Lengthening and deformity correction in vascularized fibular autograft for a patient with Ewing sarcoma

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Abstract
We report the case of a pediatric patient with Ewing’s sarcoma of the tibia treated with vascularized fibular autograft where the resulting limb deformity and leg length discrepancy (LLD) were corrected using Ilizarov external fixator. A 14-year-old girl presented to our outpatient clinic with a deformity of the right proximal and distal tibia and an 11.7 cm of LLD after tumor reconstruction surgery. Deformity correction and limb lengthening were simultaneously performed using double corticotomy on the right proximal and distal tibia. One year postoperatively, the union of the right proximal tibia had progressed, but nonunion was observed at the right distal corticotomy site. To address this, osteosynthesis with tricortical iliac bone allograft was performed after the removal of the Ilizarov external fixator. After 6 months, the union of the distal tibia was confirmed, and the varus deformity of proximal and distal tibia improved. The LLD was also decreased, but the left lower limb was still longer by 3 cm. This report shows that vascularized fibular autografts can potentially be used for the gradual correction of LLD and deformities. However, for the treatment of multiple deformities in bones previously reconstructed with vascularized fibular graft, the possibility of impaired bone forming potential of the fibular graft should be considered.

Keywords
deformity correction, distraction osteogenesis, lengthening, vascularized fibular autograft

Date received: 20 February 2020; Received revised 10 June 2021; accepted: 19 August 2021

Introduction
Malignant bone tumors are often treated with extensive excision; however, these procedures can result in large bone defects. In such cases, various reconstruction methods have been recommended to salvage the limb. These limb salvage methods include tumor prosthetic replacement, free vascularized autologous fibular graft, allogenic bone graft, allograft prosthetic composite, distraction osteogenesis, low heat treated autobone, and irradiated autologous bone graft. Of these, the free vascularized fibular graft was described by Taylor et al. in 1975 and has been used to reconstruct bone defects since then. Fibular bone grafts provide a good bone stock since a large amount of graft bone can be harvested and it contains a strong double cortical bone that can support the internal fixation device. It is known to promote the union of host and graft bone and the revascularization of the allogenic endosteal surface.

However, in pediatric patients (who have open growth plates), the amount of autologous bone that can be harvested is limited and many complications can occur at the donor sites. Vascularized fibular autografts provide adequate mechanical stability, but have been known to have slower
bone formation than iliac autografts. In addition, it may result in leg length discrepancy (LLD) and ankle instability, especially if long segments are taken in pediatric patients.\textsuperscript{15} Although vascularized fibular epiphyseal transfer, which can preserve the growth potential, may be surgical option for the reconstruction of proximal humerus and distal radius,\textsuperscript{16,17} distraction osteogenesis is a well-established procedure used to treat bone defects and LLD in viable bone tissues. However, there have only been a few reports regarding limb lengthening and deformity correction in cases with vascularized fibular autografts.\textsuperscript{18–20}

Therefore, we report a rare case of deformity correction with lengthening (for lower limb deformity and LLD, respectively) in a patient with Ewing’s sarcoma, after surgical resection and reconstruction using vascularized fibular autograft.

**Case report**

A 14-year-old girl presented to our outpatient clinic with deformity of the lower extremity and LLD. At 6 years of age, she underwent wide excision of Ewing’s sarcoma—in the right tibia—and reconstruction using vascularized fibular autograft from both sides; the procedure was performed at another hospital. She received chemotherapy consisting of vincristine, doxorubicin, and cyclophosphamide, alternating with ifosfamide and etoposide (i.e., according to the CCG 7942 protocol) preoperatively and postoperatively.\textsuperscript{21} Later, at 11 years of age, she underwent lateral malleolar epiphysiodesis for right ankle varus deformity.

Measurements done at the outpatient clinic placed her height at 151.7 cm (ninth percentile) and her weight at 60.4 kg (85th percentile). She could walk with crutch support. Physical examination revealed a varus deformity of the right lower extremity, a valgus deformity of left ankle, and LLD. Furthermore, an equinus deformity of around 30° was observed in the right ankle (Figure 1). Both knee joints showed full range of motion. The ankle dorsiflexion angles were −30° and 15°, and the plantarflexion angles were 50° and 35° on the right and left, respectively. Plain radiography confirmed the presence of a varus deformity of the proximal and distal tibia and posterior angulation of distal tibia in the right lower extremity, and a valgus deformity of the distal tibia in the left lower extremity (Figure 2). In the right tibia, the medial proximal tibial angle (MPTA) was decreased to 59°, the lateral distal tibial angle (LDTA) was increased to 117°, and the anterior distal tibial angle (ADTA) was decreased to 52°. For the left tibia, the LDTA was decreased to 52°. The LLD was 11.7 cm and was mainly in the tibia. Her growth plates in the lower extremity were closed at the age of 14 years.

