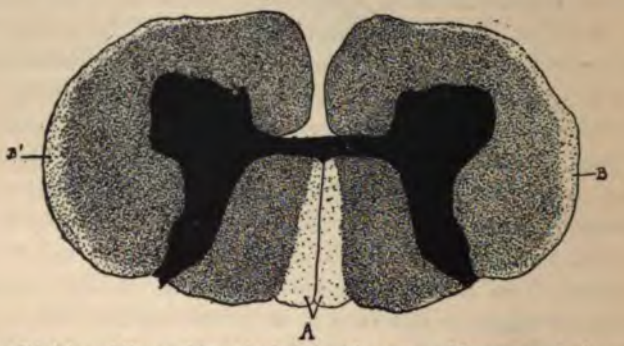

Fig. 58.—Division of the cord (cervical region) in a case of transverse lesion seated in the dorsal region. Ascending degeneration:—A, Columns of Goll; B, Direct cerebellar tract posteriorly, tract of Gowers anteriorly.

destruction of a more or less considerable portion of the columns of Clarke, may be confused. If, however, the transverse lesion is in a portion of the cord (quite the lowest part of the dorsal region) in which the direct cerebellar tract is not yet formed, although the column of Clarke is now beginning to appear, a lesion of the cells in that column is found to produce ascending degeneration in the fibres of the direct cerebellar tract, of which these cells are the trophic centre. This ascending degeneration only occurs at a certain height above the transverse lesion on account of the obliquity of these fibres. The facts indicated by Barbacci,* already mentioned, in which, after a transverse lesion at the level of the 10th or 12th dorsal nerve, degeneration of the direct cerebellar tract is only found to occur at some distance

* *Vide* p. 54 (Translator).

above the point where the zone of traumatic degeneration ceases may thus be explained.

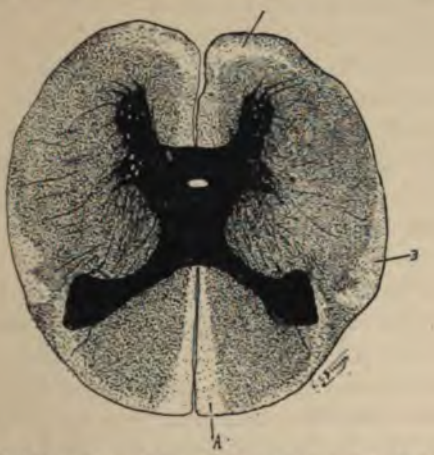

Fig 59.—Section of the spinal cord (upper part of the cervical region immediately below the decussation of the pyramids) in a case of transverse lesion of the cord in the dorsal region. A, Columns of Goll; B, Direct cerebellar tracts and tract of Gowers; C, Marginal tract of the anterior column. (Damaschino collection.)

Fig 60.

Fig. 61.

Fig. 62.

Fig. 63.

Sections of the medulla oblongata from a monkey in which a division had been made through half of the cord between the 7th and 8th cervical vertebræ (after Tooth). At the lower part of the medulla oblongata (fig. 60) the direct cerebellar tract (B) and the tract of Gowers (A) are in contact, and apparently confused together. As a higher and higher point in the medulla oblongata is reached, the two tracts tend to separate, and in fig. 63 are seen to be quite distinct from each other.

β. Tract of Gowers (antero-lateral ascending tract of English authors).—Although the altered part which resulted from degene-

nation of this tract had been previously seen and mentioned in the account of some autopsies, it may fairly be said that Gowers was the first who described it clearly and distinguished it from the change due to degeneration of the direct cerebellar tract with which it had been previously confused.

It is for this reason that I shall call this tract, as some other authors do, by the name of *the tract of Gowers*, a term which seems to me preferable to antero-lateral ascending tract, a name which some English authors employ, especially Tooth, in the interesting chapter which he devotes to the study of these fibres.

The tract of Gowers is seated at the surface of the cord, where it forms a narrow band; its posterior extremity is immediately in front of the direct cerebellar tract, while its anterior extremity terminates at the point where the anterior roots enter the cord.

The *origin* of this tract is at a much lower level than that of the direct cerebellar tract, since its fibres are found even in the lower part of the lumbar enlargement (Bechterew), and it is found to degenerate after transverse lesions seated in the lumbar region, whereas the direct cerebellar tract is unaffected. It is doubtful from which part of the cord or from what cells these fibres originate. According to certain authors, some, but not all the fibres proceed from the cells of the columns of Clarke in the same way as those of the direct cerebellar tract, while the others, which are of larger size and more numerous, take origin from the cells of the anterior horns.

However this may be, the tract formed by these fibres passes upwards along the whole length of the cord, increasing gradually in size and becoming more and more distinct from the direct cerebellar tract until it reaches the medulla oblongata. It there becomes comma shaped, the head of the comma being opposite the gelatinous substance while its tail is upon a plane posterior to that of the ascending root of the 5th nerve. It is always seated in front of the direct cerebellar tract, but instead of being in contact therewith is distinctly separated from it.

The *termination* of the fibres of the tract of Gowers is little better known than their origin. According to some authors they are in intimate connection with the *nucleus lateralis*,* in

* This nucleus is seated in the lower part of the medulla oblongata on a level with the commencement of the olive. Anteriorly to it is found a group of large multipolar cells of the same type as those which are seen in the anterior horns of the spinal cord. The nucleus itself is composed of small cells of the bi-polar type.

which a certain number of fibres terminate (Bechterew); according to Tooth such is the destination of the narrow fibres contained in

Fig. 64.

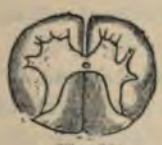

Fig. 65.

Fig. 66.

Sections of the cord from a case of fracture of the spine with destruction of the cord between the 8th cervical and 1st dorsal pair of nerves (after Tooth). Fig. 64 : 7th cervical; almost the whole posterior column is degenerated, except in the vicinity of the posterior horn, where the 8th and 7th cervical roots enter the cord introducing a contingent of sound fibres. The whole surface of the antero-lateral columns is degenerated, the degeneration at some points invading the cord to such an extent that the zone of traumatic degeneration seems to have been again reached. Fig. 65 : 4th cervical; in the posterior columns the distance of the degeneration from the grey substance is gradually increasing. In the antero-lateral columns degeneration has only occurred at the surface of the cord. Fig. 66 : 2nd cervical; in the posterior column the degeneration is almost entirely limited to the tract of Goll. That of the antero-lateral column (degeneration of the direct cerebellar tract, and of the tract of Gowers) no longer extends anteriorly as far as the margin of the anterior fissure.

this tract. The thicker fibres pass into the cerebellum through the superior cerebellar peduncle.

It will be noticed, gentlemen, that all the observations which

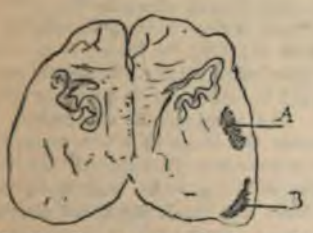

Fig. 67.—Lower part of the medulla oblongata from the man in whom there was a transverse lesion with destruction of the cord between the 8th cervical and 1st dorsal pair of nerves, the subject of figs. 64 to 66. A, Degeneration of the tract of Gowers; B, Degeneration of the direct cerebellar tract.

Fig. 68.—Middle portion of the same medulla oblongata as in fig. 67 (after Tooth). A, Degeneration of the tract of Gowers; B, Degeneration of the direct cerebellar tract.

we have just made tend to show that the direct cerebellar tract is distinct from the tract of Gowers.

This fact is confirmed by their mode of development. Bech-

terew has in fact shown that the tract of Gowers appears at a relatively late date, since it does so at the commencement of the 8th month, immediately before that of the pyramidal tract, while at that time the development of the direct cerebellar tract has already occurred.

Such anatomical considerations I wished to put before you with regard to the tract of Gowers, which contain all that refers to the degeneration of this tract after transverse lesions of the cord, and it appears to me useless to say more upon this subject.

The changes in the GREY SUBSTANCE OF THE CORD which are

Fig. 69.—Djagram representing the principal seats of ascending degeneration in transverse lesions of the cord (dorsal region). A, Column of Goll. BB, Column of Burdach; the degeneration in the part of this column adjoining the column of Goll is more pronounced than in that which is close to the posterior horn. The degeneration in the column of Burdach can only be traced to a short distance above the point where the transverse lesion occurs. C, Direct cerebellar tract. D, Tract of Gowers. This tract is largest at its posterior part, becoming gradually narrower anteriorly. The point of its termination is somewhat uncertain. It is doubtful whether the fibres in which ascending degeneration occurs by the side of the anterior fissure (they should have been prolonged along the whole length of the fissure) belong to the tract of Gowers or not. In my opinion they are distinct from that tract, the fibres seeming to be shorter than those of the tract of Gowers; they might be denominated the *sulco-marginal ascending tract.*

due to the ascending degeneration which follows transverse lesions of the cord, have as yet been little studied, and their description (Tooth, Hofrichter, Barbacci) is still incomplete. According to the latter author it is found that the grey matter above the transverse lesion has a granular appearance; that the nervous *reticulum* at this level is scanty, and may even completely be lost, while the cells disappear, or exhibit changes which are cl[illegible] retrograde in character, some of them becoming full of

pigment, while in others more or less atrophy exists. The changes are always more marked in the region of the posterior than in that of the anterior horn. As the distance from the point of division increases, these changes diminish. The granular appearance first ceases, then the lesions in the cells of the anterior horns, then those in the cells of the posterior horns, and lastly those in the nervous reticulum. As regards the true seat of the degeneration with respect to tracts or systems which are clearly determined, no knowledge at present exists which permits any opinion to be formed upon this point.

The study of ascending and descending degeneration after transverse lesions of the cord, gentlemen, is now ended, and, as might have been foreseen, all the tracts are affected in one or other direction according as their trophic centre is seated above or below the transverse lesion. There is, however, one tract which does not seem to degenerate in either direction, and which is only affected within the "zone of traumatic degeneration." This tract is that which has been termed the *lateral limiting layer* (*seitliche Grenzschicht*). The escape of this tract, however, is but apparent; it really degenerates in the same way as the adjoining tracts, but since it is composed of what appear to be very short commissural fibres their degeneration only occurs for a limited distance, and is entirely contained within the "zone of traumatic degeneration."

LECTURE VI.

DEGENERATION OF THE NERVES AND SPINAL CORD AFTER AMPUTATION OF A LIMB.

Such degeneration seems at first sight opposed to the doctrines of physiology. At the same time changes of this kind are frequent. They have been shown to occur by numerous observers. Works of Bérard, Vulpian, Dickinson, Hayem. Dejerine and Mayor, Hayem and Gilbert, Friedländer and Kranse, &c.

Changes in the *central extremity of nerves* after amputation of a limb; frequent increase in size (does not always occur), increased diameter of the fasciculi constituting the nerve. Considerable diminution, in a transverse section, of the fibres containing myelin. *Islets of degeneration* not to be confused with the *primary islets*, being in reality the traces of previously existing nerve fibres. The islets of degeneration are formed of a collection of nerve fibres composed of a small axis cylinder, and fine primitive sheath, with or without myelin.—Evolution of these islets, dispersion of the nerve fibrils, *myxoid aspect* of the partitions separating the different primary islets from each other, slight thickening of the fibrous bundles placed between the fasciculi of the nerve.

Uncertainty with regard to the condition of the *ganglia connected with the nerve roots*, and the *nerve roots* themselves.

GENTLEMEN,—The consideration of secondary degeneration in the cord would be incomplete unless a few words were said with regard to the occurrence of such degeneration after *division of the peripheral nerves*, especially when this is due to AMPUTATION OF A LIMB.

At first sight, owing to ideas derived from the study of physiology, such degeneration seems impossible. We are told that since the cells in the anterior horns of the cord are the trophic centre of the *motor* nerve fibres, the cells of the ganglia connected with the nerve roots being that of the *sensory* fibres, no degeneration of the cord can occur after the lesion of a peripheral nerve fibre. Whilst, in fact, descending degeneration could alone affect the motor fibres, ascending degeneration of the sensory fibres would be prevented by the ganglia connected with the nerve roots. The so-called fibres of recurrent sensibility are not, as it is generally acknowledged, sufficiently numerous to produce ascending degeneration which is at all extensive.

Whatever the value of these conclusions formed in the

laboratory may be, it is an undeniable fact that in most cases after amputation of a limb changes which are truly singular are found to occur in the central extremity of the divided nerve, and upon the corresponding side of the cord after a certain time. The study of these changes will form the subject of this lecture.

Without referring to the observations of Bérard (1829), which are not quite accurate, it is to Vulpian that the honour of first studying and methodically describing facts of this kind in several memoirs written between 1868 and 1872 must be given. At a later date this author inspired the interesting researches of Dejerine and Mayor (1878). In the meantime Hayem (1875) had devoted his attention to similar facts, and as you know that author has more recently (1884) been again occupied in a work written in conjunction with my distinguished colleague and friend Gilbert. It is but fair to name Dickinson, who, in the same year as Vulpian (1868), gave a most remarkable description of the lesions which he had observed in the spinal cord after amputation of a limb. Dickinson very clearly indicated the posterior column as the principal seat of the atrophy, whilst, as I shall soon observe, Vulpian specially localized that atrophy in the antero-lateral column. I do not wish, however, to insist upon this historical review, since in the course of this lecture more than one opportunity will occur of stating the name and opinion of the different authors who have made observations or written works upon the subject which now interests us.

Amongst these works there is one to which I must draw attention, namely, that of Friedländer and Krause, in which a thorough investigation may be found of the forms of degeneration which follow amputation. The articles which that work contains are numerous, and of the greatest value, while the interpretation given by the authors is new; in fact I must allude more than once to this work, though I am far from agreeing with the conclusions which are formed in it.

That you may be able to form a clear idea of the changes which occur in the cord after amputation, the study of the cord alone must be, to some extent, abandoned in these lectures, of which the special subject is "Diseases of the Spinal Cord," and the condition of the *nerve trunks* above the region in which the amputation occurred be investigated. We will not, however, go so far as to study the alterations which are shown to exist in

connection with terminal neuromata. Those who are interested in these changes will find them described in the work of Hayem and Gilbert.

Let us take as example and basis for the description of ascending degeneration after amputation a case which I have had occasion to observe during the course of this year, and from which I shall be able to put before you many interesting microscopical specimens. The patient was a man aged forty years, in the middle part of whose left thigh amputation had been performed twenty years previously, and whose death was due to phthisis while under my care.

The sciatic nerve upon the left side was considerably increased in size along its whole length, its dimensions being of about double the size of those of the same nerve upon the right side.

This fact, gentlemen, which has been already mentioned by several authors, is by no means constant; I did not, in fact, find it to exist in two cases of amputation of the arm, or in one of amputation of the foot.

In a case of amputation of the arm Hayem and Gilbert remark that "the radial nerve was slightly diminished in size, while the ulnar gradually increased from the brachial plexus to the terminal neuroma, near which it was twice as large as the sound nerve on the opposite side; the median nerve was of the same size on the two sides." It would be difficult to find a better example of the uncertainty which exists with regard to such increase in size, since in this case the three nerves of the stump were affected in a different way. It should be noticed that the increase of volume when it exists is more pronounced in the peripheral parts of the nerve, and tends to diminish or disappear gradually as the spinal cord is approached.

Leaving the macroscopic aspect of the nerve, the microscopical characters of the lesions which it presents will now be studied. If a section of the sciatic nerve be examined, which I have asked you to take as the basis of this description, it is at once evident, even to the naked eye, that the primary fasciculi which compose it have for the most part a much larger diameter (2, 3, or 4 times as large) than those in a healthy nerve, an increased degree of transparency corresponding with the increased size of the fasciculus. It will presently be explained that these fasciculi have degenerated, those which have most increased in

size, and are most transparent, being in reality those in which the greatest change has occurred. Upon examining a section stained by the hæmatoxylin of Weigert, it is at once recognized that the lesions are extremely pronounced. Instead of the regular mosaic pattern which the transverse section of a healthy nerve presents owing to juxtaposition of the sheaths of myelin, which are coloured black, a very small number of such sheaths is

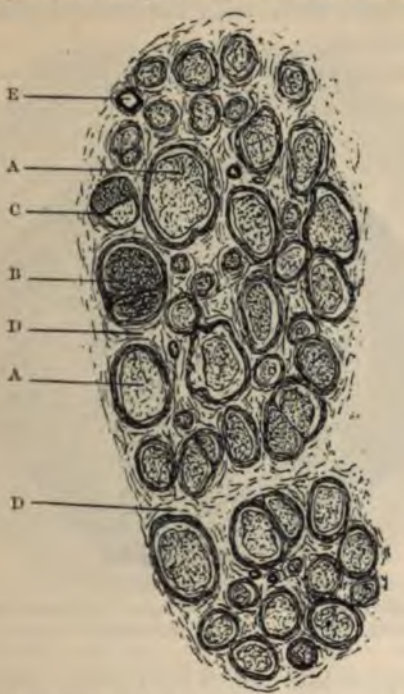

Fig. 70.—Section of the left sciatic nerve in a case of amputation of the thigh on the same side 20 years previously. A, Primary fasciculus in which degeneration has occurred. B, Primary fasciculus in a normal condition. C, Primary fasciculus in half of which degeneration has occurred, while the other half is in a normal condition. D, Portion of the nerve between the fasciculæ having a myxoid appearance. E, Blood vessel.

perceived; these sheaths are also scattered throughout every fasciculus, not being placed in groups, but completely isolated from each other. In a few fasciculi, however, two or at the most three, and these the smallest in size, the ordinary mosaic pattern indicates that their condition is normal; these come from the nervous branches proceeding to those parts of the

thigh which are seated above the point of amputation, while they are completely separate from the affected fasciculi, whose fibres were, for the most part, destined to parts of the lower extremity seated below the amputation. In some sections fasciculi may be seen divided into two distinct halves separated by a partition of connective tissue, of which one is degenerated and the other sound, the former being composed of fibres coming from the part amputated, while the latter commences above the point of amputation.

Thus the fibres which contain myelin are very rare in the most degenerated fasciculi, being replaced by small *islets* of

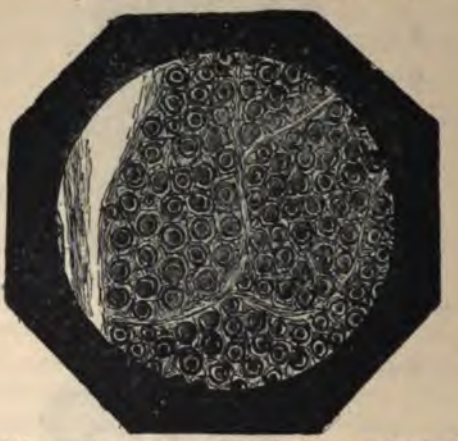

Fig 71.—Part of the section of a healthy sciatic nerve (stained by the hæmatoxylin of Weigert) (from nature, and semi-diagrammatic). The segments isolated by the white lines are the primary islets.

very singular appearance, which have been observed by different authors, but to which special attention seems only to have been drawn by Friedländer and Krause, although, in my opinion, these authors are far from attributing to them their true signification.

These islets do not become of a black colour when the method of Weigert is employed, but are distinctly coloured by carmine; they vary in size, having usually a mean diameter three or four times as great as that of a healthy nerve fibre with its sheath of myelin. They also vary in form. Where sufficiently isolated from each other, their form is completely round, while, if pressed against each other, their shape is altered on account of this reciprocal compression.

As to the composition of these islets, I completely disagree, gentlemen, from Friedländer and Krause, as you will see. If with a feeble magnifying power (objective 0 or 1 of Vérick) the section of a healthy nerve is examined, whatever colour is employed, the nerve fibres are divided by means of connective tissue into a variable number of groups (*primary islets* of some authors). These primary islets are much larger in size than the other "islets" of which I have just spoken, and to which, in order to

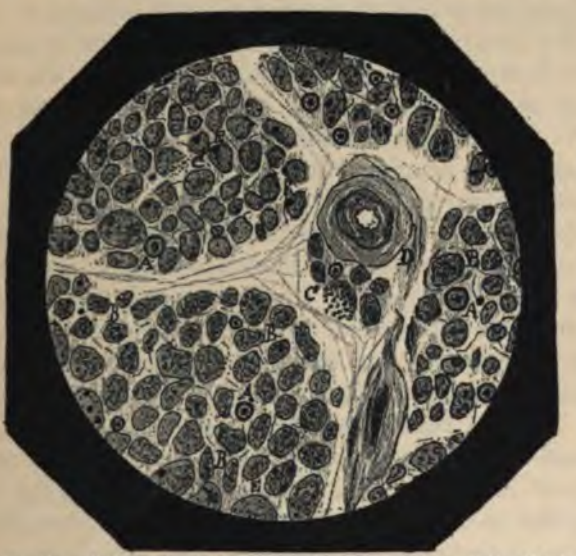

Fig. 72.—Section of the left sciatic nerve after amputation of the thigh upon the same side 20 years previously. AAA, Nerve fibres in a normal condition which have retained their axis cylinder and sheath of myelin. BB, Islet of degeneration of rounded or polyhedral form containing in their interior numerous points which are the fine nerve fibres occurring in degeneration. In some of these islets one or more black points are seen which indicate the presence of fine fibres containing myelin in the islets. CC, Islets of degeneration in the margin of which a rupture has occurred so that the nerve fibres escape and become separated from each other. D, Blood vessel with thickened wall (much exaggerated in diagram). The segments separated by large white tracts are the primary islets. (From nature, semi-diagrammatic, coloured by the hæmatoxylin of Weigert.)

distinguish them from those which are primary, I shall give the name "*islets of degeneration.*" According to Friedländer and Krause, the islets of degeneration are merely primitive islets, of which the fibres have wasted, and with considerable reduction of their size in consequence. At first sight this explanation

seems extremely probable, but upon examining it more closely I was convinced that this is not the case. The primary islets, far from being diminished in size, are found to be of larger size in a degenerated than in a sound nerve. As regards the islets of degeneration, each of them merely represents, in my opinion, a *degenerated nerve fibre*, of which the size has increased to three or four times that which it had previously. If one exposes one of these islets of degeneration to a higher magnifying power (objective 3 of Vérick) it is found to be composed of a mass of very narrow fibres with a cylinder axis, whose diameter is much smaller than would normally be the case, and a sheath of Schwann similarly reduced in size, and of great tenuity. As regards the sheath of myelin, it is usually absent; but islets of degeneration often occur in which the existence of one, two, or three extremely small black points can be recognized by the method of Weigert, indicating the existence in this part of a very narrow nerve fibre with a sheath of myelin.*

I am strongly inclined to think that each of these islets of degeneration is surrounded, at any rate when its evolution is not too advanced, by the sheath of myelin which previously surrounded the nerve fibre, of which this islet is the remaining trace. A large number of nuclei also exist within the islet, which are very clearly seen in sections made at right angles to the axis of the nerve, and even more clearly when the sections are parallel to that axis. They are scattered throughout the interstices between the nerve fibrils, and their long axis is in the same direction as that of these fibrils, namely, parallel to the axis of the nerve.

The pathology of these tracts of degeneration would be shortly as follows:—Owing to some influence which need not now be considered, one of the nerve fibres at the central extremity of a divided nerve degenerates, that is to say, its cylinder axis and sheath of myelin disappear while the sheath of Schwann remains. Consecutively within this sheath of Schwann nerve fibrils are developed having a small axis cylinder, and narrow sheath of Schwann containing a substance which but rarely gives the re-action of myelin. The number of these nerve fibrils varies, but

* The existence of these narrow nerve fibres has been mentioned by Hayem, and they were very clearly described in a memoir by Hayem and Gilbert. (*Arch. de Physiol.*, 1884, p. 430 et seq.)

is sometimes considerable (5, 10, 15, 20, or more). Owing to the development of these nerve fibrils, the sheath of Schwann, which I shall call "primary," becomes rapidly distended, and the islets of degeneration thus become of much larger size than the sound nerve fibres by which they were preceded. Owing also to the influence of the development of the nerve fibrils and the external pressure which they exert, the primary sheath of Schwann, having reached the extreme limit of its extensibility, bursts; the contained fibrils become free, and being no longer kept together and united by this sheath, separate from each other. The islet of degeneration is broken up and cannot be recognized, completely ceasing to exist when the nerve fibrils which composed it are so far apart as to have apparently no connection with each other.

Such, in my opinion, is the way in which evolution of these islets of degeneration occurs, which will be observed, gentlemen, to be in direct opposition to that professed by Friedländer and Krause; when they believe atrophy of the nerve fibres to occur, my opinion is that these fibres regenerate and multiply, and what they look upon as the indication of a primary islet, I regard as the remaining trace of a degenerated nerve fibre.

The other parts of the section of the sciatic nerve will now be examined, and for that object a lower magnifying power will be used. In considering the different nerve tracts it is found that in the degenerated tracts the *partitions* separating the primary islets from each other are twice or three times as large as those existing in healthy tracts; they seem also to be more relaxed, and have a truly *myxoid appearance*. An analogous band of myxoid tissue is also found at the surface of the most altered tracts, separating them from the fibrous sheath furnished by the neurilemma, so that, when contraction has taken place in the section, these tracts have a pronounced tendency to escape from the sheath formed by the neurilemma. The connective tissue which separates the different fasciculi from each other is moderately thickened. The spaces surrounded by many degenerated tracts are alone much increased in size, this seeming especially due to increase in the diameter of the surrounding tracts. The blood vessels contained in the degenerated tract are not increased in number, and their walls are at most but moderately thickened.

Thus the most important factor, gentlemen, in increasing the

size of the nerve trunks seated above the point of amputation is certainly the multiplication of the small nerve fibres, which has just been described.

I cannot say to what lesions atrophy of the nerve trunks is due, having never had the opportunity of making a histological examination in cases of this kind.

The changes presented by the nerve trunks are now known, healthy fibres being found in greater number as the distance from the point of amputation increases. The degeneration will now be followed in its ascending path.

As regards the GANGLIA CONNECTED WITH THE NERVE ROOTS, which some observers considered unaffected, I should prefer to give no opinion. I have examined many sections of these organs, but must confess to having formed no opinion with respect to them, and that I am unable to say whether they are or are not altered.

During a long time observations have been made with regard to lesions in the spinal nerve roots, but with very different results. Vulpian believed them to be unaffected, while Dickinson claimed to have observed a certain degree of atrophy in the posterior roots. Subsequent observers showed the same difference of opinion, a small number of whom found no lesion in the nerve roots, while some observed degeneration in the posterior roots alone, others (Hayem and Gilbert) in both the anterior and posterior roots.

In the case of amputation of the thigh which was taken as basis and example in this lecture, lesions were not very clearly seen to exist in the nerve roots. Almost the only change noted was the somewhat large increase in the number of small nerve fibres as compared with those of larger size, a disproportion which existed both in the posterior and anterior roots. As in the normal condition the narrow fibres are numerous in the roots connected with certain parts of the cord, this estimate is of such an extremely delicate nature, that the reserve with which I mention these facts will be understood.

LECTURE VII.

DEGENERATION OF THE NERVES AND SPINAL CORD AFTER AMPUTATION OF A LIMB (*continued*).

Alterations in the SPINAL CORD :—According to Vulpian the atrophy especially involves the antero-lateral column and grey matter. Dickinson believes it to be more pronounced in the posterior column. The latter opinion seems preferable to the former. Opinion of Flechsig. Friedländer and Krause describe amongst their cases both atrophy of the *posterior column* and of the *posterior horn*, disappearance of a large number of cells in the *postero-lateral group* of the grey matter, diminution in size of the *anterior horn* with a reduction in the number of its cells, atrophy of the *column of Clarke* with disappearance of many of its cells. Results furnished by examination of the spinal cord in my own autopsies. Band of sclerosis in the posterior column on the same side as the amputation. Thickening of the connective tissue in the posterior column of the opposite side.

Interpretation of these different lesions: opinion expressed by Friedländer and Krause: personal conclusions.

GENTLEMEN,—In the preceding lecture the lesions of the peripheral nerves and nerve roots after amputation of a limb were considered; the alterations which occur in the spinal cord itself will now be discussed.

It may be henceforth said that these alterations have been very differently appreciated by the authors who have studied this subject. Whilst Vulpian supposed the *antero-lateral column* and *grey substance* to be especially affected by atrophy, Dickinson believed the latter to be far more pronounced in the *posterior column*. Lastly, Vulpian, Dejerine and Mayor observed cases in which atrophy was found to an almost equal extent in all parts of the cord upon the side which corresponded to the amputated limb. Yet another opinion is held by Flechsig, who, on account of the fact that malformation, or at least want of symmetry, frequently exists in the spinal cord of those who are healthy, concludes that the atrophy in one half of the cord or one of the tracts composing it observed after

amputation, is really *congenital absence of symmetry* and totally unconnected with the amputation.*

In opposition to this opinion, which is of a very sweeping nature, the facts themselves taken collectively need alone be considered. Absence of symmetry is certainly frequent in the cord, but is not usually found as accentuated as when observed in cases of amputation. In every case of amputation Friedländer and Krause observed such atrophy, and I myself have seen it three times in three cases. After such correspondence, I would almost say constancy, in the results, in my opinion it is impossible to deny the existence of a direct and not fortuitous relation between atrophy of the cord and amputation of a limb.

The figures now before you, borrowed from the memoir of Friedländer and Krause (figs. 73, 74, 75), show the degree which may be obtained by the atrophy. In amputation of the lower extremity these authors have observed, in addition to atrophy of the *posterior column*, a diminution in the size of the *posterior horn*, at least in the lower part of the cord, and the disappearance of a large number of cells from the *postero-lateral group* of the grey matter.† With regard to the *anterior cornu* it was found to be perceptibly reduced in size in the lumbar region, and the number of cells in the ganglia were diminished by a half or two-thirds.

Lastly, the dimensions of the *column of Clarke* were smaller on the side of the amputation, and the cells less numerous by about a fifth. As regards the anterior horn reference must be again made to the work of Hayem and Gilbert, from which I borrow a diagram (fig. 76), which shows most clearly the atrophy

* In a recent work (*Des affections nerveuses centripètes consécutives a la section des nerfs et aux amputations des membres. Acad. Royale de Med. de Belgique*, 1891) Vanlair considers the alteration in the nerve roots and cord inconstant. He thinks that when existing in man they are due rather to the surgical affection which requires amputation than to removal of the limb. As regards the operations which he has performed in animals Vanlair summarises the results obtained by saying that "on account of their age and the absence of any pre-existing peripheral lesion, adult animals usually escape, to a great extent, the consequences of amputation; in them myelitis due to section of the nerves or even removal of the limbs is usually absent. It is rarely met with except in bilateral operations."

† In the observations published in *Arch. de Physiol.*, 1873, Hayem had already stated the fact that when the sciatic nerve is torn away in young rabbits rapid atrophy of the nerve cells belonging to the group in the intermedio-lateral tract is found to occur.

and rarefaction of the cells of the ganglia after the loss of an arm.

As regards my case of amputation of the thigh to which

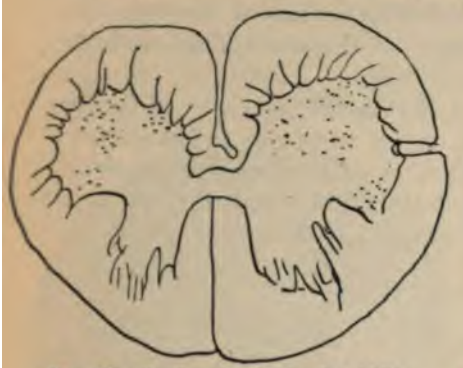

Fig. 73.—Section of the cord (middle part of the lumbar enlargement) from a case of amputation of the lower extremity on the left side (after Friedländer and Krause). It will be observed that the left half of the cord in almost every part, but especially in the posterior column and anterior horn, is smaller in size than the right half. (The notch opposite the lateral column on the right side is due to a slit made by the authors before the preparation was hardened with the object of placing the sections in a better position.)

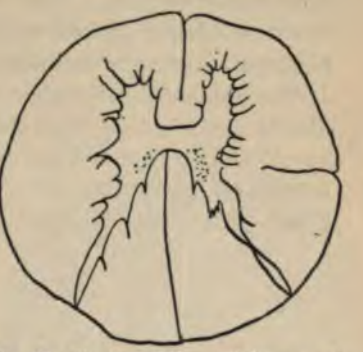

Fig. 74.—Section of the cord (lower part of dorsal region) in a case of amputation of the left lower extremity (after Friedländer and Krause). The left half of the cord is smaller than the right, and the cells of the columns of Clarke are in smaller number.

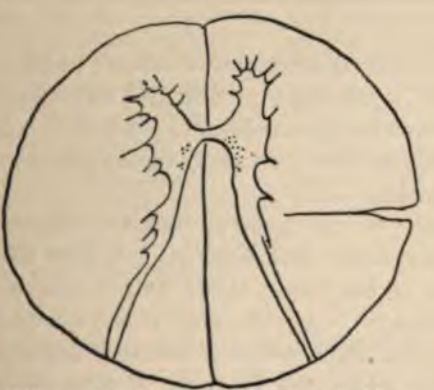

Fig. 75.—Section of the cord (middle part of dorsal region) in a case of amputation of the left lower extremity (after Friedländer and Krause). Same remarks as under fig. 74.

allusion has been frequently made, the same condition of atrophy affecting the most different parts could not be observed, while on the other hand the *posterior column* was found to be clearly diminished in size. This diminution extended through the whole or almost the whole length of the cord. (The medulla oblongata was not examined.)

The antero-lateral column on the opposite side seemed to me also of smaller size than that on the same side (a fact already mentioned by Bignami and Guarnieri). As regards the *posterior column* on the side of the amputation it is my opinion, which is based both upon that of other authors, as well as the results of my

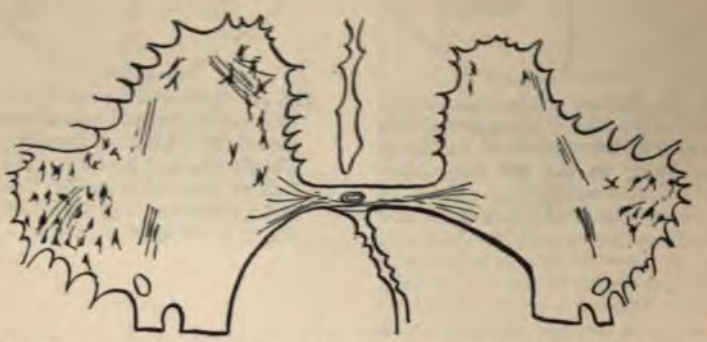

Fig. 76.—Section of the cervical cord in a case of amputation of the right arm at its lower part (after Hayem and Gilbert). This sketch only represents the two anterior horns; the small volume of the right anterior horn (the side of the amputation) and the small number of nerve cells in that part will be noticed.

own experience, that this column is the one which presents most constantly, and in the most pronounced way, the diminution in size which is now being considered. Whether this diminution affects the whole or some parts only of the posterior column will now be considered.

In opposition to the general opinion, especially that expressed by Friedländer and Krause, I believe the latter to be the true state of the case. After amputation of the arm or thigh I have always clearly observed at the centre of the wasted posterior column a band, which if not very dense, is at any rate well defined, in which the fasciculi of connective tissue were more thick and numerous. This seemed to indicate that it is specially at *that* point that atrophy of the nerve fibres coming

from the posterior roots on the same side as that of the amputation occurs. This band, of sclerosis, as it may be named, though it must be confessed that this is a somewhat exaggerated term, did not occupy exactly the same position in the cervical region after amputation of the thigh or arm. Though unable to give a detailed description of what occurs, I shall place before you sketches of this band of sclerosis at different heights in the cord.

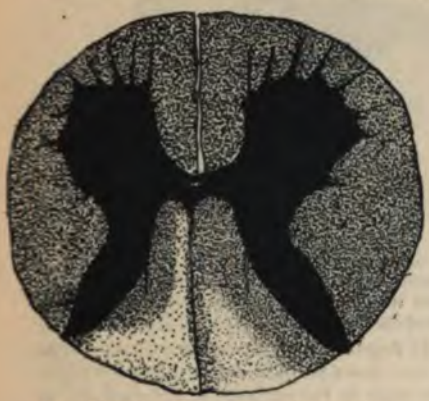

Fig. 77.—Cord in the lumbar region from a case of amputation of the left thigh performed twenty years previously. In the left posterior column, viz., on the side of the amputation, degeneration has occurred; there was also in the posterior column of the opposite side an islet of degeneration, which however was less pronounced in degree, and of smaller size. It will be observed that the zone of degeneration in the left posterior column is at the postero-external part adjoining the posterior cornu, which seems to indicate that it is due to degeneration of the fibres of the posterior roots seated below the point of section. The reduction in size of the left half of the cord is very pronounced in degree.

Fig. 78.—Cord in the dorsal region from the same case of amputation of the thigh as fig. 77. In the left posterior column (the same side as that of the amputation) degeneration has occurred. In the posterior column on the opposite side there was also an islet of degeneration, which however was less pronounced in degree and of smaller size. It will be observed that the area of degeneration is much nearer the posterior median fissure than in the lumbar cord, and that it is clearly seated in the column of Goll. The diminution of size on the left side of the cord is very pronounced.

There is one point on which I would specially insist, namely, upon the slight increase of connective tissue in the posterior column of the side opposite to that of the amputation, in a part which is in symmetry with the band of sclerosis upon the side

the posterior roots, have also observed degeneration of the posterior column on the opposite side. O. Barbacci, on the other hand, after division of half the spinal cord in the dog and cat, also observed ascending degeneration of the posterior column on the opposite side. These facts seem to agree fully with that which I am now stating, and it is for this reason that I thought it right to mention it.

Such, then, are the lesions which may be observed both in the *nerves* and *spinal cord* after amputation. Atrophy of the *cerebral convolutions*, which has been mentioned by some authors, does not come within the scope of these lectures.

How can the occurrence of such degeneration be explained since it is in opposition to the doctrines of pure or pathological physiology? The central extremity, for instance, of peripheral nerves is found to undergo the most extensive degeneration, when neither the ganglia connected with the nerve roots nor the anterior horns of the cord are affected by the wound. How does this accord with the well-known law of Waller that a nerve fibre only degenerates when separated from its trophic centre?

Friedländer and Krause have attempted to explain this in a very ingenious manner. According to them the degeneration which occurs at the central extremity of an amputated nerve involves the *sensory fibres* alone and consequently those of which the function is afferent. All the sensory fibres, however, coming from parts of the limb seated below the amputation do not degenerate but merely those *which terminate in such special terminal organs* as the tactile-corpuscles, &c., the sensory fibres which have a free extremity not presenting such a change. In support of this supposition it is stated that if the sciatic nerve is examined after amputation of the thigh, or of the leg above the malleoli, the number of degenerated fibres is found to be the same in the two cases. This, according to Friedländer and Krause, is due to the fact that the *tactile-corpuscles* being specially numerous in the skin of the foot, the number of nerve fibres which terminate in this part and are affected by ascending degeneration is almost the same wherever amputation is performed in the lower extremity, so long as it is above the malleoli.

I have myself had the opportunity of examining sections of the sciatic nerve, both after an amputation of the thigh and a supramalleolar amputation, and the result of my examination

of the atrophy. This may possibly indicate that some fibres of the posterior roots upon the side of the amputation have

Fig. 79.—Spinal cord in the cervical region from the same case of amputation of the thigh as figs. 77 and 78. In the left posterior column degeneration has occurred upon the same side as that of the amputation, there is also an islet of degeneration, which is very slight in degree, in the posterior column of the opposite side. It will be remarked that these areas of degeneration are much less extensive and slighter in degree than in the sections of the cord in the dorsal or lumbar region. The diminution in the size of the cord upon the left side is less pronounced than in the preceding figures.

Fig. 80.—Section of the cord (cervical region) after amputation of the right arm. It will be remarked that two areas of degeneration (the parts of light colour) exist, separated by a band of healthy tissue in the posterior column of each side.

The degeneration is again most manifest upon the right side, which corresponds to the seat of the amputation.

crossed each other in the posterior column of the opposite side. Tooth, Oddi and Rossi having made a section of some of

the posterior roots, have also observed degeneration of the posterior column on the opposite side. O. Barbacci, on the other hand, after division of half the spinal cord in the dog and cat, also observed ascending degeneration of the posterior column on the opposite side. These facts seem to agree fully with that which I am now stating, and it is for this reason that I thought it right to mention it.

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I have myself had the opportunity of examining sections of the sciatic nerve, both after an amputation of the thigh and a supramalleolar amputation, and the result of my examination

was in direct opposition to the statements made by these two authors. I found, in fact, that in the case of amputation above the malleoli but few fibres were degenerated, whereas you have seen that after amputation of the thigh a very large number were involved.

Everything being taken into consideration, it is in my opinion certain that with a case, such as the one which I have mentioned, before us it is impossible to maintain that the sensory fibres are alone involved. This would almost amount to saying, on account of the small number of fibres which escape, that the sciatic nerve only contains an infinitesimal number of motor fibres, whilst as you know, on the contrary, it contains a large supply of motor branches.

I certainly admit that some fibres may undergo Wallerian degeneration at the central extremity, not as Friedländer and Krause believe owing to loss of function (their *terminal organ* being destroyed), but in my opinion because this so-called terminal organ is for a large number of fibres the *nucleus of origin*, the true trophic centre.* In any case these fibres are not numerous, and an alteration in *them* would not explain such extensive degeneration as that observed in the trunk of the sciatic nerve.

How then can this divergence of opinion between different authors, I would almost say with respect to different cases observed by the same author, be explained? In my opinion the knowledge which we possess with regard to Wallerian degeneration is insufficient to solve this question. There is an unknown connected with it which the future will make clear. Perhaps the following fact may be taken as a partial explanation. All the autopsies upon which the history of these degenerations is based were connected with patients in whom an amputation had been performed a long time previously (10, 20, or 30 years), that is to say, at a time when antiseptic treatment was almost unknown. The stumps of all, or nearly all, the patients had only healed after suppuration continuing for a longer or shorter time; in consequence, the extremities of the divided nerves were exposed to many and various kinds of infection. Owing to these facts it seems possible that the

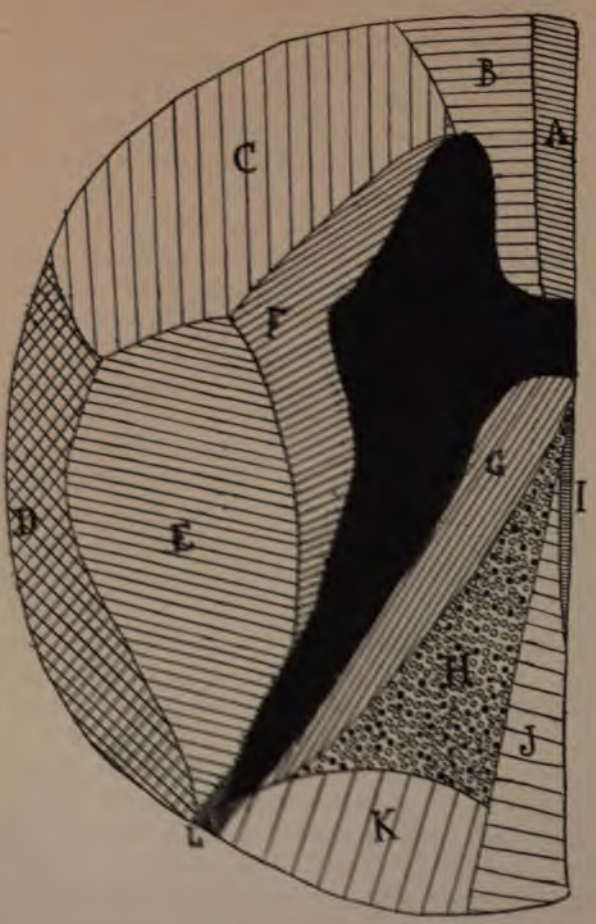
* I shall have occasion to explain this statement more fully in discussing the pathological anatomy of tabes.

alteration in the nerve fibres might be of *septic origin* (whatever the mechanism of its occurrence may be), and that some morbific agent may have penetrated into the extremity of the nerve whilst it was seated in the deep part of the wound. This is certainly but a supposition, which may not be the true one, but the occurrence of sympathetic ophthalmia being remembered, it may certainly be said that this hypothesis has at least a appearance of probability.

This, then, is my idea as to the origin of the considerable lesions which are observed at the central extremity of the divided nerve. Two causes tend to produce them: one is the occurrence of *ascending Wallerian degeneration*, which exclusively involves fibres, of which the originating nerve cell (trophic centre) is seated near the surface (in muscle, tendon, aponeurosis skin, &c.); the other, the existence of *ascending neuritis*, which occurs under the influence of the decomposing material within which the extremity of the divided nerve is placed. The nerve is then penetrated by some morbific agent,* which produces a special form of ascending neuritis at its central extremity, the sheath of myelin is soon absorbed, while the cylinder axis disappears, though I cannot say that it is entirely destroyed. A second stage then occurs, namely, that of *regeneration*.

The principal characters of this are known to you, namely, that in the sheath of Schwann, which is now without myelin, a somewhat large number of nerve fibrils (10 to 20) now appear, being formed of a thin sheath of Schwann, and a narrow cylinder axis with or without a small covering of myelin attached to it. By what means is this regeneration effected? Is it to be supposed that the nerve cells from which the fibres originate (or trophic centres) contained in the spinal ganglia throw out prolongations into the empty sheaths in greater number, and constitute the fibrils which are being considered? This is possible; but the regeneration might also be due to another cause: the fibrils which compose the cylinder axis of the nerve fibre not being all destroyed by the ascending neuritis (the communication of some of them with their trophic centre still existing) the morbid process merely separates them. Each of

* It should be stated that the microscopical examinations of nervous trunks made by Lesage, at my request, in two cases of amputation, merely gave negative results.



...ia.—Diagram of the tracts of the cord (at the junction of the cervical ...h the dorsal region) according to their mode of DEVELOPMENT (after the ...grams and descriptions of Flechsig).

...rect pyramidal tract. B, Fundamental tract of the anterior column. C, ...aining part of the lateral column. D, Direct cerebellar tract. E, Crossed ...midal tract. F, Lateral limiting zone. G, Anterior root zone. H, Middle ...l zone composed of two systems of fibres; some + + + + + = the fibres of ...first system of the middle root zone; ..... = fibres of the second system. ...dian zone. J, Column of Goll. K, Postero-internal root zone. L, Postero-...ernal root zone, or zone of Lissauer.

Fig. 80 *ter*.—Diagram of the tracts of the cord (at the junction of the cervical with the dorsal region) in connection with the study of SECONDARY DEGENERATION.

A, FIBRES LIABLE TO ASCENDING DEGENERATION: 1. Fibres of the *ascending sulco-marginal tract*. 5. Fibres of the *tract of Gowers*. 7. Fibres of the *direct cerebellar tract*. 8. Fibres of the *cornu-commissural tract*. 9. Fibres of the *column of Burdach*. 10. Fibres of the *external bandlet* properly so called. 11, Fibres of the *zone of Lissauer*. 12. Fibres of the *tract of Goll*.

B, FIBRES LIABLE TO DESCENDING DEGENERATION: 2. Fibres of the *descending sulco-marginal tract*. 3. Fibres of the *direct pyramidal tract*. Fibres of the *intermediate tract of the antero-lateral column*; some of these are stray fibres, and scattered through the tract of Gowers, the direct cerebellar tract, and, above all, the crossed pyramidal tract. 6. Fibres of the *crossed pyramidal tract*. 13. Fibres of the *comma shaped tract of Schultze*.

the nerve fibrils, which are thus dissociated, tends to reconstitute a complete nerve fibre, and it is this tendency which betrays itself by the existence of many fibrils in the previously existing sheath of Schwann.

This lecture, in which so many hypotheses have been mentioned, must not be brought to a conclusion, gentlemen, without my reminding you that these have, in every case, been put before you as mere suppositions which claim, not blind faith, but a fair examination. Such is my excuse for mentioning them. When every current theory fails, it is quite permissible to seek the truth in some other way.

LECTURE VIII.

SPASTIC PARAPLEGIA.*

History of the term *spasmodic tabes dorsalis*. Erb: *Spasmodic spinal paralysis*. Charcot: *Spasmodic tabes dorsalis*. Primary degeneration of the lateral columns. Different results obtained in the autopsies of adults. History of the disease itself. Heine: *paraplegia spastica cerebralis*. Little: *congenital spastic rigidity of limbs*. The researches of the English surgeon were for a long time unknown to neurologists. Memoirs of Rupprecht and Feer. Symptoms of the disease. Spasmodic gait: attitude of the head and limbs. Attitude in the sitting position: position of the trunk and limbs due to difficulty in flexing the thigh upon the pelvis. Attitude of the upper extremities, which are often little affected. Muscles of the front and back of the neck. Disorders of speech. Strabismus. Appearance of the face. Awkwardness of movements. Fibro-muscular retractions. Influence of fatigue, of the emotions.

GENTLEMEN,—In the course of these lectures, which are specially devoted to diseases of the spinal cord, I shall be more than once induced to speak of cerebro-spinal affections, the study of which is more properly connected with disease of the brain than with that of the spinal cord. Spastic paraplegia is one of the number, and when the proper time arrives for making this statement, when the pathological anatomy of the affection is being considered, it will be recognized that the lesions of the cord are of quite a secondary nature. At the same time it is in my opinion right to speak of this disease at the present moment, and to associate it with diseases of the spinal cord, because in the instruction given in these lectures it is necessary, in order to avoid the confusion which must otherwise inevitably occur, to respect the classifications which have been established by long usage, specially as regards well-established clinical conditions. These slight concessions may be reasonably made as regards the form of the lectures, so long as care is taken to make no error and to consent to no sacrifice to current ideas, which are in any way inexact.

It is necessary to review shortly the HISTORY of this disease in order that the position of spastic paraplegia, from a nosological

* *Synonyms:* spasmodic tabes dorsalis (Charcot). Spasmodic spinal paralysis (Erb). Congenital spastic rigidity of limbs (Little). Primary spastic paraplegia (Gowers). Spasmodic spinal paralysis (Eustace Smith). Primary lateral sclerosis (Gowers). &c.—(Translator.)

point of view, may be understood. In 1875 Erb, and a few months later Charcot, described an affection of which the special indication was pronounced spasmodic paresis in the extremities, without loss of sensation. Erb believed that in all probability this was due to primary degeneration in the lateral columns of the cord. Charcot was of the same opinion with some modifications. This idea as to the nature of *spasmodic spinal paralysis* (name given by Erb) or *spasmodic tabes dorsalis* (name given by Charcot), thus at once gave it the right of being included in nervous pathology, but owing to a singular misfortune the autopsies of adult patients in whom the diagnosis of spasmodic tabes dorsalis had been made, there was found either disseminated sclerosis, or transverse, or focal myelitis, or perhaps amyotrophic lateral sclerosis; not a single case in fact occurred in which primary degeneration of the pyramidal tract existed which had been looked upon as the basis of this new morbid condition.

An inevitable reaction followed, and thus during the last few years the diagnosis of spasmodic tabes dorsalis has been scarcely ever made.

Should this affection be definitely excluded from a nosological catalogue? If a few cases, said to have occurred in adults between the ages of 30 and 50 years, had alone to be considered I should be very inclined to answer this question in the affirmative, since, except in some cases of general paralysis of the insane (Westphal), primary and isolated degeneration of the pyramidal tract occurs but rarely in the adult. As Charcot very truly observed in his lectures in 1880, the existence of such primary degeneration in cases of so-called spasmodic tabes dorsalis had received no direct confirmation. Until the present time the same desideratum exists.

If, however, instead of looking for this disease in the adult, one seeks it in childhood it is quite otherwise. You will meet some day with a case of this kind. You will see a child suffering from general rigidity in the four limbs sufficient to embarrass the movements and impart to them special characters, but without preventing their occurrence. The tendon-reflexes will be exaggerated; at times strabismus will occur, speech will be slow, and the articulation of a spasmodic nature; some mental change or alteration in the character may occur, sensation remaining absolutely normal, and the special senses quite unaffected.

It is with regard to these young patients and them alone that in the present state of our knowledge upon this subject the name of *spasmodic tabes dorsalis* should be applied; this term will be thus perceived to have quite a different meaning from that applied to it when the first works of Erb and Charcot were written, since at that time a disease which occurs in adults was alone considered.

Such is the history of the term spasmodic tabes dorsalis; as regards the disease itself, when understood, as I have just stated, its history is very different. It was Heine who in 1840 first described some cases under the name *paraplegia spastica cerebralis*, a name which, I may say, is most suitable, and deserved to be permanent. This description however was merely suggestive, and the author, to whom we are indisputably indebted for making this disorder a true clinical entity, is the English surgeon Little, who in different works written between 1846 and 1870 clearly traced the picture of this morbid condition, and studied the causes with much talent; he termed the affection "*congenital spastic rigidity of the limbs.*"

For a long time the researches of Little were almost unknown to neurologists, and it is only in recent works, notably in that of Rupprecht, that they are mentioned with the consideration which they deserve. I shall have more than once to mention the name of Little in the study of this disease, and to quote extracts from the interesting memoirs of Rupprecht and Feer, which are monographs entirely devoted to the disease which we are actually considering.

I have already indicated, gentlemen, the principal characters of this affection, but that you may more fully understand it a detailed study of its SYMPTOMS will now be made.

Since this affection almost exclusively consists in motor disturbances of a *spasmodic nature*, the different movements and attitudes of the patient must be necessarily considered. It is specially when the patient is *walking* that rigidity is most clearly observed in almost all the muscles of the body. Even when the patient is in the upright position, as is the case with this girl whom I now place before you, the following symptoms are observed:—

The *head* and upper part of the *trunk* are inclined forward and move as if they were inseparable. The *upper extremities*

are usually in a state of semiflexion and forcibly applied to the trunk "like the wings of a fowl." The lower limbs, which are usually the most affected, present a truly strange appearance; strictly speaking this may be perceived in all forms of spasmodic paraplegia, whatever its cause may be, but is rarely so characteristic as in this disease. When such a condition occurs the two lower extremities are rotated inwards, and rigid in a

Fig. 31.—Girl suffering from spastic paraplegia (Damaschino collection). She is unable to stand without assistance. The forced adduction of the thighs, the internal rotation of the legs with the position of the foot in talipes equino-varus will be observed.

state of slight flexion at the hip and knee joints. The two thighs are permanently adducted in the most pronounced manner, being often, as it were, fixed together as far as the knees, whilst below the knees the legs are separated by a somewhat large oval space, owing to their rotation inwards. On account of the attitude of the hip and thigh the back is usually also more or less hollowed out, and the muscles in the pelvis and attached to the trochanter, and inner surface of the iliac bones [illegible]newhat rigid. The *feet* usually present, when the motor

disorders are very accentuated, a manifest tendency to assume the position of *equinus* on account of the contraction of the gastrocnemii muscles.

The little girl, upon attempting to walk, presents a *spasmodic* form of *gait* which is absolutely typical. She is seen to draw the point of each foot firmly and noisily along the ground, carrying it forward, as it were, by a semicircular movement,

Fig. 82.—Same girl as in fig. 81, seen in profile. The attitude of the lower extremities and the position of the feet in equino-varus will be remarked.

the foot which remains fixed forming, as it were, the centre of the circle. A *strong inclination* of the body towards the side opposite to that of the foot which is raised will be also observed, an inclination which is repeated in an opposite direction with each step, and thus determines a pronounced swaying movement of the whole trunk, which is specially decided at its upper part. Another special character in the gait of these patients is the more or less precipitate manner in which they advance, or rather are pushed forwards. This is entirely due to the production of a kind of foot clonus when they walk, in such a way that, owing

to reflex contraction of the calf muscles, the patient is thrown forwards at each step at the moment when the point of the foot, as it touches the ground, produces elongation of the corresponding gastrocnemius, which is immediately followed by reflex contraction of that muscle. The knees, and even the thighs, are also seen to be rubbed against each other on account of the tendency to adduction to which I have alluded, and owing to the combination of such adduction with inward rotation, the feet are often found to cross each other, so that a fall seems inevitable if the patient is not supported. This is often the case, but some infants are so accustomed to their infirmity that they become sufficiently adroit to preserve their equilibrium, notwithstanding the extremely bad conditions in which they are placed as far as walking is concerned. It is useless to add that when the disease is severe, walking and leaping are almost impossible.

There is one other attitude in which I would ask you to observe this little girl, because the difficulties connected with the position present singular characters. I mean the *seated position*. It will be observed in what a singular way she maintains that position, if she can do so at all, since she may be regarded as nothing but a solid mass upon the stool where we have placed her. It is certain that the child merely rests upon the seat by means of the ischial tuberosities, the thighs and legs remaining more or less completely extended. This awkward attitude immediately depends upon the rigidity in a state of flexion of the hip joint of which I have already spoken, a rigidity which prevents the patient from giving either to her trunk or legs the inclination which would enable her to remain firmly seated. It is for the same reason that these patients have still more difficulty in sitting upon the ground than upon a chair, since, finding it impossible to flex the thighs sufficiently upon the trunk, they necessarily fall in the reverse direction. It must be added that in some patients the spasmodic rigidity of the muscles of the hip and pelvis is so pronounced that they cannot remain seated upon a chair, and are on that account confined to the bed.

In the *upper limbs*, although the spasmodic rigidity is less pronounced, its existence is obvious. The arms are applied to the trunk, the forearms flexed, the hands in pronation and

Fig. 83.—Attitude of a patient suffering from spastic paraplegia, in the seated position (collection of Charcot). The legs, which cannot be entirely flexed, remain half extended, and hence equilibrium cannot be obtained.

more or less inclined towards the ulnar border. The hands are often extended, while hyper-extension not infrequently exists. As regards the movements of the upper extremities it is easily understood that, owing to the spasms which occur in the muscles producing them, they are difficult to perform, awkward, slow, and accompanied by rigidity of a special kind; grasping objects, supinating the hands, throwing anything to a distance, without mentioning the more delicate movements (writing, sewing, &c.), are amongst those movements which the patients find it the most difficult to execute. There are fortunately a large number of cases in which the disease is of slight or moderate severity, when the upper extremities are quite unaffected or but little involved. Thus, in a somewhat large number of cases *spasmodic paraplegia* can alone be said to exist; this is really the "paraplegia spastica cerebralis" of Heine.

It is not in the muscles of the extremities alone that this spasmodic rigidity occurs. Almost every muscle of the body, those of the trunk and abdomen being included, may, according

to some authors, be equally involved. This statement, taken in a general way, is true, but an exception must be made with regard to some muscles of the trunk, of which the isolated contraction is almost independent of the influence of the will, for in this disease the rigidity specially involves the muscles which are most subject to the action of the will, and, on the contrary, affects but little those of which the movement is more often due to reflex action through the cord than to voluntary incitement. It must be well understood, however, as the following enumeration will prove, that no absolute rule can be laid down upon this point.

The muscles of the *front* and *back of the neck* are often involved and slight extension of the head backwards, or spasmodic lateral deviation are then observed.

Difficulties in deglutition are mentioned by some authors, which are due to the existence of a tendency to spasm in the pharyngeal and œsophageal muscles, and resemble the respiratory and phonic changes known by the name of laryngismus, and which may be attributed to spasm of the laryngeal muscles.

The disorders of *speech* are also frequent, and when the affection is severe the speech is slow, drawling, jerky, and seems "to emerge with difficulty" from the lips of the patient; it must also be noted that a large number of these small patients are very backward in learning to speak.

Another group of muscles of which the functions are embarrassed is that of the *muscles of the orbit*. In more than 30 per cent. of the cases (Feer) *strabismus*, usually convergent strabismus, occurs. Many explanations of its existence have been given. According to some authors it is due to a cerebral lesion. Ziehl believes it to be caused by a simple error of refraction due to malformation of the eye, and hypermetropia is in fact known to be one of the most frequent causes of ordinary strabismus. The researches of Feer on the other hand led him to believe that the strabismus was due to the existence of muscular rigidity in the muscles of the orbit, analogous in character to that which occurs in other muscles. I myself am inclined to look upon the two latter explanations as the most satisfactory. Each of them possibly contains some truth, and not improbably many cases of strabismus occurring in this disease depend upon the association these two causes.

The muscles of the *face* are also liable to the same functional disorders, and on account of the spasmodic rigidity to which they are liable the appearance of the face is changed, and what has been mentioned by most authors is "the expression of stupidity" which the patients have, although in reality their mental power is no way inferior to that possessed by a person of ordinary intelligence. Such is the condition of the patient's face in the ordinary conditions of life, but the immobility and placid appearance of the features may, as I have myself seen, be replaced by exaggerated contraction of the muscles under the influence of passion. Thus for example in the case of a young boy who was very fond of teasing his companions, at this time, or when he became angry, the face was observed by me to assume quite a "diabolical" expression, this being accompanied by a grin which was truly frightful. Little on his part observed a case of "risus sardonicus."

Thus, gentlemen, almost the whole muscular system may be involved, the muscles, however, being affected to a different degree; without becoming notably weaker it is the *inability* and *spasmodic awkwardness* in their movement which specially prevail in this disease.

There is no true paralysis, but at most pseudo-parésis (Strümpell). Add to this that there is *no trace of muscular atrophy*, while at times the more prominent muscles become unnaturally enlarged, and a true idea of the illness will be obtained. In some cases *fibro-muscular contractions* occur which may produce permanent deformity (foot), or an abnormal position of some of the joints (knee), contractions which may also increase the difficulty which exists in using the muscles; contractions of a similar kind may be also observed in the muscles of the orbit when strabismus exists.

Such are the principal disorders connected with motion. It is also an interesting fact that the tetanic rigidity of the limbs diminishes during sleep or even after simple rest; while on the other hand it is increased by fatigue or strong moral emotions (anger, fear).

LECTURE IX.

SPASTIC PARAPLEGIA (*continued*).

Excess of tendon-reflex. Foot clonus. Normal cutaneous reflex action. Common sensibility retained. Muscular sense unaffected. Electrical irritability at times increased. Healthy condition of the bladder and rectum. Absence of intellectual change. Oddness of character. Special indications of degeneration. Attenuated forms of the disease. Tendency to progressive improvement. Congenital onset, often unperceived by the parents. Such children very backward in walking. Their intellectual development usually enables them to live subsequently in the same way as others. Nature of the affection. True spastic paraplegia must be distinguished from the tabid-spasmodic conditions; clinically the first is not accompanied by epileptic attacks, and very rarely by mental change; anatomically it is due to a failure in the development of the pyramidal tract, and not to its destruction; ætiologically it is specially due to premature birth. Diagnosis from infantile spinal paralysis, tetanus of the newly born, tetany, Thomsen's disease, the disease of Pott, transverse myelitis, insular sclerosis, spastic infantile hemiplegia. Hysteria. Treatment: methodical education of the limbs, gymnastics, massage, tenotomy.

GENTLEMEN,—As corollary to the study of the muscular rigidity which has been described in the preceding lecture the character of the tendon-reflexes must now be considered, since in the present state of our knowledge it is by them that we are able to distinguish whether a motor disorder is or is not of a spasmodic nature. In this little girl all the reflexes are considerably increased, the knee jerk is more pronounced, and muscular contraction occurs when the tendo Achillis, the tendon of the biceps or triceps in the arm, or the tendons at the wrist are struck. The deep reflexes are so much increased in this patient that the so-called periosteal reflexes occur, pronounced muscular contraction taking place not only when the tendons, but when the bones, specially those of the forearm, are tapped. The distinct occurrence of foot clonus should be noted in connection with these facts, a phenomenon which, as Pitres and de Fleury have shown, though not in every way analogous to the tendon reflexes already considered, may in all cases be looked upon as a distinctly spasmodic manifestation.

As regards the cases (notably those of idiots) in which tendon-reflexes are said not to have occurred, my opinion is, either that they were not properly sought, and I realize (*vide* the article on "*Hémiplégie infantile*," in the Dictionnaire Encyclopédique) that this examination is at times difficult, or that a mistake was made in the diagnosis, and that the patients in question were not really suffering from spastic paraplegia. In my opinion increase of the tendon reflexes should be looked upon as a constant symptom in this affection.

As regards the *superficial reflexes* little need be said, since they may be found to be in the normal condition, increased, or diminished in different patients without these variations being apparently of the slightest importance.

The *common sensibility* is absolutely unaffected, as you easily observe to be the case in this patient who feels not only when he is pricked but when lightly touched, or the temperature is changed to a very small extent.

The *muscular sense* again does not appear to be changed to any appreciable extent.

The *electrical irritability* of the muscles and nerves is normal as regards its quality; the only change which occurs is in relation to its quantity (tetanoid contraction being produced by the faradic current), and this effect seems only to be produced in isolated cases.

Vaso-motor disorders have been mentioned by some authors, the feet being said to be cold, livid, or marked with a marbling of red colour; these troubles, however, are slight and of no great importance.

The *rectum* and *bladder* are unaffected, or, at most, a tendency to spasm has at times been noted in the sphincters.

The different positive or negative characters of this affection have now been mentioned, but the *mental condition* of the patients must still be considered. Owing to the expression of stupidity of which I have already spoken when studying the aspect of the face, and which you yourselves observed in the little girl whom I placed before you, you would suppose that the mind is in the most unsound condition. This, however, would be a mistake, since the *intellectual faculties* of most of these patients are in quite a normal condition, so that at a later period they can pursue the same studies as healthy children, and adopt the same

professions. At the same time, the intelligence of some patients is but moderate in degree, although in every case it is greater than at first appears. If the intellectual condition, however, is unaffected, it is by no means the same with the *character:* this, in fact in almost every case, becomes most capricious and odd; the patients are usually irritable, violent, disagreeable, and impulsive, being also timid and sly; in short, the character responds to the description which may be given of the mental condition of those in whom general degeneration occurs, of which these patients at times present indications. It is uncertain whether this stamp of degeneration belongs to them, owing to hereditary transmission, or simply depends upon the affection from which they suffer, which, attacking them in early infancy at the time of their development, must necessarily produce in it the most pronounced alterations.

So far, I have carefully placed before you the example of a typical case of spastic paraplegia showing how, in my opinion, the affection should be understood. It must now be stated that the disease does not by any means always present these characters; *attenuated* and even *abortive forms* are not uncommon, and may be said, in fact, to be even more frequent than those which are typical. In these cases motion is not so extremely difficult as was found to be the case in the patients already considered; a certain degree of more or less pronounced rigidity exists in the limbs, which, however, is insufficient to cause much inconvenience; at times, indeed, the rigidity is so slight that it would only be observed by a careful and experienced eye. In almost every case the lower are more affected than the upper limbs; and often in the mild cases are alone involved, slight awkwardness of movement being then alone observed in the upper limbs.

It must be remembered that the tendency of this disease is to improve during its course; thus when in infants the four limbs are involved to a more or less pronounced extent during the first years of life, the motor disorders progressively diminish and the upper limbs become almost completely sound.

As regards the *onset* of the affection, since it dates from the time of birth, nothing need be said about it from a clinical point of view. At the same time, as you would expect, it is not during the first days of life that the diagnosis is made; this usually

occurs at a later date, as at the end of a few months when the parents remark that the movements of the child are not entirely free. Thus when he is bathed, instead of kicking his legs in every direction, as healthy children do, he keeps them tightly pressed one against the other, without ever completely flexing or extending them, or at a still later date, when he is eighteen months or two years old, they are astonished not to see him walk. It is the rule in fact, gentlemen, in this affection that a long time elapses before the little patients make the first step, which they succeed but imperfectly in doing, and another long period passes before their attempts to walk are crowned with success. In some cases one of the symptoms which points to the existence of this affection is the inability of these patients to remain in a seated position for reasons already mentioned. The parents alarmed by these signs of evil augury then determine to consult the physician. It is not surprising therefore, as has been already observed, that these little patients are so often seen for the first time at a somewhat long period after the time of their birth.

These observations may be made about the onset of the disease, while subsequently, as you know, the spasmodic rigidity may in some cases diminish or even entirely disappear. You will ask me, however, what becomes of these patients, and what their subsequent existence will be? Except in the case of those who are quite infirm, their existence will not be different from that of others. They may certainly be occasionally in worse circumstances at college, in the same way as is the case with any whose moral and physical development is below the average, and during life their infirmity is often enough the cause of some disappointment the eccentricity of their character may increase. With these exceptions the life of such children is of the ordinary kind; they join the same classes as other children of their age, pass through the same examinations, and belong subsequently to the same professions, those cases alone being excepted in which some special apparatus is required in order that the muscles may be used. They grow in the same way as other children, the structures and functions of the body develop in the same manner, and puberty is reached in the same way as that of their companions. Subsequently they marry, and their children are quite free from the disease which affects them,

spastic paraplegia *as you know not being a hereditary disease.* The duration of their life is of good length, and quite independent of the illness from which they suffer.

I have dwelt upon these facts in order that you may not forget that spastic paraplegia, which so far I have only put before you as it occurs in children, exists also in *adults* since the affected children reach the adult age. For the same reason it may be observed in *old age*, though the fact must not be forgotten that it has existed from the earliest years of life.

These being the clinical characters of the affection, what is its pathology? what position should it occupy in nosology? what is its true nature? This is by far the most difficult question connected with the disease, and as I explained at first totally different opinions have been held upon this point, which is far from being settled.

The cause of this difference is, in my opinion, the attempt which has been made to unite things which are quite dissimilar. If in fact it is true that an affection, sui generis, exists, which so far we have alone had in view in the description given, it is equally certain that other very analogous conditions may be observed of more or less general muscular rigidity. These conditions present almost exactly the same characters as those which have just been mentioned, but are usually associated with more or less pronounced *disorder* of the *intelligence*, and sometimes with *attacks of epilepsy*. It is also in these cases, for which I propose the name of *tabid-spasmodic conditions*, that the *malformations* of the skull are found, which are mentioned by many authors. We will first consider the *ætiology* of these different morbid conditions. It is to Little, who initiated this branch of clinical study, that we owe the knowledge of two most important facts connected with the ætiology of these spasmodic conditions: *premature birth*, or the *existence of a severe labour*. Such, gentlemen, are in fact the two great causes of the morbid conditions which we are considering, and to these must be added the *influence of certain inflammatory diseases of the fœtus* or *newly-born child*, which give rise to lesions in the meninges or brain.

The ordinary lesion in every case is *non-development of the pyramidal tract* throughout its whole length; it should be noted, gentlemen, that I do not say its degeneration. We have already mentioned as one of the characters of this affection the fact of

its being *congenital*. Now it is known that the development of the sheaths of myelin in the pyramidal tract takes place at a very late date, viz., at the end of the 9th month of fœtal existence, and that the pyramidal tract is not completely developed until a still later date; it is not, therefore, surprising that when a child is born before the proper time, as in the 7th or 8th month for instance, the development of this tract is more or less completely arrested. Again, in cases of severe labour with long duration, when compression of the head occurs during a somewhat considerable length of time, instruments being often used, it is easy to understand that the brain may be injured. Whether, as Little supposed, hæmorrhage from the capillary vessels occurs in the brain and spinal cord owing to the manual operations rendered necessary by the labour, and accompanying asphyxia, or from the occurrence of some other change is uncertain. We may, however, join Ross and Feer in remarking that in the brain of newly-born children, which is so soft as to be almost diffluent, injury or pressure, wherever they occur, necessarily affect most severely the part which has the least power of resistance. This is the pyramidal tract, the part which is the last to develop, and in which the sheath of myelin, being as yet unformed, cannot protect its fibres in the same efficient way as those in other parts of the cerebro-spinal axis.

As regards the third cause mentioned, viz., the occurrence of an *inflammatory affection* in fœtal life, or during the first few days after birth, and which gives rise to *meningitis* or *encephalitis*, which may or may not be followed by *porencephalia*, and in every case much interfering with the development of the pyramidal tract, I confess that I much hesitate to give it a place here. It is certainly true that these cases are often accompanied by spasmodic rigidity similar to that which has been described as characteristic of spastic paraplegia, but they usually present also special symptoms which clearly distinguish them from the cases which are due to premature labour. These symptoms are, on the one hand, *intellectual change*, which may reach complete idiocy; on the other hand *convulsive attacks*, which have been often noted, and which in my opinion opposed, however, to that of Feer, are merely due to *epilepsy*. In cases due to premature birth, which I have exclusively taken

as the type of my clinical description of spastic paraplegia, a fact which I consider important, no notable intellectual change or epilepsy exists. This difference in the symptoms presented justifies, in my opinion, a complete separation between the two affections.

As regards the cases which are due to severe labour I am not equally certain, nor can I say whether intellectual change and epilepsy are of frequent occurrence, or rare, or completely absent in these patients, and am therefore unable to decide whether, from a symptomatic point of view, they should be placed with those due to premature labour or not.

I should thus be inclined to isolate all the cases in which it is most improbable that any arrest in the development of the pyramidal tract occurs, cases which notably differ, as we have seen, from a clinical point of view, owing to other symptoms of extreme gravity (intellectual change, epilepsy) being associated with the disorders of motion. I should give the name of *true spastic paraplegia* to the cases which are due to premature birth, with arrested development of the pyramidal tract, while the other cases might be termed "*tabid-spasmodic conditions*" whether due to injury at birth, or to destruction of the motor region of the brain by an inflammatory disease (the most often infectious).

The advantage of making this distinction is that no confusion then exists between two groups of affections which differ from each other in many respects. As regards pathological anatomy, one of the groups, that of the "*tabid-spasmodic conditions*," consists of the destruction of a portion of the brain by encephalitis or meningitis, destruction of which the seat is not fixed, and which may only occur at the origin of the pyramidal tract, or involve other portions of the brain as is usually the case. This explains the frequency with which intellectual change occurs in this form of disease, being also the cause of the convulsive attacks, since, owing to its inflammatory nature, the lesion constitutes, as it were, a thorn within the brain, whose irritative tendency shows itself at a later date in the attacks of epilepsy. The other group, represented by one disease alone, viz., *spastic paraplegia*, is simply an arrest of development, exclusively affecting one system of fibres. It is, in fact, essentially a *systemic* disease, and it is on this account that the name of

spasmodic tabes dorsalis was expressly given by Charcot to this *primary and isolated degeneration of the pyramidal tract.* It will now be understood, gentlemen, how totally distinct these two diseases are, having in common but one symptom, the *congenital spasmodic rigidity.* This should be regarded as a mere coincidence, it being remembered of how many different affections another symptom, hemiplegia, is the indication, and it must be admitted that spasmodic rigidity has by no means the same meaning in all cases, and that these must therefore be necessarily separated from each other.

The varieties of muscular rigidity having been distinguished, the DIAGNOSIS of the affection must now be made by comparing with it other affections which resemble either spastic paraplegia or the tabid-spasmodic conditions.

Infantile paralysis has in reality no characters in common with this affection since in it the muscles are essentially flaccid, whereas in this disease the muscles are in a state of spasm.

Tetanus neonatorum need not be discussed at length, bearing but little resemblance to spastic paraplegia; it will also be remembered that it begins in the face and is rapidly fatal.

Tetany is a contracture with varying manifestations which do not resemble the spasmodic rigidity which has been so fully discussed in this lecture.

Thomsen's disease might possibly give rise to a mistake since in this affection also there is muscular rigidity which may inconvenience the free use of the limbs. It will be remembered, however, that in Thomsen's disease the rigidity only exists at the commencement of voluntary movements, whereas in spastic paraplegia the reverse is usually the case. Again, in Thomsen's disease what is so characteristic of the affection we are now studying, viz., the tendency to permanent spasm, the great increase of the tendon-reflexes and foot-clonus are notably absent.

The only affections which in my opinion it may be difficult to distinguish from this disease are those of which the symptoms include the spasmodic features which are invariably present in spastic paraplegia. Amongst these affections the first to be considered are :—

Potts' disease and the different varieties of *compression of the cord.* These are distinguished by their course, by not being

congenital, by the deformity of the spine, and by the frequent occurrence of pseudo-neuralgic pains (Charcot), &c.

Transverse myelitis, whatever its cause may be, very possibly, if chronic, bears a similar resemblance, specially in the adult. Its etiology suffices to distinguish it; the fact that the affection appears at a later, even at the adult age, proves that the disease cannot be spastic paraplegia.

The same may be said with regard to *insular sclerosis*, in which, as will be seen, the spasmodic symptoms often prevail to a special degree. Other symptoms, however, and the most frequently many of them, such as the tremor, the peculiar change in articulation, nystagmus, &c., enable this disease to be at once recognised. Lastly, there is one affection of a clearly spasmodic character with regard to which the diagnosis may be most difficult, namely, *spastic infantile hemiplegia*. I do not refer, it must be understood, to infantile hemiplegia associated with tonic contraction and pronounced atrophy of the limbs, which could not fail to be recognised, but to those forms of hemiplegia which are unaccompanied by true paralysis, but in which the movements are performed with some difficulty and attended by the pronounced occurrence of spasm. The clinical aspect then much resembles that of spastic paraplegia, and to such an extent that some of the cases described by me recently under the name of tabid-spasmodic conditions should be referred in my opinion to spastic infantile hemiplegia. These would be cases of incomplete bilateral hemiplegia due to lesions seated in both the cerebral hemispheres but involving the pyramidal tracts to a moderate degree. Having sufficiently dwelt upon the clinical aspect of that affection I need not return to the subject.

The facts recently observed by Bernhardt* should be mentioned in connection with the diagnosis of the disease. This author observed the existence of all the symptoms of spastic paralysis in many members of the same family at the conclusion of the 30th year of life, without its being exactly known to what affection these symptoms were due.

This chapter must be ended by my once more putting you on your guard with respect to *hysteria*; both in the adult and child paraplegia may occur associated with tonic contraction, which

* Bernhardt, *Beitrag zur Lehre von den familiären Erkrangkungen des centralnervensystems* (*Virchow; Arch.* 1891, 126, *p.* 59).

closely resembles that existing in spastic paraplegia, but which is really due to hysteria alone. These cases, when once known to occur, may be easily recognised by the facts that the tendon-reflexes are much less, if at all, increased, that most often the sensibility is also more or less deranged, and these combined with the other characteristic symptoms of hysteria enable this disease to be recognised.

The treatment which should be employed must now be considered; that which I would recommend, though apparently not of much energy, will undoubtedly produce appreciable results. The knowledge which we have of the nature and lesions of spastic paraplegia enables us to know that we cannot act directly upon the spinal cord, and that the cautery, blisters, heated points, and other counter-irritants, are as useless as they are painful. The development of the pyramidal tract cannot be rectified. At the same time, the fibres of the pyramidal tract whose evolution is slightly incomplete may be made to act in a proper manner. With this object *methodical education* of the limbs, *gymnastic exercises*, *massage*, and *passive movements* with or without preventive tenotomy will be your chief assistants in the treatment. Should any doubt exist as to the value of such treatment, or the mode of practising it be unknown, it may be seen in the practice of Dr. Bourneville at the Bicêtre, that, by means of this method carried on with perseverance and devotion, much improvement may be obtained in cases of spasmodic cerebral pseudo-paralysis, even when complicated by more or less pronounced idiocy.

LECTURE X.

INSULAR SCLEROSIS.

HISTORY: Cruveilhier, Rokitansky, Charcot and Vulpian, Ordenstein, Bourneville and Guerard, recent works. SYMPTOMS: 1. SPINAL. A. *Motor*. Disorders in the gait, true spastic gait, pure cerebellar gait, cerebello-spastic gait; their characters. Spastic paraplegia. Hemiplegia: characters of the hemiplegia in insular sclerosis. Tremor: its characters; occurs when voluntary movements are made, moderate rapidity of its rhythm, specially affects the muscles at the root of the limbs, but often also those of the trunk and neck, and may predominate upon one side of the body.

GENTLEMEN,—It will certainly have been observed that at the beginning of each lecture devoted to the study of a disease, I am careful to mention the names of those to whom we should be grateful for having discovered, or described, or simply studied it better than their predecessors, for having, in fact, in some way increased our knowledge. These names are usually few in number, since by no means all the works written about the disease are connected with its history, and it is a *historical account*, and not a *bibliographical index*, that I wish to give you at the commencement of the chapters. Another reason, on account of which this enumeration may appear small, is that I reserve for myself the power of indicating, as the lectures continue, the most recent works and the progress for which we are indebted to them. It will not, therefore, be surprising if at times the history of a disease is almost entirely limited to what may be called the "heroic periods" of neuro-pathology.

Such, with your permission, will be the case with regard to *insular sclerosis* and other affections which will be considered by us.

The *lesions* of insular sclerosis are undoubtedly seen in Cruveilhier's admirable atlas of pathological anatomy, in that of Carswell and in the treatise of Rokitansky. Many observations upon this affection were published by Türck, Frerichs, Rindfleisch, &c., but it was only regarded by them as a pathological curiosity and the account of the autopsies was alone given.

It is Charcot and Vulpian who deserve the honour of having given to insular sclerosis the dignity of an anatomical and clinical existence in 1866, and of describing in a masterly way the symptoms and pathological anatomy of the disease. The thesis of Ordenstein, inspired by Charcot, was written in 1867 and lastly in 1869 the memoir of Bourneville and Guerard appeared, a monograph of real value, since which time insular sclerosis has had a real existence. The works which have been recently written will be mentioned in the course of these lectures. What are the SYMPTOMS of the affection? These are usually divided into spinal, cerebral, and bulbar symptoms. This mode of arranging them is undoubtedly the most convenient and will be employed in these lectures, but it should be stated that this is merely an artificial proceeding, and that with regard to many symptoms it would be difficult to say in what part of the nervous system they originate.

The *spinal symptoms* will be first considered and studied in the following order:—

I. SPINAL SYMPTOMS.

A. MOTOR SYMPTOMS.

Amongst the disorders connected with motion the disorder of the *gait* must be specially mentioned. This assumes various forms which are quite distinct from each other, and of which the principal are the following:—

α. The form which is *clearly spastic in character.*

β. The form which is *purely cerebellar.*

γ. The cerebello-spastic form.

α. The first of these forms, which is clearly spastic in character consists of more or less pronounced paraplegia with tonic contraction and strongly marked rigidity, in short, of the same *spastic gait* which has been already described as occurring in spastic paraplegia and which will be also found to exist in other diseases. It will suffice to recall the principal features of this condition; the legs are extended and pressed against each other and the feet seem adherent to the soil so that the patient finds it extremely difficult to walk or to bring forward the foot which was previously behind the other. Since the rigidity of the leg prevents flexion of the knee the progression of

each foot is accomplished by elevation of the corresponding half of the trunk and pelvis, and even then the foot is rubbed against the ground with such force that a noise is produced which alone enables the character of the gait to be recognized from afar off. To these phenomena the other so-called "spastic" symptoms must be added. These specially consist, as you know, in increase of the tendon-reflexes (patellar tendon, Achilles tendon), and foot-clonus, the latter being often so pronounced as to occur spontaneously whenever the ball of the foot rests upon the ground during the patient's efforts to walk, to which this condition presents an additional obstacle.

In the *purely cerebellar* form (β), which certainly but seldom occurs, the appearance is quite different: what predominates is the uncertainty of the gait and the loss of equilibrium, while the movements are all free and the joints can be used without difficulty; the direction, however, given to the limbs is doubtful and uncertain, while the gait is unsteady, resembling that of an intoxicated person. In this form the tendon-reflexes are not excessive, being, on the other hand, somewhat diminished.

The *cerebello-spastic* form (γ) is by far the most frequent and characteristic form of insular sclerosis. As its name, proposed by Charcot, indicates, the characters of the cerebellar and spastic gait are associated together. The appearance of the patients is as follows: the feet are separated in the same way as those of seamen upon the bridge of a vessel, and the ball of the foot is not placed alone on the ground as in the purely spastic form, but the whole sole of the foot, and specially its posterior part. Hence the heaviness of the gait and the noise produced by the heels are explained since the patients do not place, but strike their feet against the ground. Their steps are irregular, the irregularity showing itself in different ways. As regards *rhythm*, *length*, since the steps may be longer or shorter, and *direction* which never remains absolutely the same, these patients walk in such a reeling manner as to present the appearance of an intoxicated person. These "cerebellar" phenomena are still more pronounced when the patient attempts to stop suddenly, or to turn rapidly round. The spastic phenomena are shown by the fact that the feet seem glued to the ground, on account of the difficulty with which the legs are moved, while the tendon-reflexes are more or less in excess, and foot-clonus often exists.

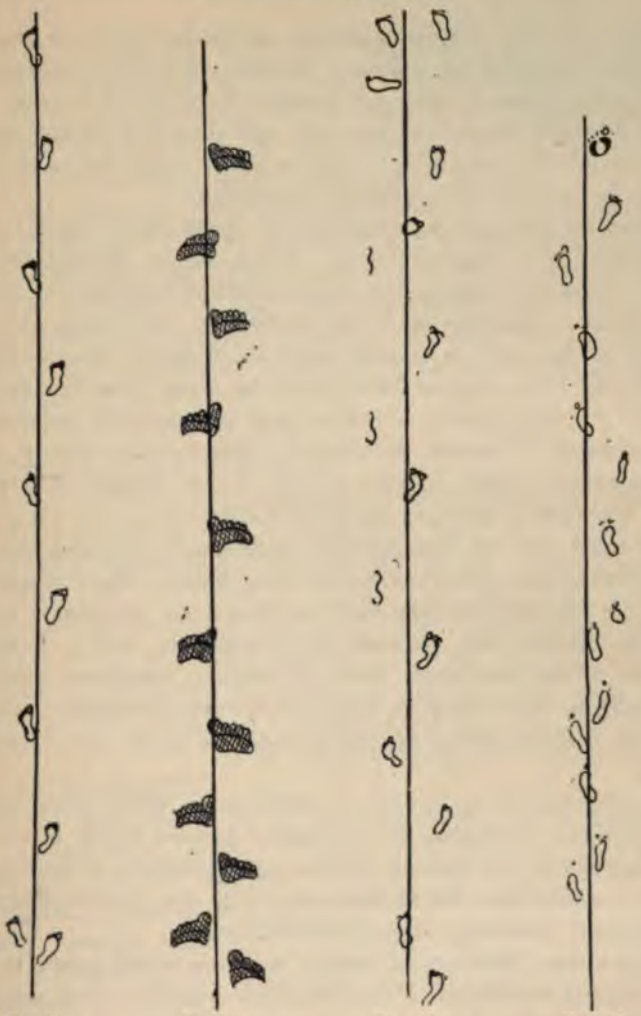


Fig. 84. Fig. 85. Fig. 86. Fig. 87.

Fig. 84.—Sketch of the normal gait according to Gilles de la Tourette. The steps are of equal length, the feet maintain the same direction in the different steps, and the same distance from the line of progress.

Fig. 85.—Diagram representing the gait of a patient suffering from insular sclerosis. Spastic gait. The steps are short; the ball of the foot alone touches the ground, the feet are turned inwards and have a tendency to cross each other (overstepping at times the median line, which represents their progress).

Fig. 86.—Sketch of the gait of a patient suffering from insular sclerosis (after Gilles de la Tourette). Cerebellar gait. The steps are very unequal in length, the direction of the feet varies in the different steps and they do not remain at the same distance from the median line, indicating the direction of their progress.

Fig. 87.—Sketch of the gait of the same patient, as fig. 86 (after Gilles de la Tourette). Cerebellar gait. The irregularity is even more marked than in the preceding sketch.

Such are the different aspects which the gait of patients suffering from insular sclerosis presents; it is obvious that the description which I have just given will only show in what way this symptom should be observed, and does not by any means include all the cases which may be seen; many forms of transition in fact exist and different associations.

Thus, for instance, walking may be much embarrassed by the existence of a general tremor which exists throughout the whole body and renders the maintenance of equilibrium almost impossible. Oppenheim, in an interesting work upon the disorders of the gait in insular sclerosis, regards these cases as belonging to a distinct form which he terms "vacillating"; I should myself prefer, in order not to multiply indefinitely the number of forms, to consider this tremor merely as a phenomenon added to one of the forms already described; this, however, is merely a question of detail.

