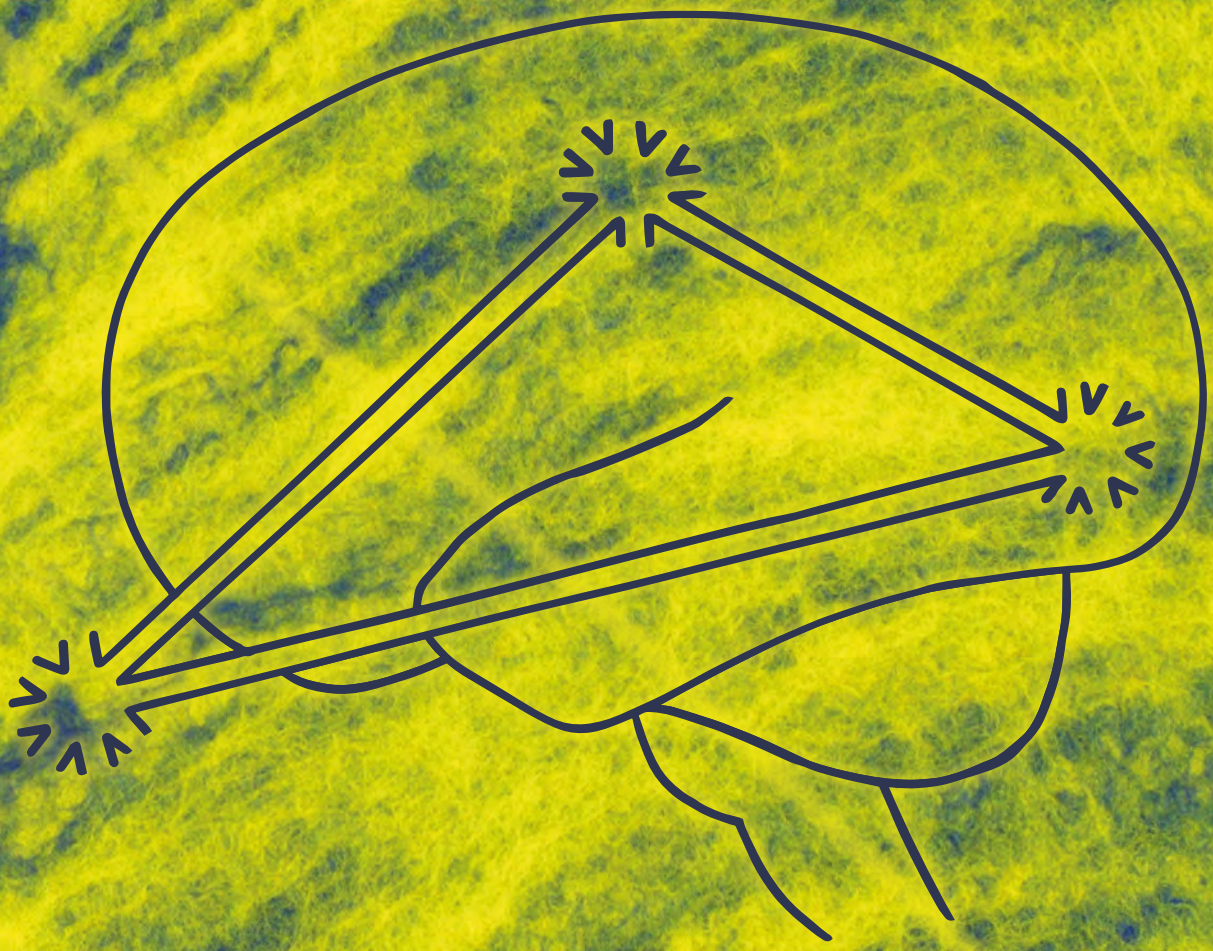
Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI)



Masoud Salavati

Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI)

Abul Salavati

Printing of this thesis is financially supported by




Royal Dutch Visio, Research Group Health Ageing Allied Health Care and Nursing, Hanze University Applied Sciences, Rijksuniversiteit Groningen, Universitair Medisch Centrum Groningen, Graduate School of Medical Sciences

Colofon

ISBN: 978-90-367-8892-2

Cover design: Anet, Sahar en Arman Salavati

Printing: Ridderprint BV - www.ridderprint.nl

The studies presented in this thesis were performed at the Novum Foundation, Huizen, The Netherlands, Royal Dutch Visio and at the Research group Healthy Ageing, Allied Health Care and Nursing Groningen, The Netherlands

Masoud Salavati

Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI)

rijksuniversiteit
groningen

Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI)

Proefschrift

ter verkrijging van de graad van doctor aan de
Rijksuniversiteit Groningen
op gezag van de
rector magnificus prof. dr. E. Sterken
en volgens besluit van het College voor Promoties.

De openbare verdediging zal plaatsvinden op

dinsdag 14 juni 2016 om 11.00 uur

door

Abul Salavati

geboren op 23 oktober 1964
te Ahvaz, Iran

Promotors

Prof. dr. C.P. van der Schans
Prof. dr. B. Steenbergen

Copromotors

Dr. A. Waninge
Dr. E.A.A. Rameckers

Beoordelingscommissie

Prof. dr. A.F. Bos
Prof. dr. S. Beurskens
Prof. dr. A.C.H. Geurts

Paranimfen: Nastaran Salavati
Sahar Salavati

Acknowledgments

This Study was financially supported by:
- Novum Foundation, Huizen, The Netherlands
- Royal Dutch Visio, The Netherlands

Contents

List of abbreviations

CP	Cerebral Palsy
CVI	Cerebral Visual Impairment
OVI	Ocular Visual Impairment
GMFCS	Gross Motor Function Classification System
GMFM-88	Gross Motor Function measure-88
GMFM-88-CVI	Gross Motor Function measure-88 for children with Cerebral Visual Impairment
PEDI-NL	Paediatric Evaluation of Disability Inventory, Dutch version
PEDI-NL-CVI	Paediatric Evaluation of Disability Inventory, Dutch version for children with Cerebral Visual Impairment
WHO	World Health Organization
ICF-CY	The International Classification of Functioning, Disability and Health for Children and Youth
CVI-MQ	Cerebral Visual Impairment Motor Questionnaire

1

General introduction

GENERAL INTRODUCTION

Children with a brain lesion (Cerebral Palsy) have an increased risk of Cerebral Visual Impairment (CVI) due to brain damage. This introduction will describe the characteristics of these children related to the delay in gross motor functioning, functional skills, and caregiver assistance. In current clinical practice, it has been experienced that children with CP demonstrate limitations at this level of motor functioning that are more comprehensive than what could be caused from, e.g., just a motor limitation. In fact, this motor limitation cannot be explained from the results of the damage of the motor area in the brain or behavioural problems. Visual perception is important for motor functioning, therefore, a deficit in this area may impact motor control, motor learning, and motor development. Hence, the presence of CVI in children with CP cannot be adequately recognized, and the current assessment instruments used by paediatric physical therapists and occupational therapists in clinical practice also do not detect the presence of CVI in children with CP. As a result, children with CP and CVI could receive lower scores on assessments and a lower estimation of their motor capacity which could negatively affects proper treatment for these children. In addition, it is important to adapt the existing assessment tools for diagnostics and the evaluation of interventions that are already familiar to professionals in order to save valuable resources and facilitate future comparative studies.

The International Classification of Functioning, Disability, and Health for Children and Youth (ICF-CY) is a conceptual framework and uses a common language and terminology for recording problems involving functions and structures of the body, activity limitations and participation restrictions manifested in infancy, childhood and adolescence and relevant environmental factors. With its emphasis on functioning, the ICF-CY can be used across disciplines to define and document the health, functioning and development of children and youth.[1]

The first aim of this thesis is to establish whether and to what degree the level of gross motor function and functional skills in children with CP and CVI as well as caregiver assistance are different when compared to the corresponding matched group of children experiencing CP without CVI.

The most commonly employed assessment tools in clinical practice are the Pediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) and the Gross Motor Function Measure-88 (GMFM-88). Reliability of the PEDI-NL and the GMFM-88 in children with CP without CVI is sufficient, however, the validity and reliability study assessment did not include children with visual impairment. Thus, the second aim is to develop an adapted version of the PEDI-NL and GMFM-88 for children with CP and CVI and determine their reliability.

Paediatric physical therapists and occupational therapists are often the first professionals to assess the level of motor functioning and to treat children with CP. This puts them in a position to identify the warning signs of CVI when screening these children. This detection allows professionals to review the impact of CVI on the observed motor behaviour and to ensure the identification of signs and symptoms of CVI in children with CP. As there are insufficient adequate tools for screening for CVI in rehabilitation centres, it is important to develop a CVI motor screening tool to identify these signs. Therefore, the third aim is to develop CVI Motor Questionnaires (CVI-MQs) for children with CP and determine their validity and usability.

The International Classification of Functioning, Disability and Health for Children and Youth (ICF-CY)

The International Classification of Functioning, Disability, and Health for Children and Youth (ICF-CY) is derived from the International Classification of Functioning, Disability, and Health (ICF).[1,2] and is designed to record the characteristics of the developing child and the influence of its surrounding environment. According to ICF-CY, the impact of neurological diseases and visual impairment is evident for children with CP and CVI (Figure 1).[1]

In Figure 1, the assessment of gross motor function is related to activities while functional skills and caregiver assistance are related to daily activity which is a component of participation level. Since the assessment of children with CP and CVI is important, it is necessary to describe the activity and participation levels of those children. Indeed, Verrel et al.[3] showed in a recent study that the

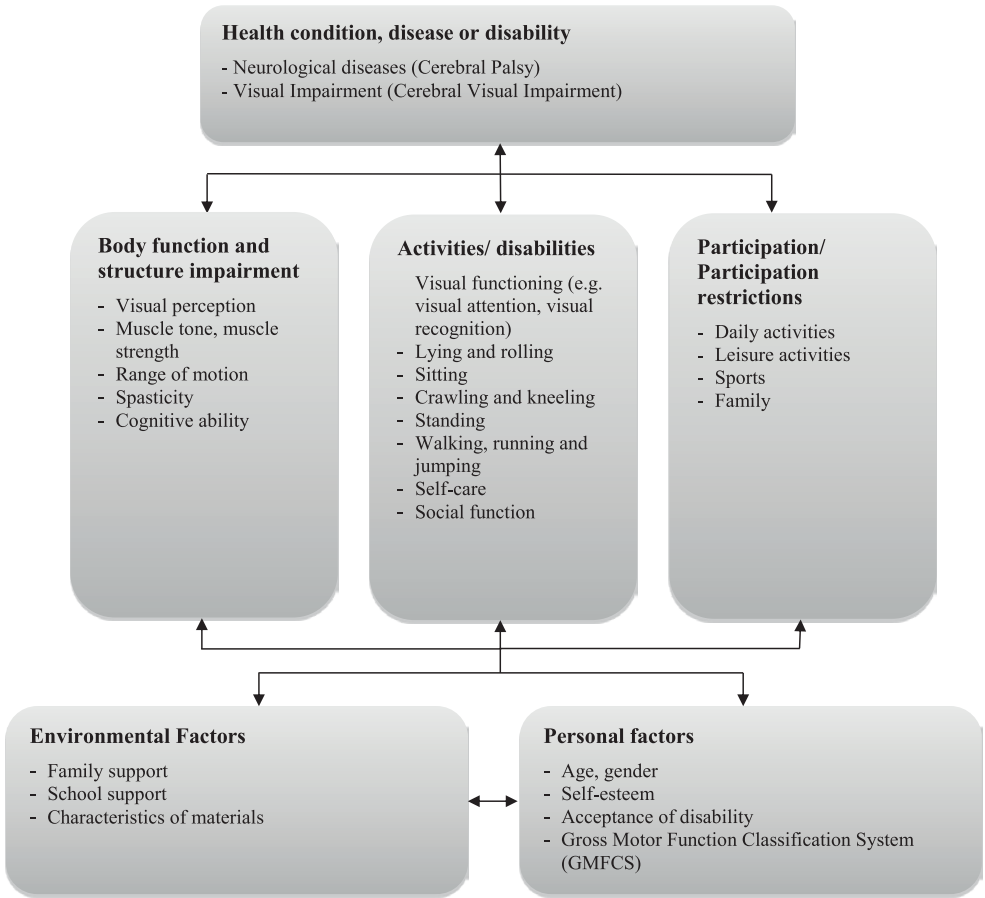


Figure 1. CF-CY model with CP and CVI components and their related activities.[1,2]

role of visual perception during motor skill is important because children with CP demonstrate increased visual perception during motor activities (walking, daily-life activities, and play), and this emphasizes the role of visual perception during motor action.[3,4,5]

Until now, there is insufficient evidence to suggest that children with CP and CVI score lower for functional skills, although there are indications that they experience increased limitations in gross motor function, functional skills, and caregiver assistance.[4,5] There are also no current valid and reliable measurement instruments to assess these limitations in children with CP and CVI, and no validated CVI motor assessment tools for screening children with CP are yet available. So far, the focus has been on screening visual dysfunction rather than motor skill abilities.

Cerebral Palsy in children

CP is a well-recognized neurodevelopmental condition that begins in early childhood, usually at less than two years of age and persisting through the lifespan. The prevalence of CP in Europe is 2-2.5 per 1000 live-birth children.[6,7,8,9] CP is a condition in which there is a motor disability caused by a static, non-progressive lesion in the brain. Other aspects of functioning are also frequently affected such as perception, vision, learning, and language, and it can cause epilepsy.[8]

A child with CP generally has one or more of the three types of neurological impairment of the motor system which are spasticity, dyskinesia, and ataxia. Spastic CP is the most common and accounts for approximately 80-90% of all cases, dyskinetic CP is experienced by 9% and ataxic CP by 2%. Spasticity is characterized by increased muscle tone which manifests as increased resistance to stretch that is

velocity dependent.[6] Dyskinesia refers to a category of movement disorders that are characterized by involuntary muscle movements. Ataxia is a movement disorder typified by uncoordinated movements and inadequate postural control that is evidenced with imbalance and walking disturbances.[6,7,8,9]

CP can be classified by severity whereby the Gross Motor Function Classification System (GMFCS) is very helpful as it indicates how much activity limitation the disorder imposes on the child with CP (Table 1).[10,11] Children at GMFCS Level 1 (mildest form) can walk and perform all of the activities of age-matched peers, albeit with limitations of speed, balance, and coordination. Children at Level 5 (most severe) must be transported, have extreme difficulties with trunk posture, and have little voluntary control of limb movement.[8,9]

Cerebral Palsy and Cerebral Visual Impairment in children

In this thesis, the focus will be on CVI which can be defined in terms of a neurological disorder in childhood caused by damage to or malfunctioning of the retrochiasmatic visual pathways (optic radiations, occipital cortex, associative visual areas) in the absence of any major ocular disease.[12,13,14,15] CVI is quite variable and has become a broad umbrella term that ranges from no light perception to normal visual acuity and, in the presence of cognitive visual dysfunction, a visual processing disorder that leads to misinterpretation of the visual world with respect to either what or where objects are.[16] There are several cortical areas involved in processing different perceptive visual functions. During the process of receiving visual information, this information is conveyed and analyzed in two separate ways to main areas, i.e., the occipito-temporal lobes and the occipito-parietal lobes. The visual pathway between the occipital lobes and temporal lobes is referred to as the "ventral steam"; it supports the process of visual recognition, orientation, and visual memory and is, therefore, sometimes called the "what" pathway. The visual pathway between the occipital lobes and the posterior parietal lobes is called the "dorsal stream" (sometimes called the "where" pathway). It includes visual spatial perception, motion perception, and simultaneous perception which can be associated with crowding.[17] CVI ranges in severity from blindness to relatively minor impairments of visual perception. Perceptual visual dysfunction and disorders of visual attention are often only minimally reduced or have normal visual acuities and are increasingly recognized as forms of CVI.[13,18,19] CVI frequently co-occurs with CP and is observed in approximately 30% of children diagnosed with various forms of CP.[4,20,21] The spectrum of visual impairments in children with CP is extremely broad and includes both Ocular Visual Impairment (OVI) such as strabismus, reduced visual acuity, ocular nystagmus, refraction disorders, and retinopathies and CVI which is a problem of central origin. CVI is among the major causes of visual impairment in children which affects early cognitive, motor, and social development.[12,18,22,23,24,25,26]

Children with CVI exhibit slow, inefficient, and highly variable visual functioning during daily-life.[13,19,4,5,27] CVI can influence the child's ability to learn and perform tasks in everyday life and is,

Table 1. Gross Motor Function Classification System (GMFCS) for Children with CP ages 6-12.[10,11]

Level	GMFCS
I	Children walk indoors and outdoors, and climb stairs without limitations. Children perform gross motor skills including running and jumping but speed, balance, and coordination are reduced
II	Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces. Children have, at best, only minimal ability to perform gross motor skills such as running and jumping
III	Children walk indoors or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a railing. Depending on upper limb function, children propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain
IV	Children may maintain levels of function achieved before age 6 or rely more on wheeled mobility at home, school, and in the community. Children may achieve self-mobility using a power wheelchair
V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. At level V, children have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations

therefore, warranted to be taken into account in therapy.[28] Considerable focus has been directed towards the detection and treatment of prematurity-caused retinopathy, however, there tends to be less of a focus on CVI which can, consequently, be overlooked. Dutton and Jacobson[13], who are focusing primarily on profound visual impairment in children experiencing CVI, state that it has an impact on all aspects of a child's development.[13] They conclude that children with CP and CVI develop more slowly in the area of self-care, mobility, and social functioning than children with CP but without CVI. Also, the presence of CVI could result in a lack of the child being able to locate its caregivers and difficulty in knowing whether the caregivers are present or absent which thereby affects the level of a child's motivation to acknowledge them.

Impaired vision as a result of CVI is evident, however, marked visual impairment may go undetected because the resulting behaviour displayed by a child is not recognized or may be marked as a behavioural impairment.[17] Lack of recognition can be problematic for a child with CVI whose inaccurate visual guidance of movement, for example, may be misinterpreted as clumsiness.[17] Meanwhile, the child may be doing its best but is continually criticized. The outcome can be disheartening for a child, leading to low self-esteem and a sense of being misunderstood. Therefore, recognizing and understanding the capabilities of children affected by CVI is essential to ensure that interventions and educational endeavors are accessible, efficient, and successful.[17] Children with CVI may not even be aware that their vision is limited. If they have always seen the world a certain way, they have no way of comparing how they see with how others see.[17] It appears that, during the support of children with CVI, they could benefit from verbal support/ instruction (e.g., what especially should be said in order to help a child accomplish a particular skill) and manual support (e.g., duration and phase of required manual support). Furthermore, those children could also benefit from adapted equipment (e.g., colourful, sound-produced, high in contrast) to receive their attention.

Ghasia et al.[4] and Da Costa et al.[5] described a relationship between both the presence of CVI and motor function impairment in children with CP and concluded that the presence of CVI in a group of children with CP and GMFCS Levels III-IV-V is higher than in children with CP and GMFCS Levels I-II.[4,5] Lueck and Dutton[17] emphasized that a child with fewer physical disabilities or other major conditions (GMFCS levels I-II) has an increased chance of undetected CVI and having unusual behaviour or motor impairment being interpreted as mental or motor impairment rather than CVI.[17] For example, the child can read large, well-spaced text and recognize people yet has significant difficulty reading smaller text, finding objects, copying text or pictures, moving through a space, or accurately reaching for objects. The associated peripheral lower visual field impairment can also lead to tripping, fear of jumping into a swimming pool, and refusal to jump off a bench. These reactions can be mistakenly interpreted as the child being clumsy and anxious. For example, if peripheral lower visual field impairment is not identified, paediatric physical therapists, who are focused on the need to maintain an erect posture, may require the child not to look down while walking. However, intermittent viewing of the ground ahead facilitates safe mobility and should be encouraged and integrated into the child's overall program.[17]

Measurement instruments

Paediatric physical therapists and occupational therapists have different measurement instruments at their disposal with various constructs which can be utilized to evaluate the outcome measure at the levels of ICF-CY such as body function, activity, or participation. For these professionals, it is important to determine whether and to what degree the presence of CVI affects the outcome extent of measurement instruments which are commonly used in clinical practice. Also, the need to use functional outcome measures is obvious as functional measures are consistent with treatment goals which emphasize independence in children with CP and CVI. Paediatric physical therapists and occupational therapists assess clinical changes in gross motor function, self-care, mobility, and social functioning of children with CP using tests such as Canadian Occupational Performance Measure (COPM), Goal Attainment Scaling (GAS), the Paediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL)[29,30,31] and Gross Motor Function Measure-88 (GMFM-88).[31,32] These assessment instruments attempt to detect all domains of motor development. Of all of these tests, the most

commonly used tests for children with CP are the PEDI-NL and the GMFM-88 due to their good reliability.[32,33] Reliability refers to the extent in which a measure is reproducible, and validity refers to the extent in which a measure is accurately assessing what it needs to assess. These assessment instruments are considered as a "gold standard" by professionals. Due to the fact that professionals are most familiar with the PEDI-NL and the GMFM-88 assessment instruments, it is obvious to adapt these instruments for children with CP and CVI. Furthermore, it is easy and time-saving to implement the adapted version of those instruments in the rehabilitation centres rather than develop a new assessment instrument for children with CP and CVI. It is stated that the content of the original PEDI-NL and GMFM-88 are developed for children with CP without ocular visual impairment (OVI) or CVI. [31,32] In conclusion, adequate visual perception is essential for the execution of most of the instruments. Additionally, professionals working with children with CP and CVI at expertise centres for blind and visually impaired individuals are of the opinion that these instruments are not appropriate for children with CVI.

The PEDI-NL is a questionnaire that evaluates the daily skills of children aged six months to approximately seven and a half years of age. It measures both capability (what the child *can* do) and performance (what the child *actually does*) of daily routine childhood activities. A structured interview with the parents or caregivers is utilized to evaluate the self-care, mobility and social function domains.[29,30,31] A child's capability can be measured by using the PEDI-NL's three functional skills scales. Performance can be determined by utilizing its three caregiver assistance scales and modification scales. According to the ICF-CY, the PEDI-NL measures both the child's capacity and the performance of essential daily activities. Capacity is measured by identifying the daily activities that the child has performed independently. Performance is measured by assessing the level of assistance needed to accomplish the daily activity that the caregiver has given to the child. This results in a complex interaction between all of the components of the ICF-CY. Reliability of the PEDI-NL in children with CP without CVI is sufficient[29], however, the validity and reliability study assessment did not include children with visual impairments.[29] The presence of CVI in children with CP results in a major challenge for paediatric physical therapists and occupational therapists when assessing and treating these children by needing to focus on both the activity and participation components of the ICF-CY.

The Gross Motor Function Measure-88 (GMFM-88) is a performance-based measure used to ascertain changes in the gross motor function of children with CP and has been commonly used by researchers.[33, 34] The GMFM-88 consists of 88 items in five dimensions: lying and rolling (GMFM-A); sitting (GMFM-B); crawling and kneeling (GMFM-C); standing (GMFM-D); and walking, running and jumping (GMFM-E). There is a 4-point scoring system for each item on the GMFM-88. The scoring is as follows: 0= the child does not initiate task; 1= child initiate task (<10%); 2= child partially completes task (10-99%); 3= child completes task (100%); NT= Not tested. The test has beneficial clinical application in that it is designed to assess gradual motor function changes or changes with intervention in children with CP. According to ICF-CY, GMFM-88 measures motor functioning at the activity level. The test has normative data, is predictive, valid, and reliable. The GMFM-88 provides information of the level of difficulty of each item which can assist the therapist in establishing realistic goals. The test is also accepted internationally. It is important to note that reliability and validity for children with CP and visual impairments is not yet known.[33,34]

This thesis focuses on children from the ages of four to 12 years who are experiencing CP and CVI and have a mild or moderate intellectual disability. With various levels of the Gross Motor Function Classification system (GMFCS), the possible differences between a group of children with CP and their peers with CP and CVI were initially investigated at the level of gross motor function, functional skills, and caregiver assistance. Second, the measurement instruments Pediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) and Gross Motor Function Measure-88 (GMFM-88) for children with CP and CVI was adapted. The objectives of both studies were to achieve consensus among a group of experts. Subsequently, the reliability of both the adapted PEDI-NL and GMFM-88 in children with CP and CVI were determined. As a result, CVI-supplements for each measurement to assess children with CP and CVI were

developed. Third, the adapted and original GMFM-88 in the same group of children with CP and CVI were compared. Fourth, and final, two CVI Motor Questionnaires (CVI-MQs) for children with CP were developed and validated, one CVI-MQ for children with CP with GMFCS I- II- III and one for children with GMFCS IV-V.

As a substantial number of children with CP also suffer from CVI, it is very important that paediatric physical and occupational therapists have a valid instrument at their disposal which measures the quantitative parameters of motor development.

Thesis outline

In Chapter 2, the differences of the level of performance in gross motor function, functional skills, and caregiver assistance in a group of children with spastic CP with CVI compared with a matched group of children with spastic CP and without CVI are described.

In Chapter 3, the adaptation of the Pediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) for children with CVI and CP is described, and its test–retest and inter-respondent reliability are determined.

In Chapter 4, the adaptation of the Gross Motor Function Measure-88 (GMFM-88) for children with both spastic CP and CVI is described, and its test–retest and interobserver reliability are determined.

In Chapter 5, a comparison between outcomes of the original and cerebral visual impairment adapted GMFM-88-CVI in children with spastic CP and CVI is described. The aim was to determine whether the adapted GMFM-88 for children with CP and CVI provides a better estimate of gross motor function per se in children with CP and CVI that is not adversely affected by their visual problems.

In **Chapter 6**, the development of two Cerebral Visual Impairment Motor Questionnaires (CVI-MQs) for children with Cerebral Palsy (CP) is described: one for children with Gross Motor Function Classification (GMFCS) levels I-II-III and one for children with GMFCS levels IV-V. Thereby their validity, usability, sensitivity, and specificity are determined.

Chapter 7 consists of a general discussion, the implications, and the research findings for occupational therapists, paediatric physical therapists, and other health care practitioners.

References

1. International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation). Dutch WHO-FIC Collaborating Centre. 1st ed.
2. World Health Organization (WHO). International Classification of Functioning, Disability and Health. Geneva: WHO, 2001. Houten: Bohn Stafleu van Loghum (www.bsl.nl) 2008.
3. Verrel J, Bekkering H & Steenbergen B. Eye– hand coordination during manual object transport with the affected and less affected hand in adolescents with hemiparetic cerebral palsy. *Experimental Brain Research* 2008*;* 187: 107–161.
4. Ghasia F, Brunstrom J, Gordon M & Tychsen L. Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: gross motor function scale. *Invest Ophthalmol Vis Sci* 2008; 49: 572-80.
5. Da Costa M, Salmao S, Berezovsky A, De Haro F & Ventura D. Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. *Behav Brain Res* 2004; 149: 145-50.
6. Surveillance of Cerebral Palsy in Europe (SCPE). Prevalence and characteristics of children with cerebral palsy in Europe. *Dev Med Child Neurol* 2002; 44(9): 6633-40.
7. Odding E, Roebroek M & Stam H. The epidemiology of cerebral palsy: incidence, impairment and risk factors. *Disabil Rehabil* 2006; 28(4): 183-91.
8. Rosenbaum P, Paneth N, Leviton A, Goldstein M & Bax M. Definition and classification of cerebral palsy. *Dev Med Child Neurol* 2007; 49: 480.
9. Bax M, Flodmark O & Tydeman C. Definition and classification of cerebral palsy. From syndrome toward disease. *Dev Med Child Neurol Supplement* 2007; 109: 39-41.
10. Palisano R, Rosenbaum P, Walter S, Russell D, Wood E & Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. *Dev Med Child Neurol* 1997; 89: 214–223.
11. Paneth N. Establishing the Diagnosis of Cerebral Palsy; *Clinical obstetrics and gynecology* 2008; 51(4): 742–748.
12. Fazzi E, Signorini SG, Bova SM, Ondei P & Bianchi PE. Early intervention in visually impaired children. International Congress Series 2005; 1282: 117–121.
13. Dutton G & Jacobson L. Cerebral visual impairment in children. *Semin Neonatol* 2001; 6: 477-85.
14. Dutton GN, Saaed A, Fahad B, Fraser R, McDaid G, McDade J, et al. Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction-a retrospective observational study. *Eye* 2004*;* 18: 27–34.
15. Expertisegroep CVI kinderen en jongeren. Visie op CVI. Koninklijke Visio. Huizen; 2013.
16. Edmond, JC & Foroozan, R. Cortical visual impairment in children. *Current Opinion in Ophthalmol 2007;* 17: 509-512.
17. Lueck AH & Dutton GN. *Vision and the Brain: Understanding Cerebral Visual Impairment in Children* 2015; ISBN: 9780891286394.
18. Fazzi E, Molinaro A & Hartmann E. The potential impact of visual impairment and CVI on child development. *Vision and the brain* 2012; ISBN: VATBCh04.

19. Dutton GN. The spectrum of cerebral visual impairment as a sequel to premature birth: an overview. *Doc Ophthalmol* 2013; 127(1): 69-78.
20. Stiers P, Vanderkelen R, Vanneste G, Coene S, De Rammelsere M & Vandenbussche E. Visual-perceptual impairment in a random sample of children with cerebral palsy. *Dev Med Child Neurol* 2002*;* 44: 370-382.
21. Huo R, Burden SK, Hoyt CS & Good WV. Chronic cortical visual impairment in children: aetiology, prognosis, and associated neurological deficits. *Br J Ophtamalo* 1999; 83: 670-5.
22. Hatton DD, Schwietz E, Boyer B & Rychwalski P. Babies Count: The national registry for children with visual impairments, birth to 3 years. *J AAPOS* 2007; 11: 351–355.
23. Rahi JS & Cable N. British Childhood Visual Impairment Study G. Severe Visual impairment and blindness in children in UK. *Lancet* 2003; 362: 1359-65.
24. Rosenberg T, Flage T, Hansen E, Riise R, Rudanko SL, Viggosson G, et al. Incidence of registered visual impairment in the Nordic child population. *Br J Ophthalmol* 1996; 80: 49-53.
25. Boonstra N, Limburg H, Tijmes N, van Genderen M, Schuil J & van Nispen R. Changes in casus of low vision between 1988 and 2009 in a Dutch population of children. *Acta Ophthalmol* (Copen) 2012; 90: 277-86.
26. Geldof CJA, van Wassenaer AG, de Kieviet JF, Kok JH & Oosterlaan J. Visual perception and visual-motor integration in very preterm and/or very low birth weigth children: A meta-analysis. *Res Dev Disabil* 2012; 33: 726-36.
27. Good W, Jan F, Burden S, Skoczenski A & Candy R. Recent advances in cortical visual impairment. *Dev Med Child Neurol* 2001; 43: 56-60.
28. Himmelmann K, Beckung E, Hagberg G & Uvebrant P. Gross and fine motor function and accompanying impairments in cerebral palsy. *Dev Med Child Neurol* 2006; 48: 417–423.
29. Wassenberg-Severijnen JE,Custers JW, Hox JJ,Vermeer A & Helders PJ. Reliability of the Dutch Pediatric Evaluation of Disability Inventory (PEDI). *Clinical Rehabilitation* 2003; 17: 457–462.
30. Wassenberg-Severijnen JE & Custers JWH. Pediatric evaluation of Disability Inventory-NL. Amsterdam, The Netherlands: Harcourt Assessment BV 2005.
31. Custers JWH,Wassenberg-Severijnen JE, Net Jvd, Vermeer A, Hart HT & Helders PJM. Dutch adaptation and content validity of the 'Pediatric Evaluation of Disability Inventory (PEDI)'. *Disability and Rehabilitation* 2002; 24: 250–258.
32. Russell DJ, Rosenbaum PL, Cadman DT, et al. The Gross Motor Function Measure: a means to evaluate the effects of physical therapy. *Dev Med Child Neurol* 1989; 31: 341–352.
33. Ketelaar M, Van Petegem-van Beek E, Veenhof C, Visser J & Vermeer A. Gross motor function measure. University of Utrecht. *Child Physical Therapy* 2003; 39: 5–7.
34. Engelen V, Ketelaar M & Gorter JW. Electing the appropriate outcome in paediatric physical therapy: How individual treatment goals for children with cerebral palsy are reflected in GMFM-88 and PEDI. *Journal of Rehabilitation Medicine* 2007; 39: 225–231.

2 |

Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI)

Masoud Salavati [1] , Eugene A.A. Rameckers [2,3,4] , Bert Steenbergen [5,6] & Cees van der Schans [7,8]

1 Royal Visio, Center of Expertise for Blind and Partially Sighted People, Haren, The Netherlands
2 Maastricht University, Department of Rehabilitation Medicine, School for Public Health and Primary Care (CAPHRI), Maastricht, The Netherlands
3 Adelante Center of Expertise in Rehabilitation and Audiology, Valkenburg and Hoensbroek, The Netherlands
4 AVANSplus, University for Professionals for Pediatric Physical Therapy, Breda, The Netherlands
5 Radboud University Nijmegen, Behavioural Science Institute, Nijmegen, The Netherlands
6 Australian Catholic University, School of Psychology, Melbourne, Australia
7 Department of Rehabilitation Medicine, University Medical Center Groningen, Groningen, The Netherlands
8 Research Group in Health Care and in Nursing, Hanze University Applied Sciences Groningen, Groningen, The Netherlands

Published as:
Salavati, M., Rameckers, E.A.A., Steenbergen, B. & Schans van der, C.P. (2014). Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). European Journal of Physical Therapy, 16(3), 159–167.

Abstract

Aim: To determine whether the level of gross motor function and functional skills in children with cerebral palsy (CP) and cerebral visual impairment (CVI) as well as caregiver assistance are lower in comparison with the corresponding group of children experiencing CP without CVI.

Method: Data aggregated from 23 children experiencing CP with CVI were compared with data from children with CP without CVI matched for Gross Motor Function Classification System, mental development and age at testing. Scores for Gross Motor Function Measure-88 (GMFM-88) and the Pediatric Evaluation of Disability Inventory-NL (PEDI-NL) were employed to compare the level of gross motor function, functional skills and caregiver assistance between both groups. The Wilcoxon Signed Rank Test was utilized with a significance level of $p < 0.05$.

Results: Children with CP with CVI, mean (± SD) age 6.4 ± 1.5, scored significantly lower than those with CP without CVI, mean age 6.3 ± 1.6, on all GMFM-88 dimensions and the total score ($p < 0.001$) and on the PEDI-NL in the sections of Functional Skills and Caregiver Assistance as well as in those of domains self-care ($p < 0.001$), mobility ($p < 0.001$) and social functioning ($p < 0.001$). Concerning the modifications scale, the scores for children with CP and CVI were significantly lower regarding mobility (no modification, $p < 0.05$), social functioning (no modification, $p < 0.05$) and social functioning (child-oriented, $p < 0.05$).

Conclusion: CVI contributes to diminished gross motor function and functional skills in children experiencing CP with CVI compared with children with CP without CVI. Children with CP and CVI also require increased support at the level of caregiver assistance. Specific interventions need to be developed for children experiencing CP with CVI in order to improve gross motor function, functional skills and caregiver assistance.

Key words: *Neurology, occupational health/ergonomics, pediatrics, population studies*

Introduction

Cerebral palsy (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, which is attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication and behavior, by epilepsy, and by secondary musculoskeletal problems (1,2). Visual disorders including cerebral visual impairment (CVI) are regularly observed in an elevated number (30–100%) of children diagnosed with the various forms of CP (3–9).

CVI can be defined as deficient visual function as a sequel of damage or malformation of the retro-geniculate visual pathways (optic radiations, occipital cortex and visual association areas) and may include deficits in central oculomotor control. CVI is a prominent sequel to premature birth, particularly when the prematurity is extreme. Considerable focus has been directed toward the detection and treatment of retinopathy of prematurity, but less attention tends to be focused on CVI, which can, as a consequence, be overlooked. Dutton et al. (9) focused primarily on profound visual impairment in children experiencing CVI, but CVI ranges in severity from blindness to relatively minor impairments of vision and perception. Perceptual visual dysfunction, reduction in visual fields and disorders of visual attention, often with only minimally reduced or normal visual acuities, are increasingly being recognized as forms of CVI as a sequel of prematurity (9–12).

Children experiencing CVI could incur difficulty at different levels such as: underdeveloped stair or curb climbing, often accompanied by the need to touch the surface with the hand; underdeveloped reaching and knocking over objects; impaired simultaneous perception manifested by an inability to locate an object in a crowded visual field such as toys in a toy box, or a parent in a crowd or difficulty reading; and difficulty seeing moving targets. Underdeveloped movements of the arms and especially the legs are compounded by bilateral inferior visual field defects and any co-existent motor deficits. They could also have problems with route finding in unfamiliar places; forgetting where objects were located; and difficulty recognizing faces, shapes or objects (12–14). More severe visual disorders have been discovered in children with spastic CP and serious motor skill limitations (5,6). However, there is no linear relationship between the presence of visual problems and the compromises in the activity level of a child (15).

Children with CP show increased visual monitoring when performing actions with the affected hand, both at the beginning and during an object transport, and this emphasizes the role of visual perception during motor action (16). CVI in children with CP might be a secondary deficit due to an impoverished environment caused by the motor limitations of the pathology that affects them. These children are significantly disabled in their physical activity, which might reduce their ability to explore their world. In addition, they often have cognitive and attention deficits associated with the motor impairment, which further reduces their experiences in general (sensory, motor, learning and memory) (5). Several studies have reported that CVI plays an essential role in motor, cognitive and emotional development (5,7,8,13). In particular, the influence that severe visual disabilities can have on motor behavior is relevant and complex, and secondary to the impairment of various areas of development. The inability to achieve normal adaptive control of posture is strongly related to maintaining dependence on sensation, mainly vision. Children with CP with CVI have specific problems with mapping between vision and proprioception (17).

Children with visual impairment have an inferior gross motor skill performance and are less physically active than their peers without visual impairment and exhibit poor performance on static and slow dynamic balance tasks (18). They have difficulty in achieving a high level of involvement in physical activity, and the development of independent walking might be more challenging for children with a visual impairment than for their normally sighted peers (18). The presence of visual impairments such as CVI may be associated with a higher Gross Motor Function Classification System (GMFCS) score (5,8). It is suggested that children with CP and CVI develop more slowly in the area of self-care when compared with children experiencing CP and without CVI (7). Although children with CP and CVI are limited in their physical activities, it is not clear to what extent CVI contributes to these limitations. Until now, no specific interventions have been implemented

for children with CP and CVI, and there is insufficient evidence whether CP with CVI children score lower on functional skills, though there are indications that children with CP and CVI experience increased limitations in gross motor function, functional skills and caregiver assistance (5,8).

Our hypothesis is that the level of functional skills and caregiver assistance in a group of children with spastic CP with CVI is lower compared with a matched group of children with spastic CP and without CVI. We also expect that the group of children experiencing CP with CVI exhibit a more inadequate performance in gross motor function as laying, rolling, sitting, crawling, walking, running and jumping.

Methods

Children enduring spastic CP with or without CVI were recruited from Royal Visio (Center of Expertise for Blind and Partially Sighted People, The Netherlands) and from primary care allied health practices. Inclusion criteria were the presence of spastic CP and age at testing (GMFM-88 (19) and the PEDI-NL (20,21) between 4 and 8 years.

Exclusion criteria were the presence of syndromes (e.g. Down syndrome) in combination with CP and hearing difficulties. Children with a (corrected) vision < 0.3 and/or a field of vision < 30° and retinopathy of prematurity were also excluded.

The study was approved by the Medical Ethical Committee (METc-2010-137) of the University Medical Center Groningen (UMCG), Groningen, and The Netherlands. Written informed consent was obtained from the children's parents.

The diagnosis of CP and the classification according to the GMFCS level were aggregated from the children's medical files and judged by a specialized child physiotherapist. The study group consisted of children who had been diagnosed with spastic CP as was cited in the medical and ophthalmological files. The diagnosis of CVI was determined based on the results of ophthalmological, (neuro-) psychological research and on the assessment data reported by a developmental coach specialized in working with children with visual impairments.

Employing a retrospective file search of data collected between March 2007 and December 2010, we were able to aggregate the data of 77 children. Based on the inclusion criteria, data of 23 children with CP and CVI (n = 11 boys) and 23 children with CP and without CVI (n = 12 boys) were analyzed (Table I). The children were matched according to their GMFCS level and the type of CP (uni- or bilateral), their mental development according to the Resing & Blok (22) method, and the age at which the GMFM-88 (19) and PEDI-NL (20,21) were administered. The tests were administered by therapists, and we exploited the raw scores of GMFM-88 (19) and PEDI-NL (20,21).

Based on the possible effect on gross motor function, functional skills and caregiver assistance, we also collected data regarding gender as well as the prevalence of epilepsy and speech/language development according to the International Classification of Functioning, Disability and Health for Children and Youth (23).

