Developmental dysplasia of the hip (DDH)

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Summary

Developmental dysplasia of the hip (DDH) is common, ranging from mild dysplasia to frank dislocation. The diagnosis can be difficult, even in experienced hands, particularly when there are bilateral dislocations. All infants should be screened clinically, but the value of other screening methods, such as ultrasound, is still debated. Initial treatment in the infant is with a Pavlik harness. If this is ineffective or if the child presents later, more aggressive treatment, such as a closed reduction, or even surgical reduction may be indicated. All hips must be carefully followed until maturity. If diagnosed and treated promptly excellent results can be obtained, but long-term sequelae occur even in patients given optimal treatment.

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Introduction

Developmental dysplasia of the hip (DDH) has been recognized from the time of Hippocrates. It is a common condition which remains controversial and confusing despite diagnostic and treatment advances. The terminology can be unclear and inconsistent, diagnosis can be subtle and there can be long-term sequelae even in patients given optimal treatment. For example, despite DDH having been documented to occur in patients who did not have dysplasia a birth, there is still an erroneous belief that all cases of DDH are present at birth and, if diagnosed later, represent a "missed" diagnosis. The value of screening methods is still commonly debated. Thus it is unsurprising that DDH is commonly associated with malpractice suits, even though results of treatment are generally quite good.

The aim of this article is to present a basic outline of the disorder, reflect upon some of the controversies, and provide an algorithm for the treatment of the child with DDH.

Terminology

Discussions of congenital or developmental disorders of the infant hip are made more difficult by conflicting terminology. This condition has been called: hip dysplasia, congenital dislocation of the hip (CDH), developmental dysplasia of the hip (DDH).
In part this is due to attempts to reflect the aetiology, which is still unclear, or at least unpredictable. Thus in 1991, the Pediatric Orthopaedic Society of North America and the American Academy of Orthopaedic Surgeons endorsed the term "DDH" in an effort to emphasize the developmental nature of the hip disorder. As The American Heritage College Dictionary defines "dysplasia" as "abnormal development or growth" "DDH" is somewhat redundant! Most commonly, DDH is used when referring to instability, dislocation or acetabular dysplasia of the hip. The abnormal development of the hip includes the osseous structures, such as the acetabulum and the proximal femur, as well as the labrum, capsule, and other soft tissues. However internationally, this disorder is often still referred to as "CDH," but DDH is the term used in this article.

More specific terms are often used to better describe the condition. A natural tendency to use phrases that refer to physical examination findings (instability), or anatomic abnormalities (subluxation or dislocation) in an aetiologic or pathophysiologic sense, complicates interpretation of existing literature. We have attempted to define these terms as follows:

**Instability:** A clinical diagnosis made on examination where passive manipulation subluxates or dislocates the hip.

**Subluxation:** An anatomic diagnosis, usually made from imaging studies, in which there is incomplete contact between the articular surfaces of the femoral head and acetabulum.

**Dislocation:** An anatomic diagnosis, usually made from imaging studies, in which there is complete loss of contact between the articular surface of the femoral head and acetabulum.

**Teratologic:** A diagnosis in which physical examination and/or imaging studies show a dislocated hip prior to birth.

### Incidence

The overall frequency is usually quoted as approximately 1 in 1000. This figure depends on how DDH is defined as demonstrated by Barlow’s classic study, in which he reported that the incidence of hip instability in the newborn examination was as high as 1 in 60. However, over 60% of these hips became stable by 1 week and 88% by 2 months. Extrapolation of these findings suggests an incidence of 1/500 (12% of the 1 in 60, or 0.2%) with residual hip instability, as compared to the usually cited figure of 1 in 1000.

### Anatomy/aetiology

The normal growth of the acetabulum is dependent on normal epiphyseal growth of the triradiate cartilage and the three ossification centres of the acetabular portion of the pubis (os acetabulum), ilium (acetabular epiphysis), and ischium. Normal growth is also dependent on normal interstitial appositional growth within the acetabulum, the presence of the spherical femoral head within the acetabulum appears to be critical for stimulating normal development of the acetabulum.

