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ORIGINAL ARTICLE

Surgical technique and outcomes for bilateral humeral lengthening for achondroplasia: 26-year experience

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Abstract

Background Elongation in patients with achondroplasia provides better overall skeletal proportionality and significantly improves such individuals' access to their perineal region to self-manage personal hygiene. This paper describes our surgical technique and outcomes for bilateral humeral lengthening in achondroplasia patients over 26 years.

Methods Ours was a retrospective study of 55 patients with achondroplasia-related short stature, in whom bilateral humeral lengthening was performed from 1990 to 2016. We describe the surgical technique and analyze mean gain in humeral length, days using an external fixator, mean percentage of lengthening, external fixation index, type of callus, and complications. Pre- and postoperative radiographic measurements were obtained. Patients also were contacted by telephone and asked about their ability to perform peri-anal self-hygiene and about their overall satisfaction.

Results In total, 110 humeri were lengthened (28 males and 27 females) with medium elongation of 9.5 cm on the right and 9.6 cm on the left, while averaging 220 days in an external fixator. We observed 14 minor complications. There was no significant association between pin position and type of callus, and elongation most often external and in the presence of a straight callus. Before elongation, 77.1% of patients reported difficulties with perineal hygiene and 85.4% could not put their hands in their pockets. Upon completion of lengthening, 100% could perform both tasks and 94.5% were very satisfied.

Conclusions Bilateral humeral elongation yields significant improvements in patient autonomy, with a relatively low complication rate and very high patient satisfaction.

Keywords Humeral lengthening · Achondroplasia · Surgical technique

Introduction

Achondroplasia is the most common form of short-limb dwarfism in humans, affecting more than 250,000 individuals worldwide. More than 95% of patients have a mutation in the gene for fibroblast growth factor receptor 3 (FGFR3) and more than 80% of these are new mutations [1].

It is short-limb dwarfism with rhizomelia, as infants have upper-extremity deformities isolated to the humerus, such that they are disproportionally short relative to their forearms, leading to functional, cosmetic, and psychological problems. Moreover, lack of full extension of the elbow is a common abnormality in achondroplasia, mainly caused by a flexion deformity involving the distal humerus and subluxation of an abnormally shaped radial head [2]. This limb disproportion and lack of elbow extension cause functional disability which manifests as problems carrying out fundamental daily activities, like taking care of peri-anal personal hygiene, which often must be performed with the help of another person or using devices specifically designed for this purpose. Moreover, this dysfunction worsens if the lower extremities are lengthened [3, 4].

Lower limb lengthening has become one of the standard modalities for treating achondroplasia patients with functional deficits. Although some centers and groups believe in cosmetic lengthening, it is not universally accepted among surgeons [5–7]. As results are encouraging, more and more achondroplasia patients are now opting for humeral lengthening after femoral and tibial lengthening.

Gradual mechanical distraction can be used for humeral lengthening, using either monolateral or circular external fixator and intramedullary nailing; however, external fixation is the preferred method for this procedure. Monolateral

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fixators are considered easier to apply and can be used when deformity correction is not necessary or less lengthening is desired [8].

Compared to lengthening of the femur or tibia, the literature regarding humeral lengthening for achondroplasia is scarce [4, 9]. Consequently, the two main purposes of this paper are (1) to describe the bilateral humeral lengthening technique we have used in our achondroplasia patients over the past 26 years and (2) to report our outcomes.

Materials and methods

We conducted a retrospective study involving 110 humeral lengthening procedures in 55 patients with achondroplasia admitted to our institution between 1990 and 2016. All humeral lengthening procedures were performed using monolateral external fixators. All 55 patients had already undergone bilateral tibial lengthening, while femoral lengthening was performed either before or after humeral lengthening. Only patients with complete medical records and radiographs were included in the study. Clinical and radiographical evaluations of the humerus were used to measure pre- and postoperative humeral lengths.

