Developmental dysplasia of the hip

Abstract

Developmental dysplasia of the hip encompasses a range of hip abnormalities in which the femoral head and acetabulum fail to develop and articulate anatomically. Developmental dysplasia of the hip is a clinically important condition, with a prevalence of 1–2/1000 in unscreened populations and 5–30/1000 in clinically screened populations. The pathology is incongruence between the femoral head and the acetabulum, which can be caused by an abnormally shaped femoral head, acetabulum, or both. This results in a spectrum of different hip abnormalities.

The precise aetiology behind developmental dysplasia of the hip is unclear, but there are a number of established risk factors. In the UK, universal clinical examination of newborns and 6–8-week-old babies is performed under the national UK newborn screening programme for developmental dysplasia of the hip (part of the Newborn and Infant Physical Examination). The physical examination of the newborn hip involves initial inspection of the infant for any of the clinical features of developmental dysplasia of the hip, followed by hip stability tests (Barlow's and Ortolani's tests). Hip ultrasound is the gold standard diagnostic and monitoring tool for developmental dysplasia of the hip in newborns and infants under 6 months of age, or until ossification of the femoral head.

Some mild cases of developmental dysplasia of the hip (and the immature hip) resolve without requiring intervention; however, there are a number of treatments, both non-operative and operative, that may be used at various stages of this condition.

Key words: Developmental dysplasia of hip; Hip abnormality; Hip development; Paediatric orthopaedics

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Introduction

Developmental dysplasia of the hip encompasses a range of hip abnormalities where the femoral head and acetabulum fail to develop and articulate anatomically. Developmental dysplasia of the hip was previously thought to be a congenital condition, but is now known to be a dynamic developmental condition, which may deteriorate or improve over time (Klisic, 1989).

Developmental dysplasia of the hip is clinically important, with a prevalence of 1–2/1000 in unscreened populations and 5–30/1000 in clinically screened populations (Sewell et al, 2009). It causes significant, long-term sequelae if not appropriately managed, and complications include gait abnormality, early degenerative change and pain. Developmental dysplasia of the hip is the reason for 29% of primary hip replacements in patients under 60 years of age (Sewell et al, 2009). These complications can be reduced if the condition is identified and managed early. The established clinical significance and complications of developmental dysplasia of the hip, alongside reliable screening tests and effective early treatment, has driven its inclusion in the Newborn and Infant Physical Examination national screening programme (Public Health England, 2018). Some centres, and in fact countries, have adopted universal screening programmes or population screening, with improved detection rate, reduced operative management and reduced costs for medical services (Clegg et al, 1999; Eastwood, 2003).

A literature search was performed using PubMed and Cochrane Reviews with the terms 'developmental hip dysplasia', 'DDH' and 'congenital hip dislocation'.

Pathology of developmental dysplasia of the hip

Normally, the femoral head and acetabulum fit each other anatomically, in a stable and congruent ball-and-socket joint. However, in developmental dysplasia of the hip, the

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femoral head and acetabulum are incongruent as a result of a mismatch to shallowness of the acetabulum in varying degrees. This results in a spectrum of different hip abnormalities:

- A stable hip joint with a dysplastic acetabulum of varying degrees
- A subluxed hip joint – the femoral head is, intermittently or permanently, only partially in contact with the acetabulum
- A dislocatable hip joint – the femoral head is intermittently fully not in contact with acetabulum, which can be elicited on clinical examination (Barlow's test positive)
- A dislocated hip joint – no contact between the femoral head and acetabulum (Ortolani's test is positive, if reducible)
- A teratology hip, which is dislocated in utero and irreducible on examination. This presents with a pseudoacetabulum and is associated with neuromuscular conditions and genetic disorders such as Ehlers–Danlos syndrome, arthrogryposis, myelomeningocele and Larsen's syndrome
- Late (adolescent) dysplasia, with a hip that is reduced and mechanically stable but dysplastic.

Aetiology

The precise aetiology is unclear, but there are a number of established risk factors that predispose babies to developmental dysplasia of the hip. These include breech presentation, female sex, a positive family history of developmental dysplasia of the hip, and first or second pregnancy (de Hundt et al, 2012; Woodacre et al, 2016). There is also mixed evidence in the literature regarding other risk factors – including high birth weight, prematurity, multiple pregnancy, mode of delivery, clubfoot deformity, packaging disorders and environmental factors (Chan et al, 1997; Perry et al, 2010; Sezer et al, 2013; Rhodes and Clarke, 2014; Pollet et al, 2017). The UK national screening programme uses two key risk factors to stratify risk in infants:

1. First degree family history
2. Breech presentation at or after 36 completed weeks of pregnancy (Public Health England, 2018).

