

CLASSIFICATION OF CONGENITAL LONGITUDINAL DEFICIENCIES

T. OGINO, M. TAKAHARA, N. KIKUCHI, K. ITOH, T. WATANABE, Y. KATO

Department of Orthopaedic Surgery, School of Medicine, Yamagata University

SUMMARY

Purpose: The purpose of this study is to propose a new classification for congenital longitudinal deficiency. **Methods:** Clinical cases and experimentally induced congenital limb anomalies by using busulfan were analyzed. **Results:** In radial and ulnar deficiencies, abnormalities occur in the hand, forearm and/or the elbow. The cause of missing digits in longitudinal deficiency was closely related to deficit of mesenchymal cells in the limb bud, which is caused by the impairment before the limb bud formation. On the other hand, central deficiency, which is defined as typical cleft hand, central polydactyly and osseous syndactyly seemed to be induced when the same teratogenic factors acted on the embryos in the developmental period of digital radiation in the hand plate. **Conclusions:** Longitudinal deficiency is classified into radial and ulnar deficiencies. However, cleft hand is different from radial and ulnar deficiencies. Cleft hand, central polydactyly and osseous syndactyly should be classified into the same entity that is abnormal induction of digital rays. *Riv Chir Mano* 2006; 2: 84-85

KEY WORDS

Cleft hand, central deficiency, radial deficiency, ulnar deficiency

TERATOGENIC MECHANISMS OF LONGITUDINAL DEFICIENCY

Congenital absence of digits confined to the long axis of the upper limb is called longitudinal deficiencies including radial and ulnar deficiencies. In animal experiments, the dead mesenchymal cells were distributed evenly inside the limb bud during formation of longitudinal deficiency. The cause of missing digits in longitudinal deficiency was closely related to deficit of mesenchymal cells in the limb bud, which is caused by the impairment before the limb bud formation (1-3).

Cleft hand is defined as central deficiency and it is closely related to central polydactyly and syndactyly, but different from radial and ulnar defi-

ciencies (4). When analyzing radiographs of the clinical cases, in osseous syndactyly between the middle and ring fingers and in polydactyly of the middle finger, if the development of osseous syndactyly occurs in the proximal direction, then it will develop toward the cleft hand. The cleft hand, central polydactyly and syndactyly, are caused by abnormal induction of the digital rays in the developmental period of digital radiation in the hand plate (5). A single cause affecting the limb bud in a certain receptive period of the development can induce these deformities. Therefore, central polydactyly, osseous syndactyly, and cleft hand may be grouped together and included in the same category of abnormal induction of digital rays (6).

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Based on clinical and experimental studies, the classification of congenital hand deformities proposed by IFSSH has been modified by Ogino et al (7). This classification has been updated and was adopted by the Japanese Society for Surgery of the Hand (8). In modified IFSSH classification, polydactyly, syndactyly and typical cleft hand are included in the same category of abnormal induction of digital rays.

Radial deficiencies

In radial deficiency, the dysplasia of the thumb was classified in 6 types including five fingered hand(non opposable triphalangeal thumb) and 5 degrees of hypoplastic thumb by Blauth (1967). On the other hand, dysplasia of the radius is classified into three types, such as: total absence of the radius, partial absence of the radius, and hypoplasia of the radius. As for elbow deformities, there are contracture or ankylosis of the elbow, radial head dislocation and radioulnar synostosis. In radial deficiency, deformity of the hand, that of the forearm and that of the elbow deformity appear in various combinations. Therefore, deformities in this category should be expressed with combination among hand, forearm and elbow deformities.

Ulnar deficiencies

In ulnar deficiency, hand deformities are classified into 5 types, such as the hypoplasia of the little finger, absence of the little finger, absence of the 2 digital rays of the ulnar side, absence of the 3 digital rays of the ulnar side, and absence of the four digital rays of the ulnar side. Dysplasia of the ulna is classified into 3 types such as, total absence, partial absence, and hypoplasia of the ulna. And the deformities of the elbow are classified into humeroradial synostosis, radial head dislocation, and flexion contracture of the elbow joint. In ulnar deficiencies, deformities should be expressed with combination among hand, ulna and elbow deformities as in radial deficiency.

Abnormal induction of digital rays

Manifestations of malsegmentation of the digital rays in the hand plate due to abnormal induction of digital rays are included in this category. Cleft of the palm means deep V shaped excessive interdigital space. It appears as an isolated anomaly but mostly it is associated with absence of central finger rays. When the index finger seems to be missing, triphalangeal thumb is associated. Clinodactyly and camptodactyly are sometimes associated with cleft hand but should not be classified into other categories, because they seem to be a secondary change due to abnormal induction of the digital rays. In this category, hand deformity can be expressed with combination of cutaneous manifestations, such as syndactyly and cleft of the palm, and skeletal manifestations, such as osseous syndactyly, central polydactyly, triphalangeal thumb, and absence of the central finger rays. Most of the hand deformities belong to this category can be expressed with these combinations.

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