CEREBRAL PALSY – PART I

AETIOLOGY

(Aetiology and methods found useful in its treatment Part II)

compiled by
Physiotherapists of the Pretoria School for Cerebral Palsy

Cerebral palsy is a condition caused by a brain lesion to the immature brain. Signs and symptoms depend on the area of the brain injured and the extent of the injury. Cerebral palsy usually involves physical abnormalities (e.g. increased or fluctuating muscle tone, retarded developmental patterns, late disappearance or pathological retention of primitive reflexes, etc.) and is sometimes accompanied by mental retardation.

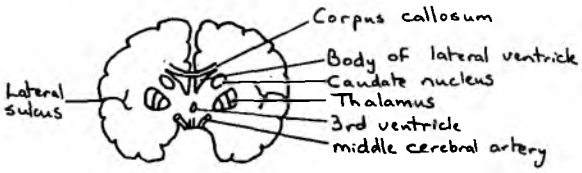

Fig. 1. Schematic representation of a section through the cerebrum.

CAUSES

A. Prenatal factors

1. *Trauma:*
   (a) Blow on the abdomen especially during 8th–9th month of pregnancy with resultant occipital contusion.
   (b) Car accidents, especially towards end of pregnancy.
   (c) Attempted abortion—by chemicals or some type of instrument, e.g. knitting needle.

2. *Diseases:*
   (a) Virus infections (e.g. measles, influenza, mumps, chickenpox). These cause a slight meningo-encephalitis concurrent with the mother's illness. It usually resolves as the mother recovers, but sometimes with a residual brain lesion caused by adhesion formation of the external fluid spaces. A cyst may form or there may be blockage of the external or internal fluid spaces with resulting hydrocephalus.
   (b) Syphilis and tuberculosis have become very rare causes.
   (c) Toxoplasmosis—early in pregnancy this causes a malformation of the foetus, but later infections cause chorioretinitis and hydrocephalus with calcification of the subventricular areas of the brain.
   (d) Rubella may cause heart lesions, cataract and deafness or microcephalus with hydrocephalus externus and internus.

3. *Circulatory disorders:*
   (a) Deficient oxygen especially in the first few weeks and months of pregnancy when no bloodvessels are found in the embryo.
   (b) Haemorrhages during pregnancy, especially with threatening abortus.
   (c) Erythroblastosis causes interference with blood-vessel permeability—due to the iso-immunization of the Rh-negative mother by the Rh-positive foetus. It is accompanied by kernicterus (the basal nuclei are stained by bile, with toxic degeneration of the nerve cells).

   Anti-Rh-agglutinins in the mother's blood pass through the placenta and agglutinate the red corpuscles of the foetus. It is treated by transfusion of O-Rh-negative blood directly after birth.

4. *Other:*
   Effects of Radiation—Röntgen rays may cause bloodvessel changes with serious brain parenchyme interference, especially during the second half of pregnancy. Even ultrasound waves have been thought to cause brain lesions.

5. *Metabolic Disorders:*
   (a) Vitamin deficiency.
   (b) Deficiency of trace-elements (e.g. copper).
   (c) Diabetes mellitus.
   (d) Protein—deficiency dystrophy of the mother.

6. *Bloodfactors:*
   (a) *Rh-factors—as described above (3c).*
   (b) Incompatibility of the bloodgroups of the mother and father (e.g. mother group B and father group O—both Rh-positive). This is very seldom seen.

B. Natal factors

1. *Mechanical lesions:*
   (a) Haemorrhage—subdural haemorrhages may be due to tears of venous vessels (e.g. sinusses or basal veins below the Tentorium) which may be caused by the friction action on the head in the birth canal.
   (b) Traumatic tissue damage is seldom seen e.g. contusion due to forceps delivery or very difficult birth through a narrow pelvic outlet.

2. *Asphyxia:*
   (a) Placenta praevia.
   (b) Prematurity.
   (c) Lengthened or difficult birth due to inadequate uterine contraction or a false rhythm of the uterine contractions e.g. during hypertonic continuous contracture with no periods of rest in between. The intra-uterine pressure during labour contractions is much higher than the systolic blood-pressure of the baby. The effect of asphyxiation is increased by the usual delay before normal breathing begins.

   These lesions are mainly found in the cortex and basal ganglia with scar formation, so-called Plaques fibromyeliniques (in the cortex) and

Status marmoratus (in the brainstem and basal ganglia). The outer layers of the brain are more sensitive to circulatory damage then the brainstem, thus the baby will survive in spite of extensive cortical damage. Diffuse scar-tissue is formed as a result of ischaemia (Fig. 2). Hypoxaemia causes the formation of more localized single areas of scar-tissue and cyst formation in the white matter of the brain.

