Journal of Hand Therapy 29 (2016) 66–72

Contents lists available at ScienceDirect

Journal of Hand Therapy

journal homepage: www.jhandtherapy.org

JHT Read for Credit Article #406.
Scientific/Clinical Article

Evaluation of muscle strength and manual dexterity in patients with Charcot-Marie-Tooth disease

Alena Kobesova MD, PhD [a,*], Marcela Nyvltova MPT [a], Josef Kraus MD, PhD [b], Pavel Kolar PaedDr, PhD, Prof [a], Angela Sardina MS [c], Radim Mazanec MD, PhD [d], Ross Andel PhD [c]

[a] Department of Rehabilitation and Sports Medicine, 2nd Faculty of Medicine, Charles University, University Hospital Motol, Prague, Czech Republic
[b] Department of Child Neurology, 2nd Faculty of Medicine, Charles University, University Hospital Motol, Prague, Czech Republic
[c] School of Aging Studies, University of South Florida, Tampa, FL, USA
[d] Department of Neurology, 2nd Faculty of Medicine, Charles University, University Hospital Motol, Prague, Czech Republic

ARTICLE INFO

Article history:
Received 10 February 2015
Received in revised form
27 November 2015
Accepted 3 December 2015
Available online 13 December 2015

Keywords:
Charcot-Marie-Tooth disease
Hereditary motor and sensory neuropathy
Hand strength
Manual dexterity
Sensory function

ABSTRACT

Study design: Matched pair study.
Introduction: Differences in hand-muscle strength/dexterity between dominant (DH) and non-dominant (NDH) hand in Charcot-Marie-Tooth disease (CMT) are not wellunderstood.
Purpose of the study: To compare muscle strength/dexterity between DH and NDH and to correlate manual dexterity, strength and sensory function.
Patients and methods: Thirty CMT patients were studied using functional muscle testing (FMT) and strength (dynamometry), dexterity (the Nine Hole Peg Test [NHPT]), and Jebsen–Taylor Hand Function [JTT]), and sensory function (the Nottingham Sensory Assessment [NSA]).
Results: Scores were worse for DH than NDH on FMT ($p = 0.043$) and NHPT ($p = 0.014$) but not on JTT ($p = 0.098$), handgrip strength ($p = 0.710$) or tripod pinch ($p = 0.645$). NSA did not correlate significantly with any tests (p′s0.05).
Conclusions: In CMT disease, DH appears more impaired than NDH in terms of function and dexterity. Greater muscle weakness in DH may also emerge as CMT progresses.
Level of evidence: 3b.

Introduction

Hereditary motor and sensory neuropathies form a heterogeneous group of genetically determined, degenerative diseases affecting peripheral nerves with a prevalence rate of 1:2500.[1] The disorder is known as Charcot-Marie-Tooth (CMT) disease, after the authors who first described it.[2] More than 50 genetic causes of inherited neuropathies have so far been identified.[3] CMT disease may be autosomal dominant or recessively inherited, but an X-linked form is also known.[1,4] The myelin or the axon is affected by gene mutations encoding their proteins, and CMT is accordingly subdivided into demyelinating CMT1 and axonal CMT2 forms.[5] It is possible to distinguish between the two types using nerve-conduction studies.

Despite genetic heterogeneity, there is a common clinical phenotype.[6] CMT patients often present with bilateral distal muscle weakness and wasting of the lower extremities, pes cavus deformity, and distal sensory loss, with abnormal stepping gait as a result. Spinal deformities are more common in CMT patients than in the general population.[6,7] CMT symptoms usually develop in first two decades of life with the disease subsequently progressing over life.[6] The upper extremities usually become involved later, however, reduced hand function limiting prehension may occur early in the progression of the disease, resulting in reliance on compensatory grasp patterns.[8] A distal to proximal progression of muscle weakness, wasting, and sensory loss occurs, ultimately resulting in a "claw hand" with disturbed dexterity. Albeit CMT patients rarely experience hand pain, limitations in upper limb functioning is perceived by the majority of patients with CMT to be strongly related

Statement of financial disclosure and conflict of interest: This study was supported by the foundation Movement without Help, Prague, Czech Republic and by scientific program PRVOUK 38. There are no conflicts of interest for any of the authors.
Statement of Institutional Review Board approval of the study protocol: This study was approved by the Second Medical Faculty, Charles University and University Hospital Motol, Prague, Czech Republic, ethical committee.

* Corresponding author. Department of Rehabilitation and Sports Medicine, Second Medical Faculty, Charles University and University Hospital Motol, V Uvalu 84, Prague 5, 159 00, Czech Republic. Tel.: +420 224 43 9264; fax: +420 224 439 220.
E-mail address: alenamudr@me.com (A. Kobesova).

0894-1130/$ – see front matter © 2016 Hanley & Belfus, an imprint of Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.jht.2015.12.002

to restricted participation at work, family role or leisure activities.[9] Greater involvement of the dominant hand as a result of overwork has been reported by some authors,[10,11] while others question its existence.[12–14] Understanding the progression of CMT with respect to the strength, dexterity and function of the DH vs. NDH has important clinical implications. If enough evidence exists that the NDH becomes superior to the DH with respect to strength, dexterity, or function (or all) as the disease progresses, clinical approach may shift to encourage training and use of the NDH over the DH.

