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Erasmus University Rotterdam
Making Minds Matter

MOVAKIC,
MOTOR ABILITIES
IN CHILDREN
WITH SEVERE
MULTIPLE DISABILITIES

SONJA MENSCH

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MOVAKIC, MOTOR ABILITIES IN CHILDREN WITH SEVERE MULTIPLE DISABILITIES;
– SMALL STEPS, BIG CHANGES –

“MOVAKIC, MOTORISCHE VAARDIGHEDEN VAN KINDEREN
MET ERNSTIG MEERVOUDIG COMPLEXE BEPERKINGEN;
KLEINE STAPJES, GROTE VERANDERINGEN”

PROEFSCHRIFT

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Chapter 3. Mensch S.M., Rameckers E.A.A., Echteld M.A., Penning C., Evenhuis H.M. (2015) Design and content validity of a new instrument to evaluate motor abilities of children with severe multiple disabilities; Movakic. Physical Medicine and Rehabilitation – International 2015; 2(9)

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Chapter 5. Mensch S.M., Echteld M.A., Evenhuis H.M., Rameckers E.A.A. (2016) Construct validity and responsiveness of Movakic; an instrument for the evaluation of motor abilities in children with severe multiple disabilities. Research in Developmental Disabilities 2016; 59: 194-201

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CONTENTS

CHAPTER 1

GENERAL INTRODUCTION

Research in Developmental Disabilities 2015; 47:185-98

   

GENERAL INTRODUCTION

This thesis is focused on the development of an instrument that measures motor abilities in children with severe multiple disabilities. Children with severe motor abilities are characterised by severe or profound intellectual disability and they have very limited motor abilities (Veugelers et al., 2005) (Mergler et al., 2012) (Rieken et al., 2011) (Putten van der, Vlaskamp, Reynders, & Nakken., 2005). Currently, evaluation of motor abilities in children with severe multiple disabilities is often based on subjective evaluation, experience with this group or on instruments developed for other groups of children with motor disabilities (e.g. Gross Motor Function Measure (Ketelaar, Petegem van-Beek van, & Visser., 1999)). Physiotherapists needed a standardized instrument to evaluate the motor abilities of these children clinical practice. The instrument that we intend to develop aims to evaluate the efficacy of therapies and monitor changes in motor abilities throughout time.

My interest in this group of children started in 1992 when Jeroen, the son of a friend, was born. Although Jeroen's parents were worried about some abnormal features they observed, the doctors told the parents that everything was normal. After three months, Jeroen had his first severe epileptic seizure and this event started the medical process. Jeroen was diagnosed with the West syndrome, which manifested itself by the occurrence of infantile spasms. From the moment the epileptic seizures started, his development stagnated. Jeroen never learned to talk or to move and he seemed to have no expression in his face. If you knew Jeroen very well, you would be able to see in his eyes or other small body signs how he felt. I started working as a nurse with Jeroen when he was a little boy of one year old. In that time I studied physiotherapy and worked as a nurse. Besides nursing Jeroen I accompanied Jeroen in a couple of his holidays acting as his volunteer. In all these years I have come to know Jeroen very well and learned so much from this beautiful boy. He taught me things such as to enjoy the small things of life and to explore the meaning of his subtle body signs. In 1999 I started working as a physiotherapist at the children's day-care centre Jeroen visited. My interest in this special group of children was growing every day and it still is. This severely disabled group of children, mostly suffering from major health problems, is a great challenge to work with. For example, a challenge is focussing on possibilities instead of disabilities, which are aimed at positively affecting quality of life. For example, Jeroen had severe motor disabilities; he was not able to e.g. move around, maintain a sitting position or to roll over. However, Jeroen also had some motor 'possibilities' or abilities. Support from others or support from an assistive device can help in facilitating a certain motor ability.

These photos give an impression of Jeroen's possibility to raise his head while sitting in his wheelchair. Jeroen did need the support of his armrest to be able to push himself up and raise his head. In this position he could for example take a bite of his food. Look at the smile on his face after completing this manoeuvre. Jeroen had a big birthday party on his 18th birthday. Unfortunately he died a few days later after complications from his umpteenth pneumonia.

Severe multiple disabilities

There is no uniform description for this group of children in the current literature. Apart from severe multiple disabilities, the terms 'severe neurological impairment and intellectual disability', 'profound and intellectual multiple disability', 'severe motor and intellectual disability' and 'severe (generalized) cerebral palsy' are used (Nakken & den Ouden., 1985) (Mergler et al., 2012) (Proesmans et al., 2015) (Munde & Vlaskamp., 2015) (Liptak et al. 2001) (Calis et al., 2010). I will use the short description 'severe multiple disabilities' and will use the abbreviation SMD in this thesis.

Whereas aetiology of the disorders is variable, severe multiple disabilities are always the result of severe brain damage. A broad range of damaging factors that can be genetic, toxic or traumatic, acting before, during or after pregnancy and birth, can cause these disabilities. Brain damage that occurs before birth, during birth or within a year after birth is called cerebral palsy (Bax et al., 2005). Brain damage, caused by genetic disorders are for example Down syndrome (Down syndrome foundation., 2016), Prader Willi syndrome (Prader-willi syndrome foundation., 2016) or progressive diseases such as the RETT syndrome (Williamson & Christodoulou., 2006), (RETT syndrome foundation., 2012) and the Tay-Sachs syndrome (Kinderneurologie., 2016). Another cause of brain damage can be malformation of the brain, for example due to corpus callosum agenesis (Paul et al., 2007). While these causes are known, in a great number of cases the exact cause is unknown or never found (van Karnebeek et al., 2005).

Children with SMD, as defined in this thesis, are characterised by a severe or profound intellectual disability, comparable to an Intelligence Quotient (IQ) <25. The *Diagnostic and Statistical Manual of Mental Disorders, 5th Edition* (DSM-5) American Psychiatric Association (APA., 2013). The American Psychiatric Association defines intellectual disabilities as neurodevelopmental disorders that begin in childhood and are characterized by (intellectual) difficulties in cognitive functioning as well as difficulties in conceptual, social, and practical areas of living. The terms 'mild', 'moderate', 'severe', and 'profound' are used to describe the severity of the condition. Severe intellectual disabilities manifests themselves as major delays in development; individuals often have the ability to understand speech but otherwise have limited communication skills. Despite being able to learn simple daily routines and to engage in simple self-care, individuals with severe intellectual disabilities need supervision in social settings and often need 'family care' in a supervised setting such as a group home. Persons with profound intellectual disability need continuous close supervision and help with self-care activities. They have very limited ability to communicate and often have physical limitations. Individuals with mild to moderate disability are less likely to have associated medical conditions than those with severe or profound intellectual disabilities.

Children with SMD also have physical limitations characterized by a low level of motor abilities, comparable to level IV or V of the Gross Motor Function Classification System (GMFCS) for children with cerebral palsy (R. Palisano et al., 2007) (Veugelers et al., 2005). Children and youth in Level IV are able to take a sitting posture (usually supported) but self-mobility is limited; they are more likely to be transported in a manual wheelchair or use powered mobility. Children and youth in Level V have severe limitations in head and trunk control and require extensive assisted technology and physical assistance. Self-mobility is achieved only if the child/youth can learn how to operate a powered wheelchair (Palisano, Rosenbaum, Bartlett, & Livingston., 2007).

Furthermore, vitality and quality of their life of these children are threatened by a range of health problems (van Schrojenstein Lantman-de Valk & Walsh., 2008) (Liptak et al., 2001) (Shevell, Dagenais, Hall, & Consortium., 2009), which start at an early age and steadily increase as they grow up. Various studies have demonstrated comorbid conditions in children with SMD such as low bone quality and fractures (Mergler et al.,

2009) (Mergler et al., 2016), dysphagia (Calis et al., 2008), gastro-oesophageal reflux (Bohmer et al., 1999), nutritional state (Rieken, Calis, Tibboel, Evenhuis, & Penning., 2010), sensory problems (Evenhuis, Theunissen, Denkers, Verschuure, & Kemme., 2001), constipation (Veugelers et al., 2010), respiratory problems (Proesmans et al., 2015) (Seddon & Khan., 2003) (Veugelers et al., 2005), scoliosis (Halawi, Lark, & Fitch., 2015) and epilepsy (Dannenberg, Mengoni, Gates, & Durand., 2016).

Prevalence and care

The large majority of Dutch children with intellectual disabilities live with their parents and visit regular or special schools. For those who are not able to attend a school, there is a network of specialised day-care centres. Reasons for admission might be severe behavioural problems, autism spectrum disorders, or severe or profound intellectual disabilities combined with sensory and health problems. In the day-care centres children with SMD are a small but significant subgroup. The prevalence of persons with intellectual disabilities is based on estimations; around 1% of the Dutch population has an intellectual disability of whom 68,000 have a severe intellectual disability (Rijksinstituut voor Volksgezondheid en Milieu (RIVM)., 2013). An inventory by the Dutch Health Care Inspectorate in 2000 showed that of the 2016 children with SMD aged under 18 receiving formal care, 1336 visited day-care centres and 680 lived in residential care (Health Care Inspectorate., 2000). The report also provided information on staffing of specialised day-care centres for this group, showing that in all centres behavioural therapists, intellectual disability physicians, physiotherapists and speech and hearing therapists were employed. In a number of centres occupational therapists and dieticians were active, which reflects the extensive motor and other neurological problems of these children. Almost 90% of the children visiting day-care centres receive physiotherapy (Health Care Inspectorate., 2000).

Paediatric physiotherapy and motor abilities in the context of the ICF-CY

The paediatric physiotherapist uses the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY) (World Health Organization., 2008) in the context of clinical reasoning, policy, documentation and reporting, mono-and interdisciplinary communication, development of guidelines, research and education (Steiner et al. 2002). The ICF-CY is a classification that describes functioning of children and youth in detail from different perspectives, and distinguishes the levels of 'body functions and anatomical characteristics', 'activities' and 'participation'. ICF-CY also contains external factors, the immediate and wider environment of a child. An illustration of the ICF with SMD as example of a health condition is given in figure 1 (World Health Organization., 2008).

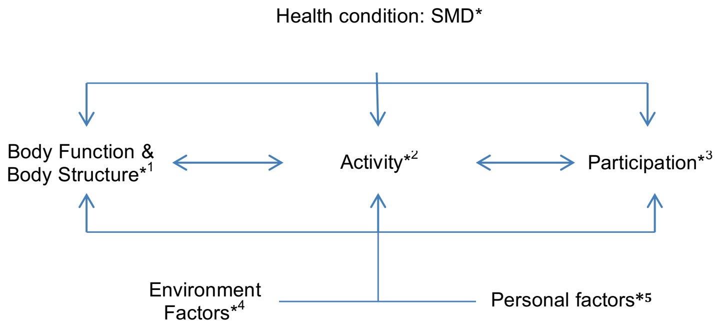

Figure 1. The ICF-CY model (World Health Organization., 2008)

***SMD= severe multiple disabilities**
***1 Body function**
"Psychomotor retardation"
Presence of problems:
Wakefulness, alertness and awareness of the child
Respiration functions
Eating and drinking functions
Motor reflexes
Muscle tone
Spontaneous movements
Postural, balance or threatening reactions

***2 Activities and *3 Participation**
"Unable to participate in daily life situations"
Presence of problems:
Exploring, watching, learning
Comprehending meaning of messages in spoken language, vocalizing
Maintain a body position, change body position, using hands and arms, moving around
Differentiation
Playing

***4 Environmental factors**
Availability of wheelchair, health-care and financial resources
Surroundings: support of other persons such as care-givers and parents

***5 Personal factors**
Coping

ICF-CY: levels. ICF-CY: levels. Treatment goals of physiotherapists will usually address the levels 'activities' and 'participation'. One of the main priorities of treatment is aimed at quality of life of children with SMD; in addition, urgent problems to be addressed are mostly pain, sputum retention, dyspnoea, and establishing the best body position for example during mealtime to avoid dysphagia/aspiration or related to comfort and prevention of contractures. Motor abilities in themselves might be considered a less relevant treatment aim in this group. Nevertheless, much time and energy, both of the children and the physiotherapists, is spent on stimulating and training specific motor abilities. Performing physical activities independently is problematic and the children need direct support of other persons to execute certain motor skills (Putten van der, Bossink, Frans, Houwen, & Vlaskamp., 2016). Despite the lack of internal motivation or initiation of these children (Munde & Vlaskamp., 2015), motor stimulation will help them (to a certain extent) to influence their surroundings such as care-givers or parents, change their position, reach or shift towards a toy, actively participate in eating and other activities and ease personal hygienic care for their parents or other caregivers. So, motor abilities provide children with SMD with a degree of control over their own life and as such may positively influence their quality of life.

In the example of Jeroen, the discovery to the fact that he could raise his head with the proper support of his arm rest resulted in being able to actively participate in feeding. Moreover, by stimulating motor abilities development of (painful) contractures such as scoliosis or hip luxation may be delayed, and bowel function and respiration stimulated.

ICF-CY: Environmental factors and personal factors. In children with SMD all of the processes needed for motor learning are disturbed due to their severe intellectual disability. Motor learning emerges from a complex set of processes including perception, cognition and action. Motor learning needs to be trained in the needed specific skills and in the specific environment that requires using these skills (Shumway-Cook & Woollacott., 1995). Therefore, especially in children with SMD, intervention needs to be implemented in all functional activities of daily life involving parents and other care-givers in order to create a maximize repetition and in turn learning.

Measurement

It is understandable that signs of psychomotor development and effects of treatment in this group will frequently be difficult to observe and may be very subtle or will develop over a lengthy period. Therefore it is possible that subtle, but nevertheless meaningful motor development or effects of intervention might be missed; the same applies to recognizing gradual deterioration. Even parents may not recognize or misinterpret subtle, gradual changes. Moreover, in clinical practice working with children with SMD there is a lack of knowledge about the relevance and importance of motor abilities for these children. Therefore, since many years, there is a need to objectively measure motor function during a longer time frame as a basis for treatment plans, collaboration with parents, but also to obtain objective insight into the quality and results of professional care. In the example of Jeroen, an instrument feasible for these children should help us focus on his relevant motor abilities, evaluation of changes and treatment goals enabling him to reach a degree of control in his own life and as such have a positive influence on the quality of his life.

In our setting we commonly use the Gross Motor Function Measure (GMFM) (Russell et al., 1989) (Russell, Rosenbaum, Avery & Lane., 2002) (Ketelaar et al., 1999). Although this instrument has been specially developed for children with cerebral palsy in mind, it has been evaluated in children with Down Syndrome (Russell et al., 1998). However, clinical experience has shown us the limitations of this instrument in children with SMD such as the use of items that describe motor functions that are not relevant in severely disabled children and the lack of sensitivity in scoring subtle, gradual changes in motor function. In addition, the study of Russell et al confirms our clinical experience; children with Down Syndrome who were younger and had mild motor impairment changed significantly more than those who were older and had moderate or severe motor impairment. In general, also internationally, in the last decade there has been a growing wish in the field of physiotherapy, to objectify changes in motor abilities in clinical practice in order to support decision-making in (expensive) interventions offered to this group (Tieman, Palisano, & Sutlive, 2005) (American Physical Therapy Association., 2016).

Therefore, in 2005, I started thinking about a measurement instrument for motor abilities of children with SMD. I was encouraged by the support of my colleague physiotherapists as well as the management in care organisation Ipse de Bruggen to develop such an instrument. The research project on its design and evaluation was performed at the Erasmus MC.

CONTENTS OF THIS THESIS

The aim of my thesis was to contribute to the physiotherapeutic care of children with SMD by developing a feasible and validated instrument that evaluates motor abilities in these severely disabled children. Before starting with the development of a new instrument we had to be sure that there was no other instrument available. Therefore, this thesis starts with a systematic review of the scientific literature, aimed at the identification of existing measurement instruments for motor abilities in children with disabilities in general (chapter 2). From this search we found eight instruments, developed for children with disabilities.

In chapter 3 we started with an evaluation of applicability of these instruments for children with SMD, we described the step-by-step designing process of a new instrument and we ended this chapter with the presentation of its first draft, as well its name: Movakic (**mo**tor e**va**luation in **k**ids with **i**ntellectual and **c**omplex disabilities).

Before implementing Movakic in practice its statistical properties had to be assessed. Chapter 4 and 5 form the core of this thesis, presenting the reliability (chapter 4) and validity and responsiveness (chapter 5) of Movakic. Data on these psychometric properties of Movakic were obtained in a longitudinal study, with the help of a range of Dutch physiotherapists and their clients with SMD.

After this study on statistical properties we applied Movakic in a clinical study. One of our hypotheses was that motor abilities affects quality of life. In chapter 6, results are presented of the relationships between motor abilities and quality of life of children with SMD.

The general discussion in chapter 7 describes the main results of this research project and other relevant findings observed during its development and evaluation, including critical reflections and recommendations for the implementation of Movakic in clinical practice as well as for intervention studies using the instrument.

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CHAPTER 2

INSTRUMENTS FOR THE EVALUATION OF MOTOR ABILITIES FOR CHILDREN WITH SEVERE MULTIPLE DISABILITIES: A SYSTEMATIC REVIEW OF THE LITERATURE

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ABSTRACT

Based on a systematic review, psychometric characteristics of currently available instruments on motor abilities of children with disabilities were evaluated, with the aim to identify candidates for use in children with severe multiple (intellectual and motor) disabilities. In addition, motor abilities are essential for independent functioning, but are severely compromised in these children.

The methodological quality of all studies was evaluated with the Consensus Based Standards for the Selection of Health Status Measurement Instruments (COSMIN) Checklist; overall levels of evidence per instrument were based on the Cochrane Back Review Group strategy.

As a result, 18 studies with a total of eight instruments, developed for children with cerebral palsy (CLA, GMFM-88 and LE85), spinal muscular atrophy (MHFMS), neuromuscular diseases (MFM), disabilities 0-6 years (VAB, WeeFIM), and one developed specifically for children with severe multiple disabilities (TDMMT) were found.

Strong levels of evidence were found for construct validity of LE85 and MFM and for responsiveness of WeeFIM, but reliability studies of these instruments had a limited methodological quality. Up to now studies of the TDMMT resulted in limited and unknown evidence for structural validity due to the poor methodological quality of reliability studies.

In a next step, the clinical suitability of the instruments for children with severe multiple disabilities will be evaluate.

INTRODUCTION

The need for a specific evaluation instrument for motor abilities of children with severe multiple disabilities (SMD) has both nationally and internationally been recognised by physical therapists. Children with severe multiple disabilities (SMD) suffer from profound intellectual disabilities (IQ <25) and have a level of motor abilities comparable to level IV/V of the Gross Motor Function Classification System (GMFCS) for children with cerebral palsy (Palisano et al., 1997; Veugelers et al., 2005): they typically are wheelchair-bound and only a few are able to crawl or move with a physical aid (Mergler et al., 2012; Rieken et al., 2011). These children are most often severely limited in maintaining their body position or transferring into another position. In addition, children with SMD may have multiple sensory disorders and other co-morbidity such as swallowing and respiratory problems, epilepsy and contractures. As a result, children with SMD are fully dependent on their social and material environment for all activities of daily life.

Motor abilities are essential for independent functioning, but are severely compromised in these children. Optimal development requires a highly structured environment with constant aid, supervision, extensive support technology and physical assistance (American Psychiatric Association, 2000). Independent mobility is only achieved if the child can learn how to operate a powered wheelchair (Palisano et al., 1997). Children with SMD often have to be manually activated and supported by caregivers to use their motor abilities. It has to be stressed that in the opinion of clinicians working in this field, even subtle motor abilities provide these children with a degree of control over their environment and may as such have influence on their quality of life.

For the purpose of evaluation of interventions, measurement of motor abilities is essential. In current clinical practice, the evaluation of motor abilities by physical therapists is mostly based on subjective assessments in children with SMD or on instruments developed for other target groups. Nevertheless, standardised instruments developed for children with SMD with adequate psychometric properties are lacking. For this reason, an overview of psychometric properties is presented in this article of instruments that are not specifically developed for children with SMD, but for generic groups of children with severe disabilities.

METHODS

We performed a systematic review of published articles evaluating psychometric characteristics of instruments for the measurement of gross motor abilities of children with severe disabilities (age range 2-18).

Search strategy

A literature search in Pubmed (1966 - January 2014), Embase (1980 - January 2014), Web of science (1975 - January 2015), and PsycINFO (1985 - January 2015) was performed in February 2015. Keywords were used alone or separately and if available medical subject heading (MeSH) terms were used. In order to maximise the chance of finding all relevant instruments, a generic set of keywords with a wide range of search terms was chosen to describe the group of persons with disabilities (disabled persons[mesh] OR disabil*[tw] OR disabl*[tw] OR retard*[tw] OR handicap*[tw] (AND (child*[tw] OR infan*[tw] OR pediatr*[tw] OR paediatr*[tw] OR juven*[tw] OR newborn*[tw] OR neonat*[tw] OR (adolescen*[tw] NOT (adult[mesh] OR adult*[tw]). Instruments with different aims such as 'evaluative', 'predictive', 'population specific' and 'disease specific' were searched, in order to provide a complete overview of available information on existing instruments (measurement*[tw] OR measuring[tw] OR measure[tw] OR scale*[tw] OR scaling*[tw] OR evaluat*[tw] OR assess*[tw] OR questionnaire*[tw] OR checklist*[tw] OR protocol*[tw]) AND (psychometr*[tw] OR reliab*[tw] OR valid*[tw] OR responsiv*[tw] OR sensibilit*[tw]). The focus of the search was based upon instruments evaluating motor abilities on the activity level (motor skill*[tw] OR gross motor function*[tw] OR functional mobilit*[tw] OR motor abilit*[tw] OR motor behavio*[tw] OR motor activit*[tw] OR physical activit*[tw] OR locomotor activit*[tw] OR motor performanc*[tw] OR physical performanc*[tw] OR physical mobil*[tw]), according to the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY) (World Health Organization, 2008).

Article and instrument selection

All English, Dutch, German or French articles including a description of an instrument for the evaluation of motor abilities in children with disabilities were selected. Articles including descriptions of instruments beyond the capacities of the target population were excluded, i.e. instruments that (a) were developed for school children (e.g. Development Coordination Disorder); (b) only measure aspects that are not relevant to children with SMD (e.g. sports, arm-hand function); (c) focus on the level of participation (such as sports or school) or on the level of body functions (such as muscle tone or range of motion) (ICF-CY); (d) have been developed as classification instruments only; (e) measure quality of life only or (f) normative measurements.

Inclusion or exclusion of articles according to these criteria was done following predetermined steps. First, one author (SM) performed a selection based on titles. Second, two reviewers (SM, ER) independently screened the abstracts of the remaining articles and third, a consensus was reached about the in- or exclusion of the articles by both raters. In case of disagreement, further judgment was based on the full text independently by both raters, after which consensus was achieved. If the information in the full text articles proved inconclusive, additional information on the instruments was collected from their authors or manuals.

Quality assessment and psychometric properties

Methodological quality of the studies was evaluated using the Consensus Based Standards for the Selection of Health Status Measurement Instruments (COSMIN) protocol, providing standardised criteria for stepwise rating of methodological quality in a range of excellent to poor (Mokkink et al., 2010; Table 1). An example of a COSMIN box in appendix A (Terwee et al., 2007) was added.

Table 1. Methodological quality criteria of the COSMIN boxes (Terwee et al., 2012).

Rating	Quality criteria
Excellent	All relevant COSMIN items are scored adequate
Good	Some things are not reported, but one can assume these issues are adequate, f.e. if it can be assumed that patients were not changed in a test-retest study, but this was not explicitly investigated
Fair	The value of the psychometric property could have been under estimated or estimated in a moderate sample or when there were other minor flaws in the design or analyses, f.e. reliability could have been underestimated due t the unstable patients or a long time interval in test-retest design
Poor	The results are not to be trusted because of major flaws in the design or statistical analyses, f.e. small sample size or inappropriate statistical methods

Statistical outcomes were rated according to the criteria, shown in Table 2, as good (+), negative (-), and indeterminate (?) (Terwee, et al., 2007).

Table 2. Quality criteria for rating of the results (Terwee, et al., 2007).

Psychometric property	Rating	Quality criteria
Internal consistency	+	Cronbach alpha >.70
	-	Cronbach alpha <.70
	?	Cronbach alpha not determined
Reliability	+	ICC >.70 OR r>.70
	-	ICC <.70 OR r<.70
	?	Neither ICC nor weighted kappa
Measurement error	+	MIC > SDC OR MID > SDC OR MIC outside the LoA
	-	MIC < SDC OR MID < SDC OR MIC equals or inside LoA
	?	MIC not defined
Content validity	+	The target population considers all items in the questionnaire to be relevant AND considers the questionnaire to be complete
	-	The target population considers items in the questionnaire to be irrelevant OR considers the questionnaire to be incomplete
	?	No target population involvement
Structural validity	+	Factors should explain **>** 50% of the variance
	-	Factors explain **<** 50% of the variance
	?	Explained variance not mentioned
Hypothesis testing	+	Correlation with an instrument measuring the same construct **> .**50 OR **>**75% of the results were in accordance with the hypotheses AND correlation with related constructs was higher than with unrelated constructs
	-	Correlation with an instrument measuring the same construct **>** 50 OR **<**75% of the results were in accordance with the hypotheses OR correlation with related constructs was lower than with unrelated constructs
	?	Solely correlations determined with unrelated constructs
Cross cultural validity	+	Original factor structure confirmed OR no important DIF between language versions
	-	Original factor structure not confirmed OR important DIF found between language versions
	?	Confirmatory factor analysis not applied and DIF not assessed
Criterion validity (predictive or concurrent)	+	Convincing arguments that gold standard is "gold" AND correlation with gold standard **>**.70
	-	Correlation with gold standard **<**.70 despite adequate design and method
	?	No convincing arguments that gold standard is "gold" OR doubtful design or method

Table 2. (continued)

Psychometric property	Rating	Quality criteria
Responsiveness	+	Correlation with an instrument measuring the same construct >.50 OR at least 75% of the results are in accordance with the hypotheses OR AUC > 0.70 AND correlation with related constructs is higher than with unrelated constructs
	-	Correlation with an instrument measuring the same construct <.50 OR <75% of the results are in accordance with the hypotheses or AUC <0.70 OR correlation with related constructs is lower than with unrelated constructs
	?	Solely correlations determined with unrelated constructs

ICC = intraclass correlation coefficient, r = Cohen (weighted) kappa, SDC = smallest detectable change, MIC = minimal important change, MID = minimal important difference, LoA = limits of agreement, DIF = differential item functioning, AUC = area under the receiver operating characteristic curve
Rating: + = positive rating, - = negative rating, ? = indeterminate rating

Two reviewers independently rated the included studies, after which consensus was obtained.

