

An Atlas of Hand Radiographs in Pediatrics

Prof. Kakarla Subbarao



KREST

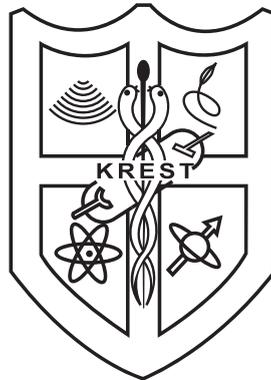


Wilhelm Conrad Roentgen
(1845 - 1923)



Reproduction of original radiograph of the
hand of **ANNA BERTHA LUDWIG**
"His wife" (Dec. 1895)





KREST

An Atlas of

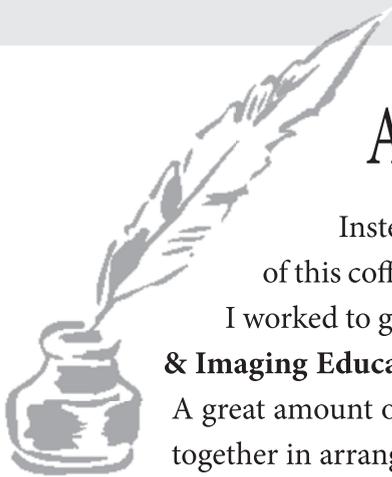
Hand Radiographs in Pediatrics

Prof. Kakarla Subbarao

Kakarla Subbarao Radiological & Imaging Educational Sciences Trust (KREST)

OBJECTIVES:

- ❖ To promote the practice of radiological and imaging sciences education, training and research
- ❖ To build institutions for training in radiology and sonography
- ❖ To network Indian and International radiologists
- ❖ To extend cost effective radiological services to the poor
- ❖ To initiate distance education through teleradiology services
- ❖ To create information and documentation services for exchange of data and expertise
- ❖ To Update professional skills through contact programmes.



AUTHOR'S FOREWORD

Instead of calling myself an author, the better term may be ‘compiler’ of this coffee-table book. I owe it to many radiologists and institutions where I worked to get the material. Establishment of **Kakarla Subbarao Radiological & Imaging Educational Sciences Trust (KREST)** enabled me to have a museum. A great amount of gratitude is due to **Dr B Rama Subba Raidu** for working hours together in arranging the films in a particular order for the benefit of the residents. This helped the examine going residents a great deal. Most of this material is from **KREST** museum. I acknowledge the help of **Prof. S Rammurthi** and **Prof. Sujatha Patnaik** for providing me with all the interesting cases from NIMS. I also thank the residents at KIMS and **Dr Joe Thomas** rheumatologist from Kochi for sending all the challenging cases. Finally, I thank **Mr G Bharath Kumar** for preparing the manuscript. I also thank **Mr D Venkatesh** & his team of **Kalajyothi** for designing this booklet.

It is well nigh impossible to include every entity regarding pediatric hands. Children have their own congenital anomalies, dysplasias and syndromes. This atlas will certainly enhance the knowledge of radiologists and pediatricians, present and future. It is said “a picture speaks thousand words”. This atlas provides maximum number of illustrations and minimum number of words to explain. Hand radiograph is only an initial study pointing out to other investigations to be followed. Plain radiography is still the foundation upon which all the other sophisticated imaging is built.

“I would give great praise to the physician
whose mistakes are small,
for perfect accuracy is seldom to be seen”

- Hippocrates

Introduction

Each hand consists of 27 bones. These bones, reflect fairly and accurately, normal skeletal development, congenital and acquired systemic disorders of the peripheral skeleton. Conventional radiographs are adequate most of the time for initial diagnosis. At times, it may be necessary to perform advanced imaging specially for identification of early calcifications and soft tissue structures. Clinical history, laboratory data and skeletal survey whenever necessary are taken into consideration to confirm the diagnosis. In skeletal dysplasias and other lesions chromosome and genetic analysis are essential for diagnosis and counselling.

Anatomy

Hand anatomy is complex and intricate. This enables hands to do gross as well as precise functions. A total of 27 bones constitute the basic skeleton of the wrist and hand [8 carpals, 5 metacarpals, 14 phalanges (3 for each digit except thumb which has 2)]. The hand is innervated by the median, ulnar, and radial nerves — each of which has sensory and motor components. The blood supply is from radial and ulnar arteries. The muscles of the hand are divided into intrinsic and extrinsic groups. The sesamoids vary with age and develop full potential after maturation

Sections - Pediatric hand radiographs

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Section-1

Bone age



Normal
10 yrs



Normal
30 yrs

Many congenital, developmental and systemic disorders affect the bones of the hand. Interpreting a hand radiograph includes not only the bones but the joints and soft tissues surrounding them. Although, ultrasonography and magnetic resonance imaging are the best modes of imaging of soft tissues, this Atlas deals with conventional radiographs. The posterior anterior views of the hand are adequate. Occasionally oblique and lateral views are added. Radiographs of both hands and wrists are included only, where ever necessary to make a definitive diagnosis. Occasionally advanced cross sectional images are included for better learning

Bone age assessment from radiographs of the hands

Bone age determination is made from radiograph of right hand in right handed people and of left hand in left handed people.

It is not wise to rely on any single area of the skeleton to assess bone age. Several areas must be assessed and the bone age is interpreted as a whole.

Bone age is an important parameter when children with growth disorders are investigated, and it is the basis for calculation of height prediction. Hand-wrist radiographs have been used for determination of maturation and subsequent evaluation of growth potential during preadolescence and adolescence.

Several methods have been developed for assessment of skeletal age on hand-wrist films. For evaluation of skeletal age, the Greulich-Pyle and Tanner-Whitehouse methods are generally used in clinical practice. The Greulich-Pyle atlas focuses on a number of maturational indicators, which represent stages of bone development or ossification events specific for each age, whereas Tanner-Whitehouse composite scores are based on osseous stages and events at each level.

Hand and Wrist

Carpal bones are much more erratic in development than tubular bones

2 yrs



8 yrs



Carpal Centres

Age in years - centres

1	-	2
2	-	3
3	-	4
4	-	5
5	-	6
10 - 11		7

+ /- few months

Skeletal Maturation

Advanced

- ❖ Adrenogenital Syndrome
- ❖ Adrenal hyperplasia
- ❖ Albright Syndrome
- ❖ Cerebral gigantism
- ❖ Hypergonadism
- ❖ Hyperthyroidism
- ❖ Low flow AVM
- ❖ Neurofibromatosis
- ❖ Pinealoma
- ❖ Tuberous sclerosis
- ❖ Miscellaneous

Retarded

- ❖ Celiac disease
- ❖ Congenital heart disease
- ❖ Cushing – steroid therapy
- ❖ Dwarfism syndromes
- ❖ Hypogonadism
- ❖ Hypopituitarism
- ❖ Hypothyroidism
- ❖ Juvenile diabetes
- ❖ Malnutrition – rickets Etc.,
- ❖ Renal, Hepatic Disorders
- ❖ Miscellaneous

Accelerated bone age with obesity. The bone age is six years, while the chronologic age is two years. Eg. adrenal hyperplasia, cushing syndrome, hypothalamic hamartoma etc.,



**3 year old child with accessory ossification
Centers which generally signify skeletal age lag.**

Delayed bone age is due to several systemic and endocrinal causes.



**Congenital
hypothyroidism (Cretin)
Delayed ossification
centres in a fifteen yr old.
Bone age is 10 yrs.**

Congenital Anomalies

Congenital - anomalies may be isolated or may be part of systemic disease.

Ulnar dimelia – Mirror hand



Ulnar dimelia

- ❖ **Rare**
- ❖ **Symmetric duplication of the limb in midline**
- ❖ **A central digit with 3 digits (long, ring, little) on either side**
- ❖ **Total 7 digits, but thumb is absent**
- ❖ **Two ulnae, no radius (Ulnar dimelia)**
- ❖ **Due to transplantation/replication of ZPA**



Dimyelia



Bifid first
metacarpal



Partial duplication of
thumb
(? Wassel type V)



12 M, Lunate
triquetrum coalition



Hamate capitate coalition

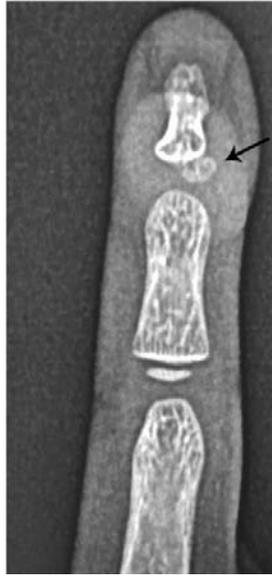


Coalition of triquetrum
and lunate

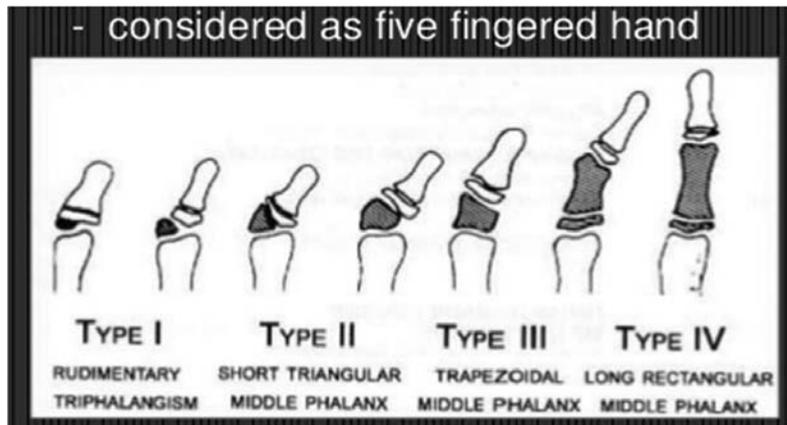


Duplication of index
finger

Triphalangeal Thumb



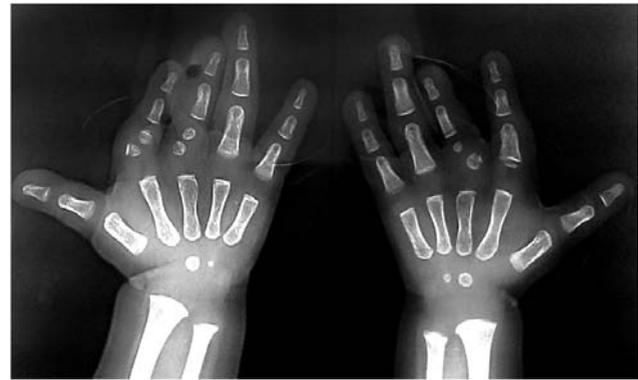
- ❖ AD
- ❖ Extra phalanx of variable size, variable shape (triangular/trapezoid/rectangular) normal appearing thumb
- ❖ Fully developed extra phalanx lying in the finger plane



From internet



Syndactyly bilateral –
no known etiology

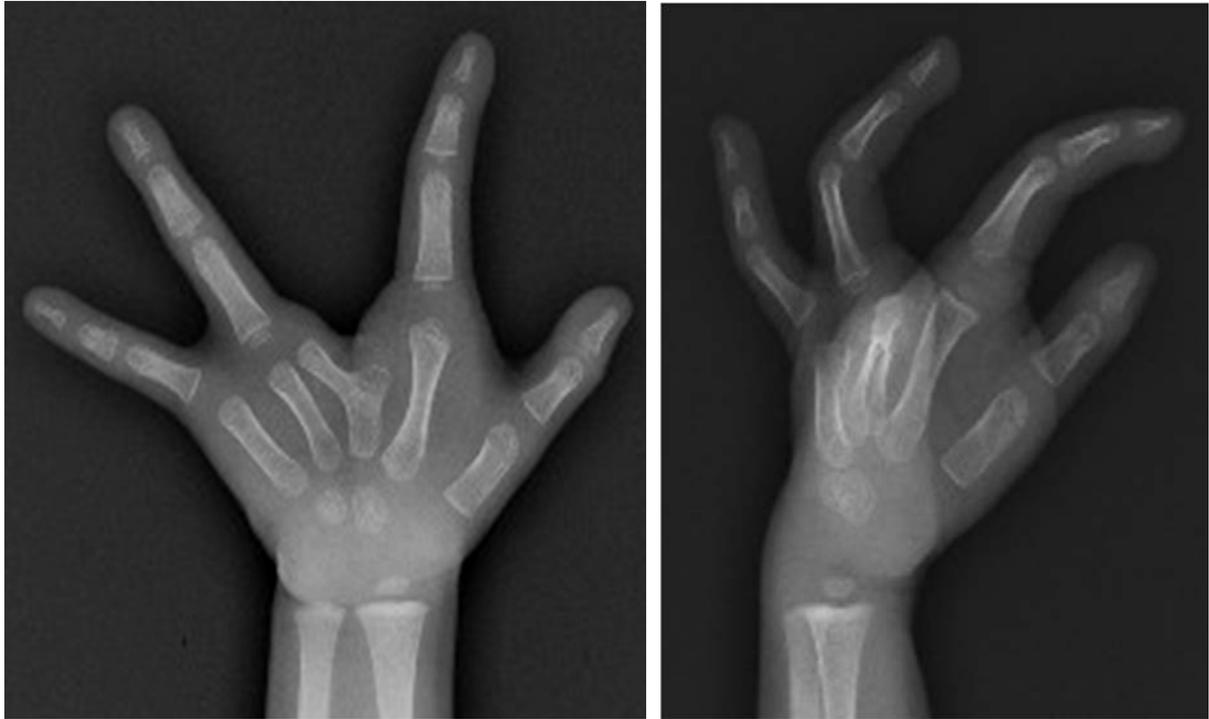


Ectrodactyly

Ectrodactyly represents a split hand, cleft hand, derived from the Greek *ektroma* (abortion) and *daktylos* (finger) involves the deficiency or absence of one or more central digits of the hand or foot and is also known as split hand/split foot malformation (SHFM).



2M – Ectrodactyly



Split hand

Congenital split hand & foot



split hand

A rare form of a congenital disorder in which the development of the hand is disturbed. ... Split hand/foot malformation (SHFM) is characterized by underdeveloped or absent central digital rays, clefts of hands and feet, and variable syndactyly of the remaining digits.



split hands & feet

Phalangeal synostosis (symphalangism)



Symphalangism refers to ankylosis of the interphalangeal joints (i.e. fusion of the phalanges) in either the toes or the fingers. Less commonly, the metacarpophalangeal joints may be affected.



Madelung deformity



Madelung

Normal

Radial & palmar angulation of distal radius due to growth disturbance of Ulnar & palmar part of distal radial physis.



It is due to premature closure or defective development of the ulnar third of the distal epiphysis of the radius. This deformity results in a radial shaft that is bowed with increased interosseous space and dorsal subluxation of the distal radioulnar joint. It can be bilateral in 50-66% of patients.

Associated disorders – Madelung and pseudo Madelung

- ❖ **Leri-Weill syndrome**
- ❖ **Turner syndrome**
- ❖ **Nail-patella syndrome**
- ❖ **Hereditary, multiple exostosis**
- ❖ **Ollier's disease**
- ❖ **Achondroplasia**
- ❖ **Multiple epiphyseal dysplasias**
- ❖ **Mucopolysaccharidoses
(Hurler and Morquio syndromes)**
- ❖ **Neurofibromatosis 1**

Madelung deformity



Leri weill dyschondrosteosis is a skeletal dysplasia characterized by short stature and an abnormality of the wrist bones called Madelung deformity.

Lateral views of both forearms



Ollier's dyschondroplasia involving the distal radius

In Leri Weill syndrome, Madelung deformity presents as a spectrum of findings. It may affect the entire radius or it may affect only the distal radius. Extremities with involvement of the entire radius have a shorter radius and ulna, decreased height, and a more severe deformity of the extremities than with involvement of only the distal radius.

