KLASIFIKACE VROZENÝCH DEFEKTŮ HORNÍ KONČETINY

CLASSIFICATION OF CONGENITAL DIFFERENCES OF THE UPPER EXTREMITY

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ABSTRACT

Swanson et al tried to classify congenital hand anomalies according to the genetic cause and reported his classification in 1976. Since then, modifications on this classification were made and this classification was adopted by the International Federation of Society for Surgery of the Hand (IFSSH). It has been used widely as an IFSSH classification. It is relatively easy to use this classification, but it has its own limitations. The biggest one occurs in the classification of ectrodactyly. In order to solve these problems, the authors conducted clinical ad experimental studies and found that there should be at least four different types of teratogenic mechanisms of congenital defect of the digits. The first one is longitudinal deficiencies due to mesenchymal cell death in an early developmental stage; the second is abnormal induction of digital ray numbers in the hand plate including cleft hand, central polydactyly and syndactyly. The third is constriction band syndrome, which is caused after digital radiations have been formed, and the fourth is transverse deficiency, in which the critical period is not known. Based on these studies, the author modified IFSSH classification and it was adopted by the Japanese Society for Surgery of the Hand and is called Japanese modification of the IFSSH classification (Japanese modification). In this paper, the author introduced the Japanese modification of the IFSSH classification and described some recommendations.

Key words: longitudinal deficiency, radial deficiency, ulnar deficiency, cleft hand, constriction band syndrome, transverse deficiency, symbrachydactyly

INTRODUCTION

When babies with congenital hand deformities are seen, the observed form is usually used as the diagnostic name. The use of various Greek and Latin names to describe common deficiencies has only described an anomaly in another language. Using this method, diagnosis is easy. For example, syndactyly is for webbed or fused digits, polydactyly is for extra digits, macrodactyly is for large digits, and ectrodactyly for defective digits. Talking ectrodactyly as an example, there are various types of ectrodactyly: those with constriction rings, those similar to amputation, those with dysplasia of the upper limb, and those with the radial side or ulnar side defect of the forearm and/or hand. Those deformities of digits are labeled as ectrodactyly, although the causes are divergent.

Swanson et al (**37**) in 1968 reported a classification of congenital hand deformities based on the concept of embryological failure. However, actually only some of the part of this classification was based on the morphology. After they reported the original classification, modification on this classification was made by Swanson (**38**) in 1976, and this classification was adopted by the International Federation of Societies for Surgery of the Hand in 1983(39). In this classification, there are seven categories as follows: Category 1, failure of formation of parts (developmental arrest), Category 2, failure of differentiation of parts, Category 3, duplication, Category 4, overgrowth, Category 5, undergrowth, Category 6, congenital constriction band syndrome, and Category 7, generalized skeletal abnormalities. This classification has been used widely and it is relatively easy to use it, but it has its own limitations (**15**).

In recent years, according to the analysis of clinical cases, cleft hand is a congenital deformity closely related to central polydactyly and syndactyly, and it is different from other absence of digits including longitudinal deficiency (**11, 21, 32**). The cleft hand is a congenital deformity, similar to central polydactyly and syndactyly, which are caused during the induction of the digital rays in the developmental period of digital radiation (**21, 32**). Based on this viewpoint, when one examines the radiographs of the clinical cases, in the case of osseous syndactyly between the middle and ring fingers and the polydactyly of the middle finger, if the development of osseous syndactyly occurs in the proximal direction, then it will develop toward the cleft hand (**32**). In the clinical cases, in the left and right hands of the same patient, the complicated combinations of central polydactyly, osseous syndactyly, and cleft hand do occur. Based on these observations, it is possible that cleft hand is the congenital deformity closely related to central polydactyly and syndactyly. The results of the laboratory-induced cleft hand indicate that if osseous syndactyly or polydactyly develops in the ring finger toward the proximal position, cleft hand also will develop. It also is clear that the time (critical period) when the cleft hand appears is consistent with that of the central polydactyly and syndactyly. A single cause affecting the limb bud in a certain receptive period of the development of the apical ectodermal ridge can induce the deformities (**13, 14**). The changes of the polydactyly and syndactyly caused by the abnormal induction of the digital ray numbers in the hand plate may be found in the central deficiency. It is difficult to classify these cases according to the IFSSH

classification. Therefore, when classifying congenital deformities of the hand based on genetics, central polydactyly, osseous syndactyly, and cleft hand may be grouped together and included in the same category of abnormal induction of digital ray numbers in the hand plate (**21**).

On the other hand, in German-speaking areas, transverse deficiency is regarded as a congenital deformity in the same category as congenital amputation, atypical cleft hand and brachysyndactyly, and these deformities are called symbrachydactyly by Müller (**12**), Blauth and Gekeler (**2**). According to the classification of Blauth et al (**2**), Grade 1 consists of short webbed fingers. Typical brachysyndactyly has shortening or absence of the middle phalanges combined with cutaneous syndactyly. Grade 2 is called atypical cleft hand, in which dysplasia of the central digital rays is severe, but complete absence of the fingers rarely occurs. There is no V-shaped depression in the palm of atypical cleft found in the typical cleft hand. Grade 3 is the monodactyly type, in which the thumb remains and there are the vestigial fingers or vestigial nails where the fingers are severely hypoplastic. Grade 4 is the peromelia type, which is also called congenital amputation and all digits are impaired (**32**). In these cases, the hypoplasia of the bones always appears across the upper limbs. Therefore, these cases are called transverse deficiencies, in contrast with longitudinal deficiency that occurs locally on the long axis of the upper limbs. Among different types of transverse deficiency, there may be intermediate types of deformities. In addition, there are some congenital deformities that can be found in all types: one side impairment; severe dysplasia of the central digital rays; bone dysplasia in the impaired digits, neighbored digits, or the entire impaired upper limb; and complication of the impairment of the pectoralis major muscle (**18, 20**). According to these observations, the sequence of deformities from brachysyndactyly, or the atypical cleft hand seems to be a morphological variant of transverse deficiency.

Based on clinical and experimental studies, the classification of congenital hand deformities proposed by IFSSH has been modified by Ogino et al (**16**) in 1986. This classification has been updated and was adopted by the Japanese Society for Surgery of the Hand (**5**) in1996. In IFSSH classification, polydactyly is classified into duplication, syndactyly into failure of formation of parts and typical cleft hand into failure of formation of parts. However, these congenital deformities may appear when the same teratogenic factor acts on embryo at the same developmental period. Therefore, they are included in the fourth new category of abnormal induction of digital rays in this classification. On the other hand, brachysyndactyly is classified into undergrowth and transverse deficiency into failure of formation of parts, and there is no item of atypical cleft hand in IFSSH classification. However, brachysyndactyly, atypical cleft hand or transverse deficiency seems to be developed as morphological variants of symbrachydactyly. Therefore, these deformities are included in the same concept of transverse deficiency as an item of failure of formation of parts. These are the main points of modification of the IFSSH classification. In this paper, the author introduces the Japanese modification of the IFSSH classification and describes some recommendations.

CLASSIFICATION CONGENITAL DIFFERENCES OF THE UPPER EXTREMITY MODIFIED BY THE JAPANESE SOCETY FOR SURGERY OF TE HAND (2000)

I.	**Failure of formation of parts (arrest of development)**		
A.	**Transverse deficiency**		
A.1	Peripheral hypoplasia type	A.8	Metacarpal type
A.2	Short webbed finger type	A.9	Carpal type
A.3	Tetradactyly type	A.10	wrist type
A.4	Tridactyly type	A.11	Forearm type
A.5	Didactyly type	A.12	Elbow type
A.6	Monodactyly type	A.13	Upper arm type
A.7	Adactyly type	A.14	Shoulder type
B.	**Longitudinal deficiencies**		
B.1	Radial deficiencies	B.2	Ulnar ray deficiencies
B.1.1	Dysplasia of the radius	B.2.1	Dysplasia of the ulna
B.1.1.1	Hypoplasia of the radius	B.2.1.1	Hypoplasia of the ulna
B.1.1.2	Partial absence of the radius	B.2.1.2	Partial absence of the ulna
B.1.1.3	Total absence of the radius	B.2.1.3	Total absence of the ulna
B.1.2	Deformities of the hand	B.2.2	Deformities of the hand
B.1.2.1	Five fingered hand	B.2.2.1	Hypoplasia of the little finger
B.1.2.2	Hypoplastic thumb: Grade 1–5	B.2.2.2	Absence of the 5th digital ray
B.1.2.3	Absence of more than two digital rays	B.2.2.3	Absence of more than two digital rays
B.1.3	Dysplasia of the elbow	B.2.3	Dysplasia of the elbow
B.1.3.1	Contracture of the elbow joint	B.2.3.1	Contracture of the elbow joint
B.1.3.2	Humeroradial synostosis	B.2.3.2	Humeroradial synostosis
B.1.3.3	Radial head dislocation	B.2.3.3	Radial head dislocation
C.	**Phocomelia**		
C.1	Complete type		
C.2	Proximal type		
C.3	Distal type		
D.	**Tendon or muscle dysplasia**		
E.	**Absent nail or skin**		

II.	**Failure of differentiation of parts**		
A.	**Synostosis**	**D.**	**Contracture**
A.1	Humeroulnar synostosis	D.1	Soft tissue
A.2	Humeroradial synostosis	D.1.1	Arthrogryposis multiplex
A.3	Radioulnar synostosis	D.1.2	Webbed elbow (Pterygium cubitale)
A.4	Synostosis of the carpal bones	D.1.3	Clasped thumb
A.5	Metacarpal synostosis	D.1.4	Windblown hand
B.	**Radial head dislocation**	D.1.5	Camptodactyly
C.	**Symphalangism**	D.1.5.1	Single digittype
C.1	Proximal type	D.1.5.1	Multiple digit type
C.2	Distal type	D.1.6	Aberrant muscles
C.3	Combined type	D.1.7	Muscle contracture
		D.1.8	Swan neck deformity
		D.1.9	Button hole deformity
		D.1.10	Nail deformity

II.	Failure of differentiation of parts *(continued)*		
D.2	Bone	**E.**	**Tumorous conditions**
D.2.1	Kirner deformity	E.1	Hemangioma
D.2.2	Delta bone (Longitudinal epiphyseal brancket)	E.2	Arteriovenous fistula
		E.3	Lymphangiom
D.2.3	Madelung deformity	E.4	Neurofibromatosisx
D.3	Others	E.5	Juvenile aponeurotic fibroma
		E.6	Osteochondromatosis
		E.7	Other

III.	Duplication		
A.	**Thumb polydactyly**	**B.**	**Central polydactyly: (Central polydactyly should be classified into Category IV.**
A.1	Type 1	**C.**	**Polydactyly of the little finger**
A.2	Type 2	C.1	Well-developed type
A.3	Type 3	C.2	Floating type
A.4	Type 4	C.3	Others
A.5	Type 5	**D.**	**Opposable triphalangeal thumb**
A.6	Type 6 (Triphalangeal type of thumb polydactyly may appear with Types 3, 4, 5, and 6.) Floating type, others (thenar muscle hypoplasia type and radially deviated type)	**E.**	**Other types of hyperphalangism**
		F.	**Mirror hand**

IV.	Abnormal induction of digital rays		
A.	**Soft tissue**	**B.**	**Bone**
A.1	Cutaneous syndactyly	B.1	Osseous syndactyly
A.2	Cleft of the palm	B.2	Central polydactyly
		B.3	Cleft hand (absence of central finger rays)
		B.4	Triphalangeal thumb
		B.5	Cleft hand complex

V.	Overgrowth
A.	**Macrodactyly**
B.	**Hemihypertrophy**

VII.	Constriction band syndrome
A.	**Constriction ring**
B.	**Lymphedema**
C.	**Acrosyndactyly**
D.	**Amputation type**

VI.	Undergrowth
A.	**Microcheiria (Hypoplastic hand)**
B.	**Brachydactyly**
C.	**Clinodactyly**

VIII.	Generalized skeletal abnormalities & a part of syndrome

IX.	Others (including unclassifiable cases)

CATEGORY I. FAILURE OF FORMATION OF PARTS (ARREST OF DEVELOPMENT)

The category of failure of formation of parts is that group of congenital deficiencies noted by failure or arrest of formation of the limb either complete or partial. In this classification, it is sub-classified into five types: transverse deficiency, longitudinal deficiency, phocomelia, absent tendons or muscles, and absent nail or skin.

A. Transverse deficiencies (so-called symbrachydactyly)

This category includes short webbed finger to amputation like deformities. The most characteristic feature in the roentgenograms of transverse deficiency was that various degrees of bone hypoplasia existed in the affected fingers, adjacent fingers and a proximal part of the affected limbs. The common features of all types of transverse deficiency were that all cases were unilateral, and in every grade there were some cases associated with the absence of pectoral muscle (**2, 27**). Finger reduction occurred mainly in the central digital rays, the thumb was least frequently affected and it had a definite pattern that progressed from brachymesophalangy, through the absence of the middle phalanx, that of the proximal and middle phalanges and that of all phalanges, finally to absence of the metacarpal bone.

