Congenital Malformation of the upper limb

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Causes of malformed U.L

* **Faulty gene 30%**
* **Environmental**

  Teratogenic agents
* **Syndromatic**
* **Unknown 60%**

1-Thalidomide Tragedy - 1960s

2-Chernobyl Disaster in Russia

Classification

Swanson A.B. JHS vol.1 no.1 p8-22

- I Failure of formation of parts
- II Failure of Differentiation
- III Duplication
- IV Overgrowth
- V Undergrowth
- VI Congenital constriction band synd
- VII Generalised skeletal abnormality

1- Failure of formation

*Amelia, Phocomelia

* Transvere arrest

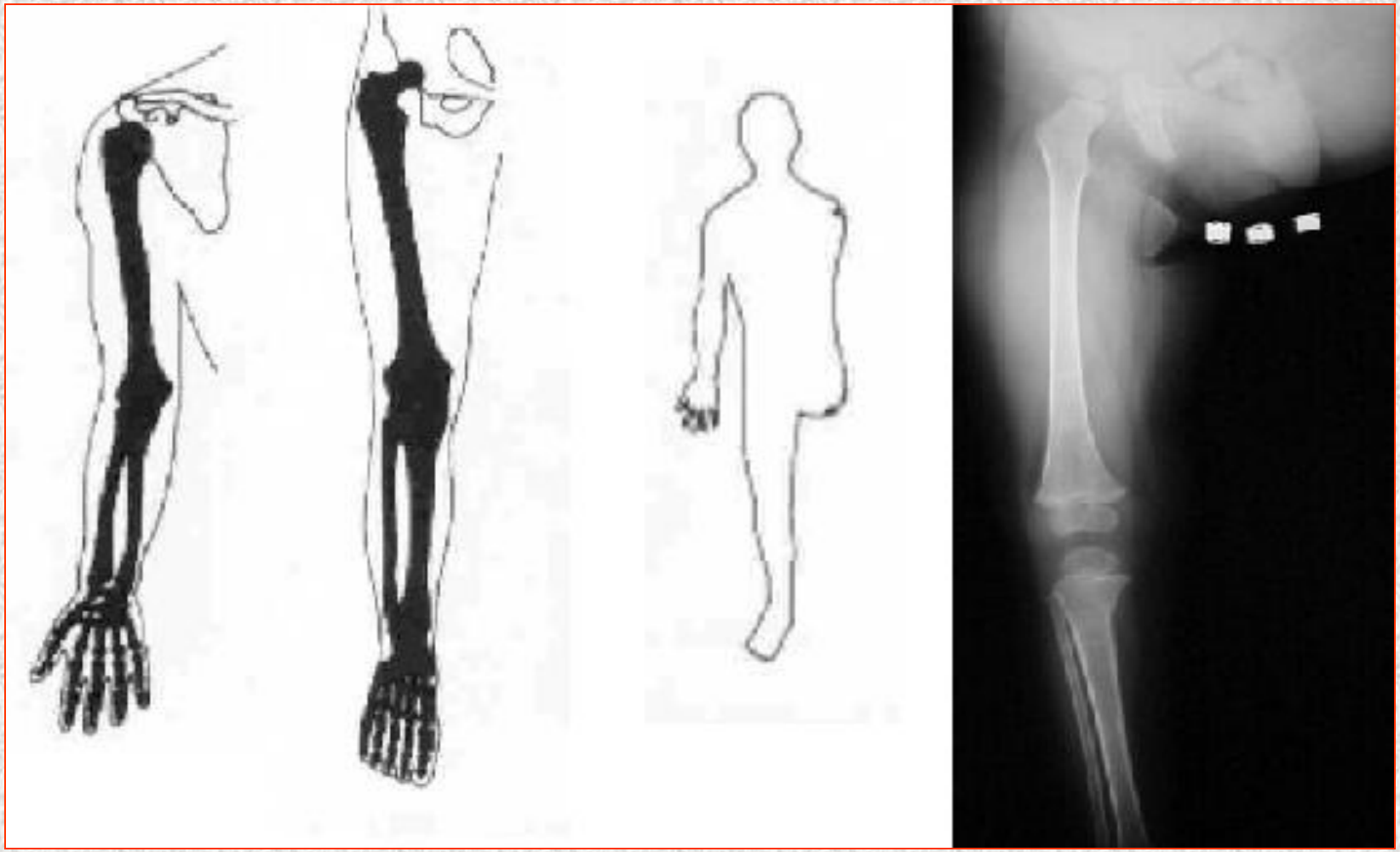
* Longitudinal arrest

* Central arrest

Amelia

Complete absence of a limb beyond a certain point →leaving a stump.

Amelia



Phocomelia

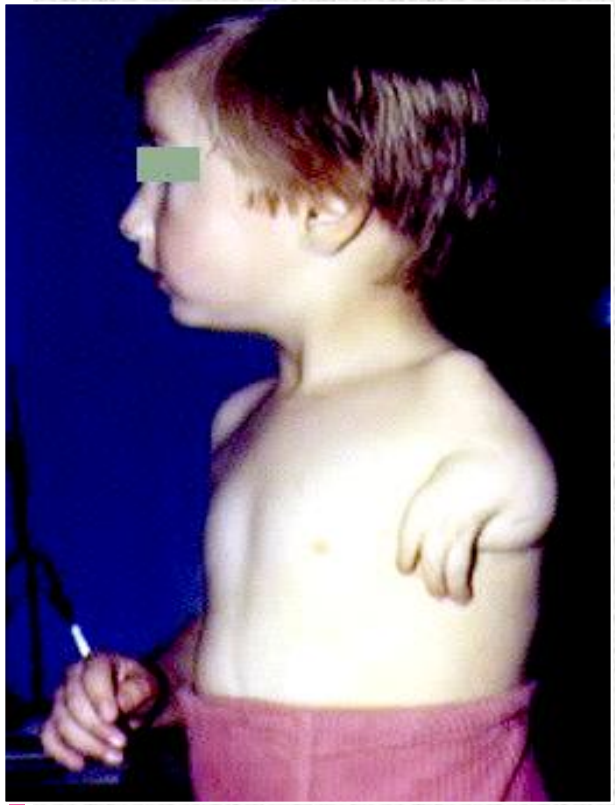
Phocomelia (seal-limb) is a congenital disorder, in which the terminal portion of the limb is attached directly to the trunk.
It is failure of formation

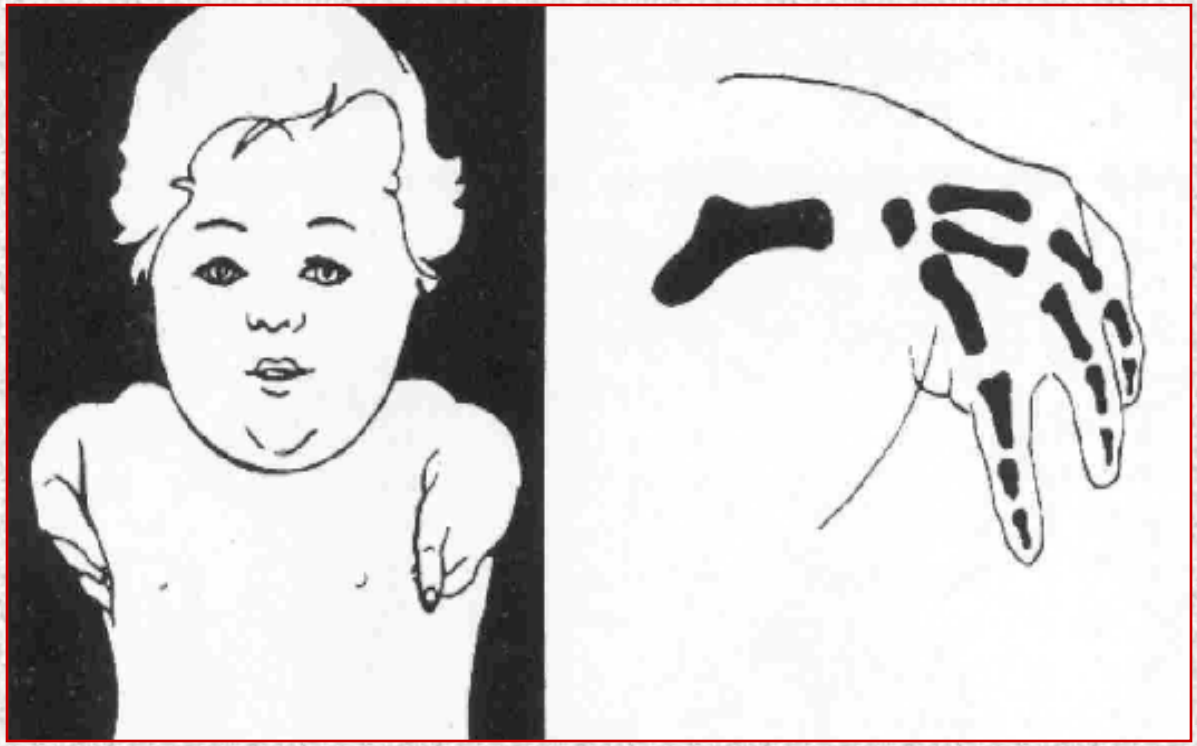
Types :

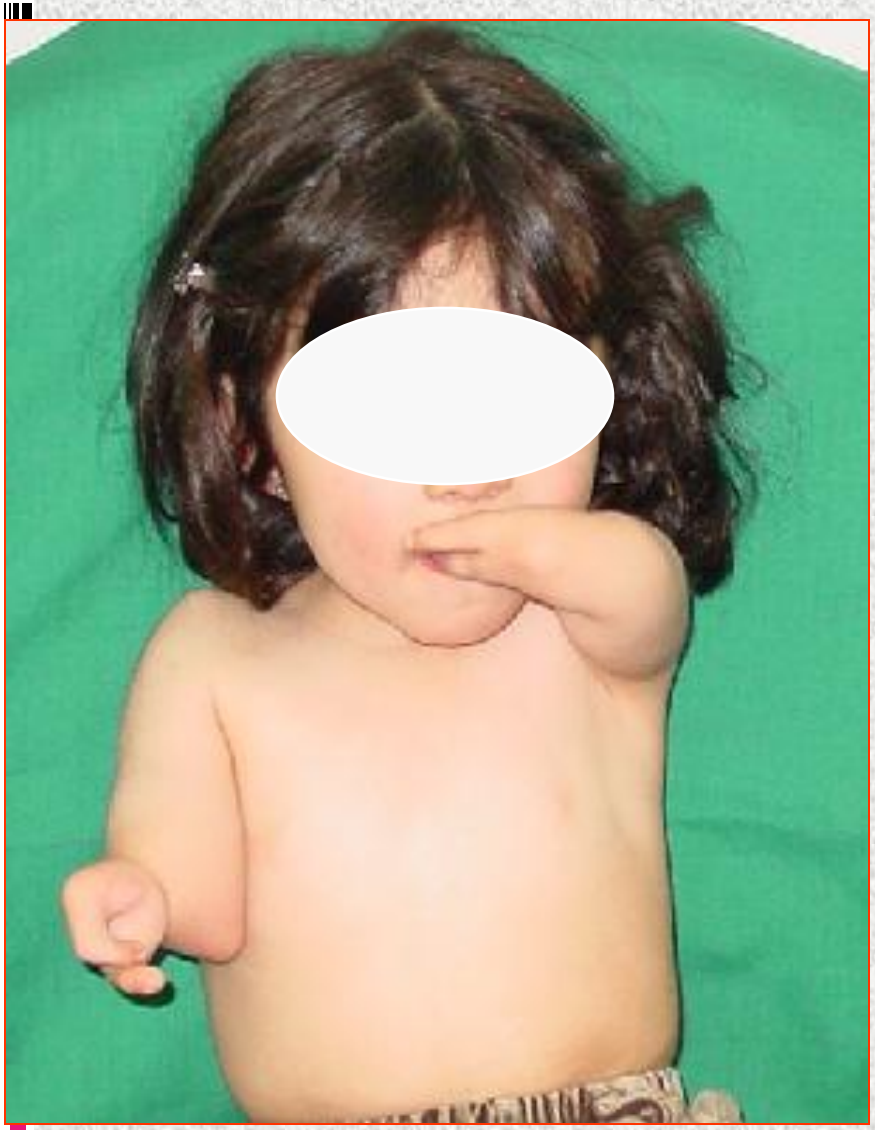
= Hand attached directly to the shoulder
= Forearm attached directly to the shoulder
= Hand attached to the arm

