Case Report

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Radiological and Functional Outcome of Management of Bilateral Congenital Dislocation of Hip in A 1-Year-Old Infant : A Case Report

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Abstract

Congenital dislocation (CDH) of the hip joint (now falling under the umbrella term Development Dysplasia of hip (DDH) has a high incidence in the Indian subcontinent. Even though the number of reported bilateral CDH cases are comparatively less, a large number go under the radar due to similar presentations on both sides due to which a medical opinion is not sought by the parents until the walking age. Herein lies the need for a more extensive screening protocol. Such cases, when presented late, require surgical correction in cases where conservative management becomes redundant. Literature is divided on the course of management of such cases and we wish to present one such case here.

Keywords: CDH, Congenital Subluxation of HIP, Derotational Osteotomy of HIP.

Introduction

Developmental dysplasia of the hip is a spectrum of structural abnormalities that involve the growing hip and represents a wide spectrum of pathologic conditions, ranging from subtle acetabular dysplasia to irreducible hip dislocation with proximal femoral displacement. It has been seen that the majority of abnormalities arise as a result of maldevelopment of the acetabulum.[1] The acetabulum is known to be the primary culprit in such cases, however, as a result of a disturbance in the normal biomechanics of the hip joint secondary to acetabular dysplasia or malformation, the femoral head is involved. It is known to be associated with other severe malformations, such as spina bifida, arthrogryposis multiplex congenita, lumbosacral agenesis, chromosomal abnormalities, diastrophic dwarfism, Larsen syndrome and other rare syndromes.[1] The incidence ranges from as low as 1 per 1,000 to as high as 34 per 1,000. Higher incidences are reported when ultrasonography is also used in addition to clinical examination owing to its sensitivity.[2] Other associated factors include ethnic background (e.g. native Americans who use swaddling that forces the hips into extension and adduction), torticollis and lower limb deformity. There are a number of predisposing factors that lead to the development of DDH, including ligament laxity, breech presentation, postnatal positioning and primary acetabular dysplasia, female new-borns, breech presentation and oligohydramnios.[3,4]

Case Report

A 1 year and 3-monthsold female child was brought to the OPD by her parents, with complaints of inability to stand or walk. Upon examination, it was noted that the attitude of bilateral hips was in flexion and adduction along with restricted extension, external rotation and abduction. The hip was otherwise painless and stable. On plain radiography of pelvis with both hips it was noted that the epiphysis of both the heads were displaced from their respective acetabulum and situated in the supero-lateral quadrant formed by the intersection of Perkin's and Hilgenreiner lines. The

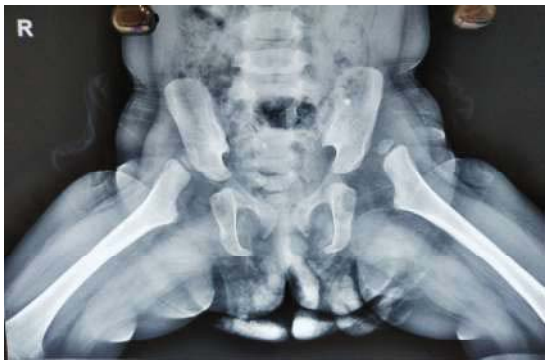

Fig. 1: Frog leg view upon presentation

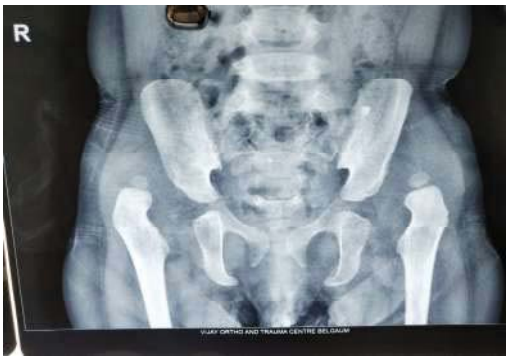

Fig. 2: Hip Antero-Posterior view showing femoral head epiphysis in the supero-lateral quadrant