A.1 Peripheral hypoplasia type

While in short webbed finger type, hypoplasia of the phalanx appears predominantly in the middle phalanges, in this type, it appears predominantly in

Peripheral hypoplasia

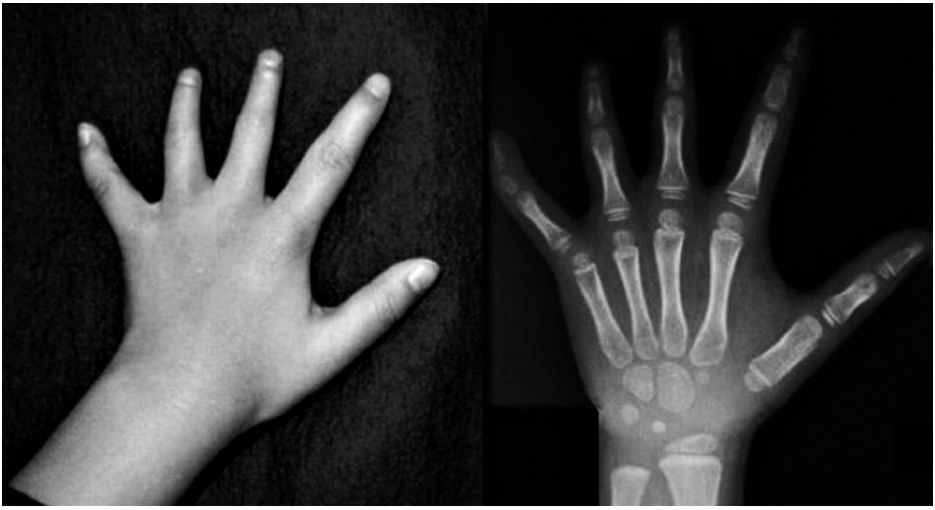

Fig. 1: Transverse deficiency: Peripheral hypoplasia type

Short webbed finger type

Triphalangeal **Diphalangeal** **Monophalangeal**

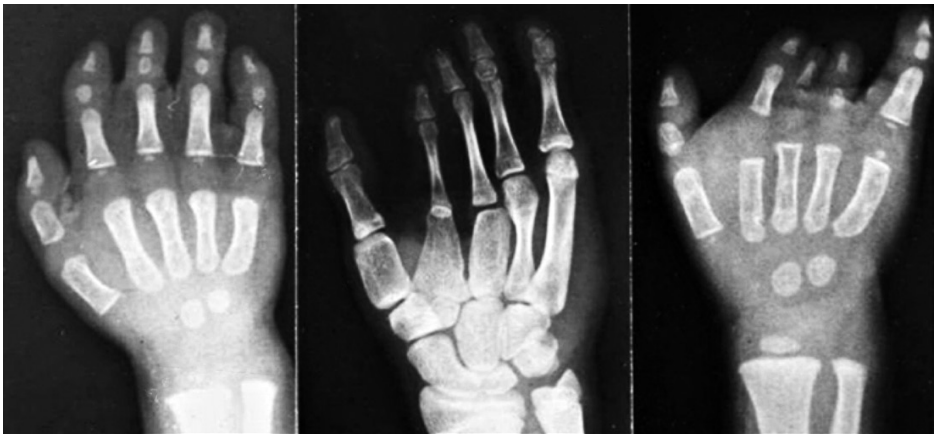

Fig. 2: Transverse deficiency: Short webbed finger type: Left: triphalangeal type, Center: diphalangeal type, Right: monophalangeal type

the distal phalanges. Therefore, hypoplasia of the distal phalanx or aplasia of the distal phalanx and absence of nails are characteristic clinical features in this type. Usually syndactyly is not associated in this type **Fig. 1**.

A.2 Short webbed finger type

Short fingers and syndactyly are essential clinical features of this type. Brachymesophalangy, absence of the middle phalanx and/or absence two phalanges including middle phalanges in a single finger ray is commonly observed. Sugiura (**35**) classified this type into three types, such as triphalangeal type, diphalangeal type, and monophalangeal type **Fig. 2**.

In triphalangeal type, there are brachymesophalangy in some finger rays, in diphalangeal type, there are absence of the middle phalanges at least in one finger ray, and in monophalangel type, there are absence of the middle and proximal phalanges at least one finger ray. However, in all types, one can find hypoplasia of the all digits including the thumb.

A.3 Tetradactyly type

All phalanges are missing in one central digit **Fig. 3**.

A.4 Tridactyly type

All phalanges are missing in two central digits.

A.5 Didactyly type

All phalanges are missing in three central finger rays **Fig. 4**. This type is used to be called atypical cleft hand. Atypical

Tetradactyly type

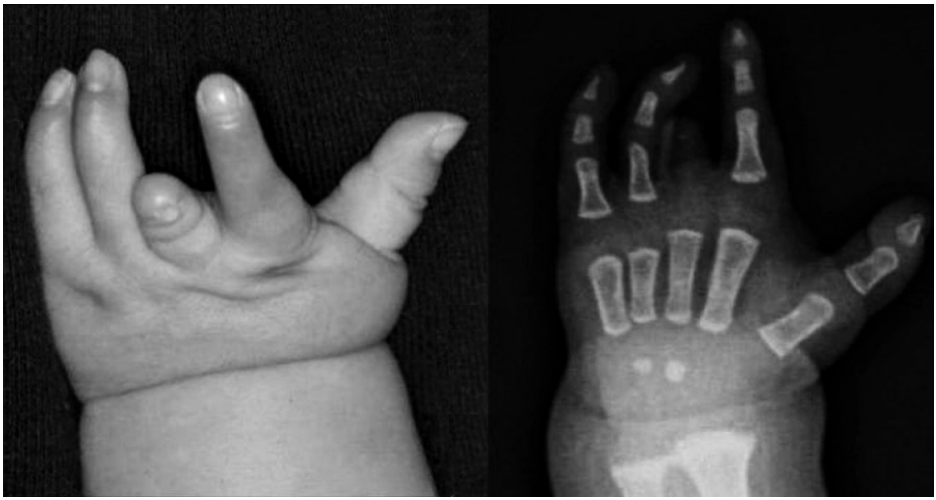

Fig. 3: Transverse deficiency: Tetradactyly type

Didactyly type

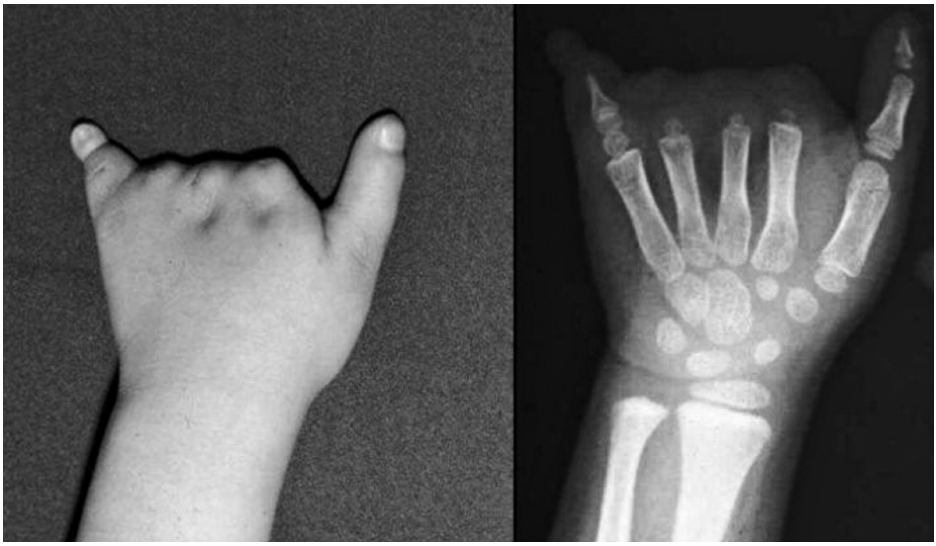

Fig. 4: Transverse deficiency: Didactyly type

Monodactyly type

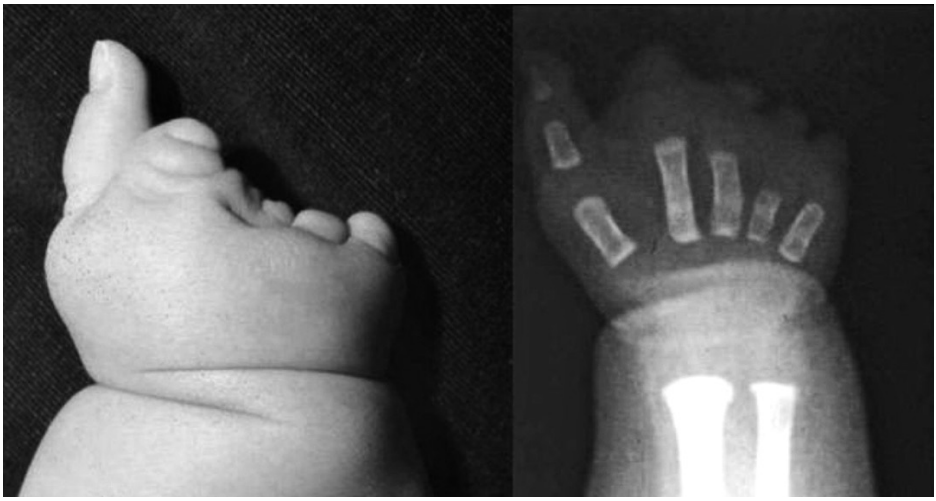

Fig. 5: Transverse deficiency: Monodactyly type

cleft hand is considered to be a severe deformity of symbrachydactyly and typical cleft hand is considered to be caused by abnormal induction of digital rays. The Congenital Committee of the International Federation of Societies for Surgery of the Hand approved the recommendation that use of the term "Atypical Cleft Hand" be discontinued (**9**).

A.6 Monodactyly type

All phalanges are missing in ulnar four digital rays. In tetradactyly type to monodactyly type, thumb is always hypoplastic compared to the opposite side **Fig. 5**. There are one or two phalanges in the thumb.

A.7 Adactyly type

All phalanges are missing in the thumb and all finger rays.

A.8 Metacarpal type

In addition to the missing of all phalanges in all digital rays, some part of the metacarpals are missing.

A.9 Carpal type

All skeletal elements distal to the carpal bones and some part of the carpals are missing.

A.10 Wrist type

All skeletal elements distal to the radius including carpal bones are missing.

A.11 Forearm type

All skeletal elements distal to the forearm bone and some part of the forearm bones are missing **Fig. 6**.

Forearm type **Elbow type**

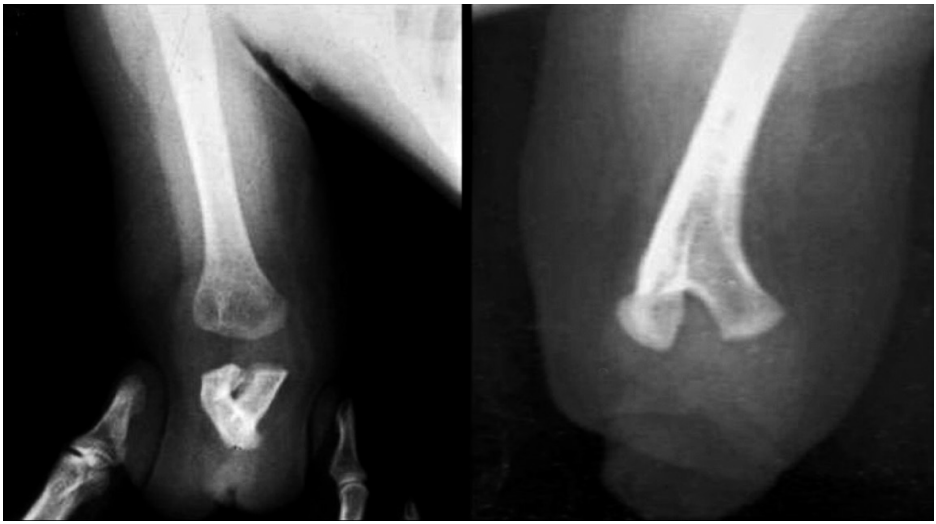

Fig. 6: Transverse deficiency: Forearm type (left). Elbow type (right)

A.12 Elbow type

All skeletal elements distal to the elbow joint forearm are missing **Fig. 6**.

A.13 Upper arm type

All or some part of the humerus is missing.

A.14 Shoulder type

All skeletal elements of the upper extremity and some part of the scalupa are missing.

