

Managing Developmental Dysplasia of the Hip

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Abstract Developmental dysplasia of the hip (DDH) involves a spectrum of hip disorders that affect hip anatomy and development and can range from mild anatomical deformity with a reduced but subluxatable hip to a frankly dislocated hip. It was previously known as congenital dislocation of the hip (CDH) but this name is no longer used due to the fact that the hip may be anatomically abnormal whilst not being dislocated. The key aim of clinical management of DDH is early diagnosis and referral as this can often mean less invasive treatment is possible, and outcomes are significantly improved if treatment is initiated at an early stage and certainly before 6 wk of age.

Keywords DDH · Dysplasia · Risk factors · Screening · Splinting · Surgery

Introduction

Developmental dysplasia of the hip (DDH) involves a spectrum of hip disorders that affect hip anatomy and development and can range from mild anatomical deformity with a reduced but subluxatable hip to a frankly dislocated hip [1]. It was previously known as congenital dislocation of the hip but this name is no longer used due to the fact that the hip may be anatomically abnormal whilst not being dislocated. DDH is thought to affect 1–3 % of newborns [2, 3] or 5.5/1000 children at birth falling to 0.5/1000 at 2 wk of age [4]. The key aim of clinical management of DDH is early

diagnosis and referral as this can often mean less invasive treatment is possible, and outcomes are significantly improved if treatment is initiated at an early stage and certainly before 6 wk of age [5, 6].

DDH may present at any age and as such varies with its clinical manifestation. The newborn may present with a positive screening test, the toddler with a limp and the adult with early osteoarthritis.

Risk Factors

Breech presentation is a strong risk factor for DDH [7] with vaginal delivery leading to a 17-fold increase in DDH risk vs. a 7-fold increase in risk with elective cesarean section delivery [8]. “What constitutes breech” varies between different studies between breech in final trimester, breech at presentation or breech at delivery and the relevance of these is not yet understood.

Family history is also a risk factor, with one affected sibling meaning a DDH risk of 6 %, one affected parent a 12 % risk and if both one parent and one sibling are affected the risk of DDH to the neonate is 36 % [9]. Genetic studies on DDH families have identified key genes (*GDF5*, *IL-6*, *TGF-β1*, *PAPPA2*, *ASPN*, *TBX4*) which may play a role in DDH [10] but this does not impact on current clinical assessment and practice. Oligohydramnios is also associated with DDH [11], although congenital talipes equinovarus that has previously been suggested to have an association with DDH, has been questioned by a recent study which suggested there is no association between the two [12].

Twenty percent of DDH cases affect both hips, with the left hip being more commonly affected than the right; in the 80 % of cases that are unilateral. Eighty percent of those affected are females [13].

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Assessment

The presence of any of the risk factors identified is useful to guide clinical suspicion and if breech presentation or positive family history, there is evidence to support ultrasound screening in these patients to prevent late hip dislocation and the need for surgery [14]. In the absence of risk factors for DDH there is a less defined role for screening and thus high quality clinical assessment is the key, in the form of both Ortolani [15] and Barlow [16] tests. The Ortolani test detects the reduction of a dislocated femoral head, with the Barlow test identifying a reduced hip that it is possible to dislocate – suggesting instability. The tests should be performed by an experienced clinician who is familiar with and well practiced in the tests, and if equivocal, should be repeated. It should be noted that the tests would not identify a hip that is already dislocated or a hip with abnormal anatomy that does not dislocate. It is possible that bilateral hip dislocations could be missed due to the symmetry. A stable hip may still have anatomical abnormalities but these can only be detected on ultrasound.

