Introduction and outline of the thesis

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Cerebral palsy (CP) is the most common cause of physical impairment in children. The prevalence of CP in Europe is approximately 2 in every 1000 live born children. The physical impairment is described as a persistent disorder of movement and posture caused by non-progressive pathological processes of the immature brain. The classification of motor disorders in CP retains the classical neurological terms for central nervous motor disorders, i.e. spastic, dyskinetic and ataxic. The physical motor impairment is the hallmark of CP. Fortunately, the last years more (and deserved) attention of the associated impairments of CP is emerging. This is reflected by the revised definition of CP in 2007, in which the authors stated that the motor disorder is often accompanied by disturbances of sensation, perception, cognition, communication, behavior, epilepsy and secondary musculoskeletal problems. In addition, by using the International Classification of Functioning, Disability and Health (ICF) framework, a more holistic approach for daily activity and participation of the child with CP is provided.

The severity of the motor impairment, as well as the associated impairments is different for each child with CP. This results in a large individual variability of functioning in all domains. It is therefore recommended that the diagnosis CP is used in combination with functional classification 1) for mobility, i.e. the Gross Motor Function Classification System (GMFCS) that describes the gross motor function on the basis of self-initiated movement with particular emphasis on sitting, walking, and wheeled mobility, 2) for manual use in daily activity i.e. the Manual Ability Classification System (MACS) that describes the manual abilities to handle objects in daily activities and, 3) for communication, i.e. the Communication Function Classification System (CFCS) that describes the effectiveness of everyday communication with familiar and unfamiliar partners. All classification systems apply a five level system with level I being least affected and level V corresponding to the most severe impairment in functioning (see table 1.1). Several studies have shown a relation between the severity of the CP (expressed in GMFCS levels) and associated impairments and frequency of disabilities in communication.

**Expressive and receptive communication**

Communication skills can be distinguished in receptive and expressive communication. Expressive communication refers to how a person (sender) conveys a message to a communication partner either by gesturing, by production of speech, by writing or signing. Receptive communication refers to what a person (listener) receives from a communication partner and understands of messages such as signs, symbols/reading comprehension and comprehension of spoken language. Disabilities in expressive and receptive communication can be associated with any type of CP, regardless of the nature of the central motor disorder. The impact of communication...
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<thead>
<tr>
<th>Level</th>
<th>GMFCS</th>
<th>MACS</th>
<th>CFCS</th>
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<tbody>
<tr>
<td>I</td>
<td>Walks without limitations</td>
<td>Handles objects easily and successfully</td>
<td>Sends and receives information with familiar and unfamiliar partners effectively and efficiently</td>
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<tr>
<td>II</td>
<td>Walks with limitations</td>
<td>Handles most objects but with somewhat reduced quality and/or speed of achievement</td>
<td>Sends and receives information with familiar and unfamiliar partners but may need extra time</td>
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<tr>
<td>III</td>
<td>Walks using a hand-held mobility device</td>
<td>Handles objects with difficulty; needs help to prepare and/or modify activities</td>
<td>Sends and receives information with familiar partners effectively, but not with unfamiliar partners</td>
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<tr>
<td>IV</td>
<td>Self-mobility with limitations; may use powered mobility</td>
<td>Handles a limited selection of easily managed objects in adapted situations</td>
<td>Inconsistently sends and/or receives information even with familiar partners</td>
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<tr>
<td>V</td>
<td>Transported in a manual wheelchair</td>
<td>Does not handle objects and has severely limited ability to perform even simple actions</td>
<td>Seldom effectively sends and receives information even with familiar partners</td>
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GMFCS, Gross Motor Function Classification System; MACS, Manual Ability Classification System; CFCS, Communication Function Classification System.
Adapted with permission from Hidecker et al. 

