Aortic disease of the acquired type is not as rare in children as is commonly supposed, and some of the cases are no doubt of congenital origin. Another point of clinical interest, a recognition of which will prevent falling into error, is that aortic disease in children is apt to be associated with a bruit which is heard loudest over the pulmonary area, and not over the aortic cartilage.

Hypoplasia of the aorta is characterized by diminution in size of the aorta and arteries throughout the body, the walls of which are thin and elastic.

A congenital origin has been claimed for this condition, because it is so commonly associated with anomalies in the generative and circulatory systems. A congenital tendency to the dwarfing of the arterial system has been ascribed to tuberculosis in the parents, and it may be due to other states of bad health.

Hypoplasia of the aorta is associated with a dwarfed heart in some instances, and in others with an enlarged heart confined to the left side or involving both ventricles. If the heart be small during childhood it hypertrophies at puberty. The period of danger in these cases appears to belong to adolescence, when heart failure is not unlikely to occur, and this is more likely to arise under physical strain.

Defects in the auriculo-ventricular valves are very uncommon. They are commonly combined with other cardiac abnormalities, uncomplicated lesions of the mitral and tricuspid valves being exceedingly rare. Of the two valves the tricuspid is more frequently attacked than the mitral, and of the lesions tricuspid atresia is the commonest.

Tricuspid stenosis in association with disease of the pulmonary valves is occasionally seen.

Mitral atresia is very rare. Cases have been recorded by Rokitsansky, Thérémis, and by Lawrence and Nabarro, and in each instance the condition has been associated with somatic abnormalities.

A specimen of *congenital mitral stenosis* was exhibited at the society for the Study of Disease in Children by Fisher in 1901.

Of *mitral insufficiency* Cautley has afforded an example in a girl aged 22 months. The physical signs were those of mitral regurgitation, and another case in a boy, aged 10½ months, has been quoted by Maude Abbott.

Hypertrophy of the heart undoubtedly arises as an idiopathic disease of congenital origin. Simmons was the first to record a case of its existence at birth. He found the heart of a newly born child, who died during the labour, to be enlarged to twice its natural size, all other organs of the body being quite healthy. Prior to that Hauser had recorded a case in a baby, aged 11 months, in which the heart was enormously dilated, with hypertrophied right and left ventricles. Virchow suggested that the hypertrophy might be congenital.

Cardiac displacements such as *dextrocardia* and *mesocardia* are the only congenital malpositions that are of interest to the physician and therefore that need be reviewed. *Dextrocardia*, accompanied by transpositions of the viscera, is commonly viewed rather as a rare developmental freak than recognized, as should be the case, as a not very out-of-the-way clinical experience.

Dextrocardia without associated transposition of the viscera is rare, and not only is it rare, but it must be viewed with suspicion, because the diagnosis of congenital *dextrocardia* has been made on some few occasions, and this opinion has not been confirmed by autopsy.

Sometimes the position of the fetal heart in the centre of the thorax is maintained, giving rise to the condition known as *mesocardia*. The apex beat will then be felt in the epigastrium, and the position of the organ will readily be demonstrated by the *x* rays.

At the eighty-first annual meeting of the West Riding Medical Charitable Society, of which Dr. Gibson, of Harrogate, is now President, it was reported that forty-eight grants, amounting in all to £1,680, had been made during the year, and that since the foundation of the charity the total sum disbursed in grants had been £46,295 10s. Of those on the society's list of grantees four had died during the year, and five new applications had been received. Mr. Herbert Rowe, who for twenty years was co-secretary with the late Mr. Claude Wheelhouse, was unanimously elected Honorary Treasurer, and Mr. Michael Teale appointed Honorary Secretary.

THE PERONEAL TYPE OF MUSCULAR ATROPHY

WITH AN ACCOUNT OF A FAMILY GROUP OF CASES.

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THE following cases of hereditary muscular atrophy belong to a family in which it appeared in at least four generations; the cases are examples of the still unclassified peroneal type first closely studied by Dr. Howard Tooth in this country, and by MM. Charcot and Marie in France.

The patients are a man, aged 45, and his two daughters, aged respectively 17 and 21; the remaining members of the group include the grandfather, mother, and younger brother of the present head of the household, all deceased.

CASE I (Father).

