

Nomenclature for Congenital Skeletal Limb Deficiencies, a Revision of the Frantz and O'Rahilly Classification¹

*Report of the Consultants to the Subcommittee on Child Prosthetics
Problems of the Committee on Prosthetics Research and Development:*

Cameron B. Hall, M.D., Los Angeles, Calif.

Claude N. Lambert, M.D., Chicago, Ill.

Ronan O'Rahilly, M.D., St. Louis, Mo.

Chester A. Swinyard, M.D., Ph.D., New York, N.Y.

Prepared by Robert L. Burtch, M.A.,² Research Scientist, Prosthetic and Orthotic Studies, New York University Post-Graduate Medical School, under the supervision of Sidney Fishman, Ph.D., Project Director, and Hector W. Kay, M.Ed.,³ Associate Project Director, Prosthetic and Orthotic Studies, New York University Post-Graduate Medical School.

At the request of the Subcommittee on Child Prosthetics Problems of the Committee on Prosthetics Research and Development, Child Prosthetic Studies, New York University, initiated a study of congenital skeletal limb deficiencies during March 1963 (2). The primary purpose of this initial effort was to determine the adequacy of the classification nomenclature for congenital skeletal limb deficiencies proposed by Drs. Charles H. Frantz and Ronan O'Rahilly (4) and of a description-classification form developed by NYU Child Prosthetic Studies. The results of the evaluation (2) indicated that 471 of 577 limbs (85 per

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² Since September 1, 1965, Mr. Burtch has been serving as Coordinator of the Physical Medicine and Rehabilitation Service, Maimonides Hospital—Coney Island Division, Ocean and Shore Parkways, Brooklyn, N.Y. 11235.

³ Since April 1, 1965, Mr. Kay has been serving as Assistant Executive Director, Committee on Prosthetics Research and Development, National Academy of Sciences—National Research Council, 2101 Constitution Ave., N.W., Washington, D. C. 20418.

cent) were classifiable within the framework of the Frantz-O'Rahilly system.

In the light of these generally favorable results, the Subcommittee on Child Prosthetics Problems appointed a group of consultants (Drs. Cameron B. Hall, Claude N. Lambert, Ronan O'Rahilly, and Chester A. Swinyard) to consider possible ways and means by which the Frantz-O'Rahilly plan might be modified to provide an even more comprehensive system for classifying limb deficiencies.

In the course of several joint meetings of the consultants and the NYU staff, a revised system was developed. The revised system generally follows the basic principles proposed by Drs. Frantz and O'Rahilly, in that: *first*, it is based on a description of *absent* skeletal parts; *second*, deficiencies are classified under the two basic headings, Terminal and Intercalary, with subgroups of Transverse and Longitudinal under each of these headings. However, the use of anatomical terms has been extended significantly and is included in the classification of all deficiencies. Thus the use of such clinical descriptive terms as hemimelia, peromelia, ectromelia, phocomelia, dysmelia, etc., has been eliminated. Only two basic de-

scriptive terms are now proposed: Amelia, or *complete* absence of a free limb, and meromelia, or *partial* absence of a free limb. The latter term is a derivative of the Greek *meros* (part or partial) and *melos* (limb).

The use of the revised nomenclature adheres to procedures set forth in the *Standard Nomenclature of Diseases and Operations* (7). The classification of a given deficiency, therefore, proceeds from the general to the specific, citing absent skeletal elements for definitive identification. For example, Meromelia: Terminal Longitudinal; Metacarpal: I, II, III describes a terminal longitudinal deficiency of the hand involving absence of digital rays I, II, and III. To provide a basis for possible international consideration, the anatomical terminology utilized in this system is consistent with *Nomina Anatomica* (3).

Since x-rays and the resulting classification may be expected to change depending on the degree of maturation (for example, tarsals and carpals), cases where ossification is continuing must be reclassified periodically.

The material related to the revised classification system is presented in five parts:

- I. A definition of the terms and symbols employed.
- II. Two charts (II. a. and II. b.) adapted from articles by Dr. Hall *et al.* (5) and Dr. O'Rahilly (6) to facilitate understanding of the basic principles involved.
- III. A detailed, illustrated description of the classification plan.
- IV. A description-classification form used for recording purposes.
- V. Instructions for use of the description-classification form.

