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A CASE OF SYRINGOMYELIA AND TWO CASES OF TABES WITH TRUNK ANÆSTHESIA.*

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Case I. Syringomyelia. L. M., a Swede, aged 38 years, an iron moulder, was first seen December 12th, 1897, through the kindness of Dr. G. W. Johnson. He had smallpox when six years old, and a year or two later sustained a severe fall, which rendered him unconscious for four or five hours and bedfast for two or three weeks, but there seems to have been no fracture or serious local injury. Although unusually strong previous to the present affection, he had indulged to excess in alcohol and venery, and at the age of 27 or 28 contracted venereal sores of indeterminate character. This was repeated three years later, there being, so far as can be learned, on neither occasion any secondaries; nor is there any history of subsequent symptoms indicative of syphilis. On several occasions he had been severely chilled by exposure to cold drafts when greatly heated by his work, having fallen asleep where he lay down to rest.

In his opinion the present trouble began during the summer of 1889 or 1890. In the spring of one of these years (he is not certain which) he was made foreman, and in consequence had no occasion to do manual labor. In the autumn he resumed his former work, and then noticed that his back and

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arms were not so strong as formerly. He had some difficulty in lifting and handling heavy objects. In all probability the disease of the cord had begun before this, for during the year preceding the first noticeable weakness he was never free from felons. These affected principally, but not exclusively, the right hand. Without apparent cause, and *without pain,* a finger would swell to about twice the natural size, feel clumsy for some days, finally discharge pus, and gradually heal. No sequestra came away, but on one occasion a good-sized slough, that looked like a tendon, separated, and when this was "pulled out by the roots" it hurt, but not excessively. In talking with the patient about it, I received the impression that this operation was very much less painful than it would have been in a normal person. What is still more conclusive is the fact that two or three years before the first motor symptoms appeared, he had a whitlow on the middle finger of the right hand, followed by diffuse cellulitis, for which three incisions were made, notably one on the dorsum of the hand about an inch and a half in length; and that these incisions were not very painful. About a year after he first noticed the diminished strength, he developed what was called erysipelas of the same hand and forearm. For this two free incisions were made and a large drainage tube drawn through. Neither the inflammation nor the operation occasioned suffering, although he cannot say that they were entirely devoid of pain.

Gradually the weakness of the back and arms increased, the left arm, he thinks, being rather worse than the right, and during the first year power in the legs also began to fail, as he can remember that sometimes after the day's work he was scarcely able to walk home, and found ascending steps particularly difficult. The ankles seemed weaker than the knees and hips. For two years after the beginning of the disability he was able to continue at his trade, with the exception of six weeks at about the end of the first year. At this time he awoke one morning to find the right arm and hand almost completely paralyzed. This seems to have been an ordinary pressure (sleep) paralysis, which allowed him to resume work in six weeks, although he thinks that the arm never fully regained its former usefulness. After this the right arm and hand were less efficient than the left. It should be remarked that three years previous to this sudden disability of the right arm, he had had a similar but less severe paralysis of the same arm, which I attribute to the same cause, and which kept him from work only two or three weeks. He was at the time of these attacks a steady and excessive drinker.

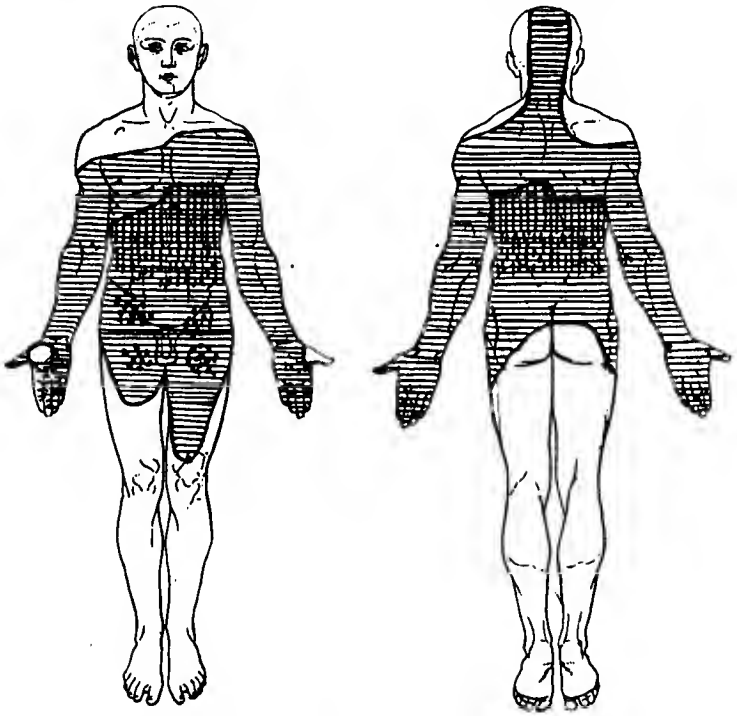
During the two years after the onset of the disease, when he was still occupied as a moulder, he constantly had blisters

