(iii) Developmental dysplasia of the hip

Benjamin Holroyd
John Wedge

Abstract
Developmental dysplasia of the hip (DDH) is a spectrum of pathologies affecting the infant hip ranging from asymptomatic subtle radiographic signs through mild instability to frank dislocations with an abnormal acetabulum. Patients with developmental hip dysplasia account for around 10% of all primary hip arthroplasties, and around 30% in those under sixty. Early detection and appropriate management can prevent or delay the requirement for total hip replacement.

In this article we aim to provide a broad overview of the aetiology, natural history, pathology and management of developmental dysplasia of the hip.

Keywords developmental dysplasia of the hip (DDH); open reduction; pelvic osteotomy; screening; spica

“It is difficult to portage a canoe with a man who limps — he dips and you don’t.”

The week beginning 23rd February 2009 marked Baby Hip Health Awareness Week organized by the STEPS charity. This coincided with a Parliamentary Early Day Motion proposed by the MP for Blaydon, Mr David Anderson, calling for the Government to improve shortcomings in screening for and patient information about developmental dysplasia of the hip (DDH).

Introduction
DDH, despite a recent change in nomenclature, is not a new diagnosis, having been described as early as Hippocrates. It is a spectrum of pathologies affecting the infant hip ranging from asymptomatic subtle radiographic signs through mild instability to frank dislocations with acetabular abnormality. Many authors have attempted to subdivide the condition into distinct entities. Although popular opinion now favors a continuous spectrum, adolescent and young adult patients presenting with acetabular dysplasia may represent as an entirely distinct entity.

Initially referred to as congenital dislocation of the hip, recognition that apparently normal hips on examination at birth did not exclude the disease led to a change of nomenclature to developmental dysplasia of the hip.

Benjamin Holroyd MBBS BSc FRCS (Tr & Orth) Orthopaedic Fellow at The Hospital for Sick Children, Toronto, Ontario, Canada MSG 1X8.

John Wedge OC MD FRCS(C) Professor of Surgery at University of Toronto and a Chair of Advisory Council for Sick Kids International, Canada.

As Connolly\(^2\) points out, DDH has both anatomical and radiological definitions. The anatomical definition refers to abnormal development of the femoral head and acetabulum, while the radiological definition refers to a break in Shenton’s line. To this definition can be added a hip with an increased acetabular index.

Aetiology
The underlying aetiology, although hypothesized, is not clear but is likely to be multi-factorial. In this review we have concentrated on non-teratological and non-neurological dislocations. For want of simplicity and clarity we have chosen to leave dislocated hips in these two groups to another discussion, as we feel there is doubt as to whether they should be included in the definition of DDH at all.

DDH is predominantly a female condition (5:1), giving rise to hypotheses pertaining to a hormonal aetiology. Relaxin is the hormone most commonly implicated. It may however be the fact that up to twice as many females as males are born breech. Males with the disease are often more resistant to treatment. It is more common in the left hip as this is the adducted hip lying against the sacrum in the most commonly occurring intrauterine position. The disease is bilateral in 20% of patients.

There is a geographical predilection. It is common in Native American Indians (1 in 20) and rare in sub-Saharan Africans, supporting postnatal influences such as swaddling as a causative factor. The close association with torticolis and foot deformities support the ‘tight packaging’ theory.

Other than female sex, risk factors for DDH and ones that may be used to rationalize ultrasound hip screening include breech presentation, primiparous babies, high birth weight, family history, multiple pregnancies and oligohydranmios.

Natural history
Why intervene at all? The reason is that patients with DDH account for 10% of all total hip replacements,\(^3\) and up to 30% in the under 60’s.

Complete dislocations may have little functional disability. Patients will have leg length discrepancies if unilateral and a Trendelenberg gait. Perceived disability is largely dependent on socio-economic factors. Walker\(^4\) reports that Navajo Indians consider dislocated hips in much the same way as urban societies view left handedness. The development of a false acetabulum is the best predictor of outcome.\(^4\) A false acetabulum undergoes degenerative change in much the same way as a native joint. Based upon an early modification of the Harris hip score only a quarter of patients with well-developed acetabula have a good result into adulthood, compared to half of those with no or poor false acetabula.

