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ORIGINAL ARTICLE

Congenital tibial deficiencies: Treatment using the Ilizarov's external fixator

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KEYWORDS
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Summary
Introduction: Congenital longitudinal deficiency of the tibia is a rare and often syndromic anomaly. Amputation is usually the preferred treatment option in complete absence of the tibia; however, a conservative management might be implemented in partial forms or in case of amputation refusal. Our experience with the Ilizarov fixator, convinced us this device was the best suited for progressive correction of lower limbs length discrepancies and articular or bone angular limb deformities (ALD). The aim of this study is to highlight the interest of the Ilizarov fixator in the multistage conservative treatment of congenital tibial deficiencies.
Material and methods: A retrospective study was conducted in nine patients suffering from Type I or II congenital tibial deficiencies (Jones) and sequentially managed using the Ilizarov technique. The functional outcome after treatment completion was then clinically assessed.
Results: The different stages of correction were recorded for each individual patient. Patients were assessed at a mean follow-up of 18,3 years (4—32 years). The mean maximum knee flexion was 35° (0°—90°) in type I deficiencies and 118° (90°—140°) in type II deficiencies. One patient underwent amputation and a bilateral knee arthrodesis was performed in another case.
Discussion: Few series in the literature report a comparable length of follow-up period in the conservative management of severe congenital tibial deficiencies. In our study, the Ilizarov fixator provided satisfactory progressive corrections of severe congenital tibial deficiencies.
Level of Evidence: Level IV therapeutic retrospective study.

Introduction

Tibial deficiency is a rare congenital condition which incidence approximates 1/1 million births [1]. The type and degree of tibial deficiency vary from complete absence of the tibia to partial deficiency with intact extensor mechanism [2]. Tibial deficiency is always associated with an equinovarus foot deformity in the most severe cases. Other associated anomalies are also reported and should be investigated.

Amputation is the recommended treatment option in patients with complete tibial deficiency [3—5]. Conservative treatment is less common. It classically includes fibular

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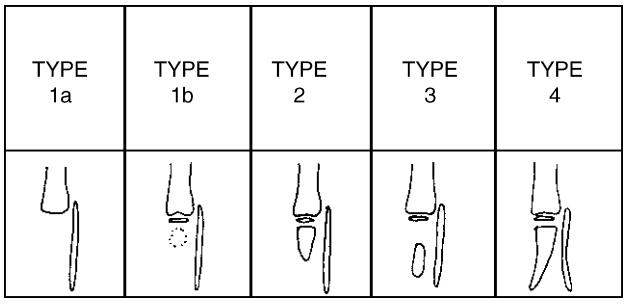

Figure 1 Classification of congenital tibial deficiencies according to Jones et al. [12].
Type I: absence of the tibia (further divided in Ia: hypoplastic lower femoral epiphysis; and Ib: normal lower femoral epiphysis). Type II: absence of the distal tibia. Type III: proximal tibia not seen. Type IV: diastasis of the distal tibiofibular joint.

tibialization, foot repositioning under the fibula and lower limb lengthening [1,6—10]. The timing and staging of these procedures vary according to the surgeon.

In the light of our experience with the Ilizarov fixator, this method appears highly efficient in the progressive correction of lower limb length discrepancies and articular or bony angular deviations [11]. The aim of that study was to demonstrate the interest of the Ilizarov external fixator in the multistage conservative management of congenital tibial deficiencies.

Patients and method

A retrospective study was conducted in nine consecutive patients suffering from congenital tibial deficiencies and managed sequentially using the Ilizarov device. There were four girls and five boys. The average age at the beginning of correction was two years and one month (1—4 years).

Patients clinical data was used for classification of tibial deficiency according to Jones et al [12] (Fig. 1) and evaluation of associated anomalies. The timing and staging of the correction was planned (Table 1). Interrecurrent complications were noted for each patient.

A conservative treatment was systematically undertaken for type II partial deficiencies. Patients with type I complete bilateral tibial deficiencies or those who refused amputation, were also managed conservatively.