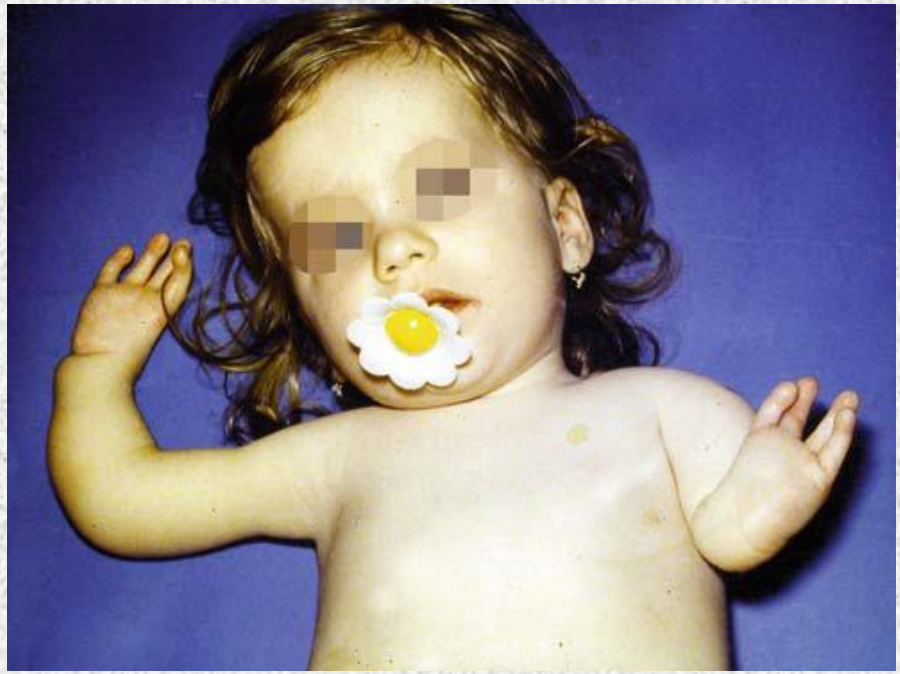
Thalidomide syndrome

Phocomelia

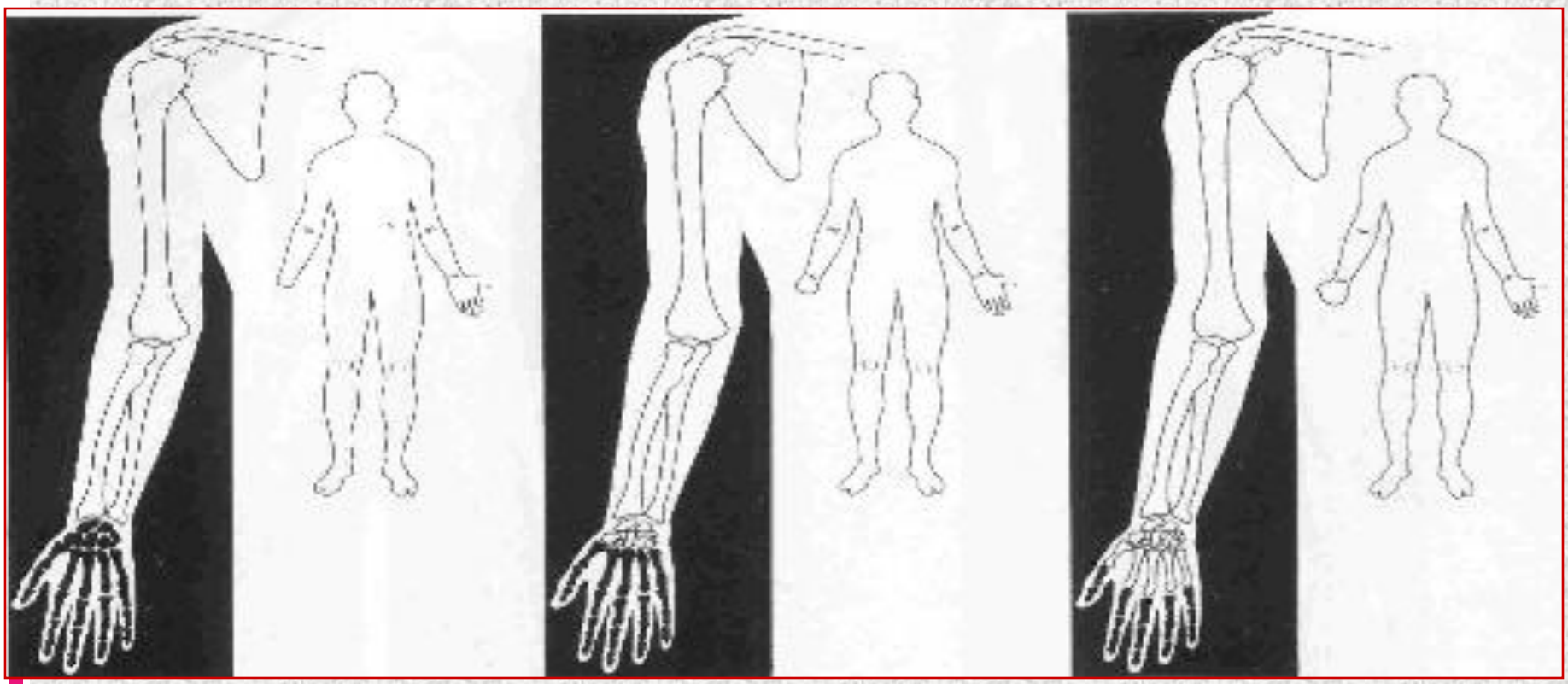


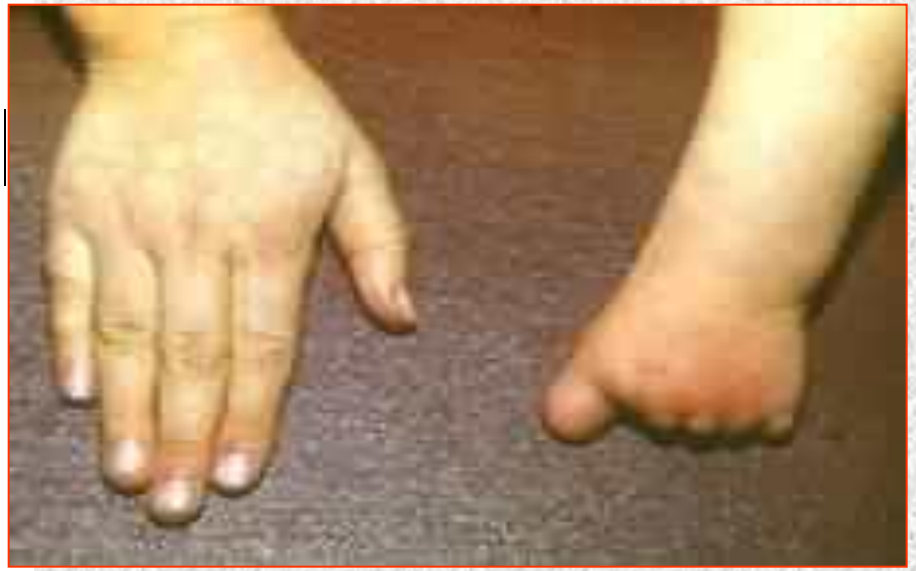


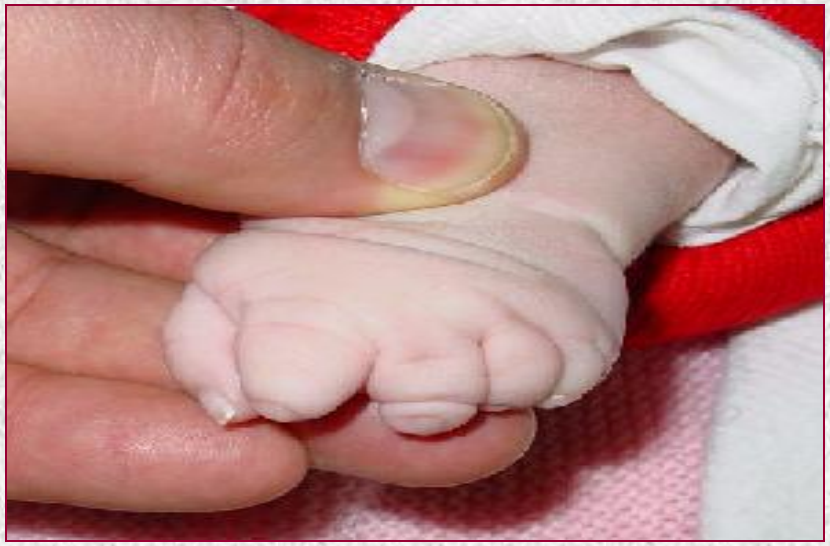


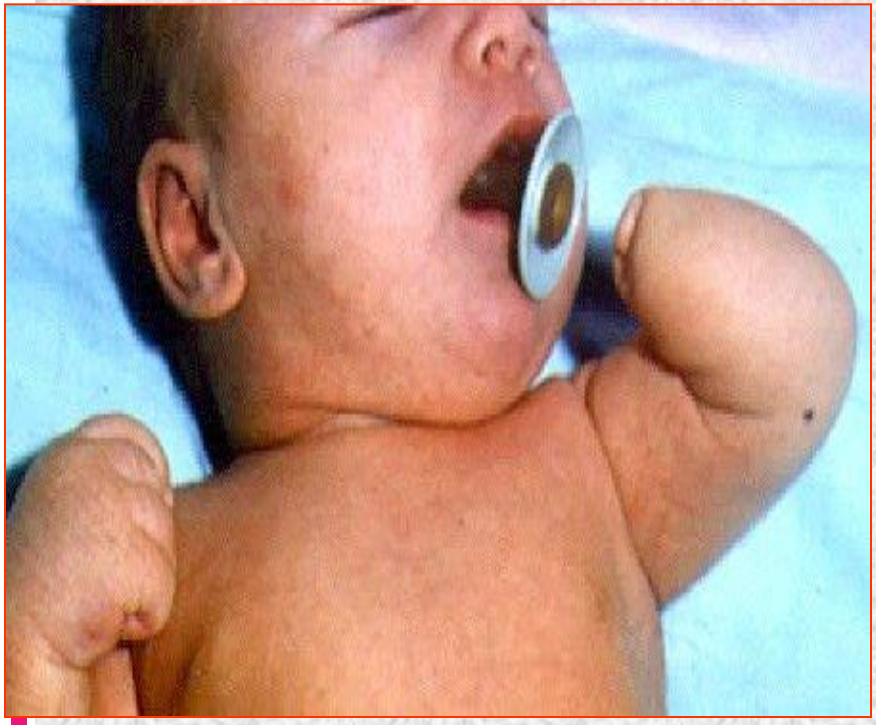


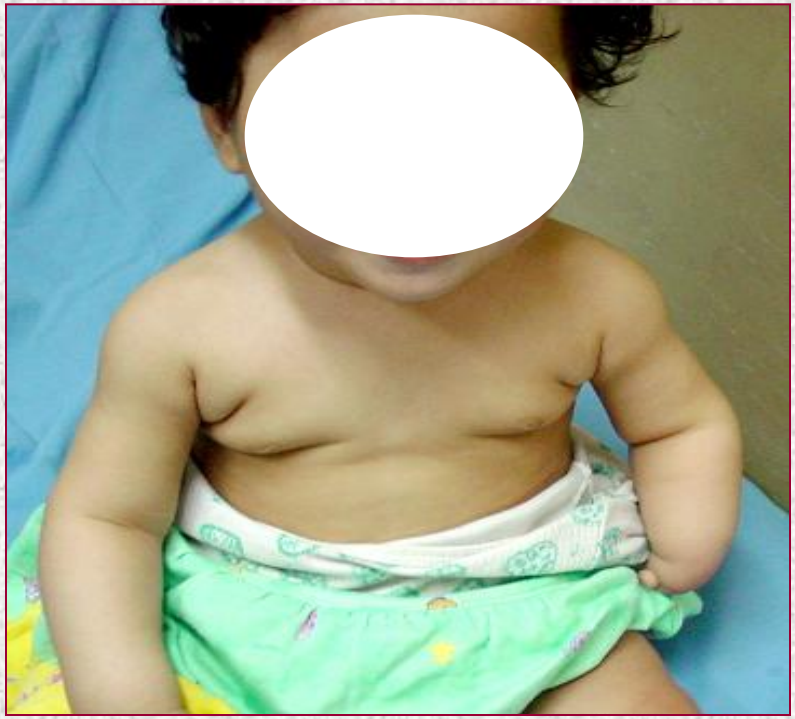
Transverse arrest









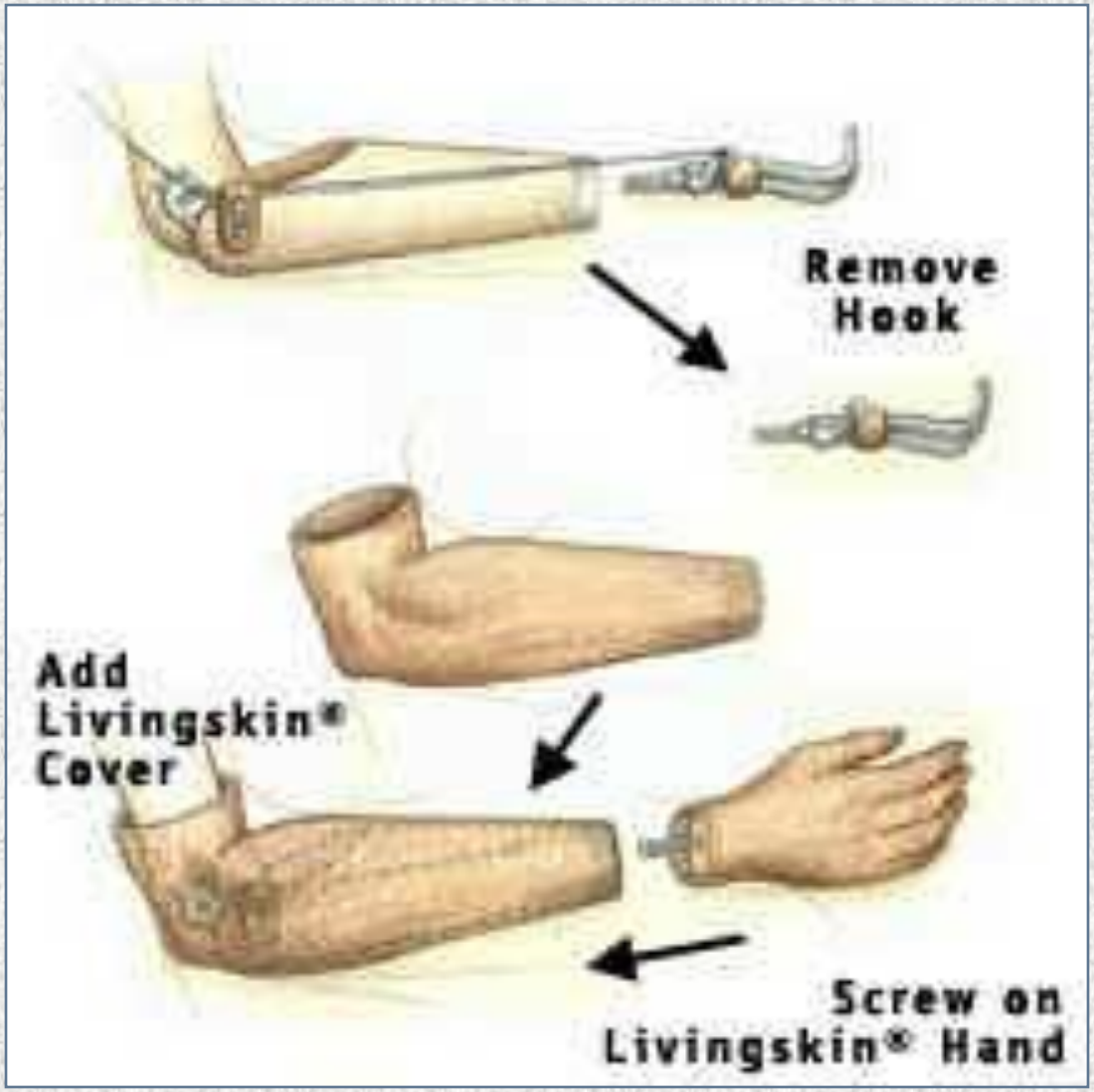


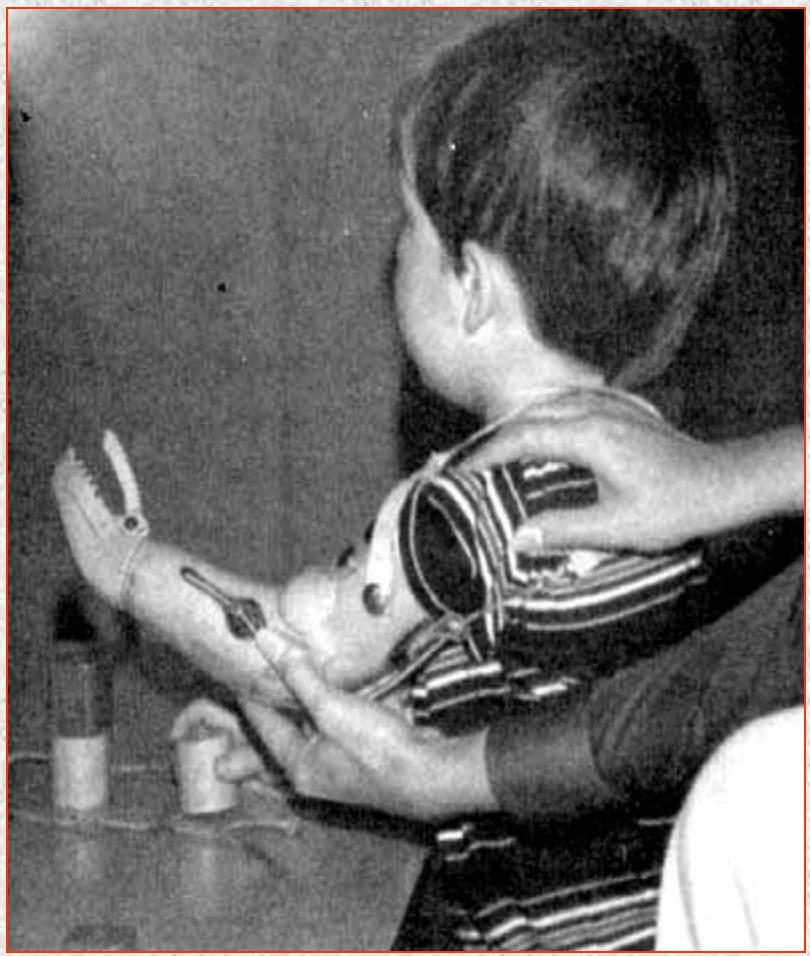
Terminal BEA

* 3-6m: Static prosthesis

-most common Cong. Amput.
-use of a passive device at
3-6 m.
(fit when they sit)

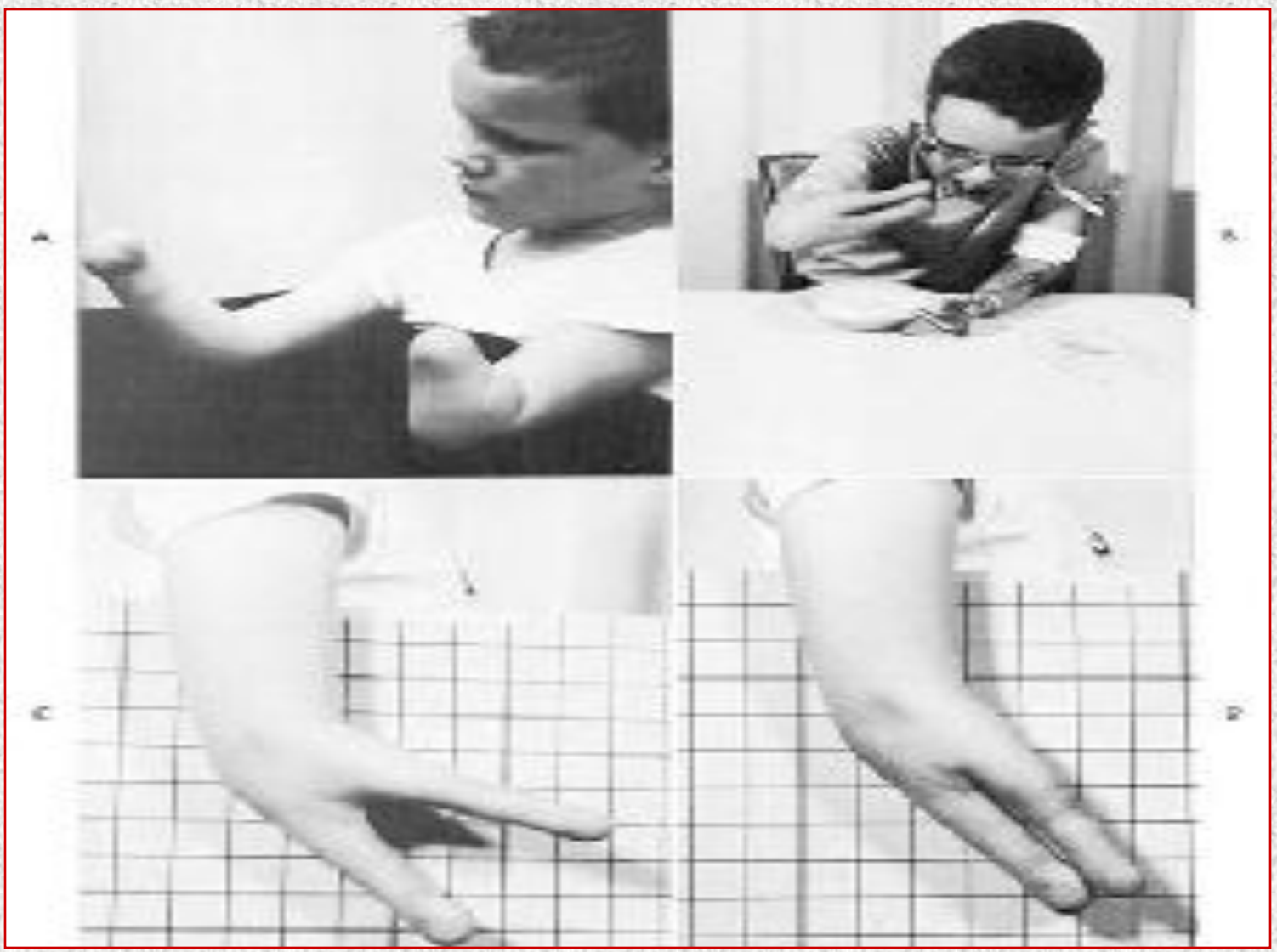
*18 m : Split Hook Prosthesis





Myoelectrical Prosthesis 3-5 years

Krukenberg in blind child



Longitudinal arrest

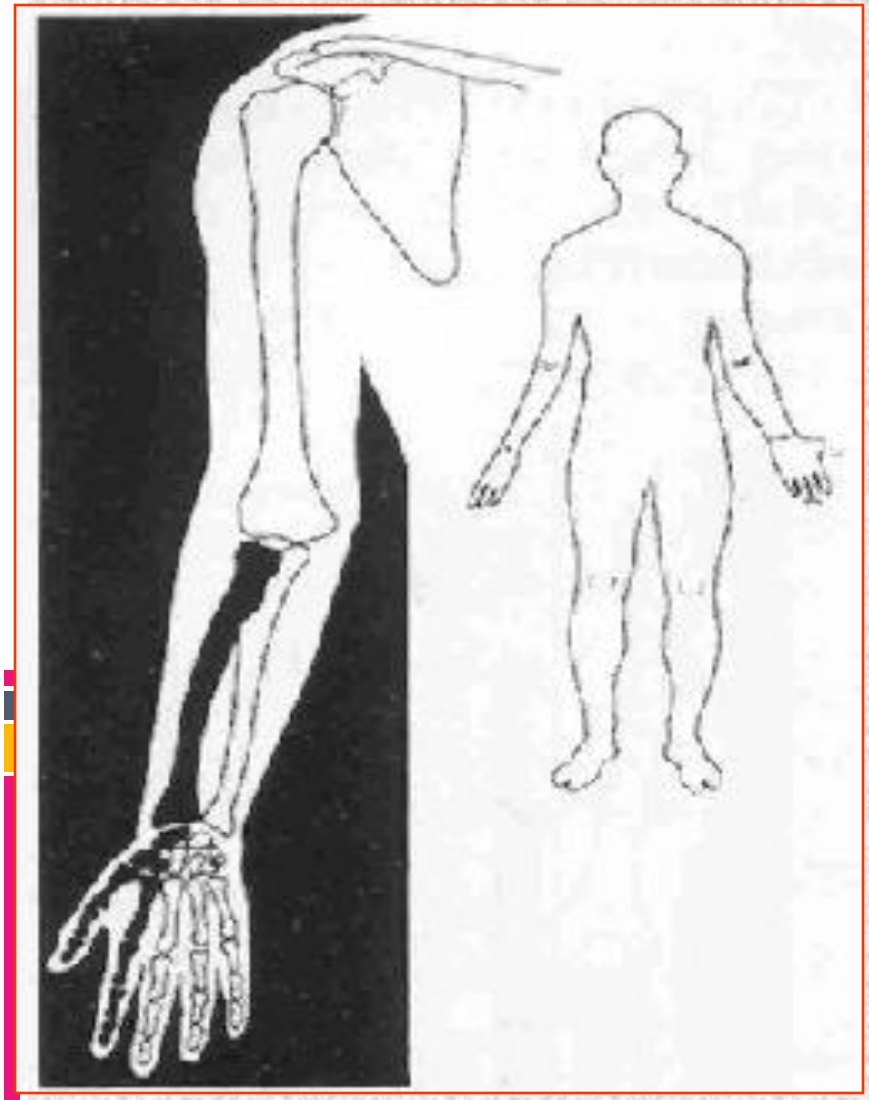
1-Radial club hand

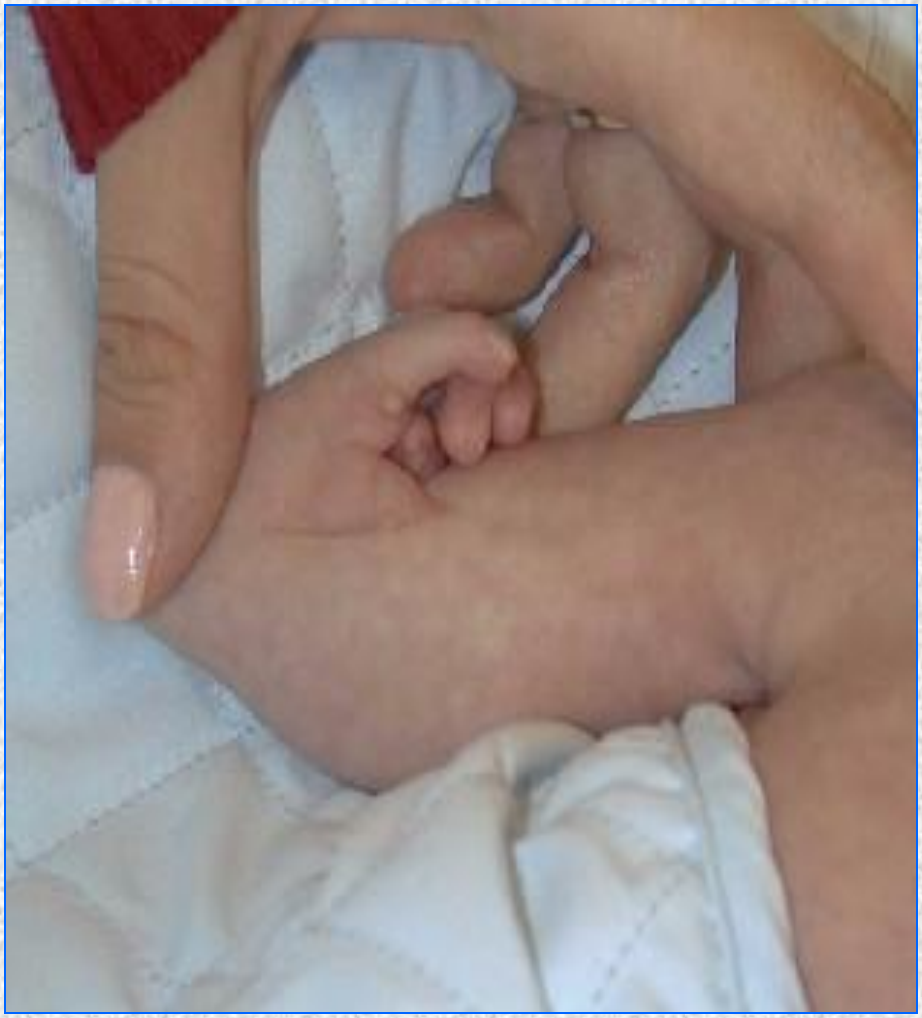
Abscent Structures

* **Long head of biceps 100%**
* **Radialy attached muscles**
* **Absent MCN, Radial A**

Available muscles

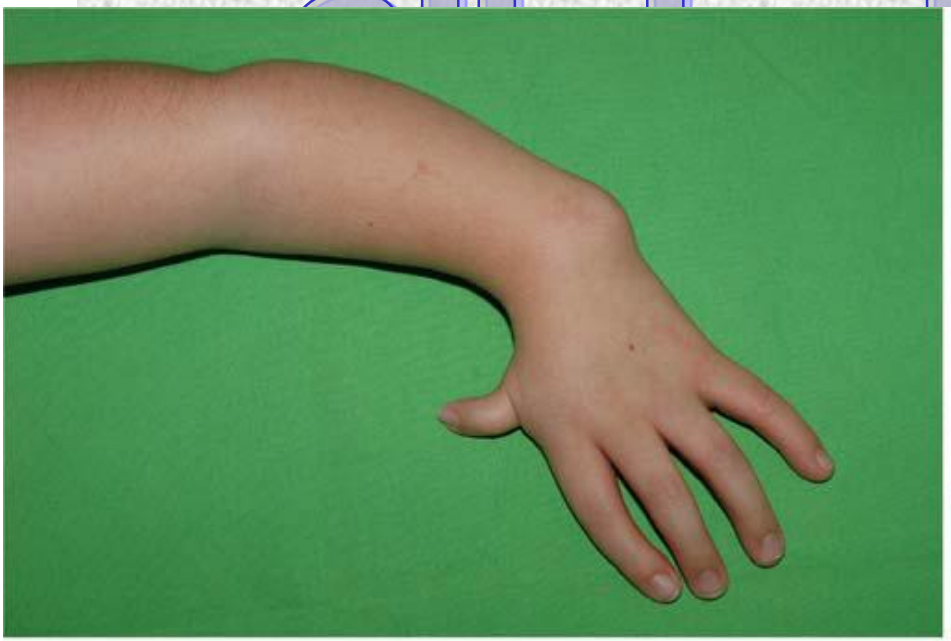
- FDP, FDS
- EDC
- Hypothenars
- Interossi, lumbricals

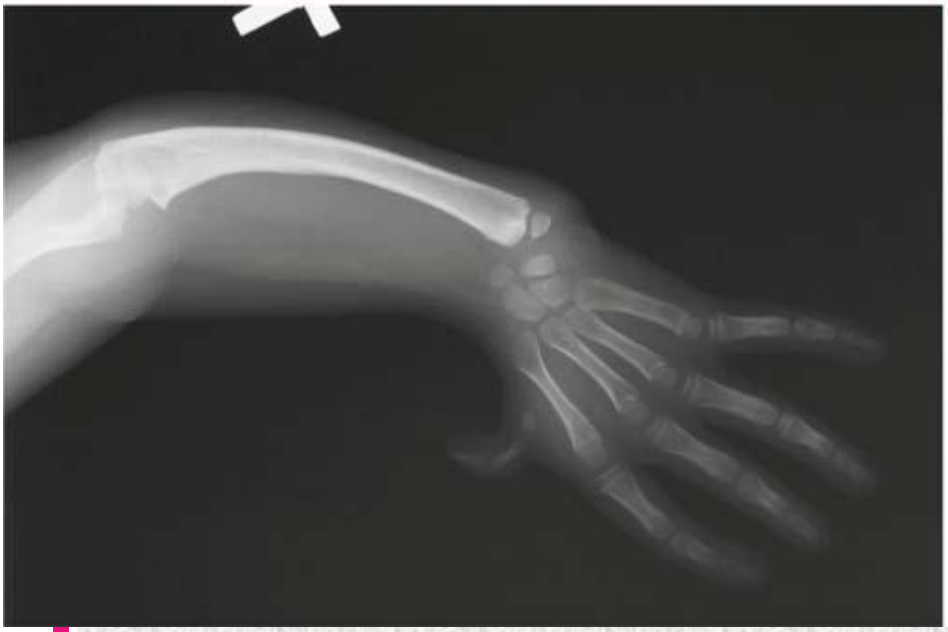
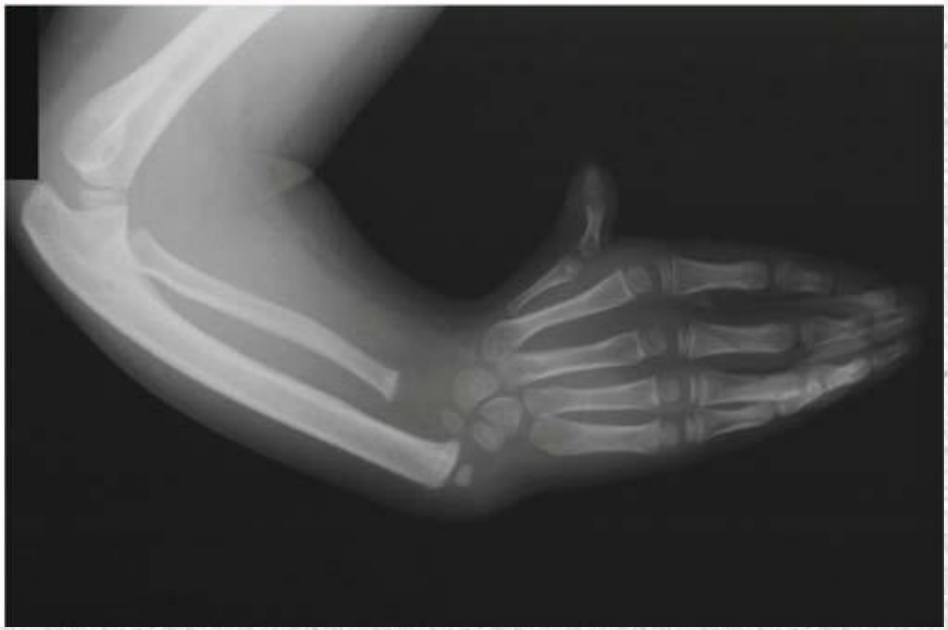


