

Congenital and developmental conditions

Developmental dysplasia of the hip

The term congenital dislocation of the hip (CDH) has been replaced by developmental dysplasia of the hip (DDH) because the latter is more representative and encompasses the spectrum of abnormalities seen in this condition.

Causes

The cause of the condition is unknown. Girls are affected more than boys (8: 1). It may be related to:

1. Hereditary factors. The risk is increased to 36% if one parent had DDH. Familial joint laxity may also be a factor in some cases.
2. Environmental factors. In those societies where it is usual to nurse the child with the legs bound together in adduction (swaddle), the incidence of established DDH is much higher than in those societies where the child is carried on the mother's back with the legs widely abducted legs.
3. Position in utero. Breech delivery is more commonly associated with DDH.

Diagnosis At birth



Clinical examination is crucial. Ortolani and Barlow's manoeuvres are carried out.

Barlow's test for hip instability

Barlow's test showing full abduction



Radiograph of late developmental dysplasia of the hip



Later in life

The leg may be short and abduction of the hip limited. The femoral head may be felt to move in and out of the joint in abduction, or may fail to reduce and be palpable in the buttock. Most late cases are spotted when the child walks. The child usually walks with a characteristic waddle (Trendelenburg gait) due to inefficiency of the abductor muscles.

Treatment

The principles of treatment are to reduce the hip and maintain reduction; this is the same irrespective of the age of the child. The older the child when the diagnosis is made, the more difficult it is to achieve the treatment goals.

Newborns and infants less than 6 months of age:

Treatment must begin as soon as the diagnosis is made (after birth). Splinting the limb in abduction usually allows reduction and stabilization of the hip. The Pavlik harness is the most commonly used device in the newborn period but others, such as the Von Rosen splint and Frejka pillow, are available. The Pavlik harness may be used until the age of 6 months.



Pavlik harness

Children 6 months to 2 years of age:

In children older than 6 months at diagnosis and those who have failed a trial of Pavlik harness, closed reduction is indicated. This procedure is carried out under general anaesthesia and in the operating room. If closed treatment fails to achieve reduction, open reduction is indicated.

Children older than 2 years of age

Open reduction is usually necessary if the diagnosis of DDH is made in children aged 2 years or older.

Congenital talipes equinovarus (club foot)

Congenital talipes equinovarus is characterized by a complex malalignment of the bones and joints of the foot and ankle. The deformities are equinus and inversion of the hindfoot, cavus (plantar flexion of the forefoot on the hindfoot) and adduction of the forefoot on the midfoot so that the sole of the foot points medially or even upwards.



Talipes equinovarus

Cause:

