THREE CASES
(PROGRESSIVE MUSCULAR ATROPHY
AND INFANTILE PARALYSIS)

ILLUSTRATING THE
LOCALISATION OF MOTOR CENTRES IN THE BRACHIAL
ENLARGEMENT OF THE SPINAL CORD.

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THE two cases which I have shown this evening are chiefly important with regard to the localisation of the centres for different groups of muscles of the upper limbs, in the different segments of the brachial enlargement of the spinal cord.

The first case is one of progressive muscular atrophy. The patient is 38 years old, and has been a goldbeater for eighteen years; he attributes his illness to violent muscular exertion, but he has been a lamplighter for the last six years and much exposed to wet and cold. There is no family or previous history of importance. Three years ago he began to have difficulty in extending the

third and fourth fingers of the right hand ; he then thinks that the muscles of the right arm began to waste, and he gradually lost power in them.

A year after the onset, the left arm became affected, and he had trouble in supinating the forearm, then in flexing the elbow, and in abducting the arm away from the trunk.

For the last six months the legs have felt weak.

Present condition.—In the *right* arm the muscles most affected are, the extensors of the wrist and fingers and thumb, the small muscles of the hand, the interossei, the flexors of the fingers, the biceps, supinator longus, deltoid, rhomboid, serratus magnus, teres major and minor.

The interossei, especially the first, and the muscles of the hand are very much wasted and the extensors of the fingers are more atrophied than the flexors. The fingers are rigidly flexed ; he cannot extend or separate, but can just flex them. In the right forearm supination is more difficult than pronation.

The biceps, brachialis anticus and supinator longus are much wasted, and though he has no power to flex the elbow-joint he can just make the two first muscles contract. On the other hand, the triceps is so strong and well developed that he can prevent the elbow-joint being forcibly flexed. The deltoid is very much wasted, specially the middle part, and he is unable to abduct the humerus away from the trunk, although the supraspinatus seems to contract. The pectoralis major acts fairly well, but the sternal better than the clavicular half, and he can with much difficulty flex the shoulder-joint, bringing the arms forwards, though not to the horizontal line. In so doing the scapula projects backwards and away from the trunk, owing to the serratus magnus being much wasted and incapable of advancing the inferior angle of the scapula. The absence of action of the serratus magnus is well seen when the arm is forcibly raised above the horizontal line, and allowed to drop, when it falls at once on removing the

support. In adducting the humerus to the trunk the latissimus dorsi acts fairly well, but not the teres major and minor. He cannot draw the scapula towards the spine by the rhomboid muscle. The upper end of the trapezius acts normally, the rest of the muscle not so well as in health, and in shrugging the shoulders the scapulæ appear to be elevated by the trapezii and not by the levatores anguli scapulæ.

In the *left* arm the changes have not proceeded so far, and there are some important differences. Here the muscles most affected are, biceps, brachialis anticus, supinator longus, deltoid, rhomboid, supra- and infra-spinati, teres major and minor, serratus magnus, extensors of thumb, and to a less degree, the extensors of the fingers. There is not so much wasting of the hand and forearm as on the right. He can flex the fingers and thumb well, and use the small muscles of the ball of the thumb, being able to touch all the tips of his fingers with the thumbs. He can extend the first finger well, but the wrist and other fingers he can only just extend, and the thumb not at all. When the hand is supported the interossei can separate the fingers. The biceps and supinator longus are much wasted and absolutely no voluntary movement can be produced; the inaction of the supinator longus is strikingly seen, in comparison with the extensors of the fingers which he can still throw into action. I may remark that this is just the *opposite* to *lead* paralysis, where the extensors are paralysed and the supinator longus, which is really a flexor of the elbow-joint, escapes; this is important, as *lead* paralysis is considered by some writers to be due to disease of the cells of the anterior cornua of the cord. On the other hand, the triceps is strong, normal, and well formed. The deltoid, the supra- and infra-spinati are much atrophied and their movements *nil*. The pectoralis major is rather weak in its action and the clavicular more affected than the sternal part, so that he is unable to bring the humerus forward so well as on the right side. In adducting the humerus to the trunk, the

latissimus dorsi acts well and better than on the right, but no action can be seen in the teres minor and major. The serratus magnus does not act at all, but there is not that protrusion of the posterior border of the scapulæ which is so marked on the other side. This is probably due to the upper fibres of the pectoralis major not being strong enough to advance the arm, so that no strain is put upon the upper part of the scapula. The rhomboid is quite powerless to draw the scapula to the spine.

