



Does baseline BMI affect progression of disability in children with Charcot-Marie-Tooth disease?

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Introduction

- Body Mass Index (BMI) has been shown to influence function in healthy children as well as those with neuromuscular disorders.
- Both high and low BMI is thought to be related to disease severity in children with CMT, however there is an absence of empirical data.
- We have previously reported the association between BMI and disability in children and adolescents with CMT.¹ Children who are severely underweight, or obese have worse baseline disability scores on the Charcot-Marie-Tooth Paediatric Scale (CMTPedS).
- The influence of BMI on progression of disability in CMT is not known.

Aims

To (1) evaluate the impact of baseline BMI on disability after 2-years in children with CMT; and (2) evaluate the impact of a change BMI on disease progression over 2 years in children with CMT.

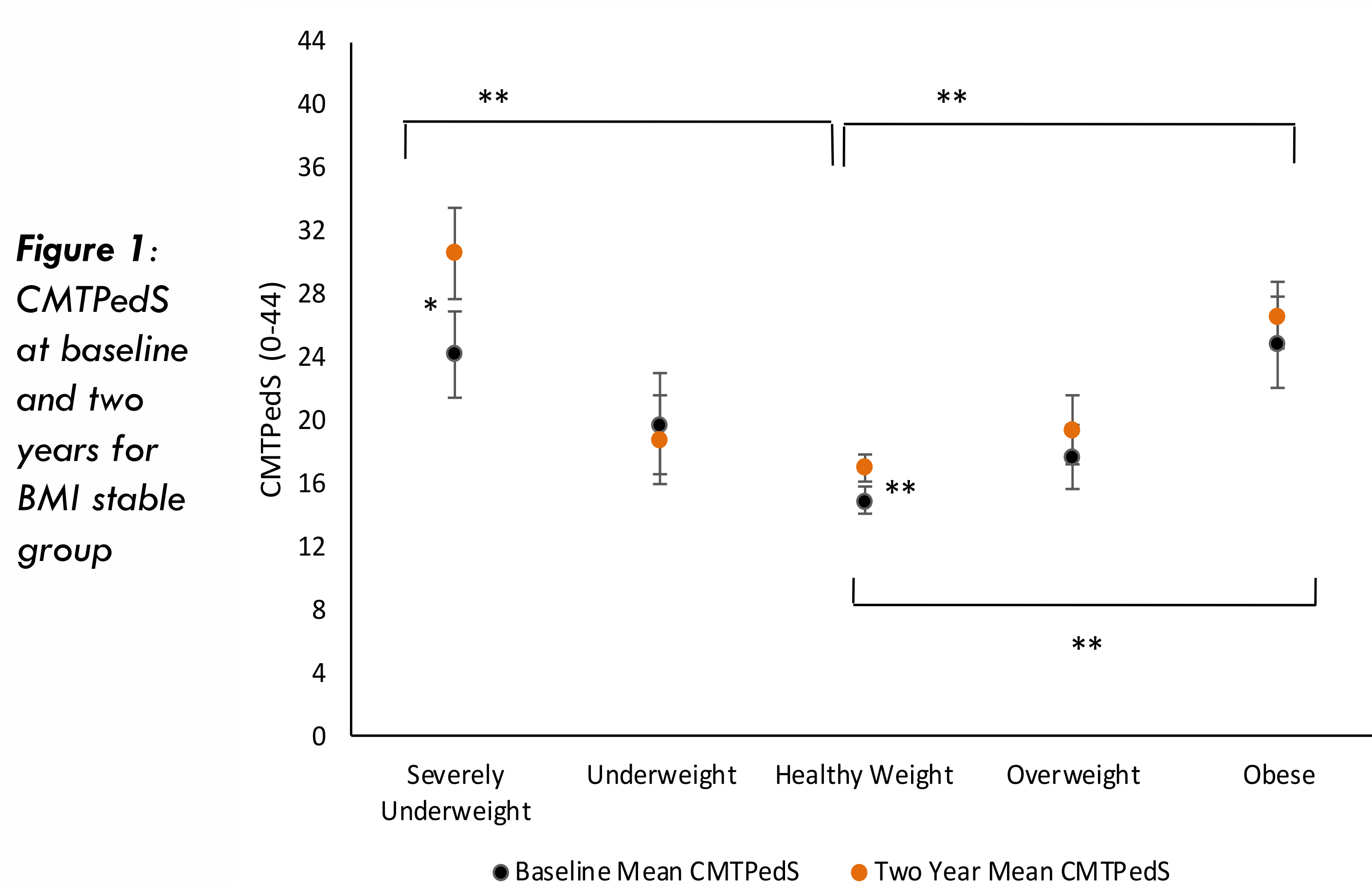
Methods

- 242 participants aged 3-20 years enrolled in the Inherited Neuropathy Consortium (INC) were assessed at baseline and after 2-years with the 0-44 point (CMTPedS).²
- BMI was classified using the International Obesity Task Force (IOTF) criteria (equivalent to an adult BMI) : severely underweight, underweight healthy weight, overweight, and obese.
- Participants were stratified according to IOTF BMI category across the two-year period as either 'BMI Stable' or 'BMI Change' groups.

Results

BMI Stable Group

- 69% of children did not change BMI category between baseline and two-years.
- The distribution across weight categories was severely underweight (2.9%), underweight (5.4%), and overweight (13.1%), and obese (5.4%) participants..
- Both baseline and two-years mean CMTPedS scores for disability assessment across the five weight categories demonstrated a u-shaped association.