Radiological findings of Madelung deformity

- ❖ Dorsal and radial bowing of the radius
- ❖ Exaggerated palmar (up to 35°) and ulnar tilt (up to 60°) of the radiocarpal articulation
- ❖ Failure of ossification of the ulnar side of the distal radial epiphysis
- ❖ Exaggerated radial inclination
- ❖ Decreased carpal angle below 118° ; normal from 118° to 139° .
- ❖ Carpal subluxation in a palmar and ulnar direction
- ❖ Lunate is gradually forced to the apex of the V-shaped radioulnar carpal joint
- ❖ “V-shaped” proximal carpal row = herniated proximal carpal row.
- ❖ Dorsal subluxation of the distal ulna
- ❖ Elongation of the ulna with positive ulnar variance
- ❖ Wedging of the carpus between the radius and ulna



Turner syndrome
(Monosomy X)
V shaped proximal carpal row
Carpal angle 130 degrees
Associated with coarctation
of aorta

Pseudo madelung



Congenital



Congenital



Congenital

- ❖ **No dorsal subluxation of the distal ulna and positive ulnar variance**
- ❖ **No wedging of the carpus between the radius and ulna**
- ❖ **No dorsal and radial bowing of the radius**
- ❖ **No failure of ossification of the ulnar side of the distal radial epiphysis**
- ❖ **No “V-shaped” proximal carpal row (herniated proximal carpal row)**

Idiopathic - Congenital



REVERSE MADELUNG DEFORMITY

Note the negative ulnar variance associated with Kienbock disease and ulnar impingement syndrome. However, in young children there may not be any change in the lunate.

Ulnar variance

Also known as Hulten variance refers to the relative lengths of the distal articular surfaces of the radius and ulna.

Neutral - both the ulnar and radial articular surfaces at the same level.

Positive - Ulna projects more distally. The level of the ulna is $> 2.5\text{mm}$ beyond the radial articular surface.

Negative - Ulna projects more proximally. The level of the ulna is $\leq 2.5\text{mm}$ below the radial articular surface.

This variance is independent of the length of the ulnar styloid process.



Neutral



+ve ulnar variance



-ve ulnar variance

No changes of the lunate at this stage



Negative ulnar variance



Negative ulnar variance

In general, ulnar variance particularly negative ulnar variance is frequently observed without any symptoms. Although it is incidental, follow-up studies were not conducted.

- ❖ **Negative Ulnar variance is associated with lunate malacia (Kienbock disease). Severe ulnar negative variance may result in ulnar impingement on the distal radius !**



Kienbock disease:

Only occurs after the fusion of the epiphyses. There are other causes of Kienbock apart from negative ulnar variance.

Eponyms such as this indicate the individual who made the original observations and were not recognized earlier.

Positive ulnar variance is associated with proximal lunate degenerative changes as indicated by the cyst formation. Also called ulnar impaction syndrome. It is associated with ulnar abutment.

Only occurs after the fusion of the epiphyses



Positive Ulnar Variance with cyst formation of lunate

Bayonet Hand Deformity



Congenital



Multiple
exostoses



Osteochondroma



Achondroplasia

Comprises of –

- ❖ Shortening of ulna
- ❖ Outward bowing of radius
- ❖ Subluxation of the inferior radioulnar joint

VACTERL - nemonic



Absent radius



V – Vertebral & vaginal defects

A – Anal atresia

C – Cardiac defects

TE – Tracheoesophageal

R – Renal, radial anomalies

L – Limb abnormalities

Patients diagnosed with **VACTERL** association typically have at least three of the above characteristic features.



Kirner syndrome

Progressive palmar radial curvature of the distal phalanx of little fingers bilaterally (deviation in 2 planes). Distortion & Widening of physeal plate along with curvature of the diaphysis of the distal phalanx.



Clinodactyly

Clinodactyly is a minor congenital malformation (birth defect). The basis for the clinodactyly is that the middle bone in the fifth finger is underdeveloped and instead of being rectangular, is wedge-shaped in a medio lateral plane. May be associated with many syndromes including Down syndrome, Klinefelter syndrome, Turner syndrome, Fanconi anemia and others.

Polydactyly



Polydactyly in utero - Ultrasonography

Central polydactyly – Three types: Type I is a central duplication, not attached to the adjacent finger by osseous or ligamentous attachments; it frequently does not include bones, joints, cartilage, or tendons. Type IIA is a nonsyndactylous duplication of a digit or part of a digit with normal components, and articulates with a broad or bifid metacarpal or phalanx. Type IIB is a syndactylous duplication of a digit or part of a digit with normal components and articulates with a broad or bifid metacarpal or phalanx. Type III is a complete digital duplication, which has a well-formed duplicated metacarpal.

Polydactyly – post axial and pre axial

More common

- ❖ Over segmentation
- ❖ Jeune syndrome (ATD)
- ❖ Ellis Van Creveld (CED)
- ❖ Trisomy 13
- ❖ Carpenter
- ❖ Lawrence – Moon – Biedl
- ❖ Meckel – Gruber
- ❖ Polysyndactyly



Super numerary and rudimentary thumb (Pre axial)

Ulnar polydactyly – The two-stage classification, according to Temtamy and McKusick, involves type A and B. In type A there is an extra little finger at the metacarpophalangeal joint, or more proximal including the carpometacarpal joint. The little finger can be hypoplastic or fully developed. Type B varies from a nubbin to an extra, non-functional little finger part on a pedicle. According to the three-type classification, type I includes nubbins or floating little fingers, type II includes duplications at the MCPJ, and type III includes duplications of the entire ray.

Radial polydactyly – The Wassel classification is the most widely used classification of radial polydactyly, based upon the most proximal level of skeletal duplication. The most common type is Wassel 4 (about 50% of such duplications) followed by Wassel 2 (20%) and Wassel 6 (12%).

Fibrodysplasia calcificans progressiva congenita

Formerly known as myositis ossificans progressiva



Note microdactyly of the thumbs due to fibrodysplasia. Similar changes are seen in the feet. Extensive calcification and ossification of muscles and soft tissue structures. A rare hereditary muscular problem.

Section-3

Skeletal dysplasias

Skeletal Dysplasias (SD) are clinically a heterogeneous group of genetic disorders characterized by the presence of generalized disorder of bone growth.

Sclerosing skeletal dysplasias

- ❖ **Osteopetrosis (Albers – Schonberg disease)**
- ❖ **Pycnodysostosis (Maroteaux – Lamy disease)**
- ❖ **Osteopoikilosis (Spotted bone disease)**
- ❖ **Osteopathia striata (Voorhoeve disease)**
- ❖ **Progressive diaphyseal dysplasia (Camurati - Engelmann)**
- ❖ **Melorheostosis**
- ❖ **Fibrous dysplasia – sclerosing type**
- ❖ **Overlap syndromes**

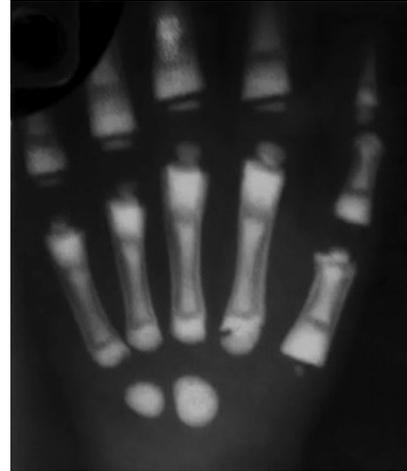
Osteopetrosis (Albers – Schonberg disease)

Four well established forms

1. Benign common form– autosomal dominant
2. Malignant form – autosomal recessive associated with death in childhood
3. Intermediate – Autosomal recessive
4. Osteopetrosis with renal tubular acidosis and cerebral calcifications

Well defined inborn error of metabolism namely deficiency of the carbonyl anhydrase 2 Isoenzyme - (Marble Brain disease)

A 5th variant with neuronal storage disease may also exist



Osteopetrosis
Bone in a bone
appearance



Intermediate form



Osteopetrosis with tubular acidosis



New born hand with rachitic changes



Autosomal recessive Osteopetrosis

Carbonil anhydrase deficiency

- 1. Osteopetrosis**
- 2. Tubular deficiency**
- 3. Cerebral calcifications**

Osteopetrosis and genes

Name	OMIM	Gene
OPTA1	607634 	<i>LRP5</i>
OPTA2	166600 	<i>CLCN7</i>
OPTB1	259700 	<i>TCIRG1</i>
OPTB2	259710 	<i>TNFSF11</i>
OPTB3	259730 	CA2 (renal tubular acidosis)
OPTB4	611490 	<i>CLCN7</i>
OPTB5	259720 	<i>OSTM1</i>
OPTB6	611497 	<i>PLEKHM1</i>
OPTB7	612301 	<i>TNFRSF11A</i>

Pycnodysostosis (Maroteaux – Lamy disease)

(Toulouse-Lautrec - French Artist had similar features)

A hybrid between osteopetrosis and Cleidocranial dysostosis. Major sites include skull, mandible, clavicles and spine. All the bones are uniformly sclerotic. However, skull findings include frontal bossing, persistent fontanel, wormian bones. An obtuse angle of mandible is noted. Clavicular hypoplasia and Acro osteolysis are present.

Gene location – CTSK^a



Pycnodysostosis



5 yr child with Pycnodysostosis



Osteopoikilosis

Osteopoikilosis is a benign, autosomal dominant sclerosing dysplasia of bone characterized by the presence of numerous bone islands in the skeleton. Often these are placed juxtaarticularly. They may assume the shape of round or oval.

Osteopathia striata is typically bilateral, although occasionally it can be unilateral, typically in tubular bones. Radiographically prominent vertical striations predominate in the metaphyses and epiphyses of the long bones (celery stalk metaphysis).



Osteopathia striata

DENSE VERTICAL METAPHYSEAL LINES

- 1. Congenital transplacental infections ('celery stalk' metaphyses) (eg, STORCH).**
- 2. Metaphyseal injury (localised).**
- 3. Osteopathia striata (Voorhoeve disease).**
- 4. Enchondromatosis (Ollier).**
- 5. Goltz S (Focal dermal hypoplasia).**
- 6. Hypophosphatasia.**
- 7. Metaphyseal chondrodysplasia.**
- 8. Mixed sclerosing bone dysplasia.**
- 9. Osteopathia striata with cranial sclerosis.**
- 10. Phenyl ketonuria**
- 11. Sponastrime dysplasia (Spondyloepimetaphyseal dysplasia).**

Progressive diaphyseal dysplasia

Camurati Engelmann disease presents during childhood with bilateral and symmetrical bone involvement, whereas ribbing disease may be unilateral and asymmetrical. In Engelmann's the skull is involved whereas in Ribbing's disease only long bones are involved. Engelmann is autosomal dominant while ribbing is autosomal recessive in fact both of them may represent phenotypic variation of the same disorder.

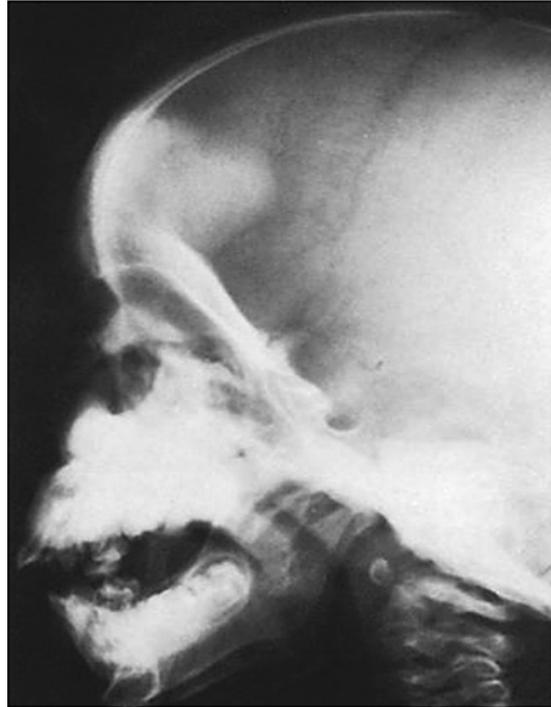


4 yrs

7 yrs

Note the dense, stubby metacarpals and phalanges

Craniodiaphyseal dysplasia (also known as CDD or lionitis) is an extremely rare autosomal recessive bone disorder that causes calcium to build up in the skull, disfiguring the facial features and reducing life expectancy.



Dense cortices with widening metacarpal phalanges.
Sclerosis of frontal and basicranium.

Fibrous dysplasia

Fibro osseous aberration by replacement of medullary area by fibrous tissue and new bone

- ❖ Monostotic
 - ❖ Polyostotic
 - ❖ Albright syndrome
 - ❖ Mazabaud syndrome (FD + myxoma of soft tissues)
-
- ❖ Ground glass / radiolucent area of trabecular alteration in long bones. Occasional calcifications
 - ❖ Bulging of the bone with endosteal scalloping & cortical thinning
 - ❖ Deformities

POLYOSTOTIC FIBROUS DYSPLASIA



Fibrous Dysplasia with Rickets explained by humoral mechanism, such as seen in oncogenic osteomalacia. Manufacture of polypeptides by mesenchymal tumors etc.,



Fibrous Dysplasia with Rickets



17 yrs

Melorheostosis, also known as Leri disease, is an uncommon mesenchymal dysplasia manifesting as regions of sclerotic bone with a characteristic dripping wax or flowing candle wax appearance. It may cross the joints and soft tissues may be involved. It may be monostotic or polyostotic

In infants, contractures of the fingers may be noted due to fibrosis and linear scleroderma



18 yrs

Nonsclerosing skeletal dysplasias

- ❖ **Chondroectodermal Dysplasia (Ellis Van Creveld Syndrome)**
- ❖ **Chondrodysplasia Punctata (Conradi-Hunermann)**
- ❖ **Multiple epiphyseal dysplasia**
- ❖ **Spondylo Epiphyseal Dysplasia With Polyarthropathy (Pseudo Rheumatoid Type)**
- ❖ **Metaphyseal Chondrodysplasia (Meta. Dysostoses)**
- ❖ **Spondylo metaphyseal dysplasia**
- ❖ **Spondylo Multiple epi metaphyseal dysplasia**
- ❖ **Pyle Disease -Frontometaphyseal Dysplasia**
- ❖ **Kniest dysplasia**
- ❖ **Cleidocranial Dysplasia**
- ❖ **Diastrophic dysplasia**
- ❖ **Mucopolysaccharidosis**

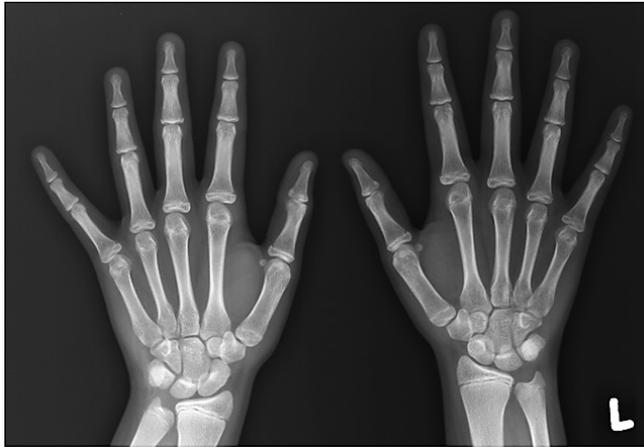
X Linked Chondrodysplasia Punctata (Conradi-Hunermann AD)



Rhizomelic – AR
Prominent metacarpals
Short humerus & femur

-
- ❖ **Stippled calci. ends of bones and ossification centers**
 - ❖ **Calci. Adjacent to vertebrae**
 - ❖ **Vertebral deformities**
 - ❖ **Asymmetric shortening of long bones**

Multiple epiphyseal dysplasia



It is a disorder of cartilage and bone development primarily affecting the ends of the long bones in the arms and legs (**epiphyses**).



**Epiphyseal dysplasia
- pseudo rheumatoid type**



16 M, Pseudorheumatoid



Molecular genetics: A novel mutation in WISP3 was detected
Interpretation: C145R is assumed to be a reason for progressive pseudorheumatoid dysplasia in this patient and confirms the diagnosis in this patient and provides specific Mutation information that can be used for carrier testing and prenatal diagnosis In the family.

Spondylo Epiphyseal Dysplasia With Polyarthropathy (Rheumatoid Type)



- ❖ Simulates R.A.
- ❖ No erosions as in R.A
- ❖ Large meta carpal heads
- ❖ Soft tissue swellings

Metaphyseal Chondrodysplasia (Meta. Dysostoses)

- ❖ Common
 - Schmid
 - Jansen
 - McKusick

Cartilage hair Dysplasia

- Meta, Chon. with panc.

Insuf and neutropenia (Schwachman)

- ❖ Uncommon
 - 10 types

Metaphyseal dysplasia



**Mckusick type of
metaphyseal dysplasia**



**Note the irregularity and
increased density of the
metaphysis**



Spondylo Metaphyseal Dysplasia

- ❖ **Peripheral arthropathy**
- ❖ **Retarded Carpal and Tarsal bones**
- ❖ **Large articular ends**
- ❖ **Metaphyseal changes**
- ❖ **Degenerative changes**
- ❖ **Spinal changes**

Spondylo epi metaphyseal dysplasia (Sponastrime dysplasia)



This is a rare type of skeletal dysplasia.

