Limb Lengthening in Children RSS and other etiologies

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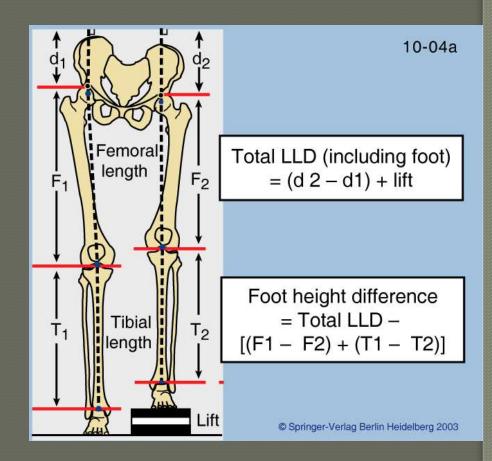




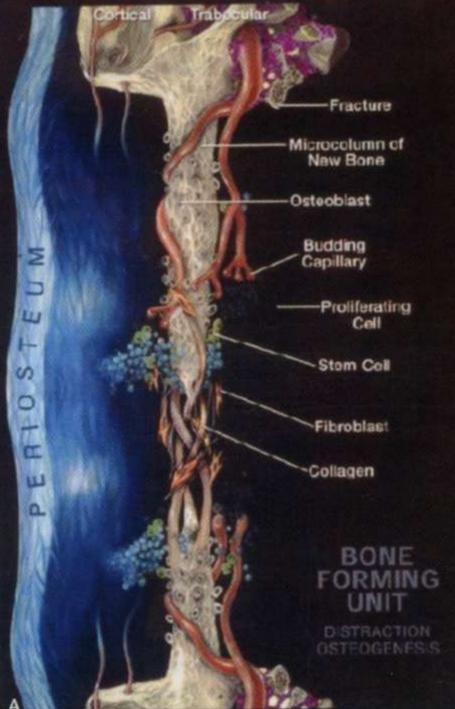
Evaluation

Causes of LLD

- Congenital
- Traumatic Growth arrest
- Tumor
- Overgrowth
- Neural inhibition









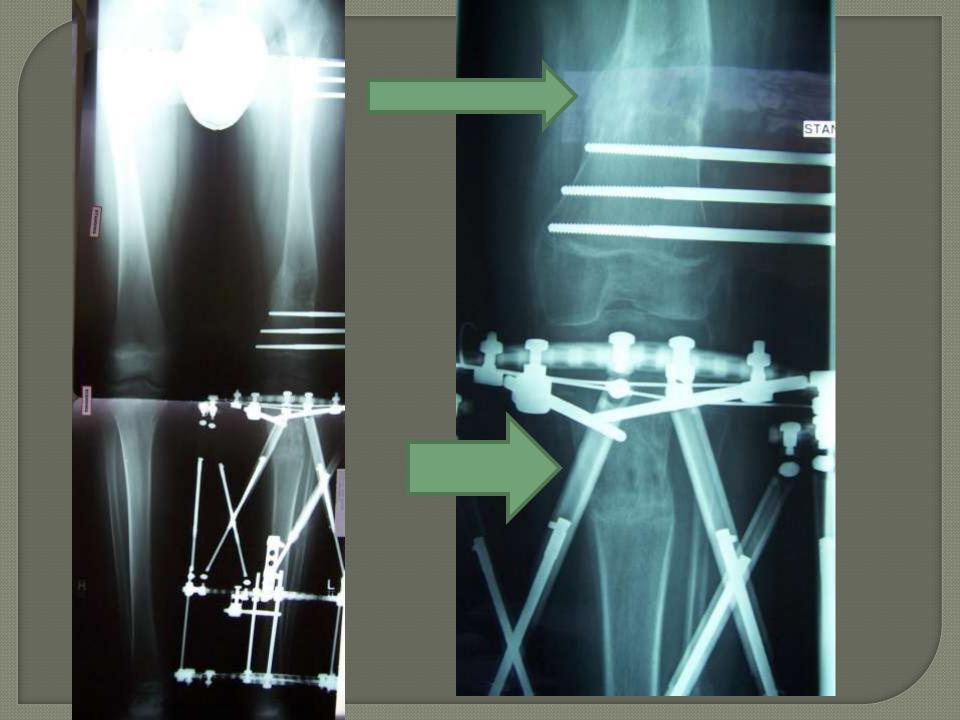


Hemiatrophy
Age 13
LLD 5 cm divided femur/tibia
Valgus deformity
Multiplier 1.12
PLLD = 5x1.12= 5.6 cm





Plan: correct valgus
2.8 cm femur
2.8 cm tibia
Overlengthen by 6 mm







Russell Silver Syndrome

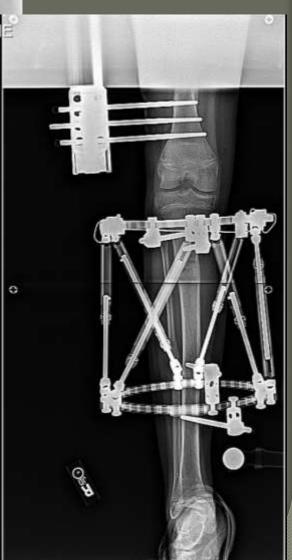
age 13
LLD 5 cm divided femur/tibia
M= 1.03
PLLD= 5.2 cm

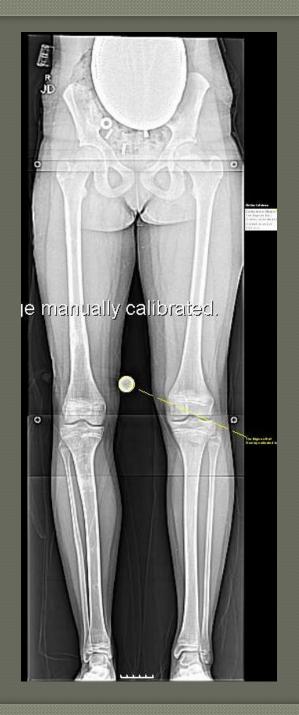
















RSS, age 8 LLD 4 cm divided femur/tibia M = 1.33

PLLD= 5.3



Puberty will be delayed and on HGH PLLD will be greater (6-7 cm)

