HEMIMELIA PERONEA

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HEMIMELIA PERONEA

- HIPOPLASIA O AGENESIA DE PERONE
- ECTROMELIA
- DEFICIENCIA PERONEA CONGENITA

HEMIMELIA PERONEA

- Malformacion somatica MAS FRECUENTE de huesos largos

- Sin herencia dominante

Casi siempre ASOCIADA a otras malformaciones : ACORTAMIENTO FEMORAL E HIPOPLASIA COND.EXT

- ALT LCA Y P INCURVACION TIBIAL Y ACORTAMIENTO

HEMIMELIA PERONEA

- Alteracion de tobillo : VALGO
- AUSENCIA MALEOLO LATERAL
- BALL AND SOCKET

- Alteracion de pie: AUSENCIA DE RAYOS LATERALES
- SINOSTOSIS AGENESIAS

Clínica

HEMIMELIA PERONEA

- SU TRATAMIENTO ESTARA CONDICIONADO POR

- MALFORMACION DE TOBILLO Y PIE
- PORCENTAJE DE ACORTAMIENTO

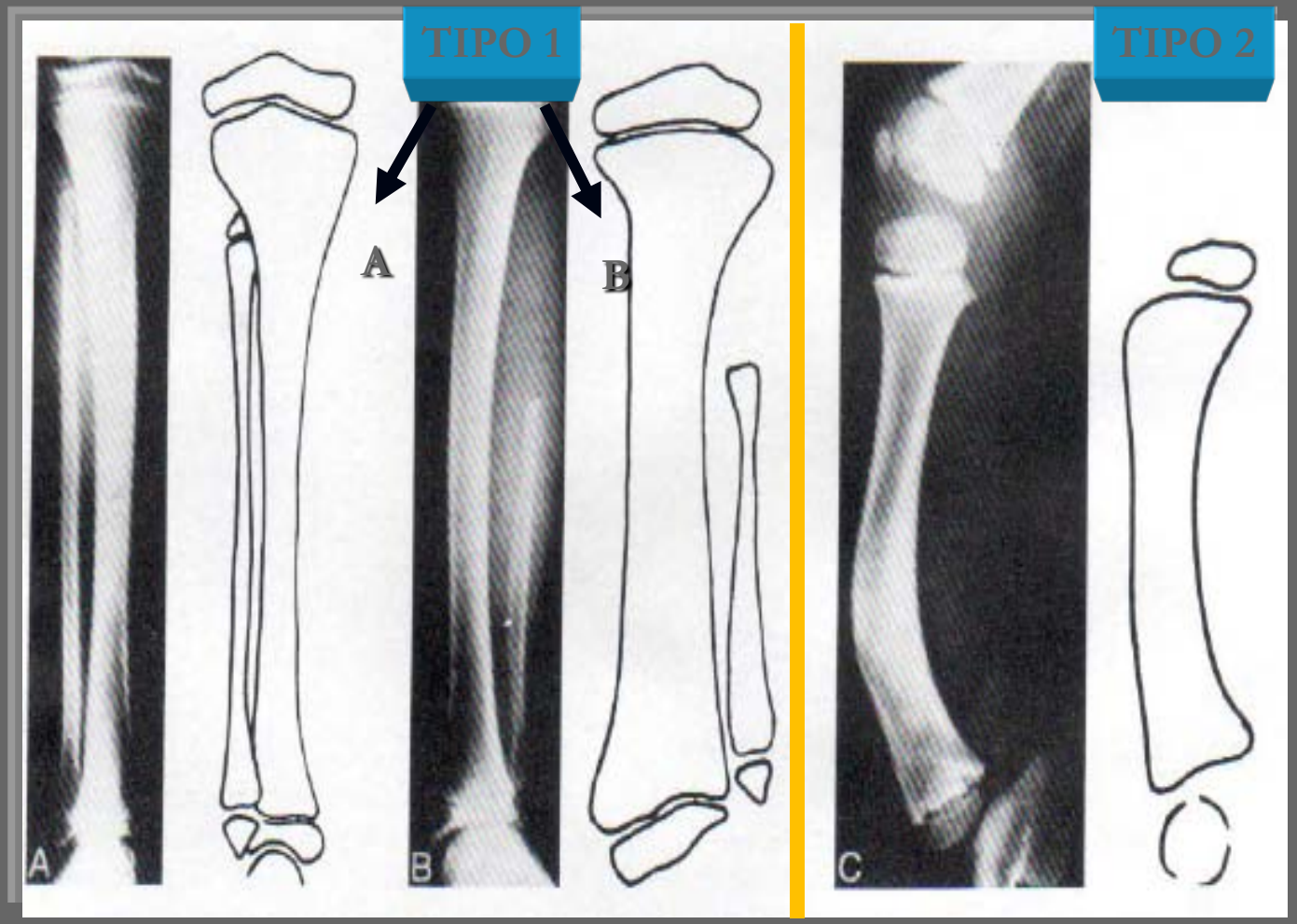
- **RECONSTRUCCION vs EQUIPAMIENTO**

HEMIMELIA PERONEA

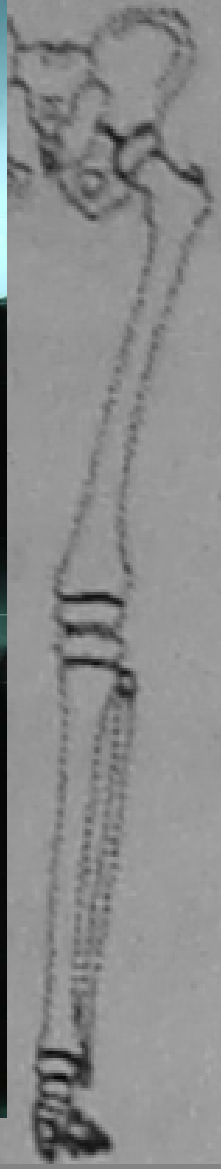
- CLASIFICACIONES
- ACHTERMAN Y KALAMCHI (descriptiva)
- DALMONTE (descriptiva orientativa)
- STANISKY BIRCH PALEY (ORIENTACION TERAPEUTICA)

Clasificación

Achterman y Kalamhi 1979 descriptiva



Clasificación (Delmonte)



Tipo I :

- Acortamiento perone y tibia minimo
- Discrepancia al final del crecimeinto e/3-8cm
- Inestabilidad de rodilla y tobillo minima o ausente
- Tobillo ball-and-socket, pero estable
- Puede faltar uno o dos rayos laterales, pero pie funcional

ficación (Delmonte)

Tipo II :

- Acortamiento perone severo, maleolo lateral no funcional.
