REMARKS

ON

SOME POINTS IN THE PATHOLOGY OF THE
PARALYSES WHICH OCCUR DURING
THE FIRST TWO YEARS
OF LIFE.¹

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It is hardly necessary for me to remark that the paralyses arising during the first two years of life form a very wide field for discussion, far too wide indeed to attempt to cover during the time allotted to the introducer of the subject. All that I can hope to do to-day is to discuss some of the points which are the most interesting and perhaps least best known in connection with the etiology and pathology of the lesions of the nervous system which occur during early life.

During the last few years much progress has been made in differentiating and classifying these palsies, and we no longer include under the name of "infantile paralysis," lesions which have little in common except that they occur during the first two or three years of life. But notwithstanding this progress towards a clearer knowledge, there is yet much to be learnt of their pathology, and more observations are required; especially do we require more *post-mortem* examinations made during the acute stages.

That there should be strongly marked lines of difference between the lesions encountered in the adult brain and in the infant is only what we should naturally expect. In the former, when middle life is well past, we find evidences of retrograde processes which tend to weaken or damage the walls of the arteries, and moreover we have cardiac and kidney lesions which tend to increase the tension of blood in the vessels and lead to cerebral hæmorrhage. The infant brain has the advantages which an immunity from degenerative changes consequent on wear and tear bring, and the disadvantages of immaturity, for the vessels of the pia mater are exceedingly delicate, and readily allow of oozing through their walls, while the brain substance is wanting in firmness and easily damaged. Moreover, damage to a brain as yet partially developed is likely to effect greater mischief in the long run by interfering with growth, than the same damage to an already well-developed brain. These differences are certain to influence the lesions which occur. In the infant we have meningeal hæmorrhage, meningeal inflammation both of brain and cord, and inflammation of the most vascular spots in the spinal cord; while we miss the large cerebral hæmorrhages, and the various forms of softening so commonly occurring in the brain of the adult towards the decline of life.

For the sake of clearness we may class the principal forms of paralysis occurring during the first two years of life under the following heads:

1. Intrauterine lesions (meningo-encephalitis).
2. Meningeal hæmorrhage.
3. Syphilitic arteritis and softening.
4. Acute cerebral paralysis (encephalitis and embolism, etc.).
5. Acute spinal paralysis (anterior poliomyelitis).
6. Peripheral paralysis.

1. *Intrauterine Lesions*.—In connection with these it is interesting to note what severe lesions may occur to the fœtus, and yet, in spite of this, it may continue to live and be born at term. Inflammation of the meninges of the brain or peritonitis may take place, and, instead of quickly proving fatal, as we should naturally expect, the fœtus survives, and a hydrocephalus secondary to the meningitis, or a sclerosis of the brain, or cicatrization and constriction of the bowels is the result. The semivenous condition of the foetal blood is not favourable to

acute inflammations; moreover, the fœtus is under much more favourable conditions for recovery from disease than in the infant after birth. The results of meningeal inflammation we see in the brains of idiotic children which show atrophy, sclerosis, or chronic hydrocephalus.

2. *Meningeal Hæmorrhage* occurs under a variety of circumstances during early life, but the invariable immediate cause is asphyxia. Let an infant pass into an asphyxiated condition, and an effusion of blood from the meningeal vessels is extremely likely to occur; the delicate vessels are readily ruptured or allow of oozing when they are distended with venous blood. The amount of bleeding varies; it may be only slight—perhaps only amounting to a few minute extravasations—or, on the other hand, the whole surface of the brain may be covered with blood. The most extensive meningeal hæmorrhages I have seen were in two newly-born infants, who had apparently been suffocated by throttling shortly after birth by forcibly compressing the larynx between the fingers and thumb—at least, the bruise marks on the neck suggested this. In both these cases there was a large extravasation of blood, both over the convex surface and also at the base of the brain. In another case in which there was extensive meningeal hæmorrhage complete birth had been delayed for more than an hour after the head was born, in consequence of the child's abdominal cavity being distended with fluid and preventing its passage through the pelvis. Doubtless the commonest cause of asphyxia—hæmorrhage—is a prolonged and difficult labour, during which the child becomes asphyxiated, the hæmorrhage taking place during the act of birth. That this is common enough I have had abundant opportunities of verifying *post mortem* in infants dying shortly after birth, where the mother has been attended by neighbours or ill-educated midwives. Meningeal hæmorrhage, as is well known, may take place during attacks of whooping-cough or during severe attacks of vomiting. There is no doubt also that it may occur during convulsions; the proof, however, is necessarily difficult, because at the *post-mortem* examination it may be impossible to say whether the meningeal hæmorrhage present is due to the convulsions or the latter are due to the irritation caused by the effusion of blood. In one case of mine, where the infant—10 weeks old—had been badly fed and suffered from convulsions, and who finally died in an attack, there were found *post mortem* minute hæmorrhages on the surface of the lungs; the brain was watery, the veins turgid, and several hæmorrhages had occurred beneath the pia mater, on the convex surface. Here there could be little doubt that the asphyxia produced by the respiratory spasm had given rise to the meningeal bleeding.

