

Late-onset hip dysplasia and dislocation in Osteogenesis Imperfecta

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ABSTRACT

Background: Skeletal deformities and multiple fractures are the most common features of osteogenesis imperfecta which frequently has been managed using intramedullary nails. Likewise, hip dysplasia is considered a rare complication of osteogenesis imperfecta. Hereby we want to describe a patient with osteogenesis imperfecta who developed late-onset hip dysplasia and subluxations.

Case presentation: Here, we presented a 7-year-old girl, known case of osteogenesis imperfecta, who was referred to our clinic for limping and limb length discrepancy. During the radiological assessments, we found increasing acetabular index and mild hip dysplasia as well as coxa valga while the previous radiographs were indicated normal hips. Salter osteotomy was performed to improve the anterior coverage of the left side and Dega osteotomy was performed on the right side to improve anterior and posterior coverages. In further follow up both hips were reduced and acetabular indexes seem normal.

Conclusion: Femoral intramedullary rods insertion can destroy the physis of the greater trochanter which conduce to coxa valga. Hyperlaxity along with valgus deformity led to hip subluxation and dysplasia.

INTRODUCTION

Brittle bone disease or osteogenesis imperfecta (OI) is understood as a rare collagen-related disorder with a prevalence rate of one in 15–20 000 births.^[1,2] The aforementioned collagen abnormality conducted to various and widely skeletal and extraskelatal manifestations depending on the age of patients as well as the severity of the disease. Regarding the skeletal presentation, patients usually presented with growth deficiency, bowing of long bones, chest wall deformities, spine curvature, coxa profunda, and pathological fractures.^[2] Likewise, the optimal management for femoral varus deformity and pathological fractures which are the common features of OI were recommended as using intramedullary fixation. The greatest results for growing bone stabilization have been reported inserting telescoping rods even though generalized ligamentous laxity which is commonly seen in these patients can lead to unwanted complications during the management.^[3, 4] Acetabular dysplasia is reported rarely in accompany with OI and its optimal treatment plan and surgical technique are often performed according to the surgeon's discretion.^[5] This perusal described the clinical manifestation and management

course of an OI patient who developed late-onset acetabular dysplasia and coxa valga along with hip subluxation and managed by corrective surgeries.

Case report

A 7-year-old girl who was known case of OI came to our clinic with the chief complaint of limping and shortness of the left lower extremity. She was the first child of the family and her parents were cousins. She had multiple fractures from 40 days of age and had a history of four previous surgeries for fixation of her right and left femur and right elbow from 3 years of age (Rash nail insertion of Right (2times), left femur, and right elbow). Her parents claimed no significant family history but on inspection short stature of her mother and her blue sclera were obvious. It's worth mentioning that the patient had prosthetic heart valves due to mitral regurgitation. The limping had been gradually worsening after the last previous surgery for exchanging the intramedullary Rash nail of the right femur. On inspection, she was a noticeably short stature girl with 110 cm height and an obvious waddling gait without any obvious rotational or deformity in lower extremities. The true length of the left extremity was 4 cm shorter than the

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right one. There was no limitation of motion or pain on motion in the knee, hips, or ankle joints. The flexion arch of both hips is from zero to 150 degrees and the rotational arch of both hips is from 40 degrees of external rotation to 90 degrees of internal rotation. She has generalized laxity and fully filled all Beighton criteria for generalized laxity. [6] Pelvic radiography showed bilateral hip subluxation and bilateral coxa valga. Both sides' acetabular indexes were 20 degrees which were above the normal range. There was the intramedullary nail in each femur inserted from the tip of the greater trochanter on each side (Figure.1).



Figure: 1 Intramedullary nail in each femur. Both acetabular indexes are above the normal range. Both hips were subluxated with severe valgus deformity. This is the first time she was referred to our clinic. (7 years old).

In the review of medical records, we found that she had multiple previous surgeries. In the pelvis x-ray that was taken 3 years prior to this admission, both hips were reduced and both sides' neck-shaft angles are lesser than the current presentation especially on the left side. Furthermore, the acetabular index was normal on both sides (Figure. 2). Further radiographic studies also revealed a gradual increase in neck-shaft angle and acetabular index.