- Deformidad en equinovalgo del pie y tobillo
- Rodilla en valgo hipoplasia condilo femoral externo
- Femur mas corto y tibia con procurvatum
- pueden faltar uno o dos rayos laterales

ificación (Delmonte)

Tipo III :

- Perone ausente solo un rudimento.
- Tibia muy corta y con desviación antero lateral
- Deformidad en equinovalgo del pie muy severa con luxacion franca.
- Rodilla con inestabilidad medial y anteroposterior
- Fémur corto con hipoplasia condilo femoral externo y con algunos signos de displasia acetabular
- Miembro pélvico con ligera extrarotacion
- Ausencia de 2,3 o mas rayos laterales

Clasificación (Delmonte)

Tipo III :

- Perone ausente solo un rudimento.
- Tibia muy corta y con desviación antero lateral
- Deformidad en equinovalgo del pie muy severa con luxacion franca.
- Rodilla con inestabilidad medial y anteroposterior
- Fémur corto con hipoplasia condilo femoral externo y con algunos signos de displasia acetabular
- Miembro pélvico con ligera extrarotacion
- Ausencia de 2,3 o mas rayos laterales

PROTOCOLO TRATAMIENTO DE PALEY

- LOS MALOS RESULTADOS SE DEBEN A LA DEFORMIDAD RESIDUAL DEL PIE

PROTOCOLO TRATAMIENTO PALEY

- Tipo 1 : alargamiento
- Tipo 2 : osteotomia supramaleolar + alargamiento
- Tipo 3: a-reseccion de la brida ,osteotomia supramaleolar y subastragalina
- b-alargamientos sucesivos mas epifisiodesis
- c-artrodesis de tobillo si no es posible estabilizar

