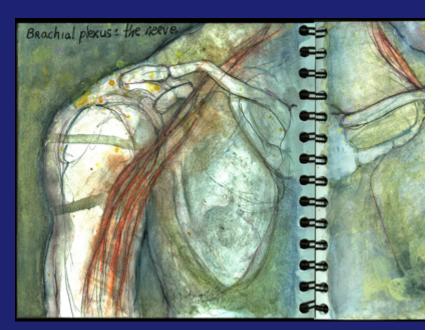
Congenital Hand Anomalies



AND



Brachial Plexus Birth Injuries

Harvey Chim, MD FACS

Associate Professor

Division of Plastic and Reconstructive Surgery University of Florida College of Medicine

Overview

- Congenital hand anomalies
 - Classification
 - Common conditions
- Brachial plexus birth injuries
 - Diagnosis
 - Management

Congenital hand anomalies

- 1 to 2% of newborns
- 10% of these have upper extremity abnormalities
- Aim is to restore function
- Common conditions
 - Polydactyly
 - Syndactyly
 - Radial deficiency
 - Hypoplastic thumb
 - Constriction ring syndrome
 - Clinodactyly
 - Camptodactyly
 - Congenital trigger thumb

Swanson classification

- Seven categories
- Defines anomalies according to embryonic failure

TABLE II Signaling Pathways During Embryogenesis				
Signaling Center	Responsible Substance Action			
Apical ectodermal ridge	Fibroblast growth factors	Proximal-to-distal limb development, interdigital necrosis		
Zone of polarizing activity	Sonic hedgehog protein	Radioulnar limb formation		
Wnt pathway		Dorsalization of limb		

 Clinical diagnosis for categorization

- I. Failure of formation of parts
 - A. Transverse deficiencies
 - B. Longitudinal deficiencies
 - 1. Phocomelia
 - 2. Radial
 - Central
 - 4. Ulnar
- II. Failure of differentiation
 - A. Synostosis
 - B. Radial head dislocation
 - C. Symphalangism
 - D. Syndactyly
 - E. Contracture
 - 1. Soft tissue
 - a.Arthrogryposis
 - b.Pterygium
 - c.Trigger
 - d.Absent extensor tendons
 - e. Hypoplastic thumb
 - f.Clasped thumb
 - g.Retroflexible thumb
 - h.Camptodactyly
 - i.Windblown hand
 - 2. Skeletal
 - a.Clinodactyly
 - b.Kirner deformity
 - c.Delta bone
- III. Duplication
 - A. Thumb
 - B. Triphalangism/hyperphalangism
 - C. Polydactyly
 - D. Mirror hand
- IV. Overgrowth
 - A. Limb
 - B. Macrodactyly
- V. Undergrowth
- VI. Congenital constriction band syndrome
- VII. Generalized skeletal abnormalities

OMT CLASSIFICATION OF CONGENITAL HAND AND UPPER LIMB ANOMALIES

Approved by the IFSSH Scientific Committee on Congenital Conditions, 3rd February 2014

I. MALFORMATIONS

A. Abnormal axis formation/differentiation entire upper limb

1. Proximal-distal axis

- i. Brachymelia with brachydactyly
- ii. Symbrachydactyly
 - a) Poland syndrome
 - b) Whole limb excluding Poland syndrome
- iii. Transverse deficiency
 - a) Amelia
 - b) Clavicular/scapular
 - c) Humeral (above elbow)
 - d) Forearm (below elbow)
 - e) Wrist (carpals absent/at level of proximal carpals/at level of distal carpals) (with forearm/arm involvement)
 - f) Metacarpal (with forearm/arm involvement)
 - g) Phalangeal (proximal/middle/distal) (with forearm/arm involvement)

iv. Intersegmental deficiency

- a) Proximal (humeral rhizomelic)
- b) Distal (forearm mesomelic)
- c) Total (Phocomelia)
- v. Whole limb duplication/triplication

2. Radial-ulnar (anterior-posterior) axis

- Radial longitudinal deficiency Thumb hypoplasia (with proximal limb involvement)
- ii. Ulnar longitudinal deficiency
- iii. Ulnar dimelia
- iv. Radioulnar synostosis
- Congenital dislocation of the radial head
- vi. Humeroradial synostosis Elbow ankyloses

3. Dorsal-ventral axis

- i. Ventral dimelia
 - a) Furhmann/Al-Awadi/Raas-Rothschild syndromes
 - b) Nail Patella syndrome
- ii. Absent/hypoplastic extensor/flexor muscles

4. Unspecified axis

- i. Shoulder
 - a) Undescended (Sprengel)
- b) Abnormal shoulder muscles
- c) Not otherwise specified
- ii. Arthrogryposis

B. Abnormal axis formation/differentiation hand plate

1. Proximal-distal axis

- Brachydactyly (no forearm/arm involvement)
- Symbrachydactyly (no forearm/arm involvement)
- iii. Transverse deficiency (no forearm/arm involvement)
 - a) Wrist (carpals absent/at level of proximal carpals/at level of distal carpals)
 - b) Metacarpal
 - c) Phalangeal (proximal/middle/distal)

2. Radial-ulnar (anterior-posterior) axis

- Radial deficiency (thumb no forearm/arm involvement)
- ii. Ulnar deficiency (no forearm/arm involvement)
- iii. Radial polydactyly
- iv. Triphalangeal thumb
- V. Ulnar dimelia (mirror hand no forearm/arm involvement)
- vi. Ulnar polydactyly

3. Dorsal-ventral axis

- i. Dorsal dimelia (palmar nail)
- ii. Ventral (palmar) dimelia (including hypoplastic/aplastic nail)

4. Unspecified axis

- i. Soft tissue
 - a) Syndactyly
 - b) Camptodactyly
 - c) Thumb in palm deformity
- d) Distal arthrogryposis
- Skeletal deficiency
- a) Clinodactyly
- b) Kirner's deformity
- c) Synostosis/symphalangism (carpal/metacarpal/phalangeal)
- iii. Complex

- a) Complex syndactyly
- b) Synpolydactyly— central
- c) Cleft hand
- d) Apert hand
- e) Not otherwise specified

II. DEFORMATIONS

- A. Constriction ring sequence
- B. Trigger digits
- C. Not otherwise specified

III. DYSPLASIAS

A. Hypertrophy

- 1. Whole limb
 - i. Hemihypertrophy
 - ii. Aberrant flexor/extensor/intrinsic muscle

2. Partial limb

- i. Macrodactyly
- ii. Aberrant intrinsic muscles of hand

B. Tumorous conditions

- 1. Vascular
 - i. Hemangioma
 - ii. Malformation
 - iii. Others

2. Neurological

- i. Neurofibromatosis
- ii. Others

3. Connective tissue

- i. Juvenile aponeurotic fibroma
- ii. Infantile digital fibroma
- iii. Others

4. Skeletal

- i. Osteochondromatosis
- ii. Enchondromatosis
- iii. Fibrous dysplasia
- iv. Epiphyseal abnormalities
- v. Others

IV. SYNDROMES*

A. Specified

- 1. Acrofacial Dysostosis 1 (Nager type)
- 2. Apert
- Al-Awadi/Raas-Rothschild/Schinzel phocomelia
- 4. Baller-Gerold
- 5. Bardet-Biedl Carpenter
- 6. Catel-Manzke

- Constriction band (Amniotic Band Sequence)
- 8. Cornelia de Lange (types 1-5)
- 9. Crouzon
- 10. Down
- 11. Ectrodactyly-Ectodermal Dysplasia-Clefting
- 12. Fanconi Pancytopenia
- 13. Fuhrmann
- 14. Goltz
- 15. Gorlin
- 16. Greig Cephalopolysyndactyly
- 17. Hajdu-Cheney
- Hemifacial Microsomia (Goldenhar syndrome)
- 19. Holt-Oram
- 20. Lacrimoauriculodentodigital (Levy-Hollister)
- 21. Larsen
- 22. Leri-Weill Dyschondrosteosis
- 23. Moebius sequence
- 24. Multiple Synostoses
- 25. Nail-Patella
- 26. Noonan
 27. Oculodentodigital dysplasia
- 28. Orofacialdigital
- 29. Otopalataldigital
- 30. Pallister-Hall
- 31. Pfeiffer
- 32. Poland
- 33. Proteus
- 34. Roberts-SC Phocomelia 35. Rothmund-Thomson
- 36. Rubinstein-Taybi
- 37. Saethre-Chotzen
- 38. Thrombocytopenia Absent Radius
- 39. Townes-Brock
- 40. Trichorhinophalangeal (types 1-3)
- 41. Ulnar-Mammary
- 42. VACTERLS association

B. Others

*The specified syndromes are those considered most relevant; however, many other syndromes have a limb component categorized under "B. Others".

