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ORIGINAL STUDY

Humeral lengthening by distraction osteogenesis : A safe procedure ?

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This study was conducted to assess the safety of humeral lengthening using an Ilizarov frame. We retrospectively reviewed 26 humeral segments in 17 patients that were lengthened at our department between 1993 and 2011. There were varying aetiologies including achondroplasia, epiphyseal dysplasia, Ollier disease, trauma or infection of the proximal humeral growth-plate, unicameral bone cyst and brachial plexus injury. Mean age at start of surgery was 17.05 years (range : 5-40).

The mean lengthening achieved was 8.85 (3-13) cm. Mean lengthening percentage was 35.3% (range : 10-48). Average healing index was 30.56 days/cm (range : 17.46-42.32). There was a significant difference in healing index between achondroplasia patients (28.79 days/cm) compared to others (33.41 days/cm). Minor problems included pin tract infection (14 segments). More important obstacles were temporary elbow flexion contracture (7 segments), premature consolidation (6 segments), radial nerve dysaesthesia (6 segments) and loosening of a Schanz screw (1 segment). Complications included one fracture and one progressive bowing after frame removal. One planned lengthening was not completely achieved.

Despite a lot of obstacles, humeral lengthening using an Ilizarov frame provided a reliable method to treat the functional or cosmetic problems of upper limb shortening.

Keywords : humerus ; lengthening ; Ilizarov ; achondroplasia.

INTRODUCTION

Bone distraction has become increasingly popular over the past three decades owing to the influence of Ilizarov's method. Experiences with lower limb lengthening and angular deformity correction have been extensively published but the application of the Ilizarov method to the upper limb appears to be less popular. This may be because upper limb deformities are expected to cause less functional impairment (*17*), but also for fear of complications that might outweigh the benefit of lengthening. To evaluate the safety of humeral lengthening, we retrospectively evaluated 30 procedures performed in our institution, with particular interest for complication rate and its relation with the amount of lengthening.

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PATIENTS AND METHODS

Between 1993 and 2011, 20 patients were referred to our department for upper limb lengthening. Because of insufficient follow-up data, 3 patients (1 with achondroplasia and 2 with septic growth arrest) had to be excluded. All remaining 17 patients, their aetiology and a short overview of their treatment are listed in Table I. In 8 patients (2 male, 6 female) with a short stature syndrome, a bilateral lengthening was performed (16 segments). The remaining 9 patients (4 male, 5 female) underwent a unilateral lengthening ; one patient underwent the procedure twice due to his young age at the first procedure and to development of a new limb length discrepancy later in adolescence (10 segments). In total, the lengthening of 26 humeral segments was retrospectively reviewed.

All operations were performed by the senior author (JL) according to Ilizarov's method with the technique as proposed by Catagni *et al*, using a proximal arch and one or two distal rings and comparable to the constructions used by the authors for the treatment of humeral nonunion (*11*). The osteotomy was always performed minimally invasive just distal to the level of the deltoid insertion, after lifting the periosteum from the bone and making drill holes to weaken the cortices. Completion of the corticotomy was done with a chisel and manual osteoclasis and was confirmed using fluoroscopy.

Physiotherapy was started on the first postoperative day and during the stay in the hospital, patients were instructed how to manage their pins and how to perform the distraction. Lengthening was started on the fifth postoperative day at a rhythm of 4 × ¼ mm per day. Clinical and radiographical evalutation of the length discrepancy, callus formation and potential complications were performed every two weeks during lengthening. After obtaining the desired length, 7 patients preferred a reduction of their ring frame to a smaller unilateral fixator because it improves comfort and function. This was done as soon as cortical bridging was seen on one side, and according to a technique previously described by the authors (*13*). After reaching the desired length, monthly follow-up was performed until bony healing was radiographically observed, allowing removal of the fixator. No further protective brace was applied. and the minimal follow-up period was at least two years. Differences in the healing rate were analysed performing a pairwise comparison between the sample means of the healing index in achondroplasia and in the other patients using the Student's t-test. Data collection and analysis was performed with Microsoft Excel.

