

Certificate Course in Hand & Upper Limb Surgery

Congenital Upper Limb Differences

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NT West cluster

Toshihiko Ogino 荻野利彦

- ✿ Born 4th November 1946
- ✿ Left us on 22nd May 2015
- ✿ A Giant has fallen.....




1946 - 2015

CULD

✿ I was asked to do this:

"..... You are invited to talk about some Congenital Upper limb anomalies such as Upper limb embryology, classifications, syndactyly, Macroductyly, brachyductyly, symbrachyductyly, polyductyly, constriction band syndrome, club hand, cleft hand, hypoplasia limb, syndromatology, arthrogryposis, phocomelia, clinodactyly, camptodactyly, clasped thumb...."

CULD



*Congenital
Malformations
of the*

HAND AND FOREARM

Edited by
DIETER BUCK-GRAMCKO

Congenital Differences of the Upper Limb



Editor:
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The Pediatric Upper Limb

Edited by
Steven Hovius

in collaboration with
Guy Foucher
Piero L. Raimondi



Federation of European Societies for Surgery of the Hand

MARTIN DUNITZ

CULD

- ✿ First encounter with patient and parents
- ✿ From Paediatrics to adulthood
- ✿ Psychological issues
- ✿ Conservative treatment
- ✿ Classifications

First encounter

- ✿ Family having a baby born with anomalies could be in great stress
- ✿ I tend not to see them in the neonatal period
- ✿ In our practice the first encounter is usually between 0 to 3 months old
- ✿ At least one of the parents should be there
- ✿ Attend by appropriate specialist
- ✿ Establish a good rapport is of prime importance

Be prepared to handle these

- ✿ Guilt feeling - **be sensitive and handle with care**
- ✿ Why did it happen?
- ✿ Chance of recurrence in next offspring
- ✿ Chance of transmitting to next generation
- ✿ Is it an isolated anomalies?
- ✿ How would it look like and function after surgery?
- ✿ What can be done about it?
 - **Don't rush to offer surgery**

Worst Case Scenario

- ✿ Complex CULD with expected poor outcome
- ✿ Other organ system anomalies
- ✿ Parents have low self esteem
- ✿ Strong feelings of inadequacy
- ✿ Blaming culture
 - Among family members
 - Pre-natal diagnosis failed to identify
- ✿ They want you to "make" the baby normal

First encounter

- ✿ Some patient might come in bringing information from the internet
- ✿ Layman literacy level in understanding the medical information on the web
- ✿ Study from Edinburgh

First encounter

☀ Take X-rays when indicated

- ▶ When you need the X-ray to help you make decision on management
- ▶ When patient is prepared for surgery
- ▶ For FU assessment

☀ USG, CT and MRI

- ▶ ?? Indication
- ▶ Associated risks of these investigations

☀ Clinical pictures

- ▶ Privacy issue

First encounter

- ✿ Family tree
- ✿ How the other affected family members were treated and what are the outcome
- ✿ What is their expected outcome for the current patient

Opposable Triphalangeal Thumb



L



R



Opposable Triphalangeal Thumb



Opposable Triphalangeal Thumb



First encounter

- ✿ The line between coincidental association or as a result of some syndrome related cause is not always obvious
- ✿ Consult liberally
 - Geneticist
 - Paediatrician
 - Psychologist
 - Social worker

First encounter

- ✿ Explain the most essential things and let them comprehend and understand
- ✿ Come back again with other questions
- ✿ Never ask for confirmation of operation in the first consultation unless urgent
- ✿ Be prepared to refer the patient out for 3^o or 4^o referral

Older patient

- ✿ Must find out why they come so late
 - New immigrants
 - Initial surgery not too satisfactory or just very limited correction
 - Change in family dynamics
 - Someone in the family forbid medical consultation
 - The child becomes an adult and wants to have his own choice

Older patient

- ✿ Respect the patient's own wish
 - "6 years old"
 - Cultural difference
- ✿ Brain development and limited remaining growth might not be helping the surgery
 - Cortical "neglect"
 - Bone and joints and soft tissue less supple
 - Secondary adaptive changes
- ✿ Adolescent patient is most difficult
 - Defensiveness and their inner feelings of conflict

Age at presentation 14



- Born in HK
- Bilateral thumb deformity
- Goes to normal school
- Normal social life
- Plans to emigrate to the States for further studies
- Wants to have it "corrected" first