The patient carries his head bent forwards and there is much prominence of the upper dorsal and last cervical spines. He has some difficulty in extending the neck and throwing the head backwards, and then when it is allowed to come forwards again it falls suddenly after it has passed the vertical position ; this is due to the weak action of the muscles keeping the head erect—the splenii, complexi, and trachelo-mastoid muscles.

The sterno-mastoids, however, act normally and can turn the head to the right or left.

The pupils are equal, but the right does not act to light, though it does to accommodation.

The left pupil reacts to light and to accommodation.

With regard to the electric reactions of the affected muscles I may state that there is no reaction to the induced faradic current on the *right* side in the interossei, extensors and flexors of fingers and thumb, the deltoid, the rhomboid, the serratus magnus, the spinator longus, and the teres major ; of these, the extensors of the fingers, their flexors and the rhomboid only react to a strong constant current and to the positive better than the negative pole ; the biceps does react to the faradic current, but in this muscle slight power still exists.

The lower and middle parts of the trapezius react to a rather strong faradic current. In the *left* side the biceps, supinator longus, deltoid, rhomboid, supra-spinatus, infra-spinatus, teres major and minor, and serratus magnus do not react to strong faradic currents ; the biceps and supinator longus react to the positive pole of a strong

constant current, as do also the teres major and minor. The pectoralis major reacts to a moderately strong faradic current, which is also required to produce contraction in the middle and lower parts of the trapezius.

The legs, though weak, do not present any marked wasting or loss of power. The patellar tendon reflexes are increased, a condition which I have seen in other cases of muscular atrophy, and especially in one which I published in 'Brain,' (Part xix) where ankle clonus was also produced. This is probably due to sclerosis of the lateral columns secondary to the primary lesion.

The second case which I have brought forward is that of a young man, aged 18, who had infantile paralysis when one year old, following scarlet fever; that is, in less time than two or three months after the fever. He was paralysed in all the limbs, but he recovered more or less with the exception of the left arm, which remained most affected. He states that, as long as he can remember, his right arm has been strong but the left arm weak; he was able to walk from two years old, but the feet have always been more or less contracted.

Present condition.—The patient is affected more or less in all his limbs, the left arm being the most disabled. *The right upper limb* below the elbow is normal and quite strong, the muscles of the hand and forearm being very well developed and possessing considerable power.

The triceps, including the long head, is very much wasted and he has not the least power to contract it; he cannot keep the elbow extended nor can he extend the elbow-joint when the forearm is allowed to hang at right angles to the upper arm. The triceps gives no reaction to electric current, either faradic or constant.

The biceps and brachialis anticus and the deltoid are normally developed and act very vigorously. The pectoralis major is very well represented in its upper clavicular part, whilst the lower or sternal part is absolutely gone, not a trace of it being seen, so that the anterior fold of the axilla passes in a line towards the upper end

of the sternum, instead of to the lower. The pectoralis minor cannot be made out. He has very little power in pressing with the right hand on a table when the arms are extended, a movement which is specially due to the action of the lower part of the pectoralis major.

The serratus magnus is not affected, and he can raise the arm high up in the air with considerable strength. The remaining muscles of the scapula are normal, including the infra- and supra- spinati, the teres minor, the subscapularis and teres major, the rhomboid, and the trapezius. The latissimus dorsi is absent and cannot be made to act; when the patient tries to adduct the humerus to the trunk against resistance, the work is done by the muscles of the scapula, which is thereby drawn forwards away from the trunk.

