AMERICAN JOURNAL ON INTELLECTUAL AND DEVELOPMENTAL DISABILITIES
2021, Vol. 126, No. 3, 260–265

DOI: 10.1352/1944-7558-126.3.260

Examination of the ADOS-2 Expressive Language Score in Fragile X Syndrome

Heather Fielding-Gebhardt, Shelley L. Bredin-Oja, and Steven F. Warren

Abstract

The development of an expressive language score for people with autism based on the ADOS-2 was recently reported by Mazurek et al. (2019). The current study examined the construct validity of the ADOS-2 expressive language score (ELS) in a sample of adolescents with fragile X syndrome ($n = 45$, 10 girls), a neurodevelopmental disorder with high rates of autism symptomology. The ADOS-2 ELS showed strong convergent validity with multiple assessments of expressive language, receptive language, and nonverbal cognition. Divergent validity was demonstrated between the expressive language score and chronological age, symptoms of anxiety/depression, and rule-breaking behaviors. This expressive language score is a promising measure of expressive language ability that can be used in research when other language assessments are unavailable.

Keywords: measurement, ADOS-2, expressive language

Recently, Mazurek et al. (2019) developed an expressive language score (ELS) based on the Autism Diagnostic Observation Schedule-2 (ADOS-2; Lord et al., 2012). The ELS is derived from the A1 item on the ADOS-2 (i.e., Overall Level of Non-Echoed Spoken Language), a rating of the individual's expressive language ability, which is based on the complexity of spontaneous language produced by the individual during the ADOS-2 assessment. This item is included on all modules; therefore, the derived ELS spans ages and abilities and may be an efficient metric of expressive language from this widely used measure of autism symptomology.

In their report, Mazurek et al. (2019) showed that the ELS was moderately to strongly correlated with most measures of expressive language, receptive language, and nonverbal cognitive ability ($r_{\text{expressive}}$: 0.51 to 0.89; $r_{\text{receptive}}$: 0.47 to 0.67; $r_{cognitive}$: 0.20 to 0.54, all $p < 0.01$). The authors demonstrated convergent validity between the ELS and parent report, clinician-rated, and direct assessment of expressive language. In their analyses, the ELS was more strongly correlated with expressive language measures than receptive and nonverbal measures, potentially indicating divergent validity between the ELS and measures of receptive language and nonverbal cognitive ability. However, divergent validity relies on non-significant and/or negative correlations between scales that are intuitively dissimilar and measure different concepts. Receptive language and expressive language are generally not considered to be dissimilar or opposing, so there are likely other constructs that would better demonstrate divergent validity with the ELS. Because expressive language is a fundamental skill, selection of constructs for divergent validity should consider those in which expressive language is not expected to differentiate severity. Two such constructs may be anxiety and rule-breaking behavior.

This short report extends the construct validity and utility of the ELS to a sample of adolescents with fragile X syndrome (FXS) by demonstrating convergent and divergent validity. FXS is a neurodevelopmental disorder that presents with high rates of comorbid autism. Estimates suggest that as many as 75 to 97% of males and 25% of females with FXS meet ADOS-2 criteria for autism or autism spectrum disorder

DOI: 10.1352/1944-7558-126.3.260

(Haebig et al., 2020; Klusek et al., 2014). Expressive and receptive language abilities are correlated in people with FXS and autism (Brady et al., 2014; Haebig & Sterling, 2017), and both language domains are generally delayed. Finally, people with FXS have elevated levels of anxiety and challenging behaviors such as rule-breaking behaviors, which are associated with higher autism symptomology (Hardiman & McGill, 2018).

Methods

Participants

Forty-five adolescents with FXS (35 boys, 10 girls) completed assessments of language, cognition, challenging behaviors, and autism symptomology with examiners during in-home visits as part of an ongoing longitudinal study (Brady et al., 2014). The FXS genotype was confirmed in all participants through blood analysis. Participants represent a sample of convenience and have been participating in the ongoing longitudinal study since the early 2000s. The average age of participants was 16.89 years. See Table 1 for participant demographics.

Measures and Procedure

Biological mothers of the adolescents completed the interview form of the Vineland Adaptive Behavior Scales (VABS-II; Sparrow et al., 2005) and the Child Behavior Checklist (CBCL; Achenbach & Rescorla, 2001). Adolescents completed the Peabody Picture Vocabulary Test (PPVT-4; Dunn & Dunn, 2007), Expressive Vocabulary Test (EVT-2; Williams, 2007), Leiter-Revised Brief IQ Scale (Leiter-R; Roid & Miller, 1997), and the ADOS-2 with assessors trained to be research reliable. The VABS-II communication scales were used to assess parent-reported expressive and receptive language and the CBCL was used to index anxiety and rule-breaking behavior. See Table 1 for information on the distribution of ADOS-2 modules and Table 2 for participant characteristics and mean scores. Raw scores were used for the VABS-II scales. For the Leiter-R, PPVT-4, and EVT-2, the growth scale value score was used in the analyses because this score best represents true ability on a given construct. The growth scale value score is along an interval scale, which provides an estimate of relative ability, and helps avoid floor effects, which are common in studies of people with developmental disabilities.

