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Case Report

Enhancing Function, Fun and Participation with Assistive Devices, Adaptive Positioning, and Augmented Mobility for Young Children with Infantile-Onset Spinal Muscular Atrophy: A Scoping Review and Illustrative Case Report

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Abstract: Recent advances in medical interventions have changed the prognosis for children with infantile-onset spinal muscular atrophy (SMA-1); however, little has been published regarding rehabilitation management. A rapid scoping review was conducted in November 2020 using Medline and CINAHL databases. Evidence supporting use of assistive devices and equipment to enhance participation, mobility, function, and posture in lying, sitting, and standing positions was sought. From 239 articles, only five studies (describing use of augmentative communication, manual and power mobility, supported standing and orthotic devices) met inclusion criteria. Results are presented alongside a case report of a 5-year-old boy (treated with Nusinersen since 7 months-of-age) who uses a variety of devices to enhance his activity and participation in family life. While reclined and tilted sitting positions as well as power mobility were previously considered for children with SMA-1, this child has progressed to supported upright standing, self-propelling a lightweight manual wheelchair indoors, communicating using multiple methods and taking steps in a dynamic mobility device. Power mobility was introduced in a switch-adapted cart at 11 months and he was independently exploring indoors and outside in his power wheelchair before 20 months. Research evidence is limited, but alongside the case report highlights the importance of a comprehensive and proactive approach to enhancing function, fun and participation with family and friends through adaptive equipment for children with significant and life-limiting disabilities.

Keywords: assistive technology; participation; durable medical equipment; wheelchair; power mobility; stander; supportive seating; postural management; supported standing; assistive devices; speech generating devices

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1. Introduction

Infantile-onset or type 1 spinal muscular atrophy (SMA-1) is the most common and severe form of this group of disorders characterized by progressive degeneration of the spinal motor neurons leading to progressive muscle weakness [1]. The first international consensus on diagnosis and management of SMA in 2007 [2] proposed dividing the phenotypes by age of presentation and highest level of function achieved and has been widely adopted. Non-sitters (SMA-1) present with symptoms before 6-months of age and never achieve independent sitting. Sitters (SMA-2) are diagnosed after 6 months-of-age and achieve independent sitting, but do not achieve independent walking. Walkers (SMA types 3 and 4) develop symptoms at older ages and have a less severe progression [2].

Children with SMA-1 were previously anticipated to have a mean survival age of less than 2 years; however, changes in medical management such as the use of non-invasive respiratory support (e.g., bi-level positive airway pressure—Bi-PAP) and nutritional support (e.g., enteral feeding) have changed the natural history of the disease with increased survival noted for cohorts born between 1994 and 2005 in comparison to historical cohorts [3].

More recently, new and emerging pharmaceutical interventions have demonstrated increased survival and improved functional abilities in children with SMA-1 leading to the emergence of a new phenotype referred to as 'treated' SMA-1 [4].

Given these changes, the Delphi consensus on diagnosis and management of SMA was updated in 2018 [1,5] with increased emphasis on proactive rehabilitation interventions [1,6]. For non-sitters (SMA-1), stretching (orthoses and standing frames), positioning (supportive seating and sleep positioning) and assistive technologies (power mobility, eye gaze systems) were recommended; however, only two references supporting these recommendations included children with SMA-1.

The International Classification of Function for children and youth (ICF-CY) [7], describes body structures and function (BSF), activity and participation as key components of health that are influenced by environmental and personal factors. Adaptive equipment and assistive devices are considered environmental modifications and enrichment aimed at enhancing participation (d465Moving around using equipment (https://apps.who.int/classifications/icfbrowser/Browse.aspx?code=d465&hsr=1, accessed on 30 November 2020) and e1201 Assistive products and technology for personal indoor and outdoor mobility and transportation (https://apps.who.int/classifications/icfbrowser/Browse.aspx?code=e1201&hsr=1, accessed on 30 November 2020)). Improving activity and participation outcomes is a primary goal in pediatric rehabilitation [8] and the ICF model has more recently been reframed for children with disabilities as Fitness, Function, Friends, Fun, Family and Future and referred to as the F-Words [9]. The emphasis is on increasing children's participation with family and friends in fun and meaningful activities that positively influence BSF, function and overall health.

For children with other significant physical disabilities, systematic reviews have been published regarding the benefits of adaptive positioning and augmented mobility to enhance activity and participation. These include reviews of night-time positioning [10], adaptive or supportive seating [11,12] and supported standing [13,14], as well as power mobility [15–17] and supported walkers or gait trainers [18]. However, while many studies describe the variable impacts of the new pharmaceutical interventions on physical function and development [19–22], little has been published regarding use of assistive devices to enhance activity and participation with children diagnosed with SMA-1. The purpose of this manuscript is to map and discuss the evidence supporting implementation of assistive devices to promote activity and participation for children diagnosed with SMA-1.

2. Methods

A scoping methodology [23] was chosen since evidence was known to be limited, of lower quality and the topic too broad to be appropriate for a systematic review. There are six stages to a scoping review [23]: (1) identifying a specific research question, (2) identifying relevant studies. In scoping reviews an iterative process is frequently undertaken to (3) select relevant studies, and (4) chart the data. The data are summarized (5) and results are reported, while stage (6) may include stakeholder consultation.

(1) The specific question guiding this review: What evidence exists to support the use of postural management equipment (e.g., lying, sitting or standing positioning devices or orthoses), mobility devices (e.g., manual wheelchairs, power mobility, supported walkers/gait trainers, tricycles) or other assistive technologies (e.g., speech-generating devices, computers, tablets) to promote activity and participation in children diagnosed with SMA-1?

(2) An electronic database search of Medline and CINAHL took place in November 2020. This was accompanied by hand-searching reference lists of systematic reviews of outcomes related to relevant interventions or devices in mixed populations and high-level expert consensus such as the recent Delphi consensus on diagnosis and management of SMA [1]. A Google Scholar search was also undertaken and population specific websites and resources such as https://curesma.org were searched. The protocol was registered on 19 November 2020 at Center for Open Science https://osf.io/tjbhf/?view_only=702074

3c014e4831becef511b53fe3a. Search terms included: spinal muscular atrophy AND child* AND rehabilitation OR wheelchair OR assistive device NOT neuromuscular agent NOT instrument development NOT reliability NOT validity.

(3) We sought to include primary source articles describing use of any device designed to enhance positioning, mobility or function in children (0–18 years) diagnosed with SMA-1. The first author conducted the electronic database search and downloaded results. The second author completed the Internet website and Google Scholar search. Both authors reviewed results independently and agreed articles meeting inclusion criteria. (4) Data were extracted by both authors independently and details presented agreed following discussion. (5) Data were summarized in table form and presentation of results discussed.

In this manuscript stage 6 takes the form of a case-report. The inspiration for this review is a male child (Tom—a self-identified pseudonym), born in 2015 and 5 years old at time of writing, who meets the description of 'treated' SMA-1. Tom's case report will be used to highlight the possibilities of the proactive use of various devices to enhance activity and participation, and an evidence-informed approach to intervention in line with the UN Convention on the Rights of the Child [24].