The differential diagnosis includes:

Congenital hypothyroidism (cretinism)

Chondrodysplasia calcificans

Congenita -

Mother on anticoagulant therapy

**Changes in the epiphyses and metaphyses,
vertebra are involved**

Pyle Disease -Frontometaphyseal Dysplasia

Involvement of metaphyses of bones of the feet, hands, ribs and clavicles. Thinning of cortices with widening, loss of corticomedullary differentiation, and ground glass opacity.



Widened metaphyses with ground-glass background

Kniest dysplasia

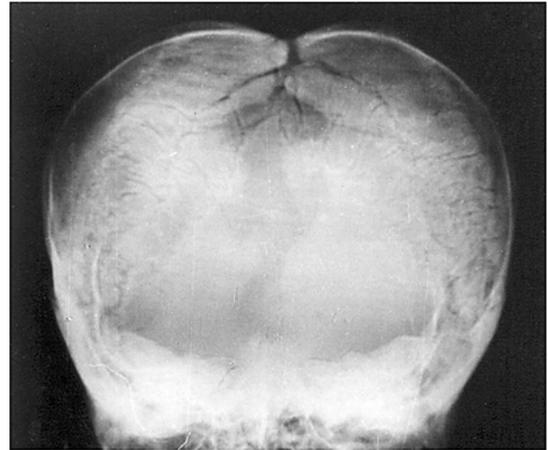


Kniest dysplasia includes broad ends of the bones at both poles, flattened metacarpal epiphyses, osteoporosis and a truncation of the tuft of the first digit. Carpal centers may also appear deformed after the age of three years. “Megaepiphyses” resemble pseudo rheumatoid type of spondylo epiphyseal dysplasia.

Cleidocranial Dysplasia



- ❖ **Pointing of tufts of terminal phalanges, acroosteolysis**
- ❖ **Brachymesophalangia of 2nd and 5th fingers (middle phalanges short)**
- ❖ **Long 2nd metacarpal with accessory epiphysis at the base**



- ❖ **Midline defects, Wormian bones in skull**

Diastrophic dysplasia - Hitch hiker's thumb



Diastrophic dysplasia is also called diastrophic dwarfism, a disorder of cartilage and bone development. Affected individuals have short stature with very short arms and legs. Most also have early-onset joint pain (osteoarthritis) and joint deformities called contractures, which restrict movement. The thumb is mobile and adducted.

Mucopolysaccharidosis – 7 types – Morquio, MPS IV

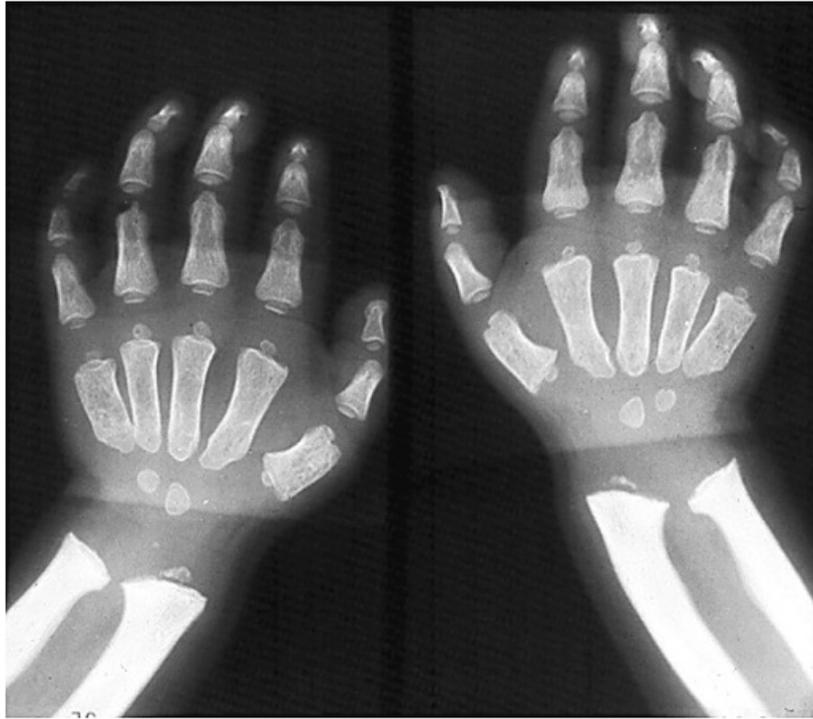


Central beaking
of the vertebral
bodies



- ❖ **Pointing of metacarpal bases**
- ❖ **Short metacarpals**
- ❖ **Carpal bones irregular**
- ❖ **Distal ends of radius and ulna are cup shaped and irregular**
- ❖ **Normal Mentation**

Hunter – MPS II A



Widening of the metacarpals with proximal pointing

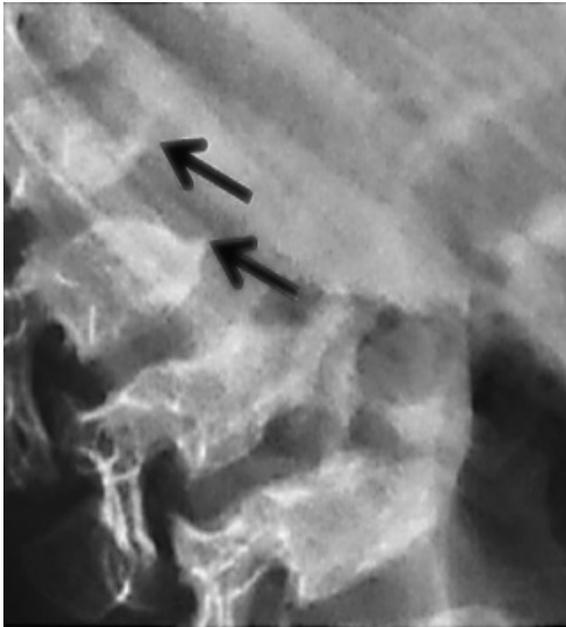
There is a deficiency in iduronate-2-sulfatase, which leads to an accumulation of heparan sulfate and dermatan sulfate. Hunter syndrome is X-linked recessive and primarily affects male subjects.

Hurler – MPS 1-H

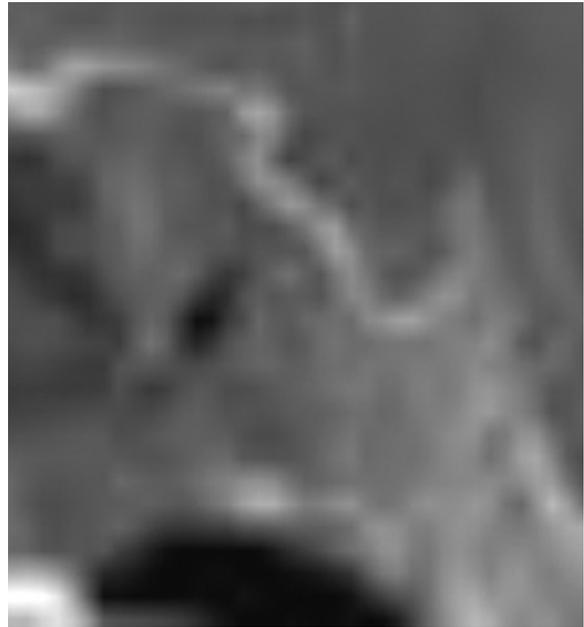


- ❖ Proximal pointed metacarpals
- ❖ Irregular, small carpals with delayed maturation
- ❖ Radioulnar surfaces slope towards each other
- ❖ Mental retardation
- ❖ **DISTAL PHALANGES ARE SMALL**

Hurler – MPS I



Inferior beaking of the vertebral bodies



"J" Shaped sella in cone down position

Short Hands & Feet

- ❖ **Peripheral Dysostosis**
- ❖ **Hypoparathyroidism**
- ❖ **Pseudo Hypoparathyroidism**
- ❖ **Pseudo Pseudo Hypoparathyroidism**
- ❖ **Fair Bank's Epiphyseal Dysplasia**
- ❖ **Diastrophic Dwarfism**
- ❖ **Ellis-Van Creveld Chondro Ectodermal Dysplasia**
- ❖ **Noonan's Syndrome**
- ❖ **Prader-Willi Syndrome**
- ❖ **Achondroplasia**
- ❖ **Ollier's**
- ❖ **Mucopolysaccharidosis (E.g., Hurler, Hunter, Morquio)**
- ❖ **Hypochondroplasia**

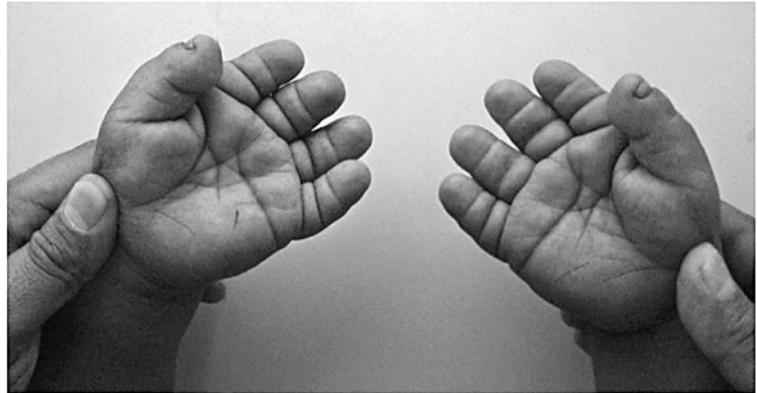
Short Hands and Feet

Trident hand



Short metacarpals and phalanges with wide epiphyses
Differentiation by interpedicular distance in lumbar spine
Gradual narrowing in achondroplasia, normal in pseudo

Achondroplasia - Most common type of dwarfism



Both palms

**Shortening of metacarpals and phalanges,
irregular articular ends**



Achondroplasia
Trumpet appearance of the
metaphyses



Pseudoachondroplasia
includes changes in the
epiphyses

Peripheral dysostosis

Singleton et al. (1960) reported a form of dysostosis limited essentially to the tubular bones of the hands and feet. The epiphyses in the fingers are conical with their apex set into the metaphyseal ends of the phalanges.



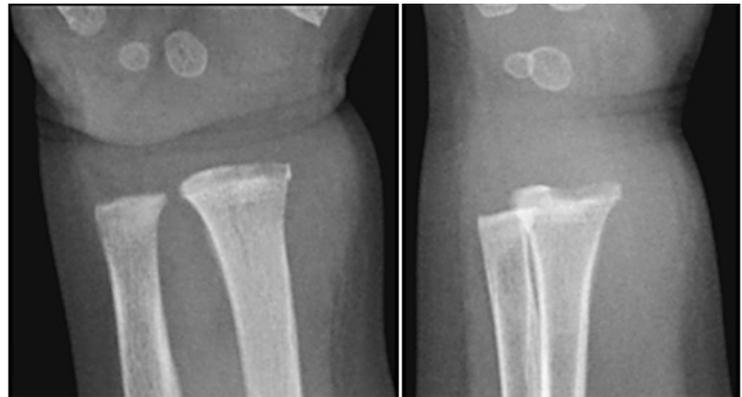
11 yr old short girl – Peripheral dysostosis

Section-4
Infections

Bacterial



17 M, Osteomyelitis of hamate with lysis. Post intravenous injections. Cuboidal bones do not exhibit periosteal reaction.



Early Osteomyelitis

Soft tissue swelling and lytic lesion in the distal end of radius

Staphylococcal infection



**Early osteomyelitis
Distal phalanx**

Post cellulitis



**Carpal osteolysis and osteolysis of
bases of metacarpals,
distal ends of radius and ulna**

Tuberculosis



Spina Ventosa – lytic lesions with expansion



TB



TB



Spina ventosa



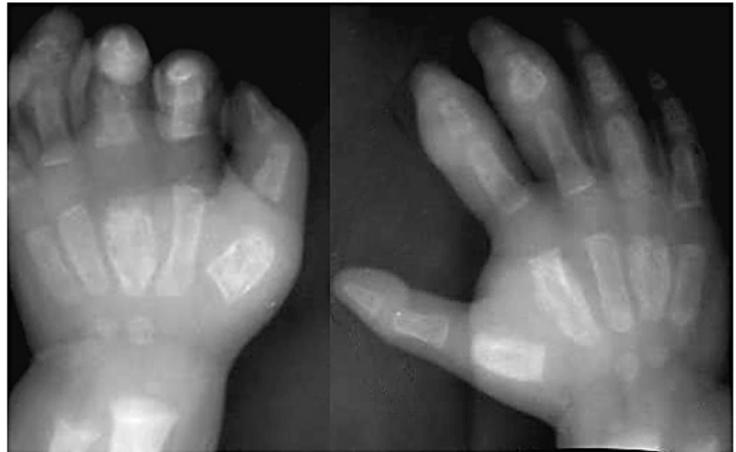
Tuberculosis dactylitis



Tubercular dactylitis



Spina ventosa



Simulates sickle dactylitis



TB middle phalanx



MRI



TB dactylitis



TB 5th metacarpal



Multicentric cystic Tuberculosis simulating enchondromata

(Torulosis)



CRYPTOCOCCUS
NEOFORMANS

Expanding lytic lesions with no periosteal reaction. Simulates spina ventosa of TB

Actinomycosis



Bitten by girlfriend

Diffuse soft tissue swelling, sclerosis and periosteal reaction of the metacarpals



Fungal infection of the hand – Actinomycetes group. Diffuse soft tissue swelling with osteoporosis of bones

Congenital Syphilis



Osteochondritis, osteitis, periostitis, osteomyelitis. Bilateral, symmetrical

Syphilitic dactylitis



Diffuse sclerosis, periosteal reaction and soft tissue swelling

Maternal Rubella syndrome



**ENCOUNTERED IN INFANTS WITH MATERNAL RUBELLA
RADIOLOGICALLY METAPHYSES ARE INVOLVED DENSE
METAPHYSES WITH CELERY STALK APPEARANCE.
NO PERIOSTEAL REACTION**



Acroosteolysis

Leprosy

The most common specific bone changes are primary periosteitis and the areas of bone destruction. Nonspecific bone changes include absorptive changes. The absorption of the terminal phalanges acroosteolysis is present.

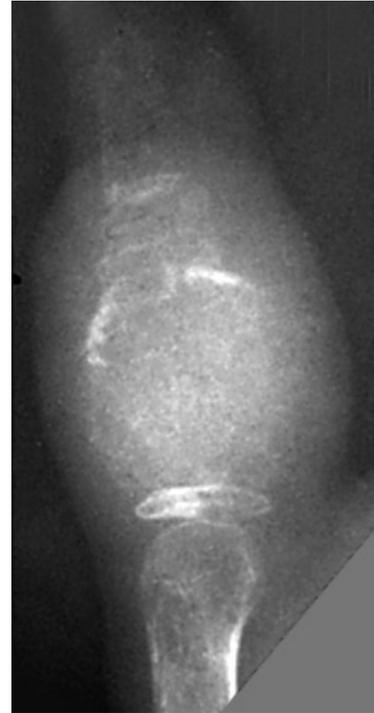


Leprosy

Osteoporosis and small cystic changes in the phalanges. Findings often simulate sarcoidosis.



