Special Interest Section Quarterly
Technology
Volume 19, Number 1 • March 2009
Published by The American Occupational Therapy Association, Inc.

Assistive Technology Use by Persons With Fragile X Syndrome: Three Case Reports

■ Laura Greiss Hess, MS, OTR/L, Kerrie Lemons Chitwood, MA, CCC-SLP, Susan W. Harris, Mariya Borodyanskaya, Randi J. Hagerman, MD, Cathy Bodine, PhD, CCC-SLP, and Michael Guralnick, PhD

When it comes to designing assistive technology (AT) interventions and services for children with intellectual disabilities and autism spectrum disorder (ASD), occupational therapists often collaborate with other disciplines, such as speech-language pathology. Children with Fragile X syndrome (FXS) have significant intellectual disabilities and often present with ASD, yet the current literature does not reflect the range of effective interventions specifically for FXS. Therefore, occupational therapy practitioners must rely on and extrapolate the literature on intellectual disability, ASD, and AT when designing and providing services for children with FXS. This article reviews some of the current literature regarding AT use by children with intellectual disabilities and ASD and presents three case reports that illustrate a broad range of AT use by children with FXS.

Fragile X Syndrome

Children with FXS have a range of cognitive deficits, from learning disabilities to mental retardation and autism. FXS affects approximately 1 in 3,600 children and is caused by a trinucleotide expansion in the fragile X mental retardation 1 gene, which is located on the bottom end of the X chromosome (Crawford et al., 2002). FXS is the most common cause of inherited mental retardation and is the leading known single-gene cause of autism (Belmonte & Bourgeron, 2006; Reddy, 2005; Schaefer & Lutz, 2006). Typical features of FXS are hyperactivity; inattention; impulsive behavior; and autistic features, such as poor eye contact, tactile defensiveness, hand flapping, and hand biting (Hagerman, 2002; Loesch et al., 2002). Approximately 30% of children with FXS meet diagnostic criteria for autism (Kaufmann et al., 2004; Rogers, Wehner, & Hagerman, 2001), and an additional 30% meet criteria for pervasive development disorder not otherwise specified (Harris et al., 2006). Approximately 10% of children with FXS do not speak by 5 years of age.

A study of academic skills in children with FXS demonstrated relative strengths in general knowledge, reflecting an ability to integrate experiential information (Roberts et al., 2005). Relative weaknesses included prewriting skills and visuospatial processing abilities, resulting in difficulty and frustration with many pencil-and-paper tasks. Although little research has been done regarding academic interventions for children with FXS, a wide range of interventions exist for treating or managing the deficits exhibited in children with intellectual disabilities and ASD. AT devices are commonly used in these populations to address problems with language and communication, memory, motor learning, and other early developmental abnormalities. Successful AT use requires only that children have "an identifiable, reproducible motor pattern and sufficient sensory skills to recognize the stimulus being provided" (Daniels, Sparling, Reilly, & Humphry, 1995, p. 92). Computer use can enhance written expression and learning in children with FXS because of these kids' specific strengths in visual processing and weaknesses in visuomotor coordination (Hagerman & Hagerman, 2002; Kogan et al., 2004).

AT for Children With Intellectual Disabilities and ASD

Early Intervention

Children with disabilities often lack the ability and opportunity to learn appropriate play skills. Low-technology devices such as picture boards for communication and Dycem (anti-skid material) for stabilizing toys, as well as more sophisticated devices such as adjustable equipment to improve positioning and access, electronic equipment and switch-adapted toys, and alternative-input devices that integrate with software (e.g., IntelliKeys keyboard, IntelliTools, Petaluma, CA) can allow children with disabilities to explore and learn new concepts (Lane & Mistrett, 1996; Wershing & Symington, 1998). Use of these kinds of devices for infants and toddlers with disabilities is best carried out within a family-centered framework.

