

Localized Pediatric Disorders of Bone and Soft Tissue

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Definition and Overview

- Process affecting the skeleton in a local or regional manner
- not unique to one area of the body
- not part of a systemic abnormality



Congenital/Developmental -soft tissue or generalized

- Klippel-Trenaunay Syndrome
- Hemangiomas
- Lymphangiomas
- Hemihypertrophy/hypotrophy
- Macrodactyly
- Constriction band syndrome



Acquired Disorders

- Myositis Ossificans
- Osteochondroses
- Quadriceps fibrosis
- Reflex Sympathetic Dystrophy



Klippel-Trenaunay Syndrome

- Diagnosis is based upon three criteria
 - Cutaneous hemangioma or nevus
 - Varicose veins
 - Hypertrophy in length or girth



Klippel Trenaunay Syndrome

- Most evident by 0-6 years old
- Lower extremities most involved



Kippel-Trenaunay Syndrome -Treatment

- Compression
 - Intermittent pneumatic at night
 - Jobst
- Surgery, only if
 - Severe dysfunction
 - Cardiac failure
 - coagulopathy



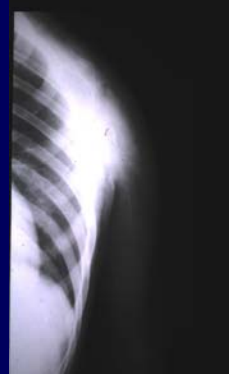
Lymphedema

- Congenital (Milroy's disease)
- Turner Syndrome
- Noonan syndrome



Gorham's Disease (disappearing bone disease)

- Progressive bone lysis without sclerosis
- Fractures do not heal



Gorham's Disease - Treatment

- Reconstruction
 - Prosthesis
 - Allograft
 - Vascularized autograft
- Amputation



Hemihypertrophy/hypotrophy

- Discrepancy > 1 cm
- Overgrowth
 - Idiopathic
 - Beckwith-Weidemann
 - Klippel-Trenaunay
 - Proteus
 - NF1



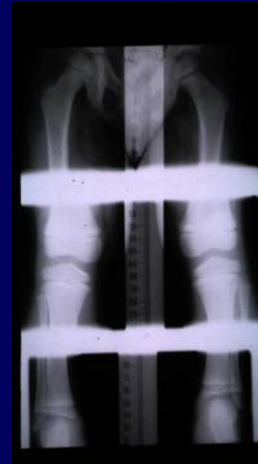
Hemihypotrophy

- Idiopathic
- Russell-Silver syndrome
- Turner mosaicism
- Neurologic



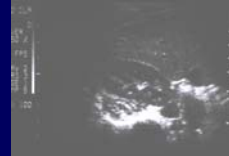
Hemihypertrophy

- Idiopathic
 - Girth and length increased
 - One limb vs. whole $\frac{1}{2}$ body
 - Rule out Wilms tumor
 - Usually proportionate discrepancy
 - Treatment according to standard principles



Idiopathic Hemihypertrophy

- Renal Ultrasound recommended
- ~Twice/year until age 5-6



- Measure discrepancy over time
 - proportionate



Macroductyly -localized gigantism

- 1° hyperplasia of all tissue elements of 1-2 rays of hand or foot
 - Usually 2nd and 3rd
- May be proportionate or progressive
- Upper > Lower extremities
- Rule Out NF1, Proteus, Hemangioma





Macrodactyly - Treatment

- Staged debulking
- Epiphyseodesis
- Shortening
- Amputation
- Ray resection



Congenital Constriction Band Syndrome

- 1/10,000
- Three elements
 - Bands
 - Acrosyndactyly
 - Amputations
- \pm vascular/lymphatic obstruction



Constriction bands

- Etiology: intrinsic defect or infarct
- Extrinsic band
- Differential diagnosis:
 - Michelin tire baby
 - Thread constriction