Determining of overall level of evidence of each psychometric property per instrument

The method of acquiring evidence for each studied psychometric property of an instrument was based on the strategy of the Cochrane Back Review Group. The overall level of evidence was scored as strong (consistent findings in multiple studies of good methodological quality or in one study of excellent methodological quality), moderate (consistent findings in multiple studies of fair methodological quality or in one study of good methodological quality), limited (one study of fair methodological quality), conflicting (conflicting findings), or unknown (only studies of poor methodological quality) (Dobson et al., 2012; Tulder van, Furlan, Bombardier & Bouter, 2003).

RESULTS

Search and article selection

The results of the literature search and selection of instruments are shown in the flow chart in Figure 1.

Quality assessment of methodology and outcomes

The investigated psychometric properties, methodological quality of the studies, and rating of their statistical outcomes are listed in Table 4.

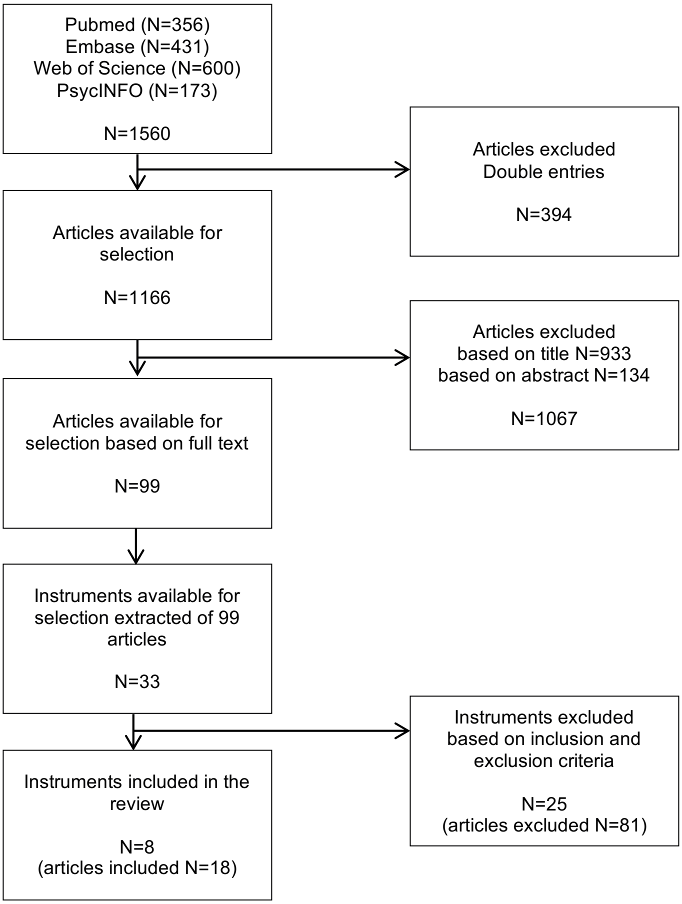

Figure 1. Flow chart for literature search and instrument selection

Instrument selection

Eight measurement instruments were identified in 18 studies for which one or more psychometric properties were evaluated in totally 18 studies: 1) The Chailey Levels of Ability (CLA), 2) Gross Motor Function Measure (GMFM-88), 3) Modified Hammersmith Functional Motor Scale (MHFMS), 4) Lower Extremity physical functioning and mobility skills (LE85), 5) Motor Function Measure scale (MFM), 6) Top Down Motor Milestone Test (TDMMT), 7) Vulpe Assessment Battery (VAB) and 8) Functional Independence Measure for Children (WeeFIM). One instrument, the TDMMT, was specifically developed for children with SMD. Characteristics such as target group, aim, method, construct of the instrument and ICF-CY domain of the eight included instruments are shown in Table 3.

Table 3. Characteristics of the 8 Selected Instruments

Name instrument	Target group	Characteristics				
		Aim[1]	Method	ICF-CY[2]	Construct	Reference
The Chailey Levels of Ability (CLA)	Individuals with motor impairments. Children and adults with low levels of physical ability	Prescriptive Evaluative	Observation	Activity	Measurement scale documenting stages of motor development in the positions of lying, sitting and standing. As part of a postural management approach aiming tot promote normal movement, improve practical ability and reduce the progression of deformity.	(Pountney, Cheek, Green, Mulcahy, & Nelham, 1999; Poutney, Mulchahy, Clarke, & Green, 2004)
Gross Motor Function Measure (GMFM-88)	Children with cerebral palsy	Evaluative	Observation	Activity	Capacity of gross motor function in 5 domains (lying and rolling (1), sitting (2), crawling and kneeling (3), standing (4), walking, running and jumping (5)); 88 items (4 point likert scale).	(Russell et al., 1989) (Ketelaar, Petegem van-Beek van, & Visser, 1999)
Modified Hammersmith Functional Motor Scale (MHFMS)	Children with spinal muscular atrophy	Evaluative Classification	Observation	Activity	Disease specific scale; 20 items based on ability to perform gross motor tasks (3-point scale).	(Krosschell, et al., 2011) (Main et al., 2003)
Lower extremity physical functioning and mobility skills (LE85)	Children with cerebral palsy	Evaluative	Observation	Activity and participation	85 items reflecting transfers, basic mobility and ambulation skills with and without assistive devices.	(Gorton et al., 2010)
Motor function measure scale (MFM)	Patients with neuromuscular diseases	Evaluative	Observation	Activity	32 item scale to assess severity and disease progression of neuromuscular diseases. 4-point likert scale based on maximal abilities without any assistance.	(Berard, Payan, Hodgkinson, & Fermanian, 2005)

Table 3. (continued)

Name instrument Target group		Characteristics				
		Aim[1]	Method	ICF-CY[2]	Construct	Reference
Top Down Motor Milestone Test (TDMMT)	Individuals with severe and profound multiple disabilities	Evaluative	Observation	Activity	Consists of the movement skills sitting, standing and walking that are assumed to be the physical skills required in order to accomplish functional skills, such as expressive language and self-care. The movement skills are structured into 16 categories. It consists in total of 74 items, sequenced according in four levels of function, which describe the amount of independent mobility.	(Putten van der et al., 2005)
Vulpe Assessment Battery (VAB)	Children with disabilities 0-6 years	Evaluative	Observation	Body function and activities	The items of the VAB-GM (Gross Motor) are similar to the PDMS-GM. Based on a 1-7 point ordinal scale Composed of six sub tests. Reflexes, locomotion (89 items), balance (30 items), receipt and throwing objects (24 items).	(Jain, Turner, & Worrell, 1994)
Functional Independence Measure for Children (WeeFIM)	Young children with physical, cognitive or developmental impairments from birth to 3 years of age	Discriminative Evaluative	Proxy report	Participation and activities	The 36 items measure early function in 3 domains: motor (16 items), cognitive (13 items) and behavioral perceptions (7 items). The motor domain measures physical functioning. Each item is rated on a 3-level ordinal scale.	(Niewczyk & Granger, 2010)

[1] Aim of an instrument: Evaluative instruments measure longitudinal change. Predictive instruments are used classify individuals against an external criterion and are intended for diagnostic, prognostic or screening purposes. Discriminative instruments are used to quantify differences between individuals when no external criterion exists. (Hankins, 2008)
[2] International Classification of Functioning, Disability and Health for Children and Youth (World Health Organization, 2008)

Table 4. Psychometrics of the included instruments

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
The Chailey Levels of Ability (CLA)	(Pountney et al., 1999)	CP	85	1.5-18	Content validity	The nine constructs outlined in the assessment were identifiable	Poor	+
			29	4.5-19	Construct validity	Pearsons AIMS .90-.97 Pearsons GMFM .85-.96	Poor	+
Gross Motor Function Measure (GMFM-88)	(Ketelaar et al., 1999; Dutch version)	CP	111	1.6-10.11	Internal consistency	ICC= .90-.98	Poor	+
			16		Test-retest reliability	ICC= .96-.99	Poor	+
			18		Inter-rater reliability	ICC= .75-.99	Poor	+
			150		Responsiveness	Kendall's tau .45 F(4,99)=2.56,p<.05 Student t-test t(27)=4.7, p<.001	Poor	+
	(Russell et al., 1989)	CP	10	<3- >9	Intra-rater reliability	ICC= .99	Poor	+
			11		Inter-rater reliability	ICC= .99	Poor	+
		CP/ ABI/ ND	47/ 22/ 28		Responsiveness	Correlations of GMFM total score with change as judged by external therapists/ parents/ blind video analysis: .65/.54/.82 (p<.05)	Poor	+
	(Russell et al., 1998)	DS	22	0.4-5.6	Test-retest reliability	ICC standard/reported score = .95/.96	Poor	+
					Inter-rater reliability	ICC standard/reported score = .96/.98	Poor	+
		CP/ ABI/ ND	106/69/ 30	0.1-6	Responsiveness	Correlation of GMFM total score with change as judged by parent/intervenor/ video: standard scoring = .16/.24/.29 Reported scoring = .32/.30/.34	Fair	-

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
Gross Motor Function Measure (GMFM-88)	(Russell et al., 1998)	CP/ ABI/ ND	13-27	<3-6	Responsiveness	Three-way ANOVA indicated a significant different pattern in GMFM and BSID-II (p<.05)	Poor	?
	(Björnson, Graubert, Kerfeld, & Clark, 1998)	CP	21	4.4-17.7	Test-retest reliability	ICC=1.00	Poor	+
	(Vos-Vromans, Ketelaar, & Gorter, 2005)	CP	55	2-7	Responsiveness	Correlations between GMFM total score and changes in global rating scale for parents total group: R= .50 (p<.01) ES .59 SRM .88	Fair	-
Modified Hammersmith Functional Motor Scale (MHFMS)	(Krosschell et al., 2011)	SMA type II	22	<2.6	Test-retest reliability	ICC = .96	Poor	+
					Measurement Error	SEM = 1.86	Poor	?
	(Main et al., 2003)	SMA	35		Inter-rater reliability	>99%	Poor	?
	(Mazzone et al., 2014)	SMA type II and III	74	2.6-28.1	Construct validity	Correlation between HMFS and MFM20 at baseline r=.73 (p<.0001)	Poor	+
					Responsiveness	Correlation between HMFS and MFM20 after 12 months .48 (p<.001)	Poor	-

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
Modified Hammersmith Functional Motor Scale (MHFMS)	(Glanzman et al., 2011)	SMA Type II and III	70	4.6-15.7	Construct validity	Correlation between HFMSE and GMFM r= .98, CI .97-.99, p<.0001	Fair	+
			56			Correlation between HFMSE and forced vital capacity r= .87, CI .78-.92, p<.0001	Fair	+
			60			Correlation between HFMSE and Knee Extension Myometry r= .74, CI .60-.84, p<.0001	Fair	+
		SMA Type II and III	58/ 12			HFMSE scores higher in subjects needed <8 hours bi-level positive airway per day (median = 23, IQR =9-50) –vs at least 8 hours per day (median =3, IQR = 0-7.5)	Poor	?
		SMA Type II and III	25/ 44			HFMSE scores higher in ambulatory subjects (median=52, IQR = 48-57, p<.0001) vs non-ambulatory (median=8, IQR 2-18.5, p<.0001)	Poor	?
		SMA Type II and III	33/ 37			HFMSE scores higher in SMA type III (median 49, IQR 29-55, p<.0001) vs SMA type II (median=8, IQR=2-17, p<.0001)	Poor	?
	(O'Hagen et al., 2007)	SMA type II and III	38/ 17		Intra-rater reliability	Combined group SMA type II and III/ SMA Type III HFMS ICC=.98 / .97 HFMSE ICC=.99/ .99	Poor	+

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
Modified Hammersmith Functional Motor Scale (MHFMS)	(O'Hagen et al., 2007)	SMA type II and III	38		Construct validity	Correlation between HMFSE type II-III and GMFM/ functional rating score r= .97 / .90	Poor	+
			17			Correlation between HMFSE type III and GMFM/ functional rating score r= .94 / .88	Poor	+
Lower extremity physical functioning and mobility skills (LE85)	(Gorton et al., 2010)	Spastic CP	308	2-20	Measurement error	The LE85 had good correlation with the Paediatric Outcomes Data Collection Instrument, Functional Independence Measure for Children, Gillette Functional Assessment Questionnaire, and Paediatric Quality of Life Inventory – CPmodule (range r=.63–.86).	Good	+
			170		Construct validity	Correlation between LE85 and FAQ walking scale r=.82, 95%CI .77-.87, p<.0001	Excellent	+
			90			Correlation between LE85 and average score 22-item skill set r=-.86, 95%CI -.8 to -.91, p<.0001	Good	+
			176			Correlation between LE85 and PODCI r=.85, 95%CI .81-.89, p<.0001	Excellent	+
			120			Correlation between LE85 and WeeFIM r=.83, 95%CI .77-.88, p<.0001	Excellent	+

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
Lower extremity physical functioning and mobility skills (LE85)	(Gorton et al., 2010)	Spastic CP	81		Construct validity	Correlation between LE85 and Paediatric Quality of Life Inventory CP overall score r=-.76, 95% CI -.65 to -.84, p<.0001	Good	+
Motor function measure scale (MFM)	(Berard et al., 2005)	Neuro-muscular diseases	303	6-62	Internal consistency	Cronbach's alpha coefficient 0.99	Fair	+
			50		Inter-rater reliability	Cohen's kappa coefficients ranged from .81 to .94 (excellent) for nine items, from .61 to .80 (good) for 20 items and from .51 to .60 (moderate) for only three items.	Poor	-
					Intra-rater reliability	Agreement coefficients for intra-rater test–retest were excellent (0.81–0.94) for 25 items, and good (0.61– 0.80) for seven items.	Poor	+
					Test-retest reliability	The intra- and inter-rater agreement coefficients (ICC) were all excellent (.96–.99) for the total score and for the three dimensional sub-scores.	Poor	+
					Content validity	Therapists estimated that the items of the scale assessed severity of motor disability correctly and thoroughly	Poor	?

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	**N**	**Age (Y)**	**COSMIN property**	**Results**	**Rating COSMIN boxes*[1]**	**Rating of results*[2]**
Motor function measure scale (MFM)	(Berard et al., 2005)	Neuro-muscular diseases	50	2-62	Construct validity	Spearman rank correlation coefficients MFM-total score –VAS physician/VAS physiotherapist/ Vignos grade/ Brooke grade/ FIM (.88/ .91/ .91/ .85/ .91)	Excellent	+
						Mean scores significantly decreased with degree of motor disability as evaluated by the Clinical Global Impression of physicians (Anova F3.299= 293.2, P< .0001—all means significantly different in pairwise comparisons).	Excellent	?
						The total score and sub-scores allowed a good discrimination between diagnosis groups (Anova F7.295= 29.1, P< .0001)	Excellent	?
Top Down Motor Milestone Test (TDMMT)	(Putten van der et al., 2005)	Multiple disabilities	3	6, 11, 13	Internal consistency	Cronbach's alpha of the categories was .95. The item-rest correlations varied between .54 (category M) and .90 (category G), with the mean .80.	Poor	+
			3	6-13	Inter-rater reliability	Cohen Kappa= .97 (.58, 1.00)	Poor	+
					Intra-rater reliability	Cohen Kappa= .95 (.54-.90)	Poor	+

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*1	Rating of results*2
Top Down Motor Milestone Test (TDMMT)	(Putten van der et al., 2005)	Multiple disabilities	66	2-16	Structural Validity	When three factors were extracted, 83% of the total variance could be explained. Factor 1 explained 71%, factor 2 explained 8% and factor 3 explained 5% of the total variance.	Fair	+
Vulpe Assessment Battery (VAB)	(Jain et al., 1994)	CP	13	1.11-5.10	Intra-rater reliability	ICC = .87, SEM 12.09	Poor	+
					Construct validity	Correlation between Peabody-GM and Vulpe-GM r=.97	Poor	+
Functional Independence Measure for Children (WeeFIM)	(Niewczyk & Granger, 2010)	Children with im-pairments	147	0-3	Internal consistency	Cronbach's alpha = .95	Good	+
					Structural validity	Rasch analyses; For the well-fitting motor items, the infit MNSQ statistics ranged from 0.61 to 1.16 and outfit MNSQ ranged from 0.19 to 1.22	Excellent	?
					Construct validity	The total WeeFIM 0-3 score was significant in predicting impairment status: B=-.79 (SE=.01),	Excellent	+
						Findings were significant, indicating 3 components which accounted for 68% of the variance in the model;	Excellent	+

Table 4. (continued)

Instrument	Study	Study population			Psychometrics		Methodological quality	
		Diagnoses	N	Age (Y)	COSMIN property	Results	Rating COSMIN boxes*[1]	Rating of results*[2]
Functional Independence Measure for Children (WeeFIM)	(Ziviani et al., 2001)	ABI Spina Bifida Chromo-somal and genetic abnormal-ities	41	1.6-9.5	Inter-rater reliability	ICC = .81-.95	Poor	+
					Construct validity	Correlation between PEDI and WeeFIM r=.53-.96	Poor	-
	(Otten-bacher et al., 2000)		174	0.11-6.3	Responsiveness	Of total score RCI=2.82, PCI=1.41, ES=.62, SRM=1.31, p=<.01	Excellent	?

Diagnfoses: CP = Cerebral Palsy, ABI = Acquired Brain Injury, ND = Non Disabled, DS = Down Syndrome, SMA = Spinal Muscular Atrophy
Results: r = Spearman Rank Score, CI= Confidence Interval, IQR = Inter Quartile Range, FAQ = Functional Assessment Questionnaire, RCI= Reliability Change Index, PCI=Proportional Change Index, ES= Effect Size (d-index), SRM= Standardized Response Means
*[1]Rating COSMIN boxes: excellent = all relevant items are scored adequate, good = some things are not reported but one can assume that these issues are adequate, fair = the value of the measurement property have been underestimated or estimated in a moderate sample size or when there were other minor flaws in the design or analysis, poor = the results are not to be trusted because of major flaws in the design or statistical analyses
*[2]Rating of results: + = positive rating, - = negative rating, ? = indeterminate rating

The table shows that intra-rater reliability, inter-rater reliability and construct validity were most frequently evaluated, whereas none of the studies addressed cross-cultural validity and criterion validity. The methodological quality was poor in 35, fair in 7, good in 4 and excellent in 10 out of 56 sub-studies. Only for the LE85, MFM, and WeeFIM, (part of) the studies had excellent or good methodological quality.

Overall level of evidence for each instrument

The overall level of evidence, synthesizing methodological quality and outcomes of all studies about that instrument, is presented for each psychometric property in Table 5. Strong levels of evidence were found for construct validity of LE85, MFM, and WeeFIM, but reliability studies were missing (LE85) or of poor quality for these instruments.

Table 5. Synthesis of Quality rating per instrument

Domain	**Reliability**					**Validity**						
Measurement property Instrument	**Internal con sistency**	**Test-retest**	**Intra-rater**	**Inter-rater**	**Measurement error**	**Content**	**Face**	**Structural**	**Construct**	**Cross-cultural**	**Criterion**	**Responsiveness**
CLA						?			?			
GMFM		+	+	+								+/-
MHFMS		+	+	+	+				+/-			+
LE85					++				++			
MFM	++	?	?	?			?		+			
TDMMT	?		?	?				+				
VAB			+						+			
WeeFIM	++			+					+++			?

Measurement property was rated* as:

+++ or --- Strong evidence. Consistent findings in multiple studies of good methodological quality or in one study of excellent methodological quality

++ or -- Moderate evidence. Consistent findings in multiple studies of fair methodological quality or in one study of good methodological quality

+ or - Limited evidence. One study of fair methodological quality

+/- Conflicting evidence. Conflicting findings

? Unknown evidence. Only studies of poor methodological quality

* + = positive rating, - = negative rating.

CLA = The Chailey Levels of Ability. GMFM = Gross Motor Function Measure. MHFMS = Modified Hammersmith Functional Motor Scale. LE 85 = Lower Extremity physical functioning and mobility skills. MFM = Motor Function Measure scale. TDMMT = Top Down Motor Milestone Test. VAB = Vulpe Assessment Battery. WeeFIM = Functional Independence Measure for Children

DISCUSSION AND CONCLUSION

This systematic review identified eight instruments for the measurement of motor abilities in children with severe disabilities on the activity level. Their psychometric properties were evaluated in 18 studies, mostly covering intra-rater reliability, inter-rater reliability and construct validity. Only two instruments were studied in all three domains (reliability, validity and responsiveness): the MHFMS (Krosschell et al., 2011) (Main, Kairon, Mercuri, & Muntoni, 2003) and WeeFIM (Niewczyk & Granger, 2010). Limited, conflicting or unknown evidence was rated for the majority of studied psychometric properties. Strong levels of evidence were only found for construct validity of LE85, MFM, and WeeFIM, but reliability studies were missing (LE85) or of poor quality for these instruments.

The strength of this review was our stepwise selection procedure. We used a generic set of key words to describe the group of persons with disabilities, after which specific in- and exclusion criteria were applied, based on the limited capacities of children with severe disabilities. In paediatric physical therapy, motor abilities of disabled children are often compared to those of children with normal development (Bond, 1996). Due to the severity of the abilities in children with SMD, norm-referenced tests are irrelevant. Evaluative criterion-referenced construct of motor abilities of the child should be the main purpose.

As was to be expected, the identified instruments were mostly developed and validated for children with many common specific diagnoses such as cerebral palsy and neuromuscular diseases. One instrument, the Top Down Motor Milestone Test (TDMMT) was specifically developed for children with SMD (Putten van der, Vlaskamp, Reynders, & Nakken, 2005). The TDMMT consists of the movement skills sitting, standing and walking, that are assumed to be the essential physical skills required in order to accomplish functional skills, such as expressive language and self-care. The design of the test seems to fit the needs of the target population, which makes it an interesting candidate for our purpose, but up to recently, its psychometric properties have been insufficiently studied: its test-retest reliability and responsiveness should be evaluated, allowing conclusions regarding the possibility of determining changes over time.

Based on this study, we conclude that currently, completely validated instruments applicable to children with severe disabilities are lacking, but several show potential. Generalization of the available instruments to children with SMD is difficult because of the limited motor capacities in these children, including the severity of the intellectual disabilities. Furthermore, these children have severe limitations in head and trunk control and often require extensive support technology, manual activation and physical support to use their motor abilities. An accurate measurement for children with SMD will have a positive effect in the therapeutic intervention whereby the appropriate level of motor abilities can be determined and therapeutic goals connected. Acquiring some motor abilities is relevant; for example, changing body position may be only possible with the help of another person. By practicing the ability to actively participate in rolling over from lying on the back to lying on the side, the child can influence its own care and this active participation will be positive for the child's quality of life. Moreover, systematic objective evaluation of such changes in motor abilities provides insight into children's functioning, and supports decision-making in goal-oriented intervention (Palisano et al., 2012). An instrument for this specific group, with the aim of longitudinal evaluation of progression, stabilization, or deterioration of motor abilities, to be applied in clinical physical therapeutic practice, is needed.

The next step towards our purpose of the ideal instrument for children with severe multiple disabilities, we will be developing criteria for clinical suitability in this specific group and applying them to the found eight instruments. Important criteria for clinical suitability will be the possibility to score manual support, the ability to score subtle changes in motor abilities and the ability to score relevant low-level motor abilities, for example raising the head and maintaining a position. This step can be a starting point in deciding whether an existing test can be used, maybe after modification and high-quality study of its psychometric properties, or a new test has to be developed. In our opinion, we need the involvement of experts working in the field of this specific target group in order to formulate suitability criteria for a test for children with SMD, which would result in a well-defined procedure and a clear definition of the construct of the test.

Declaration of interest

We certify that no party having a direct interest in the results of the research supporting this article has or will confer a benefit on us or on any organization with which we are associated and, if suitable, we certify that all financial and material support for this research and work are clearly identified in the title page of the manuscript.

