Developmental Dysplasia of the Hip

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ABSTRACT. *Objective.* The definition and early treatment of congenital dysplasia of the hip are controversial. The purpose of this study was to discuss the reasons for changing the acronym to developmental dysplasia of the hip (DDH) and to address its early detection and treatment.

Design. This multicenter study was designed to provide an updated assessment of the definition, pathologic anatomy, prevalence, etiology, natural history, early detection, and treatment of DDH.

Results. DDH more accurately describes the condition previously termed congenital dysplasia of the hip. The disorder is not always present at birth (congenital) and an infant may have a normal neonatal hip screening examination and subsequently develop a dysplastic or dislocated hip. Developmental dysplasia encompasses the wide spectrum of hip problems seen in infants and children. Physicians should understand that a normal neonatal screening examination does not assure normal hip development. The diagnosis of developmental dysplasia is made by physical examination. The Ortolani and Barlow maneuvers were designed to detect a subluxatable, dislocatable, or dislocated hip in the neonatal period. In the older child, limited abduction becomes a more reliable sign. The examination is variable depending on the type of dysplasia and changes with growth. The ultrasound is proving to be a sensitive tool in confirming the diagnosis in newborns and infants from birth to 4 months of age. The ultrasound is also valuable in older infants in terms of documenting that the dysplasia is responding to treatment. However, the ultrasound depends on an experienced sonographer and, in some cases, may be too sensitive, resulting in overtreatment. After 3 to 4 months of age, an anteroposterior pelvis radiograph can confirm the diagnosis.

Conclusions. All newborns should have a neonatal hip screening physical examination. After screening, the hips should be re-examined during health examination visits at 2 weeks, 2 months, 4 months, 6 months, 9 months, and 1 year of age. If any question arises during these visits or if there are associated risk factors, we recommend an ultrasound if the infant is <4 months of age or an anteroposterior pelvis radiograph if >4 months of age. *Pediatrics* 1994;94:201–208; *developmental dysplasia of the hip, congenital dislocation of the hip, ultrasound for hip instability.*

ABBREVIATIONS. CDH, congenital dislocation of the hip; DDH, developmental dysplasia of the hip.

The acronym congenital dysplasia of the hip (CDH) is confusing and has been used synonymously with congenital dislocation or congenital disease of the hip. This lack of precision in terminology is understood by experienced physicians who mentally define CDH according to the context in which it is used, but this abbreviation is confusing and misleading to patients, attorneys, and juries. A dislocated hip is a physical sign, not a diagnosis, and the term congenital means present at birth. Because CDH was thought to be present at birth, physicians were taught that if the neonatal examination was normal, the hip would develop normally. This is clearly not the case as several reports have documented dislocation or dysplasia of the hip occurring after a normal neonatal screening examination.[1-6] We believe the acronym developmental dysplasia of the hip (DDH) should replace CDH. The word "developmental" invokes the dimension of time acknowledging that the dysplasia or dislocation may occur before or after birth. Dysplasia means an abnormality of development and encompasses a wide spectrum of hip problems. By changing from CDH to DDH, the acronym will no longer falsely characterize a disorder in instances when the condition being described is neither congenital nor dislocated. To understand the spectrum of hip problems included in DDH, a knowledge of the embryology of the hip joint is beneficial.

METHODS

Embryology

In the embryo the limb buds first appear at 4 weeks gestation. The hip joint begins to develop at 8 weeks gestation when a cleft occurs between the acetabulum and the femoral head and development is usually complete by 11 weeks.[7] As a result, the hip is always located early in the embryologic stage because it forms from the pelvis. Hip dysplasia or dislocation may then occur in utero, perinatally, or subsequently with development. Moving from the fetal position of hip flexion to one of extension as occurs after birth increases the susceptibility of dislocation.

Definitions

DDH includes hips that are unstable, malformed, subluxated, or dislocated. Instability is the inability of the hip to resist an externally applied force without developing a subluxation or dislocation. A malformation includes any abnormality in the development of the femur and/or acetabulum. A subluxation is an incomplete dislocation with some residual contact between the femoral head and acetabulum, and a dislocation indicates complete displacement of the femoral head from the acetabulum.

typical dislocation of the hip.[8] A teratologic dislocation occurs early in utero and is associated with other malformations such as chromosomal abnormalities and neuromuscular disorders. A typical dislocation occurs in an otherwise healthy infant and may occur in utero, at birth, or after birth.

