

The Child with Lower Limb Deficiency

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Overview

- **Types**
- **Causes**
- **Child versus Adult**
- **Treatment Principles**
- **Specific levels / Prosthetic needs**

Classification

- **Frantz-O'Rahilly / ISPO**
- **Transverse deficiency (no distal segments)**
- **Longitudinal deficiency (some distal segments)**
- **Complete/partial absence of bone 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)**

Common Questions

- **Why did this happen ?**
- **When did this happen ?**
- **Can it happen again ?**
- **Whose fault ?**
- **What can be done ?**

Why did this happen ?

- **Mostly sporadic**
- **Drugs (Thalidomide)**
- **Amniotic Bands**
- **Genetic**
- **Vascular, maternal diabetes**
- **Infections (CMV, Rubella)**

When did this happen ?

Limb buds develop at 4-8 weeks, therefore insults at this time cause failure of formation. Many women are unaware of pregnancy at this time.

Can it happen again ?

- **1-3% chance**

(Slightly higher than normal)

- **Some genetic associations**

(Tibial deficiency, Ulnar-femoral syndrome)

Associated Anomalies

- **VATER (VACTERL)=multi-organ**
- **TAR=thrombocytopenia**
- **Holt-Oram=cardiac**
- **Fanconi=pancytopenia**

What can be done ?

- **Reassurance- Reinforce “Normalcy”**
- **Educate**
- **Team approach**
- **Introduce other kids/families**
- **Refer, if not comfortable**

Treatment Principles

- **Different, NOT disabled**
- **Build self-esteem**
- **Discuss all Rx options**
- **Individualize goals**
- **Timely Intervention**

Child ~~=~~ Adult Amputee

- **Congenital > Acquired**
- **More adaptable**
- **Changing development**
- **Changing size**
- **Different requirements**

Child \neq Adult Amputee

- **Changing prosthetic needs**
- **Skin more tolerant**
- **Lack of systemic illness**
- **Less careful with maintenance**
- **Stump “overgrowth”**

