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BRIEF PRACTICE

Treatment of Self-Injury in Bainbridge-Ropers Syndrome: Replication and Extensions of Behavioral Assessments

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Abstract

Bainbridge-Ropers syndrome (BRPS) is a rare and understudied developmental disorder associated with medical (e.g., sleep disruption) and behavioral (e.g., self-injury) challenges. There are no published treatments for BRPS. We targeted self-injury in a child with BRPS using a functional analysis and differential reinforcement, with several extensions to common procedures. Results present the first example of behavioral reduction for self-injury in BRPS.

- ABA strategies can reduce self-injury in BRPS
- Evaluating multiply maintained self-injury following identification of an automatic function is important.
- Sleep deficits may complicate assessment.

Keywords Bainbridge-Ropers syndrome · Self-injury · Challenging behavior · Functional analysis · Reinforcement · Competing stimulus assessment

Bainbridge-Roper syndrome (BRPS) is a rare genetic condition caused by an alteration of the ASXL3 gene, resulting in decreased protein production (UNIQUE, 2018). Approximately 32 cases exist in the published literature. BRPS is often associated with a specific craniofacial phenotype, intellectual disability, developmental delays, and issues with sleep, muscle tone, and feeding. Challenging behavior, such as self-injurious behavior (SIB), is also reported in several case reports of individuals with BRPS (e.g., Balasubramanian et al., 2017).

There are no published treatments for individuals with BRPS, including no examples of successful treatments for SIB or other challenging behavior. However, there is extensive literature on function-based treatments (based in applied behavior analysis [ABA]) for SIB in other developmental disabilities (Rooker et al., 2018). Similar strategies *should* be effective with children with BRPS, but evaluating such generality is important as comorbid symptoms common in BRPS could establish barriers to typical behavioral procedures. For example, medical complexities and sleep disruptions that are common in BRPS (UNIQUE, 2018) may serve as establishing operations for reinforcers maintaining SIB and complicate behavioral assessments (Kennedy & Meyer, 1996).

From a practical perspective, several medical insurers only cover ABA for autism spectrum disorder (ASD; e.g., Georgia Department of Community Health, 2020). Coverage gaps may be attributable to limited research on efficacy of ABA for individuals with developmental delays other than ASD (e.g., genetic conditions). Continued evidence supporting the generality of ABA across diagnoses is crucial to increase treatment accessibility. Thus, research focused on the efficacy of ABA strategies with individuals with diagnoses other than ASD may inform funders of the appropriateness of covering these types of services.

Method and Results

Participant and Setting

Bryan was a 12-year-old white male diagnosed with BRPS with Intellectual Disability, Profound (ASD rule-out). He attended ABA treatment for 30-hr a week for SIB. His SIB

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regularly resulted in bruising, swelling, and wounds and Bryan's parents used arm splints, Posey mitts, and a helmet for safety. Prior to the admission, several providers completed evaluations and ruled out medical causes of SIB.

Bryan exhibited no vocal communication and did not use alternative communication modalities. He walked with an abnormal gait and had poor muscle tone. Based on an interview with Bryan's parents, he experienced delayed sleep onset, frequent awakening, early final waking, and drowsiness. Bryan was prescribed risperidone, lorazepam, and clonidine for SIB; melatonin for sleep; and omeprazole for indigestion. Medications remained stable during this evaluation.

Behavioral therapists conducted sessions in a padded treatment room equipped with seating, condition-specific materials, and a one-way observation window.

Response Measurement and Interobserver Agreement

Bryan engaged in self-biting (teeth contacting his body), self-pinching (two of his fingers contacting his body and grasping his skin), self-hitting (hand contacting his head/body from ≥ 6 in), and head-to-surface hitting (head contacting a surface from ≥ 6 in). Parent report of common antecedents/consequences suggested SIB likely was maintained by automatic and social reinforcement. We combined all forms of SIB into one SIB category based on unstructured observations and parent reports suggesting these behaviors were likely in the same response class.

