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Impact of Prostheses on Function and Quality of Life for Children with Unilateral Congenital Below-the-Elbow Deficiency

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Investigation performed at the Shriners Hospital for Children, Sacramento, California

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Background: Children with unilateral congenital below-the-elbow deficiency present a dilemma to clinicians. Parents want the child to have a prosthesis and, because it seems that the deficiency will cause functional problems, one is customarily prescribed for infants. Use of the prosthesis is then encouraged throughout childhood. However, these children frequently abandon the prosthesis. There are no evidence-based guidelines regarding prescription of prostheses or standard methods for assessing use and function.

Methods: A multicenter outcomes study was done to assess the quality of life and function of 489 children with a unilateral congenital below-the-elbow deficiency; 321 wore a prosthesis, and 168 did not. The Unilateral Below-the-Elbow Test (UBET) was designed, validated, and administered to these children along with several outcomes measures, including the Pediatric Outcomes Data Collection Instrument (PODCI), the Pediatric Quality of Life Inventory (PedsQL), and the Prosthetic Upper Extremity Functional Index (PUFI).

Results: Use of a prosthesis was not associated with any clinically relevant differences in PODCI or PedsQL scores. Non-wearers performed either the same as or better than wearers on the UBET. When queried (with use of the PUFI) about performance of various tasks, non-wearers scored themselves higher than wearers. Children with a unilateral congenital below-the-elbow deficiency scored the same as or higher than the general population on the PedsQL. They scored significantly lower than the general population on the PODCI Upper Extremity Physical Function Domain and higher on the Happiness Domain, but the differences were small.

Conclusions: Prostheses may help with social acceptance or may be useful as tools for specialized activities, but they do not appear to improve function or quality of life, which are nearly normal for children with unilateral congenital below-the-elbow deficiency regardless of whether they wear a prosthesis. These findings call into question the standard practices of fitting infants with prostheses and encouraging young children to wear the prosthesis.

Level of Evidence: Therapeutic Level II. See Instructions to Authors for a complete description of levels of evidence.

Children with unilateral congenital below-the-elbow deficiency present a therapeutic dilemma to physicians, therapists, and prosthetists. Parents usually want a prosthesis for their child and, because it seems as though a child with a unilateral congenital below-the-elbow deficiency will have substantial functional problems, a prosthesis is customarily prescribed. Standard practice includes fitting the infant with a passive prosthesis when he or she is able to sit unsupported (at approximately six months of age) and supplying an active prosthesis when the child is between the ages of two and five years[1-7]. However, children and parents are frequently dissatisfied with the prosthesis, and many children abandon it when they are old enough to choose not to wear it[6,8,9]. There has been little research on prosthetic use by children with a unilateral congenital below-the-elbow deficiency, and there are no established guidelines regarding the prescription of prostheses or methods for assessing use and function by these children. Rejection of the prosthesis has been reported to be associated with child-related factors (a longer residual limb length, initial fitting after the age of two years, and entering into adolescence), parent-related factors (parental disappointment and limited parental involvement), and prosthesis-related factors (limited usefulness, discomfort, decreased sensation, and prosthetic weight)[6,8-12].

TABLE I Demographic Information for the Children and Adolescents with Unilateral Congenital Below-the-Elbow Deficiency

	Gender					Affected Side			
	Wearers		Non-Wearers			Non-Wearers		Wearers	
Age *(yr)*	Female	Male	Female	Male	Total	Left	Right	Left	Right
2-4	33	18	16	17	84	25	8	37	14
5-7	42	30	22	19	113	24	17	44	28
8-10	32	33	13	12	90	16	9	43	22
11-20	79	54	33	36	202	38	31	81	52
Total	186	135	84	84	489	103	65	205	116

According to an unpublished survey of Shriners Hospitals for Children limb-deficiency clinics performed in 1999, Shriners Hospitals for Children treat more than 2000 children with unilateral congenital below-the-elbow deficiency at twenty orthopaedic hospitals. Limb-deficiency teams at each hospital work together with children with unilateral congenital below-the-elbow deficiency and their parents to choose the best prosthesis for the child, but there are no evidence-based guidelines with which to make this determination. In order to provide evidence-based guidelines for the treatment of this population, ten Shriners Hospitals (in Erie, Greenville, Houston, Los Angeles, Montreal, Northern California, Philadelphia, Springfield, St. Louis, and Twin Cities) participated in a five-year multicenter cross-sectional study of the clinical outcomes for children with a unilateral congenital below-the-elbow deficiency; the parents of these children were included in this study. The major aim of the study was to determine whether use of a prosthesis by children with unilateral congenital below-the-elbow deficiency is associated with a better quality of life and better function.