Table 1
Participant Demographics and Characteristics

Level	*N*	Percent
Sex		
Male	35	78%
Female	10	22%
ADOS-2 Module		
Module 1: Pre-Verbal/Single Words	12	27%
Module 2: Phrase Speech	12	27%
Module 3: Fluent Speech (Child/Adolescent)	11	24%
Module 4: Fluent Speech (Adolescent/Adult)	10	22%
Maternal Education		
No Bachelor's	16	35.5%
Bachelor's	16	35.5%
Higher Ed. Degree	13	29%
Family Income		
< $30,000	5	11%
$30,000 to $80,000	13	29%
>$80,000	27	60%
Race		
White	42	93%
African American	3	7%
Ethnicity		
Non-Hispanic	43	96%
Hispanic	2	4%

Note. ADOS-2 = Autism Diagnostic Observation Schedule, 2nd Ed.

Mean length of utterance in morphemes (MLUm), number of different words (NDW), and number of communication units (C-units), defined as a main clause and all of its subordinate clauses, were obtained from language samples that were collected during 30 minutes of mother-child interaction. The language sample was collected during three mother-child interactions: making a puzzle, playing with an iPad, and preparing and eating a snack. For each context, the mother and child were instructed to complete the task together and that the examiners were interested in how they communicated with one another.

Language transcripts were transcribed by trained lab personnel with backgrounds in linguistics or speech-language pathology. Each file was transcribed by a primary coder, then reviewed by a secondary coder who made note of any discrep-

DOI: 10.1352/1944-7558-126.3.260

Table 2
Mean Scores and Ranges for Language, Behavior, and Autism Symptomology Measures

	N	Mean (*SD*)	Range
Chronological Age in Years	45	16.89 (1.70)	12.42–19.08
CBCL Anxious/depressed Raw Score	45	4.18 (3.18)	0–13
CBCL Rule-Breaking Behavior Raw Score	45	1.64 (1.81)	0–6
ADOS-2 Total Score	45	12.44 (5.45)	3–21
Leiter-R Growth Scale Value Score	42	460.81 (19.26)	404–502
PPVT-4 Growth Scale Value Score	43	139.00 (34.56)	84–204
EVT-2 Growth Scale Value Score	38	145.29 (27.61)	76–200
Number of Utterances	45	302.40 (173.80)	31–685
Mean Length of Utterance in Morphemes	45	2.35 (.85)	1–3.67
Number of Different Words	45	142.56 (87.40)	1–269
VABS-II Receptive Raw Score	45	32.16 (5.03)	24–40
VABS-II Expressive Raw Score	45	75.60 (26.26)	28–108
VABS-II Communication Raw Score	45	130.36 (37.96)	58–198

Note. CBCL = Child Behavior Checklist; ADOS-2 = Autism Diagnostic Observation Schedule, 2nd edition; PPVT-4 = Peabody Picture Vocabulary Test, 4th edition; EVT-2 = Expressive Vocabulary Test, 2nd edition; VABS-II = Vineland Adaptive Behavior Scales, 2nd edition.

ancies. All changes to the original transcript were resolved by consensus. When consensus could not be reached, the utterance was coded as unintelligible and excluded from the analyses. Transcripts were analyzed using the Systematic Analysis of Language Transcripts software (SALT: 2016). Research lab personnel who were trained in SALT procedures separated the utterances into C-units and marked all bound morphemes according to the SALT manual (Miller & Iglesias, 2016).

MLUm is a measure of expressive morphosyntax, an index of language development. NDW is a naturalistic measure of expressive vocabulary, and number of C-units is a measure of overall talkativeness. The ELS for each participant was derived from the A1 item on the ADOS-2, as described by Mazurek et al. (2019). The ELS is assigned into one of eight levels with higher levels indicating greater expressive language abilities. The distribution of ELS scores is provided in Table 3.

Missing Data

Two males were unable to complete the entire 30 minutes of mother-child interaction contexts due to challenging behavior; therefore, their language sample data comes from 20 and 25 minutes of mother-child interaction, respectively. Additionally, there were two male and one female participant(s) who did not complete the Leiter-R due to challenging behaviors, and two males who did not complete the PPVT-4 for the same reason. Seven participants, including one female, were unable to complete the EVT-2 due to limited expressive language skills. The sample size for each measure is reported in Table 2.