■ X-ray of another patient







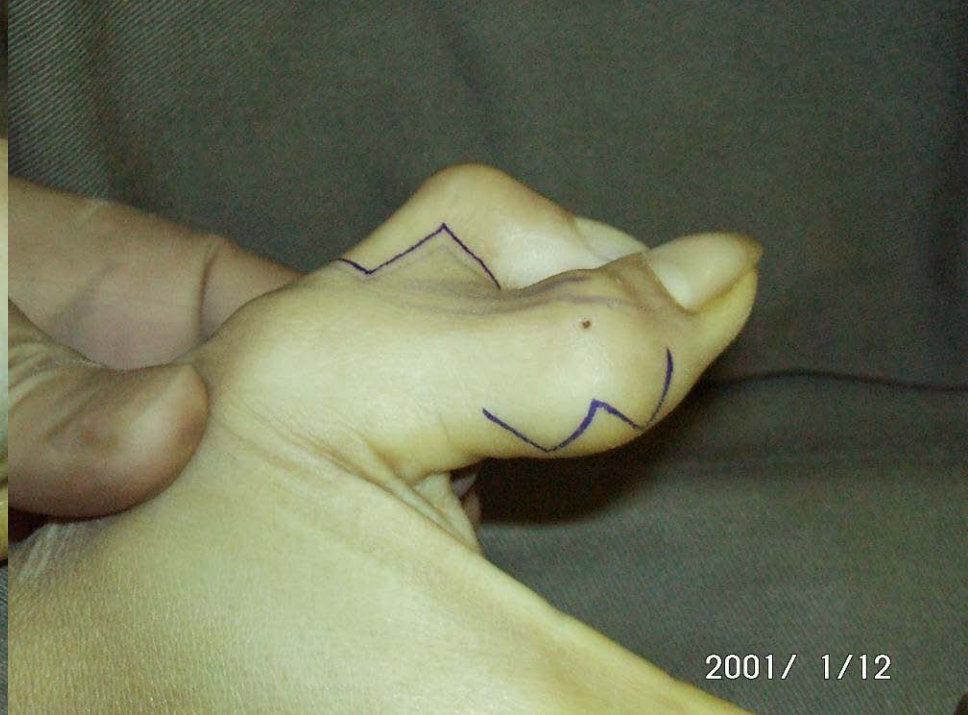
Age at presentation 37



- Thai Chinese
- Mother of two children
- Claims that it was because of the better knowledge of what surgeons can do nowadays
- Essentially no functional limitation
- No social issues



2001/ 1/12



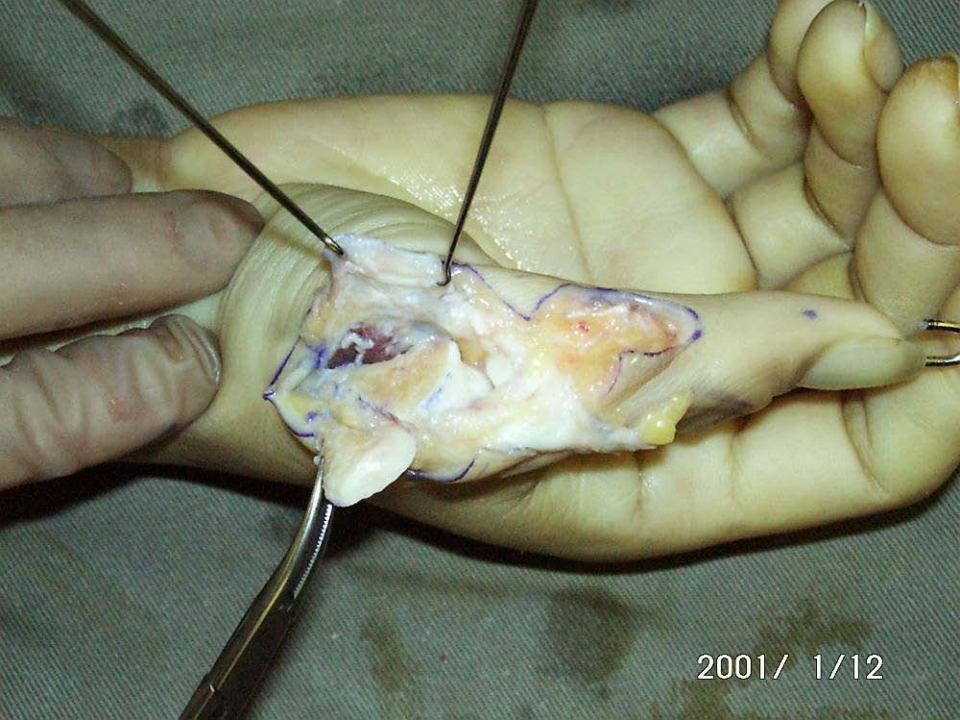
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2001/ 1/12

