Congenital anomalies of lower limb- 2

DR RAHUL UPADHYAY

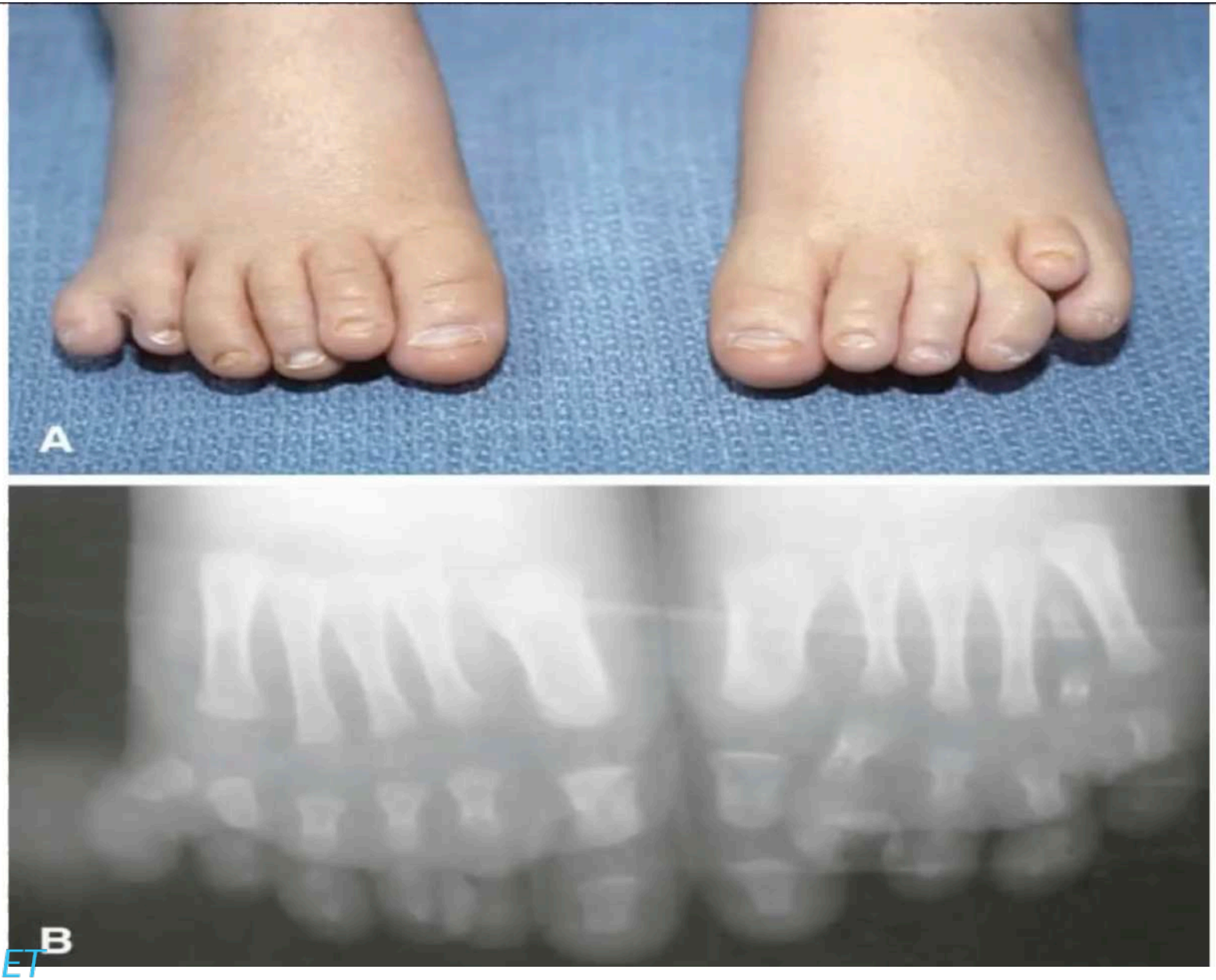
CONSULTANT ORTHOPAEDICS AND FOOT & ANKLE SURGEON

RAJASTHAN HOSPITAL, JAIPUR

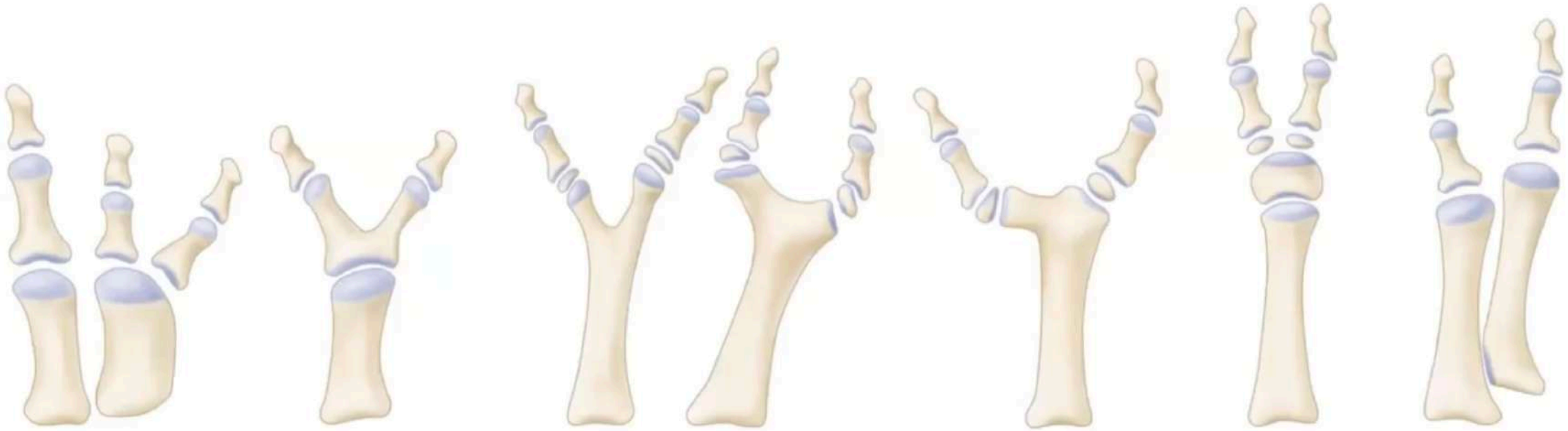
POLYDACTYLY

- MOST COMMONLY ISOLATED TRAIT
- **AD** INHERITANCE
- 2/1000 LIVE BIRTHS

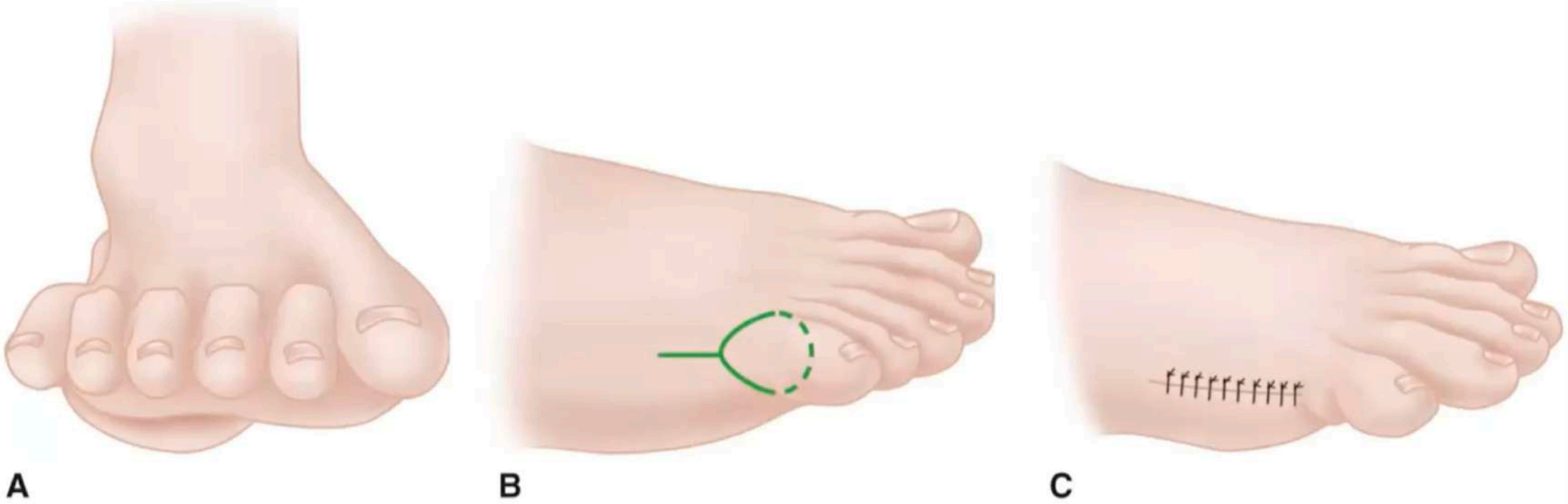
- **PATHOPHYSIOLOGY** : FAILURE OF DIFFERENTIATION IN THE APICAL ECTODERMAL RIDGE DURING 1ST TRIMESTER
- **PROGNOSIS**: SHOE FIITING PROBLEM, ANGULAR DEFORMITIES OF TOES



- Classification : **Venn- Watson**



- Treatment : **amputation of extra toe**



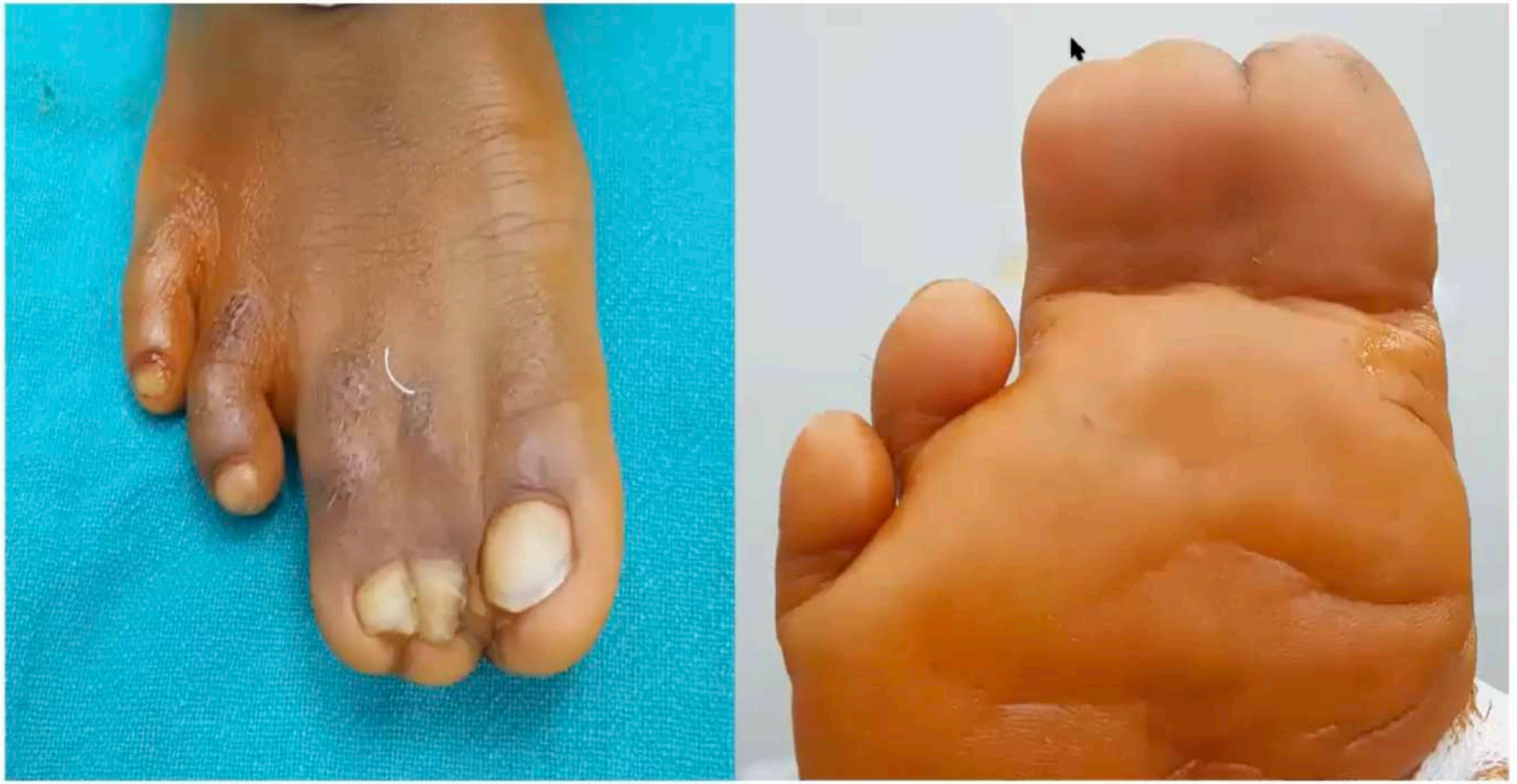
A B C

- **Modified farmer's procedure:**
  - indicated for a duplicated but hypoplastic second toe;
  - involves removal of the second toe;
  - a rectangular rotational skin flap is made in the web space;
  - starting at the proximal aspect of the medial skin flap incision, an additional incision is made across the medial aspect of the great toe MPT joint;
  - the great toe is then lateralized and is partially syndactylize to the third toe (note that the 2nd toe has been removed);
    - complete syndactyly may pull the lesser toes into varus;
  - reinsert the adductor hallucis, in order to avoid hallux varus deformity;
  - the rectangular skin flap is then rotated medially to cover the defect created by rotation of the great toe;

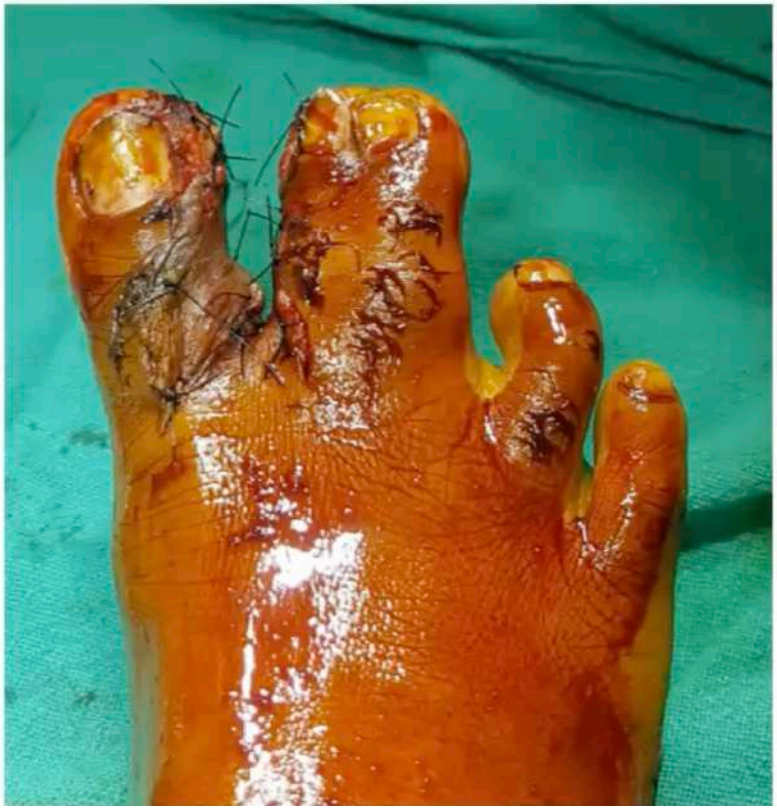
SYNDACTYLY

- Fusion of bone / skin
- 1/2000 live births
- Most common between 2nd and 3rd toe.
- Rarely interferes with functions.
- **AD inheritance**
- **Association:** Down's syndrome, Klippel- feil, familial syndactyly