Literacy

Communication impairment is the biggest barrier for children with disabilities in developing relationships and participating in home, school, and community activities. In fact, social and communication dysfunction are considered the earliest indicators of ASD (Woods & Wetherby, 2003). Many children with intellectual disabilities, ASD, and FXS have speech and language impairments that require the use of augmentative and alternative communication (AAC) aids, such as communication boards and speech-generating devices (Schlosser & Sigafoos, 2006). AAC aids may augment a child's existing speech or serve as the primary method of expressive communication (Mirenda, 2003; Wilkinson & Hennig, 2007). Although AAC has been underused with infants and young children, both aided and unaided forms of AAC are appropriate and effective for this population (Romski & Sevcik, 2005). Many AT applications address problems with literacy, mathematics, and overall academic performance (U.S. Department of Education, 2007). Children who struggle with either

the process of writing or the physical act of handwriting may benefit from computer software programs that predict and read words aloud as they are typed. Examples include the Click 4 (Crick Software, Ltd., Northampton, UK), the AlphaSmart 3000 (AlphaSmart Direct, Inc., Wisconsin Rapids, WI), and Co:Writer 4000 and Write:OutLoud (Don Johnston, Inc., Volo, IL).

Behavioral Adaptations

AT interventions can be used to promote positive behaviors and reduce negative behaviors for children with intellectual disabilities, ASD, and FXS. For example, functional communication training has been used to identify the source of the undesirable behavior and then present the child with more appropriate behavioral response options, such as using AAC or manual signs for appropriately expressing emotions (Mirenda, 2001).

Case Reports

Case 1

Casey is a 13-year-old boy with FXS, intellectual disability, and autism. His early development was globally delayed, and he is nonverbal with a history of severe expressive language delays. His behaviors include hand flapping, poor eye contact, tactile defensiveness, hyperactivity, and significant social deficits. Casey participated in occupational therapy and speech therapy privately and through the school district for several years, but he is not currently receiving services because of difficulty in finding qualified professionals within his rural community. In the past, Casey's occupational therapists and speech-language pathologists tried various forms of AAC, including sign language and picture exchange, but parent reports about the effectiveness of these modes of communication were unclear.

At 8 1/2 years of age, Casey began using a Dynamo communication device (DynaVox Technologies, Pittsburgh, PA) obtained through his private speech-language pathologist. The Dynamo has a dynamic display and digitized speech output. His parents reported that the private and school speech therapists did not collaborate or communicate well with regard to Casey's use of the device, and occupational therapy was not involved in its implementation. Because the adults who interact with Casey were not trained in the Dynamo's use, the device was not used across settings (e.g., home, school, community). Although Casey's mother received some training, she did not become proficient enough to program or troubleshoot the Dynamo, so she is trying to train herself to program and use the device. Overall, she reported that the device was appropriate, but she expressed frustration and desperation for Casey to have an effective means of communication, and she will continue to work to help improve his communication abilities.

Case 2

Shayna is a 13-year-old girl with FXS and intellectual disability. She attends a special day class on a general education campus. She demonstrates marked social anxiety and learning challenges, specifically deficits in written expression and handwriting. Her handwriting was a particular source of frustration and a barrier to her academic participation. When presented with writing tasks at school and home, she became agitated, refused to participate, and completely shut down.

Low-technology interventions. Shayna has a history of using AT throughout school, but she is very sensitive about appearing different from her peers. Although she and her occupational therapist have tried several different pencil grips to improve her motor control while writing, her parents reported that the grips made no functional difference in penmanship, and Shayna refused to use them because they felt physically and socially uncomfortable. Shayna has used other low-technology devices, such as egg and visual timers, to help her focus and participate in challenging academic activities like reading and writing, as well as in nonacademic activities like playing on the computer. Although the timers helped her to transition from one task to another, she now feels too old to use them. She continues to successfully use visual structures like checklists and charts in the home to keep her organized.

High-technology interventions. An occupational therapist and speech-language pathologist recently introduced CO:Writer and Write:OutLoud to Shayna. CO:Writer is a word prediction software program that aids with spelling and sentence composition. The software reduces the number of key strokes required to type any word by producing a list of predicted words that are refined as the user adds letters. Write:OutLoud is a word processor with text-to-speech capability, providing the user with auditory feedback as an additional method for proofreading work. As a participant in the research study that provided the software, Shayna is required to use the applications at home and at school at least 3 days a week for a minimum of 30 minutes. Although she found that CO:Writer did not meet her needs, she is using Write:OutLoud at home every day for homework and for nonacademic activities like writing stories or e-mails. Her mother reported that Write:OutLoud has "extended the depth and amount of writing taking place" in the home and that Shayna effectively accesses and uses the software at school.

