Case report

Lower extremity muscles activity in standing and sitting position with use of sEMG in patients suffering from Charcot–Marie–Tooth syndrome

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A B S T R A C T

There is very limited, evidenced data about movement possibilities in patients with high level of lower limb muscles atrophy and fatigue in patients suffering from Charcot–Marie–Tooth syndrome. Patient (age 46) suffering from Charcot–Marie–Tooth disease for 30 years with multiple movement restrictions and muscles atrophy above knees took part into the study. Tests were performed for 8 muscles of the lower limb and pelvis. Muscles electrical activity was tested in sitting and standing position (for knees extended and hyperextended). In the right leg rectus femoris, vastus lateralis obliquus, glutaeus medius and semitendinosus muscles activated at first and were working the longest time. The highest activity was observed in standing position with knees extended. In the left leg rectus femoris and biceps femoris muscles activated at first and biceps femoris was working the longest time. Activity level in left lower limb is much lower than in the right one. Muscles weakness is asymmetric. Leg is much weaker and engages antagonists and synergists muscles to compensate weaker rectus femoris, vastus medialis obliquus and vastus lateralis obliquus.

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1. Introduction

Charcot–Marie–Tooth disease is an hereditary motor and sensory neuropathy and is a demyelinating kind of disease. The most frequent form of Charcot–Marie–Tooth type1A (CMT1A) is diagnosed in about 70% of all Charcot–Marie–Tooth patients [1]. CMT1A form is caused by duplication of the short arm of 17 chromosome of gene encoding peripheral myelin protein 22. Protein 22 is largely used in myelin formation, building of Schwann cells, axons and is important for axonal transport and cells metabolism. Degeneration in
CMT disease includes mainly the largest and the biggest axons (2). In 90% disease is inherited in autosomal dominant way, rarely autosomal recessive or is X-linked [2,3]. The most important clinical symptoms associated with the Charcot-Marie-Tooth disease are distal muscles weakness (calf muscles and intrinsic fingers extensors). Calf muscles weakness and degeneration causes flat and characteristic steppage gait. Proprioception and vibration are deteriorated. Patients usually do not need a wheelchair, however gait is disordered and limbs are deformed (gait with knees hyperextended [Fig. 1], foot drop in swing, pes cavus) [4–6]. Pain and low endurance of the muscles lead to a lot of secondary limitations such as: decreasing of cardiopulmonary efficacy, increase of the energetic cost during every-day functioning, depression [7]. There is lack of effective drug treatment [2]. One of the most important treatment is rehabilitation, surgical treatment of skeletal and muscular deformations and therapy of the symptoms. Patient requires to be treated in multidisciplinary team with a lot of specialists who respect all of the limitations, and are able to handle with psychosomatic problems represented by this group of patients [3]. Most of the research with CMT1A patients include usually group of patients with no serious but only moderate limitations. This patients keep active range of motion in the ankles and are able to stay with active knee extension [4,6,7]. There are no evidenced databases with patients who lost ankles active range of motion and where muscles and nerves degeneration includes muscles above the knees (the reason of knees hyperextension what leads to knee pain and develops secondary compensatory mechanisms). Aim of the study was to determine level of lower limb muscles sEMG activity during knee extension in different positions.

2. Materials and methods

2.1. Materials

Test concerned a patient in the age of 46 years (High: 1.76 m, weight 60 kg, BMI: 19.41 kg/m²), who for 30 years has been suffering from Charcot–Marie–Tooth type 1A. For two years patient has underwent physical therapy. As an account patient is very active person in the professionally, can drive a car and walks with use of walking stick. Main restrictions connected with the disease observed during the rehabilitation are: lack of active movement in the ankles, disabled proprioception, calf and thigh muscles atrophy, hyperextension in knees, steppage gait, hyperlordosis, lack of postural control in standing position, abdomen muscles weakness, low efficiency of cardiovascular and pulmonary system, (susceptibility to hyperventilation during training), balance disturbances, painful muscles cramps and legs and back pain.

2.2. Methods

Signals were recorded with use of a 8-channel surface electromyograph TeleM 900 Noraxon connected with MyoResearch System Master Edition. EMG equipment has been made available by Technimex Company from Gliwice. Gel electrodes (Ag/AgCl bars with the diameter of 30 mm) were used according to the SENIAM recommendations. Skin was cleaned before placement of the electrodes above muscles.

