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Evaluation of Dysplasia of the Hip (Children with DDH, Adolescents, and Adults)

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Developmental dysplasia of the hip (DDH) is a congenital anomaly, characterized by insufficient development of coxofemoral joints and concomitant laxity of the capsule-binding system, which may result in partial or total loss of coxofemoral joint relationships [1, 2].

It is the most common osteoarticular deformity, presents a predilection for female sex with a female/male ratio of 61. The incidence varies between 0.7% and 2.5%, and it is very common in the north Italy, France, Spain, and the Balkans where the incidence stands at 2–4%. Incidence is significantly lower in African and Asian patients [3, 4]. In 45% of cases, it is bilateral and associated with other congenital malformations such as clubfoot. DDH often leads to early degenerative hip arthritis if not treated [5].

37.1 Clinical Presentation and Diagnosis of DDH in Newborns

In newborns, clinical hip examination is essential for the diagnosis of DDH: semeiological maneuvers such as Ortolani and Barlow tests are still considered very useful [1]. Starting from the age of 8–12 weeks of life, these maneuvers are not easily assessed and not very indicative.

Currently, the instrumental investigation, which allows an accurate and early diagnosis, is based on ultrasound, which has been widely used in recent years and which has supplanted the use conventional radiological examination of (Fig. 37.1). The ultrasound has proven to be an accurate method of diagnosis in the first months of life, allowing to highlight all the anatomical structures of the baby's hip. Among the various ultrasound classifications, the Graf classification is certainly the most widespread in Europe and allows to classify the degree of severity of the pathology.

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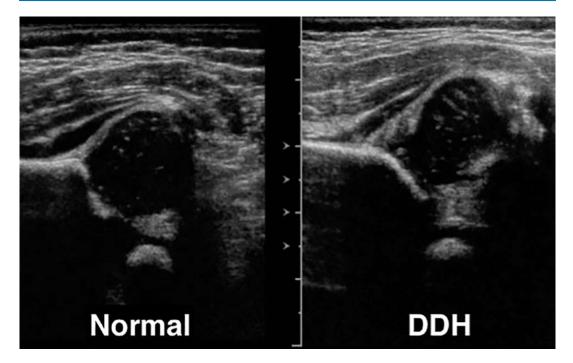


Fig. 37.1 Normal and DDH at ultrasound examination

37.2 Clinical Presentation and Diagnosis of DDH in Adolescents and Adults

The most common presenting symptoms of acetabular dysplasia in the skeletally mature individual are insidious onset of activity-related groin pain and/or lateral hip pain. Diagnosis is made through physical exam and radiographic findings [5]. Patients with hip dysplasia may initially present with mild peritrochanteric pain reflecting abductor fatigue, an almost universal finding in pre-arthritic hip dysplasia in the mature adult. Commonly, patients with dysplasia will have groin pain in the affected hip that worsens with activity. Pain worsening with weight-bearing and activity may indicate underlying joint pathology. Pain with flexion positions or prolonged periods of sitting may commonly be associated with femoroacetabular impingement (FAI). The sensation of locking or catching in the affected joint may indicate intra-articular mechanical problems, such as labral tear, chondral flap, or loose body. The physical examination of the patient with suspected hip dysplasia should also take into account general condition and body habitus, patient's sitting posture, and gait pattern. It is also important to assess for a leg-length discrepancy. Standing measurements will allow an assessment of pelvic balance and therefore are considered the best way to assess a leg-length discrepancy. Resting lower extremity rotation should be assessed with the patient lying supine on the exam table. Normal lower extremity rotation is between 10° and 30° of external rotation. Abnormal rotation may result from abnormal acetabular version, abnormal femoral version, or femoral head-neck abnormalities. Hip range of motion testing is essential for not only diagnosis but also pre-procedure planning. The examiner should steady the pelvis with one hand while performing range-of-motion testing, including hip flexion and internal and external rotation both in 90° of flexion and full extension.

In contrast to classic FAI, which may demonstrate restricted hip flexion and internal rotation, patients with classic dysplasia will generally demonstrate normal hip flexion and normal internal rotation.

