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Simultaneous bilateral lengthening of femora and tibiae in achondroplastic patients

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Objective: The aim of this study was to analyze the results and complications of simultaneous bilateral femoral and tibial lengthening in achondroplastic patients.

Methods: The study included the 44 femora and 44 tibiae of 22 achondroplastic patients (16 females, 6 males; mean age: 6.36 years, range: 3 to 11 years) that underwent simultaneous lengthening. Orthofix LRS monolateral fixators were used for femoral lengthening and either Ilizarov-type or hexapod-type circular external fixators for tibial lengthening. Tenotomies of the hip flexors and the Achilles tendon were performed to prevent injury to the growth plates and to prevent joint contractures. Results and complications were evaluated according to Paley's scoring and complication systems.

Results: Average follow-up time was 35 (range: 26 to 76) months. The femora were lengthened by an average of 7.07 cm (46.1%), and the tibiae by an average of 6.64 cm (52.9%). Patients gained an average of 16.9 cm in height, including physiological growth. The mean bone-healing index (BHI) was 31.2 days/cm for the femora (range: 17.4 to 43.3 days/cm) and 34.3 days/cm for the tibiae (range: 19.5 to 60.0 days/cm). Complications included 3 delayed maturations, 3 pin track infections, 5 transient fibular paralyses, 5 regenerate fractures, 1 late varus deformity, 1 knee contracture and 1 knee contracture secondary to knee dislocation. Functional scores were excellent in 78 segments, good in 8, fair in 1 and poor in one. There was no growth inhibition related to the lengthening.

Conclusion: Bilateral simultaneous lengthening of the femora and tibiae in achondroplastic patients provided a reduction in total treatment and external fixation time, with a low rate of complications.

Key words: Achondroplastic; femur; simultaneous bilateral lengthening; tibia.

Achondroplastic patients require serial limb lengthening procedures to achieve a height within the normal adult range. Lengthening of the lower extremities has been successful after both chondrodiatasis (distraction through the physeal plate) and callotasis (callus distraction).[1-3] Currently, simultaneous lengthening of both femora or both tibiae (transverse lengthening) is typically performed during different sessions.[1,4]

In this study, we evaluated the results of simultaneous lengthening of both femora and tibiae in achondroplastic patients.

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Patients and methods

This study included 22 patients (16 females and 6 males) diagnosed with achondroplasia who underwent simultaneous bilateral lengthening of the femora and tibiae (88 segments) between 2002 and 2008. Informed consent was obtained from all patients and the Institutional Review Board approved this study. This study is registered in Clinical Trials Registry (ClinicalTrials.gov) with the ID: NCT01328418.

Mean age at the time of the surgery was 6.36 (range: 3 to 11) years. Demographic data of the patients are depicted in Table 1. Orthofix LRS (Ortofix Srl., Bussolengo Verona, Italy) monolateral fixators were used for femoral lengthening, and either Ilizarov-type (Tasarım Med, Istanbul, Turkey) or hexapod-type (Smart Correction; Gotham Medical LLC, Fort Lee, NJ, USA) circular external fixators were used for tibial lengthening.

Hip flexor release and percutaneous lengthening of the Achilles tendon were performed in 15 patients (30 limbs). In addition to short stature, 2 patients also displayed limb-length discrepancy. The rate of callus distraction was ¾ mm per day, which was initiated on the 7th postoperative day. To prevent fracture or plastic deformation of poor regenerate, intramedullary Steinmann pins were introduced percutaneously during the fixator removal session in 4 patients (Fig. 1).

Standing X-rays of the lower extremities and photographic documentation were obtained for all patients prior to surgery and at the end of treatment (Figs. 2 and 3). The length of each segment was measured and recorded, and the malalignment test was performed to check for the presence of an associated deformity before and after lengthening (Fig. 4).

External fixation time, time for consolidation of the distraction callus and the amount of lengthening were documented. The ratio of lengthening was calculated as the amount of lengthening divided by the length of the segment. The lower extremity joints adjacent to the lengthened segments, namely the hip, knee and ankle joints, were examined for range of motion and stability. Clinical results were evaluated according to the functional scoring system described by Paley.[5] Complications were classified according to Paley's criteria; (a problem is resolved at the end of the treatment by non-operative means, an obstacle is resolved by operative means, and a sequela remains unresolved at the end of the treatment.[5]

Patients were followed up in the orthopedic department every two weeks until the end of the lengthening period and monthly until fixator removal. X-rays were obtained to evaluate regenerate bone formation and the state of the physeal growth plates. All patients were followed up in the pediatric department to monitor growth and development and to measure and record growth parameters.