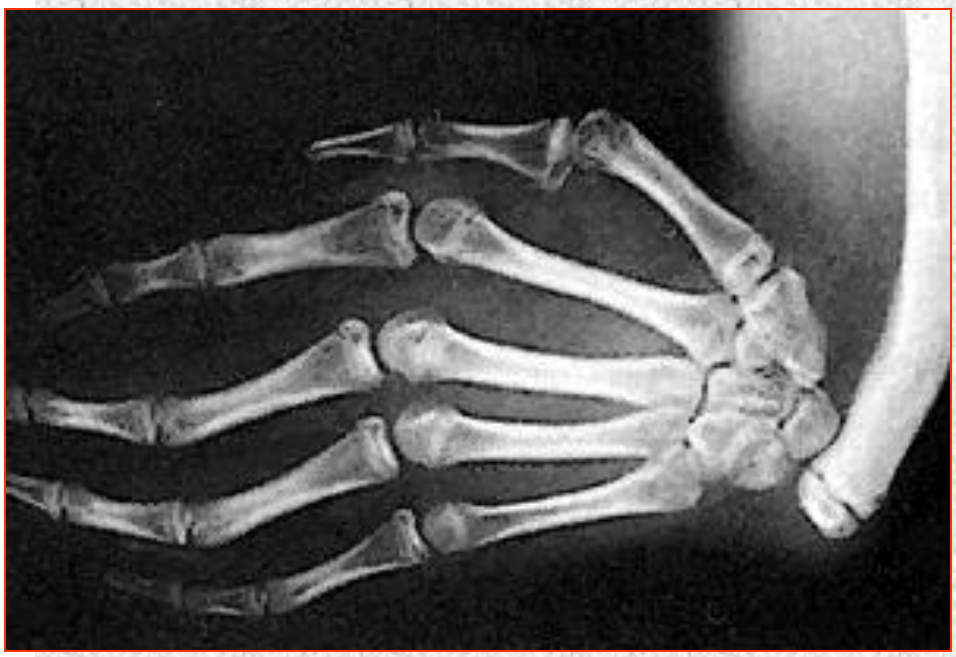


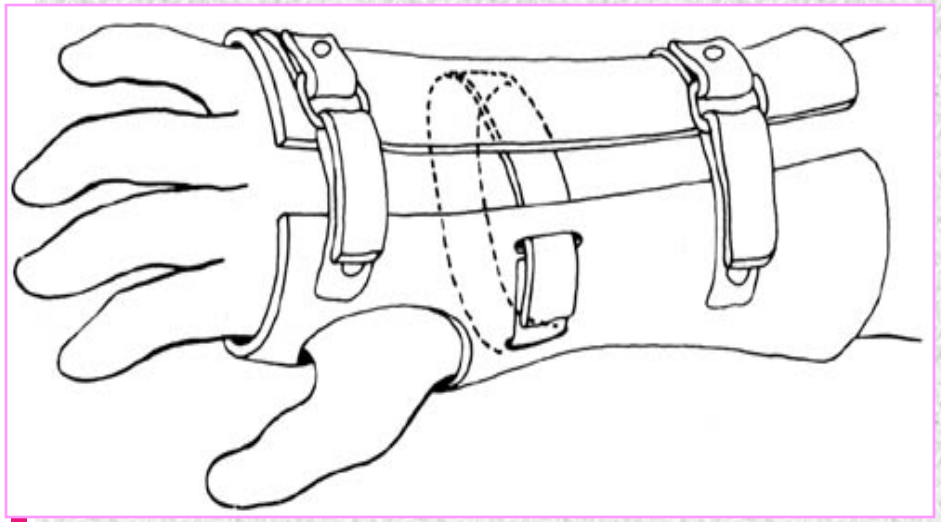
Associated anomalies

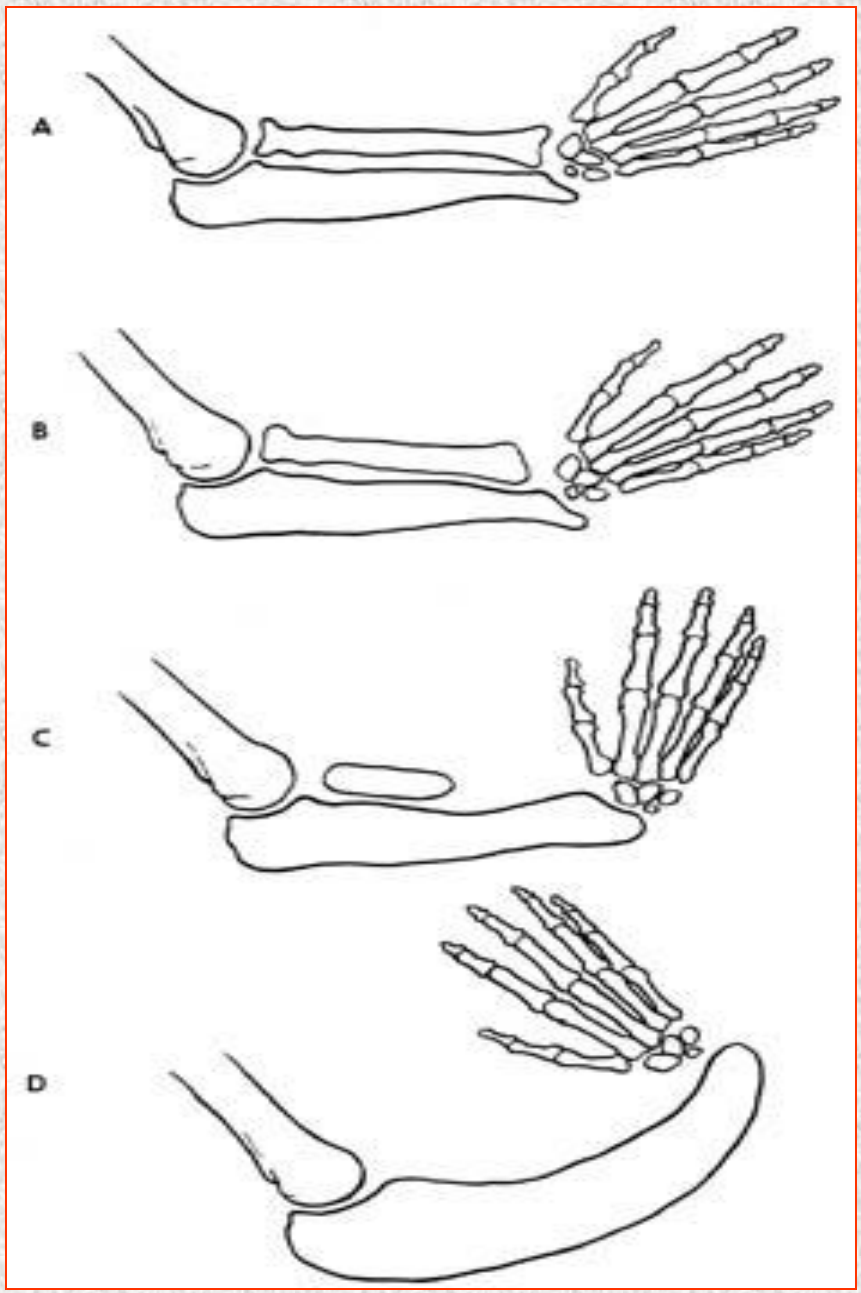
* **Heart**
* **Blood:- (Aplastic An., Thrombocytopenia)**
* **VACTERL**





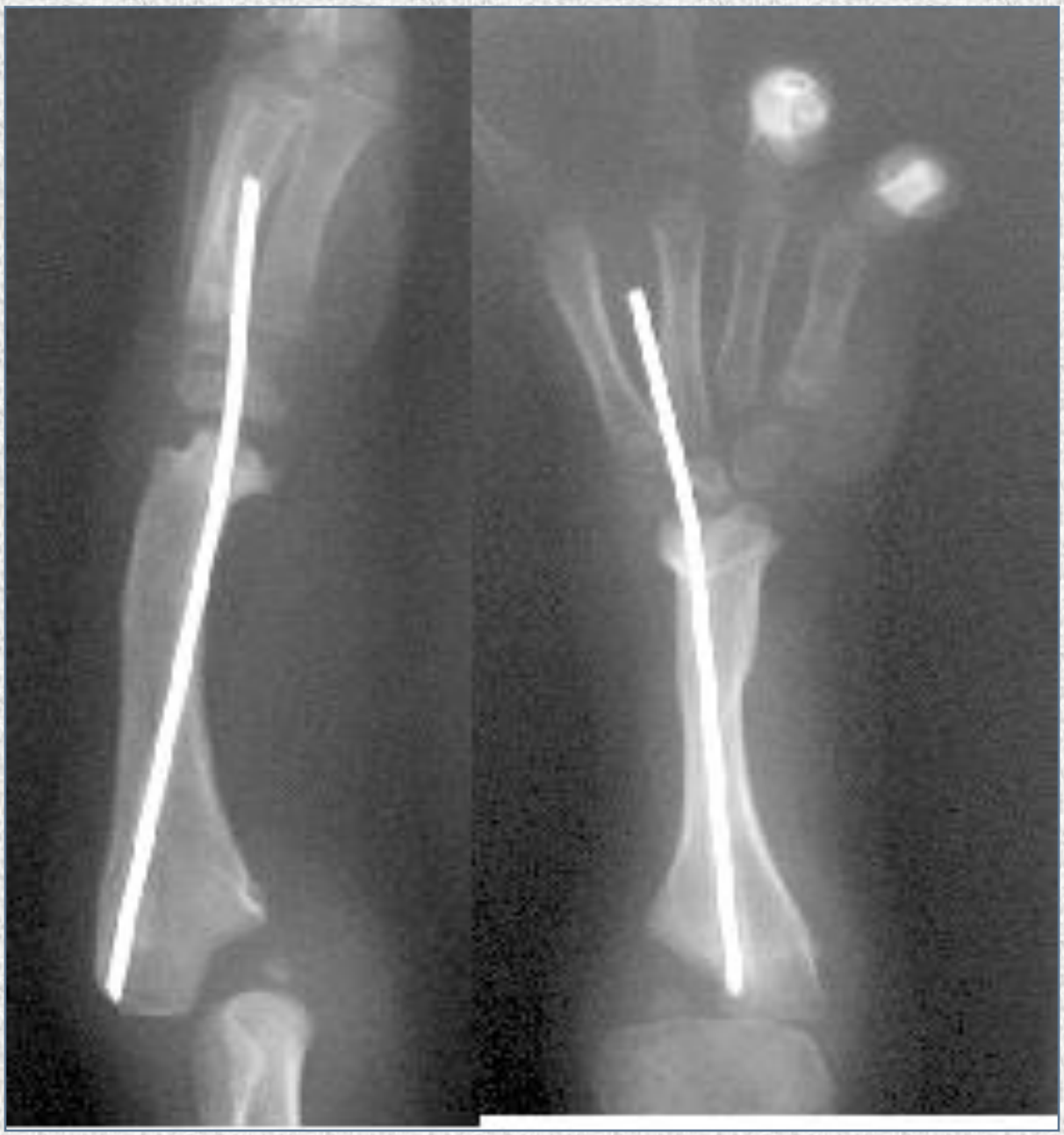



A
B
C
D

Surgery → 3 -12 M

* Centralization :3rd Mc
* Radialization :2nd Mc

(Buck Gramcko)



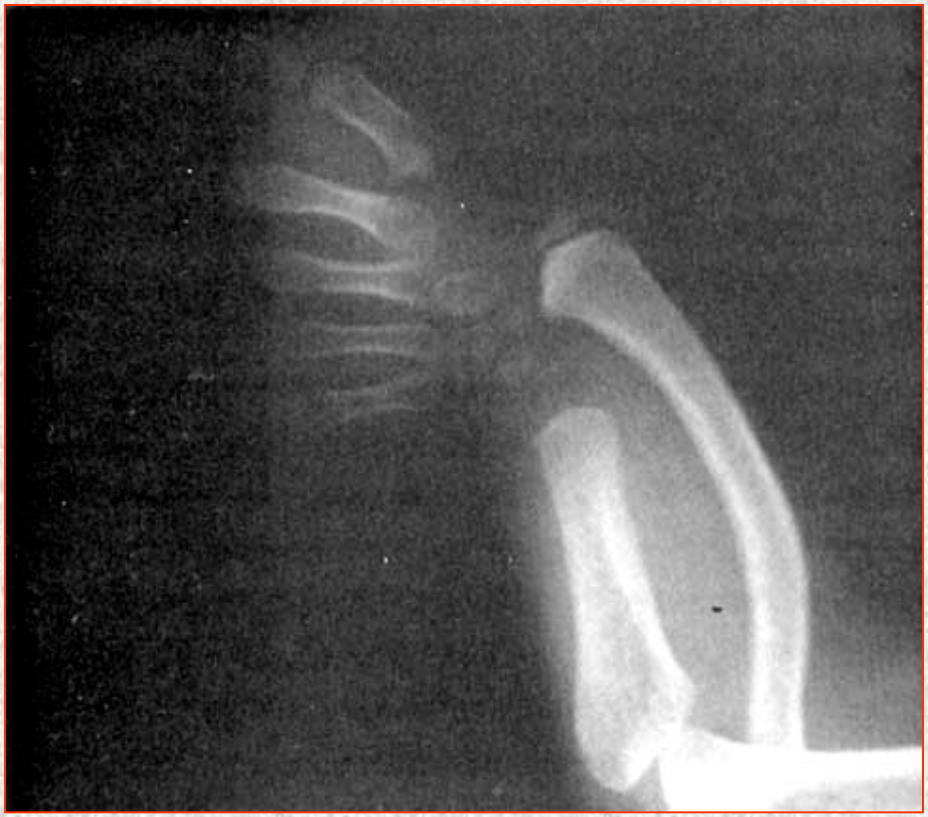
Other Procedures

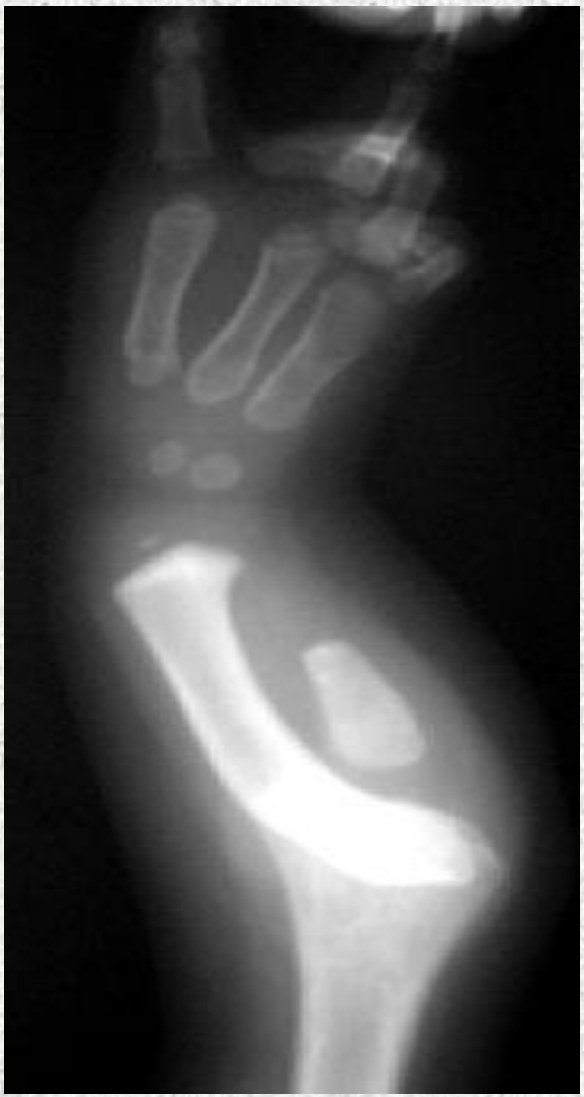
* Soft tissue release
* Ulnar osteotomy
* FCU + ECU transfer to shaft of 5th Mc
* Lengthening