A variety of generic and disease-specific measures have been developed in an attempt to address quality-of-life issues for children with a range of diagnoses. Some of these modalities are meant to measure health or functional status, while others focus on what has been termed *health-related quality of life*. Much research has also been targeted at the unique challenges of measuring pediatric and adolescent quality of life. Vitale et al. found that measures of adult health status were not valid when applied to adolescents with orthopaedic problems[13]. Roye et al. also highlighted the adaptive response of children to a chronic condition and their lack of knowledge of a different state of health, and how these factors may affect their quality of life[14].

The concept of function has been defined in many different ways. The International Classification of Functioning, Disability and Health is a conceptual framework that differentiates types of function and provides guidelines for their measurement[15]. According to this framework, unilateral congenital below-the-elbow deficiency is an impairment (problem with a body structure). As such, it may limit the child's activities (execution of tasks) and participation (involvement in a life situation). Ideally, tools that purport to measure function should address all levels of function (impairment, activity, and participation). In addition to administering questionnaires about function, it is also desirable to directly observe the activity component of function in order to eliminate the possibility of reporting bias.

In this paper, we present the results of parent and child-reported health-related quality-of-life and musculoskeletal health questionnaires as well as subjective and objective functional testing of children with unilateral congenital below-the-elbow deficiency with and without a prosthesis. In addition, the function and quality-of-life test results for the children with unilateral congenital below-the-elbow deficiency are compared with published norms for these test results in the general population.

Materials and Methods

The testing protocol for this study was established by the UCBED (Unilateral Congenital Below-the-Elbow Deficiency) Study Group (see Note at the end of the paper). Four hundred and eighty-nine children and adolescents, between the ages of two and twenty years, with unilateral congenital below-the-elbow deficiency were recruited at ten participating hospitals and tested under a protocol approved by eleven different institutional review boards. Of these 489 subjects, 55% (270) were female and 66% (321) wore a prosthesis; the left side was affected in 63% (308) of the participants (Table I).

The testing protocol included the administration of five tests (Table II). In addition to the English-language versions, all tests were translated into Spanish and French. All were converted to touch-screen computer format and were administered in a standardized fashion. There are different versions of each questionnaire, designed to test different ages on the basis of age-related cognition and abilities. We followed previously validated age-range recommendations for administration of each test. Since the different tests have different validated age-range recommendations, the age ranges for the reported results vary with the test. When available, both parent and child versions of the tests were given; we chose to analyze the parent version for children under the age of eleven to thirteen years (depending on the validated age-range recommendation of the test) and the child version for children over the age of eleven to thirteen years. Results of the demographic question-

TABLE II Tools Used in the Study

Tool	Measurement(s)	Administered to	
		Parent	Child
Demographic information	Standard demographic information	Yes	No
Pediatric Quality of Life Inventory, Generic Core Scales (PedsQL)	Health-related quality of life	Yes	Yes
Pediatric Outcomes Data Collection Instrument (PODCI)	Musculoskeletal health	Yes	Yes
Prosthetic Upper Extremity Functional Index (PUFI)	Usefulness of a prosthesis for performance of daily tasks by children with upper-limb deficiencies	Yes	Yes
Unilateral Below-the-Elbow Test (UBET)	Objective assessment of task performance by children with unilateral congenital below-the-elbow deficiency with and without prosthesis	No	Yes

naire, Pediatric Quality of Life Inventory (PedsQL) Generic Core Scales, Pediatric Outcomes Data Collection Instrument (PODCI), Prosthetic Upper Extremity Functional Index (PUFI), and Unilateral Below-the-Elbow Test (UBET) are presented here.

The PedsQL is a well-validated survey that asks twenty-three questions of both parents and children about various aspects of health-related quality of life over the past month. Published results are available for the general population[16]. The PedsQL scoring algorithm translates the available responses to questions ("never," "almost never," "sometimes," "often," or "almost always") into scores of 0%, 25%, 50%, 75%, and a maximum of 100% for each of four generic core scales (Physical Health, Emotional Functioning, Social Functioning, and School Functioning)[17]. The raw data used to obtain Emotional, Social, and School Functioning scores are averaged to obtain the Psychosocial Health Domain score, and the raw data used to obtain all four core-scale scores are averaged to obtain a Total Scale Score. Parents answer for children who are two to four years of age, and both the parents and the children answer for those who are five to twenty years of age. For this report, the parents' scores were analyzed for children under thirteen years of age, and the subjects' scores were analyzed for those between the ages of thirteen and twenty years of age.

