Newborn Skeletal Radiographs: A Practical Guide

M. S. Takahashi, W. Pryor, D. Y. Otto, F. Gual, Y. T. Sameshima, B. D. Smith, C. V. Guimaraes.

Hospital Israelita Albert Einstein, São Paulo, SP, BRAZIL

UNC Hospitals at Chapel Hill, Chapel Hill, NC

Introduction

Neonatal skeletal abnormalities are rare occurrences in pediatric practice. The wide variety of causes and often overlapping radiographic features can make this evaluation very challenging

The teaching objectives of this exhibit are:

1) **Illustrate the expected normal plain film appearance of the neonatal zone of provisional calcification**

1) **Explain specific diseases radiographic findings with key clinical and epidemiological considerations**

1) **Illustrate the different imaging patterns of the diseases**

Normal Anatomy

Zone of Provisional Calcification (ZPC)

On Histology:

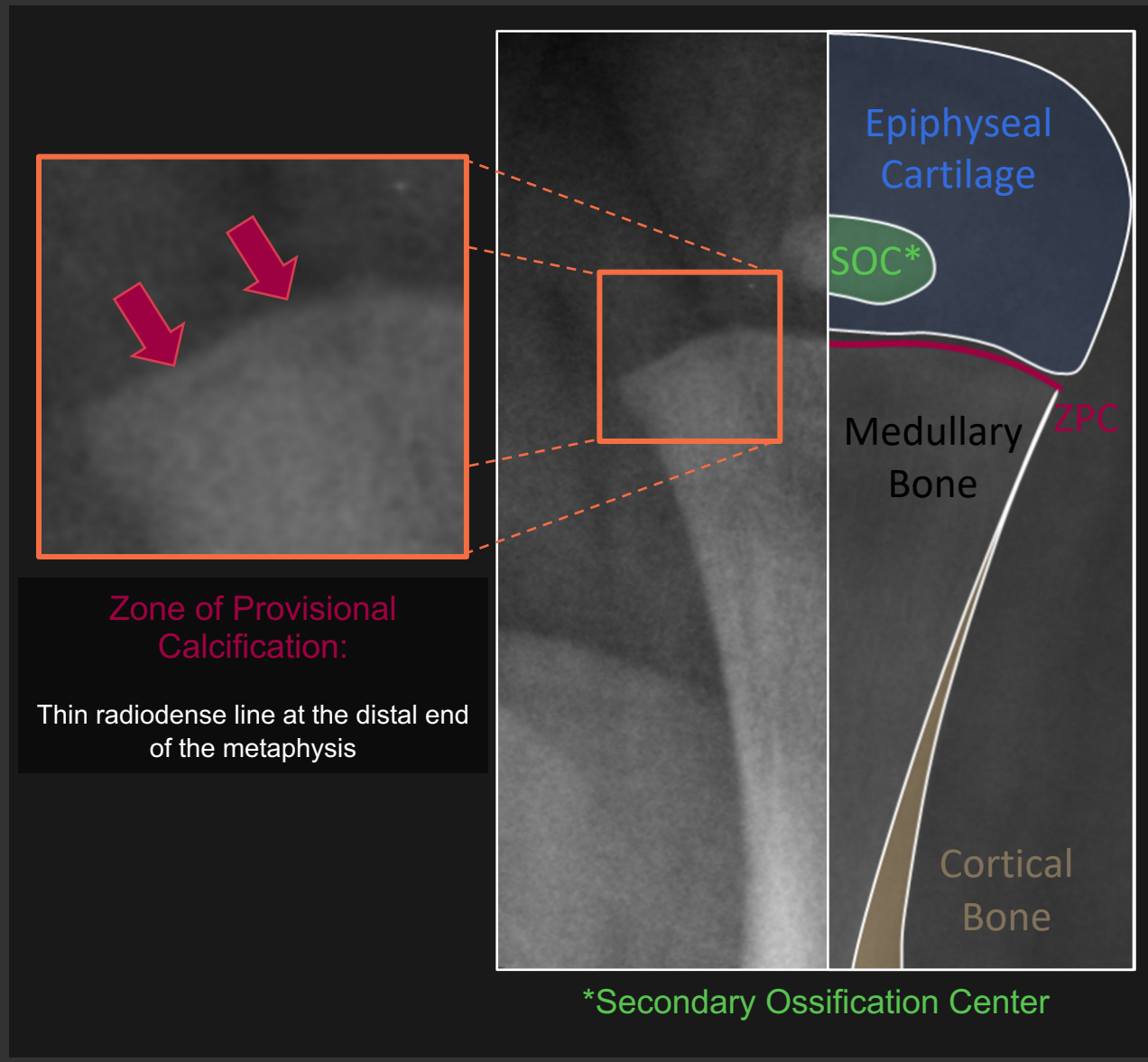
3-5 cell thick zone, which marks the transition between the bone and cartilage

On imaging:

The actual ZPC is below the resolution capabilities of normal X-Rays. Radiologists have termed the fine dense line in the transition between metaphysis and epiphysis as the ZPC

Importance:

The ZPC is an important imaging marker for some pathologies, such as rickets (decreased mineralization / discontinuity), non-accidental trauma (classic metaphyseal lesions) and congenital syphilis (metaphyseal lucent bands that spare the ZPC)



Physiologic Periostitis

Physiologic Periostitis

Normal radiologic findings in infants in up to 6 months of life.

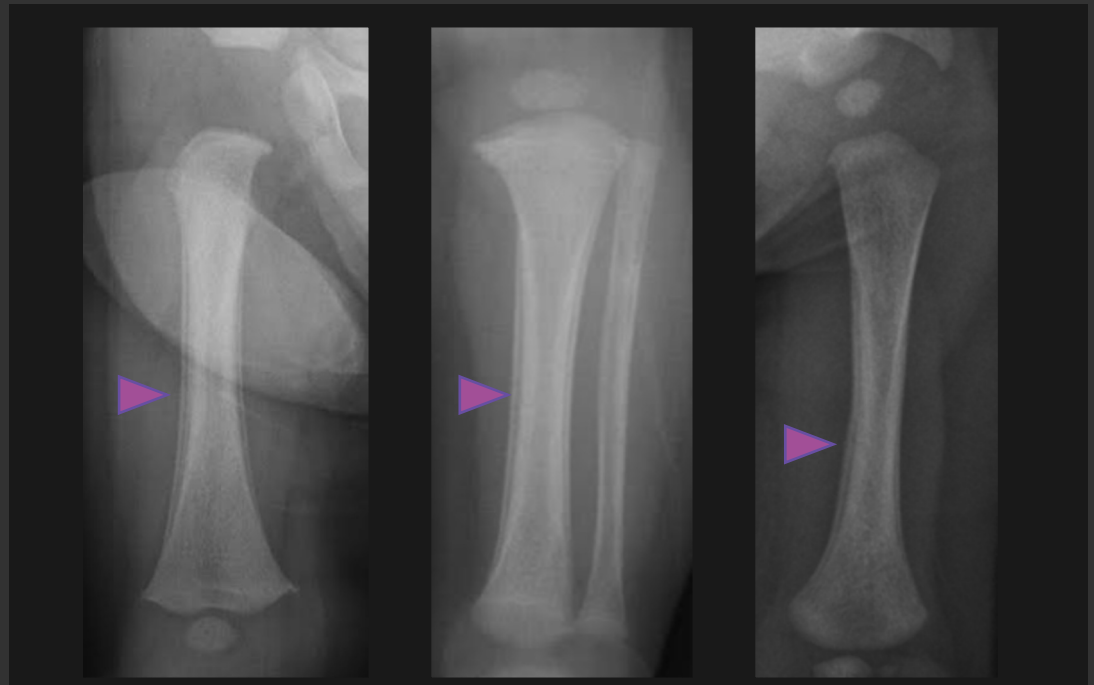
It is not as usual in newborns as in infants after 1 month of life.

