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RESEARCH PAPER


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ABSTRACT

Aim: To investigate whether the amount and distribution of lean body mass and fat mass is associated with disease severity in adults with Charcot-Marie Tooth.

Methods: Ten participants (age 46 ± 13 y, height 1.7 ± 0.1 m, and body mass 77 ± 17 kg) with Charcot-Marie Tooth disease were involved in this study. Participants were evaluated for quality of life, falls efficacy, balance, mobility, muscle strength, and power. Body composition was measured using dual energy x-ray absorptiometry. Statistical analyses were conducted on subsets of all participants.

Results: Better static balance was associated with higher lean body mass of the lower leg (r = 0.73, p = 0.03), while superior leg press strength and power was associated with greater lean body mass of the leg and lower leg (r ≥ 0.80, p ≤ 0.01). Faster habitual walking speed and enhanced quality of life was associated with lower fat mass of several regions.

Conclusion: Our study seems to suggest that assessing of body composition could assist with monitoring of disease progression in people with Charcot-Marie Tooth; however these findings need to be substantiated in a larger cohort.

IMPLICATIONS FOR REHABILITATION

- Higher lean body mass and lower fat mass of the legs is associated with better physical performances in people with Charcot-Marie-Tooth disease.
- Lower fat mass is related to greater quality of life and reduced clinical symptoms in people with Charcot-Marie-Tooth disease.
- Optimising favorable body composition profiles (higher lean body mass and lower fat mass) in people with Charcot-Marie-Tooth disease may be highly clinically relevant.

Introduction

Charcot-Marie Tooth (CMT) disease is a slowly progressive neuroopathy that leads to both distal and proximal muscle weakness with a distal-to-proximal progression [1,2]. The resulting muscle weakness gradually affects physical function and may lead to disability [3,4]. As a result of the functional limitations and loss of independence experienced by people with CMT, quality of life is reduced compared to the general population [5]. Furthermore, the increased effort required to walk as well as frequent falling and tripping caused by drop foot may lead to low levels of physical activity [6,7]. It is well established that low levels of physical activity are associated with profound changes in body composition, including loss of skeletal muscle and gain in fat mass [8]. For people with CMT, these unfavorable changes in body composition may lead to further deterioration of physical function and increase the risk of developing co-morbidities (e.g., type 2 diabetes and cardiovascular disease) [9,10].

A common manifestation of aging is a loss of skeletal muscle function and mass, known as sarcopenia [11], along with increases in whole-body and regional fat deposits [12]. Recent trends demonstrate that body adiposity compared to reductions in muscle mass play a more influential role in physical function decline in older adults [13]. Impairment in the force generating capacities of a muscle may occur with the presence of fat within (intramuscular) and between (intermuscular) muscle fibers due to interference with muscle activation [14] and/or disruption of force transmission [15]. Studies that have examined the body composition of people with CMT1A (most common type) have concentrated on the lower limb due to this region of the body being most affected [16–18]. Whilst the loss of the distal muscle strength is related mainly to the nerve degeneration [19], as the disease progresses predominantly proximal and distal loss of muscle mass and subtle fatty infiltration of calf muscle compartments occurs [20,21]. However, to date no study has examined the relationship between body composition, physical performance, and quality of life in people with CMT. Such research would be advantageous for providing a theoretical foundation for controlled trials targeting improved function and quality of life and thus expanding current treatment options.
Recent work from Roberts-Clarke et al. [22] explored the relationship between physical performance variables and quality of life in a cohort of ten participants with CMT. The main findings of this study were that leg press power was positively associated with quality of life, while seated row and hip abductor strength were related to better walking speed. The aim of the present study was to describe the body composition of the same ten participants with CMT and examine the association with physical performance variables, quality of life, and clinical indicators. It was hypothesised that better physical performances would be associated with higher lean body mass specific to the physical task (e.g., greater lean body mass of lower leg and better static balance) as well as better quality of life. Also, it was expected that higher fat mass would be associated with worse functional movement performances (e.g., greater fat mass and slower walking speed) and poorer quality of life measures.

