

# Development of a condition-specific measure of the upper extremity: Engagement of youth and families with arthrogryposis, Rasch analysis and the use of artificial intelligence

Laurie Snider PhD, OT(C.), erg.

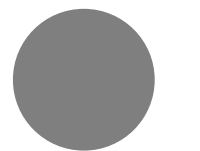
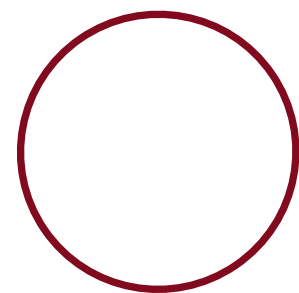
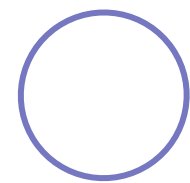
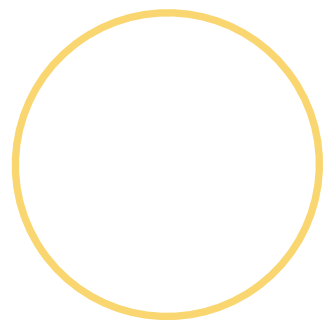
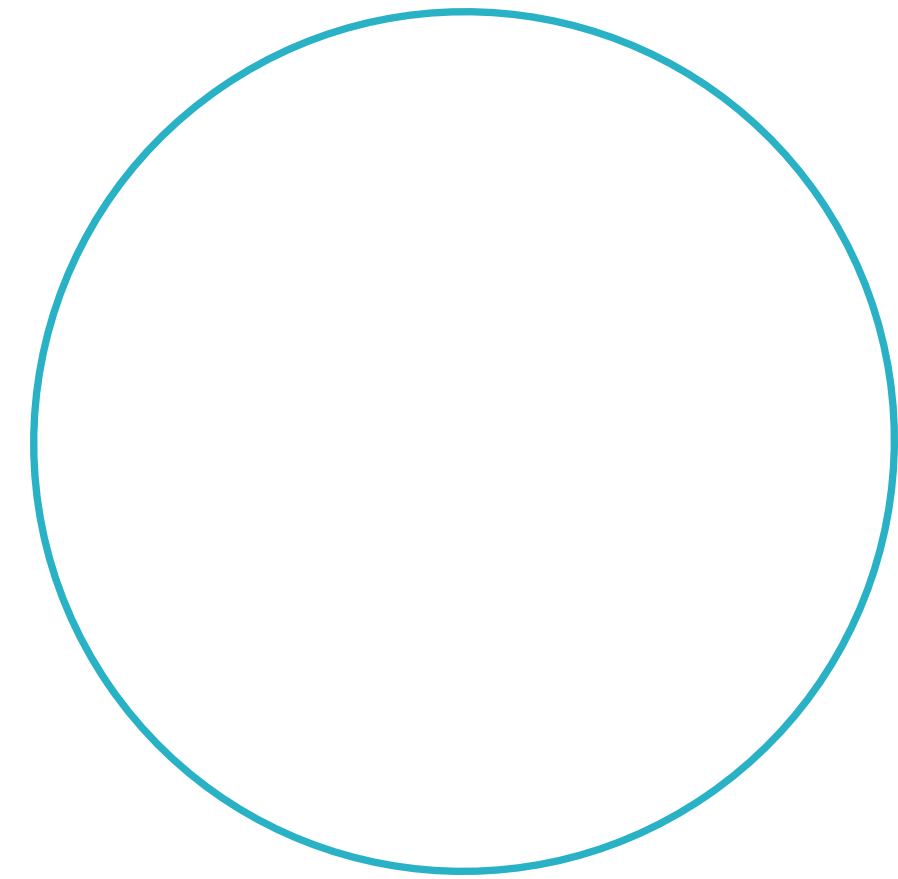
Noémi Dahan-Oliel, Caroline Elfassy, Kathleen Montpetit,  
Neda Naghdi, and Johanne Higgins