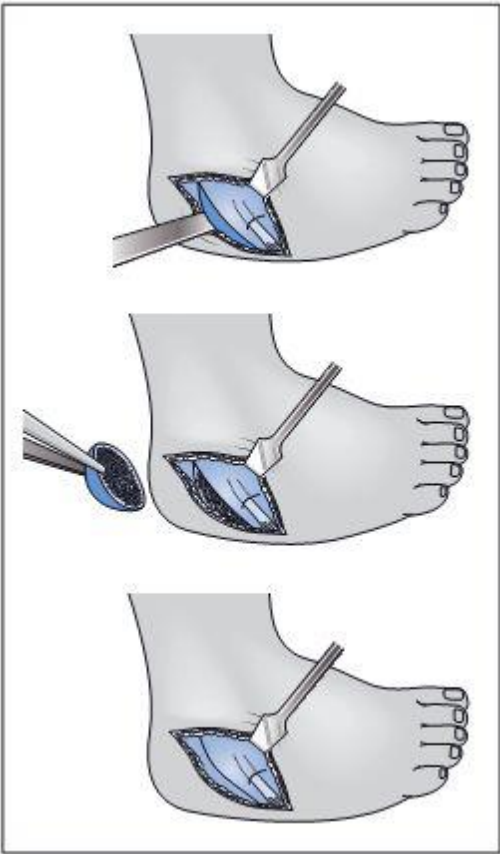
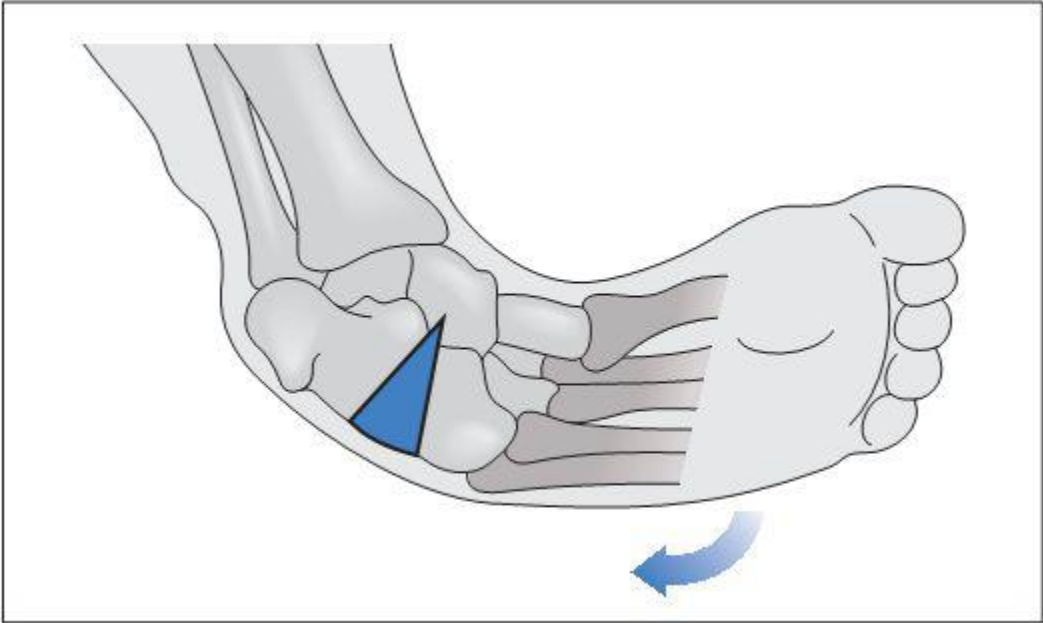
The cause is unknown but various theories have been proposed and these include primary muscle, vascular or neurological abnormalities and/or in utero molding. Club feet may be idiopathic, postural, neurogenic or syndrome related. It is important to examine the child fully to exclude other associated anomalies.

Treatment:

Treatment should begin as soon as the diagnosis is made and the goal is to achieve a plant grade, painless foot. The technique of manipulation and serial casting popularized by Ponsetti *et al* is the gold standard.

Late recurrence may occur up to the age of 4 or even later, and it may then become necessary to correct the residual deformity by operations, such as that designed by Evans, in which the calcaneo-cuboid joint is excised to produce a lateral fusion. This partly corrects the deformity and relies on further growth on the inner side to complete the correction. The operation devised by Dwyer concentrates on taking a wedge from the outer side of the os calcis to correct the inversion of the heel. He believes that growth corrects the remaining deformity.

Evans operation for relapsed club foot



Dwyer operation for relapsed club foot

Cerebral palsy

This is a disorder of movement and posture resulting from injury to the immature brain. The etiology may be intrauterine developmental defects, birth trauma, asphyxia and diseases or injuries in early life.

Cerebral palsy is divided into various types based on:

1. **Physiology** (type of movement disorder).
2. **Topography** (geographical distribution).

Physiological classification

1. **Spasticity**—this is an upper motor neurone type defect and is characterized by increased muscle tone and reflexes. This is the common form of cerebral palsy.
2. **Ataxia**—this is characterized by an inability to co-ordinate muscles for voluntary movement.
3. **Athetosis**—in this type, the limbs move at random, with jerking and uncoordinated movements.

Topographic classification

This classification is based on the degree of limb involvement.

1. **Hemiplegia**—the upper and lower limbs on the same side are involved.
2. **Diplegia**—it means involvement of both lower limbs.
3. **Quadriplegia**—all four limbs are involved.



Typical posture of a child with hemiparesis



Spastic quadriplegia with
"scissoring"

Management

The first step in the management of a child with cerebral palsy is a thorough history and clinical examination. The child's balance, sitting and gait must also be assessed. Gait analysis is helpful in assessing movement disorder in those children who can walk. Treatment must be goal-oriented and may be surgical or non-surgical. Available treatments include physiotherapy, use of orthoses (splints), intramuscular botulinum toxin injection, oral or intradermal baclofen and surgery.

Surgery:

This involves soft tissues and or bony surgery. The aims of surgery are:

1. To correct any established deformity.
2. To restore muscle balance and diminish spasticity.
3. Train the child in posture and movement.
4. Provide suitable sensory stimulation.