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Surgical Management of Exceptionally Severe Cubitus Varus Deformity in Osteogenesis Imperfecta: A Case Report

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Learning Point of the Article:

A staged surgical approach prioritizing stability is effective for managing severe cubitus varus in patients with significant bone fragility.

Abstract

Introduction: Osteogenesis imperfecta (OI) is a rare genetic disorder characterized by significant bone fragility and progressive skeletal deformities. While cubitus varus (CV) is a common complication after pediatric supracondylar fractures, deformities exceeding 40° in patients with OI are exceptionally rare. This report describes a severe case managed through a staged surgical strategy to overcome challenges associated with bone fragility.

Case Report: A 4-year-old male with OI presented with a 45° CV deformity of the right elbow following recurrent fractures. Due to extreme bone fragility and the complexity of the 3D deformity, a staged surgical plan was implemented. The initial surgery prioritized osteotomy site stability and coronal correction using a stepped cut osteotomy. Once bone union was achieved, a second corrective wedge osteotomy was performed a year later to refine the alignment. At the final follow-up at age 7, the patient maintained a 15° carrying angle and a good range of motion. The improved alignment successfully prevented further fractures.

Conclusion: For complex pediatric elbow deformities with underlying bone fragility, a staged approach prioritizing stability over immediate anatomical perfection is a realistic strategy. Establishing a shared understanding with the family regarding the potential necessity of multiple procedures is essential for clinical success and safety.

Keywords: Osteogenesis imperfecta, cubitus varus, staged surgery, osteotomy, bone fragility.

Introduction

Osteogenesis imperfecta (OI) is a rare clinical disorder, occurring at a rate of between 1/10,000 and 1/25,000 worldwide [1]. Previously, OI was thought to be caused solely by dominant mutations in the genes encoding Type I collagen (COL1A1 and COL1A2); however, with the discovery of other rare disease-causing genes, OI is now considered a "type I collagen-associated" disorder. The skeletal phenotype of OI patients is characterized by reduced bone density, increased bone fragility, recurrent fractures, and progressive skeletal deformities; however, because the pathogenic mechanisms vary among OI subtypes, clinical features are also highly diverse [2]. Furthermore, in cases with mild symptoms, such as sillence Type I, it is not uncommon for diagnosis to be delayed and confirmed only after multiple fractures have occurred.

Cubitus varus (CV) is the most frequent complication of treatment for supracondylar humeral fractures in children. It is a 3D deformity consisting of varus angulation in the coronal plane,

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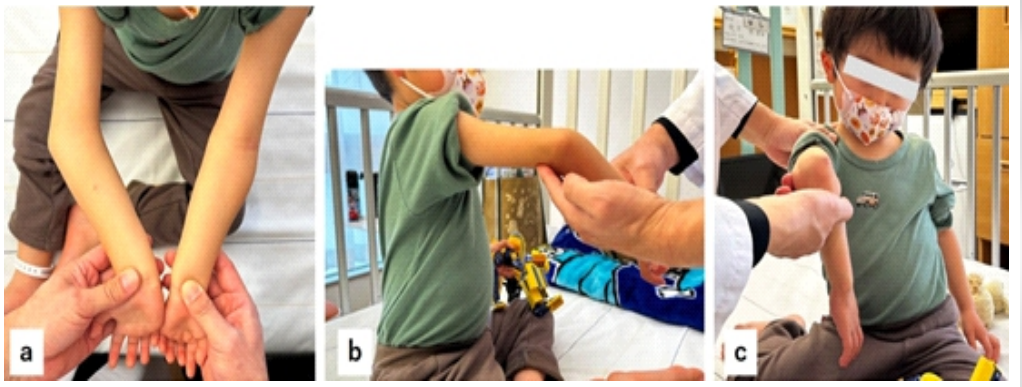

Figure 1: The patient's appearance at the initial visit. (a) The anterior view of the right elbow shows severe varus deformity. (b) The right elbow extension was approximately 20°. (c) The right humerus showed marked internal rotation.

internal rotation in the axial plane, and extension in the sagittal plane. Although CV has been conventionally described as a cosmetic deformity with little functional significance, there is growing awareness of long-term complications, including chronic pain, ulnar nerve palsy [3, 4], tardy posterolateral rotatory instability [5, 6], and an increased risk of lateral condyle and other secondary fractures [7, 8].