2.3. Test organization

In the experiment electrical activity from 8 muscles was taken into consideration: gluteus maximus muscle (GM), gluteus medius muscle (GMed), biceps femoris muscle (BF), semitendinosus muscle (ST), rectus femoris muscle (R), vastus medialis oblique muscle (VMO) and vastus lateralis oblique muscle (VLO). Reference electrode was fixed above the iliac crest. Before the test patient was instructed about the EMG measurement organization, how to prepare to the test and what the repeats look like. Patient was tested once for the right and once for the left lower extremity because of very fast and high risk of hyperventilation and a high level of muscles fatigue. The protocol was created to estimate the work and electrical activity of the muscles in the sitting position (after trunk and pelvis stabilizing) during 3 repeats of the knee extension. Every knee extension was broke by 5 s pauses. The next signals were recorded in standing position with knees extended with the assurance and next in standing position with knees hyperextended with assurance. The entire test (including pauses, 3 repeats of the knee extension in sitting position, knees extended and hyperextended in the standing position), recorded in the protocol lasted 1 min. EMG data were rectified and filtered with RMS algorithm (Root Mean Square). In the test timing formula was used to analyze onset time and
the order of measured muscles. Mean activation amplitude was used to compare qualitative right and left limb and to compare muscles activity parameters in different activity configurations. Following parameters were taken under consideration: muscles timing, mean of the 2 activities in sitting position (third repeat was loaded by EMG artifacts), muscle activity in percent during knee extension in sitting position, mean amplitude of the muscles activity in standing position with knees extended and hyperextended (asymmetry between right and left lower limb).

3. Results

3.1. Muscles order of the onset time

In the right lower limb rectus femoris muscle, vastus lateralis oblique muscle, gluteus medius muscle and semitendinosus muscle activate at first. However in the both analyzed repeats the order of the activity was different. The longest activity belonged to the gluteus medius muscle and the next to the vastus lateralis oblique muscle and to semitendinosus muscle. In the left lower limb rectus femoris muscle activated at first and next biceps femoris muscle (this muscle worked the longest time).

3.2. Qualitative comparison of the mean amplitude data in the right lower limb between sitting position knee activation and standing position with knee extended and hyperextended

During standing with knees extended the highest level of the amplitude of muscles rectus femoris, vastus lateralis obliques and vastus medialis obliques was observed compared to knees hyperextension and extension of the knee in sitting position. In the sitting position gluteals muscles and hamstrings muscles activate stronger than rectus femoris, vastus lateralis obliques and vastus medialis obliques muscles.

3.3. Qualitative comparison of the mean amplitude data in the left lower limb between sitting position knee activation and standing position with knee extended and hyperextended

In the left lower limb gluteus medius muscle activates more than other muscles but the activation of this muscle is high only at the beginning of the knee extension. In the sitting position biceps femoris muscle activates more than the other muscles. Rectus femoris muscle activates the longest time but activity level is very low.

3.4. Asymmetry between right and left lower limb during knee activation in sitting position and in standing position with knee extended and hyperextended

Asymmetry between left and right lower limb in the standing position with knees extended is very high (for every muscle asymmetry exceeds 40%) what is the evidence of the rectus femoris, vastus lateralis obliques and vastus medialis obliques muscles weakness in left lower limb compared to the right limb and higher engagement to the movement of the gluteals muscles. During standing with knees hyperextended lower activity of the rectus femoris, vastus lateralis obliques and vastus medialis obliques muscles in right limb decreases asymmetry level between both legs. In the knee extension in sitting position asymmetry level is the lowest for the vastus medialis obliques muscle and glutaeus maximus muscles, however very high levels of the left biceps femoris activity and left glutaeus medius activity were observed in this test.

4. Discussion

Surface electromyography is wildly used as a tool of the quantitative analysis of movement patterns and therapeutic interventions (gait and posture, involuntary movements). SEMG is also a kind of biofeedback used in the therapy and is a source of information about the most effective therapies [8,9]. SEMG enables to define correlation between muscles activity and clinical conditions, what let the therapist to optimize the rehabilitation goals. SEMG is sensitive and reliable tool to test muscles activity in gait analysis [6,10,11]. For patients with lower motor neuron disorders, where sEMG is wildly used, the most important data are percent of Maximal Voluntary Contraction (MVC), analysis of affected muscles fatigue, onset and offset time [6,11–14]. SEMG facilitates therapy as evaluation tool in endurance and strength training (evaluation of ability to neuromuscular recovery of tested muscles and with use of fatigue parameters [12]. It has been proven that the combination of exercises based on sEMG analysis of locomotor pattern gives a better therapeutic effect for VMO muscle recovery after injuries and surgeries [15–19]. In Charcot–Marie–Tooth disease cause of VMO muscle atrophy is different, that why it would be interesting the effectiveness of sEMG in this case.

