CONGENITAL HAND DYSPLASIA

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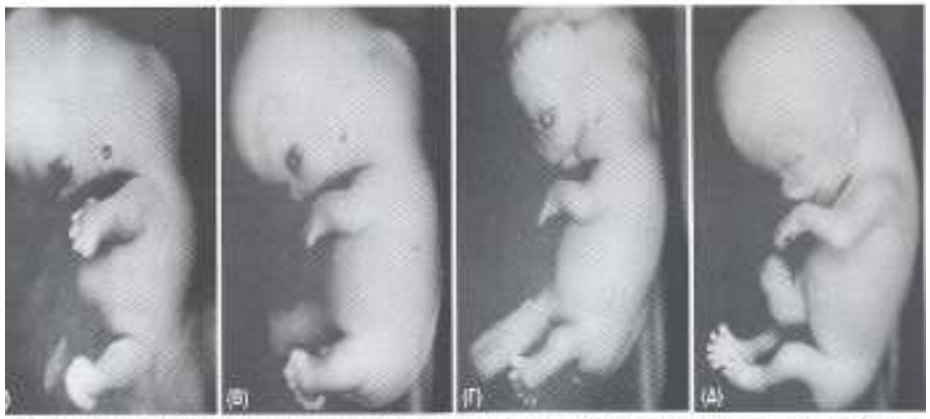
M.Ch.Orth

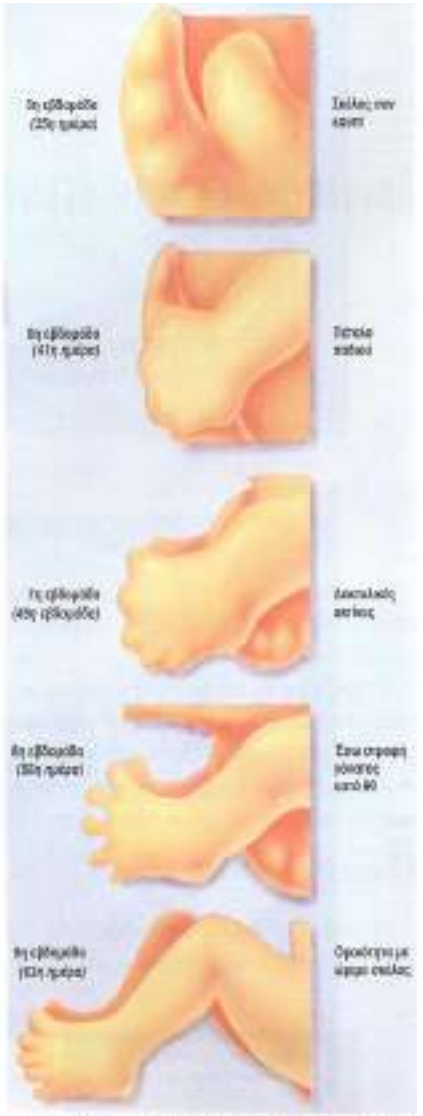
Fetus development - embryogenesis
all limb structures are present at 8 weeks

Fetus development

- lateral Buds
- Limb formation
- Digit formation



σνα 1-3. Διαχωρισμός δακτύλων κατά την 8η εβδομάδα. (Α) Έμβρυο 46 ημερών (Β) Έμβρυο 51 ημερών (Γ) Έμβρυο 54 ερών (Δ) Έμβρυο 58 ημερών (τροποποιημένη από Moore K: Έγχρωμη Κλινική Εμβρυολογία, Ιατρικές Εκδόσεις Π.Χ. Παπαζήση 1997).



Fetus development

Embryology of congenital differences

Scott H Kozin
Shriners Hospital for Children Philadelphia

J Pediatr Orthop 30: S 31 2010

signaling centers

AER proximal distally

ZPA, anteroposterior pre – post axial

signaling molecule is the sonic hedgehog protein

Wingless type center (Wnt) dorsoventral development

Hox and T-box genes

Embryology of congenital differences

proximal to distal development

AER Apical Ectodermal Ridge (thickened layer of ectoderm)

Hox and T-box genes

Congenital skeletal deficiencies

- Classification , syndrome
- Cardiovascular – kidney function
- Treatment planning considering growth
- Inheritance

CLASSIFICATION

- Absence of part failure of formation
  longitudinal or transverse
  terminal or intercalary
- Polydactyly
- Failure of separation syndactyly cleft
- Over Under Growth gigantism hypoplasia
- Malformation clinodactyly, arthrogryposis
- Unclassified constriction band syndrome

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  - Over Under Growth gigantism hypoplasia
  - Malformation CTEV, Vertical talus, CMV,
    toes abnormalities
  - Unclassified constriction band syndrome

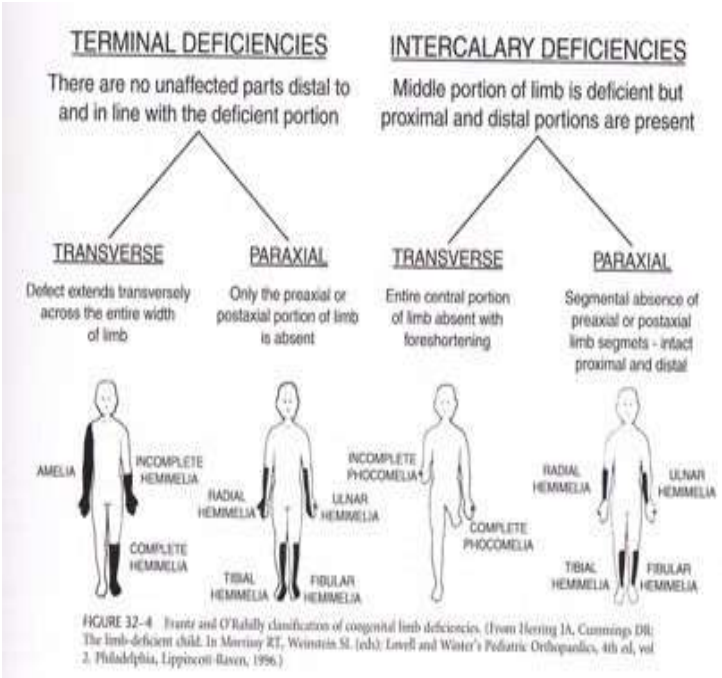


FIGURE 32-4 Frantz and O'Rahilly classification of congenital limb deficiencies. (From Herring JA, Cummings DR: The limb-deficient child. In Morrissy RT, Weinstein SL (eds): Lovell and Winter's Pediatric Orthopaedics, 4th ed, vol 2. Philadelphia, Lippincott-Raven, 1996.)