Pathologic Anatomy

Because DDH includes a wide spectrum of hip problems, the pathologic anatomy is dependent on the type, grade, and duration of the dislocation. In the neonatal period, there may be sufficient ligamentous laxity that the hip spontaneously dislocates and reduces. The newborn may dislocate and reduce the hip with kicking movements of the lower extremity. In utero the hip is in a position of flexion and abduction that results in a tightened iliopsoas tendon and anterolateral orientation of the acetabulum.[9] The tight iliopsoas, which runs across the front of the hip, may push the femoral head out posteriorly with hip extension during kicking or in swaddling the newborn. The labrum (cartilaginous rim of the acetabulum) becomes everted and flattened. If the hip spontaneously reduces within a few days, subsequent development is usually normal.

In contrast, if a subluxation or dislocation persists, then over time changes begin to develop. The femoral head becomes flattened on the posteromedial surface and femoral anteversion (internal torsion) gradually increases. Because the femoral head is not concentrically reduced, the acetabulum becomes shallow and dysplastic. The hip capsule thickens and the fibrocartilaginous labrum hypertrophies. The space between the acetabulum and femoral head is filled with fibrofatty tissue (pulvinar) that develops in the base of the acetabulum and a thickened ligamentum teres. As the femoral head moves further superolaterally, the inferior capsule is pulled upward over the lower part of the acetabular socket. The iliopsoas tendon that crosses the anterior capsule constricts it anteriorly causing the hourglass deformity. Once the capsular constriction becomes narrower than the diameter of the femoral head, nonoperative reduction becomes impossible. The fascia and muscles around the hip joint respond to the superior displacement by becoming shortened and contracted. The tight adductor muscles resist abduction and the flexor muscles hold the femoral head in the superior position making reduction difficult.

Prevalence

The prevalence of DDH varies according to racial and geographic parameters and is greater in whites than blacks. Edelstein[10] reported that he did not find one case among 16 678 African Bantu infants. The exact prevalence is controversial as some studies included only children with dislocated hips whereas others included dislocated, subluxated, and unstable hips. Barlow[11] reported that 1 newborn in 67 was born with instability of one or both hips. When these newborns were re-examined 1 week later 58% were stable, and by 2 months 90% were stable. This study emphasizes that the age at the time of examination is an important factor in reporting the prevalence of DDH.

The value of the neonatal hip screening examination remains controversial. Barlow[11] believed that widespread screening would eliminate the problem. Other investigators have identified patients who were negatively screened then later developed dysplasia or dislocation.[1-6] It now seems clear that the disorder is not always detectable at birth. In 1971, Walker[12] reported on seven children who initially had normal examinations and then subsequently developed dislocated hips. Tredwell and Bell[13] evaluated 32 480 newborns and discovered five who had normal neonatal screening examinations and then were found to have an adduction contracture with radiographic evidence of acetabular dysplasia between 2 and 8 months of age. Bialik et al[4] reported on 12 891 newborns and identified 36 who had a normal neonatal screening examination and later were found to have DDH. Davies and Walker[14] reported on 10 children who had clinically stable hips at birth but radiographs were obtained at one month of age because they had risk factors associated with DDH. In all cases, one or both hips had evidence of dysplasia on the radiographs and although clinical abnormalities were slow to appear, four hips subsequently dislocated. These studies document that, despite a negative neonatal screening examination, the hip may not develop normally. This emphasizes the importance of sequential hip examinations during health examination visits at 2 weeks, 2 months, 4 months,

Etiology

Stanisavljevic[15] reported that there are three periods during which the fetal hip is at risk for dislocation: (1) the 12th week, (2) the 18th week, and (3) the final 4 weeks of gestation. The first critical period is after the hip joint develops from the pelvis. As the limb rotates medially, the hip is at risk for dislocation. If the hip dislocates at this stage, all of the elements of the hip joint develop abnormally. The acetabulum is shallow, the capsule distends, the femoral head and greater trochanter remain small, and a false acetabulum develops. This type of dislocation is the most severe form of DDH and correlates with a teratologic dislocation. The second period is during the 18th week of gestation when the hip musculature develops. If there is an abnormality of neuromuscular development, a paralytic dislocation may occur at this stage. The third period is during the last 4 weeks of gestation when abnormal mechanical forces, such as a breech position or oligohydramnios, may predispose to DDH. In addition to these three periods, the hip may be normal at birth and then dislocate during the fourth critical period, the postnatal period.[4,12-14]

