Use and functioning of the affected limb in children with unilateral congenital below-elbow deficiency during infancy and preschool age: A longitudinal observational multiple case study

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1. Introduction

Children with unilateral congenital below-elbow deficiency (UCBED) lack a part of an arm or hand. Consequently they miss a part of typical hand function. In many cases children with UCBED are fitted with a prosthesis to replace the missing part of the limb in an attempt to improve functioning in daily activities. Yet, evidence about the functional gain of a prosthesis is conflicting [1–3]. In addition, surprisingly little reduction in performance of daily activities has been reported in children with UCBED, which contrasts with the limited function found in adults who have lost a part of their arm [4]. However, little is known about the way children with UCBED use their arms and which strategies they develop during early life to achieve adequate functioning in daily activities.

Typical motor development is characterized by variation and the development of adaptive variability [5]. Variation denotes the presence of a repertoire of strategies for a certain function. Variability is the ability to find the best strategy for a given task in a certain environment, i.e., the ability to produce adaptive motor behaviour. This ability develops through active trial and error activity and its associated multimodal sensory feedback. Reduced variation is associated with an early lesion of the brain; limited variability is considered to reflect minor or major brain dysfunction interfering with sensorimotor processing [5]. Children with UCBED have an altered repertoire of upper extremity movements which – given the anatomical restriction – may, or may not, have an appropriate size. In addition, upper extremity motility of children with UCBED may be atypical but well adapted to the situation. We wondered whether the different distal configuration of the upper limb in children with UCBED would interfere with the process of finding the best strategy in each situation, in particular as a recent study...
indicated that children with UCBED may show impaired adaptation of motor behaviour [6].

We therefore started a longitudinal observational multiple case study in order to explore how young children with UCBED use the affected limb during a standardized play session. During play we assessed the following behaviours of the affected limb: achievements (manipulation and holding of objects, use during crawling and standing up), variation and variability, and sensory exploration. Activities of daily life were assessed with the Pediatric Evaluation of Disability Inventory. In addition we evaluated neurological condition, as the presence of minor neurological dysfunction is associated with an impaired capacity for adaptive motor behaviour [6].

2. Material and methods

2.1. Design, participants and procedures

The study was designed as a longitudinal observational multiple case study. Four children with UCBED were included following a visit at the outpatient clinic of the Center for Rehabilitation Medicine of the University Medical Center Groningen in The Netherlands. Children with a deficiency of the upper arm or distal to the wrist and those who had co-morbidities interfering with the execution of the study were excluded. Parents provided informed consent for participation.

General characteristics like sex, age at first visit, level of deficiency and whether a prosthesis was prescribed or not were collected from medical charts. Every 6–8 months the children were invited for an assessment at the University Medical Center Groningen. Each assessment consisted of a standardized play session and a neurological examination. Once a year the Pediatric Evaluation of Disability Inventory—Dutch Version (PEDI-NL) was performed to evaluate functioning in daily activities. The total follow-up time was 13–49 months per child.

2.2. Assessment of the use of affected arm during a standardized play session

Each play session lasted 20 to 30 min during which the child was allowed to play with a variety of toys, such as cars, balls, puppets and toy carpentry tools. The children who had been fitted with a prosthesis played half of the session with the prosthesis, the other half without. Each play session was video-taped. On the basis of the video-recording the following behaviours of the affected limb were assessed: a) achievements: manipulation and holding of objects, use during crawling and standing up, b) variation and variability, and c) sensory exploration. Appendix A provides details of the video-assessment. The video recordings were analysed by two assessors (RvdB and MHA).

2.3. Neurological examination

At the end of each video session the child was neurologically assessed by MHA with an age specific technique for the evaluation of MND [7–9]. Two forms of MND can be distinguished: simple and complex MND. Simple MND can be regarded as a sign of a normal, but sub-optimally developing brain. Complex MND is the form which is strongly related to prenatal and perinatal adversities; moreover this form is clearly associated with developmental co-ordination disorder (DCD) and behavioural problems. The age specific assessments of the neurological examination have a satisfactory reliability ($\kappa = 0.71$–0.83) and validity [7,10,11].

