

# Humeral lengthening and deformity correction in Ollier's disease: distraction osteogenesis with a multiaxial correction frame

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**A case of Ollier's disease with deformity and shortening of the humerus is presented. Lengthening of 9 cm and deformity correction of 50 degrees were accomplished with excellent functional and cosmetic results. Unique features of this case were the use of a multiaxial correction monolateral frame and the formation of normal bone within the region of diseased Ollier's bone. *J Pediatr Orthop B* 17:152–157 © 2008 Wolters Kluwer Health | Lippincott Williams & Wilkins.**

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## Introduction

Ollier's disease is a rare, nonfamilial disorder characterized by the presence of multiple enchondromatosis [1]. It is a form of dyschondroplasia that represents a defect of endochondral calcification, proliferation of ectopic islands of chondroid tissue, and incapacity of the epiphyseal plate to mature [2]. Mutant parathyroid hormone related to a protein receptor PTHrP was found to cause endochromas in transgenic mice [3], but the exact pathogenesis in humans remains unclear.

Ollier's disease carries a 30% risk of malignancy [4]. It is regarded as a low-grade chondrosarcoma and close follow-up is mandatory. The rate of conversion to chondrosarcoma is higher in Maffucci's syndrome, a variant of Ollier's associated with soft tissue hemangiomas [5,6]. Patients with Ollier's disease usually present with deformity and length discrepancy of the affected limb. This leads to gait abnormalities in the lower extremities and difficulties with reach and compromised function and cosmesis when upper extremities are affected.

Curettage and internal fixation techniques have been used to treat lesions of Ollier's disease. These techniques, however, can only offer stabilization of the affected area without necessarily addressing the length discrepancy or deformity. In this case report, we present our experience in treating this condition utilizing the technique of distraction osteogenesis. In combination with a multiaxial correction (MAC) (EBI, Parsippany, New Jersey, USA) monolateral frame, distraction osteogenesis led to regeneration of normal bone within the region of Ollier's disease. This biological phenomenon is not commonly known. The treatment led to correction of

length discrepancy and deformity, which was achieved with satisfactory functional and cosmetic results.