Case report details were drawn from medical records and chart review with consent of Tom's parents and his verbal assent. Some data (initial power mobility skill data, activity and participation change with power mobility use and use of devices in the home setting between 16 and 22 months of age) were collected as part of a previous research study that explored introduction of power mobility with young children [25–27]. Ethics approval for collection and use of this data for research purposes was approved by the University of British Columbia Children's and Women's Research Ethics Boards. Ethics approval for conduct and publication of this case report was waived by the same Boards.

Several measures, primarily from the Activity domain of the ICF, but also addressing the Participation domain were used to address our goal of describing change in activity and participation. See Appendix A for details of measurements' scales and measurement properties.

- Pediatric Evaluation of Disability Inventory (PEDI) caregiver assistance scale [28] was selected to describe Tom's level of independence in self-care and functional mobility;
- Power mobility or driving skill measures also primarily fall under the Activity domain and were used to describe power mobility skill development over time;
- Assessment of Learning Powered Mobility (ALP) measures the power mobility skill process and is suitable for all ages, motor and cognitive abilities [29];
- Power Mobility Program (PMP) [30] measures abilities to complete a list of 34 power mobility tasks ranging from basic mobility to integrated skills in structured and unstructured environments;
- Power Mobility Training Tool (PMTT) [31] was developed to guide power mobility training for children at the very beginning stages of learning;
- One included measure is considered to address primarily the domain of Participation;
- Wheelchair Outcome Measure for Young People (WhOM-YP) [32] is an individualized, client-centered measure developed for children or young people who use wheeled mobility.

3. Results

The electronic database search was completed on 15 November 2020 identifying 237 results and a further 11 articles were identified from the hand-search. Once duplicates were removed, 223 articles remained. Most references were eliminated on title and abstract review. Following full-text review of 17 articles only five primary source articles were found to include children diagnosed with SMA-1 [33–37]. See Figure 1 for PRISMA flow chart outlining each phase of the process. Twelve articles were eliminated as they did either did not include or did not explicitly report data from individuals with SMA-1 [38–49].

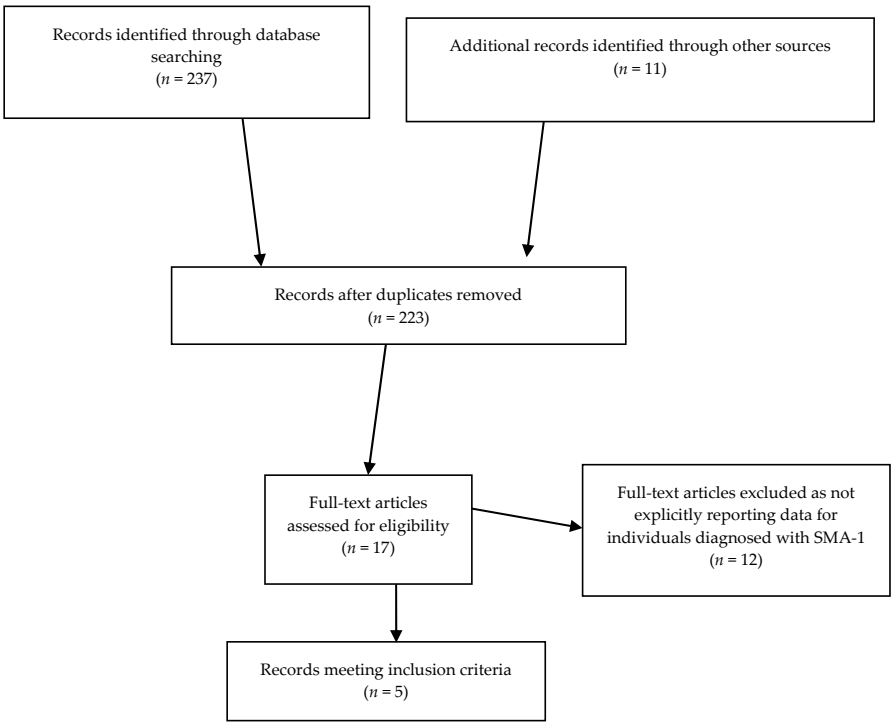

Figure 1. PRISMA flow diagram of the search results.

Five articles met inclusion criteria and are included in Table 1; one described power mobility skill development in a case-series of six children including only one child diagnosed as SMA-1 [34]; another described results from an internet-based survey of 32 parents of children with SMA-1 regarding use of speech generating devices and other augmentative communication supports [33]; a third study reported results of a retrospective cohort describing orthotic, positioning and mobility equipment use including 14 children with SMA-1 [35]; a qualitative study explored parents experiences with wheelchair provision and included parents of one child diagnosed as SMA-1 and another described as SMA-1-2 [36]; while a fifth study included data on stander use in 152 children with SMA-1 [37]. Three studies did not include individuals who could be described as 'treated' SMA-1 based on study date [34,35] or explicit statement [37]. For remaining studies, this factor was not reported [33,36].

Table 1. Studies including children diagnosed with SMA-1.

Citation	Design	Participants	*N* SMA-1	Devices	Results
Ball et al., 2019 [33]	Internet-based survey	Parents of children diagnosed SMA-1	32 parents of individuals aged 6 months–30 years	Speech Generating Devices (SGD)	12/32 used SGDs 13 used eye-gaze, 3 direct finger access and 4 switch access
Dunaway et al., 2013 [34]	Descriptive case-series	6 children (5 SMA, 1 CMD)	1 aged 34 months at PWC delivery	Power mobility	SMA-1 achieved basic skills in 260 days, directional control and doorway/corridor navigation in 458 days
Fujak et al., 2011 [35]	Descriptive retrospective cohort	194 children and adults with SMA types 1–4, recruited from one specialized clinic	14 aged 1.7–36.9 years	Orthotics Power mobility Manual mobility Standing frames	Use by SMA-1 (mean age at prescription ± SD) AFO—1 (4.7 years) TLSO—10 (6.0 years ± 4.9) PWC—6 (7.0 years: ± 3.9) Stroller—8 (ND) Stander—2 (ND)

Table 1. *Cont.*

Citation	Design	Participants	*N* SMA-1	Devices	Results
Paguinto et al., 2019 [36]	Qualitative	Parents of 14 children with neuromuscular disorders aged 1–17 years	Parents of two children: SMA-1 aged 1 year and SMA1-2 aged 3.5 years	Manual or Power mobility	PWC recommendation offered hope, increased independence and participation. Child with SMA-1-2 prescribed PWC at 14 months. Child with SMA-1 provided early power mobility trial between 9 and 12 months.
Townsend et al., 2020 [37]	Descriptive retrospective multi-center cohort	397 children SMA-1 and SMA-2 enrolled in natural history study 2004–2015	152 aged 0–4.5 years at enrolment who survived >9 months	Standing frames	20 SMA-1 stood regularly ≥3 times a week Stander use from 6 months of age

AFO: ankle foot orthoses; CMD: congenital muscular dystrophy; ND: not disclosed; PWC: power wheelchair SD: standard deviation; SMA: spinal muscular atrophy; TLSO: thoraco-lumbar-sacral orthosis.

Studies included provide descriptive rather than experimental evidence and a formal quality rating was not completed. However, the two cohort studies [35,37], included significant numbers representing our target population, and the study exploring the use of standing [37] in a natural history cohort included participants from multiple centers in the US, as well as a European and a Canadian center, suggesting results may be widely applicable. The survey of parents regarding communication [33] recruited from a large international register and is the first to explore this topic. Although 32 may seem a small number for a survey, all were parents of individuals with SMA-1 and participants from Mexico and Malaysia participated in addition to those from various regions of the USA widening applicability of results.