Beyond the age of 2–3 mo both the Barlow and Ortolani tests become difficult due to increased muscle tone, and become less reliable when assessing the hip after the age of 6 mo. At this age the most reliable clinical sign is reduced hip abduction that will be asymmetrical in unilateral DDH. The child should be examined in a relaxed state without their nappy, and in this situation the examiner should be able to abduct the hip enough that their hand touches the examination couch when holding the leg. The Galeazzi test is also useful to identify a short femur — the hips and knees are flexed to 90 degrees and the relative position of the knees noted. If one knee is lower than the other it suggests there may be an abnormality of the hip leading to a shortening of the femur. Asymmetrical skin creases is an unreliable clinical indicator and should not be used to confirm or rule out DDH as asymmetry may be seen even in a child with normal hips.

If initial clinical examination is normal then the need for follow up assessment, and the suggested frequency of this is not clear, although there is one study in which repeated assessment at 3 mo was helpful in identifying late diagnosed cases of DDH [17].

Imaging

Ultrasound imaging is useful until approximately 4 or 5 mo of age and can use either the Graf (static assessment) or Harcke (dynamic assessment) techniques [18]. There is evidence to suggest that universal ultrasound screening does not lead to a decrease in rates of late detected cases of DDH compared with selective screening of those with identified risk factors [19]. It

has also been suggested that non-selective screening may lead to over treatment [19]. Despite this some countries such as Germany have universal screening programmes and have seen a decrease in the rates of surgical reduction in DDH [20], although universal screening programmes have significant cost implications.

An experienced sonographer should perform the ultrasound as it is highly operator dependent and like all aspects of musculoskeletal diagnosis it is important that the radiological and clinical findings correlate – if there is disparity between clinical and imaging findings then both assessments should be repeated. Ultrasound scans may show abnormalities even in a normal hip up to 4 wk of age and as such scans should again be repeated if there is uncertainty over the diagnosis, ideally before 6–7 wk of age.

The Graf technique uses a coronal section through the mid acetabulum and assesses hip morphology without assessing stability, which by definition needs a dynamic assessment as an attempt is made to sublax and reduce the hip. The key measurement is the alpha angle, which is a measurement of acetabular depth, by effectively assessing the osseous acetabular roof angle (Fig. 1a and b). The angles and grading are listed in Table 1.

The second measurement, the beta angle is also measured and defines the position of the acetabular labrum, thus assessing the coverage of the femoral head by the acetabulum.

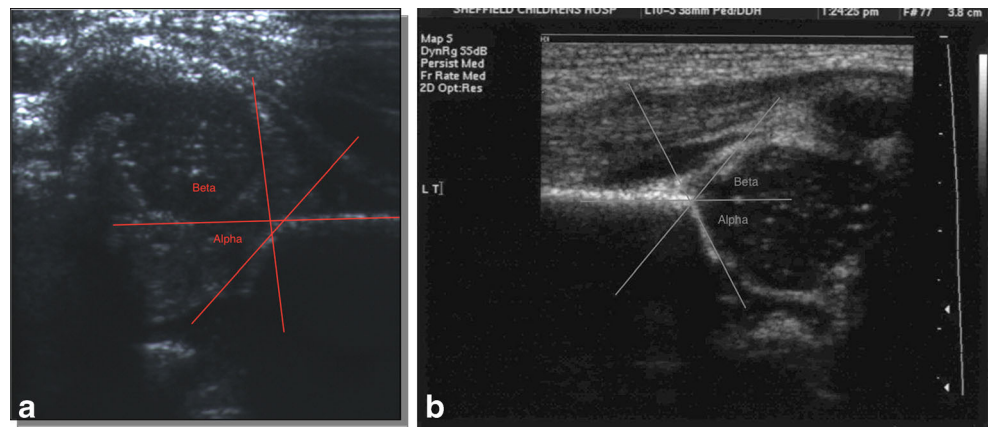
The Harcke dynamic ultrasound method uses images taken during the Ortolani and Barlow tests, to observe the relationship between the femoral head and the acetabulum as the hip is both sublaxed and reduced. Most centres use one of the two techniques rather than combining the two.

