Congenital Hand
Orthopaedic Registrar Teaching
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Aims

- How to approach children with congenital hand anomalies
- Principles of management
- Individual anomalies & management

Approach to patient

- Analyse each case individually

- MDT “on-tap”approach
  - Hand surgeon
  - Hand therapist
  - Psychologist
  - Genetics
  - Paediatrics
  - Orthopaedics

Approach to patient

Parental response

- Bereavement response
- Thorough explanation of anomalies, treatment, prognosis
- Cause/blame
- Time needed for consultations
- Multiple consultations often required

Approach to patient

- History
  - Family History
- Examination
  - Complete upper limbs
    - Include pec major muscle
  - Lower limbs- feet
  - Other
- Imaging
  - X-rays
  - USS/MR scan

Principles of management

- Aim to achieve maximum possible function
- Key functions
  - Hand to mouth
  - Toileting
  - Pinch grip
    - Tripod pinch
    - Key pinch

Principles of management

- Surgery not always the answer
- Early referral, neonatal period
  - commence physiotherapy- stretches +/- splinting
- Consider other conditions
  - Integrated management
  - May need to refer patients to other specialties
- Timing surgery
- Follow-up patients until fully grown

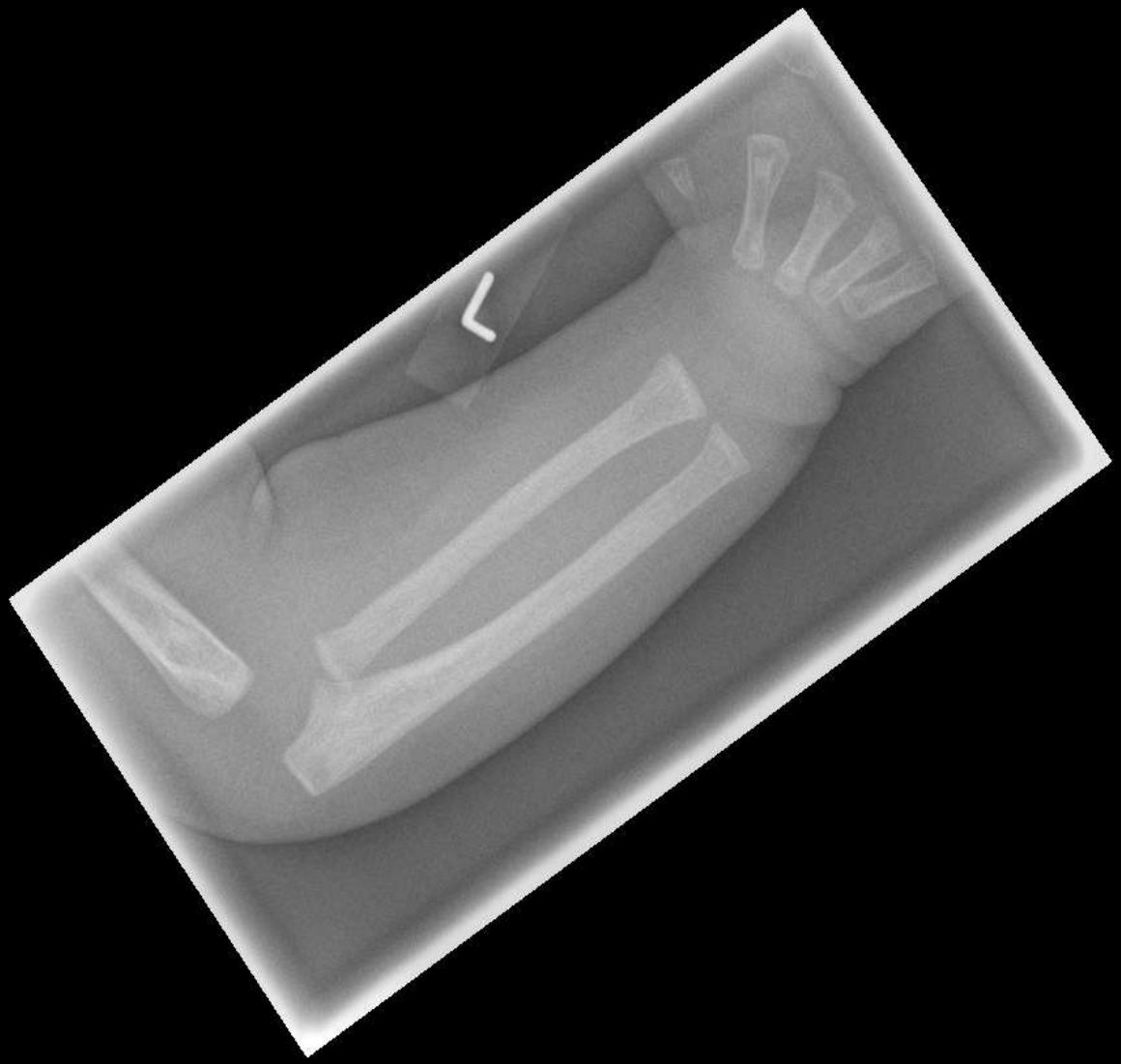
Radial Longitudinal Deficiency

- Spectrum anomalies
  - Bayne Classification
- 1:55,000 live births
- Frequently bilateral & asymmetrical
- Associated conditions
  - VACTERL
  - Holt-Oram
  - Fanconi's anaemia
  - TAR

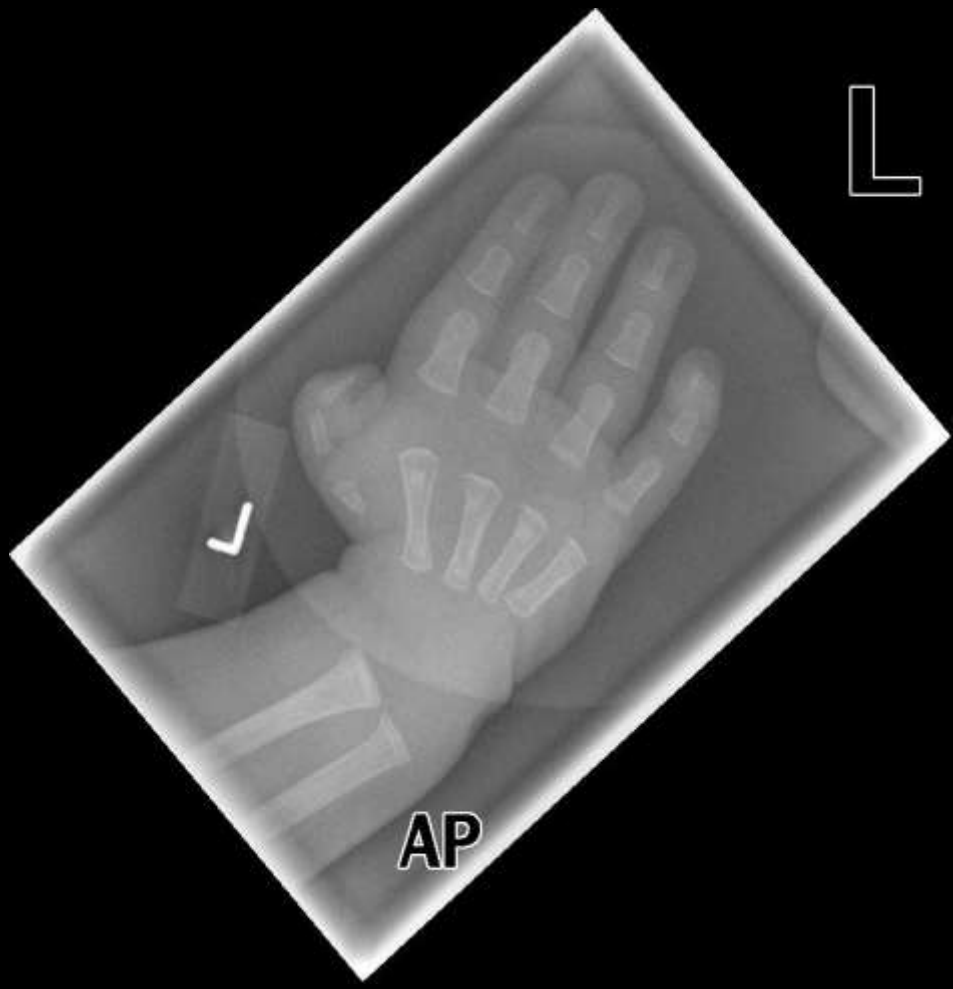
Radial Longitudinal Deficiency Classification

