# Limb reconstruction versus amputation in treatment of fibular and tibial hemimelia

Essay

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By

*Mohamed Kamel Abdrabou*
( MB.B.Ch. )

Under supervision of

**Professor Dr: Tarek Hassan Abdelaziz**
Professor of Orthopaedic Surgery
Faculty of Medicine
Ain Shams University

**Dr: Mohamed Sobhy**
Lecturer of Orthopaedic Surgery

Faculty of Medicine
Ain Shams University

Faculty of Medicine
Ain Shams University

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قَالُوا سُبْحَانَكَ لاَ عِلْمَ لَنَا إِلاَّ مَا عَلَّمْتَنَا
إِنَّكَ أَنتَ الْعَلِيمُ الْحَكِيمُ"

صدق الله العظيم

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*First of all, I wish to express my sincere thanks to* ***ALLAH*** *for his care and generosity throughout my life.*

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## Aim of the study

The aim of this study is to review the literature and to help decision making regarding the two main modalities of treatment of fibular and tibial hemimelias: amputation versus limb reconstruction.

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# INTRODUCTION

Fibular hemimelia is a longitudinal deficiency of the fibula in which there is either partial or complete absence of the fibula.[1]

It is the most common deficiency of long bones that was first described by **Gollier** 1698.[1]

Tibial hemimelia is a longitudinal deficiency of the tibia in which there is either a complete or partial absence of the tibia.[2]

The term ***melia*** is derived from the Greek Melos; meaning limb. ***Amelia*** denotes absence of the entire limb. ***Hemimelia*** refers to longitudinal loss of half of the limb.[3]

Children with congenital deficiency of the fibula may have significant shortening of the involved extremity leading to limb length discrepancy (LLD) which is the main problem of fibular deficiency. [4,5]

Fibular hemimelia syndrome includes a spectrum of problems rather than LLD including variable degrees of fibular hypoplasia, shortening of the tibia and femur, genu valgum , lateral femoral condyle hypoplasia, knee ligament laxity, tibial bowing, ball and socket ankle joint, tarsal coalitions, clubfoot and missing lateral rays of the foot.[6,7,8]

Kruger and Talbott 1961, noted that femurs are shortened in all patients with fibular hemimelia with five rayed feet and 50% in those with fewer rays.[9]

In patients with tibial hemimelia approximately 75% of all patients have associated skeletal anomalies; Developmental dysplasia of hip,lobster hand,knee flexion deformity,knee instability, fixed equinovarus and absence of one or more medial rays.[10]

The exact cause of limb deficiency is unknown but there are at least three ways in which limb deficiencies can be caused:

- Errors in the genetic control of limb development.
- Disruption of the developing arterial supply.
- Teratogenic.[11]

**Fibular hemimelia classifications have been described by many authors:**

- **Coventry** and **Johnson** classification (1952) .
- **Frantz** and **O'Rahilly** classification (1961) .
- **Achterman** and **Kalamchi** classification (1979) .
- **Catagni** classification (1991) .
- **Birch** ( Functional ) classification (1998) .
- **Stanitski** and **Stanitski** (2003) .
- **Paley** classification (2004) .

**Tibial hemimelia classifications have been also described by many authors:**

- **Frantz** and **O'Rahilly** (1961).
- **Jones** (1978).
- **Weber** (2008).

The main problems in the treatment of lower limb deficiency are the limb-length discrepancy ,deformity and instability of the foot and ankle. It is very important to realize that the discrepancy will become worse with growth and it is the ultimate discrepancy at maturity that is important.[12]

Limb reconstruction is one of the two main modalities in treatment of both tibial and fibular hemimelia.
In tibial hemimelia Brown`s procedure is widely used.[13]
while in fibular hemimelia many procedures can be done:

- Ankle/foot reconstruction[14].
- Transphyseal osteotomy (Gruca operation) [15].
- Transphyseal osteotomy for ankle valgus correction.[16]
- Bending osteotomy through distal tibial physis. [17]

Successful management aims to restore normal weight-bearing and normal limb length so that the patient can walk with normal gait as possible. In mild cases, treatment includes shoe-raises, step-in prostheses, epiphysiodeses or limb-lengthening procedures and correction of knee, ankle and foot deformities. For more severe deformities the management is controversial.[5]
Many authors recommended early amputation of the foot and prosthetic rehabilitation.The advantages of this approach is a single surgical procedure, a short hospital stay, and immediate walking and equalization of limb length so that the child can adapt quickly and lead a normal life. The major disadvantages are that amputation is irreversible and that prosthetic limbs require periodic replacement and cannot provide normal sensation and proprioception.[5]

The introduction of the Ilizarov method of limb lengthening has provided an attractive alternative to amputation. Some surgeons prefer this method because it preserves the foot and can provide simultaneous correction of limb-length discrepancy and limb deformities. The major disadvantages are the multiple and unpredictable operations needed, often including two or three separate lengthenings during growth and the prolonged stay in hospital and in rehabilitation which may have a considerable psychosocial impact on the patient and family. Moreover, these procedures sometimes fail to achieve satisfactory or cosmetically acceptable results and may end in amputation.[18]

