Diagnosis and management of developmental hip dysplasia

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Abstract

Congenital dislocation of the hip represents a spectrum of disease from transient neonatal hip instability to established dislocation. Known risk factors include breech presentation and a positive family history. Emphasis should be placed on early diagnosis and treatment to minimise the need for surgical intervention. Clinical assessment has limited accuracy which further diminishes soon after birth. Ultrasound based screening is more sensitive, but controversy remains as to which method to use and its effect on outcome. When initiated early, treatment using a simple abduction device such as a Pavlik harness has an excellent success rate. Resistant cases and late presentations usually require surgical management, which are largely age dependent. We discuss the condition, detail the screening protocol in our institution and outline the surgical options in this article.

Keywords Developmental dysplasia of the hip (DDH); open reduction; Pavlik harness; pelvic osteotomy; screening; ultrasound

Definition

The term congenital dislocation of the hip (CDH) describes a spectrum of abnormalities of the hip joint, from capsular laxity or mild acetabular dysplasia to established dislocation. Secondary hip dysplasia may occur in the absence of persistent instability in the growing hip, hence the more recent term developmental dysplasia of the hip (DDH).

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Epidemiology

The incidence of neonatal hip instability is up to 20 per 1000 live births; this varies according to diagnostic criteria, but most resolve during the first few weeks of life. The incidence of established dislocation in an untreated population is 1–2 per 1000 births, but there is considerable geographic variation (see Figure 1).

The condition is five times more frequent in girls, and the hormone relaxin has been implicated. First born babies are more often affected; instability is more common in the left hip, probably reflecting the most common intrauterine position. It occurs bilaterally in up to 20% of patients. Proven risk factors include breech positioning, particularly frank breech presentation at vaginal delivery, and a positive family history. Other established associations include congenital muscular torticollis, neonatal foot abnormalities, oligohydramnios and also postnatal positioning such as swaddling. Most cases, however, have no identifiable predisposing factor.

Pathophysiology

At birth, the neonatal hip is cartilaginous. The acetabulum has a hemispherical hyaline epiphysis with a thin fibrocartilaginous labral rim; the proximal femur has a complex growth pattern with the femoral head ossification centre appearing between four and seven months of age.

Hip instability will compromise the patency of the posterosuperior acetabulum in particular, where locally cartilage thickening produces a ridge termed the neolimbus. Later, hypertrophy is seen in the fatty tissue (pulvinar), the ligamentum teres and the transverse acetabular ligament; ultimately hourglass deformation of the inferior hip capsule occurs and impedes later reduction. Delayed ossification is evident in the femoral head. Contractures develop early in the presence of dislocation, particularly in the adductors and iliopsoas. While these early changes are confined to the soft tissues, persistent dislocation causes abnormal acetabular development, with reduced depth to width ratio, and progressive obliquity; inadequate cover of the femoral head allows progressive elevation and lateralisation.

Natural history

In established dislocation, the femoral head usually articulates with the ilium at a false acetabulum, and secondary degenerative changes inevitably occur over time. Higher dislocations without a false acetabulum, particularly if bilateral, will retain good range of motion and late onset of low back pain due to hyperlordosis. The outcome of untreated mild and moderate hip dysplasia, however, is confounded by a lack of defined long term studies. Although most cases of osteoarthritis have no underlying abnormality, dysplasia is probably the most common underlying identifiable cause in patients undergoing hip arthroplasty. Symptomatic degenerative disease is seen in up to half of patients by the end of the fifth decade.

Clinical findings

The prevention of late hip disease depends on the diagnosis of hip instability and early treatment; detection is still a matter of debate. Most clinical examinations are performed as part of a

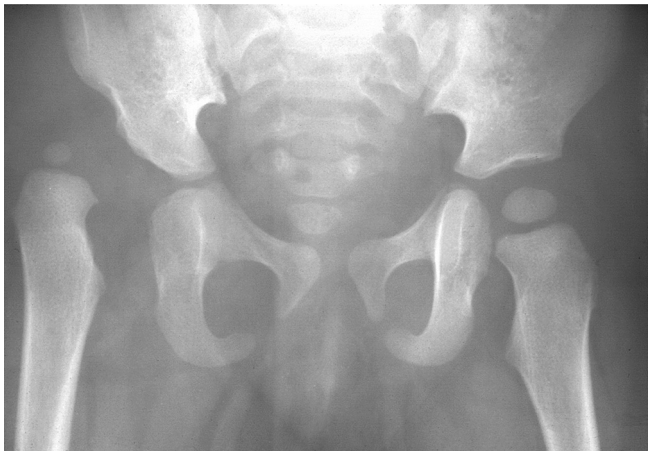

Figure 1 Established dislocation.

