Developmental Dysplasia of the Hip: Definition, Recognition, and Prevention of Late Sequelae

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The American Academy of Pediatrics (AAP), Pediatric Orthopaedic Society of North America (POSNA), and American Academy of Orthopaedic Surgeons (AAOS) currently all recognize the term developmental dysplasia of the hip (DDH) and have recommended that it replace the more traditional term congenital dislocation of the hip (CDH). The change in terminology was the culmination of decades of physician dissatisfaction with the term CDH. However, CDH was a specific entity and was easy to understand. DDH is a concept that is complex and somewhat difficult to comprehend for those who do not use it frequently. DDH is easier to understand when the change in terminology is viewed from the historical perspective.

HISTORICAL PERSPECTIVE

In 1826, a French surgeon, Dupuytren, noted in autopsy studies a type of hip dislocation in which there was “a defect in the depth or completeness of the acetabulum.” The shallow acetabulum distinguished “this variety of dislocation,” which he named “original or congenital dislocation” from acquired dislocations secondary to trauma or infection. “CDH” was accepted as a distinct clinical entity for most of the next century.

However, following the routine use of radiographs in the early 20th century, orthopedic surgeons made a number of interesting observations concerning CDH. They often noted that a child with CDH of one hip would have a located or minimally subluxated hip, with a shallow acetabulum on the contralateral side. They questioned what term should they use for the contralateral hip, which clearly was not dislocated. In 1926, Hilgenreiner, a Czech orthopedic surgeon, used the term dysplasia, which literally means abnormal growth, to describe the shallow acetabulum and soft tissue changes associated with CDH.

Putti, an Italian orthopedic surgeon from Bologna, in 1933 made the observation of “a state of pre-dislocation seen on X-ray evidenced merely by an increased tilting of the roof of the acetabulum and few clinical signs.” He suggested that some of these might progress to a complete dislocation. Putti was one of the first physicians to realize the importance of early recognition and treatment of CDH. He thought CDH could possibly be diagnosed at birth not
by physical exam but by x-ray. He recommended that all babies be screened for CDH with a pelvis x-ray at birth. However, radiographs are of limited use until age 4 months, as the newborn hip is cartilaginous and therefore cannot be visualized by x-ray. Although his recommendation of screening for CDH by x-ray was unsuccessful, Putti's unwavering advocacy for the early diagnosis of CDH influenced many physicians and students, including Mariano Ortolani.

Ortolani studied medicine in Bologna in the 1920s and was a "disciple of Putti." In 1929, he opened a pediatric practice in Ferrara, a short distance from Bologna. In 1935, a mother brought her 5-month-old child to him stating that she could feel a "click" every time the baby was washed. While examining the baby, he determined that the audible and palpable "click" occurred with reduction of a dislocated hip. He eventually learned that the "click," which occurs with gentle traction and abduction of the baby's thigh, could be detected in newborns with CDH. Subsequently, Ortolani devoted much of his career to the early diagnosis and treatment of "congenital" dislocation of the hip. He wrote many
papers and traveled widely encouraging others to use his test. In 1953, he stated: “Go to the newborn nursery and make the diagnosis of congenital hip problem, and remember that the golden time for treatment is the first day of life; any other time is not early enough.”

Many orthopedic surgeons were slow to appreciate the importance of Ortolani’s work. One of those who did was Vernon Hart of Minnesota. Hart promoted Ortolani’s concepts in the United States in the 1940s. He used the term “jerk of entry” to describe the Ortolani sign. This more accurately reflects the proprioceptive (not audible) nature of the Ortolani sign as the femoral head is reduced into the acetabulum. He also noted that by reversing the abduction maneuver, one could palpate a “jerk of exit” as the femoral head redislocated out of the acetabulum.

Subsequently, Sherman Coleman, an orthopedic surgeon from Salt Lake City, noted that there were many babies with CDH who did not have a positive Ortolani sign, but had only a “jerk of exit.” Kurt Palmen, a Swedish pediatrician, noted the same phenomenon. In some babies, the hip could be provoked to subluxate. He called this “subluxation provocation.” These babies had femoral heads that were reduced at rest but could be provoked to subluxate or dislocate with the reversal of the Ortolani maneuver. With gentle adduction and a slight compressive force, a subtle proprioceptive jerk could be felt as the femoral head dislocated out of the acetabulum.