Lengthen tibia 4 cm to correct LLD

Lengthen femur in future

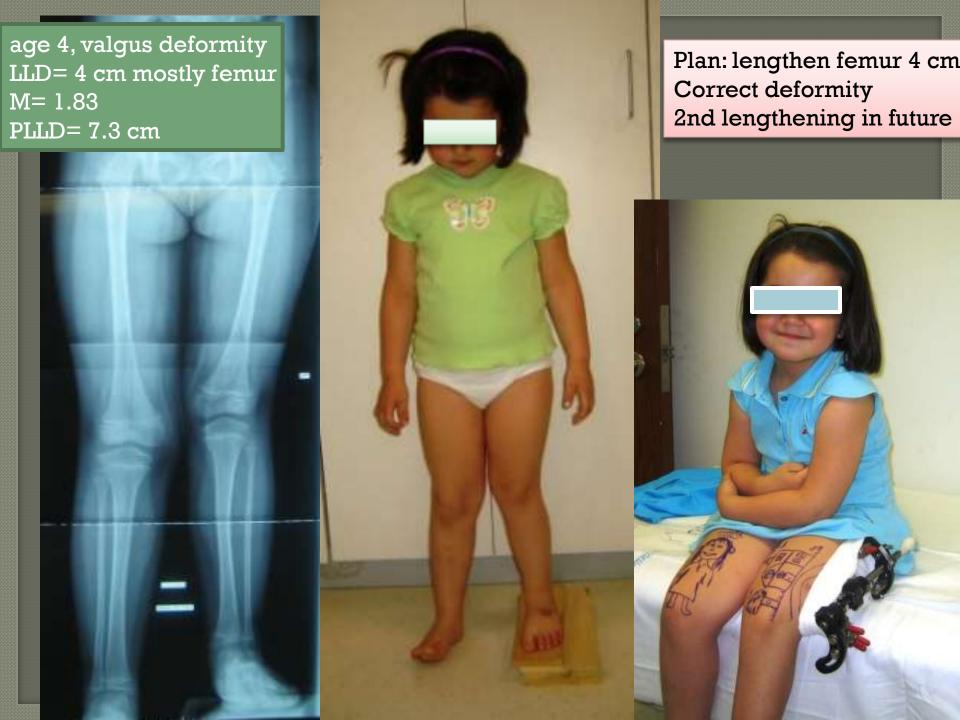


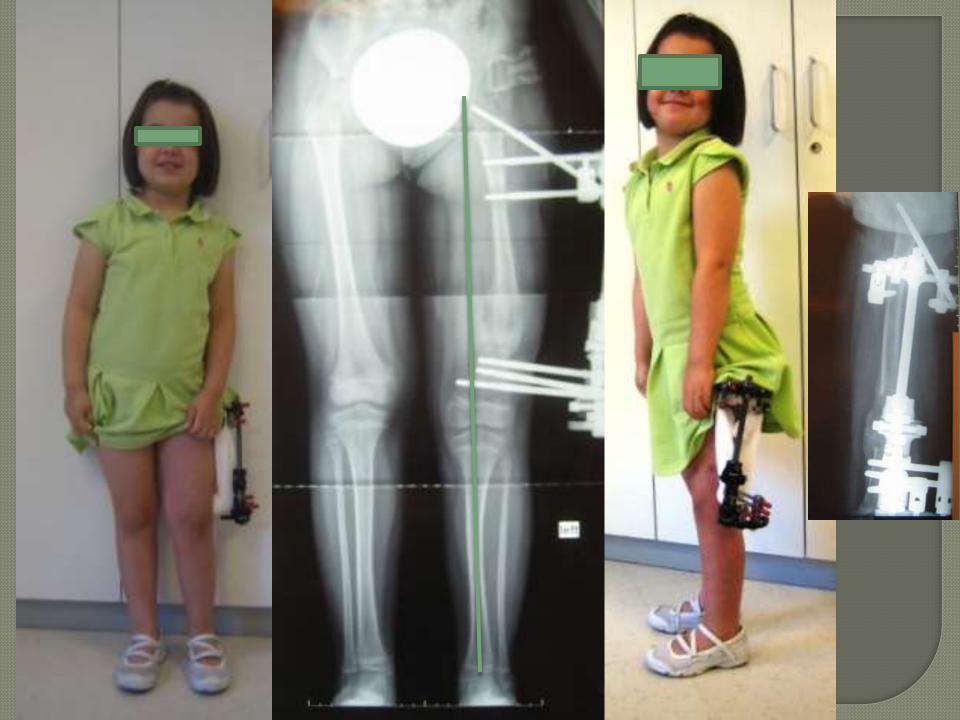






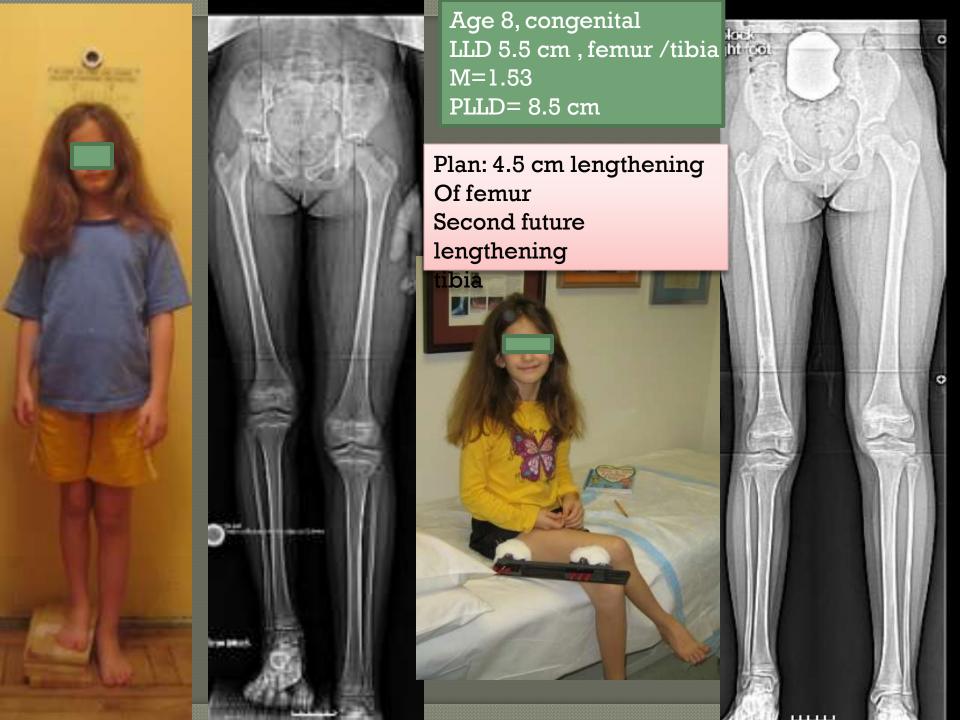












Limb Lengthening in Children with Silver-Russell Syndrome: A Comparison to Other Etiologies

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Introduction

- Silver-Russell syndrome (SRS) rare
- IUGR, difficulty feeding, postnatal growth retardation.
- LLD more than 90% of patients.
- Bone healing following lengthening is a concern (inadequate caloric intake)
- No specific data published about SRS lengthening



Growth hormone (GH)

- Abnormalities of GH secretion have been reported in many SRS children
- Human GH treatment benefits increased linear growth without concomitant increases in LLD (not limited for SRS patients)
- While hGH therapy increases total limb length it does not appear to induce limb specific catch-up growth or reduce the discrepancy between limbs.
- Given the frequency and severity of the LLD associated with SRS (reported average 3.1 cm) many patients will present for limb equalization surgery; however, epiphysiodesis is not a good option

Research question

• We asked whether pediatric patients with SRS (treated with hGH) will have uniformly good bone healing following leg lengthening.

Methods

- Retrospective comparison
- Study group SRS patients with LLD lengthening while on GH
- Control group general pediatric lengthening patients (congenital, posttraumatic, tumor)

Methods

- 7 limb segments in 5 patients with SRS
- 21 segments in 19 patients Control

Posttraumatic 8/7

Congenital 9/8

Tumor 4/4

Methods

	SRS	Control	P value
Age (years)	10.4	13	0.036
Lengthening (cm)	3.3	3.9	0.507
Follow up (months)	32 (16-38)	58 (12-130)	

Bone Healing Index

days of bone healing per cm of lengthening

	SRS	Control	P value
Bone Healing Index (BHI), days/cm	29	43	0.028

RSS patients had significantly faster bone healing during limb lengthening

Subgroup comparison of Bone healing Index

	SRS,	Trauma,	Congenital
	29	31.4	41.4
Congenital	P=0.032	P=0.068	
41.4			
Tumor,	P=0.019	P=0.04	P=0.162
66			
Trauma, (P=0.298		P=0.068
31.4			

Discussion

Function limiting LLD vs. concern about bone healing.

Scarcity of literature on SRS lengthening

hGH has known positive effect on fracture healing, not well documented for human limb lengthening

Recent animal studies also showed that GH improved muscle recovery during limb lengthening

- All SRS patients had good outcome, no significant problems
- No premature consolidation on hGH
- No hGH-related orthopedic complications (LCP, SCFE, scoliosis)

Discussion



- SRS patients treated with hGH -uniformly good healing of bone regenerate
- SRS BHI is significantly shorter than in a general pediatric population.
- hGH may
 significantly
 improve regenerate
 formation and
 consolidation