Operative technique for humeral lengthening

Every surgery was performed by the same surgeon, who already had extensive experience with lengthening procedures. Surgery was performed under general anesthesia, with the patient supine on a radiolucent table. In all patients, both limbs were lengthened simultaneously.

First, two pins were inserted into the proximal metaphyseal region. These pins were drilled through two cortices, as proximally as possible, while respecting growth cartilage. The two proximal pins were located side by side, parallel to each other percutaneously, and perpendicular to the anatomical axis of the segment. Then two supracondylar pins were placed in the same way. For this procedure, 4- or 5-mm-diameter pins were used, stainless steel pin screws before 2007 and hydroxyapatite pin screws from 2007 onwards.

In each limb, after the pin screws were inserted, an osteotomy was performed. This osteotomy was always performed percutaneously at the deltoid tuberosity, after lifting the periosteum from the bone and making drill holes to weaken the cortices, taking care to avoid all neurovascular structures. Completion of the corticotomy was performed with an osteotome, and manual osteoclasis confirmed using fluoroscopy.

In all patients, the two humeri were lengthened simultaneously using monolateral external fixators. The correct insertion of pins and external fixator position were evaluated using an image intensifier.

Postoperative care

Patients were discharged from the hospital 3 days after surgery. Actual lengthening was initiated after 7 days, at a rate of 1 mm/day (0.5 mm every 12 h). The rate was subsequently adjusted, based upon callus morphology on control radiographs performed every 6 weeks.

During the humeral lengthening process, patients were encouraged to have physiotherapy on their shoulders, elbows, and hands. They were permitted to shower daily even while wearing the external fixator.

Lengthening was considered complete when patients were able to put their hands inside their pockets and reported being able to perform personal hygiene on their own. In case of patients who failed to complete lengthening process, these patients were also included in the final statistical analyses. At the end of lengthening, the fixator was removed if either of the three or four cortices had shown satisfactory corticalization.

Radiological evaluation

Total humeral length gained was determined from radiographs taken before the distraction and immediately after removal of the fixation pins, adjusted for the effect of magnification. We analyzed the mean lengthening percentage and the external fixation index (EFI), the former calculated as the mean increase in humeral lengthening relative to the initial humeral length, expressed as a percentage, and the latter calculated by dividing the duration of external fixation (in days) by the extent of lengthening (in cm).

Callus formation was evaluated, with the shape of the callus categorized into three types, in accordance with Hamanishi's classification scheme [10]: (1) external (a barrel-like fusiform callus just wider than the original bone); (2) straight (a homogeneous callus just as wide as the original bone); and (3) attenuated (a callus narrower than the original bone with an attenuated mid portion). Callus categorization was based upon the width of the callus at the time of fixator removal. The distance from pins to the osteotomy line was measured, and complications were documented during lengthening, using Paley's classification system [11], which classifies complications into problems, obstacles, minor complications, and major complications.

Questionnaire

All subjects were contacted by telephone and asked to participate in a brief telephone interview, during which they were asked to answer either 'Yes' or 'No' to the following two two-part questions: (1A) Could you take care of your

own personal hygiene in the peri-anal region on your own prior to surgery? (1B) Can you do it now? (2A) Could you put your hands into your trouser pockets prior to surgery? (2B) Can you do it now? They then were asked (3A) to rate their level of satisfaction with the humeral elongation process, as dissatisfied, satisfied, or very satisfied, and if dissatisfied, (3B) the reason(s) for their dissatisfaction.

Statistical analysis

To compare the distances from the second pin to the osteotomy line, by the type of callus, one-factor analysis of variance (ANOVA) was used, with multiple comparison correction by Scheffé. To evaluate changes in perceived abilities before and after lengthening (for personal peri-anal hygiene and putting their hands into their pockets), McNemar's test for paired proportions was used. The relationship between satisfaction and the type of osteotomy was evaluated using Fisher's exact test. For all tests, only p values < 0.05 were considered statistically significant, and all tests were two-tailed. All analyses were performed using SPSS version 18.0 (IBM Corp.).