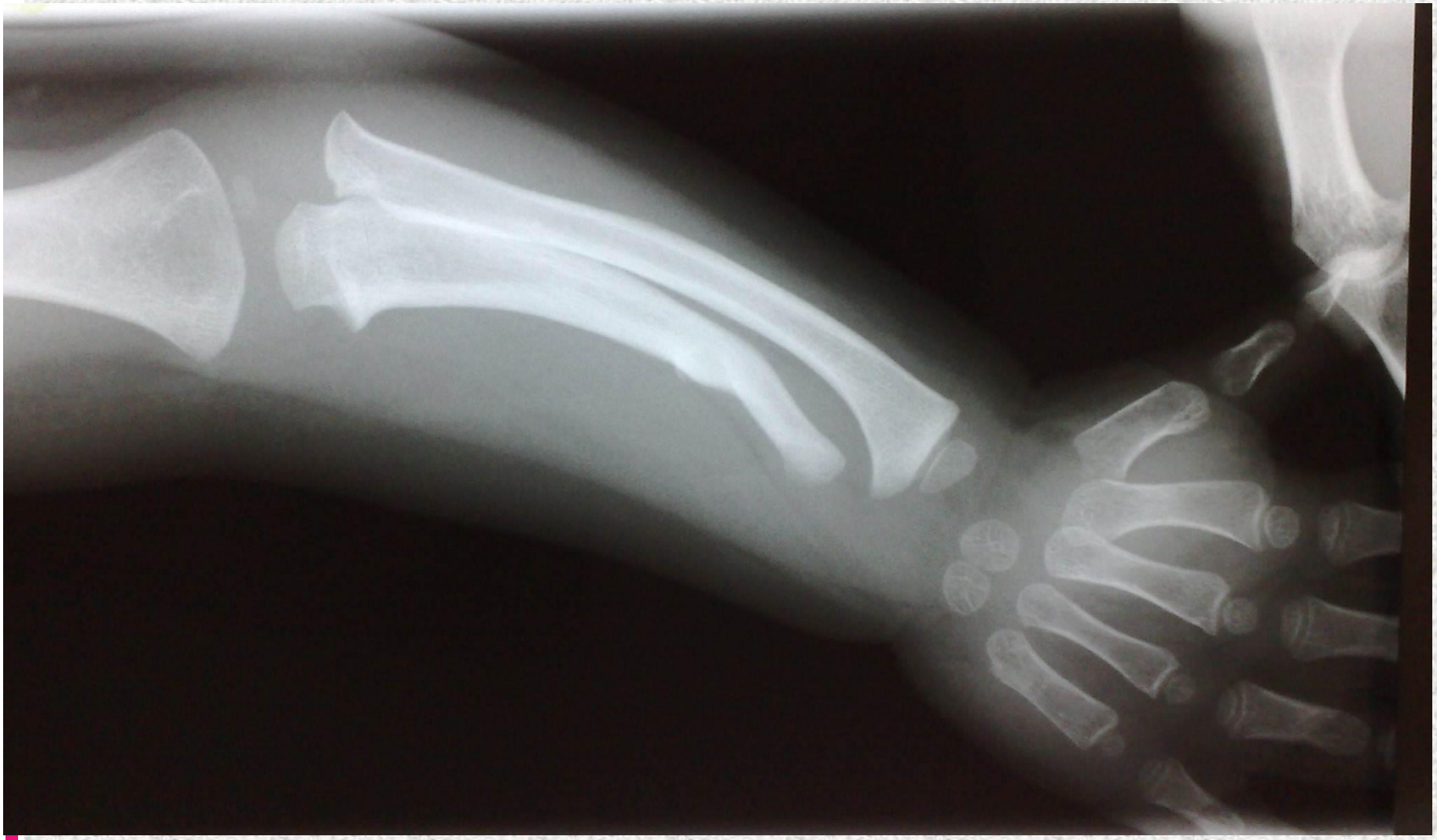
Contra indications to Surgery

* Aplastic anemia
* Stiff elbow
* Type I, II
* Adult

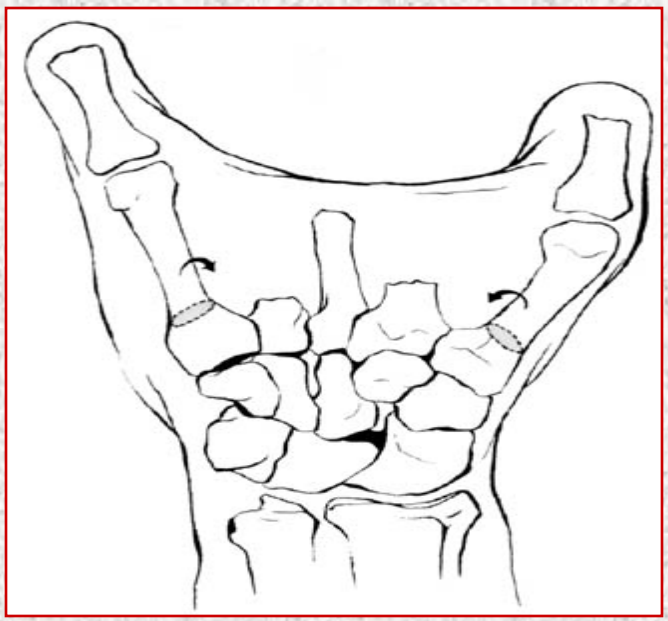
2-Ulnar club hand

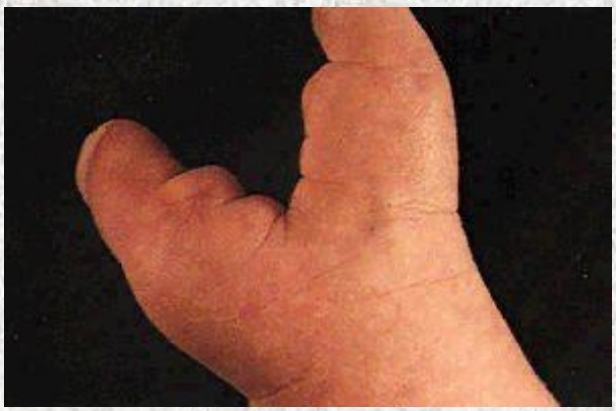






Central arrest

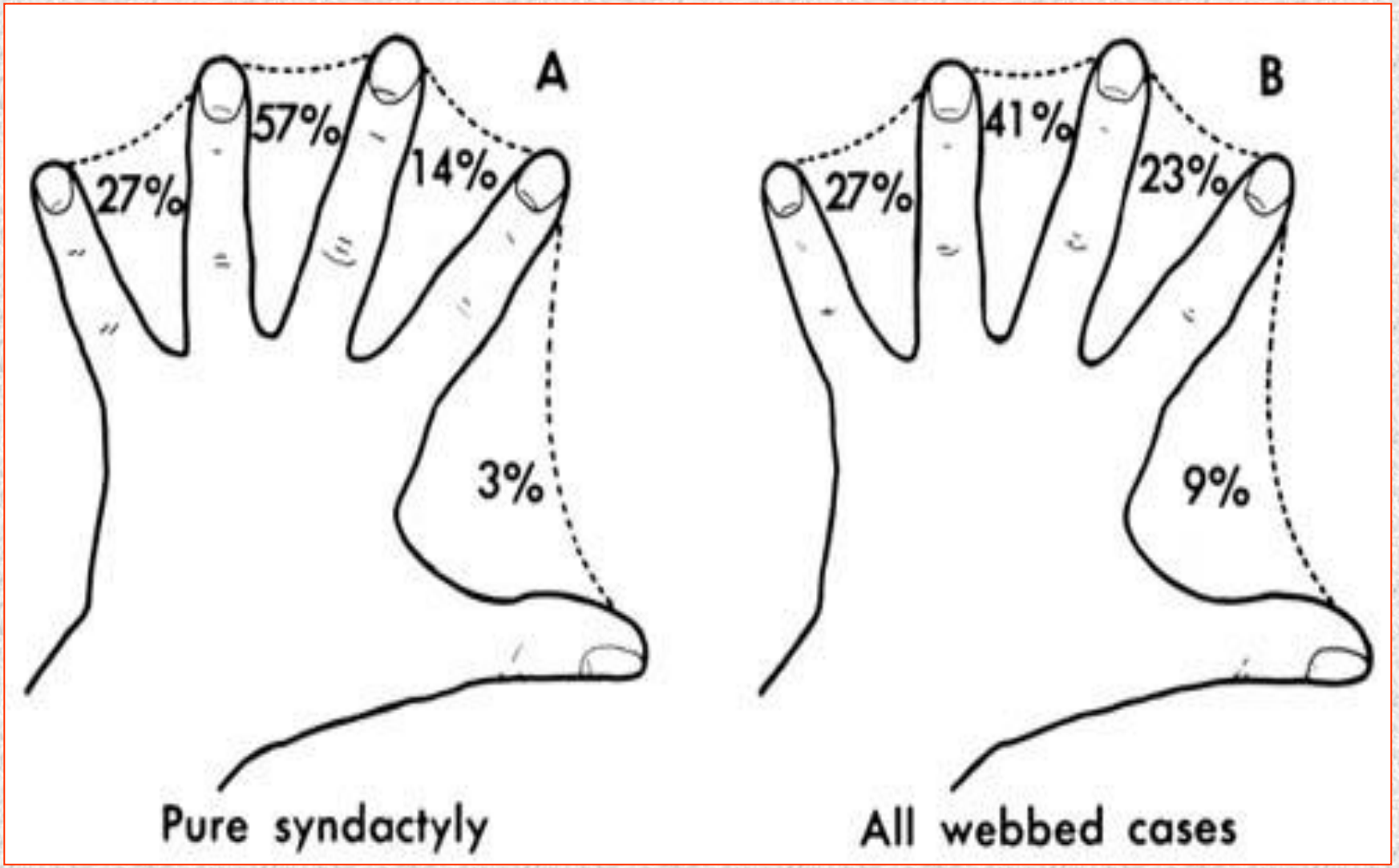




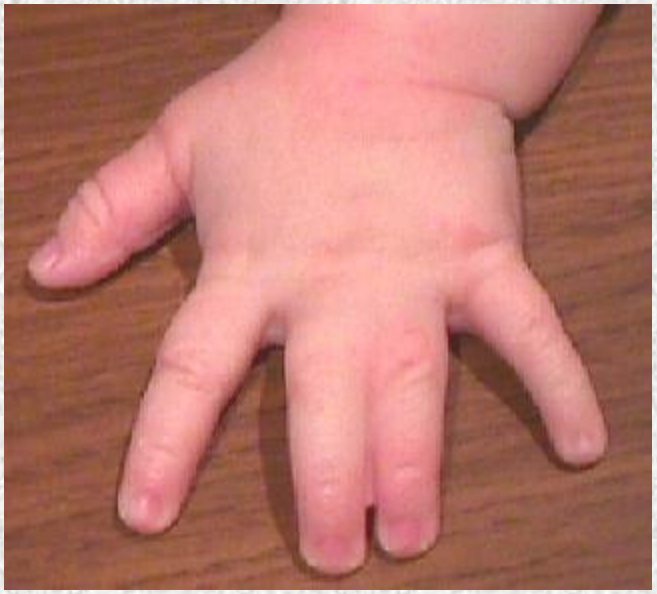
2- Failure of differentiation

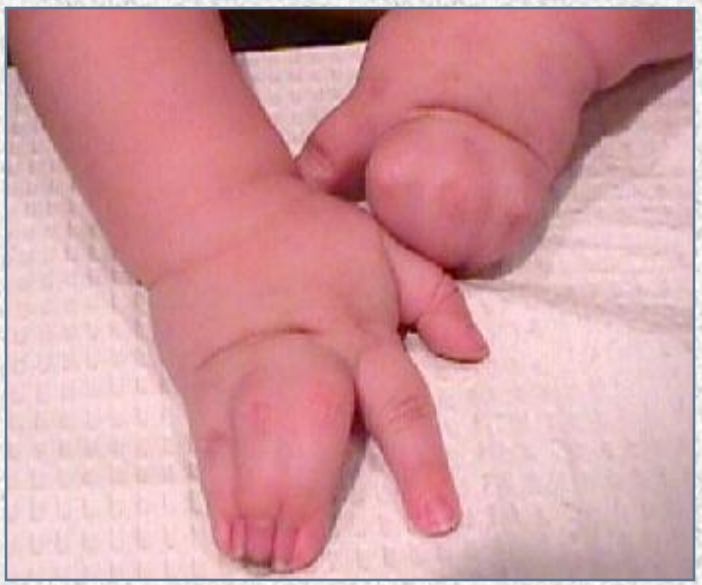
* Syndactyly
* Camptodactyly
* Synostosis

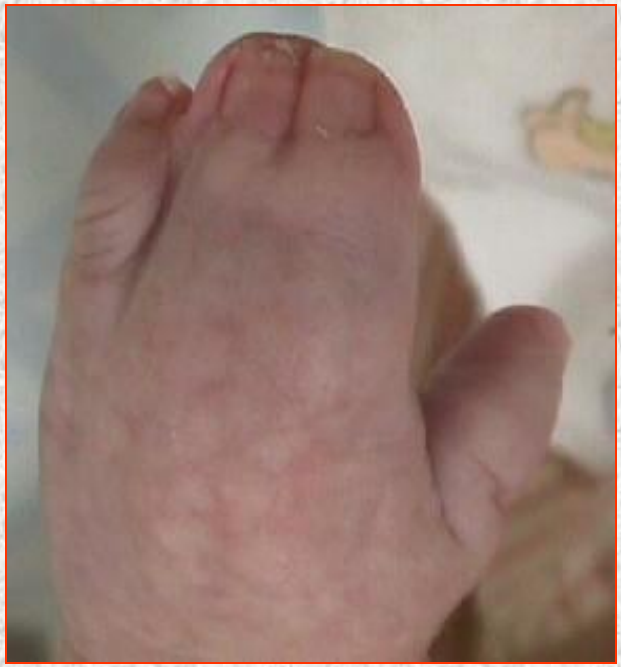
Syndactyly

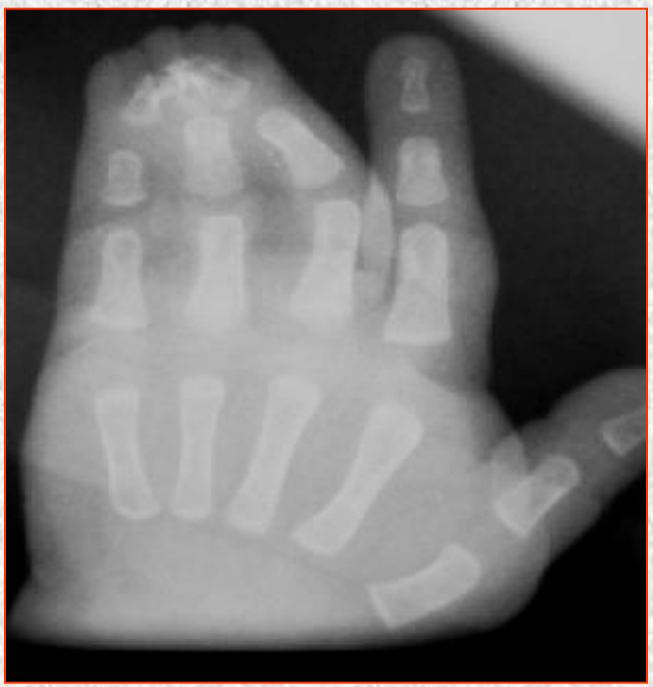


* Complete
* Incomplete
* Simple
* Complex
* Complicated
* Syndromatic : 28









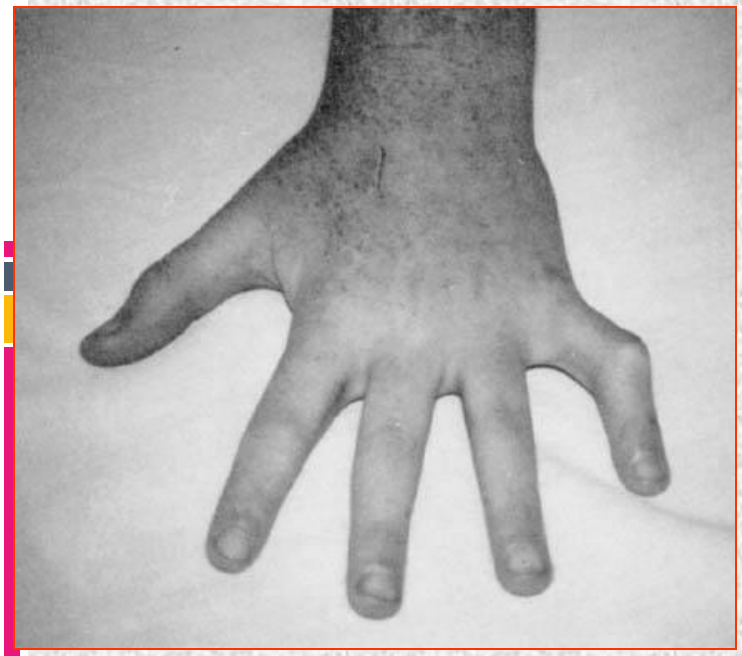
Timing of surgery

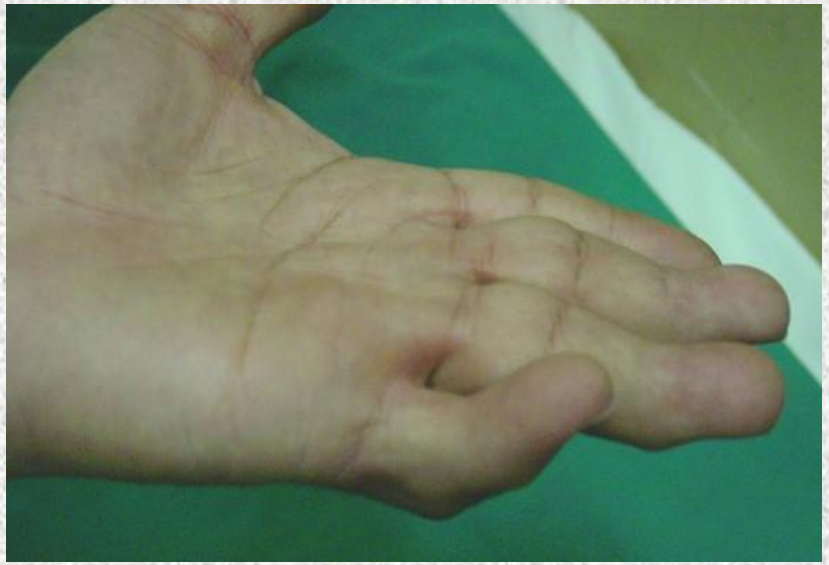
*3 - 12 M

*18 - 48 M

Camptodactly

* **Infantile**
* **Adolescent**





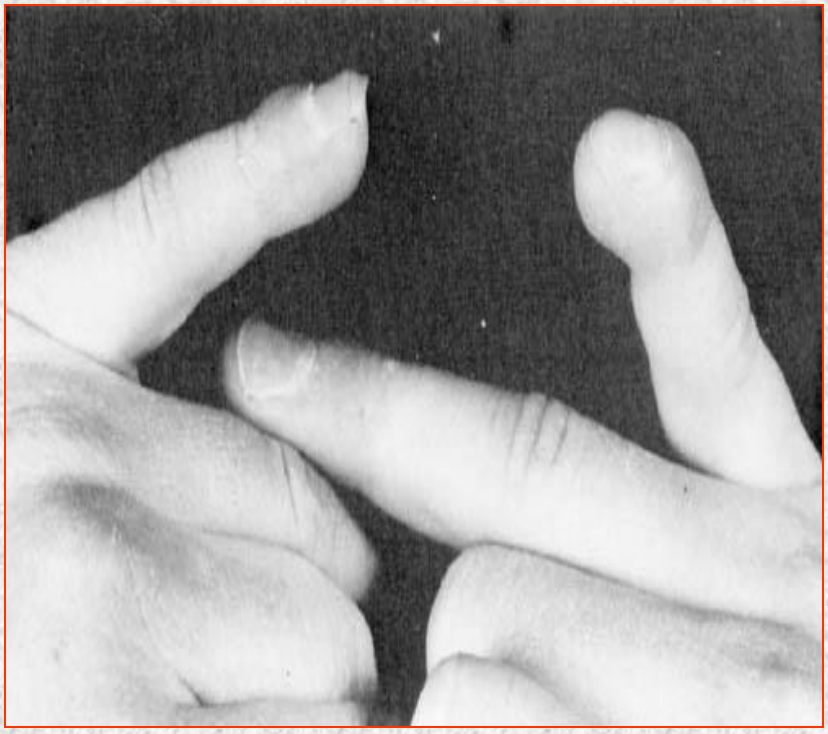
Causes

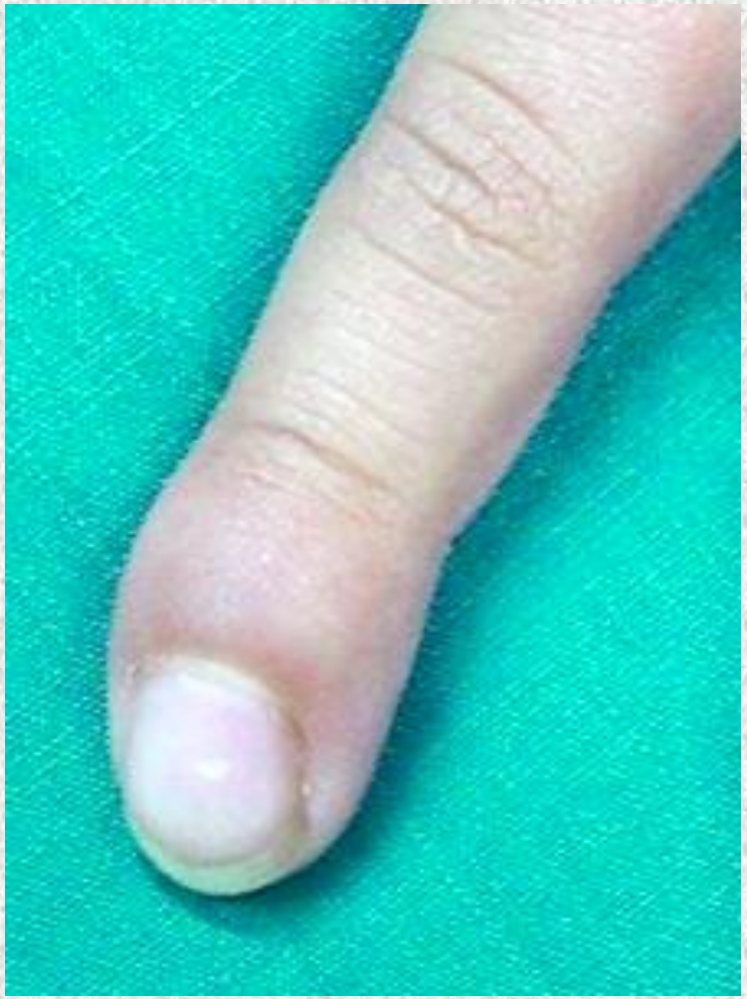
* **Imbalance**
* **FDS**
* **Lumbricals**

Treatment

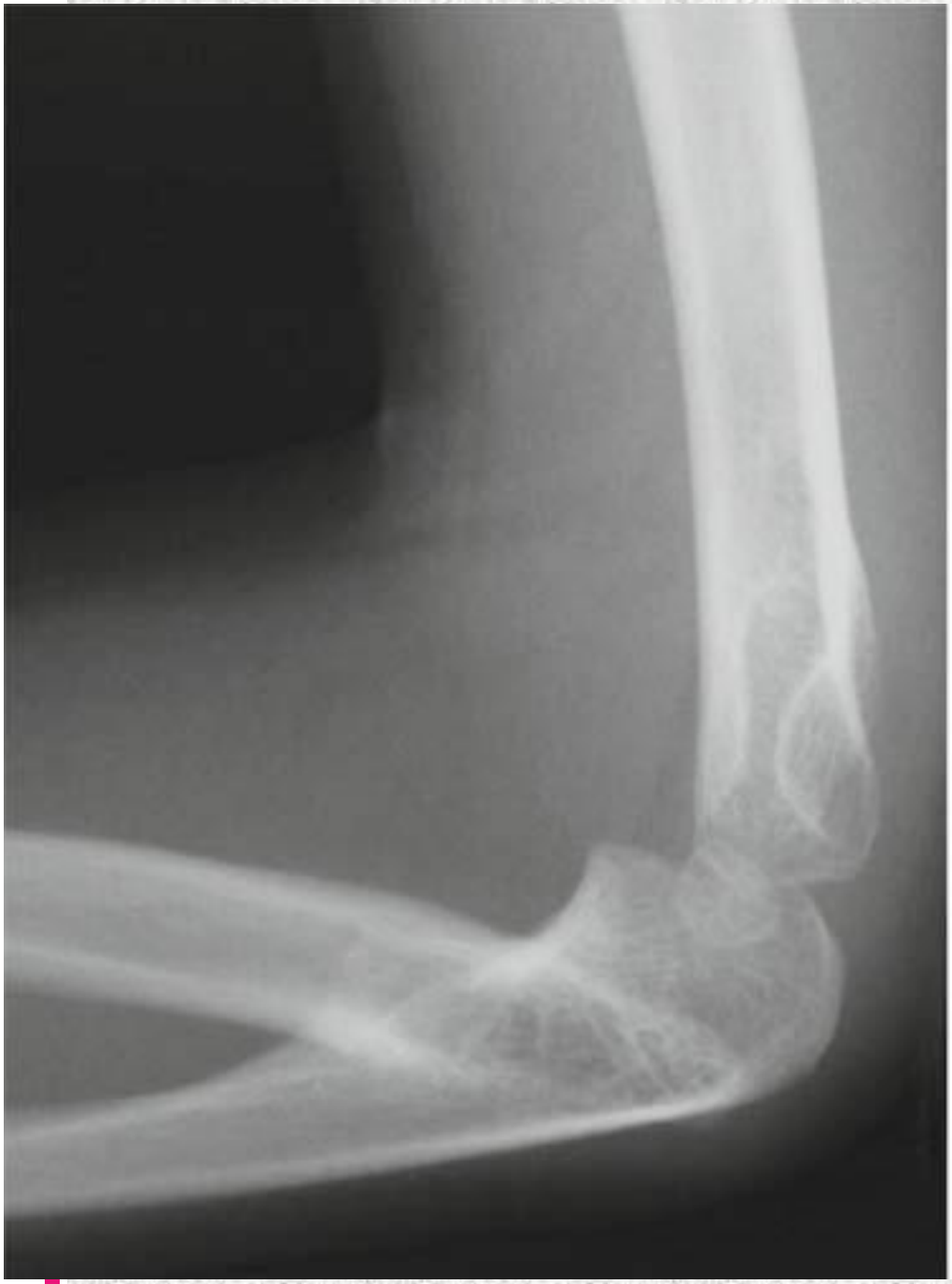
* Stretching
* Soft tissue release
* Bony

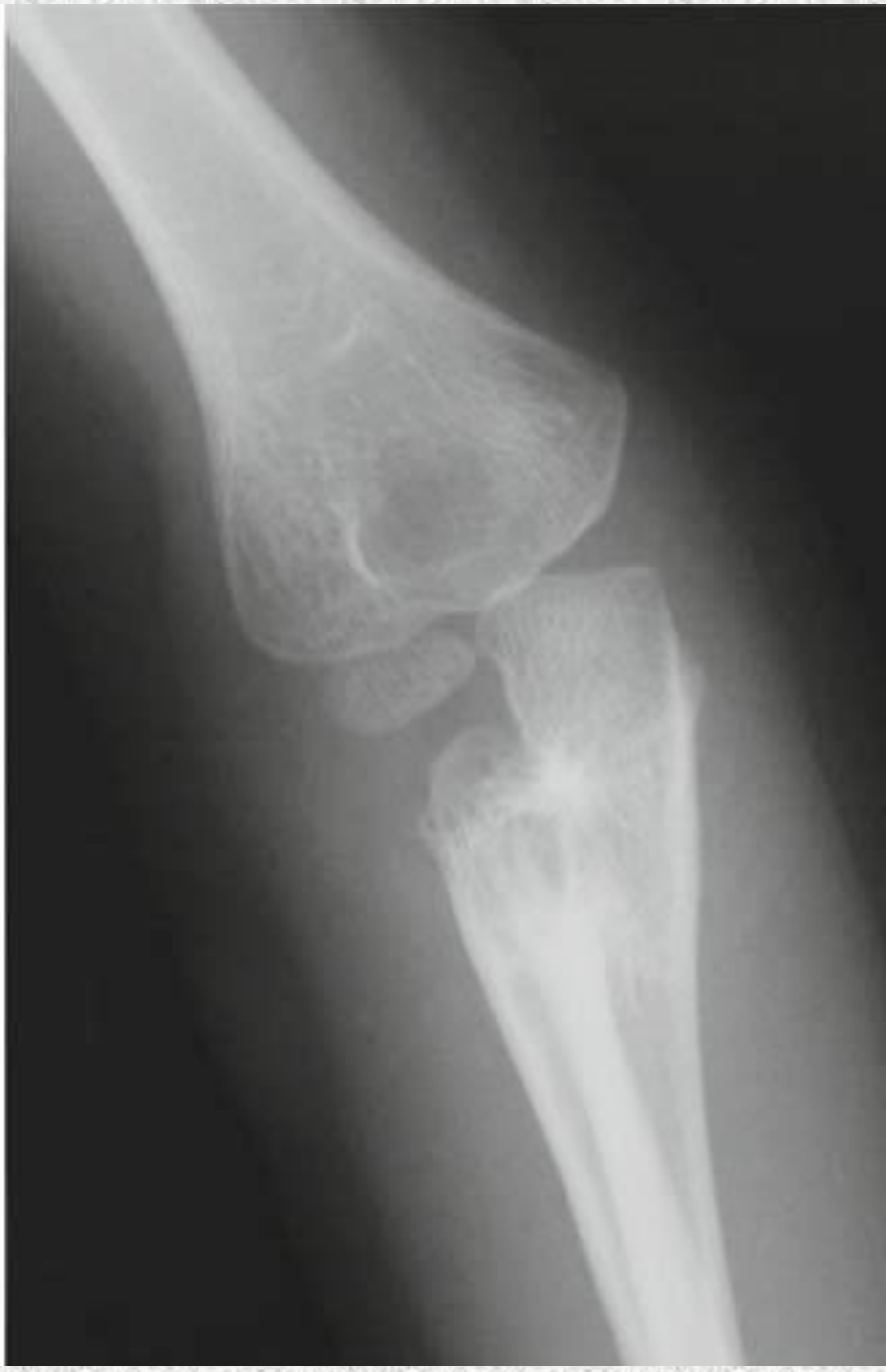
Kirner's

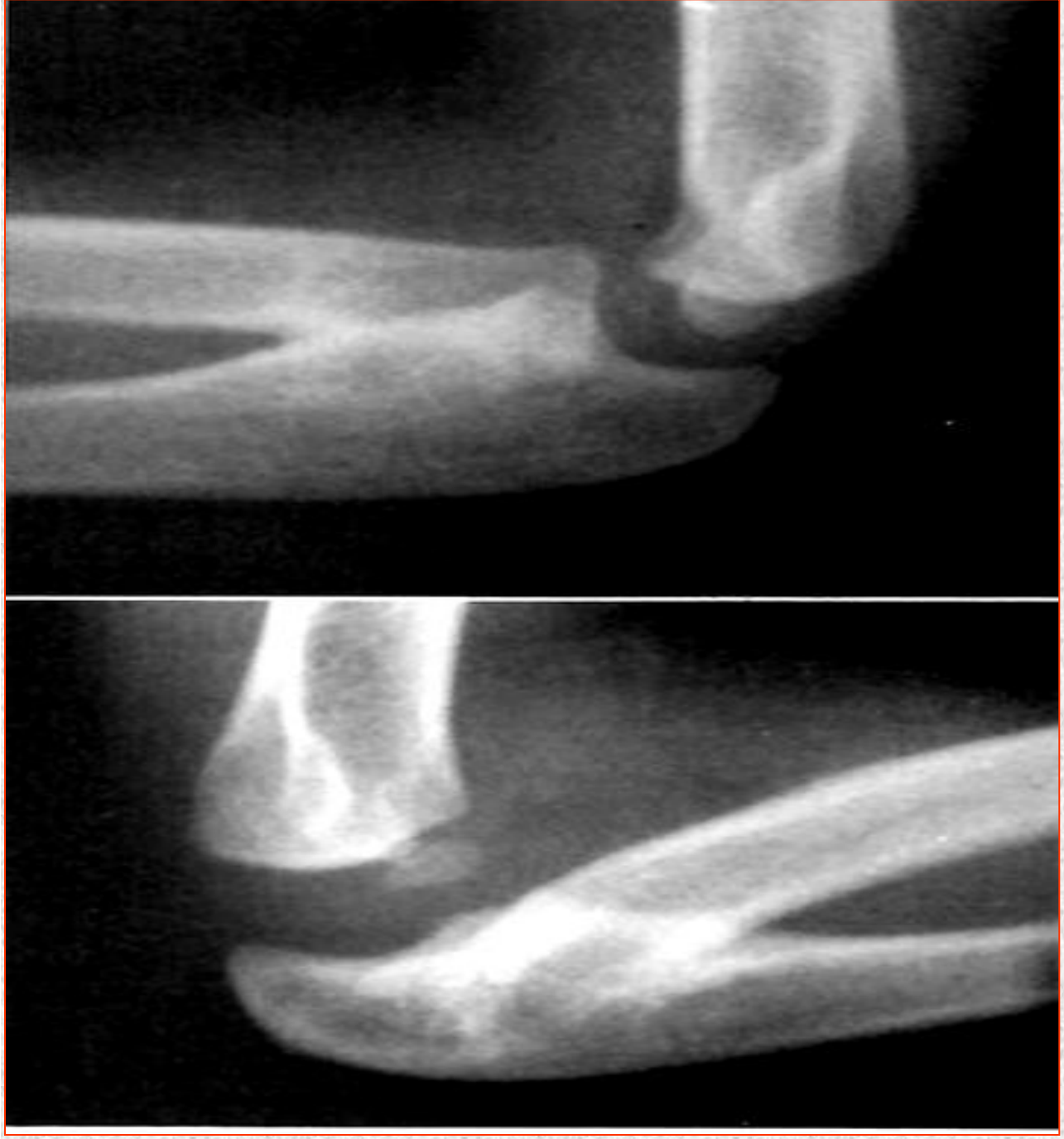




R U Synostosis







- Bilateral in 60% (most common in males).
- proximal 1/3 of forearm is most common site of bony union.