In 1962, Barlow, an English orthopedic surgeon, first used the simpler term “dislocatable” to describe the hips of babies with a “jerk of exit” or “subluxation provocation.” Barlow examined almost 10,000 newborns in the first week of life looking for instability and found that the incidence of instability (1:60) was much higher than previously had been reported. The majority of the unstable hips did not have a positive Ortolani sign. They were located at rest but were dislocatable.

In addition, Barlow made the remarkable observation that the incidence of instability was more than twice as high in babies examined between birth and 31/2 days of age (1:25) than babies examined between 31/2 and 7 days of age (1:100). It was obvious that the instability present at birth resolves quickly with time. Barlow found that 88% of the babies with instability went on to stabilize in a normal position by age 2 months. However, 12% stabilized in a position of fixed subluxation or dislocation. These babies were not born with a congenital dislocated hip; they were born with hip instability and developed a dislocated hip with time.

The percentage of babies born with hip instability that ultimately develop fixed subluxation or dislocation can be influenced by post-natal factors. Dr. Robert Salter of Toronto, Ontario, Canada, who has made innumerable contributions to our understanding of DDH, noted that a 1963 survey of “Canadian Indian tribes” demonstrated infants positioned on cradleboards had a tenfold higher incidence of “late CDH” than infants from the same tribe not positioned on a cradleboard. A cradleboard maintains the infant’s hips in a position of extension and adduction, which prevents the natural resolution of instability and dysplasia that occurs in many babies with DDH at birth.

In the late 1970s, Graf, an orthopedic surgeon from Austria, noted that the cartilaginous femoral head and acetabulum of a newborn could be imaged beautifully by ultrasound. Subsequently, Harcke, a radiologist from Wilmington, Delaware, described and promoted a dynamic examination of the infant hip by ultrasound. Harcke was able to see what Ortolani and Barlow could only feel.

During the past 20 years, numerous studies using ultrasound have confirmed the high incidence of acetabular dysplasia and ligamentous laxity accompanied by the presence or absence of instability at birth. In addition, the developmental nature of hip dysplasia with resolution of dysplasia in many infants, but progres-

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CME Educational Objectives

1. Identify the etiology and natural history of developmental dysplasia of the hip (DDH).
2. Describe physical examination techniques for DDH.
3. Discuss when referral for hip imaging or referral to a pediatric orthopedic surgeon is appropriate for the treatment of DDH.

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sion to fixed subluxation and dislocation in others, has been confirmed.\textsuperscript{14,16,17,18,20}

**DEFINITION, ETIOLOGY, AND NATURAL HISTORY OF DDH**

DDH includes a spectrum of abnormalities of the hip in infants and children that may either resolve or worsen with time. These abnormalities include variable degrees of dysplasia (acetabular and ligamentous) that may be mild or severe but are always present. In addition, there may be an abnormal relationship between the femoral head and acetabulum that includes hips that are located at rest but dislocatable or subluxatable with stress, as well as hips that are dislocated or subluxated at rest and may or may not be reducible. DDH has been defined by the American Academy of Pediatrics Subcommittee on Developmental Dysplasia of the Hip as occurring in two types, teratologic DDH and typical DDH.\textsuperscript{1}

Teratologic DDH is extremely rare, 1 in 25,000 births, and generally occurs in babies with other problems, such as Larsen’s syndrome, arthrogryposis, or spina bifida.\textsuperscript{21} A child born with teratologic DDH has a hip that is dislocated at birth and is not reducible. There are severe dysplastic changes to the shape of the acetabulum and femoral head, as well as significant soft tissue contractures. Teratologic DDH uniformly requires surgical treatment.\textsuperscript{21}

Typical DDH at birth occurs in approximately 1 of 85 babies and thus is 300 times more common than teratologic DDH. Typical DDH is a “deformation” resulting from extrinsic pressure at the end of the third trimester as opposed to an intrinsic “malformation” that occurs during embryogenesis in the first trimester.\textsuperscript{22-25} At the time of birth, the only pathologic findings in the hips of babies with typical DDH are ligamentous laxity, slight acetabular dysplasia, or both. This was demonstrated in autopsy studies of stillborns with typical DDH by Dunn\textsuperscript{25,26} and confirmed by numerous ultrasound studies during the past 2 decades. Autopsy studies of babies with teratologic DDH at birth showed severe acetabular dysplasia and soft tissue contractures similar to those Dupuytren described and those found at surgery for typical DDH in older children.\textsuperscript{26}

