Further studies on the muscular factor in infantile paralysis / William Mackenzie.

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FURTHER STUDIES ON THE MUSCULAR FACTOR IN INFANTILE PARALYSIS.

WILLIAM MACKENZIE, M.D., F.R.S. Edin.

Lecturer on Applied Anatomy, Melbourne University.

THE STABILITY OF MUSCLE IN THE ANIMAL ECONOMY.

Since John Hunter delivered his celebrated Croonian Lectures on "Muscular Motion," in which he drew attention to the consideration of every muscle in an animal body as a simple independent power, scant attention seems to have been paid to this subject. On the contrary, we find much energy devoted to the study of bone, whose existence, as shown, not only from the laws of evolution, but by numerous instances in the comparative world, is dependent on muscle. To that apathy is due the total disregard hitherto shown to the muscle in infantile paralysis. Since Hunter wrote, perhaps the most important work is that of Harrison, who by removal of the spinal cord in a series of frog embryos finally determined that muscular tissue is already pre-determined in the early gastrula, or even in the egg itself, and not dependently as was thought on stimuli from the nervous system. The stability of muscle cells in the animal economy is familar to all histologists, and we know of course that protoplasmic contraction and relaxation occurred before bone or nerve tissue came into being, and in a low variety of Mollusc, the central nervous system has beem completely removed without interference with muscular action; and further under narcosis, loss of muscular movement is preceded by loss of consciousness. The view usually held, we take it, is that muscles remaining hopelessly paralysed are so owing to destruction of the corresponding cornual cells; for if the cells had recovered, in the absence of mecahanicl interference the muscles should commence to "functionise." And similarly the coldness of the limb, its shortness and atrophy, are simply indications of loss of trophic function of the cells. The inherent stability of the muscle cell has never been regarded as worthy of consideration in this disease, nor the line of recovery as being along the avenue of muscular action and irrespective possibly of the consideration of great destruction of central cells. Furthermore, also, the muscle in this disease, apart altogether from sequential

degeneration has never been looked on as possibly pathological, but always regarded as normal.

The Bacteriological Position.

Now, in spite of the bacteriological results on monkeys—a species distinct from man, of lower intellectuality, and without the assumption of the erect attitude—which bacteriological findings we must remember, were inaugurated by the inoculation, into the central nervous system through a trephine opening, of an emulsion of the spinal cord from a fatal case, there is another picture obtained by observation on the living human species at variance with the generally conceived pathology of this condition, and with which the bacteriologists working on this disease and hoping to evolve a curative serum seem unacquainted. The whole question of cure—and in this respect infantile paralysis is unique—resolves itself into one, not merely of subsidence of cord affection, but of the restoration of lost muscular function—for we must remember that in this manner is the disease manifested. The effects of a serum could scarcely be expected to evolve lost muscular action; and although occasionally as a prophylactic it might be applicable in a hospital, with numerous cases around, it would be impossible to foresee in every instance that this is the disease for which the child is sickening, apart from the known difficulty, even with loss of power present, of differentiating this disease, *e.g.*, from an infant with epiphysitis. Without doubt the disability arising from infantile paralysis in the human subject has been greatly exaggerated, and I look on it as extremely rare for a muscle to be regarded as "permanently paralysed," unless in some old-standing deltoid cases. If a case be seen early, and treated on the "zero" lines previously defined, that case should go on to recovery, though trouble may arise from the retrogressive muscles of the leg. Furthermore, as I have repeatedly demonstrated, a case of the upper extremity, looked on in the light of the old pathology as the most hopeless for recovery, seen early runs on to a rapid and complete restoration of function. To regard the muscles as normal, and to be kept so till the cell recovered, has been a disastrous policy for this extremity, as Jones and Tubby state.

Does the Disease follow Segmental or Evolutionary Lines?

