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Chapter 46.3.2 Developmental dysplasia of the hip

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Part of "Chapter 46.3 - Paediatric orthopaedics"

Developmental dysplasia of the hip

Developmental dysplasia of the hip encompasses a spectrum of abnormalities in which any or all of the components, including the capsule, the proximal femur, and the acetabulum, are defective. The condition manifests as subtle clicks to frank dislocation, which usually occurs at delivery or in the perinatal period. The etiology is believed to be related to laxity induced by the maternal hormone relaxin, which enables the cervix to dilate during labor. Though its exact role has yet to be determined, it is thought that relaxin crosses the placenta, presumably affecting the ligaments of the hip capsule. The incidence of developmental dysplasia of the hip in the United States is approximately 10 cases per 1000 live births, with a male to female ratio of 1:6. It has been estimated that developmental dysplasia of the hip is the cause of 20 to 50 per cent of degenerative arthritis of the hip in adults.

Normal hip development results from balanced growth of the acetabular, triradiate, and proximal femoral growth centers in response to a properly located femoral head. The goal of treatment of a dysplastic hip is thus the attainment and maintenance of a concentric reduction. The key to successful management is early treatment. During the first few days of life, and possibly longer, the hip can slip in and out of the acetabulum. When the femoral head is held reduced early, then, as the ligaments tighten, the hip remains concentrically located. If, however, the hip is left dislocated, reduction becomes more difficult as the ligaments tighten.

Additional factors are thought to play a part in the genesis of developmental dysplasia of the hip. Intra-uterine position is important, particularly with oligohydramnios and breech deliveries, when the femoral head may be levered out of the socket. Hyperextension has also been implicated. Our studies have shown that, even when the musculature is removed from the hip of a stillborn fetus, the joint remains stable. This is largely due to a negative intra-articular pressure; if air is introduced into the joint by a needle, the pressure equilibrates, and the hip is easily dislocated. Salter has demonstrated that hyperextension of a joint leads to breaking of the seal, allowing gas to enter. Thus, the old custom of slapping the newborn on the buttocks has been abandoned for fear of dislocating the hip.

At birth, most patients with developmental dysplasia of the hip have ‘dislocatable hips.’ If these are allowed to remain out of the socket for several weeks, they become dislocated, and Ortolani's sign becomes negative, as the hip is not easily reduced (Fig. 1). Some hips,

however, are dislocated at birth. These are often teratologic in origin, resulting, for example, from arthrogryposis or muscle imbalance. If the hip has been dislocated *in utero*, a false acetabulum may already be present at birth (Fig. 2). Closed treatment usually fails to reduce these hips.

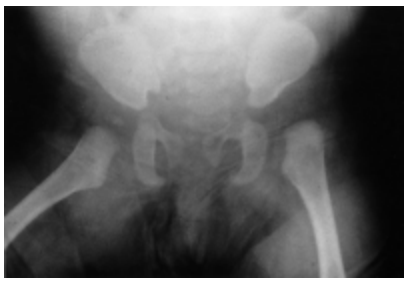


Fig. 1. An infant with dislocation of the left hip and subluxation of the right hip. The left hip has been out for several months. Note the early formation of a false acetabulum on the left.

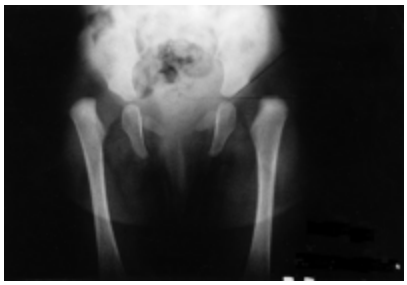
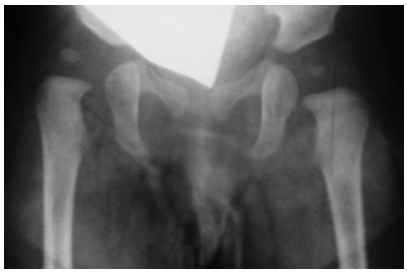


Fig. 2. Bilateral dislocations in an arthrogrypotic. There is almost no true acetabulum on either side. The false acetabulums were present before birth.

The subluxed hip represents another category of dysplasia. Unfortunately, this term has been used for widely disparate conditions. It

most appropriately designates a femoral head that is in the acetabulum, but not perfectly seated (Fig. 3). This condition is most commonly the sequela of an adducted position *in utero*, which persists after birth. The resulting limited abduction prevents the socket from developing well.

Fig. 3. In this radiograph, the right hip is subluxed. The femoral head is displaced laterally, but not dislocated. Note the elevated acetabular index.