on his hands, and was greatly puzzled because, as he expresses it, "his hands blistered so much easier than those of the other workmen." This was doubtless due in great part to the already existing analgesia. He was never conscious of burning himself; had not the warning of normal sensation, and consequently the blisters seemed to appear without adequate cause. I am inclined to think, however, that the skin of these patients may in reality blister with abnormal facility, as is the case in many other forms of paralysis. The tendency is seen in examination of the patient, for ordinary pin-pricks cause great weals to arise within a few minutes, and with a test tube of hot water I raised several blisters.

The power of the upper extremities has slowly decreased to the present time, the disability being much more pronounced about the shoulders than in the hands, but the patient affirms that the strength of the legs has improved somewhat during the last year. About a year and a half ago the hands, more especially the right, began to show vasomotor disturbance. They would frequently get red or dusky, and become somewhat swollen. The feet also became slightly enlarged, so that the patient was obliged to increase the number of his shoes from nine to ten. There has been no pain at any time, and no spasm or twitching, although he has noticed that after grasping an object for some time, the hand tends to cramp in the same position. Sexual power has been lost for four years, having begun to fail quite a year before its extinction. This seems relatively early for syringomyelia.

Examination reveals a sufficiently typical picture of this disease. Both shoulders droop, the right rather more than the left, the arms are pendulous, suggesting by their position and motion as the patient walks progressive muscular atrophy or dystrophy; the gait is typically spastic, but he "toes out" a little beyond the normal, and the feet are everted and flat, the right more so than the left. From the lower part of the neck down, all muscles seem to be more or less paretic, but very unequally so. The legs are more spastic than weak, the patient still being able to walk two or three miles at a slow rate, while the musculature of the upper arms and shoulder girdle is almost completely useless. Although he shrugs the shoulders with considerable power, the right trapezius above the shoulder, and the left one higher up, are decidedly atrophic and correspondingly weak. Indeed, at these parts they cannot be seen to contract at all. Both supraspinati and the right infraspinatus are wasted; the scapulæ seem too far from the spine, the borders are not parallel, and the patient is practically unable to approximate them, that is, to draw the shoulders well back. The deltoids are markedly atrophic, but although

the left is apparently more wasted than the right, he can raise the left arm almost to the horizontal for a moment, while abduction of the right is reduced to almost nil. Rotation is exceedingly poor, and worse on the right side. Biceps and triceps are wasted, flabby and very weak, the posterior muscles being rather weaker than the anterior, but even flexion is so feeble that feeding himself is a considerable task. Although the two sides in this location are much alike (circumference, R. 11¼, L. 11 inches), some movements at the shoulder and el-