Although Dunaway's study [34] is small and descriptive, it was the first study to describe power mobility skill progression in a group of children with SMA and highlights the need to accommodate seating and access needs for children with SMA-1. The qualitative study [36] reported results from only one region in Australia, but demonstrated rigor in its conduct. Findings suggest a difference in response between parents of children with the more severe forms of SMA to wheelchair prescription in comparison with parents of children who were ambulatory where the wheelchair was seen as symbolic of loss of function. It is encouraging that early power mobility experience was introduced before 12 months and power wheelchair prescription considered as young as 14 months. This contrasts with the case series [34] where the youngest prescription was at 16 months and the youngest power wheelchair delivery at 24 months.

Results are reported according to device category: postural management (night-time positioning, supported seating, supported standing, orthotic management), mobility (manual mobility, power mobility, supported walking/gait trainer, tricycle) and other assistive technologies (arm assists, speech-generating devices). These categories were identified either from scoping review results or case report details. Where research evidence was identified for SMA-1 (meeting inclusion criteria and reported within Table 1) this is reported first, and supplemented where necessary with relevant studies including SMA-2. Following the research evidence, results within each category are illustrated with details from Tom's case report.

3.1. Case Introduction and History

Tom is the first child born to parents with no family history of SMA. His mother is at home with him full-time, while his father works outside the home. Tom's family is supported by their large extended family, close friends, and church community. Tom was diagnosed at age 5 months after he stopped kicking and lost head control at age 3 months.

He was enrolled in a double-blind sham controlled Nusinersen drug trial at age 7 months. He received Physical Therapy (PT), Occupational Therapy (OT) and infant development consultation through his local Child Development Centre and was referred to the provincial program for positioning, mobility and assistive technology consultation.

A month following his first doses of Nusinersen Tom started moving his arms again, causing family to suspect that he was in the treatment arm of the study. He transferred to the open-label extension study from 12 months and has continued to receive Nusinersen at 4-month intervals since that time. Tom steadily gained head control and strength and established momentary control of independent sitting at 36 months. He progressed to being able to maintain cross-legged sitting for 15–20 min by age 5 years.

3.2. Current Functional Description

At time of writing, Tom's upper limb strength and control is greater than lower limbs and he is able to lift his hand to his face in sitting or supported standing. His hips have been supero-laterally dislocated since infancy, but continue to be pain free and have adequate range-of-motion (ROM) (45 degrees abduction bilaterally with hamstring tightness and mild hip flexion contractures). Tom has a 33-degree left thoracolumbar scoliosis that has been stable for the last two years.

According to the PEDI [28] caregiver assistance scale, he requires Maximal assistance in activities of daily living but is able to provide some meaningful assistance in grooming and upper body dressing and with bathing. Tom uses a urinal during the day and has a supportive shower-commode chair for bathing and toileting. He requires Total assistance for transfers, Moderate assistance indoors in a manual wheelchair and age-appropriate Supervision outdoors in a power wheelchair.

Tom continues to use non-invasive respiratory support (Bi-PAP) for sleep and naps and his nutritional needs are met via gastrostomy tube. He requires suctioning 5–8 times a day. He is a delightful, social little boy who currently communicates primarily with speech, although speech is dysarthric and more challenging for unfamiliar communication partners. At age 5 years 6 months, he started kindergarten via a home-school program. See Table 2 for details of Tom's equipment use timeline from 9–60 months.

3.3. Postural Management

3.3.1. Lying or Night-Time Positioning

Sleep positioning is specifically recommended in the 2018 consensus [1]; however, no studies or reviews specific to sleep positioning support for any type of SMA were identified in our search. One included study [35] recommends the use of lying supports to prevent frog-leg positions. Sleep positioning systems are recommended to maintain body symmetry and to enhance sleep [10]. For children with SMA-1 facilitating respiratory function is also of primary importance. As an infant, Tom slept on an air-filled mattress section, but by 43 months alignment of hips and spine was improved on a firmer memory foam mattress.

At time of writing, Tom continues to sleep in side-lying or three-quarter prone with a small roll under the upper leg to ensure neutral to abducted hip positioning. Tom can roll from supine to either side and pivot 360 degrees in supine on a firm floor mat. He cannot lift his head in supine nor clear his arms in prone. On the soft surface of his mattress, Tom is dependent on regular turning by parents or nursing staff and is positioned on either side equally to maintain symmetry and comfort. He sleeps on a high-level toddler bed that is at a functional height for caregivers.

Table 2. Equipment timeline.

Months	9–12	13–15	16–18	19–21	22–24	25–27	28–30	31–33	34–36	37–39	40–42	43–45	46–48	49–51	52–54	55–57	58–60
Night-time positioning	Air-filled mattress section. Positioned 3/4 prone with roll under top leg.																
											Memory foam mattress. Side-lying or 3/4 prone with roll under top leg						
Seating	Reclined foam seat																
		Custom seating with curved trunk laterals, medial and lateral pelvic and thigh supports and custom arm supports.															
			Wooden tilt floor base														
Standing				Floor level supine stander. Stands in 10 degrees tilt 1 h twice daily									High level transfer supine stander. Stands fully upright 1 h twice daily				
Orthotics			Solid AFO's used 2 h daily in stander initially—progressed to using 10 h a day in wheelchairs, standing and other positions. Not in walker as weight limits leg movement.														
						TLSO used for floor sitting, in floor level mobility device, manual wheelchair and in tricycle											
	Custom wrist splints used at night-time to maintain ROM. Some daytime use to enhance function																
Manual mobility		Infant stroller															
					Special needs stroller with full tilt and recline												
									Home-made self-propelled floor level manual mobility device								
												Microlite wheelchair with soft backrest and cushion.			Lightweight rigid wheelchair with cushion and curved backrest.		
Power mobility	Switch-adapted cart																
			MPW with tilt and sensitive finger joystick		Pediatric power wheelchair with tilt and seat elevate. Accessed with micro extremity joystick						Pediatric power wheelchair with tilt and seat elevate. Accessed with mini proportional joystick						
Dynamic mobility device													Hands-free device trial			Using 30 min daily	
Tricycle											Special needs tricycle with high backrest, pelvic and chest harness and shoe holders.						
Other assistive technologies	Custom suspension arm support																
					Tablet-based augmentative communication software												

AFO: Ankle foot orthoses; MPW: miniature power wheelchair; TLSO: thoraco-lumbar-sacral orthosis.

3.3.2. Seating

No studies specific to positioning with SMA-1 were identified in the search, although a study of 12 individuals (mean 16.4; range 7–24 years) diagnosed with SMA-2 reported improved respiratory function when positioned in individually prescribed and fitted supportive seating in comparison to standard wheelchair seating [44]. The Delphi consensus [1] recommends tilted or reclined supportive sitting to enhance function and custom or molded seating systems as necessary. Fujak and colleagues [35] also note that some individuals benefit from individually fitted systems.

As an infant Tom spent most of his awake time in supine due to respiratory difficulties and required frequent suctioning. A foam wedge/chair was introduced for daytime positioning at a 140-degree (reclined) seat-to-back angle and an early positioning system was introduced with small rolls to position him in side-lying. Tom had very limited abilities to move his arms against gravity and these positions allowed increased opportunities to reach, grasp and explore toys.