Although CV can indeed occur following a supracondylar humeral fracture in OI, there have been no reports of severe varus deformity exceeding 40°. Here, we report a case of very severe CV and discuss the challenges of treating fractures around the elbow in OI.

Case Report

The patient is a male who initially suffered a supracondylar fracture of the right humerus at 2 years and 6 months of age. He underwent cross-pinning at a hospital on the day of the injury, but the Kirschner wire subsequently came out shortly thereafter, resulting in mild varus deformity of the elbow. After that, the condition was monitored without surgery, but 6 months after the initial fracture, the patient fell and suffered a recurrent supracondylar fracture of the right elbow, which was treated conservatively with a cast at hospital B. Subsequently, at 12, 14, and 16 months after the initial fracture, the patient repeatedly sustained fractures of the lateral condyle of the right humerus due to falls, and pinning and cast treatments were repeatedly performed. Due to the recurrence of frequent fractures, the patient underwent evaluation at the pediatric department of A hospital, where a diagnosis of OI was made (COL1A2 c693+1G>A heterozygous splice donor variant). The patient was then referred to our facility for management of severe CV and OI.

When he first visited our facility at the age of four, his height was 90 cm, and the body weight was 11.6 Kg. He had a severe CV of approximately 45° in the right elbow, and approximately 20° of extension deformity and some internal rotation deformity were observed (Fig. 1). However, the deformity was so severe that accurate three-dimensional assessment was difficult. The patient had blue sclera and no apparent family history of OI. As shown in Fig. 2, X-rays and computed tomography scans revealed severe varus deformity of the distal humerus and a

