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Original article

Treatment of lower limb deformities and limb-length discrepancies with the external fixator in Ollier's disease

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Abstract
Background. In this study, we addressed two questions on the treatment for Ollier's disease: (1) how much callus formation occurs when an osteotomy is performed intralesionally and (2) how is the stability of the wires and half-pins that are inserted intralesionally.
Methods. Four children with Ollier's disease underwent treatment of 12 lower limb segments using distraction osteogenesis until completion of their growth. All osteotomies were performed at the centers of the deformities, resulting in a total of seven osteotomies performed intralesionally.
Results. Full correction of the deformity and full restoration of length were achieved in all cases, but a residual limb-length discrepancy of <10 mm remained. The mean external fixation index in the intralesional distraction osteogenesis group was 39.7 days/cm versus 30.8 days/cm in the extralesional distraction osteogenesis group. Conversion from abnormal cartilage to normal regenerate bone was seen in only one segment. Although approximately two-thirds of the wires and half-pins were inserted intralesionally, in all but one case (in which an iatrogenic fracture occurred) the wires and half-pins were well stabilized throughout the external fixation period.
Conclusions. Although deformity and limb-length discrepancies due to Ollier's disease were successfully resolved by distraction osteogenesis, enchondroma may arise in distracted calluses when osteotomized intralesionally. However, the stability of the external fixator was sufficient to lengthen limbs and correct deformities even when wires and half-pins were inserted intralesionally.

Introduction

Patients with Ollier's disease have deformities and distortions in the longitudinal growth of the bones, mainly in the legs.[1–3] Bowing and growth distortions occur, resulting in deformities such as genu valgus and leg-length discrepancies as well as pathological fractures. Osteotomies are performed as many times as necessary to correct deformities. However, bone stabilization is difficult to attain using internal devices, as the bone may be fragile, and the presence of pathological bone and enchondromas has been shown to preclude internal fixation.[4,5] Multiple areas of abnormal bone and difficulty obtaining normal bone for grafting also are important problems to address.

In this study, we addressed two questions on the intralesional treatment for Ollier's disease: (1) how much callus formation occurs when an osteotomy is performed and (2) how is the stability of the wires and half-pins that are inserted.

Patients and methods

From November 1990 to November 2004, twelve segments in four patients with Ollier's disease were treated at our institution (Table 1). The two boys and two girls were 4–7 years old (mean age 5.8 years) at the time of the initial surgery. All the second or multiple procedures were performed to correct a recurrence of deformity during their growth. The ring external fixators was used in nine segments (four femurs, five tibias), and the monolateral external fixators was used in three segments (two femurs, one tibia). All external fixators were installed using standard techniques. A total of 12 procedures were performed for a mean of three procedures per person: 6 in the femur and 6 in the tibia. All deformities were evaluated by the center of rotation and angulation (CORA) method,[6] and all osteotomies were performed at the CORA level. As bone weakness by chondromas were marked, more wires and half pins were used to secure that bone than would be used for the stabilization of normal bone. The achieved lengthening rate was 0.5–1.0 mm per day. The follow-up period

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Table 1. Patient demographics

Segment	Age (years)	Site	EF	Lengthening (mm)	Angulation (°)	Varus/valgus	EFT (days)	EFI (days/cm)	Intra/extra
Patient 1									
1	7	Femur	Orthofix	20	0	–	114	57.0	Extra
2	7	Tibia	Orthofix	30	0	–	114	38.0	Extra
3	13	Tibia	Ilizarov	44	20	Varus	161	36.3	Extra
Patient 2									
4	6	Tibia	Ilizarov	41	15	Varus	193	46.8	Intra
5	10	Tibia	Ilizarov	51	20	Valgus	352	69.3	Intra
6	15	Femur	Hidergerg	77	28	Varus	178	23.1	Intra
7	15	Tibia	Ilizarov	10	31	Valgus	178	178.0	Intra
Patient 3									
8	6	Femur	Ilizarov	60	23	Valgus	160	26.7	Extra
9	8	Femur	Ilizarov	35	24	Valgus	117	33.4	Intra
10	12	Femur	Ilizarov	25	18	Valgus	83	33.2	Intra
Patient 4									
11	4	Tibia	Ilizarov	46	0	–	102	22.2	Extra
12	8	Femur	TSF	52	45	Valgus	135	26.0	Intra

