Evaluation, Referral and Management of DDH

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Introduction :

It is unfortunate that the majority of the patients with Developmental Dysplasia Hip (DDH) are currently being diagnosed after the first year of life in India. By failing to diagnose them early in the infancy, we lose the opportunity to treat this entity without surgical intervention. While the incidence of DDH in India is considered less compared to the western world, we lack a uniform DDH screening or surveillance program. Different academic bodies including Indian Academy of Pediatrics, Pediatric Orthopaedic Society of India and Indian Radiology Society are working on it and soon we will be provided with the guidelines about the early detection and treatment of DDH.

Term DDH :

The original term Congenital Dislocation of Hip (CDH) described the hips which were dislocated at birth. The newer term DDH encompasses the whole spectrum of hip pathologies, including unstable hips, subluxation (partial dislocation), acetabular dysplasia and complete hip dislocation. Thus, DDH is a dynamic disorder with a potential of getting better or worse depending on the care provided to the infant.

Epidemiology :

Incidence of DDH in India varies from 1 - 9.2 per 1000 live birth, with more prevalence in northen India. DDH is responsible for 29% of primary hip arthroplasty up to age of 60 years. Hip dysplasia is the most common cause of hip arthritis in women younger than 40 years.

Left hip is more commonly dislocated than the right and 20% of cases are having bilateral hip dislocation. DDH is more commonly seen in cultures where swaddling is practised. Swaddling forces the legs in adduction and extension making it prone for dislocation. "Hip Friendly Swaddling" practise should be recommended where the child can abduct and flex the hip easily. There is 9:1 female predominance, presumably due to female hormones induced laxity of capsule. 60% DDH patients are firstborn. 20% patients are born breech. Extended breech position carries highest risk of DDH, where the hips are flexed and knee extended. There might be coexistence of Torticollis (wry neck) or Metatarsus Adductus (forefoot bent inside) with DDH as a packaging effects. There is increased incidence amongst offspring of mother who had DDH.

However, the risk factors described above, are poor predictors of DDH and a female child without other known risk factors accounts for 75% of DDH. Thus, careful physical examination of all infants is paramount to detect DDH early in infancy.

Natural History :

Mild dysplasia and instability of hip noted in the first few weeks of life is secondary to maternal relaxin hormone and is typically benign. Barlow-positive (subluxatable and dislocatable) hips resolve spontaneously.

Conversely, natural history of a child with severe hip dysplasia (dislocated hip by walking age) is less satisfactory. Without treatment they would develop limb length discrepancy and premature arthritic changes at hip, knee and spine. The rate of avascular necrosis of femoral head increases with advancing age of patient at the beginning of the treatment. Thus, early identification and treatment is important to prevent DDH related sequela.

Screening and Diagnosis :

American Academy of Pediatrics (AAP) has come up with the clinical practise guidelines for early detection of DDH. They recommended periodic newborn physical examination surveillance, rather than a routine screening of all the infants to avoid late detection of DDH after six months of life, which invariably requires surgical treatment. A simplified algorithm is generated based on positive examinations, warning signs and risk factors. (Figure -1)

