MUSCULAR ATROPHY, AFTER MEASLES, IN THREE MEMBERS OF A FAMILY.

BY J. A. ORMEROD, M.D.

THE two children whose cases form the principal part of this paper were under observation, as out-patients at the National Hospital for the Paralysed and Epileptic, from February to September 1883. In August 1884, I heard from their mother that they were still in the same condition. They had been, at an earlier stage, under the care of Dr. Barlow, who had noted the paralysis as peculiar, and who has kindly seen them for me again.

CASE I.—Annie W., *æt.* 17, was brought by her mother to me on January 30, 1883. Her legs were crippled, though she was able to walk; both hands were partially paralysed. The history given was this: she had measles between 6 and 7 years of age; about a month after she recovered from them, one foot (the right?) began to “turn,” the toes, according to the mother, pointing downwards and inwards. Tenotomy was performed at the Orthopædic Hospital. After this the other foot began to turn in the same way. About seven years ago (*i.e.* three or four years after the original attack of measles) tenotomy was again performed at University College. This affection of the legs did not come on suddenly, but took about a month to develop.

More recently the hands have become weak. This began five years ago. The mother first noticed that the girl used her hands awkwardly and stuck her knuckles out. The hands have got gradually worse, especially the thumbs, during the last two months.

She is subject to headache; also to shortness of breath and wheezing at the chest. Her menses are regular and natural, but preceded by much pain across the back and lower abdo-

men. With these exceptions her general health is perfectly good.

Her condition while under observation was as follows. Her legs were much wasted below the knees; the muscles of the thighs were not so well nourished as they should be. In cold weather there was much disturbance of the cutaneous blood-supply in the legs; sometimes the calves were blue and cold, with bright red patches on them like chilblains going to break; sometimes, in addition to the coldness and congestion, the whole leg below the knee was so swollen as quite to hide the wasting of the muscles. The tendon-reactions were present at both knees. The induction current produced no action when applied to the muscles below the knees, to the peroneal nerves—to the vasti—to the muscles at the back of the thighs (the last doubtful). The constant current also produced no action when applied to the peroneal nerves, or (so far as could be made out) to the muscles below the knees.

As to the condition of the hands and arms, for which she principally sought advice—there was well-marked wasting of the balls of both thumbs and of the intrinsic muscles of the hands generally; and wasting, though less apparent, of the muscles on the forearms, both extensor and flexor aspect. The fingers were in a position of semi-flexion at their two distal joints; their metacarpo-phalangeal joints being almost straight. All voluntary movements at the elbow- and shoulder-joints were perfectly retained; but extension of the wrist and fingers was possible, on the right side, only to a very slight degree; on the left side, not at all. Flexion of the wrists and fingers was possible, but to a slight degree only, on both sides. The left hand was said to be the worst.

Electrical examination, on two or three occasions, (it should be mentioned that she could not bear very strong currents) gave the following results:

(a) The induction current produced no action in the muscles of the thumb-ball or intrinsic muscles of the hands, nor in the muscles of the forearms; on the other hand, the muscles of the upper arms, and the deltoids, rhomboids, &c., acted normally.

(β) Reaction to constant current.

In muscles of thumb-balls present but diminished¹ [on L. side AOZ=KSZ: on R. side ASZ=KSZ]: later, no reaction could be obtained.

In extensor muscles of forearms: on L. side, absent; on R. side diminished, without qualitative change: later, no reaction on either side.

In flexors: on L. side absent; on R. side, diminished without qualitative change. But the flexor longus pollicis and pronator radii teres seemed to act normally.

In ulnar nerve at elbow, and median nerve at wrist, no reaction.

To summarize; the affected muscles showed absence of farado-contractility, and absence or diminution of galvano-contractility, with qualitative change in one instance only.

From Jan. 30th to May she took codliver-oil and syrup of phosphate of iron; from May to September, hypophosphite of soda. Meanwhile she had the constant current to the upper limbs. Complaint was made in July of incontinence of urine; not at night, but while she was walking about. This had been noticed once before; but it did not appear to trouble her much. In the condition of her limbs there was no change; and her attendances became so irregular that she had to be discharged.

CASE II.—Alfred W., æt. 9, brother of last patient, was first brought on Feb. 20, 1884, the history being that four years ago, after measles, weakness of both legs set in. This came on gradually, in about three months' time. But during the last few weeks, his mother had thought that the hands were getting weak.

The muscles of the lower limbs were much wasted below the knees, above they were apparently healthy; the patellar tendon reactions were exaggerated on both sides. The right thumb-ball was flattened as compared with the left; the induction current acted on *neither* thumb-ball. He appeared slightly feverish, and after this visit was laid up at home with bronchitis, so that I did not see him again till April. He had then

¹ I.e. required a considerably stronger current than the corresponding muscles in my own person. The estimation was made with a galvanometer, to avoid fallacies due to inaction of cells and variations of cutaneous resistance.

got thinner, and his mother thought that his hands were worse.