It must not be thought that disorders in the gait are the only symptoms observed in the *lower limbs*. Their movement may be affected in every way, so that true *paraplegia* exists, which obliges the patients to remain in bed. Extensor spasm of the two lower limbs is usually associated with the paraplegia, this being in fact but a more pronounced degree of the disorder which we have just studied as the "spastic" gait.

As regards the *upper limbs* a condition is often observed, not of paralysis, or spasm, but of spastic paresis, which often presents itself in the form of extreme awkwardness of movement, an awkwardness to the existence of which the tremor, which we shall study presently, largely contributes.

The motor disorders of insular sclerosis would again be but imperfectly understood if the *hemiplegia* which is often observed, and has peculiar characters, was not mentioned.

The *hemiplegia* which occurs in insular sclerosis was mentioned in some of the earliest observations regarding the disease; Charcot used to call the attention of his pupils to this symptom, and I have myself had the opportunity of seeing several examples of the condition in his service, of which one was extremely pronounced, and will be found described in the Thesis of Babinski. The latter author as you know devoted an important chapter in his Thesis to the description of this symptom, but the

fullest description of hemiplegia occurring in insular sclerosis is undoubtedly found in the Thesis of Miss Blanche Edwards. The conclusions at which our distinguished colleague arrived show that this symptom not uncommonly exists in the disease; that it usually occurs after an attack of apoplexy; that such an attack may take place two, three, four, or more times in the same person, and may or may not be accompanied by aphasia; lastly, that the hemiplegia in insular sclerosis is usually a transient symptom, and may completely disappear.

As regards the anatomical changes to which this hemiplegia is due, they are as yet unknown, and the course of the symptom alone enables us to recognize that the fibres of the pyramidal tract are not destroyed by the lesion, and that no secondary degeneration is associated with it.

To give an idea of the frequency of its occurrence, I should say that I myself found it to exist three times in thirteen cases of insular sclerosis in which I searched for it. The hemiplegia may affect the limbs alone, or the limbs and lower part of the face simultaneously; more rarely crossed hemiplegia occurs, lastly in some cases the face is alone affected, the muscles supplied by the lower branches of the portio dura being alone paralysed. The nature of the hemiplegia will be discussed at a future time, and you will see, gentlemen, that most often it is of hysterical origin.

The *tremor*, which is one of the most frequent and characteristic symptoms of insular sclerosis, must now be studied, of which the clinical analysis made by M. Charcot may be considered a model of its kind.

With your permission this symptom will be considered in connection with patients suffering from this disease who will be put before you.

The present attitude of these patients does not show any tremor to exist; they are seated quietly in their chairs, and no movement or effort is being made. This in fact, gentlemen, is one of the characters of the tremor in insular sclerosis, *that it does not occur while the patient is at rest*; upon drawing one of the patients forward so that he does not lean against the back of his chair, the upper part of the body is at once seen to oscillate. It is however specially during *voluntary movements* that the tremor occurs, as Charcot has shown, and hence it is

termed "*intentional tremor*." The phenomenon will be again witnessed if one of the patients is asked to take the glass which is upon this plate and carry it to his mouth. The hand seizes it somewhat abruptly so that the liquid contained in the glass is shaken; the glass is then raised from the plate, and let us examine the progress which it makes:—

The oscillations of the hand are at first small and slow, but as the glass approaches the mouth increase both in size and to a small extent in rapidity; the water in the glass is agitated and tossed in every direction, the face and clothes of the patient are drenched, and those near the patient must stand aside, or

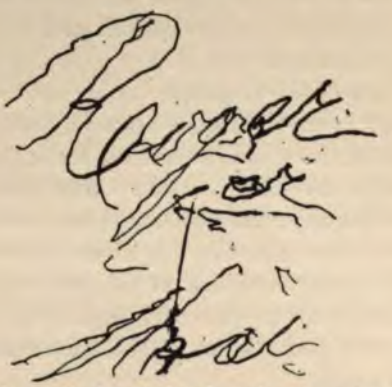


Fig. 88.—Writing of a patient suffering from insular sclerosis (service of M. Charcot).

will suffer in the same way. It is now when the hand has nearly reached the mouth that the tremor becomes considerably increased, and the oscillations of an extraordinary size [20—30 centimetres (5—7 inches), or more] and far more rapid; it will also be observed that the upper part of the patient's body which had inclined forward to meet the glass trembles in a similar manner to the hand, so that the head is moved by a series of rhythmic salutations.

An audible sound is produced when the mouth and the glass meet, and this explains how it is that at times the teeth of the patient are broken when he attempts to drink. It will be noticed, however, that in this case the glass, displaced by the oscillations of the hand, has escaped from the lips, which were unable to seize it, and carried away by its uncontrollable

movements resembling those of a pendulum, strikes at one time the nose, at another the chin; wearied by his efforts the patient has seized the right wrist with the left hand, and thus fixes the right forearm strongly against the chest and prevents it from oscillating, whilst at the same time he carefully keeps the glass in place by means of the lips and teeth, and thus with the head inclined forward succeeds in drinking a small quantity of the liquid which remains therein.

Now that the patient is at rest again there is no longer any evidence of tremor, this only occurring, as already explained, *when voluntary movements are made*. Some conditions increase this phenomenon, and these should be known that it may be understood how to produce tremor in a patient suffering from this disease. These conditions are: 1. The *extent of the movement;* thus, when a patient is directed to take hold of an object, as was done in this case, he should be placed at some distance from it in order that he may be obliged to extend his arm. 2. The *existence of emotion;* thus, the idea of being observed, or if the glass presented to them contains a liquid, the fear of upsetting it suffices in these patients to considerably increase the tremor.

It is found, when the tremor itself is studied, that from the nature of its rhythm it belongs to the tremors with *moderate rhythm* (those in which from $5\frac{1}{2}$ to $7\frac{1}{2}$ oscillations occur in a second). This fact, as I observe, gentlemen, surprises you since the tremor seems to be much more rapid than I say. You are quite right, though I have made no mistake. In reality the movements in insular sclerosis seem to be far more rapid than they really are, the cause of which is that the movements are of somewhat large size. A simple experiment will prove this: in the left hand I hold a ruler 30 centimetres (1 foot) in length, in the right hand a rod 1·5 metres (5 feet) long, and with both of these I strike the table with equal intervals, as shown by the sounds produced, the only difference is that the free extremity of the ruler is moved a much shorter distance than that of the rod. Upon watching the two instruments the movement of the rod is seen to be very much more rapid than that of the ruler (since its size is greater), although in reality the number of oscillations occurring in the two instruments is identically the same. This, gentlemen, is what occurs with respect to the tremor in insular sclerosis, but a moderate number

of oscillations occur, though they are of such a size as to give at first sight the impression that they are very rapid.

To what is the extent of these movements due? To this fact, that the tremor in insular sclerosis is not limited as in many cases of tremor, to the extremity of the limb, or to the muscles of the forearm, the flexors, and extensors of the wrist, but, as will be seen, affects almost all the muscles of the body; it may almost be said that it is specially seated in the muscles of the trunk and in those of the scapular and iliac girdles. Since, on account of having this position the tremor principally occurs near the root of the limb (as in the shoulder and elbow joints), you will understand, gentlemen, that its extent would be therefore multiplied by the whole length of the lever which the arm and forearm constitute for it, and becomes thus of considerable length.

Such, gentlemen, are the observations which I wished to make with regard to tremor in the upper limbs. As you know, the tremor is not confined to these, but occurs also in the *lower limbs*, where it is comparatively but little pronounced. The *trunk* is also liable to move violently forward or backwards when the patient places himself in an upright position, or attempts to walk, or even when he is seated, or in bed if insufficiently supported. Some patients in whom the movements are very pronounced must be "steadied" on every side before they disappear. The *neck* and *head* also move forward and backwards when an effort is made to stand in an upright position, or to walk, or, as was just observed, when the patient carries something to the mouth. In very pronounced cases the tremor in the head tends not only to occur in an antero-posterior direction but to produce, as it were, a movement of circumduction.

In some observations the tremor has been said to be *unilateral*. This, however, is rare, although it is often found to *predominate* on one side of the body, so that when this symptom is examined care must invariably be taken to examine the hands upon both sides.

CHAPTER XI.

INSULAR SCLEROSIS: SYMPTOMS.

B. *Sensory symptoms.*—Disorders of common sensibility (Freund) are rare, or but slightly pronounced: they vary in form from tingling to hemianæsthesia. Disorders of special sensibility: the hearing, taste and smell are little and rarely affected. Disorders of sight of great importance: works of Gnauck, Parinaud and Uhthoff. Nystagmus and nystagmoid movements: precautions necessary in seeking them. Paralysis of the muscles of the orbit, associated paralyses (Parinaud). Disorders of the optic disc, myosis associated with excess of iris reflex. Changes in the optic disc, disorders of sight which result from them, changes in the field of vision. Variable onset of visual disorders, absence of symmetry in them.

C. *Visceral symptoms* connected with the stomach, rectum, bladder, and genital organs (Oppenheim); these are rare, and but little pronounced.

D. *Trophic disorders*, specially amyotrophic.

II. BULBAR SYMPTOMS: disorders affecting deglutition, or mastication, trembling of the tongue, glycosuria, polyuria.

III. CEREBRAL SYMPTOMS: affections of speech, the articulation is slow, monotonous, scanning, spasmodic. Vertigo: intellectual disorders, impulsive laughter, apoplectiform, epileptiform attacks.

GENTLEMEN,—In the preceding lecture it was observed that many forms of disorder occur which are connected with motion; the effects of the disease upon *sensation* will now be discussed.

A.—COMMON SENSIBILITY.

Usually symptoms connected with common sensibility are rightly considered to be absent in insular sclerosis; at the same time, in looking through the different observations upon this subject which have been published, it is found that sensory disorders do at times occur (Charcot, Schüle, Berlin, Engesser, Vulpian, Oppenheim, &c.); recently Freund (of Breslau) has specially and completely investigated this subject.

Among the *subjective* disorders of sensibility *numbness*, *tingling*, *abnormal sensations* of *heat* and *cold*, and sometimes true *pain*, which may be somewhat diffused or lightening in character, or a

sense of constriction round the trunk may exist quite analogous to that which occurs in tabes.

The *objective* disorders consist, according to Freund, in abnormal *tactile sensibility*, a *sensation of compression*, *analgesia*, *hypalgesia* or *hyperalgesia*; various forms of *anæsthesia* may also be observed, disorders of the *sensibility to temperature*, or *muscular sense*, the latter being more rare; in some cases *hemi-anæsthesia* has been noted.

The sensory disorders are specially seated at the extremities of the limbs, viz., in the *fingers* and *toes*; they are but *slightly pronounced*, of *temporary duration*, *and liable to change*, having only been permanent in six out of thirty-three patients suffering from insular sclerosis (Freund).

These facts will not be discussed at present, since the relations between insular sclerosis and hysteria will only be considered at a later period. At the same time I would repeat what should notwithstanding this be kept in mind for daily use, the idea first mentioned by Charcot, *that sensory disorders do not form part of the clinical features of insular sclerosis.* I would not certainly deny that they may occur, but in my opinion this happens so rarely that they should merely be regarded as a clinical curiosity.

B.—Special Sensibility.

Different changes have been noted in hearing, notably a diminution of this function which may increase to complete deafness. The same may occur as regards *taste* and *smell*, but the troubles of these senses are in reality but rare and seldom at all pronounced in degree. As regards *sight* it is quite otherwise; the disorders are frequent, often much accentuated, and of great importance as regards the diagnosis of the disease. These symptoms will therefore be studied in detail. To facilitate this everything connected with the organ of vision will be now considered, its motor functions being included.

The principal symptoms associated with the *visual function* have been long known, and are mentioned in the ordinary manuals connected with this subject. During the last few years, however, attention has been again directed to these symptoms, and to this we owe the important works of Parinaud, Gnauck

and Uhthoff, works from which I shall largely quote in the description which I am about to make.

Almost all the structures of the organ of vision may be notably affected in insular sclerosis.

Amongst the disorders there is one which is specially important as regards the diagnosis of the disease, namely *Nystagmus*, which consists as you know, gentlemen, of rapid oscillations of the eyeball produced independently of the will of the patient and usually without his being aware of its occurrence. The movement always occurs in a *horizontal* direction from left to right and *vice versa;* Uhthoff, however, states that he once saw it occur in a *vertical* direction, and in two cases seen by the same author the patients believed that they saw objects move.

Nystagmus is sometimes so pronounced as to be observed at once when the attention is fixed on the eye, but at other times, is scarcely perceptible and must be carefully sought.

To do this the eye of the patient should be placed in a *strained position*, as for example by his being directed to look at the finger which is placed at some distance on the outer or inner side of the eye, without turning the head so that the eye is as near as possible to the outer or inner angle of the eyelids. The eye being thus placed in a "strained position" the rhythmical movements will be perceptible which cannot be seen in the ordinary position of the eye. This artifice should be employed in such cases, upon the utility of which Parinaud has justly insisted for many years in his conferences at the Salpetrière Hospital.

Uhthoff distinguishes two varieties of ocular movement:— 1. those which belong to *true nystagmus*, and are directly due to a lesion of the brain or medulla oblongata; 2. the so-called *nystagmoid oscillations*, which are not true nystagmus and only exist in strained positions of the eye, being due to a condition of slight paresis, involving the nerves of the ocular muscles. In 100 cases of insular sclerosis this author found true nystagmus in 12, nystagmoid oscillations in 46 cases, that is to say, rhythmical movements were found to occur in 58 per cent. of the cases. These numbers must be accepted since I know of no others to compare with them, but at the same time I must acknowledge that the number appears to me somewhat too small,

and my own impression is that from 70 to 80 per cent. are thus affected, since this is one of the symptoms most commonly observed in insular sclerosis.

Other disorders may occur in the action of the ocular muscles in this disease, viz., *paralysis of the muscles of the orbit.* Though not as frequent as nystagmus this symptom is also of great importance as regards the diagnosis of the disease (it occurs 17 times in 100 cases—Uhthoff). The condition is far more one of paresis than of true paralysis, and the paresis is partial, incomplete, and transitory, which causes it to have a characteristic nature. *External ophthalmoplegia* (paralysis of all the muscles of the orbit) may also occur, but the *6th nerve* is principally affected in this disease, either on one or both sides, as is also the *3rd nerve.*

According to Parinaud the motor disorders of the ocular muscles in insular sclerosis almost exclusively consist of paralysis or paresis of associated muscles, that is to say, both eyes are simultaneously affected, and the muscles which act together in producing a certain movement are involved.

Nystagmus is essentially of this nature. It is a tremor which indicates paresis of such *associated muscles.* The extent of the muscular movements is invariably found to be diminished when the nystagmus is pronounced. The lateral movements are usually affected, but all the muscles may be paralysed, and more or less pronounced *external ophthalmoplegia* then exists. According to the same author paralysis limited to three pairs of nerves, such as occurs in tabes and syphilis, is on the other hand very rare.

The natural consequence of such ocular paresis, if sufficiently pronounced, is *double vision,* which, however, is most often transitory, only occurs in certain positions of the eye, and corresponds, in fact, to the ocular paresis which produced it. Another consequence of this paresis is that the appearance of the patient is often characteristic, having been well described by Charcot under the name of "vague expression."

The action of the *iris* may be disordered in different ways (11 per cent. of the cases, Uhthoff). Thus *myosis, inequality of the pupils,* or a *diminution of its action under the influence of convergence or light* may occur. Parinaud specially insists upon the fact that a nervous affection in which myosis with retention,

and perhaps excess of the light reflex, as regards the rapidity of its production, exists, should be suspected to be insular sclerosis and not tabes.

The condition of the fundus oculi and ophthalmoscopic examination of the *optic disc* must now be considered. In the first descriptions made by Charcot the existence of atrophy of the optic nerve was noted in certain cases. This observer also noted the fact that such a change did not always occur, notwithstanding the existence of pronounced amblyopia, and that the lesions in the disc never corresponded in intensity to the loss of sight. Since this time Gnauck, Uhthoff, and the principal ophthalmologists who have considered this question have mentioned these facts in detail, and made interesting deductions therefrom.

The changes in the optic papilla may, according to Uhthoff, be referred to three different degrees of alteration :—

A. There is atrophy and complete decoloration of the papilla, the reddish reflection of the normal papilla having entirely disappeared.

B. There is incomplete decoloration of the whole papilla, the internal portion of which still preserves to a certain extent its red reflection.

C. The external, temporal, part of the papilla is alone discoloured, the tint of the internal portion being in every respect normal.

D. There is the appearance of *true optic neuritis* (Parinaud, Uhthoff, &c.), hyperæmia, cloudy appearance of the papilla, vessels concealed and dilated, papilla prominent, &c. This condition perhaps is merely one of the first phases of the process which leads to atrophy and decoloration of the optic papilla.

As you see, gentlemen, the changes in the fundus oculi are clearly marked in insular sclerosis, they are also frequent, Uhthoff having found that the fundus oculi was only normal in 48 out of 100 cases, while the sight was diminished in 5 of the apparently normal cases. Thus this author justly remarks that "cerebral tumours and tubercular meningitis being excepted there is no disease of the nervous system (tabes being included) which is so often accompanied by ophthalmoscopic changes as insular sclerosis."

Such, gentlemen, are the lesions of the optic nerve where it

expands to form the retina, and the symptoms due to these lesions will now be mentioned. As I have already said when

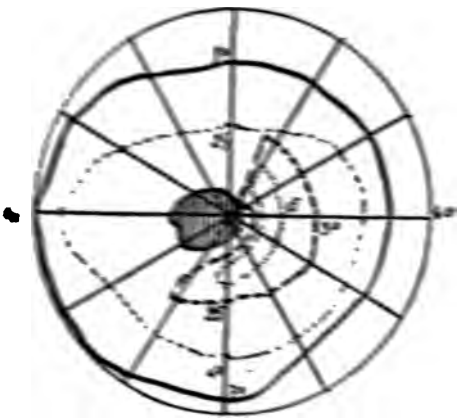


Fig. 29. left eye.

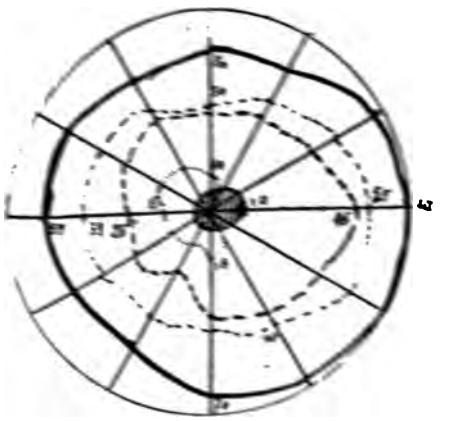


Fig. 30. right eye.

Field of vision in a case of insular sclerosis

——— white ---------- blue. ---------- red. green.

scotoma.

The periphery of the field of vision is free, while a central scotoma exists. The results are very analogous to those found in cases of toxic amblyopia. Uhthoff, from whom this figure is borrowed, states that he has only met with this case in which the field of vision had the above form.

according to the instruction given by Charcot, the two will not be found to entirely correspond in all cases.

The *disorders of sight* vary both in form and intensity. Parinaud, who has specially studied them, describes many varieties.

A. Slowly progressive diminution of visual power, possibly

Fig. 91.—Visual field of the left eye in a case of insular sclerosis (after Uhthoff
——— white. - - - - - - - - - red.
. . . . . blue. ——— green.

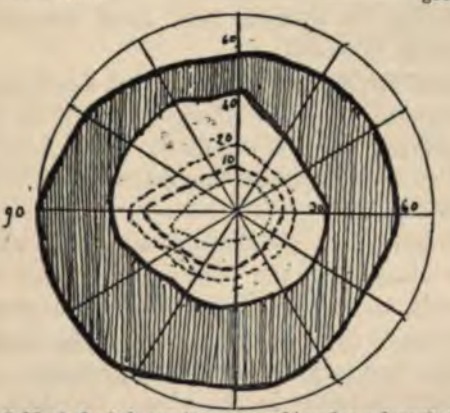


Fig. 92.—Visual field of the left eye in a case of insular sclerosis (after Uhthoff).
——— white. - - - - - - - - - red.
. . . . . blue. ——— green.

reaching from $\frac{1}{3}$ to $\frac{1}{7}$; the field of vision is normal but some dyschromatopsia exists as regards red and green, which can only be recognised by means of the photometer. This form would correspond to the commencement of decoloration in the outer portion of the papillæ.

B. The development of visual disorder is rapid; complete blindness may occur though this is but transitory; improvement takes place which is often very great; dyschromatopsia is frequent, different alterations may occur in the field of vision. The visual disorders are bilateral. These changes correspond with very pronounced atrophic decoloration of the optic nerve, which persists notwithstanding the improvement of sight.

C. The impairment of sight is unilateral, more pronounced and persistent. The field of vision presents an irregular contraction; no dyschromatopsia exists. This form depends upon white atrophy of the optic nerve which is well marked.

Uhthoff has also mentioned the different changes which occur in the *visual field*, and describes the four following varieties:—

A. Central scotoma without change in the periphery of the visual field.

B. Central scotoma associated with contraction of the periphery of the visual field.

C. Peripheral contraction of the visual field which is irregular, central vision being comparatively normal.

D. Regular concentric contraction of the visual field analogous to that occurring in hysteria (the most uncommon variety occurring once in 24 cases).

Charcot again dwelt upon the fact that *dyschromatopsia* may be very pronounced in insular sclerosis, that it is analogous to the same condition when it occurs in tabes, blue and yellow being the colours which persist longest, whereas in hysteria it is the red which does so.

These different facts, gentlemen, may be looked upon as showing that changes in the fundus oculi occur frequently in insular sclerosis, but are often moderate in degree; the visual loss which is due to them is usually but slight in degree, being liable to change and to pronounced retrogression.

An important character in the visual disorders is that they are often *unilateral*, or if *bilateral* are *asymmetrical*.

As regards their onset, Uhthoff found that in half the cases this was sudden, in the other half gradual. Sometimes the visual changes do not occur until a certain time in the evolution of insular sclerosis, while at other times they constitute the first indication of the disease. Lastly, they have at times been as it were of an intensity parallel with that of the other

symptoms of the malady, becoming more pronounced when the other symptoms increase in severity.

C.—Visceral Disorders.

Though less frequent than in tabes, for example, certain visceral symptoms are quite as liable to occur in insular sclerosis. In a memoir (presented to the Academy of Medicine for the Civrieux prize in 1885) I was already able to mention a certain number of cases in which troubles occurred in the stomach, rectum, bladder, or genital functions. Cases in fact were published in which *incontinence* or *retention of the urine* or *fæces*, or in some patients *impotence*, or more rarely *gastric crises* somewhat analogous to those of tabes had occurred.

Oppenheim, upon making special inquiry as to visceral symptoms in all the cases of insular sclerosis which came before him, found them far more frequent than observations previously made had led him to expect. According to this author they occurred in 80 per cent. of the cases. Notwithstanding the confidence that one should feel in the assertions of Oppenheim I cannot help fearing, gentlemen, that this high number was reached by his including cases of visceral disorder which were but very slightly pronounced. I would therefore repeat what I have already observed in speaking of the changes which occur in tactile sensibility:—"As regards the ordinary clinical features of the disease visceral disorders can scarcely be included among the symptoms of insular sclerosis, and even then they rarely become at all severe."

D.—Trophic Disorders.

Trophic disorders in the same way cannot be said to belong to the ordinary symptoms of insular sclerosis. At the same time several different kinds may be possibly observed.

Changes in the *nails* have been very rarely observed analogous to those which occur in tabes (?) (Domecq Turon).

Sloughing, which may occur in the glutœal region, or elsewhere are complications rather than direct consequences of the nervous affection.

Of all the trophic disorders the most pronounced is *amyotrophy*; this may affect the muscles of the hand, and specially the inter-

ossei; at other times it is more or less general, and to such an extent that these conditions of muscular atrophy in insular sclerosis have caused it to be confused with *amyotrophic lateral sclerosis;* Pitres drew attention to this fact with respect to a case which he observed; two other cases have been recorded by Dejerine and Skolosubow. The subject will be again considered when the diagnosis of the disease is discussed.

II.—Bulbar Symptoms.

It has been already observed that the division of the symptoms in insular sclerosis into spinal, bulbar, and cerebral is quite arbitrary, and only of interest from a *didactic* point of view. This statement having been made I shall class the following symptoms amongst those of bulbar origin:—

Difficulties connected (*a*) with *deglutition*, and (*b*) with *mastication*, which in some cases, of great rarity, may be so severe as to resemble those occurring in labio-glosso-pharyngeal paralysis. It will be understood that in such a case, when muscular atrophy of the limbs coexists, the diagnosis of *amyotrophic lateral sclerosis* is probably made, and an error, analogous to those of which I recently spoke, is difficult to avoid.

c. The tremor in the tongue does not seem altogether analogous to that which exists in the limbs, but rather to consist in a difficulty in protruding the tongue and in keeping it out of the mouth.

d. Glycosuria has been mentioned in a certain number of observations; their enumeration will be found in two interesting memoirs which appeared almost simultaneously, one by Richardière, the other by Miss Blanche Edwards. It is probable that in these cases the islets of sclerosis were seated in the floor of the fourth ventricle, at the point where a puncture causes the presence of sugar in the urine, unless of course true diabetes happens to exist.

e. Polyuria, which is at times observed, is probably due to a similar cause.

All the bulbar symptoms, or those which are called by this name, except perhaps the tremor in the tongue, so rarely occur in insular sclerosis that in my opinion they need only be mentioned, their description being quite unnecessary.

III.—Cerebral Symptoms.

Amongst the most important of these symptoms are the *disorders of speech*. This according to classical description becomes *slow*, *monotonous*, *scanning*, and *spasmodic*.

It is very difficult to describe these changes, and the best method of showing what they are will be to direct the patients now present to speak before you. At the same time, an attempt will be made to analyse the singular characters of the articulation. This patient, whose speech is much altered in the way already mentioned, will be asked to say a few words—this sentence for example—"toute médaille a un revers." Observe in the first place what may be called the "preparations to speak"; the lips are thrown forward, the forehead is wrinkled, and a great effort is made that all may be ready; now remark that every syllable is spoken in a laboured way, or rather is extracted from the mouth with difficulty; the effort first made is continued throughout the whole sentence, seeming to be always required. At the same time neither a vowel nor a consonant is omitted, almost precisely the same stress being laid upon each letter, and this contributes greatly to the slowness and monotonous character of the articulation. Observe, again, his great difficulty in pronouncing the syllable "daille" of the word "médaille," it might be said that a hill must be climbed upon the road which is already difficult, and that redoubled efforts must be made. This difficulty in pronouncing the vowels or diphthongs, followed by two liquid l's, is often very pronounced in insular sclerosis. The patient is now reaching the end of the sentence, and has arrived at the last syllable; this is, as it were, abruptly and incontrollably thrown out in an "explosive" manner. He seems to have badly calculated his power, and finding that there is now no syllable to retain him, makes an effort which exceeds those which he has previously made. It should be observed, gentlemen, that this description applies to those cases in which the disorders of the speech are very pronounced; in other patients these characters are but slightly indicated, and may even be entirely absent in the cases which are termed abortive.

Another symptom of cerebral origin which often occurs is *vertigo*; this symptom may only occur when the patient walks, and walking may be extremely difficult on this account; it may

exist, so to speak, continuously with but slight remissions. Lastly, in some cases, the *vertigo* exactly resembles that which occurs in *Ménière's disease*, and it will be seen that the same is often the case in tabes.

The existence of intellectual change has been noted by many observers. It is undoubtedly true that such change often exists to a more or less pronounced extent, but this does not show that all the observations in which it has been recorded are correct. Far from this I could mention certain cases in which the mental change was very pronounced, and the lesions were wrongly considered at the autopsy to be those of insular sclerosis, whereas in reality they were due to the existence of diffused cerebral sclerosis. I shall return to this point in a subsequent lecture. It should be remembered that the mental change in insular sclerosis is but moderate in degree, consisting most frequently of slight *weakness of the intellect, indifference* or *melancholia*, whereas at times these conditions may be, or appear to be, completely absent. In some cases only the mental disturbance is greater, and a kind of delirious idea which the patient has of his own importance may be observed, or even complete dementia or depravation of character if at least true insular sclerosis can be said to exist in such cases. To these mental changes the attacks of spasmodic impulsive laughter may be added which are very frequent in this disease.

One of the patients presented to you to-day has this symptom in a pronounced degree. It will be seen that the word "laughter" is scarcely spoken by me, than he begins to laugh, one must not say "with open mouth," since on the contrary the same tendency to spasm exists in this as in many other of the symptoms which occur in insular sclerosis; there are small outbursts rather than peals of laughter, although the tendency to laugh is compulsive and irresistible, and it is this which causes the peculiar appearance of the patients. In some cases the impulsive, uncontrollable nature of this symptom is so marked that the laughter continues for an unlimited time. Oppenheim, who has specially studied this symptom, states that he has seen it continue during several minutes, and be so intense as to produce alarming cyanosis of the face. This unreasonable laughter does not by any means indicate that the intellect of the patient is more seriously affected, and the answers made by this patient will show

that his intellectual condition is not so appreciably weakened as might be supposed.

Lastly, before ending what should be said with regard to symptoms of cerebral origin, the *apoplectiform* or *epileptiform attacks*, noted by Charcot, and which may occur at the onset or during the course of the disease, must be mentioned; these attacks do not by any means occur in all cases of insular sclerosis; they may almost completely resemble (and for more information upon this point, reference should be made to the thesis of Géraudeau) an attack of true epilepsy or apoplexy, except that in the apoplectiform attack of insular sclerosis the temperature usually rises to 39° and 40° C. (102° and 104° F.), whereas the reverse occurs in apoplexy due to cerebral disease.

LECTURE XII.

INSULAR SCLEROSIS (*continued*).

COURSE. DIAGNOSIS. ÆTIOLOGY.

COURSE: Onset acute, or gradual and progressive. The course of insular sclerosis may follow different types: a chronic progressive type, a chronic type with sudden exacerbations, a remittent chronic type, permanent improvement, and even recovery. Variable duration. Abortive forms. DIAGNOSIS: A. Of the *abortive forms:* with hysteria, paralysis agitans, mercurial tremor, chorea, general paralysis of the insane, cerebral tumour, transverse myelitis, compression of the cord, combined lateral and posterior sclerosis, tabes, focal cerebral lesions, amyotrophic lateral sclerosis. B. Of the *typical form* with Friedreich's disease, hysteria (coincidence of hysteria and insular sclerosis). ÆTIOLOGY: Slightly more frequent in the male sex, most frequent between the age of 20 and 30 years, does not occur in old age, very rare, if occurring at all, in childhood. The special cause is *infection*. Narration of facts, different infectious diseases during or after which insular sclerosis has been found to occur. Explanations of this fact.

GENTLEMEN,—The *symptoms* which occur in insular sclerosis having been described in the last lecture the *evolution* of the disease from a clinical point of view has still to be considered.

Firstly the *onset* occurs in different ways. It is at times sudden, even then appearing in different forms. At times an attack of *apoplexy*, either alone or complicated by hemiplegia, signalizes the commencement of the disease, or at times *hemiplegia* without apoplexy. At other times *vertigo* and *giddiness* are the first symptoms, while sometimes the disease commences suddenly and unexpectedly by *disorders of the sight*.

Sometimes, on the contrary, the onset is *slow* and *progressive*, the symptoms which first attract the attention of the patient or his friends consisting either of *increasing difficulty in walking*, or *disorders of the speech*, or *tremor* of the upper limbs; while in other cases, which at the same time are rare, the first symptoms experienced by the patient are more or less severe, and sometimes lightning *pains* resembling those which occur at the onset of tabes.

The *course* of the affection also varies very much but may, as I believe, be included in one of the following types :—

A. *A chronic progressive course*, that in which the symptoms are gradually and slowly, though constantly, aggravated. This form by no means occurs so frequently as some descriptions imply.

B. *A chronic course with sudden exacerbations.* It is in this form that some of the symptoms are found to occur which have been mentioned in connection with a rapid onset, apoplexy, hemiplegia, sudden troubles of the sight, &c., &c.

C. *A remittent chronic course.* The symptoms are but slowly progressive, if they increase at all, and remain stationary during long periods of time, after which slight exacerbations occur.

D. *Permanent improvement, or even recovery.* This not uncommonly occurs, and it is with good reason that Charcot frequently insisted upon the tendency to improvement exhibited by insular sclerosis. The disease may in fact be arrested at any period of its evolution, and not only cease to increase subsequently but even diminish to such an extent, according to Charcot, that actual recovery may occur.

In any case, gentlemen, it should be remembered that the prognosis of insular sclerosis is not so extremely grave as in some diseases of the spinal cord which I shall have to describe.

At the same time the disease does not always pursue this happy course, and independently of the fact that *death* may occur on account of some intercurrent disease, it may be due to an apoplectiform attack or some difficulty connected with deglutition and bulbar symptoms, or to the patient being completely wrecked by the disease.

The *duration* of the affection is essentially variable, and the patient perhaps dies at the end of one or two years, or may live for twenty years or more, according as the disease occurs in one or other of the forms which have been described.

We have just considered cases of insular sclerosis which are quite clear and more or less complete in character. Other cases, however, exist in which the symptoms are so little pronounced or in such an isolated form that the clinical indications of the affection can scarcely be recognised; it is these cases which Charcot termed *abortive* and in which, interesting as they are, the diagnosis is at times extremely difficult. It is these cases

which we should have specially in mind in distinguishing this disease from other affections with similar symptoms, and it is by considering them that we will begin to study the diagnosis of insular sclerosis.

A.—Diagnosis of the abortive form.

That is to say the diagnosis of those forms of the disease in which *one symptom* exists in an isolated manner or predominates over the others, to such an extent that it might on that account receive the name of *insular sclerosis, in which but one or few symptoms exist.* According as one or other symptom predominates the malady will have to be distinguished from very different diseases.

a. The predominating symptom is the tremor. The diseases which must then be distinguished from it are the following:—

(*a.*) *Hysteria*, which will be again mentioned when the typical form of the disease is considered.

(*β.*) *Paralysis agitans:* in this affection the tremor occurs more slowly than in insular sclerosis (4 or 5 oscillations occurring in a second instead of 6 or 7); it is also continuous except during sleep, exists whilst the limbs are at rest, is diminished rather than increased by voluntary movements, or whenever it increases during such movements, does so to such a slight extent as to bear no resemblance to that which occurs in insular sclerosis. Lastly it is a tremor with but very small oscillations, only affecting the wrist and fingers, the arms and forearms remaining pressed against the trunk. The other symptoms of paralysis agitans, the attitude, face, and gait, have not been mentioned, since my endeavour is to enable the cause of the symptom to be recognised by the characters which it presents itself.

γ. Mercurial tremor. This tremor has been specially studied by Charcot during the last few years; it is, according to his description, very analogous to that which occurs in insular sclerosis. The oscillations, in fact, occur with moderate rapidity (less than 8 oscillations in a second), are often of some size, and are produced and increased by voluntary movement. That, however, which enables it to be distinguished is, as Charcot expresses it, that it only ceases during rest in a *remittent* manner,

reappearing from time to time without the patient making any movement, either from the influence of slight emotion or occurring quite spontaneously, whereas in insular sclerosis the tremor is, as you know, completely absent while the part is at rest.

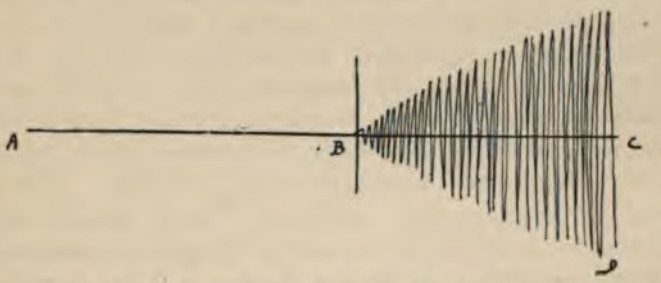


Fig. 93.—Sketch of the tremor in insular sclerosis (after Charcot). It is seen that during rest represented by the line AB no tremor occurs, but when the patient begins to execute a movement, as at B, the tremor commences and increases during its whole duration of the movement BC.

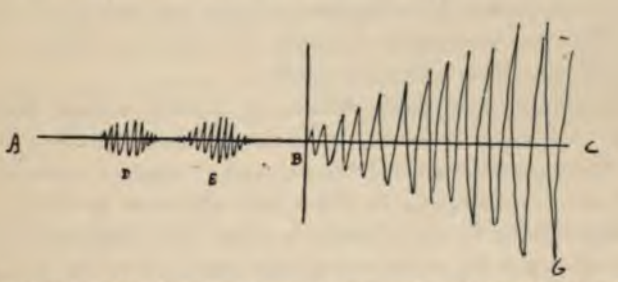


Fig. 94.—Sketch of the remittent tremor in mercurial poisoning (after Charcot). Whilst the part is at rest (AB) the trembling occurs during short periods of time (D, E), which soon cease, to occur again, and cease almost immediately. When a voluntary movement (BC) is made, the tremor is of precisely the same character as in insular sclerosis.

δ. *Chorea.*—It seems almost useless to dwell upon the diagnosis as regards this affection, in which no real tremor exists, but a series of irregular and unnecessary movements. There is no axis of direction around which the oscillations take place, nor are the movements similar to each other as are those of real tremor. It is therefore impossible to sketch them graphically with any exactness. For these reasons, without the other characters of the disease being considered, it seems to me impossible to mistake it for insular sclerosis.

b. The predominating symptom is disorder of speech with or without mental change.

The disease will then have to be distinguished from *general paralysis of the insane*. In these two affections the disorders of speech are often very similar. At the same time they may be distinguished by the fact that in general paralysis the speech is more tremulous, more hasty and indistinct, that the syllables are repeated, and that the spasmodic character which exists in insular sclerosis is absent. In order to perceive these differences it is usual to direct the patient to pronounce words which are of some length, and notably the word "artillery." While this word will be pronounced "ar-til-le-ry" by the patient suffering from insular sclerosis, the one affected by general paralysis says, "aar-tille-e-re-ry." In the first the speech is but *drawling* and *spasmodic*, while in the second it is *drawling*, *stuttering*, and *weak*, as if in the first case a hill was being ascended with difficulty but certainty, whereas in the second the gait is interrupted by false steps which occur unexpectedly, and are tripping,

c. The predominating symptom is faltering speech with or without nystagmus and optic neuritis.

These three symptoms, which, as already stated, frequently exist in insular sclerosis, by their co-existence often indicate the existence of a *cerebral tumour*, and it must be confessed that they are so analogous in these two affections as hardly to be distinguishable by the characters which they themselves possess. The existence of other symptoms must therefore be sought, either those of insular sclerosis or of a cerebral tumour, and amongst the latter, vomiting, headache, the special characters of the optic disc, &c., &c.

d. The *predominating symptom is disorder of the gait.*

a. The gait is *purely spasmodic:* the distinction must then be made from a certain number of lesions in the cord of which the principal are due, either to *transverse myelitis*, or *compression of the spinal cord* (Potts' disease, tumour, injury, &c.). Besides the fact that each of these presents its own special symptoms, it will be remembered that they are often accompanied by disorders of the sphincters, which is not the case in insular sclerosis.

β. The gait is *spasmodic* and accompanied by disorder in the *ocular muscles* (paralysis, myosis, changes in the reflex

actions of the iris). In this case the possibility of *combined lateral and posterior sclerosis* must be remembered; and spastic symptoms would then be combined with a certain number of the symptoms of tabes so that the existence of this affection could be easily recognized.

γ. The gait is *cerebellar*, besides which some *disorders* exist *connected with the eye*, and *there is absence of knee-jerk*. In these cases, which however are rare, there may be some difficulty in deciding whether insular sclerosis or *tabes* exists. In the latter affection the element "pain" is far more pronounced than in the former; besides this the ataxic gait is not precisely the same as that due to cerebellar disease, nor are the changes in the fundus oculi the same in the two diseases. It will not again be forgotten that genital and urinary disorders only occur as a complication in insular sclerosis, whilst they are one of the most important symptoms of tabes.

Lastly in some cases when *tabes* and *general paralysis of the insane* exist in the same patient, disorders of the speech are added to those of the gait and eyes, rendering the diagnosis more difficult; an investigation of the special symptoms which occur in each of these affections should then be made.

e. The predominating symptom is hemiplegia. Whether hemiplegia occurs or not after an attack of apoplexy the diagnosis between a *focal cerebral lesion* and insular sclerosis may be, specially during the first moments after its onset, of great difficulty. In many cases the subsequent course of the symptoms can alone settle the question, and with regard to this point I would remind you that hemiplegia in insular sclerosis is not followed by contracture which occurs in most cases of focal cerebral lesion.

f. The predominating symptom is amyotrophy, which may or may not be accompanied by bulbar palsy. It is these cases which, as already stated, may resemble *amyotrophic lateral sclerosis* and thus give rise to an error in diagnosis. They are, however, so rare that they may possibly never be seen. Should this, however, occur, it should be remembered that in insular sclerosis the muscular atrophy never attains a pronounced degree, that it never produces spasm in the upper limbs, or affects the chin, lips, or tongue.

B.—Diagnosis of the Typical Form.

B. Diagnosis of the typical form. When all, or nearly all the symptoms of insular sclerosis exist in the same patient, as in the *typical form*, the diagnosis usually presents no difficulty. It may do so, however, in some cases, which will now be alone considered.

a. Friedreich's disease. Without discussing at this moment Friedreich's disease, which will be specially considered in a subsequent lecture, I would remind you that many of its symptoms exist also in insular sclerosis; such are the nystagmus, the slowness of speech, the disorders of gait, and the tremor when voluntary movements are made, &c. The possibility of an error in diagnosis between these two affections is therefore easily understood owing to the resemblance of the symptoms. One of the most important differences from an objective point of view is the spasmodic character of the symptoms in insular sclerosis, whereas in Friedreich's disease the affected parts are completely relaxed. This is specially observed in the diminution or loss of the tendon reflexes (neither the irregular movements resembling those of chorea, nor the absence of ocular paralysis, nor the scoliosis will be now mentioned, their description being reserved for a subsequent lecture).

b. Hysteria. This disease now comes before us for the first time, a disease to be feared almost as much by the doctor as by the patient. It will be seen how exactly, I might almost say how artfully, it may imitate most other diseases of the nervous system, and notably those of the spinal cord, specially insular sclerosis. The thesis of Souques upon *imitative hysterical associations*, in which numerous illustrative observations will be found, may be read with advantage by those wishing for detailed information upon the imitative power of hysteria. The only fact which I will borrow from them is the following: In the six first observations of his work referring to patients who were known to be suffering from hysteria, the following symptoms were found to be associated therewith: vertigo, apoplexy, hemiplegia, intentional tremor (the interesting thesis of M. Dutil may be consulted on this point), disorder of the speech, diplopia. It will be recognized that a more perfect association of the principal symptoms which exist in insular

sclerosis could scarcely be found, and that in this case the name of *imitative hysterical association* is amply justified. Thus the mistakes which have been made are numerous, and the choice would be difficult to make even were professed neurologists alone to be quoted as a proof of this. It is possible that the occurrence of such a mistake explains the cases published by Westphal, with regard to which this author states that after observing most of the symptoms of insular sclerosis in certain patients he did not find the characteristic lesions of that disease at the post-mortem examinations. Such a mistake, gentlemen, may be avoided if it is never forgotten that hysteria exists, the fear of which should haunt you whenever a diagnosis is made, "timor hysteriæ initium sapientiæ." To know when to suspect it is the most important point; as regards its recognition this is comparatively easy, owing to the remarkable works of my master, Professor Charcot, upon the organic changes which occur in that neurosis. The "stigmata" of this disease should therefore be carefully sought: reduction of the field of vision, achromatopsia, monocular polyopia, areas of cutaneous anæsthesia, hysterogenous zones, &c., &c., and mistakes of no little importance may then be avoided.

Even, however, when the existence of hysteria is recognized it must not be supposed that all the difficulties are overcome. Cases occur, and by no means infrequently, in which hysteria and insular sclerosis are *associated together*, either from insular sclerosis occurring in a patient suffering from hysteria, or, as is much more frequent, from insular sclerosis being the cause of hysteria, an occurrence which was well discussed by Georges Guinon in his thesis for the doctorate. Whatever the chronological relation between these two affections may be they undoubtedly co-exist. When this occurs it may be difficult to recognize that such occurs, but in my opinion cases should be mistrusted in which, while symptoms of insular sclerosis exist conjointly with certain indications of hysteria, the tendon reflexes are truly excessive.

Insular sclerosis having been studied, from a symptomatic point of view, the conditions of its development will now be considered.

The ÆTIOLOGY of this affection presents, as will be seen, several points which are open to discussion, and therefore of real interest.

I am unable to indicate the exact frequency with which insular sclerosis occurs, but some idea of this can be formed from the statistics of Uhthoff, with regard to the ocular disorders which occur in this affection. These referred to 100 cases observed by him during 6 or 7 years, and which were seen in the hospitals or polyclinics of Berlin.

The two sexes are almost equally affected by the disease, the *male sex* suffering to a slight extent more than the female.

The *age* which is most affected is the *first half of adult life*, namely, between 20 and 30 years.

This indication should be carefully noted since it is a valuable ætiological fact. When you are in the presence of a patient whose illness began after the 40th year of his life, the possible existence of insular sclerosis need scarcely be considered on account of the age, and this is a most important element in the diagnosis of the disease. Though this affection does not show itself after a certain age, may the same be said with regard to the first years of life? Certainly not, since insular sclerosis may occur *in children*, and although not frequent, exists occasionally in early life. In a work devoted to this question in 1883, I was able to collect 13 cases of insular sclerosis in children, a large, and even too large number, I willingly confess, since amongst these cases, attributed by their authors to insular sclerosis, a certain number existed which I am now inclined to refer to, lobar cerebral sclerosis, or some other form of encephalitis occurring in childhood. More recently, Unger (1887), in a memoir upon insular sclerosis in childhood, collected 19 cases, and other examples were again brought forward by Nolda during the last year. The conclusion to which we ought to come owing to these facts is, that insular sclerosis may undoubtedly occur in children, but that it is very rare at this time of life; otherwise, the symptoms and course of the disease are essentially the same as in the adult.

The *causes* mentioned with regard to the ætiology of insular sclerosis in the different treatises upon nervous diseases are well known; they are always *overwork*, *exposure to cold*, *injury*, and *excess* of every kind. I am quite convinced that another cause exists which is more frequent than any of these, and without which islets of sclerosis would not be present in

the brain or spinal cord. This truly effective cause is the presence of *infection*, or rather *infections*.

With your permission this question will be discussed in some detail, since the subject is one which specially interests me. Even in my first work* upon *Insular Sclerosis and the Infectious Diseases* (1884), I thought it possible to affirm definitely that this relation of cause and effect existed between them; at that time I had collected 25 cases, of which a certain number were taken from the memoir of Kahler and Pick, in which as early as in 1879 these authors drew attention to the frequency with which insular sclerosis occurs after acute disease. It is true that all the facts stated are not equally convincing, and I admit that the prudent reserve recommended by our esteemed master, Professor Jaccond (Clinical Lectures at the Hospital de la Pitiè), was quite legitimate from a purely scientific point of view. Since that time, however, new facts have been added to those previously known. Undoubtedly some of these are more or less incomplete with regard to certain points, but it must be remembered that clinical is not the same as experimental investigation, and that being obliged to accept situations which we have not ourselves created, we must make use of whatever documents chance puts into our hands, and draw from them the best conclusions we can.

This is what usually occurs: a person of between 20 and 30 years of age contracts an infectious disease (it will presently be seen in the enumeration which I shall make how different in nature the infectious diseases may be), and either in the course of the disease, or during the period of convalescence which follows it, or perhaps some months later, the first symptoms of the nervous affection occur. In some cases, to which reference will again be made, when the pathological anatomy is considered, these symptoms improve, and may completely disappear, the morbid process not having then produced sclerosis, but having been definitely arrested. In other cases the symptoms which

* The infectious origin of insular sclerosis is almost generally admitted, and it would be ungracious on my part to find fault with those who received my work with incredulity, and in some cases in a hostile spirit. The same occurred when I suggested that *in the great majority of cases epilepsy has an infectious origin*, and until this opinion is also acknowledged to be correct, I feel bound to state that my conviction as regards the infectious origin of epilepsy remains entirely unchanged.

have occurred become aggravated, either progressively or by sudden exacerbations, and before long the whole clinical aspect of insular sclerosis which has been described presents itself.

It is certain that when the nervous symptoms only occur, as long as several months after the infectious disease, the connection between the two is far from evident. If such connection had only been observed three or four times, it might be looked upon as a mere coincidence, and this would explain to a certain point the frequency of infectious disease and the comparative rarity of insular sclerosis. When, however, the number of facts of this nature published by different authors is considered, when one compares them with those in which the nervous affection occurs during the course of an infectious disease, or the subsequent period of convalescence, it is impossible, in my opinion, to deny that they stand to one another in the relation of cause and effect. You will soon perceive that the pathological anatomy of the disease, far from being in opposition to this view, supplies an additional reason for believing it to be true.

We will now examine the facts themselves; in the following enumeration the infectious diseases will be named during, or after which, insular sclerosis has been found to occur.

Enteric fever appears to take the lead in a decided manner, since in 25 observations which I collected for my work on this subject the disease was met with 11 times.

Pneumonia is not infrequently connected with the disease. In a case which Richard had obligingly communicated to me, that of a patient named Rob, who was well known in the hospitals as a typical example of insular sclerosis, pneumonia existed on both sides of the chest.

Malaria is mentioned in several observations, and in the "Leçons du Mardi" of Charcot you will find that a patient was interrogated upon this subject, and of his own accord referred the nervous affection from which he was suffering to an attack of malaria.

Eruptive fevers must also be mentioned, namely, *measles*, *scarlatina*, and above all, *small-pox*. The cases are numerous in which insular sclerosis has been known to occur during convalescence after the latter affection: tremor in the limbs, with more or less paresis, disorder of the speech, which becomes slow and scanning, nystagmus, and in short, all the characteristic

symptoms of insular sclerosis may exist. At times these symptoms cease and entirely disappear, but they may also continue and confirmed insular sclerosis occur.

Other infectious diseases should also be named though, as regards the number of cases connected with them, they are but of secondary importance. Such are *diphtheria*, *whooping-cough*, *erysipelas*, *dysentery*, and even cholera. Lastly, at quite a recent date Charcot, in one of his clinical lectures, recorded a most interesting case, in which insular sclerosis seems to have followed *cerebral rheumatism*.

Before concluding this enumeration, a paragraph must be devoted to *unnamed infections*, so frequent, so little known, I might add so much disregarded. There are no special symptoms at the onset which indicate its existence; fever is known to have occurred, prolonged discomfort with or without gastro-intestinal symptoms, occasionally jaundice or pulmonary trouble, nothing else being known about the disease. In such a case, gentlemen, you must not doubt that this is certainly a case of infection, but of such a kind that it has not received any definite clinical name. As regards the patients in whom insular sclerosis seems to occur from the influence of injury or some other purely physical cause, my conviction is that these cases are also due to infection, but that the infection has passed away completely unperceived, while some less important but more dramatic incident has alone attracted the attention of the patient or those who are with him.

Of the manner in which insular sclerosis is produced by infection it will be better to speak when the pathological anatomy of the disease is considered.

As to the part which micro-organisms play in this process the question is an embarrassing one, scarcely any facts existing which can aid its solution. At the time of my first communications upon this subject, some of my colleagues, who undoubtedly had not done me the honour to read them, pretended to believe that I had described "the micro-organism of insular sclerosis." On the contrary, I have been careful to avoid exposing myself to such an accusation, and merely stated the fact that insular sclerosis occurs in a number of infectious diseases which are quite distinct from each other. Can it be looked upon as due to the micro-organism which is special to each of these

diseases? At first sight it seems little probable that micro-organisms which are so different could produce such similar effects. Again, if insular sclerosis was directly due to the micro-organism of each of these diseases, it should be observed far more frequently than it is during their occurrence.

For these reasons I should be inclined to think that insular sclerosis is rather the result of one of those *combined infections* which are so frequent during the course of the different infectious diseases. Is there a special micro-organism in these cases? It seems more probable that the lesions of the brain and spinal cord are caused by an ordinary pathogenic microbe, of which the special action on the brain and spinal cord is due to its being seated in those nervous centres. These questions, during the present state of our knowledge, must be left unanswered. At the same time I would say that I am inclined to believe in the second solution as being more in conformity with the knowledge we now possess as to how the micro-organisms usually act.

These, gentlemen, are suppositions, and I put them before you without unreasonably insisting upon them. The one point in this discussion which I would fix in your minds is the following fact, a fact which, thank God, has been well established, viz., "that the cause of insular sclerosis is intimately connected with infectious diseases."

LECTURE XIII.

INSULAR SCLEROSIS (*continuation and end*).

PATHOLOGICAL ANATOMY. NATURE. TREATMENT.

The meninges are usually but slightly, if at all, affected. Insular appearance of the lesions: abundance of the islets, extreme irregularity of their seat, dimensions, colour, and consistence, their seat upon the nerve roots connected with the medulla oblongata and spinal cord. Microscopical examination: the islet seems to have been cut out by means of a punch: the sclerosis essentially involves the neuroglia, containing many granular bodies in its substance, the sheaths of myelin being destroyed while the axis cylinders are preserved. Deductions which may be drawn from this fact with regard to the absence of secondary degeneration: remission or cure, the occurrence of tremor, lesions of the optic nerve, changes in the blood vessels. Pathology of insular sclerosis: inflammatory process of interstitial origin, commencing near the blood vessels. Pathological anatomy thus in accordance with ætiological data. Hypotheses connected with this subject. Distinction between insular sclerosis and diffused multilocular sclerosis. Treatment.

GENTLEMEN,—That a clear idea may be formed of insular sclerosis the study of its lesions is indispensable. The PATHOLOGICAL ANATOMY of the disease will therefore be now considered.

A. From a macroscopic point of view the affected parts of the central nervous system present the following appearance from without inwards:—

The *meninges* are neither thickened nor adherent, the transparency of the pia mater being preserved so that the most superficial islets may often be seen through the membrane. In some cases, however, meningeal changes have been said to be observed. Without being able to affirm that the meninges are never involved in insular sclerosis I must put you on your guard with respect to these cases, of which the greater number are far from being pure examples, if they belong at all to the disease which is now being considered.

The *nervous centres* themselves, when the meninges have been removed, are found to present different aspects. In some cases there is nothing abnormal at the surface, while if sections

are made through the deeper parts islets are found to be contained in them, whereas in other cases no islets are found either at the surface or more deeply seated, and it is only after

Fig. 95.—Transverse section of the extremity of a cerebral hemisphere in which 3 islets of sclerosis are seen in different parts. One of these (B) is entirely contained in the white substance. The second (C) is for the most part in the grey matter. The third (A) is seated partly in the white and partly in the grey substance, being as it were astride over one of the fissures of the cortex.

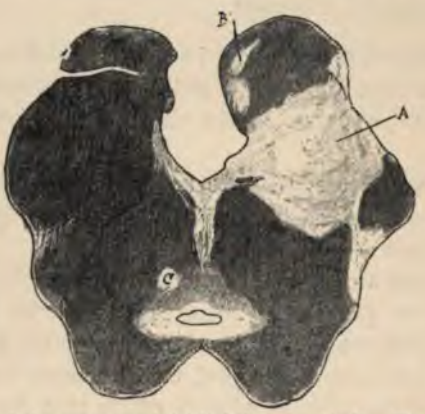


Fig. 96.—Section of the *crura cerebri* (slightly magnified). (Damaschino collection.) It is in the white substance that the islets of sclerosis have specially appeared. The islet of sclerosis (A) involves the whole thickness of the part; another islet is seated near the 3rd ventricle.

remaining for a more or less prolonged period in a solution of bichromate or being immersed for a time in a colouring liquid that such islets of sclerosis are clearly seen.

Usually the nature of what occurs is more simple, and as I have just said, even before removal of the meninges some of

Fig 97.—Transverse section of the pons varolii (slightly magnified). (Damaschino collection.) The white portions of the figure are those in which the islets of sclerosis are seated.

Fig. 98.—Section of the pons varolii in the long axis from a case of insular sclerosis (slightly magnified). (Damaschino collection.) The white portions of the figure are those in which the islets of sclerosis are seated.

the islets of sclerosis can be very easily distinguished by transparence.

The islets present some characters which it is important to

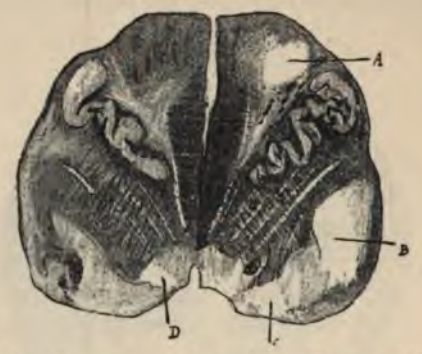


Fig. 99.—Section of the lower part of the medulla oblongata (slightly magnified). The grey and white matter of the part are both occupied by islets of sclerosis. It will be seen that neither the olivary body nor the grey substance in the floor of the 4th ventricle (D) have been respected.

Fig. 100.—Section of medulla oblongata parallel to the floor of the 4th ventricle (slightly magnified). (Damaschino collection.) The grey parts are those affected by the islets of sclerosis.

know: they are quite irregularly placed, and hence the name proposed by Charcot, and rightly adopted, of *disseminated* sclerosis. Thus it is impossible to find two cases which, from an anatomical point of view, resemble each other. In some cases the seat of the lesions is unforeseen and odd, being formed according to no rule, arrested by no obstacle, and confined to no anatomical system. At one time the islets exist abundantly at the surface of the spinal cord and convolutions, at another the deep parts of these organs are specially affected. The islets are undoubtedly more numerous in the white than in the grey substance, although it would be wrong to say, as do some authors, that the grey substance offers an insuperable barrier to the islets of

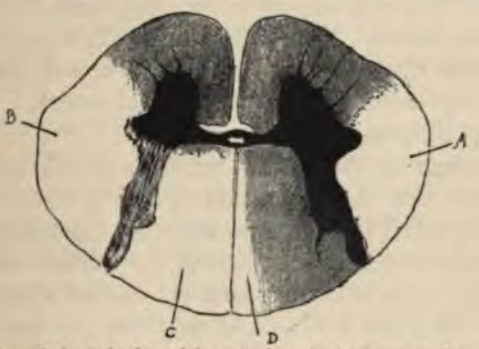


Fig. 101.—Section of the spinal cord (cervical region) in a case of insular sclerosis. Two islets are seen in this section: one (A) of moderate size, the other (B C D) of very large size is seated on both sides of the cord passing beyond the posterior fissure, and posterior horn on the left side.

sclerosis which are seated in the adjoining white substance. The different fissures of the cord again do not prevent its extension, and in the figures placed before you the lesions are seen to be at times as it were astride over the fissures. The islets are found throughout the whole length of the cerebrospinal axis, from the surface of the convolutions or ventricles, from the crura cerebri, pons varolii, or cerebellum to the *filum terminale*, though not in all parts with the same frequency; thus, for example, they rarely occupy the large ganglia of the cerebral hemispheres or olivary bodies.

The word "disseminated" indicates that the islets are of small dimensions (inselförmige sklerose in German, insular sclerosis in English; polynesic sclerosis of some authors), and they have

usually the appearance of being flattened. In many cases, however, the islets of sclerosis have a cuneiform aspect, and in this case their base is usually turned towards the periphery of the organ in which they are seated.

The number of islets varies considerably, and as many as some hundreds may exist in the same patient.

Their size also differs much, varying from that of a millet seed to the size of a two-shilling piece.

Their appearance is as follows: usually of a slate or greyish red colour, they become clearly more red after being exposed for a time to the air; their consistence differs in a similar way, being much greater in the large than in the small islets. In one section the islets may appear to be prominent, while in others they seem depressed. This difference in their appearance probably depends upon the age of the islets, and the degree of evolution of the sclerosis where they are observed.

So far, gentlemen, the islets seated in the nervous centres have been alone considered; it must be stated, however, that some authors have observed them upon the roots of the peripheral nerves which adjoin those centres. They have been also found upon the nerves *connected with the medulla oblongata*, not only upon the optic nerve of which the special structure makes it to a certain point a diverticulum from the brain, but also upon the hypoglossal, glossopharyngeal, vagus, &c. The *nerve roots* connected with the spinal cord may be also the seat of the islets.

B. Their *microscopical* appearance presents certain characters which should be mentioned.

When examining a specimen with a microscope of slight magnifying power the clearness with which the islet of sclerosis separates itself from the adjoining part is at once observed. In the section now put before you the islet of sclerosis seems almost to be punched out, and it will be difficult to find an islet of sclerosis in any organ of which the limits are as little diffused as those of the islets which we are now studying. What also contributes to give them the appearance of being "punched out" is the decided manner in which the sheaths of myelin disappear throughout the whole extent of the islet, whereas outside it, and so to speak without interval, the sheaths are preserved. The fact must therefore be carefully remembered that if in the disease

which we are studying, sclerosis presents itself in an irregular form, and essentially disseminated as regards its seat the diffuse

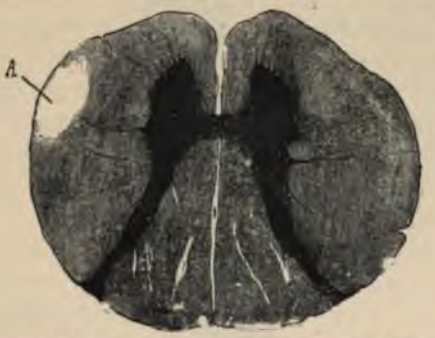


Fig. 102.—Section of the spinal cord in insular sclerosis (slightly magnified). The islet of sclerosis (white) is in this case single, and seated at the surface.

Fig. 103.—Section of the spinal cord in a case of insular sclerosis (magnified to a moderate extent) (coloured by Weigert's hæmatoxylin). This figure shows how clearly the islet of sclerosis (A) is distinguished from the surrounding healthy tissue; the white colour of this islet indicates that all its fibres have completely lost their sheath of myelin.

form exists, and should, on the contrary, be considered as a type of sclerosis occurring in foci.

As respects the inner part of the islet, it is composed of a more or less dense network of neuroglia, which is thickened, and of which the meshes are more pressed together. Its appearance has thus a special character analogous to that which would result from elongated crystals being grouped together in the direction of different axes, which cut each other at acute angles (*vide* fig. 105). It should be remarked that of all forms of sclerosis in the spinal cord insular sclerosis is, in the opinion of Weigert, that in which the proliferation of the neuroglia is most pronounced, so that if the forms of sclerosis

Fig. 104.—Longitudinal section of the spinal cord from a case in which the islet of sclerosis was seated at the surface of the cord (slightly magnified). (Damaschino collection.)

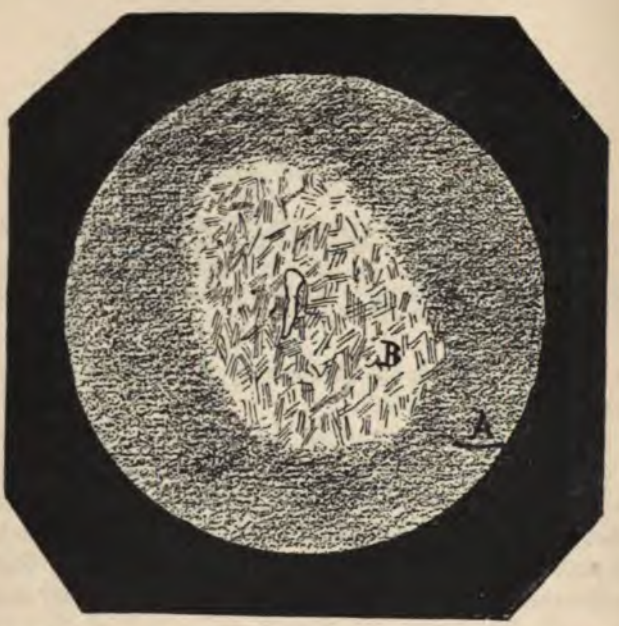


Fig. 105.—Section of an islet of sclerosis in the white substance of the brain. A, sound tissue. B, islet of sclerosis, the tissue adjoining which is composed of fibres disposed in columns, and which cross each other at the most different angles. A blood vessel is seen at the centre of the islet of sclerosis.

were classified according to the extent to which the neuroglia is involved it is insular sclerosis, and not Friedreich's disease, which would occupy the first place.

Within the meshes of the network, in a section produced by congelation, brilliant, refractive, or black and granular bodies, according to their distance from the objective, are seen to be sprinkled in large number. These are the granular bodies of which the signification will be discussed presently. I would only mention their existence at the present time, and point out that abundant and irregularly scattered as they are in islets of recent formation, they are to be found also in islets of old date, though almost solely at the surface in that case, and little, if at all, in the central parts.

In conjunction with these granular bodies within the islets of sclerosis, and as an explanation of their existence the *destruction of the sheaths of myelin* should be mentioned, such destruction being complete within the islets, whilst, as already observed, the sheaths outside these lesions are absolutely sound. It is to this destruction that the clear colour contrasting with a black ground which is produced by Weigert's coloration by hæmatoxylin in these sections is due. It will be remarked that not a single sheath of myelin remains at the centre of the islets, whilst at their peripheral part, where the patch adjoins the sound tissue, some of these sheaths are seen to be disappearing, and the process of their destruction can be studied at that part in its different stages.

As you know, gentlemen, the nerve fibres contained in the central organs differ from those in the peripheral nerves in the fact that they do not contain a sheath of Schwann, being only composed of an axis cylinder and sheath of myelin. You have heard what happens to the sheath of myelin, and what becomes of the cylinder axis? In a general way it must be admitted that the *axis cylinders are retained* in insular sclerosis. This fact, as you will see, is of great importance, and affects the whole clinical history and pathological anatomy of the disease. The honour of having first shown and proved this to be the case, and of having indicated all the consequences which resulted from this fact, must be given to Charcot.

This condition of the axis cylinders has been verified by a very large number of observers, and Babinski has given in his thesis

very exact representations of it. You can easily perceive this by observing the preparations coloured either by means of carmine or by the method of Pal; in the midst of the meshes of indurated tissue a number of coloured points will be seen which represent the section of the axis cylinders. In places you will also observe that some of these points are much larger than the others; these consist of *swollen axis cylinders*, which are not infrequently seen in islets of insular sclerosis, and also it may be said in some other varieties of sclerosis affecting the spinal cord (Hayem).

In the islets of ancient date, however, in which sclerosis exists in a pronounced degree, a certain number of the axis cylinders disappears; this, however, is merely an accessory fact, and would not be in opposition to the axiom that "in insular sclerosis the axis cylinders are retained in the midst of the affected islets, although their sheaths of myelin may have been destroyed."

Certain deductions may be drawn from this persistence of the axis cylinders:—

a. One of the principal of these is the *absence of secondary degeneration* in the path of the nerve fibres connected with the diseased part. This again is a fact which was indicated by Charcot from the time that he first studied the pathological anatomy of this disease. The absence of secondary degeneration can be easily recognized in sections of the parts seated above or below the affected islets. The same fact may be observed if longitudinal sections are made in a direction parallel to that of the nerve fibres, and the morbid changes will be seen to be limited in the same precise way above and below, as at the sides. From a clinical point of view it will now be understood how it is that the paralysis or paresis which exists, though of the spasmodic type, is not accompanied by permanent contraction, namely, that owing to the absence of secondary degeneration there is no reason for its existence.

In the rare cases of which I have already spoken, in which the axis cylinders are also finally destroyed, the existence of slight secondary degeneration is observed; this, however, shows no tendency to extend, and should be considered as purely accidental.

β. The *remission*, *improvement*, and even *cure* which may possibly occur, can be also easily understood, since the axis

cylinders not being destroyed the most important, in fact the only part of the nerve which is absolutely necessary for transmission of the nervous current is retained. When the inflammatory process diminishes, even to a slight extent, it is intelligible that the function of the nerve becomes almost completely re-established. In some cases indeed when recovery occurs, Charcot is of opinion that the sheaths of myelin, which had disappeared in the islets of sclerosis, may be restored, so that the nerve fibres return to their normal condition, not only from an anatomical, but also from a physiological point of view.

γ. According to many authors the persistence of the axis cylinders with disappearance of their sheaths of myelin throws a great light upon the pathology of the *tremor*. Comparing in fact the nerve fibres to electrical wires, their sheath of myelin to the isolating material which surrounds them, and the nerve force to a current, one is naturally led to think that on account of the destruction of the sheaths the fibres are no longer sufficiently "isolated," and that the "current may escape in places," this being physiologically indicated by the tremor. It should, however, be added that, although generally admitted, this explanation of the tremor is not accepted by every author. Some attribute it to the existence of islets in the thalamus opticus, while others look upon it as purely and simply an excessive degree of reflex action in the spinal cord.

Such, then, is the condition of the nerve fibres in the islets; as regards the *nerve cells*, on account of what has been already observed by me as to the relatively sound condition of the grey matter in this complaint, you would not expect them to be very frequently involved, which in fact is the case. At the same time, when contained in one of the islets, they are found to be diminished in size, to lose their prolongations, to present clear indications of atrophy, and even to almost completely disappear.

When islets occur in the optic nerve they present characters in every respect analogous to those of the islets in the brain or spinal cord. Uhthoff describes them as follows:—"The process of proliferation seems to take place to a very pronounced degree; in the first place, in the part occupied by the finest elements of connective tissue, seated between the nerve fibres contained within the large meshes; in the second place, in proliferation

of the nuclei; this process may also extend to the largest septa, and to the inner sheath of the optic nerve. The

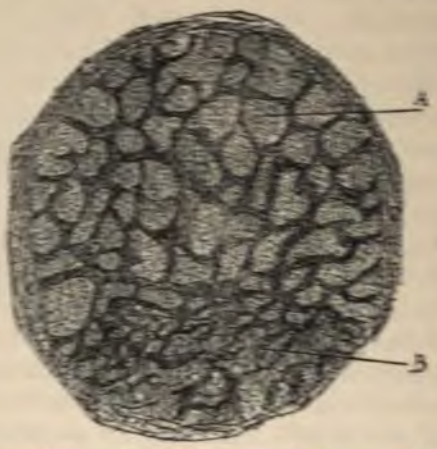


Fig. 106.—Section of the right optic nerve in a case of insular sclerosis. B, Partial focal lesion. The islet A, and the numerous ones resembling it are unaffected. (After Uhthoff.)

atrophy of the nerve tissue should, therefore, be considered as secondary. The disappearance and destruction of the sheaths occur in a relatively rapid and complete manner, while the axis cylinders entirely, or almost entirely stripped of myelin, commonly persist. The islets of sclerosis do not usually produce secondary degeneration of the fibres in the optic nerve, or if they do so to a very slight extent, as is shown by the condition of the optic disc, either as seen by the ophthalmoscope or the microscope. Even though very pronounced atrophic changes may exist behind the globe of the eye, the results of ophthalmoscopic examinations remain completely negative, and the atrophic decoloration of the optic disc is usually quite incomplete and partial, even though very extensive alterations may exist behind the globe of the eye, involving the whole thickness of the optic nerve, and having produced the most decided atrophy of this nerve." According to Uhthoff the conditions required to produce the phenomena of optic neuritis in insular sclerosis are the existence of recently formed and extensive islets in the optic nerve *immediately behind* the globe of the eye.

'er to finish the study which is now being made of the

elements contained in the islet of sclerosis the *blood vessels* must be mentioned. The alterations which they present are most clear, the thickening of their walls having been pointed out by Charcot from the time that he first wrote about this disease. This thickening specially involves the external coat, and it is on account of this that the vessels remain open, and apparently dilated in section when the sclerosis is very pronounced. At times dilatation appears really to exist and to be accompanied by increase in the diameter of the perivascular lymphatic sheaths; these in the islets which are recently formed, or in process of being formed, are absolutely full of granular bodies. Lastly, it is not useless to remark, as you will soon understand, that vessels are often found at the centre of the islet, and that the most serious changes in sclerosis occur close to these vessels.

So far, gentlemen, I have purposely omitted to speak of the PATHOLOGY of insular sclerosis, and of the place which it should hold in a classification of diseases, because I wished to put first before you every step of the process. After what has been said of the ætiology and pathological anatomy of the affection this study may be now undertaken by us.

In the first place one fact is generally admitted, namely, that the morbid process is inflammatory and of *interstitial origin*. Charcot was again the first to express this opinion, which was afterwards almost unanimously adopted. A few years ago, however (1886), Adamkiewicz, on account of results obtained from staining by safranin, thought himself justified in stating that the affection commenced in the nerve elements, and that the interstitial change was but secondary. Unfortunately the effect of staining by safranin, which was the sole origin of this idea, has found so few supporters and so many adversaries that it is unnecessary to say more about this theory, and the morbid process which exists in insular sclerosis may be safely considered as belonging to the group of inflammations which are primarily interstitial.

The origin of this primary interstitial process connected with the islets of sclerosis is *in the blood vessels*. After what has been said with respect to the infectious nature of insular sclerosis this will not surprise you, but it is an interesting fact that even without adopting this view the seat of the lesions is definite

enough to point to the same conclusion that they are of vascular origin. Rindfleisch expressed himself in favour of this supposition in 1864, but the opinion was not generally accepted. More recently a pronounced inclination has been shown in different directions to accept this theory, and amongst others Hugo Ribbart, without in any way considering the ætiology, and entirely owing to a series of deductions connected with its pathological anatomy, concluded that the lesions of insular sclerosis are caused by the presence of some irritating agent in the blood vessels. According to this author, what happens is as follows: under the influence of an irritating agent, which circulates in the vascular system, a fibrinous coagulum forms at some point in a small vessel covering most usually but a small part of its wall. Owing to the irritation which occurs at this point, and by its agency, more or less pronounced diapedesis of the leucocytes occurs. Inflammation then takes place round it which extends concentrically from the blood vessel in which it commenced. Thus, as you will understand, insular sclerosis is in no way, as some authors have stated, a diffuse form of sclerosis, but occurs essentially in "foci," of which the origin is most clear. Such is the idea, gentlemen, which pathological anatomy suggests, and though as yet unconfirmed by ætiological data, that is to say, though no irritating agent of infectious nature has yet been discovered, there is no doubt that this will be the case at some future time.

In the absence of definite facts suppositions can alone be made, and questions be asked. Thus it is doubtful whether the infectious agent is harmful in itself or by means of the materials which it secretes. It seems to me that the former mode of action is the most probable, if the dissemination of the lesions and their essentially *embolic* character be considered. As to the formation of the islets themselves two facts are in my opinion specially interesting. One of these is that, as I have already mentioned, many of the islets present at their periphery a more or less considerable number of granular bodies; this indicates that the morbid process is still in an active condition, since the products of disintegration of the nerve fibres (which form the granular bodies) have not yet been absorbed and are still being produced. The second fact to which I would draw your attention is the following: in certain cases of which the

onset dates from several years ago islets of small dimensions containing granular bodies are still found, both at the centre and periphery, having, in a word, all the characters of yellow islets, whilst in other parts of the nerve centres islets are found of undoubted sclerosis of large size, and only presenting granular bodies in any abundance at their periphery, that is to say, having every appearance of *islets of long duration*. If the former fact is interpreted in a strictly logical way we should say that the morbid agent must remain in each islet, since it seems to continue its action for an almost indefinite time. As regards the second fact it will bring us to the following conclusions: the morbid agent remaining in the organism is capable of reproducing itself even at the end of a greater or less length of time and of undergoing a totally fresh dissemination. The course of the disease confirms to a certain point this supposition, since not only may it be continuously progressive but also present sudden aggravations which seem to indicate a fresh development of the lesions. I thought it my duty, gentlemen, to mention these conclusions since they emanate directly from the facts which I have described to you, at the same time I should be the first to advise you only to accept them as suppositions; the evolution of most lesions in the nervous centres is still enveloped in so much obscurity that it would be imprudent to discuss them in any other way. The equation contains too many unknown quantities, and at the present time, as I have already stated, we can only establish the terms, and clearly state the questions to be answered.

Though however, gentlemen, we are still in doubt as regards certain points the following facts seem to be certain, viz., that insular sclerosis is of vascular origin, and apparently due to some morbid process similar to that which attends embolism; that it is liable to be produced by the influence of several infectious diseases, probably by the mechanism of combined infection.

One point, however, must still be discussed. I cannot in fact admit, as most authors do, that all the cases in which foci of sclerosis, in greater or less number and more or less disseminated, are found in the spinal cord should on this account be regarded as examples of "insular sclerosis." Two forms of sclerosis may in my opinion be quite clearly distinguished. In some autopsies

in fact the islets are found to be few in number, often seated in the spinal cord alone, and to have irregular and deeply indented borders. The microscope often shows that in these islets not only the sheaths of myelin but also the axis cylinders are more or less destroyed (it is in these cases that secondary degeneration is specially observed). Lastly their peripheral margins have not the "punched out" appearance already described; they extend more or less into the surrounding tissue, and for some reasons would be more appropriately called "diffuse sclerosis."

From a clinical point of view this morbid condition presents also certain distinctive characters; it occurs usually at a later age (specially after the age of forty years); it does not usually present the classical symptoms of insular sclerosis, specially the tremor, disorders of speech, eye-symptoms, &c., paralysis more often occurs; lastly its course is not the same as that of insular sclerosis, being often much more rapid and complicated by serious nervous symptoms which may end within a few months in death. As regards its ætiology, I can make no definite statements, but whilst believing that in this case, as in that of insular sclerosis properly so-called, infectious diseases play an important part, I am at the same time convinced that these two forms of sclerosis are due to different causes; thus, for example, syphilis which can scarcely be said to produce insular sclerosis properly so-called, may in some cases be looked upon as the cause of the second variety of sclerosis. It seems to me that the differences are both numerous and important enough to justify the distinction of these two morbid conditions, and although both are examples, in the common acceptation of the term, of "insular sclerosis," I should be inclined to apply this term to the classical variety alone and to call the other by some different name, as for example *diffuse multilocular sclerosis*.

It is certainly true that *neither one nor the other are diseases*, properly so-called, but consequences, sequelæ, of general affections. Insular sclerosis properly so-called, however, presents such characters, and, notwithstanding the variety in its symptoms, such a definite clinical aspect that it deserves, at least from a nosographical point of view, to be looked upon as a morbid condition. It is possible that at some future date the progress of bacteriology will enable this condition to be again divided, but at present it is impossible to do more than separate from it the

cases of "diffuse multilocular sclerosis," and I hope that you are convinced of the necessity of doing this.

We will now consider the TREATMENT of insular sclerosis. From what you know, gentlemen, of the nature and lesions of this affection you would naturally doubt the efficacy of such therapeutic agents as are now employed. These doubts are by no means unreasonable, and when you read of cases in which the administration of an alkaloid or hypnotism have been followed by recovery in insular sclerosis, you may unhesitatingly state that the diagnosis was incorrect, and that the patient was merely suffering from hysteria. At the same time, you are now aware that this affection tends of itself to diminish and even to improve, and this tendency can, perhaps, be to a certain extent increased by the administration of drugs suitable on the one hand to the "sclerotic," on the other to the "infective" element of the disease. You know how much good may be done by *iodide* of potassium or sodium in vascular sclerosis and these should therefore be administered in small doses, but for some length of time. As regards the second indication, which it is more difficult to fulfil, perhaps the best drug to employ is *mercury;* this you would therefore be justified in administering for some length of time in a moderate dose and in the form which seems most suitable to the condition of the patient. It must be well understood that this medicine is not given as an antisyphilitic since syphilis appears to play but the slightest or no part in the ætiology of true insular sclerosis. It is only on account of its disinfecting properties that I should recommend its administration, and the other disinfectants used internally may be also administered. I have little doubt in fact, gentlemen, that by the employment of such a substance as the vaccine matter of Pasteur or lymph of Koch the evolution of insular sclerosis will some day be rendered absolutely impossible.

LECTURE XIV.

TABES.

TERMINOLOGY. HISTORY. SYMPTOMS.

TERMINOLOGY: locomotor ataxy, tabes, &c. History: Romberg, Duchenne of Boulogne, Charcot, &c. Symptoms: A. *Motor disorders:* 1. Alterations of the muscular sense, loss of the knowledge of posture, loss of the power to detect differences in weight, difficulty in maintaining the upright position. Sign of Romberg: disorders of the gait, indications given by Fournier in connection with the investigation of these disorders; "giving way of the legs," incoordination of the upper limbs. 2. Athetoid movements and muscular tremors. 3. Paralysis: its characters; hemiplegia, paraplegia with sudden onset, local forms of paralysis.

GENTLEMEN,—The *locomotor ataxy* of Duchenne of Boulogne is now little more than a relic of the past. It is of the name alone that I speak, the place which it once rightly occupied in nosological bibliography having been taken from it by the term, less long it is true, but of more than moderate length, of *tabes*. It is necessary, and it may be said in medical matters specially, to yield to the requirements of fashion. Let us then make every sacrifice in this direction, gentlemen, in adopting with closed eyes the barbarism and solecism* to which the use of this word forcibly condemns us.

The word "*tabes*" means "*consumption*" and nothing more. You will not have forgotten, gentlemen, what an important place "consumptive diseases" held in the medicine of old times, and I believe that even at the present time in some countries, notably in England, a trace of the interest formerly taken in these affections will still be found to exist in the popular language. At the end of the last and commencement of this century many

* The barbarism consists in the fact that, as Erb remarks, since the word "tabes" makes "tabis" in the genitive, one should not say "tabetic" but "tabic," or better still, "tabid." The solecism results from the fact that in France we put, for an unknown reason, the word "tabes" in the masculine gender, whereas in Latin it is a feminine noun.

chronic affections of the spinal cord, and no doubt of other parts of the nervous system were called by the name of *tabes dorsualis* (dorsal consumption). Attempts were then gradually made to class the different cases. A progressive elimination was made, and under the term "tabes dorsualis" Romberg described in 1851 many symptoms belonging to the type of which Duchenne of Boulogne, several years later, viz., in 1858, traced so masterly a picture under the name *progressive locomotor ataxy*.

Many other names have been proposed for this disease, some of which have been temporarily adopted. *Grey degeneration of the posterior columns* (this was at the time when the opinion of Romberg was followed, and too much importance was attached to the colour of the lesions in the nervous system, softening of the nervous tissue being regarded as red, yellow, white, &c.). *Posterior leucomyelitis*, the designation proposed by Vulpian, has never been at all generally adopted. *Sclerosis of the posterior columns* is a term which was somewhat favourably received by those authors whose faith in pathological anatomy was sufficiently strong to induce them to make this the basis of their classifications. The name, however, appears to me bad for two reasons, firstly, because many other affections of the spinal cord are accompanied by sclerosis of the posterior columns, secondly, because the lesions of tabes are by no means limited to sclerosis in this part of the cord.

The reasons on account of which the term *progressive locomotor ataxy*, after receiving general approval, has now fallen into disuse are the following: in the first place the claims made by German authors in favour of Romberg, whose description of "tabes dorsualis," although very incomplete, was certainly anterior to that of Duchenne; in the second place the greater knowledge which we now possess about the nature of the disease enables us to know that inco-ordination and ataxy of movement do not necessarily occur as symptoms.

The term *tabes* has thus prevailed. On the pretext that the word "dorsualis" is bad Latin, as Erb represents, should this epithet be replaced by "dorsalis"? I have little inclination to do this, and since the word "tabes" is itself an anachronism prefer preserving this anachronism in its complete state, and shall therefore continue to use the term "*tabes dorsualis*" as the authors did in the old time, and as my master Charcot does now.

However this may be, gentlemen, the claims of Duchenne are incontestable. It is true that Hutin, Monod, Cruveilhier, Horn, Steinthal and others had previously made some observations upon pathological anatomy connected with cases of tabes, and that Romberg had for the first time sketched out the disease. The merit of Duchenne is undiminished by these facts, since not only is his description very much more complete, but he also discussed *inco-ordination* as a separate symptom. He was the first to show, as he did by means of the dynamometer, that this inco-ordination was due, not to paralysis but to loss of the muscular sense. In a word he at the same time introduced into science the notion of *ataxy* as a special phenomenon. After the name of Duchenne of Boulogne, the French names which should in all justice be recalled are those of Trousseau, Jaccoud, Topinard, Marius Carre, Charcot, and Vulpian. It is my intention to give a more circumstantial history as regards each of the symptoms which will be separately studied. But this enumeration cannot be closed without the name of Westphal being also mentioned, who contributed so much to the diagnosis of this affection by the symptoms which he discovered.

SYMPTOMS OF TABES.

The *symptoms* of tabes are so numerous that in order to study them methodically it will be necessary to arrange them in systems or functions; however artificial this plan may be you will in my opinion prefer it. The general account of the symptoms of this disease has been so often given, and by such celebrated authors, that it would be presumptuous on my part to do the same; I shall on the other hand carefully discuss each symptom separately.

A. Motor Symptoms.

The motor symptoms which may exist are (1) *disorders of the muscular sense;* (2) *involuntary movements;* (3) *paralysis.*

1. Disorders of the muscular sense. These will be investigated, gentlemen, in the patient who is now before you for that purpose. After taking the foot, which for some years it has been impossible to use in walking, and moving it in different directions as upwards, downwards, and to the right or left side

in order to deceive the patient, I hold this limb in the air and ask him to tell me in what position the foot is placed: he tells me in answer that it is by the side of the other limb, whereas the two are separated by a distance of more than 50 centimetres (19·6 inches). This investigation, which can be varied to an unlimited extent, clearly proves that passive movements are not perceived by these patients. Again, when the patients are in bed they very often have no idea in what posture their limbs are placed unless they see them. In a word "they lose their legs in bed." These troubles, which result from disorders of the muscular sense, are designated *loss of the sense of posture*.

Somewhat analogous troubles are designated by the term *loss of the sense of differences in weight*. The meaning of this is as follows: the eyes of this patient being covered I cause him to extend his two hands, and in that upon the left side I place a weight of 50 grammes (1·7 oz.) and in the right one of 30 grammes (1 oz.), asking him at the same time in which hand the heaviest weight is placed, as you see he makes a mistake and tells me that it is the right hand. On account of the disorders of the muscular sense, of which I spoke just now, he cannot sufficiently realise how great an effort is made by his muscles to support one or other of these weights, and cannot therefore compute the difference which exists on the two sides. When in health we can, as you have often yourselves realised, gentlemen, distinguish very small differences between objects contained in the two hands; most persons can estimate a difference of $\frac{1}{20}$. In tabes, on the other hand, as Lussana has shown by a number of experiments in connection with this subject, the patient may be actually unable to distinguish differences of $\frac{1}{6}$, $\frac{1}{4}$, or even more.

How may disorders of the sense of differences in weight be detected?

One necessary precaution will be to use objects having as far as possible the same dimensions, such as two receptacles of equal diameter, in which weights can be placed, or scent bags composed of different substances of unequal density. Another plan adopted in such examinations is to make use of a balance, and to place the weights upon one of the scales, whilst the limb itself is placed upon the other. The patient is then asked to press the limb upon the scale in such a way as to counterpoise the

weights, and thus ascertain their amount, and the weights being varied to indicate whatever difference he can then perceive.

The disorders of the muscular sense are seen in these cases in their most simple manner; this, however, is by no means the case with respect to other muscular actions which we have still to examine.

Difficulty in standing is one of the symptoms which was first recognized, and even to-day is one of the best diagnostic indications of tabes, being termed the *sign of Romberg*.

We will ask this patient to stand and to place the feet near each other, even to join them should he be able to do so; this, as you see, he can only do with difficulty, and after much swaying from side to side. I then ask him to close his eyes; the effect of his doing so is immediate, and if, knowing what must inevitably happen, I had not placed myself by his side for the purpose of supporting him he would undoubtedly have fallen to the ground as a mass, after swaying once or twice from side to side.

