Three cases of nervous disease / by T.K. Monro, M.A., M.D., Physician to the Glasgow Royal Infirmary.

d from Glasgow Hospital Reports, 1901.

THREE CASES OF NERVOUS DISEASE: (1) CEREBROSPINAL MENINGITIS; (2) FRIEDREICH'S HEREDITARY ATAXY; (3) MARIE'S HEREDITARY CEREBELLAR ATAXY.

BY T. K. MONRO, M.A., M.D.

THREE CASES OF NERVOUS DISEASE: (1) CEREBROSPINAL MENINGITIS; (2) FRIEDREICH'S HEREDITARY ATAXY; (3) MARIE'S HEREDITARY CEREBELLAR ATAXY.

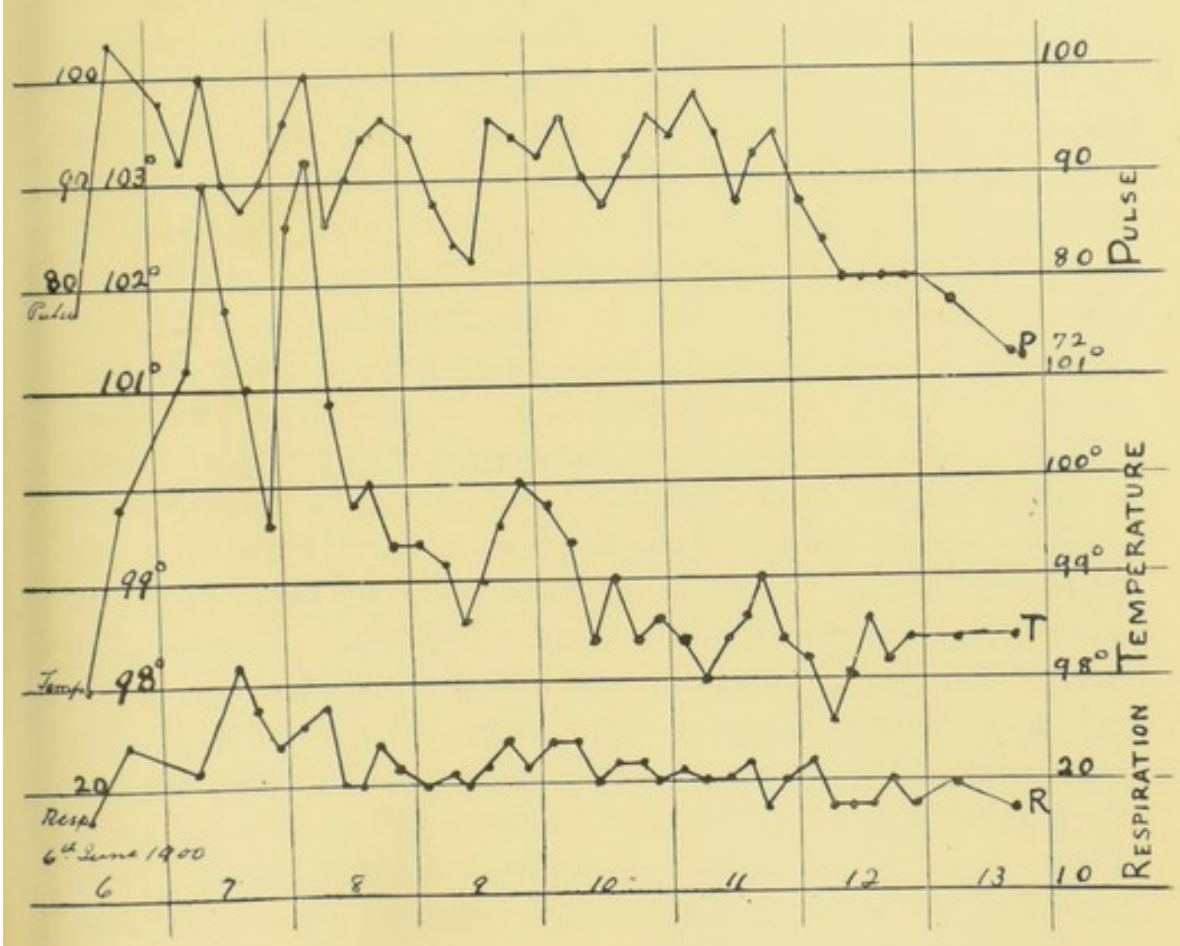
By T. K. MONRO, M.A., M.D.,
Physician to the Glasgow Royal Infirmary; formerly Examiner in the University of Glasgow.