Results

Mean follow-up time was 35 (range: 26 to 76) months. Mean amount of femoral lengthening was 7.07 (range: 4 to 11) cm and the mean amount of tibial lengthening was 6.64 (range: 3 to 11) cm. The mean lengthening ratio with respect to the original segment was 46.1% in the femora (range: 23.3 to 74.0%) and 52.9% in the tibiae (range: 22.5 to 98.0%). The mean bone-healing index (BHI) was 31.2 days/cm for the femora (range: 17.4 to 43.3 days/cm) and 34.3 days/cm for the tibiae (range: 19.5 to 60.0 days/cm). The mean lengthening per patient for one session was 16.9 (range: 9 to 25) cm and included normal growth during the lengthening period. Two patients with limb-length discrepancy (LLD)

Fig. 1. Application of intramedullary rods to both femora and tibiae to prevent fracture.

Table 1. Demographic and outcome data of the patients.

Pati. no.	Age (years)	Gend.	Height (cm)		Amount of lengthening (cm)				Ratio of lengthening (% Original length)				External Fixation Index (days/cm)				Complications
			Before leng-thening	After leng-thening	Femora		Tibiae		Femora		Tibiae		Femora		Tibiae		
					Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	Right	Left	
1	6	Female	91	110	8.0	7.5	7.0	6.2	46.1	43.3	52.6	46.6	24.5	28.0	28.0	34.6	L tibia delayed maturation, knee dislocation, knee contracture
2	8	Female	100	110	6.0	5.0	4.0	4.5	28.3	23.3	23.5	26.4	43.0	51.6	64.5	57.3	Regenerate fracture, late varus deformity
3	4	Male	85	101	5.5	5.0	6.6	7.0	42.3	39.2	54.0	56.0	35.6	39.2	26.5	28.0	L transient peroneal palsy
4	9	Female	110	125	6.5	6.5	6.5	6.5	26.5	26.0	28.3	28.8	57.6	27.3	39.2	39.2	Regenerate fracture, R tibia delayed maturation
5	3	Female	77	96	7.0	7.0	7.0	7.0	51.8	51.8	60.8	51.1	30.4	30.4	30.4	30.5	
6	4	Female	83	92	4.0	4.0	3.0	3.0	27.5	25.0	25.0	25.0	33.7	33.7	45.0	45.0	Pin track infection
7	11	Male	105	116	5.0	5.0	6.5	6.0	24.5	24.2	40.3	36.8	36.6	36.6	28.1	30.5	
8	3	Female	75	95	7.0	7.0	7.0	7.0	56.0	56.0	63.6	63.6	31.2	31.2	31.2	31.5	
9	3	Male	74	94	7.0	7.0	7.3	7.3	58.3	58.3	69.5	69.5	30.0	30.0	28.7	28.5	
10	11	Female	109	124	7.0	7.0	7.5	7.5	29.1	29.1	43.6	43.6	45.0	32.1	55.3	42.0	R tibia delayed maturation, L transient peroneal palsy
11	3	Female	80	99	7.5	7.5	9.0	9.0	51.8	55.3	58.3	66.6	26.0	25.2	26.0	22.5	L transient peroneal palsy
12	5	Female	82	104	8.0	8.0	7.0	7.0	53.3	53.3	58.3	58.3	34.6	34.6	39.5	39.5	
13	8	Male	97	101	8.0	11.0	6.0	7.5	46.5	70.9	35.7	52.3	39.0	28.3	52.0	41.6	Pin track infection, regenerate fracture
14	7	Female	91	107	7.6	7.6	6.4	6.4	52.4	52.4	51.2	51.2	43.2	43.2	51.4	51.4	Regenerate fracture
15	8	Male	96	110	7.2	7.2	5.0	5.0	43.6	44.7	37.0	37.0	42.7	42.7	61.6	61.6	Pin track infection, L transient peroneal palsy
16	4	Female	77	94	8.0	8.0	7.3	7.3	59.2	60.2	66.3	66.3	26.2	26.2	28.7	28.7	
17	4	Male	89	114	6.0	7.5	5.0	6.0	37.0	56.3	43.4	54.5	35.0	28.0	42.0	35.0	Regenerate fracture
18	11	Female	102	116	7.0	7.0	7.0	7.0	31.8	32.5	42.4	42.9	30.0	30.0	30.0	30.0	
19	4	Female	81	95	7.0	7.0	7.0	7.0	51.8	51.8	51.8	60.8	30.0	30.0	30.0	30.0	
20	8	Female	90	106	7.0	7.0	7.0	7.0	58.3	58.3	63.6	63.6	30.0	30.0	30.0	30.0	
21	7	Female	95	126	10.0	10.0	9.5	9.5	74.0	74.0	82.6	82.6	22.5	22.5	23.6	23.6	L transient peroneal palsy
22	9	Female	96	116	8.7	9.0	11.0	10.5	64.0	62.0	98.0	97.5	30.0	30.0	30.0	30.0	

Pati.: Patient; Gend.: Gender.

