Preserving play in pediatric neurodegenerative disorders: a case study

Loren F. McMahon[1,2], Abbie King[1]

[1]*Augmentative Communication Program, Boston Children's Hospital, Waltham, MA*

[2]*Tufts University, Medford, MA*

INTRODUCTION

For a child, play is a crucial occupation that supports psychosocial, cognitive, and physical development and provides opportunities to discover new areas of interest, practice advocacy, and learn group skills [1]. These social skills, including working in groups, sharing, negotiating, and conflict resolution, build a foundation for social behavior and are developed through individual and group play [2]. Play is well recognized in the field of occupational therapy as a primary domain of childhood, acting as a driver for early learning opportunities. In speech-language pathology, it is well known that receptive and expressive language development are positively correlated with functional play development [3]. Although play can involve a final product, participation is the primary goal, whether independent or with peers [4]. As the United Nations High Commission for Human Rights considers play to be a human right, modifications to the task or environment must be made to ensure that children who have physical motor challenges can access and engage in play [4]. Assistive technology (AT) and augmentative and alternative communication (AAC) are shown to improve the quality of life in individuals with complex motor profiles by enabling participation in play, communication, and access to daily occupations [5]. However, children with progressive neurodegenerative disorders (PND) require additional and consistent modifications to engage in play as their physical, cognitive, and communicative profiles change not only due to their age and typical developmental trajectory but also due to regressions across domains of their diagnosis [6]. Research has demonstrated that for individuals with PND and chronic conditions, play positively affects the children themselves, increasing resiliency and understanding of their illness. Similar trends are also documented for their entire family, positively impacting the quality of the home environment [6]. Health professionals who recommend AT and AAC for children with PNDs must consider and support play as a primary domain throughout the therapeutic process.

CLINICAL PROFILE

D was born full-term and healthy in April of 2015. At approximately 18 months old, D's parents began noticing subtle fine motor delays. These subtle fine motor delays slowly progressed into weak fine motor coordination, mild tremors, and balance difficulties. In November 2020, D saw a Neurologist, where he was subsequently diagnosed with ADHD inattentive type and sensory processing disorder. D and his family contracted COVID-19 in December 2020. Despite having mild COVID-19 symptoms, D's parents noticed a rapid decline in his overall motor skills and speech. D underwent an MRI and whole exome sequence, revealing Juvenile Metachromatic Leukodystrophy (JMLD).

JMLD is an autosomal recessive gene mutation that accumulates toxic substrates in organic tissue and causes progressive dysfunction within the central and peripheral nervous systems [7]. JMLD's prevalence within the Navajo Nation, D's ethnicity, is 1 in 2,500 individuals [8]. Commonly observed changes in motor performance include degeneration in gait, decreased fine motor skills, rigidity, and ataxia [8]. Cognitive challenges include difficulty concentrating, behavioral challenges, and loss of language skills [9]. These physical, social, and cognitive changes affect all areas of daily life and, over time, result in an inability to use oral speech or ambulate without mobility aids. Children with JMLD present with typical language acquisition until the onset of disease and experience a complete loss of expressive language between the ages of eight and thirteen [9]. The average life expectancy for an individual with JMLD is under 20 years [10].

Following diagnosis, D, now age 6 years old, presented to the Boston Children's Hospital Augmentative Communication Program (ACP) Fall 2021 for an interprofessional evaluation related to alternative access and augmentative communication with an Occupational Therapist (OT) and Speech-Language Pathologist (SLP). D's initial evaluation focused on supporting his current expressive, receptive, and pragmatic language needs using AAC with alternative access. However, in developing long-term goals, it was imperative that clinicians considered D's changing presentation. A dynamic intervention approach, informed by anticipated disease progression, patterns within his performance using his AAC system, the varied performance skills, client factors, and contexts in which he participated in his daily life, informed long-term recommendations [11].

Occupational Therapy

D has great difficulty with bilateral upper extremity hand use due to ataxia, increasing spasticity, and a fine motor delay. He experienced a regression in motor skills in March 2021, and since then presented with incomplete grasp patterns, increased use of his left hand to compensate for right hand loss of movement, less accurate fine and gross motor movements, reduced active range of movement (to shoulder height), and a discoordinated reach. He relies on seating and positioning systems as he has experienced a change in sitting balance and core strength. Due to regression in his bilateral hand skills, manual direct selection was ruled out to access AT and AAC systems. Switch scanning was also ruled out due to spasticity and ataxic movements that became more pronounced when reaching toward an on-screen target or trying to activate a switch. Upper extremity extensor tone was observed when D was excited or working on challenging tasks. Consistent with D's upper extremity movements, he did not demonstrate consistent control over lower extremity motor movements, which did not provide viable switch sites. Head tracking and head-controlled cursor control were ruled out as D could not coordinate an upright head position or engage in dynamic motor task demands simultaneously. Eye gaze was the most successful, and he accurately selected targets across all quadrants of a screen using dwell selection.