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Appendix A. Example of a COSMIN Box (Terwee, et al., 2012)

Box B. Reliability: relative measures (including test-retest reliability, inter-rater reliability and intra-rater reliability)				
Design requirements	**Excellent**	**Good**	**Fair**	**Poor**
1 Was the percentage of missing items given?	Percentage of missing items described	Percentage of missing items NOT described		
2 Was there a description of how missing items were handled?	Described how missing items were handled	Not described but it can be deduced how missing items were handled	Not clear how missing items were handled	
3 Was the sample size included in the analysis adequate?	Adequate sample size (≥100)	Good sample size (50-99)	Moderate sample size (30-49)	Small sample size (<30)
4 Were at least two measurements available?	At least two measurements			Only one measurement
5 Were the administrations independent?	Independent measurements	Assumable that the measurements were independent	Doubtful whether the measurements were independent	Measurements NOT independent
6 Was the time interval stated?	Time interval stated		Time interval NOT stated	
7 Were patients stable in the interim period on the construct to be measured?	Patients were stable (evidence provided)	Assumable that patients were stable	Unclear if patients were stable	Patients were NOT stable
8 Was the time interval appropriate?	Time interval appropriate		Doubtful whether time interval was appropriate	Time interval NOT appropriate
9 Were the test conditions similar for both measurements? e.g. type of administration, environment, instructions	Test conditions were similar (evidence provided)	Assumable that test conditions were similar	Unclear if test conditions were similar	Test conditions were NOT similar
10 Were there any important flaws in the design or methods of the study?	No other important methodological flaws in the design or execution of the study		Other minor methodological flaws in the design or execution of the study	Other important methodological flaws in the design or execution of the study

Appendix A. (continued)

Box B. Reliability: relative measures (including test-retest reliability, inter-rater reliability and intra-rater reliability)				
Statistical methods	**Excellent**	**Good**	**Fair**	**Poor**
11 for continuous scores: Was an intraclass correlation coefficient (ICC) calculated?	ICC calculated and model or formula of the ICC is described	ICC calculated but model or formula of the ICC not described or not optimal. Pearson or Spearman correlation coefficient calculated with evidence provided that no systematic change has occurred	Pearson or Spearman correlation coefficient calculated WITHOUT evidence provided that no systematic change has occurred or WITH evidence that systematic change has occurred	No ICC or Pearson or Spearman correlations calculated
12 for dichotomous/nominal/ordinal scores: Was kappa calculated?	Kappa calculated			Only percentage agreement calculated
13 for ordinal scores: Was a weighted kappa calculated?	Weighted Kappa calculated		Unweighted Kappa calculated	Only percentage agreement calculated
14 for ordinal scores: Was the weighting scheme described? e.g. linear, quadratic	Weighting scheme described	Weighting scheme NOT described		

CHAPTER 3

DESIGN AND CONTENT VALIDITY OF A NEW INSTRUMENT TO EVALUATE MOTOR ABILITIES OF CHILDREN WITH SEVERE MULTIPLE DISABILITIES: MOVAKIC

– PART I –

Physical Medicine and Rehabilitation – International 2015; 2(9)




ABSTRACT

Aim: Evaluation of motor abilities of children with severe multiple disabilities is often based on subjective assessment or on instruments validated for other target populations. A practical instrument for the evaluation of change in motor abilities is needed. In this study such an instrument is constructed and its content validity and applicability are tested.

Methods: The instruments content was developed using an expert focus group and a systematic literature review. Experts were consulted in all stages of development. Content validity was assesses using the COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN). Applicability was assessed by experienced physical therapists in a pilot study among 53 children and adults with severe multiple disabilities.

Results: Movakic (**mo**tor e**va**luation of **ki**ds with multiple and **c**omplex disabilities), a questionnaire consisting of 21 items on motor abilities, was constructed. Movakic scores are based on an assessment of motor performance of the child and have to be assessed by physical therapists. Movakic had adequate content validity and applicability.

Conclusions: Movakic's good applicability and content validity suggest that is has potential to be a useful instrument in clinical practice. Movakic's reliability is assessed in a prospective study, as reported in part II of this issue.

Key words

Severe multiple disabilities; Motor abilities; Cerebral Palsy GMFCS V; Content validity; Evaluative instrument

Abbreviations

SMD;	Severe Multiple Disabilities
COSMIN;	COnsensus-based Standards for the selection of health Measurement Instruments
Movakic;	MOtor eVAluation of KIds with multiple and Complex disabilities
GMFCS:	Gross Motor Function Classification System
CFCS;	Communication Function Classification System
MACS;	Manual Ability Classification System
IQ;	Intelligent Quotient
ICF-CY;	International Classification of Functioning, Disability and Health for Children and Youth
CLA;	The Chailey Levels of Ability
BSID III;	Bayley Scales of Infant and Toddler-Third Edition
PEDI;	Pediatric Evaluation of Disability Inventory
GMFM-88;	Gross Motor Function Measure (88 items)
MHFMS;	Modified Hammersmith Function Motor Scale
LE 85;	Lower Extremity physical functioning and mobility skills
MFM;	Motor Function Measure scale
TDMMT;	Top Down Motor Milestone Test
VAB;	Vulpe Assessment Battery
WeeFim;	Functional Independence Measure for Children

INTRODUCTION

Children with severe multiple disabilities (SMD) are characterized by a severe or profound intellectual disability and severe motor impairments. There is no universal description of this group to be found in the current literature. Apart from "severe multiple disabilities", which we selected, the terms "severe generalized cerebral palsy", "profound and intellectual multiple disability", "severe motor and intellectual disability", "severe neurological impairment and intellectual disability" are used. Support for these children in acquiring or improving motor abilities is highly relevant for participation in general care situations and a sense of self-determination or autonomy. Availability of practical and reliable instruments for the measurement of motor abilities in these children is very important. However, instruments specifically designed for the measurement of motor abilities in this target group are lacking.

Children with SMD mostly have a level of motor abilities that is comparable to level IV/V of the Gross Motor Function Classification System (GMFCS) for children with cerebral palsy (Palisano et al. 1997) (Veugelers et al., 2005): they typically are wheelchair-bound and only a few are able to move by crawling or using a physical aid (Mensch, Rameckers, Echteld, & Evenhuis., 2015) (Mergler et al., 2012) (Rieken et al., 2011). They are usually severely limited in maintaining their body position or in transfers to another position. Communicative functions are highly limited; the children are only able to communicate non-verbally or through body language, which translates to Communication Function Classification System (CFCS) level V (Hidecker et al., 2011). The child handles objects with difficulty or has severely limited ability to perform even simple actions. It requires support in almost all situations, which corresponds to Manual Ability Classification System (MACS) levels III-IV (Eliasson et al., 2006). In addition, children with SMD are often diagnosed with sensory impairments, dysphagia often leading to respiratory infections (Calis et al. 2008), gastro-oesophageal reflux disease, epilepsy, scoliosis and contractures (Veugelers et al., 2010) (Liptak et al., 2001) (Calis, Olieman, Rieken, & Penning., 2007). As a result, these children are fully dependent on their caregivers and material in their immediate vicinity for all activities of daily life.

In clinical practice, evaluation of motor abilities in children with SMD is often based on subjective assessments or on instruments developed for other target groups with motor disabilities. An instrument for this specific group, with the aim of longitudinal evaluation of progression, stabilization, or deterioration of motor abilities, which could then be applied in clinical physical therapeutic practice, was needed.

A systematic review of available instruments on motor abilities in children with severe disabilities (Mensch et al., 2015), showed that eight instruments might be potential candidates for application in children with SMD. One instrument (TDMMT) was developed specifically for this population (Putten van der, Vlaskamp, Reynders, &

Nakken., 2005). The seven other instruments were developed for children with cerebral palsy, other neurologic conditions, or motor disabilities in general. Although evaluation of psychometric properties of all eight instruments appeared incomplete, one or more of them might be suitable for children with SMD after some adaptation. Therefore, an expert focus group formulated suitability criteria and systematically judged the clinical suitability of the identified instruments for this group. The expert focus group determined the clinical suitability of the eight instruments based on five established criteria; 1) Low level of motor abilities, children with SMD are classified in level V (GMFCS) and can at best crawl. 2) Grading of scoring because of the subtle changes in motor abilities. 3) Manual and/or device support is a functional element in using motor abilities. 4) Non-verbal instruction, children with SMD have an Intelligent Quotient (IQ)<25 and do not understand verbal instruction. 5) Capability versus capacity and performance, which means the possibility, and not quality, of performing a motor ability is important. The suitability criteria were further specified in appendix A. As a result of the judgment, consensus was reached for all instruments; none of the selected instruments was found to be completely suitable in the target group. Therefore the focus group decided that development of a new instrument was needed. The procedure of the judgment is described in appendix A.

In this study we present the development of an instrument to evaluate motor abilities of children with severe multiple disabilities. The formulated suitability criteria were the starting point of the construction of the new instrument. We tested its applicability and content validity. In a companion publication in the current issue of this journal the reliability of the new instrument will be presented.

METHODS

Expert focus group

The joint development and application of new suitability criteria and growing insights into wishes for an ideal instrument for children with SMD led to a strong motivation of the expert focus group to proceed and design a new instrument. Members of the focus group were selected from therapists of the Ipse de Bruggen Care Organisation with ten years or more experience with the target population. The expert focus group consisted of six well-trained physical therapists and an occupational therapist, each with over 15 years [range 15-30 years] of specialist experience in working with children with SMD. They were trained in the use of different instruments such as the Gross Motor Function

Measure (GMFM), the Bayley Scales of Infant and Toddler-Third Edition (BSID-III) and the Pediatric Evaluation of Disability Inventory (PEDI). In addition, all have specific specializations in the field of intellectual disability and physical therapy interventions.

Theoretical framework

The content of the instrument was based on the next theoretical starting points: the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY) (World Health Organization., 2008), multidisciplinary treatment goals and monodisciplinary treatment goals. Using the ICF-CY we can distinguish the levels of 'body function and structure', 'activities', and 'participation'. Multidisciplinary treatment goals for children with disabilities are usually defined at the participation level of the ICF-CY, whereas mono-disciplinary treatment goals may concern the level of activities. In physical therapy, motor abilities (level of activity) are the primary focus for treatment, among other goals such as contracture management (level of body function and structure). This new instrument had the aim to evaluate motor abilities on the level of activities, and the users of the instrument had to be physical therapists working with children with SMD. Logically, the theoretical framework of the new instrument had to comply with the established criteria for clinical suitability.

Content validity

In the development of the new instrument, we used the general requirements (see table 1) on content validity of the COnsensus-based Standards for the selection of health Measurement INstruments (COSMIN). Assessing content validity is an important step in developing an instrument that aims to be relevant and comprehensive. An appropriate method might be to let experts judge the relevance and comprehensiveness of the items. The focus and detail of the items of the instrument need to be specifically designed to match the target population (Mokkink et al., 2012). Since the COSMIN criteria are designed to assess content validity of an instrument in a certain study, only the first four criteria needed to be met to deem the items suitable for obtained adequate content validity.

The approach in this study consisted of three steps, in which we aimed to satisfy the requirements for strong content validity: (1) listing of relevant motor abilities, (2) design of a structure and layout and (3) a first pilot study on applicability in children and adults with SMD.

Table 1. Cosmin Box D content validity (Mokkink et al., 2012)

	General requirements	Yes	No	?
1	Was there an assessment of whether all items refer to relevant aspects of the construct to be measured?	☐	☐	☐
2	Was there an assessment of whether all items are relevant for the study population? (e.g. age, gender, disease characteristics, country, setting)	☐	☐	☐
3	Was there an assessment of whether all items are relevant for the purpose of the measurement instrument? (discriminative, evaluative, and/or predictive)	☐	☐	☐
4	Was there an assessment of whether all items together comprehensively reflect the construct to be measured?	☐	☐	☐
5	Were there any important flaws in the design or methods of the study?	☐	☐	☐

Step 1: Listing of relevant motor abilities.

To identify motor abilities that are most relevant to children with SMD, we listed common treatment goals formulated in clinical practice using a survey. Twenty-five physical therapists, working with clients with SMD at a care provider service, were asked to collect all therapeutic long-term and short-term goals from their client files, formulated during the last five years.

Step 2: Structure and layout of the instrument.

As mentioned before, the five formulated suitability criteria as shown in appendix A, were the starting point for constructing the new instrument. Furthermore, the structure of the newly designed instrument was partially based on that of other instruments (e.g. start with the main positions of lying, sitting and standing) taking into account the relevance of specific situations and activities. These situations and activities were selected in concordance with collected therapeutic goals from the first step using the collective experience of the expert focus group. For the development of the structure and layout of the instrument, the expert focus group followed a repeated consensus procedure.

Step 3: Pilot study on applicability

Physical therapists working with children and adults with SMD from different care-organisations in the Netherlands were asked to participate in a pilot study. In total, twenty-six physical therapists evaluated the instrument's applicability and whether test items met the criteria for good content validity. None of these physical therapists were members of the expert focus group. They received a four-hour training on the web based computer application used to complete all items and on the user manual.

After applying the instrument to their own clients, the therapists answered questions on applicability, addressing the comprehensibility of the user manual, the layout (questionnaire and score-form), the items (relevance, comprehensibility, difficulty, number, and suggestions for other questions), the clarity of the answer categories and scoring procedure, and time needed to complete the instrument. There was room for written remarks per item, additions and further tips. The questionnaire also included a question on whether the instrument score measured corresponded to the therapist's clinical judgment, on a scale of 1 = not at all, to 10 = completely.

RESULTS

The proposed approach for the development of the new instrument outlined in the methods section of this paper was followed.

Step 1: Listing of relevant motor abilities.

The survey resulted in 355 therapeutic goals for 75 clients. After removal of duplicates and goals on the levels of body function (such as pain, contractures, dyspnoea) and participation, the remaining goals on the level of activity were translated into motor abilities by the expert focus group.

We illustrate this process with the example of the following therapeutic goal 'Client x is able to sit in his wheelchair during playing'. In this case, the generic motor ability on the level of activities is 'maintaining body position' and the specific motor ability is 'maintaining the sitting position'. This motor ability can be specified for different situations and activities of daily life. In our example, the specific situation is 'sitting in a wheelchair', whereas 'during playing' concerns the level of participation.

Step 2: Structure and layout of the instrument.

The discussions within the expert group during the repeated consensus procedure resulted in a structure and the layout of the instrument (see table 2); 1. Main body positions ('Positions'). 2. Specific situations in each body position ('Situations'). 3. Specific motor abilities in each situation ('Grouping of motor abilities'). 4. Items with questions ('Items').

Based on the relevance, the items consisting of motor abilities were clustered within the 12 situations (see Table 3).

Table 2. Structure of Movakic

Positions	Lying	Sitting	Standing
Situations ⟶ Grouping of motor abilities ↓	1 Supine 2 Supine with device* 3 Prone 4 Prone with device 5 Side 6 Side with device	7 Flat surface 8 Dangling legs 9 Chair/ sitting device 10 Feet on subsurface	11 Without device 12 With device
Maintaining position	Items (see table 3) with questions (table 4) on 1 Intensity manual or support by device 2 Activity of the child 3 Intensity of manual facilitation/ stimulation		
Activities			
Changing body position			
Moving around			

*Device: Assistive devices such as prostheses, ortheses and specialized tools and aids for personal mobility such as canes, walkers and wheelchairs. (World Health Organization., 2008)

Table 3. Items of Movakic

		Situations (see table 2)											
	Items	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**
1	Maintaining position	x		x		x		x	x	x	x	x	x
2	Duration maintaining position					x		x	x	x	x	x	x
3	Turning head	x	x	x	x	x	x	x	x	x	x	x	x
4	Upright head	x	x	x	x	x	x	x	x	x	x	x	x
5	Maintaining upright head position	x	x	x	x			x	x	x	x	x	x
6	Reaching with the arms	x	x	x	x	x	x	x	x	x	x	x	x
7	Take support (fore)arms			x	x								
8	Take support hands			x	x			x	x			x	
9	Grasping with the hands	x	x	x	x	x	x	x	x	x	x	x	x
10	Roll over to the left	x		x									
11	Roll over to the right	x		x									
12	Roll over to prone	x				x							
13	Roll over to supine			x		x							
14	Transfer from lying to sitting	x	x	x		x							
15	Transfer from sitting to lying							x	x				
16	Transfer from sitting to standing							x		x	x		x
17	Transfer from standing to sitting									x	x	x	x
18	Pivoting							x					
19	Minor voluntary postural changes								x	x	x		
20	Move on	x		x				x		x		x	x
21	Distance	x		x				x		x		x	x

In addition, the expert focus group decided that most of the items should include three questions; 1. Required level of manual support i.e. the level of palpable support that was given, or support by a device. 2. Level of activity of the child itself. 3. Level of manual facilitation, meaning active stimulation, i.e. provocation of the child with your hands. These questions address relevant elements in the actual use of motor abilities by these children during daily functioning. All questions could be scored using a four-point Likert scale (see Table 4).

Table 4. Questions and answer categories of the items

1 How much support does the child need?	0. Complete support 1. A lot of support 2. Moderate support 3. Barely support
2 Is the child active?	0. Completely passive 1. Has intention to stand up 2. Active during part of the movement 3. Completely active
3 If you use facilitation, how much do you use?	0. In spite of full facilitation there's no intention 1. A lot of facilitation 2. Variable facilitation 3. Only during start of movement

The expert focus group decided that the instrument should take the form of a questionnaire instead of an observational test. This was decided because execution of motor abilities of children with SMD may vary considerably under the influence of attention, fatigue, health, medication use, or unfamiliar circumstances. The questionnaire has to be completed by a therapist that has long-lasting experience with the child in a naturalistic setting instead of a therapist that does not know the child and uses the instrument in an isolated testing situation. Most other observational instruments are based on using a specific testing situation. Based on the difficulty to test execution of motor abilities in standardized situation, the judgment on being able to perform the motor ability in whatever shape or form in spontaneous situations during the last three months will be used to measure the motor abilities of the children with SMD.

A score-form was added to the questionnaire, containing separate pages for each specific situation and an overview page with total situation scores. The maximum total scores of the 12 situations will differ because of a different cluster of items. Because all children have different abilities or disabilities and different therapeutic goals, only situations that are relevant to the child need to be scored. Therefore, on the score-form, individual scores for evaluated situations have to be converted into percentages of the maximum scores. Apart from standardized scoring, there is room for written detail information too, because execution of motor abilities may vary in different specific situations (e.g. different types of wheelchairs). The users' manual instructs the user to write down examples of such detail information are added to the items.

The instrument was named Movakic (**mo**tor e**va**luation in **k**ids with **i**ntellectual and **c**omplex disabilities). Information for therapists was written in a first draft of a user manual, in which the guiding principles, structure and application of the instrument are explained. Descriptions of the most common terms are given, such as reaching, grasping and different forms of support. The scoring procedure is explained.

Step 3: First evaluation of applicability

Movakic was completed for 53 children, 35 boys and 28 girls, mean age 8.1 (range 2-17) years. The mean number of situations scored was 7 out of 11 (range 6-8). Mean completion time per situation was 9 minutes (range 6-15) and mean total completion time was 61 minutes (range 46-90 minutes). The content (validity) of the test items and relevance of the items was good according to all therapists. On a scale of 1 (very easy) to 10 (very difficult), mean difficulty score of the questionnaire was 2.8 (range 1-7). The mean therapists' judgment score of the correspondence of Movakic total score with the therapists' own clinical judgment was 8.5 (range 8-9). In addition, for assessing Movakic's applicability in adults, Movakic was completed by 10 physical therapists for 15 adults with SMD, seven women and eight men with a mean age of 40 years (range 19-65). Mean completion time for two situations was 18 minutes (range 10-30 minutes). The content of the test items and relevance of the items was good according to all therapists. On a scale of 1 (very easy) to 10 (very difficult), mean difficulty score of the questionnaire was 4.2 (range 2-7). The mean therapists' judgment score of the correspondence of Movakic total score with the therapists' clinical judgment was 8.1 (range 7-9).

All physical therapists were satisfied with the structure and contents of the instrument and offered practical suggestions for improvement, including the addition of an extended training. As a result of the practical suggestions for improvement the answer category is modified into a five-point likert scale, items about moving around were added and a group consisting of a cluster of items on motor abilities specific used in care-situations was added. In the appendix B the modified version of Movakic is adapted.

Content validity

From the general requirements on content validity (see table 1) all answers of item one till four were scored as yes, item five was scored as no. The relevance of the Movakic items to the construct of measuring motor abilities, to the study population and to the purpose of the instrument (item 1, 2, 3 and 4) were positively assessed using the expert focus group, by formulating the clinical suitability criteria and by using the results of the pilot study on applicability in which experts, others than the expert focus group, participated. Extraordinary care was taken to follow a comprehensive step-by-step procedure to insure good content (item 5). Movakic's content validity was thus scored as adequate.

DISCUSSION

Strengths of our approach to instrument development include highly experienced therapists, structuring the instrument and using a repeated consensus procedure, participated in the development of Movakic. By using this approach, it is highly likely that all relevant items of all relevant measurement properties are included, contributing to the content validity of the new instrument (Mokkink et al., 2012). However, since content validity is a subjective judgment, the developers cannot perform a completely unbiased judgment. Evaluation of content validity by a separate expert panel would contribute to the quality of the validity (Lynn., 1986) (Beck & Gable., 2001) (Mastaglia, Toye, & Kristjanson., 2003) of Movakic. Therefore other experts, who were not involved in the development of Movakic, participated in a pilot study and were asked to evaluate the relevance and comprehensiveness of the items of the new instrument.

The design of Movakic takes group-specific characteristics into account, which positively impacts the construction of the test. In addition the clinical criteria are formulated by clinical experts and based on the activity level (ICF-CY) (World Health Organization., 2008). Completion of a questionnaire by the therapist based on the child's performance during a longer time frame has the advantage of no extra burden of separately testing the child.

It might be argued that flaws in caregiver recall, on which much of the judgments in this study are based, may lead to inaccuracies in caregiver judgments. However, the contact between caregiver and child is very intensive, and anecdotal evidence suggests that in the care for this target group, caregivers are able to recall many (often minute) details about the care for and the condition of their clients. Experience of the physical therapist with this specific group of children and longer familiarity with the child play an essential role in effective application of Movakic. We do not expect that recall bias played an important role.

This study made an important step in the availability of an applicable, relevant and complete instrument. Movakic can be applied in a clinical setting because the construct is based on consensus of experts working in the field. In the Netherlands, most of these children visit day-care centers, where specialized on-site treatment is offered on a more or less daily basis. In such a situation, the requirements of the instrument can easily be met. We are not familiar with the involvement of physical therapists with children with SMD in other countries, but presume that most therapists treat their children often.

This study does not provide other psychometrics properties of Movakic. A prospective study that would prevent recall bias is recommended. Our study on Movakic's reliability, in collaboration with physical therapists in different care provider services, is reported in this issue in part II. Since we have demonstrated satisfactory psychometric properties, it will now be possible to use Movakic to evaluate effectiveness of current physical therapeutic interventions in children with SMD, and assess the effect of change in motor ability score on health and participation goals and quality of life.

CONCLUSION

The clinical suitability of the eight existing tests of motor abilities of disabled children for children with severe multiple disabilities (SMD) proved inadequate and as a result led us to design a new instrument. Using consensus criteria developed by an expert focus group of physical therapists a comprehensive instrument for the measurement of motor ability for children with SMD was developed: Movakic. Movakic is a questionnaire, containing of items on motor abilities, which can be objectified in a standardized procedure. Using a set of consensus based suitability requirements, Movakic was found to be applicable in clinical practice. In addition, Movakic has strong content validity.

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APPENDIX A

Procedure

The expert group formulated criteria for clinical suitability, based on their technical knowledge as well their practical experience with current instruments for the evaluation of motor abilities. In a first focus group meeting, each expert individually listed a series of requirements. This list of identified criteria was discussed and a clear definition of each criterion was drafted. In a second meeting, the criteria were ranked according to their perceived relevance and a final, limited set of criteria was agreed upon by consensus of the whole group.

Criteria for clinical suitability

The expert group formulated the following suitability requirements:

1) Low Level of motor abilities (GMFCS V). Motor functioning of children with SMD is very low, meaning these children are often unable to change body position, to take up and maintain an upright body position and moving around such as turning around, rolling and crawling. As a result, these children often stay in a supine position or sit in a fully supported wheelchair in daily living. Examples of motor abilities of a low level are for example raising the head, reaching with the arms, maintaining a sitting position or roll over. Motor abilities of a high level such as walking, running, jumping, catching, swimming and also tinkering, scissoring or washing are too complex for these children. Therefore an instrument for these children is suitable if the motor abilities are of a low level and not too difficult or complex. Motor abilities in lying or upright body positions will be an essential part of the test.

2) The grading or scoring should be sensitive to subtle changes in motor abilities. Due to the low level of motor abilities in children with SMD, in many instruments steps between changes in motor abilities are too large. Test items focused on changes in motor abilities are often related to the normal development such as the development from lying to standing in which intermediary steps are rolling over, sitting, crawling and running. Children with SMD often show subtle but relevant changes in motor abilities related to their care, eating or playing. Therefore an instrument is needed in which subtle changes have to be captured by small intermediary steps, which are relevant for these children. An example is the motor ability 'rolling over'. The desired result is coming from lying on the back to supine lying. In children with SMD a step in between can be rolling over into lying on the side or the child's cooperation during changing position in contrast to being passive.

3) Capability versus capacity and performance. An important part of instruments in general that assess motor ability is assessing the performance of motor abilities (what does the child do), which influences the score. In children with SMD the precise performance of a motor ability is of secondary importance. Of primary importance for these children is the capability in performing the desired motor ability no matter how that movement is executed (what can a child do). For example rolling over; a qualitative well-performed execution is to roll over by rotating the spine (performance). For motor functioning of these children the result and active participation is most important. Whether or not the child used rotation of the spine in the execution of rolling over is of lesser importance in these children (capability). Based on the difficulty to test the capacity (what the child can do in standardized situations) of motor abilities in a specific test situation, the capability of motor abilities of children with SMD have to be studied in a more naturalistic setting.

4) Non-verbal instruction; in children with SMD the level of understanding is very basic, which makes them barely instruction-oriented. In many instruments the execution of items is based on verbal assignments or instructions. Due to the low level of cognitive development, which is comparable to that of children with a maximum age of one year, responding to commands is not possible.

5) Manual support and/or support by devices. Children with SMD need support maintaining a position due to their severe cognitive and motor disabilities. Support can be given by devices such as a wheelchair and can be given by manual support. More specifically, a child can be supported by sitting upright in a wheelchair but can also be supported to be active in sitting upright or to maintain this position by stimulation by the hands of another person. Another example is that, on the one hand the child can be pushed from supine to prone position during diaper change and, alternatively, the child can be activated and supported by the performance of rolling over. Instruments for evaluating motor abilities are often based on observation of spontaneous or provoked motor functioning. Children are provoked to comply with specific conditions by verbal instructions or by providing triggering materials without touching the child. Due to the severe motor, cognitive and sensory disabilities, these children are often unable to understand spoken language, perform motor abilities against gravity and comply with instructions without caretakers present to stimulate attention of the child. In the performance of daily activities these children often need to be supported by their surroundings.

The experts decided that a suitable instrument should satisfy all of these criteria.