What is the effect of an extensive surface hæmorrhage on the surface of the brain if the infant continue to live? The bleeding, it must be remembered, is usually bilateral, and is most commonly found involving the parietal region, and running along the Sylvian fissures. The blood clot which forms separates the pia mater from the surface of the convolutions, tearing through the vessels which pass from the pia to the grey substance, and necessarily resulting in damage to the nerve centres. The result is certain to be that the nutrition of the nerve centres is interfered with; they fail to develop as they should, and more or less degeneration of the motor path as it passes through the brain and cord takes place.

In the large majority of cases in which "birth palsies" are present there is a history of a difficult labour, or, at any rate, the infant was born "blue," and there was more or less difficulty in bringing it round, the cyanosis remaining perhaps for some hours after birth. The causes of asphyxia are of course various; there can be little doubt, however, that there is an immediate connexion between asphyxia and hæmorrhage from the pia mater, which damages the parietal regions of the brain, and gives rise to the well known spastic paralysis (either paraplegic or hemiplegic) of infancy.

It is curious to note that in the vast majority of cases no symptoms pointing to a surface lesion are present at first; there is no paresis of the extremities, and frequently no convulsions; in fact, an extensive meningeal hæmorrhage may be present without any definite symptoms. In some cases facial and other paralyses have been noted, especially when extensive brain damage has been done or nerves have been compressed after leaving the brain. This is in harmony with what we know with regard to the undeveloped state of the cortical centres at birth; the infant's movements are almost entirely reflex, the lower grade centres are the only centres functionally active, and it is only as the child ought

¹ Read in the Section of Diseases of Children at the Annual Meeting of the British Medical Association, held in Leeds.

to be developing, learning to stand or walk, that the mother notices some backwardness in the use of its limbs, and some waywardness in its mental attainments. In some cases, at least as early as the third or fourth month, a rigidity may be noticed about the tendo Achillis, and an exaggerated tendon reflex.

Post-mortem examinations made on children who have suffered for years from "spastic paralysis," the result of a birth hæmorrhage, show atrophy of the parietal regions of the cortex, and indeed, in the worst cases, the shape of the head shows a deficiency in the parietal regions.

Syphilis.—Syphilitic disease of the brain cannot be said to be at all common in connection with hereditary syphilis occurring during the first few months of life. When it does occur it usually assumes the form of arteritis, with more or less softening of the brain and a consequent paralysis. A chronic meningitis with hydrocephalus may occur. The prominent symptoms in such cases consist in convulsions with marked stiffening of the limbs and retraction of the head, and, later, more or less rigidity and spastic condition of the extremities. As an instance of syphilitic brain disease in a young child I may relate the following: The infant was first seen at three months old, when suffering from coryza and a well-marked rash. A month later the epiphyses of the lower end of the tibia and fibula, and also the radius and ulna, were swollen and tender. When eight months old it began to suffer from convulsions, mostly left-sided at the commencement of the attack, but afterwards becoming more general. In the course of a few months the left arm and leg became drawn up and more or less rigid, the elbow was bent, the arm pronated, and the fingers flexed; later still, the right arm and leg were more or less affected in a similar way. This condition lasted till death, which took place when the child was fourteen months old. At the *post-mortem* examination arteritis, affecting the small arteries of the pia, was present, accompanied by superficial softening of the grey substance.

Acute Cerebral Paralysis, usually taking the form of hemiplegia, and depending upon various pathological conditions, is apt to come on suddenly during the first year or two of life. Considerable controversy has taken place with regard to the cause of this condition, but without doubt an infantile hemiplegia may be caused by a variety of lesions. Thus the softening accompanying tubercular meningitis, meningeal hæmorrhage, and embolism of the middle cerebral artery rank as causes.