These muscles take the place of the latissimus dorsi in placing the arm behind the back. There is no reaction in the latissimus to faradic or constant electric currents.

In rotating outwards the humerus by the teres minor and infra-spinatus, the scapula is drawn forwards, the rhomboid not having sufficient power to keep it fixed.

The *left arm* is more affected than the right, and is generally wasted, the circumference of the right forearm being eight and a half inches, whilst the left is only seven and a quarter.

The muscles of the ball of the thumb and the interossei are not wasted, and he has good movement in them. The extensor aspect of the forearm is wasted, and he can only just extend the fingers and wrist to the horizontal line; in so doing the wrist becomes flexed, while the fingers are hyper-extended. He has no power to extend the thumb. These extensors require strong faradic current for their contraction. In the upper arm, which is considerably wasted, the biceps and supinator longus act fairly well, but not so strongly as on the right side. The triceps is very much wasted and no contraction can be produced either by the will or by strong faradic or constant currents, with the exception of its long head, which

reacts to a strong faradic and to the positive pole of a strong constant current. The deltoid is also a good deal wasted, but the patient is just able to abduct the arm to the horizontal position.

The serratus magnus is somewhat affected, so that he cannot raise the arm further than the horizontal line, unless the elbow be flexed, thereby reducing the weight of the arm, but this inability may be also due to the weak action of the deltoid. In adducting the humerus to the trunk, no action is seen of the latissimus dorsi or the teres major or minor. What action there is appears to be done by the posterior part of the deltoid, which also has slight power in rotating the hanging humerus outwards. The patient cannot effect any rotation inwards, the special rotators, both in and out, being very much wasted and quite powerless.

The pectoralis major is much wasted, especially the sternal part, of which a thin band only is visible, so that he is unable to keep the left hand touching the right shoulder. The supra- and infra-spinati are very much wasted and powerless. The rhomboid is incapable of drawing back the scapula to the spine, and although the upper fibres of the trapezius act normally the rest of the muscle acts feebly.

In the lower limbs the feet are much altered in shape, the toes being hyper-extended whilst the arch of the foot is very much increased, the length of the feet being thus much shortened. This alteration seems to be due chiefly to the non-action of the interossei, and is similar to the claw shape assumed by the hand when the interossei are affected. The tendo Achillis does not appear to be contracted.

The patient can flex the toes of both feet, and slightly flex upwards the right ankle, but not the left. He cannot evert either foot by means of the peronei, and can only just invert both ankles by the tibialis anticus, the right better than the left. The electric examination shows no reaction in the peronei of the left leg to strong

faradisation and only slight contraction to a strong constant current. The tibialis anticus of the right leg reacts very slightly to a strong faradic current, whilst that in the left reacts only to a strong constant current.

There is no patellar tendon reflex in either leg, which is in marked contrast to the condition in the other case.

With regard to the electric reactions in this case, there is no reaction to a strong faradic or constant current in the *right arm*, of the triceps, pectoralis minor, or latissimus dorsi; the clavicular part of the pectoralis major reacts normally to faradisation, while the sternal half, being absent, cannot of course be tested; the serratus magnus requires a faradic current rather stronger than normal to make it contract.

In the *left arm* there is no reaction to strong currents in the triceps (with the exception of the long head which contracts to a strong faradic, and to the positive pole of a strong constant current), the latissimus dorsi, the supra- and the infra-spinatus, the teres major and minor, or the rhomboid. The clavicular part of the pectoralis major reacts normally, but the sternal part only to a strong faradic current. The extensors of the fingers, the supinator longus, biceps and serratus magnus react to strong faradic currents, as does the deltoid, but the anterior and posterior parts of this muscle react rather better than the middle part; this is of some importance as the deltoid may be looked upon as composed of three muscles, the anterior part of which acts in conjunction with the clavicular part of the pectoralis major, which in this case acts fairly well.