G. H. S., aged 45, married for eighteen years, has six children, three girls and three boys. One girl died in infancy of whooping-cough. The other children, the youngest of whom is 2 years of age, are living, but the two daughters are the subjects of the same condition as their father; the others are apparently sound. The patient is a cardboard box cutter, and for fourteen years held the position of foreman in the works. He is a man of more than ordinary intelligence, has been steady all his life, and, it may be taken as certain, has not suffered from any venereal disease.

Family History.—His father is still living and is 70 years of age; he is somewhat alcoholic and has been so for many years. The mother died when 50 years of age of heart disease; she suffered from clubbed feet. Such particulars as could be gathered of her case are set forth under Case V. An only and younger brother—there were no sisters—died, when 32 years of age, of pneumonia; he was similarly affected in the lower extremities, and particulars, so far as they could be obtained, are recorded under Case IV.

The following is the patient's family tree, details of the affected members of which follow:

History of Present Illness.—Up to 8 years of age the patient was quite healthy; he then had measles. Soon after recovery weakness of the feet was noticed, and he gradually came to walk on the outer edge of the feet. At 15 years of age he attended an orthopaedic hospital, and was given special boots, which were worn for two years. At 17 he went to another hospital and underwent an operation for the deformity, after which he wore iron supports for three years. He was able to walk fairly well on the level ground, but experienced a little difficulty in going upstairs. From adolescence up to the age of 32 the condition seems to have remained stationary, and he was able to attend regularly at work. Thirteen years ago, when 32, he began to "feel himself going all wrong," he became conscious of increasing weakness in the legs, and then noticed they were getting thinner, especially the left. At this time he also noticed commencing weakness in the hands, which showed itself in clumsiness of movement on attempting to pick up small articles. Gradually the backs of the hands became thin and hollows appeared between the metacarpal bones. He found difficulty in feeling small articles in the dark, so that he was often unable to take matches out of a matchbox, to pick up a coin, or to fasten his collar. About eight years ago an ulcer formed on the outer side of the right sole; this several times healed under treatment, only to break down again on using his foot. At varying intervals during the last thirteen years he has had attacks of pain down the limbs and also round the waist, and these have been more frequent of late. He was able to work up to February, 1904, when, in consequence of the extensive ulceration of the right foot, he was compelled to lay up. On recovery he found himself much weaker and unable to walk far without feeling exhausted.

State on Examination.—In 1904 the following notes were made: The patient's general condition in regard to his respiratory, circulatory, and digestive systems is good. Bladder and rectum are normal in their functions. Sight and hearing are good. The trunk muscles are well developed.

Lower Extremities.—Both feet are clubbed, and there is also claw-toe deformity, the big toe of each foot being most affected. He is unable to abduct or adduct the feet, is only able to extend the feet to an extremely limited degree, and there is complete loss of power to extend the toes. Both legs show marked wasting, the left more so than the right. In both legs the external fibular muscles, namely, the peronei, are markedly

Fig. 1.—Case 1. Showing the atrophy of leg muscles, more extreme in the left than in the right, with consequent loss of the natural contour; the club-foot and claw-toe; and the Aran-Duchenne hand.

atrophied, the tibialis anticus and extensor muscles have also suffered; in the left leg the calf muscles show a greater degree of atrophy than those of the right (Fig. 1). The natural contour of the legs is quite lost. Both knee-jerks are absent. On the inner aspects of both legs at about the lower third the skin is somewhat brownish and glossy. The legs always feel cold to the touch.

Upper Extremities.

—Both hands show advanced atrophy of the thenar and hypothenar muscles, and also of the interossei and lumbricales. He is unable, therefore, to extend the fingers, the attempt to do so bringing them at once into the claw-like position. In the right hand the thumb can be made to oppose all the fingers, but in the left the thumb is unable to oppose the little finger. The forearms are thin, but there is no inequality between them. Patient is able to perform all

the wrist movements without any difficulty. Fibrillary tremors have been observed in both hands. There is scoliosis of the spine with the convexity to the right.

Sensation.—Over the soles of both feet there is complete loss of sensation to touch and heat, but not apparently to cold, and there is delayed sensation to the pin-prick. Over the dorsal aspect of the feet, the front and back of the legs, there is a general impairment of sensation to touch and heat; a pin-prick is more readily recognized, and, on the whole, the right leg is better than the left, the thighs better than the legs, and the right thigh than the left thigh. Sensibility to touch, temperature, and pain is quite normal over front and back of trunk, face, head, and neck. Over the palmar and dorsal aspects of the hands there is impairment of sensation to both touch and temperature; a pin-prick is recognized as such, but there is delay. Over the arms and forearms no appreciable loss of sensation can be determined.