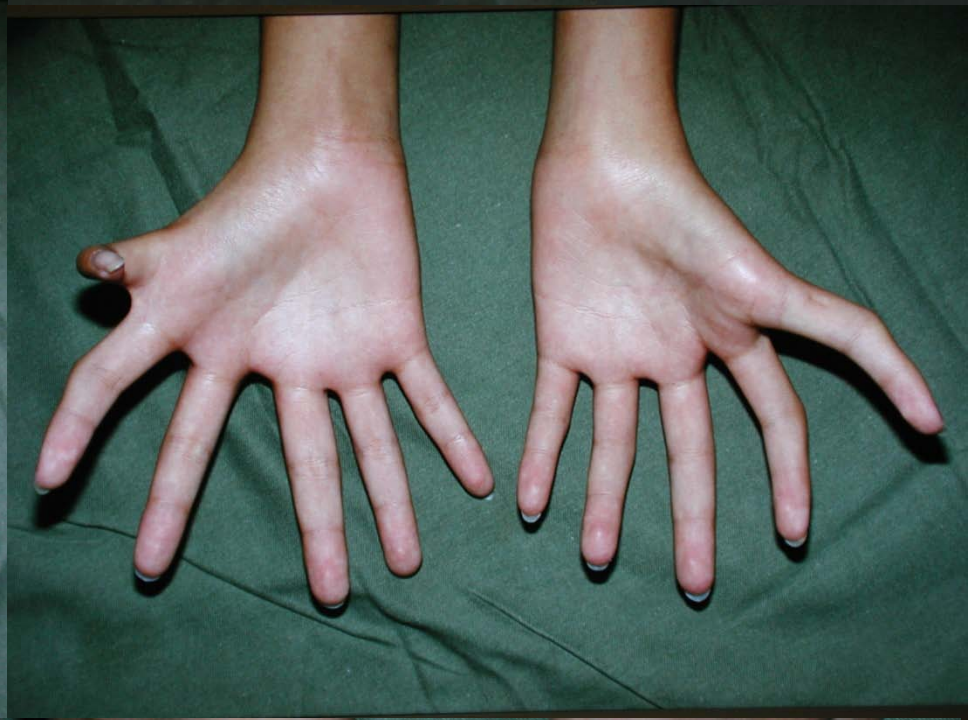


2001/ 1/12



2001/ 1/12





Specific learning points

- Not all late presenters have social or psychological issues
- Soft tissues are less pliable
 - Be prepared to use more skin flaps or grafts
- Skin more susceptible to ischaemia
 - Do not thin it out as you would in the babies
- Need more liberal osteotomies
 - Deformities do need corrections
 - That is usually what they ask for
- Somehow they do not like FU

Patient groups

- ✿ This is not a standard arrangement in my practice
- ✿ Number of patients not that many
- ✿ Most parents don't want to share with others particularly when the deformity has been corrected
- ✿ Some major centres are proud to have such groups

“I just wanted to be normal...”

- ✿ Andre Heuvelman
- ✿ Radial club hand
- ✿ First trumpet soloist in the Rotterdam Philharmonic Orchestra
- ✿ <https://www.youtube.com/watch?v=yERDKKbWx70>
- ✿ TEDx Talks

Psychological issues

✿ For the parents

- "...parents reported that they were more likely to be influenced by the way in which the surgeon communicated with them than the technical issues discussed....."

Psychological issues

- ✿ Very few studies look at the psychological and social benefits of that surgery for the child
- ✿ "...because the operation is technically successful, the child will feel happier about the hand....."
 - This is **WRONG** !!
- ✿ Psychology of children is complex

Conservative treatment

- ✿ Non-operative treatment is always an option for **ALL** kinds of CULD
- ✿ Don't underestimate the power of adaption
- ✿ Advances in prosthesis
- ✿ I would hate the doctor if he or she says to the patient "Don't come back again if you do not want surgery"