The PODCI is a well-validated musculoskeletal health questionnaire, and published results of this measure are available for the general population and for a normal population[18-20]. PODCI questions address both the activity and the participation components of function. The PODCI contains six validated scales, or domains (Upper Extremity Physical Function; Mobility/Transfers; Sports/Physical Function; Pain/Comfort; Happiness; and Global Function, which is a combination of Upper Extremity Physical Function, Mobility/Transfers, and Sports/Physical Function), with a total of 108 questions. The maximum score in any domain is 100 points, and scores below the low 80s are considered to represent below-normal function[19]. Parents answer for children who are two to ten years of age, and children and parents answer for those who are eleven to twenty years of age. In the present study, parents' scores were analyzed for children under the age of eleven years, and subjects' scores were analyzed for those between the ages of eleven and twenty years.

The Prosthetic Upper Extremity Functional Index (PUFI) was designed to record whether children with upper-limb deficiency who wear a prosthesis accomplish specific daily activities, in what manner the prosthesis is used in the activities, the ease of use of the prosthesis, the ability to perform the activity without the prosthesis, and the perceived usefulness of the prosthesis[21,22]. The PUFI addresses the activity component of function. Parents answer for children of all ages, and children over the age of eight years also answer for themselves. The PUFI has two parts. Part I requests information about whether the prosthesis is useful for several different general categories of activities, and Part II asks five different questions about the usefulness of the prosthesis for each of twenty-six specific activities (for two to five-year-olds) or thirty-eight specific activities

TABLE III UBET Completion-of-Task Scores

Score *(points)*	
4	Completes task with no difficulty. Movements are quick, smooth. Stability is maintained throughout task.
3	Completes task with minimal (25%) difficulty. Movements are quick but slightly awkward. Stability is readily regained when lost.
2	Completes task with moderate (50%) difficulty. Movements are slower and awkward.
1	Completes task with maximal (75%) difficulty. Movements are very slow and awkward. Stability is frequently lost.
0	Unable to complete task.

TABLE IV PedsQL Respondents and Scores

	Wearers	Non-Wearers	P Value
Age *(no. of subjects)*			
2-4 yr	57	29	
5-7 yr	62	35	
8-12 yr	108	27	
13-20 yr	90	41	
Total	317	132	
Mean PedsQL score* *(points)*			
Physical Health Domain	88.5	88.6	0.94
Psychosocial Health Domain†	80.8	77.5	0.012
Emotional functioning	77.9	74.3	0.04
Social functioning	82.2	80.1	0.36
School functioning	55.1	46.7	<0.001
Total Scale Score‡	83.5	81.5	0.07

*According to the PedsQL scoring algorithm[17], because the Psychosocial Health Domain represents fifteen questions and the Physical Health Domain represents eight questions, and all scores are calculated from raw data, the Emotional, Social, and School Functioning scores cannot be averaged to obtain the Psychosocial Health Domain Score and the Physical and Psychosocial Domain scores cannot be averaged to obtain the Total Scale Score. †Average of raw data used to calculate the Emotional, Social and School Functioning scores. ‡Average of raw data used to calculate the Physical and Psychosocial Health Domain scores.

(for children six years of age and older) (see Appendix). The five Part-II questions are:

A. Does your child do the activity?

B. How does your child usually do the activity?

C. How well does your child do the activity using the prosthesis?

D. How useful is the prosthesis for the activity?

E. How well does your child do the activity without the prosthesis?

For the purposes of this report, the answers to PUFI Part-II question E were compared with the UBET Completion of Task data for subjects who did not wear a prosthesis and for prosthesis wearers with the prosthesis off.