**Materials and methods**

This is a secondary analysis of data from Roberts-Clarke et al. [22] and included a convenience sample of ten participants (five male and female, respectively, age 46 ± 13 y, height 1.7 ± 0.1 m, and body mass 77 ± 17 kg) with CMT (CMT1A n = 5; CMT-X n = 2; unknown genetic origin n = 3) (Figure 1). To be eligible for study inclusion participants needed to be 18–60 y and clinically diagnosed with CMT based on genetic and electrophysiological data. The criteria for study exclusion were being insufficiently ambulatory (defined as the inability to walk 100 m in under 4 min) [23] or having preexisting conditions that could be worsened by exercise. A convenience sample of healthy sex and age-matched adults (age 46 ± 12 y, height 1.7 ± 0.1 m, and body mass 76 ± 15 kg) were obtained from study databases within the Discipline of Exercise and Sport Science, The University of Sydney, Australia. Characteristics and body composition data from the healthy sex and age-matched adults (normative data) was used to compare to the participants with CMT. This study was registered with the Australian New Zealand Clinical Trials Registry (ACTRN12614000173695). The University of Sydney Human Research Ethics Committee granted approval for this study (Project No: 2013/BS1) and all participants provided written informed consent.

**Recruitment**

The sample of community dwelling adults with CMT was recruited from Sydney, Australia between January and September 2014. Recruitment involved a variety of methods and in total, 297 letters were sent to members of Charcot-Marie-Tooth Association Australia, patients from a local neuropathy clinic, previous study participants and those who had expressed interest via word of mouth. There were 29 people that responded to the letters of which, 19 were eligible to participate in the study (detailed in Figure 1). Following a telephone screening, eight of those eligible could not commit to the study requirements (i.e., three visits to the laboratory for testing) and one was unable to commence due to medical reasons (i.e., hospitalised).

**Body composition**

A whole-body dual energy x-ray absorptiometry scanner, (Lunar Prodigy, GE Medical Systems, Madison, WI) was used to measure body composition. Scans were performed under standardised conditions (early morning, overnight fasted, and standardised body positioning on the scanning bed), by two licensed co-investigators (GW, YM). Following the scan, in-built analysis software (version 13.60.033; enCORE 2011, GE Healthcare, Madison, WI) allowed the calculation of total and regional (predefined by the software) lean body mass, excluding bone mineral content, and fat mass. A customised region of interest was created for the lower legs which included the region below the axis of knee [24], including the foot. Height-adjusted indices for body composition were calculated by dividing each participant’s lean body mass and fat mass measures in kg by the participant’s height squared in m^2 [25,26].

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**Figure 1.** Flow chart of participants with Charcot-Marie-Tooth disease recruited.
This index, expressed as kg/m², was the body composition measure used for all analyses. Sarcopenia cutoffs based on lean body mass/m² were defined as <15.5 for males and <12.6 kg/m² for females [27], respectively, which was previously established in older adults [28].

**Physical performance tasks**

The physical performance assessment tasks performed in this study have been detailed previously [22]; however, the tasks will be briefly described. Muscle strength was assessed via the one-repetition maximum for the leg press, knee extension, knee flexion, chest press, and seated row using Keiser A420 pneumatic resistance training equipment (Keiser Sports Health Equipment, Inc., Fresno, CA). For the same exercises (except for knee flexion), muscular power was assessed with the Keiser machines recording the peak power produced. Leg power was also estimated from a stair climb power test, which was calculated from the time taken for participants to ascend a flight of nine stairs as quickly as possible [29]. Power was estimated using the equation below:

\[
\text{Power (W)} = \frac{\text{[Body mass (kg)]} \times \text{vertical height of the staircase (m)} \times 9.8 \text{ (m.s}^{-2})}{\text{time (s)}}
\]

Isometric handgrip strength of the non-dominant hand was assessed using a JAMAR handgrip dynamometer (Sammons Preston, Bolingbrook, IL). Peak isometric ankle dorsiflexion and plantarflexion strength were measured on both right and left sides, as the highest of three measures, using an isometric digital dynamometer (Chatillon Dynamometer CSD200; Ametek TCI Division, Largo, FL). Balance was assessed using static standing postures (e.g., wide stance, narrow stance, half tandem, and full tandem) via tandem walking and tandem walking with a cognitive distracter (verbal fluency: naming different animals aloud) [30].