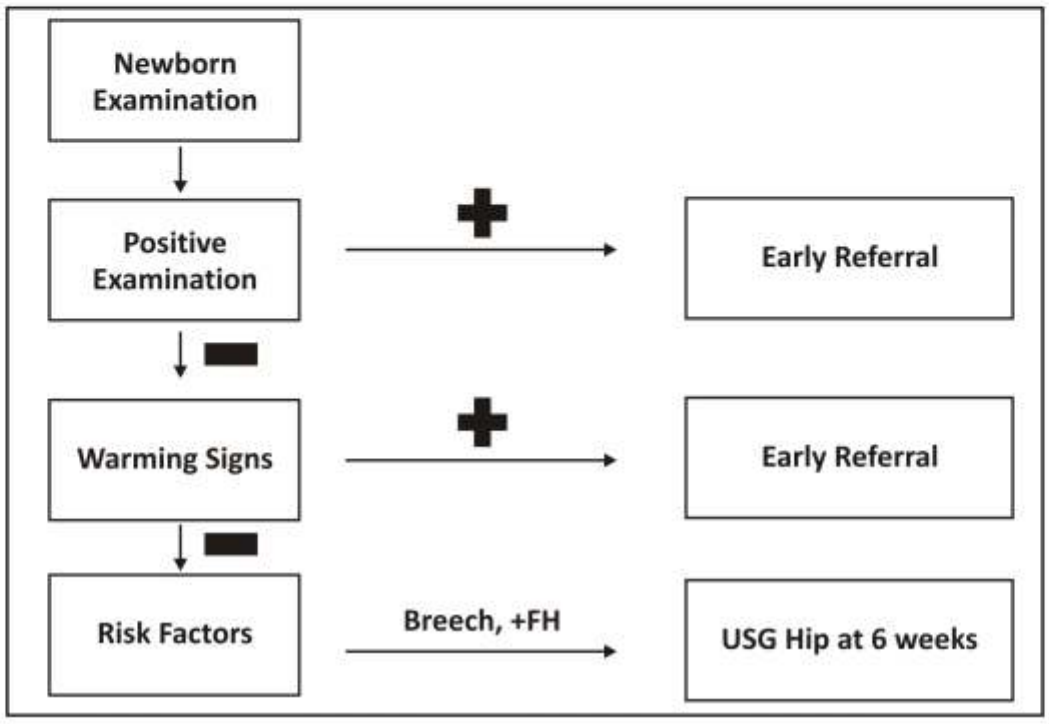


Figure 1: A simplified algorithm suggested by American Academy of Pediatrics (2006). Positive Examination includes Barlow-positive or Ortoloni-positive hips. Warning Signs include assymetric reduced hip abduction, unequal thigh or gluteal folds and short fermoal segment (Galliazi's Sign). Risk Factors include a breech delivery and positive family history. For patients with negative examination and warning signs but present risk factors, USG hip is done at 6 weeks because the maternal hormone related laxity settles by this time.

Physical Examination :

The Barlow & Ortoloni Tests: (Positive Examination Tests)

(Figure -2)

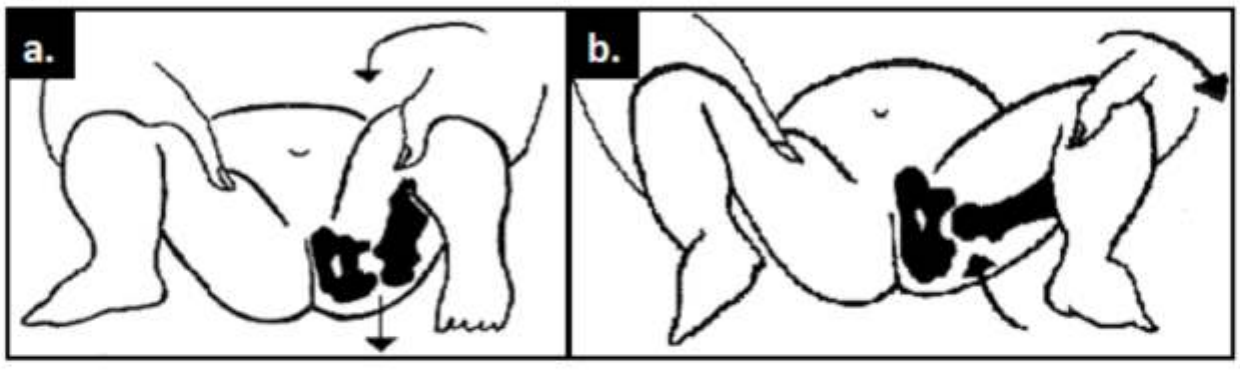


Figure 2: a. Barlow Manoeuvre is performed by gently applying posterior force while hip is adducted. This is the test of hip instability. b. Ortoloni Manoeuvre is performed by abducting the adducted thigh with anteriorly directed pressure exerted over greater trochanter. It is the test of an already dislocated hip's reducibility.

The Barlow manoeuvre is performed by adducting the hip to the midline and gently applying posterior force. A positive Barlow result is when the femoral head subluxates, and a clunk is felt. A Barlow-positive hip indicates that the femoral head is resting in the acetabulum but has pathologic instability (Subluxatable hips). Barlow manoeuvre should be performed gently to prevent iatrogenic instabilty.

With the thighs adducted and posteriorly depressed, the Ortoloni manoeuvre is performed by abducting the hips while applying anterior-directed pressure at the greater trochanters. An Ortoloni test is considered positive when the femoral head relocates with a distinct clunk. An Oroloni-positive hip is more severe than Barlow-positive hip because it indicates that the femoral head is dislocated at rest.