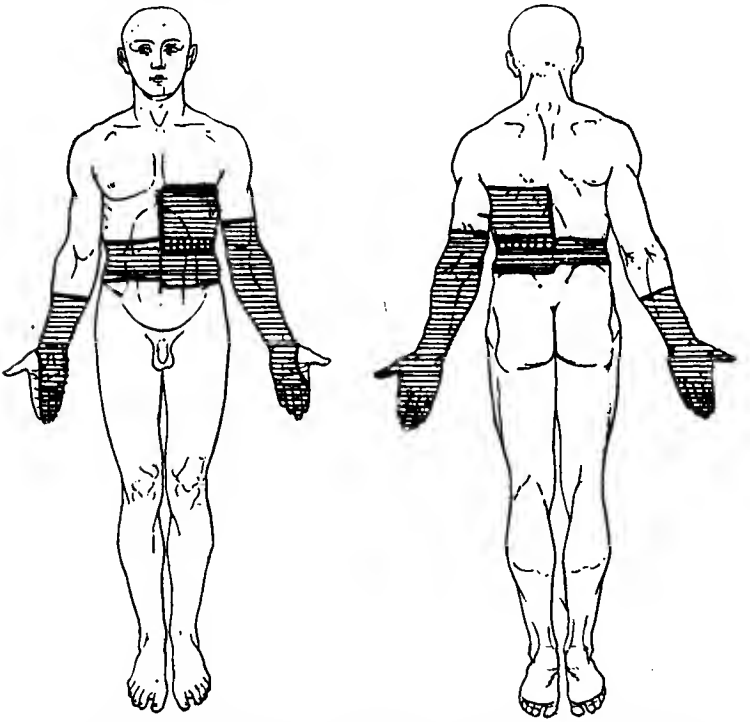


FIGS. I AND II. SYRINGOMYELIA.
Horizontal shading indicates analgesia. Perpendicular shading indicates tactile anæsthesia.

bow are better on one side and some on the other. The pectorals are not very strong, but could not be called atrophic. The forearm muscles are in a decidedly better condition than those of the upper arm. Atrophy is not apparent here and the grasp is fair, but the extensors on the left side are distinctly weaker than on the right, as in grasping strongly the hand flexes at the wrist, as it does in wrist-drop, although not to

an equal degree. Pronation and supination are quite defective, supination more so than pronation, and both worse on the right side. Spreading and approximating the fingers are feebly executed, more feebly on the left.

The cranial nerves are intact, including pupillary reactions and visual fields for white, blue, red and green. As in all cases of this disease, the sensory conditions are interesting. They are indicated in brief by the diagrams (Figures I to IV), which, however, must be supplemented by a few words.



FIGS. III AND IV. SYRINGOMYELIA. (Same case shown by Figs. I and II). Horizontal shading indicates total loss of pain sense. Perpendicular shading indicates anæsthesia to firm touches.

First, the band of tactile anæsthesia about the trunk, although unmistakable, is not absolute. That is, to demonstrate and outline it, very light touches are necessary. The lighter the touch, the broader the anæsthetic band, and if quite firm touches are made with a camels-hair brush, no anæsthetic zone is evident. At no point on the body is there complete tactile anæsthesia. I may add that, as is the case with trunk

anæsthesia in tabes, the anæsthetic area is wider when examined from its middle toward the borders than when the latter are determined by approaching the zone of anæsthesia from above or below, where sensation is normal. Aside from this variation, the lower border of the anæsthesia is so uncertain that no exact limit can be determined. Second, although diminution of the pain sense is distinct quite to the limits indicated by Figures I and II, analgesia is not complete in all of this area. In Figures III and IV is shown the extent of complete analgesia. Third, sensation for both touch and pain is decidedly better on the right than on the left side. Fourth, the tongue of analgesia extending from the dorsal area to the vertex is not unique, although examples of it seem to be very rare. Gilles de la Tourette and Zaguelmann[1] report a case in which analgesia of the nape of the neck and back of the head existed as an isolated area, and Sölder[2] has recorded two cases in which the analgesia extended upward in a way quite similar to that shown in Figure II. In one of these the area gradually spread until it included the entire head except the face, a distribution that is not very exceptional. It should be added that, aside from variations already cited, there are spots of relatively small size, apparently located at random in the anæsthetic and, especially, in the analgesic areas, where sensation is more acute than in the surrounding parts. The patient seems to be rather more sensitive to the faradic brush than to other painful impressions. Loss of the thermic sense practically coincides in distribution with the analgesia, except that it seems to be less uniform in degree.

Plantar, cremasteric and abdominal reflexes are absent. The knee-jerks are exaggerated, as are also the Achilles jerks; there is incomplete ankle clonus on both sides. The wrist-tap (radius flexor reflex) is absent. From the regions indicated by dots in the diagrams, a lively reflex is excited by pin-pricks, although these are not at all painful, and occasionally this was noted in pricking the trunk about or below the level of the umbilicus.

