Camptodactyly: A Unifying Theory and Approach to Surgical Treatment

P. J. Smith, FRCS,
A. O. Grobbelaar, FCS(SA), MMed(Plast. Reconst. Surg.), FRCS(Plast.),
London, UK

Camptodactyly is an isolated congenital flexion deformity of the proximal interphalangeal (PIP) joint. Surgical experience with 16 patients (18 fingers) between June 1983 and December 1994 is reported. Skin, fascia (retinaculum cutis), tendon sheaths, flexor digitorum superficialis tendon, lumbricals and interossei (particularly the lateral bands), joint surfaces, neck of the proximal phalanx, and central slip insertion were involved in all cases, although the degree of involvement can vary. Surgery must address all of these structures. Postoperative splinting is important. Fifteen patients had good or excellent results after surgery, with a mean gain in motion of 57° (range, 0°–90°). Surgery should be aimed at prevention of progressive deterioration and is probably not indicated in minor degrees of the deformity. Surgery should be reserved for patients with a preoperative PIP joint contracture of more than 60°. (J Hand Surg 1998;23A:14–19. Copyright © 1998 by the American Society for Surgery of the Hand.)

Camptodactyly was first described by Tamplin in 1846 in his "Lectures on the Nature and Treatment of Deformities" at the Royal Orthopaedic Hospital, Bloomsbury Square, London.[1] The term *camptodactyly* is of Greek origin and was used by Landouzy in 1906 to describe an irreducible flexion contracture affecting the proximal interphalangeal (PIP) joints in young girls.[2]

In camptodactyly, the little finger is most often involved. There can be involvement of other digits, with a diminishing incidence toward the radial side of the hand.[3,4] It occurs in less than 1% of the population,[5] although De Haas reported an incidence varying from 2 in 3,000 to 58 in 239.[6] It is often of no functional significance, counting surgeons among its numbers. It presents either in early childhood or in adolescence and may be static or progressive in degree of deformity.[1–3,5] Yet, even in the static group with deformity of no functional significance, it is surprising how often patients resist advice to leave well enough alone or to pursue only a conservative course using splinting; such are the high and unreasonable expectations of modern patients.

Siegert et al. categorized camptodactyly into simple and complex groups.[7] The simple type represents flexion deformity of the PIP joint only, while the complex group includes all the cases associated with other deformities (e.g., syndactyly, clinodactyly) Smith and Kaplan, Miura et al., and Engber and Flat all mention an association with other congenital deformities.[1,8,9]

The cause of the deformity is obscure. Hereditary factors, tuberculosis, rheumatoid disease, and ischemia have been cited.[2,3,10] Specific anatomic abnormalities have been implicated.[8]

The physician seeking to give therapeutic advice can be in a quandary, for the literature advocates 2 schools of thought. One group recommends conservative treatment using splinting, which may need to be serial, nocturnal, and continued until early adult life[11]; the other often recommends a more aggressive surgical approach.[12–14] To make matters worse, the surgical series are often confined to groups representing failed conservative treatment.[4,7,9] Those advocating surgery detail a series of isolated abnormalities

From the Department of Plastic and Reconstructive Surgery, Mount Vernon Hospital, London, UK.

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Reprint requests: P. J. Smith, FRCS, Department of Plastic and Reconstructive Surgery, Mount Vernon Hospital, Rickmansworth Road, Northwood, Middlesex, UK HA6 2RN.

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Table 1. Associated Congenital Anomalies

Anomalies	*No. of Patients*
Arthrogryposis multiplex congenita	1
Hypoplastic thumb	1
Plagiocephaly	1
Anal agenesis	1
Cleft lip and palate	1

accounting for the deformity—in 1 series, the lumbricals[15–17]; in another, the flexor digitorum superficialis (FDS) tendon[1–9]—making logical surgical intervention difficult.

The purpose of this study was to investigate the specific anatomic abnormalities found in camptodactyly and to assess whether there was an identifiable pattern that could lead to a logical and uniform surgical approach to the problem.