Some special tests may help elucidate the nature of the hip pathology. The anterior impingement test is a special maneuver that is indicative of disorders of the anterior acetabular rim. It is very sensitive for a range of anterior hip lesions, including labral tears and rim fractures, but fairly nonspecific for intra-articular disease and joint irritability. The patient is placed supine, and the affected extremity is passively flexed, adducted, and internally rotated. The test is considered positive if this maneuver reproduces the patient's pain. The apprehension test is another special maneuver that is useful to assess for anterior hip instability, though it may be positive in patients with labral lesions as well. It is performed by having the patient lie supine on the table. The affected extremity is extended and rapidly externally rotated. The test is considered positive if it elicits apprehension or anterior hip pain. Finally, patients should be assessed for hip abductor weakness. The Trendelenburg sign is elicited if the patient leans away from the affected extremity during single-legged stance on the affected extremity. Alternatively, abductor strength can be tested by having the patient lie on his/her unaffected side and asking him/her to abduct the leg against resistance.

The evaluation of an adult patient with suspected hip dysplasia is confirmed with imaging [6, 7]. The radiographic features of adult hip dysplasia may range from subtle acetabular dysplasia to complete dislocation of the femoral head from the native acetabulum. The goals of imaging are to assess the structural anatomy of the hip, determine the congruency of the articulation, examine the integrity of the joint space, and assess the soft tissues.

The anterior-posterior (AP) view of the pelvis is the single most important view for defining acetabular coverage of the femoral head, femoral head sphericity, contour of the femoral head-neck junction, height of the greater trochanter, position of the joint center, joint space, and Shenton's line (Fig. 37.2).

An acetabular undercoverage is defined by a *lateral center edge angle* minor than 22° and/or



Fig. 37.2 A-P view of DDH

an *acetabular index* >14° and/or a *femoral head extrusion index* >27%. Another feature of DDH is the internal rotation of the entire innominate bone. DDH presents also a decreased size of the lunate surface compared to normal hips and increased contact pressures (e.g., 23% increased pressure in the midstance phase of gait).

Pelvic AP X-rays allow evaluation of acetabular version: acetabular version refers to the orientation of the mouth of the acetabulum with respect to the sagittal plane. If the mouth faces forward, anteversion is present, but if the mouth faces posteriorly, retroversion is present.

In normal hips, the acetabulum usually is anteverted. Dysplastic hips are globally shallow, but the deficiency in coverage usually is more pronounced anterolaterally, thereby giving an impression of excessive anteversion. If the predominant deficiency occurs in the posterolateral wall of the acetabulum, the latter will seem retroverted. On an AP radiograph, it will be found that the posterior wall of the acetabulum meets the roof at a point medial to the junction of the anterior wall with the roof. This pattern is well recognized in certain types of dysplasias such as neuromuscular hip dysplasia, post-traumatic dysplasia, and proximal focal femoral deficiency.

Dysplastic hips are often anteverted, but DDH may also be associated with retroversion. In all cases of developmental hip dysplasia, the diagnosis of retroversion depends on the relationship of the anterior and posterior walls in the superior 1/3 of the acetabulum [8, 9].

Lateral radiographs allow better definition of the osseous anatomy of the proximal femur, anterior and posterior joint spaces, and acetabular rim. Lateral radiographs include the cross-table lateral and frog lateral. Also among the lateral views is the false profile view. This image is obtained by having the patient stand with their foot parallel to the radiographic plate and their pelvis rotated 65° relative to the film. This view is a true lateral view of the acetabulum. It allows measurement of the anterior coverage of the acetabulum and may also allow better detection of the degenerative changes that tend to begin at the anterior aspect of the joint.

Computer tomography (CT) is a complementary study in evaluating hip dysplasia when dysplastic signs have been recognized on X-rays and surgical correction is proposed. It allows reliable measurements of acetabular coverage, femoral neck anteversion, and appearance and position of the femoral head. It also allows better characterization of osseous impingement lesions.

MRI and MRI arthrogram are useful adjuvants to evaluate the labrum and should be obtained in patients with mechanical symptoms. A reactive labral hypertrophy may be detected on MRI or arthro-MRI resulting from an increased load (Fig. 37.3). The labrum often is torn along with a part of adjacent cartilage due to subluxation of the femoral head that tears the labrum from the acetabular rim together with a sleeve of cartilage.



Fig. 37.3 MRI of hip dysplasia resulting in labrum hypertrophy

Furthermore, on MRI, a progressive thinning of the acetabular cartilage may appear in early adulthood and may result in full-thickness defects at the peripheral acetabular rim due to static overload. This chondrolabral damage is typically located superiorly.

