RECENT TRENDS IN MANAGEMENT OF FIBULAR HEMIMELIA

Essay

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

صدق الله العظيم

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الطرق المستحدثة في علاج انعدام النصف الشظوى للطرف السفلى

رسالة

توطئة للحصول على درجة الماجستير في جراحة العظام

مقدمة من

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List of Abbreviations

ACL	: Anterior cruciate ligament
AER	: Apical ectodermal ridge
CFD	: Congenital fibular deficiency
K-Wires	: Kirschner wires
LCL	: Lateral collateral ligament
LLD	: Limb length discrepancy
PCL	: Posterior cruciate ligament
PETS	: Percutaneous ephysiodesis using transphyseal screws
PFFD	: Proximal femoral focal deficiency
STR	: Soft tissue release
TEV	: Talipus equinovarus
ZPA	: Zone of polarizing activity

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INTRODUCTION

Fibular hemimelia or postaxial lower limb hypoplasia is the most frequent congenital long bone deficiency with an incidence of 5.7–20 per million live births.[1]

Fibular hemimelia is a longitudinal deficiency in which the tibia is usually short with an axis deviation (anterior bowing and valgus deformity).[2]

It was first described by Gollier in 1698[3]. ThetermmeliaisderivedfromtheGreekMelos;meaninglimb.Amelia denotes absence of the entire limb. Hemimelia refers to longitudinal loss of half of the limb.[4]

Fibular hemimelia is not an isolated anomaly but rather a spectrum of dysplasia of the limb. The clinical spectrum includes partial or complete absence of the fibula with variable other anomalies, including tibial shortening and angular deformities, ball and socket ankle, tarsal anomalies, absence of the lateral rays of the foot, femoral shortening, proximal femoral focal deficiency, and occasionally hand anomalies.[5]The major functional deficits are severe shortening of the extremity and equinovalgus deformity of the ankle, knee valgus deformity and flexion contracture, anteroposterior instability of the knee and ankle. [5,6]

In observations reported by Kruger et al.[7], the most common associated disorder is femoral shortening, found in 50% of cases.

These limb anomalies affect the ipsilateral limb in approximately 75% of all cases.[1]

It usually presents clinically as limb length discrepancy, anteromedial bowing of tibia, valgus deformity of the knee, equinovalgus deformities of the foot and ankle, and absent lateral rays.[8] The primary aim of treatment is to restore normal limb alignment and length with a stable plantigrade foot for the most effective gait. The two main surgical options for treatment are reconstructive or ablative surgery.[9]

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

- Errorsinthegeneticcontroloflimbdevelopment.
- Disruptionofthe developingarterial supply.
- Teratogenic.[10]

It is theorized that a defect in the femoral-fibular-ulnar developmental field can result in a clinical spectrum ranging from complete absence of the limb or proximal femoral focal deficiency to a missing toe and simple tarsal coalition. [11]

Several classifications were proposed to address the deformity, with the Achterman–Kalamchi [12] being the most commonly used.[13]

Treatment for mild cases includes lengthening by shoe (lift), walking prosthesis, epiphysiodesis or limb lengthening

procedures and correction of foot deformities. However, treatment for advanced deformities is controversial. [14]

Many surgeons support early amputation and rehabilitation by prosthesis of the foot.[14] With the development of modern methods of limb lengthening based on distraction osteogenesis and multiplane fixators, amputations have become less popular.[15,16]