Limb Lengthening in Management of Proximal Femoral Focal Deficiency

A Systematic Review of Literature

Essay

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LIST OF ABBREVIATIONS

PFFD	Proximal femoral focal deficiency
LLD	Leg length discrepancy
LRS	Limb reconstruction system
PODCI	Pediatrics Orthopedic Data Collection Instruments
DCT/cm	Distraction consolidation time of every centimeter
HI	Healing index

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Abstract

We conducted this systematic review to evaluate the effectiveness of methods used in lengthening in cases of proximal femoral focal deficiency and compare the outcome of lengthening using external fixators and lengthening using intramedullary motorized nail.

We conducted our study by searching Medline and PubMed for studies about lengthening in cases of PFFD and we find 4 papers about lengthening using different methods of lengthening done in 2014 to 2016 which met our inclusion and exclusion criteria.

Our specific outcome measures were Achieved lengthening, corrected limb length discrepancy, complications, Range of motion of surrounding joints.

Key words:

Proximal femoral focal deficiency, lengthening, External fixator, femur, intramedullary motorized nail

INTRODUCTION

Proximal femoral focal deficiency (PFFD) is a rare congenital anomaly occurs in 1 of every 52.000 live births.[1] PFFD range from a mild shortening of the femur up to an almost absence of this bone. The most frequent form consists of a partial bone defect of the proximal femur with variable involvement of the hip joint. Most patients affected by PFFD, especially those bilaterally involved, present with other associated anomalies such as fibular hemimelia, absence of cruciate ligaments of the knee, club foot, spinal deformities, congenital cardiopathy and facial dysplasia.[2]

PFFD has a common clinical picture where the thigh is short, flexed, abducted and externally rotated. It also shows a severe apparent leg – length discrepancy (LLD), there may be also significant instability of the hip joint and in the knee joint due to absence of cruciate ligaments.[3]

Radiographic evaluation of PFFD depends on[5]:

- **X ray imaging:** a long anteroposterior and lateral film where both legs are pulled down
- **Ultrasonography:** to detect the presence of the head
- **MRI imaging:** helps in determining whether there is pseudoarthrosis or not
- **CT imaging:** only used in older age

Classification systems for PFFD[4,5]:

1. Aitken's classification consists of class A, class B, class C, class D as illustrated in table (1) and figure (1).

Table (1): Aitken's classification of PFFD[4].

Aitken's classification		
Class	***Femoral Head***	***Acetabulum***
A	Present	Normal
B	Present	Mildly dysplastic
C	Absent	Severely dysplastic
D	Absent	Absent

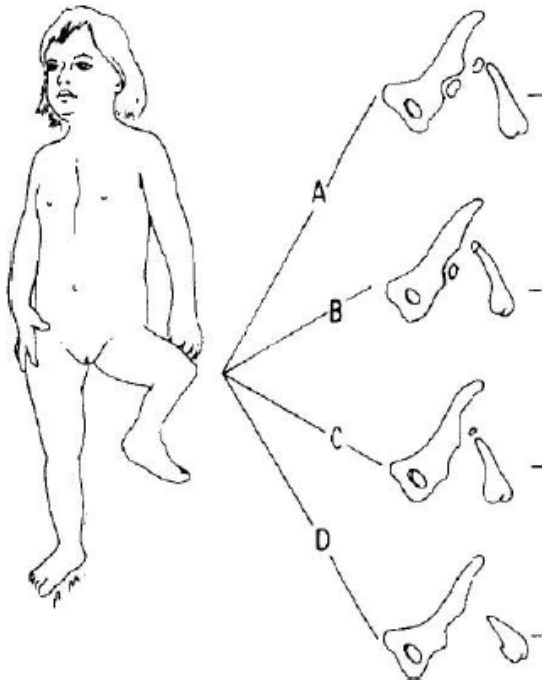

Figure (1): *Aitken's classification of Proximal femoral focal deficiency.*[4]

