



## Powered standing wheelchairs promote independence, health and community involvement in adolescents with Duchenne muscular dystrophy

Nitamarie Vorster<sup>a</sup>, Kerry Evans<sup>b</sup>, Nada Murphy<sup>a</sup>, Maina Kava<sup>c,d</sup>, Anita Cairns<sup>e</sup>, Damian Clarke<sup>f</sup>, Monique M. Ryan<sup>g</sup>, Aris Siafarikas<sup>a,h</sup>, Peter W. Rowe<sup>c</sup>, Stephanie Parkinson<sup>i</sup>, Odette Gaynor<sup>j</sup>, Linda Chiu<sup>k</sup>, Janine Anderson<sup>l</sup>, Klair Bayley<sup>m</sup>, Peter Jacoby<sup>a</sup>, Donna Cross<sup>a</sup>, Jenny Downs<sup>a,i,\*</sup>

<sup>a</sup>Telethon Kids Institute, University of Western Australia, PO Box 855, West Perth, WA 6872, Australia

<sup>b</sup>Novita, 171 Days Road, Regency Park, SA 5010, Australia

<sup>c</sup>Department of Neurology, Perth Children's Hospital, 15 Hospital Avenue, Nedlands, Locked Bag 2010, Nedlands, WA 6909, Australia

<sup>d</sup>School of Paediatrics and Child Health, University of Western Australia, Australia

<sup>e</sup>Department of Neurology, Queensland Children's Hospital, PO Box 3474, South Brisbane, QLD 4101, Australia

<sup>f</sup>Department of Neurology, Women's and Children's Hospital, 72 King William Rd, North Adelaide, SA 5006, Australia

<sup>g</sup>Department of Neurology, Royal Children's Hospital, 50 Flemington Rd, Parkville, Vic 3052, Australia

<sup>h</sup>Department of Endocrinology, Perth Children's Hospital, 15 Hospital Avenue, Nedlands, Locked Bag 2010, Nedlands, WA 6909, Australia

<sup>i</sup>School of Physiotherapy and Exercise Science, Curtin University, GPO Box U1987, Perth, WA 6845, Australia

<sup>j</sup>Therapy Focus, PO Box 20, Bentley, WA 6982, Australia

<sup>k</sup>Rocky Bay, 13 Baling Street, Cockburn, WA 6164, Australia

<sup>l</sup>Montrose Therapy and Respite Services, Level 2 20 Innovation Parkway, Buddina, QLD 4575, Australia

<sup>m</sup>Save Our Sons Duchenne Foundation, Shop 3/570, New Canterbury Road, Hurlstone Park, NSW 2193, Australia

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### Abstract

Duchenne muscular dystrophy is a common neuromuscular disorder involving progressive muscle weakness. A powered wheelchair standing device provides capacity to stand despite increasing muscle weakness. This study used qualitative methods to explore how adolescents with Duchenne muscular dystrophy used a powered wheelchair standing device in their daily lives. Semi-structured interviews were conducted with 12 adolescents, 11 parents and 11 teachers. Qualitative thematic analysis using a grounded theory framework was conducted to identify emerging domains. "Capacity to be able" was the central theme that emerged across the dataset: the introduction of the powered wheelchair standing device at a time when motor skills were declining enabled the adolescent to maintain and sometimes extend his independence. There were four underlying themes including (1) Independence, (2) Health, (3) Comfort, and (4) Community belonging and involvement. Each theme was illustrated in data collected from adolescents, parents and teachers. The device appeared to mitigate some of the challenges of progressive muscle weakness by providing the option for the individual with Duchenne muscular dystrophy to choose when and where to stand for participation in a range of activities, beyond what would be possible with existing therapeutic regimes involving standing frames. Crown Copyright © 2019 Published by Elsevier B.V. All rights reserved.

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\* Corresponding author at: Telethon Kids Institute, University of Western Australia, PO Box 855, West Perth, WA 6872, Australia.

E-mail address: [jenny.downs@telethonkids.org.au](mailto:jenny.downs@telethonkids.org.au) (J. Downs).

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## 1. Introduction

Duchenne muscular dystrophy (DMD) is a common neuromuscular disorder occurring in approximately 1 in 5000 live male births [1] and caused by a mutation on the dystrophin gene located on the X chromosome [2]. The dystrophin gene encodes the protein dystrophin, which is essential to healthy muscle growth and functioning. In its absence, muscles deteriorate and break down [3]. Health and functioning are subsequently compromised. Mobility becomes progressively more difficult with increased risk of musculoskeletal deformity [4]. Corticosteroids, which are the standard of care for DMD, contribute to lower bone density with long-term use [5]. Respiratory muscle weakness causes chronic respiratory insufficiency [6]. Pain and the risk of pressure areas due to limited movement are often underreported [7].