Because no objective function test was available to assess children with unilateral congenital below-the-elbow deficiency with and without their prosthesis, the UCBED Study Group developed, piloted, and established the interrater reliability of the Unilateral Below-the-Elbow Test (UBET)[23]. We selected bimanual tasks that are important for activities of daily living and that incorporate age-appropriate gross and fine motor skills; thus, the UBET tests the activity component of function objectively. Most tasks were selected from two well-established tests of prosthetic function, the University of New Brunswick Test of Prosthetic Function (UNB) and the PUFI[21,22]. Nine tasks were chosen for each of four age categories (see Appendix). Children were scored on their ability to complete the task as instructed. The Completion of Task score is an ordinal scale from 0 to 4 (Table III) that describes the ease of completion and the quality of movement displayed by a child with unilateral congenital below-the-elbow deficiency when performing a task with or without the use of a prosthesis. Wearers were tested twice, once with and once without their prosthesis, in a randomized order. Non-wearers were tested once, without a prosthesis. Test performance is videotaped, and scores are assigned by an occupational therapist viewing the videotaped performance. The interrater reliability of twelve occupational therapists independently assigning Completion of Task scores to the same UBET performances was high (Spearman rho coefficient = 0.98)[23].

A two-tailed t test was used to compare both PedsQL and PODCI scores between subjects who did not wear a prosthesis (non-wearers) and the subjects who did (wearers). Complete PedsQL results were available for 449 subjects (317 wearers and 132 non-wearers), and complete PODCI results were available for 467 subjects (320 wearers and 147 non-wearers). In addition, a two-tailed t test was used to compare the PedsQL and PODCI scores for the children and adolescents with unilateral congenital below-the-elbow deficiency with those scores in the general population[16,19]. Complete UBET scores were available for 385 subjects with unilateral congenital below-the-elbow deficiency (257 wearers and 128 non-wearers, with all wearers tested with and without their prosthesis). Completion of Task scores for wearers using their prosthesis were compared with the scores when the wearers did not use the prosthesis (paired t test) and with the scores for the non-wearers (unpaired t test). Mann-Whitney U tests were used to compare the UBET Completion of Task scores with the PUFI Part-II question-E data. A power analysis performed prior to the initiation of subject enrollment indicated that, with use of an alpha of 0.05 and a beta of 0.8, 144 subjects, or ninety-six prosthesis wearers and forty-eight non-wearers (given an anticipated 2:1 ratio of available subjects), should be enrolled for comparison of the two groups to ensure that a finding of no significant difference was unlikely to be

TABLE V PODCI Respondents and Scores

	Wearers	Non-Wearers	P Value
Age *(no. of subjects)*			
2-10 yr	190	93	
11-20 yr	130	54	
Total	320	147	
PODCI scores *(points)*			
Upper Extremity Physical Function	90.4	90	0.72
Transfers/Basic Mobility	98	97.2	0.03
Sports/Physical Function	90.8	90.5	0.74
Pain/Comfort	92.2	92.2	0.99
Happiness	89.9	89.5	0.80
Global Function	92.8	92.6	0.69

due to insufficient statistical power. Conversely, the multiple comparisons of measures from the various instruments increased the likelihood of making a Type-I error. As such, $p < 0.01$ was the criterion used to determine significance in all analyses.

Results

PedsQL results showed a significantly higher average score on the School Functioning Scale in the Psychosocial Health Domain for prosthesis wearers compared with non-wearers ($p < 0.001$). No other differences between the PedsQL scores of the prosthesis wearers and non-wearers reached significance at $p < 0.01$ (Table IV).

PODCI results showed no significant differences between wearers and non-wearers. Regardless of prosthetic use, the average PODCI scores were high for all age groups (>85 points except for Upper Extremity Function for two to five-year-olds) (Table V).

According to the parents' ratings for the subjects and the subjects' own ratings, the PedsQL scores for the children and adolescents with unilateral congenital below-the-elbow deficiency were slightly higher than those for the general population[16] in the Physical Health Domain (Fig. 1).

The PODCI scores for the children and adolescents with unilateral congenital below-the-elbow deficiency were also compared with those in the general population[20] (Figs. 2 and 3). Parents of two to ten-year-old children with below-the-elbow deficiency reported lower scores in the Upper Extremity Physical Function Domain (average score, 86.2 points for prosthesis wearers and 85.3 points for non-wearers) than did parents of the same-age children in the general population (average score, 92.0 points), regardless of whether the subjects used a prosthesis ($p < 0.001$ for wearers compared with the general population, and $p < 0.001$ for non-wearers compared with the general population). The eleven to twenty-year-old subjects with below-the-elbow deficiency reported lower scores in the Upper Extremity Physical Function Domain than did same-age individuals in the general population, with an average score of 96.0 points for prosthesis wearers, 97.0 points for non-wearers, and 98.7 points for the general population ($p < 0.01$ for wearers compared with the general population, and $p < 0.01$ for non-wearers compared with the general population). The parents' scores in the Mobility/Transfer Domain for two to ten-year-olds with unilateral congenital below-the-elbow deficiency were lower than the scores for the same age group in the general population regardless of whether the study participants used a pros-

