

Correction of knee and ankle valgus in hereditary multiple exostoses using the Ilizarov apparatus

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Abstract

Background Hereditary multiple exostoses (HME) is a genetic disorder that causes limb deformities due to disturbance at the growth plates.

Materials and methods Six adolescents, with symptomatic valgus deformity at the ankle and knee (seven affected legs) underwent correction procedures using the Ilizarov apparatus. In 5 legs, a bifocal Ilizarov apparatus was used, whereas in 2 legs the use of a monofocal apparatus was sufficient.

Results Correction of the mechanical axis was achieved in all cases, and limb length discrepancy was equalized in the 3 cases that underwent limb elongation. The average knee and ankle corrections were 15° and 18°, respectively. The average time from application to removal of the Ilizarov apparatus was 4.6 months. No major complication occurred.

Conclusions The use of the Ilizarov method in adolescents with HME enables successful simultaneous correction of multiplanar, multifocal complex limb deformities.

Keywords Hereditary multiple exostosis · Ilizarov apparatus · Osteochondroma · Limb deformity

Introduction

Hereditary multiple exostoses (HME) is a dominantly inherited genetic disorder, caused by point mutations in the exostosin gene family and characterized by multiple benign cartilage-capped tumors, primarily on long bones [1]. HME has a wide spectrum of clinical manifestations, the most common presentation being exostosis in the lower limbs, which is expressed by pain, disruption of osseous growth, bowing of long bones, limb length discrepancy, genu valgum, short fibula, valgus ankle, coxa valga and acetabular dysplasia [2, 3]. Various methods have been described to correct the valgus deformity at the ankle [4–6] but there is only one case report on the use of the Ilizarov method [7].

We present a series of six adolescents (seven affected legs) with HME, who were treated for valgus deformities of the knee and ankle using the Ilizarov apparatus.

Materials and methods

Six adolescents (seven affected legs) with an average age of 14.3 years (range, 10–17.5), who had valgus deformity as a result of osteochondromas at the knee and ankle (Fig. 1), underwent surgical correction using the Ilizarov method (Table 1). The average knee valgus was 13.7° (range, 9°–18°) and the average ankle valgus was 18.4° (range, 10°–30°). Preoperative planning based on long standing radiographs included a malalignment test [8], center of rotation and angulation (CORA) definition, and assembling of the Ilizarov apparatus.

In two of the seven legs, monofocal correction was sufficient to achieve mechanical alignment of the lower limb. In one of these cases the knee valgus (9°) did not

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Table 1 Patients' data, procedures and outcomes.

Patient	Age at operation (years)	Side	Knee valgus (°)		Ankle valgus (°)		Pre-op limb length discrepancy	Correction period (months)	Follow-up (years)
			Pre-op	Post-op	Pre-op	Post-op			
1	10	R	18	0	20	0	None	5.0	3
2	17.5	L	12	0	20	0	2.5 cm	4.5	8
3	15	L	25	5	15	0	2.5 cm	6.5	10
4	15	R	14	2	30	0	2.5 cm	4.0	6
5	12	R	9	NA	20	0	None	3.5	4
6	15	R	18	2	14	+3 ^a	None	4.5	3
7	16	L	15	+5 ^a	10	NA	None	4.5	2
Mean	14.3		17	13.8	18.4	20.3		4.6	5.5

^a Over-correction into varus

NA not applicable (not corrected)



Fig. 1 Anteroposterior radiograph of the lower legs of case 3. There is valgus of 15° at the ankle, and 10° at the proximal tibia, mainly due to osteochondroma of the fibula

necessitate an osteotomy and the monofocal Ilizarov system was applied following distal osteotomy. In the other case the valgus deformity was manifested only at the knee; therefore, the correction was restricted to this segment only. In the other five legs, a bifocal Ilizarov procedure was performed. The ankle correction was done acutely, while the knee deformity was corrected progressively (Fig. 2).