Evaluating the instruments

Instruments identified as potentially suitable through the systematic review (Mensch et al., 2015) were; 1) The Chailey Levels of Ability (CLA) (Palisano., 1993) (Poutney, Mulchahy, Clarke, & Green., 2004) 2) Gross Motor Function Measure (GMFM-88) (Russell et al., 1989) (Ketelaar, Petegem van-Beek van, & Visser., 1999), 3) Modified Hammersmith Functional Motor Scale (MHFMS) (Krosschell et al., 2011) (Main, Kairon, Mercuri, & Muntoni., 2003), 4) Lower extremity physical functioning and mobility skills (LE85) (Gorton et al., 2010), 5) Motor function measure scale (MFM) (Berard, Payan, Hodgkinson, & Fermanian., 2005), 6) Top Down Motor Milestone Test (TDMMT) (Putten van der et al., 2005), 7) Vulpe Assessment Battery (VAB) (Jain, Turner, & Worrell., 1994) and 8) Functional Independence Measure for Children (WeeFIM) (Niewczyk & Granger., 2010). One of these instruments has been specifically developed for children with SMD (TDMMT), and the others for children with cerebral palsy (CLA, GMFM-88, LE85), spinal muscular atrophy (MHFMS), neuromuscular diseases (MFM), and motor disabilities in general (VAB, WeeFIM).

The experts agreed that all of the criteria had to be met for an instrument to be suitable for children with SMD. They considered and discussed the possibility to adapt an existing instrument. The instrument's clinical suitability is shown in the table below.

Table: Scoring of the clinical suitability of the instruments

Instrument	Criteria of clinical suitability*					Total of positive scored criteria
	1)	2)	3)	4)	5)	
CLA	+	+	-	+	-	3
GMFM	+	-	-	-	+	2
MHFMS	-	-	-	-	+	1
LE85	-	+	-	-	+	2
MFM	-	-	-	-	-	0
TDMMT	-	-	-	+	+	2
VAB	-	+	+	-	-	2
WeeFIM	-	-	-	+	+	2

*Criteria of clinical suitability
1) Low Level of motor abilities. 2) The grading or scoring. 3) Capability versus capacity and performance
4) Non-verbal instruction. 5) Manual support and/or support by devices
+ = positive scoring. - = negative scoring
CLA = The Chailey Levels of Ability. GMFM-88 = Gross Motor Function Measure (88 items). MHFMS = Modified Hammersmith Function Motor Scale. LE 85 = Lower Extremity physical functioning and mobility skills. MFM = Motor Function Measure scale. TDMMT = Top Down Motor Milestone Test. VAB = Vulpe Assessment Battery. WeeFim = Functional Independence Measure for Children

Conclusion

None of the instruments was found to be completely suitable and therefore the focus group decided that development of a new instrument is needed. This instrument will need to adhere to all suitability requirements and should have adequate psychometric properties.

APPENDIX B MODIFIED VERSION OF MOVAKIC

Structure of the instrument

<table>
<tr><th>Positions</th><th>Lying</th><th>Sitting</th><th>Standing</th></tr>
<tr><td rowspan="2">Situations(13) ⟶
Grouping of motor abilities
↓</td><td>1 Supine
2 Supine with device
3 Prone
4 Prone with device
5 Side
6 Side with device</td><td>7 Flat surface
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<tr><td>Changing body position</td></tr>
<tr><td>Moving around</td></tr>
</table>

*Device: Assistive devices such as prostheses, ortheses and specialized tools and aids for personal mobility such as canes, walkers and wheelchairs. (World Health Organization., 2008)

Questions and answer categories

What is the intensity of manual support you give to the child?	O O O O O Full None
What is the intensity of activity of the child?	O O O O O Full Passive Full Active
What is your the intensity of facilitation to stimulate the motor ability?	O O O O O Full None

Items of Movakic

Situations (see table 'structure of the instrument')														
Items		**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**	**13**
1	Maintaining position	x		x		x		x	x	x	x	x	x	x
2	Duration maintaining position					x		x	x	x	x	x	x	x
3	Turning head	x	x	x	x	x	x	x	x	x	x	x	x	
4	Upright head	x	x	x	x	x	x	x	x	x	x	x	x	
5	Maintaining upright head position	x	x	x	x			x	x	x	x	x	x	
6	Reaching with the arms	x	x	x	x	x	x	x	x	x	x	x	x	
7	Take support (fore) arms			x	x									
8	Take support hands			x	x			x	x			x		
9	Grasping with the hands	x	x	x	x	x	x	x	x	x	x	x	x	
10	Roll over to the left	x		x										x
11	Roll over to the right	x		x										x
12	Roll over to prone	x				x								x
13	Roll over to supine			x		x								x
14	Transfer from lying to sitting	x	x	x		x								x
15	Transfer from sitting to lying							x	x					x
16	Transfer from sitting to standing							x		x	x		x	x
17	Transfer from standing to sitting									x	x	x	x	x
18	Pivoting							x						x
19	Minor voluntary postural changes								x	x	x			x
20	Move on	x		x				x		x		x	x	
21	Distance	x		x				x		x		x	x	
22	Move on and change direction									x		x	x	
23	Moving backwards									x		x	x	

CHAPTER 4

RELIABILITY OF MOVAKIC; AN INSTRUMENT TO EVALUATE MOTOR ABILITIES IN CHILDREN WITH SEVERE MULTIPLE DISABILITIES

- PART II -

Physical Medicine and Rehabilitation – International 2015; 2(9)




ABSTRACT

Aim: '**Mo**tor e**va**luation in **ki**ds with intellectual and **c**omplex disabilities' (Movakic) is a newly developed Dutch instrument for evaluating motor abilities in children with severe multiple disabilities. We have previously shown that its feasibility and content validity are satisfactory. The aim of this study is to investigate test-retest and inter-rater reliability.

Methods: Children with severe multiple disabilities were scored six times by their own physical therapists at a three month interval, and at baseline by a second therapist familiar with the child in a subset of children. For the purposes of this study, the three-month period in which no event involving the child took place was selected.

Results: Sixty children were recruited. The mean age of the children was 7.7 years (range 2-16), 45% had a cognitive development level <6 months (N=27) and 52% had Gross Motor Function Classification System level V (N=31). Test-retest reliability could be evaluated in 50 children and inter-rater reliability in 19 children. Intraclass correlations were all excellent or good (range .72-.98). Adequate absolute reliability is reflected in a small mean distance of Movakic scores and most respondents' distances for test and retest were between one standard deviation and zero. Distribution is not related to the score level, although a ceiling effect might be present in score range 90-100.

Conclusions: Movakic is a reliable instrument for measuring motor abilities in children with severe multiple disabilities.

Key words

Severe multiple disabilities; Motor abilities; Cerebral Palsy GMFCS V; Evaluative instrument; Movakic; Reliability

Abbreviations

Movakic;	MOtor eVAluation of KIds with multiple and Complex disabilities
SMD;	Severe Multiple Disabilities
GMFCS;	Gross Motor Function Classification System
COSMIN;	COnsensus-based Standards for the selection of health Measurement INstruments
IQ;	Intelligent Quotient
ICF-CY;	International Classification of Functioning, Disability and Health for Children and Youth
ICC;	intraclass correlation coefficients

INTRODUCTION

Motor abilities are of paramount importance to independent functioning, but are often severely compromised in children with severe multiple disabilities (SMD). Children with SMD suffer from profound intellectual disabilities (IQ <25) and have a level of motor abilities that is comparable to level IV/V on the Gross Motor Function Classification System (GMFCS) for children with cerebral palsy (Palisano et al., 1997) (Veugelers et al., 2005). In addition, children with SMD may have multiple sensory disorders and other comorbidity. Usually, physiotherapists are closely involved with stimulation and training of such children, because even subtle improvement of motor abilities can aid these children in developing some degree of control over their environment and may as such improve their quality of life. As in any healthcare profession, physical therapists desire to evaluate the effectiveness of their treatment methods, for which reliable instruments are needed.

However, commonly used instruments for measuring motor abilities in children with disabilities (Mensch, Rameckers, Echteld, & Evenhuis., 2015a) review are considered unsuitable for children with SMD. They are unsuitable because of their inclusion of higher GMFCS levels, the requirement of perfect execution of the motor ability, the need for verbal instruction, the use of large-step grading, and the design of items without the application of manual support or use of devices in mind. Therefore, an instrument was needed that fulfills specific suitability criteria (Mensch, Rameckers, Echteld, Penning, & Evenhuis., 2015a).

A new instrument named Movakic (**mo**tor e**va**luation in **ki**ds with **i**ntellectual and **c**omplex disabilities) for measuring and evaluating motor abilities in children with SMD was developed by a Dutch expert focus group and was found to be feasible with good content validity (Mensch et al., 2015b). Before an instrument can be used in clinical or research settings, stability across time and raters should be assessed (Vet de, Terwee, Knol, & Bouter, 2006). Therefore, in this study the test-retest reliability and inter-rater reliability of Movakic were evaluated.

METHODS

Participants, selection

Children younger than 18 years with SMD who received care in specialized day-care centers were included. Severe multiple disabilities were defined as profound intellectual disability (IQ<25) in combination with severely impaired motor abilities GMFCS level IV and V (Palisano et al., 1997).

Each of 37 experienced therapists working in the centers, who all had more than 10 years experience with the target population, selected one or two of their own clients. Informed consent by parents or legal representatives was obtained from all participants.

Movakic

Movakic is a questionnaire consisting of items on motor abilities; the complete questionnaire is shown in the appendix of the design article (Mensch et al., 2015). All terminology used is based on the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY) terminology (World Health Organization., 2008). Motor abilities are distributed over 13 'situations' (see Table 1), each representing a client's body position with or without the use of a device. Within each situation, a cluster of items addresses four groups of motor abilities: maintaining position, activities, changing body position, and moving around. Questions are asked about the extent in which manual support or support from a device was needed, the own activity of the child, and the extent of aiding the child manually i.e. 'provocation' of the child with your hands.

Table 1. Structure of Movakic

<table>
<tr><th>Positions</th><th>Lying</th><th>Sitting</th><th>Standing</th></tr>
<tr><td rowspan="2">Situations (13) ⟶
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</table>

Movakic has to be completed by professional therapists who are familiar with the clients' motor performance on the basis of longer treatment experience with the child, and not on performance of activities in a specific test situation, which is the basis of most other observational instruments. Scoring of motor abilities in this group should not depend on functioning in a single test situation only, because performance may worsen under the influence of lack of attention, fatigue, bad health, medication use, or unfamiliar circumstances (Mensch et al., 2015b). Because all children have different abilities or disabilities and different therapeutic goals, only the questions regarding situations that are relevant to the child need to be scored: A situation could be relevant if a baseline measurement is desired for future follow-up, if therapeutic changes are expected in a certain situation, or for evaluating change in motor ability.

All items are scored on a five-point Likert-scale. Scores on the left-hand side always represent lower and scores on the right-hand side represent higher scores in motor function (range 0-4 points). An example of an item is given in Table 2.

Table 2: example of an item including the sub-questions

1-Maintaining sitting					
What is the extent of manual support you gave the child to maintain this position?	O Full	O	O	O	O None
What is the child's level of activity?	O Full Passive	O	O	O	O Full Active
What is your the extent of facilitation to stimulate the motor ability?	O Full	O	O	O	O None

The maximum scores of the 13 situations will differ because of the variable cluster of items. Therefore, per situation, absolute scores are converted into percentages (situation score divided by maximum situation score x 100). For the remainder of this paper, this percentage will be referred to as the 'Movakic score'.

Movakic is provided on-line through a secure Internet site. The completion of the instrument on the screen starts by the items and their questions for the chosen situation. Per question, only one answer (one button) can be selected. Modifications are allowed until the "save" button is pressed. During the study period, participating physiotherapists could not check the results of the Movakic score after completion of the questionnaire. It was only after the study ended and all data was collected that they received a report with the scores for each child.

Procedure

The reliability study is part of a longitudinal study for which participating therapists were instructed in the use of the instrument and the study procedure during a one-day training session on the application and the user manual. Data collection was performed from August 2010 to October 2011. Therapists were requested to complete Movakic six times (T0-T5) in a period of 18 months. Intervals of three months were chosen based on standardized evaluation periods in clinical practice and changes in this period are not expected in the target population if no specific events occur. In order to evaluate inter-rater reliability, a second therapist who was also familiar with the client's motor performance, if available, completed Movakic at baseline.

For this reliability study, therapists were requested to choose a situation per child, containing a cluster of the items of Movakic. Therapists were asked to note events that might have influenced the child's scores during each interval. Because of their frequent contacts with the child, its parents, and its carers and other therapists in the day-care setting, physiotherapists were well aware of medical conditions, medication changes, changes in the care teams or at home, and other events. For the current reliability study, scores between two measurement periods in which no events took place were chosen, so called T0-T1 (see figure 1).

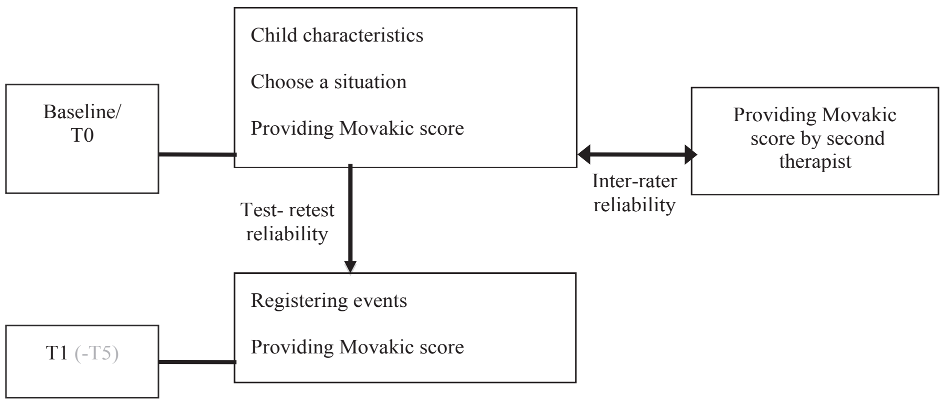

Figure 1. Flow chart of study procedure. Scores between two measurement periods in which no events took place were so called T0-T1

We recorded the child's sex, age, estimated cognitive developmental age, GMFCS level, diagnosis, comorbid conditions, and devices, which we asked the child's therapist to provide.

Analyses

For the assessment of test-retest reliability, baseline (T0) Movakic scores were compared with T1-scores at three months. In case an event was recorded that might have caused a change between T0 and T1, another 3-month period was selected in which no such events had occurred. For the assessment of inter-rater reliability, baseline measurements of two therapists were used.

To test relative reliability, which is the degree in which children maintain their position in a sample over repeated measurements, two-way mixed model intraclass correlation coefficients (ICC) were calculated with 95%-confidence intervals between the Movakic scores of T0 en T1 (for test-retest reliability) and between the Movakic scores of the two raters (for inter-rater reliability). ICC's for test-retest and inter-rater reliability were also calculated for each of the four groups of motor abilities. Reliability was classified as excellent (> 0.75), good (0.60–0.74), fair (0.40-0.59), or poor (<0.40) (Cicchetti & Sparrow., 1981).

The ICC does not provide information about the degree in which actual scores for an individual vary over repeated measurements (absolute reliability). The smaller the differences, the higher the absolute reliability. This was done by performing Bland & Altman analyses (Bland & Altman., 1986). First, we calculated for each child the distance (absolute difference) between T0 and T1 Movakic scores (for test-retest reliability) and between both T0 Movakic scores per rater (for inter-rater reliability). Next, for test-retest reliability mean Movakic scores at T0 and T1 were plotted against the individual differences between T0 and T1, for inter-rater reliability mean Movakic scores were plotted against individual differences between both raters at T0. Adequate reliability is represented by small differences from the mean (within one standard deviation (SD) of the mean).

All calculations were performed in IBM Statistical Package for the Social Sciences version 21.

RESULTS

Population

Sixty children with a mean age of 7.7 years (range 2-16 years) were selected in 15 different day-care centers. Characteristics of the study population are listed in table 3.

Table 3. Characteristics of the study population

		N = 60	%
Gender	Male Female	31 29	52 48
Age in years	1-6 6-12 12-18	20 28 12	33 47 20
Cognitive development level in months	0-6 6-12 12-18 >18 Not scored	27 12 5 2 14	45 20 8 3 23
GMFCS*[1] level	IV V Not scored	22 31 7	37 52 11
Diagnosis	Cerebral Palsy *Spastic CP* *Ataxic CP* *Dyskinetic CP* Syndromes/ gene mutations Acquired brain injury Metabolic disease Unknown	25 *21* *1* *3* 18 1 2 14	42 *84* *4* *12* 30 1.7 3 23
Comorbidity	Epilepsy PEG*[2] tube Scoliosis Visual impairment Respiratory problem Other*[3] Secondary problem/ contractures	39 22 24 44 17 17 29	65 37 40 73 28 28 48
Devices*[4]	Wheelchair Standing device Walking aid (Semi) Orthopaedic shoes Orthotics Lying device	57 42 24 24 32 16	95 70 40 40 53 27

*[1] GMFCS: Gross Motor Function Classification System (Palisano et al., 1997)
*[2] PEG tube: percutaneous endoscopic gastrostomy tube
*[3] Other: additionally noted comorbid conditions such as heart disease, hearing disorders, diabetes mellitus, gastro-oesophageal reflux disease
*[4] Device: Assistive devices and aids for personal mobility (World Health Organization, 2008)

Test-retest reliability

During the interval of three months, six children were lost to follow-up: one died, two were severely ill, two had been transferred to other day-care centers and one moved home. In addition, four children were lost to follow-up for test-retest analyses due to noted events that might have influenced reliability during all of the five 3-month intervals, such as surgery, sickness or increasing contractures. Test-retest reliability for the remaining 50 children in terms of agreement (ICC) and mean distance of Movakic scores is presented in Table 4.

Table 4. Test-retest reliability (N=50)

Baseline – 3 months	**ICC (95%CI)**	**Mean distance of Movakic scores (SD)**
Total score	.95** (.92-.97)	6.8 (6.4)
Group of motor ability		
Maintaining position	.98** (.97-.99)	
Activities	.96** (.94-.98)	
Changing body position	.91** (.82-.95)	
Moving around	.72** (.27-.90)	

ICC = intraclass correlation coefficient
95%CI = 95% Confidence Interval
SD = Standard Deviation
*p <0.05, **p<0.01

Figure 2 shows the Bland & Altman plot of all individual differences between Movakic scores at T0 and T1 scores against mean Movakic scores. The score differences varied between -25 and 24 points (SD = 9.2). Seventy-four percent of the differences were within one standard deviation (dotted lines).

Inter-rater reliability

Movakic was completed at baseline by two therapists for 19 children. Inter-rater reliability in terms of ICC and mean distance of Movakic total scores is presented in Table 5.

Figure 3 shows the Bland & Altman plot of all individual differences between Movakic scores of the two raters against mean Movakic scores. The score distances varied between -21 and 29 points (SD=11.8). Seventy-nine percent of the distances were within the standard deviation (dotted lines in the figure).

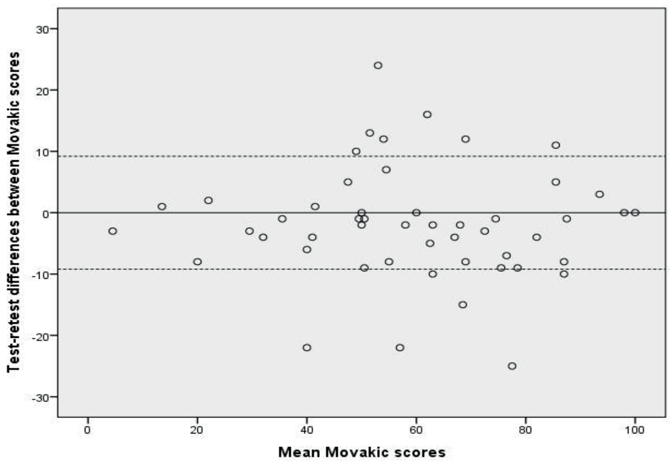

Figure 2. Individual differences of Movakic score between T0 and T1 (y-axis), plotted against mean Movakic score (x-axis) (N=50). The dotted lines represent one standard deviation of the differences in Movakic scores.

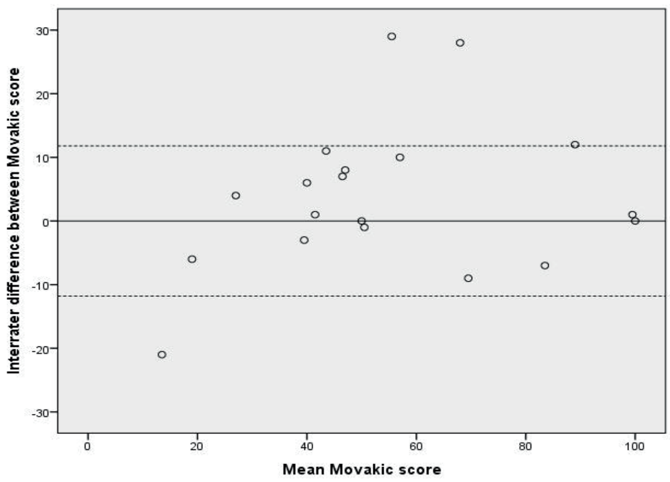

Figure 3. Individual differences in Movakic scores between two raters (y-axis), plotted against mean Movakic scores (x-axis) (N=19). The dotted lines represent one standard deviation of the differences.

Table 5. Inter-rater reliability (N=19)

Baseline	ICC (95%CI)	Mean distance of Movakic scores (SD)
Total score	.94** (.85-.98)	8.6 (8.7)
Group of motor ability		
Maintaining position	.89** (.35-.97)	
Activities	.96** (.89-.98)	
Changing body position	.85** (.57-.95)	
Moving around	.97** (.90-.99)	

ICC = intraclass correlation coefficient
95%CI = 95% Confidence Interval
SD = Standard Deviation
*p<0.05; **p<0.01

DISCUSSION

All ICC values for test-retest reliability were excellent, except for the subscale 'Moving around', of which the ICC was good. Here, the 95% confidence interval of ICC's was very wide, with the lowest limit at .27. The 95% confidence interval for 'Changing body position' was also relatively wide compared to the other ICCs. ICC values for inter-rater reliability of the subscales 'Maintaining position' and 'Changing body position' were excellent, but the 95% confidence interval of the ICC's was wide, with the lowest values at .57 and at .35, respectively. Adequate absolute reliability is reflected in a small difference of mean Movakic scores, for test-retest reliability 6.8 (SD 6.4) and for inter-rater reliability 8.6 (SD 8.7). Adequate reliability is also reflected in 74% and 79% of the test-retest and inter-rater distances falling within one SD from zero. However, a few respondents exhibited large distance scores, indicating suboptimal reliability for a small number of respondents making some caution in interpreting the results

In fact, we expected lower ICC's because, in accordance with the procedure, the therapists selected a relevant situation for each individual child. Their choice depended on various aspects such as diagnosis, comorbidity, therapeutic goals, used devices and abilities and disabilities. This procedure has ecological validity, i.e. represents the real-life situation, but also creates diversity, which may be reflected in the suboptimal ICC's with wide confidence intervals. It was hard to find a more standardized procedure for this heterogeneous and specific target group. The heterogeneity of the study population is apparent from Table 1. The children were recruited from wide spread care organizations in the country so we can assume the study population is representative and adequately reflects the diversity of the target group. It is encouraging that reliability levels were adequate despite this heterogeneity.

A lower concordance of inter-rater than of test-retest scores was to be expected, because of the relative small number of participants, but also because of the subjectivity introduced by therapists having to estimate the extent in which they used manual support. Nevertheless, the high ICC's (Tables 2 and 3) support the strong design of Movakic, including the application of manual support.

This study has several strengths. It is one of the few studies in this specific target population with relatively high participation rates e.g. (Veugelers et al., 2005) (Calis et al., 2008) (Veugelers et al., 2010) (Liptak et al., 2001) (Brug ten, Putten van der , Penne, Maes, & Vlaskamp., 2012) (Putten van der, Vlaskamp, Reynders, & Nakken., 2005) (Putten van der & Vlaskamp., 2011) (Calis et al., 2010) (Rieken et al., 2011). Moreover, all participating therapists received adequate training in the use of the instrument related to the purpose of the study. Additionally, all had extensive experience with the target population and were familiar with the included children. The therapists thus fulfilled the prerequisites for participating in the study as raters. Electronic data collection ensured that the data is of good quality and scoring errors were limited. A memory effect was highly minimalized by not showing scores during completion of the questionnaire and the wide time frame of three months. Moreover, therapists noted events that may have influenced the children's scores during the interval and such intervals were not used for analyses.

The time frame of three months may be considered a long period for showing stability of an instrument. However, based on the experience in clinical care of this group of children, it was expected that stability in motor abilities would be high.

On the other hand, the high ICC's may be explained by the extreme familiarity of the therapists with the children, the fact that they all had a long-time experience with the target group, but also by the fact that they were highly motivated because of being personally involved in the development of a new instrument. This situation may be less favorable in future daily practice, where physiotherapists with less experience with these children may have to complete Movakic, who will not always be connected to specialized day-care centers, whose time is not or to a limited extent reimbursed by health insurances, or who work in countries with different healthcare systems. Therefore, training is needed to enhance a correct application of the instrument; even in less favorable circumstances (Mensch et al., 2015b).

CONCLUSION

In this study, test-retest and inter-rater reliability were examined of a newly developed instrument, Movakic, to evaluate motor abilities of the specific subgroup of children with SMD (Mensch et al., 2015b). Test-retest reliability was evaluated in 50 children and inter-rater reliability in 19 children with SMD. For both test-retest and inter-rater reliability, intraclass correlations of Movakic scores and of sub-scores for four groups of motor abilities were all excellent or good. Adequate absolute reliability was reflected in a small mean distance of Movakic scores and in the accuracy of individual scores, representing a normal variation.

In addition, from this study we can conclude Movakic is a reliable instrument for measuring motor abilities in children with SMD. But before the implementation of Movakic for clinical evaluation of motor abilities in children with SMD, its responsiveness and construct validity have to be evaluated first.

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Declaration of interests

We certify that no party that contributed to the results of this research has or will benefit from its publication nor will any organization with which we are associated. If applicable, we certify that all financial and material support for this research and work are clearly identified in the title page of the manuscript. Conflicts of interest: none to declare.