Factors associated with DDH include ligamentous laxity, pre- and postnatal positioning, genetic factors, and environmental factors. The maternal hormone relaxin, which contributes to ligamentous laxity in the mother's pelvis during childbirth, passes freely through the placenta into the newborn. Relaxin and other maternal hormones contribute to neonatal ligamentous laxity (especially in the female fetus) and may increase the risk of DDH. Wilkinson[16] demonstrated that the hip of a female rabbit would dislocate if the knee was splinted in extension and the animal was given an injection of progesterone. This did not occur in male rabbits when the same procedure was performed. He concluded that there is a combination of hormonally induced ligamentous laxity associated with knee extension that predisposes females to a higher incidence of DDH.

Muller and Seddon[17] reported that 16% of infants with DDH had a breech presentation compared with 3% in the general population. Hip flexion and knee extension as is seen in the frank breech presentation tightens the hamstring muscles and may contribute to DDH. Michelsson and Langenskiöld[18] were able to produce DDH in young rabbits by immobilizing the knee in extension, however, when the hamstring muscles were sectioned before immobilizing the knee, DDH did not develop. Oligohydramnios, which limits intrauterine space, is associated with mechanical deformities including torticollis, clubfoot, scoliosis, plagiocephaly, and DDH.

The left side is involved three times as often as the right in DDH. The majority of newborns were positioned in the left occiput anterior position at the time of birth. In this position, the left hip lies posteriorly against the mother's spine and may predispose the left hip to a higher incidence of the disorder.

DDH is associated with congenital muscular torticollis and metatarsus adductus. Iwahara and Ikeda[19] reported that 15% of their patients with congenital muscular torticollis had DDH and Hummer and MacEwen[20] found a 20% association. Jacobs[21] found that 10% of newborns with metatarsus adductus had DDH but subsequent studies have reported a lower incidence.[20,22] Kollmer et al[23] studied 7356 infants with metatarsus adductus and were unable to demonstrate any association with DDH.

There is evidence that genetic factors play a role in the etiology of DDH. Wynne-Davies[24] studied 589 patients and their families, and reported the risk to subsequent members of a family when a dislocation occurs is as follows: 1) healthy parents with one child affected, risk to subsequent children 6%; 2) one affected parent, risk 12% and 3) one affected parent with one affected child, risk 36%.

There is a correlation between postnatal positioning and DDH. The incidence is lower in India where newborns are carried astride the waist of the mother with the hips in an attitude of flexion and abduction. This is a stable position for most hips and most treatment devices attempt to duplicate this position. In contrast, the incidence is high in American Indians who position their newborns on a cradle board with the hips swaddled in extension and adduction. Kutlu et al[25] evaluated 4173 infants and found DDH in 56, and 55 of these had been swaddled in infancy for an average of 45 days. Green and Griffin[26] reported on 18 infants who had DDH in conjunction with an abduction contracture of the contralateral

plastic hip and stretching of the abduction contracture of the contralateral hip.

Several early investigators postulated that the etiology of DDH was secondary to primary acetabular dysplasia.[24,27] Subsequent reports have demonstrated that the acetabular dysplasia is not the cause of the dislocation but is a result of the femoral head not being concentrically reduced in the acetabulum.[28–31] The acetabular dysplasia is reversible by restoring a normal relationship between the acetabulum and femoral head.[32,33] The femoral head needs to be in contact with the triradiate cartilage for normal acetabular development.

Natural History

The natural history of DDH remains controversial as newborn instability may spontaneously resolve, develop progressive subluxation, or progress to complete dislocation. McKinnon et al[34] studied 15 149 newborns and found subluxatable hips in 92. The patient profile of the newborns with subluxatable hips was similar to those with dislocated hips with an increased frequency of females, breech presentations, left-sided involvement, and associated postural deformities. They concluded that the newborn subluxatable hip arises from the same antecedents as a frank dislocation and is best managed if detected and treated early.