Most orthopedic surgeons believe that, at least initially, the bony acetabulum is normal. In hips reduced around the time of birth, radiographs remain normal. If, however, the hip is permitted to remain dislocated, there will be bony changes visible on radiograph. Furthermore, infants with *in utero* dislocations frequently have flattened acetabulums at delivery.

In the newborn, the diagnosis is made with the Ortolani and Barlow maneuvres. The Ortolani maneuvre is a reduction performed by abducting the hip, with a resulting ‘clunk’ felt by the examiner. The patient is placed in the supine position. Both hips are flexed and adducted. With the index and middle fingers over the greater trochanter, the hip is slightly abducted while lifting the femoral head forward, into the socket. If the posterior acetabular lip is shallow, the click may be subtle; pressing the hip against the acetabulum during the reduction may accentuate it.

The Barlow maneuvre is used to determine if the hip is dislocatable. Again, the infant is placed supine. The untested hip is flexed to 90 degrees and mid-abduction. The tested hip is in 45 to 60 degrees of flexion and slight adduction. With the examiners hands in the same position as for the Ortolani, the femoral head is gently pressed posteriorly, in an attempt to dislocate the hip. A positive test will be felt as a ‘clunk.’ On release of pressure, the hip will re-locate, with a second palpable ‘clunk.’ The test is then repeated for the opposite hip.

A ‘soft click’ is frequently felt during infancy and early childhood. In the past, this was thought to be due to a tendon, possibly the tensor fascia lata, snapping over the trochanter. Few orthopedists treated these patients, and most patients had no problems. With the increasing use of ultrasound, however, it has been suggested that these clicks actually represent partial dislocation, or subluxation over the cartilaginous limbus, not dislocation *per se*. While the exact significance of this finding is not yet known, some orthopedists treat these hips as if they were truly dislocatable. The approach taken by the senior author is to place these infants in a Pavlik harness (see ‘Treatment’, below) for 1 month, in the hope that the hip will tighten. If radiographs are normal at that point, treatment is discontinued and the patient is observed, although the ‘clicks,’ may persist for years.

In older children, the Barlow and Ortolani tests are negative. The main diagnostic sign is a lack of abduction. Testing both hips simultaneously can be misleading; the pelvis may rock, resulting in abduction appearing symmetric. The hips should be tested individually, with the opposite hip used to stabilize the pelvis by pushing posteriorly (Fig. 4(a) to (c)).

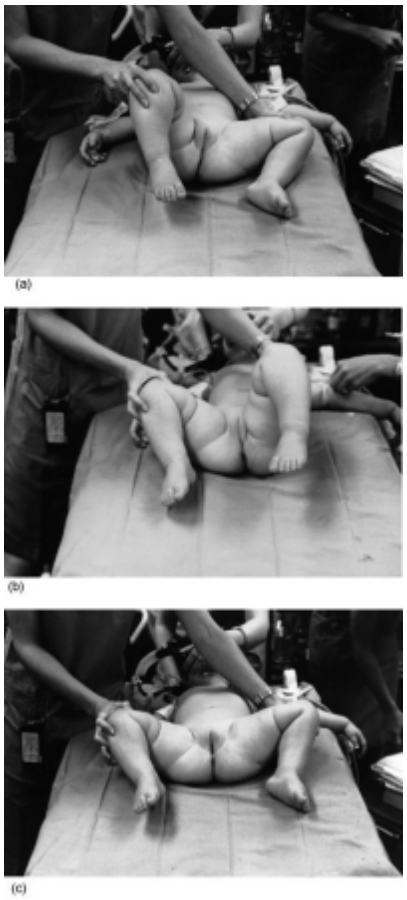


Fig. 4. Testing abduction in a child with a dislocated right hip. (a) With the right hip stabilized in neutral, the left hip is abducted. (b) The right hip demonstrates marked loss of abduction. (c) When the hips are tested simultaneously, the pelvis rocks, and abduction falsely appears symmetric.

Other physical findings include the Allis or Galleazi sign (Fig. 5) and asymmetry of the gluteal creases (Fig. 6). Galleazi's sign is elicited by examining the supine patient on a firm surface. The hips are flexed, and the buttocks are pressed posteriorly. The dislocated extremity will appear shorter than the other. However, in bilateral dislocations, these tests are unreliable. Abduction is equally limited, and leg lengths and gluteal folds may appear symmetric. A helpful finding in these cases is widening of the perineum. Normally, when the hips are adducted, the perineum is masked by the legs coming together. If the hips are both dislocated, the perineum is more exposed.