16 F, Sarcoidosis
Multiple lytic areas, some with honey combing



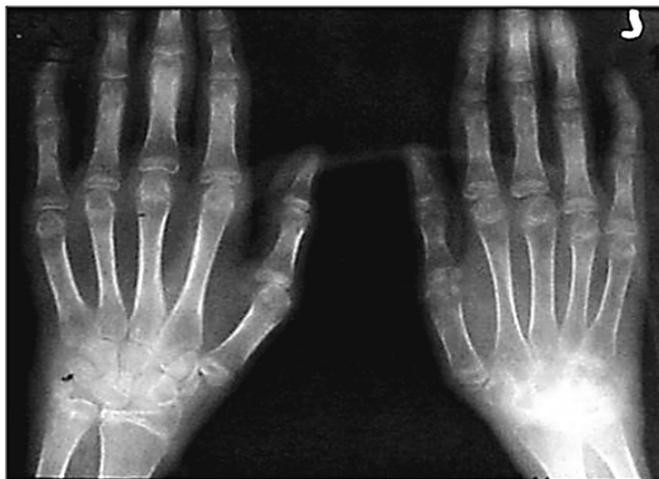
Giant cell reparative granuloma. Radiological appearances similar to giant cell tumor except the epiphysis is not involved

Collagen disorders



- ❖ Scleroderma
- ❖ Lupus erythematosus
- ❖ Dermatomyositis
- ❖ Rheumatoid
- ❖ Poly arteritis nodosa

SCLERODERMA – Soft tissue calcifications



Juvenile idiopathic arthritis

Section-5
ARTHRITIDES

**Juvenile idiopathic arthritis formerly known as
juvenile rheumatoid arthritis**

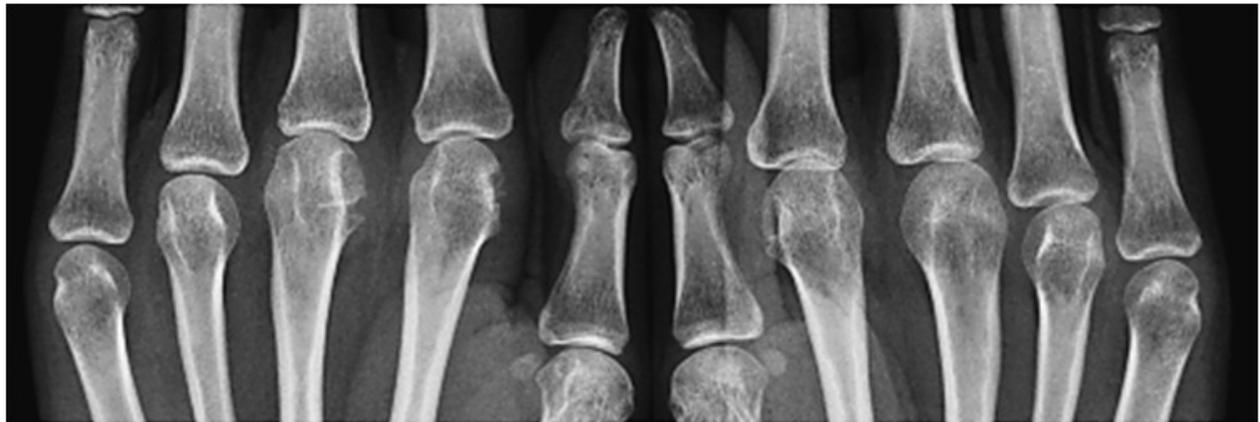


Active inflammation of isolated joints
No specific diagnostic findings



Paraarticular osteoporosis.
Erosions articular margins.
Enlarged phalanges

17 F, Juvenile Idiopathic Arthritis

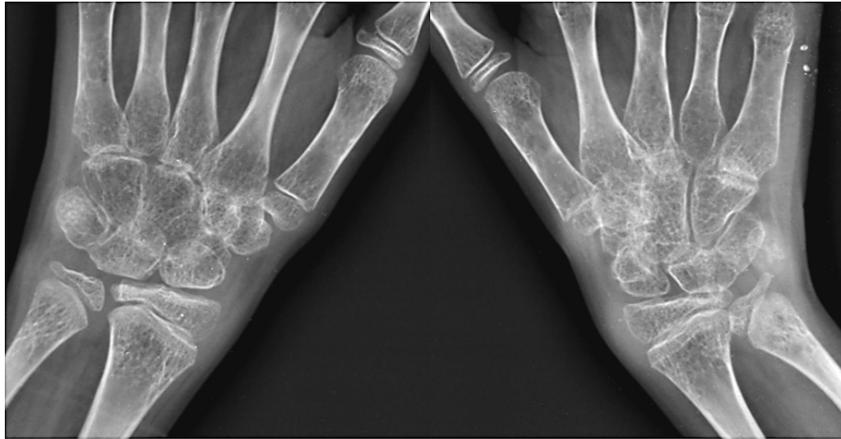


**Para articular osteoporosis, erosions of
carpal bones & metacarpal heads**

Juvenile idiopathic arthritis (JIA) is a heterogeneous group of diseases characterized by synovial inflammation. JIA has been further classified on the basis of the number of joints involved, additional symptoms, family history, and serologic findings. Imaging in patients with JIA has historically relied on radiography, which allows the accurate assessment of chronic changes of JIA, including growth disturbances, periostitis, and joint malalignment. However, radiographic findings of active inflammation are nonspecific.

- ❖ **There are several subtypes of JIA**
- ❖ **oligoarticular JIA**
 - affected ≤ 4 joints in the first six months of illness
 - peak age: 1-6 years
 - mainly affects medium and large joints
- ❖ **polyarticular JIA**
 - ≥ 5 joints are affected
 - peak age: 1-4 years; 7-10 years
 - mainly affects small and medium joints
- ❖ **systemic onset JIA** (a.k.a. Still disease)
 - see above
 - arthritis may present weeks to months after the onset of systemic symptoms

Juvenile Rheumatoid Arthritis (JIA)



12 F, JRA



16 M, JRA

Psoriatic arthritis



Psoriatic arthritis is an inflammatory arthritis associated with psoriasis. It is rare in children. It is usually negative for rheumatoid factor, and hence classified as one of the seronegative spondyloarthritides. Radiologically, no para articular osteoporosis is noted. Sausage shaped finger, reactive new bone formation with periosteal reaction are the main features.

Juvenile dermatomyositis



Note the soft tissue calcification

5 yr old, Dermatomyositis



Contractures and soft tissue atrophy



8 yr old

Jacoud arthropathy



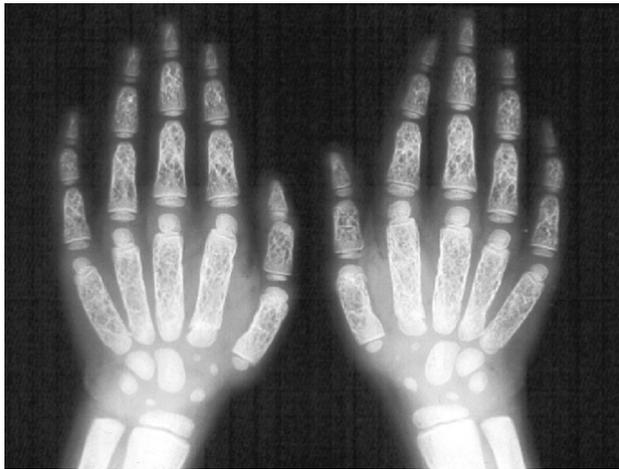
Sequelae of rheumatic fever with no erosions but subluxations mainly at the metacarpo phalangeal joints. To be distinguished from lupus arthritis.

Phalangeal Tufts Preferentially Affected

- ❖ **Scleroderma:** Soft tissue calcifications
- ❖ **Sarcoid:** Acroosteolysis can occur in 50% of patients with sarcoid of the hands. Look for small, cortical, punched-out lesions OR a permeative, lace-like pattern.
- ❖ **Psoriasis:** Erosive disease at the distal interphalangeal joints
- ❖ **Neuropathy**
 - **Leprosy:** Linear calcification of digital nerves
- ❖ **Hyperparathyroidism:** Subperiosteal resorption
- ❖ **Thermal injury**
 - **Burns:** Contractures or soft tissue calcifications.
 - **Frostbite:** Usually spares the thumb.

Section-6
Hematological

Hemoglobinopathies



THALASSEMIA

Cooley's Anemia

- ❖ **Hypermedullation**
- ❖ **Bulging cortices of metacarpals and phalanges**
- ❖ **Mosaic pattern of spongiosa**

Sickle cell hemoglobinopathy



Sickle cell dactylitis
Lytic areas with periosteal reaction



Sickle cell dactylitis
In a 1 ½ yr old child

Sickle cell anemia



Short 3rd metacarpal and proximal phalanx of the index finger due to premature fusion of epiphysis in sickle cell anemia. Ischemia due to thrombosis of nutrient vessels.

Coned epiphyses.

Leukemia



Moth eaten appearance
Osteopenia



Note osteopenia and
crumbling of metaphyseal
ends of radius and ulna



Osteopenia and
metaphyseal
changes of the
metacarpals

**9 M, Leukemia -
Chronic ALL**



Mild bone marrow changes

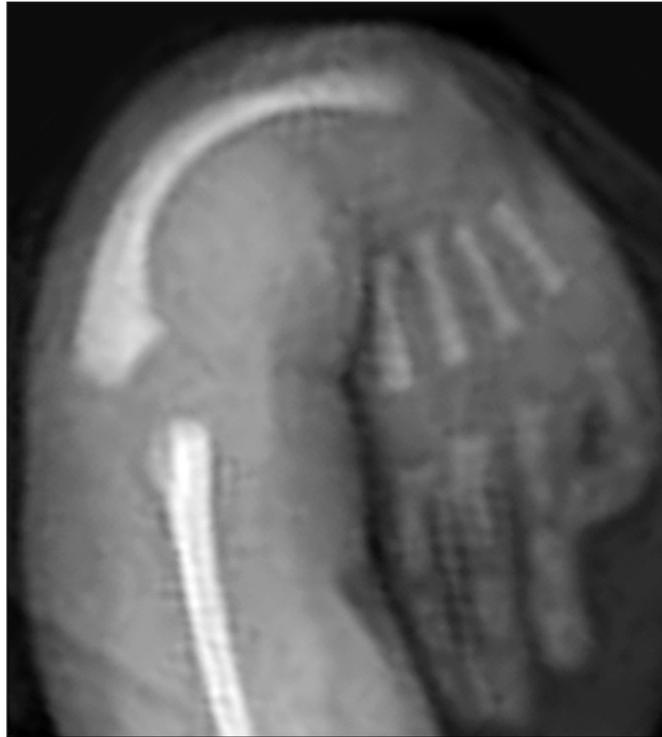
**2 yr, Acute lymphatic
Leukemia**



Osteopenia with bands of
sclerosis at the metaphyses

Radial ray anomalies – 4 types

Bayne and Klug classification



Fanconi anemia

-
- Type 1** Deficient distal radial epiphysis
 - Type 2** Deficient distal and proximal radial epiphyses
 - Type 3** Present proximally (partial aplasia)
 - Type 4** Completely absent (total aplasia – most common)

Enzymatic

Hypophosphatasia deficiency of alkaline phosphatase or phosphoethanolaminuria



- ❖ Neonatal – TAR baby
- ❖ Infantile
- ❖ Juvenile
- ❖ Adult

Metaphyseal flaring, irregular periphyseal regions with large 'chewed out' metaphyseal lucencies



Both hands

Hypophosphatasia - AR



New born



New born



2 yrs

Hypophosphatasia (HPP) is a rare genetic disorder characterized by the abnormal development of bones and teeth. These abnormalities occur due to defective mineralization, the process by which bones and teeth take up minerals such as calcium and phosphorus. Radiologically, it resembles rickets in children.

Hyperphosphatasia

Hereditary **hyperphosphatasia** is a rare genetic bone disorder (osteopathy) that becomes apparent during infancy or early childhood. It is also called Juvenile Paget's disease as the radiographic appearances simulate Paget. Increased density with widening of the bones is noted.



Autosomal recessive-
Genetic-TNFRSE11B
mutations 8

Homocystinuria



It is a disorder of methionine metabolism, leading to an abnormal accumulation of homocysteine and its metabolites (homocystine, homocysteine-cysteine complex, and others) in blood and urine. It is **due to Cystathionine Beta-Synthase Deficiency**. Radiologically, osteoporosis is the dominant feature. In the carpal bones, the capitate is larger than usual.

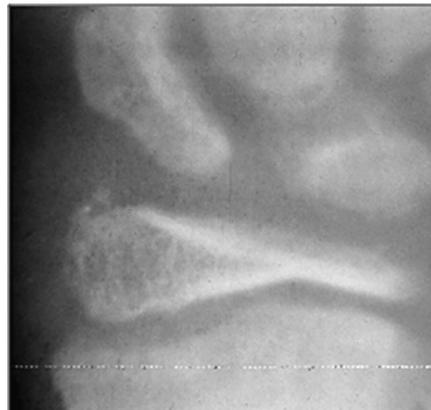
Wilson disease

Serum ceruloplasmin low. Copper accumulates in liver, kidney and cartilages

- ❖ RICKETS
- ❖ OSTEOPOROSIS
- ❖ 'FRINGED' ARTICULAR MARGINS
- ❖ FRAGMENTATION
- ❖ OSSICLE FORMATION



Radiologically, the bones show rachitic changes with cupping of the metaphysis and increased distance between epiphyses and metaphyses. Crenated articular margins is a major feature.

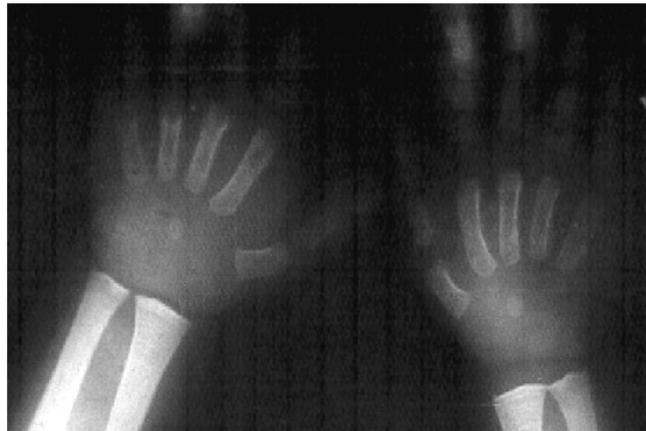


Fringing of articular margins + Rickets

Section-8
Endocrinal



Hand radiograph of a 9 ½ years old girl. The bone age is 8 years - Hypothyroidism

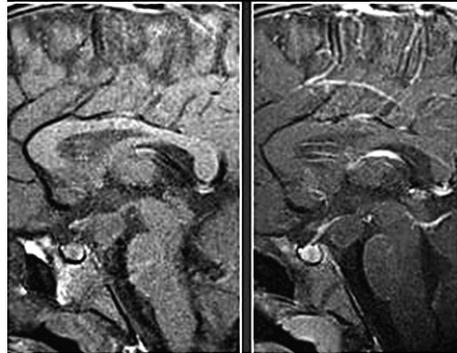


HYPOTHYROIDISM (CRETIN)
Delayed ossification centres – 2yr. old

Advanced bone age



Hypothalamic Hamartoma

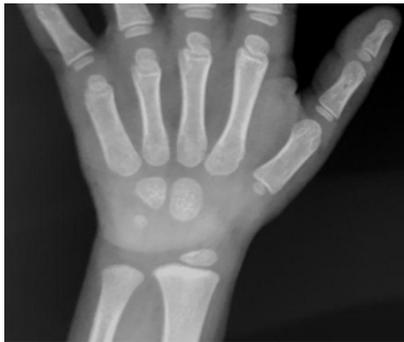


3 yr with precocious puberty, (8 carpal centers)

Hyperthyroidism - 6 yr



8 carpal centers



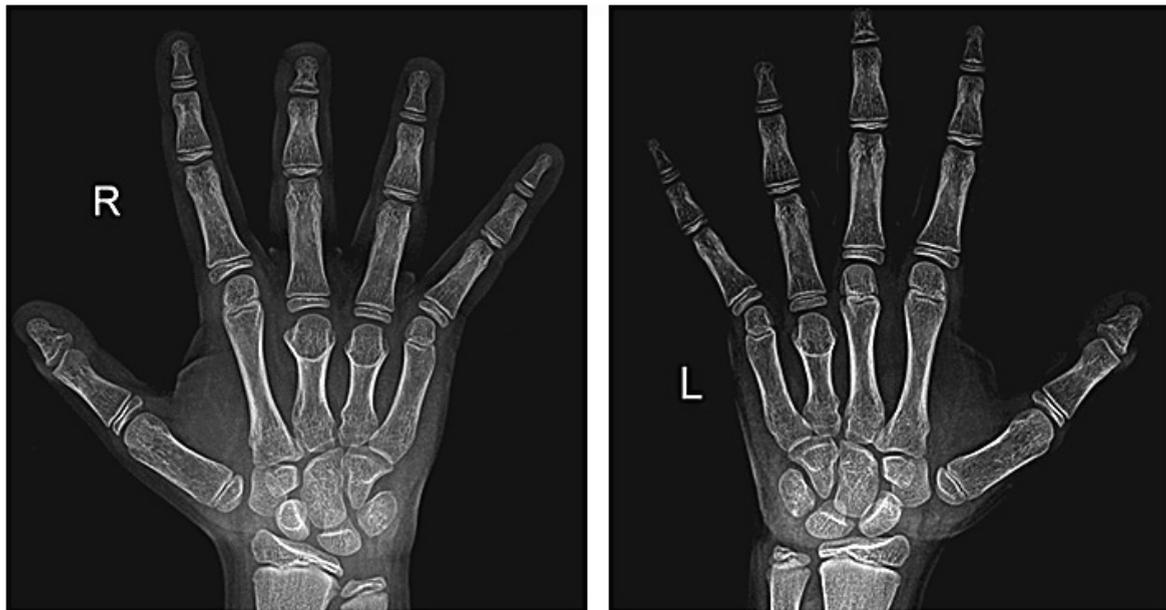
CINCA syndrome – Infant with 3 carpal centres
(Chronic Infantile Neurological
Cutaneous and Articular)



Cushing syndrome –
osteoporosis

Idiopathic Hypoparathyroidism

Hypoparathyroidism is an endocrinal disorder caused by deficiency of parathyroid hormone which in turn results in decreased serum calcium levels and raised serum phosphorus levels



Short metacarpals

Pseudohypoparathyroidism



Short 4th and 5th metacarpals



Basal ganglia calcifications



ARCHIBALD SIGN – ALBRIGHT'S
(short & dimpled 4/5th digits, i.e.,
'knuckle-knuckle-dimple-dimple' sign)
Positive metacarpal sign - TURNER

Pseudohypoparathyroidism is associated primarily with resistance to the parathyroid hormone. Those with this disorder have a low serum calcium and high phosphate, but the parathyroid hormone level (PTH) is appropriately high (due to the low level of calcium in the blood).

