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Parental Assessment of Status of Congenital Upper Limb Differences: Analysis of the Pediatric Outcomes Data Collection Instrument (PODCI):

Upper Extremity Congenital PODCI

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Abstract

Introduction—To determine the range of the Pediatric Outcomes Collection Instrument (PODCI) scores for children with a wide variety of congenital upper limb differences and to examine the scoring effect of the patient's surgical history, family history, severity of involvement, and syndromic associations.

Methods—We reviewed the PODCI scores for 109 patients, aged 2–18 years, treated for non-traumatic upper extremity conditions. Charts were reviewed for sex, age, extent of limb involvement, laterality, family history, surgical history, and syndrome association. All patients were classified based on the Oberg, Manske, Tonkin Classification with general categories of malformation, deformation, or dysplasia.

Results—Eighty of 109 (73%) patients had a malformation, 12 (11%) had a deformation, and 17 (16%) had a dysplasia. The cohort as a whole had a happiness PODCI score that was similar to the normal population, yet a lower (worse) PODCI score for upper extremity and global function. Patients with a dysplasia had a higher upper extremity function scores than those with malformations or deformations, but they had similar happiness and global function scores. Complete upper limb involvement and lower extremity involvement statistically lowered the PODCI score within our study cohort, while a positive family history and syndromic association increased PODCI scores.

Conclusion—This study showed that there was a similar level of perceived happiness between children/adolescents with congenital upper extremity conditions compared to the normal pediatric population based on PODCI scores. In contrast, the perceived upper extremity and global function was significantly decreased in patients with congenital differences compared to normal individuals. This investigation also revealed that the extent of upper extremity involvement, lower extremity involvement, family history, and syndromic association may affect PODCI scores as

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independent variables and should be taken into consideration in studies of upper extremity congenital anomalies.

Level of Evidence IV—Prognostic Study

Keywords

Congenital; pediatric; PODCI; outcomes; upper extremity

Introduction

Congenital upper limb differences affect approximately 5 to 20 in 10,000 live births [1, 2]. These conditions vary notably in clinical presentation and management. Recently, a classification system developed by Oberg, Manske, and Tonkin (OMT) was adopted by the International Federation of Societies for Surgery of the Hand (IFSSH) [4]. This classification, in contrast to the Swanson classification [3], was developed based on the current understanding of developmental biology, embryology, and genetics. The OMT classification categorizes congenital differences as malformations, deformities, or dysplasia [5]. Malformations are defined as conditions resulting from a failure in development of limb formation or differentiation. Deformities describe conditions resulting from a disruption of an already formed limb. Dysplasias are conditions grouped based on appearance and have a known associated cellular atypia or tumor formation. The OMT was designed to adapt to the advancements in our understanding of embryology, and the IFSSH recommends that the classifications be revisited every 3 years.

Given the complexity and variability of congenital upper limb differences, a systematic and standardized measurement of function and outcome would hold major clinical value. The Pediatric and Adolescent Outcomes Data Collection Instrument (PODCI) was developed by the Pediatric Orthopedic Society of America and the American Academy of Orthopedic Surgeons as a tool for assessing functional outcome for a variety of musculoskeletal disorders in children [6]. The PODCI is a validated questionnaire that assesses a child's overall health, pain, and ability to participate in light and vigorous activities [7]. The PODCI assesses the child's or adolescent's condition in 8 specific scales: upper extremity and physical function, transfer and basic mobility, sports/physical functioning, pain/comfort, treatment expectations, happiness, satisfaction with symptoms, and global functioning. The questionnaire is completed by the parent for children ages 2–18 and serves as a measure of parental perception of a child's function; there is also a questionnaire completed by adolescents themselves (not used in this study). A previous study of children with surgically corrected hand deformities used a different assessment tool and demonstrated that clinical severity does not necessarily correlate with perceived impact of the hand difference [8]. Thus a better understanding of patient (or parent) perception for upper limb differences would be valuable.

