Effective use of the assisting hand in adolescents with cerebral palsy
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General discussion
Cerebral Palsy (CP) influences the acquisition of age-appropriate independence and may negatively impact the development of the skills that children and adolescents with CP need in order to participate in activities that are important to them in the home, school and community. Different rehabilitation programmes targeting the upper extremity in CP are used to improve hand use and the performance of patient-relevant tasks. The general aim of this thesis was to improve knowledge regarding the assessment and management of upper extremity functioning in children and adolescents with CP. The studies involved children and adolescents whose neurological dysfunction and upper limb musculoskeletal impairments significantly affected their ability to use their hands to perform daily activities.

In this last chapter, the main findings of the studies are critically discussed, and implications for clinical practice and future research are given.

Main findings

Assisting Hand Assessment for Adolescents: evidence of validity and reliability

The Assisting Hand Assessment (AHA) measures and describes how effectively children with unilateral disability use their affected hand to perform tasks requiring the use of both hands. The first part of this thesis showed that the newly developed test activity, the Ad-AHA board game ‘Go with the Floe’ (Fig. 1), can be used in clinical practice to elicit bimanual performance in adolescents with unilateral CP. To evaluate the construct validity of the AHA test items, all 126 assessments of adolescents were scored using the Kids-AHA version 5.0 scoring criteria containing 20 items (AHA 5.0 scale). The AHA scale for the age range of our sample (10 to 18 years) functioned as a unidimensional measure that met quality criteria, as assessed using Rasch measurement model, and therefore we combined our data with data from an earlier AHA study involving children aged 18 months to 12 years with unilateral CP. With the combined sample (290 assessments) the scale demonstrated good construct validity, which means that the same AHA scoring criteria can be used for children and adolescents aged 18 months to 18 years (AHA 18-18) with unilateral CP (chapter 2). Four different test activities can be used (School-Kids AHA, Ad-AHA Board Game, Ad-AHA Present, and Ad-AHA Sandwich) to elicit bimanual actions involving the same underlying trait. Chapter 3 showed that the test scores for different test activities were comparable, demonstrating that reliable AHA scores can be generated when using different age-appropriate tests situations for children and adolescents aged 18 months to 18 years. The excellent measurement agreement implies that the Ad-AHA can be used to evaluate change over time, with a score change of 5 AHA units indicating a change beyond measurement error with 95% certainty, using the AHA 5.0 scale (chapter 3).

With the newly developed Ad-AHA, we fulfilled the need for a measurement tool to assess how effectively adolescents use their affected hand in bimanual activities. According to the 'International Classification of Functioning, Disability and Health' (ICF), assessment of functioning in the domain “activities and participation” should address capacity as well as performance. Capacity refers to a person's functioning in a standardized environment (can do) whereas performance refers to the performance in a real life environment (does do).

So far, tools for assessing capacity and perceived performance were only available for use in adolescents with CP. Capacity can be measured with the Melbourne Assessment, the Box and Block Test, the Jebsen-Taylor Hand Function Test and the Shriners Hospital for
Children Upper Extremity Evaluation (SHUEE), whereas perceived performance can be measured with the Abilhand questionnaire, the Canadian Occupational Performance Measure (COPM), and the Children's Hand-use Experience Questionnaire (CHEQ). Studies have shown that the SHUEE can be used to assess predominantly capacity, but it also measures the performance of activities. The SHUEE was developed to assist in targeting and measuring the outcome of spasticity and surgical management. Besides analysing grasp and release, it also assesses the spontaneous functional use of the upper extremity during standardized simple tasks (e.g. taking money from a wallet, tearing paper). The added value of the (Ad-)AHA is that it measures and describes how effectively patients use their affected hand in bimanual performance. To ensure that the person’s spontaneous use is measured, the new Ad-AHA presents the test activity as a board game, which requires bimanual handling of objects and is age appropriate and familiar to most adolescents. While the adolescent is playing the game, the therapist is allowed to interact and help if needed to keep and make it fun and engaging so as to elicit typical/habitual performance. This way of presenting an activity ensures that functioning is observed in an objective manner, without the subject even realizing that the activity is being assessed. Our study in chapter 3 showed that the different test activities are age appropriate and can be used interchangeably in adolescents with unilateral CP, which makes it more attractive to adolescents and therapists, which in turn contributes to actual (spontaneous/habitual) performance. The excellent agreement between scores obtained with different test activities (School-Kids AHA, Ad-AHA Board Game, Ad-AHA Present, and Ad-AHA Sandwich) indicates that different test activities can be used to evaluate change over time (Chapter 3). It should be kept in mind that only changes ≥5 AHA units can be interpreted as real changes beyond measurement error, which suggests that the AHA may not be sensitive enough to detect clinically relevant changes at the individual level. Information on the minimal clinically important difference for the AHA is lacking but from clinical experience it is known that smaller changes can be very meaningful to the child and that the size of a meaningful change may vary across different levels of ability. The minimal clinically important difference needs further study.

