Recommendations for Studies on Dynamic Arm Support Devices in People with Neuromuscular Disorders: a Scoping Review with Expert-Based Discussion

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Neuromuscular disorders are characterized by muscle weakness that limits upper extremity mobility, but can be alleviated with dynamic arm support devices. Current research highlights the importance and difficulties of evidence-based recommendations for device development. We aim to provide research recommendations primarily concerning upper extremity body functions, and secondarily activity and participation, environmental and personal factors. Evidence was synthesized from literature, ongoing studies, and expert opinions and tabulated within a framework based on a combination of the ICF model and contextual constructs. Current literature mostly investigated the motor capacity of muscle function, joint mobility, and upper body functionality, and a few studies also addressed the impact on activity and participation. In addition, experts considered knowledge on device utilization in the daily environment and characterizing the beneficiaries better important. Knowledge gaps showed that ICF model components and contextual constructs should be better integrated and more actively included in future research. First, integrate multiple ICF model components and contextual constructs within one study design. Second, include the influence of environmental and personal factors when developing and deploying a device. Third, include short-term and long-term measurements to monitor adaptations over time. Finally, include user satisfaction as guidance to evaluate the device effectiveness.

Keywords: neuromuscular disorders; dynamic arm support; body functions; ICF model; contextual constructs

1. Introduction

Neuromuscular disorders (NMD) are characterized by muscle weakness that limits upper extremity mobility and can affect people of all ages (Deenen et al., 2016). The worldwide prevalence of NMD is 160 per 100,000, which is similar to that of Parkinson's disease and double that of multiple sclerosis (Deenen et al., 2015). People with NMD commonly experience difficulties to perform movements against gravity,

such as lifting the arms or reaching for an object (Bergsma, Cup, et al., 2017). These functional limitations translate into problems with activities of daily life (ADL) and participation in society. Various types of dynamic arm support devices (DAS) have been developed that improve engagement in ADL by providing gravity compensation (van der Heide et al., 2015; van der Heide et al., 2014). Generally, such devices relieve upper extremity limitations that stem from muscular weakness (Coscia et al., 2014; Ellis et al., 2008; Prange et al., 2009). However, the impact might differ between devices intended for rehabilitation and research and wearable devices intended for daily life. As approximately 7 to 24% of the Dutch population with NMD, and 8.5% of people with Duchenne Muscular Dystrophy (DMD) worldwide (Janssen et al., 2014), use a DAS in daily life (Bergsma, Janssen, et al., 2017), it is relevant to understand how and for what purposes the DAS are used on a daily basis.

The perceived benefits of a DAS vary from complete satisfaction to no perceived added value, where daily life usage has been reported to be discontinued over time (Rahman et al., 2007; Shank et al., 2017; van der Heide & de Witte, 2016). Current research highlights the importance of evidence-based recommendations for DAS development and prescription. However, determining recommendations has proven to be difficult (van der Heide et al., 2015), due to the diversity in NMD, DAS, and study designs. This is further complicated by the lack of standardized and validated evaluation tools in current research (Janssen, Lobo-Prat, et al., 2019; Shank et al., 2017; van der Heide et al., 2015). To optimize DAS development and its use in daily life, it is important to investigate the users' characteristics, DAS function, and resulting user-device interactions in the short and long term. According to the Consortium for Assistive Technologies Outcome Research framework, used by Heide et al. 2015, the WHO's International Classification of Functioning, Disability and Health (ICF) model

could act as the primary guidelines (Jutai et al., 2005; van der Heide et al., 2015; World Health Organization. (2001). *International classification of functioning, disability and health : ICF*. World Health Organization. <https://apps.who.int/iris/handle/10665/42407>).

Current research indicates that DAS impact the users abilities across all ICF components (van der Heide et al., 2015). However, it is commonly assumed that a DAS primarily affects body functions through gravity compensation, which shapes the effects on activity and participation. Furthermore, Heide et al. indicated that the ability of a DAS to support ADLs does not guarantee higher performance or even utilization in a home environment (van der Heide et al., 2015). Therefore, it is important to account for the contextual differences in which the activities are performed. For example, there is a difference in what people are able to do in a standardized environment by following instructions of an examiner compared to what they actually do in their daily life. Holsbeeke et al. 2009 described three constructs, or concepts, for these contextual differences: motor capacity (can do in a standardized environment), motor capability (can do in a daily environment), and motor performance (actually do in a daily environment) (Holsbeeke et al., 2009). We propose that a combination of the ICF model as primary guidelines and the three contextual constructs as secondary guidelines would provide a suitable framework to structure the evidence of DAS evaluation and recommendations for future development.

Technological advances in DAS, such as wearable robotics, are developing rapidly and it is expected that they will become more pervasive in daily life support systems (Bergsma, Janssen, et al., 2017; Janssen et al., 2014). Yet the research on DAS evaluation in patients is relatively new and under

development (Janssen, Lobo-Prat, et al., 2019; van der Heide et al., 2015). Perspectives on the state of the art from third parties who are either engaged in development and prescription of these devices, or are end users, could provide important insights which are often lacking in the literature. The current study aims to synthesize the literature with expert opinions in order to provide an overview of current evidence and identify knowledge gaps that may limit the development of DAS. A secondary aim is to provide research recommendations to establish a standardized and validated approach for DAS evaluation in people with NMD.

2. Methods

2.1 Literature search

Inclusion criteria for the literature review focussed on studies that evaluated a DAS intended for daily life situations that supported the lower arm through gravity compensation. Studies with healthy participants only were included if the DAS was a finalized prototype designed for daily use by people with NMD. Furthermore, studies needed to report measures involving body functions described as neuromusculoskeletal and movement-related functions (*World Health Organization. (2001). International classification of functioning, disability and health : ICF. World Health Organization. <https://apps.who.int/iris/handle/10665/42407>).* Other inclusion and exclusion criteria can be found in the appendix Table 4. Both scientific and non-commercially published literature were searched.

Scientific literature was searched in PubMed and Web of Science in August 2018 and updated in February 2020. The search strategy for each database can be found in the appendix Table 4. Titles and abstracts were independently screened by two

researchers (HE, AP) where remaining articles were compared and finally in-/excluded. Included articles' authors and reference lists were then searched for additional articles.

Non-commercially literature published in the past 5 years (2015-2020) was searched from a government clinical trial database (clinicaltrials.gov), DAS suppliers' websites identified from previously included articles, and research mentioned by the experts. The search strategy for the government clinical trial source consisted of the combination of 'Neuromuscular Disorders' or 'Neuromuscular Diseases' with either 'Robot', 'Exoskeleton', or 'Arm support'. One researcher (JE) gathered the information and another researcher (AP) checked for consistency with the in-/exclusion criteria. Any inconsistency was discussed until agreement was reached.

2.2 Focus groups

Five focus groups were formed from a patient community, DAS developer, clinical, rehabilitational, and research setting. The groups consisted out of fifteen experts in total: two members of a patient community with a neuromuscular disorder, five DAS developers/suppliers, one physician, five therapists, and two researchers. At that time, one member with a NMD used a DAS in daily life and the other was orientating. The experts were most experienced with muscular weakness from atrophy/dystrophy or lesions to the central nervous system. Two developers/suppliers also had direct contact with some clients. Eight people were involved in research: two full-time researchers, one member with a NMD evaluated research proposals in a funding comity, and two developers, two therapists, and one physician were partially involved in projects involving DAS development, training, and improvement of DAS selection procedure.

The groups were interviewed by three researchers (JE, EP, ACW) in person on separate occasions. The interviews were in dutch, semi-structured, and guided by the (originally dutch) questions from the appendix. The questions were formed based on preliminary review of the literature and discussion between the research team.

Information was distilled of the focus groups' views on 1) current impairments/limitations of people with NMD, 2) effectiveness and requirements of a DAS linked to the previous, 3) previous research projects and remaining questions, and 4) research priorities. The interviews were audiotaped in support of the keywords which were noted during the interview by at least two researchers and evaluated afterwards. Keywords were formulated based on the terminology used in previous literature and descriptions were added for clarification purposes.

