# PROMIS Scores for Children with Unilateral Congenital Below Elbow Deficiency (UCBED)

Azarin Shoghi, BS, MS[1]; Anita Bagley, PhD, MPH[1,2]; Michelle A. James, MD[1,2]

[1] Shriners Hospital for Children, Northern California (SHCNC); [2]University of California, Davis School of Medicine; Sacramento, CA, USA

## INTRODUCTION

- Patients with unilateral congenital below elbow deficiency (UCBED) are born with a unilateral transverse deficiency at the level of the forearm.
- Based on previous studies,[1,2] these patients have been shown to have milder than expected disability and their impairment is not reduced by using a prosthesis.
- This work resulted in changes in practice regarding prosthetic prescription for children with UCBED. However, it did not address the social stigma caused by this impairment, nor did it provide actionable information to improve the functional deficits that these children experience.
- Although PROMIS discerns UE function in other congenital arm differences[(3)], results for children with UCBED are not known.
- This study evaluates their responses to PROMIS Short Form Upper Extremity Function (UEF), Mobility, and Peer Relationships (PR) domains.

## OBJECTIVES

1. Obtain and evaluate PROMIS scores for children with UCBED as this has not been done on this patient population previously.
2. Determine if children with UCBED report worst outcomes on the UE function, Mobility and peer relationships compared to a reference population.

## MATERIALS and METHODS

Retrospective review of children with a diagnosis of UCBED who received treatment at Shriners Hospital for Children Northern California.

Inclusion Criteria

- > 4 years old and < 18 years of age
- Diagnosis of UCBED
- Completion of PROMIS questionnaire between April 1, 2017 and March 31, 2020

Exclusion Criteria

- Contralateral upper limb deficiency
- Lower limb deficiency

Data Analysis

- Mean and standard deviation completed for the PROMIS subdomain T-scores
- Student t-test used to compare means between patients with UCBED and those of a reference population.
    - T-score ≥ 50 = normal; 40-49 = mild; 30-39 = moderate; 0-29 = severe impairment

## MATERIALS and METHODS

**Figure 1.** Patient with UCBED

## RESULTS

28 children/proxies completed questionnaires (mean 11±3.4 years). They scored the same as the reference population for Mobility and PR and reported lower scores for UEF (43.5±9.3; P=0.02).

- 5-7-year-olds (5 children) reported normal PR, mildly impaired Mobility (46.0 ± 5.6), and moderately impaired UEF (30.2 ± 3.5; P=0.003);
- 8-12-year-olds (13 children) reported normal PR and Mobility and moderately impaired UEF (42.7±8.0).
- 13-17-year-olds (10 children) reported normal outcomes on all three domains.

## RESULTS

**Table 1.** Patient demographic

| Characteristic of Study Cohort | Number (%) |
|---|---|
| **All** | 28 (100%) |
| **Age in years (mean +/- SD)** | 11 +/- 3.4 yrs old |
| **5-7-year-olds** | 5 (17.9%) |
| **8-12-year-olds** | 13 (46.4%) |
| **13-17-year-olds** | 9 (32.1%) |
| **Gender** | |
| **Male** | 16 ( 57%) |
| **Female** | 12 (43%) |
| **Ethnicity** | |
| **White** | 10 (35.7%) |
| **Asian** | 6 (21.4%) |
| **African-American** | 0 (0%) |
| **American Indian or Alaskan Native** | 0 (0%) |
| **Native Hawaiian/Other Pacific Islander** | 0 (0%) |
| **Other** | 12 (42.8%) |

## RESULTS

Figure 2. Entire cohort PROMIS

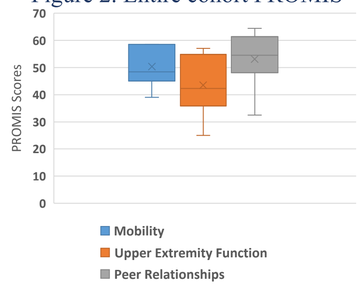



Figure 3. 5-7 years old PROMIS

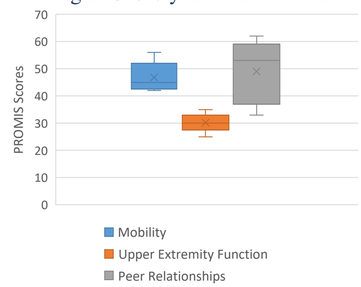



Figure 4. 8-12 years old PROMIS

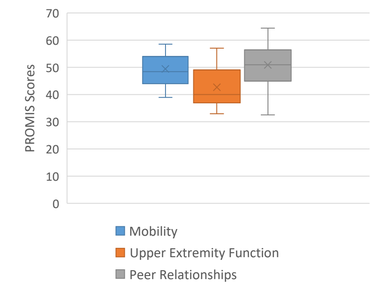
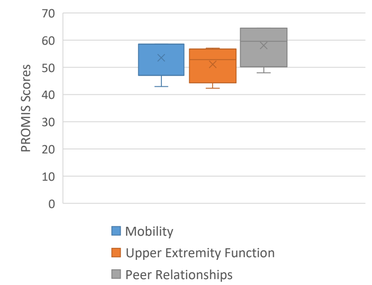



Figure 5. 13-17 years old PROMIS




## CONCLUSIONS

1. Impairment measured by PROMIS is similar to previous studies for children with UCBED.
2. 5-7-year-olds reported lower UEF scores; for children in this age group, UEF tasks may be too difficult, and/or parents may under-report function[(4)]. A study of typically developing 5-7-year-olds is underway to determine whether the UEF Short Form questions have a floor effect for this age group.
3. PROMIS UEF may have a ceiling effect for older children with UCBED.
4. Children with UCBED have a potentially stigmatizing UE difference, but do not report challenges with peer relationships.
5. This was a cohort of only 28 children. Future studies will analyze a larger group of children looking at the same domains of PROMIS scores.

## REFERENCES

(1) James MA et al. Impact of prostheses on function and quality of life for children with unilateral congenital below-the-elbow deficiency. JBJS 2006; 88:2356.

(2) Bagley AM et al. The Unilateral Below Elbow Test: a function test for children with unilateral congenital below elbow deficiency. Dev Med Child Neuro 2006; 48:569.

(3) Oishi S et al. Treatment and outcomes of arthrogryposis in the upper extremity. Am J Med Genet 2019.

(4) Sheffler LC et al. Comparison of self-reports and parent proxy-reports of function and quality of life of children with below-the-elbow deficiency. JBJS 2009; 91:2852.

RESEARCH POSTER PRESENTATION DESIGN © 2015
www.PosterPresentations.com