Results

Results are summarized in Table 1. Twenty-eight of the patients were male and 27 female. The average age of patients at the time of surgery was 16.2 years (range: 11.9–25.1). The mean gain in humeral length over all procedures was 9.5 cm for the right humerus (range: 5.7–13), increasing from a mean 16.7 cm to 26.2 cm, and 9.6 cm for the left humerus (range: 3.9–13.9), increasing from 16.6 to 26.2 cm. The average time of external fixator bearing was 220.9 days (range: 113–356).

The mean percentage of lengthening, relative to the initial humeral length, was 56.4% (28–86) in the right humerus and 57.4% (19.5–95) in the left. The external fixation index (EFI) was 24 days/cm on the right (range: 15.7–41.28) and 24.5 days/cm on the left (range: 12.94–44.5).

According to Hamanishi's classification scheme, 35 of the calluses were external, 71 straight, and four attenuated at the time of external fixator removal. The average distance from the second proximal pin to the osteotomy line was 1.5 cm in the right humerus and 1.6 cm in the left. No relationship was detected between the morphology of the callus on radiographs and the position of the humeral osteotomy line (right: $p = 0.85$, left: $p = 0.56$).

Pre- and postoperative clinical and radiographic images of one of the achondroplasia patients postbilateral lengthening are shown in Figs. 1 and 2, respectively. Two problems and 12 obstacles were experienced, for an overall 12.7% complication rate. The obstacles were one premature consolidation, two varus deviations, and nine humeral refractures. The one patient with premature consolidation required a new osteotomy during lengthening; the two varus deviations needed removal of external fixation while lengthening was completed; and the nine humeral refractures occurred after removal of the fixator, but were united following immobilization in a brace. No patient experienced any long-term adverse sequelae. The two problems were both transient radial palsy, both of which resolved within 6 months of surgery with physiotherapy.

During telephone interviews, 77.1% and 85.4% of patients reported preoperative problems with personal peri-anal hygiene and problems putting their hands into their trouser pockets, respectively. However, all patients reported being able to perform both activities, with ease, after humeral lengthening. When we asked about their level of satisfaction, 94.5% claimed to be very satisfied and the remaining 5.5% dissatisfied. The main reasons for dissatisfaction were minor complications during the lengthening process; no one expressed dissatisfaction with the ultimate functional results.

No relationship was identified between patients' satisfaction level and the morphology of the callus or position of the osteotomy line ($p = 0.52$).

Table 1 Summary of humeral lengthening results

	Right side	Left side
Sex	28 males/27 females	
Age (years)	16.2 years (range: 11.9–25.1)	
Mean gain in length (cm)	9.5 (range: 5.7–13)	9.6 (range: 3.9–13.9)
Average time of external fixator bearing (days)	220.9 days (range: 113–356)	
Mean lengthening (%)	56.4 (range: 28–86)	57.4 (range: 19.5–95)
External fixation index (days/cm)	24 (range: 15.7–41.28)	24.5 (range: 12.94–44.5)
Hamanishi's classification		
	33 external calluses	
	67 straight calluses	
	4 attenuated calluses	

Fig. 1 Achondroplasia patient **a** before lower extremity lengthening; **b** after lower extremity lengthening, but before humeral lengthening, creating a disproportionately short humerus relative to the lower extremities; and **c** upon completion of the lengthening process, demonstrating much better proportioned extremity lengths

Discussion

Functional disability secondary to upper-extremity deformities is an important problem in patients with achondroplasia. This functional disability turns into difficulties performing daily activities, which usually worsen after the person's lower extremities are lengthened [3, 4]. For that reason, humeral lengthening is indicated for achondroplasia to improve stature and proportion, especially when the lower extremities have been extensively lengthened, allowing patients to carry out daily activities more autonomously.