TABLE VI Comparison of UBET Scores Between Prosthesis Wearers Wearing Their Prosthesis and Wearers without Their Prosthesis

		Average UBET Score *(points)*		
	No. of Subjects	Not Wearing Prosthesis	Wearing Prosthesis	P Value
2-4 yr	42	3.2	3.9*	0.02
5-7 yr	57	3.1	2.9	0.06
8-10 yr	52	3.5	3.4	0.05
11-20 yr	106	3.7	3.6	<0.001
Total	257	3.5	3.3	<0.01

*The score for the two to four-year-olds wearing the prosthesis differs from that in Table VII because the score in Table VI represents only the wearers who had UBET scores for all nine tasks performed both by while wearing the prosthesis and while not wearing it. The score in Table VII represents all wearers who had scores while wearing the prosthesis. As a result of test fatigue, some two to four-year-old wearers were unable to complete the UBET test under both prosthesis conditions.

TABLE VII Comparison of UBET Scores Between Prosthesis Wearers Wearing Their Prosthesis and Non-Wearers

	No. of Subjects		Average UBET Score *(points)*		
	Wearers	Non-Wearers	Wearers Wearing Prosthesis	Non-Wearers	P Value
2-4 yr	42	24	2.9*	2.7	0.42
5-7 yr	57	33	2.9	3.3	0.002
8-10 yr	52	21	3.4	3.6	0.014
11-20 yr	106	50	3.6	3.8	<0.002
Total	257	128	3.3	3.5	<0.01

*The score for the two to four-year-olds wearing the prosthesis differs from that in Table VI because the score in Table VI represents only the wearers who had UBET scores for all nine tasks performed both while wearing the prosthesis and while not wearing it. The score in Table VII represents all wearers who had scores while wearing the prosthesis. As a result of test fatigue, some two to four-year-old wearers were unable to complete the UBET test under both prosthesis conditions.

thesis (average score, 97.3 points for wearers, 96.0 points for non-wearers, and 98.4 points for the general population; $p < 0.01$). In addition, the parents' scores for Global Function for the two to ten-year-old prosthesis wearers were lower than the scores in the general population (91.6 compared with 93.3 points; $p < 0.01$). Eleven to twenty-year-olds with unilateral congenital below-the-elbow deficiency scored significantly higher in the Happiness Domain than did the general population, regardless of whether the study subjects used a prosthesis (average score, 87.5 points for wearers and 88.1 points for non-wearers compared with 81.8 points for the general population; $p < 0.01$ for both comparisons).

Of the twenty-six tasks listed in the PUFI for two to five-year-olds (see Appendix), tying shoes was perceived by the parents to be the most difficult for both prosthesis wearers and non-wearers. There was no significant difference between wearers and non-wearers for the performance of any task without a prosthesis. In the eight to twelve-year-old group, non-wearers scored higher than wearers for the performance of three tasks without the prosthesis (putting toothpaste on a toothbrush [$p = 0.003$], cutting out a shape with scissors [$p = 0.003$], and using a ruler and pencil to draw a line [$p = 0.005$]). There were no differences between these groups for the remaining thirty-five tasks. In the thirteen to twenty-year-old group, non-wearers