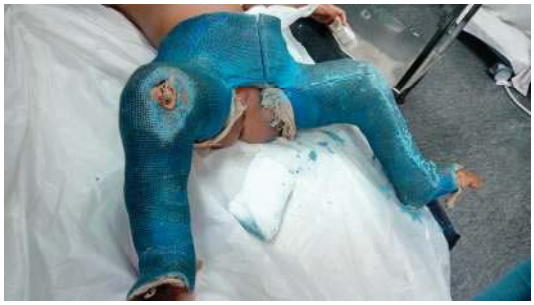

Fig. 3: Post-operative hip spica application

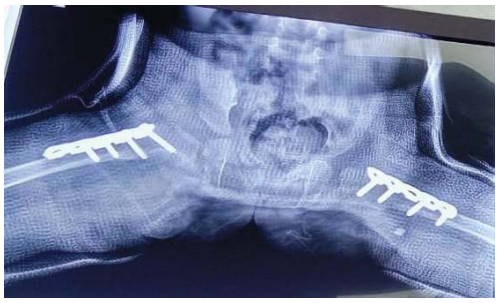

Fig. 4: Post-operative and hip spica application radiograph

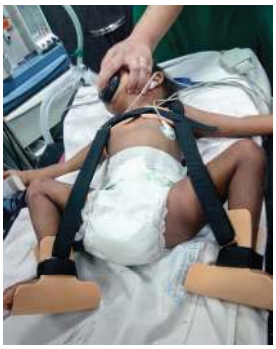

Fig. 5: Removal of hip spica and application of Pawlik harnes

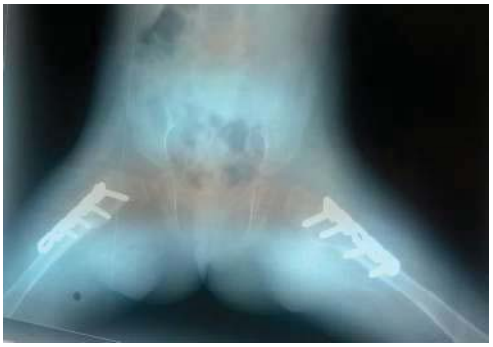

Fig. 6: Follow-up image showing well contained femoral head in frog leg view

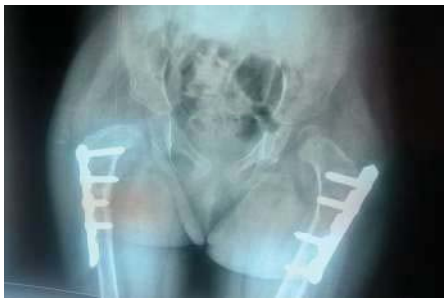

Fig. 7: Follow-up images showing well contained femoral head in antero-posterior view

heads on either side appeared to articulate with a false acetabulum superiorly in the frog-leg view (Severin type IV) (Fig. 1, Fig. 2).

Having taken the severity of the condition into consideration, surgical intervention was planned. The patient underwent percutaneous tenotomy of adductor longus along with the release of contractures on the medial aspect. This was followed by varus de-rotation osteotomy with shortening of the femur and fixation with

plate and screws. After the relocation of the head was confirmed on one side, the same was repeated on the contralateral side in the same sitting. After both the surgical sites were closed, a hip spica was applied to the patient with both hips flexed and abducted and knees flexed. (Fig. 3, Fig. 4).

The patient was discharged from the hospital in stable condition and no complications. After 3 months, the spica was removed under short general anesthesia and the patient was placed in a Pawlik harness for a further two months (Fig. 5).

At 5 months follow-up, the epiphysis of the head was well maintained in the acetabulum along with healing of the osteotomy sites bilaterally (Fig. 6, Fig. 7). Thepatient started walking without limp. The follow-up is intended to be continued for a period of 1 year.Functional outcome was assessed by the range of motion at both hip joints and ability of the infant to ambulate with a normal gait.

Discussion

The first reported case of hip instability in the literature was done by Roser in 1879. In 1910, Le Damany and Saiget described a clinical test for hip instability which was highlighted in 1937 by Ortolani. Palmen in 1961 and Barlow in 1962 developed further tests to induce dislocation or subluxation.[5] Positive Ortolani test represents the reducibility of a dislocated hip into the acetabulum. A positive Barlow's test represents potential subluxation or dislocation. After three to six months, soft tissue contractures (most commonly adductors) limit motion of the hip despite being dislocated.[6] Hence, the examination of an older child requires careful assessment of extremities for asymmetric skin folds, leg length discrepancy (in case of unilateral hip dislocation), limited abduction (in bilateral cases where Galeazzi sign is negative). In neglected cases, DDH is generally diagnosed when children approach the walking age due to a limp on the affected side (positive Trendelenburg's sign) and/or hyperlordosis. As the child reaches three to six months of age, the dislocation will be evident on plain radiographs. In a normally located hip, the medial beak of the femoral metaphysis lies in the lower, inner quadrant produced by the intersection of Perkin's and Hilgenreiner's lines, whereas in dislocated hips, it lies in the superior and lateral quadrant. In the dislocated hip, Shenton's line is broken because the femoral neck does not lie in continuity with the pubic rami. The Acetabular index is another useful measurement, formed by the junction of Hilgenreiner's lineand a line drawn along the superior aspect of acetabular surface or the roof. In normal newborns, the acetabular index averages 27.5 degrees, at six months 23.5 degrees and at two years, 20 degrees.[18] Thirty degrees is considered to be the normal upper limit.[19] In older children, the centreedge angle is a more useful measure. In children aged 6–13 years, an angle less than 19 is considered abnormal, whereas, in older children, an angle lesser than 25 is considered abnormal.[7,8]

