Treating Congenital Proximal Interphalangeal Joint Contracture

Sarah M. Yannascoli, MD, Charles A. Goldfarb, MD*

KEYWORDS

• Proximal interphalangeal joint • Contracture • Camptodactyly • Congenital

KEY POINTS

- Congenital proximal interphalangeal (PIP) joint contracture, also known as camptodactyly, is a nontraumatic PIP joint flexion deformity most commonly affecting 1 or both small fingers, sometimes involving other fingers in isolation or in conjunction with a small finger deformity, and presenting in a bimodal age distribution during periods of rapid growth (<2 years old and >10 years old).
- There is no unified consensus on the cause of camptodactyly; however, the comprehensive theory of an imbalance between the flexor and extensor mechanisms acting across the PIP joint is well accepted.
- Early, diligent passive stretching, extensor strengthening, and splinting regimens are often successful and sufficient, leading therapy to be the mainstay of treatment. These protocols must be altered based on patient age and activity level.
- Surgical treatment should proceed in a stepwise manner addressing the skin, underlying fascial structures, intrinsic muscles, lateral bands, flexor digitorum superficialis tendon, intrinsic PIP joint disorder, and secondary distal interphalangeal joint deformity.
- Patient expectations must be managed preoperatively, compliance is a requirement, and the goals of surgery should be to place the digit in a more functional and extended position, while maintaining finger flexion.

CURRENT DEFINITION

Congenital proximal interphalangeal (PIP) joint contracture is often referred to as camptodactyly. The term camptodactyly stems from a Greek word translated as bent finger.[1] Its first description was likely by Tamplin in 1846, whose *Lectures on the Nature and Treatment of Deformities* described a congenital flexion contracture.[1] Still, the term camptodactyly was not applied until 1906, when Landouzy used it to describe young girls with fixed flexion deformities of the PIP joint.[2]

The current accepted definition of camptodactyly is a nontraumatic finger PIP joint flexion deformity, often occurring bilaterally. Patients present in a bimodal age distribution during periods of rapid growth. Younger patients, less than 2 years old, are classified as having infantile camptodactyly, whereas patients greater than 10 years old are classed as having

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Department of Orthopedic Surgery, Washington University School of Medicine, 660 South Euclid Avenue, Campus Box 8233, St Louis, MO 63110, USA
* Corresponding author.
E-mail address: goldfarbc@wustl.edu

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adolescent camptodactyly. Camptodactyly has been described in conjunction with numerous other congenital anomalies. Difficulty in identifying a unifying cause and treatment recommendation has left the full characterization of the condition controversial.

CAUSE

Despite the vast array of descriptions, theories, and pathologic structures implicated in camptodactyly, the exact cause remains debatable. Some of the earliest reports on the condition by Landouzy in 1906 identified tuberculosis and rheumatic disorders to be the underlying cause of this disorder.[3] In 1954, Oldfield[2] attributed his findings to sluggish peripheral circulation found predominantly in young women. By the late 1960s, Courtemanche[3] and Smith and Kaplan[4] described a nontraumatic flexion deformity of the PIP joint without evidence of circulatory compromise. Courtemanche[3] additionally noted volar skin tightness with passive extension, as well as abnormal insertion of the fourth lumbrical in 2 of 3 cases.[3] McFarlane and colleagues[5] found similar abnormalities in the insertion of the fourth lumbrical in 21 consecutive cases with absence or diminished appearance of the palmar interossei in a small subset of patients. In a later study, he had accumulated a total of 74 consecutive cases, all with fourth lumbrical anomalies or complete absence (4%).[6] Siegert and colleagues[7] identified only 2 lumbrical abnormalities in 17 cases, suggesting that the inability of the volar skin to stretch during rapid growth was the primary pathologic cause of the deformity. Ogino and Kato[8] described a hypoplastic flexor digitorum superficialis (FDS) tendon with no continuity of the distal end with the proximal muscle in 5 of 6 patients, with others identifying tightness or slow retraction of the FDS tendon to be the cause.[4] In addition to implicating many of these structures, Smith and Grobbelaar[9] further identified adherence of the lateral bands to the proximal phalanx as a cause of loss of extension force. Congenital absence of the extensor mechanism or central slip have been described,[10] and extensor mechanism anomalies were implicated as the primary cause of camptodactyly by Koman and colleagues.[11] Smith and Kaplan[4] succinctly summarized that nearly every anatomic structure at the base of the finger has been implicated as the deforming disorder in camptodactyly.

Later publications acknowledged the heterogeneous causes, unifying the description as an imbalance in the flexion and extension forces acting on the PIP joint. Although it is possible that a single structure may be the primary cause for the development of this condition, at presentation there are often multiple structures involved in the pathogenesis of the disease, given that these abnormal forces lead to secondary changes and deformities. Thus, when considering surgical intervention, many have advocated a stepwise assessment of all potentially involved structures (skin, fascia, the FDS tendon, lumbricals, interossei, lateral bands, volar plate, accessory collateral ligaments, joint surfaces, and central slip insertion).[9,12,13]

CLINICAL PRESENTATION

The true incidence of congenital PIP joint contracture is unknown, but it has been reported to occur in less than 1% of the population.[1,4,9,14–16] It has been noted to affect male and female patients equally[3,9,11,17] but in other studies has been found to affect female patients more commonly.[1,5–8,18,19] Patients present in a bimodal age distribution, with early or infantile presentation occurring before the age of 2 years (**Fig. 1**A, B), and late or adolescent presentation occurring after 10 years of age (**Fig. 1**C, D). Engber and Flatt[1] found patient presentation to occur within the first year of life in 84%, more than the age of 10 years in 13%, with only 3% (2 patients) presenting between the ages of 1 and 10 years. Progression of the PIP joint deformity has been noted until 20 years of age[1]; however, conventional theory describes halting of progression once skeletal growth has ceased.[7] The deformity is painless, and many patients present with a concern over appearance and functional limitations such as difficulty typing, playing a musical instrument, participating in sporting activities, or wearing gloves.

The characteristic presentation is a flexion deformity of the PIP joint. However, many have noted compensatory intrinsic minus posturing of the hand with metacarpophalangeal (MCP) joint hyperextension,[1,3,5] as well as a secondary boutonnière deformity with distal interphalangeal (DIP) joint hyperextension.[12,13] The PIP joint flexion deformity may be fixed or correctable, but there is no natural history evidence that the deformity progresses from a correctable form to a fixed contracture.[6,7,9,16] Likewise, it has not been shown that a fixed deformity represents the end stage of disease. The deformity classically involves the small finger, sometimes