Congenital Femur Deficiency and Fibular Hemimelia – Amputation and Prosthetisation

Maria José Costa, Francisco Tavares
Physical Medicine and Rehabilitation
Hospital de Dona Estefânia
Lisboa – Portugal
Pediatric Amputations

• About 70% of pediatric amputee are congenital limb deficiencies:
  – Fibular Hemimelia (1 / 40,000) (1)
  – Congenital Femur Deficiency (1 / 50,000 – 200,000) (2)

• 40% of the children with congenital limb deficiencies will have multiple limb involvement (3)

(1) Limb Reconstruction Surgery for Fibular Hemimelia; Dror Paley
Pediatric Amputations
ISPO classification

- Transverse Deficiencies
  - Easy decision prothetisation

- Longitudinal Deficiencies
  - Complex problem
  - Multiple therapeutic options
  - Lower limb most frequent:
    - Fibular Hemimelia
    - Congenital Femur Deficiency
Fibular Hemimelia

• Associated with other deformities:
  – foot ray deficiency
  – sub-talar coalition
  – ball and socket ankle joint
  – ankle joint malorientation
  – diaphyseal angular deformity apex anteromedial
  – fibular deficiency
  – congenital shortening of the tibia
  – femural malformation (coxa vara, CFD, lateral femural condylo hypoplastic,...)
Fibular Hemimelia

- Clinical presentation:
  - Shorter limb (below knee)
  - Frequently with foot deformity
  - Possible ankle and knee instability
Congenital Femur Deficiency

• Spectrum of severity of femoral deficiency and deformity:
  – Lack of integrity, stability and mobility of hip and knee joints
  – Bone malorientation, bone malrotation
  – Soft tissue contractures of the hip and knee
Congenital Femur Deficiency

Clinical Presentation:

– Shorter lower limb (above knee)
– Knee apparently inexistenent or in proximal position
– Usually normal foot
CFD and FH – What they have in common?

• Abnormal stand position and gait
  – Dismetry
  – Deformity
Pediatric Amputation

- Primary goal: maximize function

- Approach based on:
  - Preservation limb length and growth plates
  - Disarticulation
  - Proximal portion stabilization/normalization
Level of Amputation – FH

- In Fibular Hemimelia:
  - Incapacity to have a plantigrad foot
  - Unstable ankle

Syme’s amputation
Elective Surgery for CFD

- Varies according to Paley’s classification

Types:
- Type 1a
- Type 1b
- Type 2a
- Type 2b
- Type 3a
- Type 3b
- Type 3c
- Type 4
Elective Surgery for CFD

• Rotationplasty

• Syme’s amputation
  – With or without knee fusion
CFD Prothetisation

- Some patients refuse chirurgical treatment
  - Requiring non-standard prosthesis
Differences between children's and adults amputations

- No sense of loss

Prosthesis: Assistive device
When to amputate?

• Before 2 years old
  – before the foot has become fully incorporated into the child’s body image
Prosthesis Fitting

- Between 6-12 months
  - Standing
  - Gait
Therapeutic training
Adequate psychomotor development
Energy Expenditure

• Through or below the knee amputation:
  – maintain a normal walking speed
  – without significantly increasing their energy

• Above the knee amputation:
  – significantly slower walk (72-80%)
  – elevated heart rate (124%) and energy cost (VO2 151%-161%)

• Bilateral BK amputation:
  – Slower velocity (87%)
  – Elevated heart rate (119%); similar energy cost

Follow-up

- Follow-up
  - 3-4 months intervals
  - Specially during growth spurts until maturity

- New lower limb prosthesis

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<thead>
<tr>
<th>Literature</th>
<th>Our experience – HDE</th>
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<td>&lt; 5 years</td>
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<td>5 to 12 years</td>
<td>Every other year</td>
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<td>13–21 years</td>
<td>Once every 3 years</td>
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<td>&gt;16 years</td>
<td>Every other year</td>
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Prosthesis Selection

• Age
  – Toddler:
    • Wide based, hip flexion and abduction, knee flexion, total foot initial contact
    • Prosthesis *without* knee component

• Function
  – Varies according to psychomotor development, personality and sports preferences
Prosthesis Components

Suspension and Sockets:

Knees:
- Monocentric
- Polycentric
- Mechanic
- Pneumatic
- Hydraulic

Foots:
- SACH
- Dynamic
- Flex Foot junior
Congenital Femur Deficiency and Fibular Hemimelia
Amputation and Prothetisation
Give children the prosthesis that works as hard as they play.