

New Classification of Congenital Malformation of Hand

L. K. Sharma, M.S., M.Ch. (Plastic)

Head of the Dept., Department of Plastic & Maxillo-facial Surgery, Medical College Nagpur

No satisfactory classification of congenital malformation of hand has been established yet. Various attempts have been made to classify the congenital anomalies of hand but they do not describe the observed patterns of deformity. The type of abnormalities of the hand are many and in each type the number of units involved, and the degree of severity vary greatly. This makes it difficult to give a satisfactory classification.

Classification of any disease should be such that it may be placed in a certain group, and each group may clearly define the type and severity of the disease. The terminology used should be simple to pronounce, and when the final diagnosis of the disease made from such classification, it should give clear picture of the type and severity of the condition.

Previous classification of hand anomalies by Mayer and Swanker (1958) Patterson (1959) and Martin, A. Entin do not give any idea of the severity of anomaly when placed in different groups, nor that they are grouped in such a manner which can be used to represent the diagnosis in simple words. There are certain types of deformities which have been completely ignored in their classification. At times the deformity placed in their classification in certain groups gives entirely different

and confused picture. Such as Mayer and Swanker (1958) places Club hand under the group of "Deformities due to Contracture".

I have studied 158 cases of congenital malformation of hand and made an attempt to classify these anomalies in a simple manner specifying the rays affected and the grade of affection without using terms long winded and difficult to define precisely.

In a classical description of the development of hand during in the 3rd week, limb buds arise from a ridge of paraxial mesoderm at either side of the trunk and by 4th week differentiation starts in these buds. In the 6th week the terminal portion of the buds flatten and four radial grooves appear. The rapid growth which occurs in five thick areas separated by these grooves produces the appearance of the fingers as they project beyond the main mass. The axial part of the mesenchyme of the limb bud condenses into its cartilaginous skeleton and the bones of the limbs are formed by their ossification. The mesoderm surrounding the developing skeleton provides the musculature of the limb.

Gegenbaner (1864) believed that it was the suppression of the primitive rays which was responsible for the deformities. He assumes that the upper extremity is

composed of main stem and four accessory rays. The main stem includes Humerus, Ulna, two carpal bones, 5th metacarpal and 3 phalanges of little finger. 2nd, 3rd, 4th ray include index, middle and ring fingers with their carpals and metacarpals. 1st ray (Radial ray) include radius, navicular, trapezium 1st metacarpal and 2 phalanges of the thumb.

Although this hypothesis is difficult to be believed by the anatomist in the present era as they believe more in the defect in the developing genes rather than the theory of evolution, but clinically it is found that most of the times the anomaly extends from the thumb to radius on the radial side little finger to ulna on the ulnar side. While

anomaly in the Index, middle and ring only extends upto the carpal bones. Ulna or Radius are never found involved, when middle rays are malformed. According to these clinical manifestations I have classified the malformation of hand taking each ray as one unit i.e. radial ray, ulnar ray and middle rays, and graded them according to the severity.

Classification of Malformation of Hand

(A) Aplasia or Hypoplasia : (Fig. 1)

Under this group the components involved may be :—

- (1) Whole limb may be under developed (Hypoplasia) or completely absent (aplasia).



Fig. 1—Photograph showing aplasia and hypoplasia of hand.

- (2) Proximal section—arm & forearm.
- (3) Distal section (Hand)—In case of the hand either.
 - (a) all rays may be involved or
 - (b) Particular ray may be involved. This group is further classified as :

- 1. Radial Ray Hypoplasia
Aplasia

- 2. Ulnar ray Hypoplasia
Aplasia
- 3. Middle rays Hypoplasia
Aplasia

(1) (a) Radial Ray Hypoplasia (Fig. 2 and 3) :

According to the severity of the condition it is further classified into various grades.