- Type N: Isolated thumb anomaly
- Type 0: Deficiency of carpal bones
- Type I: Short distal radius
- Type II: Hypoplastic radius in miniature
- Type III: Absent distal radius
- Type IV: Complete absent radius
- Type V: Complete absent radius and manifestations in the proximal humerus

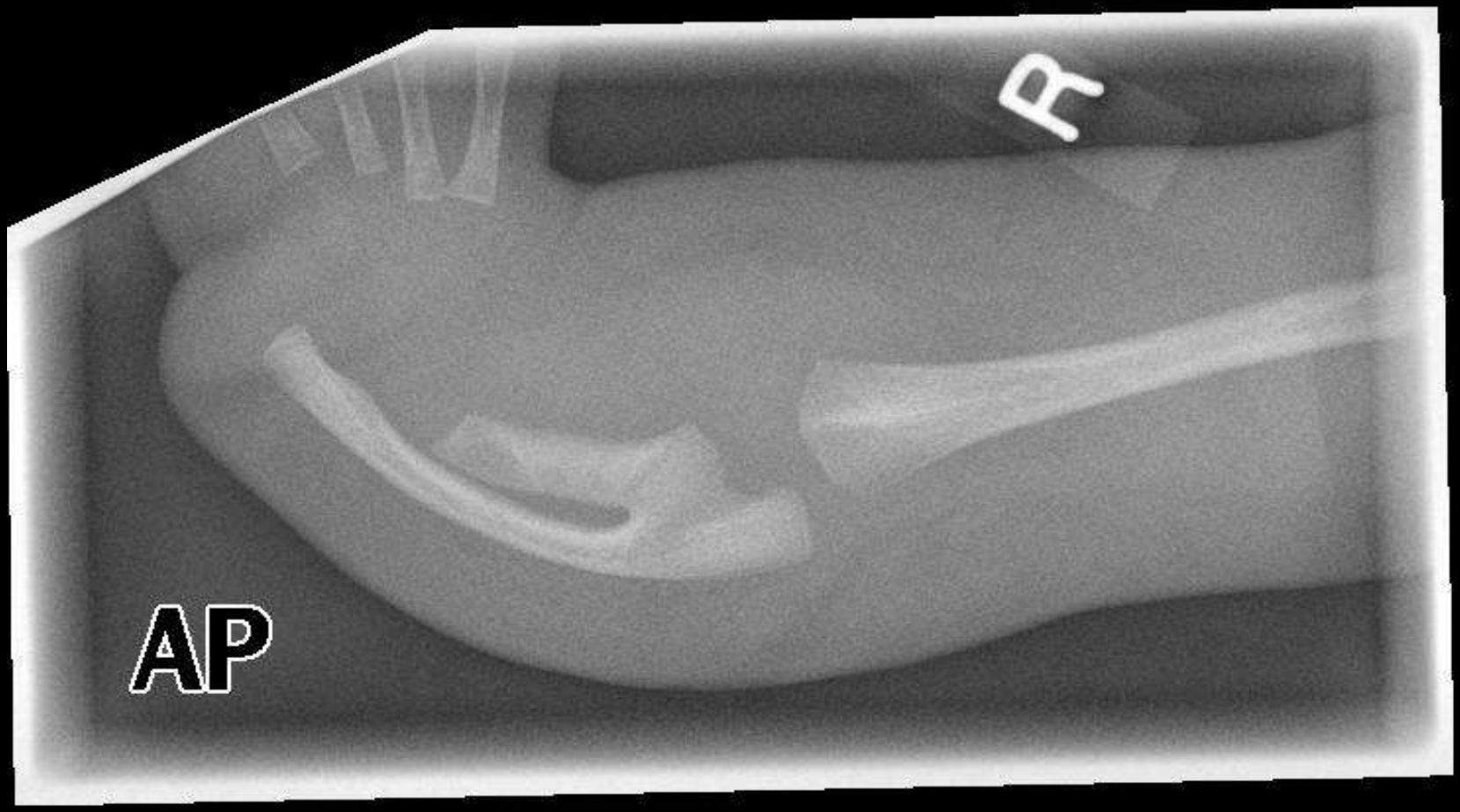
Type I: Short Distal Radius



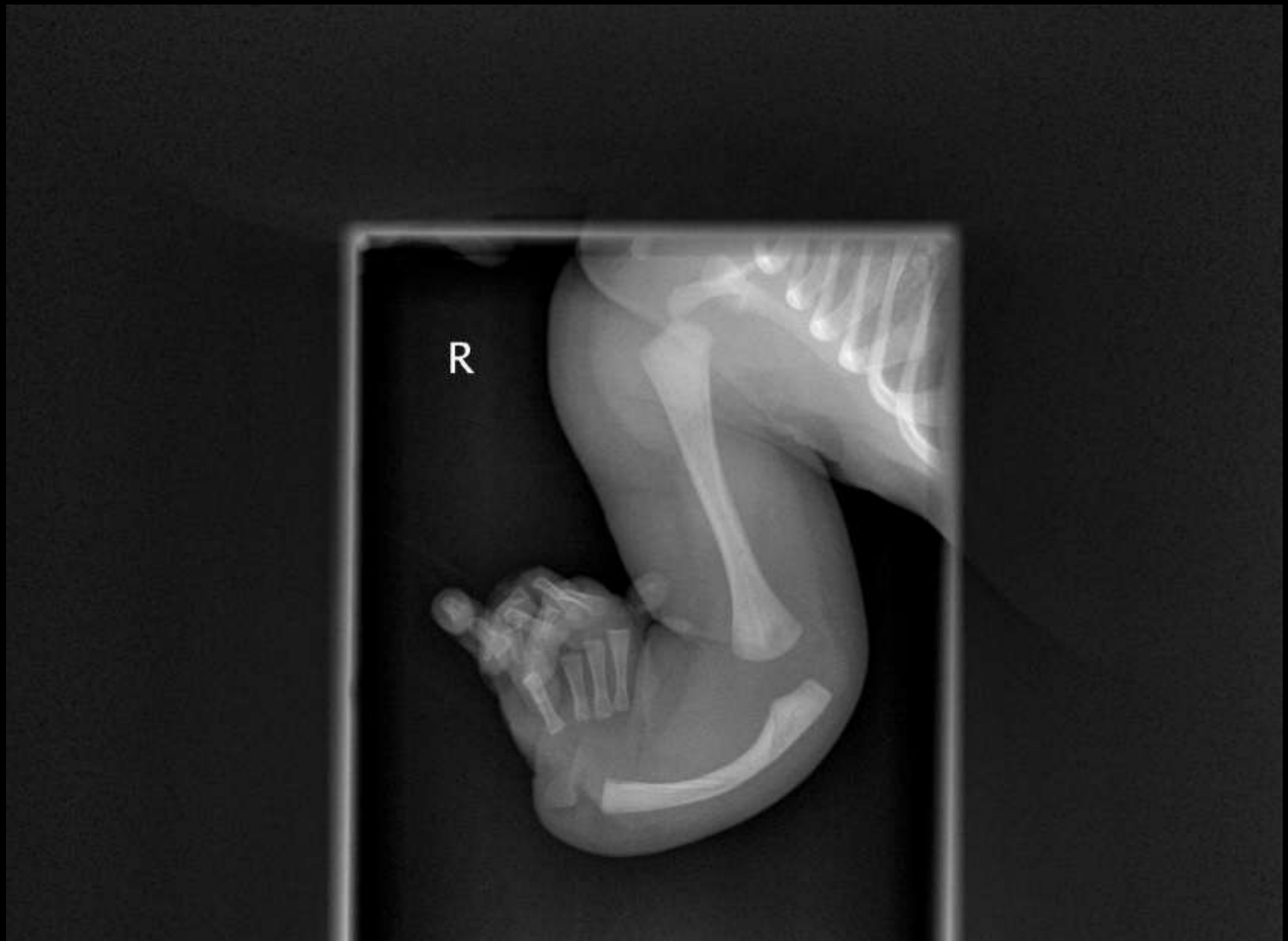
Type II: Hypoplastic Radius



Type III: Partial Absence of Radius



Type IV: Total Absence of Radius



Clinical Spectrum

- Spectrum radial forearm
- Hypoplasia bone, muscles,

  tendons, ligaments, nerves,

  blood vessels
- Aberrant/absent radial wrist extensors/thumb extensors
- Absent radial nerve, median nerve present
- Absent radial artery

Clinical Spectrum

Associated anomalies

- Hypoplasia scapula, clavicle, humerus
- Proximal radioulnar synostosis
- Radial head dislocation

More common with more severe deficiency

Radial Longitudinal Deficiency

Management

- Genetics referral
- Stretching & splinting
- Distraction device
- Centralisation/ Radialisation
- Thumb reconstruction
- Ulnar lengthening

Management

- Aim- position hand in space & maximise hand function
- Functional adaptation often good despite poor aesthetic appearance
- Individualised treatment
  - Realistic expectations

Management

- Correct radial deviation
- Balance wrist on forearm
- Maintain wrist and finger movement
- Promote growth
- Enhance appearance

Management

- Contraindication surgery
  - Anaesthetic risk
  - Lack of elbow movement
  - Adults with established functional patterns

Outcome

- Recurrance deviation & stiffness common
  - Stiff & straight v flexible & deviated
- Centralisation improve appearance not function

Hypoplastic Thumb

- Incidence 1:100 000 live births
- > 50% bilateral
- 60% associated with Radial Dysplasia