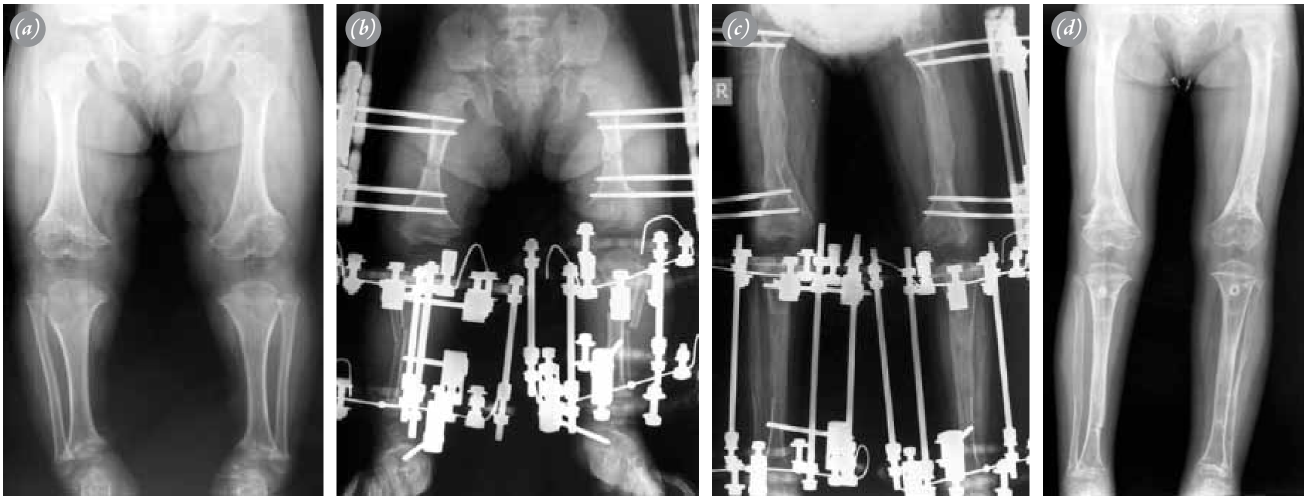


Fig. 2. X-rays showing the treatment sequence: **(a)** Preoperative standing orthoroentgenogram, **(b)** orthoroentgenogram after the surgical treatment, **(c)** orthoroentgenogram after lengthening, and **(d)** orthoroentgenogram following removal of the external fixator.

achieved equal limb lengths at the end of the treatment (LLD was 4.5 cm in one patient and 3 cm in the other).

Nineteen complications were observed; 3 segments with delayed maturation of the regenerate callus, 3 pin track infections which resolved after antibiotic therapy and local wound care, 5 transient fibular paralyses occurring immediately after surgery but diminishing spontaneously, 5 segments with fracture of the regenerate callus treated with cast application, 1 late varus deformity, 1 knee contracture, and 1 knee contracture secondary to knee dislocation.

Functional scores were excellent in 78 segments, good in 8, fair in 1 and poor in one.

Discussion

External fixation is not comfortable for patients, particularly for achondroplastic patients due to the smaller size of their extremities. Thus, because of the multiple required lengthening sessions, external fixation time is an important issue for such patients. As we performed simultaneous bilateral femoral and tibial lengthening, the total external fixation time was reduced compared