By 12 months Tom could tolerate 30–60 min a day in the foam recliner chair. Specifications were taken for a custom seating system with pelvic and trunk laterals and head support. The custom seating system with tilt-in-space floor base (see Figure 2 bottom left) were provided at 14 months and Tom was able to tolerate up to 2 h at a 100-degree seat-to-back angle and tilted to 25–30 degrees. At 16 months, this seating was used at a similar tilt angle in his first power wheelchair. Tom could not tolerate upright sitting in his custom seat until age 2 years due to head control and respiratory difficulties.

Figure 2. Assistive device examples, photos published with consent of the parents. Clockwise from top left: Side-lying with support, using overhead arm support; Lightweight manual wheelchair; Dynamic mobility device; Special-needs tricycle with supports; Pediatric power wheelchair with custom seating and very sensitive finger-controlled joystick on front of right arm support; Custom seating system on tilt-in-space floor base.

At time of writing, Tom's rib cage is mildly asymmetrical and his seating system now includes custom molded trunk laterals to provide full support. Medial and lateral thigh supports are also used to help maintain a symmetrical sitting position, to limit progression of deformities while maximizing function and participation.

3.3.3. Standing

A report was recently published describing stander use in a cohort of 397 children with SMA-1 or type 2 who had not been treated with the newer drug interventions [37]. Consistent stander use was 68% in SMA-2 in comparison to 13% for type 1 and reported for children from as young as 6 months in both groups. Supported upright standing in a supine stander was noted to be possible for a child (SMA-1) with extreme muscle weakness who required multi-joint lower extremity bracing and significant trunk and head support. Ability to turn the head when supported in the standing frame was associated with increased tolerance of supported standing and recommended to provide more opportunity for participation and inclusion in activities than from dependent supine positions.

Similarly, a retrospective review of 194 children and adults with SMA [35] reported that 2/14 with SMA-1 used standing frames in comparison to 22/133 with SMA-2. Sixteen individuals with SMA-2 used a powered wheelchair standing feature. The authors recommend individuals with non-ambulatory SMA use supported standing at least 2 h daily and incorporating this feature into the power wheelchair where possible to facilitate maximum weight-bearing.

Tom's supportive supine stander was delivered by age 15 months, but he was unable to use it until he had suitable ankle foot orthoses (AFO) at 17 months. By 18 months, he was tolerating up to 1 h at a time in the standing frame for play-time in a small amount (10-degrees) of recline. The large tray and upright position allowed Tom to manipulate and play with lightweight toys.

At time of writing Tom continues to use his stander up to 2 h daily, one hour in the morning and one in the evening. He is transferred in supine and then tilted to a fully upright position. He uses his stander to participate in school-work, art and board games with family and friends. Family would like to incorporate the standing feature into his power wheelchair in future when a model suitable to his size is available and fundable.

3.3.4. Orthotics

In the one study reporting orthotic use [35], only one individual with SMA-1 (aged 4.7 years) used lower limb orthotics in comparison to 19/133 with SMA-2 aged 1–10.2 years. The authors recommended earlier provision of AFO's to children with non-ambulatory SMA while foot position is correctible. Townsend et al. [37] recommended use of AFO's and knee ankle foot orthoses (KAFO) to ensure joints are aligned and prevent valgus forces at the knees in supported standing.

Fujak and colleagues [35] also reported that ten children with SMA-1 ranging from 1.3 to 15.3 years used TLSO's in comparison to 90 diagnosed as SMA-2. Provision of a TLSO was recommended when scoliosis reached 20 degrees to help delay progression of the curve until operative spinal stabilization was a possibility. TLSO's were noted to improve stability in sitting and increase upper limb function. The authors stressed that attention should be paid to ensuring TLSO's do not interfere with function of intercostal and respiratory muscles or diaphragm, should permit easy access to the gastrostomy site and should be regularly reviewed, adjusted and replaced during periods of rapid growth. Out of 194, only two individuals used night resting splints to maintain upper limb or wrist positioning. Participants indicated that upper limb splints limited their abilities to turn themselves at night reducing independence.

Tom's ankle foot orthoses (AFO) were introduced at 17 months and used for standing up to 2 h daily by 22 months. As time sitting in wheelchairs increased, Tom increased time wearing AFO's up to 10 h a day. They were removed for freedom of movement on the floor, when in his walker and at night.

At 27 months, a TLSO was introduced to help reduce progression of his scoliosis convex to the left and to provide stability in sitting out of his custom seating. He has continued to use this for independent floor sitting, positioning in his manual wheelchair and tricycle and his scoliosis has remained stable for more than 2 years.

At time of writing, Tom is unable to turn on his mattress at night and is fully dependent for transfers. He is at risk for losing wrist extension range due to decreased anti-gravity control and has a tendency to ulnar deviation. Tom dislikes wearing wrist splints during the day but tolerates wearing one wrist splint positioning his wrist in a neutral position each night, alternating right and left to help maintain ROM while not negatively impacting function, comfort or ability to sleep.

3.4. Mobility

3.4.1. Manual Mobility

A retrospective cohort study [35] that met inclusion criteria noted that few individuals with SMA were able to use manual wheelchairs due to limited shoulder strength and only 19 individuals (SMA-2) were prescribed these at a mean age of 4.2 years. Younger children tended to use strollers with eight children with SMA-1 and 4 with SMA-2 using either standard or special-needs models. The authors caution against extended use of strollers with this population as they do not enhance independence. They recommend an independent mobility device either self-propelled manual or power mobility by three years-of-age at the latest. The 2018 consensus [1] recommends strollers with tilt and ability to lie flat and power mobility with supportive seating for non-sitters.

As an infant, Tom used an infant stroller with full recline. By 24 months, a larger special-needs stroller with tilt and full-recline was ordered as Tom's back-up/dependent manual wheelchair option. This was used for accessing family and friend's homes that were not wheelchair accessible, for times when Tom needed to be in a more reclined position than his power wheelchair allowed, and as a changing surface when out in the community.

Around 30 months, Tom began using a floor level self-propelled manual mobility device. Tom's Grandpa created this using a foam child floor seat mounted to a wooden base with 12" wheels. Tom used his TLSO to provide trunk support and began to self-propel within the house. WhOM-YP satisfaction with mobility indoors was scored as 5/10 as he was unable to cross thresholds between rooms.

At 47 months Tom borrowed a Microlite pediatric wheelchair with a goal of being able to move from room to room independently. The importance of this goal was 8/10 but by the end of six-months satisfaction only increased to 6/10 as Tom still required assistance from an adult. Funding was requested for a lightweight rigid manual wheelchair that would meet funder required growth potential of 5 years. He received it at 56 months (see Figure 2 top center) and a new goal was set for him to be able to independently open kitchen doors to access items as a favorite game was closing cabinet doors with the loaned manual wheelchair when his parents were preparing meals. WhOM-YP satisfaction was rated by parent proxy as 9/10 after 1 year. See Figure 3 for WhOM-YP scores in all mobility devices.

