

Orthosis for Correction of Bilateral Claw-Hand In Charcot-Marie-Tooth Disease: A Case Report

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ABSTRACT

Charcot-Marie-Tooth (CMT) is a common inherited peripheral nerve disorder. Although lower extremity deformities are more common in patients with CMT, claw-hand deformities and monkey hand deformities are deformities which are also frequently seen. If these deformities are not ameliorated, they cause the impairment of the biomechanics of the hand. Orthoses made of thermoplastic material are often used to counteract these deformities. In this case, the results of the lumbrical bar orthosis applied to the bilateral claw-hand deformity in a patient with CMT are presented. Before and after the usage of the orthosis, dexterity was evaluated with the Purdue Pegboard test, hand grip strength was evaluated with a manual hand dynamometer, and pinch strength was evaluated with a pinchmeter. As a result of the evaluations, it was observed that the lumbrical bar orthosis mended the claw-hand deformity, but it did not provide improvements in hand grip strength, pinch strength and dexterity.

Key words: Charcot-Marie-Tooth, Lumbrical bar orthosis, Claw-Hand.

Charcot-Marie-Tooth Hastalığında Bilateral Pençe-Elin Düzeltilmesi İçin Ortez : Olgu Sunumu

ÖZ

Charcot-Marie-Tooth (CMT) yaygın görülen kalıtsal periferik sinir bozukluğudur. CMT'li hastalarda daha çok alt ekstremitte deformiteleri görülmesine rağmen sıklıkla pençe-el deformitesi ve maymun eli deformiteleri de görülmektedir. Bu deformitelerin düzeltilmesinde sıklıkla termoplastik materyalden yapılan ortezlerden yararlanılmaktadır. Bu vakamızda CMT'li bir hastada meydana gelen bilateral pençe-el deformitesine uygulanan lumbrikal bar ortezinin sonuçları sunulmuştur. Ortez kullanımı öncesi ve sonrası el becerisi Purdue Pegboard testi ile, el kavrama kuvveti manuel el dinamometresi ile pinç kuvveti ise pinçmetre ile değerlendirilmiştir. Yapılan değerlendirmeler sonucunda lumbrikal bar ortezinin pençe-el deformitesini düzelttiği ancak el kavrama kuvveti, pinç kuvveti ve el becerileri üzerinde iyileşmelere sebep olmadığı görülmüştür.

Anahtar Kelimeler: Charcot-Marie-Tooth, Lumbrikal bar ortezi, Pençe-el.

INTRODUCTION

Charcot-Marie-Tooth (CMT) disease is the most common hereditary peripheral nerve disorder seen in the population with an incidence rate of 1/2500 (Skre, 1974). The etiology of CMT is associated with unknown mutations in the homeotic genes involved in neural formation (Newman et al., 2007). Significant demyelination and axonal loss can be seen in nerves due to the mutation. Patients affected by CMT complain of loss of muscle strength and distal sensation in their upper and lower extremities, and the clinical course of the disease/condition; although usually slow, is mostly progressive. In addition to weakness and dysfunction in the hand, two deformities may be seen in patients with CMT: a) claw-hand deformity characterized by hyperextension of the first phalanx and flexion of the other phalanxes after relaxation of the dorsal and interosseous muscles; b) monkey hand characterized by the atrophy of the thenar and hypothenar muscles (Pareyson et al., 2006). In chronic joint deformities, bracing is one of the most commonly used conservative methods to preserve joint mechanics. In orthoses made for the hand, it is aimed to both correct the deformity and increase the functionality (Ceylan İsmail et al., 2022a, 2022b).

The aim of this study was to apply lumbrical bar orthosis to a patient who developed bilateral claw-hand deformity in his hands due to the CMT disease, while investigating changes in deformity, hand functionality and grip strength.

CASE

A 24-year-old female patient with a diagnosis of CMT was admitted to the physical therapy and rehabilitation clinic with complaints of loss of function in both hands and loss of flexible range of motion in extension (Figure 1).



Figure 1. Bilateral active extension

Written and verbal consent was obtained from the patient before starting the study. Joint angles during active finger extension were measured as shown in the table (Table 1).