Fig. 5. A positive Galleazi sign. The leg with the dislocated left hip appears shorter than the right.

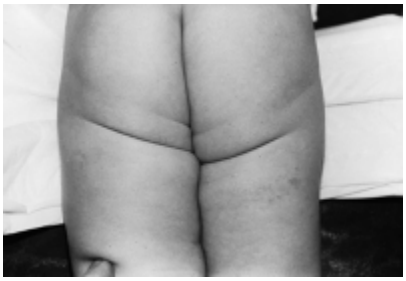


Fig. 6. Asymmetric gluteal folds in a child with a dislocated left hip.

The relationship of the femoral head to the acetabulum is used to diagnose dislocations radiographically. The head is not ossified until 3 months in girls, and until 6 months in boys. Therefore, lines are drawn to determine its position. Hilgenreiner's line is drawn through the tri-radiate cartilage of both acetabulums. Perkins' line is a vertical drawn through the lateral edge of the acetabulum. The femoral metaphysis should be equally inferior to Hilgenreiner's line and medial to Perkins' line (Fig. 7). Shenton's line is a smooth arc drawn along the medial femoral shaft and the superior border of the obterator foramen. Interruption of this line indicates that the femoral head is not concentrically seated in the acetabulum. In addition to these findings on plane radiographs, computed tomography and magnetic resonance imaging studies are occasionally used to establish the exact position of the hip. However, ultrasound is rapidly becoming the standard, as there is no radiation, and dynamic evaluation of the hip is easily performed.

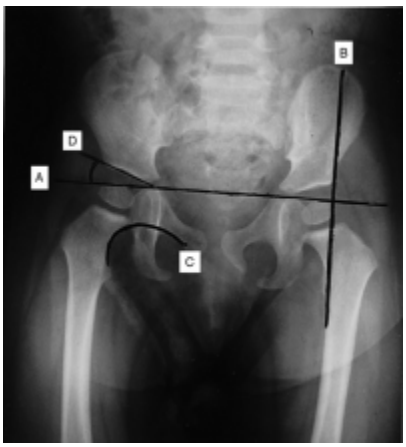


Fig. 7. Radiograph of a normal pelvis with illustration of Hilgenreiner's line (A), Perkins' line (B), Shenton's line (C), and the acetabular index (D).

On the frog-leg lateral view, the hip should point to the tri-radiate cartilage. A line drawn from the tri-radiate cartilage to the lateral edge of the acetabulum defines an angle with Hilgenreiner's line. This is the acetabular index, and is normally about 30 degrees. In the newborn, much of the acetabulum is cartilage, so slight tilting of the pelvis gives widely differing angles. Because the acetabulum is a growth plate, if the head does not make contact with it, there is no suppression of growth. When the hip has been dislocated for some time, the arc of the roof of the acetabulum becomes shallow, and the acetabular index increases above 30 degrees. If the hip remains dislocated, with the head articulating above the acetabulum, an indentation forms in the iliac wall, resulting in a ‘false acetabulum.’

Treatment

The Pavlik harness is the standard treatment for the dislocatable hip discovered at birth (Fig. 8). This device holds the hip reduced by

maintaining greater than 90° flexion and moderate abduction. While adduction is prevented, the harness should not be used to hold the hip in maximal abduction, which may cause avascular necrosis. The patient is seen weekly. Radiographs need not be repeated with each exam; progress may be followed with ultrasound.

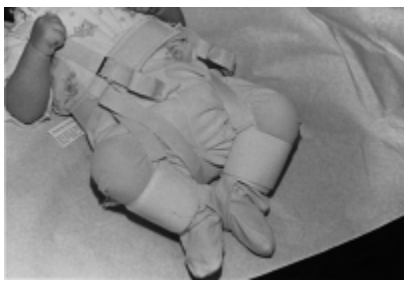


Fig. 8. An infant in a properly adjusted Pavlik harness.

Abduction improves quickly in subluxed hips; within a month, the harness may be switched to night use only. For dislocated hips, especially those that have been untreated for as long as 6 months, the harness is adjusted to hold the hip in even more flexion. The hip is prevented from adducting, and it is hoped that the child will kick the hip into position. The severe flexion should be reduced as soon as possible to prevent posterior dislocation.

If successful, the harness may be exchanged for an abduction splint after 6 weeks, although some physicians continue to use the harness for the entire 3-month treatment period. At this point, if the acetabulum is developing well, and the hip is no longer dislocatable, treatment may be stopped. When the diagnosis is made in the newborn, treatment in the harness is successful in 85 to 95 per cent of patients. If the hip has not reduced by 3 to 4 weeks, or if the hip still dislocates at 6 weeks, the Pavlik harness is

considered to have failed.