Stress is frequently laid on the "segmental or associated" grouping in this paralysis—a fact quite at variance with my own experi-

ence, and concerning which Wright, of Manchester, writes, and truly, "Very often the muscles paralysed have no relation to one another, being picked out, as it were, at random"—*e.g.*, in a child seen recently with deltoid, supinator brevis, and adductor pollicis of the right upper, and quadriceps, and leg muscles of the left lower limbs. Sherrington's work shows us that, were that factor dominant, selective affinities would rather be extensor and corresponding flexor, and hence there should be little deformity, since opposites would be affected. Deformity, markedly noticeable sometimes in twenty-four hours, we know to be a characteristic of this disease. I think it will be conceded that this disease affects the upper limb much less frequently than the lower, and I have never yet met with an adult affection there, though I have this year seen nine recent dult cases with paralysed lower extremities. And this is as one would expect. The greater the stability, and the greater the intrinsic and extrinsic adjustments of a part, the less its proneness to infection. There is a greater relative instability of the lower limb as compared with the upper—a fact well known to comparative anatomists—and, conversely, a child, since it has less co-ordinative control, has a greater proneness to infection than an adult; and, furthermore, the older the child, the less frequently have I seen cases of upper limb paralysis. Again, as has been before stated, the greater the tendinous character of a muscle the less its power of recovery; and the less the utility of a muscle, or its importance in the animal economy, the greater its liability to infection. The disease follows rather evolutionary than "segmental" lines, and not as Flexner, following the old view, would have us believe—"That the degree of affection is determined by the richness of the arterial blood-supply, whence is explained the liability of the lumbar and cervical enlargements to severe lesions"—which on logical grounds, is an evasion of the question, since it might easily be used to explain the converse. Pelvic tilting, persisting buttockal, spinal, psoas, or abdominal paralysis should never be allowed to occur. The affected muscles are actively functionising muscular masses, and if rested, as advised, on a double Thomas' splint to secure immobility, will all go on to an uninterrupted recovery—in marked contrast to those where the limb has been massaged early, and allowed to hang.

Paralysis of the Deltoid, Quadriceps, and certain Leg Muscles.

The muscles, in my opinion, that give most trouble in recovery are the deltoid, quadriceps, and the peronei and tibials of the leg,

and all these more especially on the left than on the right side. The deltoid is essentially muscular, and not tendinous, and is actively functionising, what accounts for its delay? Taken early, and subjected to scientific rest—as on the writer's upper limb splint—it invariably recovers; but if not seen within a month from the onset, the chances become more remote. The evolutionary development of function in this muscle I have already explained in detail, and will refer to later. This year I have seen two cases of upper limb paralyses of three months' standing, in girls æt. $2\frac{1}{2}$ and $2\frac{3}{4}$ years respectively, and no sign of movement till the limb was supported, after which both were able later to raise the limb above the head sitting up. What was the pathology of the central cells in these cases? Did the cells recover, and was that recovery insufficient for muscle recovery? The disastrous results of old unrested deltoid cases I regard as suggestive evidence of a pathological muscle. As regards the peronei and tibials, we are dealing with muscles whose function is becoming less important with the disappearance of pronation and supination of the foot, and altered size of the fibula.*

As regards the quadriceps, the prognosis even in old cases is always reassuring. At the outset there is always some rest, whereas in the case of the deltoid, if unsupported, there is always the dragging force of the limb, sleeping or awake. This is the muscle responsible for so much crutchdom, and that significance I pointed out some years ago. Taken early, every quadriceps paralysis should go on to an uninterrupted recovery, and even in old cases after ten years, and in one after eighteen years, I have secured perfect recovery. The more the limb has been allowed to hang, or the knee bent the less the chance of securing what I refer to later as the erect function, though in all cases the quadriceps may be educated to extend the knee from acute flexion. Under the old pathology, it was regarded as the most hopeless muscle for recovery—a view quite opposed to my own clinical facts, and which can be easily verified on any old case of contracted knee.

Effect of Stimulation of Nerve to Affected Muscle in a Recent Case.