Complications were described according to Paley's classification (*19*).

RESULTS

The average age of the patients at the start of surgery was 17.05 years (range : 5-40). Mean gain of length in all procedures was 8.85 cm (range : 3-13) and mean lengthening percentage was 35.3% (range : 10-48). The mean healing index was 30.56 days (17.46-42.32) per cm. A comparison of the healing index between achondroplasia patients (28.79 days/cm) and patients with another aetiology (33.41 days/cm) showed a statistically significant difference (p=0.04). After treatment there were no functional restrictions with a return, to the preoperative level of activity including sports practice. All patients were very satisfied, stating that they were prepared to undergo the treatment again if necessary. Pre- and postoperative clinical and radiographic images of an achondroplasia patient with bilateral lengthening are shown in Figure 1.

Most problems were related to pin tract infection (14 segments). They were a major cause of discomfort and pain, but responded well to oral antibiotic treatment according to the antibiogram, except in one patient who had to be hospitalized for an intravenous antibiotic treatment because of the development of cellulitis in the upper part of the arm. A mild elbow flexion contracture of 5-10° was seen in all patients during the treatment despite gentle physiotherapy. A significant contracture exceeding 20° developed in 6 patients (7 segments). After removal of the frame, there was a gradual recovery of the elbow function in all patients. Exuberant callus formation necessitating an increased amount of distraction to prevent premature consolidation was seen in 8 patients (14 segments), 6 of them having achondroplasia. Despite an increased distraction rhythm, a premature consolidation occurred in 4 patients (6 segments), necessitating a manipulation under general anaesthesia.

Dysaesthesia in the radial nerve sensory area developed in 5 patients (6 segments) and 2 of them (3 segments) developed a true transient neurapraxia with a drop hand. When this occurred, 3 patients (4 segments) had a distraction rate exceeding

Table I. — Patient overview

N°	Age	Sex	Side	Diagnosis	Lengthening (percentage)	Healing Index (d/cm)	Monofix	Complications
1	18	M	B	Achondroplasia	R: 12,5 (41%)	R: 19,52	NO	R: Pin tract infection, drop hand
					L: 12,5 (41%)	L: 19,52		L: Pin tract infection, pin loosening
2	15	F	B	Achondroplasia	R: 13 (47%)	R: 24,54	NO	R: Fracture in regenerate during callus maturation
					L: 11 (39%)	L: 29		R: Fracture in regenerate after removal
3	20	F	B	Achondroplasia	R: 12 (44%)	R: 39,42	NO	R: Pin tract infection, elbow flexion contracture
					L: 12 (44%)	L: 33,42		L: Pin tract infection, radial nerve dysesthesia
4	9	F	B	Achondroplasia	R: 11 (48%)	R: 42	YES	R: Premature consolidation, elbow flexion contracture
					L: 11 (48%)	L: 42		L: Premature consolidation, elbow flexion contracture
5	17	M	R	Ollier	6 (27%)	33	NO	Pin tract infection
6	13	F	B	Achondroplasia	R: 6,9 (35%)	R: 24,35	NO	R: None
					L: 7 (35%)	L: 24		L: None
7	16	F	L	Brachial plexus lesion	3 (10%)	35	NO	Pin tract infection, radial nerve dysesthesia
8	17	M	L	Osteomyelitis	5 (17%)	35,6	YES	None
9	17	M	L	Unicameral cyst	11,2 (37%)	42,32	YES	Fracture in regenerate during callus maturation
10	37	F	L	Ollier	10,2 (43%)	35	NO	Elbow flexion contracture, pin tract infection, residual discrepancy
11A	5	F	L	Traumatic epiphysiolysis	7,9 (37%)	29,14	YES	Elbow flexion contracture, pin tract infection
11B	11	F	L	Traumatic epiphysiolysis	7,2 (24%)	35,28	YES	Elbow flexion contracture, pin tract infection
12	18	F	R	Osteomyelitis	11,1 (35%)	26,67	YES	Pin tract infection
13	16	F	B	Achondroplasia	R: 7,8 (31%)	R: 25,77	NO	R: Premature consolidation
					L: 8,1 (32%)	L: 24,81		L: Pin tract infection, postoperative bowing
14	8	M	B	Achondroplasia	R: 5,2 (35%)	R: 19,81	NO	R: Premature consolidation, drop hand
					L: 5,9 (39%)	L: 17,46		L: Premature consolidation, drop hand
15	14	F	R	Epiphyseal dysplasia	7,5 (26%)	27,47	YES	Pin tract infection
16	40	M	R	Traumatic epiphysiolysis	4,1 (13%)	34,63	NO	Pin tract infection, elbow flexion contracture
17	16	F	B	Achondroplasia	R: 10,5 (45%)	R: 37,52	YES	R: Premature consolidation, fracture in regenerate during callus maturation, pin tract infection
					L: 10,5 (45%)	L: 37,52		L: Radial nerve dysaesthesia