We scored the frequency of SIB using a computerized system and analyzed SIB as a rate. We collected interobserver agreement (IOA) data for 31.25% of the functional analysis (FA), 32.58% of the treatment evaluation, and 38.89% of the competing attention assessment. We partitioned sessions into 10-s intervals and calculated mean-count-per-interval IOA. IOA was 97.44% for the FA, 87.51% for treatment, and 97.52% for the attention assessment. We scored independent and prompted Functional Communication Responses (FCRs) per trial during functional communication training (FCT) and calculated the percent of trials with independent/prompted FCRs per session. We defined FCRs as physically handing the card to the therapist (card-exchange; during the first functional communication training attempt) and depressing a button for it to emit a recording (button press; during the second functional communication training and subsequent differential reinforcement training sessions). IOA was 100% across 65.79% of trials.

Procedures

We conducted several behavioral evaluations with Bryan. We outline the order of these assessments and treatments here (also see Supplemental Material) and explain each in detail in the following sections. First, we conducted preference assessments. Next, we screened for automatic reinforcement maintaining SIB and evaluated the impact of sensory extinction in the form of protective equipment with consecutive alone sessions conducted in a reversal design with and without arm splints. We then continued the functional analysis with social test conditions (attention and escape) and eventually added a condition to evaluate SIB maintained by access to sleep.

After assessment, we initiated a treatment evaluation. First, we conducted baseline sessions where a therapist delivered access to sleep and escape from demands contingent on SIB. Next, we implemented FCT to teach a communication response for a break (first with a card exchange and then with a button press). Following FCT, we initiated a DRA treatment with mands for a break reinforced on an FR1 schedule. After several sessions of the DRA treatment, we paused these sessions and conducted an attention competing items assessment. We then incorporated attention into the reinforcement interval of the DRA treatment and restarted these sessions. We evaluated this treatment with a reversal, returning to baseline before reinitiating the DRA treatment with attention.

Preference Assessments

We conducted paired-choice and free-operant preference assessments of leisure and edible items. Bryan did not consistently make selections and engaged in low levels of interaction across all items. This outcome was consistent with home observations and parent report of limited toy play skills.

Functional Analysis

We initially conducted an extended alone condition to screen for automatically maintained SIB (Querim et al., 2013). We conducted this assessment with and without protective equipment (PE; arm splints and helmet) in a reversal design. PE reduced the physical impact of SIB but did not prevent SIB from occurring. The purpose of the screening was to (1) identify whether SIB was automatically maintained and, if so, (2) identify if PE reduced SIB. Bryan engaged in lower rates of SIB when wearing PE ($M = .16$ RPM) and elevated rates without PE ($M = 1.03$). These results were replicated (Fig. 1, Phases 1–4), suggesting SIB was at least in part maintained by automatic reinforcement.

Next, we evaluated whether Bryan's SIB also was maintained by social reinforcers with a multielement FA with attention, escape, and alone conditions. Therapists implemented contingencies as described by Iwata et al. (1994) with two modifications. First, in the attention condition the therapist provided 30 s of attention matched to commonly observed caregiver responses to SIB. Second, therapists did not implement a change-over-delay for

