Summary

Comprehension of spoken language in non-speaking children with severe cerebral palsy

Development and application of a newly developed Computer-Based instrument for Low motor Language Testing (C-BiLLT)
SUMMARY
This thesis describes the development and application of a specifically developed instrument for assessment of spoken language comprehension in non-speaking children with severe CP (classified as GMFCS IV and V). This instrument, called Computer-Based instrument for Low motor Language Testing (C-BiLLT), enables reliably and standardized investigation of spoken language comprehension in non-speaking children with severe CP. Recruited throughout the Netherlands, participants included 90 children with severe CP (aged 1;6-12;0 years) and 806 typically developing children (aged 1;2-7;6 years) to provide normed data. The following research questions were 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? (Part 1 of this thesis)
2) How adequate is the C-BiLLT for assessment of spoken language comprehension in non-speaking children with severe CP and what are its psychometric properties. (Part 1 of this thesis)
3) What are the spoken language comprehension abilities of non-speaking children with severe CP compared to typically developing children? (Part 2 of this thesis)
4) What are the associations between spoken language comprehension and type and severity of the brain damage in non-speaking children with severe CP. (Part 3 of this thesis)

Chapter 1 compromises the introduction and outline of the thesis and addresses the research questions.

Part 1 of the thesis comprises chapter 2, 3 and 4 and describes the development of the C-BiLLT to assess spoken language comprehension in non-speaking 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 language comprehension in children with CP. Twelve standardized tests and five experimental instruments were identified. All standardized tests were developed for children without limited mobility. Only the Peabody picture vocabulary test (PPVT-R), a receptive vocabulary test, was frequently used and feasible for older children with severe CP (> 9 years). The other identified tests were used occasionally. To establish utility, adaptations of standardized test procedures were necessary. In addition, no follow-up or further development of psychometric qualities of any of the experimental instruments were detected. In summary, adequate and diagnostic tools specifically designed for the assessment of comprehension of spoken language of non-speaking children with severe CP were not available and the development of such an instrument was warranted.
Chapter 3 introduces the C-BiLLT and describes the pilot study conducted to investigate the feasibility and primarily psychometric properties of the first version of the C-BiLLT. The C-BiLLT was well accepted by children with severe CP (n=18) and children with typical development (n=45) (control group). The pilot version of the C-BiLLT showed promising construct, convergent and face validity. The experimental set-up and different access methods of the C-BiLLT (such as eye-gazing, eye-gaze computer control, touchscreen, input switches operated by any body part of choice, the child’s own wheelchair head support) enabled even the most severely motor impaired children to respond autonomously to the test items of the C-BiLLT. However, significant changes (such as additional items) to the test were required and enlargements of the control group and CP group were necessary to guarantee adequate measures of reliability and validity.

Chapter 4 presents the results of the investigation of the reliability and validity of the extended and present version of the C-BiLLT (from 53 to 77 items) (see Appendix I). Reliability and validity of the C-BiLLT was investigated in 87 non-speaking children with severe CP and 806 typically developing children. The C-BiLLT was accessible for children with severe CP who are often unable to participate in standardized assessment (97% completed C-BiLLT assessment). The current version of the C-BiLLT provides evidence of good reliability (as measured by ICC and SEM), both intra-observer and inter-observer reliability showed excellent results for children with typical development (TD) and for children with severe CP. With regard to the structural validity of the C-BiLLT, the factor analyses (the factor had an explained variance of 76%) revealed a clear unidimensional structure, best characterized as comprehension of spoken language. In addition, with regard to the hypotheses on convergent and discriminant validity as measured by statistical relationships with other instruments, we observed high correlations with existing language and receptive vocabulary tests (RDLS and PPVT-III, respectively) and moderate correlation with a test for non-verbal logic reasoning (CPM). The internal consistency of the C-BiLLT was found to comply with the standards (measured with Conbach’s alpha and Guttmans Lambda 2) for a diagnostic instrument. All hypotheses concerning the differences between subgroups were confirmed. Correlation between C-BiLLT score and chronological age was high in TD children \( r = 0.82 \) and moderate in children with severe CP \( r = 0.36 \). To summarize, reliability and validity measures of the TD children on the C-BiLLT justified the calculation of normed data to establish a norm-referenced test for the assessment of spoken language comprehension in non-speaking children with severe CP. With these normed data, the C-BiLLT can provide information on (Dutch) spoken language comprehension performance of non-speaking children with severe CP relative to their peers without disabilities.
C-BiLLT assessments may offer parents and professionals important information on a child’s language comprehension skills. With the standardized administration of the C-BiLLT and different accessible methods, the C-BiLLT enables reliable measures of spoken language comprehension in non-speaking children with severe CP.

Part 2 of the thesis comprises chapter 5 and 6 describing spoken language comprehension performance of non-speaking children with severe CP compared to a norm group of typically developing children.

In chapter 5 comprehension of spoken language in 87 non-speaking children with severe CP was investigated with the C-BiLLT. Comprehension of spoken language varied considerably among children with severe CP and was overall significantly delayed compared to TD children. Despite a comparable level of functional limitations, including gross motor and communication disabilities, children with dyskinetic CP performed better than spastic children. Of the children with dyskinetic CP, in those aged under 6;6 years, more than 50% attained comprehensive language scores within the normal range. Overall data suggest that the underlying brain lesions resulting in different types of CP not only have an effect on the extent and characteristics of the motor impairment, but also on language abilities, at least on receptive abilities. The assumption that spoken language comprehension may be relatively spared in dyskinetic CP stresses the importance of distinguishing between expressive language (speech and gestures) and receptive language (language comprehension) when diagnosing non-speaking children with severe CP.