In three cases there was a limb length discrepancy of 2.5 cm due to a short tibia, which was elongated in addition to the correction of the angular deformities. The average follow-up period was 5.5 years (range, 2–10 years).



Fig. 2 Anteroposterior radiograph of left leg of case 3, in Ilizarov frame. Acute correction at the supramalleolar osteotomy, while progressive correction was done at the proximal tibial corticotomy

Surgical technique

Preoperative planning was done based on long standing radiographs, in which the mechanical axis of the limb (femur and tibia) was measured. Assembling the Ilizarov apparatus was done preoperatively based on these measurements. The operation was performed under a combination of general and epidural anesthesia and epidural anesthesia was continued for 4–5 days to control postoperative pain.

The apparatus was constructed of three rings. Two reference Ilizarov wires were applied, one at the proximal ring and the other at the distal ring. In addition, fixation



Fig. 3 Case 3. Juxta articular hinge used for translation of the tibia at the supramalleolar osteotomy

of the rings was done both by Ilizarov wires and half pins (Hybrid). The distal ring was connected to the middle ring at an angle that represented the amount of valgus at the ankle joint. Because the CORA was at the ankle joint and the supramalleolar corrective osteotomy was proximal to it, a translation of the distal tibia was performed with the application of a juxta-articular hinge (Fig. 3). A fibulotomy was also performed at the mid-diaphyseal area.

Correction of the distal valgus was achieved by resecting a medially based, supramalleolar closing wedge osteotomy. The wedge angle was equal to the valgus measurement on the long radiographs. After the wedge was removed, the osteotomy was closed with simultaneous translation (enabled by the juxta-articular hinge) and was fixed by connecting the distal ring to the middle one, resulting in parallel rings. The last step in this procedure was a corticotomy of the proximal tibia.

Hinges were also applied by an open laterally based wedge between the first and the second rings in order to progressively correct the knee valgus. In the legs that required length equalization, the elongation was done between the proximal and the middle rings.

Results

Five legs were treated with a bifocal Ilizarov procedure (Table 1). Knee valgus correction averaged 13.8° (range, 5° – 20°). Ankle correction averaged 20.3° (range, 15° – 30°). The average period from application to removal

of the Ilizarov apparatus was 4.6 months. The leg with the monofocal deformity at the knee was over-corrected with 5° of varus, while that with the monofocal deformity at the ankle was corrected to its neutral position. In all cases, the correction was maintained during the follow-up period. The functional outcome was reported to be satisfying by all patients.

Pin tract infection was seen in four legs without any case of osteomyelitis. None of the pins had to be removed or reinserted. Local treatment was sufficient to deal with these superficial infections and there was no need for systemic use of antibiotics.

In one case, iatrogenic transection of the common peroneal nerve was caused by an aberrant location of the nerve and its branches, resulting from a large osteochondroma at the proximal fibula. There was complete recovery of nerve function after an immediate microsurgical repair. In an additional case, peroneal neuropraxia developed but the patient recovered spontaneously after four months. We attribute this to the distraction at the proximal tibia.

In case 6, in whom the knee correction was performed by gradual open lateral wedge, the proximal fibula shifted into valgus and pressured the skin. This was corrected by the application of an “olive wire” which pulled the fibula back into normal position (Fig. 4).

Reconstruction of the mechanical axis, the goal of this treatment, was accomplished in five of the seven legs (cases 1–5), whereas in case 6 a varus of 5° and 3° was found at the knee and at the ankle, respectively. Equi-



Fig. 4a, b Case 6 (right leg). **a** The proximal fibula shifted laterally, in conjunction on the proximal valgus correction of the tibia, causing pressure of the skin, due to the synostosis between the proximal tibia and the fibula by the osteochondroma. **b** Correction of this condition was achieved by using an olive wire, which pulled the fibula back to normal position



Fig. 5 Anteroposterior radiograph of both legs of case 6. The right leg is five years after treatment, whereas the left leg demonstrates a valgus deformity of the proximal tibia, before monofocal Ilizarov correction

lization of limb length was achieved in all 3 patients that required it (cases 2–4).