Pseudo-pseudo Hypoparathyroidism

Pseudopseudohypoparathyroidism is when the person Looks like someone with the former (“knuckle-kunckle-dimple-dimple” hand being a notable feature) but their blood work is that of a totally normal person.



Radiological findings include short metacarpals similar to other forms of hypoparathyroidism. These three types are genetical variants.

Causes of short fourth metacarpal

Common

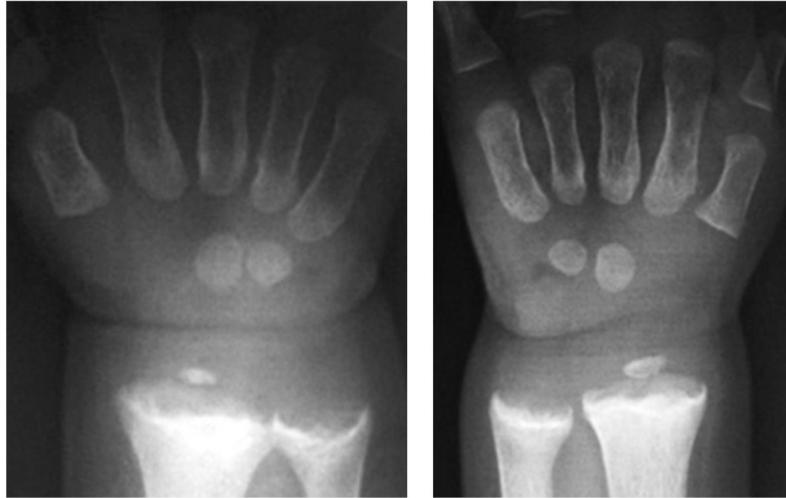
- ❖ Idiopathic, familial
- ❖ Post-infective
- ❖ Pseudohypoparathyroidism
/Pseudo-pseudo
Hypoparathyroidism
- ❖ Turner syndrome
- ❖ Post traumatic

Rare

- ❖ Gorlin syndrome
- ❖ Hereditary multiple
exostosis
- ❖ Sickle cell disease
- ❖ Homocystinuria
- ❖ Post variolar osteo
articular lesions

Nutritional, Metabolic & Toxic

Nutritional rickets



Three types of rickets related to vitamin D:

- ❖ **Vitamin D Deficient**
- ❖ **Vitamin D Dependant**
- ❖ **Vitamin D Resistant – X linked hypophosphatemia**

- ❖ **Delayed growth and maturation**
- ❖ **Widening of growth plate due to osteoid excess Metaphyseal cupping**
- ❖ **Cortical spurs projecting at right angles to metaphysis**
- ❖ **Coarse trabeculation / accentuation of primary trabeculae**
- ❖ **“Paint Brush” metaphyses**
- ❖ **Osteoid seams**

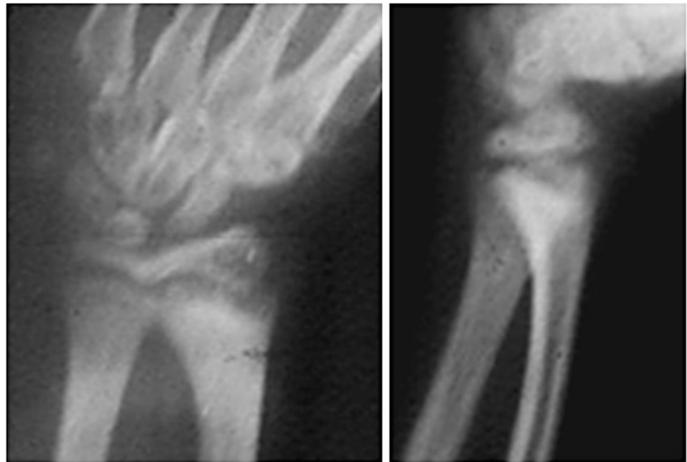
Some scientists consider vitamin D as a hormone.



17 years, Low vitamin D level

Some antiepileptic drugs interfere with hepatic enzymes resulting in rachitic changes.

Vitamin D - Activation of vitamin D is controlled by the PTH hormone (parathyroid). Any problem with this activation process of vitamin D will lead to deficiency due to poor diet, lack of sun exposure, GI, liver or kidney disease.



Dilantin therapy for epilepsy resulting in rickets

Rickets

- ❖ Delay in maturation
- ❖ Osteopenia
- ❖ Cupping of metaphyses



Florid Rickets



15 F,
Vitamin D < 10



Healing Rickets

Non nutritional rickets



Renal rickets with secondary hyperparathyroidism



Non ossifying fibroma with rickets (Humoral)



Renal Rickets



Renal Rickets +
Brown tumor



Fluorosis with rickets

X linked familial hypophosphatemic rickets



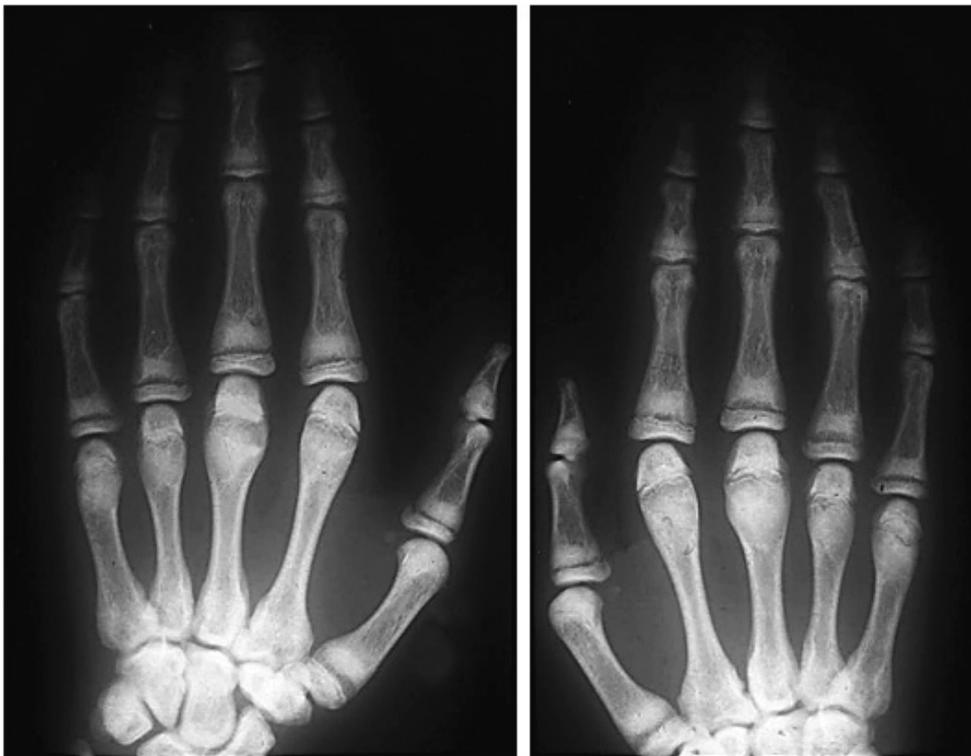
Ground glass appearance of bones with cupping of radius ulna, distally

Healed Rickets



Treated with high doses of Vitamin D

Oxalosis is a rare metabolic disorder that occurs when the kidneys stop eliminating calcium oxalate crystals from the body through the urine. Due to the renal failure, the oxalate crystals are deposited elsewhere in the body, mainly in growing bones. Kidney failure leads to renal osteodystrophy. Besides rachitic changes, the bones are dense and the epiphysis are denser.



Primary oxalosis with renal osteodystrophy



Oxalosis + tumoral calcinosis
Due to renal osteodystrophy

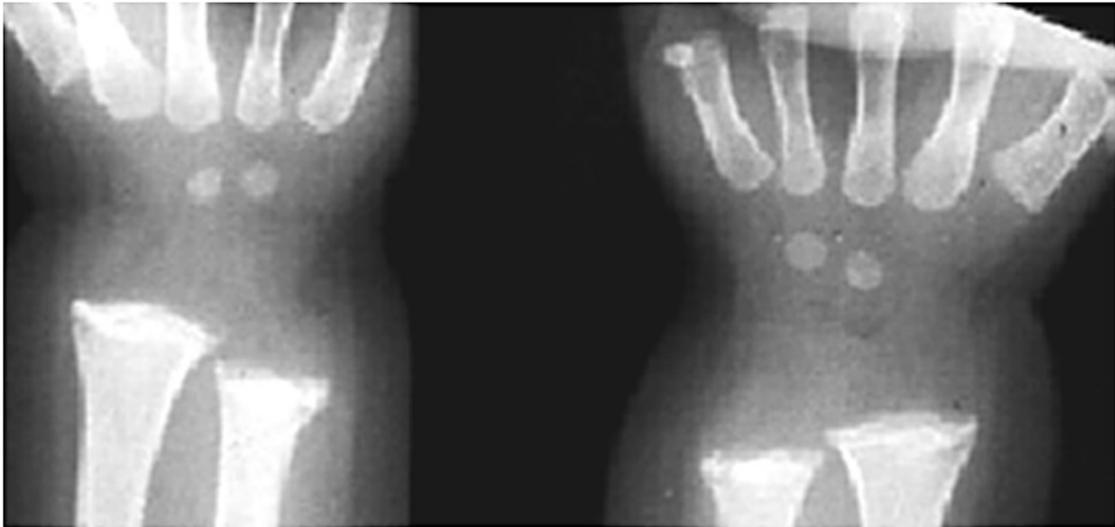


Coarse rarefaction
and cystic changes in
metaphyses, sclerotic
epiphyses with renal
osteodystrophy.

Scurvy – vitamin C deficiency

Scorbutic changes

Osteoporosis, Pelkin's spurs, dense line with a proximal radiolucent lines in metaphyses



Toxic

Lead poisoning (Plumbism)

Dense bands at the growing ends of the bones and around carpal ossification centers



Lead poisoning
(Plumbism)



Lead toxicity



Dense bones with
rachitic changes

Skeletal Fluorosis

Complex Regional Pain Syndrome - CRPS



Immobilisation for fracture radius resulting in CRPS



Para articular osteoporosis

Hyper vitaminosis A & D



Osteoporosis of the bones, dense metaphyseal bands and periosteal reaction along ulna

Frost bite



Erosive with reactive sclerosis at the bases of the middle phalanges

Epidermolysis bullosa



Marked contractures with tapering of distal phalanges – different from burns

Epidermolysis bullosa:

Hereditary disorder of marked squamous epithelial fragility and blister formation, affecting the skin and mucous membranes. Patients have numerous bullous lesions, which scar after the rupturing, leading to joint contractures, and esophageal strictures.



Skin over the shoulder

Section-10

PHACOMATOSES

The neurocutaneous syndromes are a group of inherited disorders featuring multiple, discrete lesions of 2 or more organ systems, most commonly the skin and brain.



Mesenchymal dysplasia in neurofibromatosis

- ❖ Neurofibromatosis 1 & 2
- ❖ Tuberous sclerosis
- ❖ Macrodystrophia lipomatosa
- ❖ Klippel Trenaunay Weber syndrome
- ❖ Proteus syndrome

Neurofibromatosis 1 & 2

Note the bizarre shaped bones with developmental deformities of the phalanges, metacarpals and carpals. Cystic changes in the phalanges should not be mistaken for tuberous sclerosis.

Note also growth abnormalities

NF1



Mesenchymal dysplasia
2nd & 3rd fingers

NF1



Mesenchymal defect
in 3rd metacarpal

Epiloa / Tuberos Sclerosis - AD



Little finger



**Note cortical pitting, sclerosis and cystic change in phalanges.
Clinically, adenoma sebaceum, seizures and mental retardation.**

Macrodystrophia lipomatosa



This may be part of generalised hamartomatous disorder. Anatomic location corresponds closely with the innervation of sclerotomes.

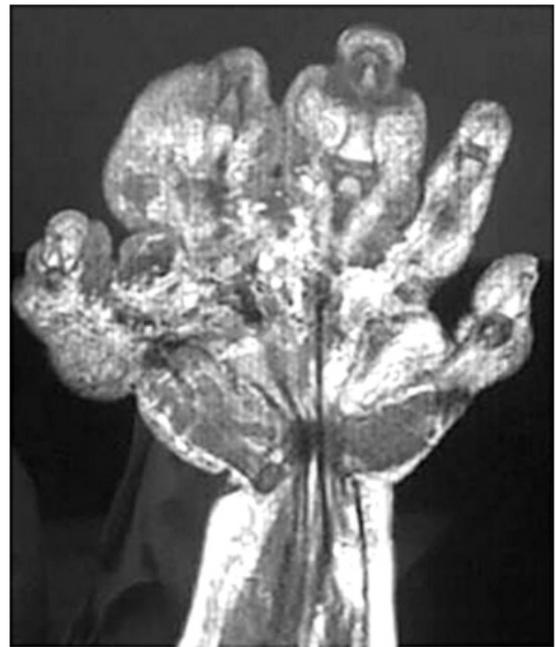
Differential diagnosis

- ❖ neurofibromatosis type 1 (NF1)
- ❖ fibrolipomatous hamartoma of the median or ulnar nerve
 - fatty tissue accumulates within the nerve sheath rather than within the region
- ❖ vascular malformation
 - haemangioma, including Maffucci syndrome
 - arteriovenous malformation (AVM)
 - Klippel-Trenaunay-Weber syndrome

Macrodystrophia lipomatosa



Involvement of thumb and index finger



MRI – medial 3 fingers are involved

Klippel Trenaunay Weber syndrome



Represents low-flow vascular malformation.



Generally involves the lower limb. Most common major finding is overgrowth of subcutaneous soft tissues. The effected fingers are thicker and longer than others. Multiple phleboliths are seen in venous channels.

Clinical



Asymmetric macrodactyly with hyperostosis

Proteus syndrome (1979) is a rare disorder named after the name of Greek God Proteus who could change his shape at will to avoid capture. It is characterized by overgrowth of the bones, skin, and other tissues. Organs and tissues affected by the disease grow out of proportion to the rest of the body. The overgrowth is usually asymmetric, which means it affects the right and left sides of the body differently. Congenital disorder in which local gigantism is associated with progressive overgrowth of all the mesenchymal elements. The involvement is almost always unilateral.

Myofibromatosis / Infantile myofibromatosis / Aggressive infantile fibromatosis



Almost 90% of cases occur within the first 2 years of life, with 50% occurring in newborns. Three categories of IM are described: solitary, multicentric without visceral involvement, and generalized with both cutaneous and visceral involvement. The multicentric form of IM involves the skin, subcutaneous tissues, muscles, and bone. The bones most commonly involved are the skull, femur, tibia, spine, and ribs. Involvement of the central nervous system has also been reported.

Section-II

Benign Neoplasms

Osteoid osteoma
phalanx with central
calcific focus



A benign osseous tumor which may be periosteal, intracortical, medullary and sub articular. A lucent nidus with surrounding sclerosis is often noted. When the nidus is not seen on conventional films, CT scan is advised. The diameter of the nidus is < 2cm.



Osteoblastoma of the hamate bone

A benign osseous tumor with a nidus > 2cm in diameter. Radiologically, four morphological patterns are noted.

