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Preservation of the foot in a five-year-old child with bilateral congenital deficiency of the tibia

İki taraflı doğuştan tibia gelişme eksikliği olan 5 yaşındaki bir çocukta ayağı koruyucu yaklaşım

Hasan BOMBACI, Mucahit GORGEC, Serhat YANIK

Haydarpasa Numune Education and Research Hospital, 1st Orthopaedics and Traumatology Department

Doğuştan tibial gelişme eksikliği alt ekstremitenin kısalık ve varusuna neden olan nadir bir anomalidir. Son zamanlarda gelişen ekstremite uzatma ve ayak deformitesi düzeltme teknikleri ayağı koruyarak fonksiyonel sonuçlar almak konusunda cesaret vericidir. Bu yazıda, Jones sınıflamasına göre iki taraflı tip II anormallik saptanan ve Syme amputasyonu olmaksızın tibia-fibular füzyon ile tedavi edilen beş yaşındaki erkek çocuk sunuldu. Hasta, dizlerinin üzerinde ancak ellerinin yardımıyla hareket edebiliyordu. Radyografik incelemede, fibulanın tam gelişmiş, distale doğru kalınlaşmış olduğu; tibianın ise yalnızca proksimal kısmının var olduğu, distal kısmının olmadığı görüldü. Ayak ve ayak bileği ekleminde sert ekinovarus deformitesi vardı. Seri ameliyatlarla her iki bacakta tibia-fibular füzyon ve fibular osteotomi, sağ ayağa subtalar artrodez ve metatarsal osteotomi, sol ayağa subtalar artrodez yapıldı. Altı yıllık tedavi ve takip süreci sonunda çocuk aksayarak da olsa yürüyebiliyordu.

Anahtar sözcükler: Fibula/transplantasyon; ayak deformitesi, doğuştan/cerrahi; tibia/anormallik/cerrahi.

Congenital tibial deficiency is a rare anomaly causing shortening and varus deformity of the lower extremity. Recent limb lengthening and foot repositioning techniques enable functional results with preservation of the foot. We present a five-year-old boy with bilateral congenital tibial deficiency of type II according to the Jones classification, who was treated with tibia-fibular fusion without Syme amputation. His ambulation depended on crawling. Radiographic examination showed a normally developed fibula with thickening in the distal portion, and only proximal tibia with absence of the distal part. He also had bilateral stiff equinovarus deformity. In a series of operations, he underwent bilateral tibia-fibular fusion and fibular osteotomy, subtalar arthrodesis and metatarsal osteotomy in the right foot, and subtalar arthrodesis in the left foot. At the end of a six-year treatment and follow-up, walking was achieved despite some degree of limping.

Key words: Fibula/transplantation; foot deformities, congenital/surgery; tibia/abnormalities/surgery.

Congenital deficiency of the tibia (CDT) is among the rarely seen deformities. The current treatment modality for cases with type I b and II (according to Jones' classification) with deficiency of tibial distal part, is fusion of the distal part of the fibula to the present proximal part of tibia. Performing a Syme amputation in the foot provides a lower extremity without a foot but eligible for prosthesis.[1-5] Although this treatment has been seen as successful in terms of function, it is not considered as a positive choice from family front of view due to loss of an extremity and the child leads his entire life dependent to prosthesis. Especially in bilateral cases, short-range mobilization of the child, presents severe difficulties. Lately, the recent developments in extremity lengthening procedures enable us more frequently applying other methods than amputation for such deformities. In the present case, in order to

Correspondence to: Dr. Hasan Bombacı. Haydarpasa Numune Education and Research Hospital, 1st Orthopedics and Traumatology Department,Turkey
Phone: +90 216 - 414 45 02 / 1592 Fax: +90 216 - 345 61 81 e-mail: bombacih@hotmail.com

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prevent social, functional, and psychological problems to take place, the treatment modality, which preserves the foot, was preferred.

As a result of our search in the related literature written in English and Turkish, we found no cases in which deformities of type II CDT were treated with an approach that included preservation of the foot. In the present study, such a case is presented with a follow-up of 6 years.