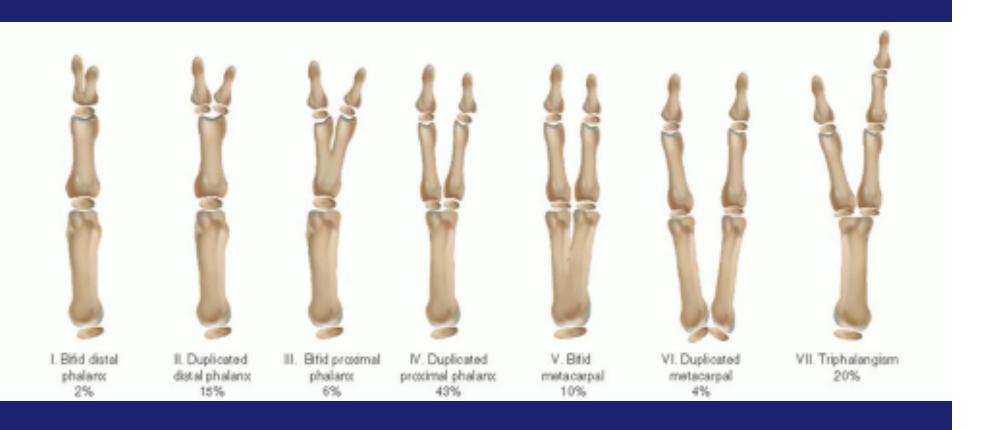
OMT Classification

- 3 groups
 - Malformations
 - Deformations
 - Dysplasias
- Extent of involvement
 - Whole of limb affected or hand plate alone
 - Whether insult involves one of 3 axes of limb development or non-axial

Polydactyly

- Preaxial (radial)
 - Asian
 - White
- Postaxial (ulnar)
 - African American
- Central
 - May be combined with syndactyly

Preaxial polydactyly/ Duplicated thumb

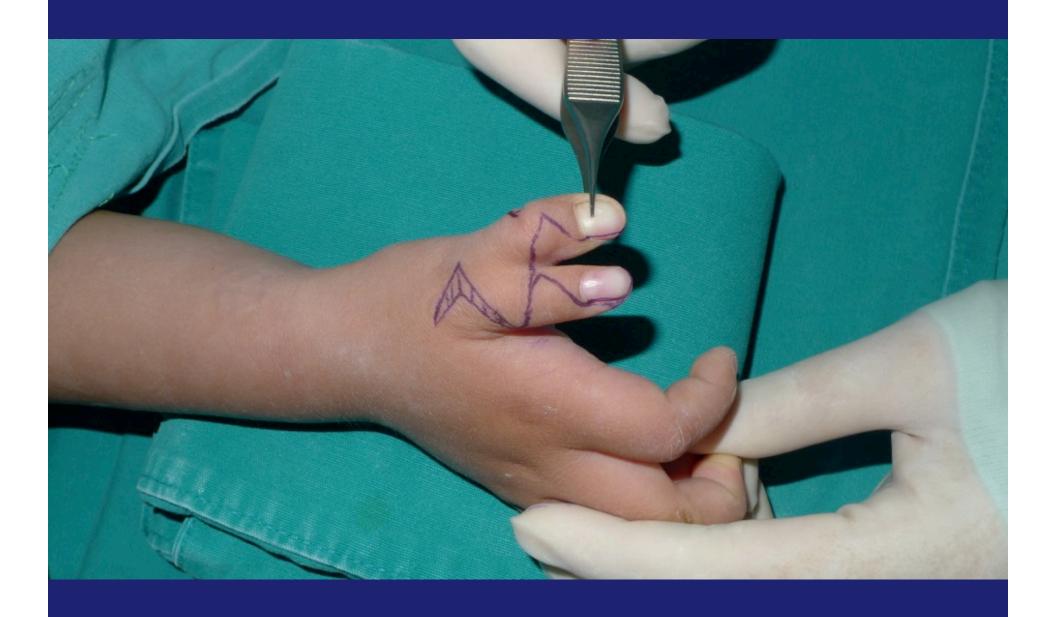


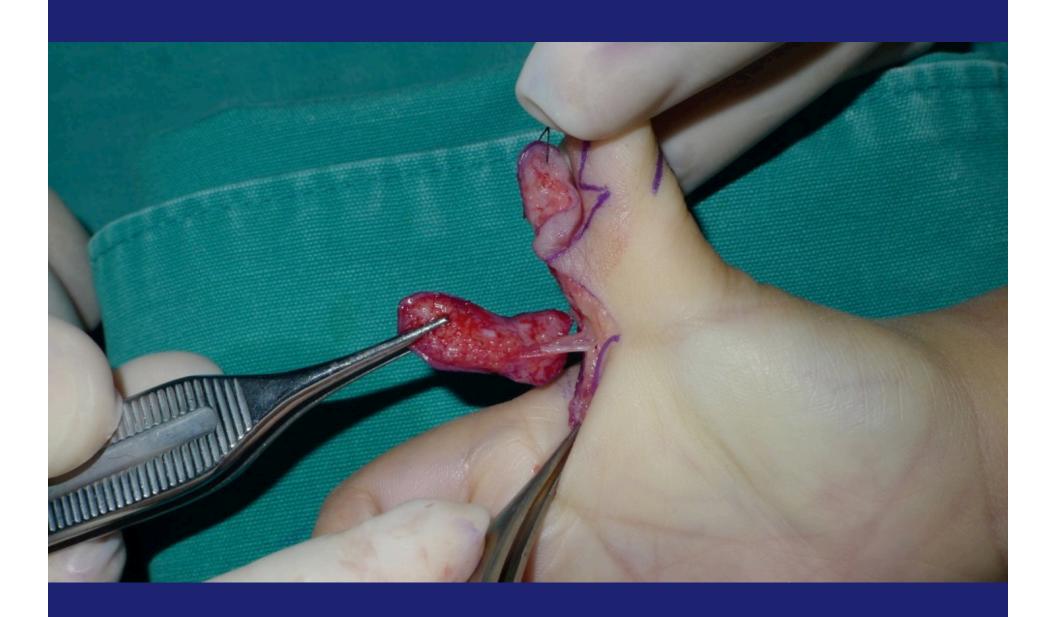
Wassel IV thumb duplication



Treatment

- All patients will need surgery
- Timing
 - 12 to 18 months
- Goals
 - To make a thumb that is at least 80% of size of the contralateral thumb
 - Preserve/ reconstruct collateral ligament of smaller thumb
 - Make functional thumb with pinch