In the majority of cases which occur during the first two years of life, there is some acute febrile disease present, usually also convulsions, and then suddenly, to the surprise of the attendants, the child is found to be hemiplegic. Such at least is the history we usually obtain, for careful observations of the primary illness are not numerous, and we have usually to depend upon the memory of the patients' friends when we want information on this point. The cause of this primary illness is often uncertain; it is attributed by friends not infrequently to "teething," to "brain fever;" in some cases measles or pneumonia, or both, are present; there is often hyperpyrexia; in a case which came under my notice, measles and pneumonia were present; the temperature was 107°; severe convulsions occurred, and hemiplegia followed in a few hours. How far in these cases the high fever is responsible for the mischief it is not possible at present to say. Then convulsions are almost certainly present, and are prolonged and severe; they are mostly unilateral, beginning in the side which is afterwards paralysed, but usually becoming general. Drowsiness, and at times coma, are present; these may be of short duration, or the child may lay in a semicomatose condition for weeks or more. It is a very significant fact that similar attacks occur, being accompanied by convulsions, fever, and coma; no paralysis follows, but the child shows a loss of intelligence or backwardness which dates from the period of the attack, or it may become idiotic. There seems to be little doubt that in these cases damage has been done to the brain, but the motor centres have escaped. Again, in other instances there is an attack of so-called "brain fever;" the child recovers, but from that date it is completely deaf. Some damage has evidently been sustained to the auditory centres, or a double labyrinthine inflammation has occurred. What is the lesion present in these cases? Observations made many years after the paralysis has occurred in the majority of cases clearly shows that more or less brain damage has been sustained to the cortex in the neighbourhood of the fissure of Rolando, and that, as a result, there has been a cicatrization or sclerosis at the seat of the old injury.

Has this been the result of an embolism of the middle cerebral,

a local encephalitis, thrombosis? If not, what has been the nature of the lesion? Embolism of the middle cerebral and a secondary softening has undoubtedly occurred in some cases, and in other cases possibly a thrombus has formed in the artery leading to a like result. This was the case in one of my own patients, a boy of a year old, who was suffering from cyanosis due to a contracted pulmonary artery. Embolism also occurs, especially in older children suffering from endocarditis, and in some of the recorded cases this has occurred secondary to endocarditis not infrequently present in scarlatinal nephritis.

On the other hand, the theory of embolism or obstruction of the middle cerebral does not explain some of the cases which have occurred, and in which *post-mortem* examinations have been obtained some years afterwards. In some of these the lesions have been confined to the convolutions bordering on the fissure of Rolando, and the lesion has extended no great depth into the brain substance. In another the superior parietal lobule only; in others the gyrus fornicatus or occipital lobe has been affected (Osler). If embolism had been the cause we must assume that some of the branches of the middle cerebral have been obstructed by a clot; and, while this is perfectly possible, it is improbable.

It has been suggested by Strümpell that an inflammation of the grey matter of the cortex, a poliomyelitis analogous to an anterior poliomyelitis takes place. This theory is certainly most suggestive, and it may reasonably be supposed that patches of inflammation may occur on the surface giving rise to coma, hyperpyrexia, convulsions, and paralysis. Hardly anything, however, is known of such a condition *post mortem*; and the only case that I know of which at all supports this theory is one recorded by Hensch. In this case a boy, aged 3 years, was suddenly seized with convulsions accompanied by fever, which ended fatally. At the *post-mortem* examination there were patches of meningitis on the parietal regions on both sides, and hæmorrhagic infiltrations were found in the grey substance. More evidence, however, is required before we can accept Strümpell's theory as explaining these cases of hemiplegia. Still I am by no means inclined to deny the possibility of a convex meningo-encephalitis occurring and giving rise to the symptoms described.