At the level of speech/language development, the collected data were: d330 = speaks; d3350 = uses body language; d3351 = uses signs and symbols; d3100 = reacts to human voice; d3101 = understands simple spoken messages; and d331 = babbles.

Test instruments

The GMFM-88 (19) (used for measuring gross motor function) and the PEDI-NL (20,21) (used for measuring functional skills and caregiver assistance) were both recommended in the CP guidelines of the Dutch Society of Rehabilitation Physicians (2) and, therefore, were employed in this study.

GMFM-88 is designed to evaluate change in gross motor function over time or with intervention for children with CP (24,25). The 88 in the name of the test refers to the 88 items that are investigated. They relate to five dimensions: laying and rolling (GMFM-A); sitting (GMFM-B); crawling and kneeling (GMFM-C); standing (GMFM-D); and walking, running and jumping (GMFM-E). The reliability and validity of this test are sufficient (19,24,25). The severity of the impairment in gross motor skills was classified in accordance with the GMFCS into five different levels where Level I indicates the least functionally hindered, and Level V is the most functionally hindered (26).

The PEDI-NL (20,21,27) is a questionnaire that evaluates the daily skills of children aged 6 months to 7.5 years. The PEDI-NL is suited to measure both capability (what the child can do) and performance

(what the child actually does) of daily routine childhood activities in the self-care, mobility and social function domain (20,21,27). The capability of a child can be measured utilizing the three functional skills scales of the PEDI-NL. The performance of a child can be measured using the three caregiver assistance scales and modification scales of the PEDI-NL. The modification scales are measurements of environmental modifications and equipment used by the child in daily routine activities (20,21,27). The reliability of PEDI-NL is sufficient for all three scales on the three domains: self-care, mobility and social function domain (21). The PEDI-NL was developed as a discriminative and evaluative measuring instrument and is capable of recording relevant changes during a 6-month period in children with CP (20,21,27).

Statistical analyses

The partial and total scores of the GMFM-88 and the PEDI-NL were calculated and compared in order to discover (possible) differences between the children experiencing CP with CVI and those with CP without CVI. Since the distribution of the differences deviated extensively from the normal distribution, the Related Samples Wilcoxon Signed Rank Test with a significance level of $p < 0.05$ was utilized to detect possible significant differences between both groups.

Results

Table I exhibits the characteristics of the included children. The two groups were matched on GMFCS levels and mental development according to the Resing & Blok (22) method, and the age at which the GMFM-88 and PEDI-NL were administered. The children could be classified into four of the five GMFCS levels (Table I). The mean (± SD) age of the children was 6.3 ± 1.6 years for the children experiencing CP without CVI and 6.4 ± 1.5 years for those with CP with CVI. The difference in age at testing was 0–7 months. No difference was apparent between 14 pairs concerning epilepsy, but nine pairs displayed a difference in the presence of epilepsy.

At the level of speech/language development, significant differences were evident between the two groups regarding d330 = speaks, d3350 = uses body

Table I. Differences in characteristics between children with cerebral palsy (CP) without cerebral visual impairment (CVI) and those with CP with CVI

Characteristic	Children with CP without CVI	Children with CP with CVI
Age at testing, mean (SD) (years)	6.3 (1.6)	6.4 (1.5)
Gender (F /M)	12 / 11	11 / 12
GMFCS I (n)	8	8
GMFCS III (n)	6	6
GMFCS IV (n)	4	4
GMFCS V (n)	5	5
Speech/language development ICF-CY, d3100 = reacts to human voice (n)	1	1
Speech/language development ICF-CY, d3101 = understands simple spoken messages (n)	22	22
Speech/language development ICF-CY, d330 = speaks (n)	16	13
Speech/language development ICF-CY, d331 = babbles (n)	2	2
Speech/language development ICF-CY, d3350 = uses body language (n)	4	8
Speech/language development ICF-CY, d3351 = uses signs and symbols (n)	1	0

F, female; M, male; GMFCS, Gross Motor Function Classification System; *n*, numbers; ICF-CY, International Classification of Functioning, Disability and Health, Child and Youth version (Dutch translation).

language and d3351 = uses signs and symbols. No differences were evident between the two on d3100 = reacts to human voice, d3101 = understands simple spoken messages and d331 = babbles (Table I).

GMFM-88

The children with CP and CVI scored significantly lower in all dimensions of the GMFM-88 compared with the children experiencing CP and without CVI. As otherwise stated, there was a significant difference in all gross motor functioning between the two groups. GMFM-88 dimensions: A (laying and rolling, $p < 0.001$); B (sitting, $p < 0.001$); C (crawling and kneeling, $p < 0.001$); D (standing, $p = 0.009$); E (walking, running, jumping, $p = 0.002$); Total (A + B + C + D + E, $p < 0.001$) (Figure 1 and Table IV).

Table II demonstrates that in only one matched pair was the child with CP and without CVI slower to crawl and kneel (GMFM-C) than the child with CP and with CVI.

PEDI-NL

The children experiencing CP with CVI scored significantly lower on the PEDI-NL in the sections on Functional Skills and Caregiver Assistance and in the domains of self-care ($p < 0.001$), mobility ($p < 0.001$) and social functioning ($p < 0.001$). Concerning the modifications scale, the scores for children with CP with CVI were significantly lower for mobility (no modification, $p < 0.05$), social functioning (no modification, $p < 0.05$) and social functioning (child-oriented, $p < 0.05$) (Figure 2, Tables III and IV).

Table II. Gross Motor Function Measure-88 (GMFM-88): the difference in raw score per pair between (a) children with CP without CVI and (b) those with CP with CVI.

Pairs	GMFM-A (laying down and rolling)	GMFM-B (sitting)	GMFM-C (crawling and kneeling)	GMFM-D (standing)	GMFM-E (walking, running, and jumping)	GMFM-Total
1a-1b	0	3	12	74.4	48.4	31.6
2a-2b	0	0	7	31	20	11.6
3a-3b	0	3	5	41	47	21
4a-4b	0	0	-5	10	8	1.5
5a-5b	0	3	26	15	0	3.4
6a-6b	0	6	0	0	4	2
7a-7b	6	8	19	7	3	8.6
8a-8b	14	10	31	12	38	21
9a-9b	6	30	53	18	10	23.6
10a-10b	4	25	50	38	8.5	25.5
11a-11b	2	16	44	44.5	20.5	25
12a-12b	0	0	9	6.5	3.5	4
13a-13b	64	42	31	0	0	27.5
14a-14b	6	12	26	25.5	9	15.4
15a-15b	37	62	0	0	0	15.3
16a-16b	22	83	98	8	0	42
17a-17b	15	6	9	7.5	0	7
18a-18b	51	25	64	15	0	31
19a-19b	60	6.5	0	0	0	13
20a-20b	1	6	5	0	0	2.6
21a-21b	6	7	4.6	0	0	2.7
22a-22b	29	36.6	14.2	0	0	14.8
23a-23b	6	0.5	0	0	0	0.9

A negative value indicates that a child with CP without CVI received a lower score than the child with CP and CVI.

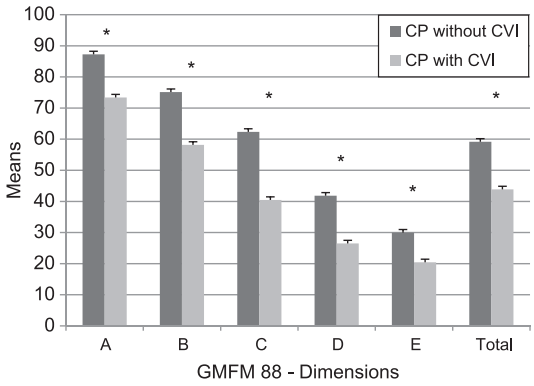


Figure 1. The Gross Motor Function Measure-88 (GMFM-88). X-axis: GMFM-88 dimensions; Y-axis: GMFM-88 mean and SD of raw scores of gross motor function (GMFM-88 SD of raw score CP without CVI 21; GMFM-88 SD of raw score CP and CVI 19). *Significant difference. GMFM-88 dimensions: A (laying and rolling, p< 0.001); B (sitting, p<0.001); C (crawling and kneeling, p<0.001); D (standing, p=0.009); E (walking, running, jumping, p=0.002); Total (A+B+C+D+E, p<0.001).

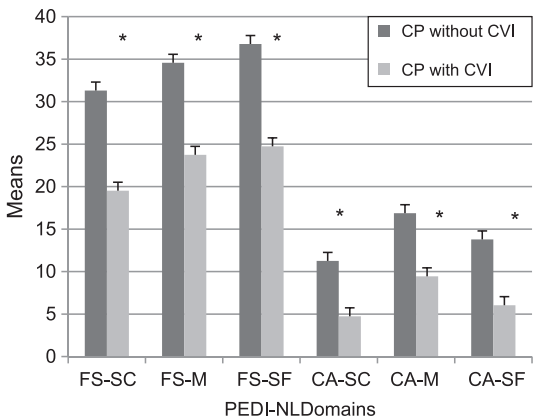


Figure 2. The Paediatric Evaluation of Disability Inventory-NL (PEDI-NL, Dutch version). X-axis = PEDI-NL domains; Y-axis = PEDI-NL raw scores mean and SD of raw scores are presented. (PEDI-NL SD of raw score CP without CVI 11; PEDI-NL SD of raw score CP and CVI 9). *Significant difference. FS-SC (Functional Skills-Self-Care, $p < 0.001$); SF-M (Functional Skills-Mobility, $p < 0.001$); FS-SF (Functional Skills- Social Functioning, $p < 0.001$); CA-SC (Caregiver Assistance-Self-Care, $p < 0.001$); CA-M (Caregiver Assistance-Mobility, $p < 0.001$); CA-SF (Caregiver Assistance-Social Functioning, $p < 0.001$).

At the level of modifications scale, there were no significant differences between the two groups except in mobility (no modification, $p < 0.05$), social functioning (no modification, $p < 0.05$) and social functioning (child-oriented, $p < 0.05$).

Table III indicates that, in five pairs of children, the child with CP and without CVI experiences a developmental delay compared with the child with CP with CVI. Such a delay was discerned in the section on Functional Skills (domain self-care, three pairs; mobility, one pair; social functioning, one pair) and Caregiver Assistance (domain self-care, three pairs).

Discussion

The first aim of this current study was to investigate whether the level of functional skills and caregiver assistance in a group of children with one type (spastic) of CP and CVI were lower compared with that of a matched group of children with spastic CP and without CVI. The results of our study indicated that children with CP with CVI scored lower on the PEDI scale in functional skills and caregiver assistance than children with CP without CVI. Our study demonstrated that children with CP with CVI obtained a lower score for self-care, mobility and social functioning. This could account for the limitations observed in daily activities and the slow processing and performance speed of children with CP with CVI.

This present study demonstrated that, based on a comparison of PEDI-NL scores, self-care, mobility and social functioning in the group of children with CP with CVI are significantly more affected than in the group with CP and without CVI and that this difference can be explained by the presence of CVI.

Our study demonstrated that children with CP with CVI clearly achieved lower scores in all dimensions of gross motor function including laying and rolling, sitting, crawling and kneeling, standing, walking, running and jumping (GMFM-A, -B, -C, -D and -E) in the GMFM-88 when compared with children with CP without CVI. For example, children with CP with CVI scored lower in the dimension of crawling and kneeling (GMFM). The crawling and kneeling stage is one of the most important phases in a child's motor skill development – it is when a child begins to move from one place to another. Reduced visual information probably has significant influence on targeted movement of a child with CP and CVI (18). Lack of visual information processing could result in limited crawling and kneeling (GMFM-C).

Our study is the first study that compared a group of children with spastic CP with CVI with a matched group of children with spastic CP without CVI by exploiting the data of GMFM-88 and PEDI-NL. We

also compared matched groups at different domains according to their GMFCS level and the type of CP (uni- or bilateral), their mental development according to the Resing & Blok (22) method, and the age at which the GMFM-88 (19) and PEDI-NL (20,21) were administered. Based on the matched control setup, this study demonstrated that CVI, indeed, causes a delay in the development of gross motor function, functional skills and caregiver assistance.

The results of our study and Schenk-Rootlieb et al. (7) and Da Costa et al. (5) demonstrate that children with CP with CVI were significantly more limited in their physical activities than those with CP without CVI. Our study confirms these results but also indicates that gross motor function and functional skills in children with CP with CVI as well as caregiver assistance are more limited compared with children with CP without CVI. The results of our study also support the conclusions of Da Costa et al. (5) and Ghasia et al. (8) that physical activities assisting in the exploration of the environment are limited in children with CP with CVI. Both Da Costa et al. (5) and Ghasia et al. (8) described a relationship between the presence of visual impairment including CVI and the presence of motor function impairment in children with CP and concluded that the presence of visual impairments such as CVI in a group of children with CP and GMFCS Levels III, IV or V is higher than in children with CP and GMFCS Levels I and II. Our study indicates that children with CP with CVI experience difficulty on all of the GMFCS levels, and CVI could occur at all GMFCS levels.

In accordance with the study of Da Costa et al. (5) and Ghasia et al. (8), we included comorbidities such as hearing problems, epilepsy, the level of mental development and speech/language development in our study because they may affect the gross motor function and functional skills in children as well as caregiver assistance for them (5,8). Therefore, we can conclude that there are many aspects of CP that could cause limitation in physical activities and CVI, indeed, could be one of the causes of limited physical activities in children with CP.

Furthermore, the current measuring instruments that are employed in rehabilitation do not consider the presence of CVI in the group of children with CP. Evidently, reliable information cannot be obtained regarding the level of functioning of children experiencing CP with CVI. Therefore, it is important that, in addition to the development of reliable measuring instruments, specific intervention programs should be developed that take into consideration the presence of CVI in children with CP.

Moreover, in the future research, it would be interesting to examine whether current management strategies for CVI would also improve gross motor function and functional skills in these children as well as their caregiver assistance (i.e. does treating CVI also treat gross motor function and functional skills?).

Limitation

Our study involved a specific target group, i.e. children with spastic CP who are characterized with an increased level of muscle tension. Previous studies also included children with ataxic and dyskinetic types of CP (6,8). Children with ataxic CP experience a loss of normal muscle coordination, which results in movements with abnormal strength, rhythm and precision while those with dyskinetic CP present with involuntary, uncoordinated and recurring movement including times at rest. Thus, movement patterns of children with either ataxic or dyskinetic CP differ from those of children with spastic CP. Since children with different types of CP possess varying motor performance, the inclusion of different types of CP decreases the ability to generalize research results. For this reason, children with ataxic, dyskinetic and spastic CP cannot be compared in regard to gross motor function, functional skills and caregiver assistance. Despite these results, children with spastic CP in relationship to their visual functioning could be generalized to children with ataxia or dyskinesia in future research. This needs to be investigated in other types of CP as well.

In our study, significant differences in the three levels of speech/language development were evident between the two groups. These differences may be partly responsible for the diminished gross motor function and functional skills as well as increased caregiver assistance of the group of children with CP and CVI. The other reason could be the fact that both groups were not matched at the level of speech/language development which will be significant in future research.

Table III. The Paediatric Evaluation of Disability Inventory-NL (PEDI-NL, Dutch version): the difference in raw score per pair between (a) children with CP without Cerebral Visual Impairment (CVI) and (b) those with Cerebral Palsy (CP) with CVI.

Pairs	Functional skills			Caregiver assistance		
	Self-Care	Mobility	Social Functioning	Self-Care	Mobility	Social Functioning
1a-1b	6	35	5	12	4	5
2a-2b	-1	14	-24	5	3	1
3a-3b	18	21	10	4	13	11
4a-4b	- 3	17	17	-3	8	9
5a-5b	36	15	25	14	16	15
6a-6b	26	7	30	6	22	14
7a-7b	33	19	38	18	23	20
8a-8b	30	27	24	13	3	9
9a-9b	11	26	12	3	11	7
10a-10b	15	6	1	3	3	2
11a-11b	35	19	11	27	18	5
12a-12b	4	9	3	3	1	2
13a-13b	7	2	7	4	2	6
14a-14b	19	-1	23	10	4	10
15a-15b	3	3	2	2	2	1
16a-16b	16	5	37	8	8	16
17a-17b	11	1	14	3	1	14
18a-18b	8	1	16	18	9	19
19a-19b	3	14	2	2	3	2
20a-20b	4	2	17	1	9	14
21a-21b	0	0	1	-1	0	0
22a-22b	-11	17	1	-2	12	0
23a-23b	1	10	5	0	5	3

A negative value indicates that a child with CP without CVI received a lower score than the child with CP with CVI.

In our study, the presence of epilepsy was not evenly distributed between both groups of children. Epilepsy may have an impact on the visual ability, such as less visual attention, resulting in affects at the level of gross motor function, functional skills and caregiver assistance in children with CP and CVI. Future studies should take this into consideration because it may explain some of the differences between both groups in the outcome of our study.

Recommendations

During the treatment and supervision of this type of group of children, it is important to discover which sensory (auditory, proprioceptive, tactile, vestibular) compensation strategies a child utilizes to support his/her visual perception (28). The ability to visually perceive movement is rarely damaged in children with CVI, and it is advisable to include movement as an aid during testing and treatment (28). Additionally, support in the area of the gross motor function, functional skills and caregiver assistance are often hands-off. This group of children would benefit from hands-on and verbal support in a task-oriented environment.

Clinical experience suggests that providing verbal and manual support allows children with CP with CVI to achieve motor development milestones easier and assists them in performing daily activities. Therefore, in order to improve the level of gross motor function, functional skills and caregiver assistance, it is

Table IV. Gross Motor Function Measure-88 (GMFM-88) and Pediatric Evaluation of Disability Inventory-NL (PEDI-NL, Dutch version) (PEDI-NL) scores in children with cerebral palsy (CP) without cerebral visual impairment (CVI) and CP with CVI

	CP without CVI		CP with CVI		*p*-value
	Mean (SD)	Median (min-max)	Mean (SD)	Median (min- max)	
GMFM-A	87 (19)	100 (27-100)	73 (29)	86 (14-100)	p<0.001
GMFM-B	75 (32)	93 (7-100)	58 (38)	65 (0-100)	p<0.001
GMFM-C	62 (41)	80 (0-100)	40 (40)	26 (0-100)	p<0.001
GMFM-D	42 (41)	28 (0-100)	27 (35)	3 (0-90)	p=0.009
GMFM-E	30 (35)	14 (0-91)	20 (29)	4 (0-83)	p=0.002
GMFM-Total	59 (30)	63 (6.4-97)	44 (32)	38 (4-94.5)	p<0.001
PEDI –FS Self-Care	31 (19)	29 (5-65)	20 (11)	18 (3-49)	p<0.001
PEDI- FS Mobility	35 (20)	28 (3-62)	24 (15)	26 (2-49)	p<0.001
PEDI-FS Social Functioning	37 (18)	45 (3-61)	25 (16)	22 (1-56)	p<0.001
PEDI –CA Self-Care	11 (9)	10 (0-32)	5 (5)	3 (0-19)	p<0.001
PEDI- CA Mobility	17 (12)	12 (0-35)	9 (8)	8 (0-24)	p<0.001
PEDI-CA Social Functioning	14 (7)	15 (0-24)	6 (6)	4 (0-19)	p<0.001

GMFM, Gross Motor Function Measure: A, laying and rolling, B, sitting, C, crawling and kneeling, D, standing, E, walking, running, jumping); PEDI-NL, Paediatric Evaluation of Disability Inventory-NL; FS, Functional Skills, CA, Caregiver Assistance.

important to use verbal and manual support, e.g. using slow-tempo speech while training a skill, simplifying daily tasks and presenting them in a low tempo, offering structure and predictability, and using rituals can support a child with CP with CVI in optimally performing a task. Furthermore, the use of bright colors, shiny and fluorescent material, a light source, and moving objects can help increase the child's attention visually.

Conclusion

Children with CP with CVI appear to be more limited in their gross motor function, functional skills, caregiver assistance and in their level of independence when performing daily activities compared with children with CP without CVI.

Children with CP with CVI clearly achieve lower scores in all dimensions of gross motor function including laying, rolling, sitting, crawling, kneeling, standing, walking and running.

The children with CP with CVI score significantly lower than those with CP without CVI on the PEDI-NL in the sections Functional Skills and Caregiver Assistance in the domains of self-care, mobility and social functioning.

Limitations in physical activities in children with CP could be caused not only by a delay in motor or mental development but also by the presence of CVI.

Therefore, it is vital that physicians, counselors and parents take into consideration that, when a child with CP exhibits a limitation of daily activities and slow processing and performance speed, it may not only stem from a delay in motor and/or mental development. He or she may be experiencing a visual impairment such as CVI.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

References

1. Rosenbaum P, Paneth N, Leviton A, Goldstein M, Bax M. Definition and classification of cerebral palsy. Dev Med Child Neurol. 2007;49:480.
2. Dutch Institute of Rehabilitation Paediatricians. Guidelines for the diagnosis and treatment of children with spastic cerebral palsy. Postbus 9696, 3506 GR Utrecht; 2007.

3. Stiers P, Vanderkelen R, Vanneste G, Coene S, De Rammelsere M, Vandenbussche E. Visual–perceptual impairment in a random sample of children with cerebral palsy. Dev Med Child Neurol. 2002;44:370–82.
4. Schenk-Rootlieb AJF, Van Nieuwenhuizen O, Van Waes PFGM, Van der Graaf Y. Cerebral visual impairment in cerebral palsy: Relation to structural abnormalities of the cerebrum. Dev Med Child Neurol 1994;25:68–72.
5. Da Costa MF, Salmao SR, Berezovsky A, De Haro FM, Ventura DF. Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. Behav Brain Res. 2004;149:145–50.
6. Schenk-Rootlieb AJF, Van Nieuwenhuizen O, Van der Graaf Y, Wittebol-Post D, Willemse J. The prevalence of cerebral visual disturbance in children with cerebral palsy. Dev Med Child Neurol 1993;34:473-80.
7. Schenk-Rootlieb AJF, Van Nieuwenhuizen O, Schiemanck N, Van der Graaf Y, Willemse J. Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. Children Care Health Dev 1993;19:411–23.
8. Ghasia F, Burnstroom J, Gordon M, Tychsen L. Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. Invest Ophthalmol Vis Sci. 2008;49:572–80.
9. Dutton GN, Jacobson LK. Cerebral visual impairment in children. Semin Neonatol. 2001;6:477–85.
10. Fazzi E, Signorini SG, LA Piana R, Bertone C, Misefari W, Galli J, Balottin U, Bianchi PE. Neuro-ophthalmological disorders in cerebral palsy: Ophthalmological, oculomotor, and visual aspects. Dev Med Child Neurol. 2012;54:730–6.
11. Dutton GN. The spectrum of cerebral visual impairment as a sequel to premature birth: An overview. Doc Ophthalmol. 2013;127:69–78.
12. Goodale MA. Separate visual systems for perception and action: A framework for understanding cortical visual impairment. Dev Med Child Neurol. 2013;55:9–12.
13. Dutton GN, Saaed A, Fahad B, et al. Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction - A retrospective observational study. Eye. 2004;18:27–34.
14. Milner AD, Goodale MA. Two visual systems re-viewed. Neuropsychologia. 2008;46:774–85.
15. Looijestijn PL, Zuidhoek S. Cerebral visual impairment. Visual Perception. 2007;51:6–8.
16. Verrel J, Bekkering H, Steenbergen B. Eye–hand coordination during manual object transport with the affected and less affected hand in adolescents with hemiparetic cerebral palsy. Exp Brain Res. 2008;187:107–16.
17. Guzzetta A, Mercuri E, Cioni G. Visual disorders in children with brain lesions: Visual impairment associated with cerebral palsy. Eur J Paediatr Rehabil Neurol Society. 2001;5:115–9.
18. Houwen S, Hartman E, Visscher C. Physical activity and motor skills in children with and without visual impairments. Med Sci Sports Exerc. 2009;41:103.
19. Ketelaar M, Van Petegem-van Beek E, Veenhof C, Visser J, Vermeer A. gross motor function measure. University of Utrecht. Child Phys Ther. 2003;39:5–7.
20. Wassenberg-Severijnen JE, Custers JWH. Pediatric Evaluation of Disability Inventory-NL. Amsterdam: Harcourt Assessment B.V.; 2005.
21. Wassenberg-Severijnen JE, Custers JW, Hox JJ, Vermeer A and Helders PJ. Reliability of the Dutch Pediatric Evaluation of Disability Inventory (PEDI). Clin Rehabil. 2003;17: 457–62.
22. Resing W, Blok J. The classification of intelligence score. Psychol Sci. 2002;244–9.
23. International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation). Dutch WHO-FIC Collaborating Centre. 1st ed. Houten: Bohn Stafleu van Loghum (www.bsl.nl); 2008.
24. Russell DJ, Rosenbaum PL, Avery LM, Lane M. Gross Motor Function Measure (GMFM-66 and GMFM-88) user's manual. London: MacKeith Press; 2002.
25. Alotaibi M, Long T, Kennedy E, Bavishi S. The efficacy of GMFM-88 and GMFM-66 to detect changes in gross motor function in children with cerebral palsy (CP): A literature review. Disabil Rehabil. 2014;36:617–27.
26. Gorter JW, Boonacker CWB, Ketelaar M. Gross Motor Function Classification System (GMFCS). Dutch J Phys Ther. 2005;4:115–6.
27. Haley SM, Coster WJ, Ludlow LH, Haltiwanger JT, Andrellos PJ. Pediatric Evaluation of Disability Inventory: Development, standardization, and administration manual. Boston, MA: New England Medical Centre Inc. and PEDI Research Group; 1992.
28. Steendam M. Do you know what I see? Cerebral visual impairment in children: A manual for professionals. Haren: Royal Visio Huizen; 2007.

3 |

Reliability of the modified Paediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment

M. Salavati[a,b], A. Waninge[a,b], E.A.A. Rameckers[c,d,e], A.C.E. de Blécourt[f], W.P. Krijnen[b], B. Steenbergen[g,h], C.P. van der Schans[b,f]

[a] Royal Dutch Visio, Centre of Expertise for Blind and Partially Sighted People, The Netherlands
[b] Hanze University of Applied Sciences Groningen, Research and Innovation Group in Health Care and in Nursing, Groningen, The Netherlands
[c] Maastricht University, Department of Rehabilitation Medicine, School for Public Health and Primary Care (CAPHRI), Maastricht, The Netherlands
[d] Adelante Center of Expertise in Rehabilitation and Audiology, Valkenburg and Hoensbroek, The Netherlands
[e] AVANSplus, University for Professionals for Paediatric Physical Therapy, Breda, The Netherlands
[f] University Medical Center Groningen, Department of Rehabilitation Medicine, Groningen, The Netherlands
[g] Australian CatholicUniversity,SchoolofPsychology,Melbourne,Australia
[h] Radboud University Nijmegen, Behavioural Science Institute, Nijmegen, The Netherlands

Published as:

Salavati, M.,Waninge, A., Rameckers, E.A.A., Ble′court, A.C.E., Krijnen, W.P., Steenbergen, B., et al. (2015). Reliability of the modified Paediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment. Research in Developmental Disabilities, 37, 189–201.

Abstract

Purpose: The aims of this study were to adapt the Paediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) for children with cerebral visual impairment (CVI) and cerebral palsy (CP) and determine test-retest and inter-respondent reliability.

Method: The Delphi method was used to gain consensus among twenty-one health experts familiar with CVI. Test-retest and inter-respondent reliability were assessed for parents and caregivers of 75 children (aged 50-144 months) with CP and CVI. The percentage identical scores of item scores were computed, as well as the interclass coefficients (ICC) and Cronbach's alphas of scale scores over the domains self-care, mobility, and social function.

Results: All experts agreed on the adaptation of the PEDI-NL for children with CVI. On item score, for the Functional Skills scale, mean percentage identical scores variations for test-retest reliability were 73-79 with Caregiver Assistance scale 73-81, and for inter-respondent reliability 21-76 with Caregiver Assistance scale 40-43. For all scales over all domains ICCs exceeded 0.87. For the domains self-care, mobility, and social function, the Functional Skills scale and the Caregiver Assistance scale have Cronbach's alpha above 0.88.

Conclusion: The adapted PEDI-NL for children with CP and CVI is reliable and comparable to the original PEDI-NL.

1. Introduction

Cerebral palsy (CP) describes a group of permanent disorders of movement and posture development, causing activity limitation; they are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. Gross motor function of children with CP is classified using the Gross Motor Function Classification System (GMFCS) into five different severity levels, where level 1 indicates the least and level 5 the most functional limitation (Dutch Institute of Rehabilitation Paediatricians, 2007; Rosenbaum, Paneth, Leviton, Goldstein, & Bax, 2007). Motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour; by epilepsy; and by secondary musculoskeletal problems (Dutch Institute of Rehabilitation Paediatricians, 2007; Rosenbaum et al., 2007).

In general, children with visual impairments have more difficulty to express themselves, they are usually delayed in language skills, appear to be at a disadvantage when performing reading tasks, and learning to write can also be difficult compared to children without visual impairment. Furthermore, developmental milestones that normally require vision e.g. reaching and walking are often delayed in children with visual impairments (Good, Jan, Burden, Skoczenski, & Candy, 2001).

The spectrum of visual impairments in children with CP is extremely broad and includes both ocular visual impairment (OVI), such as strabismus, reduced visual acuity, ocular nystagmus, refraction disorders, and retinopathies, and cerebral visual impairment (CVI), which is a problem of central origin. CVI is observed in approximately 30% of children diagnosed with various forms of CP (Da Costa, Salmao, Berezovsky, De Haro, & Ventura, 2004; Dutton & Jacobson, 2001; Ghasia, Burnstroom, Gordon, & Tychsen, 2008; Schenk-Rootlieb, Van Nieuwenhuizen, Van der Graaf, Wittebol-Post, & Willemse, 1993a; Schenk-Rootlieb, Van Nieuwenhuizen, Schiemanck, Van der Graaf, & Willemse, 1993b; Schenk-Rootlieb, Van Nieuwenhuizen, Van Waes, & Van der Graaf, 1994; Stiers et al., 2002). CVI can be defined as deficient visual function, as a sequel of damage or malformation of the retrogeniculate visual pathways (optic radiations, occipital cortex and visual association areas); in the absence of damage of the anterior visual pathways or any major ocular disease. Also, CVI is diagnosed by exclusion of OVI. CVI is a prominent sequel to premature birth, particularly when prematurity is extreme (Dutton & Jacobson, 2001). The focus of this study will be on CVI because the need of children with OVI is different to those with CVI (Da Costa et al., 2004; Dutton & Jacobson, 2001; Ghasia et al., 2008; Schenk-Rootlieb et al., 1993a, 1993b, 1994; Stiers et al., 2002).

The role of visual perception during motor action and development is very important, because children with CP show increased visual monitoring during motor activities (walking, daily-life activities and play), and this emphasises the role of visual perception during motor action (Verrel, Bekkering, & Steenbergen, 2008). Considerable focus has been directed towards the detection and treatment of prematurity-caused retinopathy, but there tends to be less of a focus on CVI, which can consequently be overlooked. Dutton and Jacobson (2001), focused primarily on profound visual impairment in children experiencing CVI. CVI ranges in severity from blindness to relatively minor impairments of visual perception. Perceptual visual dysfunction and disorders of visual attention, often with only minimally reduced or normal visual acuities, are increasingly recognised as forms of CVI (Dutton & Jacobson, 2001; Fazzi et al., 2012; Dutton, 2013). It is stated that CVI has an impact on all aspects of child's development, and children with CP and CVI develop more slowly in the area of Self-care, Mobility and Social Function than children with CP and no CVI. However, children with CVI exhibit slow, inefficient, and highly variable visual performance during daily-life activities (Da Costa et al., 2004; Dutton & Jacobson, 2001; Dutton, 2013; Ghasia et al., 2008; Good et al., 2001; Salavati, Rameckers, Steenbergen, & Schans van der, 2014; Schenk-Rootlieb et al., 1993a). It is important to evaluate daily-life activities for Self-care, Mobility and Social Function in children with CVI by a reliable and valid test. This is the subject of our study.

The PEDI-NL (Wassenberg-Severijnen & Custers, 2005; Wassenberg-Severijnen, Custers, Hox, Vermeer, & Helders, 2003; Custers et al., 2002) is a questionnaire that evaluates the daily skills of children aged 6 months to about 7.5 years. It measures both capability (what the child can do) and performance (what the child actually

does) of daily routine childhood activities in the Self-care, Mobility and Social Function domains, using a structured interview with the parents or caregivers (Wassenberg-Severijnen & Custers, 2005; Wassenberg-Severijnen et al., 2003; Custers et al., 2002). A child's capability can be measured utilising the PEDI-NL's three Functional Skills scales, performance using its three Caregiver Assistance scales and Modification scales. Reliability of the PEDI-NL in children with CP without CVI is sufficient (Wassenberg-Severijnen et al., 2003), yet the validity and reliability study assessment did not include children with visual impairment (Wassenberg-Severijnen et al., 2003). Professionals working with children with CVI and CP at Royal Dutch Visio and Bartiméus, centres of expertise for blind and partially sighted people in The Netherlands, experienced that the PEDI-NL does not account for the presence of CVI and might not be appropriate for children with CVI due to their difficulties at different levels of functioning. Also, as a consequence of CVI a child might not be able to show his/her full capacity in the domains of Self-care, Mobility and Social Function during the standardised assessment of a developmental test (Salavati et al., 2014; Wassenberg-Severijnen et al., 2003; Haley, Coster, Ludlow, Haltiwanger, & Andrellos, 1992; Visser, Ruiter, Meulen van der, Ruijssenaars, & Timmerman, 2014). In line with this, there are no reliable and valid instruments available to measure these domains in children with CP and CVI (Custers et al., 2002). Considering the fact that a high number of children with CP also has CVI, it is very important for paediatric physical therapists and occupational therapists to have at their disposal an instrument that is adapted for children with CVI, because the original PEDI-NL renders inaccurate performance outcome results and negative impacts for treatment. In this study we developed an adapted version of the PEDI-NL and determine its test-retest reliability and inter-respondent reliability for children with CVI and CP.

2. Methods

The study was carried out in two phases. First, the PEDI-NL (Table 1) was adapted for children with CVI. Second, the psychometric properties of the adapted PEDI-NL were examined in children with CP and CVI. Table 1 gives an overview of topics in the PEDI-NL. Both the capability of the child (Functional Skills scale, 205 items) and the amount of help he/she gets from his/her parents (Caregiver Assistance scale, 20 items) as well as the equipment used (Modifications scale, 20 items) in daily tasks are measured by a structured interview with parent(s) and caregivers. Functional Skills are determined in three domains: Self-care, Mobility and Social Function.

Items in the Functional Skills scale are dichotomous and are scored as either 'capable' or 'not capable'. Summed scores can be computed in every domain and transformed to standardised scores. Both the Caregiver Assistance Scale and the Modifications Scale are ordinal scales, ranging from 'totally dependent' to 'totally independent' (Wassenberg-Severijnen et al., 2003). The Modifications Scale measuring the equipment used is an ordinal scale, and consists of "no modifications", "child-oriented modifications", "specialised rehabilitation equipment", and "extensive modifications" (Wassenberg-Severijnen et al., 2003).

2.1. Phase 1

2.1.1. Adaptation

The Delphi method was used to gain consensus among a panel of experts. It is applied in a series of sequential questionnaires or 'rounds', interspersed by controlled feedback, in order to seek for the most reliable consensus of opinion of a group of experts (Powell, 2003).

Firstly, we identified relevant disciplines and their skills, giving those experts the opportunity to participate by e-mail in the study of adaptation of PEDI-NL. We invited experts with different backgrounds to the study of adaptation of PEDI-NL if they had experience with children who have CVI. Al of them worked at Royal Dutch Visio and Bartiméus, centres of expertise for blind and partially sighted people in the Netherlands. Secondly, we explained to the expert the purpose of this study and the required procedures. All of the invited experts confirmed their desire to participate. The experts were asked for their age, profession, working experience and familiarity with the PEDI-NL.

2.1.2. Data collection for adaptation

The health experts familiar with CVI participated in the adaptation process by studying the PEDI-NL, and when required adapted the instruction part of

Table 1. Content of the PEDI-NL (Wassenberg-Severijnen, Custers, Hox, Vermeer, & Helders, 2003).

Domains	Functional Skills scale/subscales	Number of items	Caregiver Assistance scale Modifications scale
Self-care	Types of food textures	4	Eating
	Use of utensils	6	
	Use of drinking containers	5	
	Tooth brushing	5	Grooming
	Hair brushing	4	
	Nose care	5	
	Hand washing	5	Bathing
	Washing body and face	5	
	Pullover/front-opening garments	5	Dressing upper body
	Fasteners	5	
	Pants	5	Dressing lower body
	Shoes/socks	5	
	Toileting task	5	Toileting
	Management of bladder	5	Bladder management
	Management of bowel	5	Bowel management
Mobility	Toilet transfers	5	Chair/toilet transfers
	Chair/wheelchair transfers	5	
	Car transfers	5	Car transfers
	Bed mobility/transfers	5	Bed mobility/transfers
	Tub transfers	5	Tub transfers
	Indoor locomotion methods	3	Indoor locomotion
	Indoor locomotion-distance/speed	5	
	Indoor locomotion-pull/carriers objects	5	
	Outdoor locomotion methods	3	Outdoor locomotion
	Outdoor locomotion-distance/speed	5	
	Outdoor surfaces	5	
	Up stairs	5	Stairs
	Down stairs	5	
Social Function	Comprehension of word meanings	5	
	Comprehension of sentence complexity	5	
	Functional use of communication	5	
	Complexity of expressive communication	5	
	Problem resolution	5	
	Social interactive play (adults)	5	
	Peer interactions (child of similar age)	6	
	Play with objects	5	
	Self-information	5	
	Time orientation	5	
	Play with objects	5	
	Self-protection	5	
	Community function	5	

the content for children with CVI. Specific feedback information could be written about any of the items. E-mail was used to process and resubmit all experts' comments in three rounds. The predetermined goal was to reach a consensus of 65% among the experts after the first round, 75% second round and 85% third round for agreement with the content of the adapted version of PEDI-NL. Also, the experts were asked to explain and justify their comments on each question and instruction, without changing the original questions and instructions. No items were eliminated from the original PEDI-NL during the adaptation process.

2.1.3. First round

All experts were invited to study the instruction part of the PEDI-NL, then gave comments individually from their viewpoint on each question and original instruction. In accordance with other studies (Ghasia et al., 2008; Dutton & Jacobson, 2001; Dutton, 2013; Steendam, 2007) on CVI, the experts were specifically asked:

Which verbal support/instruction or manual support needs to be added to the instruction of the PEDI-NL to make it suitable it for children with CVI? At the equipment level: what kind of adjustment needs to be added to the instruction of the PEDI-NL to make it suitable it for children with CVI?

For use of manual support they were asked, as applicable, to describe the amount of manual support (e.g. duration and phase of needed manual support given) in order to help a child accomplish a particular skill. At the level of verbal support they were asked, as applicable, what especially should be said in order

to help a child accomplish a particular skill. For use of materials they were asked which characteristics a material needed to have (e.g. colourful, sound-produced, high in contrast) to receive the attention of a child with CVI in order to use this specific material.

2.1.4. Second and third rounds

After receiving the experts' comments on the questions and instruction of PEDI-NL, we processed all of the suggestions in the instruction part of PEDI-NL and resubmitted this twice to the experts. We asked again the same questions as in the first round.

2.2. Phase 2

2.2.1. Participants

Children with CP and CVI were recruited from Royal Dutch Visio and allied health care practices. Inclusion criteria were presence of all types of CP and CVI, mild or moderate intellectual disability, and age at testing of the modified PEDI-NL for children with CVI between 4 and 12 years. Level of intellectual disability was reported from the children's medical files; Children with a syndrome (e.g. Down syndrome) and hearing difficulties (>30 db) were excluded. Also, children with severe or profound intellectual disability (IQ < 60) were excluded. The diagnosis of CP and the classification according to GMFCS level were aggregated from the children's medical files and judged by a rehabilitation specialist.

CVI is a heterogeneous diagnosis with large variability between children and it can present in many forms. The diagnosis of CVI was determined based on the results of ophthalmological and (neuro-) psychological research, and on the assessment data reported by a developmental coach specialised in working with children with visual impairments (Fig. 1). According to these, the diagnosis of CVI was determined by the following criteria: a normal or near normal eye exam performed by ophthalmologist; a history or presence of neurological problems; presence of behavioural responses to visual stimuli which are unique to CVI. This results in strong colour preference, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non purposeful gaze, difficulty with distance viewing, absent or atypical visual reflexes, difficulty with visual novelty, and absence of visually guided reach. Also, Children with (corrected) vision < 0.3 and/or field of vision 30° were excluded. In our study, all children with CVI were included.

This study was approved by the Medical Ethical Committee (METC-2013.104) of University Medical Center Groningen (UMCG), the Netherlands. Written informed consent was obtained from the children's parents.