Other physical performance assessment tasks included gait speed (habitual and maximal) over a two metre distance, distanced covered during a six minute walk and time required to perform five sit-to-stands (chair rise) as quickly as possible. Some of the physical performance tests were not completed by all participants due to a variety of circumstances. One participant was unable to complete the muscular power tests for the chest press and seated row due to concerns over a previous shoulder injury. Another participant was unable to complete muscular strength and power for chest press and seated row as well as the two tandem walks tests due to unrelated illness. Technical error also affected data collection of one participant for the tandem walk with cognitive distracter and another participant for the dorsiflexion and plantarflexion muscular strength tests. Therefore, the physical tests that were not completed by all 10 participants were the tandem walk (n = 9), tandem walk with cognitive distracter (n = 8), plantar flexion and dorsiflexion (n = 9), chest press power (n = 8), seated row power (n = 8), chest press strength (n = 9), and seated row strength (n = 8).

**Indicators of quality of life**

Scores from the Short-Form 36 Health Status Survey (SF-36; version 2) were summarised into two components: the physical component score and mental component score to assess quality of life [31]. A modified version of the Tinetti falls efficacy scale was used to assess fear of falling [32,33]. The modification involved the inclusion of four additional items to the first 10 items described by Tinetti et al. [33]. Administration, scoring and normative values for these assessment tools have been previously described [22]. Briefly, higher physical and mental component scores reflected better quality of life, while higher falls efficacy scale scores were equivalent to lower confidence or efficacy.

**Statistical analysis**

Statistical analyses were performed using SPSS version 22.0 for Windows (IBM Corp. Armonk, NY). Data distributions were inspected visually and statistically for normality. All data except the six minute walk was normally distributed. Therefore, data for the six minute walk was logarithmically transformed (log_{10}) to obtain a close-to-normal distribution prior to further analysis. History of CMT (age of symptom onset, duration of symptoms, age of diagnosis, and CMT exam score) and quality of life indicators (falls efficacy scale scores, physical, and mental component scores) were compared between sexes using unpaired t-tests. Characteristics of participants with CMT including age, height, body mass, waist circumference, and body composition were also compared between an age-matched group of healthy adults for each sex, and respective sexes (males and females with CMT) using unpaired t-tests. Partial correlation analyses (adjusting for sex) were performed to examine relationships between height-adjusted body composition indices and both physical performance measures, quality of life, and clinical indicators. Strength of correlations were qualitatively assessed using the following criteria: trivial (r < 0.1), small (r > 0.1–0.3), moderate (r > 0.3–0.4), strong (r > 0.5–0.7), very strong (r > 0.7–0.9), nearly perfect (r > 0.9), and perfect (r = 1.0) [34]. Data are presented as mean standard deviation (±). Significance was set at p < 0.05; however, because of the relatively small study sample, trends were declared at p ≤ 0.05–0.10.

**Results**

History of CMT and quality of life indicators of participants with CMT are detailed in Table 1. Age of diagnosis was later for females compared to males (p < 0.05), while no differences between sexes were found for age of symptom onset, duration of symptoms, and CMT exam score (Table 1). Males compared to females were found to have a higher physical component score suggesting better health-related quality of life for this SF-36 component (p < 0.05), and no difference between sexes was found for the mental component score. There was also a trend for males to have lower falls efficacy scale scores compared to females with CMT suggesting greater confidence or efficacy (p = 0.06).