The high capacity for bone healing that achondroplasia patients have allows surgeons to perform extensive lengthening that usually is well tolerated because of ligament and other soft tissue laxity [7, 12]. External fixators, including unilateral, multiaxial or circular, and, recently, intramedullary lengthening devices, have been introduced for this purpose [13–15]. We have used unilateral external fixators for every one of our achondroplasia patients undergoing bilateral humeral lengthening over the last 26 years with good long-term results, in terms of average extent of lengthening, consolidation, complications, and patient satisfaction.

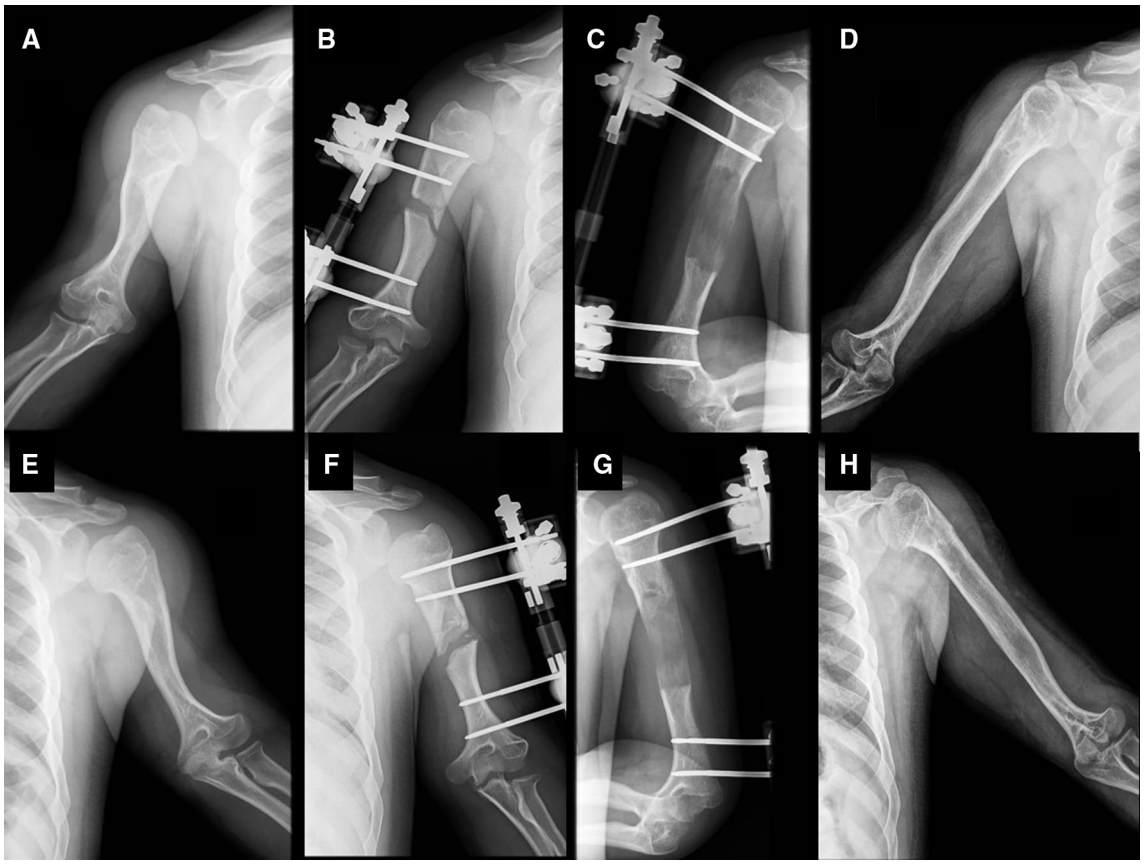

Fig. 2 Right humeral shaft **a** before lengthening; **b** immediately after external fixator placement; **c** at three-month follow-up; and **d** at seven-month follow-up after external fixator removal. Left humeral shaft **e** before lengthening; **f** immediately after external fixator placement; **g** at three-month follow-up; and **h** at seven-month follow-up after external fixator removal