Table 1.1 (GMFCS, MACS en CFCS classification systems)
problems in the life of the child with CP and their family has far-reaching consequences and is associated with reduced quality of life in relationships with parents\textsuperscript{15} and problems with psychological adjustment.\textsuperscript{6,16} In addition, unintelligible speech (severe dysarthria) or no development of speech (anarthria) is associated with restrictions in communication between child and caregiver.\textsuperscript{17,18} Disabilities in expressive and receptive communication are present in 50 to 75\% of children with CP, and increasing to 100\% in children with severe CP (corresponding to GMFCS levels IV and V).\textsuperscript{6,10,12,19} Indeed, in children with severe CP, oro-motor impairments (i.e. severe dysarthria or anarthria), are generally accompanied by severe limited mobility (WHO, 2001).\textsuperscript{5} Because of the challenges associated with communication for children with severe CP, discrepancies between their expressive communication skills (production of speech, use of augmentative and alternative communication [AAC] systems) and their receptive communication skills (comprehension of spoken language) may exist.\textsuperscript{20-22} Yet, early insight into such a discrepancy is important to the support of the participation and development of the child. For example, insight into a child’s comprehension of spoken language has immediate consequences for how the communication partners of the child can most effectively interact with the child. Moreover, it can influence or provide valuable information on which level and/or type of education program and/or which augmentative and alternative communication system should be used.\textsuperscript{23-25}

**Spoken language comprehension**

Most studies involving communication skills in CP have focussed on expressive language, (verbal) cognition and overall communication abilities of children with CP described in all GMFCS levels.\textsuperscript{6,10,12,19} Limited information is available about the development of (spoken) language comprehension in children with severe CP. Comprehension of spoken language is defined as the ‘The act or fact of grasping the meaning, nature, or significance of a system of conventional spoken language’. Comprehension skills offer a base for acquiring meanings of symbols (graphic and other non-vocal symbols) and are necessary for the development of the child’s language and communication abilities.\textsuperscript{26} Moreover, comprehension of spoken words (receptive vocabulary) and later sentences (receptive grammar) can develop even when the child is not speaking.\textsuperscript{20,27} For children for whom expressive speech is not possible, comprehension of spoken language contributes to communicative exchanges and provides the basis for learning about language and the environment.\textsuperscript{25,26} Therefore, assessment of spoken language comprehension in children with severe CP is important.

In clinical practice, various diagnostic tests are available to assess a child’s comprehension of spoken language (for review: Geytenbeek et al.).\textsuperscript{28} However, none of these tests are designed specifically for children with severe motor impairment and anarthria or dysarthria. Investigation of spoken language comprehension is a real challenge in children with
severe CP because many diagnostic tests include fine motoric tasks such as manipulating with small material and finger pointing to small pictures. It is obvious that these tasks are difficult or impossible to perform by children with severe motor impairments. As a result, the information from the assessment may not be a true reflection of the child’s ability, as it remains unclear whether test errors arise from the child’s difficulty in performing the needed motoric skills to provide a response (e.g., pointing to a response item) or from poor language comprehension. In addition, line drawings often used as stimuli in tests may be more difficult to recognize by young children and/or children with cognitive impairments, often present in children with severe CP. Consequently, the use of standardized assessments may lead to an underestimation of the real comprehension abilities of the child. Yet, underestimation of spoken language comprehension may lead to under stimulation and social deprivation with its immediate negative effects on a child’s emotional wellbeing. In addition, the risk of overestimation exists when trying to judge a child’s comprehension without testing. For instance, in daily activities, circumstances can exist in which children appear to comprehend certain structures when in fact they do not. Therefore, the assessment of comprehension skills requires the use of appropriate measurement.

Research questions of the thesis
Prompted by the lack of an instrument meeting test requirements in children with severe CP, our group developed a computer-based diagnostic instrument. This instrument, named “Computer-Based instrument for Low motor Language Testing” (C-BiLLT) is an instrument to assess spoken language comprehension in children with severe CP, corresponding to level IV and V of the GMFCS. The C-BiLLT was designed in a way that (1) it requires a minimum of motor action (performed by any body part) to respond to the questions of the test, and (2) items pertain to spoken language comprehension abilities on a sentence level and (3) items refer to the experiential environment of the child with severe impaired mobility.

