



# CAMPTODACTYLY AND CLINODACTYLY – NEW UNDERSTANDING OF KNOWN DEFORMITIES

Matija Matošević<sup>1</sup>, Lovro Lamot<sup>1,2</sup> and Darko Antičević<sup>3,4</sup>

<sup>1</sup>University of Zagreb, School of Medicine, Zagreb, Croatia;

<sup>2</sup>Department of Pediatrics, University Hospital Centre Zagreb,  
University of Zagreb, School of Medicine, Zagreb, Croatia;

<sup>3</sup>Josip Juraj Strossmayer University of Osijek, Faculty of Dental Medicine and Health, Osijek, Croatia;

<sup>4</sup>Sveta Katarina Special Hospital, Zabok, Croatia

**SUMMARY** – Camptodactyly and clinodactyly are most commonly considered just cosmetic defects, but they can pose a major diagnostic and therapeutic challenge, mainly because of their apparently similar clinical presentation. For years, experts have been arguing over definitions, descriptions, and therapeutic approaches to these deformities, with some favoring surgical approach, some advocating conservative treatment, while others are prone to use a combination of the aforementioned approaches. This article provides an overview of the current literature on two different entities, with emphasis on differences in clinical presentation and treatment modalities. This may improve the understanding and recognition of these deformities in children, and help the attending physician select the most appropriate therapy for the individual patient.

**Key words:** *Camptodactyly; Clinodactyly; Clinical presentation; Therapy*

## Introduction

Although basically both represent a bent finger, camptodactyly and clinodactyly are two etiopathologically disparate disorders that require different diagnostic and therapeutic approaches (Table 1). According to the proposed classification of developmental anomalies, camptodactyly and clinodactyly are malformations causing a defect of formation and/or differentiation in the indeterminate axis, differing in the presence of defect in the soft or bony tissue, respectively<sup>1</sup>.

## Camptodactyly

### *Classification and diagnosis*

The pathogenesis of camptodactyly has not been fully elucidated<sup>2</sup>. Almost all structures surrounding the

proximal interphalangeal joint exhibit abnormalities leading to camptodactyly, while changes in the superficial flexor and lumbrical muscle (4<sup>th</sup> lumbrical muscle) are reported as common (Table 2)<sup>3,4</sup>.

Insertion of the 4<sup>th</sup> lumbrical muscle to the tendon of the superficial flexor muscle of fingers (specifically the 5<sup>th</sup> finger) or to the capsule of the metacarpophalangeal joint is cited as the most common anatomic anomaly in camptodactyly<sup>4</sup>. Due to such abnormal insertion point, hyperextension in the metacarpophalangeal joint and flexion in the proximal interphalangeal joint occurs, resulting in clinical presentation of camptodactyly<sup>4</sup>. Other changes that contribute to and participate in the development of camptodactyly include extensor insufficiency and contractures of the collateral and palmar ligament of the fingers<sup>5,6</sup>. In long-term deformities, bone changes affecting the proximal and middle phalanges have been radiographically identified<sup>3</sup>.

Camptodactyly can also occur in some genetic disorders such as Freeman-Sheldon syndrome and Camptodactyly Arthropathy Coxa Vara Pericarditis (CACP)

Correspondence to: *Lovro Lamot, MD, PhD*, University Hospital Centre Zagreb, Kišpatićeva 12, HR-10000 Zagreb, Croatia  
E-mail: lovro.lamot@gmail.com

Received August 26, 2020, accepted April 28, 2021

Table 1. Comparison of clinical characteristics and therapeutic modalities for the treatment of camptodactyly and clinodactyly

	Camptodactyly	Clinodactyly
Characteristics	Flexural deviation in the proximal interphalangeal joint	Deviation in the radio-ulnar plane, distal to the metacarpophalangeal joint
Distribution	It involves predominantly the 4 <sup>th</sup> and 5 <sup>th</sup> fingers, while the 1 <sup>st</sup> finger is almost always spared	It can involve all fingers, including the 1 <sup>st</sup> finger
Treatment	Splints and exercises can give good therapeutic response, while surgical therapy is recommended for severe forms	Therapy is more often surgical, while splints and exercises have not proved useful

Table 2. Structures deformities of which are most commonly cited as the cause of camptodactyly<sup>4</sup>