Hips that sublux usually develop significant osteoarthritic changes in the third or fourth decades. Stable hips with radiological evidence of dysplasia are less predictable, although it is uncommon to see hips without any degenerative change beyond the fifth decade. The concentration of forces through a reduced weight bearing area is the primary cause of early onset osteoarthritis, although the inherent quality of the cartilage may also play a role. The more dysplastic a hip, the smaller the weight bearing area between articular surfaces.

MINI-SYMPOSIUM: CHILDREN’S HIP PROBLEMS

ORTHOPAEDICS AND TRAUMA 23:3

162

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Presentation
A patients’ presentation can broadly be categorized into early or late. Arbitrarily, six months of age is often used as a cut-off between the two groups, although it may again be considered a spectrum.

Early presenting patients may arrive in the hip clinic as a result of findings on baby checks, as detected by screening of at risk babies, or because of parental concern. Parents often report clicking in a baby’s hip when handling the child, particularly when changing nappies. This reported sign is a notoriously poor predictor of hip pathology as it often arises from the knee, the iliopsoas tendon moving over the femoral head or the fascia lata moving over the greater trochanter. Abnormality of soft tissue creases also corresponds poorly with DDH, despite being an often quoted finding in textbooks. Leg length discrepancy as assessed by Galeazzi’s test may not pick up more subtle differences, and is obviously absent in bilateral dislocations. We along with many authors have found a limitation of hip abduction and obvious tightness in the adductor longus tendon to be the most sensitive examination to detect a dislocated hip. Very infrequently does a child with a dislocation have full, symmetrical abduction.

Late presenting hips often present at the time of walking. There is a weak association between delayed walkers and DDH. A Trendelenberg gait may be noted, although in toddlers this may be difficult to appreciate even to the experienced. Leg length discrepancy is a common reason to present but children never present with pain. The first referral may be as a result of symptomatic OA in the young adult.

Radiological findings
Before something can be considered pathological it is important to consider what is normal. The work by Tonnis from 1975 described normal values in both adults and children and is considered the classic description.

The acetabular index changes with age from 30° (+/−5°) at birth to 20° (+/−5°) at 5 years. Tonnis reported that values falling two standard deviations above the mean were definitely pathological, but that a grey area existed between one and two standard deviations. He concluded that all those above a single standard deviation should be treated as only 25% become normal. As with all radiographic measures inaccuracies are common if positioning of the child allows postural artefacts to occur.

The centre edge angle of Wiberg is notoriously difficult to measure in the under 5’s, as it is difficult to pinpoint the centre of the femoral head. For this reason there are no meaningful data of normal values and it is therefore of very limited clinical value at this age. It does, however, offer an indication of femoral head lateralization. Tonnis described the lower limit of normal, in other words one standard deviation below the mean to increase the likelihood of iatrogenic damage. Even in experienced hands, it has been postulated that a disruption of the negative intra-articular pressure within the hip joint can lead to dysplasia in an otherwise healthy joint. Jones examined ten neonatal hips in stillborns after repeated Barlow testing and found that the posterior capsule was not a strong or important structure, but that the vacuum fit between femur and acetabulum was.

Positioing a hip to take a forced, frog lateral X-ray is enough to disrupt the labral seal containing the negative intra-articular pressure as evidenced when an air arthrogram is inadvertently created.

Other than iatrogenic hip injury, examination as a hip screening tool is problematic on account of its low sensitivity. Jones reports sensitivity likely to be less than 60% despite the high specificity, approaching 100%, as there are few false positives. Combining clinical examination with an ultrasound investigation (Figure 1) increases the sensitivity to approximately 90%.10

Some authors would argue that screening all births by examination and ultrasound leads to parental anxiety. But Schoenecker11 points out that this is “a fallacious reason not to screen, as with any healthcare issue in infancy in which early detection can lead to a simple and definitive treatment of a potentially pathological condition”.