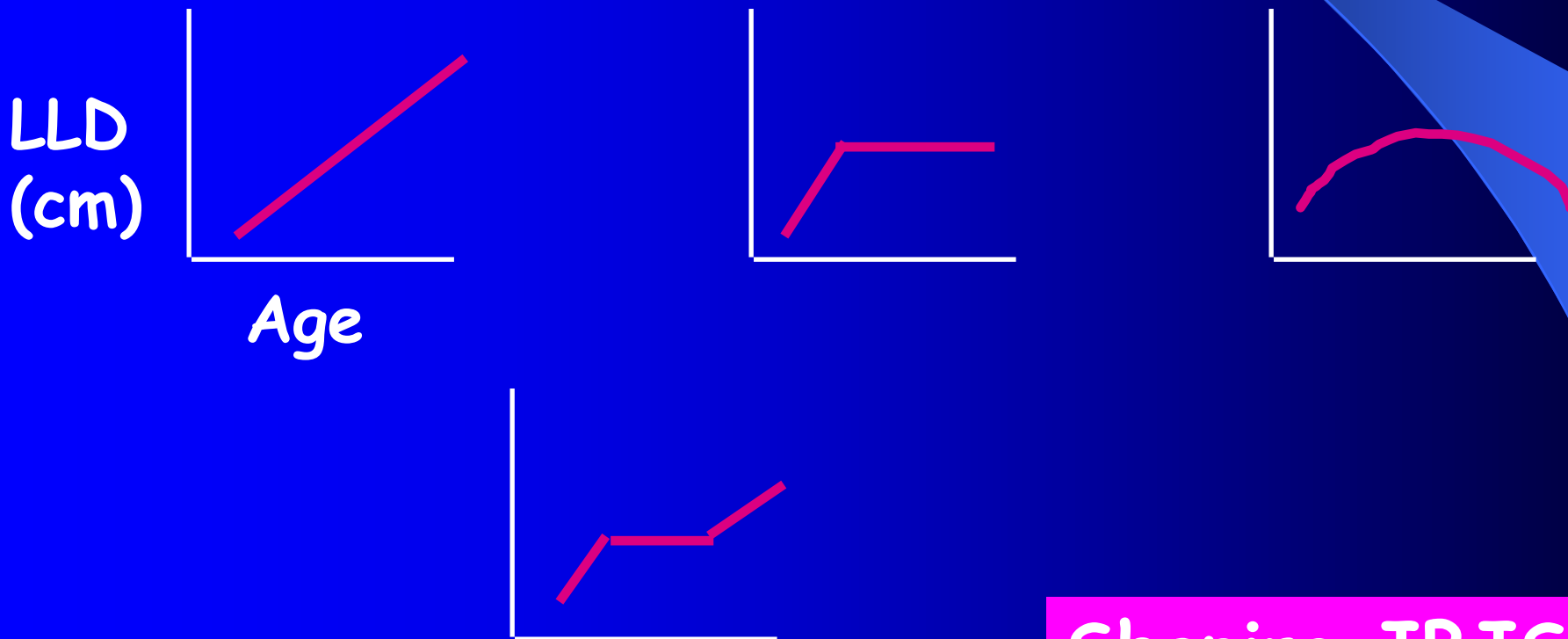
Decision Making

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

Limb Length Discrepancy

- **True / Apparent**
- **Congenital / Acquired**
- **Pattern of inhibition**
- **Bone segments involved**
- **Associated problems**

Patterns of LLD



Shapiro JBJS 1982

LLD at Maturity

● Birth	LLD x 4
● 1 yr.	LLD x 3
● 3 yr.(Girl)	LLD x 2
● 4 yr. (Boy)	LLD x 2

Compensatory Mechanisms

LLD

- **Equinus foot (lengthens limb)**
- **Knee flexion, contra-lateral (shortens)**
- **Pelvic tilt**

LLD: Consequences

- **Limp: increased energy, cadence**
- **Excessive wear opp. hip: ? DJD**
- **Scoliosis: controversial**

Management

LLD at Maturity

Rx

< 2 cm

? Shoe lift

2-5 cm

Epiphyseodesis or
Fem. shortening

5-20 cm

Lengthening

> 20 cm

Prosthesis

Syme's at 1 year



Syme's at 12 years



Amputation

- **“Conversion” amputation**
- **Site - bone versus joint**
- **Only 10% of UE need surgery**
- **Anticipate LLD at maturity**
- **Account for prosthetic needs**

Surgical intervention Goals

- **Maintain muscle dev. of limb**
- **Prevent progressive deformity**
- **Retain growth plates**
- **Preserve proximal joints**
- **Stabilize proximal joints**

Stump Overgrowth

- **Through bone amputations**
- **Appositional bone growth**
- **Humerus > Tib/fib > Femur**
- **Acquired > Congenital**
- **Prevention- Biologic “Capping”**

Heterotopic Bone Formation



Prosthetic Fitting Advantages

- **More functional ?**
- **Less psychological stress ?**

Prosthetic Fitting Disadvantages

- **Cover sensate areas**
- **Wear and tear (skin, joints)**
- **Nuisance factor**
- **“Loss” of body part**

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**

Fitting a Prosthesis

Goals

- **Optimum function**
- **Comfort**
- **Ease of use**
- **Longevity**
- **Cosmesis**

