Congenital Hands

Stewart Watson, Manchester

Sue Kennedy

- 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 patern and prognosis
Consider the child as a whole
Discuss at length with the parents

Make your treatment plan with the input of your team
intergrating with the child's other medical and developmental needs

I. FAILURE OF FORMATION

A. Transverse arrest

1. Shoulder
- *a. Shoulder level (amelia)*
- *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 (acheiria)*

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

7. Metacarpal
- *a. Metacarpal level (adactyly)*

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

B. Longitudinal arrest

1. Radial ray (preaxial)
- *a. Radial ray deficiency*
  - (1) Normal radius
    - (a) Thumb hypoplastic - functional
    - (b) Thumb hypoplastic - nonfunctional
    - (c) Thumb absent
  - (2) Hypoplasia of radius (complete but small)
    - (a) Thumb hypoplastic - functional
    - (b) Thumb hypoplastic - nonfunctional
    - (c) Thumb absent
    - (d) Madelung's deformity
    - (e) Other
  - (3) Partial absence of radius (distal and absent)
    - (a) Thumb hypoplastic - functional
    - (b) Thumb hypoplastic - nonfunctional
    - (c) Thumb absent
  - (4) Complete absence of radius
    - (a) Thumb hypoplastic - functional
    - (b) Thumb hypoplastic - nonfunctional
    - (c) Thumb absent
  - (5) Absent/hypoplastic thenar muscles
  - (6) Absent/hypoplastic extensor muscles
  - (7) Absent/hypoplastic flexor muscles

2. Ulnar ray (postaxial)
- *a. Ulnar ray deficiency*
  - (1) Normal ulna
    - (a) Metacarpals, digits hypoplastic
    - (b) Metacarpals hypoplastic, digits absent
    - (c) Metacarpals, digits absent
  - (2) Hypoplasia of ulna (complete but small)
    - (a) Metacarpals, digits hypoplastic
    - (b) Metacarpals hypoplastic, digits absent
    - (c) Metacarpals, digits absent
  - (3) Partial absence of ulna (distal and absent)
    - (a) Metacarpals, digits hypoplastic
    - (b) Metacarpals hypoplastic, digits absent
    - (c) Metacarpals, digits absent
  - (4) Complete absence of ulna
    - (a) Metacarpals, digits hypoplastic
    - (b) Metacarpals hypoplastic, digits absent
    - (c) Metacarpals, digits absent
  - (5) Defect of ulna with humeroradial synostosis
  - (6) Absent/hypoplastic hypothenar muscles
  - (7) Absent/hypoplastic extensor muscles
  - (8) Absent/hypoplastic flexor muscles

3. Central ray (cleft hand)
- *a. Central ray deficiency*
  - (1) Typical type (deficiency type)
    - (a) Metacarpals, digits hypoplastic
    - (b) Metacarpals hypoplastic, digits absent
    - (c) Metacarpals, digits absent
  - (2) Atypical type
    - (a) Syndactylous type
    - (b) Polydactylous type
    - (c) Monodactyly
    - (d) Other

4. Intersegmental (intercalated) type of longitudinal arrest
- *a. Phocomelia*
  - (1) Proximal type (hand - to - forearm - to - trunk)
  - (2) Distal type (hand - to - arm - to - trunk)
  - (3) Total type (hand - to - trunk)
- *b. Other*

II. FAILURE OF DIFFERENTIATION (SEPARATION) OF PARTS

A. Soft tissue involvement

1. Disseminated
- *a. Arthrogryposis (including multiplex congenita)*
  - (1) Severe
  - (2) Moderate
  - (3) Mild

2. Shoulder
- *a. Undescended shoulder*
  - (1) Sprengel's shoulder
- *b. Absence of thoracic muscles (including Poland's syndrome)*
  - (1) Pectoralis major
  - (2) Pectoralis major and minor
  - (3) Other

3. Elbow and forearm
- *a. Aberrant muscle*
  - (1) Aberrant muscles of long extrinsic flexors
  - (2) Aberrant muscles of long extrinsic extensors
  - (3) Aberrant intrinsics of the hand
  - (4) Other

4. Wrist and hand
- *a. Cutaneous syndactyly (complete and incomplete)*
  - (1) Radial (1st interdigital space)
  - (2) Central (2nd/3rd interdigital space)
  - (3) Ulnar (3rd interdigital space)
  - (4) Combination of (1) ± (2) or (3)
- *b. Congenital flexion contracture (camptodactyly)*
  - (1) Fifth digit
  - (2) Others
- *c. Thumb-in-palm deformity*
- *d. Deviated finger without bony deformity (laxity secondary to differentiation of muscle ligament or capsule)*
  - (1) Radial/ulnar
    - (a) Isolated digit
    - (b) Congenital ulnar drift (including "windblown hand")
  - (2) Other
- *e. Congenital trigger digit or thumb*
- *f. Other*

5. Skin and appendages
- *a. Pterygium (webbing) of axilla or elbow*
- *b. Cutis aplasia congenita*
- *c. Congenital clubbing of nails*
- *d. Tuft nail deformity, volar nail*
- *e. Other*

B. Skeletal involvement

1. Shoulder
- *a. Congenital humerus varus*
- *b. Other*

2. Elbow
- a. Elbow synostosis
  - (1) Humeroradial
  - (2) Humeroulnar
  - (3) Total elbow
- b. Elbow ankylosis (joint segmentation present)

3. Forearm
- a. Proximal radioulnar synostosis
  - (1) Without radial head dislocation
  - (2) With radial head dislocation
- b. Distal radioulnar synostosis

4. Wrist and hand
- a. Synostosis of carpal bones
  - (1) Lunate-triquetrum synostosis
  - (2) Capitate-hamate synostosis
  - (3) Scaphoid-lunate synostosis
  - (4) Others
- b. Synostosis of metacarpal bones
  - (1) Ring-small synostosis
  - (2) Others
- c. Synostosis of phalanges (intrinsic syndactyly, complex syndactyly)
  - (1) Radial (1st – 2nd rays)
  - (2) Central (2nd – 3rd, 3rd – 4th rays)
  - (3) Ulnar (4th – 5th rays)
  - (4) Mitten hand (including Apert's hand)
  - (5) Other
- d. Symphalangism
  - (1) Proximal interphalangeal joint
  - (2) Other
- e. Congenital deletion (clinodactyly)
  - (1) Hypoplastic clinodactyly
    - (a) Little finger (including delta phalanx)
    - (b) Thumb (including delta phalanx)
    - (c) Others
- f. Hypersegmentation
  - (1) Triphalangeal thumb
  - (2) Others

C. CONGENITAL TUMOROUS CONDITIONS

1. Vascular system
- a. Hemangioma
- b. Malformations
  - (1) Capillary
    - (a) Port wine stain
    - (b) Others
  - (2) Venous
  - (3) Vein/lymphatic
  - (4) Arterial (including AV fistula)
  - (5) Lymphatic
  - (6) Others

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

3. Connective tissue
- a. Juvenile (aponeurotic) fibroma
- b. Other

4. Skeletal (not including overgrowth syndromes)
- a. Osteochondromatosis (including multiple hereditary exostosis)
- b. Enchondromatosis
- c. Fibrous dysplasia
- d. Epiphyseal abnormalities
- e. Other

III. DUPLICATION*

1. Whole limb
2. Humerus
3. Radius
4. Ulna
   - a. Mirror hand
   - b. Other

5. Digit
- a. Polydactyly
  - (1) Radial (preaxial, 1st ray, including triphalangeal thumb)
  - (2) Central
  - (3) Ulnar (postaxial, 5th ray)
  - (4) Combinations

