

Contents lists available at [ScienceDirect](#)

Acta Orthopaedica et Traumatologica Turcica

journal homepage: <https://www.elsevier.com/locate/aott>

A rare case of tibial hemimelia, surgical technique and clinical results

Marco Basso ^{a, *}, Valentina Camurri ^a, Piero Frediani ^b, Silvio Boero ^c^a Clinica Ortopedica – IRCCS Azienda Ospedaliera Universitaria San Martino – IST, Istituto Nazionale per la Ricerca sul Cancro, Genova, Italy^b Clinica S. Gaudenzio di Novara, Milano, Italy^c UOC Ortopedia – Ospedale Pediatrico Gaslini, Genova, Italy

ARTICLE INFO

Article history:

Received 28 February 2017

Received in revised form

10 July 2017

Accepted 12 November 2017

Available online xxx

Keywords:

Tibial hemimelia

Leg reconstruction

Correction of pediatric deformity

Paley classification

Weber classification

Jones classification

ABSTRACT

We report a nine-year-old boy with a type IIIa tibial hemimelia, according to the new Paley classification. We describe the x-ray findings, the surgical treatment technique, and the prognostic course of the patient. Descriptions of such cases are very infrequent in the literature and type of treatment is still object of debate.

© 2017 Turkish Association of Orthopaedics and Traumatology. Publishing services by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Introduction

Tibial hemimelia (TH) is a rare condition with a wide range of clinical presentations, ranging from a hypoplastic tibia to complete deficiency of tibia.¹ The incidence of TH is 1 in 1,000,000 live births.² Several authors tried to classify this pathology, they attempted to recommend proper surgical options for each types of TH.³ In 1861, Billroth firstly described the “tibial hemimelia”.⁴ This description was then modified by Dankmeijer in 1935.⁵ TH can occur for an autosomal dominant or recessive transmission.^{6,7} TH is associated with several syndromes such as Werner's syndrome,⁸ Langer–Giedion syndrome, tricho–rhino–phalangeal syndrome (TRPS II),^{9,10} tibial hemimelia diplopodia syndrome,¹¹ tibial hemimelia and split hand and foot syndrome¹² and tibial hemimelia micromelia trigonal brachycephaly syndrome.¹³ Other known potential cause is mother assumption of talidomide during pregnancy. Weber described a classification system for TH and recently Paley has performed a more accurate evaluation system.^{3,14} All these classifications are useful to recognize if tibial reduction defects or

cartilaginous anlage are present. Nevertheless, there is a lack evidence about the correct approaches according to the grade of TH. Therefore, types of treatment are usually demanding, even for expert surgeons. We present a case report, trying to discuss the anatomical findings associated with a rare type of TH.

Case report

A nine year old male presented with TH and clubfoot of the right leg (Fig. 1). According to Jones and Weber classifications, TH was a type II. More precisely there was a distal diastasis of the tibia and normal hip joint, normal femur, normal patella, dysplastic fibula, and normal muscle function (abbreviated *coI+/feI+/pal/tiII+/fiIII/pell*) with a score of 33 points according to Weber score system for TH. In addition fibula dysplasia was difficult to evaluate, in fact the third distal portion was thick as a tibia and articulated with talus (Fig. 2). On the other hand, the foot was internally rotated and equinus with a medial contracture. We considered this case as a

* Corresponding author. Clinica Ortopedica – IRCCS Azienda Ospedaliera Universitaria San Martino – IST, Istituto Nazionale per la Ricerca sul Cancro, Largo Rosanna Benzi, 10, 16132, Genova, GE, Italy. Fax: +39 010 5556763.

E-mail address: marco.basso24@gmail.com (M. Basso).

Peer review under responsibility of Turkish Association of Orthopaedics and Traumatology.

<https://doi.org/10.1016/j.aott.2017.11.004>

1017-995X/© 2017 Turkish Association of Orthopaedics and Traumatology. Publishing services by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



Fig. 1. Pre-operative clinical picture of right affected leg.



Fig. 2. Pre-operative X-Ray evaluation of the affected leg. Talus joint with distal fibula and distal tibia is posteriorly placed.

Paley III a TH. Before surgery, we tried to mobilize medial soft tissues with serial casting without satisfactory results.

Due to the particular clinical setting of deformity, we decided to treat the patient using two surgical approaches to the ankle. Using the antero-medial approach (Fig. 3), we removed 4 cm of distal tibia (Fig. 4) and we lengthen the Achilles tendon with a Z-plasty technique. Soft tissues between tibia and fibula were gently dissected and moved from the syndesmosis. After this surgical step, we performed an antero-lateral approach to resect the distal portion of fibula and the dome of talus, to better achieve a functional ankle position. Before stabilization of ankle in proper position with K-wire, a tibiofibular synostosis was carried out using two screws (Figs. 5 and 6). Alignment of talus in both projections and closure of tibiofibular diastasis were checked using fluoroscopy. Post-operative X-rays demonstrated a good functional alignment (Fig. 7). We protected correction of deformity with cast bracing for 3 months. Weight bearing was not allowed for the first 45 days.

Discussion

Several classifications for TH have been proposed. Jones in 1978 firstly published his classification based on X-ray findings. Thirty years later, a new classification system was introduced by Weber taking into account X-ray and cartilaginous anlage.^{3,15}

According to Jones classification, our case corresponds to type IV. Jones divided TH in four types, ranging from the most to the last deficient. Type IV is shortened tibia with distal tibia-fibular diastasis. Considering the Weber classification, our case is type 2, “distal diastasis” of tibia and fibula”. For this type of TH, the incidence according to Weber is 5%.³ Weber also introduced a score system for TH, ranging from zero to 39; the higher the score, the less impairment grade suffered from the patient. Scoring patient's type of TH, we obtained 33. Paley et al recently has noted classification gaps between Jones and Weber system. They evaluated 113 THs according to Jones classification and they reported: 47 type Ia, 5 type Ib, 18 type II, 2 type III and 10 type IV. 31 cases (27.4%) were considered unclassifiable. On the other hand, following Weber classification: 18 type I, 11 type II, 3 type IIIa, 17 type IIIb, zero type IVa, 2 type IVb, 5 type Va, zero Vb, VIa, VIb, 4 type VIIa and 47 type VIIb. Using this system, only six cases (5.3%) were evaluated as unclassifiable. On this basis, Paley has introduced a new classification system developed to clarify treatment options and related prognosis.¹⁴ There are 5 types and 11 subtypes with modifiers to better represent TH associated deficiencies or duplications. Following Paley classification, our case is a type IIIa, reporting a deficiency of tibial plafond, medial and lateral malleolus still present, varus bowing tibia, relative fibular overgrowth and foot internally rotated with talus positioned between the tibia and fibula centered under the fibula. Paley found that if TH is classified using his system no unclassifiable cases are found.

Surgical treatments for TH are various. Depending on type of TH, surgical management could vary from different approaches such as amputation, leg reconstruction and correction of deformity, ankle arthrodesis, tendons lengthening or transpositions.^{6,16} In 1965, Brown published a new procedure for Jones type I providing fibular centralization.¹⁷ Observing his results, Brown introduced transposition of patellar tendon to the fibula and femoral shortening if needed to allow knee extension.¹⁸ Nevertheless, TH treatment is often challenging also for trained surgeons and frequently linked to poor results.¹⁹ Most surgeons think that through-knee amputation for Jones type I and below-knee amputation for other Jones types

Download English Version:

<https://daneshyari.com/en/article/8958576>

Download Persian Version:

<https://daneshyari.com/article/8958576>

[Daneshyari.com](https://daneshyari.com)