R = right, L = left, d = days.

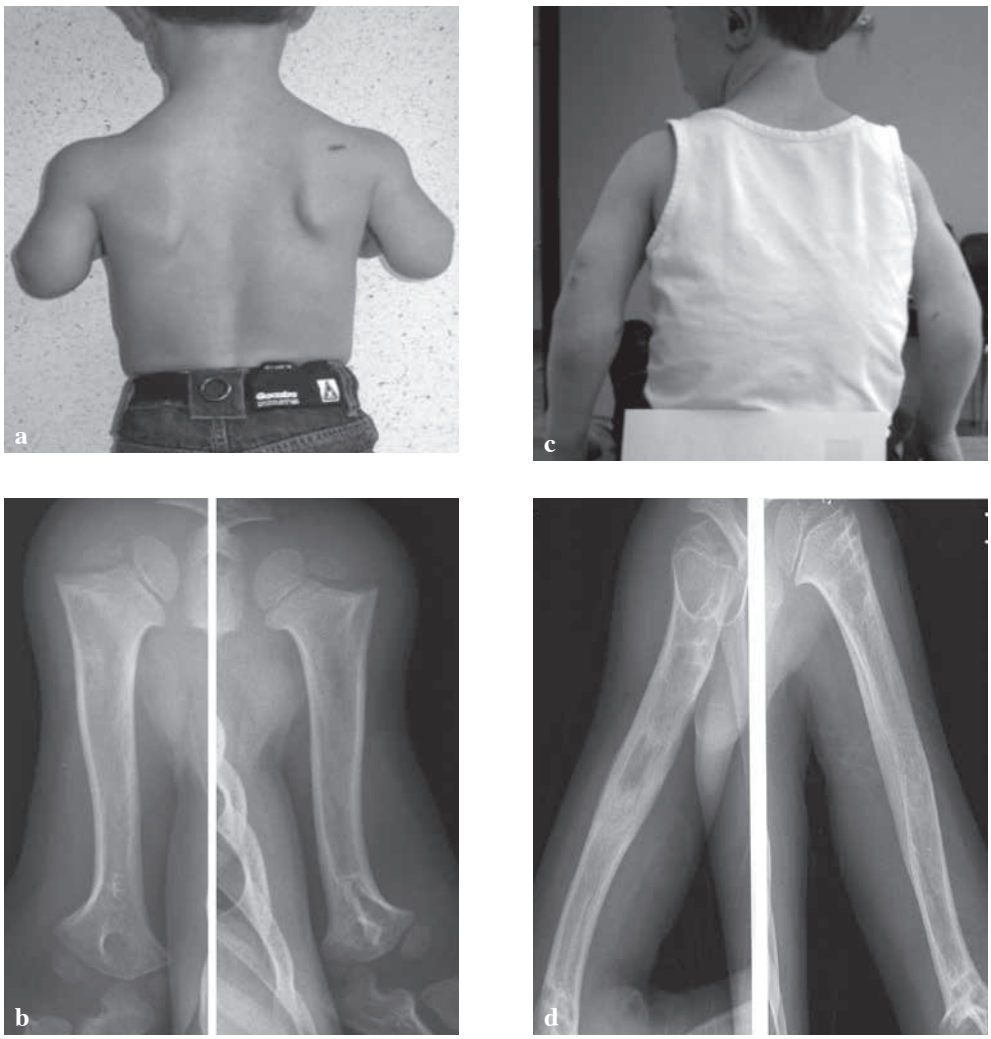

Fig. 1. — Example of an achondroplastic patient who had a lengthening of 5.2 cm of his right humerus and 5.9 cm of his left humerus (a,b : preoperative clinical image and radiograph – c,d postoperative images).

1.5 mm/day to overcome a potential premature fusion. A short interruption of distraction during 2 weeks was sufficient for recovery of the nerve function. After recovery, lengthening was continued at a slower rate. Three patients (3 segments) had a fracture in the distraction callus after a minor injury during bony maturation. The amount of lengthening in those patients was exceeding 10 centimetres. Loosening of a Schanz screw was noted in one patient.

Two patients experienced a complication after removal of the apparatus. One patient sustained a fracture that was fixed with a unilateral frame until bony healing ; in the other patient, a progressive bowing occurred, for which re-osteotomy, acute correction of the bowing and fixation with a unilateral frame was performed. In one patient a residual discrepancy of 4 cm was accepted.

DISCUSSION

The first publication of a humeral lengthening in English-language literature was by Dick and Tietjen who performed a distraction of the upper arm in 1978 according to Wagner's technique (5). One year later, the first article in French appeared, authored

by Paneva-Holevitch and Yankov, who described two cases of cubitus varus correction with lengthening (*20*). According to their description one patient healed spontaneously whereas the other had to undergo bone grafting.

The philosophy about distraction bone healing definitely changed with the introduction of the Ilizarov method. Due to the use of stable circular fixation, percutaneous corticotomy, and slow distraction with three-dimensional correction possibilities, the correction and lengthening of the upper limb became safer and easier. Since then, several reports have shown successful humeral lengthening