Electrical Reactions.—With the faradic current the interossei muscles of the left hand give no reaction, and the thenar and hypothenar muscles act feebly. In the right hand there is a faint reaction in the interossei and in the thenar and hypothenar muscles. In the forearm muscles there is diminished reaction. The upper arm muscles act feebly, but only with a strong current.

In the lower extremities no reaction is obtainable in the muscles of either the thighs or legs. In the intrinsic muscles of both hands the galvanic reaction gives KCC > ACC, and the reaction is not sluggish. In the leg muscles there is no reaction to galvanism, even with a strong current. An especially noteworthy feature is that, while there are electrical changes in the atrophied muscles, those muscles not obviously atrophied, such as the thigh muscles, cannot be made to act with strong currents, such

as could not be borne by a normal healthy person. There is inco-ordination of muscular movements both in the upper and lower extremities; Romberg's sign is present. The pupils are small and unequal, react very sluggishly to light but briskly to accommodation.

State on Examination, November 11th, 1907.—He was then admitted into the Prince of Wales's General Hospital, N., under the care of one of us, largely to attempt to obtain sound healing of the right foot. It was a long ulcer on the outer and under surface of the right sole. There was the scar of another in a similar position on the left foot, which had just healed. (See Fig. 2.) On that date he had a fairly healthy appearance, but was somewhat pallid, was inclined to be stout, and had a rather expressionless face, although his intelligence was unquestionable. The body organs were again found to be healthy. The pupils were unequal, the left being larger than the right, which was "pinpoint," and both showed the Argyll Robertson phenomenon. The optic discs were of a good pink colour, and vision was quite good. The atrophy and weakness were merely somewhat more marked than on the previous examination. Fig. 3 shows the change in bulk. There was tactile anaesthesia downwards from just above the knee on the right side and from the junction of the middle and lower third of the leg on the left side. Sensibility to pain was preserved, but there was marked delay on both sides as high as the middle of the thigh. There was tactile anaesthesia of the hands as high as just above the wrists. There was no ulnar anaesthesia in the forearms. There was apparently no thickening of any accessible nerve trunk. A special electrical examination was made by both of us with Dr. Gerald Burke, the house-physician, on January 27th, 1908. With the hand of one of us on the patient's cheek over the exit of the facial nerve and the coils separated 1½ in., a vigorous contraction was obtained in the thenar muscles of the examiner, and a very distinct sensation, but there was no contraction in the patient's facial muscles and no sensation. A good reaction in the examiner's facial muscles was obtained with one electrode in the left hand and the other in the right hand, the back of which was applied to the side of his own face, and that with the coils separated less than 2 in. With the coils pushed nearly home, the electrodes being one in the patient's hand and the other in the palm of the examiner's hand, the back of which was placed on the patient's face, there was an extremely vigorous contraction

Fig. 3.—Case 1. Showing double club-foot, double claw-hand, wasting of muscles, and pigmentation of the skin.

Fig. 2.—Case 1. Showing perforating ulcer of right foot and scar of another on left foot.

and painful sensation on the part of the operator, but on the part of the patient hardly any sensation and no muscular contraction. To galvanism a good contraction was obtained in the examiner's first interosseous muscle of the hand, using the negative pole and the indifferent electrode in the other hand with 2.75 m.a. of current. With the indifferent electrode on the patient's back and the other electrode applied to the same muscle of the operator's hand, which was resting on the

patient's biceps, a contraction was obtained in the examiner's muscle with 2.5 m.a. With the active electrode then applied directly over the patient's biceps no reaction was obtained with nearly 20 m.a., neither with ascending nor descending currents. A doubtful reaction was obtained in the first interosseous and a clear one in the third of the patient's hand, with about 20 m.a. and ACC = KCC. An examination made by Dr. Howard Pirie, electrician to the hospital, on February 3rd, 1908, gave the following results: A maximum faradic current causes slow and slight contraction in the thenar muscles, the first, second, and third interossei, and the hypothenar muscles of the left hand. No muscle of the left forearm would react. A tender spot was elicited with this current over the deltoid motor point, but no contraction. No reaction was obtained from any face muscles except a slight one from the right *risorius*. No contraction was obtained from any muscle of either thigh or leg. The right hand gave the same negative result, but a contraction was obtained from the right flexor carpi ulnaris and the flexor minimi digiti with some flexion of the other two inner fingers. Several tender spots were localized in the left forearm.