Structure	Deformity	Cause of deformity
Superficial flexor muscle of fingers	Hardened, tense or underdeveloped muscle tendon or abnormal muscle tendon origin	Congenital
Lumbrical muscles of hand	Abnormal origin or insertion point	

syndrome<sup>7</sup>. In 2008, Malik *et al.* described a case of a German family from Hessen, in which there were thirteen cases (eight women and five men) of little finger camptodactyly in four successive generations. Based on that study, the authors determined that a possible gene locus for camptodactyly could be located at 3q11.2-q13.12<sup>8</sup>. Couser *et al.* have shown that there is a link between the appearance of camptodactyly and deletion of the 22q11.21 chromosome. Later on, ten cases of children with 22q11.21 deletion presenting with camptodactyly were described in the literature<sup>9</sup>.

Although there have been various definitions of camptodactyly over years, some of which share common characteristics, camptodactyly is usually considered to be a progressive, non-traumatic flexion deformity that most commonly affects proximal interphalangeal joints of the 4<sup>th</sup> and 5<sup>th</sup> fingers. It can involve more than one finger, including 2<sup>nd</sup> and 3<sup>rd</sup> finger, while the 1<sup>st</sup> finger is spared<sup>2,3,10</sup>. In 1994, Benson *et al.* divided camptodactyly into 3 types, as shown in Table 3<sup>3,11</sup>.

In addition to Benson *et al.* classification, Foucher *et al.* proposed, in the same year, to further divide types I and II into types Ia and Ib (early correctable and early stiff) and IIa and IIb (late correctable and late stiff)<sup>5</sup>. Photos show typical clinical appearance of the 5<sup>th</sup> finger bilateral camptodactyly in a 4-year-old boy. Dorsal view is shown in Figure 1a, palmar view in Figure 1b, while preoperative measurements are shown in Figure 1c. This is an example of type III camptodactyly according to Benson *et al.* classification modified by Foucher *et al.* (Fig. 1).

Carefully taken medical history and thorough clinical examination are essential for the diagnosis of camptodactyly. It should be noted that camptodactyly

Table 3. Benson *et al.* classification of camptodactyly, with Foucher *et al.* additions<sup>3,5,11</sup>

	Type I	Type II	Type III
Localization	Unilateral	Unilateral	Bilateral
Onset period	Infancy	Adolescence	Congenital
Affected group	Affects male and female children equally	Affects female children more than male	Affects male and female children equally
Presentation	Contracture of the proximal interphalangeal joint of the 5 <sup>th</sup> finger; further divided into type Ia (stiff) and Ib (correctable)	Same as type I with further division into type IIa (stiff) and IIb (correctable)	Severe contractures affecting multiple fingers on both hands, often with other birth defects



*Fig. 1. A 4-year-old boy presenting with 5<sup>th</sup> finger camptodactyly on both hands; dorsal view (a); palmar view (b); preoperative measurements (c).*

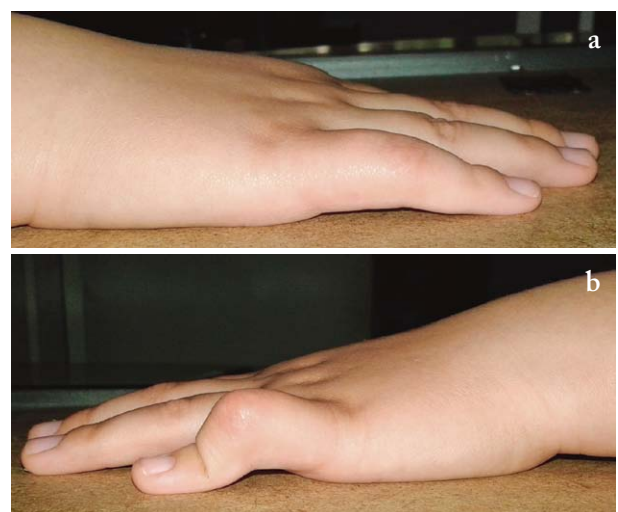
progresses slowly, most commonly is isolated, and it is not related to trauma, inflammation or visible lesions. On differential diagnosis, it is important to rule out disorders similar to camptodactyly such as trigger finger (stenosing tenosynovitis), juvenile palmar fibroma-

tosis, Dupuytren's disease, boutonnière deformity, inflammatory arthritides, symphalangism, arthrogryposis, and pterygium syndrome. When examining a patient with camptodactyly, it is important to determine active and passive range of motion of the proximal interphalangeal joint and tension of the superficial flexor muscle of hand tendon<sup>3</sup>.

