The trunk in neuromuscular disorders

A neglected part of the chain

DONDERS SERIES

Laura Peeters

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The research described in this thesis was supported by the Netherlands Organisation for Scientific Research (NWO), Duchenne Parent Project, stichting FSHD, Foundation to Eradicate Duchenne, Parent Project Muscular Dystrophy, Focal Meditech, BAAT Medical, Summit revalidatietechniek, Hankamp Rehab, Leavo and Yumen Bionics.

Printing this thesis was financially supported by the Radboud University, Donders Institute for Brain, Cognition and Behaviour, Focal Meditech BV, Hankamp Rehab and Summit Benelux BV.

ISBN 9789462841994

Cover image Codex Anatomy

Layout Laura Peeters

Print Ridderprint | www.ridderprint.nl

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Proefschrift

ter verkrijging van de graad van doctor

aan de Radboud Universiteit Nijmegen

op gezag van de rector magnificus prof. dr. J.H.J.M. van Krieken,

volgens besluit van het college van decanen

in het openbaar te verdedigen op woensdag 23 oktober 2019

om 14.30 uur precies

door

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geboren op 20 september 1989

te Venray

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“The whole is other than the sum of its parts.”
- Kurt Koffka

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CHAPTER 1

GENERAL INTRODUCTION

Abbreviations:

CP	Cerebral Palsy
DMD	Duchenne Muscular Dystrophy
GMFCS	Gross Motor Function Classification Scale
NMD	Neuromuscular disorders
SCI	Spinal Cord Injury
SMA	Spinal Muscular Atrophy
TCMS	Trunk Control Measurement Scale
UE	Upper Extremity

Unconsciously we move our trunk very often during the day when performing seated activities, for instance while reaching forward, writing a note, eating, looking over our shoulder when driving a car, et cetera. Can you imagine how this will change if we would not able to move our trunk and what the consequences would be for moving our arms?

This thesis focuses on trunk function and the interaction between trunk, upper extremity and head movements when performing seated daily tasks in patients with neuromuscular disorders. Before further introducing this topic, trunk anatomy and function in healthy people are discussed for a basic understanding.

THE TRUNK AS CENTRAL SEGMENT

The trunk (or torso) is the central segment of the body which connects the legs ("lower extremities (LE)"), arms ("upper extremities (UE)") and the head. Although it is often captured as one segment, it is a complex, multisegmented part of the body. The skeleton of the trunk consists of the ribcage and the spine, which is caudally connected to the iliae through the sacroiliac joints. The spine consists typically of 33 vertebrae that are divided into different regions: the cervical spine (7 vertebrae), thoracic spine (12 vertebrae), lumbar spine (5 vertebrae), sacrum (5 fused vertebrae) and the coccyx (4 fused vertebrae) (Figure 1A). The large number of joints provide the spine with substantial flexibility, which implies that the trunk cannot be seen as one, rigid segment. The cervical spine is also called the neck and rotations of its vertebrae considerably contribute to overall head movement [1]. Deep back muscles (e.g. rotators, multifidus, interspinales, and intertransversarii) are assumed to be involved in stabilizing the spine, whereas the superficial muscles (e.g. erector spinae, oblique abdominal muscles, and rectus abdominis) are mainly responsible for generating the large moments of force needed to hold and move the trunk against gravitational forces (Figure 1B). The origin of several back muscles can be found at the sacrum and pelvis, which emphasizes the close connection between the trunk and the pelvis.

Since the trunk is the central part of the body, it is indispensable when performing daily tasks. In a seated position, the trunk interacts with the UE as part of a kinematic chain and by providing a stable base. Involvement in the kinematic chain is most pronounced when reaching beyond arm length distance, as moving the trunk enlarges the workspace [2]. But trunk displacement is also observed when reaching within arm's length, or when performing other daily tasks than reaching [3]. In addition, the trunk adapts its posture to maintain stability during voluntary UE movements. UE movement results in displacing the body center of mass, thereby initiating a perturbation to posture and body balance [4]. As a result, in terms of

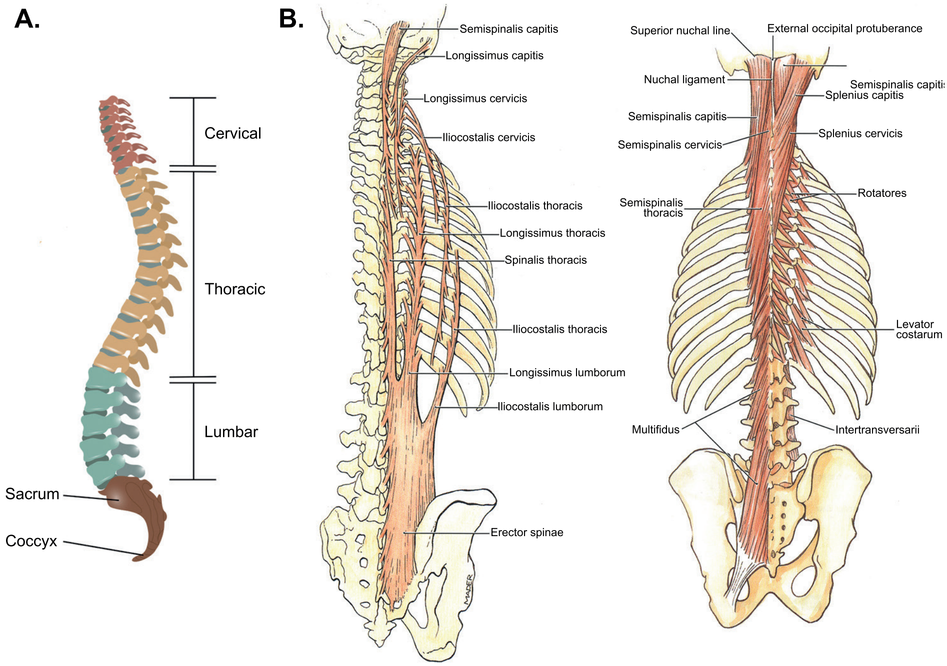

Figure 1 Illustrations of the trunk skeleton with the five different segments of the spine (A) and origin and insertion of several back muscles (B) (reprinted with permission of Wolters Kluwer Health, Inc. [6]).

stability, trunk control greatly determines the precision of UE movement [5]. Trunk movement and trunk stability also influence movement of the head and vice versa. Head orientation is generally kept constant in space for fixation of gaze on the target and for visual feedback of task performance [7]. This means that the head often shows a countermovement relative to the trunk movement.

TRUNK IMPAIRMENT

Trunk function is impaired in patients with a flaccid trunk. A flaccid trunk is typically associated with (severe) muscle weakness due to primary muscle disease (e.g. Duchenne muscular dystrophy (DMD)) or motor neuron disease (e.g. spinal muscular atrophy (SMA)) [8, 9], but can also be present in patients with central neurological disorders with bilateral paresis, like cerebral palsy or spinal cord injury [10]. Besides impairment of trunk function due to muscle weakness, (structural) deformities of the spine and/or rib cage can influence trunk function. When these disorders become symptomatic during (early) childhood and trunk function becomes impaired, two additional factors may negatively affect motor capacity. First, in typically developing children, interaction between the trunk and the UE develops with age due to maturation [7, 11]. Thus, when trunk function becomes impaired in early life, it may affect general motor development. Second, children are more prone than adults to develop spine deformities due to muscle weakness as long as the spine is growing. These deformities also affect trunk movement and stability [12]. Spinal deformities are often seen in patients with neuromuscular disorders (NMD), like SMA and DMD, and are related to factors such as type of SMA and age at loss of ambulation [13, 14].

As the trunk is indispensable for performing seated daily tasks, trunk impairment will likely result in changed interactions between trunk, UE and head movements when performing such tasks or even in an inability to perform certain tasks. However, knowledge of reduced motor capacity to perform seated activities is scarce, especially in patients with NMD (Chapter 2). Research has shown that UE function gradually decreases in patients with DMD and SMA over time [15, 16]. Therefore, trunk movement might gradually increase to compensate for reduced arm function when performing seated tasks. On the other hand, trunk impairment might also lead to an incapacity to maintain postural stability and control center-of-mass displacements when performing voluntary UE movements, leading to decreased task performance.

Since most of the patients with DMD, as well as patients with symptomatic SMA in early childhood, will not be able to walk once they have reached adulthood, it is of utmost importance to gain more insight in the interactions between trunk, UE and head movements when performing seated tasks. This knowledge is essential for developing dynamic assistive devices for the trunk and the head to support people with NMD in performing daily tasks. Clinically applicable dynamic devices for the trunk do not yet exist and are scares for the head. Development of a dynamic trunk and head assistive devices that could be integrated with an arm assistive device in the future was the key aim of the Symbionics project (Box 1).

The focus of this thesis is on trunk function in two types of NMD: DMD and SMA. These neuromuscular disorders are described below.

Duchenne muscular dystrophy

DMD is an x-linked, recessive neuromuscular disorder with an incidence of approximately 1 in 6000 live male births.[9] DMD is characterized by symmetrical, progressive muscle weakness, caused by the lack of the dystrophin protein in the muscle cells [17]. Proximal muscles are affected earlier than distal muscles (Figure 2a), resulting in a mean loss of ambulation around 11 years with the use of corticosteroids in The Netherlands [18]. UE function is already decreased in the early disease stages of DMD and overhead reaching is lost around the same age as ambulation is lost [15, 19]. The life expectancy is reduced to approximately 30 years on average [20]. Since there is no cure available, current treatment focuses mainly on slowing down disease progression (e.g. using corticosteroids [19]) and symptom management (e.g. physical therapy, nocturnal ventilation, cardiac medication [21]).

Box 1 Symbionics project

The studies performed in this thesis are part of the Symbionics program (project 2.1). The aim of this project was to develop dynamic support for trunk and head for persons with neuromuscular disorders to assist them when performing daily activities. To achieve this goal, several Dutch universities (i.e. University of Twente, Vrije Universiteit Amsterdam and Radboud university medical center) have worked closely together.

Both passive and active trunk and head supports were developed with the vision that these supports should be close to the body and should be as inconspicuous as possible. The passive supports assist to balance the trunk or head with the use of springs, so the user needs less muscle effort to move. The active supports provide additional assistance as muscle strength decreases further by the use of actuators and control strategies embedded with the spring-based mechanism.

The Radboud university medical center focused on the clinical perspective. Insight was gained on trunk and head function during seated arm tasks in the user groups to specify requirements for the supports. The Vrije Universiteit Amsterdam was responsible for design and development of prototypes and evaluation. Expertise of the University of Twente was on the control interfaces and control methods for the active supports.

SYMBIONICS

MANIPULATION • BODY SUPPORT • FORM ADAPTATION

Spinal muscular atrophy

SMA is a recessive neuromuscular disorder with an incidence of approximately 1 in 10000 [22]. SMA is characterized by muscle weakness (proximal more than distal, Figure 2b) and atrophy, caused by a low level of full-length, stable survival motor neuron 1 (SMN1) protein resulting in progressive degeneration of motor neurons in the spinal cord [23]. SMN2 protein copy number is the most important biomarker for disease severity; a higher copy number is related to less disease severity [23, 24]. Patients are categorized based on disease onset and maximum acquired milestones (e.g. achieving independent sitting or walking) [8]. Type 1 (infantile onset) is the most severe type and results in a median survival of less than 2 years. Children with SMA type 2 (onset at 7-18 months of age) will be able to sit independently, but they will never stand or walk independently. Children with SMA type 3 (onset from 18 months to adulthood) can achieve independent standing and walking, although many patients lose these abilities later on with disease progression. In SMA type 4 disease onset is during adulthood. However, the entire clinical spectrum is represented more by a gradual spectrum of functional capacities rather than by distinct subtypes [26]. The natural course is now changing due to effective treatment with Spinraza® (nusinersen) [27]. This drug is reimbursed by the Dutch government since the summer of 2018 for

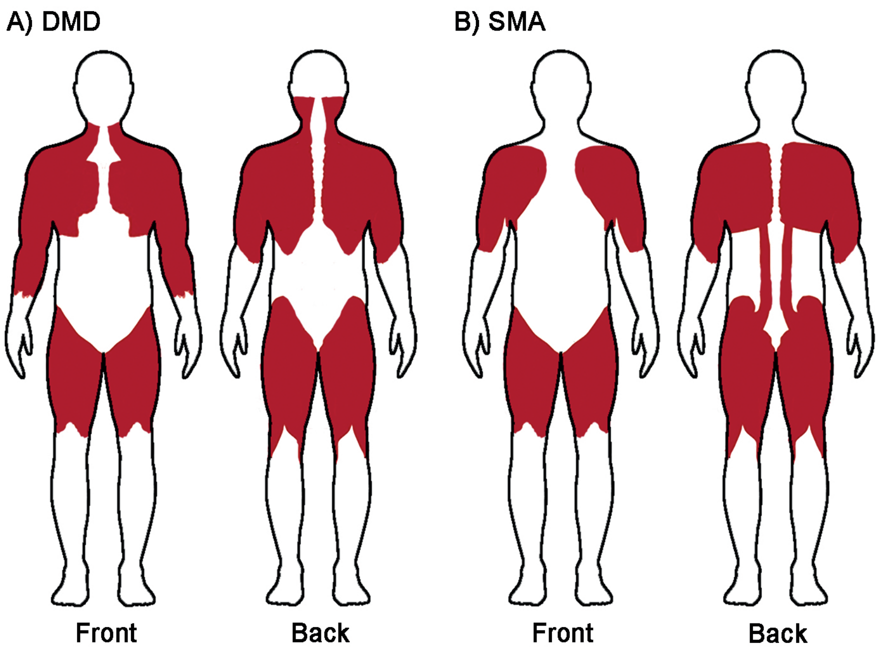

Figure 2 Characteristic muscle involvement patterns in patients with DMD (A) and SMA (B) (adapted from [25])

children below the age of 9.5 years. New natural history studies have to give insight in the long-term effects of the treatment.

AIM OF THIS THESIS

For the development and evaluation of assistive devices and other interventions for the trunk and UE in people with NMD, it is important to increase our knowledge of trunk function and the functional interactions between trunk, head and UE movements when performing seated daily tasks. Specifically, better insight in the commonalities and differences in trunk function among patients with different types of NMD, and between NMD patients and healthy controls is essential. Such knowledge provides clinicians and engineers with clues about which aspect of treatment (e.g. physical training, seating adjustments or assistive devices) can be generalized to all patients and where individual adaptations are needed.

The following research questions were formulated:

- What is known about the interactions between trunk, head and UE movements in patients with a flaccid trunk?
- How are the trunk and head involved in performing seated tasks with the UE in typically developing children and young adults; and more specifically, what is the role of specific trunk segments?
- How does trunk function differ between patients with DMD, patients with SMA, and healthy subjects during seated tasks with the UE?

MEASUREMENT METHODS

In this thesis, several outcome measures are useto answer the above questions, such as joint torque, joint range of motion, and muscle capacity. These outcome measures were determined with the use of muscle strength testing, movement analysis, and electromyography.

Muscle strength testing

Muscle strength testing is often used in the clinic to monitor disease progression. Muscle strength is clinically measured with the Medical Research Council (MRC) ordinal scale [28]. More precise measurement of muscle strength can be done with the use of a hand-held dynamometer (HHD) or with a static frame myometer. The HHD is less reliable amongst different examiners and for assessing large muscle groups, because it is influenced by the examiner's strength [29]. The static frame myometer can overcome this disadvantage, however this technique cannot easily be used in a clinical setting since it is not portable. In this thesis, muscle strength is measured with the static frame myometer, primarily as force (N). However, because the segment length has a great influence on the effective force that can be delivered, strength was adjusted for segment length and reported as joint torque (Nm).

Movement analysis

In a clinical setting, quantitative movement analysis is often performed to obtain insight in (abnormal) movement patterns for either diagnostic or evaluative purposes. Several systems are available, such as video cameras, accelerometers, or optical motion capture systems. In this thesis, an optical motion capture system is used with passive markers placed at anatomical landmarks. These markers are captured with infra-red cameras to determine 3D coordinates of all markers in space. At least three single markers are needed on each body segment to define its 3D orientation and, subsequently, to calculate joint angles from two body segment orientations [30, 31].

Electromyography

Electromyography is a widely applied technique for measuring electrical activity produced by the muscles during contraction [32]. For surface electromyography, sensors are placed at recommended locations above the target muscle [33]. Because sensors are placed on the skin, only activity of superficial muscles can be determined. Several outcome measures can be determined from the electrical signals, such as muscle activity onset and signal amplitude. In this thesis, we focus on the amplitude of the electrical muscle activity. Since the measured amplitude is dependent on sensor location and skin thickness, it can vary between persons [34, 35]. Therefore, we normalized the level of muscle activity level against the maximum amplitude determined during maximum voluntary isometric contraction, yielding muscle activity as percentage of the maximum muscle capacity of a specific muscle.

THESIS OUTLINE

In **chapter 2**, a literature review is presented addressing the current knowledge of trunk-UE and trunk-head interactions in patients with a flaccid trunk and in healthy children. This chapter clearly indicates the need for further research on this topic in patients with DMD and SMA.

Because knowledge of trunk motion is also scarce in healthy children and youngsters, especially with regard to movement of individual trunk segments, **chapter 3** provides insight in the interactions between trunk, UE and head movements in typically developing children when performing seated daily tasks. This chapter specifically focuses on movements of individual trunk segments.

Chapters 4 and 5 report studies on trunk function in boys/men with DMD (chapter 4) and people with SMA (chapter 5) when performing seated UE tasks. Trunk movement, muscle activity, and maximum joint torque are investigated and compared to healthy controls.

Chapter 6 summarizes and discusses the work described in this thesis. A comparison will be made between the different disorders described in the individual chapters to answer the last research question. Furthermore, it will elaborate on the clinical implications of the studies in this thesis and provide recommendations for future research.

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CHAPTER 2

CHANGES IN TRUNK MOVEMENT AND STABILITY WHEN PERFORMING SEATED ACTIVITIES IN NEUROLOGICAL PATIENTS WITH A FLACCID TRUNK – A REVIEW

L.H.C. Peeters, I.J.M de Groot, A.C.H. Geurts

Gait and Posture 2018; 62:46-55

Abbreviations:

CP	Cerebral Palsy
DMD	Duchenne Muscular Dystrophy
GMFCS	Gross Motor Function Classification Scale
NMD	Neuromuscular disorders
SCI	Spinal Cord Injury
SMA	Spinal Muscular Atrophy
TCMS	Trunk Control Measurement Scale
UE	Upper Extremity

ABSTRACT

Background: Trunk control is essential during seated activities. The trunk interacts with the upper extremities (UE) and head by being part of a kinematic chain and by providing a stable basis. When trunk control becomes impaired, it may have consequences for the execution of UE tasks.

Aim: To review trunk involvement in body movement and stability when performing seated activities and its relation with UE and head movements in neurological patients with a flaccid trunk, with a focus on childhood and development with age.

Methods and procedures: A search using PubMed was conducted and 32 out of 188 potentially eligible articles were included.

Outcomes and results: Patients with a flaccid trunk (e.g. with spinal cord injury or cerebral palsy) tend to involve the trunk earlier while reaching than healthy persons. Different balance strategies are observed in different types of patients, like using the contralateral arm as counterweight, eliminating degrees of freedom, or reducing movement speed.

Conclusions and implications: The key role of the trunk in performing activities should be kept in mind when developing interventions to improve seated task performance in neurological patients with a flaccid trunk.

INTRODUCTION

Control of upper body movement is essential when performing daily activities in a seated position. Trunk control is indispensable during seated activities, because it interacts with control of the upper extremities (UE) and the head by being part of a kinematic chain and by providing a stable base. In the kinematic chain of UE movement, trunk movement enlarges the workspace [1], but trunk displacement is also observed when reaching within arm length [2]. Voluntary UE movement will disturb posture, which is compensated for by postural reactions to maintain stability [3]. The trunk is involved in this postural chain when performing UE movements. Therefore, in terms of stability, trunk control greatly determines the precision of UE movement [4]. With regard to head movement, trunk movement enlarges the range of head motion in space. Lastly, trunk stability is essential for head balance as well as for accurate visual and vestibular control of posture and voluntary movements of (parts of) the body, such as the arm and hand [5, 6].

Trunk control is impaired in patients with a flaccid trunk, affecting their performance of daily activities. In addition, during their development, children with a flaccid trunk have a higher risk in developing scoliosis, which further complicates the interaction between the trunk and UE. A flaccid trunk is typically associated with (severe) muscle weakness due to primary muscle disease (e.g. Duchenne muscular dystrophy (DMD)) or motor neuron disease (e.g. spinal muscular atrophy (SMA)), but it may also be present in patients with central neurological disease with bilateral paresis [7, 8]. For instance, patients with 'high' spinal cord injury (SCI) (above thoracic level 6) may have spastic muscles below lesion level, particularly in their extremities, but often their trunk muscles lack normal (reticulospinal and vestibulospinal) control of postural tone mediated by the brainstem via the medially descending spinal tracts [9, 10]. As a result, these patients lack automatic trunk control which, in complete spinal cord lesions, cannot be compensated by the medial corticospinal descending neurons. Likewise, patients with severe (mostly bilateral) cerebral palsy (CP) may suffer from lack of postural tone as well as voluntary control of trunk muscles through lesions of their medially descending corticospinal and bulbospinal tracts [11].

When trunk control becomes impaired early in life, it may severely affect motor development in general and, through delayed and limited motor skills, even affect the cognitive and emotional development in children. Because many of the conditions mentioned above may become symptomatic during (early) childhood and because a substantial proportion of these children will not be able to walk once they have reached adulthood, studying the consequences of trunk impairments for the performance of seated UE activities is of utmost important. Undoubtedly, the interaction of the trunk with the UEs and the head will depend on the type and the stage or severity of the disease. In children with CP, the UEs are often spastic, ataxic or dyskinetic, whereas in DMD and SMA muscle weakness is most prominent, which may result in differently disturbed interactions with a flaccid trunk. SCI most often occurs in adults, but it may also be present in childhood due to e.g. trauma, neoplasma or infection. Depending on the lesion level, a flaccid trunk may coincide with normal UE function (high thoracic lesions) or impaired UE function (cervical lesions). Therefore, the interaction between trunk, UE and head movements may differ between diagnoses. The impact of a flaccid trunk is probably also dependent on age. First, the interaction between trunk, UEs and head changes with age due to maturation [12, 13]. Second, children are more prone than adults to develop spine deformities due to muscle weakness, which also affects their trunk movement and stability [14].

The goal of this review was to provide an overview of the changes in trunk movement and stability when performing UE activities in a seated position, and their relation with UE and head movements in neurological patients with a flaccid trunk compared to healthy subjects. A special focus will be given on childhood and development with age.

METHODS

PubMed was used as an electronic database to search for studies up to September 2016. Four search term categories were used in the search strategy: (1) population,(2) tasks, (3) body segments, and (4) outcomes (i.e. kinematics or stability). The key terms for each category were:

1. "muscular dystrophies", "spinal muscular atrophy", "Duchenne", "cerebral palsy", "spinal cord injuries", " spinal dysraphism", "spina bifida" or "healthy"
2. "reach", "reaching", "drinking", "activities of daily living", "ADL", "daily activity" or "pointing"
3. "upper body" or "arm" combined with either "trunk", "torso" or "head", "upper extremity" combined with either "trunk", "torso" or "head" or "trunk" combined with "head"
4. A. Kinematics: "movement", "motion", "kinematics", "motor skills" or "coordination"
   B. Stability: "postural balance", "balance", "stability", "postural control", "sway" or "postural adjustments"

The literature search was performed according to the PRISMA guidelines [15]. Studies were included when written in English. The articles were sorted in two phases. First, articles were screened by title and were included if the topic was potentially relevant.

Studies related to standing activities, gait, or therapy evaluations were excluded. Subsequently, the abstracts were read by the primary researcher (LP) and full articles were included when they met the following criteria: 1) covering the topic of task performance in a seated position, 2) involving both trunk and arm or head movement, and 3) presenting outcome measures related to kinematics (range of motion in three planes, movement trajectory, and/or spatiotemporal parameters such as movement velocity and timing of movement) or stability (center-of-mass/center-of-pressure displacement, trunk sway parameters, and/or force profiles). Relevant cited, yet unidentified, articles that met the inclusion criteria were included in second instance.

RESULTS

Search results

The literature search and article inclusion are shown in figure 1. Out of 188 potentially eligible articles, 32 articles were eventually included in this review. The study characteristics are shown in table 1.

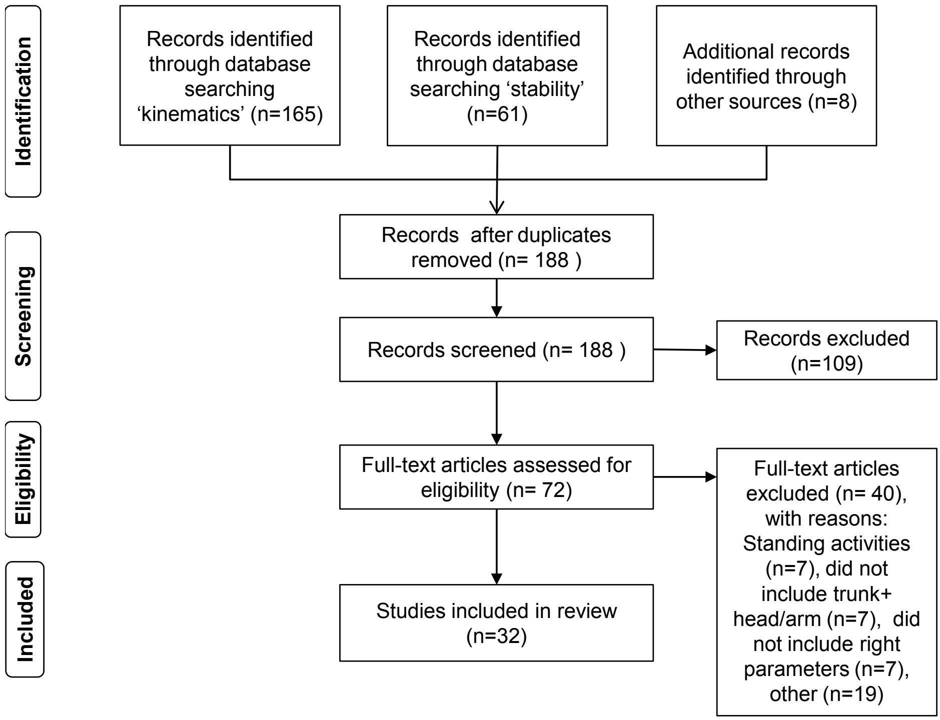

Figure 1 Flow chart of the literature search and included articles, based on PRISMA guidelines [15]

Table 1 Characteristics of included studies

Author	Year	Kinematics or stability	Trunk, head, arm	Population (no. subjects, age)	Supported/ unsupported sitting$^{\text{Foot support}}$	Equipment used	Tasks	Outcome measures
Aprile	2014	KIN	T+A	Healthy (n=6, 52-74y) Hemiparatic (n=6, 64-84y)	Supported?$^{+}$	3D optical motion tracking system	Drinking	Displacement, joint angles
Boswell-Ruyss	2009	STAB	T+A	SCI (SCI level C6-L2, n=30, 18-66y)	Unsupported^{+}	Sway meter	Upper body sway, maximal balance range, coordinated stability, reaching, t-shirt test	Body sway, duration of task, reach distance
Butler	2010	KIN	T+A	Spastic hemiplegic CP (n=2, 14-15y) Healthy (n=25, 5-18y)	Unsupported?$^{+}$	3D optical motion tracking system	Reach and grasp	Joint kinematics
Chen	2003	STAB	T+A	SCI (SCI level T3-T12, n=30, 33.97±10.7y)	Unsupported^{+}	Force plates	Static and dynamic sitting, functional tasks	sway, weight-shift and movement time
Cheng	2013	KIN	T+A	CP (MACS I-III, n=14, 7-17y)	Supported^{+}	Digital cameras, EMG	Writing	Joint angle, muscle activity
Choi	2004	KIN	T+A	Healthy (n=12)	Supported^{+}	Video camera, force transducer	Reaching	Boundaries between reach mode (arm-shoulder, arm-trunk, standing)
Coluccini	2007	KIN	T+H+A	Spastic hemiplegic CP (n=5, 11y) Dyskinetic movement disorders (n=5, 11.6y) Healthy (n=5, 11.0y) Healthy (n=5, 22.0y)	Unsupported?$^{?}$	3D optical motion analysis system	Picking blocks and transport it toward target position	Joint kinematics, task duration
Dean	1999	KIN + STAB	T+A	Healthy (n=6, 25.2±3.7y)	Unsupported^{+}	Videocamera, force plates, EMG	Reach and grasp, reach and press button	Segmental and joint angles, peak ground reaction forces, muscle activity