I. TERMS AND SYMBOLS

TERMS

| | |
|------------------------|---|
| Amelia | <i>Complete</i> absence of a free limb (exclusive of girdle). |
| Meromelia | <i>Partial</i> absence of a free limb (exclusive of girdle). |
| Terminal Deficiency | Absence of all skeletal elements distal to the proximal limit of the deficiency, along the designated axis (longitudinal or transverse). |
| Intercalary Deficiency | Absence of middle part(s) lying between a proximal-distal series of limb components; elements proximal to and distal to the absent part(s) are present. |
| Transverse | Absence extending across the width of the limb. |

| | |
|--------------|--|
| Longitudinal | Absence extending parallel with the long axis of the limb (forearm and/or hand, or leg and/or foot), either pre-axial, postaxial, or (as in the hand or foot) central in nature. |
| Pre-axial | Absence of the portion of the forearm and/or hand, or leg and/or foot on the thumb or the great-toe side of the limb (radial or tibial portion). |
| Postaxial | Absence of the portion of the forearm and/or hand, or leg and/or foot on the side of the limb opposite the thumb or the great toe (ulnar or fibular portion). |
| Central | Absence of one or more of the intermediate digital rays (for example, Ray III). |
| Rudimentary | A remnant of an osseous element. If the remnant is identifiable (for example, the humerus), the term "rudimentary humerus" would be applicable. If the remnant cannot be identified, the symbol "X" (unknown) would be cited (for example, "rudimentary X"). |
| Ray | A digit. |

SYMBOLS

| | | | |
|----------|-----------------------|-----------------------|--|
| I | Intercalary. | ? | Questionable identity of element cited (for example, radius ?)■ |
| T | Terminal. | | |
| — | Transverse. | | |
| / | Longitudinal | | |
| Pre-Post | Pre-axial. Postaxial. | X | Unknown (unidentifiable). |
| | | :I, II, III, IV, or V | Digital ray(s) involved, starting from pre-axial side to postaxial side of limb. |

SKELETAL ELEMENTS

Capital letters are used to identify skeletal elements that are *completely* absent; small (lower case) letters are used to identify skeletal elements that are *partially* absent. If the word identifying the skeletal element is written out, the first letter of the word is capitalized when the element is completely absent (for example, Humeral), and in lower case when only partially absent (for example, humeral).

| | | | |
|----------|----------|----------|-------------|
| HU or hu | Humeral. | TI or ti | Tibial. |
| U or u | Ulnar. | FI or fi | Fibular. |
| R or r | Radial. | TA or ta | Tarsal. |
| CA or ca | Carpal. | MT or mt | Metatarsal. |

| | | | |
|----------|-------------|----------|----------------------|
| MC or mc | Metacarpal. | PP or pp | Phalanx Proximal. |
| PH or ph | Phalangeal. | PM or pm | Phalanx Middle. |
| FE or fe | Femoral. | PD or pd | Phalanx Distal. |

Skeletal Segments

- P Proximal third of element cited.
- M Middle third of element cited.
- D Distal third of element cited.

The symbols P, M, and D are used to indicate thirds of the skeletal elements cited, which may be completely or partially absent. Utilization of the three symbols requires the following clarification:

Terminal Transverse (T-) Deficiencies

- P absence of *part* of the proximal third of the skeletal element cited *and everything distal to it*.
- M absence of *all or part* of the middle third of the skeletal element cited *and everything distal to it*.
- D absence of *all or part* of the distal third of the skeletal element cited *and everything distal to it*.

Terminal Longitudinal (T/) Deficiencies

- P absence of *part* of the proximal third of the skeletal element cited and everything distal to it *parallel with the same axis*.
- M absence of *all or part* of the middle third of the skeletal element cited and everything distal to it *parallel with the same axis*.
- D absence of *all or part* of the distal third of the skeletal element cited and everything distal to it *parallel with the same axis*.

Intercalary Transverse (I-) Deficiencies and Longitudinal (I/) Deficiencies

- P absence of *all or part* of the proximal third of the skeletal element cited.
- M absence of *all or part* of the middle third of the skeletal element cited.
- D absence of *all or part* of the distal third of the skeletal element cited.

II. a. BASIC SCHEMA FOR CLASSIFICATION OF CONGENITAL SKELETAL LIMB DEFICIENCIES

Figure 1 presents a basic schema for the classification of congenital skeletal limb deficiencies which has been adapted from one originally presented by Dr. Cameron B. Hall *et al.* (5).

II. b. BASIC SCHEMA FOR CLASSIFICATION OF CONGENITAL SKELETAL LIMB DEFICIENCIES

Figure 2 presents a basic schema for the classification of congenital skeletal limb deficiencies which has been adapted from one originally presented by Dr. Ronan O'Rahilly (6).

III. CLASSIFICATION NOMENCLATURE

- A. Terminal Transverse (T—) Deficiencies (Figs. 3 and 4)
Amelia—complete absence of a free limb (exclusive of girdle).
(For example, Amelia: T—; Upper Right.)