- Simple : soft tissue fusion
- Complex : bony fusion

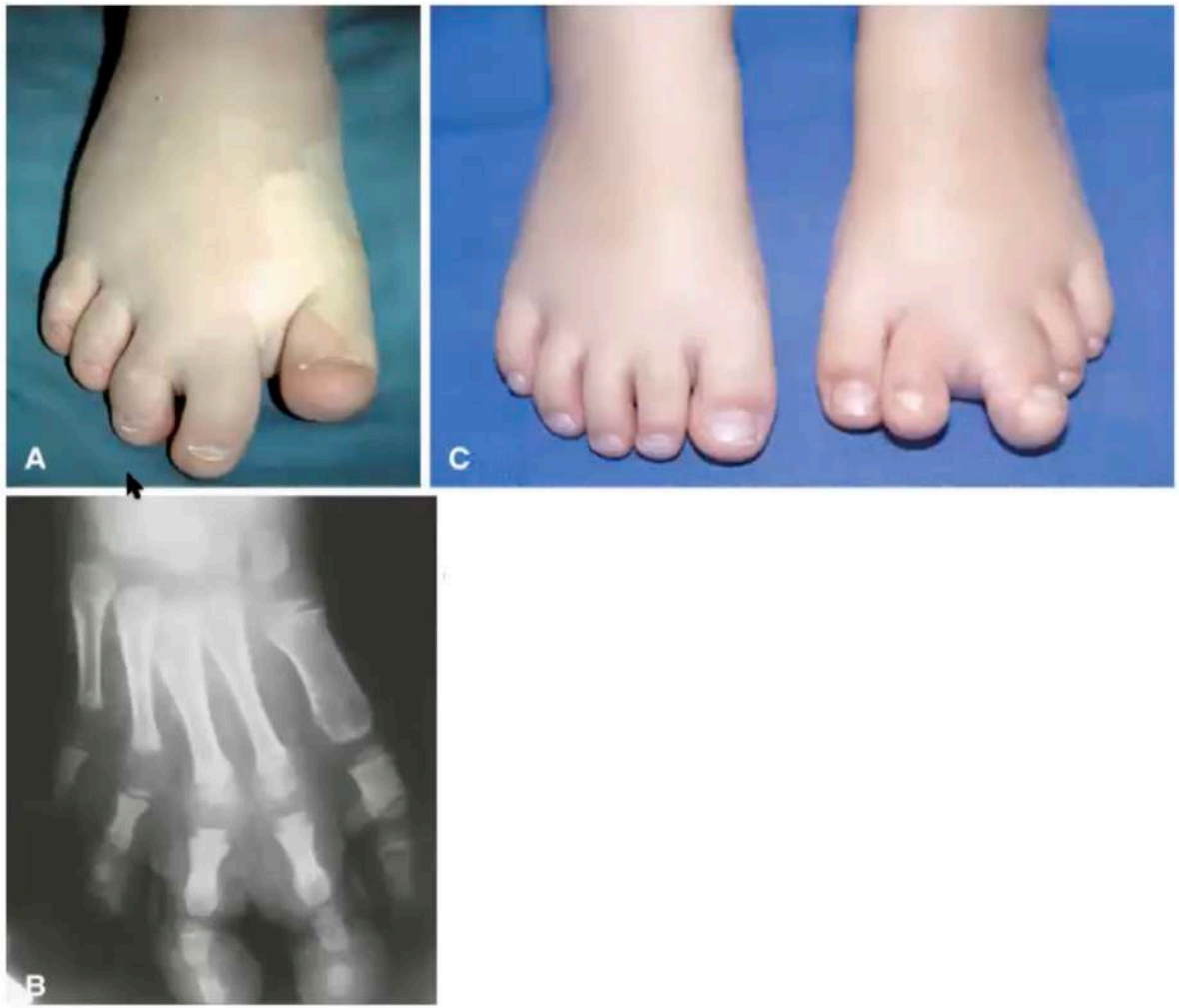


- Treatment : excision of fusion



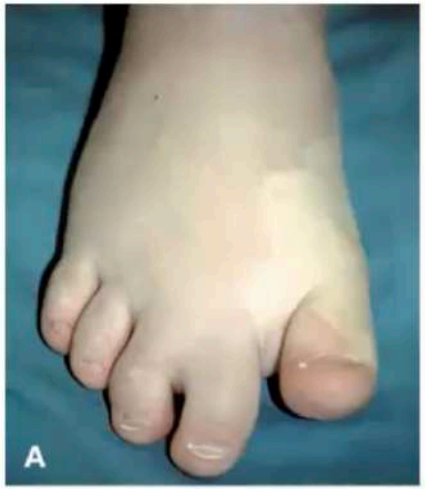
MACRODACTYLY

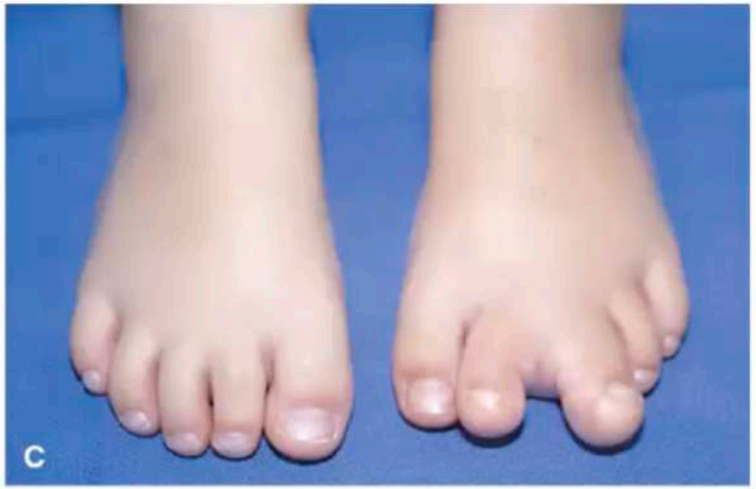
- Hypertrophy (hamartomous enlargement) of one or more toes.
- Aka **local gigantism**
- Associated conditions :
  1. **Neurofibromatosis,**
  2. **hemangiomatosis,**
  3. **congenital lipofibromatosis.**
  4. **Wilm's tumor, adrenal carcinoma, and hepatoblastoma**
  5. **Beckwith-Wiedemann syndrome**
  6. **Macromelia**
  7. **Proteus syndrome**

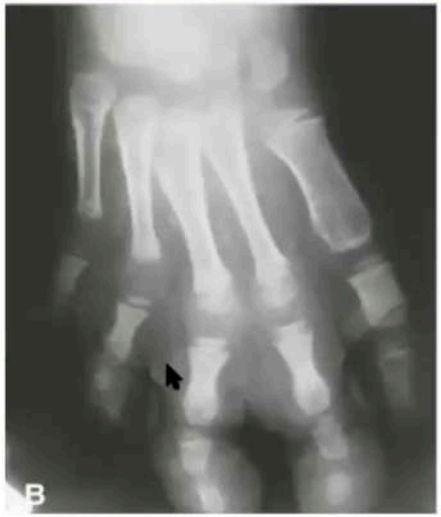


- Differential diagnosis :

  - **AV malformation**
  - **congenital lymphangioma;**
  - **macrodystophia lypomatosa progressive:**
    - rare form of localized gigantism characterized by a congenital and progressive overgrowth of all the mesenchymal elements in digit, with a disproportionate increase in the fibroadipose tissue







- **Macrodactyly simplex congenita:** enlargement includes skin, subQ tissue, nerve, joint, and bone (tendons and blood vessels are of normal size);
  - most often phalanges are involved and metatarsals are spared;
- **Theories : "nerve territory-oriented macrodactyly":**
  - some authors believe that digital nerves cause disproportionate growth of the toes and fingers and that excision in children will reduce growth with minimal neurologic sequelae
- **Symptoms:** pain, footwear issues
- **Cosmetic goal:** similar looking foot

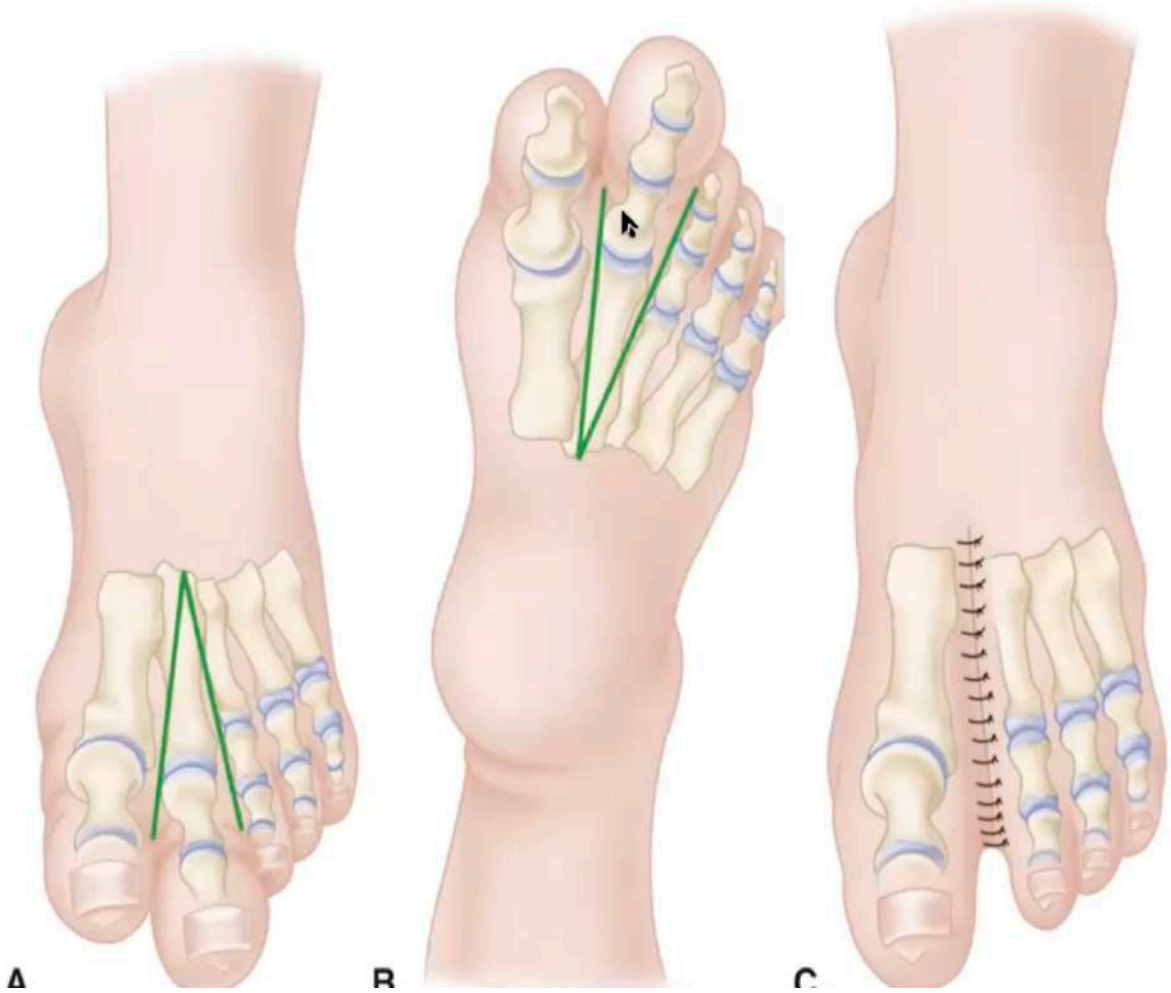
Treatment

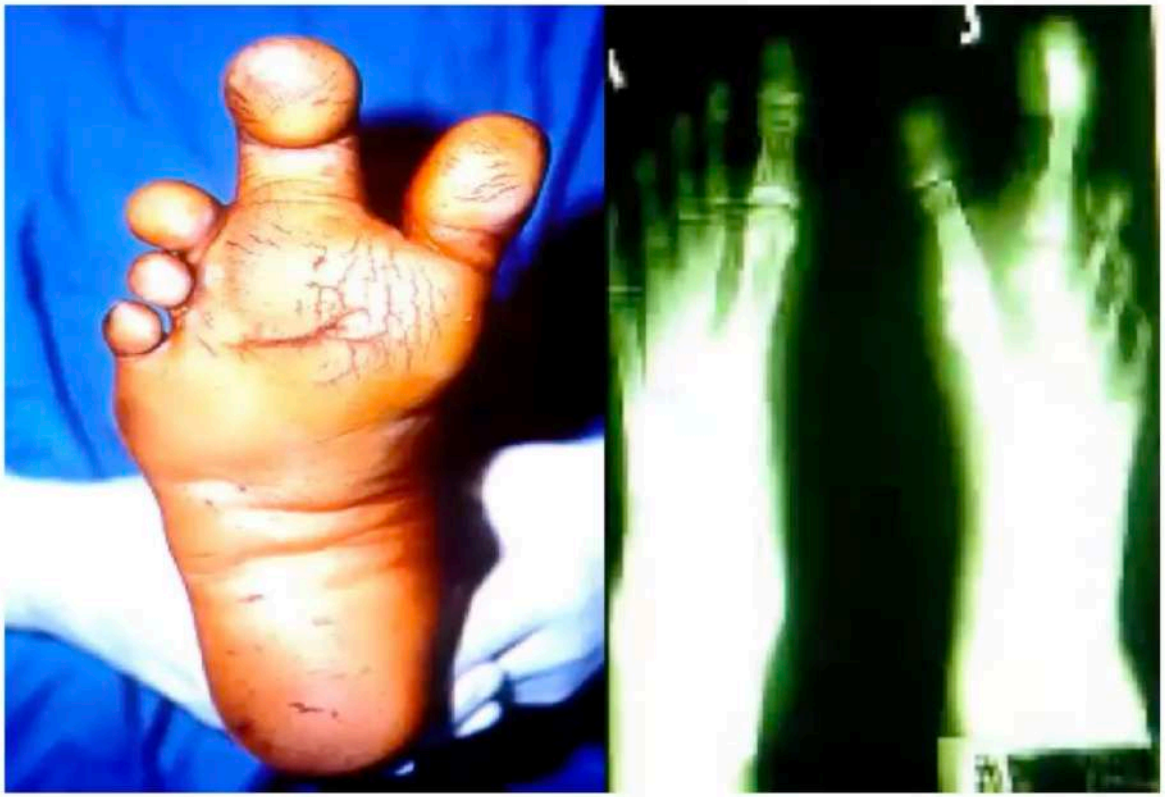
- **Reduction syndactyly**
- **Soft tissue debulking** with ostectomy/ epiphysiodesis (100% recurrence)
- **Ray resection with repeated debulking**
- **Kotwal & Farooque** technique for big toe: staged defatting