Field-Fote	2010	KIN + STAB	T+A	SCI (incomplete, n=10, 46.7 ± 5.8y) Healthy (n=10, 41.4 ± 13.6 y)	Unsupported+	3D optical motion tracking system	Maximal reaching	Limits of stability, center of pressure excursion (C7+wrist)
Flatters	2014	KIN + STAB	T+H+A	Healthy (n=61, 5-10y)	Supported+	Infrared cameras, balance board	Tracking, aiming and tracing with pen	Spatio-temporal accuracy, center-of-pressure, head movement
Heyrman	2013	STAB	T+A	hemiplegic, diplegic and quadriplegic CP (GMFCS I-IV, n=100, 8-15y)	Unsupported-	Trunk Control Measurement Scale	Static and dynamic sitting	Score for sitting balance on 15 items
Huang	2014	KIN	T+H+A	Bilateral spastic CP (GMFCS I-III, n=13, 10.1±2.1 y) Healthy (n=20, 9.0±1.6y)	Unsupported?+	Electromagnetic tracking system, force plate	Grasping and throwing Boccia balls	Maximal reach distance, joint kinematics, CoP displacement
Ju	2010	KIN + STAB	T+A	Spastic diplegic CP (GMFCS II-IV, n=8, 9.1±2.0y) Healthy (n=16, 9.5 ± 1.6y)	Unsupported+	3D optical motion tracking system, force plates	Reaching	Movement time, straightness ratio hand, peak velocity, number of movement units
Ju	2012	STAB	T+A	spastic diplegic CP (n=12, 9.0±2.1y) Healthy (n=17, 9.2y)	Unsupported+	Force plates	Reaching	COP displacement, sway, GRF
Kim	2010	KIN	T+A	SCI (SCI level T4-L4, n=10, 39.0±13.7 y) LBP (n=10, 45.9±12.2 y) Healthy (n=11, 27.5±10.8 y)	Supported+	3D optical motion tracking system, elektromagnetic system	Reaching	Joint angles (trunk)
Klotz	2014	KIN	T+A	Spastic CP (n=16, 9-17y) Healthy (n=17, 9-17y)	Supported??	3D optical motion tracking system	6 daily tasks	Joint angles

Table 1 Continued

Author	Year	Kinematics or stability	Trunk, head, arm	Population (no. subjects, age)	Supported/ unsupported sitting$^{\text{Foot support}}$	Equipment used	Tasks	Outcome measures
Kreulen	2007	KIN	T+A	Hemiplegic CP (n=10, 11-27 y) Healthy (n=10, 11-27 y)	Unsupported^{+}	Video cameras	Drinking, grasping and maximal range of motion forearm	Joint angles
Levin	2002	KIN	T+A	Healthy (n=11, 55.0±13.7 y) Hemiparethic (n=11, 54.8±13.9 y)	Unsupported^{+}	3D optical motion tracking system	Reaching	Movement time and velocity, joint displacement,
Mackey	2006	KIN	T+A	Hemiplegic CP (n=10, 10-17y) Healthy (n=10, 6-12y)	Supported?$^{?}$	3D optical motion tracking system	Functional tasks	Joint kinematics
Mark	1997	KIN	T+A	Healthy (n=54, undergraduates) *in 3 experiments	Supported?$^{?}$	Video cameras	Reaching	Reach distance, joint movement
Pereira	2014	STAB	T+A	Healthy (n=10, 51.5±5y) Stroke (n=8,60.5±5y)	Unsupported^{+}	3D optical motion tracking system	Reaching	Trunk displacement, hand peak velocity
Reft	2002	KIN	T+A	SCI (SCI level C7-T4, n=5, 23 - 35 y) Healthy (n=5, 23-30 y)	Unsupported and supported$^{?}$	3D optical motion tracking system	Reaching	joint kinematics
Saavedra	2009	KIN + STAB	T+H+A	CP (GMFCS I-III, n=10, 6-16 y)	Unsupported and supported$^{?}$	Electromagnetic tracking system, eye tracking	'Eye only', 'eye-hand' and 'hand only' tasks on computer screen	Reaction time and movement time eye and hand, amplitude head azimuth and view distance, hand peak velocity and submovements

Saavedra	2010	STAB	T+H	CP (GMFCS I-III, n=15, 6-16y) Healthy (n=11, 22-30y; n=26, 4-14y)	Supported^{+}	Electromagnetic tracking system	Quiet sitting with eyes open and closed	Head displacement, velocity, speed
Saavedra	2015	STAB	T+H	CP (GMFCS IV-V, n=15, 4-16y)	Supported^{+}	Electromagnetic tracking system	Quiet sitting	Head position, speed
Santamaria	2016	KIN + STAB	T+H+A	CP (GMFCS III-V, n=17, 2-15 y)	Unsupported and supported^{-}	Electromagnetic tracking system, video recording	Reaching	Movement time, joint kinematics, straightness ratio hand, number of movement units,
Schneiberg	2002	KIN	T+A	Healthy (n=38, 4-11y; n=9, 55±13,7y)	Unsupported^{+}	3D optical motion tracking system	Reach, grasp and bring to mouth	Joint kinematics
Schneiberg	2010	KIN	T+A	Spastic CP (n=13, 6-11y)	Supported?$^{+}$	3D optical motion tracking system	Reach, grasp and bring to mouth	Joint kinematics
Steenbergen	2006	KIN	T+A	Hemiparetic CP (n=5, 14-17y) Healthy (n=5, 22,6±1,5 y)	Supported?$^{?}$	3D optical motion tracking system, EMG	Reach and grasp	Joint kinematics, normalized EMG amplitude
Sveistrup	2008	KIN	T+H+A	Healthy (n=44, 2-11 y; n=6, 22-32y)	Unsupported^{+}	3D optical motion tracking system	Reach, grasp and bring to mouth	Joint kinematics, head stability, movement directions
van der Heide	2005	KIN + STAB	T+H+A	CP (GMFCS I-V, n=58, 2-11 y) Healthy (n=29, 2-11 y)	Unsupported (if possible)$^{-}$	3D optical motion tracking system	Reaching	Spatial angles of head, pelvis, arm and trunk at movement onset and angular displacement
Verheyden	2011	KIN	T+H+A	Healthy (n=20, 65±1.1y) Stroke (n=24, 66±3.1y)	Unsupported^{+}	3D optical motion tracking system	Reaching	Movement sequence, joint angle at maximum reach, peak velocities

Abbreviations: KIN = kinematic outcome measures, STAB = stability outcome measures, T = trunk, H = head, A = arm, EMG = elektromyography, ? symbol indicates that it was not claer whether the measurement was performed supported or unsupported, with or without foot support.
Joint kinematics can include joint angles and information about position, velocity or acceleration.

Interaction trunk – arm

Target distance and object weight have been identified as determinants of trunk involvement during reaching in healthy adults [16]. The trunk is already involved in movement when reaching at approximately 90% of arm length distance [2, 17, 18] and when performing daily tasks within arm length [19]. Healthy children up to the age of 10 years, used their trunk significantly more compared to adults when reaching forward within arm length and also showed more variability [12, 20]. Children with CP showed even more trunk movement and decreased elbow extension when performing various arm tasks compared to healthy children [20-29]. Increased trunk movement is regarded as a compensatory strategy for impaired elbow extension and supination, particularly when reaching in the sagittal plane. Even when reaching forward with the least affected side, increased trunk flexion has been reported in children with CP, albeit non-significant [28]. In addition, increased trunk rotation has been described by Kreulen, et al. [24] when performing a drinking task.

With greater target distance, trunk movement increased in all planes in healthy children, but only trunk flexion increased in children with CP [28]. Increased trunk flexion was associated with more elbow extension in healthy children, whereas it was associated with less elbow extension in children with CP [28]. Besides differences in trunk movement, the movement of the reaching arm was slower and less straight, and peak velocity was lower in children with CP compared to healthy subjects [26, 30].

Postural stability has been shown to be influenced by task demands in healthy subjects [4, 18, 31]. Increased stability was seen when a large degree of precision was required (e.g. tracing task) and decreased stability when performing UE movements which perturb posture more (e.g. aiming task) [4]. Children with CP showed postural imbalance while sitting as indicated by decreased maximum reaching distances and/or reaching performance [30, 32, 33], increased body sway [34], or a decreased Trunk Control Measurement Score [35]. Postural stability was found to be worse during task performance compared to quiet sitting in children with CP [20, 35]. However, worse postural control did not always influence the accuracy of task performance during throwing, as shown by Huang, et al. [32]. Postural stability was worse when reaching laterally compared to reaching forward in children with CP [30, 34, 35]. Saavedra, et al. [26] and Santamaria, et al. [29] studied the influence of external support on trunk stability and arm function. Adding external trunk support improved reaching performance and posture. The adequate level of support was dependent on disease severity; patients with Gross Motor Function Classification Scale (GMFCS) levels I or II already benefitted from pelvis support, whereas patients with GMFCS level V needed support at axillary level [36]. Importantly, adverse effects on reaching performance and posture were seen when the level of support was higher than the trunk level at which postural deficiencies were observed [29]. Differences in postural stability between different types of CP were also found by Heyrman, et al. [35]. Children with bilateral CP of the lower extremities were less impaired in terms of trunk stability, compared to those with bilateral CP of the lower and upper extremities. Children with bilateral CP of the lower extremities had only minor problems of static sitting balance, whereas children with bilateral CP of the lower and upper extremities had significantly impaired postural control while sitting. Children with bilateral CP of the lower limbs had more difficulties when reaching laterally compared to reaching forward [35].

In people with SCI, trunk movement and stability during reaching has been studied by a few research groups [37-41]. Patients with SCI (injury level C7-L4) adapted their trunk and arm movement strategies mainly to maintain trunk stability [40, 41]. Both arm and trunk movement paths were less straight and the peak speed was much lower compared to healthy adults [41]. The reduction in movement speed remained when the back was strapped to the backrest. When reaching forward to near targets, trunk extension was seen in some patients with SCI (injury level T4-L4) [40], whereas reduced trunk flexion was seen compared to healthy adults when reaching to distant targets (injury level C7-T4) [41]. Kim, et al. [40] concluded that these findings might be related to the need for counterbalance in SCI when stretching the arm for reaching, which means that the trunk moved in the opposite direction of the arm to maintain balance. This counterbalance could also be provided by using the other arm. Yet, the variation in movement patterns was large, which was most likely associated with the level of injury [40]. Patients with low thoracic lesions (injury level T8-T12) have grossly intact trunk muscle innervation. These patients performed better in terms of dynamic sitting stability compared to patients with high thoracic lesions (injury level C7-T7) [37, 38]. In addition, reaching distance was larger in patients with low compared to high thoracic lesions. However, no differences in quiet sitting ability were observed in patients with different (thoracic) levels of SCI by Chen, et al. [37]. Trunk length significantly influenced trunk stability [37, 38]. The center of gravity shifts upwards with increasing trunk length and, as a result, it is more difficult to maintain the center of gravity over the base of support. Comparable to the patients with CP, the limits of stability in patients with SCI (injury level above T12) were smaller when reaching laterally compared to reaching forward [39].

Interaction trunk – head

In healthy children, it has been shown that head movement is not influenced by a decrease in trunk stability due to task demands [4]. This is related to the preferred head movement strategy, namely to stabilize the head in space rather than with respect to the trunk [13]. Children with CP may have both impaired trunk and impaired head stability, and deficits in trunk control may indirectly affect head stability as well. Adding external trunk support improved head stability in children with CP, but this result was very much dependent on disease severity and level of support [29, 42, 43]. Head stability improved with higher support level in healthy children, healthy adults and in children with CP (GMFCS levels I – III). Still, head stability in the sagittal plane was worse in children with CP compared to healthy persons even when supported at thoracic level [20, 42], indicating that children with CP had deficits in both trunk and head stability. Support at mid-thoracic level resulted in improved trunk stability, postural alignment and reaching performance in children with GMFCS level IV, whereas support at axillary level restricted them in their active movements and, therefore, negatively affected their posture and reaching performance [29, 43]. For children with GMFCS level V, head stability did not improve significantly with external support at axillary level, indicating that even this level of support was not sufficient for improving reaching performance. Santamaria, et al. [29] found improvement in head alignment with support at axillary level, whereas Saavedra and Woollacott [43] did not find any effect.

No studies were found regarding the interaction between trunk and head in patients with SCI.

Interaction trunk – head – arm

No studies were found describing the interaction between all three segments combined in any group of neurological patients. However, a few studies in healthy children provided results on the interaction between the trunk, head and arm. Head and trunk movement directions with respect to the arm and strategies for head movement relative to the trunk, varied across movement planes and reaching distances (Figure 2) [13]. These interactions also mature at different ages.

The trunk starts moving prior to the arm movement when reaching forward beyond arm length [12, 17, 18], however, the literature is contradicting whether this also applies to reaching within arm length. When reaching laterally, a top-to-bottom sequence (head-trunk-pelvis) was found in healthy adults [44].

DISCUSSION

This review provides an overview of trunk movement and stability when performing upper extremity activities in a seated positionin neurological patients with a flaccid trunk compared to healthy subjects. Overall, in most studies, the study samples were relatively small and rarely exceeded 15 participants per group. Larger studies including more than 30 subjects, were mostly performed on healthy persons [2, 4, 12, 13], although we were able to identify two such studies of patients with SCI [37, 38] and one study including patients with CP [33]. No studies were found on patients with neuromuscular disorders like DMD, SMA or spinal dysraphism/spina bifida.

Nevertheless, there were common key findings that will be discussed below.

Trunk-head-arm movement during reaching

Reaching distance seems to be a crucial factor in choosing a movement strategy for both healthy persons and neurological patients (see Figure 3). Different movement strategies were found for different groups when reaching towards targets within 90% arm length (see figure 3A). Healthy adults showed no trunk flexion [2], whereas healthy children showed some trunk movement [12, 20], and children with CP even more trunk flexion [20, 21, 24, 25, 27, 28]. In contrast, patients with SCI tended to move their trunk backwards [40]. Apparently, different patients use different movement strategies to compensate for their impairments. For instance, it may be easier to accurately perform a reaching task when the arm is not completely extended (due to a smaller moment arm), which implies that more trunk flexion is needed. This strategy is seen in patients with CP, but also to some extent in healthy children. For patients with SCI, the main challenge is to maintain sitting balance while reaching. Hence, stretching out the arm (resulting in a forward center-of-mass displacement) is compensated by trunk extension to maintain balance. Remarkably,

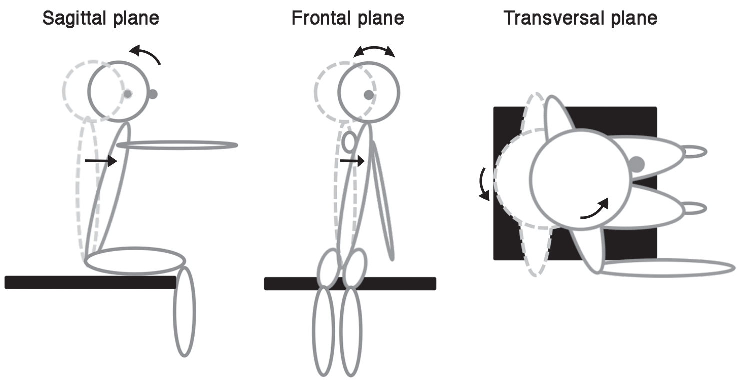

Figure 2 Movement directions of the trunk and head when reaching forward at arm length according to the study by Sveistrup, et al. [13]. In the sagittal plane, the trunk flexes in the direction of reach, while the head bends backwards when reaching at arm length or beyond. In the frontal plane, the trunk moves away from the reaching arm, while the head moves either towards or away from the reaching arm. In the transversal plane, the trunk and head predominantly rotate away from the direction of reach in children (> 3 years) and adults.

these differences in movement strategy seem to diminish with increasing reaching distance (see figure 3B) [12, 13, 28, 41]. When reaching farther, children with CP seem to be capable to apply the same movement strategy as healthy children and no longer use compensatory strategies. However, this may well occur at the expense of movement quality in terms of speed, accuracy or efficiency to explain why they do not use comparable strategies at shorter distances. In addition, their maximal reaching distance is limited compared to healthy persons [30, 32, 33]. Besides, only CP children with sufficient trunk control (i.e. mild CP) are capable of reaching farther than arm length distance. Patients with SCI showed less trunk flexion when reaching farther, because their ability to displace the center-of-mass within the base of support is lower than in healthy subjects. Limiting trunk flexion probably prevents them from falling over, but it also decreases their maximum reaching distance [38, 39]. Adding object weight when reaching results in even earlier trunk involvement than described above [16]. It is, therefore, important that patients are able to use trunk compensation strategies in order to achieve daily tasks, even when performing tasks within arm length distance.

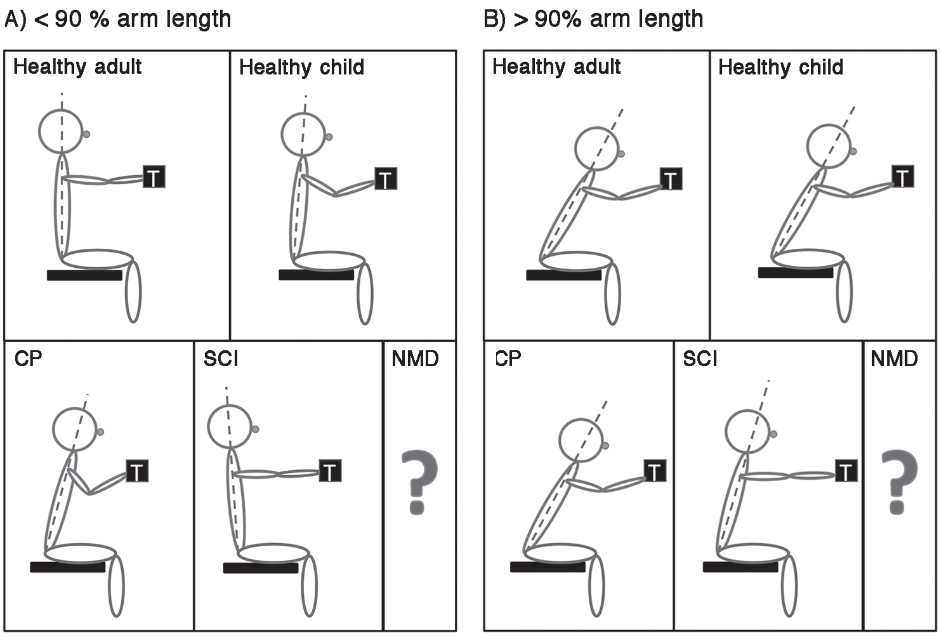

Figure 3 Schematic overview of differences in movement strategies when reaching forward to near targets (A) and targets further away (B). ? represents the unknown movement strategies, T represents the target and the arrows represent the head movement.
CP = cerebral palsy, SCI = spinal cord injury, NMD = neuromuscular disorders.

Stabilizing the head in space (i.e., not with respect to the trunk) was the most common strategy used by healthy children and adults [4, 13]. This strategy facilitates the fixation of gaze on the target while reaching, and thus optimizes visual feedback of task performance. For patients with a flaccid trunk it appears difficult to stabilize their head in space because of trunk instability, as was shown by Saavedra, et al. [42] in children with CP (GMFCS levels I-III). Head stability improved when external trunk support was given to these patients; nevertheless, some of them still showed head instability during trunk support probably due to reduced control of the neck muscles. Unfortunately, no research was available on head-trunk interaction in patients with other conditions, such as SCI or NMD. Thus, further research should take into account head stability problems due to both trunk instability and neck-head stability.

Based on the literature, it is beyond doubt that normal interaction between trunk, head and UE movements typically develops during childhood [12, 13]. Hence, the developmental stages with age of trunk-head-arm interactions must be kept in mind when looking at neurological patients with a flaccid trunk during childhood. It is, however, difficult to give precise age boundaries for the various stages, as movement strategies vary between different planes and age groups [13]. Variability seems to be highest in the sagittal plane when reaching to nearby targets in healthy children below the age of 10. This implies that when a physician examines a 7-year-old child with CP reaching forward to a nearby target and observes trunk extension, this might be interpreted as ‘abnormal’ compared to a healthy adult, but (s)he should realize that approximately 30% of the healthy children of the same age show the same movement strategy.

Postural stability and influence of reaching direction

The definition of (postural) stability varied among the included studies or was sometimes even lacking. The most common definition was keeping or returning the center of mass over the base of support while performing self-initiated actions [18, 37]. Thus, ‘stability’ refers to a ‘dynamic’ situation, whereas ‘balance’ most often refers to a ‘static’ situation. Using the same definitions in research is important to be able to validly compare data and increase our understanding of postural control in various disorders.

Two types of strategies can be distinguished to maintain trunk stability during reaching in patients with a flaccid trunk. To minimize the demands on trunk control, patients can either reduce the proximal degrees of freedom of the reaching synergy or they can reduce the movement speed of the arm. Both strategies are seen in patients with CP [26, 28, 30, 43]. While reduction of the proximal degrees of freedom reduces motor complexity, it also narrows the workspace because movement of the trunk cannot be fully used to reach at the farthest distances. Slower arm movements reduce the perturbing forces exerted on the trunk induced by arm motion and, thus, require less trunk control, but they may be insufficient to complete UE tasks within certain time constraints. Reduction of arm movement speed is also seen in patients with SCI [41]. Remarkably, this strategy remains when the trunk is strapped to the backrest, which justifies the question whether it is used to minimize demands on trunk control. It may also be related to the performed UE task because, according to Fitts' law, movement speed is dependent on the required precision of task execution [45].

To minimize center-of-mass displacement, patients with SCI use their trunk or contralateral arm to counterbalance the perturbing effect of the reaching arm [40, 41]. According to the literature, a different strategy is to change the base of support by displacing the legs and feet while sitting to increase the limits of stability [18, 34]. This strategy is primarily observed in healthy children and adults, because it is much more difficult for patients with poor motor control of the lower extremities. Proper feet positioning in a wheelchair is, therefore, extremely important to maximize the limits of stability while reaching. However, despite proper feet positioning, it may be that force transfer through the legs and feet is still different in paraparetic patients compared to healthy individuals.

In most patients, trunk stability was more affected in the frontal plane than in the sagittal plane because of the smaller mediolateral base of support while sitting and the more complex reaching movements (i.e. requiring both axial rotation and lateral bending of the trunk). Patients with CP had difficulties in both respects and, therefore, found lateral reach more challenging than forward reach [30, 34, 35]. When reaching laterally, healthy children widen the distance between their feet to create a larger base of support, but particularly children with bilateral CP of the lower extremities [34, 35] and patients with SCI [38-40] have trouble to adopt this strategy, which reduces their maximal reaching distance. It should be kept in mind that for wheelchair-bound patients, even those who are able to adjust their foot position, it may not be possible to widen the base of support due to the constraints imposed by footrests. Therefore, providing additional support in the frontal plane may be necessary for sufficient trunk stability

Influence of trunk support

When external trunk support is given to patients with a flaccid trunk, an optimal balance should be sought between providing stability and allowing movement of the trunk. The level of support can vary based on individual needs, ranging from pelvic support to complete thoracolumbosacral orthoses, and from rigid to flexible structures [46]. Such orthoses can provide the required stability to optimize UE function in specific tasks, but they may (partly) prevent movement of the trunk, especially when the support level is too high or the orthosis is too rigid [29]. This can limit patients in performing daily activities due to restrictions in the use of compensatory movements [29, 47, 48], even when performing tasks within arm length. Hence, there is a need for dynamic orthoses that allow all the trunk movements necessary to perform seated activities, while at the same time ensuring trunk stability.

Clinical implications of compensatory trunk movements

To optimize task performance, several compensatory trunk movements are seen in patients with a flaccid trunk as described in this review. These are strategies either to increase movement (i.e. to compensate for impaired arm function) or to maintain postural stability. By applying compensatory trunk strategies, daily tasks can often be executed which could otherwise not be achieved or tasks can be performed more efficiently (in terms of speed, accuracy or energy expenditure). However, the frequent use of compensatory trunk movements may also have a downside in the long-term due to overexertion of certain trunk muscles and joints and/or disuse of arm muscles and joints [49]. While overexertion may lead to muscle and joint pain, disuse should be avoided because of enhanced risk of muscle wasting and development of contractures. Hence, a balance needs to be found by patients and clinicians between, on the one hand, using compensatory strategies to achieve optimal task performance and, on the other hand, prevent long-term adverse effects of using such strategies.

Study limitations

This review has some limitations. First, although we performed a rigorous systematic search, the type and quality of the identified studies allowed a narrative rather than a systematic review. Second, we included only articles written in the English language using PubMed as an electronic database, and these were selected and read full text only by the first author (LP). However, all articles were carefully checked for relevant references to include unidentified studies. Further, we chose not to report and compare the exact movements of the trunk as these were expressed in degrees, movement path, or sometimes even in other measures. As a consequence, and because absolute measurements of trunk movement depend on trunk length, accurate comparisons of trunk movements were not possible. In addition, different definitions of arm length were used between the studies (e.g. from acromion to wrist, finger tip, or some other anatomical hallmark). This influences the interpretation of the timing of trunk involvement in the reaching strategy. Most studies stated that the trunk was already involved when reaching at 90% of arm length distance but, when the definition of arm length differs, the actual reaching distance is also different.

Future directions

Future studies should take into account the following factors. First, standardized clinical measures are missing, especially a valid trunk control scale that is applicable to all patients with a flaccid trunk. Although trunk control scales do exist, they are mostly developed for a specific patient group or unsuitable for patients that fully depend on a seated position. The Trunk Control Measurement Scale (TCMS) [50] might be a suitable trunk control scale which assesses both static and dynamic trunk control while sitting. However, it has been validated only in children with CP who can sit without trunk or feet support for at least 30 minutes. To use this scale in all patients, it must first be validated in other patient groups. The scale should also be validated for patients who already have difficulties with sitting without trunk or feet support. For children with CP who cannot sit independently, the Segmental Assessment of Trunk Control [51] could be a suitable scale to assess discrete levels of trunk control. It could be used complementary to the TCMS, but it needs to be validated in other patient groups as well.

Importantly, future studies should adequately discriminate different age groups as the interactions between trunk, head and UE control mature during childhood and are influenced by trunk length. Lastly, future studies should include other patient groups with a flaccid trunk, such as patients with NMD and spinal dysraphism/spina bifida, with the aim to discriminate generic from disease-specific mechanisms of trunk and head instability and of reaching incapacity. Especially if generic mechanisms can be identified, it will be possible to develop common intervention strategies to support postural stability and optimize seated task performance across different patient groups.

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CHAPTER 3

TRUNK, HEAD AND PELVIS INTERACTIONS IN HEALTHY CHILDREN WHEN PERFORMING SEATED DAILY ARM TASKS

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Experimental Brain Research 2018; 236: 2023-2036.

Abbreviations:

NMD	Neuromuscular disorders
ROM	Range of motion
UE	Upper Extremity

ABSTRACT

Development of trunk and head supportive devices for children with neuromuscular disorders requires detailed information about pelvis, trunk and head movement in interaction with upper extremity movement, as these are crucial for daily activities when seated in a wheelchair. Twenty-five healthy subjects (6 – 20 years old) were included to obtain insight in the physiological interactions between these segments and to assess maturation effects. Subjects performed maximum range of trunk and head movement tasks and several daily tasks, including forward and lateral reaching. Movements of the arms, head, pelvis, and sub-sections of the trunk were recorded with an optical motion capture system. The range of motion of each segment was calculated. Contributions of individual trunk segments to the range of trunk motion varied with movement direction and therefore with the task performed. Movement of pelvis and all trunk segments in the sagittal plane increased significantly with reaching height, distance and object weight when reaching forward and lateral. Trunk movement in reaching decreased with age. Head movement was opposite to trunk movement in the sagittal (>50% of the subjects) and transverse planes (>75% of the subjects) and was variable in the frontal plane in most tasks. Both trunk and head movement onsets were earlier compared to arm movement onset. These results provide insight in the role of the upper body in arm tasks in young subjects and can be used for the design of trunk and head supportive devices for children with neuromuscular disorders.

BACKGROUND

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Children with neuromuscular disorders (NMD) suffer from progressive muscle weakness. Generally, they first lose the ability to walk, followed by a decrease in trunk and arm function. Some children, e.g. with spinal muscular atrophy type I or II, may never have the ability to walk while patients with Duchenne muscular dystrophy lose the ability to walk around the age of 12 years [1]. When seated in a wheelchair, autonomy and level of independence are highly dependent on arm function [2]. Patients report that eating and drinking, reaching for objects, writing and personal hygiene are most problematic in daily life and therefore assisting performance of these tasks with supportive devices is of key importance [3].

In addition to control of upper extremity movement, trunk and head control are necessary in accomplishing daily tasks. The interaction between trunk and arm movements is likely most pronounced when reaching to objects beyond arm length distance [4, 5]. However, in healthy children, trunk movement is also seen when performing tasks within arm length distance [4, 6]. Furthermore trunk motions are often needed to maintain postural stability during daily tasks [7]. In healthy children and adults, the head generally shows a countermovement relative to the trunk resulting in a constant head orientation in space [5]. Head movement is also important for visual control of task performance. Maturation affects the interactions between arm, trunk and head movements in children. Interactions in younger children are more variable than in older children [5].

When developing supportive devices for patients with NMD, trunk and head as well as arm movement should be taken into account. Therefore, detailed information is needed about pelvis, trunk and head movement in coordination with arm movements, both in healthy children and in children with NMD. However, literature on these segmental interactions is scarce [8]. In our study, healthy children in the same age range as children with NMD were included to obtain insight in the interaction between upper body segmental movements, prior to studying this in children with NMD.

While there is some knowledge on the interactions of the upper body in healthy children, the trunk is mostly regarded as one rigid segment. The movement of the thorax is often measured, with respect to the pelvis or the world, and is seen as representative for the overall trunk movement. However, the trunk has great flexibility and can probably not be seen as a rigid segment for development of dynamic supportive devices. Clearly, for the development of supportive devices or spinal orthoses it is important to have insight in the movement of the trunk in more detail than as a single segment. This information could result in requirements concerning selection which trunk segment movements should be allowed to move or be supported when performing daily activities.

Therefore, our aim was to obtain more insight in the interaction of trunk, head and arm movements in healthy children with a specific focus on the segmental nature of the trunk.