CASE II (Daughter).

A. L. S., daughter of the patient just described, is 16 years of age. She had whooping-cough when 3 years old. From this time up to the age of 10 she enjoyed perfect health, was able to run about and play with other children. Soon after she was 10 it was noticed she was becoming less active, and there was a tendency to walk on the sides of her feet. About a year later weakness was noticed in the hands.

State on Examination, February, 1904.—The patient was pale and somewhat thin, but intelligent and cheerful in disposition. The muscles of the legs below the knees were distinctly wasted, but measurement did not reveal any inequality between the two limbs. When the patient was lying down or sitting with the legs dangling the loss of power in the peroneal and extensor muscles was more manifest, both feet showing foot-drop. Slight talipes varus was present with some pes cavus (Fig. 4). She was unable to dorsiflex the right foot, and the left foot only to a slight extent, and she was not able to stand on tiptoe. The knee-jerks were absent, even with reinforcement. Cutaneous sensibility over the soles showed some impairment, but elsewhere it was quite normal. In the upper limbs there was no obvious atrophy. There was, however, commencing weakness in the hands, which had been evidenced by the fact that whereas a few months before the patient was able to open the spring of a neck fur with her hand she was now unable to do so, but pressed the spring open with her teeth. There was commencing scoliosis with convexity to the right. There was no inco-ordination of movements; the pupils were normal; there was no nystagmus.

Electrical Reactions.—The intrinsic muscles of the hands reacted to faradism, although somewhat feebly. In the right leg the peroneal and extensor muscles reacted fairly, the tibialis anticus faintly. In the left leg the results were substantially the same.

To Galvanism.—The right and left thenar muscles responded only fairly, but KCC > ACC. Owing to the patient's extreme nervousness the electrical examination was difficult, but the general result was that there was diminished irritability to faradism.

State on Examination, November, 1908.—Since the foregoing notes were taken the girl has increased greatly in weight and height. Her legs now drag markedly in walking, the great toes are constantly turned up (claw-toe), she wears away the soles of her boots on the inner side before the rest, and her hands have become flapper-like, so that when using them to remove her clothing she is obliged to use the whole hand, not the fingers alone.

CASE III (Daughter).

The third case is that of the elder daughter of the first patient. She is 21 years of age, and a big, well developed woman. It does not appear that she has suffered from any acute illness in the past. She is healthy looking and well nourished. She is apparently (November, 1908) just beginning to follow the course of her younger sister. She is somewhat clumsy in her gait, the calf muscles are well developed, but there is weakness and some wasting of the muscles on the outer side of the legs below the knees, and the knee-jerks could not be obtained even with any method of reinforcement. An electrical examination was not made because of the apparently insuperable objection of the patient to that method. The hands are not obviously affected.

CASE IV (Brother).

W. J. S., younger brother of the first patient, was quite healthy up to the age of 16, then clubbing of the feet began. At the age of 17 he underwent an operation for the deformity. He

then wore iron supports for several years. He had considerable difficulty in getting about. Between the ages of 20 and 25 the hands became affected and claw-shaped. There was lateral curvature of the spine. Ulcers were present on both feet, and owing to extreme ulceration of the right foot it was amputated when he was 23. He died from pneumonia at the age of 32. He was unmarried. It is not known if he had any acute illness in childhood.

CASE V (Mother).

Mrs. S., the mother of patients I and IV, began to develop clubbed feet when about 17 years of age, and had an operation for the deformity when 18. She was married at 35, and in her family had only the two sons already referred to. That her hands were not affected is apparent from the fact that she was able to play the piano up to a short time before her death. She died at 50, from heart disease. No particulars can be obtained as to her having suffered from any illnesses in early life.

CASE VI (Grandfather).

Mr. A., the father of Mrs. S. and grandfather of the first patient, was known to have some deformity affecting both feet. No further information can be obtained, but it may be assumed that it was the same affection seen in the later generations.

As long ago as 1875 Leyden and others noted that there were cases of what might truly be called progressive muscular atrophy in which the wasting began, not in the intrinsic muscles of the hands, as in the Aran-Duchenne type, but in the lower limbs, and these cases, he remarked, were often hereditary. But it was not until 1886 that Howard Tooth¹ in this country, and Charcot and Marie² in France, identified these cases of hereditary lower-limb atrophy as a separate disease, subsequently known as the peroneal type of muscular atrophy, or by some as progressive neural muscular atrophy, after Hoffmann.¹⁸

The foregoing group of cases corresponds closely with this now well-recognized type, and in the following account the clinical picture represented in them is filled in with some detail.

