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THE CARPUS IN CONGENITAL ANOMALIES OF THE HAND

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Congenital anomalies of the carpus associated with congenital hand and upper limb anomalies were investigated from the point of view of the development of the hand. Defects of the carpus were categorized as follows: 1) Radial ray defect: loss of the scaphoid, trapezium and trapezoid. 2) Central ray defect: loss of the capitate and loss of a part of the trapezium and hamate. 3) Ulnar ray defect: loss of the triquetrum, pisiformis and hamate. Deficiencies of the carpus and more distal parts of the hand proved to be secondary to defects of the forearm bones in both the radial and ulnar ray. On the other hand, deficiency of the carpus was secondary to defects of the phalanx and metacarpal of the middle finger in the central ray. Thus, from the standpoint of the development of the carpus, bipolar development of the bones of the hand originates proximally in both the radial and ulnar rays and distally in the central ray.

Key words: carpal bones; congenital anomaly; radial, central, ulnar rays

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Congenital anomalies of the carpal bones are rare. Isolated carpal anomalies such as fusion, absence, separation and accessory bone have however been documented (Hammond 1947, Rushforth 1949, Waugh & Sullivan 1950, Cockshott 1966, Roche 1967, Szaboky 1969, Garn et al. 1971, Srivastava & Kochhar 1972). On the other hand, carpal bone anomalies are frequently observed in association with other congenital hand and limb anomalies, thus indicating the relationship of carpal bone development to the development of the whole limb (O'Rahilly 1953, Poznanski & Holt 1971, Kelikian 1974). However, the development of the carpus, in relation to the development of the whole limb or the hand, has not been clearly documented. In what way deficiency of the carpus is related to limb deficiency, or correlates with finger deficiency has not been elucidated.

Limb anomalies have been classified in various ways. Frantz & O'Rahilly (1961) divided limb defects into two groups, viz., terminal and intercalary defects. Entin et al. (1966) and Swanson (1966) outlined the original classification based on embryological considerations in the committee report to the American Society for Surgery of the Hand. Kay and associates (Kay et al. 1973), a working group of the International Society for Prosthetics and Orthotics proposed an international terminology for the classification of congenital limb deficiencies,

according to the longitudinal and transverse deficiency based upon embryological considerations.

In the present paper an attempt will be made to classify anomalies of the carpus in relation to the precursor defect of each anomaly in order to establish the developmental relationship between the carpus, the whole limb and the hand.

PATIENTS

Among the Japanese patients with congenital anomalies of the hand, who attended the Hand Clinic at Osaka University Hospital during the past 16 years, 19 cases of radial ray defects, 8 cases of ulnar ray defects and 30 cases of cleft hands were investigated (Table 1). Of these cases, 19 were sufficiently developed to permit X-ray evaluation.

Table 1. Fifty-seven cases with congenital anomalies of the hand.

	Total cases	Mature cases
Radial ray defect	19	12
Cleft hand	30	5
Ulnar ray defect	8	2

FINDINGS

Radial ray defect

Riordan (1955) divided congenital absence of the radius into two main groups, i.e., total absence and partial absence and described the associated carpal bone anomalies. Heikel (1959) also reported three types of radial ray defects combined with carpal bone anomalies. According to the extent of the longitudinal defect of the radius, we divided the radial ray defect into four groups:

1) total radius defect (very severe)
2) partial radius defect (severe)
3) hypoplastic radius (mild)
4) normal radius (minimal)

Carpal bone anomalies in each group were investigated. Ten cases were bilateral and nine unilateral. There were eleven males and eight females. Carpal anomalies occur in various ways depending on the grade of radial ray defect. The most severe type shows total radius defect with or without absence of the thumb and index finger (Figure 1). In this type, the scaphoid, trapezium and trapezoid bones are generally absent, but the other carpal bones are always intact. Occasionally, the central carpal bones are fused.

The next group shows a partial radius defect associated with a total defect of the scaphoid and trapezium (Figure 2). In these cases, floating thumb, hypoplastic thumb or other severe anomalies of the thumb with a defect of the first metacarpal are usually present.

The third group presents a hypoplastic trapezium and/or defect of the first metacarpal (Figure 3). Eaves & Campiche (1922) reported a similar case.

In the fourth group there is a normal radius with a hypoplastic thumb. This anomaly is considered to be the most benign and presents only a hypoplasia of the scaphoid and trapezium (Figure 4). Hanley & Conlon (1957) described the benign form of the radial defect. Thus, the carpal bone anomaly appearing in each grade of radial ray defect ranges from very severe to minimal. It varies in extent and severity depending upon the radial ray defect (Figure 5). The development of the radial carpal bones, which include the trapezium, trapezoid and scaphoid is apparently closely related to the development of the radius. The development of the thumb and first metacarpal appears to be related to the development of more the distal parts.

