FUNCTIONAL STATUS OF CHILDREN WITH A CONGENITAL UPPER LIMB REDUCTION DEFICIENCY

Laurien M. Buffart, Josemiek M.F.B. Pesch-Batenburg, Vera G. van Heijningen, Marij E. Roebroeck, Henk J. Stam

Erasmus MC, University Medical Centre Rotterdam, the Netherlands.

INTRODUCTION

Congenital reduction deficiencies of the upper limb are rare. Nearly 50% of children with unilateral transverse upper limb reduction deficiencies (ULRD) wear a prosthetic device to enhance the ability to perform functional activities. Unfortunately, empirical evidence as to whether prostheses yield improved functional outcomes in these children is lacking. Assessment of arm and prosthetic functioning mostly relies on clinical observation of task performance. The purpose of this study was to assess the functional status of children with ULRD as measured by these standardized instruments.

To adequately measure arm and prosthetic functioning in children with ULRD, standardized measures at activity level are required. The difference between what a child "can do" in a clinical setting and "does do" in daily life is well known, also recognized as capacity and performance of activities [1]. Therefore both aspects should be measured. Capacity can be measured with functional tests and to measure performance of activities, assessment of spontaneous arm use or self-reported or parent-reported questionnaires are options.

Several instruments have been developed to measure arm function in children with various kinds of diagnoses. For adequate measurements of arm or prosthetic function functional tests and questionnaires have to be attractive for children, they have to assess bimanual activities of daily living and measure quality of movement and/or difficulty in performing activities [2]. The Assisting Hand Assessment (AHA) [3], the Unilateral Below Elbow Test (UBET) [4, 5], the Prosthetic Upper extremity Functional Index (PUFI) [6, 7] and the ABILHAND-Kids [8] questionnaires met these criteria.

METHOD

20 children with ULRD between 4 and 12 years old (mean: 8.7 ± 2.9 years) participated in the study. One child had below shoulder reduction deficiency, 16 below elbow and 3 had a partial hand. Eight children used a myoelectric device, one child used a passive device and eleven children did not use a prosthetic device (non-users).

Each child performed the UBET and an explorative assessment using the AHA. Parents and all children of 8 years and older were asked to fill out the PUFI and ABILHAND-Kids. For better comparison between instruments, results of sum scores on tests and questionnaires are presented as percentage of the total possible score. Sum scores of users with prostheses were compared to sum scores of non-users (without prostheses) with an independent t-test.

Instruments

Assisting Hand Assessment (AHA)

The AHA [5] was developed and tested for children who have one well functioning but one dysfunctioning hand (cerebral palsy and obstetric brachial plexus palsy). The test evaluates performance of arm/hand function as an assisting hand, since the affected hand primarily serves as an assisting hand rather than a non-dominant hand. Scoring of the quality of movement of 22 items is completed from the video using a 4-point scale from "effective (=

4)" to "does not do (= 1)". In the AHA version for children with ULRD, the item "moves fingers" is left out. Sum scores range from 0 to 100.

Unilateral Below Elbow Test (UBET)
The UBET [6, 7] was developed to measure arm or prosthetic functioning in children with ULRD 2 to 21 years old. The test has 4 versions for different ages, each consisting of 9 bimanual activities. Method of arm or prosthetic use is scored on a nominal scale describing 4 methods of grasp and stabilisation. The completion of task is scored on a 5-point scale from "no difficulty (= 4)" to "unable to complete the task (= 0)". Sum scores range from 0 to 36.

Prosthetic Upper extremity Functional Index (PUFI)
The PUFI [8, 9] evaluates the extent to which a child actually uses the prosthetic limb for daily activities, the comparative ease of task performance with and without the prosthesis and its perceived usefulness, which are respectively scored on a 6-point nominal scale, 5-point ordinal scale and 3-point ordinal scale. Higher scores represent less difficulty in performance and higher usefulness of the prostheses. Sum scores range from 0 to 100.

ABILHAND-Kids
The ABILHAND-Kids [10] was developed to measure the ability of children with cerebral palsy to user their hands in daily living. This questionnaire consists of 21 bimanual activities of daily life. The perceived difficulty in performing activities irrespective of the limb(s) actually used to perform the activity is rated on a 3-point scale ranging from "impossible (= 0)" to "easy (= 2)". Raw sum scores range from 0 to 42.

RESULTS
All 20 children and their parents completed the tests and questionnaires, except for one prosthetic user who did not complete the AHA without prostheses and one prosthetic user did not complete the AHA with prostheses. Since no differences were found between parent reported and child reported versions of the PUFI and ABILHAND-Kids, only data of parent-reported questionnaires are presented.