Speech-Language Pathology

D presented with higher receptive language compared to expressive language skills. D used a variety of aided and unaided communication modalities throughout his day. These included facial expressions, body language, gestures, vocalizations, and speech generating device (SGD) use. Using these communication modalities, D demonstrated an understanding of cause/effect, single words, basic categories, simple directions within his physical means, and wh- and yes/no questions related to his immediate wants and needs, past events, and motivating future events. Following his initial evaluation, D was provided with a Tobii Dynavox I-13 SGD with a built-in eye tracking module and TD Snap communication software on long-term loan from the ACP Lacy Lab Lending Library. Using his highly customized 12-location user area and unaided means, D made requests, gained attention, asked and answered questions, participated in communication exchanges, commented, directed activities, indicated pain/feelings, greeted hello/bid farewell, provided information, and demonstrated age-appropriate humor.

Occupational Profile Related to Play

Prior to disease onset, D had no concerns with occupational engagement or self-care and engaged in age-appropriate daily occupations. His day consisted of attending school, socializing with peers, being a son, and exploring areas of interest through play and leisure exploration. His mother reported that D's favorite activities and play topics included: performing magic, learning about animals, independent reading, telling jokes, and drawing. Through informal assessment, it was clear that D had retained these age-appropriate pragmatic skills and a strong sense of humor. Although his change in motor status had affected his ability to engage in play physically, it was essential to preserve participation in meaningful independent, group, and pretend play using his recommended SGD with customized page sets, specialized software, and applications.

PRESERVING PLAY THROUGHOUT DISEASE PROGRESSION

The primary purpose of an SGD is to create speech output as a modality of communication for individuals who do not use oral communication as their primary means of communication due to due-to-motor, -language, -cognitive, and/or-sensory-perceptual-impairments [12]. While communication for children is vital, it is just as essential to give children access to play and opportunities to participate in leisure activities. The capabilities of AT and AAC technologies have expanded significantly in the past decade to allow children with complex physical and communication profiles to participate in play independently and with peers. Interprofessional practice within the ACP clinic allows OTs and SLPs to work together to find AAC systems that increase an individual's ability to communicate, advocate for themselves, and play. Across the literature cited in this paper, there are various accepted definitions and types of play [1-4, 6]. For the purpose of this paper, three domains of play will be explored in the context of interprofessional intervention: independent, group, and pretend play.

Independent play

Independent play is defined as engagement in an activity without an adult or peer facilitation. Before his diagnosis, D enjoyed independently drawing and coloring in his free time. Using specialized software such as Look to Learn® and Magic Eye FX® D once again participated in these preferred activities. Using Look to Learn® software and engaging in the explorative "Graffiti" game, D chose paint can colors and added "graffiti" to a brick wall. Using the same software, D played games similar to his age-matched peers like "Fruit Punch", a target-based game comparable to the mobile iOS application *Fruit Ninja*. D also had access to independent book

reading activities within his TD Snap pageset. Adapted books, created by interdisciplinary teams of OT doctoral students and engineering students in a graduate-level AT class at Tufts University, allowed D to independently read aloud and electronically turn pages. Most off-the-shelf voice-activated smart home devices can understand synthesized speech on SGDs. In the future, it is anticipated that D will be able to access these smart home devices using his SGD to play his favorite songs, ask for jokes, play pranks (e.g., "Alexa, open a box of cats"), control lights, and change the channels on his television.

Group and Social Play

Group play includes the individual and at least one other participant. This type of play allows for natural communication opportunities for all involved. Using pre-made pages on his custom TD Snap user area, D participated in board games and shared reading activities. Using the 'Card Game,' page, D independently participated in the game Go Fish using pre-programmed game phrases including "go fish!", "yes," and "no." Using his Reading' page, D commented on shared read aloud activities (e.g., "I do not like that"), directed the activities (e.g., "turn the page"), and requested his favorite books.