Grade 1 : Hypoplastic thumb with

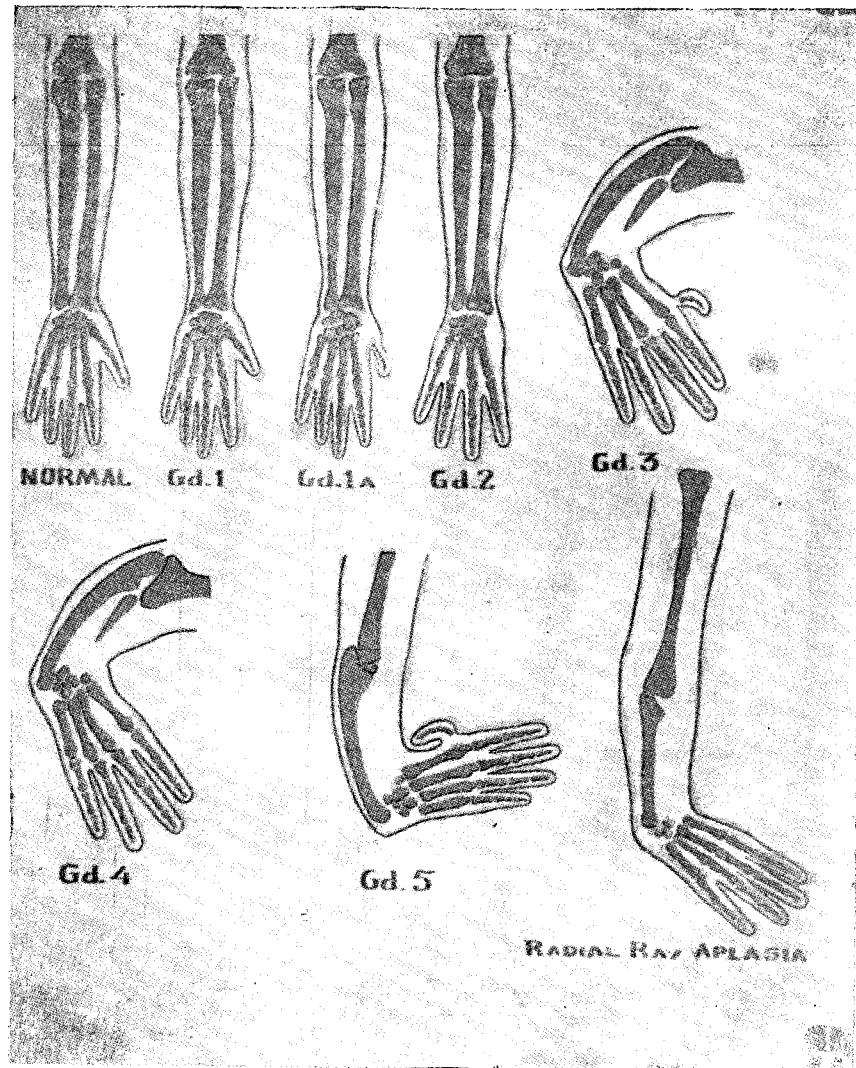


Fig. 2—Diagrammatic illustration showing various grades of radial ray hypoplasia

- all normal bones and joints.
- Grade 1 (a) : Hypoplastic thumb with rudimentary bones and no proper joint (Flail thumb)
- Grade 2 : Thumb absent radius normal.
- „ 3 : Thumb hypoplastic with partial absence of radius.
- „ 4 : Absent thumb with partial absence of radius.
- „ 5 : Hypoplastic thumb with complete absence of thumb.

- Grade 3 : Hypoplastic little finger with partial absence of ulna.
- „ 4 : Little finger with partial absence of ulna.
- „ 5 : Hypoplastic little finger with complete absence of ulna.

(b) Ulnar ray Aplasia.

