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CASE REPORT PEER REVIEWED | OPEN ACCESS

Surgical hip dislocation and retinacular flap for pediatric femoral neck fracture in osteogenesis imperfecta

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ABSTRACT

Introduction: Osteogenesis imperfecta is a genetic disorder of bone formation, characterized by increased bone fragility and low bone mass. Treatment is based on a multidisciplinary approach with medical therapy and occasional orthopedic surgery.

Case Report: We report on an 11-year-old child with osteogenesis imperfecta, which presented a transcervical femoral neck pseudarthrosis after minor trauma. This condition was successfully managed with open reduction, autologous tricortical iliac bone graft and internal fixation using a safe hip surgical dislocation with retinacular flap.

Conclusion: There is very limited information concerning the treatment of a femoral neck fracture in children with osteogenesis imperfecta. The authors report on a successful case treated with an open reduction and internal fixation using a hip surgical dislocation and retinacular flap, showing how this technique could be applied to such difficult and unusual cases as this one.

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INTRODUCTION

Osteogenesis imperfecta (OI) is a genetic disorder of bone formation, characterized by increased bone fragility and low bone mass [1–4]. Most OI cases have an autosomic dominant inheritance, linked to mutations in collagen type I genes, and as such, patients with this disease have greater susceptibility to bone fractures and deformities that compromise the motor function of the affected segment [1–6]. The severity of this disease is variable, ranging from extremely severe or lethal phenotypes, to mild forms allowing good autonomy and an almost normal life [3].

The diagnosis is usually straightforward in individuals with bone fragility, positive family history, and/or extraskeletal manifestations [1–4]. However, in the absence of these features, diagnosis may be difficult and dual energy X-ray absorptiometry (*DEXA*) allied with plain radiographs are good assets to support the diagnosis and establish disease severity [1–6].

Treatment for OI is based on the intervention of a multidisciplinary team, with orthopedic surgery and bisphosphonates as the mainstay for the management

of these patients [7–11]. Herein, the authors present an unusual but successful technique applied for a unique case of a femoral neck fracture in a child with OI. We believe that this surgical option could represent an optimal solution for particular cases as the one presented here.

CASE REPORT

We present the case of an 11-year-old female child, with osteogenesis imperfecta, diagnosed at 9 years of age after an investigation performed in the setting of several fragility fractures. Following the diagnosis, a treatment based on calcium, vitamin D, and pamidronate, according to our institution protocol, was initiated.

In February 2014, the child suffered a fall from a small wall while playing, which caused a direct blunt trauma against the right hip. From that day on an inguinal pain developed, associated with right lower limb external rotation, shortening, and limited gait ability. The radiography performed revealed a transcervical fracture of the right femur (Figure 1), and as such, an orthopedic consult was scheduled. Given the patient clinical condition and fracture type, an open reduction and internal fixation using a surgical hip dislocation and retinacular flap was proposed and subsequently performed (Figure 2A and B). A month later, a loss of reduction was observable in the follow-up X-rays (Figure 3) and a corrective surgery was proposed. The surgical procedure included a new safe hip surgical approach with a retinacular flap, and a review of the initial osteosynthesis. The screws were removed, the fracture site debrided, and using an autologous tricortical iliac crest bone graft, a new anatomical reduction and fixation with screws was performed (Figure 4A and B). A spica cast was also added in the end of the surgery, which was removed three months post-operatively. Nine months after the second procedure the patient was painless, presented full ability to weight bear the affected limb, had near normal range of motion (110° in flexion, 20° in internal rotation, and only 30° in external rotation), residual limb shortening (6 mm) and plain X-rays showing consolidation (Figure 5). Five years after surgery the patient is still pain free and with full range of motion in the hip. Despite an increased limb shortening (24 mm), the 16-year-old teenager is able to perform normal daily life activities, including sports practice (Figure 6A and B).