After waiting several years, an opportunity was recently afforded me of treating a male adult, æt. 25, in the Melbourne Hospital, thirty-six hours after the onset of the paralysis, with affection of the

* Mackenzie, Observations on the Comparative Anatomy of the Fibula, Proceedings Royal Society of Victoria, Vol. XXIII.

left lower extremity. Even in that short time marked foot-drop was noted. On this patient we were enabled to carry out an interesting experiment, for the details of which I am indebted to Alex. Thwaites, M.Sc., of the Physiological Laboratory. An incision was made over the extensor group of muscles some 8 cm. long. The deep fascia was incised and mostly removed. The tibialis anticus had been previously decided on for the experiment since it was obviously paralysed, and the foot-drop rather favoured its relaxation, and hence there was no mechanical opposition to its movement. In spite of controls, a direct stimulation to the muscle, or through the nerve to the muscle direct, failed to elicit any definite contraction, a result which should not, according to Sherrington, even with complete segmental destruction, have occurred for at least six days, and suggestive of a block at the junction of nerve and muscle in that little known region designated the receptive substance. In this case we were enabled to obtain in six weeks a recovery in the toe and hip muscles, and also the quadriceps to extension from full flexion, but not the ability to raise the heel when lying on the back, which he is being gradually educated to do. He left the institution walking well on a light caliper knee splint. He is now, eleven weeks from the onset, able to raise the heel with knee extended. This result is in marked contrast to two other adult cases in the hospital treated under old principles of massage and electricity, one of which has been an inmate for some fourteen months.

The Evolution of Muscular Function in the Quadriceps and Deltoid.

If the knee be bent, lying down, the function of the quadriceps is to straighten it, and further with the normal quadriceps we are enabled to lift the heel off the ground with the knee extended. When we stand, the quadriceps maintains that stiffening of the knee necessary for the erect attitude. This further power of the quadriceps has been a later acquirement in man's evolution. With one quadriceps intact, the erect position may be maintained. Loss of this function, and hence solution of continuity at the knee, is, as the writer pointed out some years ago, responsible for the large amount of crutchdom following this disease seen in any large city, and is obviated by the conversion of the limb into one rigid piece, *e.g.*, by a Thomas' caliper splint. The method of restoration of muscle function in the quadriceps I have previously dealt with.

Curiously enough, we may have patients able to flex and extend the knee powerfully in bed, but who are unable to maintain the erect position without support. This function of the quadriceps acquired latest disappears first, and is the last and most difficult to acquire in the education of the affected muscle. And similarly in the case of the deltoid, in which we may have perfect movement lying down, but with the adoption of the erect position the limb hangs powerless to the side.

It is inconceivable to imagine what effects either drugs or serum would have in the evolution of this function, yet no quadriceps but can be educated back, and no deltoid seen early but will recover, and occasionally even the function of old deltoid paralysis may be restored. We pay our price for the erect position.

Division of Deep Fascia over Muscles.

Apart from the quadriceps, it is common enough to see other muscles—and I refer particularly to the muscles of the leg and forearm—condemned both on electrical and utilitarian grounds as hopeless, yet the muscles may ultimately be brought into action. The cells presumably are gone—often for years—yet why should the muscle be ultimately coaxed to work. Yet that it can is the rule, not the exception. In my first series, five instances of paralysis, affecting the extensors of the foot, without movement of the toes for two years, were seen, in which an incision over the muscle origin revealed in all abundance of muscle tissue; and in two cases of division, and three of removal of the deep fascia, room was given to the muscles to expand, the fascia being simply contracted down on the affected muscle, and allowing, owing to its unyielding character, no room for enlargement. Recently I have carried out this procedure in many cases, and regard it as the possible solution of the delayed recoveries of the leg muscles, and have performed it early in the left lower limb. No leg muscles should be condemned as hopeless till this procedure has been carried out, and the good condition of "condemned" muscle, and the results of recovery of action, are surprising. It should always accompany the straightening of an acquired talipes equino-varus. For old forearm condition, with apparently little distinguishable, except skin and radius and ulna, it is the only procedure, and after the application of warm saline, it is a surprise to find that we demonstrate the individual muscles, and subsequently bring them into action along zero lines.

