

# Classification of Congenital Upper Limb Anomalies

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Congenital anomalies of the upper limb, with a prevalence of 1 in 506 births<sup>1</sup> and more than 90 diagnostic conditions with 20 among them having their own specific classifications, are extremely diverse in their manifestations and the combination of these anomalies seems to defy a classification (*fig -1*). But these anomalies demand a reproducible and consistent terminology, for example, aborted fingers rather than *ectrodactylism*, in a simple yet precise language that would be understandable by all in the English-speaking world and can be readily translated into other languages which will allow us to discuss the clinical entity, treatment modalities and comparison of results.



**Figure 1:** Anomalies of the hand presenting as wide spectrum and sometimes difficult to classify

For many years, a wide assortment of Greek and Latin words was used to describe the congenital upper limb anomalies. Those were descriptive classification, based on the deformity, such as the radial club hand, which describes a radially deviated hand looking like a 'golf club'. They could be useful for diagnosis in clinical practices but boast little scientific value. These terminologies led to perplexity among those who were brought into the research of congenital limb deficiencies following the thalidomide epidemics in 1960 and among statisticians in finding the true incidence and prevalence.

Apart from being descriptive of the deformity, an ideal classification should include aetiology, detailing the topography of the lesion, guiding treatment and provide prognosis. It should be widely acceptable and used, such that communication among embryologist, anatomist, pathologist, surgeon, paediatrician, statistician, parents and/or the children is easily possible. It should utilise a reproducible and consistent terminology.

### ***Historical perspective***

The earliest attempt of classifying limb anomalies was by Isidore Geoffroy St.Hilaire in 1832, who classified them as *ectromeliens* and *symeliens* and introduced the terms such as "*phocomele*" (seal limb), "*hemimele*" (part of limb missing) and "*ectromele*" (limb absence)<sup>2</sup>. In the words of St. Hilaire, the *ectromeliens* are characterized "by more or less complete malformation of one or more limbs."

Hundred years later, Muller noted that malformations could present as a continuum in varying degrees or in different stages of development<sup>3</sup>. This concept of "teratological progression" allowed anomalies to be

graded according to the morphologic severity and it also allowed grouping of anomalies that might have different morphologic appearances, thus simplifying classification schemes.

The further significant progress was made in 1951 when, O'Rahilly proposed a simple classification of long bone deficiencies, depending on whether the defect resulted in total absence of all structures distal to a certain point (terminal) or whether defect was segmental leaving a normal distal component (intercalary defect)<sup>4</sup>. He used the term 'hemimelia' to denote the partial absence of a limb.

Ten years later, in 1961, the first widely accepted and useful classification was proposed by Frantz and O'Rahilly<sup>5</sup>. They introduced the terms terminal and intercalary defect and further sub classified them into transverse and longitudinal deficiency (pre or post axial). Transverse terminal defects were described as *amelia* (total absence of arm), *hemimelia* (partial absence of limb), *acheiria* (absence of hand), *adactyly* (absence of digits), or *aphalangia* (absence of phalanges), depending on the level of deficiencies. Transverse intercalary defects were described as *phocomelias* ('seal limb'). Longitudinal defects were described as pre and post axial and all missing bones were listed. In 1966, Burtch and Kay revised the classification, retaining the four main categories of terminal transverse, terminal longitudinal, intercalary transverse, and intercalary longitudinal defects<sup>6</sup>. The term 'amelia' was retained but all other terms were removed and replaced with the term 'meromelia'. The concept of central longitudinal deficiency (cleft hand), in addition to pre and post axial was introduced.

When Alfred Swanson felt that it was difficult to devise a classification between one which is so general that it has no value and one which is more detailed that its use becomes impossible, he proposed his idea of

grouping these anomalies **based on the parts that have been primarily affected by certain embryologic failures**<sup>7</sup>. The classification provided a comprehensive, yet simple system, which allowed the classification of simple and complex abnormalities alike, in a straightforward language avoiding the usage of confusing Latin and Greek terms. This was universally accepted and was recommended by International Federation of Societies for Surgery of the Hand (IFSSH) in 1974<sup>8</sup>.