Ulnar ray defect

Straub (1965) reported five cases of ulnar defect and Ogden et al. (1976) eleven cases. An analysis of the carpal

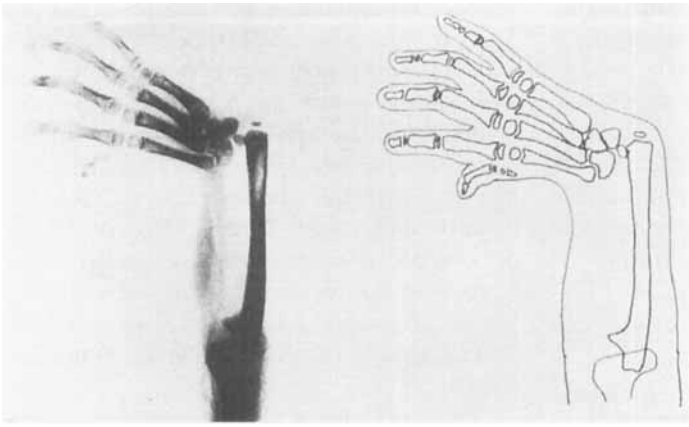

Figure 1. Roentgenogram of the right hand of an 8-year-old girl showing total radius defect, floating thumb and a most severe anomaly of the carpal bones, i.e., defect of the trapezium and scaphoid and fusion between the capitate and trapezoid.

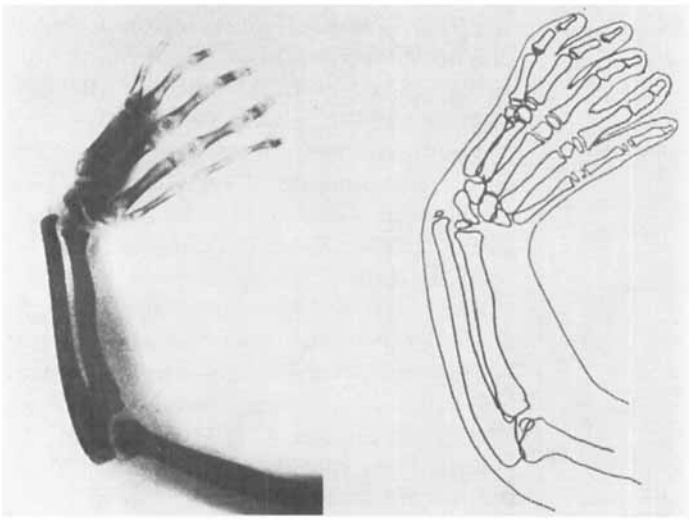

Figure 2. Nine-year-old girl. Note the partial defect of the radius, five fingered hand and severe anomaly of the carpal bones, i.e., total defect of the scaphoid and trapezium and fusion between the lunate and triquetrum.

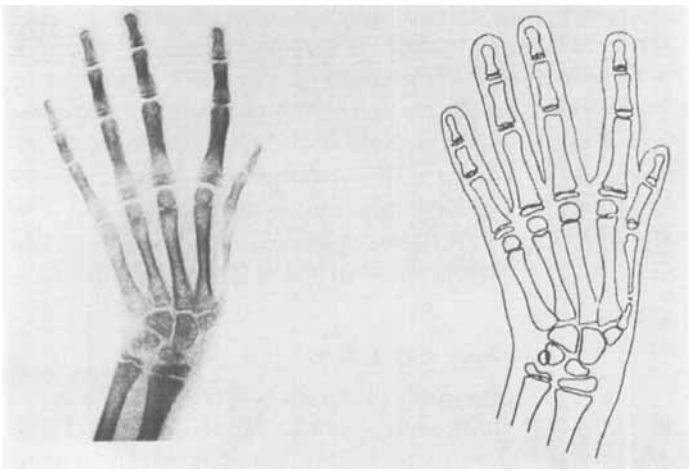

Figure 3. Ten-year-old girl. Note the hypoplastic radius, partial defect of the first metacarpal, defect of both the scaphoid and trapezoid, and hypoplastic trapezium.