METHODS

Participants

Twenty-five healthy children and young adults (13 males, 6-20 years) participated in this study. The subjects were evenly distributed over the age range. None of the participants had a history of disorders affecting movement of the upper body. In addition, they had no scoliosis and no pain in arm(s), trunk, neck or head at the time of participation.

Participants were recruited from local primary schools, high schools and university. Prior to participation, informed consent was given by participants when over 12 years old, and by the children's parents or guardians for all participants younger than 18 years old. The study was approved by the medical ethics committee Arnhem-Nijmegen (NL53143.091.15) and all data were handled according to the guidelines of good clinical practice.

Experimental setup

All subjects were seated on a height adjustable chair with a multi-celled air cushion (Starlock, Star Cushion Products, Freeburg, IL), without back- or armrests. Before the measurement, the cushion was formed to each individual shape by releasing air to provide some additional sitting stability and comfort. The sitting height was adjusted so that the knees were flexed 90° and both feet were flat on the ground.

First, subjects were asked to perform a maximum flexion movement of their trunk from a seated position, immediately followed by a maximum extension movement of their trunk (keeping both feet on the ground). They were instructed to move from the upright position to the maximum position at a slow pace (three seconds) and repeated this flexion-extension movement three times. The same was done for maximum axial rotation and lateral bending. The arms were crossed at the chest when performing the flexion-extension and rotation task, and were rested on the upper legs when performing the lateral bending task. No instructions were given regarding pelvis or hip movement. Subsequently, movements were repeated for the head. Here the instruction was to keep the rest of the body as quiet as possible and only move the head. Thereafter, a series of tasks was performed with the dominant hand at a self-selected speed. No instructions were given for the other hand. Several reaching (and placing) tasks were performed: reaching forward, sideways and contra-lateral at a 45 degrees angle in the transverse plane. The subjects were asked to touch a reference frame positioned at the desired position, or to place a weight on the reference frame (Figure 1). Reaching distance, height and object weight were varied, resulting in the following combinations for forward and lateral reaching: nearby-shoulder height-0 gram ("N-S-0"), nearby-shoulder height-500 gram ("N-S-500"), far-shoulder height-0 gram ("F-S-0"), nearby-eye height-0 gram ("N-E-0"), nearby-eye height-500 gram ("N-E-500"), far-eye height-0 gram ("F-E-0"). Contra-lateral reaching was only performed nearby-shoulder height-0 gram and nearby-shoulder height-500 gram. Nearby was defined as 100% arm length, far as 133% arm length. Arm length was defined as the distance from mid-acromion to mid-hand. Furthermore, subjects were asked to perform four daily tasks: displace a porcelain plate from left to right on a table with both hands ("Plate"), bring a cup of 200 grams to the mouth ("Drink"), trace a path with a pencil ("Draw") and place a finger on a number diagram while holding the diagram with the other hand ("Dexterity"). The drink, draw and dexterity task were based on the instructions of the Performance of the Upper Limb [9]. No instructions were given on how to perform the tasks.

Figure 1 Reference frame with 500 gram weight used for performing the reaching tasks.

Data acquisition

Marker positions were recorded at 100 samples/s using an optical motion capture system (Vicon, Oxford, UK). Twenty-five reflective markers were placed on the skin to define the position of the head, trunk, pelvis and both arms (Figure 2). The trunk was divided into four segments (upper thoracic, lower thoracic, upper lumbar and lower lumbar) to obtain a detailed representation of trunk movement [10]. Markers on the head, pelvis and arms were placed according to the Vicon Plugin-Gait model. For 15 subjects, two additional markers were placed on both sides on the iliac crest, as we noticed that the anterior superior iliac spine markers often became invisible when flexing the trunk or moving the arms. The upper thoracic segment was defined by markers on spinous processes of the 7th cervical vertebrae (C7), spinous processes of the 6th thoracic vertebrae (T6), jugular notch and xiphoid process of the sternum. The lower thoracic segment was defined by markers on T6, spinous processes of the

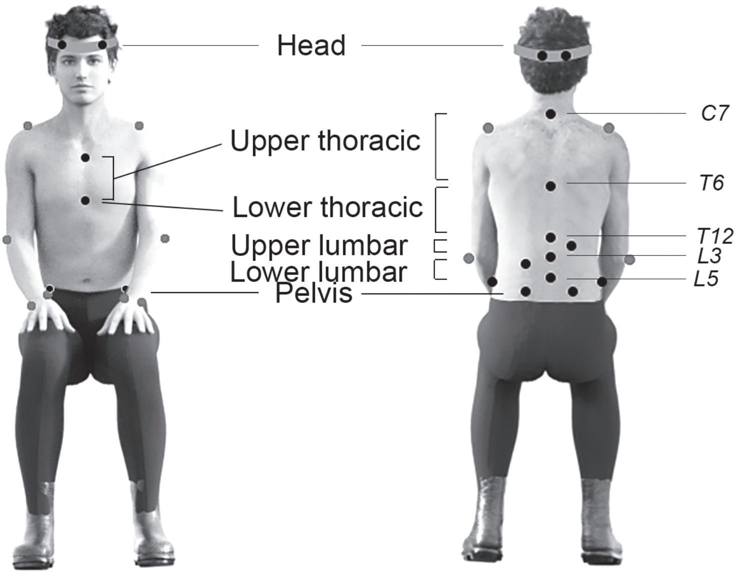

Figure 2 Illustration of marker placement

12th thoracic vertebrae (T12) and the xiphoid process. The upper lumbar segment was defined by markers on T12, spinous processes of the 3rd lumbar vertebrae (L3) and a laterally placed marker at the level of L1/L2. The lower lumbar segment was defined by markers on L3, spinous processes of the 5th lumbar vertebrae (L5) and a laterally placed marker at the level of L4.

Data analysis

Data were filtered with a bi-directional 4th order Butterworth low-pass filter (cutoff frequency of 6 Hz). A biomechanical model was used to calculate the movements of the body segments [11]. Joint coordinate systems were based on the ISB-guidelines [12, 13]. The longitudinal axis was created first for the trunk segments and the following kinematic variables were extracted using Euler decomposition in the following order:

- Pelvis angle: angle of the pelvis relative to the global coordinate system (anterior/posterior tilt – lateral tilt – axial rotation)
- Individual trunk segment angles: angle of a trunk segment relative to the more caudal segment (flexion/extension – lateral bending – axial rotation).
- Neck angle: angle of the head relative to the upper thoracic segment (flexion/extension – lateral bending – axial rotation)

Flexion, lateral bending to the right and rotation to the right were defined as positive. Movements of the different trunk segments were named after the more cranial segment (e.g. upper thoracic angle represents the orientation of the upper thoracic segment relative to the lower thoracic segment). 'Total trunk movement' is used for the summation of all trunk segments.

Pelvis, trunk and neck angles during a recording while sitting quietly (i.e. sitting upright with both hands on the legs (see Figure 2)) were used to zero angles in the movement trials. This was done by post-multiplying the orientation matrix of all segments with the inverse of the orientation matrix while sitting quietly. All kinematics for the two left-hand dominant subjects were transformed to match the kinematics for the right-hand dominant subjects.

To determine maximum ranges of trunk motion, the trial in which the summed angle of all trunk segments and pelvis was maximal in the requested movement plane was selected. Similarly, the trial with the maximum range of neck motion was selected.

For all reaching tasks, the instant of task execution that was used for analyses of segment angles was defined as the first instant where the wrist velocity reached zero after the maximum wrist velocity. For the drink task, this instant was at the point where the hand was the closest to the mouth (i.e. peak of the wrist movement path) and for the plate task, this was where the hands grabbed the plate on the left side (i.e. peak of right wrist movement path). For all of these tasks, the start was identified as the instant where the velocity of the wrist exceeded 5% of its peak velocity. All instants were selected by a computer algorithm and afterwards visually confirmed. For the drawing and dexterity tasks, the instant at task execution was midway between start and end. Start and end were defined manually with the use of video and kinematics recordings, since rendering automatic detection was unfeasible due to low wrist velocity. The ROM was defined as the segment angles at the instant of task execution, subtracted by the segment angles at the start position of the same task. Kinematics of the arms are not reported in this article.

Head movements relative to the upper thoracic segment were categorized in three different strategies: no relative movement between head and trunk, relative movement of the head in the same direction as the trunk, or relative movement of the head in opposite direction to the trunk. The range where the head movement was defined none, was in between plus or minus two times the standard deviation obtained from the head movement during the quiet sitting task. The maximum standard deviation of all participants, in each direction was used for this. For each subject and trial, the head strategy was determined and the percentage of subjects using each strategy was calculated.

Movement onsets of the head and trunk were defined relative to hand movement onset for the reaching tasks, based on 5% of their respective peak velocities. The midpoint between the front head markers was used to determine movement onset of the head, the marker at the jugular notch of the sternum for the movement onset of the trunk and the midpoint between the wrist markers for the movement onset of the hand.

All analyses were performed using Matlab R2014b (Math Works, USA) software.

Statistics

Statistical analyses were performed in SPSS 22.0. Non-parametric tests were used since most of the data was not normally distributed. One-way ANOVA with a Bonferroni-corrected post hoc test, was used to assess differences in ROM between segments when performing maximum trunk movements and when performing daily tasks. Wilcoxon signed rank tests were used to evaluate differences between rotations to the left and right for both trunk and head. A Friedman test, followed by a Wilcoxon signed rank test in case of a significant effect, was used to evaluate the effect of reaching height, distance and object weight on the ROM.

Linear regression analysis was performed to evaluate the correlation between subject age and trunk movement and the effect of age on trunk movement, when performing forward and lateral reaching tasks.

A one-sample Wilcoxon signed rank test was used to evaluate whether the trunk and head movement onset differs from zero (i.e. arm movement onset). The difference between trunk and head movement onsets was evaluated with a two-sample Wilcoxon signed rank test.

The statistical level was set at $\alpha = 0.05$ for all analysis.

RESULTS

Each movement task was successfully completed by all subjects, with the exception of reaching laterally, 1.3 times arm length at eye level. In this task, none of the subjects was able to reach the target and the target was repositioned to their maximum reach distance. Out of 25 subjects, subject data for one subject (12 of 128 kinematic outcomes), for two subjects (13 of 128 kinematic outcomes), and for tree subjects (7 of 128 kinematic outcomes) were excluded due to missing marker data. Kinematic outcomes consist of all segments and tasks.

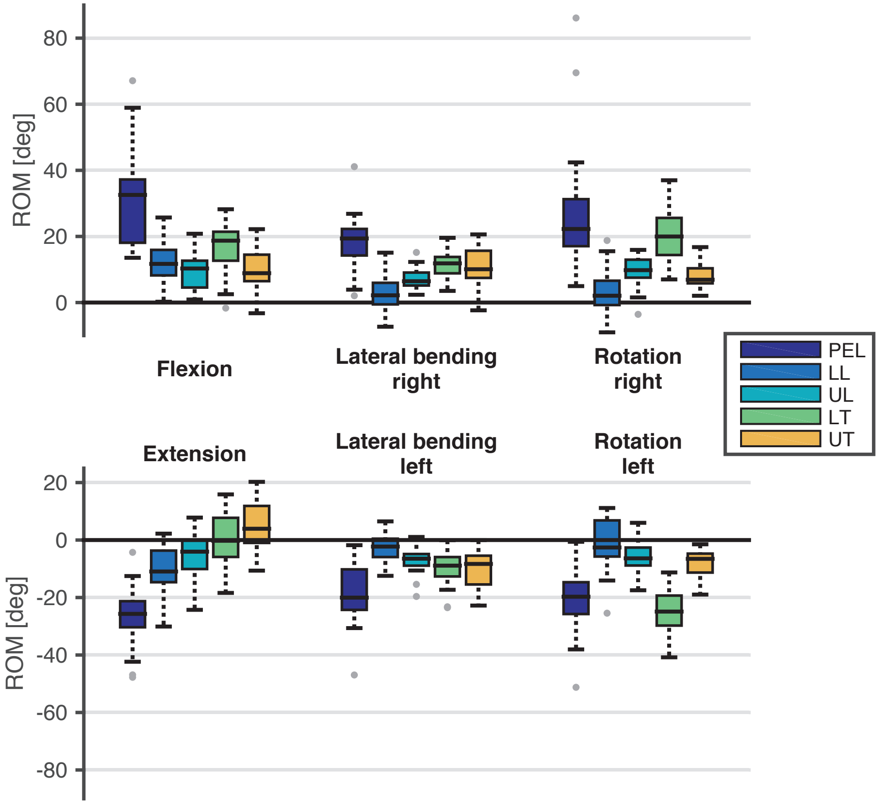

Figure 3 Range of motion (ROM) for pelvis and various trunk segments in the frontal, sagittal and transverse plane, when performing a maximum trunk flexion, extension, lateral bending or axial rotation tasks, respectively.
Boxes represent 25th, 50th and 75th percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range).
Abbreviations: *PEL* pelvis, *LL* lower lumbar segment, *UL* upper lumbar segment, *LT* lower thoracic segment, *UT* upper thoracic segment.

Maximum range of motion tasks

The maximum pelvis and trunk ROM when performing maximum trunk movement tasks are shown in Figure 3. In all movement directions, except for the trunk axial rotation task, the pelvis had a significantly larger contribution than all trunk segments ($p<0.05$). The pelvis and the lower thoracic segment had the largest contribution (i.e. significantly different from the other trunk segments ($p<0.05$)) in the axial rotation task, but were not significantly different from each other. The thoracic segments contributed more in the lateral trunk movement, compared to the lumbar segments. This difference was significant when comparing the lower lumbar segment with both thoracic segments (both $p<0.05$). For the trunk flexion task, the contribution was distributed uniformly over all trunk segments. However, when extending the trunk, the contribution decreased from caudal to cranial segments, and the difference between the two thoracic segments and the lower lumbar segment was significant (both $p<0.005$). The interquartile ranges for both thoracic trunk segments crossed zero, indicating that some participants showed thoracic flexion instead of extension when performing a maximum trunk extension task.

There was no significant difference between left and right total range of motion, both for lateral bending ($p=0.135$) and axial rotation ($p=0.545$). There was a significant difference between flexion and extension ($p<0.001$).

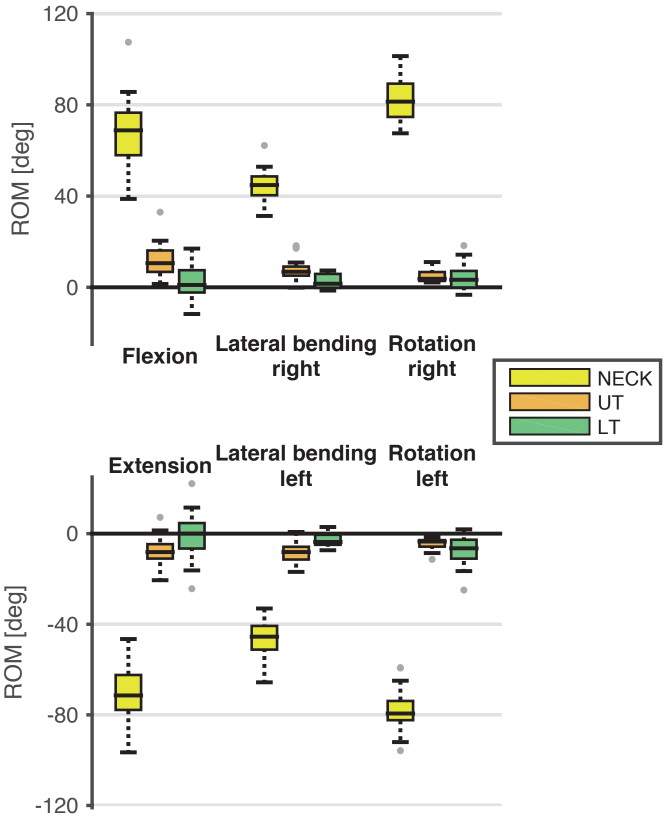

Figure 4 Range of motion (ROM) for neck, upper thoracic (UT) and lower thoracic (LT) trunk segments in the frontal, sagittal and transverse plane, when performing a maximum head flexion, extension, lateral bending or axial rotation task, respectively.
Boxes represent 25^{th}, 50^{th} and 75^{th} percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range).

The median and interquartile ranges for maximum neck ROM are shown in Figure 4. Notable is that also upper thoracic movement (median of 11.6°) was seen when performing the head movements. There was no significant difference between left and right lateral bending (p=0.281) and axial rotation (p=0.386), and flexion - extension (p=0.463).

Trunk movement in reaching and ADL

When reaching forward to a target, trunk ROM in the sagittal plane increased with reaching height, distance and object weight (Figure 5). This increase was significant for almost all segments and with all reaching conditions (Table 1). The more caudal segments (pelvis and lower lumbar segment) showed a flexion movement when reaching forward, while the more cranial segments (upper lumbar and both thoracic segments) showed an extension movement. Lateral bending significantly increased for both thoracic segments and for some reaching conditions in the lumbar segments with all reaching conditions, however this was inconsistent between the reaching conditions (Table 1). There was no consistent, significant increase in axial rotation ROM between the reaching conditions and segments, however quite some trunk axial rotation could be seen in all reaching tasks.

Comparable results were found when reaching laterally (Figure 6). The thoracic segments showed a significant increase in ROM with reaching height, distance and object weight in the frontal plane (Table 2). The pelvis showed a significant increase in ROM with reaching distance and object weight in this plane. In the sagittal plane, both lumbar segments and the upper thoracic segment showed a significant increase with reaching height, distance and object weight. In the transverse plane, only the pelvis showed a consistent, significant increase in ROM with reaching distance and object weight, but not for reaching height.

Trunk movement could be seen in all planes when performing daily activities (Figure 7), even though the activities were within arm length distance. However, the median ROM was often close to zero. Of all the performed tasks, drawing seemed to be the only task where the more cranial trunk segments showed a flexion movement.

Statistical analyses for differences in ROM between segments when performing reaching or ADL tasks, were not performed because of the high variance due to the fact that no specific instructions were given how to perform the tasks. This made it questionable what a significant difference would indicate. Nevertheless, note that the distribution of ROM over trunk segments in all reaching and daily tasks seems quite comparable with the contribution found when performing the maximum trunk movement tasks. The thoracic segments were mostly involved in lateral bending, the lower thoracic segment was mostly involved in axial rotation and the distribution

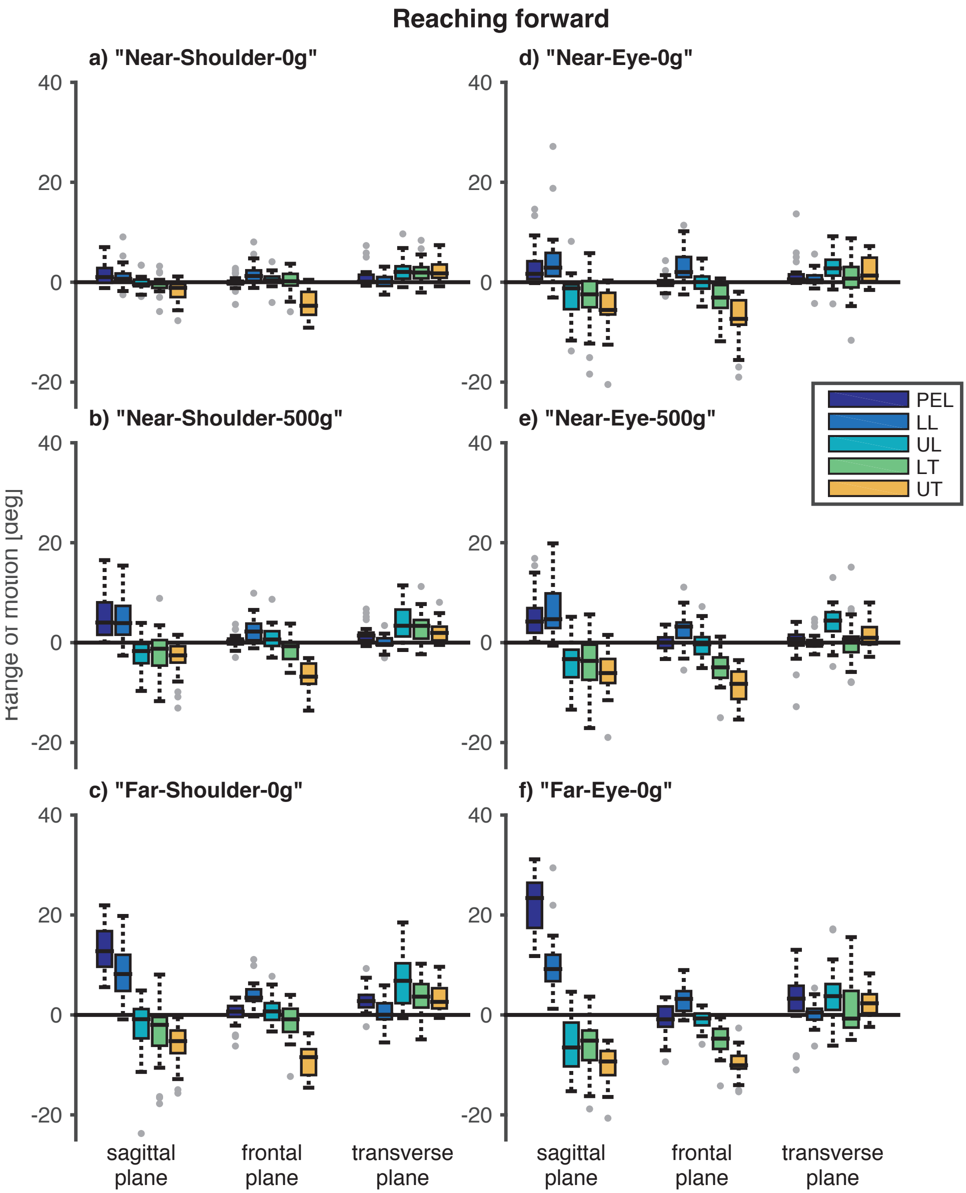

Figure 5 Range of motion (ROM) when reaching forward at different reaching heights, distances and object weights. Positive values indicate respectively flexion, lateral bending to the right and rotation to the right.
Boxes represent 25th, 50th and 75th percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range).
Abbreviations: *PEL* pelvis, *LL* lower lumbar segment, *UL* upper lumbar segment, *LT* lower thoracic segment, *UT* upper thoracic segment.

in ROM in the sagittal plane was approximately equal between all trunk segments. However, the movement direction of the segments differed in the sagittal plane; the more caudal segments showed flexion, while more cranial segments showed extension.

Table 1 P-values for the effects of reaching height, distance and object weight on segment range of motion, when reaching forward.
Abbreviations in reaching tasks: N = near target, F = far target, S = shoulder height, E = eye height, 0 = 0 gram object weight, 500 = 500 gram object weight.

	Reaching height			**Reaching distance**		**Object weight**	
	N-S-0 / N-E-0	N-S-500 / N-E-500	F-S-0 / F-E-0	N-S-0 / F-S-0	N-E-0 / F-E-0	N-S-0 / N-S-500	N-E-0 / N-E-500
Sagittal plane							
Pelvis	**0.040**	0.961	**0.006**	**<0.001**	**<0.001**	**<0.001**	**<0.001**
Lower lumbar	**0.003**	**0.010**	0.140	**<0.001**	**0.001**	**<0.001**	**0.002**
Upper lumbar	**0.009**	**0.002**	**0.014**	0.097	**0.025**	**0.016**	0.122
Lower thoracic	**0.009**	**0.002**	**0.037**	**0.006**	**0.005**	0.150	0.201
Upper thoracic	**<0.001**	**0.001**	**<0.001**	**<0.001**	**<0.001**	**0.016**	0.183
Frontal plane							
Pelvis	0.882	0.527	0.073	0.290	0.394	0.128	0.249
Lower lumbar	**0.048**	0.277	0.223	**<0.001**	0.378	**0.028**	0.592
Upper lumbar	0.078	**0.004**	**0.001**	0.689	**0.028**	0.600	0.657
Lower thoracic	**<0.001**	**<0.001**	**<0.001**	**0.035**	**0.002**	**0.009**	**0.001**
Upper thoracic	**0.001**	**0.005**	0.104	**<0.001**	**0.008**	**<0.001**	0.088
Transverse plane							
Pelvis	0.200	0.506	0.605	**<0.001**	0.144	**0.045**	0.445
Lower lumbar	0.061	0.236	0.884	0.447	0.627	0.397	0.338
Upper lumbar	0.495	0.861	**0.021**	**<0.001**	0.353	**0.020**	0.300
Lower thoracic	0.158	**0.002**	**0.004**	0.880	0.946	**0.042**	0.737
Upper thoracic	0.563	0.065	**0.002**	**<0.001**	0.264	0.619	0.581

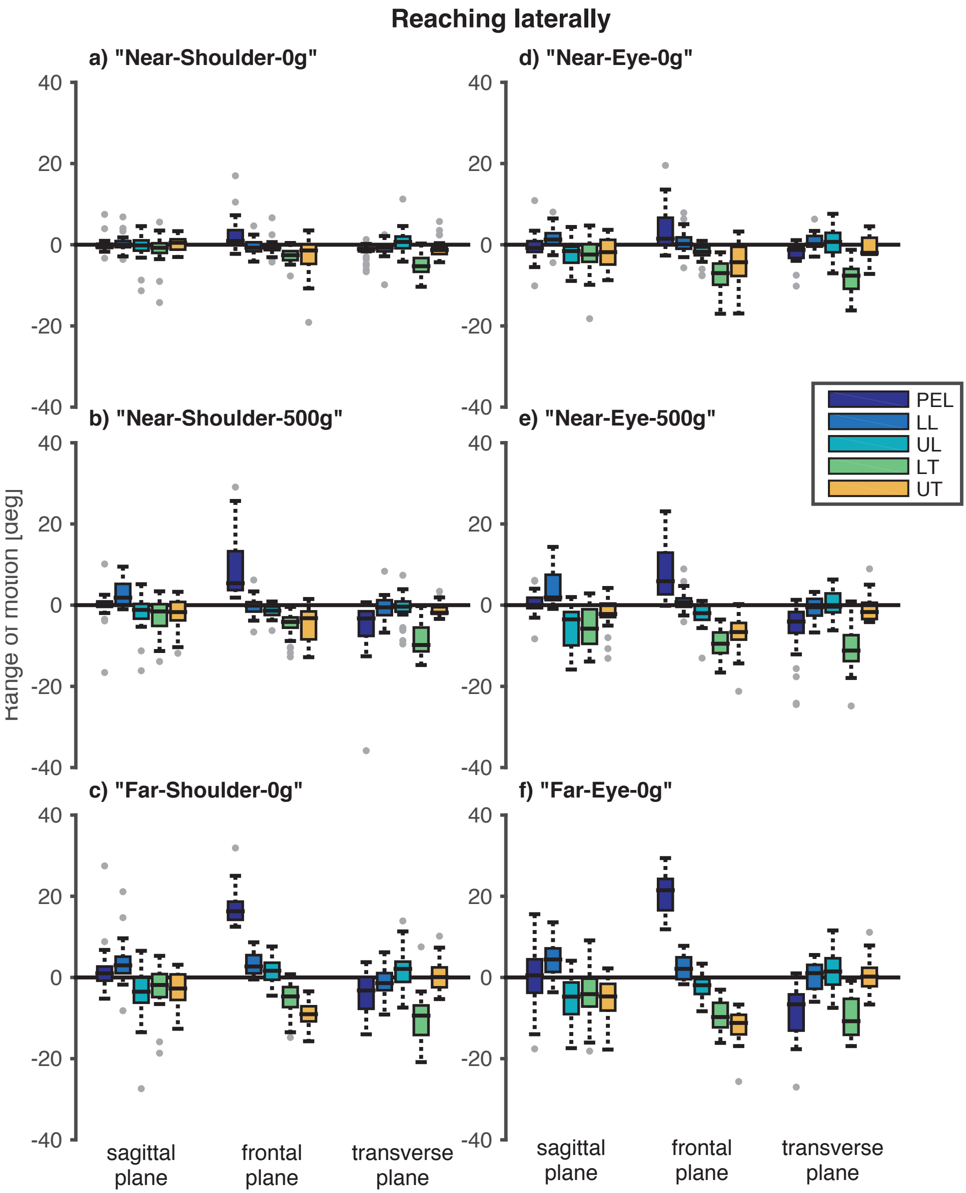

Figure 6 Range of motion (ROM) when reaching laterally at different reaching heights, distances and object weights. Positive values indicate respectively flexion, lateral bending to the right and rotation to the right.
Boxes represent 25th, 50th and 75th percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range).
Abbreviations: *PEL* pelvis, *LL* lower lumbar segment, *UL* upper lumbar segment, *LT* lower thoracic segment, *UT* upper thoracic segment.

Maturation

Figure 8 shows the correlation between age and total trunk movement when reaching forward and laterally. Significant, moderate to strong correlations were found in 10 out of 12 reaching tasks, where younger children used more trunk movement compared to older children. However, a relatively high variability could be seen in the younger children and this variability was higher in reaching forward compared to reaching laterally. The slopes of the regression lines indicated a decrease of trunk ROM of maximal -1.94 degrees/year for the ”F-S-0” task forward and minimal of -0.54 degrees/year for the “N-S-500” task laterally.

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Table 2 P-values for the effects of reaching height, distance and object weight on segment range of motion, when reaching laterally.
Abbreviations in reaching tasks: N = near target, F = far target, S = shoulder height, E = eye height, 0 = 0 gram object weight, 500 = 500 gram object weight.