Case report

Bilateral type II CDT (according to Jones' classification) was determined in the 5-year-old boy presented to our hospital in 1999. Achilles lengthening and medial release had been done in another hospital. The child was able to walk only on his knees with the help of his hands and the skin area located in the anterior part of his knee was thickened considerably. Hamstring and quadriceps functions were nearly complete and he was able to perform knee flexion and extension. In both knees, fibular head was palpable and displaced proximally and laterally. Both tibias were short and the legs were in varus position. Rigid equinovarus deformity was present in the foot and ankle joint. In the radiographical examination, while fibula was found to be completely developed and thicker than normal distally, only proximal part of the tibia was present and distal part was missing. In both of the ankles, fibula was articulating with talus with a large surface.

First, in order to form fusion between distal half of fibula and tibia, fibular osteotomy and tibio-fibular fusion operations have been done (Figure 1a). Five months later, posteromedial release was done to the right foot and ankle. Centralizing of the talus was performed beneath the fibula through an osteotomy performed on the lower end of fibula. One year after the first operation, fusion procedure was performed also on proximal tibia and distal fibula in the left lower limb (Figure 1b). Meanwhile, the contra lateral fusion line was fused. And the child was able to walk by giving his weight to this side. One year after the last operation, posteromedial release was performed on the left foot and ankle. In addition a corrective osteotomy performed on lower end of the fibula in order to correct the present deformation and centralizing the talus under fibula. Following either the tibio-fibular fusion or fibular osteotomies performed on both of the legs, even in cases that osteotomy has not been completely appropriate, no fusion problems were experienced (Figure 1a, b). The proximal parts of fibula were not removed in both of the operations. Seven months after the last operation, subtalar arthrodesis and metatarsal osteotomy were performed for rigid equinovarus deformities of the right foot. Seven months following this operation, subtalar arthrodesis was performed on left ankle. In order to avoid any risk that would compromise the circulation of the foot, metatarsal osteotomy was not performed on in the same session. In order to preserve the growth potential of the leg, fibula was chosen as the operation site of the region above ankle and subtalar region was preferred for the procedures below this level, to correct the deformities.

In the final follow-up, varus deformity was present in both feet but smaller in the side which metatarsal osteotomy has been applied. The child was able stand on the lateral margin of his foot. He was complaining of a pain in his 5th metatarsal basis when stayed on his foot for a long period. The knee range of motion (ROM) was nearly complete, ankle was immobile but stable and painless (Figure 2a, b, 3a, b). Both feet had metatarsus adductus and varus deformity, but especially on the left side on which metatarsal osteotomy has not been performed, they were more advanced (Figure 4a, b). There was two

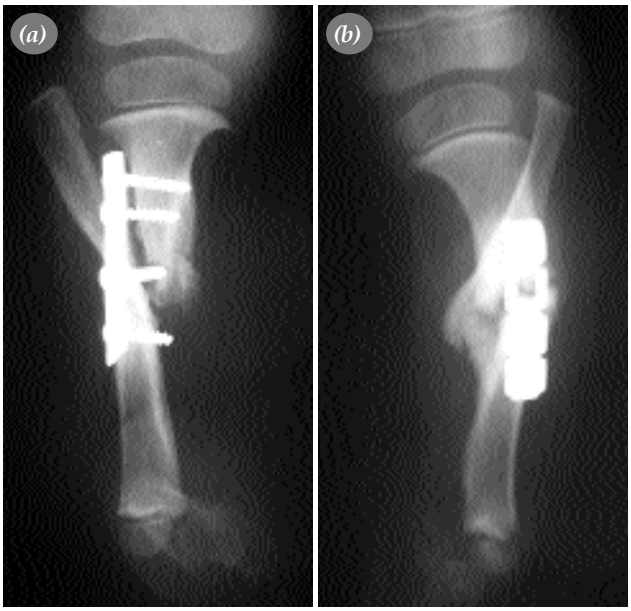

Figure 1. AP radiograph of right **(a)** and left **(b)** limb just after the operation of tibio-fibular fusion.