- > 75% of children have associated abnormalities
  - VACTERL association
  - Holt-Oram Syndrome
  - Fanconi's anaemia

Hypoplastic Thumb

Modified Blauth classification

I Minimal shortening and narrowing

II Thumb-Index web space narrowing/ Hypoplastic intrinsic thenar muscles/ MCPJ instability

III A Type II features, plus extrinsic tendon abnormalities/ Hypoplastic metacarpal/Stable carpometacarpal

III B Type II features, plus extrinsic tendon abnormalities/Partial metacarpal aplasia/Unstable carpometacarpal

IV Floating thumb

V Absent thumb

Type I

- Undersized normal components
- Fails reach beyond mid-point proximal phalanx
- Mild hypoplasia thenar muscles
- No significant functional deficit
- Leave alone

Type II

- Tight 1st web space
- Hypoplastic or absent thenar muscles
- Unstable MCPJ

Type III

- Hypoplastic 1st MC
- Absent intrinsic & extrinsic thumb musculature
- IIIA – Stable CMCJ
- IIIB – unstable CMCJ

Type IV

- Floating thumb or pouce flottant
- No muscles
- Rarely tendons
- Few rudimentary bones

Type V

- Absent thumb

Treatment Aims

- Stable
- Mobile
- Opposable
- Good longitudinal alignment
- Adequate 1st web
- Adequate size
- Cosmesis

Type II

1. 1st web release
2. Opponensplasty
    - Huber transfer-ADM
    - FDS (ring) transfer
3. MCPJ instability
    - UCL reconstruction
    - Chondrosesis

Release 1st web

- 4 flap Z-plasty
  - ABCD
  - CADBury

Type IIIA

- Stable CMCJ
- As Type II & extensor tendon transfer

Type IIIB

- Unstable CMCJ
- Pollicisation

Type IV/V

- Early amputation floating thumb
- Pollicisation

Type V

- Pollicisation

Pollicisation

- Reposition index finger to act as thumb
- Metacarpal divided & rotated into pronation & abduction
- Metacarpal head fixed to base
- 1st dorsal & palmar interosseous muscle become abductor & adductor