Good Classification System

- ✱ Descriptive framework for a group of related clinical conditions
- ✱ Facilitate communication among clinicians
- ✱ Simple and easy to use
- ✱ Wide acceptance
- ✱ High Inter- and Intra-observer reliability
- ✱ Mechanisms of etiology
- ✱ Guidance to treatment
- ✱ Prognosticate the condition
- ✱ Allow for adaptation and expansion

Classifications of CULD

☀ Swanson classification

- ▶ Based on a morphological rather than etiological system
- ▶ Proposed in 1960
- ▶ Adopted by IFSSH from 1976 – 2013

☀ Ogino's modifications of Swanson

- ▶ Incorporated some knowledge of embryology
- ▶ Adopted by JSSH

☀ OMT classification

- ▶ Heavily based on embryopathology knowledge
- ▶ Adopted by IFSSH since Feb 2014
- ▶ <http://ifssh.info/download.html>

OMT Classification

- ☀ **O**BERG, Kerby

- Molecular Embryopathologist
- USA

- ☀ **M**ANSKE, Paul

- Hand surgeon
- USA

- ☀ **T**ONKIN, Michael

- Hand surgeon
- Australia

OMT Classification

- ✿ First proposed in 2010
- ✿ Separates malformation from deformations and dysplasia
- ✿ Integrates existing morphological understanding with current knowledge of the etiology, molecular genetics and developmental biology
- ✿ "Best Fit" principle
- ✿ Regularly updated

OMT Classification

I. Malformation

- Abnormal formation of a body part or complex tissue

II. Deformation

- Results from insults that come after normal formation

III. Dysplasia

- Abnormality in the size, shape, and organisation of cells

IV. Syndromes

OMT Classification

I. MALFORMATIONS

A. Abnormal axis formation/differentiation
– entire upper limb

1. Proximal-distal axis
2. Radio-ulnar (anterior-posterior) axis
3. Dorsal-ventral axis
4. Unspecified axis

B. Abnormal axis formation/differentiation
– hand plate

1. Proximal-distal axis
2. Radio-ulnar (anterior-posterior) axis
3. Dorsal-ventral axis
4. Unspecified axis

OMT Classification

II. DEFORMATIONS

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

B. Tumourous conditions

- 1. Vascular
- 2. Neurological
- 3. Connective tissue
- 4. skeletal

III. DYSPLASIAS

- A. Hypertrophy
 - 1. Whole limb
 - 2. Partial limb

IV. SYNDROMES

- A. Specified
- B. Others

(I presented this in 2004 IFSSH)

Pattern of CULA in Hong Kong

Dr. YY Chow

Cluster Chief of Service

New Territories West Cluster

Tuen Mun Hospital

Hong Kong SAR



Lecture Outline

- Introduction
- IFSSH / JSSH classification systems
- Report of our series
- Illustrative cases

Classification of CULA

- CULA Congenital Upper Limb Anomalies
- CULD Difference
- Classification is difficult
- IFSSH system (Swanson)
- JSSH system (Ogino, Japan)
- Non-classifying recording method (Luijsterburg, The Netherlands)

Classification IFSSH

- **IFSSH Classification 1976 (Swanson)**
 - **I** Failure of formation
 - **II** Failure of differentiation
 - **III** Duplication
 - **IV** Overgrowth
 - **V** Undergrowth
 - **VI** Congenital constriction band syndrome
 - **VII** Generalized skeletal abnormalities

Classification

Japanese Society for Surgery of the Hand (Ogino)

- I Failure of formation
- II Failure of differentiation
- III Duplication
- IV Abnormality of induction of digital rays
- V Overgrowth
- VI Undergrowth
- VII Constriction band syndrome
- VIII Generalized skeletal abnormalities
- IX others

Luijsterberg JHS Br 2003

- Non-classifying recording method
- Standardized detail recordings of the deformity from shoulder to finger tip
- Can be transferred to any classification system

Hospital _____ Please fill in white boxes

I. GENERAL

Date of this registration _____
 Patient identification number _____
 Date of birth _____
 Gender Male Female Unknown
 Name of physician _____
 Clinical genetics consulted Yes No Unknown
 If affirmative, please specify location _____
 Adoption or foster child Yes No Unknown

Caucasian father Yes No Unknown
 Caucasian mother Yes No Unknown
 Consanguinity Yes No Unknown
 If affirmative, please specify _____
 Occurrence among relatives Yes No Unknown
 If affirmative, please specify _____
 Birth weight (grams) _____
 Gestational age (weeks) _____
 Remarks about pregnancy Yes No Unknown
 If affirmative, please specify _____