Characteristics and body composition (total and regional) of participants with CMT and an age-matched group of healthy adults are detailed in Table 2. Healthy males compared to males with CMT had greater total, leg, and lower leg lean body mass (p < 0.05). Greater % body fat (p < 0.05) and a trend for greater lean body mass of the arms (p < 0.06) was also found for healthy CMT.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Males (n = 5)</th>
<th>Females (n = 5)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of symptom onset (y)</td>
<td>8.3 ± 6.4</td>
<td>26.0 ± 20.0</td>
</tr>
<tr>
<td>Duration of symptoms (y)</td>
<td>38.0 ± 5.7</td>
<td>22.4 ± 20.4</td>
</tr>
<tr>
<td>Age at diagnosis (y)</td>
<td>13.8 ± 5.9</td>
<td>39.6 ± 14.8*</td>
</tr>
<tr>
<td>CMT exam score</td>
<td>12.7 ± 4.0</td>
<td>10.8 ± 1.3</td>
</tr>
<tr>
<td>Falls efficacy scale score</td>
<td>18.8 ± 4.9</td>
<td>26.8 ± 6.6</td>
</tr>
<tr>
<td>Physical component score</td>
<td>51.6 ± 5.5</td>
<td>41.4 ± 6.6*</td>
</tr>
<tr>
<td>Mental component score</td>
<td>54.6 ± 7.7</td>
<td>49.8 ± 6.4</td>
</tr>
</tbody>
</table>

CMT: Charcot-Marie-Tooth.

*Significant difference between males and females at p < 0.05.
males compared to males with CMT. There were no significant differences (or trends) between healthy males and males with CMT for age, height, body mass, waist circumference, trunk lean body mass, total fat mass, and regional fat mass.

Healthy females compared to females with CMT had lower % body fat, and less android, legs, and lower legs fat mass (p < 0.05). There was a trend for healthy females compared to females with CMT having less total fat mass (p = 0.06). Healthy females compared to females with CMT were found to have greater lower limb lean body mass (p < 0.05). There were no significant differences (or trends) between healthy females and females with CMT for age, height, body mass, waist circumference, total lean body mass, trunk lean body mass, arms lean body mass, legs lean body mass, total fat mass, and arms fat mass.

Female compared to male participants with CMT had greater total, % body fat, and regional (arms, legs, and lower legs) fat mass (p < 0.05). Females with CMT also had greater lean body mass of the lower legs compared to males with CMT (p = 0.01), while males with CMT were taller (p ≤ 0.01) and had greater lean body mass of the arms compared to females with CMT (p = 0.02). No other differences were found between males and females with CMT for characteristics and body composition.

### Body composition, quality of life, and clinical indicators

As shown in Table 4, very strong positive correlations were found between age of symptoms onset and total, arms, legs, and lower legs fat mass (r = 0.74 to 0.87, p ≤ 0.01–0.04). There was a trend for a strong positive correlation found between age of symptoms onset and android fat mass (r = 0.67, p = 0.07). A very strong negative correlation was found between physical component score and total fat mass (r = −0.71, p = 0.03), while a strong negative correlation was found between physical component score and fat mass of the legs (r = −0.67, p = 0.04). A trend for a strong positive correlation was found between falls efficacy scale scores and android fat mass (r = 0.60, p = 0.09), as well as a trend for a strong negative correlation between mental component score and lower leg fat (r = −0.59, p = 0.09).

### Discussion

In the present study we found that body composition was associated with better physical performances, quality of life, and clinical indicators in a small sample of adults with CMT. In agreement with the original hypotheses, better static balance was strongly associated with greater lean body mass of the lower leg, while superior leg press strength and power was associated with greater lean body mass of the leg and lower leg. Also in agreement with the hypotheses, faster walking speed was associated with less fat mass of the lower leg and greater quality of life (physical component score) was associated with less total and leg fat mass. Later
Muscle strength
Muscle power

Shahrizaila et al. [37] found that greater volume and thickness of lean body mass (lower fat mass) in people with CMT may be a correlation at the

correlation is significant at p < 0.05.
correlation is significant at p < 0.01.
correlation at p < 0.05 (trend).
Tandem walk CD: tandem walk with cognitive distracter; L: left; R: right.