The disease usually manifests itself between the early years of childhood and puberty, and the first sign is generally some weakness in the legs and feet, shown by a tendency for the "legs to drag" or the "feet to turn in." In most cases the affection has appeared to begin in the peroneal muscles, and it is certainly this which gives rise to the early

development of the characteristic foot deformity, the most usual form being talipes varus with some degree of pes cavus, a deformity which is clearly due to gradual loss of power in the peronei, and not to secondary contractures. In some cases, however, atrophy has been noticed in the intrinsic muscles of the feet from the beginning, and it is a question whether the point of onset is not more often than has been thought in the small muscles of the feet, a position in which the effects of the atrophy would not speedily become obvious. There is usually not only club-foot, but claw-toe as well.

The next muscles to be affected are usually the tibialis anticus, the extensor longus hallucis, and the extensor longus digitorum; and following these the flexor group, comprising the gastrocnemius, soleus, and tibialis posticus.

After the leg muscles those of the lower thigh are next attacked, especially the vastus internus, giving what is known as the bottle shape (with the neck downwards) to the thighs. The upper extremities are usually involved after a short interval, affecting first the intrinsic muscles, giving the claw-hand. This interval may vary within rather wide limits—Sainton gives six weeks and fifteen years. Thus in Case II the hands showed weakness only eighteen months after the first symptoms; in Case IV the hands became affected between four and five years after, while in Case I they were apparently free for twenty-four years, which is, as far as we are aware, the longest interval on record; and in the mother of this patient (Case V) they escaped altogether, as she was able to play the piano up to nearly 50 years of age, a short time before her death. As Sir William Gowers, in his well-known textbook, has remarked, claw-hand in early life is

Fig. 4.—Case II. Showing the tendency to drop-foot and pes cavus (February, 1904).

so rare from any other cause that it may be taken, together with club-feet, as almost pathognomonic of the condition.

The wasting may then proceed to the muscles of the forearm and upper arm; the supinator longus is spared, as in lead paralysis, and the muscles of the shoulder, neck, and back generally escape. They, however, may become affected and the wasting even general. The wasting is, moreover, as a rule more or less symmetrical, but it may begin earlier on one side, and that consequently show a greater degree of atrophy.

The electrical reactions may at this date be termed characteristic. In the Aran-Duchenne type there is a gradual diminution of irritability to the faradic current, and a degree of qualitative change in the galvanic reaction appears, the changes bearing a proportion to the degree of muscular atrophy. The fully developed "reaction of degeneration" may, however, be obtained, but when it does occur it is usually a late feature. In the "peroneal type" electrical changes take place early, before atrophy has become extreme, and the full reaction of degeneration appears early. Perhaps the most constant feature is either an early and greatly diminished irritability or complete loss of faradic excitability. In two of our cases, in which electrical examination was feasible, the diminished irritability to faradism was well marked, not only in atrophied muscles, but also in those not obviously atrophied, and there was, in the absence of analgesia, a marked diminution in what may be called faradic sensibility. Of course, it must be borne in mind that atrophy may be present in a muscle before it is apparent—in fact, the gross atrophy which is obvious can only be the outward expression of an antecedent atrophy which has been steadily progressing until sufficiently advanced to be observed.

In Case 1 it may be remarked where there was a diminution in electrical excitability of the apparently unwasted muscles, equivalent on a rough estimate to one-seventh of the normal—that is, a loss of six-sevenths—and including the facial muscles in this diminution, there was a distinct defect of tonicity in the muscles of the face, as manifested by an expressionless physiognomy, which might fairly be taken, we think, as evidence of a defect in the nutrition of the face muscles. In one of the cases recorded by Dubreuilh the face muscles were apparently incapable of expressing emotion.

Oppenheim,⁸ it may be mentioned, refers to a case in which this loss to faradic excitability was found in almost every muscle of the body accessible to electrical stimulation, although there was obvious wasting in the lower limbs alone. The galvanometer observations in Case 1 show that the diminution in the reaction to galvanism could not be due to excessive skin resistance, and, by using the patient's body as part of the faradic circuit in stimulating the observer's muscles, it was demonstrated that there was no excessive skin or tissue resistance to the faradic current. Both Hoffmann and Dubreuilh⁴ record cases in which the facial muscles could not be thrown into activity by either current when applied either through the nerve trunk or directly to the muscles.

