Open Reduction at 15 Months of Left Hip Dislocation in a Male Infant Diagnosed with Arthrogryposis

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Patient: Male, 4-week-old
Final Diagnosis: Arthrogryposis
Symptoms: Limited hip motion
Medication: —
Clinical Procedure: Open reduction of hip and femoral shortening
Specialty: Orthopedics and Traumatology

Objective: Unusual or unexpected effect of treatment
Background: Arthrogryposis is a congenital condition of multiple contractures of joints associated with hip dislocation. The outcome of open reduction of hip dislocation in arthrogryposis patients is debatable. Open reduction of arthrogryposis is challenging for shallow acetabulum and extensive adhesions and fibrosis. For this reason, a careful extensive release must be carried out to achieve the open reduction of the hip in arthrogryposis patients. The literature lacks surgical recommendations for open reduction of the hip in arthrogryposis patients and how to deal with cases of the extruded bone segment during open reduction.

Case Report: The patient presented in the first few weeks of life with bilateral clubfoot and left hip dislocation. Clinical diagnosis of arthrogryposis was made after referral to a genetics specialist. The hip was clinically irreducible. The patient underwent open reduction and femoral shortening using the Smith Peterson approach at the age of 15 months, with accidental extrusion of the proximal femur, which was retained immediately. The clinical outcome showed a painless, good range of motion. Radiographically, features of avascular necrosis and healed osteotomy site were evident.

Conclusions: A difficult hip reduction was expected in this arthrogryposis patient, which required careful dissection of surrounding fibrosis and appropriate femoral shortening. Careful dissection should be carried out during open reduction to avoid jeopardization of femoral head vascularity or even complete devitalization of the proximal femur.

Keywords: Arthrogryposis • Femur Head Necrosis • Hip Dislocation, Congenital

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Background

Arthrogryposis is a congenital, non-progressive joint contracture that can present in isolated contracture form or multiple involvements [1]. Arthrogryposis affects 1: 3000 live births [1]. Arthrogryposis is characterized by normal neurological examination and the patients have normal intelligence [2]. Patients can present with upper and lower extremity contractures [2]. Clubfoot is considered one of the most common presentation pictures [2]. Hip involvement is seen in 80% of arthrogryposis patients; either in the form of hip contracture or hip dislocation [3,4]. Hip dislocation in arthrogryposis is considered a teratological type of hip dislocation since the pathology starts during intrauterine development [5]. The typical rotation of the dislocated hip is anteversion, but in arthrogryposis cases, rotation varies from anteversion to retroversion [5]. The acetabulum is considered shallow, which is a predisposing factor to hip dislocation in arthrogryposis patients [5]. Hip dislocation is present in 30% of arthrogryposis patients, and 50% of them present with bilateral hip dislocation [6]. Closed reduction and splinting are reported to have a poor outcome in the maintenance of reduction [4]. In unilateral hip dislocation, open reduction is recommended to minimize pelvis obliquity, ease mobilization, and prevent scoliosis [6]. In contrast, for bilateral hip dislocation, some authors suggest conservative management since the pelvis is balanced and mobilization is satisfactory, whilst others suggest a bilateral open reduction to restore hip biomechanics and to avoid future hip pain or stiffness [7]. Open reduction is the recommended surgical option; furthermore, femoral shortening and derotational osteotomy must be considered [5]. In this case, we describe a 15-month-old boy with arthrogryposis who presented in the first weeks of life with bilateral club foot and left hip dislocation. The bilateral clubfoot was treated initially with serial casting and orthosis. The patient underwent open reduction with femoral shortening and derotational osteotomy with accidental intra-operative proximal femoral extrusion.

Case Report

A 4-week-old male was referred to our pediatric orthopedic clinic with multiple joint contractures of the upper extremities and bilateral foot deformity. From the history, the patient was delivered via cesarean section for breech presentation. His mother had gestational diabetes. Upon examination, no dysmorphic features were noticed. Bilateral elbow and hand contractures were evident. Left hip examination was suggestive of irreducible hip dislocation. Bilateral clubfoot was evident. The patient was referred to the genetics clinic where he was diagnosed clinically with arthrogryposis with multiple joint involvement, without further investigation. Serial bilateral foot casting was initiated, followed by percutaneous bilateral Achilles tenotomy and orthosis, which were uneventful, and full correction was achieved. His workup included a plain pelvis X-ray which showed high dislocation of the left hip and shallow acetabulum coverage (Figure 1).

At the age of 15 months, the decision for surgical intervention to treat his unilateral hip dislocation was made. The patient was prepared for surgery, and the preoperative workup was unremarkable.

Operative Technique

The Smith Petersen approach was utilized, with the fat dissected and lateral femoral cutaneous nerve secured. A window between the tensor fascia lata and sartorius was utilized. The rectus femoris was detached from the origin. Tenotomy of the iliopsoas tendon was performed with careful identification of the neurovascular bundle. Excessive femoral anteverision with diffuse adhesions was noticed around the capsule, and these were released. Capsulotomy was performed by removing intracapsular obstacles including pulvinar, ligamentum teres, transverse acetabular ligament, and inverted limbus. The head was not reducible yet, so the decision to undertake femoral osteotomy was made. A lateral proximal femur incision was made. The fat was dissected until reaching the tensor fascia band, which was splitted. Then, the vastus lateralis was splitted until reaching bone, at which point shortening of 2.5 cm with derotational osteotomy was done. The osteotomy site was fixed with a 4-hole plate and 4 conventional screws. A trial of reduction failed. The plate was removed, along with...
the screws, temporarily. A tight posterior capsule and extensive adhesions were noticed, over the posterior aspect of the proximal femur, and these contributed to the difficult reduction. During the release of extensive adhesions around the proximal femur to achieve reduction, the proximal femur was completely extruded out of the wound in the sterile surgical area and did not fall to the ground. Sterility was maintained for the extruded segment and in general during the whole course of the procedure. Before reimplanting the extruded segment, irrigation of the wound and the extruded piece was done using normal saline with the maintenance of sterility of the extruded bone. The reduction was achieved under portable X-ray after retaining the extruded part, and was fixed with a 4-hole plate and 4 screws. Capsule plication was performed.

