Thesis for M.D. degree of the University of Edinburgh by
John Charles Atkinson, M.B.
on
"Some forms of paralysis in children, with special reference to those of congenital origin.

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I propose to discuss three forms of Paresis based on cases which have come under observation:—viz.,
A: Congenital Spastic paralysis.
B: Posterior-Lateral Spinal Sclerosis.
C: Anterior Poliomyelitis.

A: Three distinct classes of cases may be recognised:

1. Where there is arrested development of the motor tracts in the brain or spinal cord.
2. Intra-cranial disease of nervous system.
3. Where brain or spinal cord has been injured during delivery.

The three most frequent conditions are:
a. Porencephalus cerebralis
b. Injury to motor cerebral centres by instrumental delivery.
c. Injury to Spinal Cord by traction during delivery.

In Porencephalus there is a deep depression on the surface of the cerebral hemisphere at the bottom of which is an opening leading into one or both of the Lateral ventricles. In all cases there is defective development in the Rolandic area.
If the injury is during birth, it is most frequently due to pressure of forceps, involving the upper part of the Rolandic area, i.e. the leg centres either as the direct effect of pressure or by a meningeal haemorrhage, limited to the upper part of the central convolutions.

As a result of this, a descending degeneration of the pyramidal tracts, which are distributed to the lower extremities, is set up.

The contractions and rigidizable irritability is somewhat different from that generally observed in paralysis depending on one-sided cerebral disease resembling rather that which is met with in the upper extremity in cases of hemiplegia with late rigidity.

The innervation of the legs is interfered with from both hemispheres.

In a number of cases of Infantile Spastic Paraplegia collected by Dr. Ross the presentation at birth was always a "Breech" and he believes that injury was inflicted on the vertebral column and Cord by excessive traction.
Injury to a vertebra or a spinal meningeal haemorrhage may set up a transverse myelitis, causing a descending degeneration of the Pyramidal tracts.

In cases of congenital paraplegia where there is no history of injury, the lesion arises from intra-uterine disease or arrest of development of the motor tracts.

The following case of congenital spastic paralysis apparently originated by an injury to the brain during instrumental delivery.

A boy, about 16, paralysed since birth; family history good; born by aid of forceps; complains of weakness and rigidity of both legs; he is thin, but intelligent and looks healthy; walks with a waddling gait with a tendency to twist his body to the left; both knees are flexed owing to contraction of hamstrings and drawn inwards by contraction of adductors, the feet therefore incline outwards; the limbs in progression describe arcs with the convexity outwards. As these arcs intersect in front the progression is cross-legged. The inner border of the
foot first comes in contact with the ground and is scraped forwards. The left leg on measurement is found to be an inch shorter than the right, which accounts for the tendency to twist the body to the left when walking.

The superficial reflexes are absent but the deep are greatly exaggerated. Ankle Clonus is however not always present and it cannot be obtained when the contracture of the Calf muscles is increased, this contracture causing a fixed posture of Talipes Equinus. When the patient is placed on his back the foot is rigidly extended on the leg and the leg upon the thigh, whilst the great toe is flexed at the metatarsal phalangeal joint. Sensibility is not impaired. The limbs are thin but there are no evidences of trophic atrophy. There is some rigidity of the Right upper extremity, the forearm being slightly flexed upon the arm and the extremity cannot be completely extended. The right elbow reflex is exaggerated and the heads of the paretal tones being flattened, meeting at an acute angle.
The distribution of the paresis, in this case, shows it to be of cerebral origin; the history of instrumental delivery and the existence of paresis from birth point to an injury of the upper part of the central convolutions affecting the leg centres and to a certain extent that of the right upper extremity. The case seems to confirm the statement that in congenital spastic paraplegia, the patellar-tendon reflex is exaggerated whilst ankle clonus cannot be produced. This may be due to arrested development in the parts of the Pyramidal tracts which are distributed to the legs and probably the contraction of the Calf muscles opposes the development of the condition upon which ankle clonus depends.

The absence or diminution of superficial reflexes in cerebral disease was pointed out by Rosenbach and Jacsonwitz.

B: Posterior Lateral Spinal Sclerosis of Congenital Origin.

This condition also known as Friedreich's Disease: Hereditary or Generic ataxia
has of late years come in for more general consideration.

The disease affects certain families and was described by Friedrich in 1861. It is a degeneration of the posterior columns of the spinal cord and the posterior nerve roots, and in some cases the anterior and lateral columns are involved. In microscopic sections, the cords show marked degeneration in the Columns of Goll and Burdach; the crossed and direct Pyramidal fibres being also implicated.

The disease is generally seen in females. Symptoms generally begin in childhood, 4 to 7 years, or they may not appear until puberty.

At the Commencement of the disease there is incoordination of movement or a sense of weakness in both upper and lower extremities; rapid development; sometimes the ataxia is confined to the upper extremities. Lancinating pains are absent or developed late in the disease; muscles of the head, speech and eyeballs may be affected.
pupil phenomena and defects of vision are usually absent; tendon reflexes are usually absent; sensory functions not much impaired but there is usually tactile anaesthesia; the plantar reflex is often retained although knee-jerk is abolished.

