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Publication Date
2020

Data Availability
The data associated with this publication are not available for this reason: N/A

Patient Reported Outcomes Measurement Information System (PROMIS) Scores for Children with Arthrogryposis Affecting the Upper Extremity

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INTRODUCTION

- Children with arthrogryposis involving the upper extremity (UE) commonly have impaired motion and strength due to contractures; however, they develop compensatory movements to achieve a higher level of function than may be anticipated[1]
- The World Health Organization (WHO) International Classification of Function – Children and Youth[2] recommends evaluation of body structure & function, ability to perform activities and participate in life roles, and quality of life
    - Clinicians typically evaluate body structure & function (strength & range of motion), but this does not necessarily characterize a child's ability to perform activities and participate in life roles
    - The Patient Reported Outcome Measurement System (PROMIS)[3,4] is designed to measure all WHO domains for people with chronic disease
    - PROMIS Pediatric Short Forms include 5-8 questions that measure Mobility, UE Function, Pain Interference with activities, and Peer Relationships using T scores that compare to a reference population
- PROMIS has been used in two studies of arthrogryposis[5,6] :both were encouraging but small
- A study of a larger cohort could better describe function and validate PROMIS in this population

OBJECTIVES

1. Determine whether children with arthrogryposis report worse mobility and upper extremity function than the reference population
2. Determine whether children with arthrogryposis report worse peer relationship quality and higher pain interference than the reference population

METHODS

- Institutional Review Board approval was obtained for a single center retrospective study of children with arthrogryposis who received treatment at Shriners Hospital for Children Northern California (SHCNC)

Inclusion Criteria

- > 4 years and < 18 years of age
- Diagnosis of arthrogryposis (arthrogryposis multiplex congenital or distal arthrogryposis)
- Treated between April 1,2017 and May 20, 2019

Data Collection

- Demographics including age, sex, race, and area deprivation index (ADI)
    - ADI is a numeric scale from 1-10 (least to most) that helps quantify degree of disadvantage for a given neighborhood
- Individual PROMIS questions/answers and PROMIS subdomain T-scores
    - Proxy report for age 5-7
    - Self-report for age 8-17

Data Analysis

- Basic statistics (mean, SD) completed for demographics and PROMIS subdomain T-scores
- Student's t-test used to compare means between study participants with arthrogryposis and the reference population

Table 1. Characteristics of the Study Cohort

Characteristic	Number (%)
All	68 (100%)
Age in years (mean ± SD)	9.8 ± 3.8
Gender	
Male	34 (50%)
Female	34 (50%)
Ethnicity	
White	41 (60.3%)
Asian	8 (11.8%)
African- American	2 (2.9%)
American Indian or Alaskan Native	2 (2.9%)
Native Hawaiian/Other Pacific Islander	2 (2.9%)
Other	13 (19.1%)
CA Area Deprivation Index (mean decile ± SD, median)	7.4 ± 2.2, 8

RESULTS

- Subjects with arthrogryposis reported:
    - Moderately impaired Mobility (38.0±8.9) and UE Function (31.8±12.2) scores, both significantly different than the reference population (p<0.00001)
    - Excellent Peer Relationships (54.0±8.3), significantly different than the reference population (p=0.015)
    - Normal Pain Interference (49.3±10.0), not significantly different than the reference population (p=0.709)

Figure 1. Distribution of PROMIS Subdomain Scores

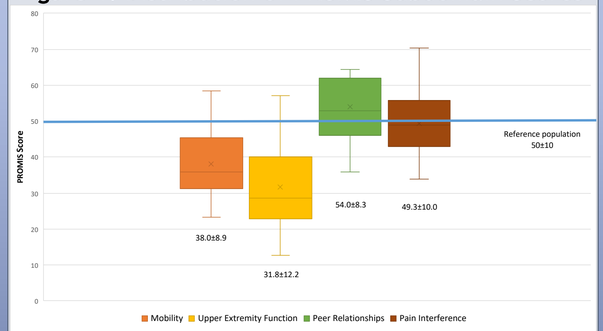
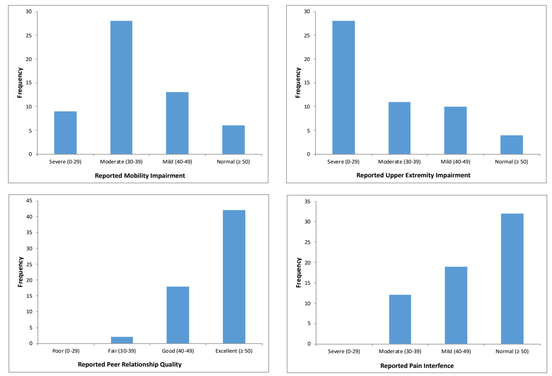


Figure 2. Frequency of PROMIS Subdomain Scores



CONCLUSIONS

- Children with arthrogryposis affecting the UE report significant impairments in Mobility and UE Function compared to the reference population, and similar (and possibly stronger) peer relationship scores than the reference population; their Pain Interference scores are similar to the reference population
- Future work will analyze the relationship between a patient's ADI and PROMIS scores

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ACKNOWLEDGEMENTS

Thank you to Dr. Michelle James, Dr. Claire Manske, Ms. Elizabeth Molnar, Ms. Kory Bettencourt, and Ms. Nancy Abarca at SHCNC