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CONGENITAL AND INFANTILE SPASTIC PALSY

BY E. MANSEL SYMPSON, M.A., M.B. CANTAB.

Lincoln.

IN the *Practitioner* for February 1888, I communicated a number of cases illustrating the symptoms and pathology of this affection: in the following paper I propose to discuss four other cases which throw some further light on the subject. For *Cases III. and IV.* I am indebted to the kindness of Dr. Harrison and Mr. Charles Brook, of the Lincoln County Hospital. Some remarks on the probable pathology follow the brief record of the cases.

Case I. Spastic Diplegia.—Walter T., aged 13 (notes taken August 23, 1888).

History. His mother had an easy labour with him, and no instruments were used: he is the youngest of the family. He has been paralysed since birth, and the spasm and movements have come on since. He understands all that is said to him (mother's statement), though he cannot speak, write, walk, or sit.

Present Condition. Both hands are over-flexed on wrists, the fingers are in constant slow movement (athetosis), while there is mobile spasm of both arms, fore-arms, the muscles of neck, and the *orbicularis oris*. No deep reflexes could be got, owing to the spasm and restless movements.

He has athetosis also of his toes, his feet are over-extended and in a position of slight equino-varus, the legs being crossed. When one leg is lifted the other one rises too, and the pelvis "rocks" with any movement of the legs. No ankle-clonus could

be obtained;¹ the child had never walked. The muscles were small but well nourished. The head symmetrical in shape. He could overcome the spasm of his arms by tightly pressing them to his sides. He was cleanly in habits, and seemed to have no trouble of micturition or defæcation.

Case II. Spastic Hemiplegia.—Walter M., aged 25 (notes taken October 8, 1888).

History. When he was nine months old he had convulsions; ever since his left arm and leg have been palsied.

Present Condition. Left fore-arm semi-flexed, pronated and rigid, the wrist flexed, and the fingers over-extended. No atetosis or involuntary movements were noticed. The rigidity probably prevented the deep reflexes being obtained.

Left leg. "Clasp-knife" rigidity; clonus on tapping patellar tendon; no ankle-clonus. Both arm and leg are smaller in girth and shorter somewhat than on the other side.

Face. Naso-labial fold more evident on the left side, while laughing affects both sides equally. Tongue protruded rather to left side. *Eyes* natural to the ophthalmoscope. His head is small, but symmetrical. He is fairly intelligent, though laughing easily, and quite cleanly in his habits. He can walk very fairly now.

*Case III. Spastic Paraplegia.*² Lucy R., aged 10, one of fourteen, of whom only four are living. (In Lincoln County Hospital May 21 to July 23, 1888.)

History. Convulsions soon after birth: the condition of legs has been the same ever since.

Present Condition. Only partially intelligent: passes everything under her. Walks stiffly, inco-ordinately, and cannot keep her balance.

Legs. "Clasp-knife" rigidity. Reflexes (patellar and ankle-clonus) greatly exaggerated. Muscles react well to faradisation.

Roof of mouth arched.

Eyes. Right pupil dilated more than left (but both large); normal to the ophthalmoscope.

¹ See Dr. Gowers, *Diseases of the Nervous System*, vol. ii. p. 333.

² For notes of this case I have to thank Mr. W. H. B. Brook, M.B., House Surgeon, Lincoln County Hospital.

She improved mentally while in hospital; could walk without assistance, her gait being more co-ordinate.

Case IV. Spastic Hemiplegia.—Ada M., aged 9 (notes taken December 19, 1888).

History. Her mother states that she was her first child, and that she had a long and hard labour; that from birth it was noticed that the right hand was little used, and when she began to walk the right leg failed her. When she was two and a half years old she had "congestion of the brain," with fits, generally with "snatching" of the right side. She had several a year, once bit her tongue, and often foamed at the mouth. These have ceased for the last twelvemonth, but every now and then she has curious "sensations," and a sense of fear comes over her (? *petit mal*).

Present Condition. *Head:* left side markedly smaller than right; circumference, eighteen inches from one auditory meatus to the other, over the head nine and a half inches, and from root of the nose to the occipital tuberosity ten inches. Bulging forehead. Palate rather high. Slavers a little, mental state poor, easily excited to laughter. Cleanly in habits. Eyes natural.

Right hand generally flexed, fore-arm also flexed and pronated. The supinator, biceps, and triceps reflexes can be obtained at times. The right hand follows all the movements of the left—grasping as it grasps, and relaxing as it relaxes. Muscles somewhat small, as is the limb generally.

