

Analysis of Causes for Congenital Ulnar Deviated Thumbs at the Distal Phalanx Level in 157 Thumbs

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Purpose To investigate anatomic abnormalities causing a congenital ulnarly deviated thumb at the distal phalanx.

Methods A total of 122 children with 157 congenital ulnarly deviated thumbs at the distal phalanges were reviewed, including those with isolated deformity or polydactyly. We analyzed the incidence and characteristics of the underlying anatomic abnormalities as well as the differential diagnoses.

Results Three main causes of an ulnarly deviated thumb were observed. Abnormal hypertrophic epiphyses were found in 96 thumbs. An extra phalanx lying between the normal proximal and distal phalanges was found in 59 thumbs. A previously undescribed cause was found in 2 thumbs with Wassell IV polydactyly, in which an obliquely angled articular surface of the proximal phalanx manifested with ulnar deviation at the interphalangeal joint. Radiographic analysis showed that in cases with abnormal epiphyses, the epiphysis was in good apposition and good alignment with its relevant distal phalanx; the distance from the abnormal epiphysis to the phalanx was usually less than 1 mm. In contrast, in cases of extra phalanges, the distance from the epiphysis to the phalanx averaged more than 2 mm and there was poor apposition between the distal phalanx and the extra bone.

Conclusions Abnormal hypertrophic epiphysis and triphalangeal thumb are the 2 main causes of a congenital ulnarly deviated thumb. A distance of more than 2 mm between the abnormal bone and the distal phalanx suggests a triphalangeal thumb. (*J Hand Surg Am.* 2019;44(10):860–867. Copyright © 2019 by the American Society for Surgery of the Hand. All rights reserved.)

Type of study/level of evidence Therapeutic IV.

Key words Abnormal epiphysis, thumb deformity, triphalangeal thumb, ulnar deviated thumb.

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CONGENITAL ULNARLY DEVIATED thumbs at the level of the distal phalanx are uncommon. In some patients, these are accompanied by other deformities, the most common of which is thumb polydactyly or a triphalangeal thumb.^{1–4} Another published association is an abnormal epiphysis of the distal phalanx.^{5–7} It is difficult to make a correct anatomical diagnosis and to select the appropriate surgical management because there are no apparent differences between these 2 types, either in clinical appearance or radiographically, before the secondary ossification center of the distal phalanx appears.

The primary aim of this study was to investigate causes of a congenital ulnarly deviated thumb. A secondary objective was to perform radiographic measurements to enable a preoperative diagnosis of the underlying cause of the ulnar deviation.

PATIENTS AND METHODS

Patients

We obtained approval for this study from the Institutional Review Board of the Children's Hospital of Chongqing Medical University. Informed consent was obtained from the parents of each child. From January, 2011 to December, 2017, we identified 122 children with a congenital ulnarly deviated thumb at the interphalangeal (IP) joint level, who had surgery in our hospital. Of these 122 children, 35 had bilateral involvement. Median age of the children at surgery was 16 months (range, 3–152 months). The series included 75 boys and 47 girls. Eighteen children had a family history of congenital ulnarly angled thumb at the distal phalanx.

Criteria for patient inclusion were (1) isolated deformity with ulnarly deviated distal phalanx; and (2) deformity accompanied by thumb polydactyly, with the ulnarly deviated distal phalanx occurring on the ulnar thumb. This thumb would be preserved in the surgery, with polydactyly occurring proximal to the IP joint (Fig. 1). Exclusion criteria were (1) deformity accompanied by Wassel type II thumb polydactyly; and (2) deformity accompanied by thumb polydactyly, with the ulnarly deviated distal phalanx occurring on the radial thumb, which is most often excised (Fig. 2).

A total of 157 ulnarly deviated thumbs were included in the study, 48 of which were isolated malformations. The other 109 thumbs were associated with thumb polydactyly. Within these 109 thumbs with polydactyly, 39 thumbs had ulnar deviation at the metacarpophalangeal (MCP) joints and the first web was comparatively narrow. Thirteen of the duplicated thumbs had been resected before our institution evaluated them, which left the ulnarly deviated IP joints untreated. All involved thumbs with polydactyly were smaller to varying degrees than those on the contralateral side, but their nails were of normal shape. Three cases were associated with mild hypoplastic thenar muscles. There was a supination deformity involving the distal phalanx in 5 thumbs.