3.4.2. Power Mobility

The only intervention study meeting inclusion criteria was a small case-series [34] involving six children, one of which was diagnosed with SMA-1. This child had seating and access difficulties that were not accommodated adequately when his power wheelchair was set up at 34 months-of-age. These challenges may have contributed to the length of time (15 months) taken to achieve competent control and contrasts with other case reports where children diagnosed with SMA-2 demonstrated proficient use within 6 weeks after starting at 20 [38] and 22 months [50]. For non-sitters, the 2018 consensus [1] recommends use of power mobility with tilt/recline, the retrospective cohort study [35] recommends power mobility introduction before age 3, while case-series authors [34] recommend introduction between 7 and 24 months as close to the typical onset of independent mobility as possible.

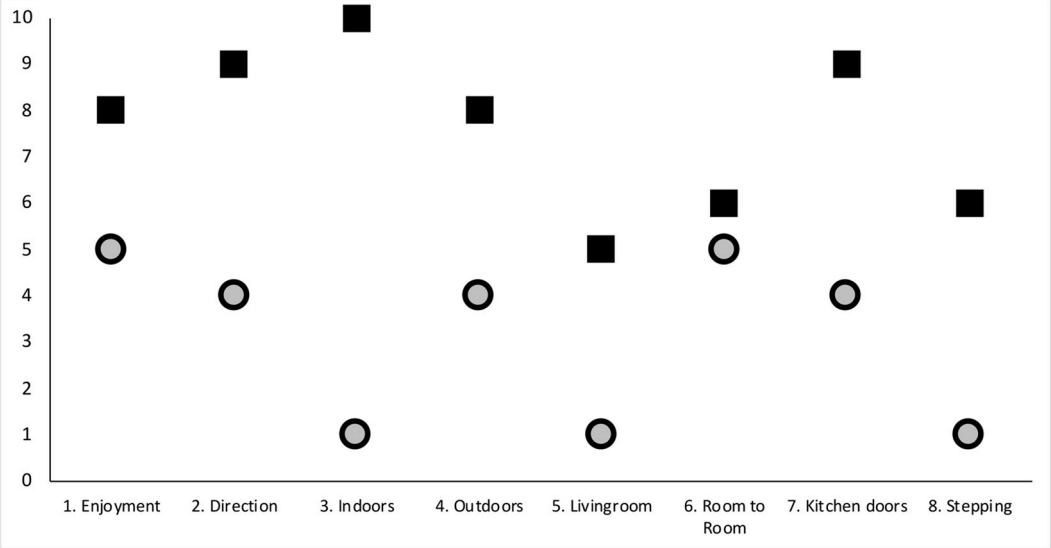

Figure 3. WhOM-YP Goal achievement. Circle: goal satisfaction when device was introduced; Square: goal satisfaction after an average of 6-months experience. 1. Enjoying movement. 2. Understanding direction and 3. Ability to move indoors: goals set for 6-month loan of Tiger cub. 4. Independent mobility outdoors: goal for power wheelchair in comparison to Tiger Cub. 5. Ability to move independently in the home: floor-level manual mobility device. 6. Ability to move and play indoors, moving between different rooms and crossing thresholds: microlite wheelchair loan. 7. Ability to maneuver in kitchen, open and close lower cabinet doors during play: lightweight rigid manual wheelchair. 8. Satisfaction with enjoying upright stepping and moving in the dynamic mobility device.

At 11 months, Tom was introduced to power mobility in a switch-adapted cart. He was positioned at a 140-degree reclined sitting position in his foam chair. His right arm was supported with an overhead arm suspension system and very sensitive mechanical switches were positioned on a lightweight tray. Tom initially enjoyed using the switches to spin in circles in the basement, and once the weather improved, to drive forward down his driveway.

His power mobility skills steadily progressed in the cart from being unaware that the switch made the cart move (ALP phase 1) to having a solid understanding of cause-effect with a single sensitive switch (ALP phase 3). At 14 months, Tom was enrolled in an early power mobility study [25,51] and had his first experience using a joystick in a power wheelchair. He was only able to tolerate sitting at a 30-degree tilted angle with full head support and required a very sensitive joystick. In this initial experience, he showed emergent understanding that he was controlling the power chair with the joystick (ALP phase 2).

At 16 months Tom participated in the second phase of the same research study [26,27]. His custom seating system was set up in a miniature pediatric power wheelchair (Invacare Tiger cub-previously available from Invacare.com, now discontinued) for a 6-month loan (see Figure 2 bottom center). A very sensitive mini joystick was positioned directly in front of his right arm trough. Tom accessed the joystick using index finger movement. Tom's power mobility skill quickly progressed and at 22 months, he achieved proficient use (ALP phase 7). See Table 3 for detail on power mobility skill progress from 11 to 22 months.

Table 3. Power mobility skill progress.

	11 Months	14 Months	16 Months	17 Months	20 Months	22 Months
ALP	1	2	3	6	7	7
PMTT non-motor	5/16	9/16	12/16	16/16	16/16	16/16
PMTT motor	4/12	5/12	6/12	12/12	12/12	12/12
PMTT driving	3/20	8/20	10/20	20/20	20/20	20/20
PMP beginning	4/30	8/30	9/30	25/30	25/30	25/30
PMP directional	2/35	4/35	5/35	30/35	30/35	30/35
PMP speed control	0/20	0/20	0/20	0/20	10/20	10/20
PMP functional	0/15	0/15	0/15	0/15	15/15	15/15
PMP ramps	0/30	0/30	0/30	0/30	5/30	18/30
PMP sidewalks	0/10	0/10	0/10	0/10	0/10	0/10
PMP community	0/30	0/30	0/30	0/30	0/30	12/30

ALP: Assessment of Learning Powered mobility use; PMP: power mobility program; PMTT: power mobility training tool.

Three goals were set for the 6-month loan using the WhOM-YP: Enjoying movement; Understanding direction; and Moving around in the house. Importance of these goals ranged from 8–10/10 and Satisfaction was rated as 5 for enjoyment, 4 for direction and 1 for independently moving in the home at loan-start. By loan-end satisfaction increased to 8 for enjoyment (still could not reach toys), 9 for understanding directions and 10 for ability to move around in the home (see Figure 3).

Immediately following completion of the research study, Tom was set up with a pediatric mid-wheel drive power wheelchair with power tilt and seat elevate through the provincial loan program. This chair had small sensitive proportional joystick, although less sensitive than the one used in the research study. However, Tom immediately demonstrated ability to drive safely in his home, through doorways and in and out of small spaces in this larger wheelchair.

Tom's parents set a new goal with the WhOM-YP of being able to move around independently outdoors. They had found some limitations over soft or more uneven ground with the Tiger cub and rated satisfaction with its performance as 4/10. Following 4 months experience in the new power wheelchair at age 26 months, they rated satisfaction with performance outdoors in the new power wheelchair as 8/10. This rating was not higher as there were still some limitations in terrain capability for this wheelchair's 10" wheels.

3.4.3. Gait Trainers and Dynamic Mobility Devices

No studies were identified describing use of gait trainers or supportive walkers propelled by stepping in children with SMA-1, although Fujak and colleagues [35] report the use of long-leg splints and a swivel walker with one individual with SMA-1 and five with SMA-2. When Tom was 45-months old, we trialed an early intervention sized hands-free dynamic mobility device (similar to a gait trainer) and he was able to take some steps. We hoped it would increase his ability to play and explore in an upright position. Funding for the larger child-sized version that would accommodate future growth was requested and received by 58 months (see Figure 2 top right).