Lateral curvature of the spine may appear early (as in Friedreich's ataxia) possibly in relation with the early development of club-foot. The affected limbs are often cold and livid, or pigmented. Cutaneous sensibility in some cases remains unimpaired, but, as a rule, there are patches of anaesthesia, especially in the region of the atrophied muscles and over the soles of the feet. Pains and cramps are often complained of. Fibrillary tremors are frequently present, especially in the hands.

Among the 52 cases collected by Sainton,⁵ the knee-jerks were absent or much diminished in 50 per cent, increased in 10 per cent., and normal in 40 per cent. As a rule, there is no sphincter trouble.

Some cases approximate very closely to "locomotor ataxy," so much so that one writer regards the "peroneal type" as a hybrid between "progressive muscular atrophy" and "locomotor ataxy." In Case 1 not only were the knee-jerks absent, but there were, in addition, patchy anaesthesia, the Argyll Robertson pupil, myosis in one eye, some inco-ordination of both upper and lower extremities, perforating ulcers on both soles, and at intervals there were general shooting pains in the limbs, only comparable with the lightning pains of locomotor ataxy.

As just suggested, this case of peroneal atrophy is by no

means unique in its manifestation of what may be called tabetic signs; in one point alone we had thought Cases 1 and 4 were unique—namely, in the exhibition of perforating ulcer; but Oppenheim⁸ refers to a case in which also there was perforating ulcer. Among other cases on record showing tabetic symptoms may be mentioned those by Siemerling⁶ (Argyll Robertson pupils), myosis (Dubreuilh⁴), lancinating pains and optic atrophy (Vizoli⁷), disturbance of micturition (Sainton,⁵ Ormerod,⁹ Egger⁹). It may again be repeated that in Case 1, after careful examination and consideration, the occurrence of syphilis was confidently excluded.

It is of interest, in this relation, to remember that though localized muscular atrophy in tabes is unusual it is not extremely rare. One of us,¹⁰ in publishing in *Brain* the notes of two cases with atrophy of the deltoid, biceps and supinator longus, collected from the case records of the National Hospital, Queen Square, among 200 consecutive cases of tabes, 29 with paralysis of muscles of the limbs or of muscles other than the external muscles of the eyes and of the laryngeal muscles, and 16 cases of obvious localized atrophy of the skeletal muscles. Among many similar cases in the literature not a few are stated to have had clubbed foot (Condoleon and others)—cases presenting affinities with Friedreich's ataxia, as indeed do some of the recorded cases of peroneal atrophy.

The etiological factors must now be considered.

Sex.—Males are more liable than females, suffering, according to Sainton, five times as frequently. In the family group on which this paper is based the transmission is very clearly from the great-grandfather to his daughter, from the grandmother to her sons, and from the father to his daughter. So here, at any rate, an ordinary transmission of nervous and mental diseases of the parent of one sex to the children of the other seems to hold good.

Age.—The disease would appear to occur most commonly between the ages of 2 and 15 years; it seldom begins after 20 and does not appear to have been observed before 2 years of age. Of 52 cases, for example, collected by Sainton⁵ the onset in 40 was before the age of 22, the extremes were 2 and 40.

Predisposing Causes.—Hereditary influences are very powerful. The malady is, indeed, essentially a familial and hereditary one. Isolated cases, of course, must occur, but they are either extremely rare or often unrecognized. According to Sainton there are only 10 isolated cases on record, but this is probably a too low estimate. The number of persons attacked in one family may be large—for example, Herringham¹¹ records 26 in one group.

Exciting Causes.—In a large number of the cases on record there has been a previous history of some acute specific infection, especially measles. In Ormerod's cases of a father, son and daughter, all had had measles in early life. In our cases there is a definite history of measles in one, and of whooping-cough in another. On the other hand, a son who suffered from both measles and scarlet fever has remained free, and one daughter who has escaped as far as can be ascertained all acute infectious diseases has been attacked. Cases are on record developing after small-pox and typhoid, and an influence of intoxication by lead, syphilis, and alcohol, is said to have been apparent in some cases.

PATHOLOGY.