The hands are "pudgy," thick, clumsy looking, a little on the "spade" order, inclined to be cold and cyanosed, but they are not deformed. The puffiness is resistant, and does not pit on pressure, and there is no enlargement of the bones. The feet present a similar appearance, and there is, besides, pes planus with, on the right side, so much eversion of the foot and protrusion of the inner bones of the tarsus as to suggest an arthropathy. There is no other joint trouble and no more lateral curvature of the spine than occurs in many normal

[1] Nouv. Iconog. de la Salp., vol ii., p. 311.
[2] Neurolog. Centralb., June 15th, 1898, p. 571.

persons. In the examination nothing has been discovered except the anæsthesia that would suggest leprosy. The patient says that often rotation of the head is accompanied with a grating in the neck, and this can be felt by the observer placing a hand upon the patient's head or neck. The feeling is something between that of crepitus and a click.

Changes in the electric reactions are not striking. All the atrophic muscles show diminished response to the faradic current, and in the atrophic portions of the trapezius the contraction is distinctly slow. These same portions (of the trapezius) show degeneration reaction to galvanism; that is, the response is very slow and the contraction long persisting. Other muscles respond by quick contractions.

CASE II. TABES. A. K., married, 52 years old, was first seen March 13th, 1898, through the kindness of Dr. E. R. Bennett. About 25 years ago he contracted a venereal sore, followed by a suppurating bubo and a number of enlarged glands in either groin. The inguinal and postcervical glands are now slightly enlarged, also the epitrochlear on the right side. There is no further evidence of syphilitic disease to be discovered either in the history or examination.

The first intimation of the present disease seems to date back about seven years, when, after dancing at a picnic, the legs felt tired out of all proportion to the exertion, and there was a sensation of tension or drawing about the calves and popliteal spaces. (This seems to be a frequent symptom of incipient tabes.) After this the same feeling was noticed when he was at work; as the patient expresses it, he felt as if he had walked a thousand miles. In the course of a year or so bladder symptoms appeared. With a sudden call to micturate there would be incapacity to start the stream promptly, and involuntary escape of a few drops of urine was not infrequent. A little later he began to have at longer or shorter intervals a sensation as if some one had suddenly gripped the left calf, and later still an occasional stinging pain in the leg or foot. Typical shooting pains have never been present. Five years ago, from no apparent cause, the left great toe became greatly swollen and congested; after a few days it "broke," giving exit to a little dark, bloody serum, and then rapidly became gangrenous. It was amputated by Dr. Bennett, and the wound healed promptly. There was some pain at the time of the acute swelling, but no particular attention was paid to the sensory conditions at that time. Shortly after the operation the left leg became enormously swollen, almost to the knee, but this swelling disappeared in a few days, and after several weeks he went back to work. It was then noticed that the left foot was everted, and that there was a

marked bony protrusion at the inner side of the instep. A few months after the operation a dusky swelling appeared on the stump, which discharged a dark serum for a short time, and then healed. At about this period the patient found that in putting on his trousers he had to steady himself by bracing his head against the wall. Nine months after the loss of the great toe the second toe on the other foot became swollen and dark, then turned black, and it was amputated three weeks after the first change. Union was prompt and perfect. By the time he had recovered from this incident control of the legs was too poor to allow resumption of his occupation. For about a year longer he got about with the aid of sticks and (later) crutches, but for the last three years has been confined to a wheeled chair, which he propels with the arms. He can now neither walk nor crawl, has had no sexual power for a year, and for the last two months there has been rectal incontinence when the bowels were loose.

Achilles-jerk, knee-jerk, wrist-tap and plantar reflex are absent, the abdominal reflexes are exaggerated. The left pupil is larger than the right, and there is reflex iridoplegia. Incoördination is very pronounced in the lower extremities, less marked but distinct in the upper extremities, and seems unusually prominent in the pelvo-femoral muscles. Sense of position is very much impaired and muscle tonus greatly diminished. The left trapezius muscle is atrophied in its upper (cervical) part, and the left arm is also somewhat wasted, being 1½ cm. less in circumference than its fellow. There is entire absence of the pectoralis major on the right side. Of the latter muscle, there is a strong strand arising from about 2½ inches of the clavicle; not a vestige below this. I consider this anomaly to be a congenital defect, as in the aplastic parts there is not the least trace of muscle, and yet the patient has never been conscious of any disability. As a carpenter he used plane and saw without inconvenience, and as a young man struck out from the shoulder like his companions. He says, too, that his mother in making clothes for him when a child remarked that he had a crooked chest.