6. Epiphyseal (extra)
- a. 1st ray
- b. 2nd ray
- c. Other

IV. OVERGROWTH*

1. Whole limb
- a. Hemihypertrophy
- b. Associated with vascular condition
- c. Other

2. Partial limb
- a. With associated vascular condition
- b. Other

3. Digit
- a. Macrodactyly
  - (1) With associated vascular condition
  - (2) With neurofibromatosis
  - (3) With bone or cartilage excrescence
  - (4) Other

V. UNDERGROWTH*

1. Whole limb
2. Forearm and hand
3. Hand alone
   - a. Entire
   - b. Partial
4. Metacarpal
   - a. Brachymetacarpia
     - (1) Fifth ray
     - (2) Other
   - b. Other
5. Digit
   - a. Brachymetacarpia
     - (1) With associated absence of thenar muscles (Poland's syndrome)
     - (2) Without associated absence of thenar muscles
   - b. Brachydactyly
     - (1) Defect of middle phalanx only (brachymesophalangia)
     - (2) Defect of two or more phalanges
     - (3) Defect of either proximal or distal phalanx
     - (4) Other

VI. CONSTRICTION RING SYNDROME*

1. Focal necrosis
- a. Constriction band (partial or circumferential)
  - (1) With lymphedema
  - (2) Without lymphedema
- b. Acrosyndactyly

2. Amputation ("intrauterine")
- a. Wrist
- b. Metacarpal
- c. Digit
- d. Combination a and b or b and c
- e. Other

VII. GENERALIZED ABNORMALITIES AND SYNDROMES*

Fig. 19.1 IFSSH classification of congenital hand anomalies. (Modified from Upton J. Congenital anomalies of the hand and forearm. In: McCarthy JG, ed. Plastic surgery. Philadelphia: WB Saunders; 1990:5240–398.)

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-osphageal,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 under standing 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 treament is to maximise

function (bilateral anomaly is very different to unilateral)

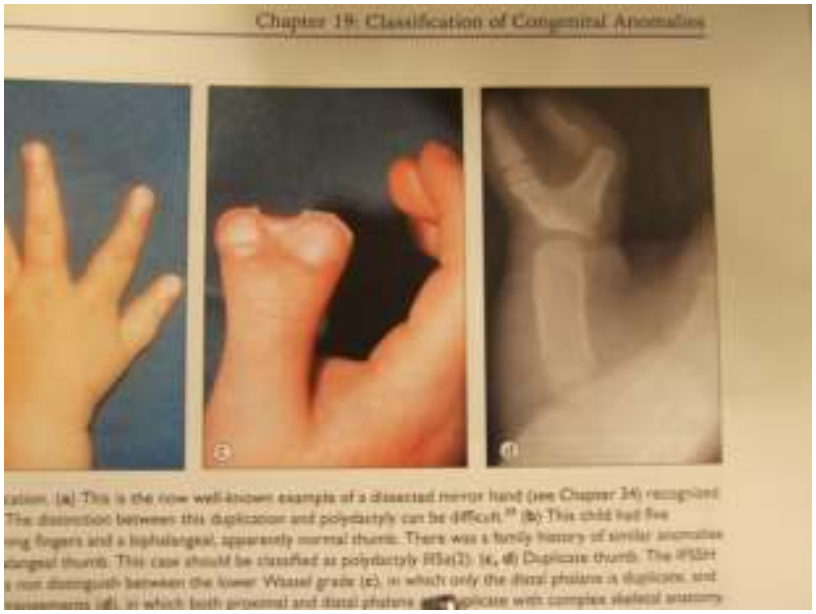
cosmesis (movement an important part of cosmesis)

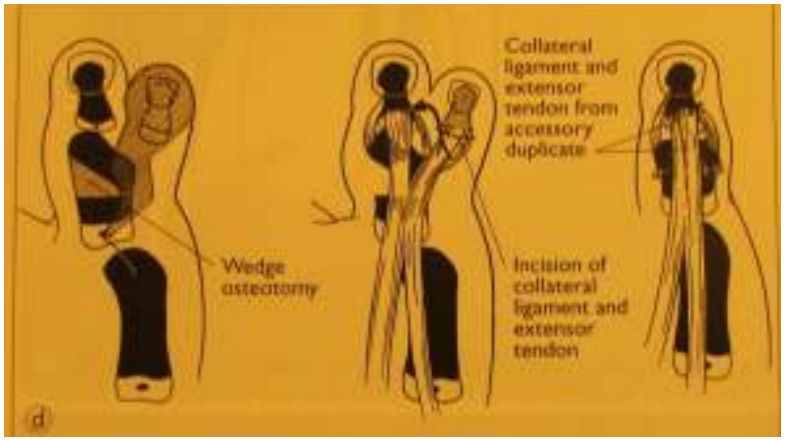
maximise growth (a scar is a powerful deforming force during growth)

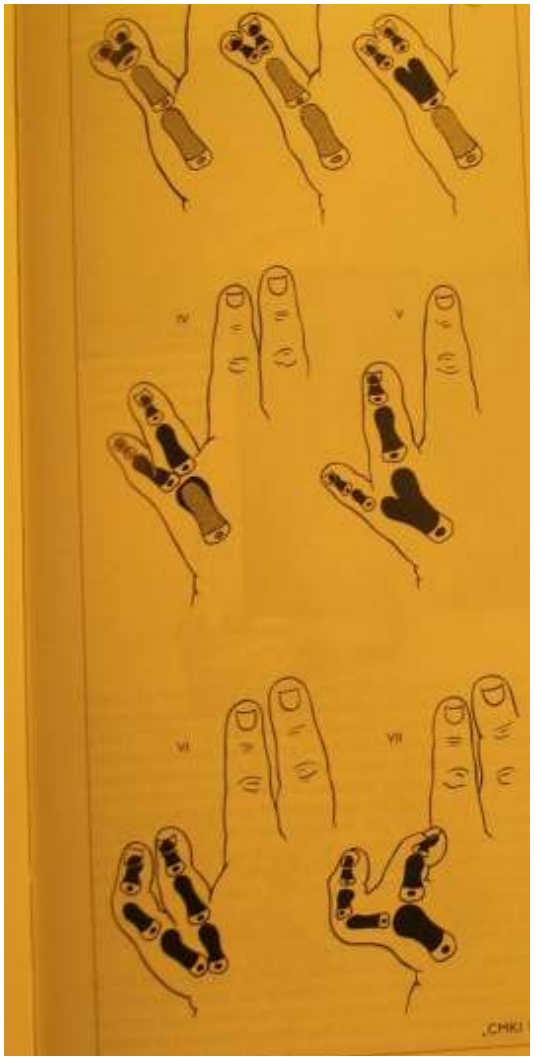
The Congenital Hand Clinic needs
Therapists particularly splintage
Wound dressings
Psychology advise on hand

