

46. ARTHRITIS AND MUSCULO-SKELETAL CONDITIONS IN CHILDREN

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	Related conditions considered in other chapters:	
	Spinal Injury	Chapter 18
46.2	Trauma	
46.2.1	<p>Trauma including fractures of bones and head injury is not uncommon in childhood. It may give rise to the need for increased attention (eg. when the child is in a plaster cast or receiving intensive physiotherapy at home). The period of increased attention is usually limited and, depending on the condition, is unlikely to last for more than three months. In those cases in which considerable attention needs persist because of the nature and severity of the fractures and resulting disability it is unlikely to be for longer than one year.</p>	
46.3	Arthritis	
46.3.1	<p>Juvenile chronic arthritis (Still's disease) is the most common form of arthritis in children. Pain, swelling and limitation of movements of affected joints are cardinal features. The number, site and degree of involvement of affected joints will determine the extent of disability and the needs it gives rise to. When the condition is in the acute stage the majority of cases will need extra attention. As the condition remits the attention may well cease to be needed. Once attention needs have become established, they will normally remain unchanged for about two years.</p>	
46.4	Congenital Dislocation of the Hip	
46.4.1	<p>Congenital dislocation of the hip is usually diagnosed, and treatment commenced, within the first year of life. Its management may be protracted, particularly if the diagnosis is delayed beyond early infancy, requiring more than one operation, each followed by periods of immobility and/or splinting of the hip. Except for children aged 1 year or less, extra attention over and above that of a healthy child will normally be required during the day. The periods of immobility and/or splinting vary between 1 and 12 months, interspersed with hospital admissions for adjustment of splints, replacement of casts, etc. Night attention is usually infrequent.</p>	

46.5 Perthes' Disease

46.5.1 This is a disease of the hip which affects children between the ages of 2 and 12 years. Most (80%) of the affected children are boys. Usually only one hip is affected in this condition. The initial treatment involves immobilization and traction (ie pulling on the leg with weights) of the painful hip. This does not usually last longer than a week. Thereafter the child is commonly fitted with a non-weight-bearing caliper or brace which protects the affected hip. The child fitted with such an appliance should be able to live a near normal life both at school and at home. The child will likely require assistance during the day with putting on and taking off the caliper; but this is unlikely to exceed a few minutes.

46.6 Spina Bifida

46.6.1 Clinical Description

- (i)** Spina bifida is a developmental abnormality. The neural tube (ie. that part of the developing baby which will give rise to the spine) fails to unite leaving a gap over which the skin is defective. In the severest form the baby is born with a protruding sac at the base of its spine containing either cerebrospinal fluid (meningocoele) or cerebrospinal fluid and nerve tissue (meningomyelocoele).
- (ii)** In this severe form the disablement includes paralysis of the lower limbs, with or without spasticity, and sensory loss which exposes the child to risk of damage to the skin and lack of control of bowel and bladder. The condition is often, though not invariably, associated with hydrocephalus (enlarged head due to increased content of cerebrospinal fluid) and with mental retardation.
- (iii)** There is also a very mild form of the condition called spina bifida occulta. Here there is only a minor defect in the spine in the low back region which is covered-over as normal with body tissues and skin. In general spina bifida occulta is an isolated, insignificant finding, but less commonly it can be associated with all the problems and complications accompanying the classic, open spina bifida.

46.6.2 Care Needs

In the severe forms of spina bifida substantial extra attention is required to care for the skin, prevent infection, attend to the bladder and bowel and train such functions as can be utilised to compensate for the disability. These needs will be required at night as well as during the day, and are unlikely to change until the child is at least 8 years of age - but will vary in individual cases depending on the severity of the lesion, any associated complications, and the needs posed by them.

46.6.3 Duration of Care Needs

- (i)** Some children with paralysis of the lower limbs may have been trained to

transfer to and from and to operate a wheelchair and to manage urinary apparatus, thus remaining clean and dry for most of the day. Despite this, the severity of the disablement usually requires a child to be supervised in these activities for most of the time. At night the needs posed by the disabilities are unlikely to require attention and/or watching-over, except during limited periods which follow corrective surgery or troublesome urinary infection. Some children with severe spina bifida will need turning at night to avoid pressure sores.

- (ii) In the absence of any associated mental retardation, a child may have achieved a degree of independence by the age of 16 years which may very substantially reduce the requirements for attention and/or supervision. At this age the needs are likely to be determined by similar factors to those described in the Chapter on spinal injury and paraplegia [Chapter 18].