- **At baseline children who were obese were significantly more disabled than those of healthy weight (p=0.023), whilst at two-years those who were severely underweight (p=0.014) or those who were obese were significantly more disabled than those of a healthy weight (p=0.015 and p=0.027).**

- **Those who were categorised as severely underweight at baseline , had a mean change in CMTPedS scores of 6.4 (3.2) over two-years, reflecting a 26.4% change from baseline (p=0.011).**
- **Within this cohort therefore, being severely underweight at baseline appears to independently influence CMT progression according the CMTPedS assessment as a marker of disability, in those whose BMI remained stable over time.**

BMI change Group

- 30.6% of participants BMI category changed between baseline and two-years.

- BMI Groups were classified according to BMI direction of change.

- Those who moved from being underweight towards a healthy weight had the highest mean CMTPedS scores at baseline.

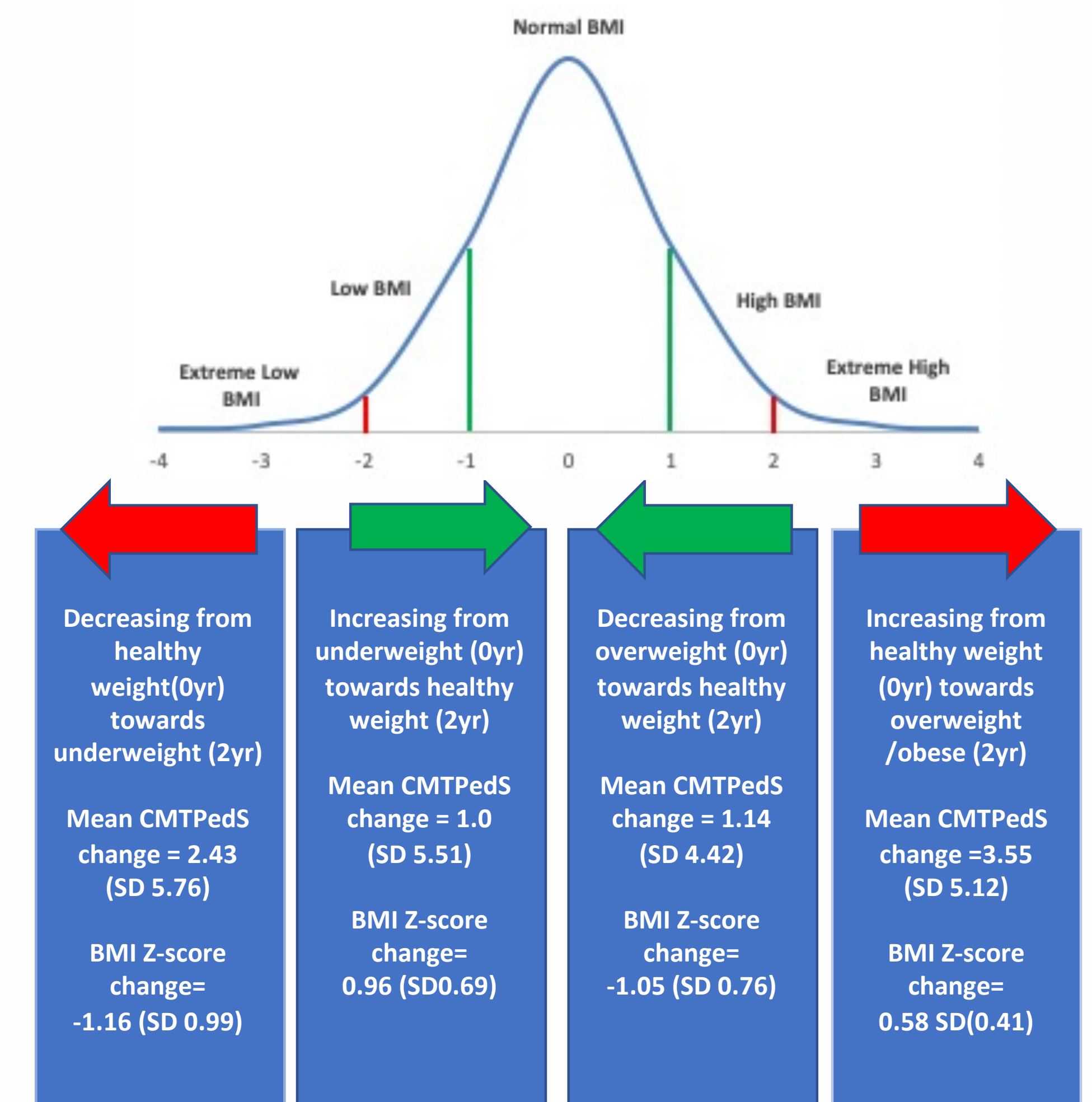


Figure 2: Changing BMI impacts disability

- **Those who moved closer towards a healthy weight experienced a lower change in CMTPedS scores (3-7%), compared to those who moved further away from a healthy weight range who experienced a higher change in CMTPedS scores (13-22%).**

Significance

- In a multicentre longitudinal analysis of 242 children and adolescents with CMT, progression of disability was dependant on BMI. Disability of severely underweight children with CMT progressed faster over two-years, than healthy, overweight and obese children.
- BMI appears to be an important factor in understanding natural history studies and the design of clinical trials.
- Clinicians aim for a BMI that targets towards the healthy weight range, as opposed to being on the extreme ends of weight for age where disease severity is increased.
- Maintaining a healthy BMI may prevent disability progression and good nutrition should be targeted to improve outcomes for children and adolescents with CMT.

References

1. Donlevy G, Garnett S, Cornett K, McKay MJ, Baldwin JN, Burns J*, (PNS 2019) Menezes MP* on behalf of the Inherited Neuropathies Consortium, Influence of Body Mass Index on disability in children with CMT
2. Burns et al. Validation of the Charcot-Marie-Tooth disease pediatric scale as an outcome measure of disability. *Ann Neurol.* 2012;71(5):642-652.

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