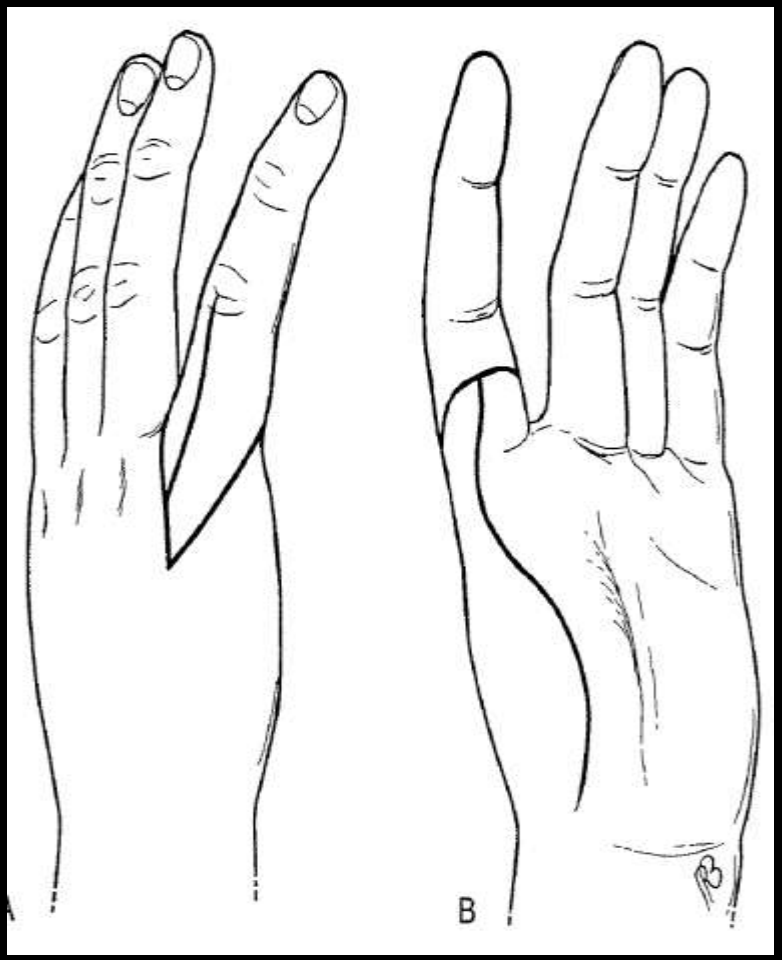
STEPWISE APPROACH TO POLLICIZATION

Step	Rationale	Technique
Gentle exsanguination	Visualization of the vasculature	
Skin incision	Creation of a thumb-index web space, adequate exposure of all structures	Exact design
Isolation of the palmar neurovascular bundles	Preserve sensibility and circulation to the index finger	Loupe magnification, meticulous dissection, and direct exposure
Microdissection of the common digital nerve	Mobilize the nerve for pollicization	Intrafascicular dissection
Ligate the proper digital artery to the radial side of the long finger	Maintain the proper digital artery with the index and long fingers perfused with the ulnar proper digital artery	Suture or ligature clip. Ligature clip preferred to allow visualization throughout the procedure
Release the first annular pulley to the index finger	Prevent buckling of the flexor tendons	
Elevation of dorsal skin from the volar incision with preservation of the dorsal veins	Veins have filled with blood to facilitate exposure and preserve venous egress	
Incise the dorsal skin	Veins have been protected	
Extensor tendons freed from adjacent finger	Ensure appropriate line of pull to the index finger pollicization	
Extensor and flexor tendons are not shortened	Shorten themselves over time	
Incise the intermetacarpal ligament	Allow better access to the palmar interosseous muscle	
Elevation of the first dorsal and palmar interossei muscles from the index metacarpal and MP joints	The metacarpal will be excised and muscles advanced to the PIP joint	Sharp dissection
Release the first dorsal and palmar muscles with a strip of the extensor hood	Tendons are sutured to lateral bands about the PIP hood	
Identify and tag the radial and ulnar lateral bands about the PIP joint	Identification before bone resection is easier and safer Later transfer of the first dorsal and first palmar tendons	Pull on the lateral band until the desired function is evident and tag the band with a suture
Shorten the index finger by removing the majority of the metacarpal bone, including physis ablation	Index too long for a thumb; must prevent continued metacarpal growth	Fine bladed saw to cut the metacarpal perpendicular to the bone through its metaphyseal portion Distal cut through the physis (epiphysiodesis)
Reposition the index MP joint into hyperextension	A normal index MP joint hyperextends and a normal thumb CMC joint does not hyperextend	Fixation of the index MP joint into hyperextension with a nonabsorbable suture placed through the epiphysis and dorsal capsule
Double-ended Kirschner wire driven through the metacarpal physis and out the PIP joint or end of the thumb	Easy fixation; provides a joystick for thumb positioning	Double-ended Kirschner wire to allow retrograde and antegrade passage
Align the index finger into the thumb position	Position into 45 degrees of abduction and between 100 and 120 degrees of pronation	The metacarpal epiphysis is placed volar to its remaining base to bring the thumb out of the palm
Tendon transfer to restore intrinsic function to the pollicization	Maximize function in grasp and pinch	First dorsal interosseous sutured into the radial lateral band and the first palmar interosseous sutured into the ulnar lateral band
Inset skin	Adequate coverage without scar in the thumb-index web space	Skin inset along the palmar aspect of the pollicization while avoiding neurovascular bundles Inset the web space skin Trim any excess skin
Deflation of the tourniquet and meticulous postoperative dressings	Ensure circulation, protect the pollicization, decrease chances of inadvertent removal of dressings	Bulky hand dressing with a long arm splint

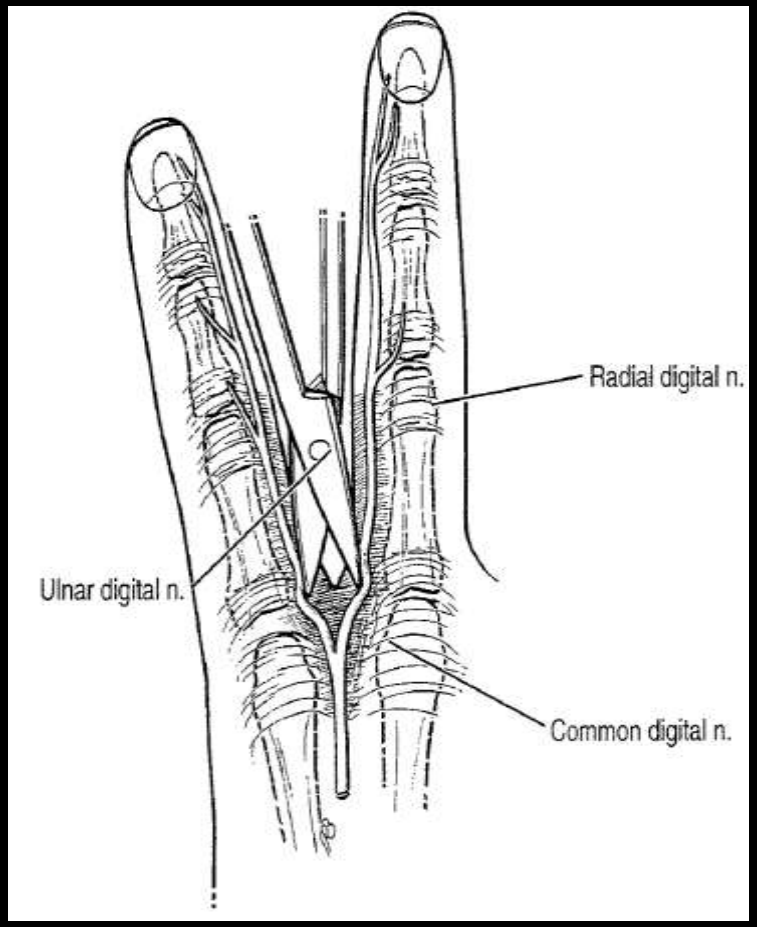
CMC, carpometacarpal; MP, metacarpophalangeal; PIP, proximal interphalangeal.