Methods

Before surgery, we measured the angle of ulnar deviation of the distal phalanx with the IP joint of the

thumb placed in neutral extension. Radiographs were also obtained in this position. On posteroanterior x-ray, we measured the angle between the axis of the proximal and distal phalanges. We measured the distance between the distal phalanx and its corresponding ossification center, whether it was a hypertrophic epiphysis or an extra phalanx (Fig. 3). Finally, we measured the length of both the ulnar and radial sides of the abnormal ossification center. The shape of the abnormal ossification center was recorded. We also recorded the Wassel type and other associated anomalies of the polydactylous thumbs, such as ulnar deviation at the MCP joint. We statistically tested the null hypothesis that variances of the distance between the distal phalanx and the abnormal bone were different between children with abnormal hypertrophic epiphyses and a triphalangeal thumb. The Levene test was employed to assess equality of variances between these groups of patients.

After preoperative assessment, one senior surgeon performed surgical correction. The IP joint was exposed through a dorsoradial approach; in a case of a triphalangeal thumb, both IP joints were exposed. The articular surfaces of the joints were inspected. Visualization of cartilaginous articular surfaces was considered to represent a true joint. Otherwise, this was considered to be a physeal plate. According to these intraoperative findings, the thumbs were divided into 3 groups: (1) hypertrophic epiphysis, (2) extra phalanx, and (3) Wassell IV polydactyly without an extra phalanx or hypertrophic epiphysis but with an unequal length of the radial and ulnar sides of the proximal phalanx. We then performed surgical correction according to the underlying etiology. Based on the Levene test, we found that variances in the distance between the distal phalanx and the abnormal bone were different between children with abnormal hypertrophic epiphyses and a triphalangeal thumb. Hence, Student's *t* test was used to evaluate for a statistically significant difference in this distance between groups.

RESULTS

According to the intraoperative diagnosis, 96 thumbs (61.1%) had a hypertrophic epiphysis (group 1), 59 (37.6%) had an extra phalanx (group 2), and 2 (1.3%) had Wassell IV polydactyly without an extra phalanx or hypertrophic epiphysis but with an unequal length of the radial and ulnar sides of the proximal phalanx (group 3). Mean age of patients in groups 1 to 3 was 23, 28, and 19 months, respectively. After surgery, mean angles of ulnar deviation in groups 1 to 3 were

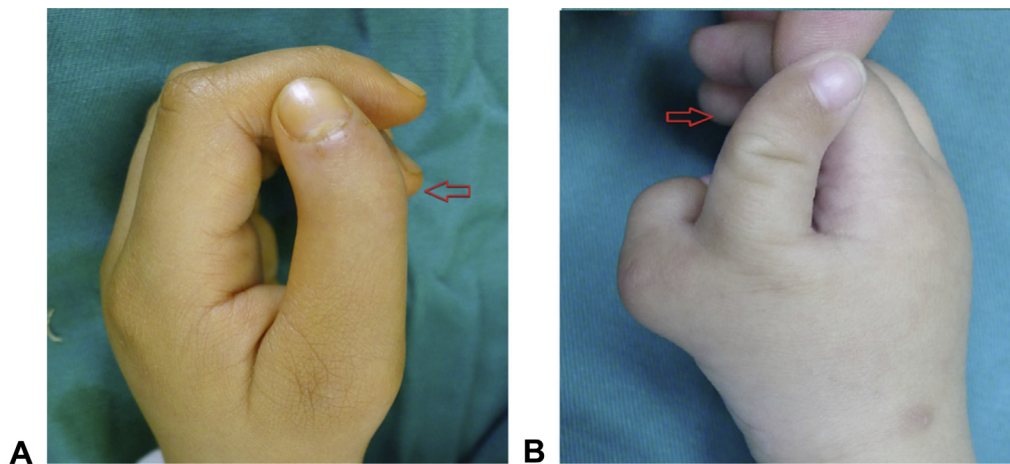


FIGURE 1: Patients included in study: **A** Isolated thumb deformity with an ulnar angulated distal phalanx. **B** Polydactyly with ulnar deviation of preserved ulnar digit.