1. Giant osteoid osteoma,
2. Aneurysmal bone cyst,
3. Fibrous dysplasia,
4. Osteosarcoma.

Cartilaginous tumors

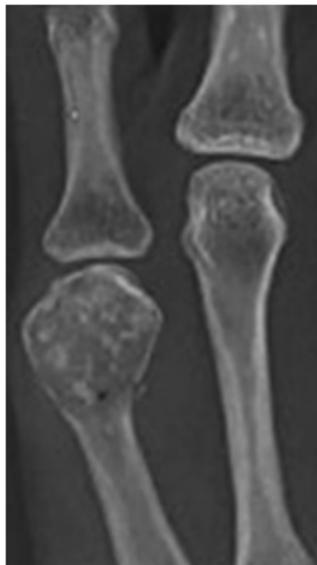


Ollier's – Multiple enchondromata

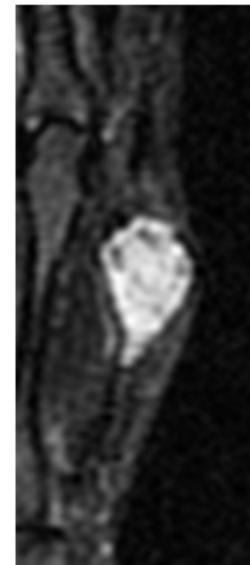


Enchondroma of fifth metacarpal head

Note multiple calcifications with pathological fracture



18 yr old, enchondroma 2nd metacarpal head



MRI

Enchondroma Protruberans 4th metacarpal



MRI

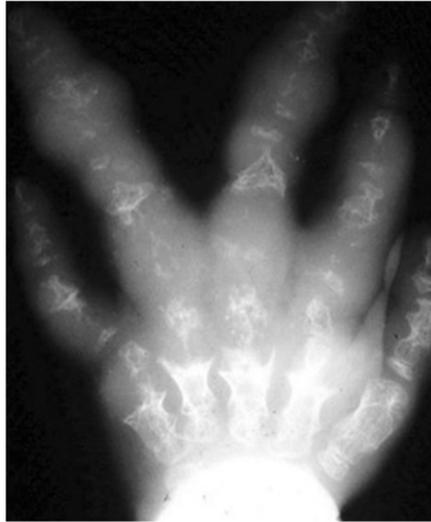
Multiple enchondromata



Benign cartilaginous lesions are more common in children:

1. Enchondroma
2. Multiple enchondromatosis
3. Osteochondroma
4. Multiple exostosis
5. Ollier's dyschondroplasia
6. Periosteal chondroma
7. Chondroblastoma
8. Chondro myxoid fibroma
9. Trevor disease
(Dysplasia epiphysealis hemimelica)
10. Intra cortical chondroma

Ollier's dyschondroplasia



Multiple enchondromata, cartilaginous dysplasia with linear columns of hyaline cartilage. Shortening of the bones may be seen.



Periosteal chondroma

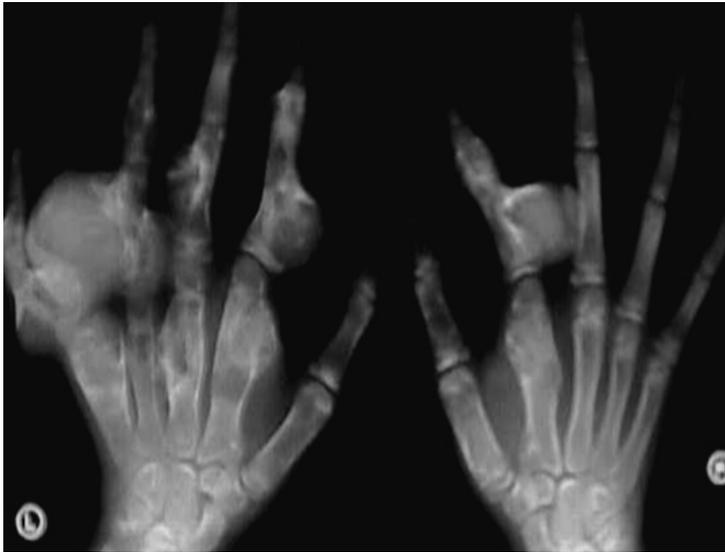


Little fingers

2nd metacarpal

It is also called ‘Juxta cortical chondroma’ and is a chondromatous lesion arising from the periosteum. It may grow into the medullary cavity. A characteristic cortical hook, similar to the hook noted in gout indicates the cartilaginous nature. MRI is rarely necessary. Nodular calcification may be noted in the matrix. Note the out-growths in the proximal phalanges with hook shaped deformity in the diaphyses. Chondroid calcification may be present.

Muffucci syndrome



Multiple chondromata with soft tissue hemangioma

Note Phleboliths

Multiple Hereditary Osteochondromata



Note multiple, scattered calcific densities with deformities at the wrist. Note pseudo Madelung deformity



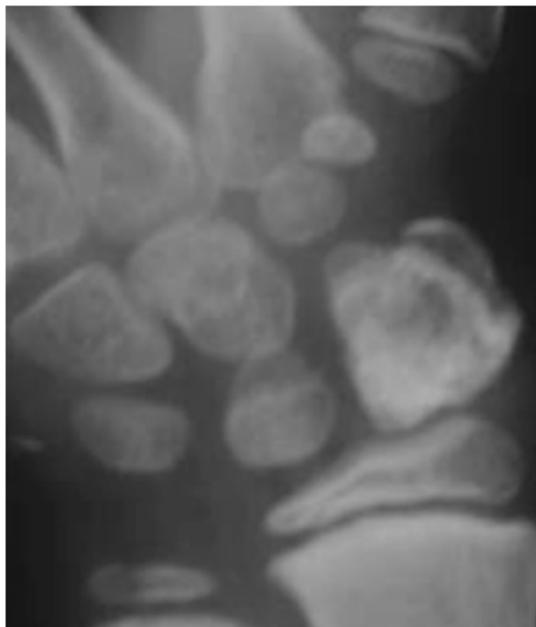
Bilateral, almost symmetrical



**Multiple hereditary osteochondromata with
Pseudo Madelung deformities**

Trevor disease

Dysplasia Epiphysealis Hemimelica (DEH)



A rare cartilaginous lesion arising from the epiphysis or ossification centre. It may involve the adjacent bone and generally has an unilateral distribution.

Vascular lesions of bone

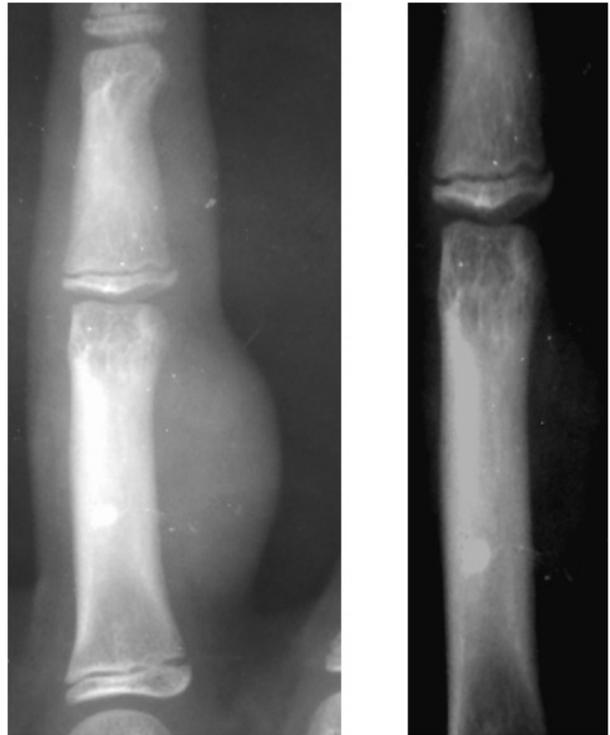
BENIGN:

- Hemangioma

- ❖ Cavernous
- ❖ Capillary
- ❖ Venous
- ❖ Sclerosing

To be differentiated from
arteriovenous and venous
malformations

Sclerosing hemangioma



Soft tissue swelling, phlebolith and
sclerosis of the bone

Hemangioma

Cavernous
Hemangioma
of middle finger
simulating
spina ventosa

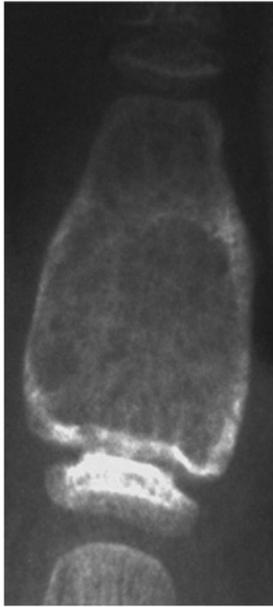


Hemangiomata



Lymphangioma

Primary aneurysmal bone cyst

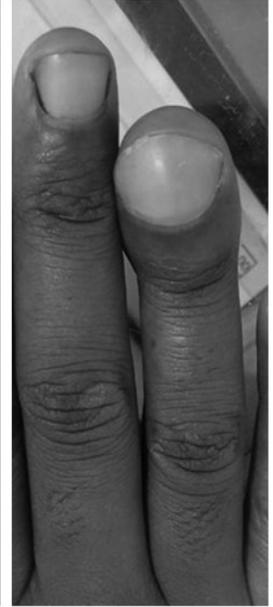


Proximal Phalanx



5th metacarpal

Glomus tumor



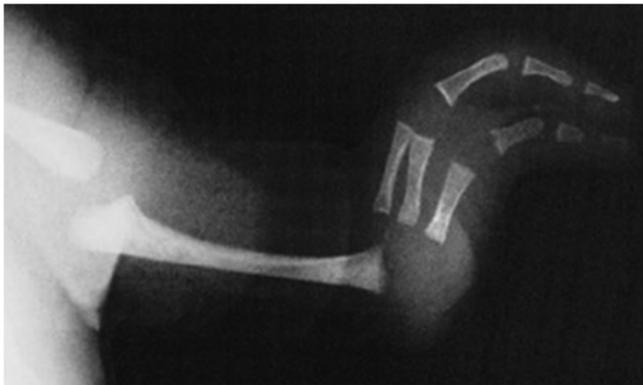
Expansion & sclerosis, clubbing

An expanding osteolytic lesion with thinning of the cortex with a peripheral shell. The zone of transition is narrow but can be wide and aggressive. Should not be mistaken for enchondroma or giant cell tumor. Secondary aneurysmal bone cysts can occur in giant cell tumors, non ossifying fibroma, chondroblastoma, osteoblastoma, telangiectatic osteosarcoma. Fluid – Fluid levels are seen on MRI.

Section-12 Syndromes

Holt Oram syndrome

Syndrome is a set of medical science and symptoms that are correlated with each other and often with a particular disorder. There are several forme fruste examples.



Cardiac limb syndrome

- ❖ Thumb anomalies – finger like
- ❖ Absent metacarpals
- ❖ 5th digit – short middle phalanx
- ❖ Hypoplasia of radial carpals
- ❖ Radial ray anomalies
- ❖ Shoulder and clavicle may also be involved

ASD / VSD

Fanconi Anaemia

Stunted growth and
loss of appetite
3rd degree
consanguinity

Radial ray deficiency



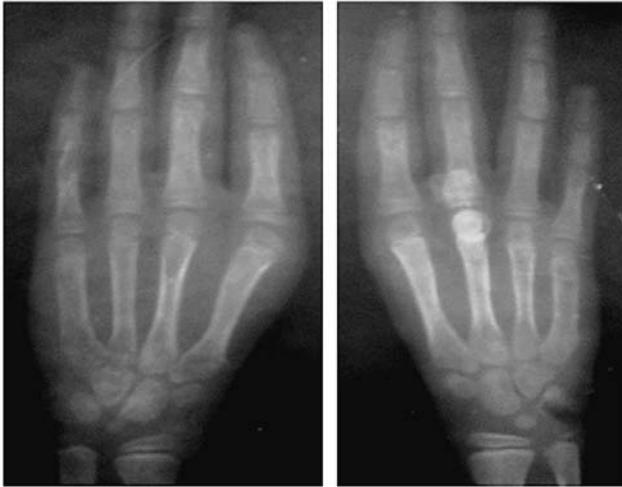
Type 5



Type 1

- ❖ FANCONI ANAEMIA
- ❖ HOLT ORAM
- ❖ VACTERL
- ❖ TAR

Hypocellular marrow with erythroid prominence and suppressed myelomegakaryopoiesis. Negative for granulomas or infiltrates. Features are suggestive of evolving hypocellular marrow. Kindly correlate clinically and with the cytogenetics



Fanconi's anaemia, Hb – 5 g%



Absence of radial ray



Fanconi anemia
– aplasia of first
metacarpal and thumb

Type I - the thumb is small, normal components are present but undersized. Two muscles of the thumb, the abductor pollicis brevis and opponens pollicis, are not fully developed ,.

Type II - is characterized by a tight web space between the thumb and index finger which restricts movement, poor thenar muscles and an unstable middle joint of the thumb metacarpophalangeal joint.

Type III - thumbs are subclassified into two subtypes by Manske. Both involve a less developed first metacarpal and a nearly absent thenar musculature. Type III-A has a fairly stable carpometacarpal joint and type III-B does not. The function of the thumb is poor. Children with type III are the most difficult patients to treat because there is not one specific treatment for the hypoplastic thumb. The limit between pollicization and reconstruction varies. In this group careful attention should be paid to anomalous tendons coming from the forearm (extrinsic muscles, like an aberrant long thumb flexor – flexor pollicis longus).

Type IV - is called a pouce flottant, floating thumb. This thumb has a neurovascular bundle which connects it to the skin of the hand. There's no evidence of thenar muscles and rarely functioning tendons. It has a few rudimentary bones. Children with type IV are difficult to reconstruct. This type is nearly always treated with an index finger pollicization to improve hand function.

Type V - is no thumb at all and requires pollicization.

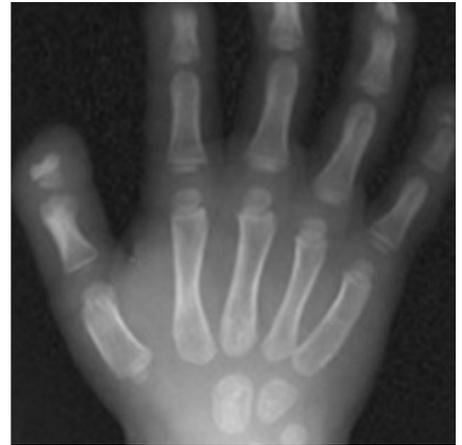
Constriction band syndrome (Streeter's dysplasia)

It is not so uncommon and the etiology refers to constricting amniotic bands

Classification (Patterson)

- a) Simple constrictions (partial/circumferential)
- b) Constrictions with distal deformity
(lymphedema may or may not be present)
- c) Constrictions with acrosyndactyly characteristic
(Fenestrated syndactyly)
- d) Intrauterine amputation

Amniotic band syndrome



Amniotic band constriction (also known as “**amniotic band syndrome**”, “**ADAM complex**”, “**Amniotic band sequence**”, “**Congenital constriction bands**” and “**Pseudoainhum**”) is a congenital disorder caused by entrapment of fetal parts (usually a limb or digits) in fibrous **amniotic bands** while in utero.



Amniotic band syndrome



Amniotic Band Syndrome 2 ½
month old baby



Amniotic band Syndrome

Symbrachydactyly



- ❖ Type 1 – Triphalangeal type
- ❖ Type 2 – Diphalangeal type
- ❖ Type 3 – Monophalangeal type
- ❖ Type 4 – Aphalangeal type
- ❖ Type 5 – Ametacarpia type
- ❖ Type 6 – Acarpia type
- ❖ Type 7 – Forearm amputation type

A congenital abnormality, characterized by limb anomalies consisting of brachydactyly, cutaneous syndactyly and global hypoplasia of the hand or foot. In many cases, bones will be missing from the fingers and some fingers or toes may be missing altogether.

