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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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ABSTRACT

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, gluteus 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. Left leg is much weaker and engages antagonists and synergists muscles to compensate weaker rectus femoris, vastus medialis obliquus and vastus lateralis obliquus.

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

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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 feet 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.

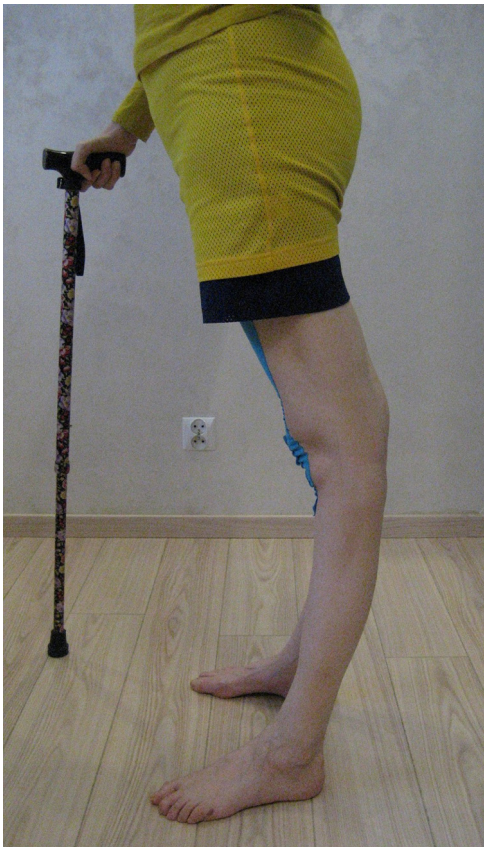

Fig. 1 – Charcot–Marie–Tooth syndrome. Knees hyperextended in standing position.

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/m^2), 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