In addition, neuroimaging plays an important role in elucidating the aetiology of CP. Patterns of brain damage explain the major period of pathogenesis in CP. The most common pattern of brain abnormality in preterm born children with CP is white matter injury, (including periventricular leukomalacia [PVL]). The most common pattern of brain abnormality in term born children with CP is basal ganglia and thalamic lesions (BGN) and malformations of the brain. In understanding the impact of these lesions on brain function MRI findings may fulfil an important role in predicting the neurodevelopmental outcome of spoken language comprehension of the child with severe CP. More specific, what is the effect of the preferred damage of the white matter in PVL and grey matter in children with BGN lesions on spoken language comprehension in children with severe CP. Thus far, information on the relationship between these brain abnormalities and language comprehension in severe CP is lacking.
In this thesis, the following research questions are addressed:

1) Which language tests are available for the investigation of spoken language comprehension in non-speaking children with severe CP, and are these adequate?

2) How adequate is the newly developed computer-based instrument for low motor language testing (the C-BiLLT) for assessment of spoken language comprehension in non-speaking children with severe CP and what are its psychometric properties?

3) What are the spoken language comprehension abilities of children with severe CP compared to typically developing children?

4) What are the associations between spoken language comprehension and type and severity of the brain damage in children with severe CP?

Outline of the thesis

Followed by the introduction in Chapter 1, which presents the topic of this thesis and addresses the research questions, Part 1 of the thesis describes the development of the computer-based instrument to assess spoken language comprehension in children with severe CP. In Chapter 2 a systematic review of the literature was conducted in order to identify the use and utility of language tests applied to measure spoken language comprehension in children with CP. Chapter 3 introduces the computer-based instrument for low motor language testing (C-BiLLT). In a pilot study, consisting of 18 children with severe CP (aged 1;6 – 12;0 years) and a control group of 45 typically developing (TD) children (aged 1;2 – 6;6 years), the feasibility of the newly developed C-BiLLT is investigated. In this study the administration and preliminary psychometric measures of the C-BiLLT are reported. In Chapter 4 the expanded and current version of the C-BiLLT is presented. The psychometric properties of the C-BiLLT were tested on a large cohort of 806 TD children (aged 1;6 – 7;6 years) and 90 children with severe CP (aged 1;7 – 11;11 years). Results on reliability and validity of the C-BiLLT are described comprehensively.

Part 2 of the thesis presents the results of the assessment of spoken language comprehension in children with severe CP with the C-BiLLT. In Chapter 5 spoken language comprehension in 87 unintelligible or non-speaking children with severe CP is compared with normed scores of TD children to determine their comprehension related to the norm. Measures of spoken language comprehension were assessed with the current version of the C-BiLLT. The apparent difference in development of spoken language comprehension found between children with spastic and dyskinetic CP is described. Chapter 6 describes the development of sentence comprehension in children with severe CP compared to TD children and describes the role of motor type of CP and associated factors such as gender and parental education on sentence type comprehension in children with severe CP. In this cohort of 68 non-speaking children with severe CP (aged 1;9 – 11;11 years), comprehension of seven different sentence types (covering 60 items of the C-BiLLT) are examined and compared with the percentages correct of 806 TD children (aged between 1;6 – 7;6 years).
Part 3 of the thesis addresses the relation between findings of neuroimaging and spoken language comprehension abilities in children with severe CP. In Chapter 7 MRI findings of 80 children with severe CP (aged 1;6 – 12;0 years) were compared with outcomes of spoken language comprehension assessed with the C-BiLLT. The influence of MRI pattern as well as severity of white matter and grey matter injury on spoken language comprehension abilities are described. Chapter 8 describes an explorative study which included 5 children with severe CP (aged 5-23 years) and a control group of 10 children without CP (aged 5-18 years). The arcuate fasciculus and the extreme capsule, two important language tracts between Wernicke’s and Broca’s area, were visualized by using Diffusion Tensor Imaging (DTI). Spoken Language comprehension was assessed with the C-BiLLT. A possible relation between language tracts and language skills was examined.

Part 4 of the thesis contains the general discussion and summary and discusses the results of the four research questions. Chapter 9 contains the general discussion which emphasizes the possibilities and future benefits of the appliance of the C-BiLLT and potential applications of C-BiLLT measures and MRI findings in order to follow the development of spoken language comprehension in non-speaking children with severe CP. Implications for further research are discussed.

References


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