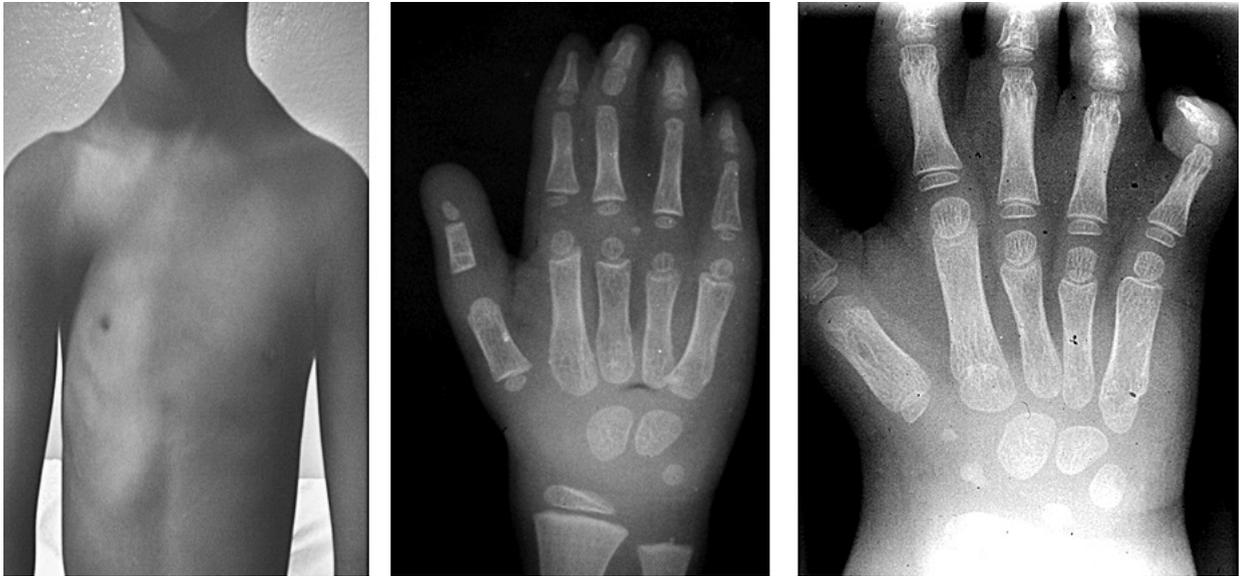
Hand deformities in generalized skeletal disorders

Most common – multiple exostoses

Other syndromes –

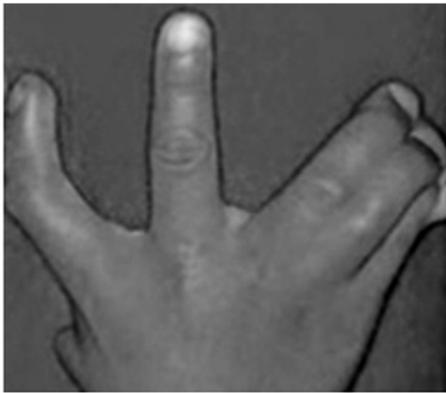
- ❖ **Poland syndrome (symbrachydactyly)**
- ❖ **Apert syndrome (complex syndactyly)**
- ❖ **Haas syndrome**
- ❖ **Freeman sheldon syndrome (wind blown hand)**
- ❖ **Mohr Wriedt syndrome (radial clinodactyly of index finger)**
- ❖ **Pierre – Robin syndrome (clasped thumb)**

Poland Syndrome (1841)



It is a congenital defect characterized by an underdeveloped chest muscle and short webbed fingers on one side of the body. Short ribs, less fat, and breast and nipple abnormalities may also occur on that side.

Haas syndrome



**Presence of 6 metacarpals
is characteristic > 5 digits
All having 3 phalanges**

Tricho rhino phalangeal syndrome (Giedion)



**Rare inherited disorder
Note the coned epiphyses
Clinically, sparse frontal hair, pear
shaped nose and deformities of the
phalanges**

Acrodysostosis is a rare skeletal dysplasia, first described by Maroteaux and Malamut in 1968. Though most case reports have occurred sporadically, it is believed to be an autosomal dominant condition.



The metacarpals and phalanges show typical cone-shaped epiphyses, which fuse prematurely. Cone shaped epiphyses are a rare and distinctive clue to the diagnosis. Acrodysostosis, pseudohypoparathyroidism and acromesomelic dwarfism may present with cone shaped epiphysis. Even though these conditions share some important clinical features, advanced bone age and first ray hyperplasia are two important findings which distinguish acrodysostosis from the other conditions(3). A pointer in neonates with suspected acrodysostosis is the extensive epiphyseal stippling, which almost always disappears by 8 months of age(3).



**Familial
Carpotarsal osteolysis
with nephropathy**



**Pointed bases of
metacarpals, carpal
bones destroyed**



Winchester Grossman syndrome



Carpal and metacarpal osteolysis, phalangeal resorption. A disease that should be classified as a nonlysosomal connective-tissue disturbance and not as a form of acid mucopolysaccharidosis.

Nasu Hakola Syndrome



Cerebral calcification and cystic changes in bones. Polycystic Lipomembranous osteodystrophy with sclerosing leucoencephalopathy. Genes, DAP12 or TREM2.

A Band of Lysis In Middle of Distal Phalanx



Hajdu-Cheney syndrome:

Hajdu-Cheney syndrome: rare disorder of bone metabolism. The diagnosis is made by finding acroosteolysis with any three of the following: Wormian bones, open skull sutures, platybasia, micrognathia, mid-facial flattening, premature loss of teeth, coarse hair and short stature.

Rubinstein Taybi Syndrome



- ❖ Short and wide terminal phalanges of thumb and great toe
- ❖ Short, wide and tufted terminal phalanges of the fingers
- ❖ Flaring of ilia
- ❖ Retarded bone age
- ❖ ASD / VSD
- ❖ Urinary tract anomalies

Arthrogrypsis Multiplex Congenita



Asymmetrical subluxed joints



Companion case

Arthrogrypsis multiplex congenita (AMC) refers to the development of multiple joint contractures affecting two or more areas of the body prior to birth.

Arthrogryposis (Larsen syndrome)

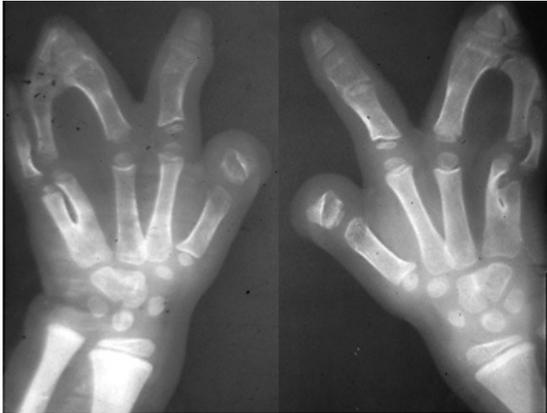
It is a descriptive term comprising disorders due to contractures and dislocation and includes Larsen syndrome

Syndromes with Multiple Carpal Bones



- ❖ Larsen syndrome
- ❖ Brachydactyly A1
- ❖ Ulnar dimelia
- ❖ Hand-foot-uterus syndrome
- ❖ Holt-Oram. syndrome,
- ❖ Oto - palato syndrome,
- ❖ Chondroectodermal dysplasia,
- ❖ Diastrophic dysplasia,
- ❖ Gorlin's syndrome,

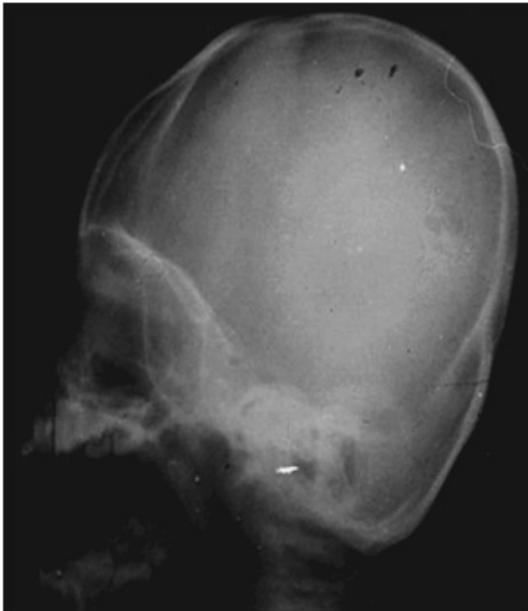
Apert syndrome



Gloved hand



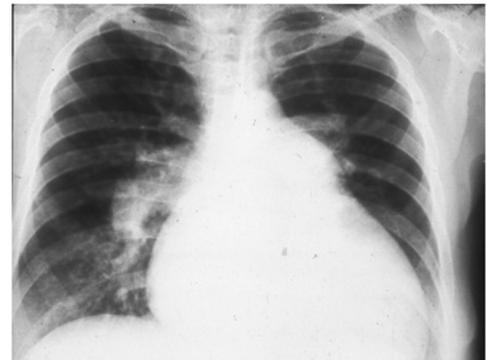
Craniostenosis



A rare congenital disorder characterised by craniostenosis, syndactyly of the hands and feet with tendency for fusion of osseous structures.

Ellis Van Creveld Syndrome

Chondroectodermal dysplasia



ASD

Mesomelic dwarfism with polydactily, syndactily and Septal defect

Post axial Polydactyly, Syndactyly, Syncarpus, deformed teeth &

Scanty hair of the scalp

Chondroectodermal dysplasia/ Ellis –Van Creveld syndrome

Short stature

Short limbs more marked distally

Absent / hypoplastic nails polydactyly, syndactyly

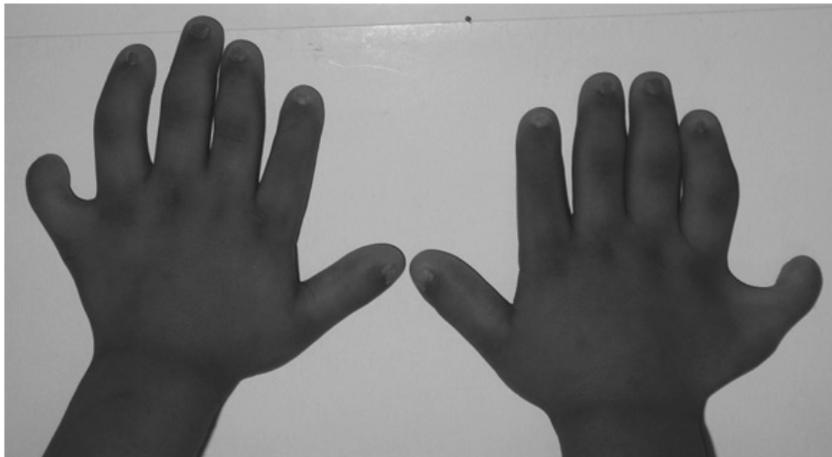
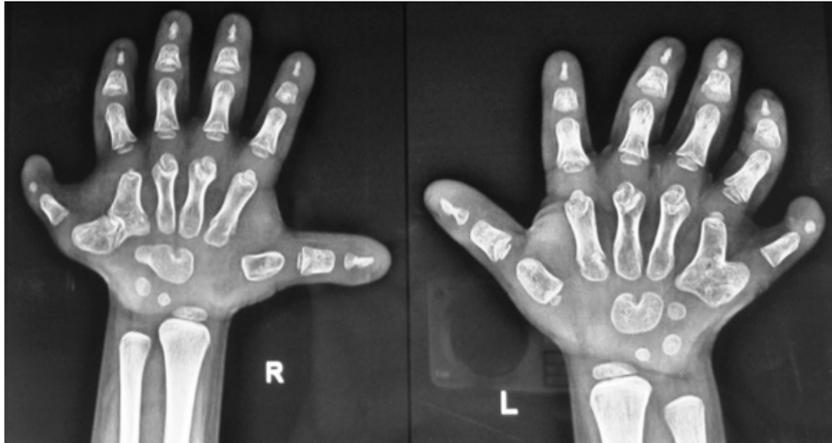
Dysplastic teeth

Upper lip anomalies

Cardiac-ASD, single atrium



5 yr old child with polydactyly, syndactyly



Chondroectodermal dysplasia

Marfan syndrome (AD)



**Arachnodactyly with cardio
vascular defects
Elongated metacarpals and
phalanges, metacarpal
index high**

- ❖ The metacarpal index (MCI) is a radiographic measurement that can be used to confirm the presence of disproportionate metacarpal length.
- ❖ The first method of MCI calculation is by obtaining the average of the ratios of length / width of metacarpals 2-5.
- ❖ The second method involves dividing the sum of the lengths by the sum of the widths of these metacarpals.
- ❖ Values of between 8.4 and 9.4 have been used as cut-off for abnormal values.



Steinberg sign

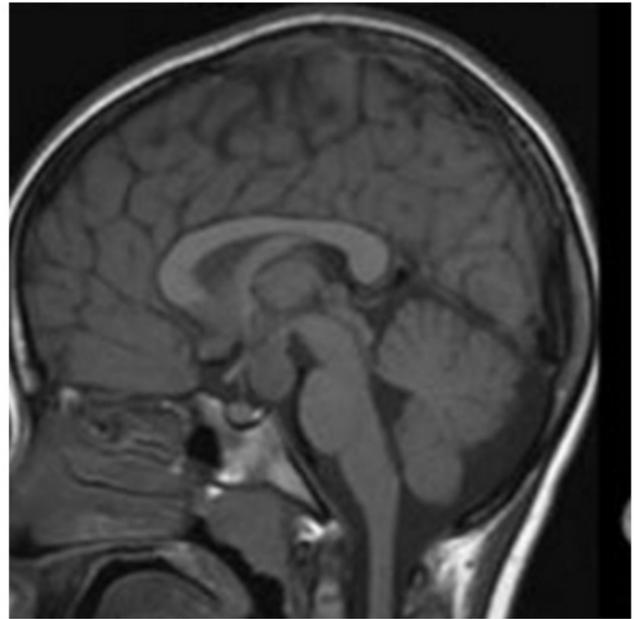


The Steinberg sign, also known as the thumb sign, is one of the clinical examination tests for Marfan syndrome in the hands. It is a clinical test in which the tip of the thumb is visible medial to the little finger when it is clasped in the clenched hand.

SYNDACTYLY WITH SYNDROMES

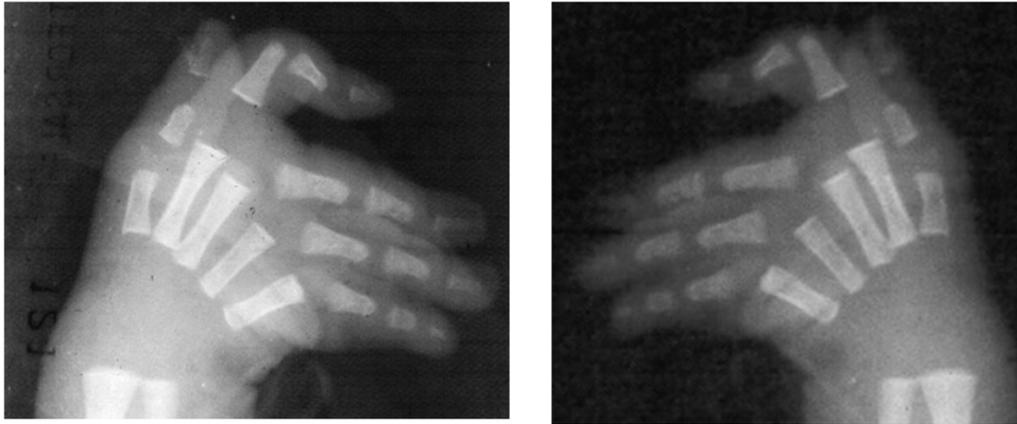
- ❖ Carpenter's
- ❖ Apert's
- ❖ Oculodento digital
- ❖ Conradi
- ❖ Down
- ❖ Neurofibromatosis
- ❖ Pierre – robin
- ❖ Prader willi
- ❖ Pseudo hypoparathy
- ❖ Trisomy – 13
- ❖ Trisomy – 18
- ❖ TAR
- ❖ Russel – silver
- ❖ Ellis Van Creveld
- ❖ Fanconi
- ❖ Goltz
- ❖ Hallerman-Streiff
- ❖ Orodigito facial
- ❖ Popliteal pterygium
- ❖ Otopalato digital

Pallister Hall Syndrome



Hypothalamic hamartomas

Hypothalamic hamartomas with digital malformations (here: short 3rd, 4th and 5th metacarpals , clinodactyly i.e. short mid phalanx of 5th digit and apparent soft tissue syndactyly between 2nd and 3rd digits)



Trisomy 18 (Edwards syndrome)

Hands typically present with overlapping digits, in which the second and fifth fingers override the third and fourth fingers, respectively. V-Shaped deformity between 2nd and 3rd metacarpals. Flexion deformity of the index finger.



Down syndrome Trisomy 21

**Short middle
phalanges of little
fingers, bilaterally.
C1-C2 subluxation**

Camptodactyly-Arthropathy-Coxa Vara-Pericarditis Syndrome



A rare genetic disorder mapped to chromosome 1q25–31. Classically this occurs at the proximal interphalangeal joints specifically of the little finger of the hand, although any finger may be affected. Coxa Vara of hips .