2. ABERRATIONS OF THE UPPER LIMB

L= left R= right	H* U* R* C*	Ray I				Ray II				Ray III				Ray IV				Ray V																
		MC*	P1	P2	P3	N*	MC*	P1	P2	P3	N*	MC*	P1	P2	P3	N*	MC*	P1	P2	P3	N*		MC*	P1	P2	P3	N*							
Absence		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Absence
Malformed ¹		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Malformed ¹
Hypoplasia ²		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Hypoplasia ²
Hyperplasia ³		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Hyperplasia ³
Synostosis		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Synostosis
Syndactyly / fusion																																		Syndactyly / fusion
Duplication / polydactyly		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Duplication / polydactyly
Clinodactyly																																		Clinodactyly
Camptodactyly																																		Camptodactyly
Ring constriction sec		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Ring constriction sec
Tumor		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Tumor
Trigger																																		Trigger
Flexion impairment sec		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Flexion impairment sec
Extension impairment sec		L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	L	L	L	L	R	R	R	R	Extension impairment sec

Abnormal shoulder Left Yes No Unknown Right Yes No Unknown
 Other aberrations of the upper limb, not appropriate above Left Yes No Unknown Right Yes No Unknown
 (Preliminary) diagnosis of the left arm Yes No Unknown
 (Preliminary) diagnosis of the right arm Yes No Unknown

3. OTHER ABERRATIONS

Circulatory system Yes No Unknown _____
 Respiratory system Yes No Unknown _____
 Digestive system Yes No Unknown _____
 Urogenital system Yes No Unknown _____
 Central nervous system Yes No Unknown _____
 Vertebral column Yes No Unknown _____
 Body wall Yes No Unknown _____
 Head and neck are Yes No Unknown _____
 Skin Yes No Unknown _____
 Lower limbs Yes No Unknown _____
 (Preliminary) common diagnosis Yes No Unknown _____

1 = present, wrong shape; appropriate configuration, undersized = 2, oversized = 3
 * H = humerus; U = ulna; R = radius; C = carpal bones; MC = metacarpal; N = nail

Please send form to Mw Dr Chr. Vermeij-Keers; Research Unit of the Department of Plastic and Reconstructive Surgery; Room EE-1591; Erasmus MC - University Medical Center Rotterdam; P.O. Box 1738; 3000 DR Rotterdam; The Netherlands

ISBN: 90-76580-07-3

Group (I) Failure of Formation

IFSSH

A Transverse arrest

- Anatomical levels

B Longitudinal arrest

- Radial
- Ulnar
- Central
 - cleft hand
- Interegmental

JSSH

A Transverse deficiency

- **symbrachydactyly**
- 10 anatomical subtypes

B Longitudinal deficiency

- Phocomelia
- Radial ray
 - Five fingered hand
 - Blauth classification of hypoplastic thumb
- Ulnar ray
 - Elbow deformity

C Tendon muscle dysplasia

D Nail dysplasia

JSSH Classification

- Group I

- Transverse deficiencies - Symbrachydactyly

- Tendon, muscle dysplasia

- Radial dysplasia, five fingered hand, thumb hypoplasia



Group (II) Failure of Differentiation

IFSSH

A Soft tissue involvement

- Disseminated
 - arthrogryposis
- Anatomical grouping
 - Cutaneous syndactyly
 - Trigger digits

B Skeletal involvement

- Anatomical grouping
 - Osseous syndactyly
 - Clinodactyly
 - Triphalangeal thumb
 - Hypersegmentation

C Tumorous condition

JSSH

A Synostosis

B Radial head dislocation

C Symphalangism

D Contracture

- Soft tissue
 - Arthrogryposis
- Skeletal
 - Kirner deformity
 - Delta bone
 - Madelung deformity

E Tumorous condition

JSSH Classification

- **Group II**
 - Radial head dislocation
 - Madelung deformity
 - Group VIII if it is associated with other syndromes, Dyschondrosteosis
 - Trigger digit
 - Not considered a congenital anomalies



Group (III) Duplication

IFSSH

Anatomical

- Whole limb
- Humeral segment
- Radial segment
- Ulnar segment
- Digits
 - Central
- Epiphyseal



JSSH

- A Thumb polydactyly
 - Wassel
 - Floating type
- B Central polydactyly
 - → Group IV
- C Little finger polydactyly
- D Opposable triphalanbeal thumb
- E Other types of hyperphalangism
- F Mirror Hand