Etiology is uncertain

Is not related to gestational age (can affect both term and preterm babies)

Importance:

As a normal radiologic finding, physiologic periostitis is an imaging pitfall, which can be confused with pathologies such as accidental trauma, non-accidental trauma, osteomyelitis and Caffey disease



Physiologic Periostitis in a newborn.
Thin, smooth and symmetrical periosteal reaction on the long bones (femur, tibia and humerus

Differential Dlagnosis:

Prostaglandin Induced Cortical Hyperostosis

Prolonged use of prostaglandins (maintaining ductus arteriosus patency in congenital heart disease patients) can cause limb swelling and long bone periosteal reaction, which usually resolves after the drug is withdrawn

Radiographic Findings:

- Smooth and thin periosteal reaction (< 2 mm thick).
- Affect diaphysis of long bones
- Usually bilateral and symmetric

Mechanical birth-related trauma

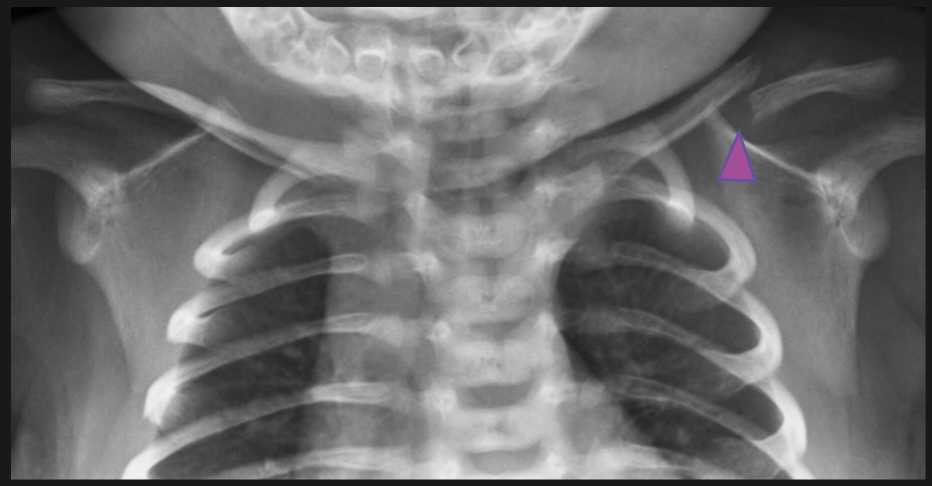
Clinical Summary

Mechanical birth-related trauma (MBRT) are injuries sustained by the newborn owing to forces of labor and delivery

The incidence of MBRT is estimated in less than 7% and less than 8% of neonatal deaths are consequence of MBRT

Risk factors include macrosomia, malpresentation and shoulder dystocia

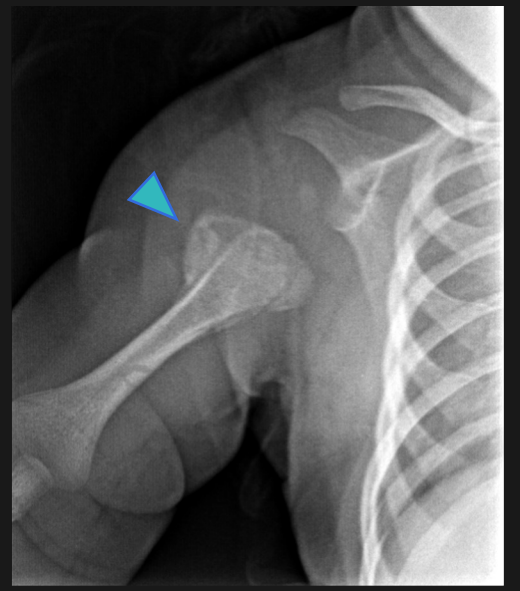
MBRT can affect multiple systems, mainly central nervous system and musculoskeletal system



Newborn with traumatic birth.
- Fracture of the clavicle

Radiographic Findings:

- Clavicle fractures are one of the most common musculoskeletal MBRTs and they can co-exist with brachial plexus injuries
- Humeral fractures can also occur, either on the humeral diaphysis or as a chondro-epiphyseal separation (Salter Harris I)
- Femoral fractures are very rare in this context



20-day old macrosomia newborn with history of traumatic birth
- Healing fracture of the humeral metaphysis

Non-Accidental Trauma / Child Physical Abuse

World Health Organization Definition :

Inflicting injury upon a child, such as burning, hitting, punching, shaking, kicking, beating or otherwise harming a child. The parent or caretaker may not have intended to hurt the child

Clinical Summary:

Estimated yearly incidence of 2 in 1,000 children in the US

Fractures are the second most common injury (skin injury / bruising are the most common)

Physical abuse is the cause of 12-20% of fractures in infants and toddlers

Understanding the type and dating of fracture, the mechanism of trauma and the child's physical development are key in determining the likelihood of physical abuse

While some fracture types have a higher specificity than others for physical abuse, less specific fracture can be used on its own to confirm or discard the diagnosis

Radiologic Finding	Peak
Soft tissue swelling resolution	4-10 days
Subperiosteal new bone formation	10-14 days
Soft callus	14-21 days
Hard callus	21-42 days
Bone remodeling	1 year

Highly Suggestive Fractures
Classical Metaphyseal Lesions (CML)
Posterior ribs
Scapula
Spinous Process
Sternum
Multiple fractures with differing ages