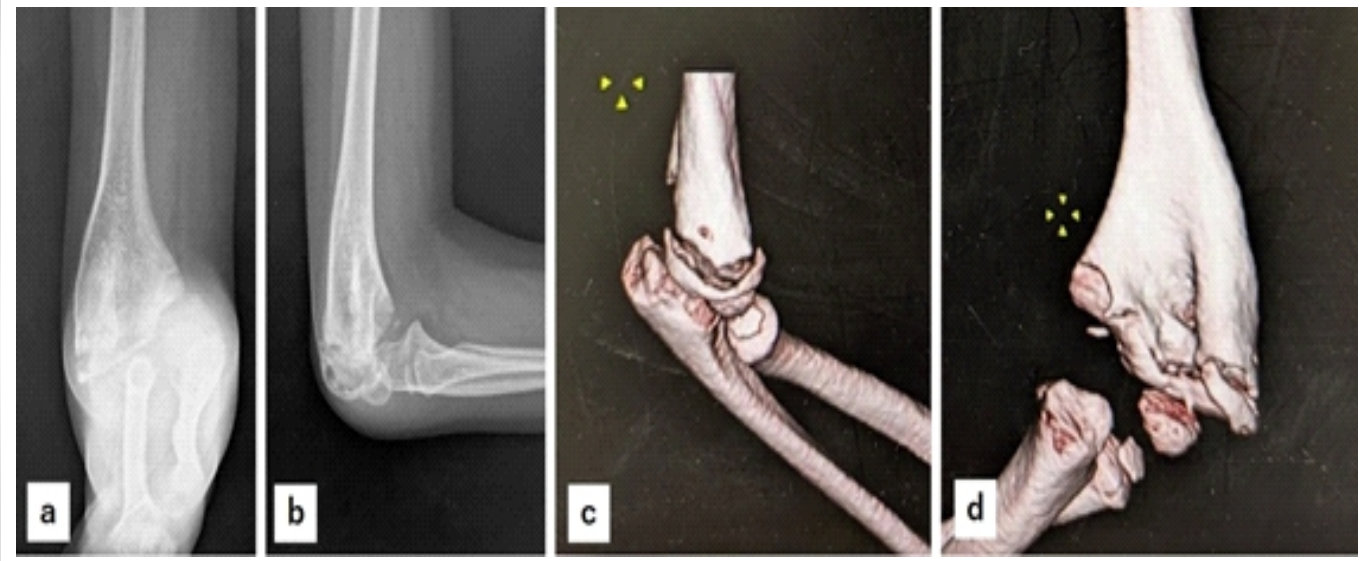

Figure 2: X-ray and 3D computed tomography (CT) images at the time of initial visit to our hospital. X-ray (a): Anteroposterior view, (b): Lateral view, and 3D CT images (c): Lateral view, (d): Posterior view show marked varus deformity of the humerus and non-union of the lateral condyle fracture.

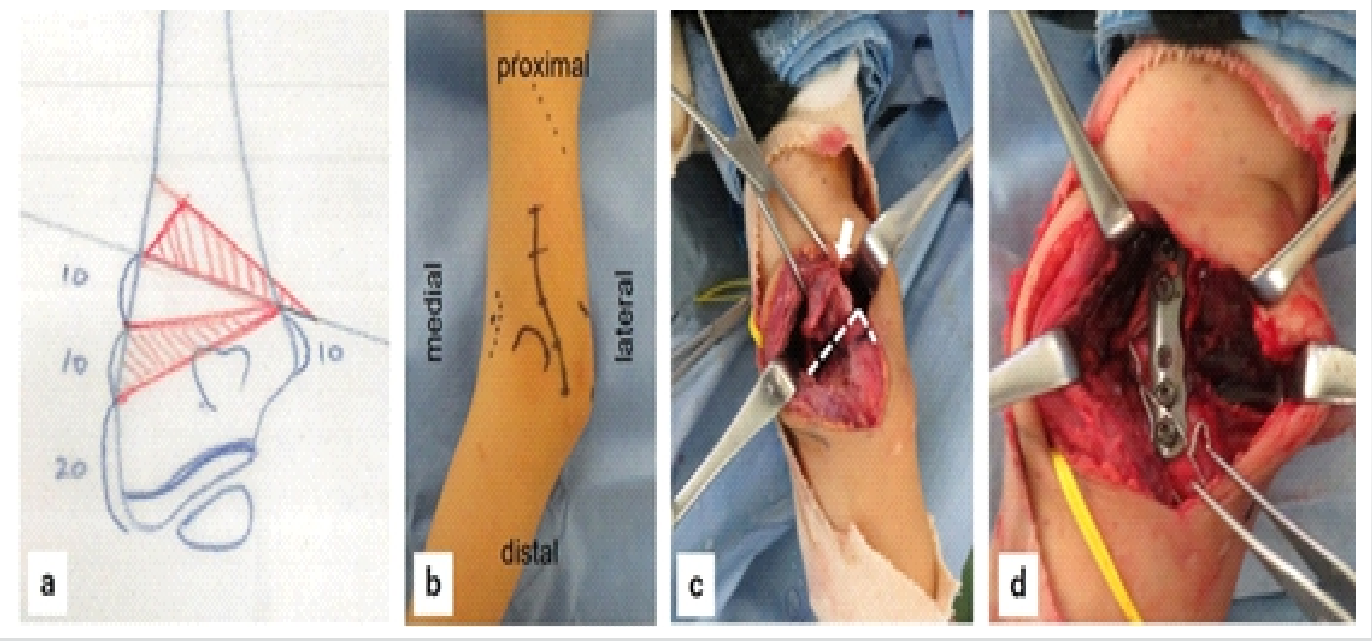

Figure 3: Surgical plan and surgical findings. (a) Osteotomies were performed in a step-like fashion in the sagittal plane so that the red-lined areas would overlap, with a planned valgus angle of approximately 45°. (b) A curved skin incision was made extending from the posterior to the lateral side of the right elbow. (c) The distal humerus was osteotomized in a stepped manner on the sagittal plane and in a triangular manner on the coronal plane, so that the osteotomy sites would make contact over a wide area. The white dotted lines indicate the edges of the osteotomy sites on the distal fragment. (d) The osteotomy site was stabilized using a plate and screws, as well as a Kirschner wire and soft wire.

