Expressive Communication Skills of Patients with Angelman Syndrome **Tayler Simonds, BA**

BACKGROUND

Angelman syndrome (AS) is a rare neurodevelopmental disorder which is caused by one of four known genetic mechanisms, all of which disrupt the UBE3A region of the maternally inherited chromosome 15. It is associated with severe developmental delay, movement and balance disorders, behavioral characteristics (e.g., frequent laughter), and communication difficulties, particularly in expressive communication (Quinn & Rowland, 2017). The Carolina Institute for Developmental Disabilities (CIDD) has housed an Angelman syndrome clinic since 2012.

The Communication Matrix (CM) is a free online tool which assesses expressive communication skills. It accepts nonverbal behavior and any modality, including speech, sign language, and AAC. Based on a caregiver, educator, or provider's answers to questions, it organizes skills into four reasons to communicate (to refuse, obtain, interact socially, and exchange information) and seven levels of communication that typically occur between birth and 2 years: preintentional communication (Levels 1 and 2), intentional presymbolic communication (Levels 3 and 4) and symbolic communication (Levels 5 and 6). Each of the 80 combinations of reasons and levels of communication is a "message," which is considered "mastered," "emerging," or "not used" depending on consistency of use (Quinn & Rowland, 2017).

OBJECTIVES

- Consolidate Communication Matrix data of former patients of the Angelman Syndrome Clinic at CIDD and analyze trends
- Compare with data collected nationally

PARTICIPANTS

Inclusion criteria:

- Former patient of Angelman Syndrome Clinic
- Caregiver consented to research data collection
- Communication Matrix was completed during visit

NC-LEND

SCHOOL OF MEDICINE

Sample Characteristics

- n = 58 (male = 29; female = 29)
- Age range: 1;5-34;0

METHODS

- Created a new CM database for patients who met inclusion criteria
- or 0 for "not used" for each of the 80 messages
- Conducted the following analyses: 3.
 - sample
 - Compared AS clinic data with Quinn & Rowland (2017) data using an independent samples t-test
 - compare with data accessible on CM "Shared Science" webpage
 - Calculated average total scores by genetic etiology and ran a one-way ANOVA to determine significance of mean differences

RESULTS

Total Scores Analysis

AS Clinic Total Scores		Quinn & Rov	Quinn & Rowland (2017) Total Scores			
Range	14 - 94	Range	5 - 131			
Mean	50.95	Mean	53			
SD	18.05	SD	25			
n = 58; age range: 1;5–34;0		n = 300; age range	n = 300; age range: 0;0-21;11			

0.738739	Analys
0.460552	sample
1.96665	 No sig sample
-	0.460552

Percent of Sample with Mastered Use

Level 1	A1 Expresses Discomfort		Ð	A2 xpresses Comfo	rt			A Expresses Int Per		
Pre-Intentional Behavior	100 94			98 97					00 4	
Level 2 Intertional	B1 Protests		32 es Action	Obtair	B3 ns More of Som	nething		B Attracts	4 Attention	
Behaviour	100 94	-	95 91		100 91			9	- 1	
Level 3 Unconventional Communication	C1 Refuses, Rejects	C2 Requests More Action	C3 Requests New Action	C4 Requests More Object	C5 Makes Choices	Có Requests New Object		C8 Requests Attention	C9 Shovis Affection	-
Communication	95 81	90 74	69 44	97 74	83 66	90 62		98 70	98 72	Top Bot
Level 4 Conventional Communication	C1 Refuses, Rejects 45 31	C2 Requests More Action 55 25	C3 Requests New Action 36 18	C4 Requests More Object 55 26	C5 Makes Choices 45 26	C6 Requests New Object 53 26		C8 Requests Attention 28 17	C9 Shows Affection 88 43	C10 Greet People 36 18
Level 5 Concrete Symbols	C1 Refuses, Rejects 5 14	C2 Requests More Action 3 11	C3 Requests New Action 10 9	C4 Requests More Object 9 12	C5 Makes Choices 19 14	Có Requests New Object 12 13	C7 Requests Absent Objects 2 10	C8 Requests Attention 0 1	C9 Shavis Affection 0 2	C10 Greets People 0 3
Level 6 Abstract Symbols	C1 Refuses, Rejects 16 6	C2 Requests More Action 24 11	C3 Requests New Action 3 6	C4 Requests More Object 26 10	C5 Makes Choices 0 7	Có Requests New Object 0 7	C7 Requests Absent Objects 0 5	C8 Requests Attention 7 4	C9 Shows Affection 2 2	C10 Greet People 14 8
Level 7 Language	C1 Refuses, Rejects 1 1	C2 Requests More Action 1 2	C3 Requests New Action 1 2	C4 Requests More Object 1 2	C5 Makes Choices	C6 Requests New Object 1 2	C7 Requests Absent Objects 1 1	C8 Requests Attention 0 1	C9 Shows Affection 0 1	C10 Greets People 1 2
	Refuse			ОЫ	tain					

Division of Speech and Hearing Sciences, The University of North Carolina at Chapel Hill

```
Accessed patients' CM profiles and coded scores of 2 for "mastered," 1 for "emerging,"
```