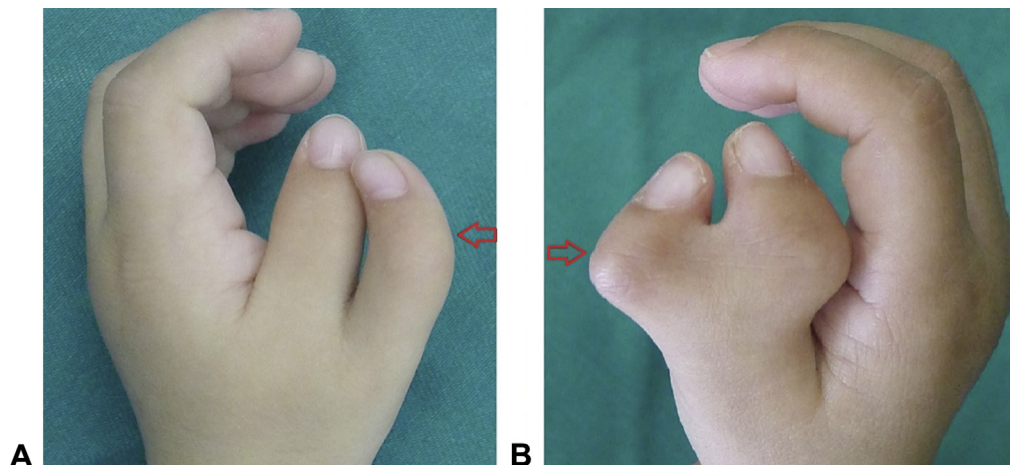


FIGURE 2: Patients excluded from study, with ulnar deviation of the radial thumb duplicate, which was excised during reconstruction. Arrows indicate the radial thumb duplicate. **A** Patient 1. **B** Patient 2.

32°, 38°, and 30°, respectively. Figure 4 summarizes the main patient subgroups.

Abnormal hypertrophic epiphyses

Abnormal hypertrophic epiphyses were found in 96 thumbs and were the most common cause of ulnar deviation at the distal phalanx of the thumb. Among these 96 thumbs, 71 were associated with polydactyly, 20 of which also had ulnarly deviated MCP joints. Radiographs showed that 60.4% of these abnormal epiphyses (58 thumbs) were triangular and 35.4% (34 thumbs) were oval, whereas the rest were trapezoidal, round, or irregularly shaped (Fig. 5).

We measured the length of the radial and ulnar sides of the hypertrophic epiphyseal plate on radiographs. The length on the radial side had a mean of

3.8 mm (range, 2.2–6.1 mm), which was greater than the ulnar side, with a mean of 2.5 mm in length. In addition, we found that the distal phalanx was in good apposition and alignment with its corresponding epiphysis. The average distance between the ossification center and the distal phalanx was 0.8 mm (range, 0.5–1.1 mm).

In cases with epiphyseal deformity, 12 thumbs had a bifid distal phalanx. Eight had a hypertrophic epiphysis consisting of 2 ossified tissues, and 7 of 8 cases were associated with a triphalangeal thumb on the contralateral side. A total of 27 thumbs had obliquely oriented articular cartilage at the distal end of the proximal phalanx. In 3 thumbs, the abnormal epiphysis appeared to be fused with its distal phalanx. In one case with bilateral hypertrophic epiphyses, we noted fusion of the abnormal epiphysis and its distal