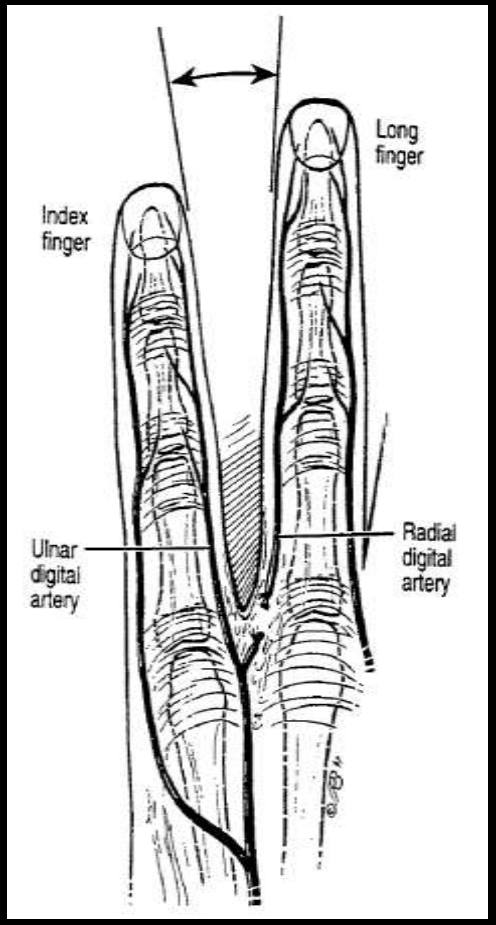
Table 41.3 Stepwise Approach to Pollicization

Pollicisation – skin incision

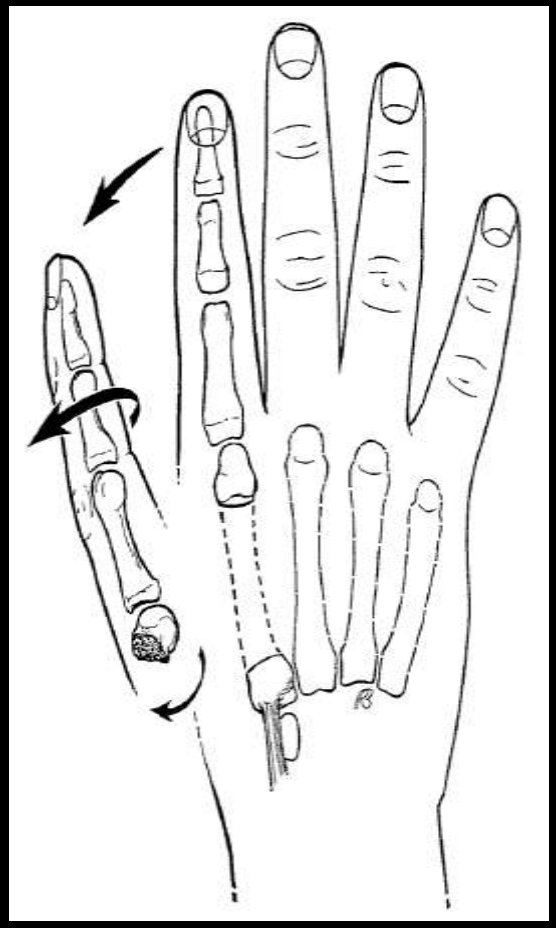


Neurovascular dissection





Skeletal adjustment



Syndactyly

- Variable fusion soft-tissue/skeletal elements digit
- 1:2000
- Majority sporadic
  - FH 10-40%
- Syndromal

  eg Apert's syndrome

CLASSIFICATION

Degree of webbing
Complete : to fingertips

Incomplete : proximal to fingertips

Bone fused or not
Simple: soft tissue only

Complex: osseus syndactyly

NB Xray

Complicated

Classification

- Complete/Incomplete

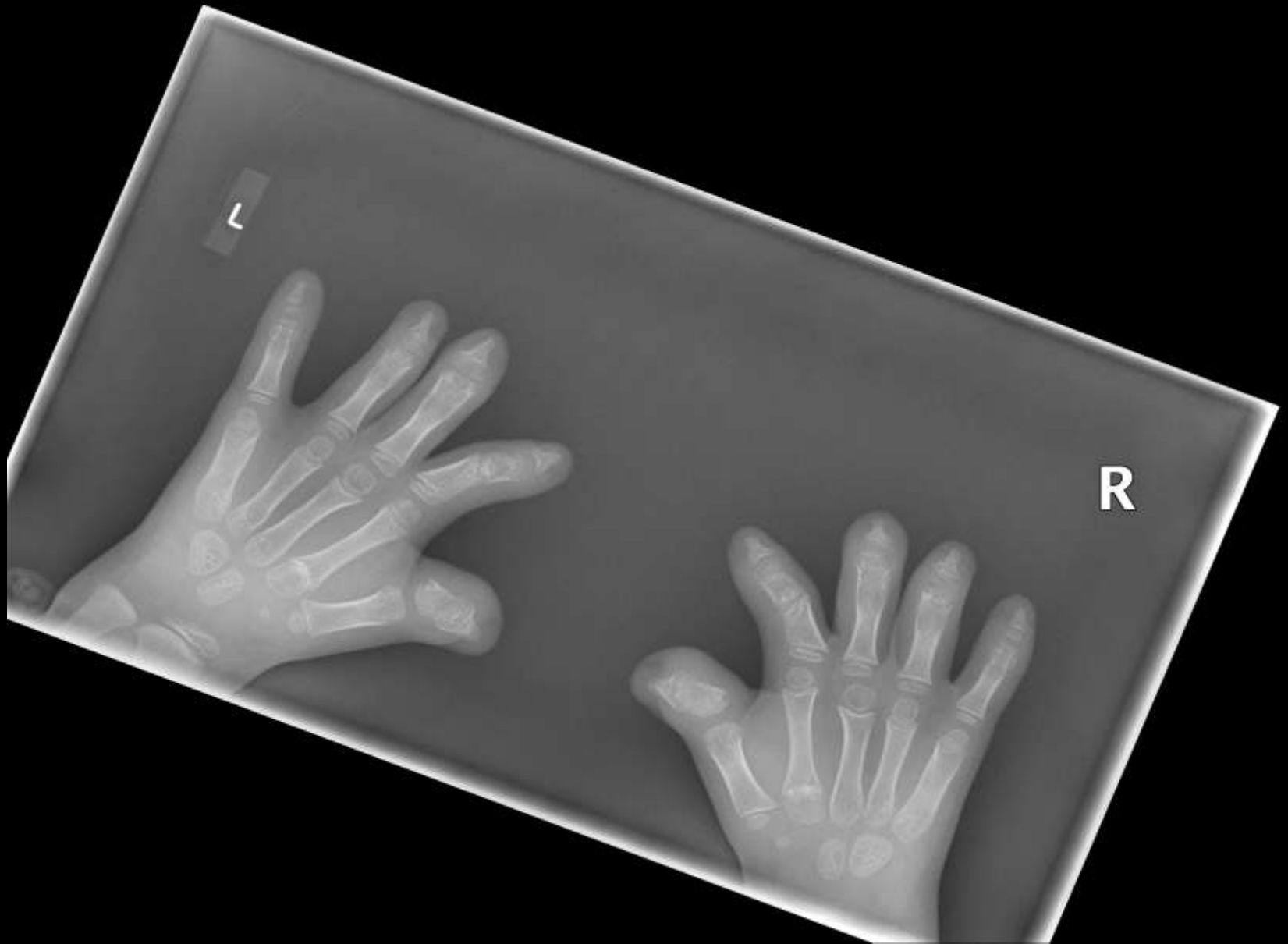
- Simple/Complex/ Complicated

COMPLEX SYNDACTYLY


LT

POLYSYNDACTYLY

APERT SYNDROME



POLANDS SYNDROME

Poland's Syndrome

- Chest wall deformity may include
  - hypoplasia breast
  - aplasia pec minor/LD
  - skeleton anomalies
- Hand deformity can include
  - Brachydactyly
  - Hypoplasia hand
  - Syndactyly

MANAGEMENT

Why treat?

