CONGENITAL HAND - 2

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DYSPLASIA - Overgrowth

MACRODACTYLY:

- Not hereditary
- Very rare 0.2/1000
- PROTEUS syndrome , Olliers, Maffuci
- Most common a/w hypertrophy of nerve
- Median nerve hypertrophy

Flatts classification:

1. Gigantism & LIPO fibromatosis
2. " & NEURO fibromatosis
3. " & digital hyper OSTOSIS
4. " & HEMI hypertrophy

Surgery -

Arrest Growth - EPIPHYSODESIS - most common

- Single digit, once it reaches adult size of same sex parent

Reduce tissue - Debulking

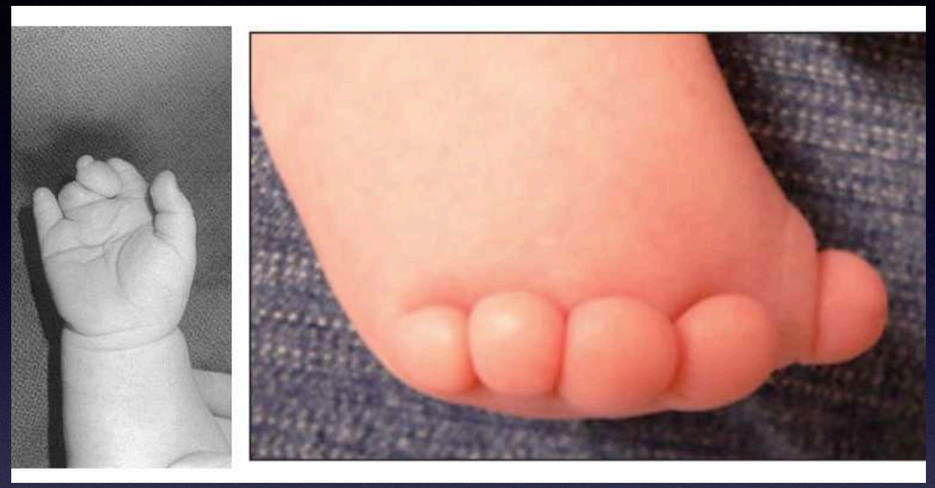
Deviation - closing wedge osteotomy

Thumb - MP arthrodesis

Ray amputation - last resort

DEFORMATION - CONSTRICTION BAND SYNDROME

- Amniotic band syndrome
- Streeter’s dysplasia
- Fingers and toes
- Digit proximal to band is well developed
- Distal NVD
- Multiple Z plasties



MADELUNG Deformity

- Radial deviation of hand
- Dyschondrosis of radial growth plate
- Secondary overgrowth of distal ulna
- Stiffness
- Bayonet appearance of hand

Madelung Deformity: Vickers Ligament

Findings

- Madelung deformity
- Ligament structure connecting lunate and TFCC to distal radius

Key points

- Madelung deformity treatment: Release the ligament

Operative??

Radial corrective osteomy
+/- distal ulnar shortening osteotomy

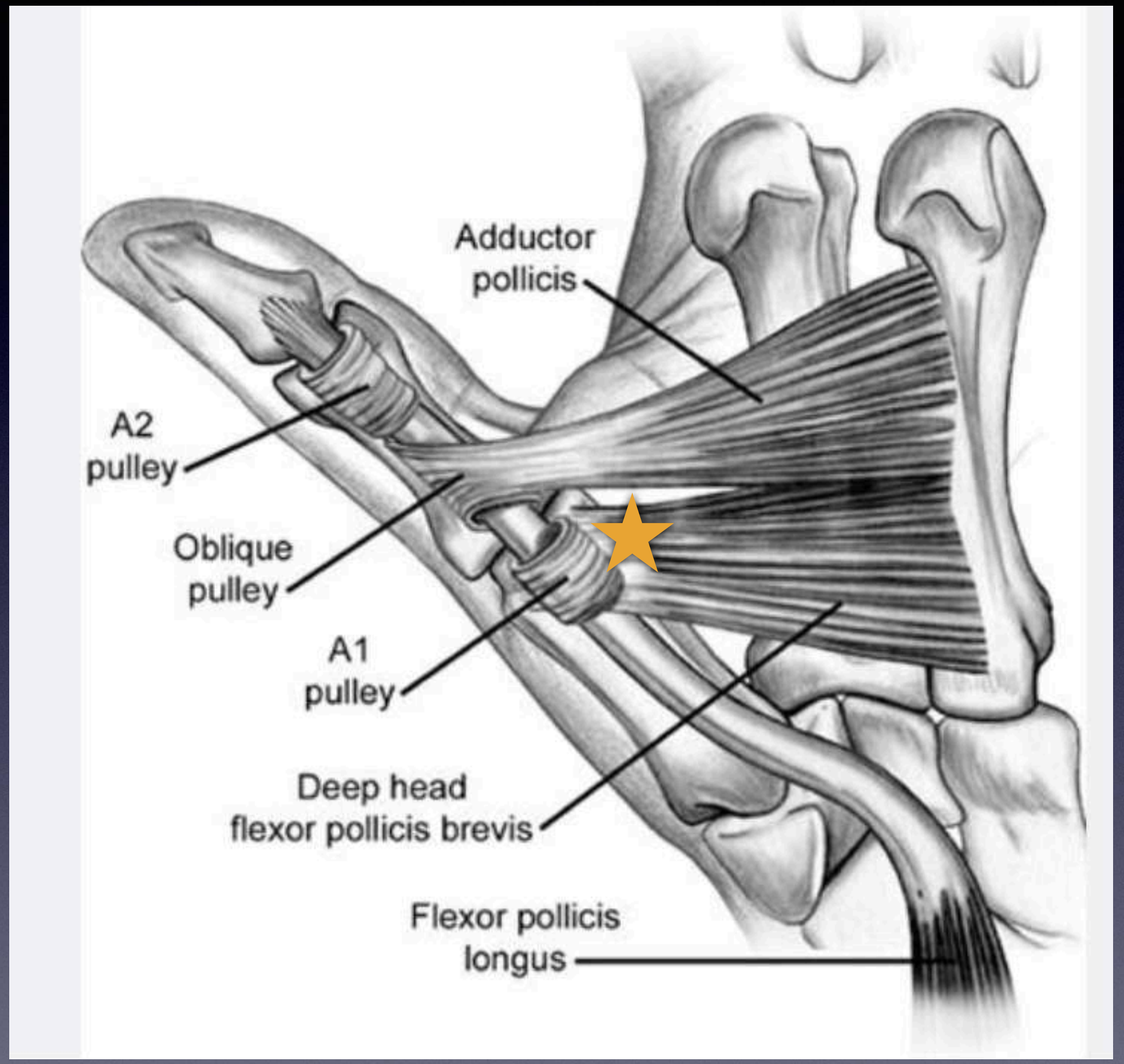
wrist pain or decreased range of motion
cosmetic deformity
functional limitations