The treatment of DDH is age-related and the goal is to achieve and maintain concentric reduction of the femoral head into the acetabulum. The outcome is in direct correlation to how early the treatment is initiated. Due to a dearth of awareness and training, surveillance (use of ultrasonography), and pediatric orthopaedic surgeons, the developing countries lag behind.[9] When conservative management is preferred, the child should then be re-evaluated both clinically and by ultrasound at three weeks of age to confirm concentricity. Hips that are still dislocated need further treatment. Orthoses, such as Erlanger, Thübinger or Pavlik, can reduce the incongruous pressure to the anterolateral acetabulum and is preferred in this age group. Application of an orthosis should be followed by bi-weekly clinical examination, and ultrasonography, if required. If the hip is reduced at three weeks follow-up, the patient may continue to wear it for a further three weeks. After six weeks, if the hip is reduced then the orthosis can be discontinued. The dislocated hip, even after 3–4 weeks of orthosis use, should be evaluated and may be treated with an abduction brace.[10,11] The Pavlik harness is contraindicated when there is major muscle imbalance, as in myelomeningocele (L2 to L4 functional level); major stiffness, as in arthrogryposis, ligamentous laxity, as in Ehlers-Danlos syndrome' or where the chances of non-compliance are high. If the hip fails to reduce with orthosis then other options should be considered, such as an abduction (Von Rosen) splint. The main aim of treatment is to achieve concentric reduction and to prevent complications such as avascular necrosis. Nakamura *et al*. reported his results of 115 patients with 130 hips. The mean age was 4.8 months and there was a mean follow up of 16 years. Patients were treated with a Pawlik harness with a mean duration of treatment of 6.1 months. Twenty-two hips required supplementary surgery for residual dysplasia, the choice of surgery depending on the state of joint and surgeon's preference. A satisfactory outcome (Severin classes I and II) was achieved in 119 patients.[12]

Children of age group 6 months to 2 years may be treated with either closed or open reduction, followed by a spica cast, as has been done in our case. The aim is to achieve reduction without damaging the femoral head. We undertook this procedure as there are several studies favouring a reduction of hips after the appearance of the ossific nucleus.[13] Segal *et al*. reported 57 hips in 49 children under 12 months of age. Thirty-eight hips were reduced closed while 17 were reduced by an open method. One patient with bilateral hip dislocation was treated initially by closed means and later treated by open reduction at three months. Avascular necrosis (AVN) developed in only one of 25 patients in which a nucleus was present while 17 of 32 patients developed AVN when a reduction was performed before the appearance of an ossific nucleus at a mean follow up of 59 months.[14] There are studies which contradict this approach too such as that by Konigsberg *et al*. wherein 40 patients in whom an open reduction through a medial approach was performed. The average age was 7.7 months, ranging from 2.4 to 18.9 at the time of surgery, with a mean follow up of 10.3 years. Only one of 20 hips which was reduced before the age of six months, went into AVN.[15] Belated reduction of hips leads to unsatisfactory remodelling due to reduced growth potential.[16]

Open reduction may be achieved through a medial (preferred due to minimal dissection and ease of contracture release).[17] Disadvantages of the medial approach include inadequate exposure, risk to medial circumflex femoral vessels and inability to perform capsulorrhaphy. Post-operatively, a cast is recommended for a total period of 3 months and changed after six weeks. A medial approach is recommended for children with a maximum age limit of 18 months in expert hands. Treatment of older child (two years of age and older). In older children, the femoral head lies more proximally. Sankar *et al*. studied the factors predicting the need for femoral shortening in 72 hips which underwent open reduction. He concluded that the patients over the age of 36 months and patients with a vertical displacement greater than 30% of the width of the pelvis were more likely to require femoral shortening.[18] The aim of femoral de-rotation varus osteotomy is to achieve concentric reduction. Spence *et al*. compared two groups of patients undergoing open reduction through an anterior approach either with a femoral de-rotation osteotomy (38 patients with 47 hips) or innominate osteotomy (33 patients with 37 hips).[19] Adequate post-reduction coverage of the femoral head is essential which was achieved in both the procedures. If femoral head coverage is inadequate, then pelvic osteotomy (Salter innominate or Pemberton) should be considered.[20] In cases of areduction under 18 months of age, the child should be followed up untill the age of 3.5–4 years. The incidence of AVN varies widely from 0–73% depending on the age, mode of treatment and criteria used to describe AVN.[21] Four per cent incidence was reported by Weiner *et al*. in children under the age of three months.[22] Literature says that AVN can easily be prevented by performing femoral shortening which is why we undertook this approach.

Inability to diagnose DDH may lead to any of the following four sequelae: the hip may reduce spontaneously, it may subluxate, it can develop frank dislocation, or it can develop dysplastic features whilst remaining located.[23] Long-term follow up of treated subluxated and dysplastic hips also revealed a higher incidence of degenerative joint disease in hips. The reasons behind this phenomenon are probably mechanical and related to a high degree of contact stress over-time on a relatively small surface area.[24]

Conclusion

A high degree of suspicion is required to diagnose DDH in its early stages to prevent surgical management. In delayed cases, despite there being many schools of thought on the management of bilateral DDH in children, there is little consensus as to which is the most appropriate approach. In this case, we operated the patient bilaterally in one sitting, at 1 year and 3 months of age, owing to the severity of displacement and soft tissue contracture. The fact that the patient has improved, both clinically and radiologically after removal of the Pawlik harness justifies this approach and provides evidence for its use in the future. The weakness in the reporting of this case is that it is to be viewed as a separate entity and a further research on a larger population is warranted.

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