Many factors may contribute to DDH. The high incidence of DDH among Native Americans and Laplanders (nearly 25–50 per 1000) and low incidence among the southern Chinese and African-American populations, as well as increased incidence among children whose parents have DDH, suggests underlying genetic influences.

Otherwise normal neonates with findings of hip instability and ligamentous laxity on physical examination may subsequently develop radiographic evidence of hip dysplasia. This has led many to conclude that ligamentous laxity is a significant aetiologic factor. However, DDH is not characteristic of collagen abnormalities with significant ligamentous laxity, such as Ehlers-Danlos syndrome or Marfan’s syndrome. Barlow noted that most neonates with instability have normal hip development without treatment. It may therefore require more than just ligamentous laxity for DDH to result. At birth, Caucasian children (who have a higher incidence of DDH) tend to have a shallower acetabulum, as compared to children of other races. Therefore, there may be a susceptible period at birth in which abnormal positioning or a brief period of ligamentous laxity combined with a shallow acetabulum may result in hip instability.

Other factors related to DDH include intrauterine positioning and gender, and some of these are interrelated. Female gender and breech positioning are all associated with an increased incidence of DDH. Eighty percent of those with DDH are female, and the incidence of breech positioning in children with DDH is approximately 20% (compared with 2–4% in the general population.) The incidence of DDH in females born breech is as high as 1 in 15.

Other musculoskeletal disorders of intrauterine malpositioning or crowding, such as metatarsus adductus and torticollis, have been reported to be associated with DDH. Oligohydramnios is also reported to be associated with an increased incidence of DDH. The left hip is more commonly affected by DDH, and this may be related to the common intrauterine position of the left hip against the mother’s sacrum, placing it into an adducted
position. Children in cultures where the mother swaddles the baby, keeping the hips adducted, also have a higher rate of hip dysplasia.

Abnormal development of the hip also occurs in children with underlying neuromuscular disorders, such as cerebral palsy, myelomeningocele, arthrogryposis, and Larsen’s syndrome, although hip dysplasia in these children is not usually classified as DDH.

Clinical presentation

Careful examination of the hip in the newborn period will identify the majority of the infants with DDH, but not all children with DDH have clinical signs at birth, even when examined by physicians with specialty training or when augmented by other imaging modalities. This may be due to the difficulty of making the diagnosis, or the hip may not have been unstable or dislocated at birth.

Ortolani and Barlow designed physical diagnostic tests to detect unstable hips in infancy. The Ortolani test is performed by placing the thumb over the inner thigh and the index finger on the greater trochanter. The hip is abducted and gentle pressure is placed over the greater trochanter. A “clunk”, like turning a light switch on or off, is felt when the hip is reduced. This should be done gently, such that the fingertips do not blanch. Ortolani felt that a positive test was associated with reduction of the hip into the acetabulum, but it may also be caused by passage of the femoral head over a biconcave ridge in the acetabulum. The Barlow test is performed with the hips in an adducted position and slight gentle posterior pressure applied to the hips. A clunk should be felt as the hip subluxes out of the acetabulum. A high-pitched ‘click’ (as opposed to a ‘clunk’) in all likelihood has little association with acetabular pathology. To perform these maneuvers correctly, the patient must be relaxed, and only one hip should be examined at a time.

Clinical examination when the child is 3–6 months of age is quite different. At this age, findings with the Ortolani and Barlow exams are more difficult to discern. The Galeazzi sign (or Allis sign) is a classic identifying sign for unilateral hip dislocation. This is performed with the patient lying supine and the hips and knees flexed (Fig. 1). Examination should demonstrate that one leg appears shorter than the other. Although this is usually due to hip dislocation, it is important to realize that any limb length discrepancy will result in a positive Galeazzi sign. Additional physical examination findings for late dislocation include asymmetry of the gluteal thigh or labral skin folds, although thigh folds have been reported to be abnormal in as many as 20% of all newborn children. Limited abduction of the affected hip is also a common finding in children with unilateral DDH. Children commonly stand or walk with external rotation, and this is most likely due to contractures of the external rotators, but it can also be a sign of a dislocated hip, especially if it persists beyond early walking age.