Conclusion

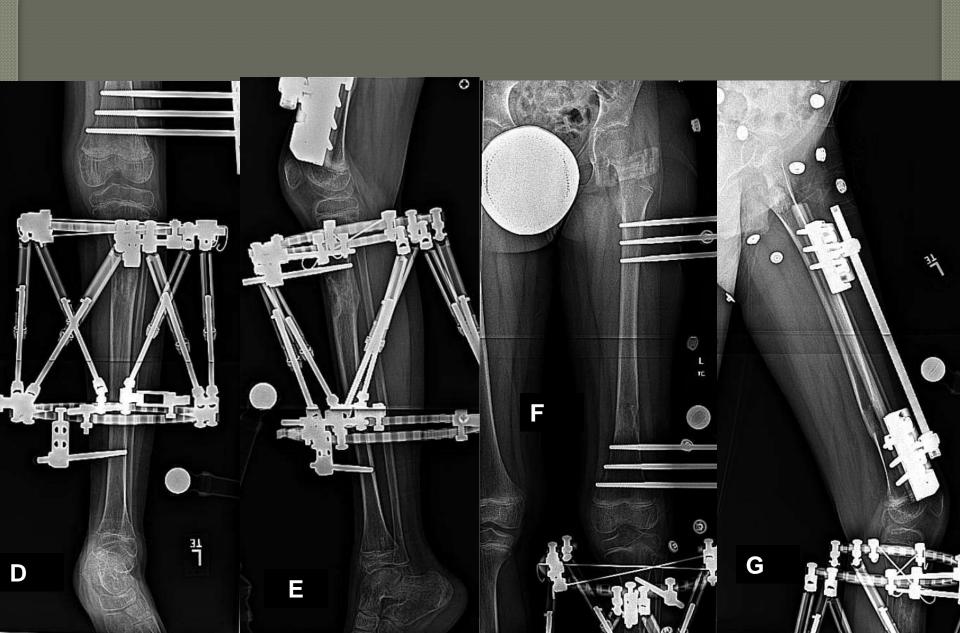




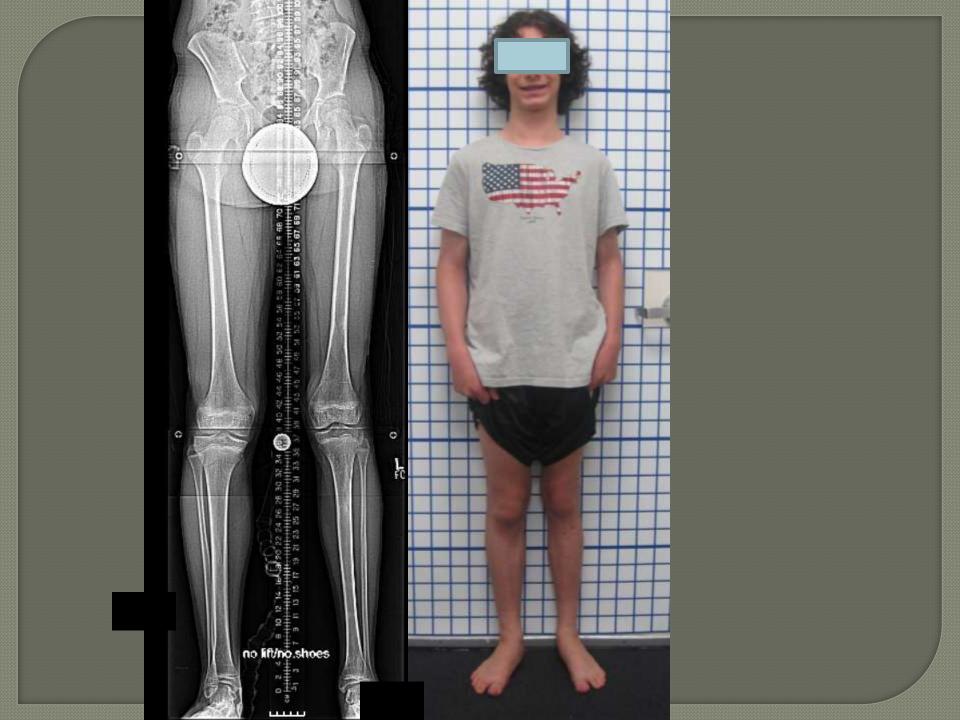
Age 12 yo LLD 39 D/45 ID mm Δ F 27 mm Δ T 18 mm PLLD \sim 5.3 cm











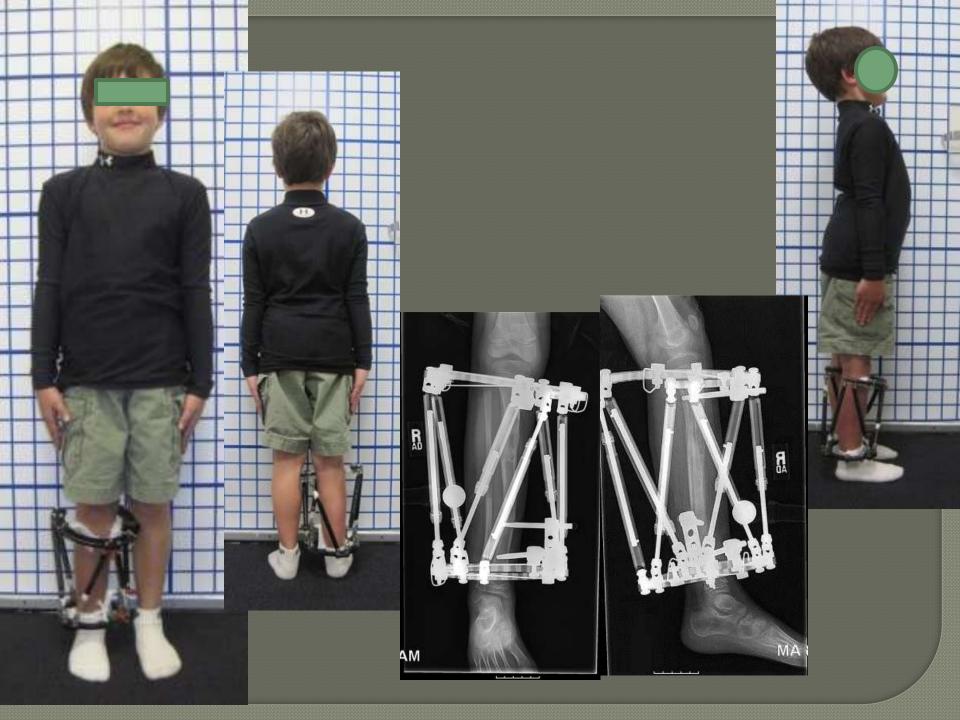




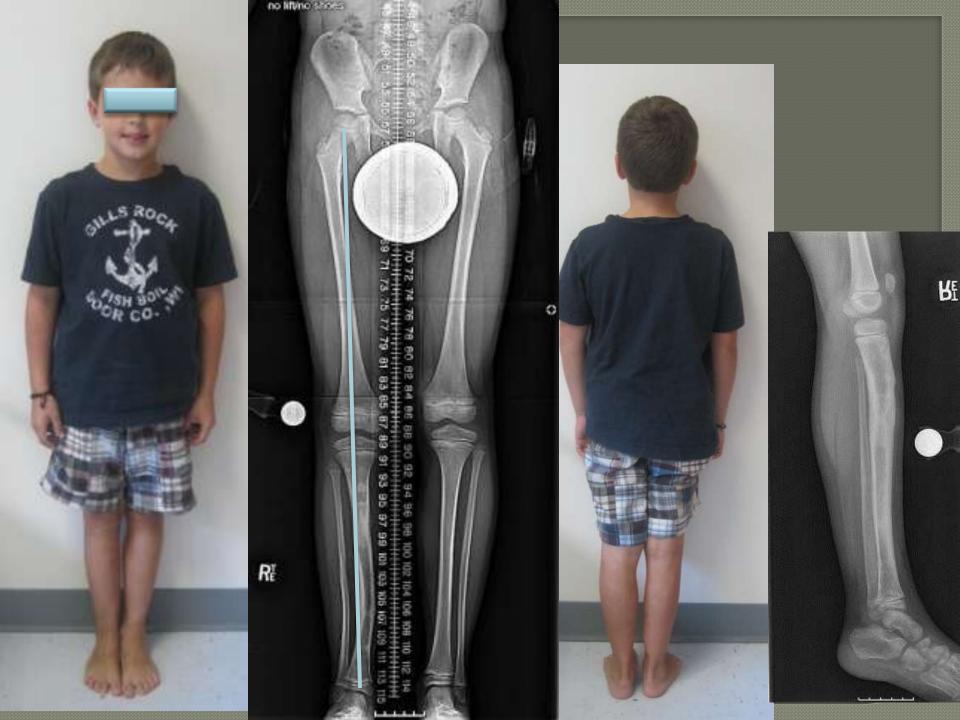
Posteromedial bow, age 6 LLD= 36 mm, all tibia

Plan: lengthen tibia 3.6 cm Correct some prox tibia varus Second lengthening in future