Can we regard convulsions as playing an important part in the production of these brain lesions? It is a remarkable fact that almost universally the paralytic attack is preceded by a series of prolonged and severe convulsions. It not infrequently happens that a child has an attack of convulsions, perhaps more marked on one side, from which it perfectly recovers; later on it is again seized with convulsions of the same side, followed by paralysis. It is well known that after severe and repeated epileptiform attacks a hemiparesis follows, affecting the side which has been most convulsed. The paresis lasts a variable time, from a few days to a few months. In some cases the paralysis is almost complete, but, as a rule, recovery quickly takes place. The cause of this temporary paresis is uncertain; possibly it may be due to an exhaustion of the nervous centres, or to some organic lesion, such as a punctiform hæmorrhage from ruptured capillaries. That convulsions, if severe, do give rise to meningeal hæmorrhage there is undoubted *post-mortem* evidence to show. In a remarkable case recorded by A. Money convulsions were apparently the cause of thrombosis of the cerebral veins and punctiform into the cortex. We are so much in ignorance of the nature and cause of convulsions that it would be rash to speculate too much, and when we find a lesion present after death it may be by no means easy to say whether it is the cause or consequence of the convulsions. Still, there cannot be a doubt that during a convulsion the cerebral veins are distended in consequence of the respiratory spasm, and a capillary hæmorrhage may readily result. Moreover, the nerve discharge is almost certain to be associated either before or after by disturbed circulation. We must bear in mind, however, that cases of hemiplegia following hemi-convulsions have occurred in uræmia, and, at the necropsy, no lesion has been found (Dunin, Cassel). One practical point, it is well to remember, is that in many of these cases hyperpyrexia is present. It is exceedingly probable that the severe convulsions may be due to this cause, hence the necessity of combating it with all the means in our power.

Acute Atrophic Paralysis.—It is worthy of notice that in the typical initial attack, the symptoms closely resemble those of the acute form of cerebral paralysis already described. There is often the same association with other diseases, as pneumonia, measles, diarrhoea; there is usually feverishness, perhaps drowsiness, coma, and convulsions, and the discovery comes as a surprise to the

friends and attendants that one or more of the limbs are limp and powerless. It is also interesting to find that a child may suffer from a similar attack and get well again without any paralysis taking place; then a few weeks later perhaps he is seized again with similar symptoms, which this time are followed by paralysis. In some cases reported the primary attack has been slight or indeed absent; in cases in which the lesion has not been extensive it is easy to understand the symptoms may not be marked and be readily forgotten or overlooked by the friends. With regard to the actual lesion, it is perfectly clear that there is an acute inflammatory lesion involving the anterior horns and more or less of the grey matter of the cord, more especially the lumbar and cervical enlargements. There is an inflammatory engorgement of the arteries which pass from the anterior spinal arteries of the cord to the anterior horns, accompanied by exudation of liquor sanguinis into the nerve substance, and apparently also in some instances by actual rupture of the capillaries and consequent minute hæmorrhages. The evidence points in the more severe cases at least to an engorgement of the cord, the most vascular parts, namely, the anterior horns being the spots where the greatest damage occurs; part of the damage done to the nervous mechanism is recoverable as time goes on by absorption of the effused inflammatory material, and part of the damage is irretrievable, on account of the breach made, so to speak, in the grey matter of the cord. That the anterior horns which contain such important groups of nerve cells, and consequently are exceedingly vascular, should bear the brunt of the damage is not surprising, still there is evidence to show that the rest of the cord does not entirely escape, for in one of my own cases there was some loss of sensation during life, and distinct evidence of slight inflammatory lesion of the vessels supplying the rest of the cord. It is important to bear in mind that evidences of inflammatory action in the shape of gorged vessels surrounded by leucocytes may be found six weeks or two months after the onset of the paralysis, this would point to the inflammatory action being subacute, the inflammatory stage lasting long after the symptoms of the primary attack have passed away. This must be remembered in any line of treatment we adopt.

Peripheral Paralysis.—Of these it is not necessary for me to speak at any length; as far as we know, they do not play an important part in the paralyses of early life. Injury to the brachial cords is apt to take place during labour by traction on the arm by an unskilled attendant, and a troublesome paralysis result. In congenital syphilis we occasionally see a peripheral paralysis due to some nerve lesion, though it need not be said we must be careful not to confuse the inability to use a limb in consequence of epiphysitis or periostitis with paralysis. In rickets periosteal tenderness and muscular weakness give rise to paresis, though even here we are still uncertain if the muscular weakness may not deserve the name of a paresis due to a nerve lesion or the effects of a poison. Of diphtheritic paralysis, and similar paralyses due to the effect of toxic matters in the blood, it is unnecessary for me to speak.