Constriction Bands

- UE > LE
- Distal > Proximal
- Central Rays most affected
- \pm syndactyly, polydactyly
- Clubfoot



Constriction Bands

-treatment

- Band excision/z-plasties
 - May be done all at one stage
- Elective or urgent
- Fasciotomies if needed



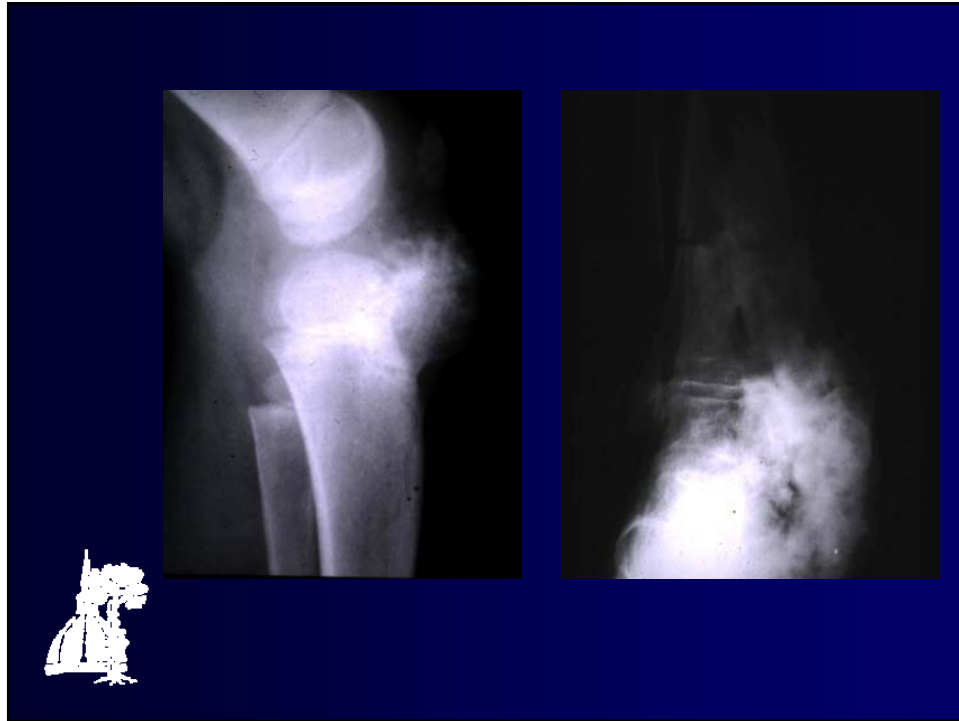


Dysplasia Epiphysialis Hemimelica

Trevor's Disease, Epiphyseal
Osteochondroma

- Usually before 14 years
- Presents with joint angulation,
locking
- Knees, ankles most commonly involved
- Widened joint space on Xray
- MRI helpful





Dysplasia Epiphysialis Hemimelica - Treatment

- Resection
 - Preserve joint surface
 - Intra-epiphyseal wedge if possible



Congenital Pseudoarthrosis of the Tibia

- An anterolateral bow
 - Usually 1st or 2nd year
 - Later onset cases do better (up to 11)
- Apex of bow more distal than genu varum
- Usually unilateral
- NF1 in 55%



Congenital Pseudarthrosis of the Tibia

- Radiographic features
 - Defect at junction mid-distal 1/3
 - Lytic/sclerotic features
 - Tapered ends
 - Usually fibular defect also



Congenital Pseudoarthrosis Tibia - Treatment

- Protective bracing if no fracture
- IM rod & bone graft
- Vascularized fibula bone graft
- Ilizarov
- Electrical stimulation
- Amputation