DDH occurs 4 times more commonly in girls than boys, is extremely uncommon in black babies, and is bilateral 25% of the time. When DDH is unilateral, the left hip is affected 4 times more than the right.\textsuperscript{28} This reflects the deformational etiology of DDH, as most babies have their left hip adducted in utero (an unstable position for the hip) secondary to pressure from their mother’s sacrum. Genetic factors are also important, especially for infant girls. There is a definite tendency for typical DDH to occur more commonly in certain families. Genetic studies of families with DDH have shown that the acetabular dysplasia and ligamentous laxity may be in part inherited as a multiple gene system.\textsuperscript{27}

Babies with significant DDH at birth have instability between the femoral head and acetabulum (neonatal hip instability; NHI) secondary to the ligamentous laxity, slight acetabular dysplasia, or both.\textsuperscript{29} The instability resolves in all babies.\textsuperscript{3} The majority of unstable hips resolve within a few days or weeks, with the femoral head perfectly located in the acetabulum, and ultimately develop into normal hips. Approximately 10% to 20% of unstable hips stabilize with the femoral head in a dislocated or subluxated position.\textsuperscript{10}

The incidence of late presentation (after walking age) of a developmentally dislocated hip in an unscreened population is approximately 1.5 per 1,000 children.\textsuperscript{1,18} Babies found to have a dislocated hip after walking age were not born with a congenital dislocation of the hip; they were born with dysplasia, and transient neonatal hip instability developed into a fixed dislocation with time. As the dislocation develops, the severity of the acetabular dysplasia worsens because induction of acetabular development requires the femoral head to be properly positioned within it. The ligamentous laxity resolves and is replaced by a significant contracture of the hip capsule, psoas and adductor tendons. The hip pathology is then similar to the pathology in teratologic DDH at birth.\textsuperscript{21,25}

A small percentage of unstable hips stabilize in a subluxated position. Babies with neonatal hip instability whose hips stabilize in a subluxated or dislocated position generally, but not always, develop an adduction contracture (loss of abduction) as the instability resolves and the adductor muscles become shortened.\textsuperscript{1}

**TABLE.**

**Incidence of Developmental Hip Dysplasia (Neonatal Hip Instability) At Birth**

<table>
<thead>
<tr>
<th>Risk Group</th>
<th>Approximate Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>All Newborns</td>
<td>1:85</td>
</tr>
<tr>
<td>Infant boys — no risk factors</td>
<td>1:250</td>
</tr>
<tr>
<td>Infant girls — no risk factors</td>
<td>1:50</td>
</tr>
<tr>
<td>Infant boys — positive family history</td>
<td>1:150</td>
</tr>
<tr>
<td>Infant girls — positive family history</td>
<td>1:33</td>
</tr>
<tr>
<td>Infant boys — breech presentation</td>
<td>1:33</td>
</tr>
<tr>
<td>Infant girls — breech presentation</td>
<td>1:8</td>
</tr>
</tbody>
</table>
Children with slight developmental subluxation may not have any signs or symptoms of hip pathology until they develop pain secondary to degenerative arthritis in adolescence or after skeletal maturity. Adult orthopedic surgeons have known for years that the etiology of degenerative arthritis of the hips in many adults is acetabular dysplasia. In a normal hip, the femoral head sits perfectly centered and is nicely contained by the acetabulum. With weight bearing, stress is evenly distributed over the head and socket. If the acetabulum is shallow, and especially if the femoral head is subluxated, stress is concentrated in a small area at the edge of the socket. This ultimately leads to degenerative arthritis and pain. It seems likely that a significant proportion of adult acetabular dysplasia is the result of DDH that did not resolve totally. However, the exact relationship of adult acetabular dysplasia with DDH has not been documented prospectively.