Adolescence is typically characterised by increasing independence, the physical changes of puberty, and marked emotional and social development with increased propensity to mental health difficulties [8]. In DMD, loss of ambulation and increased dependence upon carers for many aspects of daily living coincides with adolescence and transition to secondary school. This is an emotionally difficult time and as their peers are gaining strength and independence, those with DMD are losing theirs. They must adapt to altered ways to functioning and participation in activities of personal importance including those with their peers [9].

There is a general belief that assisted standing for individuals who have limited or no ability to walk is beneficial for physical health. Individuals with spinal cord injury have reported health benefits such as better bowel and bladder function, reduced spasticity and improved sleep [10,11]. In small numbers of non-ambulant children with cerebral palsy, standing regimens have been associated with increased hamstring length [12]. There has been scant consideration of the role of standing wheelchairs in promoting mental health. Standing frames and wheelchairs with standing function form part of recommended practice for individuals with DMD provided contractures are not too marked and the standing device is tolerable [13]. A Powered Wheelchair Standing Device (PWSD) allows standing within the powered wheelchair structure and provides opportunities for the individual to stand by controlling a joystick and without need for a transfer. There has been limited evaluation of the PWSD to date with one single subject design study investigating a standing regimen, ranging in duration from 1.3 to 3.3 h per week over six to 12 months, in four adolescent boys with DMD [14]. Three of the adolescents tolerated the standing regimen well, bone density remained unchanged and there were some increases in hip and knee flexor muscle length [14].

PWSDs are increasingly being used by individuals with DMD but there has been no systematic evaluation of how the standing function of the PWSD is used by individuals with DMD for health, wellbeing and participation in social, educational and recreational activities. The aim of the current

Table 1

Characteristics of the adolescents with Duchenne muscular dystrophy ( $n = 12$ ).

	Number
Mobility level when received the PWSD	
Walking around the house without assistance	4
Walking around the house with help	2
Walking a few steps with help	3
Unable to walk	3
Mobility level at the time of interview	
Walking around the house without assistance	4
Walking around the house with help	2
Walking a few steps with help	1
Unable to walk	5
Use of night splints	9
Previous fracture	6
Medications	
Corticosteroids <sup>#</sup>	12
Vitamin D supplement	10
Calcium supplement	8
Bisphosphonates	4
Daily pain	12
Location of residence	
Urban	11
Rural	1

<sup>#</sup> 7 prescribed deflazacort, 5 prescribed prednisolone.

study was to explore the use of the PWSD in daily life in DMD using qualitative methods.

## 2. Participants and methods

This qualitative study used semi-structured interviews to collect data. Data were collected from multiple sources including the adolescent with the PWSD, his parent and teacher to enable triangulation [15]. Each respondent provided informed consent to participate in the study.

### 2.1. Participants

Twelve adolescents with a genetically-confirmed diagnosis of DMD were recruited once their therapists had recommended that they be provided with a PWSD (Levo C3; [www.levo.net.au](http://www.levo.net.au)). At the time of recruitment, their mean (SD) age was 11.6 (2.2) years, EK score [16] was 5.5 (2.8) and BMI was 25.0 (2.7). At recruitment, nine adolescents were in the late ambulatory phase and three were non-ambulant. At interview, 7 adolescents were still in the late ambulatory phase and 5 were non-ambulant, two losing the ability to walk after they had received the PWSD. All adolescents invited to participate in the interview were recruited. Other descriptive characteristics are presented in Table 1. Eleven parents were interviewed; each was the mother of an affected adolescent. Teachers were also recruited: 10 teachers and one education assistant were female and one teacher was male. One adolescent only used his PWSD at school being unable to transport the PWSD in the family car, but the remainder used the PWSD in home, school and community settings.

Table 2

Semi-structured stem and leaf telephone interview questions for the adolescents and their parent and teacher.

*Core open-ended questions*

1. How did you/he feel when first using the PWSW?
2. How do you use/does he use the PWSW?
3. When things are going well in standing, what sorts of things are happening?
4. Have there been any problems with the PWSW?
5. Would you recommend a PWSW to another boy with Duchenne muscular dystrophy?