Case 3

William is a 14-year-old boy with the Prader-Willi subphenotype of FXS (Nowicki et al., 2007), intellectual disability, and pervasive development disorder not otherwise specified. His early development was globally delayed; he was walking at 19 months and speaking at 2.5 years. He displayed hand flapping, excessive chewing of objects, poor eye contact, tactile defensiveness, perseveration of thoughts, daily tantrums, attention problems, shyness, anxiety, and depression. Intervention teams have included occupational therapy, speech therapy, and special education. Throughout his special education history, William has used different levels and types of AT with varying success. In preschool, he used a touch screen and cause-and-effect software with support from occupational therapy and speech therapy. His family also had access to an AT lending library through the Council on Mental Retardation.

Visual structure. Since preschool, William has used low-technology strategies in his daily program, including a visual schedule. He began with photos and later progressed to Boardmaker icons (Mayer-Johnson, Inc., Solana Beach, CA) paired with typed words.

Technology
Special Interest Section Quarterly
(ISSN 1093–7137)

Chairperson: Kimberly Hartmann
Editor: Katherine Post
Production Editor: Jennifer Hart

Published quarterly by The American Occupational Therapy Association, Inc., 4720 Montgomery Lane, Bethesda, MD 20814-3425; ajotsis@aota.org (e-mail). Periodicals postage paid at Bethesda, MD. POSTMASTER: Send address changes to *Technology Special Interest Section Quarterly,* AOTA, PO Box 31220, Bethesda, MD 20824-1220. Annual membership dues are $225 for OTs, $131 for OTAs, $75 for Student-Plus members, and $53 for Standard Student members. All *SIS Quarterlies* are available to members at www.aota.org. The opinions and positions stated by the contributors are those of the authors and not necessarily those of the editor or AOTA. Sponsorship is accepted on the basis of conformity with AOTA standards. Acceptance of sponsorship does not imply endorsement, official attitude, or position of the editor or AOTA.

The picture icons were faded, leaving a written schedule without pictorial support, which he relies on for daily organization; William's family expects that Boardmaker likely will be his lifelong AT support. The implementation of visual supports has continued to be a highly interdisciplinary process, with the special education teacher, speech-language pathologist, and occupational therapist working in concert on William's behalf.

Written expression and literacy-related AT. In elementary school, William's occupational therapist provided enlarged, color-coded labels to visually organize a keyboard for typing. This strategy was helpful both at home and at school to enhance literacy skills, initially targeting the typing of single words, then entire sentences. In fourth grade, William was introduced to CO:Writer and Write:OutLoud, but these aids were not used maximally at school until the next year, when he received individualized training with the software. An AlphaSmart portable word processor was introduced in the fifth grade, but William did not want to use it because he believed that it made him different from the other students. William is now in seventh grade and completes written assignments with a regular computer with word processing software and a spell checker. Overall, reducing handwritten work has greatly improved William's participation in academic programming.

AT environmental adaptations. The lock on William's locker has line-up-style wheels instead of the typical spin dial, which reduces fine motor and motor planning demands and facilitates independence. Similarly, clothing modifications, such as hook-and-loop fasteners and elastic waistbands, have promoted his independence in self-dressing. In the classroom, William uses a chair with arms to reduce the effort needed to maintain posture and arousal because of his low muscle tone. Finally, he rides a three-wheel bicycle for leisure and fitness to accommodate for the gross motor deficits that preclude him from riding a two-wheeler.

Discussion

Occupational therapists play an important role as members of interdisciplinary teams that design and implement AT applications for children with intellectual disabilities, including FXS. The frequency, duration, and format of occupational therapy services within an interdisciplinary team must be highly individualized and fluid to meet the needs of the child. These services may increase during times of transition or after obtaining an AT device or system in order to incorporate functional strategies into daily occupations. Teams often exert much effort in evaluating and obtaining AT devices or systems but do not dedicate enough time to implementing technology, as highlighted in Casey's story. Shayna benefited from a blend of occupational therapy and speech therapy as well as from her enrollment in a research project for AT applications. William received occupational therapy services throughout his school career, including early AT for computer access and ongoing occupational therapy support through high school for activities of daily living and leisure and fitness pursuits. These three cases illustrate how AT can benefit students with FXS in the areas of play and leisure, communication, academic and school participation, written expression, computer access, environmental design and access, independent task completion, self-care, and positioning. Individualized education plan teams often do not consider the breadth and scope of AT applications, which frequently are limited to computer use or communication needs. These teams should discuss student needs and consider both low- and high-technology AT applications across all domains. Success in AT use requires that teams recognize and respond to the variable demands of individual consumers and families, with administrative support and continuing education for teachers, therapists, and support staff. ■

