Clinical Features and Operative Findings of Congenital Flexion

Deformity of Multiple Digits

Toshihiko OGINO¹, Seiichi ISHII² and Hiroyuki KATO³
¹Department of Physical Therapy, School of Health Sciences and ²Department of Orthopedic Surgery, School of Medicine, Sapporo Medical University, Minami-3, Nishi-17, Chuou-ku, Sapporo 060 and ³Department of Orthopedic Surgery, Kushiro Rousai Hospital, Nakazono-cho 13-23, Kushiro 085, Japan

ABSTRACT Thirty-six cases with congenital flexion deformities of multiple digits were classified into six types such as congenital contractural arachnodactyly, distal arthrogryposis, Freeman-Sheldon-like syndrome, congenital aplasia of the extensor muscles of the digits, ulnar drift type and multiple camptodactyly type. Many common clinical features of the hands were observed among these deformities. In eleven cases, 18 hands were treated surgically and were followed up for more than a year. During surgery, complete correction or significant reduction of the deformity was achieved in most cases. From these operative findings, it was assumed that the main cause of congenital flexion deformity of multiple digits was contracture of the palmar skin and retaining ligaments of the skin. At follow up, complete correction was achieved in 10 hands, and incomplete or minimal correction in eight hands.

Key words: hand, anomaly, camptodactyly, arachnodactyly, Freeman-Sheldon syndrome, muscle aplasia

Congenital flexion deformity of the digits is divided into two types according to the affected digits (Curra- rino and Waldman, 1964). One type is a flexion deformity of a single digit while the other affects multiple digits. Congenital clasped thumb and camptodactyly of the little finger belong to the former type. Among the latter type, congenital windblown hand (Wood and Biondi, 1990), congenital contractural arachnodactyly (Beals and Hecht, 1971), Freeman-Sheldon syndrome (Freeman and Sheldon, 1938; Burian, 1963), congenital cutaneous ulnar drift, congenital cutaneous multiple camptodactyly (Zancolli and Zancolli, 1984) and congenital aplasia of the extensor muscles of the digits (Crawford et al., 1966; Takahashi et al., 1978) have been reported. There have been few reports of treatment for congenital flexion deformities of multiple digits. The clinical features and pathogenesis of these deformities are obscure. In order to clarify the clinical features and

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pathogenesis of congenital flexion deformities of multiple digits, our own cases with these deformities were analyzed.

**MATERIALS AND METHODS**

Thirty-six patients with congenital flexion deformity of multiple digits were examined over a 12-year period. Twenty-five cases were male and 11 cases female. In all cases, both hands were affected. Out of 36 cases, 11 had congenital contractural arachnodactyly (CCA), nine had distal arthrogryposis (DA), four had Freeman-Sheldon like syndrome (F-S syndrome) and two had congenital aplasia of the extensor muscles of the digits (congenital extensor aplasia). The other 10 cases were impossible to classify into definite types. Seven of them, in which flexion deformity was predominant, were classified into the multiple camptodactyly type and the other three cases, in which ulnar deviation was predominant, into the ulnar drift type. In these cases, sex, family history, associated anomalies and the deformity of the hand itself were investigated. Operative findings and results after surgery were analyzed in 11 cases with a total of 18 hands treated surgically.

**Surgical treatment:** Two different procedures were used for correction of adduction and flexion deformities of the thumb. A rotation flap constructed from the dorsoradial surface of the index finger was used in five hands. The combination of a palmar transverse incision and a free full thickness skin graft was used in seven hands. For ulnar deviation and flexion deformities of the fingers, single or multiple palmar transverse skin incisions were made to release the skin contracture and its retaining ligaments, and the raw surface was covered with a free skin graft in 11 hands. Tendon transfer was performed after correction of the flexion deformities of the digits by splinting in four hands with congenital extensor aplasia.

