



Six months of strength training reduces progression of dorsiflexor muscle weakness in children with Charcot-Marie-Tooth disease [commentary]

Commentary

Charcot-Marie-Tooth is the most common hereditary neuromuscular disease, and is sporadically encountered by physiotherapists in neurology and orthopaedic clinics. It is a slowly progressing condition with changes experienced over years rather than months. This presents challenges from a research perspective, as tools to measure changes in the natural history need to be very sensitive. Quantitative strength testing using dynamometry and magnetic resonance imaging have successfully shown deterioration in a 1- to 2-year period.^{1,2}

The rationale, to date, for strength training in this progressive neuropathy has been to reverse disuse atrophy in the less affected proximal muscles. The unanswered question has always been: can the more affected distal muscles be strengthened? This is the first randomised, controlled trial to specifically explore this question. By the time people with Charcot-Marie-Tooth reach adulthood, they can have much reduced strength in their distal muscles. Adults with the most common type, CMT1A, have 30% of the strength of matched controls in their dorsiflexors,¹ supporting the rationale of this trial to strength train children and teenagers with Charcot-Marie-Tooth disease.

This was a rigorously conducted, well powered trial and I applaud the team for the participant blinding with a sham intervention, as a believable sham can be challenging in exercise trials. The striking

finding is the difference in unadjusted z-scores for dorsiflexion strength between the training and sham group, particularly at 12 and 24 months. The difference appears to be due to persistence of an initial training effect in the training group, and deterioration according to natural history in the sham group. The lack of change in muscle volumes points to the possibility that changes were neural rather than due to muscle hypertrophy. Central activation failure has been observed in people with Charcot-Marie-Tooth disease,³ and although the authors hypothesise peripheral neural changes as a mechanism to explain their results, central changes cannot be ruled out and are worthy of further investigation.

Provenance: Invited. Not peer reviewed.

Gita Ramdharry

School of Allied Health, Midwifery & Social Work, Kingston University and St George's University of London, United Kingdom

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