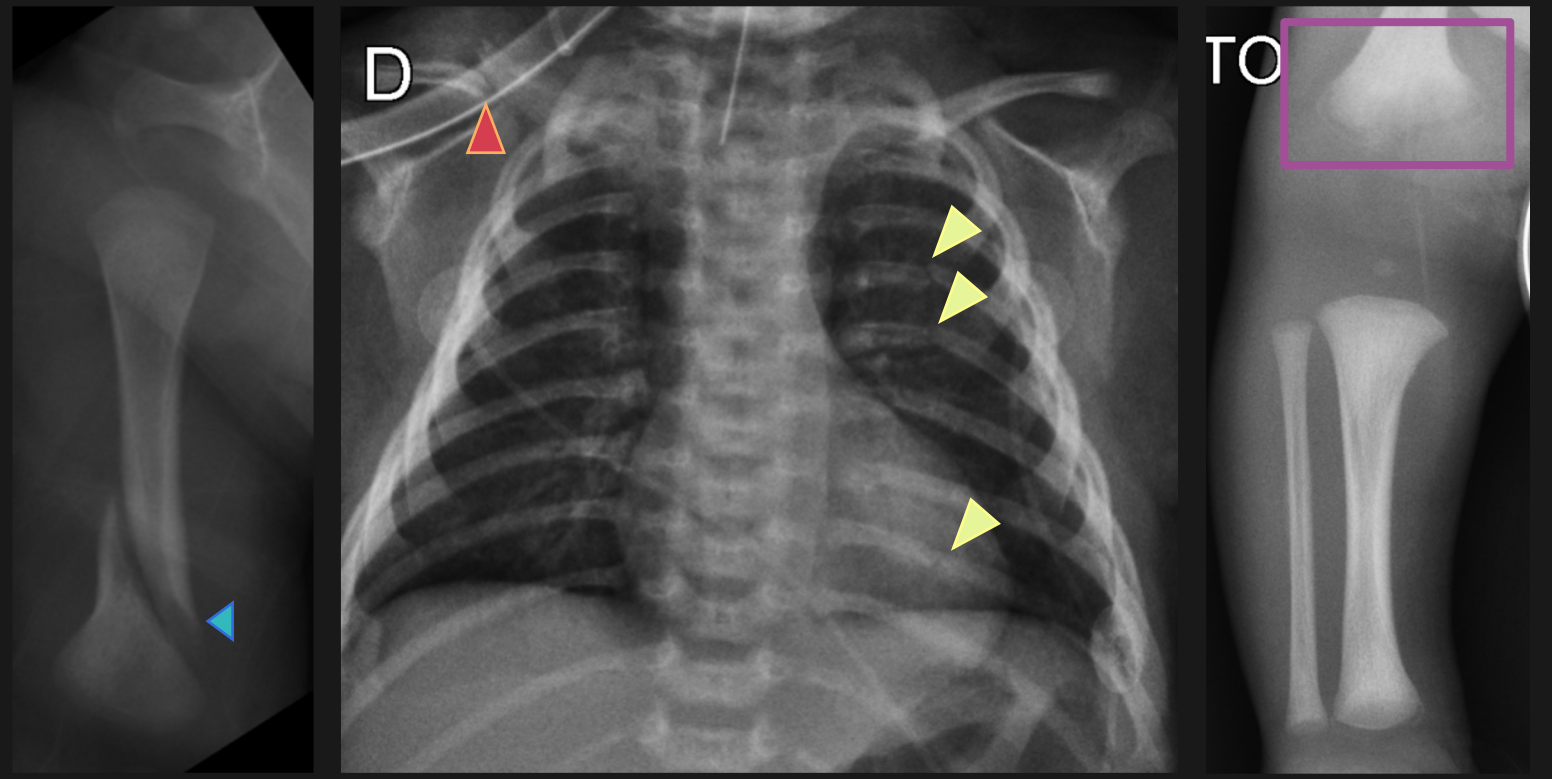
Non-Accidental Trauma / Child Physical Abuse

Imaging

ACR Appropriateness Criteria for Suspected Physical Abuse – Child:

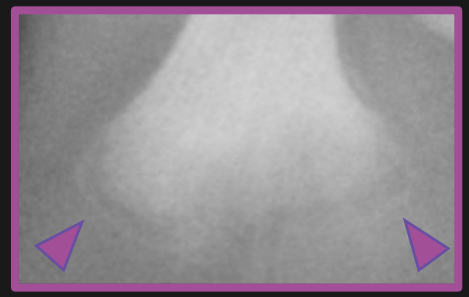
- "Skeletal survey is the universal screening examination in children 24 months of age and younger"
- "Images should be obtained using high-detail imaging systems and coned to the specific area of interest"
- "A repeat skeletal survey performed approximately 2 week after the initial examination can provide additional information … in up to 12% of children"

Skeletal Survey Protocol
Skull (frontal and lateral)
Cervical, thoracic and lumbar spine (frontal and lateral)
Long bones, hand, feet chest and abdomen (frontal)
Ribs (oblique)



Twenty-day old newborn, with normal prenatal exams, brough to the Emergency Department due to lethargy, latter confirmed as non-accidental trauma.

- Humerus diaphyseal fracture.
- Multiple posterior rib fractures
- Right clavicle fracture
- Classic metaphyseal lesions (CML) – "Bucket Handle"



Rickets

Clinical Summary:

Growth plate mineralization and ossification disorder:

Exclusive to children:

- Cartilage mineralization failure Hypertrophy of cartilage

Etiology:

- Nutritional vitamin D deficiency (most common, peak: 3-24 months)
- Other causes: Malabsorption, liver disease, renal disease, genetic syndromes

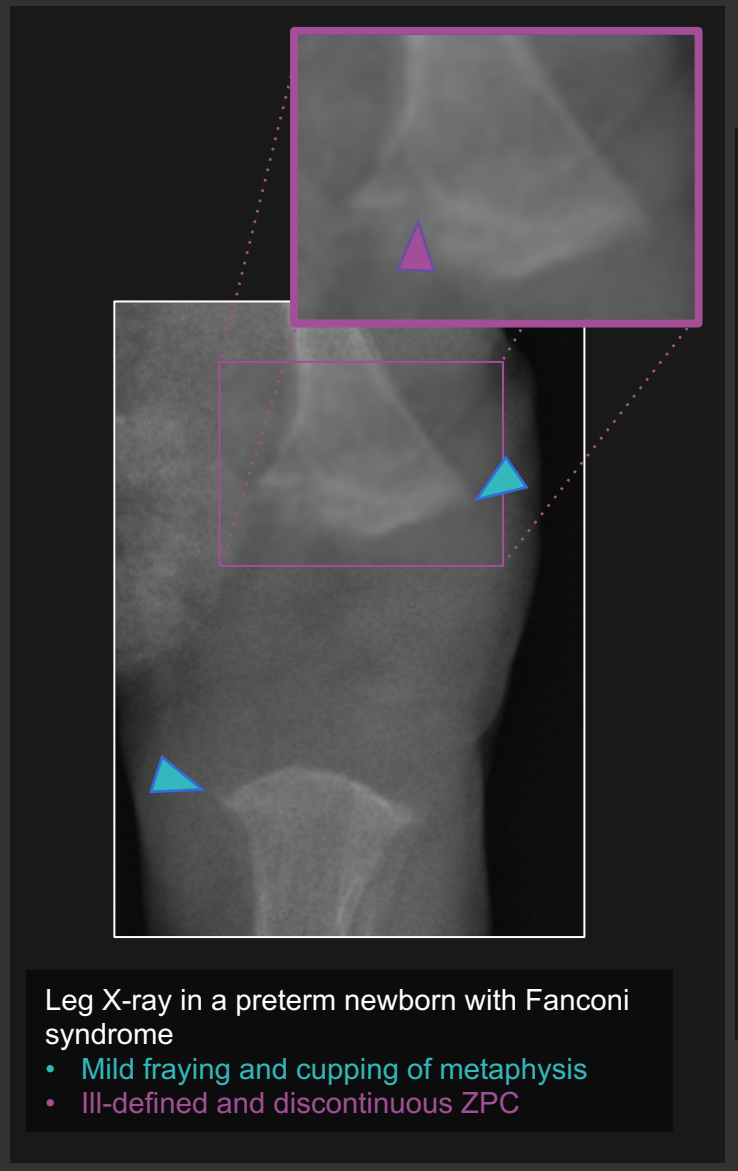
Radiographic Findings:

Periosteal reaction

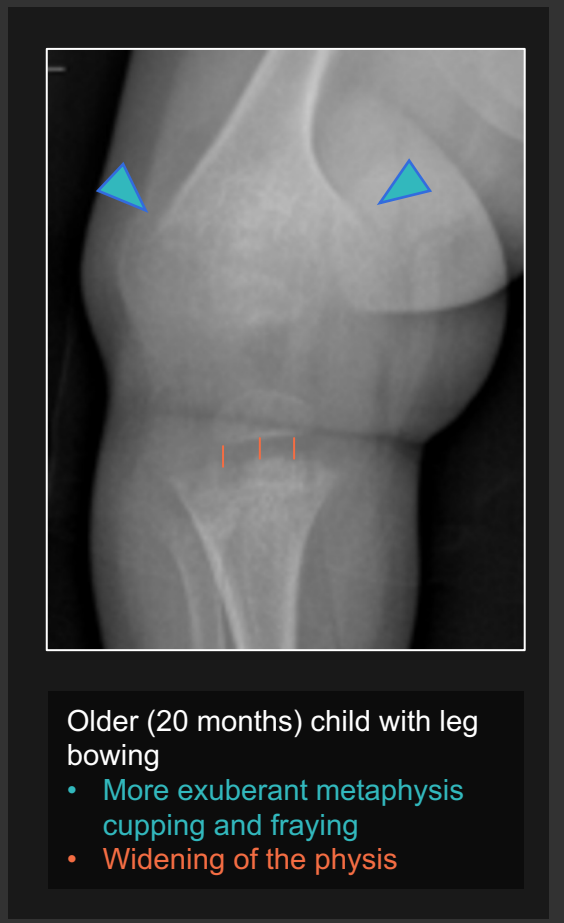
Metaphysis of fast-growing bones (limbs and ribs):

- Widening of the physis
- Fraying and cupping of the metaphysis
- **Decreased mineralization / discontinuity of the zone of provisional calcification (ZPC)**

Rickets differs from osteomalacia, which is actually a failure of osteoid to mineralize and can affect both adults and children



Leg X-ray in a preterm newborn with Fanconi syndrome
- Mild fraying and cupping of metaphysis
- Ill-defined and discontinuous ZPC



Older (20 months) child with leg bowing
- More exuberant metaphysis cupping and fraying
- Widening of the physis

Metabolic Bone Disease of Prematurity

Clinical Summary:

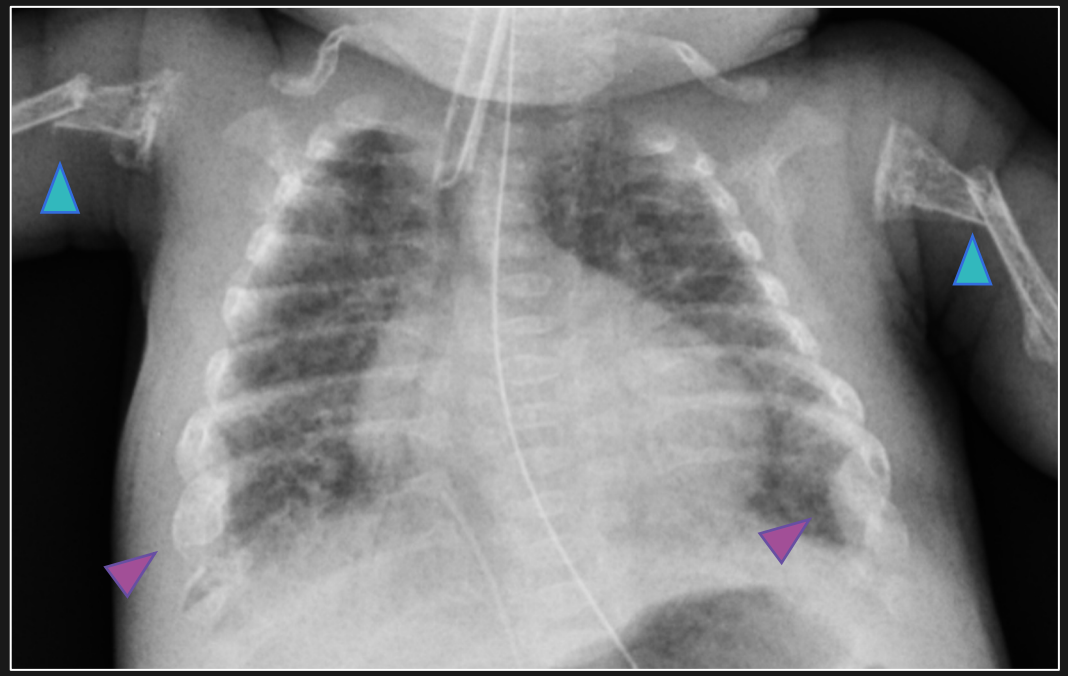
Metabolic Bone Disease of Prematurity (MBDP) refers to the undermineralization of the preterm skeleton. Patients with MBDP can present with both osteopenia and rickets

MBDP is multifactorial. However, as the third trimester is the period in which over 80% of skeletal calcium accumulation occurs, **gestational age is considered the strongest independent factor**

MBDP Risk Factors
Prematurity
Low birth weight
Reduced physical activity
Medication: - Antacids - Glucocorticoids - Loop diuretics
Parenteral nutrition
Other neonatal diseases: - Necrotizing enterocolitis - Bronchopulmonary dysplasia

Radiographic Findings:

- Bone rarefaction
- ZPC ill-definition / discontinuity
- Cortex Thinning
- Fraying and cupping of the metaphysis



Chest X-ray in a 1-month-old preterm (born at 24 weeks with 980 g)
- Diffuse Bone Rarefaction
- Bilateral Humeral Fractures
- Anterior Rib Cupping

Infantile Cortical Hyperostosis / Caffey's Disease

Clinical Summary:

Infantile Cortical Hyperostosis / Caffey's disease (ICH) is a rare disease usually occurring between 6-11 months of age

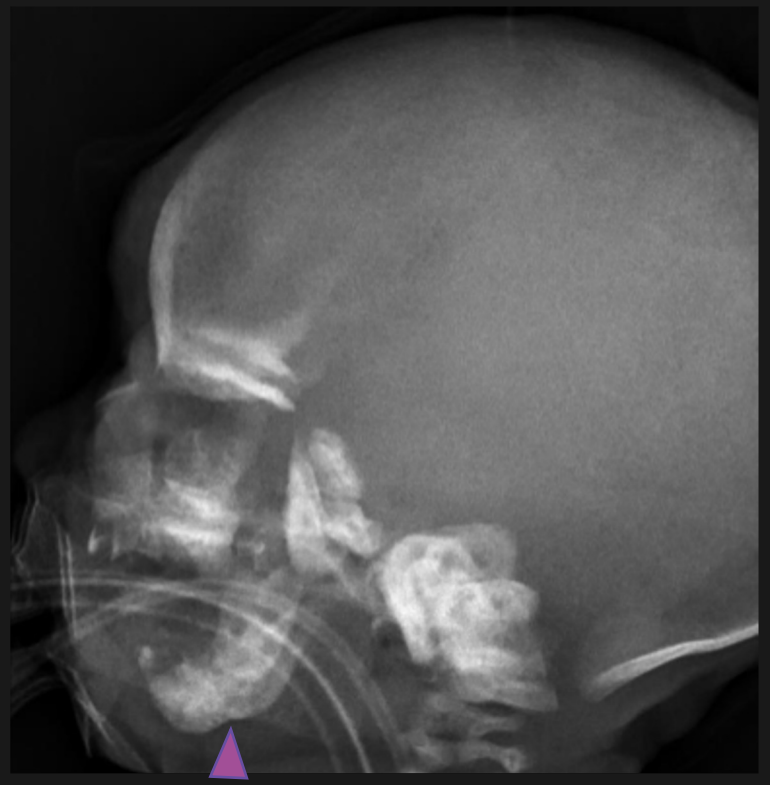
ICH presents with soft tissue swelling (face and/or extremities), fever and irritability. It is usually a self-limited disease that spontaneously resolves by 6-9 months of age in most cases