Acknowledgments

This work was supported by the National Institute on Disability and Rehabilitation Research under the U.S. Department of Education, grant #H133E040019; the Coleman Institute for Cognitive Disabilities, Administration of Developmental Disabilities grant for Center of Excellence in Developmental Disabilities #90DD05969; and the National Institute of Child Health and Human Development, grant #HD02274. We thank the families and children who have participated in this research. We also thank Laura Meyer, formerly of Assistive Technology Partners, and the research team at the University of California Davis Medical Investigation of Neurodevelopmental Disorders (M.I.N.D.) Institute.

References

Belmonte, M. K., & Bourgeron, T. (2006). Fragile X syndrome and autism at the intersection of genetic and neural networks. *Nature Neuroscience, 9*, 1221–1225.

Crawford, D. C., Meadows, K. L., Newman, J. L., Taft, L. F., Scott, E., Leslie, M., et al. (2002). Prevalence of the fragile X syndrome in African-Americans. *American Journal of Medical Genetics, 110*, 226–233.

Daniels, L. E., Sparling, J. W., Reilly, M., & Humphry, R. (1995). Use of assistive technology with young children with severe and profound disabilities. *Infant-Toddler Intervention, 5*(1), 91–112.

Hagerman, R. J. (2002). Physical and behavioral phenotype. In R. J. Hagerman & P. J. Hagerman (Eds.), *Fragile X syndrome: Diagnosis, treatment, and research* (3rd ed., pp. 3–109). Baltimore: Johns Hopkins University Press.

Hagerman, R. J., & Hagerman, P. J. (2002). *Fragile X syndrome: Diagnosis, treatment, and research* (3rd ed.). Baltimore: Johns Hopkins University Press.

Harris, S. W., Goodlin-Jones, B., Nowicki, S. T., Hessl, D., Tassone, F., Barbato, I., et al. (2006). *Autism profiles of young males with fragile X syndrome*. Paper presented at the 10th International Fragile X Conference, Atlanta, GA.

Kaufmann, W. E., Cortell, R., Kau, A. S., Bukelis, I., Tierney, E., Gray, R. M., et al. (2004). Autism spectrum disorder in fragile X syndrome: Communication, social interaction, and specific behaviors. *American Journal of Medical Genetics, 129A*, 225–234.

Kogan, C. S., Boutet, I., Cornish, K., Zangenehpour, S., Mullen, K. T., Holden, J. J., et al. (2004). Differential impact of the FMR1 gene on visual processing in fragile X syndrome. *Brain, 127*(Pt 3), 591–601.

Lane, S. J., & Mistrett, S. G. (1996). Play and assistive technology issues for infants and young children with disabilities: A preliminary examination. *Focus on Autism and Other Developmental Disabilities, 11*(2), 96–104.

Loesch, D. Z., Huggins, R. M., Bui, Q. M., Epstein, J. L., Taylor, A. K., & Hagerman, R. J. (2002). Effect of the deficits of fragile X mental retardation protein on cognitive status of fragile X males and females assessed by robust pedigree analysis. *Journal of Developmental and Behavioral Pediatrics, 23*, 416–423.

Mirenda, P. (2001). Autism, augmentative communication, and assistive technology: What do we really know? *Focus on Autism and Other Developmental Disabilities, 16*(3), 141–151.

Mirenda, P. (2003). Toward a functional augmentative and alternative communication for students with autism: Manual signs, graphic symbols, and voice output communication aids. *Language, Speech, and Hearing Services in School, 34*, 203–216.

