

# 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

## LLD at Maturity

< 2 cm

2-5 cm

5-20 cm

> 20 cm

## Rx

? Shoe lift

Epiphyseodesis or  
Fem. shortening

Lengthening

Prosthesis

# 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

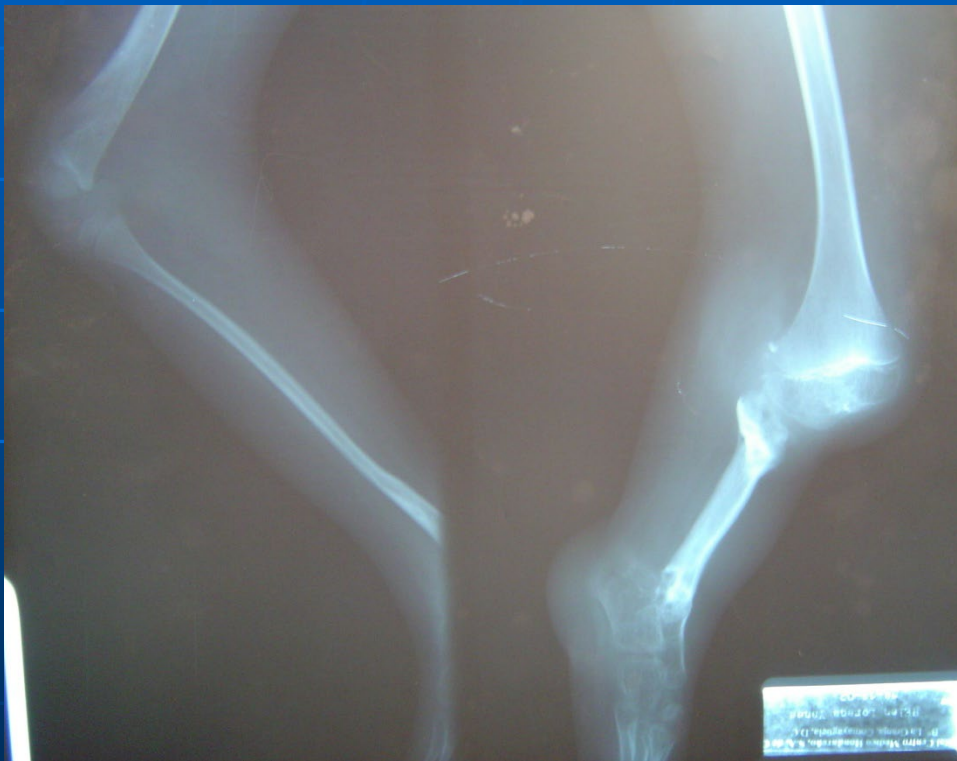
- Jones classification (based on tibial length remaining)
- Can be genetic
- Knee flexed and unstable, ankle varus deformity
- Rx based on severity