(1) CEREBROSPINAL MENINGITIS.

Maggie C., aet. 14, was admitted to the Royal Infirmary on June 6th, 1900, on account of pain in the head and vomiting. She had been seized on the evening of the 3rd with shivering and giddiness, and an hour or two later with severe headache and vomiting, which persisted, with short intermissions, for about twelve hours. A doctor who was then summoned administered a powder, with the result that patient remained in a state of stupor for nine or ten hours. From the end of that period until her admission, she was very restless, and only conscious at times. She would start up and grind her teeth, and there was twitching of the limbs. The bowels were confined, and the urine was high coloured. No cause could be suggested for her illness, but it is perhaps worthy of note that in the middle of the day on which she took ill, she took onions, an article which, she said, never agreed with her, though she felt no inconvenience until nine or ten at night. Her previous history was a good one: she had measles in infancy, but never scarlet fever, or rheumatism, or any nervous ailment. The family history was excellent.

7th June.—Patient is at present to all appearance completely sensible, but is very restless, moving her trunk, tossing her limbs, and working her lower jaw almost constantly. The

face is much flushed, and the tongue dry and somewhat foul. Her principal complaint at present is of the bad taste in her mouth. Unless when questioned, she does not complain of pain, but she seems to have some about the back of the head or neck and in the left flank, and there is in addition tenderness on pressure in the left ovarian region and in the epigastrium. Nothing abnormal is detected on examination of the heart or lungs. There is an abundant eruption of herpetic vesicles around the right margin of the mouth: this appeared



on the morning of the day of admission. In the sitting posture, the head is thrown back, and patient is unable to move the chin downwards towards the sternum. When the head is passively depressed, patient complains of pain in front of the neck. The difficulty here is not due to any obvious spasm of the muscles of the neck. Patient generally lies with the knees and hips flexed, but it is possible for her to lie with these joints completely extended. Whenever she sits up,

however, flexion sets in, and when attempts are made to extend the limbs passively, patient either complains of pain or falls backwards. The abdomen is somewhat, though not extremely, retracted, and it appears that the bowels have not been moved since the 2nd inst. No rash is observed on the skin. The pupils are medium-sized or rather small, and contract actively to light. There is no strabismus and no detectable palsy of the face or tongue. It is noted that part of the right border of the tongue as well as the mucous surface of the upper and lower lips is involved in the herpes. A rough testing of hearing with the watch points to a considerable loss in each ear. There is no optic neuritis. There is distinct nystagmus when the eyes are rotated far to the left, and slightly on rotation upwards. There is none on rotation to the right. There is some tenderness over the spine immediately below the occipital bone, but not at any other point. The temperature rose from 98° on admission to 101·2° this morning, the corresponding pulse rates being 78 and 99, and respirations 19 and 22. Urine: specific gravity 1024, reaction acid, colour amber; flocculent sediment; slight trace of albumen; no sugar or blood.