2.3 Evidence synthesis

Fundamental topics from literature and interviews were tabulated and summarized to synthesise current evidence with expert opinions. The tabulation framework was formed with the ICF model components (body functions, activity and participation, and environmental and personal factors) as rows and contextual constructs (motor capacity, - capability, and - performance) as columns. Furthermore, ICF model components were further divided as categories and sub-categories to increase the level of diversity.

Current evidence matching expert opinions were indicated in each table cell by the reference number. Cells where current evidence was lacking, despite experts' interests, were considered knowledge gaps and indicated by a dash symbol. We formulated our synthesized current evidence, knowledge gaps, and research recommendations according to the terminology used for conceptual modelling of assistive technology device outcomes (Jutai et al., 2005).

3. Results

3.1 Synthesized evidence

Current evidence and knowledge gaps were synthesized per category based on literature findings and expert opinions and presented in Table 3. There were five categories identified for body functions, three for activity and participation, and one for environmental and personal factors. The user-device interaction was considered handling an object and therefore categorised under activity and participation. Body functions was consequently linked to user-device interaction as a sub-category. Literature covered roughly nine out of 51 cells within the body functions component, which also covered eight out of 19 cells within the activity and participation component, but none for the cells within the environmental and personal factors.

3.2 Literature review

The literature search resulted in 599 hits (PubMed: 194, Web of Science: 405) of which 557 unique articles (Figure 1). Then, 503 articles were excluded based on title, another 39 based on abstract, and another ten after a full read of the article. Two articles were added after reference cross checks. Finally, seven articles were included for reviewing after the inclusion process (Table 1) (Dunning et al., 2016; Essers et al., 2013; Estilow et al., 2018; Haumont et al., 2011; Kooren et al., 2015; Lebrasseur et al., 2019; van der Heide et al., 2017). Two studies, ongoing until December 31st 2020 and 2019, respectively, were identified from the government clinical trial source (Table 2) (Bendixen, 2019; Pedrocchi, 2019).

Figure 1. Flowchart of mixed-methods process

3.3 Dynamic arm support evaluations

The evaluated DAS were a prototype A-gear (*Flexension*; Kooren et al., 2015), and commercially available devices Armon Ayura (*Armon Products*; Bendixen, 2019; Pedrocchi, 2019), Gowing (*Focal Meditech B.V.*; Lebrasseur et al., 2019), JAECO WREX (Bendixen, 2019; Estilow et al., 2018; Haumont et al., 2011; *JAECO Orthopedic*; Pedrocchi, 2019), SLING (Essers et al., 2013; *Focal Meditech B.V.*; van der Heide et al., 2017), and Top-Help (*Focal Meditech B.V.*; van der Heide et al., 2017), and a JAECO WREX modified with a trunk support prototype (Table 1) (Dunning et al., 2016). These DAS provide gravity compensation through adjustable counter-weights (SLING) or springs (A-gear, JAECO WREX, Top-Help) with an additional actuator that adjusts the springs' tension (Armon Ayura, Gowing). All studies investigated the body functions, several also investigated activity and/or participation (Bendixen, 2019; Pedrocchi, 2019; van der Heide et al., 2017), and one the satisfactory levels of using the DAS (Lebrasseur et al., 2019). All study designs were set up to cross-sectionally investigate motor capacity during functional and ADL tasks w/o a DAS. Effects were mostly quantified for muscle function, joint mobility, and upper

body functionality. Main effect identified were: DAS can lower up to 15% activity of the Trapezius muscle in healthy people; DAS can increase arm elevation from 25 up to 100° and elbow flexion from 10 up to 120° in people with NMD; Performance of the Upper Limb (PUL) scores were improved by 20 up to 30% with a device in people with NMD and showed a large effect (>1.15) in the modified Upper Extremity Performance Test for the Elderly (TEMPE); and the ability to perform ADL w/o a support device was participant specific where a device could have a limiting and beneficial effect. The ongoing studies perform longitudinal interventions of eight and five weeks, respectively, to field-test two DAS (Table 2) (Bendixen, 2019; Pedrocchi, 2019). We considered these studies to investigate the motor capacity, - capability and - performance regarding upper body functionality and activity/participation levels.

3.4 Expert opinions

Experts expressed their opinions mainly in identifying DAS effects on muscle function, joint mobility, and upper body functionality (Table 3 and appendix Table 5). They viewed people with NMD as impaired in these aspects of body functions and believed a DAS could compensate specific impairments. For example, a DAS improves joint mobility and lowers muscle efforts. This combination allows a user to perform multiple repetitions in a greater workspace needed in self-care activities such as eating and drinking. However, a DAS often does not fully support the complete hand to mouth motion and is then considered limited in their effectiveness. Therefore, experts believe further research is necessary to support DAS development not only on body functions but on all ICF components and contextual constructs. Furthermore, experts advocated to expand and improve the descriptions of the (active) population and disease progression. Specifically, device utilization in the daily environment was of interest to bridge the knowledge from motor capacity to motor capability and motor performance.

Reference	Research aim	Study design, population, DAS, and tasks	Outcome parameters and interpretations	Research Recommendations
Dunning (2016)	To explore the effect of the JAECO WREX with trunk motion capability on the total range of motion and balancing quality of the arm support.	<u>SD</u> : Cross-sectional <u>P</u> : Healthy (N=3) <u>DAS</u> : JAECO WREX with prototype trunk motion capability <u>T</u> : Functional task and ADL (single joint motions, reaching and touching various places, and drinking w/o DAS)	- Range of Motion (kinematics) - Normalized muscle activity levels Range of motion increased by 10% and less muscle activity is needed with support.	Use the same degrees of freedom in the DAS as a human arm which allows for better alignment and natural behavior. The effect of a body-bound DAS with trunk motion capability was perceived as important by both healthy subjects and three DMD patients who shortly tried the DAS without measurements. Movements were easier and more natural to perform and DMD patients mentioned that it also allowed for making compensatory movements.
Essers (2013)	To compare the influence of gravity compensation between joint moments in populations performing a reaching task.	<u>SD</u> : Cross-sectional <u>P</u> : Muscular Dystrophy (Facioscapulohumeral Dystrophy) (N=3) and Healthy (N=3) <u>DAS</u> : SLING <u>T</u> : Functional tasks (reaching at shoulder level w/o DAS)	- Joint moments (kinetics) - Execution time (kinematics) Joint moments were lower with DAS but execution time was longer.	Designing better support devices should include biomechanical considerations. Further research should be focused on expanding the investigation on the influence of gravity compensation in muscular dystrophy subjects in order to quantify the real benefits of an arm support device.
Estilow (2018)	To evaluate the efficacy of the JAECO WREX on improvement of active range of motion of the upper extremity and ADL performance.	<u>SD</u> : Cross-sectional <u>P</u> : Muscular Dystrophy (Duchenne) (N=9) <u>DAS</u> : JAECO WREX <u>T</u> : Functional task and ADL (active shoulder motions, eating/drinking, simulated facial grooming, and item retrieval w/o DAS)	- Active Range of Motion (kinematics) - Manual muscle strength testing Improvements in active range of motion of shoulder and elbow joints depended on the patients' strength.	Further exploration of agonist and antagonist strength ratios is needed with respect to the muscle groups that receive assistance. Longitudinal studies are necessary to evaluate the WREX's regarding muscle strength and ARange of motion preservation. Future research should include the use of diagnosis-specific measures and an ordinal ADL scale. In addition, research is needed to identify clinical measures that predict successful performance with the WREX to allow clinicians to appropriately evaluate and recommend the WREX for patients with neuromuscular diseases.
Haumont (2011)	To describe the JAECO WREX and to assess the functional improvement in the upper extremity.	<u>SD</u> : Cross-sectional <u>P</u> : Arthrogryposis Multiplex Congenita (N=1) and Spinal Muscular Atrophy (N=2) <u>DAS</u> : JAECO WREX <u>T</u> : Functional tasks and ADL (shoulder motions, reaching, and eating/drinking w/o DAS)	- Range of Motion (kinematics) There was greater mobility of the upper extremity in motions against gravity with the DAS.	Future DAS should assist in pronation or supination as these motions are an integral part of the feeding movement.