Here, we present the view that skin, fascia (retinaculum cutis), tendon sheaths, FDS tendon, lumbricals and interossei (particularly the lateral bands), volar plate, accessory collateral ligaments, joint surfaces, neck of the proximal phalanx, and central slip insertion are involved in all cases, although the degree of involvement can be variable. This unifying theory of camptodactyly leads to a unified approach to surgical treatment. We present the concept and notions of such an approach.

Materials and Methods

The cases of 16 patients (18 fingers) who underwent surgery between June 1983 and December 1994 at Mount Vernon Hospital and the Hospital for Sick Children were analyzed. There were 16 little fingers, 1 middle finger, and 1 ring finger involved. There were 10 male and 6 female patients. Two patients had 2 fingers each that were repaired; 7 patients had multiple fingers involved. Associated congenital abnormalities are listed in Table 1. The mean patient age at surgery was 8 years (range, 3–16 years).

The entrance criterion for this study was the presence of camptodactyly that necessitated surgical treatment. This was deemed necessary when 6 months of conservative treatment had failed to arrest progression of the deformity. In general, this meant that the patients had a 60° flexion contracture at the PIP joint when the metacarpophalangeal (MP) joint was held in flexion and that this deformity was still causing deterioration. There were 2 patients with lesser deformities at the PIP joint but who were included because of a very rapidly progressing contracture.

Surgical Technique

Surgical treatment followed a standardized pattern that we apply to all camptodactyly cases. In this pattern, a volar linear incision is used and converted to multiple z-plasties, placing the central limbs over the flexion creases of the joints. On reflecting the skin, the shortened retinaculum cutis (or tough linear fibrous bands of digital fascia, as described by McCash[18]) is visualized; it runs from the pretendinous band distally and includes the lateral digital sheets. They are all released, including the bony attachment of Grayson's ligaments. At this stage, the lateral bands of the intrinsic apparatus and interosseous muscles are freed from their abnormal and widespread attachment to the sides of the proximal phalanx. It is this attachment that prevents extension of the PIP joint. On its release, proximal pull on the lateral bands will confirm that PIP extension can now occur. Some attenuation of the central slip may have occurred and is probably secondary, as in other forms of prolonged PIP joint contracture. Its presence can be confirmed by the central slip tenodesis test.[19] This test has been described elsewhere by the senior author (P.J.S.)[19]; it involves flexing the wrist and the MP joints. In the normal hand, the PIP joint will automatically fully extend, owing to tension on the central slip insertion. If the central slip is attenuated, there will be an extensor lag at the PIP joint. In such patients, the central slip can be treated by appropriate postoperative splinting. It is important that the reader understand that we do not surgically explore and visualize the central slip; this is a closed test. The lumbrical muscle is abnormally inserted and often adherent to the proximal phalanx. It may also have an abnormal origin and occasionally may be inserted into the FDS tendon proximal to the A1 pulley. The FDS tendon is tested by a tenodesis test to ascertain whether it is short. If the PIP joint cannot be fully extended when the wrist is in extension, the FDS is short and must be released. Two types of FDS abnormalities exist: (1) one in which the FDS is merely short, and (2) the other in which only the distal portion of the FDS is present, there is proximal aplasia, and the distal part of the FDS acts as a tenodesis, producing a flexion contracture of the PIP joint. In such cases, complete surgical release by division is undertaken. If the FDS is normal proximally, then a lengthening and transposition of the FDS insertions is performed at chiasma level. The FDS insertions are both divided—but eccentrically, such that a long proximal and radial insertion can

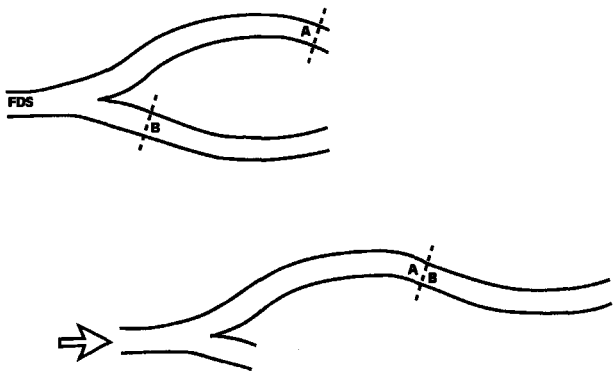

Figure 1. Lengthening of the flexor digitorum superficialis (FDS) tendon. The dotted line indicates the resection of (A) the long proximal radial insertion and (B) the long distal ulnar insertion.

then be sutured to the ulnar insertion in such a way that lengthening is achieved (Fig. 1).

