**Pediatric Orthopedic Pathology**

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**Congenital Defects - Clubfoot (congenital equinovarus)**

* Forefoot is adducted and supinated
	+ Positional equinovarus
	+ Idiopathic congenital equinovarus
	+ Teratologic equinovarus

**Congenital Defects - Developmental Dysplasia of the Hip (DDH)**

* Abnormality of the hip that can affect the femoral head, acetabulum, or both
	+ Risk factors:
		- Female sex
		- Positive family history
		- Breech presentation
	+ The hip can present as subluxated, dislocated, or acetabular dysplasia
	+ Manifestations:
		- Asymmetry of skinfolds at groin crease
		- Galeazzi sign
		- Limitation of hip abduction
		- Positive Ortolani sign
		- Positive Barlow test
	+ Clinical management
		- Outcome becomes poorer with age
		- Pavlik harness
		- Closed reduction with spica casting
		- Surgery

**Osteogenesis Imperfecta - “Brittle bone disease”**

* ****Defect in type I collagen production
	+ Bone and vessel collagen
* Clinical manifestations:
	+ Osteopenia
	+ Increased rate of fractures
	+ Bone deformity (bowing)
	+ Short stature
	+ Blue sclera and poor dentition
	+ Aortic aneurysm
* Clinical management:
	+ Surgical
		- Intramedullary and telescoping rod placement
	+ Medical
		- Increased calcium and vitamin D
		- Biphosphates

**Osteomyelitis**

* Bone infection from bacteria or tuberculosis (granulomatous)
* ****Acute hematogenous osteomyelitis in children frequently begins as a blood abscess in the metaphysis of the bone
* The abscess ruptures under the periosteum and spreads along the bone shaft or into the bone marrow
* Clinical manifestations:
	+ Pain, swelling, warmth, fever
	+ Elevated white blood cells, C-reactive protein, and erythrocyte sedimentation rate
* Clinical management:
	+ Antibiotics for 6-week regimen
	+ Surgical debridement

**Septic Arthritis**

* Caused by bacteria or granulomatous
* Surgical emergency
* Occurs primarily or secondary to osteomyelitis
* Lysosomes destroy articular cartilage and interrupt blood supply
* Clinical manifestations:
	+ Pseudoparalysis
	+ Inability to bear weight
	+ Guarded motion of the joint
	+ Malaise
	+ Anorexia
* Clinical management:
	+ *Staphylococcus aureus* most common bacteria
	+ Surgical debridement
	+ Antibiotic therapy
	+ Long-term follow-up

**Juvenile Rheumatoid Arthritis (JRA)**

* ****Childhood form of rheumatoid arthritis
* The basic pathophysiology of JRA is the same as the adult form
* Three distinct modes of onset:
	+ Oligoarthritis
	+ Polyarthritis
	+ Stills disease
* Differences in JRA and adult RA:
	+ Large joints are affected
	+ Chronic uveitis
	+ Low detection of rheumatoid factor
	+ Subluxation and ankylosis of the cervical spine
* Treatment
	+ Supportive with anti-inflammatories and methotrexate

**Osteochondrosis**

* Avascular diseases of the bone
* Decrease blood supply
	+ Trauma
	+ Change in clotting sensitivity
	+ Vascular injury
* Legg-Calvé-Perthes disease
	+ Interrupted blood supply to the femoral head
	+ Deformation due to ischemia is permanent
	+ Clinical manifestations:
		- Spasm on rotation of hip
		- Limited internal rotation or abduction of hip
		- Trendelenburg gait
	+ Clinical management:
		- Anti-inflammatories
		- Serial radiographs
		- Surgery



**Osgood-Schlatter Disease**

* Tendinitis of the anterior patellar tendon and osteochondrosis of the tubercle of the tibia
* One of the most common ailments in children involved in sports
* Clinical manifestations:
	+ Pain
	+ Swelling
* Clinical management:
	+ Restricted activity
	+ Bracing and knee immobilizer



**Scoliosis**

* Scoliosis is a curvature of the spine that involves both lateral curvature and rotation
	+ Idiopathic (80% of cases)
	+ Congenital
	+ Teratogen
* Medical management:
	+ Bracing
	+ Surgery

**Muscular Dystrophies**

* Group of inherited disorders that cause degeneration of skeletal muscle fibers
* The muscular dystrophies cause progressive, symmetric weakness and wasting of skeletal muscle groups

**Duchenne Muscular Dystrophy**

* Most common of the muscular dystrophies
* X-linked recessive inheritance
	+ Deletion of a segment of DNA or a single gene defect on the short arm of the X-chromosome
* Generally affects boys
* Duchenne muscular dystrophy gene
	+ Encodes for the dystrophin protein
	+ Dystrophin maintains the structural integrity of the cytoskeleton
* Manifestations of the disorder begin to appear by approximately 3 years of age:
	+ Slow motor development
	+ Progressive weakness
	+ Muscle wasting
	+ Sitting and standing are delayed
	+ The child is clumsy, falls frequently, and has difficulty climbing stairs

**Musculoskeletal Tumors**

* Benign bone tumors
* Osteochondroma
	+ Inherited syndrome of hereditary multiple exostoses
* Nonossifying fibroma
	+ Sharply demarcated, cortically based lesions of fibrocytes
* Musculoskeletal Tumors

**Malignant bone tumors**

* Osteosarcoma
	+ Most common tumor in childhood
	+ Originates in mesenchymal cells - Linked to deletion of genetic material
	+ Bulky tumor extending into soft tissue
	+ Clinical manifestations:
		- Night pain, swelling, warmth,
		- Cough, dyspnea, and chest pain if lung metastasis
	+ Clinical management:
		- Graded according to malignancy
		- Surgery and chemotherapy
* Ewing sarcoma
	+ Most lethal bone tumor
	+ Translocation of chromosome
	+ Breaks through bone to form soft tissue mass
	+ Metastasizes to nearly every organ
	+ Clinical manifestations:
		- Pain that increases in severity
		- Fever, Malaise, Anorexia
	+ Clinical management:
		- Radiation and chemotherapy
		- Surgical debridement

**Nonaccidental Trauma**

* “Corner” metaphyseal fractures
	+ Long bone fractures caused by a twisting force
	+ Transverse tibial fractures are the most common
	+ Associated with child abuse, but osteogenesis imperfecta must be ruled out
* Legally mandated to report child abuse