In 1988, after 27 years of research, Foucher *et al.* proposed an algorithm to diagnose and select optimal treatment for camptodactyly<sup>5</sup>. The algorithm is based on clinical examination and consists of 6 tests: active proximal interphalangeal joint extension with wrist in neutral, dermodesis test, flexor tenodesis test, metacarpophalangeal joint flexion test (the Bouvier maneuver), functional flexor digitorum superficialis test, and central band or extensor tenodesis test<sup>5</sup>.

#### *Treatment options*

Treatment for camptodactyly may be conservative (non-surgical) or non-conservative (surgical), and the choice depends on the severity of the contracture<sup>3</sup>. If the contracture is less than 60 degrees, conservative therapy is recommended<sup>10</sup>. Non-surgical therapy involves the use of passive or dynamic splints, or a combination of both. Rhee *et al.* suggest a stretching protocol that includes 5 minutes of passive stretching 20 times a day until the contracture is corrected, with additional exercises 5 to 10 times a day<sup>12</sup>, while Benson *et al.* propose wearing static splints for 15-18 hours a day<sup>3,11</sup>. On the other hand, Hori *et al.* propose the us-



*Fig. 2. A 7-year-old boy (from Figure 1) at two-year postoperative follow-up: right hand (a); left hand (b).*

age of a dynamic splint 24 hours a day for the first few months of treatment, followed by using splints for 8 hours a day<sup>13</sup>. However, upon discontinuation of wearing the splint, it has been shown that contracture has a tendency to return<sup>3,6</sup>. According to Foucher *et al.* algorithm, three outcomes of treating splinters are possible, as follows: achieving good complete mobility, passive improvement, and no significant improvement. In the case of passive improvement or no significant improvement, Foucher *et al.* suggest surgical intervention<sup>5</sup>. In surgical approach, it is important to capture and repair any pathologic changes, not just to repair the contracture as such<sup>6</sup>. Postoperative care and rehabilitation, as well as patient co-operation, are crucial for a favorable outcome of surgery<sup>3,6</sup>. Figure 2 shows clinical result in the patient from Figure 1 at two-year follow-up. Soft tissue release was performed with the addition of some surgical solutions proposed by Foucher *et al.*<sup>5</sup>. The patient's right hand as the dominant hand was treated first (Fig. 2a). Left hand has been scheduled for surgery, but had to be postponed due to COVID-19 pandemic. Left hand is shown for comparison (Fig. 2b).

The risk of flexion loss in the interphalangeal joint and limited improvement in extension in the joint are important factors when deciding whether to take surgical approach to treatment<sup>3</sup>. Given the outcome of conservative therapy, Foucher *et al.* in their algorithm propose several surgical solutions, as follows: 1) modified Malek's operation; 2) Zancolli's 'lasso' procedure and repositioning of lumbrical muscles; 3) transfer of lumbrical muscle or flexor digitorum superficialis muscle onto extensor hood; or 4) transfer of superficial flexor muscle of fingers on medial band<sup>5</sup>. Postoperative recovery involves sustained finger extension for 4 weeks<sup>6</sup>. Finally, it should be emphasized that conservative splint therapy is still the first choice in the treatment of camptodactyly and that the benefits of surgery, which include small improvements of motion and aesthetic correction, are often overshadowed by the risks<sup>3,10</sup>.

## Clinodactyly

### *Classification and diagnosis*

Unlike camptodactyly, clinodactyly occurs bilaterally as radial deviation of the little finger, but can also