Right leg. Foot in position of equino-varus, with the big toe drawn up. Foot always much colder than the left one. "Clasp-knife" rigidity of the knee, the deep reflexes to be obtained occasionally.

Case I. is almost certainly one of birth palsy due to meningeal hæmorrhage. Probably the motor areas of his brain are now sclerosed and shrunken. *Case II.* is one of infantile cerebral palsy, due to meningeal hæmorrhage (?) in early life, extending over the leg, arm, and probably face centres of the right hemisphere. In *Case III.* the history is not very clear; it is probably one of congenital paraplegia, due to meningeal hæmorrhage over the leg centres on both sides of the brain. *Case IV.* belongs to the same order, remarkable in that the child can speak fairly well,

though the left side of the brain is injured, as well as for the coldness of her palsied leg and foot. A general rule is that only in infantile spinal paralysis is there marked coldness of the limbs. The epileptiform convulsions now sinking apparently from the "*grand mal*" to the "*petit mal*" type are noticeable.

Since cases are met with, both in congenital and infantile spastic palsy, of all degrees of severity, varying from the slightest affection of arm and leg to the spastic diplegia associated with idiocy, athetosis, mobile spasm, and epileptoid convulsions, and since the results of both are so similar, an attempt to show that all *spastic* cerebral palsies own the same pathology may be excusable.

First, as to the locality of the lesion. This is almost certainly in the *cortex cerebri*, for the following reasons among others. Theoretically, spasm would attend irritation or partial destruction of the cortex in the motor areas. And this has been proved by facts.¹ Then again, in many of these cases the spasm ceases during sleep or under chloroform; this, without permanent contractures, points to functional activity of the cerebral cortex.² Then epileptiform convulsions, which are present in the severer cases, sometimes unilateral, sometimes general, are to be referred to the intermittent activity of partially damaged cortical cells, as in Jacksonian epilepsy. Also, the mental poverty of many of these patients, who are occasionally almost idiotic, tells in favour of this view.³ The motor affection and the state of intelligence, however, do not go *pari passu*, nor do the convulsions; apparently the mental state would vary more or less with limited or wide-spread cortical damage. The conditions of life of many of these children prevent their sharpening their wits with their fellows, either at work or play; *e.g.*, in *Case I.* the involuntary movements prevented him from speaking or writing. Lastly, the records of *post-mortem* investigations are greatly in favour of a cortical lesion, those quoted by Dr. Gibney and by Dr. McNutt⁴

¹ Dr. Gowers, *op. cit.*, vol. ii. p. 82, and footnote.

² Dr. Broadbent, *Lancet*, 1887, vol. i. p. 174.

³ Dr. T. Claye Shaw, *St. Bartholomew's Hospital Reports*, vol. ix. p. 130.

⁴ Dr. Gibney, *Medical Record* (New York), vol. xxx. p. 393. Dr. McNutt's case also is illustrated in Dr. Gowers' *Diseases of the Nervous System*, vol. ii. p. 384.

especially so. For individual cases two very interesting ones are to be found in the *St. Bartholomew's Hospital Reports*, vols. xv. and xvi., both cases of spastic diplegia, due to sclerosis of the motor area chiefly in one, and of the general surface of the brain in the second.¹ A reference to an interesting case may not be out of place here. A man had convulsions when eight years old; was paralysed in his left arm and leg; was lazy and useless, and latterly for the last twenty years suffered from daily fits (*petit mal*). *Post mortem*, there was a cyst containing fluid pressing on the brain and causing atrophy of the motor area on the right side, in the ascending parietal and frontal convolutions. Another case recorded by Dr. Ashby² of spastic diplegia due to diffuse sclerosis of the cerebrum is also in point.

Causation.—Little need be said on this score about the congenital cases. Hæmorrhage into the meninges will explain most of the cases satisfactorily. This may take place before or at birth; if fatal soon, the blood will be found effused, as in Dr. McNutt's case; later on there will be atrophy of the convolutions involved. Whether the so-called porencephalus is really due to actual failure of development seems doubtful; it seems quite likely to be caused by hæmorrhage into the meninges early in foetal life. "The brain-substance and the meninges in the neighbourhood of the defect often show changes similar to those which in later life are known to follow upon sub-meningeal anæmic and inflammatory softening."³ Dr. Bramwell's⁴ case may be referred to, where birth had been helped by instruments, a depression on the right side of the head was noted by the father, and left hemiplegia with constant epileptoid convulsions followed. With regard to infantile or acquired spastic palsy of children, a few reasons for the theory of meningeal hæmorrhage may be given. Fatty degeneration of cerebral vessels is common, according to Nothnagel,⁵ in cachectic children, and may render the occurrence of hæmorrhage more

¹ *Brain*, vol. xi. p. 225.