As a result of the clinical evaluation, it was decided to make a bilateral lumbrical bar orthosis for the patient. The reason why we use the lumbrical bar orthosis is that to stabilize the hand and correct the extension directional deformity. It was aimed to correct the deformity by facilitating the tenodesis reflex with an orthosis made of thermoplastic material. While providing trunk support with the volar part, the deformity was corrected during active extension by applying pressure on the proximal phalanges dorsally. With the velcro support provided from the dorsal metacarp, both the insertion and removal of the device was facilitated and stabilization was achieved (Figure 2-3). The patient was asked to wear the

Table 1. Active finger extension range of motion values

	Joints	Right hand			Left hand		
		MP	PIF	DIF	MP	PIF	DIF
Fingers	2	-30° ext	20° flex	20° flex	-40° ext	30° flex	20° flex
	3	-30° ext	30° flex	20° flex	-35° ext	30° flex	20° flex
	4	-30° ext	30° flex	20° flex	-30° ext	30° flex	20° flex
	5	-20° ext	30° flex	20° flex	-20° ext	30° flex	20° flex
	1	0°	0°		0°	0°	

MP: Metakarpphalangeal; PIF: proksimal interfalangeal; DIF: Distal Interfalangeal; ext: extension; flex: flexion

orthosis intermittently for a minimum of 2 hours and a maximum of 5 hours a day for 6 weeks. The patient was not included in any rehabilitation program during this period.



Figure 2. Lumbrical bar orthosis (lateral)



Figure 3. Lumbrical bar orthosis (anterior)

After correcting the deformity, the extent to which the orthosis affected the patient's hand functions was evaluated. The patient's hand dexterity was measured with the Purdue-Pegboard test, bilateral hand grip strength was measured with a hand dynamometer, and pinch strength was measured with a pinchmeter (Ceylan et al., 2021). The Purdue-Pegboard test is often used to assess fine motor skills in upper extremity problems. The score of the test is determined by the number of pins inserted during the given time period (Genç et al., 2002). The measurements were performed on the day of the orthosis usage, with and without the orthosis (Figures 4-5-6).



Figure 4. Purdue-Pegboard Test

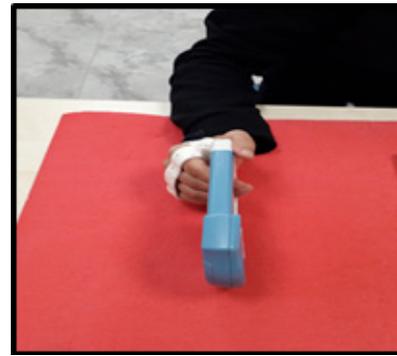


Figure 5. Hand grip strength assessment

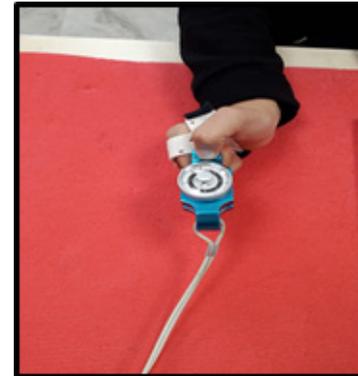


Figure 6. Pinch force assessment

RESULTS

The clinical evaluation results before and after 6 weeks of orthosis usage are given in Table 2. As a result of the evaluations, it was observed that there was no change regarding the obtained scores from the Purdue Pegboard test with the use of orthosis. In addition, reduction in pinch strength and grip strength was observed in both hands.

Table 2. Clinical evaluations before and after orthosis use

		Before orthosis use	After orthosis use
Purdue Pegboard Test	Dominant Hand (score)	7	7
	Non-dominant Hand (score)	8	8
	Both Hands (score)	3	3
	Right + Left + Both Hands	18	18
	Assembly (score)	12	12
Pinch Force	Left (kg)	1.5	1
	Right (kg)	2	1.5
Hand Grip Strength	Left (kg)	7	6.4
	Right (kg)	8	6.8

DISCUSSION

According to our study findings, the lumbrical bar orthosis we applied for the claw-hand deformity in the patient with CMT corrected the deformity. However, it was observed that this orthosis was not effective on dexterity, hand grip strength and pinch strength in an instant manner.

When the literature is examined, it is seen that the conducted studies mostly focus on ankle orthoses in CMT patients. Studies have reported that ankle foot orthosis reduces the frequency of falls (Ramdharry et al., 2012), and night splints result in an improvement in ankle dorsiflexion angle (Uygur et al., 2001). Videler et al. stated that the thumb opposition splint in patients with CMT increases functionality with regards to daily living activities and occupational performance (Videler et al., 2012).

AUTHOR CONTRIBUTION

Idea/Concept: İC; Design: İC, MC; Data Collection and/ or Processing: İC, MC; Analysis and/or Interpretation: İC, MC; Writing the Article: İC, MC.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

FINANCIAL DISCLOSURE

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ETHICAL STATEMENT

Informed consent was obtained from the patient.

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