Results

There was considerable between-person variability on all measures, which reflects the heterogeneity seen in FXS. The distribution of ELS was spread across levels, with a greater concentration in the higher scores. To test for convergent and divergent validity, Spearman's rank correlations were calcu-

Table 3
ADOS-2 Expressive Language Score Distribution

ADOS-2 ELS	*n* (%)
1	5 (8.9)
2	3 (5.4)
3	1 (1.8)
4	6 (10.7)
5	4 (7.1)
6	9 (16.1)
7	6 (10.7)
8	11 (19.6)
Total	45 (100)

Note. ADOS-2 = Autism Diagnostic Observation Schedule, 2nd edition.

DOI: 10.1352/1944-7558-126.3.260

Table 4
Correlations Demonstrating Convergent and Divergent Validity

Measure	*N*	Spearman's Rho	*P*-value
Convergent Measures			
Parent-Report			
VABS-II Communication Raw Score	45	0.79	0.00
VABS-II Expressive Raw Score	45	0.82	0.00
VABS-II Receptive Raw Score	45	0.62	0.00
Direct Assessment			
PPVT-4 Growth Scale Value Score	43	0.70	0.00
EVT-2 Growth Scale Value Score	38	0.64	0.00
Leiter-R Growth Scale Value Score	42	0.61	0.00
ADOS-2 Total Score	45	−0.53	0.00
Language Sample			
Mean Length of Utterance in Morphemes	45	0.79	0.00
Number of Different Words	45	0.72	0.00
Total Number of Utterances	45	0.50	0.00
Divergent Measures			
CBCL Anxiety/Depression Raw Score	45	0.14	0.35
CBCL Rule-breaking Behavior Raw Score	45	−0.18	0.23
Chronological Age (Years)	45	−0.15	0.32

Note. VABS-II = Vineland Adaptive Behavior Scales, 2nd edition; PPVT-4 = Peabody Picture Vocabulary Test, 4th edition; EVT-2 = Expressive Vocabulary Test, 2nd edition; ADOS-2 = Autism Diagnostic Observation Schedule, 2nd edition; CBCL = Child Behavior Checklist.

lated. Spearman's rank correlation, also called Spearman's rho, is used when assessing the correlation between ordinal, rank, or interval variables. The ELS is an ordinal variable, and all assessments that utilized growth scale value scores are interval variables. Evidence for convergent validity was demonstrated when the correlations were strong and significant. Divergent validity was demonstrated by correlations that were weak and not significant.

Convergent Validity

Convergent validity was demonstrated between the ELS and parent-reported, direct-assessment, and language sample measures of expressive language, receptive language, and nonverbal cognition, see Table 4. The VABS-II communication raw score was strongly and significantly associated with the ELS. Additionally, VABS-II expressive raw score and receptive raw score were both strongly and significantly associated with the ELS. When testing for the relative strength of correlation, analyses indicated that the VABS-II expressive score was significantly more strongly correlated with the ELS than was the VABS-II receptive score ($Z = 2.60$, $p < 0.001$). These findings replicate those reported by Mazurek et al. (2019), suggesting good convergent validity between parent-reported expressive language and the ELS. Similarly, both the EVT-2 and PPVT-4 growth scale scores were significantly correlated with the ELS. When testing for the relative strength of correlation with the ELS, the analysis indicated that there was no difference between the PPVT-4 and the EVT-2 ($Z = -1.21$, $p > 0.05$). Both MLUm and NDW were strongly and significantly associated with the ELS, see Table 4. This suggests good convergent validity between the ELS and MLUm and NDW. Total number of C-units was also significantly correlated with the ELS, however the strength of the correlation was moderate. Finally, the Leiter-R Brief IQ scale (Roid & Miller, 1997) was used to assess nonverbal cognition. Nonverbal cognitive growth scale value scores were significantly correlated with the ELS.

Divergent Validity

Chronological age, anxiety, and rule-breaking behaviors were tested as divergent validity measures. All were not significantly associated with the

DOI: 10.1352/1944-7558-126.3.260

ELS, suggesting that the ELS best measures language, rather than non-linguistic constructs.

Discussion

This report extends findings of Mazurek et al. (2019) by assessing the construct validity of the ELS in a sample of adolescents with FXS. As in the original article, parent-reported expressive language and directly-assessed expressive vocabulary were highly correlated with the ELS. Direct assessment of receptive language was more highly correlated with the ELS than direct assessment of expressive language, however the difference between receptive language and expressive language was not significant. The ELS was not correlated with chronological age, anxiety, or rule-breaking behavior.