**RESULTS**

**Clinical features:** Sex, incidence, family history, the deformity of the hand itself and associated anomalies are shown in Table 1. In each type except congenital extensor aplasia, males were predominantly affected. Family history was positive in nine cases. In CCA, DA, F-S syndrome and ulnar drift type, flexion deformity of the fingers, adduction and flexion deformity of the thumb and shortening of the digitopalmar skin were observed in all cases. In these cases, various degrees of ulnar drift of the fingers were observed in all cases except three of the CCA cases. In CCA, DA, F-S syndrome and ulnar drift type, four fingers were affected in almost all cases. However, in multiple camptodactyly type, the middle and ring fingers were affected in three hands, the ring and little fingers in two hands and the three ulnar fingers in two hands. In multiple camptodactyly type, there was no significant adduction contracture of the thumb or ulnar drift of the finger. In this type, shortening of the digitopalmar skin was observed only in two cases but it was minimal (Fig. 1). In two cases of congenital extensor aplasia, deformity of the hand resembled multiple camptodactyly upon first examination, but in one case, the deformity came to resemble ulnar drift type after a growth spurt.

There were associated anomalies in other systems of the body in CCA, DA and F-S syndrome. In CCA, crumpled appearance of the external ear was associated in all cases and scoliosis of the spine and joint contracture of the lower extremities were also relatively common. The contracture in these cases appeared at the hip joint in one case, knee in three cases and toe in two cases. In DA, congenital foot deformities, such as congenital club foot or congenital vertical talus were associated in all cases. In F-S syndrome, microstomia
Table 1 Clinical features of congenital flexion deformities of multiple fingers

<table>
<thead>
<tr>
<th></th>
<th>Congenital contractural arachnodactyly</th>
<th>Distal arthrogryposis</th>
<th>Freemen-Sheldon syndrome</th>
<th>Congenital apasia of extensor muscles of the digits</th>
<th>Ulnar drift type</th>
<th>Multiple camptodactyly type</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>11</td>
<td>9</td>
<td>4</td>
<td>2</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Male: Female</td>
<td>8 : 3</td>
<td>7 : 2</td>
<td>3 : 1</td>
<td>1 : 1</td>
<td>3 : 0</td>
<td>4 : 3</td>
</tr>
<tr>
<td>Positive family history</td>
<td>6</td>
<td>1</td>
<td>2</td>
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<tr>
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<tr>
<td>Adduction flexion deformity of the thumb</td>
<td>11</td>
<td>9</td>
<td>4</td>
<td>1</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Shortening of the digitopalmar skin</td>
<td>11</td>
<td>9</td>
<td>4</td>
<td>2</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Ulnar drift of the finger</td>
<td>8</td>
<td>9</td>
<td>4</td>
<td>1</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Joint contracture</td>
<td>6</td>
<td>3</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Arachnodactyly</td>
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<td>0</td>
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<td>Associated anomalies</td>
<td></td>
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<td>Leg:</td>
<td></td>
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<tr>
<td>Club foot</td>
<td>0</td>
<td>8</td>
<td>2</td>
<td>0</td>
<td>0</td>
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<tr>
<td>Vertical talus</td>
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<td>4</td>
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<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
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<tr>
<td>High palate</td>
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<td>1</td>
<td>0</td>
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</tbody>
</table>

Note: Joint contracture of the leg: hip in one case, knee in three cases, toe in two cases.
and micrognathia were associated in all cases and bilateral congenital club feet in two cases.

