

The Effect of Concurrent Use of Lower Extremity and Spinal Orthotics Intervention to Improve Activity Daily Living and Quality Of Life in Charcot-Marie-Tooth Disease: A Single-Subject Design

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Abstract

Introduction: The Charcot-Marie-Tooth (CMT) disease causes the significant muscular deficits in the affected patients, inhibits activities daily living, and involves a severe disability. The aim of this study was to evaluate the effect of concurrent use of a knee-ankle foot orthosis along with a body jacket orthosis to improve activities daily living in a person with CMT. **Methods and Materials:** A boy aged 16 years old with CMT was evaluated in this case report. He had a sever Kypho-scoliosis deformity in his spine and sever equines in his feet with unstable knee. In order to control of knees and ankles simultaneously, an ankle foot orthosis with the supracondylar trimline was designed. The control of his sever spinal deformity was also provided by a body jacket over shoulder. To evaluation of activities daily living, subject was asked to filled Barthel index out during a week. After one month, the individual was asked to fill the Barthel index out again. Finally, user friendly of orthotics were evaluated by a satisfaction questionnaire. **Results:** The results indicated that the orthoses was capable to improve activity daily living (25 percent increase after using orthosis in Barthel index). Also, results showed 30 percent increase after using orthosis for the QOL questionnaire. The mean of scale in satisfaction questionnaire were 4.5 based on Likert scale. **Conclusion:** This single-case study demonstrated the control of spinal and lower limb is simultaneously necessary for the improvement of activity daily living.

Key words: ADL, QOF, Charcot-Marie-Tooth, Orthotics And Spinal Deformity

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Introduction

Charcot-Marie-Tooth (CMT) disease termed as a progressive muscular atrophy initiates in the foot-ankle complex which affects the upper limbs after a few years (1). It is not a single disease, but a group of congenital disorder in the proteins of the peripheral nerve myelin sheath(2). The disease was first illustrated in 1886 by the French neurologist Jean Martin Charcot in England(3). Nowadays, a large variety of specific genetic disorders have been identified with the disease(2). Also, it is the most common hereditary peripheral neuropathies throughout world, with about 10 to 28 cases every 100,000 live births. CMT which initiates in late adolescence causes progressive muscle weakness and degenerative below the knee, resulting in equinus foots, and it

gait abnormality and reduction of ADL(4).

Results of Vinci et al. study (2005) demonstrated that People with CMT had lower scales in their quality of life (QOL) questionnaire in all cases than normal people. Since the short form of quality of life questionnaire (SF-36) is a simple and valid tool that can be used for the comparison of CMT patients with patients having other disorders(5). Juliana *et al.* (2013) confirmed that functional capacity of SF-36 also tended to be significantly affected in CMT patients (6).

Due to the challenging outcomes of surgical intervention, Rehabilitation has become as a key treatment. Orthotics is one of rehabilitation treatments used in patients with CMT (7). AFOs have been proposed as a fundamental intervention in the rehabilitative care of CMT(8). AFOs are commonly prescribed in CMT disease to improve walking quality and reduce the risk