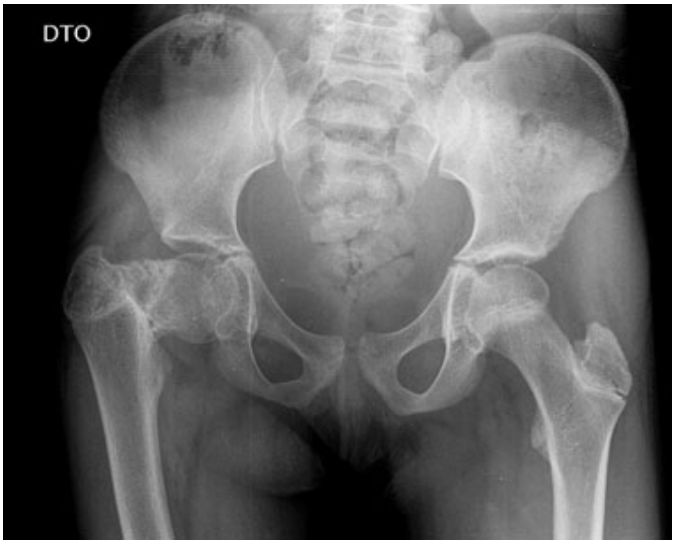

Figure 1: Pre-operative anteroposterior pelvis X-ray showing a right transcervical femoral fracture in an 11-year-old child.

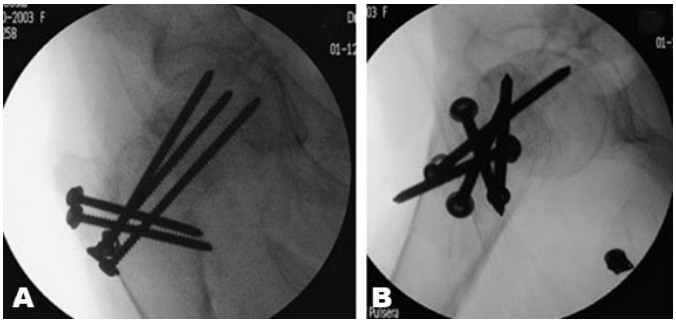

Figure 2: Final intra-operative X-rays showing the reduction achieved in the anteroposterior (A) and lateral (B) views.

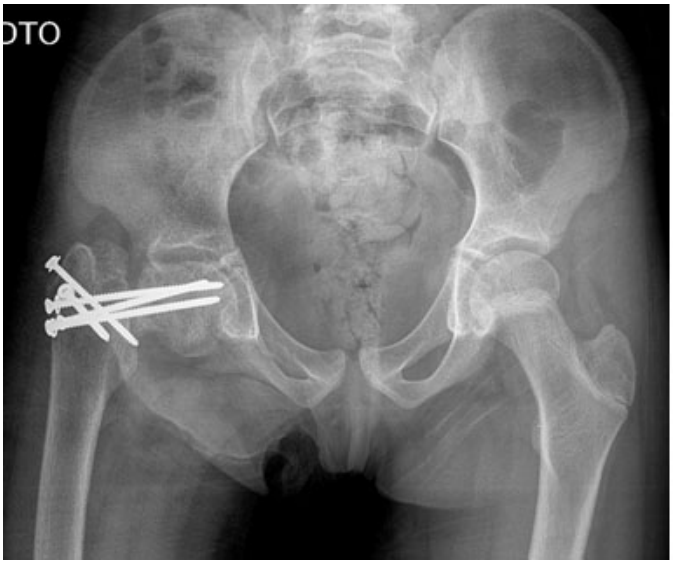

Figure 3: Post-operative anteroposterior pelvis X-ray showing osteosynthesis failure.