The senior author's preference for late dislocations, from 6 months to 1 year, is traction, adductor tenotomy, and spica cast application. This method is tried earlier if reduction is not obtained with the harness. The goal is to obtain gentle closed reduction, as forced reduction frequently leads to avascular necrosis. Various forms of traction may be employed. All are applied with the hip in a flexed position to relax the iliopsoas and prevent it from lying across the acetabulum and blocking reduction. Furthermore, experimental evidence suggests that blood flow to the hip is improved in the flexed position. Though one approach is to use Bryant's traction, with both hips pulled straight up, we prefer Buck's skin traction, with the addition of bolsters under the thighs, to relax the hamstrings at the same time that the hips are flexed. Increasingly, traction has been successfully utilized in a home program.

Two or 3 weeks are usually required for traction to be effective. Radiographs should demonstrate that the hip is being pulled down to the level of the acetabulum. Once the head is opposite the socket, or when it is easily reducible, a subcutaneous adductor tenotomy is performed. While some have abandoned tenotomy, it is useful for relieving pressure on the head, minimizing the risk of avascular necrosis. An arthrogram is performed to ensure the hip is reduced and well seated, without a soft tissue block to reduction (Fig. 9).

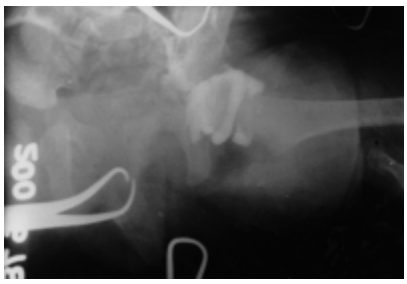


Fig. 9. An arthrogram of a hip with interposed soft tissue blocking full reduction, demonstrated by an increased dye space.

The hip is best seated in 90° or more flexion, and abducted 10° to 20° past the point of reduction. Over-abduction stretches the medial circumflex vessels, increasing the risk of avascular necrosis. As the fat and muscles atrophy during cast immobilization, the hip becomes prone to posterior dislocation. To prevent this, the surgeon must push anteriorly on the trochanter, and the cast should be molded

accordingly. Malvitz and Weintstein (1994) found that functional results have been excellent at up to 30 years follow-up when traction and adductor tenotomy are used in concert. The frequency of degenerative changes is markedly decreased compared with hips treated without tenotomy. Stromqvist and Sunden (1989) reported avascular necrosis in fewer than 5 per cent of patients treated between 2 and 12 months of age. They reported

good results in 97.5 per cent of patients at 2 to 11 years of follow-up.

When closed reduction cannot be obtained, an open procedure becomes necessary. This is usually the case in teratologic dislocations, those in children over 1 year of age, as well as in early dislocations that prove irreducible, or that have a soft tissue block to concentric seating. The medial approach allows release of the often-obstructing transverse acetabular ligament. While some reported series show a high risk of avascular necrosis with this approach, it is still utilized in many parts of the United States. We prefer an anterior Smith-Peterson approach, between the sartorius and tensor, elevating both heads of the rectus femoris, and cutting the psoas tendon to permit reduction. This facilitates a capsulorraphy, which holds the head better reduced. The capsule is opened with a 'T' incision, and the ligamentum teres is used as a guide to the true acetabulum. With the head reduced, the capsule is imbricated to secure the reduction (Fig. 10).

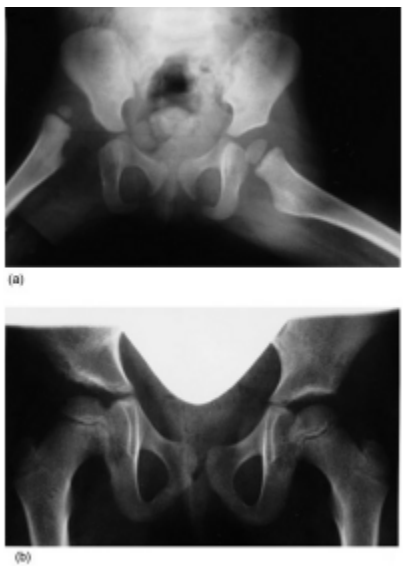


Fig. 10. (a) Unilateral right hip dislocation in an 18-month-old child. The acetabulum is minimally developed, and the femoral head is developmentally delayed. (b) The same child at 5 years of age, after an open reduction and capsulorraphy.

