ORIGINAL PAPERS

Adv Clin Exp Med 2014, **23**, 3, 399–402
ISSN 1899–5276

Maciej Urban[1, 3, A–F], Roman Rutowski[1–3, A–F], Józef Urban[3, A–C, E], Piotr Mazurek[3, B, C], Sebastian Kuliński[3, B, C], Jerzy Gosk[1, 3, A–F]

Treatment of Camptodactyly Using Injection of Botulinum Neurotoxin

[1] Department of Traumatology, Clinic of Traumatology and Hand Surgery, Wroclaw Medical University, Poland
[2] Department of Sports Medicine, University of Physical Education of Wroclaw, Poland
[3] Clinic of Traumatology and Hand Surgery, Wroclaw University Hospital, Poland

A – research concept and design; **B** – collection and/or assembly of data; **C** – data analysis and interpretation; **D** – writing the article; **E** – critical revision of the article; **F** – final approval of article; **G** – other

Abstract

Background. Camptodactyly is usually painless, not caused by trauma, often appearing bilaterally, gradually progressive flexion contracture of the proximal interphalangeal joint mainly on the 5th fingers.
Objectives. The aim of the study was to analyze the efficacy injecting botulinum neurotoxin in short muscles of the hand responsible for the contraction of the proximal interphalangeal joint.
Material and Methods. The clinical material consisted of 12 patients (8 women, 4 men) treated with injections of botulinum neurotoxin in 2009–2012. Patients were monitored respectively for 2 weeks, 3 and 6 months and then every six months after the procedure. The observation period after injection of toxin ranged from 18 to 36 months. Our proposed method of treatment is inducing a temporary paralysis of muscles (lumbrical, interosseous) by means of botulinum neurotoxin (Botox).
Results. In the majority (10) of patients an improvement and stabilization was achieved just after one injection and there were no disease progression in subsequent controlled studies. These patients continued treatment with usage of redressing extensive splints. In case of the other two patients it was required to repeat the injections.
Conclusions. The preliminary results obtained are promising. This method of treatment requires further studies and long-term follow-ups every six months until release of symptoms of the disease will be achieved. The operative treatment is reserved for severe deformities (**Adv Clin Exp Med 2014, 23, 3, 399–402**).

Key words: camptodactyly, botulinum neurotoxin, botox, finger contracture.

Camptodactyly is a usually painless, not caused by trauma, often bilateral, gradually progressive flexion contracture of the proximal interphalangeal joint mainly on the fifth fingers. This ailment was initially described by Tamplin in 1846, in his book "Lectures on the Nature and Treatment of Deformities" [1]. The term "camptodactyly", originating from the Greek language, was used by Landuozy in 1906 to describe the irreversible contracture of the fingers on the PIP joint encountered in young females [2].

Camptodactyly usually affects less than 1% of the population so it is not a very common disease [3]. In about two thirds of the cases, the deformity is present bilaterally, although deformation severity can vary [4]. Usually, the degree of contracture (if any plural deformation is present) decreases radially towards the thumb (Fig. 1).

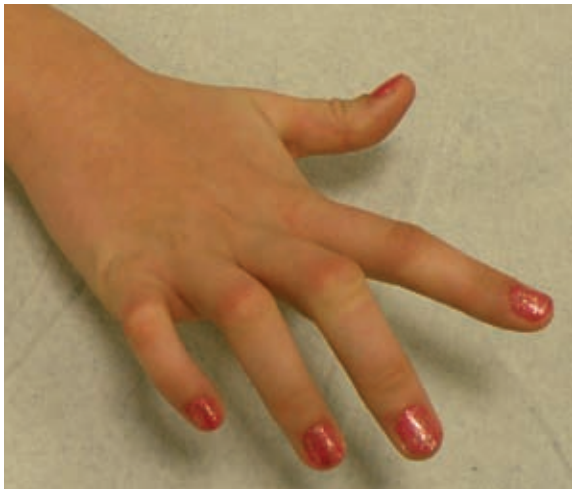

Fig. 1. Clinical picture of camptodactyly – contracture of PIP joints of the fingers 3–5

Clinical camptodactyly is similar to the boutonniere deformity (flexion contracture of the PIP joint) but there is no hyperextension at the distal interphalangeal joint. The metacarpophalangeal joint is usually free, but during the progression of the contracture it could also be affected as a result of compensation. X-rays usually do not show any pathology in the early stages of the disease [3]. The secondary changes in the bone and joint configuration of the PIP joint can develop in response to persistent flexion [3].