The two cases which have been brought forward may be both considered together, for although one is a case of progressive muscular atrophy and the other is one of infantile paralysis, they are both due to a similar cause, viz. a lesion affecting the cells of the anterior cornua of the spinal cord, and especially of the brachial enlargement. With regard to progressive muscular atrophy it has been considered by Friedreich that the disease lies primarily

in the muscles themselves, but at present most writers consider that the cells of the anterior cornua are the seat of disease.

My object in bringing forward these cases is to prove clinically the correctness of the experiments of Professors Ferrier and Yeo on monkeys. These observers divided the several motor roots of the brachial and lumbar plexuses and on stimulating the peripheral ends various groups and combinations of muscles were put into action. I have drawn up a list of these muscles taken from Ferrier's paper in the 'Proceedings of the Royal Society,' 1881, and also from a paper of his in 'Brain,' Parts 14 and 15; and parallel with this list I have given lists of the muscles affected in my two cases. The condition of the two arms is given separately in each case, so that it can be seen at a glance how each is affected.

There has always been a difficulty in explaining why in diseases like progressive muscular atrophy and infantile paralysis certain muscles should be picked out and others left. The groups of muscles affected do not correspond to the nerve supply of any one peripheral nerve; and the extensors as a whole do not suffer more than the flexors, although they are generally the weaker muscles; for while the extensors of the wrist and fingers are usually the first to suffer, the triceps has always been remarkable for its immunity from attack, and has frequently been noticed as the last to be affected.

On looking at these two cases, I think it will be observed that broadly the one is the counterpart of the other; I do not mean absolutely so, for it is not easy to find hard-and-fast stereotyped limits to clinical cases. Comparing the muscles affected in each case, it will be seen that in that of progressive muscular atrophy they correspond to the muscles assigned by Ferrier and Yeo to the upper and lower ends of the brachial enlargement, whilst the case of infantile paralysis corresponds to the muscles governed by the middle of that enlargement. Although the experiments of Ferrier and Yeo were made on the motor nerves,

the groups of muscles assigned to the different roots correspond also to the anterior cornual cells supplying them.

On looking at the list of muscles affected in the case of progressive muscular atrophy it will be seen that there are some differences between the two arms.

In the right arm the muscles innervated by the fourth and fifth cervical, according to Ferrier and Yeo, are all affected, with the exception in the fourth cervical of the supra- and infra-spinati muscles which have escaped, so that, excluding these two muscles and the diaphragm—which has the nuclear origin of the phrenic in the medulla and not in the cervical cord—not a single muscle has escaped. On looking at the sixth cervical group, the pronators of the wrist are slightly affected, but the serratus magnus is the only muscle seriously damaged, and this is also supplied by the affected zone of the fifth cervical. In the seventh cervical, all the muscles have escaped except the flexors of the wrist and fingers and the teres major; and it is interesting to note that the muscles which are supplied by, and only by, the sixth and seventh cervical have escaped; this is in striking contrast to the case of infantile paralysis, where the same muscles are the only ones which are paralysed. The eighth cervical zone has all its muscles affected excepting the long head of the triceps and the pectoralis major, about which Ferrier has some doubt as to its proper position here. The first dorsal, representing the hand muscles, is entirely damaged.

When we examine the left arm we find that the disease, while following the general course of the other arm, has shifted its position a little higher up, so that we see, contrary to what is usual, that the small muscles of the hand supplied by the first dorsal have escaped, and the disease does not seriously begin till we reach the fifth cervical zone.

The pectoralis major is affected, but in the upper or clavicular part more than the lower or sternal half—in contradistinction to what is found in the other patient—

and I certainly think that the pectoralis major may have its two halves assigned to different parts of the cord, the clavicular half going along with the anterior fibres of the deltoid, of which it is a continuation, whilst the sternal half is more associated with the triceps group. This grouping is also illustrated by physiological action, for in pressing the two hands together the clavicular part of the pectoralis major, the anterior fibres of the deltoid and the biceps are associated, whilst in pressing the hands downwards, against a table, the sternal part of the pectoralis major and the triceps act together, the biceps not being used in this movement, which may be illustrated by pressing on the table to assist in rising from a chair. In the fourth and fifth cervical groups all the muscles are involved, as on the opposite side, but with the exception that the supra- and infra-spinati are also affected, whilst the extensors of the fingers have more power than on the right side.