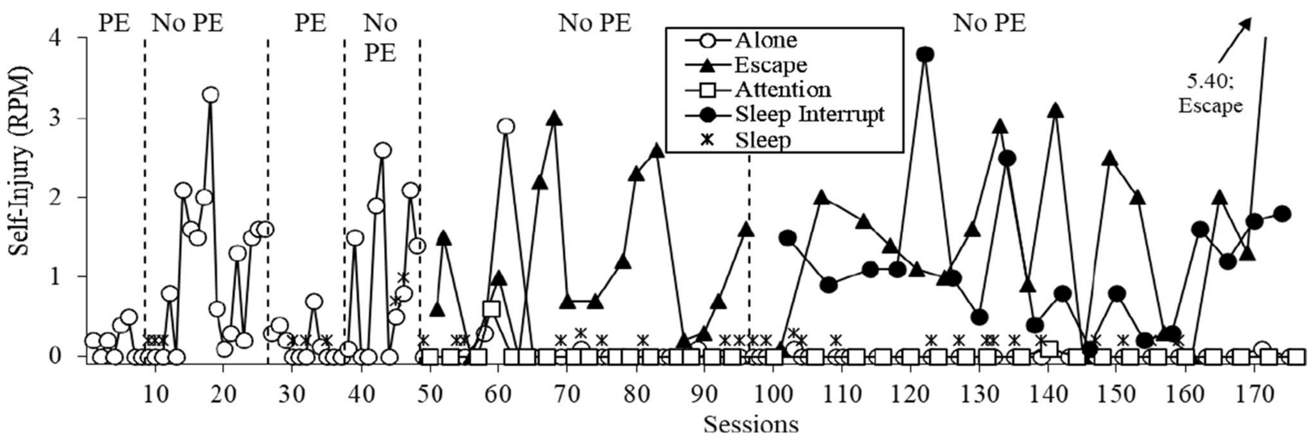

Fig. 1 Functional analysis of self-injury

SIB. Demands (stacking blocks and motor imitation) were based on a caregiver interview. We started the multielement FA without PE with the goal of using this data as a baseline to later compare to conditions with PE to evaluate multiply-maintained SIB, as has been done in past research (Scheithauer et al., 2017). However, despite SIB occurring during extended alone sessions without PE, SIB did not persist in alone sessions without PE when interspersed with other conditions in the multielement FA. Thus, we used the multielement FA, with the alone as a control, to assess for social reinforcers without the inclusion of PE. Bryan engaged in elevated rates of SIB in the escape condition compared to alone, suggesting SIB was in part maintained by escape from demands (Fig. 1, Phase 5).

In the multielement assessment, Bryan slept (defined as lying down with his eyes closed and minimal movement for 2 consecutive minutes) during more sessions (46.67% of alone sessions in multielement) compared to the previously conducted extended alone assessment (16.67% of sessions). Bryan did not meet the definitions for sleep during escape session in the multielement assessment, likely because the least-to-most prompting interrupted sleep during the physical prompt. In all other conditions, sleep was uninterrupted. Thus, the elevations of SIB in the escape condition, compared to the other conditions, could have been attributed to sleep interruption (and subsequent access to sleep during the reinforcement interval) as opposed to escape from demands. To address this, we added a condition to test for SIB maintained by access to sleep. The therapist used verbal and physical prompts to interrupt sleep on a VT 2-5 s schedule with no other demands. Contingent on SIB, sleep-interruption paused for 30 s. We observed elevated rates of SIB in the escape (M = 1.63 RPM) and access to sleep (M = 1.18 RPM) conditions compared to the alone condition (Fig. 1, Phase 6).

Treatment Evaluation and Competing Attention Assessment

During baseline, the therapist presented the same demands as the escape condition in the FA and continued the sleep-interruption schedule (i.e., the therapist physically prompted Bryan to sit up during demands). Contingent on SIB, the therapist stopped demands and sleep-interruption for 30 s. Therapists included a picture card in the initial baseline and a card and button (see FCT section) in the second baseline but ignored FCRs. Bryan engaged in elevated rates of SIB during baseline (M = 3.35 RPM) and no FCRs (Fig. 2, middle and bottom panels).

Following baseline, the therapist implemented FCT with 10 trials per session. In each trial, the lead therapist delivered the same demands as in baseline and a second therapist, positioned behind Bryan, physically prompted him to engage in the FCR. This started at a 0-s delay, with the physical prompt immediately after initiation of the first demand. The therapist ignored SIB and reinforced FCRs by removing all task materials and pausing sleep-interruption for 30 s. We increased the delay between presentation of demands and prompting the FCR using a progressive time delay until Bryan engaged in the FCR independently on 90% of trials for two consecutive sessions. The FCR was initially a card exchange, but Bryan did not master the response after several sessions (Fig. 2, top left panel). Thus, we switched to a button press that exhibited the phrase “break please.” Bryan mastered the button-press FCR at a 32-s time delay (Fig. 2, top right panel).

