APPLICATION OF THE IFSSH[3]-CLASSIFICATION FOR CONGENITAL ANOMALIES OF THE HAND ; RESULTS AND PROBLEMS

L. DE SMET[1], G. MATTON[2], S. MONSTREY[2], E. CAMBIER[2], G. FABRY[1]

The extended classification proposed by the IFSSH was used to classify 1013 hand anomalies in 925 hands of 650 patients.
We found associated anomalies in 26.7%.
The classification was straightforward in 86%, difficult in 6.6% and not possible in 7.8%.
In group I the radial and ulnar deficiencies, limited to the hand and without forearm deficiencies should be included.
Group II was the most important group including 513 anomalies.
We propose to include in this group the Madelung deformity, the Kirner deformity and congenital trigger fingers and trigger thumbs.
Triphalangeal thumbs are a problem, we suggest to list this anomaly in group III and to consider it as a duplication in length.
It is not always possible to evaluate the (transverse) absence of the fingers or hand. Longitudinal deficiencies (group II_B), symbrachydactyly group (V) and amniotic bands (group VI) occasionally develop a phenotype similar to the genuine transverse deficiency (group I_A)

Keywords : congenital ; hand ; classification.
Mots clés : congénital ; main ; classification.

INTRODUCTION

The purpose of a classification for clinical problems is to allow all investigators and surgeons involved to speak the same language. So-doing, syndromes can be delineated, frequencies of occurence can be established and results of different treatments can be compared.

A classification system should be simple to use, reliable and uniformly accepted. It should allow space for adaptations and/or excursions.

The IFSSH (1976) proposed a 7 categories classification based on the original classification of Swanson *et al.* (9, 10) in 1968 (see appendix 1). The aim of this survey is to review a large series of cases, using the IFSSH classification, to compare it with previously published series, to note difficulties and problems encountered in using this classification, and to make suggestions for adapting the classification.

MATERIAL AND METHODS

The files, radiographs and photographs of all patients consulting for congenital anomalies of the hand from 2 university hospitals were reviewed. The extended classification proposed by the IFSSH and presented in 1988 in Paris (11), was used. Classification of a case was considered *obvious* when there was no doubt, *difficult* when one had to look more carefully to label the case (mostly because a diagnosis could fit into 2 or more categories) and was considered *impossible* when the diagnosis was not at all withheld in the IFSSH classification.

Difficult and impossible cases were debated among the different authors.

[1] Dept. Orthopaedic Surgery, University Hospital Pellenberg, K.U. Leuven, Belgium.
[2] Dept. Plastic Surgery, University Hospital Ghent, Belgium.
[3] International Federation for Societies for Surgery of the Hand.
Correspondence and reprints : L. De Smet, U.Z. Pellenberg, Weligerveld 1, B-3212 Pellenberg.

RESULTS

We could review 1013 anomalies in 925 hands of 650 patients. There were 320 females (49.3%) and 330 males (50.7%) ; in 454 (49.2%) patients only the right hand, in 467 (50.7%) patients the left hand and in 274 patients (41%) both hands were involved. All hands were treated at Leuven University (LDS) (553 patients), and at Ghent University (GM, SM, EC) (372 patients).

Associated anomalies were seen in 243 hands (26.7%). An operative treatment was performed in 466 hands (51.8%).

A classification was considered obvious in 85.6% of the anomalies, *difficult* in 6.6% (61) and *impossible* in 7.8% (68). A more detailed distribution of the anomalies is seen in table I.

Table I. — Distribution of the different congenital anomalies among the 7 groups

I.	*Failure of formation* :	184 (19.8%)
	A) Transverse arrest :	44
	B) Longitudinal arrest :	140
II.	*Failure of differentiation* :	513 (55.4%)
	Soft tissue :	348
	Skeletal :	128
	Congenital tumors :	37
III.	*Duplication* :	143 (15.4%)
IV.	Overgrowth :	7 (0.7%)
V.	Undergrowth :	77 (8.3%)
VI.	Constriction band syndrome :	60 (6.4%)
VII.	Generalized skeletal anomalies :	29 (3.1%)

Group I : failure of formation of parts (arrest of development).

Transverse failures of formation (Subgroup I_A) most frequently occur at the forearm level. Those at the finger level were only accepted after exclusion of other causes of congenital "amputations" such as constriction bands and severe symbrachydactyly (grade 3 and 4) (Fig.1). The soft tissue envelope should be markedly less involved than the bone, and the rest of the proximal upper limb should not be involved at all.