**RISK FACTORS**

Babies carried in the breech position and baby girls with a positive family history of DDH are at greater risk for having DDH (neonatal hip instability) at birth. The Table (see page 95) gives the incidence of DDH (neonatal hip instability) at birth in babies with and without risk factors. It is important to remember that 60% of infants born with DDH have no risk factors, and therefore all babies should be considered to be at risk for DDH.

**DIAGNOSIS**

The late presentation of DDH cannot be prevented, but it can be minimized by early detection and treatment. At this time, the cornerstone of early detection is repeated, careful examinations at birth and throughout the first year of life. The examination of the infant hip consists of an evaluation of hip instability by performing the Ortolani and Barlow tests and evaluation of the range of abduction and length of the thigh. It has been stated that performing these tests (i.e., examination for neonatal hip instability) is easy. It is not. Training, practice, and experience are required to be proficient. Even in the most experienced hands, neonatal hip instability can be undetectable.

As the instability may resolve very quickly, the first exam should be done within 24 hours of birth. This may be the only chance of identifying those babies at risk for developing fixed subluxation or dislocation. When performing the exam, it is essential that the baby be warm and content and the examiner relaxed and not rushed. The exam must be performed with the diaper off, and one hip is tested at a time. The most common mistake in performing the exam is holding the baby’s leg tightly. A tight grip prevents the examiner from feeling sometimes subtle “jerk” as the femoral head moves into (or out of) the acetabulum. The baby’s legs should be held with the same pressure one would use holding an egg.

The examiner tests one hip at a time, stabilizing the pelvis with the opposite hand. The hip to be examined is held as in Figure 1, with the knee flexed, the tip of the long finger over the greater trochanter, and the thumb on the inside of the thigh. The leg is very gently abducted with light pressure over the greater trochanter with the fingertip and a very slight traction force. If the hip is dislocated at rest, it will reduce with a palpable, not audible, clunk or jerk. This is the positive Ortolani sign of a reducible dislocated hip. The test is then reversed.
(Figures 3 and 4). The leg is gently adducted with light pressure on the inside of the thigh with the thumb and a very slight compressive force. If the hip is dislocatable, it will dislocate with the same palpable, not audible, clunk or jerk. This is a positive Barlow sign that occurs with dislocatable hips.

The difference between the two tests is the position of the hip at rest. When the child has a positive Ortolani, the hip is dislocated at rest, reduces with the Ortolani maneuver, and then redislocates spontaneously. When the child has a positive Barlow, the hip is located at rest but is dislocatable with the Barlow maneuver. The hip then tends to relocate spontaneously. The baby with a positive Ortolani has a more severe degree of dysplasia than the baby with a positive Barlow.

It is not unusual even for experienced examiners to detect instability with the two tests, but to be uncertain as to the position of the hip at rest. Therefore, many pediatric orthopedists prefer to use the term neonatal hip instability to include both dislocated but reducible and dislocatable hips. All babies with unstable hips at birth are at risk for developing a fixed subluxation or dislocation and need to be identified.

When examining the infant for instability, high-pitched clicks that are often audible as well as palpable are frequently encountered. These clicks, which can be confused with true instability, are benign and resolve with time.

If the baby’s hips are determined to be stable, they are next examined for an adduction contracture (loss of abduction). Both hips are examined at the same time, as seen in Figure 5 (see page 98). With the hip and knee flexed, the legs are gently abducted. Any asymmetry of abduction may represent dysplasia. It is very important to ensure that the baby’s pelvis is level with the exam table. It is easy to mask asymmetrical abduction by tilting the baby’s pelvis. (Figure 6, see page 98). In addition, as hip dysplasia may be bilateral in 25% of cases, the baby may have a symmetrical loss of abduction.

A question frequently asked is, “How tight is too tight?” There is no absolute answer to this as the range of abduction of babies with normal hips varies widely. After evaluating many babies, the examiner will develop a feel for what “normal” is. Certainly, any baby who has less than 120 degrees of symmetrical abduction, as well as any baby who has asymmetric abduction, should be referred.

Following the exam for contracture, the baby’s thigh lengths are checked, as in Figure 7 (see page 99). If the knees are not at the same level, the baby should be referred. This is known as a positive Galeazzi sign. The hip on the “low side” may be subluxated or dislocated posteriorly.