Table 1. Study characteristics of completed studies. SD: Study Design, P: Population, N: number of participants, T: Tasks, DAS: Dynamic Arm Support, w/o: with and without.

Reference	Research aim	Study design, population, DAS, and tasks	Outcome parameters and interpretations	Research Recommendations
Heide (2017)	To evaluate the Motion and Muscle Ambulatory Activity System at a home setting and investigate range of motion and ADL performance w/o DAS.	<u>SD</u> : Cross-sectional <u>P</u> : Stroke (N=2), amyothrophic lateral sclerosis (N=1), muscular dystrophy (N=1), and spinal stenosis (N=1) <u>DAS</u> : Top-help and SLING <u>T</u> : ADL (reaching and eating/drinking w/o DAS)	- Range of Motion (kinematics) Shoulder range of motion increased with DAS but varied greatly between individuals.	To understand the real effects of these devices it is recommended to take this individual variation into account and that benefits are also assessed in a broader daily life context. Insight into benefits of DAS and influencing factors could support the provision and the selection process of DAS.
Kooren (2015)	To develop, pilot test, and present the characterization and validation of the A-gear.	<u>SD</u> : Cross-sectional <u>P</u> : Duchenne (N=3) and Healthy (N=1) <u>DAS</u> : A-gear (prototype) <u>T</u> : Functional tasks and ADL (shoulder and elbow motions, eating/drinking, use phone/computer, self-care, and PUL w/o DAS)	- Range of Motion (kinematics), - Distance covered (kinematics), - Comfort (self-reported) PUL score was increased with less compensatory movements. Arm movements forward and upward became easier. The DAS was comfortable.	One-hundred percent weight compensation is not always preferred by patients. One of the patients wanted less supporting force, which felt more comfortable to him. The reduction of compensatory movements is very important, as compensatory movement consumes a lot of energy and therefore they restrict the endurance to perform daily activities. Two sided support is preferred to avoid a skew posture. Forward lean capability is much appreciated. DAS preferably does not run between arm and trunk, or add considerable volume underneath the forearm and elbow. Such components create an uncomfortable environment for arm relaxation and can clash with tabletops.
Lebrasseur (2019)	To evaluate the usability of the Gowing power-assisted arm support.	<u>SD</u> : Cross-sectional <u>P</u> : Multiple sclerosis Spinal (N=3), muscular atrophy (N=3), and muscular dystrophy (N=3) <u>DAS</u> : Gowing <u>T</u> : Functional tasks and ADL w/o DAS	- Disabilities of the Arm, Shoulder and Hand (DASH) - modified Upper Extremity Performance Test for the Elderly (TEMPA) - Quebec User Evaluation of Satisfaction with assistive Technology (QUEST) The DAS improved TEMPA scores and two third were quite or very satisfied with the device.	Different actively actuated devices could be compared in manual tasks and in the long-term to assess long-term benefits. Future designs should include hand exoskeleton or wrist support on a DAS to investigate the influence of fine movement improvements. Proper use of DAS in research might depend on correct installation and should receive attention in the experimental setup.

Table 1 (continued). Study characteristics of completed studies. SD: Study Design, P: Population, N: number of participants, T: Tasks, DAS: Dynamic Arm Support, w/o: with and without.

Reference	Research title and aim	Study design, population, DAS, and tasks	Outcome parameters	Research relevance
Bendixen (2019)	<p>Title: Use of Dynamic Arm Support Devices for Upper Limb Function in Non-Ambulatory Men With DMD</p> <p>Aim: To promote participation in activities of daily living in non-ambulatory individuals with DMD with upper extremity weakness.</p>	<p>SD: Longitudinal, randomized, and interventional</p> <p>P: Duchenne Muscular Dystrophy</p> <p>DAS: Armon Ayura and JAECO WREX</p> <p>T: - Baseline (two weeks)</p> <p>- Device trial (four weeks)</p> <p>- Post device trial (two weeks)</p>	<p>Primary:</p> <p>- Change in Upper Extremity Acceleration through Actigraphy</p> <p>- Change in Upper Extremity Position through Actigraphy</p> <p>Secondary:</p> <p>- Goal Attainment Scale</p> <p>- Physical Motor Assessment</p> <p>- PUL</p>	<p>Upper extremity performance will be further quantified with use of a physical motor assessment, the PUL assessment, and patient reported outcomes.</p> <p>Data gleaned will provide important knowledge and objective results regarding the potential benefit of DAS in individuals with DMD with limited functional use of their upper extremities.</p>
Pedrocchi (2019)	<p>Title: USEFUL: User-centred Assistive SystEm for Arm Functions in neUromuscuLar Subjects</p> <p>Aim: To field-test the improvement in arm functions provided by DAS and assessing their impact to users' quality of life and independence</p>	<p>SD: Longitudinal, cross-over, and interventional</p> <p>P: Muscular Dystrophies (Duchenne, Becker, Limb-Girdle Type 2)</p> <p>DAS: Armon Ayura and JAECO WREX</p> <p>T: - Baseline (cross-sectional)</p> <p>- Short Training DAS 1 (three days)</p> <p>- Domestic use DAS 1 (two weeks)</p> <p>- Short Training DAS 2 (three days)</p> <p>- Domestic use DAS2 (two weeks)</p>	<p>Primary:</p> <p>- PUL</p> <p>Secondary:</p> <p>- Motor Function Measures scale</p> <p>- Brooke scale</p> <p>- ABILHAND</p> <p>- PedsQL</p> <p>- PROMIS FATIGUE</p> <p>- Personal Adjustment and Role Skills Scale III</p> <p>- Technology Acceptance Model</p> <p>- System Usability Scale</p>	<p>Field-testing is essential to assure a widespread accessibility to these devices for most of the potential users, possibly providing health providers with direction and guidance towards Health Technology Assessment.</p>

Table 2. Study characteristics of ongoing studies. DMD: Duchenne Muscular Dystrophy, SD: Study Design, P: Population, T: Tasks, DAS: Dynamic Arm Support.

ICF component	Category	Sub-category	Motor Capacity	Motor Capability	Motor Performance	Expert views (E) and literature findings (L)	Synthesized research recommendations
Body function	Muscle	Activity	1	—	—	E: for which muscles can a DAS reduce muscle activity? L: activity was reduced in the Trapezius, Deltoid, and Pectoralis Major muscles. E: does a DAS require compensatory strategies?	Explore the effects of external force generated by DAS on muscle function of the musculoskeletal system.
		Coordination	—	—	—		Determine whether an external force introduces a burden on other muscles or if it relieves pain and stiffness.
		Fatigue	—	—	9	E: how can we measure and prevent fatigue? L: no findings available from ongoing studies.	
		Pain	—	—	—	E: can a DAS relief muscle pain?	
		Stiffness	—	—	—	E: can a DAS reduce muscle stiffness?	Investigate the relation between muscle effort reduction and muscle coordination of the shoulder girdle.
		Strength	3	—	—	E: what are required strengths to operate a DAS? L: potential DAS effects are related to residual muscle groups' strength.	Prescribe user strength requirements to a device.
	Joint	Kinematics	1, 2, 4, 5, 6	—	—	E: can we map the differences in motion w/o DAS? L: a DAS positively affects the reachable workspace.	Explore the effects of an external force generated by DAS on the joints of the musculoskeletal system.
		Kinetics	3,	—	—	E: can we map the interacting forces for motions with a DAS? L: gravity compensation reduces joint torques and thus effort.	Investigate the relation between improvements in joint load reduction and mobility and joint stability, pain, and stiffness.
		Pain	—	—	—	E: can a DAS relief joint pain?	
		Stiffness	—	—	—	E: can a DAS overcome joint rigidity and feelings of stiffness?	Investigate the relationship between levels of external force, joint loading, and mobility across the range of motion.
Upper body	Functionality		7, 8, 9	—	—	E: can we distinguish different aspects of upper body functionality improvements? L: the DAS had a large positive effect on most items. No findings available from ongoing studies.	Investigate the effects of a DAS on the upper body as a whole by including other regions than the shoulder area.
		Hand/Wrist function	—	—	—	E: what is the influence of a DAS on hand/wrist function?	
		Motion/Posture	2	—	8	E: how can we support natural motion/support? L: an integrated DAS with trunk support positively affected participants' posture. No findings available from ongoing studies.	Explore what can be improved by integrating devices that support other regions than the shoulder area.