2.3. Design

For test-retest reliability the same caregiver familiar to the child, was interviewed twice by a therapist, within three weeks. For inter-respondent reliability we used the data of one of the parents and the caregiver within a few days, in order to prevent overload of the parents. The therapists conducted all interviews with parents and caregivers. The caregiver was a person with knowledge and skills on how take care of the child e.g. at school and was familiar with the child. The percentage identical scores of item scores were computed, and interclass coefficients (ICC) and Cronbach's alphas for domain Self-care, Mobility and Social Function were calculated.

2.4. Data collection

We collected data on gender as well as prevalence of epilepsy and speech/language development according to the International Classification of Functioning, Disability and Health for Children and Youth (Dutch Translation, 2008) to account for the possible effect on Self-care, Mobility, and Social Function. At the level of speech/language development, the collected data were: d3100 = reacts to human voice; d3101 = understands simple spoken messages; d3102 = understands complex spoken messages; d330 = speaks; d331 = babbles; d3350 = uses body language and d3351 = uses signs symbols (Table 3). The data of children were registered according to GMFCS level and type of CP (unilateral or bilateral), level of intellectual disability, and age at which the PEDI-NL were administered.

2.5. Statistical analyses

The data were analysed using the Statistical Package for Social Sciences (SPSS), v.22 software. Inter-

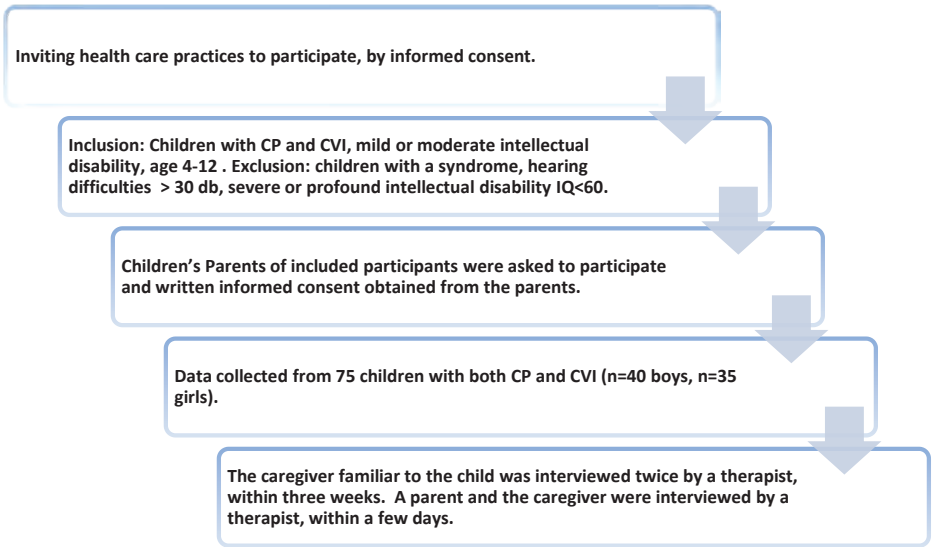


Figure 1. Procedure for inclusion.

respondent reliability and test-retest reliability were established using the partial (item) and total (scale) scores of PEDI-NL-CVI from children with CP and CVI. To calculate reliability of the modified PEDI-NL for children with CVI, we used the proportion of identical answers on every item. Also, intraclass correlation coefficients (ICC, two-way random, absolute agreement, single measure) and their confidence intervals of test-retest reliability and inter-respondent reliability for each domain were calculated. The two-way random variant of ICC for single measurements was chosen to measure degree of absolute agreement because the children as well as the observers were a random factor in the design (McGraw & Wong, 1996; Lexell & Downham, 2005). P-values below 0.05 were considered statistically significant. To test for systematic bias, the mean of the differences and the standard deviation of differences were calculated by the paired t-test. Limits of agreement (LOA) for inter-respondent reliability and test-retest reliability of the Functional Skills (FS) and Caregiver Assistance (CA) scales (domains Self-care, Mobility, Social Function) were calculated.

Internal consistency of each domain in the Functional Skills scale and the Caregiver Assistance scale was assessed by Cronbach's alpha based on the caregiver measurement. The value of Cronbach's alpha (Cronbach, 1951) was interpreted as: poor < 0.5, moderate 0.5-0.75, good 0.75-0.9, excellent > 0.9. Test-retest reliability for the Functional Skills and Caregiver Assistance scales was examined using intraclass correlation coefficients (ICCs). The ICC was used to analyse the agreement between the interviews from each of two domains. The ICC value was interpreted as follows: poor < 0.5, moderate 0.5-0.75, good 0.75-0.9, excellent > 0.9.

3. Results

3.1. Phase 1

3.1.1. Adaptation

Twenty-one health experts familiar with CVI participated in the adaptation process of the PEDI-NL; sixteen of them (male:female = 2:14) were paediatric physical therapists, two (male:female = 0:2) occupational therapists, one female speech and language therapist, and two (male:female = 1:1) behavioural scientists. Mean (SD) age of the experts was 50 (10) years and their mean years (SD) of experience with children with CVI was 18 (8). Nineteen of them were familiar with the PEDI-NL.

3.1.2. Delphi first round

Sixty-five percent of the experts agreed on the instruction part of PEDI-NL. With respect to the original instruction, most adaptations were at the

Functional Skills scale and Caregiver Assistance scales, on domain Self-care and Mobility. Also, the largest agreement amongst experts was encountered on domain Self-care and Mobility. For example, at the level of using 'a cup or a spoon' the child needed to know about the object in advance in order to locate it. It is thus assumed that the child with CVI receives verbal and manual support. Furthermore, during a toilet transfer the child uses the hands in order to orient itself to walk to the bathroom. Hence while the child with CVI walks to the bathroom, it is allowed to use the hands for orientation and not for support.

The least agreement was found on domain of Social Function. For 'Comprehension of sentence complexity', a child with CVI could have difficulty localising an object well by using visual information. It is therefore assumed that the child 'knows' the location of an object. The experts agreed that, depending on the visual impairment, children with CVI often have difficulty with of 'using a bicycle in an unknown environment'. This involves moving in an outdoor familiar environment. Adult supervision is thus required for this activity, for safety reasons.

For the Modification scale, the experts gave some suggestions to add adapted equipment, such as coloured toothbrush or a coloured cup for children with CVI.

3.1.3. Delphi second round

Seventy-five percent of the experts agreed on the instruction part of PEDI-NL after the second round. Most comments were on domain of Social Function, where, in terms of inconsistency of visual perception, a child could have difficulty 'playing safely at home' or 'following the rules'. With regard to this item an additional explanation is given (Table 2). Generally, when a task is more complicated, the child will have more difficulty performing that particular task. For example, the child has difficulty running errands independently or finding his/her way home. If there is a lack of visual information, a child may have difficulty imitating. Hence the experts agreed to use verbal support in order to explain the content of visual information to a child with CVI.

3.1.4. Delphi third round

After receiving the experts' comments we processed the suggestions, mostly on domain of Social Function in the instruction part of PEDI-NL, and resubmitted to the experts. The experts responded, indicating 100% satisfaction with the resulting overall content of the adapted version of PEDI-NL (PEDI-NL-CVI). With respect to CVI the professionals applied specific feedback information, which is added as an appendix to the instruction of the original PEDI-NL. This appendix (Table 2) can be used for children with CVI. Considering the fact that visual perception contributes to the performance of daily-life activities, it is important to use the version of PEDI-NL-CVI for children who have CVI.

The experts agreed on content of the PEDI-NL-CVI, and considered that this version presented all the important issues in the specific domains for children with CVI. The adapted version consists of additional introductions for all domains such as Self-care, Mobility, Social Function, and modification or used adaptive equipment.

3.2. Phase 2

3.2.1. Inter-respondent reliability and test-retest reliability

Children were tested between June 2013 and April 2014. Mean time between the two interviews was 14 days (SD 3 days). Mean time between researcher interviews of parent and caregiver was 7 days (SD 2 days). We collected the data from 75 children with both CP and CVI (n = 40 boys and n = 35 girls). Table 3 shows the children's characteristics. All children with CVI were included in our study and no selection was done based on subtypes. Therefore, we assume that different subtypes are represented in our study.

Tables 4 and 5 present summed scores (mean scores and standard deviation) for each domain of the Functional Skills scale and the Caregiver Assistance scale from the test-retest and inter-respondent reliability assessments in children with CP and CVI.

For test-retest reliability, the mean percentage (SD) of identical scores for the Functional Skills scale was 73 (2.5) for the domain of Self-care, 76 (0.9) for Mobility and 79 (1.4) for Social Function, indicating moderate-to-good identical scores. The mean percentage (SD) of identical scores for the Caregiver Assistance scale

Table 2. Summed changes proposed by professionals at the level of the Functional Skills scale.

Domains	Functional Skills scale/subscales	Adaptation
Self-care	Types of food textures	It is assumed that the child knows that the food will be announced verbally in advance, and will be offered at a leisurely pace
	Use of utensils	It is assumed that the child knows the position of the cutlery
	Use of drinking containers	Because children with CVI, compared to peers without CVI, have difficulty drinking without spilling, it is assumed that tasks like picking up a cup or drinking will be verbally announced
	Tooth brushing	It is assumed that the parents/caregivers will verbally announce the position of the toothbrush and toothpaste if the child doesn't know their position
	Hair brushing	It is assumed that the parents/caregivers use verbal support to help the child with this task
	Nose care	It is assumed that the parents/caregivers use verbal support to tell the child the position of the handkerchief; if necessary, they give the handkerchief to the child
	Hand washing	It is assumed that the parents/caregivers use verbal support to tell the child the position of the soap, faucets, sink, washcloth and towel
	Washing body and face	It is assumed that the parents/caregivers use both verbal and manual support to help the child find the position of the soap, faucets, sink and towel
	Pullover/front-opening garments	It is assumed that the parents/caregivers use verbal and manual support to help the child find the position of the clothes, and the clothes lie on a place that is known by the child. The child is able to scan the front and the back of the cloth, e.g. a pullover or a t-shirt
	Fasteners	It is assumed that the parents/caregivers use verbal support to help the child with these tasks
	Pants	It is assumed that the parents/caregivers use verbal and manual support to help localise the position of the clothes and to help the child dress and undress
	Shoes/Socks	It is assumed that the parents/caregivers use verbal support to help the child localise the socks and the shoes
	Toileting task	It is assumed that the parents/caregivers use verbal and manual support to help the child with these tasks
	Management of bladder	It is assumed that the parents/caregivers use verbal and manual support to help the child with these tasks
	Management of bowels	It is assumed that the parents/caregivers use verbal and manual support to help the child with these tasks
Mobility	Toilet transfers	Because the intention to move in relation to the transfer is less present in a child with CVI, it is assumed that the child uses the arms/hands to orient itself (G33). It is assumed that the parents/caregivers use verbal support to help the child with these tasks. Also, the lighting in the bathroom is adequate
	Chair/wheelchair transfers	Because of the lack of depth perception the child has an adverse influence on the transfer. It is assumed that the child is familiar with the chair, knows its location, and uses a chair with armrests. The child uses these armrests as an orientation point (see 'Caregivers Assistance' and 'Modifications Scale')
	Car transfers	The child can use its arms/hands for orientation. It is assumed that the parents/caregivers use verbal support to help the child with these tasks
	Bed mobility/transfers	Because the intention to move in relation to the transfer is less present in a child with CVI, it is assumed that the child uses the arms/hands for orientation
	Tub transfers	Because the intention to move in relation to the transfer is less present in a child with CVI, it is assumed that the child uses the arms/hands for orientation
	Indoor locomotion methods	E.g. in situations in which the child moves or is stimulated to move by visual support in the form of fluorescent toys, toys with a big contrast and/or toys that produce sound. 'Inside' means the familiar environment of the child
	Indoor locomotion-distance/speed	E.g. in situations where the child moves and follows fluorescent toys or toys that produce sound or have a big contrast, all while being verbally supported and stimulated to move. It is assumed that there are no unknown obstacles or abrupt transitions (in floor types or colours) between two rooms along the child's route
	Indoor locomotion-pull/carrier objects	It is assumed that the child must have a hand/arm free to orientate (C12), and that there are no unknown obstacles along the child's route
	Outdoor locomotion methods	Because the child has difficulty perceiving depth in relation to moving on different types of surfaces, the use of arms/hands to orientate is allowed (J46). This involves moving outside the familiar environment, and it is assumed that there are no obstacles along the child's route
	Outdoor locomotion-distance/speed	This involves moving in an outdoor familiar environment. An activity done 'without help' is still supervised by an adult for safety reasons. It is assumed that the child uses certain objects (such as a wall) to move. These objects are not used as support
	Outdoor surfaces	Because the child has difficulty perceiving depth in relation to moving across different kinds of surfaces, the use of arms/hands to orientate is allowed
	Climbing stairs	It is assumed that the child is familiar with the stairs and the height of the steps.
	Going down stairs	It is assumed that the child is familiar with the stairs and the height of the steps. In this situation the supervisor walks backwards down the stairs, verbally inviting the child to move down (depending on the demand, crawl and slide on the buttocks or walk)

Table 2. Continued.

Domains	Functional Skills scale/subscales	Adaptation
Social Function	Comprehension of word meanings	Specific concepts such as the name of a toy that is familiar to the child
	Comprehension of sentence complexity	Because a child with CVI cannot localise an object well, it is assumed that the child 'knows' the location of an object. For example: the child has both the cup and the plate in its hands and the researcher asks the child, 'give me your cup'. In this situation it is assumed that the child is able to give the cup to the researcher by knowing what a cup is and what a plate is. The child understands and executes the task 'the ball is under the table' because he/she has done it before and knows the route to the ball
	Functional use of communication	For all of the items listed below it is assumed that the child receives verbal support to help localise an object such as a cup
	Complexity of expressive communication	No adaptation
	Problem resolution	No adaptation
	Social interactive play (adults)	It is allowed for the child to make contact in other ways than just 'visual' contact with an adult or with a toy. It is assumed that the child receives both verbal and manual support to help find toys such as balls or building blocks (G33 and G34)
	Peer interactions (child of similar age)	The child makes not only 'visual' contact but also 'attention' contact with other children. It is assumed that the child is familiar with age-appropriate games that are played beforehand in a one-on-one situation
	Play with objects	It is assumed that in all situations the parents/caregivers use verbal support to help the child understand the game beforehand, and that the child is familiar with the rules regarding the toy
	Self-information	No adaptation
	Time orientation	No adaptation
	Self-protection	It is assumed that a child with CVI crosses the street under guidance/supervision
	Community function	This relates to both inside and outside the home and familiar surroundings. It is assumed that the child is familiar with the environment outside the house. For all items listed below it is assumed that the child receives both verbal and manual support to help with the process of familiarisation with the objects without helping him/her complete the task: for example, when the child climbs on top of the furniture it is expected to come down from it safely

Table 3. GMFCS, Gross Motor Function Classification system; n, numbers; ICF-CY, International Classification of Functioning, Disability and Health, Child and Youth version (Dutch translation).

Characteristic	Children with CP and CVI
Age, months, min.–max., mean (SD)	50–144, 114 (29)
Gender, male/female (*n*)	40/35
Type of cerebral palsy	
Spastic (*n*) (%)	71 (95)
Dyskinetic (*n*) (%)	3 (4)
Ataxic (*n*) (%)	1 (1)
GMFCS I (*n*) (%)	Bilateral 17 (23), unilateral left 2 (3), unilateral right 1 (1)
GMFCS II (*n*) (%)	6 (8) bilateral
GMFCS III (*n*) (%)	10 (13) bilateral
GMFCS IV (*n*) (%)	19 (25) bilateral
GMFCS V (*n*) (%)	20 (27) bilateral
Speech/language development ICF-CY, d3100 = reacts to human voice (*n*) (%)	75 (100)
Speech/language development	75 (100)
ICF-CY, d3101 = understands simple spoken messages (*n*) (%)	
Speech/language development	47 (63)
ICF-CY, d3102 = understands complex spoken messages (*n*) (%)	
Speech/language development	44 (59)
ICF-CY, d330 = speaks (*n*) (%)	
Speech/language development	31 (41)
ICF-CY, d331 = babbles (*n*) (%)	
Speech/language development	51 (68)
ICF-CY, d3350 = uses body language (*n*) (%)	
Speech/language development	37 (49)
ICF-CY, d3351 = uses signs and symbols (*n*) (%)	
Level of intellectual disability: mild/moderate (*n*) (%)	33 (44)/42 (56)
Presence of epilepsy: yes/no (*n*) (%)	12 (16)/63 (84)
Use of epilepsy medication: yes/no (*n*) (%)	11 (15)/64 (85)
Percutaneous endoscopic gastrostomy tube feeding: yes/no (*n*) (%)	12 (16)/63 (84)

GMFCS, Gross Motor Function Classification system; n, numbers; ICF-CY, International Classification of Functioning, Disability and Health,Child and Youth version (Dutch translation).

was 73 (1.3) for the domain of Self-care, 81 (6.0) for Mobility and 79 (2.3) for Social Function, indicating moderate-to-good identical scores. Further, all ICC values ranged from 0.87 to 1.00 for test-retest reliability (Table 4).

For test-retest reliability, the mean percentage (SD) of identical scores for the Modifications scale was 95 (0.7) for the domain of Self-care, 98 (0.6) for Mobility and 96 (0.4) for Social Function, indicating excellent identical scores. The ICC values were 0.90 for Self-care, 0.96 for Mobility and 0.96 for Social Function. Agreement of the two assessments was good or excellent in all domains of the PEDI-NL-CVI used for children with CP and CVI.

For inter-respondent reliability, the mean percentage (SD) of identical scores for the Functional Skills scale was 21 (2.6) for the domain of Self-care, 76 (0.9) for Mobility and 23 (2.2) for Social Function, indicating poor-to-good identical scores.

For inter-respondent reliability, the mean percentage (SD) of identical scores for the Caregiver Assistance scale was 40 (1.8) for the domain of Self-care, 43 (5.9) for Mobility and 43 (1.2) for Social Function, indicating moderate identical scores. Further, all ICC values ranged from 0.87 to 0.99 for inter-respondent reliability (Table 5). Agreement of the two assessments was good or excellent in all domains of the PEDI-NL-CVI used for children with CP and CVI.

Cronbach's alphas for the Functional Skills scale were above 0.98 (Self-care), above 0.98 (Mobility) and above 0.97 (Social Function). Cronbach's alphas for the Caregiver Assistance scale were above 0.95 (Self-care), above 0.95 (Mobility) and above 0.88 (Social Function), indicating good or excellent internal consistency within each domain of the PEDI-NL-CVI. Cronbach's alpha for the Caregiver Assistance scale ranged from 0.94 to 0.95 (Self-care), from 0.95 to 0.96 (Mobility) and from 0.88 to 0.90 (Social Function), indicating good or excellent internal consistency within each domain of the PEDI-NL-CVI.

The mean percentage (SD) of identical scores for the Modifications scale was 71 (1.8) for the domain of Self-care, 89 (1.2) for Mobility and 86 (1.0) for Social Function. The ICC values were 0.55 for Self-care, 0.75 for Mobility and 0.75 for Social Function, indicated moderate-to-good agreement.

The ICC values indicated absolute agreement of the Modifications scale on three measurements for Self-care ranged from 0.87 to 0.90 (no modifications), from 0.85 to 0.91 (child-oriented modifications), from

Table 4. Test-retest reliability. Results of the Functional Skills (FS) and Caregiver Assistance (CA) scales. Summed scores: mean (M), standard deviation (SD), paired t-test, p-value, limits of agreement (LOA), percentage of identical scores, and ICC (CI) (n = 75).

PEDI-NL-CVI	1st interview *M* (SD)	2nd interview *M* (SD)	*t*-value	*p*-value	Mean of difference (LOA)	% identical scores: *M* (SD)	ICC (LB-UB)
FS – self-care	34 (17.5)	34 (17.3)	1.74	0.086	0.50 (±4.94)	73 (2.5)	0.99 (0.98–0.99)
FS – mobility	31 (18.5)	31 (18.6)	−1.03	0.305	−0.10 (±1.75)	76 (0.9)	1.00 (1.00–1.00)
FS – social function	37 (14.0)	37 (14.2)	−1.04	0.287	−0.10 (±2.74)	79 (1.4)	0.99 (0.99–1.00)
CA – self-care	17 (9.3)	17 (9.4)	0.43	0.665	0.06 (±2.60)	73 (1.3)	0.99 (0.98–0.99)
CA – mobility	15 (12.2)	15 (11.1)	−1.03	0.305	−0.14 (±2.48)	81 (6.0)	0.87 (0.80–0.91)
CA – social function	12 (4.7)	12 (4.9)	1.34	0.169	0.12 (±1.60)	79 (2.3)	0.88 (0.82–0.92)

ICC, intraclass correlation coefficient; LB, lower bound; UB, upper bound; CI, confidence interval.

Table 5. Inter-respondent reliability. Results of the Functional Skills (FS) and Caregiver Assistance (CA) scales. Summed scores: mean (M), standard deviation (SD), paired t-test, p-value, limits of agreement (LOA), percentage of identical scores, and ICC (CI) (n = 75).

PEDI-NL-CVI	1st interview *M* (SD)	2nd interview (mother or father *M* (SD)	*t*-value	*p*-value	Mean of difference (LOA)	% identical scores: *M* (SD)	ICC (LB-UB)
FS – Self-care	34 (17.5)	34 (17.0)	−1.99	0.050	−0.58 (±5.00)	21 (2.6)	0.99 (0.98–0.99)
FS – Mobility	31 (18.5)	31 (18.7)	−0.34	0.737	−0.80 (±4.02)	76 (0.9)	0.99 (0.99–1.00)
FS – Social Function	37 (14.0)	37 (14.2)	−1.45	0.151	0.36 (±4.21)	23 (2.2)	0.99 (0.98–0.99)
CA – Self-care	17 (9.3)	16 (9.0)	−3.97	0.000	−0.82 (±3.53)	40 (1.8)	0.98 (0.96–0.99)
CA – Mobility	15 (12.2)	15 (11.2)	−0.34	0.737	−0.08 (±2.74)	43 (5.9)	0.87 (0.81–0.92)
CA – Social Function	12 (4.7)	12 (4.8)	0.29	0.169	0.04 (±2.37)	43 (1.2)	0.97 (0.95–0.98)

ICC, intraclass correlation coefficient; LB, Lower Bound; UB, Upper Bound; CI, confidence interval.

0.23 to 0.34 (specialised rehabilitation equipment), and from 0.69 to 0.82 (extensive modifications). The ICC values indicated absolute agreement of the Modifications scale on three measurements for Mobility ranged from 0.86 to 0.92 (no modifications), from 0.91 to 0.92 (child-oriented modifications), from 0.96 to 0.97 (specialised rehabilitation equipment), and from 0.93 to 0.95 (extensive modifications). The ICC values indicated absolute agreement of the Modifications scale on three measurements for Social Function ranged from 0.94 to 0.99 (no modifications), from 0.68 to 0.71 (child-oriented modifications), from 0.83 to 0.84 (specialised rehabilitation equipment), and from 0.84 to 0.85 (extensive modifications).

4. Discussion

The aims of our study were to develop an adapted version of the PEDI-NL and to determine the inter-respondent reliability and test-retest reliability of this adapted version for children with CVI and CP. The CVI adapted version of the PEDI-NL appears to be a reliable instrument for measuring the daily skills of children with CP and CVI. The results indicate high agreement about the overall content of the adapted version of PEDI-NL. All experts agreed on the PEDI-NL-CVI for the children with CVI. From the high intraclass correlation coefficients and Cronbach's alpha's we conclude that reliability was good, not only at the scale scores but also the individual items. The results of test-retest reliability of the Functional Skills scale and the Caregiver Assistance scale indicate a good identical scores. The results of inter-respondent reliability of the Functional Skills scale and the Caregiver Assistance scale indicate poor-to-good identical scores.

4.1. Adaptation

Using the PEDI-NL-CVI for children with CVI helps measure a specific task without changing the question or instruction of original PEDI-NL. The experts suggested that 'it is assumed that the parents/caregivers use verbal and manual support to help the child perform a skill'. This is due to the fact that a child with CVI has difficulty imitating by looking, which may result in difficulty imitating actions based on visual imitation. Using verbal or manual support will help the child describe an action that occurs.

During the adaptation process, experts proposed most suggestions on the Functional Skills scale (Self-care, Mobility, Social Function) (Table 2). Less adaptation was needed on the Caregiver Assistance and Modifications scales. The experts did give suggestions for use of equipment, especially for children with CVI, and adaptation of environment such as lighting at home or at school. In line with other studies (Ghasia et al., 2008; Dutton & Jacobson, 2001; Dutton, 2013; Salavati et al., 2014; Steendam, 2007) on CVI and the experts' viewpoints, the adaptation of PEDI-NL for children with CVI was on verbal support/ instruction, manual support, types of equipment and environment. Regardless of adaptations, it remains important to register the type of visual and manual support a child with CVI needs in order to accomplish a certain task and the characteristics that materials need to have (e.g. colourful, sound-produced, high in contrast) in order to receive the child's attention using specific materials. For Self-care and Mobility, our experts suggested that children need to know in advance about positions such as a leisurely pace or a toothbrush. For transfers, the child is allowed to use the arms for orientation and to use a chair with armrests. In accordance with other investigators (Wassenberg-Severijnen et al., 2003) and our experience using PEDI-NL-CVI, therapists are not familiar enough with specific Self-care activities, Social Function skills and amount of Caregiver Assistance at home (Wassenberg-Severijnen et al., 2003). For example, several therapists found it to be difficult to answer questions about 'self-protecting' or 'functioning at home' (Functional Skills scales, Social Function domain). It is therefore important to interview the parents in order to receive reliable information about these areas of functioning.

Despite the fact that for adaptation and reliability of PEDI-NL-CVI, children included with all subtypes of CVI, the researcher should take into account which type of CVI is present. Because, CVI is quite variable in its range from no light perception to normal visual acuity, and with cognitive visual dysfunction, a disorder of visual processing that leads to misinterpretation of the visual world with respect to what objects are or where they are (Edmond & Foroozan, 2006). However we cannot ensure that all subtypes are represented. Further research is needed to determine the psychometric properties of this adapted questionnaire for children with different subtypes of CVI.

Also, a familiar environment can result in successful execution of skills, in contrast to an unknown or less familiar environment. For this reason, it is important to use the PEDI-NL-CVI to evaluate the child's level of functioning at every turn, in the same environment.

There are several limitations caused by CVI that have to be taken into account when judging daily activities. Firstly, the professionals agreed that children with CVI have limitations on recognising localisation of equipment and in areas of eye-hand, eye-foot coordination, depth perception and spatial awareness, due to the fact that visual information is not processed adequately in the brain. This could result in difficulty with 'asking for help or support', as the child may not be aware of possible levels of support. This makes it difficult for the child to indicate or find a solution. Secondly, due to difficulty in the areas of spatial orientation, a child may experience limitations in finding the road in known space or finding the toilet in a known building. Furthermore, children with CVI make contact with other children differently than by using the visual system, using instead auditory (by locating the sound) or tactile (by trying to touch) perception. Thirdly, owing to the lack of depth perception, children with CVI often have difficulty with cycling and related sub-skills, such as getting on and off the bike. Cycling is a complex skill and requires proper motor skills as well as an adequate visual perception. Fourthly, children with CVI often have difficulty completing an item within the prescribed time. Likewise, 'thoroughness of washing hands' or 'drinking without spilling' will be affected by a lack of visual perception and the extent to which such a child has been taught to deal with limited visual perception. Because, CVI results in difficulties experienced with distance viewing e.g. "looking at the clock" (Social Function, time orientation), the child is allowed to use his/her (adapted) watch to check time. Lastly, doing puzzles is difficult for the child when one single colour is used for different images of the puzzle. To this end, the professionals gave some suggestions for use of adapted puzzles for children with CVI. The severity of CVI can be determined by the amount of support needed/given. In turn, a higher degree of assistance may explain the presence of the CVI. In future studies it will be important to determine the relation between the current adaptation of PEDI-NL-CVI and different types of visual impairment.

4.2. Test-retest reliability and inter-respondent reliability

Reliability of PEDI-NL (Wassenberg-Severijnen et al., 2003) is above 0.90 on a scale score for all ICC's and Cronbach's alpha above 0.88 for all domains, indicating good to excellent reliability within each domain of the PEDI-NL-CVI. This is comparable with de original study of PEDI-NL (Wassenberg-Severijnen et al., 2003). Inter-respondent reliability of the Modifications scale indicated good ICC values for all domains, except for Self-care (0.55). This could be due to the presence and use of equipment in different environments of a child, such as the home or school. However, an adapted environment may result in more use of equipment compared to an environment that is not adapted to that specific child. Table 5 shows a t-value of 3.97 ($p = 0.000$) for Self-care (Caregiver Assistance scale). However, clinically speaking the size of the difference (1.0) between the first (mean 17) and second (mean 16) interview seems small. This makes the outcome of Self-care acceptable.

In our study, the results on percentage of identical scores do not always match with those on ICC. A reason for this is that the level of agreement is close to perfect but not completely identical. If for instance, the agreement is based on 2 points above or below zero, then the percentage identical score for inter-respondent reliability, domain Self-care (Functional Skills scale) is 81 instead of 21 (mean: 0.58 and SD: 2.56), and for domain Social Function (Functional Skills scale) is 89 instead of 23 (mean: 0.36 and SD: 2.15).

The Bland and Altman plots in Figs. 2 and 3 illustrate variations around the zero line. Except for a few values, these plots demonstrate roughly equal distribution 2 points above and below the zero line for domain Self-care (Functional Skills scale) and Social Function (Functional Skills scale) with regard to inter-respondent reliability. The few higher and lower values (for Self-care SD: 5.00 and for Social Function SD: 4.21) are the scores of parents. If we compare the scores of caregiver for Self-care (Functional Skills scale) and Social Function (Functional Skills scale), with regard to test-retest reliability, the scores of the same subjects are 2 points above or below zero.

On an item score for the Functional Skills scale the PEDI-NL (Wassenberg-Severijnen et al.,

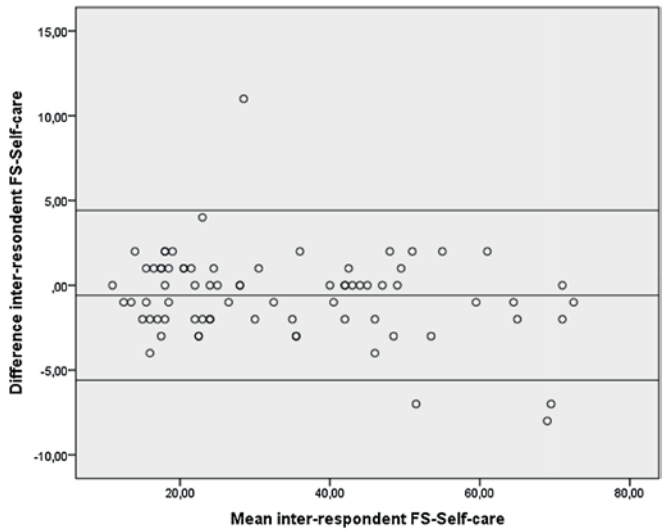


Figure 2. Bland and Altman plot for inter-respondent reliability of functional skills (FS), domain self-care. The mean difference is 0.58 5.00 (LOA) (5.59; 4 41).

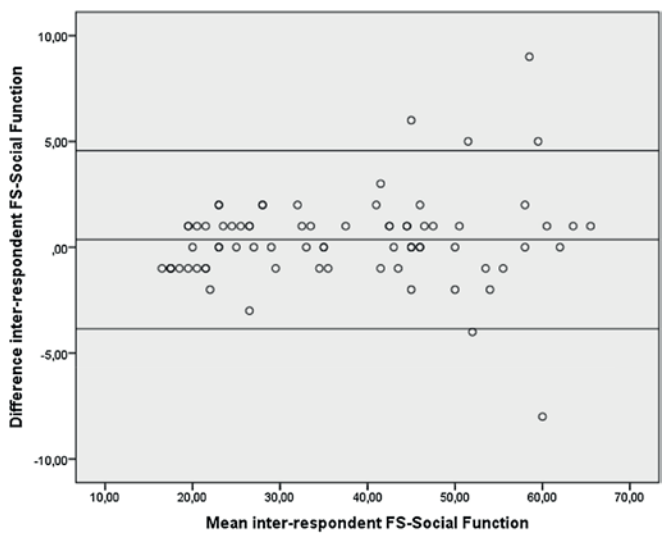


Figure 3. Bland and Altman plot for inter-respondent reliability of functional skills (FS), domain social function. The difference is 0.36 4.21 (LOA) (3.85; 4.57).

2003) shows that the mean percentage identical scores were moderate to excellent. The current study of PEDI-NL-CVI indicates moderate-to-good mean percentage identical scores for test-retest reliability, and poor-to-good mean percentage identical scores for inter- respondent reliability. In accordance with other investigators (Wassenberg-Severijnen et al., 2003), when using the PEDI-NL-CVI for children with CVI it is important to interview the same parent or caregiver to ensure that differences between two measures are the result of real changes in functional status rather than the result of differences in judgement between parents or caregivers (Wassenberg-Severijnen et al., 2003).

The results of mean percentage identical scores for the Modifications scale ranged from 0.23 to 0.34 on three measurements for Self-care (specialised rehabilitation equipment), indicating poor mean percentage identical scores. This may be caused by differences in the presence of specialised rehabilitation equipment between school and home.

Our study included children with different types of CP and various degrees of severity, who might have different profiles of functional limitations in daily life. However, most participants were children with spastic CP. The reliability of the PEDI-NL-CVI could be investigated further in the group of children with different types of CP. The PEDI-NL (Wassenberg-Severijnen et al., 2003) is used for disabled children with various diagnoses, aged between 7 and 88 months. The current study investigated the PEDI-NL-CVI for children with CVI and CP, aged between 50 and 144 months, as children with CP take longer before they achieve a higher level of functional skills. We therefore assessed the PEDI-NL-CVI in a group of older children with CP and CVI.

5. Conclusion

The adapted version of PEDI-NL is a useful and reliable instrument for professionals who work with children with CP and CVI to measure functional performance in self-care, mobility and social function in these children.

Acknowledgements

We gratefully thank the children's parents, caregivers and therapists for their participation. Financial support for this study was given by the Novum Foundation (OI0323), a Dutch nonprofit organisation providing financial support to (research) projects that improve the quality of life of individuals with visual impairments (www.stichtingnovum.org).

References

1. Cronbach, L. J. (1951). Coefficient alpha and the internal structure of tests. Psychometrika, 16, 297-334.
2. Custers, J. W. H., Wassenberg-Severijnen, J. E., Net Jvd, Vermeer, A., Hart, H. T., & Helders, P. J. M. (2002). Dutch adaptation and content validity of the 'Pediatric Evaluation of Disability Inventory (PEDI)'. Disability and Rehabilitation, 24, 250-258.
3. Da Costa, M. F., Salmao, S. R., Berezovsky, A., De Haro, F. M., & Ventura, D. F. (2004). Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. Behavioural Brain Research, 149(2), 145-150.
4. Dutch Institute of Rehabilitation Paediatricians (2007). Guidelines for the diagnosis and treatment of children with spastic cerebral palsy, Utrecht, the Netherlands. International classification of functioning, disability and health, child and youth version (2008). (1st ed.). The Netherlands: Dutch WHO-FIC Collaborating Centre, Bohn Stafleu van Loghum www.bsl.nl (in Dutch).
5. Dutton, G. N. (2013). The spectrum of cerebral visual impairment as a sequel to premature birth: An overview. Documenta Ophthalmologica, 127(1), 69-78. Dutton, G. N., & Jacobson, L. K. (2001). Cerebral visual impairment in children. Seminars in Neonatology, 6, 477-485.
6. Edmond, J., & Foroozan, R. (2006). Cortical visual impairment in children. Current Opinion in Ophthalmology, 17, 509-512.
7. Fazzi, E., Signorini, S. G., Piana, L. A., Bertone, R., Misefari, C., Galli, W., et al. (2012). Neuro-ophthalmological disorders in cerebral palsy: Ophthalmological, oculomotor, and visual aspects. Developmental Medicine and Child Neurology, 54, 730-736.
8. Ghasia, F., Burnstroom, J., Gordon, M., & Tychsen, L. (2008). Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. Investigative Ophthalmology and Visual Science, 49, 572-580.
9. Good, W. V., Jan, J. E., Burden, S. K., Skoczenski, A., & Candy, R. (2001). Recent advances in cortical visual impairment. Developmental Medicine & Child Neurology, 43(1), 56-60.
10. Haley, S. M., Coster, W. J., Ludlow, L. H., Haltiwanger, J. T., & Andrellos, P. J. (1992). Pediatric Evaluation of Disability Inventory: Development, standardization, and administration manual. Boston, MA: New England Medical Centre Inc/PEDI Research Group.
11. Lexell, J. E., & Downham, D. Y. (2005). Howe to assess the reliability of measurements in rehabilitation. American Journal of Physical Medicine and Rehabilitation, 84, 719-723.
12. McGraw, K. O., & Wong, S. P. (1996). Forming inferences about some intraclass correlation coefficients. Psychological Methods, 1, 30-46. Powell, C. (2003). The Delphi technique: Myths and realities. Journal of Advanced Nursing, 41(4), 376-382.
13. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M., & Bax, M. (2007). Definition and classification of cerebral palsy. Developmental Medicine and Child Neurology, 49(6), 480.
14. Salavati, M., Rameckers, E. A. A., Steenbergen, B., & Schans van der, C. (2014). Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). European Journal of Physical Therapy, 16(3), 159-167.
15. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Van der Graaf, Y., Wittebol-Post, D., & Willemse, J. (1993). The prevalence of cerebral visual disturbance in children with cerebral palsy. Developmental Medicine and Child Neurology, 34, 473-480.
16. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Schiemanck, N., Van der Graaf, Y., & Willemse, J. (1993). Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. Child: Care, Health and Development, 19, 411-423.
17. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Van Waes, P. F. G. M., & Van der Graaf, Y. (1994). Cerebral visual impairment in cerebral palsy: Relation to structural abnormalities of the cerebrum. Developmental Medicine and Child Neurology, 25, 68-72.
18. Steendam, M. (2007). Do you know what I see? Cerebral visual impairment in children: A manual for professionals The Netherlands: Royal Visio Huizen ISBN: 9789077680124.
19. Stiers, P., Vanderkelen, R., Vanneste, G., Coene, S., De Rammelsere, M., & Vandenbussche, E. (2002). Visual-perceptual impairment in a random sample of children

with cerebral palsy. Developmental Medicine and Child Neurology, 44, 370-382.
20. Verrel, J., Bekkering, H., & Steenbergen, B. (2008). Eye-hand coordination during manual object transport with the affected and less affected hand in adolescents with hemiparetic cerebral palsy. Experimental Brain Research, 187, 107-161.
21. Visser, L., Ruiter, A. J., Meulen van der, F., Ruijssenaars, A. J. J. M., & Timmerman, E. (2014). Validity and suitability of the Bayley-III Low Motor/Vision version: A comparative study among young children with and without motor and/or visual impairments. Pediatric Physical Therapy, 26(1), 57-67.
22. Wassenberg-Severijnen, J. E., & Custers, J. W. H. (2005). Pediatric Evaluation of Disability Inventory-NL. Amsterdam, The Netherlands: Harcourt Assessment BV. Wassenberg-Severijnen, J. E., Custers, J. W., Hox, J. J., Vermeer, A., & Helders, P. J. (2003). Reliability of the Dutch Pediatric Evaluation of Disability Inventory (PEDI). Clinical Rehabilitation, 17, 457-462.

4

Reliability of the modified Gross Motor Function Measure-88 (GMFM-88) for children with both Spastic Cerebral Palsy and Cerebral Visual Impairment: A preliminary study

M. Salavati[a,b], W.P. Krijnen[b], E.A.A. Rameckers[c,d,e], P.L. Looijestijn[f,g], C.G.B. Maathuis[h], C.P. van der Schans[b,h], B. Steenbergen[i,j]

[a] Royal Dutch Visio, Centres of Expertise for Blind and Partially Sighted People, The Netherlands

[b] Hanze University of Applied Sciences Groningen, Research Group Healthy Ageing, Allied Health Care and Nursing, Groningen, The Netherlands

[c] Maastricht University, Department of Rehabilitation Medicine, School for Public Health and Primary Care (CAPHRI), Maastricht, The Netherlands

[d] Adelante Center of Expertise in Rehabilitation and Audiology, Valkenburg and Hoensbroek, The Netherlands

[e] AVANSplus, Advanced Training for Professionals Working with Children with Movement Disabilities, Breda, The Netherlands f University of Groningen, Special Needs Education & Youth Care, Groningen, The Netherlands

[f] University of Groningen, Special Needs Education & Youth Care, Groningen, The Netherlands

[g] De Noorderbrug, Centre for Persons with Disabilities and Chronic Illnesses, Groningen, The Netherlands

[h] University of Groningen, University Medical Center Groningen, Department of Rehabilitation Medicine, Groningen, The Netherlands i Radboud University Nijmegen, Behavioural Science Institute, Nijmegen, The Netherlands

[i] Radboud University Nijmegen, Behavioural Science Institute, Nijmegen, The Netherlands

[j] Australian Catholic University, School of Psychology, Melbourne, Australia

Published as:

Salavati, M., Krijnen, W.P., Rameckers, Looijestijn, P.L., Maathuis, C.G.B., Schans van der, C.P. & Steenbergen, B. (2015). Reliability of the modified Gross Motor Function Measure-88 (GMFM-98) for children with both Spastic Cerebral Palsy and Cerebral Visual Impairment: a preliminary study. Res Dev Dis, 45-46, 32-48.