*Additional core questions for the adolescent*

1. How does your body feel during standing?
2. What do you do in standing at home?
3. What do you do in standing at school?
4. What do you do in standing with your friends?
5. What do you do in standing when you go out?

*Additional questions for the parent*

1. Has standing helped his body to stay flexible?
2. Has the PWSW affected how you supported him with personal care?
3. Has the PWSW affected what you son does at home and when going out?

*Additional questions for the teacher*

1. How has he participated in educational activities in standing?
2. Has being able to stand affected participation at school?
3. Did the PWSW affect the delivery of educational activities?

*Probing questions*

1. What was going on when this happened?
2. What would I see if I was there?
3. How do you know?
4. Why do you think that is?
5. How did you/he react?

## 2.2. Interviews

Interviews were conducted between December 2015 and June 2018 by one researcher (NV) who is a physiotherapist with extensive neuromuscular expertise and not involved in the delivery of the participant's clinical care. Interviews were held individually over the telephone or face to face, taking approximately 20 min with the adolescents and 30 min for primary caregivers and teachers. Interviews were conducted a median of 8 months (range 4 to 5 months) after the adolescent received his PWSW. Within participants, the teacher interviews were conducted a median of 2 months (range 1 to 8 months) after the adolescent and parent interviews. With participant permission, the interviews were digitally recorded and then transcribed.

A semi-structured interview schedule was developed, informed by the International Classification of Functioning, Disability and Health (ICF) model of disability [17]. The questions included aspects of health, activities, and participation in the home, school and community, and were provided to the respondents prior to the interview. Adolescents were also asked about their reasons for and feelings about using the PWSW as well as any problems. Additional questions for primary caregivers and teachers asked for specific examples of behaviours that were observable to support their proxy report of the adolescent's satisfaction, enjoyment or problems (e.g. "How do you know this?"). Lastly, respondents were asked if they would recommend the chair to other individuals with DMD. The interview schedule is shown in Table 2.

Member checking was used to promote the accuracy of the information shared by participants, giving the opportunity to add information that was overlooked or to edit information. The transcribed interviews were provided to participants for member checking which was completed by nine (75%) of adolescents, eight (73%) of parents and five (50%) of teachers.

## 2.3. Data analysis

A grounded theory approach was used where themes were fully informed by the data rather than any pre-existing theory or framework, enabling key themes to evolve naturally from the interview transcripts. Coding was subject to constant assessment and contrasting of different ideas and observations as new data were analysed [15]. To fully understand the dataset, transcripts were read and re-read by NM and NV and data were then coded using NVivo [18]. Themes of PWSW use as well as elements within the themes were ascertained. Three researchers (NM, NV, JD) reviewed and interpreted the themes and coding within the transcripts. Joint discussion was then used to establish a consensus of the primary themes and elements within themes.

## 2.4. Ethical considerations

Ethical approval of this study was prospectively reviewed by the Human Research Ethics Committee at the Child and Adolescent Health Services, Western Australia (2015012EP); the Lady Cilento Children's Hospital, Queensland (HREC/16/QRCH/27); the Women's and Children's Hospital, South Australia (HREC/16/WCHN/103); the Royal Children's Hospital, Victoria (36372A); as well as Departments of Education in Western Australia, South Australia and Victoria, and local Catholic Education authorities in New South Wales and Queensland. Participants provided consent to participate in this study.

## 3. Results

"Capacity to be able" was the central theme that emerged across the dataset. The introduction of the PWSW at a time when gross motor skills were declining enabled the adolescent to maintain and sometimes extend his independence. Fig. 1 illustrates an adolescent participating independently in an