(B) Syndactyly : (Fig. 4, 5 & 6)—The fusion of the digits may be associated with hypoplastic hand (Fig. 6) or it may

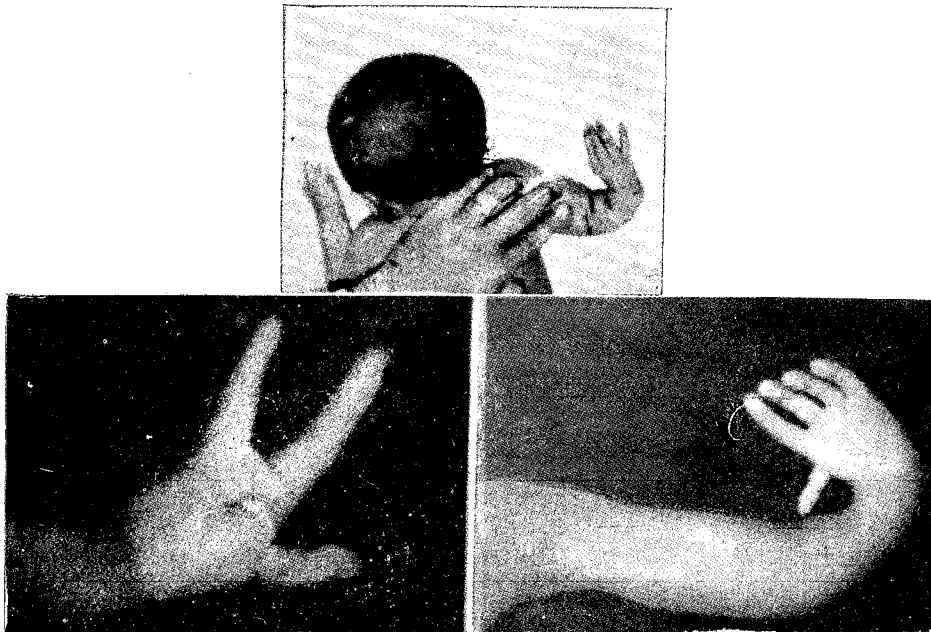


Fig. 3—Photographs showing radial ray and middle ray hypoplasia

(b) Radial ray Aplasia.

- 2 (a) Ulnar ray Hypoplasia :
- Grade 1 : Hypoplastic little finger with all bones and joints.
- 1 (a) : Hypoplastic little finger with rudimentary bones and no proper joint.
- „ 2 : Little finger absent with normal ulna.

be present in otherwise normal hand (Fig. 5). The fusion may extend according to the severity to different levels, from proximal to the distal end of the digit. Thus they are grouped as (Fig. 4). :—

- Grade 1 : Fusion upto middle joint.
- „ 2 : Fusion upto distal joint.
- „ 3 : Fusion beyond distal joint.

Grade 4 : Fusion upto tip including nails and bones.

(C) Duplication : (Fig. 7, 8, 9) : In this group the extra digit may be partially separated from the main or it may form a complete independent unit. The condition is graded according to the severity as (Fig. 7, 8) :—

- Grade 1 : Blob of soft tissue only.
- 1 (a) : Blob of soft tissue with bony element but without proper joint.
- Grade 2 : Duplication from tip to just short of distal joint.
- 2 (a) : Tip to distal joint.
- Grade 3 : Duplication from tip to just short of middle joint.

- 3 (a) : Duplication from tip to middle joint.
- Grade 4 : Duplication from tip to proximal joint. In case of Ulnar digit.