You will observe how much the difficulty in standing is increased when the eyes are closed, and the movements cannot be controlled by sight. This control by means of sight can be to some extent estimated; thus, for example, if instead of asking him to close the eyes the feet are merely concealed, by placing a screen or sheet of cardboard horizontally directed in front of the sternum, there is less swaying than when the eyes are closed, but more than when no screen is interposed. It is often quite by chance that patients suffering from tabes become aware of the existence of these difficulties. When obliged to rise at night, there being no light in the room, they find to their great surprise that it is most difficult to maintain an upright position.

Another patient will tell you that he observed, whilst dressing in the morning, that when he placed anything over the eyes, as a sponge or wet towel, he commenced at once to lose his balance, and was obliged to take hold of the toilet table with one hand to prevent himself from falling; in this case again it was the closure of the eyes which produced the difficulty in standing. The same is again the case with some patients who are in danger of losing their balance at the moment when, in order to put on their shirt they pass it over their heads, and thus cover

the eyes during a few moments. The most numerous examples of this kind might be given, but I only wish to mention their existence, since they will enable you to fix in a retrospective manner the date at which the inco-ordination commenced.

I have used the word "inco-ordination," gentlemen, in speaking of the maintenance of the upright position, since this is only possible when the different muscles of the foot, leg, and pelvis are in continual action; hence the celebrated aphorism, "immobility is the finest movement of the soldier," is from a physiological point of view merely an expression of the pure truth. On account of the disordered co-ordination the contraction of the different muscles does not take place with the necessary precision, either as regards time or degree; equilibrium in the upright position is not unconsciously maintained, but becomes the object of special attention on the part of the patient. In this condition sight is of great service to the patient, by enabling him to rectify any bad position which loss of the muscular sense prevents him from otherwise perceiving, and hence occurs the difficulty or even impossibility of remaining in the upright position when the eyes are closed. As to the swaying movements which occur when a screen is interposed between the eyes and the feet, this is obviously due to a similar cause.

The motor disorders in this disease are due to analogous causes. They are the more interesting on account of the fact that in the first description of Duchenne of Boulogne, as you know, these held the leading place.

Before describing the motor disorders connected with the *lower limbs* I must first show you the gait of some of the patients whom I have caused to be brought to this amphitheatre. The ataxic gait* is well known to you all, and twenty times already the teachers in the hospital with which you are connected will have pointed out its characters. I will not, therefore, dwell at any length upon this point. You see with what difficulty this unfortunate patient takes the few steps which he has to make. He has just risen from his chair, and it is not without some difficulty that he maintains his equilibrium in the upright position. You will have observed that he

* MM. Demeny and Quenu have studied the ataxic gait in an interesting way by means of the photo-chronographic processes of M. Morey (Academie des Sciences, May, 1889).

D. The patient is then asked to hop, the eyes being open or closed.

E. He is then directed to descend a staircase and place himself at its foot, so that the least disorder in the gait may be perceived, while the awkwardness of the patient is increased in consequence of his being observed.

Such, gentlemen, are the ingenious methods which will sometimes be found of real service, and in doubtful cases I most strongly recommend your having recourse to what may be termed "Exercice à la Fournier."

Disorders of gait are not the only symptoms connected with the lower extremities. Charcot dwells in his lectures upon a singular phenomenon which not infrequently occurs in tabes. The patient, who had never experienced any difficulty in walking, suddenly feels his legs give way beneath him, and falls quite unexpectedly, perhaps in the middle of the road at the risk of being run over; his foot has not struck anything, nor has he stumbled, but the legs bent without being influenced by the will, or rather withdrew themselves entirely from its control. A moment later the will regained its power, and the patient rises and continues to walk as before. This is the "giving way of the legs" of English authors (Buzzard). It may occur early in the disease before the period of inco-ordination or they may both exist at the same time.

As regards the *upper extremities*, inco-ordination may be as pronounced in them as in the lower, but on account of the difference of their functions the symptoms produced by it are also different.

Speaking in a general way, extreme *awkwardness* is found to exist in the different movements. I have asked some of the patients now before us to button their jackets, and you perceive that they have great difficulty if they are able to do so at all. The same difficulty occurs in tying a knot, shaving, &c., and holding any small object between the fingers is in their case a real task, and requires all their attention. If the eyes are closed, or they are in any way prevented from seeing the hand, the object is almost instantly dropped, and usually without their perceiving it.

One motor disorder in insular sclerosis to which Charcot never fails to draw attention in his lectures is the *mode in which*

these patients take up an object. If you ask one of them to take up an object of small size, as, for instance, a pencil, you

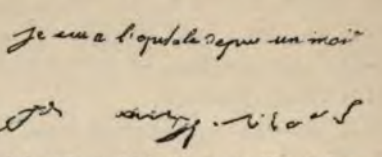


Fig. 107.—Example of the effect which the controlling influence of sight has upon the inco-ordination of movements in tabes. The upper line was written by a patient whose eyes were open. The eyes were then closed and he was asked to write the same words again (lower line). The difference between these two lines shows how much the inco-ordination had increased in the second case. (Damaschino collection.)

will see that he does so in a peculiar way. Whilst you or I reach the object almost insensibly and without abruptness by moving towards it one hand, of which the thumb is opposed to the fingers, and only separated from them by a distance equal to 2 or 3 times the diameter of the pencil, the patient opens the hand, completely separates the thumb from the fingers, and then, moving the hand close to the object to be taken, causes it to "hover," as it were, for a moment above it, and then falling rapidly to seize it unawares as if it were capable of taking flight.

2. INVOLUNTARY MOVEMENTS *resembling those which are termed "athetoid."* Such, gentlemen, are the principal manifestations of inco-ordination; but these are not the only disorders connected with the muscular system which may be observed in tabes; I must mention a symptom which is less known, but which may undoubtedly exist. I mean the movements resembling those which are termed "athetoid," and muscular shaking which occur in certain patients suffering from this disease. Rosenbach* had already mentioned them in 1876, and in his *Revue* of 1877, upon Athetosis, Grasset† called attention to this observation. In a memoir specially devoted to this subject, Audry‡ studied these movements, which he attributes to lesions in the lateral columns of the cord, corresponding with those in the posterior

* Rosenbach, *Virchow's Archiv.* LXVIII.

† Grasset, *Montpellier médical*, 1877.

‡ Audry, *Revue de médecine*, 1887. During the publication of these lectures a new work appeared by Audry upon double athetosis, in which the subject of these movements in tabes was very completely investigated.

column. Lastly, at a more recent date, in 1890, Laquer* published the account of two other cases in which these movements occurred. Other cases have been noted by B. Stern,† and by Oppenheim.‡ I myself had the opportunity of observing a very clear case, in which these movements occurred, (not published) in 1885, since which time I have seen it twice, though not to such a pronounced extent as in the former case. From the works to which I have just alluded, and the knowledge obtained from these few cases, I feel justified in telling you that objectively the condition is not one of true athetosis, but that a series of involuntary movements occurs somewhat frequently repeated, and usually more or less identical with each other in the same patient. Thus whilst in one patient, for instance, a finger will be raised, and almost immediately fall again, in another, some degree of pronation or supination of the hand will occur. In reality these movements do not present in any way the obligatory, and, to a certain extent, spasmodic character of those which occur in true athetosis. Nothing in their form specially characterises the movements, wrongly called athetoid, occurring in insular sclerosis; the only fact about them which specially attracts attention is that they are involuntary, and most often unperceived by the patients. On account of the absence of spasm I am not inclined to attribute these movements to extension of the lesions to the lateral columns; I am more inclined to connect them with loss of the muscular sense, the amount of nerve force influencing the muscles being regulated in a very imperfect manner.

3. Paralysis. Lastly *paralysis*, a symptom of quite a different character, is one of the motor disorders occurring in tabes which must be studied. Until the present time, gentlemen, in order to fix more completely in your mind the true character of the motor disorders in tabes, you have been told that these were solely due to inco-ordination, and that even when they were most pronounced the muscular power was in no way diminished. To such an extent was this the case that in an examination with the dynamometer the numbers often indicate considerable muscular strength. This, gentlemen, was,

* Laquer, *Wanderversamml. der S. W. deutschen Neurologen*, 1890.
† B. Stern. *Arch. f. Psychiatrie*, XVII.
‡ Oppenheim, *Berl. klin. Wochenschrift*, 1889, p. 965.

and is still, perfectly true, and a clear distinction must be made. Not only inco-ordination, but perhaps associated with, and at the same time quite independently of it, certain paralytic symptoms may occur in tabes. Such a fact is by no means rare, and in 224 cases observed by Fournier, he found this complication to exist in no less than 41 patients. The enumeration of these cases is so interesting that I must place it before you :—

Hemiplegia	18 cases
Unilateral paralysis of the portio dura	8 „
Paresis of the tongue	3 „
Monoplegia	3 „
Laryngeal paralysis	2 „
Paralysis of the extensor muscles of the wrist	1 „
Paralysis of the deltoid muscle	1 „
Paraplegia	5 „

You will notice that in this list no allusion is made to paralysis of the ocular muscles, which will be discussed in a subsequent lecture.

With such a number of cases before you it will be easy to understand, gentlemen, that paralysis in tabes has been recorded by numerous authors, of whom I need only mention Marius Carre, Pierret, Grasset, Debove, &c.

The seat of the paralysis varies, but it presents certain characteristic features which were clearly indicated by Fournier.

It is usually *benign* in character, and should often be termed paresis rather than paralysis.

Its *duration* is *short*, varying from a few days to a few weeks, after which time the paralysis often entirely ceases to exist even spontaneously and without treatment.

Exceptions, however, may occur, and it must not be expected that in every case the paralysis which exists in tabes will so completely disappear. The subject of paralysis in tabes being usually treated with few details in medical works, I shall ask your permission, gentlemen, to say a few words about *hemiplegia*, *paraplegia with sudden onset*, and *paralysis* affecting but one nerve, or a somewhat limited region.

Hemiplegia has been specially studied by Debove* and Miss

* Debove, *Progrès médical*, 1881.

Blanche Edwards.* It shows itself in three forms: A. *Permanent* hemiplegia which may be accompanied by secondary contracture; B. *Transient* hemiplegia unassociated with hemianæsthesia; C. *Transient* or *permanent* hemiplegia *with anæsthesia* of the *skin and special senses.* As regards the pathological anatomy a focal lesion may be found in the cerebral hemispheres or pons varolii (hæmorrhage, softening), or there may be no appreciable lesion; it is certain that in the latter case, specially after hemianæsthesia, as Charcot observed, the patients were merely suffering from *hysterical hemiplegia* associated with tabes, and analogous to that already observed to be associated with insular sclerosis.

However this may be, gentlemen, one general rule is observed in both varieties: the tendon reflexes, if lost before the onset of hemiplegia, remain absent after its occurrence; the spasmodic tendency, in short, which usually accompanies hemiplegia is unable to counteract the atonic tendency which exists in tabes. In some cases, however, the patellar tendon reflex has been found to return, and even to be excessive† after hemiplegia.

Paraplegia with sudden onset is a symptom which should be known to occur or serious errors in diagnosis may be made. I myself well remember the surprise which I felt in 1879 when, coming into the service of Charcot with the knowledge which was then current about this disease, I found myself in the presence of such a case. Yes, gentlemen, notwithstanding the classical opinion that paralysis only occurs in the later stages of tabes, in the period which has been arbitrarily termed the third period; notwithstanding this opinion, I say, you will sometimes find that paraplegia is the first symptom which attracts the attention of the patient. In his lectures upon the præataxic period of tabes, Fournier, who has carefully studied the different cases, quotes a very instructive observation made by Albert Robin. The case is that of a man who, "whilst hunting on foot, when strong and in good health, attempts to leap over a ditch, and in doing so falls heavily into it. At first he attributes this to clumsiness, or to taking off badly, but soon feels his legs to become weak and benumbed. He limps, and has no longer

* Blanche Edwards, *Thèse de Paris*, 1889.

† Goldflam, *Ueber das Wiedererscheinen von Sehnenreflexen bei Tabes, &c. Berliner Klin. Wochschft*, 1891, No. 8.

the strength to walk, while the ground appears to sink under his feet. A second ditch is reached, and instead of clearing it at one leap, as is his habit, he is obliged to descend and climb up the other side step by step. He feels that every moment it becomes more difficult to continue his course. It is only by a great effort that he can return home, dragging himself along rather than walking. He goes to bed and wakes the next morning suffering from 'almost complete paraplegia.'" I, for my part, have also had the opportunity of observing a case which was quite analogous. The patient was a native of South America, who, whilst visiting his property, wished to leap a ditch and suddenly felt his legs give way beneath him, fell, and became affected by paraplegia, as he remained during several months; when able to get up he visited Europe, presenting at that time all the symptoms of confirmed tabes. In other cases paraplegia occurs at night, and the patient, who until then had walked fairly well, is very surprised to find in the morning that he can no longer maintain the upright position.

This form of paraplegia often obeys the general rules which I have just mentioned, that is to say, it is slight in degree and after a certain time more or less completely ceases to exist, this, however, not being always the case as the paralysis is in some cases found to persist.

To what is this symptom due? Scarcely anything has been written upon the subject, and one can only make suppositions. Is there a focal lesion in the grey matter of the spinal cord associated with changes in the posterior columns? This is possible but has not by any means been proved to be the case.

As regards *paralysis affecting but one nerve* or a limited region, the most frequent form, as you will see, is that affecting the ocular muscles. This will be considered at a future time; at the present moment I will only allude to *unilateral paralysis of the facial nerve*, which sometimes occurs and may affect all the branches of the nerve, as when due to a peripheral cause. It must be observed, however, that this so-called facial paralysis in tabes is often only the glosso-labial hemispasm of hysteria. *Paralysis of the musculo-spiral nerve* observed by numerous authors, *paralysis of the thenar muscles supplied by the median nerve* (E. Remak, Möbius), unilateral *paralysis of the muscles of mastication* (Schultz) must also be mentioned. These forms of

paralysis are also, as in the first mentioned varieties, benign in character, and liable to diminish or cease of their own accord. The electrical reactions are very variable, being at times normal, while in some cases the reaction of degeneration exists in a more or less complete form, even though the paralysis is of but temporary duration.

With respect to these cases of local paralysis I must repeat, gentlemen, the observations already made in connection with the hemiplegia which occurs in tabes. It is very probable that a certain number of these cases also are due to *hysteria* alone, and you must always be on your guard, and consider this alternative in forming your diagnosis.

LECTURE XV.

TABES.

SYMPTOMS (*continued*).

B. SENSORY SYMPTOMS. I. SENSORY SYMPTOMS WHICH ARE PURELY SUBJECTIVE.—*a. Intermittent pain:* α. Seated in the trunk, the limbs, or the face: lightning, shooting, wrenching, burning, often paroxysmal. β. Pain connected with the viscera (to be specially studied when the disturbances connected with each organ are considered). γ. Sudden loss of muscular power. *b. Persistent pain:* girdle-pain, bracelet-pain, gaiter-pain, burning-pain. *c.* Abnormal sensation of tingling or numbness. II. SENSORY SYMPTOMS AMENABLE TO OBJECTIVE CONTROL.—*a. Anæsthesia* (analgesia) researches of M. Oulmont. *b.* Hyperæsthesia; hyperæsthesia in islets, relative hyperæsthesia, relative anæsthesia. *c.* Paræsthesia: retarded transmission of impressions, altered character of sensations, localization of sensations impossible, dissociated anæsthesia, return of sensations, non-correspondence between the number of sensations and that of the impressions. α. Sensory tetanus. β. Polyæsthesia: summation of impressions, loss of power to respond to impressions.

SENSORY disorders form the leading part of the clinical symptoms in tabes, the lesions of which, as you know, are specially connected with the sensory tract. These disorders, mentioned by all the authors who have written about tabes, differ much from each other and vary considerably in different patients. They should therefore be somewhat methodically described, and on that account I shall make use of a classification which, I must warn you, will be more or less artificial.

I.—SENSORY SYMPTOMS OF A PURELY SUBJECTIVE NATURE.

a. INTERMITTENT PAIN. ***a.*** *Seated in the limbs, trunk, or the face.* These pains may fairly be classed among the earliest and most important symptoms of this affection. Their character varies. The *lightning* pains are, as their name indicates, pains which pass away like a flash of lightning, and on account of their short duration have ceased by the time that the cry is heard which they often draw from the patient. They do not remain localized in one point, but extend for a certain distance sometimes along the whole, or almost the whole length of a limb.

The *shooting* pains are also rapid and transient, but they are more locally seated, occurring and ceasing in one spot, and being often compared by the patients to the prick of a pin or even the stab of a dagger. This, gentlemen, is not merely a mode of speech, and, in fact, patients are not rarely met with who tell you that at first they often turned sharply round to see who had assaulted them, so much did the sensation resemble that of being stabbed by a knife.

The *wrenching* pains are also localised but distinguished from the preceding by the fact of their occurring less rapidly. They can be, to a certain extent, analysed, the patient having the "sensation of something being screwed into the tissues," the penetration seeming to be accompanied by a twisting motion.

The *burning pains* consist of sensations resembling those of being burnt, and are at times very pronounced. These vary in the same manner as the forms of pain already mentioned.

The *seat* of pain may be, as already stated, the limbs, specially the lower limbs, the pain being felt either in the thigh, or below the knees. In the upper extremity the seat of election is usually the inner side of the forearm and hand; pain is liable to occur also in the trunk, and at times is felt in the face.

According to the description of the greater number of patients, they appear to arise less in the skin itself than underneath it, and even in the muscles, bones, or articulations.

The pains are at times isolated, and arise one by one; at other times they arise at the same time in many points, and then constitute, as the patients say, "volleys of musketry."

Most often they occur in *paroxysms*, during the interval between which the patient is at times completely free from pain; then when the paroxysm commences, the pains occur in more or less compact groups; each paroxysm varies in duration from half an hour to several days. They are usually of spontaneous origin, sometimes occurring so regularly that the patients expect them at a definite period; sometimes, however, they may be due to exposure to cold, change of temperature, fatigue, or emotion.

The intensity of the pain varies much, and corresponds in no absolute degree with the severity or duration of the disease; usually, however, the pain occurs early in the complaint, and often more or less completely ceases when the period of inco-

ordination commences, though they may persist during the whole course of the disease.

β. Visceral pains. Paroxysms of pain at the anus, or in the testis, ovary, clitoris, urethra, bladder, stomach, intestine, &c., should be described, but it seems to me preferable to speak of these when the other visceral symptoms occurring in the course of tabes are considered; and at the present time I shall only mention certain pains connected with the muscles known by the name of—

γ. Paroxysms of muscular exhaustion. These were first mentioned by Pitres* in 1884, and have been met with in a certain number of cases since that time. According to Pitres they specially exist at the onset of the disease; occurring abruptly, and without obvious cause, persisting during some hours or days, and ceasing without being followed by any remaining sensation of fatigue. These paroxysms occur in the same way as those already described at varying intervals. They consist of a most painful sensation of fatigue and muscular exhaustion, analogous to that felt by healthy persons after too violent or prolonged exercise. The seat of the sensation is in the limbs or erectores spinæ muscles, and it may be so severe that energetic and vigorous persons are obliged to take to their bed in the middle of the day, and rest in the dorsal position until the paroxysm has ended.

b. PERSISTENT PAIN. The character of these pains varies, but they often tend to exist in a circular form; such is the famous "*girdle-pain*," a feeling of constriction as if a bodice of iron was tied tightly round the chest or waist of the patient, and which is sometimes of extreme severity. Fournier quotes a case of this kind, in which the girdle-pain was severe enough to produce dyspnœa, resembling that which occurs in serious affections of the heart or lungs. In the limbs the feeling of constriction may resemble that produced by a *bracelet* in the upper, and by a *gaiter* in the lower extremities. Not infrequently areas exist in the limbs, or more often in the trunk, of greater or less size, in which a persistent and burning sensation of the most painful nature exists.

c. ABNORMAL SENSATIONS. The most usual of these sensations are those of *tingling* and *numbness* which are felt in certain

* Pitres, *Progrès médical*, 1884.

parts, specially in the limbs, and most often along the *ulnar border* of the forearm and hand. Sometimes, again, the patients feel a strange sensation of cold between the skin and subjacent tissues, "as if water was passing under the skin"; lastly, but more rarely, an itching sensation is felt in different parts of the limbs.

None of this class of symptoms, except the feeling of numbness upon the ulnar side of the forearm, is of any great importance as far as the diagnosis of the disease is concerned. In some cases, however, the existence of an area of numbness, such as the sensation of a veil, or spider's web, upon the face or penis, has indicated the existence of tabes at the most early period of the complaint (Charcot).

II.—Sensory symptoms amenable to objective control.

(*a*) *Anæsthesia* (analgesia). Anæsthesia occurs not only at the surface of the skin but also in the deeper parts, viz., the muscles, bones, and articulations. Thus it is possible to make the muscles contract in some patients by means of a strong electric current without producing pain, and, as I shall have occasion to remind you, fractures and dislocations are most often painless in patients suffering from tabes. Lastly it will be seen that this analgesia favours the occurrence of certain trophic disorders (callosities, perforating ulcers, arthropathy, &c.).

When it is very pronounced and its existence is accidentally revealed to the patient he may know that this analgesia exists, but most often it has to be sought, and in the most careful manner.

Although the seat of the anæsthesia varies and may occur in any part of the body, and in different forms, it is most often found clinically to prefer certain parts and to be disposed in a special way. Oulmont has published some interesting remarks upon this subject from which, though absolute rules cannot be laid down, useful information may be gained. In a few words these are the conclusions at which he has arrived:—

Of 20 women suffering from tabes under the care of Charcot, taken recently by hazard at the Salpêtrière Hospital, he found anæsthesia to be totally absent only in 3 patients.

With respect to its seat it existed 17 times in the *limbs*, 16 in the *trunk*, and 13 in the *head*.

As regards *symmetry* in the seat of the anæsthesia he states that in the limbs anæsthesia existed almost exactly in the same part, over a corresponding area, and in almost identically the same form; the special difference being in the intensity. In the *trunk* the areas sensibly correspond upon the two sides, or if it is in the middle line the area extends for the same distance on either side. In the *head*, on the contrary, there is usually no symmetry, the areas of anæsthesia being often unilateral, or if existing upon the two sides being of very different intensity.

The parts of the body at which the anæsthesia most often occurred (the points of election) were found to be:

In the *head*, specially the *cheeks* and *suborbital regions*.

Anæsthesia seldom occurs in the neck.

In the *trunk, anteriorly* the islets of anæsthesia are specially found in the *breasts*, and at times round *the umbilicus*, while the parts which usually escape are an area shaped like a band in front of the sternum, and one of similar form in each inguinal region. *Posteriorly* anæsthesia is not infrequently found to exist over the *shoulders*. Lastly an islet of *hyperæsthesia* is not rarely observed between the shoulders, and another in the lumbar region; and these two islets sometimes join, in which case a long band or islet of *hyperæsthesia* may exist along the whole length of the spine.

In the *upper limbs* the areas of anæsthesia occur most often in the *fingers*, even when found in no other part of the limb. In the forearm the *ulnar anæsthesia* must be specially mentioned, which was first brought to light by Charcot. Its seat is along the inner border of the forearm, and it may extend along the inner margin of the hand and little finger. The *arm* is usually less affected than the forearm. Even when the anæsthesia is very extensive in its distribution, areas nearly always exist in the palm of the hand and at the bend of the elbow, in which the sensibility is preserved.

In the *lower limbs* it is interesting to find that the sensory disorders are nearly always more pronounced on the posterior than on the anterior surface. The areas of anæsthesia are most often found in the *sole of the foot* and upon the *heel* or *toes*,

whilst the back of the foot is seldom affected. Very frequently there is some *hyperæsthesia* upon the arch of the foot. In the legs, the parts most often affected are the *knees* and *malleoli*. The thighs are less liable to be affected by anæsthesia than the legs below the knees, their *inner surface* being the part which remains longest unaffected in the whole zone which corresponds to the adductor muscles.

You will observe, gentlemen, in the figures which I now place before you, the areas of anæsthesia found in different patients suffering from tabes, that the seat of these areas is not over the course of a nerve or nerve branch, nor is it strictly limited to the area of distribution of the latter; most often, on the contrary, the affected part extends over areas supplied by different cutaneous nerves, and you will find that in some cases the area of anæsthesia has the shape of a leg of mutton, but there is nothing else in its appearance analogous to that described by Charcot as existing in hysterical paralysis.

b. Hyperæsthesia, or rather *hyperalgesia*, since, as Leyden truly observes, the tactile sensibility never becomes more acute in tabes than in the normal state, the sensibility to pain being alone increased.

The hyperæsthesia usually exists in areas, which are of much smaller size than those of anæsthesia. At the end of a certain time again the hyperæsthesia may be followed by anæsthesia, the duration of the former condition being usually far less than that of the latter.

In these areas the hyperæsthesia may exist in a most excessive degree, the gentlest friction with the hand or clothes, even a current of air may produce sensations of acute pain. In some cases this condition is one of torture to the patient, and the symptom of which he specially complains.

Sometimes again these areas of hyperalgesia constitute a centre from which the lightning pains start; a reservoir, as it were, of pain, which is but too often inexhaustible.

According to Erb, when hyperæsthesia exists to a somewhat pronounced degree in tabes, the meninges probably participate in the morbid process.

Under the name of *relative hyperæsthesia* Leyden, who has carefully studied the sensory disorders which exist in tabes, has

described cases in which a slight prick is scarcely perceived, whilst one which is at all stronger produces very intense pain which in no way corresponds with the severity of the injury.

On the other hand, Berger has found that in other patients suffering from tabes, pricks of moderate strength are well perceived, whilst those which are more decided produce no pain. As compared with the preceding condition this symptom might be termed *relative anæsthesia*.

So far we have only spoken of the hyperæsthesia which is connected with the effect of a prick. It may also be observed after other impressions upon the skin, specially those produced by change of *temperature*, this form of hyperæsthesia being specially pronounced after the influence of *cold*.

c. Paræsthesia. Under this term I shall include numerous disorders of sensibility which I am about to enumerate, confining myself, however, to the few observations which it is necessary to make in order that you may understand the subject which is being considered. These disorders have been specially studied by Leyden, Berger, Binswanger, and the different authors who have devoted their attention to tabes.

The *retardation of sensations* is frequent in tabes, specially in the lower limbs. It is easily discerned by directing the patient to call out as soon as the pain produced by a prick is felt, and measuring the length of time which passes between the moment at which the prick was made, and that at which the cry announcing its sensation occurred. The length of this interval is usually 2 or 3 seconds, but in some cases it amounts to 8 or 10 seconds, or even more. Richet states that in the same patient the interval is of greater length when the foot than when the thigh is pricked. Is this difference due to the greater length of the path through the nerves in the one case than in the other, or, as seems more probable, to the fact that the sensory disorders are very much more pronounced in the foot than in the thigh? It must be observed that this difference is liable to very great variation in the same patient from one moment to another.

The sensations produced by an impression may be all retarded, or this may perhaps occur in only one or more forms of sensation. Thus, for example, a patient suffering from tabes with closed eyes is pricked, and he states that he experienced the

sensation of contact at once, but it is only after 3 or 4 seconds or more that he shows by the contraction of his features, and by his cry that a sensation of pain is now felt. In this case the sensibility to touch was well preserved, while the sensibility to pain was much retarded. The same fact may be observed in connection with sensibility to temperature; when a fragment of ice is placed upon the skin the patient feels at once a sensation of contact, and it is only after an interval that the ice is perceived to be cold.

Sensory metamorphosis is said to exist when patients suffering from insular sclerosis cannot clearly recognise the nature of a sensation perceived. Thus, for example, a prick or pinch may seem to them a more or less severe burn.

The *inability to recognise the seat of sensations* is not an infrequent symptom, and patients suffering from tabes may feel the sensation of a prick in the foot when in reality a pin has been passed into the calf. It may, again, happen that impressions are not perceived over the whole part where they exist. Thus a scratch some centimetres (1 centimetre = ·394 in.) in length may only be felt as a simple prick.

The following symptom is called by the name of "*dissociated anæsthesia.*"

In a patient suffering from tabes who cannot feel a prick the sensibility to temperature is preserved, or *vice versâ* (M. Parmentier has recently published such a case), or while the sensation of a prick is preserved compression is not perceived. Tactile sensibility is usually lost later and in a less degree than sensibility to pain, though it is also liable to be affected. This notably contributes to increase the awkwardness in using the upper or lower limbs (the ground is felt by the patient as a carpet, cotton-wool, indiarubber, &c.).

The *recurrence of sensations* should be specially mentioned, since, from a clinical point of view, a knowledge of its occurrence is indispensable in order that a complete examination of the sensibility may be made in these patients. When, in fact, during the examination of a patient suffering from tabes a certain number of pricks has been made, a moment often arrives when the patient will feel the sensation of a prick although none has been made, in a word he feels that he is pricked when this is not the case. The previous impressions

sensation." If, for example, what happens when that prick which is of such strength produces a sensation while the second or third action of exactly the same strength as the preceding,

are clearly perceived by the patient. This symptom is of the same nature as those in which impulses, which are of insufficient strength when isolated to produce the effect desired (sensation, muscular contraction) join their effect together, and experiencing that which physiologists term a "summation," finally exert upon the nerve centres sufficient influence to determine the effect desired.

The *loss of power to respond to impressions* is also a symptom, the existence of which should be known, since it may completely vitiate the results of an examination; it is exactly the reverse of the "summation of impressions"; in this case in fact the sensibility is diminished by continuation of the same exciting force. E. Remak, B. Stern, have studied these facts, and the first of these authors showed that in some patients suffering from tabes, a feeble electric current which was at first well perceived soon ceases to be so; if the strength of the current is then increased, it is again perceived, soon however ceasing to be felt, the same occurring again and again as the strength of the current is changed. At times this diminution in the power of appreciating impulses occurs in eclipses; the temperature of a hot object applied to the skin is at first clearly felt, but after some moments is no longer perceived; if, however, the object remains in contact with the skin it is again felt, although no change in the temperature has occurred, then, again, it is not perceived after some moments, the same changes occurring as time goes on.

In all these examinations, which require the most careful attention, it must not be forgotten that, owing to the effect of certain purely physical influences, the sensibility may be much modified; it is thus that anæsthesia may be increased by cold, whilst, on the contrary, heat, friction of the skin, the passage of the electric current may cause it to cease for a certain time, or at any rate diminish its intensity.

Such, gentlemen, are the different sensory symptoms which occur in tabes, to which I wished to call your attention; if I have described a few of these at some length, considering the importance which they assume in the course of this disease, and the frequency with which they occur, it is because they are, as a rule, but little known, and special interest is thus attached to their description. Nor do I doubt that rare as some of these

symptoms seem to be, they are found to exist more frequently if carefully sought in all the patients suffering from tabes who come before you.

I must ask you to remember, gentlemen, in every case, that in discussing the sensory symptoms of tabes, my object has been of a didactic rather than clinical character. It is little probable that you will ever find these different symptoms associated together in the same patient, and one cannot too much insist upon the irregularity and variety of the sensory symptoms in this disease. In some patients the symptoms are extremely pronounced, while in others they scarcely exist; in a certain number after having been very pronounced they finally almost completely disappear. Unfortunately, whilst we can recognize the various forms in different patients, we have to confess our ignorance as regards the cause of these varieties; we are actually unable to indicate the anatomical cause on account of which one patient suffers intensely whilst another is almost entirely free from pain.

LECTURE XVI.

TABES.

SYMPTOMS (*continued*).

C, DISORDERS OF REFLEX ACTION. *Patellar tendon reflex.* Precautions to be taken in the attempt to obtain it: position to be given to the thigh; percussion of the tendon, means of detecting the muscular contraction which is thus produced, principal causes of error. Pseudo-tendon reflex of Westphal. Nature of tendon-reflexes; anatomical notions in connection with the nerves and nerve endings in the tendons. Region of the cord in which the afferent path of the patellar tendon reflex ends: external bandlets (Wurzeleintrittszone), the limits of this region in the transverse and longitudinal direction. Centrifugal paths. Conditions which may influence the production or intensity of the patellar tendon reflex: (a) local, (b) general. History of the discovery of the patellar tendon reflex; sign of Westphal, theories in connection with this subject, reasons confirming its reflex origin.—*Plantar reflex.*

GENTLEMEN,—The disorders connected with reflex action are amongst the most important which occur in tabes from a diagnostic point of view; the symptoms which are thus in fact produced occur at the moment, and in the conditions which we ourselves choose; such production is the cause of their existence, and on that account a degree of precision is acquired which the different "symptoms" developing spontaneously, and occurring, so to speak, when they please, cannot pretend to have.

You are aware, gentlemen, that reflex movements may be obtained throughout the whole length of the cord, and even by means of the brain; some of these, again, resemble each other so much that they may be classed together. This, in fact, is the case with the *tendon reflexes*, and it has been usual to investigate specially those of the knee, the tendo-Achillis, that of the wrist, elbow, and not the tendon reflexes of all the different muscles. In the same way, as regards the cutaneous reflexes, the *plantar* and *abdominal reflex* are specially tested, without much attention being paid to the reflex movements which may be produced by irritation of the skin in the intermediate region. Such is the case in studying tabes, in which disease some of the reflexes are of special interest, and it is these only, gentlemen, which will be now considered. As you know the *reflexes connected with the*

and are amongst the most important, and should perhaps be considered in this series. I prefer, however, to speak of them at a later date when the other ataxic symptoms occurring in tabes will be considered.

PATELLAR TENDON-REFLEX. My experience both at the hospital and in examinations has enabled me to know that amongst the junior members of the medical profession there is not a student who fails to examine the patellar tendon-reflex; very few, however, do so methodically: a very small number know or are interested in what it really means and its true significance. It will not therefore be useless in my opinion, to review rapidly, in speaking of tabes, the principal facts which are now known about this symptom.

The patellar tendon-reflex (knee-jerk, knee-phenomenon) consists roughly in the contraction of the quadriceps femoris, which is produced by percussion of the Ligamentum Patellæ.

Certain conditions are necessary in order that it may be obtained. Hence, from a practical point of view, certain precautions must be taken.

In the first place the fibres of the *quadriceps femoris* must be as completely *relaxed* as possible. To effect this the lower limb is placed in one of the following positions: the leg to be tested is placed by the patient across that of the opposite side: this is the plan usually adopted, and is in most cases sufficient; in some cases, however, the patients have so little intelligence, or are so sensitive that relaxation of the quadriceps cannot be obtained in this way. Should this happen, recourse must be had to one of the following plans.

a. The patient is made to sit upon a hard and sufficiently heated seat (usually a table), in order that the feet may not touch the ground, and care is taken that the patient is seated completely, and in such a way that the edge of the seat or table touches the hollow of the knee.

b. The observer sits upon a chair close to the table or other seat of the patient, and placing one knee below the calf of the leg to be tested raises it to such an extent that the foot does not touch the ground.

c. The observer raises the thigh by means of the hand placed below the tendons of the biceps and semi-membranosus, while the patient is directed to rest the leg upon it: the

extension of the thigh upon the pelvis which the patient thus produces has often for its consequence to relax the quadriceps femoris which is itself a flexor of the thigh upon the pelvis.

When the patient, instead of being seated, is in bed, he is enjoined to leave the limb relaxed, and raising the thigh the lower limb is placed in such a position that the hip and knee joint are both in a state of flexion; percussion is then effected. If the patient cannot relax the limbs sufficiently he is made to sit upon the edge of the bed, care being taken that he is so entirely seated that the edge of the bed touches the hollow of the knee.

When the relaxation of the quadriceps is obtained by one of the means which I have just mentioned, the second part of the process must be effected, viz., *percussion of the Ligamentum Patellæ*. This percussion should as far as possible be made upon the middle part of the tendon at the same distance from each of its insertions; if by simple inspection of the region this point is not accurately indicated it can be detected by means of the finger. The blow may be given by means of the ulnar border of the hand or the extremity of one or more fingers, or some instrument; that which is most often used is the percussion hammer of Skoda; in England a stethoscope is often used, of which the ear piece is hollowed out in such a way as to contain an indiarubber band. Whichever plan is adopted the blow on the tendon should be slight at first but always "sharp," becoming gradually stronger if necessary; the strength of the blow necessary to produce muscular contraction is in fact an important consideration. You will be able to see that when the reflexes are excessive a much feebler blow is required than when they are diminished.

The percussion having been effected, it is necessary to *detect the muscular contraction which is thus produced* (patellar tendon-reflex). Most often this is easily done since the contraction of the quadriceps produces a more or less pronounced movement of extension in the leg, and, as on account of the relaxation which follows in all the muscles of the lower limb, the leg almost immediately falls again, an oscillating movement of the whole lower extremity is clearly perceived. Sometimes, however, no such movement is produced; it must not be inferred from this that the reflex is absent; the contraction of the quadriceps may in fact occur, but be too weak to produce a

movement of the leg. Hence it is necessary to lay bare the leg and to keep the eyes fixed upon the anterior surface of the thigh when the blow is given, in order to see whether any prominence of the muscle occurs at that moment. In some cases when this prominence is slight, and the patient somewhat stout, it may not be perceived even by a watchful eye. Palpation may then be of service, and the left hand being placed upon the body of the muscle enables it to be known whether even the slightest contraction has been produced in the quadriceps by the blow given with the right hand.

A certain number of plans and instruments have been invented for registering or measuring the patellar tendon reflex; these, however, are but rarely used at the bedside, and I shall therefore only mention to those of you who may be specially interested in this subject the process employed by Brissaud, and described in his thesis, and the recent works of Waller and Bechterew.

You now know, gentlemen, the plan which must be adopted in order to detect and observe the patellar tendon reflex; do not, however, suppose that this is always an easy matter as at first sight it appears to be, and I must now mention the principal causes of error.

In some cases, even after taking all the precautions mentioned, no reflex movement can be obtained on account of its having been impossible to place the muscles of the lower limb in a sufficiently relaxed condition, and you must be careful not to consider these cases as examples of abolition of the reflex. As a general rule, whenever you are not absolutely certain during the observation that the fibres of the quadriceps are sufficiently relaxed, before coming to a definite conclusion about the condition of the reflex, repeat the examination in various ways until you are certain that the muscles of the leg are flaccid; it is only when this is the case that you are justified in forming a positive opinion.

One is often liable to mistake for a contraction of the quadriceps the movement of the leg, which is produced in a purely mechanical way by simple communication from the blow given. With slight attention, and by varying the strength of the blow, this mistake can be easily avoided.

Lastly, Westphal has given the name of *pseudo-tendon-reflex* to cases in which a blow upon the skin of the knee produces a

reflex movement of the leg analogous to that which occurs in the patellar tendon reflex. If this is thought to be possibly the case, it would suffice, in order to solve the question, to make a somewhat thick fold of the skin with the fingers of the left hand in front of the patellar tendon, and to make the blow fall, not upon the tendon, but upon the fold of the skin; should a movement of the leg be thus produced, it is certainly due to the pseudo-tendon-reflex of Westphal.

Such are the principal points to which attention must be paid in making this investigation. Let us now attempt to determine the nature of what occurs, and to ascertain what signification should be attributed to it.

It is said to be a *reflex movement*. What then is a reflex movement? a movement due to the motor portion of the cerebrospinal axis being caused to act by irritation of a sensory organ transferred to that axis by afferent nerve fibres. A *tendon reflex* is a reflex of which the origin is a special form of irritation of the sensory organs contained in the tendon.

Let us consider what in this special case (patellar reflex) will be the seat of the irritation and consequent reaction.

The first part of the process, as you have seen, is the blow on the patellar tendon; this blow constitutes the primary irritation. That this irritation may be transferred to the nerve centres it must so influence the peripheral receptive organs that the vibration will react upon the cord, through intermediate nerves.

Do the tendons then contain nerves* and nerve endings,† which can act as organs of peripheral reception? The anatomical knowledge obtained during the last twenty years enables us to answer this question with certainty.

A certain number of authors have already noted the presence of nerves in tendons; in 1875 Sachs stated that these nerves possessed free extremities seated in the thickness of the tendinous laminæ, and at their surface; a short time afterwards Tschiriew noted the existence of swellings in these extremities. On the other hand Golgi* discovered in 1878 the existence of special organs in the tendons (musculotendinous bodies), analogous to, and in close relation with the *Pacinian corpuscles*, which are also found in large number in the tendinous laminæ.

* Sachs, *Die Nerven der Sehnen.—Reichert's und Du Bois Reymond's Arch.*, 1875.
† Tschiriew, *Arch. f. Psych.*, V, p. 803.

According to the very detailed description of Cattaneo,† the principal characters of these *corpuscles of Golgi* are as follows:—

They are fusiform bodies enlarged, but flattened on their two faces parallel to the surface of the tendon, of variable dimensions (from 80 to 100 μ in length, and 50 to 400 μ in breadth), and can consequently be detected by means of a magnifying glass. They are usually seated upon that surface of the tendon in which the muscular fibres are inserted, and principally close to where the muscular fibres join the tendons. One of its extremities

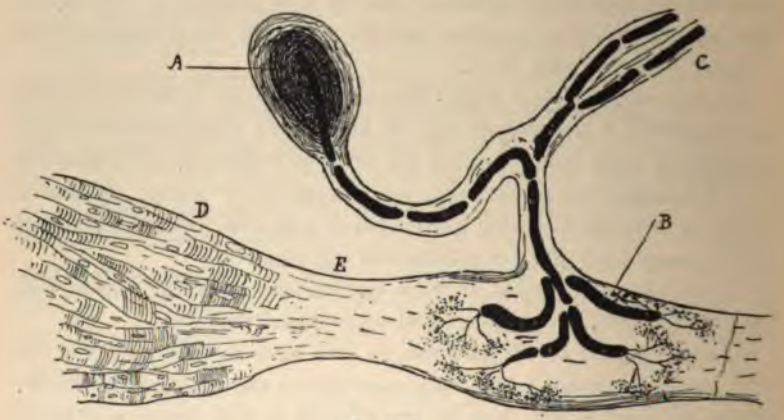


Fig. 108.

Tendinous corpuscle, after Cattaneo (semi-diagrammatic).

B. Corpuscle of Golgi with terminal ramifications of the nerve which divides within it. Its right extremity is continuous with the tendinous fibres; its left extremity E with the muscular fibres D, which come to be inserted upon it. A is a Pacinian corpuscle, which receives one of the branches of bifurcation of the nervous filament C, of which the other branch passes into the corpuscle of Golgi.

becomes gradually confused with the tendinous fibres, whilst the other somewhat thicker and longer, gives insertion at different levels to a few muscular fibres. As regards structure the corpuscles of Golgi are formed of fibrillar connective tissue containing scattered nuclei.

These fibrils are parallel to the axis of the corpuscle; they

* Golgi, *Intorno alle distribuzione e terminazione dei nervi nei tendini del uomo e d'altri vertebrali* (*Gaz. méd. italo-lombarda*, 1878) et *Mémoire de l'Acad. des sc. de Turin*, série II, t. XXXII.

† Cattaneo, *Sugli organi nervosi terminali muscolo-tendinei in condizioni normali, e sul loro modo di comportarsi in sequito*, &c. *Accad. Reale delle Scienze di Torino*, 9 Janvier, 1887.

anhistous casing coated with endothelium, somewhat to that described by Ranvier upon the sheath of this again takes part in the formation of the casing, and nies the nervous fasciculus which is connected with the le. In the interior of the corpuscle, the nerve fibres have penetrated either into its middle portion, or more into one of its extremities, divide either dichotomously, or ore ramifications which spread over the whole corpuscle; rils which go to its surface having lost their sheath of , and forming a fine reticulum. The nervous filaments thus ramify in the corpuscles are in close connection

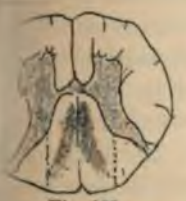


Fig. 109.

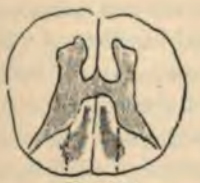


Fig. 110.

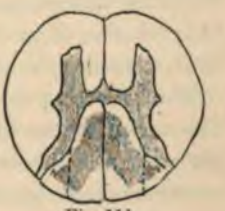


Fig. 111.

109.—Section of the spinal cord (at the union of the dorsal and lumbar gions) from a case of tabes in which the patellar tendon reflexes could still e obtained. It will be observed that in this case the lesions of the posterior olumn do not extend beyond the pointed line which passes from the bend of he posterior horn to the surface of the cord.

g. 110.—Section of the spinal cord (at the union of the dorsal and lumbar regions) from a case of tabes in which the patellar tendon reflexes could be obtained until a few days before death. In this case the lesions barely extend beyond the pointed line.

Fig. 111.—Section of the spinal cord (at the union of the dorsal and lumbar regions) from a case of tabes in which the patellar tendon reflexes had entirely disappeared for some years. In this case the lesions extend considerably beyond the pointed line and penetrate into the territory of the external bandlets (after Westphal).

either with the *muscular spindles of Kühne* (*fuseaux musculaires de Kühne*), or with the *Pacinian corpuscles* which are also found in the human tendons.*

If I have somewhat dwelt upon this delicate anatomical point it is because of my belief that you did not all perfectly know these details. At the same time I have by no means the pretension to affirm that amongst the different nerve endings in the thickness of the tendons it is the function of one form rather

* With regard to the question of the different nerve endings in muscle, see also an interesting work by M. Pilliet, in the *Journal de l'Anat. et de l*

than another to transmit the impulse which originates the tendon reflex. I only wished to show that such transmission was possible and that the only question to be decided is through which organ it is effected.

What path does the impulse now follow in order to reach the nerve centre? When once the nerve fibres of the tendon, which are afferent, are reached, it passes with them into the spinal cord. In the special case which we are considering (patellar tendon reflex) these fibres are in the sensory portion of the *crural nerve*.

In the spinal cord we can again, on account of the works of Westphal, indicate the exact point in the white substance through which the fibres pass before joining the grey matter of the posterior horn; it is in the zone of entrance of the roots (Wurzeleintrittszone, the external bandlets of Charcot and Pierret).

The following is the exact seat as indicated by Westphal:— The seat of these fibres is external to a line which is parallel to the posterior median fissure, and passes from the bend of the posterior horn to the posterior surface of the cord; they are therefore placed between this line and the posterior horn. Every lesion seated in this part* would produce some change in them, and may, in consequence, according to its size, produce diminution or absolute loss of the patellar tendon reflex.

It must not however be thought that the fibres which constitute the afferent path of this reflex exist throughout the whole length of the cord; they are only found in a certain part, and Westphal has also determined their seat both in the longitudinal and transverse direction. According to him these fibres enter the cord exclusively *at the union of the dorsal and lumbar regions of the cord.*

You will therefore understand, gentlemen, that the loss of this reflex will not be caused by every lesion of the posterior columns of the cord, but only in those in which the seat of the lesion is

* The importance of this localisation by Westphal must not be exaggerated. It amounts, in fact, to a statement that the afferent fibres of the patellar tendon-reflex enter the cord through the posterior roots of the middle or upper part of the lumbar region, and that they are short fibres. We know, in fact, that most fibres of the posterior roots enter the cord by means of the tract termed "the external bandlets." The short fibres divide here while the long fibres and those of medium length pass on in order to join the column of Burdach and that of Goll. A lesion in the external bandlets alone being able to cause the cessation of this reflex, it is very probable that the afferent fibres by which the impulse passes do not leave these bandlets, and are in consequence short fibres.

in the region of the external bandlets and quite at the upper part of the lumbar region. This localisation is in the case of tabes of considerable importance. Let us return to the study of the patellar tendon reflex considered in a general way. We have traced the path of the primary impulse whose origin was irritation of the patellar tendon as far as the grey substance of the posterior horns. When the grey matter has been reached what is then its path?

In all probability the nerve fibres by which the impulse passes transmit it to one or several cells of the posterior horns, and on account of the means of communication existing between these and the cells of the anterior horns it passes into the latter, or very possibly into the posterior root fibres which place themselves directly in contact with the cells of the anterior horns. The effect of the impulse is to cause the anterior horns to act according to their function, that is to say, by producing contraction of those muscular fibres over the action of which they preside. The primary afferent impulse is thus transformed into an efferent motor act; the motor impulse passes on its part by the following path: the anterior roots, sacral plexus, motor fibres of the *crural nerve* supplied to the quadriceps femoris whose contraction produces the movement of the leg which you perceive.

This, gentlemen, is a somewhat long and complicated path, and you will easily understand that if at any point some disorder occurs connected with the working of the organs through which the impulse passes, a diminution or even cessation of the reflex may occur. You will also understand that causes which affect either the circulation in the cord, or the muscular tone, may have considerable effect upon it.

Let us first examine (*a*) the LOCAL CONDITIONS which may affect the production of the patellar tendon reflex when seated in one or other of the segments of its path.

As regards the *patellar tendon* itself, adhesions or displacement of the ligamentum patellæ or condyles may occur after arthritis, dislocation or some other cause which prevent the necessary impulse being produced by percussion, and hence the reflex cannot occur.

In the *afferent peripheral nerve* any disorder whatever of the nerve trunk or its branches (peripheral neutris) may produce diminution or cessation of the reflex.

Disorders seated in the posterior roots through which the impulse passes may have the same effect.

In the *posterior column of the cord.* The lesions seated in this part have already been considered, and, as you know, changes in the *external bandlets* at the union of the dorsal and lumbar regions of the cord may produce cessation of the reflex.

In the *grey matter of the spinal cord.* When either the nerve cells which take part in the production of the reflex, or

Fig. 112.—Diagram of the path taken by the patellar tendon reflex. A. Patellar tendon struck by the percussion hammer, the impulse passes towards the cord by following the direction of the arrow, penetrates by the posterior roots into the posterior column and thence into the posterior horn, it then passes through the grey matter to the cells of the anterior horns, which it causes to act. From these cells an efferent motor impulse is transmitted to the quadriceps femoris C. The letter B indicates the patella.

the nerve fibres which connect them, are destroyed, the reflex is diminished or ceases; this occurs in the course of acute or chronic poliomyelitis, and specially in *infantile spinal paralysis.*

In the efferent motor nerve. Any lesion of this nerve, whether traumatic or due to peripheral neuritis from any cause will prevent the motor impulse from being transmitted to the muscle and consequently produce cessation of the reflex.

In the *muscle.* Atrophic muscular, or other changes which diminish their contractility produce first diminution and then cessation of the patellar tendon reflex. This specially happens in *idiopathic muscular atrophy.*

Under the name of GENERAL CONDITIONS (*b*), which may influence the production of the patellar tendon reflex, I shall place all those which do not immediately pass along some part of the path by which the reflex passes.

Of these conditions some tend to produce DIMINUTION, others CESSATION of the patellar tendon reflex.

Age has a striking effect, and thus P. J. Möbius observed that in 56 persons free from disease of the nervous system, who were more than 80 years old, this reflex was lost in 9 cases, and very slight in 7 others. On the other hand, Pelizœus, who tried to obtain the reflex in 2,400 children, never failed to do so. It is usually more pronounced in children than in adults.

Fatigue has a similar effect. Orchanski having showed that when muscular exertion is commenced the knee reflex increases whilst on the occurrence of fatigue it diminishes, and even ceases when the fatigue is such as to cause trembling of the limb.

Sleep has also the effect of diminishing the reflex.

Anæmia of the spinal cord (?) from any cause would have the same effect.

Different *infectious diseases* (diphtheria).

Some forms of *acute intoxication*, notably that by *chloroform*, may totally prevent the appearance of the reflex, this effect, though complete, being transient.

Certain forms of transverse *lesion of the spinal cord* seated at some distance from the lumbar region of the cord (in the dorsal or cervical region) are associated with paraplegia, in which the muscles are flaccid, and there is cessation of the patellar tendon reflex. Ch. Bastian,* then Bowlby,† recently called attention to these facts, and Babinski ‡ has just published an observation of the same kind. According to Bastian, this cessation of the reflexes would only occur in cases in which both motion and sensation (notably with respect to pain) are absolutely lost, and the transverse lesion is consequently complete.

The general conditions which may cause the patellar tendon reflex to be EXCESSIVE are of different kinds.

Certain forms of *strong emotion*, such as that produced by loud music, would have this effect, according to Lombard, who has investigated the condition of the reflex in himself in 239 different conditions.

* Bastian, *Royal Medical and Surgical Society*, 25th February and 17th May 1890. *Brit. Med. Journ.*, 1890, I, p. 480, 1132.

† Bowlby, *Ibidem*.

‡ Babinski, Paraplégie flasque par compression de la moelle. *Arch. de méd. expér.*, 1891, p. 228.

Certain *acute affections* (tuberculosis, enteric fever, rheumatic fever, pneumonia). Some forms of *acute intoxication*, specially that due to strychnine, have this effect. Intoxication by atropine produces the same result.

In rare cases of *polyneuritis* such excess is also observed (Strümpell and Möbius).

Muscular exertion notably increases the knee jerk. E. Jendrássik, who discovered this fact, has been able to utilize it clinically in a very ingenious way. Whenever the patellar tendon reflex cannot be produced in the ordinary way this author recommends the employment of the following method, which has been very rightly termed *that of Jendrássik*. Whilst the tendon is being struck the patient is directed to place the four fingers of each hand in a "claw-like" position, and to lock the hands together in such a way that the back of one hand and the palm of the other regard his chest, the fore-arms being in a horizontal position. The patient is then asked to make an effort, as if he wished to separate the two hands, care being taken, however, that he is prevented from doing so by the last phalanges which are tightly hooked together. The effort which is thus made by the two upper limbs probably determines a special condition of tone in the spinal cord; in any case the effect of this act is that the patellar tendon reflex, which was to all appearance lost or much diminished, clearly occurs again, or in the case of a healthy patient is manifestly increased.

Lesions in the pyramidal tract, whether in some part of the brain, medulla oblongata, or spinal cord above the lower part of the dorsal region, cause a considerable increase of the different tendon reflexes. It is to this class that the excess found to exist in a large number of cases of transverse myelitis, compression of the spinal cord, and amyotrophic lateral sclerosis belongs. In these cases there is a very different cause. In all probability the inhibiting effect which is normally exercised by the fibres of the pyramidal tract upon the centres in the grey matter of the cord ceases to exist, and these centres being less restrained in action become more irritable, this condition giving rise to increase of the patellar tendon reflex.

Such are the principal *facts* upon which the pathology of this reflex is based. I must, however, say a few words in connection with *theories* which exist or have existed upon this subject, and

at the same time shall avail myself of this opportunity to mention the history of this interesting question.

In the first place allow me to remind you that from very early times children have played the following practical joke upon their companions: when they are standing in an upright position their companions amuse themselves by sharply striking the tendons by the side of the hollow of the knee with the side of the hand, the immediate result of which is that they fall suddenly and unexpectedly to the ground; or when their friends are seated they lightly strike the knee of some one near them, whose legs are crossed, in the same sudden way, being amused by the abrupt movement which is thus produced in the foot. These are but the games of children, of which physiologists, being seriously inclined, were careful to know nothing, absolutely ignoring, in fact, all phenomena of this kind. It was necessary for the medical profession to take the matter in hand, this not being the first time that such a thing has occurred, and it is to be hoped in the interest of physiology that it will not be the last.

In 1875, Erb, in *Archiv für Psychiâtrie*, devoted some pages to the description of certain reflex movements, which he terms *tendon reflexes*, special attention being paid to that which takes place in connection with the knee, to which he gives the name of patellar tendon reflex (patellarsehnenreflexe).

By a singular coincidence in the same number of this treatise, and immediately after the article of Erb, was one by Westphal upon the *knee phenomenon*, which is nothing in reality but the patellar tendon reflex. The explanation of its occurrence however is quite different in the eyes of Westphal. In his opinion the contraction of the quadriceps is due to the irritation produced in this muscle by irritation of the tendon which is struck. As it is in this tendon that all the fibres of the muscle end, it is not surprising that its irritation should specially affect the muscle. That the reflex action however may occur, the muscle must be in a certain state of *tone*, upon which Westphal much insists, although he does not clearly explain its nature. At the same time, it is the loss of this tone which causes abolition of the patellar tendon reflex to occur.

The comparison of these two memoirs is on the whole to the advantage of the former. It is certain that Erb was the first to describe the phenomenon, and it is equally true that he was

far-sighted enough to indicate its true nature, viz., that it was a tendon-reflex. Notwithstanding this, however, the absence of patellar tendon-reflex in patients suffering from tabes has been called, and has kept the name of the *sign of Westphal*. It was certainly Westphal who first insisted upon the absence of this reflex in those suffering from tabes, adding a new symptom of extreme importance to the clinical history of the disease, so that from a practical if not from a theoretical point of view he has earned the gratitude of the medical profession.

In France, the study of the patellar-tendon-reflex was introduced into the Salpêtrière Hospital by Charcot, a short time after the publication of the works which I have just named. Joffroy was also interested in this symptom, and in 1879, Brissaud, in his remarkable thesis for the doctorate, devoted a certain number of pages to it which I cannot too strongly recommend you to read.

The opinion that this reflex had a tendinous origin had rapidly become more and more generally adopted, and Tschirjew, who at the instigation of Westphal had studied it from a physiological point of view, specially maintained it, although as I have just said, it was contrary to that of the professor of Berlin.

In England a certain number of authors, and by no means the least distinguished, amongst whom I would quote Gowers and Waller, are of opinion that this is an example of *idio-muscular contraction*. In support of this theory they specially allude to the fact that the interval for the patellar tendon reflex (that is to say, the time which passes between percussion of the tendon and contraction of the quadriceps) is only from 30 to 35 thousandths ($\frac{1}{33}$ to $\frac{1}{29}$) of a second (Tsirjew), it cannot be admitted that a reflex act can possibly be effected in so short a time, since according to physiologists the rapidity of the nervous current is only 30 metres (1 metre = 39·37 in.) per second. More recently these authors have also recognized that the spinal cord has some influence, and state that in order that the patellar tendon reflex may be produced, the integrity of the reflex arc is absolutely necessary. Jackson and Bastian believe that the action of the *cerebellum* is indispensable in order that the tone may be maintained, which enables the tendon-reflexes to be produced.

It may be replied to these objections that on the one hand the

rapidity with which the nervous impulse passes is not exactly known, while on the other hand the interval for the patellar tendon reflex is longer than 30 to 35 thousandths ($\frac{1}{33}$ to $\frac{1}{29}$) of a second, measuring in reality from 48 to 52 thousandths ($\frac{1}{21}$ to $\frac{1}{19}$) of a second (Brissaud). Tschirjew, in his observations of subjects suffering from spastic paralysis, was able to show that the interval was in their case shorter than in healthy persons. A certain number of other arguments will also be mentioned in support of the theory that the knee and other tendinous phenomena are reflex in nature:—

a. The loss of the patellar tendon reflex when the spinal cord becomes anæmic (as from compression of the aorta).

b. The loss of the same reflex in diseases during which the direct irritability of the muscles is scarcely modified at all (tabes, general paralysis of the insane).

c. Its production by the summation of irritations (Jarisch and Schiff)* upon condition that these very small shocks take place at the rapidity of at least one in each second.

d. Its production by percussion of the periosteum (periosteal reflex) in certain regions, notably that of the wrist. In these parts, in fact, it cannot be said that traction of the muscle by the tendon causes its irritation; and idiopathic contraction, since the blow is not made upon a tendon but upon a firm body, incapable of being moved by traction, namely, the bone itself.

We have passed in review, gentlemen, the principal theories which have been suggested in connection with the patellar tendon reflex, and almost exactly the same ideas have been formed as regards the other tendon reflexes (that of the Achilles tendon, of the triceps in the arm, of the flexors of the foot, of the wrist, of the masseter, &c.). These notions will now be applied to the study of the patellar tendon reflex in the course of tabes.

Usually *there is loss of this reflex in tabes* from the very beginning of the affection.

To this rule, however, there are some exceptions.

In some patients suffering from tabes the reflex *persists* although the signs of tabes are already quite characteristic. In these cases the lesions most often occupy almost exclusively the upper part of the spinal cord (upper form of tabes). You must know, however, gentlemen, that in these cases its preservation

* Jarisch et Schiff, *Untersuchungen über das Kniephænomen*, 1882.

is far from being constant. We have already seen that, from an anatomical point of view, it is due to the fact that the external bandlets (bandlettes externes) of the posterior column at the upper part of the lumbar region of the cord are unaffected.

In other patients suffering from tabes the reflexes still exist, but are clearly of *diminished* intensity. Not infrequently they are of *unequal* strength. Goldflam* rightly considers this inequality in the patellar tendon reflexes as an early sign of Duchenne's disease. At the same time, as he remarks elsewhere, this fact is not absolutely characteristic of tabes since Lombroso found the same inequality to exist in 14 per cent. of the criminals whom he examined in connection with this point.

Lastly, in some patients suffering from tabes the patellar reflex is apparently lost, but by the process of Jendrassik may again be produced. This artificial restoration of the reflex is not special to tabes, but may occur in other affections, a fact to which I shall return when considering the diagnosis of the disease. It has been already observed that in some patients suffering from tabes in whom hemiplegia occurred the patellar reflex has been found to return after this had happened. Thus, gentlemen, the patellar reflex may present different changes in tabes, and it is the duty of the clinical observer to understand how to observe and interpret them.

In order to conclude the study of *reflex action* in tabes the condition of the other reflexes should be now considered: the *cremaster reflex, the bulbocavernocus reflex*, &c. I prefer, however, deferring their description until the different systems are considered with the functions of which they are respectively connected.

I need only state in a few words that the *plantar reflex* is of most irregular occurrence in this disease, and the same may be said as regards the greater number of cutaneous reflexes. The plantar reflex is at times preserved during a long period, but may be completely lost. In the latter case there is often more or less pronounced anæsthesia of the sole of the foot. Such being the case, it is useless to observe that the presence or absence of this reflex is of little value in connection with the diagnosis of tabes.

* Goldflam, *Ueber die Ungleichheit*, &c. *Neurologisches Centralblatt*, 1888, Nos. 19 et 20.

LECTURE XVII.

TABES.

SYMPTOMS (*continued*).

D. ORGANS CONNECTED WITH THE SENSES.

1. *Visual organs.* I. *Those which are external to the globe of the eye.* Paralysis of the external muscles of the eyeball, ptosis, shedding of tears, epiphora, exophthalmos, narrowness of the palpebral opening, diminution of ocular tone. II. *Internal organs of the eye.* Condition of the pupils, inequality, myosis, mydriasis, deformity. Reflex actions of the iris: on exposure to light, on accommodation, sign of Argyll-Robertson, reflex due to pain. Optic nerve; optic neuritis; its onset, symptoms, and characters.
2. *Auditory organs.* Diminution in the power of hearing; deafness, its characters; Ménière's disease: excessive irritability of the auditory nerve by electric currents.
3. *Olfactory organs.* Anosmia.
4. *Gustatory organs.* Loss of taste.

Of the functional disorders which affect the ORGANS CONNECTED WITH THE SENSES in tabes, those which occur in the *visual organs* are amongst the most frequent and important. All, or almost all parts of the eye and its appendages may be affected in tabes.

I. ORGANS EXTERNAL TO THE GLOBE OF THE EYE.

Paralysis of the external muscles of the eyeball. This form of paralysis is very frequent; some statistics (Moeli, Berger) indicate its occurrence in 39 per cent. of the cases of tabes; Gowers states that four-fifths of the patients under his care who suffered from tabes presented this symptom at some time in the disease, specially before the period of inco-ordination. The figures given by Uhthoff* differ considerably from these, since he states that in 100 cases of tabes he met with paralysis of the ocular muscles in but 20 patients; it is most probable that he only alluded to patients in whom this paralysis was in actual existence, since if the cases are included in which this

* Uhthoff, *Arch. f. Psych.*, t. XXI, fasc. 2.

form of paralysis has occurred at some period of the disease the number would, in my opinion, be much higher.

As regards the greater frequency of paralysis in one or other of the muscles a difference of opinion exists. According to Berger and Woinow the external rectus is most often involved; according to de Watteville and Landolt it is also the *external rectus*. Then come the *superior rectus*, *inferior rectus*, *inferior oblique*, and lastly the *superior oblique*. In some cases the paralysis,

Fig. 113.—A man suffering from tabes with double ptosis. This photograph was taken while the patient was in the attitude which he assumed when not wishing to look at any object. (Collection of Charcot.)

instead of being confined to one muscle, affects them progressively one after the other, until the eye cannot make the slightest movement; *progressive external ophthalmoplegia* is then said to exist.

Of the nerves of the eye it is the third nerve which is most often affected. There will usually be no difficulty, gentlemen, in recognizing the existence of these forms of paralysis on account of the deviation of the ocular axes which they produce. In the cases in which no deviation exists when the eye is in the

ordinary position, it must be sought by causing the patient to follow the finger with his eye whilst it is moved in different directions. In addition to this the inquiry must be made whether *diplopia* has existed, and one of the first questions which must be asked is the following: do you see two objects instead of one? have you done so at any time?

Such a form of paralysis may be either unilateral or bilateral. Fournier, who has given an excellent description of this symptom,

Fig. 114.—A man suffering from tabes with double ptosis. This photograph was taken when the patient was in the attitude which he assumed when looking at an object; he was then obliged to throw his head back until the object corresponded with the narrow opening which existed between the two eyelids.

rightly insists upon some of its characters, namely, that it is *dissociated, partial, perhaps very limited in degree, transient, ephemeral*, and at times of *almost instantaneous duration*, that is to say, the paralysis usually ceases spontaneously after such a short time as a few weeks or days, or even a still shorter time; it is however *liable to recur*.

Of these forms of paralysis some, in the opinion of oculists, are due to a peripheral cause, others to a central cause of

nuclear origin. These facts, however, have not been clearly proved.

Ptosis is a symptom which is frequently observed at the onset of tabes. In most cases it is unilateral, but may exist on both

Fig. 115.—Asymmetry of the axes of the eyes on account of ocular paralysis in tabes. (Damaschino collection.)

sides; it is usually associated with deviation of the eye outwards, this being the result of paralysis of the third nerve; sometimes it is isolated and unassociated with any paralysis of the ocular muscles. Sometimes it is incomplete, but at times is so pronounced that even when bending the head backwards the patient is unable to see with the affected eye; he is obliged to raise the eyelid directly with the finger, or if his work requires the use of the two hands by means of a bandage or eye-speculum.

Sometimes *watering of the eyes* or *epiphora* are associated with ocular paralysis. In certain cases these symptoms are not due to paralysis, but to a disordered condition of the secretion, of which we shall again speak.

Lastly, other symptoms may be observed to which attention has only been recently drawn, and which are consequently but little known.

Exophthalmos, of which we shall speak at greater length when [illegible] the cardio-vascular complications of tabes.

A slight narrowing of the palpebral opening (Jacobson and Berger), due to paralysis of the smooth fibres of the sympathetic nerve supplying the eyelid, and analogous to that which is found to occur after some lesions of the cervico-brachial nerve roots. This symptom is, in fact, somewhat frequent.

Diminution of the ocular tone (Berger) may exist, on account of a diminution of the intra-ocular tension in consequence of paralysis of the sympathetic nerve.

II. Internal Organs of the Eye.

The *iris* is liable to many disorders which are of great importance in the diagnosis of tabes.

The *size* of the pupils is very often either relatively or absolutely altered. Sometimes there is only a slight *inequality*

Fig. 116.—Diagram representing the size of the pupils in tabes. The black figure in the middle represents the pupil of a healthy man moderately illuminated. The black figure on the left side represents the pupil when mydriasis exists (it may be of even larger size). The black figure on the right side represents the pupil when myosis exists.

between the two pupils, without its being possible to say that one is too large or the other too small; sometimes the pupil is much reduced in size, the condition to which the term *myosis* is applied. The pupil may in these cases be no larger in diameter than that of the head of a small pin, notwithstanding which fact the patients make no complaint whatever of weakness of sight.

In *mydriasis* the reverse is the case. It consists of dilatation of the pupil, which may be carried to an extreme degree in tabes. It is rarely equally pronounced in the two eyes; mydriasis may in fact be present in one eye, while myosis exists in the other. The latter condition is more frequent than the former, and it is by means of its existence that the disease hitherto unrecognised may sometimes be known to exist.

The form of the pupil may again be changed, and it may become of a somewhat *obliquely oval* shape, that is to say, instead of being round, it becomes more or less oval (Berger) with the

large diameter from above downwards, or from without inwards, the two sides remaining symmetrical.

We have not, however, yet ended the disorders affecting the iris in tabes, and its different reflex actions must still be considered. You know, in fact, that reflex contraction of the iris may be produced by three forms of irritation.

(*a*) *Light reflex.* When a light is brought near the eye, or when, after having covered the eye by the hand, one quickly removes it opposite a window in full daylight, the pupil is seen to contract at once in a decided way, and to a considerable extent. In tabes this reflex contraction produced by light is absent in a large number of cases, viz., from 80 to 90 per cent. whether myosis or mydriasis exist. This disorder usually occurs almost simultaneously in the two eyes. Sometimes the loss of the light reflex is not absolute; the first effect of the light is to produce contraction of the pupil, but this is almost immediately followed by dilatation which persists.

(*b*) *Accommodation reflex.* This reflex occurs normally when a *near* object is looked at, specially if that object is placed very close to the eye, that is to say, when the eye must necessarily accommodate in order to see the object.

This reflex also consists in contraction of the pupil. It often remains until an advanced period of the disease. This persistence of the accommodation reflex associated with loss of the light reflex constitutes an excellent sign of tabes at the onset; although this fact had been already mentioned by Vincent and Coingt, many authors continue to call it by the name of the sign of *Argyll-Robertson.* In connection with this point I must give you one piece of advice which may be of practical use; when the condition of the light reflex is investigated, be careful to ask the patient to look at some distant object and not at the light which is close to his eyes; otherwise, on account of the eye being fixed on a near object the pupil is seen to contract, and you are liable to mistake for light reflex the change in the pupil which is really due to accommodation.

The accommodation reflex is, as I have just observed, usually retained during the first stages of tabes, but after a certain time it usually also ceases to exist.

(*c*) *Pain reflex.* This reflex consists in the fact that when irritation is produced by pain in a healthy person (by means of

a pinch, &c.), and the pupil is examined at the same time it is seen to dilate momentarily. As Erb has shown, this reflex is very often lost in tabes, ceasing to occur in certain cases at an earlier period of the disease than the light or accommodation reflex.

The *optic nerve* is often affected in tabes, and its lesions are specially formidable inasmuch as they may induce complete loss of sight. Statistics differ somewhat as to the frequency with which *optic neuritis* occurs; at the same time the number cannot be said to vary more than between 10 and 20 per cent. of the cases. The eye seems to be the part specially involved in some patients with tabes, since optic neuritis is more frequent in those who have previously suffered from paralysis of the ocular muscles.

Optic neuritis is rare in cases which are already of some years duration, and is most usually found during the preataxic period. At the same time Gowers states that he has never observed it as the first evidence of tabes: in all the cases seen by him it had been preceded by loss of the tendon reflexes or lightning pains. Optic neuritis is almost always, if not always, *bilateral* in tabes; at the same time its onset may not be absolutely simultaneous in the two eyes, and in that case the left eye is usually first affected.

The length of time which passes between the onset of this affection of the optic nerve and the occurrence of *loss of sight* is variable; the mean time was found by M. Berger to be 3 years, but it varies from 2 months to 17 years. In some cases the affection of the optic nerve remains in a stationary condition; loss of sight is not, therefore, constant, and at times the condition actually improves.

Lastly, some authors, notably Gowers, have remarked that a kind of antagonism exists between optic neuritis and inco-ordination. It is certain that in the cases in which optic neuritis occurs at an early period of the disease amaurosis rapidly follows, but there is little or no inco-ordination; on the contrary in the cases in which optic neuritis only exists, after inco-ordination there is usually but slight amblyopia, and very rarely amaurosis. It has even been said that when optic neuritis occurs the inco-ordination diminishes. The latter assertion seems to me somewhat ungrounded. For my part I am quite ready to admit the first part of the assertion, namely, that the cases of tabes in

which the optic nerve is involved, constitute, to a certain extent, a special form of the disease, the *upper form of tabes*, or *cerebral tabes*; in this variety the evolution of the affection occurs favourably as far as any organs are concerned, except those connected with vision. But as regards any direct action of optic neuritis upon the course of the disease this seems to me very improbable.

The symptoms which indicate the existence of optic neuritis are the following:—

a. Diminution in the acuity of vision. In this case the patients often complain that there is, as it were, a veil or mist in front of the eyes diminishing their acuity, and an examination by means of different typographical scales enables the extent of this diminution, which may be considerable, to be estimated.

b. Dyschromatopsia. This condition specially exists with regard to certain colours. Thus yellow and blue are usually seen by the patient during a certain time after he has become unable to see the other colours.

c. Contraction of the visual field. Authors are by no means agreed as to the form which this takes. Leber speaks of a concentric contraction; Galezowski of peripheral contraction; von Graefe of nasal contraction; Schweigger of temporal contraction. It would not be my place, gentlemen, to give an opinion on this point, but in the presence of such different statements I must confess that I feel strongly inclined to agree with Berger that no form of contraction of the visual field is characteristic of tabes. According to the latter author temporal contraction is most liable to occur. Charcot found that in the majority of cases an irregular form of concentric contraction existed.

d. A *scotoma* may be present over half or a section of the field of vision in certain cases, and may even be seated in such a position that hemianopia, due to a central cause, seems at first sight to exist. Ophthalmologists are of opinion that this form of scotoma, in the same way as amblyopia, without apparent lesion of the pupil depends upon neuritis, due to some cause connected with the part of the brain seated above the medulla oblongata.

As regards the *ophthalmoscopic* appearances of these lesions I will briefly quote their principal characters as indicated by

Berger, who has specially studied the ocular disorders which occur in the course of tabes.

Quite at the onset the papilla is at times of a uniform red colour. Most frequently the first change observed is that the

Fig. 117.—Extreme concentric contraction of the field of vision in a case of tabes. (Damaschino collection.)

Fig. 118.—Contraction of the field of vision in the left eye, in a case of tabes with atrophy of the optic nerve. (Damaschino collection.)

Fig. 119.—Scotoma simulating contraction of the visual field with the hemiopic form in a case of tabes. (Damaschino collection.)

nasal portion of the optic papilla becomes of a somewhat grey colour, so that instead of being, as in its normal condition, more red than the temporal portion it becomes of almost the same hue. This grey colouration may have a bluish or pearl-grey tint. Afterwards, when the lesions are very advanced, it becomes white, like mother-of-pearl, but even at this period its edges are

still well defined. The vessels are considerably altered; quite at the onset the arteries and veins are normal; the arteries then become contracted and the veins swollen; lastly, considerable atrophy of both arteries and veins occurs towards the end of the disease.

Auditory Organs.

The AUDITORY ORGANS are almost as frequently involved as the visual, and the troubles which they present are numerous;* they often occur early in the preataxic period of the disease.

The *diminution in the acuity of hearing* is in most cases slowly progressive, being at first but slight, increasing slowly, and possibly becoming most pronounced; but the *deafness* may occur rapidly, as in a few days, or even suddenly. The diminution in the acuity of hearing is almost always bilateral, although it may be more pronounced upon one side than upon the other. One important character of the deafness is that it is absolute, and presents neither the irregularity nor the varieties which are so apt to occur when it is due to disease of the middle ear or its appendages. In tabes in fact the disorder is not one connected with the transmission of sound, the receptive organ, viz., the *auditory nerve* being involved; the *auditory neuritis of tabes*, analogous to the optic neuritis of the same disease, may exist. This explains the fact that when the external portion of the ear is examined it is found to be free from disease. At the same time, since the middle ear may be affected in tabes, it is better to ascertain how far the deafness depends upon the transmitting, and how far upon the receptive portion of the ear. Recourse should therefore be had to the method of Rinne,† and to that of

* M. Morpurgo (*Arch. f. Ohrenheilkunde*, 1890), who examined the patients of Marina, and some others who were suffering from tabes, with regard to associated disorders of the auditory organs admits that 81.13 per cent. of these patients suffer from some aural disorder; according to him the receptive portion of the ear is most often affected, changes in the transmitting portion being less frequent, and of but secondary importance; affections of both parts may however coexist, and both kinds may depend upon trophic disturbance.

† The method of Rinne consists in placing a vibrating tuning fork in contact with the mastoid process, and holding it there until it ceases to be heard; it is then placed opposite the external auditory meatus, and if it is again heard, the "positive Rinne" is said to exist, as when the ear is in a normal condition; if it is no longer heard, the "negative Rinne" exists, this indicating the probable existence of an obstacle in the middle ear; if therefore more or less pronounced diminution in the acuity of hearing is associated with the "positive Rinne," and no lesion can be detected by means of the otoscope, it is probable that some lesion exists in the internal ear.

Weber,* which may give interesting if not absolutely reliable information.

Subjective sounds are frequently heard in tabes, being of varied character: at times very violent and painful buzzing sounds are heard, or it may be the sound of hissing, or different musical noises.

Aural vertigo (*Ménière's disease*) is by no means rare, if one includes under this name all the cases of tabes in which vertiginous sensations exist, but do not occur in true paroxysms as in *Ménière's disease* properly so called. This fact was first mentioned by Pierret,† and by Charcot; I myself studied this subject from a special point of view in conjunction with Walton in 1883.‡ Since that time many works have been written on this subject, one of the last and most important being that of A. Marina.§

The cases of aural vertigo which occur in tabes may in my opinion be classed in two groups.

A. In some cases there is considerable diminution in the acuity of hearing, and different lesions may exist in the transmitting portion of the ear. In these the same happens as in cases of ordinary aural vertigo when tabes does not exist. This disease has in fact but little if any effect upon it, though perhaps it may produce increased irritability in the nerves supplied to the labyrinth.

B. In other cases there are neither diminution in the acuity of hearing, nor lesions in the transmitting portion of the ear. It is in connection with these cases that Walton and I proposed the hypothesis that a lesion probably existed in a special portion of the fibres of the auditory nerve, which is termed by some anatomists and physiologists the *nerve of space*, a lesion in all appearance directly due to the existence of tabes.

The excessive irritability of the auditory nerve to electric currents

* The *method of Weber* consists in placing a vibrating tuning fork in contact with the vertex, which is usually more distinctly heard upon the side which is the seat of the diseased ear, when the middle or external ear are affected.

† Pierret, Contribution à l'étude des phénomènes céphaliques du *tabes dorsualis*. Symptômes sous la dependance du nerf auditif. *Rev. mensuelle de méd. et de chir.*, 1877, p. 101.

‡ P. Marie et Walton, Des troubles vertigineux dans le tabes (Vertige de Ménière tabétique). *Revue de Médecine*, 1883, p. 42.

§ Al. Marina, *Zur Symptomatologie der Tabes dorsualis mit*, etc., *Archiv f. Psych.* t. XXI, p. 156.

in tabes has been specially studied by Marina. That author has shown that this condition is by no means rare in tabes, since he found that in 8 cases out of 11 electrical reactions were produced in the auditory nerve by means of currents whose intensity was less than 15 milliampères, an intensity at which they only commence to occur in healthy persons. This excessive irritability to the electric current may be looked upon possibly, to some degree, as explaining the relative frequency of vertiginous sensations in some patients suffering from tabes in whom the ear is but very slightly affected.

Olfactory Organs.

Olfactory organs.—The patients are at times troubled by more or less disagreeable odours of purely subjective origin. In rare cases *true* anosmia exists which is probably due to a lesion of the olfactory nerves which is special to tabes.

Gustatory Organs.

Gustatory organs.—Some patients experience strange sensations of taste, notably a more or less persistent taste of sugar, for the existence of which there is no reason. Sometimes, again, there is complete *loss of taste*, this being also probably due to some change in the nerves of taste.

In conclusion, it may be said that these sensory disorders are rare, slightly pronounced, and but little known.

LECTURE XVIII.

TABES.

SYMPTOMS (*continued*).

E. TROPHIC DISORDERS.—*a. Disorders of the general nutrition. b. Spontaneous fractures:* nature of these fractures: absence of pain, tendency to consolidation, tendency to form a large quantity of callus: special ætiological conditions: the slightest injury may produce them, seat of these fractures, fractures of the apophyses, fractures of the vertebræ, their nature. Lesions of the bones: porosity, thinning of the compact substance, dilatation of the medullary cavity, dilatation of the Haversian canals, decalcification of the lamellæ of bone, changes in the osteophytes, embryonic transformation of the marrow in the bones, diminution of the non-organic, increase of the organic material. Nature of these alterations.

GENTLEMEN,—The study of TROPHIC DISORDERS in the course of tabes will occupy us during several lectures. They are so varied as to constitute a certain number of entirely distinct groups, as distinct, in fact, as a fracture of the thigh is from a cutaneous affection. We will therefore consider them apart from each other and describe each one separately.

a. Disorders of general nutrition. These disorders have as yet been very little studied. With the exception of some works upon the quantity of excrementitious matter contained in the urine of patients suffering from tabes, works, again, which have contained no very conclusive evidence, scarcely anything has been written upon this subject. And yet, gentlemen, one fact connected with it is of great importance. This the good sense of the medical profession has quite understood: *tabes = consumption*, ataxy is a consumptive disease; this is what first struck the observers. They were right, since those attacked by tabes are in the large majority of cases persons suffering from consumption, from "medullary phthisis," as it was termed in old times. It is this condition which causes them to have so characteristic an appearance: they are thin, their muscles are flaccid, their skin has an earthy tint or one of bistre, the eyes are somewhat sunken and surrounded by a dark circle, the features are pinched, in short the appearance is entirely that of

persons suffering from a high degree of malnutrition. It will be observed, however, gentlemen, that this appearance, though very usual is not absolutely constant. Some patients will be found to have the appearance of being in excellent health; they are stout, and their aspect is most satisfactory. This, however, is the exception, and the latter patients are those in whom the medullary process is apparently exhausted and remains in a stationary condition. However this may be, I determined, gentlemen, to direct your attention to these disorders of general nutrition in the course of tabes, since they form part of the clinical aspect of this disease. The question may be asked whether at some future time, on account of the incessant progress which it makes, pathological anatomy will not explain this phenomenon. It will, perhaps, demonstrate that this consumptive tendency of tabes is due to the degeneration of certain extra or intra-medullary nerve fibres analogous to the *visceral fibres* described by Gaskell in the posterior columns of the spinal cord.

b. Spontaneous fractures. These fractures occur somewhat frequently in patients suffering from tabes, much more so, in fact, than one would be at first inclined to think. Mentioned for the first time by Weir Mitchell, their existence was not generally recognised until 1873, after they had been studied by Charcot. The works of Forestier, Richet and Raymond contributed to increase our knowledge upon this subject, whilst an excellent critical survey by Talamon generalised it. We shall in these lectures review the works of Regnard and Blanchard; speaking also of the opinion expressed by Volkmann and of the effect which it had upon the development of the question of tabid fractures in Germany; I shall also have more than once to quote the conclusions formed in some of the investigations recently made, to which I shall call your attention as the lectures progress.

These fractures present in most cases, without considering their ætiological conditions, certain objective characters which are sufficiently pronounced to distinguish them, and to justify their being separately described.

One of the principal amongst these characters is the *entire absence of pain due to the fracture;* this absence of pain is so complete [illegible]ny of the patients do not even perceive that

the fracture has occurred, and still continue to move the limb as if nothing had happened; you know, gentlemen, how painful fractures are in healthy individuals, so much so in fact that the least shake, the slightest movement, are quite intolerable.

In some cases the patients state that during a certain time before the fracture pain was felt in the place where this subsequently occurred; such would therefore be a premonitory pain. In other cases the fracture occurs during a paroxysm of lightning pains; in all these cases, however, the pain is never due to the fracture itself. This, as I have already said, remains painless in most cases, in some, however, pain exceptionally occurs, but it rarely becomes as pronounced as in healthy individuals. The absence of pain is in all probability due to the analgesia which exists in the deep parts, and specially in the periosteum.

When the fracture has taken place the swelling of the part is considerable, often greater than in an ordinary case, and of longer duration.

The course of these fractures also presents certain special characters; the *tendency to consolidation* is somewhat rapid, but it must not be thought, as some authors have said, that in the spontaneous fractures occurring in tabes it takes place more rapidly than in persons in good health. The consolidation may in fact take place at quite a late date, and even in some cases a false articulation (pseudarthrose) is formed.

As regards the *tendency* to produce *much* callus, this is not constant, only occurring in the fractures in which the immobility has not been complete. In such cases, in fact, on the one hand the adaptation of the fragments is very incomplete, and the size of the bone consequently increases at this part; on the other hand the following incident occurs upon which Kredel rightly insists: on account of the absence of pain the patient allows the extremities of the bone to be rubbed one against the other without being troubled by their being so; a process which is employed by surgeons for the purpose of accelerating union when the callus is slowly produced. The effect of such reciprocal friction is that the process of bone formation is much hastened, and hence the fragments become of larger size as well as the callus which joins them together.

It has been also stated that the spontaneous fractures which occur in tabes are specially liable to be associated with shortening

of the limb. This fact is generally true, but may easily be explained without any special property existing in the callus. These fractures not being attended by pain are liable to be unperceived both by the patient and his medical attendant, and even when recognized to exist may be left more or less untreated; the patient, suffering no pain, does not trouble himself to keep the limb quiet, and the fragments are liable to be displaced even after having been put in a good position. Owing to the fragments passing one over the other, the callus forms abnormally

Fig. 120.—Fracture of the lower extremity of the humerus in tabes, displacement of the fragments, large quantity of callus. (Collection of Charcot.)

Fig. 121.—Fracture of the two bones of the forearm in a case of tabes, large quantity of callus. (Collection of Charcot.)

(in excess), and the limb is shortened to a more or less considerable extent. There is also another cause of the shortening observed in fractured limbs, not only in ordinary cases, but when the arthropathy which occurs in tabes is associated with fracture of the head of the femur or humerus; on account of the tendency to absorption of the bony extremities which exists in arthropathy so that the head of the bone may completely disappear. The result of this would be a considerable reduction in the length of the leg or arm.

Before commencing to study the lesions to which these symptoms are due, we must briefly consider their *ætiological* conditions; you will again see that in this case one important fact confirms what is said by those who maintain, as Charcot was the first to do, that the spontaneous fractures which occur in tabes constitute a separate group. This fact is the *disproportion* which exists between the *slightness of the injury* and the fracture of the bone, which is so pronounced that these were consequently termed "*spontaneous fractures.*"

The history of tabes contains numerous observations in which one or several fractures have been produced by mere traction or

Fig. 122.– A woman suffering from tabes with fracture of the left femur (the figure having been turned round it is represented as the right femur): after this fracture so much absorption took place at the extremity of the femur that the length of the thigh was considerably shortened. (Collection of Charcot.)

Fig. 123.—A, A normal femur. B, femur in a case of arthropathy in tabes. C, femur in a case of fracture in tabes. Such absorption of fragments occurred that the bone scarcely attained half the length of that of a normal femur. The bone belonged to the patient represented in fig. 122. (Collection of Charcot.)

a trifling shock. Upon some occasions the patient was merely crossing his legs, or drawing off his boots, or perhaps lying in

bed with the head resting on the elbow when the fracture occurred. A fracture of the inferior maxillary bone has even been known to occur while the patient was breaking a piece of sugar between the teeth. Lastly, in some cases fractures have occurred without the patient being able to ascribe them to any definite act, being probably due to some irregular movement. It is in fact incontestable that the movements of patients suffering from tabes are often so abrupt and violent that fractures may easily result from them. To look upon this occurrence, however, as some authors do, as the principal and even exclusive cause of fracture in the course of tabes is, in my opinion, to make a mistake. The true cause of these accidents is the fragility of the bones, produced by morbid conditions of the bone, which will be considered by us presently.

