

Testing overwork weakness in Charcot-Marie-Tooth (CMT) disease: is it true or false?

<u>Prada V</u>, Mori L, Francini L, Accogli S, Ursino G, Gemelli C, Schizzi S, Grandis M, Bellone E, Mandich P, Schenone A.

Department of Neuroscience, Ophtalmology, Rehabilitation, Genetics and Child/Maternal Sciences, DINOGMI, University of

Genova, Italy



INTRODUCTION

The overwork weakness (OW) problem in CMT disease has been debated for long time. It has been reported that the non-dominant hand (NDH) of patients with CMT disease is stronger than the dominant hand (DH) as a result of OW and some authors verified this hypothesis using MRC on different muscles (Van Pomeren, 2009). More recently, Piscosquito et al. (2014) found that the OW phenomenon does not exist using a dynamometer and the 9 hole peg test, a dexterity test. We propose our evaluation with Thumb Opposition Test, Strenght Test with a dynamometer and a Sensor Engineered Glove Test (SEGT).

MATERIALS & METHODS

Patients enrolled:

- n: 56 CMT patients (28 M; 28 F)
- N: 24 healthy controls (9 M; 13 F)
- Age range: 22 79

<u>Tests Performed</u> and compared



Tripod pinch & Hand grip strength

In the Healthy Subjects, DH is significantly stronger than NDH, on the contrary CMT patients strength is similar in both hands (and on average lower than Healthy controls).





- Thumb Opposition Test (TOT)
- Dynamometer (tripod pinch & hand grip) • SEGT (FT & IMRL @ MV)



SEGT is an engineered glove which measures severity of hands dysfunction in CMT patients (Alberti et al., 2015)



SEGT protocol:

- Patient:
- Eyes closed
- Relaxed
- Alternating Hands
- Duration: 30 sec
- Protocols:
- FT at max velocity (MV)
- Sequence (IMRL) @ max velocity (MV)

Parameters evaluated:

- Touch Duration (TD) in ms
- Inter-tapping Interval (ITI) in ms
- Movement Rate (MR, 1/TD+ITI) in Hz

Hands strength is fundamental for ADL. The 10% rule states that in normal subjects strength must be higher in the DH.

50·

SEGT

In the two task proposed (FT & IMRL sequence), Healthy Subjects have a DH performance better than the NDH performance. Instead, CMT patients performed the tasks similarly with the both hands.

Healthy Controls

CMT patients

Thumb opposition test

In the Healthy Subjects there is no significant difference between DH and NDH, on the contrary in CMT patients TOT is significantly higher in the NDH.





In conclusion, this is the first study which supports the existence of the overwork weakness in CMT that matches different kind of measures. Overwork weakness is evident in the TOT. Furthermore, the strength of the hand is compromised too. In fact, according to the 10% rule (Noguchi & Demura, 2009) we should expect a higher strength in the DH. Interestingly, even the measures of hand dysfunction, as is the SEGT, support this hypothesis. We speculate that compensating movements in the weaker hand of CMT patients impairs the dexterity on both hands. Finally, our results support the



SEGT measures the dexterity of the hands. In normal subjects, even the dexterity is better in the DH. Instead, in CMT patients the dexterity of the DH decreases and is similar to the dexterity of the NDH.