Post polydactyly

Pre axial polydactyly



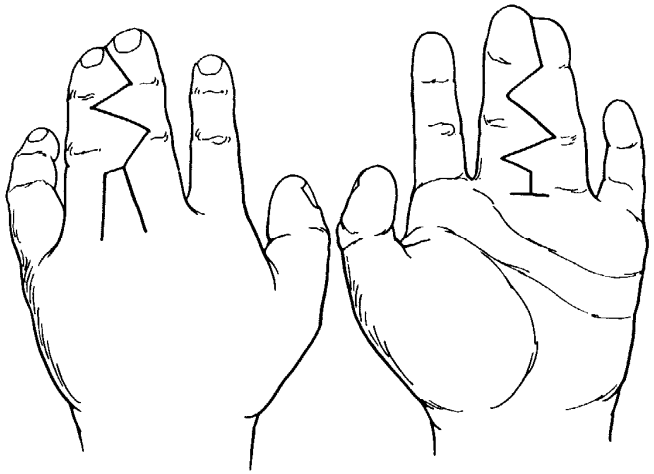


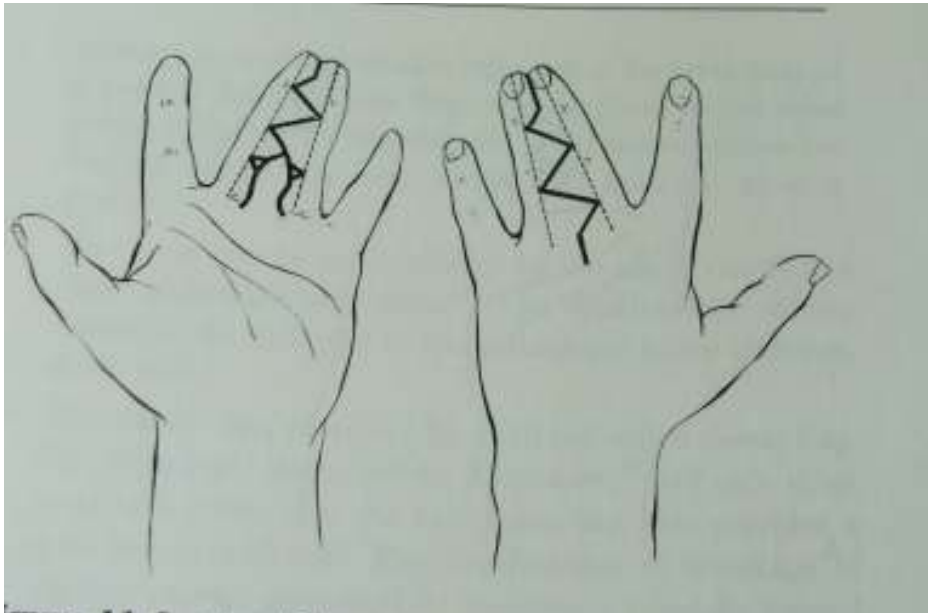


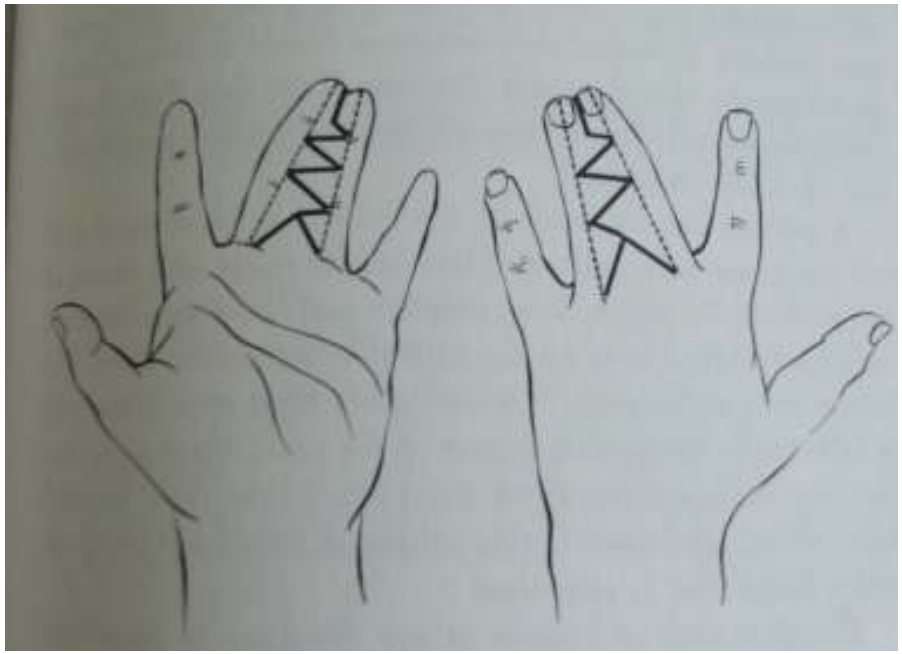
Wassel I to VII

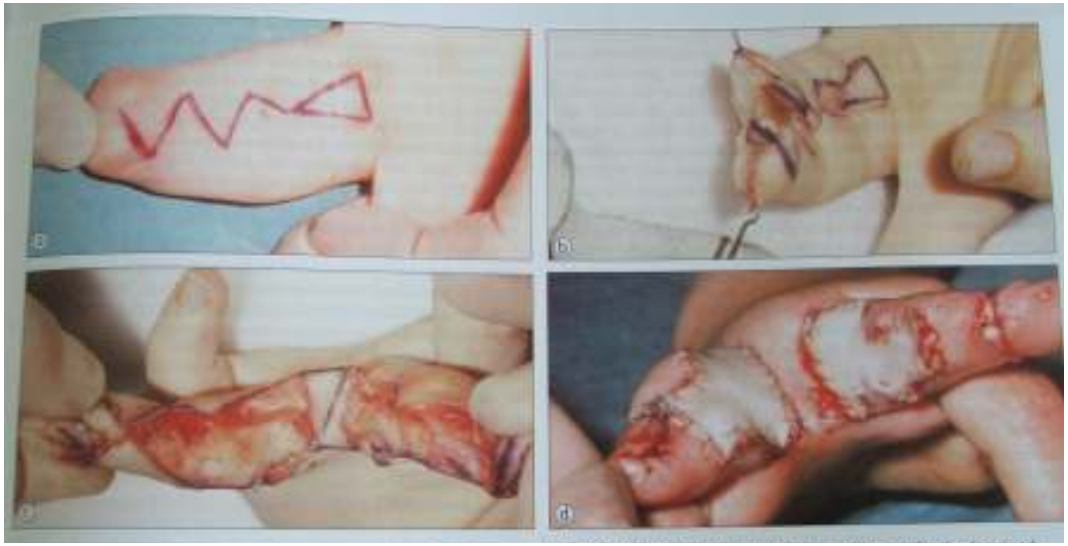
Tri phalangeal thumbs