These risk factors have led to two theories about the aetiology of developmental dysplasia of the hip (Woodacre et al, 2016). First, that developmental dysplasia of the hip is a 'packaging' problem. This theory suggests that breech presentation, high birth weight, late gestational age and the primiparous womb creates a constrictive intra-uterine environment that leads to malposition and maldevelopment of the hips. Second, that developmental dysplasia of the hip is hereditary. This theory suggests that the shape of the acetabulum and femoral head, and ligamentous laxity, are inherited factors. This is supported by the increased risk of developmental dysplasia of the hip with a positive family history.

Clinical presentation

Clinically, patients with developmental dysplasia of the hip may present with the following features and at varying age:

- Asymptomatic
- Asymmetry of skin folds (groin and buttocks)
- Asymmetry in abduction of hips during nappy changes
- Unequal leg length
- Abnormal gait in the weightbearing child
- Pain (skeletally mature).

Assessment

Universal clinical screening of developmental dysplasia of the hip was introduced in Europe throughout the 1950s and 1960s, with some countries also introducing universal ultrasound screening for developmental dysplasia of the hip (Olsen et al, 2018). In the UK, universal clinical examination of newborns and 6–8-week-old babies is performed under the national UK newborn screening programme of developmental dysplasia of the hip (Figure 1).

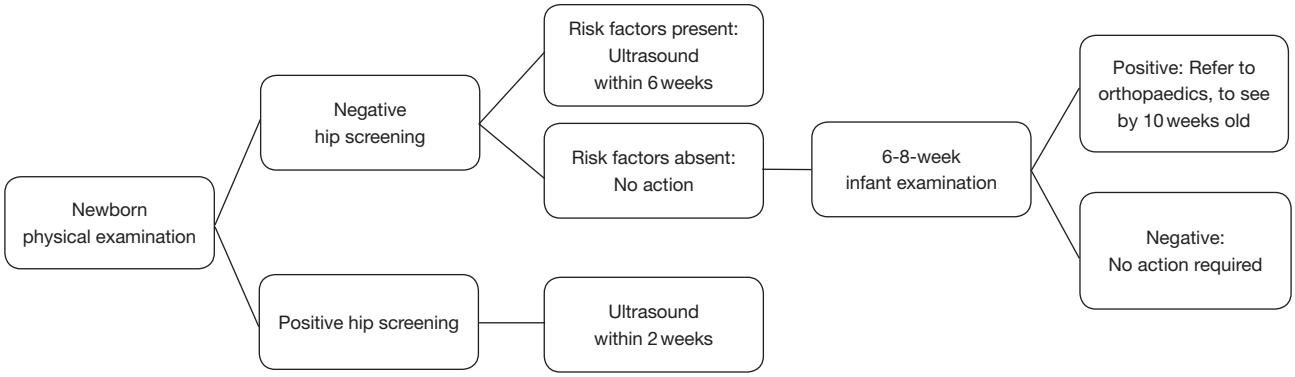

Figure 1. Examination and investigation of the newborn hip according to the UK National Screening Programme. From Public Health England (2018).

Examination

Physical examination of the newborn hip involves initial inspection of the infant for any of the clinical features of developmental dysplasia of the hip, followed by Barlow's and Ortolani's tests (Dezateaux and Rosendahl, 2007).

Barlow's test is used to assess for a dislocatable hip, where pressure causes an enlocated hip to dislocate posteriorly. The baby's hips are flexed to 90° in adduction, and posterior pressure is applied to the infant's flexed knees. The test is Barlow's positive if the hip dislocates, often without a palpable or audible clunk.

Ortolani's test is used to reduce a dislocated hip back into the joint. The hips are flexed to 90° in abduction and pressure is applied to the hips in an anteromedial direction, with fingers on the greater trochanter. The test is Ortolani's positive if the hip relocates and there is usually an audible clunk.

The specificity and sensitivity of these clinical examinations are highly dependent on the examiner (Committee on Quality Improvement, 2000; Shorter et al, 2011). A number of studies have investigated how reliable these examinations are at identifying true developmental dysplasia of the hip; the experience and training of the examiner has an impact on the sensitivity and positive predictive value of the test (Finne et al, 2008; Sulaiman et al, 2011).