In addition, in contrast to earlier studies that report on most evident restrictions in communication in children with the most severe motor impairments (i.e. GMFCS levels IV or V), our data are less explicit regarding how GMFCS levels are associated with language comprehension. C-BiLLT raw scores were found to be poorer in children classified as level V than in level IV, and the level of gross motor functioning made a significant contribution to the variability in spoken language comprehension. Moreover, when calculated in z-scores, a significant association was found between GMFCS level and spoken language comprehension abilities. However, when calculated in age-equivalent (AE) scores, the association between GMFCS level and spoken language comprehension was no longer significant. This inconsistency in outcome may be due to the differences between the subsamples and/or because of several limitations associated with AE scores.

Chapter 6 describes an in-depth study of sentence comprehension of phrases, simple and complex sentences in 68 non-speaking children with severe CP compared to TD children. These 68 children, who passed the pretest of the C-BiLLT (matching at least five spoken words to the corresponding objects) and advanced to the computer-test part of the
C-BiLLT, were investigated for comprehension of seven different sentence types. Analyses showed that for children with severe CP who have reached a basic linguistic level, sentence comprehension appears to develop up to at least age 12 years and tends to follow the developmental trajectory of typically developing (TD) children, albeit at a much slower rate. Delays in sentence type comprehension increased with sentence complexity and showed a large variability between individual children and between subtypes of CP. Comprehension of phrases showed similar results between children with spastic and dyskinetic CP while comprehension of simple and more complex sentences were significantly better in children with dyskinetic CP than in children with spastic CP. Of the children with dyskinetic CP, 10-13% showed comprehension of complex sentences within the percentage correct of TD children as opposed to none of the children with spastic CP.

Part 3 of the thesis comprises chapter 7 and 8 describing findings of neuroimaging in non-speaking children with severe CP related to outcomes in spoken language comprehension.

In Chapter 7 the relation between MRI pattern, grey matter (GM) and white matter (WM) lesions and spoken language comprehension in 80 non-speaking children with severe CP was investigated. In our study, associations between MRI patterns and language comprehension were not very explicit, other than that children with a basal ganglia necrosis (BGN) pattern generally scored better than children with a periventricular leucomalacia (PVL) pattern. Language comprehension of children with a miscellaneous pattern or malformations showed similar results. For the additional investigation of the relationship between brain lesions and spoken language comprehension, associations between WM and GM lesions and language comprehension scores, were determined in a subgroup of 62 children (malformations excluded) and controlled for chronological age. Analyses of specific brain lesions of WM and GM areas, and cerebellum, revealed that, especially WM lesions, are highly associated with moderate to severe delays in language comprehension. In contrast to GM lesions, our findings emphasise the consistent relationship between WM lesions and neurodevelopmental outcomes. In particular, corpus callosum, and frontal and parieto-occipital WM areas, were associated with spoken language comprehension. In our cohort, diffuse thinning of the corpus callosum, and diffuse reduction of the frontal and parieto-occipital WM areas, consistently corresponded with poor spoken language comprehension, indicating the importance of inter- to intrahemispheric connectivity for language development. In severe CP, the brain abnormality pattern on MRI seems not important for understanding language comprehension, whereas concomitant damage to especially WM areas is. Language comprehension was most affected in children with WM lesions in the subcortical and then periventricular areas, most characteristic for children with PVL.
In Chapter 8 using diffusion tensor imaging (DTI), two important language tracts (i.e. the arcuate fasciculus and the extreme capsule) were visualized in both hemispheres. Most children with severe CP (n=5) had detectable language tracts, though smaller and with lower fractional anisotropy (FA) than healthy controls (n=10), suggesting a lower integrity. This disturbed integrity of language pathways coincided with severely hampered language comprehension. Moreover, in one child with severe white matter loss on conventional MRI, who did not respond to the C-BiLLT questions at all, the language tract in the left hemispheres could not be visualized at all and the right tract was small with a very low FA value. Structural identification of language tracts using DTI could be important in understanding language impairment in CP, justifying further research in this field.

Part 4 of this thesis comprises chapter 9 which contains the general discussion.

In chapter 9, the general discussion addresses the main findings of this thesis. Methodological considerations and implications for clinical practice are discussed. In addition recommendations for future research are addressed.

The C-BiLLT, with its standardized assessment procedure including different access methods, enabled reliable measures of spoken language comprehension in children with severe CP. The main findings of the studies were that the C-BiLLT showed good validity and reliability results. Adequate and early insight of a child's language comprehension skills can provide valuable information to optimize the child's language environment in daily activities and participation.

Furthermore, results of spoken language comprehension in children with severe CP revealed that despite a comparable level of functional limitations, including disability in communication, children with dyskinetic CP performed better than spastic children. In addition, in the present study, sentence comprehension of children with CP (who have reached a basic linguistic level) was delayed rather than deviant and seemed to develop up to at least 12 years. The results of the present study show that it is important to follow the linguistic development of the child with severe CP more gradually, even beyond age 12 years, and to continue to offer tools for development of spoken language comprehension. Moreover, our study emphasizes the importance of not only determine the severity of motor impairment but also the nature of the underlying brain injury, with a particular focus on WM lesions for initiating appropriate and accurate stimulation of the child and its environment.

Recommendations for future research included continued work on the C-BiLLT to maintain a constructive instrument and follow-up research with longitudinal data of C-BiLLT test results to monitor development of spoken language comprehension in children with severe CP. Finally recommendations for practice are presented.