The *period of the disease* in which these fractures occur varies. Most often they take place during the stage of inco-ordination, but frequently also before that time; this being another proof that inco-ordination is not the only cause of these accidents.

As regards *sex* one fact deserves to be mentioned, namely, that these fractures are more frequent in women. To such a point is this the case that of 32 cases of spontaneous fracture occurring in tabes, collected from medical literature by Max Flatow, 15 were found to occur in the female sex. When the infrequency of tabes in women is considered this is a large number, and the great predominance of fractures in that sex must be recognized, unless one admits that the value of these statistics is to a certain extent impaired by the first observations made at the Salpêtrière Hospital, into which for a long time only women were admitted. Even, however, when this correction is duly considered, one must, gentlemen, in my opinion, allow the fact that these fractures are far more common in the female sex.

So far fractures have only been considered in a general way, let us now study them in connection with the different bones, considering in the first place in what part they are most often seated.

In connection with this question I will quote the statistics given by Max Flatow :—

Fractures of the femur	19 cases : taking place specially in the diaphysis, fracture of the neck occurring in 4 cases.
Fractures of the leg below the knee	17 cases.
Fractures of the forearm	7 cases.

It will be observed, gentlemen, that fractures of the lower limb are incomparably more frequent than those of the upper. In my opinion there are two reasons for this: 1. Tabes being an affection which specially involves the lower half of the spinal cord, it is not surprising that the lower limbs should be more affected in every respect than the upper. 2. It is in the same bones of the lower limb, specially the femur, that fractures are most liable to occur in those whose bones have been changed in their anatomical or chemical composition, as for example from the effect of age. I must also observe, and this is the objection which occurs most readily to the mind, that these are the bones which are most exposed to injury, though, when everything is considered, the latter fact does not seem sufficient to explain the predominance with which the spontaneous fractures of tabes occur in the lower limb.

The different statistics do not seem to show that the fractures specially occur on either side of the body. It is possible, however, that they are more likely to take place upon the side in which the symptoms of tabes are most pronounced whenever some asymmetry in the symptoms can be recognized to exist.

It not infrequently happens that *multiple fractures* exist in the same patient, either from many bones being simultaneously fractured, which in some cases has happened in as many as 5, or even 6, or from more than one fracture occurring in the same bone (as many as 3 in one case). This also shows, in my opinion, that the fractures are due to a special condition of the bone.

In connection with the long bones a special variety of fractures should also be mentioned, which differs from those which have been hitherto considered; it is that which ocurs when a *bony prominence* or *apophysis* is torn off by the muscles inserted upon it. On account of the absence of immobility and the friction which is facilitated by the analgesia of which I have spoken, an excessive amount of callus is formed, whence, according to Volkmann (quoted by Kredel), the bony prominences are produced which penetrate into the muscles and tendons. I am ready to admit that this may occasionally happen, but in some cases I have seen the intra-muscular osseous prolongations attain such a length that I cannot help attributing them to *ossifying myositis*. However this may be, gentlemen, I call your attention to these

facts from feeling sure that they would be more frequently observed was their existence better known.

Another class of fractures which is also very interesting is that which includes *fractures of the vertebræ*. These were first mentioned by Charcot, and the diagram which I now put before you, and which comes from his collection, is the representation of one of the first, if not the first case in which a fracture of the vertebræ was observed. Pitres* published a case in 1885; since then some other cases have been mentioned, notably by Krönig,† who on his part studied three such cases. These fractures usually present a collection of characteristic symptoms; there is more or less considerable *deformity* of the spine, a deformity which could only form progressively, and in a somewhat slow manner. At the seat of the fracture *angular curvature* is found to exist posteriorly, owing to projection of one or more vertebral spines; anteriorly a hard body is occasionally felt in the abdomen by *palpation*, owing to displacement of one of the vertebræ. On account of these changes in the form of the spine, the *attitude* of the patients is considerably altered, the upper part of the body being curved or rather *bent* anteriorly or laterally, while on the other hand, owing to diminution in the length of the spine the upper part of the trunk is as it were forced into the abdomen and pelvis, giving rise to the formation of strange horizontal folds in the skin.

I should be afraid, gentlemen, of giving you a wrong idea with regard to these fractures of the spine, if I omitted to mention that they rarely exist alone; they are most often complicated by more or less pronounced *arthropathy* affecting the inter-vertebral articulations; to such an extent is this the case that the lesions may often be classed indifferently as joint affections, or fractures. Besides these fractures involving the *bodies* of the *vertebræ*, they may also occur in the *arches* or *processes*.

After the description which has been given of these different varieties, you will easily understand that if the *diagnosis* of spontaneous fractures in tabes is often easy to make, it is at times very much the reverse. Many difficulties may present

* Pitres, *Société de Biologie*, 21st November, 1885.

† Krönig, *Wirbelerkrankungen bei Tabikern. Zeitschrift für klinische Medicin*, 1888, XIV, p. 51.

themselves to the observer. At times the analgesia is of such a character, as I have already stated, that the patient is not aware of its existence, and if the fracture interferes but little with his movements, as when he is bedridden, it may be completely unperceived. Or again, the œdema, which in the fractures of those suffering from tabes is usually more pronounced than in

Fig. 124.—Fracture of the spine. On account of the sinking of the vertebræ which has occurred there is considerable lateral curvature of the spine. (Collection of Charcot.)

ordinary fractures, may cause so much swelling as to make any examination of the part impossible.

The diagnosis will lastly be found very difficult or even impossible when the fracture involves an epiphysis of which the articular portion is affected by arthropathy; in that case, again, the swelling of the adjoining parts and mobility of the bones prevent a proper examination from being made. You will not therefore be astonished, gentlemen, that at the autopsy of those who have suffered from tabes, fractures and bony consolidation are often observed to exist, of which there was not the least suspicion during life; it is especially in connection with the flat bones which are situated somewhat deeply (sternum, pelvis) that these surprises are wont to occur.

Such observations may be made about the clinical varieties of these fractures; the lesions which accompany them will now be considered, and these will explain some of the characters which cause the fractures occurring in tabes to have a special character.

The *changes in the bones* which are visible to the naked eye are the following:—

The *open cancellous structure* at the surface causes the bone to have a "worm-eaten" appearance in certain parts. At times this condition is so pronounced that marked ulceration exists (Féré); when this condition is very evident the bone can occasionally be indented by the pressure of the finger. If a longitudinal section of a bone which is thus altered be made by means of a saw it will be found that—

The *compact substance is more thin*, being possibly so much reduced as not to have more than half or a third part of its normal thickness; this also causes the bone to give way when it is pressed upon, the compact tissue being the part which is the most capable of resisting pressure.

The *dilatation of the medullary canal* which exists, so that the hollow part at the centre of the bone is enlarged, also diminishes the solidity of the bone.

The microscopical examination shows other causes of the friability which exists in the bones; and it is to R. Blanchard that we owe the precise and detailed knowledge which we have of these lesions from a HISTOLOGICAL point of view. These consist in—

Dilatation of the Haversian canals, specially of those which are seated most near to the medullary canal. When this *dilatation* is at the surface of the bone it is to this which produces the "worm-eaten" or "lacunar" aspect already mentioned; when very pronounced near the medullary canal it contributes to produce its enlargement.

Absorption of the osseous material near the Haversian canals is shown to occur by the fact that the diseased bone is coloured more deeply by picro-carmine than that which is healthy. This absorption seems one of the first symptoms which occur in the bones in tabes, since it is detected near the Haversian canals, which are not as yet dilated; this would therefore be one of the primary lesions.

Changes in the osteoplasts. These suffer more or less from atrophy in certain points, owing to granular and fatty degeneration, presenting also a tendency to lose their angles and to present rounded outlines.

Embryonic transformation of the medulla in bones (Richet). This exists in great abundance, filling all the cavities produced by dilatation of the Haversian canals. These changes would indicate, according to Richet, somewhat pronounced osteitis.

Lastly, to these histological alterations an enumeration of CHEMICAL CHANGES found to exist in the bones in tabes by P. Reynard should be added.

Diminution of inorganic materials. Instead of the normal proportion of 66 per cent. of the total weight, the inorganic

Fig. 125.—Longitudinal section of the upper extremity of a long bone (humerus) affected by the disease which exists in the arthropathy of tabes, showing the reduced size of the compact tissue and cartilage, where the arthropathy (A) exists. (Damaschino collection.)

portion of the bone only forms 24 per cent., a large deficiency in phosphates being specially found to exist (10 per cent. instead of 50 per cent.).

Increase of organic materials. These normally form 33 per cent. of the total weight; in tabes they form 76 per cent.; the proportion of fatty matter is specially great (37 per cent.).

None of these chemical or histological changes, however, is exclusively characteristic of tabes, they show at most the existence of *rarefying osteitis*, an affection which may exist in many chronic affections of the bones.

The only lesion special to tabes is that which Pitres and Vaillard found to exist in a case of spontaneous fracture of the tibia. The nerve filament which enters the nutritive orifice of that bone, according to these authors, presents obvious changes. Siemerling afterwards observed the same lesion in an analogous

case. In consequence of these facts it is easy to understand why it is that the changes in the bone are ascribed by some of the medical profession to peripheral neuritis. It cannot be denied that the coincidence is most suggestive, but it scarcely suffices to show that, in opposition to the opinion of Charcot, the changes in the nutrition of the bone are not dependent upon medullary alterations.

However this may be, the fact upon which I would lay special stress, gentlemen, is that the changes which I have just described are found not only in the fractured bone, but in all or nearly all the bones of the skeleton, so that in certain patients suffering from tabes *all, or nearly all, the bones are "liable to fracture,"* and multiple fractures may therefore occur in the same patient. It should be added that lesions of the bone do not exist in every case of tabes, nor can it be indicated, even to an approximative degree, in what proportion of cases these lesions occur.

It will be explained, gentlemen, in the next lecture, that the lesions in the bone are precisely those which are found at the commencement of the arthropathy which occurs in tabes, so that fractures and arthropathy are, in reality, but different expressions of the same pathological condition, and cannot be separated from each other, at any rate from an anatomical point of view.

LECTURE XIX.

TABES.

SYMPTOMS (*continued*).

F. ARTHROPATHY IN TABES.—*History:* "Charcot's joint disease"—opposite opinion of Volkmann. *Onset* more or less sudden: swelling, doughy condition, absence of true œdema; crepitation; absence of pain. *Course* of the affection mild and severe form. *State of the articulation* when the arthropathy has existed for a certain time; flattening of the part, abnormal relation of the articular surfaces, abnormal mobility, absence of pain. *Complications:* fracture, pain, passage of the articular extremities through the skin, suppuration. *Ætiology:* occurrence at different periods, greater frequency in the female sex; seat in different joints. *Pathological anatomy:* articular capsule, intra-articular ligaments, synovial membrane, effusion, floating bodies in the joints, extremities of the bones; atrophic type, hypertrophic type, cases of combination of these two types. *Pathology of tabid arthropathy:* different theories. A. The primary cause of the arthropathy is a nervous lesion (spinal cord, medulla oblongata, peripheral nerves). B. The primary cause is not a nervous lesion (traumatic, syphilitic, rheumatic arthritis, arthritis deformans). *Conclusions.*

GENTLEMEN,—*Tabid arthropathy* is called by another name in England, that of "Charcot's joint disease." The fact is that Charcot not only discovered the existence of that affection, but described it from the very first in so complete a manner that this name is in every way appropriate. His first works upon this subject were written in 1868. In the following year Ball already mentioned 18 cases. In the same year (1869) Clifford Albutt published an account of the first case described in England, the second being recorded by Buzzard at a somewhat later date (1873). In Germany the idea of tabid arthropathy developed but slowly. Professor v. Volkmann having stated that the joint affections in tabes were purely and simply due to injury or twisting of the joints in the irregular movements of the disease, his authority was sufficient to prevent any other explanation from being admitted. Since that time a great change has occurred, and I shall more than once have to quote from important works written by German authors. The following numbers, taken from a memoir by Weizsäcker,

published in 1887,* represent more or less what has been written about this disease:—of 109 cases to which this writing alludes, 53 are French, 36 English, and 18 German.

I shall put before you several patients suffering from this affection, and we shall be able to observe in them some of the characters which it presents. They are all cases in which arthropathy has already existed for a certain time. Since I had not the opportunity of showing you a patient in whom the symptoms were just commencing to occur, I must describe to you in full the first stage of the affection.

The *onset* of tabid arthropathy varies but little. In some cases it is absolutely *sudden*. A patient while walking perceives crepitation in the hip; his legs give way under him, and he falls. Almost immediately, or after some hours, considerable swelling occurs in the region of the joint, which soon extends over the whole thigh. In other cases the onset is not quite so sudden, though it is always *abrupt*. Thus, while the patient is undressing, he by chance perceives that one of his joints is swollen without, however, having suffered any pain; or upon rising in the morning he makes the same observation without having had any abnormal sensation during the night. It may, again, happen that during the few days which precede the onset of the affection premonitory crepitation occurs in the joint which will be involved.

However this may be, one of the principal initial symptoms is the *swelling* of the joint, and soon afterwards of the whole limb. This swelling presents certain characteristic features. As I have just said, it occurs more or less suddenly, and acquires its maximum degree in a few hours or days. It almost always affects the whole segment of a limb or even the entire extremity which becomes of truly enormous size, having the same appearance as exists in elephantiasis. It is certain that this is not due to ordinary œdema, since the swelling is firm, unyielding to the finger, does not pit on pressure, or leave an indentation after being pressed with the finger. The skin of the part is pale, often shining, the veins are dilated, while redness, heat, pain, and in fact all the signs of inflammation are absent. In order to explain this singular appearance Debove

* Weizsäcker, *Die Arthropathie bei Tabes. Beiträge zur klinischen Chirurgie von Bruns*, 1887.

has made the very probable suggestion that there is not only an effusion into the articular cavity but also rupture of the capsule, which allows the liquid to penetrate throughout the entire limb.

The subsequent history of the swelling is as follows:—After some days, or rather weeks, or sometimes many months, it diminishes in size and becomes confined to the joint and adjoining structures. It may remain in this part for some time, after which it completely disappears, although the strangest forms of dislocation may be subsequently found to exist in its place, of which I shall speak presently.

Crepitation often, but not always, occurs in the diseased joints; usually occurring on the onset, and subsequently ceasing to exist at the time when effusion into the joint occurs.

The *movements* of the joint, strange to say, are little if at all affected, specially at the onset; the patient merely complains that the limb seems to be heavy and that he is soon fatigued.

But of all these symptoms, that which combined with the swelling is most characteristic of tabid arthropathy, is the *absence of pain*, however pronounced the articular lesions may be, a fact which is quite in opposition to that which occurs in most joint affections. We have also seen, gentlemen, that the same occurs in the spontaneous fractures of tabes, of which it was the special character to be unaccompanied by pain.

As to the *course* of tabid arthropathy, as Charcot pointed out, two forms may be distinguished:

In the *mild form* the effusion disappears in a short time, and the joint returns to its normal condition, slight crepitation being the only trace of the affection. At the same time, however mild the affection may be, it must be known that *relapses* are liable to occur, relapses which may be far more serious than the first attack; so that a mild form may be subsequently followed by a serious attack.

In the serious form the œdema of the limb does not disappear as rapidly as in the preceding form; it remains over the joint and adjoining parts; at the same time the lesions in the bones and ligaments occur, and soon become so much developed as to interfere more or less completely with all the functions of the limb.

It is from this form that the patients who are assembled here suffer, though it has not the same form in the different cases.

In this woman, for example, the arthropathy has only existed for a few months in the left hip joint; the œdema, as you will observe, still occupies the whole thigh, although as the patient tells us, it has much diminished; the movements are somewhat difficult, but still possible, and there is little if any crepitation.

In the second woman one of the knees is affected; in this

Fig. 126.—Recent tabid arthropathy in the left hip joint. (Damaschino collection.) The enormous increase in the size of the left thigh will be observed. The lesion having existed but a short time, the upright position can be maintained without pronounced deviation of the lower limb.

case there is but little swelling, but on account of the lesions in the bone, some deviation is already beginning to occur in the direction of the limb, and upon my attempting to produce lateral movements in the joint, I can do so, as you see, without difficulty, whereas when the joint is in a healthy condition the lateral movements are barely perceptible; hence there is apparently much difficulty in walking.

Lastly, in the third patient, the woman who is lying upon

the litter, the affection has, so to speak, reached its most serious point. It has lasted certainly for many years, but the difference of its appearance from that of the other cases is great; in this patient there is no swelling, but, on the contrary, flattening in the region of the hip joint so that the condition of the articulation can be easily examined. Its abnormal mobility is first

Fig. 127.—Tabid arthropathy in the left knee. (Damaschino collection.) The knee and thigh are swollen, the knee presents prominences which cause it to appear of a square form. The lateral movements were very pronounced, and could be produced by slight pressure of the hand upon the foot.

observed; the most impossible movements can be given to the thigh, in comparison with which the well-known straddling position of the most famous acrobats would cut but a poor figure; at the same time, however extensive these movements

Fig. 129.—Recent tabid arthropathy in the shoulder (from a patient named Berthel). (Collection of M. Charcot.) Much enlargement of the shoulder. The condition of the shoulder joint a few years later is seen in the following figure.

Fig. 130.—Ancient tabid arthropathy in the shoulder (from a patient named Berthel). (Collection of M. Charcot.) The head of the humerus, which is no longer in relation with the acromion, forms a prominence beneath the skin in front of the angle of the inferior maxilla.

may be, no resistance is felt in the joint. The reason of this is that the joint can be distended to an extreme degree, the articular surfaces in which a great change has occurred being no longer in close relation with each other. You yourselves can again easily understand what occurs, even at a distance, since the subcutaneous prominence which you perceive in the hip is really the upper extremity of the femur, which changes its place whenever the thigh is moved, and to an extent which corresponds with the amplitude of the movement. Owing to its

Fig. 128.—Tabid arthropathy in the knee. (Damaschino collection.) The most extraordinary dislocations can be produced in the lower limb of this patient without the slightest pain being produced.

being possible in this way to bring the upper extremity of the femur, so to speak, underneath the skin, it is easy to feel its different parts, and to recognize that both the head itself and a large part of the neck have almost completely disappeared, and that this extremity is almost solely represented by the great trochanter. It is unnecessary to add, gentlemen, that with such lesions the patient is quite unable to walk; but the fact which I am anxious to repeat and impress upon you is, that however much I move or drag the limb no pain whatever is felt by the patient; if she does not walk it is solely because the mechanical conditions of the joint do not enable it to support the weight of the body.

Fig. 129.—Recent tabid arthropathy in the shoulder (from a patient named Berthel). (Collection of M. Charcot.) Much enlargement of the shoulder. The condition of the shoulder joint a few years later is seen in the following figure.

Fig. 130.—Ancient tabid arthropathy in the shoulder (from a patient named Berthel). (Collection of M. Charcot.) The head of the humerus, which is no longer in relation with the acromion, forms a prominence beneath the skin in front of the angle of the inferior maxilla.

What may be called the pathognomonic characters of confirmed tabid arthropathy are thus, as you have just seen, *abnormal mobility*, in excess of any that could be imagined, and

Fig. 131.—Tabid arthropathy in both knees. (Collection of M. Charcot.) In this case the deformity which existed was lateral; though pronounced in degree it did not prevent the patient from walking a few steps with the help of two sticks.

Fig 132.—Tabid arthropathy in the two knees. (After M. Dreschfeld.) This patient presents in the most pronounced way the deformity which is known by the name of "genu recurvatum."

considerable change in the articular surfaces, neither of which conditions produces the slightest pain.

Such are the ordinary features of this affection; in some cases other symptoms occur which should perhaps be regarded as complications. Thus—

A *spontaneous fracture* may occur either in the diaphysis or more often at the extremity of a bone giving rise to the formation of a foreign body in the joint (specially the head of the femur).

Pain, which, as already observed, is usually almost entirely absent, may occur in some cases and persist during several days or weeks, this being specially the case when arthropathy affects the small joints (of the hand or foot).

The *articular extremities may pass* through the skin, as has been observed in some cases, and their mobility enables one to understand that this may be the case; in reality, however, it very rarely happens.

Suppuration, again, is a complication which is so rare that its occurrence need not be feared. At the same time, I might mention cases in which it has happened either after an imprudent puncture, or during the course of a general affection (pneumonia), or without obvious cause, but I would again say, gentlemen, that its occurrence is quite exceptional.

In order to finish what is connected with the clinical aspect of tabid arthropathy, gentlemen, its ÆTIOLOGY will be now discussed.

As regards the *direct causes* of its existence I shall have to speak of the part which *injury* plays in its production at a later period when the nature of the arthropathy is considered.

At the present time the conditions in which it occurs will be specially considered. The *time at which it takes place* is very variable. Kredel* found that in 132 cases it appeared—

21 times in the premonitory period of tabes.
38 times between the 1st and 3rd year of tabes.
32 times between the 5th and 10th year of tabes.
41 times after the 10th year of tabes.

It is therefore very difficult to specify the time of its occurrence, the truth being that it occurs at any period of the disease.

* Kredel, *Die Arthropathien und Spontanfracturen bei Tabes. Volkmann's Sammlung klinischer Vorträge*, 1888.

With respect to the age of the patients, no definite information can be given. At the same time, a case under the care of Charcot and Féré should be mentioned in which arthropathy occurred at the age of 20 years, tabes having commenced to exist in the 18th year. This is probably the first case published in which the onset of tabes occurred at so early an age.

As regards *sex*, in 100 cases of Weizsäcker 72 of the patients were males, and 39 females. On account of the infrequency of tabes in females one must conclude from this that arthropathy is more frequent in the female than in the male sex, even when the correction is made with respect to the female cases which were published at the Salpêtrière Hospital.

What is the *frequency* of arthropathic cases as compared with all the cases of tabes? With respect to this question I can only

Fig. 133.—Ancient tabid arthropathy in the knee (in a patient named Berthel). (Under the care of Charcot.) The lower extremity of the femur projects considerably above the upper extremity of the tibia and fibula.

quote the numbers given by Erb, who found 2 cases of arthropathy in 56 cases of tabes. These numbers represent, in my opinion, the true state of the case, and 4 or 5 cases of arthropathy may be supposed to exist in 100 cases of tabes.

In connection with the seat of the arthropathy several interesting facts are brought to light in the following statistics of Max Flatow, which include 149 cases, in 41 of which there was bilateral arthropathy. It was most frequent in the *knee*, which was affected in 60 patients, in 13 of whom it was bilateral. After the knee it occurred most often in the *foot*, which was affected in 39 patients, and in 8 of them bilaterally. The *hip* follows, having suffered in 38, and on both sides in 9 patients, and the *shoulder* in 27, and on both sides in 6 patients. The *elbow joint*, the *joints of the hand* and *fingers*, and the *temporo-*

maxillary joint were far more rarely affected, giving numbers which vary between 6 and 4. You will have observed that all

Fig. 134.—Tabid arthropathy in the thumb. The last phalanx of the thumb is turned abruptly outwards, so as to be almost at right angles with the first phalanx. The metacarpal bones connected with the fingers are somewhat swollen at their lower extremity.

Fig. 135.—Tabid arthropathy in the right elbow.

the joints may be affected, but that they are so with a varying degree of frequency.

The *pathological anatomy* of tabid arthropathy will now be considered.

The articular capsule, the lax condition of which was indicated by the clinical symptoms, is flaccid, softened, dilated, often ruptured, and at times more or less completely destroyed.

The *intra-articular ligaments*, such as the long portion of the biceps, the crucial ligaments of the knee, and specially the round ligament of the hip-joint, are the seat of somewhat accentuated lesions, and may completely disappear.

The *synovial membrane* is usually pale, its surface is often covered by vascular fringes, it is thickened, adherent to the adjoining parts; like the capsule it may entirely disappear. In

Fig. 136.—Tabid arthropathy in the shoulder. (Damaschino collection.) The capsule, which is extremely dilated, has been opened, and the head of the humerus is seen to be dislocated into the subscapular fossa.

some cases plates or nodules of bone are found in the thickened membrane.

The *effusion* into the joint cavity is usually serous, transparent, fluent, and of a clear yellow hue; it sometimes contains fibrinous flakes, in a few cases blood, and very rarely pus. Abundant at the onset, it disappears after a certain time. So much was said about these facts when the symptoms were discussed that it is quite unnecessary to dwell upon them now.

The existence of *loose bodies in the articulation* has been often mentioned, commonly of osseous structure, and either free, or possessing a pedicle. Usually newly formed within the joint they are sometimes due to a fracture of the epiphysis. In the former case these newly formed osseous bodies are frequently

many hundred in number, so that the articulation may give to the finger the sensation of a bag of walnuts.

As regards the *extremities of the bones* they present very different aspects, which may be referred to two large types :—

A. The *atrophic* type, the most frequent, the most characteristic as regards the pathological anatomy of the disease, in which there is more or less complete atrophy of the cartilage and articular

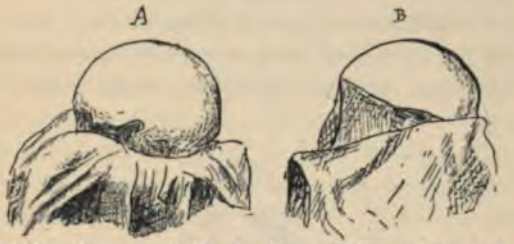


Fig. 137.—Tabid arthropathy in the shoulder. A. Normal head of the humerus. B. Head of the humerus upon the side in which arthropathy exists; the articular surface of the head of the humerus has much diminished in size upon this side. (Damaschino collection.)

Fig. 138.—Tabid arthropathy in the hip-joint. (Collection of Charcot.) The lower part A of the articulating portion of the head of the femur is completely absorbed and has disappeared, a plane surface alone remaining in its place.

Fig. 139.—Arthropathy in the shoulder. (Collection of Charcot.) The glenoid cavity of the scapula can scarcely now be seen, and the head of the humerus has almost completely disappeared.

surfaces of the bone, the wasting being possibly so considerable that a somewhat long portion of the epiphysis may entirely disappear. Thus, in the femur, for instance, not only the whole head, but even the neck of the bone may be completely absorbed. In this way, as in the patient whom I showed you just now, the great trochanter, which is itself much eroded, may be the only

part which remains; the femur has then, as has been said, an appearance resembling that of a "drum-stick."

Atrophy of a similar kind occurs in the *articular cavities*, their margins are effaced, their surface flat, their walls become thin and are at times depressed (the cotyloid cavity) by the epiphysis which rests upon them.

B. The *hypertrophic type*, also termed the type with the form of *arthritis deformans*. In this type there arise, specially at the edges of the cartilages and bone, either more or less numerous and irregular *outgrowths of bone*, or merely isolated *osteo-cartilaginous* projections, closely resembling those which are seen

Fig. 140.—Tabid arthropathy in the elbow, of the hypertrophic form. A considerable number of bony nodules are seen in the cavity of the joint, some in a free condition, others fixed in the periarticular tissues. The articular extremities are thickened.

in the joints affected by chronic arthritis deformans. At times the *velvet-like dissociation* of the cartilage is also observed, from this being worn away at the points of friction, the bone presenting in the same parts an *open cancellous* appearance or that of *eburnation*. The *synovial fringes* become *affected* by hypertrophy, and a frequent corollary to this event is the formation of *intra-articular osseous masses*, either free or pediculated. The *capsule* also presents changes; it is thickened, sometimes ossified in certain points, and may even become adherent to the bones and articular extremities.

Though these two types are quite different from each other,

however, they are undoubtedly both examples of true tabid arthropathy. To what then is the difference in the appearance due? Solely, in my opinion, to the character of the affected joint. Some joints respond to the morbid process which exists in tabid arthropathy of the type A, while others respond to that of the type B. Kredel, whose works I have already had occasion to mention in connection with the disease in which we are now interested, has remarked that certain joints, notably the hip and shoulder joints, tend specially to suffer from *atrophy* when they are the seat of some morbid process, specially that of *tubercular arthritis*, whereas others (above all the knee and elbow joints) are more disposed to suffer in a similar case from hypertrophy. This is precisely what happens in tabid arthropathy; when it is seated in the shoulder joint the *atrophic type* A is almost invariably observed, whereas when the knee is affected the *hypertrophic type B* is more frequently found to exist. And the proof that the anatomical difference is more dependent upon the character of the joint, than the special quality of the arthropathy, is that the two types may be combined in the same patient. If the hip and knee are simultaneously affected, the atrophic form will occur in the former, the hypertrophic in the latter joint.

Besides these two forms the variations produced by *combination of the two types* should be mentioned, in which by the side of atrophic lesions bony outgrowths or osteo-cartilaginous projections of more or less considerable size are found to exist. The appearance of the joint may again be changed in the cases in which an *intra-articular fracture* co-exists; the fragments of bone which are thus produced vary in dimensions from the size of a pea to that of a bean or more; they may either come from the epiphysis, or from the condyle, or may even be due to the whole condyle or epiphysis being torn away. The fragments may either remain free and constitute a foreign body in the joint, or be absorbed, or lastly, and specially in the hip joint, may become closely adherent to the articular surface of the other bone of the joint (the head of the femur to the acetabulum).

Such are the different anatomical conditions in tabid arthropathy; they are only found, it must be understood, in the joints in which this condition existed in a pronounced degree. At the same time it must not be supposed that the other joints are

absolutely free from disease. Jürgens has in fact shown that in many patients suffering from tabes all or almost all the joints, even when no arthropathy exists, present dilatation of the capsule and elongation of the ligaments; the vessels of the synovial membrane are also dilated, and the intra-articular ligaments of a somewhat red hue; in short most of the joints in tabes are frequently in a condition of "virtual arthropathy." The existence in the bones of these patients of arthropathy, and all the osseous lesions enumerated in the chapter devoted to the consideration of spontaneous fractures, should be compared with these facts (dilatation of the medullary canal and of the Haversian canals, decalcification, changes in the osteoplasts, transformation of the marrow in the bones, &c.).

Now that the principal characters of tabid arthropathy are known, its *nature* may be discussed, and I shall make use of this opportunity to say a few words about the different theories which exist, or have existed about this interesting question.

These theories may be ranged in two large classes:

A. *The primary cause of tabid arthropathy is a nervous lesion:*

Charcot, when he described arthropathy, regarded it as a trophic disorder occurring in the bones of the joints, and depending upon a lesion of *the spinal cord*, probably its anterior cornua, a lesion which in all probability had extended to the cornua from the posterior columns of the cord; Charcot and Joffroy, Pierret, Liouville, Seeligmuller have observed cases which testify in favour of such a lesion in the anterior cornua.

Other authors have assigned a different seat to the nervous lesion; Buzzard, stating that laryngeal and gastric crises occur frequently in patients with tabes who suffer from arthropathy, concludes from this fact that the trophic centre of the bones and joints is seated in the *medulla oblongata*, these lesions in the bones being associated with disorder in that part. This supposition however, as far as I know, has not been verified anatomically, and the coincidence which he mentions does not appear to exist, since Weizsäcker remarks that in the 109 cases of tabid arthropathy from which his statistics are formed, gastric crises only occurred in 30.

Lastly, certain authors, basing their ideas upon the researches of Pitres and Vaillard, Westphal, Siemerling, who in some cases of spontaneous fracture or arthropathy have found lesions in

the nerve seated in the nutritious canal of the bones, are led to believe that the symptoms connected with the joints are due to some change in the *peripheral nerves*.

B. *The primary cause is not a nervous lesion.*

Amongst those who have held this opinion the first who must be mentioned is Volkmann; according to this author the arthropathy is purely and simply a lesion of *traumatic origin*, and the reason of its occurring in those suffering from tabes is solely because injuries to the joints are far more frequent in these patients on account of the inco-ordination which exists in the movements. Either on account of falls, or because the joints are being constantly irritated by the irregular movements, they are more liable to suffer than those in healthy persons; to these unfavourable conditions the influence of analgesia must be also added (Jonathan Hutchinson).

Strümpell, on account of tabes occurring frequently as a late effect of syphilis, is of opinion that the arthropathy in tabes is really *syphilitic arthropathy*, and he consequently connects this affection of the joints with syphilis alone, and in no way with the disease of the nervous system.

Lastly, there is an opinion, which exists widely in England, that the arthropathy in tabes is merely *chronic rheumatic arthritis*. With regard to the ideas which exist in England about this affection I cannot help mentioning to you that expressed by one of the most eminent surgeons of the United Kingdom, namely, Sir James Paget.

According to this surgeon the arthropathy in tabes is not only a recently discovered, but a really new affection; it would only have existed so to speak during the last 25 or 30 years. In support of this opinion Sir J. Paget quotes the fact that in the rich collections of the Royal College of Surgeons, where examples of all the osseous or other lesions which may be met with have accumulated during so many years, no specimen whatever is found of tabid arthropathy. Although I do not form the same conclusion as Sir James Paget, this fact, at the same time, appears to me interesting, and we find in it special confirmation of the aphorism of Charcot, "one only sees that which one has learnt to see."

To return to the supposition that the arthropathy in tabes is of a purely rheumatic character I must also quote the opinion

of Virchow, according to which this affection is only a form of *arthritis deformans*. In fact, according to that learned pathologist, in both these affections the morbid process commences in the cartilage, which it causes to swell, and then to split and disappear. The only difference is that in tabes, from the effect of bad nutrition, the lesions occur more rapidly. There are many who hold this opinion, and the points of resemblance between tabid arthropathy and *arthritis deformans* are certainly at first sight numerous. But the differences are still more pronounced. Already in 1875 it was the object of Michel to explain this in his thesis. (*a*) In arthritis deformans the onset is slow, gradual, and usually accompanied by pains, which are at times most severe; (*b*) there is no effusion, or if one exists is in very small quantity; (*c*) the process is from the first one of hypertrophy, whereas in the arthropathy of tabes atrophy first exists; (*d*) the movements of the joints are difficult and limited, on the contrary, we have seen that in the arthropathy of tabes the mobility is much increased; (*e*) pronounced deformity may occur in both affections, but its character is quite different. Other points of distinction might be mentioned, specially in connection with their pathological anatomy, but in my opinion those to which I have alluded are sufficient to show that tabid arthropathy and arthritis deformans are totally distinct affections.

Such, gentlemen, are the principal theories which exist with respect to the nature of tabid arthropathy, they are, as you see, numerous, and differ much from each other. It is, in my opinion, useless to increase the difficulty in which this enumeration places us, and though I cannot oblige you to have the same opinion I must be allowed to add that it is still the first theory, that of Charcot, which seems to me the best. The arthropathy cannot be simply and purely due to the traumatic injuries occasioned by the inco-ordinate movements which occur, since, as we have already seen, it often occurs in the prodromic period when no inco-ordination of movement exists. Nor can it be simply the result of syphilis since unfortunately the number of persons affected by that complaint is legion, and yet unless tabes also exists they never present this singular variety of arthropathy.

With regard to the connection of tabid arthropathy with

peripheral neuritis as its initial lesion, I must confess, gentlemen, that I cannot believe this to exist, and for a reason analogous to the one which I have just given with respect to its purely syphilitic origin. We see, in fact, gentlemen, a large number of cases of so-called peripheral neuritis, and yet never, even in those who present the most characteristic symptoms of that affection, does any symptom occur which resembles tabid arthropathy. On the contrary an affection is known, due to a special

Fig. 141.—The innominate bone and femur in a case of tabid arthropathy of the hip joint. (Collection of Charcot.) The acetabulum has almost entirely disappeared, and its margin barely remains, the bottom of the cavity being continuous with the adjoining surface of the bone. The head and neck of the femur have also completely disappeared, and the great trochanter alone remains.

Fig. 142.—The innominate bone and femur in a case of chronic rheumatism (arthritis deformans). The margin of the acetabulum is much increased in size, and the cotyloid cavity much deeper than in its normal condition. The head of the femur is also increased in size.

lesion in the spinal cord, syringo-myelia, which is sometimes accompanied by arthropathy very similar to that which occurs in tabes.

For all these reasons I would refer the initial lesion of this affection of the joints in tabes to change in the spinal cord, and,

as I have already observed, certain autopsies seem to confirm the opinion of Charcot, who places this lesion in the grey substance. This being well established I find no difficulty in supposing that in some cases traumatic injury, either directly inflicted or due to the irregular movements, is an occasional cause of tabid arthropathy, and determines its seat.

LECTURE XX.

TABES.

SYMPTOMS (*continued*).

The foot in tabes: History.—Characters. Somewhat sudden onset, swelling of the foot, thickness of its inner margin, flattening of the arch of the foot, deviation of the metatarsus, shortening of the foot, enlargement of the malleoli, ankylosis of the joints of the foot, absence of pain.

Pathological anatomy: spongy appearance of the tarsal and metatarsal bones, their destruction.

Trophic disorders in the fibrous tissue: articular capsules; intra-articular ligaments; tendons, their rupture.

Trophic disorders of the skin: different eruptions—Perforating ulcer, its characters; Gangrene. Spontaneous ecchymoses; Loss of the nails and teeth, Hyperidrosis; anidrosis. Nature of these trophic disorders.

GENTLEMEN,—A separate place must be made by the side of tabid arthropathy and spontaneous fractures for an affection which, as I have already said with regard to the vertebral lesions, proceeds with almost the same frequency from either of these two morbid states—I mean the *tabid foot*.

The *tabid foot* was first described by Charcot and Féré. Immediately afterwards numerous observations made by Boyer, A. Chauffard, &c., showed the accuracy of this description; in England, one of the first cases published was that of Page; in Germany, that of Bernhardt. In all these cases the result of the examination of the parts was almost identically the same.

This condition usually exists at a comparatively *early* period, occurring most often at the end of the first or commencement of the second period of tabes, in other words in the præataxic stage. The *onset* is usually sudden, resembling that already described in connection with arthropathy; within twenty-four hours an abnormal swelling appears upon the back of the foot, after which the other morbid conditions gradually occur.

The principal characters of this affection are as follows:—

A somewhat considerable *swelling* forms, whose chief seat is the back of the foot, specially pronounced near the tarso-metatarsal

articulation, and causing this region to present a uniform curve, and not allowing any of the bones in the foot to form a prominence above its surface. It must be also observed, gentlemen, that pressure does not cause this swelling to disappear or produce any subsequent depression in it; this is precisely what has been already stated in connection with other forms of arthropathy.

The *inner border of the foot* becomes *thickened*, and at the same time as it were of rounded form, presenting occasionally a somewhat pronounced *prominence*, answering to the tuberosity of the scaphoid and internal cuneiform bone.

The *arch of the foot is flattened* and loses more or less com-

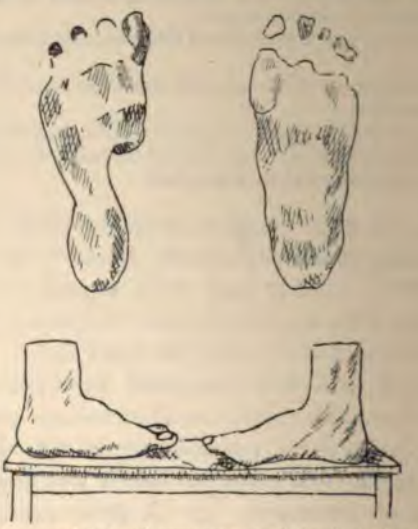


Fig. 143.—Impressions and lateral appearance of the feet of a patient suffering from a tabid foot on the right side. (On the left side the foot was sound; the right diseased.) (After Féré.)

pletely its curve, so that in some cases the sole of the foot is absolutely flat, and at times even slightly convex in form. This deformity is clearly seen in the impressions made by the foot. The arch of the foot is not always flattened (Troisier, Pavlidès), but on the contrary may be raised (Chinese foot; Damaschino, Pavlidès).

Displacement of the metatarsus outwards is sometimes observed, being less constant than the changes already mentioned.

Pronounced *shortening of the foot* exists in some cases, and this contributes to make the foot have a thickened, cubical, and in all respects singular appearance.

The *malleoli* are often enlarged and swollen, and may be the seat of co-existing arthropathy; the same may be said of the toes, which at times suffer also from arthropathy in those in whom the "tabid foot" exists.

The *mobility* of the different segments of the foot is diminished, and at times complete ankylosis occurs.

Crepitation, whether spontaneous or during passive movement of the joint, occurs but little if at all.

The *absence of pain* on pressure is as complete as in other forms of spontaneous fracture and arthropathy. Sometimes anæsthesia also exists when the back of the foot is pricked, while sensation exists in the lower part, the sole of the foot retaining its sensibility both as regards contact and temperature.

The result of the *pathological anatomy* of the condition is really singular, although the lesions found to exist are analogous to those with which we have already become acquainted in connection with other forms of tabid arthropathy. What are most surprising are the intensity of the lesions, and the amount of destruction which is produced by the morbid process in this form of arthropathy. In order to give an idea of this I now put before you the photograph of a specimen in the collection of Charcot, regretting that I cannot show you the specimen itself (since it is a most instructive one), on account of its having been recently stolen from the museum at the Salpêtrière Hospital. Charcot described the osseous lesions found in this case in the following way: "The articular facets upon the inferior surface of the astragalus, and os calcis are eroded and worn away, their margins being covered by granulations; the astragalus is fractured transversely at its neck; the scaphoid and cuboid bones are worn away, and can scarcely be recognized. Numerous separate fragments of bone exist, amongst which the external cuneiform bone can be with difficulty recognized. All the bones of the metatarsus and tarsus present a spongy appearance, and are unusually fragile and light." The fact is that in the tabid foot not only do wasting and atrophy of the different bones of the tarsus and metatarsus occur, but their absolute disintegration, to such an extent that at certain points a gruel-like mass is

alone found, containing small fragments of bone; one can therefore understand and agree with the following statement of

Fig. 144.—Bones of the foot in tabes showing the os calcis to be formed of extremely loose cancellous tissue, while the bones of the tarsus are almost completely destroyed. Fragments of bone which have passed beneath the foot are of loose cancellous tissue and jagged, coming from the bones of the tarsus, and found at the autopsy to be separated from each other. (Collection of Charcot.)

Fig. 145.—Tabid arthropathy in the foot. The os calcis forms, as it were, a thin osseous cup at its lower and anterior part, into which the astragalus has descended. (After Tuffier and Chipault.)

A. Chauffard, made in his excellent laconic expression, "In the tabid foot there is not more *osteopathy* than *arthropathy*."

What is finally the condition of the tabid foot? Much more simple than would have been expected when the intensity of the symptoms and the osseous lesions are considered. The swelling gradually subsides after a somewhat long time, but the deformities persist, and the almost absolute ankylosis which exists in the foot does not prevent the patient from using it, if not to a full, at any rate to a sufficient extent. Suppuration does not occur more in the tabid foot than in other forms of arthropathy, or in spontaneous fractures. It is unnecessary to say more about these points, which have been already sufficiently discussed, and I have spoken separately of the tabid foot in order that I might mention in detail the different objective characters which cause it to have such a peculiar appearance.

Trophic Disorders in the Fibrous Tissue.

Whilst considering these changes in the bones and articulations I should say a few words about the *trophic disorders in the fibrous tissue*.

In speaking of the different forms of arthropathy the lesions which occur in the *articular capsules* have already been enumerated; we have seen that they become relaxed, distended, and at times perforated, while atrophy may occur in certain parts they have invaded, in others newly formed bone exists. It is useless to dwell more upon this subject.

In a similar way we know that the *intra-articular ligaments* may be much altered, and even completely disappear.

What occurs in the *tendons?* In arthropathy more or less extensive lesions may occur in them in connection with the joints over the mobility of which they preside.

There is nothing surprising in this, which might have been foreseen. It is, however, another example of trophic disorder which should be mentioned, namely, the *rupture of the tendons* which occurs in tabes.

This symptom has not yet been observed in very many cases, but I have no doubt that it will be more frequently seen to occur when attention is drawn to this point.

I must quote the case of Hoffmann, in which rupture of the tendo Achillis occurring when the patient made the simple

movement required to enable him to turn round in the street.

In the case of Lépine there was rupture of the quadriceps extensor cruris. The same muscle was ruptured in the case of Löwenfeld whilst the patient was walking quietly.

Although the number of cases in which rupture of the tendons has been known to occur is small it is still possible to see that there is obviously some analogy between the rupture of the tendons and that of the bones (spontaneous fractures). The fact that they specially occur in the lower limbs, and are produced by causes which are quite insignificant when compared with the effect which they produce is to be noticed in both cases.

Trophic Disorders of the Skin.

These disorders have been noted by many observers, amongst whom the names of Charcot, Fournier, and Leloir should be specially mentioned. My intention is not to describe these in detail, but simply to enumerate them, since, owing to their infrequency, and but slight severity, they belong rather to the province of pure dermatology.

Many forms of eruption have been observed on the skin of those suffering from tabes, of which the name alone will indicate the character. Thus, lichenoid, herpetic, ecthymatous, erythematous, urticated, and pemphigoid, &c., eruptions may occur. Zona has been occasionally met with, being specially seated upon the trunk. Vitiligo (Leloir) may also be observed. Ballet and Dutil* noted hypertrophy of the epidermis, which was thickened and desquamating, the hypertrophy affecting not only the papillæ, but even the whole thickness of the skin. This is dry and can be raised in large folds, specially that of the upper limbs (the back of the hands). It is this condition to which authors have applied the term *ichthyosiform state of the skin.*

One of the most interesting trophic disorders affecting the skin, at any rate on account of its frequency, is the *perforating ulcer.*† It commences usually in a corn, which, after a variable

* Ballet and Dutil, Note sur un trouble trophique de la peau, &c. (*Progrès Médical*, 1883, p. 379.)

† Ball and Thibierge were the first to call attention to these facts in 1881. *On perforating ulcer of the foot as connected with progressive locomotor ataxy.* (*International med. Congress*, 7th session, London, 1881, vol. ii., p. 52—55.)

time ulcerates in its central part, and this ulceration may either remain superficial and heal, owing to the effect of rest, after a time which is of no great length, by the formation of fresh layers of epidermis; or on the contrary, extend more deeply to the subjacent articulations and bones, producing such lesions that the surgeon may be obliged to amputate the affected parts. This second form is certainly rare, being the form which I would term *surgical*, and which Chipault* had doubtless alone in view when, in his interesting *Review* upon the perforating ulcer, he stated that it was of rare occurrence. As regards the first form, on account of which the patients never require surgical treatment and of which the symptoms are often too mild even to attract attention, it is undoubtedly frequent, and may even be said to be very frequent. It is undoubtedly true that an ulcer which only consists of slight ulceration is not really a perforating ulcer; this, however, simply applies to the name, and an abortive form of so many affections exists that we should not be justified in refusing to look upon this as an example of such a kind. In some cases Tuffier and Chipault have shown that arthropathy of the joint immediately adjoining the perforating ulcer is already in existence when the onset of this complication occurs (specially in the metatarso-phalangeal joint). The two lesions are independent of each other and the primary arthropathy still exists when the perforating ulcer has been cured.

The perforating ulcer specially affects certain parts. Some cases have been mentioned as occurring in the *hand* (Peraire), but this was before the clinical symptoms of syringomyelia were well known, and the observations might perhaps now be revised as to this point. As regards the supposed perforating ulcer in the *viscera*, I must confess, gentlemen, that I am more inclined to consider it an erroneous expression than a reality. However this may be, the perforating ulcer, as a general rule, is seated in the *foot*, usually below the head of the 1st, and often below that of the 5th metatarsal bone, or sometimes below the heel. In some cases again it is found at other points in the sole of the foot. Since the researches of Duplay and Morat most authors attribute the perforating ulcer to some change in the

* Chipault, Le mal perforant. General Review in the *Gazette des Hôpitaux*, 1891, No. 83, p. 765.

cutaneous nerves, and the anæsthesia, which so often exists in and around the corn, is a strong argument in favour of this supposition. It is, however, evident that the mechanical effect

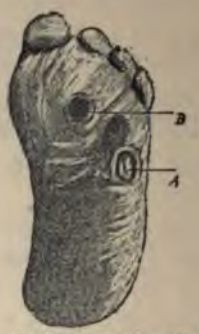


Fig. 146.—Sole of the foot, in which is a perforating ulcer, A, and traces of two other similar ulcers of more ancient date are seen.

produced upon the sole of the foot by walking or standing upright, takes a large part in its production, since the ulcer always occurs where the greatest pressure is made, either against the ground or the boot. Both feet may be affected, although frequently only *one* foot suffers from the complication, or two or more perforating ulcers may again occur in the same foot.

Gangrene has been observed by some authors in the course of tabes, not only towards the end of the disease, which would not be surprising since this is liable to occur in all wasting diseases, but at the beginning of the complaint. These would be examples of early gangrene with a rapid course: spots of blackish, violet, or reddish blue spots are first observed upon the buttocks, or over the trochanters or heels; vesicles then form in the same parts, the liquid of which soon becomes turbid and of a red or brown colour; the vesicles rupture and leave a bare, excoriated gangrenous surface of violet or black colour; this part is then eliminated with suppuration, and all the septic troubles with which you are acquainted. It is certain, gentlemen, that in all these cases some disorder of the nervous system exists, since it is to such an affection that the diminished nutrition of the skin which enables these complications to occur would in all probability be due. On the other hand, the progress of bacteriology has enabled us to know that gangrene, or mortification occurring

with such rapidity would not occur without the intervention and presence of more or less special septic micro-organisms. My opinion therefore is, gentlemen, that in the gangrene of tabes as, in fact, when it occurs in other affections of the nervous system, each of these two pathogenic elements has its special effect. At the same time the symptom is itself rare.

Spontaneous ecchymoses form an interesting episode in the history of the trophic and vasomotor disorders of tabes. They were first mentioned and studied by I. Straus.* It was shown by this author that they are in most cases obviously connected with the paroxysms of lightning pain, occurring when these cease and at the moment of their cessation. They are not, however, seated in the same place as the pains, but somewhat above the point at which these occur. This symptom, though not extremely frequent, is less uncommon than might be supposed to be the case; at the same time, from its requiring investigation, you will not be surprised that it is very frequently quite unperceived.

By the side of these trophic disorders of the skin you will allow me, gentlemen, to place others which may be logically classed with them; these are *shedding of the nails*, and *loss of the teeth*, as also the existence of *hyperidrosis* and *anidrosis*.

The *shedding of the nails* has been often observed in tabes, and I would specially quote the observations and memoirs of Joffroy, Fournier, Domecq-Turon, who have studied the conditions in which this complication occurs.

The nails of the toes are usually involved, and often in a symmetrical manner; this often happens more than once. The nail is generally shed without the association of other decided changes, becoming gradually detached from the free part towards the base, and shed without the occurrence of pain, "separating" in the same way "as an ecthymatous scab" (Fournier). Sometimes, however, tingling or slight pain is felt by the patient; more rarely, true lightning pains precede the shedding of the nails. Lastly, in some cases a patch of ecchymosis appears beneath the nail before it is shed, usually without the occurrence of pain. By the side of this shedding of the *nails* must be placed their *dystrophy* which sometimes occurs. They then

* I. Straus, Des ecchymoses tabétiques à la suite des crises douloureuses, *Arch. de Neurologie*, 1880—81, No. 4.

present the same appearance as that which occurs in some cases of eczema or psoriasis. Fournier, who has carefully studied them in these cases, describes varied forms. The nails, specially in the toes, are found to be much thickened, and the free extremity seems to be stratified, the surface is irregular and presents longitudinal or transverse streaks; the nails are usually hard and brittle. In short, they are affected by true dystrophy and pronounced alteration of nutrition due in all probability to the lesions of the nervous system.

The *loss of the teeth* has been observed for many years, since 1868 and 1869. Labbé and Dolbeau have mentioned cases of its occurrence. Amongst the most important memoirs on this subject those of Vallin, Demange, Galippe and David should also be mentioned, and the different opinions which are held will be stated to you.

This is what usually happens in such a case: without the patient having perceived that his teeth were in any abnormal condition, they gradually become loose and fall out without pain, and "as if they had been plucked," while after their separation the saliva may be but slightly tinged with the colour of blood, and the patient scarcely perceives what has happened. It sometimes occurs that when the tooth falls the corresponding fragment of the alveolar edge does so also; thus it has been possible (Dolbeau) to introduce through the opening which is thus produced, the finger or some instrument into the maxillary sinus, besides which considerable atrophy of the alveolar margin is found to occur after a certain time in the case of all the patients whose teeth have been lost.

Very different opinions have been formed as to the mechanism of this loss of the teeth; on the one hand Demange looks upon it as purely and simply due to trophic disorder depending upon change in the fifth nerve. David, whose opinion is somewhat analogous, found that a change occurred in the pulp of the teeth, and that there was atrophy of the crown. Comparing these lesions with those found in other organs connected with the senses he believes them directly due to the nervous disorders which exist in tabes, and the more so on acccount of the pulp being undoubtedly a sensory organ. On the other hand, Galippe does not consider that the nervous lesions sufficiently explain the existence of this symptom; in his opinion it is

principally due to periostitis and osteitis affecting the alveoli and teeth, as well as to the loss of the latter, and destruction of a portion of the alveolar arch, with which the presence of micro-organisms in the teeth is associated.

The last symptoms of which I would now speak are properly speaking disorders of secretion rather than of nutrition; but since we have specially considered in this lecture the symptoms connected with the skin which exist in tabes, it does not seem to me in any way illogical to speak at this time of the disorders which affect the *sudoriparous glands*.

Hyperidrosis, or an increase of their function, is not extremely rare in tabes; it may be unilateral or exist on both sides, and in the latter case may occur to such an extent as to be most inconvenient; thus, quite recently, I had the opportunity of seeing a patient suffering from tabes, who told me that he was obliged to change his flannel vest almost every half hour on account of the abundance of the perspiration. It should also be stated that in some patients this increase of function occurs in paroxysms which vary in duration, and are more or less completely absent during the intervals.

The opposite condition, *anidrosis*, or absence of perspiration, is also observed in tabes, and may also be unilateral, or exist upon both sides; the skin is then dry, and at times more or less covered by scales; even after a fatiguing exertion not a drop of sweat is secreted. It should be stated that in some patients who suffer during a certain period from hyperidrosis, more or less complete anidrosis subsequently occurs.

These are by no means the only disorders affecting *secretion* which occur in tabes, and in connection with such troubles I might mention different changes occurring in the composition of the urine, specially *glycosuria* which sometimes exists, the excretion of an exaggerated quantity of urine, a strange form of tabid diarrhœa, the hypersecretion of acid in the stomach in the gastric crises, &c., but I prefer deferring the study of these disorders until we speak of the different viscera, and of the complications and symptoms which occur in connection with them.

Lastly, an enquiry should be made as to the pathology of these trophic disorders in the skin. Are the nervous changes, upon which these disorders more or less directly depend, seated in the spinal cord or peripheral nerves? This is a question

which it is very difficult to answer; during the last few years the tendency of many authors has been to refer these symptoms to the existence of peripheral neuritis, which is observed in the course of tabes. Lesions in the spinal cord had been previously held responsible for their occurrence.

It is certain that with the means of investigation which we possess it is impossible to decide what part is taken by each of these. Allow me, however, to remark that on the one hand, notwithstanding the continually increasing number of cases of polyneuritis from some cause or other, trophic troubles, analogous to those which I have just described as occurring in tabes, have scarcely even been mentioned; on the other hand, in an affection which seems clearly connected with the spinal cord, syringomyelia, very similar symptoms do at times exist. This argument, as I admit, is of an indirect character, but, as it appears to me, it enables us to state that some, at any rate, of these trophic disorders has for its origin a lesion in the spinal cord.

LECTURE XXI.

TABES.

SYMPTOMS (*continued*).

Muscular atrophy: Distinction from the conditions of muscular emaciation, which are due to consumption. Is specially seated in the lower limbs, and most often bilateral; occurs also in the upper limbs and in the muscles supplied by nerves coming from the medulla oblongata. Onset. Extension. Fibrillar contractions. Electrical examination. Evolution. Tabid club-foot of Joffroy. *Pathological Anatomy* of these forms of amyotrophy; muscular lesions; opinions as to the nature of the nervous lesion (in the spinal cord, in the peripheral nerves). *Hemiatrophy of the tongue:* its symptoms. Appearance of the tongue, paralysis of the soft palate, and inferior vocal cord upon the same side. Lesions of the medulla oblongata in cases of hemiatrophy of the tongue. Conclusions which may be drawn owing to the co-existence of paralysis of the soft palate and vocal cord. Attempt to range the forms of amyotrophy occurring in tabes in two distinct groups.

GENTLEMEN,—In the preceding lectures we have reviewed the trophic disorders which occur in tabes in connection with the bones, joints, and skin; nor are the muscles exempt from them, and the *muscular atrophy* which occurs deserves special consideration. This symptom was some years ago studied in the most complete way by Condoléon,* the pupil of Joffroy. I shall have occasion to quote more than once from his remarkable inaugural dissertation.

Condoléon was preceded in the study of this subject by several authors, amongst whom I would quote Charcot and Pierret, who were the first to call attention to this symptom, the occurrence of which they explained, Leyden, Westphal, Dejerine, Eulenburg, Joffroy, &c., who published interesting or important observations upon the theories which will be brought before your notice by me.

Before commencing to describe the *muscular atrophy* which occurs in tabes, it must be understood, gentlemen, that by this

* Condoléon, Contribution à l'étude pathogénique de l'amyotrophie tabétique *Thèse de Paris*, 1887.

name we term the condition of muscular *emaciation*, *flaccidity*, and *weakness*, which so often occurs in patients suffering from this disease, specially in its later stages. As I have said more than once, tabes = consumption; this is a condition of the muscles which is found to exist in every consumptive disease, and has nothing in it which is special to tabes. Thus, *true muscular atrophy* will now be alone considered.

This muscular atrophy, according to Condoléon, is usually *seated* in the *lower limbs*, specially in the muscles of the *leg below the knee*, and the *foot*; it usually occurs on *both sides* but is sometimes *unilateral*.

In some cases, however, it occurs in the *upper limbs*, usually then occupying the *small muscles of the hands*. More rarely it exists in the *forearms*, or *arms*, at the *shoulder*, and in certain muscles of the *back*.

The *muscles supplied by nerves coming from the medulla oblongata* may be similarly affected; thus Schultze found it to occur in the muscles supplied by the *motor root of the 5th nerve*. We shall soon see that it also occurs somewhat frequently in those supplied by the *hypoglossal nerve*, facts to which I shall return, as they are of considerable interest, on account of the definite seat of the lesions to which they are due. Very different opinions have been held as to the *frequence* of this symptom. Eulenburg, for example, considers it rare, and says that it is only found to occur once in 250 patients suffering from tabes. This, gentlemen, is a mistaken estimate. The symptom is far less uncommon, I would almost say that it occurs with some frequency, specially in the lower limbs, and cases of the *tabid club-foot* of Joffroy are notably observed in large numbers.

The *onset* usually occurs in a late period of the disease. In some cases, however, it has been found to occur in a much earlier stage, and even during the prodromic period (Charcot, Fournier).

The degree to which the muscles waste is sometimes considerable, and you will easily recognize this for yourselves in the photographs which I put before you. The hands may present a somewhat "claw-like" attitude, while the width of the muscles of the shoulder and trunk is found to considerably exceed the thickness.

The *fibrillar contractions* are usually, but not always, absent;

when they exist it is usually when the amyotrophy occurs early in the disease.

Fig. 147.—Atrophy of the muscles of the right shoulder and of the hands in a case of tabes. (Collection of Charcot.)

The *electrical examination*, according to Condoléon, usually shows considerable diminution of electrical irritability. The response of the muscle to the electric currents is otherwise unchanged; Joffroy, however, in a case of club-foot found that the reaction of degeneration partially existed in the peronei and extensor muscles of the toes (retention of faradic and galvanic irritability in the peroneal nerve, diminution of faradic irritability in the muscles, slight increase of galvanic irritability with stronger contraction at the positive pole).

The *course* of these forms of amyotrophy is but relatively progressive; that is to say, the atrophy may gradually increase in

one group of muscles, but in other ways it rarely if ever shows any tendency to progressive change, and will not be found to extend slowly from one group of muscles to another, as occurs in some forms of progressive amyotrophy.

Such are the general facts which the present state of our knowledge enables us to state with regard to the muscular atrophy which exists in tabes. I would now consider in a somewhat detailed way a special form of amyotrophy—the *tabid club-foot*.

The *tabid club-foot* has only been studied and described within the last few years by Joffroy. The observers who met with such cases before that time merely noted their occurrence in a summary way, attributing them to the existence of tonic contraction, the effect of which was combined with that produced by the weight of the bedclothes upon the feet.

I cannot better describe the deformity which constitutes the tabid club-foot than by quoting the words which Joffroy himself uses. He alludes to the evolution of this deformity and its characters in the following terms:—

"The affection begins in the foot being permanently placed in the position of pronounced extension; in addition to this the extremity of the foot is inclined inwards, the inner side of the sole being hollowed out and raised, so that in reality *talipes equino-varus* exists, and since the lesion is usually symmetrical, the result is, that when the patient is in the dorsal decubitus, the heels are separated by 10 centimètres (4 inches), the two great toes as they approach each other leaving an angular space between the feet.

"When the condition becomes more accentuated the toes are bent to a very pronounced degree; the toes cannot be then straightened, or the foot be bent upon the leg; the impediment is clearly seated in the position of the tendo Achillis, but the calf-muscles are not necessarily contracted, as Leyden supposes, since palpation of the wasted muscles shows them to be in the same relaxed flaccid condition as the antero-lateral muscles of the leg, in addition to which, if the leg is raised and moved about, lateral displacement of the foot can be easily obtained, which can scarcely be effected when the calf muscles are contracted, and extend the foot. Electrical irritability is retained in all the muscles of the leg. (It has already been observed,

however, that in one case Joffroy found partial reaction of degeneration to exist.)

Fig. 148.—Tabid club-foot of Joffroy. (Collection of Charcot.)

"The tabid club-foot neither results from contracture, for the reason already stated, nor from such atrophy as exists in infantile spinal paralysis, nor are the bones concerned with its production, but it is a club-foot produced by flaccidity—an atonic club-foot. In these conditions the foot, owing to the weight of bedclothes, being constantly in the position of flexion, elongation of the anterior ligament of the tibio-tarsal articulation, or rather of the fibrous sheaths enclosing the tendons and taking the place of ligaments, occurs; this elongation is facilitated by the flaccidity and atony of the muscles of the leg; on the other hand, the contrary occurs in the region of the tendo Achillis, which is shortened either on account of adhesions, fibrous bands, or muscular contraction; in no case does contracture of the gastrocnemii muscles occur, which are as flaccid as the muscles in the anterior region of the leg.

"The prophylactic treatment is to prevent the bedclothes from weighing down the foot, or if necessary, to apply a simple orthopædic apparatus."

Such is the *tabid club-foot*, gentlemen, which, notwithstanding the similarity of the names, you must not confuse with the *tabid foot*; the latter is, as you have seen, the consequence of an osteo-arthropathy, whereas the former is due, as we have just said, to atrophy of the muscles of the leg. I should add, that besides

this muscular atrophy, another element takes part in its production; this element is constituted by the trophic disorders of the articular ligaments which Charcot has taught us to know, notably in cervical hypertrophic pachymeningitis and alcoholic paralysis, and in which he has shown us the good effect of tenotomy.

What, as regards its *pathological anatomy*, are the characters of the muscular atrophy which occurs in tabes?

The *muscles* are found to be wasted, of yellowish hue, and have sometimes the colour of dead leaves; but it should be observed that in the midst of the degenerated muscular fasciculi others are nearly always found in greater or less number in a healthy condition. Thus by dissociation fibres differing in appearance may be obtained:—

(*a*) Fibres which are much degenerated, varicose, and are totally deprived of their striæ enclosing a large number of the nuclei of the sarcolemma and fine granulations of a yellowish brown colour and strongly refractive.

(*b*) Fibres which are much reduced in diameter but retain their normal striation.

(*c*) Fibres which are absolutely healthy. You will observe, gentlemen, that the different fibres of a muscle are not usually in the same condition; but on the other hand, as in the microscopical preparation which I put before you, the different varieties of fibres form distinct fasciculi which degenerate independently of each other.

This association of healthy and diseased fibres will explain (Rumpf) the fact that the reaction of degeneration is usually absent since the muscle nearly always contains enough healthy fasciculi to produce the normal response to electrical irritation.

What, however, is the cause of this muscular atrophy? Upon what lesion of the nervous system does it depend? According to some authors it is due to *peripheral neuritis*, the spinal cord, anterior roots, and large nerve trunks remaining unaffected. These authors specially rely on the researches of Westphal, Pierret, Dejerine, &c., who have shown that lesions frequently exist in the peripheral nerves in tabes.

The opinion first promulgated by Charcot and Pierret, professed by Leyden, and accepted by numerous authors, is that muscular atrophy in tabes is due to a lesion in the *grey*

matter of the anterior horn; this lesion according to Charcot and Pierret is consecutive to extension of the change which occurs in the posterior columns.

Fig. 149—Section of a portion of the thenar muscles from a patient with tabes suffering from very pronounced amyotrophy. (From a preparation taken from the patient who forms the subject of fig. 151.) AA. Muscular fibres in a healthy condition; BB. degenerating muscular fibres; C. band of connective tissue between the fasciculi. It will be observed that the fasciculi of wasted fibres alternate with those which are sound, and that the degenerated fibres are disseminated in the midst of those which are sound.

On account of this difference of opinion the researches of Condoléon are of great interest, since the special object of these was to ascertain which of these two theories was most in accordance with the existing facts. The results to which they led Condoléon were as follows: lesions were found to exist in the cells of the anterior cornua with very slight changes in the anterior roots and large nerve trunks, and on the other hand pronounced lesions in the intra-muscular nerves. Thus Condoléon is of opinion that the hiatus in the seat of the lesions fully explains the fact that in certain cases the change in the cord was ignored, and the peripheral neuritis alone recognized.

In support of the opinion that the lesion which causes the muscular atrophy of tabes is seated in the nerve centres, and only secondarily in the peripheral nerves, I shall mention to you, gentlemen, in my turn, as an argument which cannot be refuted, facts which have been shown by many authors and by myself in connection with the *hemiatrophy of the tongue* which occurs in tabes.

In this case the anatomical investigation is made in conditions which are specially favourable since it is known in which definite group of cells in the medulla oblongata the lesion should be sought, and the hemiatrophy of the tongue being a unilateral symptom the nuclei of the medulla oblongata can be compared with each other, and the least alteration which occurs in one of them be definitely recognised. The result of these researches was to show clearly that characteristic lesions existed in the nerve centres. Before mentioning these it would not, I think, be useless to describe somewhat in detail this *hemiatrophy of the tongue* which occurs at times in tabes and is accompanied by an interesting series of symptoms. As was the case with regard to most of the trophic disorders in tabes which we have already considered it is again Charcot who first called attention to the *hemiatrophy of the tongue* which occurs in that affection; this subject was again taken up at a later date and developed by Gilbert Ballet; amongst the principal observations upon this symptom I must quote that of Raymond and Artaud, with the autopsy and histological examination very carefully made by Mathias Duval; the thesis of Arnaud* contains interesting documents from a clinical point of view. I had the good fortune to obtain possession of the diseased parts taken from the patient under the care of Charcot and observed by Ballet and Arnaud; P. D. Koch (of Copenhagen) and I made the microscopical examination, and this enables me to put before you certain original sketches which represent the appearance of the patient and of the lesions found at the autopsy.

Hemiatrophy of the tongue is not extremely rare in the course of tabes, and, as we shall see, occurs also without being connected with this affection. Its onset occurs slowly, without the occurrence of any inflammatory symptom, so that the patients

* J. Arnaud, De l'hemiatrophie de la langue dans le tabes dorsal ataxique. *Thèse de Paris*, 1885.

seldom know that any diminution has occurred in the size of their tongue. It is the medical examination which reveals it to them.

In every, or almost every, case the clinical aspect is identically the same. The appearance of the tongue when protruded is found to be that of a curved surface, in the form of a small crescent enclosed in one which is larger. The side affected by atrophy (the small crescent) is shrivelled, reduced in size, placed

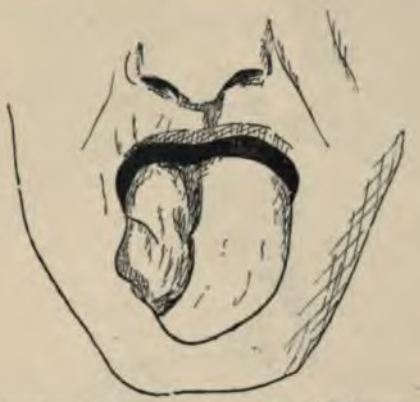


Fig. 150.—Hemiatrophy of the tongue in a case of tabes (in this diagram the hemiatrophy is seated on the right side, the figure having been turned round; in reality it was on the left side). (Collection of M. Charcot.)

at a somewhat lower level than that of the sound side. A series of furrows, which are more or less twisted, exist in it, which give to it the appearance of convolutions affected by atrophy. The tip of the tongue is distinctly turned towards the side of the atrophy.

When the tongue is taken between the fingers (J. Hutchinson) it is found that when that organ moves the half in which atrophy has occurred does not harden, whilst, on the other hand, the ordinary change occurs in the consistence of the other half.

Observe, gentlemen, that however considerable the deviation and atrophy of the tongue may be, all its movements are still possible, except that of giving itself a grooved surface. Hence no appreciable functional disorder exists. The speech, mastication, and deglutition are unaffected, and, as I have just said, the patients remain ignorant during an indefinite time of the existence of hemiatrophy in this organ.

In some cases slight fibrillary movements are found to occur upon the affected side.

Lastly, and this is a fact which is specially remarkable, and upon which Koch and I dwelt in our memoir upon hemiatrophy

Fig. 151.—A patient suffering from tabes with hemiatrophy of the tongue on the right side: the tip of the tongue is turned to the right side, the whole organ being "in the form of a crescent." In this patient there was also ptosis in the right eye (collection of Charcot). This is the patient whose autopsy P. D. Koch and I made, and whose medulla oblongata we examined.

of the tongue, there exist in the same patients *paralysis of one side of the soft palate*, upon the same side as the atrophy of the tongue, and of the corresponding *inferior vocal cord*. I shall endeavour in a few moments to explain this singular coincidence.

The course of the affection is otherwise benign; although

progressive it remains limited to the side affected and never changes to labio-glosso-pharyngeal paralysis; that is to say severe, and possibly fatal bulbar paralysis does not occur.

Fig. 152.—Section of the tongue from a case of hemiatrophy on the right side (from the patient who is the subject of fig. 151). (After P. D. Koch and P. Marie.)

I have already said, gentlemen, that hemiatrophy of the tongue occurs with the same characters in other affections than tabes; it has been observed in *general paralysis of the insane*, which is not surprising when the close analogy which exists between these two affections is remembered, which causes them to have more than one symptom in common. It has been also observed, and these perhaps are the most frequent cases, in *syphilis*, affecting the pons varolii and medulla oblongata. This is a fact upon which I specially insisted in my work written in collaboration with P. D. Koch. Gouguenheim and Leudet had also mentioned it some weeks previously. This ætiology is, in my opinion, of very great interest, since, it being allowed, as we shall see in a subsequent lecture, that the greater number of those who suffer from tabes or general paralysis of the insane have been affected by syphilis, it may be asked whether the hemiatrophy of the tongue is due to syphilis or to tabes. In other words the question is whether hemiatrophy of the tongue and tabes are connected affections, and the former depends directly upon the latter, or the two are merely associated disorders which should be considered as different manifestations of one and the same general disease, syphilis.

However this may be, the lesions of the bulb in cases of hemiatrophy of the tongue are clear. The *large nucleus* of the hypoglossal nerve on the side corresponding to that of the lingual hemiatrophy is much smaller in size than that on the opposite side, a certain number of nerve cells having disappeared, as seen in the diagram which I now place before you. In addition to this the *accessory nucleus* upon the same side shows analogous changes

(Mathias Duval and Raymond, Westphal, P. D. Koch and P. Marie); the *afferent fibres* in the nucleus of the hypoglossal

Fig. 113.—Section of the medulla oblongata at its lower part from a case of hemiatrophy of the tongue upon the left side. (After P. D. Koch and P. Marie.) The middle part of the floor of the 4th verticle is alone represented (the sinuous line below the figures); above this are the nuclei of the hypoglossal nerve. That on the left side is much reduced in size, as are the cells which it contains and the fibres which emanate from it. (In its reproduction the figure has been turned round, it is therefore the nucleus of the hypoglossal nerve upon the right side which is wasted.)

nerve, described by P. D. Koch, are not apparently affected. On the contrary, the *radicular fibres* of the hypoglossal nerve present clearly atrophic changes upon the same side as that of the lingual hemiatrophy.

It is a singular fact that no changes similar to those in the nucleus of the hypoglossal nerve are found either in that of the pneumogastric or spinal accessory nerve, although, as we have already seen, there is paralysis of the corresponding vocal cord, and half of the soft palate. In order to explain this anomaly P. D. Koch and I made a different suggestion from that first proposed by Hughlings Jackson and Henschen. In our opinion the muscles of the larynx and soft palate, besides the fibres which connect them with the nuclei of the pneumogastric and spinal accessory nerves, receive fibres also from the hypoglossal nucleus. Something of an analogous nature occurs in the eye; thus, for example, the internal rectus receives at the same time fibres from the nuclei of the 3rd and 6th pair of nerves

(Graux-Féréol, Mathias Duval). When a change has occurred in the nucleus of the latter nerve this muscle is found to be

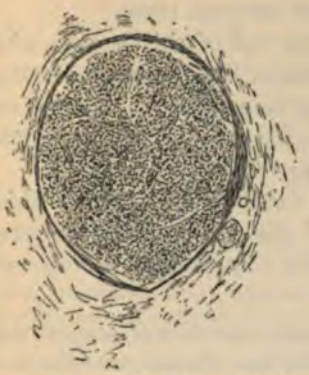


Fig. 154.—Section of the left hypoglossal nerve (normal) from a case of hemiatrophy of the tongue in tabes. (After P. D. Koch and P. Marie.)

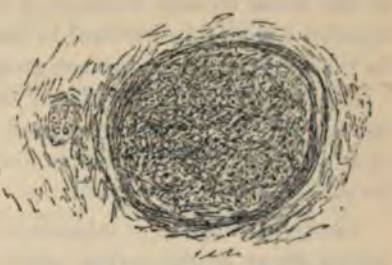


Fig. 155.—Section of the right hypoglossal nerve (diseased) from a case of hemiatrophy of the right side of the tongue in tabes. (After P. D. Koch and P. Marie.) The nerve fibres which are in a normal condition and have retained their sheath of myelin (black points) are very few.

paralysed to a certain extent, viz., in respect to certain positions of the eye, whilst its functions as regards other positions are totally unaffected. The same is the case as regards paralysis of the soft palate and larynx in tabes, which result from this change in the hypoglossal nucleus. Considerable disorder is seen to exist in these two organs, although there is, so to speak, no functional trouble. The fibres coming from the hypoglossal nucleus act therefore in a special way which differs from the action of the fibres issuing from the nuclei of the 10th and 11th pair of nerves. I cannot give you more information as to this point since these facts are as yet but little known; all I can do at present is to call your attention to their existence.

This is what I wished to say about lingual hemiatrophy. In connection with the disease which we are now studying, its consideration cannot be looked upon as a mere digression, since these cases show us in the most undeniable way that one form at least of the amyotrophy which occurs in tabes is accompanied by obvious change in the central grey nuclei.

I said "one form at least of the amyotrophy which occurs in tabes," since I must confess, gentlemen, that I am doubtful

whether all the cases of amyotrophy in tabes are of the same kind, and whether it is not possible, at any rate from a clinical point of view, to establish a decided difference between them.

As regards my own views, if at the end of this lecture, after having mentioned the actual condition of the question according to the ideas formed about it, I may be permitted to state my opinion I should be inclined to range the varieties of amyotrophy in tabes in two large distinct groups :—

In one (A) would be placed the forms of amyotrophy which usually occur at a late period of the disease, are disposed symmetrically, and in which fibrillar contractions can scarcely be said to occur.

In the other (B) I would place those varieties of amyotrophy which occur at an earlier period of the disease, of which it may be specially said that they are not disposed symmetrically, and in which fibrillar contractions are more frequent, and the reaction of degeneration exists more frequently than in those of the group A.

The tabid club-foot of Joffroy is the type of the forms of amyotrophy which belong to the first group, and which are in consequence very analogous to those which occur in different forms of intoxication, which have been ranged in quite a transitory way, as I think, among the forms of multiple neuritis ; it is to this type alone that the opinion of authors who contend with Dejerine that the amyotrophy of tabes is due to peripheral neuritis can refer.

The forms of amyotrophy belonging to the second group would include lingual hemiatrophy, some varieties of atrophy affecting the muscles of the shoulder, the back, and perhaps the hands; they would be secondary to lesions which clearly exist in the grey matter of the medulla oblongata or spinal cord.

The classification which I have just sketched, gentlemen, is simply a suggestion of which the value must not be exaggerated ; it seems to me that from a clinical point of view the differences which existed were sufficiently pronounced to justify the formation of these groups, and that, even when the pathological anatomy of this condition is considered, such a distinction makes it possible to explain the different opinions which have existed. This suggestion, gentlemen, I leave in your hands ; the future will judge of its value.

LECTURE XXII.

TABES.

SYMPTOMS (*continued*).

VISCERAL DISORDERS.—I. *Digestive system:* (*a*) *Stomach.*—α. Gastric crises, their character; pain, uncontrollable vomiting, excess of secretion, and acidity of the gastric juice, researches of Sahli, Rosenthal, Hoffmann, state of nervous depression, its abrupt onset, and sudden disappearance; tendency to relapses. Abnormal forms.—β. Tabid annorexia.—(*b*) *Intestine.*—α. Intestinal tenesmus.—β. Tabid diarrhœa,

II. *Vascular system.*—(*a*) *Heart:* lesions of the mitral, and aortic valves.—(*b*) *Blood vessels:* Arteriosclerosis; angina pectoris; exophthalmic goitre, works published upon this subject.

GENTLEMEN,—Until the present time we have specially considered the symptoms in tabes which are connected with the limbs or organs of the senses; other symptoms have still to be discussed, which occur almost as frequently, are as important, and also form part of the ordinary clinical aspect of the disease; these are the derangements which occur in the viscera.

THE VISCERAL DISORDERS in tabes are numerous, and scarcely any system is free from them; they will be reviewed as they occur in the different organs.

I.—DIGESTIVE SYSTEM.

A. STOMACH.—Amongst the symptoms which occur in tabes in connection with this organ, the most interesting is undoubtedly the gastric crisis.

a. Gastric crisis. Although examples of its occurrence are found in the work of Topinard, and in that of Delamarre (1861), it is again to Charcot (1872) that we owe the knowledge of this singular symptom; the description which he gives in his first lectures remains always classical; more recently in his "Leçons du Mardi"* the eminent professor of the Salpêtrière

* J. M. Charcot, *Leçons du Mardi*, 1888—89, p. 333.

Hospital again discussed this symptom, adding other facts and extending his classification; it is this description which should guide us in the study which we are about to commence; I must mention also to you certain facts which were brought to light by Fournier.

Many of you have without doubt been already present at one of the dramatic episodes of tabes which are termed *gastric crises* since these are far from rare; in whatever medical ward you may prosecute your studies, one case at least is almost certain to come before your notice.

The two symptoms which specially characterise the gastric crises in tabes are: (*a*) the *pain*, (*b*) the *uncontrollable vomiting*.

(*a*) The *pain* corresponds to that of gastralgia; it is felt in the epigastrium, being at times limited to that part, but more often extending in different directions, towards the abdomen, the side or the back; very severe in the great majority of cases, it is at times absolutely agonising, seeming to exceed what a human being can bear in the degree of suffering.

(*b*) The *uncontrollable vomiting* is at first of the food taken, but soon afterwards exclusively of slimy liquid, or may perhaps consist of a clear liquid, mucous in character and more or less abundant; at times the vomited matter is coloured by the bile or more rarely by blood; it may quite exceptionally present the appearance of "coffee grounds" (a case under the care of Vulpian, one under that of Charcot). Usually the vomiting occurs very frequently, but varies in abundance; at times but a few grammes (1 grm. = 15·432 grs. Troy) of liquid are vomited on each occasion, but sometimes a great deal more; these different characters vary according to the patient and in the same patient from day to day.

During the last few years different works have appeared about the composition of the liquid vomited in the gastric crises; the conclusions formed are certainly not as yet quite established, but that is no reason whatever, gentlemen, why you should fail to be interested in the researches which have taken place.

In 1885 M. Sahli* stated that an *excess of the secretion* and of *the acidity* of the gastric juice occurred but without giving a quantitative analysis of the secretion. In the fo
(1886), upon examining the liquid vomited during

* H. Sahli, *Correspondbl. f. Schweizer Aerzte*, 1

of the gastric crisis, Rosenthal found during the first days 0·30 per cent. of hydrochloric acid, and during the last days only 0·10 per cent.: now according to Ch. Richet the mean quantity of hydrochloric acid in normal gastric juice is 0·174 per cent. Thus the figure obtained by Rosenthal clearly indicates, as you see, that there is an excess of acid in the liquid vomited. A similar excess of acid in the vomit during the existence of the gastric crisis in tabes was also independently mentioned by Simonin (of Lyons) in 1886.

More recently, Hoffmann, a distinguished pupil of Erb, again studied this subject; in one of his patients he also found that the gastric juice was secreted in excess, and contained temporarily an excess of acid; the quantity of hydrochloric acid varied, often amounting to twice the ordinary amount, and being rarely less than the normal quantity, and this only when owing to the abundance of the matter vomited the acid was somewhat largely diluted.

On the other hand Hoffmann was not able to prove the fact stated by Rosenthal, that the amount of hydrochloric acid diminished or increased according to the intensity of the crisis. A cause of error in this respect was very aptly suggested by Hoffmann, consisting in the fact that the more severe the vomiting the greater is the thirst, and the amount of liquid injected consequently more considerable, the gastric juice being diluted, and therefore less acid in reaction.

Such great technical difficulties are associated with the study of these facts, and so few attempts have been made to elucidate them, that it is quite intelligible that no definite results have yet been obtained, though it seems most probable that such will soon be the case.

In order to terminate this question of the acidity of the gastric juice in gastric crises, I must add that in some cases besides the *hydrochloric acid*, *lactic acid* has been met with in small but varying quantities.

You will easily understand, gentlemen, the sensation of *weariness* which is produced in t unate patient by the severe pain and r would have but an incomplete d to describe this third symp

(*c*) A ounced,

varying from simple indolence, from an indifference on the part of the patient to all which surrounds him, absorbed as he is in his own suffering, to true stupor. In the cases of exhaustion from these terrible shocks the unfortunate patients become almost unconscious, not answering when they are spoken to, or having even the strength to replace their bedclothes when they are displaced. At the same time this precaution is by no means useless, since when the affection is of this severity the patient is not rarely found cold, in a state of *algidity*, while the surface becomes of a livid colour, and to such an extent, gentlemen, that medical advisers who have not been forewarned, being struck by the severity of the vomiting and the algid condition, have made the diagnosis of cholera. In most cases the strongest and most obvious resemblance is to those who suffer from sea sickness, and it occurs in some people who are specially susceptible to it.

Another character of the gastric crises is the rapidity of their evolution.

(*d*) *Sudden onset. Abrupt cessation.*—The onset is usually quite unexpected, and the crisis may continue without remission by night and day during 2, 3, 4, 8, 15 days, rarely more, becoming more severe when the least attempt is made to take food, or to swallow liquid, although the thirst is often truly distressing. Then as if by enchantment the vomiting and pain cease, a sensation of relief is felt by the patient; the appetite sometimes returns almost immediately, and in this case the patient, who during several days had not been able to retain the smallest part of his food, is able to take a somewhat abundant meal without being at all inconvenienced by it.

The gastric crises are usually early symptoms in tabes, often occurring even in the prodromic period.

They are rarely isolated, that is to say, the patient seldom suffers from but one crisis of this kind; usually, when they have once occurred, they *tend to recur* more or less frequently, at times indeed taking place somewhat periodically; as for example every year, every 6 months, or every 3 months. As regards what ultimately happens, either of the two following sequelæ are possible: not infrequently the gastric crises after a few years gradually diminish in number and intensity so as to completely cease; or they may persist during the whole duration of the disease, death at times occurring during one of the crises,

whilst the patient is in the collapsed condition, or after the onset of coma.

Such is usually the aspect which the gastric crisis presents, but you should know, gentlemen, that it does not always occur in this manner. In his lectures Charcot did not fail to study its abnormal forms.

A. The gastric crisis retains all the fundamental characters of the type, but the *pain* is so agonising as to simulate that which occurs in hepatic or nephritic colic, or from poisoning.

B. The crisis is accompanied by such a degree of *collapse* that the symptoms are attributed to severe malarial fever, cholera, or even to an organic cerebral affection.

C. *There is no vomiting*, the only symptom being pain, often very severe, and occurring in paroxysms, or at any rate presenting exacerbations (the cramp-like pain of Fournier).

D. On the contrary *there is no pain*, but vomiting exists, presenting the characters which have been already mentioned (Vulpian, Pitres, Fournier).

E. Fournier has described a *flatulent variety* of gastric crisis, characterised by the large quantity of gas discharged by eructation.

F. The crisis is sometimes of *very* and exceptionally short duration, lasting at the most but a few hours. On the other hand Blocq, who observed facts of this nature, remarks that the crises may occur daily, and continue during a somewhat considerable length of time.

The *duration* of the gastric crisis may again be *prolonged*, and continue during from 15 or 20 days to a month, or even more. A patient observed by Buzzard informed that author that during 9 months consecutively he had suffered from gastric crises. It should be added that in such cases the symptoms are not equally severe during the whole period, but present times of exacerbation or remission.

β. Another interesting derangement of the stomach which occurs in the course of tabes is that which Fournier has described under the name of *tabid anorexia*. This symptom, however, is very rare, even in the opinion of this author. It consists in the fact that the patient has lost the sensation of hunger; at the same time slight vomiting occurs at times without any appreciable cause. The patient, so to speak, ceases to

be nourished; he has an insuperable aversion to food, and only eats or drinks from a sense of duty, since, as I have already said, the feeling of hunger is completely absent. Notwithstanding this loss of appetite, and the vomiting which occasionally occurs, the tongue remains moist, clean, and free from deposit.

This, briefly speaking, is a more or less exact reproduction of the singular condition which Charcot made known to us by the name of hysterical anorexia. Owing to its complete resemblance, owing to the frequent co-existence of tabes and hysteria in the same patient, it seems to me, gentlemen, that the occurrence of tabid anorexia may at times be due to the association of these two affections. Subsequent observations will enlighten us upon this point.

B. INTESTINE.—Fournier described not only the derangements of the stomach which I have just mentioned, but also intestinal symptoms which occur in tabes, which he carefully studied, and which he has graphically delineated. With this author we may divide these symptoms into two principal groups:—(1) intestinal tenesmus; (2) tabid diarrhœa.

a. Intestinal tenesmus. This consists in a desire to evacuate the bowels, which, as Fournier expresses, is remarkable in three ways: (1) on account of its strong character (it must be gratified at once; while fearing to soil his clothing the patient finds it absolutely impossible to wait); (2) on account of its unusual frequency (4, 6, or 10 times in the same day); (3) on account of the absence of any cause for it (when the patients have the opportunity of relieving their pressing need very little or perhaps nothing at all is passed). The defecation is again quite inactive, not being associated either with spasmodic contraction of the sphincter, or the sensation of a foreign body in the rectum.

β. Tabid diarrhœa. This symptom is somewhat frequently observed in the prodromic period (Fournier); it thus usually occurs as an early symptom. It consists of frequent pasty or liquid stools, usually in small quantity; the diarrhœa being in most cases attended by little or no pain, and unaccompanied, so to speak, by colic.

