IFSSH Scientific Committee on Congenital Conditions

Chair: **M Ezaki (USA)**

Committee: **GH Baek (Korea)**
E Horii (Japan)
S Hovius (Netherlands)

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Classification of Congenital Hand and Upper Limb Anomalies

Our committee recommends adoption of a revised classification system for Congenital Anomalies of the Hand and Upper Limb. The Swanson Classification was proposed in the 1960s and was subsequently adopted by the IFSSH Congenital Conditions Committee. This classification has been of significant benefit, but remains based on a morphological rather than etiological system which is unable to adapt to increasing knowledge.

Dr. Paul Manske, along with Drs. Michael Tonkin and Kerby Oberg, began working on a classification system that would incorporate newer knowledge of the etiology, molecular genetics, and developmental biology into our thinking about these conditions. After Dr. Manske's death, the work has been refined by Drs. Tonkin and Oberg, and vetted by the members of the committee and an international study group. The current document has been discussed extensively. While no classification system is perfect, this OMT system has the ability to be flexible and respond to new developments and additional conditions. The classification of some conditions remains controversial and, for these, the principle of "best fit" has been utilized. It is designed to be a working classification that can be revised and/or modified.

It should be noted that the classification system, as proposed below, describes main groups and sub-groups, but allows for addition of surgical sub-classifications (such as those for thumb duplication, thumb hypoplasia, cleft hand, etc) within an expanded version.

The IFSSH Scientific Committee on Congenital Conditions recommends adoption of the OMT classification with reviews at 3-year intervals according to responses from the hand surgery community. The Committee invites comments and advice based on improvements in knowledge at the time of these reviews when the classification may be modified.

NB: This report contains the OMT Classification as approved by the IFSSH Scientific Committee on Congenital Conditions (February 3rd, 2014). This classification will be updated from time to time.

The current updated version of the OMT Classification can be accessed by referring to the link within the 2014 Congenital Conditions section of the Scientific Committee Reports page (http://ifssh.info/download.html)

OMT CLASSIFICATION OF CONGENITAL HAND AND UPPER LIMB ANOMALIES

Approved by the IFSSH Scientific Committee on Congenital Conditions, 3rd February 2014

I. MALFORMATIONS

A. Abnormal axis formation/differentiation—entire upper limb

1. Proximal-distal axis

i. Brachymelia with brachydactyly

ii. Symbrachydactyly
   a) Poland syndrome
   b) Whole limb excluding Poland syndrome

iii. Transverse deficiency
   a) Amelia
   b) Clavicular/scapular
   c) Humeral (above elbow)
   d) Forearm (below elbow)
   e) Wrist (carpals absent/at level of proximal carpals/at level of distal carpals) (with forearm/arm involvement)
   f) Metacarpal (with forearm/arm involvement)
   g) Phalangeal (proximal/middle/distal) (with forearm/arm involvement)

iv. Intersegmental deficiency
   a) Proximal (humeral – rhizomelic)
   b) Distal (forearm – mesomelic)
   c) Total (Phocomelia)

v. Whole limb duplication/triplication

2. Radial-ulnar (anterior-posterior) axis

i. Radial longitudinal deficiency - Thumb hypoplasia (with proximal limb involvement)

ii. Ulnar longitudinal deficiency

iii. Ulnar dimelia

iv. Radioulnar synostosis

v. Congenital dislocation of the radial head

vi. Humeroradial synostosis - Elbow ankyloses

3. Dorsal-ventral axis

i. Ventral dimelia
   a) Furhmann/Al-Awadi/Raas-Rothschild syndromes
   b) Nail Patella syndrome

ii. Absent/hypoplastic extensor/flexor muscles

4. Unspecified axis

i. Shoulder
   a) Undescended (Sprengel)
   b) Abnormal shoulder muscles
   c) Not otherwise specified

ii. Arthrogryposis

B. Abnormal axis formation/differentiation—hand plate

1. Proximal-distal axis

i. Brachydactyly (no forearm/arm involvement)

ii. Symbrachydactyly (no forearm/arm involvement)

iii. Transverse deficiency (no forearm/arm involvement)
   a) Wrist (carpals absent/at level of proximal carpals/at level of distal carpals)
   b) Metacarpal
   c) Phalangeal (proximal/middle/distal)

2. Radial-ulnar (anterior-posterior) axis

i. Radial deficiency (thumb - no forearm/arm involvement)

ii. Ulnar deficiency (no forearm/arm involvement)

iii. Radial polydactyly

iv. Triphalangeal thumb

v. Ulnar dimelia (mirror hand – no forearm/arm involvement)

vi. Ulnar polydactyly

3. Dorsal-ventral axis

i. Dorsal dimelia (palmar nail)

ii. Ventral (palmar) dimelia (including hypoplastic/aplastic nail)

4. Unspecified axis

i. Soft tissue
   a) Syndactyly
   b) Camptodactyly
   c) Thumb in palm deformity
   d) Distal arthrogryposis

ii. Skeletal deficiency
   a) Clinodactyly
   b) Kirner's deformity
   c) Synostosis/symphalangism (carpal/metacarpal/phalangeal)

iii. Complex

a) Complex syndactyly
b) Synpolydactyly— central
c) Cleft hand
d) Apert hand
e) Not otherwise specified

II. DEFORMATIONS

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

III. DYSPLASIAS

A. Hypertrophy

1. Whole limb
i. Hemihypertrophy
ii. Aberrant flexor/extensor/intrinsic muscle

2. Partial limb
i. Macrodactyly
ii. Aberrant intrinsic muscles of hand

B. Tumorous conditions

1. Vascular
i. Hemangioma
ii. Malformation
iii. Others

2. Neurological
i. Neurofibromatosis
ii. Others

3. Connective tissue
i. Juvenile aponeurotic fibroma
ii. Infantile digital fibroma
iii. Others

4. Skeletal
i. Osteochondromatosis
ii. Enchondromatosis
iii. Fibrous dysplasia
iv. Epiphyseal abnormalities
v. Others

IV. SYNDROMES*

A. Specified

1. Acrofacial Dysostosis 1 (Nager type)
2. Apert
3. Al-Awadi/Raas-Rothschild/Schinzel phocomelia
4. Baller-Gerold
5. Bardet-Biedl Carpenter
6. Catel-Manzke
7. Constriction band (Amniotic Band Sequence)
8. Cornelia de Lange (types 1-5)
9. Crouzon
10. Down
11. Ectrodactyly-Ectodermal Dysplasia-Clefting
12. Fanconi Pancytopenia
13. Fuhrmann
14. Goltz
15. Gorlin
16. Greig Cephalopolysyndactyly
17. Hajdu-Cheney
18. Hemifacial Microsomia (Goldenhar syndrome)
19. Holt-Oram
20. Lacrimoauriculodentodigital (Levy-Hollister)
21. Larsen
22. Leri-Weill Dyschondrosteosis
23. Moebius sequence
24. Multiple Synostoses
25. Nail-Patella
26. Noonan
27. Oculodentodigital dysplasia
28. Orofacialdigital
29. Otopalataldigital
30. Pallister-Hall
31. Pfeiffer
32. Poland
33. Proteus
34. Roberts-SC Phocomelia
35. Rothmund-Thomson
36. Rubinstein-Taybi
37. Saethre-Chotzen
38. Thrombocytopenia Absent Radius
39. Townes-Brock
40. Trichorhinophalangeal (types 1-3)
41. Ulnar-Mammary
42. VACTERLS association

B. Others

*The specified syndromes are those considered most relevant; however, many other syndromes have a limb component categorized under "B. Others".