Next, we evaluated a differential reinforcement of alternative (DRA) behavior treatment (Lalli et al., 1995). Sessions were 10 min. Therapists provided a 30-s break contingent on FCRs (no prompting was included) and ignored SIB. SIB continued at variable rates (M = 2.29 RPM) despite an increase in FCRs. SIB occurred both during the presentation of demands and during reinforcement delivery (e.g.,

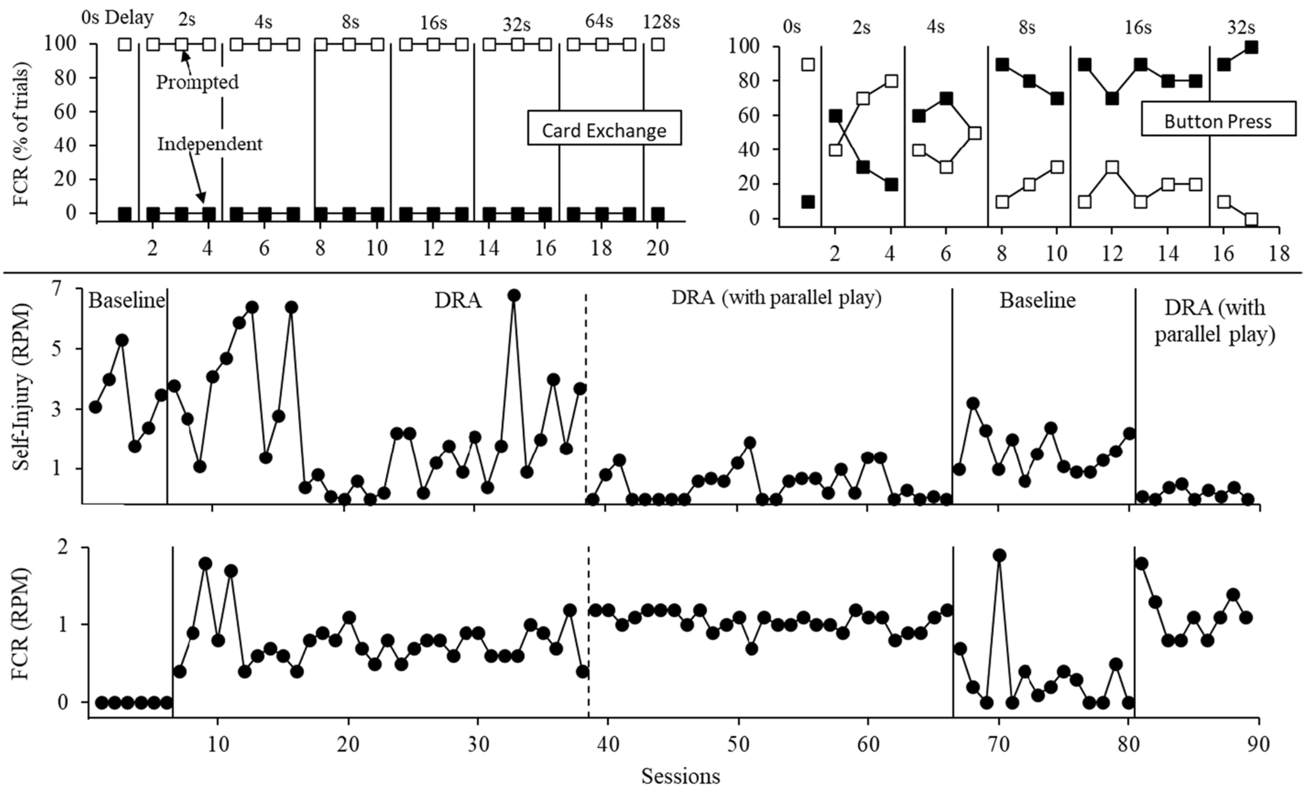

Fig. 2 Treatment evaluation, including functional communication training (top two panels), self-injury (second panel) and FCRs (bottom panel) during the DRA treatment evaluation

1.61 RPM of SIB during reinforcement in the last five sessions prior to the competing attention assessment). Thus, we hypothesized that SIB maintained by automatic reinforcement (as identified in the initial extended alone evaluation) may have reemerged and was affecting the effectiveness of treatment. Observations outside of treatment suggested attention might compete with the reinforcers maintaining SIB during this time. Thus, we paused the DRA treatment to evaluate whether attention might compete with the hypothesized automatic reinforcement produced by the SIB.