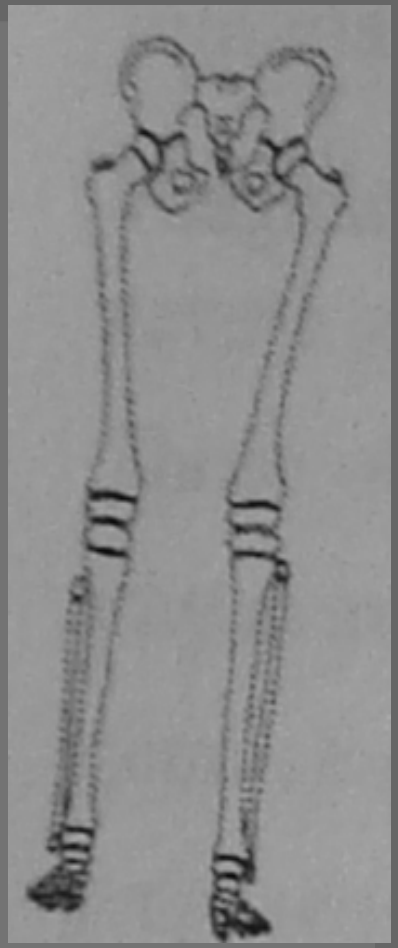
PROTOCOLO TRATAMIENTO PALEY

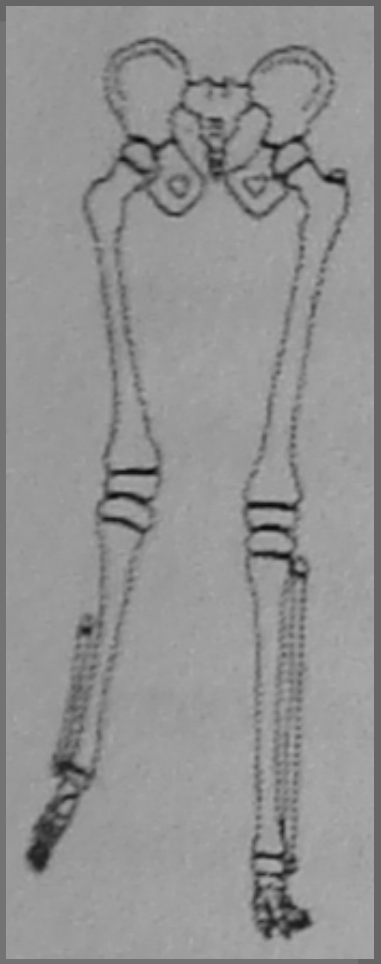
- 18 a 24 meses : superankle y alargamiento de 5 cm
- 7 a 10 años :2° alargamiento de 5 a 8 cm
- 12 a 14 años : 3° alargamiento de 5 a 8 cm y /o

