Dr. Upendra Yadav et al., IJSIT, 2018, 7(3), 474-480

International Journal of Science Inventions Today
www.ijsit.com
ISSN 2319-5436

Review Article

SCREENING THE NEWBORN FOR DEVELOPMENTAL DYSPLASIA OF THE HIP: REVIEW

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ABSTRACT

Developmental dysplasia of the hip is the preferred term to describe the condition in which the femoral head has an abnormal relationship to the acetabulum. Developmental dysplasia of the hip includes frank dislocation (luxation), partial dislocation (subluxation), instability wherein the femoral head comes in and out of the socket, and an array of radiographic abnormalities that reflect inadequate formation of the acetabulum. The precise cause of DDH is unknown, with a combination of genetic and environmental influences associated with DDH and hip dislocation including family history, fetal crowding, vaginal delivery, breech presentation and female gender [1]. Early screening for DDH has the potential to prevent long term hip dysplasia and arthritis requiring hip replacement. Delayed diagnosis requires more complex treatment and has a less successful outcome than dysplasia diagnosed early. For all infants, a competent newborn physical examination using the Ortolani maneuver is the most useful procedure to detect hip instability. Early treatment of an unstable hip with a Pavlik harness or similarly effective orthosis is effective, safe, and strongly advised. Despite having had normal newborn and infant hip examinations, there remains the possibility of a late-onset hip dislocation needing treatment in approximately 1 in 5000 infants.

Keywords: developmental dysplasia of the hip (DDH), newborn, dislocation
INTRODUCTION

Early diagnosis and treatment of developmental dysplasia of the hip (DDH) is important to provide the best possible clinical outcome. DDH encompasses a spectrum of physical and imaging findings, from mild instability and developmental variations to frank dislocation. DDH is asymptomatic during infancy and early childhood, and, therefore, screening of otherwise healthy infants is performed to detect this uncommon condition. The incidence of developmental dislocation of the hip is approximately 1 in 1000 live births. The incidence of the entire spectrum of DDH is undoubtedly higher but not truly known because of the lack of a universal definition. Important risk factors for DDH include breech position, female sex, incorrect lower-extremity swaddling, and positive family history. These risk factors are thought to be additive. Other suggested findings, such as being the first born or having torticollis, foot abnormalities, or oligohydramnios, have not been proven to increase the risk of “nonsyndromic” DDH. Breech presentation may be the most important single risk factor, with DDH reported in 2% to 27% of boys and girls presenting in the breech position. Traditional methods of screening have included the newborn and periodic physical examination and selected use of radiographic imaging. The term Congenital Dislocation of the Hip (CDH) was superseded by the new name of Developmental Dislocation of the Hip (DDH) in 1989. This was in recognition of the fact that not all cases of pathological hip conditions associated with DDH were present at birth. This opinion has important legal ramifications. If some hip joint conditions that are stable at birth deteriorate and are diagnosed at a later date as an irreducible hip dislocation, they cannot be considered to be “missed” cases following negative neonatal clinical hip screening by a competent screener. DDH is a dynamic condition in which the hip abnormality may improve or deteriorate with growth. The spectrum of presentation varies from hip dysplasia, to reducible subluxation/dislocation and eventually irreducible hip joint dislocation. Neurological, neuromuscular, syndromes and skeletal dysplasia’s are excluded, as the hip abnormality is secondary to a primary pathology and is not idiopathic. The traditional outcome measure is that of irreducible hip dislocation. The diagnosis of hip pathology in DDH screening may be clinical and or sonographic.

DISCUSSION

Neonatal screening in dedicated centres has led to a marked reduction in missed cases of DDH. Risk factors such as family history, breech presentation, oligohydramnios and the presence of other congenital abnormalities are taken into account in selecting newborn infants for special examination and ultrasonography. Ideally all neonates should be examined, but if the programme is to be effective those doing the examining should receive special training.

SCREENING METHODS

Serial clinical examination:

Serial clinical examination includes the Ortolani and Barlow tests during the first several months of life and testing for limited hip abduction or leg length discrepancy in older infants and children.
test involves flexion and abduction of the hips. This movement relocates the dislocated hip into the normal acetabular position and is accompanied with a palpable “clunk.”[10-12] The Barlow test is a provocative test of dislocation of the hip joint. The hips are tested individually, both in the flexed position. The tested hip is adducted, with gentle pressure exerted on the upper femur in a posteriolateral direction. Key components of the serial clinical examination include leg length discrepancy (Galeazzi sign), limitation of normal abduction of the hip and asymmetry of posterior thigh or gluteal folds.[11, 12] identified too late for nonsurgical therapy to be effective. In clinically screened populations, the detection rate of hip joint instability at birth has ranged from 5 to 20 cases per 1000 infants, depending mainly on age at testing and examiner experience. An observant mother may spot asymmetry, a clicking hip, or difficulty in applying the napkin (diaper) because of limited abduction. With unilateral dislocation the skin creases look asymmetrical and the leg is slightly short (Galeazzi’s sign) and externally rotated; a thumb in the groin may feel that the femoral head is missing. With bilateral dislocation there is an abnormally wide perineal gap. Abduction is decreased. Contrary to popular belief, late walking is not a marked feature; nevertheless, in children who do not walk by 18 months dislocation must be excluded. Likewise, a limp or Trendelenburg gait, or a waddling gait could be a sign of missed dislocation