Nowicki, S. T., Tassone, F., Ono, M. Y., Ferranti, J., Croquette, M. F., Goodlin-Jones, B., et al. (2007). The Prader-Willi phenotype of fragile X syndrome. *Journal of Developmental and Behavioral Pediatrics, 28*, 133–138.

Reddy, K. S. (2005). Cytogenetic abnormalities and fragile-X syndrome in autism spectrum disorder. *BMC Medical Genetics, 6*(1), 3.

Roberts, J. E., Schaaf, J. M., Skinner, M., Wheeler, A., Hooper, S., Hatton, D. D., et al. (2005). Academic skills of boys with fragile X syndrome: Profiles and predictors. *American Journal on Mental Retardation, 110*(2), 107–120.

Rogers, S. J., Wehner, E. A., & Hagerman, R. J. (2001). The behavioral phenotype in fragile X: Symptoms of autism in very young children with fragile X syndrome, idiopathic autism, and other developmental disorders. *Journal of Developmental and Behavioral Pediatrics, 22*, 409–417.

Romski, M., & Sevcik, R. A. (2005). Augmentative communication and early intervention. *Infants and Young Children, 18*, 174–185.

Schaefer, G. B., & Lutz, R. E. (2006). Diagnostic yield in the clinical genetic evaluation of autism spectrum disorders. *Genetics in Medicine, 8*, 549–556.

Schlosser, R. W., & Sigafoos, J. (2006). Augmentative and alternative communication interventions for persons with developmental disabilities: Narrative review of comparative single-subject studies. *Research in Developmental Disabilities, 27*, 1–29.

U.S. Department of Education. (2007). *Effectiveness of reading and mathematics software products: Findings from the First Student Cohort*. Retrieved October 14, 2008, from http://www.ed.gov/ids.

Wershing, A., & Symington, L. (1998). Learning and growing with assistive technology. In S. L. Judge & H. P. Parette (Eds.), *Assistive technology for young children with disabilities* (pp. 45–75). Cambridge, MA: Brookline.

Wilkinson, K. M., & Hennig, S. (2007). The state of research and practice in augmentative and alternative communication for children with developmental/intellectual disabilities. *Mental Retardation and Developmental Disabilities Research Reviews*, 13(1), 58–69.

Woods, J. J., & Wetherby, A. M. (2003). Early identification of and intervention for infants and toddlers who are at risk for autism spectrum disorder. *Language, Speech, and Hearing Services in Schools, 34*, 180–193.

***Laura Greiss Hess**, MS, OTR/L, is Occupational Therapist, M.I.N.D. Institute, University of California Davis Medical Center, 2825 50th Street, Room 1356, Sacramento, California 95817; Laura.greiss-hess@ucdmc.ucdavis.edu.*

***Kerrie Lemons Chitwood,** MA, CCC-SLP, is Speech Pathologist, M.I.N.D. Institute, University of California Davis Medical Center, Sacramento, California.*

***Susan W. Harris,** is Staff Research Associate, M.I.N.D. Institute, University of California Davis Medical Center, Sacramento, California.*

***Mariya Borodyanskaya,** is Post-Graduate Researcher, M.I.N.D. Institute, University of California Davis Medical Center, Sacramento, California.*

***Randi J. Hagerman, MD,** is Medical Director, M.I.N.D. Institute, and Professor of Pediatrics, University of California Davis Medical Center, Sacramento, California.*

***Cathy Bodine, PhD, CCC-SLP,** is Executive Director, Assistive Technology Partners, and Associate Professor, Department of Physical Medicine and Rehabilitation, University of Colorado Health Sciences Center, Denver, Colorado.*

***Michael Guralnick,** PhD, is Director, Professor of Psychology and Pediatrics, Center on Human Development and Disability, University of Washington, Seattle, Washington.*

Hess, L. G., Chitwood, K. L., Harris, S. W., Borodyanskaya, M., Hagerman, R. J., Bodine, C., & Guralnick, M. (2009, March). Assistive technology use by persons with fragile X syndrome: Three case reports. *Technology Special Interest Section Quarterly, 19*, **TK**.

FPO

The American Occupational
Therapy Association, Inc.
PO Box 31220
Bethesda, MD 20824-1220

PERIODICALS
POSTAGE
PAID AT
BETHESDA
MD