EF, external fixator; EFT, external fixation time; EFI, external fixation index; Intra, intralesional osteotomy; Extra, extralesional osteotomy; TSF, Taylor Spatial Frame

ranged from 64 to 168 months (mean 118.5 months). The preoperative leg-length discrepancy ranged from 2 to 9 cm (mean 4.4 cm).

We used the external fixation index (EFI) to evaluate our results. The EFI is defined as the entire duration of external fixation divided by the length gained. We considered consolidation completed when anteroposterior and lateral radiographs found that three of four cortices of regenerated bone in the distraction gap were bridged.[7]

Statistical significance was determined using the unpaired Student's *t*-test. $P < 0.05$ was considered to be statistically significant.

The patients and their families were informed that data from the case would be submitted for publication and gave their consent.

Results

The deformity correction followed by lengthening was performed in nine segments and the limb-lengthening in three segments. The mean amount of length gained was 40.9 mm (range 10–77 mm), and the mean angular correction was 24.9° (range 15°–45°). The mean external fixation period was 157.3 days (range 83–352 days), and the mean EFI was 49.2 days/cm (range 22.2–178.0 days/cm). The limb-length discrepancy at the latest follow-up was <10 mm in all cases (Fig. 1). All osteotomies were performed by the CORA method, so seven osteotomies were performed intralesionally followed by distraction osteogenesis. In general, the lengthening <3 cm was significantly large compared with that >3 cm in the EFI.[9] The mean EFI excluded the lengthening <3 cm in the intralesional distraction osteogenesis group was 39.7 ± 18.9 days/cm versus 30.8 ± 7.6 days/cm in the extralesional distraction osteogenesis group. There was no significant difference between the two groups ($P = 0.41$). Radiographs from the distraction osteogenesis group, including enchondromal masses, showed only one conversion of the enchondromal mass into normal new bone regenerate (Table 2). However, this conversion was not confirmed by histological analysis. Over the 12 total external fixations, 48 wires and 38 half-pins

Fig. 1. A 6-year-old boy with Ollier's disease had affected right upper and lower limbs with multiple enchondromatoses. **A** Initial radiogram at age 6 shows a limb-length discrepancy of 41 mm with a valgus deformity of 15° in the right ankle and chondromas in the proximal and distal femur and tibia. **B** Radiogram shows the first correction with the Ilizarov apparatus in the tibia. Illustration shows the enchondromas, the level of the wire insertions, and the level of the osteotomy, which was performed intralesionally. **C** Radiogram shows correction of the deformity and limb-lengthening. Conversion of cartilage to normal bone by distraction osteogenesis was observed. **D** Radiogram showing the second correction at age 10. A Valgus deformity of 20° and a limb-length discrepancy of 51 mm recurred. Acute correction using a focal dome osteotomy was performed intraoperatively, and gradual limb-lengthening was performed postoperatively. **E** Radiogram shows correction of the deformity and limb lengthening. Conversion of cartilage to normal bone was not observed. **F** Radiogram shows the third correction at age 15. A valgus deformity of 31° in the tibia, a varus deformity of 28° in the femur, and a limb-length discrepancy of 87 mm were present. Acute correction using a focal dome osteotomy was performed in the tibia. **G** Radiogram shows correction of the deformity and limb lengthening. Conversion of cartilage to normal bone by distraction osteogenesis was not observed. After skeletal maturation the limb-length discrepancy was 5 mm