A Unique Case of Recovery of Leg Paralysis.

Some twelve months ago I was asked to see a boy, aged 11½ years, with paralysis of the right leg. This was thin and cold, and no movement of the toes, either flexor or extensor, could be detected. The foot, owing to gravity effect, was in the position of equinus. He certainly seemed a fit subject for one of the numerous surgical procedures advised for flail ankle. Treatment by various methods had been carried out for nine years. The foot was placed at right angles, in an arc shoe, to overcome the gravity effect. With the heel supported, the boy was encouraged many times daily to endeavour to flex or extend the toes. Though at first without result, movement gradually returned in the toes, and in six months a definite tendo Achillis had developed. The flexor toe action was more powerful than the extensor, and although the drop had lessened, there was still little power over flexion of the ankle, in spite of twelve months' relaxation. Last week I exposed the extensors, and removed the deep fascia — the procedure being followed by immediate improvement, so that in six days he was not only able to powerfully flex and extend the toes, but flex the ankle almost fully as well, and now needs only muscular action to secure a strong foot.

The Question of Prophylactic Treatment.

In this direction, we would expect a serum to be most efficacious, especially when the disease occurred in epidemic form. Nevertheless, although one can never speak positively about prevention, I feel almost assured that in at least five cases affection of limbs has been aborted by placing them at physiological rest, *e.g.*, commencing loss of function in two limbs, with a third paralysed. This raises the question of adequate rest of limbs in children with intestinal affection during the summer months, and, like a possibl serum, could be utilised in epidemic cases, when suspicion would be early entertained, and at least the severity of the affection be greatly mitigated.

The Zero Position.

In previous communications, I have referred to the term "zero" position in explaining principles of treatment, drawing attention particularly to the action of the quadriceps muscle. A muscle is rested only when it is placed in a position where its work is zero, and its origin and insertion are as nearly as possible on a level, since

the resumption of function, which should be gradual, is always easiest along a straight line. The writer's treatment depends on the recognition of the zero position, in connection with which confusion is apt to occur. Though anatomically we speak of a muscle as being flexor or extensor, we really refer to the maximum function of a muscle; and the question arises as to the recognition of the minimum of muscular function—for it is only by the recognition of that minimum that the muscle may ultimately be coaxed up to the maximum—the ideal. The zero position of a muscle corresponds to that position in which the muscle is incapable of performing further work. If, on the other hand, the knee be contracted owing to a paralysed quadriceps, it is obvious that —the latter being at a disadvantage for functionising — equilibrium has been destroyed. Hence, we place the quadriceps in a position of relaxation—not necessarily complete—prevent the overaction of its opponent when resting by means of a splint, or when exercising by position, and daily work it to restore the lost equilibrium, by recognising that it is incorrect to flex the knee beyond the point at which extension is possible, *i.e.*, at which effort becomes manifest—thus recognising the cessation of its opposition to its opponent.

A paralysed muscle is not "dead," nor is it unless placed in the zero position, at rest. Since rest is the basic treatment of inflammation, only by the zero position can the door be closed on all sources of irritation, and cord, nerve, receptive substance, and muscle, be at physiological rest.

This disease, it is important to remember, destroys muscular adjustments, and, *ab initio*, one is not merely dealing with paralysed muscles, but with what is really the abnormal action of non-affected ones.

Recovery in Cases of Contracted Knees of Long Standing.