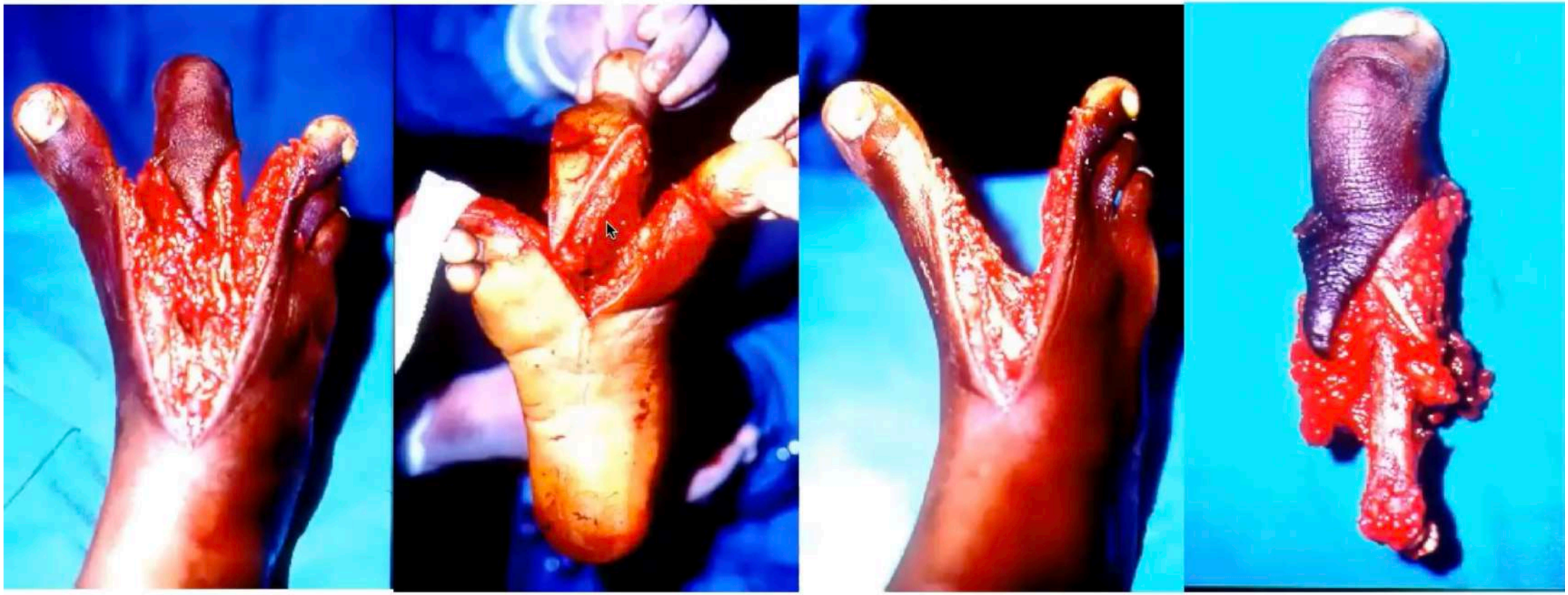
  first stage: convex side (10-20 % reduction)

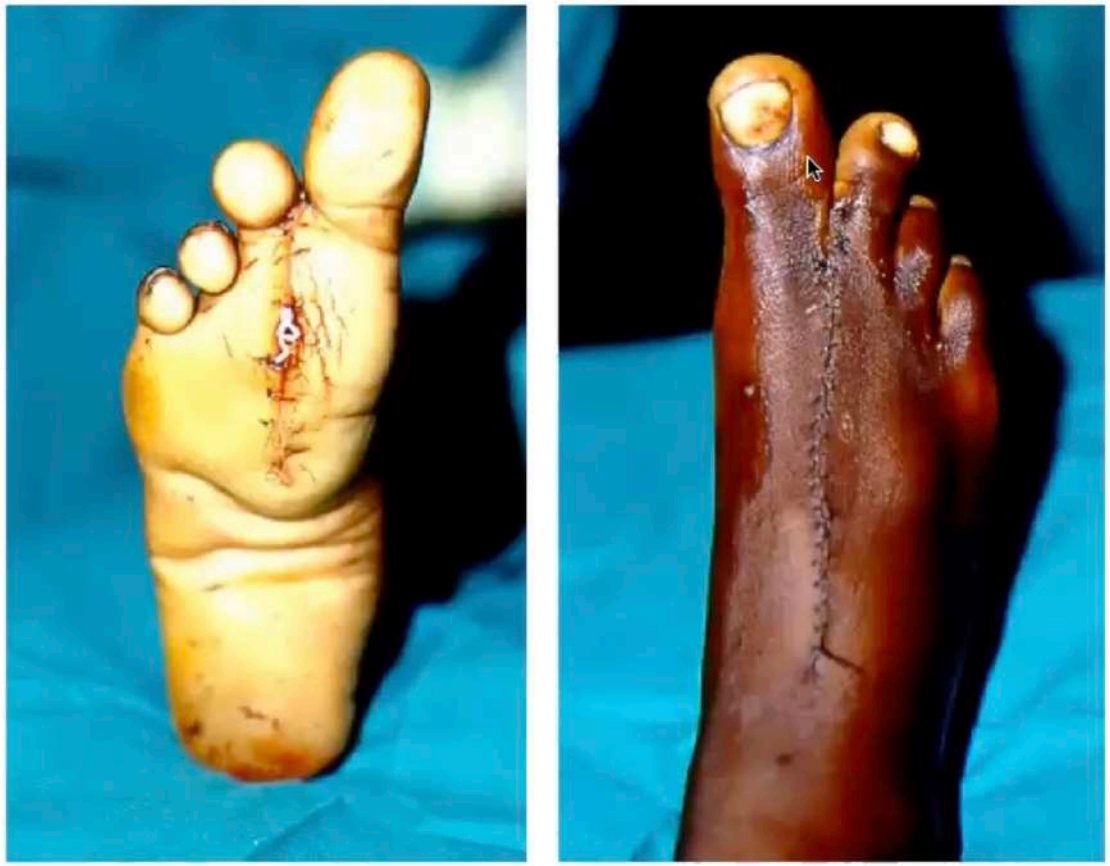
  stage two: after 3 months on other side along with bone shortening

A: dorsal incision
B: plantar incision
C: closure



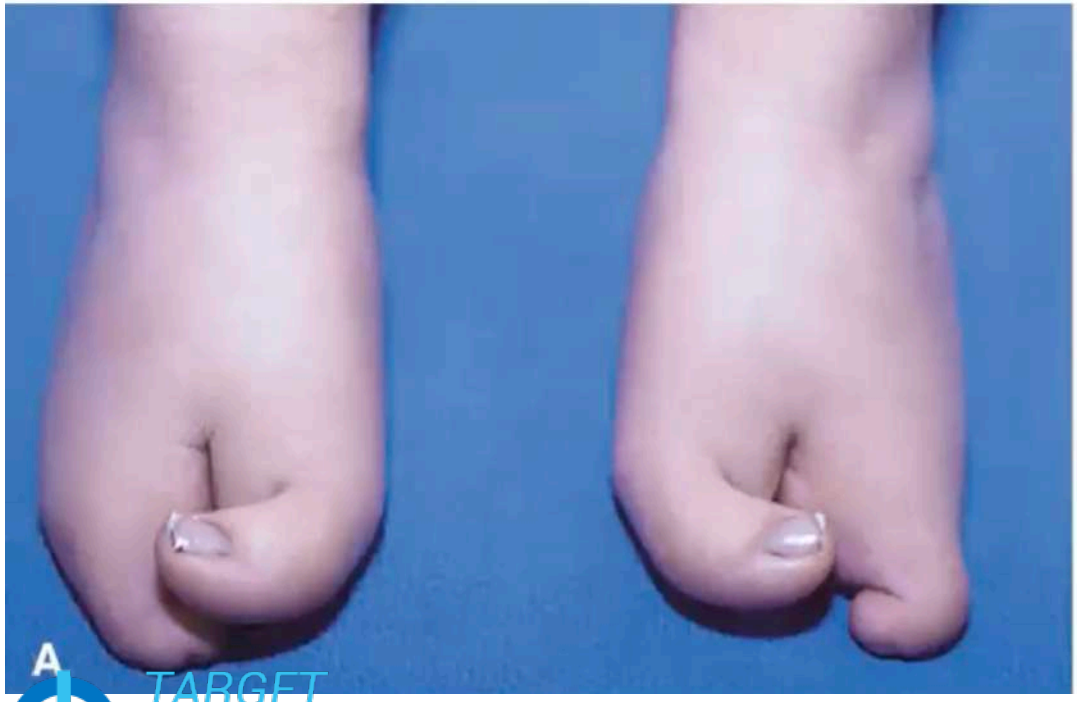


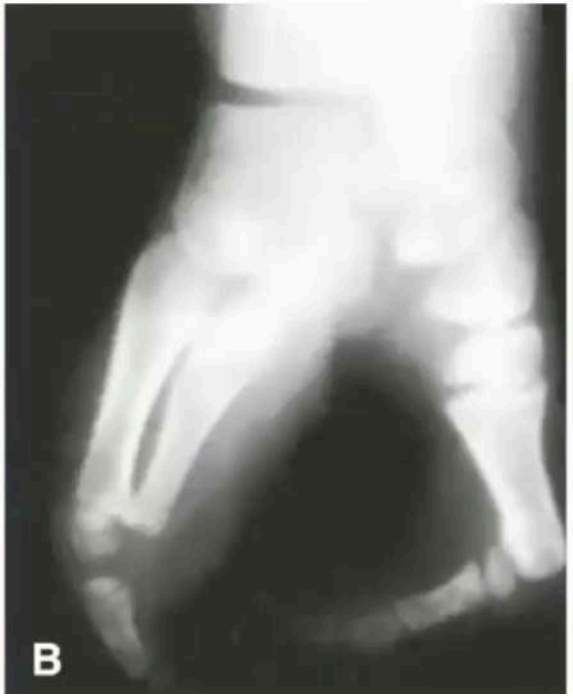


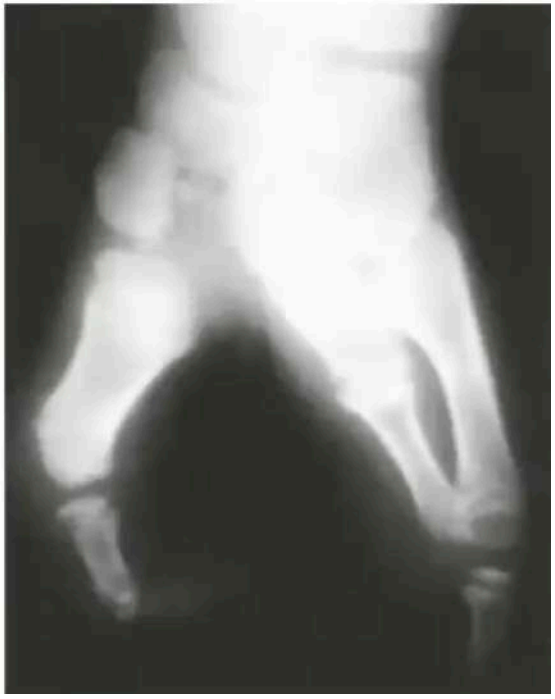


CLEFT FOOT (PARTIAL ADACTYLY)

- Aka **Lobster foot**
- Single cleft extends proximally into the foot







- Multiple deformities
- Usually 1st and 5th rays are present.
- **Blauth & borisch:**

