■ CHILDREN'S ORTHOPAEDICS

Bilateral humeral lengthening in achondroplasia with unilateral external fixators

IS IT SAFE AND DOES IT IMPROVE DAILY LIFE?

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A retrospective study was performed in 18 patients with achondroplasia, who underwent bilateral humeral lengthening between 2001 and 2013, using monorail external fixators. The mean age was ten years (six to 15) and the mean follow-up was 40 months (12 to 104).

The mean disabilities of the arm, shoulder and hand (DASH) score fell from 32.3 (20 to 40) pre-operatively to 9.4 (6 to 14) post-operatively (p = 0.037). A mean lengthening of 60% (40% to 95%) was required to reach the goal of independent perineal hygiene. One patient developed early consolidation, and fractures occurred in the regenerate bone of four humeri in three patients. There were three transient radial nerve palsies.

Humeral lengthening increases the independence of people with achondroplasia and is not just a cosmetic procedure.

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Achondroplasia causes disproportionate short stature with rhizomelic involvement. Upper limb lengthening[1-6] has been reported less extensively than lower limb lengthening. Although upper limb lengthening is better tolerated, it is seen as a cosmetic issue, rather than a functional necessity, by some authors.[3-10] The goals of lengthening are to restore normal proportions, improve reach, and improve the ability to perform perineal personal hygiene.[6]

Humeral lengthening with circular external fixators,[2,11-15] monolateral frames[6,16,17] and multiaxial frames[18-20] has been described. In achondroplasic patients, such lengthening is usually accomplished with circular and monolateral frames.[6,12,14,17,21] Although a monorail system seems more comfortable for patients, and its use has been described in case reports,[16,22] there is insufficient evidence regarding its efficacy and safety.

Humeral lengthening with circular external fixators have been reported to provide lengthening of 8 cm to 9 cm within seven to eight months of fixation.[2,6,12] Complications include transient radial nerve paresis in 6% to 10%,[2,6] refracture in 10% to 14%,[6,12] and a variable rate of pin-tract infection. Pawar et al[6] showed that humeral lengthening with monolateral external fixation is well tolerated by patients and is an effective means of improving patient function with a rate of complication similar to that of circular frames.

We assessed functional improvement after lengthening both humeri with monorail external fixators in patients with achondroplasia in order to evaluate the safety of the procedure. In addition, we determined the extent of humeral lengthening (in cm), percentage terms and the ability to enable the patient to reach the perineum for the purpose of personal hygiene.

Patients and Methods

The study was approved by the Institutional Review Board. We retrospectively reviewed patients with achondroplasia, who underwent bilateral humeral lengthening between 2001 and 2013, using monorail external fixators. There were 36 humeri in 18 patients (ten females, eight males) with a mean age of ten years (six to 15).

Patients were recalled to determine the disabilities of the arm, shoulder and hand (DASH) score,[23] which is designed to measure physical function and symptoms in people with musculoskeletal disorders of the upper limb. Other data were obtained from medical records and radiographs of the Istanbul University Istanbul Medical Faculty Department of Orthopaedics and Traumatology.

Operative technique. Under general anaesthesia, the patient is placed supine on a radiolucent table. The radial nerve is routinely located via a lateral incision over the distal humerus and protected during placement of the hydroxyapatite-coated Shanz screws (Tasarımmed, Istanbul, Turkey). The four half pins are placed under fluoroscopic guidance in the midaxial

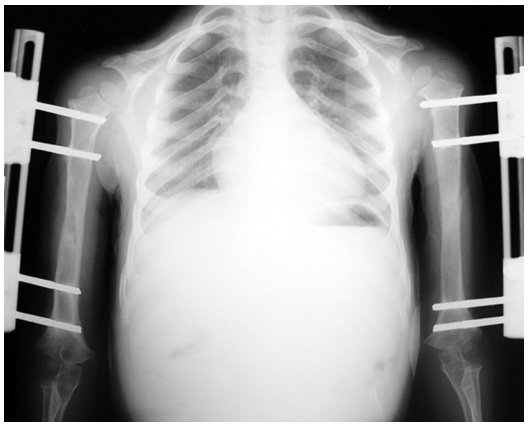

Fig. 1

Radiograph demonstrating consolidation of the humeri after bilateral lengthening with monorail external fixators in a patient with achondroplasia just before the removal of the external fixators after six months *in situ*.