Longitudinal arrest (subgroup I_B) is subdivided into radial, ulnar, central and intercalated (Table II). We added thumb hypoplasia and aplasia to

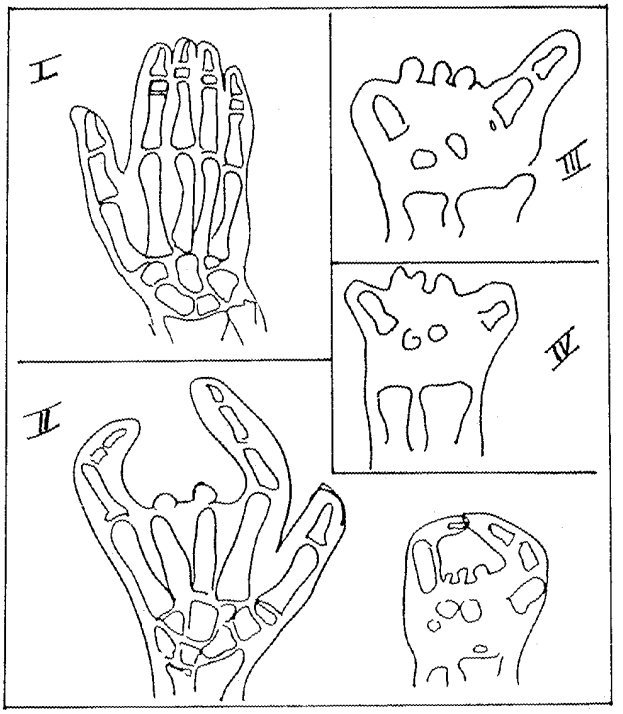

Fig. 1. — Teratological involution series of symbrachydactyly.

Table II. — Longitudinal deficiencies (A), transverse deficiencies (level of absence) (B)

A) LONGITUDINAL DEFICIENCIES		
Radial :	69	
	hypoplastic radius :	5
	partial aplasia :	3
	complete aplasia :	15
	thumb anomaly :	46
Ulnar :	41	
	partial hypoplasia :	1
	complete aplasia :	8
	with humeroradial synostosis :	3
	ulnar finger aplasia :	29
Central :	29	
	typical :	17
	atypical :	12
Intercalated (phocomelia) :		1
B) TRANSVERSE DEFICIENCIES		
	Upper arm :	2
	Forearm :	16
	Wrist :	1
	Carpus :	5
	Metacarpals :	8
	Phalanges :	12

the radial deficiency since it is missing in the IFSSH classification. The same was done for the deficiency of ulnar fingers.

The atypical cleft hand is considered by several authors as a grade 2 of symbrachydactyly. In this survey it was left included in group I_B.

Group II : failure of differentiation (separation) of parts (Table III).

This is the largest group (513 anomalies or 55.4%). In the IFSSH classification congenital trigger fingers and thumbs were missing, but we included them in this group. Syndactyly is the most frequent anomaly (200 hands), followed by camptodactyly (80), trigger fingers (63) and clinodactyly (57).

Table III. — Failure of differentiation

Syndactyly :	200	
cutaneous :		167
osseous :		33
Campodactyly :	80	
digit 5 :		57
others :		23
Trigger finger :	63	
Clinodactyly :	57	
delta bone :		14
idiopathic :		39
without bone deformity :		4
Synostosis :	45	
elbow :		1
radioulnar :		13
carpal :		16
symphalangism :		15
Congenital tumorous condition :	37	
Others :		
arthrogryposis :		14
Sprengel :		5
aberrant muscles :		1
thumb in palm :		7

Group III : polydactyly.

This is the second largest group, with a higher frequency of preaxial (142 hands) versus postaxial polydactyly (57 hands). Central polydactyly remains rare (9 cases). One mirror hand was also observed.

Group IV : overgrowth.

This congenital difference is the rarest : only 7 cases were seen in both departments.

Group V : undergrowth

Only those anomalies in which hypoplasia was the major feature (and the major reason to search medical advice) were included in this group. In fact most hands with congenital anomalies are smaller than the normal ones.

Brachymetacarpia was seen in 25 hands, brachydactyly in 20 hands and symbrachydactyly in 32 hands. Ten patients with symbrachydactyly had an absence of the ipsilateral pectoral muscle and were considered as Poland syndrome.

Group VI : congenital constriction band syndrome.

Most hands (35) in this group had combined features of the constriction band syndrome. Constriction bands only were seen in 7 hands, acrosyndactyly in 8 hands and intra-uterine amputations in 10 hands.

Group VII : generalized skeletal abnormalities.