8th June.—Giddiness is still present, though not so severe as at the onset. There is no intolerance of light or sound. There has been no delirium since admission. Patient slept during most of last evening and the early part of the night, getting perhaps in all seven hours' sleep, but since then she has been restless and uneasy and suffering much from pain all over the head, but specially about the occiput and back of the neck. There is great tenderness over various parts of the back, including two or three of the upper cervical spines and several of the lumbar spines. The tenderness is perhaps still greater at a little distance from the spine, and especially on the right side. It is also present in the suprascapular region on each side and in the lateral regions, and very specially in the right lumbar region external to the erector spinae. There is spontaneous pain also, especially in the lumbar region of the back. The pain is aggravated by movement, and may even be aggravated in the head by movement of the spine. In the front of the trunk there is tenderness to-day in the right ovarian region and

epigastrium. Patient speaks of spontaneous pain in the hips also. There is tenderness in the knees, but these joints can be passively moved without pain. The ankles are free from pain. The knee jerks are present. In the plantar reflex there is extension of the left great toe and also of the right (yesterday there was flexion of the right and extension of the left). The abdominal reflexes are deficient, but their investigation is interfered with by what appears to be slight hyperaesthesia of the skin of the abdomen, although, speaking generally, there is little or no hyperaesthesia of the skin. The eruption of herpes has been already alluded to. In addition, a certain amount of livid erythema was noted yesterday over the extensor surface of the left elbow. A similar appearance is present to-day over the right elbow as well, and to a certain extent over the extensor aspect of the knees. A similar condition is better seen to-day on the dorsal aspect of the right forearm at its lower part. This extends on to the back of the wrist, and is present in a more patchy form on the back of the forearm and hand, and is also faintly recognisable on the back of the left wrist. Patient lies preferably on the right side, with the hips flexed almost to a right angle and the knees flexed to about 135°. An effort to sit up this morning revealed continued retraction of the head, and almost immediately induced severe pain over the back and in the region of the stomach, which compelled patient to lie down at once. There is no noticeable rigidity of the limbs, and there is no discoverable palsy. There is no trismus. There have been no convulsions, either general or local. About 2 a.m. to-day, patient suddenly wakened out of sleep with a cry which seems to have been due to a sudden attack of pain which has been more severe in the head than elsewhere. The bowels were moved yesterday by enema, and have since then moved spontaneously. The bladder seems to be somewhat irritable, as patient makes more frequent attempts at micturition than are necessary. The herpes which is present on the right side of the face has not been associated with subjective sensations. Apart from the herpes no trophic lesion is detected in the area of distribution of the fifth nerve, but a slight conjunctivitis is indicated by a small quantity of pus at each inner canthus. There is no

enlargement of liver or spleen. Tache cérébrale, if present, is not very persistent. There is no hiccough or sighing respiration. The tongue, though still much furred, is moist and considerably improved as compared with yesterday, when patient was complaining of a bad taste. The parts on which patient rests are reddened in either a diffuse or a papular manner, and this is doubtless largely contributed to by her very restless condition. There is nothing, however, in the way of actual necrosis of tissue. There have been no mental symptoms since admission.

Patient states spontaneously that her throat has been sore for several days; it troubles her not so much in swallowing as in speaking.

9th June.—Patient is a good deal easier this morning, and is now able to sit up for a time. She is still unable to lower the chin to the sternum, and it is now ascertained that this disability is associated with spasm of the complexi muscles. The splenii, sterno-mastoidei, and trapezii are free from spasm, but there is much tenderness over the right sterno-mastoid and supra-spinous regions. Tenderness has passed away from the upper cervical spines, but is still present lower down. The only present seat of spontaneous deep pain is the knees. Constipation has now passed away.

11th June.—Great improvement in symptoms accompanied the fall in temperature, and, as far as the subjective aspect of the case is concerned, patient is now perfectly well. She is able to carry out normal movements without discomfort. The herpes on the face and tongue is rapidly healing. Some of the other cutaneous lesions are still recognisable, and in particular livid erythema on the right upper limb, red papules over the left olecranon and on each knee, and a vesicle below the right external malleolus.

No abnormality was ever detected in connection with heart or lungs.