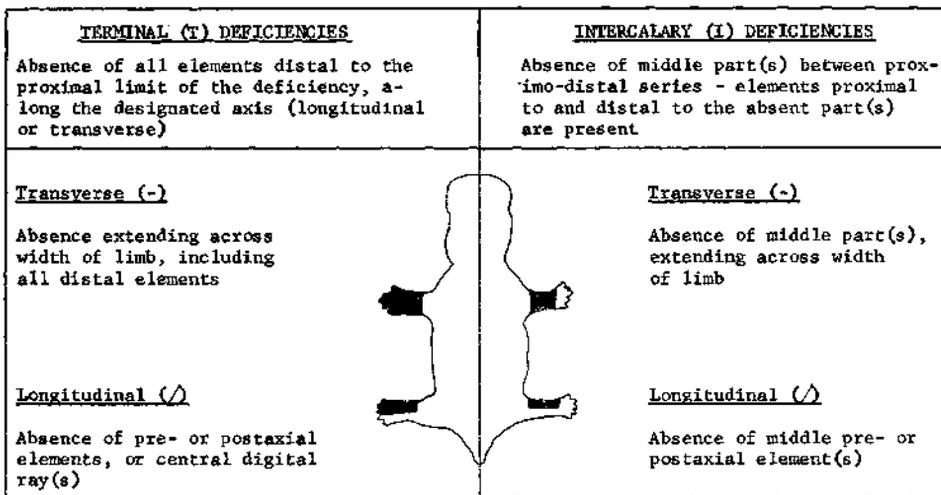


Fig. 1. Basic schema adapted from Dr. Cameron B. Hall *et al.* (5).

MEROMELIATERMINAL (T) DEFICIENCIES

Absence of all elements distal to the proximal limit of the deficiency, along the designated axis (longitudinal or transverse)

INTERCALARY (I) DEFICIENCIES

Absence of middle part(s) between proximo-distal series - elements proximal to and distal to the absent part(s) are present

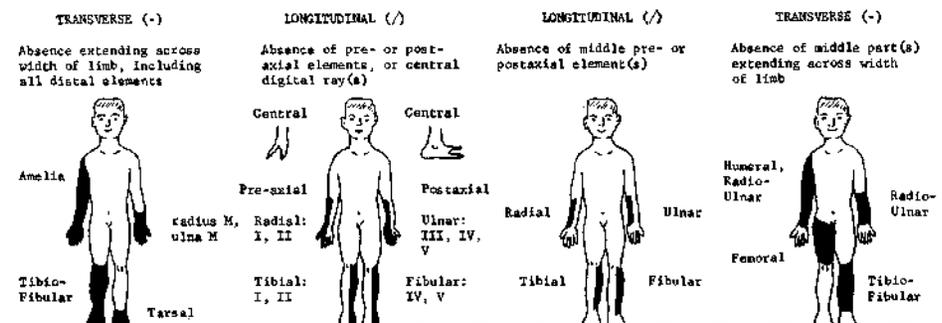


Fig. 2. Basic schema adapted from Dr. Ronan O'Rahilly (6). The term "meromelia," denoting *partial* absence of a free limb, is applicable to all examples in the schema with the exception of the transverse deficiency of the *complete* limb which has been denoted as "amelia."

Meromelia—partial absence of a free limb (exclusive of girdle).

1. Humeral or Femoral (P, M, or D)

Partial absence of the humerus or femur and all distal elements.

(For example, Meromelia: T—; humeral D (distal third above-elbow-type stump).)

2. Radio-Ulnar or Tibio-Fibular

a. Complete absence of the Radius and Ulna or Tibia and Fibula, and all distal elements.

(For example, Meromelia: T—; Radio-Ulnar (elbow-disarticulation-type stump).)

b. Partial absence of the radius and ulna or tibia and fibula, and all distal elements. Use P, M, or D, as appropriate.

(For example, Meromelia: T—; radio-ulnar M (short below-elbow-type stump).)

c. Complete absence of *one* of the forearm or leg elements, and all distal elements.

(For example, Meromelia: T—; Radius (wrist-disarticulation-type stump).)

3. Carpal or Tarsal

Complete absence of all hand or foot elements. (For example, Meromelia: T—; Tarsal (ankle-disarticulation-type stump).)

4. Carpal or Tarsal, Distal

Absence of the distal row of carpals or tarsals, and all other hand or foot elements distal to this level.

(For example, Meromelia: T—; carpal, Distal (mid-carpal-type stump).)

5. Carpal or tarsal, Pre- or Postaxial

Absence of either the pre- or postaxial carpal or tarsal bones, *and all other hand or foot elements*. (For example, Meromelia: T—; carpal, Pre-axial (carpal-metacarpal-type stump).)

6. Metacarpal or Metatarsal

a. Absence of all metacarpals or metatarsals and all hand or foot elements distal to this level. (For example, Meromelia: T—; Metatarsal (tarsal-metatarsal-type stump).)

b. Absence of a portion of metacarpals or metatarsals and all hand or foot elements distal to this level. Use P, M, or D to indicate absent segment(s) of each metacarpal or metatarsal. (For example, Meromelia: T—; metacarpal: I D, II D, III D, IV M, V M (trans-metacarpal-type stump).)