**Radiography:**

X-rays of infants are difficult to interpret and in the newborn they can be frankly misleading. This is because the acetabulum and femoral head are largely (or entirely) cartilaginous and therefore not visible on x-ray. X-ray examination is more useful after the first 6 months, and assessment is helped by drawing lines on the x-ray plate to define three geometric indice. Limited evidence supports obtaining a properly positioned anteroposterior radiograph of the pelvis. If the pelvis is rotated or if a gonadal shield obscures the hip joint, then the radiograph should be repeated. Hip asymmetry, subluxation, and dislocation can be detected on radiographs when dysplasia is present. There is debate about whether early minor radiographic variability (such as increased acetabular index) constitutes actual disease. [13] Radiography is traditionally indicated for diagnosis of the infant with risk factors or an abnormal examination after 4 months of age. [4, 14]

**Ultrasonography:**

Ultrasound scanning has replaced radiography for imaging hips in the newborn. The radiographically „invisible“ acetabulum and femoral head can, with practice, be displayed with static and dynamic ultrasound. Sequential assessment is straightforward and allows monitoring of the hip during a period of splintage.

Ultrasonography can provide detailed static and dynamic imaging of the hip before femoral head ossification. The American Institute of Ultrasound in Medicine and the American College of Radiology published a joint guideline for the standardized performance of the infantile hip ultrasonographic examination.[15]. Static ultrasonography shows coverage of the femoral head by the cartilaginous acetabulum (α angle) at rest, and dynamic ultrasonography demonstrates a real-time image of the Barlow and Ortolani tests.
COMPLICATIONS

Failed reduction multiple attempts at treatment, with failure to achieve concentric reduction, may be worse than no treatment. The acetabulum remains undeveloped, the femoral head may be deformed, the neck is usually anteverted and the capsule is thickened and adherent. It is important to enquire also why reduction failed: is the dislocation part of a generalized condition, or a neuromuscular disorder associated with muscle imbalance? The principles of treatment for children over 8 years are the same as those discussed above. Avascular necrosis a much-feared complication of treatment is ischaemia of the immature femoral head. It may occur at any age and any stage of treatment and is probably due to vascular injury or obstruction resulting from forceful reduction and hip splintage in abduction. The effects vary considerably: in the mildest cases the changes are confined to the ossific nucleus, which appears to be slightly distorted and irregular on x-ray. The cartilaginous epiphysis retains the shape and physical growth is normal. After 12–24 months the appearances return to normal. In more severe cases the epiphyseal and physis growth plates also suffer; the ossific nucleus looks fragmented, the epiphysis is distorted to greater or lesser extent and metaphyseal changes lead to shortening and deformity of the femoral neck. Prevention is the best cure: forced manipulative reduction should not be allowed; traction should be gentle and in the neutral position; positions of extreme abduction must be avoided; soft-tissue release (adductor tenotomy) should precede closed reduction; and if difficulty is anticipated open reduction is preferable. Once the condition is established, there is no effective treatment except to avoid manipulation and weight bearing until the epiphysis has healed. In the mildest cases there will be no residual deformity, or at worst a femoral neck deformity which can be corrected by osteotomy. In severe cases the outcome may be flattening and mushrooming of the femoral head, shortening of the neck (with or without coxa vara), acetabular dysplasia and in congruency of the hip. Surgical correction of the proximal femur and pelvic osteotomy to reposition or deepen the acetabulum may be needed.

MANAGEMENT
THE FIRST 3–6 MONTHS:

Where facilities for ultrasound scanning are available, all newborn infants with a high-risk background or a suggestion of hip instability are examined by ultrasonography. If this shows that the hip is reduced and has a normal cartilaginous outline, no treatment is required but the child is kept under observation for 3–6 months. In the presence of acetabular dysplasia or hip instability, the hip is splinted in a position of flexion and abduction and ultrasound scanning is repeated at intervals until stability and normal anatomy are restored or a decision is made to abandon splintage in favor of more aggressive treatment. If ultrasound is not available, the simplest policy is to regard all infants with a high-risk background or a positive Ortolani or Barlow test, as „suspect” and to nurse them in double napkins or an abduction pillow for the first 6 weeks. At that stage they are re-examined: those with stable hips are left free but kept under observation for at least 6 months; those with persistent instability are treated by more formal abduction splintage until the hip is stable and x-ray shows that the acetabular roof is developing satisfactorily (usually
There are two drawbacks to this approach: (1) the sensitivity of the clinical tests is not high enough to ensure that all cases will be spotted [16]; and (2) of those hips that are unstable at birth, 80–90 per cent will stabilize spontaneously in 2–3 weeks. It therefore seems more sensible not to start splintage immediately unless the hip is already dislocated. This reduces the small (but significant) risk of epiphyseal necrosis that attends any form of restrictive splintage in the neonate. Thus: if a hip is dislocatable but not habitually dislocated, the baby is left untreated but reexamined weekly; if at 3 weeks the hip is still unstable, abduction splintage is applied. If the hip is already dislocated at the first examination, it is gently placed in the reduced position and abduction splintage is applied from the outset. Reduction is maintained until the hip is stable. This situation must be avoided; if the hip fails to locate, splintage should be abandoned in favor of closed or operative reduction at a later date.