  type 1: cleft with all 5 normal metatarsals

  type 2: cleft with all 5 hypoplastic metatarsals

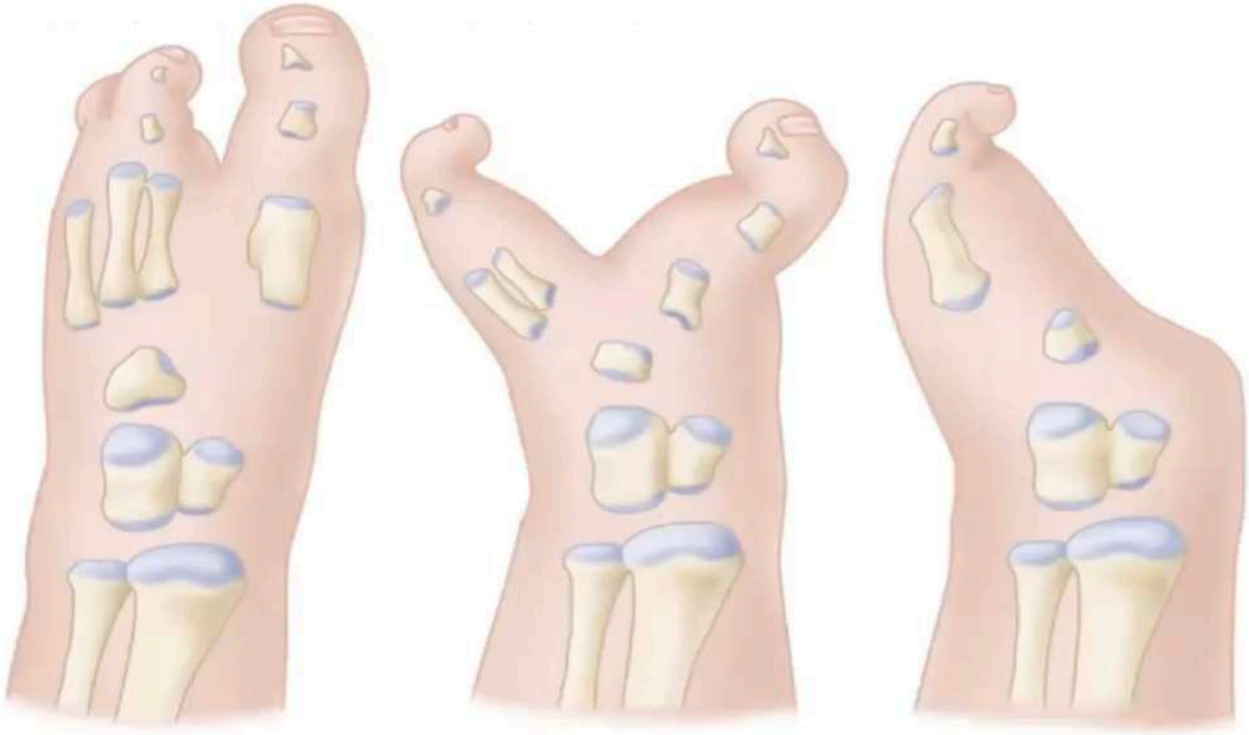
  type 3: cleft with 4MTs

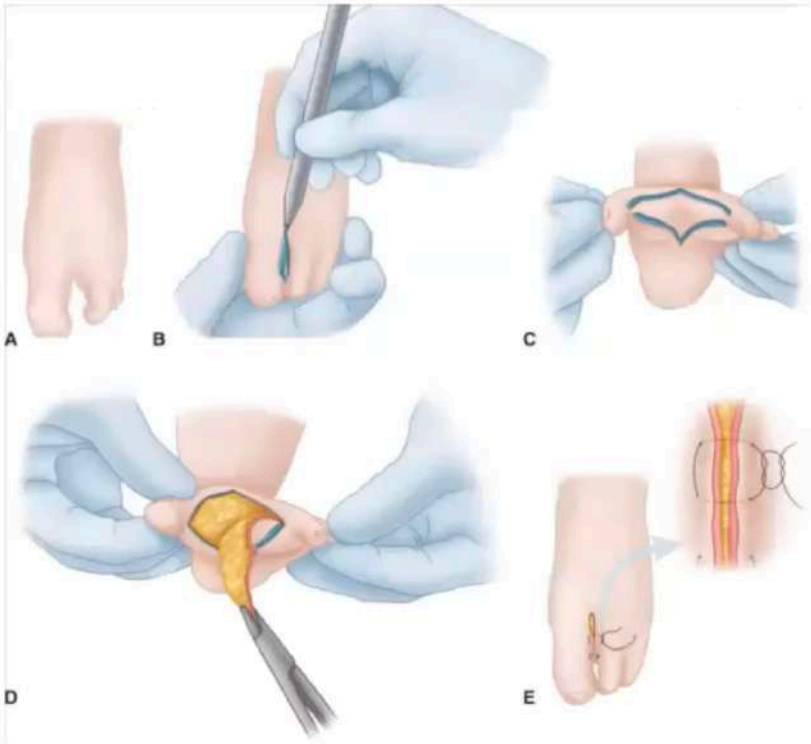
  type 4: cleft with 3 MTs

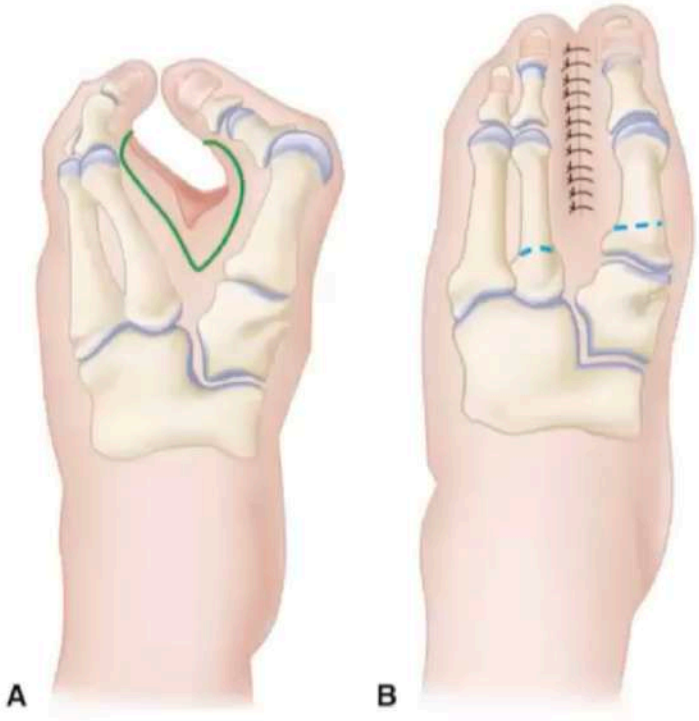
  type 5: cleft with 2 MTs

  type 6: single MT

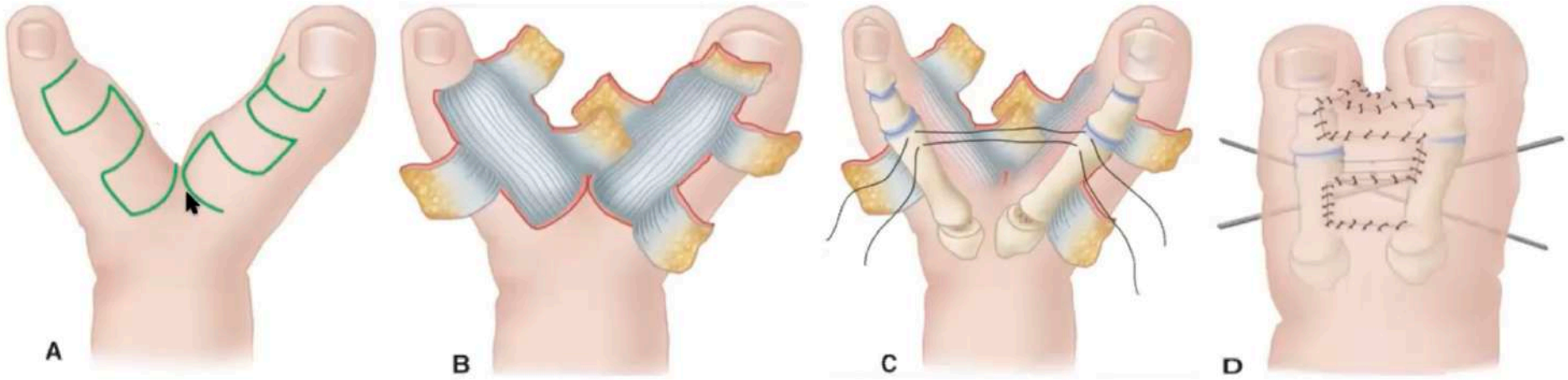
- **Abraham et al**

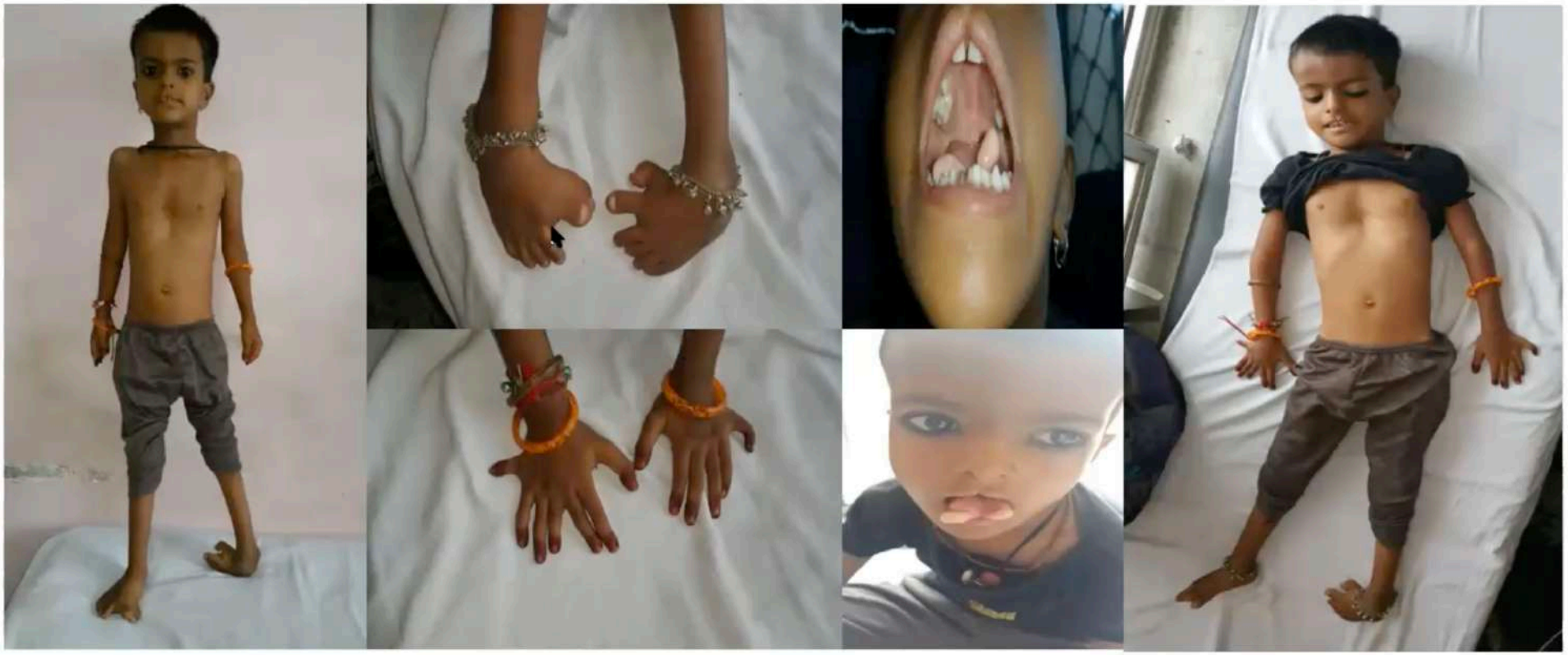






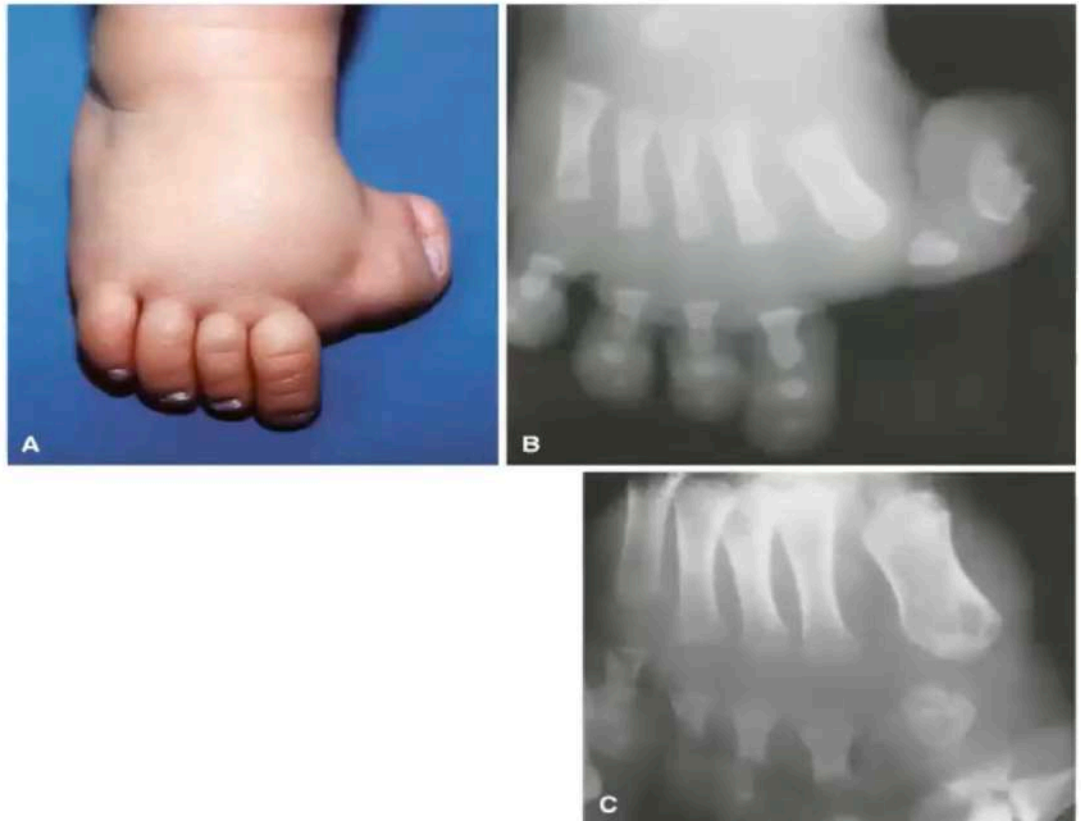
Cleft closure- wood,peppers,shook method





CONGENITAL HALLUX VARUS

- **Atavistic great Toe: medial angulation of great toe**



- Mostly **unilateral**
- Associated conditions:

1. short thick 1st metatarsal

2. accessory bones or toes

3. varus deformity of one or more of the four lateral MT

4. firm fibrous band that extends from medial side of great toe to the base of 1st MT

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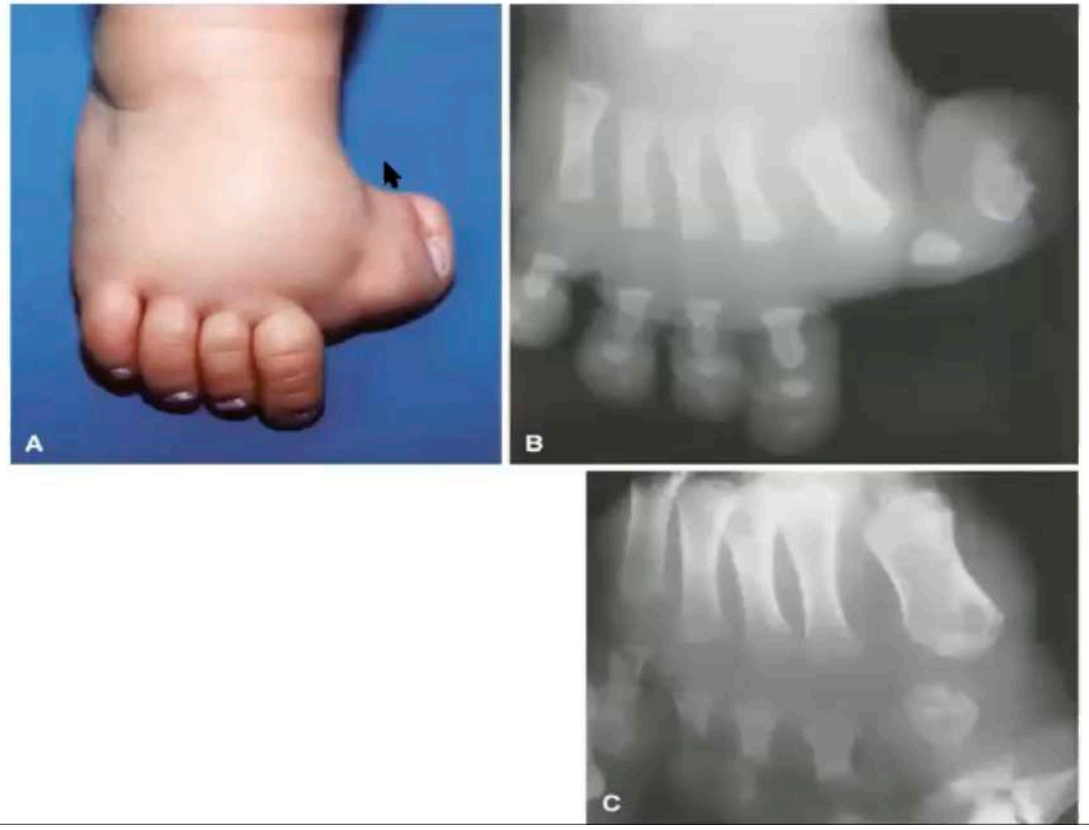