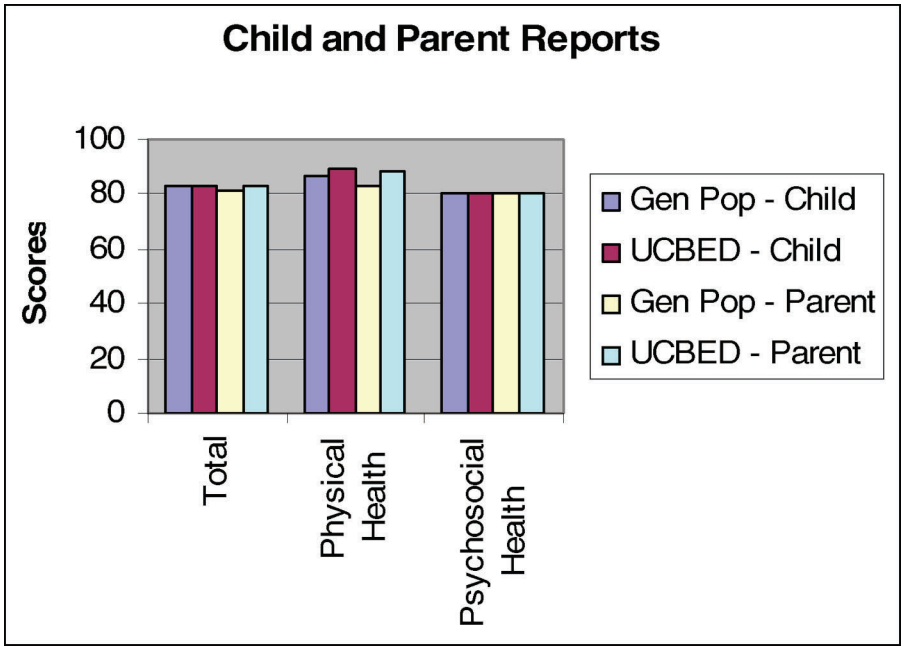

Fig. 1

PedsQL scores for the subjects with unilateral congenital below-the-elbow deficiency (UCBED) compared with those in the general population. With the Physical Health Scale, the children with unilateral congenital below-the-elbow deficiency scored themselves significantly higher than did the children in the general population ($p < 0.001$) and the parents of children with unilateral congenital below-the-elbow deficiency assigned significantly higher scores than did the parents of children in the general population ($p < 0.01$); however, these differences did not reach the minimal clinically relevant difference as defined by Varni et al.[16]. The general population data were derived from the study by Varni et al.[16].

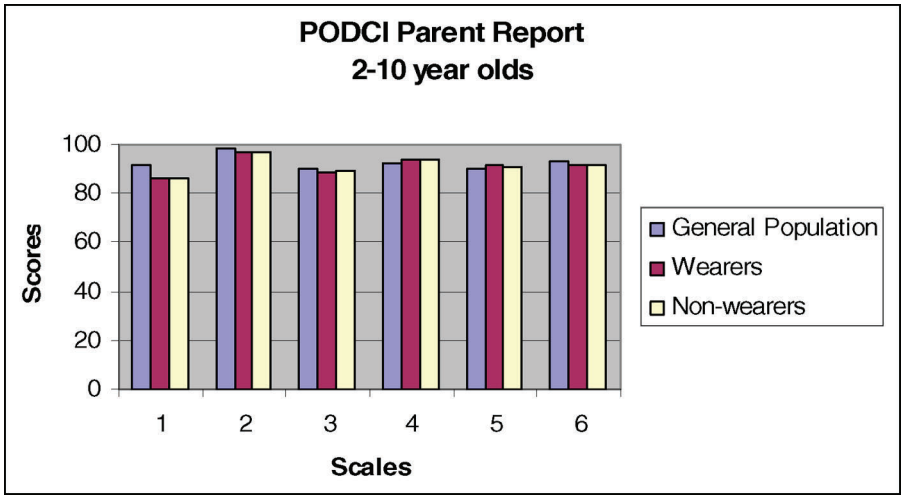

Fig. 2

Parent-reported PODCI scores for two to ten-year-old children with unilateral congenital below-the-elbow deficiency (prosthesis wearers and non-wearers) compared with scores for the same age group in the general population[20]. The parents of the prosthesis wearers and non-wearers with unilateral congenital below-the-elbow deficiency assigned significantly lower scores than did the parents of children in the general population for Scale 1 (Upper Extremity Physical Function, $p < 0.001$) and Scale 2 (Mobility/Transfers, $p < 0.01$). The parents of the prosthesis wearers assigned a significantly lower score than did the parents of children in the general population for Scale 6 (Global Function) ($p < 0.01$). The general population data were derived from the study by Hunsaker et al.[20]. Scale 3 = Sports/Physical Function, Scale 4 = Pain/Comfort, and Scale 5 = Happiness.

scored higher than wearers for the performance of five tasks without the prosthesis (zipping a jacket [$p = 0.004$], putting on gloves [$p < 0.001$], peeling back the plastic cover of a snack pack [$p = 0.001$], raking leaves [$p = 0.01$], and throwing a basketball [$p = 0.006$]). There were no differences between these groups for the remaining thirty-three tasks.