Table 3. Tabulation and summary of literature (L) and expert views (E) and synthesis of research recommendations. Current evidence, or knowledge gaps, within experts' interests were indicated in each cell by the reference number(s) or a dash, respectively. Blank cells either lacked experts' interests or were considered theoretically impossible. NMD: NeuroMuscular Disorders, ADL: Activities of Daily Living, DAS: Dynamic Arm Support, w/o: with and without. 1. (Dunning et al., 2016) 2. (Kooren et al., 2015), 3. (Essers et al., 2013), 4. (Estilow et al., 2018), 5. (van der Heide et al., 2017), 6. (Haumont et al., 2011), 7. (Lebrasseur et al., 2019) , 8. (Bendixen, 2019), 9. (Pedrocchi, 2019).

ICF component	Category	Sub-category	Motor Capacity	Motor Capability	Motor Performance	Expert views (E) and literature findings (L)	Synthesized research recommendations
Body function	Population description	Disease progression	—	—	—	E: can we expand knowledge of the (active) population on muscle/joint/upper body aspects?	Limited knowledge on disease progression is presented on a population level for DAS users. Perform a review on disease progression in NMD on functionality and disability.
	Others	Short term tests	—	—	—	E: current measurements are too time-consuming for participants.	Performance tests are considered too long due to the rapid onset of fatigue within this population. Furthermore, disease progression or DAS effects are not captured to a sufficiently distinctive level in a cross-sectional design. Develop tests which require less from the population and can be applied on multiple occasions.
		Long term tests	—	—	8, 9	E: cross-sectional studies are not representative for disease progression. L: no findings available from ongoing studies.	
		Test accuracy and discriminative properties	—	—	—	E: current methods do not capture disease progression properly enough.	
Activity & Participation	User-device interaction	Body function aspects	1, 2, 3, 4, 5, 6	—	8, 9	E: what are the effects of a DAS on body function aspects and how do they interact? L: a DAS reduces efforts and increases reachable workspace, depending on residual muscle strength. No findings available from ongoing studies.	Research should combine multiple aspects to investigate interactions within body functions.
		Functionality	5, 7, 8, 9	—	9	E: what types of ADL are possible, impossible, or limited w/o a DAS? L: the DAS positively affected the collectively measured ADL. No findings available from ongoing studies.	Relate the performance of ADL to body functions and identify areas for device development.
		Quantified device utilization			—	E: can we quantify device utilization and recognize non-usage?	Identify non-usage in daily life as an indicator for disease progression. Monitor performance and identify negative adaptations.
		Quality of performance			8, 9	E: can we monitor performance of ADL at home? L: no findings available from ongoing studies.	

Table 3 (continued). Tabulation and summary of literature (L) and expert views (E) and synthesis of research recommendations. Current evidence, or knowledge gaps, within experts' interests were indicated in each cell by the reference number(s) or a dash, respectively. Blank cells either lacked experts' interests or were considered theoretically impossible. NMD: NeuroMuscular Disorders, ADL: Activities of Daily Living, DAS: Dynamic Arm Support, w/o: with and without. 1. (Dunning et al., 2016) 2. (Kooren et al., 2015), 3. (Essers et al., 2013), 4. (Estilow et al., 2018), 5. (van der Heide et al., 2017), 6. (Haumont et al., 2011), 7. (Lebrasseur et al., 2019) , 8. (Bendixen, 2019), 9. (Pedrocchi, 2019).

ICF component	Category	Sub-category	Motor Capacity	Motor Capability	Motor Performance	Expert views (E) and literature findings (L)	Synthesized research recommendations
Activity & Participation	Population description	Needs and goals		—	8,9	E: what are the needs and goals of people with NMD? L: no findings available from ongoing studies.	Describe the population so it connects the desired motor capabilities and performance to the capacity. Integrate aspects of personal and environmental factors.
		Functionality	5,7,8,9	—	8,9	E: what types of ADL are people with NMD capable of and do they perform those at home? L: observations revealed participant-specific possibilities for ADL. No findings available from ongoing studies.	
	Others	Ambulant/ Body mounted	—	—	—	E: what is the influence of being ambulant and/or wearing a body-mounted DAS on device utilization?	User-device interaction evaluations should be applicable to a large variety of users, yet discriminative.
		Test accuracy and discriminative properties	—	—	—	E: current methods are limited in capturing user-device interaction properly.	It is suggested to have a flexible and tailored method, such as wearable sensors, to measure continuously in varying settings.
Environmental and Personal factors		Home setting and outdoors		—	—	E: what are factors in the user's environment that influence the user-device interaction?	Environmental and personal factors are under-investigated influencers of user-device interactions. Studies should consider barriers and facilitators for activity monitoring.
		Psychological drives			—	E: what is the influence of motivation on user-device interaction?	
		Distribution of energy			—	E: can we estimate energy costs through activity monitoring and effort reduction by DAS?	

Table 3 (continued). Tabulation and summary of literature (L) and expert views (E) and synthesis of research recommendations. Current evidence, or knowledge gaps, within experts' interests were indicated in each cell by the reference number(s) or a dash, respectively. Blank cells either lacked experts' interests or were considered theoretically impossible. NMD: NeuroMuscular Disorders, ADL: Activities of Daily Living, DAS: Dynamic Arm Support, w/o: with and without. 1. (Dunning et al., 2016) 2. (Kooren et al., 2015), 3. (Essers et al., 2013), 4. (Estilow et al., 2018), 5. (van der Heide et al., 2017), 6. (Haumont et al., 2011), 7. (Lebrasseur et al., 2019) , 8. (Bendixen, 2019), 9. (Pedrocchi, 2019).

4. Discussion

4.1 Current evidence and knowledge gaps

The aim of this scoping review was to provide research recommendations for DAS evaluation based on a synthesis of literature and expert opinions. We primarily focused on body functions and secondarily on other ICF model components: activity and participation, and environmental and personal factors. To structure the evidence and identify gaps we used a framework that combined the ICF model components with contextual constructs: motor capacity, capability, and performance. Most included studies focused on the user-device interaction within the framework cells of body functions and motor capacity. Typically, they studied the introductory phase of using a DAS, with just a few studies addressing the long-term adaptations. The lack of standardized evaluation tools posed difficulties in creating comparable evidence (Lebrasseur et al., 2019) and the synthesis of current evidence (van der Heide et al., 2015). The following knowledge gaps were identified: first, we poorly understand the adaptations that may ensue following skill acquisition, fatigue, or disease, which alter the support requirements over time. Second, it is yet unclear how abilities across ICF components and contextual constructs are related. For instance, it is unclear how changes in body functions influence the activity and participation and how these changes are, in turn, the result of environmental setting and task requirements. Finally, various aspects, such as comfort levels, were considered important by the experts that have not yet gained sufficient attention in the scientific literature.

4.2 Adaptations over time

From this review it is clear that the ability to adapt following skill acquisition, fatigue, or disease's progression, have not yet been investigated over time. In order to benefit

from a DAS, it is crucial that the user acquires the skills to operate the device and retains them over time. Previous studies have shown that training with a DAS is feasible in people with NMD (Heutinck et al., 2018; Jansen et al., 2015). However, it is currently unclear which skills are needed and which ones need to be learned to increase a device's benefits. Moreover, due to significant loss in upper extremity functionality and increasing fatigue due to the progressive nature of some NMD, handling the device can become increasingly difficult over time (Rahman et al., 2007; Shank et al., 2017; van der Heide & de Witte, 2016). The perceived benefits, which vary between users, can even decrease so much over time that the user decides to stop using the device completely. Similarly, a recent systematic review on the short-term benefits of wearable devices found that as the disability level changes the device benefits change as well (Gandolla et al., 2020). To prevent discontinuation, researchers and developers are promoting intuitive and adaptable DAS that counteract pathological changes due to disease progression (Kooren et al., 2015; van der Heide et al., 2014). However, such developments require extensive insights into a user's ability to adapt motor capacity, capability, and performance over time. Activity monitoring through wearable sensors might provide a solution to acquire evidence over such long periods and some recent advances have been made in this field (Bendixen, 2019; Pedrocchi, 2019; van der Geest et al., 2019).