The following notes were taken between April and September.

There is wasting of both lower limbs, worst on the right side; but only below the knees. There is no vaso-motor disturbance, as in his sister's case. He wears orthopædic boots; when he walks without them, the right foot turns in. He can flex and extend the knee-joints freely; the patellar tendon-reactions are lively on both sides. As to the upper limbs, there is now evident wasting of the muscles of *both* thumb-balls, but still most marked on the right side. He cannot move the right thumb so freely as the left. The fingers tend to assume a position of flexion at their two distal joints, and extension at their metacarpo-phalangeal joints. This is particularly noticeable while he is using the hands. It is exactly the pose¹ which the mother noticed in an early stage of his sister's disease. He can, however, flex and extend the fingers at will. The forearms and upper arms are unaffected; the tendon reactions at wrists and elbows are lively.

Systematic electrical examination was impossible, on account of the child's nervousness, but the following facts were made out.

(α) A fairly strong induction current (which acted readily on my own thumb-muscles) had no action on his thumb-muscles, nor on the first dorsal interossei (the latter doubtful), but acted on the muscles of the forearms, both extensor and flexor aspect. It did not act on the muscles below the knees, either anterior or exterior group, nor yet on the vasti, though these latter did not appear wasted.

(β) A moderate constant current acted on the muscles of the right² forearm, extensor aspect, KSZ > ASZ; but had no action on the thumb balls.

The constant current did not act on the muscles of the legs, but acted on some at least of the thigh muscles.

The electrical conditions then, so far as they could be made

¹ Due to paresis of the interossei.

² The left forearm, and the other muscles of the right, were not examined, but may perhaps, from their normal appearance and faradic reactions, be presumed normal also.

out, were the same as in his sister's case; viz. absence or much diminution of contractility, in the affected muscles, to *both* currents.

He was treated with faradisation to his hands and forearms, and with the same medicines as his sister. He did not improve: indeed the peculiar pose of his hands became, if anything, more marked.

In neither of these cases was any complaint made of sensory disturbance. There were no fibrillary twitchings, and no rigidity. The affected muscles were, as I have said, wasted or wasting; and I did not notice enlargement of any other muscles; though I admit that I did not specially look for it. Being told by the mother that the father had been paralysed in one leg from infancy, I visited him at his cottage, and made the following notes.

CASE III.—William W., æt. 56, cab-cleaner, healthy in appearance, though grizzled and looking older than his years. Ever since he can remember, his right leg has been in its present condition. He was told by an elder sister¹ that he had measles very badly when 12 months old, and that the right foot began to turn when he was about three years old. His family seem to have connected the affection of the leg with the measles.

There is a decided difference in girth between the two lower limbs, especially below the knee, as follows:

Largest girth of left calf,	12·5 inches	.. of right,	9·5 inches,
" " " thigh,	16	" .. " "	15 "

In length, no difference between the two. Patellar tendon reactions well marked on both sides, but the left greater than the right. The muscles below the right knee are extremely feeble; the foot hangs quite limp at the ankle, he cannot point the toes at all; he says he always walks on the heel; and he can only evert the foot slightly.

He cannot extend the right knee-joint with the same facility as the left.

¹ This was told him before the birth of his two children, now paralysed. I mention this to show, that the sequence of events in his case was not made to suit theirs.

The left leg is perfectly natural, the arms and hands are natural, and he is positive that he never had anything the matter with them. I had no opportunity of making an electrical examination.

Evidently his condition may have been due to an old attack of infantile paralysis, but I do not feel certain that it was so. The history, such as it was, pointed to a gradual onset; and the bones had grown to the same length as those of the other limb. In former years a doctor has taken much interest in his condition, had him photographed, &c.

Of five other children (besides my two patients), one was smothered in bed, four are alive and healthy. Two of these are grown up. All had measles at one time or another. The mother was one of a remarkably large family of children, and she knows of no nervous complaint among them. The father (Case III.) was one of four, all healthy; of these a sister committed suicide at 15, but he is confident she was provoked to it, and was not insane. His father, mother, and paternal grandfather, were all healthy.

The points that deserve notice are

- (1.) The connection with measles.
- (2.) The youth of the patients.
- (3.) The fact that brother, sister, and father were paralysed.
- (4.) The possible nature of the disease.

The measles were not apparently severe in the children's case. No doctor was called in; but from the description the mother gave me of the coryza and the rash, and from the fact that measles was known to be in the neighbourhood, I have little doubt that this disease was, as she said, measles. The two children who became paralysed did not have them at the same time; one of the others had them at the same time as Alfred, but did not become paralysed; neither did any of the neighbours' children, so far as was known to the mother. There was therefore nothing in the nature of the epidemic specially tending to cause paralysis. On the other hand, considering the uncommon nature of the paralysis, it seems unlikely that the connection with measles should have been in the case of both children (and in that of their father?) a mere chance sequence. The exciting cause was probably the

measles; the predisposing cause, a family proclivity to this form of paralysis.