The UBET scores for wearers using their prosthesis were compared with the scores for wearers not using their prosthesis (Table VI) and with the scores for non-wearers (Table VII). When the UBET scores for all ages were combined, the non-wearers scored better than the wearers wearing their prosthesis and the wearers scored higher when they did not wear their

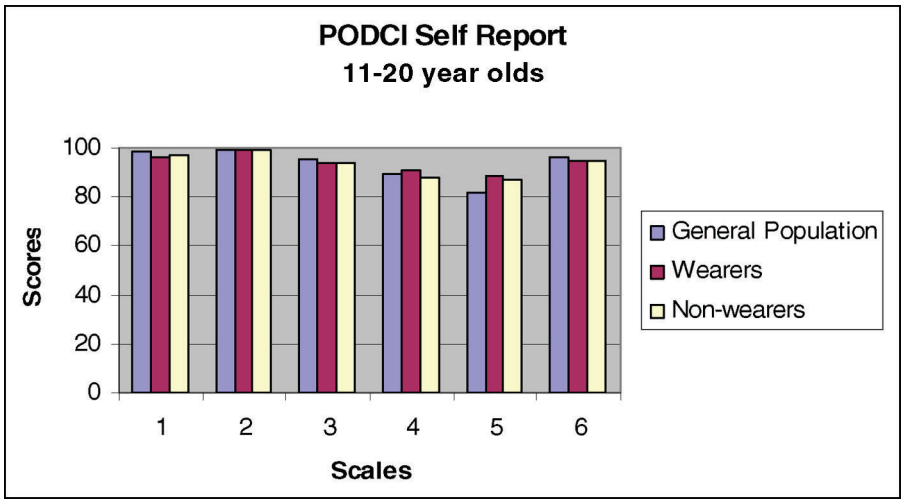

Fig. 3

PODCI scores reported by eleven to twenty-year-old subjects with unilateral congenital below-the-elbow deficiency (prosthesis wearers and non-wearers) compared with scores in the general population[20]. Prosthesis wearers and non-wearers with unilateral congenital below-the-elbow deficiency scored themselves lower than did the general population for Scale 1 (Upper Extremity Physical Function; $p < 0.01$ for both wearers and non-wearers). Both wearers and non-wearers scored themselves higher than the general population for Scale 5 (Happiness). The general population data were derived from the study by Hunsaker et al.[20]. Scale 2 = Mobility/Transfers, Scale 3 = Sports/Physical Function, Scale 4 = Pain/Comfort, and Scale 6 = Global Function.

prosthesis than when they did wear it. In the five to seven-year-old group, non-wearers had higher scores than wearers using their prosthesis. In the two to four-year-old and eight to ten-year-old groups, there were no significant differences (at $p < 0.01$) between non-wearers and wearers using their prosthesis. Similarly, in the two to four, five to seven, and eight to ten-year-old groups, there were no differences between wearers with and without their prosthesis (Table VI). In the eleven to twenty-year-old group, non-wearers scored better than wearers, and wearers scored better without their prosthesis (Table VII).

Discussion

We found that use of a prosthesis by children with a unilateral congenital below-the-elbow deficiency was not associated with relevant differences in function or quality of life, an observation that calls into question the accepted practice of early and routine prescription of prostheses to improve function or quality of life of children with unilateral congenital below-the-elbow deficiency.

Although the health-related quality of life of children and adolescents has been studied, few investigators have focused on individuals with an upper-extremity amputation. In a study of more than 1000 adults with an upper or lower-extremity amputation, Demet et al. found that, although the respondents had a diminished health-related quality of life associated with physical disability, pain, and energy level, both a young age at the time of the amputation and an upper-limb amputation were associated with a better quality of life in this population[24]. Little information regarding the health-related quality of life of children and adolescents with a limb deficiency is available.

Our study strongly suggests that, compared with the general population, children with unilateral congenital below-the-elbow deficiency, and their parents, do not perceive their health-related quality of life to be diminished and that use of a prosthesis does not have an impact on health-related quality of life as measurable by the PedsQL. Although wearers scored significantly higher than non-wearers on the School Functioning Scale in the Psychosocial Health Domain of the PedsQL, this score is based on responses to questions that are likely to be related to prosthesis wear, such as "Is it hard for you to pay attention at school?", "Do you forget things?", and "Do you miss school because of not feeling good?"[16]. Furthermore, the 8-point difference between wearers and non-wearers did not meet the definition, by Varni et al.[16], of the minimal clinically important difference for this scale (9 points). Similarly, although children with unilateral congenital below-the-elbow deficiency scored significantly higher than the general population on the Physical Health Scale of the PedsQL, the differences between these two groups (5 points when scored by the parents and 2 points when scored by the subjects) did not meet the definition, by Varni et al.[16], of the minimal clinically important difference for this scale (7 points).