The working group of International Society for Prosthetics and Orthotics met in Dundee in 1973 and in Montreux in 1974, made the conclusion that all intercalary defects had some aberration of the terminal segments of the limb, henceforth there was no actual transverse intercalary defect, and these should be considered as longitudinal defects<sup>9</sup>. Therefore, all limb deficiencies are grouped into either transverse or longitudinal deficiencies.

In 1976, Swanson published a refined version of IFSSH classification as previously proposed after a period of clinical testing<sup>10</sup>. The category of ‘failure of formation of parts’ were subdivided into longitudinal and transverse deficiencies following recommendation of the working committee as described above. The category of ‘failure of differentiation of parts’ was expanded and a complete new category ‘undergrowth’ was introduced there by making seven categories that are present in the current classification.

	<b><i>IFSSH/Swanson Classification (1976)</i></b>
I	Failure of formation of parts (arrest of development)
II	Failure of differentiation of parts (separation)
III	Duplication
IV	Overgrowth (gigantism)
V	Undergrowth (hypoplasia)
VI	Congenital constriction band syndrome
VII	Generalised skeletal abnormalities

In 2000, the Japanese Society for Surgery of the Hand suggested a modification, adding two additional groups: abnormal induction of rays and unclassifiable cases<sup>11</sup>. They attempted to include abnormal induction of rays as a causative factor for a group of anomalies.

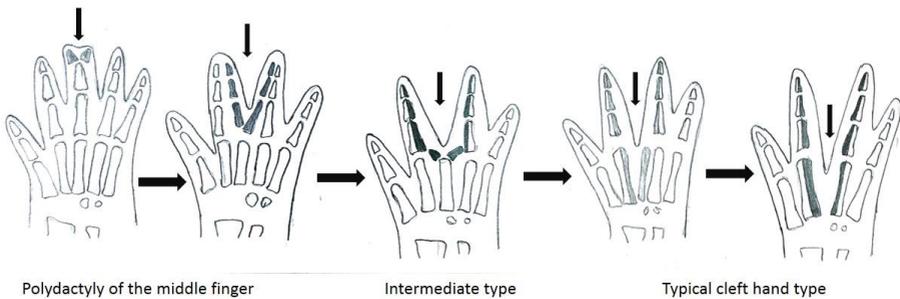
Tonkin contended that grouping according to such categories as failed formation, failed differentiation, or duplication may be inappropriate. He proposed focusing the classification purely on descriptive features, with the primary classification noting the location (i.e., arm, forearm, wrist, or hand) and subcategories listing the tissue involved (bone or soft tissue) as well as the specific morphologic features of the anomalies<sup>12</sup> (Annexure - 1). Later, in attempts to combine the molecular basis of the causation of the deformity and the description of the deformity, Oberg Manske and Tonkin (OMT) classification succeeded the Swanson classification and is being recommended by the IFSSH since 2014<sup>13</sup> (Annexure - 2).

### ***The Rise and fall***

The classification proposed by Swanson, an achievement, what could be considered as ground breaking in the field of congenital upper limb anomalies has endured a large volume of criticism since its inception. With the premise of grouping the anomalies according to the parts primarily affected by certain embryological failure, this classification enjoyed warm welcome and was embraced by the IFSSH with modifications. The earlier proposal had six groups which was later expanded to seven groups with subcategories, sub-classifications and anatomic levels of anomalies and diagnoses. It has been effective and useful and was recommended by the IFSSH as an appropriate system for use by the surgeons involved in this field.