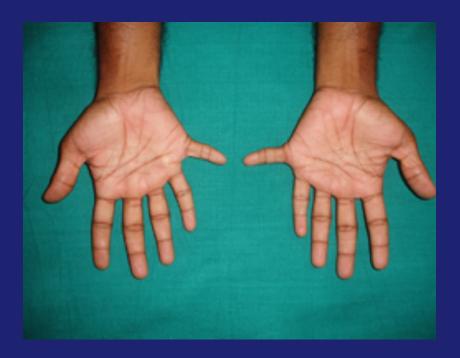






Postaxial polydactyly

- Type A
- Well formed digit



- Type B
- Rudimentary skin tag



Treatment

- Excision in OR
- Not clipping by bedside
 - Painful neuroma when older

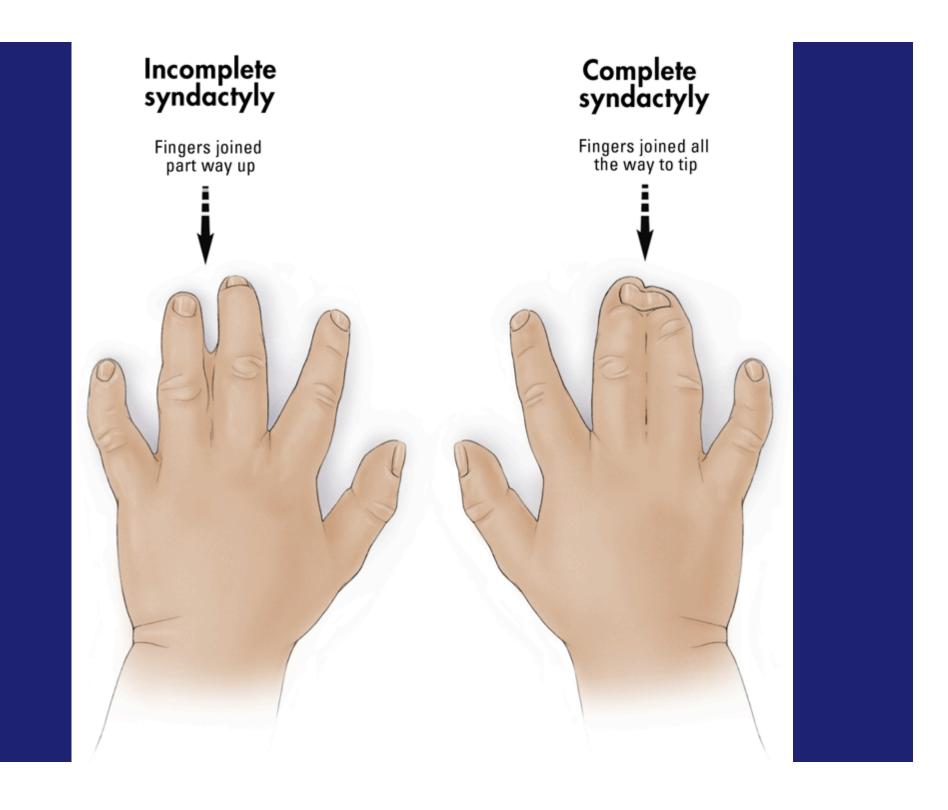


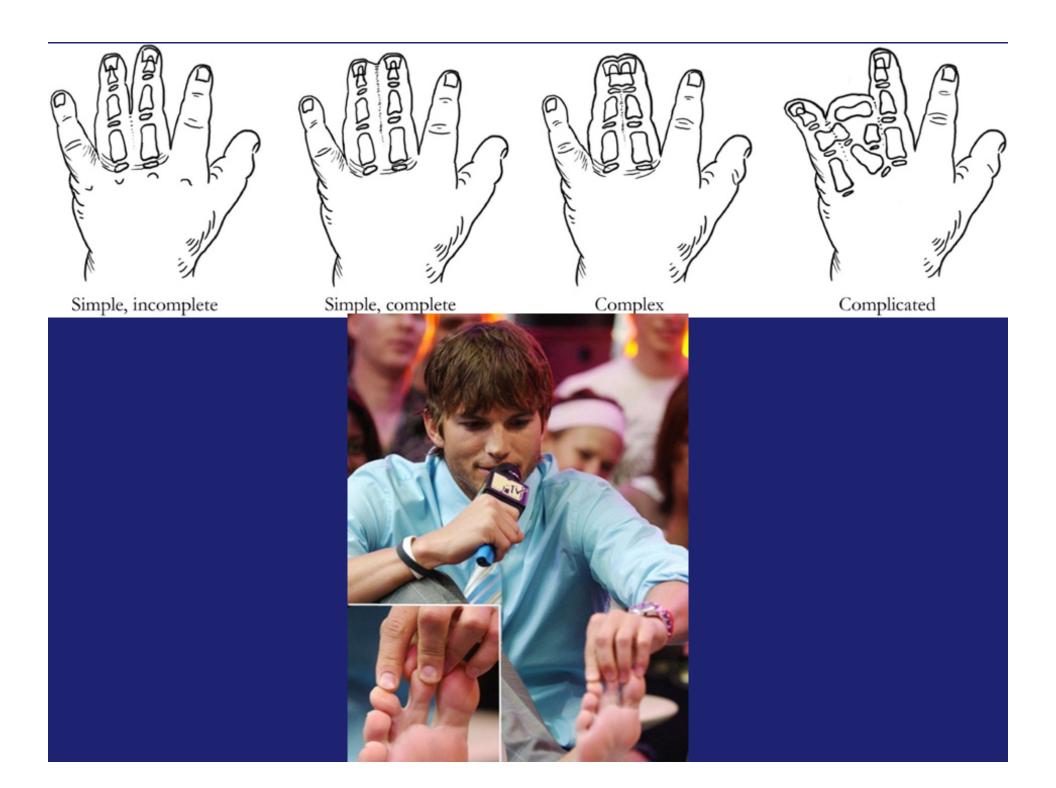
Central poly(syn)dactyly

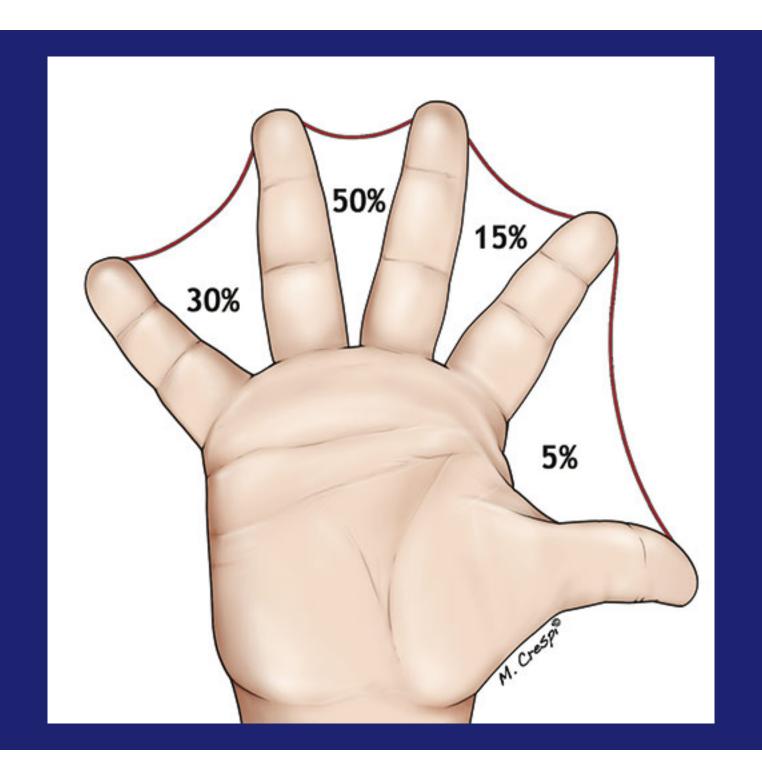


Syndactyly

- Abnormal interconnection between adjacent digits
- Very common
- Incidence 1 per 2000 to 3000 live births
- Inheritance AD or sporadic
- Surgery indicated for all cases
- Timing
 - 12 to 18 months of age

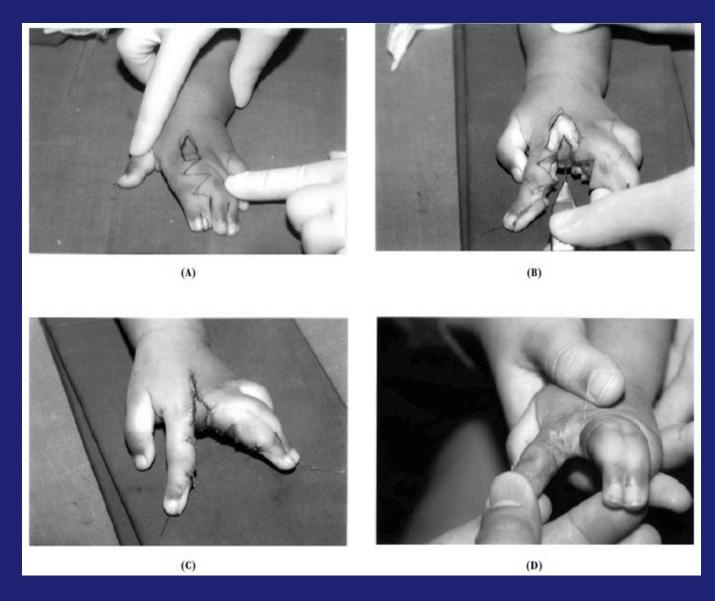






Incomplete syndactyly

- Flaps
- Less skin deficiency



Single-Stage Separation of 3- and 4-Finger Incomplete Simple Syndactyly With Contiguous Gull Wing Flaps: A Technique to Minimize or Avoid Skin Grafting