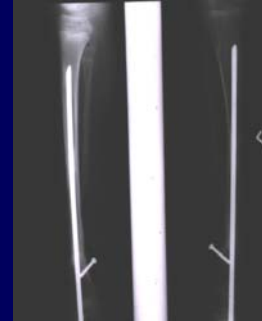
Congenital Pseuarthrosis Tiba

- Brace if not fractures
- “Bypass graft”-not recommended



IM rod and bone graft

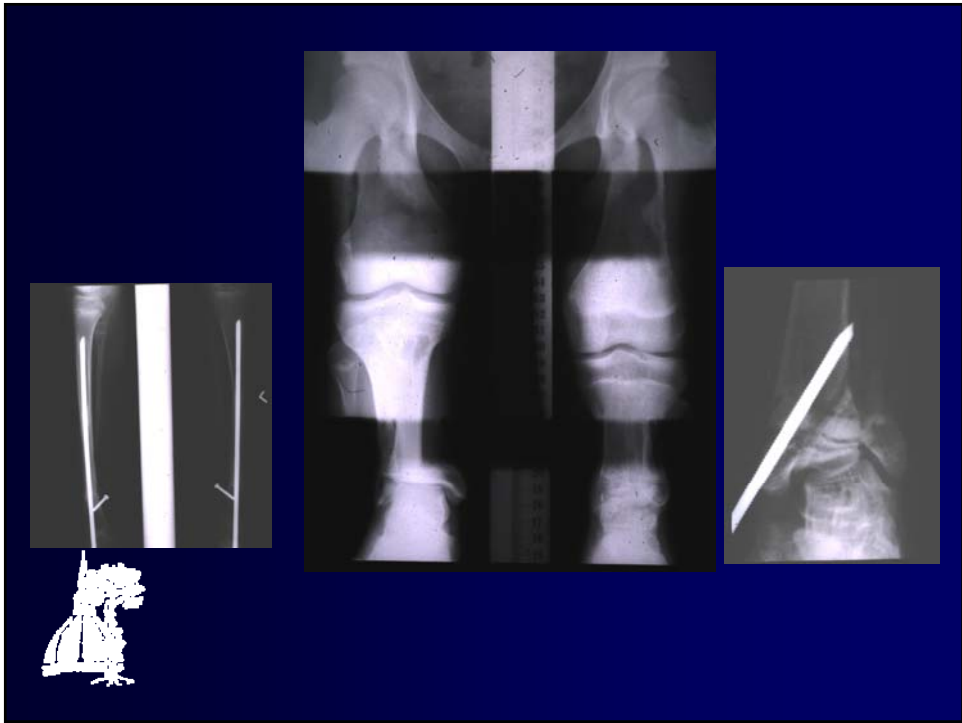
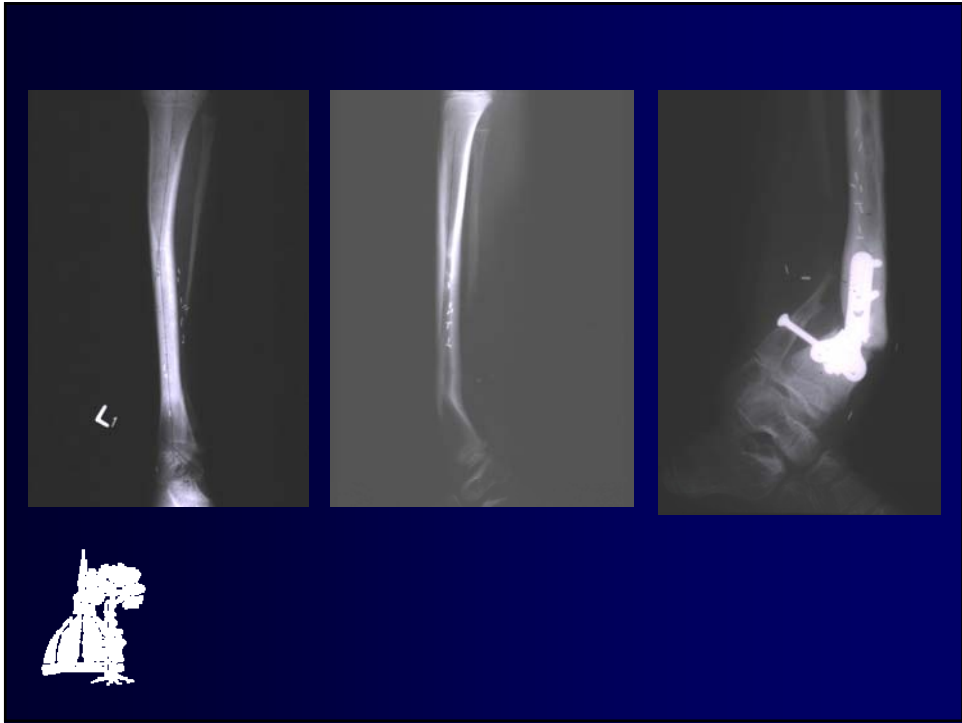
- Corrects mechanics and biology
- Protects against refracture
- Rod left through calcaneus
- 95% union
- 2 cm shortening
- ...a good initial procedure