Barlow12 demonstrated hip instability in 1 in 60 of newborns. Untreated, 60% will stabilize in the 1st week, 88% by 2 months. As it is not possible to predict which of the unstable hips will normalize, the consequences of not treating an unstable hip are severe and the risks associated with early treatment relatively low, most authors recommend that all unstable hips should be treated. However, follow up with regular ultrasound and delaying treatment until four to six weeks of age is probably safest.

MINI-SYMPOSIUM: CHILDREN’S HIP PROBLEMS
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The exception to this is the hip that is dislocated at rest, with an entry clunk, which should be treated urgently (Figure 2).

There have been many ultrasound techniques described, those by Harcke, Terjesen, Suzuki and Graf being the most commonly used. The Graf technique is the most popular technique employed in the UK. He popularized hip ultrasound in 1978 with his classification from 1, a normal hip, through to 4, a dislocated hip with no discernable acetabulum. The ultrasound must meet strictly defined criteria in order to avoid faulty diagnosis. Graf reports a misdiagnosis of normal hips that later require surgical intervention to be as high as 20% in patients who he felt had inadequate ultrasound scans. Clearly adequate training of ultrasonographers or clinicians is integral to the success of the diagnosis and hence treatment.

Pathophysiology

From studies that have examined aborted foetuses, we know that the normal hip begins to form as early as the 7th week of gestation. Primitive mesenchymal cells give rise to the limb buds that differentiate into the four extremities. A cleft first appears in the mesoderm at the end of 8 weeks and represents a cartilaginous model of the hip. By the 11th week the femoral head is spherical, the acetabulum reciprocally shaped and the capsule, synovium and ligamentum teres identifiable.

The acetabulum continues to develop through intrauterine life, the labrum contributing significantly to its depth. Having initially been deeply set, it becomes shallowest and therefore least stable in late pregnancy, presumably to facilitate delivery. After birth, it again becomes more deeply set. This perinatal period is the time of greatest risk of instability.

Sequential ultrasound investigations around this time can demonstrate how in a matter of weeks the hips can significantly deepen. The immature hip may only need observation as it normalizes, rather than commencing potentially damaging treatment.

During the adolescent growth spurt, three secondary ossification centres develop, further deepening the joint. The os acetabuli in the pubis contributes to the anterior wall. The acetabular epiphysis of the ilium forms a major part of the superior margin of the acetabulum. This structure is at risk from the inexperienced surgeon when performing open reductions and pelvic osteotomies, and must be given due respect. Finally, a secondary centre on the ilium contributes to the posterior wall.

Most of the primary pathological changes that happen occur on the acetabular side of the hip joint. The abnormal growth and development is related to primary pathology of the acetabular growth plates. Secondary changes from misdirected pressure from the femoral head also contribute.

Somerville felt that femoral neck version played an important role and that changing this and the valgus/varus angle to redirect the head to the centre of the hip would facilitate remodeling on the acetabular side.

It is worth mentioning that the same author amongst others believe ligamentous laxity may play a role. Without adequate pressure from the anterior hip ligaments, femoral neck anteversion remodeling is less likely to occur, increasing the chances of eccentric loading of the acetabulum and altering the growth potential on the acetabular side.

Salter was not concerned by femoral anteversion, provided that a concentric reduction could be maintained during walking, as he considered the version would correct spontaneously.

Although we have described DDH as a continuous spectrum we would specifically like to highlight one type of dislocation that we believe merits special consideration. We have observed in our own institution as well as elsewhere, surgeons experience greater trepidation when dealing with the higher dislocation. We suggest that the low dislocation that “slides” out requires more careful consideration. In our experience these hips are more likely to re-dislocate or not resolve as satisfactorily post-reduction. This may be because of pressure on the lateral acetabular epiphysis as the hip dislocates, inhibiting growth potential of the acetabulum post-reduction. The high dislocation exerts little or no pressure on this important growth centre, so once the femoral head is reduced and covered by the acetabulum the socket can grow and remodel reasonably well.