We conducted a modified version of a competing stimulus assessment (Haddock & Hagopian, 2020) with various forms of attention (i.e., a competing attention assessment). Attention types included physical touch, singing, talking, talking with physical touch, and parallel play (therapist interacted with items while facing Bryan, but did not talk or provide physical touch). Alone and ignore conditions served as controls. Sessions were 10 min. The therapist implemented each type of attention and both control conditions in a random order within series for six series. At the session start, the therapist provided the designated attention continuously. Bryan did not wear PE and the therapist ignored SIB. Because it is difficult to operationally define interaction with attention (a variable usually measured in competing items assessments with toys; Haddock & Hagopian, 2020), we based results solely on rates of SIB. Bryan engaged in the lowest rates of SIB during parallel play ($M = .08$ RPM; Fig. 3).

We included parallel play in the reinforcement interval of the DRA treatment starting at Session 39. Following this, Bryan engaged in low rates of SIB ($M = .49$ RPM), and we replicated this with a reversal (Fig. 2).

Discussion

To our knowledge, this is the first documented example of treatment with a child with BRPS, demonstrating generality of ABA strategies for SIB to this rare genetic

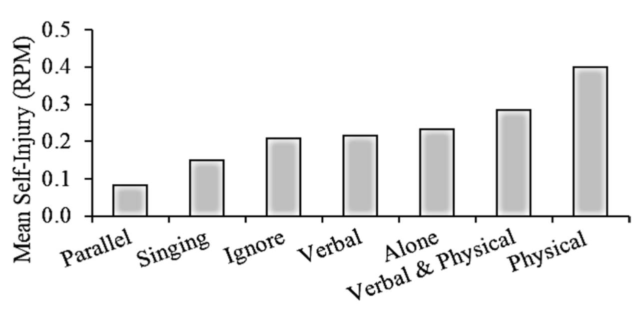

Fig. 3 Competing attention assessment

syndrome. Given the importance of replicating the efficacy of ABA with diagnoses other than ASD to promote accessibility of services, this is an important finding.

Bryan's assessment included the systematic replication of common strategies (e.g., screener for automatic reinforcement, use of protective equipment for sensory extinction, functional communication training, differential reinforcement) with several extensions. We replicated a screening for automatically maintained behavior (Querim et al., 2013) with an evaluation of PE to produce sensory extinction. Bryan engaged in elevated rates of SIB during extended alone sessions without PE and PE suppressed SIB, lending evidence to an automatic function. Despite this, he engaged in low rates of SIB during the alone condition of the subsequent multielement FA with no PE. This pattern of responding is counter to previous findings where elevated rates of SIB in a screener usually predicts elevated rates in subsequent alone sessions in a multielement FA (Querim et al., 2013). There are several possible explanations for our findings. First, the results of the extended alone assessments could be erroneous. However, this is unlikely given rates of SIB remained elevated across several extended alone sessions and we demonstrated a reduction in SIB when using PE in a reversal (Scheithauer et al., 2017). An alternative explanation is that social test conditions may have altered the establishing operation for automatic reinforcement produced by SIB. For example, after experiencing high rates of SIB evoked by the presence of demands, Bryan may have satiated on the automatic reinforcement produced by SIB, resulting in reduced rates during subsequent alone sessions. Another possible explanation is shifts in the motivating operations for sleep that affected automatically maintained SIB. If the establishing operation for sleep was stronger on the days the multielement FA was implemented (compared to the previous days where the extended alone sessions were implemented), sleep might have competed with the automatic reinforcement produced by SIB during the multielement FA (but not during the extended alone assessment). It is also possible that other physiological symptoms influenced SIB, sleep, or both (e.g., Bryan had issues with constipation and feeding). In sum, a limitation of this study is that we cannot determine the exact mechanism responsible for the discrepancy between SIB in the extended alone evaluation versus the alone sessions in the multielement FA. Additional research should test the impact of several physiological factors on automatically maintained SIB, especially for children with disorders associated with medical comorbidities such as BRPS.