Congenital Pseudarthrosis Tibia Vascularized graft

- Fibula or rib
- Ipsilateral fibula can be used if intact
- 95% eventual success
- Small distal fragment tends to angulate





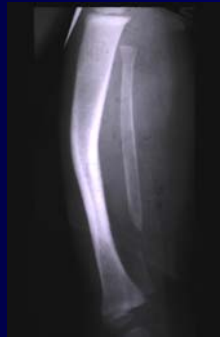
Congenital Pseudarthrosis Tibia

- Amputation
- Syme disarticulation
- Allows tibia to heal
- Allows length equalization



Congenital Pseudarthrosis Fibula

- Varus or valgus
- Rx: IM rod or synostosis



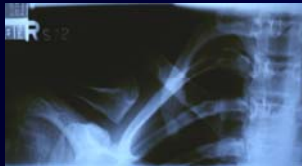
Congenital Pseudoarthrosis of the Clavicle

- Most are right sided
- Left with dextrocardia or cervical ribs
- ?due to subclavian pulsations
- Many have pain when older



Congenital Pseudoarthrosis of the Clavicle – Treatment

- Elective for pain or deformity
 - Bone graft and plate or threaded pin
 - Periosteal suture alone

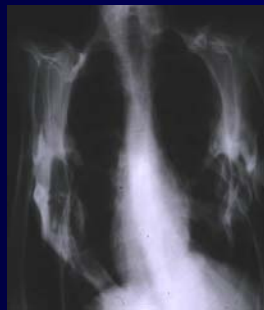


Fibrodysplasia Ossificans Progressiva

- Diagnosis
 - Progressive systemic HO and ankylosis
 - characteristic great toe deformity



- Etiology: defective regulation of induction of ossification



Fibrodysplasia Ossificans Progressiva - Treatment

- Surgery has no role
 - Don't biopsy
- Bisphosphonates \pm
- Mobility devices
- Nutrition



Acquired Disorders



Osteochondroses

Disturbance of endochondral ossification
at joint surface or physes

Classification

articular/nonarticular

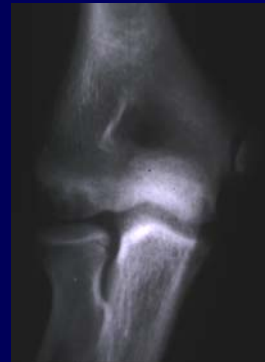
Primary/secondary



Osteochondroses

– Examples

- Panner's
- Perthes
- Scheuermann's
- Blount's
- Kohler's



Osteochondroses - Treatment

- Rest
- ROM
- Realignment PRN
- ? Drilling
- Grafting



Myositis Ossificans

- 25% of cases occur in children
 - Most over 10
- History
 - may follow trauma, injection, or be spontaneous