The surgical strategy was adapted to each case. However, from our experience, this strategy evolved toward a progressive correction combining centralization of the fibula under the femur and a ''one-stage'' repositioning of the foot under the fibula, using the Ilizarov fixator. The external fixation device comprised: an upper femoral ring with a couple of Kirschner wires and a third wire mounted on a flag; a second fibular ring and a third calcaneal ring. These rings were connected to one another by distraction rods. The upper part of the frame ensured progressive lowering of the fibula and the lengthening effect occurring between the two distal rings provided lowering of the foot. These procedures were performed simultaneously. The rhythm of ring distraction was 1 or 2 mm per day according to the child tolerance. This progressive correction of the deficient limb allowed further surgical repositioning procedures.

In type I deficiencies, stabilization of the fibula under the femur was performed according to the Brown technique [1]. Residual components of the extensor mechanism were fixed on the proximal part of the fibula. In type II deficiencies, either the whole fibula or only its distal part was lowered after a proximal osteotomy. The choice between these two techniques depended on the programme of correction. When no equalization had been planned, the objective was to provide a minimal length for the achievement of a stable equipment. Therefore, lowering the entire fibula did not appear essential. Conversely, when equalization was planned, lowering the whole fibula was crucial to provide the limb with maximum length. In these conditions, in type II deficiencies, the fibula was surgically connected to the proximal tibia with less tensioning, thus realizing a tibiofibular synostosis.

Foot repositioning consisted in a periarticular release which was often difficult due to major retractions. Among the seven cases undergoing surgical repositioning of the foot under the fibula, two cases were prepared with the Ilizarov fixator. The Ilizarov fixator was used to lower the foot and prepare the talofibular release-arthrodesis without the need for a shortening osteotomy. In both cases (cases n^{os} 8 and 9), an isolated release of the retracted tissues thus ensured the correction. In any other case, a shortening osteotomy of the fibula was required to ensure reduction. A monolateral fixator first maintained the temporary shortening, thus enabling further progressive limb length restoration. Surgical release consisted in the resection of all retracted fibrous and tendinous elements. Reduction of the foot was maintained using two talofibular wires. The arthrodesis was usually performed at the talus posterior part. The limb was then immobilized during a three-month period in a long-leg cast.

In our series, five patients underwent progressive lengthening at the end of correction using a monolateral external fixator or Ilizarov fixator with a mean correction of 9 cm (5—17 cm).

In case n^{o} 1, fibular widening was performed. This procedure was initially described by Ilizarov. A longitudinal anteroposterior fibular osteotomy was performed. Olive wires were placed from outside to inside, at different levels, on the whole height of the fibula. Each wire was lateraly fixed on a threaded rod which ensured a transverse traction.

At the end of correction, the lower limb was systematically maintained in a long-leg splint, first permanently to avoid any recurrence of deformity, then partially depending on the knee stability.

Evaluation of functional result at the end of correction was based on clinical criteria. Physical examination included maximal flexion and knee stability as well as the type of walking-aid device. Residual limb length discrepancy was clinically and radiographically assessed.

Results

All patients were managed from birth, four of them had a type I complete tibial deficiency according to Jones et al. [12] and one had a bilateral deficiency. Five patients had a type II partial deficiency (Fig. 2a). Five patients had an associated anomaly (Table 1).

Table 1 Stages of the reconstructing procedure for all cases. Associated deformities are noted for each patient.