Functional hand orthosis

The second part of this thesis showed that a functional hand orthosis (static bracing of the wrist and thumb) immediately improved the spontaneous use of the affected upper limb for bimanual activities in selected children with spastic unilateral CP (chapter 4).

Since this study, two studies with higher levels of evidence that evaluated the effect of functional hand orthoses have been published. These two randomized controlled trials (RCTs) found a functional hand orthosis not to have an effect on capacity, assessed using the “Melbourne Assessment quality of upper limb movement” and the Box and Blocks Test (BBT). These findings seem to be in contrast with our findings, but are not directly comparable because we assessed predominately the typical/habitual performance of activities and not the capability to execute activities. In agreement with the findings of these RCTs, we found large individual differences in the effects of using the orthosis. This stresses the need for individualized tailored assessment and therapy. The item hierarchy of the items of the AHA provide insight into the type of activities for which the individual child will most likely benefit from using the orthosis (chapter 4). Our study and the two RCTs evaluated the effects of intermittent use of a functional hand orthosis, so no conclusion can
be drawn about the effects of intensive use (during the whole day) of an hand orthosis in the short and long term. Little is known about the long-term effects of a hand orthosis because of the scarcity of relevant studies, but positive and negative results of long-term use can be anticipated. One study observed decreased activation of the forearm muscles during grip and suggested that bracing for a long period may lead to atrophy of the forearm muscles. However, this adverse long-term effect was less apparent when a functional hand orthosis was used intermittently. It is expected that long-term intermittent use a functional hand orthosis will result in an increase in hand use and performance, when combined with goal-directed training, the value of which has been demonstrated by Elliot et al. However, further studies are needed to obtain insight into the long-term effectiveness of wearing a functional hand orthosis in combination with training and, in possible side effects.

**Upper extremity surgery**

Our review (chapter 5) showed that there is insufficient evidence to recommend upper extremity surgery to improve hand use and performance in children and adolescents with CP. The quality of evidence was very low for each postoperative outcome after upper extremity surgery, mainly because of heterogeneous surgical interventions, different activity-based outcome measures, and poor-quality studies with uncontrolled designs. However, the consistent positive results of upper extremity surgery presented in the 12 included studies justify the initiation of additional, preferably controlled, studies. These studies showed that the ability to use the hand(s) to perform tasks on request (capacity) or spontaneously (performance) and the perception of the patient's ability to use the hand(s) improved significantly after upper extremity surgery. An RCT is the best study design for determining treatment effectiveness. However, different factors, such as the invasive and irreversible character of surgery, and patient preferences, may limit the feasibility of RCTs for studies on upper extremity surgery in children and adolescents. This might be the reason that only 1 of the 12 included studies in our review evaluated the effectiveness of upper extremity surgery in a RCT. This comparative study showed that tendon transfer surgery was more effective than botulinum toxin injections or regular, ongoing therapy in children with upper-extremity CP. The next best design to evaluate the effects of upper extremity surgery may be a cohort study design with a long follow-up, in which patient selection criteria are described in detail and pre-treatment characteristics are studied to identify predictors of (long-term) treatment outcome. A potential predictor of treatment success may be the age at which upper extremity surgery is performed. Currently, there is no evidence available about the timing of upper extremity surgery and the optimal age range for surgical intervention in patients with CP. This requires careful long-term follow-up and monitoring into adulthood.