7. Phalangeal

a. Absence of all phalanges from all five digits. (For example, Meromelia: T—; Phalangeal, Upper Right (metacarpo-phalangeal-type stump).)

b. Complete or partial absence of *one or more phalanges from all five digits* (but not all phalanges from all five digits).

(For example, Meromelia: T—; phalangeal, Upper Right: I, II; III PM, IV PM, D; V PD (trans-phalangeal-type stump).)

B. Terminal Longitudinal (T/) Deficiencies (Fig. 5)

1. Major Long Bones

a. Complete absence of one of the forearm or leg elements and of the corresponding portion of the hand or foot. The skeleto-anatomical terms Radial (R), Ulnar (U), Tibial (TI), or Fibular (FI) are used to indicate the absent long bone. In order to provide greater precision, the identifying number of each absent ray is included in the nomenclature.

(For example, Meromelia: T/; Radial: I, II.) If all but one unidentifiable ray or rudimen-

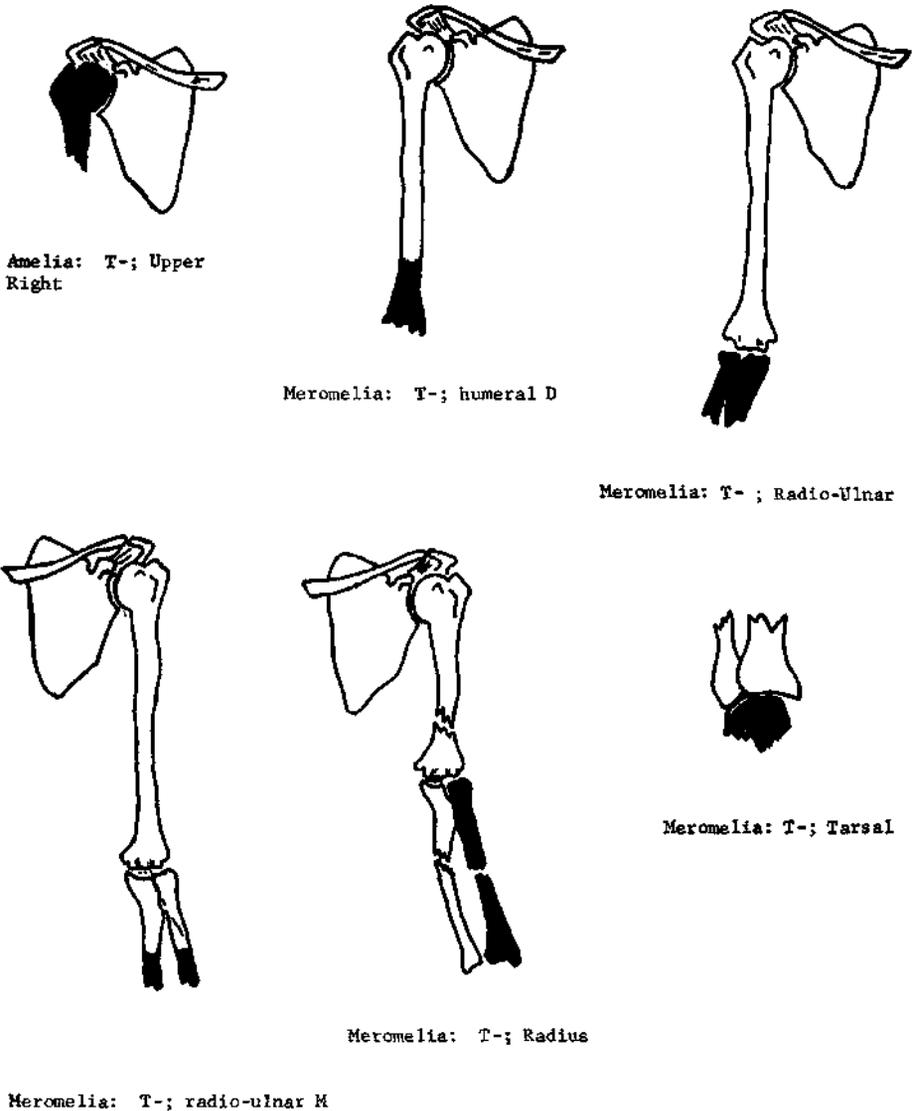


Fig. 3. Terminal transverse (T—) deficiencies. The shaded areas in the example sketches represent absent elements or parts thereof.

- tary ray is absent, the symbol "X" (unknown) or term "rudimentary X" is used.
- b. Partial absence of one of the forearm or leg elements and absence of the corresponding portion of the hand or foot. P, M, or D is used to indicate the absent segment(s) of the long bone. Lower case letters are used, and the absent ray(s) is cited.
(For example, Meromelia: T/; fibular M: IV, V.)
2. Carpal or tarsal, Pre- or Postaxial
Absence of *either* the pre- or postaxial carpal or tarsal bones, *and* corresponding digital rays.
(For example, Meromelia: T/; carpal, Pre-axial: I, II.)
 3. Metacarpal or metatarsal (P, M, or D)
 - a. Absence of all phalanges of one to four digits *and* complete or partial absence of their respective metacarpals or metatarsals.
(For example, Meromelia: T/; metacarpal: I, II, III, V.)
 - b. In the case of partial absence of a specific metacarpal or metatarsal, P, M, or D is used to indicate the absent segment(s).
(For example, Meromelia: T/; metatarsal: I, II; III D; V M.)