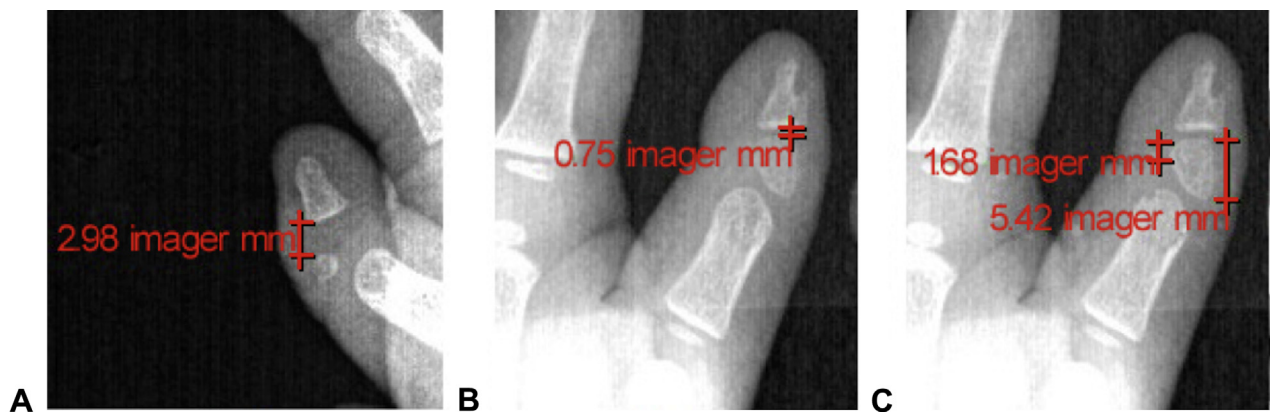


FIGURE 3: Measurements obtained on radiographs in representative patients. Distance measured between the ossification center and the distal phalanx in a **A** triphalangeal thumb (2.98 mm) and **B** obvious hypertrophic epiphysis (0.75 mm). **C** The length of the ossification center on the radial (5.42 mm) and ulnar (1.68 mm) side was measured.

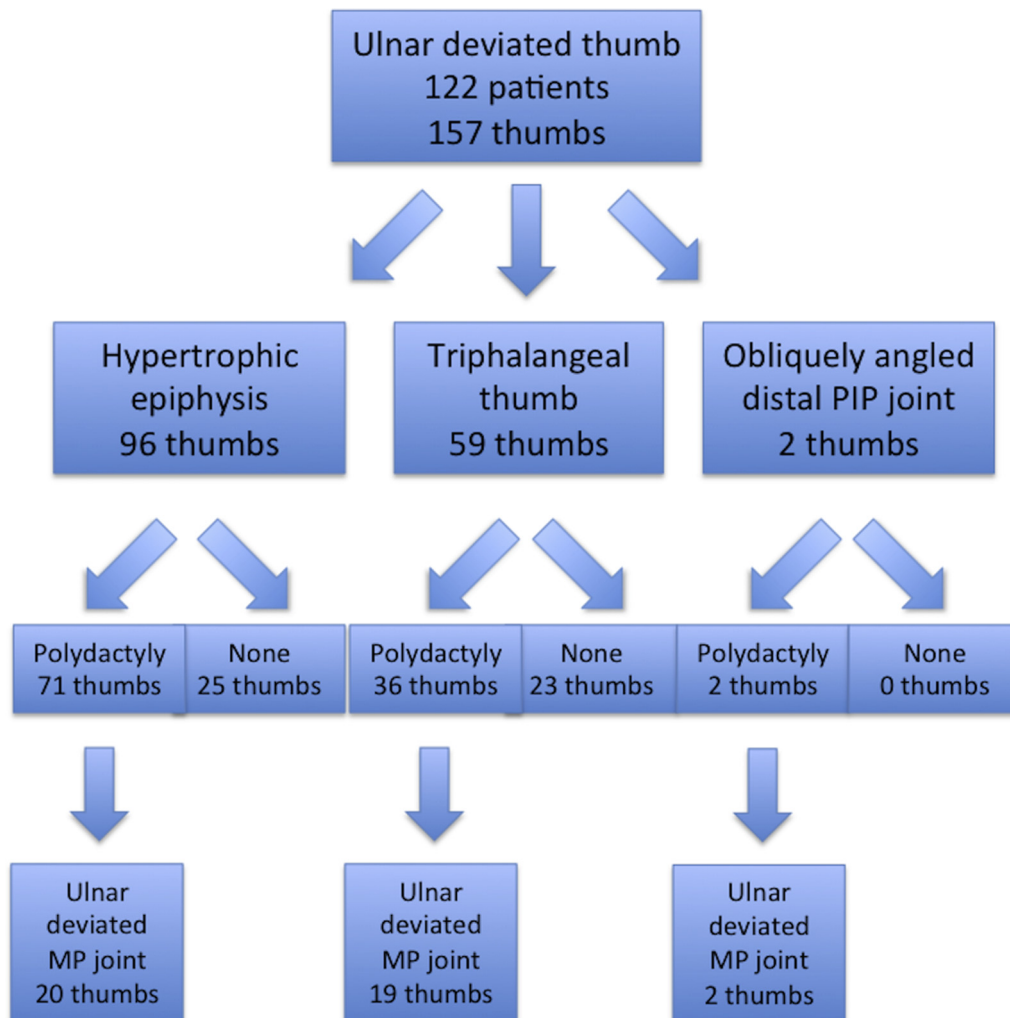


FIGURE 4: Patient subgroups. MP, metacarpophalangeal; PIP, proximal interphalangeal.

phalanx on the right thumb when the patient was seen at age 6 months, whereas its distal phalanx was notably shorter than the left.