routine assessment, although some parents will notice a clicking hip during changing, which is usually benign. Clinical examination incorporates two established manoeuvres. The Barlow test is provocative, attempting to identify an unstable hip by gentle posterior force; the Ortolani test attempts to reduce the dislocated hip, a clunk being appreciable as the femoral head passes the neolimbus. The importance of these tests diminishes soon after birth, when reduction of hip abduction becomes the most sensitive clinical finding; the signs of emerging leg length discrepancy are subtle, and are not present in bilateral cases. Most children with established dislocation will walk at a normal time, and older children are usually referred with a limp. Recently the relationship between incorrect swaddling methods and development of dysplasia has been identified. Tight wrapping of the legs in extension may cause subluxation and dislocation of the hips. Healthy hip swaddling technique allowing the infant's legs to have room for correct positioning, is advocated by international experts. Further details and information for parents can be found at http://hipdysplasia.org/developmental-dysplasia-of-the-hip/hip-healthy-swaddling/.

Screening

Although a delay in the diagnosis of hip dysplasia progressively increases the likelihood of surgical intervention, neonatal screening remains controversial. Clinical examination is dependent on the examiner and has limited accuracy, with a sensitivity of probably less than 60%. Ultrasonography as an adjunct to clinical examination was introduced in Austria by Graf, with classification by angular measurement of static angles dictating subsequent treatment. The incorporation of real time evaluation of stability during a modified Barlow test defines the dynamic method, which may have a better correlation with clinical outcome. Timing is a critical factor, as untreated instability at birth will resolve in 60% of patients within one week and 90% by nine weeks; it is not possible to determine which hips will normalise. Universal screening has been employed in Austria and Germany and several studies describe improved outcome. However its benefits over a selective screening programme, limited to neonates with risk factors, have been questioned. Selective screening has been advocated in the UK and is currently undertaken on a regional basis. In the US, recommendations are at variance. Critics of screening argue that it does not influence the rate of late dislocation or of surgical intervention. A recent meta-analysis concluded that there was insufficient evidence to recommend how to screen. The protocol in Southampton is to sonographically examine clinically abnormal hips at 2 weeks to allow for transient instability; other referrals undergo examination at 6 weeks.

Investigations

Radiology for hip dysplasia is only useful after 4 months, and several methods have been described. The most utilised are the acetabular index, which changes with age, and the centre-head distance discrepancy (CHDD). The height of dislocation can be estimated using the method described by Tonnis. In the older child ultrasound is less efficient due to the morphology of the hip. A new classification for the assessment of hip dysplasia has been validated by the International Hip Dysplasia Institute (IHDI), which grades severity of dysplasia without evidence of the ossific nucleus on radiograph. For later dysplasia the centre-edge angle of Wiberg can be used in older children, and the Severin classification is used to quantify outcome of dysplasia, albeit with poor levels of reliability. Arthrography remains a useful adjunct in the assessment of stability and adequacy of reduction.

Management

The management of hip dysplasia is age dependent, and can be divided into neonatal, early and late groups.

Neonatal hip instability is ideally diagnosed early and treated appropriately. The use of an abduction device allows maintenance of hip location, and the Pavlik harness has become established ahead of rigid splints (see Figure 2). The harness dynamically maintains the hip at about 100° of flexion and 60° of abduction, a favourable position in preventing avascular necrosis of the femoral head due to compression of its tenuous blood supply. Ability to reduce the hip is a prerequisite using the Pavlik harness, and ultrasonography is effective in monitoring the progress of treatment. The device is contraindicated in neurological dislocation. The success rate of harness treatment is 90–95%, and the infant can be weaned from the harness after six weeks of continuous relocation. A dedicated clinic incorporating fitting, monitoring and patient support assists compliance. Complications are also addressed early, in particular avascular necrosis and femoral nerve palsy due to hip hyperflexion. Ultrasound detection of hips not responding within three weeks allows discontinuation of treatment and later assessment under anaesthesia. Serial radiological follow-up should monitor the progression of the acetabular index, which usually normalises after successful treatment by 18 months; we recommend review until 5 years, when centre-edge angle can be measured and late abnormalities in physeal closure can be recognised.

Early management between 3 and 12 months includes cases that have failed Pavlik harness treatment, or have been irreducible. This may occur in the presence of capsular constriction, labral interposition or contracture of the abductors or iliopsoas. This group also includes late presentations. Surgical reduction is now indicated, as there is a correlation between residual dysplasia and the age at reduction. Some authors argue that