Partial Foot Deficiency



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**

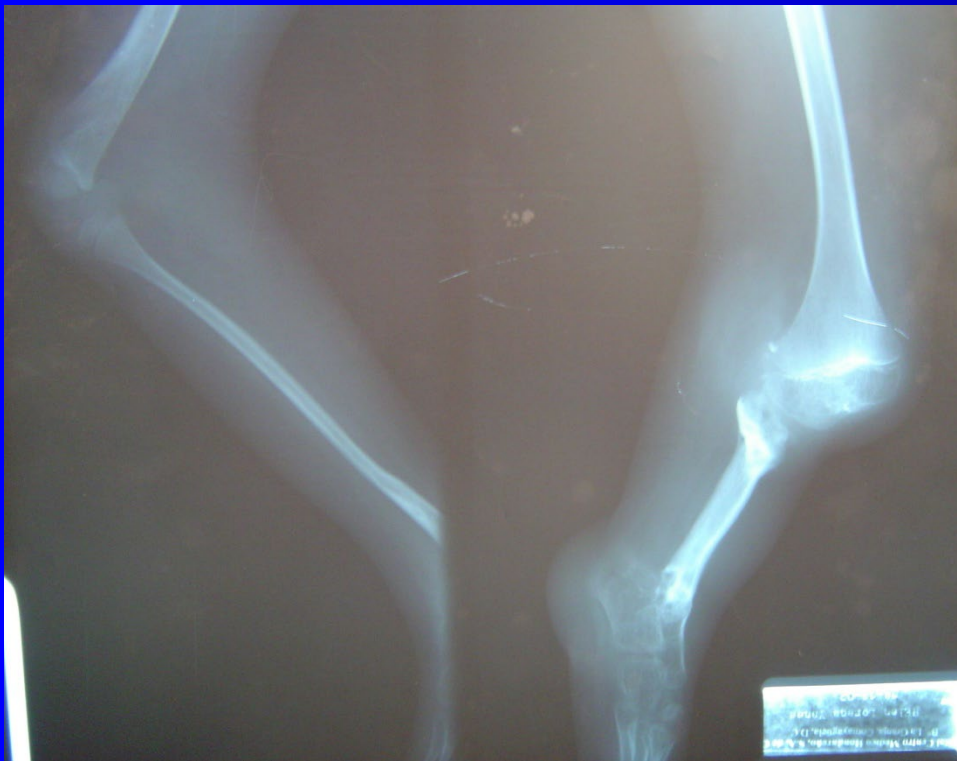
Tibial Deficiency

- **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



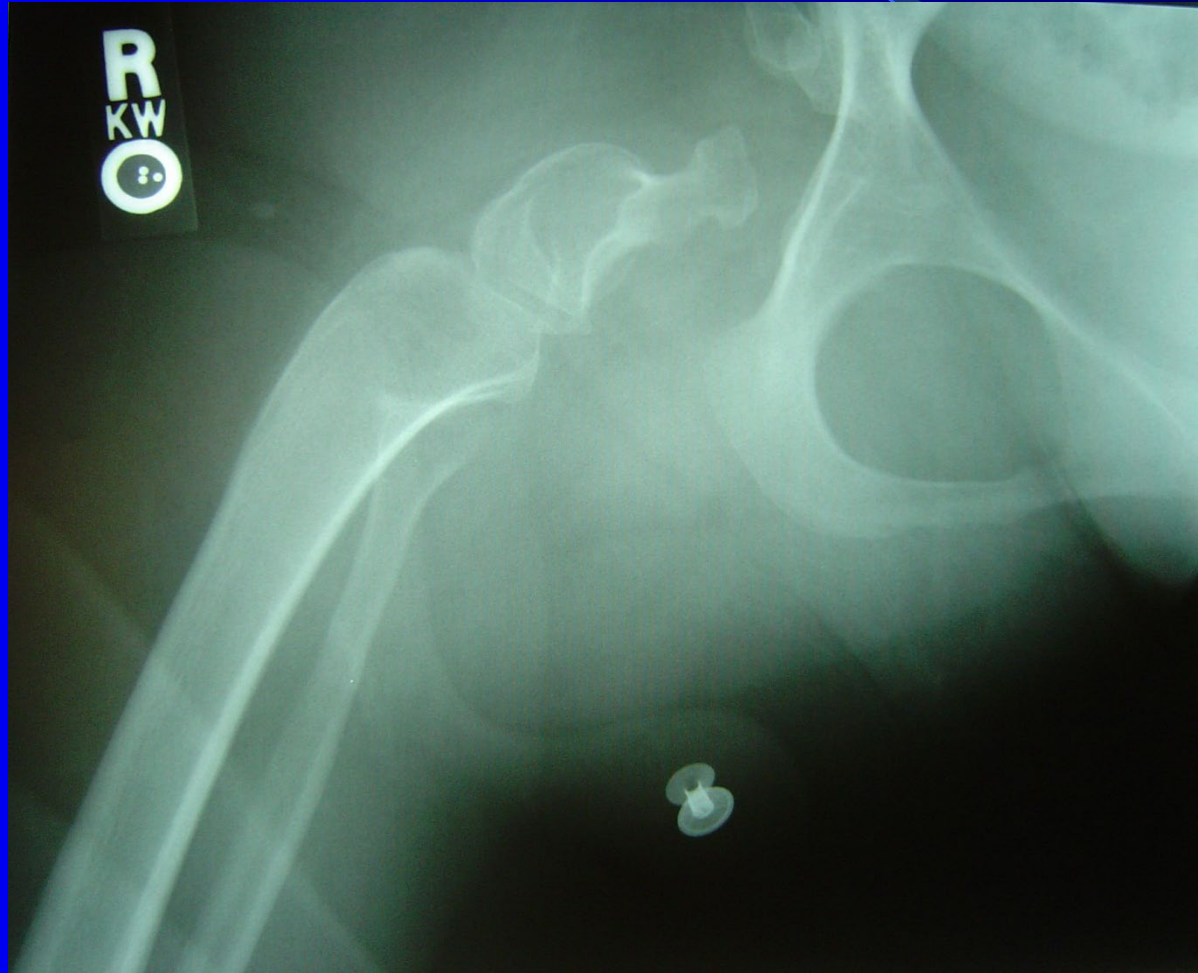
PFFD



PFFD Prosthesis



Proximal Focal Femoral Deficiency



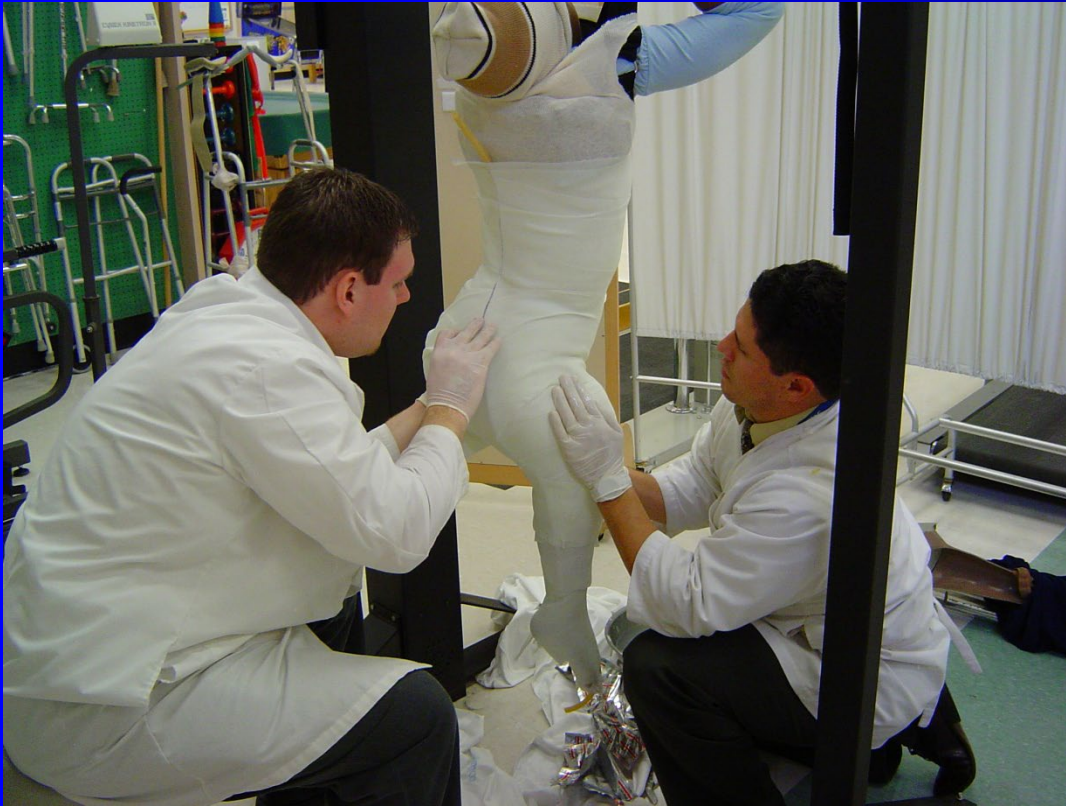
Congenital Hip Disarticulation



PFFD and Hip Disarticulation



Casting for new prostheses