Although, at time of writing, Tom still requires some adult assistance to move the walker forward, he can take small steps with his feet and spends 30 min daily engaged in games like hockey or skittles and imaginative play like pirate treasure hunts. A goal was set with the WhOM-YP (see Figure 3) to be able to take steps and explore indoors.

3.4.4. Tricycle

We did not identify any studies discussing use of tricycles with children diagnosed with SMA-1. A study including five school-aged children with SMA-2 found that arm cycling was well tolerated with increased distance and speed over time and non-significant motor skill changes on standardized testing [52]. Tom's family wished to explore use of a

tricycle as an age-appropriate recreational activity and at 43 months funding was received for a fixed-gear special-needs tricycle and supportive seating (see Figure 2 bottom right).

Tom currently enjoys using this outside on his quiet street as a different means of mobility to his power wheelchair. He loves interacting with the other children and pets in the neighborhood. Adults help Tom move the pedals by pushing with the rear-steering caregiver handle and this assisted-active range of motion is part of enhancing his fitness and maintaining BSF within a meaningful and age-appropriate activity.

3.5. Assistive Technologies

3.5.1. Augmentative and Alternative Communication

One of only five articles meeting inclusion criteria explored use of augmentative and alternative communication (AAC) and speech generating devices (SGD) [33]. An internet-based survey was sent to 3000 parents included in an international SMA register and from 35 respondents 32 parents of children diagnosed with SMA-1 were included. Participants children ranged in age from 1–30 years (mean 7.75 years; median 5.4 years). Results confirmed that receptive language and eye movements presented no difficulties, but severe difficulties with expressive language and finger control were reported. SGDs were used to gain attention, request, comment, reject, provide quick or detailed messages and to protest, however only 12/32 had their own device and 5 used low-tech symbol-based communication boards. Twenty-two children used gestures, 19 used speech while three used a combination of methods. Barriers to acquiring an appropriate SGD included funding challenges and lack of clinician support, but 9 parents reported that the SGD improved their child's quality of life.

Tom has very low tone and a myopathic facial appearance with minimal tongue movement. His speech is severely dysarthric and he used augmentative and alternative communication strategies from a young age. Between ages two and three years, more formal dynamic symbol-based communication vocabulary was introduced on tablet-based systems. Initially, a more specialized system with a keyguard was required, but as Tom gained strength and accuracy in targeting with his finger, he transitioned to a commonly used tablet.

At time of writing, Tom uses the tablet for games only and communicates primarily through speech. He hopes to attend his local school next year and may require the tablet for clarification with unfamiliar communication partners or for delivery of longer verbal presentations due to fatigue. Tom can hold a pencil and is learning to print however technology will likely be used to support written communication as academic demands increase in future.

3.5.2. Arm Assists

Reports of development and use of mobile arm assists were found for use with individuals with a variety of neuromuscular conditions including SMA-2 [41,53] These reported the device improved reach and ability to pick up objects and complete functional tasks such as self-feeding, participation in school and recreation such as playing chess, music or operating the TV remote.

A custom overhead sling arm-assist was created by Tom's OT using a commercial drawer slide mounted on a freely rotating post (see Figure 2 top left). This was introduced at 11 months to assist with play skills and handling of lightweight toys. It also facilitated Tom's access to a commercial tablet for cause-effect and educational games. These activities were used to help develop his targeting and choice-making abilities in preparation for AAC software. The arm-assist was used until age 3 years when Tom developed sufficient anti-gravity arm movement for it to be no longer required.

4. Discussion

This scoping review identified only five studies in peer-reviewed literature describing use of assistive devices to promote activity and participation in children diagnosed as

SMA-1. The limited results were enhanced and illustrated by a case report that described how the use of various types of adaptive equipment (including postural support in lying, sitting and standing positions, adaptive fitness and mobility equipment, and devices to assist with communication and fine motor control) enhanced the ability of one child and his family to participate in natural and age-appropriate routines.

Three of the five studies that met inclusion criteria discussed power mobility. This is not surprising as power mobility has been consistently recommended as the only means of providing efficient independent mobility for this population [1,2,35]. This case report documents Tom's power mobility skill progress starting at 11 months in a switch-adapted cart and then in a power wheelchair from 16 months. All basic wheelchair skills were achieved within 5 weeks of experience using a joystick and he was competent within the home environment as young as 20 months-of-age. However, without appropriately supportive seating (including full arm support), tilt-in-space and very sensitive alternate access options (switches first and then joystick) this success would not have been achieved.

The ALP [29] and PMP [30] were the most appropriate power mobility skill measures for documenting Tom's progress and he is considered a functional learner [54]. Tom's progress (Table 3), illustrates some of the issues with the content of the PMP. Due to physical weakness, he was unable to turn the chair on or off and lost 5 points in the Beginning skills section. Although Tom could maneuver in and out of small spaces, as a 22-month-old he either did not understand, or was unwilling to back-up on command for at least 2 feet and lost points on the directional control section. At 17 months, Tom was still driving outdoors only, and opportunity to test stopping at a line or a door did not present itself (speed control section). The ramp and sidewalks as described in the test were also not available in Tom's environment. The PMP was useful for guiding his training, but dividing total score by 34 as suggested by the authors was problematic. Tom's total of 120/34 gives a summary score of 3.5. This score would suggest that he requires stand-by assistance for most tasks and would be an underestimate of his abilities. These results support use of raw scores as described in this paper and by other researchers [55,56].

The cost-effectiveness of life-course altering interventions for SMA is being debated in the literature [57], however very little has been published regarding the need for assistive devices to augment abilities and enhance participation in age-appropriate and meaningful activities and life-experiences. Outcomes for these new interventions are frequently framed as increased life expectancy, or length-of-time to invasive respiratory support while quality of life and participation are rarely discussed.

As discussed earlier, the ICF has more recently been reframed as the F-Words for childhood disability—Fitness, Function, Friends, Family, Fun and Future (see Figure 4). The remainder of the discussion is structured according to the implications and relevance of this model to Tom's case study and the wider literature regarding postural and mobility management for children with significant physical disabilities.

4.1. Fitness

Until recently, orthopedic interventions such as hip and spine surgery were previously rarely recommended for children with SMA-1 due to limited life-expectancy and a more palliative approach [1]. Hip subluxation has been shown to occur most rapidly in SMA-1 in comparison to other SMA types [58] and screening as well as pro-active orthopedic management is now recommended for children with SMA-1. For other populations at risk for hip subluxation, hip surveillance is recommended to begin between 12–24 months and continue at least yearly [59]. Children and adults who are unable to change position at night, tend to remain in one position that can negatively influence body shape, lead to windswept hip deformities and scoliosis [60]. In addition to maintaining BSF, postural management in lying, sitting and standing positions enhances activity and participation. By preventing contractures and deformities children can more easily be positioned for play and other age-appropriate activities.