The baby’s hips should be re-examined carefully at every well-baby visit. The AAP recommends repeat exams at age 2 to 4 days, 1 month, 2 months, 4 months, 6 months, 9 months, and 12 months.

SCREENING

Since the 1960s, as examining babies for instability and contracture became generally accepted as part of the newborn exam, it was hoped that late presentation of DDH would disappear completely. Unfortunately, this did not happen. In many communities, the incidence of late presentation stayed the same or decreased very little. However,
in a few locations (Sweden38; Sale, Australia32; New Plymouth, New Zealand39; Vancouver, British Columbia, Canada40; Oklahoma City, OK41; and Poole, England42), there was a marked decrease in the late presentation of DDH. The one factor that these seemingly dissimilar places had in common was that they all had formal DDH screening programs in which the screeners were either very experienced or were given prolonged detailed instruction on the examination for neonatal hip instability. These programs used orthopedic surgeons, pediatricians, obstetricians, family practitioners, nurse practitioners, and physiotherapists as primary screeners. The most important aspect of a screening program is the experience of the screener, not the initials after the screener’s name.

As ultrasound is more sensitive than physical examination, ultrasound screening programs have become popular. Similar to screening by physical exam, some programs have been successful while others have not. As in screening by physical examination, the experience of the ultrasonographer is crucial to the success of an ultrasound screening program.19 Hip ultrasonography by an inexperienced ultrasonographer is worse than no ultrasound at all.

Currently, three methods of DDH screening have been advocated:

- Universal clinical screening by physical examination (clinical screening).
- Universal clinical screening by physical examination supplemented by selective screening with ultrasound or x-ray (if ultrasound is unavailable) for children with risk factors (selective ultrasound/x-ray). The risk factors that have generally been used are questionable physical exam, breech presentation and positive family history. There is some evidence that torticollis and congenital foot deformity may be risk factors as well.
- Universal screening of all babies by ultrasound, or x-ray if ultrasound is unavailable (universal ultrasound/x-ray).

Hundreds of articles have been published concerning screening infants for developmental hip dysplasia during the past 20 years. All three methods of screening have been successful in certain circumstances. However, the success of any screening program is dependent on the experience and ability of the screener.32,42,44-46 At this time, in the United States, few locations have dedicated experienced hip ultrasonographers. In addition, the cost for universal ultrasound with follow-up ultrasound for the many infants with minor instabilities would be significant. There has been little enthusiasm for universal screening at age 4 months with x-ray43 due to the radiation exposure and delay in diagnosis compared with ultrasound.

The American Academy of Pediatrics Subcommittee on Developmental Dysplasia of the Hip has recommended careful clinical screening of all babies at birth and at well-child exams during the first year, looking closely for instability and contracture, supplemented by selective ultrasound at age 3 to 4 weeks (or x-ray at 4 months if ultrasound is not available) for babies with risk factors or questionable physical findings.1 Ultrasound (or x-ray) is recommended as an adjunct, not a replacement for clinical screening.1 Ultrasound at birth has a very high false-positive rate. There-
Therefore, ultrasound is recommended at age 3 to 4 weeks rather than at birth to allow the many trivial instabilities present at birth to resolve. Infant girls who were in the breech position and who have a negative clinical exam should undergo either an ultrasound at age 3 to 4 weeks by an experienced ultrasonographer or an anteroposterior pelvis x-ray at age 4 months if ultrasound is not available. Imaging with ultrasound at age 3 to 4 weeks or x-ray at age 4 months is considered optional for baby boys who are breech and baby girls with a positive family history.

The importance of the newborn hip exam cannot be stressed enough. In addition to the AAP Subcommittee on DDH, the Canadian Task Force on Preventative Health Care and the National Screening Committee of the United Kingdom also recently reviewed the vast literature concerning screening for DDH. They agree that clinical screening by an experienced examiner is the most important strategy currently available to prevent the late presentation of DDH.

Many infants have high-pitched audible and palpable tendinous clicks that can be confused with true neonatal instability. These clicks often disappear in the first few weeks of life. However, positive Barlow and Ortolani signs also quickly resolve in more than 80% of infants with neonatal hip instability. Therefore, the resolution of questionable findings is not a guarantee that the findings did not represent true instability. In my opinion, any child with a questionable exam should be referred to a pediatric orthopedist or have an ultrasound by an experienced ultrasonographer at age 3 to 4 weeks or an x-ray at age 4 months, if ultrasound is not available.