**Operative findings and results after surgery:** In 14 hands of nine patients, contracture of the palmar skin was released during surgery. After release of the palmar skin and retaining ligaments of the skin, nearly complete correction of the deformity of the digit could be achieved in 10 hands of five patients during surgery. In four hands of the other four patients, significant improvement of the deformity was achieved but complete correction was not possible. In four hands, shortening of the neurovascular bundles of the affected digit was observed after release of the skin contracture. In two of these patients it limited extension of the digit. In one hand, there was shortening of the flexor digitorum superficialis of the middle and ring fingers. The age at operation in cases with shortening of the neurovascular bundles ranged from 2.6 to 7 years with an average of 3.6 years, apparently higher than that of cases without shortening of the neurovascular bundles, the latter ranging in age from 4 to 14 years with an average of 9.3 years. However, there was no statistical difference between these ages. Four hands of two cases with congenital extensor aplasia were treated with tendon transfer. In one of them, there was aplasia of the extensor indicis proprius in both hands and of the extensor digitorum communis in the right hand as well as hypoplasia of the extensor tendon of the extensor digitorum communis in the left hand. In this case, the extensor digitii minimi was transferred to the extensor of four fingers in the right hand and the half of the extensor digitorum communis of the middle finger was transferred to the extensor hood of the index finger in the left hand. In the other case, there was aplasia of the extensor digitorum communis in both hands and of the extensor indicis proprius in the right hand. In this case, the extensor carpi ulnaris was transferred to the extensor digitorum communis in the left hand and the distal end of the flexor digitorum superficialis of the ring finger was divided into three parts and transferred to the extensor hood of the index, middle and ring fingers.

In 11 cases, a total of 18 hands could be followed up after surgery. The follow-up periods ranged from one year to 12 years with an average of 4.2 years. At follow-up, deformities were nearly completely corrected in 10 hands, incomplete correction was achieved in six hands, and improvement was minimal in two hands (Figs. 2, 3 and 4).
DISCUSSION

When congenital flexion deformities of multiple digits are examined, if ulnar deviation is predominant, it is diagnosed as windblown hand, whereas if flexion deformity is predominant, it is diagnosed as camptodactyly with multiple affected fingers, and if arachnodactyly is present, it is diagnosed as CCA. However, many common clinical features were observed among these deformities: bilateral and symmetrical involvement, clenched finger at birth, shortening of the digitopalmar or digital skin, and slight or mild contracture of finger joints (Figs. 4, A and 5). In almost all cases, passive extension of the interphalangeal joint causes flexion and ulnar deviation of the finger at the metacarpophalangeal joint. Furthermore, passive extension of the metacarpophalangeal joint causes flexion of the finger at the proximal interphalangeal joint. From these facts it is sometimes difficult to differentiate between windblown hand, ulnar drift type, and multiple camptodactyly.
Fig. 3  External appearance of the hand before and after surgery in ulnar drift type.
A: Flexion and ulnar deviation were observed before surgery.
B: The deformities were corrected incompletely at follow-up.

Fig. 4  External appearance of the hand before and after surgery in Freeman-Sheldon like syndrome.
A: Clenched finger of the right hand at birth.
B: External appearance before surgery. Active extension was improved by splint therapy.
C: Active extension of the finger five years after surgery. Improvement of the deformity of the thumb was minimal.
Congenital flexion deformity of multiple digits

Congenital flexion deformities of multiple digits are often associated with anomalies in other systems of the body. Wood and Biondi (1990) reported that descriptions of multiple defects in these cases contributed to confusion about terms. However, these deformities could be classified into six types according to the associated anomalies in other systems of the body as follows; congenital contractual arachnodactyly, distal arthrogryposis, Freeman-Sheldon like syndrome, congenital aplasia of the extensor muscles of the digits, ulnar drift type and multiple camptodactyly type. CCA was first described by Beals and Hecht (1971) and is characterized by multiple congenital joint contracture, arachnodactyly, crumpled ears and kyphoscoliosis. It is transmitted as an autosomal dominant trait but is not associated with cardiovascular and ocular complications as in Marfan's syndrome. In this series, the hand deformity associated with this syndrome was flexion deformity of the fingers with ulnar deviation and adducted thumb as reported by Arroyo et al. (1985). They reported that arachnodactyly was not a constant feature of this syndrome. From these facts, it is impossible to make a differential diagnosis from other syndromes according to the hand deformity. The deformity of the external ears seems to be the most characteristic feature of this syndrome and is therefore valuable for differential diagnosis (Ogino et al., 1993).

