Congenital Hands

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- Occupational therapist
- Children’s Hand therapist
- innovator
- teacher
- co founder of the South Manchester Hand Therapy Clinics
Classification is of very little value
it doesn’t dictate the treatment
it doesn’t tell you the aetiology
(Wassel an exception)

Discribe what you see in each patient
Identify known syndromes
Predict the growth pattern and prognosis
Consider the child as a whole
Discuss at length with the parents

Make your treatment plan with the input of your team
integrating with the child’s other medical and developmental needs
I. FAILURE OF FORMATION

A. TRANSVERSE ARREST

1. Shoulder
   a. Shoulder level (scalp)
   b. Clavicle

2. Upper arm
   a. Upper arm level
      (1) Long above elbow
      (2) Short above elbow

3. Elbow
   a. Elbow level

4. Forearm
   a. Forearm level
      (1) Long below elbow
      (2) Short below elbow

5. Wrist
   a. Wrist level (arched)

6. Carpal
   a. Carpal level (no metacarpals present)
      (1) Proximal carpal row
      (2) Distal carpal row

7. Metacarpal
   a. Metacarpal level (dactyls)

8. Phalanx
   a. Phalangeal level
      (1) Proximal level
      (2) Middle level
      (3) Distal level

B. LONGITUDINAL ARREST

1. Radial ray (proximal)
   a. Radial ray deficiency
      (1) Normal radius
      (2) Hypoplasia of radius (complete but small)
      (3) Partial absence of radius (distal and absent)
      (4) Complete absence of radius

2. Ulnar ray (proximal)
   a. Ulnar ray deficiency
      (1) Normal ulna
      (2) Hypoplasia of ulna (complete but small)
      (3) Partial absence of ulna (distal and absent)
      (4) Complete absence of ulna

3. Intersegmental (intercalated) type of longitudinal arrest
   a. Phalangeal
      (1) Proximal type (hand - no - forearm - no - hand)
      (2) Distal type (hand - no - forearm - no - hand)
      (3) Total type (hand - no - forearm - no - hand)
   b. Other

II. FAILURE OF DIFFERENTIATION (SEPARATION) OF PARTS

A. SOFT TISSUE INVOLVEMENT

1. Disseminated
   a. Anechogenic (including multiple congenital)
      (1) Severe
      (2) Moderate
      (3) Mild

2. Shoulder
   a. Disassociated shoulder
      (1) Suprascapular shoulder
   b. Absence of these muscles (including Phalen's syndrome)
      (1) Pronator major
      (2) Pronator major and minor
      (3) Other

3. Elbow and forearm
   a. Absent muscle
      (1) Absent musculature of long extrinsic muscles
      (2) Absent musculature of long extrinsic tendons
      (3) Absent musculature of the hand
      (4) Other

4. Wrist and hand
   a. Complex syndactyly (complete and incomplete)
      (1) Radial (1st intermetacarpal space)
      (2) Central (2nd-3rd intermetacarpal space)
      (3) Ulnar (3rd-4th intermetacarpal space)
      (4) Combination of (1) + (2) or (1) + (3)
   b. Congenital flexion contractures (complex dislocation)
      (1) Ring flexion
      (2) Other
   c. Hypoplastic thumb and/or index fingers
      (1) Hypoplastic thumb
      (2) Hypoplastic index fingers
      (3) Hypoplastic middle fingers
      (4) Hypoplastic ring fingers
      (5) Hypoplastic little fingers

B. SKELETAL INVOLVEMENT

1. Shoulder
   a. Congenital humerus drink
   b. Other
2. Elbow
   a. Elbow syndrome
      i. Lateral epicondylitis
      ii. Medial epicondylitis
      iii. Cubital tunnel syndrome
   b. Elbow epiphysis (joint segmentation process)

3. Forearm
   a. Radial nerve palsy
      i. Wrist motor innervation
      ii. Wrist motor innervation
   b. Distal radial epiphysis

4. Wrist and hand
   a. Synostosis of carpal bones
      i. Lange-Fraccaro syndrome
      ii. Carpal-dactyly syndrome
      iii. Hydronephrosis syndrome
      iv. Others
   b. Synostosis of metacarpal bones
      i. Ring-nail syndrome
      ii. Others
   c. Synostosis of phalanges (proximal, middle, or distal phalanges)
      i. Radius (1st-2nd rays)
      ii. Central (3rd, 4th, 5th rays)
      iii. Ulna (3rd, 4th, 5th rays)
      iv. Others
   d. Syndactyly
      i. Partial syndactyly
      ii. Others
   e. Congenital absence (choordactyly)
      i. Heptadactyly (sextapla)
      ii. Tetradactyly (tetrapia)
      iii. Triadactyly (tripodia)
      iv. Others
   f. Hypoplasia
      i. Proximal interphalangeal joint
      ii. Others

C. CONGENITAL TUMOROUS CONDITIONS
1. Vascular system
   a. Hemangioendothelioma
   b. Hemangioma
      i. Capillary
      ii. Port wine stain
   c. Others
   d. Venous
   e. Nervenymphoma
   f. Arteriovenous (including arteriofacial)
   g. Sympathetic
   h. Others

2. Neurologic
   a. Neurofibromatosis
   b. Neuroblastoma
   c. Others

3. Connective tissue
   a. Juvenile (neurofibromatosis) (Brachosyndactyly)
   b. Others

4. Skeletal (not including overgrowth syndromes)
   a. Osteosarcoma/hematoma (including multiple hereditary osteomas)
   b. Enchondromas
   c. Fibrous dysplasia
   d. Epiphyseal abnormalities
   e. Others