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CHAPTER 5

CONSTRUCT VALIDITY AND RESPONSIVENESS OF MOVAKIC; AN INSTRUMENT FOR THE EVALUATION OF MOTOR ABILITIES IN CHILDREN WITH SEVERE MULTIPLE DISABILITIES

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ABSTRACT

Movakic is a newly developed instrument for measurement of motor abilities in children with severe multiple disabilities, with a satisfactory feasibility and content validity and good inter-observer and test-retest reliability.

The objective of this study was to investigate its construct validity and responsiveness to change.

Sixty children with severe multiple disabilities (mean age 7.7 years, range 2-16) were measured using Movakic six times during 18 months. Construct validity was assessed by correlating Movakic scores with expert judgment. In order to assess responsiveness, scores during 3-months intervals were compared (mean score-changes and intraclass correlations) during which some children experienced meaningful events influencing motor abilities and during which others experienced no such event.

Forty-five percent of children had a lower cognitive development level than 6-month, 52% had Gross Motor Function Classification System level V and 37% had level IV. For 27 children all measurements were completed, six children dropped out. Construct validity was good (r= .50 - .71). Responsiveness was demonstrated by significantly larger score changes after events than when such events did not occur.

Movakic is a valid instrument for measuring motor abilities in children with severe multiple disabilities. Results suggest responsiveness to change in motor abilities after meaningful events.

What this paper adds: The study on psychometric properties of this newly developed instrument supports the use of Movakic in clinical practice in children with severe multiple disabilities. Physiotherapists will be able to detect changes in motor abilities in these children with serious comorbid conditions often leading to a big amount of meaningful events such as surgery, pain or medication change.

Key words: Severe multiple disabilities; Motor abilities; Cerebral Palsy GMFCS IV-V; Evaluative instrument; Movakic; Validity

Highlights

- Movakic is a valid instrument for measuring motor abilities in children with SMD.
- Responsiveness seems significant to change in motor abilities after events that likely affect motor abilities.
- Physiotherapists are able to use Movakic in clinical practice for the Dutch population.
- The English version of Movakic is in progress.

INTRODUCTION

Children with severe multiple disabilities (SMD) suffer from profound intellectual disability, limited motor abilities and serious comorbid conditions (Mensch et al., 2015c; Mensch, Rameckers, Echteld, & Evenhuis., 2015b).

Physiotherapists are closely involved with stimulation and training of children with SMD, because even subtle improvement of motor abilities can aid these children in developing some degree of control over their environment (Houwen, van der Putten, & Vlaskamp., 2014). In addition, increased physical activity can have positive influence on health issues e.g. constipation and fitness and may, in turn improve their quality of life.

Palisano et al studied the amount and focus of physical therapy and occupational therapy in children with CP. Children in level I were receiving fewer minutes per month of physical therapy than children in levels IV–V. (Palisano et al., 2012). However, the effect of higher frequency of physical therapy is not assessed in this study. In order to evaluate the effect of frequency of therapy it is important to measure effectiveness of interventions. In order to evaluate effectiveness, they need validated instruments to measure change in relevant parameters of intervention outcome such as motor abilities. No instruments exist that specifically measure motor abilities in children with severe involvement (Mensch, Rameckers, Echteld, & Evenhuis., 2015a). MOtor eVAluation in Kids with Intellectual and Complex disabilities, Movakic, was developed by a group of experts (Mensch et al., 2015c), to be used by physiotherapists in planning and evaluation of intervention for children with severe motor involvement. A feasibility study demonstrated that the instrument is considered clinically relevant and suitable for the target population (Mensch et al., 2015c). Its test-retest and inter-rater reliability were satisfactory (Mensch, Rameckers, Echteld, & Evenhuis., 2015b).

Before Movakic can be implemented as an evaluation tool in clinical practice, its validity needs to be studied. Therefore, in the current study, Movakic's construct validity and its responsiveness to change were evaluated (Mokkink et al., 2010). Responsiveness is an important characteristic of instruments designed to evaluate the effectiveness of intervention.

METHODS

Participant selection

Children included in this study had SMD, defined as a profound intellectual disability (IQ<25) and motor abilities comparable with a Gross Motor Function Classification System (GMFCS) level IV or V (Palisano et al., 1997; Palisano, Rosenbaum, Bartlett & Livingston., 2008). Children younger than 18 years with SMD who received care in specialized day-care centres were included. Each of 37 experienced therapists working in the centres, all of whom had more than 10 years experience with the target population, selected one or two of their own clients. Informed consent by parents or legal representatives was obtained from all participants.

Study design

This was a prospective cohort study with a follow-up of 18 months. Measurements were performed at baseline and up to 5 times during follow-up at 3-month intervals (labelled T1 to T5), representing the standard evaluation period in the day-care centres.

Validity testing

Construct validity of an instrument is the degree to which the scores of such an instrument are consistent with hypotheses regarding the association between the target instrument and other instruments measuring the same construct (Terwee et al., 2012; Mokkink et al., 2010). Because no instruments exist to measure motor abilities in children with SMD, construct validity of the Movakic was tested against expert judgment of children's motor ability.

Responsiveness is the ability of an instrument to detect change over time of the construct of interest (Terwee et al., 2012; Mokkink et al., 2010). In an ideal situation, we would investigate responsiveness against the effect of a single intervention without confounding variables such as other events or interventions. However, because of the complex study population receiving different interventions, both medical and physiotherapeutic, responsiveness was tested making use of the fact that some children would experience meaningful events influencing motor abilities while others would not. For this study, the underlying supposition was that we do not expect changes in motor abilities in a time frame of three months, unless events occurred. The possible occurring events were defined based on the experience of the participating therapists. The next events were selected; surgery, increase of contractures, pain, medication change, change in assistive devices, and other, unexpected events. The proportion of positive versus negative changes in scores was inventoried.

Instrument

Motor abilities were measured by the Movakic instrument (Mensch et al., 2015c). Movakic is a digital questionnaire system (currently only available in Dutch), to be administered on-line through a secure internet portal. Information about the instrument, the instruments' structure, the items and an example of an item is provided in appendix A. Motor abilities are distributed over 12 situations.

Each situation represents a body position with or without the use of an assistive device, consisting of a cluster of items addressing four groups of motor abilities as defined in the International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY) (World Health Organization., 2008): maintaining position, activities, changing body position, and moving around. Questions are asked about the extent to which manual support or support by an assistive device was needed, the activity initiated by the child, and the intensity of active stimulation, i.e. 'provocation' of the child with your hands.

Because all children have different abilities and different therapeutic goals, only those situations of Movakic that are relevant to the child need to be scored. Therefore, therapists are requested to choose situations relevant to the evaluation of interventions or monitoring of motor function. A situation could be relevant if a baseline measurement is desired for future follow-up, if therapeutic changes are expected in a certain situation, if the therapist is interested in general level of change in motor ability, or a combination of these reasons. Items on the Movakic instrument (Appendix A) are scored on a five-point Likert-type scale, ranging from 0 points, representing the lowest motor function to 4 points, representing the highest motor function. Only one answer per question can be selected by clicking a button. The maximum total scores of the 12 situations differ because of the variable number of items. Therefore, situation scores and total Movakic scores are converted into percentage scores (range 0-100).

Procedure

Physiotherapists underwent training in use of the Movakic instrument and the study procedure. Participant selection and data collection were performed from August 2010 through October 2011. At baseline, therapists provided information on the child's sex, age, cognitive developmental age, GMFCS level, diagnosis, comorbid conditions, and assistive devices used. Therapists also provided expert judgment of the child's motor abilities and Movakic score at baseline. At each of five, 3-month intervals (T1 to T5), therapists again provided expert judgment, repeated the Movakic, and reported on any events theorized to influence motor ability during the previous 3 months.

Therapists were requested to choose one relevant Movakic situation, e.g., body position, for each child that was expected to remain stable during follow-up, taking into account unexpected events that could change the chosen situation. At each measurement, the therapists were requested first to provide their judgment of the child's motor abilities in the chosen Movakic situation, using the Visual Analogue Scale (VAS) (Reips & Funke., 2008). The clinical question to be answered was: 'Thinking of motor abilities relevant for children with SMD in the chosen Movakic situation, how well is the child able to perform these motor abilities at this moment?' Physiotherapists specified their judgement by indicating a position along a continuous line between two end-points, the left most side representing the lowest overall motor ability. This resulted in a score between 0-10 centimeters. Therapists then scored the items for the chosen situation. After Movakic completion, participants could not see or review the results of Movakic scores.

After each 3-month interval, therapists were asked to note the occurrence of random events potentially positively or negatively influencing motor abilities. Events that could be registered were surgery, increase of contractures, pain, medication change, change in assistive devices, and other, unspecified events. Because of the frequent contacts of therapists with the children, their parents, and caring staff in the centres, therapists were well aware of such information.

In summary, the following data were collected: at baseline child characteristics, expert judgment and Movakic score, and at T1 to T5 expert judgment and Movakic scores, as well as any events theorized to influence motor ability during the previous 3 months.

Analyses

To test construct validity, Pearson correlation between Movakic total scores and expert judgment scores was calculated. Validity was considered good when r>.40 (Cohen., 1992). Responsiveness was tested by comparing Movakic score changes during 3-month intervals with identified events and score changes during intervals without such events. For this purpose, intervals of all children were entered into a new database, with only two measurements, pre-test (T0) and post-test (T1). The new database had an event group and no-event group and could consist of the same children at different time points. An example is given in Table 1.

With the new dataset we were able to investigate whether Movakic scores changed in the appropriate interval in the presence or absence of events. We realized that complete data for all six intervals might not be available for all children. It was expected that participants with high numbers of measurements would be overrepresented in comparison with participants who were lost to follow up earlier. As a result, the number of collected measurements of participants might mediate the change in Movakic scores, because children in whom less data were collected on events might be representative of a subgroup of children with characteristics that could also influence change in motor abilities. To determine whether this effect occurred, classification in the event/no event group was correlated with the number of measurements, using point-biserial Pearson's correlation (p<.05).

Table 1. Part of the data set after restructuring for responsiveness testing. Nx are data points

Participant	T0	T1	Event
1	N1	N2	Yes
1	N2	N3	Yes
1	N3	N4	No
1	N4	N5	No
1	N5	N6	No
2	N7	N8	No
2	N8	N9	No
2	N9	N10	Yes
3	N11	N12	Yes
3	N12	N13	Yes
4	N14	N15	Yes
4	N15	N16	No
4	N16	N17	Yes
4	N17	N18	Yes
4	N18	N19	No

Responsiveness was analysed using two approaches: comparison of mean Movakic score-changes in intervals with and without events, and comparison of intraclass correlations of T0 and T1 scores in intervals with and without events. In the first approach, responsiveness would be demonstrated if the mean absolute score-change in the event group was larger than in the no event group. The difference between the means was analysed using Student's t-test, and the magnitude of the difference was expressed as Cohen's *d* effect size (small >.2, moderate >.5, large >.8) (Cohen., 1992). In the second approach, agreement between T0 and T1 scores was tested using two-way mixed model ICC's. Responsiveness would be demonstrated if the ICC's in the event group were lower than in the no event group, as shown by non-overlapping 95%-confidence intervals (CI). The proportion in the event group of the direction of change was calculated.

Statistical analyses were performed using IBM SPSS Statistics v. 22.

RESULTS

Participants

Sixty children with SMD from 15 different care settings were included in the study (Table 2). Mean age of the children was 7.7 years (2-16), 45% had a cognitive development level of less than 6 months of age and 51% of the children were classified as GMFCS level V. Only baseline data were obtained for six children who were not included in the longitudinal analyses; one child died, two children were severely ill, two children had been transferred to another day-care centre and one moved home.

Events

Physiotherapists noted a large number of meaningful events, however many events occurred without a clear influence on motor abilities e.g., change in type of medication. Because of this lack of information, we only used the event categories 'surgery', 'increase of contracture' and 'pain' for the analyses. Using these categories, a total of 68 events were recorded during the course of the study for 49 out of 54 children.

Construct validity

Scores were obtained for 60 children at baseline, however only 27 children remained in the study after 15 months. Children were lost to follow up mainly due to severe illness, transfer to other day-care centres, or circumstances of the participating therapists. Correlations of Movakic and expert judgment scores are presented in Table 3.

Responsiveness

Classification in the event or no event group was not correlated with the number of measurements per participant (r=.06; p=.27). Mean absolute Movakic score-changes were significantly larger in the event group (Table 4) (t=2.21; p=.03), with a moderate effect size (Cohen's d=.62, 95%-CI [.23, 1.00]). Intraclass correlation coefficients (ICC) of T0 and T1 scores in the event group (ICC = .78, 95%CI .64-.86) was lower than in the no-event group (ICC = .95, 95%CI .94-.96), as presented in Table 4. The confidence intervals of these ICCs do not overlap, indicating that the agreement between the two measurements was stronger in the no-event group than in the event group.

Table 2. Characteristics of the study population

		N = 60	%
Gender	Male Female	31 29	52 48
Age in years	1-6 6-12 12-18	20 28 12	33 47 20
Cognitive development level in months	0-6 6-12 12-18 >18 Not scored	27 12 5 2 14	45 20 8 3 23
GMFCS*1 level	IV V Not scored	22 31 7	37 52 11
Diagnosis	Cerebral Palsy *Spastic CP* *Ataxic CP* *Dyskinetic CP* Syndromes/ gene mutations Acquired brain injury Metabolic disease Unknown	25 *21* *1* *3* 18 1 2 14	42 *84* *4* *12* 30 1.7 3 23
Comorbidity	Epilepsy PEG*2 tube Scoliosis Visual impairment Respiratory problem Secondary problem (contractures) Other*3	39 22 24 44 17 29 17	65 37 40 73 28 48 28
Assistive devices*4	Wheelchair Standing device Walking aid (Semi) Orthopaedic shoes Orthotics Lying device	57 42 24 24 32 16	95 70 40 40 53 27

*1 GMFCS: Gross Motor Function Classification System (Palisano et al., 1997). To indicate the level of motor abilities for the total group of children with SMD we used the GMFCS
*2 PEG tube: percutaneous endoscopic gastrostomy tube
*3 Other: additionally noted comorbid conditions such as heart disease, hearing disorders, diabetes mellitus, gastro-oesophageal reflux disease
*4 Assistive devices and aids for personal mobility (World Health Organization, 2008)

Table 3. Pearson correlation of Movakic score and expert judgment score at baseline, 3, 6, 9, 12 and 15 months

	N	r
Baseline (T0)	60	.55**
3 months (T1)	54	.71**
6 months (T2)	45	.59**
9 months (T3)	41	.50*
12 months (T4)	33	.66**
15 months (T5)	27	.67**

*$p<0.05$; **$p<0.01$
r = Pearson correlation coefficient

Table 4. Mean score changes and ICC's in event and no event groups

Mean Movakic scores			
Event/No event	**N**	**Absolute mean score change (*SD*)**	**ICC (95%CI)[1]**
Event	68	12.38 (14.97)	.78 (.64 - .86)
No event	235	6.84 (6.92)	.95 (.94 - .96)

[1]ICC for agreement of T0 and T1 Movakic scores, with 95%-confidence intervals
ICC = Intra Class Correlation
SD = Standard deviation

As shown in table 5 the proportion of change in score between 3-months intervals increased in 42.7% and decreased in 48.5% of the event group.

Table 5. Direction of Movakic score changes in event group

Direction of change*	**Event group N=68**	
	%	**N**
Increase score	42.7	29
Decrease score	48.5	33
Stable score	8.8	6

* Direction of Movakic score change between 3-months intervals in event group

DISCUSSION

This 18-month prospective cohort study was one of the initial prospective studies in children with SMD with adequate participant sample size. It was designed to obtain first data on validity of the Movakic instrument, a questionnaire-based instrument to monitor motor abilities in this specific group. The results show that Movakic has good construct validity and is significantly responsive to change resulting from events that impact motor ability such as surgery.

The sample in this study reflects the complexity of the population in terms of the variety of comorbidities and events such as surgery, medication changes, or diseases that potentially influence motor abilities. Because of this complexity we could not evaluate Movakic's responsivity to specific intervention effects. Therefore, we had to rely on whether or not Movakic scores of motor abilities were sensitive to events that were theorized to influence motor ability. In order to minimize confounding, therapists were instructed to score motor abilities they believed would be stable over time. Based on clinical experience we have tried to estimate the influence of meaningful events. The many events impacting these children over time will always complicate evaluation of intervention effects, both in clinical practice and in research. Therefore, intervention studies should examine the effects of botulinum toxin treatment, specific physical therapy interventions, orthopedic surgery or assistive devices. In clinical practice, careful information on unexpected events should be explicitly considered as part of the evaluation of motor ability. In addition, cohort studies of children with serious comorbid conditions may have a higher chance of loss to follow up. To this extent, we decided to add a standardized form to the Movakic instrument to note events and their potential implications for motor ability or lost to follow up.

As almost half of the children with SMD in this study were diagnosed with CP we considered the Gross Motor Function Measure (GMFM) (Russell et al., 1989) to be used for these children. The GMFM is specifically developed for children with Cerebral Palsy and has also been used to create gross motor function growth curves for children with Down syndrome (Palisano et al., 2001). The study on clinical suitability of currently used instruments in practice (Mensch et al., 2015c) showed us that the GMFM scored low level of motor abilities and support by assistive devices was allowed. However, we note the following limitations in the suitability of using the GMFM in clinical practice for children with SMD: 1- the grading of scoring may not be sensitive to subtle changes in motor abilities; 2- the difficulty of testing motor abilities in unnatural settings, and 3- the difficulty of understanding and following directions when administering verbal instructions. These are some of the same concerns that lead to the development of the Movakic.

Although the use of ICC's for responsiveness study is unusual, given the unusual data set and unusual instrument, we allowed ourselves to find solutions that lay outside the box. In this case we hypothesised that the concordance between Movakic scores before and after an 'event' would be less than the concordance between Movakic scores without an event in between. Concordance between interval data such as Movakic scores is adequately tested with ICCs. Comparing the magnitude of ICCs with confidence intervals is also appropriate. The low sample size is also an issue, because it may lead to wide confidence intervals, which means that the results are conservative. We do not believe that the difference in sample size makes it difficult to compare the ICC values, because they do not stem from a single analysis or sample, only the overlap of confidence intervals was tested.

To study the construct validity of Movakic, the instrument was tested against expert judgement using the VAS. In accordance with COSMIN guidelines, both Movakic and VAS were scored by the same rater, which may partly account for high correlations. However, completing Movakic requires much more detailed information than the single-item VAS, which will reduce the effect of subjectivity. Respondents are no longer free to decide on the content of their ratings. Furthermore, we reduced the influence of scoring both instruments by the same rater by blinding the rater for the results of Movakic.

Another point of discussion is whether overrepresentation of respondents in the event or no-event group could affect the results. We checked whether this was the case by correlating classification into either the event or no even group with the number of measurements. This correlation was low and not statistically significant, indicating a low risk of bias.

We developed Movakic to measure changes in motor abilities in children with SMD for which no other test is sensitive. Our first step was to test responsiveness to change in motor abilities by comparison of mean Movakic score-changes in intervals with and without events, and comparison of intraclass correlations of T0 and T1 scores in intervals with and without events. In this study we did not collect detailed information on the content of the events and the direction of the effect they were expected to have on motor abilities. As a next step, we recommend an intervention study with the aim to assess the relationship between carefully inventoried events, interventions and change in motor abilities measured with Movakic. Further studies about responsiveness should include the interpretation of the changes and the meaning for children and parents.

CONCLUSION

Evaluating change in children with SMD is a challenging task due to the complexity of comorbid conditions and variety of interventions aimed to improve function or increase comfort. Given Movakic's feasibility and content validity (Mensch et al., 2015c), reliability (Mensch, Rameckers, Echteld & Evenhuis., 2015b), and the results of the current study, we recommend the instrument for use in clinical practice and research. Physiotherapists should have ample experience with children with SMD, and should be familiar with the individual child they measure using Movakic. In the process of implementing Movakic in clinical practice, therapists need to be trained in correct use of Movakic. A two-day training course was developed in the Netherlands, and translation of the instrument in English is in progress. Furthermore, in future studies using specific interventions, we recommend focusing on clarifying the interpretation of the changes in Movakic score.

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Declaration of interests

We certify that no party that contributed to the results of this research has or will benefit from its publication nor will any organization with which we are associated. We agree this paper meets the standards of expected ethical behaviour. If applicable, we certify that all financial and material support for this research and work are clearly identified in the title page of the manuscript. Conflicts of interest: none to declare.

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APPENDIX A. Additional information about Movakic (Table A.1, A.2, A.3)

Table A.1. Structure of Movakic

Positions	Lying	Sitting	Standing
Situations ⟶ Grouping of motor abilities ↓	1 Supine 2 Supine with device* 3 Prone 4 Prone with device 5 Side 6 Side with device	7 Flat surface 8 Dangling legs 9 Chair/ sitting device 10 Feet on subsurface	11 Without device 12 With device
Maintaining position	Items (see table A.2) with questions (table A.3) on 1 Intensity manual or support by device 2 Activity of the child 3 Intensity of manual facilitation/ stimulation		
Activities			
Changing body position			
Moving around			

*Device: Assistive devices such as prostheses, ortheses and specialized tools and aids for personal mobility such as canes, walkers and wheelchairs. (World Health Organization., 2008)

Table A.3. Example of questions and answer categories of an item

What is the extent of manual support you gave the child to maintain this position?	O O O O O Full None
What is the child's level of activity?	O O O O O Full Passive Full Active
What is the extent of facilitation to stimulate the motor ability?	O O O O O Full None

Table A.2. Items of Movakic

	Situations (see table A.1)												
	Items	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**
1	Maintaining position	x		x		x		x	x	x	x	x	x
2	Duration maintaining position					x		x	x	x	x	x	x
3	Turning head	x	x	x	x	x	x	x	x	x	x	x	x
4	Upright head	x	x	x	x	x	x	x	x	x	x	x	x
5	Maintaining upright head position	x	x	x	x			x	x	x	x	x	x
6	Reaching with the arms	x	x	x	x	x	x	x	x	x	x	x	x
7	Take support (fore)arms			x	x								
8	Take support hands			x	x			x	x			x	
9	Grasping with the hands	x	x	x	x	x	x	x	x	x	x	x	x
10	Roll over to the left	x		x									
11	Roll over to the right	x		x									
12	Roll over to prone	x				x							
13	Roll over to supine			x		x							
14	Transfer from lying to sitting	x	x	x		x							
15	Transfer from sitting to lying							x	x				
16	Transfer from sitting to standing							x		x	x		x
17	Transfer from standing to sitting									x	x	x	x
18	Pivoting							x					
19	Minor voluntary postural changes								x	x	x		
20	Move on	x		x				x		x		x	x
21	Distance	x		x				x		x		x	x
22	Move on and change direction									x		x	x
23	Moving backwards									x		x	x

x= situation containing the item

CHAPTER 6

THE RELATIONSHIP BETWEEN MOTOR ABILITIES AND QUALITY OF LIFE IN CHILDREN WITH SEVERE MULTIPLE DISABILITIES

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ABSTRACT

Background: The aim of this study was to determine the relationship between motor abilities and quality of life in children with severe multiple disabilities.

Methods: In this cross-sectional study, motor abilities of 29 children with severe multiple disabilities (IQ<25, girls 45%, GMFCS V 45%, mean age 9.8 (SD 4.6)) were measured using the questionnaire **mo**tor e**va**luation in **k**ids with **i**ntellectual and **c**omplex disabilities (Movakic). Quality of life was measured with the 'Quality of Life - Profound Multiple Disabilities' (QoL-PMD) questionnaire. The child's physical therapist completed Movakic and the parents of the child completed the QoL-PMD.

Results: A significantly moderate to high correlation (r = .40, p = .03) was found between the total scores on the Movakic and the QoL-PMD, indicating that higher scores in motor abilities are associated with a higher level of quality of life. Furthermore, significantly moderate to high correlations were found between the total score on Movakic and the dimension Physical Well-Being, Development, and Activities of the Qol-PMD. In multiple linear regression models, all significant bivariate relationships between Movakic total scores and QoL-PMD dimensions remained statistically significant after controlling for GMFCS-level.

Conclusions: Motor abilities, measured by Movakic, are moderately related to the quality of life, measured by the QoL-PMD, in children with severe multiple disabilities.

Key-words: Severe multiple disabilities or profound intellectual and multiple disability, Motor abilities, Quality of life, Cerebral Palsy GMFCS IV-V, Movakic.

BACKGROUND

In general, motor abilities play a very important role in independent functioning (Janssen et al. 2010). Adequate motor function in people with motor disabilities contributes to a sense of independence and autonomy, and improves the level of participation in daily life (Raz-Silberger et al. 2015; Badia et al, 2014; Schoenmakers et al. 2005). In children with severe multiple disabilities (SMD), motor abilities are severely compromised, comparable with Gross Motor Function Motor Classification System (GMFCS) level IV/V (Palisano et al. 1997). They are often wheelchair-bound and dependent on their social, instrumental and material environment for all activities of daily life. In addition, children with SMD have a profound or unspecified intellectual disability (IQ<25) (American Psychiatric Association (APA). 2013). Communicative functions are severely limited; children with SMD only communicate non-verbally or through body language. Furthermore, Mensch et al., (Mensch et al. 2015c) reported that these children have sensory disorders such as visual impairment and other co-morbidity such as feeding and respiratory problems, epilepsy and contractures.