The characters specially characteristic of this form of diarrhœa are, on the one hand, the *absence of cause*, the diarrhœa occurring in fact quite spontaneously, and without the occurrence of any

irregularity in the diet, exposure to cold or other cause, being, as Fournier expresses it, a form of "essential diarrhœa." On the other hand it *persists* almost indefinitely, and may continue during 2, 3, or 4 years, certainly with intermissions and exacerbations. In addition to this, and which is also a new character, treatment is absolutely useless as regards this form of diarrhœa, which you will understand, although not specially serious as far as the general health is concerned, is a very great inconvenience to the patients who suffer from it.

II.—Vascular System.

Lesions and derangements frequently exist in this system, much more often, in fact, than is usually believed to be the case, since, as must be confessed, the patients suffering from tabes are too seldom examined with regard to the occurrence of such symptoms. If this was more often done their existence would be more frequently recognized. The lesions and derangements are numerous, and affect the heart and blood vessels in different manners. In connection with the vascular derangements I shall have to speak to you of a complication very recently observed, viz., exophthalmic goitre.

(*a*) *Heart.* A somewhat large number of works have been devoted to the study of the lesions which may occur in the heart during the course of tabes, but I shall not enumerate them, and the more so as the conclusions formed in them are far from being the same; according to some, in fact, lesions of the *mitral* valve are by far the most frequent; according to others lesions of the aortic valve; I must add, gentlemen, that in my opinion there is no doubt about this point. An examination made in numerous cases of tabes has shown me that lesions of the aortic are infinitely more frequent than those of the *mitral* valve. With regard to the latter, stenosis is specially found to exist, whilst in the former incontinence is the most common.

Although statistics do not enable me to state the frequency of cardiac lesions in tabes, they may certainly be supposed to exist in a somewhat large number of cases; I would even say that if four or five patients be taken fortuitously, who are suffering from tabes at an advanced period of the disease, more or less

definite indications of a cardiac lesion would be found in at least one of them.

(*b*) *Blood vessels.* The vascular lesions in the course of tabes are comparatively less known than the cardiac; one fact is however certain, namely, that they are frequent, and may be observed in the small arteries as much as in the large ones. *Arterio-sclerosis*, with its different aspects and various consequences, occurs with some frequency in tabes. The existence of this vascular lesion has indeed been the base of certain theories formed about the nature of tabes, and, a point which now specially interests us, about the nature of the cardiac changes of which I have just spoken.

The following are the opinions which have been formed in order to explain the production of cardiac lesions:—

According to some authors (Berger, Rosenbach, Grasset, &c.), these lesions are due to real trophic disorder of tabid origin, which affects the valves or the heart itself. J. Teissier goes so far in this respect as to pronounce the expression "perforating ulcer of the aortic valve."

According to other authors (Adamkiewicz, Rumpf, Hippolyte Martin, Letulle, &c.), tabes and the vascular lesions would progress in a parallel manner, the lesion in the cord being the direct consequence of vascular lesions localised therein, and occurring at the same time as in other regions of the body.

I must confess, gentlemen, that I am not quite satisfied with either of these opinions, and for my part am much inclined to doubt whether tabes and the cardio-vascular affections are thus intimately connected as cause and effect; the explanation of the frequency with which cardio-vascular lesions occur in tabes is more probably, in my opinion, the fact that in tabes the great majority of patients have suffered from syphilis; now syphilis is one of the most important causes of cardio-vascular lesions, specially of those connected with the aortic valve; it is not therefore surprising that in tabes lesions of the spinal cord and of the heart and blood vessels occur simultaneously in the same way as symptoms connected with the skin and mucous membranes.

As regards cardio-vascular disorders there is one of which a few words should be said, namely, *angina pectoris* (Vulpian, Leyden), which not unfrequently occurs in tabes, and presents the usual

symptoms of that complaint; the sense of constriction and pain behind the sternum, often agonising, the extension to the left arm, the feeling of approaching death, &c. According to Leyden, this "angor pectoris" is due to the fact that the cardiac branches of the vagus are involved in the morbid process of tabes, and the professor of Berlin remarks in support of this idea that the symptom is at times associated with gastric crises (case of Vulpian); on the other hand Oppenheim has observed in certain cases of gastric crisis a premonitory sensation of agonising pain and numbness in the left arm; these different characters would indicate, according to Leyden, that the branches of the vagus participate in the affection. This is quite possible when the multiplicity of the nervous lesions which exist during the course of tabes is borne in mind, but in my opinion the question may be asked whether this is always the case, and whether the lesions of the aorta, of which I have recently spoken, are not in themselves sufficient to produce the symptoms of angina pectoris without its being necessary to associate it with the morbid process of tabes.

With the different cardio-vascular symptoms I will connect, in order to facilitate the classification, but without wishing to dogmatise, another singular phenomenon which has been recently observed—I mean *the coincidence of the disease of Graves and tabes.*

Attention was drawn to these facts in the same session of the "Société des Hôpitaux,"* by Barié and Joffroy. The latter communicated six observations of their occurrence at the same time. Much surprise was caused, but there was no doubt about the fact; the patients were undoubtedly suffering from tabes, since the lightning pains, the symptom of Westphal, the symptom of Romberg, the inco-ordination of movement, the plantar anæsthesia, and the loss of sexual power were present; again, the disease of Graves existed since the exophthalmos, the rapidity of the heart's action with hypertrophy of this organ, the paroxysmal enlargement of the thyroid, the tremor and polyuria, &c., were all recognized to exist.

This coincidence of the disease of Graves and tabes, is it of extremely rare occurrence? Certainly not, gentlemen, and since we have "learnt to see" cases of this kind fresh ones are being continually found, and for my part I have very recently observed

* Barié, Joffroy, *Soc. méd. des hôp. de Paris*, séance du 14th December, 1888.

one. Besides those presented to the Société des Hôpitaux by Barié and Joffroy I can name many others: one of Marcus Gunn,* which occurred in 1883, another of A. Marina†; Charcot also presented a case to his audience; lastly, a recent thesis,‡ written under the inspiration of Mendel, has for its subject a case of this affection.

As to how this singular connection between tabes and the disease of Graves should be understood opinions differ considerably. It must be remarked, however, that nothing is settled, from a chronological point of view, between these two affections. Thus, in the case of Barié, tabes had preceded the occurrence of the symptoms of the exophthalmic goitre, whilst the contrary took place in several cases of Joffroy. Naturally the opinion of these two authors was influenced by this difference in their observations. According to Barié these are cases of tabes during the course of which lesions occurred in the medulla oblongata which produced secondarily the disease of Graves. Joffroy believes, and Ballet is of the same opinion, that there is simply a combination of two distinct nervous diseases, neither of which is in any way the cause of the other.

For my part, gentlemen, I had the opportunity of making the autopsy of the case which I had been able to observe. From a clinical point of view I can affirm positively that a typical case of the disease of Graves was combined with one of confirmed tabes; there is no doubt whatever of this fact. The microscopical examination of the cord revealed changes which were characteristic of tabes at the onset of the disease. But I have not yet been able to examine the medulla oblongata, and regret that I cannot therefore definitely settle this interesting question. At the same time, I am strongly inclined to think that it is by the influence of tabes that the disease of Graves was caused to

* Marcus Gunn.—I have not been able to find this work, which was only indicated to me secondhand. (The account of this case is found in the *Trans. Ophthalm. Soc.*, Vol. III., p. 236.—TRANSLATOR.)

† Al. Marina, *Arch. f. Psch.*, XXI.

‡ J. Weiner, *Ueber einen Fall von morbus Basedowii mit tabes incipiens. Inaugural Dissert.*, Berlin, 1891.

During the publication of these lectures another case has been published by Möbius, *Ueber die Basedow'sche Krankheit. Deutsche Zeitschrift für Nervenheilkunde*, 1891, t. I. p. 423. I have also been able to consult since writing this lecture the following work: *Art. Secchieri, communicazione fatta al congresso di medicina tenutosi in Padova nel Settembre*, 1888, in *Rivista veneta di scienze mediche*, November, 1889.

exist. Either this is simple "instigation," as, for example, in the cases in which another form of neurosis, hysteria, coincides with tabes, or perhaps the symptoms of exophthalmic goitre are directly consecutive to the tabid lesions in the medulla oblongata as Barié believes.

It must also be observed that, if the coincidence of tabes and the disease of Graves is not very frequent, symptoms are occasionally observed in connection with one of these diseases when it exists in an isolated form, which recall the other affection. Thus, in his first works upon tabes, Charcot remarked the fact that *tachycardia* was frequent, the heart being often found in tabes to contract 100 or 120 times in the minute. Kahler was able to verify this assertion, and in 50 per cent. of his cases of tabes the frequency of the pulse was found to vary from 80 to 120 times in the minute; one of the cardinal symptoms of exophthalmic goitre, *tachycardia*, is thus frequent in tabes. On the other hand, in the course of studies connected with the abortive forms of exophthalmic goitre I twice found the *patellar tendon reflex to be absent*. In these cases one of the symptoms of tabes was found to exist in the disease of Graves (possibly these were cases of unrecognized tabes, since at that time my attention was in no way directed to cases of this kind). These facts are interesting, and if this connection does not settle the question it should at any rate be noted.

Whatever explanation is given of the coincidence, it should be remembered, gentlemen, that the fact itself is indisputable, and we should be grateful to those who first recognized its existence and made it known, since every fresh discovery is a step towards the victory of scientific truth.

LECTURE XXIII.

TABES.

SYMPTOMS (*continued*).

VISCERAL DISORDERS. III.—PHARYNGO-LARYNGEAL SYSTEM.

A.—*Pharynx.* Researches of Fano. Pharyngeal crises of Oppenheim.

B.—*Larynx.* Tabid Laryngismus: (1) Acute symptoms of tabid laryngismus; laryngeal crises, their description, laryngeal ictus, nature and mode of production of laryngeal crises. (2) Chronic symptoms of tabid laryngismus, whistling inspiration, unnatural voice, having two tones. Laryngeal paralysis: the most frequent is that of the posterior crico-arytenoid muscles. Lesions of chronic laryngismus connected with the nerves and medulla oblongata.

IV.—URINARY SYSTEM.

A.—*Derangements of the urinary secretion:* glycosuria; changes in the amount of urea, phosphoric acid, &c.; excessive secretion of urine.

B.—*Derangements of the urinary excretion:* "false urinary patients" of M. Guyon; relative retention, complete retention; absolute incontinence, relative incontinence; versical colic; nephritic crises.

V.—GENERATIVE SYSTEM.

A.—*Derangements in the male:* impotence; generative excitement; cremaster reflex, its research, its signification; bulbo-cavernosus reflex; atrophy and anæsthesia of the testis.

B.—*Derangement in the female:* genital depression; genital excitement; pain in the genital organs; vulvo-vaginal crises.

GENTLEMEN,—The visceral disorders, or rather those of the internal organs, which we shall now study are those which affect the *pharyngo-laryngeal system.*

III.—THE PHARYNGO-LARYNGEAL SYSTEM.

A. PHARYNX.—The derangements of the pharynx have been recently described by several authors. Their existence at the present time, however, is far from being generally recognized, and works controlling their existence not having been written, I look upon it as my duty to at least enumerate the derangements which may exist.

In 36 patients suffering from tabes, who formed material for the researches of A. Marina to which I have already alluded,

Fano found *hyperæsthesia* or *anæsthesia of the soft palate* to exist in 30; in 14 cases the *sensibility of the pharynx was diminished*, and in 9 cases that of the *larynx*. In 4 patients this author found the *pharyngo-laryngeal reflexes to be excessive and associated with hyperæsthesia of the pharynx*. In 5 cases the *epiglottis* was much *lower* in position. It would be interesting to know whether all these changes are really due to tabes, or would be also found to occur in patients suffering from different consumptive affections, and if so with what frequency, this being a question which unfortunately I cannot answer.

On the other hand, Oppenheim* has described by the name of *pharyngeal crisis* a symptom consisting of a series of deglutition very frequently repeated (24 to 32 in the minute) and accompanied by more or less sonorous indications of its occurrence. These crises in the case observed by Oppenheim continued during from 10 minutes to half an hour with short intervals: they were accompanied by much pain, and associated with congestion of the face and profuse perspiration.

B. Larynx.—The derangements of the larynx which may occur during the course of tabes are numerous, of varying intensity, transitory or permanent, and present considerable modifications in their aspect; Charcot includes them all under the name of "tabid laryngismus," which is most convenient in practice.

1. *Acute symptoms of tabid laryngismus: Laryngeal crisis.* The first author who mentioned these was Fereol (1868). Jean specially called attention to their existence, and they were then carefully studied by Charcot and Krishaber, and it is under the influence of these two authors that the monograph of Cherchewsky was written, which presents a full account of this subject. Since that time it has been observed that these symptoms occur frequently in a more or less pronounced form, and numerous works or observations have been published about this condition.

The laryngeal crisis consists specially in *dyspnœa* of varying intensity and existing in an isolated form, or in association with fits of coughing. In the former case the respirations are short, superficial, rapid, and during their occurrence one inspiration takes place which is deeper and accompanied by a sound which

* Oppenheim, *Neue Beiträge zur Path. der Tabes dorsalis* (*Arch. f. Psych.* XX., 1888).

recalls the "kink" of whooping cough. In the latter case, a cough is associated with the dyspnœa which occurs in paroxysms presenting at times, on account of the noisy inspiration to which I have just alluded, all the characters of the cough which exists in pertussis; the cough is dry and unaccompanied by expectoration, except at times when it occurs in small quantity at the end of the paroxysms.

The *dyspnœa* which occurs in the two cases and which really constitutes the laryngeal crisis differs much in character, being sometimes slight and of short duration, whilst at other times it is pronounced, and continues for a longer time, whereas in some cases it is extremely severe, constituting the *laryngeal ictus* of Charcot. This symptom is then truly alarming, and for my part I shall never forget the conditions in which I witnessed it for the first time. It was in 1879, during the first year of my being house physician at the Salpêtrière Hospital, and I was in the room of the superintendent of "incurable patients," when a female patient entered to ask a question of my esteemed friend and colleague Brissaud. The patient was slightly out of breath whilst she was speaking, each inspiration being accompanied by a pronounced whistling sound; in my mind I made the diagnosis of "compression of the recurrent nerve" by some mediastinal tumour; when suddenly, whilst Brissaud was explaining matters, the patiently fell violently to the ground in an unconscious condition, making convulsive movements and with the face of a purple colour. I feared that the patient was about to die suddenly, and undoubtedly betrayed my emotion, since Brissaud hastened to reassure me and explained that this patient was liable to attacks of *laryngeal ictus*.

This was an attack of such a nature, and the patient was one of those of whom Charcot made use when describing this complication. The following, according to this author, are the characters of the *laryngeal ictus:* usually a burning sensation is first felt in the region of the larynx, representing a form of aura, which is followed by a dry cough occurring in short hacks, the patient then falls to the ground in a state of unconsciousness, and most often general flaccidity, whilst epileptiform convulsions sometimes occur, causing the attack at times to resemble those which occur in epilepsy; the face is usually livid. The laryngeal ictus generally lasts but a few seconds, at the end of which

the patient rises of his own accord and immediately regains his senses without the occurrence of any condition resembling the "confusion," which is liable to occur after an attack of true epilepsy.

You have seen, gentlemen, how severe the attacks of dyspnœa may be in the laryngeal crisis. Fortunately this is not always the case, and the symptoms may be limited to those which I first described. It must also be stated that the duration of these crises varies from a few minutes to several hours. However severe the attacks may be they usually end favourably, the dyspnœa gradually diminishes and then ceases to exist, sometimes, however, in a much more abrupt manner. It must not, however, be supposed that the crises always terminate in this favourable manner. In some cases death has been known to occur within a short time. This fact, gentlemen, should not be forgotten, and if the complication becomes really serious preparations should be made which enable the operation of tracheotomy to be performed if necessary.

As regards the frequency and course of laryngeal crises it is difficult to make any precise statement, on account of the variations which occur; frequent in some patients, they occur seldom in others; constantly increasing in frequence and severity in some patients they diminish in others, and are even found at times to cease altogether.

Nor are authors agreed as to the *pathology* of this symptom. Some believe it to be due to paralysis of the muscles which dilate the vocal chords. On the other hand, Charcot and Krishaber have shown that the crises are due to reflex contracture of the muscles adjoining the glottis, a contracture which is due to hyperæsthesia of the mucous membrane, these authors having been able to reproduce it by touching that mucous membrane with a soft body. This explains the fact of these crises being at times produced by a current of air, by the contact of a cold body, by the act of speaking, by that of walking quickly, &c. Oppenheim has recently indicated another means of artificially producing the laryngeal crises. This consists of pressing upon a sensitive point situated between the sterno-mastoid muscle and larynx, at the level of the cricoid cartilage. I cannot give any personal opinion upon the value of this mode of procedure, which I have never adopted, but according

to Fano, who attempted to verify it in 36 patients suffering from tabes, and whom he examined in regard to the laryngeal symptoms, this sensitive point was only found to exist in one case, and this was in a patient who was not subject to laryngeal crises.

The supposition of muscular spasm, due to hyperæsthesia of the mucous membrane, accords also with the favourable effect which is produced by anti-spasmodic remedies (ether, chloroform, cocaine, and bromides), and upon the duration and intensity of the laryngeal crises.

2. *Chronic symptoms of tabid laryngismus.* In some patients true laryngeal crises do not occur, but only a sensation of pricking and uneasiness in the region of the larynx. The breathing of these patients is often more or less permanently *whistling* in character, and the voice often becomes unnatural and *of two different tones.* At times there is also slight dyspnœa, the inspirations at least being difficult and painful, while the cough, should one exist, may be muffled, unnatural, and accompanied by a tendency to eructation, the symptoms, in fact, of *laryngeal paralysis.*

Laryngeal paralysis is comparatively frequent in the course of tabes, and deserves brief consideration. The *posterior crico-arytenoid* muscles are most often affected, which, as you know, dilate the glottis, and are therefore essentially respiratory muscles. They are not infrequently paralysed on both sides, and according to some authors this condition is essential for the production of the whistling breath sounds, due to the fact that abduction of the vocal cords is impossible; this symptom would not result from unilateral paralysis of the muscle.

The posterior crico-arytenoid muscle being, as has just been observed, a muscle connected with respiration and not with speech, you will easily understand, gentlemen, that its paralysis does not produce loss of voice, though it probably contributes to make it unnatural, and to produce the double tone to which allusion has been made.

Other laryngeal muscles may be also paralysed during the course of tabes, as for example, the *thyro-arytenoid* or *lateral crico-arytenoid*, but these muscles are paralysed less often, and to a slighter degree than the posterior crico-arytenoid muscles.

The coincidence of the laryngeal crises with paralysis of one or more laryngeal muscles is not quite constant, though frequently observed.

As regards the *ataxy* of the vocal cords mentioned by some authors, it seems to be rare, and not to have been very clearly observed.

The *lesions* to which these different symptoms are due will now be considered.

As regards the *muscles*, secondary lesions exist in them which

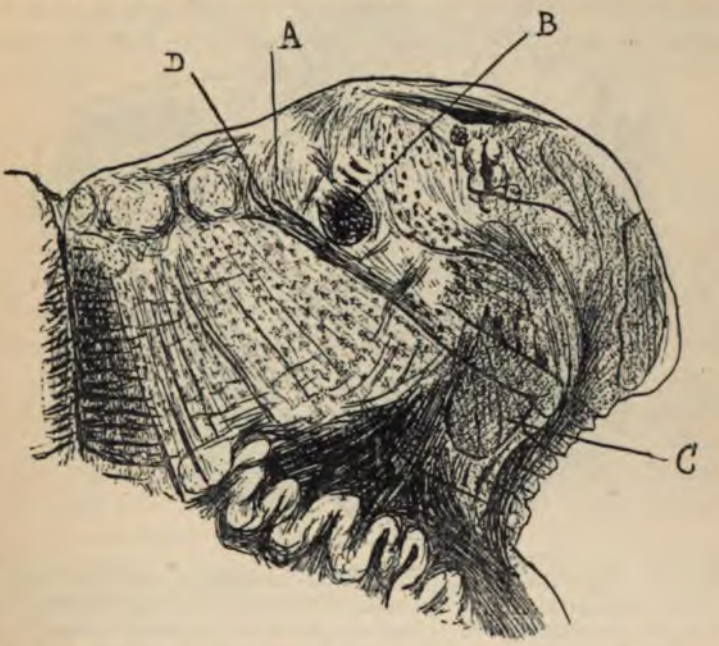


Fig. 156.—Right half of a section of the medulla oblongata in a *sound* condition. The upper transverse line represents the floor of the 4th ventricle; the vertical line which limits the figure on the left side is the septum. A, nucleus of the pneumogastric nerve; B, ascending root of the glosso-pharyngeal nerve, in which are fibres of the pneumogastric and spinal accessory nerves—the "slender column"; C, ascending root of the 5th pair of nerves; D, root fibres of the pneumogastric nerve. (After Oppenheim.)

differ in no material degree from those already studied in connection with the muscular atrophy which occurs in tabes.

In the *nerves*, changes have been found to exist in certain nerve trunks, either in an isolated condition, or conjointly with

lesions of their *roots* or their *nuclei* themselves in the *medulla oblongata*. These nerve trunks are the vagus, glosso-pharyngeal, and spinal accessory; Oppenheim has also been able to show that the electrical irritability of the *recurrent* nerve is lost.

In the *medulla oblongata*, atrophy of the *nuclei* of these same nerves (the vagus, glosso-pharyngeal, and spinal accessory) has been mentioned by a somewhat large number of authors (Jean,

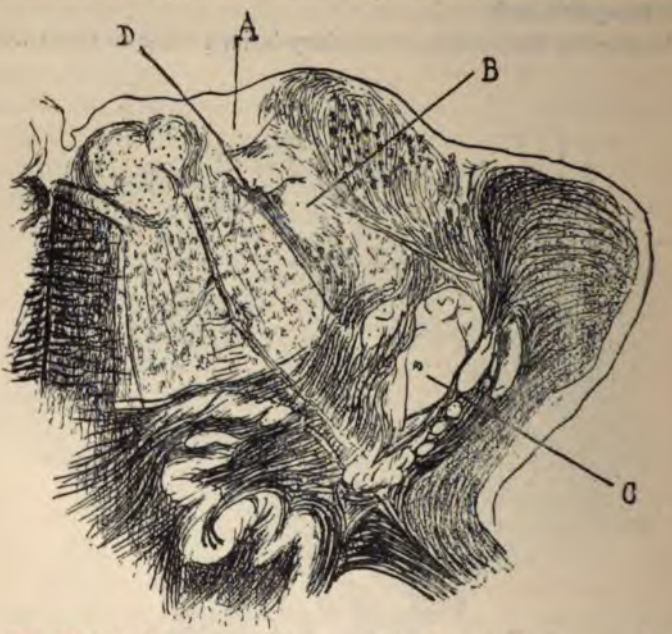


Fig. 157.—Right half of a section of the medulla oblongata, from a case of tabes with laryngeal crises. (To be compared with the preceding figure.) A, nucleus of the pneumogastric nerve, which is altered and no longer contains nerve fibres; B, ascending root of the glosso-pharyngeal nerve, in which are fibres of the pneumogastric and spinal accessory nerves—the "slender column"; this tract is very much altered, its nerve fibres having completely disappeared; C, ascending root of the 5th pair of nerves which is affected and of a white colour; D, the root fibres of the pneumogastric nerve have almost disappeared. (After Oppenheim.)

Demange, Landouzy and Dejerine, J. Ross, Kahler and Oppenheim).

To these facts I must add those which I have already mentioned, in which hemiatrophy of the tongue coincides with laryngeal paralysis, or rather with atrophy of the vocal cord on the same side. In these cases the laryngeal symptom would, in

the opinion of P. D. Koch and myself, be solely due to the lesion in the nucleus of the hypoglossal nerve, and the groups of cells connected with it.

Lastly, another lesion has been mentioned, and in a special way by Oppenheim, namely, that which occurs in the *ascending root of the lateral mixed system*, or *slender column*.

These different lesions may either occur conjointly, or be isolated from each other; in some cases the nuclei, in others the nerve roots, in others again the peripheral nerves are affected; in this case, as when muscular atrophy of the limbs exists, we meet with the same irregularity, the same opposite conditions.

IV.—Urinary System.

A. *Derangements of the urinary secretion.* These may be qualitative or quantitative.

Amongst the former I must mention *glycosuria*,* which is perhaps due to the existence of tabid lesions in the floor of the 4th ventricle. Other derangements are undoubtedly due to the consumptive tendency, of the existence of which in these patients I have spoken more than once. Such are a *diminution in the amount of urea*, and in the total quantity of *phosphoric acid*, with a proportionate increase in the earthy phosphates. Lastly there is a great variety in the quantity of chlorine eliminated, with a tendency to the production of hypochlorites, after the researches of Livon and Alezais.

Albert Robin (verbal communication) has remarked that in certain patients suffering from tabes the quantity of *phosphorus* eliminated by the urine in an *incompletely oxidised state* is more considerable in the early morning than at bedtime. This difference would be due to the nervous irritation produced by the lightning pains during the day, and the sedative influence which is due to sleep at night.

As regards the quantitative changes, the excessive secretion of urine (many pints in the 24 hours) must be mentioned, which occurs in certain patients, and often in a paroxysmal manner,

* G. Guinon and Souques, in the *Archives de Neurologie*, 1891, maintain the interesting opinion that in some patients suffering from tabes glycosuria might be the expression of a hereditary diathesis, since as Charcot has shown, tabes is frequently met with in families which suffer from diabetes.

giving rise to the term "urinary crisis," which was proposed by M. Féré for cases of this nature. This is one of those fluxes (gastrorrhœa, sialorrhœa, persistent diarrhœa, &c.) which are far from rare in tabes.

B. *Derangements of urinary excretion.* These are extremely frequent, and may occur in the earliest period of the disease, a fact which, from their being conjoined with other special symptoms, enables them to be classed amongst the best signs of tabes at the onset.

They constitute at times the first indication of tabes, and the patients naturally almost always consult a surgeon who specially treats affections of the urinary passages. A local examination is made, and, as would be expected, no lesion of the urethra, prostate, or bladder is detected, to which the disorder from which the patient suffers might be due. This is the reason why Guyon very rightly terms these persons "false urinary patients."

When tabes is suspected the patient should always be asked whether he has remarked anything unusual to happen when he passes urine. Sometimes, specially in the case of those who observe but little, the patients will say that they have not remarked anything abnormal. You must not, however, be satisfied with this reply, but put the definite question, "Are you obliged to wait, or to strain in order to pass urine?" This time the patient, who now understands the question, will give an affirmative answer, a fact which I can guarantee before hand, and often he will add characteristic details.

The fact is, that though the bladder may not be paralysed at the onset of tabes, the control over that organ is partially lost; it is, as it were, an instrument upon which the patients cannot play with the necessary precision; instead of passing water freely they are obliged, so to speak, to "pump," to *strain* with all their might, and before the first drop of urine is seen to appear they have to *wait*, not a few seconds only, but one or two minutes, or even more. As Fournier says, "these patients only pass water in several acts," and I would add that the intervals between the acts are often so long that the spectators, should there be any, are unable to wait. Thus Fournier gives the history of many of his patients, who dared not pass water in the public urinoirs, innumerable quarrels arising with those who were about to use them, and who, desiring to follow in their

turn, were provoked on account of the long time which passed before the place was unoccupied.

Complete retention of urine sometimes occurs, though it is rare, and most often of temporary duration; I have however known it require the daily introduction of a catheter.

Absolute incontinence of urine may also be observed, specially in the advanced period of the disease, but is also rare; when it occurs it is often like retention of temporary duration. On the other hand *relative incontinence* is frequent; under the influence of emotion, effort, or even during sleep, and without apparent cause, a few drops of urine dribble upon the shirt or into the trousers, the patient in some cases, on account of more or less anæsthesia of the mucous membrane of the urethra, only perceiving that this accident has happened owing to the sensation of moisture which is thus produced.

In some cases micturition is abnormally frequent, and "pollakiuria" exists, or, on the contrary, it may be infrequent, so infrequent in fact that some patients suffering from tabes no longer feel the desire to urinate, and only pass water, as Fournier says, from "a sense of duty."

I have just spoken, gentlemen, of the *anæsthesia* which sometimes exists in the mucous membrane of the urethra, and probably also in that of the bladder; it must not be supposed that this excludes the occurrence of pain in these regions. Pain, in fact, is observed to occur either of a lightning character, or of the persistent type (sensation of a foreign body); these are seated either in the urethra, or at the neck of the bladder, or in the bladder itself, and in certain patients, according to Fournier, true *vesical colic* may exist. This colic according to that author may be compared with the gastric "colic" or crisis. Lastly some authors have mentioned cases in which the character and seat of the pain were very analogous to that which occurs in nephritic colic, which explains the name of nephritic crises which has been applied to them.

V.—Generative System.

The derangements of this system should be studied separately in the two sexes, not because there is any essential difference, but on account of the special rôle of the male or female sex in the sexual act causing them to have peculiar characters.

a. In the MALE.—It is in the male sex, as you will easily understand (the part taken being more active, the investigation more easy), that the derangements of the generative system have been specially observed.

a. Impotence. This is the most frequent symptom of tabes connected with the generative organs. At first this impotence is but relative, *erection* occurs but slowly, and the patient confesses, while shaking the head with an air of resignation, that "he has lost much power in that respect," and when erection occurs it has an unsatisfactory character, and should it by chance be sufficiently pronounced it is the *ejaculation* which is but slowly produced should it fortunately occur at all. This disorder gradually, or at times rapidly, increases until absolute impotence exists. Fortunately for these unhappy persons *sexual inappetency* increases in a corresponding manner. A patient suffering from tabes who was quite impotent observed to me once at the hospital, "When, sir, hunger no longer exists it is of little consequence that the teeth are lost." Those suffering from tabes are "no longer hungry." It is on this account, as I said just now, that these sufferers from impotence are resigned, in which they differ from many affected by disease of the nervous system who, though impotent, are starving.

This, however, is not always the case. In some cases the onset of tabes is marked by the opposite condition, and the period of impotence is preceded by one of excitement.

b. Genital excitement. This, which is a rare condition, may affect the different stages of the sexual act at the same time (the desire, the erection, the ejaculation), and the patients in whom it occurs feel sometimes flattered on account of their power, but more often somewhat alarmed, and not unreasonably so. Usually, however, this excitement barely exists except in appearance, and is a false indication; the frequent persistent erection amounting in some cases to real priapism is often unassociated with any desire and followed by no ejaculation, or perhaps the ejaculation is premature, occurring at the first contact and producing no pleasure, but sometimes, in fact, actual pain. In some patients pollution simply occurs, and Fournier, who has known the ejaculations to occur in groups at intervals of time more or less distant from each other, suggests that they are really "crises"

more or less analogous to the different visceral crises with which we are already acquainted.

So far all the information which it is necessary to have with respect to the different genital derangements must be obtained from the patients, who should be questioned, their answers being then believed to be true. We have, however, the means of controlling their statements up to a certain point, and at least of knowing whether the patients are, or are not, impotent. These means are furnished to us by the examination of the different reflexes connected with the genital organs.

1. *Cremasteric reflex.* No practical examination being usually made with regard to the existence of this reflex, at any rate in France, it would be useful, I think, at any rate to some of you, gentlemen, if I indicated the modus faciendi. One of the two persons whom I now place before you is in good health, while the other is a tabid patient suffering from impotence; the condition of the reflex will be examined in each of the two patients. I have caused the pubes of the healthy man to be exposed, and the thighs as far as the knees in order that you may see the scrotum clearly, as it is this part which you must closely watch, with the object of observing what takes place.

I rapidly rub the inner surface of the thigh from above downwards by means of the nail or some soft body, gentle pressure being made upon the skin; this is only done once, and the immediate effect is that the testis upon the same side rises towards the inguinal canal; as often as I repeat this action the same elevation of the testis occurs; this is what is termed the *cremasteric* reflex. We will now examine the condition of the same reflex in the tabid patient suffering from impotence; I may perform the same action again and again along the inner surface of both thighs without producing the slightest result; no movement in the testis occurs, and the cremasteric reflex is completely absent. There is in fact, gentlemen, an intimate, I dare not say absolutely constant connection (these facts not having as yet been so thoroughly tested as one would wish) between the absence of cremasteric reflex and impotence, when the latter is due to an organic lesion of the nervous system, as in tabes; on the other hand, when impotence exists in other nervous diseases and is not due to an organic lesion of the nerve centres, the

cremasteric reflex persists, and it may even be hoped that the impotence will subsequently cease to exist.

There is another mode of testing the cremasteric reflex, with which I should make you acquainted; instead of the inner surface of the thigh being rubbed, somewhat strong pressure need alone be made with the thumb upon the inner surface of the thigh opposite the opening which exists in the adductor magnus muscle for the passage of the femoral vessels; the same result occurs. Both these means of producing cremasteric reflex should be tried before it is pronounced to be absent. Lastly, I would put you on your guard against the error which consists in mistaking contraction of the dartos for that of the cremaster muscle. This contraction of the dartos, being due to the contact of the air when the part is uncovered, has no connection with the cremasteric reflex; the fact that it is not seated in the testis in the same way as the latter reflex, but generally occupying the whole scrotum enables it to be distinguished; again, the movement which is produced is slow and vermicular, whereas the elevation of the testis in the cremasteric reflex is on the contrary abrupt and rapid.

2. *Bulbo-cavernosus reflex.* This reflex has been recently described by Onanoff,* who also mentioned the changes to which it is liable in tabes; these and the mode of investigating them are in the words of Onanoff as follows:—"The index of the left hand being placed over the region of the bulb of the urethra, the right hand rapidly rubs the dorsal surface of the glans with the edge of a small piece of paper, or again, the mucous membrane is slightly pinched by it; in these conditions the index which is applied to the bulb feels the movement to occur, which is due to contraction of the ischio- and bulbo-cavernosus muscles." This reflex which constantly exists in healthy persons, and even in those suffering from nervous diseases, in which the genital functions are unaffected, ceases to occur in some cases of tabes; in these complete erection no longer occurs according to Onanoff. This author is the first to admit that this statement is liable to some modifications, his researches only having been made as yet in cases of tabes.

3. Atrophy and anæsthesia of the testis. These two symptoms

* Onanoff, Du réflexe "bulbo-caverneux," *Société du biologie*, séance of May 9.

have been mentioned and studied by Pitres and Rivière.* The atrophy of the testis was found by them to exist in 3 out of 20 cases; with regard to the anæsthesia of this organ when exposed to pressure they found it to exist in 16 out of 20 cases; this form of anæsthesia may occur in a very pronounced degree.

These, gentlemen, are symptoms of tabes connected with the genital organs, of which there is direct evidence, and which give valuable information as to the condition of their functions in the male sex.

β. In the FEMALE. In the female sex, as I have already observed, the derangements connected with the genital organs are almost analogous to those occurring in males, but the difference of the organs causes them to have special characters.

(*a*) *Genital depression.* This may exist in a greater or less degree; the previous disposition of the patient must naturally be considered, a somewhat large number of women in a healthy condition having no sexual desire whatever. In females suffering from tabes this desire may either be diminished or perhaps completely lost, as in the male sex. In this case again the symptom is at times preceded by indications of genital excitement.

(*b*) *Genital excitement.* This sometimes occurs to a very pronounced degree, as for example, when the condition occurs which is termed by Pitres† the *clitoridean crisis.* The best means of making you understand what occurs in these crises is for me to quote verbally one of the observations of Pitres: "The voluptuous sensations occurred, according to the account of this woman, at any moment in the day, specially when she was inactive, and without being preceded by any provoking cause or lascivious thought. It began by a sensation of vibration within the vagina. The sensation extended to the clitoris, which entered into a state of erection, this being soon followed by a true erotic spasm accompanied by ejaculation, in precisely the same way as during ordinary sexual connection. This spontaneous crisis was almost always repeated three or four times during the same day, after which one or two weeks

* Rivière, De l'anesthésie et de l'atrophie testiculaires dans l'ataxie locomotrice, *Thèse de Bordeaux*, 1883.

† Pitres, Des crises clitoridiennes au début ou dans le cours de l'ataxie locomotrice. *Progrès médical*, 1884, No. 37, p. 729.

passed without its returning. It was followed by a painful sensation of languor affecting the stomach. It should be noted that at this time the patient was living with her husband, with whom sexual connection occurred to a full though not immoderate extent." The clitoridean crises therefore, as this example shows, consist merely of pollution, accompanied by the ordinary voluptuous sensations. The singular point about them is that they occur spontaneously, and usually in paroxysms. They are quite unaccompanied by pain, which is not the case with regard to the other variety of crisis which I have still to mention.

(*c*) *Pains in the genital organs.* I have already had occasion to mention the occurrence of pain in the male sex, and have alluded to the painful ejaculation, the sensation of a foreign body in the urethra or bladder, &c. In the female tabes may also be accompanied by pain in the genital organs, which at times are more or less analogous to those just mentioned. In other cases they are of a special character, having been described by Morselli* by the name of *vulvo-vaginal crises*. In these there is no erotic element, as in the clitoridean crises, the patients experiencing severe pain, which seems due to a very painful spasm of the constrictor vaginæ muscle. Thus Morselli compares the vulvo-vaginal crises with those connected with the larynx, in which, as we have seen, the spasmodic element is found to preponderate.

Such, gentlemen, is an enumeration of the principal derangements of the genital function, which are observed during the course of tabes in the male and female sex. Before this chapter is ended it should be mentioned that these symptoms vary to a considerable degree in different subjects, although some resemblance always exists between them. It should not be forgotten again that these symptoms often occur at quite an early stage of the disease, belonging to the præataxic period, during which we have seen so many tabid symptoms occur, and which in the opinion of every observant medical practitioner may be regarded as really a stage of confirmed tabes.

* Morselli, *Sulle crisi vulvo-vaginali nell' attasia locomotrice. Giornale di Neuropatologia*, 1890, March—June, p. 117.

LECTURE XXIV.

TABES.

SYMPTOMS (*continued*).

VI. CEREBRAL SYSTEM.—Hemiplegia. Apoplectiform attacks. Epileptiform attacks. Acute symptoms connected with the medulla oblongata. Psychical derangements. Coincidence of general paralysis of the insane.

COURSE OF TABES.—Prodromal period. Second period, period of inco-ordination. Third period, confinement to bed. Forms of tabes: upper or cervical, cerebral, mild, and severe forms.

VI.—CEREBRAL SYSTEM.

The *hemiplegia* which somewhat frequently occurs in the course of tabes will not be again considered, this complication having been discussed with the other motor disorders. The cerebral disorders which will be now discussed are the *apoplectiform* or *epileptiform attacks* which sometimes occur, and the *psychical derangements*. I would also remind you that many authors (Berger, &c.) have observed *migraine* to indicate, so to speak, the onset in certain cases of tabes.

The *apoplectiform attacks* in tabes have been specially studied and described by Lecoq and Giraudeau. Kahler has also considered these complications, and terms them collectively cerebral tabes; the symptoms vary from simple vertigo to complete loss of consciousness, and may be accompanied by aphasia, or, as we have said, by hemiplegia of the face or limbs.

The epileptiform attacks may be of the severe or mild form, and Jacksonian epilepsy at times occurs.

By what mechanism are these complications produced? It is probably multiple; in some cases foci of real hæmorrhage or softening exist, or some morbid process of long duration in the meninges or ependyma, or vasomotor disorders due to some change in the medulla oblongata or pons varolii. My opinion, however, is that such lesions should not be considered due to the existence of tabid lesions alone, and it seems far more

probable that they are connected with syphilis, which, as you will soon see, is the origin of tabes.

It seems that the *acute symptoms connected with the medulla oblongata*, mentioned by Hanot and Joffroy* as occurring at the onset of tabes, may, at least as far as their clinical aspect is concerned, be compared with those which precede.

The *psychical derangements* which occur in the course of tabes are not infrequent but vary considerably, and Dieulafoy was able to apply the term "tabid insanity" to the condition which existed in some patients; while others present similar symptoms which are but slightly pronounced, and may be either transitory or permanent, or possibly occur "in paroxysms" (Fournier). I should not like to exaggerate what happens, and on that account it is, I think, necessary to distinguish the moral from the intellectual derangements; the former are, I must allow, frequent, I would almost say constant, but whether they are due to the existence of tabes is doubtful. Is it not rather the consumptive tendency of this affection, the weak condition in which the patients are placed, which are the cause of this change in their character? However this may be some apathy, some indifference, are usually found to exist, which in certain exceptional cases may reach a condition of melancholic stupor, or more or less pronounced irritability. Notwithstanding this tendency, and the agonizing pains which the patients frequently feel, they are rarely found to commit suicide, whilst on the contrary, in certain affections, notably those of the bladder, suicide is relatively more common; this is an interesting point in connection with the psychology of the tabid patient. As regards the intellectual disorders, these, gentlemen, are far less frequent, and if truly pronounced depend upon the association of cerebral lesions with those of the spinal cord; tabes is in that case most often found to be complicated by *general paralysis of the insane*. This, gentlemen, is a real combination, and in most cases intellectual disorders, if at all pronounced in a tabid patient, should be referred to this complication. Such cases, however, are somewhat rare, I would almost say exceptional; on the contrary the mental condition of the tabid patient is usually found to be good, as good in fact as before the onset of the

* Hanot and Joffroy. Des accidents bulbaires aigus au début du tabes. *Congrès d'Alger*, 1881.

disease, whatever some authors may say to the contrary. Consider facts which have occurred, gentlemen, and decide this question for yourselves; there is no liberal profession to which tabid patients are not known to belong in some numbers, and to be in full possession of their intellectual powers: the one is a famed musician, the other a learned scholar, the third a statesman, the fourth a man of business, I mean with the management of important matters. In mentioning but those who are dead: the artist Manet suffered from tabes, as did Henri Heine, and remember, gentlemen, that at Bapaume Faidherbe already did so. After this enumeration, truly martyrological, it must be allowed, I think, that intellectual disorders are not usually associated with locomotor ataxy.

Course and forms of the Disease.

I have been careful, gentlemen, when speaking of a symptom, to indicate as far as possible the period of the disease in which it occurs; and I have no wish to repeat what has already been said; at the same time we must necessarily consider, from a retrospective point of view, how the principal symptoms occur, that is to say, what is the COURSE of the disease.

In ordinary cases the progress of the disease is very slow, and we are more or less justified in distinguishing certain periods: the prodromic period, the ataxic period, the period of confinement, and that of paralysis. These stages, gentlemen, must not be considered an article of faith, their real merit being that of being classical. In reality this is not of great importance, and is often in complete disaccord with the facts of the case, at the same time it somewhat facilitates the description of the disease, and on that account I shall ask for permission to adopt it.

Prodromic period.—This is pre-eminently the time at which the lightning pains occur, which are most often referred by the patient to some totally different cause, and he voluntarily terms them "rheumatic." Thus reassured as to their nature he troubles himself but little about their existence, except at the time when they occur, and does not consult a medical man unless the pain is very severe. It is thus but rarely that the occurrence of these lightning pains enables the existence of tabes to be recognized. In most cases something more is required, some things

which has more effect upon the patient's mind. This is usually either the onset of some form of ocular paralysis, or one of the numerous derangements affecting the internal organs, of which I spoke in my previous lectures, such as an altered character of the micturition or genital functions, or perhaps the larnygeal or gastric crisis, &c., or lastly some difficulty in moving the lower extremities.

This, it must be understood, is not true, confirmed ataxia, being an indication which an observant patient alone remarks. Certain movements are not performed with the necessary precision; thus, for example, they find it difficult to descend a staircase, and can no longer walk at night owing to the darkness, being also unable to indulge in certain exercises, such as riding. With these exceptions the gait in ordinary circumstances presents nothing abnormal, and an experienced eye can alone recognize that a certain amount of hesitation exists at times when the patient is obliged to turn rapidly round.

No considearable change occurs in the symptoms during a time which may be of some length. In some cases in fact the disease makes no further progress, it is to these latter cases that the name of *mild tabes* is given. Then at the end of 2, 3, 5 years or more the symptoms become progressively aggravated, or ataxy of the movements suddenly occurs; this is the second period. Sometimes in the cases of so-called *acute tabes*, this second period may occur almost immediately after the onset of the affection, it is thus impossible to make any definite statement as regards the duration of the *first period*.

In the *second period* the ataxy of movement is almost always confined at first to the lower limbs, and it is only at a later date as at the end of several years that it extends progressively to the upper extremities, which in fact does not invariably occur. It must be understood that numerous other tabid symptoms coincide with these during the period of motor ataxy, and the most different visceral symptoms as well as the ocular affections which we have already considered may be specially observed at this time. In some cases this period brings the patient by slow or rapid aggravation of the symptoms to the third stage, while in others it perpetuates itself, so to speak, and the patient remains in almost a stationary condition.

The *third period* is constituted by the almost absolute loss of

control over the movements of the lower limbs. The patient is then obliged to remain either in bed or upon a chair; he has become totally powerless. To this should be added an aggravation of the urinary disorders, the existence of cystitis with pyuria, at times sloughing of the skin in the gluteal region or heel, a more or less pronounced change in the general nutrition of the patient, which also diminishes his power of resisting the different sources of infection (pneumonia, erysipelas, phthisis) which may occur, and you will at once understand to what great dangers the patients suffering from tabes are exposed. There are some patients, however, who can still withstand the effects of the disease during many years, however bad the conditions may be in which they are placed. Thus, the duration of tabes with its three classical periods may certainly be 10, 15, 20 years, or even more. For a still stronger reason the duration may in some cases be longer, viz., when the first or second period of the disease alone exist, since the causes of death are notably fewer at these times than in the third period.

A few words should be also said, gentlemen, about the different *forms* of tabes. Of these forms some are connected with the seat of the morbid process, others with its mode of development. Although the abnormal cannot be separated from the ordinary form by any definite character it is obviously better to distinguish them from a clinical point of view. The principal forms will alone be mentioned by me.

The *superior* or *cervical* form of tabes is characterized by its onset, which consists of pains in the upper extremities; the lower limbs are but little affected and ataxy rarely exists, or if so, is but little pronounced; in this form muscular atrophy often occurs (Leyden). As to the persistence of the patellar tendon reflex, Weir-Mitchell, Martins, Bernhardt, Eichhorst, who have published cases of this form, are far from giving the same account of what occurred, since according to some the patellar tendon reflex existed, while according to others it was absent; thus it is difficult to make a definite statement with regard to this fact. In my opinion the persistence of this reflex does not seem to me at all impossible. In every case you must be very careful, gentlemen, to avoid the mistake which consists in regarding cases which belong exclusively to syringo-myelia as examples of cervical tabes.

The *tabes with cerebral form* of some authors is that in which the symptoms connected with the cranial nerves predominate, without the existence of psychical derangement being much more frequent than in the other forms. All authors, however, do not admit that the special characters of this form are sufficiently decided to justify its separation. It must at the same time be acknowledged that the cases of tabes with atrophy of the optic nerve and persistence of the patellar tendon reflex have a very special character.

The *mild form* of tabes (Charcot) is that in which the symptoms are not very pronounced, and either remain stationary or improve. In these patients the lightning pains are but slight, the loss of the knee-jerk, the inactivity of the iris-reflex, and the symptoms of Romberg are but little pronounced. These would be almost the only indications of tabes, and the symptoms far from increasing as time progresses, remain of almost precisely the same severity during 8, 10 years, or more. It is evident that in these cases, which, however, are not very frequent, the morbid process has ceased to *increase*, and the patient may perhaps be said to recover; as regards, however, recovery consisting in "restitutio in integrum," I must confess, gentlemen, that this seems to me impossible, on account of the destructive degeneration which has occurred in the spinal cord.

Some authors have thought it right to connect certain symptoms with the prognosis of the disease. M. Remak contended that when the pain is very intense in tabes, "tabes dolorosa," the subsequent course of the disease is seldom very severe. This fact may be true in some cases, but cannot certainly be applied to every patient. Other authors, and of this I have already spoken, while discussing the ocular disorders which occur in tabes, assert that the patients suffering from tabes, in whom the symptoms of optic atrophy occur early in the disease, may be assured that the affection of the spinal cord will not be severe. This observation has some truth in it, but the theory which these authors deduce from it must be accepted with some reserve, as I have already explained to you.

The *severe forms* of the disease are far from being rare, this being specially the case as they may be produced by morbid processes of different kinds.

In some cases they are severe on account of the *intensity* of

the tabid symptoms, and of their generalization; in some cases the patients seem to suffer from every symptom which can occur during the course of this affection. These were the cases described by Duchenne of Boulogne; they are truly patients who are most suitable for the study of the disease, but at the same time they deserve our fullest sympathy, since they may be considered the most unhappy of men.

In other cases the form is severe on account of the *rapid* course of the disease. These cases have been already mentioned under the name of *acute tabes*. Within some months pronounced ataxy of movement occurs in these patients or they may be confined to bed after a short lapse of time.

It is sometimes the *consumptive tendency* which develops to an unusual degree and constitutes the danger; the patients are then found to be extremely emaciated, with hollow eyes, pinched features, and an earthy complexion; should an epidemic occur, or the patients be placed in contact with the tubercle-bacilli (which is not infrequent in the wards of our hospitals) they are already destined to be victims of this disease.

Lastly, the severity of the disease may be due to the coincidence of *general paralysis of the insane* with tabes, of which you already know; the head and lower limbs then suffer simultaneously, the unfortunate patient is entirely helpless, and death inevitably occurs.

LECTURE XXV.

TABES.

ÆTIOLOGY.

ÆTIOLOGY.—Common causes: *Wet and cold; Diatheses: arthritic, herpetic; sexual excess; injury.* The true ætiological element of tabes is *syphilis.* The discovery of this fact is due to Fournier (1876). Erb (1879) adopts and defends this opinion. Statistics published by different authors as to the percentage of tabid patients known to be affected by syphilis. Recent statistics of Erb including 360 cases giving 89 per cent. of syphilis in the ætiology of tabes. Nine tenths at least of tabid patients suffer therefore from syphilis.—Increased number of adversaries to this opinion.—Reputation of their arguments.—Influence of *hereditary pre-disposition to nervous diseases* shown by Charcot. Doubtful effect of *hereditary syphilis. Age:* greatest frequency of onset between 30 and 45 years. *Race. Profession:* most often occurs in the liberal professions; its infrequency in the clerical profession.

GENTLEMEN,—The knowledge of tabes from a clinical point of view, and of its pathological anatomy will not enable an exact idea to be formed as to its nature; the study of the ÆTIOLOGY will alone enable this to be done. As you will see, I do not allude to the commonly given ætiology, far from it. There are certainly authors who have invoked, in connection with the ætiology of tabes, a long list of possible causes; allow me, gentlemen, to pass rapidly over these and only to dwell upon the opinions which appear to me to be of some interest.

I shall therefore say nothing more about the effect of wet and cold, the arthritic or even the herpetic diathesis.

Sexual excess is generally believed to play an important rôle in the ætiology of the disease; the reason of this is that tabes is ordinarily considered to be a "consumption of the spinal cord"; while again genital derangements are frequent in this affection, so that it is easily understood that popular malice has taken advantage of this fact to apply the aphorism "punishment affects the parts by which an error has been committed." In my opinion, gentlemen, the mistake and effects of sexual

excess have been much exaggerated, as also of onanism, sexual connection in an upright position, or too frequent venereal congress. That such excess may have the effect of producing such pronounced organic lesions of the nervous system is a fact which I absolutely deny. The only concession which I can make is by allowing that a condition of more or less accentuated nervous instability may be thus produced, and the more so since it is only those whose nervous system is in a weak condition who give way, at any rate habitually, to such sexual excess as to suffer from disease. In this opinion, however, namely that tabes may be produced by sexual excess, as in all popular beliefs, there is some basis of truth. It is a fact that a large number of tabid patients have been somewhat profligate. At the same time, by the simple application of the law of probabilities, they have also been more liable than others to contract syphilis, as I shall presently be able to show you. "Tabes venerea" is possibly the cause, but upon the condition that a larger place is made for an impure than for an immodest Venus.

The effect of *injury* has been invoked by many authors, amongst whom I would name Verneuil, Spillmann, Parisot, and Klemperer, who has specially studied this mode of causation; traumatic causes in general are considered in this case. Guelliot, Bernhardt, and some other authors have specially, and typically, incriminated the trepidation produced in the whole body by the use of the sewing machine. This, however, but little accords with the greater frequency of tabes in the male sex, and to me this mechanical action seems of too slight a nature to produce such considerable lesions as those which occur in tabes. I should be more inclined to attribute the origin of the disease in these cases to the fact that those who work the machines are not usually of spotless virtue. It is by no means impossible that the patients whose history has been recorded were simply suffering from syphilis.

With much the same idea as to the causes of the disease Hoffmann states that he has seen tabes occur in a nail-making workman who worked upon a machine, where he was exposed to somewhat violent shocks varying in number from 6,000 to 10,000 daily; symptoms of tabes occurred after 3 months of this work. In this case also, gentlemen, I can scarcely believe that these shocks were the real cause of the disease.

I should be more disposed to agree with the theory propounded by Straus, according to which certain facts, without proving the absolute connection of cause and effect between injury and tabes, at the same time show that traumatic injury may exercise some influence on the seat of the first tabid symptoms (the lightning pains first occurring in the limb which has been injured). It is also possible that injury to the sacral region may in certain cases hasten the occurrence of impotence in this disease.

For my part I have had the opportunity of seeing patients suffering from tabes who date the commencement of their disease from the time of a fall. One, for example, whilst hunting fell into a ditch, from that moment he had difficulty in walking, and ataxy of movement rapidly occurred. In another patient the first symptoms of tabes occurred after a fracture of the leg. By careful inquiry I was soon enabled to ascertain that both these patients were previously suffering from tabes, and presented symptoms of the disease at the time when the accident occurred; in the second, in fact, a tabid fracture had actually occurred. Be careful, therefore, gentlemen, not to consider as causes of the disease in the spinal cord fractures which are really its consequence.

All these ætiological elements are therefore invalid, or may be neglected. The true, I would almost say the sole, cause of tabes is *syphilis*. This opinion as to the syphilitic nature of the disease is due to one of the most eminent masters of our faculty, Fournier, who, in 1876, commenced to teach this in his lectures, and since that time has written works, which you all know, in order to demonstrate its truth.

In 1879 and 1881 Erb expressed himself in favour of this opinion, and the number of adherents who followed those authors is very large, so large in fact that I will not undertake to name them, contenting myself with mentioning to you, as the lectures continue, the authors of the statistics which contain the most information, since it is by means of statistics that we must advance in a question of this nature.

The number of tabid patients in whom syphilis can be found to have previously occurred is, according to Fournier, from 91 to 98 per cent. According to Erb (first statistic) it is 88 per cent.; according to Rumpf from 80 to 85 per cent.; according

to Althaus 90 per cent. By the side of these numbers I ought to mention, in fairness, those of Gowers and Seguin, which vary from 70 to 53 per cent. With regard to the statistic of Westphal, the number, which only reaches 14 per cent., differs so much from those which precede, and from my own observations, that I cannot but believe it to be vitiated by some important cause of error, and to be of no value as a guide.

Quite recently Erb* has published a second statistical account, including 369 new cases of tabes observed by him between 1883 and 1891. This work contains information which is so important, and in my opinion so convincing as regards the opinion maintained by me, that I shall claim your permission to borrow largely from it.

Of the 369 cases 300 refer to patients belonging to the higher class of society, 50 to the lower class, and 19 to women.

The 300 first cases are composed as follows :—

A. Cases in which it was impossible to find any indication of syphilitic infection ... ... = 11%
B. Cases in which syphilitic infection had occurred ... = 89%
The latter included :—
α. Cases in which symptons of secondary syphilis had existed ... ... = 63·3%
β. Cases in which the chancre had alone existed, no secondary syphilide having occurred ... ... = 25·7%

The 50 patients of the poorer class were composed as follows :—

A. Cases in which it was impossible to find any indication of syphilitic infection ... ... = 24%
B. Cases in which syphilitic infection had occurred ... = 76%
The latter included :—
α. Cases in which symptoms of secondary syphilis had existed ... ... = 52%
β. Cases in which the chancre had alone occurred, no syphilide having existed ... ... = 24%

Thus, in the first 300 patients suffering from tabes the total of 89 per cent. who had suffered from syphilis agrees completely with the figures of Fournier and Erb in his first statistical record. It is true that in the lower classes the number is not

* W. Erb, *Zur Ætiologie der Tabes. Berliner klinische Wochenschrift*, 1891, No. 29, p. 713. This work was published after the time at which the lecture was given. I thought it my duty to rearrange it in order to add these paragraphs, of which the importance will be evident to all.

so large, being only 76 per cent. This difference, however, is another reason for believing in the truth of the numbers first given, since it shows that syphilis is more often found to exist in tabes in proportion as the patients are more observant, and their intelligence greater.

It is doubtful whether the total of 89 per cent. indicates the exact proportion of those affected, since as you know, gentlemen, or soon will know, that persons who are very intelligent may certainly in their first youth have a chancre which is quite unperceived, or merely regarded as an insignificant abrasion, the existence of which is quite forgotten. Thus, everything being considered, the percentage of those suffering from syphilis in tabes would be considerably higher, and it is impossible to say at what number the limit would be reached.

Is the fact then definitely established that *nine-tenths of those affected by tabes have previously suffered from syphilis?* Some authors have looked upon this as a simple coincidence, while others by many reasons, or rather much reasoning, have sought to prove that tabes could not be due to syphilis.

Their mode of arguing is usually as follows:—

1. At the autopsy of tabid patients the lesions in the spinal cord are not found to resemble gummata in the slightest degree, nor are they in any way analogous to the ordinary specific lesions, no such lesion being again discovered in the other viscera.

To this it may be answered that many syphilitic affections of the skin or viscera exist which are also quite unaccompanied by lesions having a specific character.

2. The failure of treatment by mercury and mercurial preparations is an argument against the syphilitic nature of tabes.

The fact, which is perfectly true, does not however disprove our statement. In the first place writers on syphilis are agreed as to the point that certain syphilitic symptoms exist against which treatment by mercury and potassic iodine is almost inactive. Again it seems scarcely possible that it could have much effect when the lesion consists in complete destruction of certain fibres of the spinal cord. Would one, for instance, expect cerebral softening which is due by syphilitic arteritis to be cured by the influence of specific treatment? Why then should one suppose the spinal cord to act differently from the

other viscera when syphilis often produces in them lesions which are quite ineffaceable ?

3. Lastly, a great argument specially directed against the statistics themselves is the following statement: Syphilis is so common in the class from which the hospital patients come, that this, so to speak, is a common element, and cannot be invoked as a cause of the disease more than any other chronic disease might be. This argument is easily answered, and in this respect it suffices to ascertain how many persons suffering from syphilis are found amongst those who are free from tabes. Many authors have made statistical observations of this nature.

Erb, in a first statistical record, found that in the collection of 500 hospital patients who were free from tabes, 77 per cent. were *certainly unaffected by syphilis*, whilst it must not be forgotten, gentlemen, that 88 per cent. of the patients suffering from tabes were ascertained to be thus affected.

Levinsky found that in 620 hospital patients who were free from tabes, an analogous number, 80 to 85 per cent., were free from syphilis.

Nœgeli in 1,450 patients arrives at an even higher percentage.

Quite recently (1891), Erb, in the work from which I have already quoted, returns to this subject; he has examined 5,500 patients who passed through his hands with respect to the existence of syphilis, taking care to eliminate those suffering from tabes, and all the patients who came to consult him specially on account of syphilitic affections to the exclusion of every other affection. From this large number he obtained the following results: 22·5 per cent. of these patients were syphilitic, in the remaining 77·5 per cent. there neither was nor had been any indication whatever of syphilis. Compare these two numbers: 22·5 per cent. of syphilitic patients amongst those who did not suffer from tabes, and 89 per cent. amongst those who did so, and you yourselves will easily form an opinion as to the connection of tabes with syphilis.

Such are the principal arguments invoked by the authors who will not admit the connections which so evidently exist, in my opinion, between these two affections. I hope that I have shown you, gentlemen, that the refutation of these arguments is not really very difficult.

It would be an exaggeration to maintain, in the present state of our knowledge, that syphilis is the only cause of tabes, since

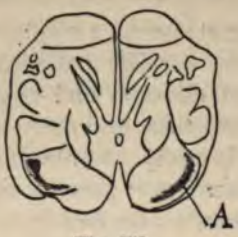


Fig. 158.

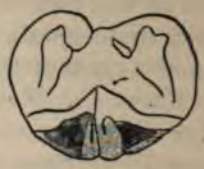


Fig. 159.

Fig. 160.

Fig. 161.

Fig. 162.

Spinal cord of a patient in whom degeneration of the posterior columns had occurred after intoxication by ergot of rye (according to Tuczek). The most severe lesions are designated by a black mark, those which were less pronounced being indicated by a grey tint. Fig. 158.—Section of the lower part of the medulla oblongata; the lesion is found upon both sides at A. Fig. 159.—Section of the spinal cord at the level of the decussation of the pyramid. Fig. 160.—Section of the spinal cord at the level of the second cervical nerve-root. Fig. 161.—Section of the spinal cord at the level of the 8th cervical nerve-root. Fig. 162.—Section of the spinal cord at the level of the 11th dorsal nerve-root. The lumbar and sacral regions present alterations of the same kind.

Tuczek has shown that *intoxication by ergot of rye* may produce lesions which are quite analogous to those of tabes. Symptoms of the same kind may occur in those affected by *pellagra*. It must therefore be admitted that from a theoretical point of view tabes may be due to certain special forms of *infection* or *intoxication*. Practically, however, you may feel sure of one thing, namely, that in ordinary practice *tabes invariably has a syphilitic origin*.

In addition to syphilis a very important place should be given to *hereditary predisposition* to nervous affections, to that singular condition of the organism which is transmitted from generation to generation, and owing to which the nervous system is specially vulnerable. As regards this tendency Charcot has truly observed that in a large number of tabid patients this hereditary tendency is very pronounced. It may also be observed, gentlemen, that the heredity in these cases need not be direct, that is to say,

tabid fathers have not necessarily tabid children, but may be indirect, the most different diseases of the nervous system possibly existing in the ancestry of these patients. Epilepsy, hysteria, hemiplegia, chorea, mental alienation, and psychical degeneration, in all their forms are found, according to Charcot, in the near relations of tabid patients. Another affection, diabetes, the pathology of which is also intimately allied to that of nervous disorders, is also one of the most frequent hereditary antecedents of locomotor ataxy (Charcot).

Heredity must also be invoked in the ætiology of tabes with regard to *hereditary syphilis*, Fournier considering that this affection may be the sole cause of the existence of tabes. I cannot say that this fact has been definitely proved; the future will show what should be thought as regards this connection, and enable us to know more certainly whether the affections of the spinal cord which are due to hereditary syphilis belong or not to the ordinary forms of tabes. It is possible that they may present special clinical features and lesions, as, for example, the disease of Friedreich (???).

Leaving, gentlemen, the question of the causes of tabes, which are now I hope understood by you as far as the great majority of cases is concerned, the other ætiological conditions of tabes will now be discussed.

The *age* at which the onset of the affection occurs is variable; it most often commences at a certain time of life (between 30 and 45 years). It is rarely observed before the age of 25 years, although cases have been recorded at the ages of 16 or 17 years (??); it is rare also after the age of 55 years.

In connection with its onset a most interesting question is that relating to the period of time which elapses between the existence of the chancre and the onset of tabes. Erb has given a statistical table of 300 patients in connection with this point, and arrived at the following results—

The onset of tabes occurred—

1 to 5	years	after the syphilitic infection	in	12·3	per cent.	of the cases.
9 to 10	"	"	"	37	"	"
11 to 15	"	"	"	24·7	"	"
16 to 20	"	"	"	14·2	"	"
21 to 25	"	"	"	4·8	"	"
26 to 30	"	"	"	1·9	"	"
31 to 35	"	"	"	0·7	"	"

The onset of tabes occurs therefore most often from 6 to 15 years after the syphilitic infection and it may be said in a general way, as Erb observes, that tabes may occur during the 20 years which follow the existence of syphilis (88 per cent. of the cases).

This knowledge, or the length of time which passes between the infection and the onset of the affection in the spinal cord, will enable you, gentlemen, to understand why it is that tabes specially occurs between the ages of 30 and 45 years. It is, in fact, between the ages of 20 and 30 years that syphilis is usually contracted; by adding to this period 6 to 15 years, which may be called the time of the *incubation of tabes*, the age of from 30 to 45 years is reached, during which clinical experience shows that tabes most often occurs. When again the infection has taken place at some other age, tabes is also found to appear at a correspondingly altered period; thus one of Erb's patients who had contracted syphilis at the age of 19½ years suffered from tabes at the age of 22, whilst in two others who only suffered from syphilis when mature age was approached, tabes occurred at a later age; in the one syphilis took place at 57, tabes at 66 years; in the other syphilis at 54, tabes at 59 years.

It will be interesting to consider the relation which exists between the form of syphilis and the onset of tabes. According to the information furnished by most of the patients it is the mild forms of syphilis which are most often followed by tabes. The chancre had not been severe and had healed in the normal length of time, secondary symptoms were entirely absent or but little pronounced, while tertiary symptoms have scarcely ever occurred; it is very rare that patients suffering from tabes present any evidence of a severe attack of syphilis. The mild character of the syphilitic symptoms explains, gentlemen, the hesitation with which some of the medical profession have recognized the connections which unite this disease to tabes, and the opposition which some authors still manifest with regard to this idea.

With respect to *sex* an important fact must be mentioned, namely that tabes is far more frequent in males than in females; so great is the difference, in fact, that in a number of cases collected since 1883 by Erb 350 males and only 19 females are found to be affected. Must this infrequency of tabes in the

female sex be referred to the fact that, other things being the same, this sex possesses a special immunity with respect to this affection of the spinal cord? This opinion does not seem very probable; it is more likely that the females being less often affected by syphilis than the males are on that account much less exposed to the danger of contracting tabes.

Perhaps also the greater frequency with which the nervous system in females is over-wrought should be taken into consideration. Syphilis, as Mobius has shown, plays the same part in the ætiology of tabes in females as in males; thus of the women suffering from tabes observed by Erb syphilis was indicated or its existence was extremely probable in 89·5 per cent.

Does race exercise any influence upon the frequency of tabes? It has been said to be more frequent in persons with blue eyes; for my part I believe that I have been able to recognize the truth of that assertion, but my observations have not been sufficiently numerous to enable me to form a definite conclusion. No works have been written upon this question of race, and I can only mention one statistical table of Minor which again is, as you will see, susceptible of another interpretation. This distinguished physician, who practises in a region of Russia where the Jews are somewhat numerous, examined how many tabid patients were found in an equal number of Jews and Russians; he found that in the latter there were 2·9; in the former only 0·8 per cent.; it thus appears that the *Jewish race*, in which from so many points of view the power of resistance and vitality are so remarkable, is gifted with a special immunity as regards tabes. Such, however, is not the opinion of Minor; this author has in fact reasonably observed that the Jews on the other hand are extremely liable to suffer from every affection of the nervous system; while he attributes the infrequency of tabes among them solely to the fact that on account of their special mode of life they are rarely affected by syphilis. This, then, would be another important, though indirect, proof of the part played by syphilis in the ætiology of tabes. This fact will be brought before us in another way during the study which we are about to make in connection with the effect of the professions upon tabes.

The different *professions* suffer most unequally from loco-

motor ataxy. If the relative number of persons in society belonging to the different professions is considered it is found that tabes is most frequent in *officers* and those belonging to the *liberal professions:* artists (those connected with the theatres, musicians, painters), clerks, journalists, &c. What then explains the greater frequency of tabes in these professions? Is it excessive use of the brain? Perhaps,—but is it not more probably inconsiderate and indiscreet activity of the feelings? I can bear witness to the great frequency of syphilis in these professions! An exception to the rule which exists with regard to the liberal professions should however be made, which is of some importance, namely that connected with the *priests of the different religions.* Thus in the statistics of Erb but one person belonging to the church is found, while there are at least 50 officers, and 26 in the medical profession. You will certainly divine, gentlemen, the reason of this immunity; it is that syphilis, it must be allowed, is less frequent in priests than in other classes of society. I should add that the only priest included in the statistics of Erb suffered also from syphilis; in this case again, as in all the facts which I have put before you in regard to the connection between tabes and syphilis, the exception confirms the rule. It is really, gentlemen, this concord in all the writings about the disease which has induced me, after mature deliberation, as you may be sure, to believe in the syphilitic origin of tabes.

LECTURE XXVI.

TABES.

DIAGNOSIS.

DIAGNOSIS: Difficulty in making a complete and methodical diagnosis. Differential character as regards: *Cerebellar affections, Insular sclerosis, Astasia-abasia, Syringo-myelia;* as regards *Pseudo-tabes:* A. *Toxic Pseudo-tabes*, alcohol, arsenic; B. *Diabetic pseudo-tabes;* C. *Neurasthenic pseudo-tabes.* THERAPEUTICS: A. *Internal remedies;* nitrate of silver, strychnine, aconitine, atropine, ergot of rye; anti-syphilitic agents: mercury, iodides; B. External remedies: counter irritation applied to the skin, electricity, massage, nerve-stretching, suspension, hydrotherapy; general summary of the treatment adopted.

The DIAGNOSIS of tabes, if completely considered, would require scarcely less than a volume to expound it; the whole of pathology would have to be reviewed. The most different disorders, either connected with the limbs and skin or with the different organs, may open the scene, and owing to their intensity occupy it exclusively, in such a way that the other symptoms due to the affection of the spinal cord are entirely masked by them.

Thus, for example, the *gastric disorders* of tabes may, as we have seen, stimulate the most severe organic affections of the stomach, or even cause an attack of cholera to be suspected.

Thus again the *genito-urinary derangements* of these patients often cause mistakes to be made, and lead to an organic affection of the urethra or bladder being wrongly suspected to exist, whence the designation of "false urinary patients" which F. Guyon rightly gave to them.

The same may be said of the *larnygeal disorders*, and of the *lightning pains*, so often termed rheumatoid, as also of the *ocular* or *auricular* troubles.

I have already, gentlemen, spoken to you of these different facts, and shall not therefore return to them.

Nor shall I now consider the diagnosis as regards certain affections of the spinal cord, with which you are not yet supposed to be acquainted, and which will be described in the course of

my lectures; in these cases it will be better to discuss the diagnosis when the diseases themselves are specially considered.

We will therefore only regard tabes at the present time under the aspect which it most usually presents, and compare it with certain diseases which present more or less exactly the same appearance.

The *cerebellar affections* have at times a very similar appearance; that is to say the patients who suffer from them present the same disorders in connection with the gait and upright position as well as various ocular troubles. A more careful examination, however, quickly reveals the differences which exist; there is no true inco-ordination but chiefly staggering; the lightning pains are nearly always absent, being often replaced by headache; there are no sensory disorders, the knee-jerk has not ceased to exist, and as regards the lesions in the fundus oculi these consist specially in the optic neuritis and not in optic atrophy.

Insular sclerosis may, as we have already seen, assume some of the characters of tabes. But in this disease also the changes in the gait resemble those which occur in cerebellar, or cerebellar spastic disease, and are not purely ataxic; the knee-jerk is usually retained or perhaps excessive, the sensory disorders or pains are but slightly pronounced; in addition to this numerous other symptoms exist (the tremor, nystagmus, scanning utterance, &c.) which are not observed in Duchenne's disease.

The disorders of the gait in tabes might, if the physician is not on his guard, be confused with those which were described by Charcot and Richer, and by P. Blocq under the name of Astasia-abasia, which consists in a loss of the memory as regards the movements necessary in walking. These cases, however, are distinguished by the fact that the knee-jerk is retained, and that the pains and other symptoms of tabes are absent. Again, whenever the patient is found to be seated or lying down the movements of the lower limbs are unaffected; and not only does he know exactly in what position these are placed, but can also direct them accurately towards a point indicated to him—in a word there is no inco-ordination, no true ataxy. The other modes of progression again are completely unaffected; thus as Charcot showed, the patient is quite able

to advance either by crawling or leaping or moving his chair; that which he cannot do is to walk. In tabes, on the contrary, inco-ordination, when it exists, affects every movement of the lower limbs whatever it may be.

Syringo-myelia, in which the symptoms are often numerous and, so to speak, unforseen, so varied is the seat of the lesions which constitute that affection, is often attended by phenomena, which are more or less analogous to those occurring in tabes: these consist of trophic derangements, sensory disorders, pains, and at times the loss of the knee-jerk. Usually the existence of the sensory disassociation which occurs in syringo-myelia will enable that affection to be distinguished from tabes; at the same time the latter disease may in some cases (Parmentier) be attended by this disassociation; the intensity and nature of the trophic disorders will be also a guide (the whitlows, the loss of one or several phalanges), as will the frequency and forms of amyotrophy. No exaggeration however must be made, the diagnosis between syringo-myelia and tabes being usually unattended by difficulty.

The same is not the case with regard to a group of affections to which, owing to their resemblance to tabes, the name of *Pseudo-tabes* has been given. These pseudotabid affections, depending upon morbid conditions which are very different from each other, have been specially studied in France, from the special point of view which we are now considering, by Leval-Picquechef; this group is classified as follows:—

A. *Toxic pseudo-tabes.*—These are produced by certain forms of intoxication, notably those which are due to *alcohol* and *arsenic*. It is to this class that the cases described under the name of neuro-tabes belong, which are in reality but cases of alcoholic pseudo-tabes.

In these cases, gentlemen, there is often most severe pain, very obvious disorder of the gait, and considerable diminution or loss of the knee-jerk; ocular disturbances consisting in impairment of vision and at times, but more rarely, ocular paralysis and ptosis exist.

When these symptoms occur the difficulty is at times very great. How may these cases of pseudo-tabes be distinguished from the real disease? A careful analysis of the symptoms will enable this to be done.

The fact is that the uncertain gait in these cases of pseudo-tabes is much less due to inco-ordination of movement than to a certain degree of paralysis or paresis which exists in the muscles of the leg and foot; these patients do not "mow" with the feet but "step," that is to say, on account of the paresis in the extensor muscles of the foot, they are obliged, in order to lift it from the ground and carry it forward, to raise the knee much higher than a healthy person would do, and hence the special mode of their progress which exists.

As regards the pains, they have not precisely the same characters as the lightning pains of tabes: they are rather lancinating and cramp-like, being specially seated in the muscles, and increased by pressure; lastly, they are more persistent, and less fugitive than the lightning pains, and occur rarely as these do in paroxysmal attacks.

The visual disorders also differ from those which occur in tabes, usually consisting at any rate in cases of alcoholism in more or less pronounced central dyschromatopsia, whilst with the ophthalmoscope atrophic discoloration is found limited to the temporal side of the optic disc.

B. *Diabetic pseudo-tabes.*—In this affection the gait is heavy, painful, undecided, much more often than ataxic; sometimes sensory disorders and powerlessness exist, often pain, which may be very severe, and lastly, loss of the knee-jerk, as Bouchard has observed, and of which he has shown the importance as regards the prognosis of the disease. I have just said, gentlemen, that the disorders of the gait when they exist rarely present an atoxic character; as regards the pains they are usually described by the diabetic patients as neuralgia (Worms, Dreyfous, &c.), and especially bilateral neuralgia. I will not dwell upon these points since the examination of the urine will remove every doubt. It must not be forgotten, however, gentlemen, that, as I have had occasion to observe before in the proper place, glycosuria sometimes, but rarely, occurs in the course of tabes.

Until the present time, with the exception of alcoholic pseudo-tabes, the forms mentioned are but seldom met with in practise: the last group of pseudo-tabes, of which I have still to speak, is far more important both on account of the complete way in which it resembles tabes and of the frequency with which it

occurs. I am referring, gentlemen, to an affection with which you often meet, to whatever class of society your patients may belong. I would even say whatever the class of diseases may be, which you specially treat; I mean, in one word, *Neurasthenia*.

In order to give you an idea of neurasthenic pseudo-tabes I shall not have to tax my memory to any great extent, so numerous and clear are the cases which I can recall. You scarcely know, gentlemen, how many of your colleagues are thus affected. Those in the medical profession, in fact, on account of their knowing the dangerous character of the disease, are the more ready to exaggerate the importance of the symptoms which they feel, or at least to interpret them in an erroneous manner.

Cases of neurasthenic pseudo-tabes present symptoms which are essentially tabid: pains having the "lightning" character at times, in some cases with a girdle-like sensation; a difficulty in the gait, which is clumsy, often staggering and resembling that due to intoxication, while at times there is more or less pronounced vertigo; in most cases there is also a more or less pronounced diminution in the genital power; lastly the knee reflex may be diminished or lost. Almost invariably, a fact which should be well noted, gentlemen, the patients come to you with the absolute conviction, boldly stated by them, that they are suffering from disease of the spinal cord. As you perceive, the condition of things is an embarrassing one for the physician, and those who are most distinguished have been led into error. If you would avoid mistaking neurasthenic pseudo-tabes for the true disease, this is how I would recommend you to act:—

In the first place, before anything else is done, put to your patients this searching question: "Have you ever suffered from syphilis?" If it is a man of education, whose sincerity you can trust, and the response is negative, consider that in all probability it is purely and simply a case of neurasthenic pseudo-tabes, and endeavour to show that this is the case by a careful examination of the following characters:—

(*a*) The pains, however acute they may be, never have the terrible severity which is so distressing in those due to tabes.

(*b*) The weakness of the genital functions, even when impotence exists, is not associated with loss of the cremasteric

reflex; I would ask you, gentlemen, to bear this remark specially in mind, as it may be of real service to you.

(*c*) The loss of the knee-jerk is not so absolute as it appears to be, that is to say if the precautions which I mentioned to you, when the reflexes were considered, are adopted. Thus, for instance, slight contraction of the quadriceps may be felt to occur by the left hand placed over that muscle at the moment when the blow is given to the tendon; or better still, if the patient is directed to exert the muscles in some way at the same time as the tendon is struck, according to the plan suggested by Jendrassik, the knee-jerk will be felt to be clearly produced. In a word, the knee-jerk is not absent, but only so much diminished that it cannot be produced in the ordinary way, though more delicate means of observation at once reveal its existence.

(*d*) Other neurasthenic symptoms exist, which you well know, and upon which I need not now dwell (headache of a special character, gastric derangements, psychical changes, &c.).

(*e*) Nothing is found in the pupil which recalls what is termed the symptom of Argyll-Robertson.

In a few words, gentlemen, it is not very difficult, as you perceive, to distinguish tabes from the affections which resemble it; the essential condition, that any error may be avoided, is to know that a mistake may be made.

An account of the methods of treatment which have been employed against this disease must now be rapidly given in order to complete the study of tabes.

THERAPEUTICS have unfortunately in most cases but a very theoretical value; the greater reason, gentlemen, for your knowing all the means which may be employed against this disease.

A. *Internal remedies.*—From a historical point of view two phases may be distinguished in connection with these remedies:

In the first those medicaments were almost indifferently employed which for some reason were believed to act specially upon the nervous centres. Among these the place of honour may be given to *nitrate of silver*; this salt was usually given to the amount of from 1 to 5 centigrams (gr. $\frac{1}{7}$ to gr. $\frac{5}{7}$) or more daily. Some patients took this medicine for months or years; I insist upon this point as you may meet with cases of the kind,

and should be forewarned in order not to be astonished at the dusky hue which the skin of these patients presents. This tint is in no way due to the trophic change of the skin which occurs in tabes, as one might at first suppose, but solely to the employment of silver as a medicine which the patient has taken during a long period of time.

Chloride of gold, arsenic, the salts of zinc and other metals have been recommended by a large number of authors.

Of the alkaloids, strychnine, aconitine, and atropine must be specially mentioned.

Another remedy drawn from the vegetable kingdom deserves special consideration, namely *ergot of rye:* this is almost the only medicine which has any perceptible effect, at any rate upon certain symptoms. It is to Charcot that we owe the knowledge of its influence, and of the cases in which it should be employed. The urinary disorders of tabes in fact are very successfully opposed by ergot of rye, and it is when they exist that this drug should be specially used, though at times it has also a favourable effect upon some of the other symptoms which occur in tabes.

In the employment of this remedy, however, certain precautions must be taken, and, as you are aware, gentlemen, the ingestion of ergot of rye in too large doses, or continued for too long a time, may produce the most unhappy effects; gangrene of a limb has been actually known to be produced by such administration of the drug. Ergot of rye should therefore be given as Charcot recommends, in moderate doses, and the patient should only be exposed to its effects during a short period of time; thus, for example, 2 or 3 doses of 30 centigrams (4½ grs.) of the powder of ergot of rye should be given *upon each of the first* 3 *days of the week* during a month or 6 weeks. The action of the ergot will be thus obtained without fear of the dangers which I have just mentioned.

The second or recent therapeutic phase is that in which, under the impulse of Fournier's discovery, a form of treatment based upon the ætiology of the disease was instituted, a form in which, in one word, the *anti-syphilitic* remedies were employed. I cannot certainly, gentlemen, be suspected of treachery after having declared myself as I have done, an absolute believer in the syphilitic nature of tabes; but at the same time it must be

candidly acknowledged that this treatment has apparently no beneficial effect upon the symptoms of tabes. On the other hand, if it does no good, it is only right to say that it may do harm, specially to the patients who are cachectic.

At the same time when a patient who is under my care seems able to support the anti-syphilitic treatment, I do not hesitate to employ it; nor should this practice be considered inconsistent or illogical. This form of treatment is not adopted as a cure for the actual symptoms of tabes, as I know but too well that it would be powerless in this respect. I only prescribe these remedies in the hope of making my patients secure from other lesions of a syphilitic nature, which at times are severe complications in tabes, as for example chronic arteritis, which is often followed by cerebral hemorrhage or general paralysis of the insane, the result of syphilitic affection of the meninges or brain.

I also adopt the anti-syphilitic treatment in hopes that the progress of tabes will be thus arrested. I dare not maintain that this is not an illusion, but it seems to me that during the last few years the patients in whom the symptoms are severe and pronounced, those suffering from "tabes major," as it is termed, have become less numerous; if this is true, would it not be due to the fact that in many cases the administration of specific treatment has been able to arrest the course of the disease; and it has in these cases remained stationary instead of being pitilessly progressive.*

After this profession of faith in these remedies, I will dwell no longer upon them, leaving you free, gentlemen, to adopt whichever mode of treatment seems the best. If you decide to use the specific remedies, I should recommend a mixed form of treatment, mercury being employed by means of friction; as regards iodides, you should prescribe them at the dose of at least 30 or 45 grains (2 or 3 grammes). It must be remembered, however, that in certain cases this form of treatment cannot be borne, and bad results occur, in which case it must be at once abandoned. This must be done, gentlemen, immediately and without hesitation, since, as I at first observed, there are some

* I am aware that in certain cases a severe attack of tabes has been seen (Fournier, Charcot) to occur in patients in whom anti-syphilitic treatment had been adopted and continued in a satisfactory way, from the time when the chancre existed; are not these, however, exceptions?

patients suffering from tabes in whom the specific treatment, far from being useful or inactive, is absolutely injurious.

B. *External remedies* comprise a large number of agents, among which I should mention the following:—

(*a*) *Counter irritation through the skin.*—*Setons*, the *cautery* and most forms of counter irritation have been almost entirely abandoned. One form, however, must be excepted which has justly been and is still held in great esteem. I mean the application of *heated points* which Charcot has always strongly recommended. These points should be applied, not over the spinal processes, but over the laminæ of the vertebræ; they should be numerous, very superficially placed, and re-applied about once a week.

(*b*) *Electricity* will consist either in the form of the *faradic current applied to the skin*, a plan which Rumpf states that he employed with real success, or of the voltaic current applied to the vertebral column either in the stable, weak or combined form, or at first stable and then weak, associating with it, if need be, as Erb recommends, galvanism of the peripheral nerves.

(*c*) *Massage and electro-massage* may produce some effect, but only upon certain special symptoms.

(*d*) *Nerve-stretching.*—If I speak of this process it is only from a historical point of view, since it is now entirely abandoned; the process consisted in dividing the skin and soft parts of the gluteal region or thigh until the trunk of the sciatic nerve was reached; more or less violent efferent traction (stretching) was then exercised upon this nerve. This plan, if I am not mistaken, had been previously employed by Nüssbaum in a patient suffering from obstinate neuralgia (and who in short was hysterical, and hence the brilliant success). Langenbuch applied the same process to tabes; if, however, some success was obtained in connection with the lightning pains, such complications were produced (paralysis of the lower extremity, death from meningitis, hæmato-myelia, shock, &c.) that it was soon abandoned.

(*e*) *Suspension.*—It is not necessary, gentlemen, that I should speak at much length about this mode of treatment; you all know it, and I need merely refer those amongst you who wish for a detailed account to the numerous reviews and articles which have been published on this subject in the *Progrès Médical* in 1889 and 1890. I need merely remind you that suspension

has a special effect upon certain symptoms, such as the lightning pains, the inco-ordination, and the genito-urinary disorders. Nor must it be forgotton that different contra-indications may exist to its employment in tabes, as in patients in whom either cardio-vascular affections, or atheroma, or enphysema, or advanced pulmonary phthisis, or, lastly, obesity, or even simply a very bad state of the teeth exists.

(*f*) *Hydrotherapy.*—Every process has been recommended in the treatment of tabes, and it must be acknowledged that some have proved effectual as regards one or other symptom, hot or cold *douche*, hot or cold *baths*, and even baths of steam or electrical baths; residence at different *thermal stations* is also recommended by many authors. Amongst these I should mention Balaruc, Néris, Plombières, Uriage, and specially Lamalou which seems to be specially efficient in the treatment of tabes.

To sum up, gentlemen, and to guide you in the right direction through the therapeutic labyrinth, how should one usually treat a case of tabes?

On the one hand, as you have seen, the most different symptoms exist; on the other, a nervous affection essentially due to syphilis.

Since one is as powerless as we now are with regard to tabes itself, it would be ridiculous and culpable to *treat the cause alone.* Remedies must be prescribed carefully in connection with the *symptoms* of the disease, the indications which I have just mentioned being borne in mind.

For the *inco-ordination* and *genital disorders* you will specially recommend *suspension.* When *urinary derangements* exist you will give *ergot of rye*, the precautions being taken upon which I have, I think, sufficiently dwelt.

The *lightning pains*, which in too many cases cause the patients to be true martyrs, will be opposed by all the anodynes which therapeutics have or do put into our hands: antipyrin, antifebrin, exalgin, phenacetin, salicylate of soda, aconitine, &c. Recourse will also be had to the different *narcotics:* hydrate of chloral, cannabine, hyosciamine, sulphonal, &c., and the preparations of opium, codeine, narceine, laudanum, thebaic extract. In connection with these remedies I must mention *subcutaneous injections of morphine.* Ought they to be practised or not in

cases of tabes? Considering the terrible influence which they have upon these patients, I should recommend you, gentlemen, to resist the entreaties of your patients as long as possible, otherwise you will undoubtedly cause them to be inveterate morphine maniacs. The injection of morphine is only excusable in cases in which the pain is really terrible, or only occurs in paroxysms (the gastric crises for example), it being then possible only to give the subcutaneous injections in quite a transitory manner.

The lightning pains may also be treated by numerous other external means than suspension: prolonged hot baths, douches, friction with ether, chloroform, the different anodyne balsams, the spray of ether or chloride of methyl, &c.

As regards the different *visceral crises*, they should be treated, as I have already said, by injections of morphine or other narcotics, by ice, by the different counter-irritants, and in some cases (the gastric crises) by the application of heated points or a blister to the *seat of pain* (Charcot).