Distal arthrogryposis was first described by Hall and Greene (1982) and was characterized by congenital contracture of the hands and feet. He classified this syndrome into two types. Type 1 is not associated with anomalies except in the extremities and type 2 is associated with other anomalies. Type 2 may include several
different syndromes. In this paper, DA means only type 1. Type 1 syndrome, termed of distal arthrogryposis, includes digitotatal dysmorphism. The hand deformity of this syndrome resembles that of CCA. Foot deformities such as congenital club foot or vertical talus seem to be characteristic features useful for differential diagnoses.

Freeman-Sheldon syndrome is characterized by a whistling face and windblown hands (Freeman and Sheldon, 1938). In this series, there was no typical whistling face and the cases with microstomia and micrognathia were diagnosed as F-S syndrome. There was overlapping of clinical features between DA and F-S syndrome. The deformity of F-S syndrome seems to be more severe than those of the other types.

Various pathologies have been reported as causes of flexion deformity of the digits. An imbalance of the forces between the flexor and extensor sides might cause flexion deformity of the digits (Millesi, 1974). Some authors have suggested that the primary cause might be on the flexor side, either contracture of the palmar skin, congenital fibrous substrata beneath the palmar skin in the subcutaneous tissue (McCash, 1966) or contracture of the flexor digitorum superficialis (Smith and Kaplan, 1968). Others have suggested that the primary cause might be on the extensor side, either abnormal insertion of the lumbrical muscles (Maeda and Matsui, 1985) or faulty development of the extensor aponeurosis over the PIP joint (Berger and Millesi, 1975). However, most of these authors did not differentiate between flexion deformity of multiple digits and that of a single digit. However, the pathogenesis of camptodactyly of the little finger is different from that of flexion deformity of multiple digits (McFarlen et al., 1983; Ogino and Kato, 1992). With regard to flexion deformity of multiple digits, Wood and Biondi (1990) suggested that windblown hand is a variation of some type of arthrogryposis, considered to be caused by faulty development of the joint. In this series, flexion deformity was completely corrected or significantly reduced in most cases after release of the skin contracture and retaining ligaments of the skin during surgery. Thus, the main cause of flexion deformities with multiple digital contracture seems to be contracture of the palmar skin and retaining ligaments of the skin, as reported by Zancolli and Zancolli (1984). They reported that shortening of the palmar skin and midpalmar fascia cause ulnar deviation and that shortening of the palmar skin and digital fascia causes flexion deformity of the fingers. The former was called congenital cutaneous ulnar drift, the latter, congenital cutaneous multiple camptodactyly. On the basis of these theories, it is easy to understand that there are many common clinical features among hand deformities with multiple digital flexion deformity.

In this series, full extension of the digit could not be achieved during surgery in some cases, because of shortening of the neurovascular bundles. The age of the group with shortening of the neurovascular bundles was higher than without shortening of neurovascular bundles, although there was no statistical difference between the two groups. Thus, the neurovascular bundle seemed to be retracted secondarily. On the other hand, in this series, some cases had unsatisfactory results at follow-up although complete correction of the deformity had been achieved by release of the contracted palmar skin and its retaining ligament. As described above, faulty development or secondary elongation of the extensor of the digits might be related to the cause of the flexion deformity in these digits. The deformity of the hand in congenital extensor aplasia resembles that of camptodactyly of multiple digits. In congenital extensor aplasia, contracture of the digitopalmar skin and retaining ligaments of the skin might be secondary change. It is possible for congenital extensor aplasia to be diagnosed as camptodactyly of multiple digits. We must also pay attention to this problem. It is unclear whether or not contracture of the palmar skin and its retaining ligaments is actually a primary cause. In order to clarify the pathogenesis of these deformities, further study is required.
REFERENCES


