Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI)
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Document Version
Publisher's PDF, also known as Version of record

Publication date:
2016

Link to publication in University of Groningen/UMCG research database

Citation for published version (APA):
Salavati, A. (2016). Assessing gross motor function, functional skills, and caregiver assistance in children with cerebral palsy (CP) and cerebral visual impairment (CVI) [Groningen]: Rijksuniversiteit Groningen

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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
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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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.\textsuperscript{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. 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.

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). 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.

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. 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. 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. 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. CVI frequently co-occurs with CP and is observed in approximately 30% of children diagnosed with various forms of CP. 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.

Children with CVI exhibit slow, inefficient, and highly variable visual functioning during daily-life. 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.

<table>
<thead>
<tr>
<th>Level</th>
<th>GMFCS</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>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.</td>
</tr>
<tr>
<td>II</td>
<td>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.</td>
</tr>
<tr>
<td>III</td>
<td>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.</td>
</tr>
<tr>
<td>IV</td>
<td>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. 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.</td>
</tr>
</tbody>
</table>
therefore, warranted to be taken into account in therapy. 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, 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. 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. Lack of recognition can be problematic for a child with CVI whose inaccurate visual guidance of movement, for example, may be misinterpreted as clumsiness. 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. 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. 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. and Da Costa et al. 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. Lueck and Dutton 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. 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.

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) and Gross Motor Function Measure-88 (GMFM-88). 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. 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. 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. 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 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. 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.

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.

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