Pretend and Object-based Play

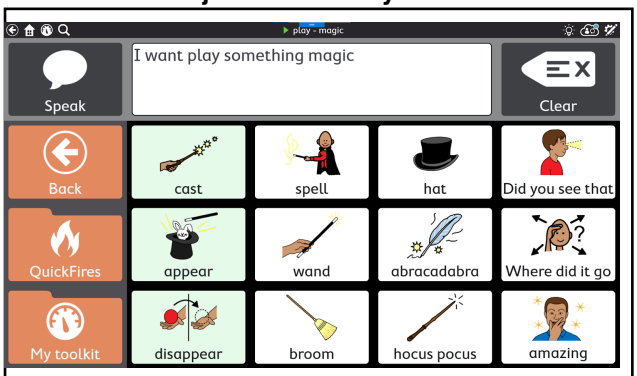

Figure 1. Customized magic pageset in TD Snap

Pretend play can be completed individually or with a peer to engage in imaginative play sequences. Prior to his diagnosis, D enjoyed performing magic tricks for anyone who would watch. Using pre-made 'Magic' pages (Figure 1) on his SGD, D directed others to help him "perform" magic tricks. D directed his play partners to, "cast" + "spell" + "disappear" while the clinician placed a toy car in a bucket. After D said, "abracadabra + Where did it go?", the bucket was emptied out of sight giving the illusion that the car had disappeared. These pretend sequences elicited laughs and happy vocalizations from D as he could again participate in one of his favorite activities. D also directed play partners in object-based play. Using his 'Cars' topic display, D directed actions "crash + fast," asked questions, "did you see that?", and described "fast + car."

Access & Positioning Considerations


Figure 2. Orthoplast hand anchor prototype

As D's condition progresses, changes in his motor and cognition status have to continue to be considered to ensure access to play and communication across domains. Changes in physical motor status, due to increased ataxia and spasticity and reduced active range of motion have already resulted in the need for external positioning supports to successfully engage in SGD use. D's wheelchair has become his primary seating system. A Rehadapt SGD wheelchair mount was recommended with an adjustable mounting system to position the device at a different depth, angle, or height as D continues to grow and experiences changes in his visual field. With a quick-shift plate on the back of his device and his mount in a consistent location, caregivers do not need to continuously recalibrate gaze interaction and can instead quickly position his device for immediate access. His device was positioned for consistent and repeated re-positioning to support independent eye gaze access. D's upper extremity ataxia led to increased uncontrolled motor movements, affecting his ability to engage with his eye gaze device while suppressing extraneous movements. When he was in a seating system with no tray, he compensated for additional movements and decreased postural stability by holding onto a caregiver's hand or reaching around his armrest. While this provided stability, it was not sustainable as he needed someone as external support, or when holding his armrest, his trunk shifted laterally and placed him in a position that affected his alignment with his mounted SGD. Across therapy sessions in the clinic, dynamic intervention offered included custom Orthoplast™ hand anchors to increase upper extremity stabilization. An Orthoplast™ cuff was fit to his thigh, with a 2-3" diameter digit anchor attached via Dual Lock™ (Figure 2). The removable components allowed D's caregivers to modify the system's positioning based on spasticity and changes in grasp patterns. Commercial supports were recommended when he was seated in a seating system with a tray, such as removable grab bars designed to

serve as a shower handle. These are able to be quickly repositioned and support force when D pulls against them for trunk and head stability.

DISCUSSION AND IMPLICATIONS FOR PRACTICE

While evaluating an individual with complex communication needs for an AAC system, it is essential to consider not only communication but also their ability to access play. This is especially true when working with a child with a PMD. As the child's presentation changes, OT and SLP recommended AT and AAC systems need to change with them to provide dynamic intervention as the child experiences declines across physical, cognitive, and/or communicative domains. By including play at the start of an evaluation, we can ensure access to all domains even as clinical focus changes from functional communication to quality of life.

Ultimately, implications for integrating play into SGD use and recommendation include:

- The use of customized page sets and SGD applications can support engagement in independent, social, group, pretend, and object-based play.
- An interprofessional team consisting of SLPs and OTs can address cognitive, linguistic, access, and positioning-based changes to support continued engagement in play with the use of an SGD.
- Play is a human right and needs to be available for all children to engage in as it is a primary occupation of childhood. For children with complex motor profiles, the use of AT can provide access to play and be modified to support changes in access and language for children with PNDs.

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