Abstract

Purpose: The aims of this study were to adapt the Gross Motor Function Measure-88 (GMFM-88) for children with Cerebral Palsy (CP) and Cerebral Visual Impairment (CVI) and to determine the test-retest and interobserver reliability of the adapted version.

Method: Sixteen paediatric physical therapists familiar with CVI participated in the adaptation process. The Delphi method was used to gain consensus among a panel of experts. Seventy-seven children with CP and CVI (44 boys and 33 girls, aged between 50 and 144 months) participated in this study. To assess test-retest and interobserver reliability, the GMFM-88 was administered twice within three weeks (Mean = 9 days, SD = 6 days) by trained paediatric physical therapists, one of whom was familiar with the child and one who wasn't. Percentages of identical scores, Cronbach's alphas and intraclass correlation coefficients (ICC) were computed for each dimension level.

Results: All experts agreed on the proposed adaptations of the GMFM-88 for children with CP and CVI. Test-retest reliability ICCs for dimension scores were between 0.94 and 1.00, mean percentages of identical scores between 29 and 71, and interobserver reliability ICCs of the adapted GMFM-88 were 0.99-1.00 for dimension scores. Mean percentages of identical scores varied between 53 and 91. Test-retest and interobserver reliability of the GMFM-88-CVI for children with CP and CVI was excellent. Internal consistency of dimension scores lay between 0.97 and 1.00.

Conclusion: The psychometric properties of the adapted GMFM-88 for children with CP and CVI are reliable and comparable to the original GMFM-88.

1. Introduction

Cerebral Palsy (CP) describes a group of permanent disorders of movement and posture development that cause activity limitations; they are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. Gross motor function of children with CP is classified into five different severity levels using the Gross Motor Function Classification System (GMFCS), where level 1 indicates the least and level 5 the most functional limitation (Rosenbaum, Paneth, Leviton, Goldstein, & Bax, 2007). Motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour; by epilepsy; and by secondary musculoskeletal problems (Rosenbaum et al., 2007).

The spectrum of visual impairments in children with CP is broad and includes forms of ocular visual impairment (OVI) such as strabismus, reduced visual acuity, ocular nystagmus, refraction disorders and retinopathies; and Cerebral Visual Impairment (CVI), which is a problem of central origin. CVI is observed in approximately 30% of children diagnosed with various forms of CP (Da Costa, Salmao, Berezovsky, De Haro, & Ventura, 2004; Dutton & Jacobson, 2001; Ghasia, Burnstroom, Gordon, & Tychsen, 2008; Schenk-Rootlieb, Van Nieuwenhuizen, Van der Graaf, et al., 1993; Schenk-Rootlieb, Van Nieuwenhuizen, Schiemanck, et al., 1993; Schenk-Rootlieb, Van Nieuwenhuizen, Van Waes, & Van der Graaf, 1994; Stiers et al., 2002). CVI can be defined as deficient visual function, as a sequel of damage or malformation of the retrogeniculate visual pathways (optic radiations, occipital cortex and visual association areas) in the absence of damage to the anterior visual pathways or any major ocular disease. CVI is diagnosed by exclusion of OVI (Dutton & Jacobson, 2001; Dutton et al., 2004), and ranges in severity from blindness to relatively minor impairments of visual perception. Perceptual visual dysfunction and disorders of visual attention, often with only minimally reduced or normal visual acuities, are increasingly recognised as forms of CVI (Dutton, 2013; Dutton & Jacobson, 2001; Fazzi et al., 2012). Children with CVI exhibit slow, inefficient and highly variable visual performance during daily-life activities (Good, Jan, Burden, Skoczenski, & Candy, 2001). It is established that CVI has an impact on all aspects of a child's development, and children with both CP and CVI develop more slowly in the areas of self-care, mobility and social function than children with CP and without CVI. (Da Costa, Salmao, Berezovsky, De Haro, & Ventura, 2004; Dutton & Jacobson, 2001; Dutton, 2013; Ghasia et al., 2008; Good et al., 2001; Salavati, Rameckers, Steenbergen, & Schans van der, 2014; Schenk-Rootlieb, Van Nieuwenhuizen, Van der Graaf et al., 1993).

The role of visual perception during motor action and development is very important because children with CP show increased visual monitoring during motor activities (reaching, walking, daily-life activities and play), and this emphasises the role of visual perception during motor action (Good et al., 2001; Guzzetta, Mercuri, & Cioni, 2001;Verrel, Bekkering, & Steenbergen, 2008; Palisano et al., 1997). Compared to their peers, children with CVI have inferior gross motor skill performance, are less physically active, and exhibit poor performance on static and slow dynamic balance tasks (Houwen, Hartman, & Visscher, 2009; Salavati et al., 2014, 2015). Early assessment and accurate detection of visual disorders is therefore of paramount importance in children with CP (Fazzi et al., 2012). Limitations in physical activities in children with CP may not be caused solely by a delay in motor or mental development but also by the presence of CVI (Salavati et al., 2014). As children with both CVI and CP experience limitations in daily activities (Da Costa et al., 2004; Salavati et al., 2014), it is important to evaluate their gross motor function.

The Gross Motor Function Measure-88 (GMFM-88) is used to measure changes in gross motor function in children with CP and has been commonly used by researchers (Chrysagis, Skordilis, Stavrou, Grammatopoulou, & Koutsouki, 2012; Scholtes et al., 2010). The GMFM-88 consists of 88 items in five dimensions: lying and rolling (GMFM-A); sitting (GMFM-B); crawling and kneeling (GMFM-C); standing (GMFM-D); and walking, running and jumping (GMFM-E). The GMFM-88 comprises 88 items, of which only seven were not found to be at the level of activities and participation of the ICF (WHO, 2001). (Engelen, Ketelaar, & Gorter, 2007) and are therefore not classified: Domain A: Lying and Rolling - item 1: Supine, head in midline: turns head with extremities symmetrically; item 3: Supine: lifts

head 45 degrees; item 4: Supine: flexes right hip and knee through full range; item 5: Supine: flexes left hip and knee through full range; item 10: Prone: lifts head upright. Domain B: Sitting - item 21: sitting on mat, supported at thorax by therapist, lifts head upright, maintains for 3 s; item 22: Sitting on mat, supported at thorax by therapist, lifts head to midline, maintains for 10 s (Engelen et al., 2007; WHO, 2001).

The reliability and validity of this test are sufficient (inter-rater reliability: ICC = 0.75-1.00; test-retest reliability: ICC = 0.96-0.99) (Engelen et al., 2007; Ketelaar, Van Petegem-van Beek, Veenhof, Visser, & Vermeer, 2003). The GMFM-88 is a criterion-referenced instrument constructed to evaluate the development of motor skills in children with CP, designed and validated for these children by using principles of classical test theory. It is used widely as a clinical and research outcome measure and there is considerable evidence of its reliability, validity and responsiveness (Avery, Russell, Raina, Walter, & Rosenbaum, 2003). The GMFM -88 is responsive to changes in motor functioning, and can be used to measure changes in fundamental gross motor skills over time in children with CP as well as evaluate physiotherapeutic intervention for these children (Engelen et al., 2007; Ketelaar et al., 2003). Importantly though, its reliability and validity for children with visual impairments is unknown (Engelen et al., 2007; Ketelaar et al., 2003). Experts working with children with both CVI and CP experienced that the GMFM-88 does not account for the presence of visual impairments - that is, it might not be an appropriate assessment method for children with CVI due to the confounding effect of visual impairments on levels of motor functioning. Given these restrictions, it is a potentially less reliable and valid measure of motor functioning for children with CP and CVI. During an assessment with the GMFM-88, the paediatric physical therapist assumes that the child with CP and CVI is visually able to locate a toy it is reaching for (GMFM-88, nos. 6, 7, 25-27). As children with CP and CVI have an inherent problem with proper identification and processing of visual information, and the original GMFM-88 does not take this aspect of functioning into account, the results on this test most likely do not reflect the true motor capacity of these children. Also, because of visual impairments a child might not be able to show its motor functioning abilities during a standardised assessment of motor development (Haley, Coster, Ludlow, Haltiwanger, & Andrellos, 1992; Salavati et al., 2014; Visser, Ruiter, Meulen van der, Ruijssenaars, & Timmerman, 2014; Wassenberg-Severijnen, Custers, Hox, Vermeer, & Helders, 2003).
The aims of this study were twofold. Firstly we set out to develop an adapted version of the GMFM-88 for children with CP and CVI, and secondly we wished to determine its test-retest and interobserver reliability.

2. Methods

The study was carried out in two phases. In the first phase the Delphi method was applied to an adaptation process of the GMFM-88 for children with CVI, and in the second phase the psychometric properties of the adapted GMFM-88 were examined.

2.1. Phase 1

2.1.1. Adaptation

The Delphi method was used to gain consensus among a panel of paediatric physical therapists. It was applied in a series of sequential questionnaires or 'rounds', interspersed by controlled feedback, in order to identify adaptations and obtain the most reliable consensus of opinion on these adaptations among a group of experts (Powell, 2003). The Delphi method was used because it is well-established for consensus-building. Delphi, in contrast to other data gathering and analysis techniques, employs multiple iterations designed to develop a consensus of opinion on a specific topic. One of the primary characteristics and advantages of the Delphi process is participant anonymity, which can reduce the effects of dominant individuals - a frequent concern when using group-based processes to collect and synthesise information (Chia-Chien & Sandford, 2007; Gracht von der, 2012).

We started by identifying all paediatric physical therapists who worked at centres of expertise for blind and partially sighted people and had experience with children with CVI, and gave them the opportunity to participate in the adaptation study. All of them worked at Royal Dutch Visio and Bartiméus, centres of expertise for blind and partially sighted people in the Netherlands. Next, we explained to the experts the purpose of this study and the required procedures. All of the invited experts agreed to participate. They were

asked for their age, profession, working experience and familiarity with the GMFM-88.

2.1.2. Data collection for adaptation

We included 16 paediatric physical therapists who had experience with CVI to participate in the adaptation process of the GMFM-88. Mean (SD) age of paediatric physical therapists was 49 (11) years and their mean years (SD) of experience with children with CP and CVI was 18 (8). In the Netherlands, paediatric physical therapists are expert clinicians who regularly use the instrument in children with CP (Salavati et al., 2015). In addition to following specialised training on how to use the GMFM-88, when required the therapists had to adapt the instructional part of the content to children with CVI. Specific feedback information could be written about any of the items. Electronic mail was used to process and resubmit all the experts' comments in three rounds.

The authors (Gracht von der, 2012; Powell, 2003) defined agreement among the majority as exceeding 50% of the respondents, in accordance with the Delphi method (Gracht von der, 2012; Powell, 2003) The predetermined goal of the Delphi method was to reach a consensus of 65% among experts after the first round, 75% after the second round and 85% after the third round on the proposed adaptations for the content of the GMFM-88. These agreement thresholds are arbitrary, but we considered that a vast majority of the experts should agree in order to reflect sufficient agreement. The experts were also asked to explain and justify their comments on each question and instruction without changing the original questions and instructions. No items were eliminated from the original GMFM-88 during the adaptation process (Engelen et al., 2007).

2.1.3. First Delphi round

All experts were invited to study the instruction part of the GMFM-88, and subsequently gave individual comments on each question and original instruction. In accordance with other studies (Dutton & Jacobson, 2001; Dutton, 2013; Ghasia et al., 2008) on CVI, the experts were asked the following specific questions: Which verbal support/instruction or manual support needs to be added to the instruction of the GMFM-88 to make it suitable for children with CVI? And at the equipment level: What kind of adjustment needs to be added to the instruction of the GMFM-88 to make it suitable it for children with CVI? For use of manual support (GMFM-88, e.g. nos. 18, 21 and 22) they were asked, if applicable, to describe the amount of manual support (e.g. duration and phase of needed manual support given) in order to help a child accomplish a specific skill. At the level of verbal support they were asked, if applicable, what the instructor should articulate in order to help a child accomplish a specific skill. For use of equipment they were asked which characteristics one needs to have (e.g. colourful, sound-produced, high-contrast) to obtain the attention of a child with CVI.

2.1.4. Second and third Delphi rounds

After receiving the experts' comments on the questions and instruction of the GMFM-88, we processed all the suggestions in the instruction part and resubmitted this twice to the experts. We asked the same questions as in the first round.

2.2. Phase 2

Children with CP and CVI were recruited from Royal Dutch Visio and allied healthcare practices. Inclusion criteria were presence of any type of CP and CVI, mild or moderate intellectual disability, and age at testing of the modified GMFM-88 for children with CVI between 4 and 12 years. Level of intellectual disability was reported from the children's medical files. Our study included children with different types of CVI, as the diagnoses from the medical files did not always specify clearly the type of CVI present. Children with a syndrome (e.g. Down syndrome), hearing difficulties (>30 db hearing loss), severe or profound intellectual disability (IQ < 40), and corrected vision <0.3 and/or field of < 30° were excluded (Fig. 1). In accordance with a study examining the Dutch translation of the GMFM-88 (Ketelaar, van Petegem-van Beek & Visser, 1995), we used inclusion criteria such as age at testing. We chose to include mild or moderate intellectual disability because profoundly intellectually disabled children with CP will have difficulty with the standardised performance of gross motor tasks. The Gross Motor Function Classification System (GMFCS) excels in categorising degree of ambulatory capacity to describe degree of activity limitation in children with CP. We used GMFCS as applied to children from ages 4 to 6 and ages 6 to 12.

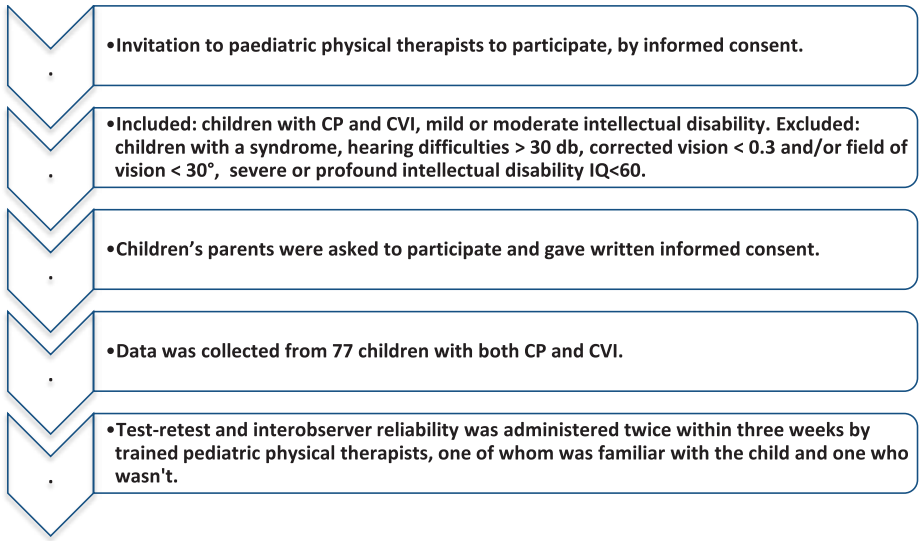


Fig. 1. Procedure for inclusion.

The diagnosis of CP and the classification according to GMFCS level were obtained from the children's medical files and assessed by a rehabilitation physician. The diagnosis of CVI was determined based on the results of ophthalmological and psychological/ neuropsychological research and on the assessment data reported by a developmental coach specialised in working with children with visual impairments, using the following criteria: a normal or near-normal eye exam (corrected vision <0.3 and/or field of vision < 30°) performed by an ophthalmologist; a history or presence of neurological problems; and presence of behavioural responses to visual stimuli that are unique to CVI. This results in strong colour preferences, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non-purposeful gaze, difficulty with distance viewing, and absent or atypical visual reflexes (Dutton & Jacobson, 2001; Stiers et al., 2002).

This study was approved by the Medical Ethical Committee (METC-2013.104) of University Medical Center Groningen (UMCG), the Netherlands. Written informed consent was obtained from the children's parents.

2.2.1. Design

Test-retest and interobserver reliability were conducted and administered twice within three weeks by trained paediatric physical therapists, one of whom was familiar with the child and one who wasn't. Cronbach's alphas and intraclass correlation coefficients (ICC) were computed at each dimension level.

2.3. Data collection

Based on possible effects on motor functioning, we also collected background data on gender as well as prevalence of epilepsy and speech/language development according to the International Classification of Functioning, Disability and Health for Children and Youth (Dutch Translation, 2008). At the level of speech/language development, the collected data were: d3100 = reacts to human voice; d3101 = understands simple spoken messages; d3102 = understands complex spoken messages; d330 = speaks; d331 = babbles; d3350 = uses body language, and d3351 = uses signs symbols (Table 2). We registered level of GMFCS and type of CP (unilateral or bilateral), level of intellectual disability and age at which the GMFM-88 was administered.

2.4. Statistical analyses

The data were analysed using Statistical Package for Social Sciences (SPSS), v.22 software. The proportion of identical answers for every item was calculated. Test-retest and interobserver reliability were established using the partial and dimensional scores of the adapted

GMFM-88 from children with CP and CVI. ICC (two-way random, absolute agreement, single-measure) and its confidence intervals were computed to assess test-retest and interobserver reliability for each dimension separately as well as for the total (McGraw, kenneth, & Wong, 1996). p-Values below 0.05 were considered statistically significant. The ICC value was interpreted as follows: poor <0.5, moderate 0.5-0.75, good 0.75-0.9, excellent >0.9. In line with the reliability studies of the GMFM-88 (Ko & Kim, 2013; Shi et al., 2006) we used ICC 0.90 as threshold for acceptable reliability. To test for systematic bias, the mean of the differences and the standard deviation of differences were calculated using the paired t-test. Limits of agreement (LOA) for test-retest reliability and interobserver reliability of five dimensions (lying and rolling (GMFM-A); sitting (GMFM-B); crawling and kneeling (GMFM-C); standing (GMFM-D); and walking, running, jumping (GMFM-E) were calculated. Internal consistency of each domain of adapted GMFM-88 was assessed by Cronbach's alpha based on the paediatric physical therapist's measurement. The Cronbach's alpha value (Cronbach, 1951) was interpreted as: poor <0.5, moderate 0.5-0.75, good 0.75-0.9, excellent >0.9.

3. Results

3.1. Phase 1

3.1.1. Adaptation

Sixteen paediatric physical therapists (male:female = 2:14), familiar with children with CVI participated in the adaptation process of the GMFM-88. Mean (SD) age of the experts was 49 (11) years and mean (SD) years of experience with children with CVI was 18 (9) years. All the therapists were familiar with the GMFM-88.

3.1.2. Delphi first round

Seventy percent of the experts agreed on the instruction part of the GMFM-88. Most proposed adaptations of the original instruction were on the 'crawling and kneeling' (GMFM-C) and 'walking, running, jumping' (GMFM-E) dimensions. The experts disagreed most on the GMFM-E dimension, suggesting that the higher complexity of motor functions such as jumping, the more the need for visual support to perform them. They also suggested that limitation of depth perception results in difficulties at the level of jumping from a certain height (GMFM-88, no. 80), so we added an extra instruction to the GMFM-C and GMFM-E dimensions.

The experts agreed most on the dimensions of lying and rolling (GMFM-A), sitting (GMFM-B) and standing (GMFM-D). They suggested that when 'reaching for the toy', (GMFM-88, nos. 6 and 7) a child with CVI could have difficulty visually localising an object, therefore the child should be told of the toy's location in advance.

The experts suggested that during use of the adapted version of the GMFM-88 for children with CVI, the paediatric physical therapists needed to be conscious of their own body position while they invite the child to move. Hence during motions like rolling over (GMFM-88, nos. 8 and 9), it is important to be positioned on the side towards which the child will be rolling (Table 1). Finally, on the GMFM-E dimension the experts agreed that during motions like independent walking or climbing stairs it was allowed to use the hands for orientation (GMFM-88, no. 85, e.g. touching the stairs and the rails to be aware of the stairs' height and the position of the rails) but not for supporting purposes (e.g. holding on to the rails of the stairs).

3.1.3. Delphi second round

Eighty percent of the experts agreed on the instruction part of the GMFM-88 by commenting on this part after the second round. Most comments were about the walking, running, jumping (GMFM-E) dimension. At this level, in terms of inconsistency of visual perception, the child could have difficulty with something like 'running' (GMFM-88, no. 77) in an unfamiliar environment. An additional explanation is given of these items. Generally, when a task is more complex, such as running or jumping, the child will have more difficulty performing that particular task. For example, the child may have difficulty 'stepping over a stick at knee level' (GMFM-88, nos. 75 and 76). The child could also have difficulty 'walking forward between two parallel lines'. A child with CVI will have difficulty 'kicking a ball with the foot' (GMFM-88, nos. 78 and 79). For these reasons, the experts agreed to use verbal support when explaining the content of visual information to children with CVI (Table 1).

Table 1. Summary of changes proposed by experts.

Adaptation Item	Dimension A: lying and rolling
1	Sit on the side that the head of the child should turn towards. During the exercise use both manual and verbal support (e.g. researcher's voice) to ensure that the child turns its head sideways. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast.
2	Practice the movement together with the child for the first time. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast. Invite the child to move its hands to the toys that are held in its midline.
3	During the practice phase exercise the movement together with the child. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast. During the exercise use verbal support (e.g. researcher's voice).
4	During the practice phase exercise the movement together with the child. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast (e.g. use bells attached to ankles or colourful socks).
5	During the practice phase exercise the movement together with the child. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast.
6–7	Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice).
8–9	Sit on the side the child should roll towards. Use manual and verbal support (e.g. researcher's voice) during the practice phase to invite the child to roll towards a side. Use toys that have moving parts, produce sound and/or are fluorescent/high-contrast.
10	During the practice phase use manual support to lift the head of the child (note that the head is often sensitive to being touched); the child then has to able to be visually/aurally attracted by a toy or a familiar face (e.g. face of a parent). For safety reasons sit close to the child. Use verbal support (e.g. researcher's voice) to invite the child to lift its head. Use toys that move, produce sound and/or are fluorescent/high-contrast/reflective or have lights.
11	During the practice phase use manual support to stretch the arm of the child, thereafter the child should focus aurally and visually on the toy. Use toys that move, produce sound and/or are fluorescent/high-contrast/reflective or have lights.
12	During the practice phase use manual support to reach for the toy and to focus both aurally and visually on the toy. During the practice phase stimulate the child to touch the toy with its left hand. Use toys that move, produce sound and/or are fluorescent/high-contrast/reflective or have lights.
13	During the practice phase use manual support to reach for the toy and to focus both aurally and visually on the toy. During the practice phase stimulate the child to touch the toy with its right hand. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast/reflective.
14–15	Sit on the side the child should roll towards. During the practice phase use manual and verbal support (e.g. researcher's voice) to invite the child to roll towards a side. Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast. If necessary, practice the movement together for the first time.
16–17	Sit on the side the child should pivot towards. During the exercise phase use manual and verbal support (e.g. researcher's voice) to invite the child to pivot. Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast.
Item	Dimension B: Sitting
18	Practice this beforehand by giving manual and verbal instructions.
19–20	Sit on the side the child should roll towards before sitting. Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast.
21	Use a second person (e.g. a parent); sit in front of the child if it mainly focuses on the other person. Use objects if the child mainly focuses on them. Use reflective, big, moving, sound-producing and/or fluorescent, high-contrast toys that have lights. Use verbal support (e.g. researcher's voice) to focus the child on the toy so that the child will lift its head.
22	Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice) to let the child focus either on the toy or the researcher's head so that the child will lift its head.
23	Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice) to let the child focus either on the toy or the researcher. Practice the support function with the child beforehand.
24	Use toys that have lights, moving parts, produce sound, and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice) to focus the child on the toy. If needed, invite the child to reach for the toy without holding it. If needed, place the child in a favourable position and invite the child to hold this position.
25	Use verbal support (e.g. researcher's voice) to focus the child's attention on a toy that has lights, moving parts, produces sound, and/or is fluorescent/high-contrast. First practice the 'touch the toy' movement together with the child so it knows where the toy is. Avoid eye contact with children who focus on faces by looking at the toy at the same time.
26	Use verbal support (e.g. researcher's voice) to focus the child's attention on a toy that has lights, moving parts, produces sound, and/or is fluorescent/high-contrast. First practice the 'touch the toy' movement together with the child so it knows where the toy is. During the practice phase the child is allowed to touch its right hand (stroke the toy across the right hand/arm) with the toy so that it will subsequently reach for the toy.
27	Use verbal support (e.g. researcher's voice) to focus the child's attention on a toy that has lights, moving parts, produces sound, and/or is fluorescent/high-contrast. First practice the 'touch the toy' movement together with the child so it knows where the toy is. During the practice phase the child is allowed to touch its right hand (stroke the toy across the right hand/arm) with the toy so that it will subsequently reach for the toy with the goal of 'touching' it.
28	If the child possesses a sufficient balance response, one can chose to let it hold with both hands a toy that has lights, moving parts, produces sound, and/or is fluorescent/high-contrast to stimulate it to sit with its arms 'loose'. If needed, put the child in the ideal position and motivate it to hold the position.
29	If the child possesses a sufficient balance response, one can chose to let it hold with both hands a toy that has lights, moving parts, produces sound, and/or is fluorescent/high-contrast to stimulate it to sit without use of arm or hand support. If needed, put the child in the ideal position and motivate it to hold the position.
30	Beforehand, perform the movement together with child; use verbal and manual support. Use toys with lights that move/produce sound and are fluorescent/high-contrast to invite the child to lie down on its abdomen.
31	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to invite the child to change its position.
32	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to lure the child.

Table 1. Continued.

33	In the original instructions it is noted to move the toy in the child's field of vision. If the child has field-of-vision problems, use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. This makes the toy visible to the child and will stimulate the child to pivot. Move the toy slowly. During the practice phase use verbal and manual support to make sure that the child understands what is expected from it and how the movement must be exercised.
34	Use verbal support to ensure that the child remains sitting. If possible, put a mirror in front of the child or offer a toy with lights out of the child's range. Sit in front of the child (below eye height) if the child focuses on the researcher's face.
35	Position the child so that it can touch/feel the couch. Let the child touch the couch with the rear of its legs. Exercise the movement together with the child during the practice phase. Use verbal support.
36–37	Position the child so that it can touch the small couch beforehand. During the practice phase exercise the movement together with the child. If needed, demonstrate 'sitting on the small couch'. Use verbal support.
Item	Dimension C: crawling and kneeling
38	Use toys with lights that move, produce sound and/or are fluorescent/high-contrast. Use verbal (e.g. researcher's voice or calling) and manual support during the practice phase to convince the child to creep forward. Invite the child to creep towards you by using verbal stimulation. Possible use of a tunnel needs to be practiced with the child beforehand.
39	Invite the child to focus on a toy that has lights, moves, produces sound and/or is fluorescent/high-contrast. Sit in front of the child and use verbal stimulation (e.g. researcher's voice or calling) and manual support to keep the child in its position.
40–43	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. Use manual and verbal support (e.g. researcher's voice or calling) during the practice phase.
44	During the practice phase use verbal support (e.g. researcher's voice or calling) to invite the child to move towards you. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. Start the exercise by placing the child close to you, gradually increasing the distance to 1.8 m.
45	During the practice phase use manual and verbal support (e.g. researcher's voice) to invite the child to move towards you. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast.
46	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to invite the child to climb up. If needed, use a second person towards whom the child can crawl. Use verbal support. During the practice phase present the stairs to the child.
47	When the child goes down the stairs backwards by crawling, for safety reasons the researcher goes down the staircase behind the child, facing it. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to persuade the child to go down the stairs. Use manual and verbal support (e.g. researcher's voice or calling). During the practice phase present the stairs to the child.
48	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to persuade the child to climb up. Use verbal support (e.g. researcher's voice or calling).
49	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to invite the child to climb up. Use verbal support (e.g. researcher's voice or calling). If needed, sit in front of the child and practice this movement and position with it.
50	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to invite the child to climb up. During the practice phase use manual and verbal support (e.g. researcher's voice or calling).
51	Sit in front of the child. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to persuade the child to 'walk' towards you on its knees. Use manual and verbal support (e.g. researcher's voice or calling) during the practice phase.
Item	Dimension D: Standing
52	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to persuade the child to rise. Use verbal support (e.g. researcher's voice or calling). During the practice phase use manual support to make the child aware of the presence of the big couch. Practice this item multiple times together with the child.
53	Use verbal support (e.g. researcher's voice or calling). During the practice phase make the child aware of the presence of objects such as the treatment couch so that the child can use this as support. During the test phase the aim is to let the child stand up without any support. For safety reasons it is important that the child knows your position with respect to it.
54–55	Use verbal support. During the practice phase make the child aware of the presence of the big couch. Let the child experience 'raising its feet', manually supported by the researcher. For safety reasons it is important that the child knows your position with respect to it.
56	Use verbal support. During the practice phase make the child aware of its position towards you. Place yourself in the position that makes the child feel most comfortable: in front or behind the child.
57–58	Use verbal support. During the practice phase make the child aware of its position towards you. Let the child experience 'raising its feet' manually, supported by the researcher. Place yourself in the position that makes the child feel most comfortable: in front or behind the child.
59	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast to invite the child to rise its feet. Use verbal support (e.g. researcher's voice). During the practice phase use manual support and conduct a trial of the movement together with the child.
60–61	Use verbal support (e.g. researcher's voice or calling). During the practice phase make the child aware of its position towards you. Place yourself in a position that makes the child feel most comfortable: in front or behind the child. Conduct a series of trial exercises during the practice phase.
62	Use verbal support (e.g. researcher's voice or calling). Make the child aware of its position towards you. Conduct a series of trials during the practice phase.
63	Use verbal support (e.g. researcher's voice or calling). Make the child aware of its position towards you. Place yourself in a position that makes the child feel safe: in front or behind the child. Conduct a series of trial exercises during the practice phase.
64	Use toys that have lights, move (to catch the child's attention), produce sound and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice or calling). Make the child aware of its position towards the toy and towards you. Conduct a series of trials during the practice phase.
Item	Dimension E: walking, running and jumping
65–66	Use toys that have lights, move (to catch the child's attention), produce sound and/or are fluorescent/high-contrast. During the practice phase use manual and verbal support (e.g. your voice or calling). Make the child aware of its position towards you. Conduct a series of trials during the practice phase.
67	Use verbal support (e.g. your voice or calling) to stimulate the child. If needed, the researcher can walk in front of the child while inviting it to walk towards the researcher.

Table 1. Continued.

68	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice or calling) to stimulate the child. The researcher walks backwards and invites the child to move towards him/her. Conduct a series of trials during the practice phase.
69	Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. Use verbal support (e.g. researcher's voice or calling). Make the child aware of its position towards you; the researcher should stay close to the child during this exercise. The researcher walks backwards and invites the child to move towards him/her. Conduct a series of trials during the practice phase.
70	Use an attractive goal for the child. Let the child walks towards you. Use toys that have lights, move, produce sound and/or are fluorescent/high-contrast. Use manual and verbal support (e.g. your voice or calling) during the practice phase. Make the child aware of its position towards you. Conduct a series of trials during the practice phase.
71	Use manual and verbal support (e.g. your voice or calling) during the practice phase. Make the child aware of its position towards you. Conduct a series of trials during the practice phase. Stand behind the child to give it confidence.
72	Use verbal support. Ask the child to walk towards you. Conduct a series of trials during the practice phase. Keep in mind that when children with CVI carry a large object they can have extra balance disturbances.
73	Use verbal support (e.g. researcher's voice or calling). The two parallel-placed strips have to be touchable and high-contrast so that the child can walk on these strips barefoot. Stand next to the child during the practice phase and ask it to take ten steps without holding on to you. Conduct a series of trials. If needed, use a white cane during these trials; the cane should only be used for orientation purposes. Use thicker parallel strips for better palpability. Keep in mind that children with CVI can have more difficulties with their balance.
74	Use verbal support (e.g. researcher's voice or calling). The high-contrast lines on the floor ought to be felt so that the child can walk on them barefooted. Let the child touch the strips on the floor beforehand. Conduct a series of trials. If needed, use a white cane during these trials; the cane should only be used for orientation purposes.
75–76	Use verbal support (e.g. researcher's voice or calling). Before stepping over the stick, invite the child to touch the stick with its hands to be aware of its high position. Conduct a series of trials.
77	Use verbal support (e.g. researcher's voice or calling). Stand next to the child during the practice phase and ask the child to run without holding on to your hand. Conduct a series of trials. Invite the child to run to you, while you stand 4.5 m away from it.
78	Use verbal support (e.g. researcher's voice or calling). The child puts the ball in front of its right foot. Use balls that produce sound and/or are fluorescent/high-contrast. Conduct a series of trials.
79	Use verbal support (e.g. researcher's voice or calling). The child puts the ball in front of its left foot. Use balls that produce sound and/or are fluorescent/high-contrast. Conduct a series of trials.
80	Use verbal support (e.g. researcher's voice or calling). Conduct a series of trials by standing in front of the child. The child supports its hands on the hands of researcher without holding on to them. During the practice phase, if required the researcher can jump simultaneously with the child to make the child aware of this item. During practice use of a trampoline is allowed to make the child aware of this item.
81	Use verbal support (e.g. researcher's voice or calling). Conduct a series of trials by standing in front of the child. Use thicker strips for better palpability. During the practice phase, to make a jump forward the child supports its hands on the hands of researcher without holding on to them.
82–83	Use verbal support (e.g. researcher's voice or calling). Conduct a series of trials by standing in front of the child. Use thicker strips for better palpability to hop on the right foot. During the practice phase, the child supports its hands on the hands of researcher without holding on to them.
84	Use verbal support (e.g. researcher's voice or calling). During the practice phase, invite the child to touch the stairs and the rails to be aware of the stairs' height and the position of the rails. Conduct a series of trials by standing behind the child. If required, a second researcher can stand in front of the child to invite it during the walk-up. It is assumed that the child is familiar with the stairs.
85	Use verbal support (e.g. researcher's voice or calling). During the practice phase, invite the child to touch the stairs and the rails to be aware of the stairs' height and the position of the rails. Conduct a series of trials by standing in front of the child to invite it during the walk-down. It is assumed that the child is familiar with the stairs.
86	Use verbal support (e.g. researcher's voice or calling). During the practice phase, invite the child to touch the stairs to be aware of the stairs' height. Conduct a series of trials by standing behind the child. If required, a second researcher can stand in front of the child to invite it during the walk-up. It is assumed that the child is familiar with the stairs.
87	Use verbal support (e.g. researcher's voice or calling). During the practice phase, invite the child to touch the stairs to be aware of the stairs' height. Conduct a series of trials by standing in front the child to invite it during the walk-down. It is assumed that the child is familiar with the stairs.
88	Use verbal support (e.g. researcher's voice or calling). During the practice phase, invite the child to touch the stairs to be aware of the stairs' height. It is assumed that the child is familiar with the stairs. During the practice phase it is allowed to provide manual support to the child.

3.1.4. Delphi third round

After receiving the experts' comments we processed the proposed adaptations, mostly on the walking, running, jumping (GMFM-E) dimension in the instruction part of GMFM-88, and resubmitted to the experts. The experts responded on the adapted version of GMFM-88 (GMFM-88-CVI), indicating 100% agreement with the resulting overall content. For CVI the experts applied specific feedback information, which is added as an appendix to the instruction of the original GMFM-88 (Table 1). Most of the adjustments related to higher motor skills such as jumping, climbing stairs and cycling (Table 1). The experts also suggested that the equipment used needed to be colourful, sound-producing, and high in contrast, in order to obtain the attention of the child who is to move towards the material. In line with the original GMFM-88, during the testing stage it is allowed to provide manual and verbal support in order to enable the child to perform a motor skill. The experts agreed that children with CVI have limitations in the areas of eye-hand and eye-foot coordination, depth perception and spatial awareness, as visual information is not processed adequately in their brains. This could affect tasks such as 'kicking a ball with the foot' (GMFM-88, nos. 78 and 88) or 'standing on a 15-cm step, jumping off with both feet

Table 2. Characteristics of children with both CP and CVI.

Characteristic	Children with CP and CVI
Age in months, mean (SD), min–max	114 (29), 50–144,
Gender, male/female (*n*) (%)	44 (57)/33 (43)
Type of cerebral palsy:	
Spastic (*n*) (%)	74 (96)
Dyskinetic (*n*) (%)	2 (3)
Ataxic (*n*) (%)	1 (1)
GMFCS I (*n*) (%)	Bilateral 17 (22), unilateral left 2 (3), unilateral right 4 (5)
GMFCS II (*n*) (%)	6 (8) bilateral
GMFCS III (*n*) (%)	9 (12) bilateral
GMFCS IV (*n*) (%)	19 (25) bilateral
GMFCS V (*n*) (%)	20 (26) bilateral
Speech/language development:	
ICF-CY, d3100 = reacts to human voice (*n*) (%)	77 (100)
ICF-CY, d3101 = understands simple spoken messages (*n*) (%)	77 (100)
ICF-CY, d3102 = understands complex spoken messages (*n*) (%)	49 (64)
ICF-CY, d330 = speaks (*n*) (%)	46 (60)
ICF-CY, d331 = babbles (*n*) (%)	30 (39)
ICF-CY, d3350 = uses body language (*n*) (%)	50 (65)
ICF-CY, d3351 = uses signs and symbols (*n*) (%)	37 (48)
Level of intellectual disability: mild/moderate (*n*) (%)	32 (42)/45 (58)
Presence of epilepsy: yes/no (*n*) (%)	14 (18)/63 (82)
Use of epilepsy medication: yes/no (*n*) (%)	12 (16)/65 (84)
Percutaneous endoscopic gastrostomy tube feeding: yes/no (*n*) (%)	12 (16)/65 (84)

GMFCS, Gross Motor Function Classification System; n, numbers; ICF-CY, International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation).

simultaneously' (GMFM-88, no. 88). Considering the fact that visual perception contributes to the performance of motor activities, it is important to use the GMFM-88-CVI version for children with CVI.

3.2. Phase 2

3.2.1. Test-retest reliability and interobserver reliability

Children were tested twice between June 2013 and April 2014 with a mean of 9 days between test-retest. We chose to administer the test-retest within two weeks because it is highly unlikely that gross motor function of children with CP will change within such a time period. We collected data from 77 children with both CP and CVI (n = 44 boys and n = 33 girls). Table 2 shows the children's characteristics. All children with CVI were included in our study and no selection was done based on subtypes. We therefore assume that different subtypes are represented in our study.

The ICC was used to analyse the agreement between two trained paediatric physical therapists from each of two dimensions. Table 3 presents the ICC per dimension and total scores of the GMFM-88-CVI. The test-retest reliability ICCs of dimension scores were 0.94-1.00 and the mean percentages of identical scores varied from 29 to 71. Table 3 shows the lowest mean percentage of identical scores (test-retest reliability: paediatric physical therapist not familiar with the child: 29; paediatric physical therapist familiar with the child: 30) from the 'Total' percentage score of adapted GMFM-88. Interobserver reliability ICCs for the GMFM-88-CVI were 1.00-1.00 for dimension scores and the mean percentages of identical scores vary from 53 to 91 (Table 4). Test-retest and interobserver reliability of the GMFM-88-CVI for children with CP and CVI was excellent.

The internal consistency of dimension scores is between 0.97 and 1.00 (dimension A: 0.97-1.00, dimension B: 0.99-1.00, dimension C: 1.00-1.00, dimension D: 1.00-1.00, dimension E: 1.00-1.00 and Total: 1.00-1.00), so the dimensions are reliable. Tables 3 and 4 show the results of limits of agreement (LOA) for test-retest reliability.

Tables 5 and 6 present the ICCs per dimension and total scores of the GMFM-88-CVI at each GMFCS level for test-retest and interobserver reliability.

Figs. 2 and 3 illustrate the percentages of identical scores for test-retest and interobserver reliability for the 'Total' dimension: 30 (test-retest reliability: paediatric physical therapist familiar with the child) and 29 (paediatric physical therapist not familiar with the child).

GMFCS level is a determining factor of gross motor function in children with CP. Variation in patterns of gross motor function among children within each GMFCS level will result in different GMFM scores (Wood & Rosenbaum, 2000). In our

Table 3. Test-retest reliability (paediatric physical therapist who was familiar with the child). Results of adapted version of the GMFM-88-CVI. Summed scores: mean (M), standard deviation (SD), paired t-test, p-value, limits of agreement (LOA), percentages of identical scores, ICC (CI) (n = 77).