DRUJ arthroplasty???

highly controversial
painful DRUJ instability and limited supination/pronation
significant deformity may require staged procedures

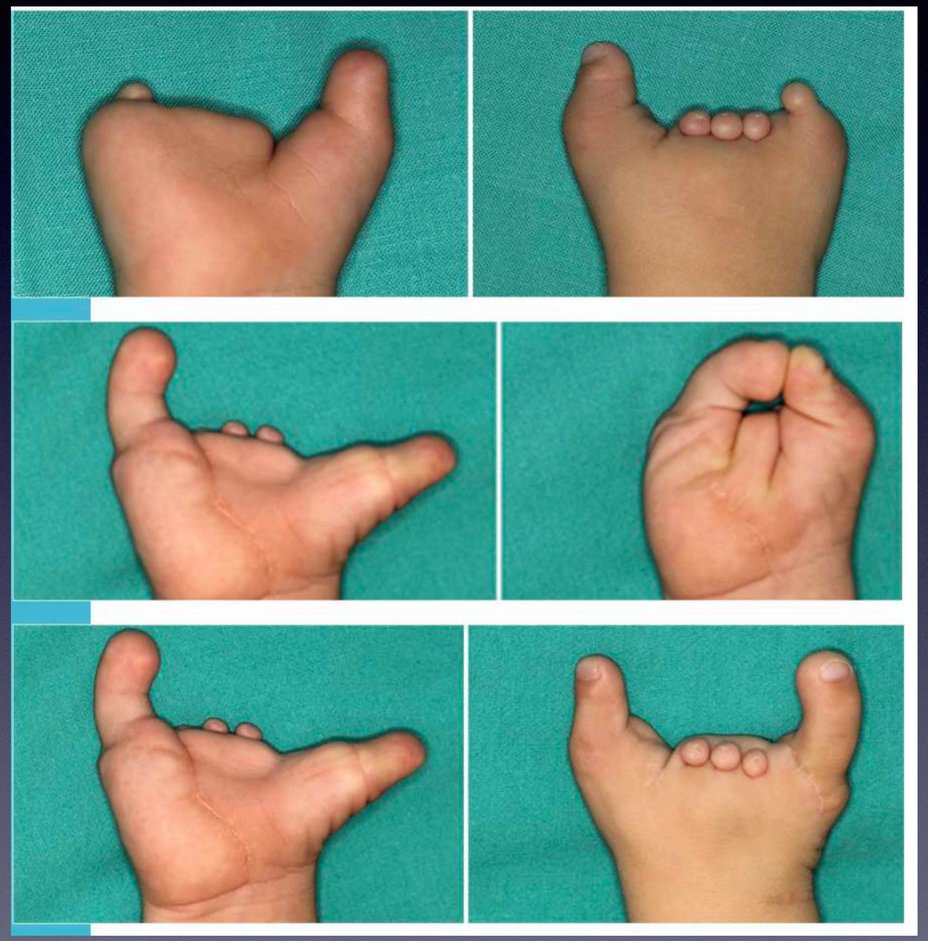
CONGENITAL TRIGGER THUMB

- Very common
- Flexion at IP
- Tenosynovitis of FPL
- A1 pulley
- Notta’s node proximal to A1 pulley
- Release of A1 pulley



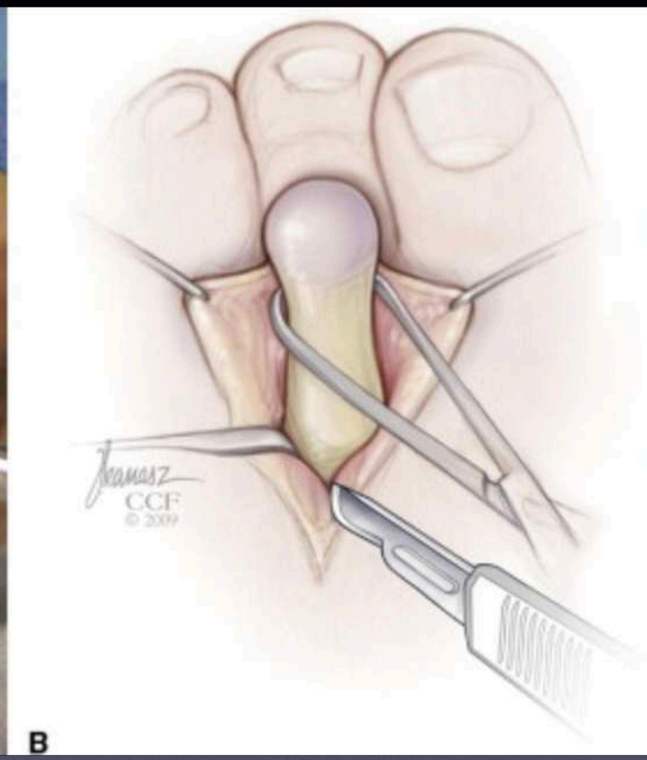
FAILURE in hand plate formation & differentiation

- Transverse arrest - humerus, forearm
- carpal and metacarpal
- BRACHYDACTYLY: (metacarpal)
- Brachymesophalangy
- Brachymetacarpia
- Symbrachydactyly (atypical cleft hand)



Management:

- Deepen web space (phalangization)
- Free phalangeal transfer from toe
- Distraction lengthening +- bone graft
- Vascularised toe transfer