Xiaofei Tian, MD,*† Jun Xiao, MD,*† Tianwu Li, MD,*† Wei Chen, MD,*†

Qiu Lin, MD,*† Harvey Chim, MD‡

Purpose Staged separation of 3- and 4-finger syndactyly is commonly performed owing to concerns about vascular supply to the central digit and availability of flap skin. We performed single-stage separation of patients with incomplete syndactyly of multiple digits with adjacent contiguous dorsal gullwing flaps and avoided skin grafts in the majority of cases.

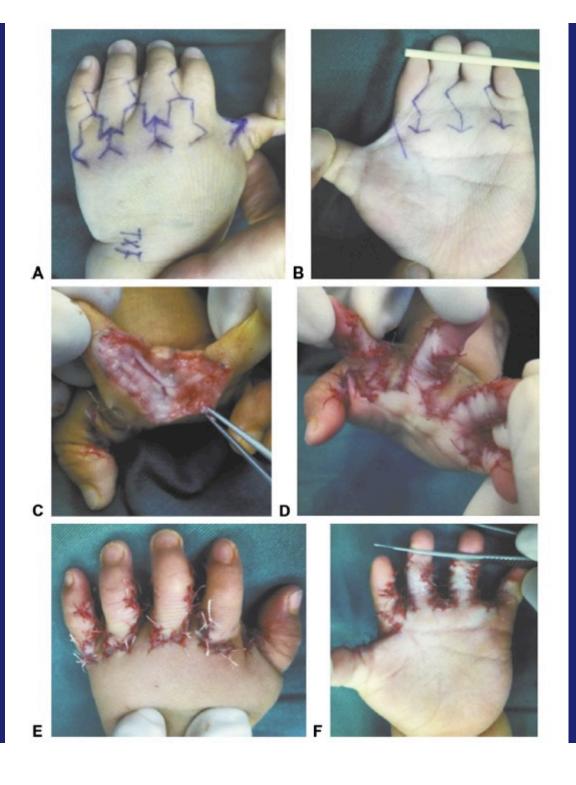
Methods Seventy-four webs of 31 patients with more than 2-finger incomplete syndactyly were included. Median age at surgical separation was 12 months (range, 5—123 months). All cases were incomplete syndactyly that did not extend to the fingernail level, with no bony involvement. A dorsal gullwing flap was used for all cases, which reconstructed the interdigital webs and partly covered the lateral side of the proximal phalanx. The technique relies on perfusion of the flap through the dorsal metacarpal artery perforator to aid flap mobility and double radial and ulnar z-plasties on each side of the flap to aid flap advancement. Skin grafts were needed if there were any remaining skin defects.

Results In 30 of 31 cases, a single-stage procedure was accomplished. One case was staged owing to abnormal digital arterial anatomy found on exploration. No skin graft was required in 21 out of 31 patients (67.7%). Median postoperative follow-up was 12 months (range, 6–36 months). All finger web depths were normal or slightly deepened.

Conclusions One-stage separation for 3- and 4-finger syndactyly with a dorsal gullwing flap is feasible and safe as long as at least 1 proper digital artery is preserved in each finger. The need for skin grafting is minimized. (*J Hand Surg Am. 2017;42(4):257–264. Copyright* © 2017 by the American Society for Surgery of the Hand. All rights reserved.)

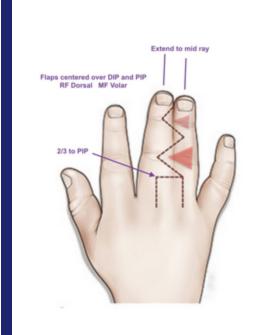
Type of study/level of evidence Therapeutic IV.

Key words Syndactyly, Poland syndrome, congenital hand, synbrachydactyly.

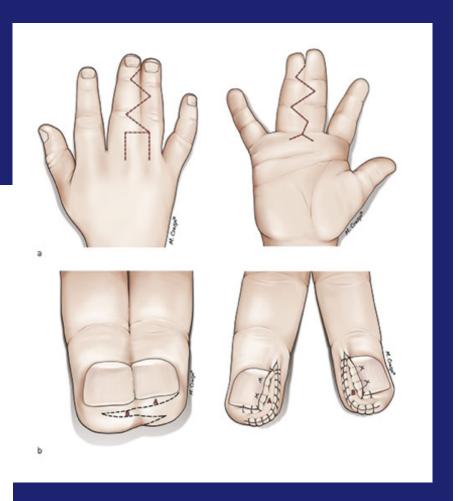


Complete syndactyly

- More skin deficiency
- Skin grafts







Hypoplastic thumb

- Can be present in isolation or with radial deficiency
- Second most commonly encountered thumb anomaly after thumb duplication
- Rare- 1 in 100,000 infants
- Equal in males and female
- 60%- both thumbs affected

Syndromes

- Holt-Oram
 - Hand-heart syndrome
- VATER (vertebral, anal, tracheal, esophageal, phalangeal and renal)
- Fanconi anemia
- Thrombocytopenia absent radius syndrome (TAR)

Workup

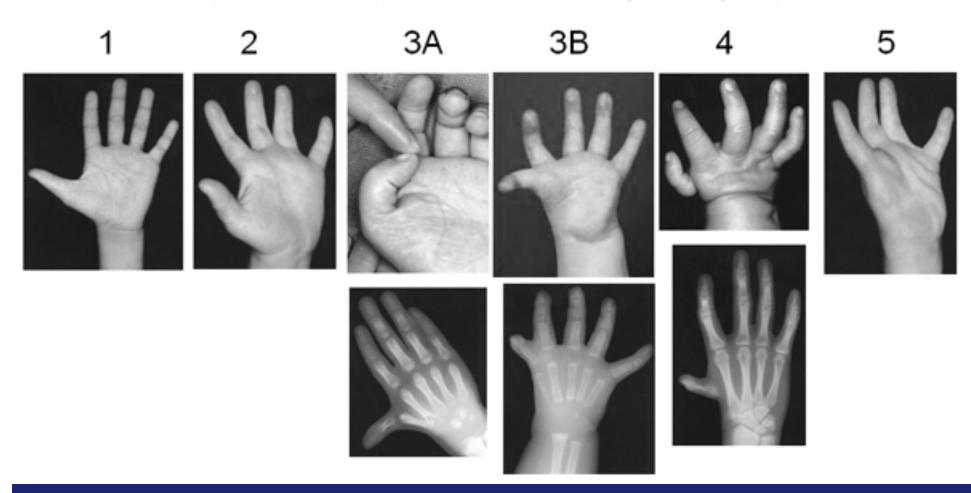
- XR hands
- Genetic testing
- Echocardiogram
- Renal ultrasound

Thumb Hypoplasia/Aplasia I. II. Narrowed veeb space Smaller bones Underferviloped or absent thumb muscles IV. V. No supporting connection for detail bones Absent thumb muscles Absent thumb

- Type 1
 - Minor hypoplasia
 - Everything smaller in size
- Type 2
 - MCP jt ulnar collateral ligament instability
 - Thenar hypoplasia
- Type 3
 - Absence of active motion at MCP or IP joint
 - A- CMC joint intact
 - B- Deficient CMC joint
- Type 4
 - Pouce Flouttant
- Type 5
 - Complete absence of thumb