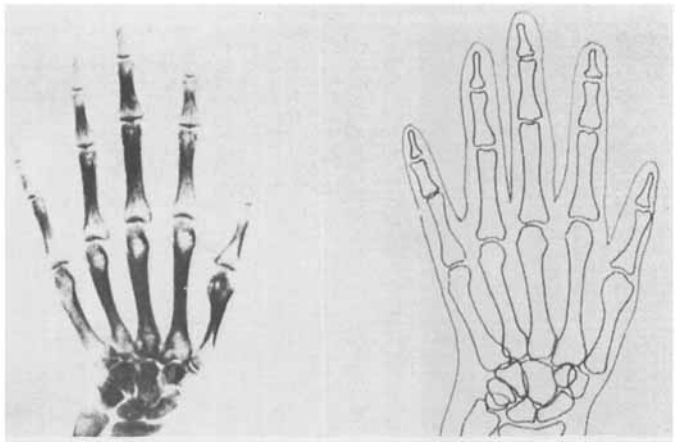

Figure 4. Twenty-three-year-old female, complained of hypoplasia of the left thumb and thenar muscles. Note the hypoplasia of the scaphoid and trapezium also.

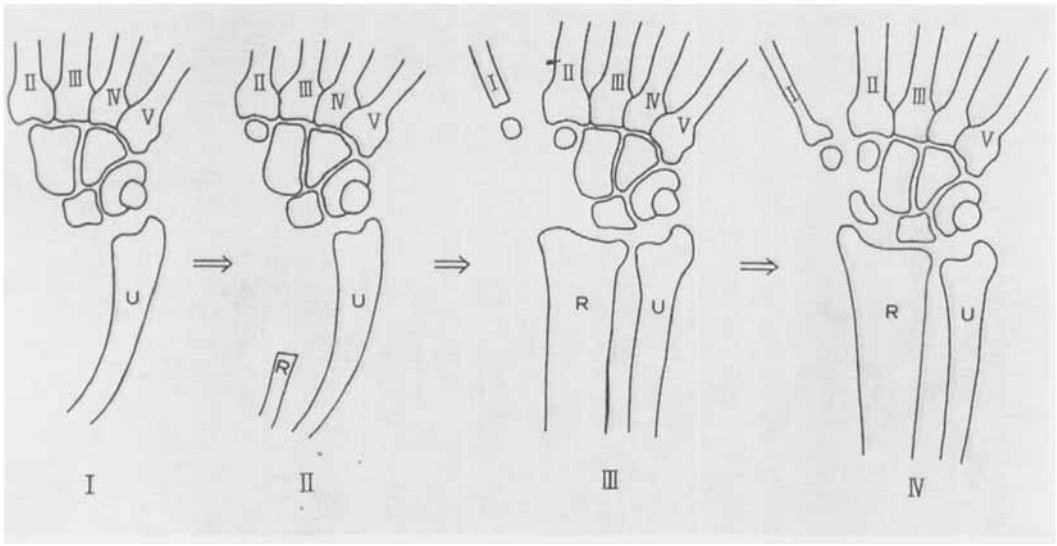

Figure 5. Illustration of the grades of severity of the radial ray defect. Abnormality of the radial carpal bones is also classified into four grades according to the severity of the defect of the radius itself.

bone anomaly, however, was not included in their reports. In the present work we studied eight cases in which carpal bone anomalies were combined with ulnar ray defects. The ulnar ray defect is classified according to the longitudinal defect of the ulna but is a less common anomaly than the radial ray defect. This defect involves the ulna, carpal bones and the two ulnar fingers, and ranges from total ulnar defect to mere hypoplasia of the little finger alone. The triquetrum, pisiformis and hamate are absent, hypoplastic or fused depending on the grade of ulna defect. In our series, eight cases were registered but only two cases were mature enough to be classified as partial defect of the fifth metacarpal associated with carpal anomalies. In one of these two cases there was fusion of capitate-hamate and hypoplasia of triquetrum and pisiformis (Figure 6). The severity of this case resembles that seen in the third group of patients with a radial ray

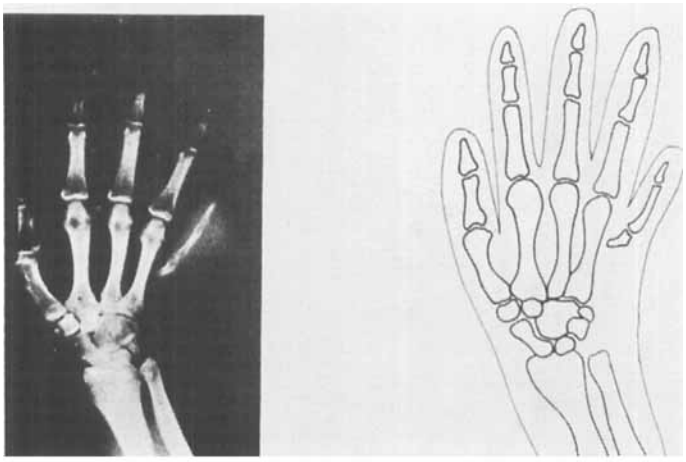

Figure 6. Seventeen-year-old girl with hypoplasia of the ulna, partial defect of the fifth metacarpal and carpal bone anomalies.