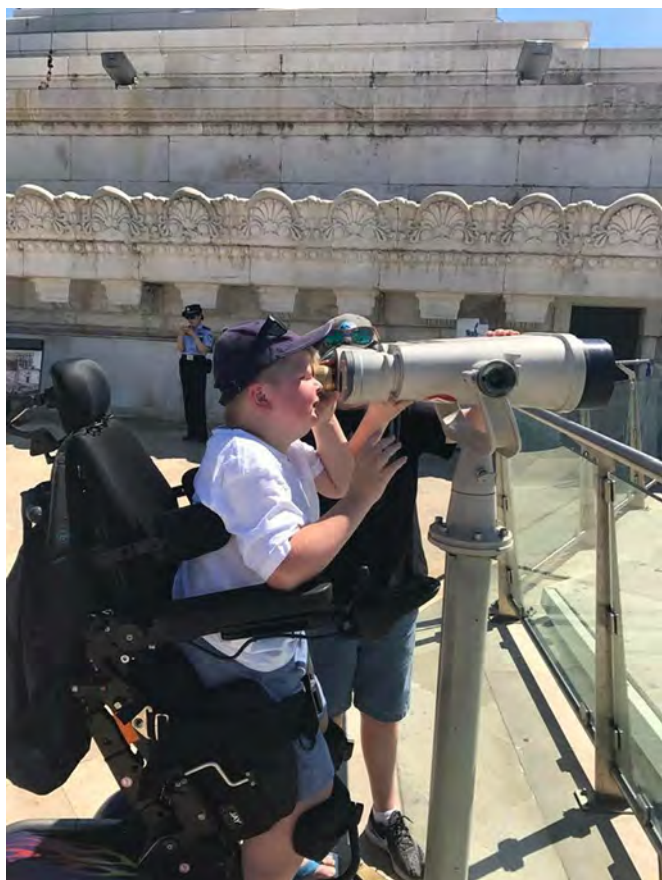


Fig. 1. Adolescent using a PWS to stand independently and access a community resource.

activity that required the standing position, similar to his peers. Four underlying themes emerged from this overarching notion: (1) Independence, (2) Health, (3) Comfort, and (4) Community belonging and involvement. These underlying themes were illustrated in data collected from adolescents, primary caregivers and teachers, described below with additional sample quotes shown in Tables 3–6.

### 3.1. Independence

#### 3.1.1. Independent actions

The adolescents described being able to do things without having to ask for assistance. For example, they could choose when to stand or sit down again to adjust their comfort level, reach for objects, assist with domestic tasks such as cooking, help themselves to snacks or refreshments, or stand when purchasing goods in shops. One summed this up as life changing. *“It increased my independence because I am able to do more things on my own rather than getting other people to do it.”*

Increased independence was similarly observed by parents who observed that this contributed positively to their son’s self-esteem, reduced demands for their care including the need for manual lifting or hoisting, and as the adolescents themselves had described, facilitated an active contribution to

domestic tasks. Teachers reported that the adolescents used the standing function to participate in classroom activities that were usually conducted in standing such as art classes and in the science laboratory. Abilities in standing included reaching independently for library books, collecting needed equipment, using smart boards and generally functioning more independently. Teachers observed that standing was associated with greater self-confidence and one teacher remarked that the individual’s increased independence meant she had more time to teach.

#### 3.1.2. Independence in personal care

Some adolescents retained independence with some personal care tasks whereas other were fully dependent at the time of interview. Many made use of the standing function to use a bottle or toilet to pass urine but did not comment in detail. One adolescent said *“Yes, I use standing in the bathroom before I go to school to brush my teeth and do my hair”*.

Most parents and teachers provided greater detail regarding positive impacts on personal care activities. For one parent, the chair enabled *“great functional improvement”* because of her adolescent’s capacity to pass urine into a bottle or toilet in standing and to reach for objects such as toothbrush. Two adolescents were independent for toileting as they could walk into the toilet whereas the others passed urine in standing whilst in the PWS, although one parent reported that her son preferred not to use the standing function in this regard. When at school, five regularly used the standing function to pass urine and this was considered extremely valuable. Outings were also less problematic – *“And when we go out, I find the chair a lot easier. He doesn’t have to go right up into standing position, even if he just stands up a little bit it is still easier.... That is much easier than doing it from a seated position.”* Other personal care routines were generally faster, required less use of hoists and transfers, and faster toileting at school also meant less missed class time.

### 3.2. Health

The standing wheelchair was perceived as contributing favourably to both physical and mental health.

#### 3.2.1. Physical activity & wellbeing

Increased opportunity for physical activities was not commonly reported by the adolescents although some used the chair to engage in activities such as Frisbee or simply stood to stretch their legs. One adolescent used the chair diligently for standing activities including extending his duration of standing to build his physical condition and enjoyed feeling that his breathing was easier in standing.

Parents were generally aware of the potential for the chair to support their son’s physical health and observed better circulatory and digestive functioning. The PWS supported regular stretching and skin pressure management. One parent observed that the standing function had helped her son recover physical condition after a serious bout of illness. In contrast,

Table 3

Sample quotes for the theme of independence, from the teenagers, their parents and teachers.