The procedure up to this point will fully release the majority of contractures. A small minority may require release of the flexor tendon sheath, volar plate, or even the accessory collateral ligaments. The central slip attenuation responds to postoperative extension splinting.

We do not explore any preoperatively detected bony abnormalities at the neck of the proximal phalanx or involving the articular surfaces of the PIP joint. Past experience has lead us to believe that there is no benefit to be gained from complicated osteotomies.

The involvement of the structures noted in Table 2 was recorded for each digit operated on. The postoperative results were graded according to the classification used by Siegert et al.[7] (Table 3).

Table 2. Structures Involved in Patients With Camptodactyly

No. of Digits	*Structures Involved*	*Percentage of Cases*
18	Skin	100
12	Flexor digitorum superficialis and tendon sheaths	66.6
10	Retinaculum cutis	55.5
4	Lumbricals	22
3	Bone (abnormal proximal interphalangeal joint surfaces and neck of proximal phalanx)	16.6
3	Volar plate	16.6
2	Central slip	11
2	Adherence of lateral bands to proximal phalanx	11
1	Accessory collateral ligaments	5.6

Table 3. Classification of Results by the Method of Siegert et al.[7]

Classification	*Explanation*
Excellent	Correction to full extension, with <15° loss of PIP joint flexion
Good	Correction to within 20° of full PIP joint extension and >40° increase in PIP joint extension, with <30° loss of flexion
Fair	Correction to within 40° of full PIP joint extension and >20° increase in PIP joint extension, with <45° loss of flexion
Poor	<20° of improvement in PIP joint extension, with <40° of total PIP joint motion

PIP, proximal interphalangeal.

Postoperative Regimen

The postoperative program involved maintaining the PIP joint in extension for 4 weeks. Early in the series, we used Kirschner wires (K-wires), but later, we relied more upon dorsal and volar plaster of Paris slabs. Admittedly, K-wires maintain the PIP joint correction, but in cases of prolonged flexion contractures, their use can lead to vascular embarrassment due to the tethering of the neurovascular bundles by fibrous tissue. The slight movement of the finger within the tendinous structures allowed by plaster of Paris would seem desirable. After 4 weeks, a soft Capener splint (Exeter Mobility, Exeter, UK) was used for a further 2 weeks during the day, allowing active resisted flexion. If the FDS had been lengthened, the initiation of active resisted flexion would have been postponed to week 8. At 6 weeks, free daytime motion was combined with night splinting (continued for 4–6 months). This postoperative regimen will deal with any central slip attenuation that is detected by the closed test.[19]

Results

As noted in Table 2, the skin was always involved, a short FDS tendon and tendon sheath was present in 67% of the patients, and the retinaculum cutis was involved in 56%. Lumbrical involvement occurred in 22%. Bone and joint abnormalities that were determined purely by x-rays and not by exposure of the joint surfaces at surgery accounted for a 17% incidence, and not surprisingly, a volar plate tightness was identical in incidence. Central slip attenuation and adherence of the lateral bands to the proximal phalanx were both equal at 11%; again, this is not surprising, as both deformities would tend to occur at the same time. Tightness of the accessory collateral

Table 4. Siegert Grades

Siegert Grade	*No. of Patients*
Excellent	6
Good	9
Fair	2
Poor	1

ligaments was the rarest of all the problems encountered. Patients were assessed at a mean of 2.8 years after surgery (range, 8 months to 9 years).

It is important to remember that this series represents a sequential series of patients who were observed and documented while we were developing a theory about the degree of anatomic involvement of the various structures. As a result, early in the series we did not detect deformities that became quite apparent to us later in the series, and thus the results do not reflect a true estimate of the incidence of the involvement of some structures, namely, the FDS tendons, retinaculum cutis, lumbrical abnormalities, and interossei abnormalities. Later in the series, it became apparent that these structures were always involved. We will report on a further series of patients in the future. It will be interesting to determine whether our impression that these structures are always involved is correct.