Group (IV) Abnormality of Induction of digital rays

~~IFSSH~~

JSSH



A Soft tissue

- Cutaneous syndactyly
- Cleft of the palm

B Skeletal

- Osseous syndactyly
- Central polydactyly
- Cleft hand
- Cleft hand complex



IFSSH Group IV

Overgrowth

- Whole limb
 - Hemi-hypertrophy
- Partial limb
- Digit
 - macrodactyly

JSSH Group V

Overgrowth

- Macrodactyly
- Hemi-hypertrophy



IFSSH Group V

Undergrowth

- Whole limb
- Forearm and Hand
- Hand alone
 - Entire or partial
- Metacarpal
- Digit

JSSH Group VI

Undergrowth

- Microcheiria
 - Hypoplastic hand
- Brachydactyly
- **Clinodactyly**



IFSSH Group VI

Constriction band syndrome



JSSH Group VII

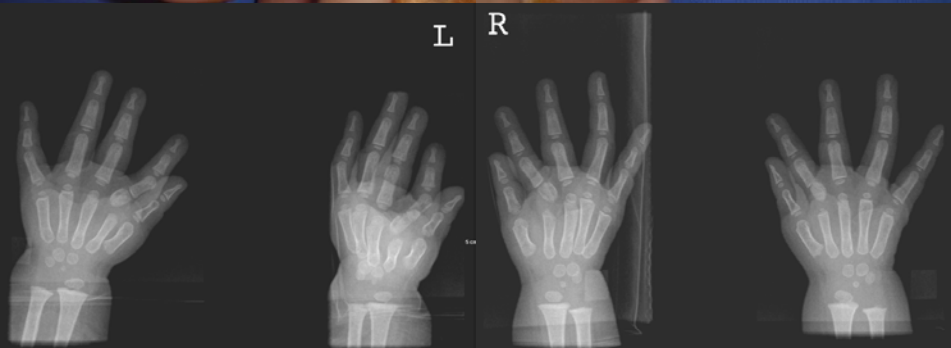
Constriction band syndrome

- Constriction ring
- Lymphedema
- Acrosyndactyly
- Amputation type



IFSSH Group VII

Generalized skeletal abnormalities



JSSH Group VIII

Generalized skeletal abnormalities & a part of a syndrome

- Dyschondrosteosis
- Pierre Robin Sequence

JSSH Group IX

Others

- Including unclassifiable cases

Our Series based on JSSH System

(Continuation of Cheng & Hung series)

Group I	186 (18%)	Group VI	36 (3.5%)
Group II	249 (24%)	Group VII	23 (2.2%)
Group III	418 (40%)	Group VIII	25 (2.4%)
Group IV	63 (6.1%)	Group IX	20 (1.9%)
Group V	16 (1.6%)	<i>Total:</i>	1036

Comparison with Reported Literature

	Flatt (1994)	Lamb (1982)	Ogino (1986)	Giele (2001)	Our series (2004)
Failure of Formation	15	18	11	15	18
Failure of differentiation	41	41	52	32	30
Duplication	15	20	19	38	40
Overgrowth	1	1	1	1	2
Undergrowth	9	14	9	8	4
Constriction ring	2	4	5	3	2
Generalized	4	2	3	3	2
Unclassified	13	--	1	--	2

Take home message

- ✿ Difficult and vast topic
 - You have to read a lot
- ✿ Take the first encounter seriously
- ✿ Good communication is the key
- ✿ "Total patient care" approach to the whole family
- ✿ Be prepared to face a dis-satisfied parent despite you have done a good job

References

- ✿ Dieter Buck-Gramcko, *Congenital Malformations of the Hand and Forearm*, 1998
- ✿ Toshihiko Ogino, *Congenital Differences of the Upper Limb*, 2000
- ✿ Steven Hovius, Guy Foucher et al, *Paediatric hand*
- ✿ Tonkin M, et al, *Classification of Congenital Anomalies of the Hand and Upper Limb: Development and Assessment of a New System*, 2013 ASSH Current Concepts
- ✿ Ezaki M, IFSSH Scientific Committee on Congenital Conditions, 2014
- ✿ Naruse T, Takahara M, Takagi M, Oberg KC, Ogino T. 2007. Busulfan-Induced Central Polydactyly, Syndactyly and Cleft Hand/Foot: A Common Mechanism of Disruption Leads to Divergent Phenotypes. *Dev Growth Diff* 49:533-41.



Thank you