fracture line at the lateral condyle. The dominant hand of the patient is right and suffered significant limitations in activities of daily living. Furthermore, due to the poor alignment of the right elbow, the patient had fallen into a vicious cycle where fractures recurred immediately upon falling and bracing with the hand; therefore, a plan was established to correct the deformity via osteotomy. Rather than forcibly correcting rotational or extension deformities, the plan prioritized achieving stability by increasing the contact surface at the osteotomy site. Therefore, a stepped cut osteotomy in the sagittal plane and 45° valgus correction in the coronal plane were planned (Fig. 3a), followed by plate fixation. It was decided that if, after union was achieved, there were insufficient corrections such as rotational alignment, the case would be re-evaluated in detail before performing additional precise osteotomies and internal fixation; this was fully explained to the patient's parents before the surgery.

The first surgery was performed under general anesthesia in the prone position at 4 years and 10 months old. An incision was made as shown in Fig. 3b, and the distal humerus was exposed from the posterior approach. After performing the osteotomy as planned (Fig. 3c), the bone was fixed with a plate from the posterolateral side; and due to concerns about stability, additional fixation was provided using Kirschner wires and soft wires (Fig. 3d and 4). However, due to poor bone quality, the Kirschner wires gradually loosened over time, causing slight displacement at the osteotomy site; nevertheless, bone union was achieved by extending the duration of external fixation. Six months postoperatively, mild varus deformity persisted, and there were concerns about re-fracture due to falls, so we decided to perform a second corrective osteotomy.

The second surgery was also performed in the prone position using the same incision at 5 years and 8 months old. Since the bone thickness at the planned osteotomy site had increased following the initial surgery, it was determined that a standard wedge osteotomy would provide sufficient contact between the osteotomy surfaces. A wedge-shaped osteotomy was performed

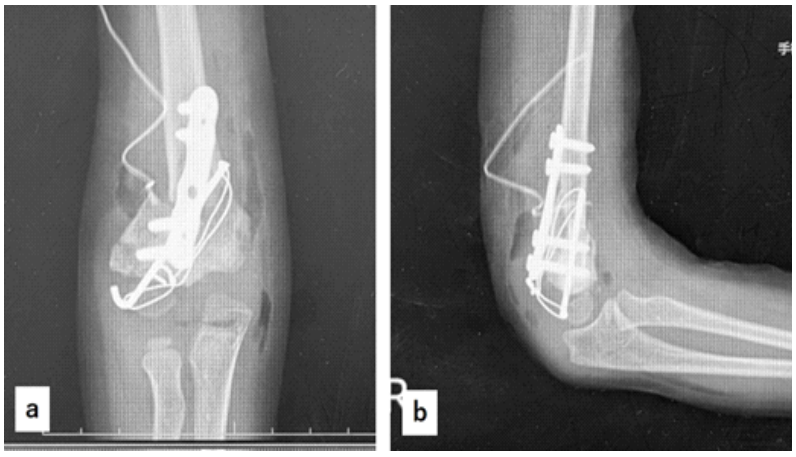

Figure 4: X-ray findings after the first surgery. (a): Anteroposterior view, (b): Lateral view.