For me, the interest in the case is almost confined to the sensory conditions and the spontaneous loss of the toes. Cases I and II taken together and considered in connection with some cases of syringomyelia, afford food for reflection and future comparison. A year ago I reported to this association[3] a case of syringomyelia

[3] Journal of Nervous and Mental Disease, October, 1897.

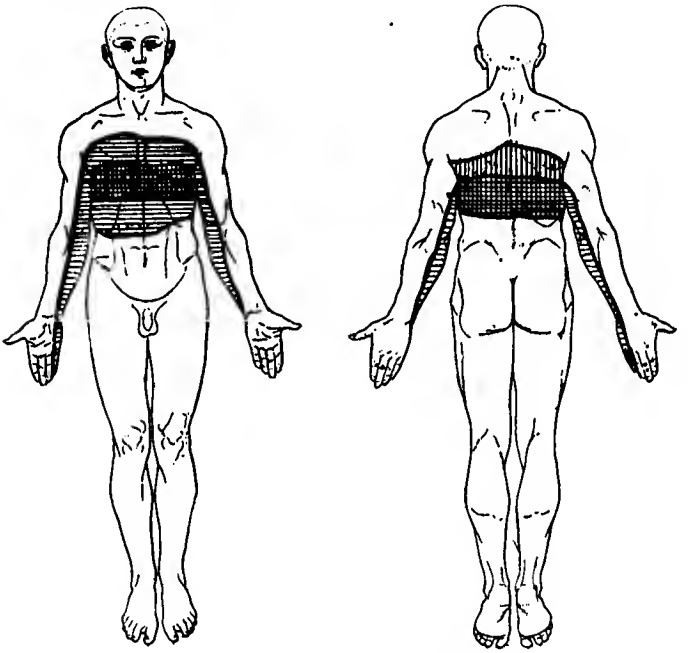
with trunk anæsthesia, and the preceding case (Case I.) constitutes an additional example of the same thing. The first one showed anæsthesia and analgesia more closely corresponding to the tegumentary sensory representation of the segments of the spinal cord than any reported up to that time, and, so far as I know, such a striking case has not been reported since. It showed the identical distribution often found in tabes, with the striking difference that the area of analgesia was large and that of tactile anæsthesia a narrower zone in the middle of the analgesic surface. The exact reverse ordinarily obtains in the trunk anæsthesia of tabes, as shown by Laehr,[4] Bonnar[5] and myself.[6]

The present case and the following one constitute striking exceptions to this rule. At least in the present state of our knowledge of this symptom in tabes, the topographical relation of analgesia and tactile anæsthesia shown by these two cases must be regarded as very unusual, and much more closely resembling that found in syringomyelia.

In A. K. (Case II.) the analgesia extends in front from the lower border of the second rib almost to the umbilicus, and includes the inner surface of the arms, stopping at the wrist on the left side, but covering the little finger of the right. (Figures V and VI). The analgesia on the arms, however, is not so marked as on the trunk, and the line bounding it is most indistinct; it is impossible to say exactly where it begins. The zone of tactile anæsthesia extends from just above the nipple to the xiphoid cartilage. Behind, the sensory condition is more like that prevailing in tabes. The tactile blunting extends higher, while its lower border practically coincides with that of the analgesia. Queerly enough, the latter extends on to the arms, and the former does not. Considering this anom-

[4] Arch. f. Psych., 1895, Bd. xxvii., Heft 3.
[5] New York Medical Record, May 22d, 1897.
[6] New York Medical Journal, February 6th, 1897.

alous distribution, it is proper to say that a number of examinations revealed the same condition. On the legs there is the usual dulling of both tactile and painful impressions, the former being the more in evidence. Sensory conduction is delayed, and impressions not at first painful often produce a burning sensation that is decidedly disagreeable.



FIGS. V AND VI. TABES.
Horizontal shading indicates analgesia. Perpendicular shading indicates tactile anæsthesia. The ordinary anæsthesia of the lower extremities is not indicated in the diagrams.