Table 4. Classification of clinodactyly<sup>3,22,23</sup>

Burke and Flatt	Familial clinodactyly Clinodactyly related to other congenital abnormalities Clinodactyly due to epiphyseal injuries Clinodactyly associated with triphalangeal thumb
Cooney	Simple clinodactyly (affects the bone) Complex clinodactyly (affects the bone and surrounding soft tissue) Complicated clinodactyly (finger curvature greater than 45°)
Ali <i>et al.</i>	Group one (<5°) Group two (5-10°) Group three (15-30°) Group four (>30°)

affect the thumb and ring finger. The curvature of the finger up to 10 degrees is tolerated as normal, while the upper limit is 15 degrees<sup>3</sup>, although some authors believe that this limit can be moved to up to 20 degrees<sup>14,15</sup>. As an autosomal dominant disease with variable expression, clinodactyly occurs mainly as an isolated deformity. Nevertheless, it can also be associated with more than 60 different syndromes, such as Down's, Klinefelter, Turner, Apert, Andersen-Tawil and Kirner syndrome, as well as with familial brachydactyly and symphalangism<sup>3,16-19</sup>. The most common change observed in clinodactyly is an abnormal shape of the middle phalanx<sup>3</sup>. The reason for this is the longitudinally placed epiphysis which extends along the phalanx, due to which the phalanx can have a trapezoidal or triangular shape. In its most severe form, the C-shaped epiphysis encloses the phalanx and forms a 'bracket', thus preventing longitudinal growth of the phalanx. This form of phalanx is known as delta phalanx<sup>3,14</sup>. The aforementioned 'bracket' is radiologically visible after the age of three years. Clinodactyly can also be caused by trauma, fracture, frostbite, inflammatory arthritis, and tumors, all of which are factors that can affect normal development of the growth plate<sup>3,20</sup>. Clinodactyly of the 5<sup>th</sup> finger itself is not a cause of functional disorders if deviation of the phalanx is less or equal to 10 degrees. In this case, such flexion change can be compensated for by abduction of the affected finger. Larger deviations present a problem in certain





*Fig. 3. A two-year-old boy presenting with clinodactyly of the fifth finger bimanually (a); right hand close-up for better visualization (b).*

activities, such as playing a musical instrument or working on a keyboard, however, most patients seek medical attention for aesthetic reasons<sup>3,14,15,21</sup>. In patients with clinodactyly of the thumb, flexion and pinch present a significant clinical problem<sup>3</sup>. There are three different classifications of clinodactyly, each with its advantages and disadvantages (Table 4).

Burke and Flatt propose classifying clinodactyly into four groups: 1) familial; 2) related to other congenital anomalies; 3) related to epiphysial injuries; and 4) related to the triphalangeal thumb<sup>20,22</sup>. This classification is useful in differential diagnosis but is not suitable for determination of appropriate treatment<sup>6</sup>. The most commonly used classification has been suggested by Cooney; this classification takes into account the

curvature of the phalanx and involvement of the surrounding tissue<sup>3</sup>. It distinguishes between simple (in which only the bone is affected) and complex clinodactyly (in which, in addition to bone, the associated soft tissue is also affected), and each of them can be further classified as complex (if the curvature of the phalanx is greater than 45 degrees)<sup>3,5,23</sup>. A more recent classification is proposed by Ali *et al.*<sup>23</sup>, based on the severity of angular deformation, which is divided into four groups: the first group includes physiological curvature up to 5 degrees, the second mild curvature (5-10 degrees), the third moderate curvature (15-30 degrees), and the fourth group severe curvature (greater than 30 degrees)<sup>3,23</sup> (Fig. 3). Clinical appearance of bilateral clinodactyly of mild degree according to classification by Ali *et al.*<sup>23</sup> is shown in Figure 3a, with more detail in the right hand of the same patient shown in Figure 3b.

#### *Treatment options*

As with camptodactyly, treatment of clinodactyly can also be divided into surgical and non-surgical. Non-surgical treatment involves observation and follow-up, as the use of splints and passive stretching has no effect on clinodactyly<sup>3</sup>. Such an approach is possible in patients with smaller phalangeal curvature, whereas surgical treatment is indicated in patients with phalangeal curvature greater than 20 degrees<sup>21</sup>. Surgical procedures used in the treatment of clinodactyly involve four types of osteotomy (opening wedge, closing wedge, partial excision greenstick (PEG) and reverse wedge osteotomy), and physiolysis<sup>3</sup>. In children under the age of six years, physiolysis is recommended because it is simple and fast, and allows for natural correction of phalangeal curvature through growth after removal of the abnormal epiphysis<sup>15,21</sup>. In children older than six years, osteotomy is the treatment of choice, especially closing wedge osteotomy, as it is a simple procedure and provides excellent correction of deformities<sup>21,23</sup>. However, this osteotomy is not recommended in children who have brachydactyly due to additional shortening of the finger, which is why it is necessary to wait for the growth of the finger to be completed<sup>21</sup>. Opening wedge and reverse wedge osteotomies prolong phalangeal length but are limited by soft tissue damage<sup>15,21</sup>. Surgical intervention is not recommended for such patients as it carries the risk of losing the range of motion and unnecessary scarring.