B. Longitudinal deficiencies

In IFSSH classification, there are two major categories of congenital absence of digits. One is transverse deficiency and the other is longitudinal deficiency. Congenital absence of digits confined to the long axis of the upper limb is called longitudinal deficiency. In IFSSH classification, longitudinal deficiency is classified into three types. The absence of digits on the ulnar side is called ulnar deficiency, which of the radial side is called radial deficiency, and that of the central part is called central deficiency or cleft hand. The longitudinal deficiencies considered to be caused due to mesenchymal cell death in an early developmental stage (**32**). However, many investigators suggested that teratogenic mechanisms of central deficiency were different from those of radial and ulnar deficiencies (**32**). Therefore, the author classified longitudinal deficiency into radial and ulnar deficiencies (**16, 29**). Central deficiency has been excluded from this category. Longitudinal deficiency

Hypoplastic thmb (Blauth)

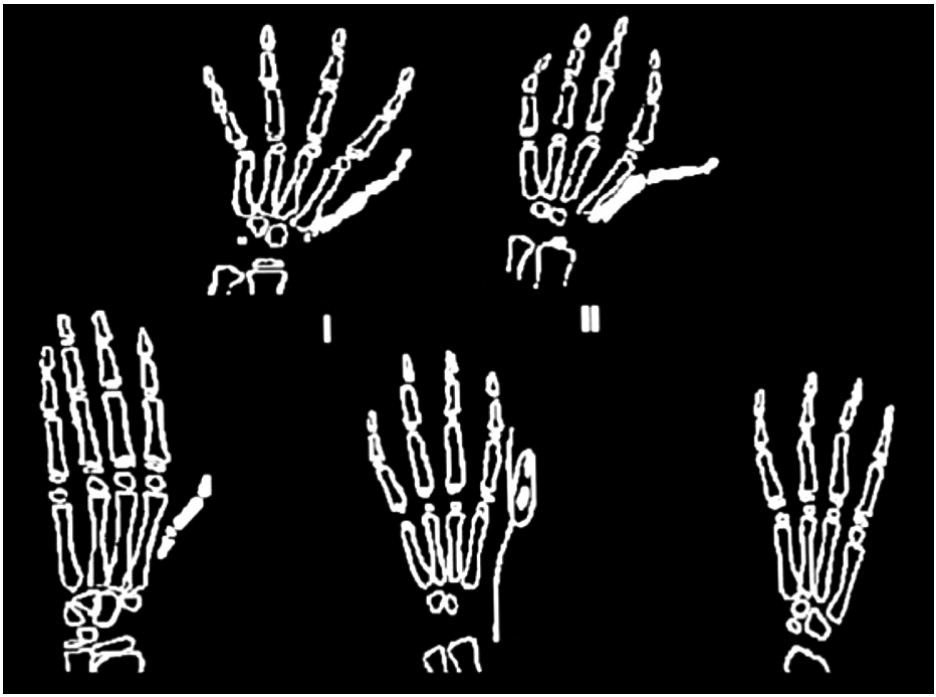

Fig. 7: Blauth's classification for hypoplastic thumb in radial deficiency

means the deformities in which hypoplasia or aplasia of the forearm bone and/or that of the digits are located in radial side or ulnar side of the limb.

B.1 Radial deficiencies

In radial deficiency, the skeletal changes appear in the hand, forearm and elbow in clinical cases. In the forearm, there are 4 types of dysplasia of the radius, such as total absence of the radius, partial absence of the radius, hypoplastic radius and normal radius with hypoplastic thumb (**32**). In some cases of small children with total absence of the radius, X-ray film does not show any radius but hypoplastic anlage of the radius becomes to be ossified when they grow up. In such cases, type of the radius dysplasia changes during growth. Hand deformities in radial deficiency are classified according to Blauth's classification (**1**) **Fig. 7**. In his classification, **Grade 1**: the mildest form and hypoplasia of the thenar muscles without functional disturbance, **Grade 2**: hypoplasia of the thenar muscles associated with adduction contracture of the thumb, **Grade 3**: hypoplasia of the thenar muscles with absence of the first metacarpal base, **Grade 4**: floating thumb, **Grade 5**: the most severe form, total absence of the thumb. Non-opposable triphalangeal

thumb, which is called five-fingered hand, is also one of the types of hypoplastic thumb **Fig. 8**. In some cases, radial two digits such as, the thumb and index finger are absent **Fig. 8**. There are also special type of hypoplastic thumb, in which hypoplastic/absent thumbs and cutaneous syndactyly of the most radial digits (**34**). In syndactylous type of hypoplastic thumb, complete fusion of the radius and the ulna appears in some cases. Pollex abductus is also sometimes associated with hypoplastic thumb. Pollex abductus is an anomaly in which the flexor pollicis longus attaches not only at its customary insertion, but also into the extensor by a tendon that passes around radial aspect of the thumb (**40**). Contraction of the anomalous flexor abducts the thumb.

Radial deficiency is sometimes associated with elbow deformities **Fig. 9**. Limitation of the elbow flexion, ankylosis of the elbow, radial head dislocation and radio-ulnar synostosis may be associated with radial deficiency (**32**). In radial deficiency, deformity of the hand, that of the radius and that of the elbow appear in various combinations. Therefore, deformities in radial deficiency of a patient should be expressed with combination among dysplasia of the radius, hand and elbow deformities. Most of the deformities belong to radial deficiencies could be expressed with combinations of these deformities. This classification system successfully reflects the multiplicity of the defects of radial deficiencies.

B.2 Ulnar deficiencies

The absence of the finger of the ulnar side is called ulnar deficiency. In severe ulnar deficiency, absence of the digits is sometimes associated with dysplasia of the ulna and/or elbow deformity. Dysplasia of the ulna is classified into 3 types such as, total absence, partial absence, and hypoplasia of the ulna. In some cases,

Radial deficiency: Hand

Five fingered hand Absence of more than 2 digits

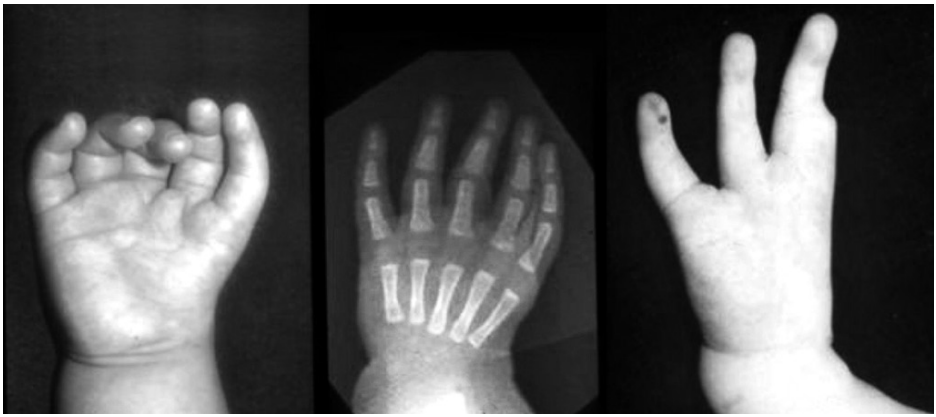

Fig. 8: Hypoplastic thumbs in radial deficiency: Left, Center: Five fingered hand, Right: absence of the thumb and index finger

patients have normal ulna with hypoplastic/absent ulnar fingers. Hand deformities of ulnar deficiencies are classified into 5 types, such as the hypoplasia of the little finger, absence of the little finger, absence of the 2 digital rays of the ulnar side, absence of the 3 digital rays of the ulnar side, and absence of the four digital rays of the ulnar side (**32**). Deformities of the radial digits sometimes associated with absence of the ulnar digits. Thumb is always present although it is sometimes hypoplastic. And the deformities of the elbow are classified into humeroradial synostosis, radial head dislocation, and flexion contracture of the elbow joint. In ulnar deficiencies, deformities of the hand, dysplasia of the ulna and deformities of the elbow occur in various combinations. Therefore, deformities must be expressed with combination among hand deformity, dysplasia of the ulna and deformities of the elbow as in radial deficiencies. This classification system also successfully reflects the multiplicity of the defects of ulnar deficiencies.

B.3 Proximal type of longitudinal deficiency

Goldfarb et al (**6**) reported deficiencies of the proximal part of the arm in longitudinal deficiencies. Radial deficiency of the proximal part of the arm includes: a. Hypoplasia and aplasia of the humeral head; and b. Hypoplasia of the glenoid.

Ulnar deficiency of the proximal part of the arm includes; a. Severe radiohumeral synostosis with distal humeral duplication, and b. severe radiohumeral synostosis with a large medial exostosis that resembles large medial condyle. The congenital hand committee of the IFSSH recommended that these types also should include into longitudinal deficiency. However, these are very rare conditions and I wonder whether we should include these items as a part of classification or

Radial deficiency: Elbow

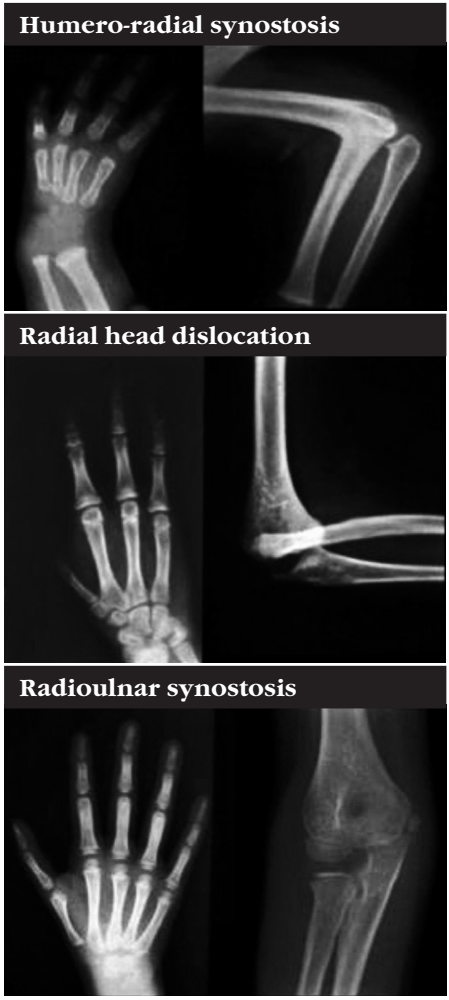

Fig. 9: Combination of elbow deformities and hypoplastic thumb in radial deficiency

Congenital ankylosis of the elbow between humerus and:

radius **ulna** **radius & ulna**

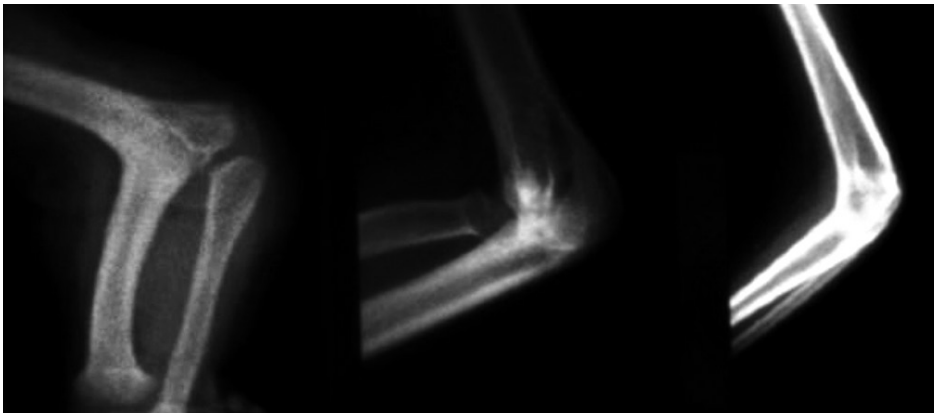

Fig. 10: Congenital ankylosis of the elbow joint: Left: Humeroradial synostosis, Center: Humeroulnar synostosis, Right: Humeroradial and humeroulnar synostoses

not. The Japanese modification does not include these conditions in this category.

C. Phocomelia

Phocomelia is classified into three types as follows: a. Complete type, b. Proximal type, c. Distal type. The major manifestation of complete type phocomelia is absence of the humerus and forearm bones and the hand attached directly to the shoulder girdle.

Proximal type of phocomelis is absence of the humerus, and distal type of phocomelia is absence of the forearm bones.

D. Absent tendons or muscles

Absent of flexor pollicis longus, finger that of extensors and that of pectoralis muscles are included in this item.

E. Absent nail or skin.

Congenital hypoplasia or aplasia of the fingernail is classifies into this item.