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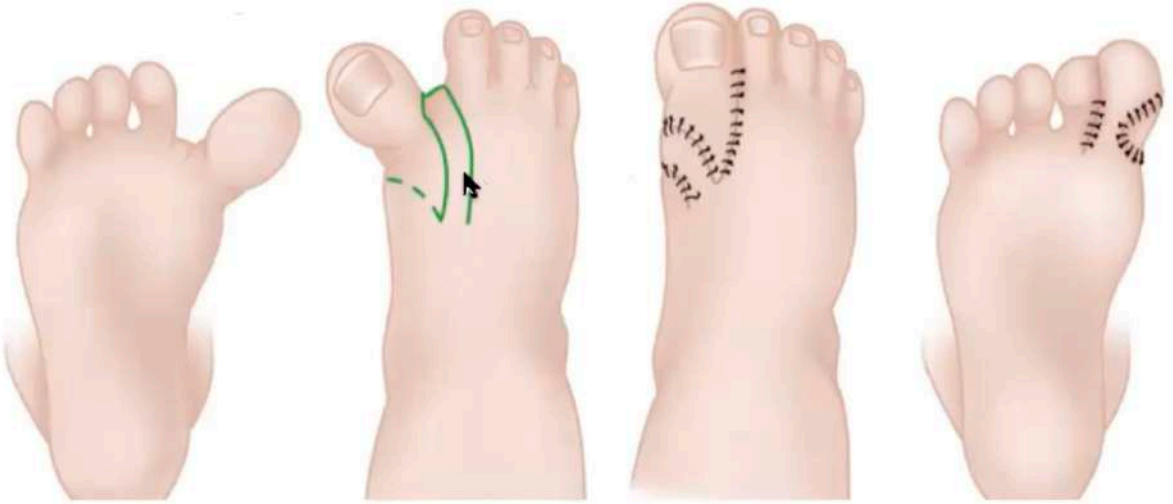
- **Pathoanatomy :**

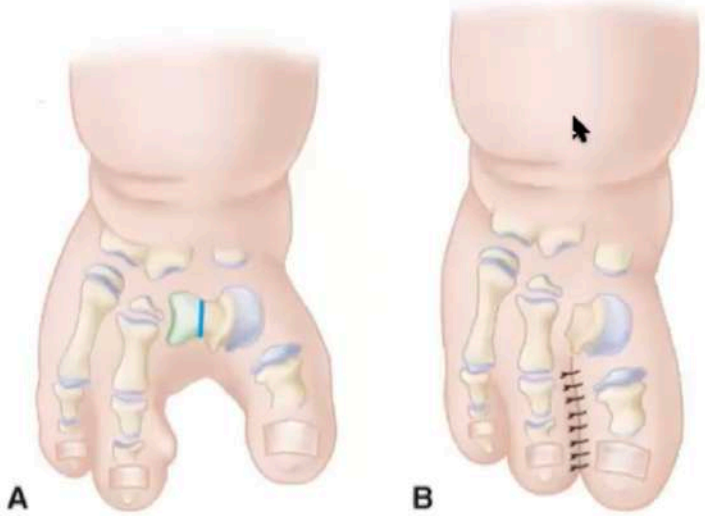
2 great toes in utero → Failure of medial to develop ↓

Rudimentary medial with fibrous tissue forms a band ← Bowstringing to pull medial toe into varus

- Treatment :
- **Farmers procedure : mild to moderate deformity**



- **Kelikian operation: severe deformity with excessive short MT**



CONGENITAL METATARSUS ADDUCTUS

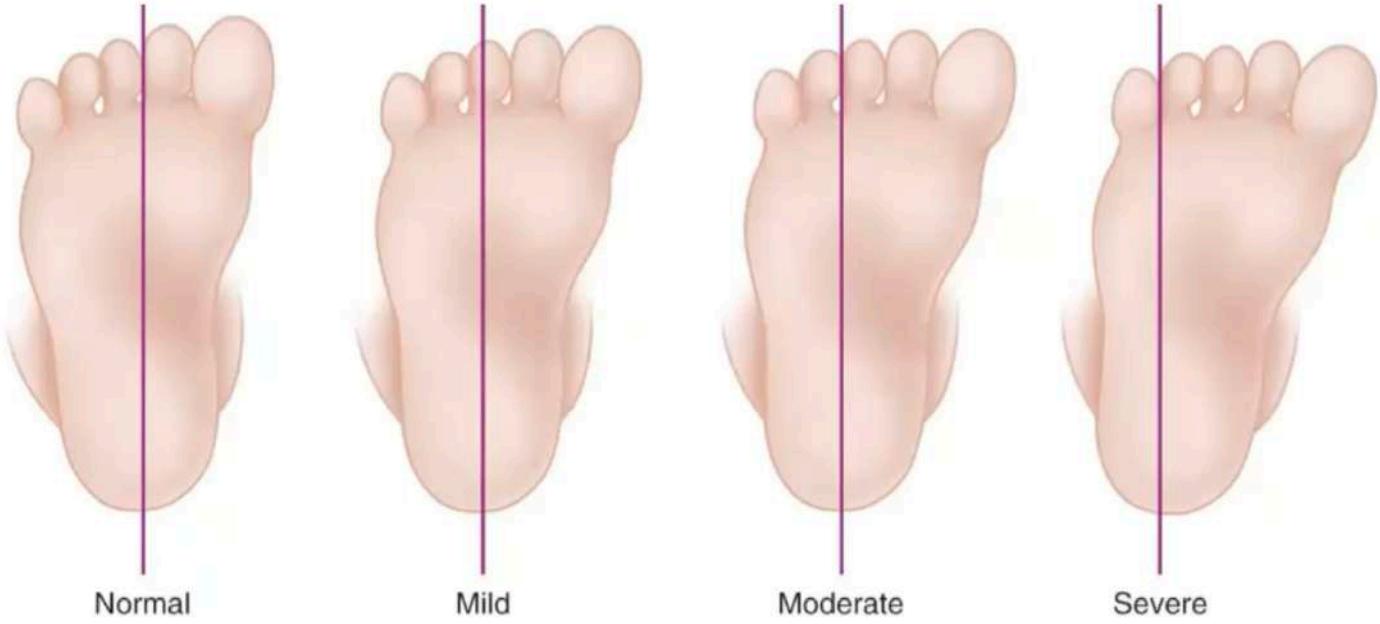
- Adduction of forefoot with normal midfoot and hindfoot alignment
- **In toeing gait**
- Isolated or with CTEV
- Types :

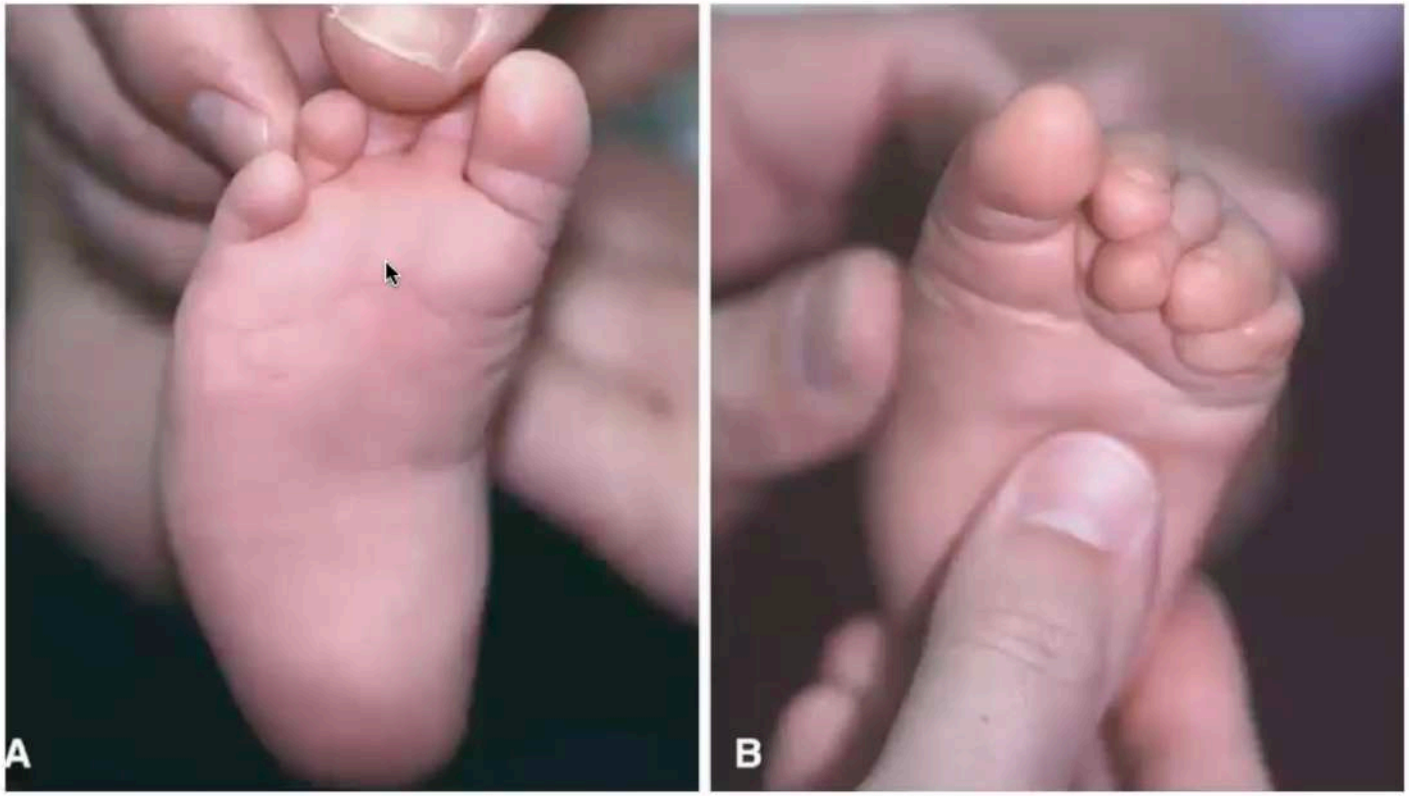
  Mild: forefoot clinically abducted to midline and beyond

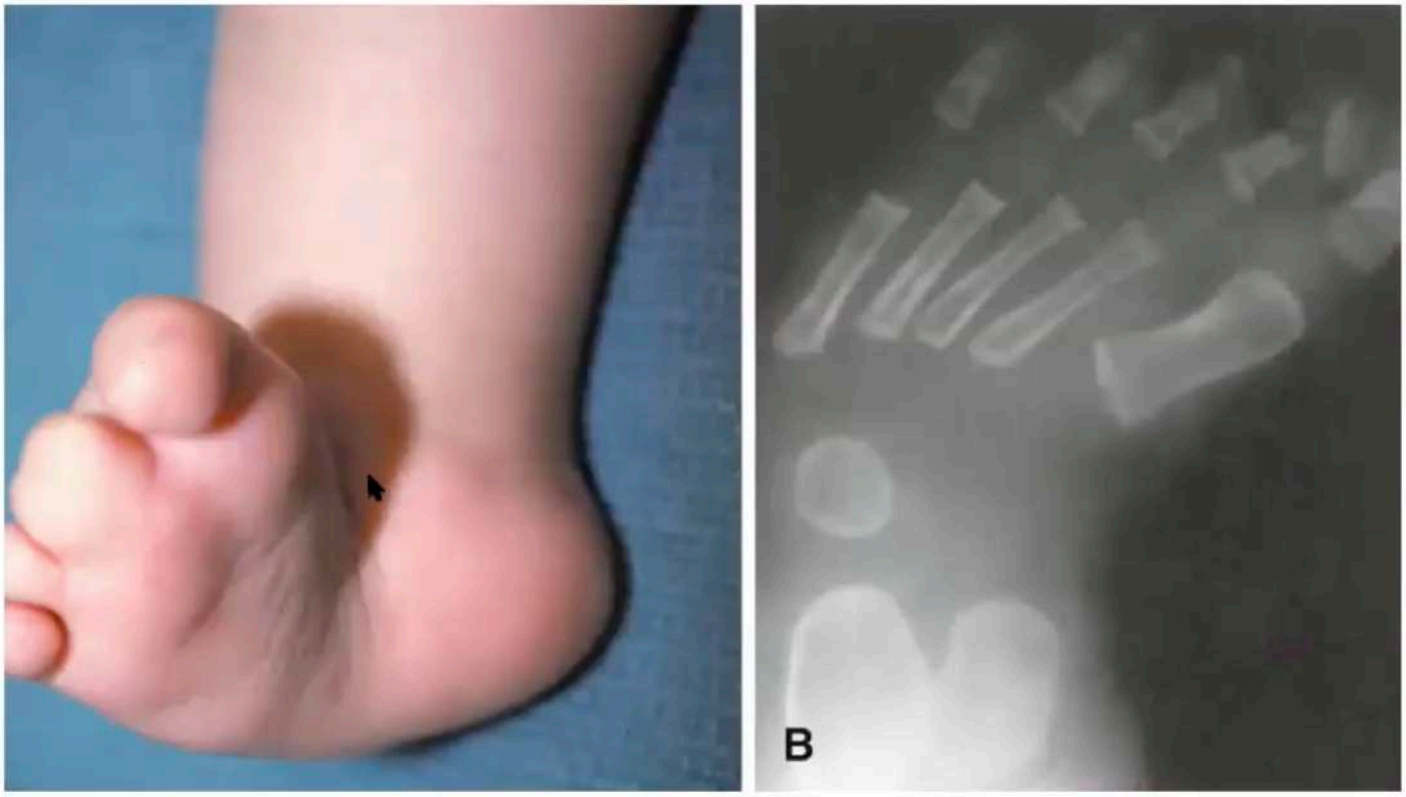
  Moderate :up to midline

  Severe/rigid: cannot be abducted at all

Heel bisector line







Treatment considerations

- Residual deformity in clubfoot
- Rigid/Flexible
- Dynamic : imbalance due to anterior tibial tendon during gait
- Concerns : None/cosmetic/ shoe wear