After patient recovered, her hearing was tested more carefully than was possible in the height of the fever, and was found to be very defective. Thus, as tested with a watch, R. $\frac{1}{2}\frac{1}{3}$, L. $\frac{1}{4}$. Rinne's experiment gave a positive result, but the excess of air conduction over bone conduction was

diminished. The membranes were found to be greatly retracted. Nevertheless patient asserted that her hearing was perfect before the illness which has just been described. She never had any discharge from the ears. The presumption is therefore that there was an old-standing dry affection of the middle ear, and that the present disease caused further impairment of hearing by damaging the internal ear.

Treatment in the febrile stage of the disease was by means of laxative enemata, two grains of calomel every eight hours, cold applications to the head and spine, and rest on either side or in the prone position instead of on the back. Later on, salicylate of sodium was given in combination with perchloride of iron.

The symptomatology in this case seems sufficiently characteristic. The headache, vomiting, delirium, retraction of the head, Kernig's sign, constipation, spinal tenderness, pain in the back aggravated by movement, etc., all point to cerebro-spinal meningitis; and the sudden onset with shivering and giddiness, the fever, the herpes, erythema, and other cutaneous lesions, the pains in the joints, and deafness complete the resemblance to the epidemic disease, of which this may be regarded as a sporadic example. Bacteriology might have settled the diagnosis beyond question, but an attempt at lumbar puncture was unsuccessful, one of the difficulties being the severe burning sensation experienced by the patient on the application of a freezing agent to the skin of the back.

(2) FRIEDREICH'S HEREDITARY ATAXY.

Jane H., aet. 25; no occupation. Admitted to the Royal Infirmary, 2nd April, 1900, complaining of dizziness, with weakness and stiffness of the legs.

Past history and condition on admission.—Patient belongs to a healthy family, and had no serious illness until the present trouble set in at the age of 15, after a severe fright which she sustained through being chased by two men on a dark night. From that time she was unable to carry on her occupation as domestic servant. Her first symptoms are described as dizziness and light-headedness, and these were constantly present,

but were specially troublesome when she walked in the dark, thus causing her to stagger and sometimes to fall. The next symptom was occasional pain about the upper part of the sternum, gradually passing upward into the neck and giving rise to a sense of constriction of the throat; this only troubled her on physical or mental excitement, and was relieved by a drink of water or by loosening the neck of her dress. When aged about 20, she began to have slight shooting pains in the lower limbs from the knees downward, and especially in the ankles and feet; these pains, which are still felt at times, were of momentary duration, and did not occur oftener than once a day.

About the same time, she first suffered from a feeling, sometimes painful, as of a band round the body; this has become more severe of late, and is constantly present, though aggravated by excitement or exertion. It is sometimes associated with a sense of abdominal distension, which compels her to loosen her dress at the waist. The principal seat of this girdle-sensation is the epigastrium. The upper limit is about the level of the fourth costal cartilages, and the lower limit about midway between the xiphoid and the umbilicus.

Since the New-Year, patient has been subject to a severe pain, which occurs in paroxysms at the right lower costal margin, towards the back, and when this sets in patient ceases to be sensible of the abnormal sensation in the epigastrium. She states that the severe pain in the side comes on only when she is walking and happens to look to the ground; it is associated with dizziness so severe that she falls backwards. She has occasionally had pain in the stomach after food, but did not vomit at these times. The bowels have long been constipated. When she is going about on her feet she has an almost constant inclination to pass water, and occasionally, if she does not make haste to empty the bladder, urine will escape involuntarily, though not without her knowledge. There is no trouble of this kind when she is in bed, and there is no delay when she wishes to evacuate the bladder. Menstruation has always been regular.

There is well-marked slowness of speech, of which patient is quite aware, though she has no idea when it commenced. At

times there may be noted a tendency to separate the syllables of words, but the main defect is simply slowness of utterance.