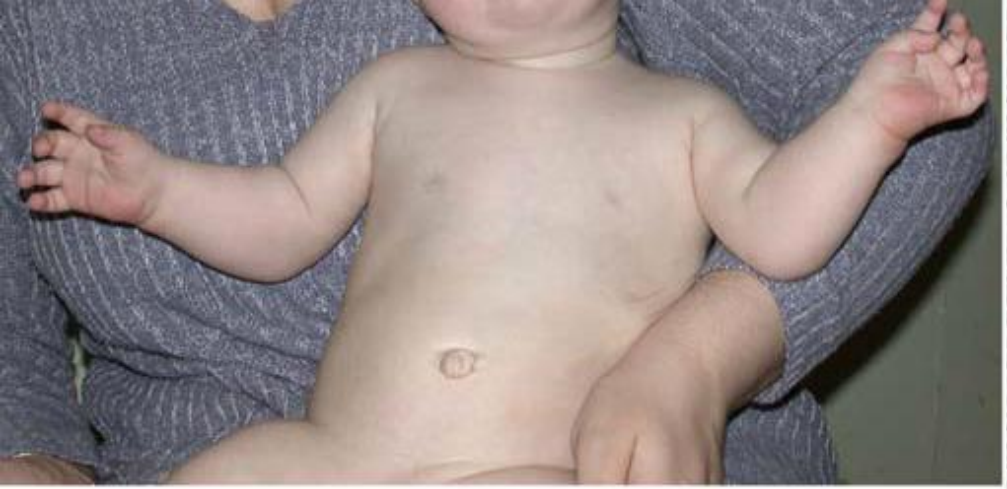
- **Non operative treatment:**
  - patients with a unilateral deformity or with a bilateral deformity & minimal pronation do not need surgery;
- **Surgical treatment:**
  - for severe pronation deformities (more 60 deg of pronation)
  - in pts w/ severe bilateral hyperpronation, osteotomy of nondominant extremity, to create supination, is indicated.
  - derotational osteotomy through the area of synostosis is recommended, placing one side in 10 to 20 degrees of pronation and the other forearm in a neutral position or slight supination for function;

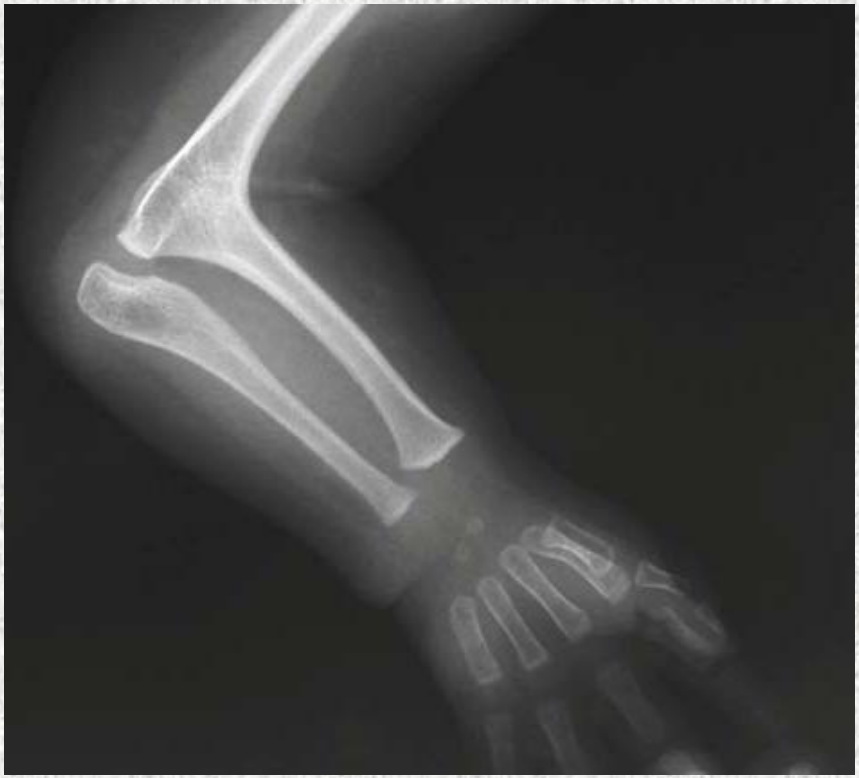
Congenital Radiohumeral Synostosis

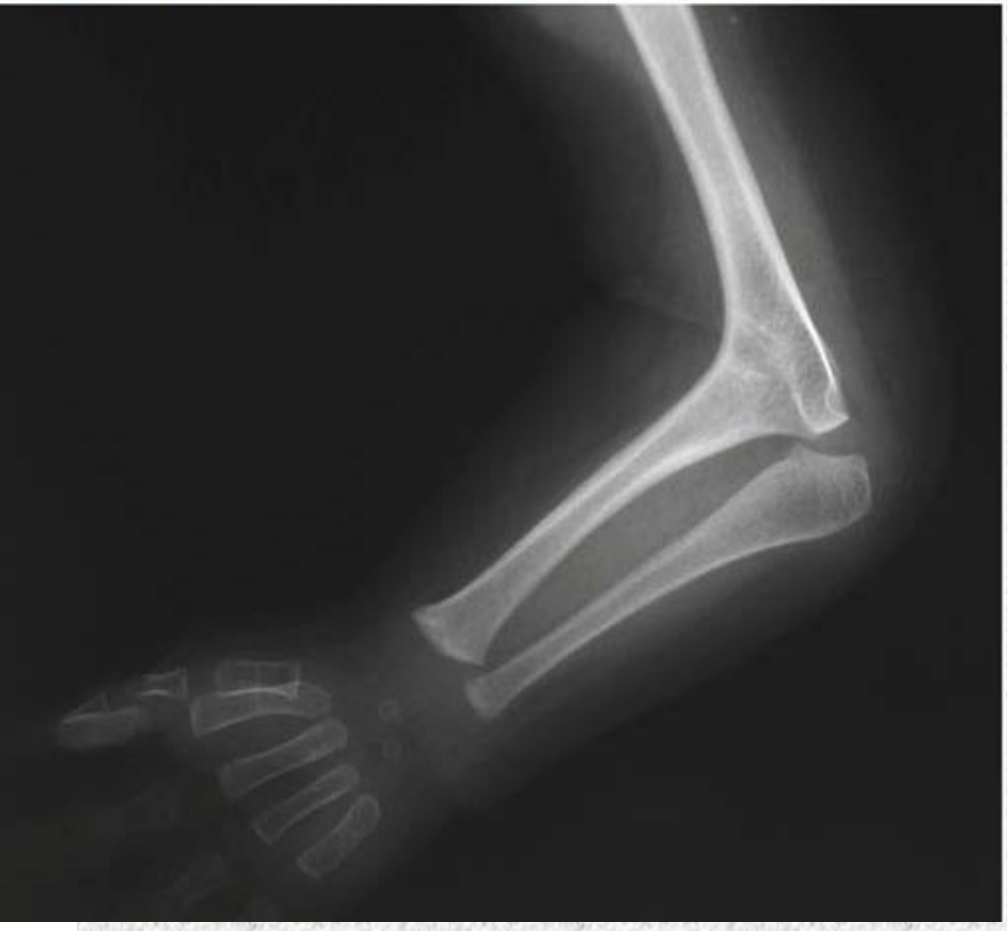
Synostosis may exist between the humerus and one of the forearm bones, most frequently of the radius.

Two third of the cases are unilateral

One third of the cases are associated with general skeletal abnormalities, such as hip dislocation; knee anomalies; clubfoot; polydactyly; syndactyly; Madelung deformity; thumb hypoplasia; carpal coalition; and problems of the cardiac, renal, neurological, and gastrointestinal systems

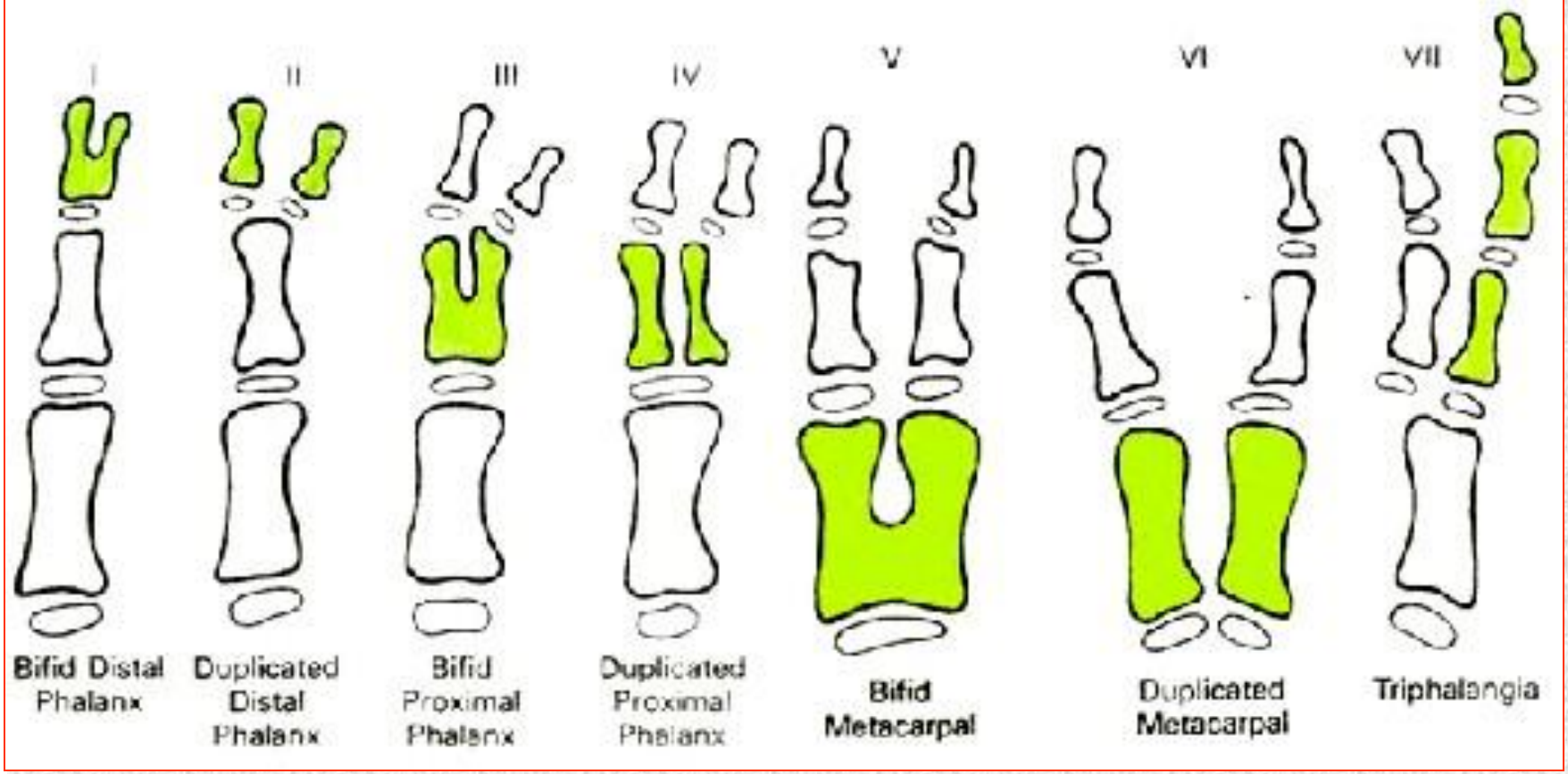




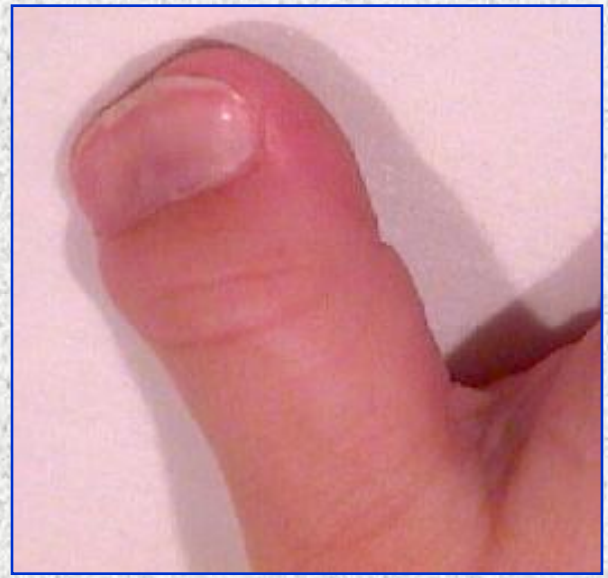


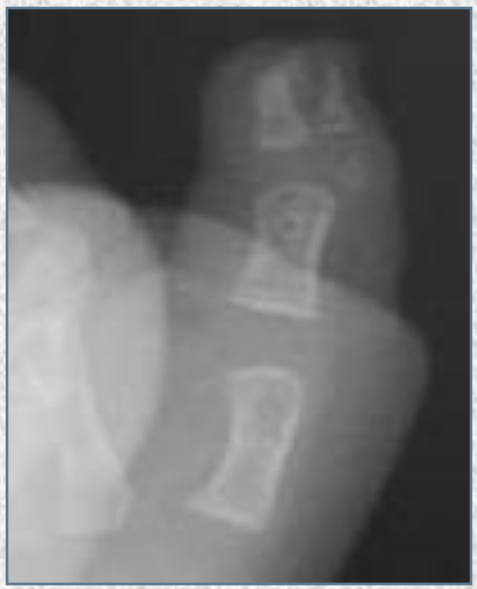
3-Duplication

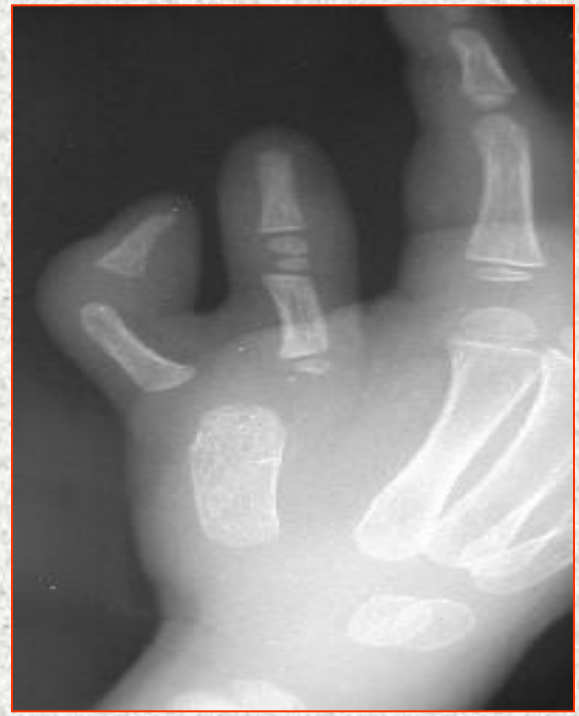
* Polydactyly

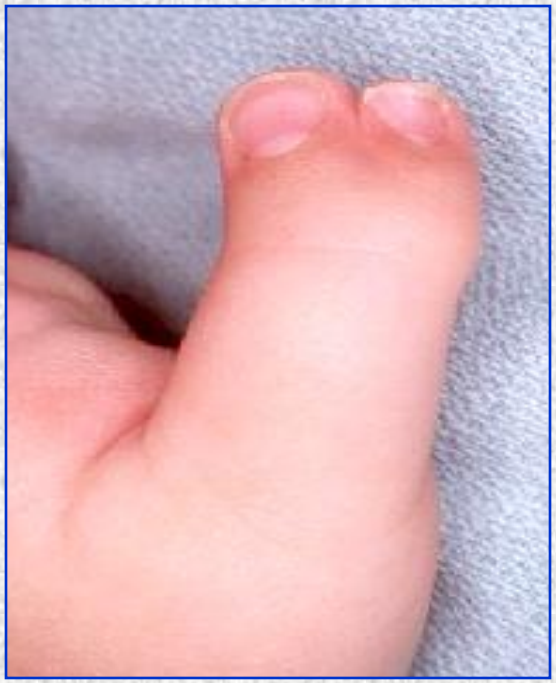


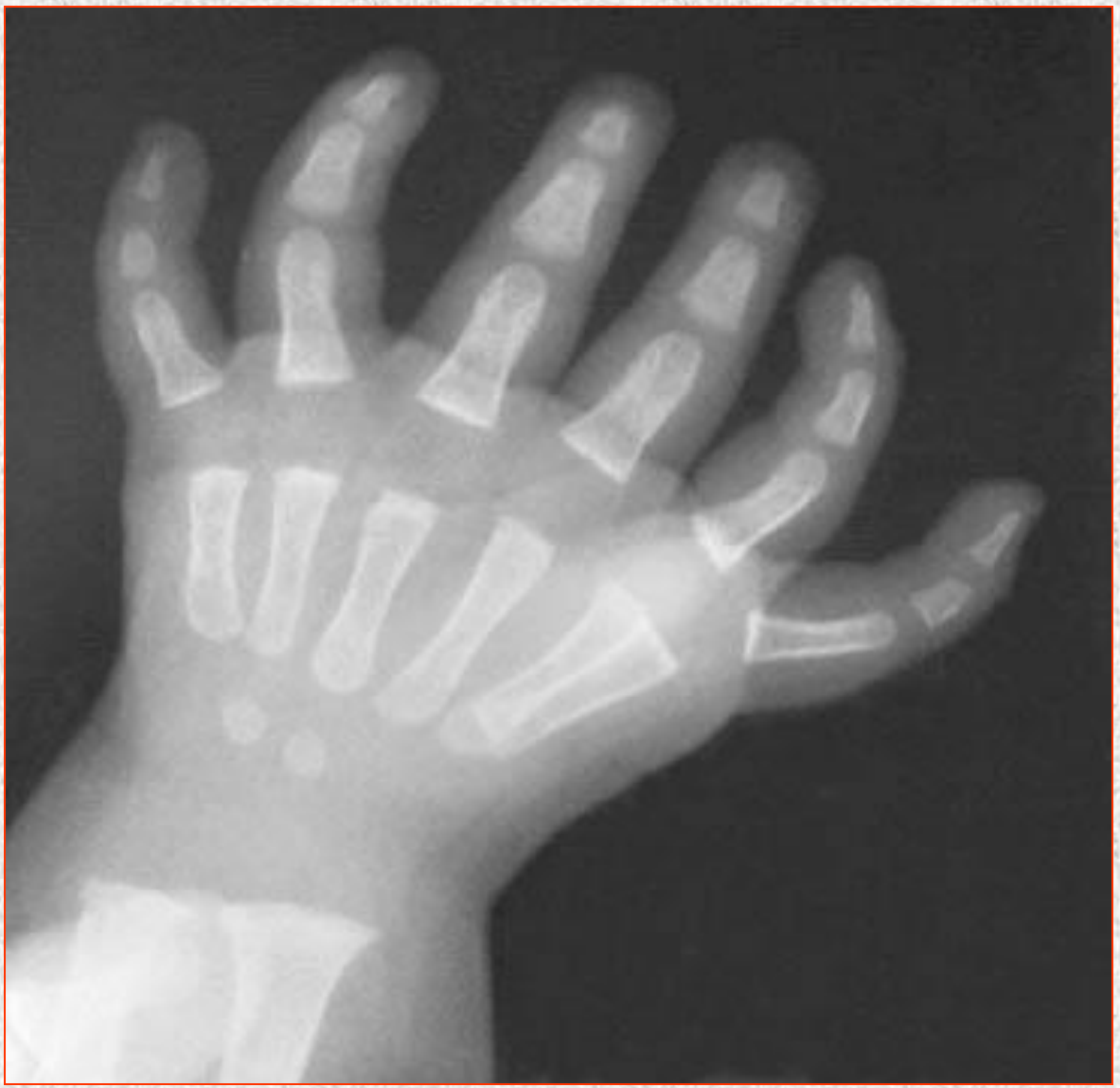
*Radial







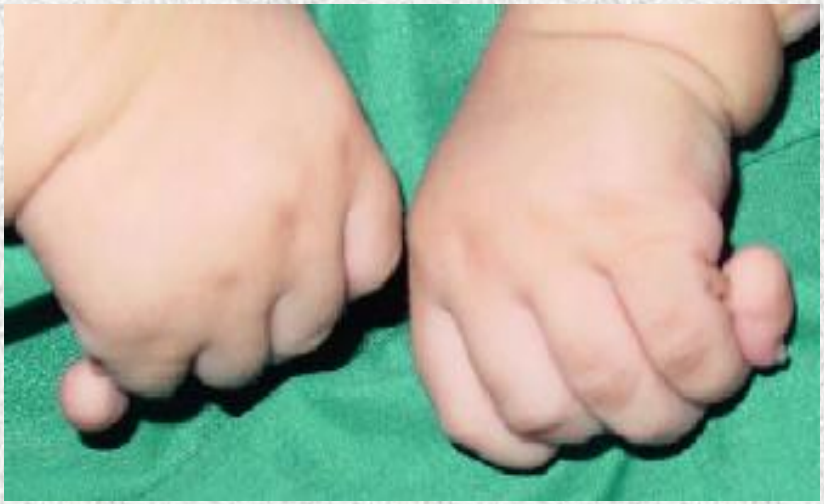




Ulnar Dimelia

Timing of surgery

* Nursery :
* 12 - 18 m.