4.3 Relations across framework cells

Evidence from experts suggests that the ICF model and contextual constructs are considered during the design process and the formulation of device requirements, however, this is not reflected in the literature findings. It is commonly assumed that a DAS primarily affects body functions, which influences performance in the activity and participation. However, most literature focused mainly on the technical and design

requirements necessary to overcome body function impairments, mostly neglecting the relationship between motor ability and ADL performance. Only one study directly investigated the relation between joint mobility and the ability to perform common ADLs, concluding that improvements in joint mobility alone does not directly translate to changes in ADL performance in a home environment (van der Heide et al., 2017). For instance, increased arm elevation with a device from 26.4° to 67.1° did not result in the ability to comb one's hair, while peers could execute the same task at an elevation angle of 44.6° with and without a device. Furthermore, Heide et al. also indicated that environmental and personal factors, such as adjustments in the home setting and compensatory movements, have an important influence on ADL performance in multiple studies (van der Heide & de Witte, 2016; van der Heide et al., 2015; van der Heide et al., 2017). These factors could affect the relationship between changes in joint mobility and the ability to complete tasks. As a result, we propose that future research considers the device requirements of multiple ICF model components and contextual constructs, i.e. motor ability, capability and performance, within the same study design.

4.4 Unaddressed framework cells

Our mixed-methods approach revealed that literature focuses on selected framework cells which are often considered separately, thus limiting the evidence on the interaction between cells. For instance, current evidence shows that muscle activity and joint mobility are both influenced by load reduction (Dunning et al., 2016; Essers et al., 2013; Estilow et al., 2018; Haumont et al., 2011; Kooren et al., 2015; van der Heide et al., 2017). However, as also pointed out by experts, the interaction between 1) muscle activity and joint mobility and 2) how this is affected by disease and 3) how this could be restored by the device should be investigated. Stabilizing and facilitating the shoulder girdle requires relatively complex muscle coordination, which is affected in

people with NMD (Bergsma et al., 2014; de Baets et al., 2014; Essers et al., 2019). Experts believe that insights into how muscle coordination is affected would benefit the development of a universal DAS and to optimize a device to fit the individual requirements (Trigili et al., 2019; van der Heide et al., 2014). Other symptoms, such as stiffness, pain, and early fatigue, were also regarded as important factors by experts and were present in the literature findings, however these topics were not clearly addressed in the study designs. Two studies investigated comfort levels with limited evidence on stiffness, pain, or fatigue (Kooren et al., 2015; Lebrasseur et al., 2019) and were therefore not represented in the respective cells. In contrast, while pain and stiffness are highly prevalent and should be reduced to comfortable levels, they might not be the limiting factors for ADL performance in people with NMD (Bergsma, Janssen, et al., 2017). Bergsma et al. 2017, found that participants who had high pain and stiffness levels also reported relatively few activity limitations, which indicates an overuse in their body functions. However, it is unclear whether a DAS positively relieves these symptoms and how effects differ across motor capacity, capability, and performance. Therefore, future research should consider the importance of these unaddressed cells for device development as possible influencers or as main device requirements.

4.5 Research recommendations

From our analysis it is clear that integration and inclusion of ICF components and contextual constructs are needed to bridge the knowledge gaps in the development and evaluation of DAS. To realize these two tasks, we propose four steps with each a focus point, examples from our analysis, and suggestions for the design of future studies.

First, we propose that future research incorporates multiple ICF components and contextual constructs within one study design. It is commonly assumed that body functions are primarily affected by a DAS, which shapes the

effects on activity and participation. Therefore, we suggest to focus on the relation between these two components before proceeding to examine the effect of environmental and personal factors. Our analysis shows that muscle activity and joint mobility are affected by load reduction, but their relation has not been investigated nor linked to ADL performance. Gandolla et al. deduced similar conclusions from their focus on activity and participation (Gandolla et al., 2020). Therefore, we recommend to investigate the relation between the two ICF components in a biomechanical framework under various levels of load reduction during functional tasks to optimize effort reduction and mobility improvements during common ADLs.

Secondly, the influence of environmental and personal factors should be investigated when deploying a device. Previously, barriers within these factors have been ascribed to personal preferences, such as performing ADLs without support or conserving energy altogether, and home setting, such as limited space or a fixed location of the device (van der Heide & de Witte, 2016).

Thirdly, research should include short term, such as within- and between-day repeatability, and longitudinal measurements, such as yearly follow ups, to monitor adaptations over time. For example, it is expected that a DAS delays fatigue onset and reduces fatigue, but it is unclear if and how this affects ADL performance throughout the day. From two ongoing studies and previous literature we consider activity monitoring a method to quantify device utilization and a proxy for motor performance (Bendixen, 2019; Koene et al., 2016; Pedrocchi, 2019; van der Geest et al., 2019). In addition, characteristics of ADLs, such as a diminished frequency and variety, can be used to monitor adaptations in activity and participation (van der Geest et al., 2019). Furthermore, we propose to include muscle strength and joint mobility measures to monitor disease progression (Han et al., 2015; Janssen, Harlaar, et al., 2019). A

longitudinal cohort study should investigate the relationship between disease progression and adaptations in daily activity of people with NMD over the course of a year. Disease progression and daily activity could be sampled every few months. Daily activity should then be averaged over the timespan of several days to include a range of ADL. The relationship could be expressed as a correlation between disease progression factors, muscle strength and joint mobility, and device utilization, and motor performance.

Lastly, experts and literature agree that user satisfaction, such as perceived benefits and comfort, should be taken as guidance to evaluate the device effectiveness. Gandolla et al. also recently promoted the use of objective and subjective measures as both measures provide equally important evidence of the functional status of the user (Gandolla et al., 2020). Therefore, device requirements should align with the needs and goals of the user and additionally aim to relief symptoms of pain, stiffness, and fatigue. Furthermore, future research should validate and incorporate subjective measures related to the respective ICF model components and contextual constructs. A possible longitudinal study could include a questionnaire that links symptoms of pain, stiffness, and fatigue in people with NMD to the motor capability and capacity to perform ADL w/o a DAS and perceived benefits over the course of a year.

4.6 Recommendations for developing evaluation tools

When the above evidence is combined, it will provide the basis for understanding how standardized device benefits result in daily device utilization accounting for changes in disease progression and users' needs and goals over time. Evaluation tools developed along these insights should be standardized and validated with focus on international consensus, as indicated by recent research (Janssen, Lobo-Prat, et al., 2019). We

suggest several minimal requirements for the development of such tools. First, the tools require an integration of translatable cells that cover at least two of the contextual constructs. Second, the tools should be applicable alongside the development and after deployment of DAS. Third, subjective measures, such as perceived benefits and comfort, should be included in a device's evaluation of effectiveness near the product final development stages.

5. Conclusion

Three knowledge gaps were identified and given synthesized research recommendations based on the integration and inclusion of ICF model components and contextual constructs. First, adaptations due to altered support requirements over time are poorly understood. Second, relations between ICF model components and contextual differences are limited. Finally, several framework cells, such as comfort levels, were brought to our attention by experts that were not covered sufficiently in scientific literature. We promote the use of multiple ICF model components and contextual constructs within research to benefit the development of DAS. Research should quantify device benefits and daily device utilization with respect to disease progression and users' needs and goals over time. Furthermore, we suggest several minimal requirements for the development of evaluation tools of DAS. The tools are required to cover multiple framework cells and to be applicable in various environments, for various users, and on multiple time points. Moreover, the tools should integrate objective and subjective measures to evaluate device effectiveness.

6. Acknowledgements

This work was supported by the Netherlands Organisation for Scientific Research (Den Haag, NL) under Grant: Symbionics Perspectief Program project 13523 ADAPT. The

authors would like to thank the focus groups for their contributed expertise, and Evy Paulussen and Anna-Carolin Wijnands for their contributions in data collection.