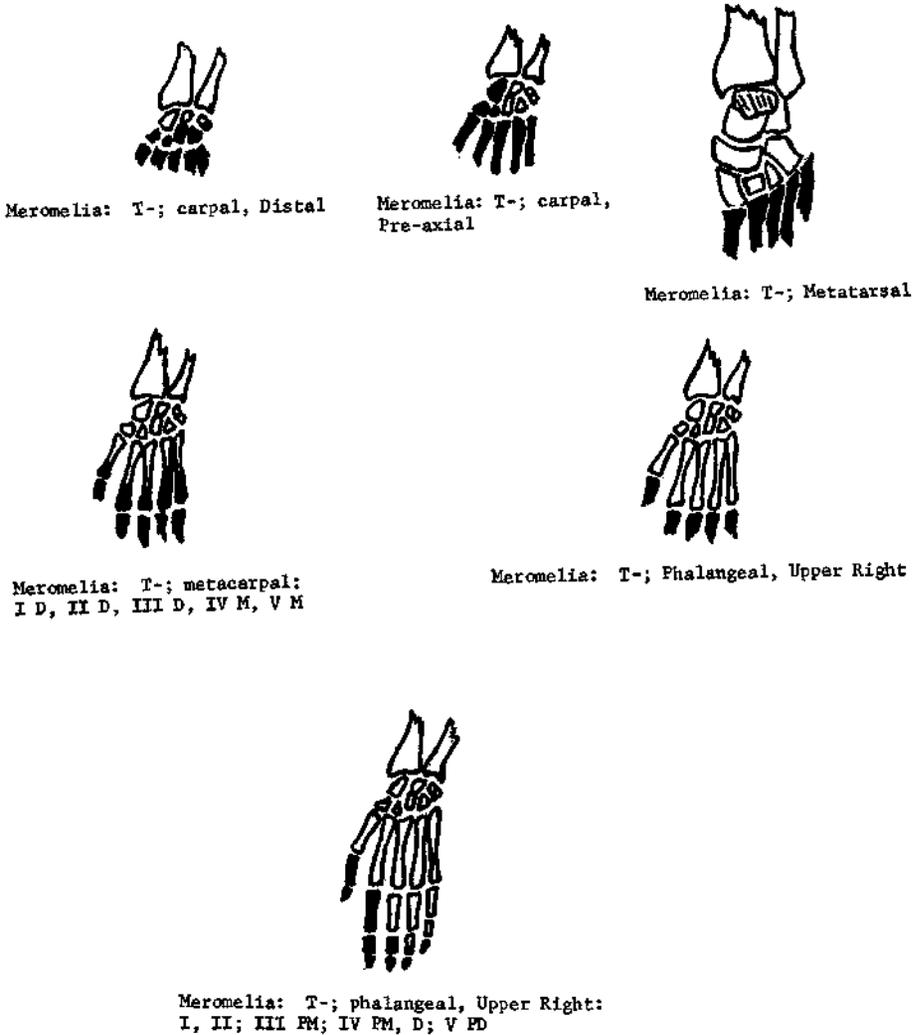


Fig. 4. Terminal transverse (T—) deficiencies (continued). The shaded areas in the example sketches represent absent elements or parts thereof.

4. Phalangeal

Absence of all or part of one or more phalanges from one to four digits.

(For example, Meromelia: T/; phalangeal, Upper Right: I, II, III.)

C. Intercalary Transverse (I—) Deficiencies (Figs. 6 and 7)

A minimum of at least two digital rays (two metacarpals or metatarsals and their associated phalanges) must be present to permit classification as an Intercalary Transverse (I—) deficiency of the major long bones. In such cases, the hand or foot deficiencies (if any) are classified separately. Where there are fewer than two complete digital rays, the deficiency is classified as Terminal Transverse (T—), with a description of the distal digital elements that are absent (for example, "all but one ray absent").

1. Major Long Bones

a. Humeral, Radio-Ulnar; or Femoral, Tibio-Fibular

Complete absence of all three major long bones in the limb with hand or foot elements attached directly to the trunk.

(For example, Meromelia: I—; Humeral, Radio-Ulnar.)

a'. Concomitant hand or foot deficiencies are classified independently of the major long bone deficit.

(For example, Meromelia: I—; Humeral, Radio-Ulnar; plus T/; metacarpal: I, II, V.)

b. Humeral or Femoral

Complete or partial absence of the long bone cited.

(For example, Meromelia: I—; Humeral.)