Our clinical cohort study (chapter 6) showed that a comprehensive multidisciplinary assessment and the use of strict selection criteria contributed to the effectiveness of surgery in our patient population. From this study, we can conclude that children and adolescents with unilateral CP who are able to handle most objects with somewhat reduced quality and/or speed (MACS level II) will probably benefit from surgery, based on assessments evaluating hand use and performance. Given our high proportion of successful outcomes, it can be questioned whether our selection criteria for upper extremity surgery were too strict and more patients might have benefited from surgery. To establish better
criteria for patient selection, larger cohort studies with patients of different ages, CP subtypes, and CP severity are needed. However, some selection criteria are in common use. For example patients with dystonia or athetosis are frequently excluded from surgery, because the outcome of surgery is unpredictable owing to the variable muscle imbalance.

The first selection of patients who are referred to a multidisciplinary upper extremity surgical team is made by the child’s own physician. Medical professionals and therapists working with patients with CP have the possibility to choose from a range of different interventions. Selecting the intervention that is most appropriate for the individual patient is based on best research evidence, clinical expertise/experience and patients values and preferences. Surgery was once one of the interventions of first choice. In the last decades, more studies have compared the effect of non-surgical interventions, such as constraint-induced movement therapy (CIMT), hand-arm intensive bimanual training (HABIT), intramuscular injections of botulinum toxin A (BoNT-A), and intensive occupational therapy (OT). Because of the high-level evidence supporting these interventions, they are the first to be chosen by the medical professionals, and only a select group of children are referred to an upper extremity surgery team. It can be questioned whether this pre-selection for upper extremity surgery by the referring physician contributes to the exclusion of patients who might also benefit from surgery. We expect that this pre-selection influenced our outcomes, because we may have assessed only those children and adolescents in whom one of the other interventions did not provide (long-term) improvement in hand use and performance.

Methodological considerations

Interpretation of Assisting Hand Assessment scores

At the time we evaluated the effect of a functional hand orthosis (chapter 4) AHA scores were mostly reported as raw scores, and changes in raw scores (sum of item scores) were presented rather than logit-based scores. A change on the AHA of 4 or more points between two test sessions (with 95% certainty) was considered a clinically relevant change. In 2012, Krumlinde recommended the use of AHA units instead of raw scores when interpreting change after interventions at an individual level. She concluded that simply adding up raw scores of unknown intervals might misrepresent results. For raw scores, changes at the lower or upper ends of a scale may underestimate the real increase in ability as compared with changes in the middle of the scale. When applying these new insights we found that we had overestimated the effect of a functional hand orthosis - now 32% instead of the reported 52% of the children had a significant clinically relevant change. Thus one in three children benefited from static bracing of the wrist and thumb when it came to using their affected hand to perform tasks requiring the use of both hands.

Different constructs

To obtain full insight into the performance of activities of a child/adolescent, two different performance-based assessments are needed, reflecting two different constructs. In chapter 6, we used outcome measures for perceived performance (COPM, ABILHAND, and VAS) as well as the AHA measuring spontaneous (habitual/typical) use of the affected hand during bimanual task performance. Although one might expect perceived and typical performance
to be correlated, we could not demonstrate this in our sample – we did not find an association between the Abilhand and AHA ($r = 0.012, p = 0.95$). The lack of correlation indicates that different aspects of the constructs are measured when evaluating performance of activities with these two instruments. The Abilhand measures perceived performance in which the patient or parents use a questionnaire to indicate how daily activities are performed (with ease, with difficulty, or not possible to perform at all). How the patient performs the activity, i.e. with one or two hands or by using compensation strategies, is not evaluated. In contrast, with the AHA a therapist observes and scores how effectively the affected hand is used to perform activities requiring the use of both hands. A difference may occur because the AHA provides an objective measure of functioning, so that hand use may be considered less effective with the AHA than with the Abilhand. This difference highlights the need to evaluate the performance of activities with outcome measures that evaluate both perceived and typical/habitual performance, in order to refine treatment.