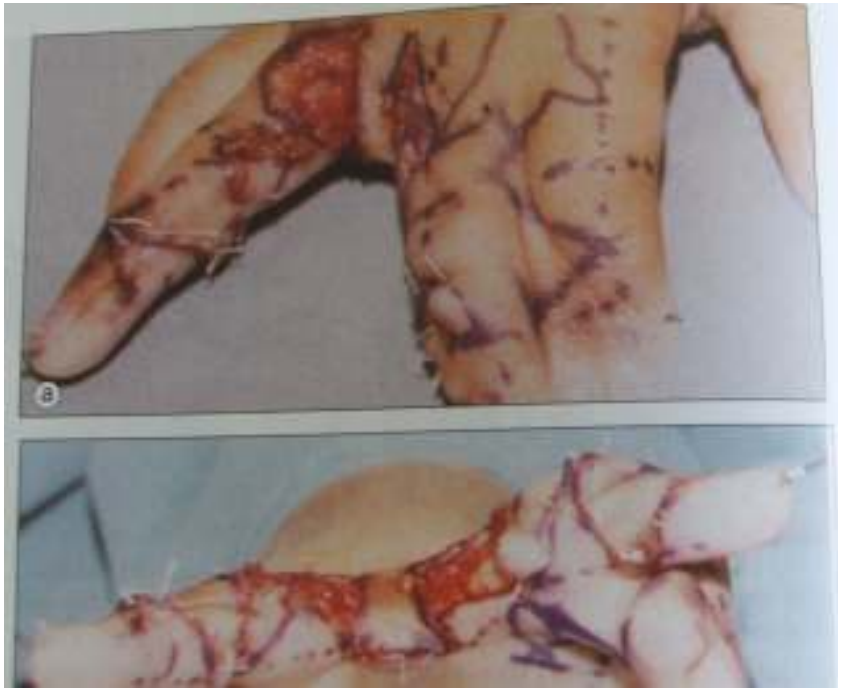
Syndactyly

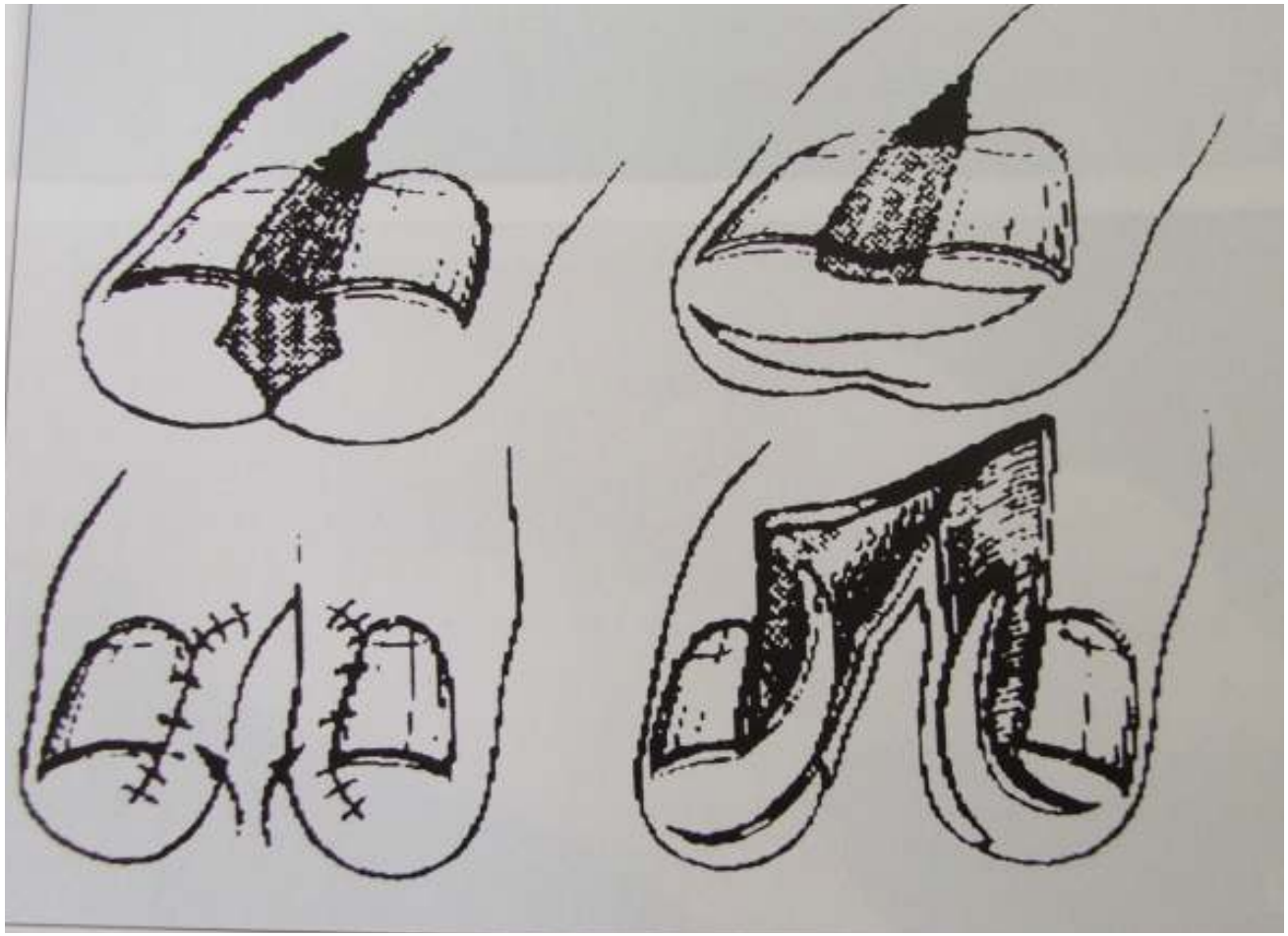












Full thickness grafts



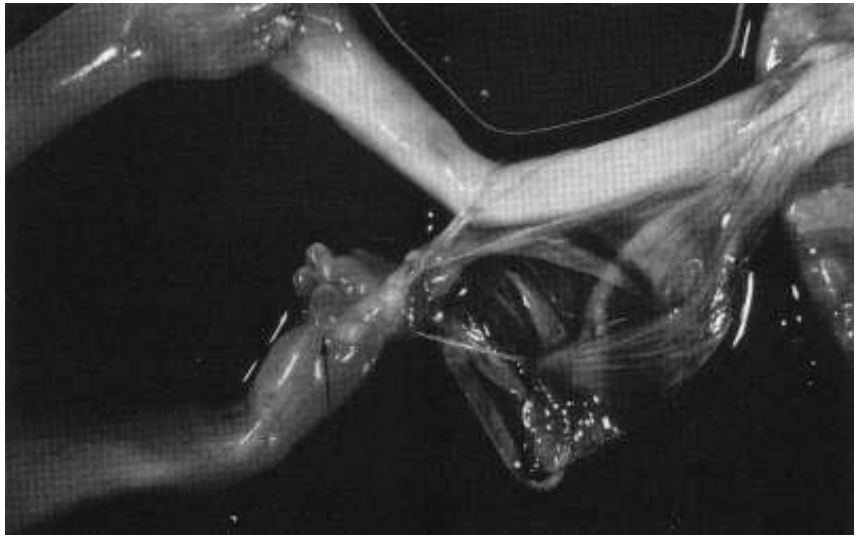

Full thickness skin grafts can fail even in the best Units!