CATEGORY II. FAILURE OF DIFFERENTIATION OF PARTS

Failure of differentiation or separation of parts is this category, in which the basic unit of the hand and arm have developed, but the final form is not completed. In IFSSH classification, syndactyly is included in this category, but it appears as a result of abnormal induction of the digital ray numbers of the hand plate (**13, 14, 21**). Therefore, syndactyly is excluded from this category. This category includes incomplete formation of the joint, contracture and deformities due to failure

Congenital radioulnar synostosis

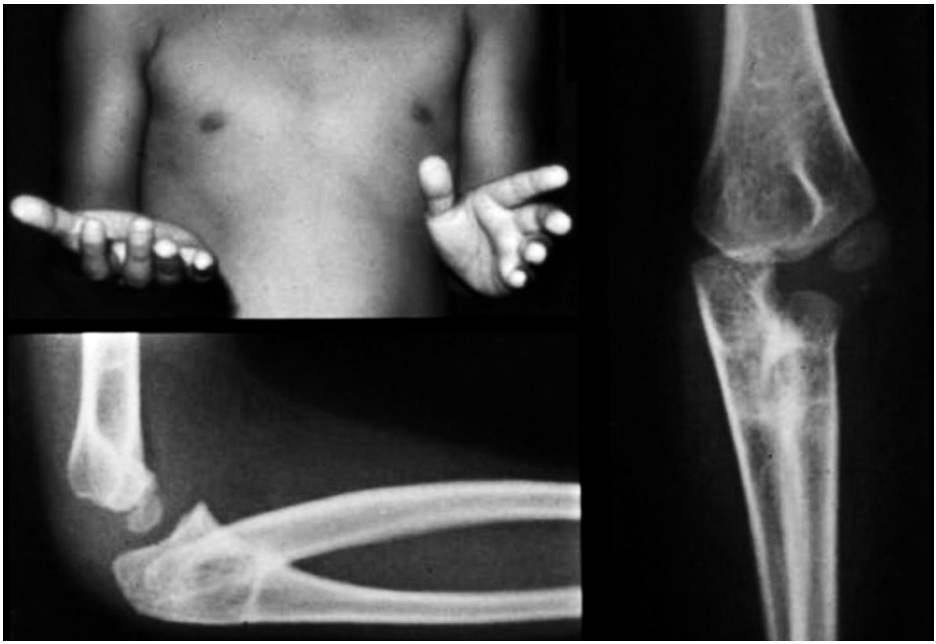

Fig. 11: Congenital radioulnar synostosis: The left side was affected and the forearm was ankylosis in pronation position.

of differentiation of parts, and tumor-like conditions including hamartomas. Incomplete formation of the joint may result in synostosis, congenital dislocation, and ankylosis of digital joint.

A. Synostosis

Synostosis includes humero-ulnar synostosis, humero-radial synostosis, radioulnar synostosis, carpal coalition, and metacarpal synostosis. The humero-ulnar synostoses, and humero-radial synostosis, which cause ankylosis of the elbow joint, often appear as a part of ulnar deficiency or that of congenital syndromes, such as Antley-Bixler syndrome and Apert syndrome (**28**) **Fig. 10**. Radio-ulnar synostosis occurs mostly in the proximal radioulnar joint but it also occurs in the distal radioulnar joint (**17**) **Fig. 11**. In some cases, the radius and the ulna fused side by side completely, but it is rare condition (**34**). Carpal coalition may occur among all carpal bones. The metacarpal synostosis is often occurs in the ring and little finger rays **Fig. 12**. It may occur as an isolated anomaly and also as a part of ulnar deficiency (23). In this deformity, bone fusion occurs only at the base of the metacarpals in some cases, but complete fusion of the adjacent metacarpals may also occur.

Metacarpal synostosis

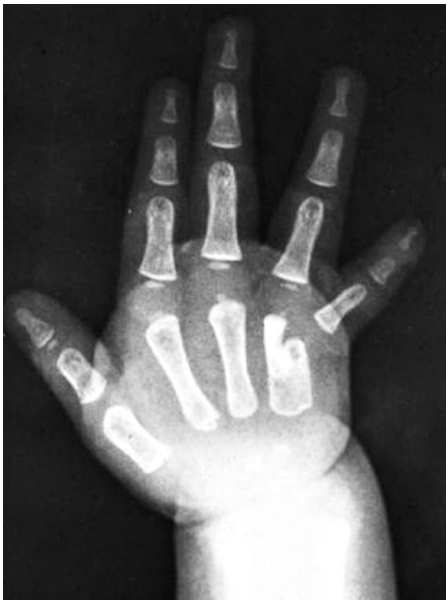

Fig. 12: Congenital metacarpal synostosis of the ring and little fingers: incomplete fusion: The proximal parts of the ring and little finger metacarpals were fused osseously.

B. Radial head dislocation

In congenital dislocation, radial head dislocation is the most common, but the diagnosis is not easy when it appears as an isolated dislocation that is not associated with other anomalies. In such cases, it is difficult to make differentiation from traumatic one (**10**).

C. Ankylosis of digital joints

In ankylosis of digital joint, there are symphalangism and congenital ankylosis of the metacarpophalangeal (MP) joint. Symphalangism is a congenital ankylosis of the finger joint. It includes ankylosis of the proximal interphalangeal (PIP) joint, which is a proximal type, ankylosis of the distal interphalangeal (DIP) joint, which is a distal type, and combination of proximal and distal types (**25**) **Fig. 13**. In congenital ankylosis of the MP joint, the ankylosis is a partial one and the patient can extend the MP joint of the finger but the patient cannot flex the MP joint completely.

D. Contracture and deformities

Contracture and deformities are caused due to soft tissue abnormalities and skeletal abnormalities. The former one includes arthrogryposis multiplex congenita, pterygium cubitale, clasped thumb, windblown hand, and camptodactyly, aberrant muscles, muscle contracture, swan-neck deformity, buttonhole deformity, and nail deformity. The later one includes Kirner's deformity, delta bone, and Medelung's deformity. Clinodactyly may be included in this category.

D.1 Soft tissue

D1.1 Arthrogryposis multiplex congenital

Bilateral upper limbs are involved. The shoulders are adducted, the elbows are extended and the wrists are flexed. The fingers are usually extended and the thumb is extended and adducted position. Muscle weakness of the both arms is evident.

D.1.2 Pterygium cubitale

This is very rare condition and there is a cutaneous web on the anterior elbow

Symphalangism

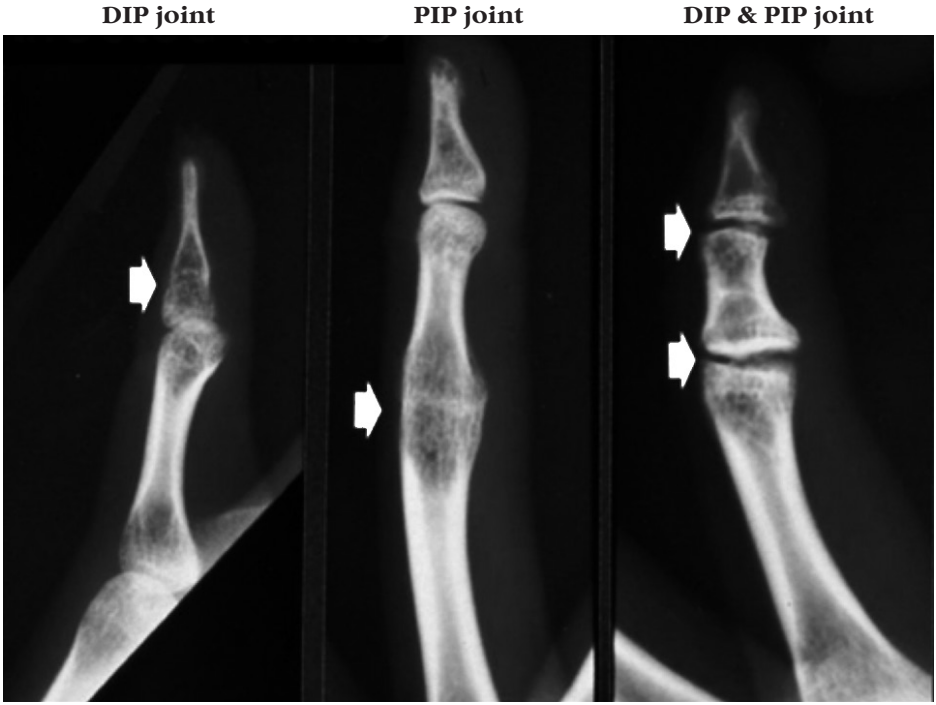

Fig. 13: Symphalangism: Left: distal symphalangism, Center: Proximal symphalangism, Right: Incomplete symphalangism of the distal and proximal interphalangeal joints

when it is extended, but he elbow extension is severely restricted. Hypoplasia of the elbow is often associated and the radial head is usually dislocated **Fig. 14**.

D.1.3 Clasped thumb

Thumb in palm position is seen in infant, but the thumb can be extended passively. Gradually patient can extend the IP joint of the thumb, but not MP joint of the thumb. If it is followed without treatment, flexion contracture of the MP joint occurs and the IP joint becomes hyper-extended **Fig. 15**.

D.1.4 Windblown hand

Both hands are involved. The MP and PIP joint of the fingers are flexed and there is ulnar deviation of the MP joints. Thumb in palm position is characteristic in this deformity **Fig. 16**. If windblown hands are associated with clubfeet or vertical talus, the case might be distal arthrogryposis (**7**). If they are associated with micrognathia, the case might be Freeman-Sheldon syndrome. Hand deformity in congenital absence of the extensor muscles of the digits shows similar to the windblown hand deformity.

Webbed elbow: Pterigium cubitale

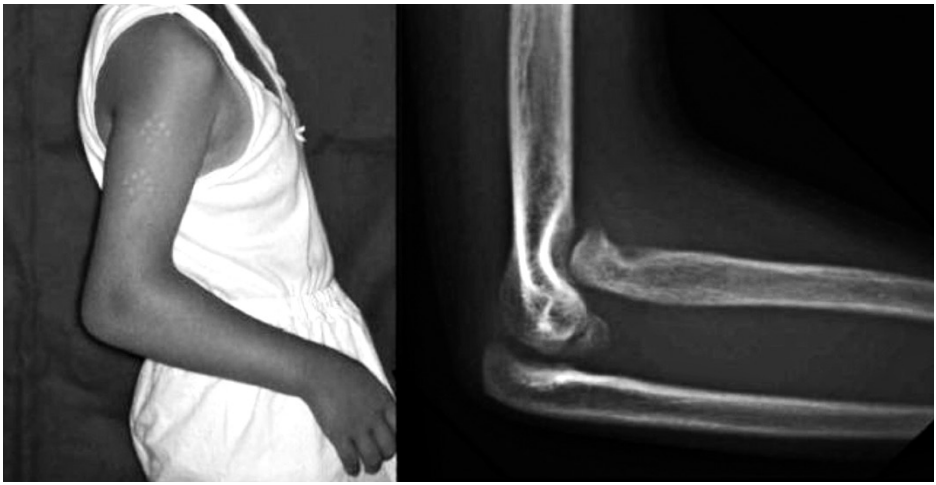

Fig. 14: Pterigium cubitale: The right elbow had a cutaneous and soft tissue webbing of the anterior elbow associated with dislocation of the radial head and hypoplasia of the elbow joint

Clasped thumb

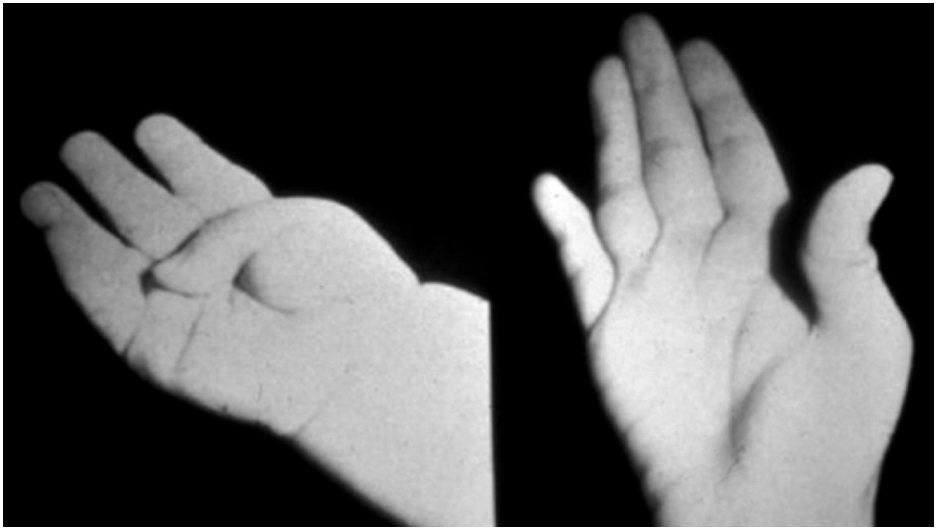

Fig. 15: Clasped thumb: Left: There was thumb in palm deformity, but a flexion deformity of the metacarpophalangeal joint was correctable manually in this infant. Right: The patient could flex the interphalangeal joint, but the flexion deformity of the metacarpophalangeal joint of the thumb was fixed at the school age