	Reaching height			**Reaching distance**		**Object weight**	
	N-S-0 / N-E-0	N-S-500 / N-E-500	F-S-0 / F-E-0	N-S-0 / F-S-0	N-E-0 / F-E-0	N-S-0 / N-S-500	N-E-0 / N-E-500
Sagittal plane							
Pelvis	0.221	0.879	0.346	0.061	0.627	0.475	0.248
Lower lumbar	0.058	**0.026**	0.548	**<0.001**	**0.005**	**<0.001**	**0.001**
Upper lumbar	**0.002**	**0.002**	0.189	**0.002**	**0.034**	**0.030**	**0.003**
Lower thoracic	0.122	**0.002**	0.083	0.093	0.158	0.069	**0.003**
Upper thoracic	**0.045**	0.946	**0.005**	**<0.001**	**0.001**	**0.006**	0.619
Frontal plane							
Pelvis	0.443	0.761	**0.007**	**<0.001**	**<0.001**	**<0.001**	**<0.001**
Lower lumbar	0.054	**0.013**	0.527	**<0.001**	**0.006**	0.668	0.121
Upper lumbar	0.054	0.093	**<0.001**	**0.007**	0.932	0.074	0.757
Lower thoracic	**<0.001**	**<0.001**	**<0.001**	**0.003**	**0.032**	**0.001**	**0.003**
Upper thoracic	**0.017**	**0.026**	**0.004**	**<0.001**	**<0.001**	**0.001**	**0.007**
Transverse plane							
Pelvis	0.201	0.301	**0.001**	**0.032**	**<0.001**	**<0.001**	**0.001**
Lower lumbar	**0.025**	0.855	**0.016**	0.376	0.833	0.732	0.055
Upper lumbar	0.696	0.476	0.648	0.209	0.549	0.288	0.427
Lower thoracic	**0.001**	0.109	0.382	**<0.001**	0.201	**0.001**	**0.006**
Upper thoracic	0.677	0.925	0.459	**0.020**	**0.013**	0.242	0.201

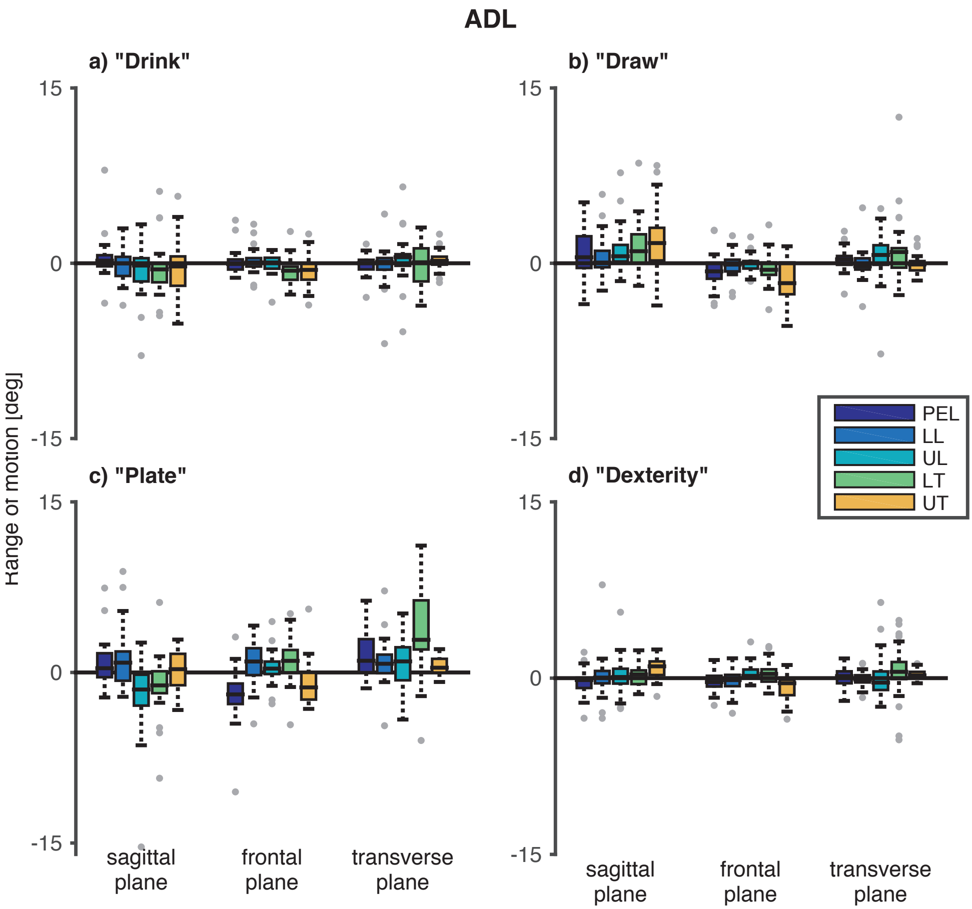

Figure 7 Range of motion (ROM) when performing four activities of daily life. Positive values indicate respectively flexion, lateral bending to the right and rotation to the right.
Boxes represent 25th, 50th and 75th percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range).
Abbreviations: *PEL* pelvis, *LL* lower lumbar segment, *UL* upper lumbar segment, *LT* lower thoracic segment, *UT* upper thoracic segment.

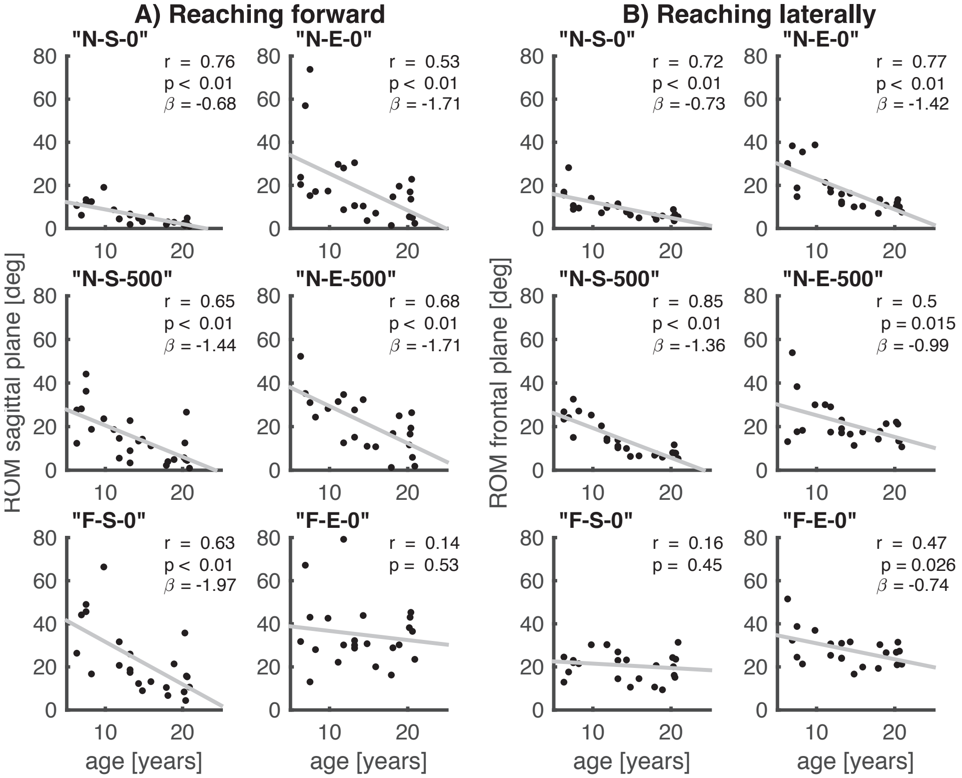

Figure 8 Linear regression between subject age and total trunk range of motion (ROM) in the sagittal plane when reaching forward (A) and in the frontal plane when reaching laterally (B) at different reaching heights, distances and object weights.
Abbreviations in reaching tasks: N = near target, F = far target, S = shoulder height, E = eye height, 0 = 0 gram object weight, 500 = 500 gram object weight.

Head movement strategies

Different head movement strategies were found in the daily activities (figure 9). There was no missing data. Two times the maximum standard deviation of quiet sitting was equal to 2.24° (frontal plane), 2.80° (sagittal plane), 2.02° (transverse plane), and was used as range within the head movement was categorized as none.

In almost all tasks, a variety in head movement strategies was used by the participants. Most consistency could be seen in the transverse plane. Axial rotation movement of the head was in opposite direction to the axial rotation of the trunk when reaching forward and contra-lateral (on average across tasks, 88% of the participants), while the rotation was in the same direction when reaching laterally (on average across

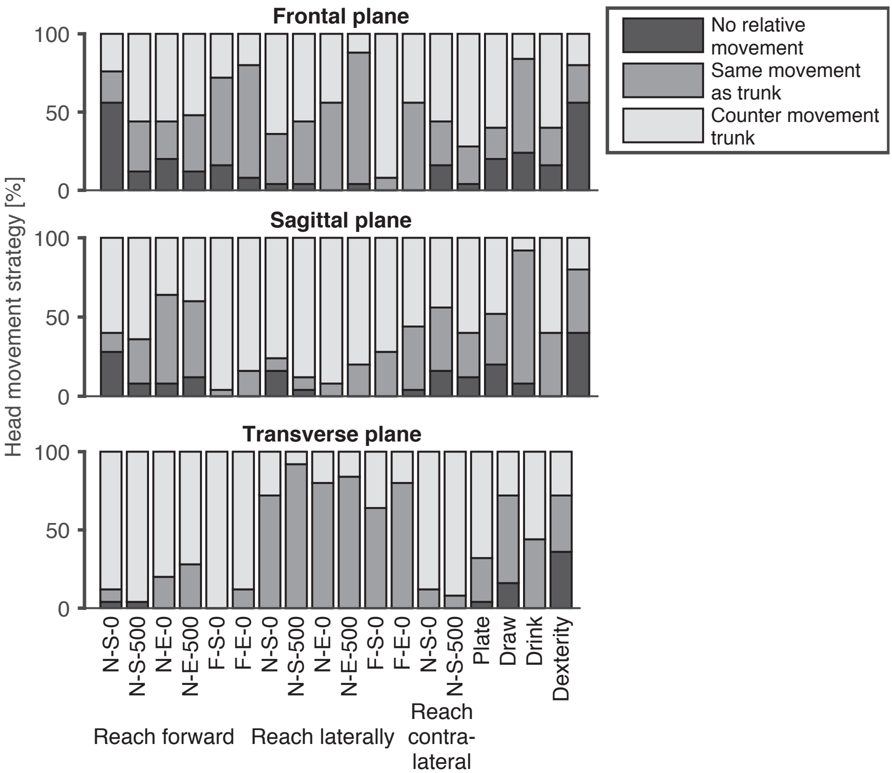

Figure 9 Head movement strategies used by the subjects as percentage of the total group when performing daily tasks. The bars indicate no relative movement between head and trunk, relative head movement in the same direction as the trunk movement and relative head movement in opposite direction of the trunk movement.
Abbreviations in reaching tasks: N = near target, F = far target, S = shoulder height, E = eye height, 0 = 0 gram object weight, 500 = 500 gram object weight.

tasks, 79% of the participants). In the sagittal plane, more than 50% of the participants moved their head in opposite direction to the trunk. However, both for reaching forward and contra-laterally a substantial part of the participants moved their head in the same direction as the trunk (on average of tasks, 30% of the participants). In the frontal plane, 22% of all participants did not move their head relative to the trunk when reaching forward and contra-laterally and when performing daily tasks. This was higher compared to the other movement planes.

For the four daily tasks, the head movement strategy varied. For the dexterity task, more than 36% of the participants did not move their head relative to the trunk in all planes, and when drawing more than half of the participants moved the head in the same direction as the trunk movement in all planes.

Movement onset

When trunk and head onset were equal to the start of the recording, data were excluded from analysis. It could not be guaranteed that these movements were related to the performed task. The number of included subjects is shown in Figure 10.

Compared to the arm motion onset, the head movement onset was significantly earlier in all reaching tasks, and the trunk movement onset was significantly earlier in most reaching tasks (Figure 10). In some tasks when reaching forward, the head onset was also significantly earlier than the trunk onset, resulting in a 'head-trunk-arm' movement sequence. However, the inter quartile ranges were large and also passed the arm movement onset line, indicating that the movement onset for head and trunk was not prior to the arm movement for every subject.

DISCUSSION

The results of this study give insight in the interaction between arm, trunk, head and pelvis movements in children when reaching and performing daily tasks, and in the contribution of different trunk segments to the task in children and young adults.

When performing maximum trunk movement tasks, contributions of individual trunk segments varied with movement direction. In flexion, the contribution was roughly equal among all segments, but in lateral bending the thoracic segments contributed more compared to the lumbar segments, and in trunk axial rotation the lower thoracic segment contributed most. This is in agreement with the study of Preuss and Popovic [14] for axial rotation, where subjects performed target-directed trunk movements. Their results contradict our results in the other two planes. They found the highest contribution in both flexion-extension and lateral bending from the most

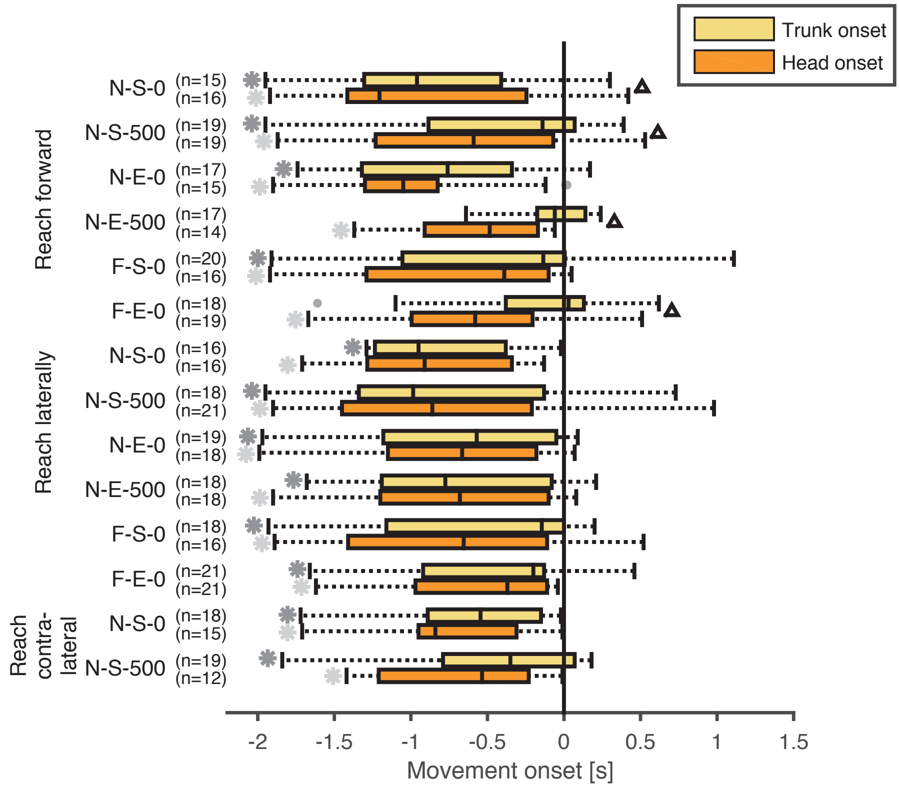

Figure 10 Trunk and head movement onset relative to the arm movement onset for all reaching tasks. Boxes represent 25th, 50th and 75th percentile, wiskers minimum and maximum of non-outlier values, and dots indicate outliers (greater than 1.5 times the interquartile range). * p<0.05 for trunk or head with respect to zero (e.g. arm movement onset), Δ p<0.05 between trunk and head onset. Abbreviations in reaching tasks: N = near target, F = far target, S = shoulder height, E = eye height, 0 = 0 gram object weight, 500 = 500 gram object weight.

caudal segments. These difference are likely due to differences in task instructions. Subjects moved their head along with the trunk in our study, whereas they had to touch a reference with their head in the study of Preuss and Popovic [14]. The pelvis also contributed greatly in all movement directions in our maximum trunk movement tasks, indicating that it has a great influence on the maximum trunk movement.

In accordance with a previous study [15], trunk movement increases with reaching distance and object weight when reaching forward. In addition, we found that this also applies for reaching laterally and for different reaching heights. Moreover, it applies to most trunk segments and the pelvis in the sagittal plane and for the thoracic segments in the frontal plane. It is noticeable that despite the large standard deviations in ROM, subjects adapt similarly to differences in reaching conditions in terms of trunk movement. The trunk segments that showed a significant increased ROM with reaching height, distance and object weight, correspond to the segments contributing the most in the maximum trunk movement tasks: in the frontal plane the thoracic segments and in the sagittal plane all trunk segments, with an exception of the lower thoracic segment when reaching laterally. In the transverse plane, there was no consistent, significant increase in trunk movement between all segments and reaching conditions. This could be explained by the fact that too much trunk rotation will cause an overshoot in arm alignment with the target. Although there was no consistent, significant difference found in axial rotation between the different reaching conditions, axial rotation of the lower thoracic segment was present in each reaching task and therefore seems to be necessary. Again, this is consistent with the finding that the lower thoracic segment contributed the most in the maximum trunk rotation task.

When performing the reaching and daily tasks, anterior tilt of the pelvis and flexion in the lower lumbar segment was seen, while extension was seen in the thoracic segments, indicating that subjects prefer to erect their trunk (decrease thoracic kyphosis and lumbar lordosis) when performing arm tasks. This is in line with suggestions that an erect sitting posture has benefits compared to a slumped posture when performing arm tasks, as it elongates the spine so less arm elevation is needed, and consequently less arm muscle strength, and it ensures a larger shoulder range of motion [16]. Also, the maximum range of axial rotation of the trunk itself increases with a more erect sitting posture [17].

Strong correlations were found between total trunk ROM and age when reaching forward and laterally. Younger children used more trunk movement compared to older children and the variability was higher, indicating maturation of coordination between trunk and arm movements. The strongest correlations were found when reaching near, at shoulder height and without weight, but the effect (in degrees per year) was the least. This maturation effect with age is in line with findings of Schneiberg, et al. [4] and Sveistrup, et al. [5], and should be taken into account when evaluating children with NMD. Age-matched comparison is very important to distinguish between natural and pathologic trunk movements.

Interactions between trunk and head could already be seen when performing maximal head movements, where the upper thoracic segment contributed quite substantial in (mainly) the maximum neck flexion and extension movement, in agreement with Tsang, et al. [18]. When performing daily tasks, the chosen strategy for head movement relative to the trunk, likely depends on maintaining, or achieving, gaze on the target [19]. This could be seen in the transverse plane, where axial rotation of the head was used in the opposite direction to the trunk when reaching forward and contra-laterally, compared to movement in the same direction when reaching laterally. In the sagittal plane, the strategy to move the head in opposite direction of the trunk was most frequently present. However, also a quite substantial percentage of participants did move the head in the same direction as the trunk in several tasks. Variations in strategy might be explained by the relatively small trunk movements, which do not strongly influence the gaze on the object when the head would not move relative to the trunk at all.

Movement onset of the head and the trunk generally seemed to be earlier than the movement onset of the arm when reaching. Only in a few forward reaching tasks, there was also a significant difference between head and trunk onset, resulting in the onset sequence "head-trunk-arm". These findings correspond to previous literature [19, 20], however, the variability of movement onset was very large in our study. This could be caused by the chosen method in this study; we did not instruct participants to sit as quietly as possible before the start. Especially for the younger, more energetic subjects it was difficult to sit quietly. We did ask the participants to look ahead at the beginning of each trial, but especially younger children did not always comply. We tried to eliminate these movements unrelated to the task performed, by excluding the trials in which subjects already moved their head or trunk at the start of the recording before performing the task.

The following considerations should be taken into account when developing new trunk or head supportive devices. Allowing movement between the pelvis and lower lumbar segment is of importance for all movement directions. Based on the relative motions of the lower thoracic segment, allowing movement between the lower thoracic and upper lumbar segments is important for both lateral bending and axial rotation. In additions, since movement of the upper thoracic relative to the lower thoracic segment is quite substantial when bending laterally and when flexing forward, some movement should also be allowed between upper and lower thoracic segments. Although the four trunk segments taken into account in this study still represent a simplification of reality, this analysis provides insight in the minimal degrees of freedom that should be allowed for performance of daily tasks. For a head supportive device, it is important to realize that the head is often moving in the opposite direction of the trunk. Therefore, supportive devices should allow for head rotations independent of the trunk movement. When developing actuated trunk and head supportive devices, they cannot be controlled based on the arm movement when timing of movement is seen as important factor, since the trunk and head generally started to move prior to the arm movement.

Several other limitations of this study warrant some discussion. First, reaching distance and height were set based on the sitting posture of the subject at the given moment, while small changes in posture may influence reaching distance and height. This may have resulted in variance between trials within and between subjects. However, we were interested in self-selected movements of the trunk and hence chose not to standardize initial sitting posture. Second, although the age of the subjects was uniformly distributed over the whole age range, we had only a few participants for each age. Especially because the variability was larger in the younger children, a larger group size would have allowed for a more sensitive analysis of age effects. Third, surface markers were used to identify movement of the segments. Soft tissue movement can result in artifacts in the movement estimation and is a well-known disadvantage of this measurement technique. Especially the soft tissue movement artifacts of the trunk can be quite substantial [21]. However, this influence should be minor when evaluating the range of motion instead of absolute angles according to Zemp, et al. [21]. Last, results of the lumbar segment movement in the younger children should be interpreted with caution, because the markers were placed at a small distance from each other and therefore small artifacts can result in substantial errors.

In conclusion, the contribution of individual trunk segments to the ROM varied with the movement plane with specific task aspects such as distance, height and weight handled. Range of trunk movement decreased with age when performing reaching tasks and this should be kept in mind when evaluating the interaction between trunk and upper extremity movements in children. Increased reaching distance, height and object weight all resulted in increased trunk movement in reaching forward and laterally. Generally, the head moved in opposite direction to the trunk (except in the transverse plane when reach laterally), but the head movement strategy was highly variable in the frontal plane and was also dependent on the task performed. Head and trunk movement onsets were generally earlier than arm movement onset when reaching. Only in a few tasks head movement onset was significantly different from trunk movement onset.

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CHAPTER 4

DON'T FORGET THE TRUNK IN DUCHENNE MUSCULAR DYSTROPHY PATIENTS: MORE MUSCLE WEAKNESS AND COMPENSATION THAN EXPECTED

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Journal of NeuroEngineering and Rehabilitation 2019; 16(1); 44

Abbreviations:

DMD	Duchenne Muscular Dystrophy
HC	Healthy Controls
ROM	Range of motion
sEMG	Surface Electromyography
UE	Upper Extremity

ABSTRACT

Background: Performing daily activities independently becomes more difficult in time for patients with Duchenne muscular dystrophy (DMD) due to muscle weakness. When performing seated daily activities, the trunk plays an indispensable role besides the upper extremities. However, knowledge is lacking on the interaction between trunk and upper extremities. Therefore the aim was to investigate whether patients with DMD use trunk movement to compensate for reduced arm function when performing seated tasks, and whether this is related to increased muscle activity.

Methods: Eighteen boys with DMD and twenty-five healthy controls (HC) performed several tasks when sitting unsupported, like reaching (and placing) forward and sideward, drinking and displacing a dinner plate. Maximum joint torque and maximum surface electromyography (sEMG) were measured during maximum voluntary isometric contractions. Three-dimensional movements and normalized sEMG when performing tasks were analyzed.

Results: Significantly decreased maximum joint torque was found in DMD patients compared to HC. Trunk and shoulder torques were already decreased in early disease stages. However, only maximum trunk rotation and shoulder abduction torque showed a significant association with Brooke scale. In all reaching and daily tasks, the range of motion in lateral bending and/or flexion-extension was significantly larger in DMD patients compared to HC. The trunk movements did not significantly increase with task difficulty (e.g. increasing object weight) or Brooke scale. Normalized muscle activity was significantly higher in DMD patients for all tasks and muscles.

Conclusions: Boys with DMD use increased trunk movements to compensate for reduced arm function, even when performing relatively simple tasks. This was combined with significantly increased normalized muscle activity. Clinicians should take the trunk into account when assessing function and for intervention development, because DMD patients may appear to have a good trunk function, but percentage of muscle capacity used to perform tasks is increased.

BACKGROUND

For patients with Duchenne muscular dystrophy (DMD), performance of daily activities becomes more difficult over time due to progressive muscle weakness. DMD is an x-linked neuromuscular disorder with an incidence of approximately 1 in 6000 live male births [1]. Mean loss of ambulation is around 11 years with use of corticosteroids in the Netherlands [2], but patients report difficulties in performing daily activities involving arm movements already earlier [3].

Decreased upper extremity function is already visible in early stages of DMD and precedes the decline in activity performance [4]. Trunk weakness seems to occur in later disease stages. Trunk function seems relatively good and stable in the ambulatory phase, but starts to decrease when boys become non-ambulant [5, 6]. However, both measures used (Segmental Assessment of Trunk Control [5] and Motor Function Measure [6]) are influenced by upper extremity function too and therefore might not completely represent trunk function alone.

Knowledge concerning the relation between upper extremity movement and trunk movement in patients with DMD is completely lacking at present [7]. In healthy adults and children, coordination of upper extremity and trunk motion is essential for accomplishing daily tasks [8, 9]. For DMD patients this may be even more, because clinically they show increased trunk movement to compensate for reduced arm function. Understanding the use of compensatory trunk movements could be beneficial for the development of interventions, such as physical exercise training, seating adjustments and assistive technology.

Therefore, the aim of this study was to investigate how DMD patients use trunk movement to compensate for reduced arm function. We hypothesise that compensatory trunk movement is dependent on task difficulty, disease progression and related to increased trunk muscle activity.

METHODS

Participants

Eighteen male DMD patients and twenty-five healthy controls (HC) (13 males) participated in this study. Participants were included if they were between 6 and 21 years of age, able to show arm motor skills at request and could sit independently (without back or arm rests) for at least 10 minutes. DMD patients needed to have a genetically confirmed diagnosis of DMD. Participants were excluded if they had (other) diseases affecting the arm, trunk or head movements, and if they had received spinal fusion surgery.

DMD participants were recruited through advertisements by two patient organizations (Duchenne Parent Project and Spierziekten Nederland) and through the outpatient clinic of the Radboudumc in Nijmegen. HC were recruited from local primary schools, high schools and university. Prior to participation, written informed consent was given by participants when over 12 years old, and by the children's parents or guardians for all participants younger than 18 years old. The study was approved by the medical ethics committee Arnhem-Nijmegen (NL53143.091.15) and all data were handled according to the guidelines of good clinical practice.

Procedures

We used the same procedure as the one employed in a previous study with healthy children [10]. All participants were seated on a height adjustable chair with a multi-celled air cushion (Starlock, Star Cushion Products, Freeburg, IL), without back- or armrests. The sitting height was adjusted so that the knees were flexed 90° and both feet were flat on the ground.

First, to determine maximum trunk range of motion, participants were asked to perform a maximum active flexion movement of their trunk from a seated position, immediately followed by a maximum active extension movement of their trunk (keeping both feet on the ground). The same was done for maximum axial rotation and lateral bending. Thereafter, a series of tasks were performed with the dominant hand at a self-selected speed. Several reaching (and placing) tasks were performed at shoulder height: reaching forward, sideways and contra-laterally at a 45 degrees angle in the transverse plane. Participants had to touch a reference frame positioned at the desired position, or to place an object on the reference frame. Reaching distance and object weight were varied, resulting in the following combinations for forward, lateral and contra-lateral reaching: nearby-0 gram ("N-0"), nearby-500 gram ("N-500"), far-0 gram ("F-0"). Contra-lateral reaching was not performed at a far distance. Nearby was defined as the distance that could be reached by stretching the arm (i.e. 100% arm length for HC, but could be closer for DMD) and far as 133% of arm length when possible, otherwise as maximum reaching distance. Arm length was defined as the distance from mid-acromion to the centre of the hand. Furthermore, subjects were asked to perform two daily tasks: displace a porcelain plate (circa 600 grams) from left to right on a table with both hands ("Plate") and bring a cup of 200 grams to the mouth ("Drink"). The drink task was based on the instructions of the Performance of the Upper Limb [11]. No instructions were given on how to perform the tasks.

Outcome measures

Participant characteristics

The following participant characteristics were noted based on self-reports: age, weight, height, arm preference, age of diagnosis (if applicable), use of corticosteroids, wheelchair confinement, pain in upper body at time of participation and occurrence of scoliosis. Sitting height was measured and, for DMD patients, the Vignos lower extremity scale [12] and Brooke upper extremity scale [13] were used for clinical assessment of leg and arm function, respectively.

4

Three dimensional motion analysis

We used the same data acquisition and analysis as employed in a previous study with healthy children [10]. An optical motion capture system (Vicon, Oxford, UK) was used to record 25 single reflective markers, which were placed on the skin to define positions and orientations of the head, trunk, pelvis and both arms during task performance. The markers divided the trunk initially into four segments (upper thoracic, lower thoracic, upper lumbar and lower lumbar), because the trunk cannot be seen as rigid segment [10]. However to make the data more concise, we decided to report here the trunk movement as one segment (i.e. summation of the trunk and pelvis segment angles). Distribution of movement patterns over the individual trunk segments was essentially the same among HC and DMD.

All kinematics data were filtered with a bi-directional 4th order Butterworth low-pass filter (cutoff frequency of 6 Hz). Trunk joint angles are expressed relative to the global coordinate system, and are described in all three movement directions: flexion-extension (i.e. sagittal plane), lateral bending (i.e. frontal plane) and axial rotation (i.e. transversal plane).