# Overview of Pediatric Rare Disease

Development of the SHAPE-UP

- WHO estimates a population of 93 million children aged 0-14 with moderate to severe disabilities (WHO, 2004)
- Orphanet database states that approximately 4% of the total world population is affected by a rare disease at any given time (Nguengang et al 2020)
- Musculoskeletal (MSK) conditions are among the most prevalent type of chronic pediatric conditions worldwide. (Bradley et al, 1994 Brooks 2006)
- Pediatric rare diseases are often complex and chronic with limited treatment options (EURODIS, 2022)
- Arthrogryposis Multiplex Congenita (AMC) is a rare group of conditions characterized by joint contractures in two or more body areas (Cacheco et al, 2019)
- Occurring in 1/3000 live births, AMC describes multiple contractures at birth. These contractures affect joint mobility and muscle development in the upper and lower limbs, spine and jaw, with involvement of the upper extremity (UE) in 73% of children, thus limiting independence in daily activities and mobility



# Stakeholder Engagement in Outcome Measure Development

Development of the SHAPE-UP

## Stakeholder Engagement in Outcome Measure Development

- Instruments to measure function should be developed in close cooperation with experts (clinician experts AND patients' lived experience) (de Vet et al., 2011)
- Tasks included in outcome measures should include both unilateral and bilateral activities, consider the quality of the movement completed, and be valid and reliable to the specific population targeted (Higgins et al., 2010)
- Children and youth with AMC describe their ability to participate based on their environment, their selection of leisure activities based on their physical abilities and interests (Elfassy et al., 2019)
- Scoping review on participation of children and youth with AMC demonstrated lack of information regarding participation such as sporting activities, employment, and education (Elfassy et al., 2020)



Elfassy et al.  
*Research Involvement and Engagement* (2023) 9:64  
<https://doi.org/10.1186/s40900-023-00479-6>

Research Involvement  
and Engagement

RESEARCH

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Stakeholder engagement  
in the development of an upper extremity  
outcome measure for children with rare  
musculoskeletal conditions

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Noémi Dahan-Oliel<sup>1,2</sup>



# Content Development

Development of the SHAPE-UP

## Stakeholder Engagement

- Nominal Group Technique with people with lived experience (parents)
- Three-round Delphi survey (clinicians)

*Stakeholder engagement in the development of an upper extremity outcome measure for children with rare musculoskeletal conditions (Research Involvement and Engagement, 2023)*



# Outcome Measures Currently in Use

Development of the SHAPE-UP

## What outcome measures are currently being used in rehabilitation for children with AMC?

- PODCI, PedsQL, PROMIS, ROM, Strength (Oishi et al., 2019)
- AIMS, ASKp, PODCI, Wee-FIM, ROM, Strength (Elfassy et al., 2020)
- ROM, Strength, post-op protocols (Wagner et al., 2019)





- **HOW?**
- **Which joint or muscle is limiting function?**
- **Unable to compare to normative data**

Original Article

**Pediatric Performance-Based Outcome Measures for Upper Extremity Function: A Scoping Review and Linking to the International Classification of Functioning, Disability, and Health**

Mesures des résultats basées sur le rendement pour la fonction du membre supérieur en réadaptation pédiatrique : étude de portée et arrimage avec la Classification internationale du fonctionnement, du handicap et de la santé (CIF)

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Canadian Journal of Occupational Therapy  
2024, Vol. 91(4) 325-373  
DOI: 10.1177/00084174241233513

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# Content Development

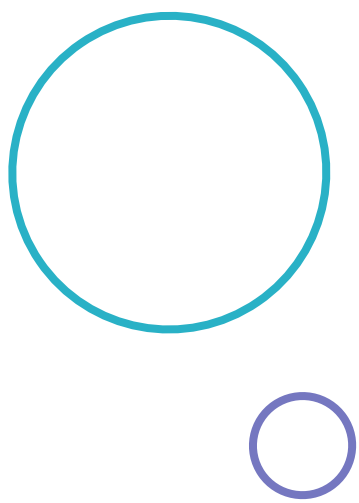
Development of the SHAPE-UP

## Scoping Review

- Identified pediatric performance-based outcome measures for evaluating UE function
- Linked items/constructs to the International Classification of Functioning, Disability & Health (ICF)
- Majority of items in these outcome measures were linked to mobility and fine hand use by the ICF