Windblown hand

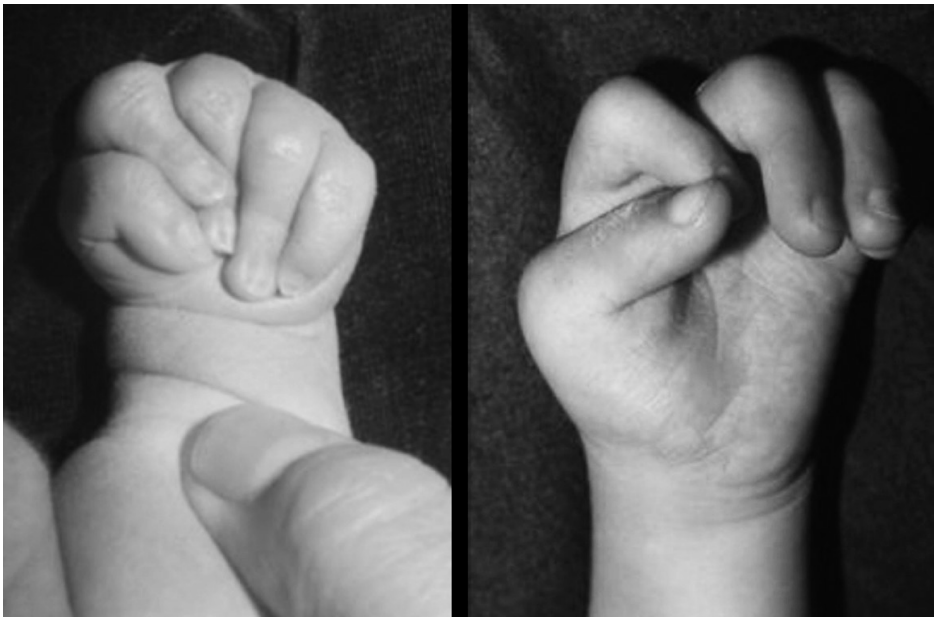

Fig. 16: Windblown hand: Left: The both hands had similar deformity. All digits were flexed and there was overlapping of the fingers soon after birth. The flexion deformity of the metacarpophalangeal joint of the thumb was improved slightly.

D.1.5 Camptodactyly

There are two types of camptodactyly **Fig. 17**. In single digit type, only single digit is involved and usually the little finger is affected. The deformity is noticed in infant in some cases but it is also first noticed in teen-agers in some cases (**22**). Multiple digits type means multiple digit involvement (**26, 42**). If multiple digit type is associated with crumpled ear, the case might be Beal's syndrome, which is congenital contractual arachnodactyly (**24**). Camptodactyly is often associated with cleft hand or thumb hypoplasia. This is not a true camptodactyly. The deformity must be classified according to the underlying conditions.

D.1.6 Aberrant muscles

It is considered that the characteristic clinical features of aberrant muscles are unilateral muscular hyperplasia, aberrant muscles or accessory muscles, ulnar drift of the fingers in the MP joints, flexion contractures of the MP joints, extension contractures of the wrist, and bulky palmar surface (**31**) **Fig. 18**. However, bilateral involvement of this condition has been reported. Roentogenogram reveals widening of the intermetacarpal spaces. Crossing of the fingers and/or intrinsic plus deformity are sometimes observed.

Camptodactyly

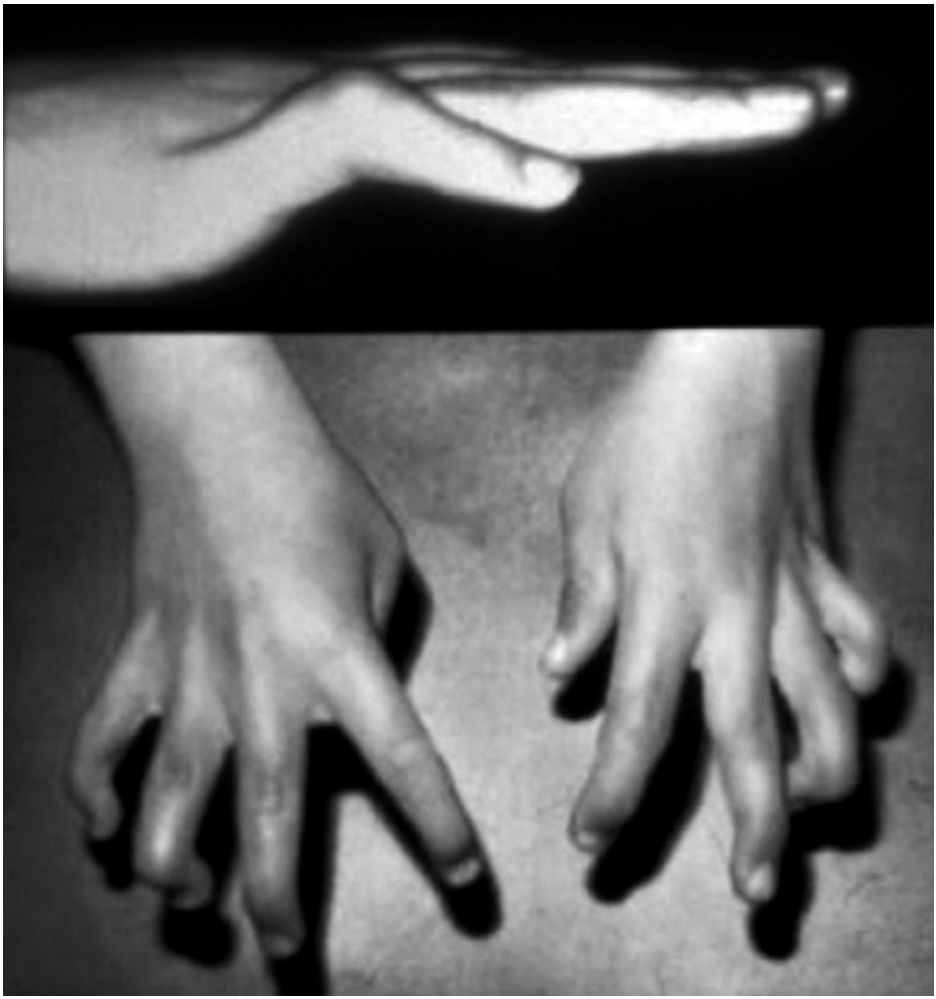

Fig. 17: Camptodactyly: Above: camptodactyly of the little finger; There was a flexion contracture of the proximal interphalangeal joint. Below: Camptodactyly of the multiple digits. There was a limitation of the extension of the digits.

D.1.7 Muscle contracture

In a case of muscle contracture, the range of motion of the joint, where the affected muscle is a mover, is limited. Usually the affected muscle has short muscle belly and long tendinous portion. The excursion of the muscle is smaller than normal.

Abberant muscle syndrome

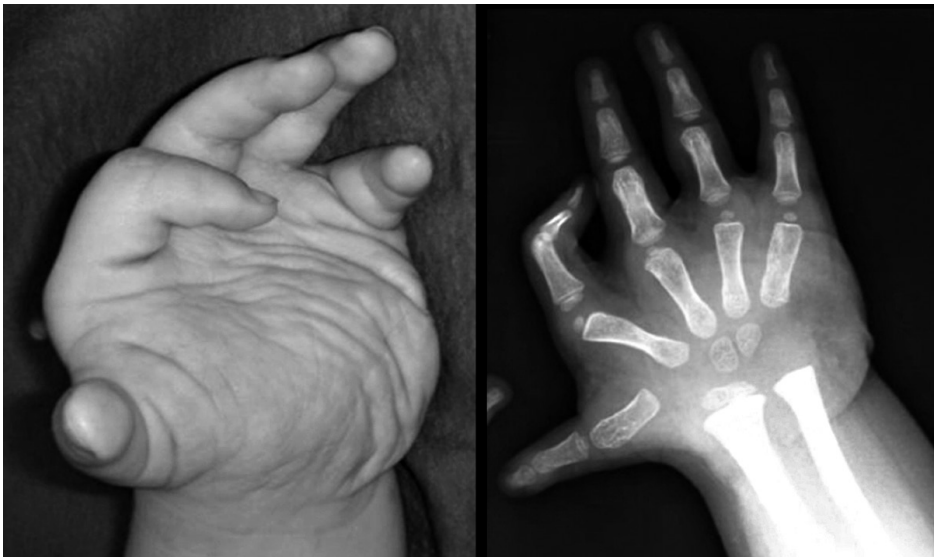

Fig. 18: Abberant muscle syndrome: Left: There were enlargement of the hand, ulnar drift of the fingers in the metacarpophalangeal joints, and bulky palmar surface. Right: Roentogenogram reveals widening of the intermetacarpal spaces. The thumb was in extended and abducted position.

D.1.8 Swan-neck deformity

The congenital swan-neck deformity gives rise to the locking phenomena after a few years of progressive laxity of the volar plate and the collateral ligaments and becomes worse with aging.

D.1.9 Buttonhole deformity

Multiple fingers of both hands are affected and the deformity becomes contracture as other deformities.

D.1.10 Nail deformity

Congenital nail deformity includes claw nail, hook nail , clam nail and circumferential nail.

D.2 Bone

D.2.1 Kirner deformity

Kirner deformity is a condition, in which distal phalanx of the little finger has palmar radial curvature. The nail has a similar deformity to the distal phalanx **Fig. 19**. Bilateral fingers are involved more than half cases. The deformity rarely appeared in other finger.

D.2.2 Delta bone

The Delta phalanx is defined that the phalanx in which the epiphyseal growth plate runs longitunally, although normally, it runs perpendicular to the longitudinal axis of the phalanx **Fig. 20**. The terminol-

Kirner deformity

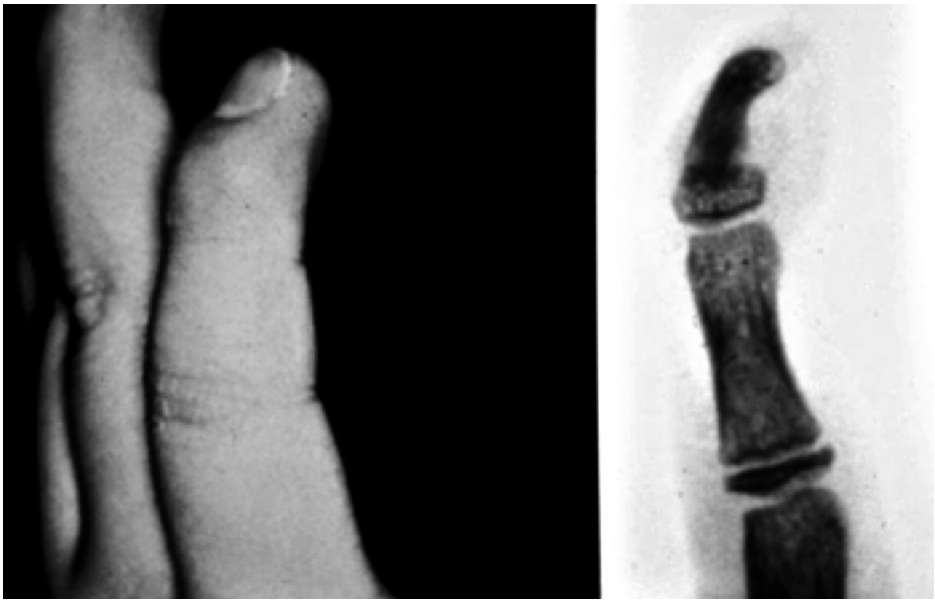

Fig. 19: Kiner deformity of the little finger: The distal phalanx of the little finger had palmar radial curvature.

ogy of delta phalanx is used in the IFSSH classification for congenital hand differences. Some investigators think that the Delta phalanx is not the appropriate terminology and recommend using the longitudinal bracketed diaphysis or the longitudinal epiphyseal bracket (**4, 8**). The same kind of deformity appears in metacarpal or metatarsal bones. Therefore, the terminology of the Delta bone has been used in the Japanese modification of the IFSSH classification. Delta bone is often associated with other deformities, such as polydactyly and syndactyly. These cases must be classified according to the underlying conditions.

D.2.3 Madelung deformity

The majority of cases of Madulung deformity are caused by hereditary dyschondrosteosis at the wrist. These cases with mesomeric dwarfism must be classified into Category 9 .

E. Tumorous conditions

They includes, haemangioma, arteriovenous fistula, lymphangioma, neurofibromatosis, and juvenile aponeurotic fibroma.