## Conclusion

This article describes differences in clinical presentation and therapeutic approach to camptodactyly and clinodactyly. Camptodactyly is a flexion deformity of unexplained origin that responds well to conservative treatment if the deformity does not exceed 60 degrees; if it does, it is recommended to select an appropriate surgical procedure according to Foucher algorithm. Clinodactyly is an autosomal dominant disease of variable expression characterized by radial deviation of the affected finger. Since conservative treatment shows no effect, surgical approach is warranted. One of the four types of osteotomy is recommended for children above the age of six years. As for children under the age of six years, physiolyysis is recommended, mostly because of easier recovery. It is important to emphasize that both deformities are an aesthetic problem, which is important to consider when selecting therapeutic approach.

## Acknowledgment

We would like to thank Karla Matošević Schaller, Ivan Banovac, MD, and Christian Torbica for providing technical support and much appreciated criticism in making of this paper.

## References

- Manske PR, Oberg KC. Classification and developmental biology of congenital anomalies of the hand and upper extremity. *J Bone Jt Surgery-American Vol* [Internet]. 2009 Jul;91(Suppl 4):3-18. doi: 10.2106/JBJS.I.00008
- Wang AMQ, Kim M, Ho ES, Davidge KM. Surgery and conservative management of camptodactyly in pediatric patients: a systematic review. *Hand* [Internet]. 2019 Mar 22;155894471983465. doi: 10.1177/1558944719834654
- Tse R, Ljungquist K. Camptodactyly and clinodactyly. In: *Congenital Anomalies of the Upper Extremity* [Internet]. Boston, MA: Springer US; 2015. p. 209-17. doi: 10.1007/978-1-4899-7504-1\_16
- McFarlane RM, Curry GI, Evans HB. Anomalies of the intrinsic muscles in camptodactyly. *J Hand Surg Am* [Internet]. 1983;8(5):531-44. doi: 10.1016/S0363-5023(83)80120-8
- Foucher G, Lorea P, Khouri RK, Medina J, Pivato G. Camptodactyly as a spectrum of congenital deficiencies: a treatment algorithm based on clinical examination. *Plast Reconstr Surg* [Internet]. 2006 May;117(6):1897-905. doi: 10.1097/01.prs.0000218977.46520.55
- Smith PJ, Grobbelaar AO. Camptodactyly: a unifying theory and approach to surgical treatment. *J Hand Surg Am* [Internet]. 1998 Jan;23(1):14-9. doi: 10.1016/S0363-5023(98)80082-8
- Madhusudan S, Gupta A, Prakash M, Matta D, Suri D, Singh S. Camptodactyly-arthropathy-coxa vara-pericarditis (CACP) syndrome: a mimicker of juvenile idiopathic arthritis. *Scand J Rheumatol* [Internet]. 2016 Jan 2;45(1):77-8. doi: 10.3109/03009742.2015.1085085
- Malik S, Schott J, Schiller J, Junge A, Baum E, Koch MC. Fifth finger camptodactyly maps to chromosome 3q11.2-q13.12 in a large German kindred. *Eur J Hum Genet* [Internet]. 2008 Feb 14;16(2):265-9. doi: 10.1038/sj.ejhg.5201957
- Couser NL, Pande CK, Walsh JM, Tepperberg J, Aylsworth AS. Camptodactyly and the 22q11.2 deletion syndrome. *Am J Med Genet Part A* [Internet]. 2017 Feb;173(2):515-8. doi: 10.1002/ajmg.a.38029
- Evans BT, Waters PM, Bae DS. Early results of surgical management of camptodactyly. *J Pediatr Orthop* [Internet]. 2017; 37(5):e317-20. doi: 10.1097/BPO.0000000000000967
- Benson LS, Waters PM, Kamil NI, Simmons BP, Upton J. Camptodactyly: classification and results of nonoperative treatment. *J Pediatr Orthop* [Internet]. 1994;14(6):814-9.
- Rhee SH, Oh WS, Lee HJ, Roh YH, Lee JO, Baek GH. Effect of passive stretching on simple camptodactyly in children younger than three years of age. *J Hand Surg Am* [Internet]. 2010;35(11):1768-73. doi: 10.1016/j.jhsa.2010.07.032
- Hori M, Nakamura R, Inoue G, Imamura T, Horii E, Tanaka Y, *et al.* Nonoperative treatment of camptodactyly. *J Hand Surg Am* [Internet]. 1987 Nov;12(6):1061-5. doi: 10.1016/S0363-5023(87)80112-0
- Piper SL, Goldfarb CA, Wall LB. Outcomes of opening wedge osteotomy to correct angular deformity in little finger clinodactyly. *J Hand Surg Am* [Internet]. 2015;40(5):908-913.e1. doi: 10.1016/j.jhsa.2015.01.017
- Medina JA, Lorea P, Elliot D, Foucher G. Correction of clinodactyly by early physiolyysis: 6-year results. *J Hand Surg Am* [Internet]. 2016 Jun;41(6):e123-7. doi: 10.1016/j.jhsa.2016.02.006
- Fairbank SM, Rozen WM, Coombs CJ. The pathogenesis of Kirner's deformity: a clinical, radiological and histological study. *J Hand Surg (European Vol)* [Internet]. 2015 Jul 30; 40(6):633-7. doi: 10.1177/1753193414551911
- Adeb M, Hayashi D, Kumar Y. Kirner's deformity of the fifth finger: a case report. *BMC Musculoskelet Disord* [Internet]. 2016 Dec 21;17(1):440. doi: 10.1186/s12891-016-1300-8
- Song J, Luo S, Cheng X, Yue D, Zhu W, Lin J, *et al.* Clinical features and long exercise test in Chinese patients with Andersen-Tawil syndrome. *Muscle Nerve* [Internet]. 2016 Dec;54(6): 1059-63. doi: 10.1002/mus.25169
- Salazard B, Casanova D. La main du syndrome d'Apert: stratégie thérapeutique. *Chir Main* [Internet]. 2008 Dec;27(SUPPL. 1):S115-20. doi: 10.1016/j.main.2008.07.014 (in French)