Both the mean lengthening achieved (9.5 and 9.6 cm in the right and left humerus, respectively) and the external fixation index (24 and 24.5 days/cm on the right and left) were comparable to the mean extent of humeral lengthening reported in the literature, which has ranged from 5.0 to 11.1 cm for one-stage procedures not using fibular grafts or plating, and mean external fixation indices for the humerus from 23.6 to 30 days/cm [9, 13, 16, 17]. The overall percentage lengthening we achieved, relative to the initial humeral length, was greater than reported by other authors, at 56.4% and 57.4% versus previously reported percentages from 27 to 50% [13, 16–19].

Regarding the type of callus—utilizing Hamanishi's classifications [10] of external, straight, and attenuated, based on callus width at the time of fixator removal—we cannot compare our results with those of other studies, since no published literature exists describing callus types when removing external fixators. Despite this limitation, we observed no association between the osteotomy line and type of callus.

In terms of complications, only one of our 55 patients experienced premature consolidation, similar to the one case of premature consolidation Nakano-Matsuoka et al. [17] reported in their 54 bilateral humeral lengthening procedures. This contrasts with the higher premature consolidation rate reported by Kim et al. [16], for whom two of twelve patients developed premature consolidation. We observed nine humeral refractures in 110 humeral lengthening procedures (rate of 8.2%), similar to the eight refractures in 108 humeral lengthening procedures (7.4%) reported by Nakano-Matsuoka et al. [17], and the one in 20 (5.0%) reported by Kashiwagi et al. [4]. Other authors have reported slightly higher refracture rates, however, including two refractures in 19 patients (10.5%) reported by Lee et al. [20], and two in 16 (12.5%) described by Hosny et al. [15]. In our patients, all refractures occurred after external fixator removal, and all were treated successfully with brace immobilization without long-term sequelae.

Two of our 55 patients (3.6%) had transient radial nerve palsy developed during surgery, which is also concordant with rates previously reported [4]. The most important finding related to obstacles and complications in our series, however, is that all were transient or successfully treated with no lasting sequelae.

It is important to consider that humeral lengthening is both a difficult procedure and lengthy procedure, requiring a mean 220.9 days in our series (range: 113–356). For this reason, one prerequisite for limb lengthening surgery in achondroplasia patients is a highly motivated patient who understands the benefits and risks of the process. On the other hand, despite the long time required for limb lengthening, patients were able to continue going to school wearing fixators, with little disturbance to their daily activities. One of the main benefits of humeral lengthening for every patient was their ultimate ability to self-manage personal perineal hygiene once the lengthening process was completed. Moreover, no persistent functional restrictions occurred to prevent them from returning to their preoperative level of activity, including athletics.

Our study has limitations. First, ours was a retrospective review. Moreover, there is potential bias in our selection of patients, because one prerequisite for limb lengthening is a motivated patient, which could interfere with postoperative satisfaction rates. Finally, all surgeries were performed by the same experienced surgeon, so our outcomes might not be generalizable to other surgeons and institutions. This said, our complication rates were generally consistent with those already reported in the literature.

In conclusion, the findings of this retrospective review of 55 achondroplasia patients undergoing simultaneous bilateral humeral lengthening using a monolateral external fixator suggest that this procedure is both reliable and safe, with a relatively low complication rate and most complications minor and self-limiting. Most importantly, patients generally appear very satisfied with the surgical results, which include ultimately being able to manage their own personal perineal hygiene and potentially improving their overall autonomy for routine daily activities.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval Hospital ethics committee approval was obtained by the 'Comité de Etica Asistencial Hospital Quiron Salud.'