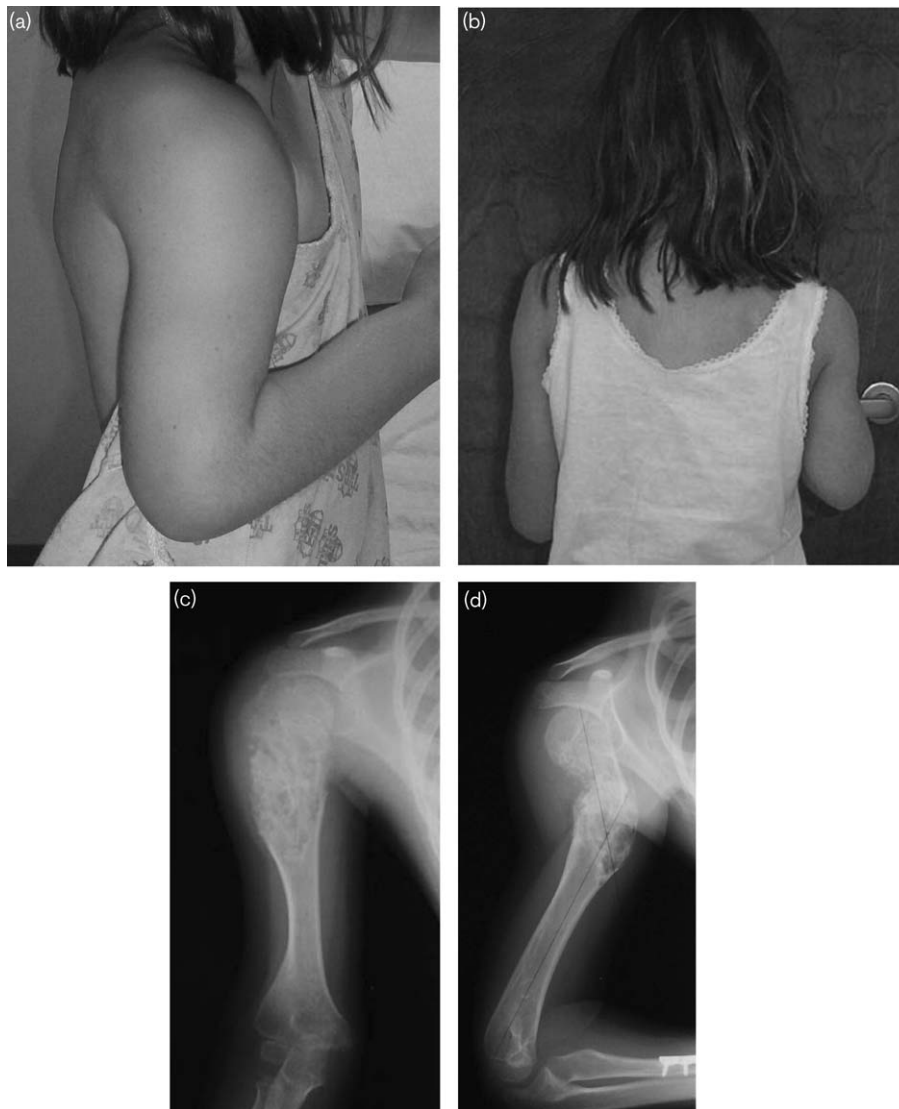
## Case report

A 7-year-old girl child with a short right humerus associated with deformity was referred by the Hand and Tumor service at our institution. She was free of pain, but was unhappy with the appearance resulting from the deformity and shortening and complained of inability to reach with the right arm owing to short arm span. She had a history of Ollier's disease, but there was no family history of the disease. Ollier's disease involved her humerus, radius, index finger, and scapula all on the right side. The child did have lower extremity involvement. The child had undergone surgery on her radius for curettage and osteotomy and plating at the age of 6 years with satisfactory results. She had no significant medical history, and a review of systems was otherwise negative.

## Physical exam

At presentation, the child was noted to be 4 feet tall and weighed 56 lb. The discrepancy in length between the humeri was 6 to 7 cm (Fig. 1a and b). Examination of her right shoulder revealed forward flexion to 180 degrees, abduction to 160 degrees, external rotation to 90 degrees, and internal rotation to the thoracolumbar spine. Elbow motion was full extension to 135 degrees of flexion, 90 degrees of pronation, and 90 degrees of supination. Wrist motion was 90 degrees of palmar flexion, 80 degrees of dorsiflexion, 25 degrees of ulnar deviation, and 40 degrees of radial deviation. Range of movement of the elbow and wrist was equal to the contralateral limb. Hand and finger motion were noted to be normal. Normal motor and sensory function of the axillary, median, radial, and

Fig. 1



(a) Preoperative side view showing the apex anterior deformity. (b) Back view showing the amount of shortening of the right arm span compared with contralateral side. (c, d) Anteroposterior (AP) and Lateral views of right humerus showing the extent of Ollier's disease and the 50 degree apex anterior deformity.

ulnar nerves was observed. Radial and ulnar pulses were present.