	Physical activity and wellbeing	Personal care
Adolescent with DMD	<p>"I was able to stand up and order my own ticket at the counter. It is easier than having to get my friends to pay for it and give me change or anything. So, I do that all myself."</p> <p>"I can now reach higher places at home. Like one day mum found me getting a packet of chips out of the cupboard."</p>	<p>"Yes, I use it in the bathroom before I go to school to brush my teeth and do my hair."</p> <p>"I used it a couple of times, standing up, to go to the toilet. Not at home though. Outside."</p> <p>"Standing to go to the toilet is great and very important because it helps me to be independent at school."</p>
Parent	<p>"So, as I said before it enabled him to reach higher places. I found him getting chips out of the cupboard. Helping himself to food which is great. I haven't actually asked him to do anything like hang out washing but he could do it if he was asked"</p> <p>"The timing of the chair's arrival was good as he was going off his feet, so it gave him the opportunity to continue with the activities he was capable of when he was still walking, so maintained his independence."</p>	<p>"It was a lot easier using the urine bottle in standing as no need to lift him. He is more independent with dressing, adjusting his clothes and grooming. It is easier as the chair is not as low and I do not have to bend down low e.g.: brushing his teeth"</p> <p>"Probably the main thing for us is the toileting. Definitely being able to stand, he stands and uses a urine bottle. That is definitely something that makes it a lot easier."</p>
Teacher	<p>"From an overall point of view it has been positive for him, for his communication, for his social engagement, participation in classroom and for him to have control and choice over whether he is standing or sitting as what is the most suitable for his circumstances at the time."</p> <p>"It has improved and maintained his independence as he can reach the chalk board, get equipment out of the cupboard himself and work at different surfaces like all the other kids."</p>	<p>"When going to the toilet he is able to stand to urinate into a bottle making it much easier as he does not have to be transferred from a chair to the toilet seat".</p> <p>"It is also helping him to keep his independence with toileting as the standing facility enables him to transfer more easily to the toilet."</p>

Table 4

Sample quotes for the theme of health, from different teenagers, their parents and teachers.

	Physical activity and wellbeing	Mental wellbeing
Adolescent with DMD	<p>.. played games in standing with my iPad, reading and watched TV and different things and moved around in my chair."</p> <p>"It feels good to stand up and breathe as I get tired later in the day. Then I feel "folded up" after sitting too long. Standing helps me to stretch out around my chest and abdomen and can breathe easier."</p>	<p>"I was happy that I was able to stand up easier whenever I wanted and to be taller than I was before."</p> <p>"I felt a bit nervous and excited, and after about a year's wait the "royal blue" coloured chair arrived in March 2015."</p>
Parent	<p>"After a recent hospitalization with gastroenteritis he was quite deconditioned and getting back in to the chair made a really positive difference for him to recuperate faster."</p> <p>"Yes, when we went outside and we were playing outside and we had a few games. We messed around with the cricket and tried to do cricket standing up. That was funny. It was interesting and a fun for him to try."</p>	<p>"I do not like to do it in front of people."</p> <p>"His behavior is probably that frustration I guess. It is probably the biggest thing. He doesn't get as frustrated now because he has the independence to be able to do what he wants to do."</p> <p>"So I think for him that he his self-conscious enough being in a wheelchair that he feels that he stands out. I think for him, although we try to encourage him, he doesn't want to stand out at school because he thinks it makes him even more different."</p>
Teacher	<p>"During PE lessons, he is able to stand for activities like table tennis, archery on a recent camp trip and activities like golf. Normally there would be lots of activities that he cannot participate in."</p> <p>"Teachers could make a point of including some 'standing time' for the whole class to stretch out ligaments and internal organs for the student in the chair."</p>	<p>"This was the first thing he did when he received his new chair and it certainly brought a big smile to his face and ours when we saw him standing to access these books. This was something they did not even think about and it was like him turning around and 'saying look at me' I can reach the book on the top shelf."</p> <p>"I have talked straight out to him and he said he felt embarrassed and does not like people looking at him. He wants to blend in and in standing he feels he stands out. He is not super tall, but thinks he is going to feel that everybody is looking at him so that is going on inside his head."</p>

Table 5

Sample quotes for the theme of comfort, from different teenagers, their parents and teachers.