Ellis Van Crevald Syndrome



hemiepiphysiodesis







































Age 9 5 cm LLD



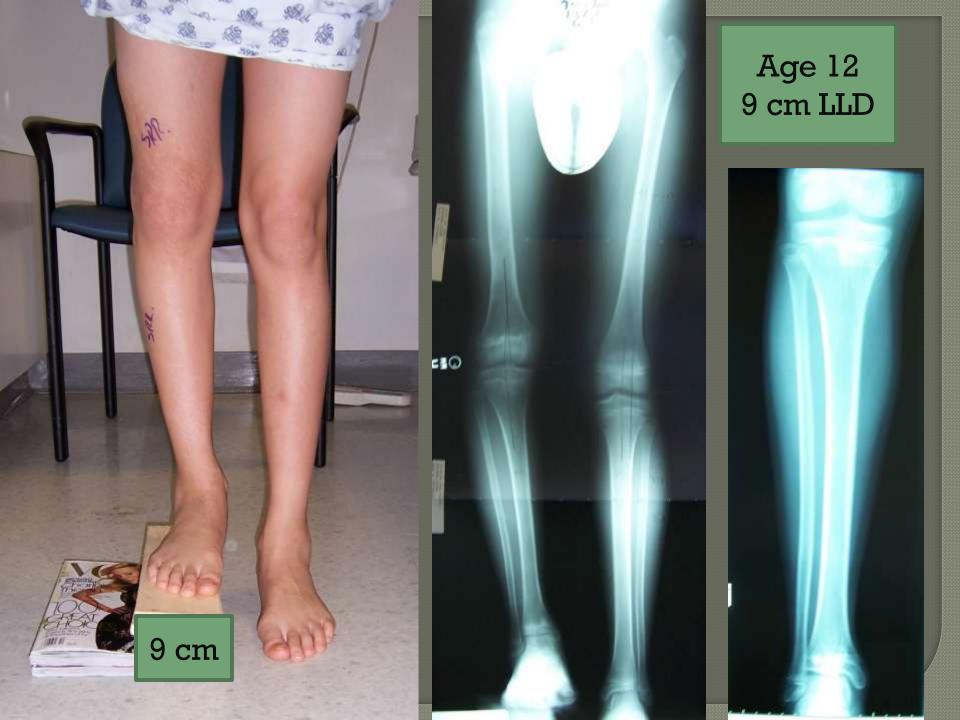




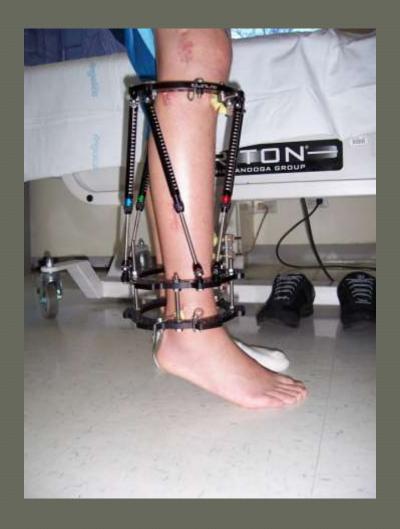












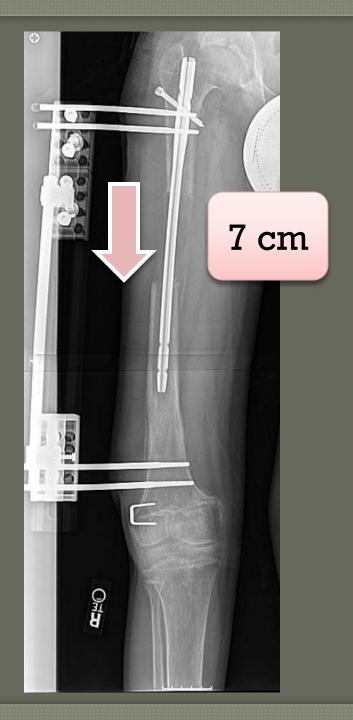


7 cm LLD At age 15

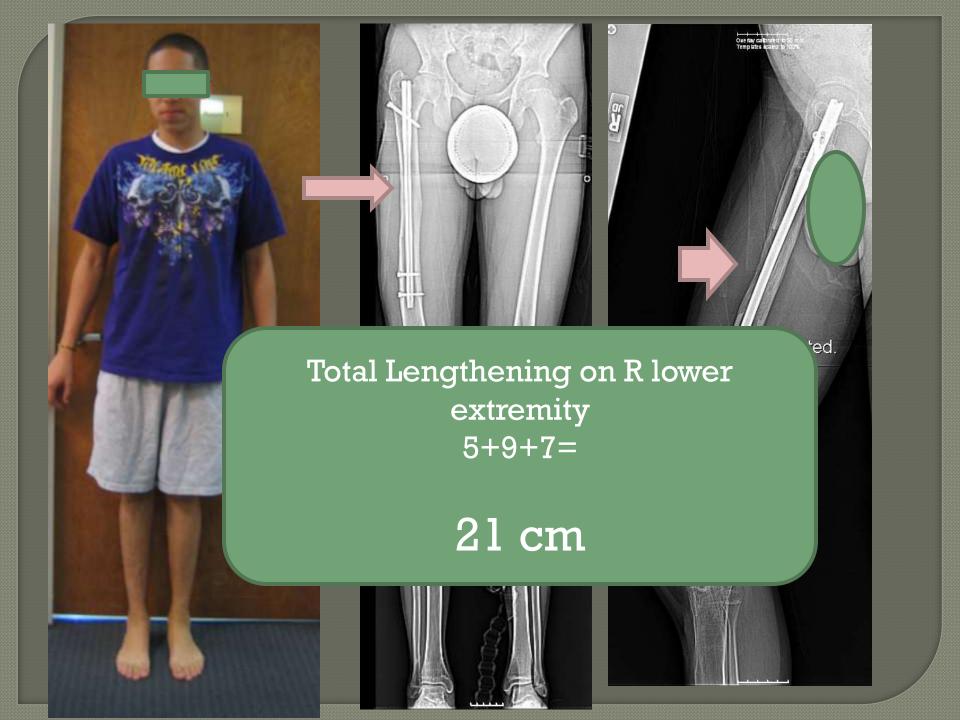






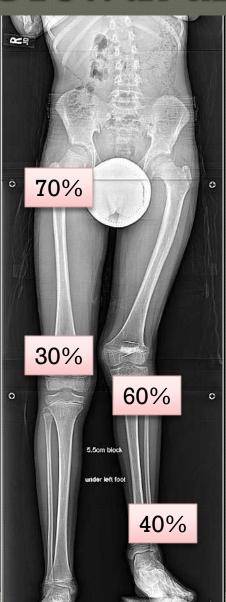






Growth arrest

Relativecontributions ofvarious growthplates





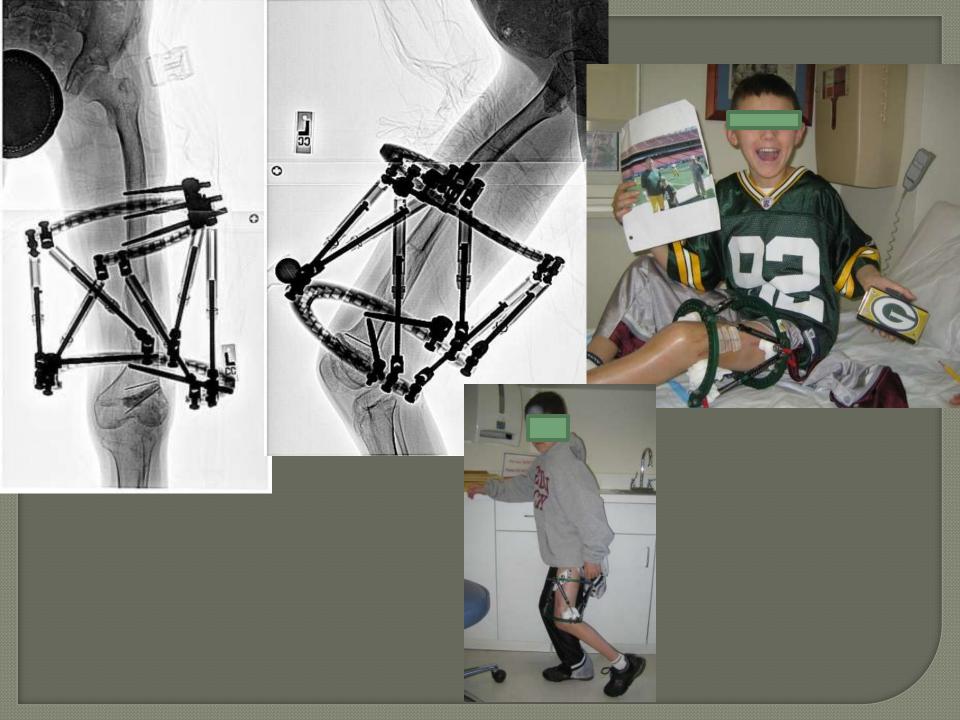


Age 8
Distal femur growth arrest
Proximal tibial also
LLD 7 cm
Valgus deformity

PLLD

M= 1.47 R femur= 350 x 1.47 R femur will be 515 515-350= 165 mm 165 x 70%= 11.5 cm