Follow-up whatever policy is adopted, follow-up is continued until the child is walking. Sometimes, even with the most careful treatment, the hip may later show some degree of acetabular dysplasia.

**PERSISTENT DISLOCATION: 6–18 MONTHS:**

If, after early treatment, the hip is still incompletely reduced, or if the child presents late with a „missed”, dislocation, the hip must be reduced – preferably by closed methods but if necessary by operation – and held reduced until acetabular development is satisfactory. Closed reduction Closed reduction is suitable after the age of 3 months and is performed under general anesthesia with an arthrogram to confirm a concentric reduction. To minimize the risk of avascular necrosis, reduction must be gentle and may be preceded by gradual traction to both legs. Failure to achieve concentric reduction should lead to abandoning this method in favor of an operative approach at approximately 1 year of age. The hips should be stable in a safe zone of abduction, which may be increased with a closed adductor tenotomy.

**Splintage** The concentrically reduced hip is held in a plaster spica at 60 degrees of flexion, 40 degrees of abduction and 20 degrees of internal rotation. After 6 weeks the spica is changed and the stability of the hips assessed under anaesthesia. Provided the position and stability are satisfactory the spica is retained for a further 6 weeks. Following plaster removal the hip is either left unsplinted or managed in a removable abduction splint which is retained for up to 6 months depending on radiological evidence of satisfactory acetabular development. Operation if, at any stage, concentric reduction has not been achieved, open operation is needed. The psoas tendon is divided; obstructing tissues (redundant capsule and thickened ligamentum teres) are removed and the hip is reduced. It is usually stable in 60 degrees of flexion, 40 degrees of abduction and 20 degrees of internal rotation. A spica is applied and the hip is splinted as described above. If stability can be achieved only by markedly internally rotating the hip, a corrective subtrochanteric osteotomy of the femur is carried out, either at the time of open reduction or 6 weeks later. In young children this usually gives a good result.

**PERSISTENT DISLOCATION: 18 MONTHS – 4 YEARS:**

In the older child, closed reduction is less likely to succeed; many surgeons would proceed straight to arthrography and open reduction. Traction Even if closed reduction is unsuccessful, a period of traction (if
necessary combined with psoas and adductor tenotomy) may help to loosen the tissues and bring the femoral head down opposite the acetabulum. Arthrography An arthrogram at this stage will clarify the anatomy of the hip and show whether there is an inturned limbus or any marked degree of acetabular dysplasia. Operation The joint capsule is opened anteriorly, any redundant capsule is removed along with any other blocks to reduction including the hypertrophied ligamentum teres and transverse acetabular ligament and the femoral head is seated in the acetabulum. Usually a derotation femoral osteotomy held by a plate and screws will be required. At the same time a 1 cm segment can be removed from the proximal femur to reduce pressure on the hip (Klisic and Jankovic, 1976). If there is marked acetabular dysplasia, some form of acetabuloplasty will also be needed – either a pericapsular reconstruction of the acetabular roof (Pemberton’s operation) or an innominate (Salter) osteotomy which repositions the entire innominate bone and acetabulum. Splintage After operation, the hip is held in a plaster spica for 3 months and then left unsupported to allow recovery of movement. The child is kept under intermittent clinical and radiological surveillance until skeletal maturity.

**DISLOCATION IN CHILDREN OVER 4 YEARS:**

Reduction and stabilization become increasingly difficult with advancing age. Nevertheless, in children between 4 and 8 years – especially if the dislocation is unilateral – it is still worth attempting, bearing in mind that the risk of avascular necrosis and hip stiffness is reported as being in excess of 25 per cent. The principles of treatment are as described immediately above. Unilateral dislocation in the child over 8 years often leaves the child with a mobile hip and little pain. This is the justification for non-intervention, though in that case the child must accept the fact that gait is distinctly abnormal. If reduction is attempted it will require an open operation and acetabular reconstruction. These procedures are best undertaken in centres specializing in this area. With bilateral dislocation the deformity – and the waddling gait – is symmetrical and therefore not so noticeable; the risk of operative intervention is also greater because failure on one or other side turns this into an asymmetrical deformity. Therefore, in these cases, most surgeons avoid operation above the age of 6 years unless the hip is painful or deformity unusually severe. The untreated patient walks with a waddle but may be surprisingly uncomplaining.

**CONCLUSION**

Late diagnosis of DDH might cause serious public health problems. Screening for and treatment of hip dislocation (positive Ortolani test result) and initially observing milder early forms of dysplasia and instability (positive Barlow test result) infants should be examined for hip instability at birth, with periodic follow-up exams until the child is walking, which helps to prevent further complication of hip joint. Early diagnosis and treatment of developmental dysplasia of the hip (DDH) is important to provide the best possible clinical outcome.
REFERENCES