As long ago as 1855 Virchow¹² found anterior horn changes in what must be identified with the peroneal cases, and as a result they were classified with progressive muscular atrophy. The first systematic examination of their morbid anatomy was made as late as 1889, three years after the publication of the clinical accounts by Tooth, Charcot, and Marie. Finding an atrophic neuritis of the peripheral nerves, the condition became classed in the multiple neuritis group. In 1893 Bernhardt¹³ found, in addition to the neuritic changes, sclerosis in the posterior columns of the cord. At the present time, especially since Sainton and Marinesco,¹⁴ first examined the nervous system in these cases by modern methods, the pathological evidence supports the view that the anatomical basis of the disease is on the type of a system lesion, especially of the reflex-arc system, and might be formulated as comparable with those of a hereditary locomotor ataxia (Friedreich) with muscular atrophy due

to a superadded cornual lesion. The actual findings in the recorded necropsies agree in degeneration of the nerves, the muscles, and the posterior columns of the cord; these three may be said to be constant as far as present knowledge goes. Among other lesions not constantly present or recorded are those of the posterior horns (recorded by Gombault and Mallet,¹⁵ and Marinesco), the anterior horns (by Friedreich, Gombault, Déjerine and Marinesco, and Sainton), sclerosis of the pyramidal tracts (by Sainton and Dubreuilh), and atrophy of the cells of Clarke's column by Sainton; while Déjerine,¹⁶ Gombault, and Marinesco have described some hypertrophy of the interstitial substance of the nerves leading to manifest thickening.

DIFFERENTIAL DIAGNOSIS.

The differential diagnosis offers little, if any, difficulty. Oppenheim and Cassirer¹⁷ have recorded a case which showed all or most of the distribution characters of a myopathy; but the reaction of degeneration (a degree of which is present often in the muscles not obviously wasted), the pains and anaesthesia if present and the distribution of the wasting are sufficient to differentiate the peroneal from the myopathic type. The similarity with progressive muscular atrophy is not close enough to occasion perplexity; the absence of the knee-jerks and the sensory phenomena are sufficient to obviate that. There is much greater difficulty in eliminating the possibility of multiple neuritis. Some of its characteristic symptoms developing after a toxic fever are suggestive, but the progressive and prolonged course of the peroneal atrophy are definitely distinctive, and the occurrence of electrical changes before and in excess of the atrophy are characteristic of the peroneal type alone.

In respect of treatment there is unfortunately but little to be hoped for. In the very early stages the administration of thyroid gland substance seems worthy of trial, on the theory that it might counteract the hereditary vulnerability of the central nervous system. Carefully adapted tenotomies and orthopaedic methods may certainly add many years of active existence to the patient's life, and the significance of this is especially important in view of the protracted course of the disease. For example, a case is referred to by Sainton who at 81 could still walk about although the disease began in adolescence.

REFERENCES.

- ¹ Tooth, H. H.: The Peroneal Type of Progressive Muscular Atrophy, Camb. Univ. Thesis, 1886; also St. Bart. Hosp. Reports, vol. xxv, and *Brain*, vol. x.
- ² Charcot and Marie: *Revue de médecine*, 1886, p. 97.
- ³ Oppenheim, H.: *Lehrbuch der Nervenkrankheiten*, Vierte Auflage, Erste Band, S. 266.
- ⁴ Dubreuilh, W.: *Revue de médecine*, 1890, p. 441.
- ⁵ Sainton, P.: *Nouvelle iconographie de la Salpêtrière*, Tome xii, 1899, p. 206 and p. 317.
- ⁶ Siemering: *Arch. f. Psychiat.*, Bd. xxxi, S. 105.
- ⁷ Vizoli: *Bulletin de la R. Acad. Med. Ch.*, Napoli, 1897.
- ⁸ Ormerod, J. A.: *Brain*, vol. vii.
- ⁹ Egger: *Arch. f. Psychiat.*, Bd. xxix, Hft. 2, S. 400.
- ¹⁰ Whiting, A. J.: *Brain*, Part 84, 1898, p. 494.
- ¹¹ Herringham, W. P.: *Brain*, vol. ix.
- ¹² Virchow, R.: *Virchow's Archiv*, Bd. viii, 1855, S. 537.
- ¹³ Bernhardt: *Virchow's Archiv*, Bd. cxxxiii, 1893, S. 287.
- ¹⁴ Marinesco, G.: *Archives de médecine expérimentale*, Tome vi, 1894, p. 320.
- ¹⁵ Gombault and Mallet: *Archives de médecine expérimentale*, Tome i, 1889, p. 385.
- ¹⁶ Déjerine et Soltas: *Progrès méd.*, 1893, No. 12.
- ¹⁷ Cassirer and Oppenheim: *Neurol. Centralbl.*, 1896, p. 718.
- ¹⁸ Hoffmann: *Deutsche Zeitschr. f. Nervenheilk.*, Bd. i, S. 95; and *Archiv f. Psychologie und Nervenkrankheit.*, Bd. xx, S. 560.