Treatment of Metatarsus Adductus

None—mild deformities resolve

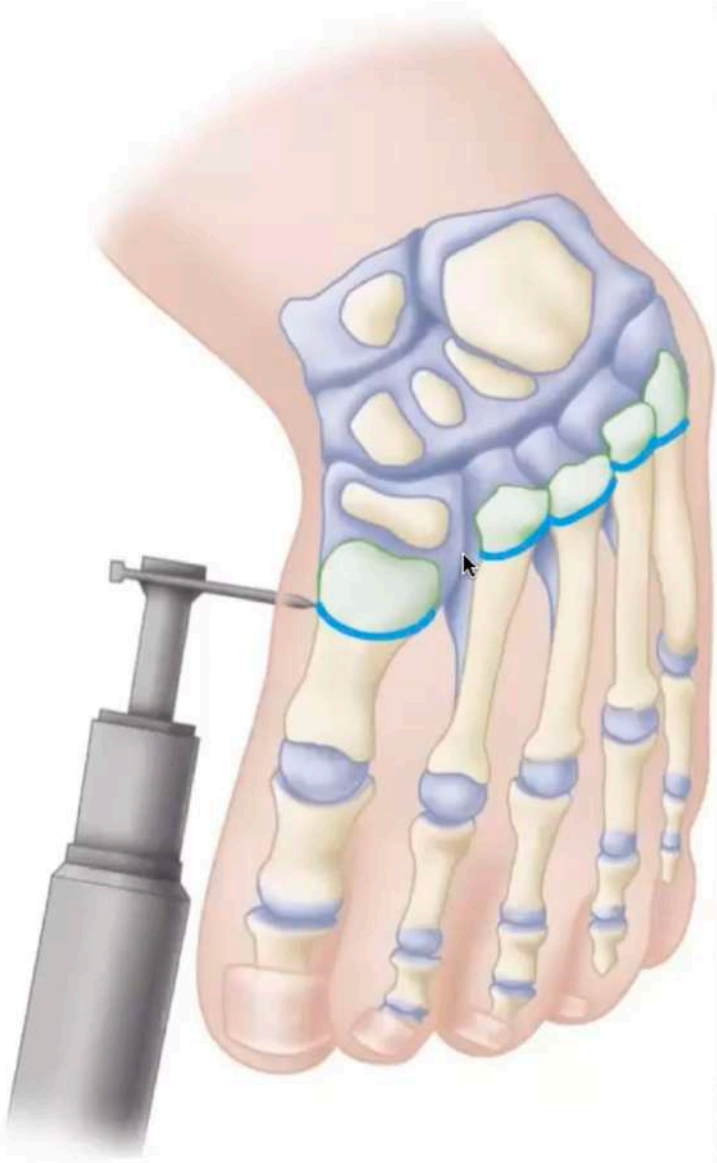
Serial stretching and casting—rarely for moderate and severe deformities

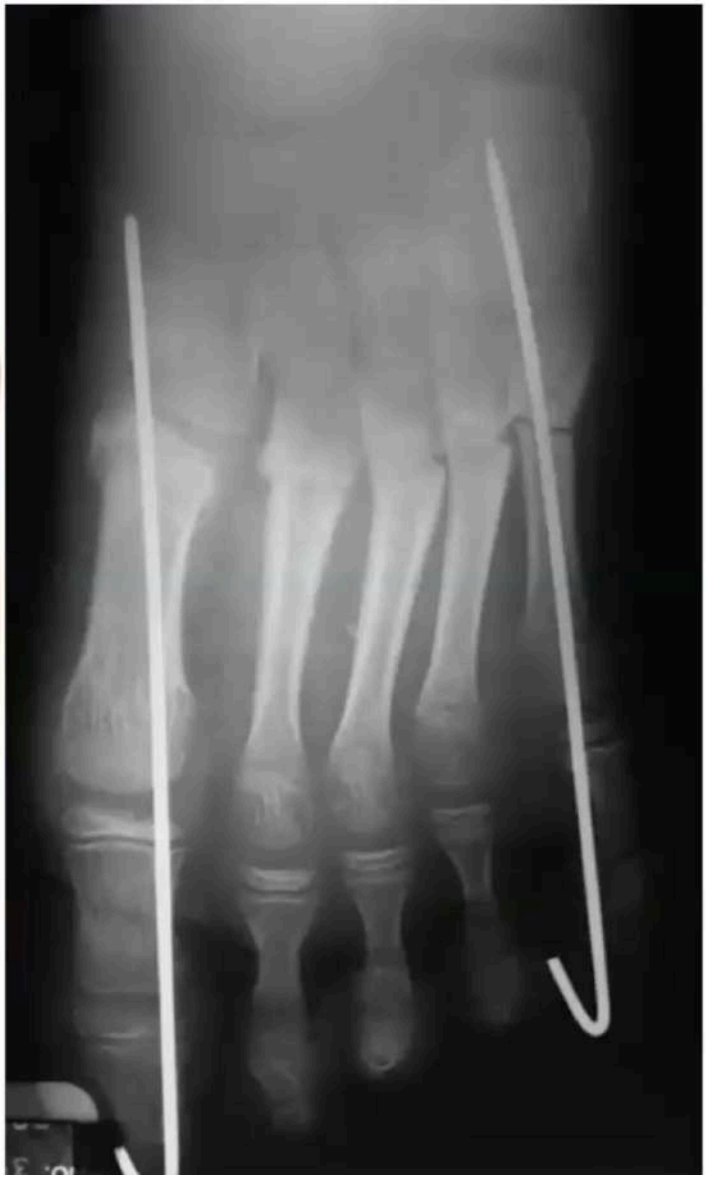
Surgery—severe deformity uncorrected by conservative treatment

- Pain
- Objectionable appearance
- Difficulty in fitting shoes

2-4 yr: Tarsometatarsal capsulotomies (Heyman, Herndon, and Strong)

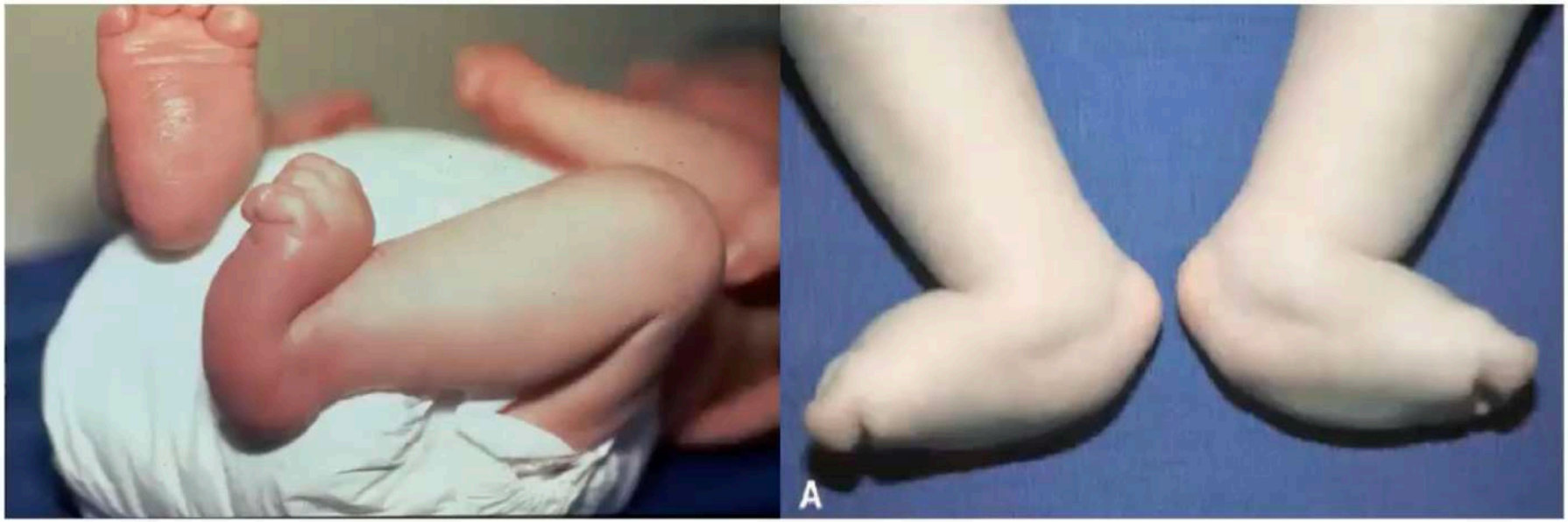
≥4 yr: Multiple metatarsal osteotomies (Berman and Gartland); medial cuneiform, lateral cuboid double osteotomy





CONGENITAL VERTICAL TALUS

- Aka **rocker bottom flat foot/ congenital rigid flat foot**
- Associated with **AMC, myelomeningocele.**
- **Irreducible dorsal dislocation of the navicular on the talus**



- Clinical presentation:

  At Birth:

  Rounded prominence of medial and plantar surface

  talus : **plantarward and medially**

  calcaneus: mild **equinus**

  forefoot : **dorsiflexed** at midtarsal joint

  navicular: **dorsal** aspect of talus

  sole: convex

  deep creases on dorsolateral aspect of foot

- **At weight bearing :**adaptive changes

  talus: hourglass, equinus with longitudinal axis same as tibia

  calcaneus: equinus and posteriorly displaced

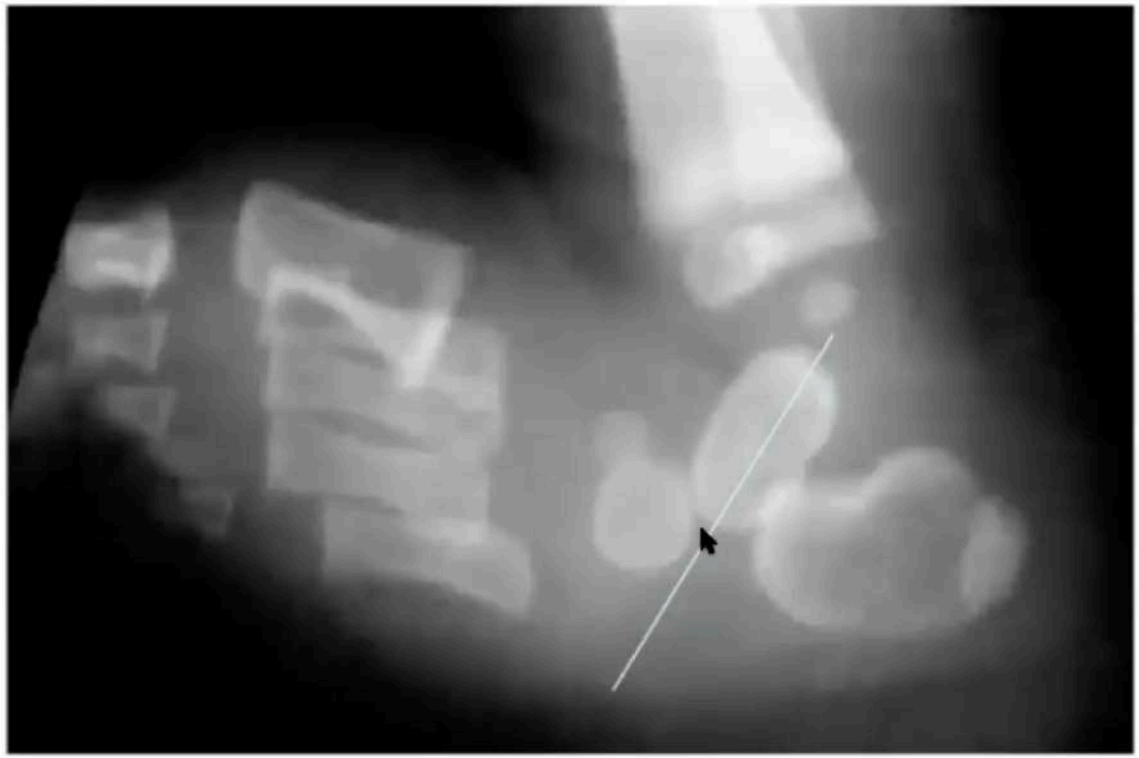
  callosities: ant end of calcaneum and medial border of foot

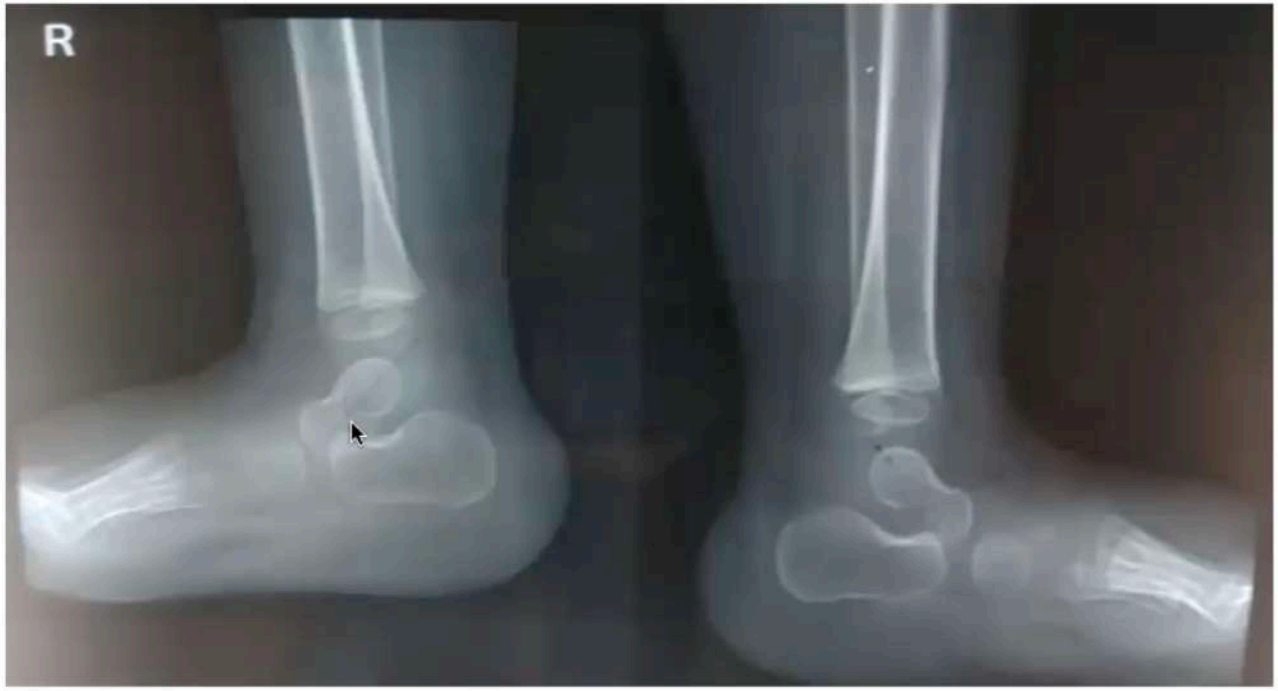
  forefoot: abducted

  all capsules, ligaments and tendons on dorsal are contracted and function abnormally

  DD: rigid pes planus, posteromedial tibial bowing, paralytic flat foot

- gait abnormality : **Peg leg gait-** poor push off





Meary's angle > 20° (between line of longitudinal axis of talus and longitudinal axis of 1st