DISCUSSION

Osteogenesis imperfecta increases bone fragility, however, is classified in different subtypes based on genetic, radiographic, and clinical features, which can be significantly different [12–15]. In patients with identified molecular defects, OI is mainly caused by mutations in genes encoding alpha 1 and alpha 2 chains of type I collagen [16]. Type I collagen is a fundamental structural protein for the bone, tendon, ligament, skin, and sclerae, and depending on the type, clinical manifestations may

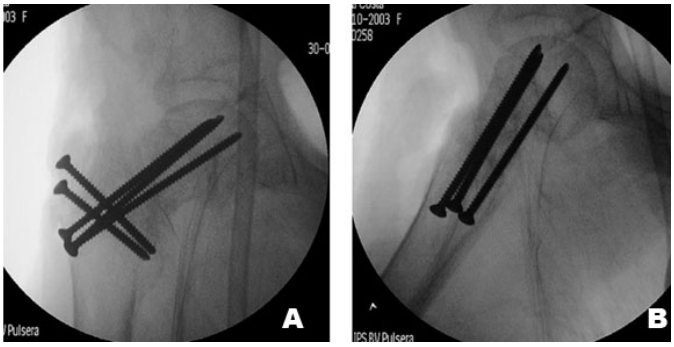

Figure 4: Final intra-operative X-rays showing the optimal reduction achieved in the second surgical procedure (anteroposterior (A) and lateral (B) views).

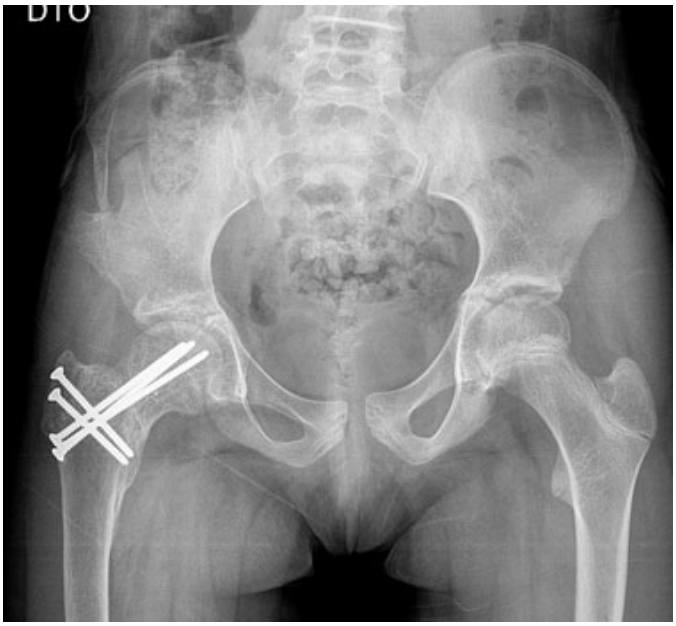

Figure 5: Anteroposterior pelvis X-ray nine months after the second surgery showing fracture consolidation.

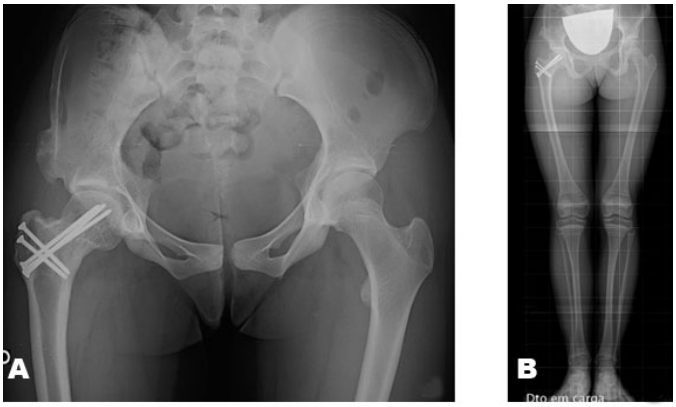

Figure 6: Anteroposterior pelvis X-ray (A) and long leg view (B) (with a right lower limb 24 mm shorter) at five years of post-operative follow-up.

vary substantially even within the same family. As a consequence, identifying a mutation in a particular gene does not necessarily result in a clear clinical diagnosis [17].