Cooperman et al[35] studied 20 adults (32 hips) with acetabular dysplasia to determine the natural history of the disorder. After an average follow-up of 22 years, 30 (94%) had severe or moderate osteoarthritis. Wedge and Wasylenko[36] reported on 54 adults (80 hips) who had previously been affected by dysplasia, subluxation, or dislocation. At follow-up, only 41% of dislocated hips and 42% of subluxated and dysplastic hips received a good score. In contrast to the dislocated group, the patients with subluxation and dysplasia tended to be younger and had more pain.

History

The previously held view, that if the newborn was examined at birth all dislocated hips would be diagnosable, is both outdated and inaccurate. There are dislocations that are not detected and dislocations that occur late. If several basic principles are followed, the ability to detect DDH can be improved.

Asking the family about potential risk factors including a family history of hip dislocation, ligamentous laxity, pre and postnatal positioning, and genetic and environmental factors is recommended. A history of breech position, even if it was transient or if the delivery was by cesarean section, is an important risk factor. Conditions including plagiocephaly, clubfeet, metatarsus adductus, and congenital muscular torticollis may be associated with DDH. The mother may note a difference in the length of the legs or that one leg will not come out far enough (limited abduction) when changing diapers. In the older child, a limp, toe-walking, intoeing, or outtoeing may be secondary to DDH.

RESULTS AND DISCUSSION

Physical Examination

In the neonatal period, the diagnosis is made by physical examination. The physical signs change and none are pathognomonic. The examination requires patience and skill and the newborn must be relaxed on a firm surface. It is impossible to perform an adequate physical examination on a crying, fussy newborn. When performing the examination the physician looks for asymmetry between the lower extremities. Asymmetric thigh folds, a short leg appearance, a prominent greater trochanter, or limitation of abduction or adduction may be significant findings. In the supine position, with the hip in 90° of flexion and one hand stabilizing the pelvis, each hip should easily abduct to 75° and adduct to 30° past the midline.

The examiner attempts to reproduce the dislocation or subluxation using the Ortolani and Barlow maneuvers. With the newborn supine the tips of the
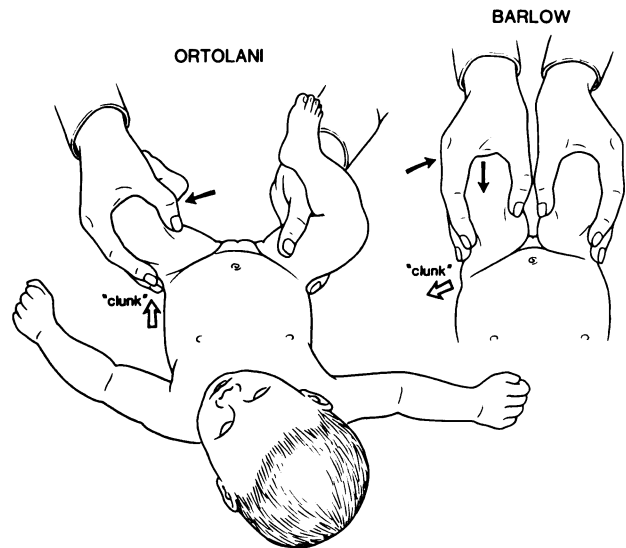

Fig. 1. Drawings demonstrating the technique for performing the Ortolani and Barlow maneuvers. The Ortolani sign is obtained by gently abducting the leg and a palpable "clunk" is felt as the femoral head slides over the posterior rim of the acetabulum into the socket. This is called the sign of entry. The Barlow provocative test is obtained by abducting the hip and pushing gently on the knee and a palpable "clunk" is felt as the femoral head slides over the posterior rim of the acetabulum and out of the socket. This is called the sign of exit.