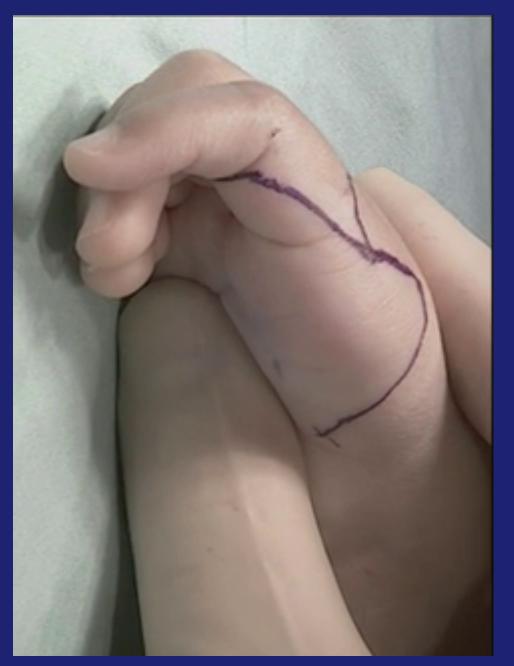
Thumb Hypoplasia Classification

Blauth and Schneider-Sickert Classification, Modified, 2004



Treatment

- Depends on severity
- Type II, IIIA
 - Stabilization of MCP joint
 - Deepening of 1st webspace
 - Opponensplasty
- >=Type 3B: pollicization
- Timing
 - 12 to 18 months of age









Radial deficiency

- Ranges from mild thumb hypoplasia to complete absence of the radius
- Forearm shortening
- Radial deviation of the wrist or hypoplasia
- Absence of a thumb

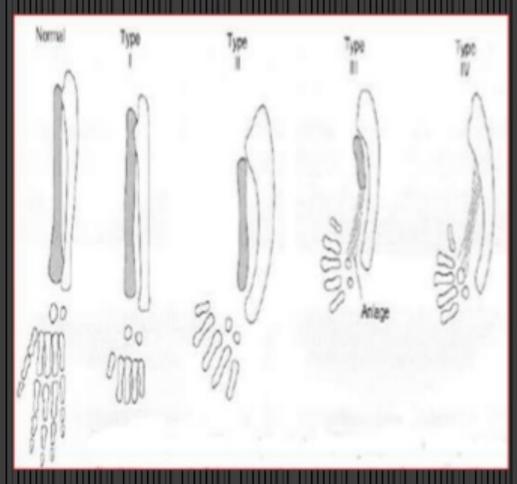


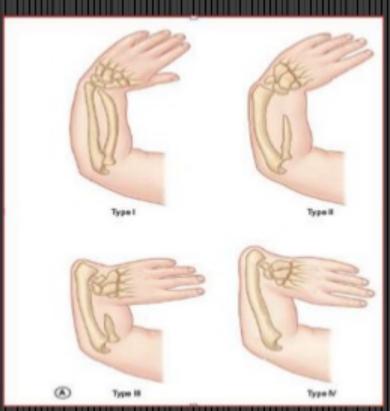


ABLE III Global Classification of Radial Longitudinal Deficiency					
Туре	Thumb Anomaly	Carpal Anomaly*	Distal Part of Radius	Proximal Part of Radius	
N	Absence or hypoplasia	Normal	Normal	Normal	
0	Absence or hypoplasia	Absence, hypoplasia, or coalition	Normal	Normal, radioulnar synostosis, or radial head dislocation	
1	Absence or hypoplasia	Absence, hypoplasia, or coalition	>2 mm shorter than ulna	Normal, radioulnar synostosis, or radial head dislocation	
2	Absence or hypoplasia	Absence, hypoplasia, or coalition	Hypoplasia	Hypoplasia	
3	Absence or hypoplasia	Absence, hypoplasia, or coalition	Absence of physis	Variable hypoplasia	
4	Absence or hypoplasia	Absence, hypoplasia, or coalition	Absence	Absence	

TABLE IV Syndromes Associated with Radial Deficiency			
Syndrome	Characteristics		
Holt-Oram	Heart defects, most commonly cardiac septal defects		
Thrombocytopenia-absent-radius syndrome	Thrombocytopenia present at birth but improves over time		
VACTERL	Vertebral abnormalities, anal atresia, cardiac abnormalities, tracheoesophageal fistula, esophageal atresia, renal defects, radial dysplasia, lower-limb abnormalities		
Fanconi anemia	Aplastic anemia not present at birth; develops at about 6 yr of age. Fatal without bone- marrow transplant. Chromosomal challenge test now available for early diagnosis		

Bayne & Klug classification of radial longitudinal deficiency





Treatment

- Depends on severity
- Timing
 - 1 year to 4 years of age
- Centralization
- Soft tissue distraction
- Transfer of 2nd toe to support radial wrist
- Late (salvage) procedures