² Dr. Ashby, *Brit. Med. Journal*, 1886, vol. i. p. 155.

³ Ziegler, *Special Pathological Anatomy*, by Dr. MacAlister, sect. xi. art. 630.

⁴ Dr. Byrom Bramwell, *Brit. Med. Journal*, 1886, vol. ii. p. 1035.

⁵ Nothnagel, *Ziemssen's Cyclopædia of Medicine*, vol. xii. p. 80.

probable. Many cases date the origin of palsy from acute eruptive fevers, particularly measles; some after diphtheria, where the special paralysis is so strikingly unlike this; and a few follow whooping-cough. These, then, are supposed to exert injurious action on the walls of blood-vessels. Now, it is in the early years of life that tubercle selects the meninges of the brain for special attack.¹ Is there any reason for this? During infancy, of all the organs of the body the brain is in the most actively developing state, changes the most delicate are going on, evolving the complex system of cells and nerve-cords out of comparative simplicity. This may serve to explain the apparent preference, and at the same time suggest that, if a child's blood-vessels are disposed to rupture, this will be likely to take place in the meninges. Now convulsions immediately preceding the palsy are a common feature in the history of these cases. Convulsions, of course, are so common in children that too much stress must not be laid upon their occurrence. But meningeal hæmorrhage causes convulsions, frequently of an epileptiform nature.² Or, *vice versâ*, the convulsions may cause the hæmorrhage, according to Dr. Goodhart. When these cases come to be examined *post mortem*, sclerosis and atrophy of the motor areas or more of the brain is the most frequent condition. Dr. Gee's second case³ may be mentioned as one instance, the case quoted above from *Brain*⁴ will serve for another and a somewhat different one. Mr. Hutchinson's case of diplegia⁵ may be due to the same cause, a cyst on either side pressing on the cortex, the cysts being a remnant of old hæmorrhage.⁶ Dr. West's case seems to belong to this class, as well as Dr. Holt's⁷ and Dr. Taylor's,⁸ whether the causation I have mentioned be the correct one or not. Dr. Wolfenden's⁹ cases are very similar to several given or quoted

¹ See table given by Dr. Gee in article on Tuberculous Meningitis, *Reynolds's System of Medicine*, vol. ii. p. 372.

² Nothnagel, *Ziemssen's Cyclopædia*, vol. xii. p. 175.

³ Dr. Gee, *St. Bartholomew's Hospital Reports*, vol. xvi. pp. 32, 33.

⁴ *Loc. cit.*

⁵ Mr. Hutchinson, *Trans. Path. Soc.* xxxiii. p. 27.

⁶ Dr. West, *Brit. Med. Journal*, vol. i. p. 157.

⁷ Dr. Holt, *Med. Record* (New York), vol. xxx. p. 393.

⁸ Dr. Taylor, *Trans. Clin. Soc.* vol. xvi. p. 243.

⁹ Dr. Wolfenden, *Practitioner*, vol. xxxvii. p. 161.

here, and perhaps can be explained as due to the same pathological changes.

If infantile spastic paraplegia be purely a spinal disease, like the adult form described by Erb and Charcot, there should surely be "an absence of all cerebral disturbance."¹ But in one of Dr. Gee's cases, John B., there was athetosis of the right hand, while Margaret B. had rigidity and mobile spasm of the arm and legs. The mental state of *Case III.*, above, was very poor, the like being noted by Dr. Gee of three of his; and Dr. Mackay² says of his patient that "she smiled idiotically." Of course there may be deficiency of brain as well as of spinal cord, but it is more reasonable to invoke only one lesion. It would be interesting to make out in all these cases the state of the superficial reflexes, which are generally supposed to be diminished or abolished in cerebral disease.³ And should athetosis of the toes occur—athetosis being invariably a cerebral symptom—this would lend greater force to the argument for the cerebral origin of most if not all of the cases of congenital or infantile spastic paraplegia.

¹ Erb, *Ziemssen's Cyclopædia*, vol. xiii. p. 623.

² Dr. Mackay, *Brit. Med. Journal*, 1887, vol. i. p. 66.

³ *Trans. Clin. Soc.* xvi. p. 243.