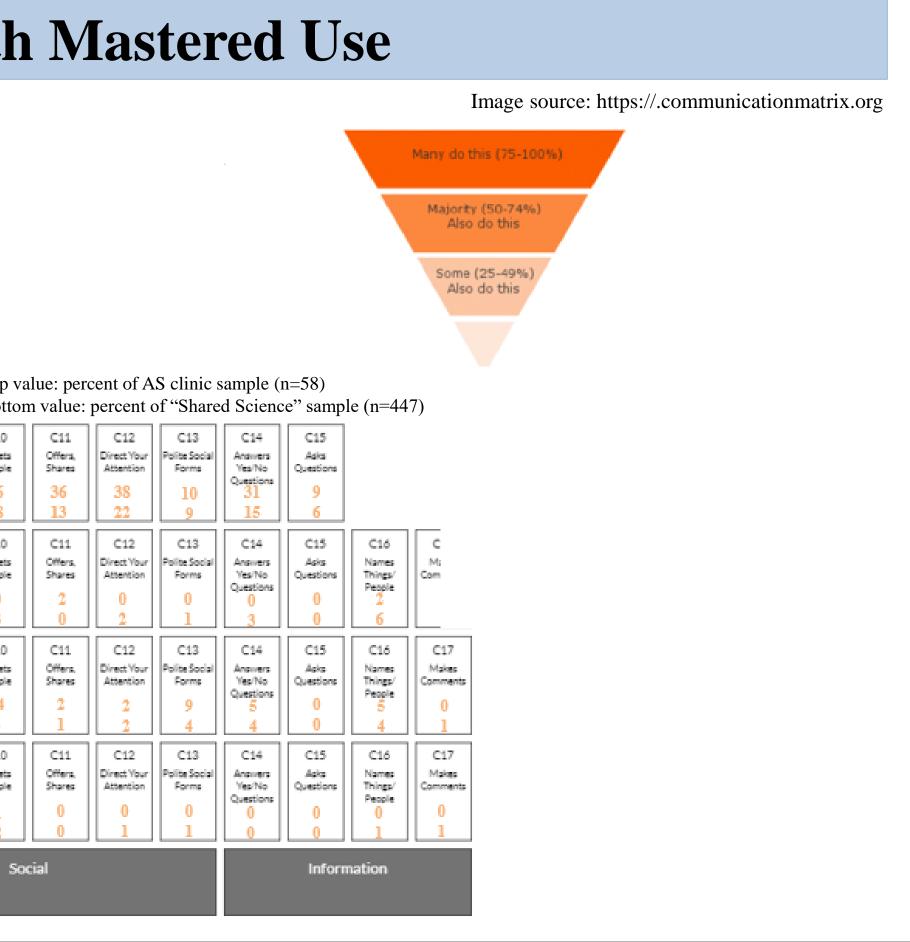
Calculated total scores and range, mean, and standard deviation for AS clinic

Calculated percent of AS clinic sample which had mastered each message to

Job, aze ranze. 0,0 21,1

vsis limited as t-test was conducted with only mean, le size and standard deviation gnificant differences between means of the two

ples with alpha = .05



Etiology Uniparent Imprinting UBE3A m Deletion (Other (5)

Between G Within Gro Total

		Mean Difference (l-			95% Confide	ence Interval
(I) VAR00002	(J) VAR00002	J)	Std. Error	Sig.	Lower Bound	Upper Bound
1.00	3.00	15909	6.76075	.981	-13.7525	13.4343
	4.00	27.75000	6.14076	.000	15.4032	40.0968
	5.00	10.75000	7.76752	.173	-4.8676	26.3676
3.00	1.00	.15909	6.76075	.981	-13.4343	13.7525
	4.00	27.90909	4.04705	.000	19.7719	36.0462
	5.00	10.90909	6.24532	.087	-1.6480	23.4661
4.00	1.00	-27.75000	6.14076	.000	-40.0968	-15.4032
	3.00	-27.90909	4.04705	.000	-36.0462	-19.7719
	5.00	-17.00000	5.56822	.004	-28.1957	-5.8043
5.00	1.00	-10.75000	7.76752	.173	-26.3676	4.8676
	3.00	-10.90909	6.24532	.087	-23.4661	1.6480
	4.00	17.00000	5.56822	.004	5.8043	28.1957

Communication Matrix is useful in educating caregivers and providers about the scope of communication Accessible for free online and accepts multiple modalities for symbolic communication, so it can be used to assess response to intervention (e.g., AAC) Data should be continually added to this new database and analyzed in different ways to develop a detailed understanding of the communication profiles associated with Angelman syndrome

Quinn, E.D., & Rowland, C. (2017). Exploring expressive communication skills in a cross-sectional sample of children and young adults with Angelman syndrome. American Journal of *Speech-Language Pathology, 26*, pp369-382.

Rowland, C., Ph.D. (2018). Communication Matrix. Retrieved April 11, 2018, from https://communicationmatrix.org/

Ruthy Xu, B.A. Cindy Navis, SLP

RESULTS (CONT.)

Etiology and Total Scores

	Ν	Mean
tal Disomy (1)	4	68.75
g (2)	1	31
nutation (3)	10	68.91
(4)	32	41
	5	58

One-Way ANOVA

	Sum of Squares	df	Mean Square	F	Sig.
roups	8227.783	3	2742.594	20.455	.000
ips	6435.659	48	134.076		
	14663.442	51			

*. The mean difference is significant at the 0.05 level

DISCUSSION

REFERENCES

ACKNOWLEDGEMENTS

Margaret DeRamus, M.S., CCC-SLP

Stephanie Fox, Ph.D. Shelby Waldron