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

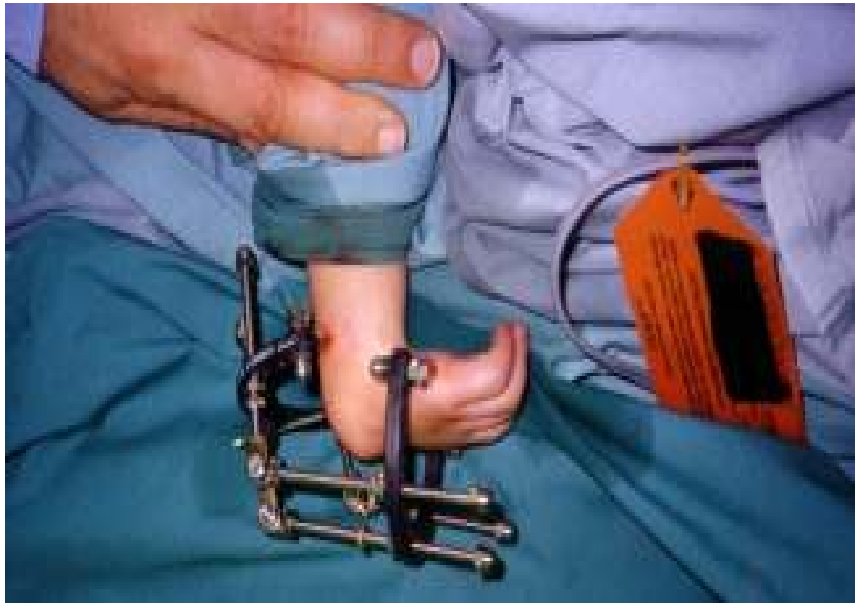
Radial Club Hand

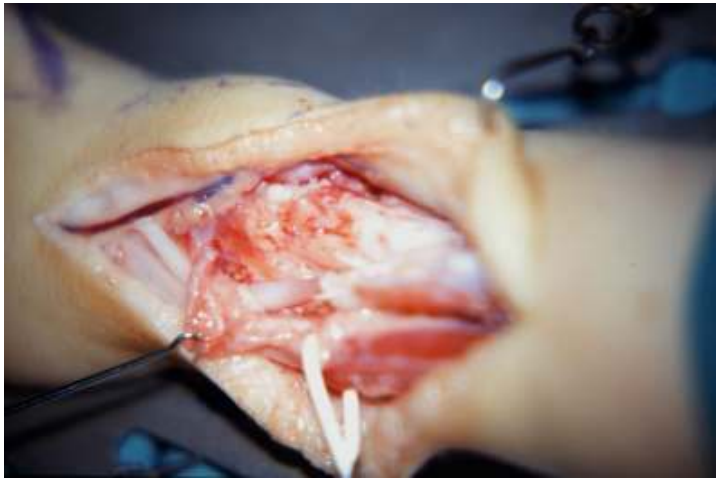
Type I distal radial hypoplasia
Type II
Type III
Type IV complete absence of radius

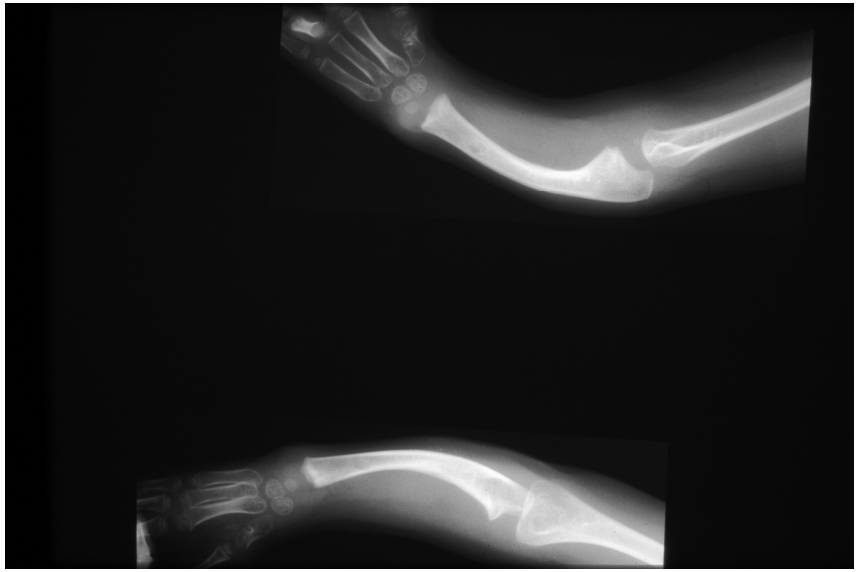
Initial treatment is frequent stretching and splinting

Aphorism

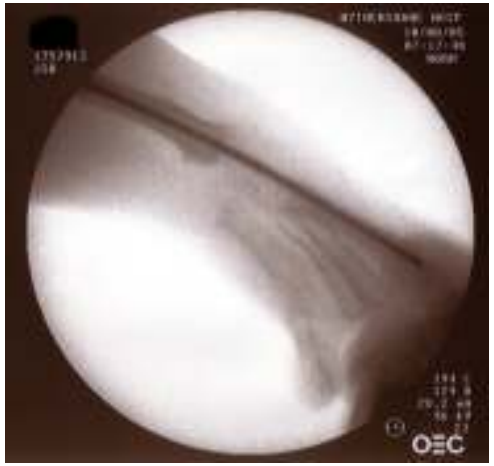
Always do what your therapists
tell you to do.