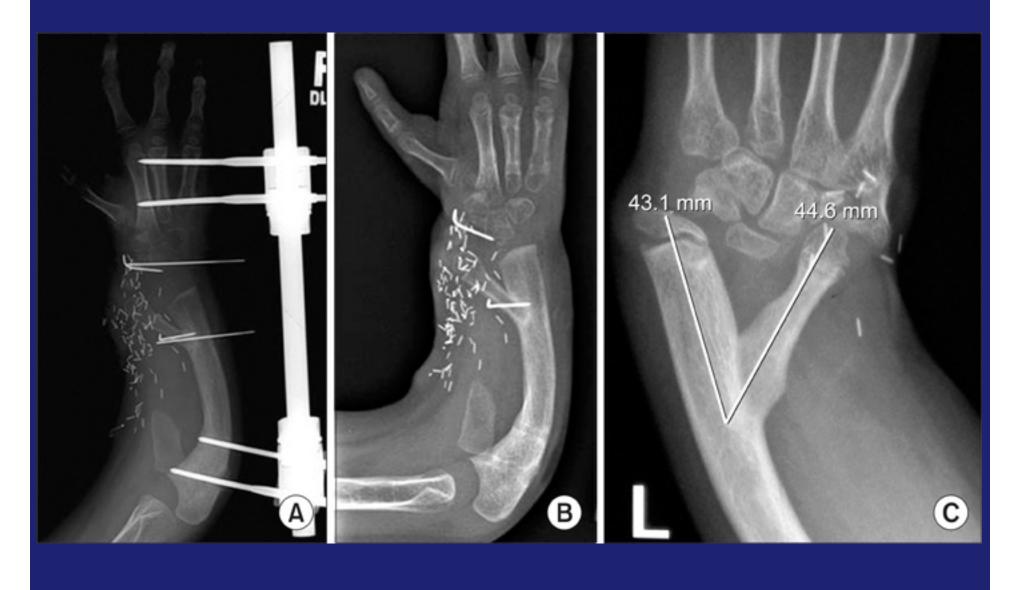
Centralization

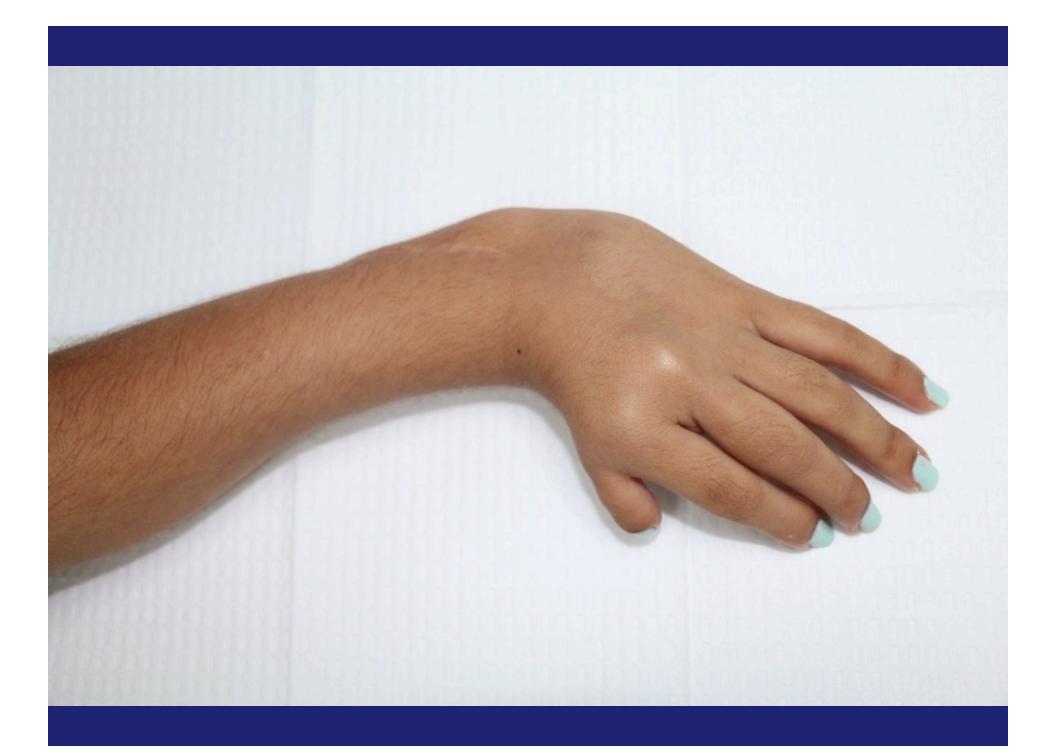


Soft tissue distraction

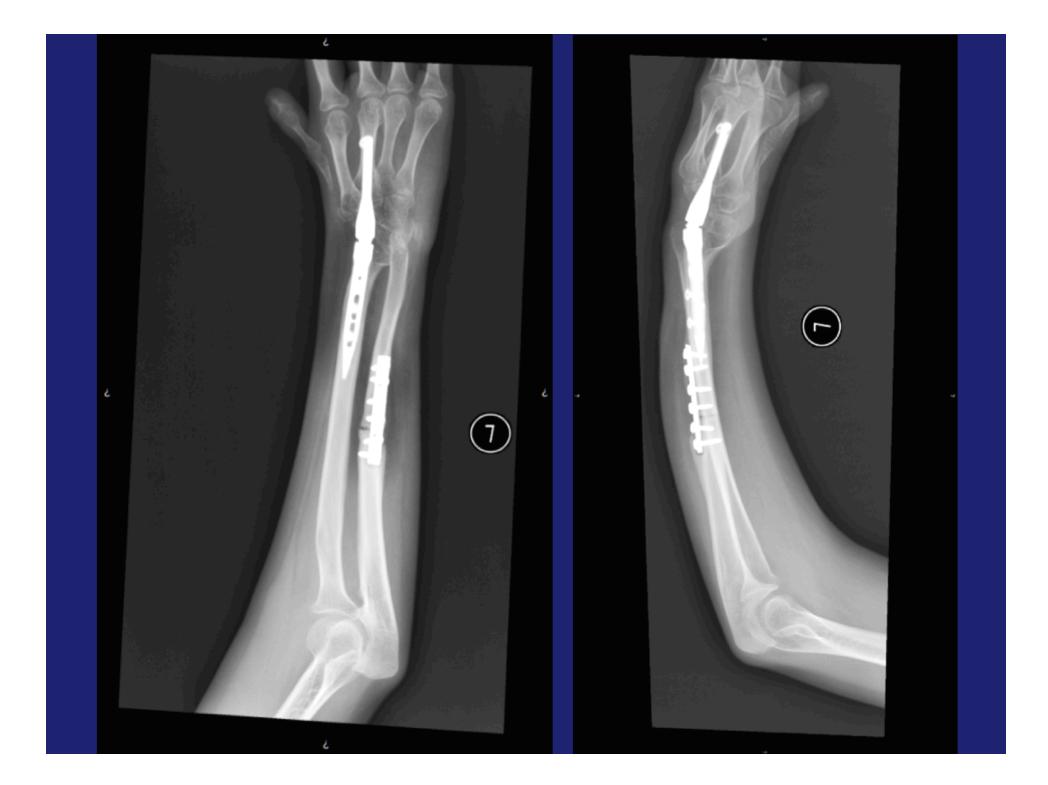


2nd toe transfer (Vilkki procedure)











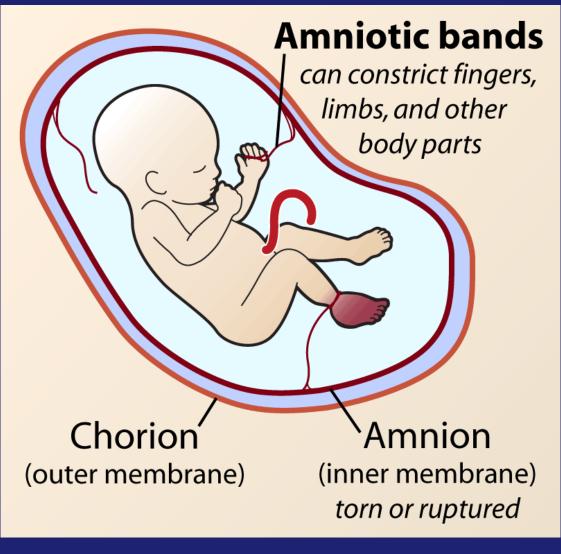


Constriction ring syndrome

- Rare
- Incidence 1 in 1200 to 15000 births
- Index, middle, ring fingers most often
- Limbs or digits of fetus become entangled with strands of amnion
- Part of finger distal to constriction ring often small or absent
- Timing
 - Soon after birth (if circulation compromised)
 - 6 to 12 months of age











Scar

Incisions for W-Plasty (around the scar)

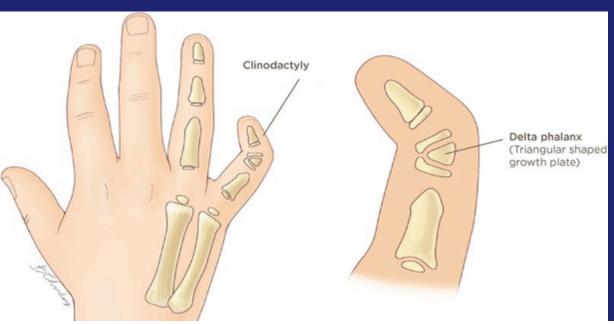
Closure of W-Plasty



Clinodactyly

- Definition
 - Abnormally bent or curved finger
- Rare (3%)
- 1 in 4 children born with Downs syndrome
- Males
- Small finger





Classification

- Type 1
 - Minor angulation with normal length
 - Most common
- Type 2
 - Minor angulation with short length
- Type 3
 - Significant angulation and delta phalanx
 - C-shaped epiphysis
 - Longitudinal bracketed diaphysis

Treatment

- Surgery
 - More than 30 degree curvature
- Physiolysis and fat interposition
 - Ideally less than 5 years of age
- Opening wedge osteotomy
 - Older children

Camptodactyly

- Congenital flexion
 deformity that usually
 occurs in the PIP joint of
 the small finger
- Causes
 - Abnormal lumbrical insertion/ origin
 - Abnormal FDS insertion
- Genetics
 - Sporadic
 - AD



Treatment- Splinting

Type

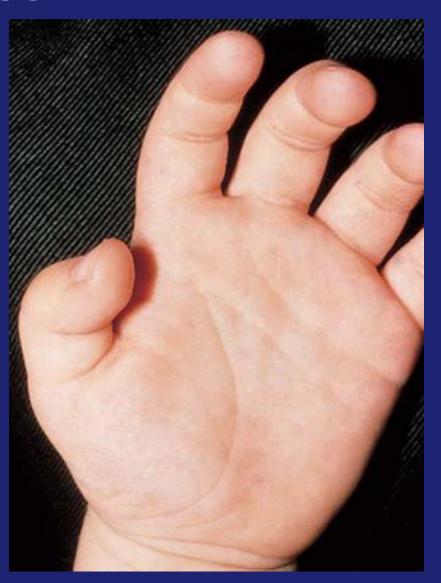
- 1
 - Isolated anomaly small finger
 - Presents in infancy
- 2
 - Presents in adolescence
- 3
 - Severe contractures, multiple digits
 - Presents at birth

Treatment

- Stretching/Splinting
 - Best for PIP contracture < 30 deg
- FDS tendon explored to radial lateral band
- Non-operative