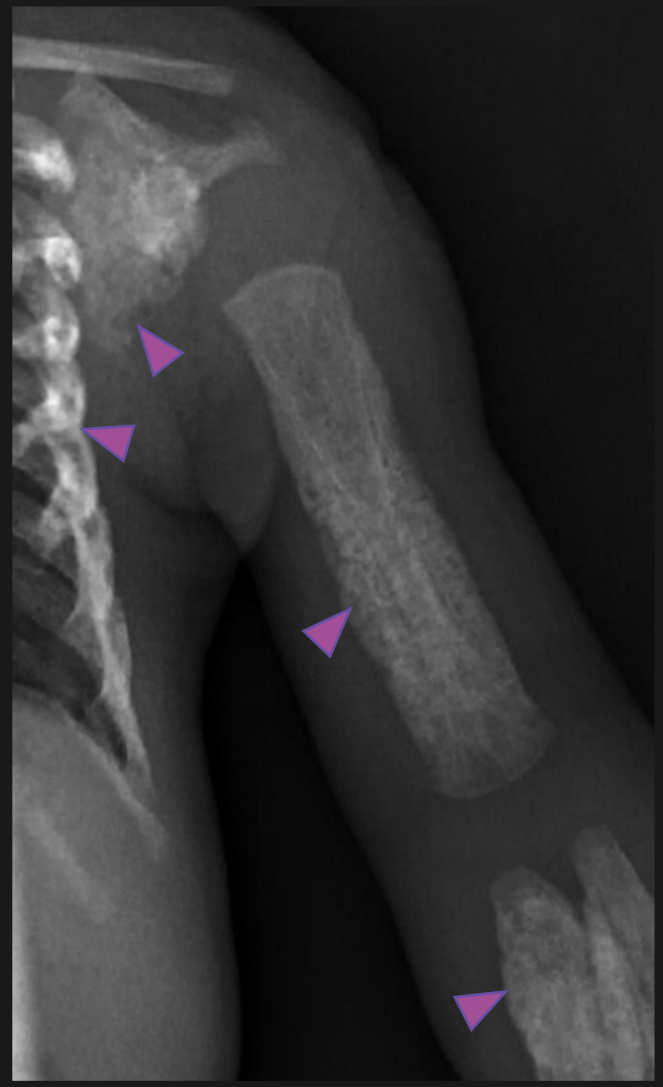
ICH usually affects mandibles (up to 80% of cases), ribs, clavicles, scapula and diaphysis of long bones

Radiographic Findings:

Diffuse or asymmetric subperiosteal bone deposition, which expands the bone and cortical hyperostosis

In latter phases the cortical hyperostosis is incorporated, which leads to widening of the medullary and can even evolve into limb bowing





Five day old preterm with diffuse swelling of the face and limbs and irritability.

- Extensive cortical hyperostosis involving the mandible, ribs, scapula, humerus, radius and ulna

Osteopetrosis

Clinical Summary:

Osteopetrosis, also known as Albers-Schönberg disease or marble stone disease is a rare genetic condition in which there is abnormal osteoclast function, leading to:

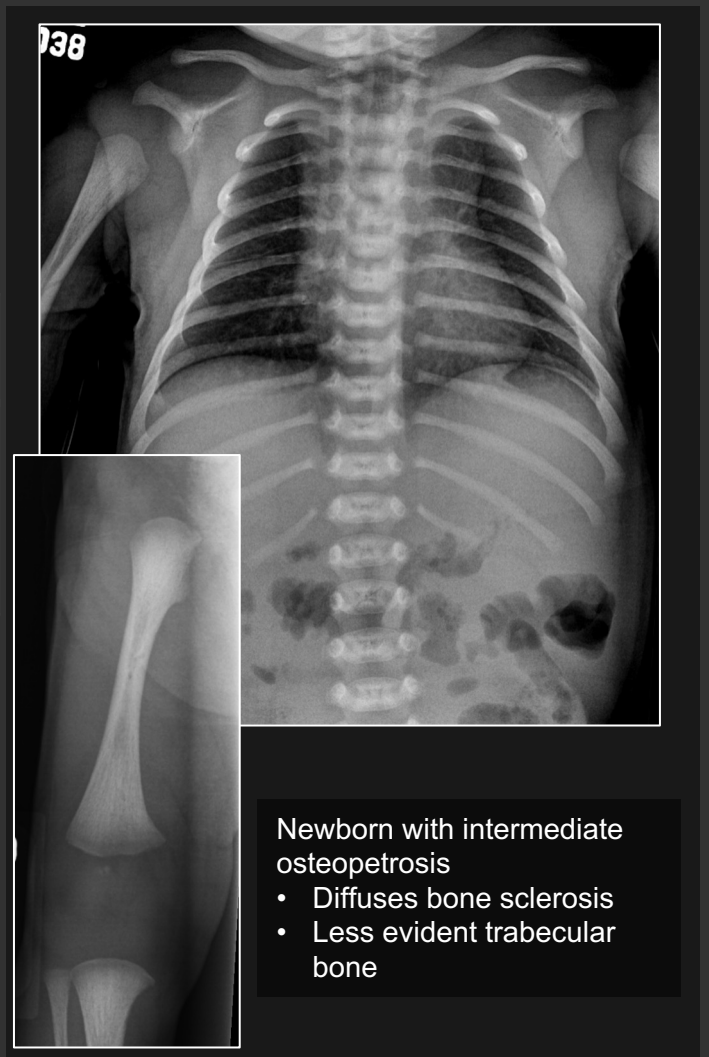
- Greater bone formation than destruction
- Fragile bones
- Bone marrow failure

Osteopetrosis is usually divided into three types:

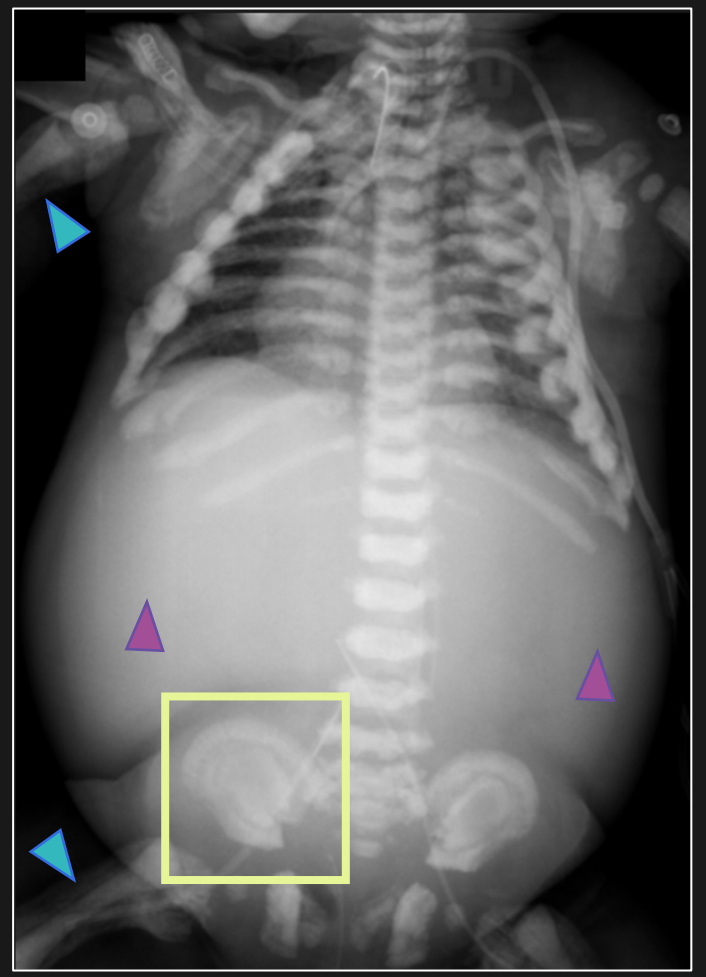
Infantile Type ("Malignant")	Intermediate Type	Adult Type ("Benign")
Autosomal Recessive More aggressive & high mortality Pancytopenia, Hepatosplenomegaly, immunosuppression	Autosomal Recessive Symptoms less severe than in infantile type Usually lives into adulthood.	Autosomal Dominant Milder form Anemia, pathologic fractures and early onset osteoarthritis