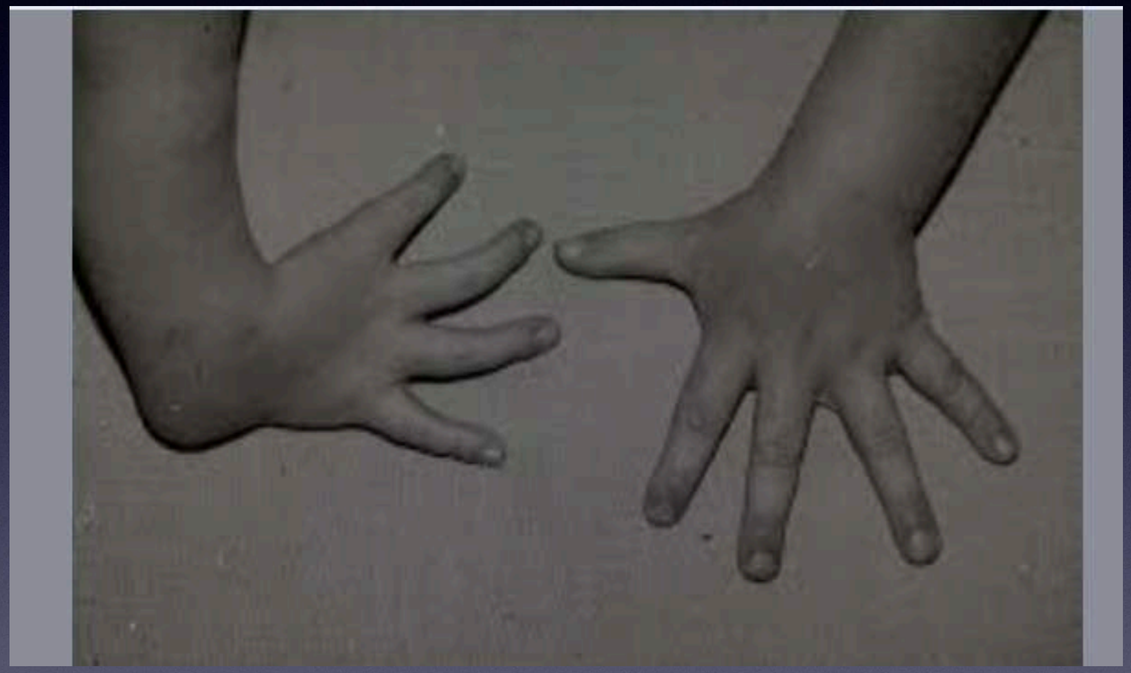
Phocomelia

- Failure in axis formation & differentiation

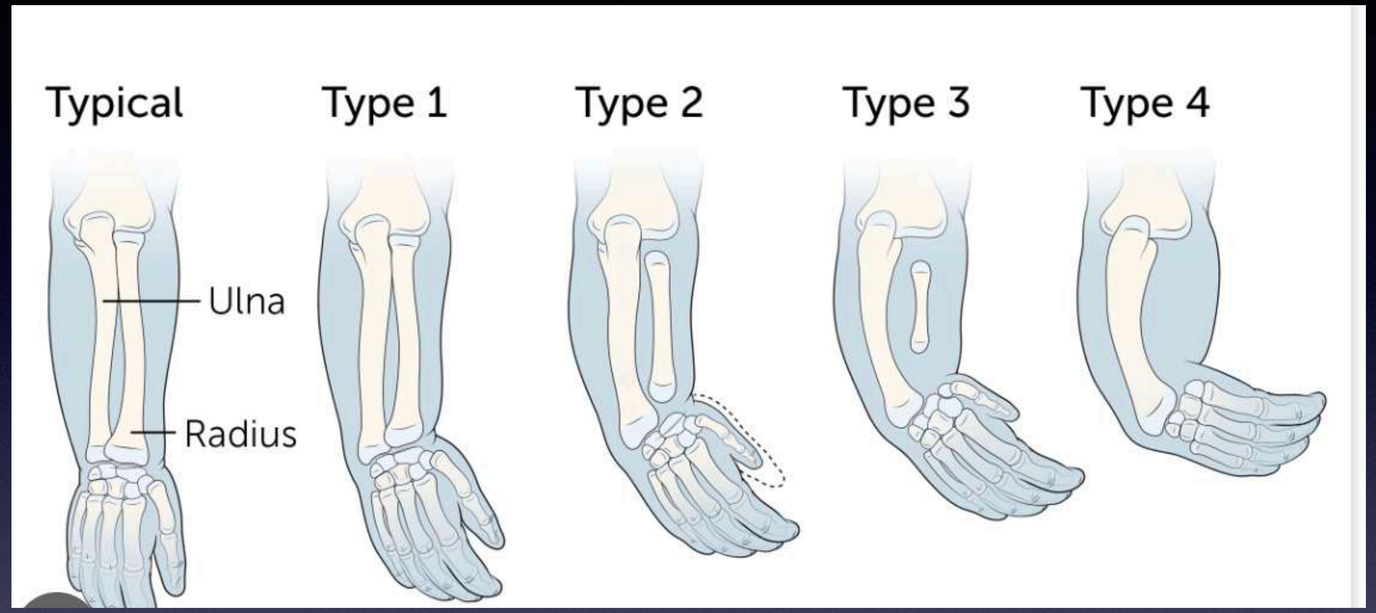
- Intercalated deficiency
- Seal limb
- Distal structures present, long bones absent
- Roberts syndrome
- Thalidomide
- Orthotics/ prosthetics

Radial A/ hypoplasia

- Longitudinal radial deficiency
- Radial club hand
- Preaxial border
- Hand deviated radially,
- flexed & pronated at forearm



Bayne & Klug :



Radial longitudinal deficiency
Short distal radius
Hypoplastic radius
Partial absence of radius
Total absence of radius