Case	Type	Associated anomaly	Age (years)	Stages of correction
1	1	None	1	Open repositioning of the fibula under the femur according to Brown
			1	Foot repositioning under the fibula via posteromedial release and talofibular arthrodesis
			2	Fibular derotational osteotomy for excessive internal rotation of the foot
			14	Progressive fibular widening using the Ilizarov fixator
			16	Open iterative correction of hindfoot varus deformity
			17	LLD 13 cm: 14 cm fibular lengthening with Ilizarov
			20	Osteotomy for genu recurvatum
2	1	None	1	Open repositioning of the fibula under the femur according to Brown
			10	LLD 10 cm: 10 cm fibular lengthening with Ilizarov
			11	Genu flessum and posterior dislocation of the tibia: Correction with Ilizarov fixator
			11	Proximal metaphyseal fibular osteotomy for genu recurvatum
3	2	None	2	Foot repositioning via posteromedial release and talofibular arthrodesis 15 mm shortening osteotomy of the fibula
			3	Tibialization of the fibula via tibiofibular synostosis
			5	Percutaneous epiphysiodesis of fibular proximal physis for hypertrophy of the fibular head
			8	LLD 5 cm: 5 cm fibular lengthening and iterative foot repositioning using the Ilizarov fixator
			8	Fracture of the regenerate bone
			13	LLD 5 cm: 5 cm fibular lengthening with Ilizarov
4	1	Bilateral Type 1	2	Foot repositioning via posteromedial release and talofibular arthrodesis combined with fibular shortening osteotomy
			2	Correction of flessum according to Wagner
			10	Open repositioning of the fibula under the femur according to Brown
			15	Knee arthrodesis
			18	Varus malunion: valgus osteotomy
			19	Nonunion secondary to osteotomy: compression with Ilizarov fixator
5	2	Bilateral congenital hip dislocation Interventricular communication	4	Foot repositioning via posteromedial release and talofibular arthrodesis 15 mm shortening osteotomy of the fibula
			4	Tibialization of the fibula via tibiofibular synostosis
			9	LLD 16 cm: 5 cm fibular lengthening with Wagner
			13	LLD 10 cm: 6 cm fibular lengthening with Ilizarov
			18	LLD 9 cm: 6 cm fibular lengthening with Ilizarov
6	2	Syndactyly of third and fourth left toes	3	Fibular centralization with Ilizarov
			3	Tibialization of the fibula via tibiofibular synostosis Foot repositioning via posteromedial release and talofibular arthrodesis
7	2	Type IV left congenital tibial deficiency	2	Foot repositioning via posteromedial release and talofibular arthrodesis Fibular shortening osteotomy
			2	Fibular centralization with Ilizarov
			2	Tibialization of the fibula through tibiofibular synostosis
			3	Nonunion: decortication
			14	LLD: 5 cm lengthening with Ilizarov
			15	Nonunion: decortication + graft
8	1	None	4	Correction of genu flessum and fibular centralization with Ilizarov
			4	Femoral shaft fracture
			4	Removal of external fixator Foot repositioning under the fibula via posteromedial release and talofibular arthrodesis Open repositioning of the fibula under the femur according to Brown
			5	Fibular fracture
			5	Recurrence of genu flessum: correction with casts
			6	Thigh amputation
9	2	None	2	Correction of genu flessum and fibular centralization with Ilizarov
			2	Foot repositioning via posteromedial release and talofibular arthrodesis
			3	Tibialization of the fibula via tibiofibular synostosis
			5	Internal rotation gait: 40° external derotation osteotomy
			6	LLD: 4 cm

LLD: leg length discrepancy.

Table 2 Knee functional outcome at the end of correction.

Case	Type	Maximal flexion (°)	Stability	LLD (cm)
1	Type 1	90	Unstable	0
2	Type 1	50	Unstable	−3
3	Type 2	140	Stable	−1
4	Type 1	0	—	—
5	Type 2	90	Unstable	−2
6	Type 2	90	Unstable	Lost to follow-up
7	Type 2	130	Stable	0
8	Type 1	0	—	—
9	Type 2	140	Unstable	−4

LLD: leg length discrepancy.

Evaluation was performed at a mean follow-up of 18.3 years (4—32 years). One patient was lost to follow-up (case nº 6). One (case nº 8) had a knee disarticulation due to the recurrence of deformity. One patient (case nº 4) underwent a bilateral knee arthrodesis. These two patients had a complete congenital tibial deficiency. Excluding these two patients, mean range of maximal knee flexion was 106°. All patients having kept their knee mobility could achieve complete extension and walk using a femoral leg splint with knee immobilization. The mean maximal knee flexion was 35° (0°—90°) in type I deficiencies and 118° (90°—140°) in type II deficiencies. In two patients (cases nºs 3 and 7), knee stabilization was achieved at the end of correction (Table 2) (Fig. 3).