7. Declaration of interests

None

8. Appendix

8.1. Literature search terms, criteria, and strings.

	Search Terms	Inclusion criteria	Exclusion criteria
Population	Neuromuscular Diseases Neuromuscular Manifestations Muscular Dystrophies Musculoskeletal Diseases Disabled Persons	Neuromuscular Diseases Neuromuscular Manifestations Muscular Dystrophies Musculoskeletal Diseases Disabled Persons	Animals Cadaver
Impairment	-	-	Amputees Fractures, Bone Cognition Disorders Muscle-Tendon problems
Region	Upper Extremity Shoulder Arm Elbow Scapula	Upper Extremity Shoulder Arm Elbow Scapula	Lower Extremity Spine Hand Head Psychological*
Intervention	Self-Help Devices Orthotic Devices Gravity Compensation Exoskeleton* Robot* Support Devices Arm Support Arm Assistance	Self-Help Devices Orthotic Devices Gravity Compensation Exoskeleton* Robot* Support Devices Arm Support Arm Assistance	Medicine General Surgery Electrical Stimulation Drug Therapy Transcranial Magnetic Stimulation Magnetic Field Therapy Virtual Reality Exposure Therapy
Evaluation	Investigative Techniques (PubMed only)	Investigative Techniques Activities of Daily Living Range of Motion, Articular Torque Reachable Workspace Joint Loading Muscle Activity Body Function Activity	-
Others	-	-	Publication Date: 2000 until 2020
Search string (PubMed)	(Neuromuscular Diseases[All Fields] OR Neuromuscular Manifestations[All Fields] OR Muscular Dystrophies[All Fields] OR Musculoskeletal Diseases[All Fields] OR Disabled Persons[All Fields]) AND (Upper Extremity[All Fields] OR Shoulder[All Fields] OR Arm[All Fields] OR Elbow[All Fields] OR Scapula[All Fields]) AND (Self-Help Devices[Title/Abstract] OR Orthotic Devices[Title/Abstract] OR Gravity Compensation[Title/Abstract] OR Exoskeleton*[Title/Abstract] OR Robot*[Title/Abstract] OR Support Devices[Title/Abstract] OR Arm Support[Title/Abstract] OR Arm Assistance[Title/Abstract]) AND (Investigative Techniques[All Fields]) AND ("2000/01/01"[PDAT] : "2019/12/31"[PDAT])		
Search string (Web of Science)	ALL=(Neuromuscular Diseases OR Neuromuscular Manifestations OR Muscular Dystrophies OR Musculoskeletal Diseases OR Disabled Persons) AND ALL=(Upper Extremity OR Shoulder OR Arm OR Elbow OR Scapula) AND TS=(Self-Help Devices OR Orthotic Devices OR Gravity Compensation OR Exoskeleton* OR Robot* OR Support Devices OR Arm Support OR Arm Assistance) Timespan=2000-2019		

Table 4. Literature search terms, criteria, and strings.

8.2. Interview questions.

- (1) Describe your relation to the population?
- (2) What are the most common problems within the population?
- (3) How are these problems solved by the currently available dynamic arm supports?
- (4) Which problems are not solved by the currently available dynamic arm supports?
- (5) Describe your relation to the respective research field?
- (6) Give a short description of the previous/current research projects?
- (7) What are the current unanswered research questions that are not being investigated?
- (8) What are your interests regarding using dynamic arm supports?
- (9) How would you regard (in)effective device usage? Please describe a respective measurement unit?
- (10) What would a user's interests of use be? Considered over which time period: hour, (partial) day, week, month, year?
- (11) What are the minimum requirements of dynamic arm supports? What are currently available dynamic arm supports capable of and what are opportunities for improvement?
- (12) What are the current gaps in knowledge for further dynamic arm support

development? How would you approach these gaps to generate knowledge?

Combine this question with #13.

(13) How familiar are you with the following examples of evaluation techniques?

a. Questionnaires and scoring tests:

- i. MAS: Modified Ashworth scale
- ii. FMA: Fugl-Meyer Assessment
- iii. ARAT: Action Research Arm Test
- iv. Jebsen test of hand function
- v. WMFT: Wolf Motor Function Test
- vi. MAL: Motor Activity Log
- vii. SIS: Stroke Impact Scale
- viii. ABILHAND
- ix. ULFI: Upper-Limb Functional Index
- x. PUL: Performance of the upper limb scale
- xi. RMA: Rivermead arm score
- xii. Barthel index

b. Biomechanical tests:

- i. Muscle activation (electromyography); amplitude, timing, synergy, fatigue.
 - ii. Kinematics; workspace, Range of Motion, tracking error, movement accuracy, joint angles.
 - iii. Kinetics; torque, force-interaction.
- c. Activity monitoring:
 - i. Activity sensors; compliance, (in)active, area under curve, peak, bouts.
 - ii. Diaries; usage duration, type of activities.

(14) Other remarks?

8.4. Expert views.

ICF model	Impairments & Limitations	Effectiveness & Requirements	Previous projects & Remaining questions	Research priorities
Body functions	<p><u>Common:</u> Difficulty with performing motions against gravity; shoulder pain; muscle weakness/ pain/ fatigue, coordination; inactivity from muscle groups or other detrimental effects due to compensational strategies; joint rigidity; hand function</p> <p><u>Solved:</u> Certain level of range of motion; shoulder pain; muscle fatigue and coordination; posture</p> <p><u>Unsolved:</u> Larger range of motion; hand function; muscle weakness/ pain/ fatigue (required residual forces for device usage); finer coordination; integration with wrist support; joint rigidity; DAS induced inconvenience; limited shoulder motion/ stabilization; pronation/ supination</p>	<p><u>Effective:</u> device usage is intuitive; it supports and motivates usage of affected side; effectiveness is mostly subjective to user</p> <p><u>Ineffective:</u> DAS has limited range of motion; it requires additional energy; disease and device utilization shows progression; not used as intended according to specifications; user has compensation strategies; ineffectiveness is mostly subjective to user</p> <p><u>Minimum requirement:</u> DAS has controlled interaction from joints (shoulder, elbow, wrist driven control); promotes normal posture, natural motion, and range of motion especially above shoulder/ head level; provides adaptive support; usable in training; frictionless; usage is intuitive; provides shoulder support; decreases fatigue</p>	<p><u>Previous projects:</u> Influence of gravity compensation; basic demands for ADL tasks (gravity compensation, range of motion); arm positioning/ balancing; biomechanical interaction with support; force transposition from DAS to user; head support; arm brace for shoulder stabilization; improvement for proprioceptive feedback</p> <p><u>Remaining questions:</u> Supporting posture; interaction disease-device; kinematic profile (abilities, limitations); fatigue/ pain/ residual strength; required residual force/ strength; compensation support and muscle rigidity; feedback regarding shoulder motion</p>	<p><u>Interests:</u> Biomechanics of usage (interaction forces, utilization of range of motion, feedback regarding shoulder motion as therapy tool); interaction ambulant user-device; fatigue</p> <p><u>Device user's interests:</u> Upper extremity performance (RoM/ motion progression, frequency of usage, speed/effort, training, time/ effort); how to prevent inactivity/ fatigue</p> <p><u>Gaps:</u> Biomechanical interaction user-device (forces); required residual strength capacity; supporting natural posture/ motion; accurate description/ indication disease progression; need for short-term measurements; adequate tests for pain/ stiffness/ fatigue/ functionality in combination with DAS; cross-sectional tests are not representative, need for longitudinal tests</p>
Activity	<p><u>Common:</u> Limitations in repetitions; independence/ self-care (eat/ drink, writing); interaction with environment; DAS acceptance (embarrassment); computer work</p> <p><u>Solved:</u> Certain level of dependency/ self-care (eat/drink, computer work)</p>	<p><u>Effective:</u> DAS changes users' inability to ability or improves a realistic user need (repetitions); completing tasks within acceptable range of motion at comfortable level</p> <p><u>Ineffective:</u> Mostly subjective to user</p>	<p><u>Previous projects:</u> Role of DAS in independency/ self-care; motion intention detection; obscure interaction with environment; evaluation of ADL support; motion freedom w/o DAS</p>	<p><u>Interests:</u> What are individuals' subjective needs or goals; device usage duration/ frequency; area map of gravity compensation linked to range of motion; identification and performance evaluation of self-care activities at home (eating/ drinking, computer/ tablet usage)</p>

Table 5. Expert views as tabulated keywords from the collective focus group interviews. DAS: Dynamic Arm Support, w/o: with and without.