OEC


OEC

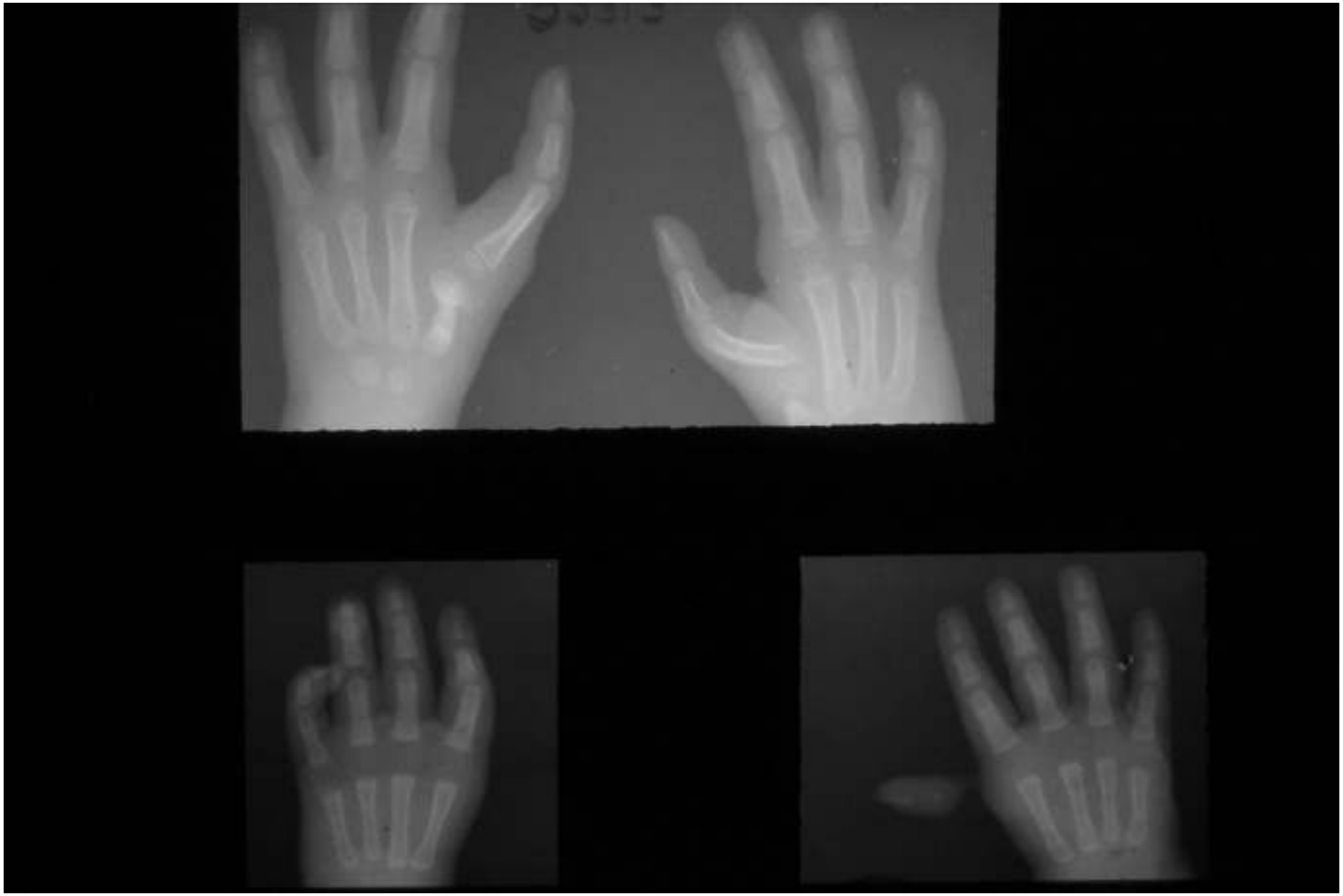

OEC


OEC

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

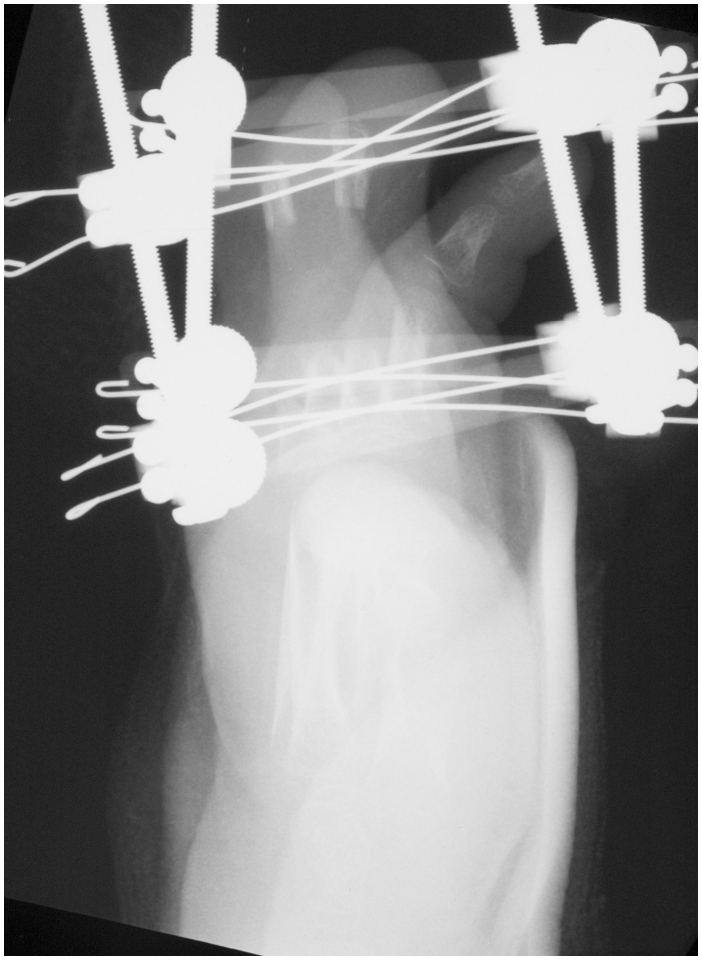
Free double second toe transfer

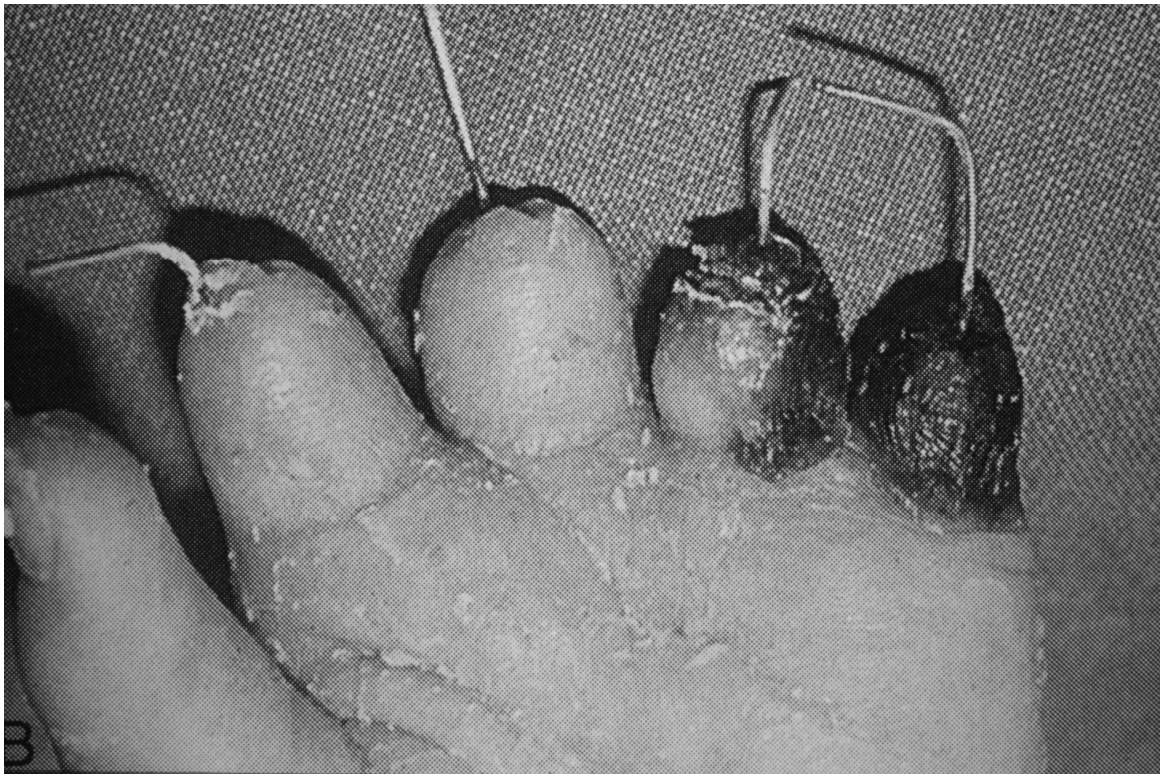
Free second toe to thumb

Free non- vascularised donor toes

Right non-vascularised
Left free vascularised second toe

distraction

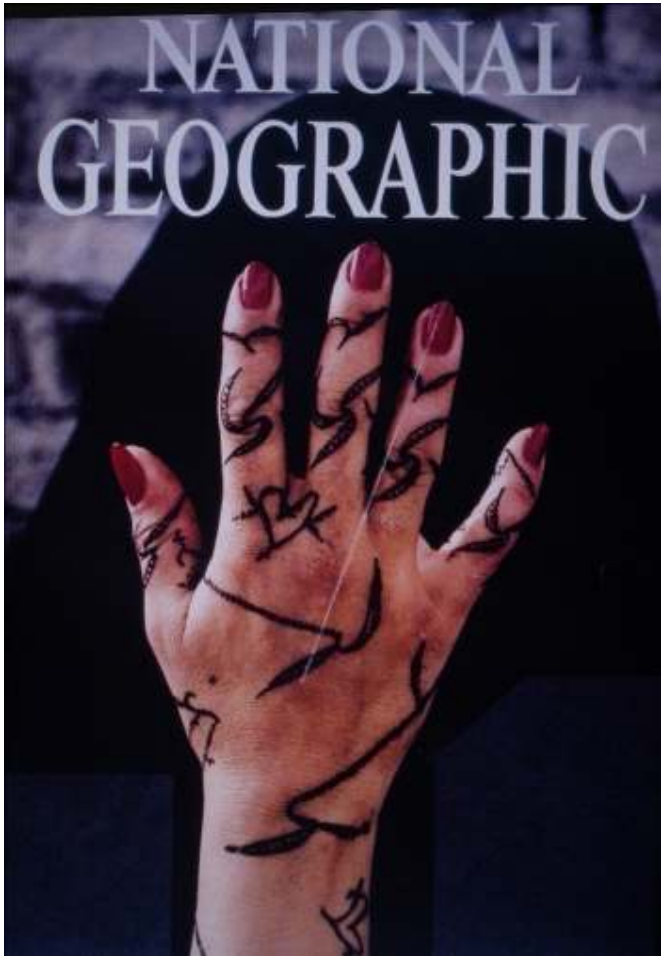



B

MANCHESTER AIRPORT 21/4/2010

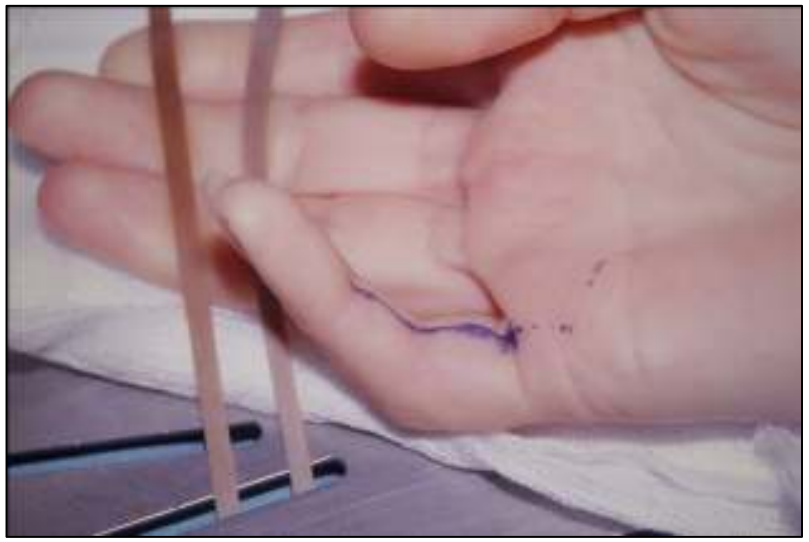