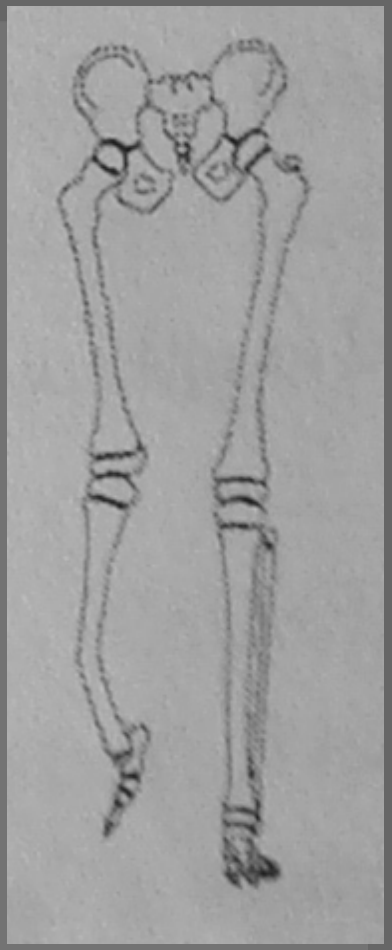
  epifisiodesis

Elongacion final de 12 a 26 cm con procedimientos sucesivos combinados

Protocolo terapéutico

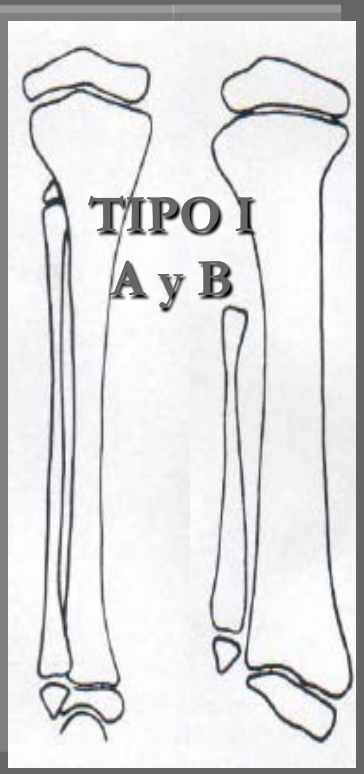






Protocolo terapéutico

<1 año Ortesis

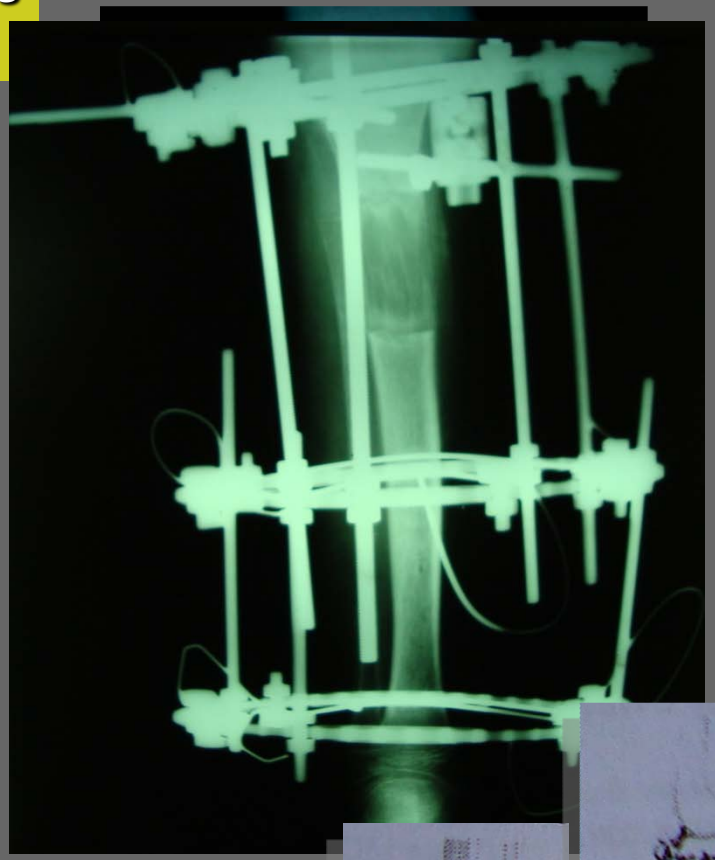


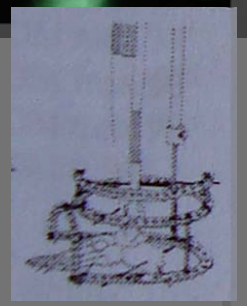
Tobillo epifisis
horizontal

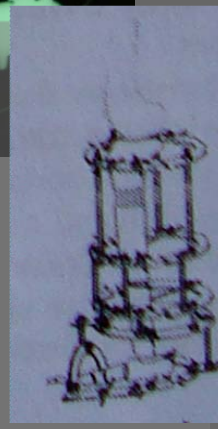
Liberación blanda
Y banda peronea
Al año de vida

Ortesis y realce
Hasta los 5 años

Elongaciones óseas
Sucesivas de acuerdo a forma
De epífisis tibial proximal







Como se hace la elongación ?

- 1º Calcular la discrepancia al final del crecimiento (Met. Moseley o el de Multiplier), incluir fémur y tibia en los cálculos.
- El fijador es discutible, pero se tiene mas experiencia con Illizarov.
- Siempre tomar calcáneo, proteger la rodilla cuando las espinas tibiales son hipoplasicas.
- Si es necesario se libera la sinostosis astrágalo calcáneo.
- Se puede realizar hemiepifisiodesis transitoria femoral interna.
- Ritmo de elongación lento ½ mm día, no superar los 5 cm.

Complicaciones ?

Protocolo terapéutico

TIPO II

<1 año
Esperar evolución

Yesos, ortesis o nada

¿ Reconstruir Vs. Amputar ?

Escuela Norteamericana

Discrepancia al año de entre 3,5 y 5 cm
Con una final calculada de alrededor de 10 a 12,5 cm.
Con un pie rígido con 2 dedos o menos

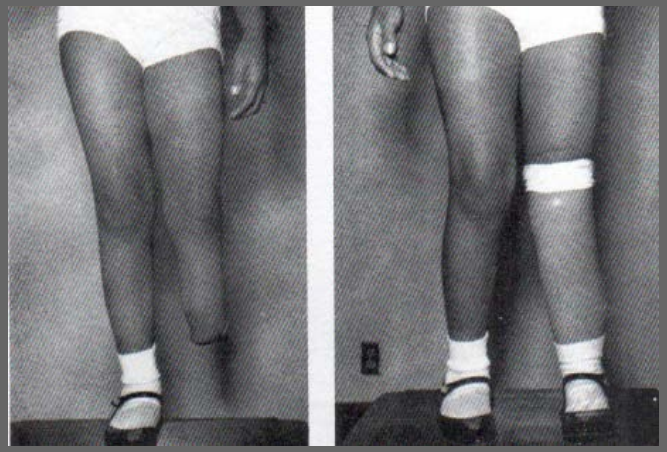
Protocolo terapéutico

TIPO II

< 1 año
Esperar evolución

¿ Reconstruir Vs. Amputar ?