CASE OF CHRONIC INFANTILE PARALYSIS OF TEN YEARS' STANDING: TREATMENT: RESULT.

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CASES of chronic infantile palsy may be roughly divided into three classes: (1) Those of great severity, in which there is much wasting, extensive deformity, and, it may be, complete loss of the power of locomotion. Here it is obvious that surgery offers the only hope of betterment. (2) Cases in which the wasting is slight, and deformity absent; but where there is pain and aching after using the affected limb. Such are liable to be confounded with morbus coxae, but once their true nature is recognized, it is evident that no operation can be of benefit. (3) A group midway between these extremes—a lower limb, for example, is shortened, more or less wasted, and to some extent misshapen.

Cases falling into this third category are of fairly frequent occurrence in general practice. The diagnosis presents no difficulties, and the lesion is not so severe as to negative home treatment. Minor surgical measures may seem to be indicated, but in many instances the parents will not consent to anything of the kind. The question arises, what can be done?

The following case, occurring in my own practice, I venture to hope may prove not without interest:

N. H., a girl, aged 14, had infantile paralysis of ten years' standing, affecting chiefly the left lower extremity.

History.

At the age of 4 the patient, after exposure to a hot sun, was seized with an acute attack of vomiting and diarrhoea, with febrile symptoms. This lasted two days. On the child recovering from the gastro-intestinal disturbance the left leg was found to be almost completely paralysed. The left arm and back muscles of the same side were also slightly affected. There was nocturnal incontinence of urine, with which the child had not previously been troubled. The paralysis followed the usual course in such cases. That is to say, there was gradual spontaneous recovery up to a certain point, and after a few months the patient was able to get about again. The limb, however, remained generally feeble. So far as I could gather, there was not at this time complete paralysis of any group of muscles, but the movements of dorsiflexion and eversion of the foot were relatively the most impaired.

The parents, who were reasonably well-to-do, took the child to more than one neurologist, and received in every case practically the same opinion—that the disease was infantile paralysis; that the growth of the limb would be more or less checked; and that nothing could be done except to massage the affected muscles. Electrical treatment apparently was not mentioned; at any rate, it was not impressed on the parents as a vital necessity, nor does it seem that any warning was given as to the likelihood of deformities supervening unless suitable supporting apparatus were employed.

In accordance with medical advice the limb was rubbed systematically by the child's mother, and an attempt was made to carry out graduated exercises. Some improvement resulted from these measures, but in the course of a few months it was evident that the limit of improvement had been reached and the parents made up their minds that nothing further could be done. As the years went by, relative shortening increased, contractures of the calf muscles occurred, and from a state of paresis the peronei passed gradually into a condition of almost complete paralysis.

Physical Examination.

When I saw the patient, ten years after the original lesion, the left lower limb was much shrunken and wasted; there was $3\frac{1}{2}$ in. of actual shortening, as measured from the anterior superior spine to the inner malleolus. The buttock muscles were flattened and the gluteal fold lost. The foot could not be bent to a smaller angle than about 120 degrees with the leg. There was complete absence of voluntary power in the peronei, but no contractures of their opponents; the foot assumed the varus position on an attempt being made to place a weight on it. The limb was clammy to the touch and of lower temperature than its fellow.

The boot worn had no stiffening of any sort, and but 2 in. of sole extension.

Walking was possible only by the aid of a stick (Fig. 3), and the patient was liable to severe falls. As a consequence of the uncorrected shortening plus the paralytic varus, there existed marked downward (compensatory) tilting of the pelvis to the affected side, together with lateral spinal curvature and general faulty carriage.

Electrical Reactions.—Response to the faradic current was fair throughout the limb, except in the case of the muscles before specified. Even here, however, although all voluntary power was lost, a faint reaction indicated some remaining vitality.

Treatment.

Tenotomy of the tendo Achillis, together with shortening of the overstretched peronei was an obvious preliminary measure;

Fig. 1.—Illustrates shortening and talipes equinus. The patient was supported by a person not visible in the photograph.

Fig. 2.—Paralysed leg, showing foot drop and equino-varus.

Fig. 3.—Showing patient standing in original boot, steadying herself by the aid of a stick.