

Expressive Communication Skills of Patients with Angelman Syndrome

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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

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
- Accessed patients' CM profiles and coded scores of 2 for "mastered," 1 for "emerging," or 0 for "not used" for each of the 80 messages
- Conducted the following analyses:
 - Calculated total scores and range, mean, and standard deviation for AS clinic sample
 - Compared AS clinic data with Quinn & Rowland (2017) data using an independent samples t-test
 - Calculated percent of AS clinic sample which had mastered each message to 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

| | |
|-------|---------|
| Range | 14 - 94 |
| Mean | 50.95 |
| SD | 18.05 |

n = 58; age range: 1;5-34;0

Quinn & Rowland (2017) Total Scores

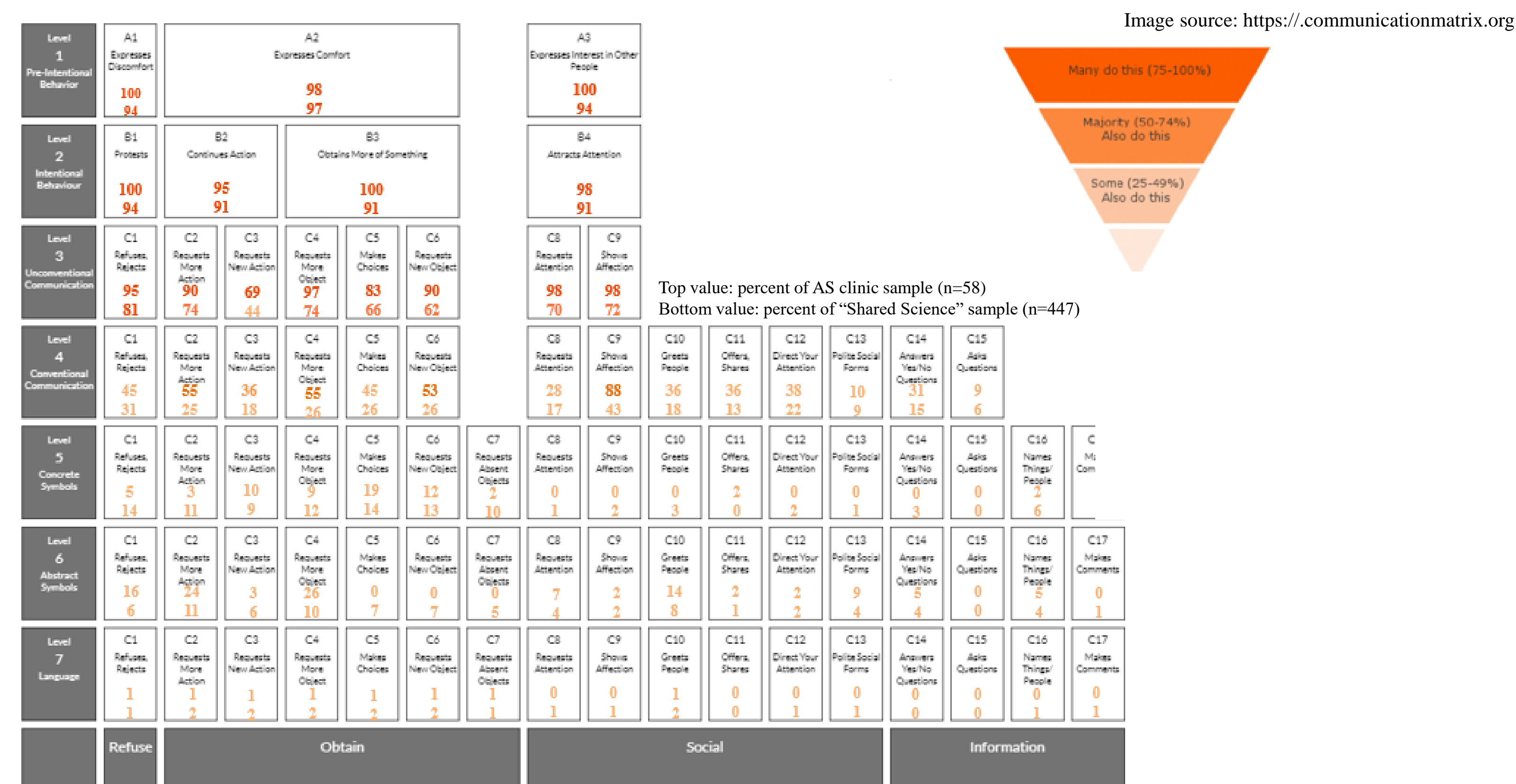
| | |
|-------|---------|
| Range | 5 - 131 |
| Mean | 53 |
| SD | 25 |

n = 300; age range: 0;0-21;11

| | |
|----------------|----------|
| t-statistic | 0.738739 |
| p value | 0.460552 |
| critical value | 1.96665 |

- Analysis limited as t-test was conducted with only mean, sample size and standard deviation
- No significant differences between means of the two samples with alpha = .05

Percent of Sample with Mastered Use



RESULTS (CONT.)

Etiology and Total Scores

| Etiology | N | Mean |
|------------------------|----|-------|
| Uniparental Disomy (1) | 4 | 68.75 |
| Imprinting (2) | 1 | 31 |
| UBE3A mutation (3) | 10 | 68.91 |
| Deletion (4) | 32 | 41 |
| Other (5) | 5 | 58 |

One-Way ANOVA

| | Sum of Squares | df | Mean Square | F | Sig. |
|----------------|----------------|----|-------------|--------|------|
| Between Groups | 8227.783 | 3 | 2742.594 | 20.455 | .000 |
| Within Groups | 6435.659 | 48 | 134.076 | | |
| Total | 14663.442 | 51 | | | |

| | | Mean Difference (I-J) | | Std. Error | Sig. | 95% Confidence Interval | |
|--------------|--------------|-----------------------|---------|------------|------|-------------------------|-------------|
| (I) VAR00002 | (J) VAR00002 | | | | | 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 |

*. The mean difference is significant at the 0.05 level.

DISCUSSION

- 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

REFERENCES

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/>

ACKNOWLEDGEMENTS

Margaret DeRamus, M.S., CCC-SLP
Ruthy Xu, B.A.
Cindy Navis, SLP

Stephanie Fox, Ph.D.
Shelby Waldron

NC-LEND



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