The inclusion of expressive language measures (i.e., MLUm, NDW, and number of C-units) obtained through naturalistic language samples and extensive analysis extends and strengthens the construct validity of the ELS. Naturalistic expressive language obtained from lengthy language samples can be difficult and time-consuming to collect and transcribe. The ELS may serve as a proxy measure of expressive syntax and vocabulary, as indicated by the strong correlations between it and MLUm and NDW. The data reported here suggest that this measure may be an efficient way of assessing expressive language ability in adolescents with FXS when no other language measures are available or when time constraints prohibit more comprehensive language assessment. However, there are several limitations to the ELS. First, limited variability of ADOS-2 modules and participants' language levels may negatively impact the validity and utility of this measure because it is dependent on the ADOS-2 module administered. Additionally, the ELS may not be suited to longitudinal examination because it may lack sensitivity to small changes in language ability. Although the ADOS-2 ELS is unlikely to be sensitive enough to change over time for clinical decision-making or for intervention outcomes, it may, nonetheless, be a starting point. Further examination of the validity of the ELS in other neurodevelopmental disorders and across populations is needed, as are direct comparisons between populations.

As addressed by Mazurek et al. (2019), consideration of language ability is paramount in studies of children with neurodevelopmental disabilities. Given the widespread use of the ADOS-2, the creation of a language score derived from the ADOS-2 is logical and warranted. Although not a substitute for more comprehensive language assessment, this score may nonetheless be useful as a general estimate of language. The use of this relatively efficient score may have potential benefits in research studies of children with autism, FXS, or other neurodevelopmental disorders associated with autism symptomology.

References

Achenbach, T. M., & Rescorla, L. A. (2001). *Manual for the ASEBA School-Age Forms and Profiles*. University of Vermont, Research Center for Children, Youth, and Families. https://doi.org/10.1044/1092-4388(2013/12-0341)

Brady, N., Warren, S. F., Fleming, K., Keller, J., & Sterling, A. (2014). The effect of sustained maternal responsivity on later vocabulary development in children with Fragile X syndrome. *Journal of Speech Language Hearing Research*, *57*(1), 212–226. https://doi.org/10.1044/1092-4388(2013/12-0341)

Dunn, L. M., & Dunn, D. M. (2007). *Peabody Picture Vocabulary Test, 4th Edition*. Pearson Assessments.

Haebig, E., & Sterling, A. (2017). Investigating the receptive-expressive vocabulary profile in children with idiopathic ASD and comorbid ASD and fragile X syndrome. *Journal of Autism and Developmental Disorders, 47*(2), 260–274. https://doi.org/10.1007/s10803-016-2921-3

Haebig, E., Sterling, A., Barton-Hulsey, A., & Friedman, L. (2020). Rates and predictors of co-occurring autism spectrum disorder in boys with fragile X syndrome. *Autism and Developmental Language Impairments, 5*, 1–19. https://doi.org/10.1177/2396941520905328

Hardiman, R. L., & McGill, P. (2018). How common are challenging behaviors amongst individuals with fragile X Syndrome? A systematic review. *Research in Developmental Disabilities, 76*, 99–109. https://doi.org/10.1016/j.ridd.2018.02.020

Klusek, J., Martin, G. E., & Losh, M. (2014). Consistency between research and clinical diagnoses of autism among boys and girls with fragile X syndrome. *Journal of Intellectual*

DOI: 10.1352/1944-7558-126.3.260

Disability Research, 58(10), 940–952. https://doi.org/10.1111/jir.12121

Lord, C., Rutter, M., DiLavore, P. C., Risi, S., Gotham, K., & Bishop, S. (2012). *Autism Diagnostic Observation Schedule, Second Edition*. Western Psychological Services.

Mazurek, M. O., Baker-Ericzen, M., & Kanne, S. M. (2019). Brief report: Calculation and convergent and divergent validity of a new ADOS-2 expressive language score. *American Journal of Intellectual and Developmental Disabilities, 124*(5). https://doi.org/10.1352/1944-7558-124.5.438

Miller, J. & Iglesias, A. (2016). *Systematic analysis of language transcripts (SALT)*. [Computer Software]. SALT Software, LLC.

Roid, G. H., & Miller, L. J. (1997). *Leiter International Performance Scale- Revised: Examiner's Manual*. Stoelting.

Sparrow, S. S., Cicchetti, D., & Balla, D. A. (2005). *Vineland-II: Vineland Adaptive Behavior Scales, Second Edition*. Pearson.

Williams, K. T. (2007). *Expressive Vocabulary Test, 2nd Edition*. Pearson Assessments.

Received 3/19/2020, accepted 10/6/2020.

The authors wish to thank the families who have participated in the ongoing study. This work was supported by NIDCD T32 DC000052 and NICHD R01 HD084563.

Authors:
Heather Fielding-Gebhardt, Shelley L. Bredin-Oja, and **Steven F. Warren,** University of Kansas.

Correspondence concerning this article should be address to Heather Fielding-Gebhardt, University of Kansas, 1000 Sunnyside Avenue, Lawrence, KS 66045 (email: fielding.h@ku.edu).