GMFM-88-CVI	Paediatric physiotherapist familiar with the child							Paediatric physiotherapist not familiar with the child						
	M (SD) Test	M (SD) Retest	*t*-Value	*p*-Value	Mean of difference (LOA)	% Identical score: (*M* (SD))	ICC (LB-UB)	M (SD) Test	M (SD) Retest	*t*-value	*p*-value	Mean of difference (LOA)	% Identical score: (*M* (SD))	ICC (LB-UB)
Lying, rolling (GMFM-A)	78 (26.9)	78 (25.8)	−0.52	0.62	−0.47 (±13.10)	47 (8.19)	0.95 (0.93–0.97)	78 (26.9)	78 (25.5)	−0.65	0.52	−0.40 (±17.04)	48 (8.98)	0.94 (0.91–0.97)
Sitting (GMFM-B)	64 (35.4)	64 (35.9)	−0.58	0.56	−0.41 (±12.29)	47 (8.19)	0.99 (0.97–0.99)	64 (35.4)	64 (35.9)	−0.57	0.57	−0.42 (±12.30)	46 (6.24)	0.99 (0.98–0.99)
Crawling, kneeling (GMFM-C)	44 (42.1)	45 (41.9)	−1.49	0.14	−1.00 (±11.57)	55 (5.91)	0.99 (0.98–0.99)	44 (42.1)	45 (42.0)	−1.35	0.18	−0.90 (±11.58)	53 (5.91)	0.99 (0.98–0.99)
Standing (GMFM-D)	35 (39.3)	36 (39.7)	−1.09	0.28	−0.723 (±11.42)	66 (5.83)	0.99 (0.98–0.99)	35 (38.9)	35 (39.2)	−0.39	0.69	−0.22 (±9.67)	65 (4.94)	0.99 (0.99–1.00)
Walking, running, jumping (GMFM-E)	29 (36.2)	30 (37.0)	−2.65	0.01	−1.02 (±6.64)	66 (3.39)	1.00 (0.99–1.00)	29 (36.5)	30 (37.2)	−2.75	0.01	−0.98 (±6.17)	71 (3.15)	1.00 (0.99–1.00)
Total	50 (32.5)	51 (32.8)	−2.27	0.03	−0.77 (±5.91)	30 (3.02)	1.00 (0.99–1.00)	50 (32.4)	51 (32.8)	−1.79	0.08	−0.68 (±6.63)	29 (3.4)	0.99 (0.99–1.00)

ICC, intraclass correlation coefficient; LB, lower bound; UB, upper bound; CI, confidence interval; GMFM-88, Gross Motor Function Measure-88.

Table 4. Interobserver reliability. Results of adapted version of GMFM-88-CVI. Summed scores: mean (M), standard deviation (SD), paired t-test, p-value, limits of agreement (LOA), percentages of identical scores, ICC (CI) (n = 77).

GMFM-88-CVI	Test							Retest						
	M (SD) First observation	M (SD) Second observation	*t*-Value	*p*-Value	Mean of difference (LOA)	% Identical score: *M* (SD)	ICC (LB-UB)	*M* (SD) First observation	*M* (SD) Second observation	*t*-Value	*p*-Value	Mean of difference (LOA)	% Identical score: M (SD)	ICC (LB-UB)
Lying, rolling (GMFM-A)	78 (26.9)	78 (26.9)	2.55	0.01	0.39 (±2.62)	91 (1.34)	1.00 (1.00–1.00)	78 (25.8)	78 (25.4)	0.45	0.65	0.45 (±5.07)	74 (3.77)	0.99 (0.98–0.99)
Sitting (GMFM-B)	64 (35.4)	64 (35.3)	1.57	0.12	0.20 (±2.27)	83 (1.16)	1.00 (1.00–1.00)	64 (35.9)	64 (35.8)	1.08	0.28	0.22 (±3.50)	86 (1.79)	1.00 (1.00–1.00)
Crawling, kneeling (GMFM-C)	44 (42.1)	44 (42.1)	0.000	1.00	0.00 (±4.50)	53 (5.91)	0.99 (0.98–0.99)	45 (41.9)	45 (42.0)	0.42	0.68	0.09 (±3.76)	87 (1.92)	1.00 (1.00–1.00)
Standing (GMFM-D)	35 (39.3)	35 (38.9)	1.93	0.06	0.44 (±3.93)	81 (2.01)	1.00 (1.00–1.00)	36 (39.7)	35 (of 39.2)	2.06	0.04	0.94 (±7.90)	84 (4.03)	1.00 (0.99–1.00)
Walking, running, jumping (GMFM-E)	29 (36.2)	29 (36.5)	-0.19	0.85	-0.02 (±2.42)	84 (1.24)	1.00 (1.00–1.00)	30 (37.0)	30 (37.2)	0.09	0.92	0.01 (±2.28)	84 (1.16)	1.00 (1.00–1.00)
Total	50 (32.48)	50 (32.4)	2.21	0.03	0.23 (±1.82)	71 (0.93)	1.00 (1.00–1.00)	51 (32.8)	51 (32.8)	2.29	0.02	0.32 (±2.43)	65 (1.24)	1.00 (1.00–1.00)

ICC, intraclass correlation coefficient; LB, lower bound; UB, upper bound; CI, confidence interval; GMFM-88, Gross Motor Function Measure-88.

Table 5. Test-retest reliability (paediatric physical therapist who was familiar with the child). Results of adapted version of GMFM-88-CVI. ICC (CI) (n = 77).

GMFM-88-CVI	Paediatric physiotherapist familiar with the child					Paediatric physiotherapist not familiar with the child				
	ICC (LB-UB) GMFCS 1	ICC (LB-UB) GMFCS 2	ICC (LB-UB) GMFCS 3	ICC (LB-UB) GMFCS 4	ICC (LB-UB) GMFCS 5	ICC (LB-UB) GMFCS 1	ICC (LB-UB) GMFCS 2	ICC (LB-UB) GMFCS 3	ICC (LB-UB) GMFCS 4	ICC (LB-UB) GMFCS 5
Lying, rolling (GMFM-A)	0.89 (0.77–0.95)	0.79 (0.04–0.97)	0.98 (0.92–1.00)	0.97 (0.91–0.99)	0.92 (0.81–0.97)	0.89 (0.77–0.95)	1.00 (1.00–1.00)	0.98 (0.90–1.00)	0.90 (0.76–0.96)	0.91 (0.78–0.96)
Sitting (GMFM-B)	0.99 (0.97–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)	0.98 (0.94–0.99)	0.88 (0.71–0.95)	0.99 (0.97–0.99)	1.00 (1.00–1.00)	0.99 (0.97–1.00)	0.98 (0.95–0.99)	0.87 (0.71–0.95)
Crawling, kneeling (GMFM-C)	0.96 (0.91–0.98)	0.96 (0.79–0.99)	0.96 (0.81–0.99)	0.99 (0.94–0.99)	0.80 (0.57–0.91)	0.98 (0.95–0.99)	0.96 (0.73–0.99)	0.95 (0.81–0.99)	0.96 (0.90–0.98)	0.80 (0.57–0.91)
Standing (GMFM-D)	0.83 (0.63–0.92)	0.86 (0.06–0.98)	0.87 (0.53–0.97)	0.96 (0.89–0.98)	1.00 (1.00–1.00)	0.86 (0.71–0.94)	0.89 (0.40–0.98)	0.89 (0.57–0.97)	0.97 (0.93–0.99)	1.00 (1.00–1.00)
Walking, running, jumping (GMFM-E)	0.98 (0.96–0.99)	0.91 (0.48–0.99)	0.96 (0.85–0.99)	0.92 (0.80–0.97)	1.00 (1.00–1.00)	0.99 (0.96–1.00)	0.93 (0.62–0.99)	0.94 (0.78–0.99)	0.89 (0.73–0.95)	1.00 (1.00–1.00)
Total	0.97 (0.93–0.99)	0.96 (0.69–0.99)	0.98 (0.93–1.00)	0.99 (0.98–1.00)	0.96 (0.90–0.98)	0.97 (0.93–0.99)	0.96 (0.73–0.99)	0.97 (0.88–0.99)	0.98 (0.94–0.99)	0.95 (0.88–0.98)

ICC, intraclass correlation coefficient; LB, lower bound; UB, upper bound; CI, confidence interval; GMFM-88, Gross Motor Function Measure-88; GMFCS, Gross Motor Function Classification System.

Table 6. Interobserver reliability. Results of adapted version of GMFM-88-CVI. ICC (CI) (n = 77).

GMFM-88-CVI	Test					Retest				
	ICC (LB-UB) GMFCS 1	ICC (LB-UB) GMFCS 2	ICC (LB-UB) GMFCS 3	ICC (LB-UB) GMFCS 4	ICC (LB-UB) GMFCS 5	ICC (LB-UB) GMFCS 1	ICC (LB-UB) GMFCS 2	ICC (LB-UB) GMFCS 3	ICC (LB-UB) GMFCS 4	ICC (LB-UB) GMFCS 5
Lying, rolling (GMFM-A)	0.99 (0.99–1.00)	1.00 (1.00–1.00)	1.00 (1.00–1.00)	0.99 (0.99–1.00)	0.97 (0.99–1.00)	0.99 (0.99–1.00)	1.00 (1.00–1.00)	0.99 (0.98–1.00)	0.95 (0.87–0.98)	0.99 (0.96–1.00)
Sitting (GMFM-B)	1.00 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	1.00 (1.00–1.00)	1.00 (1.00–1.00)	1.00 (1.00–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)	0.99 (0.99–1.00)
Crawling, kneeling (GMFM-C)	0.99 (0.99–1.00)	0.99 (0.95–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.95–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)	1.00 (1.00–1.00)
Standing (GMFM-D)	0.98 (0.95–0.99)	1.00 (1.00–1.00)	0.98 (0.91–1.00)	0.97 (0.89–0.98)	1.00 (1.00–1.00)	0.97 (0.94–0.99)	0.98 (0.85–1.00)	0.87 (0.56–0.97)	0.86 (0.68–0.95)	1.00 (1.00–1.00)
Walking, running, jumping (GMFM-E)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.97 (0.86–0.99)	0.98 (0.97–0.99)	1.00 (1.00–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)	1.00 (1.00–1.00)	0.96 (0.89–0.98)	1.00 (1.00–1.00)
Total	0.99 (0.99–1.00)	1.00 (1.00–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.99–1.00)	0.99 (0.91–1.00)	0.99 (0.98–1.00)	0.99 (0.99–1.00)	0.99 (0.98–1.00)

ICC, intraclass correlation coefficient; LB, lower bound; UB, upper bound; CI, confidence interval; GMFM-88, Gross Motor Function Measure-88; GMFCS, Gross Motor Function Classification System.

study, the correlation between the GMFCS level and GMFM score was for GMFM-A= -0.52; GMFM-B= -0.77; GMFM-C = -0.85; GMFM-D= -0.94; GMFM-E= -0.91; GMFM-Total= -0.91.

4. Discussion

The aims of our study were to develop an adapted version of the GMFM-88 and to determine test-retest and interobserver reliability of this adapted version for children with CP and CVI. The adapted version of the GMFM-88 is based on solid agreement of experts and shows very good test-retest and interobserver reliability. From the high intraclass correlation coefficients and Cronbach's alphas we conclude that reliability was good, not only at the dimension scores but also for the individual items.

4.1. Adaptation

The current adaptation of the GMFM-88 for children with CVI helps measure a specific task without changing the question or instruction of the original GMFM-88 (see Table 1). In line with other studies (Dutton & Jacobson, 2001; Dutton, 2013; Ghasia et al., 2008; Salavati et al., 2014) on CVI and the viewpoint of experts, the adaptation of the GMFM-88 for children with CVI was at the level of verbal support/instruction, manual support, types of equipment and environment.

The experts suggested that before touching the toy the child needs to be aware of the position of the toy, which is placed at a 45^0 angle from the child. Furthermore, when 'sitting on a small bench and achieving standing position without using its arms' (GMFM-88, no. 59), the child is still allowed to use its arms for orientation to locate the chair or table, and makes use of a chair with armrests. The armrests will be used as an orientation point for the child and not as a means of support. In addition, using verbal or manual support will help the child to describe an action that occurs.

The focus of this study was on CVI because the need of children with ocular visual impairment is different than that of those with CVI (Da Costa et al., 2004; Dutton & Jacobson, 2001; Ghasia et al., 2008; Schenk-Rootlieb, Van Nieuwenhuizen, Van der Graaf, et al., 1993; Schenk-Rootlieb, Van Nieuwenhuizen, Schiemanck, et al., 1993; Schenk-Rootlieb et al., 1994; Stiers et al., 2002). CVI is quite variable, ranging from no light perception to normal visual acuity and in the presence of cognitive visual dysfunction, a visual processing disorder that leads to misinterpretation of the visual world with respect to either what or where objects are (Edmond & Foroozan, 2006). We cannot ensure representation of all subtypes though. Further research is needed to determine the psychometric properties of this adapted GMFM-88-CVI for children with different subtypes of CVI. Furthermore, children with CVI could have difficulty at different levels on occasional fixation on large objects, faces or movements in the environment, and on variable visual function, but some moments of good visual fixation as indicated by the ability to see small objects (Good, Jan, Burden, Skoczenski, & Candy, 2001). A familiar environment can result in successful performance of skills, in contrast to an unknown or less familiar environment. It is therefore important to use the GMFM-88-CVI to evaluate a child's level of functioning in the same environment.

The severity of CVI can be determined by the amount of support, such as the child touching or holding the hand of a parent/caregiver in order to orient itself or during transfers. The experts gave suggestions for use of manual support during the practice phase to familiarise the children with CVI with a specific motor task. On the dimension 'lying and rolling', for an item such as 'bring hands to midline, fingers touching', the experts suggested using specific equipment that is colourful, sound-producing, and high in contrast to receive the attention of the child. They also suggested that the physical therapists needed to sit at the side the child rolls over or pivots towards (GMFM-88, nos. 8, 9, 14-17). For the items 'sit on mat, touch toy' (GMFM-88, nos. 26, 27), the experts suggested using a toy that is colourful, sound-producing and high in contrast. During the practice phase the child is allowed to touch the toy in order to locate it. On the 'crawling, kneeling and standing' dimension use of verbal support is allowed in order to give the child information to move in the direction of the therapist. On the 'walking, running and jumping' dimension the experts suggested using special material as well as verbal and manual support to help the child accomplish the task.

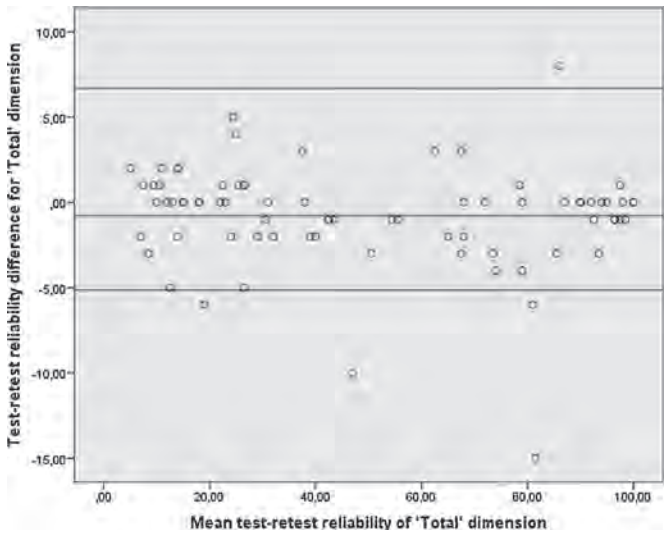


Figure 2. Bland and Altman plot for test–retest reliability on the 'Total' dimension for paediatric physiotherapist familiar with the child. The mean difference is -0.77 (±5.91) (LOA)(-5.14; 6.68).

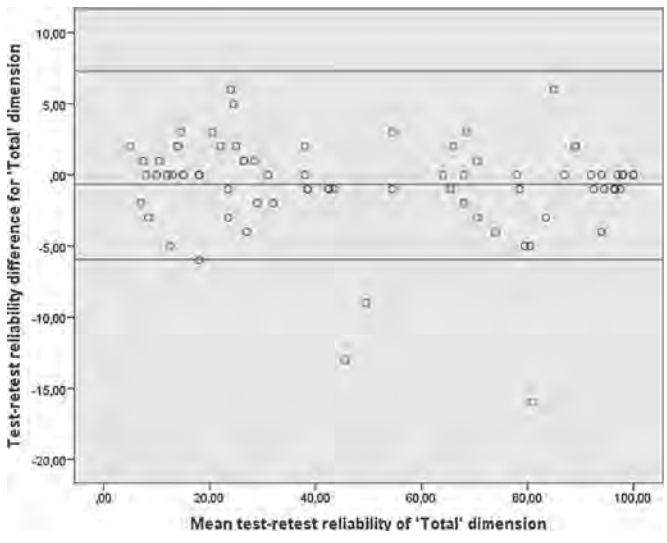


Figure 3. Bland and Altman plot for test-retest reliability on the 'Total' dimension for paediatric physiotherapist not familiar with the child. The mean difference is -0.68 (±6.63) (LOA) (-5.95; 7.31).

4.2. Test-retest reliability and interobserver reliability

The test-retest and interobserver reliability ICCs of dimension scores for the GMFM-88-CVI were excellent and are comparable with the original GMFM-88 study (Ketelaar et al., 2003; Engelen et al., 2007). The internal consistency of dimension scores for the GMFM-88-CVI as well as the original GMFM-88 were equally high (Ketelaar et al., 2003; Engelen et al., 2007).

In our study, the percentages of identical scores for test-retest and interobserver reliability for the 'Total' dimension were 30 (test-retest reliability: paediatric physical therapist familiar with the child) and 29 (paediatric physical therapist not familiar with the child) (Table 3). If, for instance, the agreement is based on 2 points above or below zero, then the percentage of identical scores for test-retest reliability (paediatric physical therapist familiar with the child) on the 'Total'

dimension is 75 instead of 30 (mean: -0.78 and SD: 3.01) (Fig. 2). For test-retest reliability (paediatric physical therapist not familiar with the child) on the 'Total' dimension it is 74 instead of 29 (mean: -0.69 and SD: 3.38) (Fig. 3). The Bland and Altman plots in Figs. 2 and 3 illustrate variations around the zero line. Except for a few values, these plots demonstrate roughly equal distribution 5 points above and below the zero line for the 'Total' dimension for test-retest reliability. If we compare the scores of the 'Total' dimension for test-retest reliability, the scores of the same children are 5 points above or below zero.

5. Limitations

Our study included participants with different types of CP in various degrees of severity, who might have different profiles of motor functioning. Most of the participants (96%) were children with spastic CP. The reliability of the GMFM-88-CVI could be investigated further in a group of children with different types of CP.

CVI includes different types of visual impairments such as visual attention, depth and visual field problems. A higher degree of assistance may explain the presence of CVI as well as its severity. For future studies it is important to determine the relation between the current adaptation of the GMFM-88-CVI and different types of visual impairment, as different types of CVI could result in different visual behaviours in daily life.

6. Conclusion

The adapted version of the GMFM-88 is a useful and reliable instrument for paediatric physical therapists who work with children with both CP and CVI. Considering the fact that visual perception contributes to the performance of motor functioning, it is important to use the version of GMFM-88-CVI for children who have CVI, so that their motor functioning can be measured.

Acknowledgements

We gratefully thank the parents, children and therapists for their participation. Financial support for this study was provided by the Novum Foundation (O10323), a Dutch non-profit organisation that grants financial support to projects and research that improve the quality of life of individuals with visual impairments (www.stichtingnovum.org) and Royal Dutch Visio, centre of expertise for blind and partially sighted people (www.visio.org).

References

1. Avery, L. M., Russell, D. J., Raina, P. S., Walter, S. D., & Rosenbaum, P. L. (2003). Rash analysis of the gross motor function measure: Validating the assumptions of the Rasch model to create an interval-level measure. Archives Physical Medicine & Rehabilitation, 84, 697-705.
2. Chia-Chien, H., & Sandford, B. A. (2007). The Delphi technique: Making sense of consensus. Practical Assessment, Research & Evaluation, 12, 1.
3. Chrysagis, N., Skordilis, E. K., Stavrou, N., Grammatopoulou, E., & Koutsouki, D. (2012). The effect of treadmill training on gross motor function and walking speed in ambulatory adolescents with cerebral palsy: A randomized controlled trial. American Journal of Physical Medicine & Rehabilitation, 91(9), 747-760.
4. Cronbach, L. J. (1951). Coefficient alpha and the internal structure of tests. Psychometrika, 16, 297-334.
5. Da Costa, M. F., Salmao, S. R., Berezovsky, A., De Haro, F. M., & Ventura, D. F. (2004). Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. Behavioural Brain Research, 149(2), 145-150.
6. Dutton, G. N. (2013). The spectrum of cerebral visual impairment as a sequel to premature birth: An overview. Documenta Ophthalmologica, 127(1), 69-78.
7. Dutton, G. N., & Jacobson, L. K. (2001). Cerebral visual impairment in children. Seminars in Neonatology, 6, 477-485.
8. Dutton, G. N., Saaed, A., Fahad, B., Fraser, R., McDaid, G., McDade, J., et al. (2004). Association of binocular lower visual field impairment, impaired
9. simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction—a retrospective observational study. Eye, 18, 27-34.
10. Edmond, J., & Foroozan, R. (2006). Cortical visual impairment in children. Current Opinion in Ophthalmology, 17, 509-512.
11. Engelen, V., Ketelaar, M., & Gorter, J. W. (2007). Electing the appropriate outcome in paediatric physical therapy: How individual treatment goals for children with cerebral palsy are reflected in GMFM-88 and PEDI. Journal of Rehabilitation Medicine, 39, 225-231. Fazzi, E., Signorini, S. G., Piana, L. A., Bertone, R., Misefari, C., Galli, W. P. E., et al. (2012). Neuro-ophthalmological disorders in cerebral palsy: Ophthalmological, oculomotor, and visual aspects. Developmental Medicine & Child Neurology, 54, 730-736.
12. Ghasia, F., Burnstroom, J., Gordon, M., & Tychsen, L. (2008). Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. Investigative Ophthalmology & Visual Science, 49, 572-580.
13. Good, W. V., Jan, J. E., Burden, S. K., Skoczenski, A., & Candy, R. (2001). Recent advances in cortical visual impairment. Developmental Medicine & Child Neurology, 43(1), 56-60.
14. Gracht von der, H. A. (2012). Consensus measurement in Delphi studies: Review and implications for future quality assurance. Technological Forecasting & Social Change: An International Journal, 79(8), 1525-1536.

15. Guzzetta, A., Mercuri, E., & Cioni, G. (2001). Visual disorders in children with brain lesions: Visual impairment associated with cerebral palsy. European Journal of Paediatric Neurology, 5, 115-119.
16. Haley, S. M., Coster, W. J., Ludlow, L. H., Haltiwanger, J. T., & Andrellos, P. J. (1992). Pediatric evaluation of disability inventory: Development, standardization, and administration manual. Boston, MA: New England Medical Centre Inc and PEDI Research Group.
17. Houwen, S., Hartman, E., & Visscher, C. (2009). Physical activity and motor skills in children with and without visual impairments. Medicine & Science in Sports & Exercise, 41(1), 103.
18. International Classification of Functioning Disability and Health, Child & Youth version (Dutch translation) (2008). (1st ed.). The Netherlands: Dutch WHO-FIC Collaborating Centre, Bohn Stafleu van Loghum.
19. Ketelaar, M., van Petegem-van Beek, E., & Visser, J. J. W. (1995). Gross motor function measure manual: Nederlandse Vertaling. Utrecht, The Netherlands: Utrecht University.
20. Ketelaar, M., Van Petegem-van Beek, E., Veenhof, C., Visser, J., & Vermeer, A. (2003). Gross motor function measure. University of Utrecht. Child Physical Therapy, 39, 5-7.
21. Ko, J., & Kim, M. (2013). Reliability and responsiveness of the Gross Motor Function Measure-88 in children with cerebral palsy. Journal of the America Physical Therapy Association, 93(3), 393-400.
22. McGraw, Kenneth, O., & Wong, S. P. (1996). Forming inferences about some intraclass correlation coefficients. Psychological Methods, 1, 30-46.
23. Palisano, R., Rosenbaum, P., Walter, S., Russell, D., Wood, E., & Galuppi, B. (1997). Development and reliability of a system to classify gross motor function in children with cerebral palsy. Developmental Medicine & Child Neurology, 39, 214-223.
24. Powell, C. (2003). The Delphi technique: Myths and realities. Journal of Advanced Nursing, 41(4), 376-382.
25. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M., & Bax, M. (2007). Definition and classification of cerebral palsy. Developmental Medicine & Child Neurology, 49(6), 480.
26. Salavati, M., Rameckers, E. A. A., Steenbergen, B., & Schans van der, C. (2014). Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). European Journal of Physical Therapy, 16(3), 159-167. Salavati, M., Waninge, A., Rameckers, E. A. A., Blécourt, A. C. E., Krijnen, W. P., Steenbergen, B., et al. (2015). Reliability of the modified Paediatric Evaluation of Disability Inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment. Research in Developmental Disabilities, 37, 189-201.
27. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Van der Graaf, Y., Wittebol-Post, D., & Willemse, J. (1993). The prevalence of cerebral visual disturbance in children with cerebral palsy. Developmental Medicine & Child Neurology, 34, 473-480.
28. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Schiemanck, N., Van der Graaf, Y., & Willemse, J. (1993). Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. Child Care, Health and Development, 19, 411-423.
29. Schenk-Rootlieb, A. J. F., Van Nieuwenhuizen, O., Van Waes, P. F. G. M., & Van der Graaf, Y. (1994). Cerebral visual impairment in cerebral palsy: Relation to structural abnormalities of the cerebrum. Developmental Medicine & Child Neurology, 25, 68-72.
30. Scholtes, V. A., Becher, J. G., Comuth, A., Dekkers, H., Van Dijk, L., & Dallmeijer, A. J. (2010). Effectiveness of functional progressive resistance exercise strength training on muscle strength and mobility in children with cerebral palsy: A randomized controlled trial. Developmental Medicine & Child Neurology, 52(6), 107-113.
31. Shi, W., Wang, S. J., Liao, Y. G., Yang, H., Xu, X. J., & Shao, X. M. (2006). Reliability and validity of the GMFM-66 in 0- to 3-year-old children with cerebral palsy. American Journal of Physical Medicine & Rehabilitation, 85, 141-147.
32. Stiers, P., Vanderkelen, R., Vanneste, G., Coene, S., De Rammelsere, M., & Vandenbussche, E. (2002). Visual-perceptual impairment in a random sample of children with cerebral palsy. Developmental Medicine & Child Neurology, 44, 370-382.
33. Verrel, J., Bekkering, H., & Steenbergen, B. (2008). Eye-hand coordination during manual object transport with the affected and less affected hand in adolescents with hemiparetic cerebral palsy. Experimental Brain Research, 187, 107-116.
34. Visser, L., Ruiter, A. J., Meulen van der, F., Ruijssenaars, A. J. J. M., & Timmerman, E. (2014). Validity and suitability of the Bayley-III Low Motor/Vision
35. version: A comparative study among young children with and without motor and/or visual impairments. Pediatric Physical Therapy, 26(1), 57-67.
36. Wassenberg-Severijnen, J. E., Custers, J. W., Hox, J. J., Vermeer, A., & Helders, P. J. (2003). Reliability of the Dutch Pediatric Evaluation of Disability Inventory (PEDI). Clinical Rehabilitation, 17, 457-462.
37. Wood, E., & Rosenbaum, P. (2000). The gross motor function classification system for cerebral palsy: A study of reliability and stability over time. Developmental Medicine & Child Neurology, 42, 292-296.
38. World Health Organization International Classification of Functioning (2001). Disability and health: ICF. Geneva: WHO.

5 |

Gross motor function in children with spastic Cerebral Palsy and Cerebral Visual Impairment: A comparison between outcomes of the original and the cerebral visual impairment adapted Gross Motor Function Measure-88 (GMFM-88-CVI)

M. Salavati, MSPT[a, b], E.A.A. Rameckers, PhD PPT[c,d,e], A. Waninge, PhD[a, b], W.P. Krijnen, PhD[b], B. Steenbergen, PhD[f, g], C.P. van der Schans, PhD, CE, PT[b,h]

[a] Royal Dutch Visio, centre of expertise for blind and partially sighted people; The Netherlands.

[b] Hanze University of Applied Sciences Groningen, research group Healthy Ageing, allied health care and nursing; Groningen, The Netherlands.

[c] Maastricht University, Department of Rehabilitation Medicine, School for Public Health and Primary Care (CAPHRI); Maastricht, The Netherlands.

[d] Adelante Center of Expertise in Rehabilitation and Audiology; Valkenburg and Hoensbroek, The Netherlands.

[e] AVANSplus, University for Professionals for Paediatric Physical Therapy; Breda, The Netherlands

[f] Radboud University Nijmegen, Behavioural Science Institute; Nijmegen, The Netherlands.

[g] Australian Catholic University, School of Psychology; Melbourne, Australia.

[h] University of Groningen, University Medical Center Groningen, Department of Rehabilitation Medicine; Groningen, The Netherlands.

Submitted as:

Salavati, M., Rameckers, E.A.A., Waninge, A., Krijnen, W.P., Steenbergen, B. & Schans van der, C.P. Gross motor function in children with spastic cerebral palsy and cerebral visual impairment: A comparison between outcomes of the original and cerebral visual impairment adapted Gross Motor Function Measure-88 (GMFM-88-CVI).

Abstract

Purpose: To investigate whether the adapted version of the Gross Motor Function Measure-88 (GMFM-88) for children with Cerebral Palsy (CP) and Cerebral Visual Impairment (CVI) results in higher scores and is a better reflection of their gross motor function per se without the influence of impaired visual abilities.

Method: The scores of the original GMFM-88 and the GMFM-88-CVI were compared in the same group of children (n= 21 boys and n= 16 girls), mean (SD) age 113 (30) months with CP and CVI, within a time span of two weeks. To compare outcomes of the original GMFM-88 and the GMFM-88-CVI, a paediatric physical therapist familiar with the child assessed both tests in random order. GMFCS level, mental development and age at testing were also collected. The Related (paired) Samples Wilcoxon Signed Rank Test with a significance level of $p<.05$ was used to detect possible significant differences in mean scores of both tests.

Results: The comparison between scores on the GMFM-88-CVI and the original version of GMFM-88 in children with CP and CVI yielded higher or similar scores on all dimensions of gross motor function, including lying, rolling, sitting, crawling, kneeling, standing, walking and running, as well as the total score ($p<.001$).

Conclusion: The GMFM-88-CVI provides a better estimate of gross motor function per se in children with CP and CVI that is not adversely affected by their visual problems. On the basis of these findings, we recommend using the GMFM-88-CVI to measure gross motor functioning in children with CP and CVI.

1. Introduction

The Gross Motor Function Measure-88 (GMFM-88) is a widely used instrument to assess motor capacity in children with Cerebral Palsy (CP) (Chrysagis, Skordilis, Stavrou, Grammatopoulou & Koutsouki, 2012; Scholtes, Becher, Comuth, Dekkers, Van Dijk & Dallmeijer, 2010). However, it does not account for the presence of visual impairments, which may reduce validity for children with CP and visual impairments. We previously adapted the GMFM-88 for children with CP and Cerebral Visual Impairments (CVI). This adapted version (GMFM-88-CVI) takes into account the presence of higher visual impairments in children with CP and is reliable for measuring motor functioning in children with CP and CVI (Salavati, Krijnen, Rameckers, Looijestijn, Maathuis, Schans van der & Steenbergen, 2015b). The GMFM-88-CVI supports a specific task without changing the question or instruction of the original GMFM-88. The adaptation of the GMFM-88-CVI for children with CVI is at the level of verbal support/instruction, manual support, types of equipment and environment (Salavati et al. 2015b).

CP represents a large group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain (Rosenbaum, Paneth, Leviton, Goldstein, & Bax, 2007). Gross motor function of children with CP can be classified into five different severity levels using the Gross Motor Function Classification System (GMFCS), where level 1 indicates the least and level 5 the most functional limitation. Generally, children at GMFCS level 1 walk indoors and outdoors and climb stairs without limitations, children at GMFCS level 2 walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines, children at GMFCS level 3 walk indoors or outdoors on a level surface with an assistive mobility device, children at GMFCS level 4 sit on a chair but need adaptive seating for trunk control, and children at GMFCS level 5 have physical impairments that restrict voluntary control of movement (Rosenbaum et al., 2007). Motor disorders of CP can be accompanied by disturbances of sensation, perception, cognition, communication and behaviour, as well as by epilepsy and secondary musculoskeletal problems (Rosenbaum et al., 2007).

Visual impairment can have a major impact on motor development and skills acquisition. A delayed onset of different motor milestones, such as sitting, crawling, standing or walking, has been reported in visually impaired children (Prechtl, Cioni, Einspieler, Bos & Ferrari, 2001; Elisa, Josee, Oreste, Claudia, Antonella, Sabrina, et al., 2002; Levtzion-Korach, Tennenbaum, Schnitzer, & Ornoy, 2000). CVI is observed in approximately 30% of children diagnosed with various forms of CP (Ghasia, Burnstroom, Gordon, & Tychsen, 2008; Stiers, Vanderkelen, Vanneste, Coene, De Rammelsere & Vandenbussche, 2002). CVI can be defined as deficient visual functioning, resulting from a sequel of damage or malformation of the retrogeniculate visual pathways (optic radiations, occipital cortex and visual association areas) in the absence of damage of the anterior visual pathways or any major ocular disease (Dutton & Jacobson, 2001; Dutton, Saaed, Fahad, Fraser, McDaid & McDade, 2004). CVI ranges in severity from blindness to relatively minor impairments of visual perception. Children with CVI exhibit slow, inefficient and highly variable visual performance during daily-life activities (Good, Jan, Burden, Skoczenski, Candy, 2001). CVI has an impact on all aspects of a child's development, and children with CP and CVI develop more slowly in the areas of self-care, mobility and social function than children with CP without CVI. (Da Costa, Salmao, Berezovsky, De Haro, Ventura, 2004; Dutton & Jacobson, 2001; Dutton, 2013; Ghasia et al., 2008; Good et al., 2001; Salavati, Rameckers, Steenbergen & Schans van der, 2014; Schenk-Rootlieb, Van Nieuwenhuizen, Van der Graaf, Wittebol-Post, & Willemse, 1993; Salavati, Waninge, Rameckers, de Blécourt, Krijnen, Steenbergen & Schans van der, 2015a). Children affected more severely by CP have greater reduction in visual acuity (Da Costa et al., 2004; Fazzi et al., 2012).

The GMFM-88 consists of 88 items in five dimensions. The reliability and validity of this test are sufficient (inter-rater reliability: ICC = 0.75-1.00; test-retest reliability: ICC = 0.96-0.99) (Engelen et al., 2007; Ketelaar, Van Petegem-van Beek, Veenhof, Visser & Vermeer, 2003). The GMFM-88 is a criterion-referenced instrument constructed to evaluate the development of motor skills in children with CP, and designed and validated for these children by using principles of

classical test theory. It is widely used as a clinical and research outcome measure and there is considerable evidence of its reliability, validity and responsiveness (Avery, Russell, Raina, Walter & Rosenbaum, 2003). Importantly though, its reliability and validity for children with visual impairments is unknown (Ketelaar et al., 2003; Engelen et al., 2007). Experts working with children who have both CVI and CP experienced that the GMFM-88 does not account for the presence of visual impairments – that is, the assessment may not be suitable for children with CVI because outcome scores are likely to be negatively affected by visual impairments. As such, it is a potentially less valid measure to assess motor functioning in children with CP and CVI. These children have an inherent limitation with proper identification and processing of visual information (Haley, Coster, Ludlow, Haltiwanger & Andrellos, 1992; Salavati et al., 2014). Also, because of visual impairments a child might not be able to show its motor functioning abilities during a standardised assessment of motor development, leading to a possible underestimation of its true motor capacity (Visser, Ruiter, Meulen van der, Ruijssenaars & Timmerman, 2014; Salavati et al., 2014).

The GMFM-88-CVI for children with CP and CVI (test-retest reliability: ICC = 0.94-1.00; interobserver reliability: ICC = 1.00-1.00; internal consistency= 0.97-1.00) takes into account higher visual impairments in children with CP (Salavati et al., 2015b). Based on the importance of visual processing on motor performance we hypothesise that the original GMFM-88 gives an underestimation of the gross motor functioning of children with CP and CVI.

The aim of our study was to investigate whether the GMFM-88-CVI for children with CP and CVI results in a higher score of their gross motor function via a comparison with the original GMFM-88 in the same group of children with CP and CVI.

2. Methods

2.1 Participants

Children with CP and CVI were recruited from Royal Dutch Visio (centres of expertise for blind and partially sighted people) and allied healthcare practices. Inclusion criteria were presence of any type of CP and CVI, mild or moderate intellectual disability (IQ 70-40), and age at testing between 4 and 12 years. Level of intellectual disability was obtained from the medical files. Children with another comorbid syndrome (e.g. Down syndrome) or hearing difficulties (> 30 dB hearing loss), severe or profound intellectual disability (IQ<40) and (corrected) vision < 0.3 and/or field of vision < 30° were excluded. Children who had planned surgery between the two tests were also excluded.

The diagnosis of CP and the classification according to GMFCS level were obtained from the medical files and judged by a rehabilitation physician. The diagnosis of CVI was determined based on the results of ophthalmological and psychological/ neuropsychological assessment and on the assessment data reported by a developmental coach specialised in working with children with visual impairments. According to them, the diagnosis of CVI was determined by the following criteria: a normal or near-normal eye exam performed by an ophthalmologist, a history or presence of neurological problems, and presence of behavioural responses to visual stimuli which are unique to CVI. These responses constitute strong colour preference, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non-purposeful gaze, difficulty with distance viewing, absent or atypical visual reflexes, difficulty with visual novelty, and absence of visually guided reach (Dutton & Jacobson, 2001; Stiers, 2002). Children with all types of CVI were included in our study and no selection was carried out based on subtypes.

Permission to conduct the study was obtained from the Medical Ethical Committee (METC-2014.438) of University Medical Center Groningen (UMCG), the Netherlands. Written informed consent was obtained from the children's parents.

2.2 Test instrument

2.2.1 The original GMFM-88

The GMFM-88 is a standardised functional assessment tool used by therapists to examine the achievements and limitations of gross motor function of children with CP, monitor progress of the individual child, and evaluate the outcomes of treatment programs of this population. The GMFM-88 is responsive to changes in motor functioning, and can be used to measure changes in fundamental gross motor skills

over time in children with CP as well as evaluate their physiotherapeutic interventions (Ketelaar et al., 2003; Engelen et al., 2007). The test consists of 88 items grouped into five dimensions of gross motor functions: lying and rolling (GMFM-A) 17 items; sitting (GMFM-B) 20 items; crawling and kneeling (GMFM-C) 14 items; standing (GMFM-D) 13 items; and walking, running and jumping (GMFM-E) 24 items. Each item is scored on a 4-point scale. A percentage score is calculated for each dimension and for the total score of the five dimensions. It is possible to score with or without support (walker, crutches and canes) or orthoses (ankle foot control, knee control or shoes). There are no age limits, and a 5-year-old child with normal motor abilities can accomplish all items (Harries, Kassirer, Amichai, Lahat, 2004; Russell, D.J., Rosenbaum, P.L., Avery, L.M., Lane, M., 2002; Avery et al., 2003).

2.2.2 The GMFM-88-CVI

The GMFM-88-CVI is an appendix to the instruction of the original GMFM-88. Most of the adjustments relate to higher motor skills such as jumping, climbing stairs and cycling. Equipment use in GMFM-88-CVI is colourful, sound-producing and high in contrast in order to get the attention of the child who is to move towards the material (Salavati et al., 2015b).

The test-retest reliability ICCs of dimension scores are 0.94-1.00 and the inter-observer reliability ICCs for the GMFM-88-CVI are 1.00-1.00 for dimension scores. Test-retest and interobserver reliability of the GMFM-88-CVI for children with CP and CVI are excellent. Internal consistency of dimension scores is: dimension A 0.97-1.00, dimension B 0.99-1.00, dimension C 1.00-1.00, dimension D 1.00-1.00, dimension E 1.00-1.00 and Total 1.00-1.00; the dimensions are thus reliable (Salavati et al., 2015b).

Table 1. Characteristics of the participants.