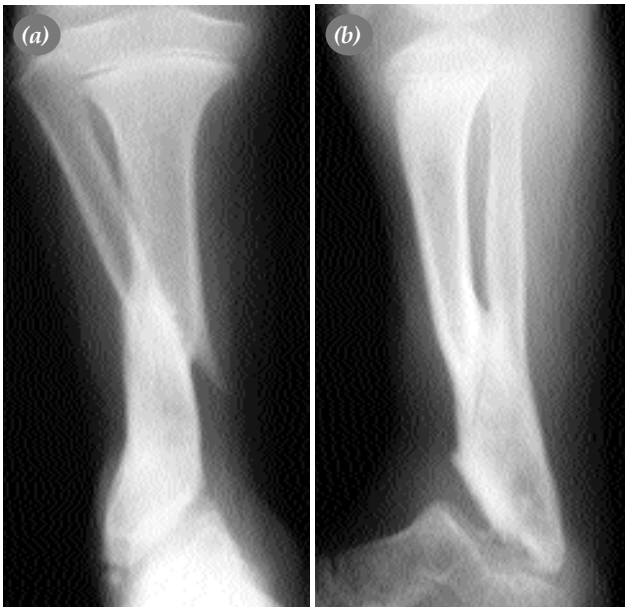

Figure 2. AP **(a)** and lateral **(b)** view of the right lower limb six years after first operation. There is complete union between tibia and fibula, fibula has been enlarged considerably.

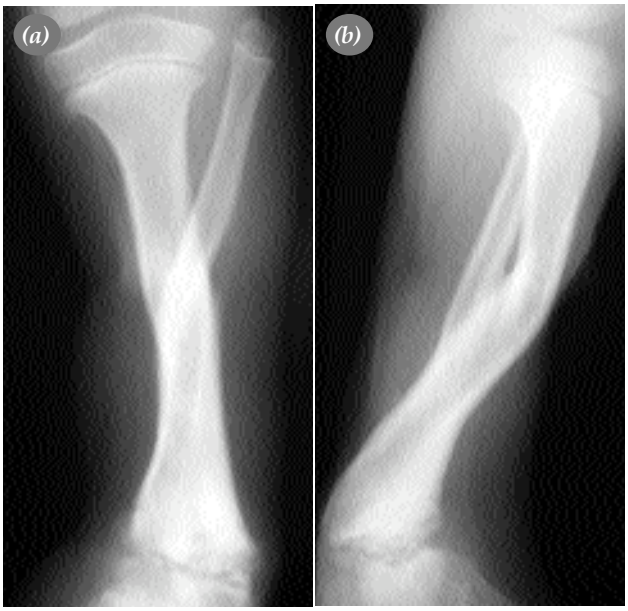

Figure 3. AP **(a)** and lateral **(b)** view of the left side at the last follow up. Radiographic findings resembling right side are seen.

centimetres of shortness in the right lower limb. The child was able to walk by himself although limping. A corrective procedure was planned for the metatarsus adductus and varus deformity in the feet.

Discussion

Knee disarticulation and Brown operation are some of the treatment modalities which have varying success rates in rarely seen and sometimes bilateral CDT.[6-9] If proximal part of the tibia is present,