Other signs are a positive Galleazzi test, presenting as apparent limb length discrepancy with hips flexed to 90° and the feet on table, and is the result of a unilaterally dislocated hip. The femur appears shortened on the affected side (Noordin et al, 2010).

In children above 3 months of age, limitations in hip motion are more obvious as contractions begin to occur, with leg length discrepancy predominating. In children over the age of 1 year who are walking, findings include pelvic obliquity, lumbar lordosis in response to hip contractions in bilateral dislocations, a Trendelenburg gait as a result of abductor insufficiency, and toe walking to compensate for shortness on the unilateral affected side.

Following clinical examination, under the UK newborn screening programme, children who test positive or have pertinent risk factors for developmental dysplasia of the hip are referred for an ultrasound (Public Health England, 2018).

Investigation

Hip ultrasound is the gold standard diagnostic and monitoring tool for developmental dysplasia of the hip in newborns and infants under 6 months of age, and has been adopted as a routine screening method for developmental dysplasia of the hip. On hip ultrasound, the hip joint is typically evaluated in two main ways (Omeroglu, 2014). First, by the Graf method, which is a static image of the hip with quantitative measurements (Graf, 1984). This involves the identification of key anatomical points, including the femoral head, hip joint capsule and labrum, acetabular cartilage and the bony roof of the acetabulum, followed by the measurement of angles between these landmarks and classification into subtypes depending mainly on the alpha angle (with the beta angle contributing to the severity)

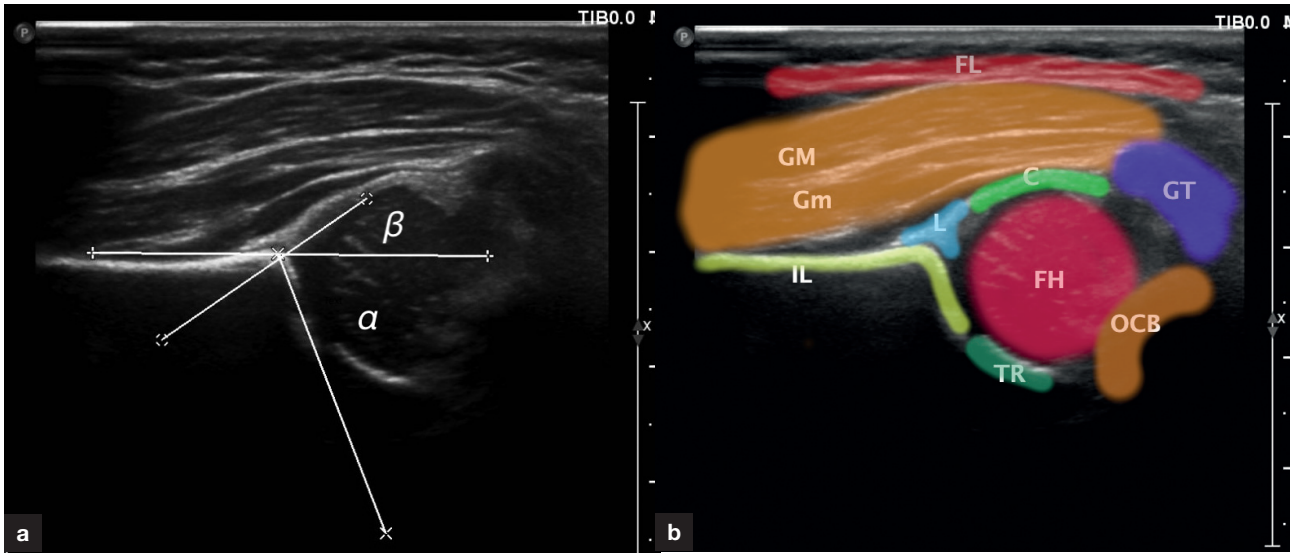

Figure 2. a. An ultrasound scan detailing the measured alpha and beta angles. b. The different structures identified: C = capsule; FH = femoral head; GM = gluteus medius; Gm = gluteus minimus; GT = greater trochanter; IL = ileum; L = labrum; OB = osteochondral border; TR = triradiate cartilage.

(Figure 2). Second, by the Harcke method – a dynamic image of the hip with qualitative evaluation of joint movement (Harcke et al, 1985). This allows the clinician to assess the position, stability and morphology of the hip joint (Omeroglu, 2014).