Generalizability

Both effect studies (chapters 4 and 6) included a selected group of patients with CP, which limits the generalizability of findings to the overall CP population. We selected a specific age range in both studies. The first study (chapter 4) included children up to 12 years of age. This age range was chosen because at the start of this study, the AHA had been validated in children aged 18 months to 12 years only, and thus the effect of a functional hand orthosis could not be ascertained in adolescents with CP older than 12 years. The other study (chapter 6) included a high proportion of adolescent (79%, age range 13 to 19 years). The positive effects of upper extremity surgery on hand use and performance found in this study were mostly based on outcomes measured with the Ad-AHA in this older age group. Therefore, we are not able to draw conclusions about the effect of surgery in young children, who also might benefit from surgery.

A substantially higher proportion of children and adolescents with unilateral CP were included in studies of chapter 4 (100%) and chapter 6 (87%) than would be expected on the basis of the distribution of CP subtypes in Europe, in which just over a quarter of all children with CP (29.2%; 95% CI 27.9 to 30.4) have unilateral spastic CP. Due to the low number of children with bilateral CP no conclusions can be drawn about the effect of a functional hand orthosis or upper extremity surgery in this population, in which more than 60% of the children have decreased hand function. This suggests that there is a need for more evidence about the effects of these interventions in patients with bilateral CP.

We included mainly patients with a relatively high ability to handle objects in important daily activities, as classified according to the MACS. However, patients with lower or even higher ability level might also benefit from upper extremity surgery. Our study showed that the proportion of patients classified as MACS level II (patient able to handle most objects but with somewhat reduced quality and/or speed of achievement) was 72%, which is not consistent with the earlier reported proportion of 20-25%. These differences in patient population (age range, type of CP and MACS level) may have caused an overestimation of the effects of upper extremity surgery or use of a functional hand orthosis.
Measuring performance in a clinical setting

Both intervention studies (chapters 4 and 6) investigated the impact of CP on an individual's ability to perform activities, as assessed with the AHA.²⁰

In the ICF, performance is understood as “involvement in a life situation” or “the lived experience” of people in the actual context in which they live.⁷ Thus assessing performance in a clinical setting seems contradictory. The use of (semistructured) test kits and the presence of a therapist violates the social context. Therefore, in strict sense capacity is evaluated.

That said, it is not always practical or feasible for therapists to observe and assess patients in their own environment.³² With the (Ad-)AHA it is possible to assess spontaneous use of both hands in a semistructured context, using different test activities (Ad-AHA Board game, Ad-AHA Sandwich and Ad-AHA Parcel) of which the adolescent may choose. In the case of the AHA, the best context is a room with a table, chair, and an activity which evokes the spontaneous use of both hands and which can be assessed afterwards, using a video recording of the activity. The setting itself, the clinic, is of less influence because the activity involves a play session with a board game allowing interaction with family members. The Ad-AHA board game ensures that functioning is observed objectively, without the subject even realizing that the activity is being assessed.³ All these aspects together makes the (Ad-) AHA an appropriate instrument to capture how well the affected hand is spontaneously used as an assisting hand during bimanual task performance, this in contrast to instruments measuring a person's best capacity.⁴,³,¹⁷-¹⁹

Implications for clinical practice

Assisting Hand Assessment for Adolescents

The newly developed and validated test activity, the Ad-AHA board game (Chapter 2 and 3), is already available and in use in clinical practice to elicit bimanual performance of activities in adolescents with unilateral CP.³,⁶ With the AHA 18-18 it is possible to evaluate interventions, monitor development from childhood to young adulthood, and guide intervention planning.³ (Fig. 1)
The Ad-AHA has proven its merits as outcome measure for adolescents with unilateral CP in clinical practice, and in the study evaluating the effect of upper extremity surgery on hand use and the performance of daily activities (Chapter 6). The AHA 18-18 covers the age range 18 months to 18 years, which makes it possible to follow the development of the effective use of the affected hand while performing bimanual tasks and provides information about the possible improvement or deterioration of hand skills with age. In clinical practice, it is important to monitor how hand skills change from infancy to adulthood, because hand skills develop rapidly during the first few years of life, followed by a refinement of these skills until adolescence, after which further refinement occurs at a lower rate until adulthood. Skills are refined when children/adolescents learn new age-related skills that are important for daily life or after an intervention at older ages. In addition to this age-related refinement, hand skills may also deteriorate with age. Both fine and gross motor development has been reported to decrease in children with low ability after 11 years of age. The impaired function may persist into adulthood, and the ability to use the affected hand may deteriorate further in later life, as a result of contractures, muscle weakness, and atrophy, all of which may increase in adulthood. Being able to measure this development (improvement or deterioration) in hand skills will help professionals to plan appropriate interventions.

