Fixator-Assisted Lengthening and Deformity Correction Over an Intramedullary Nail in a Patient with Achondroplasia

Akondroplazili Hastada Eksternal Fiksatör ile Çivi Üzerinden Uzatma ve Deformite Düzeltme

Lengthening and Deformity Correction in a Patient with Achondroplasia

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Özet

Akondroplazi orantısız boy kısalığının en sık görüldüğü rizomelik kısalık formudur. Hastalar günlük yaşamda büyük oranda fiziksel ve psikolojik sorunlarla karşılaşırlar. Akondroplazili hastalarda boy uzatma ve deformite düzeltme birçok farklı metodla yapılabilir. Bu vaka sunumunda, 17 yaşındaki akondroplazili bayan hastada iki taraflı alt ekstremite uzatma ve iki taraflı tibial varus deformitesi düzeltme tedavisi sunulmaktadır.

Anahtar Kelimeler

Akondroplazi; Deformite; Kısalık; Düzeltme; Uzatma

Abstract

Achondroplasia is the most frequently encountered form of nonlethal skeletal dysplasia and a type of rhizomelic dwarfism. It results in considerable physical and psychologic handicaps owing to the disproportionate stature of the body and difficulty in performing routine activities of daily living. They also have major musculoskeletal problems including symptomatic malalignment of the lower limbs. Limb lengthening has been used in patients with achondroplasia by different techniques (Intramedullar nailing, monolateral or circular external fixator). We report our treatment of a patient 17 years of age with achondroplasia for bilateral lower limb length discrepancy and bilateral tibial varus deformity.

Keywords

Achondroplasia, deformity, shortness, correction, leghthening

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Introduction

Achondroplastic dwarfism results in considerable physical and psychologic handicaps and difficulty in performing routine activities of daily living [1]. These individuals often suffer from emotional disturbances, and are prone to have inferiority complexes [1]. They also have symptomatic malalignment of the lower limbs. Some patients eventually undergo serious surgery for correction of their malalignment to decrease pain and prevent early-onset degenerative arthritis [2,3]. Lower limbs can be lengthened with chondrodiastasis (the growth plate distraction) and callotasis (the callus distraction) successfully [4]. Nowadays in common practice both the tibia and femur are simultaneously lengthened in different sessions [6]. In this study, We report the result of using a combination of fixator-assisted nailing with lengthening over an intramedullary nail in a 17 year old patient with achondroplasia with tibial deformity and shortening.

Case Report

17 years old patient referred for leg lengthening for cosmetic purposes and difficulty of walking due to bilateral tibial varus deformity. Anterior posterior and lateral radiography of both legs were taken to determine the calf length, extremity length, the appropriate nail characteristics and the tibial varus angle. The varus deformity was 18° for both tibias and extremty length for both sides was 59 cm. At the initial radiologic examination there were Harris Muller plates in both proksimal femurs from an operation that she had at two years of age (Figure 1). We notified the patient and her family about the complete procedure, the possible complications as well as the duration of the process. Informed consent was obtained from the patient and her family. We planed to operate extremities as a cross in different sessions. Firstly we removed both plates and lengthen the right femur with intramedullar nailing with fixator assisted method; and the contralateral tibia with the same method correcting the tibial varus deformity (Figure 2). For the right femur we used intrammedullar nail (IMN) and because of the small calf of the tibia two of 3 mm IM elastic nails were used for tibial fixation (Figure 2). Under general anestesia, on supin position; from the proximal enterence the right femur was reamed of the intramedullary canal over an intramedullary nail (of the diameter of 10 mm, 260 mm length). Then osteotomy for femur was made from the subtrochanteric region and maximum possible size of nail was performed regarding the length of femur. After locking the femoral nail distally, monolateral external fixator was used; two pins were inserted proximally and three for distally. For the tibial deformity of contralateral side, after inserting two intramedullary elastic nails of 3 mm to the canal, acut correction of tibial varus deformity was performed with tibial opened wedge osteotomy and fibular osteotomy performed through an incision of 1 cm, the procedure took approximately 2.5 hours. The conditions of the subjects were assessed during the first visit. The lengthening process was initiated after 5–7 days at a rate of 2 mm/day at the first week followed by 0.25 mm every 6h later. Simultaneously, the physical therapy was started to stretch and empower hip abductors, quadriceps femoris, calf muscles and Achilles tendon. Postoperatively when pain and swelling were reduced the patient was mobilized and after three weeks load bearing is allowed for both sides. The bone regeneration and deformity correction was usually assessed every two weeks by taking X-ray images. The rate of lengthening was adjusted to ensure the bone formation. The subjects were carefully investigated for possible complications. Once the desired length was achieved for both bones, one screw were inserted into the medullary nail at the proximal side of the femur, and the external fixators were both removed almost six weeks after the operation. The intramedullary nail supported the bone during the consolidation phase and allowed the removal of external fixator after the distraction phase of lengthening. Cefazolin (1 g/TDS) was administered during hospitalization, 7–10 days oral antibiotic was also prescribed. During the lengthening, in case of clinical diagnosis of infection (including pin-tract infection, osteomyelitis), appropriate antibiotic was administered. NSAIDs were prescribed for pain management as needed. The patient was followed up for six months so that their range of motion,