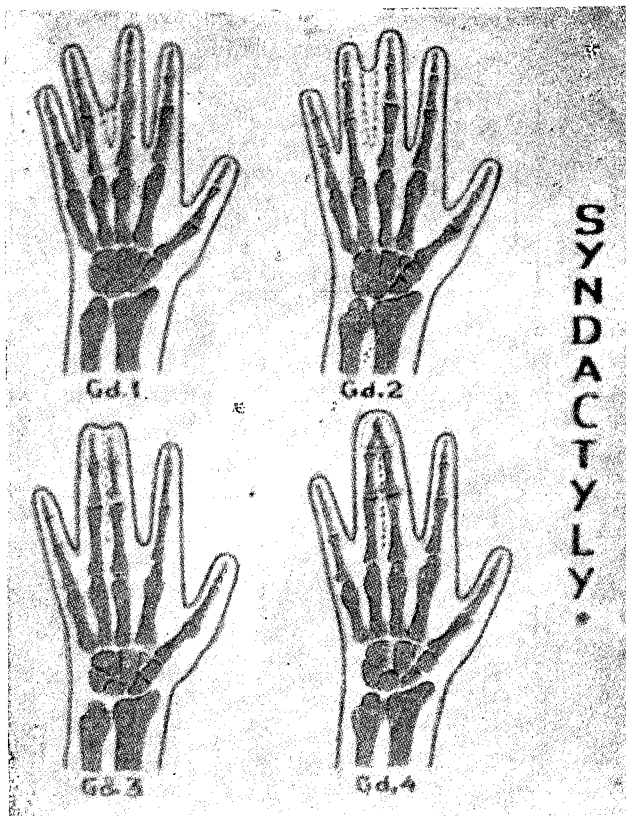


Fig. 4—Diagrammatic illustration of various grades of syndactyly



Fig. 5—Photograph showing syndactyly in a normal hand

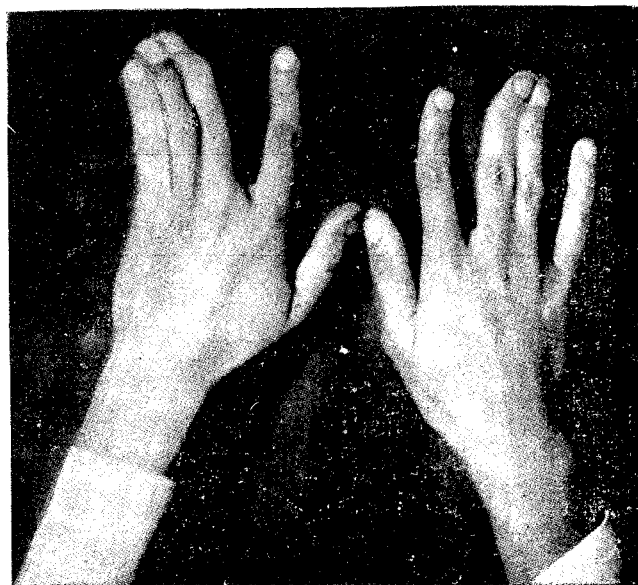


Fig. 6—Photograph showing syndactyly in a hypoplastic hand

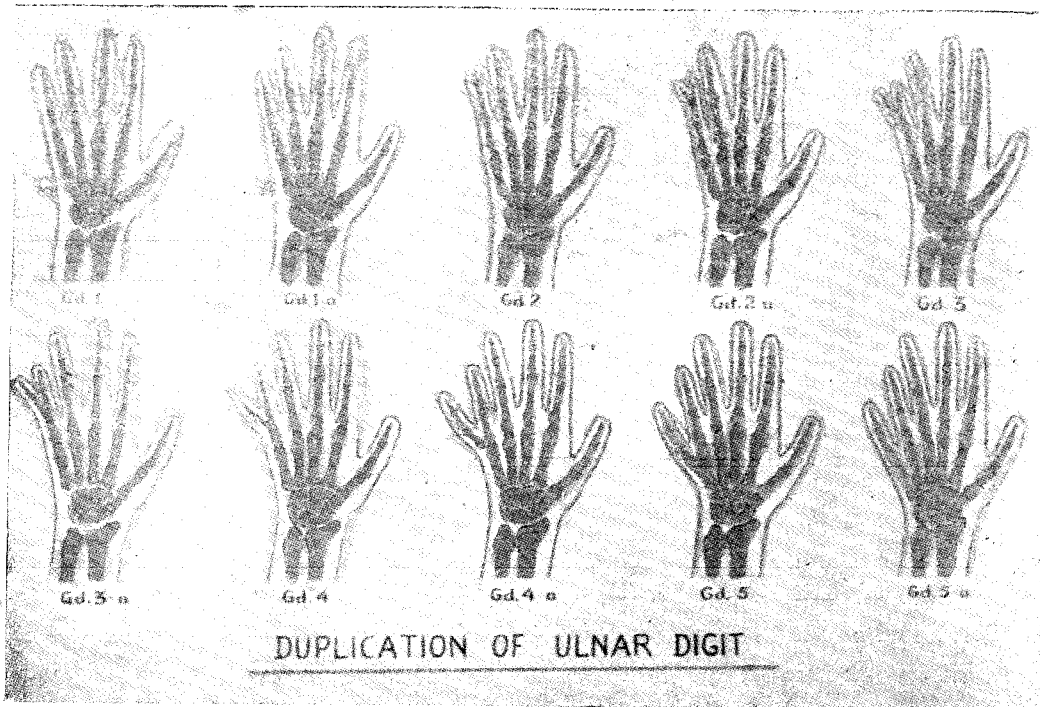


Fig. 7—Diagrammatic illustrations showing various grades of duplication of radial digit