2.4. PEDI

To evaluate the child’s independence in daily skills the Pediatric Evaluation of Disability Inventory — Dutch Version was performed annually [12]. The PEDI is an instrument designed to measure the functional status of children of 6 months to 7.5 years of age. In a structured interview, the parents are asked to provide information on their child’s functional capability and assistance given by care givers in three domains: selfcare (74 items), mobility (65 items) and social functioning (66 items). In the PEDI-NL one point is given when the child is able to perform the activity and no point when the child is unable to perform the activity. The amount of assistance given by caregivers is reflected in a 6-point Likert scale in which 0 is totally dependent of help and 5 is totally independent. Finally, the sum score of the activities per domain (functional capability and assistance by care givers) can be transferred into normative scores and scale scores using a table. In the normative scores the individual test results per domain are compared to those of healthy children of the same age. The normative score is indicated as normal between the 30th and 70th percentile. In the scale scores the functional attainment of the child in time and an estimation of the functional capacities of the child in the continuum of items of which the scale is composed from 0 (none of the items can be performed) to 100 (all items can be performed) can be seen. In the PEDI-NL the ability to accomplish the activity is evaluated, regardless of whether or how the arms and hands are used. The discriminative validity of the PEDI-NL between children with and without disability, the inter-interviewer reliability (ICC 0.99), test–retest reliability (ICC 0.91–0.98) and inter-respondent reliability (ICC 0.91–0.99) are good [13,14].

2.5. Data analysis

The behavioural scores of the affected arm during standardized play over time of the four children were summarized in graphs, which were analysed visually. The changes in PEDI scores over time were plotted in relation to the norms.

3. Results

The children were assessed two to nine times during infancy and preschool age (median value 6.5 times). The age at the first assessment varied from 3 to 16 months. Clinical information on the four children is provided in Table 1. Two children had been fitted with a prosthesis shortly before the emergence of crawling behaviour.

3.1. Use of the affected arm during the play session

Achievement without prosthesis: All children frequently used the affected arm to manipulate toys (Fig. 1A), to hold objects between the affected arm and trunk (Fig. 1B) and – at the appropriate developmental stage – for crawling. Less consistent behaviour was observed in holding objects with the affected arm, for instance in the fold of the elbow. Only child 1 showed this behaviour frequently once she had developed this strategy, while in the other children the behaviour was observed to a varying degree across time (Fig. 1C). When the children developed the ability to stand up, three children (children 2, 3 and 4) consistently used the affected arm to get up from the floor, while the remaining child (1) initially used the affected arm only occasionally to get up. At later ages this child did use her affected arm in standing up. After the age of 34 months none of the children used their arms to get up from the floor.

Achievement with prosthesis: Achievements of the affected arm of the two children who had been fitted a prosthesis (children 1 and 2) varied considerably over time when they were wearing their prosthesis. This inconsistent behaviour differed from the consistent behaviour of all four children without prosthesis. With prosthesis the arm was sometimes used for manipulation and holding an object, but at other occasions the arm with prosthesis was more or less neglected (Fig. 1D, E, F). Both children used their arm as a support during crawling with prosthesis. Until the age of 20 months both children used their affected arm with prosthesis more often during getting up from the floor than without prosthesis. At later age this difference disappeared as the children no longer used the arms to get up from the floor.
Variation and variability: Without prosthesis, children 1, 3 and 4 in general showed an appropriate repertoire of strategies with adaptive behaviour, while child 2 showed a limited repertoire which he used in the most appropriate way. When the two children who had a prosthesis wore the device, they used few strategies with inconsistent efficiency, i.e. they exhibited reduced variation and reduced variability (Fig. 2A and B).

Sensory exploration: Without prosthesis, all children were frequently observed to explore objects with the stump circumference or the stump end point of their affected arm. The two children with a prosthesis showed less exploration when wearing the prosthesis than without the prosthesis, and this varied over time (Fig. 3). While wearing the prosthesis they used the hand and the shaft of the prosthesis to explore objects.

In general, the children enjoyed the play session. However, we noted that from the age of 24 months child 1 got moody and rather reluctant to play when she had to wear her prosthesis.

3.2. Neurological condition and PEDI

The neurological condition of child 1 was normal, child 3 showed simple MND and children 2 and 4 showed complex MND (Table 1).

The PEDI-NL indicated that all children showed typical functional capabilities in the domains selfcare, mobility and social functioning. Results of selfcare are given as an example in Fig. 4A. Capacities in all domains increased with increasing age (Fig. 4B). The four children with UCBED required the same amount of assistance by care givers as children of the norm population.

