Gait & Posture 62 (2018) 262–267

Contents lists available at ScienceDirect

Gait & Posture

journal homepage: www.elsevier.com/locate/gaitpost

Full length article

Gait and footwear in children and adolescents with Charcot-Marie-Tooth disease: A cross-sectional, case-controlled study

Rachel A. Kennedy[a,b,c,*], Jennifer L. McGinley[b,c], Kade L. Paterson[b], Monique M. Ryan[a,c], Kate Carroll[a,c]

[a] Department of Neurology, The Royal Children's Hospital, Parkville, Victoria, Australia
[b] Physiotherapy Department, Melbourne School of Health Sciences, The University of Melbourne, Parkville, Victoria, Australia
[c] Murdoch Children's Research Institute, Parkville, Victoria, Australia

ARTICLE INFO

Keywords:
Charcot-Marie-Tooth disease
Pediatric
Gait
Footwear
Gait variability

ABSTRACT

Objective: Children with Charcot-Marie-Tooth disease (CMT) report problems with gait and footwear. We evaluated differences in spatio-temporal gait variables and gait variability between children with CMT and typically developing (TD) children, and investigated the effect of footwear upon gait.
Method: A cross-sectional study of 30 children with CMT and 30 age- and gender-matched TD children aged 4–18 years. Gait was assessed at self-selected speed on an electronic walkway while barefoot and in two types of the child's own footwear; optimal (e.g., athletic-type runners) and suboptimal (e.g., flip-flops).
Results: Children with CMT walked more slowly (mean (SD) −13.81 (3.61) cm/s), with shorter steps (−6.28 (1.37) cm), wider base of support (+2.47 (0.66) cm; all $p < 0.001$) and greater base of support variability (0.48 (0.15) cm, $p = 0.002$) compared to TD children. Gait was faster in optimal footwear than suboptimal (−7.55 (1.31) cm/s) and barefoot (−7.42 (1.07) cm/sec; both $p < 0.001$) in the combined group of children. Gait in suboptimal footwear was more variable compared to barefoot and optimal footwear. Greater base of support variability and reduced balance was moderately correlated for both groups (CMT and TD).
Conclusion: Gait is slower with shorter, wider steps and greater base of support variability in children with CMT. Poor balance is associated with greater base of support gait variability. Suboptimal footwear negatively affects gait in all children (CMT and TD), which has clinical implications for children and adolescents with CMT who have weaker feet and ankles, and poor balance.

1. Introduction

Children and adolescents ("children") with Charcot-Marie-Tooth disease (CMT) often report gait difficulties including problems with balance, frequent trips and falls [1], and experience problems walking in certain footwear types. A genetically and clinically diverse group of neuropathies, the primary lower limb impairment of CMT is progressive distal weakness affecting the feet and ankles [2]. A systematic review of gait in paediatric CMT found limited evidence, from a small number of barefoot gait studies, that children with CMT walk more slowly than their typically developing (TD) peers, with shortened step length, reduced calf muscle power at push-off and foot drop in swing [3].

Additionally, no study has evaluated the effects of CMT on gait variability. Normal gait is intrinsically relatively stable in linear preferred-speed walking. The magnitude of variation between one step and the next is regarded as a sensitive indicator of dynamic balance [4]. Increased step-to-step gait variability is associated with gait problems and poor balance in children with other neurological disorders [5] and older adults who are at increased risk of falls [4]. Associations between gait, balance and step-to-step variability have not been evaluated in children with CMT.

Footwear and its influence on gait are of key interest in children with health conditions that affect foot and ankle strength and gait function. Typically, children wear shoes in their everyday environments, and footwear is often discussed between families of children with CMT and health professionals. At times, some children may wear

Abbreviations: 6MWD, six-minute walk distance; 6MWT, six-minute walk test; BMI, body mass index; BOS, base of support width; BOT, Bruininks-Oseretsky Test of Motor Proficiency; CMT, Charcot-Marie-Tooth disease; CMTPedS, Charcot-Marie-Tooth Pediatric Scale; DS%GC, double support time as a percentage of gait cycle; FAS, Footwear Assessment Score; FPI, Foot Posture Index; SS%GC, single support time as a percentage of gait cycle; TD, typically developing

* Corresponding author at: Department of Neurology, 3West Clinical Offices, The Royal Children's Hospital, 50 Flemington Rd, Parkville, Victoria, 3052, Australia.
E-mail addresses: rachel.kennedy@rch.org.au (R.A. Kennedy), mcginley@unimelb.edu.au (J.L. McGinley), kade.paterson@unimelb.edu.au (K.L. Paterson), monique.ryan@rch.org.au (M.M. Ryan), kate.carroll@rch.org.au (K. Carroll).