2. Paley's classification[5] (figure 2) :

Type 1: "intact femur" with mobile hip and knee

a. Normal ossification proximal femur

b. Delayed ossification proximal femur

Type 2: "Mobile pseudarthrosis" with mobile knee

a. Femoral head mobile in acetabulum

b. Femoral head absent or stiff in acetabulum

Type 3: "Diaphyseal deficiency" of femur

a. Knee motion 45° or more

b. Knee motion less than 45°

c. complete absence of femur

Type 4: "Distal deficiency" of femur

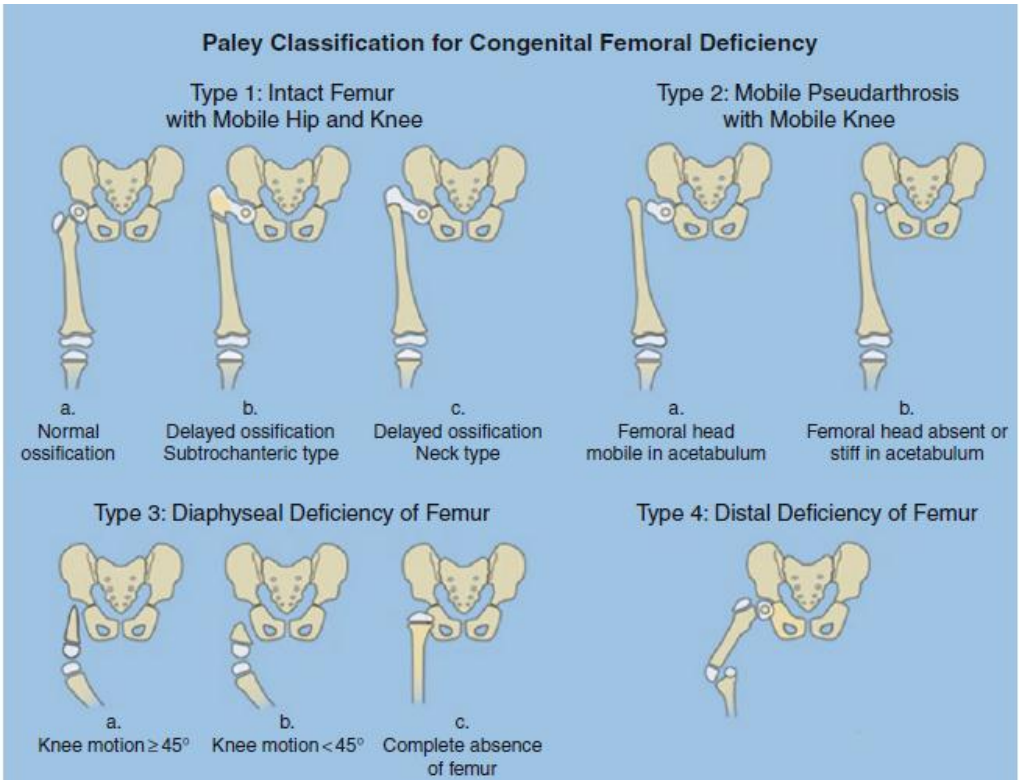

Figure (2): *Paley's classification of congenital femoral deficiency.*[5]

Treatment for a PFFD patient depends mainly on limb length discrepancy, bilaterality or unilaterality, presence of functional knee and foot, severity degree[3].

For a child with a predicted discrepancy of >20 cm, treatment is typically about facilitating and enhance prosthetic fitting. Options might include prosthetic fitting only, ankle disarticulation or modified Boyd amputation, knee fusion, rotationplasty, or femoro-pelvic arthrodesis[5].

If the predicted discrepancy is <20 cm, treatment may be through limb lengthening strategies[5].

For lengthening to be successful there are certain prerequisites[5]:

- Predicted discrepancy not exceeding 20 cm.
- Stable hip or hip that could be stable by surgeries
- Functional knee, foot and ankle.

Lengthening in congenitally short femur is done by means of distraction osteogenesis either by using external fixation system or intramedullary motorized nail[6].It depends on gradual distraction of bone leading to

formation of new bone in the distraction gap. Lengthening passes by three phases, the first phase is latency phase which resembles early fracture healing, the second phase is distraction and the third one is consolidation which occurs at the end of distraction and in which the distraction gap has healed in a way permitting removal of the external fixator[7].

The intramedullary motorized nail is an intramedullary nail that is capable of 40 mm – 80 mm of distraction according to the manufacturer. The motorized nail contain electric spindle motor and a subcutaneous antenna which receives impulses through a transmitter[11].

Lengthening is challenging and may be associated with many complications, such as pin tract infections, residual leg length discrepancy, delayed union, ankle and knee stiffness, re-fractures, joint subluxation/dislocation, residual sagittal or coronal deformity and psychosocial disturbances.

Surgical technique of lengthening

There are two main steps in lengthening procedure which is:

1- Femoral osteotomy which can be done percutaneous or open through anterolateral approach, it is usually done by drilling holes in femur by a 3.2 mm drill bit and completion of the osteotomy with an osteotome, the osteotomy is mostly a distal femoral osteotomy but in cases of lengthening over intramedullary nail a proximal osteotomy may be used[8].

2- Application of lengthening device which is either monolateral external fixator or circular fixator as in (figures 3,4) or motorized ante-grade intramedullary nail as in (figure 5).

Figure (3): *Showing gradual distraction done to lengthen a femur using external fixator*.[9]

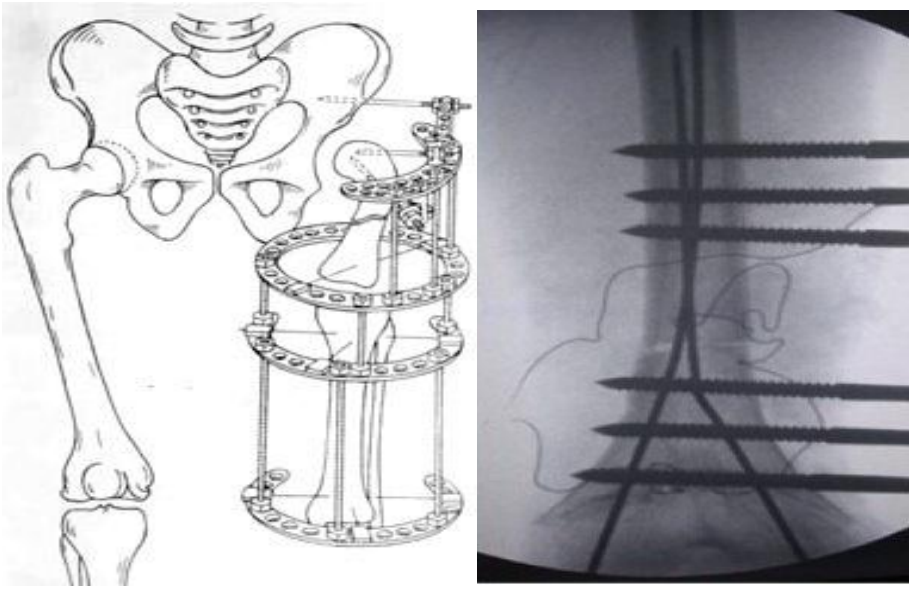

Figure (4): *Lengthening of femur using monolateral external fixator*[(8)] *or circular external fixator*

Figure (5): *Lengthening of femur using intramedullary motorized nail*[11]**.**