Several studies found a relationship between motor functioning and QoL. In a study of Dickinson et al. on self-reported quality of life of 8–12-year-old children with cerebral palsy (N=500, GMFCS IV-V 14%, IQ<70 27%) severely limited self-mobility was found to be significantly associated with reduced mean score for physical well-being (Dickinson et al. 2007). In a systematic review of Tsoi et al. some positive effect was identified of certain medicinal and motor control interventions on quality of life in children with cerebral palsy (N=347, average age 9.1) (Tsoi et al. 2012). Badia et al. found in their study on pain, motor function and health related quality of life in 91 children (age 8-19Y, GMFCS level IV-V 51,6%, severe cognitive impairment 30,8%) that motor impairment scores were significantly positively associated with physical and autonomy domains of quality of life (Badia et al. 2014). Maher et al. found a positive association between physical activity, social and physical quality of life, and happiness in young people with cerebral palsy (N=70, mean age 13y, GMFCS level IV-V 16%) (Maher et al. 2016). We found one study in people with SMD (N=49, average age 23.7 SD12.2) where Petry et al. (2009) studied the association between QoL, motor function, personal characteristics and characteristics of care settings. They found characteristics of the medical conditions of the children turned out to be most strongly associated with the QOL-PMD scores. In their study other personal characteristics such as age, gender, motor limitations and sensory limitations were not found significantly related to the QOL-PMD scores.

In addition, there is some limited evidence that impaired motor abilities will impact quality of life (QoL). We hypothesize this might be especially true in children with SMD, whose motor abilities are severely limited. By improving their motor abilities these children might feel more sense of control over their environment, which might in turn influence their QoL. However, the knowledge of a relationship between motor function and QoL in this target population is largely lacking.

This study explores our hypothesis that a higher motor ability level is related to a higher level of QoL in children with SMD. Specifically, we studied the association between different QoL dimensions and motor abilities in different positions (lying, sitting, standing). In addition, we studied independent associations, controlling for possible confounders i.e. gender, age and GMFCS levels.

METHODS

Study design and study population

This was a cross-sectional study. Children younger than 18 years with SMD who received care in specialised day-care centres were recruited from nine different care organizations for people with intellectual disabilities in the Netherlands. Children with SMD, as included for this study, had a profound intellectual disability (IQ<25), motor abilities comparable with a GMFCS levels IV or V (Palisano et al. 1997) and sensory or other comorbid disorders.

Thirty-four physical therapists working in the day-care centres were asked to select one or two of their clients with SMD and 27 agreed to participate. All participating physical therapists had over 10 years of experience working with children with SMD and had to be the therapist of their child with SMD for at least 3 months. They selected 56 children in total who fulfilled the inclusion criteria. The parents or legal representatives of 29 children provided informed consent for participation. Table 1 shows the characteristics of the study sample. Mean age of the children was 9.8 years (range 2-18) and 45% of the children were classified as GMFCS level V.

Table 1. Demographic and diagnostic data of the study population

		N = 29	%
Gender	Male Female	13 16	45 55
Age in years	1-6 6-12 12-18	3 10 16	10 35 55
GMFCS*[1] level	IV V Not scored	12 13 4	41 45 14
Diagnosis	Cerebral Palsy Syndromes/gene mutations Meningitis Metabolic disease Unknown	8 12 1 1 7	28 42 3 3 24
Cognitive developmental level in months	0-6 6-12 12-18 Unknown Not scored	6 4 2 6 11	21 14 7 21 38
Comorbidity	Epilepsy PEG*[2] tube Scoliosis Visual impairment Respiratory problem Secondary problem (contractures) Other*[3] Not scored	12 7 7 14 4 8 7 11	41 24 24 48 13 27 24 37
Assistive devices*[4]	Wheelchair Standing device Walking aid (Semi) Orthopaedic shoes Orthotics Lying device Not scored	16 11 7 9 10 3 11	55 38 24 31 35 10 38

*[1] GMFCS: Gross Motor Function Classification System (Palisano et al. 1997) Palisano et al., 1997]. To indicate the level of motor abilities for the total group of children with SMD we used the GMFCS
*[2] PEG tube: percutaneous endoscopic gastrostomy tube
*[3] Other: additionally noted comorbid conditions such as heart disease, hearing disorders, diabetes mellitus, gastro-oesophageal reflux disease
*[4] Assistive devices and aids for personal mobility (World Health Organization. 2008)

Instruments

Motor abilities

Motor abilities were measured by the **mo**tor e**va**luation in **k**ids with **i**ntellectual and **c**omplex disabilities (Movakic) instrument, a digital instrument that was specifically developed for the evaluation of motor abilities in children with SMD (Mensch et al. 2015c). This instrument was developed because instruments applicable to children with severe disabilities were lacking (Mensch et al. 2015b) and the existing tests have been proven inadequate (Mensch et al. 2015c). The structure of Movakic is shown in table A1 of the appendix. Motor abilities are distributed over 12 situations, each representing a client's body position: lying, sitting and standing, with or without the use of a device. Each situation consists of a cluster of items addressing four groups of motor abilities: maintaining position, activities, changing body position, and moving around. Questions were asked about the extent to which manual support or support from a device is needed, the own activity of the child, and the extent of stimulating the child manually, i.e. 'facilitation' by enabling a child to actively participate in a certain motor ability.

Because all children have different abilities or disabilities and different therapeutic goals, only situations that are relevant to the child need to be scored, based on the therapist's experience of the child's motor abilities. All items are scored on a five-point Likert-scale. The maximum total scores of the 12 situations differ because of the variable number of items. Therefore, situation scores and total Movakic scores are converted into percentage scores (range 0-100). A higher score is associated with better motor abilities within each chosen situation. Additional information on Movakic's items and sub-questions is given in appendix A.

Movakic was judged clinically relevant and suitable for the target population and has adequate content validity (Mensch et al. 2015c). Its test-retest and inter-rater reliability are excellent or good (ICC .72-.98) (Mensch et al. 2015a), its construct validity is good (r= .50 - .71) and Movakic is responsive to change resulting from events that might impact motor ability (Mensch et al. 2016).

Quality of Life

For measuring the QoL in children with SMD, we used the 'Quality of Life - Profound Multiple Disabilities' (QoL-PMD) (Petry et al. 2009a; Petry et al. 2009b; Petry et al. 2008). Because of the communication difficulties of children with SMD proxy measures of QoL in multiple dimensions were assessed. The QoL-PMD is a multidimensional questionnaire consisting of 55 items. These items consist of statements related to the life of the child with SMD and are divided into six subscales: physical well-being, material well-being, communication & influence, socio-emotional well-being, development, and activities. The items are scored on a 4-point scale (agree, partly agree, disagree and undecided). A score of 0-100% can be obtained, both on the total score and on the subscale score. A higher score indicates a better QoL. Psychometric examination of the QoL-PMD (Petry et al. 2009b; Petry et al. 2008) showed a good internal consistency for the total questionnaire (α = 0.90-0.92) as well as for the subscales (α = 0.63-0.88). In addition, correlations between the QoL-PMD and the MIPQ (Mood interest and Pleasure Questionnaire) ($r = 0.31$, $p < 0.001$) and between the QoL-PMD and a general measure of the QoL (using a 10-point scale) ($r = 0.44$, $p < 0.001$) demonstrated a moderate construct validity of the QoL-PMD in children with SMD.

Procedure

Informed consent by parents or legal representatives was obtained for all participants. Physiotherapists underwent training in the proper use of the Movakic instrument and the study procedure. The researcher and the expert group who developed the instrument trained the therapists. At baseline, therapists provided information on the child's gender, age, cognitive developmental age, GMFCS level, diagnosis, comorbid conditions, and assistive devices used. The motor abilities were measured with Movakic, completed by the child's physiotherapist; QoL was measured with the QoL-PMD, completed by the parents of the child. Both measures had to be completed within a period of a maximum of three months. During this time period, no changes were expected. To control for possible changes, all physiotherapists were asked to report on factors such as medication change, surgery, pain or increase of contractures that might influence motor abilities in the interim. If an event occurred in a child, this child would be excluded from further analysis.

Analyses

Characteristics of the study sample were described. Movakic scores were obtained in two different ways: 1) Per child the average of all completed situations of Movakic was calculated in a total Movakic score. 2) The scores of the chosen situations of Movakic were averaged for the three body positions 'lying' (situation 1-6), 'sitting' (situation 7-10) and 'standing' (situation 11-12) and for each child the average score per body position was calculated.

Normality of the Movakic and QoL-PMD data was checked using the Kolmogorov-Smirnov and was found sufficient. Using Pearson's correlation coefficient (r), Movakic sub-scores in the different body positions (lying, sitting and standing) were correlated with both QoL sub-scores on dimensions and total QoL score. The same analysis was used to correlate the Movakic total score with both QoL sub-scores and total QoL score. The magnitude of correlation coefficients was interpreted using guidelines by Cohen (1992) (Cohen. 1992) (.10-.29=small, .30-.49=medium, >.50=large). A positive correlation means that high scores on motor abilities often co-occur with high scores on quality of life; this is also the case for low scores. Independent relationships between motor abilities and QoL were tested using multiple linear regression analysis models with the QoL dimensions (dependent variables) that correlated significantly with Movakic sub-scores on body positions (independent variables). In model 1 gender, age and GMFCS-scores were entered into the equation after which in model 2 the Movakic motor abilities total and subscale scores were entered into the equation. In order to control for the covariates gender, age and GMFCS-score, explained variance (R square) was calculated for both models. The uniquely explained variance in QoL scores using only the Movakic scores can now be calculated by subtracting the R square of the total model (model 2) from the model that only uses the covariates (model 1). Multicollinearity was tested using the variance inflation factor (VIF); multicollinearity was considered to be present if VIF was higher than 10. P-values of .05 or smaller were considered to indicate statistical significance.

All analyses were done using SPSS/PASW Statistics version 21 (SPSS Inc, Chicago, Illinois).

RESULTS

Movakic and QoL-PMD were measured within a period of 2-12 weeks (mean 7.2 weeks); no meaningful events were recorded within this measurement interval for any child.

Bivariate correlations between Movakic and QoL subscales and total scores are shown in Table 2. A significantly moderate high correlation (r=0.40, p=0.03) between total Movakic score and total QoL-PMD score was shown. Significant bivariate moderate correlations were also found between the total Movakic score and the QoL-PMD dimensions Physical Well-Being, Development and Activities. Motor abilities in the lying and sitting body position were significantly positively correlated with the QoL-PMD dimension Development (moderately to substantially). Motor abilities in the sitting body position were also significantly positively correlated (moderately) to the dimension Physical Well-being and total QoL-PMD score. Motor abilities in the standing body position had a strong significant positive correlation to the dimension Activities and total QoL-PMD score.

Mean QoL-PMD and Movakic scores as well the results of the multiple regression analysis are reported in Table 3. As significant bivariate correlations were found between total Movakic score and the QoL-PMD dimensions Physical Well-Being, Development and Activities and between Movakic body position scores and these same QoL-PMD dimensions, these QoL dimensions were used for the multiple regression analyses. The scores within the Physical Well-being, Development and Activities dimensions were relatively high. The mean total score was 65.3% (SD 14.7). After controlling for gender, age and GMFCS-levels in multiple linear regression models, all significant bivariate relationships between Movakic total scores and QoL-PMD dimensions and the QoL-PMD total score remained statistically significant. The relationship between the dimension Activities and the body position standing and between total QoL-PMD and the body positions sitting and standing lost their significant correlation in this analysis. The addition of the Movakic variables on body positions into the regression models resulted in significant additions to the proportion of explained variance. Most models explained fair amounts of variance. The relationships between gender, age, GMFCS-levels and QoL-PMD variables were not statistically significant in any of the regression models, except for GMFCS in the regression model with the body position lying and QoL dimension physical well-being (β=.66; R^2=.23; p=<.05).

Table 4 shows the results of the last step of the multiple regression analysis, that show independent relationships between total Movakic and the different domains of Quality of Life scores (total, physical well-being, development and activities). After controlling for the covariates, the MOVAKIC total score explained a significant 17% more of the variance in the total QoL score (ß = 0.48; p = 0.05) than the model using only the covariates. In addition, the model accounted for 30% more of the variance in the QoL domain 'physical well-being' (ß = 0.64; p = 0.01), 20% in the QoL domain 'development' (ß = 0.52; p = 0.03) and 19% in the QoL domain 'activity' (ß = 0.51; p = 0.03). The total model accounted for 25% of the variance.

Table 2. Bivariate correlations between Movakic and Quality of Life scores

Movakic	Quality of life dimensions						
	Physical well-being	**Material well-being**	**Communication & Influence**	**Socio-emotional well-being**	**Development**	**Activities**	**Total QoL-PMD**
Body Positions	r^1 (*p*)	r^1 (*p*)	r^1 (*p*)	r^1 (*p*)	r^1 (*p*)	r^1 (*p*)	r^1 (*p*)
Lying (N=20)	.43 (.06)	.18 (.46)	-.17 (.49)	.30 (.20)	.51* (.02)	.34 (.15)	.38 (.10)
Sitting (N=25)	.49* (.01)	.16 (.46)	.06 (.78)	.29 (.16)	.45* (.03)	.36 (.08)	.41* (.04)
Standing (N=17)	.30 (.25)	-.05 (.85)	.46 (.06)	.43 (.09)	.23 (.40)	.61** (.01)	.53* (.03)
Total Movakic (N=29)	.43* (.02)	.09 (.65)	.08 (.67)	.32 (.09)	.46* (.01)	.38* (.05)	.40* (.03)

[1] Pearson (bivariate) correlation coefficient; *$p \leq .05$; **$p \leq .01$

Table 3. Mean Movakic scores and independent relationships (standardised regression coefficients) between Movakic and Quality of Life scores

Movakic		Quality of life dimensions							
		Physical well-being		**Development**		**Activities**		**QoL total**	
		(Mean=55.3 ; SD=20.5)		**(Mean=64.8 ; SD=21.2)**		**(Mean=70.4 ; SD=17.0)**		**(Mean=65.3 ; SD=14.7)**	
Body Positions	**Mean (SD)**	β^1	$R^{2\,2}$	β^1	$R^{2\,2}$	β^1	$R^{2\,2}$	β^1	$R^{2\,2}$
Lying	.60 (.22)	.66*	.34*	.60*	.29*	.44	.15	.55*	.24*
Sitting	.56 (.22)	.75**	.38*	.57*	.22*	.48	.15	.49	.16
Standing	.60 (.19)	.19	.03	.01	.00	.46	.16	.34	.09
Movakic total	.58 (.20)	.64**	.30**	.52*	.20*	.51*	.19*	.48*	.17*

[1] Standardised regression coefficient controlling for gender, age and GMFCS levels; [2] Proportion explained variance of the regression model (R^2); the significance level indicates a significant change in the F statistic after entering the Movakic dimension; *$p \leq .05$; **$p \leq .01$

Table 4. Regression analyses showing independent relationships between Movakic and Quality of Life (total and relevant domain scores)

	R^2	ΔR^2	ΔF	*p* ΔF	*Indep var*	B	*SE*	β	*t*	*p*
QoL-PMD total										
Movakic total	.25	.17	4.56	.05	Gender Age GMFCS Movakic total	-3.32 .47 2.34 31.83	5.44 .59 6.34 14.9	-.12 .16 .08 .48	-.61 .79 .37 2.14	.55 .44 .72 .05
Movakic lying	.27	.24	4.85	.04	Gender Age GMFCS Movakic lying	.88 -.01 9.60 30.30	5.61 .55 6.13 13.76	.04 -.01 .40 .55	.16 -.03 1.57 2.20	.88 .98 .14 .04
Movakic sitting	.28	.16	3.61	.08	Gender Age GMFCS Movakic sitting	-4.62 .44 2.38 31.18	6.42 .76 7.58 16.38	-.16 .13 .08 .50	-.72 .58 .31 1.90	.48 .57 .76 .08
Movakic standing	.43	.09	1.67	.22	Gender Age GMFCS Movakic standing	-4.8 1.23 -6.52 19.25	7.62 .91 5.74 14.92	-.23 .48 -.31 .34	-.63 1.35 -1.14 1.29	.54 .20 .28 .22
QoL-PMD Physical well-being										
Movakic total	.34	.30	9.16	.01	Gender Age GMFCS Movakic total	-3.53 .32 10.95 61.15	7.38 .81 8.60 20.21	-.09 .08 .27 .64	-.48 .40 1.27 3.03	.64 .69 .22 .01
Movakic lying	.39	.34	8.36	.01	Gender Age GMFCS Movakic lying	2.25 -.31 19.32 56.16	7.93 .78 8.66 19.43	.06 -.09 .52 .66	.28 -.40 2.23 2.89	.78 .69 .04 .01
Movakic sitting	.45	.38	10.88	.01	Gender Age GMFCS Movakic sitting	-3.90 .40 14.26 68.34	8.12 .96 9.58 20.72	-.09 .08 .34 .75	-.48 .42 1.49 3.30	.64 .68 .16 .01
Movakic standing	.17	.03	.35	.57	Gender Age GMFCS Movakic standing	-12.01 2.03 -4.79 19.65	16.94 2.02 12.75 33.15	-.31 .44 -.12 .19	-.71 1.01 -.38 .59	.49 .34 .71 .57
QoL-PMD Development										
Movakic total	.31	.20	5.81	.03	Gender Age GMFCS Movakic total	10.43 .37 2.42 53.26	8.07 .88 9.41 22.10	.24 .08 .06 .52	1.30 .42 .26 2.41	.21 .68 .80 .03
Movakic lying	.40	.29	7.10	.02	Gender Age GMFCS Movakic lying	14.01 -.09 11.57 61.63	9.44 .93 10.31 23.13	.30 -.02 .26 .60	1.48 -.10 1.12 2.67	.16 .92 .28 .02

Table 4. (continued)

	R^2	ΔR^2	ΔF	*p* ΔF	*Indep var*	B	*SE*	β	*t*	*p*
QoL-PMD Development										
Movakic sitting	.32	.22	5.10	.04	Gender Age GMFCS Movakic sitting	13.03 .26 5.02 56.44	9.80 1.15 11.56 25.00	.28 .05 .11 .57	1.33 .22 .43 2.26	.20 .83 .67 .04
Movakic standing	.36	.00	.00	.97	Gender Age GMFCS Movakic standing	14.49 .50 -14.11 .99	12.72 1.52 9.57 24.89	.43 .13 -.42 .01	1.14 .33 -1.47 .04	.28 .75 .17 .97
QoL-PMD Activity										
Movakic total	.26	.19	5.25	.03	Gender Age GMFCS Movakic total	1.34 .50 3.71 36.86	5.88 .64 6.85 16.10	.04 .16 .12 .51	.23 .78 .54 2.29	.82 .45 .59 .03
Movakic lying	.18	.15	2.74	.12	Gender Age GMFCS Movakic lying	2.17 .21 7.53 29.97	7.39 .72 8.07 18.10	.07 .07 .25 .44	.29 .29 .93 1.66	.77 .77 .37 .12
Movakic sitting	.24	.15	3.20	.09	Gender Age GMFCS Movakic sitting	-.15 .34 .92 31.03	6.80 .80 8.02 17.35	-.01 .09 .03 .48	-.02 .43 .11 1.79	.98 .68 .91 .09
Movakic standing	.50	.16	3.48	.09	Gender Age GMFCS Movakic standing	7.45 .36 -8.20 35.04	9.60 1.14 7.23 18.79	.26 .11 -.29 .46	.78 .31 -1.13 1.87	.46 .76 .28 .09

R^2 = R square
ΔR^2 = R square change
ΔF = F Change
P ΔF = Sig F. change
B = Unstandardized coefficient values
SE = Standard Error
β = Beta
t = t-test
p = Significance

DISCUSSION

This first specific study into the effect of motor abilities on quality of life (QoL) of children with severe multiple disabilities (SMD) shows that a relatively higher level of motor abilities is significantly related to higher QoL. Motor ability is most strongly correlated to the QoL dimension 'physical well-being' and to a lesser extent to the dimensions 'development' and 'activities'. Multiple regression analyses showed significant independent relationships between motor abilities and QoL for almost all domains of motor abilities.

Quality of life is an important aim of treatment for children with SMD. The results of our study show that different motor abilities have a relation to QoL that is not explained by GMFCS level. Supporting children in improving their motor abilities, which enables them to have more sense of control over their environment, can thus contribute to QoL. This study gives a clear justification for the assumption that stimulation of motor abilities, even for those children with a very low level, is most likely to promote their sense of well-being. As QoL is considered a multidimensional construct; in literature no consensus on the relevant domains or the content of the QoL domains exists (Tsoi et al. 2012). In a review of Albers et al. 29 questionnaires on QoL were found, various domains such as physical, psychological, social, spiritual, emotional, communicative and material aspects have been identified as domains that are of importance to a person's total well-being (Albers et al. 2010). In our study positive relationships were found between the QoL domains 'physical well-being', 'development' and 'activities' and motor abilities. In contrast, no significant relationships were found between motor abilities and the QoL domains 'material well-being', 'communication & influence' and 'socio-emotional well-being'. The QoL domains that were found to be positively correlated to the level of motor ability in our study seem relevant to the study population. Although QoL is a broad concept with many possible domains, it is important to predetermine your goal of measuring it in a specific population. The three QoL domains 'physical well-being', 'development' and 'activities' correlate positively with motor abilities in this population, but that is not to say that the same will be true in another population.

Our findings are consistent with earlier studies, where motor abilities are associated with QoL in other groups of children with disabilities. In a study of Dickinson et al. the authors found that children with Cerebral Palsy with poorer walking ability had poorer physical well-being scores (Dickinson et al. 2007) and Shelly et al. concluded that physical well-being domains (f.e. abilities in participation and using arms and legs) of QoL are more strongly associated with functioning in general than psychosocial well-being domains (Shelly et al. 2008). Schoenmakers et al. showed that in children with Spina Bifida being able to move independently appeared to be much more important for daily life function and quality of life than other medical indicators of the disorder

(Schoenmakers et al. 2005). The conclusion of a study of Raz-Silbiger et al. was that gross motor skills of children with Developmental Coordination Disorders do appear to be somewhat related to the physical and school dimensions of their QoL (Raz-Silbiger et al. 2015). In a study of Petry et al. (2009), where the association between QoL, motor function, personal characteristics and characteristics of care settings were studied in people with SMD, the authors, surprisingly, found no significant relation between motor limitation and QoL scores. However, the absence of such a relationship was insufficiently discussed in the study since motor limitations were operationalised by the number of physical limitations in body parts, muscle tone and deformities and not by measuring the level of motor abilities.

Adding to this body of knowledge, the results of our study underline the importance of stimulating motor abilities in this severely disabled group of children. Moreover, small changes in motor ability in this specific population do matter since we found such an important positive relationship between motor ability and the QoL domain of physical well-being.

We should realise that motor abilities of children with SMD are severely compromised and support is often needed in order for them to complete daily activities. This was an important fact in the development of Movakic. The expert group concluded that motor abilities in this group of children are mostly based on support by other persons through facilitation of movement and support through assistive devices in combination with activity of the child itself (Mensch et al. 2015c). As shown in this study, other existing tests evaluating motor abilities, applicable to children with motor disabilities, do not allow for manual support. Therefore, change in motor abilities, as measured by Movakic, is to a great extent dependent on change in level of support or facilitation, but also on change in level of activity of the child itself. This again supports our statement that active participation of the child during all activities of daily life, i.e. by stimulating motor abilities, is a prerequisite to be more active in controlling parts of the environment of these children, which influences their QoL.

A limitation of our study was the small sample size, which may influence the correlation analyses. As a result standard deviations for both scores seem large; this is likely due to a combination of a low number and heterogeneous composition of respondents. This usually causes a lower chance of a significant result. There is thus a chance that e.g. moderate relationships could have been significant in higher numbers and / or a more homogeneous group.

Despite the small size of the study population, children of nine different care organizations were included which makes the results more representative of the Dutch population. Another issue is that motor abilities of these children were measured in a body position that is relevant to the individual child. The total score and the body position scores of Movakic for each child are based on the scores of these relevant situations and thus can differ per child. Therefore, the resultant score of motor abilities does not express the motor abilities in general. On the other hand, the theoretical framework of Movakic was based on the fact that instruments measuring general motor abilities are not relevant and applicable to children with SMD (Mensch et al. 2015c). So, there simply is no instrument that measures general motor abilities in this population and it is infeasible to create such an instrument because of the characteristics and heterogeneity of these children.

Based on the knowledge that so many variables could influence QoL, positively or negatively, we would like to urge health professionals to apply every bit of solid evidence to make the lives of these children more pleasant. More specifically, based on the results of this study we recommend physical therapists to invest in their interventions focussed on stimulating and activating children with SMD with the aim of increasing their motor abilities. Parents and care-givers play an important role in the success of this intervention. Therefore, after completing Movakic, we recommend involving parents and caregivers. Discussing the scores per item will help them in better understanding the need for investing their time in stimulating motor abilities and in achieving goals in the domain of motor abilities.

In this first study in children with SMD we only studied cross-sectional relationships and not longitudinal relationships. A longitudinal study design is recommended to judge the influence of changes in motor abilities on QoL. A Randomised Controlled Trial (RCT) with a large study population and a long period of intervention, aimed at improving motor abilities, is recommended. An earlier study on the responsiveness and validity of Movakic showed the importance and multitude of life events that children with SMD have to deal with (Mensch et al. 2016). Therefore, the relationship between carefully inventoried events such as change in medication, disease, surgery and interventions and its influence on QoL should be considered in future studies in this severely disabled group of children.

CONCLUSION

This study shows that motor abilities in children with severe multiple disabilities are related to quality of life. Better motor abilities were moderately associated with higher QoL levels, indicating the importance of motor abilities, even in this severely disabled group of children.