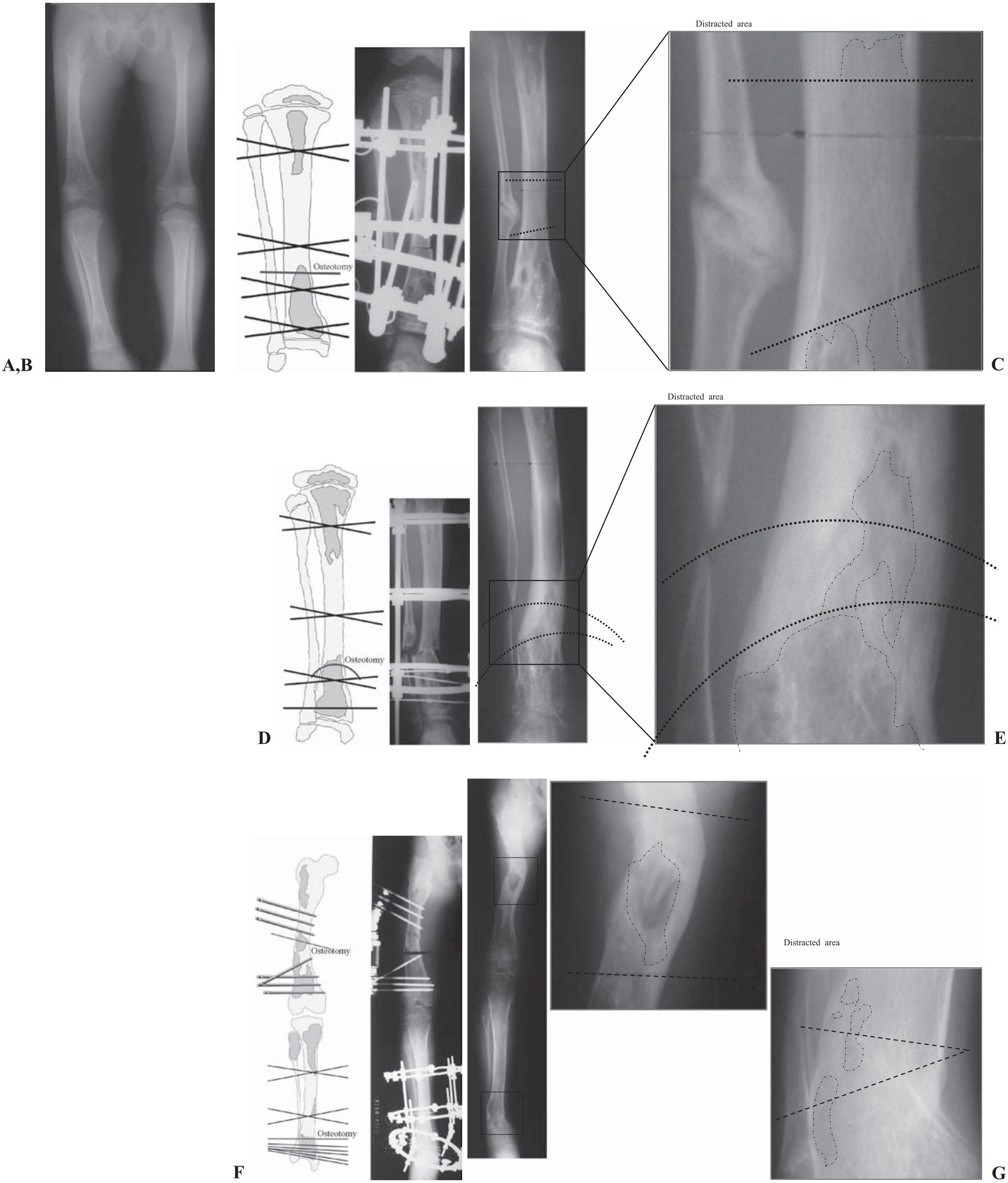
A,B
Osteotomy
Distracted area
C
Osteotomy
Distracted area
D
E
Osteotomy
Osteotomy
Distracted area
F
G