Although the mechanisms influencing Bryan's assessment results need future research to clarify, the procedural modifications to our assessment might prove useful for other clients that present similarly to Bryan. That is, if a child is engaging in elevated rates of SIB during alone conditions at some times but not others, it might be possible to assess for social functions during times that the establishing operation is not present for automatically maintained behavior. If multiply maintained challenging behavior is identified, therapists can then tailor treatment to address multiple functions or conduct additional assessments to identify the establishing operations responsible for fluctuations in the automatically maintained behavior.

A second novel extension was the test for SIB maintained by access to sleep. Past research suggests drowsiness may act as an establishing operation for other functions for challenging behavior (Kennedy & Meyer, 1996). However, to our knowledge, this is the first demonstration of a test for SIB maintained by access to sleep directly. Given the condition contingencies, it is not possible to determine whether this condition technically identified SIB maintained by access to sleep/laying down, escape from sitting up, or escape from the social interaction associated with the sleep interruption. Regardless, these data suggest that SIB was maintained by some variable associated with escape from the therapist physically interrupting sleep, in the absence of other demands, providing additive information to the escape condition alone. This novel FA condition might be important for clinicians working with children who experience sleep issues, a symptom common among children with BRPS and other genetic disorders (UNIQUE, 2018).

A limitation of our evaluation is the lack of direct measure of Bryan's sleep quality at home. When considering the impact of sleep, future researchers and clinicians should use validated measures for gathering information about sleep. For example, actigraphy measures collecting automated data on sleep; nightly diaries and logs completed by parents on basic sleep information; and validated sleep questionnaires are methods recommended for assessment of sleep in children with disabilities (for additional details on sleep measurement, see Abel et al., 2017). Collecting these measures would allow for a more comprehensive assessment of sleep and the impact that sleep may have on behavior. In addition, it is unclear from our data why Bryan slept more during some evaluations compared to others. It is possible that sleep at home affected sleep in the clinic, in which case more validated measures of sleep data as described above would be helpful. However, it is also possible that other physiological factors influenced variability in sleep.

As another extension, we implemented a DRA treatment described in past research (Lalli et al., 1995). Unlike past studies, this treatment was unsuccessful on its own. This was possibly attributed to automatically maintained SIB that persisted during reinforcement. A common treatment for automatically maintained behavior is the use of competing stimulus (Haddock & Hagopian, 2020). However, our preference assessments suggested Bryan did not regularly interact with items. Given anecdotal evidence that attention might be preferred, we implemented a competing stimulus assessment (Haddock

& Hagopian, 2020) with the novel modification of including different forms of attention. We felt this novel extension was necessary because Bryan lacked prerequisite skills necessary for previously established methods of preference-assessments for attention (e.g., Morris & Vollmer, 2020). For example, Bryan lacked the prerequisite skills for choice-based assessments (e.g., array scanning), and given his limited mobility, we had concern requiring movement to demonstrate choice (e.g., Strohmeier et al., 2018). Thus, presenting forms of attention in a free-operant context and measuring SIB was the best option for Bryan's presentation. Our competing attention assessment successfully identified a form of attention (parallel play) that resulted in reductions in SIB when used in treatment. It is interesting that parallel play is not often included in preference assessments for attention. These procedures (competing attention assessment) and inclusion of parallel play may be important considerations for clients with a similar presentation as Bryan.

Unfortunately, we did not fully evaluate reinforcement schedule thinning, home generalization, or caregiver training prior to Bryan discontinuing services (Bryan withdrew early due to COVID-19). Despite the lack of generalization and maintenance data, the results are important as they are the first indication of behavior reduction using ABA strategies in a child with BRPS and highlight numerous extensions to empirically supported strategies that may be further evaluated in future research and clinical work.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s40617-022-00749-x.