Benson et al. proposed the following division of camptodactyly into 3 categories:

I – congenital (infant): present in infancy, limited only to the fifth finger, equally affects males and females;

II – pre-adolescence (acquired): developing normally between 7–11 years of age, progressive form which may transform into a severe deformity of up to 90°; it affects girls more often than boys;

III – associated with a variety of syndromes (syndromic camptodactyly): involves multiple digits of both extremities, severe deformity which occurs in conjunction with craniofacial disorders (orofacial-digital syndrome, craniocarpotarsal dystrophy, oculodentaldigital dysplasia), chromosomal disorders (trisomy 13–15), short stature (mucopolysaccharidosis, camptomelic dysplasia, facial-digital-genital syndrome), and other syndromes (Zellweger syndrome, Blau syndrome, Tel Hashomer camptodactyly syndrome, Jacobsen syndrome, Weaver syndrome and many more) [3, 5–7].

Camptodactyly may develop as a part of over 150 hereditary syndromes [7, 8]. In medical practice there are sometimes cases observed that do not fall into these 3 categories [9, 10]. There are two forms of camptodactyly observed: reversible (flexible) and irreversible (fixed) [3, 11].

Material and Methods

The clinical material consisted of 12 patients (8 women and 4 men) treated with injections of botulinum neurotoxin in 2009–2012. The age of the treated patients varied from 12 to 18 years (median of 15.3 years). In most cases (10) the illness related to the bilateral fifth finger, and in two cases fingers 3–5 were involved. One patient presented a unilateral deformation.

Patients were qualified for the appropriate procedure based on the degree of contracture and the number of fingers with the contraction. For each patient, an individually matched treatment was prepared (the number and location of injections). In each case, the degree of contracture and range of motion was recorded (TAM). Patients were monitored respectively at 2 weeks, 3 and 6 months and then every 6 months after the procedure. Throughout the whole period of monitoring, the patients used a redressive dynamic splint of the PIP joint of the contractured finger. The observation period after injection of the toxin ranged from 18 to 36 months in the case of 11 patients and one patient was monitored for 3 months (median of 27.7 months).

The method of treatment involved injecting botulinum neurotoxin into the short muscles of the hand responsible for the contraction of the proximal interphalangeal joint. For this purpose, 10–25 units of Botox were injected into the head of each interosseous muscle, dorsal and palmar, and the hypothenar muscles of the fifth finger. Injections were made in the fourth interphalangeal space on the dorsal side and on the ulnar side of the hypothenar muscles. In the case of pathological changes in fingers 3 and 4, the injections of botulinum neurotoxin were made in the second and third interosseal space of the hand.

Results

In the majority (10) of patients, an improvement and stabilization was achieved after just 1 injection and there was no disease progression in subsequent controlled measurements (Fig. 2). These patients continued treatment with the use of redressing extensive splints. In the cases of the other 2 patients, it was required to repeat the injections. In the case of one of them, despite 6 consecutive injections, a positive outcome was not achieved. This patient was qualified for surgery because of significant PIP joint contracture due to extra-articular changes and a large transposition of the parts of the extensor mechanism. Surgical treatment consisted of arthrolysis of the joint and reconstruction of extensor apparatus.

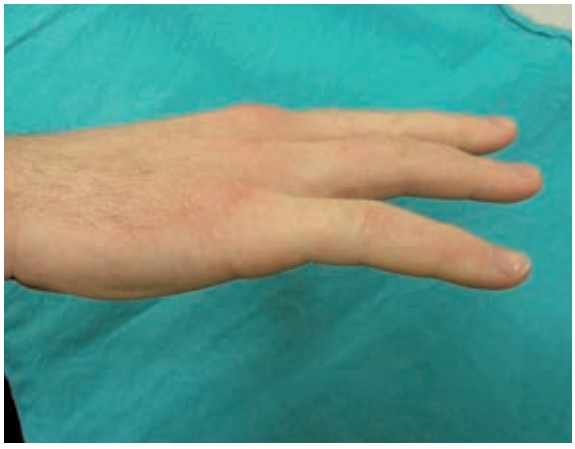

Fig. 2. The result of botulin toxin injection treatment – a decrease of contracture degree in the PIP joint