Atrophy of muscles about the neck and shoulders is not frequent in tabes, and the wasting of the upper part of the trapezius is of some interest, as being almost the exact counterpart of that found in the case of syringomyelia (L. M.). Probably still more exceptional is gangrene of the toes in this disease. Pitres[7] has reported an

[7] Revue Neurol., 1893, p. 202.

nstance, and so has Kornfeld.[8] The case of the latter, however, was one of acute neuritis added to tabes. Joffroy and Achard[9] have recorded a case of spontaneous gangrene in tabes, but the disease was already in the terminal stage; the patient had "pied-bot tabetique" with extreme tension of the skin due to the deformity, and died three days after the gangrene supervened. Besides, the gangrene in this case was not "in mass," but simply a gangrenous ulcer. Indeed, it seems strange that the occurrence is not more frequent, considering some of the other severe trophic accidents of tabes, such as the perforating buccal ulcer with bone exfoliation reported by Letulle and Lérmoyez,[10] Hudelo[11] and Leo Newmark,[12] and considering that there would seem to be principally differences of degree between perforating ulcer of the foot, whitlow with sequestrum and gangrene of the toes. The case that I report was once shown in a medical society as a probable example of Morvan's disease, and although I am not aware that the diagnosis between this disease and locomotor ataxia has given rise to difficulty, that between tabes and syringomyelia has been embarrassing to more than one observer.[13] Even in the present instance, the sensory symptoms, the localized atrophy of the trapezius and the trophic lesion of the toes might, at a certain stage of the affection, have caused the diagnostician to hesitate.

CASE III. TABES. This case is sufficiently typical in every respect, excepting the sensory conditions on the trunk and arms. The patient is a man, 45 years old, who contracted a small venereal sore about 25 years ago, can recall no further symptoms indicative of syphilis, and first noticed slight incoördination of the legs seven years ago. Included in the history are pains in the lower extremities, transient ptosis and

[8] Semaine Méd., November 9th, 1892, p. 442, and Wien med. Presse, December 11th, 1892, p. 1,986.

[9] Arch. de Méd. exper et d'anatomie path., 1889, No. 2, p. 24.

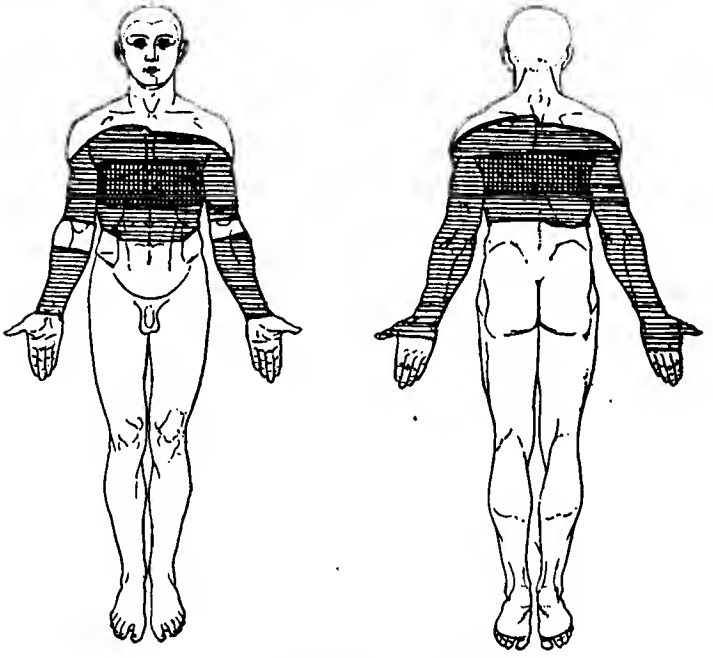
[10] Med Week, 1894, p. 355.

[11] Soc. franc. de derm. et de syph., May 13th, 1893.

[12] Med. News, January 26th, 1895.

[13] Parmentier: Nouvelle Iconographie de la Salpêtrière, vol. iii., 1890, p. 213. Bruns: Neurolog. Centralb., 1897, p. 511.

diplopia, impaired sexual power, and a moderate degree of cystic incompetence. At present there are small Argyll-Robertson pupils, loss of the knee-jerks, well marked ataxia of the lower extremities, slight incoördination of the upper extremities and complete analgesia of the ulnar trunk. On the legs the sensory blunting is that usually found in cases of tabes not far advanced; that is, tactile anæsthesia is very slight, analgesia very distinct, and both gradually decrease in degree



FIGS. VII AND VIII. TABES.
Horizontal shading indicates analgesia. Perpendicular shading indicates tactile anæsthesia. The anæsthesia on the lower extremities is not indicated in the diagrams.

from the feet upward. On the trunk and arms the distribution of impaired sensation is much like that already described as occurring in syringomyelia (Figures VII and VIII). Not only is the area of analgesia much more extensive than that of tactile anæsthesia extending from the trunk on to the arms (the tactile anæsthesia being limited to the former), but on the latter it presents the segmental distribution, with border at right angles to the limb, often seen in syringomyelia, but rare in tabes. In this case also the persistence of the peculiar sensory conditions has been confirmed by repeated examinations.