Figure 1. At the initial radiologic examination there were Harris Muller plates in both proksimal femurs from an operation that she had at two years of age.

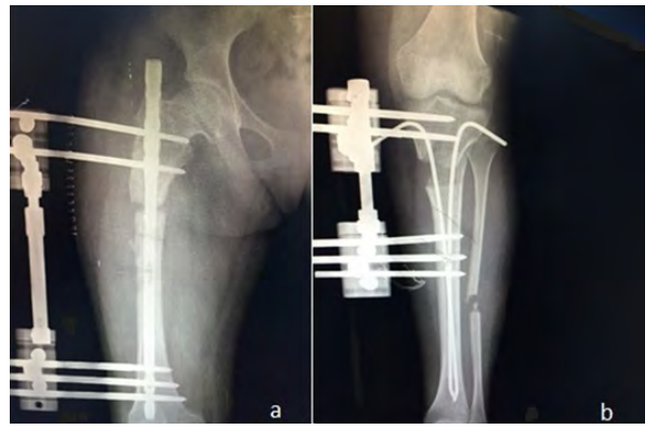


Figure 2. (a) After removing both plates right femur was leghthened with external fixator over an IM nail; (b) and the contralateral tibia with the same method correcting the tibial varus deformity. Because of the small calf of the tibia two of 3 mm IM elastic nails were used for tibial fixation.

level of pain and regenerated bone quality could be evaluated. Six mounts after the first operation we performed the same procedure for the left femur and the controlateral tibia. After one year follow up the patient was free of pain and had full range of motion for both lower extremities. The final results of the patient after removal of the lengthening device showed straight, re-aligned legs (Figure 3). Thus, feet are notably better positioned than prior to the correction, long-leg standing radiograph of the frontal plane mechanical axis showed optimal limb alignment and satisfactory length (5 cm for both) for the patient. There were no complications for the hole prosedure.



Figure 3. The final results of the patient after removal of the lengthening device showed straight, re-aligned legs with satisfactory length for the patient.

Discussion

Achondroplasia is the most common condition associated with disproportionate short stature. Surgical treatment to re-align the lower limbs in achondroplasia is generally indicated for cases who present either a severe, cosmetically unacceptable or clinically symptomatic limb deformity. Varus malalignment is generally more common than genu valga in this syndrome. Realignment can generally be achieved by gradual correction using external fixation devices [6]. Surgical correction is needed for tibial deformities higher than 10 degrees at coranal plane in adolescent with unexpected remodeling potential and adults [7]. Because of the the small size of the extremities especially in terms of patients with achondroplasia, external fixation itself can create discomfort. Because this application must be performed in multiple sessions, It is important for patients how much time they spend with external fixator. We performed simultaneous femoral and contralateral tibial lenghtening with correction of the tibial deformity. Thus we have shortened the duration of external fixation with fixator-assisted lengthening over an intramedullary nail. With earlier removal of the external fixator, the complication rates related to the pins, including pin tract infections and joint stiffness, are substantially diminished [8].

In conclusion fixator-assisted lengthening and deformity correction over an intramedullary nail in patients with achondroplasia is thought to be an appropriate and successful method with patients' high satisfaction.

Competing interests

The authors declare that they have no competing interests.

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