Maximum trunk joint angles in all three movement directions were determined when performing the active range of motion (ROM) tasks for trunk. For the reaching tasks and daily tasks, the trunk ROM between the start and end of the task was determined. Start and end of a task were defined as the time where the velocity of the wrist exceeded/got below 5% of its peak velocity. Direction of the movement was defined for all reaching tasks by subtracting the trunk joint angle at the time of touching the reference frame, from the joint angle at the start. This defined whether the movement was in a positive or negative direction (i.e. flexion or extension, or towards dominant or non-dominant side). Towards dominant side reflects the side of the hand used to perform the tasks.

Joint torque and surface electromyography

Muscle activity was measured with the use of surface electromyography (sEMG) (Zerowire EMG, Aurion, Italy) and was recorded with a sample frequency of 1000 Hz. Electrodes were placed on the following muscles on both sides of the body: iliocostalis (6 cm from spinous processes of the 1st lumbar vertebrae), longisimus (3 cm from spinous processes of the 3rd lumbar vertebrae), external oblique (3 cm from axillae midline at height of umbilicus), trapezius descendens (1/2 on the line from the acromion to the spinous processus of the 7th cervical vertebrae) and medial deltoid (1/3 on the line from acromion to lateral epicondyle of the elbow) [14]. The trapezius and deltoid muscles were included to get an estimate of shoulder muscle effort when performing tasks. Electrodes on the iliocostalis muscle were not placed on the smaller participants (n=9), due to space limitations on the back.

Maximum force was measured using a static frame myometer. The frame consisted of a KAP-E Force Transducer (range 0.2 - 2000 N) (Angewandte System Technik, Dresden, Germany) and a height and position adjustable frame (custom made at the VU medical centre, Amsterdam, the Netherlands). The force signal was filtered with a bi-directional 4th order low-pass filter of 30 Hz. Afterwards the measured maximum force signal was converted to joint torque by multiplying the force with the segment length (i.e. moment arm) and additionally resulting torques were also corrected for body weight.

Maximal voluntary isometric contractions (MVICs) were performed to determine maximal joint torques and corresponding sEMG amplitudes. Participant's positions for MVIC measurements were adapted to seated positions so all participants with DMD could perform the measurements. Participants performed two MVIC tasks for each of the following movements: trunk flexion, trunk extension, lateral bending trunk (left and right), shoulder elevation (left and right) and shoulder abduction (left and right). Participants were encouraged to push as hard as they could for 3 seconds. When the maximum force of the MVIC tasks varied more than 10% between the two trials, an additional trial was recorded. A 4th order Butterworth filter (20-450 Hz) was used to filter the sEMG signals, followed by rectification and low-pass filtering (3 Hz) of the signals to obtain the linear envelopes.

The maximum sEMG amplitude for each trunk muscle was taken as the highest amplitude from the four MVIC tasks of the trunk, for the trapezius as the highest amplitude from the shoulder elevation task and deltoid as the highest amplitude from the shoulder abduction task.

Normalized sEMG amplitudes were used to describe the percentage of muscle capacity used during task performance. These were calculated by dividing the sEMG amplitudes during task performance, by the corresponding maximum sEMG amplitudes. Subsequently, average muscle activity of the back muscles (i.e. longissimus and iliocostalis both sides) and average activity of the abdominal muscles (i.e. external oblique both sides) were calculated. If there were more than two missing values, i.e. trials that failed due to inability of the participant to perform the task, or technical errors such as missing signals due to loose electrodes, the average muscle activity was defined as missing value.

All analyses were performed using Matlab R2014b (Math Works, USA) software.

Statistics

Non-parametric tests were used since most of the data were not normally distributed. Median values and interquartile ranges were used to describe the participant characteristics. Wilcoxon rank sum test was used to assess differences between DMD patients and HC and the Kruskal-Wallis test to assess differences between DMD patients with different scores on the Brooke scale.

To test the hypotheses that compensatory trunk ROM would increase with task difficulty, trunk ROM when reaching without weight was subtracted from trunk ROM when reaching with 500 gram object for each individual. Afterwards the change in trunk ROM between DMD patients and HC was assessed with the Wilcoxon rank sum test.

The range of motion is depicted in graphs, where the boxes represent 25th, 50th and 75th percentile, whiskers minimum and maximum non-outlier values and dots indicate outliers (greater than 1.5 times the interquartile range). All statistical analyses were performed using Matlab R2014b (Math Works, USA) and the statistical significance level was set at $\alpha = 0.05$.

RESULTS

Subject demographics.

Participant characteristics are described in Table 1. Of the corticosteroid users, three patients used Deflazacort and the others used Prednisolone. The Vignos classifications of the DMD participants were: 1 (n=1), 2 (n=1), 3 (n=1), 4 (n=1), 5 (n=2), 7 (n=2) and 9 (n=9); and the Brooke classifications: 1 (n=6), 2 (n=6), 3 (n=5). One DMD participant left the assessment before the protocol was finished and was therefore excluded from the analysis. Termination of the measurement was unrelated to the protocol or measurement itself.

Table 1 Participant characteristics

	Healthy		DMD	
	n	median IQR	n	median IQR
Age [years]	25	13.2 [9.4-18.0]	17	13.1 [11.7-15.8]
Gender [male/female]	13/12		17/0	
Weight [kg]	25	48.6 [30.5-63.5]	15	48.0 [40.0-51.5]
Height [cm]	25	160.0 [136.5-171.0]	15	150.0 [145.5-157.0]
Sitting height [cm]	25	62 [50.5-65.6]	12	50.0 [47.8-56.3]
Pain at time of participation [n]	0		0	
Age of diagnosis [years]			16	4 [3-5]
Corticosteroid use [n]	0		15	
Wheelchair confinement indoors [n]	0		10	
Wheelchair confinement outdoors [n]	0		14	
Scoliosis [n]	0		2	

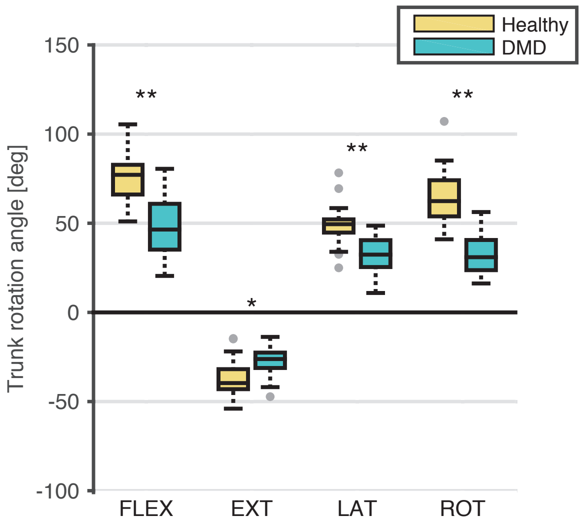

Figure 1 Maximum trunk angle during active trunk movements.

Active range of motion and joint torque

The maximum trunk angles were significantly lower ($p<0.05$) in all movement directions for DMD patients compared to HC (Figure 1). Only trunk axial rotation showed a significant relation ($p=0.014$) with Brooke scale, where smaller angles were seen with a higher Brooke scale (Additional file 1).

DMD patients had significantly ($p<0.01$) lower joint torques compared to HC in all muscle groups and tasks both with and without normalization to body weight (Table 2). However, a significant ($p<0.05$) effect of Brooke scale was only found when correcting the joint torques for body weight, except for the trunk extension torque (Table 2, Additional file 2). Both with and without correction for body weight, trunk torque was already approximately two times smaller in DMD patients with Brooke scale 1 compared to HC.

Performing daily activities

Trunk ROM in one or more movement directions was significantly higher in DMD patients compared to HC in all of the reaching and daily tasks (Figure 2, Additional file 3). Increased lateral bending was seen for all tasks, except for reaching laterally at nearby distance, and increased trunk flexion-extension was seenfor most of the nearby reaching tasks, i.e. reaching at arm length distance. However, the change in trunk ROM with task difficulty (e.g. object weight), was not significantly different

Table 2 Maximum joint torques (in Nm and Nm/kg) of trunk and shoulder.

		Healthy			DMD			p-value HC vs DMD	p-value Brooke scale
		n	median	IQR	n	median	IQR		
Joint torque [Nm]	Trunk (flexion)	25	47.5	[24.5-58.2]	17	20.3	[14.8-25.0]	0.001	0.120
	Trunk (extension)	25	43.9	[19.8-78.6]	16	21.4	[14.3-30.8]	0.003	0.673
	Trunk (lateral bending)	25	44.4	[27.5-65.2]	17	23.6	[17.0-33.1]	0.001	0.355
	Shoulder elevation	25	50.0	[30.1-95.8]	17	18.2	[12.6-24.4]	<0.001	0.244
	Shoulder abduction	25	30.6	[20.0-46.9]	17	11.9	[4.4-14.8]	<0.001	0.019
Joint torque [Nm/kg]	Trunk (flexion)	25	0.94	[0.73-1.09]	15	0.43	[0.34-0.52]	<0.001	0.009
	Trunk (extension)	25	0.81	[0.72-1.25]	14	0.41	[0.31-0.52]	<0.001	0.585
	Trunk (lateral bending)	25	0.99	[0.88-1.14]	15	0.45	[0.39-0.64]	<0.001	0.014
	Shoulder elevation	25	1.10	[0.9-1.76]	15	0.34	[0.26-0.56]	<0.001	0.027
	Shoulder abduction	25	0.69	[0.61-0.8]	15	0.2	[0.08-0.28]	<0.001	0.012

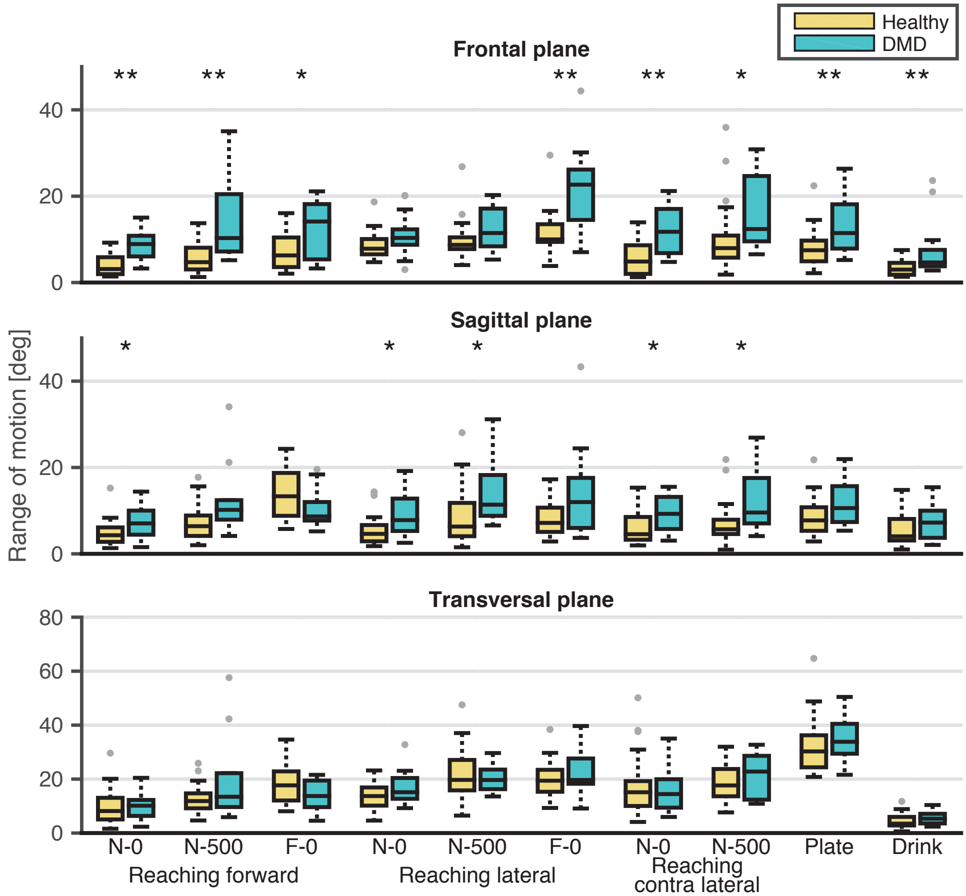

Figure 2 Trunk range of motion in DMD patients and healthy controls when performing daily activities. Abbreviations: N = near, F = far, 0 = without object weight, 500 = 500 gram object weight, * p<0.05, ** p<0.01

between DMD patients and HC (Table 3), except when reaching forward were DMD patients used significantly more trunk flexion-extension movement compared to HC. The change in trunk axial rotation tended to be higher in DMD patients when reaching forward (p=0.061) and contra-laterally (p=0.062).

A significant increase in trunk ROM with Brooke scale was only found for the drinking task (in the frontal plane) (p=0.007) and when reaching contra-lateral with 500 gram object (in the frontal plane) (p=0.025) (Additional file 3).

The direction of movement was largely the same for all DMD participants (Figure 3). The largest variation in movement direction could be seen in flexion-extension when reaching forward. Both flexion and extension movements were made by

Table 3 Change in trunk range of motion (ROM) (in degrees) between reaching with 500 gram object weight and without object weight.

		Healthy		**DMD**		
Trunk ROM direction	Reaching direction	n	median IQR	n	median IQR	p-value HC vs DMD
Flexion-extension	Forward	25	8.5 [3.2-21.1]	9	26.6 [13.4-31.1]	0.017
	Sideward	25	5.8 [2.5-9.8]	8	11.6 [5.1-21.8]	0.303
	Contra-lateral	25	5.6 [2.5-10.8]	8	11.7 [4.1-18.0]	0.231
Lateral bending	Forward	25	4.1 [1.2-8.3]	9	7.3 [2.1-16.8]	0.458
	Sideward	25	7.5 [5.3-11.4]	8	15.6 [4.7-21.8]	0.284
	Contra-lateral	25	3.0 [-2.0-5.0]	8	8.4 [0.4-16.2]	0.125
Axial rotation	Forward	25	4.7 [1.6-6.6]	9	11.0 [3.2-16.5]	0.061
	Sideward	25	8.6 [3.6-15.3]	8	8.4 [3.7-10.7]	0.850
	Contra-lateral	25	5.0 [-2.9-9.0]	8	11.4 [5.8-14.5]	0.062

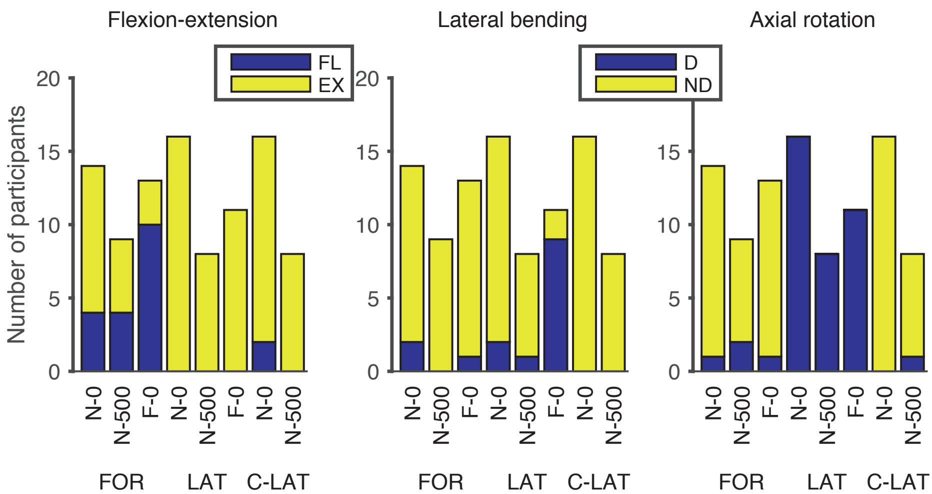

Figure3 Movement direction of the trunk in DMD patients when performing daily activities. Abbreviations: N = near, F = far, 0 = without object weight, 500 = 500 gram object weight, FOR = forward reaching, LAT = reaching laterally, C-LAT = reaching contra-laterally.

the DMD participants, while trunk extension was seen in the other tasks. Lateral bending was mainly performed towards the non-dominant side, in other words opposite to where the arm was lifted for reaching, except for far lateral reaching. Axial rotation was performed towards the dominant side when reaching laterally and towards the non-dominant side when reaching forward and contra-laterally. The movement direction for DMD participants was essentially the same as in the HC.

Normalized muscle activity was significantly higher in all muscles and all tasks for DMD patients compared to HC (Figure 4, Additional file 4). Static sitting (without back or armrests) already required approximately twice as much of trunk muscle capacity in DMD patients than in HC. The ability to perform a task was related to the percentage of muscle capacity used. This could for example be seen when comparing reaching forward without object and with a 500 gram object (Figure 4, Additional file 4). All DMD patients with Brooke scale 1 were able to perform the task with a 500 gram object, but only half of the DMD patients with Brooke scale 2 and none of the subjects with Brook scale 3 could. However, those patients with Brooke scale 2 needed around 100% of their back and arm muscle capacity to execute the task.

DISCUSSION

This study provides new insights in the role of trunk movements and used muscle capacity in DMD patients when performing seated tasks. During arm tasks the trunk shows a larger range of motion in DMD patients compared to healthy controls, combined with increased normalized trunk muscle activity. This reflects that due to compensatory movement, demands on trunk muscles are increased which is compounded by trunk muscle weakness.

Both maximum active trunk ROM and maximum trunk joint torque were significantly decreased in DMD patients compared to HC, indicating that their overall trunk capacity is already less compared to HC. Although this finding is not surprising, this is the first study to show it in a quantitative manner. However, the limitations found in maximum ROM are unlikely to result in restrictions when performing tasks such as tested here, because the maximum trunk ROM (Figure 1) was less than generally used to perform daily tasks (Figure 2).

Interestingly, we found that boys in early disease stages (e.g. Brooke scale 1) already showed lower trunk joint torque compared to HC. Additionally, trunk joint torque (in Nm) did not significantly decrease with Brooke scale, while shoulder abduction torque did. The latter was also found in previous research [4]. This could indicate that arm function (i.e. Brooke scale) is decreasing first or that the decrease in trunk function is independent of the decrease in arm function. However, as bodyweight

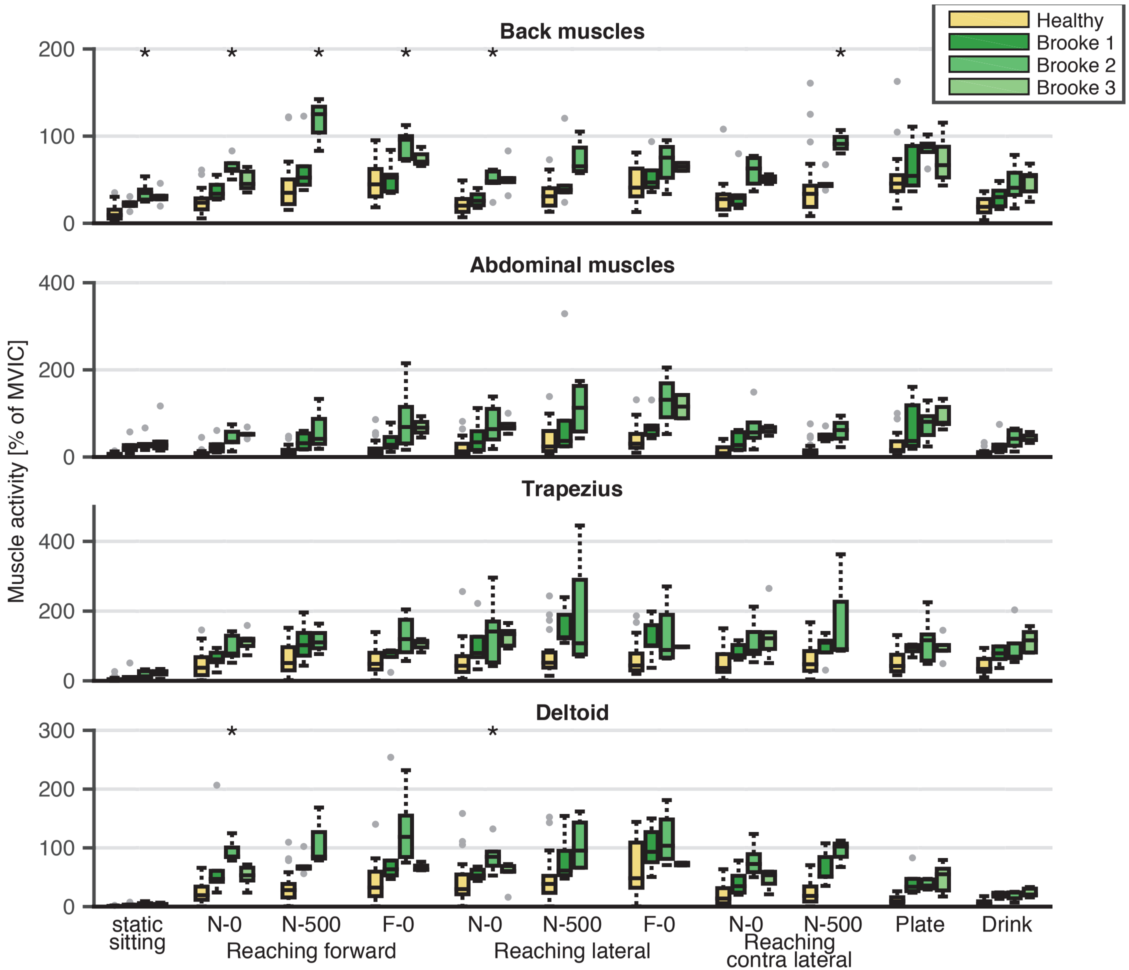

Figure 4 Normalized muscle activity in healthy controls and DMD patients with different Brooke scales.
Abbreviations: N = near, F = far, 0 = without object weight, 500 = 500 gram object weight, * p<0.05 between Brooke scales

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increases with age, and joint torque does not increase with body weight, function decreases [15]. Indeed, when we corrected trunk joint torque for body weight in DMD patients, we found a significant decrease with Brooke scale, implying that functional trunk strength does decrease with disease stage.

Increased trunk lateral bending and/or flexion-extension was found in DMD patients in all tasks. It is remarkable that this was even found when reaching within arm length distance, since the reaching distance could be shorter for patients. DMD patients likely reduce shoulder and upper arm muscle activity using increased trunk lateral bending towards the non-dominant side to reduce shoulder flexion and abduction. By leaning towards the non-dominant side, the dominant shoulder and arm are automatically positioned higher so less shoulder muscle effort is needed to lift the arm for reaching. Opposite to what was initially expected, the increased ROM in trunk flexion-extension was mainly in extension direction. This could mean that the DMD participants lean backwards in order to keep balance, as is also seen in patients with spinal cord injuries [16], or that patients extend their spine from an initially more slumped posture. This also positions the shoulder higher to reduce shoulder muscle effort and allows for a greater ROM of the shoulder [17].

These compensatory trunk movements are likely crucial to accomplish a task when arm function is insufficient. Compensatory trunk movements are also seen in children with cerebral palsy when performing daily tasks and were related to decreased upper extremity function [18, 19]. Unexpectedly, we did not find a significantly larger increase in compensatory trunk movements with task difficulty (e.g. object weight) in DMD patients compared to HC. It could be that patients already use the most optimal strategy in the easiest tasks (e.g. reaching nearby without weight) and therefore further increasing trunk movements is not beneficial. Alternatively, trunk function could limit increasing the compensatory movements as muscle activity levels did approach the maximum values. However, the median change in trunk ROM was often twice as high in DMD patients compared to HC. It is therefore also possible that we did not find a significant increase due to lack of statistical power, also due to the fact that DMD patients with less good arm function could not perform the more difficult task. No significant differences were found in trunk ROM between patients with different scores on the Brooke scale, although it was expected that compensatory trunk ROM would increase with Brooke scale. This is likely caused by small numbers of subjects in all categories.

Normalized muscle activity was significantly higher in patients with DMD compared to HC for all muscles and all tasks. Normalized muscle activity also increased until the task could not be performed. Despite possible overestimation due to non-maximum MVIC, we found that normalized back muscle activity was around 100% when the maximum arm muscle (e.g. deltoid and trapezius) activity was reached.

This indicates that back muscle function plays a more important role than thought, so the arm might not be the only limiting factor accomplishing tasks. It is likely that compensatory trunk movements are limited by increasing back muscle activity with disease progression, due to which patients lose the ability to accomplish the task.

The percentage of trunk muscle capacity used when sitting upright was already two times higher in patients with Brooke scale 1 compared to HC and this normalized activity level is even higher when performing tasks. This indicates early trunk muscle weakness in relation to motor function, which contrasts with previous studies indicating that trunk function is good in the ambulatory phase [5, 6]. When a higher percentage of the maximum muscle capacity is used, this leads to faster development of fatigue and possibly to overloading of the muscles [20]. Clinicians should take this increased muscle activity into account for function assessment and development of interventions. Proper seating, back rests or the use of other trunk supportive devices can reduce trunk muscle fatigue during the day [21]. However, it is important that patients are still able to move their trunk, despite increased activity, to accomplish tasks independently. Also physical muscle strength training might reduce fatigability [22].

There are several limitations to this study. The sample size was small when subcategorizing the DMD patients based on Brooke scale. Therefore, the power to detect differences in trunk ROM between these categories may have been too low. Furthermore, only patients with relatively good arm function could perform the more difficult tasks, which reduced statistical power. The control group was not completely matched with the DMD patients in terms of gender. However, there were no significant differences between boys and girls in the HC group. The normalized trunk muscle activity was based on standardized seated MVIC tasks, which probably does not correspond to the actual maximal values for trunk muscle activity. As a consequence, 100% muscle activity does not necessarily correspond to the maximum capacity, but is likely an overestimation. However, since the MVIC tasks were standardized across all participants, it showed that DMD patients used significantly more muscle activity compared to healthy subjects. Reaching distances were based on the distances that could be reached without moving the trunk. Consequently, the reaching distances varied between subjects and groups. In general, patients with weaker arm muscles reached towards shorter distances, however even though the distance was shorter they showed increased trunk movement compared to HC. Lastly, as described before [10], reaching distance and height were set based on subjects' sitting posture. Small changes in posture could already influence the distance and height and cause variability between tasks within and between subjects. Since we were interested in self-selected movements of the trunk, we did not choose to standardize sitting posture.

CONCLUSION

Trunk capacity (joint torque and active ROM) is reduced in DMD patients compared to HC. They used compensatory lateral bending and trunk flexion-extension movements to accomplish daily tasks, in combination with increased normalized muscle activity. The compensatory movements did not significantly increase more with task difficulty (e.g. increasing object weight) compared to HC and also did not increase with Brooke scale, although differences could be seen. Percentage of muscle capacity used was higher in patients with DMD for all muscles and in all tasks, which could result in early development of muscle fatigue. Clinical interventions are necessary to reduce the muscle fatigue, like development of dynamic assistive devices or implementing proper seating. However, (compensatory) trunk movements should not be restricted because this will likely lead to limitations in accomplishing tasks independently.

SUPPLEMENTARY MATERIAL

See online version of this article at https://doi.org/ 10.1186/s12984-019-0515-y or scan the QR code.

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CHAPTER 5

PATIENTS WITH SPINAL MUSCULAR ATROPHY USE HIGH PERCENTAGES OF TRUNK MUSCLE CAPACITY TO PERFORM SEATED TASKS

Accepted: Peeters LHC, Janssen MMHP, Kingma I, van Dieën JH, de Groot IJM.

American Journal of Physical Medicine and Rehabilitation.

Abbreviations:

HC	Healthy Controls
HC_6y	6 years old HC participants
MVIC	Maximum Voluntary Isometric Contractions
ROM	Range of motion
sEMG	Surface Electromyography
SMA	Spinal Muscular Atrophy
SMA_6y	6 years old SMA participant
UE	Upper Extremity

ABSTRACT

Objective: To investigate trunk function during seated upper extremity tasks in patients with spinal muscular atrophy (SMA) type 2 and 3.

Design: 17 persons with SMA and 15 healthy controls (HC) performed several tasks when sitting unsupported, such as reaching (and placing) forward and sideward. Joint torque and muscle activity were measured during maximum voluntary isometric contractions (MVIC). Three-dimensional kinematics and normalized muscle activity were analyzed when performing tasks.

Results: Trunk joint torques were significantly decreased, approximately 45%, in patients with SMA compared to HC. Active range of trunk motion was also significantly decreased in all directions. When performing tasks, the average back muscle activity was 27% and 56% of MVIC for respectively HC and SMA, and for abdominal muscles respectively 10% and 44% of MVIC. Trunk range of motion did not differ when performing daily tasks.

Conclusion: The trunk of patients with SMA is weaker compared to HC, reflected by reduced trunk torques and decreased active range of motion. Additionally, patients with SMA use high percentages of their trunk muscle capacity to perform tasks. Clinicians should take this into account for intervention development, because using high percentages of the maximum muscle capacity results in fatigue and muscle overloading.

BACKGROUND

Spinal muscular atrophy (SMA) is characterized by progressive degeneration of motor neurons in the spinal cord, leading to muscle weakness and atrophy [1]. As a result patients experience limitations in performing daily activities independently [2, 3]. Patients are categorized based on maximum acquired milestones and disease onset, but clinically it is more a gradual scale of functional abilities [4, 5]. The natural course of children with SMA is characterized not only by weakness of upper and lower extremity muscles, but also by (severe) weakness in the trunk leading to scoliosis at young age [4]. However, the natural course is now changing due to effective treatment with Spinraza [6].