Previous studies have characterized PODCI scores for patients with specific upper extremity conditions, including brachial plexus birth palsy, arthrogryposis, hand/wrist injury, and unilateral upper extremity deficiency [9–12]. These studies consistently demonstrated lower PODCI scores in children with upper extremity disorders compared to normal. However, the

perceived impact for a broad range of congenital upper extremity differences is unclear. The purpose of this study was to determine the PODCI scores for children with a wide range of diagnoses and to examine the effect of the surgical history, family history, severity of involvement, and syndromic associations on the PODCI score. We hypothesized that children with more severe limb involvement, extended surgical history, negative family history, and syndromic association would demonstrate a lower perceived outcome as measured by the PODCI.

Methods

The PODCI questionnaire has been given to the parents of all patients undergoing upper limb surgery for congenital differences at the St Louis Shriner's Hospitals for Children since 2007. We used only the parent-completed PODCI scores for both the child and adolescent cohorts. Between 2007–2015, there were a total of 288 treated patients over the age of 2 years, but only 186 PODCI scores were located within the medical records. Of these there were 111 fully completed parent PODCI questionnaires of patients aged 2–18 years. Parent PODCI, not child PODCI, was used to be consistent across ages. We excluded patients treated before the age of 2 years and those with an incomplete questionnaire. For patients with multiple PODCI questionnaires reflective of multiple surgeries, only the first valid PODCI was scored. This was chosen for consistency and to assess each child only once. One patient was excluded due to concurrent diagnosis of cerebellar agenesis and another due to the preoperative diagnosis of an upper extremity difference that was complicated by osteomyelitis. The remaining 109 patients with a completed questionnaire were included in the study.

After institutional review board approval was obtained, the medical charts were reviewed and the PODCI questionnaires were assessed. We recorded all relevant diagnoses as well as sex, age, adopted or natural born, extent of upper limb involvement (hand plate versus entire limb, as defined by the OMT classification), laterality, presence of lower limb involvement, family history, and surgical history. The PODCI domains of upper extremity function (UE), happiness, and global function were assessed in this study, as we felt that they were the most pertinent to this population.

For analysis, the study cohort PODCI scores were compared to previously published normal values established from children/adolescences with no orthopedic involvement [7]. Additionally, scores from individuals with previous surgeries were compared against those without prior surgical history to assess for effect of surgery on PODCI score. Positive family history was compared to no family history. A positive family history was defined as the presence of a limb anomaly in any family member as noted on the medical record. Limb severity was defined as entire limb involvement versus hand plate only, as categorized by the OMT classification, and compared.

The student *t*-test was used to compare subjects by OMT classification, surgical history, family history, extent of limb involvement, presence of lower extremity involvement, syndromic association, and adoption status. Statistical significance was defined as $P < 0.05$.

Results

Fifty (45%) of the 109 patients in the study were females. There were 93 (85%) subjects between the age of 2–10 years (child/children) and 16 (15%) between the ages of 11–18 years (adolescents). Only parent-completed PODCIs were considered.

Based on the OMT, 80 (73%) patients had a malformation, 12 (11%) had a deformation, and 17 (16%) had a dysplasia. Table 1. The mean PODCI score for upper extremity function was 77 (+/- 24), for global function was 85+/-15, and 90+/-14 for happiness. Table 2 reports PODCI scores by sex, age, and diagnosis.

Child cohort vs normal

Upper extremity function in our cohort was found to be significantly lower than previously published normal children ($P < 0.05$), except for those with dysplasia ($P=0.51$). Global function was also found to be lower in the study cohort ($P < 0.05$), except for deformations ($P=0.13$). The happiness with physical condition score was similar between previously published normals and most of the study cohort ($P > 0.05$) [13]. One difference was identified, children with hand plate only involvement did produce a higher than normal score compared to normal ($P=0.03$). Details are recorded in On-line Appendix 1

Adolescent cohort vs normal

The adolescent population, as a group, had an UE and global function scores lower than the mean scores ($P < 0.05$), while happiness was not statistically different, $P=0.92$. Specifically UE function was perceived to be less in all groups except those with lower limb involvement. With regards to global function, adolescents with lower limb involvement, no prior surgeries, a positive family history, associated syndrome, and dysplasia had statistically lower scores ($P < 0.05$). Happiness was higher in adolescents with lower extremity involvement, no prior surgery, syndromic association, and dysplasia ($P < 0.05$). (On-line Appendix 1)

Intra-cohort comparison

Patients diagnosed with dysplasia by the OMT criteria had a higher UE function score than patients with malformations and deformations ($P < 0.05$). There was no statistical difference between UE function in malformations and deformations ($P > 0.05$). And there was no difference between the 3 groups in happiness or global functioning ($P > 0.05$). (Table 3).