### Radiographs

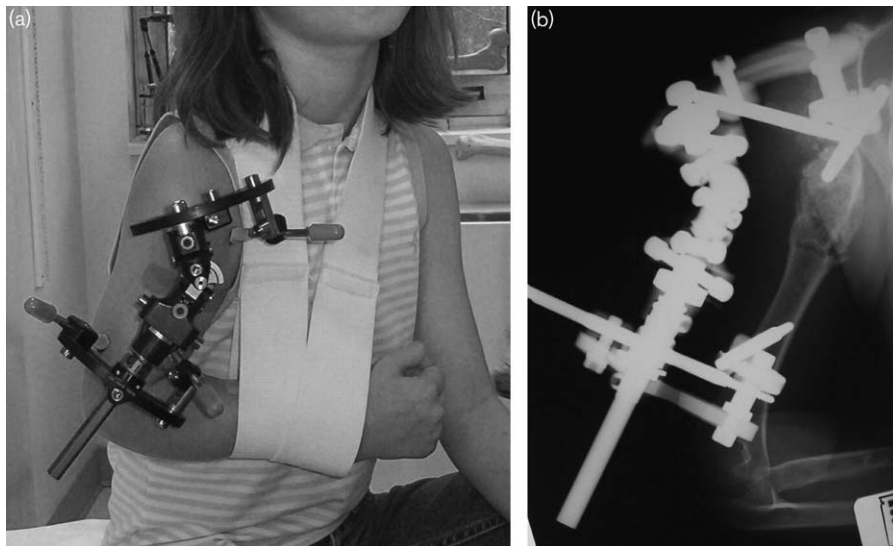
The child had a 50 degree apex anterior deformity at the proximal one-third of the humerus. The length of her normal humerus was 23 cm and the length of the right humerus was 17 cm. Ollier's disease at the proximal humerus with possible involvement of the growth plate was evident. The apex of the deformity was located within the region of diseased bone (Fig. 1c and d) at the base on an upper limb multiplier [7] of 1.37 and the

length of the left humerus of her projected left humeral length was calculated to be 31 cm. If the child were to have no additional growth at the proximal growth plate in the right humerus, the projected discrepancy at maturity would be 14 cm.

### Treatment

Gradual lengthening and correction of deformity were performed using distraction osteogenesis. The procedure was performed under regional block and sedation. The MAC frame (EBI, Parsippany, NJ) was applied, using two proximal and two distal half pins. The frame was

Fig. 2



(a) Early postoperative clinical picture showing how the frame mimics the deformity. Note the hinge at the level of the deformity. (b) Lateral radiograph showing fracture in the proximal humerus within the region of Ollier's bone and at the apex of deformity.

mounted with 50 degrees apex anterior parameter dialed onto the frame to mimic the humeral deformity, and the hinge was placed directly over the apex of deformity (Fig. 2a and b). The proximal pins were placed laterally and anterolaterally using 4.5-mm hydroxyapatite-coated pins. The distal pins were placed from anterolateral and posterolateral direction using 6-mm hydroxyapatite-coated pins. Percutaneous osteotomy just distal to Ollier's bone was performed. The rationale of choosing a distal location was to avoid lengthening through Ollier's bone. It was also felt that the osteotomy location proximal to deltoid tuberosity may alter the shoulder biomechanics. Distraction started on the 8th day after surgery at the rate of  $\frac{1}{4}$  turn corresponding to  $\frac{1}{4}$  mm four times daily. Premature consolidation occurred at the osteotomy site, and the rate of distraction was increased to  $\frac{1}{4}$  mm five times a day to overcome the problem. Spontaneous fracture occurred more proximally through Ollier's bone (Fig. 2b). This led to the decision to continue lengthening through Ollier's bone. We observed new bone regeneration within the region of diseased bone.

### Outcome

A total of 9-cm length was achieved with full correction of the apex anterior deformity (Fig. 3). The frame was worn by the child for 165 days, with a healing index of 18.3 days/cm. The range of movement of the shoulder and the elbow was normal, and there was no evidence of neurovascular compromise.

At 2-year follow-up, there was no recurrence of the deformity or Ollier's bone (Fig. 4). The child had equal