Treatment

We do not feel that describing our treatment algorithm in depth would be useful as there will always be debate as to how and when
to initiate various treatments. We do however want to highlight the basic treatment principles and options available, along with the reasoning behind commonly adopted timings for interventions.

It would seem logical that an early concentric reduction would give the optimal result. Several authors have reported that this is not necessarily the case and that hip reduction does not automatically lead to normal acetabular growth. Using the acetabular index as a marker of growth, Harris concluded that “the later the age of congruity, the more likely it is that the acetabulum will be dysplastic”. He went on to state that “the critical age for obtaining a congruous reduction in the functional position is four years and that above this age an unsatisfactory acetabulum is the probable outcome”.

The timing of a reduction appears to be key to the prognosis. Albinana observed the timing of appearance and morphology of the teardrop in unilateral DDH treated non-operatively, using the normal side in his study as the control group. He found that a widened teardrop in the dislocated hip, presumably because of abnormal joint reaction forces, correlated strongly with a poor outcome at skeletal maturity. The use of the teardrop unfortunately cannot be used to time interventions, as by the time it is widened the intervention window has been missed. It may be useful however to predict outcome.

Albinana also found that the age at which the hip reduction was achieved was the most important factor predicting outcome by Severin class at maturity, and that the earlier the reduction is achieved, the greater the remodeling capacity of the hip. Kalamchi however reports better results with waiting until patients are older than six months, as he feels this decreases the risk of avascular necrosis.

Although an early reduction is desirable this has to be offset against technical difficulties associated with operating on younger patients. Salter for example does not recommend performing an innominate osteotomy before the age of 18 months.

The most controversial timing of intervention is between six and eighteen months. It is becoming an increasingly popular approach and can identify residual soft tissue obstructions which accept no medial pooling of contrast when judging the reduction with radiographic evaluation may be all that is required to achieve a satisfactory reduction. Although it is common practice to use an intra-operative arthrogram, we believe that this is not an essential requirement, and that the reduction can be assessed without radio-opaque medium. Proponents of arthrography accept no medial pooling of contrast when judging the reduction position and can identify residual soft tissue obstructions which may impair the acetabular response after femoral head reduction. The potential risk of anaphylaxis and sepsis with this procedure are over-stated.

Non-operative treatment

Once identified at a baby check, a child with an unstable hip is often placed into double nappies whilst awaiting an ultrasound scan or referral to an orthopaedic surgeon. The idea of increased abduction is a logical one but with little scientific basis. The same may be said for children spending time prone whilst awake and being observed. In the light of sudden infant deaths, few if any would continue to recommend prone sleeping. These two activity modifications although unproven to be beneficial are unlikely to cause damage, so may in our opinion continue to be employed but with advice to parents that they are an adjunct rather than as the primary treatment modality.

Pavlik first described the use of his harness to the Czechoslovak Orthopaedic Society in Prague in 1946. He developed the harness because of disappointment at the high rates of avascular necrosis (AVN) with other methods of conservative treatment, predominantly using “passive mechanical treatment”. Pavlik felt that movement was essential to the treatment of dysplasia. With the hips flexed, infants are unable to keep the hips adducted because of fatigue. This positioning leads to a gradual, spontaneous and non-violent reduction of the dysplastic hip. Movement of the legs generates a cyclical loading of the acetabulum, stimulating remodeling.

The Pavlik harness is only suitable for hips that can be reduced on examination. An irreducible hip is a contraindication and an operative course of treatment should be initiated. The high rate of AVN that Pavlik observed was likely to be the result of forced abduction and/or flexion. Ramsey describes a “safe zone” when managing hips non-operatively. This is between the abduction and flexion necessary to maintain reduction, but not so far as to lead to interruption of the vascular supply of the femoral head.