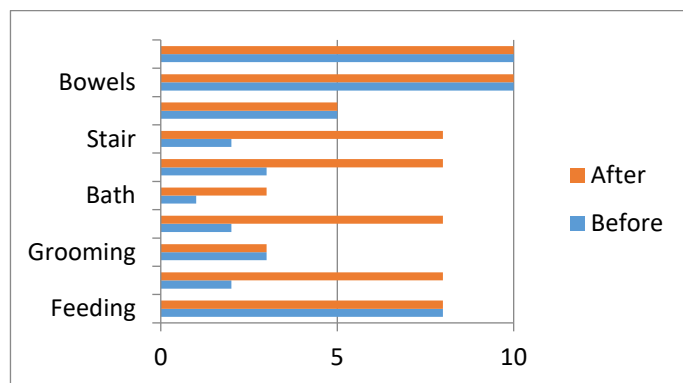


Chart 1. Shows the impact of Orthotics on ADL

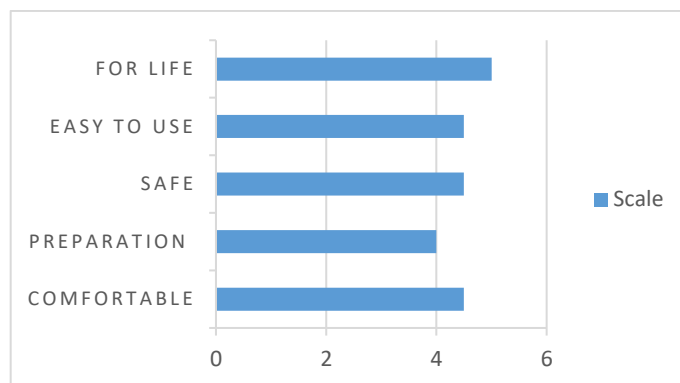


Chart 2. Shows scale for each question

of falling due to the foot drop (9). Bean *et al.* (2001) evaluated the effect of an AFO on submaximal aerobic performance and submaximal perceived exertion in a participant with CMT. Their findings showed the AFO can enhance physiologic performance (10).

There are several studies that evaluated the effect of orthotics devices on gait and performances in CMT patients, but no studies found that evaluated the effect of spinal and lower limb orthosis on activity daily living (ADL) and QOL simultaneously (7, 8, 10). Purpose of present study was to evaluate the concurrent use of a knee-AFO and a body jacket orthosis on ADL and QOL in a person with CMT.

Methods and Materials

Subject's description:

Participant in this study was a 16-year-old boy with 12 years history of CMT. He had the severe scoliosis and kyphosis deformities. The scoliosis pattern of was right thoracic (Cobb angle=63°) and left lumbar (Cobb angle=64°) curves. Figure 1 showed radiography of spinal deformities in the subject. Also, his feet were in the severe equinus (plantar flexion, supination and in toeing) with gastrocnemius spasticity in 3 level according modified Ashworth scale. His knees were unstable in medial-lateral directions so that he was not able to stand without any assistances. Because of the severe deformity in subject's spinal and ankles, his trunk was not in an upright position and his soles were not fully in contact with the floor. So, he needed to an assistance for standing and walking.

Orthotics construction:

Lower limbs part

In order to control knees and ankles simultaneously, a long-leg brace was necessary. Since participant had several disabilities,

the use of long brace similar to a knee ankle foot orthosis (KAFO) can cause the high restriction and early fatigue for the subjects. So, an ankle foot orthosis (AFO) with supra condylar trim line (knee-AFO) was constructed. This custom molded orthosis was designed for both leg. The orthosis could be fitted in his shoelaces. Figure 2 shows the designed knee-AFO for this case report.

In order to control severe deformity in spinal, a **custom molded** thermoplastic (4 millimeter) body jacket over his shoulders was constructed. A thoracic pad was located on scapula in right direction to prevent winging scapula, and a pad was set in sternum to prevent the deformity progression. Orthosis molded in the upright position to control kyphosis deformity. Figure 3 shows the orthotics device used by the subject.

Evaluation

To verify the performance orthosis before and after using, modified Barthel index (in ten items) was assessed for ADL (11, 12). At first, items of Barthel index and QOL-physical function were explained for participant and his family. For QOL, the SF-36 was used which consists of ten subscales that are physical function (PCS-36) scores. Then he filled those in during a week, while he had not any assistive devices. After using knee-AFO and body jacket orthotics for one- period, he filled Barthel index and QOL- physical function.

Finally, user friendly of orthosis was assessed by satisfaction questionnaire. The participant was asked to fill out a questionnaire of satisfaction (Table 3 in appendix part). The questionnaire included five items which participants could answer questions according to Likert scale (13), a 6-point Likert scale (14), where each item was scored from 0 (orthotics were undesirable for the addressed aspect) to 5 (orthotics were excellent for the addressed aspect). A score of 2.5 was selected as the desired threshold (13).



Figure 1. Deformity of spinal showed, scoliosis deformity, Frontal view showed 63 degree of right thoracic curve and 64 degree of left lumbar curve (left figure). Sagittal view showed 94 degree of kyphosis angle (right figure)



Figure 2. Knee-AFO without foam in posterior shell to reduce spastic gastrocnemius muscle (15).