Our test concerns a patient with high level of the disability and muscles atrophy above knees. Unfortunately research about the patient with this advanced Charcot–Marie–Tooth syndrome are very limited and there are a lot of difficulties to compare the data to the others which involve a younger patients suffering from Charcot–Marie–Tooth syndrome but with moderate level of the disability and muscle dysfunction [1,7,8]. Most of the participants in the studies with participants suffering from Charcot–Marie–Tooth disease are patients with active range of motion in the ankle. This is important information, because they are able to achieve a lot of different testing strategies: gait up and down, heel and toes walking and during the test they do not need orthopedic supplies [6]. Muscles used to be tested are VMO, VLO, RF, BF [12–15] and gluteals and calf muscles [6]. Charcot–Marie–Tooth disease concerns both legs symmetric, but our test showed high asymmetry between muscles activity in right and left leg. It can be the result of unequal loading of both lower limbs because the patient uses walking stick which is hold always in the right arm and has been essential for 13 years to keep the balance. In the left lower extremity a lot of compensatory mechanisms to make the knees extension easier, have been observed. One of them is rise of the hamstrings activity during knees extension in sitting position, despite of trunk and pelvis stabilization, and quicker activity of the biceps femoris muscle to better stabilize the knee during the movement. Hamstrings muscles are the knee extension antagonists and in the
situation of the knee dysfunction and quadriceps femoris weakness or atrophy, these muscles activation increases to compensate deficiency of the quadriceps femoris strength [9]. The next compensation mechanism is a very high activity of the gluteus medius muscle. High activity and long activation time of this muscle is a characteristic component of steppe gait, represented by Charcot–Marie–Tooth patients [6]. Low activity of the vastus medialis obliquus, vastus lateralis obliquus and rectus femoris muscles and lack of joint stability connected with quadriceps femoris atrophy lead to often occurring left knee inflammation of the soft tissues. Research with use of EMG consist of an important information for the doctors and therapists to optimize rehabilitation program for patients with heredity motor and sensory neuropathy. Proper rehabilitation is essential part of the treatment program to limit disability and disease progression. According to our research strengthening of the lower limbs muscles in sitting position cannot be effective because of the high level of synergists and antagonists activity during the movement. It would be reasonable to repeat this test to determine efficiency of the rehabilitation process. The question is, are we able to activate in the greater degree the muscles which are so atrophic, or maybe muscles degeneration and demielisation make it impossible to increase the work by quadriceps femoris muscle. Strengthening of the quadriceps femoris muscle is important to unload the muscles which compensatively work harder and can also degenerate because of their hypertrophy (e.g. gluteus medius muscle). Maybe the most important goal of the rehabilitation is to use the supplies to balance the use of the muscles, improve cardiopulmonary efficiency and the gait (to eliminate knees hyperextension, pain and inflammation) by use of crutches and knees unloading. Important factor which determines muscles weakness in patients suffering from Charcot–Marie–Tooth disease is change of the I and II muscles fibers (I fibers predominance). Sometimes after the training, level of the biceps femoris cocontraction can decrease, what decreases the need of neuronal inhibition (whose main goal is to secure the joint when the agonist muscle start to work) [3,7]. Charcot–Marie–Tooth disease is a common heterogenetic neuropathy. Multiplicity of this disease produces a lot of questions and every new research can lead to controversies and make it impossible to assess the activities undertaken to minimalize Charcot–Marie–Tooth disease effects. Patients suffering from this disease require a lot of attention and flexible management with account of their psychosomatic conditions.

In conclusion: muscles weakness in both legs is asymmetric. Left lower extremity is much weaker and underwent higher level of muscles atrophy than the right one. In both legs rectus femoris muscle works the longest time, but the activity level is low. Standing with knees extended enable to generate higher electrical activity of vastus medialis, vastus lateralis and rectus femoris musces, than standing with knees hyperextended and eliminates compensatory mechanisms which facilitate knee extension.

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**Ethics**

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

**References**


[15] Ng GY, Zhang AQ, Li CK. Biofeedback exercise improved the EMG activity ratio of the medial and lateral vasti muscles in

**Conflict of interest**

We wish to confirm that there are no known conflicts of interest associated with this study.