Functional hand orthosis

Static bracing of the wrist and thumb improves hand position for performing manual tasks, and thus braces should be considered as a treatment option to improve functional performance (Chapter 4). It should be realized that not all children with unilateral CP will immediately benefit from a functional hand orthosis. In our study, some children were less able to extend their fingers and to grasp and release objects while wearing the functional hand orthosis, and this should be taken into account when a functional hand orthosis is considered as a means to improve hand use and the performance of activities. Some children will need time to get used to an orthosis. By evaluating performance in terms of individual AHA items, the therapist will be able to suggest specific activities for which the functional hand orthosis will be beneficial. For example, a more neutral position of the wrist may help the child to fixate paper more effectively while writing, or when combined with a thumb out of the palm the orthosis will enable the child to grip bicycle handlebars more effectively with both hands.

Upper extremity surgery

Eligibility for upper extremity surgery should be based on extensive multidisciplinary assessment. Different instruments should align surgical and postoperative treatment plans in order to achieve the best outcome in terms of patient-specific goals related to the performance of activities in daily life. As the heterogeneity of study outcomes precludes meaningful meta-analysis of data, it would be advisable to use a core set of instruments to improve selection criteria, to support the choice of surgical intervention, to assess the effect of surgery, and to predict outcomes.

Use of a core set of instruments would also make it easier to compare the results of different studies. In addition to the instruments used in our study (COPM, AHA, ABILHAND, and BBT, chapter 6), a validated instrument measuring patient satisfaction with cosmetic appearance would provide important information about patient-specific goals.
Furthermore, adding the “Shriners Hospitals Upper Extremity Evaluation” dynamic positional analysis (SHUEE-DPA) allows to measure and link “function” and “activity” when evaluating the result of surgery, by analysing function and describing the position of the thumb, fingers, wrist, forearm, and elbow during 16 different activities. The COPM can be used to identify patient-specific “hand-use oriented” goals, which may be linked to the different surgical procedures. Information from the Abilhand, AHA, and BBT should be collected to obtain insight into the ability of the patient to use the affected hand and the likelihood that COPM goals will be achieved. If the results from the Abilhand, AHA, and BBT show a very low ability to use the affected hand effectively (i.e., not able to fixate or hold objects), some goals might be very difficult to achieve. For example, when the goal of an adolescent with no forearm supination is to hold a plate with drinks and to shake hands, the patient should refine his/her goals and expectations with the help of the surgical team.

The same core set (COPM, AHA, ABILHAND, BBT, SHUEE-DPA) can be used to guide postsurgical treatment. Activity-based therapy should be used to improve hand function and the performance of functional hand activities after surgery, and may include intensive therapy, modified constraint-induced movement therapy, or bimanual task-oriented therapy.

Implications for future research

Assisting Hand Assessment for Adolescents

In children with CP, several interventions have proven to be effective in improving hand use and performance. The next step would be to use the Ad-AHA when evaluating these interventions in adolescents and extend study participations to patients up to the age of 18 years. The continuous and large age range of the AHA 18-18 makes it possible to use the same scale to monitor the development or possible deterioration of hand use from early childhood to young adulthood. Future research should focus on possible changes in adolescents with CP, in order to improve interventions that can prevent a deterioration of hand skills with age and which can facilitate further refinement of acquired skills. Information about improvement or deterioration will also aid the professional when planning interventions to improve hand use and performance. Furthermore, studies of the long-term effect of interventions from childhood to adulthood, and of changes in hand function during the childhood-to-adolescence-to-adulthood transitions are needed. The logical next step would be to extend the AHA for use in adults aged 18 years and older, in order to evaluate the effects of interventions in adults with CP (Fig. 2).