Informed consent Informed consent was obtained from parents/LAR of all participants included in the study.

References

1. Horton WA, Hall JG, Hecht JT (2007) Achondroplasia. Lancet Lond Engl 370(9582):162–172
2. Kitoh H, Kitakoji T, Kurita K, Katoh M, Takamine Y (2002) Deformities of the elbow in achondroplasia. J Bone Joint Surg Br 84(5):680–683
3. Damsin JP, Ghanem I (2000) Upper limb lengthening. Hand Clin 16(4):685–701
4. Kashiwagi N, Suzuki S, Seto Y, Futami T (2001) Bilateral humeral lengthening in achondroplasia. Clin Orthop 391:251–257
5. Vilarrubias JM, Ginebreda I, Fernandez-Fairen M (1988) 500 cases of lower limb lengthening using a personal technic in achondroplasia. Acta Orthop Belg 54(4):384–390
6. Cai G, Yang L, Saleh M, Coulton L (2007) The effect of tibial diaphyseal lengthening on the longitudinal growth of the tibia. J Pediatr Orthop Part B 16(6):403–407
7. Paley D (1988) Current techniques of limb lengthening. J Pediatr Orthop 8(1):73–92
8. Cattaneo R, Villa A, Catagni M, Tentori L (1988) Strategies for limb lengthening in achondroplasia using the Ilizarov method–the experience of the hospital of Lecco, Italy. Basic Life Sci 48:381–388
9. Shadi M, Musielak B, Koczewski P, Janusz P (2018) Humeral lengthening in patients with achondroplasia and in patients with post-septic shortening: comparison of procedure efficiency and safety. Int Orthop 42(2):419–426
10. Hamanishi C, Yasuwaki Y, Kikuchi H, Tanaka S, Tamura K (1992) Classification of the callus in limb lengthening. Radiographic study of 35 limbs. Acta Orthop Scand 63(4):430–433
11. Paley D (1990) Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. Clin Orthop 250:81–104
12. Yasui N, Kawabata H, Kojimoto H, Ohno H, Matsuda S, Araki N et al (1997) Lengthening of the lower limbs in patients with achondroplasia and hypochondroplasia. Clin Orthop 344:298–306
13. Malot R, Park K-W, Song S-H, Kwon H-N, Song H-R (2013) Role of hybrid monolateral fixators in managing humeral length and deformity correction. Acta Orthop 84(3):280–285
14. McLawhorn AS, Sherman SL, Blyakher A, Widmann RF (2011) Humeral lengthening and deformity correction with the multiaxial correction system. J Pediatr Orthop Part B 20(2):111–116
15. Hosny GA (2016) Humeral lengthening and deformity correction. J Child Orthop 10(6):585–592
16. Kim S-J, Agashe MV, Song S-H, Choi H-J, Lee H, Song H-R (2012) Comparison between upper and lower limb lengthening in patients with achondroplasia: a retrospective study. J Bone Joint Surg Br 94(1):128–133
17. Nakano-Matsuoka N, Fukiage K, Harada Y, Kashiwagi N, Futami T (2017) The prevalence of the complications and their associated factors in humeral lengthening for achondroplasia: retrospective study of 54 cases. J Pediatr Orthop B 26(6):519–525
18. Kiss S, Pap K, Vízkelety T, Terebessy T, Balla M, Szőke G (2008) The humerus is the best place for bone lengthening. Int Orthop 32(3):385–388
19. Kitoh H, Mishima K, Matsushita M, Nishida Y, Ishiguro N (2014) Early and late fracture following extensive limb lengthening in patients with achondroplasia and hypochondroplasia. Bone Jt J 96–B(9):1269–1273
20. Lee FY-I, Schoeb JS, Yu J, Christiansen BD, Dick HM (2005) Operative lengthening of the humerus: indications, benefits, and complications. J Pediatr Orthop 25(5):613–616