Congenital trigger thumb

- Abnormal flexion at IP joint thumb
- 3 per 1000 children by age 1 year
- 25% bilateral
- Treatment
 - Conservative/ splinting
 - 50 to 60% resolution
 - Surgery if not better by 2 years



More congenital hand anomalies

- Cleft hand
- Macrodactyly
- Arthrogryposis
- Synostosis (radioulnar, elbow)
- Amputations/ failure of formation

Treatment

- Based on restoring function
- Individualized for each child





Daily Mail (UK) 12/22/16

 "Man, 32, born without a hand undergoes 'world first' surgery to have new limb attached

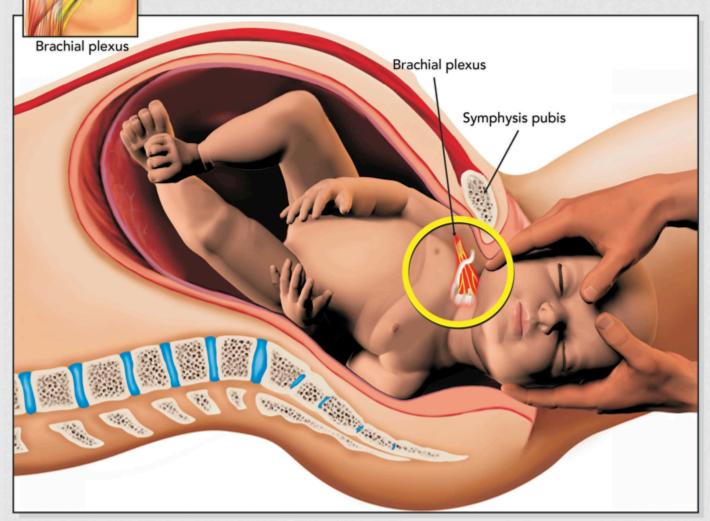


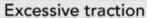
Brachial plexus birth injuries

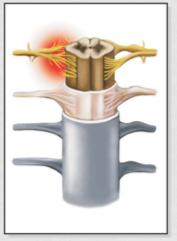
Brachial plexus birth injury

- 0.5 to 4.64 every 1000 births
- Often related to difficult delivery
 - Breech
 - Shoulder dystocia
 - Macrosomia
- Many patients have no movement of the arm at birth
- Most improve spontaneously

BRACHIAL PLEXUS INJURY



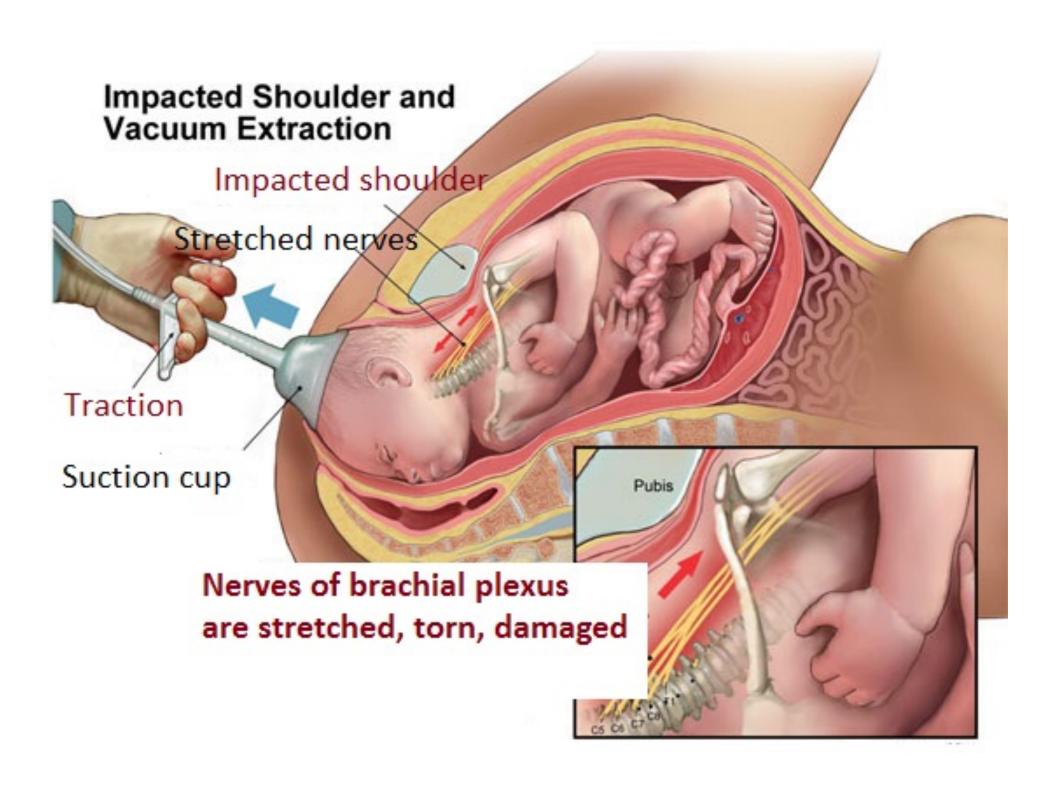




Torn nerve endings at the base of the spinal cord



Stretched nerves of the brachial plexus

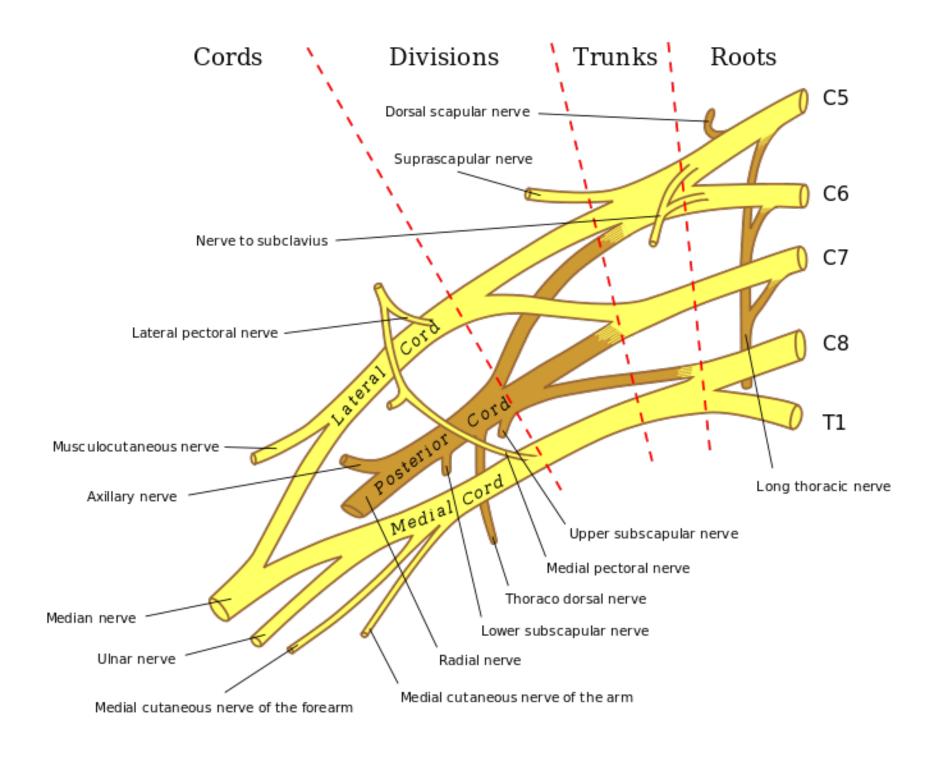


Symptoms & Signs

- Decreased or absent movement of upper extremity at birth
- Physical Examination
 - Severity of the injury
 - Any movement in the hand, wrist and elbow
 - Condition of shoulder and elbow
 - Acute fracture or dislocation
 - Clavicle, humerus

Treatment

- Referral to brachial plexus surgeon by 3 months of age
- Referral to brachial plexus therapist ASAP
- EMG 3 months
 - Assess recovery
- MRI 5 months
 - Surgical planning (if no recovery)