Results

ADL

Results for Barthel index was obtained by different percent before and after using orthosis. Results showed increase 25% after using the orthosis compare to before the orthosis in Barthel index (without orthosis: 46 and with orthosis: 71). Chart 1 shows changes to each item.

QOL questionnaire (physical function)

Results for QOL (10 items) questionnaire was obtained by different percent before and after using orthosis. Results showed 30% increase after using the orthosis compare to before the orthosis in QOL questionnaire (physical function).



a. frontal view

b. sagittal view

Figure 3. Shows participant stand up with orthosis and without any other assistance

Satisfaction

According to the results, participant considered a score higher than 2.5% for each item. Also, the mean of all questions was 4.5 based on Likert scale. Chart 2 reveals the results of each question.

Discussion

Purpose of this single-case study was to evaluate a knee-AFO along with a body jacket orthosis in order to assist the ADL and the physical function of QOF in a person with CMT. The knee-AFO can prevent ankle plantar flexion thereby stabilizing knees. Results of Barthel index demonstrated that orthotics device was effective to increase ADL (sum of scales increased from 48 to 75). Total Barthel score of -20 suggests total dependence, 21-60 severe dependence, 61-90 moderate dependence, and 91-99 slight dependence. Since score 60 is considered as an acceptable threshold in the Barthel Index (11), it can be stated that orthotic intervention in the participant has achieved an acceptable level in the Barthel Index. Also, results of QOF- physical function demonstrated the increased QOF after using the orthotics. It seems that orthotics devices could improve of trunk and ankle alignments to more optimal position thereby improving the subjects' ability. So, QOL has probably increased due to better alignment in legs and trunk.

Results of satisfaction questionnaire demonstrated that orthotics intervention was considered as an user-friendly criteria. Results confirmed that participant preferred to keep on the use of orthotics. In fact, it can be stated that controlling the trunk upright position and controlling ankle plantar flexion simultaneously can increase the patient's functional level. Also, supporting the ankles and trunk resulted in safer feeling during standing.

The limitation of this study was the lack of gait analysis tests to evaluate kinetic and kinematic changes during the gait. In future studies, it is suggested this orthotics device is performed on more participants with CMT.

Conclusion

To improve the ADL and QOL, the potential benefits of orthotics were achieved on a subject with CMT. This single-case study demonstrated that trunk control and lower limbs is simultaneously necessary for improvement of the ADL and QOL.

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Conflict of interest:

None

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Authors' contributions:

Both authors made substantial contributions to the conception, design, analysis, and interpretation of data.

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Appendix

Table 1. Satisfaction questionnaire (13)

NO	Questions	Score: 0-5
1	Do you feel comfortable with the orthosis?	
2	Is the process of preparation comfortable for you?	
3	Do you feel safe with the orthosis?	
4	Has been easy to learn how to use it?	
5	Would you use the orthosis in your daily life activities?	

Table 2. Barthel index

No		Unable	Attempts But Unsafe	Moderate Help	Minimal Help	Fully Independent
1	Feeding	0	2	5	8	10
2	Chair/bed transfers	0	3	8	12	15
3	Grooming	0	1	3	4	5
4	Toilet	0	2	5	8	10
5	Bathing	0	1	3	4	5
6	Walking	0	3	8	12	15
7	Stairs	0	2	5	8	10
8	Dressing	0	2	5	8	10
9	Bowels	0	2	5	8	10
10	Bladder	0	2	5	8	10

Table 3. QOL questionnaire (physical function)

No	Item		Answer	
1	Vigorous activities, such as running, lifting heavy objects, participating in strenuous sports	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
2	Moderate activities, such as moving a table, pushing a vacuum cleaner, bowling, or playing golf	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
3	Lifting or carrying groceries	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
4	Climbing several flights of stairs	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
5	Climbing one flight of stairs	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
6	Bending, kneeling, or stooping	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
7	Walking more than a mile	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
8	Walking several blocks	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
9	Walking one block	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all
10	Bathing or dressing yourself	Yes	, Limited a lot Yes	Limited a Little No Not Limited at all