Success rates with Pavlik harness treatment are as high as 95% with corresponding AVN rates as low as 0.3% if used early in the course of the disease. Other than AVN and occasional parental compliance issues, the only significant side effect of harness treatment is femoral nerve palsy associated with hyperflexion. This usual resolves on adjustment of the harness. It is easily detected because active knee extension is lost, a movement that the harness does not restrict.

Operative treatment

If a hip has failed to respond to non-operative measures, is irreducible, or is diagnosed late, operative interventions are indicated.

Closed reductions A hip manipulation under general anaesthesia with radiographic evaluation may be all that is required to achieve a satisfactory reduction. Although it is common practice to use an intra-operative arthrogram, we believe that this is not an essential requirement, and that the reduction can be assessed without radio-opaque medium. Proponents of arthrography accept no medial pooling of contrast when judging the reduction position and can identify residual soft tissue obstructions which may impair the acetabular response after femoral head reduction. The potential risk of anaphylaxis and sepsis with this procedure are over-stated.

In the thirty year follow up paper of closed reductions, Malvitz placed his patients in hip spicas with 90°—100° of flexion and 60° of abduction, with the cast down to the ankle on the affected side and to above the knee on the unaffected side, thus stabilising the pelvis. The spica was left on for twelve weeks. It is fairly common practice to change the spica after six to eight weeks, to account for growth and soiling. Attention has to be paid to the ‘safe zone’ mentioned earlier, and to performing an adductor tenotomy either percutaneously or open if this arc is insufficient.

Confirmation of reduction in the spica is usually achieved with either MRI or CT scans. CT scans provide a clear view of bony architecture, but involve ionizing radiation, whereas an MRI involves no radiation but often requires general anaesthesia in order for the child to be still enough to gain meaningful images.
The rate of AVN varies widely from 0 to 73%.25 The true rate clearly lies in between and depends very much on the diagnostic criteria and how rigorously and for how long they are sought. It is likely that the hips that go on to develop avascular necrosis are those that are dislocated rather than dislocatable.

**Open reductions** The requirement for open reductions has declined since hip screening programs have become more widespread. Failure to achieve a closed reduction by the methods described previously, or late presentations, may necessitate an open procedure. In the child under six months of age, our preference is for a medial approach. Over eighteen months a concomitant innominate osteotomy can be performed through an anterior approach. The treatment between six and eighteen months as mentioned above is more controversial. We tend to favour an anterior open reduction, often postponing the pelvic osteotomy until after eighteen months of age if the acetabular index is not improving towards the expected normal values.

The predominant structures that prevent reduction and need addressing at the time of open reduction, regardless of the approach used are the iliopsoas tendon, ligamentum teres, transverse acetabular ligament and pulvinar. The labrum and capsular infolding do not usually cause a block to reduction if the capsule is opened and later plicated to remove the “dead space” into which the hip can redislocate. A very deformed, inverted labrum can be brought out over the femoral head if radial cuts are inserted into it, without damaging the lateral acetabular epiphysis. The thickened ligamentum teres often requires to be excised, after its proximal end has been used to define the true acetabulum in the high dislocation. The transverse acetabular ligament and any other medial bands should be released prior to reduction, but the pulvinar (fat pad) is a useful structure and does not merit excision as it is not obstructive.

**Pelvic osteotomies** As discussed under hip pathophysiology, the primary pathology lies on the acetabular side of the joint, with secondary changes occurring on the femoral side, such as femoral head deformation and increased anteversion. It is for this reason that we believe the principal correction should also take place on the acetabular side.

There are numerous pelvic osteotomies described in the literature. Broadly speaking, they can be divided into two categories. There are those that rely on rotating the acetabulum around the open pubic symphysis, therefore not changing the volume of the joint but merely redirecting it. These osteotomies are used when a congruent reduction can be achieved and additional head coverage is required. The second group rotates the acetabulum about the tri-radiate cartilage, reducing the volume of the joint. These may be appropriate when there is an incongruence between the femoral head and acetabulum after reduction.