Discrepancia al año de entre 3,5 y 5 cm
Con una final calculada de alrededor de 10 a 12,5 cm.
Con un pie rígido con 2 dedos o menos



Protocolo terapéutico

TIPO II

< 1 año
Esperar evolución

Yesos, ortesis o nada

Reconstruir

Al año
liberacion blanda +
Osteotomia cora tibial
(correctora de antecurvatun y valgo)
Fijada con kirchner transcalcaneo.
Con o sin operación de Gruca

Elongaciones óseas
Sucesivas de acuerdo a forma
De epífisis tibial proximal

Protocolo terapéutico

¿ Reconstruir Vs. Amputar ?

TIPO II

Bending osteotomy through the distal tibial physis in fibular hemimelia for stable reduction of the hindfoot

G. Ulrich Exner

Fibular hemimelia is associated with a deformity of the distal tibial epiphysis resulting in a convexity with lateral and posterior slope of the distal joint surface. The deformity results into an equinovalgus position of the foot and ankle and frequently consecutive dislocation of the foot. A new procedure is presented, in with a metaphyseal osteotomy is performed towards the physis in order to bend the postero-lateral third of the distal tibial epiphysis forming a concave distal tibial joint surface. This procedure was done in four feet in three patients at ages between 7 and 20 months combined with soft tissue releases and rebalancing the tendons. At a follow-up between 6 and 42 months the position of the feet is well retained. *J Pediatr Orthop B* 12:27–32 © 2003 Lippincott Williams & Wilkins.

Journal of Pediatric Orthopaedics B 2003, 12:27–32

Keywords: Fibular hemimelia, metaphyseal osteotomy, tibial epiphysis

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Introduction

In fibular hemimelia the hindfoot lacks lateral support from the fibular malleolus; additionally the distal tibial epiphysis is wedge-shaped and the resulting convexity adds to the instability of the hindfoot, which tends to slip off the tibia laterally and dorsally. This was documented recently by Choi *et al.* [1].

To provide stability a technique of forming a fork by splitting the distal tibia was described by Rincheval [2]. This author learned the technique from Bardenheuer, which was reported later on by Gruca, and cited and described by Thomas and Williams [3]. We have performed the Bardenheuer–Gruca procedure in a few cases and reported on our first case [4]. The Bardenheuer–Gruca procedure is effective; however, complications may result from fusion of the epiphysis and loss of further growth. Today we consider this procedure rather a salvage, when the proper timing for the new procedure presented in this report, has been missed.

The new osteotomy was invented to change the convexity of the distal tibia to a concave shape in order to improve the support of the talus underneath the tibia. The osteotomy is performed through the lateral third of the distal tibial metaphysis directed onto the growth plate, which allows bending of the lateral third of the epiphysis distally. The short-term results appear very promising and we therefore wish to communicate this technique.

Patients and methods

Three patients were treated with this method (two girls and one boy). Both girls had unilateral absence of the fibula and short femur, the boy had bilateral absence of the fibula and mildly short femurs. The feet had four rays in one patient and three rays in the others. According to the classification of Achterman and Kalamchi [5] all patients had type II deficiencies with unstable feet in equinovalgus position. From the intraoperative findings a talo-calcaneal coalition at least of the fibrous type is to be assumed. Surgery was performed in patient A.L. (Fig. 1) at 7 months, in patient L.Z. (Fig. 2), at 18 months of age, in patient F.L. at 15 months on the right and at 20 months on the left.

The principle of the surgery is shown in Figure 3. A wide exposure of the fibular and lateral structures of the foot is used as shown in Fig. 2d. This exposure is used in order to be clear about the anatomy, to ensure identification of the fibular anlage, if present, and to fully resect it, as to decide for appropriate transposition of tendons at appropriate length; e.g. peroneal tendon transposed more proximally, lengthening of tendo achilles. A soft tissue release is performed around the hindfoot to allow for reduction of the foot, carefully avoiding any unnecessary devascularization. The distal metaphysis of the tibia is identified and the planned osteotomy is marked with a K-wire (Figs. 1c, 2d and 2e) and then marked with a saw and completed with a chisel, carefully avoiding entering the growth plate. The postero-lateral part of the epiphysis is next bent through the growth plate and a small bone block, taken a little more proximally from the tibia, is interposed to correct for valgus as well as equinus deformity of the distal tibial joint surface. The foot is transfixed in the reduced position with a K-wire. A long leg cast with the knee in 90° flexion is applied for 6 weeks. The K-wire is then removed and an

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Limb lengthening in fibular hemimelia type II: can it be an alternative to amputation?