46.6.4 Mobility Considerations

Children with total paralysis of the legs are unable to walk. Many affected children without total paralysis of the legs are nonetheless severely restricted in their walking abilities because of weakness in leg muscles. Walking problems which are present at 5 years of age are likely to persist into adult life.

46.7 Muscular Dystrophy

46.7.1 Introduction

There are several different types of muscular dystrophy, most of which are rare. All are progressive, hereditary disorders, in which muscle fibres degenerate and are replaced by fibrous tissue and fat, resulting in gradually increasing weakness. Only those muscular dystrophies most commonly encountered will be described in detail. Their effects depend on the muscles involved and on the rate of progress of the disease.

46.7.2 Duchenne Muscular Dystrophy

- (i) This is the commonest and most severe form of muscular dystrophy. In about two-thirds of affected individuals the condition is inherited through the mother, who is a carrier. In the remainder the condition is a result of a change in genetic structure, (mutation) in the egg (ovum). Only boys are affected by the disease though a few female carriers show some of the symptoms.
- (ii) The condition usually becomes apparent between one and four years old, with delay in walking, a clumsy gait, inability to run properly, frequent falls and difficulty managing stairs. Learning disability is a common accompaniment, but is usually mild to moderate. [See Chapter 35].
- (iii) Weakness of the leg and hip muscles gradually increases, and walking becomes

more difficult, despite the use of aids, until a wheelchair is necessary. This stage is usually reached in early adolescence. Slowly the arms become weaker, as do the muscles of the trunk, which may require a brace. Eventual involvement of chest and even heart muscle leads to recurrent chest infections, heart failure, and death, which may occur in the late teens or early twenties.

46.7.3 Becker Type

This is a variant of Duchenne, again affecting only boys. It is milder in form and later in onset, appearing usually from late childhood to early twenties. The pattern of weakness is somewhat similar but much slower in its progression so that often the affected person lives well into the thirties or forties and sometimes longer. Walking may be possible into the thirties, or even to middle age. Heart muscle is not involved, and many people, though disabled, have a normal life span.

46.7.4 Limb Girdle Dystrophy

This is similar to Becker in patterns and severity, but affects both sexes. It usually appears in the second decade of life, often in adolescence, but sometimes later, and occasionally not until middle life. Weakness may begin in the muscles of either the hips or the shoulders, but eventually both groups are involved. The disease progresses more quickly when the legs are affected first, but most people are severely disabled within twenty years of onset, and many may die early.

46.7.5 Facioscapulohumeral Dystrophy

This is generally a milder disorder than those already mentioned, and affects both sexes. Onset may be at any age from childhood to adult life, but comes on most commonly during adolescence. The muscles of the face and shoulders are most affected, causing increasing difficulty in lifting the arms, weakness of eye closure, lack of facial expression and poor pronunciation with nasal speech. Progress varies and so does the degree of disability, but most people with this form of muscular dystrophy remain active.

46.7.6 Myotonic Dystrophy

- (i) This differs from other forms in being a multi-system disorder. The muscular problem may be accompanied by others involving the eyes, heart, lungs, hormone and immune systems, and the brain. As well as weakness, there is difficulty in relaxing muscles after voluntary effort.
- (ii) It affects both sexes and is commonly found in adults, developing between the ages of twenty and fifty, though it may occasionally arise in childhood or even

at birth (in children of affected women) when it tends to be very severe and associated with learning disabilities. The first symptoms are usually weakness of the hands and difficulty with walking. Poor vision, weight loss, increased sweating and drowsiness may develop, and there may be dulling of the intellect or dementia. Progression is slow, severe weakness occurring only at a late stage. Few people are confined to a wheelchair before their fifties, but the presence of cardiac and respiratory symptoms with a slow deterioration in mental and physical energy may add to the level of disability. For most people this is severe within fifteen to twenty years of onset, and death from respiratory or cardiac failure may occur early.