Similar reservations about clinical importance arise when the PODCI scores for children with unilateral congenital below-the-elbow deficiency are compared with those of the general population. For instance, although subjects with unilateral congenital below-the-elbow deficiency scored significantly lower in the Upper Extremity Physical Function Domain than did the general population, the differences between these groups were small (1 to 2 points) and all scores were high (96.0 points [of a possible 100 points] for wearers, 97.0 points for non-wearers, and 98.7 points for the general population). In addition, although the average PODCI scores for the two to ten-year-old subjects were statistically and possibly clinically significantly lower than those in the general population, they were still within the range defined as normal (above the low 80s[19]). Thus, this study shows that, as measured with the PODCI, the self-reported musculoskeletal health and function of older children with unilateral congenital below-the-elbow deficiency is not different from the musculoskeletal health and function of older children in the general population. This finding may indicate that the PODCI has a ceiling effect for older children with unilateral congenital below-the-elbow deficiency; it is possible that a tool with more specific questions about more complex activities might uncover functional deficits in this population. Parents of younger children with unilateral congenital below-the-elbow deficiency reported lower scores in the Upper Extremity Physical Function Domain than did parents of children in the same age group in the general population; however, the scores for the subjects who wore a prosthesis did not differ significantly from the scores for those who did not wear a prosthesis.

The significant differences measured by the PUFI Part-II Question E between the prosthesis wearers when they wore their prosthesis and the wearers when they did not wear the prosthesis, and between the wearers and the non-wearers, were quite small and probably not meaningful, although all favored non-wearers and wearers without their prosthesis. It seems safe to conclude that prostheses do not help children and adolescents with unilateral congenital below-the-elbow deficiency to function, as measured by the tests used in this study.

According to the UBET scores, the five to seven-year-old and eleven to twenty-year-old non-wearers performed better than did the wearers, and the eleven to twenty-year-old wearers performed better with their prosthesis off. There were no significant differences in the other age groups, although the average scores for all age groups combined reflected the results for the eleven to twenty-year-olds. As was the case for the PedsQL and PODCI, however, these significant differences were small and possibly not clinically relevant. Again, use of a prosthesis did not appear to help the subjects to perform the UBET tasks.

This study was cross-sectional, but the disadvantages of this methodology are offset somewhat by the large number of children who were studied. In addition, in the Shriners Hospitals system, children and families choose whether or not to wear a prosthesis and most are allowed to choose the type of prosthesis, diminishing the chance of a bias associated with children being constrained to use a type of prosthesis that did not appeal to them or their parents.

The results of this study do not indicate that any individual child will not benefit from a prosthesis. They do indi-

cate that children and adolescents with unilateral congenital below-the-elbow deficiency have close to normal but not normal function, and they imply that current prostheses are not normalizing function, leaving room for improvement in upper-extremity prosthetic technology.

Instead of asking why children and adolescents with unilateral congenital below-the-elbow deficiency frequently stop using a prosthesis, perhaps we should be asking why some people with this type of deficiency continue to wear a prosthesis. Prostheses may promote social acceptance by altering appearance, and they may be useful as tools for specific high-level activities, such as some sports. Children and adolescents may benefit from being allowed to choose the type of prosthesis that fits their individual needs, and when they are allowed to do so freely, they may frequently choose a passive hand prosthesis[25], indicating that appearance may be more important than the function of an active terminal device.

This study shows that, according to well-validated quality-of-life and musculoskeletal health questionnaires, children and adolescents with unilateral congenital below-the-elbow deficiency score the same as or better than the general population, or within the normal range, in all domains. All of the children who were tested had a specific type of unilateral hand malformation. The fact that they functioned within the normal range with only one hand calls into question any reported functional improvements associated with reconstruction of other types of unilateral hand malformations. Prostheses have been assumed to improve the function of children with unilateral deficiency, but they did not do so in this study. Assumptions about the benefits of surgical treatment to improve the function of children with unilateral upper-extremity malformations may also require further evaluation. Reported functional improvements following any intervention in children with congenital malformations should be demonstrated by standardized subjective and objective measures and a comparison of developmentally adjusted postoperative function with preoperative function and with age-appropriate function in the general population.

Appendix

eA Tables showing the specific elements of the PUFI and the UBET are available with the electronic versions of this article, on our web site at jbjs.org (go to the article citation and click on "Supplementary Material") and on our quarterly CD-ROM (call our subscription department, at 781-449-9780, to order the CD-ROM). ■

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