using the Ilizarov technique, the monolateral fixator (*16,21*) and more recently, the Taylor Spatial Frame (*1*). Nevertheless, although lower limb lengthening is performed on a daily basis in specialized centres, upper limb lengthening is perfomed less often : few large series of humeral lengthenings were reported in the literature during the last decades (*1,4,7,8,10, 14,16,21*). There are several reasons making distraction of the upper arm less popular. First of all, a unilateral shortening of the humerus does not always cause a functional impairment, unless there is a major difference. Discrepancies of less than five centimetres do not seem to impede patients' normal function and are not elected by the surgeon for correction (*17*). Furthermore both congenital and acquired shortenings of the humerus are often associated with an abnormal shoulder joint configuration and limited function compared to the other side, which cannot be overcome by the lengthening procedure. Finally, external fixation and lengthening of a limb is known to cause complications such as radial nerve problems. However, shortness of the upper extremity may lead to problems with personal hygiene and less functional performance, and there is sufficient evidence that upper limb lengthening is an excellent option for moderate to severe discrepancies (*9,10,21,22*). Nevertheless, in our institution, they form a minority of all limb lengthenings, as we could only retrieve 20 patients with 30 humeral lengthenings over the past two decades.

Regarding functional outcome, upper limb distraction seems to pose less problems than lower limb lengthening (*9,14*). In our series, after treatment, all patients returned to their preoperative level of activity including sports practice with preservation of shoulder and elbow function, despite an elbow flexion contracture of 5-10° during lengthening in all patients and even greater than 20° in 6 patients, which recovered after removal of the frame with gentle physiotherapy.