Radiographs are useful in assessing for DDH beyond 6 mo of age when ultrasound cannot be used. They rely on the ossification of the femoral head and use a standardised AP radiograph of the pelvis that should be taken with the hips in a neutral position (Fig. 2). The acetabulum is assessed using a series of radiological lines, with the key measurement being the angle formed between Hilgenreiner's line and the line forming the roof of the acetabulum. This measurement determines the effective depth of the acetabulum, and is termed the acetabular index with normal values being <35 degrees at birth and <25 degrees at 1 y.

Treatment

The key goals of treatment in DDH are to maintain function, prevent disability and avoid complications of treatment. Many unstable hips at birth will settle without intervention by 6 wk of age [21]. Treatment should be started by 2–6 wk of age though to ensure optimal outcomes, with initial treatment usually taking the form of abduction bracing, most commonly with the Pavlik harness.

Fig. 1 Ultrasound scan showing a) a dislocated femoral head and b) a centered femoral head with a dysplastic hip and lack of femoral head cover



Pavlik Harness

The most commonly used treatment method is the Pavlik harness (Fig. 3) [22], a dynamic splint that maintains the hip in flexion and abduction. The success of treatment is between 80 % and 96.7 % in the literature [23] with rates of avascular necrosis (the most significant complication of harness treatment) quoted between 0 and 28 % [23]. There is no consensus on the duration of treatment in harness but the reported length of treatment varies from 11 to 28 wk, and there is also no consensus on whether treatment should be ceased immediately or the time in harness reduced over a gradual period. During the main treatment period the harness should be worn at all times, and parental compliance may sometimes be an issue due to the difficulties it produces with caring for their child. If parental compliance is a concern, more regular follow-up should be arranged.

The harness is not suitable for use in those with teratological hip dislocation (Cerebral Palsy and other neuromuscular disorders) and those with irreducible hips.

Alternative Methods

There are other methods of bracing treatment that also rely on the same principle of maintaining the hip in an abducted position to maintain reduction and allow the normal acetabular development as it remodels around the femoral head. The Frejka pillow has a success rate of 90 % vs. 88 % for the Pavlik harness in one published study by Atar et al. [24] with an avascular necrosis (AVN) rate of 7 % vs. 6 % for Pavlik

harness. The differences in rates of both success and AVN did not reach statistical significance between the two groups. The Von Rosen splint has also been used to treat DDH with success rates of 99 % and an AVN rate of 0.6 % [25], but the recording of AVN rates may not be truly accurate and successful treatment was defined as lack of further intervention, in the absence of a patient outcome measure.

Although the Pavlik harness is widely used and gets good results, there is a lack of high quality evidence on both duration of treatment and outcome and complication rates, and also comparison studies with other treatment techniques. There is a definite role for large-scale prospective randomized studies going forward.

Surgical Treatment

The role of surgery in DDH is usually for those who have proved resistant to brace treatment or those who have previously been diagnosed at a late stage. The later the age at diagnosis the greater the magnitude of surgical intervention

Table 1 Graf angles and classification

Alpha angle	Classification
> 60 degrees	Normal
50–60 degrees	Immature
43–50 degrees	Mild dysplasia
< 43 degrees	Significant dysplasia

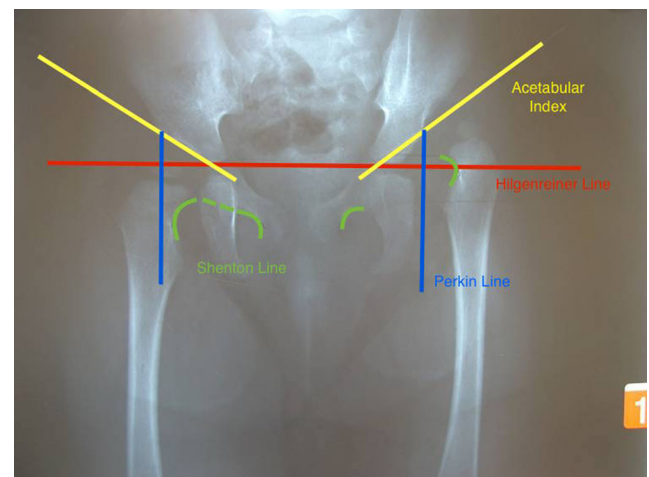


Fig. 2 Radiograph showing a left dislocated hip and severe dysplasia of the acetabulum. Shenton’s line is broken and has an increased acetabular index on the left