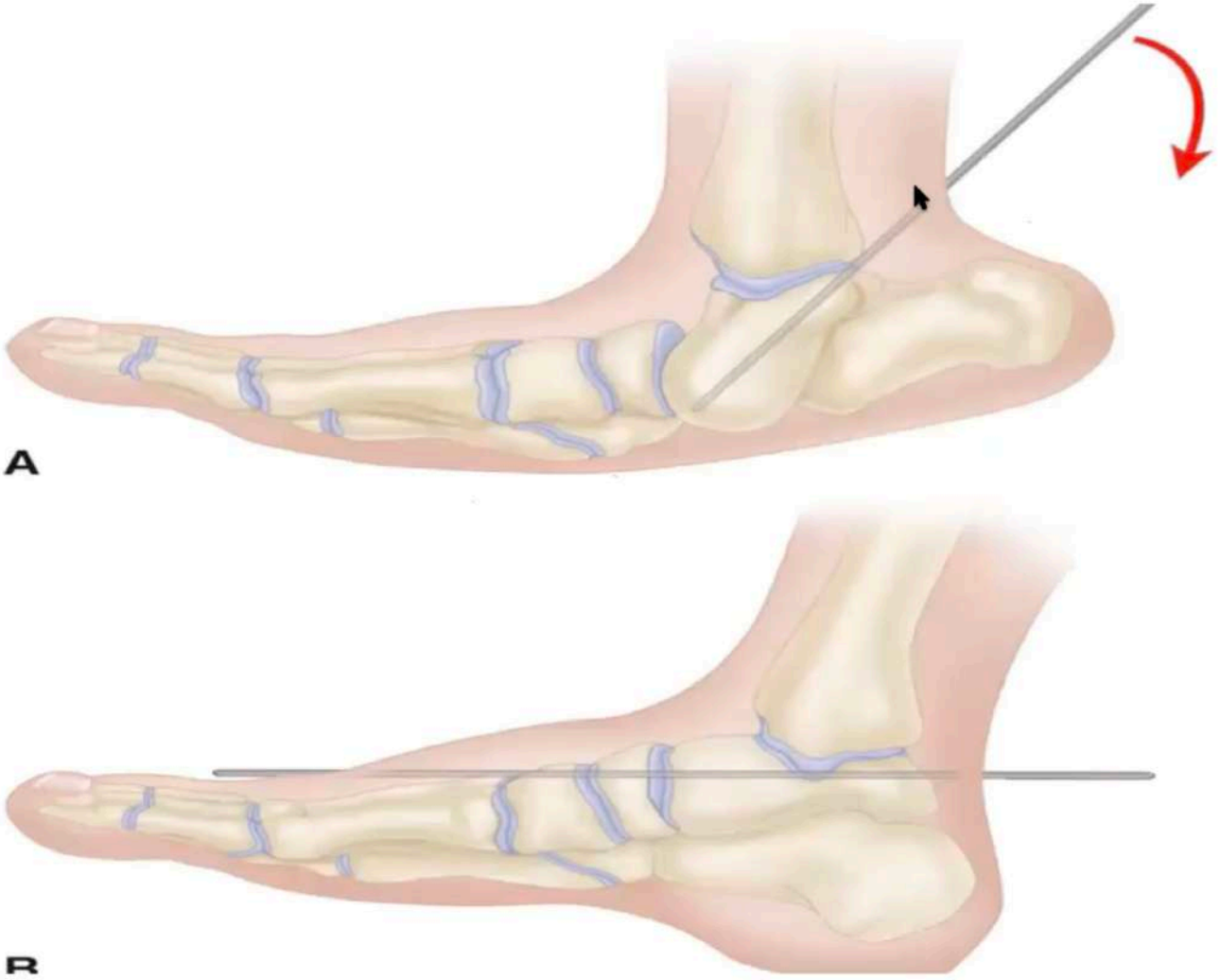
Treatment

- **Serial manipulation and casting:** usually pre op

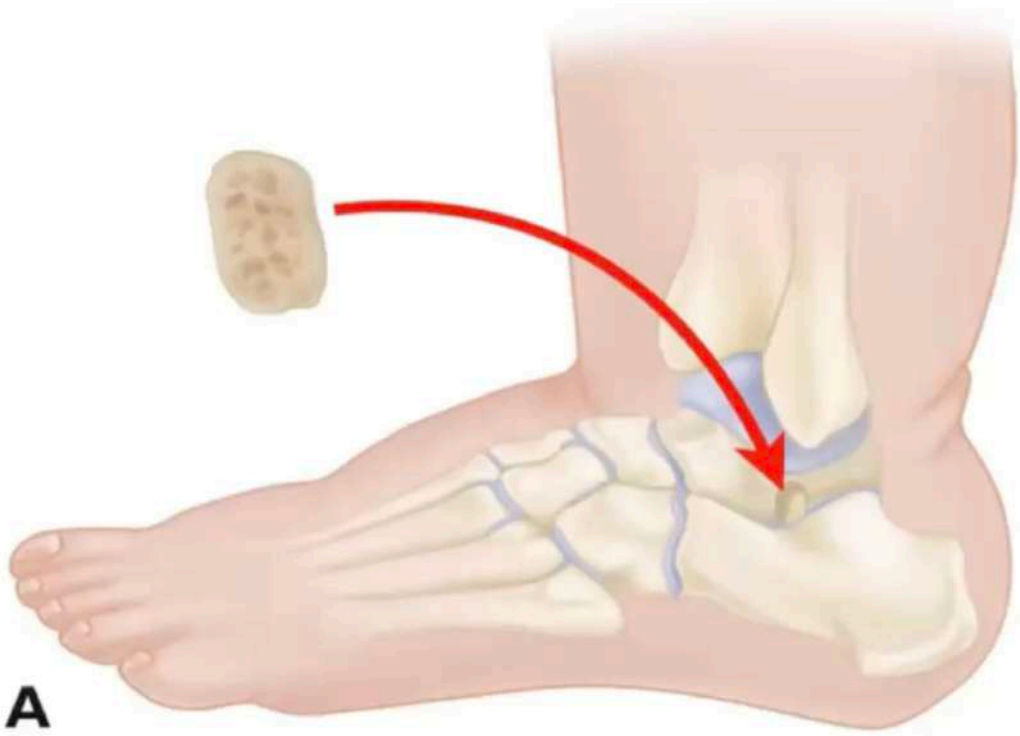
  foot is inverted and plantarflexed
- Surgery : open, Mini invasive, Talectomy, triple arthrodesis

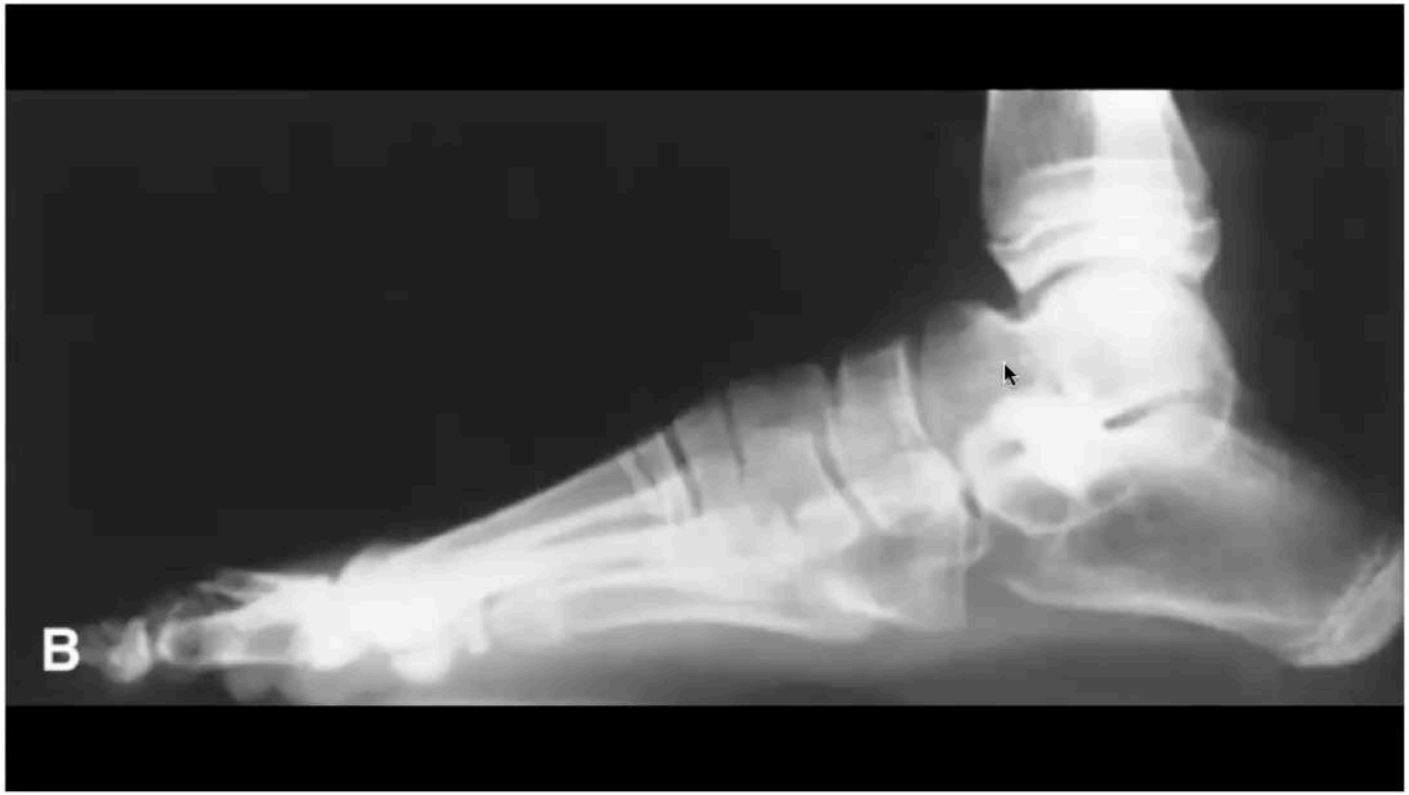
- **1- 4 years** : open reduction and realignment of TN & ST joints

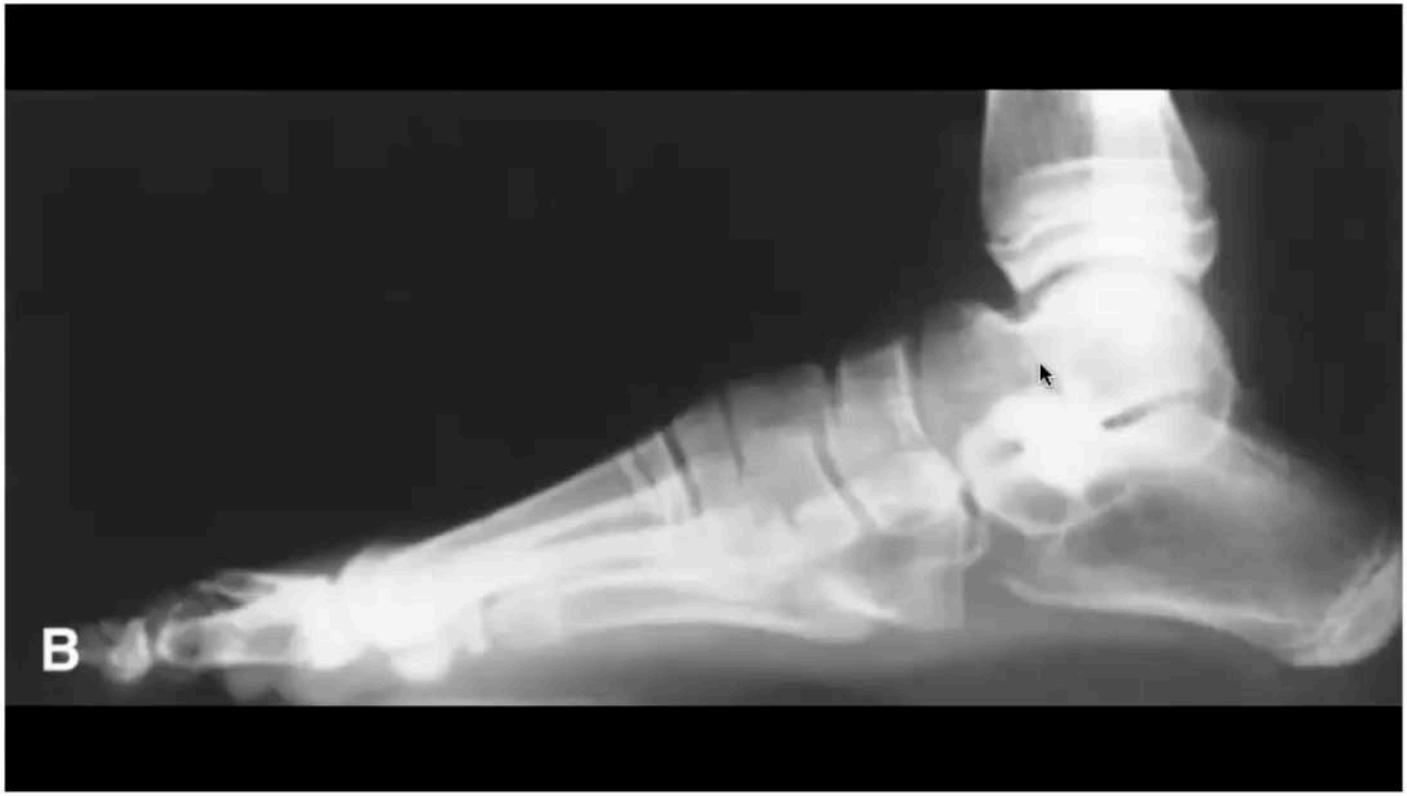
  if deformity is **severe- navicular excision** can be done (after 3 years)
- **4- 8 years** : open reduction and soft tissue procedure with extra articular subtalar arthrodesis
- **> 12 years**: triple arthrodesis