![Figure 1](image-url). Preoperative clinical photographs of the patient.
Our surgical team attempted to solve all the deformities using the minimum number of operations. A supramalleolar closing wedge osteotomy was performed on the distal tibia for the valgus deformity of the left ankle; a locking compression plate was used. In the right lower extremity, tendo-Achilles lengthening was performed for the equinus deformity. Thereafter, we simultaneously performed both deformity correction and lengthening by performing a double corticotomy on the proximal and distal tibia (Figure 3). The corticotomy was performed at the level of the center of rotational angulation (CORA).

An Ilizarov fixator was applied to the right tibia and foot, using four full rings and one-half ring. The first ring was mounted proximally from the proximal tibial corticotomy site parallel to the knee joint line using one 1.8 mm Ilizarov tension wire, one 1.8 mm olive-stopped wire, and one-half pin. The second ring was mounted just distal to the proximal corticotomy site using one-half pin. The third ring was mounted on the middle of the tibia, below the second ring, using one-half pin. The fourth ring was mounted above and parallel to the ankle joint line using one 1.8 mm Ilizarov tension wire and three-half pins. The foot was fixed with half rings, using two 1.8 mm Ilizarov tension wires, in a neutral ankle position. All Ilizarov tension wires were tensioned to 130 Nm. The proximal distraction rod was placed medially, and the distal one was placed anteromedially. The proximal hinge device was placed laterally, and the distal hinge was placed posterolaterally. Corticotomies were performed at the level of the proximal and distal

**Figure 2.** Preoperative radiographs showed leg length discrepancy (11.7 cm), varus deformity of the right proximal tibia, varus deformity and posterior angulation of the right distal tibia, and valgus deformity of left distal tibia.
metaphyses through the neo-tibia from the previous vascularized fibular autograft, using a multiple drill-hole technique.

Gradual correction of the deformities was initiated at the proximal and distal corticotomy sites 2 weeks after the operation. For angular correction, the motor systems on the concave side at the proximal and distal tibia were lengthened at the rate of 2 mm/day. At 6 weeks after the operation, the varus deformity of the tibia was corrected and the hinge device was changed to a simple rod device for lengthening. Union of osteotomy site at left distal tibia was achieved and partial weight-bearing was started. The distraction rate at the corticotomy site was changed to 1 mm/day for lengthening. At 10 weeks after the operation, poor regenerate bone formation was found; thus, the distraction rate of the proximal corticotomy site was changed to 0.5 mm/day. At 16 weeks postoperatively, delayed union of the distal tibia was observed. We attempted the accordion method for 1 month to promote bone union. Finally, the deformity correction and lengthening was achieved after 4 months.

One year postoperatively, the union of the proximal tibia had progressed, but nonunion was observed at the distal corticotomy site (Figure 4). To address this, osteosynthesis with commercially available human tricortical iliac bone allograft and reconstruction plate was performed after the removal of the Ilizarov external fixator. She was immobilized with a long leg cast for 6 weeks. After 6 months, the union of the distal tibia was confirmed, and the varus deformity of the right proximal and distal tibia improved; the LLD was also decreased to only 3 cm (Figure 5).

On the teleradiogram, the varus deformity of the right lower extremity was improved (MPTA: 81° and LDTA: 95°), and posterior angulation of distal tibia was improved.

**Figure 3.** Postoperative radiographs of bilateral lower legs: The Ilizarov external fixator was applied after the corticotomy of the right proximal and distal tibia, and medial closing wedge osteotomy of the left distal tibia was performed.

**Figure 4.** Plain radiograph and coronal CT image of right lower leg at 1 year after surgery showed nonunion in the distal tibia (arrow).
The valgus deformity of the left ankle was also improved (LDTA: 80°) (Figure 5). No complications, including nerve palsy, vascular injury, pin site infection, and joint stiffness, occurred after surgery or during the distraction period; the mean external fixation index was 50.1 day/cm. Finally, she could walk without crutch support. The ankle dorsiflexion angles were 0° and 15°, and the plantarflexion angles were 30° and 35° on the right and left, respectively.