# Shriners Hospital Arthrogryposis Pediatric Evaluation SHAPE-UP



Development of the SHAPE-UP



Shriners Hospital Arthrogryposis Pediatric Evaluation – UPper Extremity (SHAPE-UP)		
Task #	Task Prompt	Joints focused
1	Pick up a Cheerio, bring it to your mouth, place it back down in front of you, and let it go.	Thumb, Fingers, Elbow
2	Grasp a water bottle, bring it to your mouth, place it back down in front of you, and let it go.	Thumb, Fingers, Elbow
3	Open the jar, pour out a few beads, pick up the bead and string 3 beads onto a string, and close the jar.	Thumb, Fingers, Wrist
4	Pick up the marker, draw a circle or lines or write you name on this piece of paper with the marker, then cut or snip the paper (2 or 3 cuts in the paper) using the scissors.	Thumb, Fingers, Wrist
5	Pick up the fork, stick it into the Play-Doh, and bring the fork to your mouth.	Wrist, Forearm
6	Put on and take off a t-shirt.	Elbow, Shoulder
7	Put on this vest, attach the zipper, and pull the zipper all the way up and then all the way back down.	Elbow, Shoulder
8	Put on this sock.	Thumb, Elbow
9	Throw the tennis ball underhand. Repeat task, throwing ball overhead.	Elbow, Shoulder
10	Throw the dodgeball overhead with both hands.	Elbow, Shoulder
11	Pull down the shorts, reach toilet paper, and pull the shorts back up. your bottom from the back while holding 2 squares of toilet paper, also reach between your legs from the front while holding 2 squares of	Thumb, Wrist, Elbow

## Original 5-point Task Completion Scale

0	The child is unable to attempt to complete or complete any part of the subtask.
1	The child scores a 1 when they are attempting to complete the task with minimal success.
2	The child scores a 2 when they can actively partially complete the task with any physical assistance.
3	The child scores a 3 when they can passively complete the task when using external aid (not physical aid from a person), compensatory movements, or a device.
4	The child scores a 4 when they can actively complete the task without any external aid or compensatory movement.

# SHAPE-UP PILOT STUDY

Development of the SHAPE-UP



Study ID: \_\_\_\_\_  
 DOB: \_\_\_\_\_  
 Date of completion: \_\_\_\_\_

9. Throw the tennis ball underhand. Repeat task, throwing ball overhead.  
 Select R or L in column "Arm" to describe the side the child primarily used throughout each subtask.  
 (Evaluator hands ball to child.)

Arm	Item	Score	Analysis of Joint Motion and Position					
			Thumb	Fingers	Wrist	Forearm	Elbow	Shoulder
<input type="checkbox"/> R <input type="checkbox"/> L	Wind up in preparation to throw ball underhand (into extension position to throw)	NA 0 1 2 3 4			<input checked="" type="checkbox"/> Flexion <input type="checkbox"/> Neutral <input type="checkbox"/> Extension	<input type="checkbox"/> Pronation <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Supination	<input type="checkbox"/> No elbow extension <input type="checkbox"/> Partial elbow extension <input checked="" type="checkbox"/> Full elbow extension	<input type="checkbox"/> Flexion <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Extension
<input type="checkbox"/> R <input type="checkbox"/> L	Throw ball underhand	NA 0 1 2 3 4			<input checked="" type="checkbox"/> Extension <input type="checkbox"/> Neutral <input type="checkbox"/> Flexion	<input checked="" type="checkbox"/> Pronation <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Supination	<input type="checkbox"/> No elbow extension <input type="checkbox"/> Partial elbow extension <input checked="" type="checkbox"/> Full elbow extension	<input type="checkbox"/> Extension <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Flexion
<input type="checkbox"/> R <input type="checkbox"/> L	Wind up in preparation to throw ball overhead (into flexion position at the shoulder to throw)	NA 0 1 2 3 4			<input type="checkbox"/> Flexion <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Extension	<input type="checkbox"/> Supination <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Pronation	<input type="checkbox"/> No elbow flexion <input checked="" type="checkbox"/> Partial elbow flexion <input checked="" type="checkbox"/> Full elbow flexion	<input type="checkbox"/> Extension <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Flexion
<input type="checkbox"/> R <input type="checkbox"/> L	Throw ball overhead (into extension posture at the shoulder when you release)	NA 0 1 2 3 4			<input checked="" type="checkbox"/> Extension <input type="checkbox"/> Neutral <input type="checkbox"/> Flexion	<input type="checkbox"/> Supination <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Pronation	<input type="checkbox"/> No elbow extension <input type="checkbox"/> Partial elbow extension <input checked="" type="checkbox"/> Full elbow extension	<input type="checkbox"/> Flexion <input type="checkbox"/> Neutral <input checked="" type="checkbox"/> Extension

Remarks: please describe any compensatory movements: use of opposite arm, a particular starting position (standing versus sitting) or assistive device such as sock aid, larger zipper pull, etc)