Plan: lengthen femur 7 cm, correct valgus, Close growth plate. Second lengthening of about 5 cm. femur and / or tibia

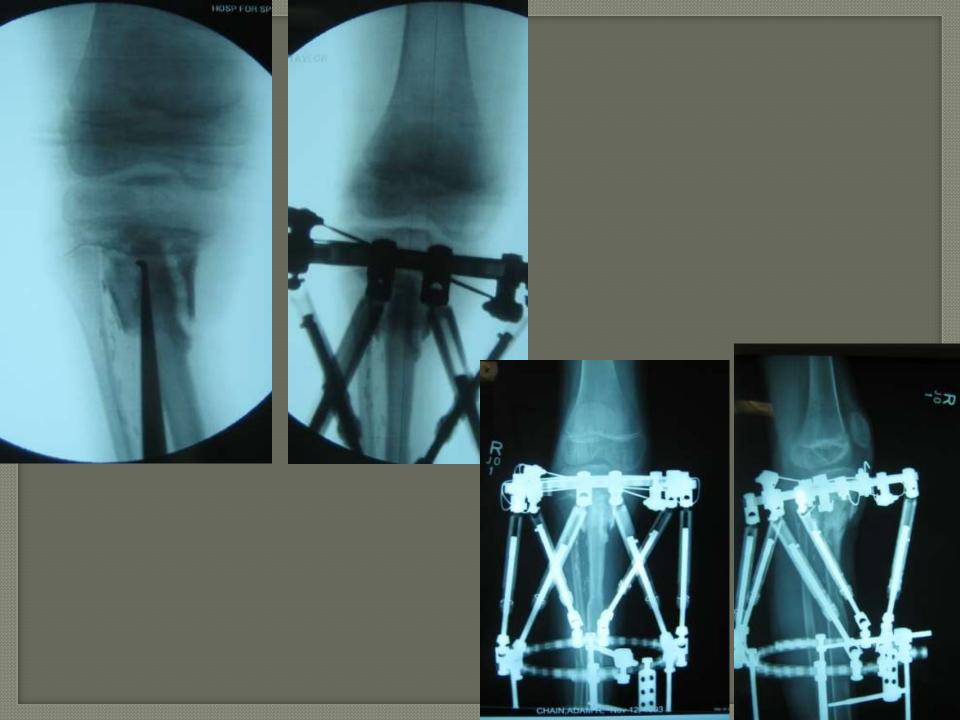


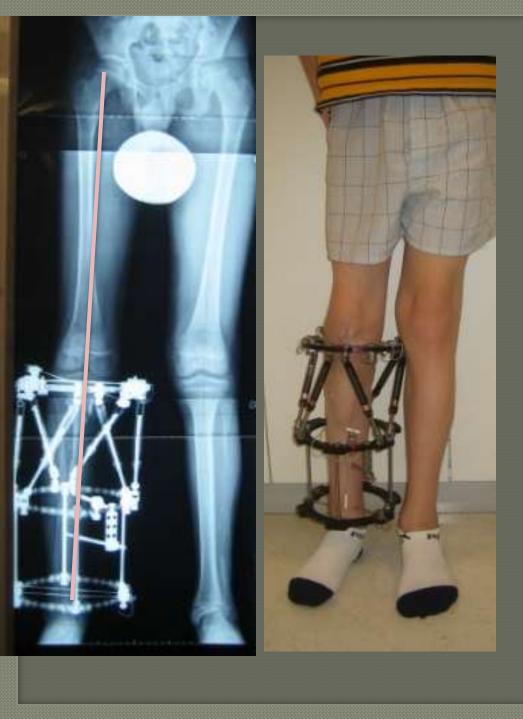












L ankle valgus from free fibula Donor site

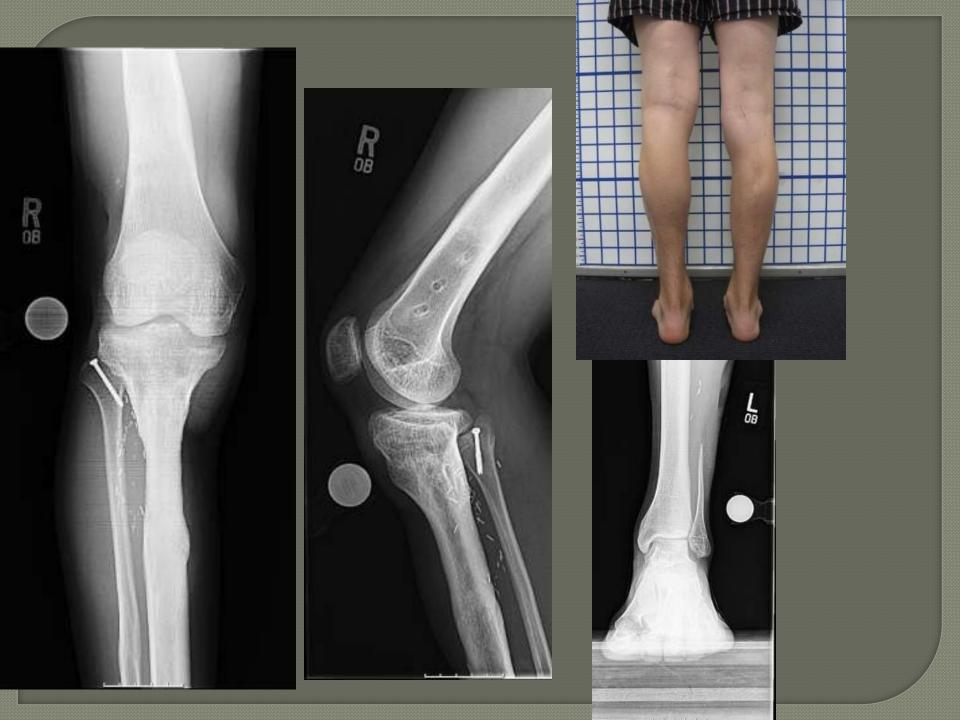