Discussion and conclusion

Bone tumors often require extensive resection, which then results in large bone defects; various methods have been introduced to compensate for these defects. The method of multiple vascularized fibular autografts after extensive resection of bone tumors has been in use since the 1990s. However, there are some disadvantages of donor complications and microsurgical techniques. Shpitzer et al. reported that vascularized fibular autografts may involve many complications at the donor site like postoperative infection, ankle instability, weakened flexion, donor pain, and swelling.

In general, valgus deformities of the ankle after fibular resection or in fibula pseudarthrosis are known to occur in pediatric patients. Ankle instability is also known to occur owing to loss of the inter-osseous membrane secondary to bone harvesting. In our case, the left ankle lost its lateral support owing to the fibular resection, which then may cause a valgus deformity in the ankle. The function of the right distal tibial growth plate was lost during tumor excision, but that of the lateral malleolar growth plate was retained, which may result in a varus deformity in the right ankle. To correct this varus deformity, lateral malleolar epiphysiodesis was performed; but correction was not achieved. To prevent such an ankle deformity, surgeons
should consider performing syndesmosis fusion during the fibular grafting surgery.

Vascularized fibular autografted bone has no growth potential in pediatric patients, resulting in short limbs. Lengthening of the femur or humerus can be performed with good results after vascularized fibular autografting. Ilizarov et al. and Pilge et al. reported the cases of a 13-year-old boy and an 18-year-old girl, respectively, where the humerus was lengthened using a monolateral external fixator following autologous tumor resection and fibular graft transplantation. Han et al. reported limb lengthening in a 10-year-old boy with osteosarcoma who had been treated with free vascularized fibular graft and reconstruction of the femur; using distraction osteogenesis in two staged operations, the total leg length was extended by 13.3 cm.

Various surgical options may be available for our patient, such as lengthening of the normal right femur, shortening of the left lower limb, and acute correction at the distal tibia and gradual correction at the proximal tibia. However, lengthening of the normal right femur or shortening of the left femur could not be considered because these procedures result in knee height asymmetry. Previous studies have suggested that knee height asymmetry less than 4 cm does not lead to cosmetic and functional deficits. A previous study recommended that tibial shortening be limited to 3 cm. Acute correction at the distal tibia and gradual correction at the proximal tibia may prevent nonunion at the distal corticotomy site. However, a previous study recommended bifocal osteotomies for longer lengthening (>5–6 cm) to reduce the external fixation index. The LLD in our patient was approximately 11 cm; thus, we performed bifocal osteotomy of the right tibia.

In the present case, LLD, varus deformity of proximal tibia, and varus deformity and posterior angulation of distal tibia were simultaneously corrected. The right tibia was lengthened by approximately 8.7 cm—the right MPTA increased by 16°, LDTA decreased by 22°, and ADTA increased by 14.1°. The results show that even after extensive bone resection and autologous fibular grafting, the surgical site has enough bone remodeling potential to allow distraction osteogenesis and deformity correction at the same time. We think that simultaneous correction of multiple deformities can be a viable therapeutic option for pediatric bone tumor patients with limb shortening and other deformities following tumor resection.

Osteosynthesis using an allogenic bone graft was performed during the final operation because of the poor bone formation at the corticotomy site in the distal tibia. We believe that excessive deformation correction of the graft may degrade bone formation ability when performing multilevel distraction osteogenesis. Kristiansen and Steen showed that more complications, such as ankle joint contracture and pseudarthrosis in the callostasis site, occurred with bifocal lengthening than with monofocal lengthening. Aarnes et al. found that bifocal lengthening exposed the entire soft tissue to large loads, resulting in increased tissue stiffness and reduced ability to adapt to the increased length. In our case, since the external fixation index was higher than for general cases, a longer fixing period of about 1 year was required. We think that earlier bone graft and cast immobilization may reduce the treatment time in the Ilizarov external fixator if the stability of the callostasis site was confirmed. Therefore, surgeons should consider the possibility of poor regeneration of the bone formation when performing multiple deformity correction with limb lengthening using bifocal callostasis. For complicated cases, such as ours, earlier revision surgery can be considered if delayed healing at the callostasis site is expected.

In conclusion, the vascularized fibular autograft provided bone stock for reconstruction of the bone defect, and it has potential to allow future procedures for the correction of LLD and deformities. However, for the treatment of multiple deformities in bones previously reconstructed with vascularized fibular graft, the possibility of impaired bone forming potential of the fibular graft should be considered.

Declaration of conflicting interests
The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding
The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical approval
This research has been approved by the institutional review board of our hospital. Written informed consent was obtained from the patient and his parents for this study.

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