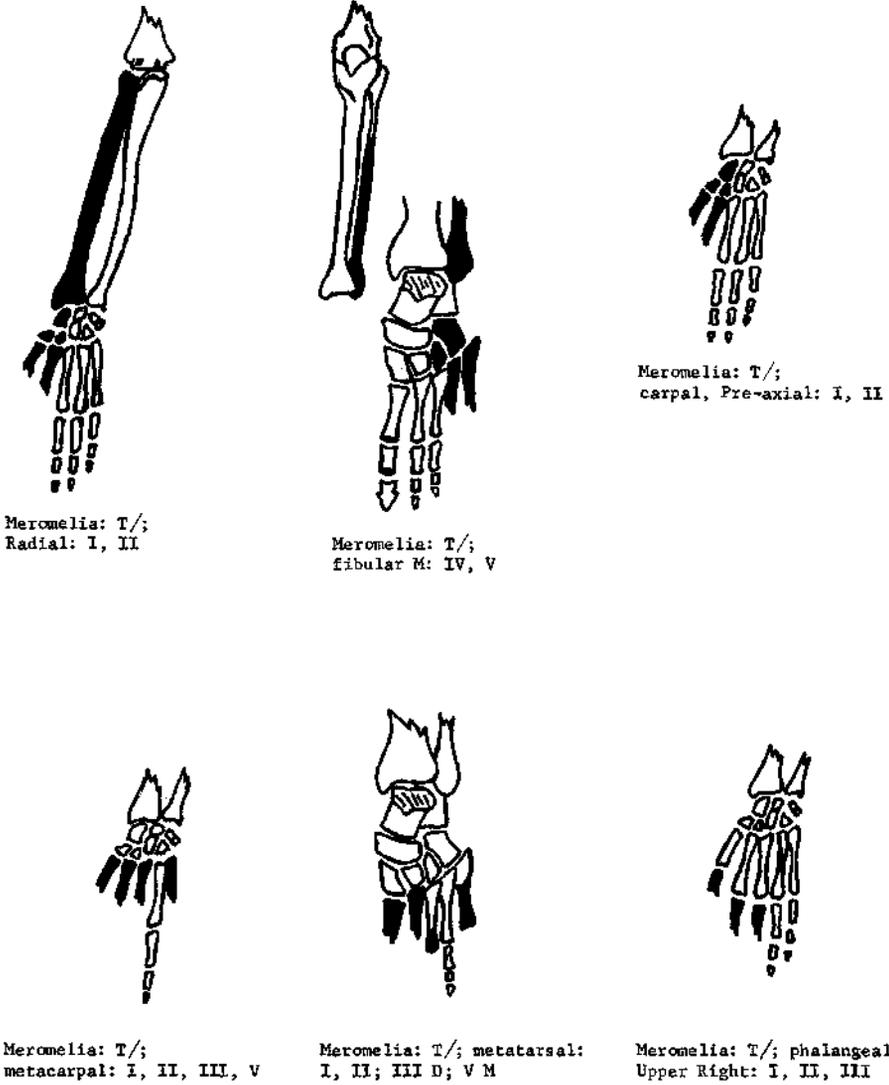


Fig. 5. Terminal longitudinal (T/) deficiencies. The shaded areas in the example sketches represent absent elements or parts thereof.

- b'. If a partial absence exists, P, M, or D is added to indicate the absent segment(s) of the bone cited.
(For example, Meromelia: I—; humeral M, D.)
- c. Radio-Ulnar or Tibio-Fibular
Complete or partial absence of the long bone cited.
(For example, Meromelia: I—; Radio-Ulnar.)
- c'. If a partial absence exists, P, M, or D is used to indicate the absent segment(s) of each bone.
(For example, Meromelia: I—; tibio-fibular P, M.)
- d. Humeral, radio-ulnar; or femoral, tibio-fibular
Partial absence of *all three major long bones* in the upper or lower limb. P, M, or D is used to indicate the absent segment (s) of each long bone.
(For example, Meromelia: I—; humeral D; radio-ulnar M, D.)
- 2. Carpal or Tarsal
Complete absence of the carpal or tarsal bones, with proximal and distal skeletal elements present.
(For example, Meromelia: I—; Carpal.)