Both these tests are difficult to elicit after 3 months of age, as the dislocated hips become fixed by this time. "Hip clicks" without the sensation of instability are clinically insignificant.

Warning Signs :

The 2000 AAP guidelines mentioned leg length discrepancy, thigh-fold asymmetry and limited hip abduction as warning clinical signs for detection of DDH. Leg length discrepancy (LLD) should be assessed with infant in supine position, hips and knees flexed to 90°. A discrepancy is indicated by unequal knee heights, which is termed as Galeazzi's sign (Figure -3). Asymmetric thigh folds and assessment of LLD is prone to error and inaccuracy. The most sensitive examination for unilateral dislocation is asymmetric diminished hip abduction (Figure -4).

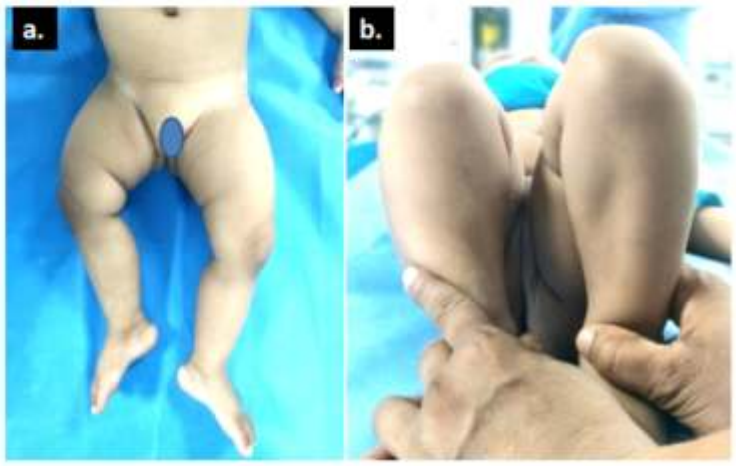


Figure 3 : Warning Signs. a. Unequal thigh folds. b.Apparent shortening of thigh (femoral) segment – Galeazzi's sign. This is measured with hip flexed at 90°.

Figure 4. Warning Sign. Restricted abductor spread is evident on left side compared to the right.

The walking child may present with a short limb or a Trendelenburg gait. Child might keep the longer limb (unaffected) flexed at the knee and the affected limb straight while standing. At times, they might keep the affected side heel off while walking. Children with bilateral hip dislocation demonstrate waddling gait, increased lumbar lordosis and prominent lower abdomen. (Figure -5)

Figure 5. A 3 years old girl presented with prominent buttocks, prominent lower abdomen, waddling gait and exaggerated lumbar lordosis. X-ray revealed bilateral hip dislocation and acetabular dysplasia.

Diagnostic Imaging :

Ultrasonography : (Figure - 6)

Hip ultrasonogoraphy (USG) is the recommended imaging modality in infants < 4 months of age because the infant hip is predominantly cartilagenous, precluding clear visualisation on conventional x-rays. USG can be performed in a static or dynamic manner. In a typical static hip USG image, three parameters should be observed. Alpha angle (α -angle) or the acetabular roof angle should be more than 60°. Beta angle (β -angle) suggests lateral labral coverage of femoral head and less than 50° is considered normal. More than 50% of femoral head should be covered by acetabulum. A dynamic ultrasound can demonstrate stability of joint with stress manoeuvres. USG is used for initial diagnosis and to follow the progress in infants receiving Pawlik Harness treatment.