Twenty-nine patients had a multiple congenital anomaly (MCA) syndrome, of whom 5 had chromosomal abnormalities.

DISCUSSION

From this review it is obvious that the IFSSH classification is a useful tool for a majority of the congenital deformities. In the current series several frequently observed anomalies could not be fitted into the IFSSH-classification. Therefore the authors propose to add several subgroups to the IFSSH- classification, taking care to respect the whole concept.

In group I_B (longitudinal deficiency) hypoplasia and aplasia of the thumb, without forearm or wrist deficiency, should be added.

Kirner's deformity, Madelung's deformity, congenital trigger finger and trigger thumb should be added to group II (failures of differentiation).

Triphalangeal thumbs present a problem for classification. We propose to include this anomaly in group III (polydactyly) since it is frequently associated with duplication deformities (12) and the isolated triphalangeal thumb can be seen as an excess of material in the length, but it can also be seen as a failure of differentiation with equal arguments.

In group VII (generalized-skeletal-syndromes) most hand deformities can be classified into one of the former groups. In Swanson's publication (10) Madelung deformity is included in this group, but isn't that a failure of differentiation also ?

Absence of a part of the hand poses a problem. Is it a genuine transverse deficiency (Group I_A) - a further involvement of a (ulnar or central) longitudinal deficiency (Group I_B), a consequence of amniotic ring syndrome (Group VI) or an extreme hypoplasia, i.e. symbrachydactyly grade IV (Group V) (1) ?

We suggest that "amputations" proximal to the wrist, without other signs of constriction rings, should be considered as a transverse deficiency. When fingers are missing in a hypoplastic hand and upper limb, it is usually a symbrachydactyly (Group V), certainly when central digits are missing, when they are syndactylous and/or when the middle phalanges are mostly involved. When fingers are missing and skeletal involvement is more important than soft tissue deficiency, and hand size is almost normal, we consider it as a transverse deficiency (Group I_A).

The evolving ideas about symbrachydactyly extended this "embryological- teratological" failure and 4 groups can be considered : the typical symbrachydactyly (group 2 or 5), the atypical cleft hand (group I_B), the monodactylous hand Group I_B) and the amelia (Group I_A) (Fig. 1). In 1990 Ogino described a teratological linkage between central polydactyly (Group 3), osseous syndactyly (Group 2) and typical cleft hand (Group I_B) (8).

Despite all these considerations the IFSSH classification cannot be used in 15.4% of the anomalies and inclusion of all these - complex - cases should extend the list too much to make it useful for clinical use.

The frequency of congenital anomalies in different populations has been studied by several authors. In 1982 Lamb *et al.* (4) published a first report, based on information from members of The Congenital Malformations Committee of the IFSSH. This report reflects the worldwide distribution of congenital anomalies (Table IV). Other large series have been summarized in tables IV and V. Syndactyly and polydactyly are the most frequently encountered anomalies, although polydactyly and longitudinal deficiencies are very frequent in the Chinese and Japanese populations. This Belgian population survey does not differ fundamentally from the others, except for a relatively high incidence of longitudinal deficiencies (Table V).

Table IV. — Distribution among different populations

	N	Transv. arrest	Long. arrest	Hypoplasia	Ring constrict	Polydact.	Macrodact.	Group II
Hong Kong (2)	728	5.6%	6.6%	4.3%	6.5%	35.9%	0.5%	31.8%
Lamb et al. (4) (Multicenter all over the world) (1976-1978)	1804	4.8%	13%	14.4%	4%	20.2%	1%	41.1%
Masada et al. (6)	374	not included	13.4%	15.8%	9.4%	33.4%	1.6%	22.5%
This series	1013	4.3%	13.8%	8.3%	6.4%	15.4%	0.7%	55.4%

Table V. — Most frequent anomalies in different populations

Iowa (3) (N=1476)		Yokohama (13) (N=250)		Hong Kong (5) (N=326)		Sapporo (7) (N=955)		This Series (N=1013)	
Type	%	Type	%	Type	%	Type	%	Type	%
Syndactyly	(19.1)	Polydactyly	(25.0)	Polydactyly	(39.3)	Trigger finger	(21.0)	Syndactyly	(19.7)
Polydactyly	(14.8)	Syndactyly	(15.6)	Syndactyly	(14.9) 3	Polydactyly	18	Polydactyly	(14.1)
Cong. Amput.	(11.0)	Arthrogryposis	(11.0)	Syndromes	(11.9) 5	Camptodactyly	6.	Longitudinal arrest	(13.8)
Campodactyly	(6.5)	Brachydactyly	(6.0)	Transverse arrest	(6.8)	Clapsed thumb	(5.5)	Camptodactyly	(7.9)

