

Metacarpal Hypoplasia Associated With Congenital Constriction Band Syndrome

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Purpose The amputation of digits that occurs in association with congenital constriction band syndrome (CCBS) is often described as similar to that resulting from trauma. Some previous studies have suggested that no bone hypoplasia is present in the affected limb proximal to the amputated part. The purpose of this study was to report our series of patients with metacarpal hypoplasia in amputated digits associated with CCBS.

Methods We reviewed a total of 37 hands of 24 patients diagnosed as having amputated fingers owing to CCBS and evaluated the clinical features of the metacarpal bone hypoplasia.

Results We found coexistent metacarpal hypoplasia in 11 hands in 9 patients. Three of the patients were male and 6 were female. The average age at the time of examination was 9 years (range, 4–20 y). We observed metacarpal hypoplasia in cases of amputation at the level of the proximal phalanx and not at the level of the middle phalanx.

Conclusions We found metacarpal hypoplasia in 38% of patients and in 30% of hands with amputated digits owing to CCBS. (*J Hand Surg* 2012;37A:760–763. Copyright © 2012 by the American Society for Surgery of the Hand. All rights reserved.)

Type of study/level of evidence Prognostic IV.

Key words Congenital anomaly, amputation, metacarpal, hypoplasia, brachymetacarpia.

CONGENITAL CONSTRICTION BAND syndrome (CCBS) is diagnosed using 4 main criteria and clinical features: constriction ring, lymphedema, acrosyndactyly, and amputation.¹ Digital amputation associated with CCBS is often described as similar to that resulting from trauma. Some previous studies have suggested that no bone hypoplasia occurs in the affected limb proximal to the amputated part.² Isolated hypoplasia of the metacarpal is rare in patients

with brachymetacarpia.³ However, we have experienced a case of isolated metacarpal hypoplasia associated with CCBS.

In the present study, we investigated the incidence and clinical features of metacarpal hypoplasia associated with amputated digits resulting from CCBS.

MATERIALS AND METHODS

We reviewed 27 patients associated with CCBS in our series and excluded 3 patients who did not have digital amputation. In the remaining 24 patients, we investigated 37 hands affected by digital amputation resulting from CCBS. The investigated parameters included the incidence of metacarpal hypoplasia, affected side and digits, and clinical features.

Four orthopedic surgeons (2 attending hand surgeons, 1 trainee hand surgeon, and 1 orthopedic specialist) observed radiographs of both hands of patients associated with digital amputation owing to CCBS.

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TABLE 1. Clinical Features of Metacarpal Hypoplasia Associated With Amputated Digits Owing to Congenital Constriction Band Syndrome

Case	Age at Observation (y)	Gender	Dominant Hand	Site of Metacarpal Bone Hypoplasia	
				Right Hand	Left Hand
1	6	Female	Unclear	4	
2	4	Male	Right	4, 5	2, 3, 4
3	8	Female	Unclear	4	
4	17	Male	Right	4	
5	5	Female	Right		4
6	20	Female	Unclear	2, 3	3, 4
7	9	Female	Right		3, 4
8	7	Female	Right		3, 4
9	8	Male	Right		4

**FIGURE 1:** Radiograph of both hands of a 4-year-old boy. Metacarpal hypoplasia is evident in the right ring and small and left index, middle, and ring metacarpals (black arrows).

Metacarpal hypoplasia was determined to be present when all 4 examiners diagnosed hypoplasia.

RESULTS

We found metacarpal hypoplasia in 11 hands in 9 patients. Table 1 details the patients' age, gender, hand dominance, and metacarpals involved. We

observed a constriction band on the lower extremities in 4 cases, acrosyndactyly in 7, and lymphedema in 3. There was no proximal banding of the hand and no family history of CCBS or brachymetacarpia. We observed metacarpal hypoplasia in 11 digits when there was amputation at the level of the proximal phalanx, and none when

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