Characteristic	**Children with CP and CVI**
Age in months, mean (SD), min-max	113 (30), 54-144
Gender, *n (%)* male / *n (%)* female	21 (57) /16 (43)
Type of cerebral palsy: *n (%)* spastic / *n (%)* dyskinetic	36 (97) / 1 (3)
GMFCS I, *n* (%)	bilateral 10 (27), unilateral left 1 (3)
GMFCS II, *n* (%)	6 (16) bilateral
GMFCS III, *n* (%)	3 (8) bilateral
GMFCS IV, *n* (%)	7 (19) bilateral
GMFCS V, *n* (%)	10 (27) bilateral
Speech/language development:	
ICF-CY, d3100 = reacts to human voice, *n* (%)	37 (100)
ICF-CY, d3101 = understands simple spoken messages, *n* (%)	37 (100)
ICF-CY, d3102 = understands complex spoken messages, *n* (%)	27 (73)
ICF-CY, d330 = speaks, *n* (%)	21 (57)
ICF-CY, d331 = babbles, *n* (%)	16 (43)
ICF-CY, d3350 = uses body language, *n* (%)	32 (87)
ICF-CY, d3351 = uses signs and symbols, *n* (%)	28 (76)
Level of intellectual disability, *n* (%) mild / *n* (%) moderate	14 (38) / 23 (62)
Presence of epilepsy, *n* (%)	3 (8)
Use of epilepsy medication, (*n* (%)	3 (8)
Percutaneous endoscopic gastrostomy tube feeding, *n* (%)	4 (11)

GMFCS, Gross Motor Function Classification System; ICF-CY, International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation).

Table 2. Difference in percentage score between GMFM-88 and GMFM-88-CVI for each child.

Child	GMFCS level	Level of intellectual disability*	GMFM-A (lying & rolling)	GMFM-B (sitting)	GMFM-C (crawling & kneeling)	GMFM-D (standing)	GMFM-E (walking, running & jumping)	GMFM-Total
1	1	2	12	2	25	10	5	11
2	1	1	10	12	10	7	7	10
3	1	2	9	15	16	10	14	13
4	1	1	9	11	27	14	4	13
5	1	1	3	12	10	5	6	6
6	1	2	12	19	0	3	3	7
7	1	1	20	3	7	0	0	6
8	1**	1	31	23	3	0	0	11
9	1	1	8	2	5	0	0	3
10	1	2	6	8	0	0	0	3
11	1	1	0	0	0	3	10	3
12	2	2	14	20	10	2	16	12
13	2	2	8	10	29	5	3	11
14	2	2	17	24	10	0	0	10
15	2	1	0	5	5	0	0	2
16	2	2	0	0	8	3	4	3
17	2	2	6	2	0	0	0	2
18	3	1	23	9	16	2	3	11
19	3	1	5	3	7	7	2	5
20	3	2	0	0	12	8	5	5
21	4	2	14	12	24	10	10	14
22	4	2	12	17	7	12	7	12
23	4	2	9	7	3	5	5	6
24	4	2	20	15	4	0	0	8
25	4	1	0	2	1	0	0	1
26	4	2	0	0	6	6	7	4
27	4	1	5	0	0	0	0	1
28	5	2	8	7	20	1	37	15
29	5	2	3	4	7	3	5	5
30	5	1	18	19	15	11	-1	12
31	5	2	8	7	-1	15	7	6
32	5	2	26	12	5	0	0	8
33	5	2	1	1	4	0	0	1
34	5	2	20	4	0	0	0	5
35	5	2	33	11	0	0	0	9
36	5	2	30	18	-1	0	0	10
37	5	1	0	0	0	0	3	1

*A positive value indicates a higher score for GMFM-88-CVI. GMFM-88, Gross Motor Function Measure-88; GMFM-88-CVI, Gross Motor Function Measure-88 for children with CVI; CP, cerebral palsy; CVI, cerebral visual impairment; GMFCS, Gross Motor Function Classification System; *1, mild intellectual disability; 2, moderate intellectual disability; **unilateral left.*

2.3 Design

The paediatric physical therapist familiar with the child administered the original GMFM-88 and the GMFM-88-CVI in random order as either first or second test, within a two-week period. We choose to administer the test-retest within two weeks because it is highly unlikely that gross motor function of children with CP will change within such a time period.

2.4 Data collection

Based on the possible effect on motor functioning, we also collected background data on prevalence of epilepsy as well as speech/language development according to the International Classification of Functioning, Disability and Health for Children and Youth (Dutch Translation, 2008). At the level of speech/language development, the collected variables were: d3100 = reacts to human voice; d3101 = understands simple spoken messages; d3102 = understands complex spoken messages; d330 = speaks; d331 = babbles; d3350 = uses body language and d3351 = uses signs symbols (Table 1). The data of children were registered according to GMFCS level and type of CP (unilateral or bilateral) and level of intellectual disability. In addition, the gender and age at which the GMFM-88 and GMFM-88-CVI were administered was noted. All paediatric physical therapists were familiar with both the original GMFM-88 and the GMFM-88-CVI. The tests were administered by trained paediatric physical therapists and the dimension as well as total scores of the GMFM-88 and GMFM-88-CVI for children with CP and CVI were used for further analysis.

2.5 Statistical analyses

Data were analysed using the Statistical Package for Social Sciences (SPSS), v.22 software. Since the distribution of the differences deviated extensively from normal, the Related Samples Wilcoxon Signed Rank Test with a significance level of $p<.05$ was used to detect possible significant differences in mean scores of both tests.

3. Results

All children were tested with both tests between November 2014 and February 2015. Mean (SD) duration between the two tests was 10 (6) days. We took data from 37 children with both CP and CVI (n= 21 boys and n= 16 girls) for analysis. Table 1 provides the characteristics of the included children.

Table 2 shows the individual differences in percentage scores between the GMFM-88 and the GMFM-88-CVI for children with CP and CVI. Fourteen children at GMFCS 1-5 with mild or moderate intellectual disability showed a positive difference in percentage scores on all dimensions: lying and rolling (GMFM-A); sitting (GMFM-B); crawling and kneeling (GMFM-C); standing (GMFM-D); and walking, running and jumping (GMFM-E). Six children at GMFCS 1 with mild and moderate intellectual disability had a positive difference in percentage scores on almost all dimensions. Seven children at GMFCS 1, 2, 4, 5 with mild or moderate intellectual disability had a positive difference in percentage scores for the dimensions GMFM-A, GMFM-B and GMFM-C. Two children showed a positive difference in percentage scores for only dimensions GMFM-B and GMFM-C. No differences in percentage scores were found for the dimensions GMFM-D and GMFM-E. Three children at GMFCS 2, 3, 4 with moderate intellectual disability showed a positive difference in percentage scores for only the dimensions: GMFM-C, GMFM-D and GMFM-E. Four children at GMFCS 1, 2, 5 with moderate intellectual disability showed a positive difference in percentage scores on dimensions GMFM-A and GMFM-B, but no difference on dimensions GMFM-C, GMFM-D and GMFM-E.

One child at GMFCS 1 with mild intellectual disability showed a positive difference in percentage scores for the dimensions GMFM-D and GMFM-E. One child at GMFCS 5 with mild intellectual disability showed a positive difference in percentage scores for the dimension GMFM-E only. One child at GMFCS 4 with mild intellectual disability showed a positive difference in percentage scores for the dimension GMFM-A only.

Table 3 shows the comparison between the GMFM-88 and the GMFM-88-CVI for the separate GMFM dimensions. The Related (paired) Samples Wilcoxon Signed Rank Test shows that the differences between both test outcomes on all dimensions as well as the Total scores are significant ($p<.001$). The children tested with GMFM-88-CVI scored significantly higher

Table 3. Related (paired) Samples Wilcoxon Signed Rank Test. Median (min-max) and Mean (SD) of GMFM-88 and GMFM-88-CVI scores, Z–value, median, and 95% CI of differences. N=37

GMFM dimension	GMFM-88		GMFM-88-CVI		*Z* –value[a]	Median of the differences (95% CI lower-upper)
	Median (min-max)	Mean (SD)	Median (min-max)	Mean (SD)		
A	79 (10-100)	74 (25)	96 (28-100)	85 (21)	-4.79	12.5 (9.0-16.5)
B	77 (2-100)	64 (33)	92 (10-100)	73 (32)	-4.86	10.0 (7.0-13.0)
C	43 (0-100)	44 (39)	56 (0-100)	52 (42)	-4.62	9.0 (6.0-13.0)
D	14 (0-100)	35 (38)	26 (0-100)	40 (39)	-4.02	6.5 (4.5-8.5)
E	7 (0-97)	29 (34)	11 (0-100)	33 (38)	-4.08	6.0 (4.0-8.5)
Total	39 (2-99)	50 (31)	49 (8-100)	57 (32)	-5.31	7.0 (5.5-8.5)

GMFM-88, Gross Motor Function Measure-88; GMFM-88-CVI, Gross Motor Function Measure-88 for children with CVI; CP, cerebral palsy; CVI, cerebral visual impairment. A: lying and rolling; B: sitting; C: crawling and kneeling; D: standing; E: walking, running and jumping; Total (A+B+C+D+E); CI, confidence interval.

[a] *All corresponding P-values are <.001.*

on all dimensions (*p<.001)* compared to the children tested with original GMFM-88 (Table 3).

4. Discussion

The aim of our study was to investigate whether the GMFM-88-CVI for children with CP and CVI results in a higher score for gross motor function via a comparison with the original GMFM-88 in the same group of children with CP and CVI. Our study showed that the GMFM-88-CVI results in higher scores for gross motor functioning than the original GMFM-88, hence it is a better test to assess motor capabilities per se for children with CP and CVI. The reason for the higher scores using the GMFM-88 CVI is the adaptation of the instruction of the GMFM-88-CVI. This enables a child with CP and CVI to perform a motor skill and helps the paediatric physical therapist take a more realistic measure of the gross motor function per se that is not confounded by visual impairments. For instance, to enable a child with CP and CVI to 'roll to supine over a side' (GMFM-88, nos. 14 and 15), the paediatric physical therapist used the additional instruction: 'sit on the side the child should roll towards and during the practice phase the paediatric physical therapist uses manual and verbal support (e.g. researcher's voice) to invite the child to roll towards a side'. Also, the paediatric physical therapist 'used toys that have lights, moving parts, produced sound, and/or were fluorescent/high-contrast' (Salavati et al., 2015b). As another example, CVI results in a limitation of depth perception and this causes difficulty performing a task such as 'kicking a ball with the foot' (GMFM-88, nos. 78 and 79) or 'standing on a 15-cm step, jumping off with both feet simultaneously' (GMFM-88, no. 88). The additional instructions, such as verbal and manual support, thus enable the child with CP and CVI to successfully perform the motor skills (Salavati et al., 2015b).

Generally, the additional instruction in the GMFM-88-CVI is based on the amount of manual support (e.g. duration and phase of needed manual support given), verbal support and special equipment (e.g. colourful, sound-producing, high-contrast) needed to obtain the attention of a child with CVI, in order to help the child accomplish a specific skill. The lower score using the original GMFM-88 is thus probably a reflection of visual impairment rather than motor impairment. By using the GMFM-88-CVI, the developmental level of motor performance can be monitored more accurately, which should lead to more realistic planning of appropriate level of motor skills in intervention programs. Interventions can be better adjusted to the needs and capabilities of the child, leading to increased efficacy of such programs. As a consequence, the use of verbal or manual support by the paediatric physical therapist during the intervention will help the child to describe and accomplish an action that occurs. For example, on the 'walking, running and jumping' dimension, using special material as well as verbal and manual support helps the child accomplish the task. Additionally, a familiar environment can result in successful performance of skills, in contrast to an

unknown or less familiar environment. It is therefore important to evaluate a child's level of functioning in the same environment (Salavati et al., 2015b).

The results of our previous study on comparing a group of children with CP with and without CVI (Salavati et al., 2014) showed that children with CP and CVI scored significantly lower ($p<.009$) on all dimensions of the original GMFM-88 than children experiencing CP without CVI. The results of our present study comparing both tests show that by using the GMFM-88-CVI children with CP and CVI score significantly higher ($p<.001$) on all dimensions of the GMFM-88-CVI.

We found that in all GMFM dimensions the scores of the GMFM-88-CVI were higher compared to the GMFM-88. However, those differences were smaller for dimensions D and E. A possible explanation is that only children at GMFCS levels 1 and 2 are able to perform motor tasks on dimensions D and E. Children at GMFCS levels 1 and 2 usually have less severe CVI.

The results from the difference in percentage score between the GMFM-88 and GMFM-88-CVI for each child show that 10 children at GMFCS levels 1 and 2 present no or small differences on GMFM-D and GMFM-E. The reason could be that these children have fewer adverse effects from CVI when they perform motor skills such as standing or walking. These motor skills place a high demand on sustained visual attention (Dutton & Jacobson, 2001; Stiers, 2002).

Da Costa et al. (2004) and Ghasia et al. (2008) showed that visual acuity was lowest for children at GMFCS level 5 and improves progressively for children at GMFCS levels 4, 3, 2 and 1. The results of comparing mean score on all dimensions of both tests indicate that the mean scores on dimensions C, D and E are lower than those on dimensions A and B. A reason could be the fact that in our study a smaller number of children (approximately 20) is able to perform the gross motor tasks on dimensions C, D and E for both tests.

5. Limitations

CVI is quite variable in its range from no light perception to normal visual acuity, and with cognitive visual dysfunction, a disorder of visual processing that leads to misinterpretation of the visual world with respect to either what objects are or where they are (Jane C. Edmond & Rod Foroozan, 2006). In our study we included all types of CVI. In general, each type of CVI could result in different motor performance and outcome for the GMFM-88-CVI. It is important that future studies notice which type of CVI each included child has, therefore the paediatric physical therapist should take into account which type of CVI is present.

Furthermore, children with CP and CVI also have a lack of visual information, so they use the auditory information to better understand their environment. A highly variable visual performance during daily-life activities could result in different performances on two different testing days. To achieve reliable test results, it is important to repeat measuring motor functioning on different days. Also, a familiar environment will result in successful execution of a particular motor skill.

It is important to use the GMFM-88-CVI for children with CP when a child shows a higher level of motor functioning during the therapy but may not be able to show its motor functioning abilities during a standardised assessment of motor development.

6. Conclusion

Assessment with GMFM-88-CVI results in higher scores in children with CP with CVI that are affected by visual problems. On the basis of these findings, we recommend using the GMFM-88-CVI to measure gross motor functioning in children with CP with CVI.

7. Acknowledgements

The authors kindly acknowledge and thank the parents, children and therapists for their participation. Financial support for this study was given by the Novum Foundation (O10323), a Dutch non-profit organisation providing financial support to (research) projects that improve the quality of life of individuals with visual impairments (www.stichtingnovum.org), and Royal Dutch Visio, centre of expertise for blind and partially sighted people (www.visio.org).

8. References

1. Avery, L. M., Russell, D. J., Raina, P. S., Walter, S. D., & Rosenbaum, P. L. (2003). Rash analysis of the gross motor function measure: Validating the assumptions of the Rasch model to create an interval-level measure. *Archives Physical Medicine & Rehabilitation,* 84, 697-705.
2. Chrysagis, N., Skordilis, E. K., Stavrou, N., Grammatopoulou E., Koutsouki, D. (2012)"The effect of treadmill training on gross motor function and walking speed in ambulatory adolescents with cerebral palsy: a

randomized controlled trial," *American Journal of Physical Medicine & Rehabilitation*, 91(9), 747-760.
3. Da Costa, M.F., Salmao, S.R., Berezovsky, A., De Haro, F.M., Ventura, D.F. (2004). Relationship between vision and motor impairment in children with spastic cerebral palsy: new evidence from electrophysiology. *Behavioural Brain Res,* 149(2), 145-150.
4. Dutton, G.N., Jacobson, L.K. (2001). Cerebral visual impairment in children. *Semin Neonatol*, 6, 477–485.
5. Dutton, G.N., Saaed, A., Fahad, B., Fraser, R., McDaid, G., McDade, J., et al. (2004). Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction-a retrospective observational study. *Eye*, 18, 27–34.
6. Dutton, G.N. (2013). The spectrum of cerebral visual impairment as a sequel to premature birth: an overview. *Doc Ophthalmol*, 127(1), 69-78.
7. Elisa, F., Josee, L., Oreste, F., Claudia, A., Antonella, L., Sabrina, S., et al. (2002). Gross motor development and reach on sound as critical tools for the development of the blind child. *Brain & Development,* 24, 269–275.
8. Ghasia F., Burnstroom J., Gordon M., Tychsen L. (2008). Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. *Invest Ophthalmol Vis Sci,* 49, 572-580.
9. Good, W.V., Jan, J.E., Burden, S.K., Skoczenski, A., Candy, R. (2001). Recent advances in cortical visual impairment. *Dev Med Child Neurol,* 43(1), 56-60.
10. Haley, S.M., Coster, W.J., Ludlow, L.H., Haltiwanger, J.T. & Andrellos, P.J. (1992). *Pediatric Evaluation of Disability Inventory: Development, standardization, and administration manual.* Boston, MA: New England Medical Centre Inc/PEDI Research Group.
11. Harries, N., Kassirer, M., Amichai, T., Lahat, E. (2004). Changes over years in Gross Motor function of 3-8 year old children with Cerebral Palsy: Using the Gross Motor Function Measure (GMFM-88). *IMAJ,* 6, 408-411.
12. *International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation). (2008). (1sted.).The Netherlands: Dutch WHO-FIC Collaborating Centre, Bohn Stafleu van Loghum.*
13. Jane, C. Edmond & Rod, Foroozan. (2006). Cortical visual impairment in children. *Current Opinion in Ophthalmol*, vol 17, 509-512.
14. Ketelaar, M., Van Petegem-van Beek, E., Veenhof, C., Visser, J., Vermeer, A. (2003). Gross Motor Function Measure. University of Utrecht. *Child Phys Ther,* 39, 5-7
15. Levtzion-Korach, O., Tennenbaum, A., Schnitzer, R., & Ornoy, A. (2000). Early motor development of blind children. *Journal of Paediatrics and Child Health,* 36, 226–229.
16. Prechtl, H., Cioni, G., Einspieler, C., Bos, A., & Ferrari, F. (2001). Role of vision on early motor development: Lessons from the blind. *Developmental Medicine and Child Neurology,* 43, 198–201.
17. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M., Bax, M. (2007). Definition and classification of cerebral palsy. *Dev Med Child Neurol,* 49(6), 480.
18. Russell, D.J., Rosenbaum, P.L., Avery, L.M., Lane, M. (2002). *Gross Motor Function Measure (GMFM-66 and GMFM-88) User's Manual.* London, United Kingdom: MacKeith Press.
19. Salavati, M., Rameckers, E.A.A., Steenbergen, B., & Schans van der, C.P. (2014). Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). *European Journal of Physical Therapy*, 16(3), 159–167.
20. Salavati, M., Waninge, A., Rameckers, E.A.A., de Blécourt, A.C.E., Krijnen, W.P., Steenbergen, B & Schans van der, C.P. (2015a). Reliability of modified paediatric evaluation of disability inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment. *Res Dev Dis,* 37, 189-201.
21. Salavati, M., Krijnen, W.P., Rameckers, E.A.A., Looijestijn, P., Maathuis, C.G.B., Schans van der, C.P. & Steenbergen, B. (2015b). Reliability of the modified Gross Motor Function Measure-88 (GMFM-88) for children with both Spastic Cerebral Palsy and Cerebral Visual Impairment: A preliminary study. *Res Dev Dis,* 45-46, 32-48.
22. Schenk-Rootlieb, A.J.F., Van Nieuwenhuizen, O., Schiemanck, N., Van der Graaf, Y., Willemse, J. (1993). Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. *Children Care Health Dev,* 19, 411-423.
23. Scholtes, V.A., Becher, J.G., Comuth, A., Dekkers, H., Van Dijk, L., Dallmeijer, A.J. (2010). Effectiveness of functional progressive resistance exercise strength training on muscle strength and mobility in children with cerebral palsy: a randomized controlled trial. *Dev Med Child Neurol,* 52(6): 107-113.
24. Stiers, P., Vanderkelen, R., Vanneste, G., Coene, S., De Rammelsere, M., Vandenbussche, E. (2002). Visual-perceptual impairment in a random sample of children with cerebral palsy. *Dev Med Child Neurol,* 44, 370-382.
25. Visser, L., Ruiter, A.J., Meulen van der, F., Ruijssenaars, A.J.J.M., Timmerman, E. (2014). Validity and suitability of the Bayley-III Low Motor/Vision version: A comparative study among young children with and without motor and/or visual impairments. *Pediatr Phys Ther*, 26 (1), 57-67.

6 |

Development and validity of a Cerebral Visual Impairment Motor Questionnaire for children with Cerebral Palsy

M. Salavati[a, b], A. Waninge[a, b], E.A.A. Rameckers,[c, d, e], J. van der Steen, PhD[a, f], W.P. Krijnen[b], C.P. van der Schans[b, g], B. Steenbergen[h, i]

[a] Royal Dutch Visio, centres of expertise for blind and visually impaired people, The Netherlands

[b] Hanze University of Applied Sciences Groningen, research group Healthy Ageing, allied health care and nursing; Groningen, The Netherlands

[c] Maastricht University, Department of Rehabilitation Medicine, School for Public Health and Primary Care (CAPHRI); Maastricht, The Netherlands

[d] Adelante Center of Expertise in Rehabilitation and Audiology; Valkenburg and Hoensbroek, The Netherlands

[e] AVANSplus, advanced training for professionals working with children with movement disabilities; Breda, The Netherlands

[f] Erasmus MC, Department of Neuroscience, Rotterdam, The Netherlands

[g] University of Groningen, University Medical Centre Groningen, Department of Rehabilitation Medicine; Groningen, The Netherlands

[h] Radboud University Nijmegen, Behavioural Science Institute; Nijmegen, The Netherlands

[i] Australian Catholic University, School of Psychology; Melbourne, Australia

Submitted as:

Salavati, M., Waninge, A., Rameckers, E.A.A., Steen van der, J., Krijnen, W.P., Schans van der, C.P. & Steenbergen, B. Development and validity of a Cerebral Visual Impairment Motor Questionnaire for children with Cerebral Palsy.

Abstract

Purpose: The objectives of this study were:

- to develop two Cerebral Visual Impairment Motor Questionnaires (CVI-MQ's) for children with Cerebral Palsy (CP): one for children with Gross Motor Function Classification (GMFCS) levels I-II-III and one for children with GMFCS levels IV-V; second,
- to describe their face validity and usability;
- to determine their sensitivity and specificity.

Methods: The initial versions of the two CVI-MQ's were developed based on literature. Then the Delphi method was used among two groups of experts, one familiar with CVI, in order to gain consensus about face validity and usability. The sensitivity and specificity of the CVI-MQ's were subsequently assessed in 82 children with CP with (*n*= 39) and without CVI (*n*=43). With the Receiver Operating Curve (ROC) the cut-off scores were determined to detect possible presence or absence of CVI in children with CP.

Results: Both questionnaires showed very good face validity (percentage agreement above 96%) and good usability (percentage agreement 95%) for practical use. The CVI-MQ version for GMFCS levels I-II-III had a sensitivity of 1.00 and specificity of 0.96, with a cut-off score of 12 points or higher, and the version for GMFCS levels IV-V had a sensitivity of 0.97 and a specificity of 0.98, with a cut-off score of 8 points or higher.

Conclusion: The CVI-MQ is able to identify at-risk children with CP for the probability of having CVI.

1. Introduction

Cerebral visual impairment (CVI) is the major cause of visual impairment in developed countries (Good, Jan, DeSa, Barkovich, Groenveld & Hoyt, 1994; Ortibus, Verhoeven, Cock De, Sunaert, Casteels, Laenen, Schoolmeesters, Buyck, & Lagae, 2011; Liew, Michaelides, & Bunce, 2015). Approximately 30% of children diagnosed with various forms of cerebral palsy (CP) also suffer from CVI (Schenk-Rootlieb, Van Nieuwenhuizen, Schiemanck, Van der Graaf, & Willemse, 1993; Dutton, & Jacobson, 2001; Stiers, Vanderkelen, Vanneste, Coene, De Rammelsere, & Vandenbussche, 2002; Da Costa, Salmao, Berezovsky, De Haro, & Ventura, 2004; Ghasia, Burnstroom, Gordon, & Tychsen, 2008). CP is defined as a group of permanent disorders of movement and posture development that cause activity limitations (Rosenbaum, Paneth, Leviton, Goldstein, & Bax, 2007).

CVI is generally defined as a deficiency in visual function, due to damage or malfunction of visual pathways and visual centres in the brain, including the optic radiations, the occipital cortex and visual associative areas. CVI is a result from impaired processing of visual information in the presence of a (nearly) intact ophthalmological system (Dutton, & Jacobson, 2001; Goodale, & Milner, 1992; Dutton, Saaed, , Fahad, Fraser, McDaid, McDade, 2004; Edmond, & Foroozan, 2006; Fazzi, Bova, Uggetti, Signorini, Bianchi, Maraucci, Zoppello, & Lanzi, 2004; Fazzi, Signorini, LA Piana, Bertone, Misefari, Galli, Balottin, & Bianchi, 2012). CVI has an impact on all aspects of a child's developmental milestones, including reaching and walking, and children with CP and CVI develop more slowly in the areas of self-care, mobility and social function than children with CP without CVI. In addition, CVI can result in a delayed motor development in children with CP (Da Costa *et al.,* 2004; Ghasia *et al.,* 2008; Fazzi, Signorini, LA Piana, Bertone, Misefari, Galli, Balottin, & Bianchi, 2012). When a child with CP exhibits a limitation of daily activities and slow motor processing and performance speed, this may not only originate from a delay in motor and / or mental development but also from a visual impairment (Boot, Pel, Evenhuis, & van der Steen, 2012).

Paediatric physical therapists and occupational therapists are often the first professionals to assess and treat children with CP at the level of motor functioning. This puts them in a position to identify red flags for CVI (higher visual risk factor) when screening these children. Such red flags allow professionals to review the impact of CVI on the observed motor behaviour and to ensure the identification of signs and symptoms of CVI in children with CP. Because red flags for CVI are lacking in rehabilitation centres, it is important to develop a CVI screening tool to identify these signs. These children could be referred to an ophthalmologist and paediatric neurologist in order to establish full diagnosis and exclude the presence of ocular visual impairments.

Input from visual systems are important sources of information about the body's position and movement in space with respect to gravity and the environment. CVI has a large impact on motor development and it hinders normal visuomotor development by affecting aspects such as accuracy of distance estimation, thereby influencing visually guided motion (Ortibus *et al.,* 2011). Children with CP have many limitations, which hamper a thorough standardised assessment of visual functioning, and the current assessments do not account for the presence of visual impairments in children with CP. Hence professionals have to rely on observations or findings from the child's history to diagnose CVI (Ortibus *et al.,* 2011; Salavati, Rameckers, Steenbergen, & Schans van der, 2014).

A motor screening tool consisting of items related to the contribution of visual perception to perform a motor activity may be helpful for paediatric physical therapists and occupational therapists. Early identification of CVI may lead to an emphasis on the right determinants and a proper focus of the comprehensive treatment, which helps the children in their development. Thus far, the available CVI screening tools have focused on screening visual dysfunction and no validated CVI screening tool is yet available to screen children with CP to identify the possible contribution of CVI on motor impairment(Ortibus *et al.,* 2011; Dutton, & Jacobson, 2001; Steendam, 2008; Dutton, Calvert, Cockburn, Ibrahim, & Macintyre-Beon, 2012). Paediatric physical therapists and occupational therapists could benefit from having a screening tool at their disposal to determine the extent to which CVI contributes to delays in motor disabilities in children with CP (Boot *et al.*, 2012; Salavati *et al.,*

2014; Dutton, 2013; Salavati, Waninge, Rameckers, de Blécourt, Krijnen, Steenbergen, & Schans van der, 2015a; Salavati, Krijnen, Rameckers, Looijestijn, Maathuis, Schans van der, & Steenbergen 2015b).

Gross motor function of children with CP can be classified into five different severity levels using the Gross Motor Function Classification System (GMFCS), where level I indicates the least and level V the most functional limitation (Rosenbaum *et al.,* 2007). Because of these large functional differences, that is, children who can walk and children who are wheelchair-dependent, we decided to develop two different CVI Motor Questionnaires (CVI-MQ): the CVI-MQ for children with CP with GMFCS I-II-III and CVI-MQ for children with GMFCS IV-V. The content of the CVI-MQ for children with GMFCS levels I-II-III includes motor items for children about higher motor skills such as walking, stair-climbing and jumping, while the CVI-MQ for children with GMFCS levels IV-V contains motor skills such as rolling over and reaching. Developing one single CVI-MQ for all children with GMFCS I through V would have had the effect that a high number of items would be 'not applicable' for children with GMFCS IV and V, and it would also take more time to fill in the questionnaire. Developing two different CVI-MQ's thus meets the motor capabilities needs of both groups.

Our aims were first to develop two CVI-MQ's for children with CP, second to describe their face validity and usability, and third to determine their sensitivity and specificity to detect a possible presence of CVI in children with CP.

2. Methods

The study was conducted in three phases. First, based on existing literature and according to the GMFCS levels, we developed two CVI-MQ's for children with CP: one for GMFCS levels I-II-III and one for GMFCS levels IV-V. Second, the Delphi method was used to gain consensus about the face validity among a panel of experts and to gain insight into the usability of the two MQ's. The predetermined goal for the usability was to reach a consensus of 95% agreement on the two CVI-MQ's among experts that were not familiar with CVI. Third, data of children with CP with and without CVI were used to calculate sensitivity and specificity of the two CVI-MQ's in order to detect the probability of presence or absence of CVI in children with CP and to determine cut-off scores.

2.1. Phase 1 – developing CVI-MQ's

We electronically searched for relevant literature published between May 1995 and December 2015 using the PubMed, PsychLit, EMBASE, PEDro and MEDLINE databases using Medical Subject Headings (MeSH) terms and text words. The following queries were used: "Cerebral Palsy" [MeSH] *AND/OR* "Cerebral Visual Impairment" *OR* "Cortical Visual Impairment" *OR* "Cortical blindness" *OR* "vision disorder" [MeSH], in combination with *AND* "Gross Motor Classification System I, II, III, IV, V" *AND* "motor activity" *OR* "functional skills" *OR* "self-care" *OR* "mobility" *AND* "screening" *OR* "observation" *OR* "questionnaire".

The developed multiple-choice items in the two CVI-MQ's were carefully selected on the basis of the current available questionnaires used by 1) the home intervention team for children with CVI at Royal Dutch Visio and Bartiméus (centres of expertise for blind and visually impaired people in the Netherlands); 2) the visual skills inventory available from the studies of Dutton et al (Dutton, & Jacobson, 2001; Dutton *et al.,* 2004; Dutton *et al.,* 2012 & 2013); 3) literature reviews of features of CVI in children; 4) the adapted version of the paediatric evaluation of disability inventory, Dutch version (PEDI-NL); and the Gross Motor Function Measure-88 (GMFM-88) for children with CVI (Edmond, & Foroozan, 2006; Fazzi *et al.,* 2004; Dutton, 2013; Salavati *et al.,* 2015a; Salavati *et al.,* 2015b; Russell, & Rosenbaum, 2002; Haley, Coster, Ludlow, Haltiwanger, & Andrellos, 1992).

2.2. Phase 2 – Adaptation of CVI-MQ's

2.2.1. Adaptation

The multiple-step Delphi method was used to gain consensus about the content of both developed CVI-MQ's among a panel of experts familiar with CVI and a panel that was unfamiliar with it. Both groups of experts were familiar with CP. The Delphi method was applied in a series of sequential questionnaires or 'rounds', interspersed by controlled feedback, in order to seek the most reliable consensus of opinion from a purposeful sample of experts (Powell, 2003; Gracht von der, 2012). In this study, face validity was defined

Table 1. Cerebral Visual Impairment Motor Questionnaire (CVI-MQ) for children with Cerebral Palsy (CP), GMFCS I-II-III. Results of percentages consensus experts after third Delphi round. Percentage of agreement for test result on each question: 'Yes', 'No', 'Not applicable'.

CVI-MQ for children with GMFCS I-II-III		Score:			Percentage consensus experts after third Delphi round
Item	**Gross motor skills**	**Percentage YES**	**Percentage NO**	**Percentage not applicable**	
1	The child belly-crawls if stimulated by movement*, sound production*, fluorescence*, high-contrast* toys, or verbal support*. **Circle as applicable*	70%	23%	7%	89.5%
2	The child crawls if it is stimulated by movement*, sound production*, fluorescence*, high-contrast* toys, or verbal support*. **Circle as applicable*	74%	23%	3%	89.5%
3	The child bumps into moved toys or furniture when it crawls.	44%	54%	2%	100%
4	The child is more uncertain when it walks in an unfamiliar environment compared to a familiar environment.	51%	49%	0%	100%
5	The child has difficulty anticipating differences in height when it walks, for example when stepping down from the sidewalk onto the road.	56%	42%	2%	94.7%
6	The child walks slower in unfamiliar environments.	56%	44%	0%	94.7%
7	The child will walk up an unfamiliar staircase one step at the time, always leading with the same foot, whereas it will walk up a familiar staircase with alternating feet at each step.	54%	37%	9%	94.7%
8	The child will walk down an unfamiliar staircase one step at the time, always leading with the same foot, whereas it will walk up a familiar staircase with alternating feet at each step.	54%	37%	9%	94.7%
9	The child bumps into obstacles/persons when it walks.	44%	56%	0%	94.7%
10	The child bumps into obstacles/persons when it runs.	44%	47%	9%	94.7%
11	The child walks significantly slower when there is no person to follow.	44%	54%	2%	94.7%
12	The child hesitates when it moves from one room to another; this occurs when the child both leaves and enters a room.	44%	54%	2%	100%
13	The child falls* and/or trips* over obstacles. **Circle as applicable*	49%	44%	7%	100%
14	The child does not jump off an elevated platform.	46%	33%	21%	89.5%
15	The child does not jump forwards*, sidewards *or backwards*. **Circle as applicable*	44%	35%	21%	89.5%
16	When catching a ball, the child misses a non-sound-producing* and/or non-fluorescent*, non-high-contrast* ball more often than a sound-producing*, fluorescent*, high-contrast* ball. **Circle as applicable*	44%	51%	5%	100%
17	The child kicks behind/next to the ball when kicking a non-sound-producing*, non-fluorescent*, lower-colour*/-contrast* ball.**Circle as applicable*	44%	49%	7%	100%
18	The child rolls*/throws* a ball towards a person if there is verbal support. **Circle as applicable*	56%	39%	5%	89.5%
19	The child has difficulty estimating the distance and speed of other road users.	58%	35%	7%	100%
20	The child has difficulty finding the route to the class or the school playground when walking at school.	44%	54%	2%	100%
Fine motor skills / Reaching and grasping					
21	The child reaches behind/bumps into small objects. The child only grabs the object after touching it.	44%	54%	2%	100%
22	The child manipulates the toy with its hands instead of exploring it with its eyes.	51%	49%	0%	100%
23	The child has difficulty copying figures with a pencil.	30%	42%	28%	100%
24	The child reaches more precisely when reaching for moving objects.	47%	51%	2%	100%
25	The child reaches more precisely towards sound-producing*, high-contrast*, fluorescent*, illuminating* objects compared to non-sound-producing*, non-high-contrast*, non-fluorescent*, non-illuminating* objects. **Circle as applicable*	47%	51%	2%	100%
26	The child does not reach for and look at an object at the same time.	47%	53%	0%	100%
27	The child reaches towards toys but has difficulty finding the toys in a crowded background. For example, finding a block on a full table or in a basket filled with toys.	47%	51%	2%	100%

as an opinion of CVI experts on the CVI-MQ's, who commented from their perspective on the developed CVI-MQ's. We therefore asked the experts familiar with CVI whether or not the CVI-MQ's measured presence or absence of CVI in children with CP.

To investigate the face validity of developed CVI-MQ's, first we invited a group of experts familiar with CP and CVI by e-mail. These experts worked at Royal Dutch Visio and Bartiméus. To assess usability of CVI-MQ's, we also invited a group of experts not familiar with CVI by posting an invitation on the website of their organisation and by e-mail. Due to the fact that mostly the experts not familiar with CVI will use CVI-MQ's, it is important that these questionnaires were appropriate for those experts. The experts unfamiliar with CVI worked at private practices and healthcare practices. Purpose of the study and required procedures were explained to both groups, and they were subsequently asked for consent to participate in the study. Following consent, the experts were asked about their age, profession and working experience.

2.2.2. Data collection for adaptation

Firstly, the experts familiar with CVI participated in the adaptation process by studying the two developed CVI-MQ's, and giving their comment on the content of questionnaires. Feedback information could be written about any items, and the experts were specifically asked whether each item was appropriate for children with CVI. If it wasn't, we asked what needed to be added or changed to make it appropriate. We also asked the experts if they thought that items needed to be added to the CVI-MQ's.

The predetermined goal was to reach an experts' consensus of 65% on each item after the first round, 75% after the second round and 85% after the third round for agreement with each item as well as content of the CVI-MQ (Powell, 2003; Gracht von der, 2012). The experts familiar with CVI were also asked to explain and justify their comments on each item for both CVI-MQ's (Tables 1 and 2).

2.2.3. First Delphi round

The experts familiar with CP and CVI were invited to study both CVI-MQ's (Tables 1 and 2), and then gave comments individually on the content of each item. We asked these experts which items needed to be changed or added to the two CVI-MQ's and why, in order to make the content appropriate for children with CVI.

2.2.4. Second and third Delphi rounds

After receiving the comments of experts familiar with CVI on the items of both CVI-MQ's we processed all of the suggestions in the questionnaires and resubmitted them twice to these experts. We asked them whether the content of each item and instruction part of CVI-MQ's was appropriate for children with CVI. To determine usability, we also asked how long it took to answer the items on each CVI-MQ, when the expert was familiar with the child with CP.

After the third Delphi round we asked the experts not familiar with CVI to comment on the two CVI-MQ's and gave their individual comments. Our goal was to investigate the usability of developed CVI-MQ's for those experts not familiar with CVI. We asked those experts whether the items and the instruction part of CVI-MQ's were clearly stated. We also asked how long it took to answer all the items on each CVI-MQ, when expert was familiar with the child with CP.

2.3. Phase 3 – Sensitivity and specificity of CVI-MQ's

Children with any type of CP with and without CVI were recruited from Royal Dutch Visio and allied health care practices. Inclusion criteria were presence of all types of CP and CVI, mild or moderate intellectual disability, and age at testing of the CVI-MQ for children between 4 and 16 years. Level of intellectual disability was drived from the children's medical files. Children with a syndrome (for example Down syndrome) and hearing difficulties (>30 db hearing loss) , and corrected vision <0.3 and/ or field of vision < 30° were excluded. Children with severe or profound intellectual disability (IQ<40) were also excluded. The diagnosis of CP and the classification according to GMFCS level were taken from the children's medical files and judged by a rehabilitation specialist. Based on possible effects on motor functioning, we also collected background data on gender as well as prevalence of epilepsy and speech/language development according to the International Classification of Functioning, Disability and Health for Children and Youth (International Classification of Functioning, Disability and Health, Child & Youth version, 2008). At the level of speech/ language development, the collected data were:

Table 2. Cerebral Visual Impairment Motor Questionnaire (CVI-MQ) for children with Cerebral Palsy (CP), GMFCS IV-V. Results of percentages consensus experts after third Delphi round. Percentage of agreement for test result on each question: 'Yes', 'No', 'Not applicable'.

CVI-MQ for children with GMFCS IV-V		Score:			
Item	**Gross motor skills**	**Percentage YES**	**Percentage NO**	**Percentage not applicable**	**Percentage consensus experts after third Delphi round**
1	The child turns it head to follow, if encouraged by sound production*, fluorescence*, high-contrast* toys or verbal stimulation. **Circle as applicable*	64%	31%	5%	94.7%
2	The child lifts its head when lying on its stomach, if encouraged by some production*, fluorescence*, high-contrast* toys or verbal stimulation. **Circle as applicable*	64%	31%	5%	94.7%
3	From a sitting position the child lifts its head, if encouraged by sound production*, fluorescence*, high-contrast* toys or verbal stimulation.**Circle as applicable*	64%	31%	5%	94.7%
4	The child belly-crawls if encouraged by some production*, fluorescence*, high-contrast* toys or verbal stimulation. **Circle as applicable*	42%	28%	30%	94.7%
5	The child bumps into moved toys or furniture when it belly-crawls.	31%	36%	33%	100%
6	The child crawls/belly-crawls slower in an unknown environment with the same surface as a known environment.	28%	36%	36%	94.7%
7	The child has difficulty finding the route to the class or school playground when driving a wheelchair (mechanic/electric).	36%	51%	13%	100%
8	The child bumps into obstacles/persons when driving a wheelchair (mechanic/electric).	43%	44%	13%	100%
Fine motor skills/ reaching and grasping					
9	The child reaches more precisely for moving objects than for non-moving objects.	51%	49%	0%	94.7%
10	The child reaches more precisely for sound-producing*, high-contrast*, fluorescent*, illuminating* objects than for non-sound-producing*, non-high-contrast*, non-fluorescent*, non-illuminating* objects.**Circle as applicable*	51%	49%	0%	100%
11	The child looks away when it grabs an object.	54%	46%	0%	100%
12	The child reaches for a toy but has difficulty finding the toy in a crowded background. For example, finding a block on a full table or in a basket filled with toys.	54%	46%	0%	100%
13	The child grabs an object if it produces sound.	64%	36%	0%	89.5%
14	The child explores*/manipulates* toys with its mouth or hands instead of exploring it with its eyes. **Circle as applicable*	46%	54%	0%	89.5%

d3101 = understands simple spoken messages; d3102 = understands complex spoken messages; d330 = speaks (Table 3). We registered level of GMFCS and type of CP (unilateral or bilateral), level of intellectual disability and age at which the CVI-MQ was administered.