ICF model	Impairments & Limitations	Effectiveness & Requirements	Previous projects & Remaining questions	Research priorities
Activity	<p><u>Unsolved:</u> Activities outside range of motion; interaction with environment; natural motion hand to mouth after elevation (eating/ drinking, pouring a drink); writing; easy don/ doff; limited activities supported; problems with computer work still exist; other types of DAS are sometimes better (adjusted spoon), perhaps a combination is necessary</p>	<p><u>Minimum requirements:</u> Mostly subjective to user; DAS should support independent self-care; improvement of ADL; allow control of wheelchair; enable writing; motivate users</p>	<p><u>Remaining questions:</u> What are the kinematic profiles (activities and limitations) w/o DAS; how to increase the set of supporting activities</p>	<p><u>Device user's interests:</u> Recognizing non-usage; types of activities (can/not and w/o DAS)/ activity profile; user-device/needs-specs match; distribution of energy over activities/ motivation; activity monitoring with moderated feedback (users indicate they are aware of their activities/ energy)</p> <p><u>Gaps:</u> Recognizing use/ non-use; activities/ functional capacity w/o DAS in home setting with respect to ADL needs; description of active population; environmental factors</p>
Participation	<p><u>Common:</u> Dependency on caregiver; trying to blend in; reaching individual goals; inactivity due to body function problems; age and disease dependent; eating at a restaurant</p> <p><u>Solved:</u> Certain level of dependency/ self-care (eat/ drink at restaurant); reaching out hand for contact</p> <p><u>Unsolved:</u> Integration with wheelchair; trying to blend in/ embarrassment; limited social contact due to caregiver dependency and hand contact; DAS not accessible/ transportable</p>	<p><u>Effective:</u> Mostly subjective to user needs and social context; if a user becomes more independent</p> <p><u>Ineffective:</u> Mostly subjective to user needs and social context; progression of usage; environmental factors limiting usage</p> <p><u>Minimum requirements:</u> DAS is transportable; allows interaction with environment; double-sided DAS to stimulate both arms</p>	<p><u>Previous projects:</u> Inquiring user needs; ADL and home usage; Quality of Life; user-DAS match procedure/ evaluation (does it work and can it be optimized?)</p> <p><u>Remaining questions:</u> Relation between device usage and psychomotor factors (motivation, mood, good/bad motion profiles, energy costs); environmental factors (home setting and outdoors) and personal factors; aesthetics/ embarrassment</p>	<p><u>Interests:</u> device usage in home settings; DAS in ambulant users as body mounted</p> <p><u>Device user's interests:</u> Psychological drives; social interaction; experiencing normal puberty; barriers of confrontation</p> <p><u>Gaps:</u> What is important/ positive for users; individual wishes and possibilities; quality of performance at home</p>

Table 5 (continued). Expert views as tabulated keywords from the collective focus group interviews. DAS: Dynamic Arm Support, w/o: with and without.

ICF model	Impairments & Limitations	Effectiveness & Requirements	Previous projects & Remaining questions	Research priorities
Other	<p><u>Common:</u> Users are very intuitive with compensational strategies and masking problems</p> <p><u>Solved:</u> Mounted to wheelchair provides constant access</p> <p><u>Unsolved:</u> Aesthetics/ hardware/ size/ transportation; DAS not adaptive to disease progression; limited knowledge from user/ therapist on DAS; costs/ repair; user-device mismatch due to shortcomings in selection procedure/ disease progression; variation in user capabilities/ no universal method</p>	<p><u>Effective:</u> DAS wear/utilization (actual usage); acceptance; user motivation</p> <p><u>Ineffective:</u> DAS prototypes that do not reach the market</p> <p><u>Minimum requirements:</u> Mechanistic balance; application to monitor user-device match; smaller size; smaller increments/ adjustments for gravity compensation; adaptive support; proper manual/ guidance device usage</p>	<p><u>Previous projects:</u> General DAS information (also for insurance companies); involvement of young users</p> <p><u>Remaining questions:</u> Fine-tuning prototypes with simulation modeling; transposing applied force to user; reducing internal friction/ required force from user; terminology of all DAS related items; aspects of user-device match (individual assessments; differences between DAS)</p>	<p><u>Interests:</u> Time DAS motor is active; commands send to readjust motors; integration of devices and other DAS; aesthetics; use/ disuse and affected/ unaffected side;</p> <p><u>Device user's interests:</u> Mostly subjective; allocate time to develop DAS acceptance in selection procedure</p> <p><u>Gaps:</u> Problem cases; intensity of device utilization; DAS as measuring tool; matching wishes/ needs users; limited knowledge from care providers about DAS; structured method to decide which DAS and when; easy yet universal device for users and therapists; limited choice in DAS (insurance/ costs); test period at home; communication between professionals should improve to find the best user-device match</p> <p><u>Other:</u> Psychomotor and complexity of advice procedure should be taken into account; need to know actual usage regarding ADL; what are other obscurities; user-device adaptation; therapy to improve usage; website with all DAS information; adequate tests to indicate need for DAS; more research for practical things (selection procedure, costs, aesthetics); rehabilitation can observe ADL but progress is training dependent</p>

Table 5 (continued). Expert views as tabulated keywords from the collective focus group interviews. DAS: Dynamic Arm Support, w/o: with and without.

9. References

- Armon Products. Retrieved February 2020 from <https://armonproducts.com>
- Bendixen, R. M. (2019). *Use of Dynamic Arm Support Devices for Upper Limb Function in Non-Ambulatory Men With Duchenne Muscular Dystrophy (DMD)*. <https://clinicaltrials.gov/ct2/show/study/NCT03531788>
- Bergsma, A., Cup, E. H., Janssen, M. M. H. P., Geurts, A. C., & de Groot, I. J. M. (2017, Feb). Upper limb function and activity in people with facioscapulohumeral muscular dystrophy: a web-based survey. *Disabil Rehabil*, 39(3), 236-243. <https://doi.org/10.3109/09638288.2016.1140834>
- Bergsma, A., Janssen, M. M. H. P., H., G. A. C., Cup, E. H., & de Groot, I. J. M. (2017). Different profiles of upper limb function in four types of neuromuscular disorders. *Neuromuscul Disord*, 27(12), 1115-1122.
- Bergsma, A., Murgia, A., Cup, E. H., Verstegen, P. P., Meijer, K., & de Groot, I. J. (2014, Sep). Upper extremity kinematics and muscle activation patterns in subjects with facioscapulohumeral dystrophy. *Arch Phys Med Rehabil*, 95(9), 1731-1741. <https://doi.org/10.1016/j.apmr.2014.03.033>
- Coscia, M., Cheung, V. C. K., Tropea, P., Koenig, A., Monaco, V., Bennis, C., Micera, S., & Bonato, P. (2014). The effect of arm weight support on upper limb muscle synergies during reaching movements. *J Neuroeng Rehabil*.
- de Baets, L., Jaspers, E., Janssens, L., & van Deun, S. (2014). Characteristics of neuromuscular control of the scapula after stroke a first exploration. *Front Hum Neurosci*.
- Deenen, J. C., Horlings, C. G., Verschuuren, J. J., Verbeek, A. L., & van Engelen, B. G. (2015, Jul). The epidemiology of neuromuscular disorders: a comprehensive overview of the literature. *J Neuromuscul Dis*, 73-85.
- Deenen, J. C., van Doorn, P. A., Faber, C. G., van der Kooi, A. J., Kuks, J. B., Notermans, N. C., Visser, L. H., Horlings, C. G., Verschuuren, J. J., Verbeek, A. L., & van Engelen, B. G. (2016, Jul). The epidemiology of neuromuscular disorders: Age at onset and gender in the Netherlands. *Neuromuscul Disord*, 26(7), 447-452. <https://doi.org/10.1016/j.nmd.2016.04.011>
- Dunning, A. G., Janssen, M. M. H. P., Kooren, P. N., & Herder, J. L. (2016). Evaluation of an Arm Support With Trunk Motion Capability. *J Med Device*.