a/w thumb hypoplasia

Syndromic - Fanconi, TAR, Holt Oram

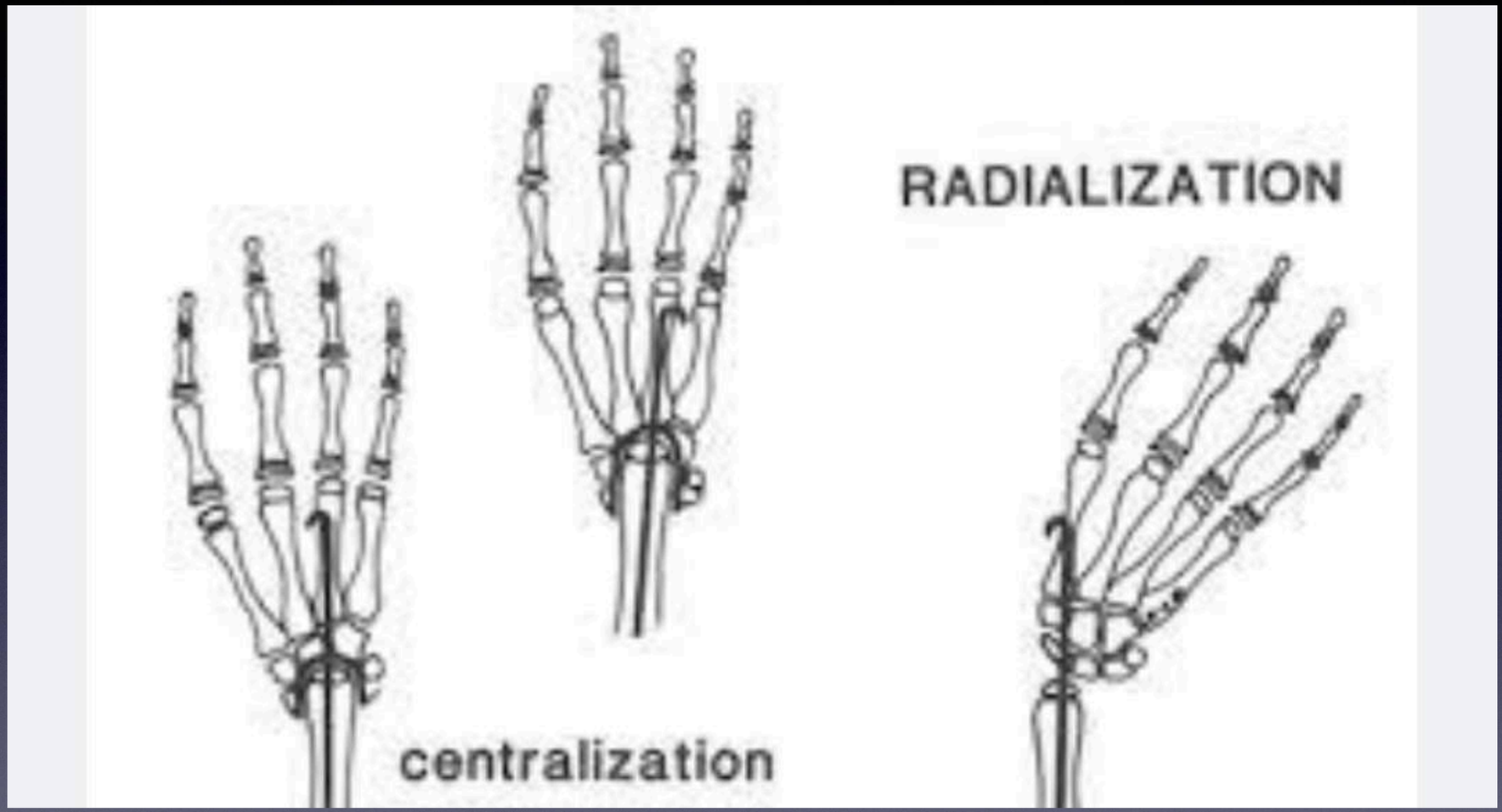
Management?

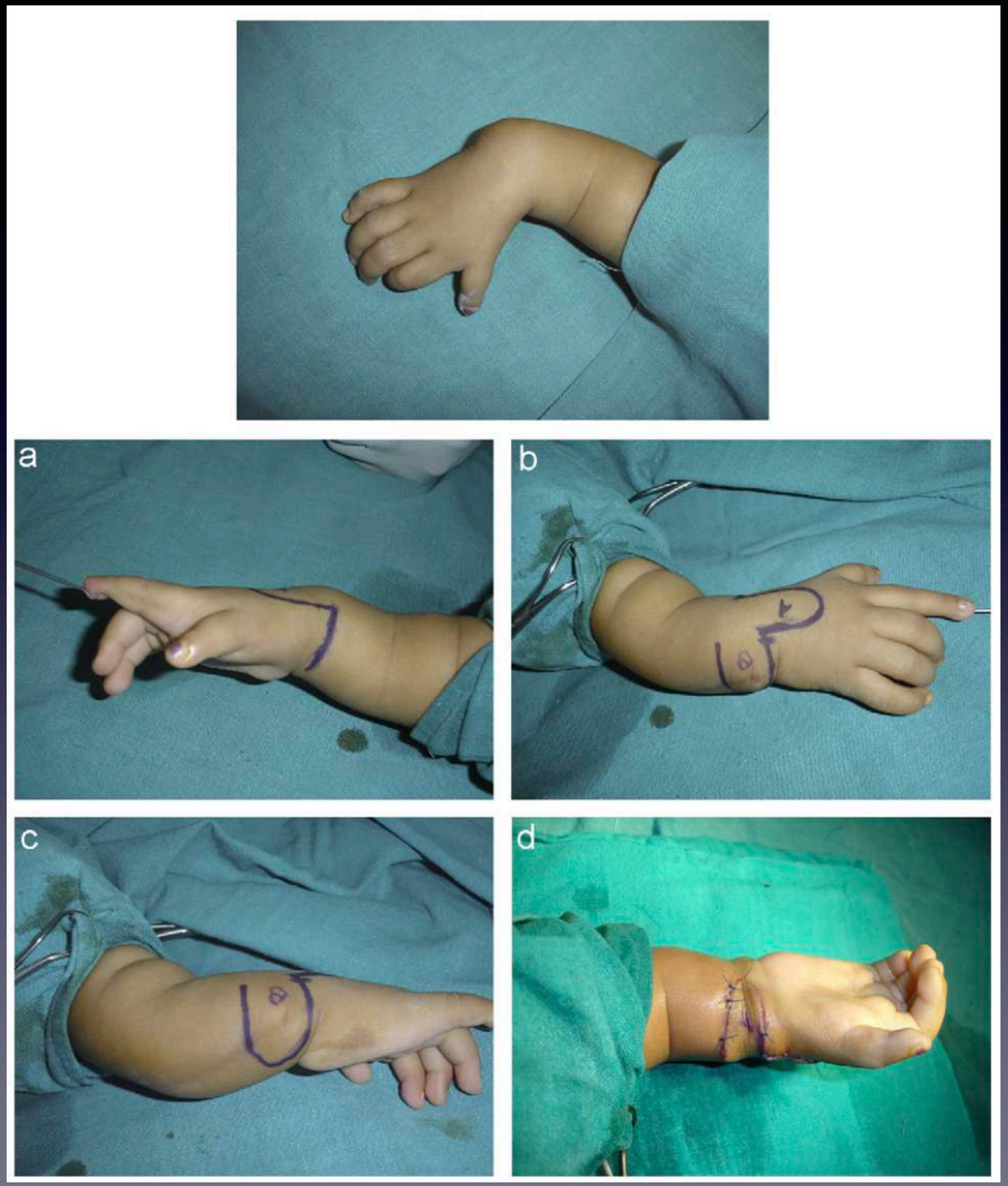
- Splintage
- Tendon balancing
- Convert type 2-3 to type 4
- Excision of radial analge