As the classification was gaining importance, the Japanese Society for the Surgery of the Hand (JSSH) came into the light. With the knowledge gained through elegant experimental studies, especially involving the diagnostic conditions of cleft hand and symbrachydactyly, classified under failure of formation of parts and undergrowth respectively, Miura (1978), proposed that abnormalities of the embryo at the stage when the digits are separated or when the interdigital space is formed were the grounds of basis for the typical cleft hand, syndactyly and central polydactyly<sup>14</sup>. When the failure of separation is limited, the middle finger develops into two parts, each fusing to the adjacent digits followed by hyper-regression of the embryonic central interdigital tissue, resulting in the typical cleft hand. He concluded that these conditions should not be classified as failure of differentiation of parts but as failure of separation of parts. Ogino, in 1986, tested the validity and practicality of Swanson's classification and classified 955 congenital upper limb anomalies. He noted that leaving the syndactyly associated with constriction band syndrome, which is caused after the formation of digital rays, all the syndactyly without associated anomalies and those associated with polydactyly and typical cleft hands should be considered the same resulting from the failure of formation of digital rays<sup>15</sup>(fig-2). He further added, through his clinical and experimental studies on rats where he induced polydactyly, syndactyly and cleft hand, that clefting will also develop when the osseous syndactyly or polydactyly develops toward the proximal position of the ring finger. He suggested that these anomalies may appear when the same teratogenic factor acts on the embryo at the same developmental period and based on a common teratogenic mechanism- failure of induction of finger rays<sup>16</sup>. Based on these findings, the JSSH modification proposed a new category "Abnormal induction of rays" which included central polydactyly, syndactyly and

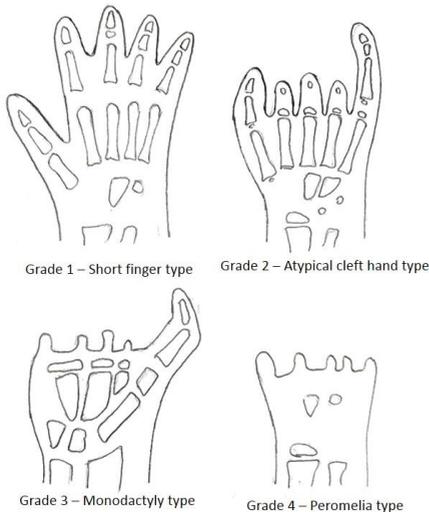
cleft hand that were originally considered by Swanson as duplication, failure of differentiation and failure of formation respectively. This category included those manifestations of mal-segmentations of the digital rays, i.e., the abnormal central finger number induction in the central part of the hand plate due to cleft formation without apoptosis caused by the inactivation of the apical ectodermal ridge<sup>17</sup>. It also incorporated a group of unclassifiable cases that had four anomalies.



**Figure 2:** *Cleft hand formation process from polydactyly.*

Symbrachydactyly, a condition characterized by shortening of the middle phalanges of the central three digits to start with, progressing to absence and then increasingly severe deficiency of the adjoining parts of the hand, is the second condition placed in the category of undergrowth was considered to be misplaced in the Swanson's classification. The JSSH committee believed that symbrachydactyly is synonymous with transverse failure of formation. Miura, in 1994, noted that when the mesenchyme is damaged severely, it resulted in transverse deficiency and if the damage is mild and formation has continued, it resulted in intercalated transverse deficiency<sup>18</sup>. Based on these observations, the sequence of deformities included in

brachysyndactyly or atypical cleft hand, appear to be morphological variants of transverse deficiency (*fig-3*). Henceforth, the condition was included under “transverse deficiency” as manifestations of failure of formation of parts.



**Figure 3:** Classification of symbrachydactyly according to Blauth.

Tonkin recognized that problems arise when attempting to incorporate our current understanding of causation into a morphology-based classification. He pointed out the shortcomings with the JSSH classification like not including the radial and ulnar polydactyly which are morphologically akin central polydactyly in the abnormal induction of rays group and symbrachydactyly which has a component of syndactyly remain in transverse failure of formation. Based on gene mapping studies, he proposed that, complex interactions of gene abnormalities would decide whether clefting may, or may not, occur with syndactyly or polydactyly rather than a teratologic sequence. He felt that rather than moving around things within the classification, a major reassessment would benefit the lot.

He re-wrote the IFSSH classification in a descriptive manner based upon appearance alone and does not attempt to explain the cause which would allow a universal language of communication, collection of data, assessment of associations and avoids losing data when the clinical presentation is unable to be placed in an appropriate single classification group (Annexure -1).