The diagnosis of CVI was determined based on the results of ophthalmological and psychological/ neuropsychological assessments and on the assessment data reported by a developmental coach specialised in working with children with visual impairments. On this basis, the diagnosis of CVI was determined by the following criteria: a normal or near-normal eye exam (corrected vision >0.3 and/ or field of vision >30°) performed by an ophthalmologist; history or presence of neurological problems; presence of behavioural responses to visual stimuli unique to CVI. These include strong colour preferences, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non-purposeful gaze, difficulty with distance viewing, absent or atypical visual reflexes, difficulty with visual novelty, and absence of visually guided reach (Dutton, & Jacobson, 2001; Stiers *et al.,* 2002).

The study was approved by the Medical Ethical Committee (METc-2015-048) of University Medical Center Groningen (UMCG), Groningen, The Netherlands. Written informed consent was obtained from private practices and healthcare practices.

2.4. Design

Data of the two CVI-MQ's were used to calculate sensitivity (the proportion of positive cases that were classified as positive) and specificity (the proportion of negative cases that were classified as negative) to detect the probability of presence or absence of CVI in children with CP (Rosner, 2000). The experts familiar with children with CP with and without CVI administered the two CVI-MQ's (CVI-MQ for children with GMFCS I-II-III and CVI-MQ for children with GMFCS IV-V).

2.5. Statistical analyses

Data were analysed using Statistical Package for Social Sciences (SPSS), v.22 software. We used the Receiver Operating Curve (ROC) depicting of sensitivity versus 1-specificity (1 – true positive proportion) for different values of the cut-off point. The Area Under the Curve (AUC) represented an overall accuracy measured covering all possible interpretation thresholds. An area of 0.9-1.0 represented an excellent value for a test, a value between 0.8-0.9 is good, between 0.7-0.8 fair, between 0.6-0.7 poor and between 0.5-0.6 fail. AUC values closer to 1 are preferable (Eng, 2005). An optimal cut-off point was determined with sensitivity and specificity rates set at good value (0.8-0.9). We analysed the CVI-MQ's data to investigate their predictive value to presume the presence of CVI in children with CP. Sensitivity and specificity of the two CVI-MQ's were analysed from children with CP, with and without CVI. Because the CVI-MQ's were meant to identify at-risk children with CP for the probability of having CVI, and to refer for full diagnosis, it was important to maximise sensitivity so as to miss the fewest possible number of cases. To evaluate diagnostic accuracy we calculated positive predictive value, negative predictive value, positive likelihood ratio, negative likelihood ratio and confidence interval of both CVI-MQ's. We also created scatter plots to visualize the distribution of CVI-MQ measurements for children with CP where CVI is present and absent. To report percentage of agreement we gave this percentage or each item of the two CVI-MQ's.

3. Results

3.1. Adaptation of CVI-MQ's

Nineteen health experts familiar with CP and CVI confirmed their willingness to participate in the development of the two CVI-MQ's; five were occupational therapists, 13 paediatric physical therapists, and one a behavioural scientist. Mean (SD) age of the experts was 51 (10) years and their mean years (SD) of experience with children with CP and CVI was 20 (9).

In addition, to determine usability of two CVI-MQ's, 20 health experts familiar with CP but not familiar with CVI participated in the development of the two questionnaires; sixteen of them were paediatric physical therapists and four occupational therapists. Their mean (SD) age was 46 (11) years and mean years (SD) of experience with children with CP was 19 (11).

3.1.1. First Delphi round

On the CVI-MQ for children with GMFCS I-II-III (Table 1), 68% of the experts familiar with CVI agreed about the content of items 5, 6, 9, 10, 12, 17, 18, 20, 21, 24-28 (agreement percent 74%-89%). Most comments were about items 1-4, 7, 8, 11, 13-16, 19, 22, 23 (agreement percent 42%-63%). For example, they suggested that for item 3 it is important to add the word 'moved' to the item: 'The child bumps into moved toys or furniture when it belly-crawls'. The experts suggested changing item 4 from 'The child holding on to a person in an unfamiliar environment when it walks' into 'The child is more uncertain when it walks in an unfamiliar environment compared to a familiar Environment'.

Item 5 was changed by deleting 'the child has difficulty without verbal support' from the item, 'The child has difficulty anticipating differences in height when it walks, for example when stepping down from the sidewalk onto the road'. With respect to stairs (items 7-8, Table 1), the experts suggested having two separate items, one for climbing stairs and another for walking down stairs.

On the CVI-MQ for children with GMFCS IV-V (Table 2), 71% of the experts familiar with CVI agreed about items 5-8, 10-13 (agreement percent 74%-89%).

The experts suggested combining two items by deleting 'talking at the same time' from item 8 'The child bumps into obstacles/persons when driving a wheelchair (mechanic/electric)', to make it suitable for children with CVI. Most experts' comments were about items 1-4, 9, 14 (agreement percent 53%-63%). They suggested that by adding information on instruction those items would be suitable for children with CVI. The experts suggested adding the item 'The child grabs an object if it produces sound' (item 13, Table 2).

3.1.2. Second Delphi round

Ninety-seven percent of the experts familiar with CVI agreed on the content of items of the CVI-MQ for children with GMFCS I-II-III (Table 1), and 96% agreed on the items of the CVI-MQ for children with GMFCS IV-V (Table 2). Because of the high percentage of agreement we sent both CVI-MQ's to experts familiar with CVI as well as to those unfamiliar with CVI.

On the CVI-MQ for children with GMFCS I-II-III (Table 1), the highest level of agreement (100%) among experts was for items 3, 4, 12, 13, 16, 17, 19-23, 25-27. The percentage of agreement for items 1, 2, 5-11, 14, 15, 18, 24 was between 89.5% and 94.7%. One expert suggested that these items could also be used for children with ocular visual impairment.

On the CVI-MQ for children with GMFCS IV-V (Table 2), the highest level of agreement (100%) among experts was for items 5, 7, 8, 10-12. The percentage of agreement among experts for items 1-4, 6, 9, 13, 14 was between 90% and 95%. One expert suggested that item 14 could also be caused by a behavioural impairment.

3.1.3. Third Delphi round

After receiving the experts' comments we processed the proposed adaptations and resubmitted them to the two groups of experts. The usability results showed a consensus of 95% agreement on each CVI-MQ among those experts not familiar with CVI. The experts familiar with CVI indicated 12 (5) mean (SD) minutes to administer the CVI-MQ for children with GMFCS I-II-III and the experts not familiar with CVI indicated 14 (10) mean (SD) minutes to administer it (Table 1). The experts familiar with CVI indicated 9 (4) mean (SD) minutes to administer the CVI-MQ for children with GMFCS IV-V and the experts not familiar with CVI indicated 14 (10) mean (SD) minutes (Table 2).

3.2. Sensitivity and specificity of CVI-MQ's

The CVI-MQ's filled out between June 2015 and November 2015. We collected data from 82 children with both CP and CVI (*n*=57 boys and *n*=25 girls). Table 3 shows the children's characteristics. All children with CVI were included in our study and no selection was done based on subtypes. We therefore assumed that different subtypes are represented in our study.

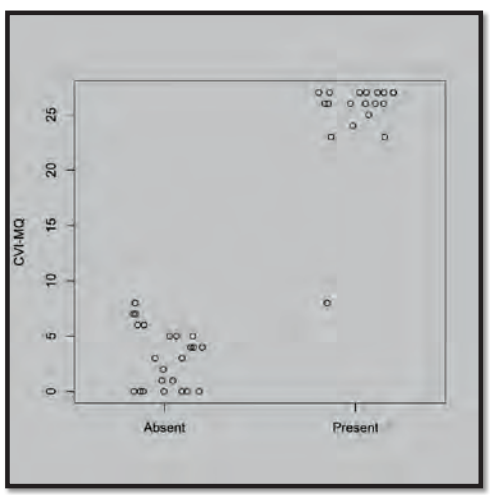


Figure 1. Scatter plot of CVI-MQ scores for children with GMFCS I-II-III. Absent, CVI is absent; Present, CVI is present; CVI-MQ, Cerebral Visual Impairment Motor Questionnaire for children with CP.

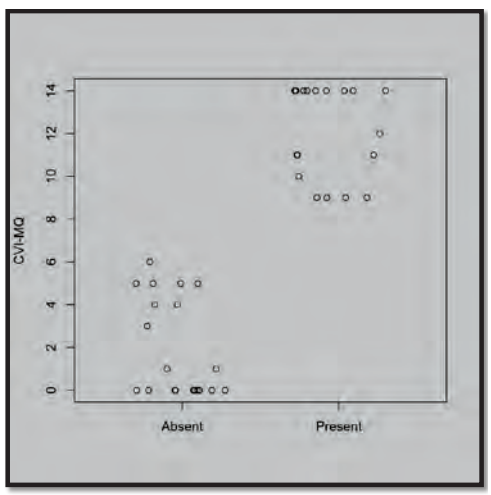


Figure 2. Scatter plot of CVI-MQ scores for children with GMFCS IV-V. Absent, CVI is absent; Present, CVI is present; CVI-MQ, Cerebral Visual Impairment Motor Questionnaire for children with CP.

Table 3. Characteristics of CP children with and without CVI

Characteristic	Children with CVI	Children without CVI
Age in months, mean (SD), min-max		
- GMFCS I-II-III	119 (44), 50-192	120 (43), 60-192
- GMFCS IV-V	119 (36), 62-189	134 (42), 72-201
Gender, male /female (*n) (%*)		
- GMFCS I-II-III	11 (55) /9 (45)	16 (70) /7 (30)
- GMFCS IV-V	15 (79) /4 (21)	15 (75) /5 (25)
Type of cerebral palsy (GMFCS I-II-III-IV-V):		
spastic (*n)* (%)	36 (92)	41 (95)
dyskinetic (*n)* (%)	3 (8)	2 (5)
GMFCS I (*n*) (%)	bilateral 11 (28), unilateral left 1 (3)	bilateral 3 (7), unilateral left 1 (2), unilateral right 4 (9)
GMFCS II (*n*) (%)	2 (5) bilateral	6 (14) bilateral
GMFCS III (*n*) (%)	6 (15) bilateral	9 (21) bilateral
GMFCS IV (*n*) (%)	9 (23) bilateral	16 (37) bilateral
GMFCS V (*n*) (%)	10 (26) bilateral	4 (9) bilateral
Speech/language development (GMFCS I-II-III-IV-V):		
ICF-CY, d3101 = understands simple spoken messages (*n*) (%)	35 (90)	42 (98)
ICF-CY, d3102 = understands complex spoken messages (*n*) (%)	34 (87)	38 (88)
ICF-CY, d330 = speaks (*n*) (%)	22 (56)	38 (88)
Level of intellectual disability (GMFCS I-II-III-IV-V): mild/moderate (*n*) (%)	14 (36)/25 (64)	23 (54)/20 (46)
Presence of epilepsy (GMFCS I-II-III-IV-V): yes/no (*n*) (%)	7 (18)/32 (82)	5 (12)/38 (88)

GMFCS, Gross Motor Function Classification System; n, numbers; ICF-CY, International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation).

Table 4. Sensitivity and specificity values and cut-off scores for the two CVI-MQ's.

	GMFCS I-II-III	GMFCS IV-V
Cut-off scores	12	8
Sensitivity (point estimates and 95% CI)	1.00 (0.76- 1.00)	0.97 (0.79- 1.00)
Specificity (point estimates and 95% CI)	0.96 (0.78- 1.00)	0.98 (0.80- 1.00)
Area Under the Curve (AUC) value	0.99	1.00
Standard error	0.002	0.000
Positive predictive value (point estimates and 95% CI)	0.95 (0.76-1.00)	0.97 (0.79- 1.00)
Negative predictive value (point estimates and 95% CI)	1.00 (0.78-1.00)	0.98 (0.80- 1.00)
Positive likelihood ratio (point estimates and 95% CI)	23.00 (3.38- 156.39)	40.95 (2.65- 633.88)
Negative likelihood ratio (point estimates and 95% CI)	0.00 (0.00- 0.00)	0.03 (0.00- 0.40)
Asymptomatic significance	<0.001	<0.001
Asymptomatic 95% C.I. (lower bound-upper bound)	0.000-0.006	0.000-0.000

C.I., confidence interval.

The scatter plots show the distribution of CVI-MQ's scores of children with CP, with and without CVI for both GMFCS groups (Figs. 1 and 2). Figure 1 shows that children without CVI and GMFCS I-II-III have a score below 10 and children with CVI and GMFCS I-II-III have a score above 10, except for one child. Figure 2 shows that children without CVI and GMFCS IV-V have a score below 8 and children with CVI and GMFCS IV-V have a score above 8. A cut-off score of 12 or higher (Figure 1, Table 4) indicates probability of presence of CVI in a child with CP and GMFCS I-II-III. A cut-off score of 8 or higher indicates probability of presence of CVI in a child with CP and GMFCS IV-V (Figure 2, Table 4).

Tables 1 and 2 show the frequency in percentage scores (Yes, No, Not applicable) for each item on the both CVI-MQ's. For a large number of items, the experts gave as answer 'Not applicable'. The reasons were that the child was too young or too old to perform a motor skill, or that the motor skill was too difficult to perform in relation to a child's GMFCS level.

The results of the ROC curve for CVI-MQ, GMFCS levels I-II-III are sensitivity 1.00 and specificity 0.96, and for CVI-MQ, GMFCS levels IV-V sensitivity 0.97 and specificity 0.98. This indicates excellent sensitivity and specificity for identifying at-risk children with CP with the possibility of having CVI.

Figure 1 shows that one child (GMFCS I) with CVI received a lower score on the items of the CVI-MQ for children with GMFCS I-II-III, compared with the other children with CVI.

The two CVI-MQ's measure the degree of CVI in children with CP, where higher scores indicate higher levels of probability to predict the presence of CVI in children with CP. The sum score of each CVI-MQ represents this interpretation if all items are applicable. Each CVI-MQ counts the number of CVI cases implied by the items. If not all items were applicable, then the non-applicable items were not taken into account and the sum score was applied to the applicable items. Table 4 presents the values of sensitivity and specificity and corresponding cut-off scores for both CVI-MQ's.

4. Discussion

The CVI-MQ's for children with CP, with GMFCS I-II-III and GMFCS IV-V both have good face validity and are potentially usable tools to detect children suspected of having CVI. They have excellent sensitivity and specificity as well as a positive/ negative predictive value with feasible cut-off scores.

During the Delphi rounds, the CVI experts suggested several issues that may influence validity. First, the difficulty with moving in an unfamiliar environment compared to a familiar environment (items 4, 6-8, 11, 14, 15, Table 1; item 6, Table 2) could be due to not feeling safe/secure enough. On the other hand, moving without difficulty in a familiar environment could be the result of automated motor patterns rather than the familiarity of the environment. Different types and strengths of lighting (halogen, LED, fluorescent) in different rooms may cause the child to hesitate when it moves to a different room (item 12, Table 1). In addition, shining light points or shadow spots on the floor affect the perception of surroundings (Cohen-Maitre, & Haerich, 2005). Also, the child may be trying to first explore the area, before entering or leaving the room. It is therefore important to take these aspects into account.

The questionnaire also includes some complex tasks (item 20, Table 1; and item 7, Table 2), caused for example by difficulty with depth perception, distance viewing or absence of visually guided reach (item 20, Table 1). With respect to the item 'The child has difficulty estimating the distance and speed of other road users' (item 19, Table 1), the difficulty is caused not only by the child moving but also by changes in the environment.

CP results in pathological reflexes, disorders of movement and posture development that cause activity limitations, falling into various severity levels of motor functioning (Rosenbaum *et al.,* 2007). Also, successful execution of fine motor skills, including reaching and grasping, is dependent on visual, motor, cognitive and other sensory processes such as tactile perception. For items 21 and 22 (Table 1) it is important to assess if the child uses visual guidance before reaching for a small object, which could be a result of visual support. When the child adapts the size of its hand to the size of the object after touching it, it could be a result of tactile support rather than obtained visual information. In the

item 'The child does not reach for and look at an object at the same time' (item 26, Table 1), the presence of CVI could affect serial processing in the brain, resulting in difficulty with multitasking (looking and reaching at the same time).

The results of the analyses of both CVI-MQ's show an excellent predictive value to predict the possible presence or absence of CVI in a child with CP. Early detection of developmental problems such as CVI is needed for a professional to facilitate an early start in appropriate intervention for these children and support for their parents. This has been proven to be beneficial and improves outcome (Ortibus *et al.,* 2011; Malkowicz, Myers, & Leisman, 2006; Visser, Ruiter, Meulen van der, Ruijssenaars, & Timmerman, 2015). Using the CVI-MQ's makes it possible to quickly achieve information on the risk of CVI in children with CP. Using these screening tools can also help paediatric physical therapists and occupational therapists to assess children with CP when additional certainty is desired about whether the current impairments of a child with CP are not only caused by motor or mental delay but perhaps also by the presence of CVI. Presuming the presence of CVI as a result of a positive score on the CVI-MQ's could be the first step towards an early diagnostic for a child with CP. In the absence of red flags, it also prevents unnecessary comprehensive testing of children and is cost- and time-efficient. Use of these CVI-MO's for children with CP is therefore relevant and warranted.

The cut-off score for GMFCS I-II-III is 7.5 points and higher if the sensitivity rate is set at 1.00 and the specificity rate at 0.96, or 15.5 points and higher if the sensitivity rate is 0.95 and the specificity rate 1.00. We therefore chose a score of 12 as cut-off value in order to meet a maximal sensitivity and specificity for children with GMFCS levels I-II-III. One child with CVI and GMFCS I (Table I) has a score below 10. We don't know the reason for this lower score but it could be caused by less suffering from presence of CVI or because of this child's high level of mobility.

Paediatric physical therapists and occupational therapists are familiar with motor screening and they can screen children with CP in approximately 10 minutes to detect children suspected of CVI and to refer for further assessment. To extend insights into the probability of the presence of CVI in a child with CP, we recommend using the CVI-MQ's as a part of comprehensive research with other screening tools for CVI. To obtain reliable information, it is also important that the same expert who is familiar with the child with CP administers the CVI-MQ.

We recommend taking into account that the content of both CVI-MQ's consists almost entirely of items at the level of motor functioning related to depth perception. CVI could result in for example a strong colour preference, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non-purposeful gaze, difficulty with distance viewing, absent or atypical visual reflexes, difficulty with visual novelty, or absence of visually guided reach (Dutton, & Jacobson, 2001; Stiers *et al.,* 2002).

5. Conclusion

The CVI-MQ's are a valuable addition for paediatric physical therapists and occupational therapists working with children with CP to detect the presence of CVI. Implementing CVI-MQ's as part of clinical reasoning is important in order to screen children with CP and identify red flags for CVI.

6. Clinical Messages

- CVI can result in a delayed motor development in children with CP.
- The red flags allow professionals to review the impact of CVI on the observed motor behavior in children with CP.
- The CVI-MQ's are a valuable addition for professionals working with children with CP to detect presence of CVI.

7. Conflict Of Interest

The authors declare that there is no conflict of interest.

8. Acknowledgements

We gratefully thank the children's parents and the therapists for their participation. Financial support for this study was provided by the Novum Foundation (O10323), a Dutch non-profit organisation that grants financial backing to projects and research that improve the quality of life of individuals with visual

impairments (www.stichtingnovum.org), and Royal Dutch Visio, centre of expertise for blind and visually impaired people (www.visio.org).

9. References

1. Boot, F.H., Pel, J.J.M., Evenhuis, H.M. & van der Steen, J. (2012) Factors related to impaired visual orienting behavior in children with intellectual disability. *Res Dev Dis,* **33:** 1670-1676.
2. Cohen-Maitre, S.A. & Haerich, P. (2005) Visual Attention to Movement and Color in Children with Cortical Visual Impairment. *Journal of Visual Impairment and Blindness,* **99(70),** 389-402.
3. Da Costa, M.F., Salmao, S.R., Berezovsky, A., De Haro, F.M. & Ventura, D.F. (2004) Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. *Behavioural Brain Res*, **149(2),** 145-150.
4. Dutton, G.N. & Jacobson, L.K. (2001) Cerebral visual impairment in children. *Semin Neonatol,* **6,** 477–485.
5. Dutton, G.N., Saaed, A., Fahad, B., Fraser, R., McDaid, G., McDade, J., (2004) Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction-a retrospective observational study. *Eye,* **18,** 27–34.
6. Dutton, G.N., Calvert, J., Cockburn, D., Ibrahim, H. & Macintyre-Beon, C. (2012) Visual disorders in children with cerebral palsy: The implications for rehabilitation programs and school work. *Eastern Journal of Medicine,* **17(4),**178-187.
7. Dutton, G.N. (2013) The spectrum of cerebral visual impairment as a sequel to premature birth: an overview. *Doc Ophthalmol.* **127(1),** 69-78.
8. Edmond, J.C. & Foroozan, R. (2006) Cortical visual impairment in children. *Current Opinion in Ophthalmol* **17,** 509-512.
9. Eng, J. (2005) Receiver Operating Characteristic Analysis: A primer. *Acad Radiol,* **12,** 909-916.
10. Fazzi, E., Bova, S.M., Uggetti, C., Signorini, S.G., Bianchi, P.E., Maraucci, I., Zoppello, M. & Lanzi, G. (2004) Visual-perceptual impairment in children with periventricular leukomalacia. *Brain Dev*, **26(8),** 506-512.
11. Fazzi, E., Signorini, S.G., LA Piana, R., Bertone, C., Misefari, W., Galli, J., Balottin, U. & Bianchi, P.E. (2012) Neuro-ophthalmological disorders in cerebral palsy: Ophthalmological, oculomotor, and visual aspects. *Dev Med Child Neurol,* **54,** 730–736.
12. Ghasia, F., Burnstroom, J., Gordon, M. & Tychsen, L. (2008) Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. *Invest Ophthalmol Vis Sci,* **49,** 572-580.
13. Good, W.V., Jan, J.E., DeSa, L., Barkovich, A.J., Groenveld, M. & Hoyt, S. (1994) Cortical visual impairment in children: A major review. *Surv Ophthalmol,* **38,** 351– 64.
14. Goodale, M.A. & Milner, A.D. (1992) Separate visual pathways for perception and action. *Trends in Neuroscience,* **15,** 20-25.
15. Gracht von der, H.A. (2012) Consensus measurement in Delphi studies: Review and implications for future quality assurance. *Technological Forecasting & Social Change: An International Journal,* **79(8),** 1525–1536.
16. Guzzetta, A., Mercuri, E. & Cioni, G. (2001) Visual disorders in children with brain lesions: Visual impairment associated with cerebral palsy. *Eur J Paediatr Rehabil Neurol Society,* **5,** 115-119.
17. Haley, S.M., Coster, W.J., Ludlow, L.H., Haltiwanger, J.T. & Andrellos, P.J. (1992) Pediatric Evaluation of Disability Inventory: Development, standardization, and administration manual. Boston, MA: New England Medical Centre Inc/PEDI Research Group.
18. International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation). (2008). Dutch WHO-FIC Collaborating Centre, first ed Bohn Stafleu van Loghum (www.bsl.nl) The Netherlands.
19. Liew, G., Michaelides, M. & Bunce, C. (2015) A comparison of the causes of blindness certifications in England and Wales in working age adults (16–64 years), 1999–2000 with 2009–2010. BMJ Open 2014; 2015.
20. Malkowicz, D.E., Myers, G. & Leisman, G. (2006) Rehabilitation of cortical visual impairment in children. *Int J Neurosci,* **116,** 1015-1033.
21. Ortibus, E., Verhoeven, J., Cock De, P., Sunaert, S., Casteels, I., Laenen, A., Schoolmeesters, B., Buyck, A. & Lagae, L. (2011) Screening for Cerebral Visual Impairment: validation of a CVI questionnaire. *Neuropaediatrics,* **42,** 138-147.
22. Powell, C. (2003) The Delphi technique: myths and realities. *J Adv Nurs,* **41(4),** 376–382.
23. Rosenbaum, P., Paneth, N., Leviton, A., Goldstein, M. & Bax, M. (2007) Definition and classification of cerebral palsy. *Dev Med Child Neurol,* **49(6),** 480.
24. Rosner, B. (2000) Fundamentals of Biostatistics. 5th Ed. Pacific Grove: Duxbury.
25. Russell, D.J. & Rosenbaum, P.L. (2002) Avery LM and Lane M. Gross Motor Function Measure (GMFM-66 and GMFM-88) User's Manual. London, United Kingdom: MacKeith Press.
26. Salavati, M., Rameckers, E.A.A., Steenbergen, B. & Schans van der, C.P. (2014) Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). *European Journal of Physical Therapy,* **16(3),** 159–167.
27. Salavati, M., Waninge, A., Rameckers, E.A.A., de Blécourt, A.C.E., Krijnen, W.P., Steenbergen, B. & Schans van der, C.P. (2015a) Reliability of modified paediatric evaluation of disability inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment. *Res Dev Dis,* **37,** 189-201.
28. Salavati, M., Krijnen, W.P., Rameckers, E.A.A., Looijestijn, P., Maathuis, C.G.B., Schans van der, C.P. & Steenbergen B. (2015b) Reliability of the modified Gross Motor Function Measure-88 (GMFM-88) for children with both Spastic Cerebral Palsy and Cerebral Visual Impairment: A preliminary study. *Res Dev Dis,* **45(46),** 32-48.
29. Schenk-Rootlieb, A.J.F., Van Nieuwenhuizen, O., Schiemanck, N., Van der Graaf, Y. & Willemse, J. (1993) Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. *Children Care Health Dev,* **19,** 411-423.
30. Steendam, M. (2008) Do you know what I see? Cerebral visual impairment in children: A manual for professionals. Royal Dutch Visio Huizen, www.visio.org.
31. Stiers, P., Vanderkelen, R., Vanneste, G., Coene, S., De Rammelsere, M. & Vandenbussche, E. (2002) Visual-perceptual impairment in a random sample of children with cerebral palsy. *Dev Med Child Neurol,* **44,** 370-382.
32. Visser, L., Ruiter, A.J., Meulen van der, F., Ruijssenaars, A.J.J.M. & Timmerman, E. (2015) Validity and suitability of the Bayley-III Low Motor/Vision version: A comparative study among young children with and without motor and/or visual impairments. *Pediatr Phys Ther,* **26(1),** 57-67.

7

Summary and general discussion

Summary and general discussion

The overarching aim of this dissertation was to contribute to the recognition and understanding of the presence of CVI in children with CP and, as a result, to adequate care and support for these children. Being aware of the extent that CVI affects a child with CP at the level of gross motor function, functional skills, and caregiver assistance is important in order to support these children and their parents. This thesis focused especially on: (1) whether and to what degree gross motor function, functional skills, and caregiver assistance in children with CP and CVI differ from those with CP and without CVI; (2) the development of an adapted version of the PEDI-NL and GMFM-88 for children with CP and CVI and determining their reliability; and (3) the development of two CVI Motor Questionnaires (CVI-MQs) for children with CP and determining their validity and usability.

General/principal findings

The initial purpose of this study was to determine whether and to what degree the level of gross motor function and functional skills in children with CP and CVI as well as caregiver assistance were different in comparison with the corresponding group of children experiencing CP without CVI. Therefore, the data aggregated from 23 children experiencing CP with CVI were compared with data from 23 children with CP without CVI matched for GMFCS, mental development, and age at testing. Scores for GMFM-88 and the PEDI-NL were employed to compare the level of gross motor function, functional skills, and caregiver assistance between both groups. The results indicated that self-care, mobility, and social functioning in the group of children with CP with CVI were significantly more affected than in the matched group with CP and without CVI and that this difference can be explained by the presence of CVI. Also, children with CP with CVI achieved obvious lower scores in all dimensions of gross motor function including laying and rolling, sitting, crawling and kneeling, standing, walking, running and jumping when compared with children with CP without CVI.

Children with CP and CVI have an inherent problem with proper identification and processing of visual information. Functioning of these children can be quantified with the PEDI-NL and the GMFM-88. However, the original PEDI-NL and GMFM-88 do not take visual functioning into account. Therefore, the results on these tests most likely do not reflect the actual motor capacity and underestimate the potential functioning of these children.[1,2,3,4,5,6] In order to measure motor skills and functional skills in children with CP and CVI, the verbal support/instruction, manual support, types of equipment, and environment of the content of PEDI-NL and the GMFM-88 were adapted. These adapted versions appeared to be valid and reliable. These adapted versions are additional supplements available for professionals. Prior to using these supplements, professionals must first be familiar with the original instructions of the PEDI-NL and the GMFM-88.

After determining the reliability of the adapted version of GMFM-88 for children with CP and CVI, research was focused on whether the adapted version of the GMFM-88 for children with CP and CVI resulted in higher scores and was a better reflection of their gross motor function per se without the influence of impaired visual abilities. Therefore, the scores of the original and adapted GMFM-88 were compared in the same group of children with CP and CVI. The comparison between the scores on the original and adapted GMFM-88 in children with CP and CVI yielded higher or similar scores in all dimensions of gross motor function including lying, rolling, sitting, crawling, kneeling, standing, walking, and running. The adapted GMFM-88 provided a better estimate of gross motor function per se in children with CP and CVI that was not adversely affected by their visual problems. On the basis of these findings, we recommend using the adapted GMFM-88 to measure gross motor functioning in children with CP and CVI. Paediatric physical therapists and occupational therapists using the adapted versions of the PEDI and GMFM-88 could determine a better estimation of functional ability and motor functioning of the child with CP and CVI. As a consequence, the approach and treatment of the child with CP and CVI will improve which may contribute to improved self-esteem and development of these children. Thus, e.g., during treatment, the therapist should give a child with CP and CVI more time to respond to a stimulus and should use extensive verbal instruction and manual support to help the child accomplish a task. Furthermore, a familiar environment can result in the

successful performance of daily skills, in contrast to an unknown or less familiar environment. To that aim, it is important to use the adapted PEDI-NL and GMFM-88 in the same environment and the same condition to evaluate a child's level of functioning.

Children with CP who may be labeled as "noncompliant", "oppositional", or "clumsy" could also be suffering from unidentified CVI. Early identification of CVI could lead to an emphasis on the correct determinants and proper focus of the comprehensive treatment which helps children in their development. A motor screening tool consisting of items related to the contribution of visual perception to perform a motor activity may be beneficial to paediatric physical therapists and occupational therapists. Thus far, the available CVI screening tools have focused on screening visual dysfunction, and no validated CVI screening tool is yet available to screen children with CP to identify the possible contribution of CVI on motor impairment.[7,8,9,10] Paediatric physical therapists and occupational therapists could benefit from having a CVI motor screening tool at their disposal in order to determine the extent to which CVI contributes to delays in motor disabilities in children with CP. Therefore, it is important to screen all children with CP, GMFCS levels I-V by using CVI-MQs as an additional tool to identify at-risk children with CP for the probability of having CVI. The final objectives of this thesis were to develop two Cerebral Visual Impairment Motor Questionnaires (CVI-MQs) for children with CP to describe their face validity and usability and to determine their sensitivity and specificity to detect a possible presence of CVI in children with CP. Therefore, the first initial versions of the two CVI-MQs were developed based on literature. The Delphi method was then used among two groups of experts, one familiar with CVI, in order to gain consensus about face validity and usability. The sensitivity and specificity of the CVI-MQs were subsequently assessed in 82 children with CP with and without CVI. Both questionnaires indicated very good face validity and good usability for practical use. The CVI-MQs had excellent sensitivity and specificity. The CVI-MQs are able to identify at-risk children with CP for the probability of having CVI. The development of two CVI-MQs for children with CP allow professionals working with children with CP to detect CVI in an early stage. This is important for these children as the visual impairment is often undetected in children with CP. A lack of recognition can be problematic for a child with CVI whose inaccurate visual guidance of movement, for example, may be misinterpreted as clumsiness.[11] Meanwhile, the child may be doing its best, yet is continuously criticized. The outcome can be disheartening for a child, leading to low self-esteem and a sense of being misunderstood.[11] With the CVI-MQs, the paediatric physical therapists and occupational therapists who are often the first professionals to investigate and treat children with CP at the level of motor functioning are able to identify warning signals (higher visual risk factor) for CVI when screening children with CP. Professionals can review the impact of CVI on the observed motor behaviour and ensure the identification of signs and symptoms of CVI in children with CP. Consequently, children suspected of CVI should be referred for full assessment and diagnosis by an ophthalmologist and paediatric neurologist in order to determine the presence of ocular visual impairment by the child with CP and to identify and characterize the disorder, its cause, and effective strategies for its treatment.

In conclusion, it is recommended to implement both the adapted PEDI-NL and GMFM-88 as the CVI-MQs in daily practice for children with CP and CVI.

CVI diagnosis

There are many variations in the manifestation of CVI as well as in the cause. Additionally, definitions of CVI vary between research groups and generally accepted diagnostic criteria are still inadequate.[12,13] Defining features of CVI have been described across different levels of human functioning including anatomical, functional, and behavioural levels. Currently, no agreement exists in terms of the use of cerebral imaging measures, tests to assess (cerebral) visual functioning, or the use of behavioural screening questionnaires to diagnose CVI.[13]

However, in the event of a CP diagnosis, especially for children with GMFCS IV-V, it is almost impossible to make a distinction between the different affected pathways because almost all brain areas are affected. Therefore, for those children with CP, it is not relevant to use the distinction between the ventral and dorsal stream. For children with CP and CVI, the motor impairment caused from CP will interfere with

impairment from CVI resulting in new phenomena which could be different from isolated CP or CVI. For example, reflexes in children with CP such as the Asymmetric Tonic Neck Reflex (ATNR) could have a greater effect on looking and reaching at the same time. The presence of CVI could result in the same behaviour as an ATNR reflex, specifically, after locating an object, looking away from it, and thereafter reaching to grasp that object. Therefore, the major challenge is to develop a set of diagnostic instruments for professionals working with children with CP and CVI. Also, there is a need to advance the diagnosis of CVI in children with CP based on quantitative parameters.

Due to the fact that there is no linear relationship between the impairment of body functions and structures, activities, and participation, it is important to use the ICF-CY (International Classification of Functioning, Disability and Health, Child & Youth version) as a tool to establish this diagnosis in children with CP, especially at the level of both activity and participation. Furthermore, professionals from all disciplines must be involved to establish a CVI diagnosis. The screening tools for detecting CVI in children with CP also should to be focused at the level of functional approach and both activity and participation components of the ICF-CY.[14] Despite the fact that it is important to be aware of this information, the focus of our studies were at the level of both activity and participation components of the ICF-CY.[14]

Consequences of CVI and assessment

The presence of CVI could result in difficulties of being able to visual locate caregivers and difficulty knowing whether they are present or absent thereby affecting the level of a child's motivation to acknowledge them. Furthermore, the child may become clumsy and become easily distressed in crowded environments.[11] When a child with CP exhibits a limitation of daily activities, slow motor processing, and performance speed, it may not only originate from a delay in motor and/or mental development but also from visual impairment. Furthermore, CVI can also cause behavioural problems in that child which need an approach from a perspective that is different than behavioural impairment. Using the CVI-MQs as an additional tool for screening children with CP to determine the presence of CVI is beneficial for helping professionals and caregivers to better understand a child. CVI is the contributory cause of motor limitation in the daily life of that child rather than only a motor dysfunction. Professionals should use the CVI-MQs when a child with CP is able to perform a particular motor task during therapy but is not able to perform the same task during a motor assessment. Also, when a child with CP has difficulty with motor skills related to depth perception such as jumping and reaching, the use of CVI-MQs could be considered. Other reasons to employ the CVI-MQs could be that a child does not react toward a sound in the majority of instances; the child has difficulties letting go of the reference point such as contact with the floor or the bed in its environment; and the child has a common behaviour of "freezing" in response to interesting stimuli.[15] Using these validated CVI-MQs affords an opportunity to quickly obtain information of the risk of the presence of CVI for a child with CP. Furthermore, it can help paediatric physical therapists and occupational therapists to assess children with CP by using these screening tools when professionals want to ensure that the current impairments of a child with CP are perhaps also caused from the presence of CVI. It also helps to achieve a realistic impression of the capabilities of a child with CP and CVI. Presuming of the presence of CVI as a result of a positive score on the CVI-MQs could be the initial step toward an early diagnosis for a child with CP. Also, in the event of an absence of warning signals, it prevents the unnecessary comprehensive testing of children. It is also cost and time efficient. Therefore, the use of these CVI-MOs for children with CP is relevant and warranted. In addition, centres of expertise for blind and visually impaired individuals and allied healthcare practices could use the CVI-MOs for children with CP as a part of wider investigation into CVI. Finally, it helps professionals working in rehabilitation centres to identify children with CP who are at risk for the probability of having CVI.

During data collection of children with CVI, it was not always recorded in medical files which aspects of CVI were present in a child with CVI. Therefore, the centres of expertise for blind and visually impaired people are advised to develop a standard procedure for screening and recording information of children with CP and CVI. This information must include the types of CVI that are present in a child with CP. Furthermore,

it is important to explain this information to those professionals working in the rehabilitation centres in order to implement this advice during the treatment of a child with CP and CVI. The adjusted treatment for children with CP and CVI facilitate self-determination for these children and help them increase independence in daily skills. Such an intervention requires a team approach with CVI experts who work together with the experts from rehabilitation centres. Moreover, it is important to support and educate the professionals working at rehabilitation centres on how to implement knowledge on CVI in their approach for children with CP. Furthermore, factors to consider that affect visual presentation during a motor activity vary with the need of each child with CP and CVI and include the size, contrast, color, and arrangement of materials.[11] In addition to these, movement has also been determined to attract and maintain attention for children with CP and CVI.[16] It is obvious that the contribution of the parents and caregivers is the most important component of comprehensive intervention for these children.

The impact of CVI on the daily life of a child with CP could result in a delay of motor development and a greater level of dependency. It is not only important to be aware of ocular visual impairment, but also recommended to investigate the possible presence or absence of CVI in children with CP. Therefore, to meet the needs of these children, it is very important that both researchers and education centres for professionals integrate the knowledge of CVI with CP during the research studies on children with CP. It prevents a presence of bias in future studies on CP. For education of health care professionals, it is also recommended to develop special modules for children with CP and CVI which include the knowledge on CVI in children with CP and the practical implication for those children at the level of gross motor function, functional, skills and caregivers.

Future challenges

One of challenges is that the adapted PEDI-NL and GMFM-88 be implemented in the educational program for paediatric physical therapists and occupational therapists which could eventually result in the use of these assessment instruments in clinical practice. Hereafter, a child with CP and CVI and his or her therapist can benefit from these adapted versions. The child is able to show its capabilities at the level of functioning, and the therapist can estimate the child with CP and CVI at the proper level.

The other challenge is to Knowledge transfer and implementation of the CVI-MQs in the rehabilitation centres. The professionals working with children with CP have an advantage when using the CVI-MQs because all of the items in both questionnaires are related to the motor functioning of a child who is familiar to the professional. In the event of a positive outcome score, this child could be referred for a full assessment and diagnosis by an ophthalmologist and a pediatrician. Furthermore, in the future, it is very important to determine the psychometric proportion of CVI-MQs in children with CP and CVI.

The last, and also the greatest, challenge is to adapt or develop an intervention program for children with CP and CVI. CVI influences a child's ability to learn and perform tasks in everyday life and should, therefore, be taken into account in therapy and intervention.[17] In addition, the development of a visually impaired child can be delayed especially with regard to self-initiated mobility, posture, and locomotion.[17] It is established that CVI has an impact on all aspects of a child's development, and children with both CP and CVI develop more slowly in the areas of self-care, mobility, and social function than children with CP and without CVI.[2,18,19,20,21,22,23] Due to the fact that children with CP and CVI are more limited at the level of gross motor function, functional skills, and caregiver assistance, it is important that those children meet their needs during treatment by therapists giving appropriated verbal instruction and manual support.[2,18,19,20,21,22,23]

Children with CP and CVI also have an extended processing time to receive and interpret information from their environment.[11] By providing these children with extra time to understand the content of a question, they will be able to demonstrate real ability to perform a particular task. For children with CP and CVI, it is important that the environment is predictable, the decoration of a room does not frequently change, and they are involved in the layout of their room.[11,16,24,25,26,27,28] This may contribute to independent functioning in their daily life.