III. DUPLICATION∗
1. Whole limb
2. Hemiarth
3. Radius
4. Ulna
   a. Forearm
   b. Others

5. Digits
   a. Polydactyly
      i. Radial (proximal, 1st ray, including brachysyndactyly)
      ii. Central
      iii. Others (proximal, 1st ray)

IV. OVERGROWTH∗
1. Whole limb
   a. Hypercontrol
   b. Associated with vascular condition
   c. Others

2. Partial limb
   a. Associated with vascular condition
   b. Others

3. Digits
   a. Acrobrachydyody
   b. With associated vascular condition
   c. With scoliosis
   d. With bone or cartilage anomalies
   e. Others

V. UNDERGROWTH∗
1. Whole limb
2. Forearm and hand
3. Hand alone
   a. Cleft
   b. Polydactyly

4. Metacarpal
   a. Brachymesophalangy
      i. 1st ray
      ii. Others
   b. Others

5. Digits
   a. Brachymesophalangy
      i. Without associated absence of fingers and toes
      ii. With associated absence of fingers and toes
   b. Brachydactyly
      i. Defect of middle phalanges only
      ii. Contiguous (brachymesophalangy)
      iii. Defect of two or more phalanges
      iv. Defect of either proximal or distal phalanges
      v. Others

VI. CONSTRICTION RING SYNDROME∗
1. Focal necrosis
   a. Carotid artery (proximal or circumoral)
      i. Without lymphedema
      ii. With lymphedema
   b. Others

2. Amputation ("intrauterine")
   a. Wrist
   b. Metacarpal
   c. Finger
   d. Combination a and b or b and c
   e. Others

VII. GENERALIZED ABNORMALITIES AND SYNDROMES∗

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Place and timing of surgery

• It has to be in a suitable Paediatric environment
• Most anaesthetists still say that anaesthetic complications are less when the child is over one year this very much depends on where you work
• There are a few rare conditions where the first op should be done urgently or in the first few months eg tight amniotic bands, complete syndactyly flexing the fingers
• Co morbidity and other planned surgery usually takes priority
• Developmental issues eg pollicisation
Some known syndromes

VATER vertebral, anal, trache-osophageal, radial club
or expand to VECTRAL by adding cardial and renal

Holt-Oram radial club hand with ASD dominant gene

Increasingly advances in genetics are giving us more understanding of the aetiology
Antenatal diagnosis on ultra sound

- It is becoming increasingly common
- It must be difficult for the radiologist and obstetrician to break the news
- The parents grieving has happened earlier and they are prepared for the condition at birth
- The parents are well informed before they come to the surgeons clinic and the surgeons consultations are easier
Aims of treatment is to maximise

function (bilateral anomaly is very different to unilateral)
cosmesis (movement an important part of cosmesis)
maximise growth (a scar is a powerful deforming force during growth)

The Congenital Hand Clinic needs Therapists particularly splintage Wound dressings Psychology advise on hand
Post polydactyly
Pre axial polydactyly

Wassel I to VII
Tri phalangeal thumbs
Syndactyly
Full thickness grafts

Full thickness skin grafts can fail even in the best Units!

If you have any doubts about a graft take, redo it at 7 days there is nothing to loss and every thing to gain
Tissue expansion, does it help with syndactyly correction?
Amniotic Bands
Amniotic Bands
Neonatal surgery to release constriction
Acro-syndactyly, joined at the tips.

After release

After ‘on top’ plasty
A further index stump pulp transfer
Radial Club Hand

Type I  distal radial hypoplasia
Type II
Type III
Type IV  complete absence of radius
Initial treatment is frequent stretching and splinting.
Aphorism

Always do what your therapists tell you to do.
Pollicisation of the index finger to reconstruct the thumb
Huber transfer
ADM to APB

What is a Camitz transfer??
Left good position  Right poor position of ‘thumb’ metacarpal
Transverse arrest

Good thumb
Good first web
Some thumb to V stump pinch
Free non vascularised phalangeal transfers done in the first year of life
Free second toe transfer
Free second toe transfer
Free double second toe transfer
Free second toe to thumb
Free non-vascularised donor toes

Right non-vascularised
Left free vascularised second toe
distraction
MANCHESTER AIRPORT 21/4/2010

JUST AFTER THE NO FLIGHTS BAN WAS LIFTED!!
Plan your skin incisions and plan for the skin shortage
In a case with a pipj contracture the standard mid axial incision or the usual Brunner incision may not give you the extra skin that you need.
Z plasty
Drawing a Z-plasty

**line 1** - direction of scar or incision to be elongated
Drawing a Z-plasty

line 2 - non cutting line where the final transverse limb will be
Drawing a Z-plasty

**lines 3 and 4** - at 60° to the 1st line
Z plasty: drawing a Z plasty
Full thickness skin grafts can fail even in the best Units!
If you have any doubts about a graft take, redo it at 7 days.
There is nothing to lose and every thing to gain.
The proposed job cuts would affect clinical staff as much as administrators. They would be a £15bn shortfall in NHS funds by 2016. He added that on average 30,000 people a year retired and a substantial number left the NHS, so it was possible the vast majority to go would be front-line NHS staff,” he said.

Karen Jennings, head of health at Unison, the public service trade union,
Shared housework means fewer divorces, a study shows. So in this age of equality, who does what still cause so much angst? By Kate Burt
Thierry Henry handles the ball just before the decisive goal, which left Keith Andrews (top, right) in tears after Irish protests fell on deaf ears (bottom).

Dunne said: ‘The referee said he would have scored a bad day for football’. Brady said: ‘many are massive countries. There’s...’
We know what it takes to be a Tiger.