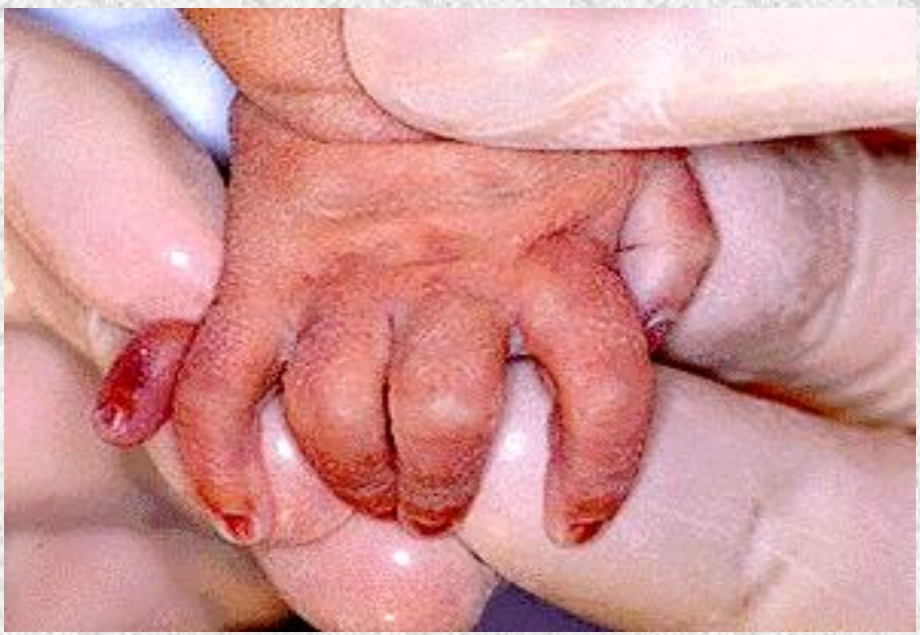
Management

1 + 2 : Wedge + Wiring

4 : Reconstsruction

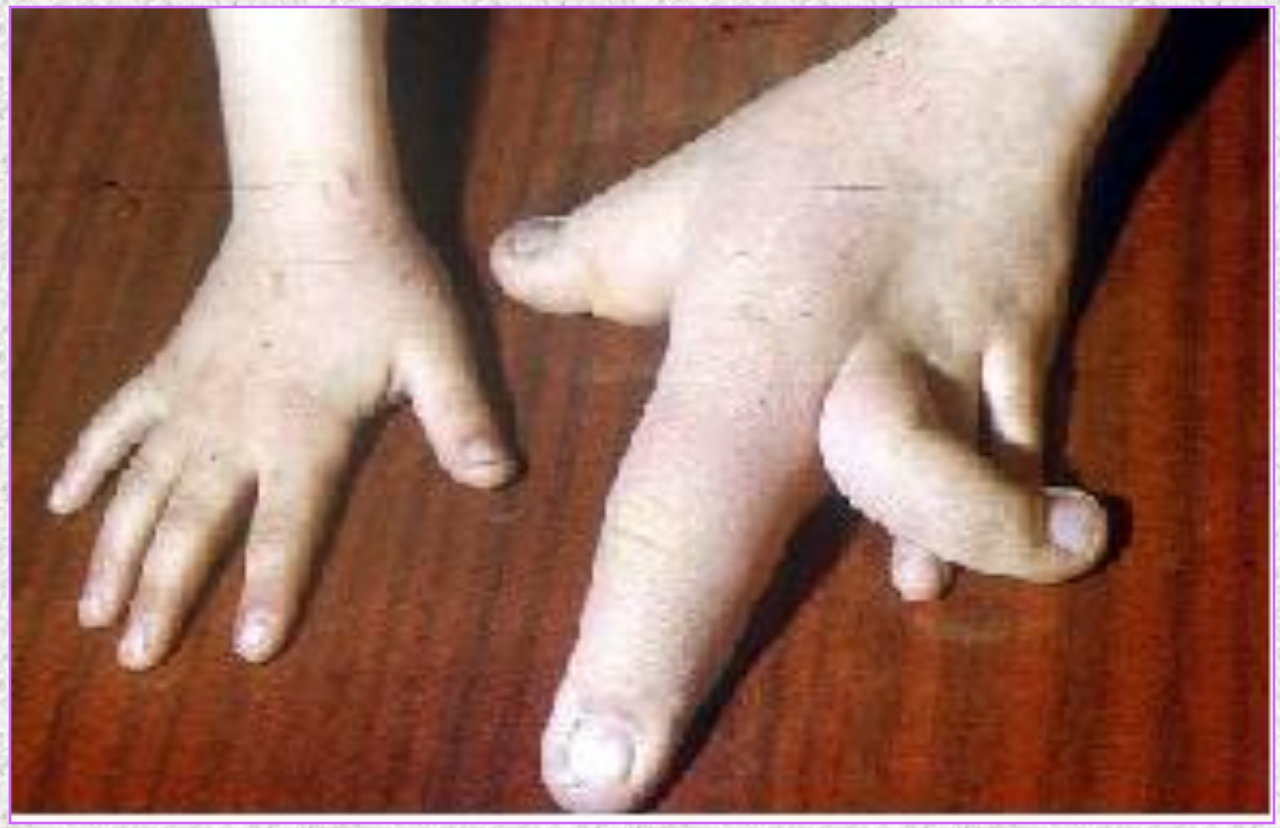
3 + 5 + 6 + 7: Amputation

Ulnar + Central



* Early surgery
* Reconstruction

4-Over growth



- A hamartomatous enlargement of soft tissue & underlying bone
- Enlargement includes skin, subcut tissue, nerve, joint, and bone (tendons and blood vessels are of normal size);
- most often phalanges are involved and metacarpals are spared

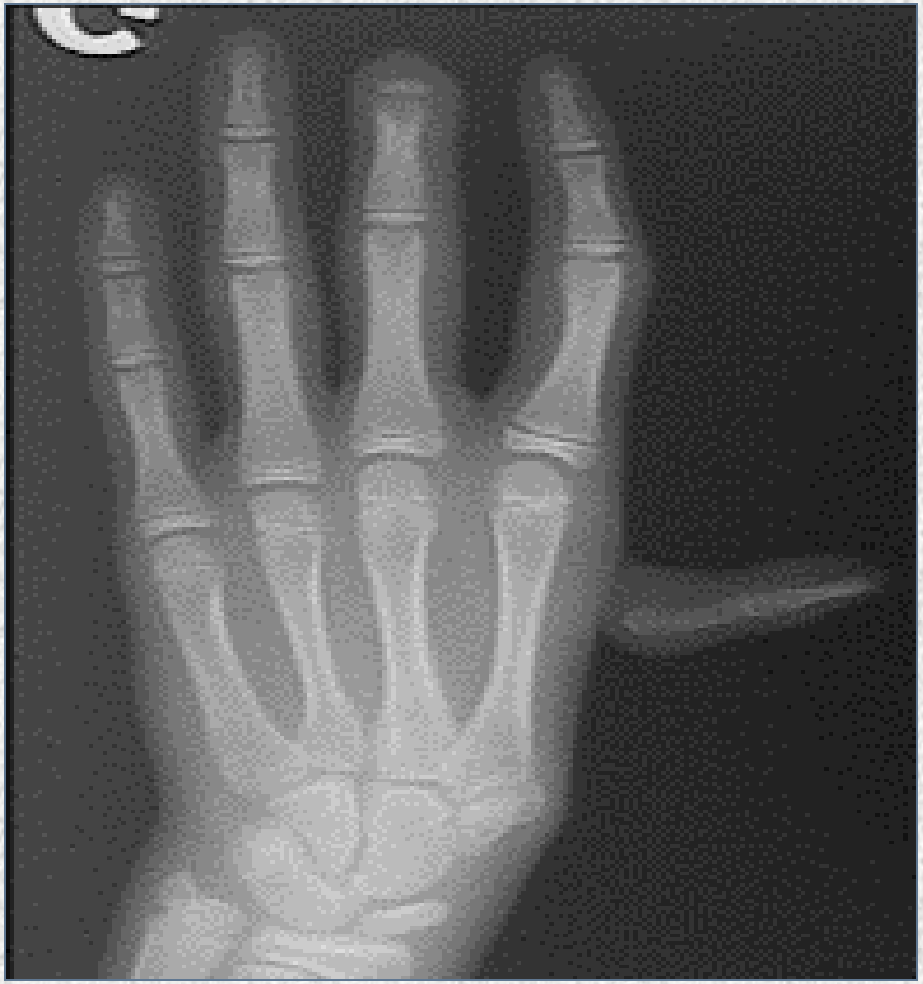
Associated conditions:

- Neurofibromatosis
- Lipofibromatosis

Treatment:

- Staged debulking, dealing with one side of digit or hand at a time, because blood supply to the skin of the enlarged digits is poor;
- **First stage**, defatting is performed on one side of digit, removing up to 20% of its thickness (convex side of digit is adressed first)
- **Second stage**, perform similar defatting procedure and consider bone shortening;
    - in severe cases, consider removal of an entire phalanx (digit must be stabilized with a K wire, extensor tendon shortened, and flexor tendon left alone)
- Appropriately timed epiphysiodesis of the involved bones is performed during growth
- alternatively, later bone resections and fusions can be carried out

5- Under growth



6-Cong. Constriction band synd.

= Annular band → focal necrosis

= Aetiology !Placental membrane

= Effects

@ intrauterine gangrene

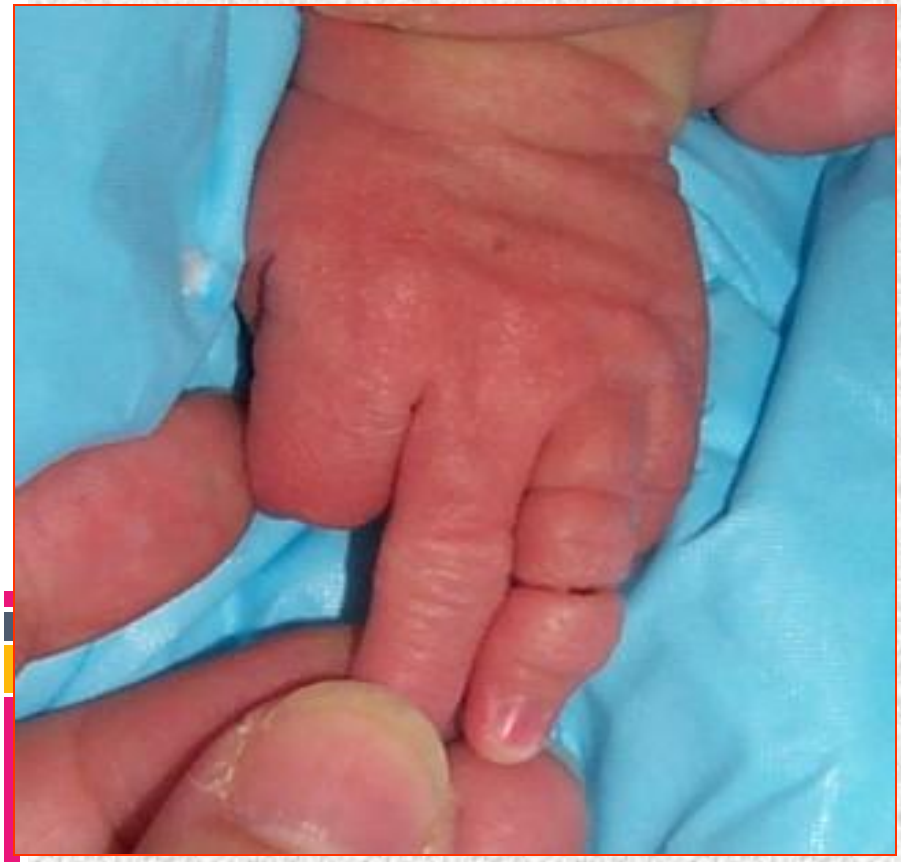
@ amputation

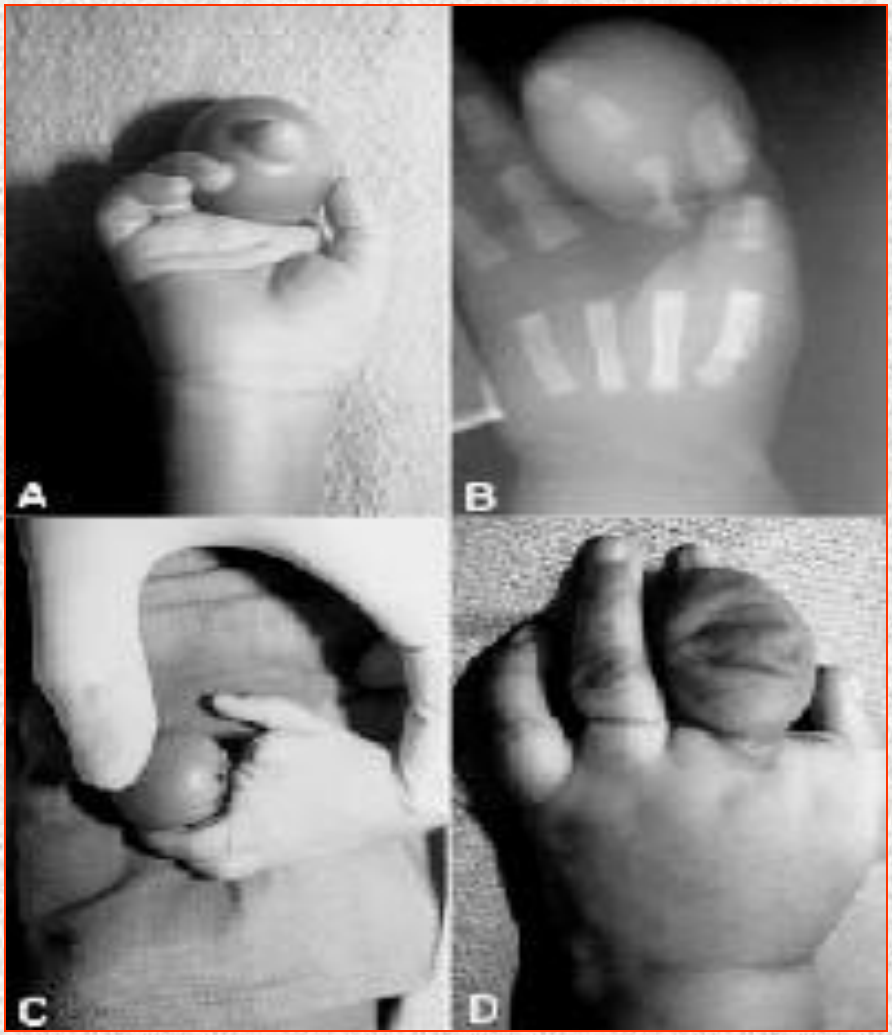
Types

I- Skin

II-Impending gangrene

III- Skin + Subcut.




A
B
C
D

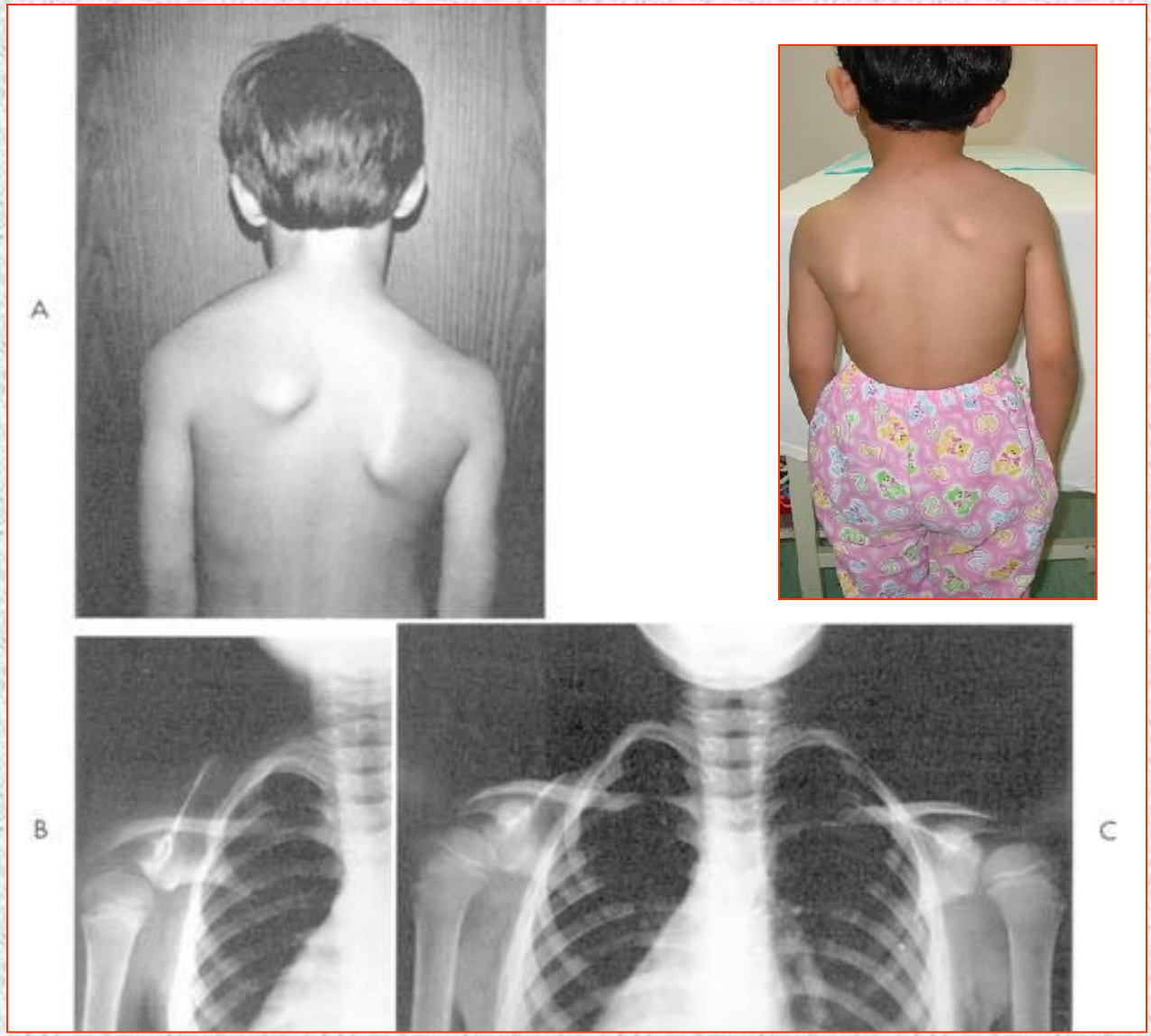
Management

* **Type II: Urgent**
  Staged Z plasty
* **Type III: 1 year**
* **Type I : Before school.**

Trigger Thumb

* 1 - 2 years.

Sprengel's deformity



- Deformity is characterized by elevation & medial rotation of inferior scapula
  - involved scapula is both smaller and more cephalad than normal;
  - in 30% of pts, the scapula is attached to the cervical spine by an omovertebral bone, cartilage, or fibrous tissue, which, when present, can severely limit scapulothoracic motion;

Associated anomalies:

congenital scoliosis, cervical ribs, torticollis, renal abnormalities, & muscular hypoplasia, especially involving the trapezius;

Surgical Treatment:

-Timing:

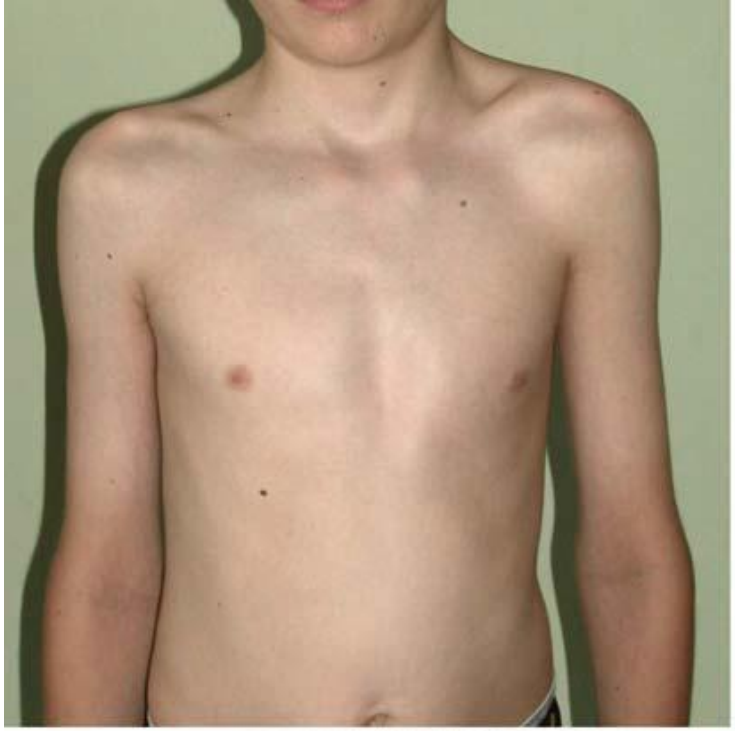
- Between 3-8 yrs of age with significant deformities, both functional and cosmetic;

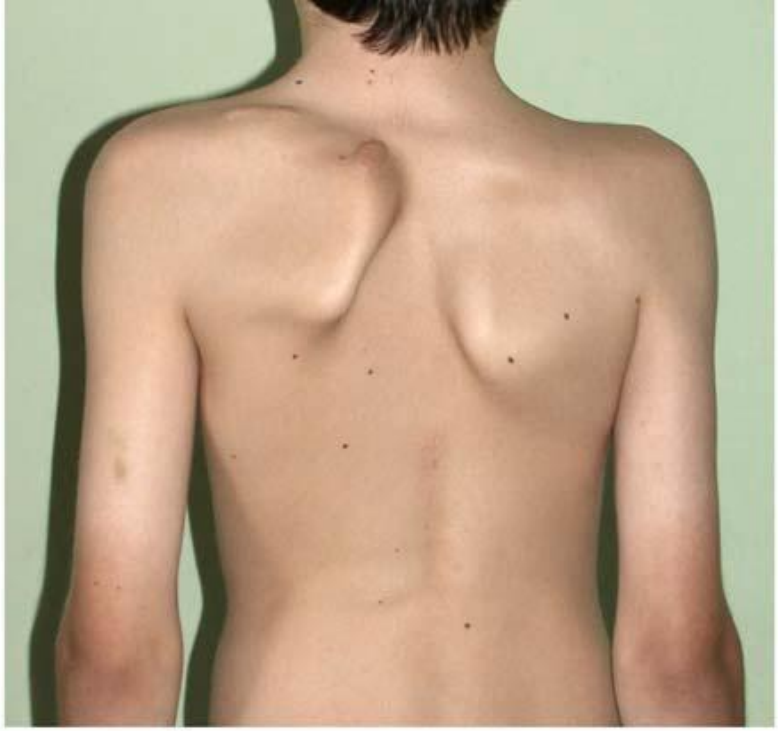
- Options:

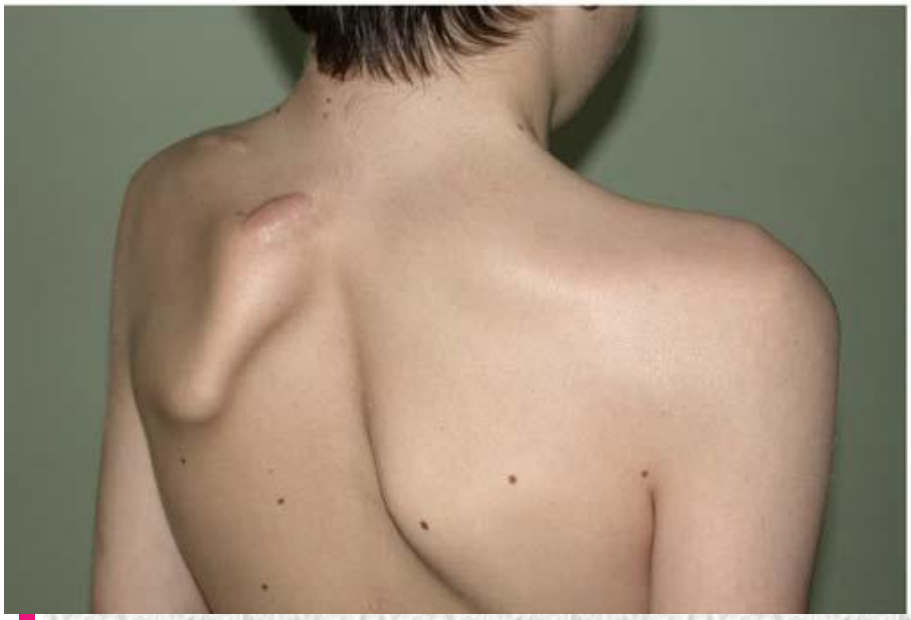
- detachment of medial & superior scapular muscles, repositioning scapula caudad, & subsequently reattaching the muscles to lowered scapula.