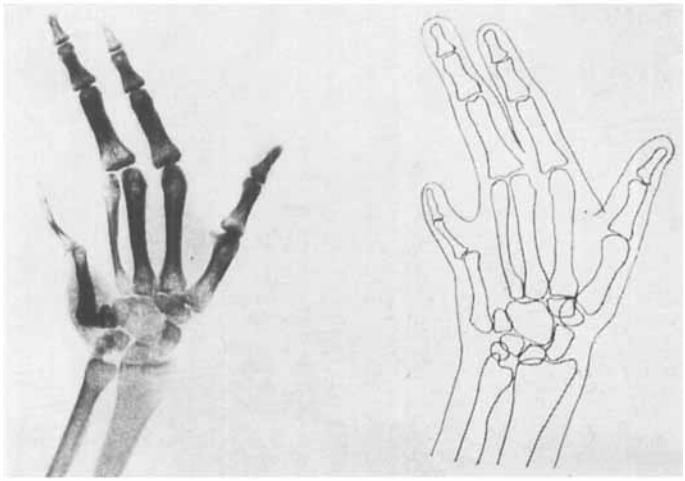

Figure 7. Eighteen-year-old girl with anomalies similar to the carpal bone anomalies of the case in Figure 6. Ossal syndactyly of middle and ring fingers is also evident.

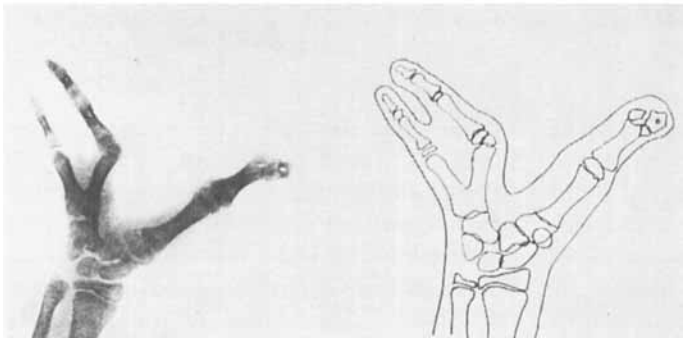

Figure 8. Eleven-year-old girl with defects of the second and third metacarpals and the capitate.

defect. In the second case, a hypoplastic fifth metacarpal with a normal ulna was associated with fusion of the capitate hamate and hypoplasia of triquetrum and pisiformis (Figure 7). Because osseous syndactyly of the middle and ring fingers

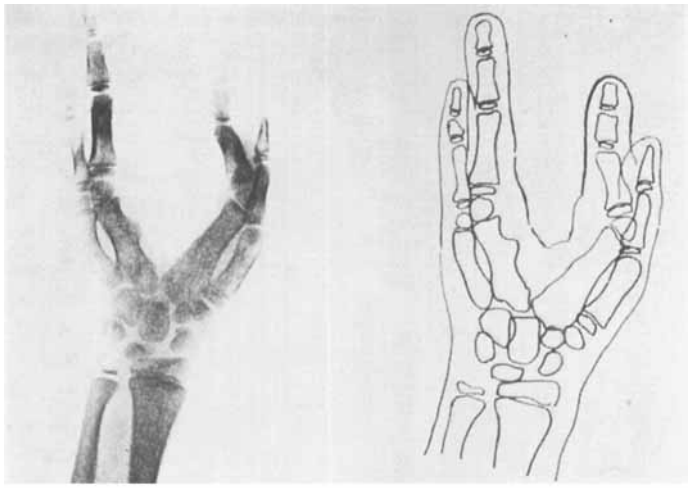

Figure 9. Roentgenogram of a 10-year-old boy with total defect of the third metacarpal and a fairly normal capitate.