Table 1

<table>
<thead>
<tr>
<th>Child</th>
<th>Sex</th>
<th>Level of deficiency</th>
<th>Affected side</th>
<th>Age at first assessment (months)</th>
<th>Prosthesis</th>
<th>Age at first prosthesis (months)</th>
<th>Number of observations</th>
<th>Neurological condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>P</td>
<td>L</td>
<td>3</td>
<td>Y</td>
<td>9</td>
<td>9</td>
<td>Consistently normal</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>P</td>
<td>R</td>
<td>3</td>
<td>Y</td>
<td>6</td>
<td>6</td>
<td>Consistently complex MND</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>W</td>
<td>L</td>
<td>4</td>
<td>N</td>
<td>–</td>
<td>7</td>
<td>Consistently simple MND</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>W</td>
<td>L</td>
<td>16</td>
<td>N</td>
<td>–</td>
<td>2</td>
<td>Consistently complex MND</td>
</tr>
</tbody>
</table>

F: female; M: male; L: left; R: right; Y: yes; N: no.
P: proximal one third below elbow; W: wrist.
MND: minor neurological dysfunction.

Fig. 1. Individual developmental trajectories of achievements of the affected arm of the four children. The upper panels (A, B and C) display achievements without prosthesis (4 children), while the lower panels (D, E and F) achievements of two children while wearing a prosthesis. The left hand panels (A and D) show observed manipulation with the affected limb (0 = not, 1 = occasionally, 2 = frequently observed). The middle panels (B and E) display information on the frequency with which holding of an object between limb and trunk was observed (0 = not, 1 = occasionally, 2 = frequently observed). The right hand panels (C and F) provide information on the frequency with which the child held an object with the affected limb (0 = not, 1 = occasionally, 2 = frequently observed). For details see Appendix A.
Variation and variability

A. without prosthesis

B. with prosthesis

Fig. 2. Individual developmental trajectories of variation and variability of the affected arm. 0 = limited variation and limited variability, 1 = limited variation and appropriate variability, 2 = appropriate variation and appropriate variability. Panel A displays the trajectories of the four children without prosthesis. Most children in general showed appropriate variation and appropriate variability, but only child 2 showed all but once a limited variation with appropriate variability. Panel B displays the trajectories of the two children who had a prosthesis while they wore the device. They virtually always showed limited variation with or without reduced variability.

4. Discussion

Many studies have been performed to evaluate functioning in daily life of children with UCBED with or without prosthesis [1,2,6,15–18]. However, our study is the first with a longitudinal and observational design starting in infancy.

The play videos showed that the young children with UCBED used the affected limb without prosthesis frequently for manipulation. From the age of one year, they also frequently exploited the affected limb for holding objects, often by holding the object between the arm and trunk, and less frequently by holding it with the affected limb only. In addition they frequently used the stump for object exploration. Two children had been fitted with a prosthesis. They effectively used the prosthesis during the second half of the first year for support during crawling and standing up. But throughout infancy and preschool age they used the affected limb with prosthesis less often for manipulation, holding an object and sensory exploration than when not wearing a prosthesis. In child 1, who was the only girl and the only child without minor neurological dysfunction, the reduced use of the affected limb when she wore the prosthesis was accompanied by moody behaviour; she became reluctant to play. As soon as the arm was free from prosthesis she started to play enthusiastically. Nevertheless, in the last video observation, when she was almost five years old, a more functional use of the prosthesis was observed. This may be a chance observation, but it may also be that functional and social gains of a prosthesis during daily activities emerge after the age of four years. Longer follow-up measurements are needed to confirm this finding.

The play videos also showed that the children with UCBED without a prosthesis mostly showed an appropriate repertoire of movement strategies of the affected limb, provided that the anatomical limitations are taken into account. In addition, they mostly showed appropriate variability, i.e., they were able to adapt their movements to the specifics of their self selected playing tasks. It showed that the minor neurological dysfunction, which was present in three of four children, did not interfere with these unchallenging activities. The presence of the prosthesis, however, resulted in a reduction of the number of movement strategies of the affected arm and a reduction of adaptive behaviour. This was true for the child without minor neurological dysfunction (number 1) and the child with minor neurological dysfunction (number 2). The less optimal motor behaviour during upper arm activities in the presence of the prosthesis may be an expression of a lack of functional possibilities of the prosthesis. In this respect our data on object exploration are also interesting. We observed that without prosthesis all children explored objects with the stump circumference or the stump end point of their affected arm. Johnson et al. [19] reported that children with UCBED have a better touch pressure sensation at the stump circumference of the affected arm than in homologous parts or in the hand of the contralateral arm. Our data suggest that children with UCBED indeed use the good sensory abilities of the stump for object exploration and that the presence of a prosthesis partially may interfere with this activity.