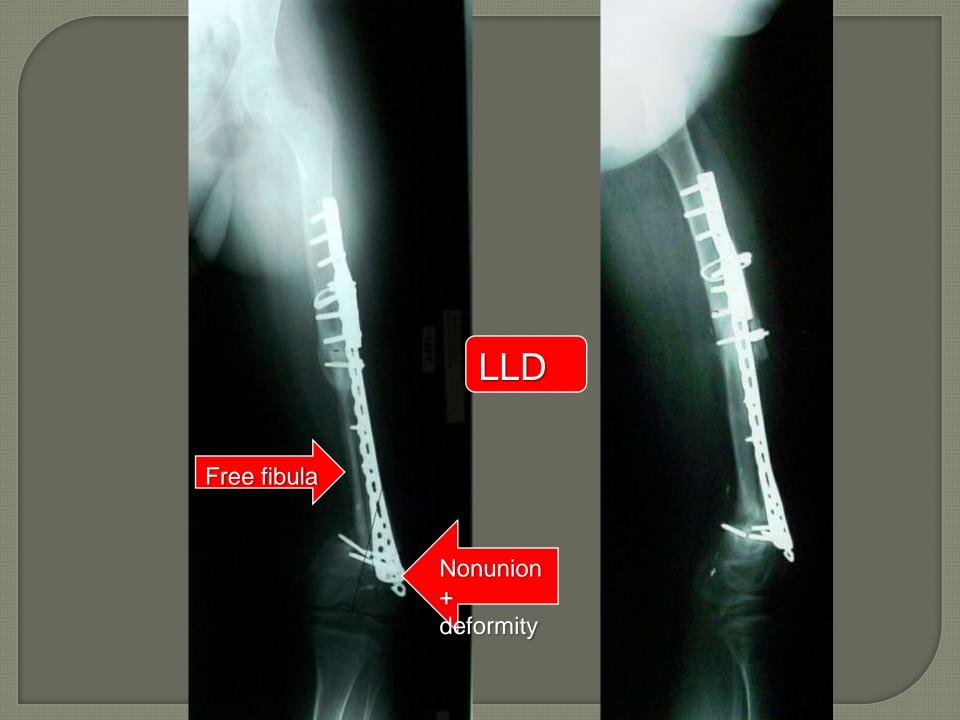


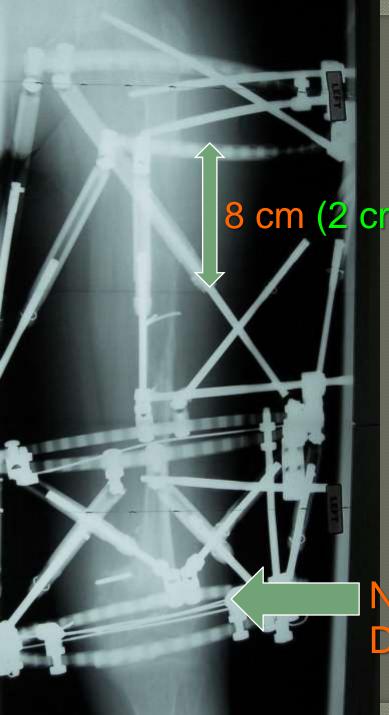








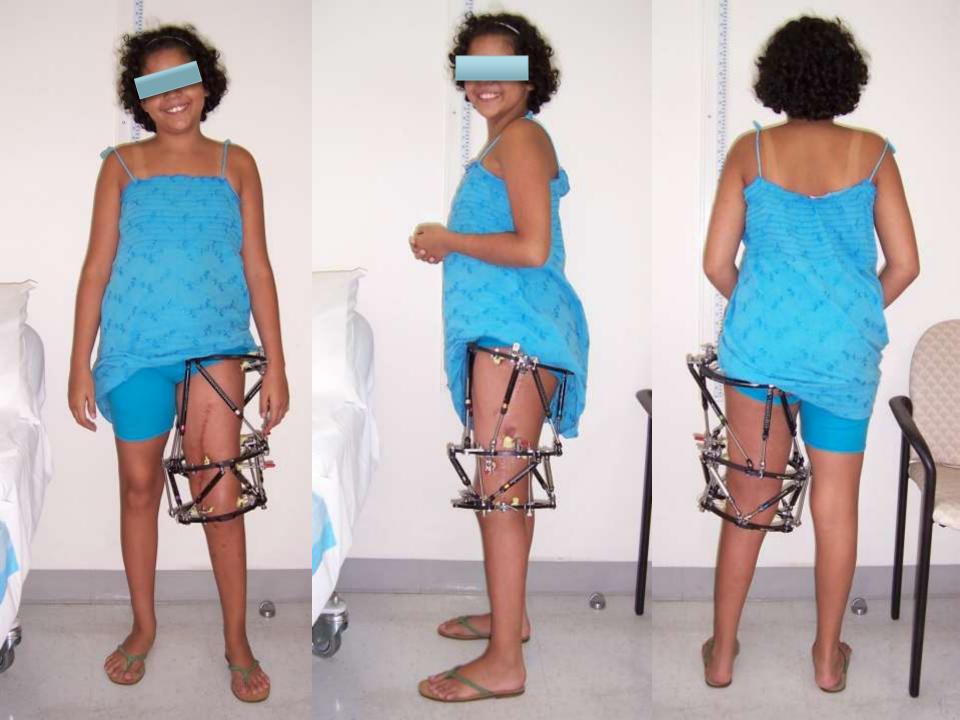


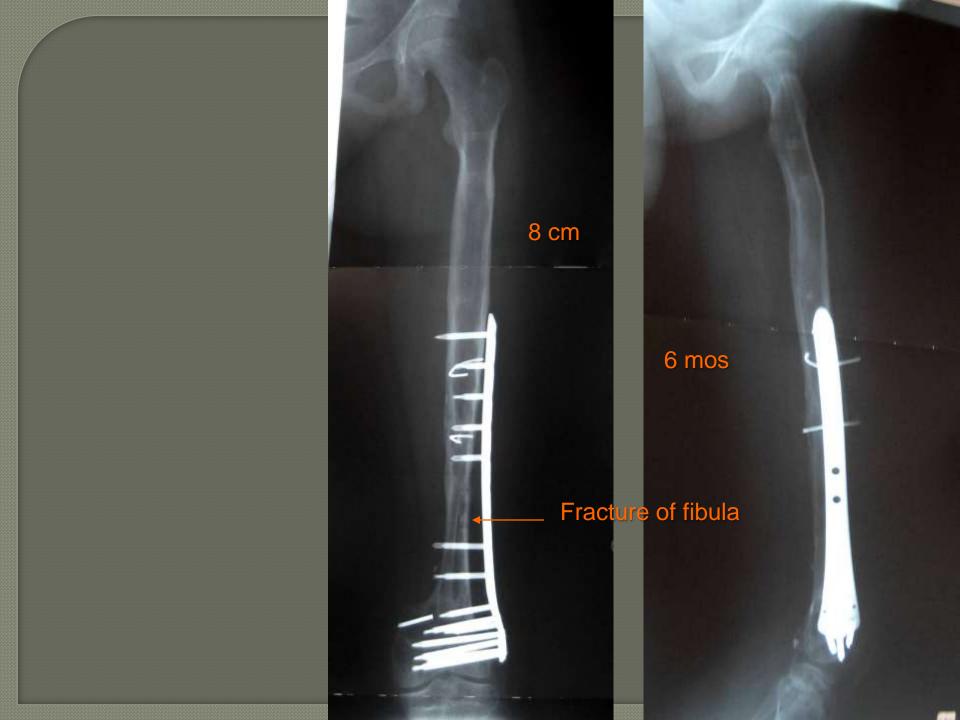




Nonunion repair
Deformity correction















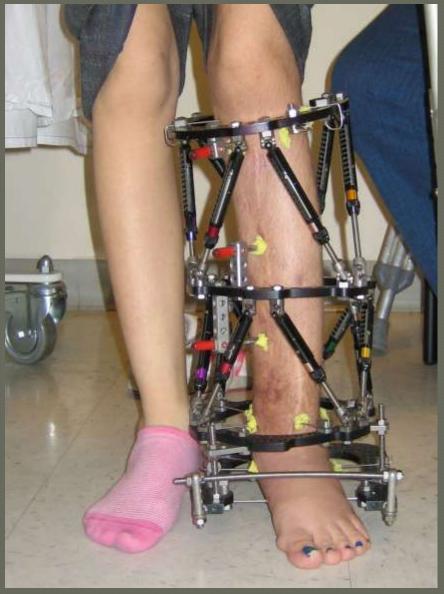
CPT- congenital pseudoarthrosis of tibia