The parents who are the decision makers are weighing the hope for their child to retain the limb against what will entail. They most likely have never seen a child or adult with an amputation, they visualize something horrible. At the same time they cannot really know what a lengthened limb will be like at the end of treatment; they imagine the limb will be normal. Although they may understand that they will need two or three lengthening procedures, they cannot know what the impact will be on their child or their family, what the complications they will encounter on the way, or how their child will look or function at the end of the treatment.[12]

Studies that compare these two treatment options are not conclusive because there are frequent advances in each treatment option (especially limb reconstruction) which change and improve the results of each option, hence making older comparative studies irrelevant.[4]

## DEVELOPMENT OF LIMBS

Normal development of the limbs begins at the end of the fourth week after fertilization with buds forming in mesoderm along the flank of the embryo (Fig.1). The limbs develop in a proximodistal direction from the limb girdle to the digits. The proximal bones of the limb girdle and the humerus/femur form prior to the differentiation of ridge ectoderm, while development of the remaining bones and digits depends upon the apical ectodermal ridge (AER)(Fig. 2). [19]

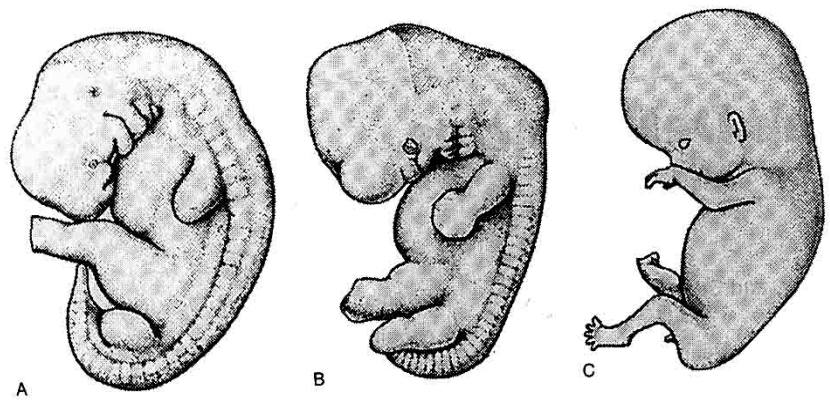
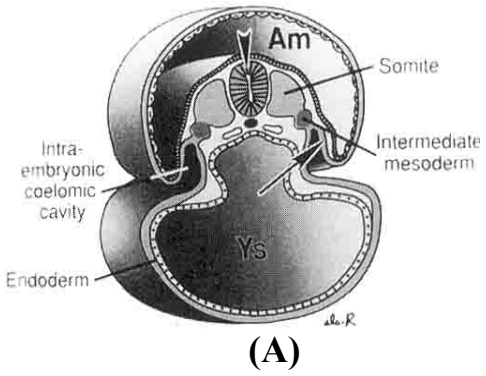
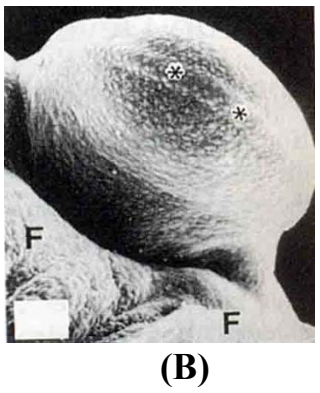



**Fig.(1):**
Development of the limb buds in human embryos. A:At 5 weeks,B:At 6 weeks and C:At 8 weeks. The hindlimb buds are less well developed than those of the forelimbs. [20]

This ridge exerts an inductive influence on adjacent mesenchyme causing it to remain as a population of undifferentiated rapidly proliferating cells, the progress zone. As the limb grows cells farther from the influence of the AER begin to differentiate into cartilage and muscle. In this manner development of the limb proceeds proximodistally.[20]




(A)

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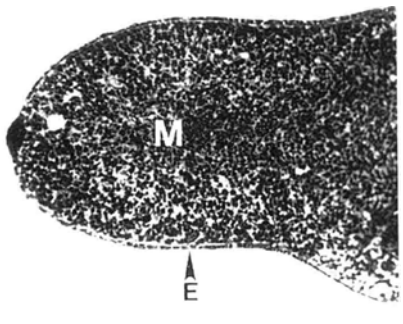


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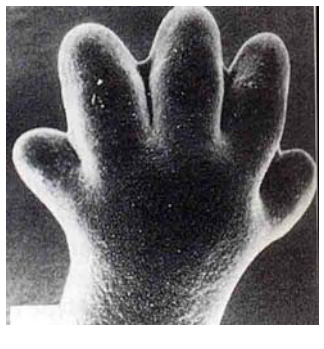

(D)

**Fig. (2):**

A : Cross section through an embryo. The intermediate mesoderm signals the lateral plate mesoderm to initiate limb development. (Am:amnion; Ys:yolk sac).

B : The early limb bud. Asterisks indicate the location of the apical ectodermal ridge, which regulates proximodistal growth of the limb bud. (F:flank).

C : Longitudinal section of the limb bud showing the ectoderm (E) surrounding a core of undifferentiated mesoderm (M).

D : Digits are forming following programmed cell death in the AER in the spaces between the digits, each digit then continues to grow under the influence of its own AER**.** [19]