Discussion

Most authors advocate knee disarticulation or transtibial amputation in the management of congenital tibial deficiencies [5,13]. In some communities, parents do not accept amputation [7,8]. These refusals lead us to suggest a conservative management as in partial deficiencies. The multiple-stage correction of the affected limb includes: tibialization of the fibula, foot repositioning and management of limb length discrepancy in a single or multiple-stage lengthening. The aim of that study is to point out the interest of the Ilizarov fixator in the sequential conservative management of severe congenital tibial deficiencies.

Tibialization of the fibula and foot repositioning were classically performed separately at an interval of several months. The order of priority for the knee or ankle correction differed according to each case. The extemporaneous surgical realignment was made difficult due to periarticular retractions. A shortening osteotomy of the fibula was systematically performed but resulted in a loss of length in an already short limb. Our experience with the Ilizarov fixator allowed us to consider a ''one-stage'' progressive repositioning of the foot and fibula while preserving the length of the affected limb.

Tibialization of the fibula was initially described by Brown [1]. This procedure was used to treat type I deficiencies [4,6—8,10]. In patients with type II deficiencies, fusion of the fibula to the tibial remnant might be performed [4,14]. In type I deficiencies, Weber et al. [15] have described a knee arthroplasty technique using the patella. This technique appears to provide a better stabilization of the knee but we lack experience about it. With no preparation, these surgical techniques induce frequent complications [16]. We used the Ilizarov fixator in four cases to achieve a progressive correction of genu flessum then to centralize the fibula. However, in all cases and more specifically in type I deficiencies, without the presence of active knee extension [10,16], the obtained correction should be rigorously maintained with an external device to avoid early recurrence of flessum. In case nº 8, the Ilizarov fixator provided complete correction of the genu flessum, but non compliance of the

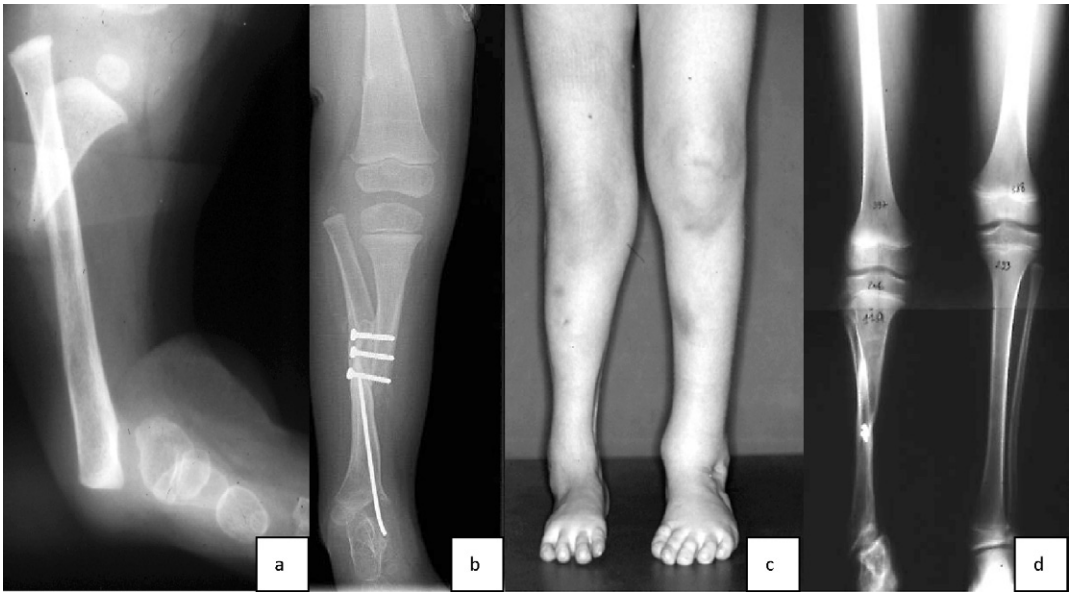

Figure 2 Type II congenital tibial deficiency (case nº 9); a: radiographic aspect in a two-year old patient; b: radiographic aspect following tibiofibular synostosis and talofibular arthrodesis in the same patient; c: clinical and (d) radiographic aspects at maturity, in the same patient.