Table 3. Correlation matrix between body composition and physical performance in participants with Charcot-Marie-Tooth disease.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Lean body mass</th>
<th>Fat mass</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>Trunk</td>
<td>Arms</td>
</tr>
<tr>
<td>Balance</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Static</td>
<td>0.43</td>
<td>0.27</td>
</tr>
<tr>
<td>Tandem walk</td>
<td>0.08</td>
<td>0.16</td>
</tr>
<tr>
<td>Tandem walk CD</td>
<td>0.36</td>
<td>0.40</td>
</tr>
<tr>
<td>Gait speed</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Habitual</td>
<td>-0.34</td>
<td>-0.42</td>
</tr>
<tr>
<td>Maximal</td>
<td>0.14</td>
<td>0.05</td>
</tr>
<tr>
<td>6 minute walk</td>
<td>0.14</td>
<td>0.01</td>
</tr>
<tr>
<td>Stair Climb (estimated power)</td>
<td>0.34</td>
<td>0.34</td>
</tr>
<tr>
<td>Chair rise</td>
<td>0.22</td>
<td>0.23</td>
</tr>
</tbody>
</table>

Muscle strength
Leg press
Knee extension
Knee flexion
Chest press
Plantar flexion (L)
Plantar flexion (R)
Dorsiflexion (L)
Dorsiflexion (R)
Hand grip (L)
Hand grip (R)

Muscle power
Leg press
Knee extension
Chest press
Seated row

Table 4. Correlation matrix between body composition and indicators of quality of life in participants with Charcot-Marie-Tooth disease.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Lean body mass</th>
<th>Fat mass</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>Trunk</td>
<td>Arms</td>
</tr>
<tr>
<td>Age</td>
<td>0.24</td>
<td>0.32</td>
</tr>
<tr>
<td>Age at symptom onset</td>
<td>0.37</td>
<td>0.48</td>
</tr>
<tr>
<td>Duration of symptoms</td>
<td>-0.23</td>
<td>-0.24</td>
</tr>
<tr>
<td>Age at diagnosis</td>
<td>0.56</td>
<td>0.53</td>
</tr>
<tr>
<td>CMT exam score</td>
<td>0.28</td>
<td>0.38</td>
</tr>
<tr>
<td>Falls efficacy scale score</td>
<td>0.56</td>
<td>0.59</td>
</tr>
<tr>
<td>Physical component score</td>
<td>0.15</td>
<td>-0.05</td>
</tr>
<tr>
<td>Mental component score</td>
<td>0.11</td>
<td>0.20</td>
</tr>
</tbody>
</table>

It has been proposed that the magnitude of fat within muscle fibers (intramuscular adipose tissue) and between muscle fibers (intermuscular adipose tissue) may contribute to impaired muscle force production [40]. Fat infiltration into the muscle is known to positively correlate with overall body fatness [41,42]. Furthermore, increased fat infiltration of leg muscles has been shown to be an independent risk factor for incident mobility limitations and may be a crucial aspect of sarcopenia that affects functional
status in the elderly [43,44]. Numerous CMT case studies have been published that show prominent fatty infiltration of lower leg muscles [16,18,20,21]. Recently, Morrow et al. [45] showed that significant intramuscular fat accumulation in the calf can occur over a 12-month period in CMT1A patients compared to match-controls. Therefore, assessment of fat infiltration of the calf muscles in people with CMT via magnetic resonance imaging could be a helpful marker of disease severity and potentially as a clinical biomarker of disease progression. Unfortunately fat infiltration of skeletal muscle could not be determined in the present study and would have required the use of magnetic resonance imaging or computed tomography. However, Davison et al. [46] previously found subcutaneous fat in the leg and lower leg regions to be positively correlated with fat infiltration of skeletal muscle. Due to the findings of the present study of no association between fat mass and muscle strength, it appears unlikely that fat infiltration of the leg muscles plays a major role in deterioration of physical capacity in people with CMT.