37.3 Etiopathogenesis

It is multifactorial:

- Acetabular dysplasia with exiguous development of the cavity and that consequently escapes the acetabular roof
- · Hyperlaxity of joint capsule structures
- Straight dysplasia: derived from muscle tensions with prevalence of the tone of flexoradductor muscle with respect to the abductor projectors
- Response of the development tissue to possible hormonal tubes
- Congenital syndromes such as Larsen, Ehlers-Danlos, and Down or neuromuscular disorders such as myelodysplasia or spina bifida
- Mechanical causes including the position taken by hips during intrauterine life, type of delivery (breeding presentation), or oligohydramnios

At birth, the components of the hip are largely made by cartilage. As for the femur, the proximal portion, epiphysis, and part of the neck are cartilage for the first 3 months of life. The ossification nucleus appears from the third to the sixth month and is settled to the femoral metaphysis at an age ranging from 11 to 16 years.

In the newborn, the angle of inclination, the angle between the axis of the neck of the femur and axis of the diaphysis, is greater than that of the adult, with $130-135^{\circ}$ in the newborn compared to 125 of the adult.

Also, the angle of declination, the angle that the axis of the neck of the femur forms with a frontal plan passing for the femoral condyles, appears wider: 35° of neonatal anteversion of the neck of the femur, $15/20^{\circ}$ in the adult. The first classification is established according to the time of diagnosis:

- From birth until 10–12 months (age of the walk) can have cases of preluxation or, rarer, embryo dislocation.
- From the beginning of the verticalization to the next development, the dysplastic hip may show itself to be dislocated or subluxed.
- From the end of the accretion, an inveterate dislocation is established.

37.4 Preluxation

At this early stage, acetabular cavity has an exiguous development, elusive acetabular roof, joint capsule laxity, and initial formation of a neolimbus (semicircular saliency located in the upper region of acetabulum). Histologically, ossification will be slowed (hypoplasia and increased obliquity of the bone roof), and there will be acetabular cartilage dysplasia with irregular chondrocytes and laxity of extracellular matrix.

37.5 Subluxation

In this stage, the femoral head moves upward from socket, anteverted coxa valga, pulvinar hypertrophy, joint capsule distension, and increased muscle tone with retraction of pelvitrochanteric muscle, adductors, and iliopsoas.

37.6 Dislocation

The femoral head exceeds the acetabular edge with neo-cotyle formation; the head is deformed and the neck becomes valgus and anteverted. A hypoplasia of the ossification nucleus is observed.

37.7 Inveterate Dislocation

Condition that occurs after the fourth–fifth years of life in the absence of treatment, with accentuation of the characteristics of dislocation and increased evidence of the neo-cotyle. Diagnosis of DDH should be early and oriented by anamnestic news such as familiarity, ethnicity, and complications of gestation, but also by clinical finding of probability represented by the asymmetry of the skin, hypotrophy and slight shortening of the preluxated limb, tendency for extra rotation, slight flattening of the buttock, and limitation of the abduction.

In suspected case, clinical diagnosis is carried out with clinical maneuvers; just remember that a dysplastic hip is a non-luxated hip yet, but can be easily dislocated.

37.8 Ortolani Test (Reduction Test)

With the child in the supine position, the examiner puts his/her hands on the newborn's thighs and applies a slight pressure at the great trochanter while the thumb is on the thighs. It abducts and extra-rotates with the knees and hips in flexion, and a reduction of femoral head is warned with a click. The test is positive when the hip is subluxated.

37.9 Barlow Test (Dislocation Test)

With the child in the upper position and thighs flexed at the right angle from the pelvis, grab the knees with the palm of the hand and lay the thumbs on the middle legs and side at the great trochanters. By adjusting the thighs and bringing them to the midline, a slight pressure is applied on the knee, leading the force in the front-rear direction. As a forced maneuver, there is a risk of damage to structures that are already intrinsically unstable, so the Ortolani test is usually preferred.

37.10 Trelat Sign

Patient in prone position with the knees flexed at 90° shows major intra-rotation in the sliced hip.

37.11 Savariaud Sign

The hypometria of the limb affected during the transition from the supine position to the sitting with the knees extended.

Between the 10th and 12th months of life, the child starts walking and goes into a dislocation condition. When dysplasia has not been diagnosed properly on time, this represents the evolution of a preluxation or subluxation condition.

At this point, the deformity can be seen with extra rotation and shortening of the limb, major deficit of walking (Trendelenburg sign or gait anserine), muscle hypoplasia, and Galeazzi sign.