When performing seated activities, the trunk plays an indispensable role as it interacts with upper extremity (UE) movement as part of the kinematic chain and it provides a stable base for UE movements [7-10]. Only a few studies describe trunk function in patients with SMA. Trunk muscle force and axial function seems to be less for patients with SMA type 2 compared to type 3 [4, 11]. But literature contradicts whether axial function decreases with age. Vuillerot, et al. [5] found only a decline in axial motor function for SMA type 2 patients based on the Motor Function Measure dimension 2, whereas Wadman, et al. [11] found a decline in motor function for all SMA types based on the Hammersmith Functional Motor Score. Both measures are not solely based on axial function (i.e. upper or lower extremity function also influences the score), which might explain differences in findings. It is remarkable that so little research has been done concerning trunk function although scoliosis secondary to muscle weakness is a major problem in childhood for patients with SMA [4].

Therefore, the aim of this study was to investigate trunk function and its relation with upper extremity movements when performing seated upper extremity tasks in patients with SMA types 2 and 3. We hypothesized that maximum trunk torques and maximum active range of movement are reduced in patients with SMA (types 2 and 3) compared to healthy controls, while trunk movements and muscle activity levels when performing daily tasks are increased to compensate for reduced upper extremity function and trunk muscle strength.

METHODS

Participants

Seventeen people with SMA and fifteen healthy controls (HC) participated in this study. Participants were included if they were older than 6 years, able to bring their hand to the mouth and could sit independently (without back or arm rests) for at least 5 minutes. Patients also needed to have a genetically confirmed diagnosis of SMA. Participants were excluded if they had (other) diseases affecting arm, trunk or head movements.

The fifteen HC were a sample of the HC group described previously [9]. Since our participants with SMA were mainly adults, we selected only HC above the age of 12 years to eliminate the maturation effect (e.g. coordination between trunk and arm movements changes in children up to the age of 10 years) as previously described [9, 12]. For the same reason, the 6-year old participant with SMA (SMA_6y) will be described and compared separately with 3 HC of 6 years (HC_6y) as a case study.

Participants with SMA were recruited through advertisements by patient organizations (Spierziekten Nederland and Prinses Beatrix Spierfonds) and through the Radboudumc outpatient clinic Nijmegen. HC were recruited from local primary schools, high schools and university. Written informed consent was given by all participants prior to participation. The study was approved by the medical ethics committee Arnhem-Nijmegen (NL58988.091.16) and all data were handled according to the guidelines of good clinical practice. This study conforms to all STROBE guidelines and reports the required information accordingly (see Supplementary Checklist).

Procedures

We used the same procedure as that employed in a previous study with healthy children [9]. All participants were seated on a height adjustable chair without back- or armrests. The sitting height was adjusted so that the knees were flexed 90 degrees and both feet were flat on the ground.

First, to determine maximum trunk range of motion, participants were asked to perform a maximum active flexion movement of their trunk from a seated position, immediately followed by a maximum active extension movement of their trunk (keeping both feet on the ground). The same was done for maximum axial rotation and lateral bending. Thereafter, several reaching (and placing) tasks were performed with a preferred hand at shoulder height: reaching forward, sideways and contra-laterally. Reaching distance and object weight were varied, resulting in the following combinations for forward, lateral and contra-lateral reaching: nearby-0 gram ("N-0"), nearby-500 gram ("N-500"), far-0 gram ("F-0"). Contra-lateral reaching was not performed at a far distance. Nearby was defined as the distance that could be reached with the arm without moving the trunk (i.e. 100% arm length for HC, but could be closer for SMA) and far was defined as 133% of arm length when possible, otherwise as maximum reaching distance. Furthermore, subjects were asked to perform two daily tasks: displace a porcelain plate (circa 600 grams) from left to right on a table with both hands ("Plate") and bring a cup of 200 grams to the mouth ("Drink"). No instructions were given on how to perform the tasks.

Outcome measures

Data acquisition and analysis were similar as used in a previous study with healthy children and patients with Duchenne muscular dystrophy, and will be described briefly [9, 13].

Participant characteristics

Patient characteristics were recorded based on self-reports and included age, weight, height, arm preference, age of diagnosis (if applicable), wheelchair confinement, pain in upper body at time of participation, scoliosis and spinal fusion surgery. Sitting height was measured and, for patients with SMA, the Vignos lower extremity scale [14] and Brooke upper extremity scale [15] were used for clinical assessment of respectively leg and arm function.

Three dimensional motion analysis

An optical motion capture system (Vicon Motion Systems Ltd, Oxford, UK) was used to record 13 single reflective markers, which were placed on the skin to define positions and orientations of the trunk and pelvis during task performance. Markers on the spinous processes of C7, T6, T12 and L3, a laterally placed marker at level L1/L2, jungular notch and xiphoid process of the sternum defined three trunk segments (upper thoracic, lower thoracic and lumbar) [9]. The pelvis markers were placed according to the Vicon Plugin-Gait model with two additional markers on the iliac crest. The markers divided the trunk initially into three segments, because the trunk cannot be seen as rigid segment. However to make the data more concise, we decided to report the trunk movement as one segment (i.e. summation of the three segment angles and pelvis) in this paper. Distribution of movement patterns over the individual trunk segments was essentially the same among HC and patients with SMA without spinal fusion surgery.

All kinematic data were filtered with a bi-directional 4^{th} order Butterworth low-pass filter (cutoff frequency of 6 Hz). Trunk joint angles are expressed relative to the global coordinate system.

In all three planes of movement, maximum trunk joint angles were determined when performing the active range of trunk motion tasks. For the reaching tasks and daily tasks, the trunk range of motion (ROM) between the start and end of the task (e.g. time where wrist velocity exceeded/got below 5% of its peak velocity) was determined.

Joint torque and surface electromyography

Surface electromyography (sEMG) (Zerowire EMG, Aurion, Italy) was used to measure muscle activity at a sample rate of 1000 samples/s. Electrodes were placed on the following muscles on both sides of the body: iliocostalis (6 cm from spinous processes of L1), longisimus (3 cm from spinous processes of L3), external oblique (3 cm from axillae midline at height of umbilicus) and medial deltoid (1/3 on the line from acromion to lateral epicondyle of the elbow) [16, 17]. The deltoid muscles were included to get an estimate for shoulder muscle effort when performing tasks. Electrodes on the iliocostalis muscle were not placed in two smaller participants with SMA, due to space limitations on the back.

Maximum force was measured using an adjustable static frame myometer with a KAP-E Force Transducer (range 0.2 - 2000 N) (Angewandte System Technik, Dresden, Germany). The force signal was sampled at 1000 samples/s and filtered with a bi-directional 4^{th} order low-pass filter of 30 Hz. Afterwards the maximum joint torque was calculated by multiplying the measured force with the segment length (i.e., moment arm).

Maximal voluntary isometric contractions (MVICs) were performed to determine maximal joint torques and corresponding sEMG amplitudes. Participants' positions for MVIC measurements were adapted to seated positions so all participants with SMA could perform the measurements. Two MVIC efforts were performed for 3 seconds by the participants for each of the following directions: trunk flexion, trunk extension, lateral bending trunk (left and right) and shoulder abduction (left and right) (Figure 1). When the maximum force of the MVIC task varied more than 10% between the two trials, an additional trial was recorded. Because patients with SMA are easily fatigued, it was not feasible to perform many MVIC trials.

A 4^{th} order Butterworth filter (20-450 Hz) was used to filter the sEMG signals, followed by rectification and low-pass filtering (3 Hz) of the signals to obtain the linear envelopes. The maximum sEMG amplitude for each trunk muscle was taken as the highest amplitude from the four MVIC tasks of the trunk and maximum deltoid sEMG amplitude as the highest amplitude from the shoulder abduction task. Normalized sEMG amplitudes (maximum sEMG amplitude during task divided by the maximum MVIC amplitude for that muscle) were used to describe the percentage of muscle capacity used during maximum active range of motion and daily tasks. Subsequently, average normalized muscle activity of the back muscles (i.e. longissimus and iliocostalis both sides) and average normalized activity of the abdominal muscles (i.e. external oblique both sides) were calculated. If more than two values were missing, due to inability of the participant to perform the task or due to technical errors such as missing signals as result of lose electrodes, the average normalized muscle activity

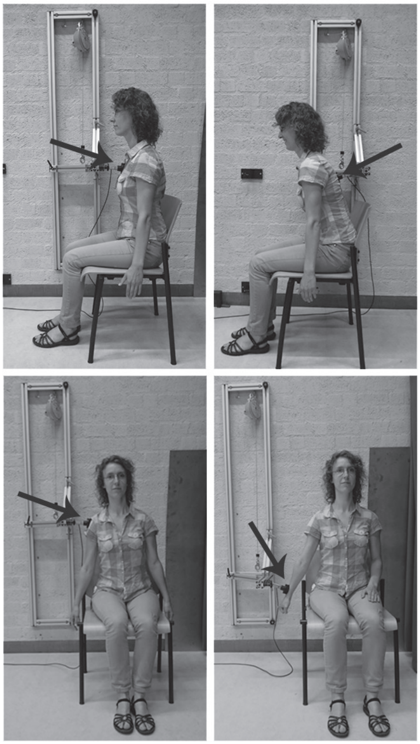

Figure 1 Participant's positioning for the maximum voluntary isometric contraction tasks with the static frame myometer (indicated by arrow). Top row: trunk flexion and extension; bottom row: lateral bending and shoulder abduction.

was defined as missing value.

All analyses were performed using custom scripts in Matlab R2014b (MathWorks, Natick, USA).

Statistics

Median values and interquartile ranges are used to describe the data since the data were not normally distributed. Wilcoxon rank sum tests were used to assess differences between patients with SMA and HC. The range of motion is depicted in graphs, where the boxes represent 25th, 50th and 75th percentile, whiskers minimum and maximum non-outlier values and dots indicate outliers (greater than 1.5 times the interquartile range). All statistical analyses were performed using Matlab R2014b and the statistical significance level was set at $\alpha = 0.05$.

RESULTS

Participant characteristics

Participant characteristics are described in Table 1. Participants who reported pain at time of the measurement, had mainly (chronic) shoulder pain which did not have major impact on their mobility in daily life. None of the participants used medication described as affecting SMA, except for one participant who used Mestinon. The 6-year old, type 2 SMA participant is not included in the table. His Vignos scale was 9, Brooke scale 1 and he had no scoliosis.

Three participants with SMA were not able to sit unsupported and perform tasks at the same time, and were excluded from the kinematic and muscle activity analysis. They all had SMA type 2, spinal fusion surgery and scored 3, 5 and 6 on the Brooke scale. One of these subjects wore a trunk brace, others did not.

Table 1 Participant characteristics

	Healthy controls			SMA patients		
	n	median	IQR	n	median	IQR
Age [years]	15	18.1	[14.4-20.4]	16	43.5	[25.5-57.2]
Gender [male/female]	7 / 8			11 / 5		
Weight [kg]	15	60.0	[51.1-66.5]	16	74.5	[56.6-88.1]
Height [cm]	14	170.5	[166.0-174.0]	15	176.0	[167.0-178.8]
Sitting height [cm]	15	64.5	[62.1-67.5]	14	63.0	[52.0-69.0]
Pain at time of participation [n]	0			5		
Type of SMA [type 2/type3]				5 / 11		
Age of diagnosis [years]				16	3.5	[2-16]
Wheelchair confinement [n]				14		
Scoliosis [n]	0			9		
Spinal fusion surgery [n]	0			6		
Vignos lower extremity scale	1	2	9			
[n]	1	1	14			
Brooke upper extremity scale	1	2	3	4	5	6
[n]	3	3	8	0	1	1

Joint torque

Trunk joint torques were significant lesser ($p<0.01$) for patients with SMA compared to HC in all directions, with median values slightly below 50% of HC (Table 2). Median shoulder torques were below 25% of HC in patients with SMA. SMA type 2 patients seemed weaker compared to type 3 patients, although the numbers were too small for statistical testing.

Active range of motion tasks

The numbers of participants with missing values in trunk ROM and muscle activity outcomes are shown in Table 3. Maximum active trunk angles were significantly lower ($p<0.01$) in all directions in patients with SMA compared to HC (Figure 2A). Median trunk flexion angle was reduced the most, approximately by 58°, followed by axial rotation (36°), extension (27°) and lateral bending (24°). There was no significant difference between lateral bending and axial rotation to the dominant or non-dominant side.

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Normalized muscle activity levels when flexing and extending were not different between patients with SMA and HC for the muscles primarily counteracting gravitational moments (e.g. back muscles for trunk flexion, abdominal muscles for trunk extension) (Figure 2B). However, there was a significant increase ($p<0.01$) in normalized antagonistic activation for patients with SMA, i.e. the abdominal muscles for flexion (up to 29% MVIC), back muscles for extension (up to 24% MVIC) and ipsilateral back muscles for lateral bending (up to 39% MVIC). Normalized muscle activity of the ipsilateral back and abdominal muscles were also significantly greater ($p<0.05$) for axial rotation (up to 24% MVIC) in patients with SMA.

Table 2 Maximum trunk joint torque (in Nm) in four directions.

	Healthy controls			SMA			
	n	median	IQR	n	median	IQR	P-value HC/ SMA
Trunk (flexion)	15	54.4	[47.8-70.7]	13	23.7	[20.7-34.4]	0.001
Trunk (extension)	15	59.6	[46.8-84.3]	13	25.9	[11.2-47.7]	0.001
Trunk (lateral bending D)	15	66.4	[52.7-86.9]	13	29.2	[17.7-42.8]	0.005
Trunk (lateral bending ND)	15	63.1	[47.5-75.0]	13	31.6	[16.2-42.7]	0.004
Shoulder abduction (D)	15	47.9	[35.4-57.0]	16	11.3	[5.1-17.4]	<0.001
Shoulder abduction (ND)	15	42.1	[31.0-54.5]	14	9.8	[4.9-13.0]	<0.001

Abbreviations: D = towards dominant side; ND = towards non-dominant side

Table 3 Number of participants (SMA/HC) who accomplished a task and included data for trunk kinematics, back and abdominal muscle activity.

tasks		Task accomplished	Trunk kinematics	Back muscle activity	Abdominal muscle activity
Maximum active ROM					
	Flexion	13/15	11/15	12/15	13/15
	Extension	13/15	11/14	12/15	13/15
	Lateral ben-ding	13/15	12/15	12/15	12/11
	Axial rotation	13/15	13/15	12/15	11/11
Reaching forward					
	N-0	13/15	13/15	12/15	13/15
	N-500	7/15	7/15	7/15	7/15
	F-0	9/15	8/15	8/15	9/15
Reaching lateral					
	N-0	11/15	10/15	10/15	11/15
	N-500	7/15	7/15	7/15	7/15
	F-0	7/15	7/15	7/15	7/15
Reaching contra-lateral					
	N-0	13/15	13/15	12/15	13/15
	N-500	7/15	7/15	7/15	7/15
	Plate	13/15	13/15	12/15	13/15
	Drink	12/15	9/15	11/15	12/15

Abbreviations: ROM = range of motion, N = near, F = far, 0 = without weight, 500 = 500 gram object

Performing daily tasks

The number of participants who could accomplish a task and the numbers of participants with missing values in trunk ROM and muscle activity outcomes are shown in Table 3. In general, no differences were seen in trunk ROM between patients with SMA and HC when performing daily tasks (Figure 3).

Normalized muscle activity levels for back and abdominal muscles were significantly greater ($p<0.01$) in patients with SMA compared to HC for all tasks, except reaching laterally far (Figure 4). Unsupported static sitting required already three time as much normalized trunk muscle activity for patients with SMA. When performing the daily tasks, the average back muscle activity was 27% of MVIC for HC and 56% of MVIC

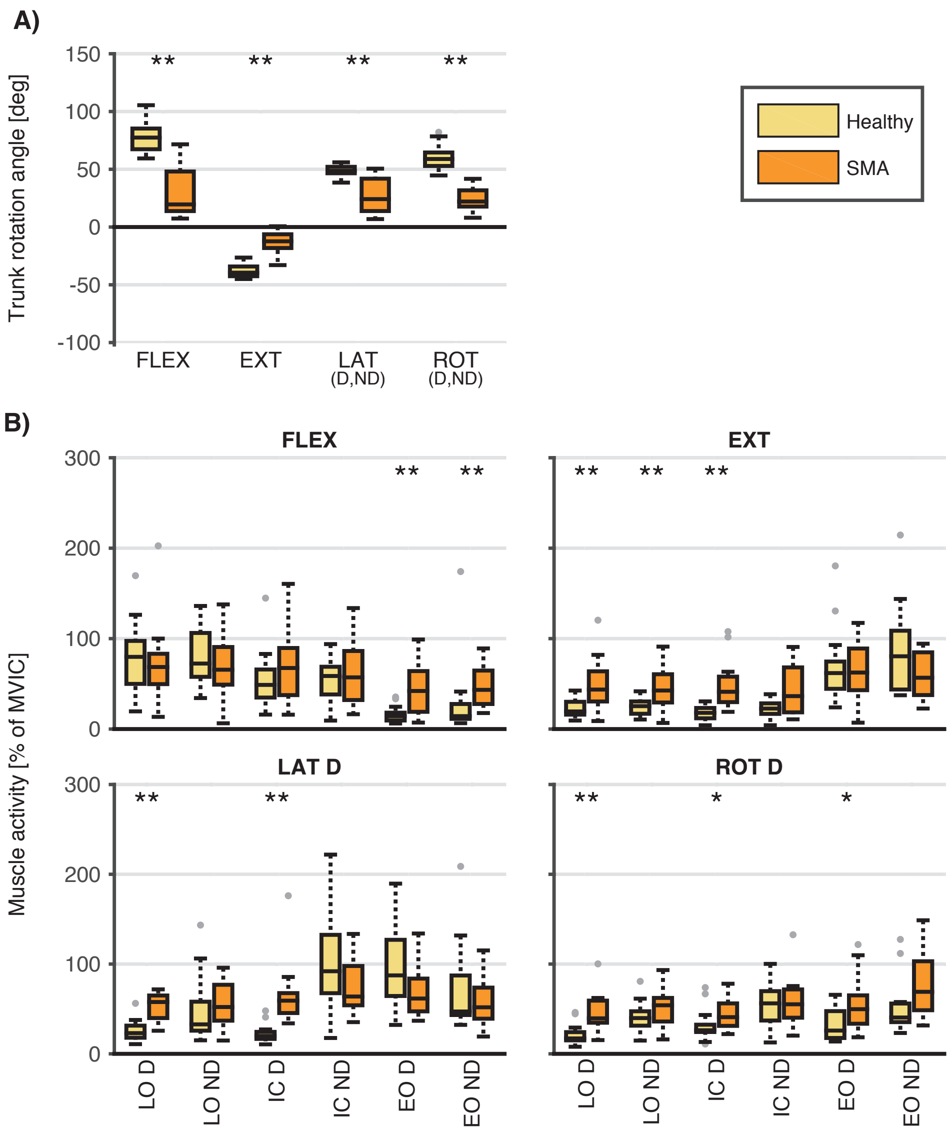

Figure 2 Rotation angles and muscle activity for active range of trunk motion tasks. A) Maximum trunk rotation angle when performing a maximum flexion (FLEX), extension (EXT), lateral bending (LAT) or axial rotation (ROT) movement. Lateral bending and axial rotation are mean values towards dominant and non dominant side. B) Maximum muscle activity levels when performing a maximum flexion, extension, lateral bending to dominant side and axial rotation to dominant side movement. Abbreviations: LO = longissimus, IC = iliocostalis, EO = external oblique, D = dominant side, ND = non dominant side, * p<0.05, ** p<0.01

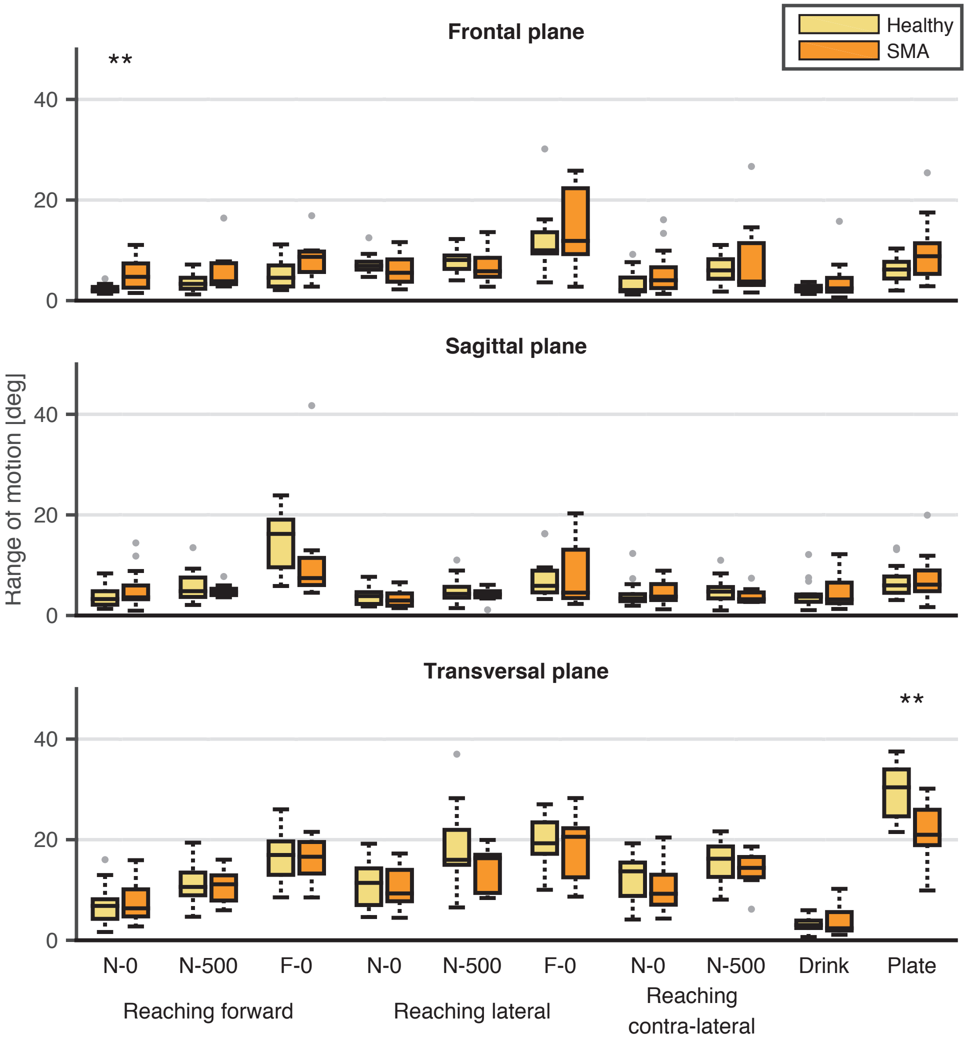

Figure 3 Trunk ROM in patients with SMA and healthy controls when performing tasks.
Abbreviations: N = near, F = far, 0 = without weight, 500 = 500 gram object, * p < 0.05, ** p < 0.01

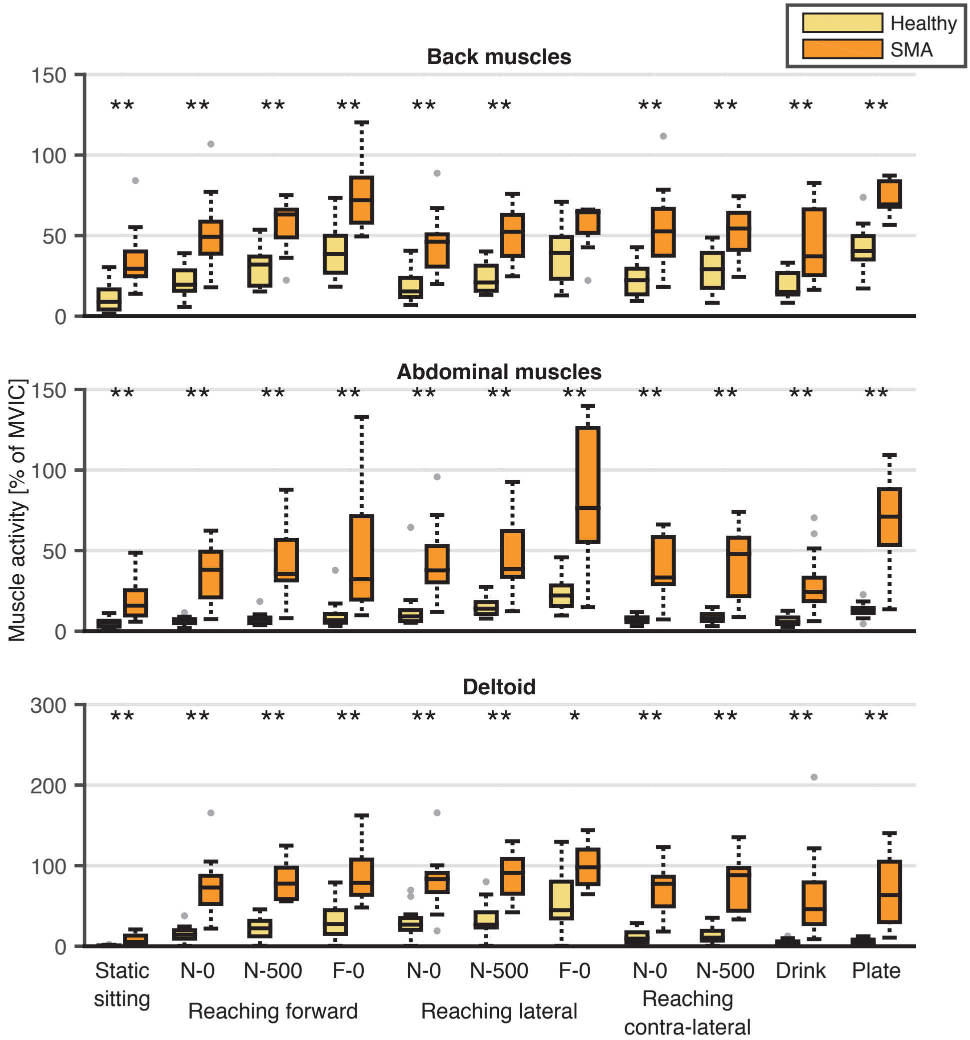

Figure 4 Muscle activity in patients with SMA and healthy controls when performing tasks. Abbreviations: N = near, F = far, 0 = without weight, 500 = 500 gram object, * $p < 0.05$, ** $p < 0.01$

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for SMA, and the average abdominal muscle activity was respectively 10% of MVIC and 44% of MVIC. In addition, median muscle activity for the deltoid muscle was around 100% MVIC and was significantly greater ($p<0.05$) compared to HC.

Case 6 years old participant

In general, little differences were found between SMA_6y and HC_6y. Both joint torque and maximum active range of trunk motion were comparable between the 6 years old participants. Trunk ROM of SMA_6y was different from the HC_6y in half of the daily tasks. However, both increased and decreased ROM was seen, and in the majority of tasks the difference was less than 3 degrees. Variability in normalized muscle activity for HC_6y was too large to reliably compare with SMA_6y.

DISCUSSION

This is the first study describing trunk function in SMA in relation to the performance of upper extremity tasks. Demand on trunk muscles is high when performing such tasks, reflected by increased normalized muscle activity levels as hypothesized, but in contrast with our hypothesis this occurred without an increased trunk range of motion.

Trunk joint torque was decreased in patients with SMA compared to HC with at least a factor two in median value. Additionally, SMA type 2 patients seemed weaker in trunk torque compared to type 3, as was also found previously [4]. On the other hand, the large interquartile ranges indicate a gradual scale in trunk function, which is in line with the fact that SMA shows a range of functional abilities rather than absolute differences between types of SMA [5]. More patients are needed to confirm whether there is a difference between types or that it is a graduate scale.

Maximum active trunk ROM was limited in patients with SMA compared to HC in all directions. To perform the ROM tasks, both groups used a comparable percentage of their maximum muscle capacity for the muscles counteracting gravitational moments in flexion, extension and lateral bending movements. This indicates that patients with SMA achieve a lower maximum ROM when using similar muscle effort of the counteracting gravitational muscles as HC. This is not surprising, since the maximum absolute muscle activity is much less for patients with SMA due to loss of motor neurons. A lower maximum absolute muscle capacity results in less force generating capacity, as reflected in the decreased joint torques.

When performing reaching and daily tasks, patients with SMA used a greater percentage of their maximum trunk muscle capacity compared to HC, although trunk movement did not increase. We expected to find increased trunk movement to compensate for reduced arm function, as for example was visible in patients with Duchenne muscular dystrophy [13]. But, although deltoid muscle activity level was close to 100% of MVIC, trunk ROM did not increase. As a consequence, patients will be restricted in their workspace and therefore in performing daily activities. The fact that patients with SMA did not increase their trunk ROM, although normalized shoulder muscle activity was very high, suggests that patients with SMA need more of their trunk muscle capacity to maintain stability in order to perform the upper extremity movements [18].

To gain more insight in mechanisms underlying the increased normalized muscle activity when performing the reaching and daily tasks, we analyzed the absolute muscle activity. This showed similar absolute muscle activity levels of the back muscles, indicating comparable back muscle activation during task performance in SMA and HC (in combination with comparable trunk ROM). Noteworthy, this still resulted in increased percentages of normalized muscle activity in patients with SMA since the absolute maximum muscle activity was decreased. On the other hand, the absolute abdominal muscle activity was significantly increased in patients with SMA, which could indicate co-contraction of the abdominal muscles during task performance and would support the hypothesis above. The co-contraction can be caused by recruitment of more motor units needed to generate enough muscle force to maintain trunk stability and/or recruitment of larger motor units due to re-innervation in SMA [19].