Wound closure was performed and a hip spica was applied. The gross picture of the extruded segment of the proximal femur is shown in Figure 2, and an intraoperative plain radiograph is shown in Figure 3.

Followup

During followup, the wound healed uneventfully. Serial radiographs taken during followup demonstrated concentric reduction of the hip. Followup X-ray 6 weeks postoperative demonstrated concentric reduction and healing of the femoral osteotomy site (Figure 4). The hip spica was maintained for 6 weeks, then converted to a broomstick for another 6 weeks. The cast was removed after immobilization for 3 months. The
The first step of the procedure is open reduction, which aims to release intracapsular obstacles (pulvinar, tight transverse acetabular ligament, ligamentum teres, and inverted labrum) to release intracapsular obstacles (pulvinar, tight transverse acetabular ligament, ligamentum teres, and inverted labrum). The anterior capsulotomy.

Usually, femoral shortening is not the only factor preventing concentric reduction; a tight capsule also participates as an obstacle to reduction [7]. The medial approach increases the risk of AVN due to the higher possibility of injuring the medial circumflex artery which is the main blood supply source to the proximal femur [8]. Recent literature showed no difference in AVN risk between the 2 approaches [9]. The Smith Petersen approach is considered a safe approach in arthrogryposis-related dislocated hips even if done prior to the age of 12 months [10]. The AVN rate may reach 23% among patients who undergo open reduction of a dislocated hip [11]. Hip dislocation in arthrogryposis patients requires an extensive surgical approach due to the nature of the extensive fibrosis surrounding the capsule, which makes the reduction difficult and increases the risk of complications, including redislocation [7]. In our patient, we utilized the anterolateral approach (Smith-Petersen) and during followup, radiographs showed AVN changes. We believe this is linked to extensive dissection and devitalization of the proximal femur from surrounding structures.

Hip dislocations in arthrogryposis patients in most cases are high dislocations and therefore they require femoral shortening with an open reduction [7]. In our patient, neither open reduction nor shortening was sufficient to reach reduction due to extensive adhesions and tight posterior capsule in an antverted position. The capsule participates in redislocation due to high tension over the capsule in a reduced position. Therefore, in addition to open reduction surgery and femoral shortening, the patient required extensive release and posterior capsulotomy.

The first step of the procedure is open reduction, which aims to release intracapsular obstacles (pulvinar, tight transverse acetabular ligament, ligamentum teres, and inverted labrum) and extracapsular obstacles (iliopsoas tendon) to free the dislocated capsule to be mobilized to its anatomical location [7]. After capsulotomy to remove intracapsular obstacles, the posterior capsule must be preserved since the medial circumflex artery passes over the posterior capsule [7]. In our case, the careful release of the capsule was carried out and the posterior capsule was preserved initially. Reduction failed with open reduction alone; therefore, femoral shortening was performed, which did not help in the maintenance of reduction. Thus, the difficult reduction required extensive release of the posterior capsule, which resulted in the complete circumferential release of the capsule and thereby extrusion of the proximal femur from the surgical wound. The extruded segment was reimplemented again, and reduction of the hip was achieved anatomically. The extruded proximal femur was fixed using rigid hardware which united the bones during followup.

Despite multiple expected postoperative complications related to hip dislocation in arthrogryposis, including redislocation, avascular necrosis, stiffness, or persistent flexion deformity [7], the postoperative course of the patient was uneventful except for AVN changes on plain radiographs.

Once a bone segment becomes devitalized, the vascular supply to the segment gets jeopardized and therefore bone healing after reimplantation of the segment is questionable [12]. Multiple factors are associated with successful re-implantation, including timing from extrusion to reimplantation, rigidity of fixation, the condition of the surrounding soft tissue, and sterility of the extruded segment [12,13]. Multiple cases of traumatic extrusion of the femoral segment with successful reimplantation have been reported [13,14]. To the best of our knowledge, no iatrogenic bone extrusion has previously been reported.

For proper planning of open reduction of hip dislocation in arthrogryposis patients, extensive fibrosis surrounding the femoral head is expected [7]. Therefore, sufficient meticulous release of the capsule, specifically the posterior aspect, is required to ease the reduction of the hip [7]. The extensive posterior release might result in AVN; therefore, a careful dissection must be carried out until reaching hip reduction [7]. Usually, femoral shortening is not the only factor preventing concentric reduction; a tight capsule also participates as an obstacle to reduction [7].

Conclusions

During open reduction of the dislocated hips of arthrogryposis patients, extensive adhesions are to be expected. The very shallow acetabulum and rotation of the femoral head can contribute to preventing concentric and stable reduction. Careful
dissection should be carried out around the capsule, and the posterior capsule should be preserved as much as possible to avoid complete devitalization of the shortened proximal femur, which can result in AVN and bone extrusion. In the case of iatrogenic extrusion, sterility of the segment must be maintained, and immediate reimplantation needs to be considered and rigid fixation needs to be utilized.

References:


Department and Institution Where Work Was Done

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