Concerning complications, it has been reported that pin tract infection is the most frequent problem and this is supported by our findings (*2,7,8,16*). For achondroplasia patients, it has been noted that bone formation was better in the upper than in the lower limb (*9,23*). Our average healing index was 30.56 days/cm, comparable with the healing index reported in literature (*7,8,16,23*). When comparing the healing index between achondroplasia patients and those with another aetiology however, we found a significant difference. To our knowledge, there is no clinical evidence so far that achondroplasia patients have a faster healing index when compared to normal subjects. This finding of excellent bony healing is also reflected in the frequent abundant callus formation noted during lengthening. This occurred in 8 patients of whom 4 had a premature fusion, and this respectively after 24 (15%), 41 (20%), 11 (7.5%) and 10 (6%) millimetres lengthening. They were all achondroplasia patients, and it is known from clinical observations that despite their disturbed endochondral bone formation due to a fibroblast growth factor receptor 3 (FGFR3) mutation, the callus formation during distraction is excellent (*3,6*). That the premature fusion in our series was not caused by an incomplete corticotomy is clearly illustrated by the fact that all patients were able to distract several centimetres with clear separation of bone fragments on radiography before the problem was encountered.

A known serious problem during upper limb lengthening is radial nerve paresis and this was also encountered in our series. It never occurred perioperatively but as a consequence of an increased distraction rate (> 1.5 mm/day) to prevent premature bony consolidation. It is known that nerves tolerate limb lengthening poorly and this is electrophysically proven (*15*). After occurrence, the distraction was halted until resolution of the neurological irritation. Afterwards, a normal distraction rhythm of

4 × ¼ mm per day was restored and no recurrent radial nerve irritation occurred, even if a large distraction was achieved. In view of this finding, we conclude that high distraction rhythm rather than total amount of lengthening is a risk for radial nerve neurapraxia. Remarkably, after cessation of the increased distraction rhythm no premature healing occurred anymore during further distraction, indicating that a short period of supraphysiological distraction is sufficient to prevent further exuberant callus formation.

A residual shortening of 4 centimetres was accepted in one patient, but the main indication was correction of a major deformity in a severely deviated limb due to Ollier's disease ; a concomitant lengthening of 10 centimetres was nevertheless achieved, illustrating the possibilities of the three-dimensional corrective possibilities of the Ilizarov method. In 3 humeral segments, a fracture line in the distraction callus was noticed on radiographs during maturation of the callus and was according to the patients due to a minor injury. These 3 segments were lengthened more than 10 cm, which may explain the vulnerability of the lengthened segment. A fracture in the bone regenerate after removal of the apparatus was documented in one segment (which already sustained a fracture in the distraction callus during lengthening). It is a known complication previously described (*7,9,14*) but it is less frequent when compared to lengthening of the lower limb (*1*). A progressive anterolateral bowing after removal of the fixator due to plastic deformation of the distracted segment was noted in one segment, necessitating a corrective osteotomy. These complications show the importance of an optimal timing for the removal of the fixator. A recent method to evaluate corticalisation of the regenerate is the pixel value ratio (*9,22*), which can lower the risk of a re-fracture or gradual bowing of the regenerate but which was not used in this series.

Our study has its limitations. It is a retrospective review of patients who had a humeral lengthening, only allowing the review of 26 segments in 17 patients. Moreover, there is a lack of true patient specific outcome measurements. Nevertheless, it represents a single surgeon's experience with humeral lengthening and gives a good review of the problems encountered during the treatment. It also proves that bony healing in achondroplasia patients is slightly better when compared to normal subjects.

CONCLUSION

A review of this series indicates that humeral lengthening is a suitable procedure with excellent results regarding functional performance and self-image. However there is a relatively high complication rate, with most of them being minor and self-limiting. This study also highlights the excellent bone forming capacity in case of achondroplasia, making these patients more prone to premature fusion and transient radial nerve paralysis because of an increased distraction rhythm.

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