

COMPREHENSIVE APPROACH IN HABILITATION OF CHILDREN WITH CONGENITAL BILATERAL DEFICIENCY OF UPPER LIMBS - A CASE REPORT

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Abstract

The incidence of congenital upper-limb deficiencies is very low. Children with upper-limb deficiencies must be referred to a multidisciplinary clinic early in life in order to achieve good functional independence. Management of children with bilateral limb deficiency differs from those with unilateral deficiency. Our aim

is to present a comprehensive approach in habilitation of a child with congenital bilateral deficiency of upper limbs. We believe that this boy was fitted early enough to establish good wearing patterns. We also believe that early fitting may prevent secondary physical disabilities during growth, such as scoliosis, and will also enhance bilateral functional skills.

INTRODUCTION

The incidence of congenital upper-limb deficiencies for United States, Canada and the UK is estimated to be 1:9400 live births (1). An 11-year total population study of Western Australia, that examined the prevalence and epidemiology of congenital upper limb anomalies, found the prevalence of babies born with upper limb anomalies to be 1 in 506 (2). Bilateral upper limb deficiency is very rare condition. By our knowledge there are no precise data on incidence available. In last eight years there were two such cases in Slovenia, one of them is going to be presented in this article.

To the knowledge of a nowadays, children with limb deficiencies must be referred to a multidisciplinary clinic within the first four months of life. The aim is a detailed assessment, evaluation and early fitting of prosthesis for unilateral upper limb deficiency (3). Kuyper et al. are reporting that only 3% of children with bilateral upper limb deficiency are fitted with prosthesis (4). "De Hoogstraat" rehabilitation centre uses a restrained prosthesis prescription policy, depending on the type of deficiency and the expected functional benefits.

An early fitting of unilateral limb deficiency is important for amputations as well as for congenital malformations (4). In the unilateral upper-limb amputee it allows the opportunity to develop bimanual skills into the body image and into useful prehensile activities at an earlier age. Early fitting may also contribute ultimately to better prosthetic tolerance and wearing patterns and may prevent an asymmetrical posture and spinal curvature (5-12).

On the other hand, management of bilateral upper limb deficiency is usually different. Lento describes infants and children with congenital bilateral upper-limb deficiencies, who generally develop a remarkable ability to adapt to their situation. They do not have a distorted sense of body form that may occur if the limb or limbs were amputated later in life. Therefore, many may not feel the need for any type of prosthesis even for cosmetic reasons. Many prefer to rely on their lower limbs in place of an upper limb(s) to perform typical activities of daily living such as bathing, dressing and feeding (12).

But there might be also some long-term problems of lower limbs use. A retrospective study performed to determine the incidence of spinal abnormalities in patients with skeletal defects of the upper extremities showed that there was a 100% incidence of scoliosis among patients with bilateral amelia. The results of bracing were poor, due to patient rejection of the brace (13).

We believe also that using of feet might not be always a socially accepted choice in different settings.

Aim: We want to present a comprehensive approach in habilitation of a child with congenital bilateral deficiency of upper limbs.

METHODS AND SUBJECTS

A boy was born after uneventful pregnancy, 8 days before the term, with bilateral amelia of upper limbs. Radiologically there were a short humerus, ulna, radius and 2 metacarpal

bones present on the left side and aplasia of humerus, short radius and ulna, two metacarpal bones and phalangeal bones for two fingers on the right side.

He was referred to our Institute at the age of 8 months. A team of specialist of physical and rehabilitation medicine, occupational therapist and prosthetic engineer conducted a detailed functional assessment. Clinically there were two short fingers on the right side coming out of shoulder, and a 10 cm long arm with two longer fingers on the left side. Neurologically there were no signs of abnormality. Motor development was slightly delayed: he just started to roll from prone to supine, was able to transfer to sitting position with help and needed some support to maintain sitting position. To support the motor development he was referred to physiotherapy. He was encouraged to use his feet in different activities.

At the age of 15 months he was still not able to transfer independently to sitting and standing position. He was using his feet to handle different objects. Four months later he was able to walk independently, but was dependant in most of daily life activities. Some adjusted eating tools for left upper limb fingers were provided.