Delta bone associated with synpolydactyly

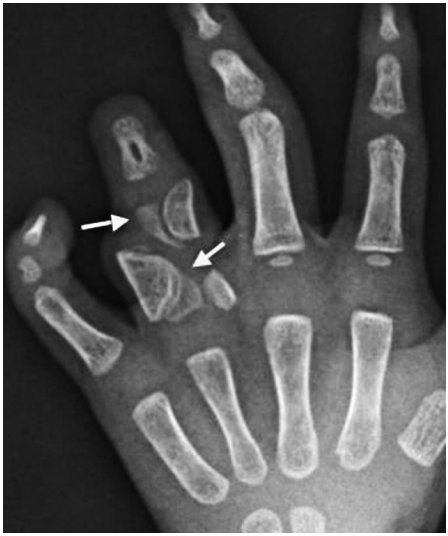

Fig. 20: Delta phalanges (Delta bones) associated with central synpolydactyly: In the ring finger, there were two Delta phalanges (small arrows), in which the epiphyseal growth plate run longitunally.

CATEGORY III. DUPLICATION

In IFSSH classification, radial, central, and ulnar polydactyly were included in this category, but central polydactyly appears as a result of abnormal induction of the digital ray numbers in the hand plate and mostly it is associated with syndactyly. Therefore, central polydactyly is excluded from this category.

A. Thumb polydactyly

Thumb polydactyly can be subclassified according to Wassel's classification (**41**) **Fig. 21** as follows; a) Type1, b) Type 2, c) Type 3, d) Type 4, e) Type 5, f) Type 6. Triphalangeal type of thumb polydactyly may appear with Types 3, 4, 5, and 6. In such cases, one should describe thumb polydactyly Type 4 radial triphalangeal type. There are other types, such as floating type, thenar muscle hypoplasia type and radially deviated type (**19**).

Wassel's classification

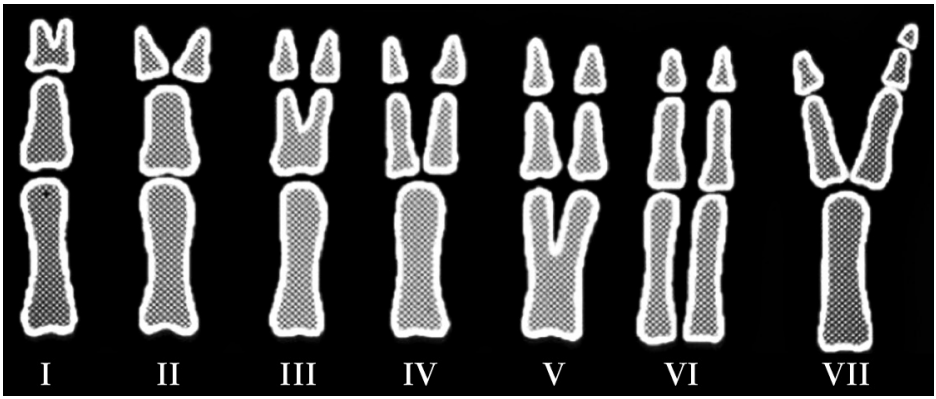

Fig. 21: Wassel's classification for radial polydactyly

Ulnar polydactyly

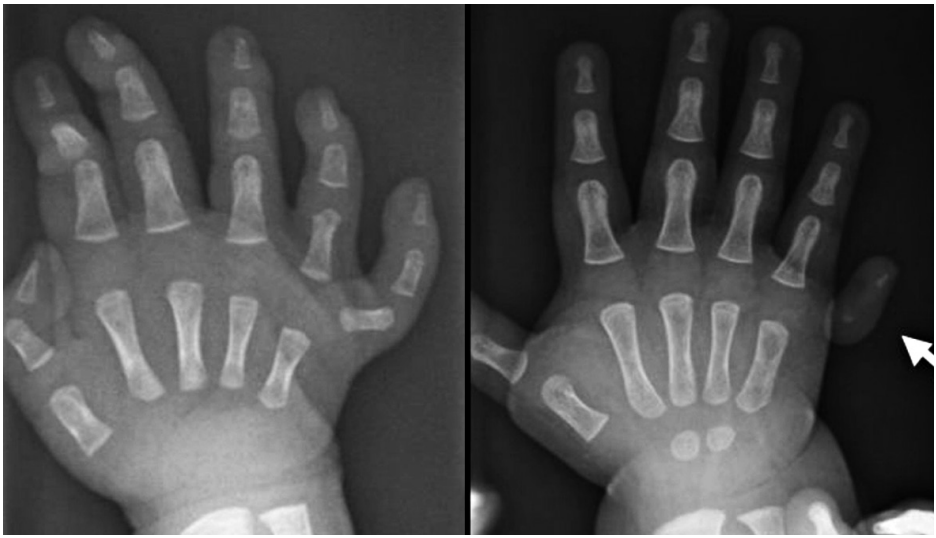

Fig. 22: Ulnar polydactyly: Left: Well formed type, Right: Floating type

B. Central polydactyly: It should be classified into category IV.

C. Polydactyly of the little finger

It is classified into floating type and well developed type, but there are cases with specific deformity, such as polydactyly of the little finger associated with ankylosis of the MP joint **Fig. 22**.

D. Opposable triphalangeal thumb

There is no item of triphalangeal thumb in IFSSH classification. There are two types of triphalangeal thumb. One is opposable triphalangeal thumb and the other is non-opposable one. Non-opposable thumb is classified into hand deformities of radial deficiency, since thenar muscle is always absent. Opposable triphalangeal thumb might be formed as a result of incomplete fusion of the duplicated thumb, since X-ray film of the true antero-posterior view of the thumb but not that of the hand often reveled evidence of thumb polydactyly (**27**) **Fig. 23**. Therefore it is classified in this category. Opposable triphalangeal thumb is often associated with cleft hand with absence of the index finger. This type of triphalangeal thumb should be classified according to the underlying diseases. In opposable triphalangeal thumb without other deformity, the deviation of the thumb is always ulnar ward, but in that associated with cleft hand it is always radial ward and the index finger is missing.

E. Other types of hyperphalangism

Opposable triphalangeal thumb

Type 1 Type 2 Type 3

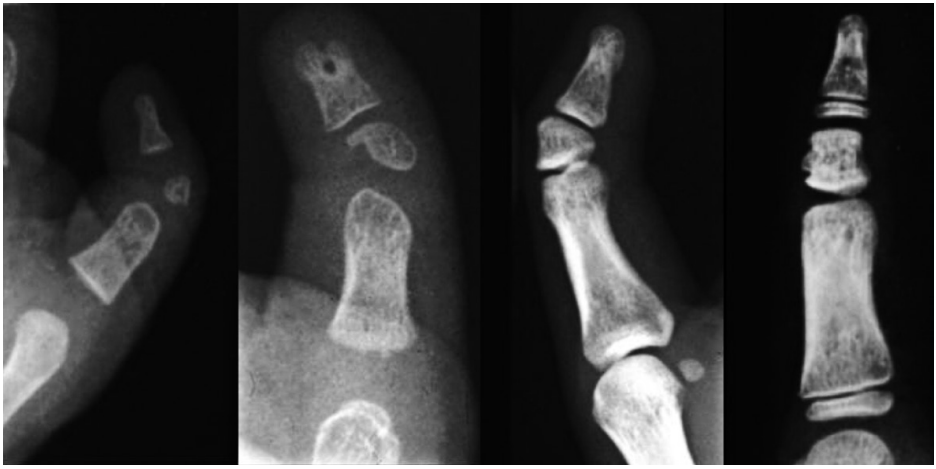

Fig. 23: Opposable triphalangeal thumb: Type 1: There was a small ossicle between the distal and proximal phalanges in small child. This type may change into Type 2 or 3 during growth. Type 2: The deformity of the epiphysis is so-called triangular epiphysis. This type is considered as a transient form between normal and independently formed middle phalanx. Type 3: There was a triangular or rectangular middle phalanx. This type is a real triphalangeal thumb.

Cutaneous syndactylyand cleft on the palm

Fig. 24: Cutaneous syndactyly and cleft on the palm: These are soft tissue deformities due to abnormal induction of digital rays.

Osseous syndactyly

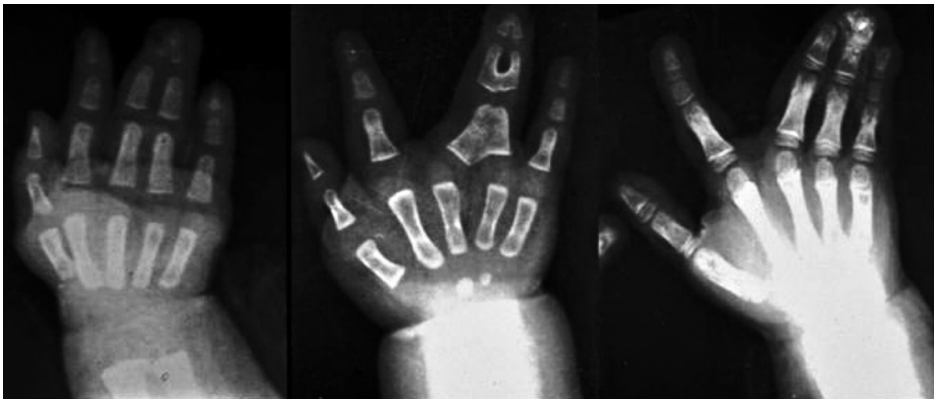

Fig. 25: Osseous syndactyly due to abnormal induction of digital: Left: Distal phalangeal type, Center: Proximal phalangeal type, Right: The ulnar three digits were involved.

F. Mirror hand

In mirror hand, thumb is always missing, but an excess number of fingers are present. In typical mirror hand, there are two ulnas and the radius is missing. In this type of deformity, the motion of the elbow is limited and there is special deformity of the distal end of the humerus. If there is no duplication of the ulna, it is classified into atypical mirror hand. This type has no elbow deformity.

CATEGORY IV. ABNORMAL INDUCTION OF DIGITAL RAYS (ABNORMAL INDUCTION OF DIGITAL RAY NUMBERS IN THE HAND PLATE)

Manifestations of malsegmentation of the digital rays in the hand plate due to abnormal induction of digital rays are included in this category (**29, 32**).

As a skin manifestation, there are cutaneous syndactyly and cleft of the palm **Fig. 24**. Cleft of the palm means deep V shaped excessive interdigital space. It appears as an isolated anomaly without absence of the central finger ray, but mostly it is associated with absence of central finger rays. As a skeletal manifestation, there are osseous syndactyly, central polydactyly, absence of the central fingers and triphalangeal thumb **Fig. 25**. Cleft hand means absence of the central fingers. It should be distinguished from cleft of the palm. In cleft hand, usually central single finger, two fingers or three fingers may be missing **Fig. 26**. But in some cases, ulnar four digits or radial four digits are missing. When the index finger seems to be missing, triphalangeal thumb is associated in all cases in our series **Fig. 27**. When only the single digit is preserved, it is necessary to distinguish it from monodactyly type of transverse deficiency and ulnar deficiency

Absent central digit & cleft on the palm

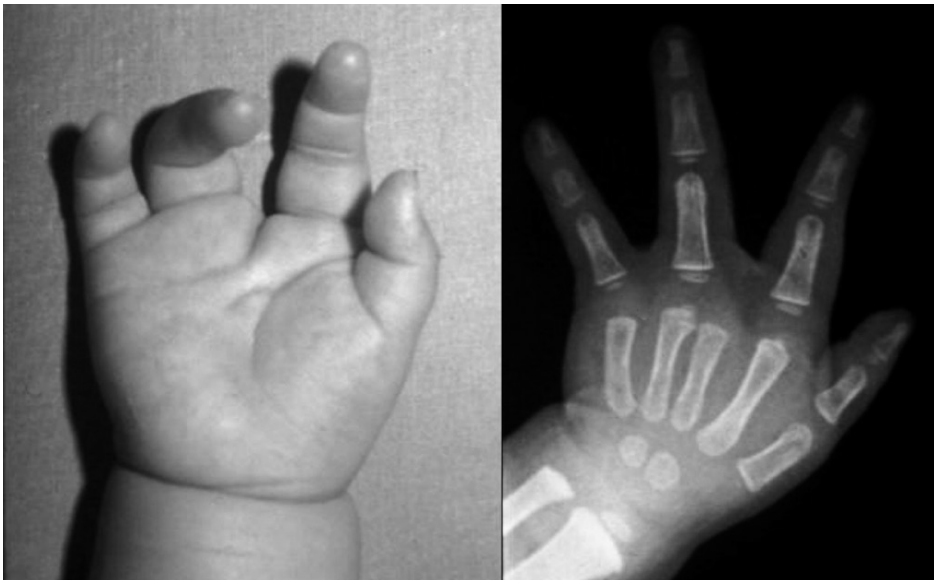

Fig. 26: So called cleft hand due to abnormal induction of digital: The middle finger was missing and there was a cleft on the palm between the middle and ring fingers.