Radiographic imaging becomes more widely used in patients beyond 6–9 months of age, following significant ossification of the femoral head. This is classically an anteroposterior pelvic radiograph, which is evaluated for displacement of the femoral ossific nucleus, and deficiency of the acetabulum depth and contour, and allows for measurement of the acetabular index (Grissom et al, 2008).

Management

Non-operative

A number of cases of developmental dysplasia of the hip resolve without requiring intervention and develop into a normal hip (Bialik et al, 1999). An early study by Barlow (1963) found that 62% of infants diagnosed with developmental dysplasia of the hip on positive examination recovered spontaneously in 1 week, and 88% recover spontaneously in the first 2 months. This has led to suggestions that intervention could begin after 30 days of age, to allow as much as possible for the natural correction of hip dysplasia (usually coined as the immature hip) (Larson et al, 2017).

Pavlik harness and abduction braces

These come in various forms, such as the Von Rosen splint and Frejka pillow. The Pavlik harness, which was first described in 1944 (Mubarak and Bialik, 2003), is the most commonly accepted and used abduction splint or brace method for patients with developmental dysplasia of the hip diagnosed at less than 6 months of age, with reducible hips. It is contraindicated in teratological hip dislocations and in patients with spina bifida or spasticity. It otherwise is a very well-tolerated dynamic splint that promotes early spontaneous reduction of developmental dysplasia of the hip. It acts by holding the baby's hips in flexion and abduction with straps, while allowing free movement of the legs within the constraints of the splint. Various protocols exist, most commonly the baby wears the Pavlik harness for 24 hours, or for 23 hours with 1 hour off each day (Hines et al, 2017). The duration of treatment varies, but is an average of 6–12 weeks (Atalar et al, 2007). Regular clinical and ultrasonographic monitoring is performed throughout the duration of wearing a Pavlik harness and treatment is stopped at 3–4 weeks if it appears to have failed. The Pavlik harness has a success rate of 90–96.8% (Uçar et al, 2004; Choudry and Paton, 2017). Severity of developmental dysplasia of the hip on initial clinical examination

was found to be the most powerful predictor of treatment success (Lerman et al, 2001). However, the Pavlik harness can lead to rare complications, including avascular necrosis of the femoral head, temporary femoral nerve palsy, pressure ulcers and parental anxiety (Shorter et al, 2011).

Hip arthrogram with closed reduction and spica casting (with or without adductor tenotomy)

This treatment is used in patients between 3 and 18 months of age, or following failure of Pavlik harness treatment. The hip is reduced under general anaesthesia and successful reduction is confirmed by an arthrogram, with note being made of the dye pooling in the medial acetabulum and evidence of interposition of the limbus (Khoshhal et al, 2005). The hip is then held in a spica cast in flexion and abduction in the safe zone (abduction 40–60° and flexion 100–110°). If there is evidence of tight adductor tendon or restriction in abduction then an open or percutaneous adductor tenotomy is performed. Postoperative imaging can include a pelvic X-ray if the dye remains, or more commonly in specialist centres magnetic resonance imaging performed on the patient from recovery, or a low dose focused computed tomography scan. The spica cast is usually changed at 6 weeks and reimaging is performed, to confirm successful treatment. The cast can stay in place from 12 weeks to 6 months dependent on age and progress. Caution is needed as extreme abduction is associated with increased risk of avascular necrosis, and pressure ulcers can occur in certain areas covered by the cast (Keret and MacEwen, 1991).

Operative management

Operative management of developmental dysplasia of the hip is only used after failure of non-operative treatment, or if treating a late presentation, commonly after 18 months of age. It is usually accompanied with a spica cast.

Open reduction

Open reduction is indicated for children that have failed closed reduction. It is commonly performed through the anterior Smith–Peterson approach, but the medial approach has also been used in patients less than 12 months old. The joint is surgically exposed and any obstruction, such as hypertrophied ligamentum teres, iliopsoas contracture, pulvinar or labral infolding, is corrected, to allow the femoral head to engage with the acetabulum (Studer et al, 2017). This reduction is then maintained by placing the hips in a spica cast. An adductor and psoas tenotomy are usually required to maintain the reduction.

Open reduction and osteotomy (pelvic, femoral or both)

This is indicated for children over 18 months of age with residual hip dysplasia. It is used to treat severe dysplasia accompanied by anatomical changes to either femur or acetabulum. These include excessive hip anteversion, coxa valga, shallow acetabulum with increased acetabular indices. The aim of surgery is to reduce the hip joint in a concentric fashion and depending on osteotomy used, it could rotate the femoral neck, place it in varus or increase the acetabular coverage. Multiple types of osteotomies have been described, with varying indications (Vaquero-Picado et al, 2019).