REFERENCES

1. Blauth W., Gekeler J. Zur Morfologie und Klassification der Symbrachydactylien. Handchir., 1971, 4, 123-172.
2. Cheng J., Chow S., Leung P. Classification of 578 cases of congenital limb anomalies with the IFSSH system. J. Hand Surg., 1987, 10-A, 1055-1060.
3. Flatt A. The care of congenital hand anomalies. Quality Medical Publishing, St. Louis, 1994.
4. Lamb D., Wynne-Davies R., Soto L. An estimate of the population frequency of congenital malformations of the upper limb. J. Hand Surg., 1982, 7, 557-562.
5. Leung P., Chan K., Cheng J. Congenital anomalies of the upper limb among the Chinese population in Hong Kong. J. Hand Surg., 1982, 7-A, 1055-1060.
6. Masada K., Tsuyugchi Y., Kawabata H., Kawai H., Tada K., Ono K. Terminal limb malformation : analysis of 523 cases. J. Ped. Orthop., 1986, 6, 340-45.
7. Ogino T., Minami A., Fukudu K., Kato H. Congenital anomalies of the upper limb among the Japanese in Sapporo. J. Hand Surg., 1986, 11-B, 364-371.
8. Ogino T. Teratogenic relationship between polydactyly, syndactyly and cleft hand. J. Hand Surg., 1990, 15-B, 201-209.
9. Swanson A., Barsky A., Entin M. Classification of limb malformations on the basis of embryological failures. Surg. Clin. North Am., 1968, 48, 1169-1179.
10. Swanson A. A classification for congenital hand malformations. J. Hand Surg., 1976, 1, 8-22.
11. Swanson A., De Groot B., Swanson G. Une classification pour les malformations des membres. In : Gilbert A., Buck-Gramko D., Lister G., eds. Les malformations congénitales du membre supérieur. Monographies du GEM. Paris : Expansion Scientifique, 1991, 1-18.
12. Wasser H. The results of surgery for polydactyly of the thumb. Clin. Orthop., 1969, 64, 175-193.
13. Yamatuchi Y. Proceedings of the 16th annual meeting of the Japanese Society for Surgery of the Hand. Fukuoka, Japan, 1973 (referred in Flatt).

SAMENVATTING

L. DE SMET, G. MATTON, S. MONSTREY, E. CAMBIER, G. FABRY. Gebruik van de IFSSH classificatie voor aungeboren afwijkingen van de hand : resultaten en problemen.

De uitgebreide classificatie voorgesteld door de IFSSH werd aangewend bij 1013 handafwijkingen in 925, handen bij 650 patiënten.
In groep I zouden ook de longitudinale deficiencies van de hand, zonder onderarm afwijkingen kunnen worden ondergebracht.
Geassocieerde afwijkingen werden gevonden in 26.7% der gevallen. Groep II is met 513 gevallen de grootste groep. Wij stellen voor om in die groep ook Madelung deformatie, Kirner deformatie en congenitale springvinger (en - duim) onder te brengen.
De triphalangeale duim blijft een probleem en kan best in groep III worden ondergebracht.
Ook bij dwarse deficienties is het niet altijd mogelijk om de subtypes te onderscheiden.
Longitudinale deficienties (groep II_B), symbrachydactylie (groep V) en amnion strengen (groep VI) kunnen zich als een gelijkaardig fenotype uiten.

RÉSUMÉ

L. DE SMET, G. MATTON, S. MONSTREY, E. CAMBIER, G. FABRY. L'application de la classification IFSSH pour les déformations congénitales de la main : résultats et problèmes.

La classification proposée par l'IFSSH a été appliquée à 1013 déformations touchant 925 mains chez 650

patients. Des anomalies associées ont été retrouvées dans 26.7% des cas. Le groupe II est le plus important avec 513 cas. Nous proposons d'y ajouter la déformation de Madelung, la déformation de Kirner et les doigts à ressaut.

Dans le groupe I il faut ajouter les déficiences longitudinales isolées de la main, sans atteinte de l'avant-bras.

Le pouce à trois phalanges reste un problème. Selon nous son incorporation dans le groupe III est la plus raisonnable.

Pour les déficiences transversales (groupe I_A) il n'est pas toujours évident de différentier les sous-types : des déficiences longitudinales (groupe I_B), les symbrachydactylies (groupe V) et les brides amniotiques (groupe VI) peuvent s'exprimer comme une déficience transversale (groupe I_A).

