



IFSSH Scientific Committee on Congenital Conditions

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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) Fuhmann/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".