the greater trochanter with the thumb along the medial thigh. The leg is positioned in neutral rotation with 90° of hip flexion and is gently abducted while lifting the leg anteriorly. With abduction one can feel a clunk as the femoral head slides over the posterior rim of the acetabulum and into the socket (Fig 1). This is the clunk originally described by Ortolani.[37] This is called the sign of entry, as the hip relocates with this maneuver. Maintaining the same position, the leg is then gently adducted while gentle pressure directed posteriorly is placed on the knee and a palpable clunk is noted as the femoral head slides over the posterior rim of the acetabulum and out of the socket. This clunk was originally described by Barlow[11] who liked to combine this provocative test with the Ortolani test (Fig 1). This is called the sign of exit, as the hip dislocates with this maneuver. Both tests are designed to detect motion between the femoral head and the acetabulum. The reproducibility of these tests is dependent on ligamentous or
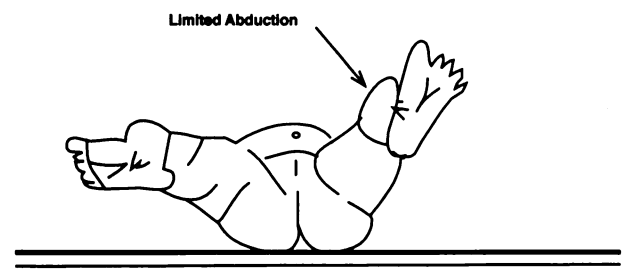

Fig. 2. A drawing demonstrating limited abduction of this infant's left hip. The infant is placed supine on a firm table and the examiner simultaneously abducts both hips. Asymmetrical abduction is often associated with developmental dysplasia of the hip.

capsular laxity, which usually disappears by 10 to 12 weeks of age.

The physical examination is performed one hip at a time using the other hand to stabilize the pelvis. The amount of force required to illicit an Ortolani or Barlow sign is minimal. It is important to distinguish the clunk of an Ortolani or Barlow maneuver from a high-pitched click, that may occur with flexion and extension in an abducted position. A subluxatable hip is characterized by a feeling of telescoping or sliding movement of the temporal head. The Ortolani and Barlow maneuvers are negative because there is no palpable clunk of exit or entry.

In the 3- to 12-month-old infant, there is less ligamentous and capsular laxity so the Ortolani and Barlow tests usually disappear. The best physical finding in this age group is limitation of hip abduction (Fig 2). A unilateral dislocation is identified by limited abduction of the hip, a positive Galleazzi's sign (relative shortening of the femur), and extra thigh folds secondary to shortening.

The diagnosis of bilateral hip dislocation is more difficult as the limitation of abduction may be symmetric. Hip abduction in bilateral DDH is typically limited to 30 to 40° although in the normal 3- to 12-month-old infant, it should be 75 to 80° with the hip in 90° of flexion. This test offers another clue to the difficult diagnosis of bilateral DDH. Nélaton's line is an imaginary line between the ischial tuberosity and the anterosuperior iliac spine. The greater trochanter in a dislocated hip lies cephalad to this line while the trochanter in a normal hip lies caudad. A high index of suspicion is required to detect bilateral DDH.

Once a child is walking the physical signs become more obvious. There is a typical limp and the child will often toe-walk on the affected side. If both hips are dislocated, increased lumbar lordosis, prominent buttocks, and a waddling gait pattern are noted. When the patient is asked to stand on the affected leg, the pelvis drops to the opposite side and the trunk leans toward the affected side (positive Trendelenburg test). This test is positive because the superiorly dislocated hip mechanically shortens the abductor muscles decreasing their strength.

The diagnosis of hip dysplasia is more difficult. The adolescent patient may note pain or discomfort after walking and may limp intermittently. The physical examination may be normal or some discomfort may be elicited at the extremes of range of motion, particularly abduction and internal rotation. If any of these signs are present, an anteroposterior pelvis radiograph is recommended.