When to treat?

What to do?

PLANNING OF TREATMENT

Patient factors

Surgical considerations

Number of webs/which webs

Simple/complex

Hypoplasia/instability

NORMAL WEB FINGERS

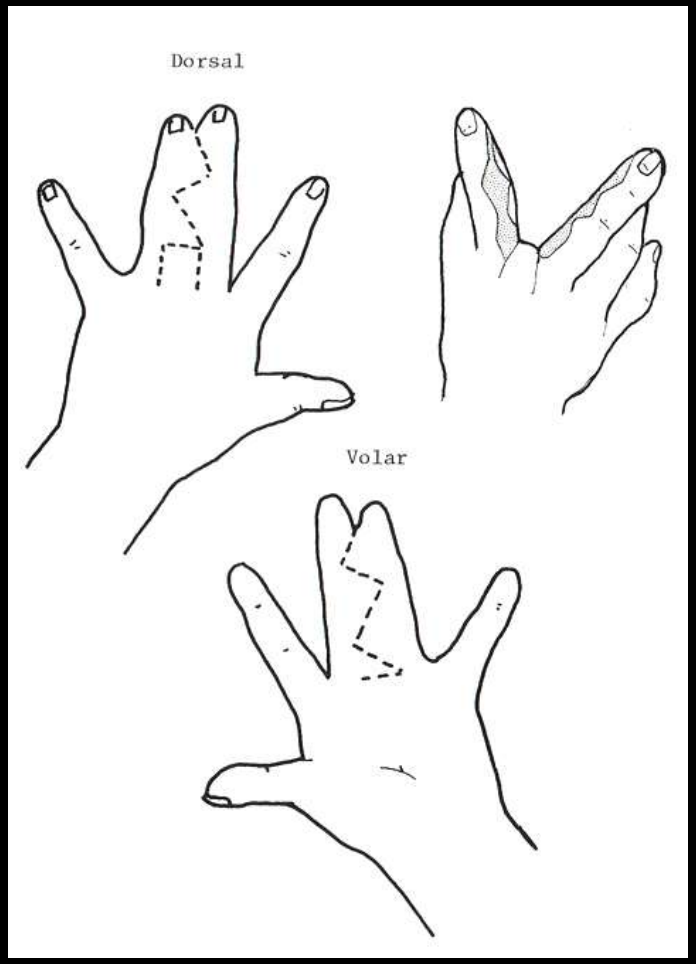
- U-shaped
- Dorsal palmar slope
- Mid-point proximal phalanx

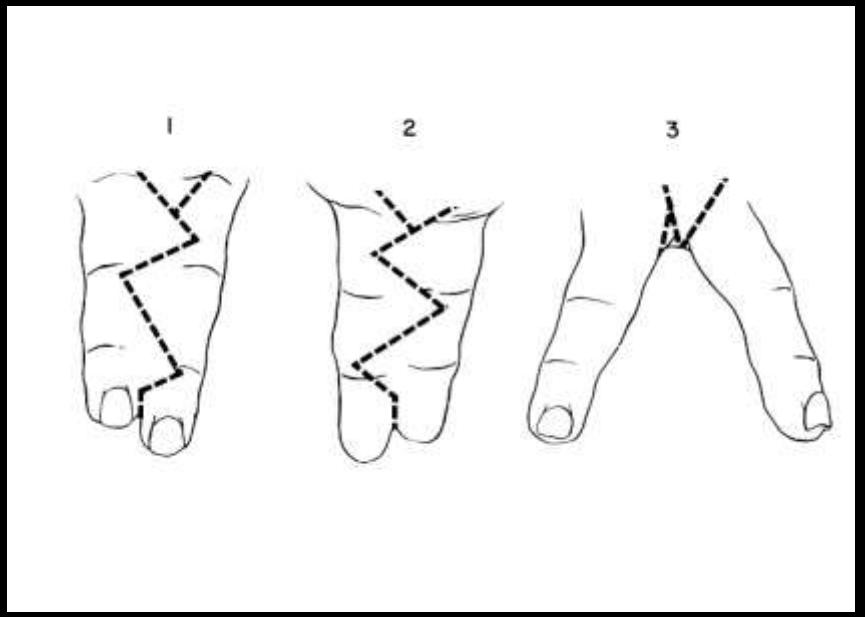
Web flap design

Placement of flap

Dorsal skin more mobile

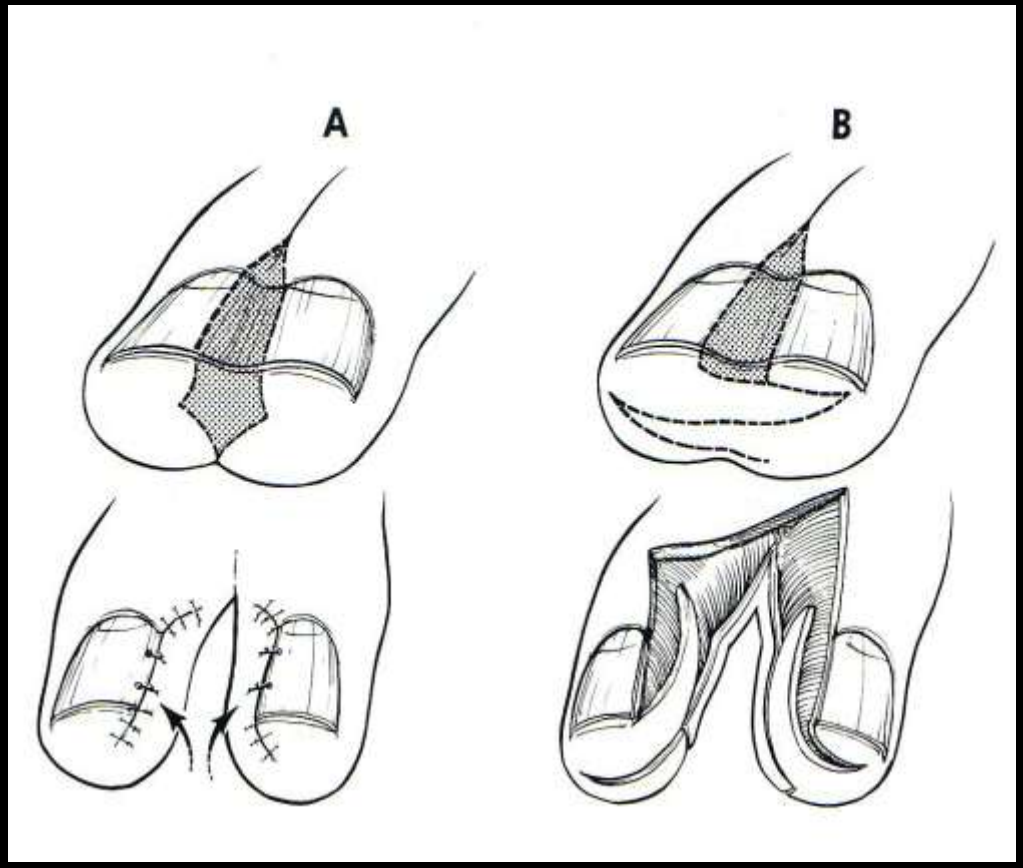
SKIN FLAPS FOR WEBS AND FINGERS +/- SKIN GRAFT