Method of performance
The way children can and do use their prosthesis or residual limb in performing activities is respectively scored in the UBET and PUFI (table 1).

Can do (UBET)				Does do (PUFI)			
Method of use prosthesis/arm	Users	Non-users		Method of performance	Users	Non-users	
Actively	30 ± 28	55 ± 30		Prosthesis actively	15 ± 18	0 ± 0	
Passively	38 ± 25	17 ± 13		Prosthesis passively	15 ± 21	0 ± 0	
Elbow/trunk	11 ± 14	27 ± 27		Residual limb	41 ± 34	85 ± 12	
One-handed	21 ± 13	*	1 ± 3	One-handed	23 ± 18	*	5 ± 6
				Some help	2 ± 4	3 ± 3	
				Cannot do	4 ± 4	7 ± 9	

Table 1. Method of use of prosthesis or residual limb of UBET and PUFI. Mean percentage of activities ± standard deviation. (* = Significant differences between users and non-users)

Results of the PUFI show that users and non-users performed respectively 94% and 90% of the activities independently. Results show a difference between how many activities a child *can* perform with the prosthesis (68%) and how many activities a child actually *does* with the

prostheses (30%). Both the UBET and the PUFI show that users perform more activities one-handed compared to nonusers (p=0.01 and p=0.00 respectively).

Difficulty and effectiveness of performance
Sum-scores of tests and questionnaires regarding effectiveness or ease of performance are presented in figure 1. Results show that children with ULRD perform well on daily activities.
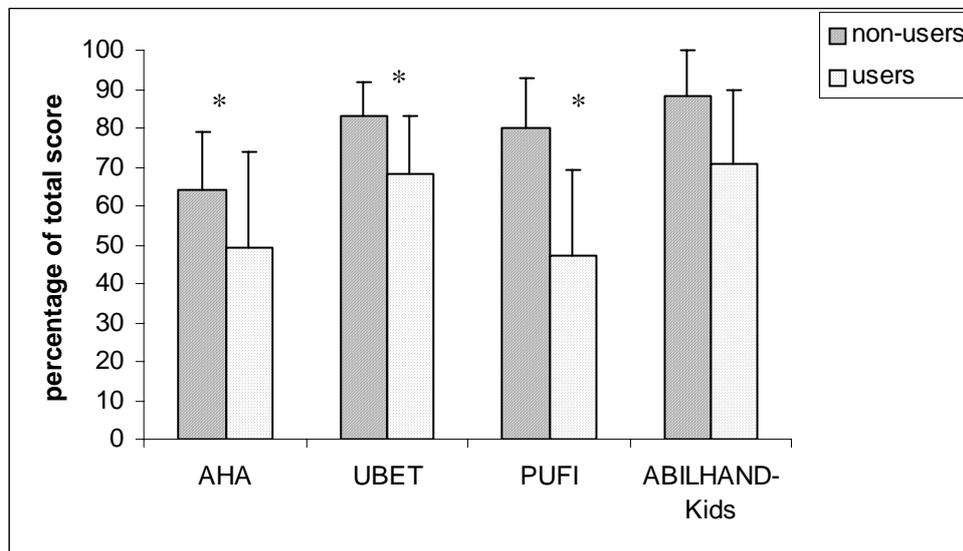

Figure 1. Sum scores of tests and questionnaires expressed as percentage of total score.

AHA results show that non-users used their arm more effectively compared to users (p=0.03). Also, non-users had less difficulty in performance of activities compared to users performing activities with prosthesis (p=0.02 UBET; p=0.00 PUFI). However, when we only take the activities into account in which the prosthesis is actually used (30% of the activities, see table 1), children perform these activities easily (sum score: 86.4 ± 9.8). Results of the ABILHAND-Kids did not show differences between users and non-users.

Usefulness of prosthesis
On average, users found the prosthesis useful in 34% of all activities as reported by the PUFI. Most activities of daily living, such as dressing tasks, are usually performed without prosthesis. Children use their prosthesis for specific activities such as riding a bicycle, playing with a beach ball and using scissors. For these activities, children find their prosthesis very useful (sum score: 79 ± 11).

CONCLUSIONS
Children with ULRD perform well on daily activities. 45% of the children wore prostheses and only 3 children wore the prosthesis more than 6 hours a day during the week. Children with ULRD find their prosthesis very useful in specific activities and can perform these activities very easily with prosthesis. Thus prosthetic devices have additional value in children with ULRD in specific activities rather than in activities of daily living in general.

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