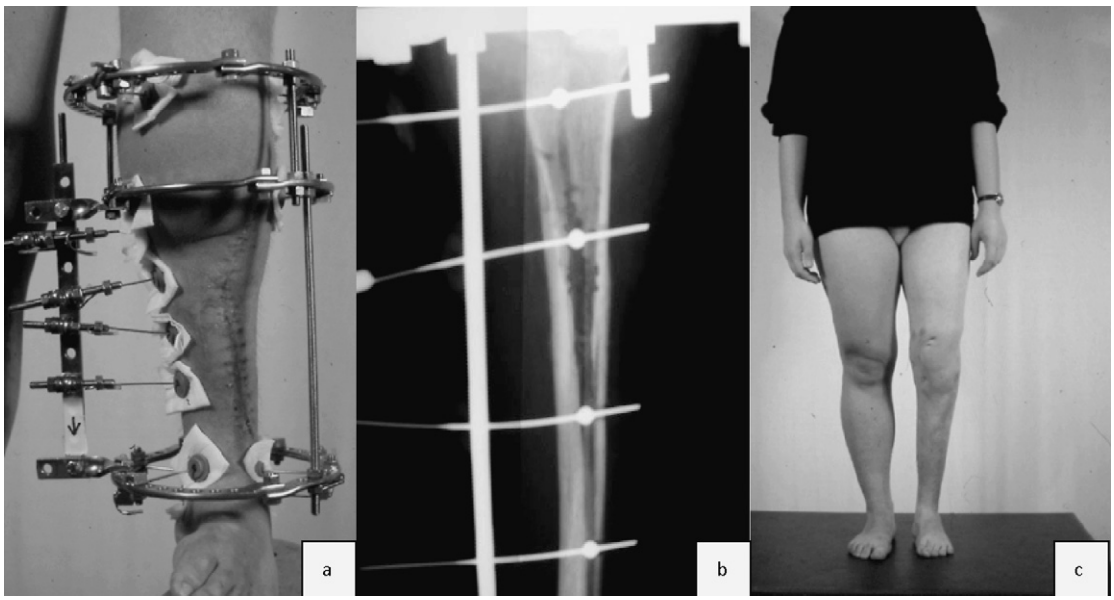

Figure 3 Congenital deficiency of the tibia type 1 (case nº 1); a: clinical and (b) radiographic aspect during transversal widening of the fibula with the Ilizarov fixator in the same patient; c; clinical aspect at maturity in the same patient.

patient with the orthopaedic treatment led to the recurrence and to the limb amputation, which had initially been suggested to the family.

The Ilizarov external fixator was initially introduced in the surgical treatment of congenital tibial deficiencies to manage residual limb length discrepancies with progressive lengthening of the centralized fibula or of the fibula fixed to the tibia [7—9,14]. In our series, the Ilizarov fixator was used at the end of correction in five patients for progressive lengthening and achieved a mean correction of 9 cm (5—17 cm). Equalization was achieved each time the program could be completed, with a mean residual shortening of 1.5 cm (0—4 cm). This residual inequality is well tolerated but might be compensated prior to maturity by a contralateral tibial epiphysiodesis. It is most of the time compensated by the residual equinus resulting from the foot deformity and only requires the need for an adapted fitting or long-leg splint. One patient is still under correction (case nº 9) and has not undergone lengthening yet. Our treatment option in the management of congenital short limbs varies according to the underlying joint stability but also to the anticipated limb length discrepancy at maturity [17]. Limb equalization does not guarantee functional improvement and limb lengthening induces many complications. This is the reason why we only resort to limb lengthening when the anticipated limb length discrepancy is expected to be less than 15 cm. Even if in some cases of our series, we did perform one-stage lengthenings of more than 10 cm through to the end, we advocate from now on a two to three-stage method.

The most challenging aspect of such lengthenings is not stabilization of the ankle through talofibular arthrodesis but knee instability. Therefore, the external fixator should bypass the knee during lengthening. However, most authors contra-indicate fibular lengthening in type I deficiencies and save it for partial forms [8,9], when the knee is more stable. All lengthening procedures described in the literature (11—17,5 cm) involve type II deficiencies. In our series, equalization could be achieved in all cases. At the end of correction, multidirectional instability of the knee generally requires the need for a long-leg splint with or without knee immobilization, depending on the quality of the quadriceps function. The presence of a remnant of the proximal tibia in type II deficiencies does not guarantee the presence of the stabilizing structures of the knee. In cases nºs 5, 6 and 9, instability was due to the absence of the central pivot of the knee. The permanent equipment enables proper knee flexion in the oscillating phase while preventing lateral and recurvatum instability during weight-bearing. Knee arthrodesis ensures stabilization of the knee but do not restore an harmonious gait pattern. It was necessary in case nº 4 due to the recurrence of genu flessum.