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APPENDIX A. Additional information about Movakic (Table A.1, A.2, A.3)

Table A.1. Structure of Movakic

Positions	**Lying**	**Sitting**	**Standing**
Situations ⟶ Grouping of motor abilities ↓	1 Supine 2 Supine with device* 3 Prone 4 Prone with device 5 Side 6 Side with device	7 Flat surface 8 Dangling legs 9 Chair/ sitting device 10 Feet on subsurface	11 Without device 12 With device
Maintaining position	Items (see table A.2) with questions (table A.3) on 1 Intensity manual or support by device 2 Activity of the child 3 Intensity of manual facilitation/ stimulation		
Activities			
Changing body position			
Moving around			

*Device: Assistive devices such as prostheses, ortheses and specialized tools and aids for personal mobility such as canes, walkers and wheelchairs. (World Health Organization., 2008)

Table A.3. Example of questions and answer categories of an item

What is the extent of manual support you gave the child to maintain this position?	O O O O O Full ... None
What is the child's level of activity?	O O O O O Full Passive ... Full Active
What is the extent of facilitation to stimulate the motor ability?	O O O O O Full ... None

Table A.2. Items of Movakic

	Situations (see table A.1)												
	Items	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**
1	Maintaining position	x		x		x		x	x	x	x	x	x
2	Duration maintaining position					x		x	x	x	x	x	x
3	Turning head	x	x	x	x	x	x	x	x	x	x	x	x
4	Upright head	x	x	x	x	x	x	x	x	x	x	x	x
5	Maintaining upright head position	x	x	x	x			x	x	x	x	x	x
6	Reaching with the arms	x	x	x	x	x	x	x	x	x	x	x	x
7	Take support (fore)arms			x	x								
8	Take support hands			x	x			x	x			x	
9	Grasping with the hands	x	x	x	x	x	x	x	x	x	x	x	x
10	Roll over to the left	x		x									
11	Roll over to the right	x		x									
12	Roll over to prone	x				x							
13	Roll over to supine			x		x							
14	Transfer from lying to sitting	x	x	x		x							
15	Transfer from sitting to lying							x	x				
16	Transfer from sitting to standing							x		x	x		x
17	Transfer from standing to sitting									x	x	x	x
18	Pivoting							x					
19	Minor voluntary postural changes								x	x	x		
20	Move on	x		x				x		x		x	x
21	Distance	x		x				x		x		x	x
22	Move on and change direction									x		x	x
23	Moving backwards									x		x	x

x= situation containing the item

CHAPTER 7

GENERAL DISCUSSION




GENERAL DISCUSSION

Main findings

This thesis describes all relevant steps in the development of Movakic, an instrument for physiotherapists to evaluate and follow-up over time motor abilities, specifically in children with severe multiple disabilities (SMD). The main reason for developing a new instrument named Movakic (**mo**tor e**va**luation of **ki**ds with multiple and **c**omplex disabilities) is the need for a practical instrument. Furthermore, based on our systematic review none of the existing instruments, evaluating motor abilities in children with disabilities, are suitable for children with SMD. In the development procedure the starting point was the experience and knowledge of an expert group, consisting of physiotherapists with longstanding specialist expertise treating this group of children. This expert group addressed/formulated the suitability criteria and designed the construct of the new instrument, resulting in a high feasibility and content validity. After the construction of Movakic a longitudinal study in 60 children with SMD was performed. We can conclude that the test-retest and inter-rater reliability were very high (range ICC=.72-.98). Furthermore, the construct validity (*r*= .50 - .71) and responsiveness for changes after meaningful events were satisfying. The study where the relationship between motor abilities and quality of life was explored showed that higher scores in motor abilities were moderately associated with increased quality of life scores.

Our main finding is that Movakic is a suitable instrument for the evaluation of motor abilities in children with SMD. It is recommended for implementation in care-organizations where persons with SMD live or receive day-care. Movakic is provided on-line through a secure internet site and after completion of the questionnaire physiotherapists receive a report with the scores for each child. The report is suitable for adding to the electronic client dossier. Based on our experience during the development process we can conclude that training of physiotherapists is required before Movakic reliably can be used in clinical practice.

Methodological issues

Movakic is a performance-based measurement and consists of items related to the performance of motor abilities of children with SMD as observed by physiotherapist that have longstanding treatment experience with the individual child. From practical experience we know children with SMD may vary considerably under the influence of alertness, fatigue, health, medication use, or unfamiliar circumstances. That is the reason a questionnaire evaluating performance in day-to-day situation is the best choice and can prevent disagreement and "bias" as opposed to testing in standardized test conditions (Dobson et al., 2012). This choice also has the added benefit of no burdening the child by having to put it through extra testing.

A relevant issue of the construct and criteria of Movakic is that is we have to face subjectivity due to the application of manual support ("hands-on"). Although inter-rater reliability of Movakic is found very high, objectifying the extent of "hands-on" remains debatable and personal. We stress again that performing physical activities independently is problematic in the target group and as a result, direct support of other persons is necessary in all activities of daily life (Putten, Bossink, Frans, Houwen, & Vlaskamp., 2016). Agreement between observers of the definition of extent of "hands-on" support is therefore very important to reliably use the Movakic instrument. To improve agreement, "hands-on" was described in the user's manual of Movakic and during the required training considerable time was spent discussing the uniform use of "hands-on". We feel reading the manual and participating in the training is needed to maximize agreement between the physiotherapists.

The absence of a gold standard, applicable to children with SMD, was of methodological importance for the validity study, leading to a compromise in testing the construct validity by comparing motor ability scores and Visual Analogue Scale (VAS) (Reips & Funke., 2008) scores. It would have been better to compare Movakic scores with those of relevant parts of existing tools. However, first of all other tools do not allow for hands-on support rendering them unusable for comparison. Secondly, the necessity of repeated physical assessments would also have meant a higher burden for the participating children. So we decided that using the VAS was the most optimal solution.

In order to evaluate responsiveness of Movakic to clinically relevant changes during the follow-up period of 18 months we asked participating physiotherapist to report events in the life of the child. A strong point of the design of this study was its close relation with usual care in these children. These children go through a lot of life events that might influence motor abilities. The expectation was that changes in Movakic scores were either results of physiotherapeutic intervention, surgery or of other relevant interventions, or the result of comorbidity or other influences. It remains difficult to establish the impact of a certain physiotherapeutic intervention in this vulnerable group of children when these children are subject to concurring illness or recovering from a medical intervention at the same time. All these influences will either have a positive or negative impact on motor ability. However, their individual share in the net effect on motor ability is hard to assess. In order to work around the complexity of analysing individual impact of events, we chose to compare Movakic score changes in two datasets: one containing 3-month intervals with events and one without events.

This meant that Movakic data of the same child could be used in both datasets, as a child could be without events in one 3-month interval but have had surgery in the next for example.

Lastly, relevance of the interpretation of changes in scores is a topic of debate for many years (Maas., 2017). In order to interpret changes in Movakic scores, there needs to be agreement on the size of impact of events on motor abilities in children with SMD. We propose to carefully record events and regularly perform measurements using Movakic in a longitudinal study. At least, the measurement should be performed after every meaningful event to minimize the chance of another event that could introduce bias in establishing the net effect of the first recorded event. Next, we recommend an intervention study that aims to establish the net effects of physiotherapeutic interventions on motor abilities measured with Movakic, which using the data of the longitudinal study could be controlled for bias/confounding introduced by co-occurring events.

Implications for clinical practice

Use of Movakic might help identify change in motor functioning, as a result of influences with negative impact on motor ability such as having pain or longer disturbances of health and fitness or influences indented to have a positive impact such as training motor abilities, the use of devices or undergoing surgery or other specific therapeutic interventions. As stated in the last paragraph of the previous section, we recommend that in clinical practice such influences should be registered with a standardised, longitudinal and preferably objective documented method. Using such a standardized method would enable objectifying the effects of interventions on motor abilities in clinical practice as well in scientific (intervention) studies.

Another potential use of Movakic is systematically recording motor ability in the individual child with SMD as part of the evaluation of multidisciplinary goals. In a multidisciplinary setting all disciplines work on the same perspective of the individual child with SMD. However, all disciplines have their own specific goals often leading to discussion of which intervention could have led to the result or which intervention has the priority. By formulating clear goals with the child's perspective in mind we can try to prevent this. Movakic consists of items on specific motor abilities clustered in relevant body positions with or without material aid, and clusters of items focussing on specific perspectives. These items on specific motor abilities on maintaining body positions, activities, changing body position and moving around can be helpful in formulating clear goals and evaluating treatment goals in multidisciplinary settings. In multidisciplinary settings such as in day-care centres, evaluating goals often follows the treatment plan cycle. Moreover, based on the practical experience in working with children with SMD we know the difficulties in formulating clear, achievable and relevant goals containing small steps. Movakic can help physiotherapists by identifying relevant goals on the domain of motor abilities. Especially in the intervention of children with SMD, intervention needs to be implemented in all functional activities of daily life involving parents and other care-givers in order to maximize repetition and in turn learning. Formulating achievable goals will help motivate working on goals. In addition, parents and care-givers reacted very positively on the fact that knowledge of possibilities in motor abilities, instead of what the disabilities of their child are, helped them focus on relevant aspects of stimulating their child. As mentioned before, the limitations of other observational instruments such as relevance of items and big steps in scores which were hard to achieve (Mensch et al., 2015a) demotivated physiotherapists, parents and care-givers.

It is generally acknowledged that physical activity is relevant for prevention of health issues such as cardio-vascular diseases, poor physical fitness and cognitive problems (Hartman, Houwen, Scherder, & Visscher., 2010) (Houwen, Putten, & Vlaskamp., 2014), (Nederlandse Hartstichting., 2017) (Bartlo & Klein., 2011). We assume that this applies to people with SMD too. Moreover, it demonstrates the importance of measurement of motor abilities and quantifying activity of the child with SMD. Unfortunately, due to the budget cuts in the Dutch care system, the number of professional care-givers is rapidly declining. Children with SMD need help from other people in all activities of daily life (Putten et al., 2016). By the decrease in number of qualified care-givers the burden of care is increasing exponentially, resulting in for example less time for the children. Children with SMD have significant lower reaction time so reduction of qualified care-givers in our care system may directly influence their quality of life (QoL). A recent study on motor activation in people with profound multiple disabilities showed that motor activation in persons with PMD in daily practice by support of persons is rather limited. The consequences can be extensive and related to nearly all domains of human functioning and as it is on their QoL (Putten et al., 2016). In addition, interesting is the relation we found between motor abilities and QoL (Mensch et al., submitted) because maximizing QoL of children with SMD is often the most important objective of treatment and care. A range of health problems often threatens QoL of these children and care or treatment concentrate on the prevention and relief of suffering. Although problems in motor abilities are not the most important aspect children with SMD have to deal with, stimulating motor abilities instead of accepting inactivity can help these children in alertness and being active during the day (Houwen et al., 2014), (Putten, Vlaskamp, Reynders, & Nakken., 2005).

Recommendations

Good quality research investigating the measurement properties, and in particular the responsiveness and interpretability of performance-based measures, is needed (Dobson et al., 2012). Responsiveness is an important measurement property when used to evaluate patients over time (Mokkink et al., 2010). We recommend a study aiming to evaluate the responsiveness of Movakic related to the impact of events and to establish reasonable estimates of relevant changes in motor abilities in order to provide practical guidelines how to interpret changes in Movakic scores.

Specifically in this target group the effects of physiotherapeutic or other interventions on motor abilities are often unknown. Movakic can be used in intervention studies that aim to establish the effect of interventions such as specific medication or surgery on motor abilities in children or persons with SMD. We know children with SMD are a small but significant subgroup of persons with intellectual disabilities (Health Care Inspectorate [Inspectie voor de gezondheidszorg (IGZ)]., 2000). In order to increase evidentiary power we recommend performing studies by adding adults in addition to children with SDM to the study population. Based on the findings from the first evaluation of applicability (Mensch et al., 2015a), Movakic is a feasible instrument for adults too.

In summary, we recommend Movakic in clinical practice of physiotherapists working with persons with SMD. Despite some methodological issues caused by the complexity of objective evaluation in this group, the results of this thesis are that Movakic has adequate feasibility, reliability, validity and responsiveness (Mensch et al., 2015a) (Mensch, Rameckers, Echteld & Evenhuis., 2015b) (Mensch, Echteld, Evenhuis, Rameckers., 2016). To use Movakic one needs to be trained and build experience in using Movakic. Practicing with individual cases is very important during training of Movakic. We plan to work with experts form care-organizations around the country to ensure this online instrument is kept available and remains up-to-date.

Personal note about this special group

In addition to the main subject of this thesis I also feel responsible to raise my concerns related to this vulnerable group of children. One of my concerns is the discrepancy in our care system. The lack of sufficient, objective, relevant and scientific evidence, specifically related to this multiple disabled group, leads to an incomplete analysis on the effectiveness of intervention. As a result there is an underestimation of the real needs and support required to help children develop and maximize their quality of life. These children are probably not receiving enough care. In addition, the lack of information and thus evidence might cause health insurance companies to reduce funding and cut budgets intended to be used in the execution of The Social Support Act (WMO).

Policymakers too increasingly require objective information. Again, as described in this thesis, I want to emphasize the complexity and importance of objective evaluation of interventions and how it affects the wellbeing of this group. However this group is relative small, often unknown and heterogeneous (Health Care Inspectorate [Inspectie voor de gezondheidszorg (IGZ)]., 2000) and therefore the acquisition of grants and funds required for research is very difficult. I would advocate relying more on practice based evidence than on evidence based practice in this group of children.

My second concern is that since January 2015 the municipalities are responsible for all youth welfare and youth care services, ranging from preventive services to child protection services. The new youth care system should be more efficient, coherent and cost-effective. A focus on prevention, the capacities of youth and parents, support at an earlier stage, care made to measure and a better cooperation between professionals was thought to decrease the use of specialised services (Bosscher., 2014). Unfortunately, children with SMD and their parents need to be helped by specialists and need specialised, often costly services. Due to the lack of objective information, the inefficiency of the new care system and the lack of specialists who know the needs of this group, these children and their parents are the victims of this transition. Paradoxically, as a group, the health of children with SMD has improved over time causing some children to survive into adulthood. From the age of 18 these children are considered adults by law, which means smaller budgets while their need for specialised and often expensive care has not changed. I will give one very distressing example. In the case of a child with SMD who doesn't have a feeding tube, and needs to be fed by a care-giver specialized in preventing aspiration causing the child to complete his meal half an hour later than a child without SMD. When this child reaches the age of 18 by law this child is formally an adult and so it will be placed in a day-care centre for adults. Although this client has the same special needs, care-givers in day-care centres for adults do not have the time needed for example to feed clients in this manner. Parents depend on the care of a specialized day-care centre so they can continue caring for their child in their own home during the evenings and weekends. It also enables them to participate in society as a whole by being able to work during the day or care for other family members that might need help. They cannot be expected to provide 24-hours care because of the child's serious health problems and the intensive care it needs. If day-care is not available because of a reduced budget, parents will be forced to place their child out-of-home or accept that in order to remain at home their child needs to be fed through a gastro-intestinal feeding tube. Parents of children with SMD are obviously aware that the life expectancy of their child is limited, but like every other parent they would like to see their child live a full life and that includes their life after the age of 18 years. However, by limiting available care after they reach adulthood, this wish of parents will not be fulfilled. In my opinion a highly unethical issue.

Take home messages

- Movakic has adequate feasibility, reliability, validity and responsiveness and can be used in clinical practice of the physiotherapist.
- Training in the use of Movakic is required for valid objective evaluation.
- Objective evaluation of treatment effects in this vulnerable group continues to be complex.
- Evidence based practice in this group should be replaced by practice based evidence.
- Although problems in motor abilities are not the most important aspect children with SMD have to deal with, using motor abilities, necessary for daily functioning, should be stimulated in order to maximize various domains in quality of life.

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CHAPTER 8

SUMMARY
SAMENVATTING
DANKWOORD
OVER DE AUTEUR
PHD PORTFOLIO

   

SUMMARY

Chapter 1. General introduction

This thesis starts by introducing the group of children with severe multiple disabilities and their severe health problems through a case description of Jeroen. Such health problems are often linked to or arise as a result of their severe motor disabilities. Motor abilities are the field of the (pediatric) physiotherapist. We describe how – although the disabilities will always remain severe - even small changes in motor abilities, with or without support, may lead to an increased control over their own life, resulting in a better quality of life. In addition to the benefits for children, the burden of care for parents and caregivers may also be diminished.

Information is given about the International Classification of Functioning, Disability and Health for Children and Youth from the World Health Organisation (ICF-CY). This classification conceptualizes functioning as an interaction between the levels of 'body functions and anatomical characteristics', 'activities' and 'participation'. Activities and participation are influenced by 'environmental factors' and 'personal characteristics'. The physiotherapist focuses mainly at improving the activity level, whereas multidisciplinary collaboration in treatment and support is mainly directed towards positively impacting at the participation level.

In current clinical practice, evaluation of the often-subtle effects of interventions, that may take longer periods of time, is primarily based on subjective information of the therapist. Nationally and internationally, the need for a method to more objectively measure and follow up clinical progression or deterioration in this group of children (and adults) is increasingly being felt in the field of physiotherapy.

Therefore, finding, adapting or developing such an objective method, applicable to this relatively small group of children with very specific problems was the aim of the studies described in this thesis.

Chapter 2. Instruments for the evaluation of motor abilities for children with severe multiple disabilities: A systematic review of the literature

Eighteen studies in groups of children with different motor conditions and disabilities were found; they described a total of eight instruments. One of these had been specifically developed for the group of children with severe multiple disabilities. Quality of all studies was judged using the COSMIN (COnsensus-based Standards for the selection of health Measurement INstruments) protocol. Reliability as well as validity and responsivity had been studied in only two instruments. Of all eight instruments, only part of the essential statistical properties had been evaluated, or methodological quality was unsatisfactory, or outcomes of different studies were contradictory.

Chapter 3. Design and content validity of a new instrument to evaluate motor abilities of children with severe multiple disabilities: Movakic

With an expert group consisting of six physiotherapists and one occupational therapist, working in intellectual disability care, all of them with over 15 years of experience with the target group, a new instrument for the evaluation of motor abilities at the level of 'activities' was systematically developed. The procedure included the following steps: 1. Formulating relevant criteria for an instrument in the target group, 2. Making a list of relevant motor abilities, 3. Designing the instrument's structure and lay-out and 4. Performing a pilot to ascertain applicability.

The expert group decided that the instrument should be a questionnaire, to be completed by the child's physiotherapist, and based upon his/her knowledge of the child's motor abilities over a longer time frame, instead of – as is the current approach – an observation test of the child's performance at a certain moment. The result was Movakic (**mo**tor e**v**aluation of **ki**ds with multiple and **c**omplex disabilities), a questionnaire with 12 situations/groups, each consisting of multiple sub-questions.

The structure of Movakic is shown in the following table:

Positions	**Lying**	**Sitting**	**Standing**
Situations ⟶ Grouping of motor abilities ↓	1 Supine 2 Supine with device* 3 Prone 4 Prone with device 5 Side 6 Side with device	7 Flat surface 8 Dangling legs 9 Chair/ sitting device 10 Feet on subsurface	11 Without device 12 With device
Maintaining position	Items with questions on 1 Intensity manual or support by device 2 Activity of the child 3 Intensity of manual facilitation/ stimulation		
Activities			
Changing body position			
Moving around			

*Device: Assistive devices such as prostheses, ortheses and specialized tools and aids for personal mobility such as canes, walkers and wheelchairs.

Because the children have different capabilities to begin with, only situations that are relevant to the child are scored.

After a 4-hour training, 26 physiotherapists who had not been members of the expert group, participated in the pilot study. They completed the questionnaire for 53 children in total and 15 adults. All participating physiotherapists judged the content and relevance of the items to be good, some items were found difficult to understand.

They offered suggestions for improvement of the wording of items, for the score system and for additions to the training. Content validity, as judged according to COSMIN criteria, was adequate.

Chapter 4. Reliability of Movakic

After improving the instrument according to outcomes of the pilot, test-retest reliability and inter-rater reliability were assessed. Thirty-seven trained physiotherapists participated, all working in specialized children's day-care centres and with over 10 years of experience with the target population. Each of them selected one or two children of their own clients, resulting in a total of 60 participating children.

Test-retest reliability could be assessed for 50 children: intraclass correlations for the total score (ICC=.95, 95%-confidence interval (CI) .92-.97) and for scores of the four groups of motor abilities (range .72-.98) were all excellent or good.

In 19 children for whom a second therapist was available who was familiar with the child too, inter-rater reliability could be established. Intraclass correlations for the total score (ICC=.94, 95%-CI .85-.98) and scores for the four groups of motor abilities (range ICC=.85-.97) were also excellent or good.

Therefore, we conclude that Movakic is a reliable instrument for children with severe multiple disabilities.

Chapter 5. Construct validity and responsiveness of Movakic

After establishing reliability, in a prospective cohort design, Movakic was completed six times during 18 months by the same 37 physiotherapists for the same 60 children. For each child, one situation had to be selected that was relevant to the child and was expected to remain stable during follow-up.

In order to measure construct validity, scores were compared with the expert judgement of the physiotherapists by means of Pearson correlations. Prior to completion of Movakic, experts had to report their scores on a visual-analogue scale; these scores could not be seen or changed afterwards. Construct validity was good (r=.50-.71).

In order to measure responsiveness to change, score-changes in 3-month-periods, in which no relevant influences had been reported for the child, were compared with score-changes in periods during which events had occurred that might have influenced the scores positively or negatively, using intraclass correlations. To this end, after each 3-month-period, the physiotherapists reported the relevant events. The following events were used for analyses; surgery, increase of contractures and pain.

Responsiveness was demonstrated by significantly larger score-changes during periods with relevant events than during periods without such influences: mean absolute Movakic score-changes were significantly larger in the group with events (t=2.21;p=.03), albeit with a moderate effect size (Cohen's d=.62, 95%-CI .23-1.00). A comparable tendency was seen for intraclass correlation coefficients: in the group with events it was .78 (95%-CI .64-.86) and in the group without events .95 (95%-CI .94-.96).

We conclude that Movakic has good construct validity and is responsive to change. However, it was unclear to what extent the events reported had really influenced the child, nor whether such influences improved or worsened motor function. Further studies should include interpretation of score-changes and what they mean clinically for children and parents. Our experiences clearly demonstrate how complex scientific research in this group of children can be.

Chapter 6. The relation between motor abilities and quality of life in children with severe multiple disabilities

In a group of 29 children with severe multiple disabilities, motor abilities were scored with Movakic by their own physiotherapists. The parents of these children scored quality of life using the Quality of Life-Profound Multiple Disabilities (QoL-PMD) questionnaire. Scores were compared by means of Pearson correlations.

A moderate but significant correlation was found between Movakic and QoL-PMD total scores (r=0.40, p=0.03). Significant correlations with total Movakic scores were specifically found for the quality of life dimensions 'physical wellbeing', 'development' and 'activities'.

We conclude that in children with severe multiple disabilities, higher scores on motor abilities are associated with higher quality of life scores.

Chapter 7. General discussion

The careful work of an expert group with longstanding specialist experience in physiotherapeutic treatment of children with severe multiple disabilities resulted in a structured method to objectively evaluate subtle changes in motor abilities. Movakic is a well applicable, reliable and valid instrument. Its scores are sensitive to change. However, scores are influenced just as much by the frequent acute and chronic comorbidity, medical accidents, changes in medicine use, surgery and other events as by physiotherapeutic intervention.

In individual clinical practice, therapists should carefully include all of these possible influences in their evaluations of interventions in these children and adults.

Furthermore, the necessity to accept and include 'hands-on' support in Movakic's structure is discussed, as well as the necessity of training of physiotherapists in the application of Movakic, and methodological barriers in the studies are described in this chapter.

Also, recommendations for clinical practice are given, specifically for work in multidisciplinary teams.

Finally, the author expresses her personal concerns about the lack of scientifically supported diagnostics and therapies for this severely disabled group. This may lead to an underestimation of care needs by e.g. policy makers and the use of ineffective methods.

Policy makers and health insurance companies increasingly require evidence-based diagnostics and treatment; lack of such evidence for time-consuming interventions by highly specialised professionals may have negative budgetary consequences. The same applies to the fact that care of this group of children is now primarily the responsibility of municipalities.

On the other hand, collecting scientific evidence in this severely disabled group remains difficult due to the complexity that became apparent in the current studies and to the lack of grant opportunities as a result of the limited size of the target group.

In clinical practice, Movakic can now be used online and an English version of the instrument is in the process of completion.

SAMENVATTING

Hoofdstuk 1. Algemene inleiding

Aan de hand van de casus van Jeroen wordt de doelgroep van kinderen met ernstige meervoudig beperkingen en hun vaak ernstige gezondheidsproblematiek geïntroduceerd. Veel van deze problemen hangen samen met of zijn het gevolg van hun ernstige motorische beperkingen. Dit is het werkterrein van de (kinder) fysiotherapeut. Toegelicht wordt hoe - hoewel de beperkingen altijd ernstig zullen blijven – ook minieme verbeteringen van de motorische mogelijkheden, al of niet met ondersteuning, kunnen leiden tot enige zelfstandigheid en daarmee een toename van de kwaliteit van leven. Bovendien kan daarmee de fysieke belasting van de ouders en verzorgenden verminderd worden.

Informatie wordt gegeven over de 'internationale classificatie van functioneren, beperkingen en gezondheid bij kinderen en jongeren (ICF-CY)' van de Wereldgezondheidsorganisatie. Daarin wordt onderscheid gemaakt tussen de niveaus van 'lichaamsfuncties en anatomische kenmerken', 'activiteiten' en 'participatie'. Activiteiten en participatie worden beïnvloed door 'omgevingsfactoren' en 'persoonlijke kenmerken'. De doelen van de fysiotherapeut liggen voornamelijk op het niveau van 'activiteiten', terwijl het multidisciplinaire samenspel van behandeling en ondersteuning vooral gericht is op het niveau van 'participatie'.

In de praktijk is de beoordeling van het vaak subtiele effect van behandelingen, die zich over lange tijd kunnen uitstrekken, voornamelijk subjectief. Fysiotherapeuten in binnen- en buitenland hebben behoefte aan een methode waarmee ze vooruitgang of achteruitgang bij deze kinderen (en volwassenen) objectiever kunnen vastleggen en volgen.

Het vinden, aanpassen of ontwikkelen van zo'n objectieve methodiek, geschikt voor deze relatief kleine groep van kinderen met zeer specifieke problemen (in Nederland ruim 2000 onder de leeftijd van 18 jaar), was dan ook het doel van de studies in dit proefschrift.