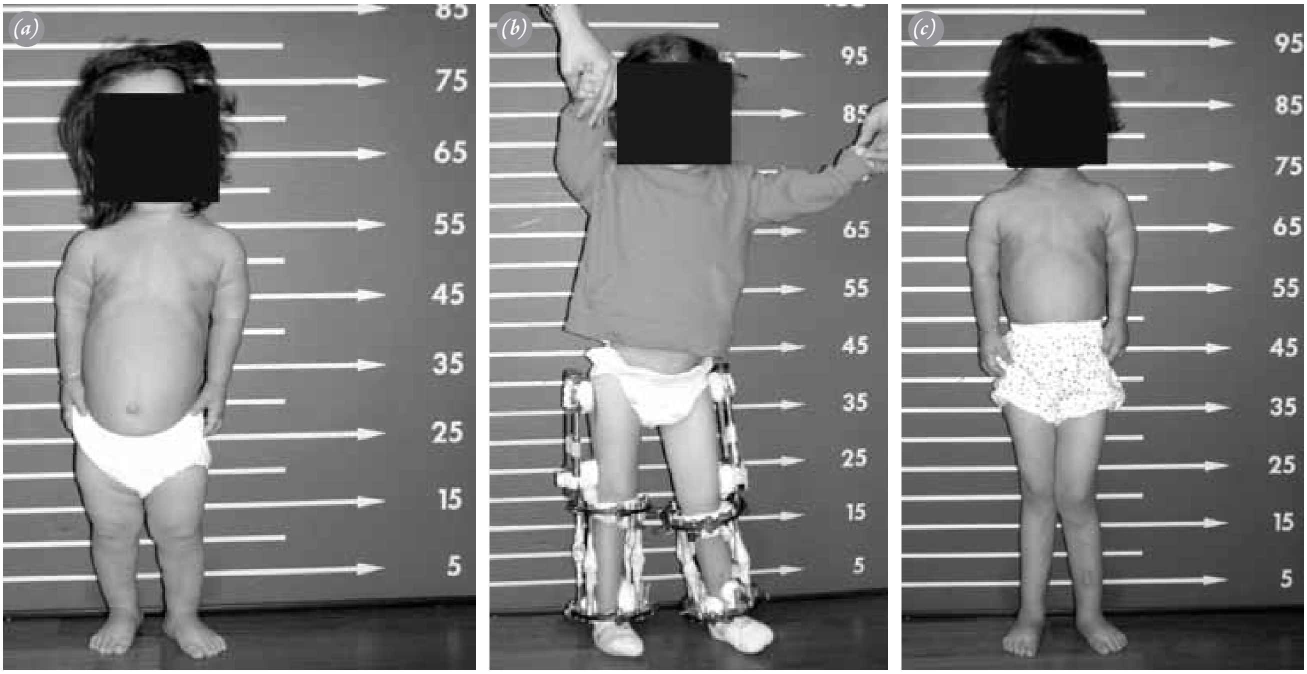
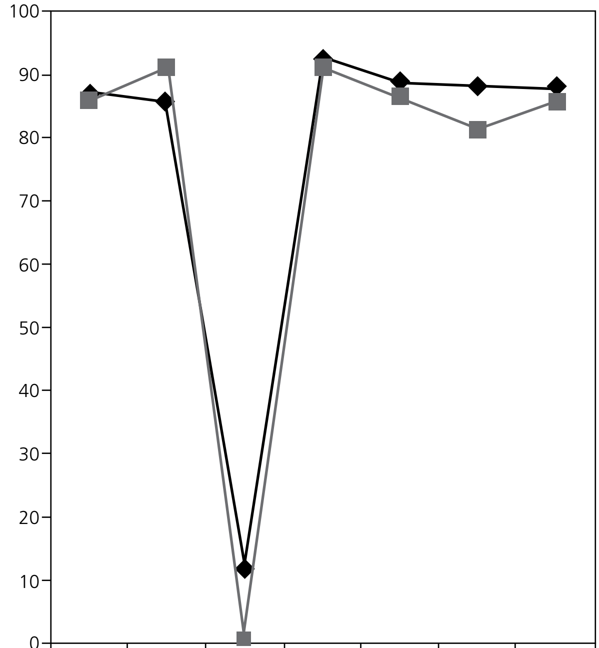


Fig. 3. Photographs of a patient **(a)** before, **(b)** during, and **(c)** at the end of lengthening.



	LDFA-a	MPTA	MAD (mm)	LDFA-m	ADTA-a	PPTA-a	PDFA
Mean preoperative alignment parameters	87	86	11.5	92.5	88.7	88.1	87.8
Mean postoperative aligment parameters	86	91	0.6	91.2	86.6	81.33	85.77

Fig. 4. Preoperative and postoperative alignment parameters for patients. ADTAa: Anatomic Anterior Distal Tibial Angle; LDFAa: Anatomic Lateral Distal Femoral Angle; LDFAm: Mechanic Lateral Distal Femoral Angle; MAD: Mechanic Axis Deviation; MPTA: Medial Proximal Tibial Angle; PDFA: Posterior Distal Femoral Angle; PPTA: Posterior Proximal Tibial Angle.

to consecutive lengthening of the tibiae and femora (in which case the external fixation times will be doubled). In our series, the mean external fixation time was 233 days for the tibiae and 228 days for the femora. Total external fixation time for separate femora and tibiae fixation would have increased by 228 days in our patients (233+228=461 days).