There was no relationship between the happiness score and lower limb involvement, extent of upper limb involvement, previous surgery, family history, adoption status, or syndromic association ($P > 0.05$). Upper extremity and global functioning results can be seen in On-line Appendix 1.

Discussion

The PODCI questionnaire is a commonly used assessment tool to evaluate outcome and function in the pediatric orthopedic population. In this investigation, we applied the PODCI to evaluate the perceived impact of congenital upper limb differences in the pediatric

population. The PODCI was sensitive in this population, demonstrating lower scores with measurement of UE function and global function.

In contrast, the happiness score was similar, in general, between the pediatric population with congenital anomalies and the normal population. This finding confirms the quite innovative findings of a study from Sweden that specifically evaluated the psychological well being of children with hand deformities, both mild and severe [8]. In this study, the 80-item Piers-Harris Children's Self-Concept Scale was used to assess child self-esteem and well being [13]. They found, similar to our result, that normal children and those with mild and severe hand anomalies showed no difference in overall happiness.

When analyzing the PODCI scores for our entire cohort, certain anatomic and demographic factors had an impact. First, there was a greater perceived impairment when the entire limb was involved compared to children/adolescents with only hand involvement. Second, when the lower limb was also involved, there was a resulting lower PODCI functional score, both UE and global. Both of these findings confirm that the PODCI is sensitive to the overall severity of involvement of the congenital anomaly, whether that includes the entire upper limb or involvement of the lower limbs as well. Lastly, individuals with a family history had a higher perceived UE function, which may be explained by a sense of a commonly shared condition and a better understanding of the functional impact. Despite all the factors evaluated, there was no difference in happiness for any of the following identified variables: hand plate- only involvement, lower limb involvement, previous surgery, family history, and adoption status or syndrome association.

A comparison of our results with those of previous studies of congenital upper extremity conditions assessed with the PODCI did not detect a statistical difference between mean PODCI scores. Lerman et al. reported a functional assessment of patients with unilateral upper extremity deficiencies using the PODCI score [12]. Their study included 13 non-congenital and 51 congenital unilateral upper extremity deficiencies producing an UE function score of 84 (+/-15). We found a score of 80 +/-19 in patients with unilateral conditions, which was similar to the finding of Lerman et al ($P > 0.05$). Also, our happiness and global functioning scores were similar to those presented by Lerman, et al (92+/-14 vs 88+/-16 and 88+/-11 vs 88+/-9).

Amor et al. studied 73 arthrogryposis patients, specifically those with amyoplasia, and reported a mean upper extremity functioning score of 60 (+/- 24) [9]. Our study, which had 18 arthrogryposis patients, revealed a mean upper extremity functioning score of 51 (+/-3). While the difference in scores is not statistically significant ($P = 0.17$), the 9-point difference is notable although a minimally clinically important difference for this population has yet to be determined. In contrast to upper extremity function, we did not see a significant difference in our results, compared to Amor et al. with regards to both happiness and global functioning ($P > 0.05$).

Bradbury et al. [14] reported the parental and child perspective of surgical outcomes of congenital upper extremity anomalies using pre-operatively collected data. Their study had a more expansive assessment of the parental psychology, and it demonstrated that the parents'

level of adjustment factored into their subjective assessment of post-operative function and outcome. They found better adjusted parents and children had better overall outcomes. While we did not attempt to categorize the baseline psychological differences within the parents and the child cohorts, we believe that this is an important consideration for those analyzing and caring for children with congenital upper extremity conditions.