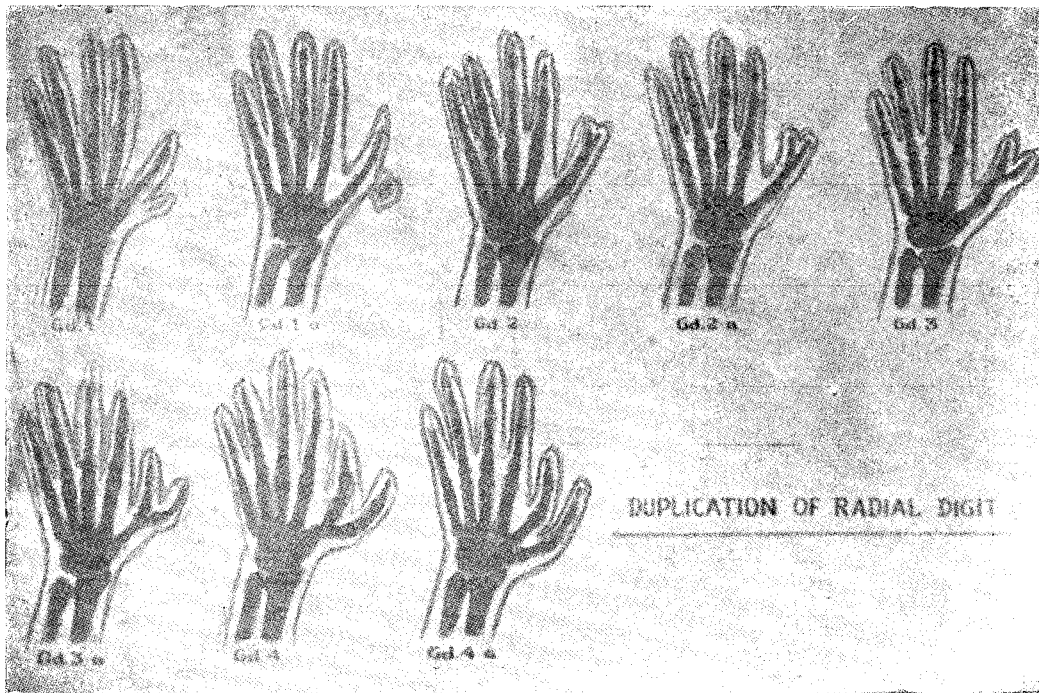


Fig. 8—Showing duplication of ulnar digit

Grade 5 : Duplication of metacarpal (Bifurcation only).

- 5 (a) : Duplication of complete ray
- (D) Twinning of the Hand (Mirror Hand)
- (E) Index Like Thumb : (Fig. 10) :

This is the deformity where there is poor differentiation of the thumb and thenar muscles. It resembles the Index ray having three phalanges and a metacarpal with its epiphysia situated distally. Such a thumb has poor opposition to the other digits.

(F) Deformities due ot Mesodermal De-ficiency : In this group the following conditions may be present :—

- 1. Constriction Ring :
- Grade 1 Shallow groove (skin and

subcutaneous tissue).

- Grade 2 Deep groove involving muscles.
- Grade 3 Deep groove involving the bone without instability.
- Grade 4 Deep groove with local absence of bone and flail distal segment.

2. Abnormal Alignment of the Joint:

- 1. Articular deviation.
 - (a) Lateral deviation.
 - (b) Antero-posterior deviation.
- 2. Soft tissue contractures (flexion contracture).
- 3. Vascular Malformations :
 - (a) Arterial aneurism (Cirroid)
 - (b) Cavernous haemangioma.
 - (c) Capillary haemangioma.
 - (d) Lymphangioma.