Old 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**

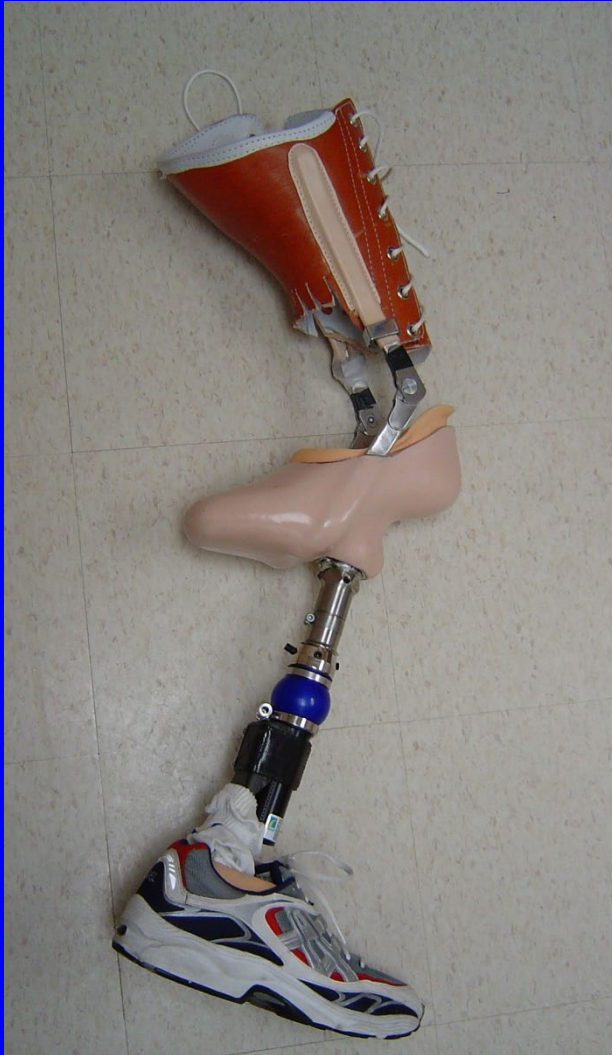
Congenital Femur Deficiency Rotationplasty

- **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)



Acquired Amputation

- **Trauma (most common)**
- **Malignancy**
- **Infection- meningococcus**
- **Vascular**
- **60% in lower extremity**

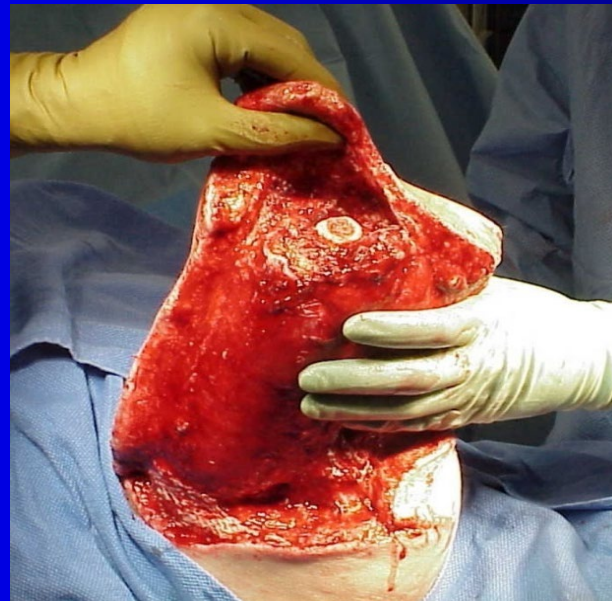
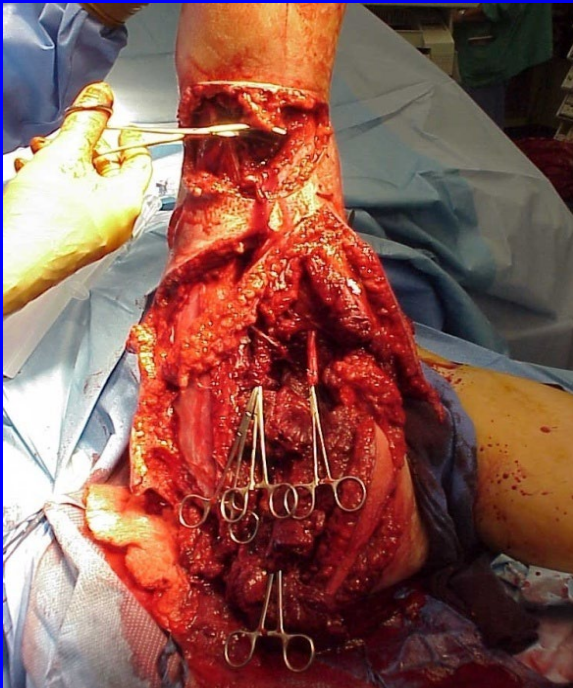
BKA due to vascular injury



Peds Trans-tib prosthesis with thigh corset



Traumatic amputation (start to finish, or just the beginning?)



Lumpy, Bumpy Limb



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

**Conjoined twin
separated at three
months**

(hemi-pelvectomy)



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)**



Kids are different, not disabled



Bilateral Trans-
humeral and
Trans-femoral
amputations.
Where do you
start ?



Bilateral Trans-femoral Amputee (video)



Thank You