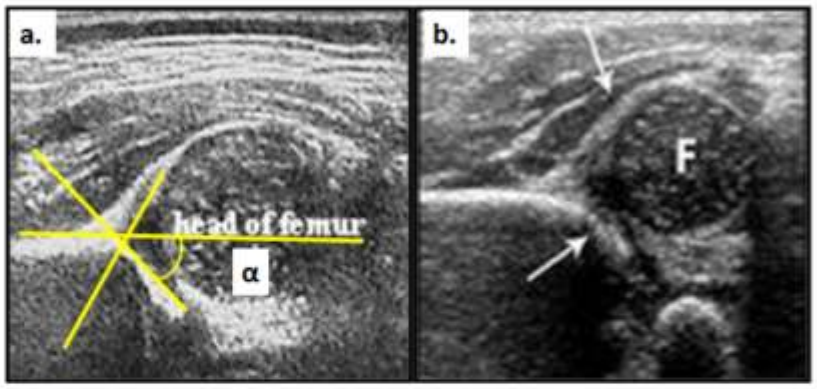
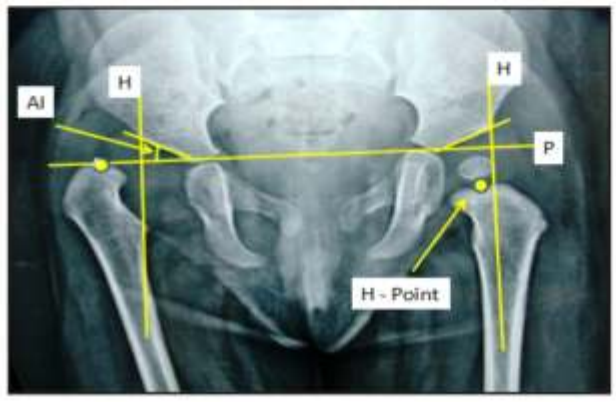


Figure 6. A typical Ultrasound of hip. a. Ultrasound of unaffected hip. Angle between the central ilial line and acetabular roof suggests alpha angle (n=>60°). Half of the femoral head lies within the confines of acetabulum. Angle between the central ilial line and labrum suggests beta angle (n=< 50°). b. Ultrasound of affected hip showing femoral head is subluxated out of the acetabular confines and acetabulum is dysplastic (alpha angle = < 60°).

X-rays :

After 6 months of age, femoral head ossification reliably appears and x-rays are the preferred method of evaluating and monitoring DDH. It is important to note that femoral head ossification is delayed in the dysplastic hips. International Hip Dysplasia Institute (IHDI) has proposed a classification of hip dislocation based on pelvis x-ray. Details of parameters are explained in Figure -7.

Figure -7. Typical measurements on pelvis with both hip x-ray. H-line is the Hilgeinreiner's line drawn from the outer margin of the acetabulum. Perkin's line passes through the triradiate cartilage. H-point is the center of proximal femoral metaphysis. When H-point lies in upper & lower outer quadrant, it is a dislocated hip (according to IHDI classification). A line is drawn from the triradiate cartilage to the outer margin of acetabulum. Angle whic it intersects with Perkin's line is known as Acetabular Index. Normal Acetabular Index is 15°. Higher angle suggests Acetabular dysplasia. (H – Hilgeinreiner's Line, P –Perkin's Line, AI – Acetabular Index).



Treatment : (Table -1)

Age of Patient	Preferred treatment
Up to 6 months	Pavik Harness
6 months to 12 months	Close Reduction of hip
12 months to 24 months	Close Reduction of hip
24 months to 8 years	Open Reduction of hip ± Femoral Osteotomy ± Pelvic Procedure
More than 8 years	No treatment advocate for bilateral dislocations For unilateral dislocations, treatment is offered with possible high complication rates.

Early Brace Treatment :

For infants up to 6 months of age, the Pavlik Harness has been classically used for stabilization of hips. Pavlik Harness of different sizes are pre-fabricated. Brace should be worn for full time and periodic follow up examination should be carried out. At one week follow up, infant is checked for any harness related skin irritation or a rare femoral nerve palsy which can happen due to excessive flexion in harness. All these issues are reversible on adjusting the harness straps.

Infants are followed every 2 weeks with a hip ultrasound to assess the regression of acetabular dysplasia and improved hip coverage. Typically, infants need 4 -6 weeks harness treatment in most cases. Harness should be discontinued when the hips do not respond to the treatment for 4 weeks. The Barlow-positive hips has shown >90% successful stabilization with a Pavlik harness. The Ortoloni-positive or initially dislocated hips have had Pavlik harness failure in about 25% of patients. (Figure -8)