After incoordination has existed for some time, paralysis and contracture develop, more marked in the lower extremities than in the upper. After a time patients are unable to walk and free movements of the limbs become impossible. Bedsores are not developed and the sphincters escape. The mental faculties are not impaired. The speech has at first a drawling character and gradually becomes unintelligible.

Vaso-motor disturbances are common, as polyuria, salivation, hyperhidrosis; the feet are frequently "clubbed" and there may be "wrist-drop".

Case E. P. Female, age 19. No history of ataxia in family. When seen there was slight tremor of the hands on voluntary
movement: slight lateral nystagmus; marked ataxia of the lower limbs with very slight loss of sensibility: articulation indistinct; no cerebral symptoms or hysteria: Symptoms commenced at 16 when menstruation became irregular and finally ceased. The pupil phenomena and visceral symptoms of Tabes were absent: the spastic features of ataxic paraplegia were absent and the tremors, nystagmus and cerebral conditions of disseminated sclerosis are not present.

Under rest, tonics and massage the case improved.

Case 2. 13 oy aged 6. There was gradually developing incoordination of movement spreading from the lower to the upper half of the body and subsequently the organs of speech became affected: ordinary sensibility, special senses and functions of the brain were not impaired: there was no paralysis of Sphincters:

O: Anterior Poliomyelitis

Synonyms: Infantile Paralysis or Essential
Paralysis of Children.
Anterior Poliomyelitis or inflammation of the Cells of the Anterior Horns, may exist in two forms:

1. Combined with Lateral Sclerosis
2. As an independent affection.
The latter is the form most frequently met with in children.
The Cells in the Anterior Cornua are the seat of inflammation which either causes Atrophy or destruction.
If acute the inflammation of the Cells is accompanied by sudden Paralysis and if severe the Cells undergo acute pigmentary degeneration. Owing to the inflammation of the Cells, their trophic function is withdrawn from the nerve fibres connected with them and these nerves degenerate, this being followed by rapid fatty degeneration and atrophy of the muscles connected with them.
The symptoms of the disease appear to show that these Cells not only preside over muscular movement but also preside over the nutrition of the muscles.
The inflammatory changes in the cells are accompanied by very different symptoms from those produced by chronic inflammatory degeneration. In anterior cornual acute inflammation only a few spinal segments are involved; in some cases one side may escape altogether. In one recorded case the disease attacked the right horn of the cervical segments and the left horn of the lumbar segments; he had paralysis and atrophy of the right arm and paralysis of the left leg.

If the case is not too severe there may be complete recovery.

Beyond a slight numbness, there is no disturbance of sensory functions and the bladder and rectum escape.

Etiology: Exposure to damp and cold; Dentition; Eruptive Fevers; Fractures; Shock etc.

The disease generally occurs in boys, before the age of two and seldom after seven, and most frequently in the hot months of the year.

The onset is marked by a sudden elevation
of temperature, malaise and sometimes convulsions; the fever - which may be continuous or remittent - lasting one or more days; there may be concomitant severe pains in the limbs, probably depending on a condition of nerve inflammation, occurring simultaneously with the changes in the Anterior Cornua (Powers); muscular twitchings or tremors; convulsions; delirium and a sense of numbness. Sudden paralysis follows the fever; occasionally, fever is absent, the paralysis coming on suddenly and being the first evidence of the disease. The paralysis is usually greatest at its commencement, but the initial loss of power will to a certain extent pass off, the permanent palsy usually affecting the legs.

The paralysis may be unilateral (Hemiplegia) or bilateral (Paraplegia); in some cases all the limbs are affected (Complete paralysis) or in rare cases only one limb (Monoplegia)

In the so called "Erb type" of the disease
The scapular muscles are especially affected. The loss of power may be in different groups of muscles in the two limbs, e.g., the muscles of the thigh in one and those of the leg in the other.

The paralyzed limb is flaccid, colder than the other and usually blue in colour, probably from vaso-motor disturbance. It has been shown that a dose of Pilocarpine will cause sweating in the healthy limb but not in the paralyzed one; the Knee-jerk is as a rule lost in the affected limb, probably because the Quadriceps is wasted, but cases are recorded where the paralysis was limited to the leg below the knee, the Knee-jerk remaining active.

The general health is good, but if the muscles of the trunk are affected, the respiration may become embarrassed and constipation is frequently troublesome. The muscles may be conveniently classified into

a. Those which have escaped the disease
b. The muscles affected.
The affected muscles may be again divided into three groups:

1. Those capable of complete recovery.
2. Those capable of partial recovery.
3. Those incapable of any recovery.

If a piece of muscle, which is progressively atrophying, is obtained by Duchenne’s trochar and placed under a microscope, it will be found to be in a condition of fatty degeneration.

C. Case: Female child, aged 3 years after recovery from a mild attack of measles. Fever came on suddenly and dyspnoea was present on the third day or the fourth day. The breathing became stertorous and the patient sunk into a stupor from which she could be roused but only to relapse. In a few days the fever and stupor disappeared when it was noticed that the muscles of the right arm and leg were affected, the right foot being dragged along the ground. There was no disturbance of speech and the child ultimately recovered completely.
References:


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