Discussion

Osteochondromas in the proximal and distal fibula cause knee and ankle valgus in patients with HME. Ankle valgus is a result of a sequence of events; the osteochondroma interferes with the normal growth of the distal fibular physis, leading to a shortened fibula. Dias et al. [9] showed a correlation between a short fibula and ankle valgus in 173 cases, of which 18 had HME. The lateral support of the ankle joint is reduced due to the short fibula, enabling a sliding of the talus in the mortise, resulting in ankle valgus. In addition, the normal distribution of pressure applied to the distal tibial physis is altered, causing increased compression pressure on the lateral portion of the tibial physis, thus inhibiting growth. According to the Hueter-Volkman law, the increased amount of distraction pressure on the medial physis accelerates growth on this side, creating a wedge shape deformity of the epiphysis, further increasing the ankle valgus [10].

Another explanation for the development of ankle valgus is inherited growth disturbance of the distal fibula rather than a mechanical etiology [11]. Solomon [12] and Fogel et al. [13] found that in both upper and lower limbs, the deformity originates from the location of the osteo-

chondromas on a small cross-sectional bone area, i.e. ulna and fibula, respectively. These bones are more vulnerable to growth disturbance associated with the presence of the local osteochondroma.

Various surgical methods exist for correcting ankle valgus, such as epiphysiodesis of the distal medial tibial physis as described by Beals [4]. Other techniques such as transphyseal medial malleolar screws [5], screw epiphysiodesis and medial tibial hemiepiphysal stapling [4, 6] have been described as solutions for the correction of ankle deformity in HME. The advantages of these latter techniques are their minimal invasiveness, technical simplicity, and rapid recovery. Their drawbacks, however, are their inability to correct severe ankle valgus and to solve limb length discrepancy by elongation when needed.

No reports in the English medical literature were found describing simultaneous surgical correction of ankle and knee valgus in HME. There was, however, a single case report by Shawen et al. [7] describing the correction of ankle valgus due to HME by the Ilizarov apparatus.

The Ilizarov method enables simultaneous correction of multifocal multiplanar deformities. Therefore, it has been chosen as the treatment modality in these complex cases. The treatment of ankle valgus in our series was performed by osteotomy and an acute correction, unlike the 10-day progressive correction described by Shawen et al. [7]. In our series, knee valgus was corrected progressively to avoid peroneal nerve injury.

We are aware that correction of ankle valgus by stapling, for example, is much easier than the Ilizarov surgical procedure. Nevertheless, acute correction using the Ilizarov apparatus immediately equalizes the forces acting on the ankle joint as well as on the physis, whereas in stapling unbalanced pressure continues to affect the joint.

Another significant advantage of the approach advocated here is the immediate post-operative weight bearing, despite the two osteotomies performed in the same segment.

According to Paley's classification of complications [14], in this series there were three "problems", two of "obstacles" and no "major complications". The problems consisted of one case of neuropraxia of the common peroneal nerve and two cases of over-correction into varus. The "obstacles" included one case with iatrogenic transection of the common peroneal nerve and one case of proximal fibular shifting into valgus resulting in pressure on the skin. Both cases required additional surgical intervention. Despite these complications, the aim of the treatment was accomplished.

The Ilizarov method's unique advantage is the achievement of simultaneous correction of multifocal multiplanar complex deformities of the lower limbs in patients with HME.

In all cases, the correction was maintained and the functional outcome was satisfying.

The alternative to this would be multistage surgical procedures done separately for each deformity, a process that would require repeated anesthesia and a prolonged period of rehabilitation. Therefore, we recommend the use of the Ilizarov method as a satisfying and beneficial surgical technique for the treatment of these deformities.

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