Discussion

The pathogenesis of camptodactyly still remains unknown [3, 9]. Every structure surrounding the PIP joint may be considered in the etiology of the camptodactyly [3, 9, 12]. It is difficult to establish which part is a primary cause and which pathological changes have only a secondary character [3]. Generally, camptodactyly is caused by a soft-tissue deficit involving many components [9]. These structures include: skin, subcutaneous tissue, collateral ligaments, volar plate, flexor tendons, intrinsic muscles, extensor apparatus and retinacular ligaments [3, 12]. The most prevailing anomalies affect the flexor digitorum superficialis and intrinsic musculature (lumbricals, interossei) [3]. McFarlane et al. found abnormal lumbrical muscle in all 53 cases treated surgically [13]. According to them, abnormalities within the intrinsic system are the principal causes of camptodactyly [13]. The involvement of the intrinsic muscles was confirmed by other authors [3, 9].

The proposed methods of treatment of camptodactyly depend on the type of deformation and degree of advancement. Conservative treatment includes stretching and static or dynamic splinting [3, 14, 15]. Rhee et al. showed the effectiveness of passive stretching without any other form of physiotherapy and splinting in children younger than 3 years of age [15]. Other authors indicate the advisability of prolonged splinting [3, 5, 14, 16]. The recommended duration of immobilization varies from 8 to 24 h a day [3, 5, 14, 16]. Splinting should be long lasting, practically, to achieve skeletal maturity [3].

Operative treatment is reserved for severe deformities [3, 9]. The first indication is the failure of conservative management (splinting) [3, 9]. The second indication for surgery is camptodactyly with flexion contracture of 90 degrees [3, 9]. At the time of surgery, different skin incisions are performed (Glicenstein's plasty, "Z" skin plasty) and full-thickness skin grafts [3]. In clinical practice, the following operating methods are performed: dissection of fascia and fibrous bands, palmar capsulotomy and release of collateral ligaments, release of the flexor tendon sheath and flexor digitorum superficialis, tendon transfer of the flexor digitour superficialis to extensor apparatus, elongation of the flexor digitour superficialis, resection of pathological changes and insertion of the lumbrical or interosseous muscles [13, 17–19]. There are also known attempts at treatment by means of external fixators [3, 20]. Operations carry a risk of neurovascular structure damage, skin loss and exposure of the tendon, also a loss of motion (flexion in PIP) due to scar formation within the tendon sheath [3].

Camptodactyly is difficult to treat [3]. Some authors have proposed algorithms for the treatment of camptodactyly [3, 12]. However, the material presented by various authors usually refers to one treatment method and forms a weak basis for a generalized conclusion.

Botulinum neurotoxin injection may be an interesting alternative to the methods described above. The purpose of this type of treatment is to improve hand function and facilitate rehabilitation [21]. The dose of neurotoxin depends on the muscle mass and muscle activity (active muscle requires a higher dose). For the safety of the patient, treatment should be limited to the specific number of muscles. The maximum dose should not be greater than 6 units/kg body weight [21].

The results of botulinum neurotoxin therapy we obtained are only preliminary and there was a limited number of patients in the study. Considering the improvement in the mobility of the fingers and the degree of reduction of the contracture that we observed, the results of the treatment should be regarded as encouraging.

The use of comprehensive treatment with botulinum neurotoxin in adolescents combined with the appropriate process of rehabilitation should be an alternative option to surgery in adulthood, when it comes to fixed joint contractures. This method can be a significant help in surgery for adults.

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] **Landouzy L:** Camptodactylie. Stigmante precoce du neuroarthritisme. Presse Med 1906, 14, 251–253.

[3] **Kozin SH, Kary SP:** Camptodactyly. In: Green's Operative Hand Surgery. Eds.: Wolfe SW, Hotchkiss RN, Pederson WC, Kozin SH, Churchill Livingstone Elsevier, Philadelphia 2011, 6th ed., 1443–1451.