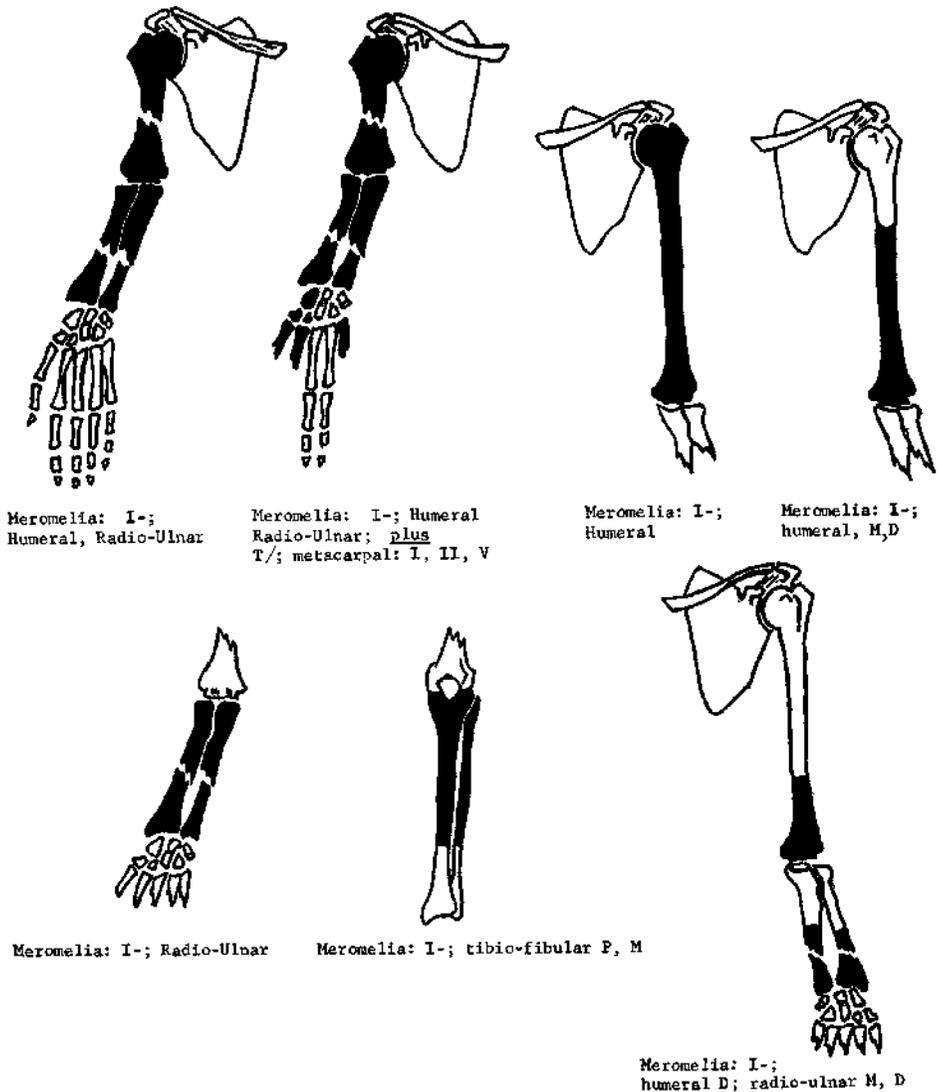


Fig. 6. Intercalary transverse (I-) deficiencies. The shaded areas in the example sketches represent absent elements or parts thereof.

3. Metacarpal or Metatarsal
Complete absence of the metacarpals or metatarsals, with proximal and distal skeletal elements present.
(For example, Meromelia: I-; Metacarpal.)
4. Phalangeal
Absence of all or part of the proximal and/or middle phalanx from all *five* digits.
(For example, Meromelia: I-; phalangeal, Lower Right: I PP; II PP; III PM; IV PM; V PP.)
- D. Intercalary Longitudinal (I/) Deficiencies (Fig. 8)
 1. Major Long Bones
 - a. Complete absence of one of the forearm (R or U) or leg (TI or FI) elements with hand or foot elements intact along the same axis as the deficient long bone.
(For example, Meromelia: I/; Fibular.)
 - b. Similar to above except that *only part* of the long bone cited is absent. P, M, or D is used to indicate the absent segment (s).
(For example, Meromelia: I/; radial P, M.)
 2. Carpal or tarsal, Pre- or Postaxial
Absence of *either* the pre- or postaxial carpal or tarsal bones with all other hand or foot elements present.
(For example, Meromelia: I/; tarsal, Pre-axial.)



Meromelia: I-; Carpal



Meromelia: I-; Metacarpal



Meromelia: I-; phalangeal, Lower Right:
I PP; II PP; III PM; IV PM; V PP

Fig. 7. Intercalary transverse (I—) deficiencies (continued). The shaded areas in the example sketches represent absent elements or parts thereof.

3. Metacarpal or metatarsal

Absence of *all or part* of one to four metacarpals or metatarsals.

(For example, Meromelia: I/; metatarsal: I, II.)

If only part of a metacarpal or metatarsal is absent, P, M, or D is used to indicate the absent segment(s) of the involved ray.

(For example, Meromelia: I/; metatarsal: I D; II M, D.)

4. Phalangeal

Absence of *all or part* of the proximal and/or middle phalanx of from one to four digits.

(For example, Meromelia: I/; phalangeal, Upper Left: I PP; II PM; IV PP.)