Fig. 3 Clinical photograph showing the Pavlik harness

usually required, further emphasizing the importance of early diagnosis. Surgery ranges from a closed reduction with or without adductor tenotomy; to pelvic and femoral osteotomy in late presenting or difficult cases. The key aim of any surgical intervention is to achieve a concentrically reduced femoral head within the acetabulum at the earliest stage possible to allow bony remodeling and normalisation of hip anatomy.

Closed Reduction

This usually involves an examination under anesthesia (EUA) and arthrogram to establish the anatomy of the hip and the contributing factors to the hip instability. It is usually followed by a period of immobilisation in a hip spica plaster but there is no agreement in the literature on how long the hip should be immobilised for. It is important that post operative imaging is obtained to ensure the hip joint is reduced in the plaster – usually in the form of CT or MRI scan. The main complications of closed reduction are AVN and redislocation or failed treatment.

The AVN quoted in the literature for closed reduction varies between 2.6 % [26] and 60 % [27], with a marked variation due to a lack of agreement between studies as to what exactly is AVN and how it should be measured. The re-dislocation rate varies from 2.8 to 13.6 % following closed reduction [23]. The timing of closed reduction also remains contentious amongst the pediatric orthopedic community with some arguing that it should only take place once the ossific nucleus of the femoral head is apparent on radiographs and others arguing that there is no need to wait for this stage.

Open Reduction

Open reduction should be used when closed reduction has failed or cannot be achieved and is the next step in the treatment ladder and is performed *via* either an anterior (Smith

Peterson) or medial (Ludloff) approach to the hip. Although the medial approach carries a theoretical risk of injury to the blood supply to the femoral head which comes *via* the circumflex vessels and potentially higher rates of AVN secondary to this, there is no evidence to support this with rates of AVN equal between the two approaches [23].

Bony Surgery

Bony surgery is used if closed and open reduction are either unsuitable or have failed. The surgery is usually in the form of pelvic osteotomy to change the shape/volume of the acetabulum, femoral osteotomy to redirect the femoral head, or both pelvic and femoral osteotomy combined. There are a number of techniques to perform both pelvic and femoral osteotomy, all of which have good published results in case series but there is a lack of randomized trials comparing different osteotomies and their outcomes.

Neglected or Missed DDH

The long-term sequelae of DDH, if left untreated or undertreated are of hip dislocation, or abnormal hip morphology that usually leads to pain and disability. Early onset osteoarthritis, low back pain and mobility problems are all seen in untreated cases. A previous Norwegian study based on the joint registry suggested that 29 % of total hip replacement surgeries in those aged less than 60 were due to DDH [28].

Conclusions

DDH is a common but highly treatable cause of disability in the pediatric population. The key to successful treatment is early diagnosis and commencing treatment, if possible before 6 wk of age. It must be emphasized that a high proportion of neonates with an abnormal hip examination at birth will resolve by 6 wk; hence vigilance and repeat assessment are key points. The role of screening programmes, except for those with established risk factors remains unproven, and the Cochrane review published in 2013 suggested there is a current lack of evidence to support universal screening [29].

There are a number of unanswered questions with regards to the treatment of DDH and there is a need for multi-centre prospective studies to look at aspects of both conservative and surgical treatment techniques.

Early hip examination by a trained clinician familiar with neonatal hip examination is paramount and if there is any suspicion of hip abnormality, repeat examination and/or referral to a pediatric hip specialist should take place within the first

few weeks of life, supported by ultrasound screening at 3–6 wk of age, if necessary. Those with a normal hip examination should continue to be assessed on a regular basis until 1 y of age.

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Compliance with Ethical Standards

Conflict of Interest None.

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