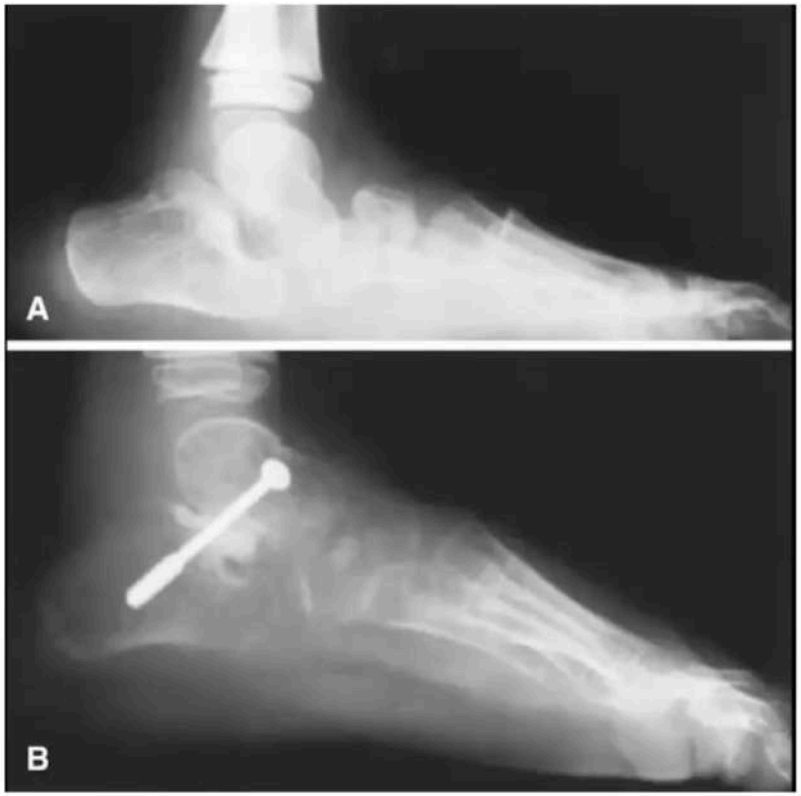


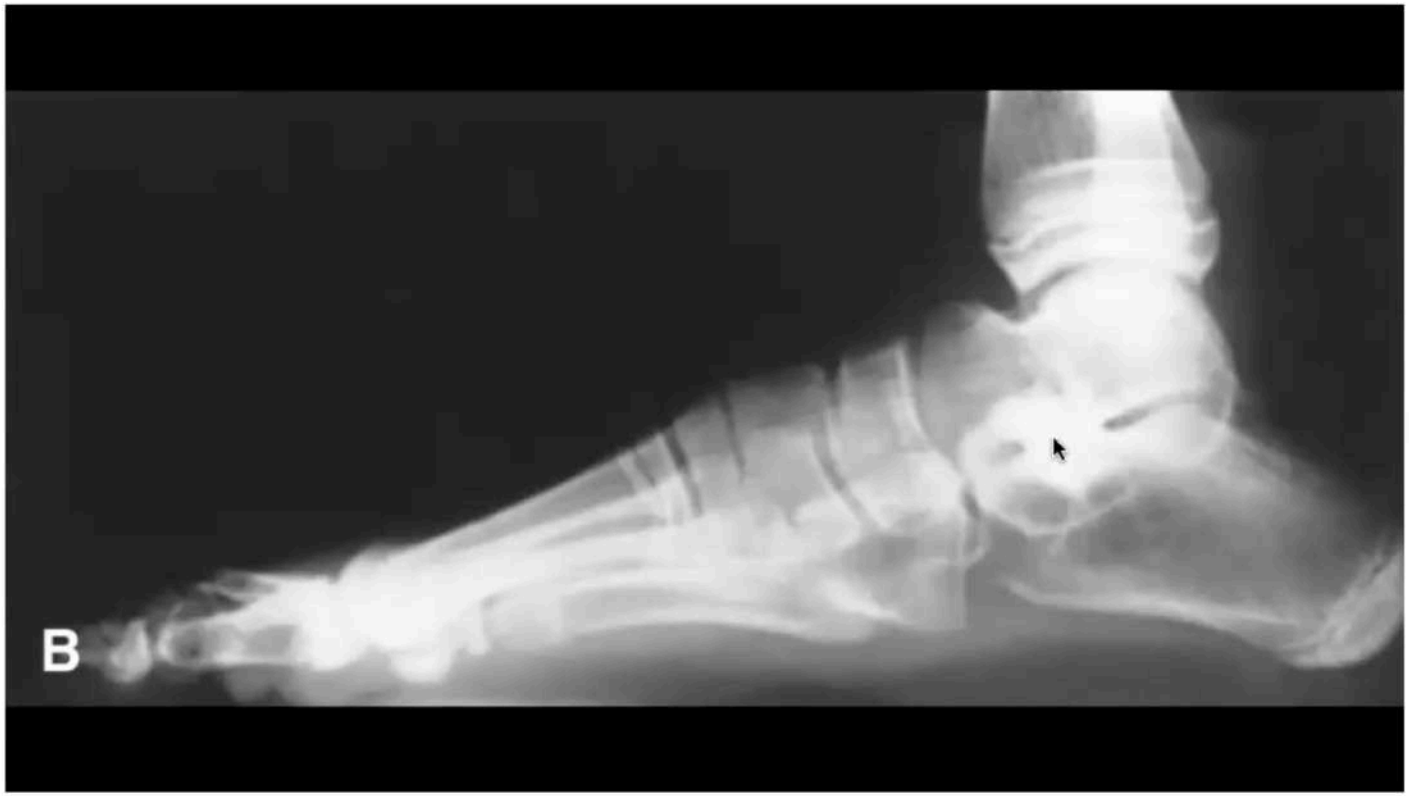
Grice and green extraarticular fusion









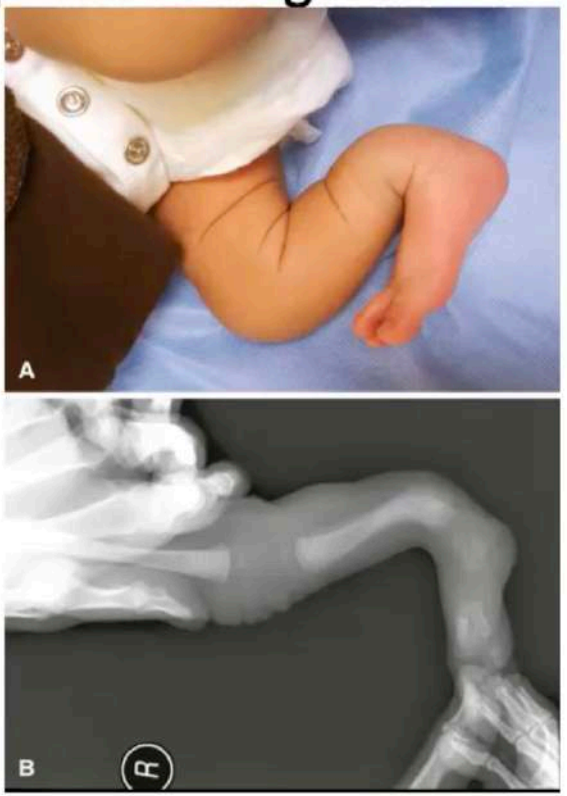


CONGENITAL ANGULAR DEFORMITIES OF LEG

- Two types:

- A. **posteromedial bowing of tibia**
- B. **anterolateral bowing of tibia- ass. With neurofibromatosis, CPT**

- **Posterior deformities**: usually resolve or improve with growth
- LLD
- Yearly examination and close follow up
- Epiphysiodesis, limb lengthening



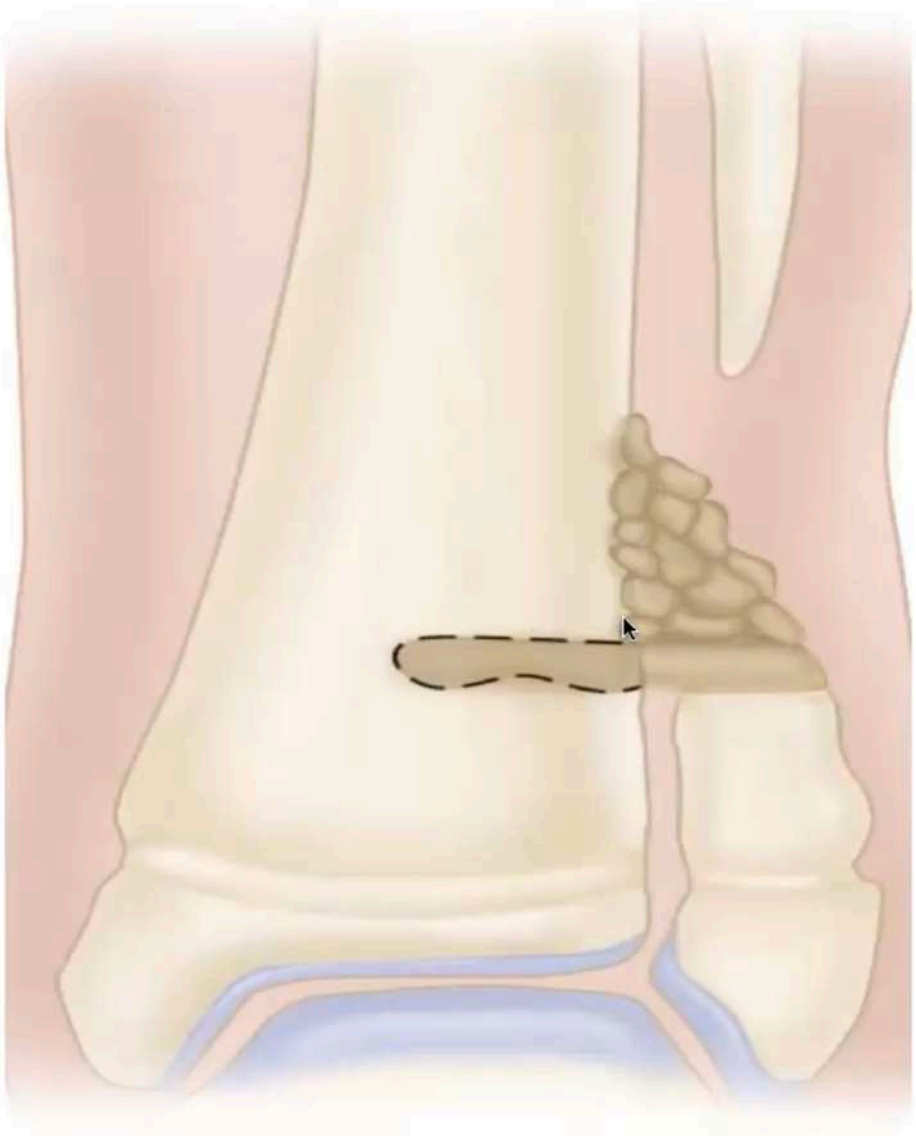
CONGENITAL PSEUDOARTHROSIS OF FIBULA & TIBIA

- Specific type of non union at birth or incipient
- Seen with **neurofibromatosis**
- **M.C – distal half of tibia**
- Cause of non union- unknown , but some hamartomous fibrous tissue with limited vascular ingrowth is seen at the site.
- Extremely rare
- 1:250000 live births
- Up to **50-90%** associated with **Neurofibromatosis Type 1**
- **15% associated with Fibrous Dysplasia**

- **Fibula :**
- Often precedes or accompanies tibia
- **Type 1: bowing of fibula without pseudoarthrosis**
- **Type 2: fibular pseudoarthrosis without ankle deformity**
- **Type 3: fibular pseudoarthrosis with ankle deformity**
- **Type 4: fibular pseudoarthrosis with latent pseudoarthrosis of tibia**

- **Until skeletal maturity**: **AFO**
- **At maturity: supramalleolar osteotomy**
- **Langenskiold operation: children to prevent ankle valgus**

  created a **tibio-fibular synostosis** using iliac crest graft



Tibia – boyd classification

- **Type 1**: pseudoarthrosis with anterior bowing and tibial defect at birth
- **Type 2: anterior bowing and hourglass constriction of tibia at birth.**

  spontaneous fracture/ after trivial trauma before 2 years

  " high risk tibia"- tapered, round, obliterated canal

  " most common", poor prognosis, neurofibromatosis
- **Type 3:** pseudoarthrosis in a **congenital cyst,** junction of middle and lower 3rd tibia , anterior bowing before or after a fracture
- **Type 4**: pseudoarth. In **sclerotic bone segment . Stress #**

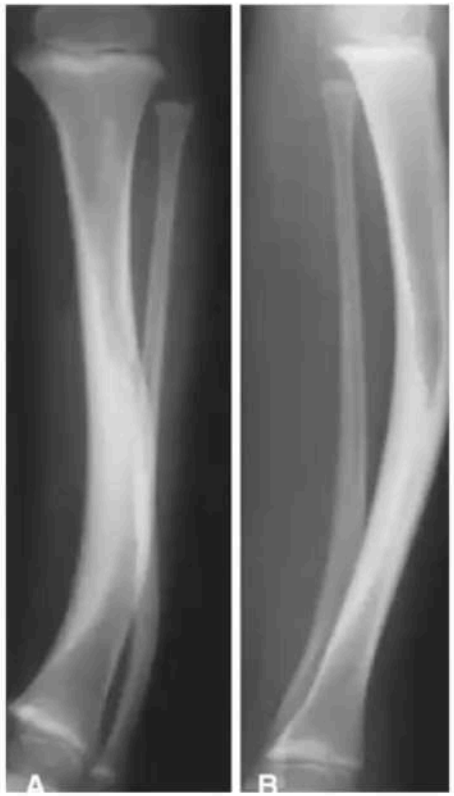
  good prognosis if insufficiency is treated before fracture becomes complete

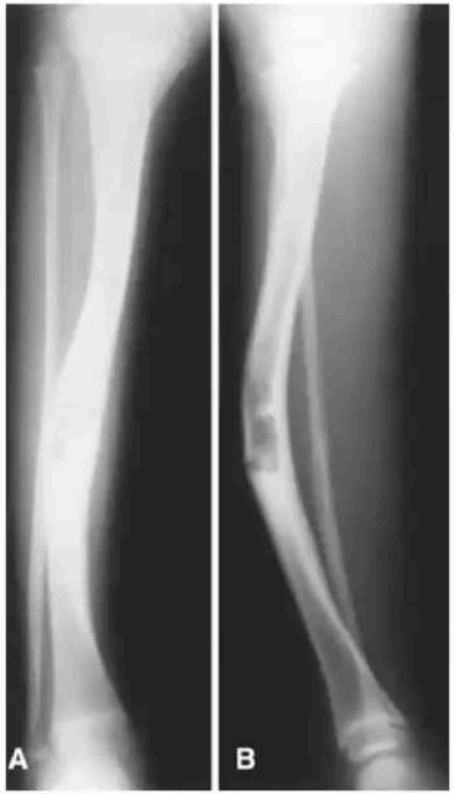
- **Type 5: pseudoarthrosis with dysplastic fibula.**

  good prognosis if lesion is confined to fibula, but if reaches tibia then acts like type 2

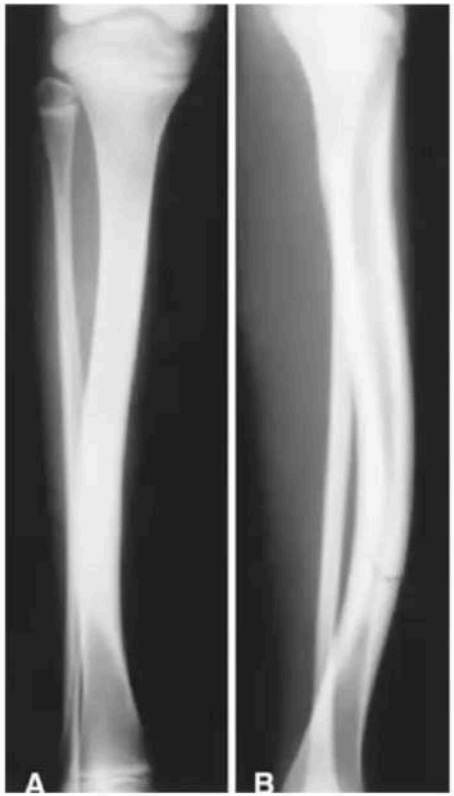
Type 6: intraosseous neurofibroma/ schwannoma

extremely rare.





Type 4



Treatment

- Age of the patient
- Presence/ absence of fracture

- **Before walking age: no treatment**

- **bracing in clamshell orthosis or patellar tendon bearing (PTB) orthosis:**

  Children of ambulatory age (weight bearing) bowing without pseudarthrosis or fracture

  spontaneous remodeling is not expected

- goal is to prevent further bowing and fractures
- Maintain until skeletal maturity

- **Surgical fixation**: bowing with pseudoarthrosis or fracture

  restoration of alignment and IM fixation

- **Amputations** :

  typically indicated after **multiple failed surgical attempts** at union

  severe limb length discrepancy(**shortening > 3 inches**)

  dysfunctional angular deformity

  Method- Syme or Boyd amputation

- **bracing in clamshell orthosis or patellar tendon bearing (PTB) orthosis:**

  Children of ambulatory age (weight bearing) bowing without pseudarthrosis or fracture

  spontaneous remodeling is not expected

- goal is to prevent further bowing and fractures
- Maintain until skeletal maturity