	Managing comfort and fatigue	Pain
Adolescent with DMD	"It is a positive in that I can readjust myself to get comfortable, rest in the recline position, stretch and it is easier with toileting."	"It is easier to stand up and stretch my back a bit I guess."  "If I stand for too long, my feet hurt, that is to be expected I think, but nothing that I didn't expect."
Parent	"He has realized that on some days when he is really tired and struggling he can still stand. He will lean a bit back and still get a good stretch without being totally upright. He lays down flatter and watches movies as well. Going the other way is good for him as well."	"Yes, he does experience some tingling if he has been standing for ten minutes or more. He tends to recline himself back ever so slightly to take some of the weight. That helps a little bit and he will still lower back down again."  "He actually re-fractured his back about 6 months after the original fracture. Just that being able to relieve pain by just changing position was huge. But, no he has not had pain from the chair."
Teacher	"He is physically more comfortable and able to work longer before he fatigues."  "So it conserves his energy and he is a very aware of that."	"Yes, he lies back reclining in his chair when he is uncomfortable. He at time moves armrests up or take footrests off. Sometimes he goes up to almost standing but not all the way up."  "No occasionally there are the sore points that are on his shins - he has said that it rubs a little bit."

Table 6

Sample quotes for the theme of community belonging and involvement, from different teenagers, their parents and teachers.

	Community belonging and involvement
Adolescent with DMD	"We went to my year eleven semi-formal and I was able to stand up and talk to my friends. It is a lot easier in a party situation otherwise I would not have been able to hear what any one was saying if I was sitting down."
Parent	"Yes, sometimes my mom will get me to stand and help her with the cooking in the kitchen."  "I was amazed we turned up to this family function .. This is my family who he doesn't see very often. And the next thing I hear my uncle came in saying: "He has just been standing up in front of everyone in his chair. He is showing everybody what it can do. He is showing the headlight and this, and tilting it. He has explained everything to everybody" and I am like "but he doesn't usually talk to people!"  "He was able to interact with people in standing during shopping, ordering at a counter etc. He was able to participate in school activities in standing face to face e.g. Speech giving, performing in the Easter play."
Teacher	"Yes, it is absolutely positive and it is not only good for him but it is good for the other boys as well. It just takes a barrier away really. Everyone doesn't have to think about whether I should be sitting down or standing up or what's comfortable. You know how he can adapt his circumstances to suit the social circumstances so therefore he is in control."  "He could go outside throw a ball around and shooting hoops in standing at the same eye level as the other students or playing games in the library/class room having the ability to do it in standing as the other students."

another parent observed that her son did not enjoy stretching in his chair when standing.

At school, most adolescents stood when participating in physical education activities such as table tennis, archery and golf although others showed very little interest in sport. One teacher was proactive and created opportunities for her student to stand during the classroom schedule, hoping it would lead to him "*stretching ligaments and [reducing pressure on] internal organs*".

### 3.2.2. Emotional wellbeing

Most adolescents said they had felt happy or excited when receiving the chair and enjoyed doing things without seeking help from others. Some adolescents reported specific experiences that for them had felt extremely special. These included being a part of a cheer squad, being able to go fast in the chair when standing, and being given special status in their peer group with affection and a nickname, "*Ironman*".

In contrast three adolescents reported being self-conscious of standing in front of others and did not enjoy appearing different and being the centre of attention. One individual was nervous at first with the sensations that standing provoked but adapted to this with practice. Another worried about standing and did not adapt to feeling safe with practice over time.

Parents observed that the greater independence and capacity to be in control provided their son with more confidence and improved self-esteem. They observed their son to be happier and more content, more mature and self-confident. Parents described fewer episodes of grumpiness, frustration and anger. However, they also reported negative emotions that were associated with the PWSD. Some parents observed their son as reluctant to draw attention to himself or being anxious about being knocked or knocked over in standing. Another parent reported that her son perceived the PWSD as a set-back - "*I am going to be in this for the rest of my life – this is the end of my walking*".

In the main, teachers observed that the adolescents became more confident as they needed less help from others and could access resources of their own accord. One boy was described as more proactive and spoke positively about his plans for his future. Some adolescents had a sense of pride in the chair and were happy to show others how it worked. Teachers also noted that for some adolescents, initial anxiety, apprehensiveness and embarrassment needed to be overcome. One teacher was aware that the introduction of the chair had been confronting for the individual with DMD, who was familiar with another student using a wheelchair fulltime because of a severe disability.

### 3.3. Comfort

#### 3.3.1. Management of comfort and fatigue

Adjusting the chair independently meant that adolescents could manage their own comfort – “*It made my legs feel nice*” when standing. The tilt and recline function enabled the adolescent to lean back and recline whilst still “standing” to achieve stretch and comfort but working within available energy levels. This option was often used in the evenings.