Radiographic Findings:

- Involvement of entire skeleton
- Uniformly sclerotic bones (epiphysis, metaphysis and diaphysis)
- "Bone within bone" appearance
- Undertubulation of long bones
- Thickening and sclerosis of cranial



Newborn with intermediate osteopetrosis
- Diffuses bone sclerosis
- Less evident trabecular bone



Newborn with infantile osteopetrosis
- Diffuses bone sclerosis
- "Bone within bone" aspect
- Undertubulation (broad femoral and humeri diaphysis
- Hepatosplenomegaly

Amniotic Band Syndrome

Clinical Summary:

Parts of the fetal body get constricted by bands of amniotic tissue (usually sporadic)
Presentation will depend mainly on two factors:
Part of the body that is affected
Gestational period in which entanglement occurs

Radiographic Findings:

Extremities are the most affected region:

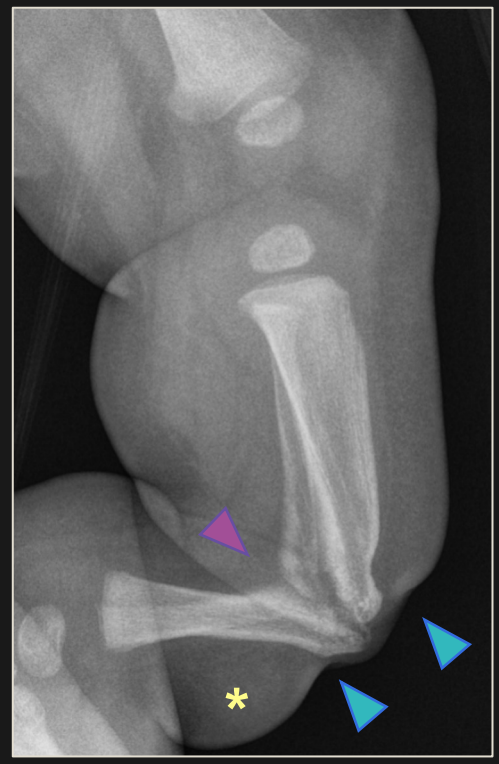
- Amputation
- Ring like constriction of soft tissue
- Pseudosyndactyly
- Distal lymphedema

Head:

- Acrania
- Anencephaly
- Facial clefts
- Asymmetric encephaloceles

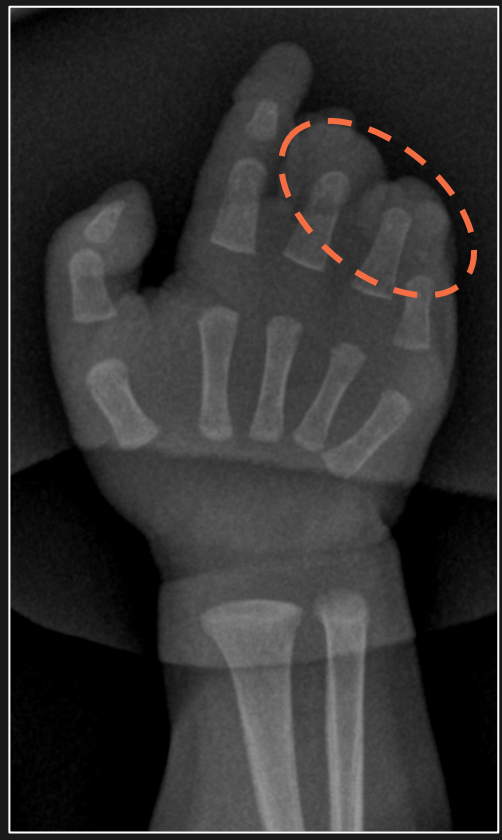
Trunk:

- Abdominal wall defects



Left leg X-ray in a newborn lower limb length discrepancy

- Pronounced angulation of both tibia and fibula with adjacent soft tissue constriction
- Distal enlargement of soft tissue



Left hand X-ray in a newborn with finger webbing and amputations

- Absence of the distal phalanxes of the second to fifth fingers
- Absence of the medium phalanxes of the third to fifth fingers
- Deformity of the soft tissue of the extremity of the second to fifth fingers

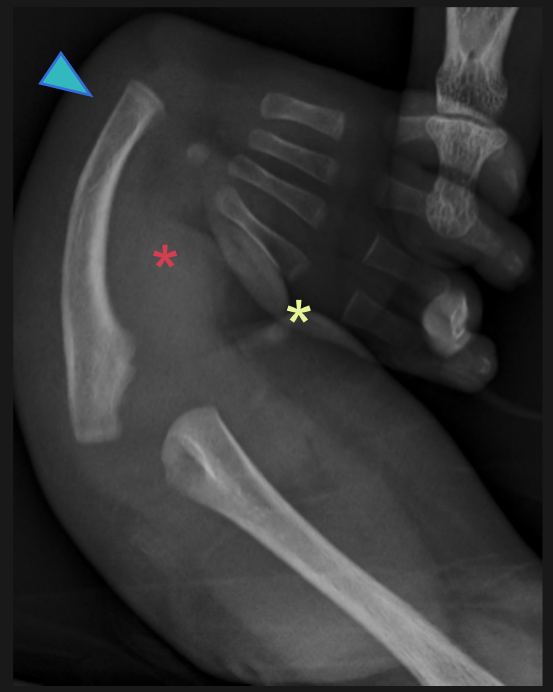
Hemimelia / Congenital Longitudinal Deficiency

Clinical Summary:

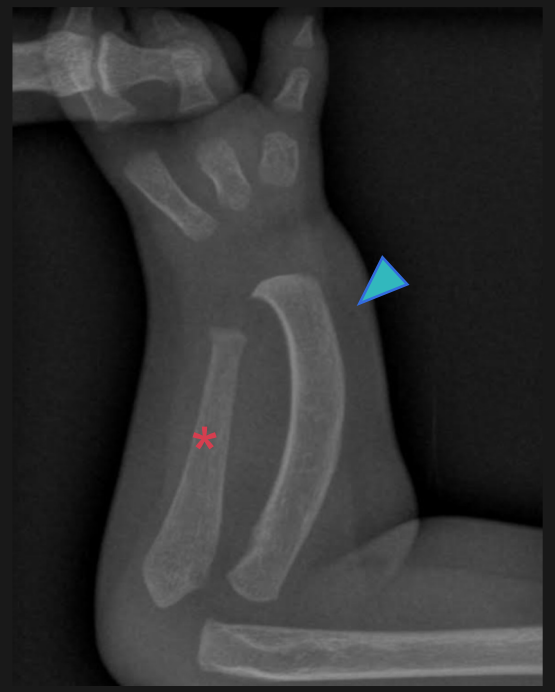
Hemimelia can also be termed as “congenital longitudinal deficiency” and consist of a congenital underdevelopment of the distal part of a limb.
It can be unilateral or bilateral and affect the tibia, fibula, ulna or radius
It is usually an isolated findings but may be associated with other skeletal anomalies or specific syndromes
Clinical presentation is often obvious at birth with limb asymmetry or deformity