Bilateral dislocation of the hip, especially at a later age, can be quite difficult to diagnose. This often presents as a waddling gait with hyperlordosis. Many of the above-mentioned clues for a unilateral dislocated hip will not be present, such as Galeazzi sign, asymmetrical thigh and skin folds, or asymmetrically decreased abduction. Careful examination is essential and a high level of suspicion is important.

Imaging studies

Ultrasound plays an important role in both the diagnosis and treatment of DDH. Most authors agree that it is an excellent tool for the assessment of children with suspected hip instability (i.e. when clinical examination is equivocal) and is useful as an adjunct in the treatment of children with hip dysplasia, especially in monitoring reduction by closed methods. However, routine ultrasound evaluation of newborns is more controversial; the primary concern is the over-diagnosis (increased false-positive results) of hip dysplasia. In addition, the routine use of ultrasound for children, even those with risk factors, has not yet been shown to
reduce the prevalence of late diagnosis of hip dysplasia. After the femoral head begins to ossify (at 6–12 months of age), the usefulness of ultrasound diminishes. Therefore, the use of ultrasound for routine screening children is not currently recommended.

Radiographic evaluation of the pelvis can be used to assess the hips, although early radiographs (within the first 6–12 weeks) may be misleading. Numerous radiographic measurements have been used to assist in the evaluation of DDH (Fig. 2). The most commonly used are:

**Hilgenreiner’s line:** Drawn from an anteroposterior radiograph of the hips, a horizontal line is drawn between the triradiate epiphyses.

**Perkin’s line:** Drawn perpendicular to Hilgenreiner’s line through the superolateral edge of the acetabulum, dividing the hip into four quadrants. The proximal medial femur should be in the lower medial quadrant or if the ossific nucleus of the femoral head, if present (usually seen between 4 and 7 months), should be in the lower medial quadrant.

**Acetabular index:** The angle between Hilgenreiner’s line and a line drawn from the triradiate epiphysis to the lateral edge of the acetabulum. Classically, this decreases with age and should measure less than 20° by 2 years of age.

**Shenton’s line:** A line drawn from the medical aspect of the femoral neck to the inferior border of the pubic rami. It should create a smooth arc that is not disrupted. If disrupted, it indicates some degree of hip subluxation. This measurement may be unreliable in infants.

Dozens of other radiographic measurements have been proposed to assist with the radiographic evaluation of DDH in infants, children, and young adults, which can be confusing. For example, the acetabular index is used to assess the acetabular depth in infants and children and is often measured in an attempt to assess the effect of treatment.

The acetabular angle of Sharp is primarily used to assess residual dysplasia in patients after the closure of the tri-radiate cartilage. It is important to understand the indications and limitations of these measurements.

As well as the radiographic measurements, there are a number of radiographic descriptions used to assess the degree of dysplasia and the effects of reduction. The acetabular teardrop is formed by the cortical surfaces of the acetabular fossa (laterally), the pelvic wall (medially) and the cotyloid notch (inferiorly). If the “teardrop” takes on a “V-shape” with widening of the superior width, the prognosis is poorer. The shape of the “sourcil,” or eyebrow, of the acetabulum can also be a gauge to the degree of acetabular dysplasia. An evenly distributed sourcil with the lateral edge horizontal or curved downward is an indication of a normally developing acetabular roof, whereas a sourcil that has a lateral triangular shape or is curved upward is more consistent with a dysplastic acetabulum. More recently, the presence of the acetabular notch (a scooped deformity on the superolateral aspect of the acetabular edge) has been associated with dysplasia.

It can be difficult to delineate radiographically acetabular dysplasia in the young adult that is severe enough to lead to early osteoarthrosis. A centre edge (CE) angle of less than 16° has often been used to predict early osteoarthrosis, but other authors have found this measurement to be less reliable. Subluxation, defined as a break in Shenton’s line, has been shown to be associated with osteoarthrosis and decreased function.