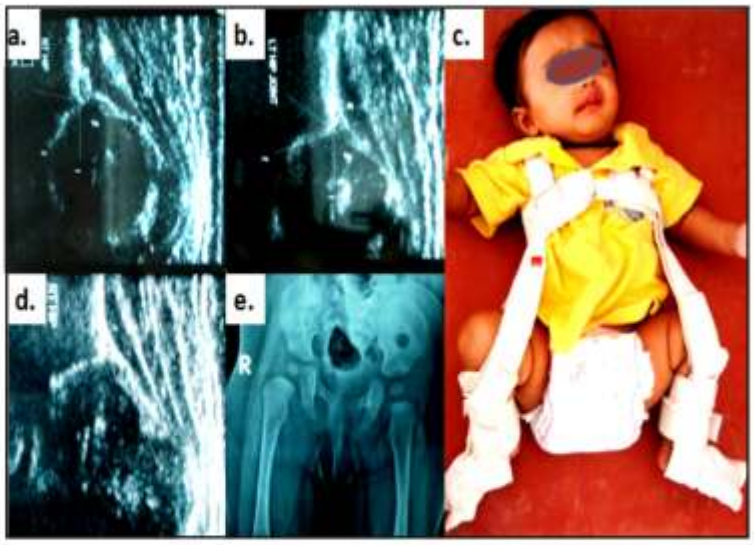


Figure -8. A 2 months old breech delivered baby presented with assymetric abduction at hips. Her right side abduction was 20° compared to 50° on the eft side. a. Right hip Ultrasound suggests subluxated hip joint with majority of the head lying outside the ilial line and dysplastic acetabulum (alpha angle=35°). b. Left hip Ultrasound suggested well centralised hip joint with > 50% of head lying within acetabulum and well developed acetabulum (alpha angle =65°). c. Patient was presecribed Pavlik Harness treatment. d. Four weeks follow up ultrasound shows improved alpha angle to 60° and head well covered by acetabulum. Harness application stopped at 6 weeks. e. Pelvis x-ray at the age of 1 year suggests well located hip & nicely developed acetabulum.

Patients with an Ortoloni-positive hips who fail to stabilize with initial Pavlik harness treatment, are offered a trial of more rigid hip abduction orthosis. In a recent series, about 82% hips got stabilized with this alternate pathway and a surgical intervention could be avoided.

Surgical Treatment :

Children between 6 and 12 months of age typically are amenable to close reduction of hip joint. Adductor tendon is percutaneously tenotomised and a stable hip reduction is ensured by checking its stability through a wide range of safety zone. When femoral head is non-ossified, a radio-opaque dye is injected in the joint (arthrogram) to visualise the outline of the femoral head and quality of reduction. A hip spica is required to be kept on for abou 8-10 weeks. (Figure -9)

Figure s9. A typical hip spica. The spica extends to the lower abdomen and both the legs to hold the hip joint in position. A bar between legs is applied to improve the stability of the spica.

Children between 12 months to 24 months may fail to reduce by close means. They need open reduction of the hip joint. Tight muscles surrounding the hip joint are released and intra-articular obstacles to reduction are removed to achieve a stable reduction. Capsulorrhaphy is performed to secure the joint stability.

Children older than 24 months to 30 months need additional femoral or acetabular procedures to ascertain stable hip reduction. All these procedures are done in a single stage. (Figure -10)

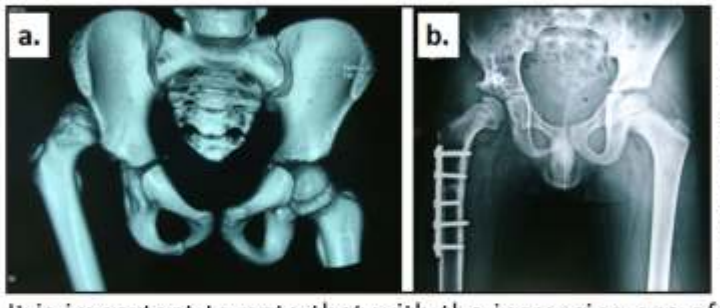


Figure -10. a. An eight years old boy presented with short limb gait and limited abductor spread on right side. Radiography suggested a high dislocation of right hip with dysplastic acetabulum. b. Patient required open reduction of right hip, femoral shortening-derotation osteotomy and acetabuloplasty to improve the inclination of the acetabulum.

It is important to note that with the increasing age of the patient, the chances of avascular necrosis and the complications related to the surgery are increased. Thus, importance of early identification and referral cannot be over emphasized.

Suggested Reading :

1. American Academy of Pediatrics: **Clinical Practice Guideline: Early Detection of Developmental Dysplasia of the Hip.** PEDIATRICS Vol. 105 No. 4 April 2000
2. Evaluation and Referral for Developmental Dysplasia of the Hip in Infants. Pediatrics December 2016, 138 (6) e20163107.