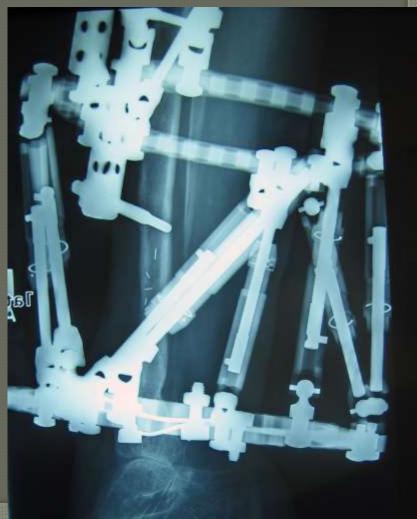














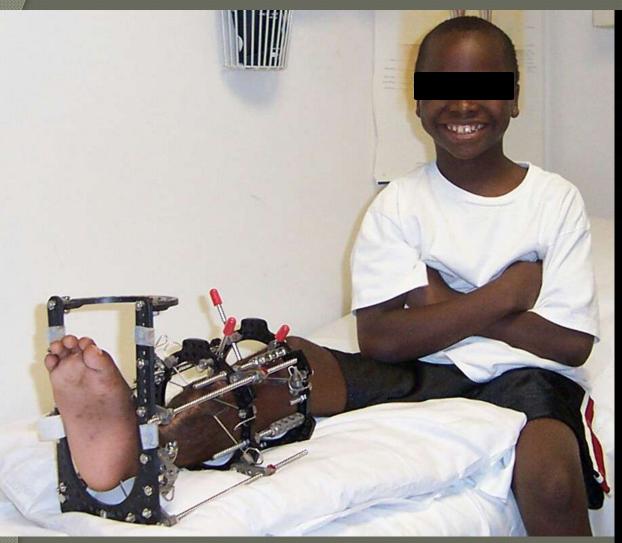




















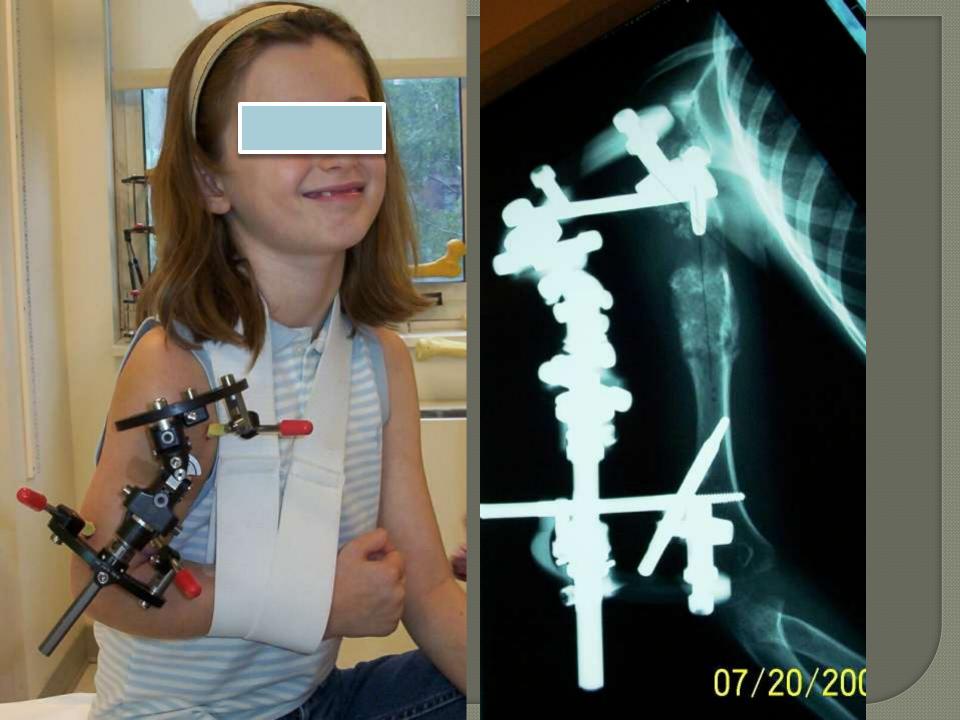


Ollier's Disease

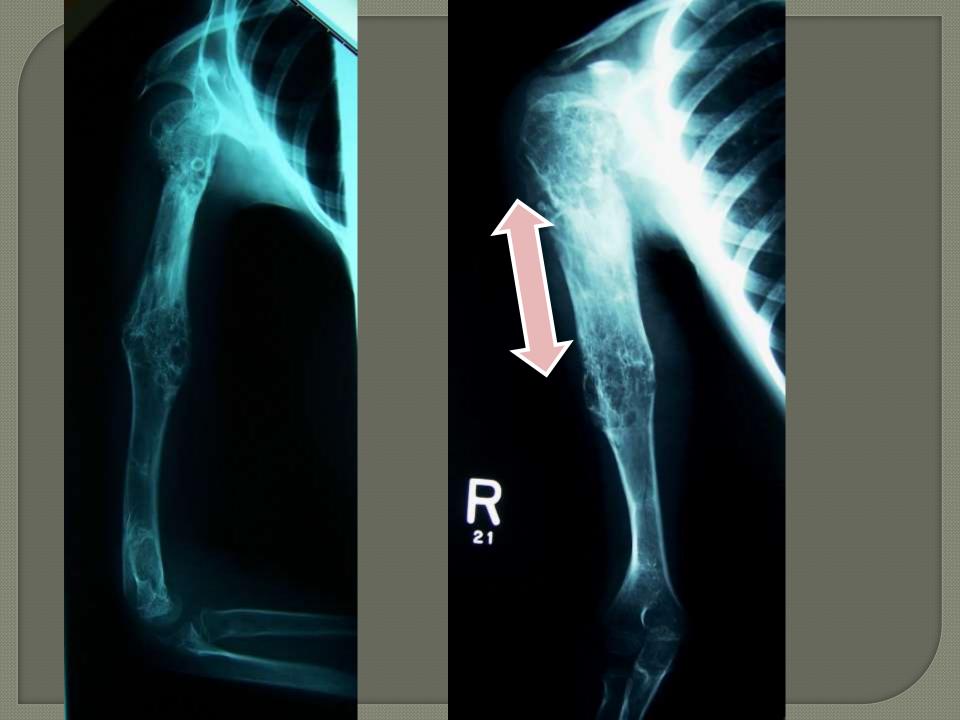


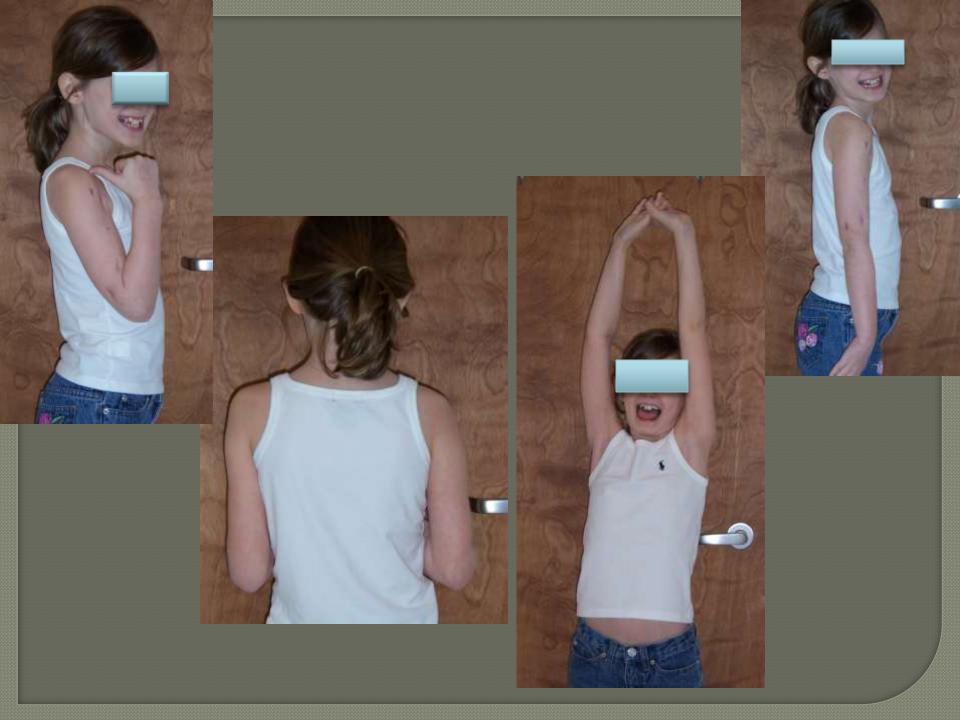


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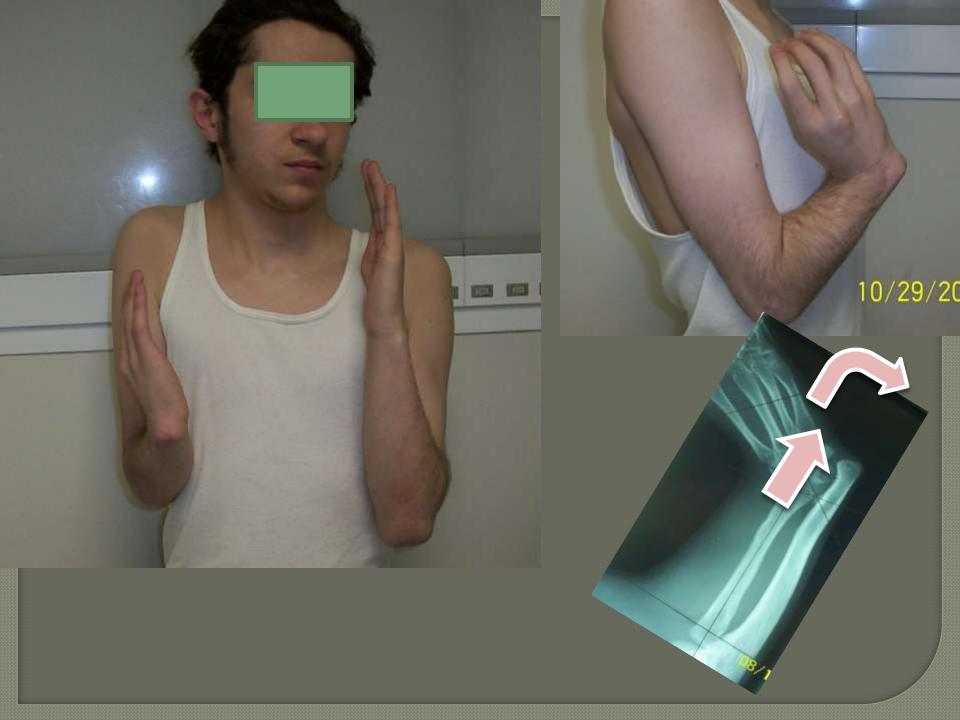