[4] **Jobe MT, Wright MT:** Congenital anomalies of hand. In: Campbell's operative orthopaedics. Ed.: Crenshaw AH. Mosby Year Book, St. Louis 1992, 3415.

[5] **Benson LS, Waters PM, Kamil NI, Simmons BP, Upton J:** Camptodactyly: Classification and results of nonoperative treatment. J Pediatr Orthop 1994, 14, 814–819.

[6] **Akawi NA, Ali BR, Al-Gazali L:** A novel mutation in PRG4 gene underlying camptodactyly-arthropathy-coxa vara-pericarditis syndrome with the possible expansion of the phenotype to include congenital cataract. Birth Defects Res A Clin Mol Teratol 2012, 94, 553–556. doi: 10.1002/bdra.23031. Epub 2012 Jun 8.

[7] **Malik S, Afzal M, Gul S, Wahab A, Ahmad M:** Autosomal dominant syndrome of camptodactyly, clinodactyly, syndactyly, and bifid toes. Am J Med Genet A 2010, 152A, 2313–2317. doi: 10.1002/ajmg.a.33552.
[8] **Choi BR, Lim YH, Joo KB, Paik SS, Kim NS, Lee JK, Yoo DH:** Camptodactyly, arthropathy, coxa vara, pericarditis (CACP) syndrome: a case report. J Korean Med Sci 2004 Dec, 19, 907–910.
[9] **Hamilton KL, Netscher DT:** Multidigit camptodactyly of the hands and feet: A case study. Hand 2013, 8, 324–329. doi: 10.1007/s11552-013-9497-6.
[10] **Wieczorek D1, Bartsch O, Lechno S, Kohlhase J, Peters DJ, Dauwerse H, Gillessen-Kaesbach G, Hennekam RC, Passarge E:** Two adults with Rubinstein-Taybi syndrome with mild mental retardation, glaucoma, normal growth and skull circumference, and camptodactyly of third fingers. Am J Med Genet A 2009, 149A, 2849–2854. doi: 10.1002/ajmg.a.33129.
[11] **Ozsahin M, Uslu M, Kutlucan A, Baki AE, Ataoglu S:** Bilateral Fifth-Finger Camptodactyly. Am J Phys Med & Rehab 2012, 91, 638. doi: 10.1097/PHM.0b013e318238a14b.
[12] **Foucher G, Loréa P, Khouri RK, Medina J, Pivato G:** Camptodactyly as a spectrum of congenital deficiencies: a treatment algorithm based on clinical examination. Plast Reconstr Surg 2006, 117, 1897–1905.
[13] **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.
[14] **Hori M, Nakamura R, Inoue MD:** Nonoperative treatment of camptodactyly. J Hand Surg 1987, 12A, 1061–1065.
[15] **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 2010, 35, 1768–1773.
[16] **Miura T, Nakamura R, Tamura Y:** Long-standing extended dynamic splintage and release of an abnormal restraining structure in camptodactyly. J Hand Surg Br 1992, 17, 665–672.
[17] **Engeber WD, Flatt AE:** Camptodactyly: an analysis of sixty-six patients and twenty-four operations. J Hand Surg 1977, 2, 216–224.
[18] **Siegert JJ, Cooney WP, Dobyns JH:** Management of simple camptodactyly. J Hand Surg 1990, 15B, 181–189.
[19] **Koman LA, Toby EB, Poechling GG:** Congenital flexion deformities of the proximal interphalangeal joint in children: a subgroup of camptodactyly. J Hand Surg 1990, 15A, 582–586.
[20] **Akoz T, Erdogan B, Gorgu M, Kapucu MR, Girgin O:** Correction of camptodactyly with an external fixator. Eur J Plast Surg 1998, 21, 308–310.
[21] **Domżał TM:** Neurotoksyna botulinowa w praktyce lekarskiej. Czelej, Lublin 2009, 77–87.

Address for correspondence:

Maciej Urban
Department of Traumatology, Clinic of Traumatology and Hand Surgery
Wroclaw Medical University
Borowska 213
50-556 Wrocław
Poland
Tel.: +48 71 73 43 820
E-mail: maciejurban@poczta.onet.pl

Conflict of interest: None declared

Received: 26.11.2013
Revised: 28.04.2014
Accepted: 9.06.2014