Absent digits – terminal transverse

Absent digits – terminal transverse

Absent digits

Absent digits 2

Absent digits 3

Brachy metacarpodactyly nik

Brachy metacarpodactyly free phalangeal fracture

Brachy metacarpodactyly 2 final result nik

brachymetacarpodactyly

- Function
- cosmesis

Brachymetacarpodactyly Pap symela

- Lengthening of fingers

Newborn with congenital deformity

- Importance of appropriate treatment with occupational therapy
- Severe medico legal problems, that was NOT diagnosed prenatal

Nail absence

Longitudinal deficiencies

- Thumb hypoplasia

Thumb hypoplasia

Thumb hypoplasia

PRENATAL DIAGNOSIS OF SKELETAL DYSPLASIAS
3 ray hand

Longitudinal deficiencies

- Radial club hand

Longitudinal deficiencies

- Radial club hand

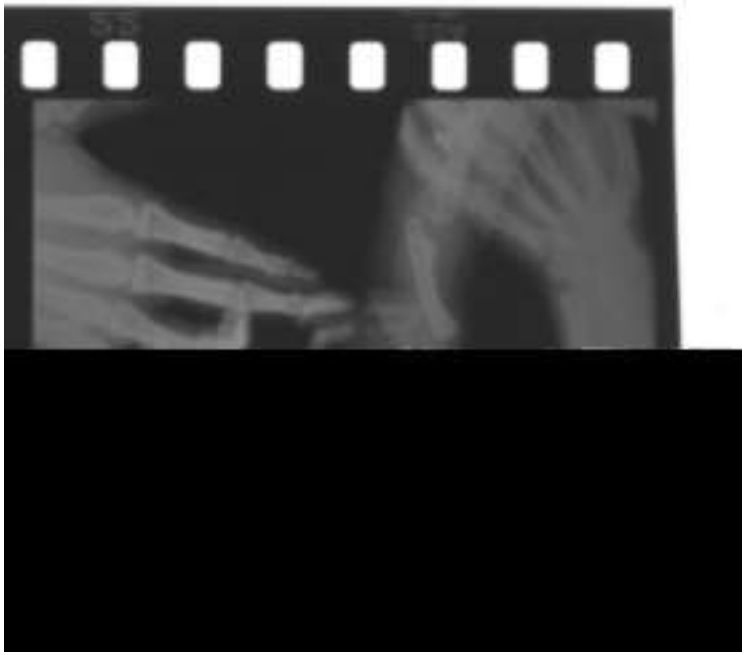
Longitudinal deficiencies

- Radial club hand

Longitudinal deficiencies

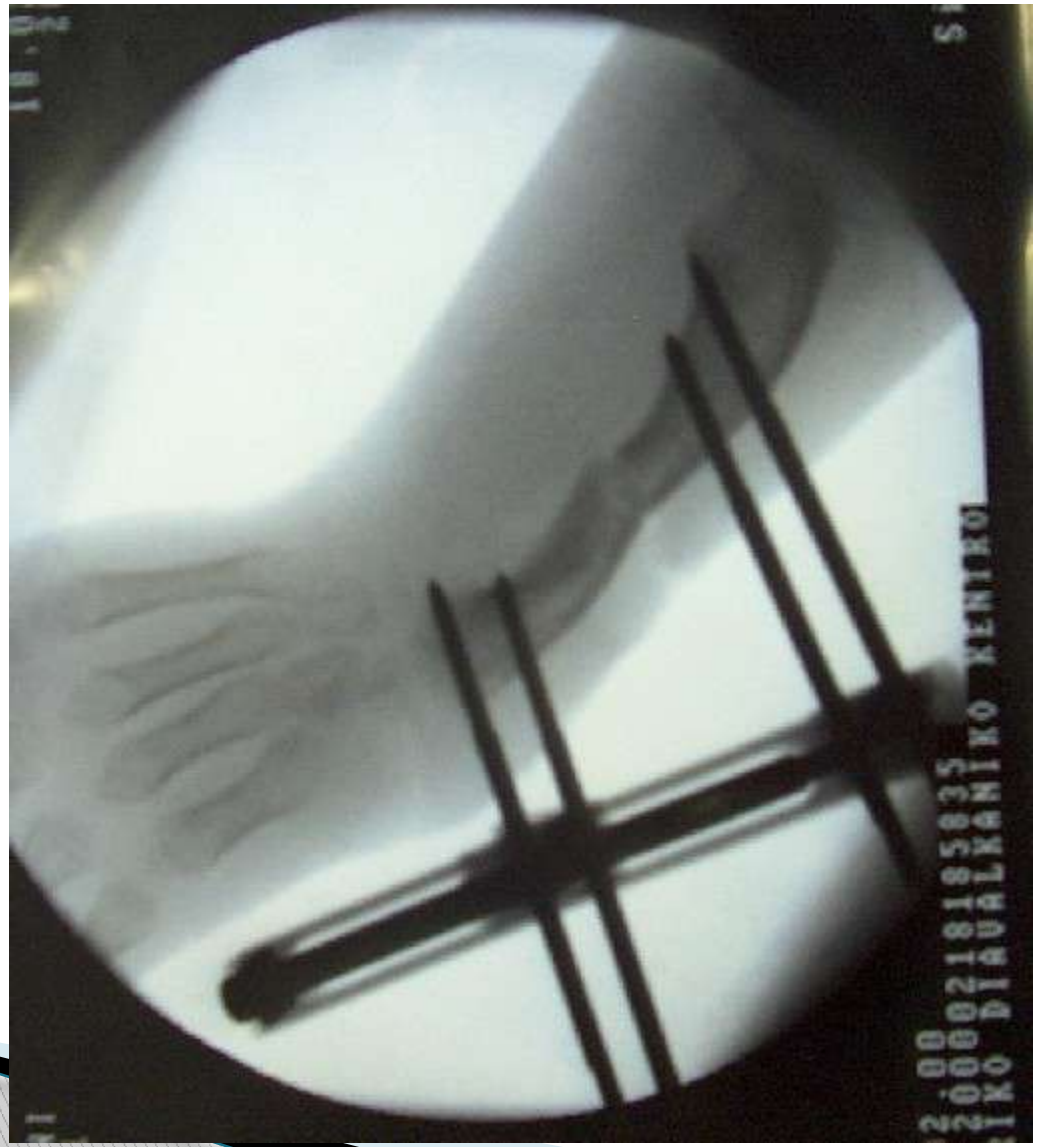
- Radial club hand function