IV. DESCRIPTION-CLASSIFICATION FORM

Figure 9 presents the description-classification form developed by NYU Child Prosthetic Studies for recording congenital skeletal limb deficiencies.

V. CLASSIFICATION OF CONGENITAL SKELETAL LIMB DEFICIENCIES

The following instructions were developed by NYU Child Prosthetic Studies to accompany the description-classification form:

1. Fill in the identification items at the top of the page.

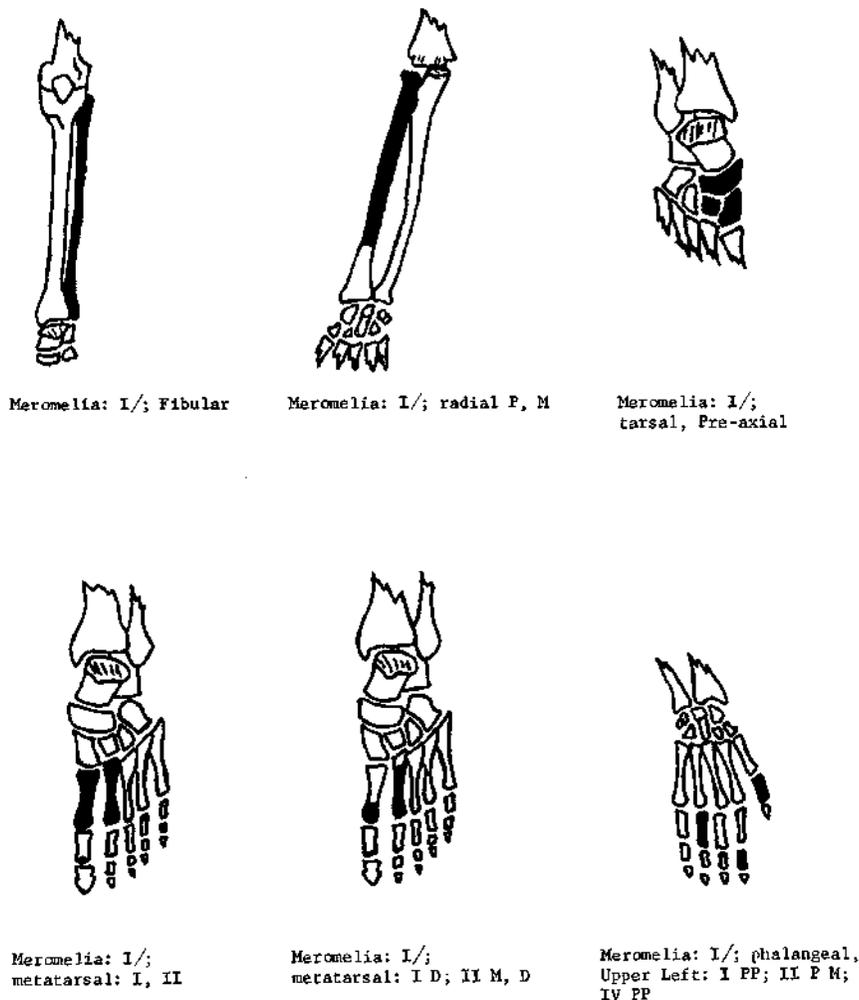


Fig. 8. Intercalary longitudinal (I/) deficiencies. The shaded areas in the example sketches represent absent elements or parts thereof.

2. Indicate in the space provided the presence or history of congenital visceral, soft-tissue or skeletal anomalies other than those of the limbs; that is, cardiac, pulmonary, gastrointestinal (esophageal and/or duodenal atresia, imperforated anus, etc.); genito-urinary, for example, cryptorchidism; cleft palate, hare lip, congenital and/or structural scoliosis, spina bifida, etc.

3. Using a *black* pencil or pen, shade in all *absent* skeletal elements or parts of elements. If an anomaly has been converted to an amputation, describe and classify the *original* anomaly. Care should be taken to retain the approximate length and girth proportions when shading in partial elements. Using a *red* pencil or pen, also indicate on the appropriate limb the approximate *site* and *date* of the surgical conversion (s).

4. In cases where prosthetic restoration is appropriate, indicate the analogous functional level of amputa-

tion for prosthetic purposes (for example, short above-elbow, short below-elbow, long above-knee, etc.) in the space provided. Consult *Upper and Lower Extremity Manual(s)* for functional amputation levels.

5. Indicate next to the appropriate skeletal part on the diagram any of the following conditions that exist. Also, include any unlisted conditions present, as well as any additional information that will enhance the completeness of the description.

| | |
|--------------|------------------------|
| Synostosis | Contracture |
| Hypoplasia | Pseudoarthrosis |
| Bifurcation | |
| Valgus | Dislocation |
| Varus | Subluxation |
| Syndactylism | Supernumerary digit(s) |
| Torsion | Soft-tissue nubbin(s) |

6. After completing the description of each affected

limb, insert in the appropriate space the appropriate classification nomenclature.

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