Daniel Zarzycki, Barbara Jasiewicz, Wojciech Kacki, Arkadiusz Koniarski, Marcin Kasprzyk, Maja Zarzycka and Maciej Tesiorowski

The purpose of our study was to analyze limb lengthening in fibular hemimelia type II. Ten patients underwent 16 tibia lengthenings. The mean tibia shortening was 5.8 cm. We used the Ilizarov technique in all cases. The mean follow-up time was 22 years. The mean lengthening was 23% of the former length. The healing index was 50.8 days/cm. In the final examination six patients were skeletally mature, equal limb length and functional foot positioning were achieved in four of them. Complications were observed during 14 lengthenings (87.5%). Although lengthening in fibular hemimelia is difficult, elongation with axis and foot correction may offer an alternative to amputation. *J Pediatr Orthop B* 15:147–153 © 2006 Lippincott Williams & Wilkins.

Journal of Pediatric Orthopaedics B 2006, 15:147–153

Keywords: limb lengthening, the Ilizarov method, fibular hemimelia

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Introduction

Fibular hemimelia is the most common longitudinal deficiency in which the tibia is usually short with an axis deviation (anterior bowing and valgus deformity) [1,2]. Fibular hemimelia can vary in its extent; from minimal fibular shortening to total absence, which is the basis of the Achterman–Kalamchi classification, with type II complete absence of the fibula [1,3].

Until recently, treatment in cases of fibular aplasia included early amputation of the limb [4–6]. The development of modern lengthening methods potentially allowed for the lengthening of a shorter limb. Amputation is still popular, however, with orthopedists who emphasize good functional results in patients who have undergone early amputation [2,4,6–8].

The purpose of our study was to analyze limb lengthening in fibular hemimelia type II as an alternative to amputation.

Materials and methods

Between 1989 and 2001 we treated 10 patients with fibular hemimelia type II according to Achterman–Kalamchi classification: five girls and five boys, who underwent a total of 16 tibia lengthenings. In one case, the patient's deformity was bilateral (both legs type II), in another it was associated with type Ib in the other limb and in the other eight cases it was unilateral. According to the Birch functional classification there were one type IB patient, four type IC patients, two type ID patients, two type IIA patients, and one type IIB patient [9]. The mean age at the time of lengthening was 10 years (range 5.1–20).

Fibular deficiency was associated with various defects of the ankle joint and foot and details are presented in Table 1. Six patients had undergone surgery previously before they were 5 years old, as a result of foot deformity; three of them did so in our hospital, one underwent foot correction twice, and one patient underwent not only foot correction, but also femoral lengthening using the Wagner technique and epiphysiodesis on the uninvolved side. Tibia lengthening was performed on three patients twice, and three times on one patient. In the case of the bilateral defect, correction of the tibia axis and foot position was carried out on both sides. On average, tibia shortening was 5.8 cm, from 0 (lengthening in advance) to 14 cm, covering 19.6% (range 0–47%) of the length of the unaffected tibia (Fig. 1). The range of mobility of the knee was normal in the case of nine patients. Ninety degrees of flexion was present in one patient (prior to two lengthenings), 10° flexion contracture was observed in two patients.

Before lengthening, anterior bowing of the tibia was noted in seven cases (mean 21°; range, 13–32), valgus deformity in two cases (10° and 12°), and valgus with anterior bowing in three cases (mean 13° valgus and 23° anterior bowing).

Equinus deformity of the foot or the absence of a dorsal flexion was observed in nine cases, dorsal contracture in one case and stiff ankle joint in four cases; a normal range of motion and foot position was present in only two cases. For the purposes of lengthening and axis correction we used the classic Ilizarov technique in all cases [10,11].

The patient with the bilateral defect underwent axis correction with foot positioning correction carried out

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Protocolo terapéutico

¿ Reconstruir Vs. Amputar ?

TIPO II

Protocolo terapéutico

¿ Reconstruir Vs. Amputar ?

TIPO II

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¿ Reconstruir Vs. Amputar ?