TWO CASES OF CONGENITAL AND INFANTILE CEREBRAL PALSY.¹

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THE patient, H. E., aged 19, was the third child of his mother, who states that she had a long and hard labour with him, but that no instruments were used. Ever since birth he has been palsied on his right side, the right leg getting fairly strong and useful, while the right arm has always been a good deal weaker than the left. About three years ago he had a sharp attack of rheumatic fever, with probably some dilatation of the heart, for there was then a mitral murmur, which I am glad to say has now vanished. For several years he has been subject to sick headaches, scarcely worthy, perhaps, to be dignified with the title of migraine, though the attacks were paroxysmal, of very severe headache, with vomiting, cold extremities, but with no affection of sight. Also about three years ago, while bathing directly after breakfast, he was suddenly seized with a fit of struggling, vomited freely, and was unconscious for some time. The next

fit took place eighteen months afterwards, the last about four months ago; there have been about five fits altogether. There is no aura or warning; he is unconscious for from three to five minutes, in apparently clonic general spasms, eyes turned upwards and rolling, face purple, no involuntary evacuations, petechiæ on face, and on several occasions he has injured himself severely.

Present Condition.—The head is obviously larger on the right side; from the vertex to the top of the left ear is more than half an inch smaller than from the vertex to the right ear; eye movements normal; heart slightly hypertrophied.

Right forearm an inch and a quarter smaller in circumference and half an inch shorter than the left. Its muscles react well to faradism. The chief wasting or want of development seem to have taken place in those arising from the inner condyle of the humerus. The interossei are very poor. The forearm is generally somewhat flexed on the elbow, the hand flexed on the wrist, the fingers overextended; supinator, biceps, and triceps reflexes are easily obtainable; the hand and arm are colder than on the left side, while the skin of the hand is markedly smooth and almost glossy; sensation seems unimpaired; grasp, though considerably weaker than that of the left hand, is fair, but he can do nothing demanding delicate touch or manœuvring.

Right leg is also half an inch shorter and three-quarters of an inch smaller than the left; knee-jerk and ankle clonus are evident; he walks with a slight limp.

The treatment latterly has been, of course, only directed to these epileptoid fits and the attacks of sick headache. I began with potassium bromide, 20 grains twice a day, afterwards small doses of tinctura belladonnæ and tinctura digitalis; finally, he is now taking potassium bromide and tinctura digitalis. For four months he has had no fit of any kind, and he has improved in every way, probably from their absence. For the sick headache I have found nothing so satisfactory in this, as in other cases, as the effervescent citrate of caffeine.

Three or four years ago he saw Sir James Paget on account of hemiplegia. Massage and galvanism were recommended, with the result that, as I have stated, the muscles are in very fair condition. The diagnosis is, I think, clear; it is a case of congenital cerebral spastic palsy, the birth palsy of Gowers, with epileptoid convulsions coming on rather late in puberty. I called it congenital because apparently it dates from birth, and cerebral because the difference of the two sides of the head, the spastic hemiplegia, the electric reaction of the muscles, and the later developed epileptoid convulsions leave me no room to doubt that the brain is responsible for his condition. I also, for the same reasons, feel pretty sure that the locality of the lesion is in the cortex cerebri, covering the leg and arm centres on the left side, and that lesion now is almost certainly a localised atrophy, the cause of that atrophy being, as Dr. Gowers says on this subject, meningeal hæmorrhage, which occurred probably during parturition. The association of those migraine-like attacks with epileptoid fits is suggestive of the near alliance mentioned by Dr. Hilton Fagge and others between migraine and epilepsy proper.

The second case I am bringing before you is that of a girl, aged 13, suffering from infantile cerebral palsy, and I am much indebted to the kindness of Mr. A. L. Peacock, of Lincoln, for an opportunity of seeing her and making notes of her condition. She seems to have been a fairly healthy child till she was 3½ years old; then, in close connection with an attack of measles—the mother's memory was rather shaky as to the exact relation in time of the two—she was seized with aphasia and right hemiplegia. Since then her speech has never been quite perfect.

Present Condition.—She is an intelligent child; head well shaped and equal on both sides; eye movements normal; sight excellent. Her right arm and leg are much weaker, and the right leg is half an inch less in circumference than on the left side. The right foot is always colder than the left. No deep reflexes are to be gained in right arm or leg, though the left knee-jerk is very obvious. There is no spastic condition of muscles of the palsied limbs. No affection of sensation was evident; micturition and defæcation were quite natural.

In this case again I think that the hemiplegia, with no great atrophy or ill development of muscles, accompanied by aphasia at its first onset, points indisputably to the lesion being cerebral. The association with measles, as occurred in seven of Dr. Gowers's, four of Dr. Abercrombie's, and four also of Dr. Osler's, forms a starting point for several diverse explanations. It is almost

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