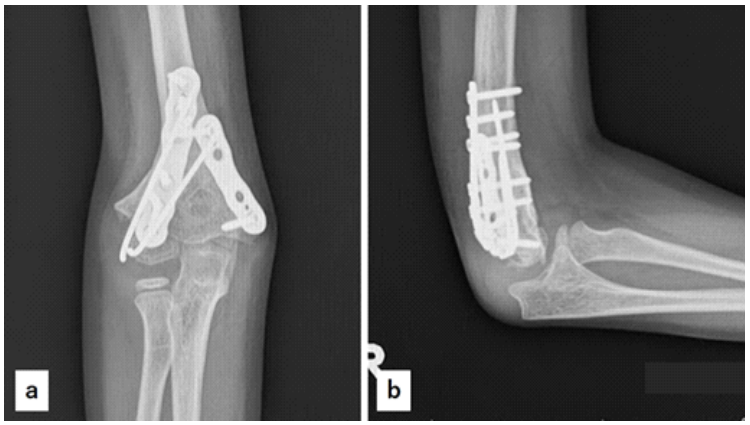

Figure 5: X-ray findings after the second surgery. The alignment of the humerus has been corrected, and the fracture site at the lateral condyle has healed well. (a): Anteroposterior view, (b): Lateral view.

to achieve 20° of valgus and 10° of flexion, and the bone was firmly fixed from the posterior aspect using two plates (Fig. 5). Sufficient bone union was achieved 6 months after the second surgery, so the internal fixation hardware was removed. At the final follow-up at age seven, the carrying angle is 15°, and the good range of motion for elbow flexion and extension is maintained (Figs. 6 and 7), thanks to the improved alignment; no re-fractures have occurred following falls.

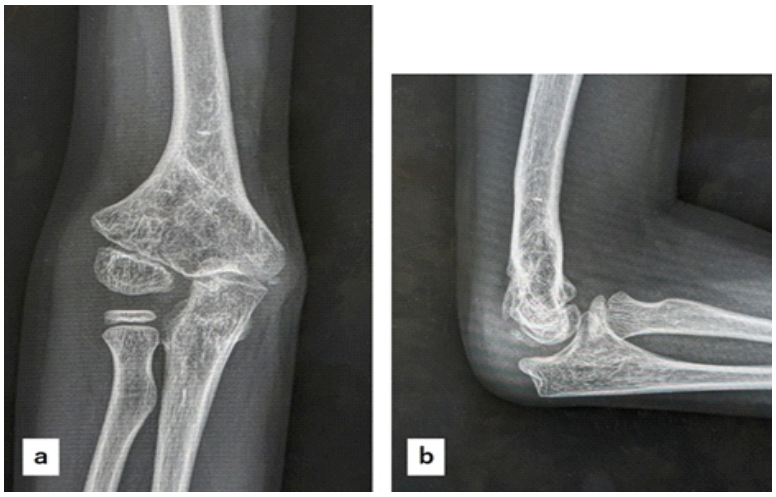

Figure 6: X-ray findings at final follows up. (a): Anteroposterior view, (b): Lateral view.

Discussion

Cases like the present one, involving severe CV exceeding 40° in patients with OI, are extremely rare even in existing reports. CV involves a complex combination of varus, internal rotation, and extension deformities. When the deformity is extremely severe, it becomes difficult to accurately assess the three-dimensional deformity even with plain radiographs or CT scans. Various osteotomy techniques for CV have been reported, and none is generally considered superior to the others [9, 10]. In the current case, due to the patient's small stature and small bones, combined with the severe deformity, we determined that attempting to achieve perfect 3D anatomical correction in a single surgery would risk reducing the contact area of the osteotomy surfaces, thereby compromising fixation stability. Therefore, given the bone fragility associated with OI, we prioritized "achieving contact and stability at the osteotomy sites" over "perfect correction of rotation and extension" during the initial surgery, limiting the procedure to simple valgus correction in the coronal plane. Furthermore, Takagi et al. have reported that good outcomes can be achieved with valgus correction alone in CV cases under 10 years of age [11]; our judgment is therefore considered reasonable from this perspective as well. Thus, for complex elbow deformities involving underlying bone fragility, a strategy that allows for staged correction – rather than aiming for "perfection in a single surgery" – is more realistic.