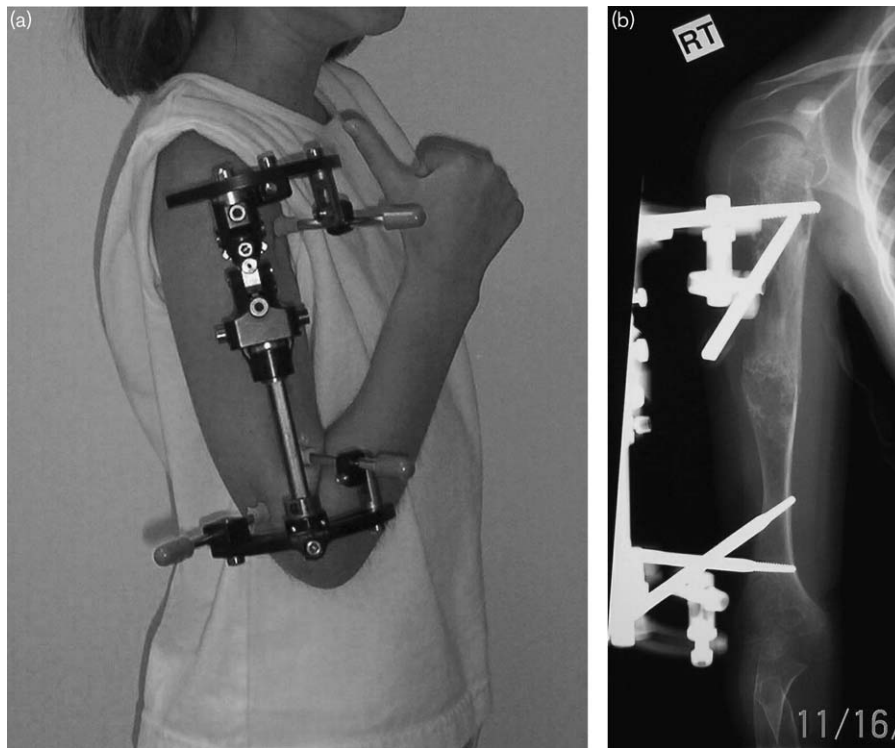
limb lengths, with full range of movement in the shoulder and elbow joint and excellent reach (Fig. 5).

### Discussion

The pattern of growth and progression of deformity in Ollier's disease is difficult to predict. Different approaches have been utilized with regard to the amount of lengthening, fixation method, and osteotomy location [8]. In this child, Ollier's disease resulted in humeral shortening and marked deformity at the center of the diseased bone. The location of the lesion and its proximity to the growth plate in a growing child have compounded the problem.

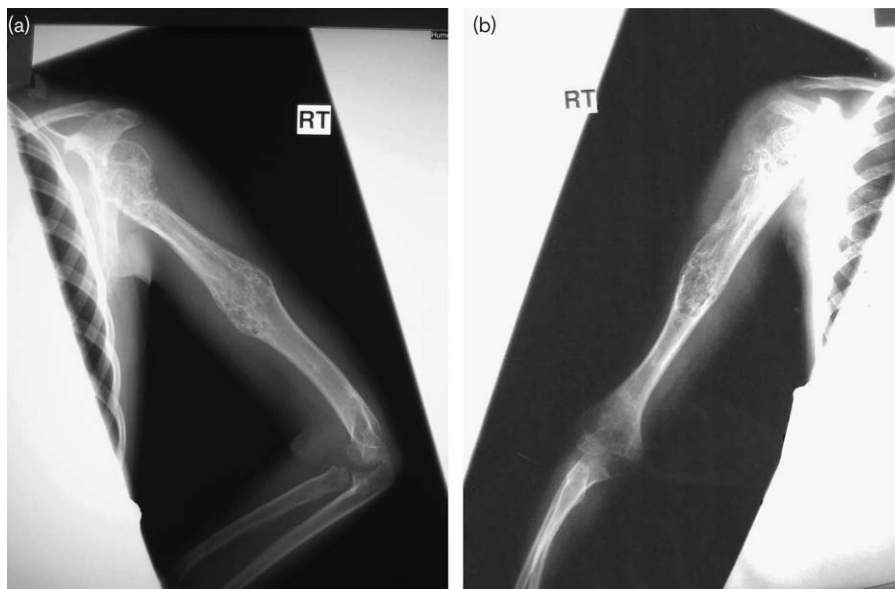
Traditionally, the methods of treatment included curettage, bone grafting, osteotomies, and internal fixation. These methods are not comprehensive in that they do not address the length discrepancy. The use of distraction osteogenesis and the Ilizarov frame in Ollier's disease have been described [9–11]. This technique enables length and deformity correction and stimulates the conversion of enchondromas into lamellar bone [9,10]. Its minimally invasive nature does not disturb the blood supply to the diseased area, and gradual correction of length and deformity is achievable. It is thought that intramembranous ossification is not disrupted in Ollier's disease [1,7,11]. Myers *et al.* [12] studied the use of distraction osteogenesis in skeletal dysplasia and confirmed that Ollier's disease can be treated by using the Ilizarov method with satisfactory results and noticed a tendency for hypertrophic regenerate in patients with Ollier's disease. Premature consolidation is a