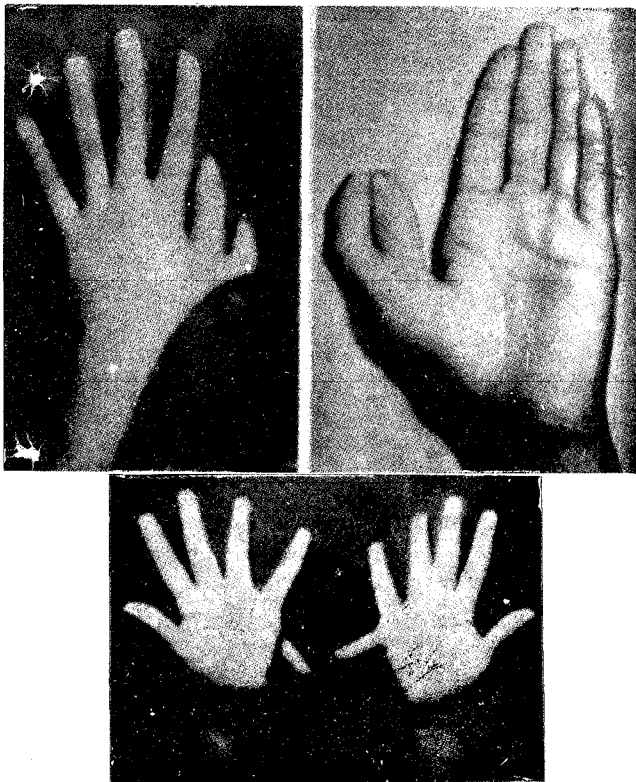


Fig. 9—Photograph showing duplication of radial and ulnar digit

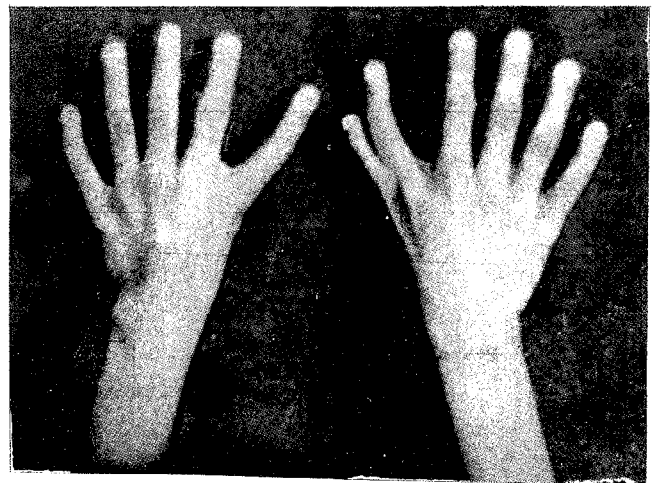


Fig. 10—Photograph showing index like thumb. Thumb is replaced by a digit which is in the same axis as other fingers. There is poor differentiation of thenar muscles. Pinch action is absent.



Fig 11—Photograph showing a tumour of hand in a child aged 5 months. The tumour was present at birth. Histologically it was proved as fibroma

(G) Macrodactyly : The condition may be due to overgrowth of fibro-fatty tissue or neuro fibromatous tissue.

(H) Miscellaneous Group of Anomalies :

1. Fibroma (Fig. 11).
2. Myxoma.

The classification presented above helps in placing various deformities of the hand in a broad group which is graded according to the severity of the condition. It helps identify the deformity rather than a guide for the line of treatment.

Summary :

158 patients of various malformation of hands have been studied and a new classification of congenital malformation of hand is presented. The classification is simple specifying the rays affected and the grade of affection without using terms long winded and difficult to define precisely. Literature on the subject is reviewed.

Acknowledgement :

I am indebted to Prof. C. Balakrishnan, F.R.C.S., who has guided me in completing the above classification.

REFERENCES

1. Mayer & Swanker : Anomaly of Infants & Children, 1958, 357.
2. Patterson, T.J.S. ; Ann. of R.C.S. Vol. 25, 306, 1959.