Congenital absence of Radius



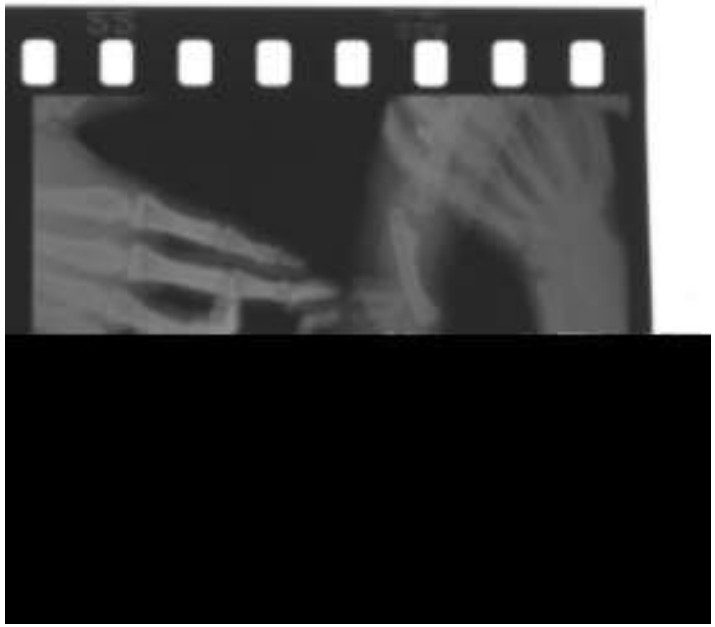
Congenital absence of Radius

Congenital absence of Radius b



Congenital absence of Radius c

Congenital absence of Radius d



Congenital absence of Radius e

Longitudinal 5th metacarpal absence

Longitudinal 5th metacarpal function stoik

Cleft hand

Cleft hand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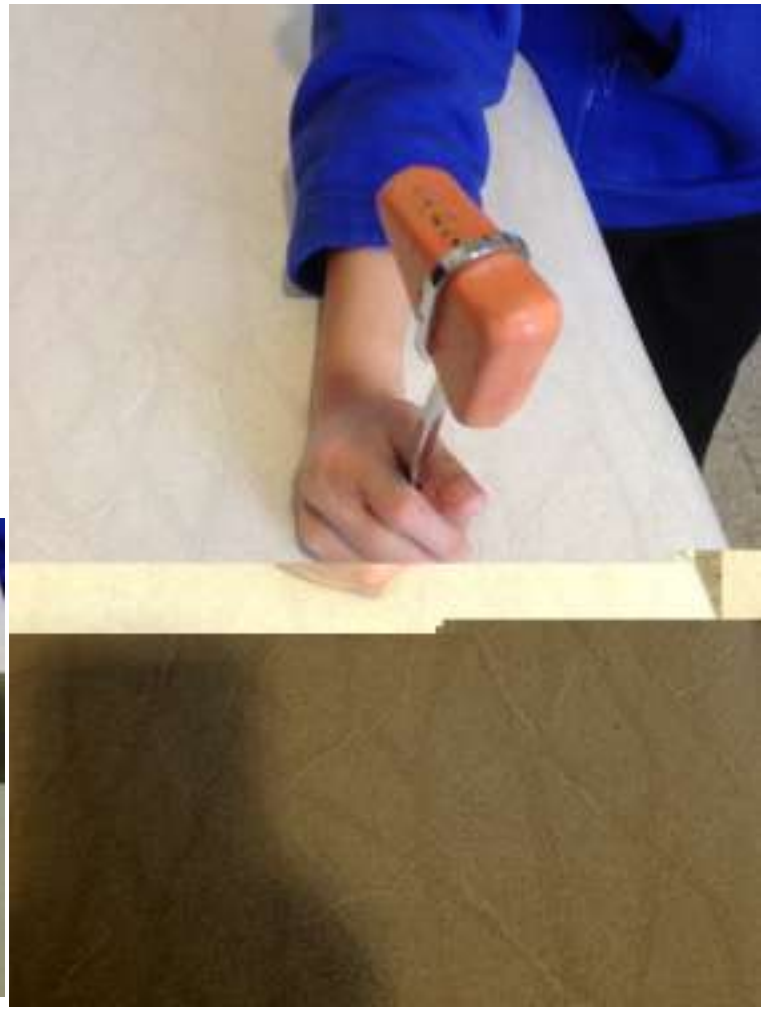
Cleft hand



Διαμαρτίες κνήμης

- Συνοδές βλάβες

Longitudinal deficiency

Διαμαρτίες κνήμης

Διαμαρτίες κνήμης

- απλασία κνήμης

Διαμαρτίες κνήμης

- απλασία -υποπλασία κνήμης
- Jones classification

polydactyly

- Duplication of thumb
- Polydactyly of little finger
- Medial rays

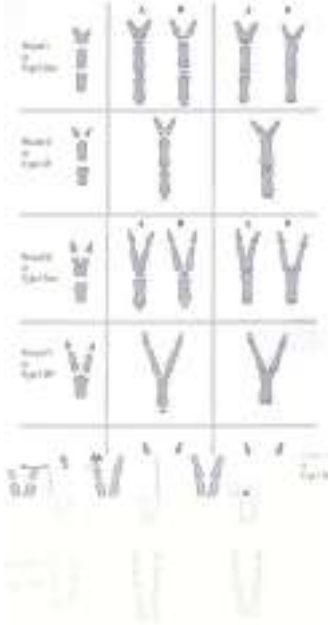
Duplication of the thumb Classification

- Duplication of distal phalanx
- Duplication of distal two phalanges
- Duplication by a minor thumb with noticeably less than normal size

