NEUROLOGIA I NEUROCHIRURGIA POLSKA 50 (2016) 195–199

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.elsevier.com/locate/pjnns

Case report

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

Natalia Maria Kuciel [a,*], Grzegorz Krzysztof Konieczny [b], Łukasz Oleksy [c], Zdzisława Wrzosek [b]

[a] Department and Division of Medical Rehabilitation, Wroclaw Medical University, Wroclaw, Poland
[b] Faculty of Health Sciences and Physical Education, Witelon State University of Applied Sciences in Legnica, Poland
[c] Department of Clinical Rehabilitation, University School of Physical Education in Krakow, Poland

ARTICLE INFO

Article history:
Received 30 June 2015
Accepted 18 January 2016
Available online 29 January 2016

Keywords:
Electromyography
Polineuropathy
Charcot–Marie–Tooth

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

* Corresponding author at: Department and Division of Medical Rehabilitation, Wroclaw Medical University, ul Borowska 213, 50-556 Wroclaw, Poland. Tel.: +48 660897427.
E-mail address: nataliakuciel@interia.pl (N.M. Kuciel).

http://dx.doi.org/10.1016/j.pjnns.2016.01.011
0028-3843/

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 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 obliquus and vastus medialis obliquus 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 obliquus and vastus medialis obliquus 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 obliquus and vastus medialis obliquus 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 obliquus and vastus medialis obliquus 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 obliquus muscle and gluteus maximus muscles, however very high levels of the left biceps femoris activity and left gluteus 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 steppage 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 demielinisation 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.

Conflict of interest

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

Acknowledgement and financial support

We wish to confirm that there has been no financial support for this work.

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

[1] Van Dijk JP, Verhamme C, van Schaik IN, Schelhaas HJ, Mans E, Bouv LJ, et al. Age-related changes in motor unit number estimates in adult patients with Charcot–Marie–Tooth type 1A. Eur J Neurol 2010;17:1098–104.

[2] Pareyson D, Marchesi C. Diagnosis, natural history and management of Charcot–Marie–Tooth disease. Lancet Neurol 2009;8:654–67.

[3] D'Ydewalle C, Benoy V, van den Bosch L. Charcot–Marie–Tooth disease: emerging mechanism and therapies. Int J Biochem Cell Biol 2012;44:1299–304.

[4] Newman CJ, Walsh M, O'Sullivan R, Jenkinson A, Bennett D, Lynch B, et al. The characteristics of gait in Charcot–Marie–Tooth disease types I and II. Gait Posture 2007;26:120–7.

[5] Shy ME. Therapeutic strategies for the inherited neuropathies. Neuromol Med 2006;8:255–78.

[6] Don R, Serrao M, Vinci P, Rabavolo A, Cacchio A, Ioppolo F, et al. Foot drop and plantar flexion failure determine different gait strategies in Charcot–Marie–Tooth patients. Clin Biomech 2007;22:905–16.

[7] El Mhandi L, Millet GY, Calmels P, Richard A, Oullion R, Gautheron V, et al. Benefits of interval-training on fatigue and functional capacities in Charcot–Marie–Tooth disease. Muscle Nerve 2008;37:601–10.

[8] Giggins OM, Persson UM, Brian Caulfield B. Biofeedback in rehabilitation. J NeuroEng Rehabil 2013;10:60.

[9] Pullman SL, Goodin DS, Morquinez AI, Tabbal S, Rubin M. Clinical utility of surface EMG. Neurology 2000;55(2):171–7.

[10] Jette AM. Physical disablement concepts for physical therapy research and practice. Phys Ther 1994;74:380–6.

[11] Ferrarin M, Bovi G, Rabuffetti M, Mazzoleni P, Montesano A, Moroni I, et al. Reliability of instrumented movement analysis as outcome measure in Charcot–Marie–Tooth disease: results from a multitask locomotor protocol. Gait Posture 2011;34(1):36–43.

[12] Lindeman E, Spaans F, Reulen PH, Leffers P, Drukker J. Surface EMG of proximal leg muscles in neuromuscular patients and in healthy controls. Relations to force and fatigue. J Electromyogr Kinesiol 1999;9(5):299–307.

[13] Menotti F, Bazzucchi I, Felici F, Damiani A, Gori MC, Macaluso A. Neuromuscular function after muscle fatique in Charcot–Marie–Tooth type 1a patients. Muscle Nerve 2012;46(September (3)):434–9.

[14] Place N, Matkowski B, Martin A, Lepers R. Synergists activation pattern of the quadriceps muscle differs when performing sustained isometric contractions with different EMG biofeedback. Exp Brain Res 2006;174(4):595–603.

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

subjects with patellofemoral pain syndrome. J Electromyogr Kinesiol 2008;18(1):128–33.
[16] Kirnap M, Calis M, Turgut AO, Halici M, Tuncel M. The efficacy of EMG-biofeedback training on quadriceps muscle strength in patients after arthroscopic meniscectomy. J N Z Med Assoc 2005;118(1224):U1704.
[17] Wise HH, Fiebert I, Kates JL. EMG: biofeedback as treatment for patellofemoral pain syndrome. J Orthop Sports Phys Ther 1984;6(2):95–103.
[18] Matjacić Z, Zupan A. Effects of dynamic balance training during standing and stepping in patients with hereditary sensory motor neuropathy. Disabil Rehabil 2006;28 (23):1455–9.
[19] Alkjaer T, Simonsen EB, Magnusson SP, Dyhre-Poulsen P, Aagaard P. Antagonist muscle moment is increased in ACL deficient subjects during maximal dynamic knee extension. Knee 2012;19(5):633–9.