### ***The Time to change***

As our knowledge progressed with studies detailing the insult to the embryo at molecular level, various authors felt that the shortcomings of the Swanson's classification could be overcome. They considered that the classification partly implies insight into the pathogenesis, the groups of "duplication", "overgrowth" and "constriction band syndrome" are simply based on the appearance. For these reasons and those mentioned earlier by Tonkin, the authors of the OMT Classification believe that the Swanson Classification does not embrace the expansion in knowledge which has occurred within the last 50 years and that attempts to modify it simply create contradictions which become illogical.

### ***The Arrival and growth***

Researchers were using the descriptive classification for a period till the OMT classification arrived. It was based on the dysmorphology framework which included:

Group 1: Malformations, an abnormal formation of tissue resulting from abnormal cell formation.

Group 2: Deformations, differ from malformations, as the insult is to cells which have already formed normally. It is a deformation of normal tissue.

Group 3: Dysplasias, a lack of normal organization of cells into tissue.

Group 4: Disruption, involving damage to tissues which have already formed.

The authors of OMT classification, considering the pathological process, included disruptions along with deformations. Malformations were subgrouped according to the part involved, either the whole limb or the hand plate alone. These were further divided based on the axis of involvement, one of the four- proximal-distal, radial-ulnar, dorsal-ventral and unspecified axis. The signalling centres for each of these axes are well known and any disruption of one signalling centre or pathway will have consequences within other signalling centres and pathways, both upstream and downstream. For example, thumb hypoplasia is malformation of the hand plate in radial-ulnar axis whereas radial deficiency is malformation of the whole limb in radial-ulnar axis. The subgroup unspecified axis caters those conditions for which the dominant axis of involvement is not defined though involvement of multiple axes is seen like syndactyly, camptodactyly, synostosis and symphalangism. Deformation includes constriction ring syndrome and trigger digit, though it is considered as developmental rather than congenital. Dysplasia include those conditions which involve limb hypertrophy and conditions that were earlier classified based on the appearance<sup>19</sup>.

In February 2014, after extensive discussions, the IFSSH Scientific Committee on Congenital conditions recommended the adoption of

the OMT classification with reviews at 3 years intervals<sup>13</sup>. The committee found the system to be flexible and had the ability to respond to new developments and additional conditions. Bae et al investigated the reliability of OMT classification and found that it exhibits substantial to almost perfect intraobserver and interobserver reliability among paediatric hand surgeons at different institutions<sup>20</sup>. They also pointed out the discrepancies in the agreement between observers in conditions like transverse deficiencies and symbrachydactyly. Goldfarb, in 2015 was able to classify all the 653 individuals based on the OMT classification and said that it is an improvement compared to the Swanson system, easy to use and it is intelligently designed in such a way to incorporate the increasing knowledge over time<sup>21</sup>.

### ***Inviting criticisms***

Though the classification received support from multiple reviews, the authors themselves acknowledge the difficulties in classifying certain conditions like symbrachydactyly and arthrogryposis. The authors claim that these conditions, the cause of which are not fully understood and any classification of conditions which are of unknown cause will be classified in a less than satisfactory manner<sup>22</sup>. Ekblom (2013) accepted that OMT classification is a needed and appropriate replacement for the Swanson's classification but did not fail to highlight the dispute involving the placement of conditions like complex syndactyly, symbrachydactyly and general hypoplasia of the limb<sup>23</sup>. He concluded that these disputes reflect the incomplete knowledge of the cause of the underlying condition rather than the inconsistency of the Swanson's or OMT systems.