In the study presented in chapter 6, 13% of the included patients were diagnosed with bilateral CP. Regrettably, the (Ad-)AHA could not be used in this group to evaluate the effect of upper extremity surgery on (bimanual) arm-hand performance because the instrument is not valid and reliable for use with children and adolescents with bilateral CP. This is particularly unfortunate because more than 60% of children with bilateral CP have decreased hand function. In the past, there was a lack of appropriate outcome measures for different ICF levels validated for use in children with bilateral CP. Recently, the Both Hands Assessment (BoHA) was developed by adapting the AHA. New evaluation studies should use the BoHA to measure bimanual performance in children with bilateral CP and to quantify a possible asymmetry between hands. Besides the Kid’s-AHA, Ad-AHA, and BoHA,
more versions have been developed within the “AHA family”, based on age groups and diagnoses (bilateral and unilateral CP). (Fig. 3)

In order to evaluate hand use and performance in the entire CP population, it is necessary to further extend the AHA and to investigate the possibility of using the same AHA scale for a broader age range. The principal researchers involved in the development of the AHA family joined in the recently set up “AHA research network” to continue these studies, to collaborate with other investigators, and to contribute to future AHA research (www.ahanetwork.se). The ultimate goal is to develop an AHA for patients with unilateral and bilateral CP who are able to handle objects, although with varying difficulty (Manual Ability Classification System (MACS) levels I-III), in order to improve intervention planning, evaluate (new) interventions, determine their effectiveness, and monitor the development of hand use.

![Small-Kids AHA](18 months to 5 years) ![School-Kids AHA](6 to 12 years) ![Ad-AHA](13 to 18 years)

**Figure 2.** The AHA 18-18

**Intervention studies that aim to optimize hand use and performance of activities**

Future studies should evaluate the effect of a functional hand orthosis, using appropriate and valid outcome measures, in order to assess benefits (more effective use of the assisting hand) as well as potential harms (such as muscle atrophy).

Furthermore, there is a need for studies evaluating the immediate and long term effect of a hand orthosis in different age groups (including adolescents). The findings of various studies suggest that a functional hand orthosis may be effective for performing certain daily tasks, which highlights the need for research into the effect of intermittent use of a functional hand orthosis in combination with training.

There is still insufficient evidence to recommend upper extremity surgery as a treatment option for improving the hand use and performance of children and adolescents with CP. To make clinical recommendations, future research should include patients of different ages and ability. In order to predict and achieve optimal outcomes after upper extremity surgery that match patient-specific goals and expectations related to the performance of activities important to the patient and to define patient selection criteria (based on e.g. age, severity), larger studies are needed, preferably RCTs comparing the effects of surgical and non-surgical intervention, possibly with a waiting-list control group and monitoring into adulthood.

Besides the choice of surgical intervention, the postoperative hand rehabilitation programme has a major impact on hand use and performance. More research is needed into
the effect of a postoperative hand therapy. Such therapy should be activity-based with a focus on patient-relevant goals in order to optimize the outcome of upper extremity surgery.

Lastly, in order to evaluate the cosmetic results of upper extremity surgery, outcome measures evaluating the psychosocial impact of a hand deformity and cosmetic appearance should be developed and validated.

The findings of this thesis have contributed to knowledge relevant to the assessment and management of upper extremity functioning in children and adolescents with CP. The newly developed and validated instrument, Ad-AHA, makes it possible to assess the actual performance of adolescents with CP, which in turn makes it possible to evaluate the effect of interventions. The continuous and large age range of the AHA 18-18 will enable investigators to monitor the development of, or changes in, hand use from childhood to young adulthood. We found that the results achieved with functional hand orthosis and upper extremity surgery were promising in terms of the improvement in hand use and the performance of activities of a selected group of patients with CP.

Figure 3. AHA family
Chapter 7

References


