

Medicine for Managers

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Cerebral Palsy

Cerebral Palsy is a general term used to describe a disorder which results from damage to the brain before, during or after birth before development is complete (usually before age 3) and is associated with disordered posture and movement. This cold definition conceals a group of individuals who so often meet its challenges with fortitude and commitment and who live to enjoy

Damage to the brain may occur as a result of a developmental abnormality or can occur as a result of infection during pregnancy, premature birth or trauma during the delivery or to anoxia (oxygen shortage) after birth.

Infections during pregnancy may include rubella (German measles), herpes virus, cytomegalovirus or toxoplasmosis (parasite). Sometimes the condition develops as a result of a genetic mutation. It may occur as a result of trauma or injury to the baby's head which can occur during the delivery itself or as a result of injury (accidental or non-accidental) during the first few months of life.

Lack of oxygen after birth can also cause damage to the very sensitive brain cells. Sadly, cerebral palsy has resulted in a large amount of litigation as parents sue obstetricians and paediatricians in an attempt to prove that the complication was avoidable during delivery or in the immediate post-delivery period.

The causes of cerebral palsy have resulted in a classification based on time of occurrence and the nature of the cause, thus:

Pre-natal

Natal

Post-natal

Or:

Congenital (the result of developmental or genetic malformations)

Acquired (infection, anoxia, etc.)

The symptoms of cerebral palsy vary according to the type of damage sustained. Some are described as **spastic** where muscle tone is increased (the body is stiff) and others as **athetoid** where muscle movements are increased (the body may be weak).

Yet others are described as **ataxic** where muscle actions are uncoordinated, dysrhythmic or lacking accuracy. Commonly the condition displays elements of all three types of movement.

The disorder is estimated to affect about 1,600 children a year. Sadly, at the present time, there is no cure.

The central feature of CP is the disordered motor (muscle) movements, in some there will be difficulties in thinking, communication, learning and sensation.

As the child grows the movements may be **hypertonic** (increased muscle tone, making them appear stiff) or **hypotonic** (making them appear floppy).

The physical disabilities, including difficulties with communication, may result in those people who do not understand the condition equating lack of co-ordination with low intelligence. In fact, intelligence is usually unaffected.

About one third of all children afflicted with cerebral palsy will have epileptic seizures.

The diagnosis of cerebral palsy is often suspected early in development as a result of postural difficulties, lack of muscle skills and weakness or stiffness of the muscles. Medical examination may reveal abnormalities in reflexes and loss or impairment of motor skills.

Normally referral is to a paediatrician who will examine the child and then undertake investigations to eliminate other disorders such as tumours of various types, blood vessel anomalies such as stroke, nerve palsies

affecting the arm(s), traumatic brain injury and muscular dystrophy (a genetic muscular disorder).

Investigations will include blood tests to eliminate thyroid disease (cretinism), chromosome studies and other chemical analyses to exclude metabolic (functional) diseases.

Also required will be ultrasound of the brain and CT or MRI brain scans. Other tests may involve testing nerve activity (evoked potentials) and brain electrical activity (EEG).

Diagnosis is usually made by the age of three but sometimes, and particularly with more subtle

abnormalities, it may be delayed until the child is four or five.

Once diagnosed the management of a child becomes complex and will involve a variety of health professionals including the paediatrician and the GP, but very importantly the musculo-skeletal therapist (physiotherapist or osteopath), occupational therapist and speech therapist.

The musculo-skeletal therapist will work to prevent the development of weakness in the muscles and to encourage the child to undertake exercises to avoid the development of contractures (fibrosis in the muscles which prevent them from working).

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Sometimes the child will be fitted with limb braces to prevent contraction and to add support.

The skills of occupational therapists may be invaluable in training children to be more self-sufficient and to undertake the daily tasks which most people find easy but with which a child with cerebral palsy may struggle (for example, tooth brushing or going to the toilet). Speech therapy may be extremely important to help the child to communicate well with those people around him or her.

The involvement of the family is crucial and good supportive parents and siblings are of considerable benefit. The whole approach to the child should be coordinated to ensure that maximum benefit is achieved and that the child's self-esteem is protected as much as possible.

Medication is valuable and may be essential. For those children with epilepsy, management with anticonvulsants may improve or eliminate the seizures.

Other medication may be used to counteract the stiffness and overactivity which muscles may demonstrate and which will cause sufferers inconvenience, annoyance and frustration. **Baclofen** is a drug which is effective at relieving muscle spasm.

It is usually given by mouth but can be administered into the nervous system through an implanted pump with an intrathecal catheter (a tube passing from the pump and carrying the drug through the covering of the brain and

spinal cord to act directly on the nervous system). The drug must be used with care because it may induce seizures and hyperthermia if withdrawn abruptly.

Diazepam (Valium) is an effective muscle relaxant but its use is limited by drowsiness, constipation and nausea.

It is normally administered by tablet. If the side effects of the diazepam limit its use, then other muscle relaxants are available. If however they are insufficiently effective or limited by side effects, **botulinum toxin** may be injected to block the nerves supplying affected muscles. The effect will last about three months.

A key complication of uncoordinated or excessive muscle spasm is skeletal distortion resulting in locomotor difficulties, posture and pain.

Surgery to correct distortion and deformity may improve the ability to move and thereby improve confidence. Better appearance through corrected stature may also raise self-esteem. During surgery muscles and tendons can also be lengthened to improve mobility further.

Cerebral palsy proves to be an enormous burden for many sufferers. The stresses on the body induced by the muscular asymmetry and distortion result in weakness, persistent tiredness, aching pain and early onset arthritis. Body organs such as the heart, lungs and circulatory system are under constant strain and can be displaying signs of failure by the age of 40-45.

For many CP patients, depression becomes a persistent problem triggered by the challenges of the condition which only those people with cerebral palsy can fully understand.

Yet for many people with the disorder, they remain happy and accept the challenges of their disability with fortitude and courage.

They can develop their intellect and intelligence to the full and many have enjoyed the physical challenges of sport and we have watched, spellbound, their successes in the recent Paralympic Games. Perhaps the miracles of gene therapy and pluri-potential (stem) cell recuperative techniques may herald a cure for this most debilitating of conditions. We live in hope.

I am most grateful to Dr David Phillips for reviewing this article and to his daughter Rachel, herself a victim of cerebral palsy, who copes with the problems it causes with courage and good grace while using her lively personality to the full to entertain and cheer those around her.

She is truly a testament to victory over adversity.

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