The Limb Deficient Child

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Classification

- Frantz-O’Rahilly / ISPO
- Complete/partial absence of bone segments
- Transverse deficiency (no distal segments)
- Longitudinal deficiency (some distal segments)
- Difficult to classify over/undergrowth, duplication and congenital bands
Congenital Limb Deficiency

- Incidence 4.1- 6.3/10,000 births
- Congenital : Acquired - 60:40
- Upper : Lower - 2:1
- 30% multiple limbs
- Male : Female - 1.8:1
- Seldom have associated diseases
- Usually normal intelligence
Genetic Considerations

- Most transverse defects have no genetic risks
- Tibial defects have the highest risk (30%)
- Renal defects associated with lower extremity deficiency
- Scoliosis (18%) and cardiac defects linked with upper extremity deficiency
Congenital

- Radial deficiency (most common)
- Fibular deficiency
- Tibial deficiency
- Femur deficiency
- Multi-limb deficiency (least common)
Can it happen again?

- **1-3% chance**
  (Slightly higher than normal)

- **Some genetic associations**
  (Tibial deficiency, Ulnar-femoral syndrome)
Decision Making

- Severity of deficiency
- Estimate limb length at maturity
- Amputation, lengthening, bracing?
- Timing of intervention (age of child)
- Psycho-social assessment
- Available expertise and finances
# Management

<table>
<thead>
<tr>
<th>LLD at Maturity</th>
<th>Rx</th>
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</thead>
<tbody>
<tr>
<td>&lt; 2 cm</td>
<td>? Shoe lift</td>
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<tr>
<td>2-5 cm</td>
<td>Epiphyseodesis or</td>
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<tr>
<td></td>
<td>Fem. shortening</td>
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<tr>
<td>5-20 cm</td>
<td>Lengthening</td>
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<tr>
<td>&gt; 20 cm</td>
<td>Prosthesis</td>
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</tbody>
</table>
Syme’s at 1 year
Syme’s at 12 years
When to fit Prosthesis?

- Match with child’s development
- Upper passive - sit ~ 4-6 mos
- Activate terminal device ~ 9-18 mos
- Activate elbow ~ 24-36 mos
- Lower non-articulated ~ 6-12 mos
- Lower articulated ~ 24-36 mos
Syme’s Amputation Prosthesis
Tibia/Fibula considerations
Fibular Deficiency

- Femoral shortening in ~50%
- 25% are bilateral
- Knee / Ankle / Foot anomalies
- Anteromedial tibial bow
- Equino-valgus foot
Fibular Deficiency
Limb lengthening if:

- Stable foot with >3 rays
- Plantigrade foot
- Stable / mobile ankle
- Predicted LLD <20 cm
- Multidisciplinary team
Fibular Deficiency
Conversion Amputation to Syme’s

- Unstable foot with <3 rays
- Unstable / stiff ankle
- Predicted LLD >20 cm
- Multidisciplinary team
Tibial Deficiency

- Jones classification (based on tibial length remaining)
- Can be genetic
- Knee flexed and unstable, ankle varus deformity
- Rx based on severity
Tibial Deficiency

Options

- Absent tibia = knee disarticulation
- Fibular centralization
- Proximal third = tibia-fibula fusion
- More than a third = syme’s
- Lengthening / reconstruction if foot and ankle are stable
Bilateral Tibial Deficiency
After Bilateral Knee Dis-articulations
Bilateral Congenital Limb Deficiency (video)
Congenital Femur Deficiency

- Femur / Acetabulum dysplastic
- Hip Flex / Abd / Ext rot.
- Hypoplastic lateral condyle
- Knee A-P laxity
- Fibular hypoplasia (~50%)
Longitudinal Femoral Deficiency Prosthesis
PFFD
Proximal Focal Femoral Deficiency
Congenital Femur Deficiency

Prosthetic Fitting

- Prosthetic Fitting > 50% deficiency
- Knee fusion + ankle disartic = knee disartic
- With VanNess Rotationplasty = Modified BKA
- Consider limb lengthening < 50% deficiency
Congenital Femur Deficiency

Rotationplasty

- Need mobile ankle (90 degrees PF)
- Gastroc-soleus = “knee extensor”
- Ankle at level of opposite knee
- Muscle strength 4/5 necessary
- Fitted as modified BKA on foot
VanNess Rotationplasty Prosthesis
Walk with Rotationplasty Prosthesis (video)
Partial Hand Deficiency
Partial Hand Deficiency
Radial Deficiency

- Transverse/partial deficiency most common
- Assess shoulder and elbow mobility
- Fit with mitt prosthesis by 6 months
- If longitudinal deficiency, consider limb salvage by centralizing wrist on ulna to preserve hand
Transverse Radial Deficiency
Comparison to normal
Transverse radial deficiency
Trans-radial Cable Prosthesis
Humeral Deficiency

- Transverse/partial deficiency most common
- Fit with non-articulated prosthesis by 6 months
- Articulate elbow by 36 months
- If longitudinal deficiency, consider limb salvage by lengthening (Illizarov)
Elbow Disarticulation Deficiency
Elbow Disartic Prostheses
Elbow Disartic Hybrid Prosthesis
Elbow Disartic Deficiency with Myo-electric Prosthesis
Kids are different, not disabled
Acquired Amputation

- Trauma (most common)
- Malignancy
- Infection- meningocococcus
- Vascular
- 60% in lower extremity
BKA due to vascular injury
Peds Trans-tib prosthesis with thigh corset
Prosthetic Replacement

- Replace prosthesis once a year from age 1 to 16 (or when growth stops).
- Occasionally the foot can be used for 2 years
- Try to plan for growth into prosthesis (pylon and socket)
- Frequent modifications are expected
Multi-Limb Deficiency

- Function with/without prosthesis
- Keep limbs / spine mobile
- Preserve feet and hands
- Adaptations for ADL’s
- Mobility versus “walking”
Multiple-Limb Amputee
(four limb deficiency)
Bilateral Trans-humeral and Trans-femoral amputations. Where do you start?
Do you fit prostheses?
How do you achieve independent self-care?
Thank You