# Methods/Results

Development of the SHAPE-UP

**Sample:** children with AMC (n=92; female=58%; age:1-21 years)

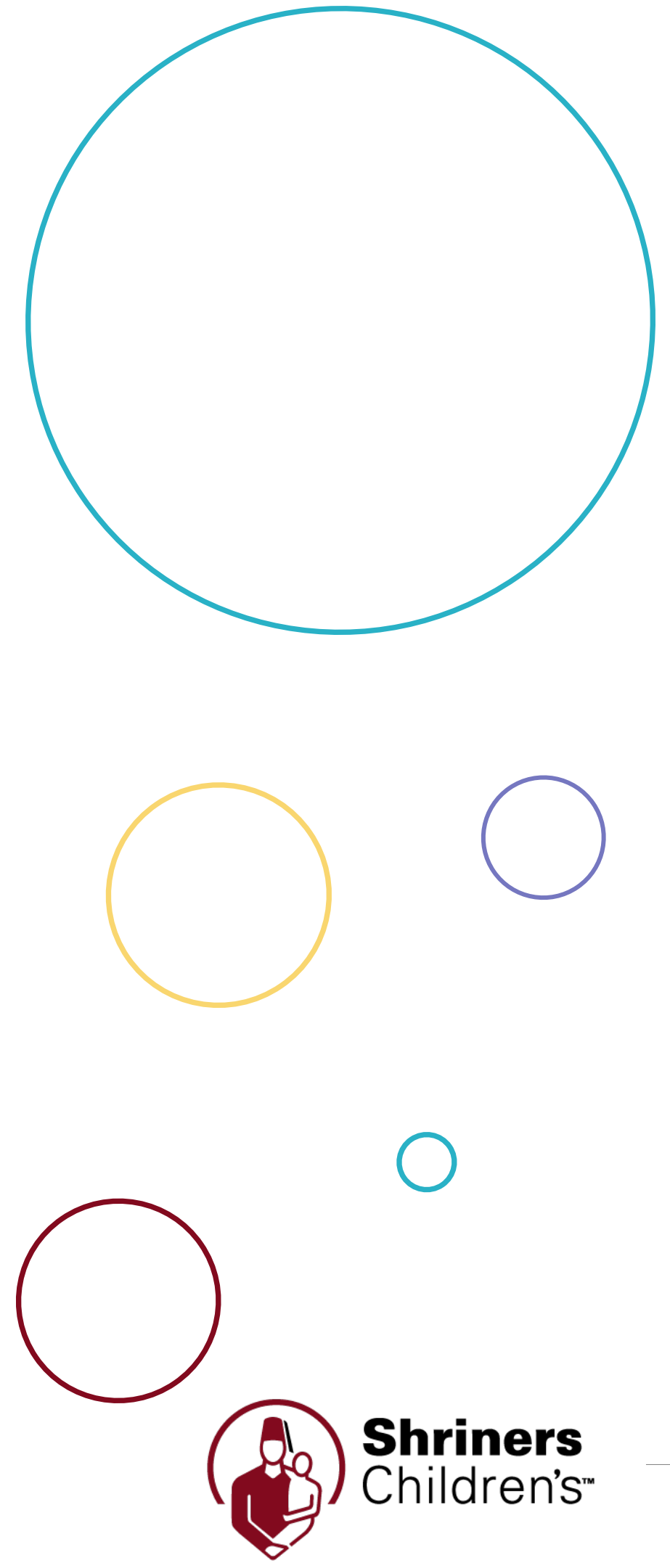
**Methods:** Pilot SHAPE-UP administered across six Shriners' sites  
- Canada, USA

**Data Analysis:** Data were assessed for 'goodness of fit' of the model (task completion) using Rasch analysis  
-evaluate its content and construct validity, reliability, appropriateness of response categories.

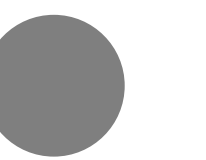
- Sample size was proportionately large for this rare condition  
-Insufficient to definitively establish unidimensionality and internal consistency of the items.