During surgery, for all patients with abnormal hypertrophic epiphyses, we found that the distance between the joint cartilage and growth plate of distal

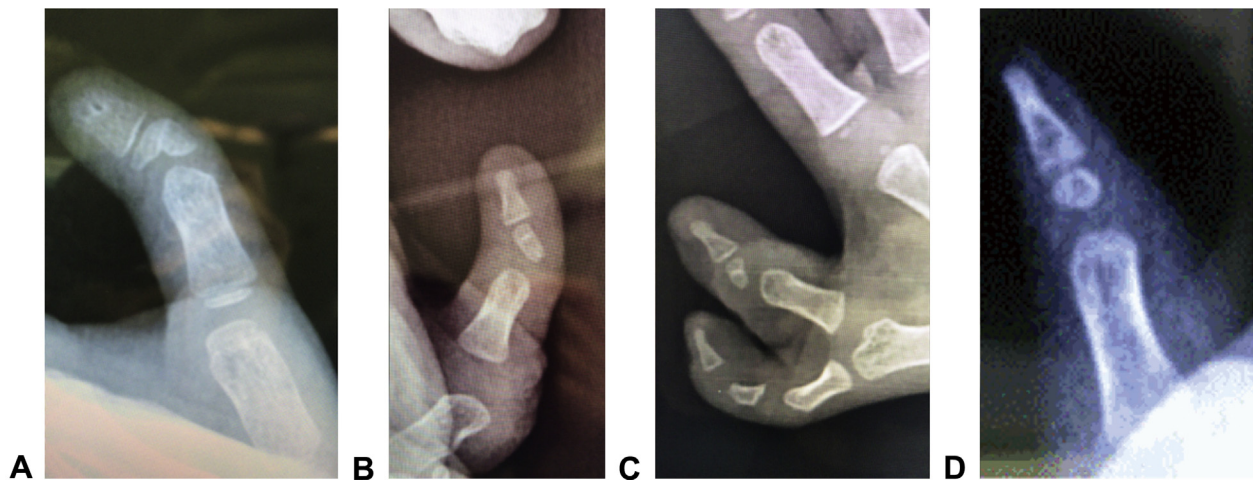


FIGURE 5: Radiographs showing different shapes of abnormal hypertrophic epiphyses, resulting in ulnar deviation of the thumb: **A** triangular, **B** oval, **C** trapezoidal, and **D** round.

phalanx was visibly enlarged, mainly on the radial side. It was ossified in the center of the enlarged part and was surrounded by cartilage. At the same time, no extra bones were seen in the IP joint, unlike with a triphalangeal thumb.

Triphalangeal thumb

Triphalangism was another important cause of the ulnar-angled thumb. It was found in 59 thumbs, 36 of which were associated with polydactyly; ulnarly deviated MCP joints were found in 19. No association was found between the presence of an ulnarly deviated MCP joint and the severity of ulnar deviation at the IP joints. The radiologic appearance of these malformed thumbs suggested that the distal epiphyseal plate was normal in shape although an extra bone block existed in the IP joints (Fig. 6). Triangular ossification was the most common shape of the extra bone, accounting for 47.5% (28 thumbs), followed by an oval shape, accounting for 22.0% (13 thumbs). In the remaining cases, the extra phalanx was trapezoidal, round, or irregularly shaped.

On radiographs, the extra bone blocks were located on the radial side of the IP joint and in poor apposition with the distal phalanx. The epiphyseal ossification center in the distal phalanx of the thumb often was not visible in radiographs until 1 year of age in some cases. However, the epiphyseal cartilage was present, so there was a larger distance between the distal phalanx and the extra bone, which measured an average of 2.8 mm (range, 2.0–4.5 mm). The length of the radial side of the extra bones averaged 3.4 mm and was always greater than the ulnar side, which averaged 2.3 mm in length.

A C-shaped epiphysis around each of the delta phalanges was found in only one patient, who had bilateral triphalangeal thumbs.

During surgery, an accessory bone block was seen lying in the IP joint as a middle phalanx; each end formed a new joint with the adjacent phalanx. However, the distal articular cartilages were dysplastic with a narrow joint space in most cases. The articular cartilage of the proximal phalanx and distal phalanx were mismatched in 24 cases.

The measured distance between the ossification center and the distal phalanx on radiographs was greater for a triphalangeal thumb compared with an abnormal hypertrophic epiphysis ($P < .05$).