Table 2. Intralesional versus extralesional distraction osteogenesis

Distraction osteogenesis	Segment	EFI (days/cm)	Conversion
Intralesional	7	39.7 ± 18.9	1/7
Extralesional	5	30.8 ± 7.6	

$P = 0.41$

Table 3. Number of wires and half-pins inserted intralesionally

Parameter	Intralesional	Extralesional
Wires	30	18
Half-pins	26	12

were used. Although 30 wires (62.5 %) and 26 half-pins (68.4 %) were inserted through the enchondromal masses, all wires and half-pins were well stabilized throughout the external fixation period except in one patient, in whom an iatrogenic fracture occurred (Table 3).

Discussion

Curettage/bone grafting and deformity correction are two different treatments with different intended goals in the treatment of Ollier's disease. Complete eradication of enchondromal masses is realistically impossible as the lesions are extensive.[1] Limbs with good mechanical alignment have the advantage of joint health, which influences truncal and contralateral limb alignment, and have sufficient support to prevent pathological fracture.[8] In our study, although approximately two-thirds of the wires and half-pins were inserted intralesionally, the stability provided was sufficient for deformity correction and limb-lengthening. Based on these results, we conclude that the most important treatment goals in Ollier's disease are to achieve good mechanical realignment and equivalent limb-length for normal walking ability and to relieve pain from pathological fracture. Distraction osteogenesis is the most effective and reliable surgical procedure for deformity correction and limb lengthening. However, with an intralesional osteotomy, healing, which is most often cartilaginous and at times with bone, occurs with standard distraction osteogenesis techniques.

Conventional surgical treatment — curettage, bone grafting, osteotomies — is not feasible in many patients with Ollier's disease because the affected bone is weakened and the amount of elongation necessary to correct the discrepancy is usually extensive. Application of the Ilizarov method provides enough mechanical strength to perform an elongation axial correction. Unilateral external fixators are easier and more comfortable for patients than ring external fixators, but their use is limited for simple limb lengthening in patients with Ollier's disease.

If the patient is young and the deformity is gross, repeated deformity correction combined with lengthening is appropriate.[9] We performed 12 external fixation surgeries in four patients. Although the proper timing for surgery depends on many factors, the procedure is usually indicated when the weight-bearing alignment of the extremity is significantly altered. Shapiro[1] reported that an angular deformity of ≥25° that is not balanced by a reverse deformity is an indication for surgery. If possible, procedures should be delayed to limit the amount of surgery required. Although it might seem that overcorrection of the deformities could decrease the likelihood for repeated surgeries, it is difficult to know how much overcorrection is desirable. We emphasize the need to plan carefully so the fewest surgical procedures possible are required.

Conversion of cartilage to normal bone that had been observed on radiograms was confirmed by Garcia[4] in a biopsy of the elongated area, where typical lamellae and bone marrow were obtained. However, we are not entirely in agreement with this opinion. Shapiro[1] reported that fully normal trabeculation did not occur after skeletal maturation in any site where there had been a lesion. Kolodziej[5] reported that the conversion of cartilage to normal bone was observed in four of five segments. In our study, this transformation occurred radiographically in only one of seven segments, but we did not confirm it histologically because of the ethical difficulty of obtaining a sample. There is no consensus on whether intralesional distraction osteogenesis in patients of enchondromatosis affects the occurrence of secondary malignancies.[10,11]

Conclusions

Deformity and limb-length discrepancies due to Ollier's disease were treated successfully by distraction osteogenesis. Enchondromas may arise in distracted calluses when they are osteotomized intralesionally. The stability of external fixators is sufficient to lengthen limbs and correct deformities even if the wires and half-pins are inserted intralesionally.

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