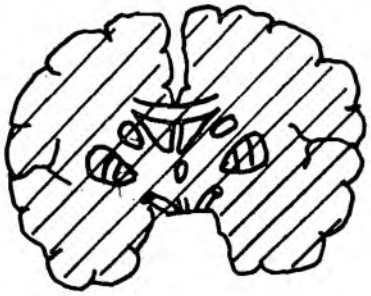

Fig. 2. Diffuse brain damage due to asphyxia and resulting ischaemia (quadriplegia with athetosis).

(d) Venous congestion leading to oedema causes a typical conic lesion of the brain, the base stretching from both sides of the longitudinal sinus. The apex may reach down to the brainstem ganglia. This causes different types of spastic diplegia with possible athetosis of the arms and face.

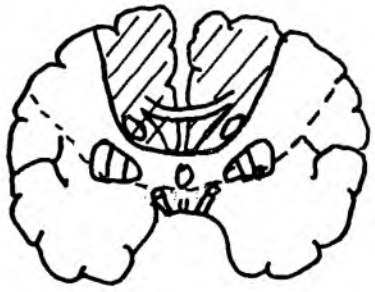

Fig. 3. Venous congestion with typical conic lesion.

////// Pure spastic diplegia.

Area marked with dotted line:
Spastic diplegia with athetosis of arms and face.

(e) Many of the typical childhood hemiplegias arise from damage during birth, causing the formation of porencephalia (the presence of cysts in the surface of the brain due to an arrest of development or to birth haemorrhage: Black's Medical Dictionary—W. A. R. Thomson, M.D.).

The condition is generally associated with serious mental defect. The lesion, usually found around the fissure of Sylvius, is probably caused by a temporary ischaemia in the area of the carotid artery. Often the formation of porencephalia is a limited tissue defect so that the rest of the brain is relatively intact resulting in only slight mental defects. More diffuse damage causes more serious mental defects.

Fig. 4. Spastic hemiplegia.

(f) Cord around the baby's neck.

C. Postnatal factors (up to fifth year of life)

1. *Circulatory disorders:*
   (a) Whooping cough eclampsia—bloodvessels are weakened and remain more permeable or large adhesions are formed.
   (b) Continuous convulsions ("stuipe") where the brain cannot cope with the angio-spastic circulatory disturbance.
   (c) Cerebral emboli.
2. *Infections:*
   (a) Perivenous encephalitis and meningitis.
      (i) It may occur concurrently with influenza, mumps, rubella and chickenpox. Diffuse lesions in the brain and spinal cord are produced.

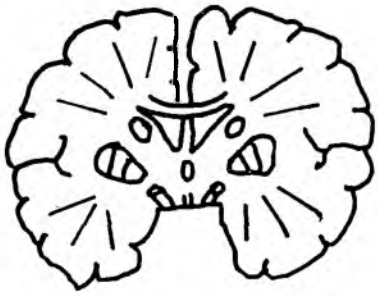

Fig. 5. Diffuse lesions due to perivenous encephalitis and meningitis. Various neuropathological pictures may result.

      (ii) It may also be caused by vaccinations, especially if done late (between 3rd–9th or up to 12th year of life). The danger lessens after 17th year.
      (iii) Penicillin and sulphonamide damage of the brain.

(b) Meningismus may cause adhesions in the external or internal fluid spaces, which may cause obstruction to the normal flow of the meningeal fluid.

3. *Metabolic disorders:*
E.g. nutritional dystrophy of the newborn.

4. *Trauma:*
Car accidents mainly, but any kind of trauma causing haemorrhage in the brain and skull fractures. Car accidents cause increasingly more brain lesions as speed and traffic increases.

5. *Brain tumors:*
E.g. Neurofibromatosis.

CLASSIFICATION

A. Spastic Type (Hypertonic)

1. *Quadriplegia*—all four limbs, neck and head, muscles of articulation and mastication and eye muscles are all affected. Usually there are perceptual problems as well as mental defects.

2. *Diplegia*—legs and lower trunk mainly affected.

3. *Double hemiplegia*—arms and headmuscles sometimes more spastic than legs. The trunk muscles are also affected.

4. *Hemiplegia*—one side of the body and the half of the head and face are affected.

5. *Triplegia*—sometimes the two lower limbs, but only one arm is affected (the other arm seems normal). It may be accompanied by spasticity of some head and face muscles. It is actually a quadriplegia with one arm fairly normal.

6. *Monoplegia*—only one limb, usually an arm, is affected. It is actually a hemiplegia with the leg fairly normal.