Image Findings

There can be partial or complete absence of the affected bone
It is important to also evaluate other associate malformations, specially in the same limb



Newborn with left forearm deformity – Radial Hemimelia
- Complete absence of radius
- Bowing of the ulna
- Associated first finger malformation



Newborn with right forearm deformity – Ulnar Hemimelia
- Hypoplastic ulna
- Bowing of the radius
- Associated metacarpophalangeal malformation

Proximal Femoral Focal Deficiency

Clinical Summary:

Congenital defect of the primary ossification center of the proximal femur

Clinical findings: Limb shortening with leg abduction, flexion and external rotation. Hip and knee instability

Radiographic Findings:

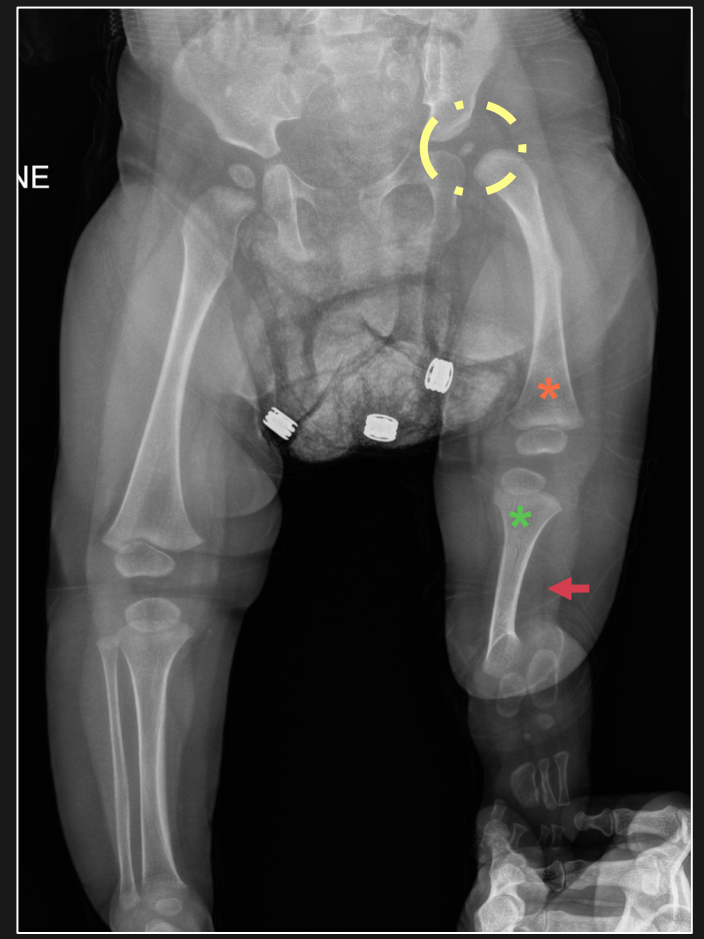
Aitken classification:

- A (38%): acetabulum and femoral head present
- B (32%): acetabulum and femoral head present. Delayed capital femoral epiphysis ossification
- C (17%) Acetabulum severely dysplastic. Absence of femoral head
- D (13%) Acetabulum and femoral head absent. Extremely short shaft

<u>MR is essential for cartilage evaluation and can affect treatment / prognosis</u>

Fibular deficiency/hemimelia associated in 70% of cases

- Congenital absence or shortening of the fibula associated with anteromedial bowing of tibia



Leg X-ray in a 4-month-old with left leg shortening
Aitken type B PFFD with fibular defiance
Shortening of the left femur
Acetabulum and femoral head present
Absence of the left fibula
Shortening and bowing of the left tibia

Developmental Dysplasia of the Hip (DDH)

Clinical Summary:

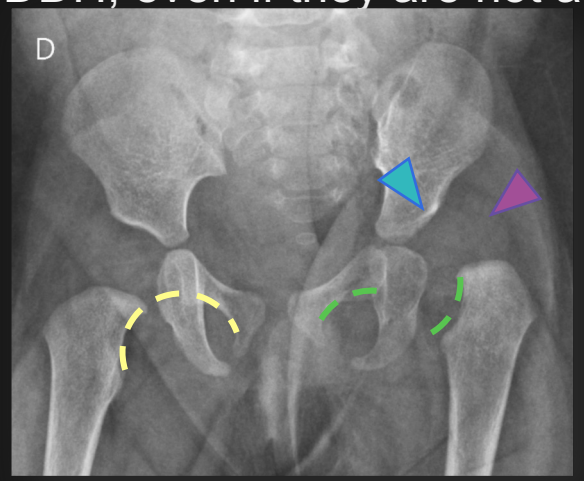
Incidence of DDH in children is estimated in 11.5/1000 live births in the US

In the newborn DDH is usually asymptomatic and diagnosis is made through screening exam, mains risk factors include:

- Female gender, breech position, and positive family history

<u>Ultrasound is the main imaging modality for screening/diagnosis in children below 6 months</u>

Frontal hip radiograph can however depict alterations in DDH, even if they are not as evident as in ultrasound



Hip X-ray in a 2-month-old girl with leg length discrepancy and confirmed DDH with femoral head luxation

- Shallow acetabulum
- Normal proximal femur
- Disruption of Shenton Line

Radiographic Findings:

Hilgenreiner line (Y line):

- Horizontal line connecting the triradiate cartilages

Perkin line:

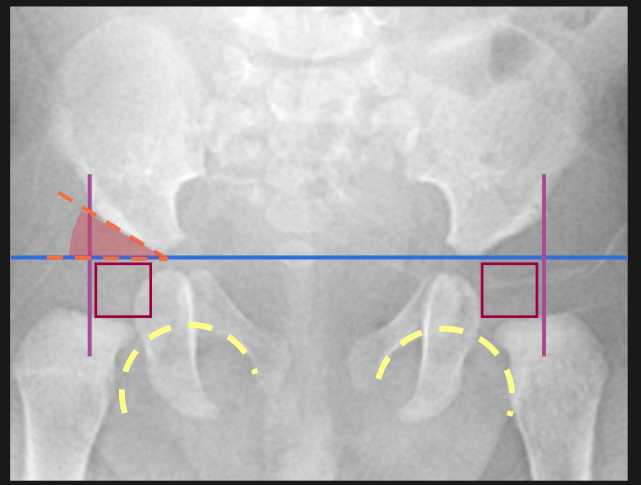
- Perpendicular to Y line, tangent acetabular roof

Shenton line:

- Curve along border of pubic ramus and femoral neck

Acetabular Angle:

- Angle between Y line and acetabular roof



Normal Hip X-ray in a 3-week-old girl.