JUST AFTER THE NO FLIGHTS BAN WAS LIFTED !!

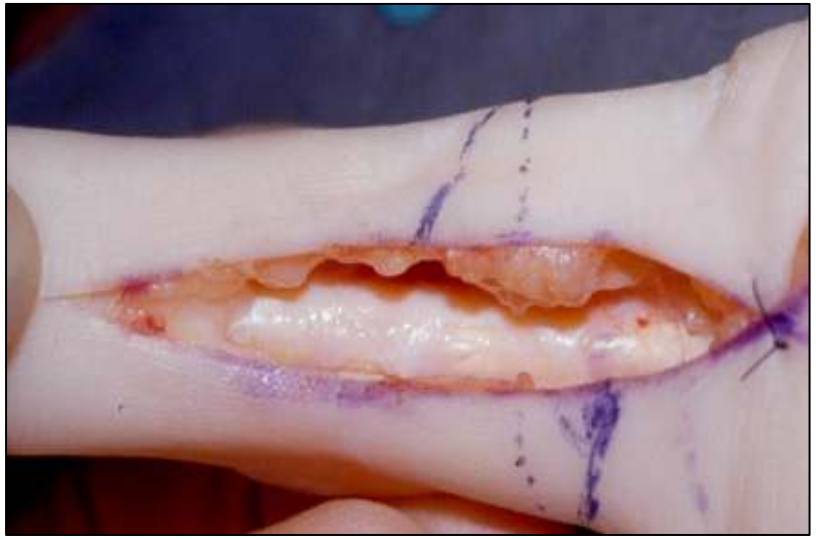
Plan your skin incisions and plan for the skin shortage



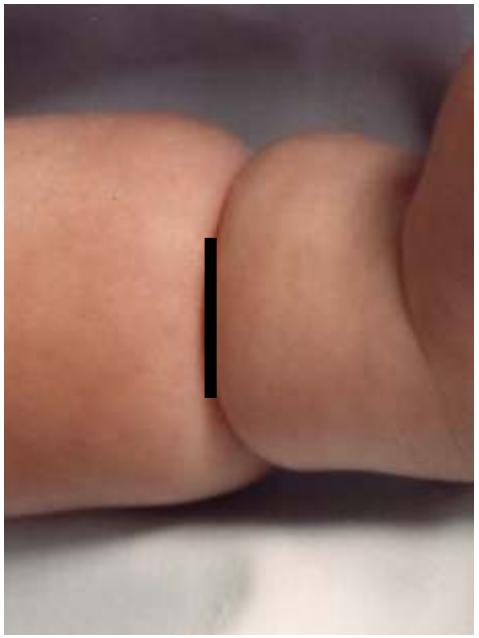
In a case with a pipj contracture the standard mid axial incision or the usual Brunner incision may not give you the extra skin that you need

Z plasty



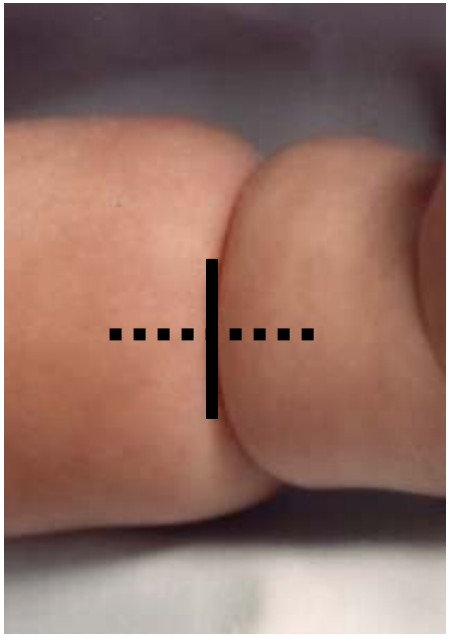


Drawing a Z-plasty



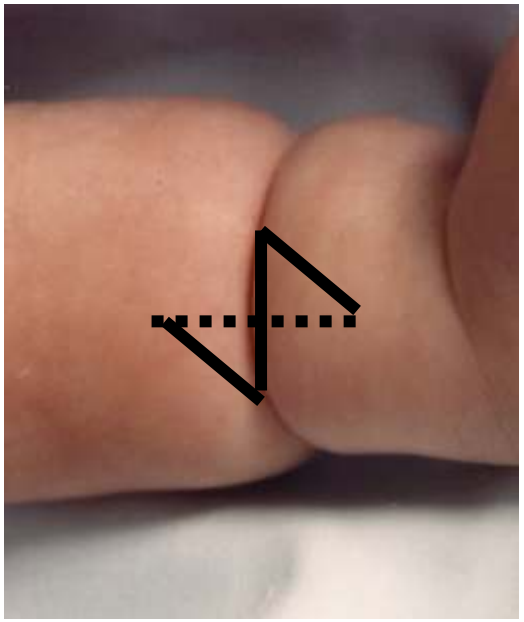
line 1 - direction of scar or incision to be elongated