The Trendelenburg sign is shown in mono podalic stand caused by hypoplasia and deficiency of hip abductor (mainly medius gluteus). During walking, the Trendelenburg sign causes the so-called anserine gait.

The **Galeazzi sign** is the asymmetry of knees by observing the patient on the bed with the knees flexed at 90° . The knee is lower on the affected side due to the rear movement in the dysplastic hip.

37.12 The Instrumental Diagnosis (Ultrasound Scan and Radiography)

The ultrasound is certainly the elective test in the first 2–3 months of life for diagnosis and screening investigation since the proximal femoral epiphysis has not yet ossified. It is a noninvasive examination, and it is repeatable and extremely sensitive (so that it is possible to overestimate the pathology). It shows the soft parts and all components of the joint and allows the control of the evolution of the effects of treatment.

With the ultrasound, three lines are set:

- 1. A straight iliac line
- 2. The tip of the acetabular labrum
- 3. The transition from the os ilium to the triradiate cartilage

From these come two angles:

- Angle α : formed by the acetabular roof and the vertical cortex of the ilium in coronal plane. It is indicative of the depth bone portion of the acetabulum roof. Normal value is equal or greater than 60 degree.
- Angle β : formed by the vertical cortex of the ilium and the triangular labral fibrocartilage. It is an indication of the development of the cartilage part of acetabulum. A Graf β angle greater than 55° is abnormal.

Based on the ultrasound characteristics and values of angles, the proposed Graf classification divides the various stages of the disease into groups and subgroups, on which the treatment is based.

- 1. **Type I:** alpha angle >60° (normal) (*normal*, *fully mature hip; the acetabular rim is angular, and the acetabular cup is deep; the cartilaginous roof covers the femoral head*).
- 2. **Type II**:
 - **Type IIa**: alpha angle 50–59° (<3 months) (*physiologically immature at less than 3 months of age*).
 - **Type IIb**: alpha angle 50–59° (>3 months) (similar to IIa but in infants older than 3 months; the joint is dysplastic and requires treatment to prevent further deterioration and dislocation).
 - **Type IIc**: alpha angle 43–49°; beta angle 70–77° (*the hip socket is severely dysplastic and is close to decentering, but the cartilaginous roof still covers the femoral head*).
 - **Type II D**: alpha angle 43–49°; beta angle >77° (*similar to IIc but the hip is decentered; the cartilaginous roof bends cranially*).
- 3. **Type III**: alpha angle >43° (*dislocated femoral head with a shallow acetabulum*).
- Type IV: alpha angle <43° (dislocated femoral head with a severely shallow, dysplastic acetabulum with labrum interposed).

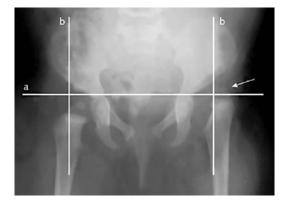


Fig. 37.4 A-P view with Hilgenreiner's and Perkin's line traced, showing the nucleus in the superior/external quadrant

The X-ray is performed after the first 4–5 months of life since the femoral head core is not ossified and therefore not visible. An A-P front projection of pelvis is performed.

We can obtain this by drawing three lines:

- (a) **Hilgenreiner's horizontal line**: traced through the superior aspect of both triradiate cartilages of the pelvis
- (b) **Two vertical lines of Perkin**: traced from the most lateral point of the acetabular perpendicular to the Hilgenreiner's line (Fig. 37.4)

The dysplastic hip has the femoral epiphysis in the superior/external quadrant rather than the inferior/medial quadrant, below the Hilgenreiner's line and medial to Perkin's line.

The whole radiographic signs, constituting the triad of putty, Fig. 37.1, enable DDH to be diagnosed: (1) Superolateral displacement of proximal femur; (2) Increase in acetabular angle; and (3) Small capital femoral epiphysis.

Distancing of the nucleus of the femoral epiphysis of the acetabular roof tends to lead to the superior/external quadrant, rather than being in the lower/internal quadrant of the Ombredanne diagrams, resulting in the interruption of the Shenton ogive (arc formed by the lower margin



Fig. 37.5 A-P view showing an increased acetabular angle and interruption of Shenton ogive

of the femoral metaphysis and the lower margin of the ileopubic branch).

Other X-ray alterations may be neo-cotyle formation; $\cos a$ valga (angle >130); and increased anteversion of the head of the femur (Fig. 37.5).

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