Early last year the writer recorded a case of recovery of the quadriceps quite unique in the annals of this disease. It was a complete recovery from an acutely contracted knee of some ten years, owing to paralysis of this muscle. This year he has treated four similar cases—two of ten years, one of two, and one of eighteen years' standing—all now able to walk, and all with the prospect of having soon a perfectly-functionising quadriceps, *i.e.*, one able to maintain the erect position unsupported, since all can extend from acute flexion and raise the heel when on the side. These cases

are quoted because they represent the worst that a paralysed quadriceps could suffer. After fruitless attempts at treatment many years ago, they were regarded as incurable—the affected anterior cells being presumably destroyed, the muscle as irrecoverable, and no better than a useless dangling limb, amputation was to be preferred. For nearly ten years those cells, even if they had recovered, were never used. Hence there were, according to the recognised pathology, two factors at work—firstly, all involvement; and secondly atrophy from disuse. Relaxation for many months does not cause recovery; yet, in one month, a scientific working of the muscle causes a regeneration or re-education of affected cells, or else there were adjacent and adaptable healthy cells called on to functionise. What position does the presumed degenerated nerve to the muscle occupy in these cases, presuming the origin to be wholly central? Pertinently, the question arises—Were the cells alone affected; was the muscle also the seat of disease? It suggests that, though the central factor is presumably always favourable (for I have yet to see the quadriceps that won't respond), the muscular factor is the dominant one in recovery.

Results of Nerve Transplantation.

If cell recovery were the all-essential in this disease, as we have been taught, then surely nerve transplantation should have here obtained its greatest successes—the conditions being particularly applicable for its use, viz., healthy muscle which only required stimuli to enable it to resume function. Nevertheless, except in occasional cases, this method seems to have been little used, and presumably has been found inapplicable—neither such authorities as Vulpius or Jones recording cases. Apart from the risk of interference with the function of non-affected muscles, mere regeneration of the nerve is *per se* insufficient for recovery, because by the time such regeneration is complete, the muscle itself, apart altogether from possible pathological change, has been functionally reduced by the circumstances attendant upon its lengthy paralysis, and must be treated on zero principles along minimum lines to the maximum. Till that be done no transplantation should be attempted, for in no other way can the amount of cell recovery be determined, and the necessity for its use be justified. This question, from the point of view of nerve injury, I hope to refer to on a future occasion.

Present Position Defined.

Flexner and Levaditi, looking at this disease from the bacteriologist's standpoint, hope, we take it, that by the discovery of a

micro-organism a serum will be produced, which will abort the disease when threatening, or modify it if present, and since this is a disease of paralysis, by that means presumably to evolve muscular function when lost. It will be generally agreed that in the case of an epidemic the same cause is at work, be it toxin or micro-organism. In sporadic cases, if due to an organism, varied ones may possibly produce an attack, of which a case seen recently by me is suggestive. A baby, 11 months old, previously healthy, which developed the disease after two and a half weeks diarrhœa, in a district where no other cases were noted. Speaking practically, whatever we might hope for, the disease as such will manifest itself to us by the loss of muscle function, and it must be extremely rare to see a case with developing paralysis. In fact, my own experience is that cases are rarely seen within five days of the onset, and all clinicians are aware that early diagnosis in infants is an extremely difficult thing. It is inconceivable to think of a serum being effectual in restoring muscular function when lost, and the followers of Owen Thomas know well how little tuberculin has done for cases say of tubercular hip, since it is adequate rest that cures the disease and prevents the deformity, without which any drug or vaccine would be ineffectual; and as before stated, cure in this disease is not merely restoring the function of paralysed muscles, but regulating the action of non-affected ones. My own view is that the disease is regarded with a seriousness to which it is not entitled, and that the central cell has been given a dominant position in treatment unjustifiably. Pelvic tilting, buttockal wasting, lordosis, and curvature of the spine, which go to make up the usually formidable and apparently hopeless picture, do not represent this disease, and are all preventable. The possibility of the affected muscle being pathological and not normal has never suggested itself, nor in spite of its position in man's evolution have the lines of its recovery of action been regarded as dominant. The old view that gave such disastrous results was that, if the cell recovered, the muscle would, and if the cell failed, the muscle could be regarded as hopelessly paralysed. Hence, in treatment the idea was to maintain the tone of the muscle by massage and electricity, but as for years, or perhaps centuries, case after case did not recover, the old view might surely have been regarded with suspicion, a suspicion that said perhaps the cell has recovered, but that does not necessarily denote recovery of muscular power of action. Whatever improvement Levaditi and Flexner may hope to produce in the

cell by means of a serum, will still be powerless to restore the function of a paralysed muscle—the desideratum.