Though patient complains of both weakness and stiffness in the *legs*, no definite weakness can be detected as she lies in bed, and no tonic or clonic spasm can be recognised in these limbs; but when she walks there is a perceptible tendency to drag the feet, and especially the right foot. The freeness of movement in the toes and the slight disorder noticed in the manner of putting down the feet suggest that the difficulty in walking is due to incoordination more than to weakness, and this is borne out by the fact that she cannot keep her balance when she tries to stand with her feet close together,—a disability which is rather increased when she shuts her eyes. She can stand however, with the feet apart, even if the eyes are shut. In walking, as in standing, the ataxy is distinct but not extreme. There is no club-foot. The right knee jerk is normal or slightly sluggish; the left is diminished. The plantar reflex is represented by flexion of the toes on the right side, and is absent or represented by extension of the great toe on the left side. The sense of posture in the legs is good, but the common muscular sense appears to be much diminished in the muscles of the calf. The ground feels normal to the feet. When the various other forms of sensation in the legs (touch, pain, temperature) are examined, it is not possible to detect any localised defect, but there is a general lack of acuteness, as shown, for instance, by the way in which patient may attribute the touch of the finger to a pin point, or may fail at one moment to recognise the piercing of the skin by a pin, while showing a little later that the sense of pain in the same part is well preserved. After a little practice, patient indicates when she is touched with almost no delay more than the normal. There is no history of perverted sensibility in the legs, such as a sensation of pins and needles. The legs when exposed readily show a livid marbling. There is no muscular wasting.

The only symptom referable to the *upper limbs* is occasional slight pain or stiffness at the right elbow and right metacarpo-phalangeal articulations, which began about a week ago. There is no localised palsy or wasting.

There is well-marked curvature of the *spine*, viz., kyphosis

in the dorsal region and lordosis lower down. In addition, there is slight skoliosis, with the convexity to the right at the level of the scapula. When she pushes with the left arm, a groove appears along the inner border of the left scapula chiefly at its lower part. No such groove is seen when the upper limbs are at rest by the side, and the effect of a strong pushing effort is not only to make the lower scapular angle more prominent, but also to tilt it slightly inwards (paresis of serratus magnus). There is no trace of any such weakness on the right side, and there is no evidence of weakness of the rhomboids. The power of the trunk muscles, as tested by efforts to raise the head and shoulders in the prone and supine positions, is regarded as normal. It is perhaps worthy of note that patient finds she can keep her balance much better in walking when her hands are placed over the front of her chest than when they are allowed to hang at her sides.

The abdominal and epigastric reflexes are well marked.

Cranial Nerves.—i. Smell is preserved.

ii. V.A. Right and Left each about $\frac{6}{36}$. This defect may perhaps be explained by the hypermetropia or hypermetropic astigmatism which is recognised by the ophthalmoscope. No hemianopia. Fundi normal. Examination of the visual fields shows only a slight restriction in the outer parts.

iii., iv., and vi. The functions of these nerves may be regarded as normal. The pupils are equal and somewhat large; each contracts directly and consensually to light; each contracts in convergence of the visual axes; and each dilates when the skin of the neck is stroked, either on the same or on the opposite side. Occasional slight jerks are seen when the eyeballs are moved laterally, but this scarcely amounts to nystagmus. There is no diplopia.

v. Motor function apparently normal, but difficult to investigate because of the difficulty in making patient understand instructions given. Common sensation in the region of this nerve normal. Taste is not very acute. Thus, while she recognises bitter and to some extent salt and acid solutions, she is scarcely able to recognise a sweet taste.

vii. There is a slight flattening of the left naso-labial furrow, but no other evidence of weakness of the muscles of

the face. The difference between the two sides is if anything less marked when patient tries to show her teeth.

viii. Hearing good in both ears.

ix., x., xi. There is no evidence, as judged by the condition of deglutition, respiration, vocalisation, etc., of any involvement of these nerves.

xii. Movements of the tongue normal.