  Hartrampf 1974

Duplication of the thumb Classification

- Wassel classification in 7 types
- Universal classification system of Buck Gramko
- I,II, III, IV,V involved ray
- Dist,DIP,MID PIP Prox MP, MET CM C



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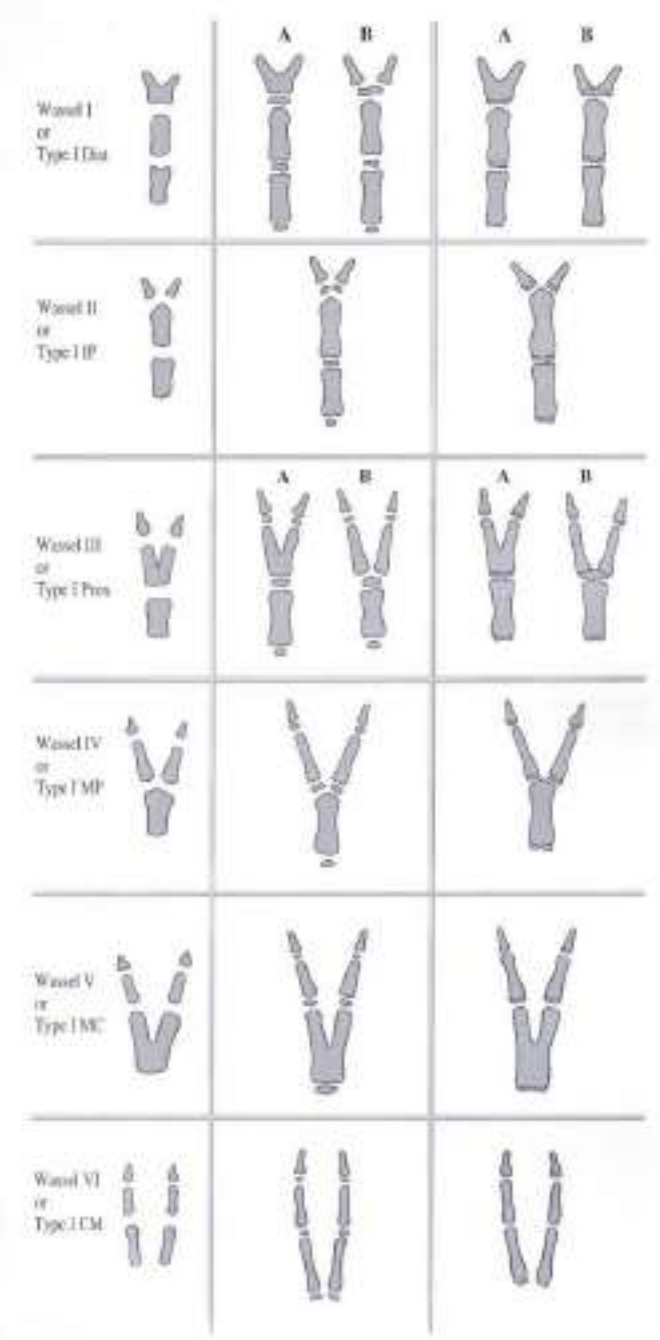
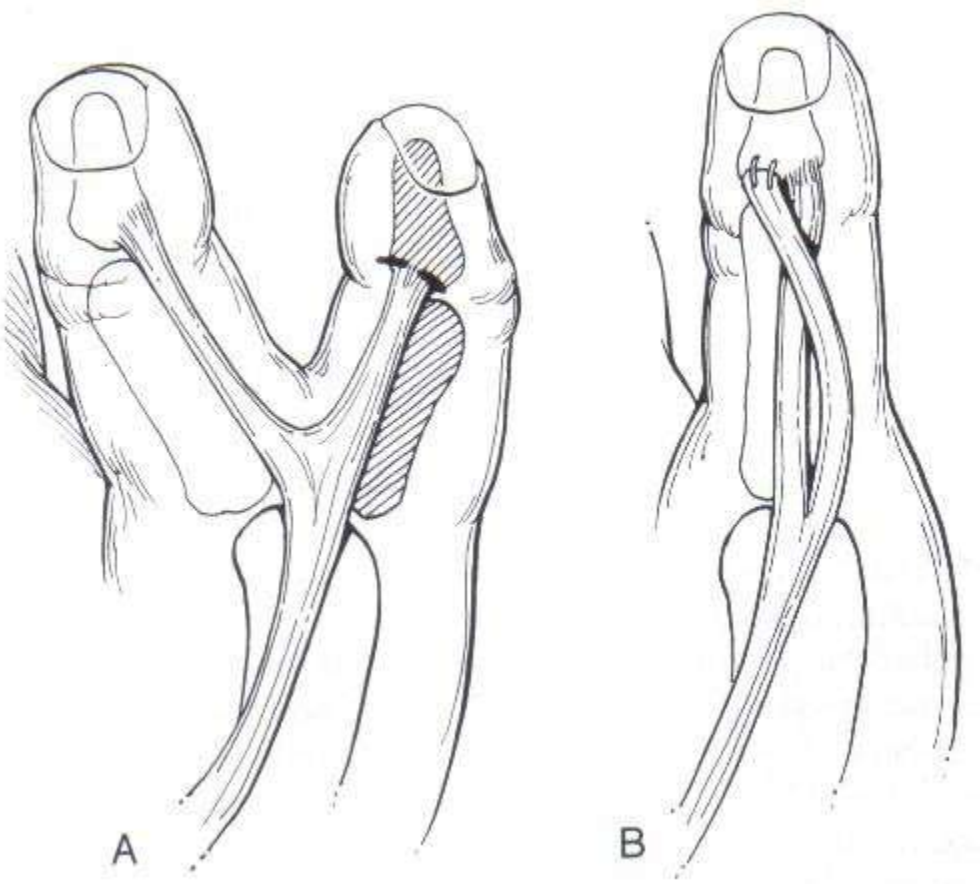