Obliquely angled distal articular surface of proximal phalanx

A previously undescribed cause of ulnar deviation of the thumb was seen in 2 thumbs with Wassel IV polydactyly, in which there was neither a hypertrophic epiphysis nor a triphalangeal thumb. Instead, unequal lengths of the radial and ulnar sides of the proximal phalanx, caused by an obliquely angled distal articular surface of the proximal phalanx, resulted in ulnar deviation of the thumb (Fig. 7).

Bilateral cases

There were 35 bilateral cases. Bilateral hypertrophic epiphyses were found in 16 cases; the other 13 had triphalangism in both thumbs. In 6 cases, a hypertrophic epiphysis was found in one thumb with triphalangism in the other; there were 3 in which the abnormal hypertrophic epiphyses consisted of 2 ossified tissues.



FIGURE 6: Radiographs showing different shapes of the extra phalanx in triphalangism, resulting in ulnar deviation of the thumb: **A** triangular, **B** rectangular, **C** oval, and **D** trapezoidal.

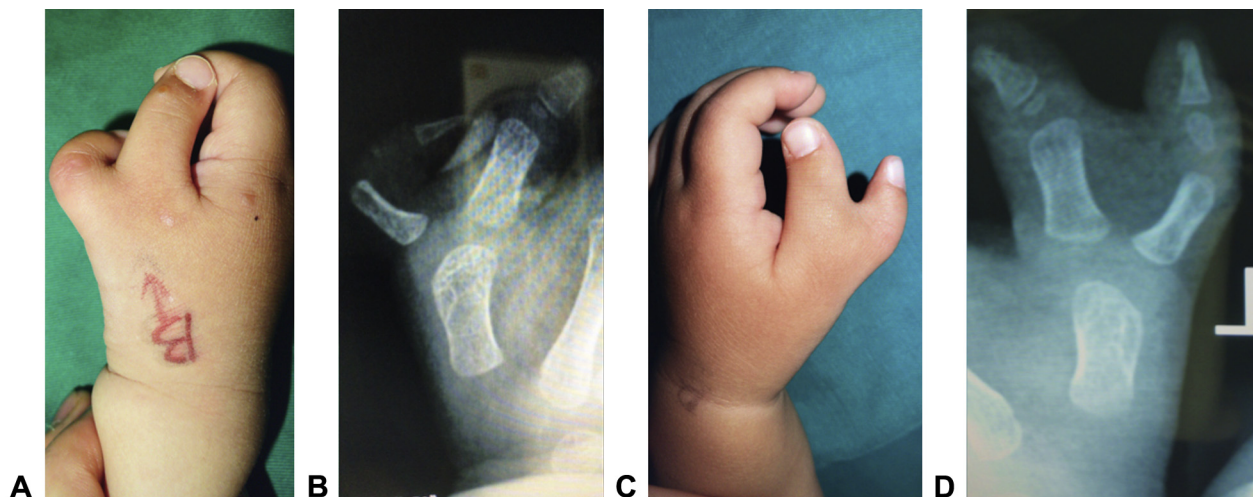


FIGURE 7: Obliquely angled articular surface of the proximal phalanx of the ulnar thumb in Wassel IV polydactyly, in the absence of a hypertrophic epiphysis or triphalangeal thumb, can result in ulnar deviation. **A, B** Patient 1. **C, D** Patient 2.

DISCUSSION

Congenital ulnarly deviated thumb at the distal phalanx may affect the appearance of the thumb and cause functional impairment. Deformity in the IP joint is usually obvious and the terminal tendon is usually inserted on the radial aspect of the base of the distal phalanx.

According to the published literature, the ulnarly deviated thumb is usually caused by an extra phalanx in a triphalangeal thumb.^{1–4} In contrast, this study found that an abnormal hypertrophic epiphysis was more common than a triphalangeal thumb, which suggests that an abnormal hypertrophic epiphysis could more commonly cause an ulnarly angulated thumb.

Theander and Carstam⁵ first noticed the difference between an epiphyseal deformity and a triphalangeal thumb in 1979. They also named the abnormal epiphysis a pseudotriphalangism. The abnormal epiphysis was been reported by Ogino et al⁶ as a subtype of delta phalanx. Baek et al⁷ introduced the term “abnormal triangular epiphysis” as another kind of deformity related to the abnormal epiphysis. In the current study, a triangular epiphysis accounted for only slightly more than half of all thumbs with epiphyseal deformity identified by preoperative radiography; the rest were oval, circular, or trapezoid. Therefore, we think it is more appropriate to define it as an abnormal hypertrophic epiphysis.