Adult presentation

Adult presentation with developmental dysplasia of the hip depends on the extent of disease progression and the degenerative changes that might have set in. Osteotomies around the hip (pelvis or femur) are always considered in the hope that they will preserve the hip and that a hip replacement will not be necessary (Noordin et al, 2010). Untreated developmental dysplasia of the hip can lead to tears within the labrum which can be painful and restrictive. This can be addressed with hip arthroscopy which is controversial because of its potential complications. Total hip replacement should be considered in patients who present with significant delay in diagnosis and established osteoarthritic and chondral damage. These are commonly complex cases because of the anatomical and morphological changes of the proximal femur and acetabulum, as well as possible limb shortening (Jakobsen et al, 2018).

Complications

Avascular necrosis can be seen with all treatment modalities, and is associated with excessive abduction in harnesses or casts. It is more likely to occur in patients who have previously undergone failed closed treatment.

Recurrence happens in around 10% of cases even with appropriate treatment, so all cases of developmental dysplasia of the hip should be followed up radiographically until maturity.

Transient femoral nerve palsy can be seen when patients are placed in Pavlik harnesses in excessive flexion.

Should we be screening for developmental dysplasia of the hip?

The importance of developmental dysplasia of the hip, and the availability of effective treatments, has led to the creation of screening programmes, to improve outcomes. However, this has proven to be a point of controversy in developmental dysplasia of the hip and it remains contentious as to whether clinical examination and ultrasonography for the at-risk group, or a population universal screening programme, is the optimal and cost-effective method of treatment. There is yet to be a clear international consensus as to the optimal method for screening and surveillance (Shorter et al, 2011; Paton, 2017). Paton maintains that the UK screening programme does not meet the World Health Organisation criteria for a screening programme, as set out in the Wilson and Jungner (1968) report and falls more in the category of surveillance.

In other countries, universal ultrasound screening of newborn hips has been implemented, but a meta-analysis by Shorter at al (2011) does not show this to decrease the number of cases of late-detected developmental dysplasia of the hip or reduce the requirement for surgery in later life, while increasing the numbers treated. On the other hand, Clegg et al (1999) demonstrated a reduction in surgical cost with routine screening as a result of earlier detection, and the overall combined cost of running the screening programme and treating the condition surgically matched the cost of other screening policies. This is the most recent calculation but in terms of treatment little has changed since 1999, and the cost of implementing surgery in later life has increased making the argument valid, if not the

Key points

- Developmental dysplasia of the hip is an abnormality of hip development, which can lead to a spectrum of hip disorders, from abnormal morphology, to subluxation or dislocation.
- The incidence ranges from 1–2/1000 in unscreened populations to 5–30/1000 in clinically-screened populations.
- The main risk factors include female sex, firstborn infant, breech presentation, family history and oligohydramnios.
- Clinical presentation can be occult, or present as skin-fold asymmetry, restriction in movement, abnormality of gait or leg length, or as pain in late presentation.
- Assessment is formed clinically by Barlow's and Ortolani's tests in the newborn infant examination. Radiologically, ultrasound is usually used before 6 months of age or until femoral head ossification, followed by radiographic assessment thereafter.
- Non-operative treatment is by Pavlik harness (or equivalent), abduction splintage, or closed reduction and plaster of Paris hip spica cast (this may involve operative adductor tenotomy).
- Operative treatment options include open reduction of the hip and hip spica, and possible femoral, acetabular or pelvic osteotomy.
- Major complications may arise such as avascular necrosis, chondrolysis and, in the long term, early degenerative hip disease.

exact numbers. The question remains whether a more sensitive, specific and cost-effective way of screening can be found and implemented in the UK.

Conclusions

Developmental dysplasia of the hip is a multifactorial disease, with complex aetiology and many interacting risk factors. It has a substantial impact on patients' quality of life and the possible need for significant surgical intervention in later life. Its importance, and the availability of effective treatments, has led to the creation of developmental dysplasia of the hip screening programmes, to improve outcomes. However, it remains contentious as to whether clinical examination and ultrasonography for the at-risk group, or a population universal screening programme, is the optimal and cost-effective method of treatment.

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Conflicts of interest

The authors declare no conflicts of interest.

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