The results were classified according to the Siegert classification and are shown in Table 4; pre- and postoperative ranges of motion (ROM) are shown in Table 5. The mean preoperative ROM at the PIP joint was an arc of 28° (range, 10°–55°). Following surgical treatment, the mean ROM was 86° (range, 45°–110°). The results revealed that 6 fingers were excellent grade using the Siegert scale, 9 were good, 2 were fair, and 1 was poor. This gave us good to excellent results in 15 of 18 patients (83%).

One patient's condition was made worse by the operation. In this patient, there was multiple involvement of the anatomic structures associated with joint stiffness and bony deformity of the PIP joint. Both our patients with fair results had more than 1 finger per hand involved. All 3 patients falling into the fair and poor categories also had bony deformities shown on x-rays. Radiologic examination revealed an abnormality of the PIP head in all 3 patients, with volar angulation of the neck of the proximal phalanx in 1. These patients also failed to comply with the postoperative regimens. Interestingly, the preoperative PIP contracture in these 3 patients was 35°, 40°, and 60°, respectively. This confirms the experience of other authors; it has been shown that increased contracture severity at the PIP joint is not associated with a poorer prognosis.[7]

Discussion

Anatomic Factors

Various factors have been blamed by different authors as the primary cause of camptodactyly.

Table 5. Pre- and Postoperative PIP Joint Range of Motion

Patient No.	*Sex*	*Age (y)*	*Finger Involved*	*Preoperative PIP ROM (degrees)*	*Postoperative PIP ROM (degrees)*	*Siegert Grade*
1	M	14	LV	70/110	0/110	Excellent
2	M	3	RV	70/95	10/90	Good
3	M	12	LV	40/95	10/90	Fair
4	F	16	RV	80/100	0/100	Excellent
5	M	5	RV	65/105	15/100	Good
6	M	4	RV	90/100	0/100	Excellent
7	M	3	LV	90/110	15/100	Good
8	F	3	LIII	80/100	10/95	Good
9	M	7	RV	60/110	35/90	Fair
10	M	5	RV	70/100	10/95	Good
11	F	4	RV	80/105	0/100	Excellent
12	F	4	LV	90/105	10/100	Good
13	M	13	RV	70/95	15/90	Good
14	F	12	RV	35/90	50/95	Poor
15	M	4	RV	90/100	0/95	Excellent
			RIV	80/105	15/100	Good
16	F	16	RV	90/100	0/95	Excellent
			LV	70/105	10/100	Good

PIP, proximal interphalangeal; ROM, range of motion; L, left; R, right; III, third finger; IV, fourth finger; V, fifth finger.

Todd[10] stated that the stiffness of the joint seemed to be entirely due to changes in the soft tissue parts and that the principal contracture seemed to be in the capsule of the joint. Oldfield also blamed the soft tissue on the flexor surface of the affected fingers.[2] The FDS has been implicated as a significant factor by Stoddard (abnormal shortness),[20] Scott (tight flexor tendon under the skin),[21] Herbert (slow retraction of the flexor tendon),[1] and Smith and Kaplan (contracture of FDS).[1] McFarlane et al. suggested that an abnormal lumbrical insertion is the major deforming force.[4] Koman et al. believed that extensor mechanism anomalies are primary and the palmar manifestations of a tight FDS tendon and contractures of the palmar soft tissue are secondary.[22] Millesi believed that abnormal development of the central slip and dorsal aponeurosis over the PIP joint was the cause of flexion contracture deformity.[4] The theory of disturbed equilibrium between flexor and extensor forces has been accepted by Engber and Flatt,[9] Koman et al.,[22] and Miura et al.,[8] although the primary cause is still unclear.[9,13,22] McCash pointed out 3 main factors: skin shortage on the volar side, congenital fibrous substrate present beneath the skin, and muscle imbalance.[18]