When once the best treatment to be applied to the symptom has been settled, the disease and the patient may be considered. As regards the disease, you know my opinion as to the necessity of anti-syphilitic treatment. Should the state of health permit it, a trial of specific remedies is, as I think, fully authorized; you will then prescribe mercury, administered by means of friction, during three weeks, and as regards iodides, you may continue their administration during a longer period. It is fully understood that you watch carefully the effects of this treatment, and that should the least complication, or even indication of one, occur these remedies are at once discontinued.

As to the patient, you will already have done much for him in avoiding the injections of morphine. You will continue to help him by administering tonics and strengthening remedies in order to delay as long as possible the "consumption" of which I have already spoken upon several occasions. Is it advisable, as some authors, and specially Weir-Mitchell, recommend, to give complete rest to patients suffering from tabes, and even to keep them in bed? This is not my opinion, and, on the contrary, they should not, as I think, forget how to walk. If required, artificial means should be used, and a go-cart, used for several years at the Hospital of Bicêtre in order to enable

patients suffering from ataxy to walk, even in the advanced disease, may be of service in this respect. It must be well understood that the exercise of the muscles which I recommend should never be carried so far as to produce fatigue.

Fig. 163.—Go-cart, used at the Hospital of Bicêtre for many years to enable patients suffering from tabes to walk. It forms, as it were, a movable bar, which surrounds the patient on all sides and, moving with him, constitutes a basis of support which is always close at hand. A bench placed at one of the extremities enables the patient to rest whenever he wishes to do so. (From a stereotype of Damaschino.)

Lastly, gentlemen, I would remind you of the suggestion* which, whatever may have been said to the contrary, has never cured any organic affection of the spinal cord, but should on no account be forgotten, and on the other hand, stated in the clearest and most agreeable manner, the expression, I mean, of such an opinion as will calm the fears of the patient, and even in the most severe case will still cause the patient not to lose hope.

* It must be well understood that no reference is made here to hypnotic practices but only to the moral action of the physician.

LECTURE XXVII.

TABES (*continued*).

PATHOLOGICAL ANATOMY.

ANATOMY OF THE PARTS IN A NORMAL CONDITION. A. ANATOMY OF THE POSTERIOR COLUMN. Description of the three kinds of fibres: long, of medium length, short. (*a*) *Posterior column properly so called.* Researches of Flechsig upon the order in which the zones composing it are developed. Origin and termination of the fibres which constitute these different zones. (*b*) Zone of Lissauer: its seat, limits, extension; its division into two segments external and internal; the fibres which constitute it; their origin; development of this zone. B. ANATOMY OF THE POSTERIOR HORN. (*a*) Posterior horn properly so called: apex; gelatinous substance of Rolando, its division into (α) the spongy zone of the gelatinous substance, and (β) the typical gelatinous substance of Rolando. Spongy substance with its two zones, the anterior and the posterior. C. ANATOMY OF THE COLUMN OF CLARKE. Its seat, limits, and extension. Within this tract are found: (*a*) nerve cells; (*b*) a network of nerve fibres.

GENTLEMEN,—We have now to commence a study which is extremely difficult, full of obscurity and uncertain—that of the Pathological Anatomy of Tabes. Fortunate indeed are those whose opinion is formed upon this point, and who recognize in the "primary sclerosis of the posterior columns" a condition which is necessary and sufficient to produce the lesions of tabes. If some of you gentlemen are included in this number, I hope that the sight of our unceasing hesitation, and doubt, will not trouble a placidity which one cannot too much envy.

The changes in the spinal cord specially affecting the tracts in the *posterior columns* and *posterior cornu* of the spinal cord should, on account of my engagements, be described to you at the commencement of this discussion, and the principal knowledge which has been acquired as to the anatomy of these different tracts.

A. ANATOMY OF THE POSTERIOR COLUMN.—We have already considered the anatomy of this part as respects the origin of the fibres which compose it, while discussing the ascending

secondary degenerations. I showed you that for the most part it was constituted by the conjunction of the fibres of the posterior roots. You will remember, gentlemen, that amongst these fibres we agreed with Singer and Münzer in distinguishing *three groups* which vary in the length of their course. One of the groups passes almost immediately into the *extremity of the posterior cornu*, the intermediate group into the *column of Burdach*, while the third group composed of the longest fibres joins the *column of Goll*.

The knowledge of these facts is sufficient when it is a question of degeneration produced by a coarse lesion of the root-fibres (division of the roots, or transverse myelitis, compression of the cord). When, however, the disease is distinctly localised, as is the case in tabes, the principal tracts which constitute the posterior column must no longer be considered together, but the different groups of fibres which compose it must be taken into account. Until the last few years the only parts of the posterior column which were clearly distinguished were the *columns of Goll*, the *columns of Burdach*, and the *external bandlets* described by Charcot and Pierret. Recent works of which the principal are due to Flechsig and Lissauer have much extended our knowledge in this respect. These works are specially interesting to us inasmuch as the knowledge which they contribute as regards the normal anatomy of the spinal cord has often a special connection with the seat of the initial lesions in tabes; I should therefore be glad to convey this knowledge of the normal anatomy of the part to you before discussing these lesions.

(*a*) *Posterior column properly so called.*—It is to Flechsig, as I have just said, that we owe the increased knowledge which now exists in connection with this part. The researches of this author were made according to the same plan as that which gave such remarkable results as regards the other white tracts of the cord being based upon the distinct development of the different groups of fibres in the fœtus.

In the embryos of from 28 to 35 centimetres (11 to 14 inches) in length, Flechsig found that in the cords of which the myelin was coloured by any means whatever the posterior columns contained zones of different hues. The parts which were least coloured, and in which the myelin was consequently least

developed, are those marked by the letters A, D, and G, in figure 164; on the contrary, in the other parts the colour was much more marked, and consequently the myelin in far larger quantity.

In G, the fibres of the *column of Goll*, as you already know, will be developed, and which as you are aware, after a very long course along the posterior fissure, join the grey substance of the nuclei called "those of the column of Goll."

The lettters A, D, represent the *postero-internal root zone;* you will observe, gentlemen, that in the lumbar region this zone is divided into two parts by, as it were, a spur coming from the part marked B, of which the most internal and largest seems to be a projection from Goll's column; according to Flechsig, this is by no means the case, the segment being in no way connected with the fibres of that column.

As regards the other regions, the myelin does not develop simultaneously in them, so that the tracts can be distinguished from each other by means of the difference in the time of their development.

A number of fibres first appears close to the posterior commissure (specially in its lateral part) and the posterior cornu almost throughout its whole extent; it is to these fibres that Flechsig gives the name of *anterior root zone of the posterior columns.* We have elsewhere had occasion to speak of the fibres contained in this region, at the edge of the posterior commissure, while discussing secondary degeneration, and I have shown you, gentlemen, that a certain number of these seemed to be commissural fibres, which join the grey substance existing at different heights; hence their name of *cornu-commissural zone.*

The part seated between the anterior root zone of the posterior columns and the postero-internal root-zone has received the name of *middle root-zone* (B, E). Flechsig observes that all the fibres contained in this zone are not developed at the same moment; from this point of view two systems can be distinguished: the fibres of the *first system of the middle root-zone* develop cotemporaneously with those of the middle zone; while the fibres of the *second system of the middle root-zone* develop cotemporaneously with those of the column of Goll.

At the inner part of the posterior column, immediately adjoining the posterior fissure, is the middle zone F, C, which

Flechsig believes to be quite distinct from the columns of Goll. In the cervical region and in the upper two thirds of the dorsal region of the spinal cord, it is seated between the columns of

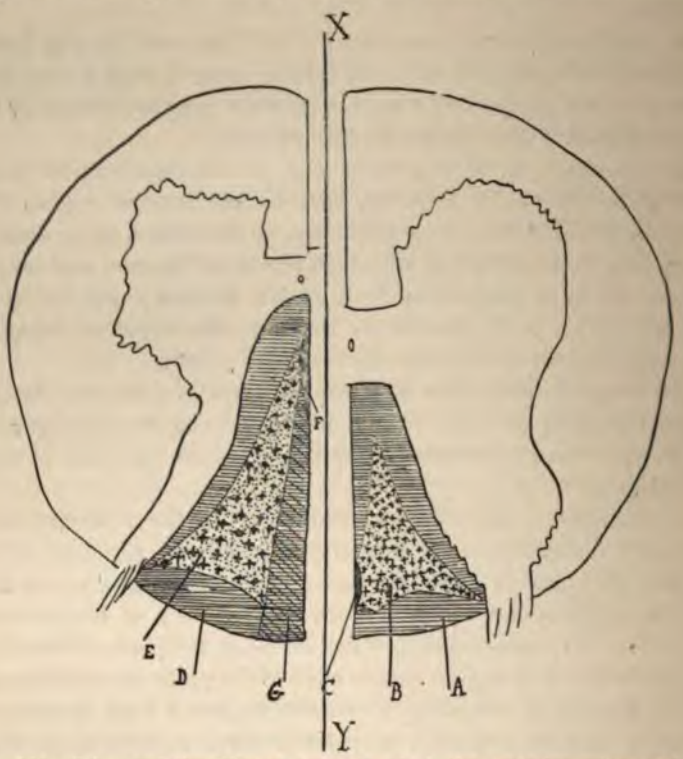


Fig. 164.—Scheme of the different zones in which the posterior columns of the spinal cord are developed. (After the description of Flechsig.) Upon the left of the line XY a section of the cord in the *cervical* region is represented in the figure; upon the right a section of the cord in the *lumbar* region; AD, *postero-internal root-zone;* it will be observed that in the spinal cord of the lumbar region, this zone A has the form of a gourd of which the smallest segment might (wrongly according to Flechsig) be considered as connected with the column of Goll. G, *Column of Goll.* B,E, *Middle root-zone containing* two kinds of fibres: + + + + = fibres of the *first system* of the middle root zone ···· = fibres of the *second system* of the middle root-zone. The part of the posterior column seated in front of and externally to BE and adjoining the posterior cornu (not represented by a number in the figure) is the *anterior root-zone.* C,F, middle zone. The column of Lissauer is not marked in the figure.

Goll and anteriorly to them, whereas below this point it is distinguished with greater difficulty from the middle root-zone.

If we recapitulate and consider the series of these different tracts in connection with the time of their development we find that the sheath of myelin first appears in the anterior root-zone, then in the first system of the middle root-zone R, at the same time as in the middle zone; the columns of Goll come next in order with the second system of the middle root-zone, as also with the postero-internal root-zone.

Such is the anatomy of these different tracts. However interesting the study may be, it would not be complete without inquiry being made as to the origin and termination of the fibres by which the tracts are constituted. In regard to these points the opinion of Flechsig will be given.

The *anterior root-zone* receives a large number of its fibres directly from the posterior roots. After a course which varies in length these fibres pass into the posterior cornua, becoming lost in their anterior portion. These fibres do not join Clarke's column, with which they are totally unconnected. On the other hand, as I have just reminded you, there are reasons for thinking that this zone contains a certain number of commissural fibres, which connect the grey substance which exists at different levels in the cord.

The fibres of the *first system of the middle root-zone* all issue from fibres of the posterior roots, and after a course, which is usually somewhat short, in the posterior columns, pass into the reticulum of Clarke's column; in the regions in which the latter have no clear existence these fibres pass with the posterior cornua between the edges of the anterior cornua.

The fibres of the *second system of the middle root-zone* in the lumbar region of the spinal cord are the origin of the fasciculi of fibres which at a higher level join together to form the columns of Goll.

As regards the *middle zone* no information seems to have been given in connection with them, and Flechsig himself states that he can give no positive opinion upon this point.

Such are the different tracts which constitute the posterior column. It must be well understood, gentlemen, that their limits and differentiation are not always so clear in reality as a description such as the one which I have just given you would indicate.

(*b*) *Zone of Lissauer.—Marginal zone* (Lissauer), *postero-ex-*

ternal root-zone (Flechsig).—A group of fibres is contained in this zone which essentially belongs, in reality, to the posterior column, but as they encroach upon the lateral column, specially as they have been unrecognized until the last few years, and their description does not seem to have made its way into the manuals which are placed in your hands, it seems to me a good plan to separate to some extent the study of these fibres from that of the rest of the posterior column.

As the name which I have just used for this tract indicates, it is to Lissauer, a pupil of Weigert, that the honour must be given of having first mentioned the existence of this zone, and of having described its structure. This zone is seated in the interval which separates the exterior angle of the posterior column from the internal and posterior angle of the lateral column. Thus it occupies the part through which the posterior roots pass in joining the cord and passing into the posterior cornu. Such are its limits in the transverse sense (as considered in a transverse section of the cord). In the antero-posterior direction this zone is limited in front by the spongy zone of the gelatinous substance which covers the extremity of the posterior cornu; posteriorly, it extends as far as the surface of the cord, which it just reaches (hence the name of *marginal zone* given by Lissauer).

The fibres of the posterior roots in entering the cord pass through the zone of Lissauer, which they divide into two unequal parts. One of these is on the outer side of the fibres of the posterior roots, the other being internal to them; the latter segment is the smallest in size, being, in fact, at times scarcely recognizable.

The *external segment of the zone of Lissauer*, which, as I have just said, is the largest in size, and which has a trapezoid form, extends along the outer edge of the posterior cornu and encroaches more or less upon the postero-interal angle of the lateral column, which it presses back to the same extent. Although essentially belonging to the posterior column, this segment appears at first sight to be an integral part of the lateral column. I shall have, while discussing the pathological anatomy, to return to the mistaken interpretation to which this singular appearance has been able to give rise.

The *inner segment of the zone of Lissauer*, of which the size

may not be more than a fifth or even a tenth part of that of the external segment, has, on the other hand, a triangular form and penetrates like a wedge between the horizontal fibres of the

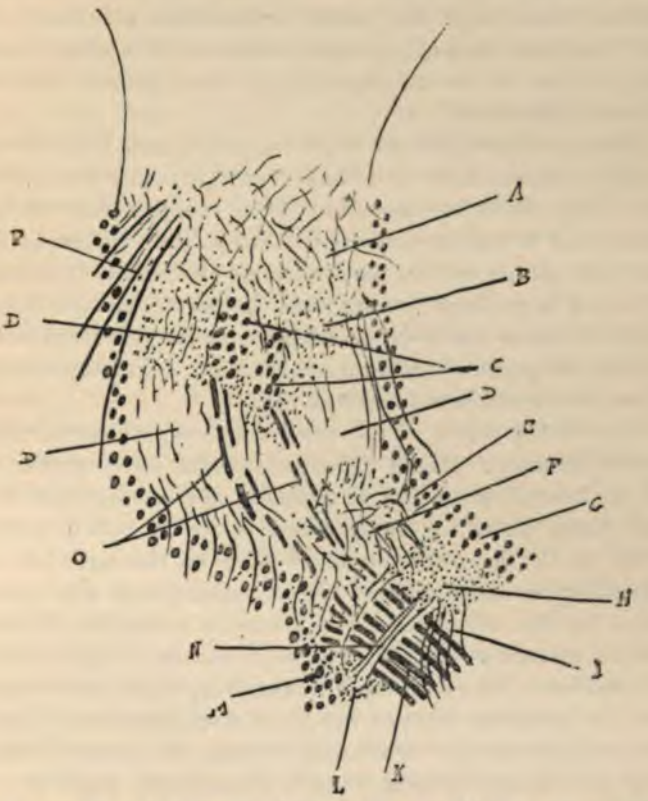


Fig. 165.—Scheme of the different tracts and fibres of which the posterior cornu is composed (lumbar region), after Lissauer. A.—Anterior zone of the spongy substance. B.—Posterior zone of the spongy substance. C.—Fasciculi of large fibres ascending in columns. D.—Gelatinous substance. E.—Fibres insinuating themselves between the spongy zone of the gelatinous substance and the fibres of the lateral column. F.—Spongy zone of the gelatinous substance. G.—Fibres of the lateral column adjoining the external margin of the posterior cornu. H.—External marginal zone (*external segment of the zone of Lissauer*). I.—Small root-fibres passing into the zone of Lissauer. K.—Posterior root-fibres. L.—Internal marginal zone (*internal segment of the zone of Lissauer*. M.—Fibres of the posterior column. N.—Fasciculi of transverse fibres becoming mixed with the posterior root-fibres at the moment of their penetration into the anterior cornu. O.—Large fibres joined in fasciculi, with a direct passage into the gelatinous substance. P.—Radiating fibres of the posterior column.

posterior roots and the vertical fibres of the postero-external angle of the posterior column.

The zone of Lissauer is found to be composed almost exclusively of *small nerve-fibres*, of much less size than those of the posterior roots or of the lateral or posterior columns; these small fibres are massed together in the zone in a larger number than is found to be the case in any other part of the white columns of the cord.

You must now however suppose, gentlemen, that the part which is named zone of Lissauer contains only small fibres; some large fibres coming either from the posterior or lateral column may be scattered therein. Not only do the small fibres which are special to this part insinuate themselves between the fasciculi of large fibres coming from the posterior roots, but they are also found in the intervals which separate the large vertical fibres of the posterior column immediately adjoining the apices of the posterior cornua from each other.

What is the *origin* of the small fibres which constitute the zone of Lissauer? These fibres come from the posterior roots and are merely, according to Lissauer, the continuation of the small fibres seated in the posterior roots in which they are known to be far more abundant than in the anterior roots. (According to some authors these small fibres are specially formed by the *collaterals* of the posterior roots, the knowledge of which we owe to the works of Golgi, Ramon y Cajal, Kölliker, von Lenhossek, &c.) However this may be, small fibres separate from the posterior roots at the point of their entrance into the cord, and pass almost horizontally towards the zone of Lissauer, being principally directed towards its external segment; they then curve upwards and pass to a certain height in the zone of Lissauer, from which they enter the gelatinous substance. Besides this group another portion of the small fibres of the posterior roots forms a second group which accompanies the large fibres of these roots, and passes with them either into the gelatinous substance of the posterior cornu, or into the interior of that cornu. In short, these small fibres, as you see, have but a short course in the zone of Lissauer which they soon leave; this explains how it is that notwithstanding the fact that fibres are incessantly brought to it by each posterior root, this zone does not increase in size from below upwards: on the contrary

it is in the lumbar region that it is most developed; its smallest size is in the dorsal region, and it becomes slightly more developed in the cervical region.

From this description, gentlemen, you will be able to understand how it is that the small fibres which constitute the zone of Lissauer, although having the same origin (the posterior roots), as most of the fibres forming the posterior column are clearly distinguished from them by a certain number of characters. The study of the development of the fibres which constitute this zone also accentuates this dissimilarity; Flechsig has, in fact, shown that these which in his nomenclature formed the *posterior and external root-zone* receive their sheath of myelin at a much later time than the other fibres of the posterior columns, namely quite at the end of fœtal life.

B. Anatomy of the Posterior Horn.—Under this name we will study the *posterior horn, properly so-called*, and the *columns of Clarke*, these two parts of the grey matter in the cord being apt to present changes in the course of tabes.

(*a*) *Posterior horn properly so-called:* the normal anatomy of the posterior horn has been the subject of many works; it is again from that of Lissauer that I shall borrow the principal facts in the description which I am about to give. (In connection with the whole of this description of the posterior horn consult fig. 165).

The extremity or "*apex*" of the posterior horn does not reach the surface in any way as you would be tempted to think when looking at a greater number of schematic figures which represent the spinal cord. It is separated from it by a *subpiamaterian layer*, which in fact occupies the whole surface of the cord (*vide* the very beautiful figures of Waldeyer in his memoir upon the spinal cord of the gorilla).

This extremity of the posterior horn is covered by the gelatinous substance of Rolando, which is itself coated externally by a special layer termed the *stratum zonale*, of which the tissue is analogous to that of the subpiamaterian layer with which in fact it is continuous.

The fibres of the *zone of Lissauer* or *marginal zone* are found to be entirely contained in the layer beneath the pia-mater through which they must consequently pass in order to reach the posterior horn.

As regards the *gelatinous substance of Rolando*, Lissauer divides it into two zones, which, however, are not separated from each other by any precise limit. One of these zones is seated posteriorly, viz., the *spongy zone of the gelatinous substance;* the other which is in front of this is the *typical gelatinous substance of Rolando.*

α. In the *spongy zone of the gelatinous substance* the root-fibres are more closely pressed together, and irregularly placed than in the rest of the gelatinous substance; it often extends to a slight distance along the inner margin of the posterior cornu. This zone is constituted by an admixture of fibres which are isolated from each other, and disposed in every direction: three principal origins may, according to Lissauer, be discovered for these fibres: 1. some fibres coming from the *marginal zone of Lissauer;* 2. some directly from the posterior roots; 3. some from the *posterior column.*

β. The *typical gelatinous substance of Rolando*, which, as I have just said, is placed immediately in front of the preceding zone, contains large fibres coming directly from the posterior roots, and narrow fibres coming from the spongy zone of the gelatinous substance. In this substance a number of *cells* are found which have been recently described and studied by Gierke and H. Virchow. Under a magnifying glass of moderate power the cells have the appearance of large spherical nuclei within round and clear spaces, since their protoplasm, which is almost colourless, can scarcely be recognized at first, though with a more highly magnifying power the protoplasm and its fine prolongations can be distinguished. The nucleolus of these cells is small, but the authors who have described it believe that it is constantly present. Owing to all these characters Gierke, H. Virchow, Waldeyer, believe these cells to differ from all other cells which exist with the exception of nerve-cells, and regard them as true *ganglion nerve-cells.* I would also add that contrary to the opinion expressed by some authors, the gelatinous substance of Rolando is, according to Weigert, the part of the spinal cord in which the neuroglia is least abundant.

If we still consider the parts from behind forwards we then meet with a new layer which, while surrounded to a great extent by the gelatinous substance of Rolando, is found to constitute the special substance of the posterior cornu.

This layer is the *spongy substance* properly so-called in which Lissauer also distinguishes two zones:

a. Posterior zone; this zone consists chiefly of the longitudinal fibres, some of which are small and belong in great part to the tract of the posterior root-fibres, while others are large and form a small number of fasciculi, seeming to be the direct continuation of the posterior roots.

β. Anterior zone; this differs from the preceding zone in consisting specially of fibres placed in a transverse direction.

According to the recent works of anatomists who have made use of the colouring process of Golgi; a large number of fibres and nervous collaterals terminate in the spongy substance, specially between it and the gelatinous substance, some ending in the latter part; it is the passage of these fibres and collaterals through the gelatinous substance which causes this to have a striated aspect; lastly, some of these fibres pass through the posterior commissure and end in the gelatinous substance of the posterior horn on the opposite side.

C. Anatomy of the column of Clarke.—These columns, notwithstanding some claims as to their having been first described in Germany (Waldeyer), would more rightly be called columns of Stilling-Clarke. Stilling, in fact, mentioned these groups of cells as early as in 1843, whilst the description of Lockhart-Clarke was only written in 1851; it should be added, however, that the latter is far the most complete both as regards the columns themselves, their seat, and their path.

These columns are, as you know, gentlemen, seated in the neck of the posterior horn, at its internal part, distinctly behind a transverse line passing through the posterior commissure, at any rate in the lower regions of the spinal cord, since at a slightly higher level these columns tend to pass more anteriorly.

They are not found in any other part of the spinal cord, but, according to the generally received opinion, only between the upper portion of the lumbar and that of the dorsal region. On the other hand Tooth, and I am much inclined to share his opinion, believes that the columns of Clarke only extend from the second lumbar to the eighth dorsal pair of nerves; at this level they cease to exist as columns, and between this point and the level of the second or first dorsal pair are only represented

by scattered cells; these cells also completely disappear in the cervical enlargement, reappearing in the upper cervical region. Masses of cells are again found, according to Tooth, in the medulla oblongata, within the nuclei connected with the columns of Goll and of Burdach, resembling those of the column of Clarke. Waldeyer also thinks that the columns of Clarke do not exist in the dorsal region of the cord alone, but throughout the whole length of the cord without exception; but that in other parts than the dorsal region it is not a large mass or real column that is found, but simply a group of two, three, or four isolated cells which so strongly resemble those of the columns of Clarke in their form and seat that they must be considered of the same nature.

In these columns we must, while considering how the pathological anatomy can be applied, specially consider two elements: (*a*) the *cells*, (*b*) the *nervous reticulum*.

(*a*) *Ganglion cells of the columns of Clarke.*—These are large cells which are consequently visible by means of a small magnifying power somewhat resembling in this respect the large cells of the anterior horns. They differ, however, considerably from the latter, since they are of smaller size, and present a much smaller number of processes; only one or two lateral processes can in fact be recognized to exist. According to the description of Obersteiner, both the upper and lower poles of these cells (in form they are elongated in the vertical sense) have a process which constantly exists; as, however, these processes form suddenly they are not fusiform in shape, and the body of the cell preserving its rounded outline they contain a large and well-marked nucleus and much pigment. In all probability it is from these cells that the fibres which constitute the direct cerebellar tract take their origin, to which they probably act as a trophic centre, having the same office perhaps in regard to some fibres of the column of Gowers.

(*b*) *Reticulum of the columns of Clarke.*—The knowledge of this reticulum is as you will see, gentlemen, very important in the study of the lesions which occur in tabes. In the normal cord coloured by the hæmatoxylin of Weigert the general tint of the section through the columns of Clarke is almost analogous (an exception being made of the brownish-yellow points representing the nerve-cells) to that of the remaining grey substance

of the anterior and posterior horns. It is somewhat dark on account of the presence of a thick network of nerve-fibres containing myelin, and it is only through this bluish network that some yellowish points or lines can be seen by transparence indicating the cells and position of the neuroglia. The nerve-fibres of this reticulum are chiefly afferent fibres coming from the posterior roots. At the present time I merely indicate these facts, to which I shall return when speaking of the pathological anatomy of tabes.

LECTURE XXVIII.

TABES (*continued*).

PATHOLOGICAL ANATOMY.

HISTORY: Hutin, Monod, Cruveilhier, Rokitansky, Türck, Romberg, Charcot and Pierret, &c. A.—SPINAL CORD. Macroscopic appearance. Appearance under the microscope.—(*a*) AT THE ONSET: *Posterior column—α. External bandlets*, their seat, their alterations; β. *Column of Goll*; γ. *Rest of the column of Burdach*; δ. *Zone of Lissauer*. (*b*) AT AN ADVANCED STAGE: 1. Posterior column—lesions of the *external bandlets*, the *column of Goll*, the *rest of the column of Burdach*. 2. Grey substance—α. *Anterior horn*; β. *Column of Clarke*; γ. *Posterior horn*; δ. *Central canal*. B.—POSTERIOR ROOTS, their lesions. C.—SPINAL GANGLIA, their lesions. D.—PERIPHERAL NERVES, their lesions. E.—BRAIN: disappearance of the nerve fibres of the convolutions.

GENTLEMEN,—We are now, I think, in a position to discuss the seat of the lesions which occur in tabes, or at any rate to form an approximative idea of where they exist. I will commence by enumerating the different changes which may be observed in tabes, and we will then endeavour to bring these lesions together as regards each other, and thus to discover the nature of the morbid process to which they are due.

You will pardon me, gentlemen, for not spending much time in speaking to you of the history of this disease, which is somewhat dull, and which, beginning at Hutin (1817), is continued in Monod, Cruveilhier, Rokitansky, Türck, Romberg, &c. The fact is that the lesions of the spinal cord which occur in tabes were known by those who studied pathological anatomy long before this affection was raised by clinical observers to the dignity of a morbid entity. A passage of Froriep quoted by Jaccoud is in this respect of great interest. It does not appear to me therefore that the idea of sclerosis of the posterior columns existing in tabes can be connected with one or other of these authors. On the other hand, however, I would specially mention the progress which has been made in the pathological anatomy of tabes when proof was afforded by Charcot and

Pierret that in this form of sclerosis *the external bandlets were first affected*, and that the morbid process was first seated in the region of these bandlets. This, in my opinion, is an important discovery, not only because it has met with general agreement but because it also indicates the way in which researches should be made in future with regard to the nature of tabid lesions.

I will commence, gentlemen, as I have already said, by stating the facts, after which the opinions which exist will be considered.

A. SPINAL CORD.—A certain number of changes in this part can be perceived even with the naked eye. Often, in fact, the cord is diminished somewhat in size, specially when the lesions are very pronounced. It is sometimes also slightly thickened, and the pia-mater is more or less opaque at the level of the posterior columns. Sometimes, again, when the membranes have been removed, these columns will be specially observed on account of their grey hue, which has the appearance of a long ribbon which extends over the whole length of the cord. In a section it is equally observed that this grey band encroaches upon the posterior columns, into the interior of which it extends more or less deeply. This, gentlemen, is what is observed, as I have just said, when the lesions are very pronounced; when they are but slightly marked the diagnoses as connected with the pathological anatomy of the disease can only be made by means of the microscope.

In order to understand thoroughly the microscopical appearance of the spinal cord in tabes it should be studied in two different stages of the affection; that of the onset, in fact, alone enables the seat of the fundamental lesions to be explained, since at a later period, when the whole posterior column is affected, no anatomical dissociation can possibly he made.

(*a*) AT THE ONSET.—When by means of a moderate magnifying power one observes the posterior columns, it is remarked that at certain points in them the nerve-fibres are far less numerous, while the tissue by which they are supported is much more abundant; a short examination is sufficient to show that these changes are really due to the formation of true zones of sclerosis.

POSTERIOR COLUMN.—Its different segments should be considered separately:

a. The *external bandlets*, that is to say the part of the posterior column which adjoins the posterior horn upon each side, not throughout its whole extent but specially in its middle

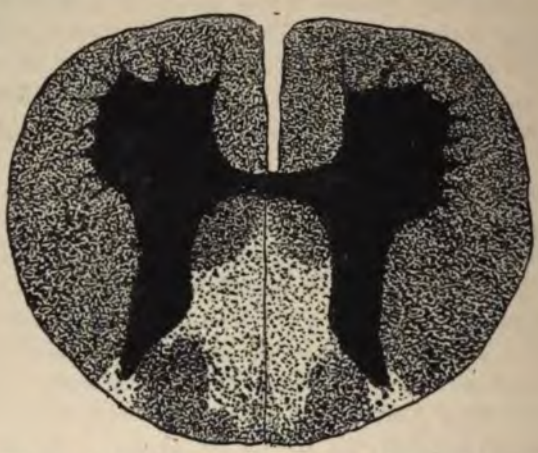


Fig. 166.—Section of the spinal cord in the lumbar region from a case of tabes at the onset. (The white parts are affected by sclerosis.) The change in the zones of Lissauer at the extremity of the posterior horns, and the seat of the sclerosis in the posterior column will be observed; this extends from the part immediately adjoining the posterior horns (external bandlets properly so-called), and spreading transversely as far as the posterior fissure, occupies the whole thickness of the posterior column.

and posterior thirds, and at which level numerous expansions of the nervous tissue are seen to pass into the middle part of the posterior horn. The form and size of these external bandlets varies somewhat according to the part of the cord at which they are observed, and I cannot now enumerate the varieties which exist; one thing, however, is certain, namely, that in all or almost all the autopsies of tabid patients, even at the onset of the disease, the external bandlets are affected. It should be observed, gentlemen, that they are not necessarily involved throughout the whole length of the cord, but may be almost or wholly unaffected in the cervical, though much changed in the lumbar region. Sometimes, however, they are affected not only in the lumbar, but also in the dorsal region, as well as in the cervical enlargement; above this point they are usually in a healthy condition.

β. The *column of Goll*.—This column is in most cases already

affected, but often to quite a slight degree, both as regards the intensity and extent of the lesions. Why they are seated in

Fig. 167.—Section of the spinal cord (dorsal region) from a case of tabes at the onset. The intensity of the lesion is proportionate to the light colour of the diagram. The column of Goll is affected: the column of Burdach on the other hand is free from disease except at its posterior part; the lesions are most pronounced in the external bandlets, properly so-called.

this part is not yet understood, and future researches will be required to explain the fact.

γ. *The rest of the column of Burdach.*—The extent to which sclerosis occurs in this part increases proportionately to the duration of the disease; in addition to this the islets become more apparent when the external bandlets of the subjacent segment of the spinal cord are much altered. The exact position of the lesions in this tract are, however, quite inexplicable to us in the present state of our knowledge; thus I prefer merely putting before you sketches which faithfully represent the appearance of the posterior columns in cases of tabes which are not yet in an advanced stage.

δ. *Zone of Lissauer.*—As you know, gentlemen, this zone is seated between the apex of the posterior horn and the surface of the cord, and is composed of a mass of narrow nerve-fibres. In tabes this zone, as Lissauer has shown, is much altered; all or almost all the small nerve-fibres which constitute it entirely disappear. Thus, in preparations coloured by the hæmatoxylin of Weigert, this region is found to have a special appearance,

seeming to be quite denuded. It is this appearance which has induced some authors to describe sclerosis of the postero-internal part of the lateral column as frequently co-existing in

Fig. 168.—Section of the spinal cord (cervical enlargement) from a case of tabes at the onset. The column of Goll is affected throughout its whole length. The middle zone of the column of Burdach is also involved, as well as the external bandlet properly so-called. The parts are affected in proportion to the lightness of their colour.

tabes. In reality the lateral column is in no way affected in these cases, the zone of Lissauer is alone changed, which, as I have already said, belongs entirely to the posterior root-zone. I should add that degeneration of the zone of Lissauer is one of the early changes in tabes; the examination of the lesions at this point, which is by no means difficult, being thus interesting in every respect.

(*b*) AT AN ADVANCED STAGE.—At this period again the appearance of the lesions presents great variations since the extent of the disease differs to an unlimited extent; I shall therefore but roughly indicate the changes which exist, feeling sure that the figures which I shall place before you will explain more than the most detailed description.

1. POSTERIOR COLUMN.—In very advanced cases nearly all this column may be affected. It then presents a singular appearance, specially in the dorsal region, where, on account of participation of the *zone of Lissauer*, the part affected by sclerosis extends beyond the posterior horn and, assuming the form of a "crescent," appears to extend at the expense of the lateral column. The *external bandlets* are, as you would expect,

completely degenerated, at any rate at the level of the lumbar and cervical enlargements, and in a large part of the dorsal region.

Fig. 169.—Section of the spinal cord in a case of tabes. The posterior column is affected throughout its whole extent, with the exception of a few healthy fibres which are completely isolated, and of which the number is slightly greater in the anterior portion adjoining the posterior commissure. The extension of the islet of sclerosis in the form of a fan will be specially observed. This appearance is due to the fact that the marginal zone of Lissauer is involved. It will be understood that on account of the occurrence of such cases as these the lesion has been supposed to extend from the posterior to the lateral columns.

The *column of Goll* is usually much affected. In some cases the lesions are found to be more pronounced in the cervical region than in other parts of the posterior column. In these cases the

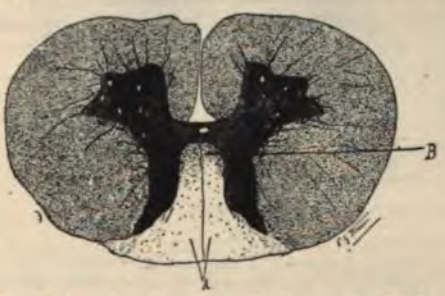


Fig. 170.—Spinal cord from the cervical region in a case of tabes (the parts light in colour are affected by sclerosis). The fibres B (commissural?) should be observed seated in the anterior part of the posterior column; extension of the lesions in the posterior column in the form of a fan on account of the zone of Lissauer being involved.

whole of the column of Goll is affected; in others, a part only of this column is involved.

Lastly, Strümpell has shown that a small fusiform tract of fibres is not rarely seen in a healthy condition at the junction of

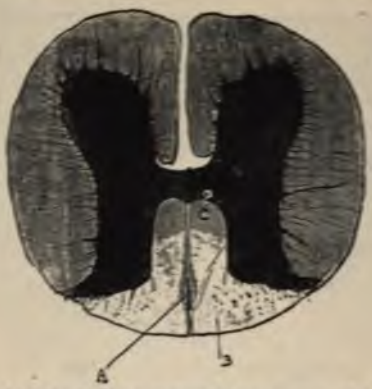


Fig. 171.—Spinal cord in the lumbar region from a case of tabes (the parts of a light colour are affected by sclerosis). The (commissural?) fibres (C) seated in the anterior part of the posterior column are sound. A small part (A) of the posterior column upon each side of the posterior fissure is also sound. The unaffected condition of this small part mentioned by Strümpell is not infrequent in the lumbar region of the spinal cord in tabes.

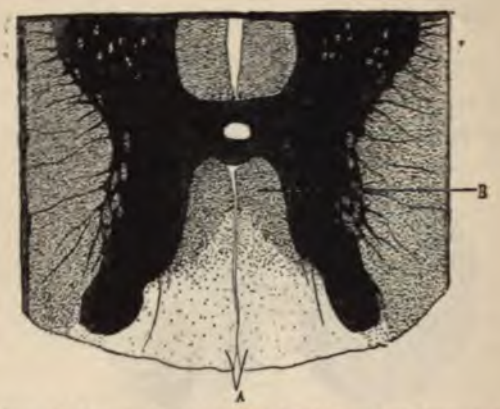


Fig. 172.—Section of the spinal cord in the lumbar region from a case of tabes (the parts of a light colour are affected by sclerosis). The (commissural?) fibres (B) seated in the anterior part of the posterior column are sound. It will be remarked that the sclerosis extends into the white columns at the extremity of the posterior horn, the small quadrilateral which is thus formed, and which one sees to be marked by small points, is the marginal zone of Lissauer affected by the degeneration; its colour should be less dark.

the posterior with the middle third of the posterior fissure. This small band of fibres corresponds to the middle zone of Flechsig.

As regards the rest of the column of Burdach it is in the middle part of this tract that the lesions are usually most pronounced, while certain parts of the column, as Strümpell has very truly observed, are only affected by degeneration at quite a late period of the disease. These are, on the one hand, the anterior part of the posterior column which adjoins the posterior

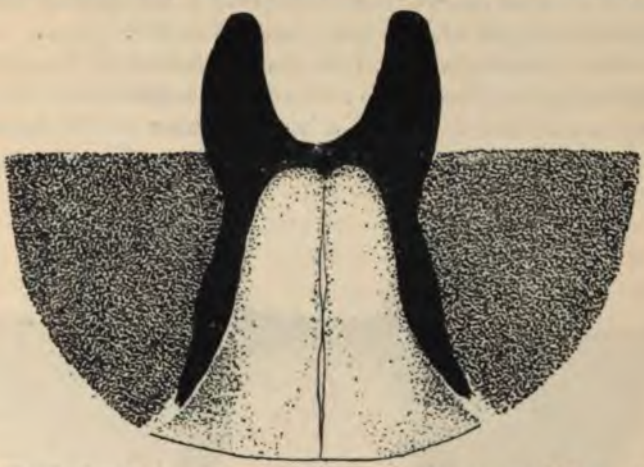


Fig. 173.—Section of the cord in the cervical region from a case of tabes. Almost the whole of the posterior column is affected with the exception of a few fibres at its posterior and external angle, which are sound, as are also the fibres seated behind the posterior commissure and along the base of the posterior cornu, the cornu-commissural zone. It will be also observed that lines of isolated fibres extend from the posterior commissure along the posterior fissure which they then quit in order to pass to the surface of the cord. These isolated fibres are the last traces of the healthy tracts which are found at this point in a less advanced stage of the disease. Compare fig. 191 with the above figure.

commissure, and the most anterior part of the base of the posterior horn*; on the other hand, the postero-external angle of the posterior column adjoining the posterior cornu. The healthy condition of the latter part is often very marked in sections of the cervical region. It should be observed, however, that this condition does not persist so long in that part as in the anterior portion.

* We have already seen, in connection with the secondary degeneration associated with transverse lesions of the cord, that a certain number of commissural fibres exist in this portion of the posterior column. It is probably these fibres which remain for a time unaffected in tabes, the so-called cornu-commissural zone.

When almost the whole of the posterior column is affected by the sclerotic lesions the posterior fissure not infrequently disappears as if the two halves of the cord were completely joined together.

The changes which I have just mentioned are usually identical in the posterior columns upon both sides. At the same time, there is in some cases obvious *asymmetry* in the islets of sclerosis as in the example which I now place before you.

From a histological point of view the lesions of the posterior columns are as follows: the meshes of *connective tissue* are much more extensive and contain a larger number of nuclei than in the normal condition. The number of *nerve-fibres* surrounded

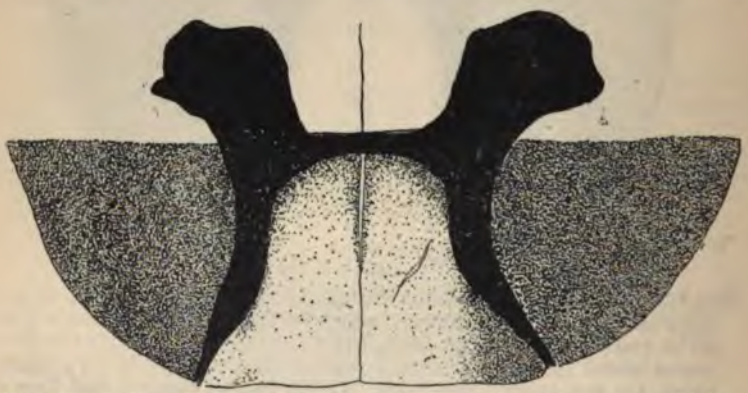


Fig. 174.—Section of the cervical region in the cervical cord from a case of tabes. This figure shows that some asymmetry may exist in the lesions of the posterior columns. Upon the right side, in fact in the angle formed by the junction of the posterior and external margins of the posterior column, a patch of sound tissue is found which does not exist upon the left side.

by this tissue is much diminished, and this diminution seems to me to occur specially and primarily in the fibres of small size which normally exist in the posterior columns. We have ourselves noticed, in fact, that in very pronounced cases the nerve-fibres of the posterior column may almost entirely disappear. It is usual to describe an abundance of granular bodies in the posterior columns. I must confess that for my part, although they have been most carefully sought, I have been rarely able to find any such bodies in the posterior columns of those who have suffered from tabes. On the other hand, I have recognized the absence of these elements both in cases in which the lesion was

commencing to exist and in those in which it was in an advanced condition. In this point I therefore completely disagree from most of those who study pathological anatomy. As regards the *blood-vessels*, their walls are much thickened at the spots in which they are surrounded by a fibrous zone, but only at these points.

I have already said a few words about the *meningeal* changes and the opacity of the pia-mater in the posterior columns. It seems that the degree of meningeal change corresponds more or less with that of the sclerosis in the cord, but the cause of this thickening of the membrane or, as it may be said, of this chronic meningitis is unknown.

2. *Grey substance.*—The changes which occur in the grey matter during the course of tabes have been frequently studied, and are the subject of some special works, specially by Pierret. The knowledge which we have upon this subject, however, is still by no means complete.

a. Anterior horn.—I spoke of the lesions which may be found in this part in a previous lecture, when discussing the different forms of amyotrophy which occur in tabes; it is unnecessary to say more about them, and other parts of the grey matter will be now considered.

β. Columns of Clarke.—The changes in this part, though mentioned by several authors, have only been really described in 1885 by Lissauer. Their character is as follows:—As you know, gentlemen, ganglion-cells on the one hand are distinguished in the columns of Clarke, and on the other a *reticulum* of narrow nerve-fibres. The reticulum is alone affected in tabes, the cells remaining free from disease, in such a way that the latter, in sections coloured by the hæmatoxylin of Weigert, instead of being placed, as in the normal condition, upon a blue ground, are found to be contained within a yellowish disc, which contrasts strikingly with the rest of the grey matter. This yellowish disc is merely the part occupied by the column of Clarke, of which the nerve-fibres which form the reticulum have lost their myelin and degenerated. Lissauer has remarked that at the time of the onset of tabes this degeneration of the fibres of the reticulum is much more pronounced upon the inner than upon the outer side of the columns of Clarke; he explains this difference in the intensity of the lesion by the fact that the

posterior root fibres which come from the lower part of the spinal cord (and which are usually the most affected) pass specially into the inner part of these columns, whereas those

Fig. 175—Column of Clarke in a healthy spinal cord (semidiagrammatic). A. Column of Clarke; B. posterior column. At the upper part and upon the left side traces of the posterior commissure are seen. Upon the right side and below the column of Clarke are seen tracts in the form of a fan which represent the posterior horn. The part marked by points which occupies the whole height of the figure upon the right side is the lateral column. The large black points in the column of Clarke represent the ganglion-cells (they should be in larger number); while the very small black points represent the reticulum of nerve-fibres which exists normally in Clarke's column.

Fig. 176.—Column of Clarke in the spinal cord from a case of tabes (semidiagrammatic). A', column of Clarke; B', change in posterior column (more evident) except, however, near the posterior horn (band of darker colour); C', posterior horn, &c.; D', posterior column which is affected (the lighter part), except in the part adjoining the posterior horn (the darker band); C', posterior horn; D', lateral column. The number of ganglion-cells is almost the same as in a healthy cord, but the reticulum of small nerve-fibres has almost entirely disappeared, and hence the clear appearance presented by the column of Clarke.

which come from the cord at a higher level join the external portion. This healthy condition of the cells in Clarke's column in tabes should be joined with the corresponding state of the direct cerebellar tracts, of which these cells are, as you know, the trophic centres. In the cases, which, however, are very rare,

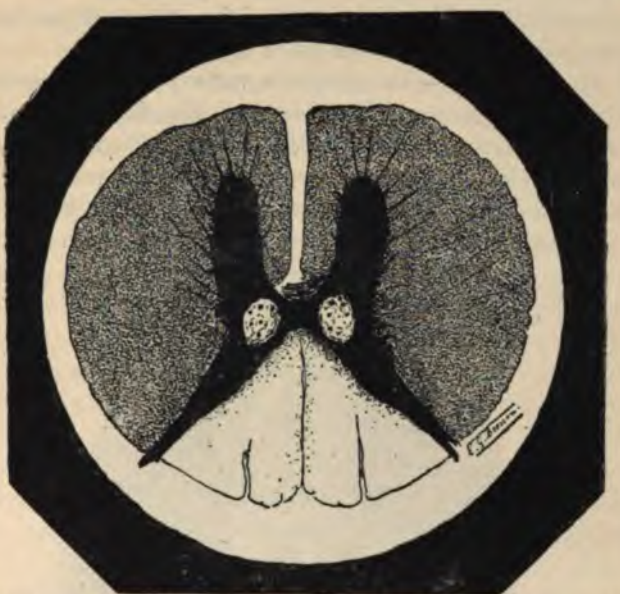


Fig. 177.—Section of the spinal cord (lower dorsal region) from a case of tabes. The whole of the posterior column is affected by sclerosis. The appearance of the columns of Clarke will be observed; their aspect is quite clear on account of the disappearance of the reticulum of nerve-fibres which exists in the healthy cord; the number of ganglion-cells has undergone no diminution.

in which the direct cerebellar tracts are affected (Jendrassik), their degeneration would perhaps be due to the fact that the cells in the column of Clarke are, by rare exception, involved.

γ. *Posterior horn.*—One would, *á priori*, expect that the grey matter of the posterior horn must inevitably be involved in the more or less pronounced lesions which occur in tabes, so intimate are the connections which exist between this part and the tracts of the posterior columns. Such is the opinion maintained by Lockhart, Clarke, Leyden, Pierret, &c., whilst Vulpian is more doubtful, and even considers that the lesions in the posterior horn are of exceptional occurrence.

In his work upon the condition of the posterior horn in tabes Lissauer analyses in a more exhaustive manner than had hitherto been done the lesions which may be met with in tabes. Amongst other things he mentions the disappearance of a large number of the small fibres seated in the *posterior zone of the spongy substance*, as also of the *radiating fibres which come from the posterior column*. As regards the *cells which are special* to the posterior horn, no precise information has been given upon this point.

δ. *Central canal.*—Its condition varies; not infrequently the cells of the ependyma in it are in such abundance as to obstruct the canal. In some cases considerable dilatation of the central canal coincides with pronounced lesions in the posterior horns. Whether this is a fortuitous occurrence in the course of tabes or an exceptional form of syringomyelia I am unable to say.

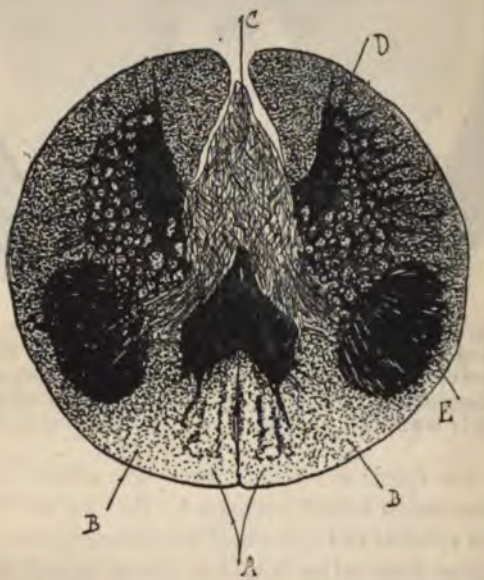


Fig. 178.—Section through the lower part of the medulla oblongata in a case of tabes (the white colour in the posterior columns indicates the parts affected by sclerosis). A. Lesions in the column of Goll. B. Lesions in the columns of Burdach. C. Decussation of the pyramids. D. Traces of the anterior horn. of the posterior horn.

rminating my remarks upon the pathological anatomy connection with the spinal cord, I would observe that

the latter may be affected throughout its whole length, from and inclusive of the *filum terminale* to the *medulla oblongata* and even higher. The study of the lesions which occur in the bulb in tabes is not within the scope of these lectures, besides which I would remind you that in another lecture I stated the facts which it was necessary to know in connection with the laryngeal crises and lingual hemiatrophy. I would only add, in order to terminate what is directly connected with the pathology of the spinal cord, that the changes in the column of Goll can be very clearly traced almost as far as the nucleus of that tract, while those of the column of Burdach can be similarly traced almost as far as the nucleus of the same name.

B. Posterior roots.—The atrophy of the posterior roots which occurs in tabes is so prominent a symptom that it has been recognized to occur from the very earliest time at which the disease was studied. Leyden is one of the authors who have most specially insisted upon the existence of this lesion in the roots of the spinal nerves, and is one of the few who have believed it to explain the origin of the changes in the spinal cord which occur in tabes. The roots of the cauda equina and lumbar enlargement specially suffer from this atrophy, which is also observed at the upper part of the spinal cord in which the roots connected with the cervical enlargement are chiefly affected.

Under the microscope the number of nerve-fibres in the roots which thus suffer from atrophy is found to be very considerably diminished. On the other hand, the number of small nerve-fibres appears to be sometimes increased, though I am unable to say whether this increase really exists or is simply relative.

It should be observed, gentlemen, that the cranial nerves, which for the most part (with the exception, certainly, of the optic and olfactory nerves) are, after leaving the medulla oblongata, but *posterior roots*, act in precisely the same way as the roots of the spinal nerves and are liable to analogous changes.

C. Ganglia of the spinal nerves.—The changes in these ganglia have been mentioned by a large number of authors who are by no means agreed as to their nature; no methodical description whatever of the lesions which occur has been written.

Some authors mention atrophy of the cells, others degeneration of the fibres which join them. In the ganglia which I have been able to examine the latter lesion has seemed to me by far the most clear, and notwithstanding my most careful investigation I was unable, to my great regret, to discover the existence of such or such an alteration of the cells. At the same time, as I shall soon say, it appears to me that some lesion very probably exists in these cells.

D. Peripheral nerves.—Extremely pronounced alterations undoubtedly exist in the peripheral nerves in tabes, and peripheral neuritis occurs with such frequency and intensity in tabes that it can scarcely be left unconsidered when the pathological anatomy of that affection is discussed. Westphal (1878) was apparently the first to mention its existence, but in quite a secondary manner, while referring to the lesions existing in the sensory branch of a nerve in an ataxic patient. Pierret followed, to whom the honour should undoubtedly be given of calling the special attention of the medical profession to the existence and cause of peripheral neuritis in tabes (1879). In 1883 Déjerine published other examples of this lesion, and ascribed to its existence a preponderating influence in the production of some of the symptoms which occur in tabes. Since that time Pitres and Vaillard, Oppenheim and Simmerling, Nonne, and many others, have shown how frequently peripheral neuritis occurs in tabes. As in all other affections in which it occurs (from the effect of a toxic agent, diabetes, pulmonary tuberculosis, different forms of cachexia, &c., &c.), the neuritis is more pronounced in the small branches of the nerves than in the large trunks, although it must not be supposed, gentlemen, as has been said, that the sensory fibres are alone affected; every branch of the nerve, whatever its functions may be, is liable to changes of this nature.

E. Brain.—It is not my intention to speak now of the focal lesions, which, as I have already said with regard to the hemiplegia which occurs in tabes, are sometimes found in the cere-
emispheres or pons varolii. There is in fact nothing
r in their nature. The cerebral changes which are special
s consist in a more or less marked disappearance of the
ibres contained in the convolutions. This change, which
analogous to that described by Tuczek as occurring in

general paralysis of the insane, was recognized by Jendrássik in two cases of tabes. In these the posterior and inferior convolutions, however, were affected, whereas in general paralysis it is usually the convolutions of the frontal lobe which are specially involved. I shall return to this subject in my next lecture, when I shall put fresh details before you.

LECTURE XXIX.

TABES (*continued*).

NATURE OF THE DISEASE.

NATURE OF TABES: Theories about this subject.—The two lesions of tabes are seated in the *brain* (Jendrássik).—Rôle of the *sympathetic nerve*.—Primary importance of the *vascular lesions*.—Rôle of the posterior meningitis.—*Primary sclerosis in the tracts of the posterior columns*.—Objections to this which is the leading opinion.—The fibres of the posterior column, in the same way as is the case with those in other columns of the cord, do not degenerate until the cells from which they part are affected.—Study of the cells from which the fibres of the posterior column originate:—Cells of the *spinal ganglia, peripheral ganglion cells*, arguments in favour of the existence of the latter, changes in the different cell elements.—Forms of *peripheral neuritis*:—Lesions of the *posterior roots*.—The lesions of the cord in the course of tabes are due to ascending secondary degeneration of the nerve fibres which come from the posterior roots.—The seat of the islets of sclerosis in the cord of tabid patients differs in the different cases on account of the fact that in patients suffering from this disease the same posterior roots are not always involved, or at any rate are not affected to the same degree, and that the degeneration first occurs in certain groups of their fibres.—Hence the different clinical types rest upon a good basis from an anatomical point of view: inferior tabes, cervical tabes, &c.—Explanation of the symmetry which usually exists in the lesions of the cord in tabes.—The *primum movens* of the change in the nerve cells to which the lesions of tabes are due is the toxic agent of syphilitic origin suspected by Strümpell.

THE NATURE of tabes is one of the points in neuro-pathology which has given rise to the most numerous and contradictory theories. Unfortunately, gentlemen, we have not yet escaped from the period of suppositions, and to my great regret I shall have little to offer you in this chapter, which treats of so important a branch of nervous pathology, which is not hypothetical.

On the other hand it is quite useless to mention all the theories which have been advanced, and you will permit me only to place before you those which are apparently of special interest.

The BRAIN, according to M. Jendrássik, is the seat of the most important lesions in tabes, and the greater number of the

symptoms of that affection are directly due to these cerebral lesions; this author in fact is much inclined to consider the

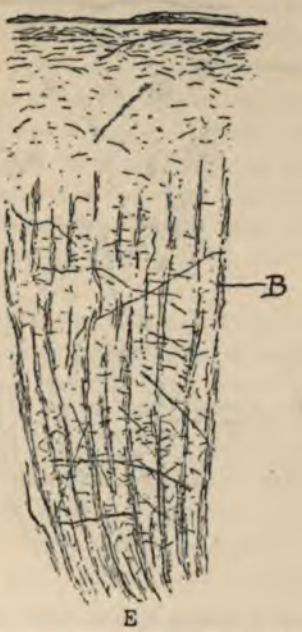


Fig. 179.—Section of the second frontal convolution (normal) made at right angles to its surface. (After Jendrássik.) The free surface of this convolution is at the upper part, while the lower part of the section (E) is continuous with the corona radiata. The vertical lines B represent the fasciculi of fibres which, in the healthy convolutions, contribute to form the rays of the corona radiata.

changes which occur in the spinal cord to be due to secondary degeneration caused by the lesions of the brain.

It is not unreasonable, gentlemen, that you hear this opinion for the first time with a feeling of surprise, so much does it differ from the views which are generally held. At the same time it should not be rejected without examination, since though even an observer of Jendrássik's importance may make a mistake, his error will certainly be associated with a large amount of truth. As I had occasion to remark in the preceding lecture, these cerebral lesions certainly exist in a large number of patients suffering from tabes, even when there is no indication of general paralysis. I am unable to say that they

are present in every patient, which I do not believe to be the case.

Fig. 180.—Section of the occipito-temporal convolution from a case of tabes made in a similar way to that in the preceding figure. (After Jendrássik.) The upper horizontal line represents the surface of the convolution.—A. Granular bodies in somewhat large number. The fasciculi of nerve-fibres (B') in this affected convolution are far less numerous, and thinner than in the preceding figure which represents a convolution in its normal condition.

The fact, however, is in itself true; as to its interpretation, I cannot accept that which is proposed by Jendrássik, which is specially based upon physiology, a frail science. The cerebral lesions which he regards as the cause of sclerosis in the cord, have in my opinion no direct connection with it; the convolutions suffer of their own accord in the same way as in certain cases a portion of the crus cerebri, pons varolii, or medulla oblongata. The cerebral lesions of tabes in fact are purely and simply due to the same cause as that which produces the lesions in the spinal cord, namely syphilis. They are very analogous to, if not identical with those which occur in general paralysis
ne; like them they are due to the direct and injurious
hilis upon the fibres of the cerebral convolutions,

independently of any destructive action it may have upon the posterior columns of the spinal cord.

If the symptoms due to changes in the cortex of the brain occur after the onset of tabes, general paralysis is said to have supervened in the course of tabes, if not, the reverse is said to have occurred. It is unknown at the same time why in certain cases general paralysis or tabes develop, either separately or conjointly, and why it is sometimes one and sometimes the other of these affections which first occurs; we are quite as ignorant of the reason why optic neuritis exists in some patients suffering from this disease, whereas in others it never occurs.

Certain alterations in the SYMPATHETIC nerve have been supposed by some authors to be the cause of the disease; Duchenne of Boulogne thought that the derangements were chiefly functional. MM. Raymond and Arthaud have been able to show, however, that organic lesions undoubtedly exist in that nerve.

In the opinion of Ordoñez, Adamkiewicz,*, Buzzard, &c., the sclerosis which exists in the posterior columns of the spinal cord is of vascular origin; the blood vessels contained in these columns, which supply them with blood, become the seat of more or less pronounced changes, and it is, in their opinion, consecutively to these alterations that sclerosis occurs in that part. This theory was for a certain time somewhat generally accepted; it seems reasonable, and, as I shall have occasion to show you, is well adapted to explain the lesions which exist in cases of diffuse combined sclerosis. When, however, an affection so clearly "systematic" as tabes is concerned, such an explanation seems to me impossible. How can it be supposed, in fact, that a disease in which the lesions are so small, and have such a definite seat, can be due to such a widely spread, I would almost say blind process, as that of perivascular inflammation?

The same arguments may be used in connection with those authors who, on account of the fact that the pia mater is found to be thickened in some cases of tabes, believe *posterior meningitis* to be responsible for the existence of the lesions which occur in tabes.

We will now consider the theory according to which *tabes is*

* This author, however, admits that sclerosis may be due to other causes.

a primary systematic sclerosis of the posterior columns; this, I may say, is the opinion which the greater number of neuro-pathologists have professed during the course of the last thirty years. Quite recently Flechsig has also discussed this subject, and owing to data which were furnished to him in his study of the development of the nervous tracts, has attempted to show that the points which are first affected in tabes correspond to fasciculi, which are quite definite from the developmental point of view.

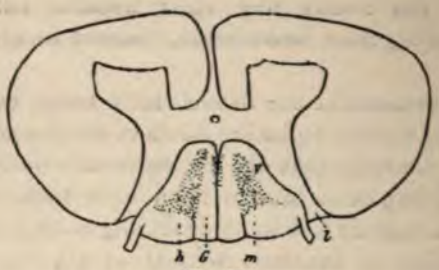


Fig. 181.

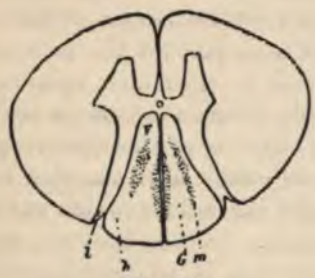


Fig. 182.

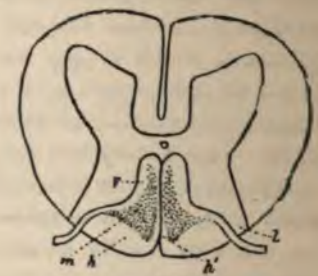


Fig. 183.

Diagrams of the spinal cord showing the lesions which exist in tabes at its onset. The pointed parts indicate the lesions; those of the zone of Lissauer have not been indicated. (After M. Flechsig.) Fig. 181.—Middle part of the cervical enlargement; the lesions would be specially seated in the zone *m* (middle root zone) and in the median zone. Fig. 182.—Upper half of the dorsal region of the cord in which lesions exist both in the middle root zone and in the median zone. Fig. 183.—Lumbar enlargement; the lesions are seated in the middle root zone. The signification of the letters is as follows: V, anterior root zone; *m*, middle root zone; G, column of Goll; *h*, external part of the posterior root zone; *h'*, internal part; *l*, zone of Lissauer. These diagrams are made from written accounts and correspond to the lesions which are most often found; they do not, however, represent every case of tabes at the onset.

cording to him, in fact, the first lesions which exist in tabes eated in the parts of the posterior column which you have

learnt to know by the names of *middle root zone* and *median zone*, the development of these two zones in the embryo being synchronous; the next lesions which occur in tabes are in the zone of Lissauer and columns of Goll, then in the *posterior and internal root zone*, and only at the last period of the disease in the *anterior root zone*.

I believe, gentlemen, that as regards our knowledge at the present time it is *impossible to admit that primary sclerosis of one or other column of the cord can possibly exist*. In order that a nerve fibre may degenerate, it must either be separated from the cell from which it takes origin, and which is its trophic centre, or the cell from which it comes must itself disappear. It would be paying far too much honour to these organs, which are simply conductors (nerves or columns of the cord), to attribute to them such an independent existence that they can suffer from primary lesions. No, gentlemen, every systematic alteration in the columns of the cord is but a secondary change; in all degenerations of a nervous tract, whether it is in the cord or has a peripheral seat, the *cell* in which disease exists must be specially sought.

Let us endeavour to apply these facts to a case of tabes. What, in that affection, is the system of fibres in the spinal cord in which the changes specially occur? It is the *posterior column*. Which, then, are the cells from which the fibres of the posterior column take origin, and do any reasons exist for supposing that these cells are diseased?

I have already, gentlemen, had occasion to discuss in some detail, in connection with secondary ascending degeneration, the origin of the fibres which constitute the posterior column. Allow me to remind you, in a few words, of the principal facts observed.

The posterior columns, we observed, do not at first form a definite part of the cord, at least during the earliest period of embryonic life. It originates in the lateral neural plates and only penetrates in a secondary manner into the cord by the medium of the posterior roots, which, budding from without inwards, penetrate into the thickness of the spinal cord; their fibres during this course are disposed in a direction parallel with each other, and the posterior column is thus formed. This, gentlemen, is a fact of the greatest importance which should

never be forgotten, namely, that the *fibres of the posterior column only belong to the spinal cord by accession.*

Which, then, are the cells from which the fibres of the posterior roots take origin? The opinion which generally exists is that all these fibres come from the nerve cells of which the *spinal ganglia are composed.* This, gentlemen, is said to be the fact as regards many, the greater number, in fact, of the posterior root fibres; is it, however, the case with respect to them all without exception? I should be very inclined to think that some of the posterior root fibres have a different origin, and come from nerve cells which, during embryonic life, are separated from the group of cells which formed the Neural Plates, and then made their way to the surface, and into the different regions of the body, being carried along by the centrifugal development of the different organs with which they had connected themselves (skin, muscles, tendons, aponeuroses, &c.). For this reason I propose to call these elements *peripheral ganglion cells.* It is very probable that all, or at any rate some of the nerve corpuscles which are called terminal, and which are found in different organs (the skin, aponeuroses, tendons), are really the peripheral ganglion cells to which I have just alluded. One difference alone, but an important one, exists between my opinion, as to the nature of these corpuscles, and that of other authors, namely, that I for my part, far from regarding them as *terminal organs*, believe them to be *cells from which the fibres take origin*, and from which they pass into the nerve trunks, posterior roots, and spinal cord, in which they contribute to form the posterior columns.

You will ask me, gentlemen, upon what I found my belief as to the existence of these fibres of peripheral origin. Unfortunately, the arguments which I shall not put before you are not, I fear, such as will at once convince you when they are heard.

In the first place, I must admit, that according to the opinion of the anatomists who most understand these questions the existence of a fibre of this nature has never been directly shown to exist. At the same time we know that in certain organs connected with the senses (olfactory mucous membrane, taste-buds, &c.) cells exist which are almost certainly seated at the origin and not at the termination of the nerves. In animals again the existence of such peripheral ganglion cells is very

much more clear; these animals are certainly far removed from us: the amphioxus, lumbricus,* &c.

On the other hand the experiments of Joseph showed him that if a nerve is divided below its spinal ganglion degeneration occurs in a certain number of fibres in that ganglion, and in the posterior root connected with it. If the results of these experiments were conceded by all those interested in pathological anatomy the truth of my supposition might be looked upon as at once demonstrated. I must admit, however, gentlemen, that these conclusions have met with great opposition.

In the third place, and I feel that this reason has a more solid foundation, since it concerns human pathology, it has been shown that after amputations of the thigh, as I have explained to you in a detailed manner, and division of a mixed nerve such as the sciatic *ascending degeneration* takes place in that nerve; this is followed by obvious atrophy of the corresponding posterior column. We must, therefore, necessarily, and upon that occasion I reminded you of how Friedländer and Krause had made the same observation, we must necessarily, as I said, admit that fibres exist in that nerve of which the trophic centre is seated in some part of the amputated limb, and of which the degeneration is consequently ascending. I would also observe, gentlemen, that in the case of amputation of the thigh, which I have taken as the basis of my description, the lesions due to the degeneration of the nerve extended as far as the spinal cord, and that in this part they were seated in the same points as those which exist in tabes (the external bandlets, the column of Burdach, the column of Goll).

I have, however, allowed this digression to carry me away from the subject of the lecture, nothing being so complicated as the explanation of a hypothesis.

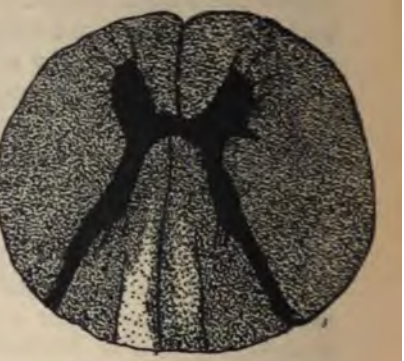
We were saying, gentlemen, that the nerve fibres contained in the posterior roots come from two distinct trophic centres: one constituted by the *cells of the corresponding spinal ganglion*, the other by the *peripheral ganglion cells*.

I have just mentioned to you the argument which I thought important as indicating change in the peripheral ganglion cells

* M. Von Lenhossek. Ursprung, Verlauf und Endigung der sensibeln Nervenfasern bei Lumbricus. Archiv. für Mikroskopische Anatomie, t. XXXIX. 1892.

in tabes. The *condition of the spinal ganglia* in that disease must now be considered.

As I have already observed to you, gentlemen, the recognition



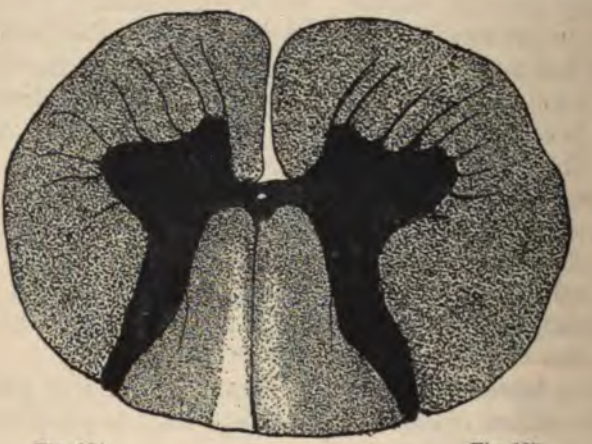


Fig. 184. Fig. 185.

Fig. 186.

Fig. 184.—Section of the cord in the lumbar region. Fig. 185.—Section of the cord in the dorsal region. Fig. 186.—Section of the cord in the cervical region. From a case of amputation of the left thigh It will be observed: 1. That in the lumbar region into which the nerve fibres coming from the divided nervous parts the external bandlet the column of Goll and that of Burdach are affected by degeneration, on the contrary in the dorsal and cervical regions, which no longer receive the affected fibres, the colums of Goll alone show signs of degeneration. 2. On the left side the degeneration of the nerve fibres produces alterations in the posterior column not only of the left but also of the right side; this fact will explain the symmetry of the lesions in tabes notwithstanding the number and independence of the centres in which the degeneration commences (spinal ganglia and posterior roots).

of the lesions in these organs is attended by some difficulties, and as, when an autopsy is made it is not habitual to keep these parts, in order that a subsequent examination may be made, the knowledge with respect to their lesions is but slight. At the same time the writings of Oppenheim and Siemerling, to speak only of recent works (observations of this kind published more than fifteen years ago have much less value on account of the progress which has been made in the mode of carrying on the examination), describe and figure considerable lesions in the cells and fibres of the spinal ganglia in tabes. For my part I could not speak so definitely in the affirmative, or say that in the one case in which I specially searched for them lesions clearly existed in these nerve cells. At the same time I quite believe that in this case the cells were less numerous than should be the case in an unaffected ganglion, and some of them seemed to present signs of degenerative atrophy. In the same case atrophy clearly existed in a somewhat large number of the nerve fibres contained in the fasciculi which pass within the spinal ganglia. I am, therefore, convinced that changes do exist in the spinal ganglia in all cases of tabes, and that these changes* are of the greatest importance as regards the origin of the process of degeneration in tabes.

It is somewhat probable, gentlemen, that these lesions in the cells of the spinal ganglia are also the cause of the disorders known by the name of *peripheral neuritis*. You know, in fact, that these cells have a trophic effect both upon the fibres of the posterior roots and upon those of the peripheral nerve trunks. The lesions of the ganglion cells would therefore be accompanied by degeneration in the peripheral nerve trunks. This, it seems, should occur in tabes, and such, in my opinion, is in part the cause of some of the changes which occur in the peripheral nerves. This, however, would not explain all the changes which exist, specially the fact that the small peripheral branches are more involved than the nerve trunks themselves.

I am convinced, for my part, that the degeneration of these

* During the publication of these lectures I learn from an article by Darier (Gaz. hebdom de Méd., 30th January, 1892) that M. Babinski also expressed the opinion in 1891 that the origin of the lesions in tabes should be attributed to the spinal ganglia. Babinski is of opinion that a *functional derangement* in the functions of the cells in these ganglia would be sufficient to produce the morbid process which occurs in tabes.

peripheral ganglion cells, of which I have frequently spoken to you, is greatly concerned in the origin of the peripheral neuritis which occurs in tabes. Nor can I possibly admit that simple organs of transmission, such as the nerves, can be primarily and spontaneously the seat of such a decided lesion as that which is known by the name of peripheral neuritis, and I am as decidedly opposed to this theory, as I have already explained, as I am to that which supposes primary affections to exist in the columns of the spinal cord.

Now that you understand, gentlemen, how and why changes

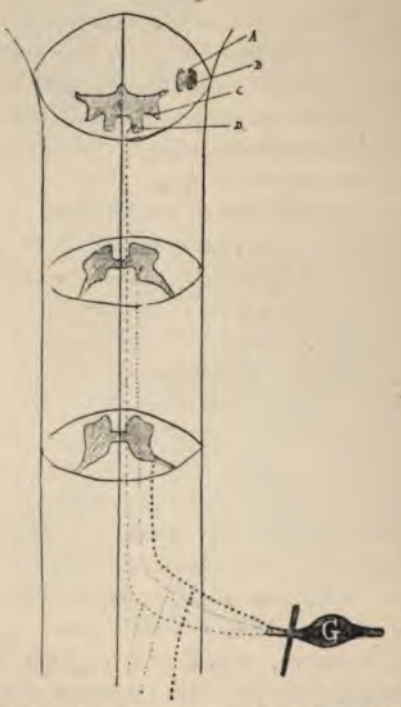
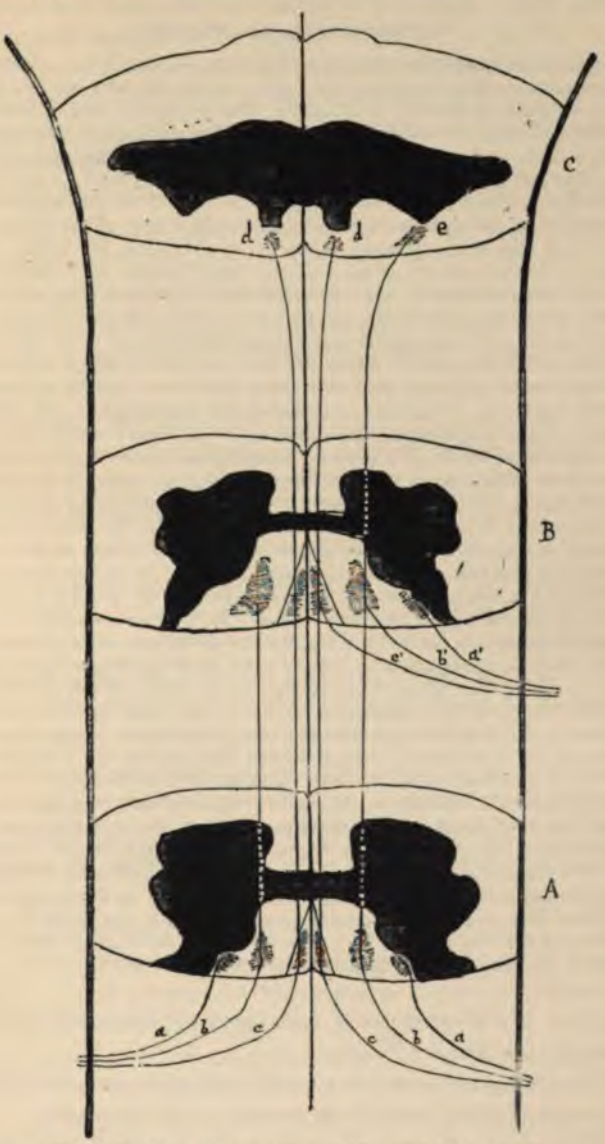


Fig. 187.—Diagram of the course and termination of the posterior root fibres. The dark fusiform patch (G) placed below and on the right side represents a spinal ganglion, the line which crosses it indicating the point of division of the posterior root which issues from it. This posterior root divides into three branches, each of which gives off secondary descending branches. + + + + Short fibres passing to the apex of the posterior horn; .... fibres of moderate length which join the posterior horn at its base after passing upwards in the column of Burdach; ---- long fibres passing into the nucleus of the column of Goll (nucleus of the class) D after passing upwards in the column of Goll. The nucleus of Burdach exists in C.

occur in the *fibres of the posterior roots*, I must show you how it is that the degeneration of these fibres which penetrate into the cord in order to form the posterior column, produces sclerosis



Figs 188–190.—The meaning will be explained directly.

in this part. But in order that you may be able to understand completely what is meant, I must first remind you of the course which the posterior root fibres pursue in the spinal cord (*vide*

Explanation of the figs. 188—190.

This diagram is meant to show that the seat of the lesions of the spinal cord in tabes varies according as the posterior roots connected with one or other pair of spinal nerves are affected, or as one or other group of fibres in that root is specially involved. A, section of the cord in the lumbar region; B, section of the cord in the cervical region; C, section of the medulla oblongata at its lower part. The left half of the figure represents a spinal cord in which the posterior roots are only affected in the lumbar region (*a*, *b*, *c*). The right half represents a cord in which the posterior roots are affected not only in the lumbar region (*a*, *b*, *c*, but also in the cervical region (*a'*, *b'*, *c'*). Upon the LEFT SIDE the change in the *short fibres* (*a*) of the posterior roots produces in the *lumbar* region a zone of degeneration in the *external bandlet*; the change in the fibres of *moderate length* (*b*) produces degeneration in the *column of Burdach*; the change in the long fibres (*c*) produces degeneration in the *column of Goll*. In the *dorsal* and more still in the *cervical* region (B), however, it is no longer the same, and the roots which pass into the cord being here in a healthy condition the *external bandlet* is completely unaffected by degeneration; the fibres of moderate length and the long fibres affected in the lumbar region alone reaching to this height in the cord produce degeneration in the *columns of Burdach and of Goll*. While other affected fibres of lumbar origin do not reach the cervical region, the long fibres extend to the medulla oblongata and produce at that part a zone of sclerosis in the column of Goll. On the RIGHT SIDE of the DIAGRAM the zones of degeneration produced by the change in the posterior roots of the lumbar nerves (*a*, *b*, *c*) are the same as on the left side; the *external bandlet, column of Burdach, column of Goll*, in the section of the spinal cord in the lumbar region (A); the *column of Burdach* and *column of Goll* in the section of the cord in the cervical region (B); the column of Goll in the section of the medulla oblongata (C). On account of the posterior roots, however (*a'*, *b'*, *c'*), being also affected on the right side which did not happen on the left side, the zone of degeneration exists in the right *external bandlet*; in addition to this (which is not marked in the diagram) the degeneration in the column of Burdach is much more extensive than on the left side, since upon that side it receives in addition the fibres of *moderate length* and *long* fibres which are affected. Lastly, in the *medulla oblongata* upon the right side these fibres of *moderate length* and *long* fibres coming from the posterior roots, which are affected in the cervical region in joining the *nucleus of Burdach*, produce in the *column* degeneration which does not exist upon the left side. It is also very probable that in any pair of spinal nerves one group of fibres can be alone affected at first, and that this group is specially that of the fibres of moderate length or of the long fibres. This explains the frequent predominance of the lesions in the column of Burdach and the column of Goll.

fig. 187). We divided them into three groups in the same way as Singer and Münzer:

1. The group of *short fibres* which pass into the posterior horn almost immediately after their entrance into the cord.

2. The group of fibres of *moderate length* which pass upwards

for a certain distance in the cord, being inclined towards the posterior fissure and then curving outwards and passing into the posterior horn nearly at its middle part; a certain number of these fibres pass into the columns of Clarke. This group of fibres is contained in the column of Burdach.

3. The group of *long fibres;* these fibres come specially from the roots of the cauda equina and pass through the whole length of the cord to join certain nuclei in the medulla oblongata; they constitute the columns of Goll.

We will now consider how all these anatomical data accord with the conclusions furnished by the autopsies of those who have suffered from tabes.

In the spinal cord the initial seat of the lesions as we have said is in the *external bandlets* and *zone of Lissauer;* this is the more easy to understand on account of these being the points at which the posterior roots (the short fibres and those of moderate length) enter the cord and pass into the grey matter of the posterior horns. As regards the *sclerosis* which occurs in the *columns of Burdach,* it is specially due to degeneration of the fibres of moderate length, the disappearance of the reticulum of nerve fibres in the columns of Clarke being also the cause. Lastly, the sclerosis of the *columns of Goll* is directly due to the degeneration which occurs in the long root fibres.

You will thus be able to completely understand, gentlemen, how the well-known zones of sclerosis of the posterior columns are formed in tabes. It should be observed also that degeneration may occur not in the fibres connected with one or two of the spinal ganglia alone, but perhaps in those connected with a large number, or possibly them all. You will thus understand why in some cases lesions occur in the external bandlets throughout almost the whole length of the cord, while all the root fibres in the upper part of the cord, the short fibres being comprised, are affected similarly to those in the lower regions.

In other cases, on the contrary, the zone of the external bandlets is not affected in the cervical region or even in the upper part of the dorsal region. In these cases the seat of the lesions is limited to the column of Goll and the middle portion of the column of Burdach (*vide* fig. 191). It may then be supposed, gentlemen, that the lesions of the posterior root fibres and consequently of the corresponding spinal ganglia only

existed in the lower part of the cord (the cauda equina, lumbar, and lower part of the dorsal region); the degeneration has therefore only involved the upper part of the cord, the fibres of moderate length and long fibres coming from the roots at a lower level, that is to say, the fibres which are exclusively seated in the column of Goll and the column of Burdach.

It may again be supposed that the groups of fibres in the different posterior roots are not simultaneously affected, but that the lesions commence in the group of long fibres, and in that of the fibres of moderate length; it will then be understood how in sections of the cord which are made in parts where the lesions are by no means in an advanced stage (the dorsal or cervical region), the changes are seated almost exclusively in the column of Burdach, and to a slight extent in the column of Goll; the external bandlets properly so-called are found to be almost

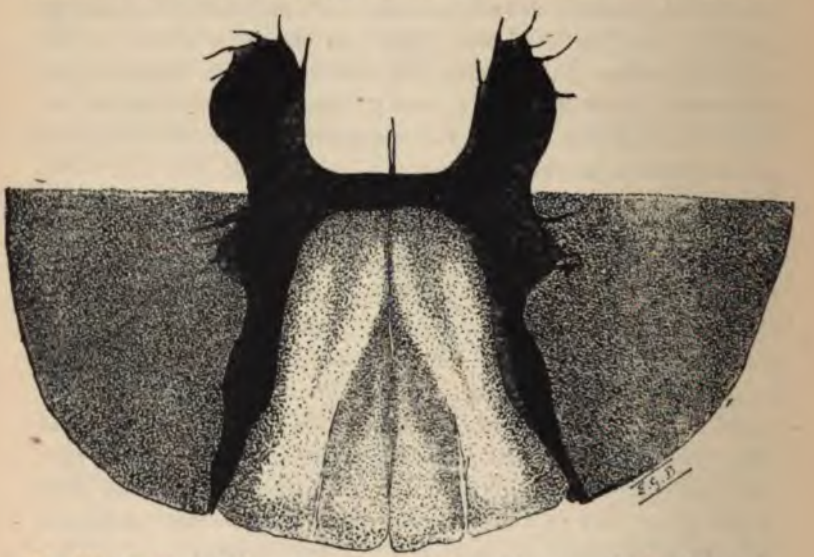


Fig. 191.—Section of the cervical region of the cord in a case of tabes (the parts are of a lighter colour in proportion as they are more affected by sclerosis). If the figure is turned round in such a way that the posterior horns are at the upper part, it is seen that the part which is chiefly affected by sclerosis has clearly the form of the letter M. The band of healthy tissue which separates the outer from the inner stroke of the letter seems to occupy precisely the same region as that in which the comma-shaped degeneration of Schultze is found to occur in some cases of descending degeneration. The posterior part of the column of Goll and the middle portion of the column of Burdach are diseased this case; the external bandlet, properly so-called, is unaffected.

exclusively unaffected. It is probable that the same occurs in the cases of tabes which complicate general paralysis of the insane, since in the diagrams which illustrate such cases and are furnished by M. Westphal and Flechsig (figs. 181, 182, 183), it will be observed, gentlemen, that in the sections of the cord above the lumbar region the lesions are not seated in the external bandlets but in the column of Burdach and in the column of Goll.

There is then, in my opinion, one theory without which the pathological anatomy of tabes cannot be described; this is the important fact that the *lesions of the spinal cord in tabes occur by*

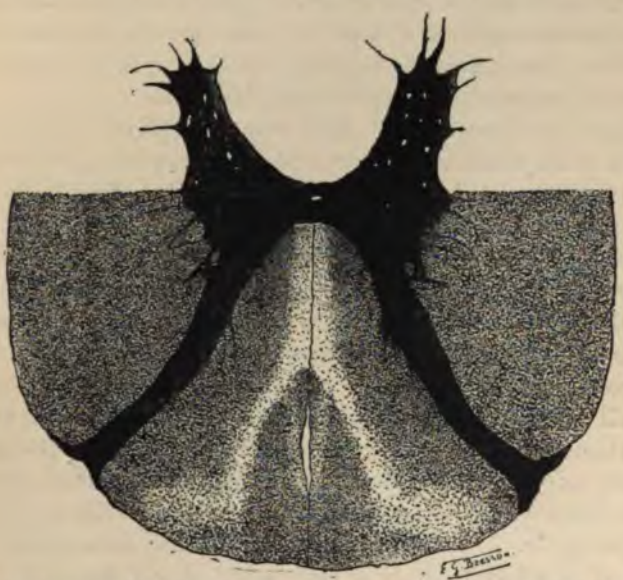


Fig. 192.—Section of the spinal cord (upper part of the cervical region) in tabes. The anterior half of the column of Goll is alone affected by sclerosis in this case; the general form of the region affected is that of a Y reversed.

segments, each posterior root bringing into the posterior column a fresh contingent of degenerated fibres.* The great difference which exists, gentlemen, between two patients suffering from tabes indiscriminately selected both as regards the clinical history and pathological anatomy of the disease, can be thus easily

* As regards the different segments of the cord, it is specially in those constituting the lumbar and cervical enlargements that the lesions of tabes occur.

understood; the same pair of nerves has not been affected by the primary lesion in the two cases. In the one *tabes has occurred in the lower limbs* and abdominal viscera; in the other, *cervical tabes* exists; in a third, the chief signs presented are those of *cephalic tabes*. This difference results from the fact that in the first the roots of the cauda equina, in the second those of the cervical enlargement, in the third those of the cranial nerves were specially and primarily affected.

These three principal segments often suffer in almost a parallel manner, though at the same time it must be remembered that one or other may be affected, if not quite exclusively, at any rate with such a predominence of the symptoms, both from a clinical and anatomical point of view, that the constitution of the three forms, viz., the lumbar, cervical, cephalic, which have been just mentioned is undoubtedly legitimate.

When the existence of this extreme difference, when the origin of the morbid process is remembered, you will be perhaps surprised that the lesions in tabes are usually symmetrical upon one side of the cord or the other. This symmetry can undoubtedly be in some degree explained by the admission, as I have already had occasion to show you in connection with the spinal cord of patients in whom an amputation had been performed, that a portion of the root fibres on one side passed into the posterior column of the opposite side.

This symmetry again should not surprise you, since we are accustomed to meet with it in affections having an anatomical substratum of a most dissociated and peripheral nature, namely, toxic forms of paralysis (lead poisoning, alcoholism, arsenical poisoning, &c.). The exposition of all these facts, gentlemen, is now concluded, and, I may add, of all the hypotheses connected with them.

Allow me to state rapidly, at the end of this lecture, the idea which I have myself formed as to the nature of tabes. From an anatomical point of view:

The lesions of the spinal cord in tabes are by no means the result of primary myelopathy and disease of the posterior columns. They are merely the result of degeneration which has occurred in the posterior nerve roots.

This degeneration of the posterior root fibres is due to change in the cells of the spinal or peripheral ganglia.

This change in the cells which is the *primum movens* of the lesions which occur in tabes, how and why are they produced? You will remember, gentlemen, what I have already said with regard to the ætiology of tabes as to the rôle played by *syphilis*. It is to the syphilitic infection that the cellular changes must be attributed. But by what means? This is no longer a case of new syphilitic growths, presenting or not the appearance of circumscribed or diffuse gummata; nor is it the interstitial tissue of the nervous system which is affected, but the nervous substance itself, the cell.* This mode of action differs considerably from that which you have been accustomed to find occur in syphilis. The progress which has been recently made in the knowledge of infectious diseases enables us, however, if not to solve, at any rate to propose a rational solution of the problem. This solution I shall borrow from Strümpell. In my opinion, we cannot help admitting, in accordance with that distinguished physician, that syphilis acts as a real organic poison, a *toxine*. It is only in this way that we can explain the fact that cells, as distant from each other as are those of the spinal and peripheral ganglia, are simultaneously affected by the same morbid process, and in the same special elective manner.