Arthrograms are dynamic studies performed by injecting dye into the hip joint and examining the patient with aid of fluoroscopy, usually with the patient under anesthesia. Although this can be performed independently, it is routinely performed in conjunction with a closed reduction. Arthrography can be helpful in determining the underlying cartilaginous profile and dynamic stability of the hip.

**Treatment**

Early diagnosis aids in the successful treatment of DDH. Prompt recognition and appropriate treatment provides the child with the greatest opportunity for a good outcome. An algorithm is outlined in Table 1.

**Pavlik harness**

The Pavlik harness is the treatment of choice for neonates with hip instability. It is designed to hold...
the hips in a position that encourages normal growth and development while permitting motion within a safe range. To ensure that the Pavlik harness is fitted properly to hold the hip in a reduced position, close supervision and frequent adjustments are essential.\textsuperscript{16} It should be placed such that the chest strap is at the nipple line with two finger breadths of space between the chest and strap. The anterior strap should be positioned at the mid-axillary line with the hips flexed to no more than 100°–110° as excessive hip flexion can lead to femoral nerve compression and inferior dislocations. The posterior abduction strap should be at the level of the child’s scapula and adjusted to allow for comfortable abduction, thus preventing the hips from adducting to the extent that the hips dislocate. Excessive abduction should be avoided because of concern regarding the development of avascular necrosis. It is important to carefully monitor the patient to insure that the harness fits and the hips are reduced. Fitting should be checked clinically within the first week and then weekly thereafter, and quadriceps function should be assessed at all clinic visits. Ultrasound is an excellent means of documenting stability and reduction of the hip in the Pavlik harness and should be performed early in the course of treatment.

If the hip is not reduced, then the Pavlik harness should be discontinued. Risk factors for failure of the Pavlik harness include: bilateral dislocations, older age at time of initial treatment, inability to reduce the hip with manipulation, and initial coverage less than 20% as determined by ultrasound.\textsuperscript{16,17}

There is little consensus as to the overall time the Pavlik should be worn, although a minimum of 3 months seems to be reasonable. If the hip is satisfactorily reduced in the harness, we maintain this treatment at least until the hip is stable clinically and by ultrasound out of the brace. Abduction splinting is maintained thereafter until there is no radiographic evidence of residual dysplasia. The use of an abduction brace if there is a failure of the Pavlik harness treatment has been suggested to be successful in over 80% of patients.

The Pavlik harness is not appropriate for use in infants in whom diagnosis is made later than 6

Table 1  A suggested algorithm for treatment of DDH.
months of age. In these patients, contractures of the hip capsule and supporting ligaments prevent gentle reduction of the hip. Interposition of the limbus or the tendon of the iliopsoas tendon may block the femoral head from entering the acetabulum.2

Closed reduction

Patients presenting between 6 months and 2 years of age are best managed by a period of skin traction followed by careful reduction under anesthesia. Closed reduction is performed with the aid of arthrography, used to determine the adequacy of the reduction. Arthrography is especially useful in determining the adequacy of closed reduction of the hip. A medial dye pool and/or an interposing limbus is associated with a poor prognosis. If, on the other hand, there is a sharp or even a blunted limbus and no medial dye pooling, prognosis is good.

The safe zone of Ramsey, the angle between maximum abduction and minimum abduction in which the hip remains reduced, should also be measured. This should be at least 25° and can be increased with release of the adductor longus. The cone of stability is a cone that involves hip flexion, abduction and internal/external rotation. If this cone measures greater than 30°, it is considered satisfactory.

Then a spica cast is applied, with care taken in molding over the posterior aspect of the greater trochanter of the ipsilateral limb. Then a post-operative CT scan is taken to ensure there is no evidence of posterior subluxation. The cast should be worn for 6–12 weeks, when the hip is reexamined and, if found to be stable, the patient is placed in an abduction brace. If still unstable, a further period in a spica cast is necessary.