The rate of callotasis should not exceed ¾ mm per day due to the potential risk of the formation of poor quality bone in these patients[2,6] and to prevent traction injury to the nerves due to the significant amount of lengthening related to the simultaneous lengthening of both the femora and tibiae. In three segments, we reduced the rate of distraction further due to poor regenerate bone formation.

In our series, the mean BHI was 31.2 days/cm for the femora and 34.3 days/cm for the tibiae. Aldegheri and Dall'Oca reported healing indices of 40.76 days/cm for the femur and 42.05 days/cm for the tibia,[3] and Vaidya et al. reported a lower healing index of 26.06 days/cm for the tibia.[7]

There were 5 regenerate fractures (5.68%) in our series, which were treated non-operatively with cast application. Venkatesh et al. reported a 15% rate of regenerate fractures in their series.[8] The lower incidence of regenerate fractures in the current study was due to the lower distraction rate of ¾ mm per day (versus 1 mm per day), which yielded a higher quality of regenerate bone, and the use of prophylactic intramedullary Steinmann pins during fixator removal in patients with poor regenerate formation.

It has been reported that patients younger than 8 years old do not cooperate well with physical therapy during lengthening.[3,4] The mean age of our patients was 6.36 years. Fifteen patients underwent prophylactic hip flexor release and Achilles tendon lengthening. Permanent knee contracture due to knee dislocation was encountered in only one patient. The low rate of joint-related complications can be attributed to the prophylactic tenotomies and general ligamentous laxity in achondroplastic patients.[8]

The difficulty for patients in undergoing simultaneous osteotomy of four long bones is the main criticism of this procedure. However, simultaneous osteotomy of the four lower-extremity segments was well tolerated in our patients. This was achieved through pain control and early mobilization. The operative time was between 3 and 4 hours per patient.

It has been reported that simultaneous femoral and tibial lengthening, or two tibial lengthenings in close succession, can lead to tibial growth inhibition.[9] Lee et al. documented that tibial lengthening of more than 40% in young female rabbits adversely affected the proximal tibial growth plate architecture.[10] These negative effects on physiological longitudinal growth of the tibia can be prevented by Achilles tenotomy.[11] In our series, 15 of 22 patients underwent tenotomies of the hip flexors and the Achilles tendon. Furthermore, we believe that the generalized ligamentous laxity in achondroplastic patients also protected the physeal plates from excessive compressive loading related to lengthening. Similarly, Noonan et al. reported more joint-related complications in patients with congenital shortening than in achondroplastic patients.[12] Song et al.[13] reported 23 achondroplasia patients who underwent tibial lengthening and were followed for physeal damage or skeletal maturity. They concluded that physeal damage occurs after limb lengthening by over 50% in achondroplasia. This damage is a gradual process that manifests itself about 2 years after surgery. However, the number of patients in the current study was small and determination of growth arrest was performed only radiologically without any clinical correlation. A prospective study on the growth curve of the tibia and femur in 'normal' patients with achondroplasia in the Turkish population and follow-up of our patients for growth arrest is necessary to draw a limit age for lengthening in achondroplasia patients.

The gastrocnemius-soleus-Achilles tendon complex is a biarticular muscle at risk during tibial lengthening.[14] Many options have been described to prevent ankle equinus contracture during lengthening. Preoperative gastrocnemius-soleus-Achilles tendon complex stretching exercises and intraoperative pin fixation of the tendon complex can be used. Pin fixation technique should be used in high risk patients (congenital shortenings, neurovascular disease and amount of lengthening more than 20% of the bone segment length).[5] According to the growth arrest theory, the generalized ligamentous laxity in achondroplastic patients may also protect the gastrocnemius-soleus-Achilles tendon complex during lengthening. Nevertheless, in our study, the lengthening ratio to the original bones was much higher than the literature. Therefore, we chose to perform tenotomy of the Achilles tendon. Belthur et al. also reported the details of a new surgical technique using an extra-articular calcaneotibial screw to prevent ankle equinus.[15] However, this technique may increase compressive loads on the articular cartilage and the growth plate during lengthening. In addition, this technique was used in a relatively small group (14 limbs in 10 patients) which may not have been large enough to reflect other possible complications such as flexor hallucis longus tendon or neurovascular complications. Furthermore, a second operation is needed for removal of the calcaneotibial screw.

In conclusion, simultaneous lengthening of both femora and tibiae in achondroplastic patients reduces the total treatment and external fixation time.

Conflicts of Interest: No conflicts declared.

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