Fig. 3

Photograph demonstrating that the same patient as in Figure 1 is able to reach the perineal and sacral regions after humeral lengthening just seven months after the operation and 1.5 months after the removal of external fixators.

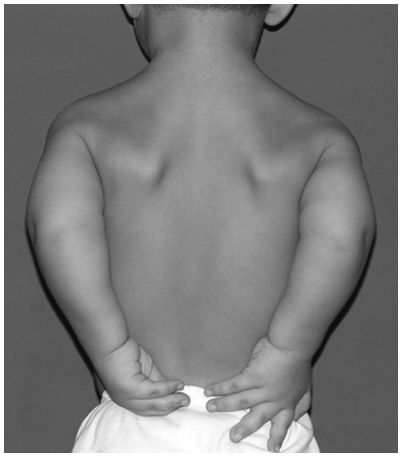

Fig. 2

Pre-operative clinical photograph of a patient aged seven years (no.1) who is not able to reach the perineal area.

line, perpendicular to the bone in the frontal plane. To ensure precise placement of the half pins, we first inserted a Kirschner wire under fluoroscopic control and checked the placement of the wire. We then drilled with cannulated drills over the wire before inserting the half pin in the correct position. This method has been referred to the cannulated wire technique.[6] The first pin is inserted proximal to the olecranon fossa. The second pin is placed under the guidance of the monolateral rail in the proximal humerus in the deltoid area, and the middle two pins are placed through the unilateral external fixator with direct visualisation of the radial nerve. In the presence of deformity, we place the half pins separately and perpendicular to the bone for acute correction. An osteotomy of the mid diaphysis is performed, distal to the insertion of the deltoid muscle, using a multiple drill-hole technique. Both humeri are lengthened, starting on the fifth post-operative day, four times per day at a rate of 1 mm/day.

An unrestricted range of active and passive movement of the elbow and shoulder is initiated on the second post-operative day. Physiotherapy is supervised three days a week and home exercises are recommended. Lengthening is continued until the patient's hand is able to reach the perineum without difficulty. The external fixators are removed when three cortices are visible on anteroposterior and lateral radiographs of the regenerate (Fig. 1).

After removal of the fixator, the humerus is secured with a personalised humeral functional brace for six to 12 weeks. During this period, the patients are not allowed to bear load through the arm.

Evaluation. Patients were reviewed every two weeks while lengthening progressed, and monthly during consolidation until removal of the external fixator.

We recorded the pre- and post-operative DASH scores,[23]and complications, bone and functional scores according to Paley et al[24] and Paley.[25] The radiological lengthening of the humerus was measured in the sagittal plane (HİB,SGB). The percentage lengthening was calculated as the length of the distracted segment divided by pre-operative length of the intact humerus. The external fixation index[26] was calculated by dividing the number of days wearing the frame by the amount of lengthening achieved (in cm).

Statistical analysis. Pre- and post-operative DASH scores were compared using two-tailed paired sample *t*-tests with SPSS v16.0 software (SPSS Inc., Chicago, Illinois). A p-value < 0.05 was considered statistically significant.