This is certainly a hypothesis, but, as I observed when commencing this series of lectures upon the pathological anatomy of tabes, everything connected with it is still in a condition of uncertainty and doubt. Fortunate are those who believe in the existence of medical questions which are "completely understood," and that tabes is one of these.

* It will be observed that although the nerve cells of the sensory organs (the skin, tendons, &c.) and of those connected with special sensation (the eye, ear, &c.) are specially involved in the toxo-syphilitic process which causes tabes, the nerve cells of the motor system seem in some cases to undergo similar change (some forms of amyotrophy, hemiatrophy of the tongue, &c.). The cells of the *brain* may also be affected, as is shown by the degeneration of the nerve fibres of the convolutions which occurs in some cases (general paralysis of the insane, cases observed by M. Jendrassik).

LECTURE XXX.

FRIEDREICH'S DISEASE.

HISTORY.—The first cases were described by Friedreich in 1861; in England, the case of Carpenter (1871), of Gowers (1880); in France, the thesis of Brousse (1882); Lecture of Charcot; thesis of Soca. SYMPTOMS.—A. MOTOR DISORDERS:—(a) *Disorders of gait.* (b) *Difficulty in maintaining the upright position.* (c) *Ataxic tremor.** (d) *Choreiform movements.* (e) *Paralytic symptoms.* B. SENSORY DISORDERS:—(a) *Pains.* (b) *Anæsthesia, analgesia.* (c) *Disorders of the muscular sense.* C. DISORDERS OF THE REFLEXES:—(a) *Cutaneous reflexes.* (b) *Tendon reflexes.* D. DISORDERS IN THE ORGANS CONNECTED WITH THE SENSES:—*Ocular disorders; nystagmus.* E. CEREBRAL DISORDERS, *vertigo; cephalalgia; state of the intellect; disorders in speech.* F. GENITO-URINARY DISORDERS. G. TROPHIC AND VASOMOTOR DISORDERS:—*Club-foot; muscular atrophy; curvature of the spine.* Course of the disease: Progressive, remissions at times occur; recovery impossible. DIAGNOSIS from:—*Tabes, insular sclerosis, chorea of Sydenham,* a *pseudo-disease of Friedreich* recently described by Nonne.

GENTLEMEN,—Having studied in detail both insular sclerosis and tabes, we are now in a position to consider the description of another disease of which some of the symptoms exist in both these affections, and which, while it is termed by some hereditary ataxy, and considered by others as a part of insular sclerosis, actually bears the name of "*Friedreich's disease.*"

A simple glance at the disease will enable us at once to recognize its similarity to these two affections.

It consists in the existence of ataxy, which is slowly progressive.

Its onset occurs at an early age, sometimes in many members of the same family; the motor trouble exists first in the lower limbs, the muscles seated in the upper extremities or at a higher level only being affected at a later period. In confirmed cases there is increasing difficulty in walking, and the

* This form of tremor, which only occurs when a movement is attempted, is called by the French "tremblement intentionnel." Gowers (Dis. of Nerv. Syst., Vol. II., p. 549, Note) truly says that this expression cannot be used in English "on account of the meaning of 'intention,'" and that "atactic tremor" would be, on the whole, the best term. (*Translator.*)

gait resembles that due to cerebellar disease and to tabes, in addition to which choreïform instability exists. The articulation of words is difficult and nystagmus exists, but none of the pupil symptoms which are observed in tabes or insular sclerosis. The patellar tendon-reflex is completely lost, but no sensory disorders or lightning pains exist.

The discovery of this affection is undoubtedly due to one of the most eminent clinical observers of German origin, Friedreich, who, as you know, was at the head of the School at Heidelberg, from which so many distinguished physicians have come. In 1861, at the Spire Congress, Friedreich presented several patients who were suffering from a singular form of ataxy, of which the character was not at all in accordance with the masterly description which Duchenne of Boulogne had just given (1858—1859) of locomotor ataxy. The German observer, after observing these cases, came to the conclusion that the description of Duchenne was inexact, and, above all, incomplete, since it did not apply to such cases as these. In a word Friedreich believed this to be purely and simply a form of locomotor ataxy.

In France, during a long period, these facts were ignored, and, owing to the influence of works devoted by Charcot, Vulpian, Bourneville, and Guerard to the study of insular sclerosis, the cases of hereditary ataxy were believed to belong entirely to that disease.

In England, Carpenter in 1871, and Gowers in 1880, had published similar cases; the disease was always "hereditary ataxy," this, however, being but a form of locomotor ataxy.

The condition of the question remained thus, and I have a special reason for remembering it, since, in a memoir which I wrote in August, 1882, for the competition in connection with resident hospital appointments, I endeavoured to draw up, according to the description given by authors, a clinical pictu:e of this affection, with which I was not myself acquainted, which no one in France had as yet observed, and which I also considered as a hereditary form of tabes. In the same year (1882) an excellent thesis was written by Brousse, a thesis composed under the influence of Grasset, and in which the author clearly expressed his opinion that this was an independent affection; considerable progress was thus made,

and Brousse proposed to call the affection by the nam
"Friedreich's disease."

The first case observed, or at any rate recognized, in P
was that of a young lad who had been admitted into the ser
of Charcot in 1884 at a time when I had the honour of be
his clinical assistant. I still remember the circumstances co
nected with his admission, which I shall mention in order
show you what was known at that time about the disease. Th
boy was brought to Charcot by a physician attached to one o
the hospitals, who was most interested in medical progress,
well-informed, and, if I may use the expression, quite on the
watch for any new discovery. This distinguished physician had
intended to exhibit the patient at the meeting of a learned
society in order to prove a fact, which was denied by some
authors, that tabes might occur in children. Before doing this,
and owing to a feeling of scientific scrupulousness which can-
not be too highly praised, my distinguished colleague wished to
have the advice of some one who was well authorized to give an
opinion upon this disease, and he consequently brought the lad
to the Salpêtrière Hospital. As was naturally the case, Charcot
at once recognized from what disease the boy was really suffer-
ing, and a few days subsequently devoted a lecture to the study
of this patient, the permission to do so being readily given by
the physician who had asked for his advice, and who was but
too pleased to escape from the error into which he had so nearly
fallen; this lecture definitely established the separate existence
of *Freidreich's disease*.

Since that time many works and reviews (Rütimeyer, Musso,
Massalongo, &c.) have been written about this disease, numerous
cases have been published, and the disease of Friedreich now
constitutes a well-defined malady. Amongst these works I must
specially name those written in the French language by Gilles
de la Tourette, Blocq and Huet, the very remarkable thesis of
Soca (1888), and a critical review by Ladame (of Geneva); and
I shall have in fact to borrow frequently from these different
authors.

When the pathological anatomy of this affection is discussed
I shall have to mention other works to you.

The SYMPTOMS of the disease of Friedreich affecting several
different systems should be described in somewhat methodical

order, and I shall follow that which is usually employed, it being at the same time understood that this classification is purely artificial.

A. *Motor disorders.*—These may be said to constitute the chief

Fig. 193.—Boy aged six years, brother of the patient who forms the subject of the next figure, suffering from the disease of Friedreich. (After R. Massalongo.) Incо-ordination very pronounced, the attitude of the legs, arms, and head will be observed.

group of symptoms, and to be those to which the attention of the patient and that of the physician are specially called.

(*a*) *Disorders of the gait.*—In confirmed cases these troubles are

very pronounced, and the patient reels in walking, the legs being wide apart, the steps irregular, awkward, and often re-

Fig. 194.—Girl aged ten years affected by the disease of Friedreich. (After R. Massalongo.) The attitude of the patient clearly indicates the inco-ordination and difficulty in maintaining the upright position which exists.

sembling those of an intoxicated person; it does not resemble the gait which exists in locomotor ataxy, since the movements

are neither so violent, so sudden, or so unreasonable as in tabes, while at the same time the unsteadiness somewhat exceeds that which is due to cerebellar disease alone; with this fact is associated a certain degree of inco-ordination which does not exist in the latter case; thus the disorder of the gait occurring in Friedreich's disease is termed *tabid-cerebellar*. The head during this time presents a series of unsteady movements, which are very analogous to those which occur in insular sclerosis.

(*b*) *Difficulty in maintaining the upright position.*—This difficulty was well perceived by Friedreich, and described by him as *static ataxy*. It is not easy for the patient to maintain the upright position, even when the limbs are separated; he is obliged to alter the position of his feet from time to time in order to preserve his equilibrium, the whole body being subject to irregular movements, while his head is affected by a somewhat long series of slightly pronounced movements of salutation. The symptom of Romberg is usually, but not always, absent.

(*c*) *Atactic tremor.*—This tremor does not occur by any means in every case, but when it exists exactly resembles that of insular sclerosis. Having discussed this symptom at some length, and while speaking of that disease, it seems to me unnecessary to repeat what was said upon that occasion.

(*d*) *Choreïform movements.*—These movements are "abrupt," somewhat resembling those which occur in the chorea of Sydenham. Usually, however, they are not so long, and occur not only in the limbs, but also in the face and neck, contributing to give to the patients an appearance of instability which is really characteristic of the disease.

(*e*) *Paralytic symptoms.*—Is there in Friedreich's disease a more or less pronounced degree of muscular paralysis? A slight difference of opinion exists upon this point, though for my part I certainly agree with Musso and Soca in thinking that such is the case. There is no difficulty in recognizing the fact that the affected muscles are absolutely inactive, but it seems to me indisputable that the strength of certain muscles is also much diminished, and that these are the seat of somewhat pronounced paresis. Such is the case, though it must be well understood, in some patients, only as regards certain muscles of the legs, trunk, shoulders, and probably of the upper limbs.

B. *Sensory disorders.*—These, differently from what occurs in

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B. *Sensory disorders.*—These, differently from what occurs in

tabes, are rare, and but slightly pronounced when they do exist:—

(*a*) *Pains.*—These are not felt in Friedreich's disease; Charcot, however, has shown that they exist in certain cases, and that their character may resemble that of the *lightning pains* in tabes.

(*b*) Anæsthesia and analgesia similarly but seldom occur; at the same time some authors, specially Soca, believe these disorders to be somewhat frequent, and that the reason for their being so seldom observed is that they pass unperceived from being but slightly accentuated. In some cases, on the other hand, absolute hemianæsthesia exists, the cause of this symptom being hysteria. This affection is sometimes associated with Friedreich's disease, in the same way as we have seen it occur in the course of insular sclerosis or tabes; in such a case care must be taken not to attribute to disease of the spinal cord what is really the mere effect of this neurosis. Dejerine, however, observed cases in which, as he says, the sensory disorders could not be attributed to hysteria.

(*c*) *Muscular sense.*—According to most authors the muscular sense is usually unaffected, at any rate as regards the knowledge of the posture of the limbs, since the symptom of Romberg, when it exists, seems due to the choreïform instability, rather than to any disorder of the muscular sense. At the same time Soca, owing to some facts observed by him, is of opinion that the latter is sometimes affected. These symptoms, however, are of such a complex nature that in my opinion it is very difficult to apportion them correctly, and nothing definite can be asserted as their cause.

C. *Disorders of the reflexes.*—(*a*) The cutaneous reflexes are usually retained; in some cases they have been found excessive. It does not seem, however, that there is ever anything characteristic in their nature.

(*b*) The tendon-reflexes are almost always lost, and it is this loss which conduces to give such a singular clinical appearance to Friedreich's disease, which consists in a mixture, as it were, of the symptoms of tabes with those of insular sclerosis. Simple diminution of the tendon-reflexes occurs in some cases. In others they are observed to be increased. Is it possible that some mistake has occurred with regard to these cases which are

not really examples of Friedreich's disease? or should this excess be attributed to the fact that the lateral columns of the cord are more affected by the morbid process than is usually the case? The first suggestion seems to me by far the most probable, and I should recommend you to have some doubt with regard to a supposed case of Friedreich's disease in which the tendon-reflexes are excessive.

D. *Disorders in the organs connected with the senses.*—These, properly speaking, do not affect the organs themselves, but their appendages, specially the muscles if any are present. This is another point of difference between this disease and tabes, analogous to that which consists in the infrequency of sensory disorders during the course of Friedreich's disease.

(*a*) *Ocular disorders.*—The most frequent and interesting disorder is:—

Nystagmus.—This in fact exists in most cases, but it must be known that as a general rule it occurs at a late period of the disease: you will only meet with it in fact two, three, or even more years after the onset of the disease. As in the case of insular sclerosis, this nystagmus is not accentuated, and may even disappear during the time of rest, but from the moment that the patient fixes an object, and specially when he is obliged to make an effort to do this (the object being placed laterally), the nystagmus at once occurs or is considerably increased.

Paralysis of the ocular muscles with or without diplopia may exist, and Joffroy has published such a case; you must not expect however, gentlemen, to meet with disorders of this nature in Friedreich's disease in the same way as in tabes; they are as frequent in the latter as they are rare in the former.

The *optic nerve* and *acuity of vision* which are so often and so severely involved in tabes, are on the contrary unaffected in Friedreich's disease.

No pupil symptoms exist; the size of the pupil undergoes its normal changes, and the symptom of Argyll Robertson is absent.

(*b*) No special observation need be made as to the power of tasting, hearing, or smelling.

E. *Cerebral disorders.*—Vertigo occurs somewhat frequently, existing at times almost permanently, while in other cases it comes on in well-marked paroxysms.

Cephalalgia has been observed in some cases, and may present the same characters as in migraine.

The *intellect* may be considered as almost unaffected; this fact must be well remembered since it will surprise you, gentlemen, when you are in the presence of these young patients. On account of their strange, unstable, and frequently stupefied appearance you will be tempted to believe them endowed with much less intelligence than a healthy child. In this, however, you will be mistaken, since if you question the parents or the children themselves you will find them to be almost as advanced in knowledge as other children of the same age. Such is the case in childhood. As regards the adult, those suffering from this disease who survive childhood are perhaps somewhat less intelligent than their healthy companions. My own observations have led me to think that their mental development is more or less imperfect, although they cannot be said to suffer from idiocy, or even backwardness. Some of them can never learn to write, but the cause of this is really the difficulty which exists in using the muscles.

The *speech* presents considerable changes in confirmed cases which are due to derangements in the motor innervation of the muscles by means of which articulation occurs. The speech is slow, hesitating, and at the same time unequal (in the same phrase some words may be spoken by the patient more rapidly than others). In addition to this the pronunciation is thick, and at times indistinct. At the same time it is not the spasmodic utterance which exists in most cases of insular sclerosis. If I had to make a comparison, I should rather say that the speech resembles the "cerebellar gait" since, like that symptom, it is heavy, uncertain, and wavering.

F. *Genito-urinary disorders.*—These are very slightly pronounced, this being another difference between Friedreich's disease and tabes; at most nocturnal incontinence of urine exists to a slight degree in very rare cases. As regards genital disorders, properly so-called, Soca, who has carefully sought for them, states that they do not exist, while impotence is never observed either in the male or female, the only derangement being more or less marked delay in the existence of the sexual instinct in the male, or of menstruation in the female.

G. *Trophic and vasomotor disorders.*—There is no trophic disorder of the skin which I need mention to you, and I may specially state that nothing exists in the foot resembling the perforating ulcer which is so frequent in tabes. I should, however, call your attention to certain singular symptoms which may be as well ranged under the head of the trophic disorders as by any other term.

A special form of club-foot is often observed in patients suffering from confirmed disease of Friedreich. This symptom consists in a deformity which recalls talipes equinus, the foot being shorter than when in a healthy condition, the metatarsus large, while the whole organ seems to be compressed in the antero-posterior direction; upon observing the foot from the side it is seen to be much curved on the plantar surface, whilst the prominence of the dorsal surface is excessive. In addition to this the foot is "claw-like," on account of the forced extension of the toes; notwithstanding this, voluntary extension is possible to a pronounced degree, the toes being then in a position of extreme extension, which causes them, as has been observed, to have a similar appearance to that which they present in cases of athetosis. These deformities are bilateral, and partly disappear when the patient is in the upright position. It must be observed, gentlemen, that this occurs at a comparatively early period in the disease, the first indication of its occurrence being retraction of the great toe, and subsequently of the others; thus in those families of which Friedreich's disease affects many members, the sign which is believed by the parents to indicate that one or other of their children suffers from it is precisely this elevation of the great toe. To what are these deformities due? It is extremely difficult to express an opinion in this respect; one thing is certain, namely, that not only the muscles, but also the bones of the foot participate in the changes which produce this condition.

Muscular atrophy sometimes occurs, and affects different muscles; in the cases which I have had the opportunity of seeing, the muscles of the shoulder girdle and pelvis were specially affected; I cannot, however, describe what occurred in detail. Joffroy and Dejerine have observed patients similarly affected, and in those named by the latter author

Fig. 195.—Feet of a patient suffering from Friedreich's disease in whom club-foot existed in a most severe form upon both sides. The extreme extension of the toes, specially of the great toes, and the excessive curve of the sole and back of the foot will be observed. This figure represents the foot of a patient under the care of Charcot; my good friend, P. Richer, was kind enough to make the sketch specially for these lectures. I gladly express my sincere gratitude for it.

the atrophy was seated in the peripheral segments of the limbs.

Considerable lateral curvature has been observed in most cases of confirmed Friedreich's disease, this being a symptom which sometimes occurs at an early period. It consists usually of scoliosis, which is most often found in the dorsal region, and may be accompanied by more or less pronounced lordosis in the lumbar region.

Such are the symptoms which cause Friedreich's disease to have a special character. As regards the order in which they occur, and the COURSE of the disease, the following observations have been made by different authors:—

The first symptom which attracts the attention of the patient or his acquaintance is almost always the disorder in the gait, being sometimes, however, that of the speech or the scoliosis. In those families in which it is feared that each fresh child may be affected by disease of the spinal cord, and in which the child is constantly watched for the occurrence of the different symptoms, extreme extension of the great toe, or the absence of the patellar tendon reflex are most often sufficient to indicate the onset of the disease in any of the children in which these symptoms occur.

However this may be at the end of a few, usually from three to five years, the ataxy which exists in the lower as well as the upper limbs is quite characteristic; this again continues to increase both in the hands and feet, so that not only is the patient unable to walk any longer, but also has great difficulty in using his fingers; he is then *confined to his bed*, this being a second period of the disease.

The condition now remains the same, and the patient is unable to leave his bed or arm-chair during fifteen, twenty years, or even more. Death occurs from the effect of some intercurrent disease and not on account of the progress of the disease in the spinal cord.

Recovery is a termination which is absolutely unknown, whatthe treatment adopted or age of the patient may be. At the same time, *remissions*, which may be of somewhat long duration, sometimes occur. On the other hand, in some cases sudden and serious aggravations occur.

I do not believe that the affection has ever been known to be

arrested. The course of Friedreich's disease must therefore be regarded as *essentially progressive*.

I have been careful, while describing the principal symptoms of Friedreich's disease, to point out the analogy which some of them have, either with those of tabes or of insular sclerosis, as also the differences which exist between them. The diagnosis of Friedreich's disease has, in fact, specially to be made as regards these two affections. I must also point out to you the characters which distinguish it from the chorea of Sydenham, since I have seen distinguished physicians mistake these two affections for each other.

Tabes differs from Friedreich's disease in the fact that the inco-ordination has not the same character, the movements being neither so extensive nor so abrupt; in addition to this they are almost as pronounced in the upper as in the lower limbs, this only occurring in very advanced cases of tabes. Irregular choreïform movements also occur more frequently and to a much more pronounced extent in Friedreich's disease than in tabes, and it is owing to these movements far more than from true inco-ordination that the symptom of Romberg exists in the former affection.

We have also seen that the gait in Friedreich's disease is of the tabid-cerebellar type, that is to say, with the inco-ordination which exists in tabes, the tottering occurs which is analogous to that which is due to cerebellar disease. In Friedreich's disease again, as you know, sensory symptoms and disorders connected with the organs of the senses, notably with the pupil, are almost entirely absent; these, on the other hand, form the basis of the clinical symptoms in tabes. As regards the nystagmus which is so frequent in the former affection, some authors have stated that it may also occur in tabes. This fact, however, is far from being generally admitted, and even should this combination be possible it must very infrequently exist (Mobius). Neither visceral derangements nor trophic disorders of the skin or joints are observed in Friedreich's disease; on the other hand, scoliosis and special disorders of the speech often occur. As regards the club foot, which is met with in both diseases, its character is quite different in one and the other. Lastly, Friedreich's disease occurs in children and young people while tabes is rarely

observed before the age of twenty-five years. The first of these two diseases is essentially a family affection, which is by no means the case with respect to the second.

As regards *insular sclerosis*, I have already sufficiently insisted upon the numerous ways in which it resembles Friedreich's disease; I must now show you that the points of difference are even more numerous. In the former the disorder of the gait is specially of the spasmodic cerebellar, in the latter of the ataxic cerebellar type; in the latter again choreïform irregularity exists, which does not occur in the former.

Lastly, as you have seen, neither ocular paralysis nor optic neuritis, which are so frequent in insular sclerosis, exist in Friedreich's disease, whereas the loss of the tendon reflexes, the scoliosis, and club-foot, which are almost constant in the first of these two affections, do not occur in the second.

The chorea of Sydenham has been known, as I have already said, to give rise to erroneous diagnosis in some cases; certain features are in fact common to these two affections, such as the so-called choreïform unstableness, and the alteration in the voluntary movements both of the limbs and face, while both affections are, so to speak, the appanage of childhood.

With some attention, however, nothing is more easy than to distinguish these two diseases, since in chorea the movements are more extensive, and usually more pronounced in the upper than in the lower limbs; neither club-foot nor nystagmus occur, the patellar tendon reflex is retained, and the symptom of Romberg is absent.

I may perhaps be allowed to mention upon this occasion the cases recently described by Nonne,* in which inco-ordination of movement and disorders of speech analogous to those which exist in Friedreich's disease were observed to occur in many members of the same family; this affection only occurred at puberty. At the autopsy of one of these cases Nonne did not find the sclerosis to exist, which is characteristic of Friedreich's disease, but only pronounced tenuity (gracilité) of the spinal cord and some other parts of the nervous system. What is the

* Nonne. Ueber eine eigenthümliche familiäre Erkrankungsform, &c., Arch. f. Psyc'., XXII., p. 283.

nature of this affection? We are completely ignorant, but whilst waiting for more information upon this subject I thought it my duty not to leave unnoticed facts recorded by so distinguished an observer, this being perhaps the indication of some fresh neuropathic condition.

LECTURE XXXI.

FRIEDREICH'S DISEASE (*continued*).

ÆTIOLOGY: It is a *family affection*.—What is the rôle of the pathological antecedents in the parents or ancestry? What is that of syphilis? The onset of Friedreich's disease nearly always occurs in childhood, and very rarely after the age of 16 years; Soca's law; is slightly more frequent in the male sex. Pathological anatomy: tenuity (gracilité) of the spinal cord. Sclerotic lesions: A. In the POSTERIOR COLUMNS as the *columns of Goll and of Burdach*. B. In the DIRECT CEREBELLAR TRACT. C. In the LATERAL COLUMN (it would not be the pyramidal fibres which are affected). D. In the ZONE OF LISSAUER. E. In the GREY MATTER: *columns of Clarke*, their nerve reticulum is scanty, their cells are in smaller number; *posterior horns* diminished in size; anterior horns also somewhat altered. The central canal of the cord may be the seat of different lesions. The condition of the *meninges*, the *posterior roots*, and *peripheral nerves* is the subject of very divergent opinions on the part of different observers. Nature of Friedreich's disease. Opinion of Dejerine and Letulle, Pitt and Grasset. Therapeutics: its want of success.

GENTLEMEN,—You will have noticed, owing to what has already been said, that from a clinical point of view Friedreich's disease is clearly distinct from the affections with which one is at first sight inclined to connect it. It has also its own special ætiology, as you will soon perceive.

It is in fact a *family disease*, that is to say, a disease which affects many members of the same family. If you read the account of the observations published by different authors you will find that three, four, or five brothers and sisters often suffer consecutively from this disease. It also appears that in some cases the disease is found to have occurred in the ancestry, and a father, mother, and several of their children may be found to present the symptoms of this malady; at times the morbid tendency skips a generation and a person apparently healthy, but in whose father or brother this form of inco-ordination existed, is the father of children who are thus affected. These cases, however, are very rare, if they exist at all, and most usually, as I have just said, its co-existence in many brothers and sisters alone stamps it as a "family" disease. This notion

had already been held by Friedreich, who recognized its great importance, as shown by its prominent place in the term *hereditary ataxy* proposed by him.

It must not be thought, however, gentlemen, that this family character is absolutely necessary; you will in fact meet with isolated cases, one child may be alone affected in a family, whilst its brothers and sisters remain in perfect health. Soca observes in connection with this fact that such isolated cases specially occur in small families, and suggests that if the number of children was greater in them some would probably have suffered from this disease.

This is very possible, but at any rate the fact is certain that Friedreich's disease does not necessarily occur in all the children of the family when one of them is affected.

As regards the pathological antecedents and their place in the ætiology of the disease nothing definite is known. In connection with the parents more or less pronounced diseases of the nervous system are somewhat frequently found to exist, but without any definite signification, and varying in nature from some kind of neurosis to the most various organic diseases.

Owing to what you now know of the ætiology of tabes you will naturally ask, gentlemen, whether *syphilis* does not also play a part in that of Friedreich's disease. No long time has passed since I in the same way asked myself that question, and when any opportunity occurred I endeavoured to ascertain whether it might not be simply and purely due to hereditary syphilis. I questioned the parents carefully, but came into possession of no facts which indicated such a connection; in the observations, again, which have been published by different authors you will rarely find that any specific infection of the parents is known to have existed. We are not, therefore, at the present time authorized to suppose that this disease is connected with syphilis.

There is little to be said as regards the affections from which the patients are liable to suffer previously, except that in some cases Friedreich's disease has seemingly occurred after some infectious disorder such as measles, scarlatina, or small-pox. There is but little probability that they are directly connected with each other as cause and effect on account of the hereditary nature of the disease; at the same time it is quite probable that

the disease is so influenced by the infectious disorder that its symptoms appear at an earlier date in consequence of its existence. The fact in itself is by no means extraordinary, and the same occurs in another affection which is essentially hereditary, myopathic atrophy, of which the onset seems at times to be also connected with one of the infectious diseases of childhood.

At what age does the onset of Friedreich's disease occur? I have already had occasion several times to say that this is specially a disease of childhood, not only of *puberty*, as Friedreich supposed, but also of *childhood*, and in 76 cases of which the age was known in not less than two-thirds, the disease had begun before the age of fourteen years; in some, in fact, the first symptoms had been observed during the first months of life. On the other hand the onset very rarely occurs after the age of sixteen years.

As regards the age at which the onset occurs Soca has made a remark which he has enunciated in the form of a law in the following terms: "In the same family the onset of Friedreich's disease occurs at the same age in the case of each member of the family who is affected by it"; each family is thus attached by Friedreich's disease in a special manner, not only as regards the age of the onset, but also it appears owing to certain clinical features being observed in some families and not in others.

The *male* is affected somewhat more frequently than the female sex (in the proportion of 68 to 47—Soca), the contrary opinion having been held at first by Friedreich.

PATHOLOGICAL ANATOMY.—This branch of the study of Friedreich's disease is, I must admit, gentlemen, by far the least advanced; few autopsies have been made as yet, and the results have not been always in accordance with each other. I will endeavour, however, to describe the different lesions which have been found in this disease to you, but as I would again say, this description should be considered as simply an attempt which recently published works will assist me to make.

A feature which should be first mentioned is the *tenuity* (*gracilité*) of the spinal cord; this change can be at once recognized to exist by the naked eye, since its diameter does not exceed the $\frac{3}{4}$ or even $\frac{2}{3}$ of that which normally exists; it seems to be specially pronounced in the dorsal region. At the same

time it is difficult to say to what this is specially due ; possibly different causes exist, such as the disappearance or diminution in size of a large number of fibres in the degenerated tracts, retraction of the part affected by sclerosis, or again some defect in the development of the different tracts.

An examination by the microscope enables a certain number of lesions to be recognized, notably in connection with the white tracts of the columns, viz., the existence of sclerosis in the cord. Let us first examine the seats of this affection :

A. In the *posterior column*. The *columns of Goll* are usually the seat of most pronounced degeneration which can be traced from the lower part of the cord as far as the nib of the calamus

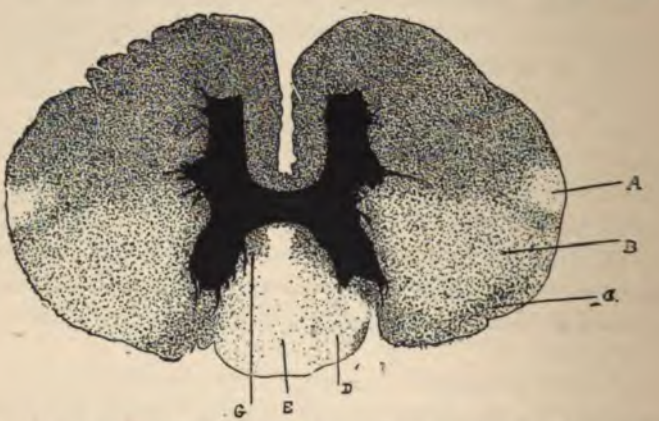

Fig. 196.—Section of the spinal cord in the middle part of the dorsal region from a case of Friedreich's disease. (After Blocq and Marinesco.) A, triangular portion much degenerated, seated in front of the direct cerebellar tract, and which according to Blocq and Marinesco, perhaps, represents the tract of Gowers ; B, lateral much degenerated ; C, direct cerebellar tract ; D, column of Burdach, degenerated ; G, band of sound fibres which adjoins the posterior horn.

scriptorius ; these tracts are thus affected throughout their whole length. The *columns of Burdach* are also involved throughout their whole extent, but in varying intensity according to the region of the cord, the affected part not presenting the same appearance at different heights; their external zone is usually unchanged. The lesion of these columns diminishes progressively as the cervical region is left to disappear altogether in the lower part of the medulla oblongata.

B. *Direct cerebellar tract.* This column is affected from its origin, that is to say, the lower part of the dorsal region; the lesion being more pronounced in the upper part of the dorsal region, while on the other hand it diminishes considerably in the lower part of the medulla oblongata, where but a few degenerated fibres are found to exist in the seat of these tracts. In some observations (Pitt, Rütimeyer, Blocq, and Marinesco) it is clearly indicated that the lesions extend anteriorly beyond the seat of the direct cerebellar tract, and that the tract of Gowers was also affected; I believe that this change may be

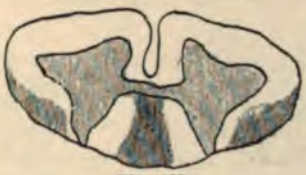

Fig. 197.

Fig. 198.

Fig. 199.

Fig. 200.

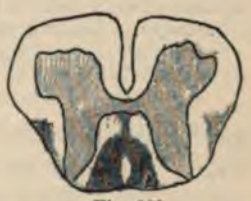

Fig. 201.

Sections of the cord from a case of Friedreich's disease. (Case I. of Rütimeyer.) Fig. 197.—Upper part of the cervical enlargement. Fig. 198.—Region seated between the cervical and dorsal portion of the cord. Fig. 199.—Middle part of the dorsal region. Fig. 200.—Region seated between the dorsal and lumbar regions of the cord.

considered as constant in Friedreich's disease when somewhat advanced in degree.

C. *Lateral columns.*—Most authors regard the lesions of the lateral columns as seated in the crossed pyramidal tracts; I confess, gentlemen, that I have some difficulty in sharing their opinion. I allow that the lesions are seated in the region of the crossed pyramidal tracts, but I cannot readily admit that the affected fibres belong to those tracts. You know, in fact, gentlemen, that different varieties of fibres exist in these tracts (large fibres, narrow fibres); there is, therefore, nothing surprising in the fact of these fibres having a different origin, and

the proof that such is the case is the fact that in cases of extensive lesion in the pyramidal system a certain number of fibres

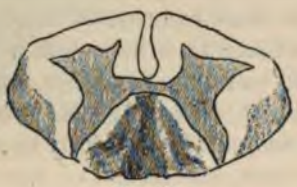

Fig. 202.

Fig. 203.

Fig. 204.

Fig. 205.

Fig. 206.

Sections of the spinal cord from a case of Friedreich's disease. (Case II. of Rütimeyer.) Fig. 202.—Lower part of the cervical enlargement. Fig. 203.—Upper part of the dorsal cord. Fig. 204.—Middle part of the dorsal cord. Fig. 205.—Lower part of the lumbar cord. Fig. 206.—Sacral cord.

are found to be quite unaffected in the middle of the diseased zone. In the disease of Friedreich the following are the reasons upon which I based my opinion that the affected fibres do not belong to the crossed pyramidal tract (FPyC): 1. The lesion in the lateral column diminishes considerably from below upwards as far as the lower part of the medulla oblongata, which would scarcely be the case if the pyramidal tract was affected. 2. In a transverse section of the cord the seat of the lesion in the lateral column does not exactly correspond to that which a lesion in the pyramidal tract would have, occupying a more external portion of the cord. 3. Nothing in the clinical symptoms recalls those which constantly occur when the pyramidal tract is affected. If, then, the pyramidal tract is not involved, which are the fibres, you will ask, whose disease produces this sclerosis in the lateral column? It is difficult to answer this question in a positive manner, but we may, I think, presume that these fibres are connected with the direct cerebellar and anterolateral ascending tracts, which they join together.

D. *The marginal zone of Lissauer.*—Changes in the nerve fibres of this region are not admitted to exist by all the different authors. Rütimeyer and Ladame state that they did not find them, while Letulle and Vaquez, Blocq and Marinesco observed them to exist; the latter authors remark that in the cases seen by them the lesions in the zone of Lissauer only existed in the lumbar region, this part being unaffected in the dorsal and cervical region. In the presence of these diverging opinions it is difficult to form a conclusion; I should at the same time be very inclined to think that this lesion of the fibres in the zone of Lissauer exists very frequently, if not in all cases of Friedreich's disease.

The grey matter is also the seat of different changes:—

(*a*) *Clarke's columns.*—The nerve fibres which constitute the nerve-network of these columns not only disappear to a more or less pronounced degree, as occurs in tabes, but the cells exist in smaller number, are of smaller size, and have usually lost their prolongations.

(*b*) *Posterior horns.*—These in the same way as all other parts of the cord are of diminished size, in addition to which the number of cells which they contain is appreciably less.

(*c*) *Anterior horns.*—Changes have been also found in these by some authors (Friedreich, Rütimeyer). Atrophy of the cells having been sometimes found to exist, these lesions will perhaps explain how it is that in some cases, as I have already said, muscular atrophy is a clinical symptom of the disease.

The *central canal of the cord* is sometimes altered, the signification of this change not being as yet understood; thus it may be *bifid*, or *lateral ectopia*, or "*peri-ependymal*" *lesions* which may occur.

The meninges of the spinal cord do not seem to present always the same appearance; thus some authors assert that they have been clearly thickened in the cases observed by them, while others affirm that such a change exists but slightly if at all. In any case, if such thickening occurs, it is most pronounced.

The *posterior roots* have been also the cause of much difference in the opinions expressed as to their condition; thus, for example, Letulle and Vaquez state that they are affected in an irregular manner, whilst Blocq and Marinesco consider them to be as much changed as in ordinary tabes.

As regards the *peripheral nerves*, still less has been written about their condition than about the changes in the cord. Letulle and Vaquez and Dejerine, supposing, somewhat a priori it may be said, that the peripheral nerves are but little, if at all involved, explain by this fact the absence of the lightning pains and sensory derangements which are usual in Friedreich's disease. Blocq and Marinesco do not agree with this opinion. The truth is that as regards the principal fact, the absence or presence of peripheral neuritis in Friedreich's disease, but little is definitely known. It is therefore somewhat venturesome to suggest hypotheses founded upon a supposition which is still quite uncertain.

Such then, gentlemen, are the lesions shown to exist in the different parts of the nervous system in Friedreich's disease as regards their seat; with respect to their nature, I must make some explanations which will put you on your guard against an erroneous opinion which has been recently circulated.

Many authors had already observed, in describing the lesions which are seen microscopically, that in certain regions of the cord the part affected by sclerosis is found to be clearly formed of long fibrils, plaited in the transverse direction, and on account of this plaiting more or less obliquely placed with respect to each other. Recently, in 1890, Dejerine and Letulle recalled the existence of these "eddies," making them the basis of an actual theory as to the nature of the anatomical process which occurs in Friedreich's disease. Comparing these "eddies" which exist in sclerosis with the lesions described by Chaslin in the cerebral cortex of those affected by epilepsy, lesions considered by that distinguished pathologist as due to changes in the neuroglia, Dejerine and Letulle thought that the change which occurs in hereditary ataxy was special to the disease; they therefore believed themselves authorized to assert "that they had discovered a form of sclerosis affecting merely the neuroglia, the only sclerosis of this nature hitherto known." In support of this view they relied not only upon the "eddies of fibrils," but also upon the sound condition of the septa of connective tissue and blood vessels in the posterior columns. Thus, according to these authors, Friedreich's disease is constituted by sclerosis of the neuroglia alone, being as it were a glioma (sclerosis of ectodermal origin) of the posterior columns. On

the other hand, sclerosis of the crossed pyramidal and direct cerebellar tracts would be always, in the opinion of Dejerine and Letulle, a "vascular form of sclerosis," since in these conditions the eddies of the neuroglia are absent, while sclerosis of the septa of connective tissue, and more or less pronounced vascular changes exist.

This thesis, which was zealously maintained by the above authors, was for a time favourably received by the medical profession, but the neurologists on the one hand and pathologists on the other became united against it. Amongst the first opponents we find Blocq and Marinesco, Achard and Weigert, authors who joined together in order to prove in different ways that there was no foundation for the statement of Dejerine and Letulle.

The following is a brief summary of the arguments which may be considered more or less conclusive against the opinion of Dejerine and Letulle: (*a*) The vascular changes in the posterior columns are not only present, but are at times so pronounced that Pitt was of opinion that Friedreich's disease should be regarded as a vascular form of sclerosis; Blocq and Marinesco again describe numerous changes in the vessels, proliferation of the nuclei, dilatation of the cell spaces, and even obliteration. (*b*) As regards the septa of connective tissue, they are far from being always unaffected, and changes have been recognized in them by some observers. Lastly (*c*), which is a more serious objection, the so-called specific nature of this form of sclerosis, owing to the neuroglia being specially affected in Friedreich's disease, has no true foundation. On the one hand, the "eddies" which exist seem, in my opinion, to be merely an indication that the sclerosis in the nervous centres fulfils the two following conditions: 1. That of being of very long duration. 2. Of having commenced in childhood at the time when the nervous centres were still being developed. (I for my part, in 1884, in a work written conjointly with Jendrássik, showed that the condition of the parts affected by sclerosis in the cerebral convolutions was very analogous to that of the eddies which exist in the cord in Friedreich's disease, and I maintain that this condition is entirely due to the two causes which I have just mentioned.) Certain special methods of staining the sections (Malassez, Weigert) have also shown, as Achard* very truly observes, that

* Achard, *Bulletin de la Société Anatomique*, 1890.

other forms of sclerosis in the spinal cord, such as that which occurs in Pott's disease, tabes, insular sclerosis, and amyotrophic lateral sclerosis, may be quite as rightly called sclerosis of the neuroglia as that of Friedreich's disease. Weigert* goes even farther, and, owing to results obtained by his new method of staining the neuroglia, is able to state that in insular sclerosis the proliferation of the neuroglia is even more pronounced than in Friedreich's disease, and that he is quite unable to understand how the sclerosis which exists in the latter affection can possibly be regarded as a form of glioma.

This prejudicial question having been thus answered, and the theory of Dejerine and Letulle being recognized as erroneous, what opinion should be formed as regards the pathological anatomy of hereditary ataxy? Friedreich looked upon it as the result of arrest in the development of the spinal cord, and believed the lesion in the lateral columns to be due to extension of the change in the pia-mater, which was itself consecutive to the alteration in the posterior columns. Pitt, accepting the opinion that the disease was due to some defect in the development of the cord, states that owing to this fact the fibres which develop at a late period are specially affected in Friedreich's disease, the diseased fasciculi, none of them receiving their sheath of myelin until after the fifth month. Pitt thus admits that the pyramidal tracts take part in the degeneration. Lastly, the opinion of Grasset is again different; according to him Friedreich's disease is a form of combined sclerosis, which, as he considers, is a term applied to a special group of diseases. I shall soon have occasion to express my opinion upon this form of disease, and you will see that it is very different from that expressed by the distinguished professor of Montpelier.

At the end of this chapter upon the pathological anatomy of the disease, I should add that I have only spoken of the lesions in the spinal cord which occur in Friedreich's disease, owing to the fact that they are the only ones as yet known. It is, however, probable that as the study of neurology advances other lesions will be found in the medulla oblongata, brain, and cerebellum, since Friedreich's disease seems to me essentially a cerebro-spinal disease. At the same time, you will allow me,

* C. Weigert, Zur Pathologischen Anatomie des Neurogliafasergerüsts.—Centralblat für Allgemeine Pathologie, 1890, p. 729 et seq.

gentlemen, to be absolutely silent in other respects as regards the nature of the disease, since I have been so far totally unable to form even an approximate opinion upon this subject.

How should Friedreich's disease be treated? Some symptoms, especially the disorder of the gait and inco-ordination, have in some cases been improved by means of *suspension* and *electricity*; *antipyrin*, recommended by some authors, has no beneficial effect. After what you have learnt as regards the origin of the disease, and the way it occurs in families, you will not be surprised to hear, gentlemen, that it is absolutely incurable.

LECTURE XXXII.

COMBINED LATERAL AND POSTERIOR SCLEROSIS.*

HISTORY: Westphal (1877), Kahler and Pick, Strümpell, Raymond and Arthaud, Babes, Ballet and Minor, Grasset, &c.; PATHOLOGICAL ANATOMY—LESIONS: In the POSTERIOR COLUMNS: (*a*) column of Goll; (*b*) column of Burdach. In the LATERAL COLUMNS: (*a*) crossed pyramidal tracts; (*b*) direct pyramidal tract. In the DIRECT CEREBELLAR TRACT; In Gowers' tract; In the GREY MATTER: (*a*) cells of the *anterior horns*; (*b*) cells of the *posterior horns*; (*c*) cells of Clarke's column. SYMPTOMS: very diffuse and variable. Two large groups according as the tabid or spasmodic symptoms predominate. DIAGNOSIS: from *tabes*, *insular sclerosis* and *transverse myelitis*. NATURE of these affections: their classification in different groups. Experience of Stenson; results obtained by Ehrlich and Brieger, by Singer and Münzer; their application to the study of the pathology of combined lateral and posterior sclerosis.

GENTLEMEN,—The symptoms of *combined lateral and posterior sclerosis*, or as it has been also termed, *combined lateral and posterior tabes*, are not such as to specially attract the notice of observers. Thus it is not the clinical aspect of this morbid condition which has specially determined its existence as was the case with respect to the other diseases about which I have already spoken, but its pathological anatomy; you will see, gentlemen, that the foundation upon which this disease rests is less assured on that account.

Although the attention of some authors (Pierret) had already been drawn to certain abnormal forms of tabes, which Erb termed "mixed forms," no precise knowledge existed with respect to this form of combined sclerosis until the work of Westphal upon the subject appeared (1877). In the same year Kahler and Pick devoted a memoir to the study of this disease, to which I shall again have occasion to allude. Since then other works have been written upon this subject, of which those of Strümpell, Raymond and Arthaud, Babes, Ballet and Minor, Dejerine, Babinski, and Charrin, Dana, &c., should be

* This disease has also received the name of "ataxic paraplegia" (Gowers), and ataxo-paraplegic tabes (Dejerine).

specially mentioned. Lastly, that of Grasset should not be forgotten in this enumeration, one of the most complete works which have appeared upon this subject; I am the more anxious to allude to this work, owing to the fact that, as you will see in the course of these lectures, I am far from agreeing with the opinions of that author.

Its pathological anatomy having, as I have already observed, been the special cause of this form of combined sclerosis being recognized, it would be best, in my opinion, to commence by studying what has been written upon this subject.

In order to have a typical example of the disease, though I would not by any means say that this type exists as often as is said to be the case, we will imagine, gentlemen, a spinal cord in which the microscopic examination reveals the simultaneous existence of the following lesions:

A. In the *posterior columns*:—

(*a*) The columns of Goll have degenerated throughout almost the whole length of the spinal cord, as far as their termination in the medulla oblongata.

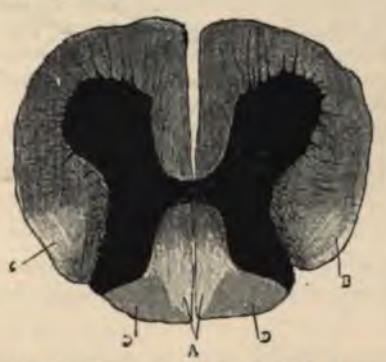

Fig. 207.—Section of the spinal cord from a case of combined lateral and posterior sclerosis (lumbar cord). The clear portions indicate the parts affected by sclerosis; they are of lighter colour in proportion as the sclerosis is more pronounced. A. Column of Goll; B, C. Posterior part of the lateral column near the pyramidal tract; D. Column of Burdach.

(*b*) The *columns of Burdach* are also involved, but usually to a slighter, less complete, and in all cases a more variable extent. According to some authors the lesion in these columns is almost always more pronounced in the dorsal than in the lumbar cord, the seat of the alterations which occur in this column in tabes

being different; you know in fact that in the latter affection it is usually in the lumbar region that the changes are most pronounced.

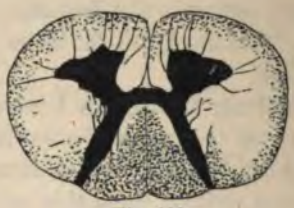

Fig. 208.

Fig. 209.

Fig. 210.

Sections of the spinal cord from a case of combined lateral and posterior sclerosis. (After Westphal.) Fig. 208.—Cervical enlargement. Fig. 209.—Inferior part of the cervical region. Fig. 210.—Middle part of the dorsal region.

B. In the *lateral columns* :—

(*a*) The *crossed pyramidal tract* is not usually, as authors say, involved throughout its whole extent; its postero internal part is usually unaffected: in the same way as regards the seat of the lesions in these tracts, it is observed that most often they can scarcely be said to exist in the upper part of the cervical region, being much more pronounced in the dorsal and lumbar cord.

(*b*) The *direct pyramidal tract* is usually unaffected; in some cases, however, an islet of sclerosis has been found in the fibres which compose it, specially at the internal part of this tract.

C. In the *direct cerebellar tract* :—

This tract is always affected, and to a somewhat pronounced degree.

D. *Gowers' ascending antero-lateral tract* :—

The lesions are usually less pronounced in this than in the preceding tract; at times, in fact, they are but very slightly marked.

E. In the *grey matter*: –

(*a*) The cells of the *anterior horns* may be involved in the morbid process, though this rarely occurs.

Fig. 211.

Fig. 212.

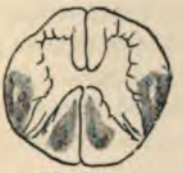

Fig. 213.

Fig. 214.

Sections of the spinal cord in a case of combined lateral and posterior sclerosis. (After Strümpell.) The lesions are indicated by zones of black or grey colour according to their greater or less intensity. Fig. 211.—Lesions in the column of Goll, column of Burdach, the direct cerebellar tract and the crossed pyramidal tract (upper part of the cervical cord). Fig. 212.—The same lesions as in the preceding figure (the middle part of the dorsal cord). Fig. 213.—The lesion in the posterior columns is less pronounced, and tends to recede from the median line (upper part of the lumbar cord). Fig. 214.—The lesion of the posterior columns can scarcely be seen (lower part of the lumbar cord). It will be observed that all these lesions diminish in intensity from above downwards.

(*b*) The cells of the *posterior horns* apparently may or may not participate in the changes which occur in the posterior columns.

(*c*) The cells of Clarke's columns suffer in some cases from more or less pronounced atrophy, but the knowledge which exists about this point is not explicit enough to enable me to make any definite statement.

Such, gentlemen, is a summary of the changes which may possibly be found to exist in the same spinal cord. You will understand what a great variety of symptoms may be produced by such a combination of lesions.

A. There are symptoms of *tabid nature* which consist in—

Inco-ordination, which is indicated by the awkward movements, ataxic gait, and symptom of Romberg. The loss of the patellar tendon reflex; the lightning or other pains; anæsthesia and paræsthesia; the ocular derangements; the disorders connected with micturition; the genital derangements, &c.

B. Conjointly with the preceding symptoms, or separately from them, symptoms of a *spasmodic nature* exist such as —

Excess of the patellar tendon reflex ; foot-clonus ; diminished motor power, paresis or paralysis ; cramps ; contractures ; spasmodic gait, &c.

Such, gentlemen, are, so to speak, a gross inventory of the symptoms which may be observed in cases of this form of combined sclerosis.

You observe what a singular combination of symptoms may exist. Do not, however, suppose that these symptoms are associated together quite indescriminately and without any directing cause. For from this they tend to exist together in groups, so that different clinical forms can be recognized. At the same time this does not imply that the combined form of sclerosis is a distinct morbid condition which may be compared with the "system diseases" of the spinal cord.

I will now mention to you the clinical groups which are most often observed.

The onset may take place by the occurrence of one or other of the following symptoms :—

Spasmodic rigidity, or inco-ordination ; or possibly paresis and cramps, paræsthesia, or pains may exist, the latter being rarely very severe.

The following is what usually occurs :—*inco-ordination* first exists, after which *spasmodic rigidity of the lower extremities* soon occurs ; *early and rapid diminution of genital power ; disorders in the bladder* are soon observed, and a certain degree of *numbness* in the *lower extremities*. As regards *pains*, I have already observed that they are usually of slight intensity and short duration. Gowers observes that while these pains are specially seated in the lower limbs, in tabes they chiefly occur in the lumbar or sacral region in the combined form of sclerosis.

The *ocular disorders* are rare, and the gastric, intestinal, and laryngeal crises scarcely ever exist.

In those cases in which spasmodic rigidity occurs in the lower limbs, the patellar *tendon-reflex* is naturally exc
much more rarely absent, and only when the cli
of the case more closely resemble those of tabes.

The *disorder of the gait* may be ch

tabid-spasmodic; that is to say, it resembles both that which exists in tabes, and that associated with spastic paraplegia.

The *upper limbs* are rarely affected, and if so to a less degree than the lower limbs.

It is in order to recall the principal characters of the affection, which have just been mentioned, that Dana proposes that the name "progressive spastic ataxia" be conferred upon it, and this from a descriptive point of view seems to me preferable to that of "ataxic paraplegia" proposed by Gowers.

The diagnosis must be specially made as regards (*a*) *tabes*, (*b*) the principal forms of *spastic paraplegia*.

(*a*) *Tabes*.—In this disease the complete absence of any spasmodic symptom, the frequence and intensity, as also the seat of the pains (which specially occur in the lower limbs) associated with the loss of the knee-jerk, and the existence of ocular disorders usually enable the diagnosis to be made. In combined lateral and posterior sclerosis again paralysis of the lower limbs exists to a somewhat pronounced degree far more often and always at a much earlier period of the disease than in tabes.

(*b*) *Spastic paraplegia*.—The forms of this disorder, which are found to be distinguished with the greatest difficulty from the combined form of sclerosis, are the following:—

(*a*) That which occurs in *insular sclerosis*. In the latter disease the absence of symmetry, the irregularity in the symptoms, the tremor which occurs, and the disorders of speech will prevent its being mistaken for the combined form of sclerosis.

(*β*) That which is due to *diffuse transverse myelitis*. In this case the grey matter is specially affected; there is more or less extensive muscular atrophy, and disorders often occur in connection with the action of the sphincters; in addition to this it can be recognized that the symptoms correspond to a transverse lesion in the spinal cord, owing to the fact that all the parts above the point at which this exists are totally unaffected.

As regards the ÆTIOLOGY of combined lateral and posterior sclerosis what has hitherto been written upon this subject does not enable us to make any definite statements; all that I can ...at most cases occur in adults between the ages of 20

I have been obliged to present this com- ...ou as a special disease; such is the

most convenient way of describing it. We will now with your permission consider what must be thought of its real nature and the place which it should occupy in a classification of diseases.

According to Westphal, Kahler and Pick, Strümpell, &c., this form of sclerosis is most often due to the co-existence of different types of system sclerosis in the same patient; it would then be in fact a true *system sclerosis*.

The opinion of Grasset is that the "combined form of tabes" is in the same way as tabes dorsalis "a combination of clinical symptoms connected with disease of the spinal cord, since it possesses a clinical and anatomical individuality characterized by the association of a system posterior sclerosis with a diffuse lateral sclerosis; it is thus a mixed form of myelitis."

According to Ballet and Minor the disease usually consists of *diffuse pseudo-systemic sclerosis*, which is very often of vascular nature, and often again due to a process of chronic meningitis (the latter opinion is also that maintained by Dejerine).

In the first place, gentlemen, one thing must be allowed, namely, that it is absolutely impossible to consider all the cases of combined lateral and posterior sclerosis as due to the same morbid process. It is necessary to class these cases in distinct groups, which may be, as you will see, very different from each other.

The CLASSIFICATION is almost the same as that formed by Ballet and Minor in their memoir, and contains four groups:—

I. There is a *combination of different forms of true systemic sclerosis*, which, for example, exists in the following:—

(*a*) In certain cases of general paralysis of the insane in which the lesions in the posterior columns, which are characteristic of tabes, are found to co-exist with very evident degeneration of the lateral tracts, containing the pyramidal tracts (?).

(*b*) Systemic sclerosis of the columns of Goll, columns of Burdach, and the direct cerebellar tracts are found to exist in combination. Cases in which this association exists have as yet been rarely observed, and have not been described in detail; you will remark, however, gentlemen, that the seat of lesions is very analogous to that which I have described as existing in Friedreich's disease.

With regard to these forms of *combined systemic scler*

should like, gentlemen, to draw your attention to an observation in pathological physiology which is most interesting. No one, I believe, has as yet applied it to the study of the lesions which we are considering, and yet it throws much light upon their origin.

You undoubtedly know, gentlemen, the celebrated experiment of Stenson, which consists in temporarily arresting the course of the blood in the abdominal aorta and then re-establishing the current, and observing the disorders in the functions of the spinal cord which result from such momentary arrest of the circulation in the lower part of the spinal axis. Ehrlich and Briger conceived the ingenious idea of making use of this observation in order to examine from a microscopic point of view the lesions which are thus produced in the cord. They noted the fact that extensive degeneration exists at that time in the grey matter and antero-lateral columns, whereas the posterior columns are almost unaffected, except at their anterior border in the part which adjoins the posterior commissure. To this very important fact another may be added, furnished by Singer and Münzer, namely, that a marginal zone which completely corresponds to the regions of the direct cerebellar and Gowers'

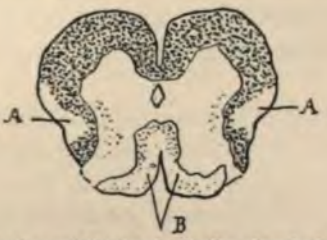

Fig. 215.—Spinal cord of a rabbit in which the circulation of the blood in the abdominal aorta was temporarily arrested (the clear parts in the lateral and posterior columns indicate the regions which are still sound; the black points represent the fibres which have degenerated). (After Singer and Münzer.) It will be observed that the region consisting of the direct cerebellar and Gowers' tract A is completely unaffected. In the posterior columns the number of degenerated fibres (black points) is very small, being specially seated in the vicinity of the posterior commissure and base of the posterior horns; they much appear to belong to that system which we have already considered to be in part composed of commissural fibres.

tract is in a healthy condition. From these researches the following conclusion may be drawn: As regards their nutrition the white columns of the cord may be divided into two groups

LECTURE XXXIII.

COMBINED LATERAL AND POSTERIOR SCLEROSIS (*continued*).

What is known as to the normal anatomy of the ARTERIES in the spinal cord. A. EXTRA-MEDULLARY branches. I. *Anterior system*: Anterior spinal artery; lateral spinal arteries. II. *Posterior system*: Posterior spinal arteries. B. INTRA-MEDULLARY BRANCHES. I. *System of the anterior spinal artery*: anterior median and commissural artery; branches to the anterior portion of the white substance; anterior radicular branches. II. *System of the posterior spinal artery*: posterior medial artery; post-intermediate artery; posterior radicular artery; posterior cornual artery; posterior lateral branches; middle lateral branches; anterior lateral branches. The lesions in certain cases of the combined form of sclerosis are seated round the vessels of the posterior cornua and the lateral arteries; connection of the lesions with the above named vascular regions. ÆTIOLOGY. THERAPEUTICS.

BEFORE entering more deeply into the study of the *pseudo-systemic forms of combined sclerosis of vascular origin*, I must ask your permission to state in a few words the principal facts which are now known about the CIRCULATION OF BLOOD IN THE SPINAL CORD. For this knowledge we are chiefly indebted, as you know, to the studies of Adamkiewicz, the different works devoted to this subject having, as you know, been more or less inspired by him; Duret has also made important investigations in connection with this subject.

The arteries of the spinal cord are derived on the one hand from the *vertebral*, on the other from the *intercostal*, *lumbar* or *sacral* arteries, and pass into the spinal canal through the intervertebral foramina with the anterior or posterior roots. To facilitate the description we will study separately: A. The extramedullary. B. The intramedullary branches.

A. EXTRA-MEDULLARY BRANCHES. I. *Anterior system.*—The *anterior spinal artery* is formed by the union of two descending branches, each of which is supplied by one of the vertebral arteries; these two branches usually join at the upper part of cervical cord. The anterior spinal artery which is thus d corresponds to the anterior median fissure, and can be

traced as far as the conus medullaris of the cord. The two above branches may join at a lower point, or the anterior spinal artery may again divide at certain points into branches which afterwards join to form a single trunk.

The anterior spinal artery is reinforced by many tributary branches from the *lateral spinal arteries* which pass through the orifices in the laminæ of the vertebræ and accompany the anterior roots in order to join the anterior spinal artery. These branches are usually of very small size, and according to Adamkiewicz there are only two or three of those vessels throughout the whole length of the cord which are large enough to be of much importance; the tributary vessels seated in the lower part of the cord are of larger size than those in its upper part.

A series of somewhat large branches pass from the anterior spinal artery at right angles to that vessel from before backwards into the anterior median fissure, constituting the *anterior median arteries*, of which I shall presently have to speak. The *anterior radicular arteries* and some *anastomotic branches* which join the system of the posterior spinal arteries, and contribute to supply the lateral parts of the cord with blood, come from the same vessel.

II. *Posterior system.*—The *posterior spinal arteries* are two in number upon each side; they also come from the two vertebral arteries, and are formed upon each side by two branches, of which one descends behind the posterior roots, namely, the *internal posterior spinal artery*, while the other, the *external posterior spinal artery*, descends in front of them.

These arteries have a large number of branches which pass into the interior of the cord or anastomose with those from the three other arteries; lastly, some branches communicate with those of the anterior system.

The anterior and posterior systems are therefore very distinct from an anatomical point of view, although of common origin; from a pathological point of view, as I shall show you, gentlemen, they are no less distinct; in some affections the anterior while in others the posterior system is involved. This localization of the lesions in a part of the cord supplied by one set of vessels alone should not surprise you, since examples of this kind are by no means rare. Thus, in the pathology of the heart the

LECTURE XXXIII.

COMBINED LATERAL AND POSTERIOR SCLEROSIS (*continued*).

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lesions which involve the coronary arteries alone are known by you; and in that of the lungs there are lesions which, as you know, almost exclusively affect the pulmonary or bronchial arteries. Lastly, in the brain none of you can be ignorant of the very predominant manner in which the region supplied by the middle cerebral artery is affected, although that artery communicates with the other arteries of the brain.

Before commencing, however, to study the different lesions of the spinal cord in connection with the position of the blood vessels I must enable you to understand the seat of the arteries within it.

B. INTRA-MEDULLARY BRANCHES. I. *System of the anterior spinal artery.* (*a*) ANTERIOR MEDIAN ARTERY.—This artery passes from the anterior spinal artery to the end of the anterior median fissure, from which point it passes either to the right or to the left of the anterior commissure; it but rarely divides into two branches (Kadyi). When the commissure is reached the vessel takes the name of commissural artery, which divides into small vessels, which ramify in the anterior two-thirds of the grey matter. This artery specially supplies the *anterior horn* by means of a special branch, which curving round from behind forwards supplies this horn with numerous twigs, and to a small extent (Kadyi) the adjoining white substance.

The branches of the commissural artery are as follows:—(1) *Anastomotic artery.*—This is the most internal branch; it passes either upwards or downwards, and anastomoses with the corresponding branch situated above or below. Thus a vessel is formed in the grey matter at the junction of the posterior commissure, with the neck of the anterior horn which is longitudinally directed and occupies the whole length of the cord; it is this vessel, essentially arterial in nature, which many of the ancient authors have considered a vein and described by the name of *central vein.*

(2) Branch for *Clarke's column.*

(3) Some *small branches* for the *white substance* which adjoins the grey matter, specially that in the neck of the anterior horn.

(*b*) BRANCHES FOR THE ANTERIOR PORTION OF THE WHITE SUBSTANCE, of which the shortest are limited to this part, whilst those which are longer become lost in the grey matter of the

anterior horns into which they penetrate either anteriorly or laterally.

(*c*) ANTERIOR RADICULAR BRANCHES (coming in great part, as we have seen, from the lateral spinal arteries); they accompany the roots into the interior of the cord, giving off twigs both to the white substance which immediately adjoins them and to the grey matter in which they terminate.

II. *System of the posterior spinal artery.* (*a*) POSTERIOR MEDIAN ARTERY.—This artery passes to the end of the posterior median fissure; in its course it gives off small branches to the adjoining parts. This artery is usually of somewhat large size, and is very easily seen in most sections of the spinal cord.

(*b*) POST-INTERMEDIATE ARTERY.—This artery passes between the column of Goll and that of Burdach in the *intermediate groove;* it is contained in a somewhat large septum as indeed are most of the intra-medullary arteries of a certain size.

(*c*) POSTERIOR RADICULAR ARTERY.—Each of these arteries accompanies the corresponding posterior root, and terminates with it in the grey matter of the posterior horn.

(*d*) POSTERIOR CORNUAL ARTERY.—This vessel passes through the column of Burdach on the inner side of the posterior roots, and terminates in the base of the posterior horn.

(*e*) POSTERIOR LATERAL ARTERY.—This branch, which is not quite constant, is distributed to the posterior part of the lateral column.

(*f*) MIDDLE LATERAL ARTERY.—This vessel, usually of some size, supplies the middle part of the lateral column.

(*g*) ANTERIOR LATERAL ARTERY.—The anterior part of the lateral column is supplied by this branch.

The 3 arteries *e*, *f*, *g*, specially the latter, are partly furnished by the anastomotic branches, between the posterior and anterior systems, but are connected with the posterior system of vessels.

Now that we have a general idea, gentlemen, of the blood supply in the spinal cord, we will again consider *combined lateral and posterior sclerosis*, for which purpose this digression was made.

I observed to you, gentlemen, that a large number of cases of this affection are of vascular origin, and this I shall now attempt to prove.

If we place the diagram of a section of the normal spinal cord

in which the course of the vessels is indicated over one representing a section of the cord affected by combined lateral and posterior sclerosis, it will be clearly seen that the parts affected

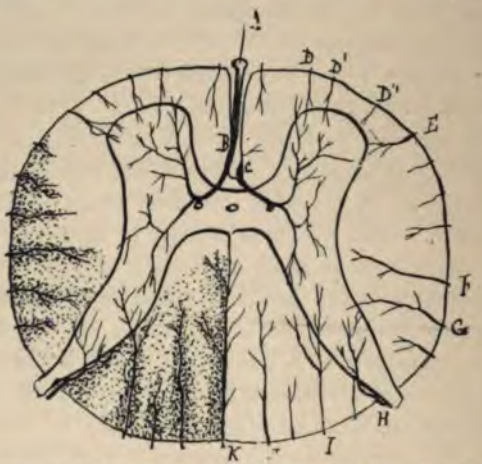

Fig. 216.—Diagram made for the purpose of showing the part taken by vascular lesions in the production of certain forms of combined sclerosis. E, F, G, anterior, middle, and posterior lateral arteries; H, posterior radicular artery; I, posterior cornual artery; J, post-intermediate artery; K, posterior median artery. On the left side of the figure the existence of sclerosis is indicated (by means of points) round each of the arteries in the posterior column and the peripheral and posterior part of the lateral column, the blood of which is chiefly supplied by the posterior spinal artery; the seat of the sclerosis which is thus indicated is a good representation of what is really observed in the combined form of sclerosis of vascular origin.

by sclerosis correspond to the parts of the cord which receive their blood from the system of the *posterior spinal artery*.

Let us consider, for example, the *posterior columns*. You observe, gentlemen, that the sc[illegible] is very pronounced in Goll's columns around the *poste*[illegible] *artery*; in the same way in the columns of Burda[illegible] sclerosis exists near the *post-intermediate* artery, and t[illegible] more inflamed of vascular changes sclerosis exists[illegible] which recalls in the most exact way by[illegible] characteristic of tabes in an advanc[illegible]

This, however, is not all that may b[illegible] seen, that which characterises the case[illegible] is the fact that *that* condition not only [illegible] columns but also in the *direct cerebella*

columns almost at the points occupied by the pyramidal tract. It can very easily be shown, gentlemen, that these lesions also

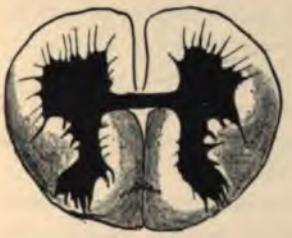

Fig. 217.

Fig. 218.

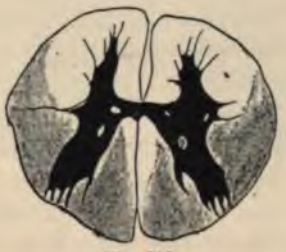

Fig. 219.

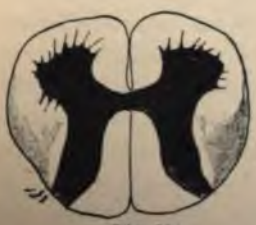

Fig. 220.

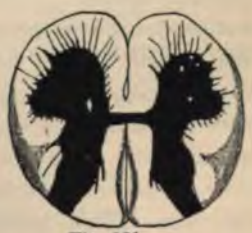

Fig. 221.

Sections of the spinal cord from a case of false systemic sclerosis in the spinal cord of vascular origin (after M. M. Ballet and Minor). The parts of the white substance affected by sclerosis are of a black or grey colour according to the intensity of the lesion. Fig. 217.—Cervical cord. Fig. 218.—Middle part of the dorsal cord. Fig. 219.—Lower part of the dorsal cord. Fig. 220.—Middle part of the lumbar cord. Fig. 221.—Lower part of the lumbar cord. It is observed that the lesions diminish from above downwards, and that in the middle part of the lumbar cord the posterior columns are completely unaffected.

a change in certain vascular regions which also
supply from the system of the posterior
ular regions are those of the *posterior*,
artery; on the one hand these

arteries being distributed to the lateral part of the surface of the cord give rise to marginal sclerosis in this part which exactly resembles degeneration of the direct cerebellar tract, or if it extends rather more forward, of the antero-lateral ascending tract of Gowers. On the other hand it is intelligible that since branches are distributed by these arteries as far even as the central part of the lateral column, while some of them extend to the grey matter, their inflammation may give rise to sclerosis in the lateral columns, which may extend far within them, and affect the fibres of the crossed pyramidal tract which exist at that part. I will not dwell upon the lesions which must necessarily exist in the grey matter, since they are little if at all known, but I insist upon the fact that, in this case, the system of the posterior spinal artery being alone involved, the internal part of the anterior column is usually free from sclerosis, that is the region occupied by the direct pyramidal tract. The fact of the direct pyramidal tract being unaffected while the crossed pyramidal tract is badly affected by sclerosis, forms, as I have already said, one of the best arguments which can be used in opposition to the theory that this form of sclerosis is of systemic nature. The system of the anterior spinal artery may, however, share in the morbid process: a diffuse form of sclerosis then exists in the cord which can scarcely be said to be in appearance either systemic or pseudo-systemic, unless, as in the case which I have put before you, the system of the anterior spinal artery is but partly affected. In that case sclerosis may exist round the *anterior median artery* in the anterior column of the cord. This artery, however, scarcely ever dividing the sclerosis, is found to be unilateral, as in the example which I now put before you, so that its pseudo-systemic character becomes even more manifest.

You see therefore, gentlemen, that the *vascular origin* of combined lateral and posterior sclerosis is in some cases quite clear and easy to demonstrate; I must add, and I have already expressed this opinion, that the vascular origin seems to me by far the most frequent.* The meaning of this is, gentlemen, that that affection is essentially secondary, and that in addition

* The cases observed by M. Demange might be mentioned here, who observed a diffuse form of sclerosis to occur in the cord in old age consecutively to atheroma of the spinal arteries, and to give rise to clinical symptoms which recall fasciculated sclerosis of the lateral columns.

to this, far from being a disease of the spinal cord, it is at most an example of a vascular inflammatory process of which the effects are localised in certain regions of the cord. These conclusions should take the first place in connection with the *ætiology* of that form of combined sclerosis, and I have no doubt that certain general diseases in which the blood vessels are specially affected, as for example *syphilis*, would be often found, if more carefully examined, to be cases of this kind.

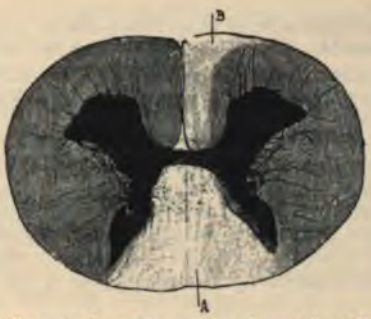

Fig. 222.—Section of the spinal cord in a case of combined lateral and posterior sclerosis (cervical cord). The white parts are those in which sclerosis exists. The two posterior columns A, and the anterior column B, upon one side are affected by sclerosis. This combination, which specially indicates a vascular origin, is not rare. Westphal, Strümpell, Sioli, Kahler, and Pick, &c., have observed and given illustrations of cases in which it existed, but an islet of degeneration was usually found to exist also in the lateral columns.

LECTURE XXXIV.

INFANTILE SPINAL PARALYSIS.

HISTORY: From a clinical point of view, Heine, Rilliet and Barthez, Duchenne of Boulogne, Laborde. From an anatomical point of view, Prevost and Vulpian, Clarke, Charcot and Joffroy. SYMPTOMS: Fever, gastro-intestinal derangements, nervous symptoms consisting of somnolence, convulsions, &c. *Paralysis*, the nature of its onset and extension, its seat, its regression. Period of *deformities*, points to be considered as regards their production; the duration of the paralysis, early age of the patient at the time of its occurrence, atrophy of the bones. Nature of the paralysis, results of an electrical examination, loss of the tendon reflexes, flaccidity of the limb. Integrity of the sensory functions, and of the superficial reflexes. Trophic disorders: subcutaneous adiposis, thinness of the skin, ulcerations, callosities, chilblains, hypertrophy of the hair, excessive secretion of sweat, fragility of the bones. Intellectual condition.

ACUTE SPINAL PARALYSIS OF THE ADULT.

History: Duchenne of Boulogne, Charcot, Moritz, Meyer, Bernhardt, Bourneville and Teinturier, E. C. Seguin. SYMPTOMS: *Onset*, fever, nervous symptoms, &c. *Paralysis*, its mode of onset and extension, its seat and regression. Briefly, the age of the patient being excepted, the symptoms are quite analogous to those which occur in infantile paralysis; the few clinical differences which exist are connected with the onset of the disease.

GENTLEMEN,—The name of *infantile spinal paralysis* is by no means the only one by which the affection of which I shall have to speak to you to-day is called; it has also been termed *atrophic infantile paralysis*, *essential paralysis of children*, *acute anterior poliomyelitis of childhood*, *anterior tephromyelitis of childhood*. As I believe the first of these names is generally adopted on account much less of its excellence than of the long time which it has existed since Heine gave a very remarkable description of this affection in 1860, which he called by this name, and the existence of which he had observed since 1840. More recently (1883) Rilliet and Barthez well described the clinical features of the disease under the very inexact denomination of *essential paralysis of childhood*.

It is only fair to state that the disease has only been thoroughly

known since the memoirs of Duchenne of Boulogne (the father and son) and the thesis of Laborde were written.

Such is the history of the disease as regards its clinical aspect. The history as regards the pathological anatomy is no less interesting, in which the works of Prevost and Vulpian, Charcot and Joffroy, of Roger and Damaschino, hold a preponderating place.

I shall return, gentlemen, to most of these works, and speak also of others. I wished, however, to commence by enumerating the works of which I have just spoken, since the names just mentioned are connected not only with the history of infantile paralysis but with that of diseases of the nervous system in general. It was, in fact, by their observations that the first ideas of medullary pathology were established, and the researches of Prevost and Vulpian, Clarke, Charcot, and Joffroy, proved undoubtedly the connections which exist between lesions in the anterior cornua of the spinal cord and muscular atrophy.

Before, however, considering the pathological anatomy of infantile spinal paralysis, interesting as it is, I would first, gentlemen, make you thoroughly acquainted with the SYMPTOMS.

The mode of onset is of great importance, a fact upon which I shall insist when discussing the pathology of the affection. In a general way it resembles in every respect the onset of acute febrile diseases; the little patient is found, in fact, to be suffering from high fever, the temperature rising, perhaps, to 103° or 104° F. (39° to 40° C.), or higher. The duration of the febrile symptoms varies, and while sometimes this does not exceed 36 or 48 hours, it may extend over several days.

At this time the *gastro-intestinal derangements* which usually exist in acute diseases occur, namely, loss of appetite, a coated tongue, diarrhœa, vomiting, &c., of which the intensity varies in different patients. These are merely the ordinary accompaniments of the febrile condition and have no special connection with this disease.

Symptoms associated with the *nervous system* often occur, which may to a certain extent be qualified as "general," consisting in a more or less pronounced condition of somnolence, sometimes amounting to coma, which terminates in death. Lastly, convulsions at times exist, but not in my opinion as often as some authors have stated to be the case.

So far, gentlemen, there is nothing in the symptoms, as you see, which indicates the existence of infantile spinal paralysis: it is purely and simply the usual history of the onset of an acute disease, when the physician can only recognize the existence of fever, and state "that patience must be exercised until the character of the disease declares itself."

In a short time, however, the scene changes. Either at the very first or, it may be, at the end or during the course of the febrile period a fresh symptom occurs which reveals the disease, namely, *paralysis*. It rapidly reaches its maximum extent, either immediately or in from 12 to 24 hours, announcing its existence by the immobility of one or more limbs.

The extent and seat of the paralysis are very variable. It is often considerable in degree. Thus the four limbs may be found to be simultaneously affected, or complete paraplegia may exist, or one of the upper and both of the lower extremities may be paralyzed, or perhaps the upper extremity upon one side and the lower upon the other. Lastly, in some cases the whole of one side of the body is paralyzed while the other is totally unaffected (hemiplegic form).

At this time the whole limb is often paralyzed without its being possible to localize the affection in one or other of the muscles. In a short time, however, this is no longer the case. At the end of a few days, in fact, some of the muscles are found to regain their power, and the patient is then in what has been termed the *period of regression* or *fixation* of the paralysis.

This period usually lasts for about one or two weeks. From day to day, so to speak, the extent of the paralysis diminishes, and instead of the whole extremity being affected some muscles of the limb are soon found to be alone affected. It is then, for the first time, possible to ascertain the exact seat of the affection and to observe which muscles are involved, this fact being most interesting as regards the seat of the lesion in the spinal cord.

Certain observations have been made as regards the affected muscles which should be mentioned, although they cannot be applied to every case.

In the *deltoid* muscle the fasciculi may be separately affected by the paralysis; its *clavicular portion* being involved at the same time as the upper part of the serratus magnus, whilst its *middle* and *posterior portions* are paralyzed at the same time as

the *infra spinatus* and *rhomboid*. The clavicular portion of the trapezius, which receives its nerve supply from the spinal accessory, is not paralyzed in conjunction with the other fasciculi of that muscle (Kirmisson).

In the lower extremity, the *tibialis anticus* may be paralyzed quite separately from the *peronei* and *extensor muscles of the toes*. At the same time, it must be remarked, that as one would be far from expecting in a disease which is so definitely localized in the cord, the dissociation of the muscular districts affected is by no means so definite as might have been supposed.

The *motor nuclei in the bulb* may also be paralyzed. Médin, in the epidemic which he observed, found that the muscles supplied by the 6th nerve, the portio dura, and the hypoglossal nerve were affected; this fact should perhaps be accepted with some reserve, although there is nothing improbable in its occurrence.

The *sphincters* are usually unaffected, at the same time incontinence may exist, but this rarely occurs except at the onset and during the period of invasion.

The period of regression continues, as I have already said, during 1 or 2 weeks, the muscles of which the motor nuclei are not or but little affected regain power, the paralysis which existed in them having been due, so to speak, to the shock produced upon them by the vicinity of the focus in the spinal cord now gradually ceasing. It is quite different, however, with respect to the muscles, whose nuclei have been more or less completely destroyed by the lesion: these not only remain paralyzed, but a new symptom, atrophy, occurs, indicating the degeneration which exists in them.

A new period in the disease now commences, the period of *deformities*. These may be of various kinds, and are due to causes which we shall have to study in a few moments.

The most frequent and characteristic of these deformities are the following:—

The *club-foot* which usually exists, is the form *equino-varus*, either alone or combined to a varying extent with flat foot.

Club-hand, of which the form varies in different patients, may exist, and is often complicated by more or less pronounced deformity of the fingers.

Various deformities occur in the *trunk*, from simple scoliosis

to the more complicated position which is termed by the popular, but at the same time expressive name of *cul-de-jatte*. In the

Fig. 223.—A patient of the male sex affected for a long time by infantile paralysis of the left arm and shoulder. The atrophy is very pronounced. (Damaschino collection.)

latter case, the inferior part of the trunk and lower extremities have been simultaneously affected, and cannot be used by the patients, either to maintain the upright position or for locomotion.

This deformity is somewhat rare, but in most persons in whom it exists, is due to infantile paralysis. The same is not the case as regards scoliosis, which may be due to a large number of causes.

In order to understand *the existence of these deformities* and the degree to which they extend, a certain number of elements must be borne in mind:—

The long duration of the paralysis has evidently some effect, but cannot certainly be considered a chief agent, since in adults who suffer from atrophic muscular lesions, the limbs are never found, even after 10, 15, 20 years or more, to be as much distorted as in infantile paralysis.

The *early age* of the patients must be specially regarded as

the true cause of the pronounced changes which occur; the patients in fact are children whose development is still taking

Fig. 224.—Infantile paralysis affecting the lower limbs and muscles of the pelvis, and having produced the infirmity which is termed "cul-de-jatte." (Damaschino collection.)

place, and every derangement in the growth of the limbs is indicated by some deformity which increases that which is due to the paralysis, and at times becomes exceedingly pronounced.

It must also be observed, that not only are the muscles affected by paralysis and atrophy, but the latter condition also exists in the *bones*, probably for the same reason as in the case of the muscles, namely, the destruction of certain trophic centres in the spinal cord. As a corollary to these muscular and osseous lesions, *dislocation* occurs in one or several joints of the paralyzed limb. These dislocations are as much due to predominance of tone in the unaffected muscles whose antagonists have ceased to exist, as to change in the direction of the articular surfaces.

The paralysis which is associated with atrophy in this disease has also a certain number of features which contribute to give it a special character.

The paralysis is *essentially flaccid*, both as regards the consistence of the muscles, the tendon reflexes, and the movement of the different segments of the limb with respect to each other.

The *tendon reflexes* in the affected muscles are usually lost, both in the upper and lower extremities.

As regards the *movement* of the different segments of the limbs with respect to each other, this is in a most abnormal condition; if, for example, the leg is grasped in a case of paralysis of the lower limb, and shaken, the foot is found to swing in every direction as if it did not belong to a living person, but to a doll; this condition has been termed the *leg of a puppet* (*jambe de polichinelle*).

An *electrical examination* made at the onset gives the following results as regards the paralyzed muscles: The *faradic*

Fig. 225.—Bones of the leg and foot in a case of infantile paralysis. Considerable deformity exists in the bones of the foot. (Damaschino collection.)

irritability is rapidly lost in the paralyzed muscles, but only in those which will be permanently paralyzed; those which, although apparently paralyzed, still contract to a certain extent, a fortnight after the onset of the affection will, in all probability, regain their power to a greater or less extent within a somewhat short space of time. As regards *galvanic irritability* this is increased at first in the muscles in which the paralysis will persist—in which the reaction of degeneration is also found to exist.

The *idio-muscular contractility* is usually increased, at any rate at the onset, in the muscles of which the paralysis will persist; at a later period it entirely ceases in these, which also occurs as regards the electric contractility.

With regard to *sensibility* it may be said, in a general way, that it is unaffected; in very rare cases, however, it has been

found to be changed, but usually to such a slight and limited an extent that it does not appear right to consider it in the description of infantile spinal paralysis.

Reflex action is also unaltered, both as regards the cutaneous and tendon reflexes in the muscles, which are not affected by paralysis.

Certain *trophic disorders* occur with varying frequence and intensity. The following are those which most often exist:—

The *growth of the limbs is arrested*, to which fact I drew your attention just now, and which explains why it is that the deformities are so considerable in degree.

The *subcutaneous adiposis* (local obesity of Landouzy) which exists sometimes to a considerable degree in the wasted muscles, so as even to entirely conceal the atrophy or even simulate hypertrophy of the limb. This subcutaneous adiposis, however, is by no means constant.

The *coldness* of the paralyzed limbs. This coldness may be very pronounced, and the parents try in vain to prevent it by covering the limbs with the warmest clothing; the children persist, however, in having, as it were, "frozen feet."

Cyanosis, redness, marbling of the skin are the usual accom-

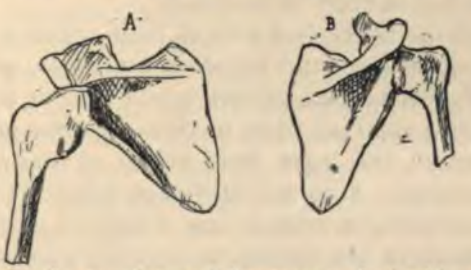

Fig. 226.—Fig. A the scapulum and humerus on the healthy side. Fig. B scapulum and humerus on the side affected by infantile paralysis. The bones are much smaller than on the sound side.

paniments of coldness of the limbs. An eruption of *purpura* has been also known to occur in the wasted limb.

The *thinness of the skin* is at times so pronounced that excoriations or ulcers are produced by the slightest injury, specially owing to the pressure of the orthopœdic apparatus which the patient has to wear, or simply from the effect of walking in boots which press slightly against the foot. When subsequently

by means of rest and careful treatment the ulceration has healed, which does not always readily occur, the cicatrix formed is too thin, and the slightest cause reproduces the ulceration in the same place.

On the other hand *callosities*, often of considerable size, may appear upon some points of the skin covering the limbs, specially in the extremities which are subjected to repeated friction.

As a consequence of this vulnerability of the skin the tendency to *chilblains* must be mentioned, from which the patients usually suffer.

In some cases, again, the limbs which are affected by paralysis and atrophy present a somewhat singular appearance on account of *hypertrophy of the hair*, and at times *excessive secretion of perspiration* in them.

Lastly, the trophic disorders, instead of affecting the skin and soft parts, may involve the *bones* themselves. I have already mentioned, gentlemen, the arrest in their growth; in certain cases they are found to be extremely *fragile*, and in regard to this I would remind you of the *fractures* which Potherat and Berbez observed to occur in persons suffering from atrophic infantile paralysis, which well demonstrate the existence of this fragility in the bones.

As regards the *intellectual state* of those affected by atrophic infantile paralysis care must be taken, gentlemen, not to believe that it is affected, as some authors would have us believe. The patients, from a *psychical* point of view, are quite on a par with healthy persons, and have been known to occupy the most eminent positions. I do not deny that some of them may be extremely irritable, or more or less strange in character; this, however, would, in my opinion, be specially due to the bodily infirmity, which prevents them from leading the same life as other children and young people of their age, and contributes in different ways to increase the nervous sensibility. It may be that the inherited neuropathic tendency has also some effect, when it exists, in increasing some of these conditions.

Acute Adult Spinal Paralysis.

I should now, gentlemen, consider with you, an affection which is in every respect analogous to the one which we have just studied; it occurs when the period of childhood is ended,

and is named on that account, *acute adult spinal paralysis*. Its symptoms, ætiology, and pathological anatomy are so completely identical with those of acute infantile spinal paralysis that it seemed to me that these two affections should be considered, so to speak, simultaneously. We shall thus have the advantage of avoiding repetition and gaining time.

As if to accentuate still more the identity of these two forms of spinal amyotrophy, one finds the names of Duchenne and Charcot connected with the origin of acute adult spinal paralysis. Amongst many other works I would also name those of Moritz Meyer, Bernhardt, Bourneville and Teinturier, E. C. Seguin, &c. . . .

The only difference, as I said, gentlemen, is in the age at which this form of acute spinal paralysis occurs, since it is entirely the same disease: thus the difference which exists in its clinical features are, as you would expect, but slightly pronounced.

The onset is the same, being sudden, accompanied by fever, pain in the loins, cervical region, and at times in the limbs, with or without the sensation of tingling. Gastro-intestinal disorders may also occur more or less resembling those due to gastric trouble. Symptoms connected with the nervous system are often also observed at this time. These, however, do not consist of convulsions, as in children (you know that in early life convulsions are, so to speak, the common mode of response on the part of the nerve centres to different forms of irritation, whereas in the adult this is no longer the case). At this time of life cephalalgia, somnolence, and more or less pronounced delirium, are more liable to exist.

Paralysis occurs either at the end of a few hours or after a day or two; the limbs are specially affected and rarely the sphincters; the same parts are liable to be affected as in cases of infantile paralysis.

In the same way again, as in the latter affection, after an interval of a few days the *period of regression* exists, in which the paralysis only persists in the muscles which will be permanently paralyzed.

The second period is followed by a third, "that of regression," during which the size of the muscles diminishes and the muscular fibres, remaining paralyzed, are destroyed.

It is useless to insist upon the character of the amyotrophy since it is almost identical with that already described with respect to acute infantile paralysis; the electrical reactions, the flaccidity, the loss of tendon reflex in the affected muscles, are the same. With respect to the deformities, the same changes do not occur as in children. In the adult, deformities are less frequent, and invariably far less pronounced in degree, as will be easily understood from the fact that growth having ceased, less extensive deformities are liable to occur in consequence of the paralysis.

The truth is that acute adult spinal paralysis rarely occurs, far more rare, in fact, than the infantile form, though I can give no reason for this fact. It is perhaps on account of the grey matter being more vulnerable in early life. This is merely a supposition, but is one which appears to me somewhat probable, on account of the clinical symptoms and pathological anatomy of the disease.

LECTURE XXXV.

INFANTILE PARALYSIS (*continued*).