# SHAPE-UP PILOT: Revisions

Development of the SHAPE-UP



- Task Completion Scale revised
  - 5 scores → 3 scores (0-2)
  - 11 tasks → 7 tasks
- Adapted Equipment and Physical Assistance
- Compensations








Study ID: \_\_\_\_\_

DOB: \_\_\_\_\_

Date of completion: \_\_\_\_\_

Arm	Subtask	Task Completion		Subtask Analysis of Joint Motion and Position							
				Prehension	Limitation				Adaptive Equipment/ Physical Assistance	Compensatory Movements	
					Wrist	Forearm	Elbow	Shoulder			
<input type="checkbox"/> R <input type="checkbox"/> L	Attach zipper 	0	1	2	<input type="checkbox"/> Scissor between the fingers <input type="checkbox"/> Fingers bending to back of thumb <input type="checkbox"/> Thumb to palm <input type="checkbox"/> Pinch <input type="checkbox"/> Other:	<input type="checkbox"/> Fixed in flexion, minimal movement <input type="checkbox"/> Flexed, some movement <input type="checkbox"/> Fixed in extension, minimal movement <input type="checkbox"/> Extended, some movement <input type="checkbox"/> No limitation	<input type="checkbox"/> Overpronation <input type="checkbox"/> Oversupination <input type="checkbox"/> No limitation	<input type="checkbox"/> Fixed in extension <input type="checkbox"/> Some flexion/extension <input type="checkbox"/> Fixed in flexion <input type="checkbox"/> No limitation	<input type="checkbox"/> Excessive internal rotation <input type="checkbox"/> No limitation	<input type="checkbox"/> If yes, describe:	<input type="checkbox"/> Bimanual: <input type="checkbox"/> Back of hand <input type="checkbox"/> Palm to palm <input type="checkbox"/> Crossover palm to palm <input type="checkbox"/> Neck flexion <input type="checkbox"/> Trunk <input type="checkbox"/> Arm swing <input type="checkbox"/> Stabilize using surface/body <input type="checkbox"/> Other:
<input type="checkbox"/> R <input type="checkbox"/> L	Pull zipper halfway to chest  	0	1	2	<input type="checkbox"/> Fixed in flexion, minimal movement <input type="checkbox"/> Flexed, some movement <input type="checkbox"/> Fixed in extension, minimal movement <input type="checkbox"/> Extended, some movement <input type="checkbox"/> No limitation	<input type="checkbox"/> Overpronation <input type="checkbox"/> Oversupination <input type="checkbox"/> No limitation	<input type="checkbox"/> Fixed in extension <input type="checkbox"/> Some flexion/extension <input type="checkbox"/> Fixed in flexion <input type="checkbox"/> No limitation	<input type="checkbox"/> Excessive internal rotation <input type="checkbox"/> No limitation	<input type="checkbox"/> If yes, describe:	<input type="checkbox"/> Neck flexion <input type="checkbox"/> Trunk <input type="checkbox"/> Arm swing <input type="checkbox"/> Stabilize using surface/body <input type="checkbox"/> Other:	