Another parent described the processes of her son finding his own position of comfort as a series of position changes: “*In terms of getting comfortable, my son fidgets and you hear the beeps, you know he is up, he is down, and then forwards and backwards, and you know he is moving around a lot.*” As another parent said, “*He is able to do a lot without tiring*” and this flexibility often resulted in fewer complaints about discomfort. Teachers also found adolescents would manage fatigue and discomfort in the classroom by adjusting the chair to a reclining position in both standing and sitting so that they did not tire as quickly during the school day.

#### 3.3.2. Pain

Some adolescents said commencing use of the standing wheelchair had not been associated with any additional pain whereas others reported foot discomfort when first using the standing function, resolved by sitting and/or reducing the duration of standing. Knee pain was usually resolved with adjustments being made to the knee-pads and leg blocks, highlighting the importance of monitoring comfort levels particularly in the initial stages of use and as the adolescent grows. Varying tilt and chair height was also used to manage pain. “*He tends to recline himself back ever so slightly to take some of the weight. That helps a little bit and he can also lower back down again.*”

Two of the individuals experienced fractures in the lumbar spine, each associated with steroid induced osteoporosis [19]. For one, “*being able to relieve pain by just changing position in the standing wheelchair was huge*” but for the other, any movement including using the standing function exacerbated his acute fracture pain.

### 3.4. Community belonging & involvement

Another outcome was the capacity to stand during interactions and activities with peers, perceived by the

adolescents as important for their sense of belonging to their peer group. Adolescents spoke of advantages to being at eye level with their friends, hearing conversations, being able to see more clearly, and feeling better able to cope in crowded situations. One boy stood when on an outing “*to increase my height to play sideshow alley games and see the shows better in a crowd*”. Another described how he used standing when joining in with activities with his friends: “*I stand up at lunchtime with my friends to talk to them easier. And we had a cheer off for Grade 12s. When the Grade 12s leave school, the Grade 11s cheer them off and I had my chair alongside the other boys and put my arms around them. That was really good because I was front and centre and cheering with the group.*”

Parents also observed that the standing function facilitated their son’s communication with others and that better hearing was facilitated by being “*eye to eye*”. The boys mostly experienced a positive reaction from people who were interested in the chair. Following the school dance: “*After that evening he was talking about being able to hear the boys better in that party situation where there was music and excitement and noise, it was far better for him to be at their level. And psychologically very good for him to be equal to them or feel equal to them. Also to communicate and speak with his partner and to go out on to the dance floor and dance with her, out there, or to be able to go over and get them a drink. It made him feel less disabled I think.*”

Teachers’ observations also suggested that being able to stand had a positive impact on the boy’s sense of wellbeing, fun and enjoyment, and being part of his peer groups. “*He was not in my class last year but he seemed to be lonely. Now he talks to a lot of people and is more independent getting himself around and join in with the kids*”. Standing was used for both teaching and ceremonial activities at school. “*When students are asked to stand, he can also stand with the rest of his class. He was excited when he could stand for the National Anthem at assemblies*”. School drama performances often took advantage of the capacity to stand. Some teachers also paid attention to helping the adolescent learn how to use his chair safely in crowds and to be aware of others around him.

## 4. Discussion

Loss of walking imposes a substantial challenge on young adolescents with DMD. Adolescence, defined by the World Health Organisation as the period ranging from 10 to 19 years [20], is a time of physical change with strong desire for independence and increased need for social relationships [21,22]. For most young people, this period is a healthy, happy experience, however for some it can be characterised by health, social and academic challenges. The quality of peer relationships is described as one of the strongest indicators of current and future psychological health in adolescents [23]. Our overall finding was that the PWSD intervention provided “*capacity to be able*” which mitigated some of the

multiple challenges for adolescents with DMD in achieving independence, mental and social wellbeing in their daily lives.

Adolescents with DMD need to be independent but simultaneously, there is increasing dependence on parents and carers because of progressive muscle weakness. The PWSD allowed continuing independence for daily tasks or components of daily tasks including providing the individual with control over the decision of when he wanted to stand. This is consistent with other studies, in which use of electronic aids such as controls for doors, lighting and computers was associated with greater independence and autonomy in persons with DMD [24]. The most oft-discussed personal care activity was using the standing function to pass urine. Urine flow and residual bladder volume in boys in early adolescence are similar whether voiding occurs in sitting or standing [25]. However, the opportunity to pass urine in standing appeared associated with other advantages. Whilst the standing function enabled some adolescents to manage toileting independently for longer, passing urine in standing was procedurally easier and faster for all. The adolescent felt more dignified and might be less likely to postpone voiding [26] or experience incontinence [27]. The acceptance and utility of standing frames can be limited by the burden of transfers into the standing frame as the child becomes heavier. For caregivers, the standing function reduced the need for transfers that can be particularly difficult when in confined spaces in the community. It is possible that an additional benefit of the standing wheelchair could include protection of caregiver health and safety.