Drawing a Z-plasty



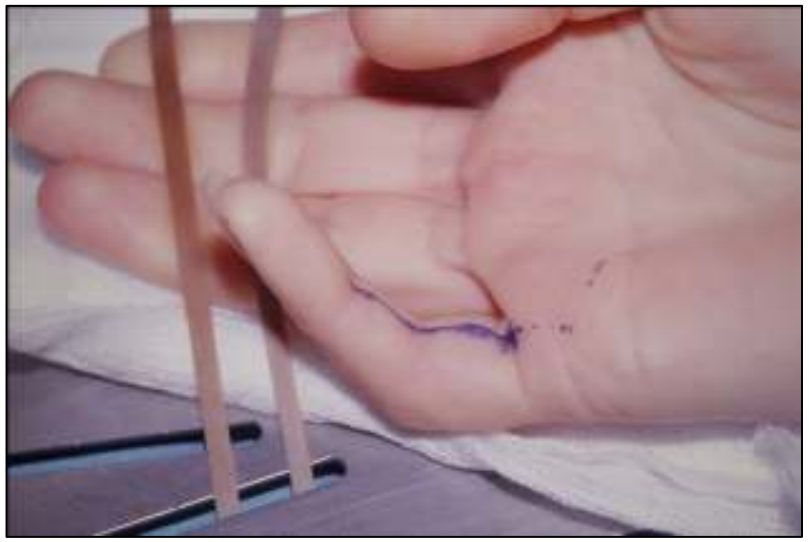
line 2 - **non cutting** line where the final transverse limb will be

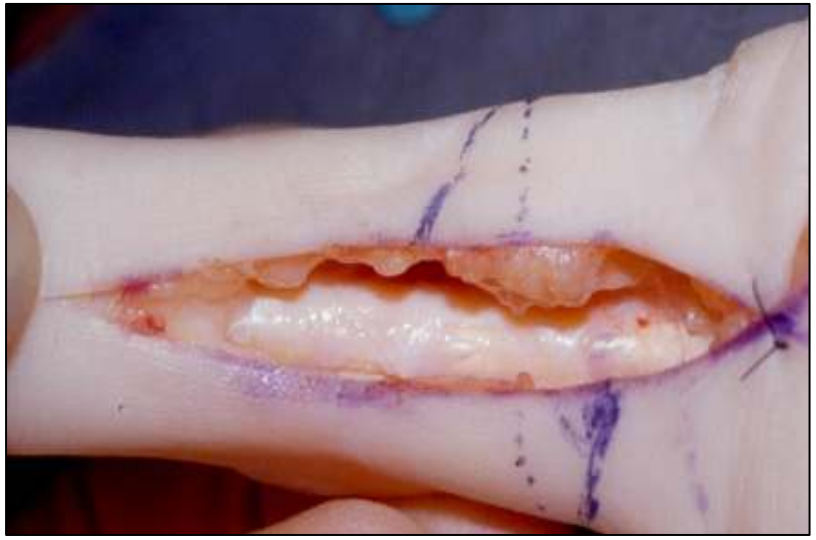
Drawing a Z-plasty



lines 3 and **4** - at 60° to the 1st line

Z plasty: drawing a Z plasty





Full thickness 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 lose and every thing to gain.

The proposed job cuts would affect clinical staff as much as administrators PA

The NHS in numbers

701,831

NHS medical staff, including 37,213 GPs; 84,595 hospital doctors; 408,160 nurses; 11,854 dentists and dental staff; 142,558 other medical staff and 17,451 members of ambulance crews

666,863

Non-medical staff, including 355,010 clinical support workers; 179,151 administrators; 39,913 senior managers; 353 other non-medical staff and 92,436 non-medical GP surgery staff

ense an NHS plan of action" but that would be considered.

Last night Mr O'Brien insisted: "Ministers have rejected the suggested pro-

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 possi-

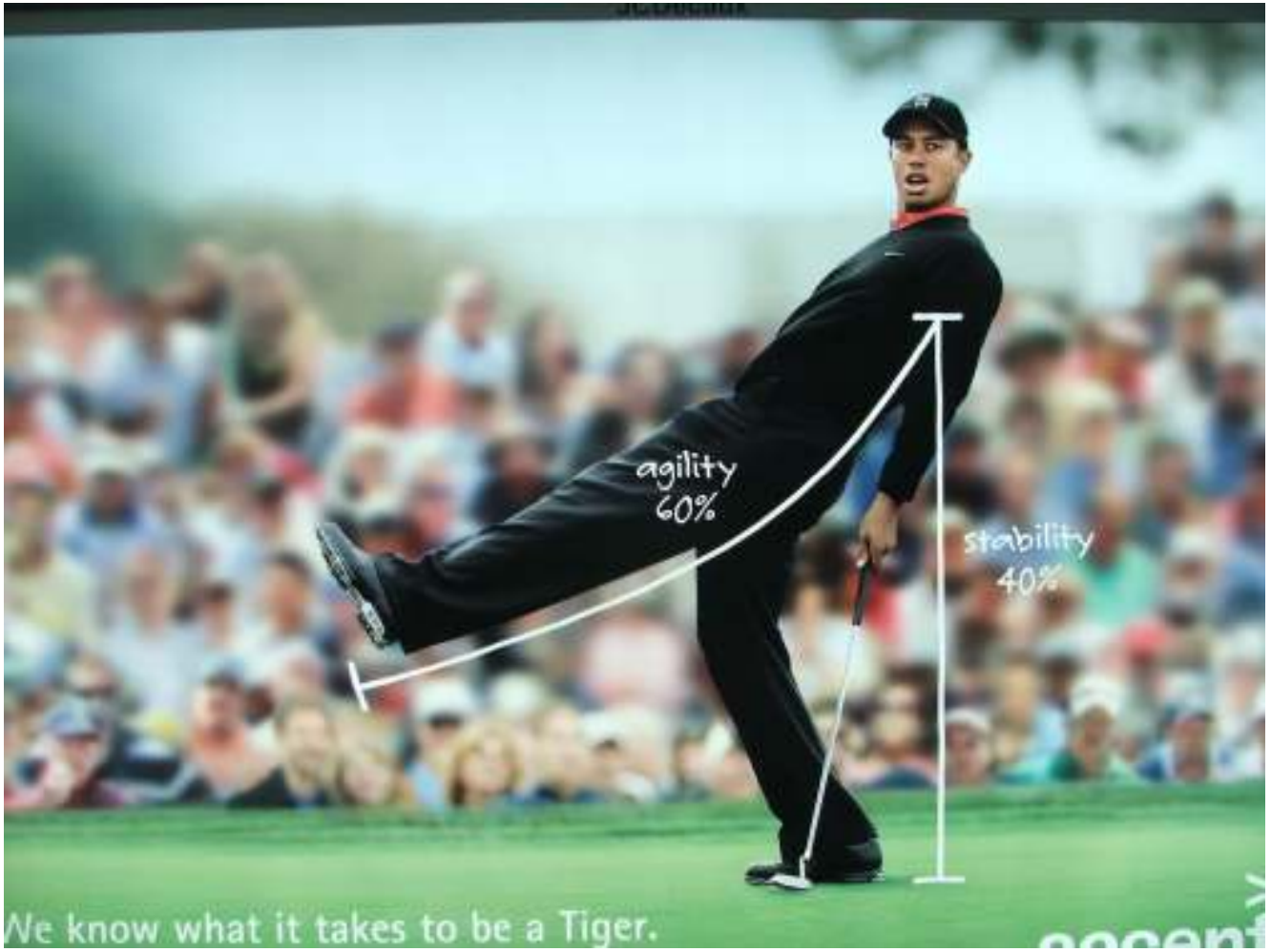
ing the vast majority to go would be front-line NHS staff," he said.

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, [illegible] the issue of 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) PA

Dunne said: "The referee said he ... win the game, we could have scored a ... bad day for football". Brady said: ... many are massive countries. There's


agility
60%
stability
40%
We know what it takes to be a Tiger.