# Next steps: SHAPE-UP + AI=?

## Scoring Workflow

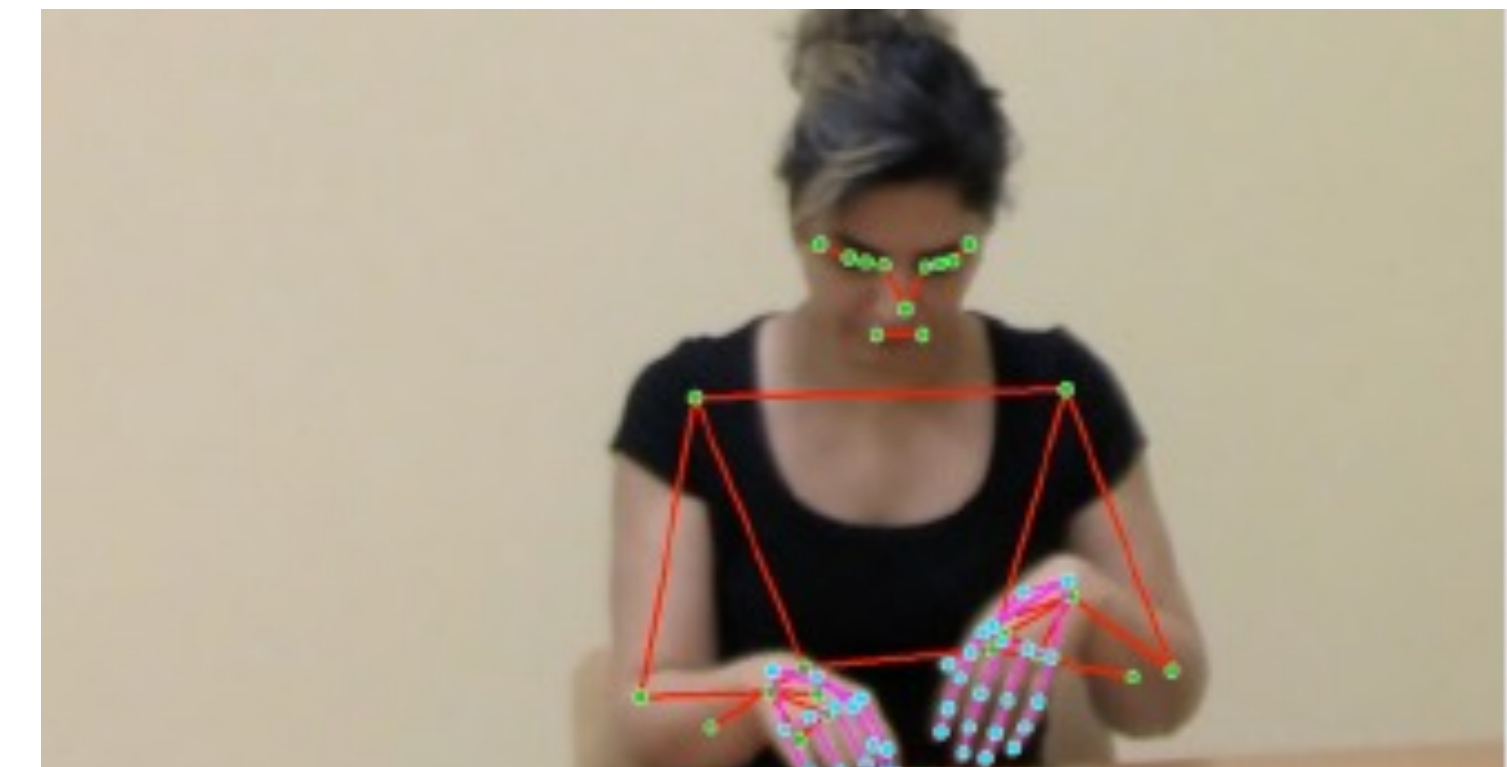
- Features **101 video assessments** of upper limb tasks and sources from across 6 Shriners Hospitals across the U.S.
- Angle estimation algorithm assigns a **score of 0-2** to each child based on depicted angle measurements for all upper-arm joints.

## Model Training Workflow

- Use of MediaPipe Pose **convolutional neural networks (CNNs)** - **objective** is to assign 3D coordinates to select frames of a given video.
- Shriners' psychometric validation framework evaluates inter- and intra-rater reliability using paired t-tests, which compare **clinician ratings and intraclass correlation coefficients (ICC)** to quantify universal scoring consistency.
- **Pearson correlation coefficients (r)** between algorithm-generated SHAPE-UP scores and clinician ratings will be calculated, categorized as excellent ( $r \geq 0.80$ ), good ( $0.60 \leq r < 0.80$ ), fair ( $0.40 \leq r < 0.60$ ), or poor ( $r < 0.40$ ).

## Broader Projected Outcomes:

- Automated scoring of SHAPE-UP tasks
- Consistency across raters and timepoints
- Scalability for use across clinics and research centers
- Remote use potential for underserved or rural patients



# Current Progress

Development of the SHAPE-UP

- IRB acceptance at all 6 participating sites
- Signed agreement with University of North Texas
- 3 participants recruited, test-retest and utility interviews completed
- Addition of another camera to get 3D images to capture compensations of the trunk

# Clinical Relevance of SHAPE-UP

Development of the SHAPE-UP



- Characterizes and documents functional status of the UE in children and youth with AMC
  - Addresses the uniqueness of children with AMC
  - Task completion
  - Joint analysis and compensations
  - Tasks consist of UNILATERAL, BILATERAL and BIMANUAL UE performance
  - Items span the complete range of difficulty

# Summary

Development of the SHAPE-UP

- This AMC-specific outcome measure informs clinical decision-making in the treatment of the upper extremity.
- The inclusion of clinicians and others with lived experience in the process of test item generation ensures that the SHAPE-UP is relevant and meaningful to the users.
- SHAPE-UP provides a standardized description of task completion and joint analysis to guide individualized treatment planning and assessment of treatment outcomes in AMC.

# Thank You!

## Funding



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