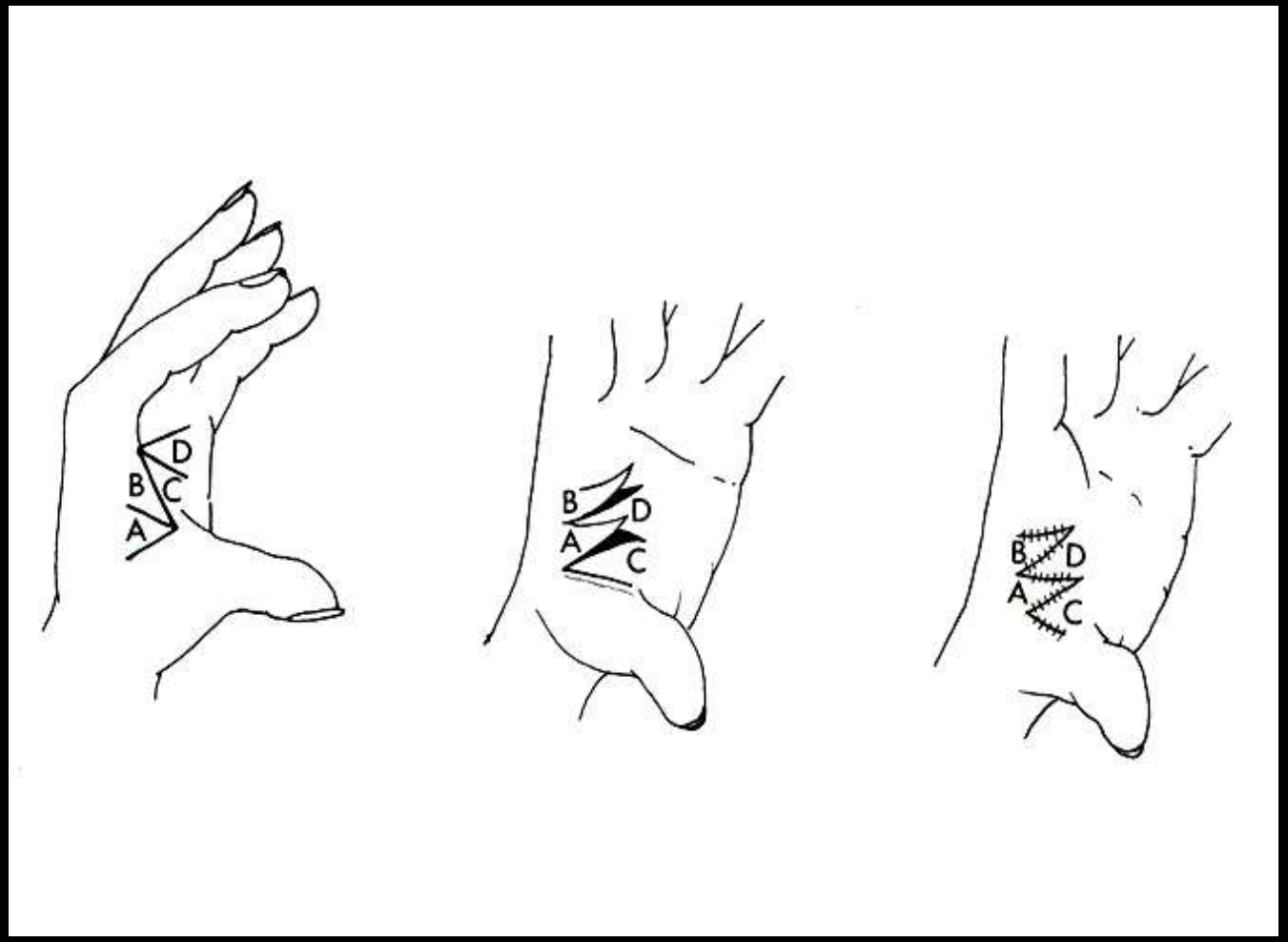
COMPLICATED SYNDACTYLY MAY REQUIRE MORE COMPLICATED FLAPS

FINGERTIPS



1st WEB RELEASE

4 FLAP Z-PLASTY



SKELETON

Separate fused bones

Remove extra bones

Osteotomy to improve longitudinal alignment

SKIN GRAFT DONOR SITES

- Expendible
- Concealed
- Pigmentation
- Hair

Groin

Inner arm

Abdomen

Prepuce

Full thickness/thick split thickness

OTHER TECHNIQUES

- Tissue expansion – not successful
- Soft tissue distraction
- Release without skin graft

Polydactyly

- Radial (pre-axial)
- Ulnar (post-axial)
- Central

Overall incidence

- 1:3000 Caucasians
  - Radial more common
- 1:300 African Americans
  - Ulnar more common