Clinical diagnosis of OI is based on signs and symptoms which includes multiple fractures with minimal or no trauma, short stature, increased laxity of the ligaments and skin, blue sclerae, or hearing loss, however, in the absence of these features, diagnosis may be difficult [1–4, 18]. There is no definitive, readily, available laboratory test for OI. Nonetheless, some laboratories made serious advances in molecular genetic testing for OI that will eventually be more accessible in a near future [1–4, 19]. We must stress that negative molecular studies for OI do not exclude the diagnosis, because false-negatives can arise in approximately 10% of cases, and some types of OI are not related to collagen type 1 mutations [18].

As stressed above, fragility fractures is one of the main features in OI, but pediatric femoral neck fractures are not frequent, and literature regarding the treatment in this particular setting is scarce [20–22]. Different surgical techniques have been proposed and applied to treat pediatric femoral neck fractures, however and to our knowledge, surgical hip dislocation and retinacular flap was never used to treat these injuries.

In 2001, Ganz et al. described a technique baptized as safe surgical hip dislocation, which is performed through a trochanteric-flip osteotomy, avoiding in this fashion disinserting external hip rotators, which contribute to the protection of the medial femoral circumflex artery (MFCA), the most important branch for femoral head vascularization [23]. For pediatric patients and following the trochanteric osteotomy, a capsulotomy (usually Z-shaped) and the development of a retinacular flap can be achieved, which contributes to preserve local blood supply, since vessels to nourish the femoral head are contained within [23, 24]. The soft-tissue flap is derived subperiosteally from the retinaculum and external rotators, allowing an entire detachment of the proximal femur epiphysis. After raising the flap, an anatomical reduction under visual control of the retinaculum will be possible, minimizing the damage to the vessels [24]. This technique was initially reported to treat slipped capital femoral epiphysis, nonetheless, and taking into account the ability to access the entire femoral head and acetabulum, without damaging the femoral head vascularization, we believe that there is room to apply this surgical option to treat other conditions such as the one herein presented [23, 24].

It is well known the poor outcome for intra-capsular proximal femur fractures with conservative treatment, and for this particular case, the poor bone quality and predictable less potential to achieve consolidation represented additional problems [25–27]. The first attempt to treat this fracture was based in an anatomic reduction and fixation. We also performed a relative neck lengthening with a greater trochanter distalization in order to prevent hip impingement and improve hip abductors biomechanics (given the impossibility to restore original neck length). However, the osteosynthesis attempt eventually failed as we could observe during the follow-up. The main reasons for this failure were

probably related not only with the chronicity of the injury, poor bone quality, and challenging anatomy, but also with insufficient reduction, mainly due to lack of bone support in the posterior and inferior aspect of the femoral neck, which also favored suboptimal screw placement. Additionally, and due to poor bone quality, a protective immobilization should also have been performed. All these features drove us toward a new surgical attempt using the same surgical technique, in which we tried to correct the correctable factors of failure, using tricortical iliac autograft to ensure the enough mechanical support and bone stock, and also promoting a proper screws positioning. In the end and to protect the osteosynthesis, a spica cast was added.

Fortunately, the second surgical procedure could provide a successful osteosynthesis, which promoted fracture consolidation and femoral neck anatomy restoration without jeopardizing the femoral vascularization. This is the main advantage of using a surgical hip dislocation with retinacular flap. To our knowledge, this is the first time that this technique is applied to treat a pediatric femoral neck fracture. It is our strong belief that this technique has the potential to become the mainstream option to treat such difficult cases as the one presented.

CONCLUSION

Literature concerning treatment of femoral neck fractures in children with osteogenesis imperfecta is sparse. The authors report on a case of a pediatric femoral neck fracture in a child with osteogenesis imperfecta, performing an open reduction and internal fixation using a hip surgical dislocation with retinacular flap. Despite considering this technique as demanding, this particular case shows how this option could be successfully applied to difficult and unusual cases as the one herein presented.

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Author Contributions

André Spranger – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

André Chambel – Acquisition of data, Analysis of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Joaquim Soares do Brito – Conception of the work, Design of the work, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Paulo Almeida – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Graça Lopes – Conception of the work, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.