Few studies address the relationship between muscle strength, sensory deficit and manual dexterity,[15–18] however, no consensus exists on how to evaluate hand function in CMT patients. The aim of our study was to compare muscle strength and dexterity in all CMT patient volunteers using a matched pair design, where the matched pair was represented by the DH and NDH of the same patient. Previously, Vinci[19] and Videler[13] suggested that overwork weakness appears in more severely affected CMT patients. To test this notion, we also classified patients into three categories of muscle weakness according to the published CMTNS criteria[20] and used our measure of handgrip strength to explore DH vs. NDH differences in handgrip strength based on severity. In addition, we present correlations of hand muscle strength and sensory function with manual dexterity.

Methods

Participants

Thirty patients, of whom 13 were male, aged 21–68 (mean age 40.2 ± 10.29) were recruited from the local Clinic of Rehabilitation and Sports Medicine. Exclusion criteria was any disease other than CMT that may cause hand weakness and impaired motor dexterity and sensitivity. Written informed consent was obtained from all participants. CMT diagnosis was confirmed by electromyography (EMG) and DNA analysis, or by EMG alone (in absence of confirmed genotype markers). Twenty-five patients were classified as CMT1 (demyelinating type) and 5 patients as CMT2 (axonal type); 29 patients declared right-hand dominance, 1 was left-hand dominant. According to Charcot-Marie-Tooth Neuropathy Score (CMTNS),[20] 6 patients fell into Group I, 14 into group II, and 10 into Group III.

Assessments

All measurements were performed by the same researcher in the following sequence.

Muscle strength evaluated by functional muscle testing (FMT)

Janda's *functional muscle testing*[21] was utilized to assess hand and forearm muscle strength. The test includes 26 measurements for each hand, as shown in Fig. 1. The Medical Research Council (MRC) Scale was used for muscle grading, each ranging from 0 to 5. We averaged scores across these 26 measurements and measured FMT with this continuous variable.

Muscle strength evaluated by dynamometry

Handgrip strength was assessed using hand-held dynamometry (Citec, C.I.T. Technics, Haren, The Netherlands) following standardized testing procedure.[22] *Tripod pinch strength* was assessed as shown in Fig. 2. Three attempts were recorded (in Newtons) for each the handgrip strength and the tripod pinch strength tests. As done previously,[11] the three trials were averaged to create scores for analysis.

Dexterity

The *Nine Hole Peg Test (NHPT)*, which involves taking pegs one-by-one from a container, placing them into holes on a board, and then replacing them back into the container. As recommended by standardized testing procedures,[23] the test was performed by the DH followed by NDH and normalized based on sex and age. In addition, *the Jebsen–Taylor Test (JTT)*[24] was performed by NDH followed by DH and normalized based on sex and age. JTT is a well-established, widely used test of hand function and dexterity consisting of seven tasks which are representative of hand activities performed during activities of daily living.

Sensory function

Nottingham Sensory Assessment (NSA)[25] was used. The NSA is used mainly to identify sensory deficits, i.e. proprioceptive, stereognostic and epicritic sensory modalities in hemiparetic patients, showing good psychometric properties.[25] For this study, only hand, wrist and elbow segments were tested. British coins suggested for stereognostic testing by NSA authors were replaced by local currency coins of similar size and shape. For further analysis, all points scored within this test were added up for DH and for NDH. The maximum score was 97 points, implying normal stereognostic function at the area of hand, wrist and elbow.

To reduce any influence of fatigue, 5-min breaks were given between tests.

Statistical analysis

Normality of score distribution was assessed using the Kolgomorov–Smirnov test. The test revealed that scores for the tripod pinch test, and JTT test were not normally distributed. Therefore, the differences between the DH and NDH were analyzed using the non-parametric Wilcoxon signed-rank test for paired samples. Student's paired-sample *t*-test was used otherwise.

Based on Vinci[19] and Videler,[13] who posit that overwork weakness appears in more severely affected CMT patients, we used CMTNS criteria[20] to classify patients based on the extent of impairment into stage I ($n = 6$), stage II ($n = 14$), and stage III ($n = 10$), and applied this classification to handgrip dynamometry. Given the small sample size, we consider these results only exploratory.

In addition, we examined correlations between the administered tests of strength, dexterity, and sensory function. Spearman rank correlation coefficients, which are less susceptible to bias due to outliers than Pearson correlation, were used with non-normally distributed variables. A p-value <0.05 determined significance. Following Svensson at al[18] we interpreted the strength of the correlation according to Munro[26]: <0.25 little if any correlation, 0.26–0.49 low correlation, 0.50–0.69 moderate correlation, 0.70–0.89 high correlation, >0.90 very high correlation. All analyses were performed using the SAS software version 9 (The SAS Institute, Cary, NC). Significance was set at a two-tailed 0.05 level.

Results

Comparison of muscle strength and function between DH and NDH (see Table 1)

With respect to muscle strength, the NDH was significantly stronger than the DH in FMT. Handgrip dynamometry and the tripod pinch test yielded no differences in strength between the NDH and the DH.

We subcategorized patients into groups according to CMTNS stages I–III (not in Table 2). Handgrip strength was greater for NDH than DH in Group II ($p = 0.002$) and Group III ($p = 0.037$), but not in Group I ($p = 0.249$).

With respect to motor dexterity, the scores were significantly better for the NDH than the DH on NHPT, and the difference in JTT scores approached significance.