Physical activity is a continuum from sedentary behaviour to high intensity activities. Standing is considered a light physical activity in individuals with cerebral palsy [28] but energy expenditure and cardiac effects of standing in DMD are not known. Participants reported sensations of stretching and sometimes easier breathing suggesting physiological effects but these also need to be quantified. Intertwined with healthy activity is comfort. Most children and adolescents with DMD experience frequent pain, especially in the back and legs [7,29]. Pain [4] and fatigue [30] align with loss of mobility and can detract from quality of life. Most adolescents in our sample used the PWSD to adjust their position both for comfort and to manage fatigue. The standing wheelchair provided opportunity for independent pain management whilst allowing fuller functioning and participation and could be considered an adjunct to pharmaceutical pain management methods. Further research is necessary to quantify these parameters also.

Many mental health problems such as depression, anxiety and stress emerge during adolescence [8,31] and place young people at an increased risk for social and health difficulties that can persist into adulthood [32]. Emotional wellbeing appears particularly vulnerable in younger adolescents with DMD as walking becomes more limited [33]. Notably, some parents reported their son's improved mood associated with his greater autonomy when performing tasks such as finding his own snacks in the pantry cupboard, purchasing a ticket to the theatre, or better quality participation during special

experiences such as the school ball. This is in keeping with the notion of Disability Paradox where severe impairment is not necessarily associated with poor quality of life [34], provided the activity and participation restrictions can be avoided or moderated by appropriate contextual interventions [35]. For the adolescents in our study, intervening with the PWSD enabled more autonomous participation in physical, social and educational activities considered important to the adolescents and appeared associated with strong emotional wellbeing.

Mental health and social wellbeing are also intertwined. Developmentally, adolescents typically shift from a reliance on parents to a reliance on peers [36] with peer support needed for their development of social and emotional health [37,38]. Effective social interactions and social competence including within the school environment allows for the development of peer and adult relationships [39], and are among the most important determinants of mental wellbeing [40]. Whilst some adolescents were extremely self-conscious of standing, most reported that the capacity to stand at their choosing was useful in social settings with family and their peers at school, who in return responded positively. The role of the PWSD in supporting the boy's transition from primary to secondary school is an important topic for future research.

Respondents also reported key messages for the professionals involved in planning and supporting PWSD use. Training programs need to take into account the temperaments and needs of individuals and assist those who were inclined to be self-conscious in using the standing option, including early practice with the PWSD to allow for the boy, his parents and teachers to become comfortable and adept with its use. Individuals who received the PWSD after they became non-ambulatory appeared to derive less benefit, possibly because they were more self-conscious and less comfortable standing after a period of no upright activity. Some suggested that the PWSD should have been introduced for standing and fatigue management prior to loss of walking, as indicated by motor assessments [41]. Timely ongoing surveillance of the fit of the PWSD and need for modifications to accommodate growth was also essential to avoid the frustrations and possible loss of ability to stand after long breaks.

We achieved a 100% recruitment fraction and thematic saturation, suggesting that we had captured all relevant themes for this topic. Themes were derived from the adolescents, parent and teacher experiences and were consistent, the multiple perspectives providing a form of triangulation. We acknowledge that this qualitative study reports lived experiences and cannot quantify the perceived effects, and that our time frame of assessment was limited to (on average) eight months of PWSD use. Collection of qualitative data in response to PWSD use over a longer time frame is an important future study, as is the collection of data to quantify associations with physical and mental health, the transition from primary to secondary school, and quality of life. There are many issues regarding PWSD use in DMD where evidence is poor including how to determine eligibility and we are currently planning this research. Building the



evidence base for PWSD use is a critical task to enable appropriate accessibility to those with need.

Our qualitative data suggests that the PWSD supported the capacity of most of the adolescents to participate in a range of meaningful activities with greater autonomy, despite the progression of their disease, beyond what would be possible with existing therapeutic regimes involving standing frames. As is the case for their peers in the general population, it would seem reasonable and necessary for adolescents with DMD to experience the independence and participation in daily life afforded by having the option of choosing when and where to stand, after they are unable to walk.

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## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.nmd.2019.01.010.

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