- Ellis, M. D., Sukal, T., DeMott, T., & Dewald, J. P. A. (2008). Augmenting Clinical Evaluation of Hemiparetic Arm Movement With a Laboratory-Based Quantitative Measurement of Kinematics as a Function of Limb Loading. *Neurorehabil Neural Repair*.
- Essers, J., Meijer, K., Murgia, A., Bergsma, A., & Verstegen, P. (2013, Jun). An inverse dynamic analysis on the influence of upper limb gravity compensation during reaching. *IEEE Int Conf Rehabil Robot*, 2013, 6650368. <https://doi.org/10.1109/icorr.2013.6650368>
- Essers, J. M. N., Peters, A., Meijer, K., Peters, K., & Murgia, A. (2019, Jul 10). Superficial shoulder muscle synergy analysis in Facioscapulohumeral Dystrophy during humeral elevation tasks. *IEEE Trans Neural Syst Rehabil Eng*. <https://doi.org/10.1109/tnsre.2019.2927765>
- Estilow, T., Glanzman, A. M., Powers, K., Moll, A., Flickinger, J., Medne, L., Tennekoon, G., & Yum, S. W. (2018). Use of the Wilmington Robotic Exoskeleton to Improve Upper Extremity Function in Patients With Duchenne Muscular Dystrophy. *Am J Occup Ther*.
- Flextension*. Retrieved February 2020 from <https://flextension.nl>
- Focal Meditech B.V.* Retrieved February 2020 from <https://focalmeditech.nl>
- Gandolla, M., Antonietti, A., Longatelli, V., & Pedrocchi, A. (2020). The Effectiveness of Wearable Upper Limb Assistive Devices in Degenerative Neuromuscular Diseases: A Systematic Review and Meta-Analysis. *Front Bioeng Biotechnol*.
- Han, J. J., De Bie, E., Nicorici, A., Abresch, R. T., Bajcsy, R., & Kurillo, G. (2015, Dec). Reachable workspace reflects dynamometer-measured upper extremity strength in facioscapulohumeral muscular dystrophy. *Muscle Nerve*, 52(6), 948-955. <https://doi.org/10.1002/mus.24651>
- Haumont, T., Rahman, T., Sample, W., King, M. M., Church, C., Henley, J., & Jayakumar, S. (2011). Wilmington Robotic Exoskeleton: A Novel Device to Maintain Arm Improvement in Muscular Disease. *J Pediatr Orthop*.
- Heutinck, L., Jansen, M., van den Elzen, Y., van der Pijl, D., & de Groot, I. J. M. (2018, 2018-Jul-06). Virtual Reality Computer Gaming with Dynamic Arm Support in Boys with Duchenne Muscular Dystrophy. *J Neuromuscul Dis*. <https://doi.org/10.3233/jnd-180307>

- Holsbeeke, L., Ketelaar, M., Schoemaker, M. M., & Gorter, J. W. (2009). Capacity, Capability, and Performance: Different Constructs or Three of a Kind? *Arch Phys Med Rehabil*.
- JAECO Orthopedic. Retrieved February 2020 from <https://jaecoorthopedic.com>
- Jansen, M., Burgers, J., Jannink, M., van Alfen, N., & De Groot, I. J. M. (2015). Upper Limb Training with Dynamic Arm Support in Boys with Duchenne Muscular Dystrophy: A Feasibility Study. *Int J Phys Med Rehabil*.
- Janssen, M. M. H. P., Bergsma, A., Geurts, A. C. H., & de Groot, I. J. M. (2014). Patterns of decline in upper limb function of boys and men with DMD: an international survey. *J Neurol*.
- Janssen, M. M. H. P., Harlaar, J., Koopman, B., & De Groot, I. J. M. (2019). Unraveling upper extremity performance in Duchenne muscular dystrophy: A biophysical model. *Neuromuscul Disord*.
- Janssen, M. M. H. P., Lobo-Prat, J., Bergsma, A., Vroom, E., & report, W. P. L. a. t. e. o. t. (2019). 2nd Workshop on Upper-Extremity Assistive Technology for People with Duchenne: Effectiveness and Usability of Arm Supports. *Neuromuscul Disord*.
- Jutai, J. W., Fuhrer, M. J., Demers, L., Scherer, M. J., & DeRuyter, F. (2005). Toward a Taxonomy of Assistive Technology Device Outcomes. *Am J Phys Med Rehabil*.
- Koene, S., Dirks, I., van Mierlo, E., de Vries, P. R., Janssen, A. J. W. M., Smeitink, J. A. M., Bergsma, A., Essers, J. M. N., Meijer, K., & de Groot, I. J. M. (2016). Domains of Daily Physical Activity in Children with Mitochondrial Disease: A 3D Accelerometry Approach. *JIMD Rep*.
- Kooren, P. N., Dunning, A. G., Janssen, M. M. H. P., Lobo-Prat, J., Koopman, B., Paalman, M., De Groot, I. J. M., & Herder, J. L. (2015). Design and pilot validation of A-gear: a novel wearable dynamic arm support. *J Neuroeng Rehabil*.
- Lebrasseur, A., Lettre, J., Routhier, F., Bouffard, J., Archambault, P. S., & Campeau-Lecours, A. (2019). Evaluation of the usability of an actively actuated arm support. *Assist Technol*.
- Pedrocchi, A. (2019). *USEFUL: User-centred Assistive SystEm for Arm Functions in neUromuscuLar Subjects*. <https://clinicaltrials.gov/ct2/show/NCT03127241>

- Prange, G. B., Jannink, M. J. A., Stienen, A. H. A., van der Kooij, H., IJzerman, M. J., & Hermens, H. J. (2009). Influence of Gravity Compensation on Muscle Activation Patterns During Different Temporal Phases of Arm Movements of Stroke Patients. *Neurorehabil Neural Repair*.
- Rahman, T., Sample, W., Seliktar, R., Scavina, M. T., Clark, A. L., Moran, K., & Alexander, M. A. (2007). Design and Testing of a Functional Arm Orthosis in Patients With Neuromuscular Diseases. *IEEE Trans Neural Syst Rehabil Eng*.
- Shank, T. M., Wee, J., Ty, J., & Rahman, T. (2017). *Quantitative Measures with WREX Usage* IEEE Int Conf Rehabil Robot, London.
- Trigili, E., Grazi, L., Crea, S., Accogli, A., Carpaneto, J., Micera, S., Vitiello, N., & Panarese, A. (2019). Detection of movement onset using EMG signals for upper-limb exoskeletons in reaching tasks. *J Neuroeng Rehabil*.
- van der Geest, A., Essers, J. M. N., Bergsma, A., Jansen, M., & de Groot, I. J. M. (2019). Monitoring daily physical activity of upper extremity in young and adolescent boys with Duchenne muscular dystrophy: A pilot study. *Muscle and Nerve*.
- van der Heide, L., & de Witte, L. (2016). The perceived functional benefit of dynamic arm supports in daily life. *J Rehabil Res Dev*.
- van der Heide, L., Gelderblom, G., & de Witte, L. (2015). Effects and Effectiveness of Dynamic Arm Supports. *Am J Phys Med Rehabil*.
- van der Heide, L., Ramakers, I., Essers, J. M. N., Gelderblom, G., De Witte, L., & Meijer, K. (2017). Is it possible to assess the effects of dynamic arm supports on upper extremity range of motion during activities of daily living in the domestic setting using a portable motion capturing device? – A pilot study. *Technol Disabil*, 29, 91-99.
- van der Heide, L., van Nijhuijs, B., Bergsma, A., Gelderblom, G., van der Pijl, D., & de Witte, L. (2014, Aug). An overview and categorization of dynamic arm supports for people with decreased arm function. *Prosthet Orthot Int*, 38(4), 287-302. <https://doi.org/10.1177/0309364613498538>
- World Health Organization. (2001). *International classification of functioning, disability and health : ICF*. World Health Organization. <https://apps.who.int/iris/handle/10665/42407> Retrieved February 2020 from