The bones of OI patients are extremely fragile, presenting challenges that differ from those of typical fracture treatments and deformity correction surgery. During the initial surgery, we reinforced the fixation with K-wires in addition to plates and screws; however, due to poor bone quality, wire laxity and displacement occurred. This strongly suggests that in OI patients, even when fixation is believed to be

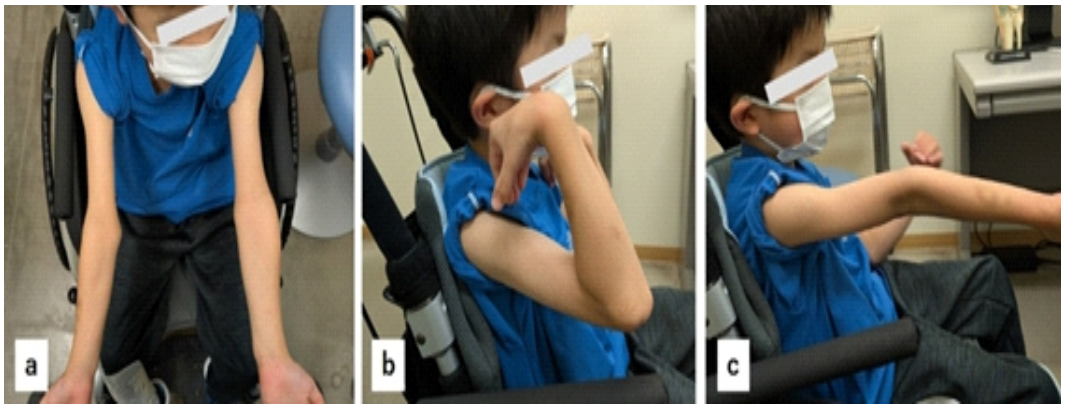

Figure 7: The patient's appearance at final follows up. The carrying angle of the right elbow has improved to normal (a), and the range of motion for elbow flexion and extension is full (b and c).

"secure," there is always a risk of post-operative loss of correction. For the second surgery, partly because the bone had thickened following the initial procedure, we opted for robust fixation using two plates. In cases involving OI, careful management is required, including preparing more backup fixation methods than usual and setting a longer period for post-operative external fixation.

In the treatment of severe deformities in patients with OI, the success of treatment depends not only on medical expertise but also on prior communication with the family. Given the fragility of the bones and the complexity of the deformity, the risk of post-operative displacement or insufficient correction is higher than usual. In the current case, it was crucial that we explained to the parents before the initial surgery that "there is a significant possibility that additional surgery will be necessary" and obtained their understanding. Ultimately, after two surgeries, we achieved good alignment and range of motion, breaking the vicious cycle of re-fractures. Precisely because this was a case with a high degree of uncertainty, incorporating a "stepwise approach" into the treatment plan from the pre-operative stage and sharing it with the patient is key to enhancing both the final treatment satisfaction and safety.

Conclusion

Fractures around the elbow in OI require special attention, as they often result in severe deformities during the healing process, unlike typical fractures. We reported a case of extremely severe CV in a patient with OI that was successfully managed through a two-stage surgical approach. In cases with profound bone fragility and complex multi-planar deformities, prioritizing osteotomy site stability over immediate anatomical perfection is a valid and realistic strategy. To minimize the risk of post-operative loss of correction, robust internal fixation and an extended period of external support are essential. Furthermore, establishing a shared understanding with the family regarding the potential necessity of staged procedures is crucial for ensuring treatment safety and satisfaction in such challenging clinical scenarios.

Clinical Message

- Fractures associated with osteogenesis imperfecta can result in severe malunion that would not normally occur.
- For extremely severe deformities, it may be beneficial to perform corrective surgery in multiple stages.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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