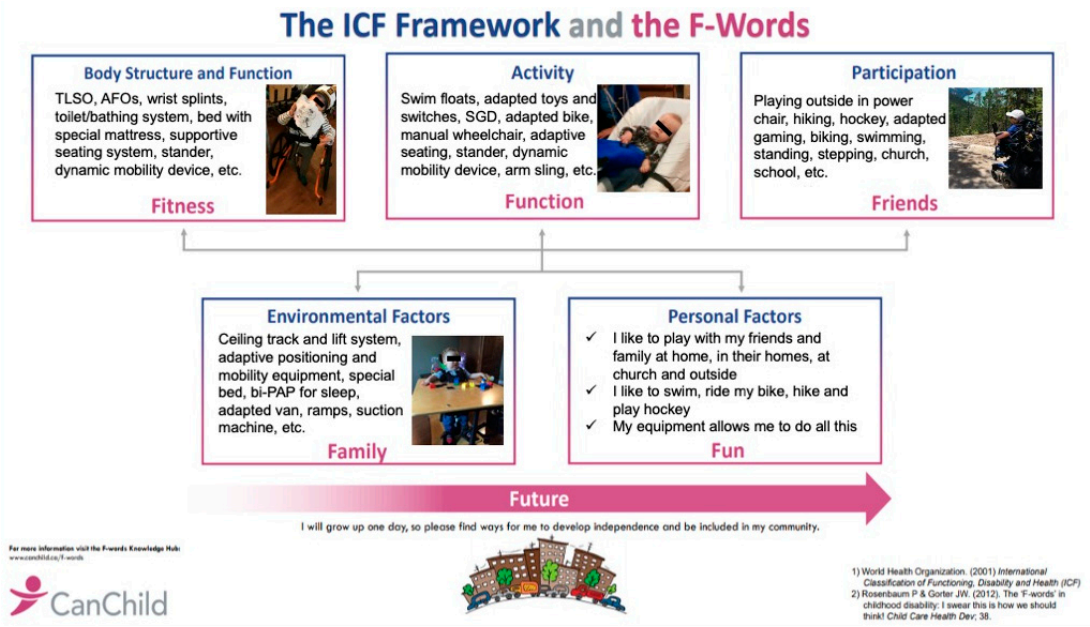

Figure 4. F-Words [9] for one child with SMA-1 adapted with permission from the authors. AFO: ankle foot orthosis; Bi-PAP: bi-level positive airway pressure; SGD: speech generating device; TLSO; Thoraco-lumbar sacral orthosis.

Standers are commonly prescribed to increase weight-bearing and upright positioning in order to positively influence bone mineral density, range of motion, hip stability and bowel function [14]. These outcomes have not been studied in children with SMA, but the natural history cohort [37] suggests that standers are well tolerated and this case report provides support as Tom continues to tolerate standing around 2 h daily. In his case, the upright position was also found to be beneficial for his respiratory function and Tom maximizes his time in upright and weight-bearing positions by also using his dynamic mobility device around 30 min daily.

4.2. Function

Although for children with neuromuscular conditions, TLSOs are more commonly recommended to slow progression of spinal curves until such time as spinal instrumentation surgery is viable [1], they have also been suggested as a means of improving sitting balance and arm function [35]. In a study of 251 children with cerebral palsy who used TLSO's, most (96%) used them with the goal of improving stability and positioning while 51% used them to promote head control and 38% to improve hand control [61]. In Tom's case, the TLSO allowed him to use mobility equipment other than his power wheelchair and it enhanced function in his manual wheelchair, tricycle and dynamic mobility device. In the first year, Tom had such limited anti-gravity control that he was positioned in side-lying with supports and an overhead arm suspension sling was used to allow access to cause-effect apps and lightweight foam toys or balloons. The standing frame was also found to facilitate play and upper limb use in comparison to a seated position. Tom could slide toys around on the large tray surface before he was strong enough to lift objects against gravity.

4.3. Friends

Participation is a primary goal for therapists working in pediatric rehabilitation [8]. Power mobility has been demonstrated to enhance participation, engagement in meaningful life experiences and social relationships for children with mobility limitations [17]. In Tom's case, when we introduced early power mobility experiences at 11 months, his prognosis was unclear, but we sought to provide independent mobility experiences at a developmentally appropriate age as recommended by Dunaway [34]. Tom's parents' recollections of their feelings at the time when power mobility was first recommended resonate

with the parents in Paguinto's qualitative study [36] who found the recommendation of power mobility to be positive following the dark period of loss and helplessness around time of diagnosis. Tom's power wheelchair allows him to be independent outdoors and in the community, and will hopefully facilitate integration at school in future.

4.4. Family

Environmental factors—including social, attitudinal and physical—can have a huge impact on young children's use of power mobility [17] as well as other assistive technologies [62], and in Tom's case, his family has been a huge support in helping him to maximize his potential. They have been very proactive in acquiring equipment and modifications, and Tom's Dad and Grandpa have modified his home, adding ramps, modifying equipment such as his mobile arm support, and even building his first floor-based manual mobility device. Applications to charitable organizations for funding support take time and dedication. Tom's parents have been supported in this process by a number of different therapists and consultants to facilitate his access to the right equipment, at the right time in his development.

4.5. Fun

Activities and interventions should be appropriate to the age of the child, their interests, support the family and be culturally sensitive. Service delivery should occur in natural environments and focus on including the child in the family's existing natural routines or routines in which they would like to engage. In other populations, practicing in context as a whole activity that is part of a naturally occurring routine has been shown to be more effective than practicing isolated exercises or pieces of a routine [63]. Participation in daily-life is enhanced by use of adaptive equipment to modify the environment. BSF (Fitness) goals can often be addressed within routines that are fun and involve family and/or friends.

Another way to look at this approach is to pull back the lens and look not just at the dance floor, but climb up to the balcony and take a full view of the whole eco-system surrounding the child. Focusing on BSF is only the first step, keep in mind that the goal of improving BSF is to enhance activity and participation. All interventions should be child-directed, and, for children who are mostly at home, caregiver delivered. By keeping children active and engaged, goals across all the ICF domains (and all the F-Words [9]) can be achieved.

4.6. Future

The aim of this paper is to raise expectations for children with life-limiting conditions, through use of equipment to augment abilities and facilitate participation in age-appropriate and meaningful activities and life experiences. For a child with SMA-1, Nusinersen drug costs alone for the first five years-of-life are estimated at USD 2,295,000 (based on USD 127,500 per dose: 6 in the first year and 3 maintenance doses for each year thereafter) [64]. The cost of the adapted equipment recommended in this case report for the same time period (see Table 4) is estimated at USD 125,500. Adaptive equipment that increases function, participation and quality of life represents approximately 0.05% of the drug cost. If society accepts the cost of life-saving drugs and other medical interventions, we propose that providing the equipment needed to support activity and participation must also be considered standard of care.

Article 23 of the UN Convention on the Rights of the Child [24] refers to the obligation of States parties and recognizes that a child with mental or physical disabilities is entitled to 'enjoy a full and decent-life, in conditions that ensure dignity, promote self-reliance and facilitate the child's active participation in the community.' More importantly, the article emphasizes the need for inclusion of children with disabilities in society and provision of basic services to eradicate stigmatization. Of note is that only one study identified in this scoping review included children from low- and middle-income countries. Children with

complex or life-limiting disabilities may be at even greater risk for violation of their rights in these contexts.

Table 4. Estimated adaptive equipment costs.