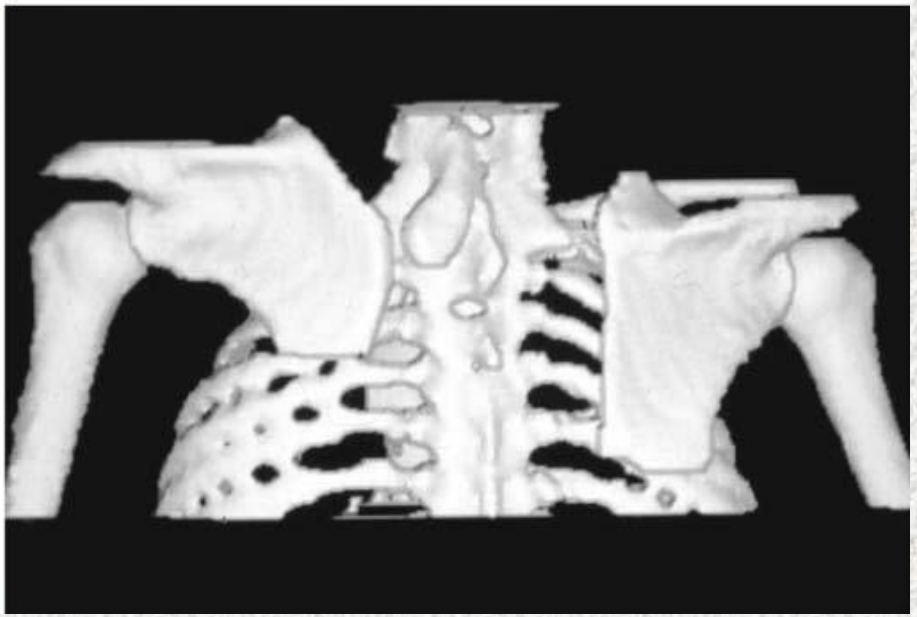
Woodward Procedure:

- procedure has 80% satisfactory functional and cosmetic results.

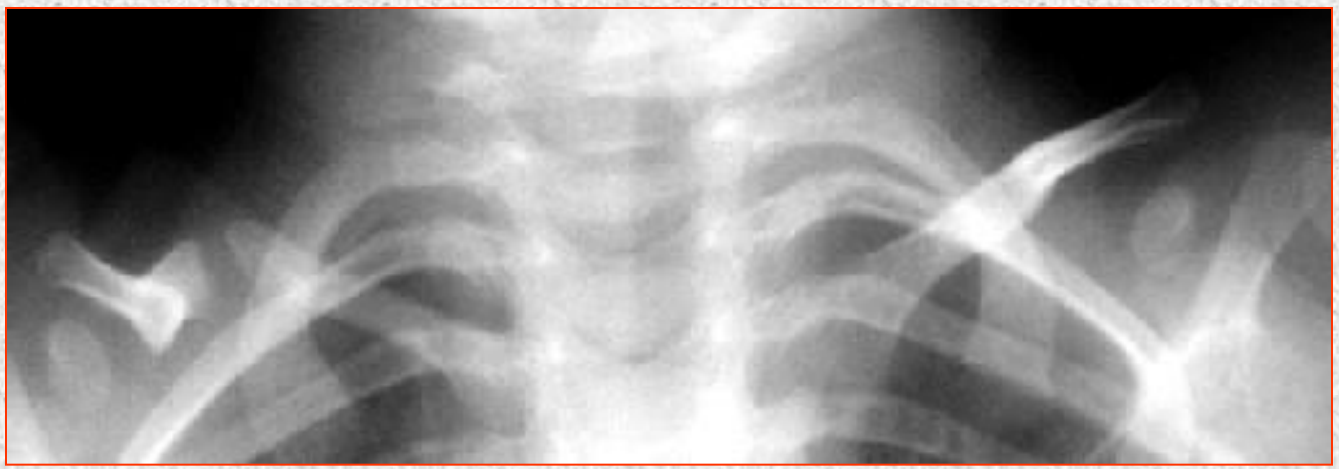


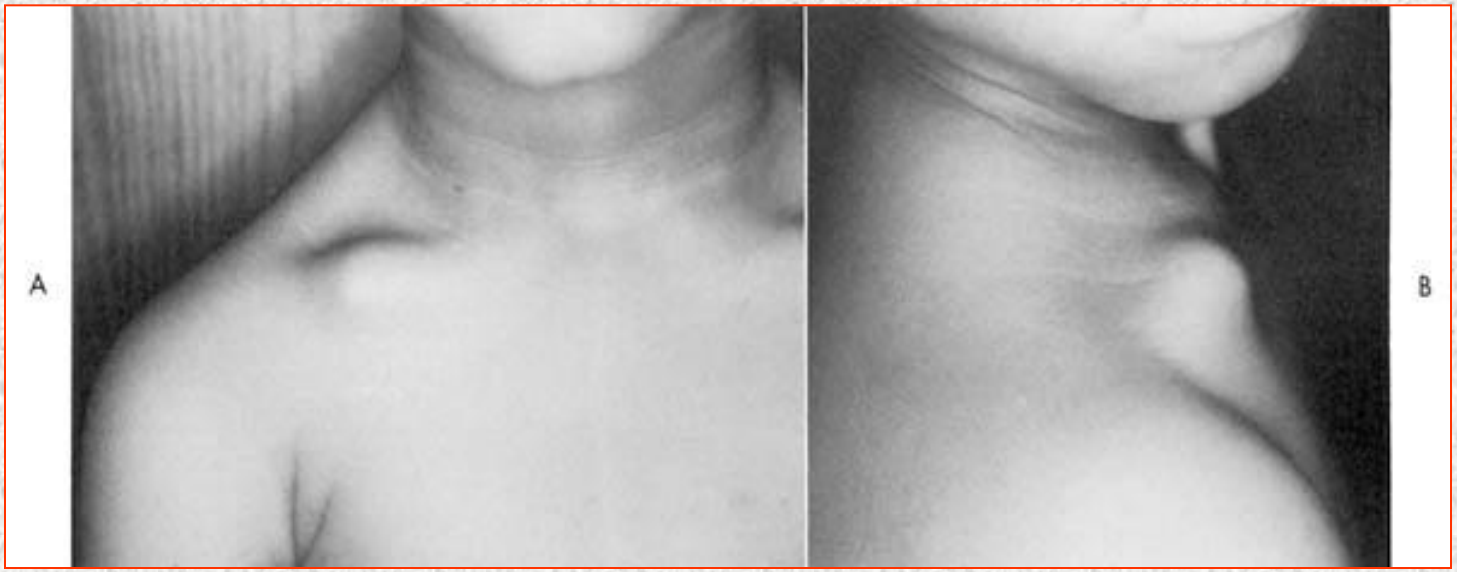






Congenital Pseudoarthrosis





Epidemiology

- almost always on right side with the exception of situs inversus
- bilateral in < 10%
- Pathophysiology

may be related to pulsation of underlying subclavian artery

- Usually asymptomatic
- Physical exam

painless, enlarging, nontender mass on right clavicle

Nonoperative

observation

minimal symptoms and cosmetic deformity

Operative

ORIF with bone grafting at age 3-6 years

indications

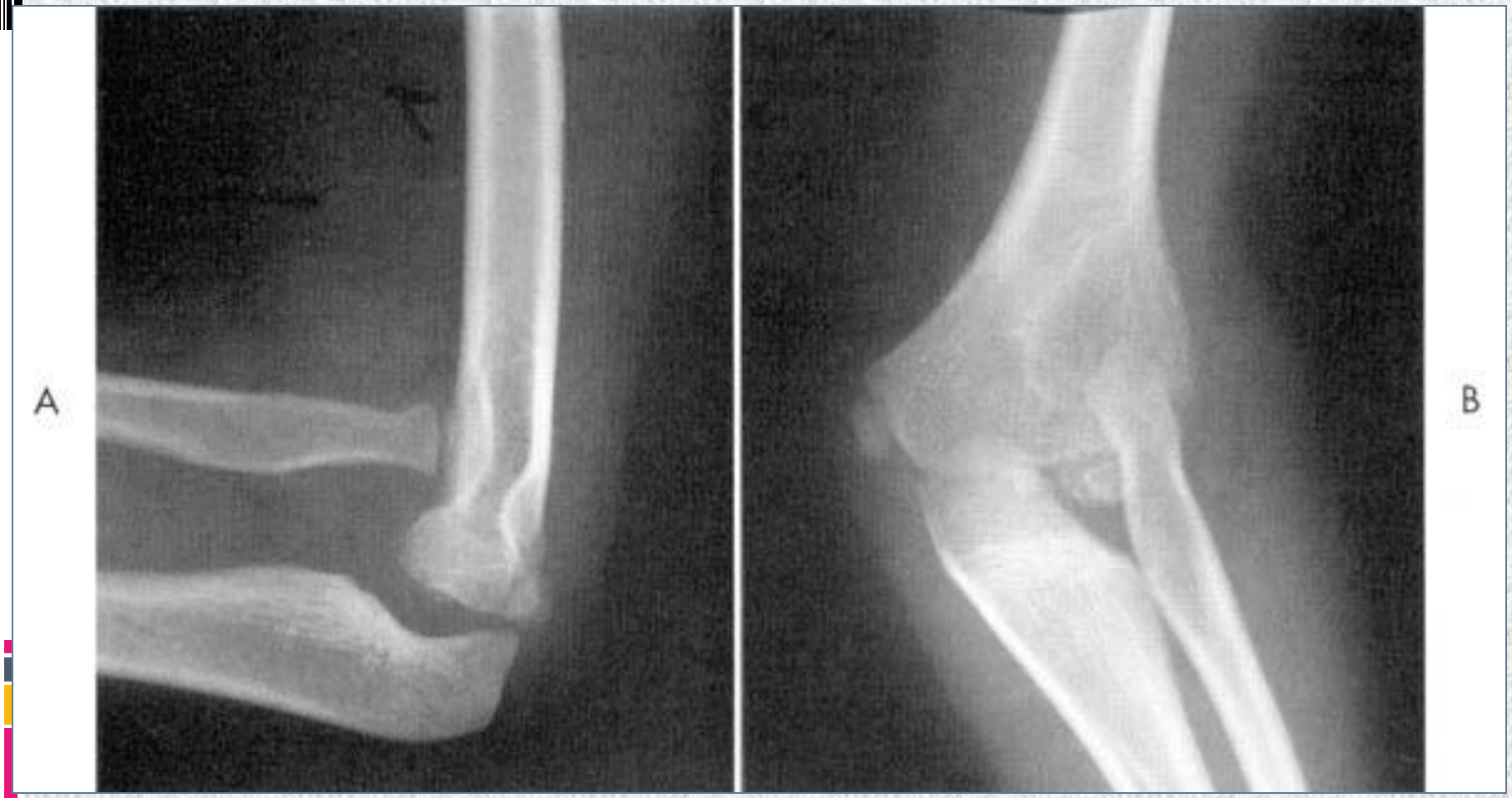
functional impairment

= cosmetic concerns

outcomes

successful union is usually obtained

Congenital radial head dislocation



It is often associated with syndromes such as nail patella syndrome, and may in some cases by genetically transmitted.

When unaccompanied by other radial or systemic conditions, it is almost always bilateral.

Radial shortening often accompanies congenital dislocation of the radial head.

It is often not noted until age 4 or 5 at which time some limitation of motion or deformity becomes evident.

Most are posterior, about 1/ 3 are anterior or lateral.

The natural history is relatively benign

The favored treatment approach at present is to defer any intervention until skeletal maturity, at which time resection of the radial head can improve appearance, but not motion.

Resection of the radial head in a child results in regrowth of the head.

Congenital Muscular Torticollis

Torticollis (wry neck) is a congenital or acquired condition of limited neck motion in which the child holds the head to one side with the chin pointing to the opposite side.

It is the result of shortening of the sterno-cleidomastoid muscle.

= In most cases, the shortening is a consequence of injury during birth.

= In early infancy, a firm, nontender mass may be felt in the mid portion of the muscle.

= The mass usually disappears and is replaced with fibrous tissue.

= If untreated, there can be permanent limitation of neck movement.

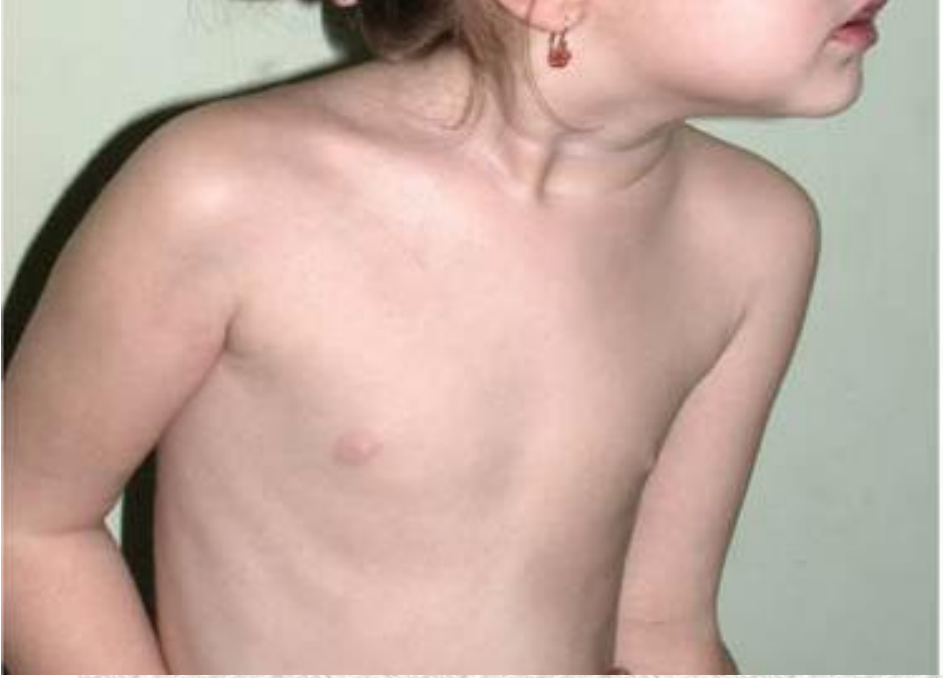
= There may be flattening of the head and face on the affected side

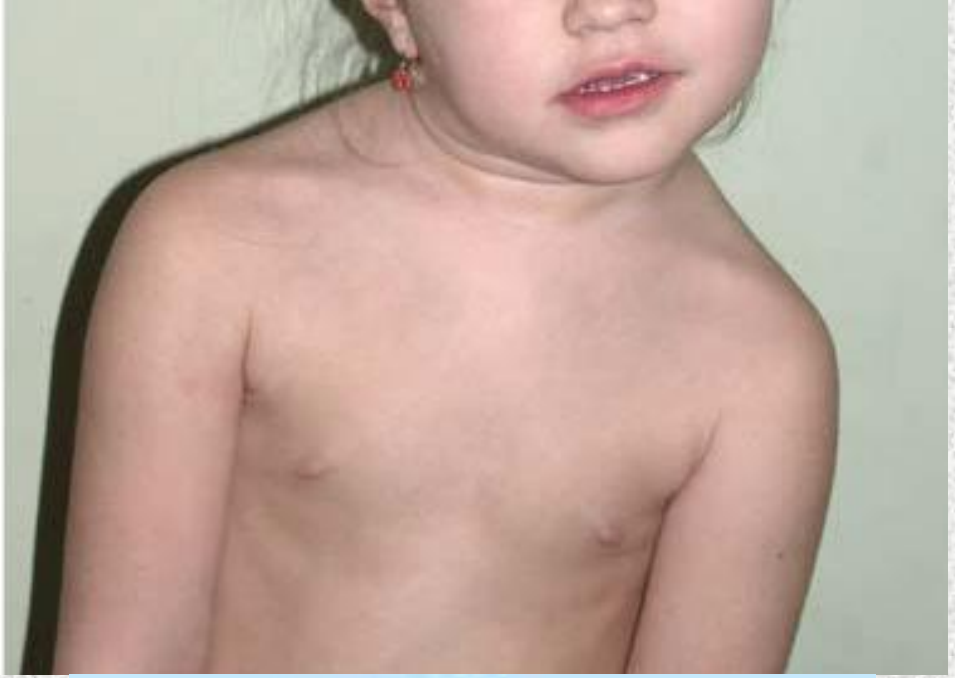
Causes

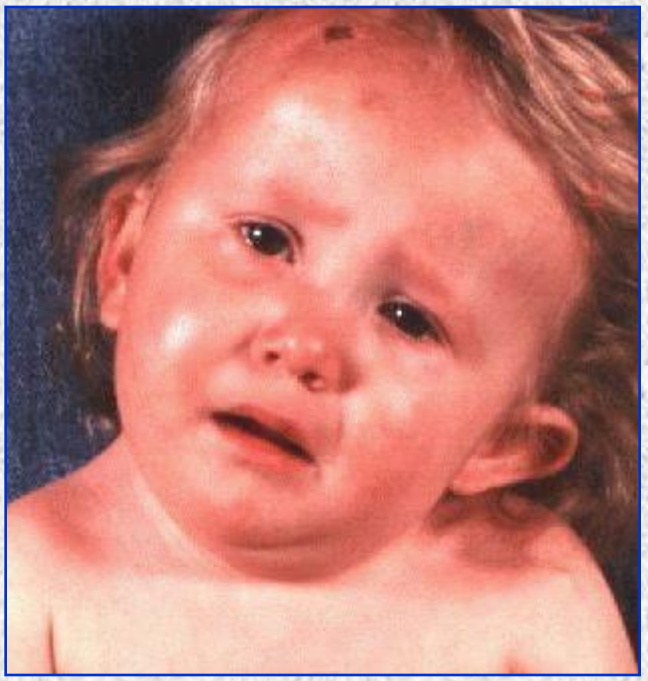
* **Trauma** to the neck or spine can lead to torticollis.
* **C1-C2 subluxation**
* **infection** of the

= head or neck secondary to inflamed glands and LN in the neck.
= abscesses of the throat and upper airway,
= infections of the sinuses, ears, mastoids, jaw, teeth.

***Eye problems** e.g Squint








SCM

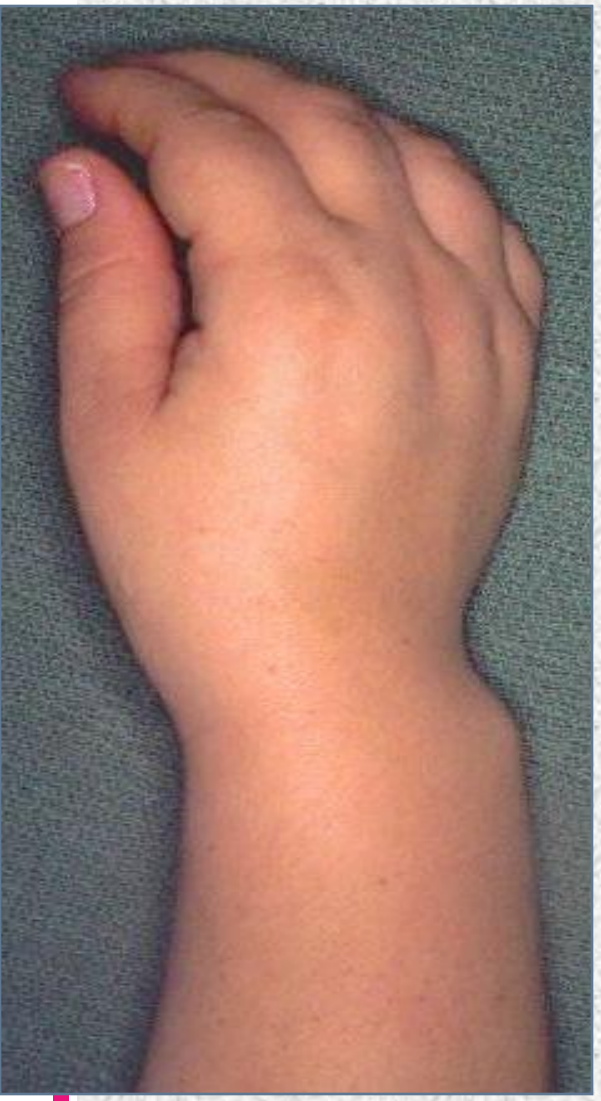
7-Generalised Skeletal abn.