https://doi.org/10.1016/j.gaitpost.2018.03.029
Received 7 August 2017; Received in revised form 6 February 2018; Accepted 18 March 2018

Fig. 1. Examples of optimal and suboptimal footwear.

footwear considered to have suboptimal fit (e.g. flip-flops) due to cultural factors and/or personal preferences. In TD children, athletic-type runners and school shoes have been shown to have a positive impact on gait, as children walk faster, with longer steps and reduced cadence relative to barefoot [6]. To date, there has only been one study investigating the effects of footwear on gait in CMT. A single report of a 55-year-old woman with CMT reported that well-fitting orthopaedic shoes increased walking speed and reduced falls [7]. No study has investigated effects of footwear on gait in children with CMT.

The aims of this study were to (i) evaluate differences in spatio-temporal gait variables, and step-to-step variability, between children with CMT and TD children; and (ii) investigate the effect of footwear on spatio-temporal gait variables and variability. We hypothesised that gait in children with CMT would be slower and have greater step-to-step variability than TD children, and that gait in optimal footwear would be faster with less step-to-step variability compared to gait in suboptimal footwear.

2. Methods

This was a cross-sectional case-controlled study in children with CMT and age- and gender-matched TD children. Ethical approval was gained from The Royal Children's Hospital (33272) and The University of Melbourne (1441639).

2.1. Participants

Thirty children aged 4–18 years with confirmed genetic or clinical diagnosis of CMT were recruited from the Neuromuscular Clinic at The Royal Children's Hospital, Melbourne, Australia between March 2014 and July 2016. Thirty age- and gender-matched TD children were recruited from Melbourne and surrounding regions. Parent/guardian informed consent and participant assent when appropriate were obtained for all participants. Inclusion criteria were the ability to walk > 75 m without gait aids (orthotics permitted). Exclusion criteria included developmental disorders, other neuromuscular/musculoskeletal disorders that could affect gait, and lower limb injury or surgery in the preceding 6 months.

2.2. Procedure and assessments

A single study visit comprised assessments of anthropometry, gait, footwear and disease severity.

2.3. Descriptive data

Anthropometric characteristics included weight, height and body mass index (BMI). Leg length was measured in supine from the anterior superior iliac spine to the medial malleolus [8]. Foot posture was assessed using the Foot Posture Index (FPI) [9].

2.4. Gait

Spatio-temporal gait assessment was conducted over a 4.27 m electronic walkway (GAITRite®, CIR Systems, Inc., Franklin, USA) with a two-metre zone at each end to capture steady state gait. The GAITRite® is a valid and reliable tool in both TD and children with motor impairment [10,11]. Six walk trials per footwear condition were recorded at self-selected speed.

Gait was assessed in three footwear conditions: barefoot, suboptimal and optimal footwear (with orthotics if normally worn). Gait variables included speed, step length, step time, cadence, base of support width (BOS), and single and double support time as a percentage of the gait cycle (SS%GC and DS%GC). Step-to-step variability was calculated as the standard deviation (SD) for the group of individual steps taken by each participant for the combined six walk trials per footwear condition. This was calculated for step length, BOS and step time.

2.5. Footwear

The Footwear Assessment Score (FAS) is a valid and reliable measure of children's footwear that assesses factors affecting fit, including shoe type, composition and fastening method, length, width, heel slip and height, and wear [12,13]. Scored out of 15, higher scores indicate better fit. Participants were assessed in their own optimal and suboptimal footwear. Optimal was defined as footwear that enclosed the foot, had a heel cup, fastened firmly with a low heel height (< 25 mm). Examples include athletic-type runners, leather school shoes or boots with laces, Velcro® or buckles. Suboptimal was defined as footwear that slipped on, didn't fasten securely around the foot, didn't enclose the heel, or a high heel height (≥ 25 mm). Examples of suboptimal footwear include flip-flops, slip-on shoes, ballet flats, slippers, wedge-heeled shoes or elastic sided boots. A photographic guide of appropriate footwear was provided (Fig. 1).

2.6. Measures of disease severity, strength, balance and gait function

Disease severity in children with CMT was measured using the CMT Pediatric Scale (CMTPedS), an 11 item valid and reliable scale [14]. Typically developing participants were assessed on selected lower limb