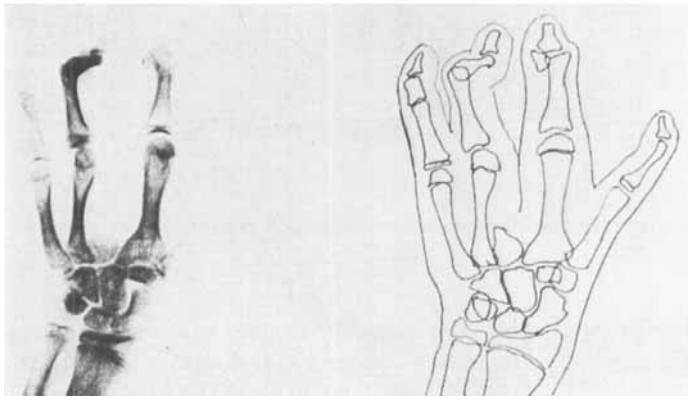

Figure 10. Fifteen-year-old boy with a partial defect of the third metacarpal and normal carpal bone.

was also present this does not appear to be a typical case of ulnar ray defect. Thus the development of the ulnar carpal bones in the ulnar defect is much the same as that in the radial ray defect.

Central ray defect: cleft hand

Thirty cases in which carpal bone anomalies were combined with a central ray defect were studied.

Cleft hand includes various types of anomalies and classification has been made by many authors. Barsky (1964) divided these into the typical and atypical types. The atypical types included other anomalies such as symbrachydactyly, congenital amputation or microdactyly. In the present study, 30 cases were considered typical cleft hand. Eight hands were sufficiently mature for evaluation and were classified according to the extent and grade of the central ray defect. Various degrees of carpal bone defect were also noted. The defects were graded and divided into three groups. The most severe group shows a total defect of the metacarpal and possibly neighbouring metacarpals (Figure 8). The carpal anomaly in this case includes total defect of the capitate and partial defect of the hamate and trapezium. By contrast the other carpal bones are fairly intact.

In the next group, only a third of the

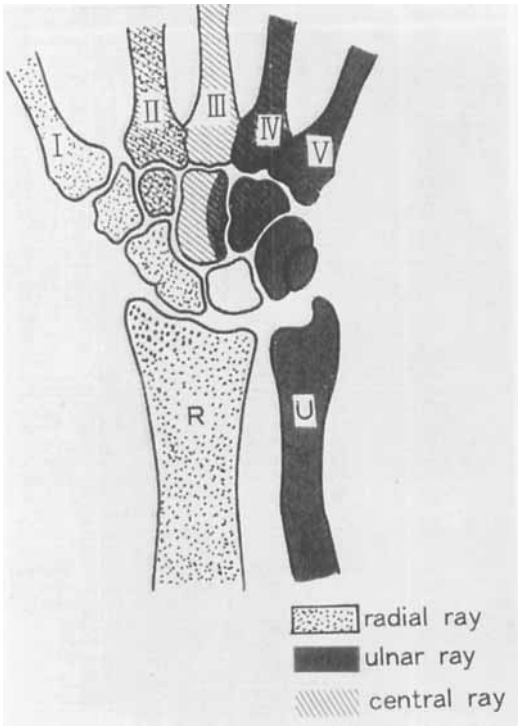

Figure 11. The three rays to which each element belongs.

metacarpal and the middle finger are absent with the other finger rays intact (Figure 9). Partial defect or hypoplasia of the capitate is sometimes observed. but in other cases it is normal. In the most benign group, a partial defect of the third metacarpal and defect of the middle finger are observed. The carpal bones are always normal in this type (Figure 10).

DISCUSSION AND CONCLUSION

In 1924 Kato postulated that the upper extremity consisted of a main stem and four accessory rays. Since his report the terms "radial ray" and "ulnar ray" have been used and applied for purposes of morphological description of limb deficiency, such as radial hemimelia or ulnar hemimelia (O'Rahilly 1951). The present concept is based on limb development with the forearm and hand consisting of three rays, i.e., radial, ulnar and central. However, exactly where each carpal bone should be placed appropriately with reference to these rays has not been defined, nor has the precursor defect of each carpal bone deficiency.

Based on the clinicoradiological study of 57 hand anomalies the following conclusions were drawn:

From the point of view of development the hand consists of three rays, viz., radial, central and ulnar. Each carpal bone is also considered to belong to one of these rays as clearly indicated in Figure 11.

The development of the radial ray division of the carpal bones was found to be related to that of the radius, based on the analysis of four types of radial ray defects in our series.

The ulnar ray was confirmed to include the ulna, triquetrum, pisiformis, hamate, a part of the capitate, the fourth and fifth metacarpals and the two ulnar fingers. The development of this ulnar division of carpal bones was also found to be related to that of the ulna in the same manner as seen in the radial ray. The central ray was confirmed to consist of the capitate, a part of the trapezoid, a part of the hamate, three central carpals and the three central fingers. In comparison with the other two divisions, carpal bones of this central ray division show a quite different mode of development. Here the development precedes from the distal part of the hand, as determined from various carpal bone anomalies in the cleft hand. An anomaly of the lunate was not detected in any patient with a central ray defect.

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