Exploration

A. Without prosthesis

B. With prosthesis

Fig. 3. Individual developmental trajectories of sensory exploration with the affected arm (0 = not, 1 = occasionally, 2 = frequently observed). Panel A displays the trajectories of the four children without prosthesis. The children virtually always showed frequently sensory exploration. Panel B displays the trajectories of the two children who had a prosthesis while they wore the device. The trajectories reveal less exploration in the presence of the prosthesis.
Our PEDI data show that children with UCBED perform daily activities within age specific norms. They apparently do so despite the presence of UCBED and the presence of a non-optimal neurological condition. This underlines the creativity of children to cope with daily life activities as can be seen in the study of Beckung et al. [20] in which a substantial part of children with mild cerebral palsy experience little impairment in daily activities. It also shows the value of the short limb as an exploratory, manipulatory and fixing tool. Scoring within the norms of the PEDI does however not preclude the presence of more specific problems in daily life activities, for instance activities evaluated with the Unilateral Below Elbow Test (UBET) or the Prosthetic Upper extremity Functional Index (PUFI) [16,21]. The first test may be more suitable to evaluate the extent to which a child actually uses an affected limb for daily activities.

A limitation of this study is the small number of participants. Therefore, a larger study is needed to generalize the findings to all children with UCBED. The strengths of the study are the longitudinal design from infancy onwards and the application of a systematic analysis of spontaneous behaviour with and without prosthesis [22].

In conclusion it can be said that throughout infancy and preschool age, children with UCBED use their affected arm frequently during spontaneous play. The short limb is used to explore and manipulate objects and as such, the affected limb is a valuable tool, which may explain the fact that these children perform activities of daily life within the normal range. Prosthetic use seemed to be associated with less exploratory and less adaptive motor behaviour of the affected arm. The presence of minor neurological dysfunction in children with UCBED deserves further attention.

Conflict of interest

None declared.

Appendix A. Definition of the observed activities and description of the scoring system of the video observations.

<table>
<thead>
<tr>
<th>Activity</th>
<th>Definition</th>
<th>Score</th>
<th>Description of score</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Motor skills</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Manipulation</td>
<td>Changing the position of an object with the affected limb or with both upper limbs</td>
<td>2</td>
<td>The child frequently uses the affected limb during object manipulation.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally uses the affected limb during object manipulation.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not use the affected limb during object manipulation.</td>
</tr>
<tr>
<td>Holding an object between limb and trunk</td>
<td>The object is placed between the affected limb and trunk to stabilize it and hold it in place</td>
<td>2</td>
<td>The child frequently holds an object between limb and trunk.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally holds an object between limb and trunk.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not hold an object between limb and trunk.</td>
</tr>
<tr>
<td>Holding an object with the affected limb in the fold of the elbow or in the prosthetic hand of the affected limb</td>
<td>The object is stabilized and held in place in the fold of the elbow or in the prosthetic hand of the affected limb</td>
<td>2</td>
<td>The child frequently holds an object in the fold of the elbow or in the prosthetic hand.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally holds an object in the fold of the elbow or in the prosthetic hand.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not hold an object with the affected arm.</td>
</tr>
<tr>
<td>Use of limb during crawling</td>
<td>The affected limb is used to support body weight and propulsion</td>
<td>2</td>
<td>The child frequently uses the affected limb for body support and propulsion.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally uses the affected limb for body support and propulsion.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not use the affected limb for body support and propulsion.</td>
</tr>
<tr>
<td>Use of limb during standing up from the floor</td>
<td>The affected limb is used for support during standing up from the floor, e.g. by holding on to or leaning on a piece of furniture</td>
<td>2</td>
<td>The child frequently uses the affected limb during standing up.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally uses the affected limb during standing up.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not use the affected limb during standing up.</td>
</tr>
<tr>
<td>Variation and variability</td>
<td>The affected limb has a repertoire of strategies and is used in the most effective way</td>
<td>2</td>
<td>The child uses the affected limb in the most effective way; the limb exhibits a typical repertoire of strategies.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child uses the affected limb in the most effective way; the limb exhibits a reduced repertoire of strategies.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not use the affected limb in the most effective way; the limb exhibits a reduced repertoire of strategies.</td>
</tr>
<tr>
<td><strong>Sensory behaviour</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Exploration</td>
<td>Touching and gently rubbing the outer surface of objects with the affected limb (whether or not a prosthesis is present)</td>
<td>2</td>
<td>The child frequently touches, feels and explores the surface and shape of an object.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1</td>
<td>The child occasionally touches, feels and explores the surface and shape of an object.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child does not touch, feel or explore the surface and shape of an object.</td>
</tr>
<tr>
<td>Affection: Mood</td>
<td>The child wants to play when invited</td>
<td>2</td>
<td>The child is eager to play.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>0</td>
<td>The child is reluctant to play.</td>
</tr>
</tbody>
</table>
References