Declarations

Conflicts of Interest The authors have no conflicts of interest to report.

Ethical Approval We received an exemption from the institutional review board of the first author's institution as it was deemed a case study. All ethical standards for human subjects research were followed.

References

Abel, E., Kim, S. Y., Kellerman, A. M., & Brodhead, M. T. (2017). Recommendations for identifying sleep problems and treatment resources for children with autism spectrum disorder. *Behavior Analysis in Practice, 10*(3), 261–269. https://doi.org/10.1007/s40617-016-0158-4

Balasubramanian, M., Willoughby, J., Fry, A. E., Weber, A., Firth, H. V., Deshpande, C., Berg, J. N., Chandler, K., Metcalfe, K. A., Lam, W., Pilz, D. T., Tomkins, S., & DDS Study. (2017). Delineating the phenotypic spectrum of Bainbridge-Ropers syndrome. *Journal of Medical Genetics, 54*(8), 537–543. https://doi.org/10.1136/jmedgenet-2016-104360

Georgia Department of Community Health, Division of Medicaid (2020). Policies and procedures for Autism Spectrum Disorder (ASD) services. Retrieved from: https://medicaid.georgia.gov/document/publication/asd-policy-manual/download

Haddock, J. N., & Hagopian, L. P. (2020). Competing stimulus assessments: A systematic review. *Journal of Applied Behavior Analysis, 53*(4), 1982–2001. https://doi.org/10.1002/jaba.754

Iwata, B. A., Dorsey, M. F., Slifer, K. J., Bauman, K. E., & Richman, G. S. (1994). Toward a functional analysis of self-injury. *Journal of Applied Behavior Analysis, 27*, 197–209. https://doi.org/10.1901/jaba.1994.27-197

Kennedy, C. H., & Meyer, K. A. (1996). Sleep deprivation, allergy symptoms, and negatively reinforced problem behavior. *Journal of Applied Behavior Analysis, 29*(1), 133–135. https://doi.org/10.1901/jaba.1996.29-133

Lalli, J. S., Casey, S., & Kates, K. (1995). Reducing escape behavior and increasing task completion with functional communication training, extinction, and response chaining. *Journal of Applied Behavior Analysis, 28*, 261–268. https://doi.org/10.1901/jaba.1995.28-261

Morris, S. L., & Vollmer, T. R. (2020). A comparison of methods for assessing preference for social interactions. *Journal of Applied Behavior Analysis, 53*(1), 918–937. https://doi.org/10.1002/jaba.692

Querim, A. C., Iwata, B. A., Roscoe, E. M., Schlichenmeyer, K. J., Ortega, J. V., & Hurl, K. E. (2013). Functional analysis screening for problem behavior maintained by automatic reinforcement. *Journal of Applied Behavior Analysis, 46*(1), 47–60. https://doi.org/10.1002/jaba.26

Rooker, G. W., Bonner, A. C., Dillon, C. M., & Zarcone, J. R. (2018). Behavioral treatment of automatically reinforced SIB: 1982–2015. *Journal of Applied Behavior Analysis, 51*(4), 974–997. https://doi.org/10.1002/jaba.492

Scheithauer, M. C., Lomas Mevers, J. E., Call, N. A., & Shrewsbury, A. N. (2017). Using a test for multiply-maintained self-injury to develop function-based treatments. *Journal of Developmental & Physical Disabilities, 29*, 443–460. https://doi.org/10.1007/s10882-017-9535-3

Strohmeier, C. W., Crysdale, C., & Schwandtner, S. (2018). Rapid assessment of attention types for the treatment of attention-maintained problem behavior. *Behavior Analysis in Practice, 11*(4), 417–423. https://doi.org/10.1007/s40617-018-00300-x

Understanding Chromosomes & Gene Disorders (UNIQUE). (2018). Bainbridge-ropers syndrome (ASXL3 "loss of function"). https://www.rarechromo.org/media/singlegeneinfo/Single%20Gene%20Disorder%20Guides/Bainbridge-Roper%20syndrome%20ASXL3%20FTNW.pdf. Accessed in 2018