Operative treatment

Indications for surgery are met if the expected results of the surgery are better than those of the natural history. The natural history of untreated hip dislocations depends, in part, on the severity of the disease, bilaterality, and whether or not a false acetabulum is formed. Unilateral dislocations result in significant leg length inequality, with a gait disturbance and possibly associated hip and knee pain. The development of a false acetabulum is associated with a poor outcome in approximately 75% of patients. Bilateral hip dislocation in a patient without false acetabuli will have a better overall prognosis. In fact, Milgram reported a case of a 74-year-old man with no history of hip or thigh pain whose dislocated hips were only discovered shortly before his death.

Relative contraindications to surgery include age over 8 years for a unilateral hip dislocation, or over 4–6 years for bilateral hip dislocation, especially if there is no false acetabulum.

Open reduction

Open reduction is the treatment of choice for the child presenting older than 2 years of age or for the younger child after failed closed reduction.

There are two surgical approaches for open reduction of the hip: the medial approach and the anterior approach.

The advantages of the medial approach are:
- if bilateral both hips can be reduced at the same time,
- obstacles to reduction e.g. psoas tendon are easily identified,
- the adductor longus can be sectioned through the same incision,
- the hip abductor muscles and iliac apophysis are avoided,
- the resultant scar is relatively inconspicuous.

Problems with this approach include:
- increased risk avascular necrosis,
- the potential lack of familiarity of surgeons with this approach,
- the inability to perform capsular plication or a pelvic procedure through this incision.

If a medial approach is used, the post-operative cast plays a much more important role than the more commonly used anterolateral approach. This can, if necessary, be combined with a capsule plication, and/or an acetabular procedure. Between the ages of 2 and 3 years, either traction or femoral shortening can be considered, but in a child older than 3 years of age, femoral shortening is typically performed instead of traction18 (Fig. 3). Proximal femoral dysplasia, such as that seen with significant anteverision or coxa valga, can also be corrected at the same procedure. After open reduction, a spica cast is worn for 6–12 weeks. As with a closed reduction, a CT scan may be necessary to confirm reduction. The patient is then placed in an abduction orthosis. The length of time in a hip orthosis remains controversial and depends on the treating physician’s experience and the individual
patient. The authors’ practice is for full-time orthosis wearing for 3 months after cast removal, and wean to night-time splinting when there is radiographic evidence of acetabular remodeling and no recurrent subluxation.

Pelvic osteotomies

A pelvic osteotomy for residual hip dysplasia may be necessary. A pelvic osteotomy can be performed at the time of open reduction, and is indicated if there is persistent instability after open reduction. As acetabular remodeling may occur after open reduction, some surgeons feel at least 12–18 months of acetabular remodeling should be allowed before an acetabular procedure is undertaken. In children over 4 years of age with significant hip dysplasia, an acetabular procedure should be considered at the time of open reduction due to the decrease in remodeling potential.

Complications

Complications include

- Redislocation,
- stiffness of the hip,
- infection,
- blood loss,
- the most devastating, avascular necrosis (AVN) of the femoral head.

The incidence of AVN varies significantly, depending on the study, from 0 to 73% and may be evident years after the surgery. Numerous studies show that extreme abduction, especially combined with extension and internal rotation, results in a higher incidence of AVN. All reductions (open or closed) have a risk of AVN, and all patients must be followed to skeletal maturity.

Results

Overall, the prognosis of children treated for hip dysplasia is good. In one study of children between the ages of 1 and 3 years, 83% had a good or excellent result (Severin class I or II).

Conclusion

Developmental dysplasia of the hip (DDH) is a common disorder. Clinical screening exams should be performed in all infants, and repeated at each evaluation until walking with a normal gait. Special attention should be directed towards children with underlying risk factors. Additional evaluation methods, such as ultrasound or radiographs may be needed. The diagnosis can be difficult, even in experienced hands, and a late diagnosis does not necessarily imply a missed diagnosis. Treatment depends on the age of initial diagnosis and success.
of previous treatment. Earlier treatment usually requires less aggressive methods. Follow up should be continued to skeletal maturity, and both hips should be carefully evaluated. Although outcomes are usually very good, long-term sequelae occur even in patients who receive optimal treatment.

References