Considering the fact that a high number of children with CP also have CVI, it is very important for paediatric

physical therapists and occupational therapists to have an effective and evidence-based intervention program that is adapted for children with both CP and CVI at their disposal. It is a fact that children with CP and CVI are different from those with CP and without CVI, therefore, it is important to develop a specific intervention program for children experiencing CP with CVI in order to improve their gross motor function, functional skills, and caregiver assistance. There is evidence of the effect of different intervention programs such as functional therapy, strength training, and condition improvement to improve mobility and self-care in children with CP.[29,30] Novak et al.[30], Franki et al.[31], Gelkop et al.[32] and Sakzewski et al.[33] concluded that intervention programs that include aspects such as goal-directed training, context-focused therapy, and home-programs are the best evidence intervention programs to improve gross motor function, functional skills, and self-care in children with CP. However, those intervention programs did not take into the account the presence of CVI and its effect on motor functioning and self-care of children with CP and, therefore, they are less suitable for children with both CP and CVI.

An intervention program must have, as a basis, the following criteria: goal-oriented, focused on activities and/or participation, task-oriented, active contributing of the child and parents in learning, discovering and finding solutions, focused on functionality instead of normality and context-specific.[29] On the other hand, it is important to integrate the aspects from CVI into those criteria to develop such an intervention program. Those aspects could be adapting the environment by adding color to the using aids or giving verbal and manual support (e.g., duration and phase of required manual support that is given) during the specific phase of the execution of a task. Furthermore, by teaching a particular task to a child with CP and CVI, it important to begin with comprehensive hands-on training of the entire task and, when the child is familiar with the task, proceed into hands-off training of the same task.

Early intervention in a visually impaired child is stressed, and treatment of sensory input impairments should begin as early as possible in a positive emotional setting that enhances the child's motivation and relationship with caregivers.[34,35] Such an intervention program needs to help a child to increase its awareness of visual stimuli, improve visual attention and teach basic visual skills as well implement compensatory strategies that are all integrated into the activities of daily life.[9] It is not only important to develop or adapt an intervention program for children with CP and CVI but also to ensure the implementation of it. To achieve such goals established for those children, there are three intervention aspects: 1) education of all caregivers and usual school staff about the child's visual functioning; 2) application of compensatory strategies; and 3) regular sessions of direct teaching by the caregivers and school staff.[9] The centres of expertise for blind and visually impaired people have significant responsibility not only by testing a child at the level of visual functioning and providing instruction and advice but also monitoring this child and being proactive. Implementing advice in the daily life of a child with CVI is a continuous process of investing in the child and its environment, and these need to be warranted by the centres of expertise for blind and visually impaired people.

In general, there are three recommendations which can be made to guide an intervention that will facilitate the development of children with CVI. First, it is critical that early identification of CVI be a focus of pediatricians and other early interventionists. Second, an intervention should focus on the use of integrated sensory information. Lastly, the intervention should be family focused.[36]

In the future, it is recommended to add a section for children with visual impairment into the "Guidelines for the diagnosis and treatment of children with Spastic Cerebral Palsy".[29] This section consists of information on CVI and OVI related to CP at the level of ICF that can support professionals' efforts to use the knowledge of visual impairments on children with CP.

Study limitations

In the studies in this thesis, the diagnosis of CVI was determined based on the results of ophthalmological and psychological/neuropsychological assessments and on the assessment data reported by a developmental coach specialized in working with children with visual impairments. However, we did not select specific subtypes of CVI because the additional information in the medical files of which type of CVI the child was experiencing was often incomplete. As a consequence, we were not able to determine the differences between the types of CVI. This may be a limitation because it is

not clear whether the different types of CVI contributed to those studies and affected the results on the adapted PEDI-NL, GMFM-88, and CVI-MQs. However, there are several issues such as environmental and personal factors which may contribute to the execution of a motor activity and the content of the adapted PEDI-NL, GMFM-88 and CVI-MQs consist of various items which could represent all types of CVI within them. Therefore, these measurements are appropriate to use for children with various types of CVI.

In addition, brain damage resulting in CP affects several areas such as motor and visual functioning which makes it difficult to distinguish damage from various areas. In the study of the adapted PEDI-NL, the adapted GMFM-88, and CVI-MQs, we included participants with different types of CP in various degrees of severity who might have different profiles of motor functioning. Most of the participants (96%) were children with spastic CP. The studies on original PEDI-NL and GMFM-88 have demonstrated that these instruments were developed for all types of CP.[6,37,38,39,40] Despite the fact that most of participants were children with spastic CP, and in accordance with other studies[6,15,16,17,18], the results of these studies could also be used for children with other types of CP such as dyskinetic CP and ataxic CP.

Concluding remarks

Professionals working in rehabilitation centres must be aware of the presence of CVI in children with CP. Due to the fact that CVI results in delays of motor development and a greater level of dependency, it is important to screen children with CP and, if needed, to refer to centres of expertise for blind and visually impaired people. In the event of the presence of CVI in a child with CP, the professionals and caregivers must adjust their approach to help the child achieve an increased level of independency. Also, the knowledge about the presence of CVI in the child with CP contributes to better estimating of functioning level.

References

1 Haley S M, Coster WJ, Ludlow LH, Haltiwanger JT & Andrellos PJ. Pediatric Evaluation of Disability Inventory: Development, standardization, and administration manual. Boston, MA: New England Medical Centre Inc. and PEDI Research Group; 1992.

2 Salavati M, Rameckers EAA, Steenbergen B & Schans van der CP. Gross motor function, functional skills and caregiver assistance in children with spastic cerebral palsy (CP) with and without cerebral visual impairment (CVI). *European Journal of Physical Therapy* 2014; 16 (3): 159–167.

3 Salavati M, Waninge A, Rameckers EAA, de Blécourt ACE, Krijnen WP, Steenbergen B & Schans van der, CP Reliability of modified paediatric evaluation of disability inventory, Dutch version (PEDI-NL) for children with cerebral palsy and cerebral visual impairment. *Res Dev Dis* 2015a; 37: 189-201.

4 Salavati M, Krijnen WP, Rameckers EAA, Looijestijn P, Maathuis CGB, Schans van der, CP & Steenbergen B Reliability of the modified Gross Motor Function Measure-88 (GMFM-88) for children with both Spastic Cerebral Palsy and Cerebral Visual Impairment: A preliminary study. *Res Dev Dis* 2015b; 45(46): 32-48.

5 Visser L, Ruiter AJ, Meulen van der F, Ruijssenaars AJJM & Timmerman E. Validity and suitability of the Bayley-III Low Motor/Vision version: A comparative study among young children with and without motor and/or visual impairments. *Pediatr Phys Ther* 2014; 26 (1): 57-67.

6 Wassenberg-Severijnen JE,Custers JW, Hox JJ,Vermeer A & Helders PJ. Reliability of the Dutch Pediatric Evaluation of Disability Inventory (PEDI). *Clinical Rehabilitation* 2003; 17: 457–462.

7 Ortibus E, Verhoeven J, Cock De P, Sunaert S, Casteels I, Laenen A, Schoolmeesters B, Buyck A & Lagae L. Screening for Cerebral Visual Impairment: validation of a CVI questionnaire. *Neuropaediatrics* 2011; 42: 138-147.

8 Dutton GN & Jacobson LK. Cerebral visual impairment in children. *Semin Neonatol* 2001; 6: 477–485.

9 Steendam M . Do you know what I see? Cerebral visual impairment in children: A manual for professionals. Haren: Royal Visio Huizen ; 2007.

10 Dutton GN, Calvert J, Cockburn D, Ibrahim H & Macintyre-Beon C. Visual disorders in children with cerebral palsy: the implications for rehabilitation programs and school work. *Eastern Journal of Medicine* 2012; 17(4): 178-187.

11 Lueck AH & Dutton GN. *Vision and the Brain: Understanding Cerebral Visual Impairment in Children* 2015; ISBN: 9780891286394.

12 Boot FH, Pel JJM, van der Steen J & Evenhuis HM. Cerebral Visual Impairment: which perceptive visual dysfunctions can be expected in children with brain damage? A systematic review. *Res Dev Dis* 2010; 31: 1149-1159.

13 Geldof CJA, Wassenaer-Leemhuis van AG, Dik M, Kok JH & Oosterlaan J. A functional approach to cerebral visual impairments in very preterm/very-low-birth-weight children. *Pediatric Research* 2015; 78: 190–197.

14 International Classification of Functioning, Disability and Health, Child & Youth version (Dutch translation). Dutch WHO-FIC Collaborating Centre. 1st ed. Houten: Bohn S*tafleu van Loghum (w*ww.bsl.nl) 2008.

15 Tadić V, Pring L & Dale N. Are language and social communication intact in children with congenital visual impairment at school age? *J Child Psychol Psychiatry* 2010; 51(6): 696-705.

16 Cohen-Maitre S & Haerich P. Visual Attention to Movement and Color in Children with Cortical Visual Impairment. *J Vis Impair Blind* 2005; 99(7): 389-402.

17 Fazzi E, Signorini SG, LA Piana R, Bertone C, Misefari W, Galli J, Balottin U & Bianchi PE. Neuro-ophthalmological disorders in cerebral palsy: Ophthalmological, oculomotor, and visual aspects. *Dev Med Child Neurol* 2012; 54: 730–6.

18 Da Costa MF, Salmao SR, Berezovsky A, De Haro FM and Ventura DF. Relationship between vision and motor impairment in children with spastic cerebral palsy: New evidence from electrophysiology. *Behavioural Brain Res* 2004; 149: (2) 145-150.

19 Dutton G & Jacobson L. Cerebral visual impairment in children. *Semin Neonatol* 2001; 6: 477-85.

20 Dutton GN. The spectrum of cerebral visual impairment as a sequel to premature birth: an overview. *Doc Ophthalmol* 2013; 127: (1) 69-78.

21 Ghasia F, Burnstroom J, Gordon M & Tychsen L. Frequency and severity of visual sensory and motor deficits in children with cerebral palsy: Gross Motor Function Classification Scale. *Invest Ophthalmol Vis Sci* 2008; 49: 572-580.

22 Good W, Jan F, Burden S, Skoczenski A & Candy R. Recent advances in cortical visual impairment. *Dev Med Child Neurol* 2001; 43: 56-60.

23 Schenk-Rootlieb AJF, Van Nieuwenhuizen O, Schiemanck N, Van der Graaf Y & Willemse J. Impact of cerebral visual impairment on the everyday life of cerebral-palsied children. *Children Care Health Dev* 1993; 19: 411-423.

24 Pel J, Does van der L, Boot F, et al. Effects of visual processing and congenital nystagmus on visually guided ocular motor behaviour. *Dev Med Child Neurol* 2011; 53(4): 344-349.

25 Wagner M, Haibach P & Lieberman L. Gross motor skill performance in children with and without visual impairments—Research to practice. *Res Dev Disabil* 2013; 34(10): 3246-3252.

26 Aki E, Atasavun S, Turan A & Kayihan H. Training motor skills of children with low vision. *Percept Mot Skills* 2007; 104(3 Pt 2): 1328-1336.

27 Fazzi E, Lanners J, Ferrari-Ginevra O, et al. Gross motor development and reach on sound as critical tools for the development of the blind child. *Brain Dev* 2002; 24(5): 269-275.

28 O'Connell M, Lieberman L & Petersen S. The use of tactile modelling and physical guidance as instructional strategies in physical activity for children who are blind. *J Vis Impair Blind* 2006; 100(8): 471-477.

29 Dutch Institute of Rehabilitation Paediatricians. Guidelines for the diagnosis and treatment of children with Spastic Cerebral Palsy. Postbus 9696, 3506 GR Utrecht; 2015.

30 Novak I, McIntyre S, Morgan C, et al. A systematic review of interventions for children with cerebral palsy: state of the evidence. *Dev Med Child Neurol* 2013; 55 (10): 885-910.

31 Franki I, Van den Broeck C, De Cat J, et al. A randomized, single-blind cross-over design evaluating the effectiveness of an individually defined, targeted physical therapy approach in treatment of children with cerebral palsy. *Clin Rehabil* 2014; 28 (10): 1039-1052.

32 Gelkop N, Burshtein D, Lahav A, et al. Efficacy of Constraint-Induced Movement Therapy and Bimanual Training in Children with Hemiplegic Cerebral Palsy in an Educational Setting. *Phys Occup Ther Pediatr* 2015; 35 (1): 24-39.

33 Sakzewski L, Miller L, Ziviani J, et al. Randomized comparison trial of density and context of upper limb intensive group versus individualized occupational therapy for children with unilateral cerebral palsy. *Dev Med child Neurol*. 2015; 57 (6): 539-47.

34 Fazzi E, Signorini SG, Bova SM, Ondei P & Bianchi PE. Early intervention in visually impaired children. *International Congress Series* 2005; Volume 1282, 117–121.

35 Houwen S, Hartman E & Visscher C. Physical activity and motor skills in children with and without visual impairments. *Med Sci Sports Exerc* 2009; 41(1): 103-109.

36 Fazzi E, Molinaro A & Hartmann E. The potential impact of visual impairment and CVI on child development. *Vision and the brain* 2012; ISBN: VATBCh04.

37 Wassenberg-Severijnen JE & Custers JWH. Pediatric evaluation of Disability Inventory-NL. Amsterdam, The Netherlands: Harcourt Assessment BV 2005.

38 Custers JWH,Wassenberg-Severijnen JE, Net Jvd, Vermeer A, Hart HT & Helders PJM. Dutch adaptation and content validity of the 'Pediatric Evaluation of Disability Inventory (PEDI)'. *Disability and Rehabilitation* 2002; 24: 250–258.

39 Russell DJ & Rosenbaum PL. Avery LM and Lane M. Gross Motor Function Measure (GMFM-66 and GMFM-88) User's Manual. London, United Kingdom: MacKeith Press 2002.

40 Ketelaar M, Van Petegem-van Beek E, Veenhof C, Visser J & Vermeer A. Gross motor function measure. University of Utrecht. *Child Physical Therapy* 2003; 39: 5–7.

S |

Summary in Dutch

Nederlandse samenvatting

De inhoud van dit proefschrift heeft betrekking op kinderen met aangeboren hersenbeschadiging, Cerebrale Parese (CP), bij wie ook sprake is van onder andere visuele waarnemingsproblemen. Bij kinderen met CP kunnen problemen op het gebied van visuele waarneming in het oog en/of in de hersenen gelokaliseerd zijn. Oculaire Visuele Inperking (OVI) heeft betrekking op die vormen van slechtziendheid welke samenhangen met oogheelkundige aandoeningen. Cerebrale Visuele Inperking (CVI) kan omschreven worden als een visuele stoornis als gevolg van al dan niet aantoonbare schade aan één of meerdere hersengebieden na het chiasma opticum of aan slechtziendheid waar geen oculaire verklaring voor bestaat. CVI kan veroorzaakt worden door het hypoxische-ischemisch letsel. Daarnaast kunnen prematuriteit, hydrocefalus, epilepsie en beschadigingen aan het centraal zenuwstel, CVI tot gevolg hebben. De klinische symptomen van CVI kunnen zich manifesteren in onder andere visueel-sensorische symptomen, oculomotorische symptomen en hogere visuele symptomen. Een ander opvallend kenmerk is verminderde gezichtsscherpte zonder oogheelkundige verklaring. CVI kan samengaan met oogheelkundige afwijkingen zoals strabismus, optische atrofie en nystagmus. Daarnaast doen visuele problemen zich met name voor op het gebied van de hogere cognitieve processen als herkenning, oriëntatie, diepte-perceptie, gelijktijdige perceptie van beweging en visuele waarneming. Kinderen met CVI kunnen bijvoorbeeld moeite hebben met het vinden van de looproutes en de overgang van licht naar donker. Als gevolg van CVI kunnen functioneringsproblemen in het dagelijks leven ontstaan maar ook problemen in het gedrag en motorische ontwikkeling van kinderen. Deze problemen kunnen op verschillende manieren tot uiting komen omdat CVI samen kan voorkomen met andere problemen, zoals gedragsproblemen, epilepsie of problemen op het gebied van aandacht en concentratie.

De inhoud van dit proefschrift wordt in **hoofdstuk 1** toegelicht en in hoofdlijnen besproken.

In **hoofdstuk 2** worden de verschillen in het grof-motorisch functioneren, functionele vaardigheden en verzorgersassistentie tussen twee vergelijkbare groep kinderen onderzocht. De ene groep bestaat uit kinderen met CP en CVI en de andere groep bestaat uit kinderen met CP zonder CVI. Uit het onderzoek blijkt dat kinderen met CP en CVI grotere achterstand op het gebied van het grof-motorische functioneren ondervinden. Daarnaast zijn de kinderen met CP en CVI meer afhankelijk van hun ouders en begeleiders, zich uitend in hogere mate van verzorgersassistentie op het gebied van zelfverzorging, ambulantie en sociaal functioneren. Dit in vergelijking met kinderen met CP zonder CVI.

In **hoofdstuk 3** worden de ontwikkeling en betrouwbaarheid van een aangepast versie van de Paediatric Evaluation of Disability Inventory, Nederlandse versie (PEDI-NL), voor kinderen met CP en CVI beschreven. De PEDI-NL is een instrument waarmee op basis van een gestructureerd interview kan worden nagegaan of er ontwikkelingsachterstand of functionele beperkingen aanwezig is bij kinderen van zes maanden tot 7,5 jaar. Enerzijds worden de vaardigheden die het kind bezit geëvalueerd. Anderzijds wordt de hoeveelheid hulp die het kind van de ouders en verzorger(s) krijgt genoteerd. Daarnaast wordt ook de uitgebreidheid van aanpassingen die kinderen eventueel gebruiken genoteerd. Op basis van het gestructureerd interview kunnen normscores worden berekend, die vergeleken kunnen worden met normscores van gezonde kinderen van dezelfde leeftijd. Daarnaast kunnen schaalscores worden berekend, die aangeven welk percentage van een bepaalde dagelijkse activiteit het kind beheerst. PEDI-NL bestaat uit: een functionele vaardigheidsschaal, verzorgerassistentieschaal en aanpassingsschaal. Iedere schaal bestaat uit drie domeinen: zelfverzorging, ambulantie en sociaal functioneren. De PEDI-NL is niet geschikt voor kinderen met een visueel waarnemingsprobleem, omdat in de vraagstellingen geen rekening is gehouden met de aanwezigheid van CVI.

Op basis van Delphi onderzoek is eerst de PEDI-NL aangepast voor kinderen met CVI. De aangepaste PEDI-NL in de vorm van het CVI-supplement kan gebruikt worden voor kinderen met CVI. Het CVI-supplement is een toevoeging op de instructie van de oorspronkelijke PEDI-NL. De oorspronkelijke vraagstelling en de score van de PEDI-NL zijn niet

gewijzigd. Het CVI-supplement heeft betrekking op de manuele ondersteuning en verbale instructie aan het kind in relatie tot de wijze waarop een taak wordt uitgevoerd. Zo geven ouders voorafgaand aan een activiteit, waar toegestaan, verbale instructies gericht op de uitvoering van een taak.

Vervolgens is de test-hertest en interbeoordelaar betrouwbaarheid van aangepaste versie PEDI-NL voor kinderen met CVI bij een groep kinderen met CP en CVI onderzocht. Uit het onderzoek naar de psychometrische eigenschappen blijkt dat de aangepaste PEDI-NL voor kinderen met CP en CVI betrouwbaar is.

In **hoofdstuk 4** worden de resultaten van het onderzoek naar de betrouwbaarheid van de aangepaste Gross Motor Function Measure-88 (GMFM-88) voor kinderen met CP en CVI beschreven. De GMFM-88 is een evaluatief instrument waarmee de veranderingen in het grof-motorisch functioneren van kinderen met CP vastgelegd kunnen worden. De GMFM-88 is een criterium gerelateerde test en is ontworpen om te bepalen hoeveel procent van een motorisch item het kind kan uitvoeren. De GMFM telt 88 items die zijn onderverdeeld in vijf dimensies van de grove motoriek: A: liggen en omrollen, B: zitten, C: kruipen en knielen, D: staan, E: lopen, rennen en springen. Er wordt van uitgegaan dat een kind van 5 jaar, met normale motorische vaardigheden, alle 88 items kan uitvoeren. De oorspronkelijke GMFM-88 houdt geen rekening met de aanwezigheid van visuele beperking bij een kind met CP.

Op basis van Delphi onderzoek is de oorspronkelijke GMFM-88 aangepast voor kinderen met CVI. De aangepaste GMFM-88 in de vorm van een CVI-supplement kan gebruikt worden voor kinderen met CVI. Tevens is de test-hertest en interbeoordelaar betrouwbaarheid van kinderen met CP en CVI onderzocht. Op basis van de uitkomsten van dit onderzoek blijkt dat de aangepaste GMFM-88 voor kinderen met CVI een valide en betrouwbaar instrument is voor het gebruik door kinderfysiotherapeuten.

In **hoofdstuk 5** worden de resultaten beschreven van het onderzoek naar het grof-motorisch functioneren van kinderen met CP en CVI, waarbij de uitkomsten van de oorspronkelijke GMFM-88 en de aangepaste GMFM-88 vergeleken zijn voor kinderen met CP en CVI. De resultaten van dit onderzoek laten zien dat de aangepaste GMFM-88 voor kinderen met CP en CVI tot een hogere score leidt, omdat deze aangepaste versie rekening houdt met de aanwezigheid van visuele waarnemingsproblemen bij kinderen met CP en CVI. Het gebruik van een aangepaste GMFM-88 voor kinderen met CP en CVI resulteert in betere inschatting van het grof-motorisch functioneren van kinderen met CP en CVI. Daarom wordt aanbevolen gebruik te maken van het CVI-supplement voor de GMFM-88 wanneer er bij een kind met CP ook sprake is van CVI.

Hoofdstuk 6 richt zich op de ontwikkeling, validiteit, gebruiksvriendelijkheid, sensitiviteit en specificiteit van twee motorische CVI screeningsinstrumenten (CVI-MQ's) voor kinderen met CP. Er is een screeningsinstrument voor de groep kinderen met Gross Motor Function Classification System (GMFCS) I-II-III en een screeningsinstrument voor de groep kinderen met GMFCS IV-V ontwikkeld. De inhoud van CVI-MQ voor kinderen met GMFCS niveau I-II-III bevat motorische items die gerelateerd zijn aan hogere motorische vaardigheden zoals lopen, traplopen en springen. De CVI-MQ voor kinderen met GMFCS niveau IV-V items bevat inhoud die gerelateerd is aan vaardigheden zoals het omrollen en reiken en grijpen.

De CVI-MQ's zijn ontwikkeld op basis van literatuur en door inbreng van experts. Na de ontwikkeling van de CVI-MQ's is de indruksvaliditeit en gebruiksvriendelijkheid bij een groep experts onderzocht. Er bleek een hoge mate van consensus te zijn onder de experts over de validiteit, en de gebruiksvriendelijk. Hierna is de sensitiviteit en specificiteit van beide screeningsinstrumenten bij twee groepen kinderen onderzocht, één groep met de diagnose CP en CVI en de andere groep met alleen de diagnose CP. De sensitiviteit en specificiteit om de eventuele aanwezigheid van CVI in kinderen met CP op te sporen blijkt zeer goed te zijn. Gezien het feit dat bij kinderen met CP het risico op aanwezigheid van CVI groot is, is het van belang dat kinderfysiotherapeuten, kinderergotherapeuten en revalidatieartsen over een motorisch screeningsinstrument beschikken waarmee ze het risico op aanwezigheid van CVI bij kinderen

met CP tijdig kunnen signaleren. Hierdoor kan een kind met CP met het vermoeden op CVI vroegtijdig voor verder onderzoek doorverwezen worden naar een kinderneuroloog en expertise centra voor mensen met een visuele beperking.

In **hoofdstuk 7** worden de resultaten van het proefschrift samengevat, bediscussieerd en tevens worden de implicaties voor de praktijk en het toekomstig onderzoek beschreven.

De belangrijkste conclusies van dit proefschrift zijn:

- CVI kan resulteren in een achterstand in motorische ontwikkeling van kinderen met CP.
- Kinderen met de diagnose CP en CVI behalen lager score op het gebied van de grof-motorische vaardigheden, zelfverzorging en sociaal functioneren. Dit in vergelijking met kinderen met de diagnose CP.
- Het gebruik van aangepaste PEDI-NL en GMFM-88 voor kinderen met CP en CVI kan kinderfysiotherapeuten en ergotherapeuten helpen om een adequate inschatting te maken van functionele vaardigheden en motorisch functioneren van kinderen met CP en CVI. Als gevolg hiervan kan de behandeling beter afgestemd worden op de mogelijkheden van dit kind welke de ontwikkeling van het kind positief stimuleert.
- CVI-MQ's kunnen gebruikt worden als screenings-instrumenten door professionals werkzaam met kinderen met CP. Hiermee kan worden nagegaan of bij kinderen met CP ook sprake is van het vermoeden op aanwezigheid van CVI.

A

Acknowledgments

Dankwoord

Een inspiratie ontstaat meestal individueel maar voor de uitvoering daarvan heb je altijd een groep voor nodig! De inhoud van dit proefschrift is dan ook het product van de ondersteuning van een grote groep mensen en hard werken.

Ik wil dan ook graag een aantal van deze grote groep mensen bedanken.

Om te beginnen gaat mijn dank uit naar de kinderen, ouders, verzorgers en collega's die geholpen hebben met tot stand komen van dit onderzoek. Daarnaast wil ik alle instellingen, revalidatiecentra en eerste lijn particulier praktijken bedanken voor het verlenen van hun medewerking aan deze onderzoeken. Voor het testen van kinderen ben ik mijn dank schuldig aan alle collega kinderfysiotherapeuten en ergotherapeuten. Dit waardeer ik zeer.

Mijn dankbaarheid is groot voor het hele onderzoeksteam. Ik wens iedereen zo'n bijzonder onderzoeksteam.

Professor dr. Van der Schans, beste Cees, je voortreffelijke begeleiding, rust, relativeringsvermogen en het met jou van gedachten te wisselen over de opzet en uitvoering van het onderzoek heb ik als zeer waardevol en heel erg leerzaam ervaren. Het was heel prettig om in deze vorm, begeleiding en ondersteuning te krijgen.

Mijn grote dank voor alles wat je voor mij gedaan hebt.

Professor dr. Steenbergen, beste Bert, ik ben je zeer dankbaar voor het leren kritisch wetenschappelijk bezig zijn en blijvend werken aan het verbeteren van mijn argumentaties. Je scherpte heeft mij bijzonder geholpen om mijn onderzoek steeds op een hoger niveau te tillen. De momenten die ik met je van gedachten wisselde heeft mij heel veel nieuwe inzichten bijgebracht, waarvoor mijn grote dank.

Dr. Rameckers, beste Eugene, wij kennen elkaar al ruim voor de start van mijn promotietraject en je ondersteuning aan mijn promotie beschouw ik ook zeer belangrijk in mijn onderzoekstraject. Je bijzonder prettige persoonlijkheid in combinatie met alle andere positieve ondersteuning waaronder je inzicht en scherpte hebben mijn ontzettend veel geholpen. Mijn grote dank hiervoor.

Dr. Waninge, beste Aly, ik ben heel blij om naast je collega te zijn, ook je prettige en waardevolle ondersteuning te mogen krijgen voor tot stand komen van mijn proefschrift. Je rust in combinatie met kritisch opbouwende vragen hebben mij enorm geholpen met het verbeteren van mijn onderzoeken. Ik wil je heel erg bedanken voor je prettige ondersteuning.

Mijn grote dank gaat ook uit naar dr. De Blécourt (postuum), dr. Krijnen, dr. Looijestijn, dr. Maathuis en professor dr. Van der Steen voor jullie bijdrage aan het schrijven van artikelen in mijn proefschrift. Jullie hebben mij vanuit de verschillende invalshoeken geholpen dit manuscript te verbeteren en hiervoor ben ik jullie allen bijzonder dankbaar.

Mevrouw Van der Driessche, beste Marie Jose, je geloof in mijn mogelijkheden en het belang van dit onderzoek voor de doelgroep personen met een visuele en meervoudige beperking heeft geresulteerd tot je inzet om me te ondersteunen bij het indienen van mijn onderzoeksaanvraag. Ook je belangstelling tijdens het verloop van mijn onderzoek beschouw ik als zeer belangrijk en waardevol. Heel erg bedankt.

Dr. Van der Steen, beste Sanny, je luisterende oor, je tips en je ondersteuning in alle fasen van mijn onderzoek beschouw ik als onmisbaar en heel waardevol. Tijdens de contactmomenten volgde je met belangstelling mijn onderzoekstraject en ondersteunde me in allerlei zaken die mijn onderzoek faciliteerden. Hierdoor kon ik mijn focus op mijn onderzoek blijven richten. Heel erg bedankt hiervoor. Ook afdeling KEI van Koninklijke Visio wil ik heel erg bedanken voor de ondersteuning die ze me verleend hebben.

Mijn bijzondere dank aan mevrouw Judith van der Boom, mevrouw Ieneke van der Jagt en mevrouw Ineke van der Hoek voor jullie ondersteuning vanuit de Hanzehogeschool in de afgelopen jaren. Het is een geruststelling om als promovendus voor allerlei vragen en adviezen op jullie terug te kunnen vallen.

Tijdens mijn heel onderzoekstraject heb ik veelvuldig beroep gedaan op mijn collega's Jacqueline Maschewski, Jikke Littel en Jasmijn van der Tuin. Jullie hebben mij enorm geholpen met veel werk uit mijn handen nemen

en kinderen te testen voor mijn onderzoek. Ik dank jullie voor deze ondersteuning.

Daarnaast wil ik Kim Tjarks en Ewoud Meijer bedanken voor het helpen testen van kinderen en Jasmijn van der Tuin, Jasmijn Tepper en Lotte Jongsma bedanken voor hun ondersteuning in het laatste deel van mijn onderzoek.

Voor het tot stand komen van een groot deel van mijn onderzoek is de ondersteuning van een grote groep experts binnen en buiten Koninklijke Visio onontbeerlijk geweest. Ik wil jullie allen bedanken voor jullie beschikbaarheid, hulp en positieve bijdrage aan mijn onderzoek. Ik waardeer het zeer hoe jullie steeds voor mij klaar stonden. Heel erg bedankt.

Zonder de ondersteuning van stichting Novum en Koninklijke Visio was dit onderzoek niet tot stand gekomen. Daarom mijn dankbaarheid voor jullie ondersteuning. Hierbij wil ik ook heel graag dhr. M. de Bruine, Voorzitter Raad van Bestuur van Koninklijke Visio voor zijn ondersteuning bedanken.

Naar mijn mening heeft iedere onderzoeker een stevig fundament en drijfveer vanuit de thuissituatie nodig om te kunnen onderzoeken.

Eén van mijn inspiratiebronnen ben jij Anet. Je hebt ook nu veel voor mij betekend. Je hebt mij gestimuleerd om het onderzoek op te zetten en uit te voeren. Dank je wel.

Nastaran en Sahar, het is bijzonder om tegelijk met jullie als mijn dochters met een onderzoekstraject bezig te zijn. Ik heb vaak met jullie over mijn onderzoek gehad en jullie met mij over jullie belangstelling voor wetenschappelijk onderzoek waar jullie nu ook mee bezig zijn. Ik geniet van jullie om jullie met zoveel passie bezig te zien en ik weet zeker dat jullie ook straks als arts en onderzoeker zullen bijdragen aan het verbeteren van de zorg.

Arman, je VWO-periode zit er bijna op. Fijn dat jij je wens om geneeskunde te willen studeren kunt realiseren. Je enthousiasme hiervoor is groot en zie ik bij jou ook een sterke interesse in de medische wetenschap.

Ik wil ook mijn familie en vrienden bedanken die ondanks de fysieke afstand altijd bij mij zijn.

CV |

Curriculum Vitae

Over de auteur

Masoud Salavati is geboren op 23 oktober 1964 te Ahvaz in Iran. In 1984 verhuisde Masoud naar Nederland, waar hij vervolgens in 1987 begon met de studie fysiotherapie. Nadat hij in 1991 de opleiding afgerond had, is hij als (kinder-)fysiotherapeut werkzaam geweest bij enkele fysiotherapiepraktijken en een kinderrevalidatiecentrum.

Sinds januari 1997 is Masoud als (kinder-) fysiotherapeut werkzaam bij Koninklijke Visio te Haren. In de loop van de jaren heeft hij ervaring opgedaan in het werken met kinderen en jongeren met een ernstige complexe meervoudige beperking. Waaronder een visuele, motorische en cognitieve beperking.

In de loop van de jaren volgde Masoud specialisaties op het gebied van kinderen met een meervoudige beperking. Daarnaast volgde hij van 1999 tot en met 2002 de opleiding kinderfysiotherapie en van 2009 tot en met 2011 de Masteropleiding kinderfysiotherapie bij de AVANSplus Hogeschool in Breda. De eindopdracht van zijn master vormde de start voor zijn promotie-onderzoek. Zijn doel is om bekendheid te geven aan de invloed van CVI op het dagelijks functioneren van kinderen met CP, en zijn kennis/ ervaring over deze kinderen en jongeren te delen met het werkveld van onder andere kinderrevalidatieartsen, kinderfysiotherapeuten, ergotherapeuten en overige hulpverleners.

SHARE

Research Institute SHARE

This thesis is published within the **Research Institute SHARE** (Science in Healthy Ageing and healthcaRE) of the University Medical Center Groningen / University of Groningen.

Further information regarding the institute and its research can be obtained from our internetsite: http://www.share.umcg.nl/

More recent theses can be found in the list below. ((co-) supervisors are between brackets)

2016

Göhner, C
Placental particles in pregnancy and preeclampsia
(prof SA Scherjon,prof E Schleußner, dr MM Faas, dr T Plösch)

Vries AJ de
Patellar tendinopathy; causes, consequences and the use of orthoses
(prof RL Diercks, dr I van den Akker-Scheek, dr J Zwerver, dr H van der Worp)

Holland B van
Promotion of sustainable employment; occupational health in the meat processing industry
(prof MF Reneman, prof S Brouwer, dr R Soer, dr MR de Boer)
13.04.2016 EXPAND

Otter TA
Monitoring endurance athletes; a multidisciplinary approach
(prof KAPM Lemmink, prof RL Diercks, dr MS Brink)

Bielderman JH
Active ageing and quality of life; community-dwelling older adults in deprived neighbourhoods
(prof CP van der Schans, dr MHG de Greef, dr GH Schout)

Bijlsma MJ
Age-period-cohort methodology; confounding by birth in cardiovascular pharmacoepidemiology
(prof E Hak, prof S Vansteelandt, dr F Janssen)

Dingemans EAA
Working after retirement; determinants and conzequences of bridge employment
(prof CJIM Henkens, dr ir H van Solinge)

Jonge L de
Data quality and methodology in studies on maternal medication use in relation to congenital anomalies
(prof IM van Langen, prof LTW de Jong-van den Berg, dr MK Bakker)

Vries FM de
Statin treatment in type 2 diabetes patients
(prof E Hak, prof P Denig, prof MJ Postma)

Jager M
Unraveling the role of client-professional communcation in adolescent psychosocial care
(prof SA Reijneveld, prof EJ Knorth, dr AF de Winter, dr J Metselaar)

Mulder B
Medication use during pregnancy and atopic diseases in childhood
(prof E Hak, prof SS Jick, dr CCM Schuling-Veninga, dr TW de Vries)

Romkema S
Intermanual transfer in prosthetic training
(prof CK van der Sluis, dr RM Bongers)

Diest M van
Developing an exergame for unsupervised home-based balance training in older adults
(prof GJ Verkerke, prof K Postema, dr CJC Lamoth, dr J Stegenga)

Waterschoot FPC
Nice to have or need to have? Unraveling dosage of pain rehabilitation
(prof MF Reneman, prof JHB Geertzen, prof PU Dijkstra)

Zijlema WL
(Un)healthy in the city; adverse health effects of traffic-related noise and air pollution
(Prof JGM Rosmalen, prof RP Stolk)

Zetstra-van der Woude AP
Data collection on risk factors in pregnancy
(prof LTW de Jong-van den Berg, dr H Wang)

Mohammadi S
The intersecting system of patients with chronic pain and their family caregivers; cognitions, behaviors, and well-being
(prof M Hagedoorn, prof R Sanderman, dr M Deghani)

Verbeek T
Pregnancy and psychopathology
(prof MY Berger, prof CLH Bockting, dr H Burger, dr MG van Pampus)

2015

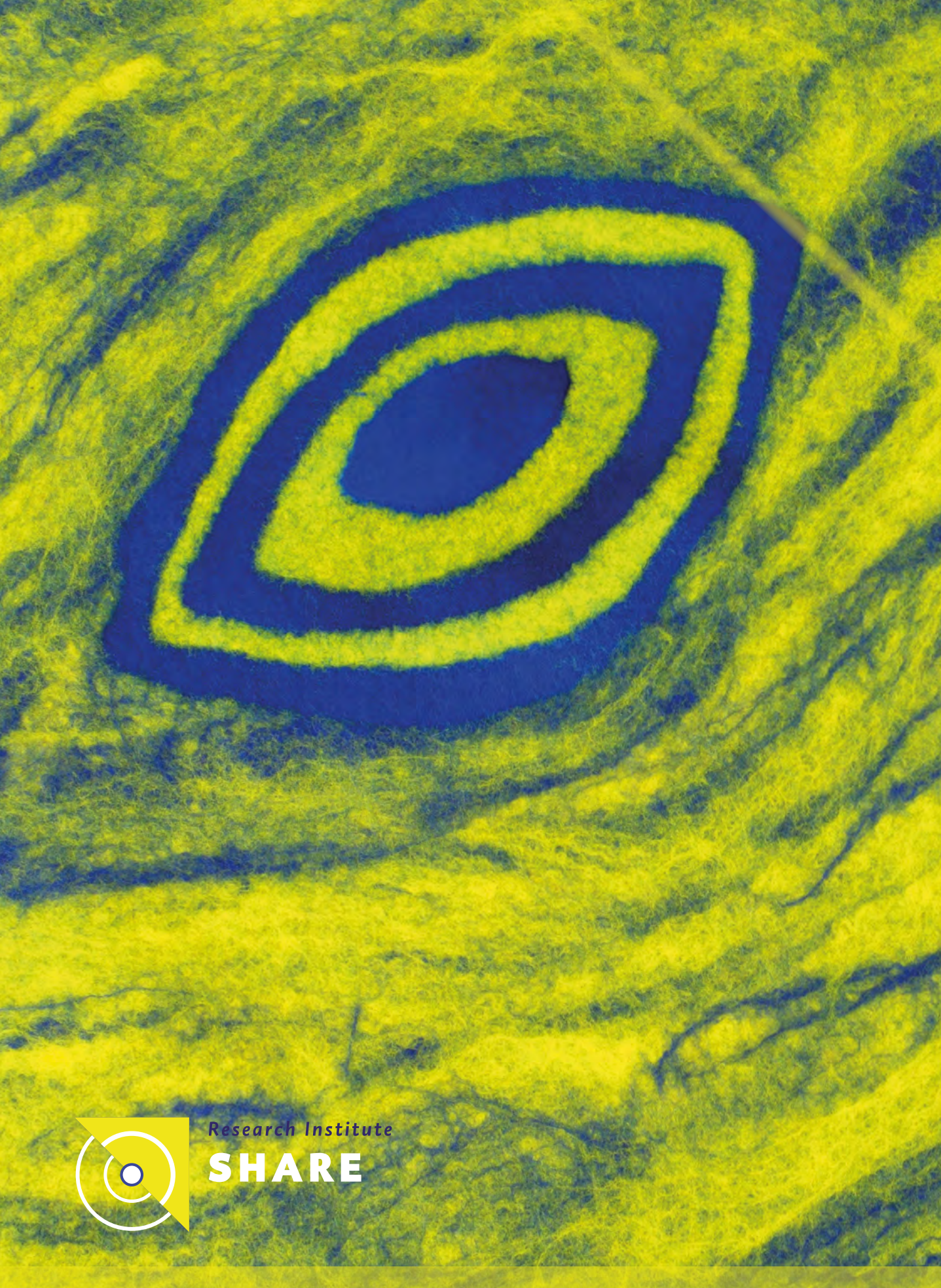
Broekhuijsen K
Timing of delivery for women with non-severe hypertensive disorders of pregnancy
(prof PP van den Berg, prof BWJ Mol, dr MTM Faassen, dr H Groen)

Tuuk K van der
Who's at risk? Prediction in term pregnancies complicated by hypertensive disorders
(prof PP van den Berg, prof BWJ Mol, dr MG van Pampus, dr H Groen))

Vitkova M
Poor sleep quality and other symptoms affecting quality of life in patients with multiple sclerosis
(prof SA Reijneveld, prof Z Gdovinova, dr JP van Dijk, dr J Rosenberger)

Sudzinova A
Roma ethnicity and outcomes of coronary artery disease
(prof SA Reijneveld, dr JP van Dijk, dr J Rosenberger)

For more 2015 and earlier theses visit our website


Research Institute
SHARE