Neuromuscular conditions of childhood

J Mark H Paterson

Abstract

In this article a review is given of the pathology, clinical features and orthopaedic management of some of the more common neuromuscular conditions affecting children. The term 'neuromuscular' will be used to cover those conditions in which there are significant musculoskeletal consequences of neurological or muscular pathology, and include cerebral palsy, myelodysplasia (spina bifida) and other spinal anomalies, poliomyelitis, Duchenne muscular dystrophy, and hereditary motor and sensory neuropathy. Emphasis is placed on the principles of management rather than the technical details in recognition of the specialized nature of the treatments involved.

Keywords Cerebral palsy; children; neuromuscular; pes cavus; tendon transfer

Cerebral palsy

Cerebral palsy (CP) is the term given to a group of conditions in which a non-progressive brain lesion at or around birth gives rise to disorders of posture and movement. The lesion may be a discrete focus of haemorrhage or cyst formation, or it may be the result of more diffuse damage from ischaemia or infection. The premature brain is especially vulnerable to ischaemic damage, and babies born under 30 weeks' gestation are at high risk of ischaemic hypoxic encephalopathy. There is also a wide range of autosomal recessive inherited syndromes with neurodisability similar to that seen in CP. Overall, CP is relatively common, with an incidence of about 2 per 1000 live births.

As a result of the central lesion, interruption of the corticospinal tracts result in impaired control of the spinal reflex arc, which in turn gives rise peripherally to the characteristic movement disorder of spasticity. This is an exaggerated response to the normal stretch reflex. Spastic muscles resist stretch, which is essential for the normal growth and development of muscle. As a consequence, spastic muscle does not grow properly, and contractures and deformity can result (Figure 1). Other movement disorders include athetosis (uncontrolled writhing movements) and dystonia (variable posture). Unlike spasticity, which is a disturbance of pyramidal system function, these latter disorders involve extrapyramidal tracts. The outcome of surgical interventions in these disorders is much less predictable than that of spasticity.

A child with CP may have unilateral involvement as in stroke (hemiplegia), involvement predominantly of the lower limbs (diplegia), or global involvement. This latter used to be called quadriplegia, but nowadays we tend to refer to total body involvement (TBI), in recognition of the important effects on the axial skeleton in the form of scoliosis, and effects on the bulbar system involving swallowing and allied functions. However severe the physical disability it is important to remember that there may be little or no cognitive impairment.

The role of the orthopaedic surgeon in the management of CP varies according to the extent of neurological involvement. Children with hemiplegia and many with diplegia will be able to walk, and the emphasis here is on treating contractures, foot and upper limb deformities, and torsional deformity in the lower limbs. Children with TBI are at particular risk of progressive hip dislocations and scoliosis.

Hemiplegia: this results from a focal lesion in the brain such as a cyst or haemorrhage. The effects are seen on one side of the body in the form of spasticity, which may lead in turn to contractures and deformity. Some muscle groups are involved more than others, giving rise to the typical upper limb posture of adduction and internal rotation at the shoulder, flexion at the elbow, pronation in the forearm, and flexion at the wrist and fingers. In the lower limb there is typically equinus or equinovarus at the foot and ankle due to involvement of the calf muscles (Figure 2).

Diplegia: this is frequently the result of ischaemic or hypoxic damage to the immature brain and is seen in children who were born prematurely. The most obvious neurological abnormalities are seen in the lower limbs, with the upper limbs and axial skeleton relatively spared. Most of these children have some standing and walking ability, and surgical interventions are directed at maximizing their walking potential. Many of them will develop calf and hamstring contractures, and may have valgus feet and ankles.

Management of both hemiplegia and diplegia in the early years involves joint ranging and stretching exercises to maintain muscle length and encourage muscle growth. Serial casting is a technique which uses the stress-relaxation properties of muscle and tendon to achieve a gradual increase in muscle-tendon length by the application of plaster casts applied in maximum stretch. Children can be helped to adopt a normal gait sequence with the use of ankle-foot orthoses. Tone reduction can be achieved on a temporary basis by use of botulinum toxin, which interrupts motor endplate function and blocks the release of acetylcholine causing a reduction in muscle tone which in practice may last for 4–6 months. Spasticity may also be reduced by selective destruction of the dorsal spinal roots (selective dorsal rhizotomy or SDR) or by continuous intrathecal infusion of baclofen (ITB), a tone-reducing drug. These are both relatively recent developments, and the precise indications for their use in cerebral palsy have yet to be established. Ultimately it may be necessary to lengthen surgically the muscle-tendon unit (Figure 3). Surgery is best avoided in the early years as it interferes with muscle growth. Surgery may also unmask the underlying weakness of the involved muscles, and extensive releases in older heavier children may make it difficult for them to rehabilitate.