# 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



# PFFD Prosthesis



# 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

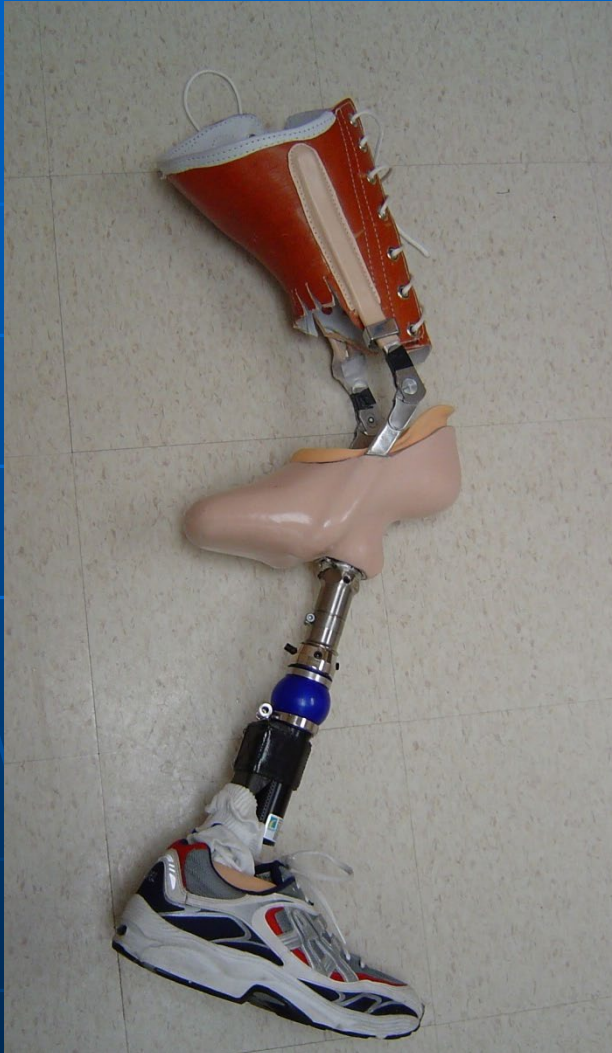
## Rotationplasty

- 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



# VanNess Rotationplasty Prosthesis



# Walk with Rotationplasty Prosthesis (video)





# Partial Hand Deficiency



# Partial Hand Deficiency



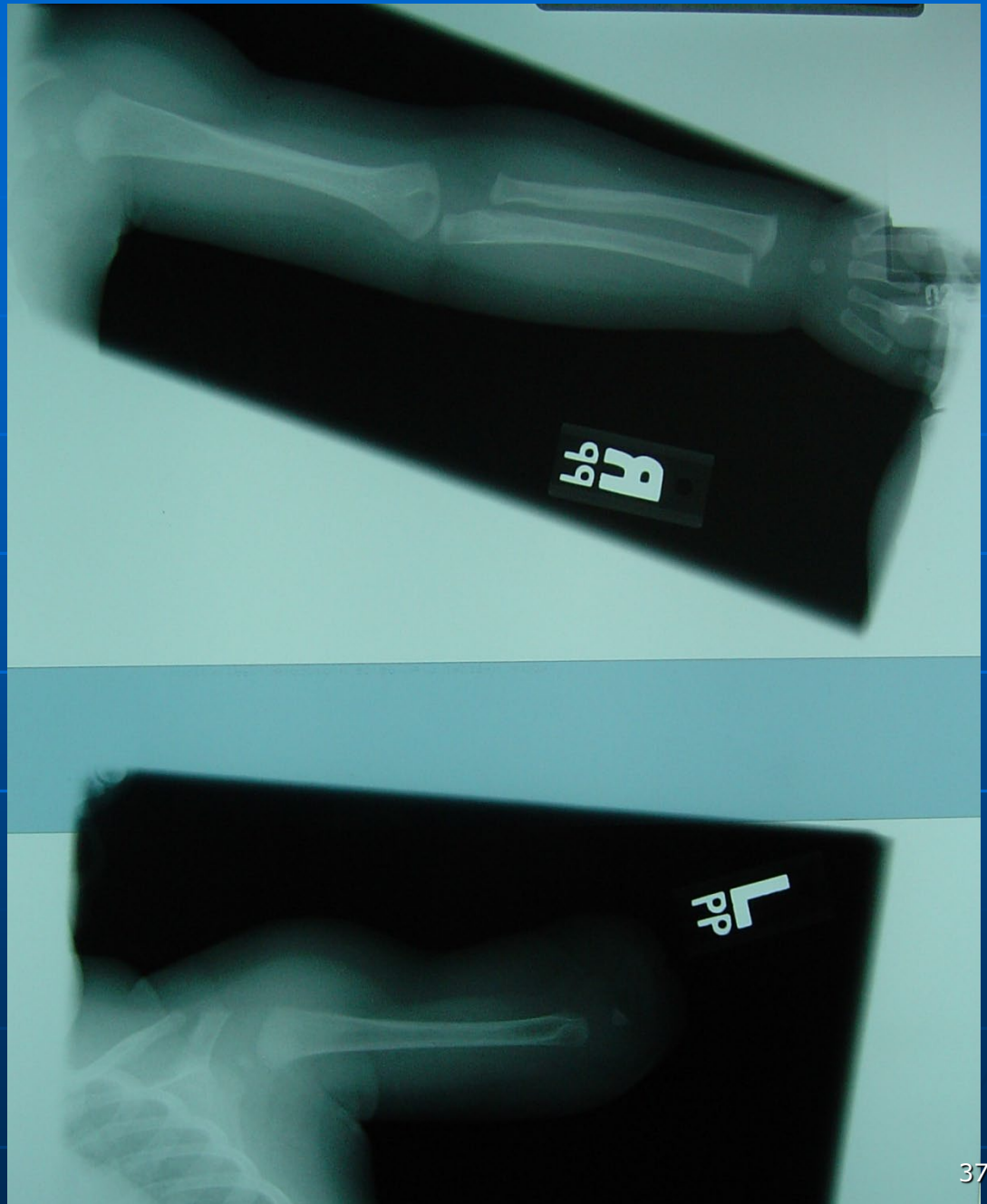
# Radial Deficiency

- 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

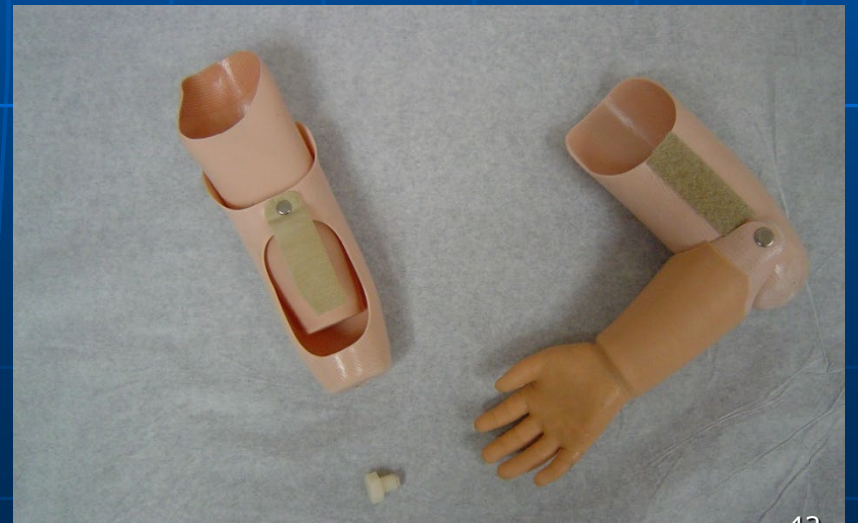
- Transverse/partial deficiency most common
- Fit with non-articulated prosthesis by 6 months
- Articulate elbow by 36 months
- If longitudinal deficiency, consider limb salvage by lengthening (Ilizarov)



# 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- meningococcus**
- **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**