B. Athetoid

The trunk, limbs and facial muscles as well as those of articulation and speech are affected.

C. Ataxia

Mainly characterized by poor balance and inco-ordination.

D. Hypotonic spastic

The so-called "floppy" child, is a quadriplegia with the head and face also affected. Hypotonia usually develops into spasticity, ataxia, or athetosis in later life (e.g. about puberty).

E. Hyperkinetic

A hyperkinetic child with fairly minimal brain damage, with little or no physical disability (except awkwardness or stumbling or impaired fine co-ordination) but with mental retardation, often presents serious perceptual problems and a very short concentration span.

F. Mixed cases.

CHARACTERISTICS

A. Spastic Type

1. *Quadriplegia:* This is due to diffuse and extensive lesions of the brain. It may be combined with athetosis or rarely with ataxia, depending on the extent of the lesion.

Quadriplegia is characterised by increased muscle tone, usually of a rigid rather than clasp-knife type. One half of the body is usually more affected than the other and spasticity is usually worse in the lower extremities.

Predominant flexion of the hips and adductor spasticity leads to scissoring of the legs. There may be flexor spasticity or hyperextension of the knees with plantarflexion of the feet, which may be in equino-varus or -valgus position. The shoulders are protracted and the arms adducted and flexed; the elbows are flexed and pronation of the forearms, the hand in ulnar deviation, fingers flexed and the thumb in the palm of the hand. There is often hyperextension at the metacarpalphalangeal or proximal or distal interphalangeal joints (the typical swan-neck deformity). The face is rigid and slow to react, speech is usually affected and a tongue thrust and mandibular stretch reflex are present. There is difficulty with swallowing which results in dribbling. Divergent or convergent squints are common and often there is very little eye movement. Normal development is retarded and mental retardation is usually present. Abnormal tonic reflex activity is present. The Moro reflex does not disappear and often is very marked. Certain primitive reflexes, normally present in the new born, do not disappear at the normal time (e.g. Asymmetric Tonic Neck Reflex and others). Other reflexes like the Sprungbereitschaft reflex appear late or are absent.

The severity depends on the extent of the brain lesion.

2. *Diplegia.* The severity depends on the extent of the brain lesion. In diplegia, the characteristics of the lower extremities and lower trunk is similar to the quadriplegia, but the upper trunk, face and upper limbs may show little or no signs of spasticity.

3. *The double hemiplegia* is also similar, except that the upper trunk and upper extremities are more severely affected than the lower trunk and lower extremities.

4. and 5. *Triplegia and monoplegia.* In triplegia and monoplegia too the affected limbs show similar characteristics of spasticity. In hemiplegias only one half of the body shows these signs. The extension element of the positive supporting reflex is more dominant in hemiplegias with the result that the leg is usually adducted, internally rotated and flexed at the hip, extended at the knee with plantarflexion and inversion of the foot. The shoulder is retracted.

B. Athetoid

The term "Athetosis" was first used by Hammond 1873 and means "without a fixed position" or more generally "without co-ordination". It is caused by damage to the extra-pyramidal system. The tone fluctuates continually causing grotesque mass movements (which are not necessarily due to increased reflex activity) especially when voluntary movements are attempted. The muscles of facial expression, the tongue, breathing, swallowing and articulation are affected. This often leads to frustration, especially in moments of emotional excitement. It is necessary to include emotional control and quietness in therapy. Athetosis may be accompanied by spasticity. Athetosis is often not observed in the very young baby, and usually appears when intentional movements are attempted later. Due to the mass movements, difficulty with articulation, mastication and swallowing as well as the common deafness, the intelligence of these children is often underestimated. Yet, when athetosis is not combined with other defects, the average athetoid usually has a higher intelligence than the average spastic. The frustrations these children experience often leads to the development of faulty social relationships. Even paranoid tendencies may sometimes develop.

Intensive physiotherapy to teach quietness and control of grotesque movements, must be accompanied by remedial teaching. Progress will be determined by the degree of existing central personality functions (e.g. how much the child understands, how much he absorbs, how much drive and interest he has to communicate with the world etc.).

C. Ataxia

Ataxia presents a disorder of the afferent and efferent pathways. It can be accompanied by spasticity. There are clear co-ordination disturbances, especially of the finer movements which are often accompanied by an intention tremor. The co-ordination disturbances are not to be confused with those which in athetosis are caused by fluctuating tone. In addition there are balance and equilibrium reaction

disturbances. These balance and equilibrium reactions develop later than normal and remain slow and weak, the children often walking with a wide base or a type of Parkinsonian gait (this is seldom seen). The whole developmental pattern is retarded. Usually there are also disturbances in sucking and swallowing in the baby; and sometimes breathing difficulties too. These signs are less clear in the small baby, but become more clear with increasing age. A very typical sign is the intention tremor present during movements of fine co-ordination.