At the age of three and half years first prosthesis was prescribed. He got a body-powered prosthesis for exarticulation of right upper limb, with Omni wrist for shoulder joint, orthotic elbow and voluntary closing children's hook (a crocodile) for some other activities. For most of daily life activities he was using his feet or he got help from parents or accompanying person. He was able to hold spoon with two longer fingers on his left side, but had problems when feeding himself. He was also able to hold a pencil, but not firmly enough to draw. At the same time he was able to draw and play very successfully by using his feet.

We decided to refer him into a two weeks in-hospital training program for training the use of different assistive devices, slowly strengthening the grip on left side and use of prosthesis on right side. At the time of precise evaluation we tried to find an important goal with the boy, to motivate him for prosthetic wearing. We found out he was very keen on playing some computer games, but was having problems, since he could use just left upper arm fingers. We prepared some adjusted tool to hold it with prosthesis while playing. We also organized a meeting with his teachers from a kindergarten, so we could transfer our knowledge and help them to provide a stimulating environment for a boy.

RESULTS

At the beginning he used prosthesis only for bike riding and occasionally for some other activities, such as carrying basket during picking up chestnut. For most of daily life

activities he was using his feet or he got help from parents or accompanying person.

He was able to hold spoon with two longer fingers on his left side, but had problems when feeding himself. He was also able to hold a pencil, but not firmly enough to draw. At the same time he was able to draw and play very successfully by using his feet. We found out he was very keen on playing some computer games, but was having problems, since he could use just left upper arm fingers. We prepared some adjusted tool to hold it with prosthesis while playing.

We organized also a meeting with his teachers from a kindergarten, so we could transfer our knowledge and help them to provide a stimulating environment for a boy.

At the age of four years he is able to hold all different objects, which are not too big or heavy, with fingers on the left side. He gained some muscular power and dexterity. He is able to eat and draw by using adjusted tools for holding spoon and pencil. He is also able to use some adjusted tool for his right side to type on a computer. He is able to ride a bicycle by using prosthesis. He is also motivated to use it during some other activities, but predominantly while playing on a computer. He still uses his feet for some activities. His spine is developing without any serious pathology.

Functional skills level evaluated by Pediatric Evaluation of Disability Inventory (PEDI, 14) are comparable to normal development in the mobility and social skills domains. He is having lower functional level score in the self-care domain. Results are on 14,8 percentile. Similar are results in caregiver assistance scales.

DISCUSSION

Our aim was to present a case of comprehensive approach in habilitation of children with congenital bilateral deficiency of upper limbs. We didn't follow the premises of early fitting, which are reported to contribute ultimately to better prosthetic tolerance and wearing patterns and may prevent an asymmetrical posture and spinal curvature (5-12). As Lento was describing infants and children with congenital bilateral upper-limb deficiencies, who generally develop a remarkable ability to adapt to their situation, we can confirm it stands also for a presented boy. He is a happy child, interested in many different activities and only motivated for a prosthesis wearing when it offers him a help in activities of his interest.

Due to a good mobility of both fingers on left side, we planned to improve their strength and dexterity and to use them as a dominant hand. Both fingers on the right side are much smaller, so we tried to fit prosthesis on a non-dominant side. To make prosthesis as light as possible we decided to use body-powered system. In one year he became less dependent

on his feet, more frequently he draw with hands and also use them better at some other activities, which are normally performed with hands.

If we are taking into account the goals that a school-aged child should achieve are (11), we can say that the development of functional activities of this boy are going quite well. He is able to hold objects with the prosthetic limb without breaking or dropping them, to operate the terminal device reliably, but still needs an assistance to don and doff the prosthesis and to dress. Parents are encouraging use of the prosthesis in some of activities.

We hope that he will gain further in the field of functional abilities. However we can not predict how it will be in the adolescence. Function and wear may occasionally be far less important than appearance in that time, especially to the adolescent with an upper-limb deficiency.

CONCLUSION

We believe that this boy was fitted early enough to establish good wearing patterns and he would not discard prostheses once he will be old enough to make his own decisions. We also believe that early fitting may prevent secondary physical disabilities during growth, such as scoliosis, and will also enhance bilateral functional skills.

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