The harsher criticism came from Lowry and Bedard, who stated that just because some of the molecular and signalling pathways involved in limb development are known does not mean that they can be incorporated into the classification system<sup>24</sup>. They strongly recommended the use of International Classification of Diseases (ICD) system and the Mendelian Inheritance in Man (MIM) to help with etiological knowledge. They also questioned the utility of the system as they found it to be devoid of prevalence data. They argue that the classification is unlikely to assist collaboration with other disciplines especially groups involved with surveillance and routine acquisition of data internationally and concluded that the OMT system falls short in providing a practical and easily utilized classification scheme. Tonkin replying to these criticisms raised, states that the OMT system could not provide prevalence data as ICD does but the latter was developed for gaining information for administrators rather than being helpful for surgeons<sup>25</sup>. He adds that the system is expandable without destroying its core structure, amenable for modification as and when the chromosomal or gene defect is known for a given condition.

## ***Conclusion***

It is vitally important that a uniform classification be adapted throughout the world in order to facilitate the monitoring of the congenital malformations, to permit comparisons of incidence between the different areas, and to assist in the research regarding possible etiological factors, prevention, and selection of most efficacious methods of management. These are mandatory for the establishment of national registries for collection of baseline data and for establishment and coordination studies carried on by individuals and institutions. Given, the wider range of manifestations of the

anomalies, any classification is expected to fall short of reaching the 'perfection'. However, at present the OMT classification appears to be a usable one. It has a scope of modifications in future as our knowledge of the cause and the molecular basis improves over time. Though the classification looks complex but when fragmented and understood well, it is easy to apply and has proved to be reliable. In its present form it would suffice for communication with the clinicians, geneticist, anatomist and more importantly, the parents.

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# **Annexure 1 - Practical description of upper limb anomalies**

## **(Tonkin):**

### **(I) Abnormalities of zeugopod** **(arm)**

- (A) Hypoplasia/ aplasia
  - 1) Shoulder
  - 2) Arm
  - 3) Poland syndrome
- (B) Transverse deficiency
  - 1) Transverse arrest
- (C) Intersegmental deficiency
  - 1) Phocomelia
  - 2) Symbrachydactyly
- (D) Synostosis
- (E) Overgrowth
  - 1) Hemihypertrophy
- (F) Duplication
  - 1) Whole arm
- (G) Anomalies of soft tissue
  - 1) Arthrogryposis
- (H) Complex osseous anomalies
  - 1) Constriction ring syndrome
  - 2) Congenital tumorous conditions

### **(II) Abnormalities of stylopod (forearm)**

- (A) Hypoplasia/ aplasia
  - 1) Radial deficiency
  - 2) Ulnar deficiency
- (B) Transverse deficiency
  - 1) Transverse absence
- (C) Intersegmental deficiency
  - 1) Symbrachydactyly
- (D) Synostosis
  - 1) Elbow synostosis
  - 2) Forearm synostosis
- (E) Overgrowth
  - 1) Hemihypertrophy
- (F) Duplication
  - 1) Ulnar duplication
- (G) Anomalies of soft tissue
  - 1) Arthrogryposis
- (H) Complex osseous anomalies
  - 1) Madelung deformity
- (I) Constriction ring syndrome
- (J) Congenital tumorous conditions

**(III) Abnormalities of autopod**  
**(wrist and hand)**

- (A) Hypoplasia/ aplasia
  - 1) Radial deficiency
  - 2) Ulnar deficiency
  - 3) Brachydactyly
- (B) Transverse deficiency
  - 1) Transverse absence
  - 2) Brachydactyly
- (C) Intersegmental deficiency
  - 1) Symrachydactyly
  - 2) Brachydactyly
- (D) Synostosis
  - 1) Carpal
  - 2) Metacarpal
  - 3) Symphalangism
  - 4) Phalangeal – complex syndactyly
- (E) Overgrowth
  - 1) Macroductyly
- (F) Duplication
  - 1) Radial polydactyly
  - 2) Central polydactyly
  - 3) Ulnar polydactyly
  - 4) Mirror hand
- (G) Anomalies of soft tissue
  - 1) Simple syndactyly
  - 2) Camptodactyly
  - 3) Distal arthrogyposis
  - 4) Clasped thumb
  - 5) Trigger digit
    - 6) Cleft hand complex
- (H) Complex osseous anomalies
  - 1) Complex syndactyly
  - 2) Clinodactyly
  - 3) Triphalangism
  - 4) Cleft hand complex
  - 5) Hyperphalangism
- (I) Constriction ring syndrome
- (J) Congenital tumorous conditions