Using increased percentages of the maximum muscle capacity and co-contraction causes earlier development of fatigue and increased risk of muscle overloading [18, 20]. Since scoliosis is related to muscle weakness and fatigue, clinicians should pay high attention to trunk function in children with SMA [21]. But also in general for functional assessment and development of interventions, there should be more awareness for the great loads on trunk muscles required to perform simple manual tasks. Interventions to reduce muscle fatigue during the day can be applied, like proper seating, use of trunk supportive devices, or physical muscle strength training to reduce fatigability [22, 23]. Rigid trunk orthoses are not recommended, because these restrict important trunk movements that are necessary to perform daily tasks. Additionally, being able to move could also prevent the muscles from degenerating faster due to disuse [3, 24]. New supportive devices that allow movement and reduce load on the trunk are needed.

For the first time in patients with SMA, a quantitative insight in trunk function was obtained. The results were consistent with clinical experience on trunk function and can therefore support clinical decision making. Furthermore, the method used in this study gives opportunities to evaluate interventions in a quantitative manner in the future. Treatment with for example Spinraza is currently evaluated with the use of the Hammersmith Functional Motor Scale, but this does not discriminate between different body segments and does not give insight in the benefits for performing activities of daily living [6, 25].

This study has several limitations. First, while we covered a broad range of the clinical spectrum of SMA, it was statistically not possible to compare for example SMA type 2 or type 3 patients, or patients with or without spinal fusion surgery due to the small sample size. It would be interesting to investigate in more detail how differences between subtypes affect task performance. Secondly, the control group was not age matched with the patients with SMA. This might have had an effect on the maximum joint torque and maximum active trunk ROM, as muscle strength and joint flexibility decrease with ageing (starting around 50 years) [26, 27]. However, differences found between the HC and patients with SMA were very high and cannot be solely attributed to age. Furthermore, the reported ROM values during the maximum ROM tasks are active ranges based on unsupported seating and it should be noted that several participants reported that they were afraid of falling when moving further. Lastly, the percentages presented for normalized muscle activity are likely an overestimation, since standardized MVIC tasks were performed from a seated position which likely resulted in lower absolute maximum muscle activity signals. However, this position was chosen so patients could perform the MVIC tasks and because it corresponded with the position in which the movement tasks were performed.

In conclusion, due to degeneration of motor neurons, patients with SMA need a greater percentage of their maximum muscle capacity to generate the same amount of force as HC. This study was the first to quantify the effects of this in performance of seated tasks. Maximum trunk joint torque and active trunk ROM were significantly reduced in patients with SMA. Further, increased normalized trunk muscle activity, without increased trunk ROM, was seen when performing daily tasks. Co-contraction of the trunk muscles is very likely present. This indicates that patients with SMA use more of their muscle capacity to maintain trunk stability compared to healthy controls. Clinicians should take trunk function into account when assessing function and interventions, as using a high percentage of the maximum muscle capacity may result in fatigue and muscle overloading. On the other hand, one must bear in mind that restrictions in trunk movement will likely cause limitations in accomplishing tasks independently and might accelerate muscle decline due to disuse.

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CHAPTER 6

SUMMARY AND GENERAL DISCUSSION

Abbreviations:

CP	Cerebral Palsy
DMD	Duchenne Muscular Dystrophy
HC	Healthy Controls
MFM	Motor Function Measure
MRI	Magnetic Resonance Imaging
MVIC	Maximum Voluntary Isometric Contraction
NMD	Neuromuscular disorders
ROM	Range of motion
SCI	Spinal Cord Injury
sEMG	Surface electromyography
SMA	Spinal Muscular Atrophy
TCMS	Trunk Control Measurement Scale
UE	Upper Extremity

SUMMARY

Trunk and head control are indispensable when performing seated upper extremity tasks. The trunk interacts with the upper extremities (UE) and the head as part of a kinematic chain and it provides a stable base for performing voluntary UE and head movements [1-3]. The trunk can become impaired due to muscle weakness (e.g. Duchenne muscular dystrophy (DMD) or spinal muscular atrophy (SMA)) or central neurological disorders (e.g. cerebral palsy (CP) or spinal cord injury (SCI)). Impairment of the trunk in (early) childhood can affect motor development and it increases the risk of developing spine deformities due to muscle weakness [4]. In turn, spine deformity also affects trunk movement and stability [4].

Although the trunk plays a vital role during seated UE tasks, little research has been done concerning trunk function in patients with DMD and SMA. It is of utmost importance to gain more insight in the interaction between trunk, UE and head when performing seated tasks, since most patients with DMD or SMA who are symptomatic in early childhood will not be able to walk once they have reached adulthood. Additionally, the scarce literature on these interactions in typically developing children only focuses on the trunk as one rigid segment and ignores the substantial flexibility of the spine. Therefore, the aim of this thesis was to gain insight in trunk function and the interaction between trunk, UE and head movements in patients with DMD and SMA using typically developing children and young adults as a reference. This knowledge is essential for developing dynamic supportive devices for the trunk and head to support people with DMD and SMA when performing seated daily tasks. Development of supportive devices for patients with neuromuscular disorders was the overall aim of the Symbionics project, of which the research reported in this thesis is a part.

Review: interaction trunk, head and upper extremity

Chapter 2 is a literature review describing patients with a flaccid trunk caused by both neuromuscular disorders (i.e. DMD and SMA) and central neurological disorders (i.e. CP and SCI), with a main focus on childhood. The aim was to evaluate what is known about trunk involvement when performing seated UE tasks and head movements. Both trunk involvement in the kinematic chain and in the context of providing a stable base were taken into account. We performed a literature search in PubMed with the use of broad key terms. Studies were included if they covered the topic of task performance in seated position, involved both trunk and arm or head movement, and presented outcome measures related to kinematics (i.e. range of motion, movement trajectory, and/or spatiotemporal parameters) or stability (i.e.

center of pressure displacement, trunk sway parameters, and/or force profiles). Out of 188 potentially eligible articles, 32 articles were eventually included in the review. No studies were found involving patients with neuromuscular disorders, like DMD and SMA. Interactions between trunk, head and UE change with age in typically developing children, where trunk movement decreases with age when performing UE tasks. Trunk involvement was also dependent on reaching distance in healthy children, as well as in healthy adults and in patients with CP and SCI. Main differences in trunk movement strategies between CP and SCI patients were seen when reaching towards targets within 90% arm length: increased trunk flexion in CP versus increased trunk extension in SCI. Various strategies were found to maintain trunk stability during reaching: reduce degrees of freedom, reduce movement speed of the arm, counterbalance the perturbing effect of UE movement by moving the trunk or the other arm in the opposite direction, and change the base of support. For the head, stabilizing its orientation in space (i.e. not with respect to the trunk) is the most common strategy in healthy children and adults. However, stabilizing the head appeared more difficult in CP children with a flaccid trunk, because of trunk instability. It is concluded that the key role of the trunk in performing activities should be kept in mind when developing interventions to improve seated task performance in neurological patients with a flaccid trunk, and that more research is needed on these interactions in patients with DMD and SMA.

Measurements: trunk function during seated upper extremity tasks

The literature review showed the gap in knowledge of interactions between trunk, UE and the head in people with DMD and SMA when performing daily tasks. To increase this knowledge, we performed laboratory measurements with healthy controls (**chapter 3**), DMD patients (**chapter 4**) and type 2 and 3 SMA patients (**chapter 5**). The methods were comparable for all groups. All participants performed two series of tasks when sitting unsupported (no back or armrests, feet on the floor). First, they performed maximum active range of motion (ROM) tasks of the trunk and head in all three movement planes. Thereafter, participants performed various daily tasks at self-selected speed, like reaching and placing objects in forward and sideward directions, drinking, and displacing a dinner plate from left to right. Movement of the trunk, pelvis, head and UE were captured with an optical motion capture system when performing these tasks, together with surface electromyography (sEMG) signals from the back (i.e. iliocostalis, longisimus), abdominal (i.e. external oblique) and shoulder muscles (i.e. trapezius descendens and medial deltoid). These sEMG signals were normalized to the maximum sEMG signals obtained during maximum voluntary isometric contraction (MVIC) measurements in a seated position, so that values represented a percentage of the maximum muscle capacity. Maximum trunk and shoulder joint torques were also collected during MVIC.

In **chapter 3,** we aimed to gain insight in trunk, pelvis and head movements when performing UE tasks in 25 healthy children and young adults (6-20 years old). We focused especially on movement of different trunk segments (i.e. upper thoracic, lower thoracic, upper lumbar, and lower lumbar), since the trunk has substantial flexibility but has previously mainly been studied as one rigid segment. We found that contributions of individual trunk segments varied with movement direction and, therefore, with the performed task. The contribution to trunk motion was approximately uniformly distributed across all trunk segments when flexing and decreased from caudal to cranial segments when extending. For lateral bending, the thoracic segments contributed more than the lumbar segments. In axial rotation, movement of the lower thoracic segment with respect to the upper lumbar segment was most important. The pelvis also contributed greatly in all movement directions, indicating that it has a major influence on the maximum trunk movement. Trunk movement significantly increased with reaching height, distance and object weight in the sagittal and frontal planes. This also applied to all individual trunk segments in the sagittal plane and to the thoracic segments in the frontal plane. Similar to the literature, we found that total trunk movement decreased with subject age in childhood when reaching forward and laterally [5, 6]. Age-matched comparison is therefore important in childhood to distinguish between natural and pathologic trunk movements. Head movement was opposite to trunk movement in the sagittal (> 50% of the subjects) and transverse planes (> 75% of the subjects) and was variable in the frontal plane in most tasks. Both trunk and head movement onsets were earlier compared to arm movement onset.

Chapter 3 showed that interaction between trunk and UE movement is essential for accomplishing daily tasks in healthy children and young adults. For DMD patients this may be even more important, because clinically they show increased trunk movement to compensate for reduced arm function. Therefore, the aim of **chapter 4** was to investigate how DMD patients use trunk movement to compensate for reduced arm function. We hypothesized that the use of compensatory trunk movement is dependent on task difficulty and disease progression, and is related to increased trunk muscle activity. Seventeen boys with DMD participated in this study, and results were compared to the 25 healthy controls (HC) as described in Chapter 3. As hypothesized, we found a significant increase in trunk movement in the frontal and/or sagittal plane in DMD patients compared to HC when performing all tasks. However, trunk movement did not significantly increase with task difficulty (i.e. increasing object weight) or Brooke scale. Normalized muscle activity was significantly higher in DMD patients compared to HC for all tasks and all muscles. On average, normalized muscle activity was almost twice as high for back muscles and 4 times higher for abdominal muscles.

These high levels could lead to fatigue and overloading. Normalized muscle activity also increased until a task could not be performed anymore. This might indicate that back muscle function plays a more important role than previously thought, and the UE might not be the only limiting factor for accomplishing tasks. Additionally, trunk and shoulder joint torques were significantly decreased (by 52% and 63%, respectively) in DMD patients compared to HC, and so was the active trunk ROM in all movement planes. Joint torques were already decreased in early disease stages. To conclude, due to increased compensatory trunk movement, demands on trunk muscles are increased in DMD patients, and this is compounded by trunk muscle weakness. Therefore, clinicians should take the increased load on trunk muscles into account when assessing function and when developing interventions such as seating adjustments or physical exercise training. Additionally, if supporting the trunk restricts (compensatory) trunk movements, this will likely cause limitations in accomplishing tasks independently, and could accelerate muscle decline due to disuse.

The aim of **chapter 5** was to investigate trunk function during seated upper extremity tasks in patients with SMA type 2 and 3. Seventeen patients with SMA participated and they were compared with a HC group above the age of 12 years old (n=15, a subgroup of the study described in chapter 3), because the majority of the SMA patients were adults. We expected to find similar results to DMD patients, since patterns of muscle weakness are often described as comparable. However, trunk ROM did not differ between SMA patients and HC when performing the tasks. So, SMA patients did not use compensatory trunk movements when performing seated tasks, although normalized deltoid activity was close to 100% of MVIC in all tasks. The normalized trunk muscle activity was significantly increased in all muscles of SMA patients when performing these tasks. The average muscle activity was almost twice as high for back muscles and 4 times higher for abdominal muscles. This indicates that SMA patients need high levels of trunk muscle capacity to maintain stability when performing UE movements. Consistent with these findings, we found decreased active trunk ROM in SMA patients compared to HC in all movement planes during the active ROM tasks, but comparable percentages of maximum muscle capacity for the muscles counteracting gravitational moments. So, comparable muscle effort coincided with less movement in SMA patients, which is not surprising since the force generating capacity is reduced due to loss of motor neurons. Decrease in maximum muscle capacity was also reflected in significantly decreased maximum trunk and shoulder joint torques in SMA patients compared to HC. Therefore, similar to DMD, clinicians should take trunk function into account when assessing overall function in SMA and when designing interventions, as increased muscular effort to perform tasks could result in fatigue and muscle overloading. Again, one must bear in mind that restrictions in trunk movement will likely cause limitations in accomplishing tasks independently and might accelerate muscle decline due to disuse.

GENERAL DISCUSSION

Trunk function in DMD and SMA

DMD and SMA are often seen as comparable conditions, since both diseases are characterized by progressive muscle weakness, proximally more than distally, and disease onset is (generally) during childhood [7, 8]. This thesis shows to what extent these disorders are comparable in terms of trunk function. Specifically, trunk function in DMD and SMA patients compared to healthy controls (HC) is described in chapters 4 & 5, respectively. It is remarkable that the DMD patients showed increased trunk movement when performing seated tasks and the SMA patients did not. We stated that DMD patients used this increased trunk movement to compensate for their reduced arm function. However, UE function was reduced in both groups compared to HC and the included SMA patients generally had worse UE function compared to the DMD patients (based on the Brooke scale). The percentage of trunk muscle capacity used when performing tasks was comparable between both groups, despite the smaller trunk motions in the SMA group. This suggests that the included SMA patients had on average worse trunk function and likely needed more muscle effort to stabilize the trunk compared to DMD patients. No literature was found to confirm or contradict this hypothesis. Nevertheless, the decreased active trunk flexion and extension in SMA patients compared to DMD patients that we observed substantiates this hypothesis. Yet, this hypothesis was not supported by the median trunk joint torques, as these were comparable between both groups, indicating that the difference in trunk function cannot solely be explained by differences in trunk muscle strength, thus, that other factors must play a role too.

Several other factors could have had an influence on trunk function and could help to explain why the trunk seemed weaker in the included SMA patients compared to the DMD patients when performing tasks, while maximum trunk torque was comparable. First, there was a large difference in body weight and trunk length between the SMA and DMD patients. The median body weight was 74.5 kg (IQR 56.6-88.1 kg) for SMA patients compared to 48 kg (IQR 40-52 kg) for DMD patients, and the median trunk length was 61 cm (IQR 52-67 cm) for SMA patients compared to 50 cm (48-56 cm) for DMD patients. Both higher trunk mass and length would result in larger torques needed to balance the trunk in the same inclination angle against gravity. Therefore, SMA patients were likely functionally weaker than DMD patients, despite similar maximum trunk torques. Secondly, there could be a difference in spinal stiffness; more SMA patients had a scoliosis which could be related to increased stiffness. Therefore, more force is needed to move the parts of the spine involved in the deformity.[9] When performing a maximum voluntary isometric contraction

(MVIC) task to determine the joint torque, stiffness does not limit performance. Third, since the trunk is a central segment, function of the head, UE and lower extremities can also influence the trunk movement (for more detail see heading 'trunk as central segment'). For example, head stability is challenged more when moving the trunk (chapter 2). If neck muscle strength is lower in SMA patients compared to DMD patients, SMA patients might reduce the trunk movement, or be more conservative in moving the trunk to prevent instability of the head. The same applies to the lower extremities as they contribute to sitting stability [10]. If lower extremity function is worse in SMA patients compared to DMD patients, more muscle effort might be required from the trunk to stabilize the body when performing seated tasks. Fourth, there could be a difference in the function of the deep trunk muscles responsible for trunk stability. Demands on trunk stability are less when leaning and pushing against a force sensor (MVIC tasks) than during voluntary UE movement. Last but not least, in the healthy control group we found a decrease in trunk movement with age when performing tasks (chapter 3). SMA patients were generally adults and might therefore initially use less trunk movement to perform tasks compared to the younger DMD patients.

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However, these results are difficult to generalize across the patient populations. SMA patients seemed to have a weaker trunk in the population tested, but also had worse UE function compared to the DMD patients. Nevertheless, even if UE function was comparable between both groups, differences in trunk function could still be anticipated between these patient groups. For instance, imaging studies for lower extremity, pelvis and UE muscles show differences in muscle atrophy and fatty infiltration patterns between patients with DMD and SMA [11-14]. However, studies evaluating back or abdominal muscles are minimally available. Two research groups found that the rectus abdominis and external oblique muscles are affected in late disease stages in both DMD and SMA type 3 patients [11, 15], and Sambrook, et al. [14] showed that the posterior spinal muscles are involved in the disease at mid-disease stage in SMA patients. Therefore, new imaging studies are necessary to gain more insight in the trunk muscle weakness patterns in both groups. This would improve the understanding of our results and provide insight in disease progression in relation to the trunk. More in-depth research into trunk stability itself can also provide more insight in the differences in trunk function that we observed. For instance, muscle reflex response times might be relevant to study, since reflexes can be absent or reduced in SMA patients [16] (impacting the maintenance of stability) and can therefore differ from DMD patients.

Measuring trunk function

Physiological measures

In this thesis, we used physiological measures, such as muscle strength testing, kinematic analysis and electromyography to assess trunk function. These measures give insight in the maximum capacity (maximum strength, maximum range of motion, and maximum muscle activity) and the movement and muscle activity used to perform daily tasks, but also have their limitations.

Markers were placed on the participants' back to gain insight in trunk movement. Since these markers needed to be visible for the optical infrared cameras, only participants were included who were able to sit independently without a backrest. This does not reflect the entire patient population and the daily life situation for patients seated in a wheelchair. Therefore, being able to measure trunk movement when seated in the wheelchair would have had added value, and might lead to different results with regard to trunk movement. Compensatory trunk movements might increase in early non-ambulatory patients, because the backrest can be used as a support surface for compensatory movements in the frontal plane, it provides safety from falling backwards, and reduces muscle effort needed to maintain posture. However, measuring trunk movement in multiple segments in a wheelchair is challenging, because measurement systems have to be very small and pressure resistant, so that they do not cause discomfort and generate signals without artifacts caused by sitting against the backrest. A few startup companies, like Bainisha (Bainisha cvba, Lokeren, Belgium) and Epionics SPINE (Epionics Medical GmbH, Potsdam, Germany), are developing new types of sensors based on strain gauge techniques that might be promising for future research in patients seated in a wheelchair. These systems might even make it possible to perform home based measurements or measure for a whole day to give insight in the performance in daily life.

Surface EMG provides insight in the capacity used by some superficial trunk muscles, however, these muscles are thought to be mainly responsible for movement and less for stability [17]. Since we hypothesized that SMA patients needed much more of their muscle capacity to stabilize the trunk, more insight is needed into function of the deep trunk muscles. An alternative to invasive needle or fine wire EMG would be the use of imaging techniques like magnetic resonance imaging (MRI) to grade the fatty infiltration and muscle volume. This is related to loss of muscle fibers and therefore also to functional grades [18, 19]. Yet, imaging studies involving trunk muscles in DMD or SMA are scarce as described above.

Measurement scales

Also standardized clinical measures would be useful to gain insight in trunk capacity or performance and are generally more easily applicable in clinical practice. However, validated measures for trunk function are scarce. The motor function measure (MFM) and the Hammersmith functional motor scale include trunk function and are often used in DMD and SMA patients [20, 21]. However, both scores are influenced by upper or lower extremity function, which makes it difficult to examine trunk function by itself. For example, MFM item 9 is defined as follows: "the patient sits on a chair and stretches his/her arms forward and maintains this position for 5 seconds". When he/she cannot stretch the arms forward for 5 seconds, the score is decreased by 1 point, however, this could be solely due to a limitation in UE function. So, scoring trunk function with these measures will only be appropriate if trunk function is worse than UE function. Based on the results of this thesis, this assumption is, at least for DMD patients, highly questionable. Additionally, scoring is often decreased when a patient uses compensatory trunk movements when moving the UE, where this could also be seen as good trunk function. A proper measurement scale should therefore have a domain focusing on trunk function alone, but also one domain combining trunk and UE movement, because this reflects daily life situations.

The Trunk Control Measurement Scale (TCMS) can be a good starting point as a measure for DMD and SMA, but its validation is needed. The TCMS has been developed and validated for children with cerebral palsy (CP) with the purpose to score both roles of the trunk during seated activities as discussed in this thesis, namely to form a stable base of support (i.e. static sitting balance) and to constitute an actively moving body segment (i.e. dynamic sitting balance) [22]. The latter aspect is further divided in the TCMS into selective movement control (i.e. trunk movement only) and dynamic reaching (i.e. involvement of the UE). Being able to use the same measurement scale for different patient groups with impaired trunk function gives the opportunity to compare diseases and generalize treatment strategies where possible.

Detail of measuring

As pointed out in the general introduction, the trunk is a complex segment and therefore people should carefully decide on which level of detail they want to evaluate the trunk. The aim and patient population should be kept in mind when making such a decision. For clinical assessment of overall trunk function, considering the trunk as one segment could be sufficient for DMD and SMA patients, because our results showed that the contribution of individual trunk segments to overall trunk movement was comparable to HC. However, differences between trunk levels can be expected in patients with a scoliosis, CP or spinal cord injury [23]. Therefore, considering the trunk as a rigid segment might not be appropriate for these patient groups, as it might mask focal impairments or compensatory mechanisms. Clinical assessment of individual trunk levels could be done with the use of the Segmental Assessment of Trunk Control [24]. However, some adjustments have to be made when used in patients with DMD and SMA, because the trunk control score is automatically zero if people cannot keep the UE in shoulder abduction [25]. When the aim is to study specific trunk movements or to develop devices or interventions, it is recommended to separately assess several trunk segments and the pelvis.

Trunk as central segment

Interaction with head

Head movement is strongly related to trunk movement as was discussed in chapters 2 and 3. We measured head movements in DMD and SMA patients as well, but chose not to incorporate these results in chapters 4 and 5, as this would have led to complex and elaborate manuscripts. DMD and SMA patients are able to move their head until in the late disease stages, despite neck flexor weakness already present in early disease stages [7, 8]. In our population, neck muscle weakness was evident in the reduced maximum neck ROM in all movement planes for SMA patients and in extension and axial rotation for DMD patients. However, all but one SMA participant did not seem to be restricted in head movement when performing the tasks (restriction in head movement in this SMA participant was caused by spinal fusion up to cervical levels). Neck muscle weakness negatively influences head stability probably earlier than the loss of ability to move the head. This is especially noticed by patients when moving in a car or wheelchair over uneven surfaces. Additionally, decreased trunk stability also challenges head stability, as previously reported for patients with CP [26].

Interaction with lower extremities

The lower extremities play an important role in sitting stability [10]. A larger base of support is created with the feet on the floor, resulting in increased limits of stability and therefore increased workspace [27]. Feet position is also adapted by healthy children and adults to meet specific task demands in terms of stability [28]. Moreover, even if the feet remain in place, the leg muscles are actively involved in performing seated tasks [10, 29]. Therefore, in patients with neuromuscular disorders with early leg muscle weakness, impaired contribution of the lower limbs to trunk stability can be anticipated. However, the impact of such impairment is unknown. We performed a first pilot study with 9 healthy boys and 9 boys with DMD to see if we could detect a difference in contribution of the legs to bodyweight distribution between both groups while sitting quietly, lifting the arms and reaching forward. We used a force plate to measure the ground reaction forces and found no difference in contribution of the legs to bodyweight support between DMD patients and HC when sitting quiet and lifting the arms, but we did find a significant decrease in contribution of the legs in DMD patients when reaching beyond arm's length, even though only 3 of the 9 boys with DMD were non-ambulant. Further research should clarify whether differences were indeed related to leg muscle weakness, or that weakness of the trunk or UE also played a role. Measurements should combine ground reaction forces with leg and trunk muscle activity, and a trunk stability measurement, to determine the passive/active contribution of the legs and to see how this relates to trunk stability. Notably, even the non-ambulant patients were still able to increase the force on their feet. Whether this is only a passive support from the legs or also an active contribution needs to be determined in further research. Clearly, irrespective of the question whether these forces are active or passive, these findings show that proper feet support while sitting in a wheelchair is necessary.

Implications for interventions

This thesis showed that the trunk in DMD and SMA patients is much weaker than it appears at first sight. Although the included patients could sit independently (without backrest) and were able to perform several arm tasks at the same time, trunk strength was strongly decreased and trunk muscle effort was much higher compared to HC. This means that clinical interventions with regard to trunk function might be required to start earlier than what would be expected based on the observable decline in trunk function.

Physical activity and seating

Physical and muscle strength training has beneficial effects on muscle strength and fatigability [30, 31]. Even if people become wheelchair dependent, physical training can be beneficial to reduce the deterioration caused by disuse [31]. Seating adjustments can be another way to reduce fatigue in wheelchair users. A proper seating cushion combined with proper back- and armrests should provide the opportunity to relax the muscles when seated in a wheelchair. Additionally, it is also important for prevention of pressure points and improving sitting stability when necessary [32]. The latter is often done by stabilizing/fixating the pelvis first in order to create a stable base for UE task performance. However, as also mentioned by Sprigle, et al. [33], providing stability with wheelchair cushions or a sitting orthosis undermines the ability to move the trunk and pelvis to perform daily tasks (chapter 3) and additionally to prevent disuse. A good balance between providing stability and allowing movement needs to be sought to optimize task performance. Therefore, more research is needed into both potential benefits and negative effects of stabilizing the pelvis and restricting trunk movement when performing daily tasks. As this balance of effects is likely to change with the disease stage, this research should be extended to a range of disease stages in DMD and SMA.

Spinal deformities

Spinal deformities are often seen in patients with DMD and SMA due to trunk muscle weakness already being present while the spine is still growing [34, 35]. Severe scoliosis can have an effect on sitting balance and cardio-respiratory function and is generally treated by spinal surgery [36]. After spinal surgery, sitting balance is often improved, but ROM is reduced because spinal flexibility is lost [37]. Despite the fact that movement between spinal segments is used to perform daily tasks (chapter 2), people with sufficient muscle strength do not experience major restrictions in performing daily activities after surgery [38, 39]. Their muscle strength is large enough to create sufficient torque to move the rigid trunk; when spinal segments are fused the amount of force needed to stabilize and move the rigid trunk increases (increase in inertia). However, patients with neuromuscular disorders will lose function due to the spinal surgery [40]. Compensatory trunk movements cannot be performed anymore and it is more difficult, or even impossible, to create enough muscle force to move the rigid trunk towards higher inclination angles.

The level of spinal fixation is determined by the surgeon and can be limited to the thoracolumbar region or can involve fixation of the lumbosacral joint too. There is still controversy whether it is necessary to extent the fixation to the sacrum. The major argument for inclusion of the lumbosacral joint is to correct for pelvic obliquity and thereby creating a leveled and stable base. However, we found that movement between the lower lumbar spine and pelvis segment contributes substantially to trunk ROM in the sagittal plane (chapter 2), so restricting movement at the lumbosacral joint would likely decrease functionality even more. In addition, some studies showed that improvement of pelvic obliquity in the frontal plane could also be seen when fixation was done down to the L5 level, although this only applied to patients with less severe pelvic obliquity [41, 42]. These arguments together would plead for fixation down to the L5 level as long as further fixation is not strictly necessary, which is also suggested in the Dutch guideline for scoliosis treatment in neuromuscular disorders [43].

Development of dynamic supportive devices

Another solution to reduce fatigability and assist patients in their trunk and head movement would be the development of dynamic supportive devices. The development of such devices was the aim of the Symbionics project. Both passive systems (i.e. spring based) and control methods for active systems (i.e. actuated by motors) were developed in the project and knowledge from this thesis helped in the development.

First of all, we saw clear trunk involvement in healthy participants when performing UE tasks (chapter 3), which confirmed that trunk movement is essential and should be supported in patients to optimize task performance. Based on the ROM found in healthy participants when performing tasks, we could set requirements for the amount of trunk and head movement that should be provided by the supportive devices. Supporting both trunk and head flexion-extension had the highest priority and axial rotation should either be allowed or supported. Control of the supportive devices should preferably not be based on displacement of the hand, since trunk and head movement onsets were earlier than arm movement onset (chapter 2). Last but not least, we saw that contribution of individual trunk segments to the ROM varied with movement direction and therefore with the task performed, so the devices should provide or allow movement in different segments or continuous movement.

In DMD patients we saw that UE function (i.e. Brooke scale) seemed to decrease before trunk function. This indicates that the trunk supportive device for DMD patients will usually be integrated with an UE supportive device. Since previous research showed that compensatory trunk movements decrease when an UE supportive device is used [44], we did not increase the requirements for the amount of trunk movement that should be provided by the device, although increased trunk movements were seen in DMD patients compared to HC (chapter 4). It remains difficult to predict whether an UE or trunk support should be provided first in SMA patients or that both should be combined from the start, because we could not compare different disease stages in SMA patients. Increasing demands of support can be expected in both groups due to progressive muscle weakness. Therefore, support levels should be adaptable to the disease stage or even during the day to prevent excessive fatigue. When the support level is higher than required, there is a risk of deterioration of function due to disuse. On the other hand, providing more support than needed can also give possibilities to perform more activities or with a longer duration. Clearly, more research is needed to quantify the required support level and to determine factors that influence this level. Ultimately, adaptive devices are needed. In other words, devices that automatically adapt their support level based on predicted user needs, or systems that adapt based on intentional user-driven signals.