Triphalangeal thumb is a congenital autosomal dominant condition with an incidence of about 1:25,000. The etiology of the triphalangeal thumb is still unclear. Some authors⁸ opined that the additional phalanx is a remnant of the base of one of the phalanges of a bifid thumb. Others¹ think that triphalangism and polydactyly are not associated.

Ulnarly angulated thumbs at the distal phalanx caused by the 2 pathologies are similar in appearance; therefore, it is difficult to differentiate them clinically. In addition, the normal epiphysis of the distal phalanx may not appear in radiographs until age 1 year. A hypertrophic epiphysis may be misdiagnosed during surgery as a delta phalanx and excised during the wedge osteotomy.

It is important to differentiate these 2 pathologies. It is known that a delta phalanx can be easily and clearly distinguished from a secondary ossification center of the distal phalanx in a child aged more than 1 year, in whom the well-defined phalangeal epiphysis is clearly evident. However, in young infants, in whom the hypertrophic epiphysis of the distal phalanx may not be radiographically evident, Baek

et al^{7,9} introduced 2 distinguishing features. First, in a triphalangeal thumb, motion exists between the abnormal middle phalanx and the distal phalanx, whereas there is no motion between an abnormal triangular epiphysis and the distal phalanx. Second, the radiologic distance between the abnormal middle phalanx in a triphalangeal thumb and the distal phalanx is greater than that between the abnormal epiphysis and the distal phalanx because the former is a real joint that includes 2 articular cartilage surfaces.

In our experience, clinical examination of joint movement is unreliable in a young child. Based on results of this study, a distance of 2 mm or more between the abnormal bone and the distal phalanx suggests a triphalangeal thumb as opposed to an abnormal hypertrophic epiphysis as the cause of the ulnar deviated thumb.

Our approach to surgical correction for the thumb is as follows. Based on preoperative radiographic assessment, a provisional differentiation is made between an abnormal hypertrophic epiphysis and a triphalangeal thumb, which is confirmed on intraoperative exploration. For patients with an abnormal hypertrophic epiphysis, an intraepiphyseal wedge osteotomy is used for most thumbs, except for those with a small triangular hypertrophic epiphysis in which the deviation cannot be corrected fully by a wedge osteotomy because of the risk of damaging the epiphyseal growth plate. To avoid the chance of the deviation recurring after only a wedge osteotomy, partial excision of the epiphysis including the articular cartilage is also performed in these cases. Our strategy parallels that described by Horii et al,¹⁰ who described treating thumbs with epiphyseal deformity by either osteotomy in the epiphysis or partial excision of the epiphysis (including the whole articular cartilage). At follow-up examination in that series, it was confirmed that the latter worked better for patients with a small triangular hypertrophic epiphysis. However, in the long term, partial excision of the epiphysis might cause traumatic arthritis and joint stiffness.

For patients with a triphalangeal thumb, different surgical methods were reported.^{2,11} Excision of the extra bone and reconstruction of the collateral ligament are a traditional surgical method.^{12,13} Our experience suggests that we should further assess how well the articular cartilage of the proximal phalanx fits with that of the distal phalanx after removing the extra phalanx, regardless of the size of the delta phalanx. If they do not match, we believe that wedge

osteotomy and arthrodesis of the dysplastic IP joint between the extra bone and the distal phalanx results in a better outcome.

We have described a new cause of the ulnarly angulated thumb at the distal phalanx: unequal lengths of the radial and ulnar aspects of the proximal phalanx, in the absence of an abnormal hypertrophic epiphysis or a triphalangeal thumb. For these patients, we performed a distal wedge osteotomy in the proximal phalanx to level the distal articular surface and correct the ulnar deviation.

A limitation of the study is that the measured distances between the ossification center and the distal phalanx could vary by age. Mean age of patients in all 3 groups was similar and the difference in measurements between the group of patients with abnormal hypertrophic epiphysis and those with triphalangeal thumb suggests that this finding is also clinically relevant.

Abnormal hypertrophic epiphysis and triphalangeal thumb are the 2 most common causes for congenital ulnarly angled thumb at the distal phalanx. In our series, we found a new third cause, that owing to unequal lengths of the radial and ulnar aspects of the proximal phalanx. Preoperative radiographs can reveal the most likely cause of the ulnar deviated thumb and therefore guide surgical treatment.

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