*Specific anomalies may be placed in more than one section as the classification is merely descriptive.*

# Annexure 2 - OMT (Oberg, Manske, and Tonkin) Classification:

## Group 1 – Malformations

### A. Abnormal axis formation and differentiation – Entire upper limb

#### 1. Proximal distal axis (AER involvement)

- a) Brachymelia with brachydactyly
- b) Symbrachydactyly –
  - Poland syndrome,
  - Whole limb excluding Poland syndrome
- c) Transverse deficiency –
  - Amelia
  - Clavicular/ Scapular
  - Humeral
  - Forearm (common)
  - Wrist (carpal absent at proximal or distal row)
  - Metacarpal
  - Phalangeal (proximal, middle, distal)
- d) Intersegmental deficiencies
  - Proximal (Rhizomelic),
  - Distal (Mesomelic),
  - Total (Phocomelia)
- e) Whole limb duplication/ triplication.

#### 2. Radial-ulnar axis (anteroposterior)

- a) Radial longitudinal deficiency
- b) Ulnar longitudinal deficiency
- c) Ulnar dimelia
- d) Radio ulnar synostosis
- e) Congenital dislocation of radial head
- f) Humeroradial synostosis – Elbow ankylosis
- g) Madelung deformity.

#### 3. Dorsal-ventral axis

- a) Ventral dimelia
  - Nail patella syndrome
  - Furhmann/ Al-Awadi/ Raas-Rothschild syndromes
- b) Absent/ hypoplasia extensor, flexor muscles.

#### 4. Unspecified axis

- a) Shoulder –
  - Sprengel deformity
  - Abnormal muscles
- b) AMC complex (Arthrogryposis Multiplex Congenita)

## **B. Abnormal axis formation/ differentiation – Hand plate.**

### **1. Proximal-distal axis (AER)**

#### **involvement)**

- a) Brachydactyly (no forearm/ arm involvement)
- b) Symbrachydactyly (no forearm/ arm involvement)
- c) Transverse deficiency – wrist, metacarpal, phalanges.

### **2. Radial-ulnar axis (anteroposterior)**

- a) Radial deficiency – Thumb
- b) Ulnar deficiency – no forearm/ arm involvement
- c) Radial polydactyly (pre axial)
- d) Triphalangeal thumb
- e) Ulnar dimelia (mirror hand)
- f) Ulnar polydactyly (post axial)

### **3. Dorsal-ventral axis**

- a) Dorsal dimelia (Palmar nail)
- b) Ventral dimelia (hypoplastic/ absent nail)

### **4. Unspecified axis**

- a) Soft tissue
  - Syndactyly
  - Camptodactyly
  - Thumb in palm deformity
  - Distal arthrogryposis
- b) Skeletal deficiency
  - Clinodactyly
  - Kirner's deformity
  - Synostosis (metacarpal and carpal)
- c) Complex
  - Complex syndactyly
  - Synpolydactyly
  - Cleft hand complex
  - Complicated syndactyly

## **Group 2 – Deformations**

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

## **Group 3 – Dysplasias**

- A. Hypertrophy
  - 1. Whole limb
    - a) Hemihypertrophy
    - b) Aberrant flexor/ extensor/ intrinsic muscles
  - 2. Partial limb
    - a) Macrodactyly
    - b) Aberrant intrinsic muscles
- B. Tumorous conditions
  - 1. Vascular – Hemangiomas
  - 2. Neurological – Neurofibromatosis
  - 3. Connective tissue –
    - Juvenile aponeurotic fibroma,
    - Infantile digital fibroma
  - 4. Skeletal – Osteochondromatosis
    - Enchondromatosis
    - Fibrous dysplasia
    - Epiphyseal abnormalities

## **Group 4 - Syndromes**

- A. Specified
- B. Others – this include unspecified syndromes having congenital hand anomalies