**Fig. 3**



(a) Clinical picture at the end of lengthening and correction. (b) AP radiographs showing correction of deformity and 9-cm regenerate. AP, anteroposterior

**Fig. 4**



(a, b) AP and lateral radiographs at 1-year follow-up showing full correction of deformity. Normal appearing bone can be seen in the middle of the Ollier's affected area. AP, anteroposterior.

Fig. 5



(a-c) Clinical pictures at 2-year follow-up showing equal upper arm lengths and normal range movement of the right shoulder and elbow joints.

problem that has been reported in the literature and was experienced in our child [8].

Only a few publications have discussed upper limb lengthening and angular deformity correction by the Ilizarov technique in this condition [9–11]. Jesus-Garcia *et al.* [9,13] used a distraction osteogenesis technique to treat deformity in two humeri. The amount of lengthening was 2 cm in the humerus with no reported complication.

The child tolerance of circular frames in the upper limb is low. Use of circular frames in the upper limb is awkward, as it requires the child to abduct the shoulder to avoid hitting the frame against the trunk. Therefore, we opted to use a monolateral frame that offers multi-axial correction. The type of fixator used offered stability and adjustability to address the length and the deformity correction. The MAC frame is a monolateral frame that has the capacity for multi-planar pin placement. It also can be used to lengthen gradually and to correct angulation and translation in both the coronal and sagittal planes [14] (Figs 2a and 3a).

Complete correction of the deformity can only be achieved if the osteotomy is performed at the apex of the deformity, in this child located within the region of Ollier's bone. Some authors have advocated performing the osteotomy at Ollier's bone location. It is thought that fractures through Ollier's bone heal in a similar fashion to normal bone. Report of conversion of Ollier's bone into a normal bone has also been published using histological studies [10,13].

In this case, we performed the osteotomy distal to Ollier's bone beyond the deltoid tuberosity. The rationale was not only to tighten the deltoid but also to avoid the diseased bone. Distraction was started 8 days after surgery, with a distraction rate of  $\frac{1}{4}$  mm four times daily. Resistance to lengthening was noticed, suggesting premature consolidation, and the rate of distraction was increased to  $\frac{1}{4}$  mm five times daily to overcome the problem. The child reported a sudden give and mild pain in the area of the osteotomy. Radiological studies had shown a fracture through Ollier's bone and persistent premature consolidation at the original osteotomy site.

Lengthening and correction through the fracture (new osteotomy) was continued. Follow-up has shown satisfactory regenerate formation. We observed that the method of distraction osteogenesis led to the transformation of enchondroma into normal bone.

The outcome of this case suggests that deformity and shortening from Ollier's disease can successfully be treated with distraction osteogenesis. This method of treatment helps the conversion of Ollier's bone into a normal bone. This phenomenon was used to achieve considerable lengthening and full deformity correction. It is also important to highlight the location of the diseased bone and the challenges it represent in terms of feasibility of full deformity correction and lengthening without disrupting the shoulder mechanics. From our experience, we feel that osteotomies can be safely done at Ollier's disease bone and can be used to stimulate Ollier's disease conversion into normal bone. We also found the MAC frame to be well suited for lengthening and deformity correction in the upper arm.

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