Weight-bearing usually ensure proper qualitative tibialization of the fibula combined with global widening and remodelling of the proximal epiphysis in a tibial plateau shape. In one case (case 1), we were brought to perform a progressive widening of the fibula in the frontal plane to improve this remodelling. The final radiographic aspect favors the remodelling of the proximal epiphysis of the fibula that takes the shape of a tibial proximal epiphysis (Fig. 4).

In our series, the conservative treatment of congenital tibial deficiencies is long and punctuated with many intercurrent complications. Our series is heterogeneous but few series in the literature report similar follow-up. Hosny [8] reports good results with a reconstructing technique using the Ilizarov fixator in type I and II congenital tibial deficiencies. Wada [7] and de Sanctis [14] show similar outcome without prior preparation using the external fixator at the cost of resections and secondary progressive lengthenings. With a mean follow-up of 18 years, the complications reported in our series are those commonly encountered when performing progressive limb lengthening with an external fixator: pin-related fractures, fractures occurring in regenerate bone, pin-track infections or malunions.

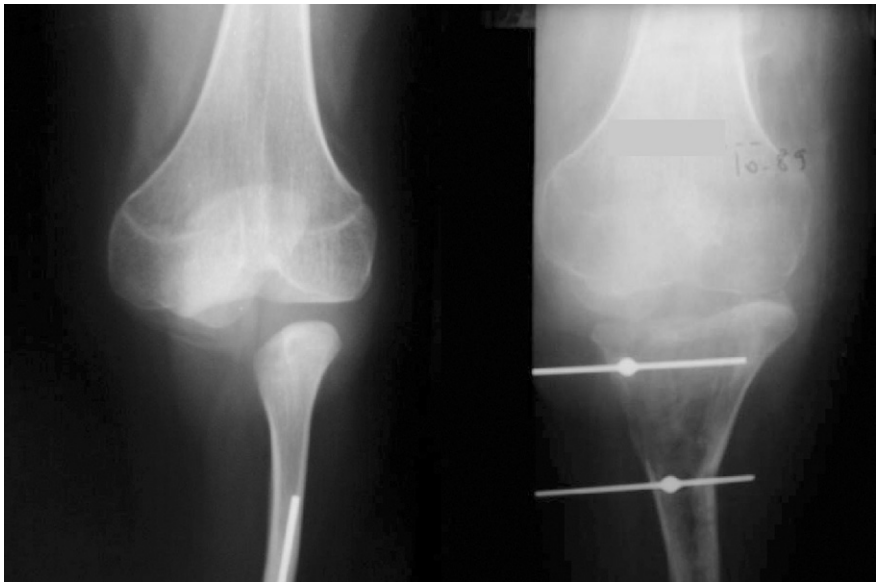

Figure 4 Congenital deficiency of the tibia type 1 (case nº 3). Radiographic aspect (A-P view) of the knee before and after transversal widening of the fibula.

Functional outcome at the end of correction is satisfactory but two failures of the conservative treatment led to a knee arthrodesis in one case and an amputation in another case. Amputation had been suggested and initially asked for by the patient but was refused by the family.

Conclusion

Due to the frequently reported associated anomalies in type I congenital tibial deficiencies, amputation remains the treatment of choice. Conservative treatment should be applied in the primary management of type II deficiencies or in case of amputation refusal in type I deficiencies. Proper restoration of limb function results from both the reconstruction of the knee and ankle joints and lower limb lengthening. Tibialization of the fibula is the main aspect of a long and difficult reconstruction programme. However, the Ilizarov fixator modularity in the management of limb length discrepancies and joint retractions is a valuable tool in optimizing the progressive correction of congenital tibial deficiencies.

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