Duplication of the thumb

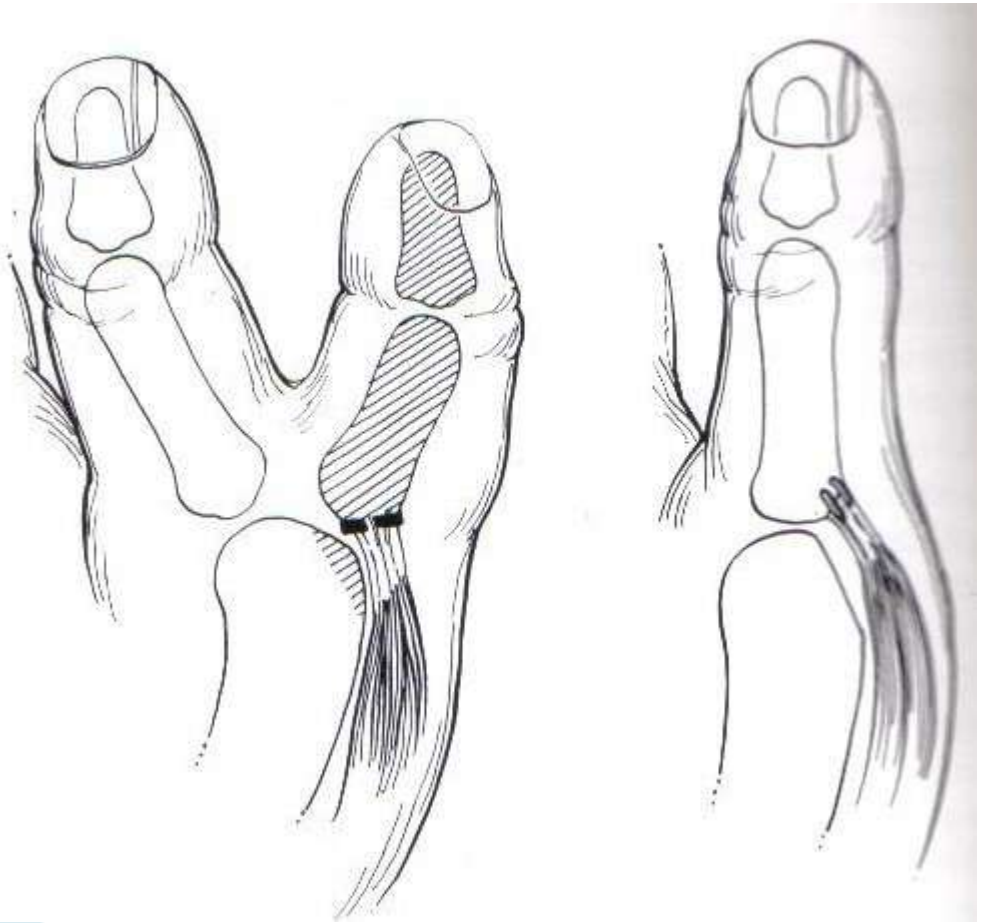
Extrinsic flexor and extensor tendons

- Separate tendons or a single common tendon that diverge in two tendons
- Eccentric insertions on the distal phalanx
- Anomalous slip of FPL to extensor mechanism creates stiffness of the IP joint flexion
- Incidence of 20% as reported from Lister



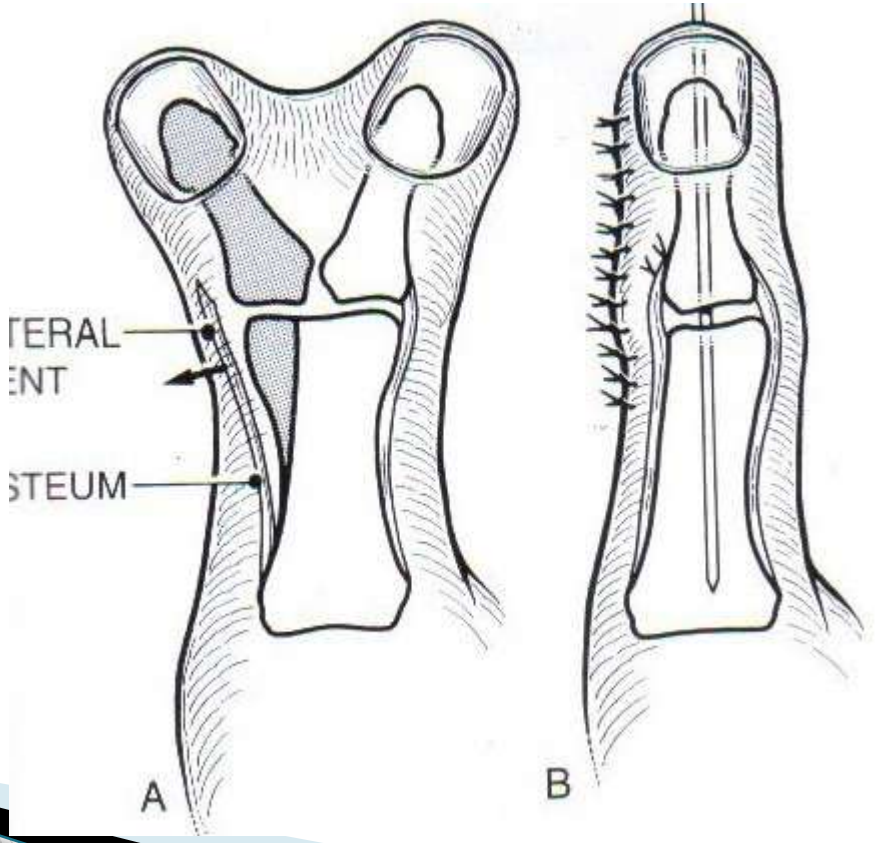
Intrinsic muscle insertion

- Abductor pollicis brevis in the radial digit



Collateral ligament reconstruction

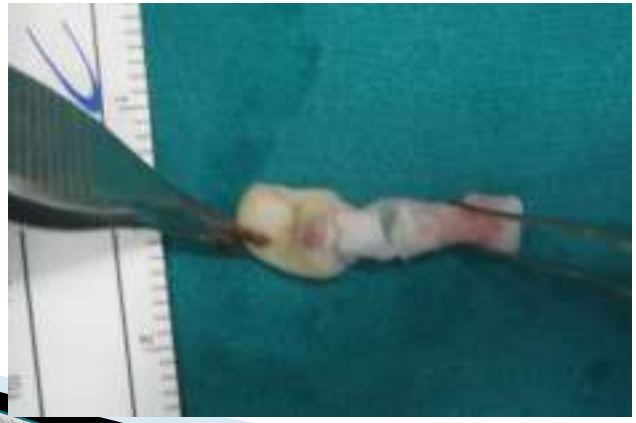
- Radial collateral ligament
- Unstable ulnar digit