There are limitations to this study. First, we used the parental PODCI and not a self-perceived report. The use of the parental PODCI may present an inherent level of bias in outcome since the PODCI scores are from the parent perspective. Secondly, we did not control for level of parental education or comprehension of the parent while completing the PODCI survey. Lastly, the PODCI itself is a limitation. Its length and resulting time required to complete the 117-item PODCI questionnaire makes it difficult to obtain all necessary data. Our study faced numerous incomplete questionnaires, limiting our data collection. This supports the current call for more user-friendly concise questionnaires. Potential candidates at this time include the Patient Reported Outcomes Measurement Information System (PROMIS) [15], a focused and validated electronic data collection instrument; this questionnaire has yet to be used at our institution, but it shows great possibilities for improving overall data intake.

In our analysis we revealed from the variables analyzed that the extent of upper extremity involvement, lower extremity involvement, family history, and syndromic association may affect PODCI scores independent of the congenital anomaly. Therefore, we recommend that these factors be considered and controlled for in analysis of outcomes of congenital upper extremity conditions in future research studies.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Table 1

Demographics of study population.

	N	%
Males	59	54%
Females	50	46%
Age < 5 for 1st PODCI	50	46%
Age ≥ 5 for 1st PODCI	59	54%
Syndactyly	8	7%
Polydactyly	11	10%
Symbrachydactyly	12	11%
Radial Longitudinal Deficiency	17	16%
Ulnar Longitudinal Deficiency	3	3%
Central Ray Deficiency	8	7%
Arthrogryposis	18	17%
Constriction Band Syndrome	10	9%
Osteochondromatosis	17	16%
Trigger Finger	2	2%
Camptodactyly	2	2%

Table 2

Comparison of PODCI scores based on upper limb involvement, lower limb involvement, surgical history, family history, syndromic association, and adoption status (N=109).

	N	Upper Extremity Functioning			Happiness with Physical Condition			Global Functioning		
		Mean	SD	P-value	Mean	SD	P-value	Mean	SD	P-value
Limb Involvement	Hand Plate Only	82	18	.02*	93	12	.07	91	9	.000*
	Upper Limb Involvement	72	27		87	16		79	17	
Lower Limb Involvement	- Lower Limb Involvement	83	18	.005*	90	15	.93	89	10	.001*
	+ Lower Limb Involvement	70	28		90	14		79	17	
Surgical History	- Prior Surgery	80	22	.26	93	10	.08	85	15	.91
	+ Prior Surgery	75	25		88	16		85	15	
Family History	- Family History	71	26	.001*	90	16	.95	83	17	.11
	+ Family History	87	16		91	13		88	12	
Syndrome	No Associated Syndrome	74	24	.03*	89	15	.73	84	16	.25
	Syndromic Association	86	22		91	13		87	11	
Adoption Status	Not Adopted	78	24	.06	90	14	.32	85	15	.30
	Adopted	62	23		85	14		80	14	
OMT Classification	Malformation	72	25	0.38 (M vs Def)	89	15	.57	83	16	.06
	Deformation	78	16	.006* (Def vs Dys)	92	13	.70	90	9	.67
	Dysplasia	95	8	.000* (Dys vs M)	90	14	.89	88	12	.15

Table 3

Comparison of mean PODCI scores based patient characteristics and diagnoses.

	Upper Extremity Mean Score	Happiness Mean Score	Global Functioning Mean Score
ALL	76.6	89.9	84.4
Males	74.3	88.9	82.9
Females	79.2	91.0	86.9
Age < 5 for 1st PODCI	70.9	95.5	84.1
Age ≥ 5 for 1st PODCI	80.7	87.0	85.2
Syndactyly	77.2	95.8	88.4
Polydactyly	93.7	93.0	97.2
Symbrachydactyly	83.8	90.4	88.5
Radial Longitudinal Deficiency	71.1	88.0	83.9
Ulnar Longitudinal Deficiency	83.3	73.3	82.5
Central Ray Deficiency	82.8	93.1	92.7
Arthrogryposis	50.8	85.5	65.2
Constriction Band Syndrome	76.3	91.3	89.3
Osteochondromatosis	95.1	90.0	88.3
Trigger Finger	91.7	100.0	97.2
Camptodactyly	89.6	87.5	89.9