Ulnar Polydactyly

- Type A
  - Digit well developed & connected via bone
  - May have associated syndromes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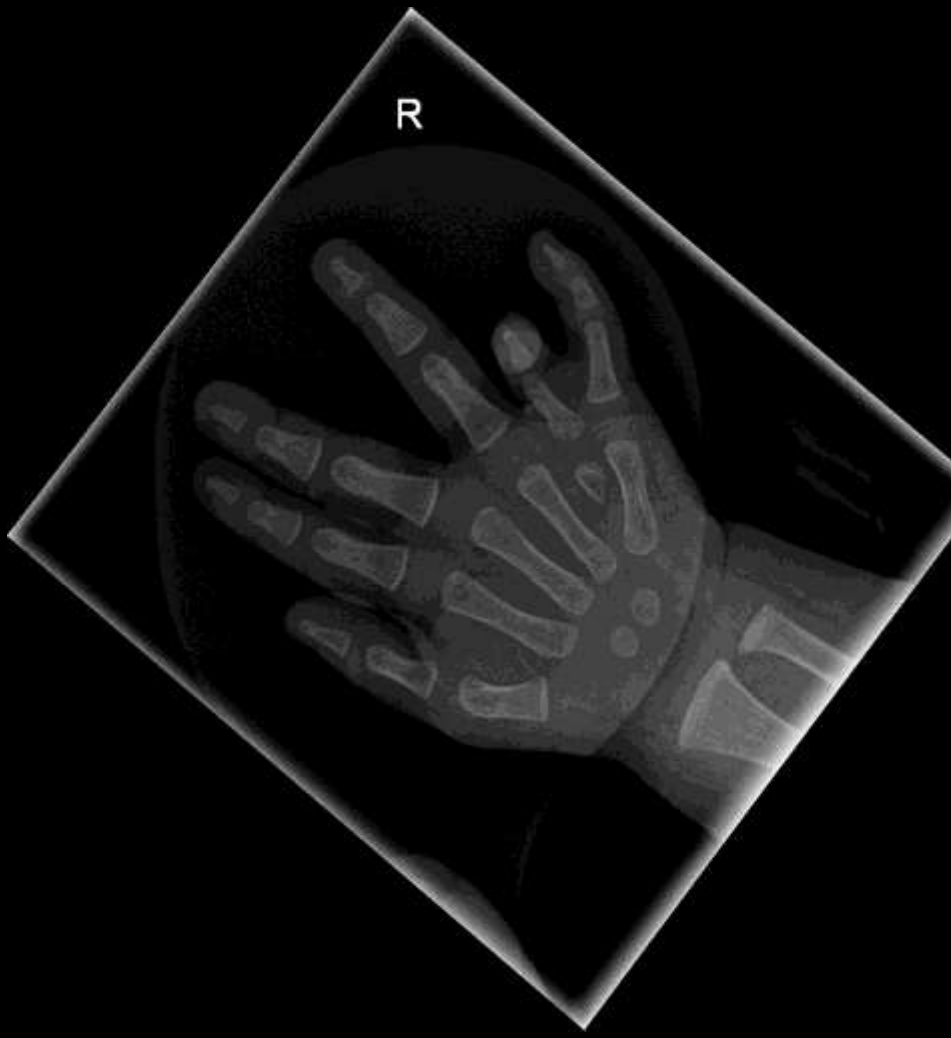
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Ulnar Polydactyly

- Type A
  - Surgical treatment based on type present

Ulnar Polydactyly

Type B

- Rudimentary & pedunculated digit, loosely attached
- More common African Americans

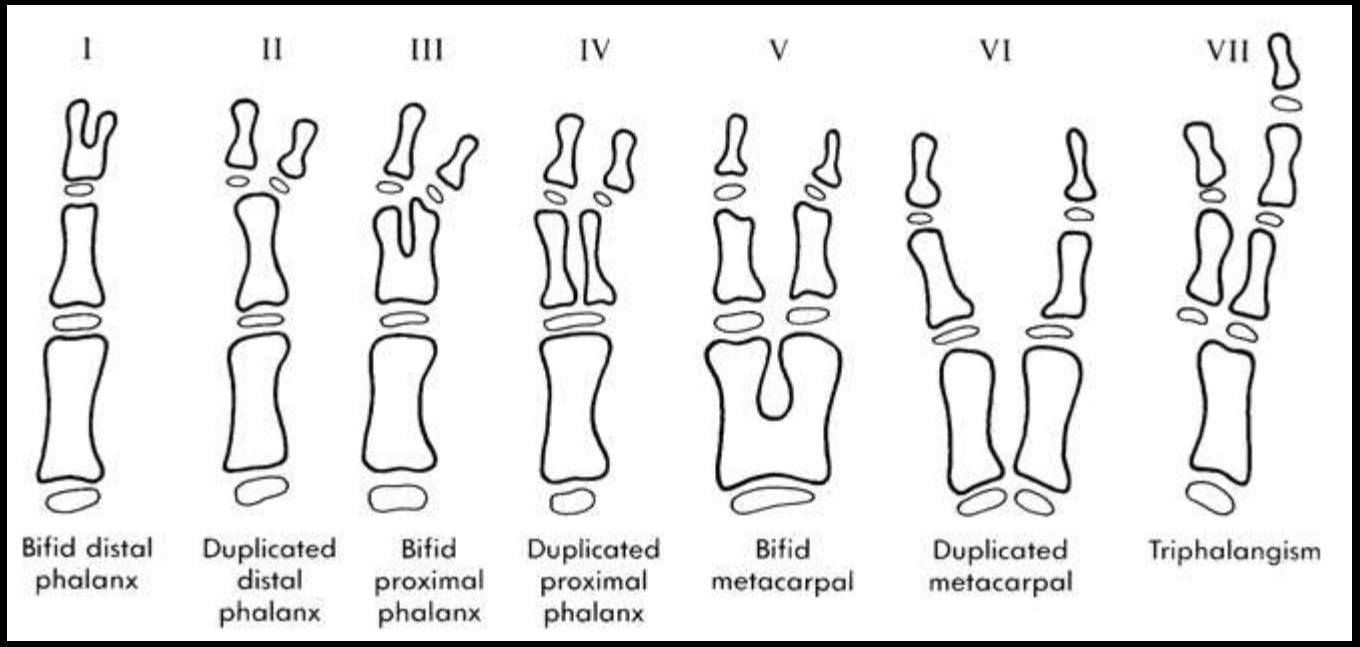
Ulnar Polydactyly

- Type B
  - Surgical excision LA in early neonatal period
  - Liga clip/ligate
  - GA excision

Radial Polydactyly

- Incidence 1:1000
- Wassel classification
  - Based x-ray appearance
  - Simplification of overall anatomy
- Wassell I-VI
  - Usually sporadic & unilateral
  - IV most common
- Wassell VII
  - Frequently AD & bilateral

Wassel Classification



Focus on radiographically visible skeleton
Disparate patients within individual groups

Treatment Aims

- Stable
- Mobile
- Opposable
- Good longitudinal alignment
- Adequate 1st web
- Adequate size
- Cosmesis

Radial Polydactyly

Management

- Select dominant thumb
- Consider structures anatomically
- Osteotomy (if required)
- Reconstruct collateral ligaments
- Re-balance tendons

Wassel IV

Ring Constriction Syndrome

- Variability in presentation
  - Annular constrictions limbs/trunk
  - Amputations
  - Acrosyndactyly
  - Oligodactyly
  - Talipes equinovarus
  - Nerve compression
  - Facial clefts
  - Body wall defects

Patterson Classification

Ring Constriction Syndrome

- Aetiology unclear
  - Intrinsic theory
  - Extrinsic theory

- Risk factors
  - Prematurity
  - Low birth weight
  - Maternal drug exposure

Pathogenesis

1965 Torpin
Premature rupture of amnion with passage of foetus into chorionic cavity. Close apposition of foetus to chorion with resulting postural deformities. Fibrous strings produced by mesodermic outer surface of amnion.

1989 Lockwood
Contested exogenous theory. Proposed a vascular aetiology.

Ring ConstrictionSyndrome Treatment

- Early surgery
  - Threatened ischaemia
  - Tethering digits
  - Neurological compromise

- Release bands/syndactyly
  - Z-plasty/straight line incision
  - Excise entire band
  - Staged surgery