Kou 1

Kou 2

Kou 3



Kou 4

Duplication of the thumb

Duplication of the thumb

Modified Bilhaut-Cloquet Procedure for Wassel Type-II and III Polydactyly of the Thumb

By Goo Hyun Baek, MD, Hyun Sik Gong, MD, Moon Sang Chung, MD, Joo Han Oh, MD, Young Ho Lee, MD, and Sang Ki Lee, MD

Investigation performed at the Department of Orthopedic Surgery, Seoul National University College of Medicine, Seoul, South Korea

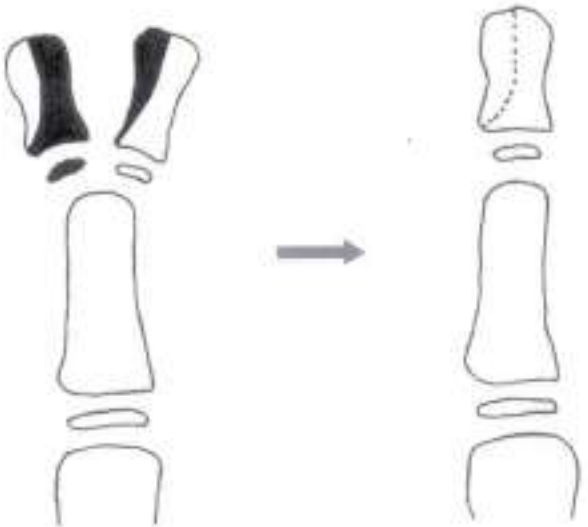


Fig. 1

The modified Bilhaut-Cloquet procedure applied to a Wassel type II (bifid) thumb. The dark area is resected, and the two distal phalangeal bones are combined extramedullarily.

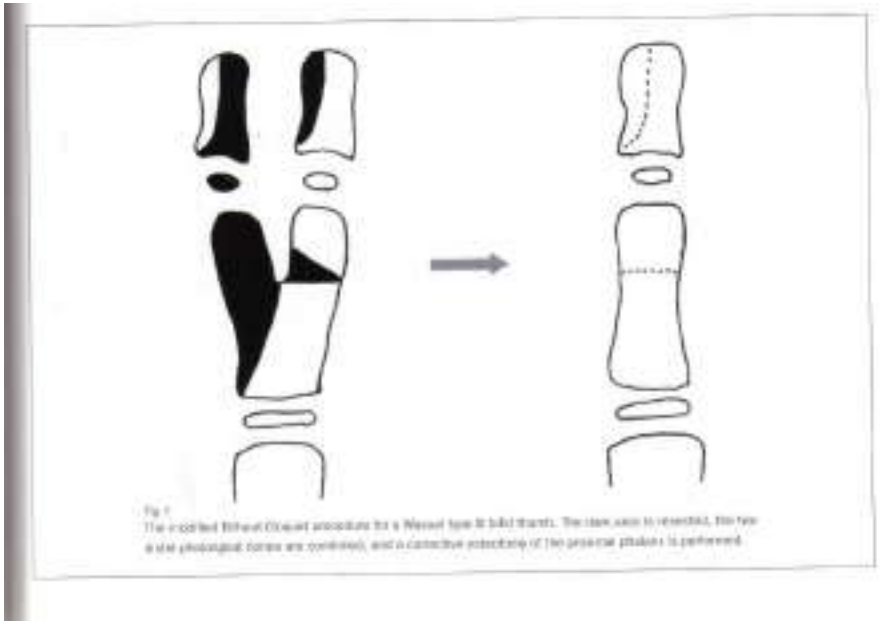


Fig. 1

The modified Bilhaut-Cloquet procedure for a Wassel type III (tri) thumb. The dark area is resected, the two ulnar phalangeal bones are combined, and a corrective osteotomy of the proximal phalanx is performed.

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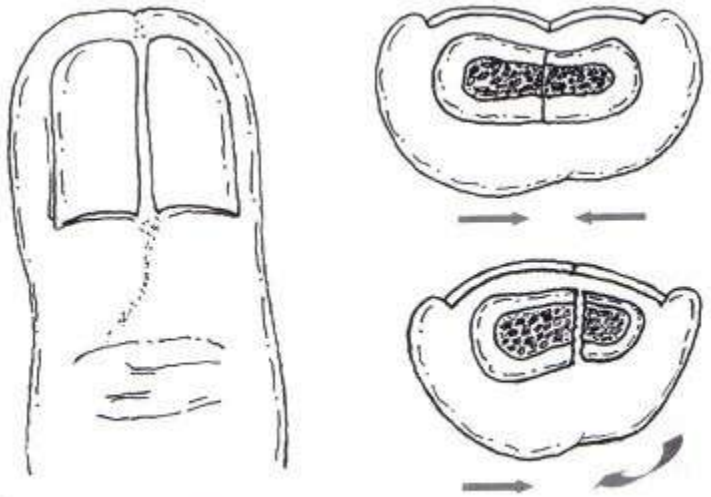


Fig. 2

Axial rotation of the distal phalangeal bone is necessary to make one semicircular nail bed (right bottom). Attaching two semicircular nails in a transverse plane creates a so-called seagull deformity (right top).

Figs. 4-A through 4-E Case 5. A four-year-old boy with type-II polydactyly of the left thumb.

Fig. 4-A Preoperative photograph (left) and anteroposterior radiograph (right).