In the list of the muscles affected in the infantile paralysis case, it will be seen that only the triceps, pectoralis major (sternal half), and latissimus dorsi are absolutely powerless and give no reaction to electric stimulation. Now these muscles, according to Ferrier's list, are innervated *only* by the sixth and seventh cervical roots, and if these are seriously damaged, or rather if the anterior cornua, the segments of the cord containing their nerve-cells, are diseased they become powerless. It is interesting to note that these three muscles are the only ones in the sixth and seventh cervical zones which are supplied by both these nerves; the teres major and subscapularis, both internal rotators, are supplied by the seventh cervical alone, and the pronators by the sixth alone, and these have escaped. I can only suggest that these may perhaps have another nerve supply from a different zone which has not yet been discovered. I think some light may be thrown on this apparent anomaly when we examine the left arm.

In the left arm precisely the same muscles are affected as in the right, but the disease has gone further and involved other zones of the cord. And here we see that

the subscapularis and the teres major, the clavicular portion of the pectoralis major, the rhomboid, the supra- and infra-spinati must be added to the list of muscles severely affected. I think it is interesting to note that the remaining muscles supplied by the sixth and seventh cervical are here invaded, as the disease is more extensive. The pectoralis major is here more affected in the sternal than its clavicular part, following a course similar to the muscle of the right side. The fact of the deltoid being somewhat affected would lead us to place the clavicular part of the pectoralis major along with the deltoid, whilst the sternal part would be assigned to the sixth and seventh dorsal zones.

I confess it is difficult to make the spinati harmonise with the fourth cervical zone.

Besides the above muscles the serratus magnus, deltoid, and extensors of the wrist are somewhat affected, though they possess voluntary movement, and all respond to strong faradisation. They would show a moderate extension of the disease to the fourth and fifth cervical.

It will be seen that the eighth cervical and first dorsal are almost intact, the small muscles of the hand and flexors of the fingers thereby escaping.

It seems probable that in the case of progressive muscular atrophy the disease began with atrophy of the right hand muscles, and so corresponded to the first dorsal segment of the spinal cord; the disease subsequently attacked the upper end of the cord, the fourth and fifth cervical, the middle part of the brachial enlargement not being affected or only very slightly.

It is very extraordinary why the extremities of this enlargement should be most attacked, and why the disease should, so to say, leap over the middle part; perhaps some difference in the blood-supply of the middle and ends of this part of the cord may in future time be discovered. It is of course possible that the disease may begin in the two extremities at the same time, and I think that in the left arm the disease seems to have commenced at the

upper part of the brachial enlargement as shown by the biceps and deltoid being first affected.

In the case of infantile paralysis, the whole of the brachial enlargement was probably entirely affected at first, the ends recovered completely in the right arm, but to a less degree in the left, while the middle has been left permanently paralysed, but why we do not at present know.

A third case which was under the care of Dr. Ferrier at the Queen Square Hospital—whom I have to thank for allowing me to publish it here—is a boy, aged 13, who had infantile paralysis when two and a half years old, affecting the right arm and the legs.

At present the right arm is affected in the triceps group of muscles corresponding to those supplied by the sixth and seventh cervical roots, and therefore corresponds to the second of the cases already described, viz. that of infantile paralysis.

In the right arm he has all the movements of the small muscles of the hand and interossei, and can extend and flex the wrist and fingers, and these muscles react normally to the faradic current. The biceps, brachialis anticus and supinator longus act well but the triceps is very much weaker than in the left arm, and it is important to note that whereas the long head of the triceps (which is placed in the eighth cervical group) reacts to nearly a normal faradic current, the short head reacts very slightly to a strong faradic current and to the positive pole of a rather strong constant current, whereas all parts of the left triceps react to the normal faradic and constant currents. The right deltoid abducts the humerus well, but the sternal half of the pectoralis major is decidedly weaker than the clavicular part, and while this clavicular part acts normally to faradisation, the sternal part requires a much stronger current than does the same part of the muscle on the left side.