- Mx of thumb hypoplasia
- Pollicization
- Distraction lengthening
- Centralisation - stability
- Radialisation - mobility

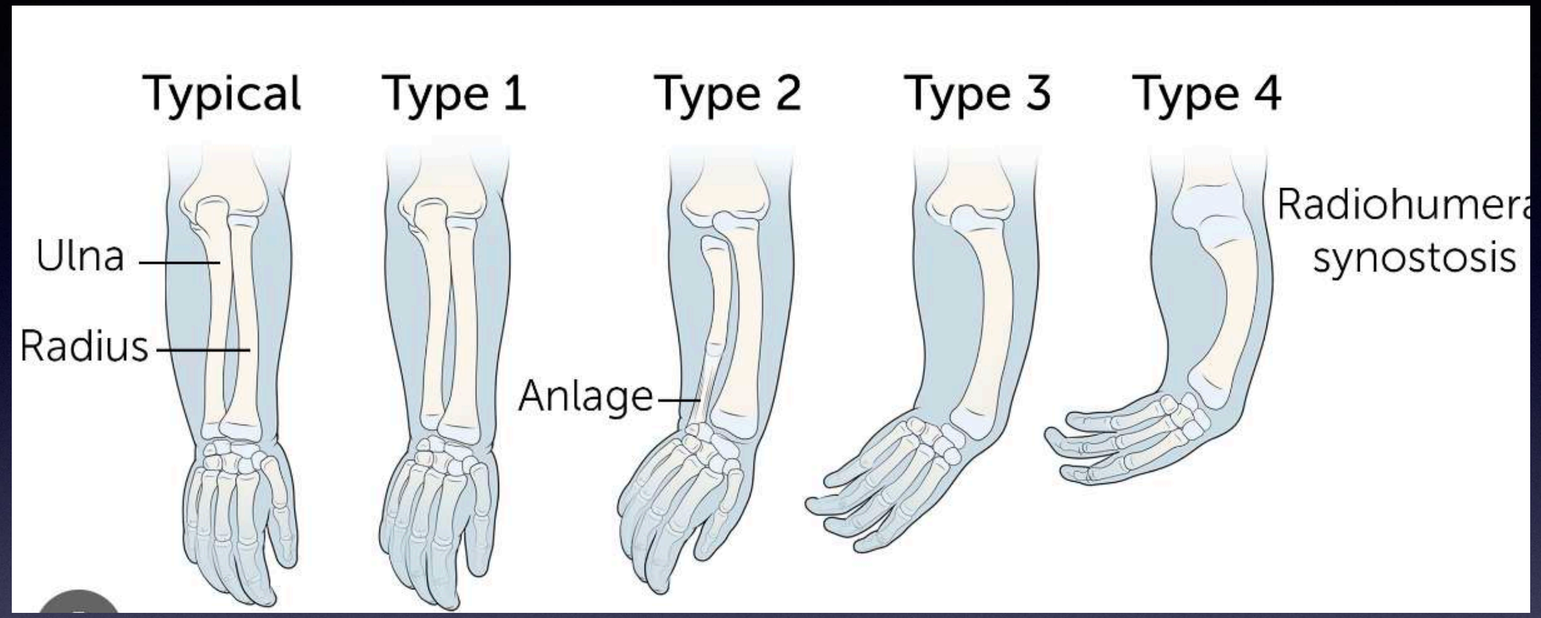
CONTRAINIDICATION?

STIFF ELBOW





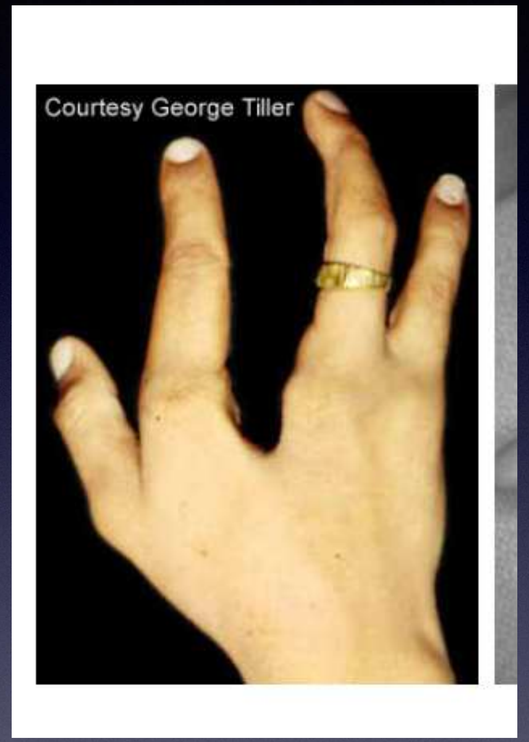
Ulnar dysplasia - rarest



- Splinting
- Distraction
- One bone forearm

Central ray deficiency

- Cleft hand
- Functionally great, aesthetically poor
- AER, apical ectodermal ridge
- Missing 3rd ray
- Male > female
- Autosomal dominant 70%



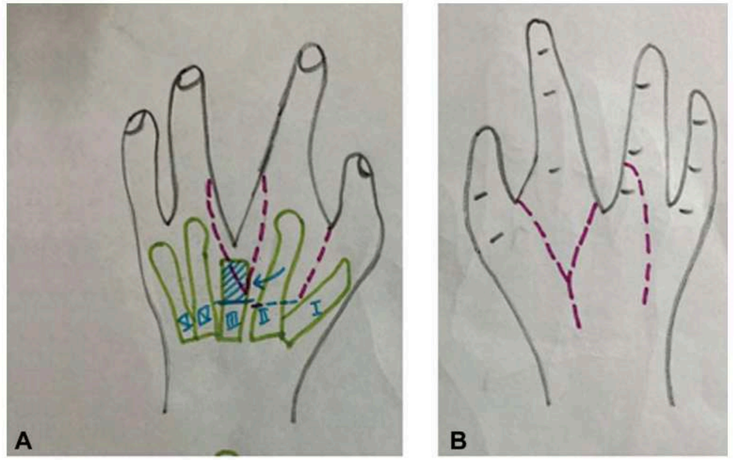
Typical	Atypical
V-shaped	U-shaped
Bilateral	Unilateral
Family heredity	Sporadic
Syndactyly is common	Syndactyly is rare
Association with harelip	No association
Absence of finger buds	Finger buds are common
Feet affected	Feet unaffected

symbrachydactyly

Mx:

- Excision of transverse bones
- Cleft narrowing
- Web space creation
- Shifting index to
- position of mid

shown in ►**Fig. 1A–D**.* All three patients had absent middle finger with its metacarpal present. The incisions were planned as shown. Preoperative photograph for case 1 is



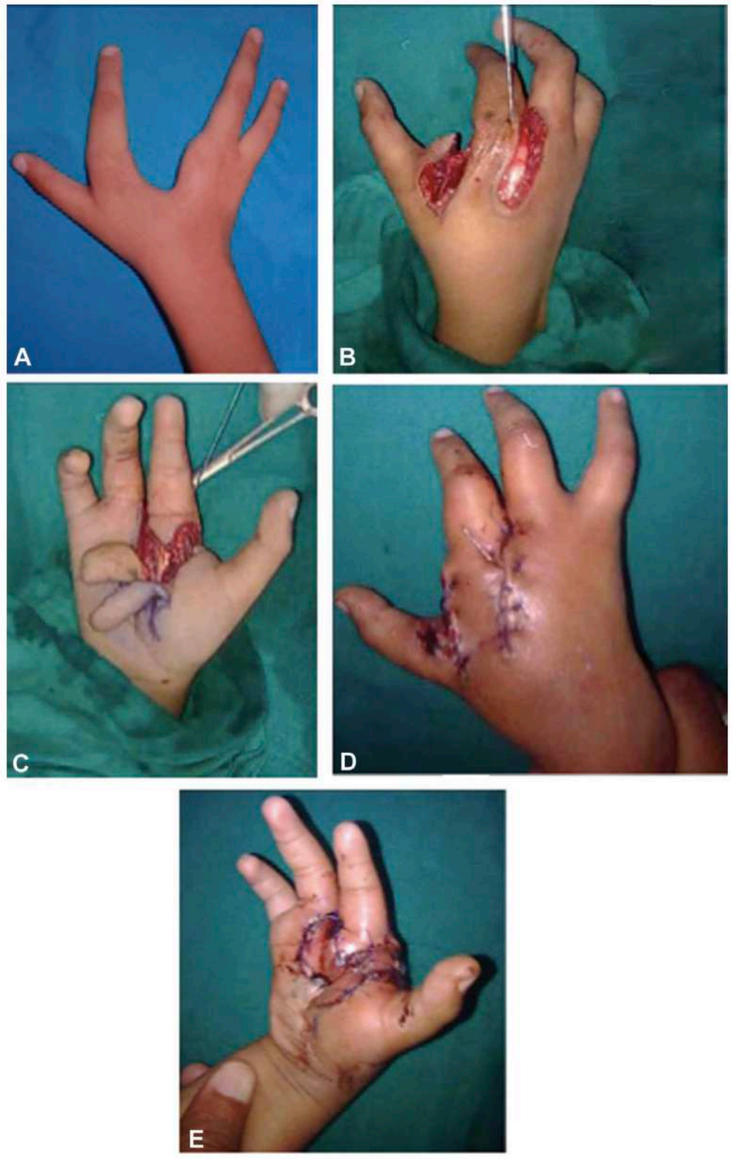


Fig. 1 Schematic diagram for the Snow–Littler procedure: (**A** and **B**)