APPENDIX 1

Swanson A. B., de Groot Swanson G. Une classification pour les Malformations Congénitales des Membres. In : Les Malformations Congénitales du Membre Supérieur. Monographies du Groupe d'Etude de la Main. Ed. R. Tubiana, Paris 1991.

1. Failure of formation of parts (arrest of development)
   - A. Transverse arrest
     1. Shoulder
        - a. amelia
     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
     5. Wrist
        - a. wrist level
     6. Carpal
        - a. carpal level
     7. Metacarpal
        - a. metacarpal level
     8. Phalanges
        - a. phalangeal level
   - B. Longitudinal deficiencies
     1. Radial ray
        - a. radial ray deficiency
          1. hypoplasia of the radius
          2. partial absence of the radial ray
          3. total absence of the radial ray
     2. Ulnar ray
        - a. ulnar ray deficiency
          1. partial absence of the ulna
          2. total absence of the ulna
          3. defect of the ulna with humeroradial synostosis
          4. defect of the ulna with a congenital amputation at the wrist
     3. Central ray (cleft hand)
        1. typical type (deficiency type)
        2. atypical type
           - a. syndactyly type
           - b. polydactyly type
     4. Intersegmental (intercalated)
        - a. Phocomelia
          1. total
          2. partial

2. Failure of differentiation (separation) of parts
   - a. soft tissue involvement
     1. Disseminated
        - a. Arthogryposis multiplex
          1. Severe form
          2. Moderate form
          3. Mild form
     2. Shoulder
        - a. Undescended shoulder
        - b. Absence of pectoral muscle or muscles
          absent pectoralis major muscle
          absent pectoralis major and minor
     3. Elbow and forearm
        - a. Aberrant muscles
          1. long flexors
          2. long extensors
          3. intrinsic muscles
     4. Wrist and hand
        - a. Cutaneous syndactyly
          1. radial (1 interdigit web)
          2. Central (2 & 3 interdigit web)
          3. Ulnar (4 interdigit web)
        - b. Congenital flexion contracture (Camptodactyly)
          1. little finger
          2. other fingers

c. Thumb in palm deformity
d. Deviated finger without bony lesions

b. Skeletal involvement

1. Shoulder
   a. Congenital humerus varus
2. Elbow
   a. Elbow synostosis
      1. Humeroradial
      2. Humeroulnar
      3. Total
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. Osseous syndactyly
      1. Radial (between 1/2 dig ray)
      2. Central (between 2/3 and 3/4 dig ray)
      3. Ulnar (between 4/5 dig ray)
      4. Mitten hand (incl. Apert)
   b. Synostosis of the carpal bones
      1. Lunate - triquetrum
      2. Capitate - hamate
      3. Seaphoid - lunate
      4. Others
   c. Symphalangism
      1. Prox interphal joint
      2. Dist interphal joint
   d. Clinodactyly
      1. Idiopathic
      2. resulting from a delta bone

c. Congenital tumorous conditions

1. Vascular tumors
   a. Port wine stain
   b. cavernous hemangioma
   c. arterio-venous fistula
2. Lymphatic tumors
   a. lymphangioma
3. Neurogenic tumors
   a. neuroblastoma
   b. neurofibromatosis
   c. others
4. Connective tissue tumors
   a. juvenile aponeuronic fibroma
   b. others
5. Skeletal tumors
   a. osteochondromatosis (multiple exostoses)
   b. others

3. Duplication

1. Whole limb
2. Humerus
3. Radius
4. Ulnar
   a. Mirror Hand
5. Digits
   a. Polydactyly
      1. Radial (preaxial)
      2. Central
      3. Ulnar (postaxial)

4. Overgrowth

1. Whole limb
2. Partial limb
3. Digits
   a. Macrodactyly
      1. Without interstitial lipoma or nerve
      2. With interstitial lipoma or nerve

5. Undergrowth

1. Whole limb
2. Whole hand
3. Metacarpal
   a. Brachymetacarpia
4. Digits
   a. Brachysyndactyly
      1. Without absence of pectoral muscles
      2. With absence of pectoral muscles (Poland syndrome)
   b. Brachydactyly
      1. Brachymesophalangia
      2. Brachydactyly : involving prox. middle and distal phalanges
      3. Brachydactyly with absence of the phalanx

6. Congenital constriction band syndrome

A. Focal necrosis
   a. Constriction bands
      1. With lymphedema
      2. Without lymphedema
   b. Acrosyndactyly
   c. Intrauterine amputation
   d. Combinations of a., b. and c.

7. Generalized skeletal abnormalities

a. Chromosal abnormalities
b. Other generalized abnormalities