The serratus magnus is very weak, and in carrying the right arm forwards to the horizontal line, the posterior

border is seen to protrude very much owing to the weak action of the serratus magnus ; the protrusion is most marked just before the horizontal line is reached, but after this is passed and the arm is raised nearly vertical, the protrusion disappears ; this may possibly be due to the lower fibres of the serratus being less affected than the upper fibres (?) ; this muscle reacts very little, if at all, to a strong faradic current. The infra-spinatus and teres minor, the external rotators of the shoulder, act better than the internal rotators, the subscapularis and teres major.

In adducting the right humerus to the trunk against resistance, the latissimus dorsi hardly appears to contract at all, and the scapula is drawn away from the trunk by the teres muscles ; the right latissimus acts very slightly to a strong faradic current. The patient has apparently some difficulty in drawing the right scapula towards the spine, but the rhomboids act to nearly a normal faradic current.

In this case it will be seen that the muscles affected belong to the groups innervated by the sixth and seventh cervical roots, the fourth, fifth, and eighth cervical and the first dorsal escaping. In the seventh cervical we have the teres major, latissimus dorsi, subscapularis, pectoralis major (sternal half), and triceps, in fact all the muscles, except the flexor of wrist and fingers, which being also supplied by the eighth cervical escapes. In the sixth cervical, the latissimus dorsi, the pectoralis major, serratus magnus and triceps are affected. It may be said that the serratus magnus ought to have escaped as it is also innervated by the fifth cervical, but it seems probable that this muscle is twofold in its action, and the upper fibres are supplied by the sixth cervical, while the lower fibres inserted into the lower end of the scapula are supplied by the fifth cervical, and this grouping is borne out by the movements produced by Ferrier and Yeo in monkeys.

FERRIER AND YEO'S LIST.	Progressive muscular atrophy.		Infantile paralysis.	
	Right.	Left.	Right.	Left.
<i>1st Dorsal.</i>				
Small muscles of hand and inter-ossei	x			
<i>8th Cervical.</i>				
Long flexors	x			
Ulnar flexors of wrist	x			
Small muscles of hand	x			
Extensors of wrists and fingers	x	—		—
Triceps, long head		—	x	—
(Pectoralis major ?).			Sternal part	
<i>7th Cervical.</i>				
Teres major	x	x		x
Latissimus dorsi			x	x
Subscapularis				x
Pectoralis major		—	Sternal part	x
			x	
Flexors of wrist and fingers (median)	x			
Triceps			x	x
<i>6th Cervical.</i>				
Latissimus dorsi			x	x
Pectoralis major		—	Sternal part	x
			x	
Serratus magnus	x	x		—
Pronators (flexor of wrist ?)	—			
Triceps			x	x
<i>5th Cervical.</i>				
Deltoid (clavicular part)	x	x		—
Biceps	x	x		
Brachialis anticus	x	x		
Serratus magnus	x	x		—
Supinator longus	x	x		
Extensors of wrist and fingers	x	—		—
<i>4th Cervical.</i>				
Deltoid	x	x		—
Rhomboid	x	x		x
Supra-spinatus		x		x
Infra-spinatus		x		x
(Teres minor)	x	x		x
Biceps	x	x		
Brachialis anticus	x	x		
Supinator longus	x	x		
Extensors of wrist and fingers	x	—		—
Diaphragm				

x Severely affected. — Slightly affected.

Ferrier has since found that the above list is one root too high ('Proc. Roy. Soc.,' vol. 35, 1883, p. 229), and the groups of muscles should refer from the

fifth cervical to the second dorsal, so that the group assigned to the fourth cervical really belongs to the fifth, and the fifth to the sixth, &c.

(For a report of the discussion on this paper, see 'Proceedings of the Royal Medical and Chirurgical Society,' New Series, vol. i, p. 430.)