Radiographic Evaluation

The anteroposterior pelvis radiograph is difficult to interpret in the neonatal period.[11,38] A normal newborn radiograph may be misleading and deceptive because much of the neonatal pelvis and femoral head are cartilaginous so the relationship between the femoral head and acetabulum is difficult to determine. In positioning the newborn for the radiograph, the technician may spontaneously reduce a
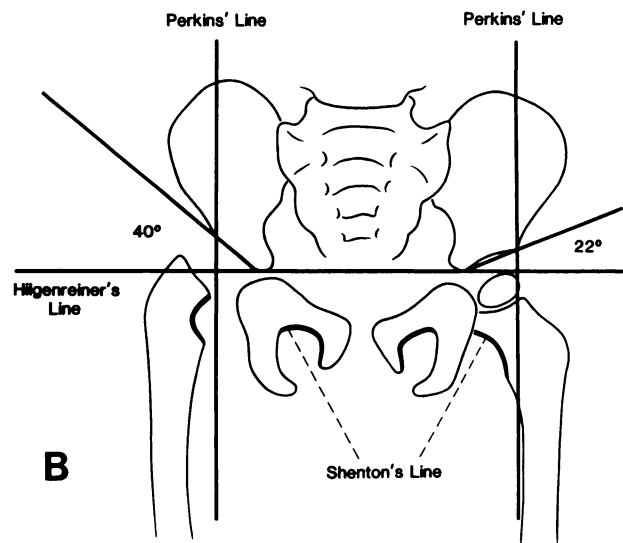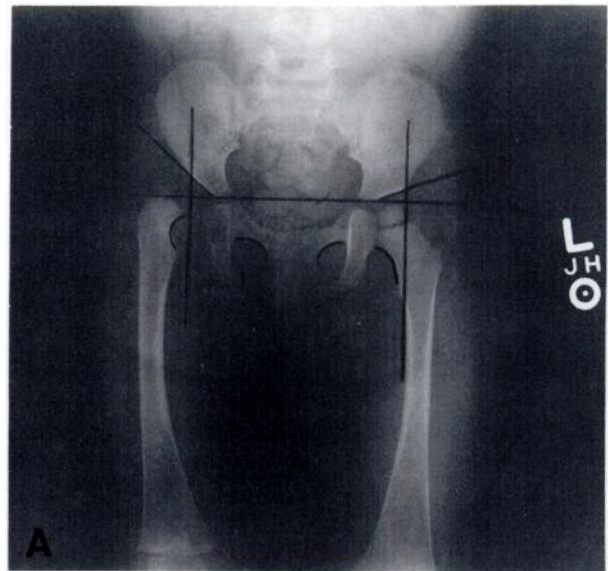

Fig. 3. A, An anteroposterior pelvis radiograph of an 8-month-old infant with DDH on the right. B, The drawing of the radiograph shows Hilgenreiner's line connecting the top of the triradiate cartilages. Perkins' line is perpendicular to Hilgenreiner's line through the lateral ossified margin of the acetabulum. Shenton's line forms a continuous contour between the obturator foramen and the medial border of the femoral neck. There is DDH on the right with a delay in the appearance of the ossific nucleus, lateral displacement of the proximal femoral metaphysis, an elevated acetabular index (40°), and a "break" in Shenton's line.

addition, if the newborn is not positioned properly, asymmetry is introduced that makes interpretation difficult.

From a properly positioned anteroposterior pelvis radiograph several parameters are used to evaluate the hips. Hilgenreiner's line is a horizontal line connecting the top of the respective triradiate cartilages. Perkins'[39] line is drawn perpendicular to Hilgenreiner's line through the most lateral ossified margin of the roof of the acetabulum. The medial margin of the proximal femoral metaphysis should lie in the inferomedial quadrant of the intersection of these two lines (Fig 3).

The teardrop shadow is a normal radiographic

months old.[40] It consists of three lines: a lateral semi-circular line corresponding to the cortical surface of the acetabular fossa, a medial line corresponding to the medial cortex of the pelvic wall, and a short, curved line connecting these two lines corresponding to the semicylindrical cortex of the acetabular notch. Delay in the ossification of the teardrop may result from a lack of stimulation from a concentrically reduced femoral head suggesting DDH. Similarly, a delay in the appearance of the ossification center of the femoral head, which normally appears between 3 and 7 months of age, also suggests DDH.

Shenton's line is a line drawn along the medial border of the neck of the femur and the superior border of the obturator foramen and should form a continuous contour. If there is superolateral subluxation of the femoral head, Shenton's line is interrupted (Fig 3).[41] The acetabular index represents the angle formed between Hilgenreiner's line and a line beginning at the lateral ossified margin of the roof of the acetabulum and extending to the intersection between the depth of the acetabular socket and Hilgenreiner's line. The acetabular index is helpful in evaluating the development of the acetabulum. In the newborn the acetabular index averages 28° and decreases to 20° by 2 years of age.[42] Coleman[43] reported on 1155 newborns who were examined both clinically and radiographically and found 77 that had abnormal clinical examinations. The acetabular index averaged 29° in the healthy newborns and 35° in those with abnormal clinical findings. It is important to remember that a radiograph with positive findings is helpful while a normal-appearing radiograph may be misleading. The diagnosis in the newborn is made by clinical examination and can be confirmed by ultrasound examination.