This brings us to the other points, which it will be convenient to consider together. What was the nature of this paralysis? The three clinical types which suggest themselves are: (1) acute anterior poliomyelitis or ordinary infantile paralysis; (2) chronic¹ anterior poliomyelitis or *paralysie spinale antérieure subaigue* (Duchenne); (3) progressive muscular atrophy.² We may dismiss the first of these, with which the onset and course of the disease entirely disagree. As to the second, chronic anterior poliomyelitis, the youth of the two patients might have been held, till recently, to justify us in dismissing this also. But cases recently published³ by Professor Erb and Dr. Hughes Bennett show that chronic anterior poliomyelitis does occur in children, however rarely. Indeed, between Dr. Bennett's first case and my own there are some close similarities. Dr. Bennett's patient suffered from whooping-cough and *measles*; a month afterwards, slight weakness and unsteadiness of the gait was noticed; still later, awkwardness of the hands; there was cyanosis of the legs, and diminished response both to faradism and galvanism. There remains, however, this difference, that in my patients there has been no attempt at recovery, though the disease is now of five and ten years' standing; whereas both Erb and Bennett emphasise the fact that their patients recovered, as being confirmatory of the diagnosis.

Moreover if it is rare for chronic anterior poliomyelitis to occur in children, it is, so far as I am aware, unknown that it should run in families. This is not the case with progressive muscular atrophy; which, as is well known, frequently attacks many members of a family. These also are just the instances in which the disease begins in early life. For recent information on the subject of "hereditary" muscular atrophy, I may refer the reader to an article by Zimmerlin, recently abstracted by Dr. Beevor.⁴ As regards the progressive muscular atrophy

¹ Better perhaps called subacute, to avoid confusion with (3).

² This last probably comprehends several forms.

³ 'BRAIN,' Nos. 21 & 23, "On Chronic Atrophic Spinal Paralysis in Children."

⁴ Zimmerlin, 'Zeitschrift für Klin. Med.,' Band I., Heft I. 'BRAIN,' No. 26. p. 285.

of youth, Erb recognises this as a distinct type and assigns to it the following characteristics.¹ It begins insidiously in childhood or youth, usually in the shoulder or arm muscles, sometimes in the legs or back. The pectorals trapezii latissimi rhomboids and long muscles of the back are at a certain stage almost always involved; biceps, brachialis, and supinator longus very frequently; and the triceps later. Among other muscles, the deltoids and *small muscles of the hands are always spared*, and the muscles of the forearms for a long time. In the lower extremity, the glutæi, quadriceps, peroneals and tibialis anticus are usually attacked. The districts of the bulbar and cerebral nerves are not invaded.² While some muscles are soft and flabby, others may remain firm. There are no fibrillary twitchings. The irritability of the muscle to mechanical stimuli disappears. The patellar tendon reflexes are preserved. Distinct reaction of degeneration has never been demonstrated to exist.³ The course of the disease is very chronic. This form of disease, Erb suggests, when it attacks several members of one family, constitutes the "hereditary" or "family" form of progressive muscular atrophy; and when it is complicated with formation of fat in lieu of the atrophied muscle, constitutes pseudo-hypertrophic muscular paralysis, which is also, as is well known, a "family" disease. He thinks that the disease begins in the muscles themselves, not in the cord or nerves.⁴

Now as regards the present cases, the age of the patients, the family predisposition, the absence of distinct reaction of degeneration,⁵ the preservation of the tendon reactions—all suggest that they should be classed as instances of this

¹ The original article will be found in 'Deutsch. Archiv für Klin. Med.,' Band 34, s. 467. I quote from an abstract by Strümpell, 'Neurol. g. Centralblatt,' 1884, p. 200.

² Duchenne states that progressive muscular atrophy in children begins in the face. E. Remak ('Neurol. g. Centralblatt,' 1884, p. 337) gives a case of Erb's juvenile type, with complete facial diplegia.

³ There is a simple diminution of contractility to both currents ('Neurol. g. Centralblatt,' 1883, p. 453).

⁴ Recent pathological observations by Landouzy and Dejerine confirm this. See 'Lancet,' Jan. 26, 1884.

⁵ Qualitative change was found once only the galvano-contractility was diminished or absent; see above.

juvenile form of progressive muscular atrophy. But the order of invasion, at least as regards the upper limbs (first muscles of hands, then of forearms), certainly does not agree with this view.

I am compelled therefore to leave the exact nature of the disease undecided; perhaps the further course of the cases may decide it; but as it seems unlikely that I shall have further opportunities of investigating them, I venture to record them as they at present stand.