There are no definite *mental* symptoms. The one feature in her case which suggests anything of the kind is her inability to comprehend some of the instructions that are given her in the course of examination. To this may perhaps be added the slowness of speech already described.

The only noteworthy feature in the chest is the presence of a V.S. murmur at the pulmonic area and to a less marked degree at the aortic area and manubrium sterni. Lungs normal. Temperature normal or subnormal. Urine contains a faint haze of albumen.

In the course of her residence, which extended over several months, patient's condition improved in some ways. Thus the slight pains in the right upper limb ceased, and the girdle-sensation also passed away. The attacks of pain in the right side became slight and infrequent, and any difference between the two sides of the face became inappreciable. The shooting pains in the legs were replaced by abnormal sensations in the feet and ankles, which at some times took the form of a sensation of pins and needles, and at other times of simple soreness. In the main, however, the symptoms referable to the nervous system persisted. The plantar reflex was for a time at least absent. The knee jerks were inconstant. The tendency was for both, but especially the left, to be deficient, even when reinforced. But the right was sometimes normal, and on one occasion seemed to be if anything exaggerated. The left knee jerk was sometimes slightly diminished, and at other times almost completely absent. There was no trophic lesion of skin or joints.

Examination by faradism and galvanism of different nerves and muscles of the lower limbs revealed no evidence of disease.

The haemic murmur became less marked, and an examination of the blood at this time gave the following results:—Fresh

blood microscopically examined presented normal appearances. Haemoglobin 70%. Red corpuscles 4,666,000, and white corpuscles 6400 per c.m. In a stained specimen, the four principal varieties of white corpuscles were found to be present in normal proportions.

The urine continued to show a slight haze of albumen.

The treatment was at first by arsenic with nux vomica. About six weeks later, this was replaced by phosphorus pill, for which, five weeks afterwards, Blaud's pill was substituted.

While the symptoms in this case are in the main sufficiently characteristic, one or two unusual features are present. The diagnosis of Friedreich's hereditary ataxia seems justified by the incoordination in the lower limbs with a comparatively slight degree of weakness, the late and trifling involvement of the upper limbs, the commencement at puberty, the slowly progressive course, the affection of speech, the spinal curvature, and it may even perhaps be added, the basic cardiac murmur. Among other evidences of spinal cord disease are the pains in the legs and trunk, the constipation, and the impaired control over the bladder. A very unusual feature, however, in this case is the preservation of the knee jerks, though it will be noted that there is deficiency on one side. Another departure from type is witnessed in the fact that this girl alone of her family suffers from the disease. A tendency to nystagmus is just recognisable. Other points which deserve attention, though not to be discussed here, are the different curves of the spine, the paresis of the left serratus magnus, the doubtful paresis of the left midface, the slight defect of taste and of tactile and painful sensation, the persistent slight albuminuria and the continuance of regular menstruation.

(3) MARIE'S HEREDITARY CEREBELLAR ATAXY.

Alexander S., aet. 15, was admitted to the Royal Infirmary on 12th April, 1900. Patient is the second child in a large family of children otherwise healthy, and born of healthy parents. He had measles in childhood, and thereafter enjoyed good health until aged about 12, when he had enteric fever. He never had scarlet fever. At about 13, it was noticed that

he became giddy after walking a short distance, and had to stop or hold on to some support. Patient seems to have been first conscious of his illness one day in school, when he was seized with shivering, which was followed by loss of feeling in the feet and legs. The disorder in the lower limbs gradually increased, so that he staggered when he walked, and sometimes fell. He was in the Victoria Infirmary for some weeks in 1899. He has had no pain or sickness, and there is little or no loss of control over the bladder and rectum. The bowels tend to be relaxed.