THUMB HYPOPLASIA

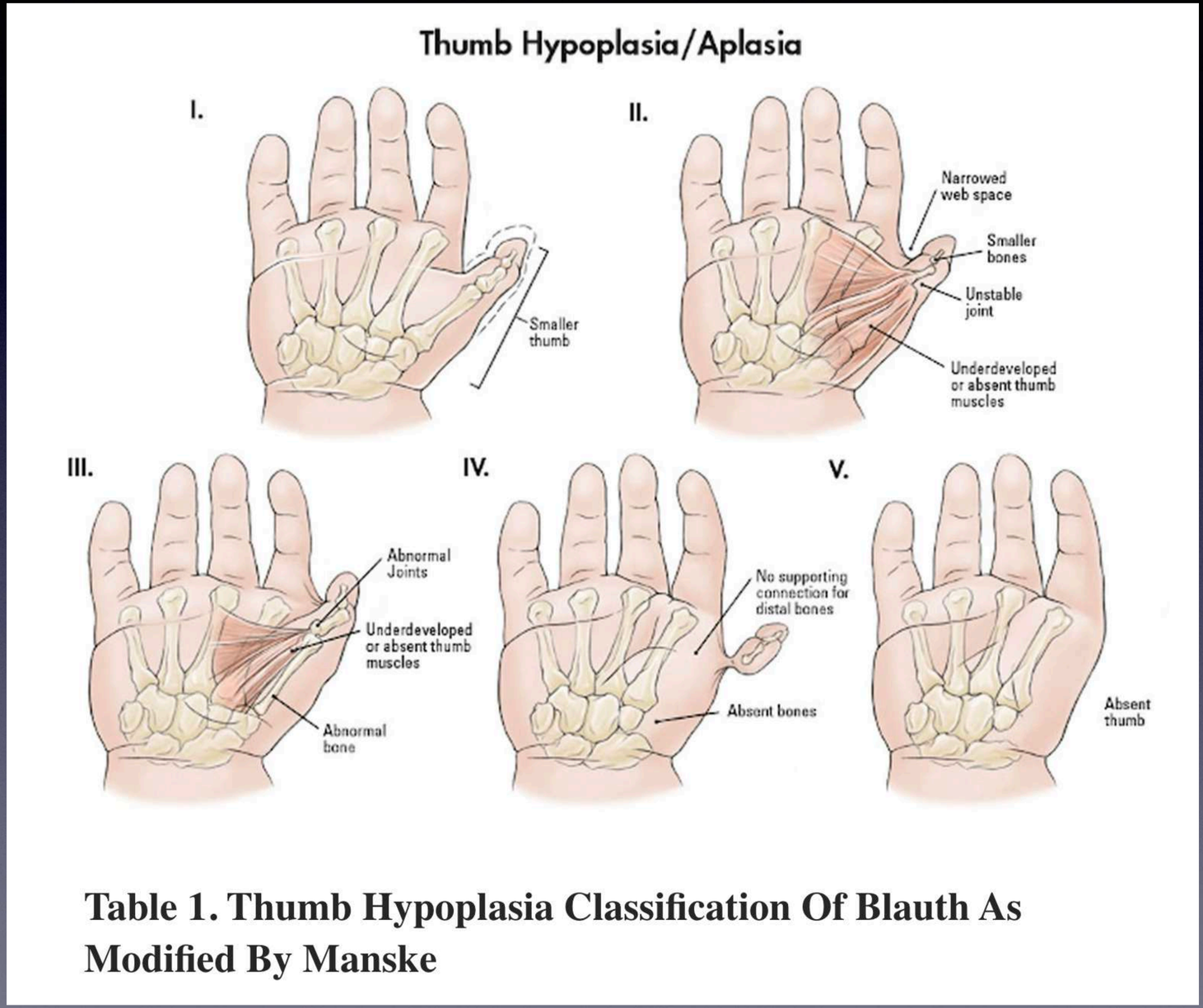


Table 1. Thumb Hypoplasia Classification Of Blauth As Modified By Manske

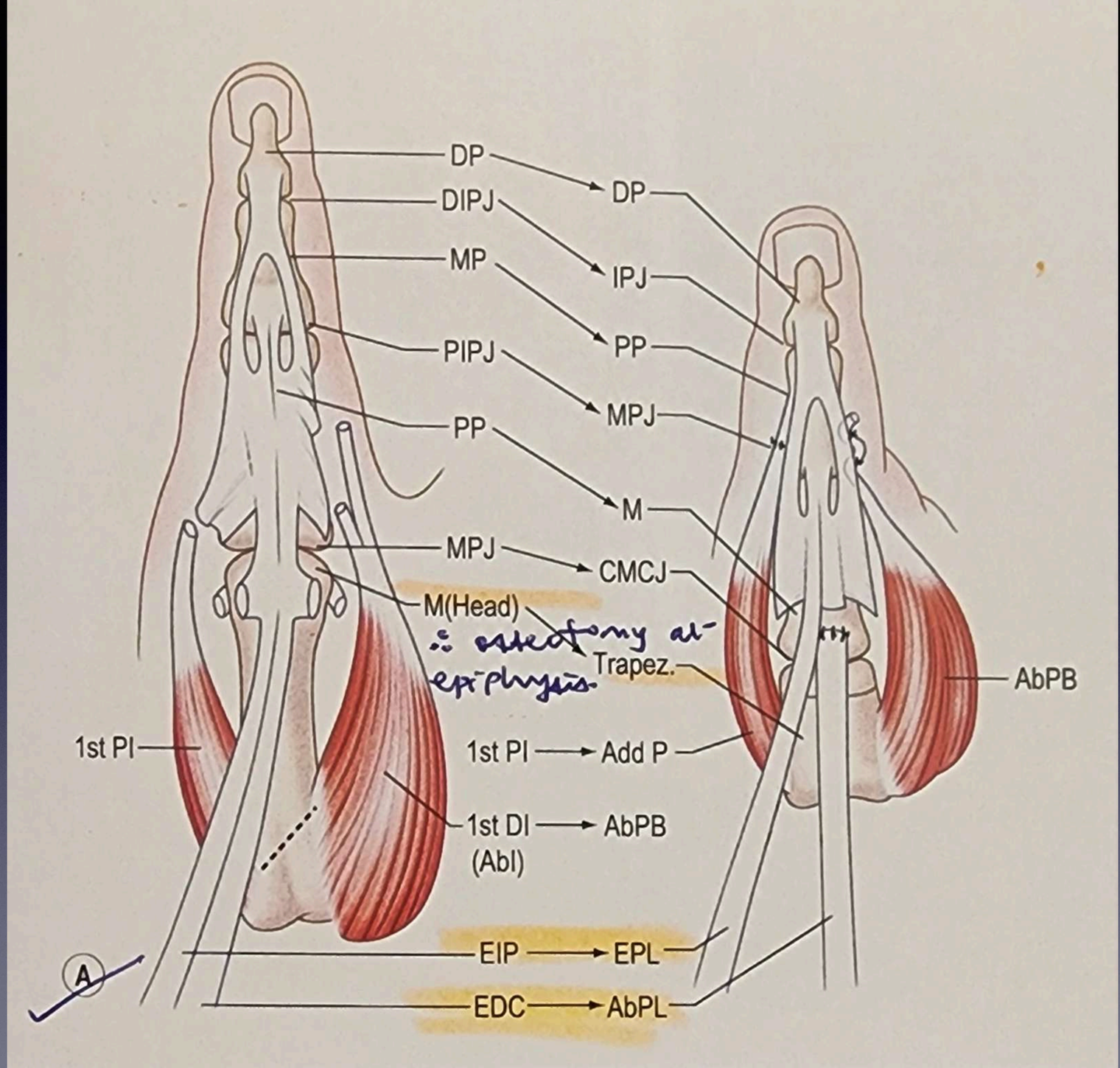
Type	Findings
I	Minor generalized Hypoplasia
II	Absence of Intrinsic thenar muscles First web space narrowing Ulnar collateral ligament insufficiency
III	Similar findings as Type II plus: Extrinsic muscle and tendon abnormalities Skeletal Deficiency A: Stable carpometacarpal joint B: Unstable carpometacarpal joint–(Manske) C: Remnant head of metacarpal only–(Buck–Gramcko)
IV	Pouce flottant or floating thumb
V	Aplasia