Hoofdstuk 2. Systematisch literatuuronderzoek naar bestaande methodieken (instrumenten) en de methodologische kwaliteit van het verrichte onderzoek

Er werden 18 studies bij kinderen met verschillende aandoeningen en motorische beperkingen gevonden, die acht instrumenten betroffen. Hiervan was er één speciaal voor de doelgroep ontwikkeld. De kwaliteit van die instrumenten werd beoordeeld met behulp van het COSMIN protocol.

Van slechts twee instrumenten waren zowel de betrouwbaarheid als de validiteit en responsiviteit bestudeerd. Van alle acht instrumenten waren of de essentiële statistische eigenschappen niet bestudeerd, of de methodologische kwaliteit schoot tekort, of de uitkomsten van verschillende onderzoeken spraken elkaar tegen.

Hoofdstuk 3. Ontwikkeling en inhoudelijke validiteit van Movakic: een nieuw instrument voor de evaluatie van motorische vaardigheden bij kinderen met ernstig meervoudige complexe beperkingen.

De bestaande instrumenten uit de systematische literatuurstudie zijn vervolgens getoetst op toepasbaarheid voor de doelgroep. Naar aanleiding van deze toetsing bleek dat geen van de instrumenten geschikt waren of aangepast konden worden.

Door een expertgroep van zes (kinder)fysiotherapeuten en een ergotherapeut, werd systematisch een nieuw beoordelingsinstrument ontwikkeld voor motorische vaardigheden op het 'activiteiten' niveau. Alle experts waren werkzaam in de zorg voor mensen met verstandelijke beperkingen en hadden meer dan 15 jaar ervaring met de doelgroep. De volgende stappen werden afgelegd: 1. Opstellen van criteria waaraan een instrument voor deze doelgroep moet voldoen, 2. Opstellen van een lijst met relevante motorische vaardigheden, 3. Ontwerpen van een structuur en lay-out van het instrument, 4. Een pilotstudie naar de toepasbaarheid.

De expertgroep besloot dat het een vragenlijst zou moeten worden in plaats van zoals gebruikelijk is, een observatietest van vaardigheden op één moment. Deze vragenlijst dient ingevuld te worden door een behandelend (kinder)fysiotherapeut en is gebaseerd op diens kennis van de motorische mogelijkheden van het kind over enige tijd. Het resultaat was Movakic (**mo**torische **va**ardigheden van **ki**nderen met ernstig meervoudige **c**omplexe beperkingen), een vragenlijst bestaande uit 12 situaties/groepen met een variabel aantal items, elk met zijn eigen sub-vragen. De items betreffen motorische vaardigheden die gaan over de onderdelen; handhaven van een houding, activiteiten in een houding, transfers (houdingswisselingen) en voortbewegen.

Zie voor de structuur van Movakic deze tabel:

Posities	Liggen	Zitten	Staan
Groepen ⟶ Groepen van motorische vaardigheden ↓	1 Op rug 2 Op rug met hulpmiddel 3 Op buik 4 Op buik met hulpmiddel 5 Op zij 6 Op zij met hulpmiddel	7 Effen ondergrond 8 Afhangende benen 9 Met zithulpmiddel 10 Met voeten op ondergrond	11 Zonder hulpmiddel 12 Met hulpmiddel
Handhaven houding	Items met vragen over: 1 Mate van ondersteuning 2 Activiteit van het kind 3 Mate van manuele facilitatie		
Activiteiten			
Lichaamspositie veranderen			
Voortbewegen			

Omdat de mogelijkheden van kinderen verschillen, hoeven alleen die situaties gescoord te worden die voor het kind relevant zijn.

Aan de pilotstudie naar de toepasbaarheid werkten 26 (kinder)fysiotherapeuten mee die geen lid van de expertgroep waren; zij kregen eerst een training van vier uren en vulden de lijst daarna in voor totaal 53 kinderen en 15 volwassenen. Alle deelnemende (kinder)fysiotherapeuten beoordeelden de inhoud en relevantie van de items als goed, sommige items werden als moeilijk beoordeeld. Zij leverden suggesties voor verbetering van de formulering van sommige items, voor het scoresysteem en voor een uitgebreidere training. De content validiteit, beoordeeld volgens COSMIN-criteria, was adequaat.

Hoofdstuk 4. Betrouwbaarheid van Movakic

Na verbetering van het instrument op basis van het toepasbaarheidsonderzoek werden de test-hertest betrouwbaarheid en de interbeoordelaarsbetrouwbaarheid getoetst. Hieraan werkten 37 getrainde (kinder)fysiotherapeuten mee, die allen werkten in kinderdagcentra en meer dan 10 jaar ervaring hadden. Zij includeerden uit hun eigen cliëntenbestand ieder een of twee kinderen, in totaal 60 kinderen. Voor 50 kinderen kon de test-hertest betrouwbaarheid bepaald worden: de intraclass correlaties voor de totaalscore (ICC=.95, 95% betrouwbaarheidsinterval .92-.97) en de vier onderdelen van motorische vaardigheden (range .72-.98) waren excellent of goed. De interbeoordelaarsbetrouwbaarheid kon worden bepaald bij 19 kinderen, voor wie een collega-fysiotherapeut beschikbaar was die het kind ook goed kende. Ook hier waren de intraclass correlaties voor de totaalscore (ICC=.94, 95% betrouwbaarheidsinterval

.85-.98) en de vier onderdelen van motorische vaardigheden (range .85 - .97) excellent of goed. Movakic is dus een betrouwbaar instrument voor kinderen met ernstig meervoudige beperkingen.

Hoofdstuk 5. Constructvaliditeit en responsiviteit van Movakic

Movakic werd vervolgens in een prospectieve cohortstudie door dezelfde 37 fysiotherapeuten bij dezelfde 60 kinderen gedurende 18 maanden 6 maal ingevuld. Hiervoor werd steeds één voor het individuele kind relevante situatie gekozen, waarvan de verwachting was dat deze stabiel zou blijven gedurende de follow-up. Voor het meten van de constructvaliditeit werden met behulp van Pearson correlaties de scores vergeleken met het expertoordeel van de therapeuten, waarvoor een score op een visueel-analoge schaal moest worden gegeven, voorafgaand aan de invulling van Movakic.

Voor het meten van de responsiviteit werden scoreveranderingen in perioden zonder bijzondere gebeurtenissen, met behulp van intraclass correlaties vergeleken met perioden met gebeurtenissen die de scores positief of negatief beïnvloed zouden kunnen hebben. Na afloop van elke 3 maanden werden door de fysiotherapeuten relevante gebeurtenissen gescoord. De volgende gebeurtenissen zijn gebruikt tijdens de analyses; operaties, toename van contracturen en pijn. De constructvaliditeit was goed (r= .50 - .71). Responsiviteit bleek uit significant grotere scoreveranderingen in perioden met relevante gebeurtenissen dan in perioden zonder zulke gebeurtenissen: de gemiddelde absolute Movakic scoreveranderingen waren significant groter in de groep met gebeurtenissen (t=2.21; p=.03), met een matige effectgrootte (Cohen's d=.62, 95% betrouwbaarheidsinterval .23 - 1.00). Dezelfde tendens werd gezien voor de intraclass correlatie coëfficiënten: in de groep met gebeurtenissen was deze .78 (95% betrouwbaarheidsinterval .64-.86) en in de groep zonder gebeurtenissen .95 (95% betrouwbaarheidsinterval .94-.96).

Echter, van de enorme berg gemelde gebeurtenissen was niet duidelijk in hoeverre ze werkelijk van invloed waren geweest en evenmin of die invloed dan positief of negatief was. Vervolgonderzoek zou gericht moeten zijn op de interpretatie van de scoreveranderingen en de betekenis ervan voor kinderen en ouders. Onze ervaringen uit deze studie tonen aan hoe complex wetenschappelijk onderzoek in deze groep kinderen kan zijn.

Hoofdstuk 6. De relatie tussen motorische vaardigheden en kwaliteit van leven bij kinderen met ernstig meervoudige complexe beperkingen

Bij 29 kinderen werden de motorische vaardigheden door de eigen fysiotherapeut gescoord met Movakic. Kwaliteit van leven werd gescoord door de ouders van deze kinderen met behulp van de vragenlijst over de kwaliteit van leven van personen met ernstige meervoudige beperkingen (Quality of Life - Profound Multiple Disabilities' (QoL-PMD). De scores werden vergeleken met behulp van Pearson correlaties. Een matige maar significante correlatie werd gevonden tussen de totaalscores op Movakic en de kwaliteit-van-leven schaal (r=0.40, p=0.03). Ook werden significante correlaties gevonden met de specifieke dimensies 'fysiek welbevinden', 'ontwikkeling' en 'activiteiten' van kwaliteit van leven.

Uit deze studie kan geconcludeerd worden dat hogere scores in motorische vaardigheden bij kinderen met ernstig meervoudige beperkingen gepaard gaan met hogere scores op de kwaliteit van leven.

Hoofdstuk 7. Algemene discussie

Het zorgvuldige werk van een groep experts met grote ervaring in de fysiotherapeutische behandeling van kinderen met ernstig meervoudige beperkingen heeft geresulteerd in het instrument Movakic. Movakic is in staat om de veelal subtiele veranderingen in motorische vaardigheden objectief in de tijd te volgen, is goed toepasbaar, betrouwbaar en valide gebleken.

De scores zijn gevoelig voor verandering. Echter, de veelvuldige acute en chronische comorbiditeit, medische incidenten, veranderingen in medicijngebruik, operaties en andere gebeurtenissen beïnvloeden de scores evenzeer als gerichte fysiotherapeutische interventie. In de individuele praktijk zal de therapeut met al deze invloeden rekening moeten houden bij de evaluatie van interventies bij deze kinderen en volwassenen.

Nader wordt ingegaan op de noodzaak om manuele ondersteuning gericht op te nemen in de structuur van Movakic, op de noodzaak van training in het gebruik van de methodiek, en op de methodologische barrières in het verrichte onderzoek.

Aanbevelingen worden gegeven voor de praktijk, met name ook voor de multidisciplinaire praktijk. Tenslotte uit de auteur haar persoonlijke zorgen over het gebrek aan wetenschappelijk ondersteunde diagnostiek en behandeling voor de groep met ernstig meervoudige beperkingen. Dit kan leiden tot een onderschatting van de zorgbehoefte en tot het gebruik van ineffectieve methoden. Beleidsmakers en zorgverzekeraars eisen steeds vaker aantoonbare effectieve diagnostiek en behandeling. Het ontbreken van zulk bewijs voor tijdrovende behandeling zou budgettaire consequenties kunnen hebben. Hetzelfde geldt voor het feit dat de verantwoordelijkheid voor de zorg voor deze groep kinderen grotendeels bij de gemeenten terecht is gekomen.

Aan de andere kant is het verzamelen van bewijs door wetenschappelijke evaluatie lastig, enerzijds door de complexiteit die ook in deze studie weer is gebleken, anderzijds door het ontbreken van subsidiemogelijkheden vanwege de beperkte omvang van de doelgroep.

De digitale versie van Movakic is inmiddels gereed en de engelse vertaling is in een gevorderd stadium.

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Natuurlijk 'mijn' klankbord bij de inhoudelijke ontwikkeling van Movakic, allemaal collega's van Ipse de Bruggen. Pauline van den Boogaard, een belangrijke kritische noot, gebaseerd op veel praktische ervaring. Miriam Caminada, bij de totstandkoming van de eerste opzet van Movakic je bijdrage geleverd vanuit je functie als ergotherapeut. Angelique Gestman, met je positieve insteek altijd bereid om te helpen en in te springen waar nodig. Margrethe van der Kleij, nooit liet je verstek gaan, altijd enorm betrokken en nog steeds een intensieve samenwerking. Ria Schmitz, ondanks je werkzaamheden voor Bangladesh toch altijd weer tijd gevonden om je inbreng te hebben in de ontwikkeling van Movakic. Wineke Vlieg, verdiepende vragen stellen, kritisch als altijd met veel expertise en zelfs nu nog betrokken blijven. Hoe kan ik jullie bedanken? Ik probeer het via "mijn welgemeende dank" uit te spreken, voor een ieder binnen haar eigen mogelijkheden en op haar eigen manier. Dat hapje en drankje houden we erin!

Het verwetenschappelijken van Movakic heeft voor een groot deel plaatsgevonden bij het Erasmus MC op de Westzeedijk. In het oude maar sfeervolle pand werkte ik samen met de collega's van de leerstoel geneeskunde voor verstandelijk gehandicapten. Van Sandra, die ook onderzoek deed binnen de kinderen met EMB lijn, en het GOUD-team dat startte met Luc, Thessa, Heidi en Marieke en later ook Alyt en Josje, heb ik veel geleerd. Maar met jullie heb ik ook veel plezier gehad! Aan de IASSID congressen in Zuid Afrika en Rome heb ik fijne herinneringen, ondanks de spanning die een praatje altijd met zich mee brengt. En Jan, de databeheerder, heeft veel tijd en energie gestoken in de digitalisering van het instrument. De samenwerking was op sommige momenten erg intensief, en ondanks dat we soms wat verschillende 'computer' taal spraken was er bij aanvang van de valideringsstudie toch maar echt een mooi en werkbaar instrument! In de tijd dat ik nog regelmatig in Rotterdam werkte heb ik veel onderzoeks-collega's van de afdeling huisartsgeneeskunde in "de 3 kamers" aan de Westzeedijk leren kennen, ik kijk terug op een gezellige en leerzame tijd met jullie. Dederieke, jij hebt je tijdens de laatste loodjes ingezet om me met raad en daad bij te staan. Mijn belangrijkste doel was dat de promotie gepaard zou gaan met de lancering van Movakic, mede dankzij jou is dit gelukt!

Tijdens het zoeken naar en ontwikkelen van een geschikt instrument en tijdens de valideringsstudie van Movakic heb ik gebruik mogen maken van de expertise van vele collega's uit het werkveld. Ongelooflijk wat een draagvlak er was! Ik begin bij de stuurgroep waar ik in 2006 mee van start ging. Naast collega's van Ipse (Marcel Schellart, Anemone Linthorst, Marijke Tonino en Pauline van den Boogaard) waren dat Marjolijn Ketelaar, Eugene Rameckers, Annette van der Putten en Stef van Buuren. Hier werd het fundament gebouwd, een heel belangrijk onderdeel tijdens de ontwikkeling! In 2008 deden we een eerste inventariserend onderzoek naar de toepasbaarheid en betrouwbaarheid van één van de vele versies van Movakic. Aan dit onderzoek deden al 31 collega's vanuit 9 zorgorganisaties mee uit het werkveld. Naast collega's van Ipse de Bruggen; Mirjam Haye, Eline Kuijpers, Annelies Goetheer, Ellen Arensman, Margrethe van der Kleij, Wietske van der Berg, Ria Schmitz, Miriam Caminada, Angelique Gestman, Wineke Vlieg, Pauline van den Boogaard en Willy van der Vegt waren Loek Steenbeek, Hestia Strijkert-Koppers en Sylvia Nieuwenhuis (Kempenhaeghe), Karlieke

Dijkstra en Nanda Ruibing (Hartekamp Groep), Maroeska Wurzer, Marcella Keijmel en Wieteke Bosker (Gemiva-SVG), Inge Maters-Dormolen (Pameijer), Maaike Henneman, Madelon Snijders en Wietske Bakker (ASVZ), Mariska Talitsch, Lieke van Esch, Ineke Groothoff en Mieke Wiersma (SWZ), Els de Weerd en Patricia Volle ('s-Heerenloo), en Marjolein Lincklaen Arriens (Sherpa) bereid deel te nemen, waarvoor mijn grote dank! Aan de training voorafgaande aan de valideringsstudie die in 2010 startte deden 53 collega's mee vanuit 17 zorgorganisaties; Els Simons, Grada Mout, Wietske Bakker, Maaike Henneman (ASVZ), Godie Schuitema (Hartekamp Groep), Anja Stroet-Duijves, Elles Lievendag, Pete de Jager, Janneke Besseling (Esdégé Reigersdaal), Els Heerschap, Wieteke Bosker (Gemiva-SVG), Annelies Goetheer, Eveline Sonneveld, Margrethe van der Kleij, Marieke Poot, Miriam Caminada, Olaf Klein, Pauline vd Boogaard, Renate van Hettema, Renske de Leeuw, Ria Schmitz, Wietske van den Berg, Willy van der Vegt, Wineke Vlieg, Jolanda Fransen, Eline Kuijpers, Angelique Gestman (Ipse de Bruggen), Sylvia Nieuwenhuis (Kempenhaeghe), Kitty Friebel, Maja Kroes (Kleur, Dichterbij), Wilma van Hoorn (nieuw Buitenzorg), Johannes Veldkamp (NOVO), Rieneke van de Meij, Jarno Koorneman (Philadelphia), Els de Weerd, Hans van der Wiel, Harriët van Geffen, Ivonne Foppen, Patricia Volle ('s-Heerenloo), Marieke Ijbema, Ferdy Oudebeek (Sherpa), Malou Houtman (de Driestroom), Cheryll-ann Cabenda, Eliane Knops, Marscha den Exter , Wieske Cronenberg (Omega), Inge Malipaard, Marcel Kouijzer (Tragel), Mariska Talitsch, Mieke Wiersma, Miriam Kuipers- Pelders, Lieke van Esch, Ineke Groothoff (SWZ), Carola Eilander, Emiel Groot Jebbink, Marianne van den Brink, Mariska Sevink, Caroline Leene, Marieke Heyneman, Wim Boon (Zozijn). Wat een enthousiasme ging er van jullie uit, mijn ontzettend grote dank hiervoor! En ik waardeer de inbreng enorm van de collega's Thijs Schilp en Jolanda de Kort (de Witte Vogel) die hebben geholpen bij het kritisch beoordelen en de fine tuning van Movakic.

Een promotie zonder promotor en copromotoren kan niet. Eugène, veel meer dan dat, een vriend, een spraakbaak, een motivator, altijd klaar staan, op ieder tijdstip van de dag.. vanaf het allereerste begin betrokken, en zelfs ver daarvoor. Michael, jij nam het over van Corine. Corine die mij hielp te leren denken op z'n 'wetenschappelijks'. Best een opgave is het voor je geweest om een praktijkmens als ik een beetje om te turnen.. En toch, dat heb je mooi voor elkaar gekregen. En die methodologische kennis en analyses, Michael aan jou de eer om daar met mij op voort te borduren. Het is nog steeds niet mijn sterkste kant, gelukkig was jij er om dat stuk met me op te pakken. Vanuit de praktijk leerde ik Heleen kennen als AVG van 'mijn' bijzonder mooie kinderen. Voor mij heel prettig dat je als mijn promotor begrip had voor mijn praktijkgevoel tijdens het wetenschappelijke stukje van mijn werkzaamheden. Je sprak vaak je waardering voor me uit. Dat motiveerde en relativeerde, ook als het even moeilijk werd. Eugène, Corine, Michael en Heleen. Ontzettend bedankt! Voor de positieve reacties bij de beoordeling van mijn manuscript wil ik prof.dr. S.M.A. Bierma-Zeinstra, prof.dr. M.W.G. Nijhuis en prof. dr. M. Hadders-Algra hartelijk bedanken. En ik vind het een eer dat dr. Marjolijn Ketelaar, dr. Corine Penning, dr. Marijke Tonino en dr. Ruud van Wijck bereid zijn deel te nemen aan de grote commissie.

Paranimfen kiezen was voor mij zo klaar als een klontje. En wat ben ik blij dat jullie deze taak op je wilden nemen! Carine, mijn collega maar vooral hartsvriendin. Vanaf mijn allereerste kennismaking bij destijds nog stichting Ipse, tot bijna 20 jaar later hebben we al veel mogen delen. Met als belangrijkste stuk onze gedeelde passie voor deze bijzonder mooie kinderen. Van verdriet rondom het overlijden van veel van 'onze' bijzonder mooie kinderen, het hebben van een professionele samenwerking op de werkvloer van KDC Zonnehof in Naaldwijk, tot het enorme plezier tijdens de vakanties van het epilepsiefonds waar we als vrijwilligsters een week voor Jeroen, Sven, Larissa en nog veel andere kinderen mochten zorgen. Ik hoop je nog jaren als vriendin en ook als collega naast me te mogen hebben! Rob, mag ik zeggen een beetje 'mijn' Robbie? Vanaf het eerste wetenschappelijke stukje van mijn carrière op de Westzeedijk heb ik je leren kennen. Al snel groeide de 'wetenschap voor dummie' vragen uit tot gezellige fietstochtjes waar we alles maar dan ook alles bespraken. Privé hebben we met onze partners een mooie vriendschap opgebouwd. En zeker ook tijdens de laatste loodjes van mijn promotie was jij degene die er voor me stond. Dat Engels zal nooit helemaal m'n ding worden en jij "editte" alle artikelen voor me. Met als resultaat dat ze alle vijf gepubliceerd zijn. Ook methodologisch hebben we samen nog gestoeid, ik begin die analyses zelfs gewoon een beetje leuk te vinden. Carine en Rob, 'mijn' paranimfen... en daar ben ik heel trots op!

En dan.. het thuisfront. Pap, mam, Hans en Marian (m'n schoonouders), Tan en Dan, altijd kon er iets geregeld worden met de opvang van de kinderen. Hoe fijn is dat! En vooral hoe fijn is het dat jullie er in deze hectische tijd voor mij waren, even een luisterend oor, een advies, even lekker koken of dan toch maar gezellig ter ontspanning samen een wijntje drinken. Edith, m'n nepzus. De omslag van mijn proefschrift met zoveel gevoel. Wederzijdse trots en samenwerken. Hoe bijzonder. Donna, Maecy en Cis, jullie hebben ervoor gezorgd dat ik ondanks de drukke tijden altijd lekker kon ontspannen, kon relativeren en genieten, want jullie zijn het allerbelangrijkste en rijkste bezit in mijn leven! Cin, mijn steun en toeverlaat, begrip als altijd als ik weer eens een 'beetje' stress had, 'even' moest werken of 'iets' moest regelen. Voor ons komt er nu echt meer rust en kunnen we samen optimaal genieten van elkaar en onze heerlijke meiden!

OVER DE AUTEUR

Sonja Mensch is op 23 september 1972 geboren in Leidschendam. In 1988 behaalde ze haar MAVO diploma aan het Maartenscollege in Voorburg. In datzelfde jaar startte ze de MDGO-vz opleiding in Leiden die succesvol werd afgerond in 1991. Direct daarna startte ze aan de Leidse Hogeschool de HBO opleiding fysiotherapie waar ze in 1995 haar diploma haalde. Na het opdoen van werkervaring in verschillende particuliere praktijken begon ze in 1998 de modulaire opleiding kinderfysiotherapie (post-HBO) aan de Hogeschool Breda. Tegelijkertijd kreeg ze een vast contract aangeboden bij de stichting Ipse de Bruggen (destijds stichting Ipse) waar ze nog altijd met veel passie werkt bij behandelcentra voor kinderen met een grote verscheidenheid aan doelgroepen. Haar expertise ligt op het gebied van de interventie bij kinderen met ernstig meervoudige complexe beperkingen. In 2001 werd ze geregistreerd kinderfysiotherapeut en in 2004 startte ze in Breda bij Avans+ de opleiding tot Master Specialized Physical Therapy waar ze in 2006 het diploma behaalde. Tijdens deze studie legde ze de basis van het meetinstrument Movakic. Vanaf 2010 is ze het officiële traject als promovenda gestart aan het Erasmus MC waar de verdere ontwikkeling en validering van het instrument heeft plaatsgevonden. Tevens verzorgt ze lessen aan de Masteropleiding kinderfysiotherapie bij Avans+ in Breda en aan de Hogeschool Rotterdam en ze begeleidt enkele studenten van deze opleidingen met hun afstudeeronderzoek.

Sonja is getrouwd met Cindy Ruijgrok en ze zijn de dolgelukkige ouders van hun 3 dochters Donna (2011), Maecy (2013) en Cis (2017).

PHD PORTFOLIO

Summary of PhD training and teaching

Name PhD student: drs. Sonja Mensch
Erasmus MC Department: Intellectual Disability Medicine, Department of General Practice
PhD period: 2011-2017
Promotor: Prof. dr. H.M. Evenhuis
Copromotor: Dr. E.A.A. Ramèckers, Dr. M.A. Echteld

1. PhD training	**Year**	**Workload (Hours/ ECTS)**
General courses		
Master of Specialized Physical Therapy (Avans+ Breda)	2004	32.0
Biomedical English Writing and Communication	2009	4.0
BROK (Basiscursus regelgeving Klinisch Onderzoek)	2012	1.0
Masterproof Teaching (didactic skills, EBP, examination)	2015	3.5
Specific courses		
Introduction to data analysis (Erasmus Summer Programme)	2007	0.9
Principles of research in Medicine (Erasmus Summer Programme)	2007	0.9
Endnote	2011	8
Presentations		
Dutch Seminar nvAVG/nvFVG (oral presentation)	2012	1.0
Dutch Seminar nvFVG (oral presentation)	2010	1.0
Dutch Seminar KNGF (oral presentation)	2005	1.0
International conferences		
3rd European Congress IASSID, Rome (oral presentation)	2010	2.0
64th International Association AACPDM (poster presentation)	2010	1.0
13th World Congress IASSID, Cape Town South Africa (oral presentation)	2008	2.0
2. Teaching	**Year**	**Workload (Hours/ECTS)**
Lecturing		
Lecturer Master Pediatric Physical Therapist Avans+ Breda	2012-2016	96
Lecturer Master Pediatric Physical Therapist HRO Rotterdam	2005-2016	96
Supervising Master's theses		
2 Master Pediatric Physical Therapy student research projects	2011/2012	80
4 Master Pediatric Physical Therapy student trainees	2014-2017	160
Supervising practicals and excusrions, Tutoring		
Examine student final projects (27 MPPT students)	2014-2016	81
Supervisor research project Physical Therapists	2008	40
Other		
Two day course Measurement training Physical Therapists	2008-2010	48

"MOVAKIC, MOTORISCHE VAARDIGHEDEN VAN KINDEREN MET ERNSTIG MEERVOUDIG COMPLEXE BEPERKINGEN; KLEINE STAPJES, GROTE VERANDERINGEN"