Myositis Ossificans

- Symptoms, signs
 - Tender, enlarging mass
 - \pm fever
 - \uparrow ESR
 - Lower extremities more than upper
 - Ca^{++} . P04 WNL



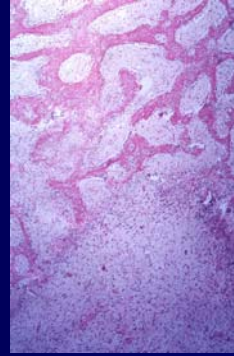
Myositis Ossificans

- Radiographs
 - Most lesions in diaphyseal region
 - Calcification from peripheral to central
 - CT shows it well



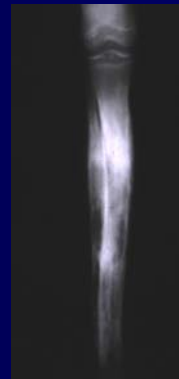
Myositis Ossificans

- Histologic Features
 - Four zones
- Treatment
 - Rest
 - NSAIDS
 - Excise if functional problem



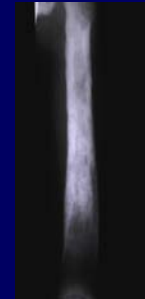
Progressive Diaphyseal Dysplasia Camurati-Engelman Disease

- Symmetrically widened, sclerotic diaphyses
- Epiphyseal sparing
- Muscle atrophy



Progressive Diaphyseal Dysplasia

- 1/ million
- Autosomal Dominant
- 1st decade: pain, fatigue, inability to run
- DDx Muscular dystrophies
- Labs, biopsy unremarkable



PDD

- Rx:
- Rest
- Steroids if severe
- Osteotomies if severe deformity



Melorheostosis

- Melos= limb
- Rhein=flowing



Melorheostosis

- Asymmetrical longitudinal hyperostotic streaks
- Limb pain
- Soft tissue contractures



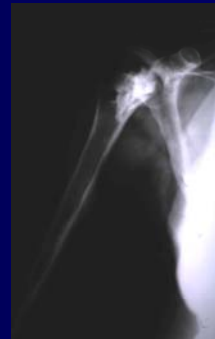
Melorheostosis

- Painless contracture in children
- Knee>ankle> hip
- Skin tight, shiny
- May resemble focal scleroderma



Melorheostosis

- Treatment:
- Analgesia
- Bracing
- Contracture releases



Osteopathia Striata

- Rare
- Asymptomatic
- Linear metaphyseal striations
- Autosomal dominant



Osteopoikilosis

- Rare
- Autosomal Dominant
- Multiple epi/metaphyseal spots
- asymptomatic



Reflex Sympathetic Dystrophy

- May occur in children
- Unexplained or exaggerated pain in an extremity with autonomic dysfunction
- Females:Males = 5:1
- Lower extremities more common



RSD -etiology

- Abnormal firing by damaged peripheral nerves
- RAS reset due to sensory input
- Abnormal synapses between damaged peripheral nerves



RSD

-diagnostic criteria

- Neuropathic pain descriptors (≥ 2)
 - Burning
 - Dysesthesia
 - Mechanical allodynia
 - Cold hypersensitivity
- Signs of autonomic dysfunction (≥ 2)
 - Cyanosis
 - Mottling
 - Edema
 - Sweating
 - coolness



RSD

-stages

- Acute
 - Intense pain, vasodilation, edema
- Dystrophic
 - Disuse, atrophy, woody edema
- Atrophic
 - Atrophic skin, Stiffness, osteoporosis



Reflex Sympathetic Dystrophy - Treatment

- Stepwise progression
- Physical, behavior therapy
- Tricyclics
- Sympathetic blocks***
- Miscellaneous (anticonvulsants, steroids, beta blockers)



RSD

- 55% have some residua at 3 years
- Best results with young age, early treatment



Thank You!