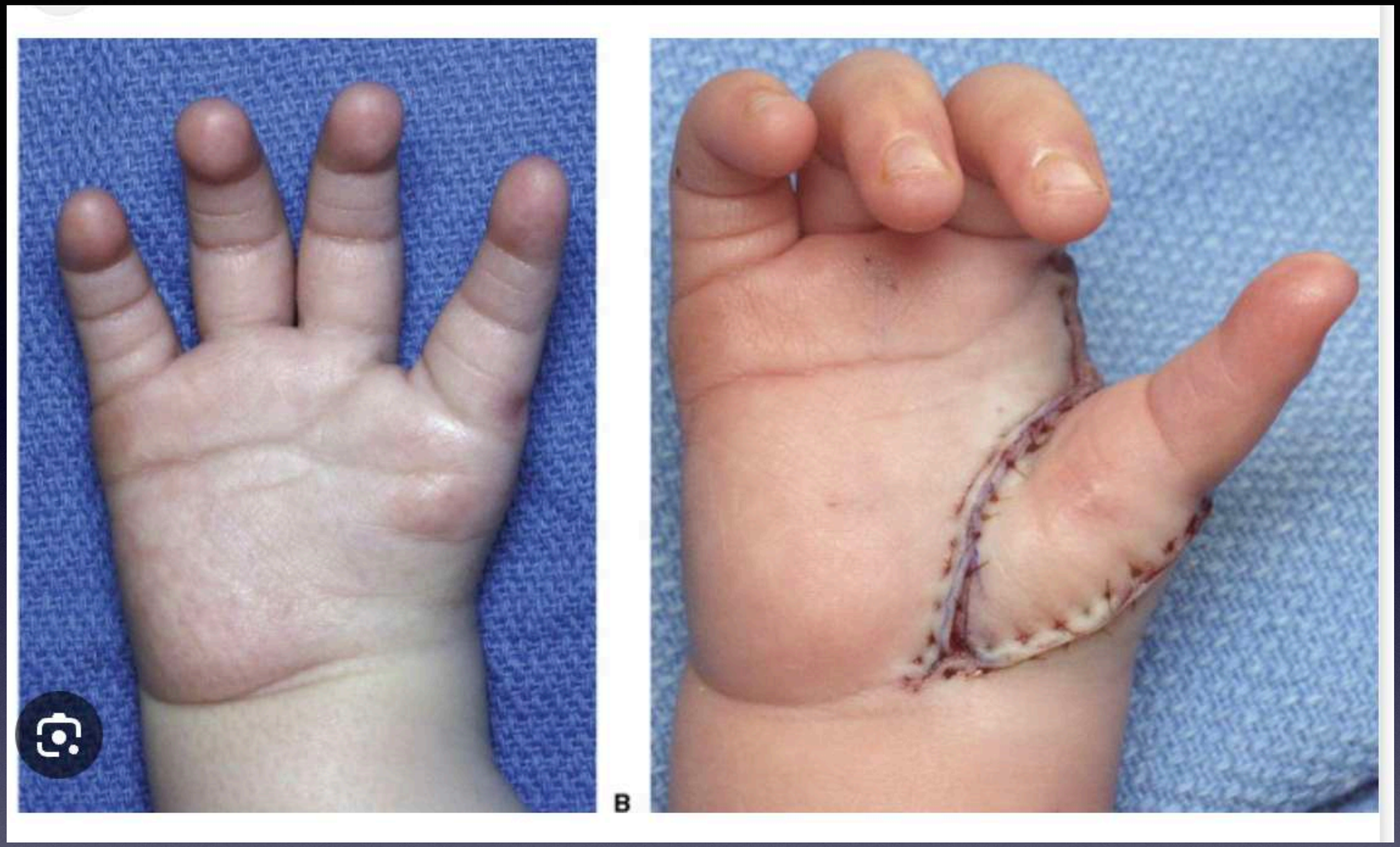
Mx:

- Web space Z plasty
- Hubers oppensplasty
- Transfer of FDS ring to thumb
- Pollicisation 12 to 18months

Pollicization principles:

- Index becomes thumb! - BRAIN PLASTICITY
- Transposition of index on NV island
- Rotation and shortening of ray
- Rebalancing of tendons and muscles
- Creation of good 1st web





Buck Gramcko - racket shaped incision

- Index metacarpal is shortened and physis ablated

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Step	Technique	Rationale
Exsanguination	Moderate	Vessel identification
Skin incision	Ezaki design	More glabrous skin along palmar aspect of thumb and excellent thumb-index web space
Isolation palmar neurovascular bundles	Loupe magnification and meticulous dissection	Preserve sensibility and circulation to index
Microdissection of common digital nerve	Intra-fascicular dissection	Mobilize nerve for tension free pollicization
Ligate proper digital artery to radial side of long finger	Ligature clip	Allows constant visualization throughout the procedure
Release A1 pulley to index finger		Prevent buckling of flexor tendons after pollicization
Incise intermetacarpal ligament		Allows repositioning of the index finger
Elevation of dorsal skin with preservation of dorsal veins		Delaying dorsal exposure allows veins to be filled with blood
Extensor tendons freed from adjacent		Confirm appropriate line of pull to index finger pollicization
Extensor and flexor tendons are not shortened		Adapt and shorten over time
Elevation of the first dorsal and palmar muscles from the index metacarpal and metacarpophalangeal joint with a strip of extensor hood	Sharp dissection	Muscles will be advanced to proximal interphalangeal joint and length is necessary
Identify and tag the radial and ulnar lateral bands about the proximal interphalangeal joint	Pull on lateral band until desired function is evident and tag band with suture	Prior to bony resection, identification is easier
Shorten the index finger by removing the majority of the metacarpal bone, including physis ablation	Fine bladed saw to cut metacarpal perpendicular to bone through its metaphyseal portion	Index too long for a thumb Physeal ablation prevents continued metacarpal growth
	Distal cut directly through physis (epiphysiodesis)	

Reposition index metacarpophalangeal joint into hyperextension	Fixation of the index metacarpophalangeal joint into hyperextension using a non-absorbable suture placed through the epiphysis and dorsal capsule	Prevents unwanted thumb carpometacarpal joint hyperextension
Kirschner wire is passed anterior to the metacarpal epiphysis, into the proximal phalanx, and out the proximal interphalangeal joint	Wire driver	This Kirchner wire is used as a joystick for index finger positioning and ultimate fixation
Align the index finger into the thumb position with 45-degrees of abduction and between 100 and 120-degrees of pronation	Metacarpal epiphysis is aligned anterior to its remaining base and Kirschner wire drilled retrograde across the metacarpal base to secure the position	Replicate thumb position
Tendon transfer to restore intrinsic function to the pollicization	First dorsal interosseous sutured into the radial lateral band and the first palmar interosseous sutured into the ulnar lateral band	Maximize function in grasp and pinch
Inset skin with absorbable suture	Skin inset advanced and inset along the palmar aspect of the "thumb"	Index appearance similar to thumb
	Inset web space skin	Avoid suture line in thumb-index web space
	Trim any excess skin	
Deflation of tourniquet and meticulous postoperative dressings	Bulky hand-dressing with long-arm soft cast	Ensure circulation, protect pollicization, decrease chances of inadvertent dressing removal

- Above elbow splint is given for 4 - 5 weeks
- K wire removed after 4- 6 weeks
- Brain plasticity encouraged by training exercises
- Large object f/b pinch
- Opening containers, picking up crayons, scribbling etc

THANK YOU!

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Instagram – drps_plastics

Youtube – DrPS