Our view of the anatomic factors involved in camptodactyly differs from that of these authors in that we believe that rather than isolated anatomic factors that differ in each individual, it is involvement of all the anatomic factors previously discussed to a variable degree that occurs in all patients. Thus, we believe that there is a skin shortage, but we are not sure whether this is primary or secondary; we believe that the underlying fascial structures that have been grouped under the generic term *retinaculum cutis* are always involved and always short and that this is primary. Part of this involvement contributes to the formation of abnormal attachments between the intrinsic apparatus and the sides of the proximal phalanx, thus robbing the PIP joint of dynamic extensor forces. The lumbrical is nearly always abnormal, as is the FDS tendon. All of the foregoing factors combine to produce a flexion contracture as well as a lack of extensor force at the PIP joint. With progressively increasing contracture associated with growth of the hand, secondary attenuation of the extensor apparatus occurs, leading to complete inability to extend the PIP joint. Bony deformity ensues, although most authors, including Smith and Kaplan and Miura et al.,[1,8] agree that flattening and tapering of the proximal phalanx shown on x-rays are secondary and due to compression of the head by the central slip, which is attenuated and stretched. The bony abnormality may well be a primary deformity in syndromic camptodactyly. Some indication of whether the abnormality is primary or secondary may be obtained by analyzing the degree of correction achieved at the time of surgical intervention: full correction may indicate a secondary abnormality, whereas failure to produce full extension may indicate a primary bony abnormality. This is an area where further study results would be interesting. We documented not the completeness of correction achieved at surgery but only the endpoint at the time of postoperative assessment. The reader should also remember that these results represent an evolution in our surgical approach. Many of the abnormal factors that we increasingly documented toward the end of the series were simply not recognized early on. We now believe that all of these structures are abnormal and that surgical attention must be directed to all of them to obtain the best possible result. The isolated correction of a FDS tendon or lumbrical abnormality will not correct camptodactyly.

Treatment

Treatment can be either conservative,[11] surgical,[2] or a combination regimen in which only certain patients undergo surgery.[7,9,12–14] Hori et al.[11] advocated using a dynamic splint worn 24 hours a day until full extension of the PIP joint was achieved, followed by a regimen in which the splint is worn 8 hours a day. Contracture tended to recur when the dynamic splint was no longer used. This raises the question of when to tell the patient to stop wearing the splint. Patient compliance seems highly unlikely.

Siegert et al. performed release of the FDS according to Smith and Kaplan's recommendations, which was followed by a palmar capsulotomy and collateral ligament release if necessary.[7] Engber and Flatt advocated a combination of volar soft tissue releases: some combination of skin, subcutaneous tissue, flexor tendon sheath, FDS, PIP collateral ligament, and volar plate release.[9] Osteotomy was used occasionally.

Various tendon transfers and lengthenings have been used. McFarlane et al. advocated FDS V transfer to the extensor mechanism after an anomalous insertion of the lumbrical muscle had been sought.[4,12] Koman et al. lengthened or transferred the FDS tendon and reconstructed the extensor mechanism.[22] Lengthening the FDS appears sensible; however, FDS transfer into the lateral bands is too powerful, in our opinion, and is unnecessary, as the central slip attachment will tighten up with splinting.

Gupta and Burke recommended the extensor indicis proprius (EIP) transfer to the radial side of the extensor expansion in an attempt to strengthen the intrinsic action.[5] EIP transfer proximal to the attenuated central slip will not work and must be accompanied by release of the lateral bands from the proximal phalanx to achieve PIP extension.

Results of treatment are difficult to compare owing to lack of objective data in most studies. We preferred to use the classification of results proposed by Siegert et al.[7] They reported only 7 good results and no excellent results, using their classification, after surgical release in 38 digits. We had 15 good or excellent results after surgical treatment of eighteen digits. McFarlane et al. reported perfect results in 22% of cases after 1 year.[12] Preoperative joint contractures play an important role in the eventual outcome. McFarlane et al. reported an average preoperative PIP joint contracture of 49°,[12] and Siegert et al., of 44°.[7] Engber and Flatt reported a preoperative average of 50° (range, 10°–110°).[9] The preoperative average in our series was 73° (range, 35°–90°). In the series of Siegert et al., the surgical results were worse if preoperative contracture was less than 60°.[7] Our series confirms this. The 2 patients with PIP joint contracture of less than 60° had a fair and a poor result after surgery. McFarlane et al. used 30° or more of contracture as an indication for surgery and did not correct those of under 30°.[12] Siegert et al. recommended surgery if the contracture was more than 60°.[7] We now reserve surgical intervention for cases with PIP joint contracture of more than 60°. This somewhat surprising observation is because of the fact that surgery on patients who have minor contractures is more likely to produce complications than to produce benefits.