TREATMENT

In the early 20th Century, CDH generally was not diagnosed until well after the walking age. Standard treatment was the Lorenz method, advocated by Adolph Lorenz, an orthopedic surgeon from Vienna. The Lorenz method involved closed reduction of the dislocated hip under anesthesia and immobilization in wide abduction and external rotation (the Lorenz or “frog” position). Years later, it was determined that the incidence of avascular necrosis, the most feared complication of treatment of DDH, is extremely high with rigid immobilization in the Lorenz position.

In 1957, Arnold Pavlik, an orthopedic surgeon from Czechoslovakia, described the functional method of treatment using a harness with stirrups. Pavlik’s method uses the baby’s natural kicking movement to encourage reduction of the hip, rather than forcibly reducing the hip and holding it in a fixed position. The “Pavlik harness” was introduced into the United States by Dr. Dean MacEwen, who was then Director of the DuPont Institute in Wilmington, DE, and one of the strongest advocates for prevention in pediatric orthopedics in North America. The harness is now considered the treatment of choice for infants younger than 6 months with DDH. The incidence of avascular necrosis when the Pavlik harness is used for an unstable hip in the neonatal period is extremely low, with no cases appearing in a number of series.

When the harness is used for hips that have stabilized in a dislocated or subluxated position, the incidence of avascular necrosis increases. In addition, some hips fail to stabilize in a located position with the harness. Closed reduction under anesthesia with adductor tenotomy and hip spica cast immobilization is recommended for children who have failed the Pavlik harness or are 6 months or older at the time of diagnosis. Children who fail closed reduction or are older than 18 months at the time of diagnosis generally are treated by surgical methods. As significant complications (avascular necrosis, femoral nerve palsy, inferior dislocation) can occur with the Pavlik harness, it should only be used by physicians who have significant experience in the treatment of DDH in babies of all ages.

Because the majority of unstable hips at birth stabilize and develop into normal hips without treatment, and because there is a small risk of treatment at birth, some pediatric orthopedic surgeons have advocated “managing” babies with neonatal hip instability with a short period of close observation, treating only those babies with persistent dysplasia. When babies are monitored purely by physical examination, the treating physician does
not know that the hip has stabilized in a normal position until an x-ray is taken at age 4 months. If there is residual subluxation or dysplasia at that age, the hip is more difficult to treat, and the risk of complications, including avascular necrosis, is greater than it would have been at birth. For this reason, the majority of pediatric orthopedic surgeons in the US recommend immediate treatment of any babies with unstable hip at birth.

However, when babies are monitored by experienced ultrasonography, hip stability and normality can be verified by age 3 to 4 weeks. Therefore, pediatric orthopedists with experienced ultrasonography available will sometimes manage infantile DDH (neonatal hip instability) with a few weeks of close observation by both physical exam and ultrasound, treating only those babies with residual abnormality at age 3 to 4 weeks.50,51

**SUMMARY**

When a child presents with a dislocated hip after the walking age, parents are upset, pediatricians are distraught, and lawsuits often follow. The prevention of late presentation is a goal that all practitioners should strive for. However, at this time, using current diagnostic techniques, the late presentation of DDH can be minimized but not eliminated. The AAP Subcommittee on DDH has estimated that approximately 15% of DDH at birth is not detectable, even by experienced examiners or ultrasonographers.1

In addition to preventing the late presentation of a developmentally dislocated hip, the prevention of premature degenerative arthritis of the hip secondary to developmental subluxation and acetabular dysplasia is equally, if not more, important in terms of morbidity and cost. It is hoped that the identification and treatment of babies with DDH at birth will have the added benefit of decreasing the incidence of degenerative arthritis of the hip in adults.

As the key to early detection remains repeated, careful examination of the infant in the first year of life, it is imperative for practitioners to become as skilled as possible in performing the exam. Unfortunately, medical school curriculum and pediatric and family practice residency programs often are deficient in teaching the neonatal hip examination.

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**REFERENCES**