In those cases of acutely contracted knee owing to paralysed quadriceps of a duration from 10 to 18 years, it was assumed the muscle did not act because the cells were destroyed. Under treatment that considered the muscle, in a comparatively short period function is completely restored to the quadriceps. Granting that the cells alone were affected and recovered again, I ask is recovery of cell sufficient for muscular function to be restored, as taught. For six months, relaxation, *i.e.*, absence of mechanical interference with action, failed to secure recovery of muscle function. Again, why should a deltoid be powerless for three months, and begin to functionise after a few days' rest on the zero principles, if the cell recovery were the all-important point in treatment. The cell must have recovered, or presumably the deltoid would not have worked, yet cell recovery did not cause the muscle, *sua sponte*, to functionise. It can be confidently stated that the prognosis can be regarded as hopeful, and the disease a recoverable one, provided the zero position of the muscle be recognised. Unlike the old view in which the amount of work a muscle was asked to do was usually in excess of cell and possibly muscular recovery, the resumption of work should be gradual, *i.e.*, presumably with cell and possibly muscular recovery. I recognise that the treatment of the muscle is the all-essential in this disease. Instead of waiting for a chance recovery in the anterior cornua, and treating in a perfunctory way the muscles with massage and electricity, we recognise that every affected muscle will work, provided that a commencement be made from zero. It is recognised that an affected limb will retain its heat and show little waste, provided that it be rested and worked within physiological limitations. Work must not be done *for* the muscle, it must itself work. And although we may think that the minimum of a recovering muscle is slight, *e.g.*, to coax the quadriceps to extend from a two-inch flexion, we must remember that it really represents the maximum function of the muscle for the time being, and as such may soon exhaust in the same way as the ordinary individual would soon tire if asked to continue flexion and extension of the forearm to the full extent. Hence, patience is required. Treating cases early, my experience has been that all go on to recovery, and even in old cases such recovery of muscle will be obtained that except perhaps in the case of the shoulder,

the procedures of arthrodesis, tenodesis, tendon transplantation, tendon shortening or excision, will all be rendered unnecessary. The doctrine of muscle rest and work on zero principles lay down for all cases a definite and scientific line of treatment, and to those who follow it in detail a new vision of this disease is opened up, to which serum and medical treatment, as in the case of a tubercular joint, must be subsidiary.

Finally, as regards the use of that lauded and condemned drug urotropine in this disease, it seems to me to be based on an empiricism. In the series of recoveries of upper limb cases I first reported, all were placed on hydrarg. internally, but I regard its utility as dubious. The luetic factor must always be remembered in severe and fatal cases as in two seen recently, one with all limbs affected and associated with a syphilitic epiphysitis, and another, a baby 12 months old, whose birth was preceded by 5 miscarriages—another child in the family being affected with a history of four previous miscarriages. I have also injected soamin in two cases that did well.

Is it too much to ask the clinical collaborators of Flexner and Levaditi to take cases of old condemned quadriceps with contracted knee, "whose cells have gone," and after the knee has been straightened, verify the means of recovery I have suggested; or the old withered leg or forearm after the deep fascia has been divided or partly removed; or the hanging upper limb after rest on the writer's splint; or, with the recent case of, say, two lower limbs, make a commencement with complete rest in a double Thomas' splint, and without massage or electricity find that little wasting occurs, and that by paying regard to the zero position, a complete recovery will ensue. Then they will recognise that the hopeless picture of this disease is not from paralysis but from inefficiency of treatment.

The line of treatment indicated evolves the recovery of function in paralysed muscles, irrespective of the use of drug or serum. The utility of such measures if introduced, will, I take it, be in the early stages, possibly to hasten cell recovery, but especially to prevent other cells being affected—and provided, of course, an early and correct diagnosis be made.

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