Figure: 2 Both hips were reduced. The acetabular index was normal on both sides. (4 years old)

To evaluate acetabular dysplasia and deficiency and any possible rotational deformity of the femur, computed tomography (CT) scan was done. It should be considered that MRI may be the preferred

imaging method in children regarding the amount of radiation, however, we choose CT scan since the patient had MR-incompatible prosthetic heart valves. The right and left side anterior acetabular deficiency and also femoral anteversion were detected. Therefore, we decided to perform open reduction and salter osteotomy to improve anterior coverage. To decrease the amount of limb length discrepancy we used a trapezoidal graft (Figure. 3).



Figure: 3 Left side open reduction and salter osteotomy with trapezoid graft.

Four months after left hip surgery, the left hip was reduced, and the right side was dislocated. Therefore, right side surgery for open reduction and acetabular osteotomy had been planned. At the time of surgery, we found that there was a global acetabular deficiency mainly anteriorly and also not enough posterior coverage. therefore, we decide to perform Dega osteotomy for the right side.

Seven months after the second surgery, both hips were reduced and there was only 2cm LLD (Figure. 4).



Figure. 4 Both hips were reduced 7 months after the last surgery (9 years old).

On the last follow-up 19 months after the last surgery, she walked independently and there was a significant improvement in limping. Both hips were still reduced. The limb length discrepancy was about 2 cm.

DISCUSSION

It has been thought for a long time that OI accompanies by the acetabular protrusion or coxa profunda till several case reports claimed that hip dysplasia can also see in these patients. Likewise, primary reports attributed the main etiological factor of hip dysplasia to joint hypermobility and laxity.[7,8] The forementioned laxity is also documented in our case which is thought to have contributed to hip subluxation. Similarly, Kishta et al. documented that their patients with developmental dysplasia of hip and OI also had joint hyperlaxity

simultaneously.⁵ Regarding the treatment method of hip dislocation, the Pavlik harness was frequently criticized, not only due to failure in hip reduction but also because of augmenting the risk of avascular necrosis and more fractures. The optimal treatment was achieved via correcting femoral deformities and close or open reduction with or without pelvic osteotomies.^[5, 9, 10] Likewise, open reduction was suggested in patients older than 18 months old.^[3] Regarding our patients, concerning anterior coverage insufficiency on the left side, salter osteotomy was done and due to anterior and posterior insufficiency of the right side, Dega osteotomy was performed. On the further follow up both hips were reduced and located appropriately.

Multiple long bone fractures and deformities are among the most common manifestation of osteogenesis imperfecta which are usually managed with intramedullary nail insertion. The aforementioned nails are needed to exchange as the child grows.^[11] Placement of an intramedullary rod in a long bone is encouraged frequently for stabilizing a severe fracture, internal support establishment for healing following correction of bone deformity as well as to interrupt fracture or disuse cycles.^[4,13] Telescopic nails were introduced as the most appropriate ones in these patients. However, due to the inaccessibility of the foregoing nails in our region, the rush nail was used. Furthermore, the femoral valgus deformity was presented in our patient, contrary to what is commonly seen in OI (varus deformity). We supposed that rush nail insertion from the origin of the greater trochanter's tip could destroy the physis of the greater trochanter. Therefore, it was considered as the main cause of the femoral valgus deformity. The uncommon valgus deformity along with generalized ligamentous laxity conducted to hip dysplasia, increasing acetabular index, and hip subluxation and dislocation. Of course, we can't certainly claim which factor may result in the aforementioned effect during the management, but the possibility of them should be considered. Moreover, the potential positive effect of drugs such as Bisphosphonates as well as bracing to help protect the patient during physical activity should be noticed. The multidisciplinary comprehensive approach not only improves the outcomes^[14] but also may decrease the need for frequent surgical management and its complications.

Multiple surgeries not only increase the risk of physis injuries but also raise the possibility of surgical related infections. Moreover, it is accompanied by excessive anesthetic drug exposure which has the related central nervous system adverse effect particularly for children^[15-17].

CONCLUSION

To sum up, everything that has been so far, we recommend avoiding multiple surgeries as much as possible and if it is needed to insert a nail from the tip of the trochanter it should be done with minimal damage to the growth plate. That's all that can prevent the femoral valgus deformity as the associated factor in hip dysplasia.

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