Equipment	
Item	Cost in USD (website source where available)
Power wheelchair	USD 12,000–30,000 replacement every 5 years
Custom adaptive seating	USD 2000–4000 replacement at least every 2 years
Supine standing frame	USD 2000–4000 replacement every 5 years (www.adaptivemall.com)
Manual wheelchair	USD 2–6000 replacement every 5 years
Gait Trainer/Dynamic Mobility Device	USD 2000–4000 replacement every 5 years (www.adaptivemall.com)
AFO's	USD 2000–4000 replacement every 1–2 years
TLSO	USD 2000–4000 replacement every 1–2 years
Wrist splints	USD 50–100 replacement every 1–2 years
Bath/Toilet Chair	USD 500–2000 (www.adaptivemall.com)
Ceiling Track lift system	USD 3500
Adapted wheelchair van	USD 60–80,000
Ramp for home accessibility	USD 400–2000
Special-needs adapted tricycle	USD 600–2500 (www.adaptivemall.com)
SGD and communication software	USD 2000–10,000 depending on model and features
Car support harness	USD 200 (www.especialneeds.com)

AFO: ankle-foot orthoses; Bi-PAP: Bi-level positive airway pressure; SGD: speech generating devices; TLSO: thoraco-lumbar-sacral orthosis.

4.7. Study Limitations

This scoping review was conducted in only two databases, and it is possible that we may have missed studies referenced in other sources, published in other languages or in grey literature. However, we included a Google Scholar search and reviewed websites relevant to SMA, as well as hand-searching references from the international Delphi consensus and systematic reviews of relevant interventions to ensure our search was relatively complete. A formal quality rating was not used since studies were descriptive and heterogeneous, and scoping reviews rarely employ quality ratings for these reasons [23]. Although the research evidence found was limited in study size and design, we contrasted this with studies including children diagnosed with SMA-2. Children who are described as 'treated' SMA-1 may have a trajectory more similar to children who develop sitting before onset of symptoms and interventions recommended for this group could also be considered.

The scoping review was supported by a single case report and results cannot be generalized to all children with SMA-1, or even all those who began treatment at a similar length of time after symptoms presented. Tom has consistently responded well to the pharmaceutical intervention and has made steady gains in strength, development and overall health. However, initially, we did not know his long-term prognosis and age-appropriate adaptive device interventions were introduced as soon as feasible and then modified and progressed as his abilities progressed. Devices introduced in the earliest days may be appropriate to consider for children who have a more severe trajectory, while those introduced as he gained in strength may be considered earlier for children on a less severe trajectory.

5. Conclusions

New and promising pharmaceutical treatments are changing the prognosis and functional trajectory for children with SMA. Scoping review results were enhanced by a case-report describing one child with SMA-1 who was diagnosed early in the disease process and accessed one of these treatments through a nearby research study. Effective medical treatment was accompanied by proactive OT and PT intervention that was evidence-informed and assisted him to access assistive devices that supported his development and maximized his function and participation. His human rights were maintained, and he was enabled to move independently inside and outside of his home, as well as to play, learn and grow.

Young children with significant and life-limiting disabilities such as SMA-1 need a variety of ways to participate in age-appropriate activities and family routines. Funding for medical interventions aimed at reducing mortality must also be accompanied by a broader perspective; one that not only includes orthopedic surveillance and interventions to promote and maintain BSF, but one that ensures provision of the devices and supports that will increase activity and participation. The provision of individually prescribed adaptive positioning, assistive devices and augmented mobility should be considered standard of care for all children with SMA-1. Although research evidence is limited, a comprehensive and proactive approach using adaptive equipment is needed to enhance function, fun and participation with family and friends.

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Conflicts of Interest: G.P. has worked as an educational consultant for various manufacturers and suppliers of standers and gait trainers. This study was unfunded and this manufacturer relationship did not influence the manuscript in any way. R.L. declares no conflict of interest.

Appendix A. Measures Included in Case Report

- Pediatric Evaluation of Disability Inventory (PEDI) caregiver assistance scale [28] describes the amount of assistance children require in activities of daily living, mobility and social skills. Items may be reported on a 6-point ordinal scale. Independent indicates the child can do the task alone; Supervision indicates verbal supervision is needed; Minimal indicates caregiver may need to provide a small amount of physical assistance, Moderate indicates the caregiver may complete up to half the task; Maximal indicates the caregiver does more than half the task but the child may provide a small amount of meaningful assistance; and Total indicates the child is completely dependent. The PEDI has demonstrated reliability and validity in multiple studies [38,65].
- Assessment of Learning Powered Mobility (ALP) measures the power mobility skill process and is suitable for all ages, motor and cognitive abilities [29]. It includes eight phases from novice (ALP 1) to expert (ALP 8). At phases 1 or 2 children are just beginning to explore effects of the switch or joystick and by phase 3 they establish cause-effect. At phases 4 and 5 children explore different effects and directional control.

Basic steering control is demonstrated at phase 6 and at phase 7 they can use the power mobility device to participate in other activities. The ALP has established validity for a wide range of ages and abilities and inter-rater reliability has recently been established between experienced professionals ($ICC_{2,1}$ 0.83) [66] and between professionals and relatives or assistants (κ_w 0.85) [67].

- Power Mobility Program (PMP) [30] measures abilities to complete a list of 34 power mobility tasks ranging from basic mobility to integrated skills in structured and unstructured environments. It is scored on a 6-point ordinal scale depending on amount of assistance required from 1 (maximal assistance) to 5 (age-appropriate supervision). Maximum raw score is 170 and, where all items can be scored, total score can be divided by 34 for a summary score of 1–5 describing average assistance required. Good inter-rater reliability (κ_w 0.87) and fair intra-rater reliability (κ_w 0.52) was reported from repeated assessment of 9 children with physical disabilities in the original study [30]. Recently other researchers reported excellent intra-rater ($ICC_{2,1}$ 0.97/0.98) and good to excellent inter-rater reliability ($ICC_{2,1}$ 0.93/0.87) from assessment of 30 children over two occasions [66]. It has been widely used in paediatric power mobility research although concerns remain regarding its structure and content [54]. To illustrate sections more challenging to score in community settings (and with very young children with significant disabilities) we reported scoring separately from each section within the tool. Basic mobility skills include three sections: beginning skills (including ability to turn the wheelchair on/off); directional control; and speed control. Integration of basic skills in structured environments also has three sections: functional skills (doorways, hallways and curving paths); negotiating ramps; and negotiating sidewalks. The final section is integration of basic skills for functional mobility in unstructured environments and includes community mobility skills.
- Power Mobility Training Tool (PMTT) [31] was developed to guide power mobility training for children at the very beginning stages of learning. Content validity was established through multiple methods including review by an international expert panel. It includes non-motor, motor and driving skills subscales and items are scored on a 5-point ordinal scale for a maximum score of 48. A score of 1 indicates that the task can only be completed with assistance, 2 indicates independent demonstration less than 50%, 3 indicates independent demonstration more than 50% and a score of 4 means the child completes the task independently more than 90% of the time.
- Excellent Spearman correlations (0.83–0.92, $p < 0.001$) have been measured between PMTT, PMP and ALP scores [26].
- Wheelchair Outcome Measure for Young People (WhOM-YP) [32] is an individualized, client-centred measure developed for children or young people who use wheeled mobility. Depending on the child's age or abilities, importance and satisfaction of meaningful participation-focused outcomes related to use of a wheeled mobility device may be rated either by the child or parent proxy. It is scored on an 11-point ordinal scale and has evidence of face and content validity with ability to discriminate known groups and good association between parent and child ratings. Test-retest reliability for parent proxy ratings is excellent ($ICC_{2,1}$ 0.85–0.90) and minimal detectible change (MDD_{95}) has been estimated as 1.8 [32].

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