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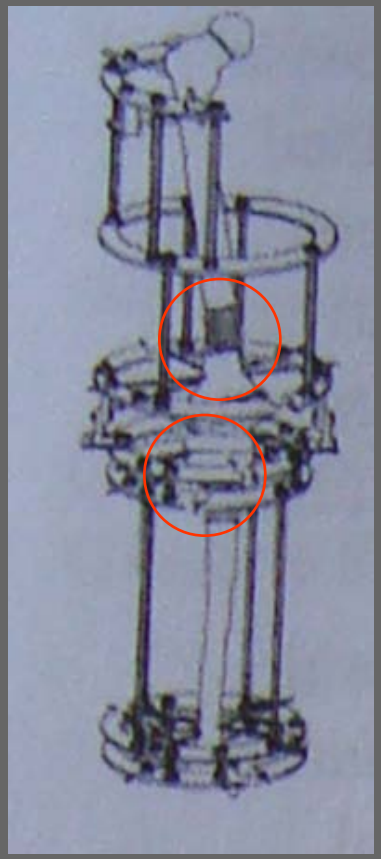
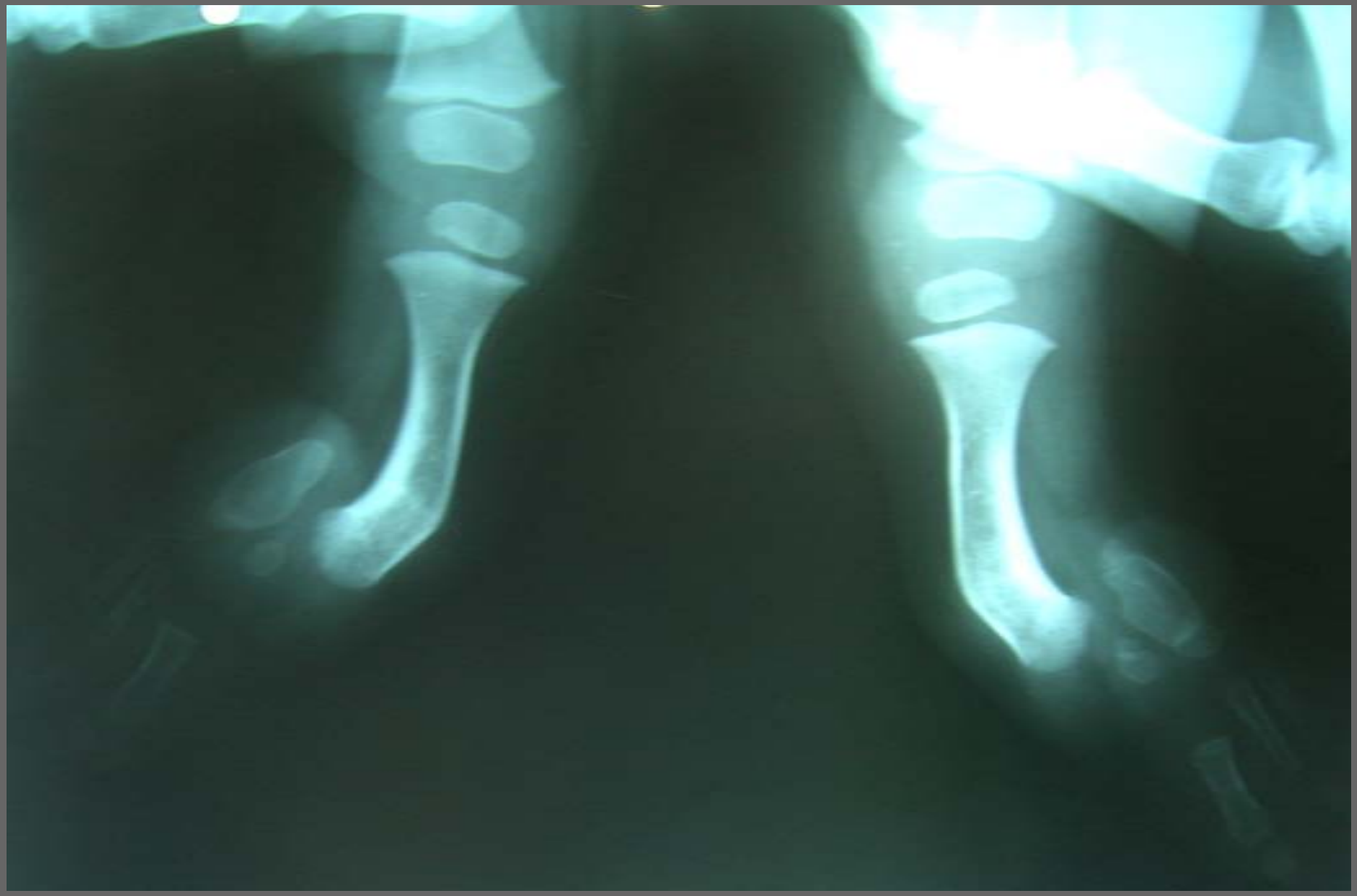
Protocolo terapéutico