Table I. Results and functional scores

Patient no	Age at operation (yrs)	Gender	Length of follow-up (mths)	Lengthening amount (%)	Lengthening amount (cm)	EFI (day/cm)	Complications	Paley's bone score R/L	Paley's functional score R/L	Post-op DASH score	Pre-op DASH Score
1	7	M	41	79	7.5	22.0		Excellent/ good	Excellent/ good	9.2	35.8
2	7	F	15	52	6.5	19.2	Late radial nerve palsy	Excellent/ Excellent	Excellent/ excellent	7.5	31.7
3	13	M	12	60	8.0	33.7		Excellent/ Excellent	Good/good	6.7	37.4
4	8	F	104	73	8.0	30.6		Excellent/ excellent	Good/ excellent	10.2	34.2
5	7	M	88	92	9.5	17.3	Grade 2 pin-tract infection, early radial nerve palsy	Excellent/ excellent	Good/ good	8.3	32.7
6	15	F	75	50	8.0	28.1	20° loss of extension, early consolidation, bilateral fracture after removal of fixators	Good/ good	Good/ good	8.3	20.4
7	7	F	43	95	10.0	18.4		Excellent/ excellent	Good /good	14.1	34.2
8	11	F	52	45	7.0	23.4	Grade 1 pin-tract infection	Excellent/ excellent	Good/ excellent	8.5	37.5
9	11	M	60	45	8.0	30.6		Excellent/ excellent	Excellent/ good	12.2	34.3
10	11	F	44	54	7.0	21.4	Early radial nerve palsy	Excellent/ excellent	Good/ excellent	8.2	40.2
11	6	M	38	80	8.0	28.7	Grade 2 pin-tract infection, humerus fracture after removal of fixator	Good/ excellent	Good/ excellent	6.5	33.3
12	15	F	43	44	7.0	30.0		Excellent/ excellent	Excellent/ excellent	8.3	37.8
13	10	F	18	56	7.0	24.2		Excellent/ excellent	Excellent/ excellent	7.5	27.5
14	10	M	32	70	8.0	18.7		Excellent/ excellent	Good/ excellent	13.2	33.2
15	7	F	17	40	7.0	29.2	Humerus fracture after removal of external fixator	Excellent/ excellent	Excellent/ good	13.1	25.5
16	10	M	19	43	7.0	23.0	Grade 1 pin-tract infection	Excellent/ excellent	Excellent/ excellent	9.7	29.2
17	14	M	12	52	8.0	24.0		Excellent/ excellent	Excellent/ excellent	10	25.1
18	11	F	13	53	8.0	17.1		Excellent/ excellent	Excellent/ excellent	6	32.5

EFI,external fixator index; Pre-op, pre-operative; Post-op, post-operative; DASH, disabilities of the arm, shoulder and hand; R, right; L, left

Results

The mean follow-up was 40 months (12 to 104). No patient was lost to follow-up. The ages, gender, length of follow-up, amount of lengthening and individual outcomes are given in Table I. The mean lengthening was 7.8 cm (6.5 to 10). The mean external fixator index was 24.1 day/cm (17.1 to 33.7). The mean external fixator time was 185 days (120 to 270). The mean proportionate lengthening was 60% (40% to 95%).

There were two grade I and two grade II pin-tract infections according to Paley et al,[24] which were treated with dressings and oral antibiotics. One patient had a unilateral 20° loss of elbow extension. No patient developed inferior subluxation of the shoulder joint. There were two patients with early transient radial nerve palsies, for which we did not stop the lengthening, and one with radial nerve palsy which occurred later, for which the lengthening was discontinued at 6.5 cm. All nerve palsies resolved without sequelae, which for the early presentations was in three and five weeks. The late presentation resolved in 14 weeks.

According to Paley's bone scoring system, 32 segments were excellent and four were good. One patient developed early consolidation that required re-osteotomy. Fractures occurred in the regenerate bone of four humeri after removal of the external fixators. In one patient this occurred after one week and in the others, three weeks. One was treated with an intramedullary Rush pin, one was treated with external fixation and the other two were treated with Sarmiento braces.

According to the Paley functional scoring system, 20 segments were excellent and 16 were good.The mean DASH score was 32.3 (20.4 to 40.2) pre-operatively and 9.4 (6 to 14.1; $p = 0.037$) at final review. Preparing a meal, placing an object over the head, washing the back and hair, going outside, daily and social activities, and feeling more capable of doing daily activities, were the most improved functional scores. All but one patient had no difficulty reaching their perineum and could use the bathroom independently (Figs 2 and 3).

Discussion

This study has some limitations. It was a retrospective case series. The operations were performed by surgeons experienced in the lengthening procedure, therefore it might not be possible to generalise our experience.