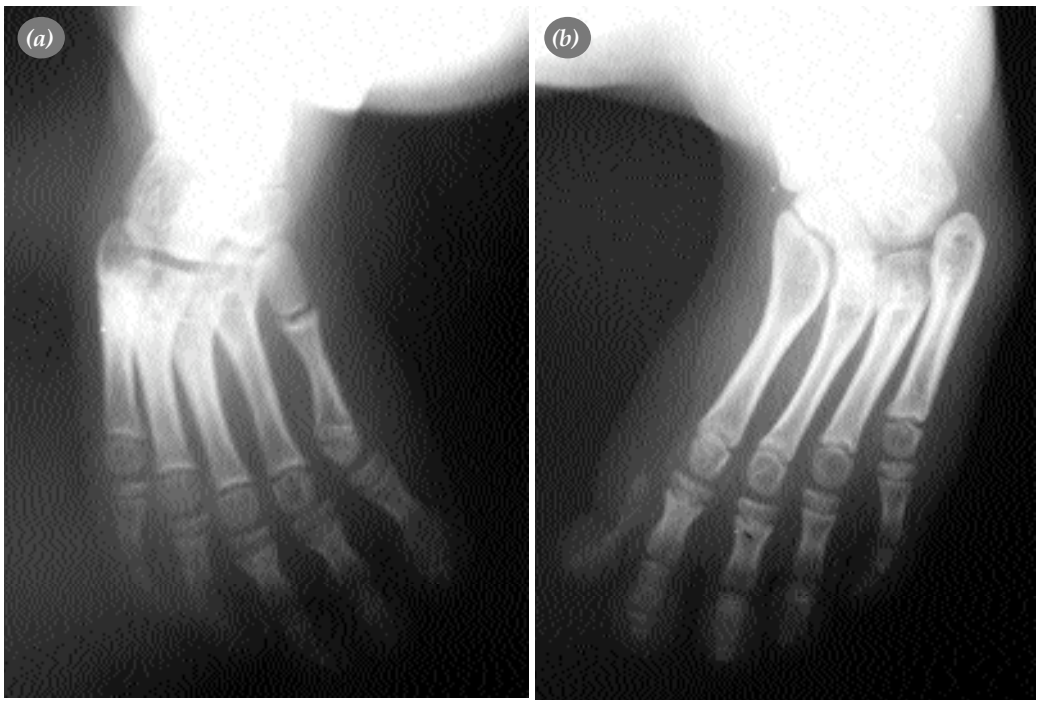

Figure 4. AP views of the right **(a)** and left **(b)** foot. There appear hypogenesis on the first ray of the left foot in addition to the metatarsus adductus deformity which involve both feet.

(Type I B and II according to the Jones classification), fusion between the distal part of fibula and the present part of the tibia and Syme amputation have resulted in satisfactory outcomes.[2,3] However, because flexion contracture is an frequently expected deformity in cases with quadriceps deficiency, disarticulation at the knee level is recommended more often.[7,8] Moreover, families could reject to the idea of amputation sometimes. If the abnormality is bilateral, the preservation of the foot bears more importance.[4] One of the amputation indications in CDT cases is, extensive lower extremity inequality.[5,10] In the present case, because both extremities had shortness, it was not considered as an indication for amputation. Furthermore, latest successful results in both lengthening procedures and correction of foot deformities encourage us to preserve the foot.

In the present case which had good knee function with a bilateral abnormality, a lower extremity with immobile ankle joint but able to stand as plantigrade feet, was intended. Thus, firstly, the procedures aiming to restore the integrity of the bone between knee and ankle were performed. The following purposes were to enable the foot to step on the ground with its plantar surface and then finally, correct the length inequality. Keeping in the mind that the abnormality was present in both extremities, the length inequality was thought to be corrected by foot elevator, arrest of epiphyseal growth, and applying either lengthening or shortening method.

Contrary to the findings of certain studies in the related literature, no union problem was experienced during tibiofibular fusion procedure of this case.[3] Again, ankle instability which has been frequently reported in type I deformities, was not present in this case and on the contrary, there was a very rigid equinovarus deformity.[5] Fibula and talus were articulating over a very large surface. In order to correct the considerable rigid equinovarus deformity in the foot, a procedure was required to be applied on the bones along with the posteromedial release intervention. For this purpose, a wedge with its base to the lateral side was removed from the subtalar region and correction and arthrodesis were performed. Fibulo-talar joint was left alone to avoid damage to the physeal line and not to increase the already present shortness. However, the inadequate nature of this procedure for complete correction of the deformity was seen during the last controls. The metatarsal osteotomy performed on the right foot, has enabled adductus deformity to be corrected in a considerable amount. In CDT, ankle instability is not a rare finding.[10] In the present case, fibula was articulating with talus over a large surface and the ankle was stable. The stable ankle facilitated the weight bearing of foot.

Upper end of the fibula is recommended to be removed following Syme amputation, for better prosthesis conformity. However, following this procedure, sagittal function of the knee might be affected by the muscular imbalance between the hamstring and quadriceps muscles and lateral stability might be affected by the involvement of fibular collateral ligament in the already instable knee.[5,9] In the present case, fibular upper part was not removed and thus, knee stability was not endangered.

In the follow-up evaluation of the presented case after 6 years, bone integrity between knee and ankle was observed to be achieved, enabling the boy to participate in daily social life, but the equinovarus deformity in the foot was not found to be corrected as much as intended. However, his parents have expressed their satisfaction despite this flaw. Whereas more extensive surgical interventions are more effective for the correction of equinovarus deformity in the foot, they may also increase further the present shortness of the extremity. Another component of this abnormality; lower extremity inequality, might increase in the long-term period despite possible involvement of both extremities; however, this is a problem that should be solved later when required. In conclusion, in order to obtain a functionally satisfying lower extremity in congenital deficiency of the tibia (CDT), surgical interventions which preserve foot, require a series of operations. Correction of the foot and ankle deformities constitutes the most difficult part of this process. On the other hand, Ilizarov technique, which has not been performed in this case, may be considered as an alternative option for correction of the deformities in foot and ankle.

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