D. Hypotonic spastic

These children show hypotonus with extremely weak head and trunk control. They are often mentally retarded and lacking in motivation. They show many of the abnormal reflex patterns even in later life. Athetoid, spastic and/or ataxic tendencies usually develop later (about puberty).

E. Hyperkinetic

These children have mainly frontal lobe lesions causing defective intelligence, various types of perceptual problems, lack of concentration and hyperactivity or clumsiness. Often a retarded motor development may be noticed—this may be due to the defective intelligence. Usually there is little or no motor disturbance.

The characteristics of the cerebral palsied are seldom observed at or shortly after birth. Soon, however, the following anomalies of the simplest vital functions may be observed:

1. Difficulty in sucking, swallowing and later chewing too.
2. Breathing anomalies—e.g. very shallow breathing, frequent bronchitis or pneumonia, etc.
3. Peculiar crying:
   (i) Very soft and without strength.
   (ii) More often a shrill, piercing cry.
4. Soon a disturbance in concentration power is noticed. The eyes of the baby cannot follow moving objects or fixate. A squint is often present.
5. The child does not kick (or kicks more with one leg and keeps the other fairly still). He does not attempt to grasp. His hand-mouth co-ordination and hand-feet motor development are slow to develop.
6. Primitive reflexes are slow to disappear. Positional reflexes appear later than normally, and the normal developmental pattern is retarded.
7. Epileptic attacks may occur in different forms and intensity.
8. According to Collis,[1] intelligence in more than 75 per cent of Cerebral palsied children, is below normal and less than 25 per cent have a normal I.Q. About 25 per cent have severe mental retardation and will never be educable, and could not even be trained to perform the simplest tasks or own bodily care. About 50 per cent have less severe mental retardation in different degrees and may be educable or may at least be trained to perform basic activities of daily living and simple tasks.

BIBLIOGRAPHY

1. Collis, E., and others: *The Infantile Cerebral Palsies.* (W. Heineman, London, 1956).
2. Lindemann, Prof. K., and others: *Die Infantilen Zerebral Paresen.* (George Thieme Verlag, Stuttgart, 1963).

This last book was used for all reference to the aetiology and aided in compiling the characteristics of the types of Cerebral Palsy.

Part II, about the treatment for Cerebral Palsy, will be published in the next magazine.

GENERAL

RETIREMENT—RESIGNATION

Miss G. Jones has retired from the post of Principal of the Physiotherapy School, King Edward VIII Hospital, Durban. Mr. C. A. Liggins, M.C.S.P., H.T., Dip.T.P., has taken over this responsible position and it is understood that Miss Jones will return to a teaching post at the school following overseas leave.

Mr. J. Stockton, M.B.E., S.R.N., M.C.S.P., H.T., Dip.T.P., has resigned from the Physiotherapy College, Pretoria and his place has been filled by Miss Anne Hendry, who is Acting Principal from April, 1971.

Mr. Stockton, who has left for overseas played a very active part in Physiotherapy in South Africa from the day he arrived in the country in April, 1965. He was appointed to the National Executive Committee and the Training and Registration Committee, where his past experience, clear thinking and quick appreciation of the importance of a situation under discussion were of inestimable value. He will be greatly missed in all fields of the profession, teaching and administrative in particular.

We take this opportunity of wishing both these ex-Principals "Good Luck" for the future and offer them our sincere thanks for the loyal service they have given to the profession in South Africa.

NOTICE TO ALL MEMBERS

CHANGE OF ADDRESS

All members of S.A.S.P. and those registered with S.A.M.D.C. are reminded of the importance of keeping the Society and the South African Medical and Council informed of any change of address. Those members who have recently married must accompany their change of address with a photostat copy of their marriage certificate.

NORTHERN CAPE SOCIETY FOR THE CARE OF CRIPPLES

RE: POST-MATRIC BURSARIES

The Executive Committee of the Northern Cape Society for the Care of Cripples has created a Bursary of R200 per annum for Coloured or Bantu Post-Matric students who wish to qualify as Social Workers or Physiotherapists at recognised Universities or Training Schools. Bursary holders would be required to work in the Cripple Care field for a suitable number of years, if necessary.

Application forms may be obtained from:

The Secretary,
Northern Cape Society for
the Care of Cripples,
P.O. Box 928,
Kimberley.