The following data were obtained after his admission to the Royal Infirmary. Patient's principal complaint is of weakness in the legs and feet, and indeed of weakness all over, but there is no localised paresis of muscles. There is no tonic or clonic spasm, and no tendency in walking to lift the feet unduly high. There is only a little tendency to drag the right foot. The difficulty with the legs is due to incoordination, which is such that he cannot stand with his feet together, and requires, when walking, to keep the feet far apart. The unsteadiness on standing is not much increased on closing the eyes. The knee jerks are greatly exaggerated, but no ankle clonus is obtained. The cremasteric, abdominal, and epigastric reflexes are diminished. The plantar reflexes are not obviously disordered, but are difficult to investigate on account of patient's sensitiveness to tickling.

The different forms of sensation in the lower limbs (touch, pain, temperature, posture, common muscular sense) are all normal, and there is no delay in the conduction of tactile impulses.

The *upper limbs* are unaffected.

The muscular weakness is most manifest in the trunk, being shown by the inability to sit upright for any length of time, but it can also be recognised in a slight degree in flexion of the ankles. The extensor muscles at the hip and knee retain great power, as is shown by the little assistance required to enable patient to step on to a bench. Both in sitting and on standing there is marked kyphosis, the head and shoulders being thrown far forwards, as if to counteract the tendency of the trunk to fall backwards. The extensor muscles of the

neck are powerful, and patient is able by an effort, when he is asked to do so, to make the back quite straight, at least when he is sitting in bed. He is quite unable, however, to maintain the sitting posture for any length of time, and tends to fall backwards and to the right. The power of raising the head from the bed, when the body is in the prone position, seems to be rather less than normal. There is no lateral curvature of the spine. There is no detectable atrophy or pseudo-hypertrophy of any set of muscles.

A certain amount of *intellectual defect* is present. Patient states that when he left school a year ago he was in the fifth standard. Whether this is correct or not, his arithmetic is now very rudimentary, and he fails to answer correctly such a question in multiplication as 7 times 5.

Another feature in his case is the defective power of attention, which is readily recognised in course of conversation with him. He is slow to answer questions, partly perhaps through lack of attention and partly through lethargy of mental processes. He is very emotional.

Although his *speech* is somewhat slow, there appears to be no defect in articulation as regards words or letters. There is no tremor of the tongue.

Cranial Nerves.—i. Smell appears to be acute, but it is difficult to investigate on account of the mental defect.

ii. Vision—Right, $\frac{5}{2\,0}$; Left, also about $\frac{5}{2\,0}$. O.E. normal.

iii., iv., and vi. There is no strabismus. Patient speaks of having once seen an object double, but no reliance can be put upon his statement. Nystagmus is present in well-marked degree on lateral movement, and to a less degree on upward movement of the eyeballs, but not spontaneously. Each pupil contracts directly and consensually to light. The pupils also contract in convergence. They are medium-sized and equal.

v. Taste in front of the tongue for sweet, salt, and bitter is preserved.

vii. Facial movements normal.

viii. Hearing good.

ix., x., xi. No symptoms referable to these nerves.

xii. Tongue normal.

Examination of the heart, lungs, spleen, liver, and abdomen generally yields normal results.

Urine normal as regards reaction, specific gravity, and absence of albumen.

The *bowels* still tend to be loose, the motions being watery. There is some defect of control over the sphincter ani, so that, although patient always gives notice, he has on one or two occasions been unable to retain the evacuation until attended to. There has been no such trouble with the bladder.

Temperature generally subnormal—never febrile.

In its more obtrusive features, such as the incoordination in the lower limbs, commencing in early life and slowly progressive in course, the muscular weakness and the nystagmus, this case is at first suggestive of Friedreich's disease. The great exaggeration of the knee jerks, however, together with the striking loss of power of the muscles of the trunk and the intellectual defect, seems to ally this case more closely with the special group which Marie called hereditary cerebellar ataxy, and which he regarded as depending upon some congenital defect in the cerebellum.