ABNORMAL FORMS. Insidious onset; onset during convalescence after an acute disease; onset announced by pain. Forms of transient paralysis. Fatal termination. Late reappearance of paralytic symptoms, or occurrence of secondary progressive muscular atrophy. Theories upon this subject. *Diagnosis:* from *birth palsy; syphilitic pseudo-paralysis;* infantile cerebral hemiplegia; myopathic atrophy; muscular atrophy of the Charcot-Marie type; hysterical paralysis with amyotrophy in childhood. ÆTIOLOGY: Cold, injury, ordinary causes; dentition probably has some influence but is not a direct cause. Infantile paralysis really occurs from the influence of a general disease, usually of *infectious character*. Enumeration of the infectious diseases after which it has been known to occur. Epidemics of infantile paralysis: Cordier, Leegard, Medin, Bergenholtz, Colmer, Briegleb. *Hereditary influence*. The age at which infantile paralysis usually occurs is between one year and eighteen months. The sexes are equally affected.

GENTLEMEN,—We considered in the preceding lesson the symptoms of infantile paralysis and the course which it followed in ordinary cases. I have insisted sufficiently, I think, upon the character of the onset, which is accompanied by the usual symptoms of a febrile affection, the occurrence of paralysis to a somewhat extensive degree, to the subsequent period of regression, and lastly, to the atrophic period, or rather condition, which may be considered the result of the preceding stages. I do not wish to repeat these facts, but would remind you that if medical pathology is based upon general facts, clinical practice is concerned with individual cases alone; the *course* which I have described is that which usually occurs in this disease; but exceptions occur, and the ordinary type of the disease is often modified. It is of these abnormal cases that I would speak to-day.

The *onset*, instead of causing great distress, may on the contrary occur so *insidiously* that those living with the child observe nothing extraordinary in its condition. I am not alluding, gentlemen, to the cases in which a child is entrusted to a nurse, and returns after being under her care in a paralyzed

and wasted condition; it is in vain that the nurse is asked in these cases for an explanation of the condition: nothing strange has been seen or noticed, except that "the limb did not continue to grow"; such is the classical and invariable answer; additional inquiry is useless, no more information will be given. The cases of insidious onset of which I wish to speak are those in which sincerity exists, and the child has received every attention; the parents, who are intelligent and careful, tell you that one morning upon lifting the child, or putting it in its bath, one of the limbs was noticed to remain still, and not to be moved in the same way as that on the opposite side, paralysis was in fact clearly recognized to exist; at the same time it is stated that nothing abnormal had been observed in the child either during the day or in the night which preceded the onset of the paralysis. These cases, gentlemen, were specially described by S. West under the somewhat appropriate name of *paralysis in the morning*.

Another variety of infantile paralysis with insidious onset is that which occurs *in the course of or during convalescence after an acute disease* (measles, scarlatina, &c.), and as you will see this is by no means rare. It is quite intelligible that when this is the case, the symptoms which precede the onset of the paralysis may be easily unperceived, and even in all probability be completely absent.

Lastly, in some cases the onset of infantile paralysis has been stated (Osw. Laurent) to be characterized by pain in the limbs, which was so severe that the true nature of the disease was quite unrecognized and acute rheumatism was believed to exist. As regards the *termination* of infantile paralysis we have already seen, gentlemen, that it usually consists in paralysis, associated with atrophy of those muscles which remained affected during the period of regression. This, however, is not always the case; in some patients the paralysis only remains for a few days and then entirely ceases. This is *the transient form of paralysis* (Kennedy). Some authors state that improvement may occur after the lapse of some months—even of several years. Does infantile paralysis ever end in *death?* Many authors are silent as regards such a termination; it seems to me, gentlemen, that such an occurrence must be regarded as possible when the lesions are very extensive and seated in the

upper part of the spinal cord or medulla oblongata, and I am convinced that some children in whom death has occurred, as it was supposed, from meningitis really suffered from unrecognized infantile paralysis, in which the lesions so rapidly produced a fatal result that the clinical symptoms of the disease could not occur.

As regards the course and termination of infantile paralysis, I must dwell upon a most singular symptom, which is not extremely rare, and the knowledge of which we specially owe to Ballet and Dutil,* although some authors had previously published examples of its occurrence (Charcot, Vulpian, Seeligmüller). The following is what occurred: a patient, who at the age of 2 or 3 years for example had suffered from infantile paralysis affecting either the upper or lower extremity, and who has attained the age of 15, 20, 30 years or more without being troubled by the disease in any additional way, finds that he is suffering in a somewhat serious way. Movements, either of the segment of the limb affected by paralysis, or of the parts which adjoin it, become more difficult; the power of the muscles, which is already much diminished in this region, becomes progressively decreased, and what is worse, this diminution of the muscular power is observed to exist not only in the muscles, which are affected by atrophy, but in those in which there is no sign of this condition. Progressive amyotrophy exists, of which the progress occurs more or less tardily, but is continuous with a pronounced tendency to generalization. It must also be remarked that in some cases this second onset of amyotrophy may be found (Rémond of Metly) to commence in the same limbs as were paralyzed in the initial stage of the disease, and which recovered power during the period of regression. These facts seem at least to show that at the time of the invasion of the disease a lesion had existed in the part of the spinal cord which corresponds to these limbs, and that at a later period the morbid process, which seemed previously to be dormant, or to have ceased, became again active. How general the affection may become I cannot say, on account of the small number of observations made; but it is certain that not only the muscles of the limbs, but also those of the trunk may be involved in this new amyotrophic process. Thus a

* Ballet and Dutil. Revue de Médécine, 1884.

patient in whom this disease was studied by Charcot* in one of his lectures was affected by atrophy affecting the muscles, which moved the scapula and shoulder joint precisely as it does in primary myopathic atrophy. However extraordinary these facts may at first appear we shall see when considering the pathological anatomy of the disease in what way an attempt may be made to explain them. This second onset of amyotrophy, which occurs some time afterwards, may be also observed in the acute spinal paralysis of the adult, which, in fact, as we have already seen, is but a variety of infantile paralysis.

Infantile paralysis is, as you know, gentlemen, by no means the only form of paralysis attended by wasting which occurs in early life, and before continuing the study of this affection the diseases should be mentioned which more or less resemble it from a clinical point of view. You will see that the diagnosis usually presents little difficulty.

In mentioning these different forms of paralysis in chronological order, *birth-palsy* must be first considered. This, which was mentioned by Danyau in 1851 as possibly occurring when the forceps was used in child-birth, was subsequently studied from a neurological point of view by Duchenne of Boulogne, who saw them occur when the forceps had not been used at birth, as after version, breech presentation, or in fact any difficulty in parturition. It is to Erb that we specially owe the knowledge of the exact seat of this form of paralysis; it is the deltoid and infra-spinatus muscles, the biceps and brachialis anticus, and often also the coraco-brachialis and supinator longus muscles which are usually affected. In one word, the nerve roots are affected in this form of paralysis. From a clinical point of view the arm which is applied against the trunk is kept very low, while the arm is rotated inwards and the forearm extended upon the arm. The electrical reactions are more or less analogous to those which occur in infantile spinal paralysis, and present no very distinctive features. The course of this paralysis is generally favourable, and it usually ceases after a short time. It need not, I think, be discussed at greater length, since the different characters which I have already mentioned are sufficient to prevent any mistake from being made.

* J. M. Charcot. Bulletin Médical, 1890.

Syphilitic pseudo-paralysis is also an affection which occurs in early life; the works of Parrot, Troisier, and Dreyfous have shown that this affection may involve one or more limbs; as for example, one of the upper and both of the lower extremities, and that the attitude of the affected limbs which hangs down in a motionless condition much resembles that which is due to paralysis, specially infantile paralysis. At the same time these authors have shown us by what means a mistake of this kind may be avoided; owing to their observations we know that in these cases true paralysis does not exist, but only a loss of power in the limbs, a false paralysis due to a lesion in the bones at the junction of the epiphysial cartilage with the diaphysis. Slight attention to the case soon enables the following symptoms to be recognized: somewhat pronounced swelling of the limb specially near the extremities of the affected bones; the crepitation near the joints; the maintenance of the electrical reaction; preservation of voluntary movements in the extremities (hands, feet, fingers); pain which is often very severe in the paralyzed limbs, but without sensory disorder in the skin; a tendency to progressive aggravation of the disease, the limbs being affected one after the other; lastly an eruption, which is clearly of syphilitic origin, often adds the last touch to a picture which is already most characteristic.

Infantile hemiplegia of cerebral origin is sometimes confused with infantile spinal paralysis, since it produces in the same way paralysis of the limbs with atrophy, and under its influence considerable disorders may occur in the muscles and bones; in some cases monoplegia and not hemiplegia exists. In this affection, however, the tendon reflexes are excessive, instead of the reverse occurring as in infantile paralysis, attacks of epilepsy often co-exist, and the most varied conditions of intellectual defects amounting perhaps to complete idiocy are not unfrequently observed, whilst, as we have already observed, nothing analogous is found to occur in infantile paralysis.

Myopathic atrophy, in whatever form it may occur, can scarcely be confused with the disease which we are now considering; it never, in fact, produces atrophy of a limb or even of the segment of a limb, but wasting, which is exactly confined to certain muscles, and which is seated in the root of the limb rather than in the limb itself.

A form of muscular atrophy must be also mentioned which occurs in early life and which is known, at any rate in France,

Fig. 227.—A girl affected during the first year of her life by infantile hemiplegia of cerebral origin affecting the left side. The left arm is shorter and less developed, pronounced club-hand existing. The same remarks may be made about the left leg. (Patient under the care of Charcot. Stereotype of Londe.)

by the name of the Charcot-Marie form, in which both the lower and upper extremities are thus affected by pronounced atrophy. What distinguishes this form of wasting from the atrophy which exists in infantile paralysis is its onset, which

is essential and progressive; the almost perfect symmetry in the muscular changes which exist upon one side of the body or the other, and the absence of, or slight degree in which trophic disorders affect the bones is also characteristic.

Lastly, the observations which I am making about the diagnosis must not be brought to a close without my reminding you that in some cases, which though certainly rare, have undoubtedly been shown to exist (A. Chauffard), children who have been long affected by *hysterical paralysis* of a limb, present, on account of absence of growth, such want of development in the size and length of the limb, that one might be tempted at first sight to believe in the existence of infantile paralysis. When this happens, the history of the case, the electrical reactions, and tendon reflexes which are retained, and the recognition of other hysterical symptoms prevent the occurrence of any mistake.

The *ætiology* of infantile paralysis must be fully considered since, as you all see, gentlemen, unless this is completely understood, the true nature of this affection cannot be realised.

The effect of *cold*, and of *injury*, have, as you may well suppose, been invoked by numerous generations of authors. I have had upon several occasions to give you my opinion as to the importance of these so-called ordinary causes; I will not repeat my observations upon this point, and merely say that at most they can only be looked upon as occasional causes.

The ætiological consideration upon which Duchenne of Boulogne for his part most willingly insisted, was *dentition*. According to that author, by far the most cases occur at the same period of infantile development, and often at the very moment when one or more than one tooth is actually piercing the gums. You know, gentlemen, how great a part "the period of dentition" played in connection with the diseases of children in old times; you also know that a reaction, certainly excessive as every reaction is, has occurred during the last few years. After being regarded as the keystone of pathology in childhood, this period has come to be looked upon as of no importance whatever. I must admit, and I believe that my opinion is shared by a certain number of clinical observers, that dentition seems to me a state which should not be disregarded. Not that it can itself be the direct cause of any disease, but because it constitutes a period during which the organism is fatigued,

overwrought, weakened, either by the pain suffered, or loss of sleep, or the diarrhœa which is so often associated with the eruption of teeth. The organism which is thus fatigued will on that account be specially exposed to the different forms of infection. Such is the part which I believe dentition to play in infantile paralysis. This is a time of life during which the organism being depressed, in consequence of its existence, is specially liable to suffer from the different infectious diseases which may be followed by infantile paralysis. Dentition is not an actual cause, but a period during which this liability exists.

My firm conviction, in fact, is, gentlemen, that infantile paralysis invariably, or almost invariably, depends upon the existence of some *general affection*, usually of an *infectious* nature. This has been my opinion for many years; and though I do not believe that it is confirmed as yet by many adherents in France, this is of little consequence, and certain presentiments lead me to believe that "the time approaches" when this will be the case. I hope, in fact, that I shall be able, if not to convince you all, at any rate to obtain some converts to my opinion, while reserving until another occasion a statement of the arguments which are furnished by its pathological anatomy, I would now place before you those which are founded upon its clinical features alone.

As regards the onset of infantile paralysis I dwelt at sufficient length upon the different general symptoms which occur at this time; it is unanimously agreed that [illegible] quite analogous to those which exist in acute febrile [illegible]ses. It is possible that the acute disease may be [illegible] resulting from some "unknown affection," [illegible] practice. In a certain number of cases, [illegible] will perhaps think more suitable for the de[illegible] theory, the nature of the infectious disease [illegible] what may be called the fundamental infectious [illegible] It is during the course of measles, s[illegible]rlatina, d[illegible] pox or whooping cough, that the [illegible] set of infan[illegible] occurs.

Lastly, one of the arguments w[illegible]h should in[illegible] have the most weight, is that wh[illegible] *nature* of infantile paralysis. [illegible] observations upon this subje[illegible]

Cordier* in a communication made to the Société des Sciences Médicales of Lyons in 1887 states that he saw in two months (June and July 1885), at Sainte-Foye-l'Argentière, in a population of 1,500 persons, the occurrence of 13 cases of infantile paralysis; whereas in other years nothing analogous to this took place.

Leegard† mentions on his part the fact that in the small town of Mandal (Norway), Oxholm and his colleagues observed 8 cases of infantile paralysis between the end of July and beginning of September, this having never happened previously.

Medin‡ has also recognized cases of the same kind, but in far greater number. In the spring of 1888 he had already seen 5 cases, and after August the number increased to such a degree, that the author met with 44 cases before the month of November. It is true that amongst these cases there are some which, as I have already said, cannot be regarded as true examples of the disease in its ordinary form. It cannot, however, be said that they were fundamentally different from the more typical cases of this disease, and, when the whole statement is considered, these cases formed but a small minority which could not seriously invalidate the comprehensive statistical observation made by Medin.

That author also recalls that in 1881 Bergenholtz had already observed a small epidemic of 13 cases at Umea, and that G. Colmer,§ having inquired of the parents of a child suffering from infantile paralysis, learnt from them that in their district, within the distance of a few miles, and in 3 or 4 months, from 8 to 10 children had been similarly affected.

As regards the 5 cases observed by Briegleb‖ in his clinical [illegible] at Iena, during the months of June and July, it should [illegible] that, as the cases came from places which were so [illegible] one or two hours were occupied by the train in [illegible], they seem of less value in connection [illegible] this fact than the preceding.

[illegible]nstration af mikr. prep.; d'après l'analyse

[illegible]l Paralyse. *Hygeia*, Septembre, 1890.
[illegible]iences, 1843.
[illegible]n Natur der acuten Poliomyelitis

Owing to these facts, the epidemic nature of infantile paralysis, at any rate in some circumstances, seems to me definitely established, and in my next lecture we shall see how this fact may be understood.

Such are the arguments which I consider of value in connection with the infectious origin of this disease. I would not terminate my observations about its ætiology without reminding you that in most cases hereditary transmission plays a very important part. For my part, as I have already said, I have no difficulty in believing that the neuropathic tendency communicated by transmission to the organism of a child may cause its nerve centres to be less able to resist the causes of disease. At the same time, I find it quite impossible to look upon hereditary predisposition as the effective and direct cause of the disease.

The *age* at which the affection usually exists is, as I have already said, the *earliest infancy*. In most cases it occurs during the first 2 years of life, specially between the ages of 1 year and 18 months. At the same time it may occur later, as in the 3rd or even the 4th year. We shall have upon another occasion to study a form of poliomyelitis which occurs in the adult, which so much resembles the infantile form that, properly speaking, no precise age can be mentioned which separates these two affections, which seem to occur at any age from infancy to the adult age, and even longer. At the same time, from a clinical point of view, infantile paralysis should be considered a distinct disease, on account of its being specially frequent in early infancy, and of the singular aspect which the affected limbs present which are in full process of development.

As regards *sex*, there is nothing special to be said, the two sexes being equally affected.

LECTURE XXXVI.

INFANTILE PARALYSIS (*continued*).

PATHOLOGICAL ANATOMY.—History of the discovery of changes in the anterior horns: Vulpian and Prevost (1865), Lockhart Clarke, Charcot and Joffroy, Parrot and Joffroy, Roger and Damaschino. (A) Character of the lesions when *the affected focus is of old date*, lesions in the *anterior horns*, nature of these lesions many foci usually exist which are rarely quite symmetrical; atrophy of the half of the spinal cord which corresponds to the paralyzed side, and in some cases of the cerebral hemisphere upon the same side (Rumpf, Colella, Fornario); lesions but slightly pronounced in the *anterior roots* and *trunks of the mixed nerves*, explanation of Joffroy and Achard: alterations in the *muscles*; alterations in the *bones*, diminution of the diameter of the Haversian *systems*; alterations in the *blood vessels*. (B) Character of the lesions when *the affected focus is of recent date:* every appearance of a true inflammatory focus then exists, this focus of acute myelitis may extend to the adjoining white matter; the reason of this fact probably connected with the distribution of blood vessels in that region. NORMAL ANATOMY of the blood vessels supplied to the anterior horn. ANTERIOR SPINAL SYSTEM.—*Anterior spinal artery;* anterior median artery; anterior radicular branches; each of these arteries may be the seat of the intra medullary vascular lesion which constitutes infantile paralysis. Identity of infantile spinal paralysis with infantile cerebral hemiplegia; my disagreement with Vizioli and Strümpell. Observation of P. J. Mobiüs. This explanation of what occurs enables the late "renewal" of the clinical course of infantile spinal paralysis, which has been considered to be understood. THERAPEUTICS.—Electricity, massage, hydrotherapy, orthopædic apparatuses.

THE pathological anatomy of infantile paralysis deserves full consideration, not only on account of its own merits, but also because it is the subject of numerous works which explain the nature of this affection, as you will see; these confirm what has been already said when the ætiology was considered.

The time is far distant at which it was generally believed, as by Rilliet and Barthez, that the disease had *no material cause*, and was a "form of paralysis essential to childhood."

At one of the first autopsies made by Cornil and Laborde, the existence of very pronounced changes was recognized in the anterior and lateral columns. I shall presently have occasion to insist upon the importance of such alterations.

The work of Prevost and Vulpian was written in 1865; these

authors stating for the first time atrophy of the anterior horns, the disappearance of a certain number of cells in them, the diminished size of the white columns upon the same side of the cord, and the fact that the lesions only extended to a certain height. In the same year, Lockhart Clarke, in England, described granular disintegration of the anterior horn.

It was in 1870, that these facts relating to the pathological anatomy of the disease were made of definite value, on account of the discovery by Charcot and Joffroy of the connection which existed between amyotrophy and the lesions of the cells in the anterior horns, since these authors deserve the whole honour of having discovered, and clearly indicated this relation which is one of the bases of medullary pathology.

In 1871 and again in 1881, Roger and Damaschino attempted to show that the lesion in the anterior horns is clearly of inflammatory nature, usually consisting in the focus of softening. The fact is that these authors being in a children's hospital, observed the disease in conditions which were quite different from those in which neurologists usually do so; they could study the disease in fact from the time of its first onset, whilst the latter, in most cases, only come in contact with the disease when the paralysis has already existed for many years.

The character of the lesions is very different, when the disease has only existed for 1 month, from that which it has 5, 10 or 20 years later.

A. We will first consider the latter case which is most usual, *the autopsy only being made after a certain number of years.*

The fact must obviously be remembered that an old cicatrix now exists, and that the lesions have also affected an organ which was in full development; this will explain the perhaps considerable degree to which wasting and atrophy exist in the spinal cord in which the primary changes are seated.

In the spinal cord the grey substance is recognized by means of the naked eye and in preparations which are properly stained to be more translucent at the seat of the affection than in other parts of the grey matter. At these points a glass of slight magnifying power shows that the minute structure of the grey substance has changed its character, that the large nerve c of the anterior horn, which is affected, have disappeare

strongly coloured masses of protoplasm without processes and with a round or obtuse margin, perhaps the relics of these cells being found in places. In the whole affected region a network

Fig. 223.—Lumbar cord in a case of infantile paralysis. (A.) Focus of old date seated in the antero-external part of the left anterior horn; it will be observed that the left half of the cord taken as a whole is smaller in size than the right half.

of fibrils of the same nature as the neuroglia and with somewhat small meshes is found to exist; some of the cells termed cells of Deiters being also found in this part. The nerve fibres, which are normally seen in every part of the grey matter of the anterior horns, are completely destroyed throughout the affected region, or at any rate in much smaller number. The blood vessels are usually thickened, seeming at times to be dilated, while in some cases they appear to be much increased in number on account of the retraction of the affected part, owing to which they are contained in a smaller space.

These changes do not at the same time affect the anterior horns throughout the whole length of the cord, they are also usually unilateral, and in the rare cases in which both sides are involved the diseased parts are never quite symmetrical, the changes occur commonly in the form of foci, which extend over a length of one or several centimetres (from a quarter of an inch to one inch) or more; 2, 3, 4, or more of these exist, and may be found upon both sides of the cord, so that, as you have seen, the left arm and right leg may be affected in the same patient; or if the cells connected with the muscles of the upper and lower extremities are contained in the foci, the infantile paralysis may assume the hemiplegic form; these foci of disease may be

found not only in the spinal cord but also in the medulla oblongata and pons varolii. This, however, occurs more rarely, or at any rate the clinical symptoms which are due to this condition are not usually ascribed to the ordinary form of infantile paralysis.

Such are the principal points which should be noted in connection with the lesions in the grey matter of the anterior horns; whether the changes which occur are quite confined to the grey substance or extend into the white matter is a question which must still be answered. It is difficult to decide this point in such cases as those which we are now considering, namely, when the affection has already existed for many years, since, on account of the retraction which takes place in the cord, the limits of the grey and white matter are not always very clearly defined; more information is gained by the examination of lesions which have recently occurred; this point will be reconsidered upon a future occasion.

On the other hand the examination of lesions of old date gives the most precise information as to the nature and seat of the degeneration which occurs after lesions in the grey substance of the anterior horns.

If a section of the spinal cord, which is made at the seat of one of these foci, is examined with the naked eye, it is easily seen that the corresponding half of the cord is of much smaller size than that of the opposite side; this difference in size occurs not only in the *antero-lateral* but also, and to a very pronounced degree, in the posterior columns; the *posterior horn* is also of smaller size than that on the sound side, and a certain number of observers (Parrot and Joffroy, Charcot, Schultze, Roger and Damaschino, &c., &c.) have found atrophy to exist in Clarke's column upon the side of the disease. This extensive atrophy in different parts of the spinal cord upon one side is probably due to many causes, on the one hand to arrest of development in these parts in consequence of a certain number of cells and nerve fibres being destroyed, and on the other to degeneration or an ascending form of atrophy analogous to that which we considered in detail in the lecture which was devoted to the changes which occur in the nervous system after amputations of the limbs.

Lastly, there is a lesion, the occurrence of which has been

noted by certain authors (Rumpf, Colella,* Fornario†), who observed that the motor convolutions of the brain, which

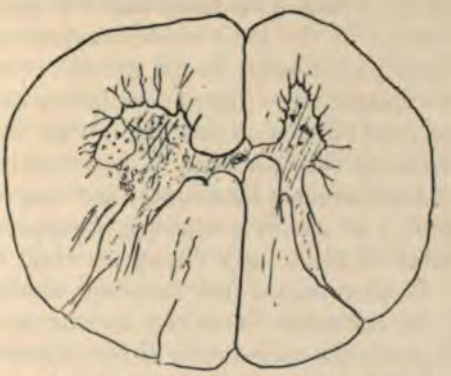

Fig. 229.—Section of the spinal cord from a case of old infantile paralysis. (After Charcot.) It will be observed that the white substance and grey matter upon the right side, which is the seat of the lesion, are affected by considerable atrophy.

corresponded to the paralysed limbs, were also affected by a certain degree of atrophy, this again resembling what occurs in amputation of the limbs (?). Theoretically this is by no means improbable; but if, gentlemen, you ever undertake a research of this kind you will soon perceive how extremely difficult it is to recognize so small a difference between the two hemispheres. The anterior roots are recognized by the naked eye to be clearly diminished in size, though under the microscope scarcely any lesion, according to Joffroy and Achard,‡ is found to exist, and islets of sclerosis are not present as would have been expected to be the case. The authors, whom I have just named, are of opinion that this is due to the fact that the lesions in the anterior horns having occurred during the time of their development the fibres affected by secondary degeneration have disappeared, the sound fibres, which previously adjoined them, having taken their place, so that no vacant place exists which might be occupied by the islet of sclerosis. A somewhat large

* Colella, La paralisi spinale atrofica infantile in rapporto con i centri corticali, &c. *La Psichiatria*, anno VII.

† Fornario, La paralisi infantile atrofica in rapporto alle vie sensitive del midollo, &c. *Giornale di Neropathologia*, May-June, 1890, p. 131.

‡ Joffroy and Achard. *Archives de Médécine Expérimentale*, 1891.

number of authors, on the other hand, have stated that very characteristic lesions exist in the anterior roots.

As regards the *trunks of the mixed nerves* I need only repeat, gentlemen, almost in the same words the observations which I have just made with respect to the anterior roots; the same remarks and explanation on the part of Joffroy and Achard, the same divergence of opinion on the part of other authors.

Very pronounced changes exist, as you would expect in the muscles, which are affected by atrophy, and may even in certain cases completely, or almost completely disappear; or again a certain number of fibres may disappear while the others are unaffected. It often occurs that the fibres which disappear are replaced by an extensive interstitial lipomatosis, which is undoubtedly of much the same origin as the subcutaneous lipomatosis, of which I had occasion to speak in one of the preceding lectures, and the description of which was given by Landouzy.

As regards the hypertrophy of certain fibres in the affected muscles, to which Dejerine has called attention, it would be due, according to some authors, to compensatory excess of action; in Erb's opinion it constitutes the first stages of the morbid process which ends in atrophy; lastly, Joffroy and Achard believe it to be a degenerative change, possibly connected with the incomplete lesion of a certain number of cells in the anterior cornua.

The trophic disorders of the limbs affected in infantile paralysis extend even to the bones. Even at first sight one

Fig. 230.—Sections of the two humeri in a case of infantile paralysis. (A.) Section of the humerus on the sound side; (B.) Section of the humerus upon the side affected by infantile paralysis; the dimensions of the latter are much smaller, and the shape is more rounded. (Damaschino collection.)

recognizes that the outline of the affected bones, instead of presenting, as in the normal condition, ridges and depressions, is irregularly rounded, and does not present the irregular surface which normally exists. In addition to this the thickness of the compact tissue is far more uniform than in a healthy bo

different points of the circumference of the transverse section of a long bone (Joffroy and Achard).

Microscopical examination shows, again according to these authors, that in a general way the *Haversian canals* have a smaller diameter than in the normal state; this diminution in their diameter is specially marked in the deeper parts, where the diameter of the system may be only half of its normal size.

Lastly, the changes which occur in the blood vessels of the paralyzed limbs must be mentioned. These are sometimes much diminished in size, their walls being much thinner, and they are affected by true atrophy, or at any rate very pronounced disorder in their development, on account of the derangement which exists in the trophic functions of the spinal cord. This atrophy in the vascular system may also affect the growth of the whole limb, and its small size should be partly ascribed to this cause.

Such, gentlemen, are the different changes which are found to exist when death has occurred several years after the onset of the affection. However interesting these alterations may be, as I have already said, they are of but slight value as an indication of the nature of infantile paralysis, whilst the study of lesions which have recently occurred has enabled the most important observations to be made; it is these which we will now consider.

B. *The autopsy takes place at the end of a few days, or at most five or six weeks:*

In these cases the induration and sclerosis, the cicatrix as it may be termed, is not found to exist in the part; on the other hand a *focus of true inflammatory softening* (Roger and Damaschino) is recognized in the anterior horn. A moderate magnifying power shows that the details of this portion of the grey matter are not clear, numerous granular bodies being seen to exist, which are either free in the spaces formed by dissociation, as it were, of the tissue by inflammatory effusion, or enclosed in the meshes of the nervous tissue, or blended together in the perivascular lymphatic sheaths.

Within these foci the nerve cells have either completely disappeared or granulo-fatty degeneration is found to exist in them, or again, simple atrophy with a tendency on the part of the processes to disappear, so that at a later period they have the form of the more or less rounded masses to which I have

the external portion of the anterior horn. These foci, due to acute inflammatory softening, are not therefore, as most authors state, essentially limited to the grey matter but may also involve the white substance.

This lesion is not therefore, really systemic in nature, as will at first sight appear, since it passes beyond the limits of the grey matter. What is the cause of this pseudo-systemic character? In my opinion, gentlemen, its cause must be sought in the distribution of the blood vessels within the spinal cord. I hope to be able to show you that by the knowledge of this distribution we shall be able to understand the singular nature which exists in the seat of the lesions in infantile paralysis.

When speaking of combined lateral and posterior sclerosis I had occasion to consider the intramedullary blood supply, but my principal object at that time was to discuss the arteries of the posterior spinal system. To-day it is the ANTERIOR SYSTEM which I shall take as my subject.

This system is composed, as you know, of the *anterior spinal artery* formed by the union, at the upper level of the cervical cord, of the two descending branches supplied by the vertebral arteries: the anterior spinal artery is seated in front of the anterior median fissure, and can be traced as far as the cornus medullaris; in its course it is joined by a certain number of branches of the lateral spinal arteries.

The *anterior spinal artery* gives off a series of somewhat large branches at right angles to its course which pass backwards in the anterior fissure and constitute the *anterior median arteries*.

Each of the *anterior median arteries* passes into the anterior horn upon one side of the cord which it supplies with blood; on that very account of this artery is the seat of a serious lesion, the corresponding anterior horn will be also affected. It should be observed, gentlemen, that the anterior median artery passes to the end of the anterior fissure, and as Kadyi has shown, either to the right or to the left into *one* of the anterior horns, and does not divide so as to supply these horns on both sides; this explains the fact that the lesions in infantile paralysis are nearly always *unilateral*. In addition to this the extension of the lesions into the antero-lateral column is quite intelligible, since one or several branches of this arteriole in the anterior

already alluded when speaking of the lesions of old date. It must be well understood that the nerve fibres are also affected, a certain number of them having the characteristic granular appearance.

As regards the blood vessels, the changes which occur in them are most obvious; in the parts of the grey matter where the affection exists they are found to be dilated. At times, in recent cases, obstruction is even found to exist in consequence of thrombosis. Proliferation occurs in the nuclei of their walls

Fig. 231.—Left half of the lumbar cord in a case of infantile paralysis. In the left anterior horn a focal lesion is seen to exist at (A) in which the blood vessels are much developed. (Spinal cord from a patient under the care of Archambault. Damaschino collection.)

which are themselves thickened. In every case the changes in the blood vessels are very clearly marked.

Thus an inflammatory process clearly occurs in this disease, a form of *acute myelitis* which produces, as I have said, focal lesions. Though there is, perhaps, little to say with respect to the seat of these foci, as regards their height in the cord, since the distance to which they extend is quite variable, the same is by no means the case as regards their seat in the antero-posterior or transverse direction.

In the antero-posterior direction these foci are clearly seated in the anterior horn and usually do not extend beyond the neck of the cornu.

In the transverse direction the lesions have apparently a certain tendency to exist in the antero-external and antero-internal portions of the anterior horn; but owing to the fact that they have a tendency to occur at its outward part, they often encroach upon the antero-lateral column where it adjoins

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median fissure are known to pass into the white substance which adjoins the anterior horn.

It may also be supposed that the primary lesion in infantile paralysis, which is constituted by a focus of acute polio-

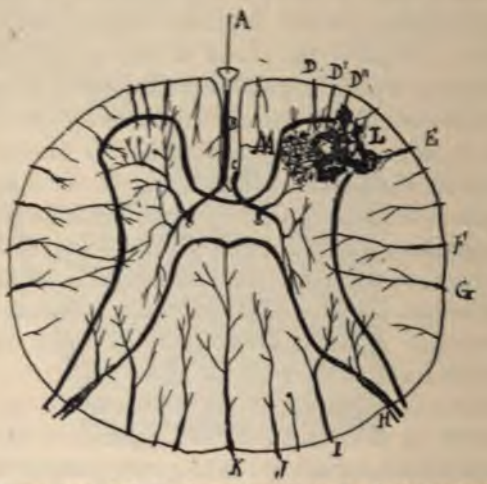

Fig. 232.—Diagram intended to show how lesions of the anterior horns are produced in infantile paralysis. A, anterior spinal artery; B, anterior median artery; C, commissural artery; D, D', D'', anterior radicular arteries; E, anterior lateral artery; F, middle lateral artery. Two foci of disease existing in infantile paralysis are represented, each of which has a different origin; one (M) is due to a primary lesion in the branch of the commissural artery which passes into the anterior horn, it is entirely comprised within the horn; the other (L) is due to a primary lesion seated in one of the *anterior radicular arteries*, it extends into the white substance of the antero-lateral column.

myelitis, does not result from an affection of the anterior median artery, but of one of the *anterior radicular arteries*. The seat of these arteries enables the fact that the antero-lateral column is affected, and the assymmetery of the lesions to be explained. It is possible that these arteries are equally liable to be the seat of the vascular lesion which now interests us, and this difference of seat explains certain varieties which exist in the symptoms. What, however, is the nature of this lesion of vascular origin? It is in the ætiology of the disease, gentlemen, that we shall find the answer to this question. You will not certainly have forgotten [illegible] of the infectious nature [illegible] the febrile symptoms—

account of these facts, solely from a clinical point of view, we decided that the disease was of an infectious nature.

Owing to what we are taught by normal and pathological anatomy, we can now push our conclusions still farther, and state that it is through the medium of the vascular system that this infectious disease involves the spinal cord. As to the exact process, by means of which the lesion is produced, it is difficult to observe it whilst this is actually taking place; at the same time it seems to me most probable that in reality an infectious form of *embolism* exists, or that *thrombosis* occurs in one or several arteries of the anterior median fissure.

It may appear to you strange that these arteries are the special seat of this morbid process in such a comparatively large number of cases. The fact, I allow, is singular; but as I previously observed to you in connection with combined lateral and posterior sclerosis, it is far from being unique, and other examples of its occurrence can be mentioned. Is it again quite certain that the anterior median and anterior radicular arteries are the only vessels affected by the morbid process? I believe, for my part, that a certain number of the acute nervous symptoms of childhood, called for the most part by the mistaken name of "meningitis," are only cases of arteritis, or infectious embolism, it occurring in different parts of the nervous system.

In connection with this subject I should mention the analogy, I would even say the identity, which exists, as regards its pathology, between infantile spinal paralysis and infantile hemiplegia, which is due to a cerebral cause. I do not mention this identity for the first time to-day; I have done so constantly since 1885, and I hope to be able to convince you, gentlemen, that this opinion is in conformity with the facts which really occur. Two eminent pathologists, Vizioli and Strümpell,* preceded me, as I am happy to acknowledge, in having this opinion, both of them clearly expressing the opinion that the two diseases of childhood, which I have just named, were closely analogous

* Vizioli only mentions the similarity which exists in the course of the two affections and the development of the symptoms; he makes no allusion to the eral pathology or pathological anatomy, and, olly distinguishes infantile spinal paralysis are liable to occur at an early age (in ther cause)."—Emiplegia cerebrale hical indication had previously

to each other. At the same time, singularly enough, with the exception of this idea as regards the analogy of the diseases, we all three have very different opinions about the nature of these two diseases.

Contrarily to Vizioli, I believe both these affections to be of an infectious character; while on the other hand I cannot agree with Strümpell that they are systemic diseases of the grey matter. Whatever may be thought of these two opinions, the fact that the two affections are identical seems to me undeniable, and if you have any doubt about this, I need merely quote an observation of Mobius which I have quoted several times; a brother and sister, whose respective ages were three years in the former, and a year and a half in the latter case, after suffering from general symptoms (fever, gastric derangement, &c.) during a few days, were both attacked simultaneously, the sister by atrophic spinal paralysis, the brother by spastic infantile hemiplegia. I will go even farther, gentlemen: I am convinced that a favourable opportunity will occur, and that infantile hemiplegia, due to a cerebral cause, and infantile spinal paralysis will some day be found associated together in the same patient, and I am confidently awaiting the publication of this typical observation which will conclusively prove the identity of these two affections.

One question still remains unanswered: what is the infectious agent which determines the existence of this morbid process in the spinal cord? In reply, suppositions can alone be made. Is it the micro-organism which specially occurs in exanthemata or other diseases, in the course of which we have found infantile paralysis? or are other micro-organisms associated with these, combined infection existing? and in this case is the pathogenic micro-organism specific or common? These are questions which cannot yet be answered, at the same time increased knowledge

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escaped the notice of the authors interested in this question; it is taken from the remarkable work of Sigm. Freud and Osc. Rie upon infantile hemiplegia. Vienna, 1891.

Strümpell, whose memoir upon this subject was written in 1884, starts with the idea that infantile hemiplegia, due to a cerebral cause, is for the same reason as infantile spinal paralysis an essentially systemic affection, seated in the grey matter of the convolutions; hence he applies the name of *polio-encephalitis* to this form of hemiplegia, in the same way as the term *poliomyelitis* was adopted for infantile spinal paralysis. According to M. Strümpell again both these affections are by nature infectious.

is indicated, as it seems to me, by the fact that they can be asked.

Upon examining the different elements of the problem, one fact must be considered specially from this point of view. We have seen, gentlemen, that in some cases, after 10, 15, or 20 years the amyotrophic process which had seemed to have completely ceased, recurred in a most unexpected manner, and became general. To what is this *recurrence* due? One might be tempted to suppose that this was an additional proof that the affection was due to the effect of micro-organisms. The microbes existing in the cicatricial foci becoming again active owing to some as yet unknown influence, and finding a soil favourable to their production in the grey matter of the anterior horns, determine by the pullulation this progressive form of amyotrophy. Examples of such long periods of rest, followed by sudden and pronounced activity of the micro-organisms contained in the bones or skin, are undoubtedly known to exist, specially in certain cases of osteo-myelitis or furunculosis; but whether this justifies the above statement must be decided by future observers.

While still basing my observations upon clinical facts, which are of more definite value, in connection with the "recurrence" of the amyotrophic process, I would direct your attention to another analogy which exists between infantile spinal paralysis and infantile hemiplegia of cerebral origin. In the latter affection it not unfrequently happens that 5, 8 or 10 years after the occurrence of hemiplegia, when the lesions may be believed to have passed into a cicatricial condition, attacks of *epilepsy* occur, often accompanied by a recurrence of the paralytic symptoms, a recurrence of paralysis which exactly resembles that which occurs in the cases of infantile paralysis, to which I have made allusion. The latter analysis should, in my opinion, be noted, since it shows that the fact is not merely a fortuitous incident, but due to a process which is the very essence of these affections.

The treatment adopted in infantile spinal paralysis is unfortunately far less beneficial than might have been expected.

During the period of invasion, the object of the treatment should specially be to reduce the pyrexia, and in fact to combat the infectious disease of which it is the index: sulphate of

quinine and other internal antiseptic agents, or at times when hyper-pyrexia exists, the use of cold baths are specially indicated.

At a somewhat later date different counter-irritants may be applied to the spine, such as tincture of iodine, diluted croton oil, &c.

Lastly, when the period of regression is definitely established, but not previously, in order that a spinal cord which is already affected by inflammation, may not be much subjected to ill-timed irritation by the electrical current employed in the treatment, a weak, a very weak current should be used that the child may not be much disturbed; and whether the galvanic or faradic current be employed, it must be remembered that but slight contraction of the muscles need be produced, and that violent contractions should not occur.

During this period of regression, massage, muscular exercises, hydro-therapy (douches, saline baths, sulphurous baths), the patient should also be watched, in order that when deformities threaten to occur, this may be prevented by the use of an appropriate orthopœdic apparatus. At the same time, care must be taken, gentlemen, not to allow the children to be covered by a too heavy casing of iron, under pretext of preventing deformity in a paralysed limb which must be always useless; they would then be unable to take the exercise which is necessary for their development and good health, and you would thus be preventing what you wish to promote.

LECTURE XXXVII.

AMYOTROPHIC LATERAL SCLEROSIS.

HISTORY:—*Disease of Charcot.* SYMPTOMS: A. *Spasmodic symptoms*: excess of tendon reflexes in the lower and upper limbs, foot-clonus, tendency of the limbs to pass into a condition of contracture, or at any rate of spasmodic rigidity. B. *Paralysis.* C. *Muscular atrophy*, its seat, existence of fibrillary contractions; electrical reactions. D. *Bulbar symptoms;* paralysis and atrophy of the muscles of the lips, tongue, and soft palate; difficulty in mastication; loss of the lateral movements of the lower jaw; distorted action of the heart; examination of the masseter tendon reflex (jaw jerk); examination of the pharyngeal reflex. No disorder of the sphincters or trophic derangement exist, frequent diminution of intellectual power, liability to emotion; the existence of symptomatic neurasthenia. COURSE of the disease; different modes of *onset:* A. By atrophy of the upper limbs; B. by bulbar symptoms; C. by spastic paraplegia. Duration of the disease. Termination inevitably fatal.

GENTLEMEN,—It may be said of amyotrophic lateral sclerosis that, like a certain goddess of antiquity, it issued from the brain of its creator in a completely armed condition; the history of this disease may be simply comprised in these three words. As early as 1865 our distinguished teacher remarked at the autopsy of some patients, who had suffered from progressive amyotrophy, that sclerosis existed in the lateral columns; in 1869, in combination with Joffroy, and in 1871 with Gombault, he observed other cases of the same kind; lastly, in 1872 and 1874, in his well-known lectures, Charcot established the existence of this affection, which he distinguished clearly from the common form of progressive muscular atrophy. In 1877 the thesis of Gombault appeared, and in 1879 the work of Debove and Gombault. From that time amyotrophic lateral sclerosis was a definite morbid condition, both from an anatomical and clinical point of view. As the lecture continues I shall name to you the principal works written about the disease since that time.

The symptoms of this disease are directly indicated by its name, which has been formed in a logical manner. Thus, *lateral sclerosis* signifies the existence, not only of paralysis, but also of spasmodic symptoms, while the term "*amyotrophic*" indicates

the occurrence of progressive muscular atrophy, so that the name is a short but exact statement of the clinical symptoms which exist in the disease of Charcot.

This affection, in fact, is one of the rare forms of amyotrophy which are associated with *spastic phenomena*. With your permission we will commence by studying these, which is the most natural course to pursue, owing to the fact that these symptoms usually occur at the onset of the disease.

The *tendon reflexes* are excessive, not only in the lower and upper limbs, but in all parts of the body, such is the case both in the foot and knee, in the wrist and elbow, and even the jaw jerk may be most excessive in degree. The excess of the tendon reflexes is carried to a degree such that associated phenomena as the *periosteal reflexes* (specially in the fore-arm, in which movements of flection and pronation are produced) and *foot-clonus*, the latter perhaps existing to a pronounced degree, may occur.

But, as you know, the knowledge of tendon reflexes has been acquired at a comparatively recent date, and yet before the discovery of Erb-Westphal, Charcot had clearly indicated the spastic nature of amyotrophic lateral sclerosis; he based this idea upon the fact, recognized by him, that more or less pronounced contractures existed in the limbs, specially the lower limbs. Some, who were opposed to the idea that amyotrophic lateral sclerosis should be considered a distinct disease, did not recognize the existence of these contractures. With regard to this difference of opinion it must be said that if we expect to meet with contractures as pronounced as those which occur in hemiplegia or spastic paraplegia, we shall often be disappointed; but if, on the other hand, we look carefully for some sign of rigidity in the upper or lower limbs this will be easily found in the great majority of cases.

This tendency to spasm has a real effect upon the *attitude* of the limbs; thus the *lower limbs* are often seen, when the patient is confined to bed, to be rigidly extended, with more or less pronounced rotation of the inner border of the foot inwards; again, when the patient walks the lower limbs may present the appearance which I have already had occasion to describe as characteristic of the spasmodic disorder of gait.

The upper limbs are usually in the position described by Charcot as follows: "the arm hangs by the side of the body,

and any attempt to separate them is opposed by the action of the shoulder muscles.

Fig. 233.—Woman affected by amyotrophic lateral sclerosis. This figure shows the atrophy of the hands and the position in which they were usually placed by the patient; the legs, in which the atrophy is much less pronounced, are close to each other, the feet being slightly rotated inwards, in short the characters of slight spasmodic rigidity are observed in the lower limbs. (Collection of Charcot.)

"The fore-arm is semi-flexed and in pronation, nor can it be placed in the position of supination without much force being employed and pain produced.

"The same may be said with respect to the wrist, which is also semi-flexed, whilst the fingers are also strongly flexed into the palm of the hand."

As an annex to these spastic symptoms tremor has been mentioned in some cases to occur in the limbs when voluntary (?) movements are made, which would be a mitigated form of the spasmodic tremor which exists in insular sclerosis. In concluding these remarks about the spastic symptoms I would call special attention, gentlemen, to the great difference of intensity which may exist in them; they exist sometimes in the form of contracture, sometimes in the form of simple rigidity of the limbs, while at other times nothing of this kind occurs, though in all

cases the tendon reflexes are found to be excessive in both the upper and lower limbs.

In addition to this spasmodic condition it must be mentioned that a certain degree of *paralysis* exists often from the first, and which is due to a lesion in the lateral columns. On this account the limbs, specially the lower limbs, are moved with much more difficulty than would be expected to occur on account of the small degree to which atrophy exists in their muscles.

This muscular atrophy, which constitutes the second phase in the symptomatic history of amyotrophic lateral sclerosis, presents a certain number of special characters.

It occurs gradually, affecting muscle after muscle, fasciculus after fasciculus, and, so to speak, fibre after fibre.

As regards its seat, the upper limbs are affected, specially the hands, in which, in fact, it commences. The thenar and hypothenar eminences are first involved; the interossei are also affected at an early date; the consequence of these muscular disorders is that the hand becomes "claw-like" ("en patte de singe") in its most characteristic form, the palm of the hand being flattened, with the first phalanx extended, whilst all the others are flexed. The fore-arm is affected in its turn, and then, as Charcot says, the upper limbs remain in pronation. As regards the arms above the elbow they are only affected later, and usually to a less degree.

The muscles of the *neck* suffer also from atrophy and paralysis, and the head is then bent forwards over the sternum, and sometimes, specially at the onset, becomes rigid in this position.

In the lower limbs the atrophy only occurs in many cases at a very late period, after being preceded, during a somewhat long time, by spastic paralysis.

Briefly, gentlemen, in order to understand the different deformities and attitudes which the limbs have in amyotrophic lateral sclerosis the following elements should all be considered: paralysis, contracture, atrophy; to this triad tendinous contraction must also be added. It may, in fact, happen that the contracture ceases to exist, whilst the muscular atrophy is sufficiently pronounced to have destroyed the greater part of the muscle, and yet the deformity which is thus produced persists;

this is due to the fact that tendinous retraction has occurred during the occurrence of these different processes.

Fig. 234.—A man affected by amyotrophic lateral sclerosis, in whom extreme spastic rigidity existed, involving all the muscles of the body. This spastic condition is indicated in the figure by the attitude of the patient, who, although the muscles of the leg were in a sound condition, could not maintain his equilibrium without assistance. The atrophy of the muscles in the hands will be also observed. (Collection of Charcot.)

The muscular atrophy which exists in amyotrophic lateral sclerosis is a form of atrophy in which the *fibrillary contractions* are extremely pronounced; this is often observed to be the case in muscles which, at first sight, appear to present no sign of atrophy; this is an important sign, which I recommend you always to investigate with great care, since it will indicate to you the muscles which will be affected in the course of the disease.

The electrical irritability varies in different cases, probably on account of the fact that, as I have already said, the fasciculi of the muscles are attacked one after the other; usually there is diminution in the electrical irritability. In some cases the reaction of degeneration is found to exist, but usually in a few muscles only at the same time, and without its being very clearly defined.

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Until the present time, gentlemen, we have specially considered muscular atrophy as it occurs in the limbs, we must now study its existence at a higher level, namely, in the parts supplied by the bulbar nerves.

The *symptoms connected with the bulb* are, in fact, of special interest in the disease which we are now considering, constituting, as they do, the special gravity of the affection.

The muscles connected with the lips (the orbicularis oris, quadratus menti, depressor labii inferioris, and levator menti)

Fig. 235.—A patient affected by amyotrophic lateral sclerosis, with most pronounced bulbar symptoms. The lips can scarcely be moved, and the mouth is always kept open; the pronounced character of the nasal furrow, the wrinkles in the frontal region, the expression of wonder in the face will be observed. It is clearly seen in the figure that the hands are affected by atrophy and spasms. (Collection of Charcot.)

are sometimes involved early in the disease; the fibrillary contractions are sufficiently pronounced in the region of these muscles to indicate the changes which have occurred in them, specially as regards the chin, which presents a series of movable depressions and elevations, which cause it to have exactly the appearance which it presents in a young [illegible] to cry.

When the changes in the mu[illegible]

owing to the effects of the paralysis and atrophy by which they are affected, the following symptoms are observed:—

The mouth is open; the lower lip droops, and seems to be pushed forward, but without being at all curved back; the upper lip is somewhat raised, so that the upper incisor teeth become visible, the whole face has a weeping appearance, which is partly due to the depth of the nasal furrow, which drags the angle of the mouth downwards; the saliva dribbles constantly from the mouth, and the patients "slaver." When they laugh

Fig. 236.—Woman affected by amyotrophic lateral sclerosis. The appearance of the mouth and chin, and the pronounced character of the nasal furrow will be observed. (Collection of Charcot.)

the lips remain at times separated from each other in such a way that they have to be brought together by means of the hand. Lastly, on account of the paralysis and atrophy of the lips the patients can neither blow, whistle, pout, or pronounce the vowels e and i.

The tongue, even when it still retains its normal size, presents
that it is first affected fibrillary movements,
notice of an experienced eye. When
ed the surface of this organ has an
ts a series of rounded eminences

and depressions, which resemble depressed cerebral convolutions. Lastly, when the paralysis reaches the most pronounced degree the tongue remains depressed and flattened behind the dental arches without being able to execute any movement.

Thus, not only does the pronounciation of the labials i, r, s, l, k, g, ch, t, d, n become very difficult, but again at certain times the speech becomes quite indistinct.

The *soft palate*, in cases in which the bulbar symptoms are very pronounced, hangs down in a relaxed condition, being often

Fig. 237.—Face of the patient affected by amyotrophic lateral sclerosis, who is the subject of fig. 233. The appearance of the mouth will be observed, which is drawn outwards, as also the depth of the nasal furrows, the wrinkles in the frontal region, and the expression of astonishment in the face. (Collection of Charcot.)

covered by dripping mucosity. The paralysis of this organ, besides making it impossible to blow or whistle, produces certain difficulties connected with speech. Thus the patients find it difficult to pronounce the letters b or p; in addition to this, and for the same reason, the tone of the voice is very much altered.

The phrases when uttered are short, and, as it were, clipped; they have lost the power of so regulating the current of expired air that a long sentence can be spoken. To this paralysis of the soft palate the troubles connected with deglutition must be partly referred, specially that which consists in the return of

food through the nostrils. Other elements, such as paralysis of the pharyngeal muscles or of those connected with the œsophagus, also take a part in increasing the troubles connected with deglutition.

At a more advanced period of the disease *mastication* is only effected with difficulty, and the *lateral movements of the lower jaw* are lost, the motor nucleus of the 5th nerve, as was shown by Duchenne (of Boulogne), being thus proved to be involved, and the lesion to be consequently seated in the upper part of the bulb.

At this time the *vagus* nerve becomes also affected, and, in consequence, troubles connected with the *respiration* (attacks of suffocation, dyspnœa) and circulation of the blood (accelerated and irregular contractions of the heart, syncope, sudden death) may occur. It is possibly on account of some alteration in the functions of the vagus nerve that the passage of alimentary particles into the respiratory tract and the *pneumonia of deglutition*, which is its consequence, are liable to occur.

We have seen, gentlemen, of how great importance the condition of the *tendon reflexes* in the limbs is in this affection, and what valuable information was derived from them as regards the state of the different segments of the spinal cord. As regards the *medulla oblongata*, similar information is given by the condition of the *masseteric tendon reflex*.

As there may be some amongst you who do not quite understand the occurrence of this reflex and the mode in which it is produced, I shall enter into some details in connexion with it from a practical point of view, since, as regards the theory of its production, there is nothing to add to what has been already said with respect to the patellar tendon reflex. This reflex is obtained by stimulation of the tendon of the masseter muscle, which may be produced in two ways. One of these consists in directing the patient to keep the mouth half open, but free from rigidity, whilst by means of a percussion hammer the tendinous origin of the masseter muscle from the posterior portion of the malar process of the superior maxilla is gently struck. Another method, described by De Watteville and Beevor, is also to cause the patient to keep the mouth half open while the medical attendant places one extremity of a paper-knife, held with the left hand, upon the teeth of the lower jaw; by means of the right hand a

slight blow is then given to the paper-knife between the left hand and the patient's teeth with a percussion hammer. Whichever method is employed, the result is that the masseter contracts, that is to say, the jaws approach each other. After but little experience it can easily be recognized whether this contraction is more or less strong, and, consequently, whether the masseteric reflex is increased or not. In amyotrophy, specially when the medulla oblongata is involved, this reflex is very excessive.

Another reflex which it is interesting to observe in cases of amyotrophic lateral sclerosis complicated by very pronounced bulbar symptoms is the *pharyngeal reflex*. This reflex is, as you know, the movement of deglutition which occurs when the back of the throat is tickled, either by the finger or a roll of paper. In cases of amyotrophic lateral sclerosis in which bulbar symptoms do not exist, or are still but slightly pronounced, the intensity of this reflex is unchanged; when the bulbar symptoms are obviously more accentuated it may be apparently diminished to a slight degree, but, whatever may be said to the contrary, it only ceases to exist, if it does so, quite in the last stage of the disease, when the muscles of the soft palate and pharynx are, so to speak, entirely destroyed.

The *sphincters* remain unaffected during the greater part of the affection.

The *cutaneous sensibility* and *special senses* are quite unaffected. As regards sensory symptoms, the only ones that can be mentioned are the very slight and also transient pains which occur in some cases; most frequently tingling alone exists.

There are no trophic derangements of any importance.

Are the *psychical functions* altered in the course of amyotrophic lateral sclerosis? Most authors deny that this is the case. I cannot, for my part, agree with them, and should not only be inclined to consider such a change as very frequent in the disease, but even as constituting one of its ordinary symptoms. Properly speaking, this change in the psychical functions does not produce any startling symptoms which *must* be noticed by the observer. If, however, one takes the trouble to seek them, a tendency to laugh or weep without any sufficient reason will almost certainly be found to exist; the liability to emotion is decidedly excessive, besides which the whole intellectual and moral condition of the patient are childish—the credulity, the

simplicity of the patient, are at times quite astonishing. Lastly, the symptoms of *neurasthenia* are sometimes observed, specially at the beginning of the disease. Always mistrust, gentlemen, the pronounced neurasthenia which occurs in a sudden and unexpected manner; when it is due neither to real misfortune, nor loss of money, nor excessive mental labour, it is almost always the unpleasant indication of a serious disease which affects the organism in its most important functions. The way in which nutrition is affected should then be carefully observed, the urine should be examined in case diabetes exists, and the different organs of the body, the nervous centres being included, should be examined. Amyotrophic lateral sclerosis is thus sometimes found to be accompanied and, so to speak, preceded by the symptoms of neurasthenia.

The *course* and symptoms of the disease vary much, both as regards its duration and mode of onset.

The onset, in fact, occurs in three ways which are quite different from each other.

A. The first symptoms which occur are in the *upper limbs*; muscular atrophy exists, which first affects the small muscles of the hands in exactly the same way as when other forms of amyotrophy affect this part.

B. The first symptoms are connected with the *medulla oblongata*; fibrillary contractions occur, the lips are not easily moved, there is slight difficulty of deglutition, &c., and it is only at a later period that the muscles of the limbs are in some way affected.

C. The first evidence of the disease consists in the occurrence of *spastic paralysis*, while muscular atrophy does not exist, or is so slightly pronounced as not to deserve notice from an objective point of view.

Thus, gentlemen, the onset of the disease occurs in three ways, which are so different that an observer who is not warned of their possible occurrence could not help believing them to be three distinct diseases.

Has this difference in the mode of onset any effect upon the course of the disease? To a certain extent it has. Thus, for example, in the cases which begin by spastic paraplegia (variety C) the amyotrophy which subsequently occurs is not usually as pronounced as in the other varieties. This, however, is not

invariably the case. Does death occur at an earlier date in the case of patients in whom the onset consists of bulbar symptoms? This is usually so, but the rule is by no means without exceptions, and it is quite impossible to connect the length of time which a patient suffering from amyotrophic lateral sclerosis has to live with the mode of its onset.

The *duration* of the disease is also, as I have already said, somewhat variable; that of the ordinary forms may be fixed at from 18 months to 2 years. In some cases, in which the progress has been very slow, it has been known to attain and even to exceed 3 or 4 years. Lastly, the course of the disease is not infrequently very rapid, and in 6 or even 3 months the disease produces a fatal result.

Death results from different causes. Some disorder of the *respiratory system* may produce it; the dyspnœa becomes extreme and, notwithstanding their great anxiety to breathe, the necessary amount of air cannot be made to enter the lungs, probably on account of some disorder in the functions of the vagus nerve. Death may, again, be due to some derangement connected with the *heart:* syncope occurs once or several times from which the patient recovers, and then takes place for the last time and the patient succumbs. Lastly, death is not infrequently due to *intercurrent disease*, either unconnected with amyotrophic lateral sclerosis or, more often, due to its existence.

Such is the case, for instance, as regards the pneumonia which is connected with the digestive process. This is due, as I have already said, to some derangement in the process of deglutition, owing to which some particles of food penetrate into the bronchi, and even into the parenchyma of the lungs. To whatever cause it may be due, the termination of the disease is inevitably death, and, whether amyotrophic lateral sclerosis is treated or not, it invariably has a fatal termination.

LECTURE XXXVIII.

AMYOTROPHIC LATERAL SCLEROSIS (*continued*).

PATHOLOGICAL ANATOMY. I. SPINAL CORD. A. Changes in the *grey matter of the anterior horns:* atrophy of the large ganglion cells, inflammation of the whole anterior horn. B. Changes in the *white substance;* (*a*) lesions of the *pyramidal tract* (direct and crossed); (*b*) lesions of the *whole antero-lateral columns*, while at times the lesions are apparently seated in the portion of these tracts which is between the anterior horn and surface of the cord, or in the part adjoining the anterior horns; lesions in the part occupied by the *columns of Goll.*

II. MEDULLA OBLONGATA. A. Changes in the *grey matter* affecting the nuclei of the hypoglossal, 5th nerve, portio dura, &c. B. Changes in the *white matter:* the pyramids, the posterior longitudinal tract (faisceau longitudinal posterieur), the band of Reil (Muratof).

III. THE PONS VAROLII.

IV. THE CRUS CEREBRI.

V. THE BRAIN. The presence of granular bodies in the internal capsule in the motor convolutions. Atrophy of the large pyramidal cells of those convolutions. These lesions at the same time are not constant. The method of seeking the granular bodies.

Theories which exist as to the NATURE of amyotrophic lateral sclerosis.

Lesions of the ANTERIOR ROOTS, the MOTOR NERVE-TRUNKS, the INTRA-MUSCULAR NERVES, and the MUSCLES.

DIAGNOSIS. A. Of the *amyotrophic form* from: the muscular atrophy of Duchenne-Aran (?); muscular atrophy due to polyneuritis; the different forms of myopathy; syringo-myelia; hypertrophic cervical pachy meningitis; arthritic muscular atrophy. B. Of the form with *spastic paralysis* from: transverse myelitis; spastic paraplegia; insular sclerosis. C. Of the form with *bulbar symptoms* from: acute bulbar paralysis; chronic bulbar paralysis; pseudo-bulbar paralysis.

GENTLEMEN,—I have already had occasion to remark that the name amyotrophic lateral sclerosis itself indicated the chief symptoms of that affection; it also contains an enumeration of the lesions which exist: *lateral sclerosis—changes in the grey matter of the anterior horns.*

I.—SPINAL CORD.

A. *Changes in the grey matter of the anterior horns.*

(*a*) The principal change, or at least the one which is most evident to the eye, consists in *atrophy of the large ganglion cells*

of the anterior horns. These cells diminish in size, lose th processes, and before long almost or quite disappear.

Fig. 238.—Normal anterior horn, with its ganglion cells, and the network of ner fibres which when coloured by Weigert's hæmatoxylin give a dark hue to th horn (semi-diagrammatic).

All the cells in the anterior cornua, at any rate in those the cervical cord, seem equally liable to be affected by th atrophic process, both those of the anterior and those of th antero-lateral group. Possibly, by studying cases of recen development it may be possible to recognize which of thes groups are first affected. In the lumbar cord many more cell as a rule remain in a healthy condition than in the cervic region.

(*b*) It would be a mistake to think that the large ganglio cells of the anterior horns are alone involved to the exclusion the adjoining parts. The *grey substance* is, in fact, severel affected throughout the whole extent of the anterior horns, an the change can be traced as far as the base of the posterio horns, and perhaps even in these parts. This alteration in th grey substance is characterized by increase in the number o nuclei, a proliferation of the interstitial tissue, and very pro

nounced disappearance of the nerve fibres, which normally form an abundant network in this part. The lesions in the grey

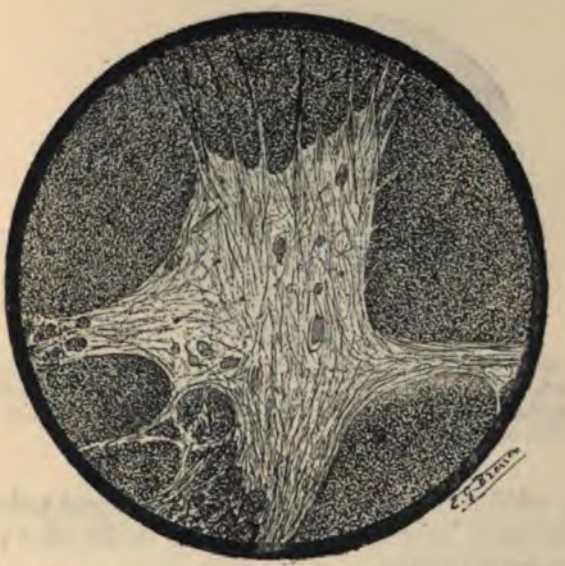

Fig. 239.—Anterior horn in a case of amyotrophic lateral sclerosis. The absence of most of the nerve cells, and the atrophy of some others will be observed: on account of the fact that a large proportion of the nerve fibres in this part have disappeared the deep colour which Weigert's hæmatoxylin gives normally to the anterior horn is not produced (vide fig. 238); on the other hand this deep hue is produced in the posterior horn (semi-diagrammatic).

matter seem also to be more marked in the middle of the anterior horn than at its periphery.

In very pronounced cases the *posterior commissure* seems to take some share in the morbid process, by which the grey matter in front of it is affected. The columns of Clarke are unaffected, both as regards their cells and reticulum.

B. *Changes in the white substance.*—(*a*) The white column, in which the lesion is by far the most pronounced, is the *pyramidal tract;* both the parts of which it is composed, the direct and crossed pyramidal tracts are affected by degeneration in amyotrophic lateral sclerosis. In this part the sclerosis is extremely pronounced, so much, in fact, that with the naked eye, and when the part has been for a short time in a bichromate solution, this lesion is the one which seems in itself, and without the use

of the microscope, to justify the diagnosis of amyotrophic lateral sclerosis in a living patient.

Fig. 240.—Section of the spinal cord (lumbar region) in a case of amyotrophic lateral sclerosis. The part affected by sclerosis seems to be almost entirely seated in the crossed pyramidal tract, being represented by a clear triangle, which exists in the lateral column.

(*b*) In addition to the degeneration in the direct and crossed pyramidal tracts lesions undoubtedly exist in the other parts of the *antero-lateral column*.

You remember, gentlemen, that when we studied the secondary descending degenerations of the spinal cord I remarked that when the degeneration was due to a transverse lesion the extent of sclerosis in the region of the crossed pyramidal tract was far greater than when this effect was due to a lesion seated in the cerebral hemisphere. In amyotrophic lateral sclerosis the same is the case, and the islet of sclerosis in the region of the crossed pyramidal tract extends far beyond the limits of that column.

The fact is, that in the same way as when descending degeneration occurs after a transverse lesion of the cord, so also in amyotrophic lateral sclerosis changes exist which extend to the extra-pyramidal portion of the antero-lateral columns. I would add that the seat of these changes is at times somewhat analogous in the two cases.

This is to say, gentlemen, that the lesions in other parts of the antero-lateral column are specially seated in the portion of that tract which is intermediate between the grey matter of the anterior horns and the surface of the cord. The sclerosis produced by them is, on the one hand, less pronounced, and on the

other less extensive than when degeneration occurs after a transverse lesion as already mentioned; for all these reasons it

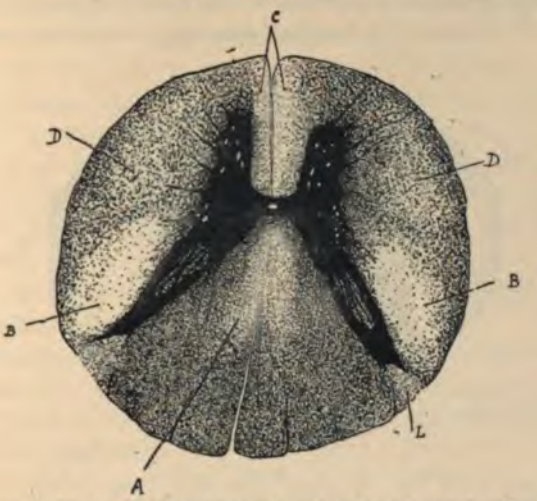

Fig. 241.—Section of the spinal cord (the dorsal region) in a case of amyotrophic lateral sclerosis. The parts affected by sclerosis are whiter in colour in proportion to the degree of sclerosis. B. Sclerosis in the region of the crossed pyramidal tract; D. sclerosis in the intermediate regions of the antero-lateral column; C. sclerosis in the region of the direct pyramidal tract; A. sclerosis affecting Goll's columns to a slight degree. L. Lissauer's tract, which, although of grey colour in the the diagram, is quite free from disease.

is difficult to define the limit of the affected part as exactly as would be desirable. In some cases the sclerosis is found not even to confine itself to the part of the antero-lateral column seated between the surface of the cord and the anterior horn, but to be seated in closer proximity to that horn, which it may almost completely surround, although always most pronounced in the lateral column properly so-called.

The condition of the *lateral limiting layer* (Seitliche Grenzschicht of Flechsig) varies; in two cases of amyotrophic lateral sclerosis, in which this part was specially observed by me, I found it almost unaffected in one, while in the other cord it seemed to be involved in the sclerotic process. The *anterior commissure* is also slightly affected in some cases.

(*c*) The condition of *Goll's columns* must be specially mentioned. These are obviously the seat of sclerosis, which is easily recognized to exist, either by the darker hue, which it

presents when coloured by carmine, or by the yellowish-brown tint which it assumes when coloured by Weigert's hæmatoxylin. This condition of Goll's columns is mentioned by many observers, and in 1885 Charcot and I myself specially noted it in connexion

Fig. 262.—Section of the spinal cord (cervical region) in a case of amyotrophic lateral sclerosis. The parts affected by sclerosis are whiter in colour in proportion as they are more severely involved. B. Sclerosis in the lateral column, extending far beyond the limits of the crossed pyramidal tract, above which a transverse band (more dark in colour) of sound tissue is found, and then, between this and the direct pyramidal tract C, which is much diseased, the whole of the antero-lateral column is affected by sclerosis to a slight degree; A. sclerosis of moderate intensity in the region of Goll's columns.

with two autopsies in which it was more clearly found to exist. I am aware, gentlemen, that in spinal cords which are free from disease Goll's columns may often be distinguished from the rest of the posterior columns by their special hue, but in that case there is no appearance of sclerosis, whereas, on the contrary, the existence of this condition is obvious in amyotrophic lateral sclerosis. I must add that in this disease the change in Goll's column is in no way accompanied by the production of granular bodies.

II.—Medulla oblongata.

We find changes of the same kind in this part as have already been found to occur in the spinal cord.

A. *Changes in the grey matter.*—The motor regions are specially altered in this part, but not all of them, since, as you

know, gentlemen, the influence of some of the motor nuclei in the bulb is so necessary to the maintenance of life that in this case death occurs before the lesions are sufficiently severe to be recognized by any of the means which we possess.

Amongst these nuclei it is that of the hypoglossal nerve in which change is most often found to have occurred (atrophy and disappearance of the ganglion cells, lesions of the very substance of the nucleus). The chief nucleus is alone affected, the nucleus of Roller, and the large cells existing along the root fibres of that nerve being in a sound condition (Muratoff).

At times the *motor nucleus of the fifth nerve* is involved (loss of the lateral movements of the jaws).

The nucleus of the *portio dura* and *posterior* nucleus *of the vagus* may also be altered to a more or less pronounced extent.

The nuclei of the nerves connected with the movements of the eyeball are unaffected.

B. *Changes in the white substance.*—In the same way as in the cord it is principally in the pyramidal tracts that the lesions occur. It should be observed that although these changes very clearly occur, their intensity cannot be compared with that which exists in cases of secondary degeneration due to a focal lesion in the brain. A much larger number of sound fibres always remains, in fact, in this part in amyotrophic lateral sclerosis than would be found in secondary degeneration associated with hemiplegia.

Muratoff also states that the *posterior longitudinal tract* of the bulb is changed to some extent, and observes the analogy which exists between this tract and the anterior column of the cord, which we have seen to be also affected in amyotrophic lateral sclerosis.

The same author mentions lesions of the same kind as existing in the raphe of the bulb and in the band of Reil (Roth).

It should be observed that in some cases lesions in the white substance are found to diminish to a considerable degree from below upwards, so that they seem to be exhausted in the medulla oblongata, above which they are no longer found to exist. In other cases, on the contrary, these lesions can very clearly follow each other from below upwards beyond the

medulla oblongata, and their existence can be recognized the pons varolii, crura cerebri, and even in the brain.

III.—Pons varolii.

When this part is affected lesions are only found to exist the white substance of the part. Lesions in the grey matt are as yet unknown. The changes in the white substance a

Fig. 243.—Section of the crura cerebri in a case of amyotrophic lateral sclerosis. C, fibres of the 3rd pair; B, cells of the black substance of Soemmering; A, lower half of the crus; D, granular bodies seated in the middle part of the crus. These granular bodies existed also in the crus upon the right side but were not indicated in order that the diagram might be more simple (half diagrammatic).

seated in the part through which the fibres of the pyramidal tract pass; the zone which they occupy is usually of very small extent.

IV.—Crura cerebri.

When the morbid process extends to this region it again affects the part through which the fibres of the pyramidal tract pass, that is to say, the middle part of the lower portion of the

crus. The lesions of the grey matter in this part, if they exist, are also unknown.

V.—Brain.

M. Koschewnikoff first noted the existence of changes in the internal capsule and white substance of the motor convolutions. The Moscow professor showed in fact, by means of a series of patient investigations, that granular bodies were found in somewhat large number in the two parts which I have just named. A short time afterwards (December, 1883), having had occasion to make an autopsy of this kind I was careful to show the existence of granular bodies in the brain and spinal cord by means of processes which enable these elements to be seen in the very seat which they occupy. The following is the result of my investigations in connection with this point, the plan of taking sections by freezing the part having been employed.*

a. In the *internal capsule* more or less numerous granular bodies are scattered through the region which anatomy has shown to be occupied by the fibres of the pyramidal tract, that is to say the anterior two-thirds of the posterior segment of that capsule.

b. In the *motor convolutions* two kinds of changes have been noted (only it must be understood in those cases in which the

* The following is the plan which I adopt: The nervous centre is placed in Müller's fluid at the temperature of the surrounding air during from 10 to 20 days. (During the last part of this period Müller's fluid will not be added but the liquid in the beaker will be maintained at the same level by pure water which is added in order to make up for the loss due to evaporation.)

When the part is sufficiently consistent (it must not be really hard, since when this is the case disaggregation of the sections is apt to occur) it is placed during from 15 to 30 minutes in a somewhat thick solution of gum arabic; after this time it is placed upon a microtome adapted to frozen tissues, and sections are made which are received into a vessel containing Müller's fluid, which is diluted and has been boiled some hours previously and then allowed to cool.

A few sections are then separated and placed in a vessel containing boiling water, Müller's fluid and the gum with which they are impregnated being thus removed from them.

The sections are then placed upon slides and may either be examined at once or, what is preferable, be stained by one of the following processes:—

A.—Mount the section in glycerine coloured by the Quinoléine (prepared by dissolving the bleu de Quinoléine in a few drops of alcohol and mixing it with glycerine, the Pierre Marie and Huet process); at the end of a few minutes it is covered by the cover glass, and under the microscope the granular bodies are found to be of a deep blue colour, which is clearly distinct from the turquoise

lesions of lateral sclerosis are found in the brain, and not in the spinal cord alone).

a. Changes in the *white substance*; this consists in the existence of numerous granular bodies disposed in radiating series like the fasciculi of fibres in that part, and directed from the centrum ovale to the cerebral cortex; in sections parallel to the large axis of the motor convolutions this disposition is very clearly seen, and it may be supposed that these series of granular bodies take the place of the nerve fibres which are affected by degeneration.

β. The change in the *grey matter* which I noted in 1883, and of which Charcot and I reported a fresh example in 1885, consists in atrophy of the *large pyramidal cells* of the cerebral cortex. In the two cases, in fact, to which I allude the section of the motor convolutions showed a much smaller number of these cells in the cortex of the motor convolutions than in a healthy person; in addition to this the large pyramidal cells which remained presented the appearance of being affected by atrophy. A short time later Koschewnikoff also stated in one of the cases of amyotrophic lateral sclerosis which he had observed the existence of these changes in the large pyramidal cells of the cortex.

Such, gentleman, are the lesions of the nervous centres which

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blue tint of the rest of the section. This colour, however, is not retained for an indefinite time.

B.—The section having been withdrawn from the water, and placed upon the slide is coloured by carmine, and after being washed, the slide is turned round so that the surface upon which the section rests is directed downwards; it is then placed upon a small cup or watch-glass which contains a few drops of a solution of osmic acid, and left exposed to the osmic vapours until it turns of a slightly brown colour; it is then again washed and mounted either in glycerine or Canada balsam.

By means of the latter process of colouring the section, which I have employed since December, 1883, permanent specimens are obtained. It consists in reality of the same elements as the "colouring of Marchi" (1885) = fluid of Müller + osmic acid. The difference between these two methods consists in the fact that in the plan which I recommend a section of the frozen tissue is made which is only coloured subsequently, whereas in the method of Marchi the section is hardened and coloured simultaneously by the combined action of the bichromate and osmic acid.

I readily admit that the latter method is the most rapid and easy for those persons who are not accustomed to the freezing process, but on the other hand I believe that the plan which I recommend is not exposed to the same causes of error and should be preferred in certain examinations.

exist in amyotrophic lateral sclerosis. How may these be connected together and their existence be explained?

After what has been said of the preference which these changes show for the whole length of the pyramidal tract this

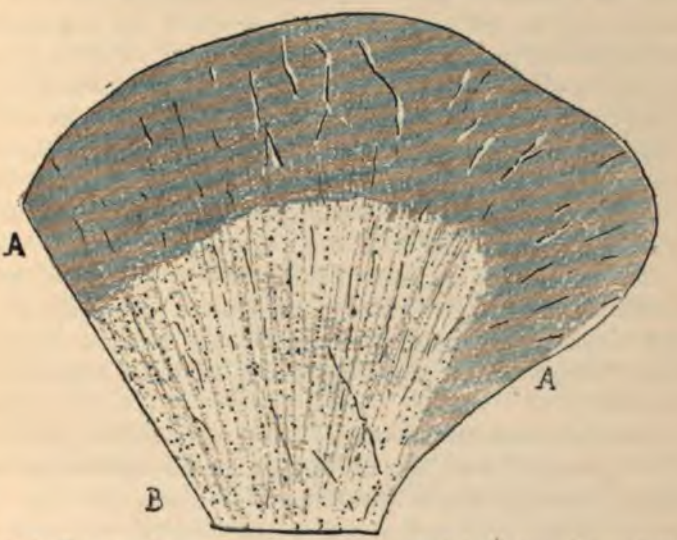

Fig. 244.—Section of a convolution (motor region) in a case of amyotrophic lateral sclerosis. This section was made in a direction parallel to that of the fibres of the white substance. Black granulations are observed in the white substance disposed in diverging linear series. The granulations represent the granular bodies which resulted from degeneration of the nerve fibres. A, Grey matter; B, white substance.

explanation is apparently quite simple; the first lesion is seated in the large pyramidal cells of the cerebral cortex; the fibres of the pyramidal tract subsequently degenerate, and this degeneration may be followed through the crura cerebri, pons varolii, and medulla oblongata as far as the spinal cord. At the same time the motor cells of the cord are affected by atrophy of the same nature as that which exists in the large motor cells of the cerebral cortex.

Unfortunately, gentlemen, this seductive theory very imperfectly explains the morbid process which produces amyotrophic lateral sclerosis, and serious objections may be made to its adoption.

We know, in fact, that, at any rate in the cord, the lesions

are by no means strictly confined to the seat of the direct and crossed pyramidal tract. In order to explain the extension of the lesions to other parts of the antero-lateral column, it has been said: 1. That fibres are affected which have been separated from the pyramidal tract. 2. That the additional fibres which are affected are commissural fibres of which the degeneration was secondary to lesions in the grey matter. 3. It may again be supposed that these fibres belong to a system which is analogous to those of which we have studied the degeneration in the antero-lateral column after transverse lesions in the cord. 4. As regards the opinion which I formerly held that the extension of the lesion is due to direct propagation of the inflammation to different parts of the cord, this I hasten to retract.

The truth is that we know nothing certain about this point; in other respects the existence of these extra pyramidal islets of sclerosis cannot be regarded as proving that columnar degeneration of the pyramidal tract throughout its whole length is impossible.

In certain cases, however—and this is more serious—the lesion of the pyramidal tract, instead of existing throughout its whole length from the motor convolutions to the spinal cord, only occurs in the cord and ceases in the medulla oblongata, the crura cerebri containing no traces of degeneration.

It might be supposed that in these cases the change in the large pyramidal cells of the cortex and the secondary degeneration, which occurs in consequence in the pyramidal tract, are first indicated by the alteration in the lower part of those fibres (intra-medullary portion of the pyramidal tract); this, however, is really most improbable and quite different from what we are accustomed to observe in cases of hemiplegia with secondary degeneration; in these, on the other hand, the changes are seen to extend from above downwards in the same direction as the fibres of the pyramidal tract.

Clinical observation again shows us that the affection may as well follow a *descending* (onset in the bulb, secondary affection of the upper and lower limbs) as an ascending path (onset in the limbs, secondary affection of the medulla oblongata).

Lastly, gentlemen, it seems to me impossible that a fasciculus of fibres can degenerate of its own accord and independently of the cells from which it takes origin (trophic cells), nor do I

believe that the pyramidal fibres can be altered in an intermediate segment of their path.

It seems so impossible to explain this difficulty that in my opinion it would be better to confess openly that we cannot explain the morbid process which exists in amyotrophic lateral sclerosis.

All that can be said is that we are no less powerless to explain why it is that in general paralysis of the insane the intramedullary degeneration of the lateral column, when it exists, can very rarely be traced as far as the crus cerebri. It appears, therefore, that the pyramidal tract is both liable to complete descending degeneration, and to ascending or descending degeneration occurring in segments, which in the actual state of our knowledge is quite absurd.

As to the admission, which would be less illogical, that the pyramidal tract is composed of two columns, the fibres of which are closely mixed together, the anatomical knowledge which we possess does not permit us to believe that such is the case.

Before ending these remarks upon the nature of amyotrophic lateral sclerosis allow me, gentlemen, to insist once more upon the fact that more than one point of contact seems to exist between this disease and the forms of general paralysis of the insane, to which I have just alluded, and in which degeneration of the pyramidal tracts, which is limited to the cord, is observed to occur (Westphal, Zacher, &c.). Analogies exist as regards the pathological anatomy; the clinical condition shows (indisputable decrease of mental power in every case of amyotrophic lateral sclerosis, even when the indication of paresis and atrophy seem confined to the limbs); these seem to me good reasons for comparing these two morbid conditions with each other. It must be well understood, gentlemen, that I do not consider these two diseases as identical or even analogous from a clinical point of view, but am convinced that they should be placed close to each other if merely considered in connection with general topographical nosography.*

Enough, however, has been said about the nature of amyotrophic lateral sclerosis, and our ignorance on this subject.

* I cannot admit, as some authors have maintained, that amyotrophic lateral sclerosis, progressive muscular atrophy, and the different forms of myopathy are varieties of one process alone, rings, as it were, of the same chain. Important differences exist between them while very few analogies bring them together.

The description of the lesions which occur in that disease will now be continued, especially those which occur in the peripheral parts of the neuro-muscular system.

The nerve fibres which compose the *anterior roots* are usually affected by atrophy to a somewhat pronounced degree.

As regards *the trunks of the motor nerves* which supply the wasted muscles, their lesions seem in complete discordance with the changes in the anterior horns on the one hand, and on the other with the alterations in the muscles, which are the consequence of these lesions; in some cases, in fact (Kronthal), these nerve trunks are apparently quite unaffected, although the muscles supplied by them may have in great part disappeared. It should be observed, gentlemen, that as Kronthal has remarked, this discordance seems only to exist as regards the nerve trunks of *medullary origin*; in the *nerves connected with the bulb*, on the contrary, very pronounced changes occur, changes which are quite analogous to those observed in the bulbar nuclei, and in the muscles which they innervate. What is the cause of this singular anomaly? A fresh problem exists which is no less insoluble than those which have already been mentioned.*

The *small intra-muscular nerves*, according to Babes and Marinesco, are affected by sclerosis to such an extent that they are no longer, so to speak, constituted by a solid cord or fibrous tissue; they contain at the same time fibres which are very narrow and fibres of large size in varying numbers. The muscles involved in the morbid process are usually affected by simple atrophy; it must not, however, be supposed that all their fibres disappear at the same time, since by the side of some fibres which are diminished in size others exist which have retained their normal dimensions. In some cases again

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It is an abuse of general pathology to join together what clinical features have so clearly separated.

On account of the failure of the different theories which have been proposed to explain the origin of amyotrophic lateral sclerosis I should be somewhat inclined to believe that vascular changes have some effect, in connection specially with the grey matter, and producing secondary changes in the white substance; it may again be supposed that one or several anatomical systems in the cord or medulla oblongata are primarily "dissolved" (??).

* Joffroy and Achard also recognized in one case an obvious want of correspondence between the degree of change in the cord and that in the peripheral nerves. (*Arch. de Méd. Expérim. et d'Anat. Path.*, 1890, II. 434.)

slight lipomatosis exists between the different fasciculi, this tendency being far more pronounced in the tongue than in other parts.

DIAGNOSIS.—The clinical features of amyotrophic lateral sclerosis are so numerous, that it will be well to consider them separately when considering the affections which simulate this disease in making the diagnosis.

A. DIAGNOSIS of amyotrophic lateral sclerosis of the FORM IN WHICH MUSCULAR ATROPHY OCCURS as regards other kinds of AMYOTROPHY:—

(*a*) The amyotrophy is combined with *diminution* or *loss* of the *tendon reflexes:*

Progressive muscular atrophy of Duchenne-Aran (?);

Muscular atrophy supposed to be due to peripheral neuritis;

Myopathies.

In these cases, the diminution in the tendon reflexes alone suffices to distinguish them from amyotrophic lateral sclerosis, and it is therefore useless to dwell upon other points of difference which enable these affections to be clinically distinguished from each other; the seat of the atrophy, the age at which the onset occurs, the presence or absence of fibrillary contractions, the electrical reactions, &c.

(*b*) The amyotrophy is combined with *excessive tendon reflexes.*

It is really as regards these affections alone that the diagnosis requires some attention.

Syringomyelia sometimes produces the same combination of symptoms, namely, excessive tendon reflexes associated with amyotrophy in the upper limbs; it is distinguished from amyotrophic lateral sclerosis by the special sensory derangements (loss of sensibility to temperature), and the trophic disorders of special nature (perforating ulcer, whitlows, dropping off of the phalanges, &c.).

Cervical hypertrophic pachymeningitis is also often accompanied by muscular atrophy in the upper limbs, and excess of the tendon reflexes; but not only are bulbar symptoms usually absent, but this affection is characterized by the existence of pseudo neuralgic pains which are never observed in amyotrophic lateral sclerosis.

As regards the more or less general amyotrophy which occurs after certain forms of infectious poly-arthritis, it suffices, gentle-

men, to recognise the existence of lesions in the joints in order to avoid any mistake being made in consequence of the exces of the reflexes.

B. *Diagnosis* of amyotrophic lateral sclerosis of the *bulbar form*, as regards other affections in which similar symptoms occur:—

Acute bulbar paralysis is distinguished by its more abrupt onset, more rapid course, by the absence in most cases of any very pronounced symptoms connected with the limbs, and the exaggeration of the tendon reflexes.

Pseudo-bulbar paralysis of cerebral origin is not accompanied by amyotrophic symptoms, but most often by a varying degree of hemiplegia affecting one or both sides; in this form of disease the pharyngeal reflex is usually much diminished or lost, whereas in true bulbar paralysis (amyotrophic lateral sclerosis) it is retained.

The analogies between *subacute or chronic bulbar paralysis* and amyotrophic lateral sclerosis are of the remotest kind, and, in my opinion, need not be discussed at greater length. At the same time I must warn you against the opinion of Leyden, according to which a form of bulbar paralysis exists which is associated with lesions of the pyramidal tract, but at the same time is quite distinct from amyotrophic lateral sclerosis. These cases in reality belong purely and simply to the latter disease, and to separate them is to inflict an injury both upon clinical medicine and pathological anatomy.

ÆTIOLOGY.—Though amyotrophic lateral sclerosis is not, properly speaking, a rare disease, it cannot be included within those which we observe daily; this will partly explain, gentlemen, some of the doubts which are connected with it.

The *age* at which it supervenes is variable, since the onset occurs at any age between 25 and 50 years, or to be more exact it may be said between 35 and 50 years; if in fact I base my observations upon cases which I have myself seen, it should be said between 35 and 45 years. It is thus a disease which occurs in the *second part of adult life;* thus you should really mistrust, gentlemen, the so-called cases of amyotrophic lateral sclerosis in which the onset occurs in adulescence or even in childhood.

The *female sex* seems more liable to this disease than the male, but without its being possible to state the exact proportion.

As regards the *cause* of amyotrophic lateral sclerosis there is nothing which indicates the reason of its existence. If inquiry is made as to the antecedents of the patients with regard to hereditary pre-disposition to affections of the nervous system no answer is usually given. Neither is the influence of any infectious disease found to be in any way connected with this disease of the nervous centres. Nor does syphilis appear to be in any way the cause. There at least remains, gentlemen, if the empty sound of a word will satisfy us, the power of stating that amyotrophic lateral sclerosis is a *disease of involution*.

What shall I say of the TREATMENT of amyotrophic lateral sclerosis?

After the observations which I have just made about the Ætiology of this affection you will spare me, gentlemen, the regret of confessing that we know nothing, absolutely nothing, of the treatment which should be employed against it. How in fact could it be otherwise when the two cardinal facts from which all efficient treatment starts the cause and the nature of this affection are equally unknown to us? Until our knowledge about these two points has increased, we must be contented, gentlemen, to observe as powerless witnesses the progress of a conflagration in the grey matter of the medulla oblongata and spinal cord which we can neither extinguish nor limit.

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