I regret to say that I have no explanation of the distribution and character of the sensory symptoms in these cases that is at all satisfactory to myself, nor have I even an opinion that is new. At the last meeting of the association Dr. Knapp[14] presented an able paper covering similar cases, and I have now nothing to add to the several hypotheses and opinions then presented, but simply present these instances as a clinical contribution to a subject not very well understood.

DISCUSSION

Dr. P. C. Knapp said that a year ago he had read a paper before the association on the subject of sensory disturbances in cord lesions; he had been unable to throw much light on the subject then, and now he knew even less about it. Certainly, the study of sensory disturbances, especially in tabes, shows that they do not follow any definite course. Sometimes we get the well defined sensory disturbance of the so-called spinal distribution, at other times disturbances of the stocking, or glove, or sleeve type; sometimes such symtoms are absent entirely, or we may get one set of symptoms one week and another the next. In some cases we get analgesia; in others, analgesia with more or less anæsthesia or hyperæsthesia. As a rule, in tabes the analgesia is more extensive than the anæsthesia.

We certainly see cases of syringomyelia with analgesia, and considerably later in the course of the disease there is anæsthesia in a more limited area than the analgesia. In hysteria it is common to find analgesia on one side of the body without any anæsthesia.

In conclusion, Dr. Knapp said it was a question in his mind whether analgesia is the result of a slight disturbance of the sensory tract and anæsthesia is the result of a much greater disturbance, or whether we are dealing with two separate tracts, one for pain and one for tactile sensibility.

Dr. B. Sachs thought the view that the well known sensory symptoms of syringomyelia were almost pathognomonic of that disease could no longer be entertained. The speaker said he had observed two cases of Pott's paralysis, with dissociated sensory symptoms confined to the extremities; in two other cases dissociated sensation occurred first in the distribution of the trigeminal nerve. In one of these cases, seen about a

[14] Journal of Nervous and Mental Disease, September, 1897.

year ago, there was absolute loss of pain and temperature sense in the distribution of the trigeminal nerve, while in other parts of the body sensation remained entirely normal. The speaker said he had no satisfactory explanation to offer for these unusual cases. In another class of cases he had observed symptoms which resemble those of syringomyelia, inasmuch as they have their origin in the cervical region of the cord, but they become absolutely stationary, and remain so for long periods—in some cases at least five years. An unusual feature of these cases is that the sensory symptoms are not as typical as they are in many cases of syringomyelia. Almost every form of sensation is somewhat involved, and there is not that sharp distinction between the analgesia and the anæsthesia that there is usually in syringomyelia.

Dr. Sachs said he wished to inquire whether cases of dissociated sensation within the distribution of the trigeminal nerve had been observed by any one else, and whether such cases developed the symptoms of syringomyelia later on.

Dr. Frank R. Fry said he had seen two cases of dissociated sensory disturbance of the fifth nerve. In neither of the two cases was any trophic disturbance noticed; there was merely a disturbance of sensation in the region of the fifth nerve. In one case this was more marked in the upper than in the lower division of the nerve. In the latter region it seemed impossible to produce pain.

Dr. Patrick, in closing, said he did not agree with the statement made by Dr. Knapp, if he referred to the impaired sensation on the trunk, that in tabes, as a rule, the analgesia is more extensive than the anæsthesia. On the trunk tactile anæsthesia is more extensive than the analgesia, and appears sooner, while in the lower extremities the reverse holds good. By some the view is held that the analgesia is due to a less pronounced involvement of the sensory tracts than that which causes anæsthesia, but this explanation is hardly adequate.

Dr. Patrick said that the dissociation of sensation on the extremities in several diseases was not very uncommon; for example, in Pott's paraplegia, multiple neuritis and tabes.