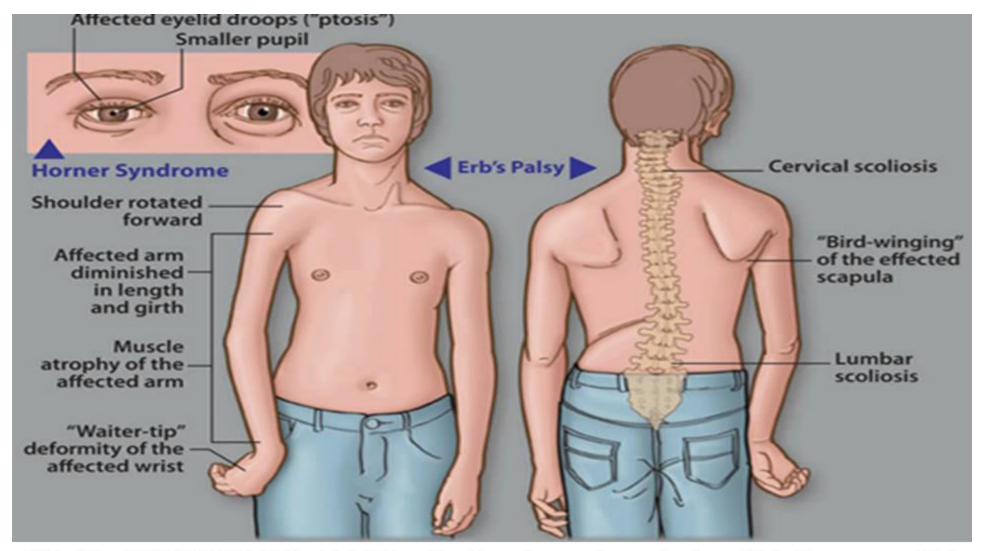
Narakas Classification

- Upper Erb's palsy (C5-C6)
- Extended Erb's palsy (C5,6,7)
- Total palsy with no Horner's sign
- Total palsy with Horner's sign

Erb's palsy

- Most common form
- No shoulder abduction
- No elbow function
- Pronated forearm
- Wrist flexed
- Movement in hands





What is ERBS PALSY - ERB'S palsy is a type of paralysis within the arm which is caused by an accident to the brachial plexus. The word brachial plexus refers to the primary network associated with nerves operating from the disposal to the entire spine. ERB'S palsy might be a result of carelessness or medical negligence at the time of the birth to the impacted child.

Surgical treatment

- Upper Erb's palsy
 - Most will recover almost complete function
 - Very few need surgery
- Extended Erb's palsy
 - Majority will recover good function
- Total palsy
 - Often will need surgery
 - Quite rare nowadays

Vancouver splint



- Prevent shoulder internal rotation contracture
- Start splinting as early as possible
- Normally up to around 1 year of age

Botox injections



- 4 months and older
- For shoulder internal rotation and adduction contracture

Internal rotation contracture



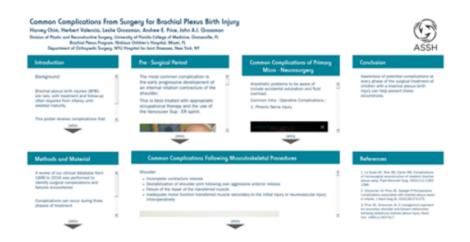
Timeline for surgery

- 6 months to 18 months: Nerve surgery
- 2 to 4 years: Shoulder surgery
- Childhood to Adolescence: Tendon transfers, secondary procedures in hand and wrist, elbow contracture release

UF Brachial Plexus Team

- Surgeons
 - Harvey Chim (Plastic Surgery)
 - Chandan Reddy (Neurosurgery)
- Pediatric Neurologist
- Hand Therapist
 - Brenda Hartwell, Portia Gardner-Smith
- Neurophysiologist
- Pathologist
- Nursing staff

Common Complications From Surgery for Brachial Plexus Birth Injury



Harvey Chim, Herbert Valencia, Leslie Grossman, Andrew E. Price, John A.I.

Grossman

Division of Plastic and Reconstructive Surgery, University of Florida College of Medicine, Gainesville, FL

Brachial Plexus Program, Nicklaus Children's Hospital, Miami, FL Department of Orthopedic Surgery, NYU Hospital for Joint Diseases, New York, NY

Reconstruction of Pediatric Brachial Plexus Injuries With Nerve Grafts and Nerve Transfers

Harvey Chim, MBBS, Michelle F. Kircher, BA, Robert J. Spinner, MD, Allen T. Bishop, MD, Alexander Y. Shin, MD

Purpose To review the demographics and injury patterns in consecutive pediatric patients with traumatic brachial plexus injury presenting to a single center over a 16-year period and to review the outcomes of nerve grafting and nerve transfers for reconstruction of shoulder abduction and elbow flexion in these patients.

Methods Forty-five pediatric patients presented for treatment of traumatic Brachial plexus injury from 1996 to 2012. Subgroup analysis of patients who had nerve grafting or nerve transfers for restoration of shoulder abduction and elbow flexion was carried out to compare outcomes of Medical Research Council (MRC) motor grading.

Results The mean age of patients was 13.8 years (range, 3–17 y). Panplexal injuries (62%) and upper plexus injuries (16%) were particularly common. There was a very high proportion of preganglionic injuries (91%). Six of the 10 of patients who underwent intraplexal nerve grafting only for restoration of shoulder abduction achieved grade 3 or better power compared with 42% (5/12) of patients who had nerve transfers. When contralateral C7 was used as a donor for nerve transfer in restoration of shoulder abduction, 1 of the 5 patients achieved grade 3 or better shoulder abduction. All 4 patients who had nerve grafts for restoration of elbow flexion achieved grade 3 or better power, compared with 11 of 12 patients who had nerve transfers. There was no statistical difference in outcome (MRC grade 3 or 4) between patients who had nerve grafts and those who had nerve transfers.

Conclusions This study shows that nerve grafts can result in similar outcomes (MRC grading) to nerve transfers for restoration of shoulder abduction and elbow flexion in traumatic pediatric BPI. The findings of this study do not support the use of contralateral C7 as a donor for nerve transfer in reconstruction of shoulder abduction in this age group. (J Hand Surg Am. 2014;39(9):1771–1778. Copyright © 2014 by the American Society for Surgery of the Hand. All rights reserved.)

Type of study/level of evidence Therapeutic IV.

Key words Brachial plexus injury, contralateral C7, nerve graft, nerve transfer, pediatric brachial plexus.

Free Functioning Gracilis Transfer for Traumatic Brachial Plexus Injuries in Children

Harvey Chim, MBBS, Michelle F. Kircher, BS, Robert J. Spinner, MD, Allen T. Bishop, MD, Alexander Y. Shin, MD

Purpose To report our technique and experience with use of free functioning muscle transfer (FFMT) in reconstruction of traumatic brachial plexus injuries (BPIs) in children as well as its complications and outcomes.

Methods Twelve patients with complete BPI underwent FFMT for reconstruction between 2000 and 2012. Eight had single-stage gracilis transfer for restoration of elbow flexion, and 4 children had double free gracilis muscle transfer for restoration of elbow flexion and prehension. Mean duration of follow-up was 27 months (range, 14–55 mo).

Results Eleven out of 12 patients achieved at least M3 elbow flexion, with 8 patients achieving M4 or greater elbow flexion. Eight of 12 patients had nerve transfers to the musculocutaneous nerve. Mean active elbow arc of motion was 79° (range, 30°–130°). Two patients aged 8 and 11 years with open growth plates developed elbow joint contractures, which limited range of motion, but they recovered M4 and M5 elbow flexion strength.

Conclusions FFMTs can result in good outcomes following reconstruction for traumatic BPI. The use of FFMT should be carefully considered in children prior to skeletal maturity because of the risk of the development of an elbow flexion contracture. (*J Hand Surg Am. 2014; 39(10):1959–1966. Copyright* © 2014 by the American Society for Surgery of the Hand. All rights reserved.)

Type of study/level of evidence Therapeutic IV.

Key words Brachial plexus injury, gracilis transfer.



Thank you!

Questions?

Email: harvey.chim@surgery.ufl.edu