Evaluation of the developed passive trunk support device for trunk flexion and extension showed reduced normalized back muscle activity by 10-35% in healthy man and boys with DMD [45, 46]. This means that patients need up to 1/3 less muscle effort to perform tasks, which would reduce development of fatigue and provide the option to increase the frequency or duration of performing tasks. Using a passive head support system also reduced normalized upper trapezius muscle activity levels. Therefore, these dynamic systems are promising solutions for interventions to optimize muscle load in DMD and SMA patients when performing daily tasks.

Recommendations

In this thesis we made the first step towards obtaining a better understanding of trunk function in patients with neuromuscular disorders during UE tasks, but we are still far away from a complete understanding of trunk function in the perspective of different disease stages. A profound MRI study would be the first step to obtain more insight in trunk muscle degeneration patterns in DMD and SMA patients and would help to explain our findings in more detail. Inclusion of DMD and SMA patients with different disease stages, and different types of SMA, will be essential. It is also important to include both superficial and deep back muscles (at different levels) and abdominal muscles to create an overall picture of trunk muscle weakness, which can be related to the movement patterns and muscle activity levels observed.

We hypothesized that trunk stability is decreased in SMA patients and is worse in SMA patients compared to DMD patients. To confirm or reject our hypotheses, more research on this topic is recommended. Evaluating trunk stability could be done by measuring center-of-pressure trajectories when being seated on a wobbly chair [47]. However, since SMA and DMD patients can have reduced or absent postural reflexes, insight in the presence, amplitude and timing of the reflexes involved in trunk stability control might be more valuable. This has not been studied before in these patient groups. van Drunen, et al. [48] described a method to quantify intrinsic and reflexive muscular contributions to trunk stabilization by providing force perturbations at the trunk and measuring frequency response functions, kinematics and reflexes (sEMG). But this method likely needs to be adapted slightly to the population at hand, like reducing the perturbation force. Combined with imaging-based information on deep and superficial muscles, this could help to further unravel trunk function, including better distinction between (control of) stability and overall trunk motion.

Development of a reliable trunk measurement scale is also recommended, so that trunk function can be evaluated reliably in clinical practice. Evaluating the feasibility and reliability of the Trunk Control Measurement Scale in patients with DMD and SMA can be a good starting point [22]. Additionally, it would be interesting to evaluate whether scores in the 'dynamic sitting stability' could be divided into subscores for the 'trunk only' tasks and tasks that also include UE movement.

Fourth, repeating the measurements performed in this thesis with other patient populations having a flaccid trunk, like (bilateral) CP and SCI, would be of great interest to extract common grounds and crucial differences for interventions. It can also increase our understanding of the interaction between trunk, UE and head movements when performing tasks. However, from a pilot study with three boys with CP (Gross Motor Function Classification Score [49] 3 and 4) we learned that the developed measurement protocol should be adapted to the patient population. The CP participants had problems with selective trunk control, which is known from literature [50, 51]. Selective movement control is essential for proper execution of the MVIC tasks. Often compensatory trunk movements were observed and it was sometimes difficult to determine whether these occurred because of the impairments in selective movement control or because instructions were misunderstood. The reliability of the measured muscle strength was therefore questionable and consequently the reliability of the maximum sEMG signals too. This implied that the sEMG signals during task performance could not be normalized. Since normalized sEMG signals are necessary to compare participants, alternatives have to be found for using the MVIC tasks in children with CP, like normalization based on a specific task. The disadvantage of this method is that the maximum contraction amplitude cannot be determined and a task should be selected that can be performed correctly by all participants (taken into account contractures and spasticity) [52].

Finally, research should be extended to the influence of the pelvis and the lower limbs on trunk movement, stability and the performance of daily tasks. Effect of cushions on pelvis and trunk movement can be examined, as well as effects of complete fixation of the pelvis in different patient populations. For the contribution of the legs, research should combine ground reaction force measurements with sEMG of the legs and trunk, to disentangle active and passive components. This knowledge will be crucial in the light of seating adjustments and positioning in the wheelchair in order to support people as well as possible in their daily activities.

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CHAPTER 7
NEDERLANDSE SAMENVATTING

Afkortingen:

CP	Cerebrale parese
DMD	Duchenne spierdystrofie
GC	Gezonde controles
MVIC	Maximum Vrijwillige Isometrische Contractie
sEMG	Oppervlaktie electromyografie
SMA	Spinale spieratrofie

NEDERLANDSE SAMENVATTING

Romp- en hoofdbeweging zijn onmisbaar tijdens het uitvoeren van zittende taken met de armen. De romp werkt samen met de armen en het hoofd als onderdeel van de bewegingsketen en het zorgt voor een stabiele basis voor het vrijwillig uitvoeren van bewegingen met de armen en het hoofd. De romp kan verzwakt raken door spierzwakte (bijvoorbeeld door Duchenne spierdystrofie (DMD) of spinale spieratrofie (SMA)) of door een centraal neurologische aandoening (bijvoorbeeld door cerebrale parese (CP) of een dwarslaesie). Als de romp verzwakt is in de (vroege) kinderjaren, kan dit ook effect hebben op de motorische ontwikkeling en kan dit de kans vergroten om een verkromming van de ruggenwervel (scoliose) te ontwikkelen. Dit heeft vervolgens ook weer gevolgen voor rompbeweging en stabiliteit.

Ondanks dat de romp een vitale rol speelt tijdens het uitvoeren van zittende taken met de armen, is er weinig onderzoek gedaan naar de rompfunctie in patiënten met DMD en SMA. Het is van groot belang om meer inzicht te krijgen in de interactie tussen romp, armen en hoofd tijdens het uitvoeren van zittende taken, omdat veel patiënten met DMD of SMA (die symptomatisch zijn in de kinderjaren), niet in staat zullen zijn om te lopen als ze volwassen zijn. Bovendien richt de schaarse literatuur over deze interacties, in typisch ontwikkelende kinderen, zich vooral op de romp als één star segment en negeert de aanzienlijke flexibiliteit van de wervelkolom. Het doel van dit proefschrift was daarom ook om meer inzicht te krijgen in romp functie en de interactie tussen de romp, armen en het hoofd in typisch ontwikkelende kinderen en jong volwassenen, en patiënten met DMD en SMA.

Deze kennis is essentieel voor de ontwikkeling van dynamische hulpmiddelen die de romp en het hoofd kunnen ondersteunen bij mensen met DMD en SMA als ze dagelijkse zittende taken uitvoeren. Ontwikkeling van deze hulpmiddelen was het doel van het Symbionics project, waar dit proefschrift onderdeel van uitmaakt.

Review: interactie tussen romp, armen en hoofd

Hoofdstuk 2 geeft een literatuur overzicht over de interactie tussen romp, hoofd en armbewegingen bij patiënten met een slappe romp veroorzaakt door zowel neuromusculaire aandoeningen (DMD en SMA) als centraal neurologische aandoeningen (CP en dwarslaesie), met een focus op de kinderjaren. Het doel was om inzicht te krijgen in de huidige kennis over de bijdrage van de romp tijdens het uitvoeren van zittende arm taken en hoofdbewegingen. Zowel de betrokkenheid van de romp in de bewegingsketen als de betrokkenheid bij stabiliteit zijn hierin meegenomen. We hebben uitgebreide zoektermen gebruikt om literatuur te zoeken in PubMed. Studies waren geïncludeerd als ze gingen over het uitvoeren van taken in zittende positie, zowel romp als arm of hoofdbeweging beschreven was, en er uitkomst maten gebruikt waren gerelateerd aan beweging (zoals bewegingsbereik, bewegingpad en/of spatio-temporele parameters) of gerelateerd aan stabiliteit (zoals verplaatsing van het centrum van de druk, romp sway parameters en/of krachtprofielen). Van de 188 artikelen die mogelijk in aanmerking kwamen, zijn 32 artikelen uiteindelijk opgenomen in het literatuur overzicht. Er zijn geen studies gevonden over patiënten met een neuromusculaire aandoening, zoals DMD en SMA. Interactie tussen romp, hoofd en de armen verandert met leeftijd in typisch ontwikkelende kinderen; de rompbeweging vermindert met leeftijd bij het uitvoeren van taken met de armen. Betrokkenheid van de romp was ook afhankelijk van reikafstand in gezonde kinderen, evenals gezonde volwassenen en patiënten met CP of een dwarslaesie. De voornaamste verschillen tussen CP en dwarslaesie patiënten in romp bewegingstrategieën werden gezien bij voorwaarts reiken binnen 90% arm lengte: meer romp flexie bij CP tegenover meer romp extensie bij dwarslaesie. Verschillende strategieën werden gevonden om romp stabiliteit te handhaven tijdens reiken: verminderen van vrijheidsgraden, verminderen van bewegingssnelheid van de arm, het compenseren van het verstorende effect van de armbeweging door de romp of andere arm in tegengestelde richting te bewegen, en het veranderen van de basis van ondersteuning. Het hoofd stabiliseren in de ruimte (en niet ten opzichte van de romp) is de meest voorkomende strategie gebruikt door gezonde kinderen en volwassenen. Echter bleek het stabiliseren van het hoofd in CP kinderen met een slappe romp moeilijker te zijn door romp instabiliteit. Concluderend moet de sleutelrol van de romp voor het uitvoeren van activiteiten in gedachten worden gehouden bij de ontwikkeling van interventies om het uitvoeren van zittende taken in neurologische patiënten met een slappe romp te verbeteren, en is er meer onderzoek nodig omtrent deze interacties in patiënten met DMD en SMA.

Metingen: romp functie tijdens het uitvoeren van zittende arm taken

Het literatuur overzicht liet ontbrekende kennis zien omtrent romp, arm en hoofd interacties bij mensen met DMD en SMA tijdens het uitvoeren van dagelijkse taken. Om kennis op dit gebied te vergroten, hebben we metingen uitgevoerd in het bewegingslaboratorium met gezonde controles (**hoofdstuk 3**), patiënten met DMD (**hoofdstuk 4**) en patiënten met type 2 of 3 SMA (**hoofdstuk 5**). De methode was gelijk voor alle groepen. Alle deelnemers hebben twee series van taken uitgevoerd tijdens niet ondersteund zitten (zonder rug- of armleuningen, maar wel voeten op de grond). Als eerste hebben zij maximum actieve bewegingsbereik taken van de romp en het hoofd uitgevoerd in alle drie de bewegingsvlakken. Daarna werden er verschillende dagelijkse taken uitgevoerd in zelf gekozen snelheid, zoals reiken en plaatsen van objecten in voorwaartse en zijwaartse richting, drinken, en het verplaatsen van een bord van links naar rechts. Beweging van de romp, het bekken, het hoofd en de armen werden opgenomen met een optisch bewegingsregistratie systeem samen met oppervlakte elektromyografie (sEMG) signalen van de rug (iliocostalis en logisimus), buik (external oblique) en schouder spieren (trapezius descendens en medial deltoid). Deze sEMG signalen werden genormaliseerd ten opzichte van de maximale sEMG signalen verkregen tijdens de maximale vrijwillige isometrische contractie (MVIC) metingen in zittende positie, zodat de spieractiviteit werd uitgedrukt als een percentage van de maximale spiercapaciteit. Maximale romp en schouder gewrichtskoppel waren ook verzameld tijdens MVIC.

In **hoofdstuk 3** wilde wij inzicht krijgen in romp, bekken en hoofd bewegingen tijdens het uitvoeren van arm taken in 25 gezonde kinderen en jong volwassenen (6 – 20 jaar oud). We hebben in het bijzonder gefocust op de beweging van de verschillende romp segmenten (hoog thoracaal, laag thoracaal, hoog lumbaal en laag lumbaal), omdat de romp een aanzienlijke flexibiliteit heeft maar eerder vooral bestudeerd is als één star segment. We hebben gevonden dat de bijdrage van individuele romp segmenten varieert met de bewegingsrichting en daardoor ook met de uitgevoerde taak. De bijdrage aan de maximale rompbeweging was ongeveer evenredig verdeeld onder alle romp segmenten tijdens flexie en verminderde van de caudale naar craniale segmenten tijdens extensie. Tijdens maximale laterale buiging hadden de thoracale segmenten een grotere bijdrage dan de lumbale segmenten. Tijdens maximale axiale rotatie was de bijdrage van het laag thoracale segment (ten opzichte van het hoog lumbale segment) het meest belangrijk. De bijdrage van het bekken was ook aanzienlijk in alle bewegingsrichtingen, wat aangeeft dat dit een grote invloed heeft op de maximale rompbeweging. De rompbeweging nam significant toe met reikhoogte, -afstand en object gewicht in het sagittale en frontale vlak. Dit gold ook voor alle individuele rompsegmenten in het sagittale vlak en de thoracale segmenten in het frontale vlak. Vergelijkbaar met de literatuur vonden wij dat de totale rompbeweging afnam met leeftijd in de kinderjaren bij het voorwaarts en zijwaarts reiken. Het is daarom belangrijk om in de kinderjaren te vergelijken met eenzelfde leeftijdsgroep om onderscheid te kunnen maken tussen natuurlijke en pathologische bewegingen. Hoofdbeweging was tegengesteld aan de rompbeweging in het sagittale vlak (> 50% van de deelnemers) en in het transversale vlak (> 75% van de deelnemers), en was variabel in het frontale vlak in de meeste taken. Zowel het begin van romp- als hoofdbeweging was eerder dan het begin van de armbeweging.

Hoofdstuk 3 liet zien dat de interactie tussen romp en armbewegingen essentieel is voor het uitvoeren van dagelijkse taken bij gezonde kinderen en jong volwassenen. Voor patiënten met DMD is dit mogelijk nog belangrijker, aangezien zij klinisch meer rompbeweging laten zien ter compensatie van een verminderde arm functie. Daarom was het doel van **hoofdstuk 4** om te onderzoeken hoe patiënten met

DMD rompbewegingen gebruiken om te compenseren voor verminderde arm functie. Onze hypothese was dat het gebruik van compenserende rompbewegingen afhankelijk was van taak moeilijkheid en het ziektestadium, en dat het gerelateerd was aan een verhoogde rugspieractiviteit. Zeventien jongens met DMD hebben deelgenomen aan dit onderzoek en de resultaten zijn vergeleken met de 25 gezonde controles (GC) die ook beschreven waren in hoofdstuk 3. Zoals verwacht vonden we een significante toename van de rompbeweging in het frontale en/of sagittale vlak bij DMD patiënten bij het uitvoeren van alle dagelijkse taken vergeleken met GC. Echter was de rompbeweging niet significant hoger bij moeilijkere taken (zwaardere objecten) of later ziektestadium (Brooke schaal). Genormaliseerde spieractiviteit was significant hoger in patiënten met DMD in vergelijking met GC voor alle taken en alle spieren. Gemiddeld was de genormaliseerde spieractiviteit twee keer zo hoog voor de rugspieren en vier keer zo hoog voor de buikspieren. Deze verhoogde spieractiviteit kan leiden tot vermoeidheid en overbelasting. De genormaliseerde spieractiviteit nam toe totdat een taak niet meer uitgevoerd kon worden. Dit geeft aan dat de rugspier functie mogelijk een belangrijkere rol speelt dan van te voren gedacht, en dat de arm functie mogelijk niet de enige limiterende factor is voor het kunnen uitvoeren van dagelijkse taken. Bovendien waren de romp en schouder gewrichtskoppels significant lager (respectievelijk 52% en 63% lager) in DMD patiënten vergeleken met GC, net zoals het maximale actieve bewegingsbereik van de romp in alle bewegingsrichtingen. Gewrichtskoppels waren al verminderd in een vroeg ziektestadium. Concluderend, als gevolg van de toegenomen (compenserende) rompbewegingen, nemen de eisen aan de rompspieren ook toe bij patiënten met DMD, en dit wordt ook nog eens versterkt door spierzwakte in romp. Daarom moeten clinici de toegenomen belasting op de rompspieren mee nemen bij het beoordelen van de algemene functie en bij het ontwerpen van interventies, zoals zitaanpassingen en fysieke training. Maar als het ondersteunen van de romp het maken van (compenserende) rompbewegingen beperkt, leidt dit waarschijnlijk tot beperkingen in het zelfstandig uitvoeren van dagelijkse taken, en kan bovendien de spierafname versnellen als gevolg van het niet gebruiken van de spieren.

Het doel van **hoofdstuk 5** was om rompfunctie te onderzoeken in patiënten met SMA type 2 en 3 tijdens het uitvoeren van dagelijkse arm taken in zittende positie. Zeventien patiënten met SMA hebben deelgenomen en we hebben de resultaten vergeleken met de gezonde controles boven de 12 jaar (n=15, een subgroep van de deelnemers beschreven in hoofdstuk 3), omdat het merendeel van de SMA deelnemers volwassen was. We hadden verwacht vergelijkbare resultaten te vinden als bij de DMD patiënten omdat de spierzwakte patronen vaak beschreven staan als vergelijkbaar. Echter was de rompbeweging bij SMA patiënten niet verschillend van GC tijdens het uitvoeren van dagelijkse taken. Dus SMA patiënten gebruikten geen compenserende rompbewegingen tijdens het uitvoeren van dagelijkse taken, terwijl de genormaliseerde deltoideus activiteit in alle taken dicht bij de 100% MVIC lag. De genormaliseerde spieractiviteit was significant hoger in alle spieren bij patiënten met SMA. De gemiddelde genormaliseerde spieractiviteit was bijna twee keer zo hoog voor de rugspieren en vier keer zo hoog voor de buikspieren. Dit suggereert dat SMA patiënten hoge percentages van de rompspiercapaciteit gebruiken om stabiliteit te behouden als ze dagelijkse arm taken uitvoeren. In overeenstemming met deze bevindingen, vonden we een verminderd actief maximaal bewegingsbereik van de romp in alle bewegingsrichtingen vergeleken met GC, maar met vergelijkbare percentages van de maximale spiercapaciteit voor de spieren die tegen de zwaartekracht in werken. Dus vergelijkbare spierinspanning resulteert in minder beweging bij SMA patiënten. Dit is niet verrassend omdat de capaciteit om spierkracht te genereren verminderd is als gevolg van het verlies van motorneuronen. Een verminderde maximale spiercapaciteit was ook zichtbaar in het significant lagere romp en schouder gewrichtskoppel bij patiënten met SMA in vergelijk met GC. Daarom moeten clinici ook bij deze doelgroep, net zoals bij DMD, rekening houden met de romp functie bij het beoordelen van algemene functie en voor het ontwerpen van interventies. De verhoogde spierinspanning die nodig is voor het uitvoeren van dagelijkse taken kan namelijk leiden tot vermoeidheid en overbelasting. Zoals ook hierboven beschreven, is het belangrijk dat men in gedachten houdt dat het beperken van de rompbeweging kan resulteren in beperkingen bij het zelfstandig uitvoeren van dagelijkse taken, en kan spierafname versnellen door het niet gebruiken van de spieren.

7

APPENDICES

DANKWOORD

Na vele jaren met enorm veel plezier te hebben gewerkt aan de onderzoeken die hiervoor beschreven staan, is het nu toch echt klaar. Ik wil iedereen bedanken die op welke manier dan ook mee heeft gewerkt om dit proefschrift tot stand te brengen.

Allereerst mijn grote dank aan alle deelnemers die mee hebben gedaan aan de verschillende onderzoeken. Zonder jullie had ik dit onderzoek niet kunnen doen en was dit boekje er nooit geweest! Ook bedankt aan alle ouders voor de betrokkenheid bij het onderwerp en de tijd die ze vrij hebben gemaakt. Marco, Frank, Frank en Hanneke, ontzettend bedankt voor jullie tijd en betrokkenheid bij het Symbionics project. Jullie ervaringen, feedback en het actief mee denken tijdens de focusgroep bijeenkomsten was zeer waardevol om goede keuzes te kunnen maken voor de ontwikkeling van de dynamische ondersteuners.

Daarnaast wil ik graag mijn promotieteam hartelijk bedanken. Imelda, dankjewel voor je enthousiasme, ondersteuning, vertrouwen en kennis de afgelopen jaren. Ik vond het erg prettig om met je samen te werken en veel geleerd van onze overleggen. Je wist altijd de positieve punten te benoemen, terwijl ik soms zelf niet zag dat het al een stap vooruit was. Bij jou staat de patiënt altijd op nummer één, iets waarvoor ik je enorm bewonder. Het is mooi om te zien dat je dat nooit verloren hebt, zelfs niet nu je pensioen langzaam nadert.

Idsart, ik ben heel blij dat je onderdeel bent geworden van mijn promotieteam. Ik had het niet gekund zonder je ondersteuning bij de analyses en interpretatie van de resultaten. Heel fijn dat ik altijd bij je kon aankloppen en je snel tijd had om iets te bespreken of te reviewen. Ik heb veel van je geleerd.

Jaap, bedankt voor je betrokkenheid bij het onderzoek. Je inbreng tijdens overleggen en op artikelen was altijd heel waardevol. Je heldere uitleg en kritische vragen hebben me geholpen om de materie beter te begrijpen en om het onderzoek te verbeteren.

Sander, ondanks dat we elkaar niet veel hebben gesproken wist je altijd de juiste vragen te stellen en met een kritische blik te kijken naar mijn onderzoek. Dit bewonder ik ten zeerste. Ik heb ook zeer veel gehad aan je feedback op het schrijven. Je wist de boodschap over te brengen in veel minder woorden waardoor de tekst krachtiger werd.

Ik wil ook de leden van de manuscriptcommissie hartelijk bedanken voor de bereidheid om met een kritische blik mijn proefschrift te beoordelen.

Nauzef, Stergios and Anoek, it was a great pleasure to work closely together in the Symbionics project. I think we have proved that you can achieve more with good teamwork. Nauzef, it is very pleasant to work with you. You work very hard, are patient and friendly. You have clear ideas in your mind but are always open for other suggestions. With your designs for the passive orthoses, you made a major step towards an actual product usable for patients with neuromuscular disorders. You can be proud of this. Stergios, you were the expert on a topic which I knew the least about. In the beginning we strugled to understand eachothers worlds and langurage, but I think we mannaged at the end, and the collaboration was good and fun. It was very nice to perform a measurement together and you have really surprised me with the great participant interactions during these measurements. Anoek, jij hebt je wat later bij onze groep gevoegd als PDEng. Ondanks dat jouw opdracht wat meer los stond, was je altijd heel betrokken bij het project en kon je goed met ons mee praten en denken. Jouw rustige, vriendelijke en enthousiaste houding zorgt ervoor dat het heel fijn is om je in de groep te hebben. Wish you all the best for the future.

I also want to thank everyone else participating in the Symbionics project. First of all the Symbionics 2.1 team (Bart, Bart, Edsko, Arjen, Micha en Hans) and all partners, thank you for your great ideas and suggestions, involvement and critical notes. I always enjoyed our meetings. All other PhD candidates, Bob, Hans, Kostas, Niek, Ronald, Roy, thank you for the 'gezelligheid' at the yearly symposium, all the conferences (including South Africa!) and our weekend trips. I will never forget the tree climbing, the great costumes for the highland games and that if you let two Greek people arrange a barbecue, you know for sure that there is a lot of (!!) meet.

Lieve kamergenoten, Mariska, Mariëlle, Rosemarie, Rosanne, Theo, Jilske, Merel, bedankt voor de goede gesprekken en gezelligheid tussen het harde werken door. Zonder jullie was het een stuk saaier geweest, maar misschien op z'n tijd wel productiever ;). Daarnaast natuurlijk ook alle andere 'onderzoekers', Lotte, Frank, Jolanda, Milou, Bas, Renee, Geert, Vera, Mitchel, Wouter, Digna en Marian. Samen lunchen, borrelen in de Aesculaaf, SYTYCC, trampoline springen, ploeteren tijdens de schrijfweken en 's avonds spelletjes spelen of sporten. Ik had het niet willen missen! Als laatste ook alle andere collega's die mij op enig moment geholpen hebben tijdens mijn promotie (in het bijzonder Yolanda, de collega's van de kinderkant, Dorien en Laurien), bedankt.

Ik heb ook nog enkele stagiaires mogen begeleiden tijdens mijn promotieonderzoek. Janne, Marissa en Maaike, bedankt voor jullie geweldige inzet en hulp bij het onderzoek. Ik hoop dat jullie er veel van geleerd hebben. Ik vond het in ieder geval erg prettig om samen te werken en samen de metingen uit te voeren.

Lieve TG chickies, Nicole, Anniek, Lidy, Laura, Annika, Marit en Maaike. Onze vriendschap is ontstaan bij de into in 2007 bij de doegroep van Piranha. Wie had er toen gedacht dat dit uiteindelijk zo bepalend zou zijn voor een hechte vriendschap. Inmiddels zijn we vele weekendjes (inclusief anschlüss de laatste 2 jaar), sinterkerstennieuws, liters thee, wandelingen, 'meet-in-the-middle' borrels en de eerste bruiloften verder, en ik hoop dat er nog vele mogen volgen van dit alles! Ik vind het ontzettend leuk dat iedereen de moeite blijft doen om elkaar te zien, ondanks dat we niet allemaal bij elkaar om de hoek wonen.

Masha en Karlien, de wereldreizigers. We kennen elkaar al sinds de middelbare school, waarna we allemaal uitgevlogen zijn naar de universiteit. En vervolgens ook naar de rest van de wereld en weer terug. Volgens mij hebben we het zelfs een keer voor elkaar gekregen om allemaal in een ander continent te zijn (exclusief Europa). Ik vind het ontzettend mooi dat we tussendoor altijd de tijd vinden om elkaar weer te zien ergens in Nederland! Onder het genot van een mooie wandeling, fietstocht (Utrecht-Horst) of een andere activiteit, hebben we altijd genoeg om bij te praten.

Bernie, sinds groep 6 zijn we al goede vriendinnen, maar ik zag laatst dat we zelfs al naast elkaar stonden op de foto van de peuterklas. Van vele spelletjes spelen, bakken (die 'zout'koekjes ga ik nooit vergeten), kamperen in de tuin, naar samen (beach) volleyballen, carnaval vieren en nu vaker gewoon gezellig bij kletsen bij iemand thuis of op het terras. Na al de jaren voelt het nog steeds vertrouwd. En ik vind het ontzettend leuk dat je mee hebt gedacht over de cover van mijn proefschrift!

Nicole en Mariska, met twee doctoren aan mijn zij zou het allemaal goed moeten komen toch? Ik vind het ontzettend leuk dat jullie mijn paranimfen willen zijn. Nicole, na al deze jaren kennen we elkaar ontzettend goed. Je bent een enorme harde werker, met grote ambities, maar tegelijkertijd maak je altijd genoeg tijd vrij om leuke dingen te doen. Dit is iets wat ik ontzettend bewonder. Mariska, wat was het fijn om een sparringspartner te hebben de afgelopen jaren waar ik altijd terrecht kon, zowel voor interpretatie van resultaten, de eeuwige rotatiematrix strijd als het zoeken van fouten in een matlab script. Daarnaast was er ook altijd tijd voor een goed gesprek en een mooie roadtrip door Zuid Afrika.

Lieve schoonfamilie, Hans, Trijnie, Femmie, Frans, Kayleigh, Fynn en Chloé. Ondanks dat jullie niet veel verstand hadden van het onderwerp waar ik mee bezig was, hebben jullie altijd wel veel interesse getoond. Bedankt daarvoor! Het is altijd leuk om de reis naar Kampen weer te maken.

Leeve pap, mam, Bas, Gijs, Imke en Vivian. Bedánkt dat jullie er altiëd vur mej zien en altiëd interesse hebbe getoënd in wat ik duj. Het velt soms neet mej um iederiën bej elkaar te kriegen, maar als dát wal gebeurt is het altiëd gezellig. Òk het bourgondische laeve wurd altiëd good gevierd, met de goije kaokkunste van pap en 'n goije fles wiën of beer. Ondanks dat ik al jaore weg bin oet Hôrs, vult 't altiëd nog as thoeskomen.

Lieve Jan Willem. Het eerste lustrum hebben we inmiddels al gevierd, maar wat gaat de tijd toch snel. We hebben al veel avonturen samen beleeft en ik hoop dat er nog veel meer mogen volgen, zowel met als zonder ons busje! Jouw avontuurlijkheid, doorzettingsvermogen en goede klus skills zorgen ervoor dat ik (letterlijk en figuurlijk) verder kom dan dat ik zonder jou had gedaan. Bedankt dat je er altijd voor mij bent. Hou van je!

CURRICULUM VITAE

Laura Peeters was born in Venray on September 20th, 1989. She graduated from secondary school in 2007 and started her study Technical Medicine at the University of Twente. Laura followed the master 'Robotics & Imaging', which included two years of internships. The internships were followed at the Radboud University Medical Center (department of Radiology and department of Nuclear Medicine), Philips Healthcare (department of X-ray and Ultrasound) and the University Medical Center Utrecht (department of Rheumatology & Clinical Immunology), and combined clinical activities with research focused on improving healthcare with the use of technological innovations. She obtained her Master of Science in Technical Medicine in 2014. Afterwards she started her PhD training at the Radboud University Medical Center Nijmegen (department of Rehabilitation). Laura's PhD was part of the Symbionics project, aiming to develop dynamic assistive devices to support the trunk and head in patients with neuromuscular disorders. The project was a collaboration between several universities (Vrije Universiteit Amsterdam, University of Twente and Radboud University Medical Center), companies and patient organizations. The role of bridging the gap between patients and engineers suited Laura very well. During her PhD, Laura presented her work at several (inter)national conferences and won 'the best abstract award' in 2016 at the Dutch conference of Technical Medicine. Laura now lives in 's-Hertogenbosch and works at Acknowledge Health Innovation as consultant health innovation.

LIST OF PUBLICATIONS

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