- Hilgenreiner
- Perkin

Femoral head should center in the lower mid quadrant formed by these two lines

- Shenton
- Acetabular Angle
  - Should be < 30° at birth

Early Congenital Syphilis

Clinical Summary:

In Utero transmission of Treponema pallidum

- Early congenital syphilis: Onset before 2 years of age
- Late congenital syphilis: Onset after 2 years of age

Congenital Syphilis cases nearly quadrupled between 2015-2019

- Fetal/perinatal demise in up to 40% of cases

Radiographic Findings:

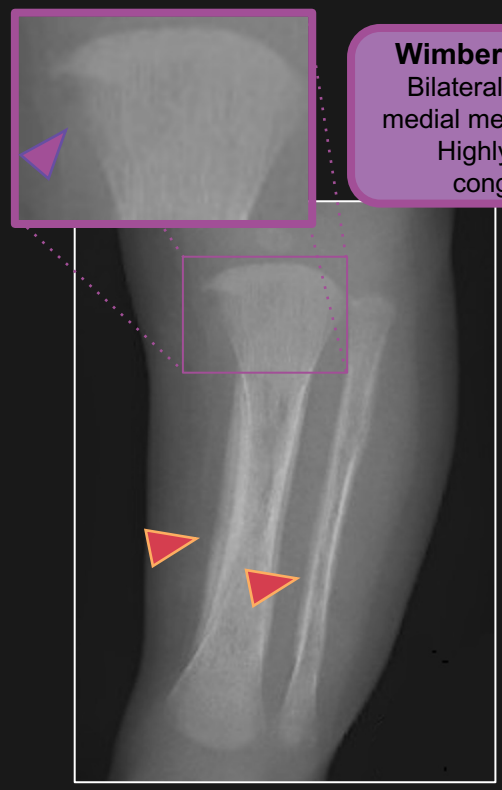
Involvement is typically widespread and symmetric, most commonly affecting long bones

Diaphysis:

- Diffuse periosteal reaction (most common finding)

Metaphysis:

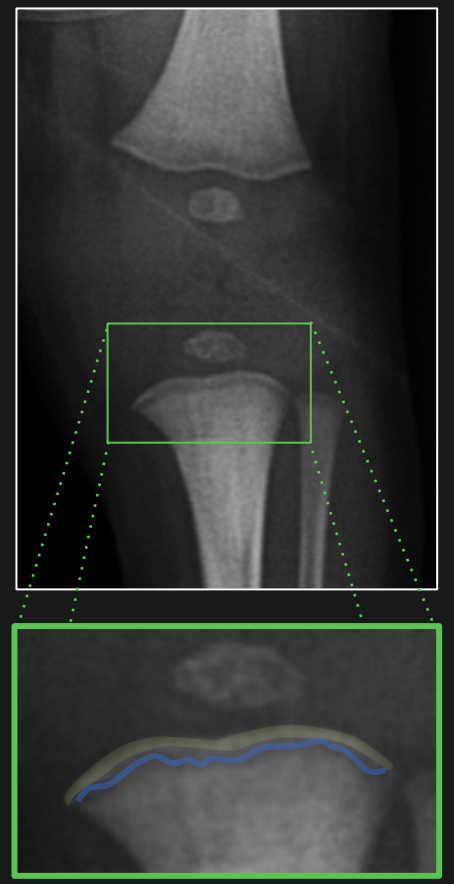
- Horizontal lucent bands (spares the zone of provisional calcification - ZPC)
- Osteolytic lesions in tibia, femur or humerus



Wimberger corner sign: Bilateral destruction of the medial metaphysis of the tibia. Highly suggestive for congenital syphilis

Leg X-ray in a newborn with confirmed congenital syphilis

- Extensive periosteal reaction in the tibia and fibula diaphysis
- Destruction of medial metaphysis of the tibia (Wimberger sign)



Leg X-ray in a second newborn with confirmed congenital syphilis

- **Horizontal lucent bands** sparing the **ZPC (thin continuous radiodense band)**

Osteomyelitis

Clinical Summary:

Acute osteomyelitis is very rare in neonates but can be catastrophic. Bone and joint infection has an incidence of 0.1 per 1000 births with a mortality rate of 7%.

Risk factors include septic arthritis (usually iatrogenic) and preterm (especially due to the use of invasive devices)

Most commonly affects the metaphysis of long bones (hematogenous infection)

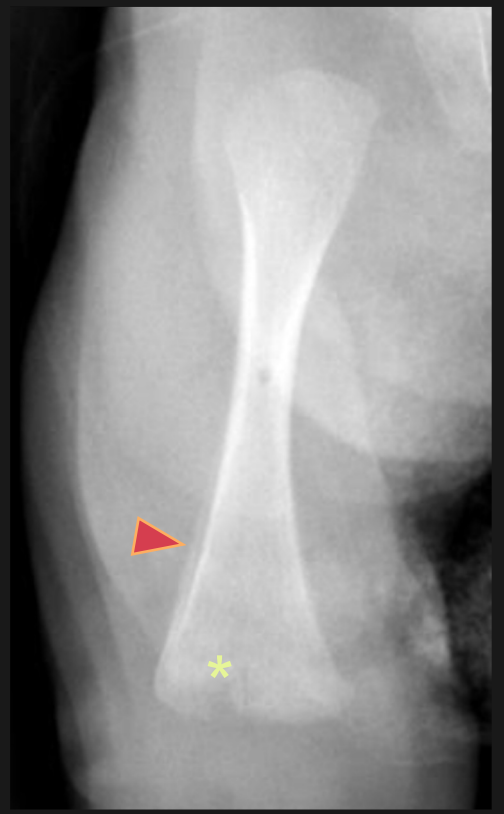
Diagnosis can be very challenging as symptoms are often unspecific

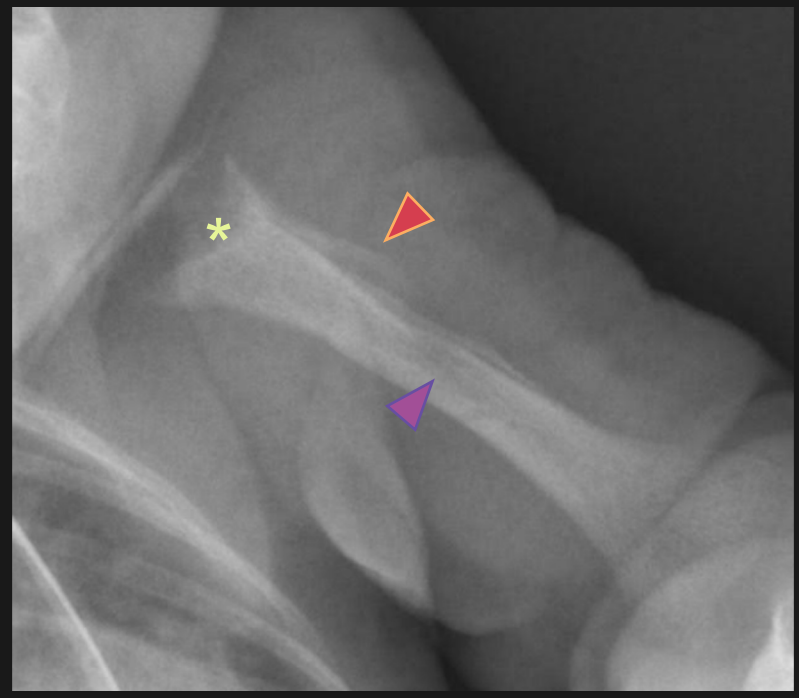
Radiographic Findings:

Th Imaging findings can be very subtle/absent until the 5-7th day of infection and include:

- Soft tissue swelling
- Join effusion
- Lytic lesions
- Periosteal reaction

MRI and Nuclear Medicine are more sensitive and specific for osteomyelitis, with abnormalities being evident earlier than on X-rays





Two different newborns with long bone osteomyelitis

- Discontinuous periosteal reaction.
- Metaphyseal lytic lesions
- Diaphyseal lytic lesion.

Take Home Messages

Periosteal reaction in the first months of life is not always related to a pathologic process

While mechanical birth-related trauma may lead to fractures, it is always important to consider other differential diagnosis such as physical abuse and metabolic bone disease of prematurity

Diffuse bone diseases in the newborns have a specific list of differential diagnosis

Bone infections on the neonates can have subtle radiographic presentation. Early radiologic findings can be key in preventing serious complications

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