Ultrasound

In 1980, Graf developed the idea of adopting ultrasound to study the neonatal hip joint.[44,45] The two methods currently in use include the static technique proposed by Graf, and the dynamic or real time method as described by Harcke et al.[46]

The static technique is performed with the infant in the lateral decubitus position and the hip in 35° of flexion and 10° of internal rotation. A coronal image of the hip is obtained and three lines are constructed from the image. A vertical line is drawn parallel to the ossified lateral wall of the ilium. A second line is drawn along the roof of the cartilaginous acetabulum from the lateral bony edge of the acetabulum to the labrum. A third line is drawn from the inferior edge of the bony acetabulum at the roof of the triradiate cartilage to the most distal point on the ilium in the center of the hip joint. The alpha angle is formed between the line parallel to the lateral wall of the ilium and the line parallel to the bony acetabulum. The lower limit of normal for the alpha angle is 55° (Fig 4).[47] Because the alpha angle reflects bony acetabular coverage of the femoral head, the smaller the angle, the greater the degree of dysplasia. The beta angle is formed between the line parallel to the lateral wall of the ilium and the line parallel to the roof
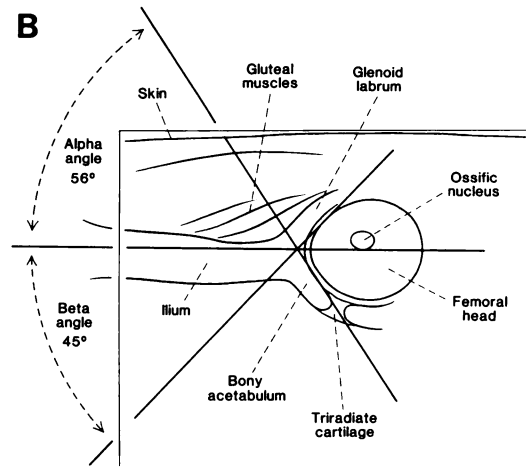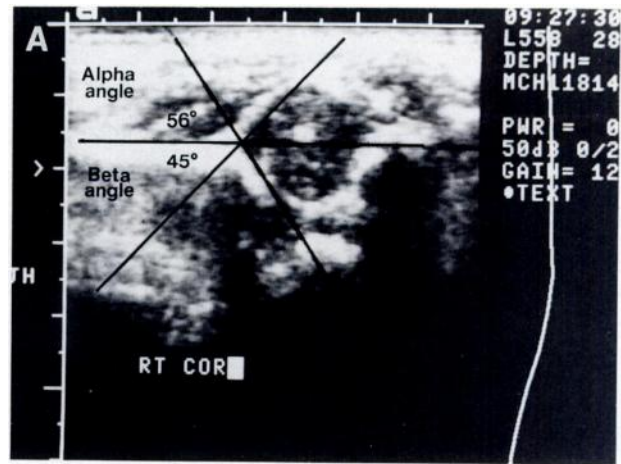

Fig. 4. A, A coronal ultrasound of the right hip in a 7-month-old infant with DDH. B, The drawing of the ultrasound shows a line drawn parallel to the lateral wall of the ilium. A second line is drawn along the roof of the cartilaginous acetabulum. A third line is drawn along the inferior edge of the bony acetabulum. The alpha angle measures 56° (Normal >55°) and the beta angle measures 45° (Normal <72°).

>72°, it indicates eversion of the labrum and subluxation of the hip (Fig 4).[47] The problem with the static method is that the alpha and beta angles are difficult to reproduce accurately and if the image is not precise, the hip may appear more dysplastic than it really is.

The dynamic or real time method attempts to visualize the Barlow and Ortolani maneuvers on the ultrasound screen. The technique is dependent on ligamentous or capsular laxity and just like the physical examination, the ultrasound examination is dependent on the operator. The dynamic technique is performed with the infant in both the lateral decubitus and supine positions and imaging in the coronal and transverse planes with and without stress. The coronal image is obtained with the hip flexed to 90° and posterior stress is applied to the knee with the palm of the hand (Barlow provocative test) and any subluxation is noted (Fig. 5). Six mm of subluxation on the left and 4 mm on the right is normal during the first few days of life.[48] If the hip subluxates or dislocates, an attempt at reduction is performed (the Ortolani maneuver). The second stage of the dy-

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