Patients complain about not being able to reach the perineum and perform personal hygiene independently; the lengthening procedure improves this aspect of their lives. A longer humerus increases the working area of the hand in space. The ability to be more social and leave the confines of the home accounts for the large decrease in DASH score.

Our study demonstrated a substantial improvement of 23 points in the mean DASH score after humeral lengthening in patients with achondroplasia. Pawar et al[6] reported a five-point improvement after 7 cm of unilateral or bilateral lengthening. We do not determine the amount of the lengthening according to the long bone ratio as suggested in the literature.[27] We aim to restore the patient's ability to perform perineal hygiene. We check the ability of the patient to reach the perineal area from the back and front with both hands without the help of others. This amount of lengthening also allows the hand to reach the proximal third of the femur.

In our study, all of the patients had achondroplasia and lengthening was achieved with monorail external fixators. In the literature on humeral lengthening, the results achieved for patients with achondroplasia are typically combined with other diagnoses,[6,9,14,20,21] lengthened with circular external fixators,[12] or monolateral external fixators with multiaxial fixation.[6] In the literature, the biggest series for humeral lengthening comes from three studies[2,12,14] involving a total of 79 segments of 55 patients with achondroplasia. For these patients, the mean external fixator index was 28 day/cm, the mean age was 15 years, the mean lengthening was 8 cm, and the mean lengthening percentage was 40% to 55%,[11] compared with a mean of 60% and a maximum of 95% achieved in our study.

Although hyperextensibility of joints is common in achondroplasia, elbow stiffness is observed in 0% to 7% of cases.[6,14,28] Loss of up to 30° of elbow extension does not greatly affect daily function, being much better tolerated than loss of extension in knee and hip joints.[29] Loss in extension in the elbow joint is a common problem found in achondroplastic patients.[30] The use of multiplanar wires and half pins with circular and monolateral frames decreases the range of movement.[6] The use of the monorail system, in which uniplanar half pins do not restrict the elbow movement, and specialist physiotherapy after surgery, were our main approaches to preventing elbow stiffness.

Radial nerve palsy can be associated with the osteotomy, introduction of pins or distraction.[12,14] In an anatomical study on the insertion of half pins in the distal humerus with monolateral frames, pins passed through, or were directly in contact with, the radial nerve in 13 of 40 cases.[31] Especially in short and thin humeri, we believe that insertion of half pins in the distal one third of the humerus and percutaneous osteotomy in such short humerei, increases the risk of radial nerve palsy. We recommend radial nerve exploration using the cannulated wire technique and fluoroscopic guidance to prevent radial nerve injuries and to restore the mechanical axis during the insertion of the half pins. Pawar et al[6] suggested the use of regional anaesthesia and neural monitoring in all humeral lengthening procedures. In our practice, we start the operation with exploration and release of the radial nerve, and we introduce the pins and perform the osteotomy under direct vision. In the event of early post-operative radial nerve palsy, we continued the lengthening as we knew the nerve was intact. In contrast, we discontinued the lengthening when a palsy occurred later on during the process of lengthening.

The potential of the humerus for effective lengthening is recognised. Callus formation during distraction occurs at a significantly higher rate than in the tibia.[32] In the lower extremity, the incidence of complications increases when the lengthening exceeds 20%,[9] but this does not occur with the humerus. The elbow and shoulder are not prone to subluxation.[9] Monolateral external fixators are more comfortable than circular ones and produce adequate stability as the humerus is a non-weight-bearing bone.[9]

In our experience, humeral lengthening is safe and effective in patients with achondroplasia. Patients are able to continue daily activities during the distraction and consolidation phases. The ability to perform personal hygiene decreases dependence on others. Humeral lengthening achieves fundamental functional and social goals, as well as a cosmetic improvement.

Author contributions:
H. I. Balci: Data collection, analysis, performed surgeries, writing the paper.
M. Kocaoglu: Analysis Performed surgery, writing the paper.
C. Sen: Analysis Performed surgery, writing the paper.
L. Eralp: Analysis, Performed surgery, writing the paper.
S. G. Batibay: Data collection, analysis, Performed surgery, writing the paper.
K. Bilsel: Data collection, analysis, Performed surgery, writing the paper.

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