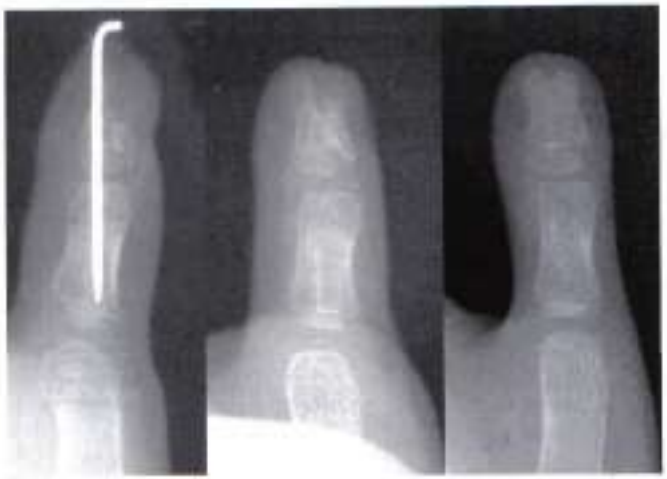


Fig. 4-B

Radiographs made immediately postoperatively (left), six weeks after surgery (middle), and eighteen months (right) after the modified Bilhaut-Cloquet procedure, showing remodeling of the distal phalanx with time.

Bilhaut cloquet

Bilhaut cloquet 2

Bilhaut cloquet 3

Thumb triplication

polydactyly

- 5th ray

polydactyly

- 5th ray

polydactyly

- 5th ray
- Reverse position

polydactyly

- Middle ray

Animals polydactyly

- Cats normally have five digits on the front paws and four on the rear. Polydactyl cats have more, and this is a moderately common condition, especially in certain cat populations.

- Polydactyly was believed to be common in the earliest tetrapods, the number of digits settling to the common five on each foot in amniotes and four to the hand and five to the foot in amphibians in the early Carboniferous

Animals polydactyly

octapus

- *The Silence of the Lambs* (1988) by Thomas Harris features the character Hannibal Lecter who has a duplicate middle finger on his left hand. This physical anomaly was never portrayed in the film adaptation

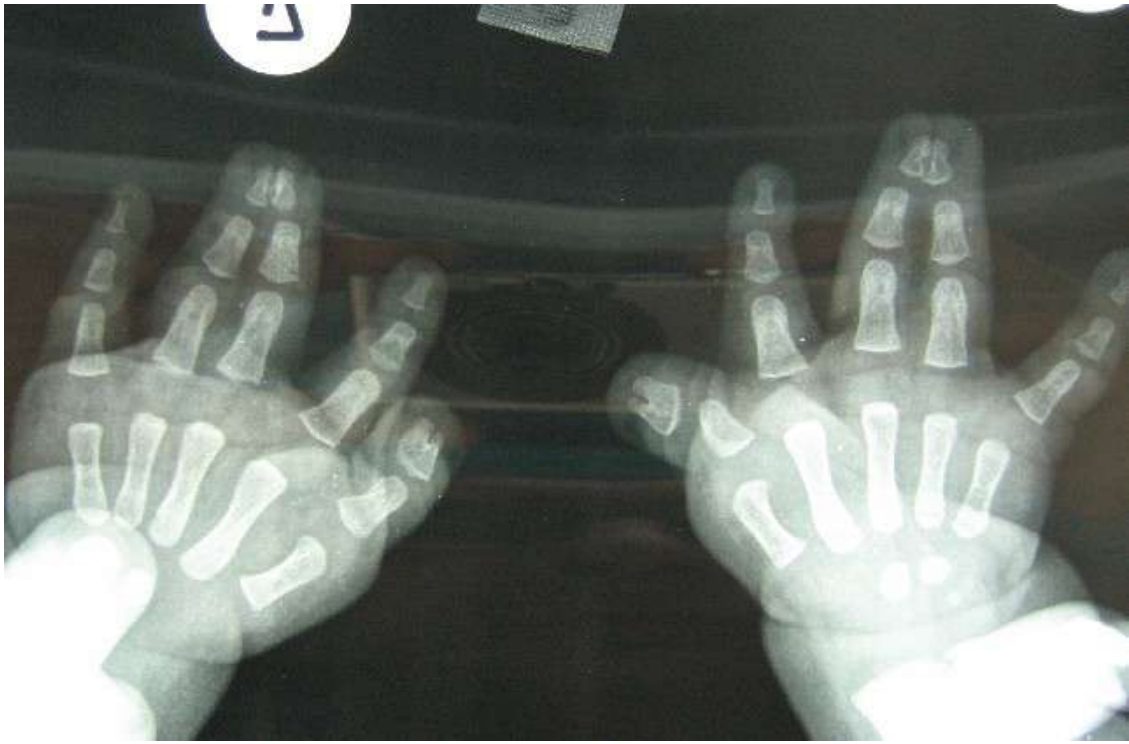
- In Dostoyevsky's *The Brothers Karamazov* the son of Grigory Vasilievich Kutuzov and Marfa Ignatyevna is born with six fingers. Devastated by the abnormal features of the baby, Grigory is reluctant to have it baptized calling the baby "a confusion of nature."