15 M, Camptodactyly-Arthropathy-Coxa Vara-Pericarditis Syndrome



Flexion contraction of the proximal interphalangeal joints, difficulty in walking, sitting down

Mild coxa vara with non specific changes in the hips.



Cornelia De lange syndrome



Short first metacarpal bone, combined with relatively long third and fourth metacarpals, deformed metaphyses of the radius

Turner syndrome

11 yr old girl with webbed neck



Short 4th metacarpal with drumstick fingers

Section-13 Miscellaneous



Hypertrophic osteoarthropathy (HOA) is a syndrome of clubbing of the digits, periostitis of the tubular bones, and arthritis. Pulmonary and non pulmonary causes.

Cyanotic heart disease with periosteopathy



16 yr with tetralogy of Fallot, clubbing of the fingers and periosteal reaction along the metacarpals and proximal phalanges.

Periosteal Reaction in Pediatrics

- ❖ **Physiological (During rapid growth)**
- ❖ **Premature birth (Prostaglandin E)**
- ❖ **Healing trauma**
- ❖ **Infection –**
- ❖ **Metabolic (Scurvy, hypervitaminosis A & D, healing rickets etc.,)**
- ❖ **Hyperphosphatasia**
- ❖ **Caffey disease**
- ❖ **Inflammation - Eosinophilic granuloma, sarcoidosis, JIA, psoriasis**
- ❖ **Infarction**
- ❖ **Skeletal dysplasias - Engelmann**
- ❖ **Neoplastic – Leukemia, malignant bone tumors, neuroblastoma, mets**
- ❖ **Idiopathic – Hypertrophic osteoarthropathy**

Bizarre parosteal osteochondromatous proliferations (BPOP)



4 F, Nora lesion
(Post traumatic)



Middle phalanx of
index finger



Focal periostitis of
unknown etiology

Pigmented Villonodular Synovitis – localised – Giant cell tumor of the tendon sheath

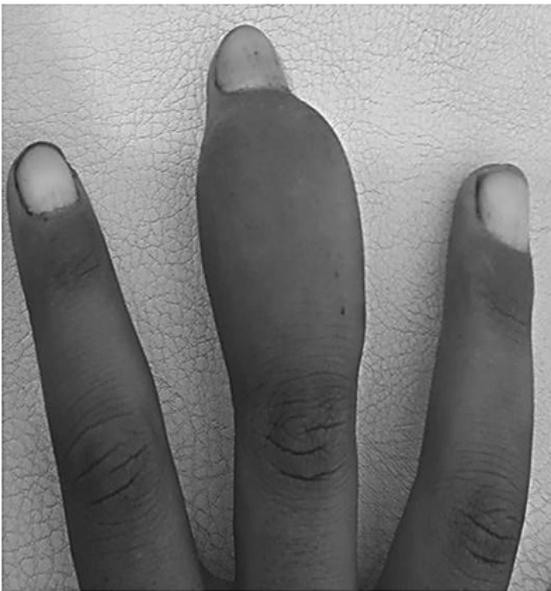


GCTTS may arise from the synovium and pressure erosion can occur on the adjacent phalanx. It is rare in children.

11 yr old with soft tissue swelling antero laterally at the proximal phalanx of the index finger with lytic lesion in the phalanx

8 F, Giant cell tumor of tendon sheath (PVNS) Focal Pigmented Villonodular Synovitis

Soft tissue swelling with minimal involvement
of the cortexExtra articular



Soft tissue venous Hemangioma



Plain



Angiography

Ten Commandments In Skeletal Trauma

- 1. Films / Digital images should be of good quality (poor films invite erroneous diagnosis)**
- 2. Know normal anatomy**
- 3. Beware of normal variants and anomalies**
- 4. Comparison views specially under the age of 18 years**
- 5. Study the soft tissues including fat pads**

Ten Commandments In Skeletal Trauma (Cont.)

6. More views, Special views and stress views
(without discomfort to the patient)
7. Differentiate chip fractures from accessory ossicles and calcifications
8. Study the whole film (in a long Bone Trauma, both distal and proximal joints should be included)
9. Benefit of doubt to the patient
10. Follow up films and further imaging e.g. CT, MRI, US etc.,



View box



Magnifying lens
(Antique)



Modern – zoom the spot

17 yr old fall on out stretched hand



Missed



10 weeks later

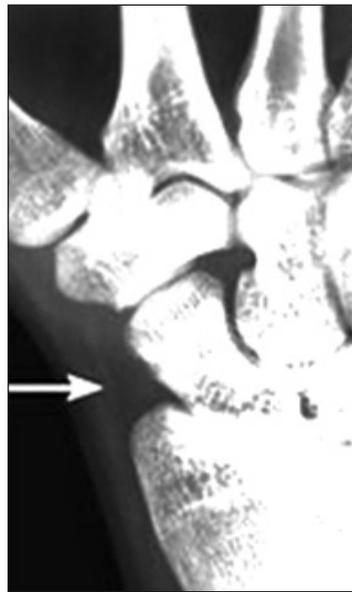
Follow-up films/images in the same position with same technique are important

Wrist is the most commonly injured part in the body. The complex shape and articulations of the carpus make it possible to miss the trauma. Carpal bone fractures account for 18% of hand fractures.

Scaphoid fat pad sign



Normal



Loss of fat pad – scaphoid fracture



Scaphoid fracture



Missed



6 weeks later



Missed



Recognized later

Rotary subluxation of scaphoid

Increased gap between scaphoid and lunate bones which should not exceed $>3\text{mm}$. The shape of the scaphoid assumes a circle and is called Signet ring sign.



Normal scaphoid
- Lunate relationship



$> 6\text{mm}$

Scapholunate dissociation with distal radial Torus fracture



Do not be satisfied with one finding, look all around

Scapholunate dissociation (SLD) is the most common type of carpal instability. Another reason is that compression forces transmitted through the capitate has a tendency to separate the scaphoid and lunate. It occurs due to injury to scapholunate interosseous ligament (SLIL). SLIL may either be injured alone or in combination with other ligaments such as radioscapohcapitate ligament or dorsal intercarpal ligament.

Acute trauma – 9 yr old



Incomplete mineralization of pisiform at this age should not be mistaken for trauma

Bennett fracture

Fracture of the base of the thumb resulting from forced abduction of the first metacarpal. It is defined as an intra-articular two-part fracture of the base of the first metacarpal bone.



Actually Salter II



Bennett fracture dislocation



Normal



**Subluxation at 1st
carpo metacarpal joint
(missed)**



**Dislocation of the first carpo
metacarpal joint.**



Fracture neck of 5th metacarpal



Fracture base of proximal phalanx of 4th finger

Fracture neck of 2nd metacarpal



Seymour Fracture



Buckle fracture metaphysis of proximal phalanx of the thumb



The Seymour fracture is a clinically important subtype of mallet finger type injury. It is comprised of a distal phalanx physeal fracture that has an associated nail bed injury commonly with unguis subluxation.



Fracture distal radius

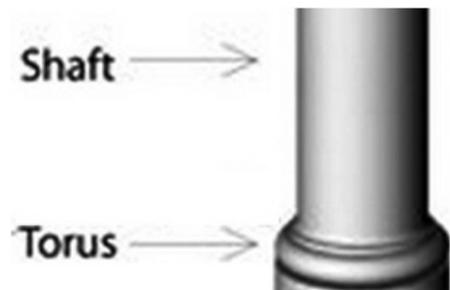


Impacted fracture radius

Torus fracture



- ❖ Due to longitudinal compressive force
- ❖ Also called as buckle fracture: focal angular deformity at metaphyseal-diaphyseal junction of long bones.



Green stick fracture



Buckling Fracture



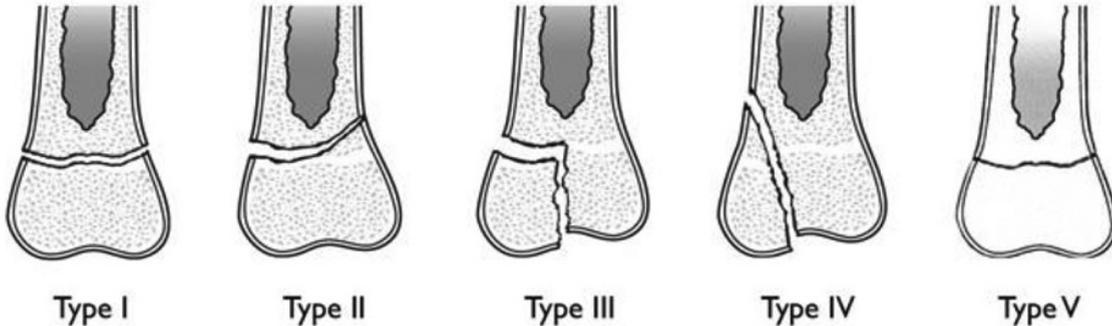
Distal radioulnar dislocation



Periosteum is intact. There is no displacement. (Green Stick # - Only one cortex is broken the other cortex is intact)

Salter Harris Physeal Fractures

Salter-Harris Classification



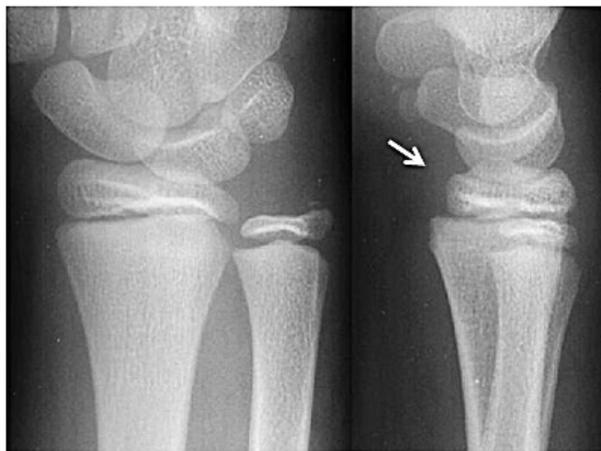
Of these types, type 1 and 5 fractures are frequently missed

Salter Harris Fracture: Type VI

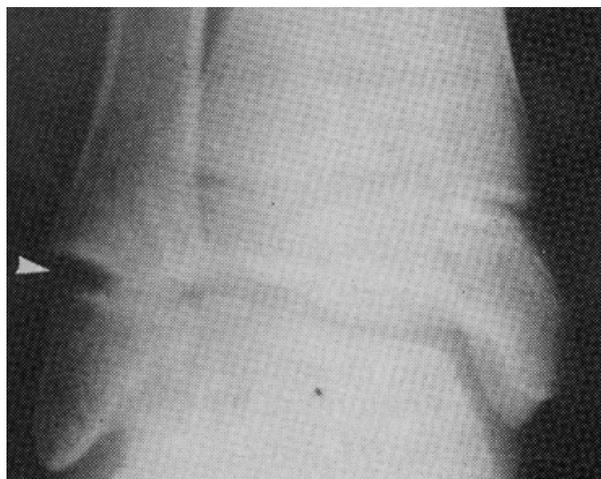
A newer classification, called the Peterson classification, adds a type VI fracture, in which a portion of the epiphysis, physis and metaphysis are missing.



Salter 1 fracture



Radius - Missed on PA view



Fibula



Base of proximal phalanx



Salter I fracture - Missed on PA view



Base of proximal phalanx



Mallet fracture Middle phalanx

Salter II fractures



Metaphysis of 1st metacarpal

**Salter Harris Fracture
Type III**



**Salter Harris Fracture
Type IV**



**Salter Harris Fracture
Type V**



Missed on PA view

Gymnast's wrist with physeal damage



Mistaken for rickets



Physeal injuries are also common in neurological states. Eg., congenital insensitivity to pain

17 yr old, osteochondral fracture, distal end of radius



Late Sequelae Of Missed Physeal Injury

Growth arrest



Madelung deformity

Physeal bar



Growth arrest



Common complication of type 5 & 6 Salter Harris fractures

Fracture mimics

Fracture mimics involve normal variations simulating fractures. An accessory centre or mach line may be called a fracture.



The normal spurlike projection of the epiphysis at the epiphyseal line may simulate avulsion injury.



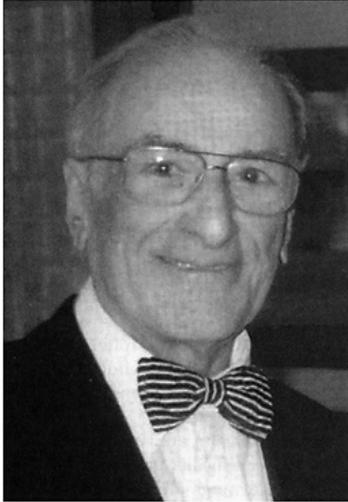
Physeal line - not to be mistaken for an impacted fracture

Causes Of Missable Fracture

- ❖ **Poor technique**
- ❖ **Inadequate investigation**
- ❖ **Perceptual error**
- ❖ **Incomplete knowledge**
- ❖ **Misjudgement**
- ❖ **Combination of all the above**

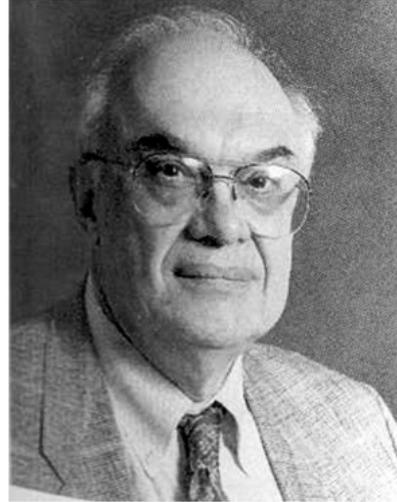
“The eye cannot see what the mind does not know”

Extra Reading



**Hooshang Taybi,
1919-2006**

Taybi H, Lachman RS Radiology of syndromes, Metabolic disorders and Skeletal Dysplasias (4th Ed.), Chicago year book. Out of 600 syndromes 500 involve hands.



Andrew K. Poznanski

**Poznanski Andrew K.
The Hand in
Radiologic diagnosis.
3rd Edition
Philadelphia. W. B. Saunders**

- ❖ **Kakarla Subbarao, X-ray palmistry in pediatrics; Proceedings of the Academy of Medical sciences, Andhra pradesh, Vol – 7 , No. 1, Jan 1965.**

- ❖ **Kakarla S, Benign tumors of the bones of the hands; JMSR, 2018; 6(2): 49-58. DOI: <http://dx.doi.org/10.17727/JMSR.2018/6-9>**

- ❖ **Kakarla S Tumoral lesions of the bones of the hands – Pictorial essay; J Med Sci Res 2018; 6(3): 86-92. DOI: <http://dx.doi.org/10.17727/JMSR.2018/6-15>**

- ❖ **Kakarla Subbarao – Pediatric Bone Tumors Chapter 31; Section VI Skeletal system: 710-746 (Selected topic from pediatric radiology 2018 – Editor MD Rahalkar's).**



Prof. Kakarla Subbarao

MBBS, MS (Rad), FFR (Eng), FACR, FICR, FICRI, DSc (Hon)

Prof. Kakarla Subbarao, 94 is a world renowned radiologist. He was a professor of Radiology at Osmania Medical College, Hyderabad. Subsequently, he left for US and he served as professor of Radiology at Albert Einstein College of Medicine, New York, and New York College of Podiatric Medicine. Upon returning to India, he worked as founder Director of Nizam's Institute of Medical Sciences (NIMS), Hyderabad, and subsequently Director, Vice chancellor of the same institute. He was Chairman of Indian College of Radiology and Imaging and National President of Indian Radiological and Imaging Association. He was Founder President of Musculoskeletal Society of India. He was founder of Susruta Society of Radiology now named as Association of American Radiologists of Indian Origin (AARI). He was Founder Chairman of Telugu Association of North America. He was Life time Trustee of Andhra Mahila Sabha. He has established an International School for children to foster the concept of holistic education and integrated personality development. He has been conferred with 'Padma Sri' award by Government of India. He has received Life time Achievement Awards from ten (10) Non-profit organisations. He has won many awards, Gold Medals and Orations in recognition of his selfless services rendered to the nation in the Field Of Medicine.

Presently he is Emeritus Prof. of NIMS, Visiting Professor of KIMS, Sec and Chairman for Krishna Foundation for Research Centre, Hon. Edi. of the Journal for Medical and Scientific Research and Chairman at KREST (Kakarla Subbarao Radiological & Imaging Educational Sciences Trust).



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