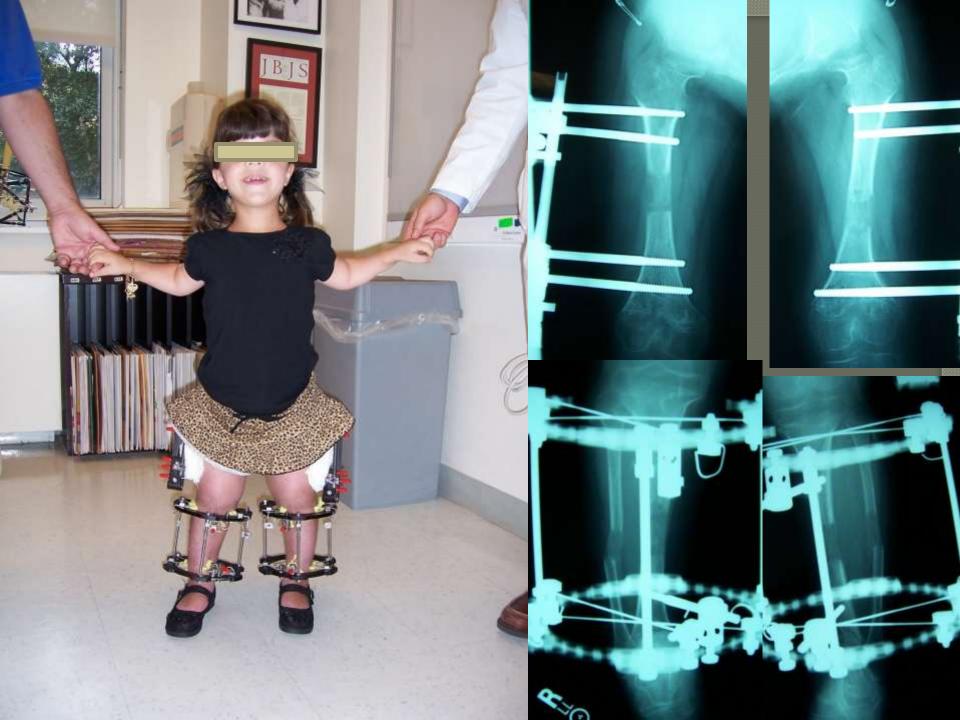




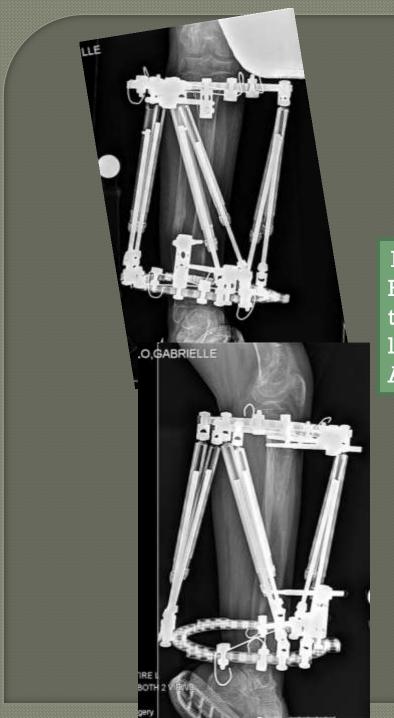




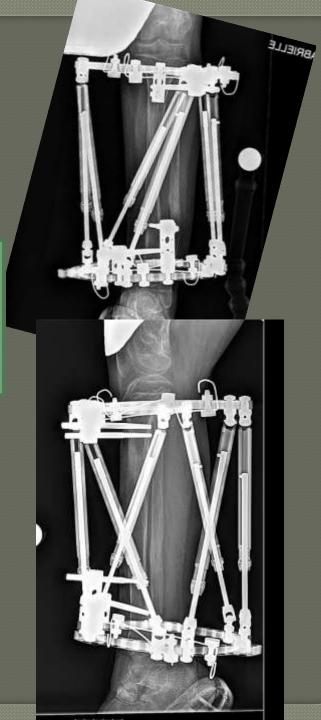








10 cm Bilateral tibial lengthening Age 13

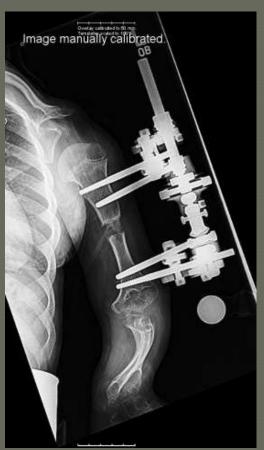


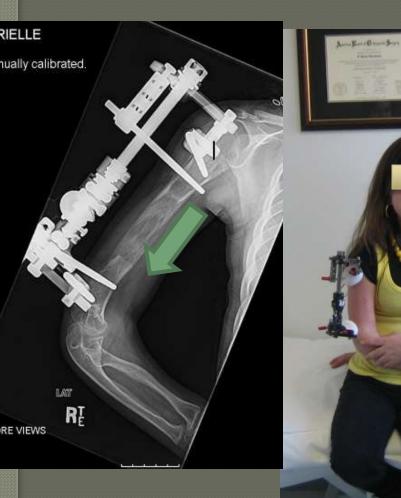




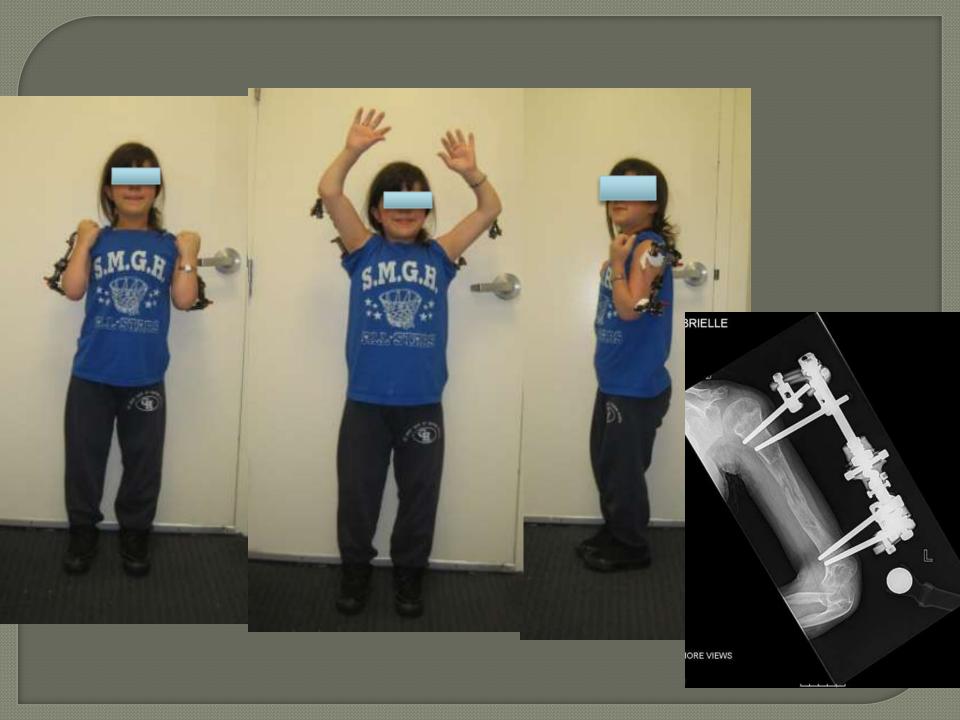






























Blount's Disease