It is essential to consider the child as a whole when assessing difficulties with standing and walking. Because of the complex nature of human gait, and the fact that some muscles cross more

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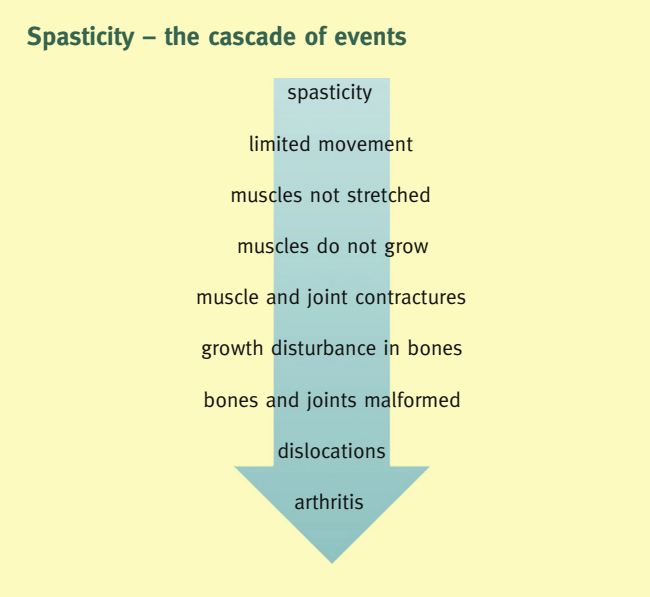

Figure 1 Sequence of problems in cerebral palsy.

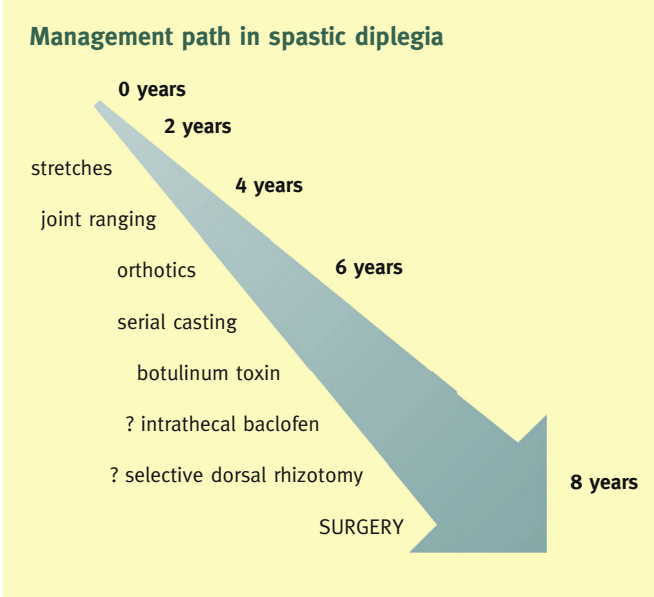

Figure 3

than one joint and have more than one action, a simple surgical intervention at one level may lead to problems elsewhere. Because of this, instrumented gait analysis has been developed in order to acquire objective gait data which is used to plan appropriate surgery (Figure 4). This may include lengthening a contracted gastrocnemius or hamstring, stabilizing a foot that has been pulled by abnormal muscle tone into a valgus position, or de-rotating a femur that has become internally rotated, again through prolonged unbalanced muscle action. In situations where there is relative overactivity of one muscle, an appropriate tendon transfer may be used to redress the balance. In general, such surgery should be performed in one sitting wherever possible, so that the whole limb can be properly balanced and the child has one rehabilitation period. This has led to the concept of single event multilevel surgery (SEMLS) in which both muscle and bone procedures at all levels in the lower limb may be performed at the same operation. This requires a heavy commitment from the patient, family and therapists, as the rehabilitation is long and hard.

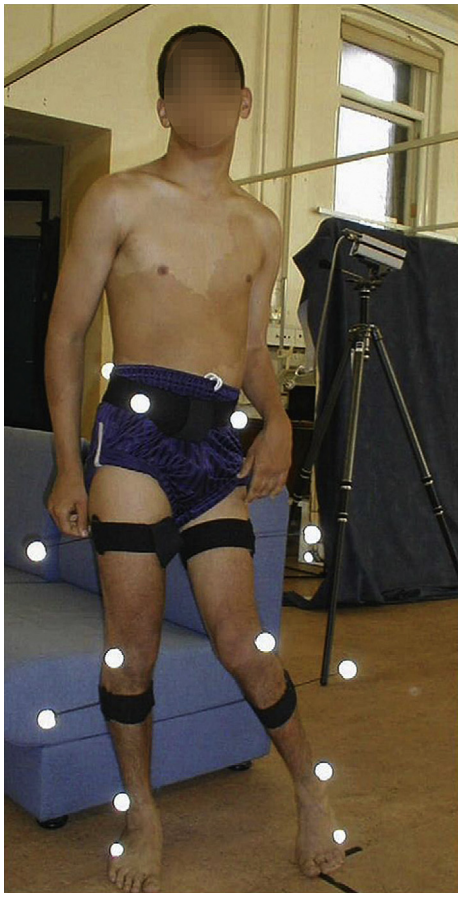

Figure 2 Teenager with right hemiplegia with typical posturing. Note reflective markers used for gait analysis.

Total body involvement: these children have a wide variety of severe brain abnormalities, including genetic and metabolic defects. They are generally not able to stand or walk independently and many will not have the ability to sit or crawl. Seizure disorders are common, and, as a result of bulbar dysfunction and swallowing problems, many also have feeding difficulties and chest problems with consequent malnourishment. These factors are of great importance when considering surgical intervention in such children.

The orthopaedic priorities are to prevent and treat progressive hip displacement and scoliosis. Hip reconstruction (Figure 5) and spinal correction are major procedures that carry a definite risk to the TBI child, who is often undernourished with a high risk of postoperative respiratory complications because of impaired swallowing and resulting aspiration. However, a dislocating hip can be a source of severe relentless pain, with associated spasm and deteriorating posture. Consequently the indications for and against surgery have to be weighed up very carefully with all those involved in caring for the child.