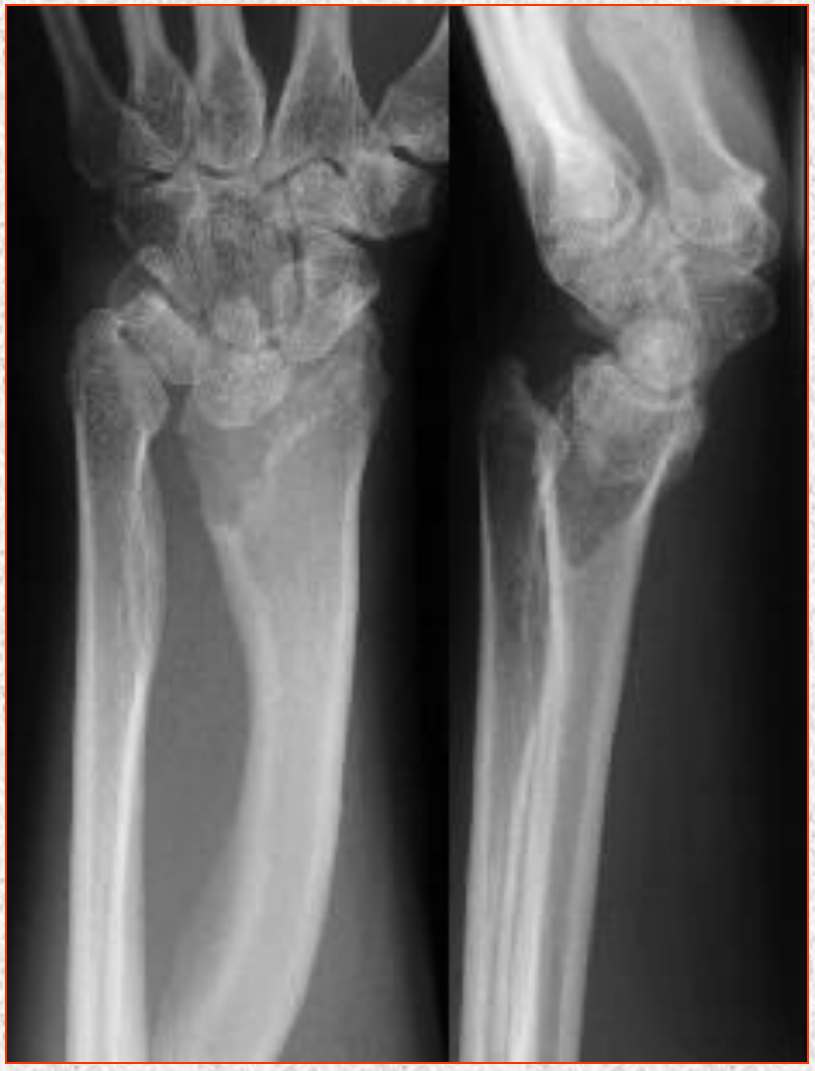
= Achondroplasia

= Marfan's

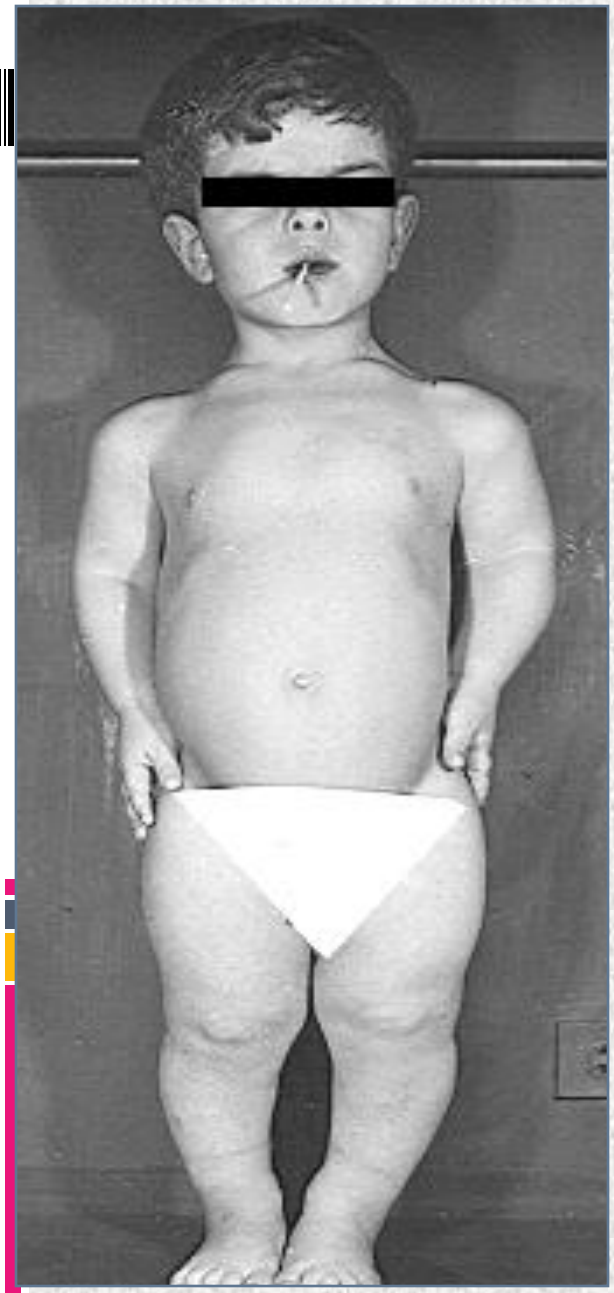
= Apert's

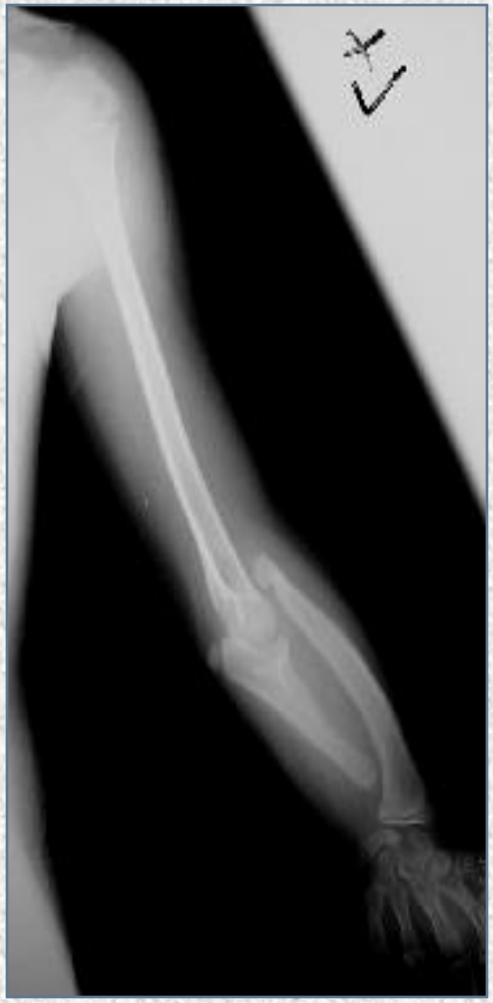
Madelung's deformity

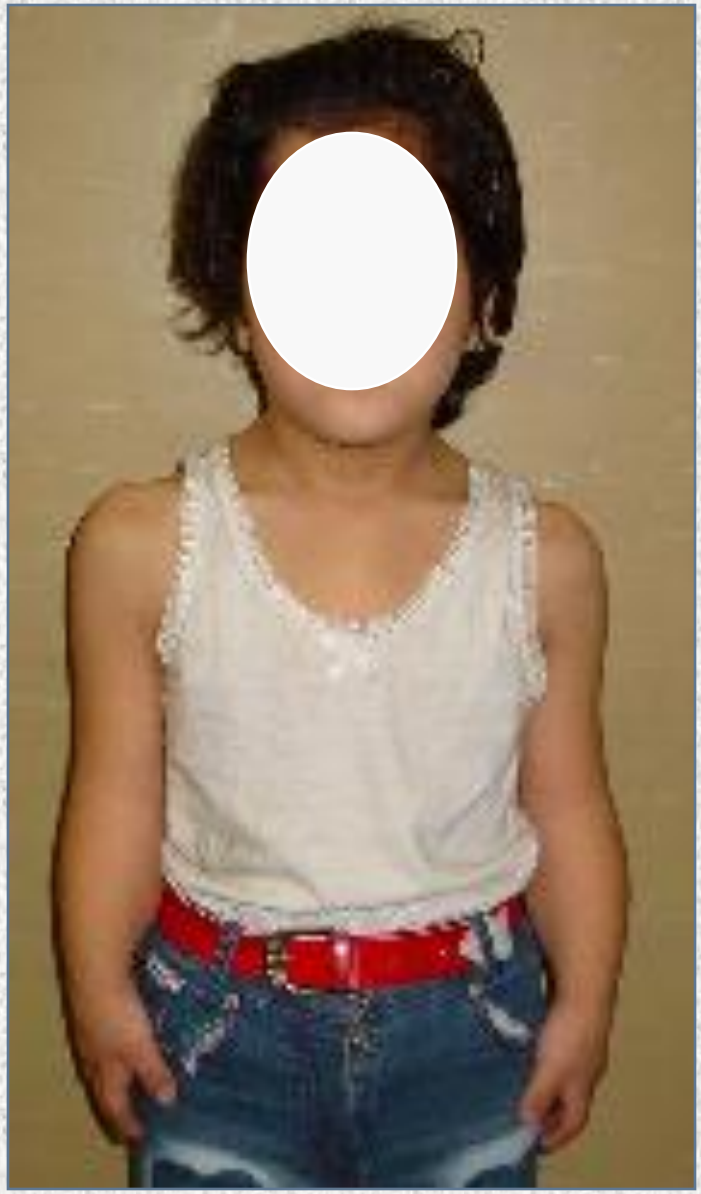




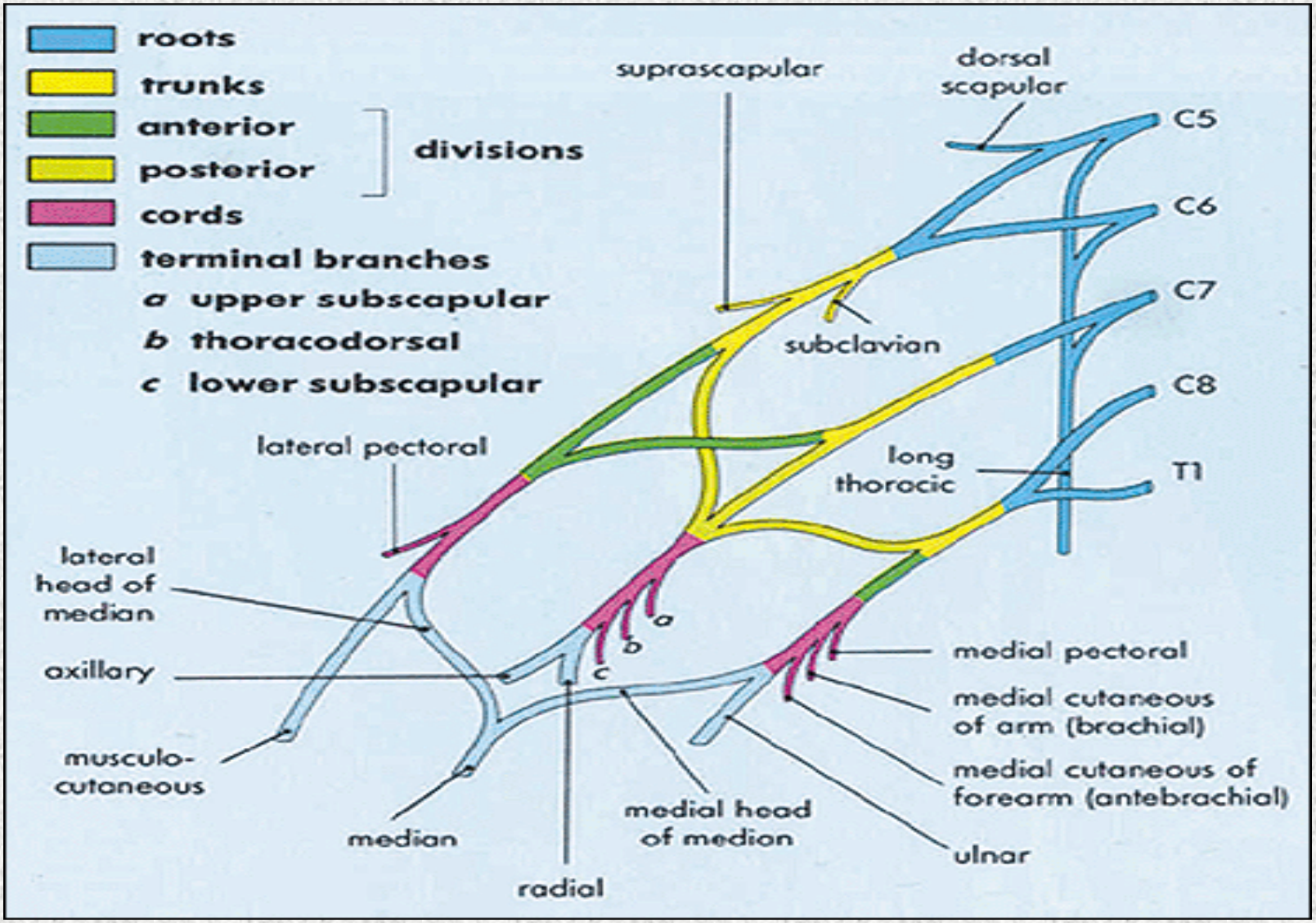
Achondroplasia







Brachial plexus injury



Obstetrical factors

* **Cephalic : Overweight, 88%.**
* **Breech : Small baby.**
* **Pressure neuropathy in uterus.**

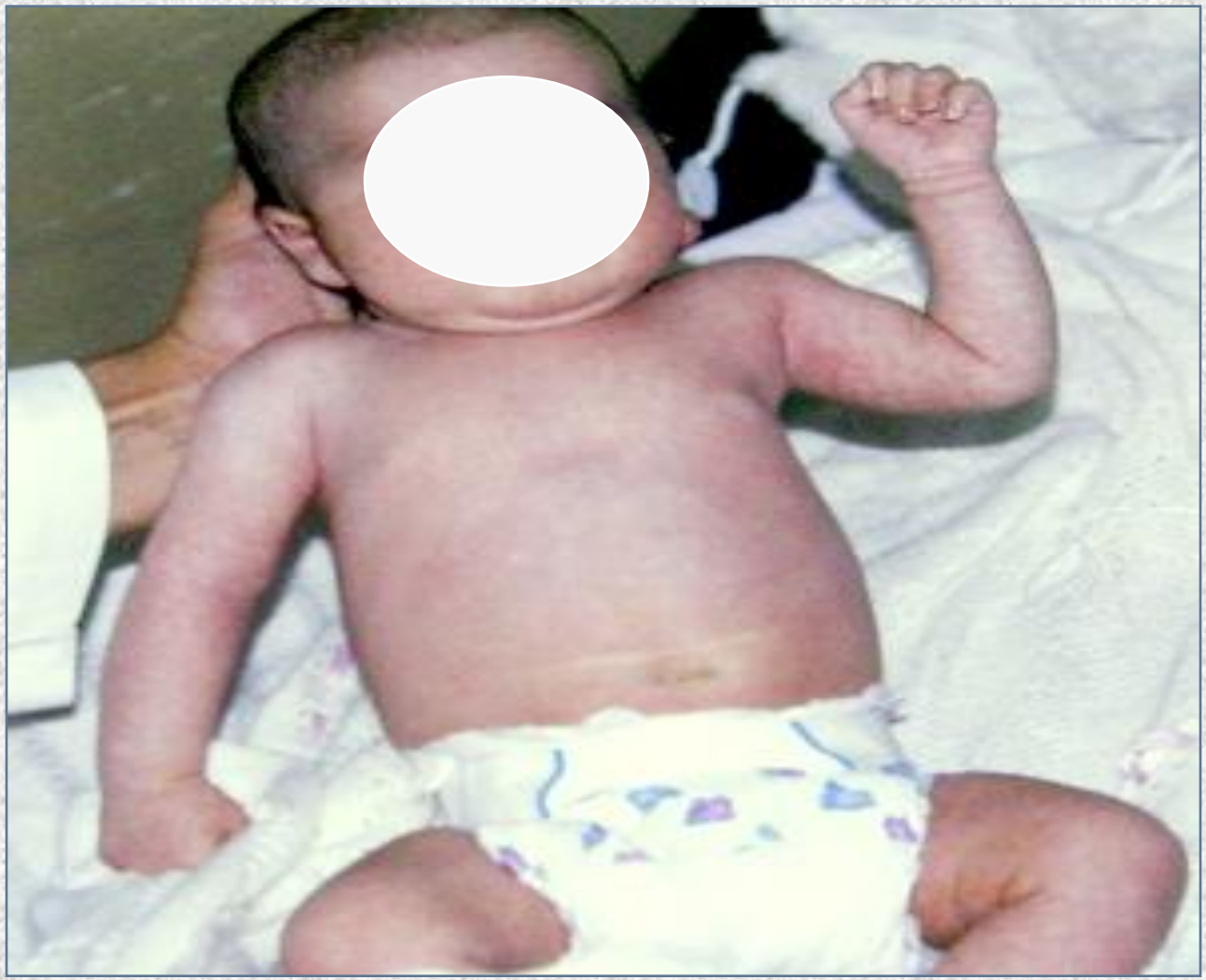
Incidence

***4:1000 in poor Obstetrics.**

0.1 - 0.3 :1000 in good care.

***1% bilateral.**

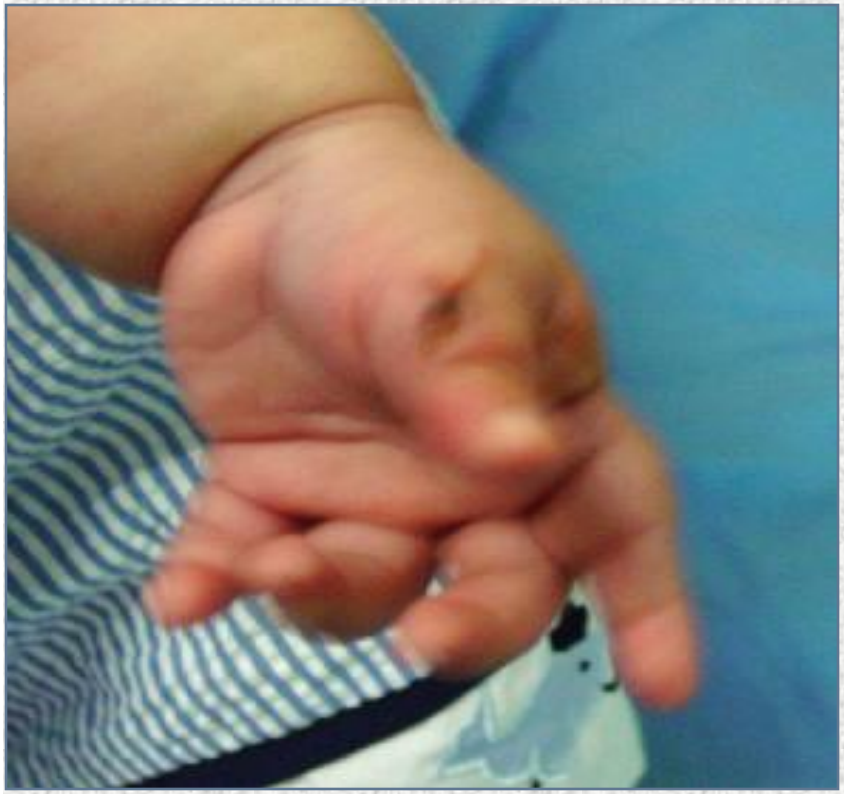
Clinical Picture

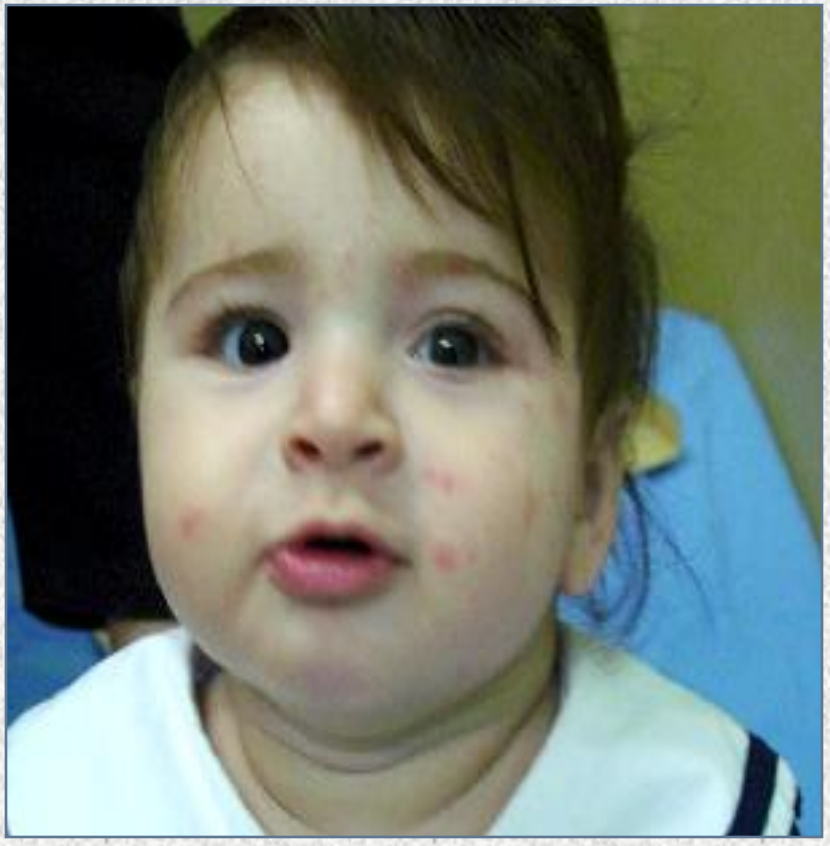


Clinical Types

* **Upper roots.**
  * **Elbow extended : C5-6.**
  * **Elbow flexed : C5-6-7.**
* **Complete paralysis**
  * **Flial arm with clinched hand.**
  * **? Horner's syndrome.**
  * **? Medullary lesion.**

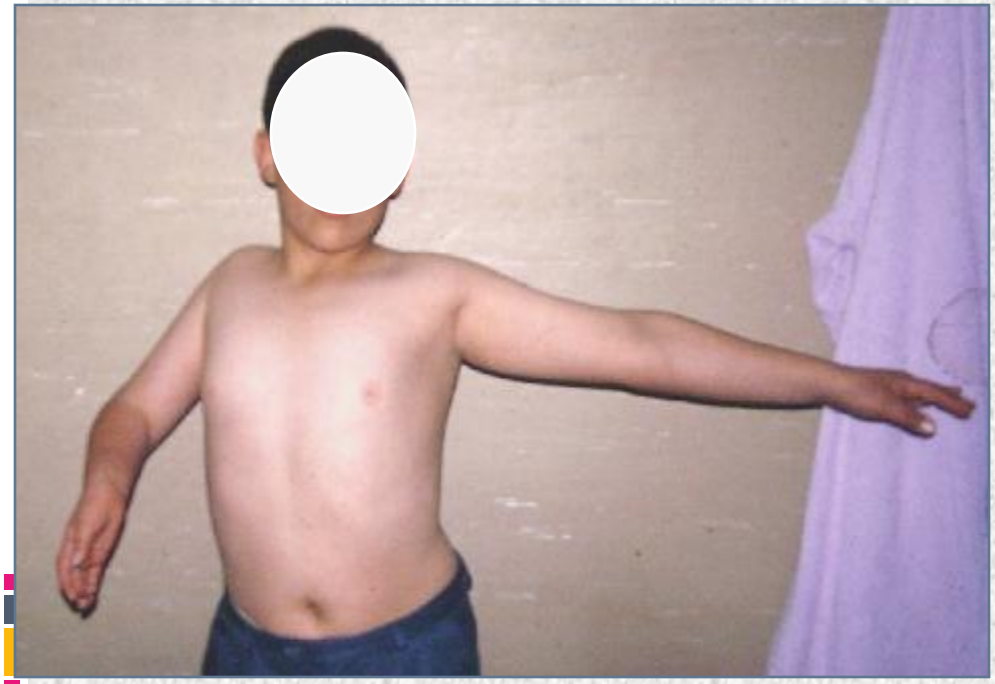
Complete paralysis



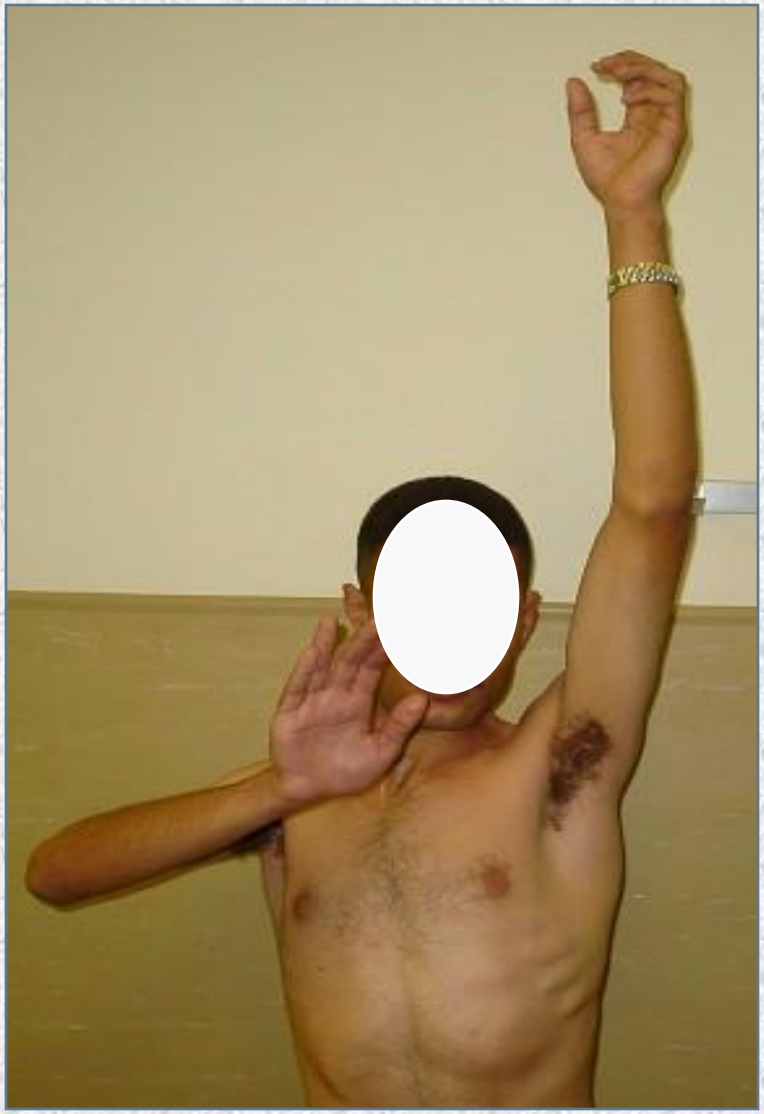


Horner’s

Complete paralysis



Upper roots

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