syndactyly

- Skin partial or complete
- bone

Congenital syndactyly mem ag

Congenital syndactyly b



Congenital syndactyly c

Congenital syndactyly feet and hand

Syndactyly hatz

Syndactyly mouz ol

Syndactyly mouz ol web creeping

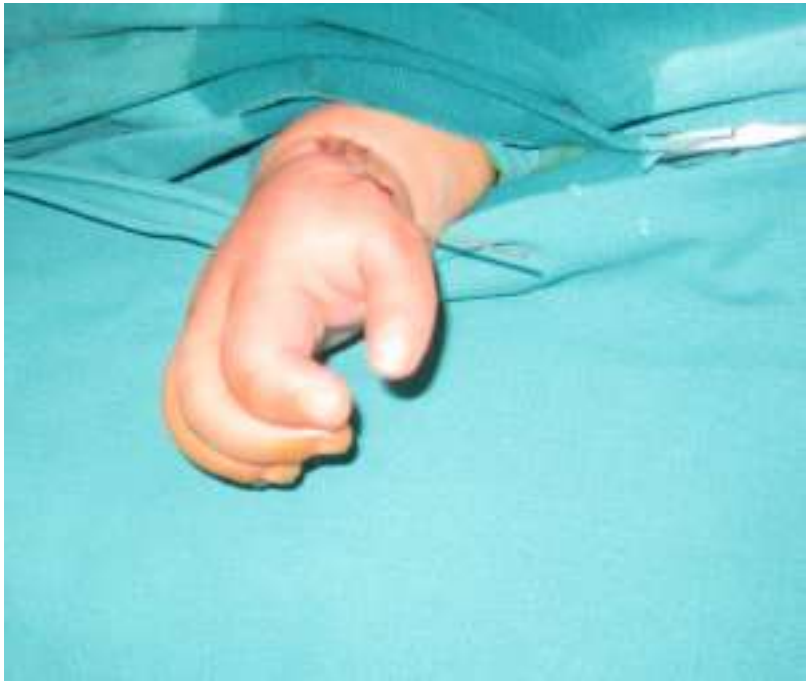
Syndactyly mouz ol final correction

synostosis

Triradiate hand

Thumb deviation

Thumb deviation



delta phalanx

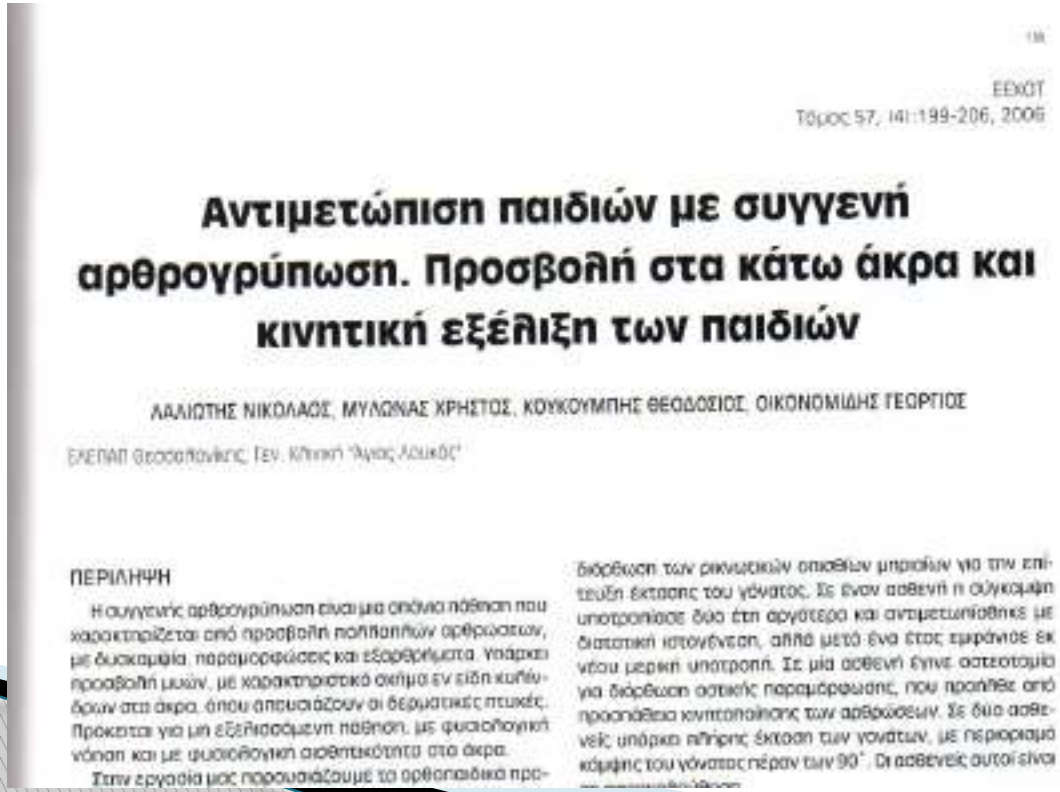
delta phalanx

thumb deviation Rubinstein Taubi

Arthrogryposis

Arthrogryposis

- Variety of severe muscoleskeletal problems
- Normal intelligence



ΕΕΧΟΤ
Τόμος 57, τ4ι:199-206, 2006

Αντιμετώπιση παιδιών με συγγενή αρθρογρύπωση. Προσβολή στα κάτω άκρα και κινητική εξέλιξη των παιδιών

ΛΑΛΙΩΤΗΣ ΝΙΚΟΛΑΟΣ, ΜΥΛΩΝΑΣ ΧΡΗΣΤΟΣ, ΚΟΥΚΟΥΜΠΗΣ ΘΕΟΔΟΣΙΟΣ, ΟΙΚΟΝΟΜΙΔΗΣ ΓΕΩΡΓΙΟΣ

ΕΛΕΠΑΠ Θεσσαλονίκης, Γεν. Κλινική "Άγιος Λουκάς"

ΠΕΡΙΛΗΨΗ

Η συγγενής αρθρογρύπωση είναι μια σπάνια πάθηση που χαρακτηρίζεται από προσβολή πολλαπλών αρθρώσεων, με δυσκαμψία, παραμορφώσεις και εξαρθρήματα. Υπάρχει προσβολή μυών, με χαρακτηριστικό σκήμα εν είδη κυλίνδρων στα άκρα, όπου απουσιάζουν οι δερματικές πτυχές. Πρόκειται για μη εξελισσόμενη πάθηση, με φυσιολογική νόηση και με φυσιολογική ευφυΐα στα άκρα.

Στην εργασία μας παρουσιάζουμε τα ορθοπαιδικά προ-

διόρθωση των εκτατικών οπισθίων μηριαίων για την επιτεύξη έκτασης του γόνατος. Σε έναν ασθενή η σύγκαμψη υποτροπίασε δύο έτη αργότερα και αντιμετωπίσθηκε με διατατική ισοντένταση, αλλά μετά ένα έτος εμφάνισε εκ νέου μερική υποτροπή. Σε μία ασθενή έγινε οστεοτομία για διόρθωση οστικής παραμόρφωσης, που προήλθε από προσπάθεια κινητοποίησης των αρθρώσεων. Σε δύο ασθενείς υπάρχει πλήρης έκταση των γονάτων, με περιορισμό κάμψης του γόνατος πέραν των 90°. Οι ασθενείς αυτοί είναι

Constriction band

Clinodactyly camptodactyly

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  terminal or intercalary
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take home message

- VARIETY of deformities
- EACH one A DIFFERENT PRESENTATION
- Evaluation of a GROWING child
- Approach of a FAMILY problem
- Adequate follow up
- DIFFERENT APPROACH OF CONGENITAL vs TRAYMATIC

Thank you