Cleft hand with triphalangeal thumb

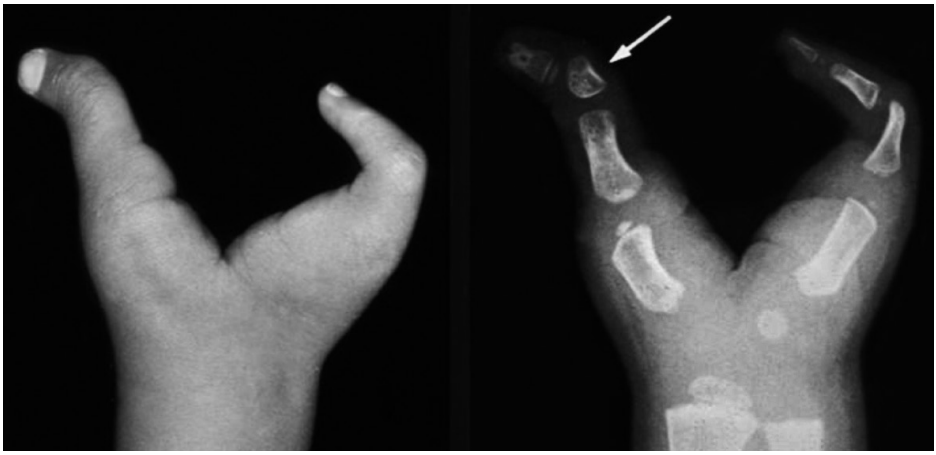

Fig. 27: Cleft hand with triphalangeal thumb due to abnormal induction of digital: There was a triphalangeal thumb and the direction of the deviation was radial, though in opposable triphalangeal thumb it was ulnar.

Central polydactyly & cutaneoud syndactyly

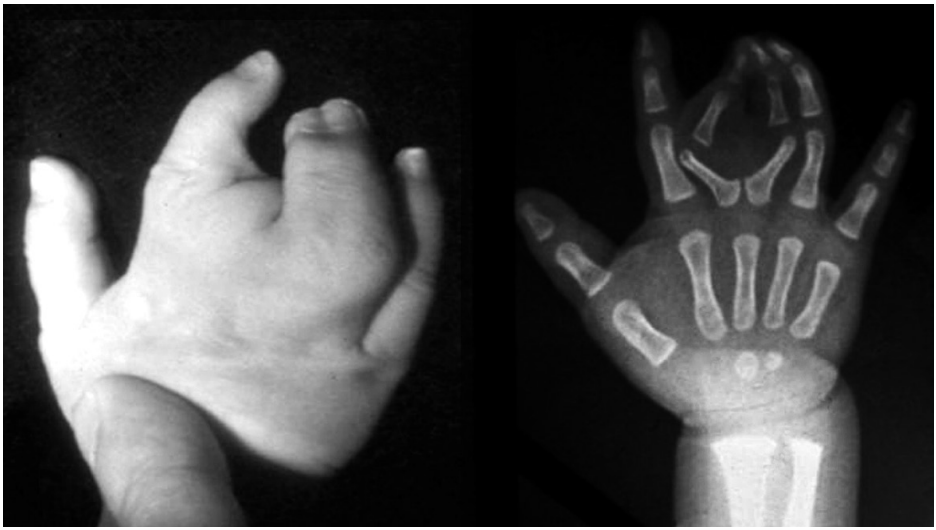

Fig. 28: Central polydactyly associated with cutaneous syndactyly

Syndactyly, central polydactyly & cleft on the palm

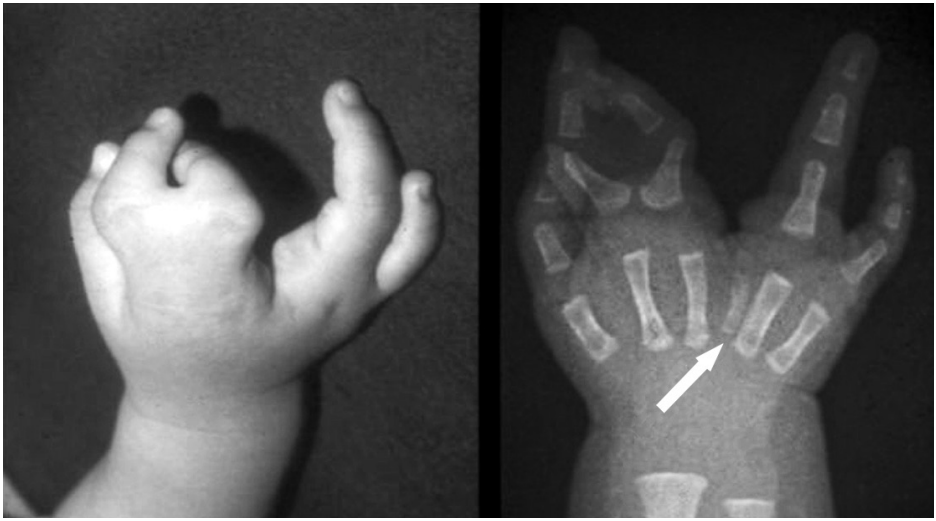

Fig. 29: So–called cleft hand complex: The deformity can be called cleft hand complex, but this hand deformity should be described with combination among syndactyly, central polydactyly, and cleft on the palm, because thre were six metacarpals.

Macrodactyly

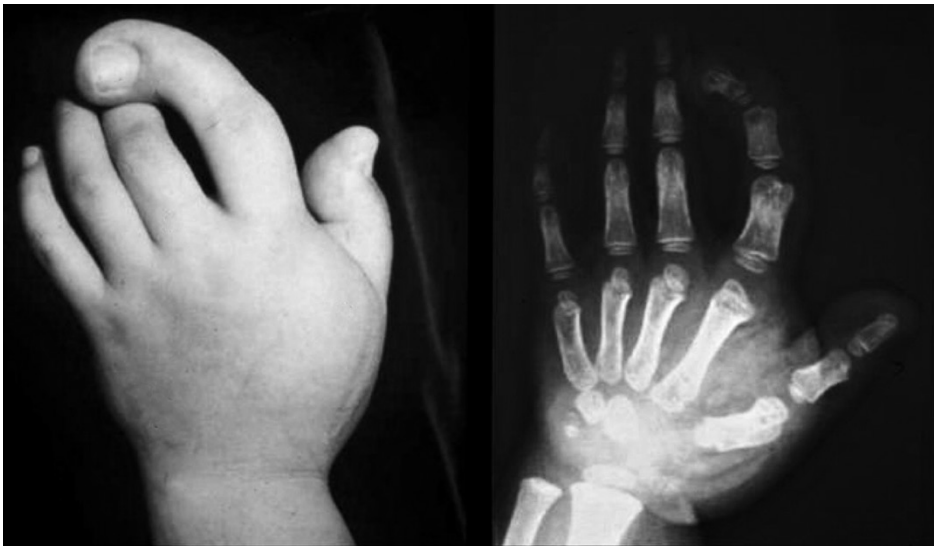

Fig. 30: Macrodactyly: Thumb and index finger were involved. The enlargement of the digital nerves were observed during surgery in this case.

with four-finger defect. In cleft hand, even four digits are absent, hypoplasia of the retained digit, the carpal bone, and the forearm bones does not exist. In ulnar deficiency with four-finger defect, the ulna is completely absent in most cases. Clinodactyly and camptodactyly are sometimes associated with cleft hand. In such cases, associated clinodactyly and camptodactyly should not be classified into other categories, because these deformities seem to be a secondary change due to abnormal induction of the digital rays.

In this category, hand deformity can be expressed with combination of cutaneous syndactyly, cleft of the palm, osseous syndactyly, central polydactyly, triphalangeal thumb, and absence of the central finger rays **Fig. 28**. A cleft hand complex means complex combination of the manifestations of abnormal induction of digital rays. The author would like to recommend that use of the term "Cleft hand complex" should be discontinued, since the multiplicity of the deformity in this category can be expressed with combination of skin and skeletal manifestations **Fig. 29**.

CATEGORY V: OVERGROWTH

This category includes hemihypertrophy and macrodactyly **Fig. 30**. The whole limb, hand or the digits may be affected by overgrowth. Overgrowth due to lymphangioma or hemangioma is classified according to the underlying disease. An aberrant muscle is a rare

Brachydactyly

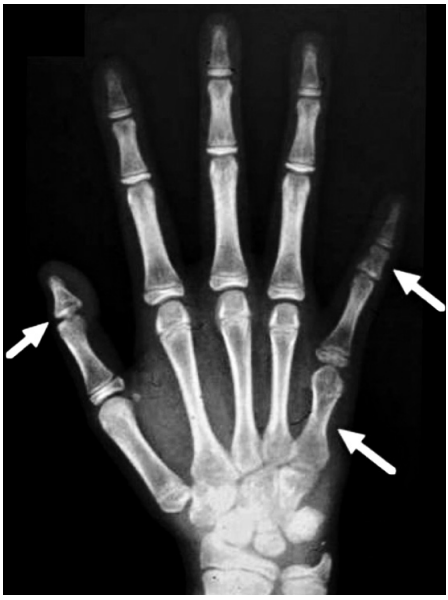

Fig. 31: Brachydactyly of the thumb and little finger: There was brachytelephalangy of the thumb, brachymesophalangy and brachymetacarpia of the little finger.

condition, which also causes enlargement of the digit and the limb, and is classified into failure of differential of part. Congenital hemihypertrophy is known as Silver-Russell syndrome, in which hemihypertrophy, low birth weight, short stature, elevated urinary gonadotrophins, and characteristic facial features, including triangular shaped face with a broad forehead and pointed, small chin with a wide, thin mouth. Asymmetry of the body is also associated.

In macrodactyly, there are two types, such as one without interstitial lipoma of the nerve and the other is one with interstitial lipoma of the nerve. However, sometimes it is not easy to distinguish between these types. Most of macrodactylies are associated with hypertrophy of the fat and/or digital nerves, median nerve, or ulnar nerve (**30**).

CATEGORY VI: UNDERGROWTH

Undergrowth is that category, in which defective or incomplete development of the parts. This category is a supplemental one for the category one, failure of formation of parts. This category includes undergrowth of the whole hand (microcheiria) including undergrowth of the whole limb, brachydactyly, and clinodactyly with shortening of the digit. Brachydactyly includes brachymetacarpia, brachybasophalangy, brachymesophalangy, and brachytelephalangy **Fig. 31**. Clinodactyly is deviation of the digit and it is usually associated with shortening of the phalanx.

CATEGORY VII: CONSTRICTION BAND SYNDROME

Constriction band syndrome is caused by the insult after digital radiations have been formed. Congenital constriction band syndrome has four different expressions, such as constriction band, lymphedema associated with constriction band, acrosyndactyly, and amputation. These expressions appear in various combinations (**33**). However, if a patient has one of these expressions, the diagnosis can be done as congenital constriction band syndrome. In the congenital constriction band syndrome, though the

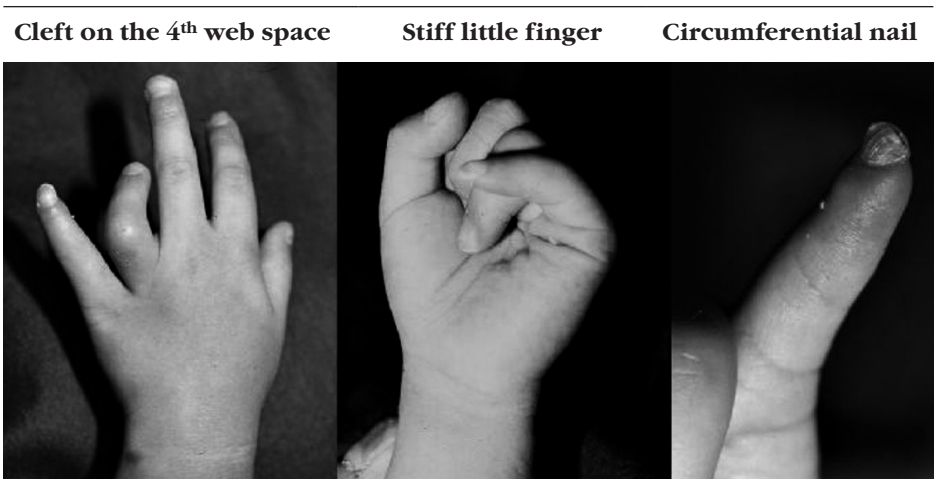

Fig. 32: Unclassifiable case: This is so called ulnar cleft hand without finger defect. In this case, mild cleft of the ring and little finger without finger defect, hypoplasia of the little finger, extension contracture and nail deformity of the little finger.

amputation may extend from the digital tip to the forelimb, or even to the proximal part of the limb, it is considered that no bone dysplasia from the amputated part to the proximal occurs. However, hypoplasia of the metacarpal bones is sometimes observed in adult patients. Hand deformity should be expressed with combination of constriction ring, lymphedema, acrosyndactyly, and amputation type.