46.7.7 Spinal Muscular Atrophy

- (i) Although not a dystrophy, this condition may at first resemble one, and it causes similar problems. It is the childhood version of Motor Neurone Disease [See Chapter 15], and has three forms, the infantile, the intermediate and the juvenile, affecting both sexes.
- (ii) The infantile form (Werdnig-Hoffman disease) is usually apparent soon after birth. There is severe weakness of all muscles, except those of the face. Most of these babies die in infancy, but for the few who survive longer, complete helplessness persists.
- (iii) "Intermediate" spinal muscular atrophy is a severe, chronic, generalised form of the disease which normally presents in the first year of life with severe muscle weakness and skeletal deformities. Despite the fact that they have severe weakness and physical disability, these children are usually intellectually normal.
- (iv) The juvenile form (Kugelberg-Welander Syndrome) is much milder, and occurs between early childhood and adolescence. It usually begins with difficulty in walking, climbing stairs and rising after a fall. Progress is spasmodic, periods of deterioration being interspersed with periods when it seems to be arrested, and the ability to walk is frequently retained into adult life. Gradually increasing disability is, however, inevitable and survival limited to some thirty years or so from onset.

46.7.8 Care Needs and Mobility Considerations

- (i) Because of the range of disabilities caused by these conditions it is impossible to generalise. Needs arise as muscle groups degenerate, and the type and level of need in any individual will depend on which muscles are involved and what stage has been reached in the disease process. Either care or mobility needs may be the first to arise and, in the more severe forms of these conditions, both may be extensive.
- (ii) It is important that these people are kept under regular review by medical, orthopaedic and physiotherapy agencies. It is essential to maintain good

posture, mobility either independently or with aids, and as good a quality of life as possible.

46.7.9 Duration of Needs

All these conditions are progressive, and cause gradually increasing disability. Needs, once established, do not abate. The level of need will increase as the disease progresses.

46.7.10 Further Evidence

In the case of a child, the most valuable source is likely to be a professional involved in community care, either at a Child Development Centre or in school; for adults, it may be a social worker or GP.

46.8 Osteogenesis Imperfecta

46.8.1 Introduction

The term osteogenesis imperfecta (brittle bone disease) refers to a group of rare inherited disorders with the common feature that bones are excessively fragile. The overall incidence of osteogenesis imperfecta is between 1 per 20,000 and 1 per 50,000 of the population. It takes several different forms and varies considerably in its severity, although the commonest form is usually relatively mild.

46.8.2 General

- (i)** In the mild forms the tendency of bones to fracture is not great and any fractures that do occur heal well leaving little or no disability. This group may have little problem in childhood, fractures occurring later in life. This form of the condition is often associated with deafness and with damage to the enamel of teeth.

- (ii)** The more severe cases suffer repeated fractures from minor injuries. They may develop severe deformities because the fractures do not heal properly. This often leads to the need for repeated operations to correct the deformities. It may even be necessary to support the skeleton externally with inflatable "space suits". An alternative approach is to accept that a severely affected child will never walk and that the provision of a suitable electric wheelchair is the best means of increasing independence and mobility.

- (iii)** The most severely affected children are born with multiple fractures and gross deformity and shortening of limbs due to these. Most die at or shortly after birth but a few survive infancy grossly disabled and unable to do anything without the risk of further fractures occurring.

46.8.3 Care Needs

These will depend on the severity of the condition. In most instances all that is needed is some extra supervision in situations when the possibility of falls is greater than usual. This extra supervision varies with individual circumstances and depends to some extent on the age of the child. In the more severely affected child, in particular those with a history of repeated fractures, there is likely to be a need for considerable additional supervision for most of childhood. If there is evidence of deformity and shortening of bones there may also be a need for help in connection with bodily functions. Those with the most severe form of the disease that survive infancy will be in need of a great deal of attention both by day and by night.

46.8.4 Mobility Considerations

These again will depend on the severity of the condition. For those with the mildest form of the disease there may be some need for supervision when walking, but in most instances this is unlikely to be significantly greater than that needed by a healthy child of the same age. For those who do need supervision to avoid the danger of falls, there may be a need for supervision when walking. There may also be a danger to health caused by the effort of walking as the "stresses" of walking may alone be sufficient to cause fractures. Those affected to this degree may well be using a wheelchair already since the severity of their disability results in their being unable to walk.

46.8.5 Duration of Need

In less severe cases where it is established that a young child needs a significant degree of supervision, this may be required to about the age of 8. By this age the child may be aware of the dangers and may be able to take appropriate action to avoid them. Each case, however, must be considered individually. In the more severe cases, particularly those with considerable care needs or those unable to walk, the disability will be permanent.

46.8.6 Further Evidence

The needs of those affected severely should be clear. In less severe cases a report from the GP or hospital attended may well provide details of the frequency and severity of previous fractures and their consequences. This information should give an indication of the likely risks in future.