Ankle reconstruction in fibular hemimelia

Essay

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BY

Mostafa Mahmoud Nagy Hassan

M.B.B.ch.

Supervised by

Prof. Dr. Salah Abd El Gawad Abou Seif

Professor of Orthopaedic Surgery

Faculty of medicine – Ain Shams University

Dr. Mahmoud Ali Mahran

Assistant Professor of Orthopaedic Surgery

Faculty of Medicine – Ain Shams University

Faculty of medicine

Ain Shams University

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Introduction

Fibular hemimelia is a congenital condition characterized by partial or total absence of the fibula. Rarely is it an isolated event, and deficiency of other long bones in the same limb is common. In fact, these other subtle deficiencies in the same limb have led to the consideration of the disease as being a 'syndrome' rather than a single long bone deficiency. Even the fibula itself shows a very wide spectrum of deficiency ranging from the almost unrecognizable deficiency where the fibula is present but mildly hypoplastic, to the complete fibular absence with marked foot deformity and severe limb shortening.

The fibula is the most commonly deficient long bone with an incidence of 1.07 per 10000 live births. Next in line is deficiency of the femur, the radius and the tibia.

The classic picture is that of a new born baby with a short limb, a tibia that is bowed anteriorly with a skin dimple at the apex of the bowing, an ankle thrust into a rigid equinovalgus position and a foot with missing two lateral rays.

As the child grows older, the discrepancy in length between the two limbs increases and the foot –if untreated- could not provide an adequate stable plantigrade fulcrum for

bearing weight. Hence, walking is difficult, with marked limping and an abnormal gait.

Prosthetic replacement to compensate for the discrepancy in length was the first logical solution to improve the gait and abolish limping. Amputation to allow better fitting and use of the prosthesis rapidly became popular and led to good results.

With advances in understanding of the deformity, its extent, its pathological background, and its natural history, new classification schemes were put down and a renewed interest in managing the problem surgically -but with conservation of the limb- emerged.

While there are still many advocates of early amputation and prosthetic fitting, a lot of specialists are now focusing on ankle and foot reconstruction to provide a stable plantigrade foot, and limb lengthening with or without contralateral epiphysiodesis to correct the limb length discrepancy. Authors are reporting good results with these techniques as well.

Whether fibular hemimelia is managed by amputation or by reconstruction and limb lengthening, it still is a tremendous challenge to the doctor, the parents and above all to the child himself.

Aim of the study

This essay will focus on ankle and foot reconstruction before lengthening in fibular hemimelia.

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List of Abbreviations

ACL	Anterior cruciate ligament.
ADTA	Anterior distal tibial angle.
CORA	Center of rotation and angulation.
CR	Computed radiography.
CT	Computed tomography.
FAH	Fibular aplasia or hypoplasia.
FFU	Femur fibula ulna complex.
FGF	Fibroblast growth factor.
HOX	Homeobox.
ISO	International Standards Organization.
LDTA	Lateral distal tibial angle.
LLD	Limb length discrepancy.
PCL	Posterior cruciate ligament.
PFFD	Proximal femoral focal deficiency.
STR	Soft tissue release.

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Definition

The term fibular deficiency implies a congenital absence of all or part of the fibula (1). The more common name fibular hemimelia describes the same deformity. It is derived from the Greek *melos*, a limb: where hemimelia means absence of a large part (*hemi*, a half) of a limb. Hemimelia may be complete (when the entire half of a limb is absent) or partial (when a greater portion of the distal half is absent). Paraxial hemimelia is used to indicate absence of one border (*paraxial,* means beside the axis) of the distal half of the limb involved. Paraxial hemimelia may be preaxial (referring to the border that includes the thumb or big toe and which is cephalic in the embryo) or postaxial (referring to the opposite border that is caudal in the embryo). Hence, fibular hemimelia is a postaxial partial hemimelia. It is agreed by convention that hemimelia is named after the affected (absent) portion of the limb (2).

Frantz et al (2) introduced the term *longitudinal* deficiency to indicate a defect along one of the preaxial or postaxial borders of the limb as opposed to *transverse* deficiency which indicates total absence of a part of the limb. Fibular hemimelia is thus a longitudinal deficiency that may be *terminal* (with affection of the lateral foot rays)

or *intercalary* (if the foot is intact). It may be *complete* (with total absence of the fibula) or *incomplete* (when part of the fibula is intact).

In 1989 the International Standards Organization (ISO) published a 'Method of describing limb deficiencies present at birth (3). It uses English anatomical names on a simple anatomical basis dividing the deficiency into *transverse* or *longitudinal*. A missing segment maybe *partial* or *total*.Hence, a proximal deficiency of the femur with an absent fibula and a three-toed foot would be described as a femur partial upper third, fibula total, tarsus partial, rays four and five total. It is an excellent descriptive method that encompasses the deformity but is clumsy to use in normal clinical practice (4).

The condition is recognized as a syndrome rather than a single bone defect and the syndrome of fibular deficiency encompasses – in addition to fibular absence - a spectrum of abnormalities affecting the femur, knee, tibia, ankle and foot. In one form, there is mild shortening of the fibula, valgus knee with anteroposterior instability and an absent fifth toe. In another form, the fibula is absent, the tibia is shortened and bowed and the foot has missing rays and is in marked equinovalgus (1).

Epidemiology

Congenital absence of the fibula is a rare disease. It was first described by Gollier in 1698 (5). Froster and Baird (6) quoted an incidence of 1.07 per 10000 live births. Others reported an incidence ranging from 7.4 to 20 per million live births (7).

It maybe difficult to accurately assess the incidence of this disease because the presentation may be so subtle that it may pass unnoticed. A case with very mild shortening less than 1 cm and an absent fifth toe may not be included in epidemiological studies.

There is consensus however that fibular hemimelia is the commonest major limb deficiency. Next in order is deficiency of the femur, the radius, the tibia, the ulna and the humerus (8).

The sex incidence is almost 1:1 (8) but there are reports of a slight male preponderance (9,10) , and the ratio of unilateral to bilateral cases is 2:1 (8,9). In unilateral lesions, the affection was twice as common on the right side (8,10).

Etiology

The exact etiology of fibular hemimelia is still unknown, yet many theories have been proposed.

***The Amniotic Theory*:** (11)

Between the fifth and eighth week, pressure of a too-tightly fitting amnion or an amniotic band interferes with the development of the exposed fibula and the outer toes. The persistent lack of growth in the whole limb is probably a result of deficiency in vessels and nerves due to early pressure.

Growth Arrest Theory:

Ollerenshaw (12) challenged the previous theories. He argued against the trauma theory stating that violence that would cause such a defect would more likely have caused abortion. He also argued against the constriction ring or amniotic theory stating that it fails to explain the presence of the defect in one bone or one part of the limb without causing distal amputations. He proposed that the predominant cause is undoubtedly a cessation of growth in one or other set of embryonic cells at a period between the fourth and sixth week of foetal life. The cause of the arrest of growth is quite unknown.