We believe that we have achieved unexpectedly good results by concentrating on the surgical treatment of all the involved structures. When surgical treatment is attempted, release of all the involved structures should be performed as described in this series. Postoperative rehabilitation is important, as loss of flexion is a disaster. It is also important to instill a sense of realism in patients at the onset of treatment by explaining that they will be fortunate to obtain a correction of the deformity and that if they do, they should regard it as an unexpected bonus. It should be explained that the real aim of surgery should be to prevent further progressive deterioration.

References

1. Smith RJ, Kaplan EB. Camptodactyly and similar atraumatic flexion deformities of the proximal interphalangeal joints of the fingers: a study of thirty-one cases. J Bone Joint Surg 1968;50A:1187–1203.
2. Oldfield MD. Camptodactyly: flexor contracture of the fingers in young girls. Br J Plast Surg 1956;8:312–317.
3. Courtemanche AD. Campylodactyly: etiology and management. Plast Reconstr Surg 1969;44:451–454.
4. McFarlane RM, Curry GI, Evans HB. Anomalies of the intrinsic muscles in camptodactyly. J Hand Surg 1983;8: 531–544.
5. Gupta A, Burke FD. Correction of camptodactyly. J Hand Surg 1990;15B:168–170.
6. De Haas WHD. Camptodactylie Nederlands tijdschr Geneesk 1957;101:2121–2124.
7. Siegert JJ, Cooney WP, Dobyns JH. Management of simple camptodactyly. J Hand Surg 1990;15B:181–189.
8. Miura T, Nakamura R, Tamura Y. Long-standing extended dynamic splintage and release of an abnormal restraining structure in camptodactyly. J Hand Surg 1992;17B:665–672.
9. Engber WD, Flatt AE. Camptodactyly: an analysis of sixty-six patients and twenty-four operations. J Hand Surg 1977;2:216–224.
10. Todd AH. Hereditary contracture of the little fingers (Kamptodactyly). Lancet 1929;2:1088–1089.
11. Hori M, Nakamura R, Inoue MD et al. Nonoperative treatment of camptodactyly. J Hand Surg 1987;12A:1061–1065.
12. McFarlane RM, Classen DA, Porte AM, Botz JS. The anatomy and treatment of camptodactyly of the small finger. J Hand Surg 1992;17A:35–44.
13. Miura T. Nontraumatic flexion deformity of the proximal interphalangeal joint: its pathogenesis and treatment. Hand 1983;15:25–34.
14. Ogino T, Kato H. Operative findings in camptodactyly of the little finger. J Hand Surg 1992;17B:661–664.
15. Maeda M, Matsui T. Camptodactyly caused by an abnormal lumbrical muscle. J Hand Surg 1985;10B:95–96.
16. Inoue G, Tamura Y. Camptodactyly resulting from paradoxical action of an anomalous lumbrical muscle. Scand J Plast Reconstr Hand Surg 1994;28:309–312.
17. Minami A, Sakai T. Camptodactyly caused by abnormal insertion and origin of lumbrical muscle. J Hand Surg 1993;18B:310–311.
18. McCash C. Congenital contractures of the hand. In: Stack HG, Botton H, eds. The proceedings of the Second Hand Club. London: British Society for Surgery of the Hand, 1975:399–401.
19. Smith PJ, Ross DA. The central slip tenodesis test for early diagnosis of potential Boutonniere deformities. J Hand Surg 1994;19B:88–90.
20. Stoddard EE. Nomenclature of hereditary crooked fingers: Streblomicrodatyly and camptodactyly—are they synonyms? J Hered 1939;30:511–512.
21. Scott J. Hammer finger with notes of seven cases occuring in one family. Glasgow Med J 1903;60:335–344.
22. Koman LA, Toby EB, Poehling GG. Congenital flexion deformities of the proximal interphalangeal joint in children: a subgroup of camptodactyly. J Hand Surg 1990; 15A:582–586.