20. Burke F, Flatt A. Clinodactyly – a review of a series of cases. *Hand* [Internet]. 1979 Oct 4;os-11(3):269-80. doi: 10.1016/S0072-968X(79)80049-2
21. Goldfarb CA, Wall LB. Osteotomy for clinodactyly. *J Hand Surg Am* [Internet]. 2015 Jun;40(6):1220-4. doi: 10.1016/j.jhsa.2015.03.003
22. Flatt AE. The troubles with pinkies. *Baylor Univ Med Cent Proc* [Internet]. 2005 Oct 11;18(4):341-4. doi: 10.1080/08998280.2005.11928094
23. Ali M, Jackson T, Rayan GM. Closing wedge osteotomy of abnormal middle phalanx for clinodactyly. *J Hand Surg Am* [Internet]. 2009 May;34(5):914-8. doi: 10.1016/j.jhsa.2009.01.007

#### Sažetak

### KAMPTODAKTILIJA I KLINODAKTILIJA – NOVO SHVAĆANJE POZNATIH DEFORMITETA

*M. Matošević, L. Lamot i D. Antičević*

Kamptodaktilija i klinodaktilija se uglavnom promatraju samo kao kozmetički nedostaci, no oni predstavljaju i velik dijagnostički te terapijski izazov, ponajviše zbog svoje naoko slične kliničke slike. Godinama se stručnjaci spore oko definicija, opisa, ali i terapijskih pristupa ovim deformitetima. Dok jedni prednost daju kirurškom pristupu, drugi zagovaraju konzervativno liječenje, a treći prednost daju kombinaciji prethodno navedenih pristupa. Stoga smo odlučili napraviti pregled literature o ova dva različita entiteta, s naglaskom na razlici u kliničkoj prezentaciji te pristupu liječenju. Cilj ovoga preglednog rada je lakše razumijevanje i raspoznavanje obaju deformiteta te naposljetku i odabir najprimjerenije terapije za pojedinačnog bolesnika.

Ključne riječi: *Kamptodaktilija; Klinodaktilija; Klinički prikaz; Terapija*