Our preferred osteotomy is the innominate osteotomy (Figures 3, 4 and 5) described by Salter. This yields predictable results as demonstrated by post-operative follow up to up to 45 years.26 Patients treated with an innominate osteotomy and open reduction had hip survival rates of 99% at thirty year follow up, 86% at forty year follow up and 54% at forty five year follow up. Two thirds of the surviving hips had little or no evidence of osteoarthritis.

The results of any author describing their own technique are almost universally better than those who try to replicate it. This however is not necessarily the case following the innominate osteotomy if the author’s original technical description is understood and adhered to.

As pelvic osteotomies rely on flexibility of either the tri-radiate cartilage or the pubic symphysis, there is obviously an age limit beyond which they should not be performed, and an adult pelvic acetabular osteotomies then considered more appropriate. We consider the upper age limit for reduction to be ten years in unilateral high dislocations and five years in bilateral, high dislocations. Bilateral dislocations treated with pelvic osteotomies should be staged to avoid creating a pelvic discontinuity.

**Femoral osteotomies** Studies trying to compare the outcomes of femoral osteotomies versus pelvic osteotomies have been undertaken, but attempting to glean meaningful conclusions from them is difficult, on account of the large number of variables.
Femoral osteotomies do have a place, but rarely, in our opinion, as the sole procedure. The commonest indication is a high dislocation that requires a femoral shortening osteotomy in order to reduce the hip without tension (Figures 6 and 7). The coronal alignment (varus/valgus) does not need much correction, and needs careful to be carefully controlled when adjusting femoral neck version.

**Sequelae**

Despite advances in our understanding of the pathophysiology and hence treatment of DDH, avascular necrosis of the femoral head remains a relatively common and serious complication. It is difficult to place a figure on the rate of avascular necrosis in DDH, not only as the treatment options are so varied, but also because diagnostic criteria for it differ, depending on author. This is likely to account for the wide range in quoted rates from 0–73%.

Kalamchi developed a classification system for avascular necrosis by reviewing 1072 patients treated for DDH, 119 of whom had avascular necrosis. He found that avascular necrosis affecting the ossific nucleus resulted in excellent long term function, whereas avascular necrosis affecting the proximal femoral physis was associated with an unpredictable but usually poor outcome. Kalamchi found the highest rate of avascular necrosis to be in those treated under six months of age. This however was in the pre-Pavlic era, when it was not uncommon to place a child in a hip spica without general anaesthesia. He correctly points out that “this is not to suggest a delay in initiating treatment, but rather a plea for extra care when treating infants”.

Ultimately the sequelae of DDH, whether affected by avascular necrosis or not, is arthritic change requiring total hip replacement. Although implant design and materials have advanced over the years, arthroplasty in the untreated DDH hip with no recognizable acetabulum or in one that is proximally placed with marked limb shortening remains a challenge. Long standing shortening is often only partially correctable, limited by soft tissue distraction tolerance. The acetabular component is often sub-optimally supported even with augmented implants or bone grafting procedures. The increase in arthroplasty options should not detract from solving the underlying problem.

**Conclusions**

The healthcare economics that surround hip dysplasia are extremely complex. However the morbidity spared the child and family as well as the potential savings for the country if a national strategy is appropriately implemented through resource availability could be substantial.

There is an enormous volume of literature written about DDH. What has become abundantly clear is that it is unlikely to be a single disease entity requiring a single simple solution. Early diagnosis is clearly beneficial to both the child and family, and to the wider community in terms of healthcare economics. What remain the greatest challenges in the management of DDH are interpretation of hip imaging, timing of the intervention and the treatment choices.

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**Figure 5** Radiograph demonstrating acetabular remodeling after innominate osteotomy.

**Figure 6** Radiograph of bilateral late presenting hip dislocations.

**Figure 7** Post operative radiograph in a hip spica following open reduction, innominate osteotomy and shortening femoral osteotomy. Note: Staging of bilateral procedures.
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