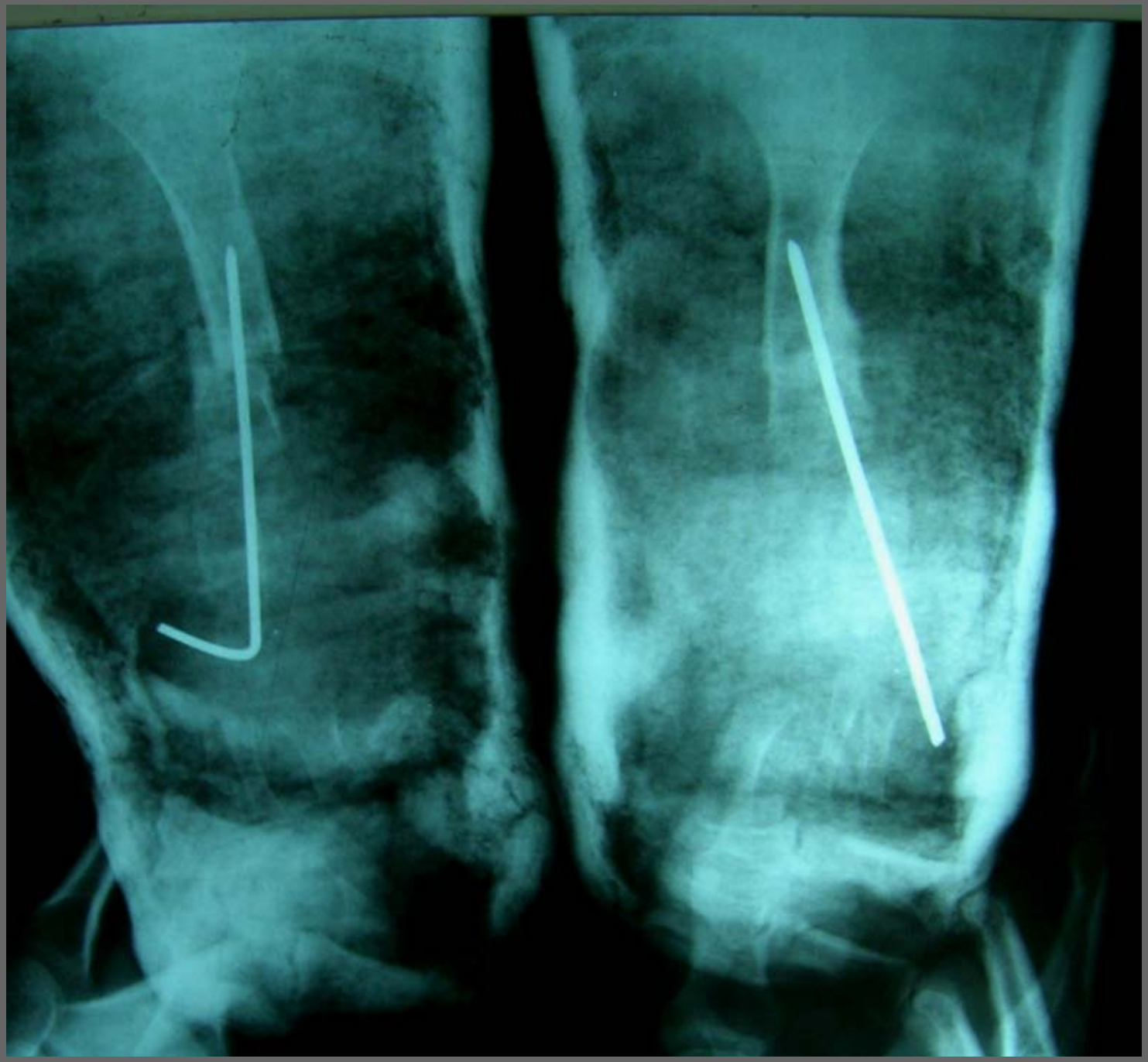
¿ Reconstruir Vs. Amputar ?

TIPO II



Edad 5,5 años
3 años pop





- MUCHAS GRACIAS