CATEGORY VIII: GENERALIZED SKELETAL ABNORMALITIES & A PART OF SYNDROME

Deformities in the hand may be a manifestation of congenital syndromes or that of generalized skeletal abnormalities caused by osteochondrodysplasias. The deformities in this category should be classified according to the underlying conditions, which are true diagnostic name.

CATEGORY IX: OTHERS

This category includes unclassifiable cases. There might be a lot of rare conditions, which cannot be classified according to this classification. There are also relatively common conditions, which cannot be classified according to this classification. In the later group, at least two major groups. One is so called ulnar cleft hand without finger defect, but characteristic clinical features of this condition are different from various types of abnormal induction of digital rays including cleft hand. So called ulnar cleft hand is characterized with combination of various degrees of cleft of the ring and little finger without finger defect, hypoplasia of

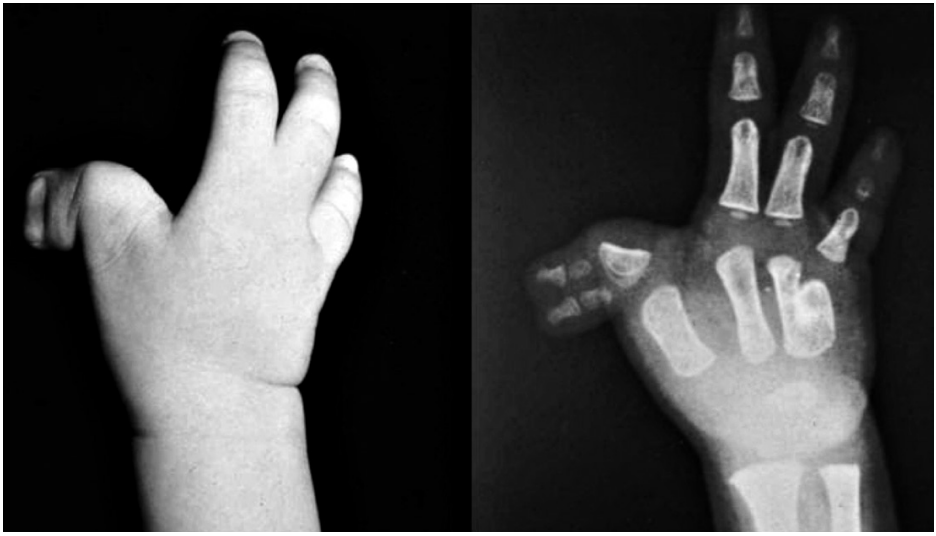

Fig. 33: Unclassifiable case: This deformity was unilateral involvement. The affected hand was hypoplastic compared with the opposite hand. One finger was absent but it is difficult to determine which finger was missing. The absence of the finger ray might be induced by abnormal induction of the digital ray

the little finger, hypoplasia of hypothenar muscles, contracture or symphalangism of the little finger and nail deformity of the little finger **Fig. 32**. In some cases of this condition is associated with ulnar polydactyly or ulnar deficiency. The other is abnormal induction of the finger ray numbers associated with hypoplsia of the affected hand. This deformity is mostly unilateral involvement. The characteristic features of this condition seem to be those of transverse deficiency and those of abnormal induction of digital rays. That means the hand has cleft of the palm, syndactyly, central polydactyly, osseous syndactyly and/or absence of the central fingers. But it is sometimes difficult to say which finger rays are missing. The other characteristic features are unilateral involvement, hypoplasia of the whole affected hand compared to the opposite hand (3) **Fig. 33**.

SUMMARY AND CONCLUSIONS

When Swanson, Barsky and Entin published the original version of the IFSSH classification (37), they stated as "A workable classification must employ a simple, easily remembered terminology. An ideal classification could be made by grouping cases according to etiology. Unfortunately, however, the etiology of many of these defects is still obscure.

Enough basic information has been collected, nonetheless, to group cases according to the parts have been primarily affected by certain embryological failure." According to the advances of molecular biology and genetics, the causes of some of the congenital hand anomalies have been clarified. But most of the etiology of many of these defects is still obscure. The situation has not been changed 33 years after they published the first version of the IFSSH classification. Swanson (**36**) also stated as "The clinician's role in clarifying the pathogenesis of limb defects should be to observe and record the various deformities as he sees them, and then attempt to relate them to the embryologists' and geneticist' studies as to their chronological ontogenetic development. The author agrees their opinion.

In IFSSH classification, polydactyly is classified into duplication, syndactyly into failure of formation of parts and typical cleft hand into failure of formation of parts. However, these congenital deformities may appear when the same teratogenic factor acts on embryo at the same developmental period. Therefore, they are included in the same category of abnormal induction of digital rays in our modified classification. On the other hand, brachysyndactyly is classified into undergrowth and transverse deficiency into failure of formation of parts, and there is no item of atypical cleft hand in IFSSH classification. However, brachysyndactyly, atypical cleft hand or transverse deficiency seems to be developed as morphological variants of symbrachydactyly. Therefore, these deformities are included in the same concept of transverse deficiency as an item of failure of formation of parts. When we use modified classification, the deformities, which belong to the same category, would not be classified into different categories. This is a profitable point of this classification.

In our modification of the IFSSH classification, sub-classification was made according to the morphology and the deformities of each limb were described with combination of the deformity of its part. This method makes possible to describe the multiplicity of the deformities of the limb in each case. This is also a profitable point of this classification.

REFERENCES

1. BLAUTH W. Der hypoplastische Daumen, Archiv für Orthopädische und Unfall-Chirurgie 62: 225–246, 1967 (In German).

2. BLAUTH W, and Gekeler J. Zur Morphologie und Klassifikation der Symbrachydaktylie. Handchirurgie 4: 123–128, 1971.

3. BUCK-GRAMCKO D, OGINO T. Congenital malformation of the hand: non-classifiable cases. Hand Surgery 1: 45–61,1996.

4. CARSTAM N, THEANDER G. Surgical treatment of clinodactyly caused by longitudinally bracketed diaphysis ("delta phalanx"). Scandinavian Journal of Plastic and Reconstrucrive Surgery. 9: 199–202, 1975.

5. Congenital Hand Committee of Japanese Society for Surgery of the Hand. Manual for classification of congenital hand deformities. Jouranal of the Japanese Socety for Surgery of the Hand 13: 455–467,1996.

6. GOLDFARB CA, MANSKE PR, BUSA R, MILLS J, CARTER P and EZAKI M. Upper-Extremity Phocomelia Reexamined: A Longitudinal Dysplasia. J Journal of Bone and Joint Surgery 87A: 2639–2648, 2005.

7. HALL JG and GREENE G. The distal arthrogryposis: Delineation of new entities – review

and nosologic discussion. American Journal of Medical Genetics 11: 185–239, 1982

8. LIGHT TR, and OGDEN JA. The longitudinal epiphyseal bracket: implications for surgical correction. Journal of Pediatric Orthopediscs 1: 299–305, 1981.

9. MANSKE PR. Symbrachydactyly instead of atypical cleft hand. Plastic and Reconstructive Surgery 91: 196, 1993.

10. MARDAM-BEY T, GEL E. Congenital radial head dislocation. Journal of Hand Surgery 4 : 316–320, 1979.

11. MIURA T. Syndactyly and split hand. Hand 8: 125–130, 1976.

12. MÜLLER W. Die angeborenen Fehlbildungen der menschlichen Hand. Georg Thieme Verlag, Leipzig, 1937. P86–99.

NARUSE T, TAKAHARA M, TAKAGI M, and OGINO T. Early morphological changes leading to central polydactyly, syndactyly, and central deficiencies: an experimental study in rats. Journal of Hand Surgery 32A: 1413–1417, 2007.

13. NARUSE T, TAKAHARA M, TAKAGI M, OBERG KC and OGINO T. Busulfan-induced central polydactyly, syndactyly and cleft hand or foot: a common mechanism of disruption leads to divergent phenotypes. Development, Growth and Differentiation, 49: 533–541, 2007.

14. OGINO T. Congenital anomalies of the upper limb in our clinic – An application of modified Swanson's classification. Journal of the Japanese Society for Surgery of the Hand 2: 909–916, 1986. (In Japanese)

15. OGINO T, MINAMI A, FUKUDA K and KATO H. Congenital anomalies of the upper limb among the Japanese in Sapporo. Journal of Hand Surgery 11B: 364–371, 1986.

16. OGINO T, and HIKINO K. Congenital radio-ulnar synostosis: compensatory rotation around the wrist and rotation osteotomy. Journal of Hand Surgery 12B: 173–178, 1987.

17. OGINO T and SAITOU Y. Congenital constriction band syndrome and transverse deficiency. Journal of Hand Surgery 12B: 343–348, 1987.

18. OGINO T, ISHII S, and MIMANI M. Radially deviated type of thumb polydactyly. Journal of Hand Surgery 13B: 315–319, 1988.

19. OGINO T, MIMAMI A and KATO H. Clinical features and roentgenograms of symbrachydactyly. Journal Hand Surgery 14B: 303–306, 1989.

20. OGINO T. Teratogenic relationship between polydactyly syndactyly and cleft hand. Journal of Hand Surgery 15B: 201–209, 1990.

21. OGINO T, and KATO H. Operative findings in camptodactyly. Journal of Hand Surgery Surgery 17B: 661–664, 1992.

22. OGINO T, and KATO H. Clinical features and treatment of congenital fusion of the small and ring finger metacarpals. Journal of Hand Surgery 18-A: 995–1003, 1993.

23. OGINO T, KATO H, OHSHIO I, and TAKAHARA M. Clinical Features of congenital contractural arachnodactyly. Congenital Anomalies 33: 85–94, 1993.

24. OGINO T, TAKAHARA M, KATO H, SUGIMOTO Y, and ISHII S. Clinical features of congenital ankylosis of the digital joints of the hand. Congenital Anomalies 33: 211–219, 1993.

25. OGINIO T., ISHII S, and KATO H. Clinical features and operative findings of congenital flexion deformity of multiple digits. Congenital Anomalies 33: 389–397,1993

26. OGINO T, ISHII S, and KATO H. Opposable triphalangeal thumb. clinical features and results of treatment. Journal of Hand Surgery 19A: 39–47, 1994.

27. OGINO T, ISHII S, and SUGIMOTO Y. Congenital anomalies of the elbow joint – clinical features and classification. Congenital Anomalies 35: 447–454, 1995.

28. OGINO T. Current classification of congenital hand deformities based on experimental research. In Current Practice in Hand Surgery

(eds P. Saffar P, Amadio CP and Foucher G) Martin Dunitz, London 1997, 337–341 p.
29. OGINO, T. Macrodactyly. In Congenital Malformations of the Hand and Forearm (ed Buck-Gramcko D) Churchill Livingstone Inc, Edinburgh 1998, 183–193 p.
30. OGINO T, SATAKE H, TAKAHARA M, KIKUCHI N, WATANABE T, IBA K, and ISHII S. Aberrant muscle syndrome: hypertrophy of the hand and arm due to aberrant muscles with or without hypertrophy of the muscles. Congenit Anom (Kyoto). 50: 133–138, 2010.
31. OGINO T. Teratogenic mechanisms of congenital absence of digits. Locomotor System Advances in Research, Diagnostics and Therapy18: 173–193, 2011.
32. PATTERSON T J S. Congenital ring-constriction. British Journal of Plastic Surgery 14, 1–31, 1961.
33. SATAKE H, OGINO T, TAKAHARA M, WATANABE T, IBA K. Radial longitudinal deficiencies with hypoplastic/absent thumbs and cutaneous syndactyly of the most radial digits. Journal of Hand Surgery 35A: 1497–1501, 2010.
34. SUGIURA Y. Poland's syndrome. Clinico-roentgenographic study on 45 cases. Congenital Anomalies 16: 7–28, 1976.
35. SWANSON A B. The treatment of congenital limb malformations. Surgical reconstruction or prostetic replacement. Kobe Journal of Medical Science 11: 41– 62, 1965.
36. SWANSON AB, BARSKY AJ, and ENTIN MA. Classification of limb malformations on the basis of embryological failures, Surgical Clinnics of North America 48: 1169–1179, 1968.
37. SWANSON A B. A classification for congenital limb malformations. Journal of Hand Surgery 1A: 8–22, 1976.
38. SWANSON AB, SWANSON GG, TADA K. A classification for congenital limb malformations. Journal of Hand Surgery 8: 693–702, 1983.
39. TUPPER JW. Pollex abductus due to congenital malposition of the flexor pollicis longus. Journal of Bone and Joint Surgery 51A: 1285–1290, 1969.
40. WASSEL HD. The result of surgery for polydactyly of the thumb, a review. Clinical Orthopopedics and Related Resarch 64 : 175–193, 1969.
41. ZANCOLLI EA and ZANCOLLI EA Jr. Congenital ulnar drift and camptodactyly produced by malformation of the retaining ligaments of the skin. Bulletin of the Hospital for Joint Diseases Orthopaedic Institute 44: 558–576,1984.

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