The only significant differences in lean body mass between participants with CMT and healthy sex and age-matched subjects were found in the legs (primarily lower legs). Due to the accentuated motor weakness and sensory loss in the lower limbs as CMT progresses, activities such as walking are difficult [3,4]. As a result, people with CMT may adopt a sedentary lifestyle which is known to increase body weight, muscle loss, musculoskeletal pain, and reduce functional capacity. In the present study the associations found for fat mass and walking ability suggests that a priority area for people with CMT is achieving and maintaining a healthy body fat range. Excess total body fat and in particular abdominal obesity is associated with cardiometabolic diseases including type 2 diabetes, hypertension, dyslipidemia, and coronary heart disease [47]. Waist circumference is often used as a surrogate marker of abdominal fat mass with increased risk associated with a waist circumference of >102 cm for males and >88 cm for females, respectively [48]. In the present study females exceeded their waist circumference cutoff and were found to have significantly greater fat mass compared to the group of age-matched healthy females. However, there were no differences in fat mass between males with CMT and the age-matched healthy males. This is in contrast to a previous study that reported greater % body fat in both males and females with CMT compared to healthy age-matched norms (37 and 44%, respectively) [49].

The results from the present study suggest that people with an earlier age of symptoms onset for CMT are able to maintain a healthier body fat level. The CMT participants with less fat mass also had a higher quality of life (physical component score) which is consistent for patients with other health conditions such as metabolic syndrome [50] and fibromyalgia [51]. Excess body fat is associated with a range of disabling musculoskeletal conditions [52] and has negative impact on physical function [53]. Also, it has been suggested that the risk of falls is increased for obese individuals because postural stability is compromised leading to limitation of movement [54]. This supports the present study findings of a trend for a significant correlation between android fat mass and falls efficacy scale scores. Also, the lack of significant correlations or trends with quality of life (physical and mental component scores) in participants with CMT in the present study is consistent with results from a previous study in healthy older men and women [55]. However, the lack of significant correlations or trends between lean body mass and falls efficacy scale scores is surprising based on greater muscle mass being associated with lower falls risk [56].

It is well known that resistance training is beneficial for increasing muscle mass. In one of the very few upper and lower body resistance training interventions involving people with CMT, Chetlin et al. [49] found no increase in the lean body mass of CMT participants following a home-based 12-week resistance training intervention despite improvements in strength and activities of daily living. However, the lack of significant improvement in lean body mass following the resistance training intervention is most likely attributed to the use of a low resistance training intensity (20–50% maximal voluntary contraction) rather than reduced resistance training adaptive capability in people with CMT. Based on total and regional lean body mass being associated with muscular strength and power in the present study, future resistance training interventions in people with CMT should consider using evidence-based prescriptions to maximise gains in lean body mass [57].

This study has several potential limitations that affect the inferences that can be made from the data. Firstly, the low number of participants was a strong source of bias in this study and it is possible that findings could be different if there were a larger number of participants. However, while the small sample size affects the generalisability of the results for people with CMT, it is likely that it contributed to the failure to detect more significant associations. Furthermore, not all the participants completed all the physical performance tests which may have affected the reliability for some of the data. Secondly, even though dual energy x-ray absorptiometry is recommended as a safe, noninvasive, inexpensive tool for managing patients with neuromuscular diseases [58], currently the precision of body composition analysis for people with CMT is unknown. Therefore some caution is required regarding the body composition results considering that the reliability of the dual energy x-ray absorptiometry measurements was not assessed. Thirdly, no correlations of muscular strength and power with lean body mass and fat mass were performed in the healthy sex and aged-matched participants. So it is unknown how the correlations found in the present study for the CMT participants would compare to healthy subjects. Finally, it is possible that the use of a modified Tinetti scale, that has not been validated, may have biased the falls efficacy scale scores.

**Conclusions**

We report for the first time that greater lean body mass and lower fat mass of specific body regions is associated with better physical performance in numerous tasks and higher quality of life in people with CMT. However caution is warranted because no correlation was found between body composition and CMT exam score. Furthermore, factors unrelated to CMT such as age-related sarcopenia may have influenced the associations found between measures due to the mean age of participants being >45 y. Future research should aim to substantiate these findings in a larger cohort, explore sex differences, and investigate whether resistance training and/or dietary interventions targeting favorable changes in body composition can lead to improvement in physical performances, quality of life, and clinical indicators in people with CMT.

**Disclosure statement**

The authors report no conflicts of interest.

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