In children over 3 years of age, traction has been forgone in favor of femoral shortening, which is thought to reduce pressure on the head by functionally lengthening the soft tissues. In 1984, Schoenecker and Strecker reported a 54 per cent incidence of avascular necrosis and 31 per cent redislocation rate in children in this age group treated with traction and open reduction, compared with no avascular necrosis and 8 per cent redislocation in those who underwent a femoral osteotomy and open reduction. The cut is made just distal to the greater trochanter, and the hip is then reduced. A small dynamic compression plate is then used to hold the osteotomy. A spica cast is placed for 3 months. Usually, further bracing is not needed, although some prefer to employ it until there is evidence of acetabular deepening. In pathologically lax patients, however, even a well-reduced hip may later have a widened teardrop-head distance. Prolonged bracing is recommended in this group of patients.

For hips that have been dislocated for more than 18 months, there may not be sufficient growth potential for the acetabulum to become normal. The pathology dictates the appropriate procedure. A hip in excessive valgus is treated by a varus osteotomy, ducking

the head into the acetabulum. For a shallow acetabulum with a normal femoral head and neck, an acetabuloplasty is performed. When both aspects of the hip are abnormal, both types of procedures are performed.

A common error is made in children with marked anteversion. These hips appear to be well-seated on internal rotation radiographs, although not in neutral or external rotation. Often, the surgeon will perform an external rotation osteotomy, with the intent of forcing the child to hold the hip internally rotated, and, thus, reduced. This does not occur. The correct procedure is to perform a capsulotomy, internally rotate, and seat the hip, then imbricate the capsule, ensuring maintenance of the reduction. A distal femoral osteotomy is then performed, externally rotating the distal segment.

There are multiple osteotomies that may be employed to change the orientation of a dysplastic acetabulum, thereby increasing the coverage of the femoral head. A Salter osteotomy of the innominate bone is generally the senior author's preference. A Gigli saw is placed in the sciatic notch, and a cut is made to a point just superior to the anterior inferior iliac spine. The inferior fragment is then rotated forward, and a wedge of bone from the anterior superior iliac spine is held between the fragments with Kirschner wires or threaded pins (Fig. 11). Salter and Dubos (1974) reported excellent results in 93.6 per cent of their patients over 15 years, and Gulman et al. (1994) had similar (88.4 per cent good to excellent) outcomes.

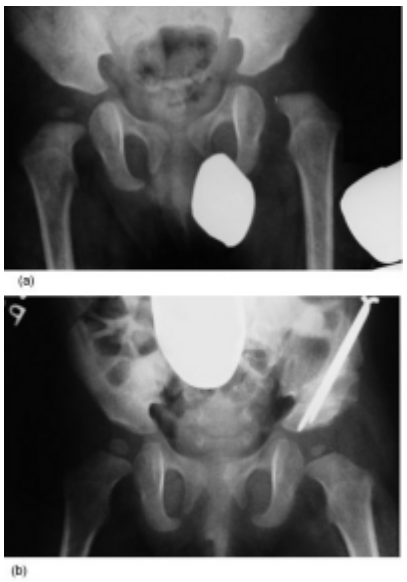


Fig. 11. (a). This child's dislocated left hip was initially treated in excessive abduction at another institution. Her hip remains subluxed, and avascular necrosis has developed. (b) The same child, after open reduction and Salter osteotomy. Note the normalized acetabular index and the healed bone graft, taken from the ipsilateral ilium.

In older children, the pubic symphysis upon which the osteotomized segment hinges is no longer supple. A Sutherland osteotomy, which cuts through the superior and inferior pubic rami just lateral to the symphysis, or a Steele triple osteotomy, with cuts through both the pubis and the ischium (Fig. 12), may then be employed. Osteotomies close to the acetabulum, such as the Dial and Wagner osteotomies, allow the acetabulum to rotate freely, but are technically very difficult, and are rarely necessary.

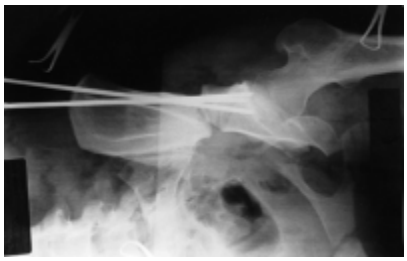


Fig. 12. Intra-operative radiograph of a Steele triple osteotomy.

Dislocated hips can generally be reduced up to 5 years of age. After this, results are poor. Bilateral dislocations are usually left in place after this point. These hips are usually quite symmetric; treatment frequently results in failure on one side, with resulting asymmetry.

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In contrast, even in older children, an attempt is usually made to reduce a unilateral dislocation, to provide symmetry.

Further reading

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