

**EDUCATING CHILDREN WITH ANGELMAN SYNDROME:
MOVING BEYOND SOCIAL INCLUSION**

by

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A project submitted to the Faculty of Education

In conformity with the requirements for

the degree of Masters of Education

Queen's University

Kingston, Ontario, Canada

(January, 2014)

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Abstract

Angelman syndrome is a genetic disorder that causes significant disabilities. Students diagnosed with this syndrome generally present in the classroom with a lack of oral language, significant developmental delay, a uniquely cheerful personality, and challenges with motor control and motor coordination. This project reviews the literature on Angelman syndrome in order to explain the learning characteristics we would expect to see in these students in the areas of communication, cognition, physical abilities, sensory abilities, affect, attention, and unique medical needs. These learning characteristics are summarized along with a review of instructional approaches that may be appropriate to assist educators to meet the needs of these students. Next, the literature review is summarized in fact sheets collected in an informational booklet for educators. The fact sheets suggest specific instructional strategies to assist educators to meet the needs of students with Angelman syndrome. Finally, the project concludes with a description of how families and educators can re-imagine the needs of students with Angelman syndrome to maximize opportunities for these students to participate in regular classrooms and to access instruction in the general education curriculum.

Dedication

This work is dedicated to my daughter, Maggie, and all individuals diagnosed with Angelman syndrome. I am inspired every day by Maggie's limitless curiosity, buoyant good spirits, and relentless drive for autonomy while developing her own skills and learning. I deeply respect Maggie's strong sense of self and her refusal to engage in work that is not meaningful to her. This project is my attempt to understand Maggie's learning needs in a way that respects both her capacity for learning and her insistence that every learning task have personal value.

This work is further dedicated to all the educators who enact the values of "local knowledge" and the presumption of competence. I am particularly indebted to the researchers and expert educators who generously shared their time, experience, and resources to develop my thinking and answer my questions, particularly Mary-Louise Bertram, Dr. Caroline Ramsey Musselwhite, Dr. Chris Kliewer, Dr. Gretchen Hanser, and Dr. Karen Erickson.

Acknowledgements

The following project, while an individual and original work, would never have been finished without the guidance, patience, and flexibility of my supervisor, Dr. John Freeman. He kept me on track when my many personal commitments were obstacles to finishing this work. He gave me the space to work through my own research questions as new information shifted my focus. His unflinching honesty and high standards for quality writing have dramatically improved this project.

Lou Brown won't remember me, but he challenged me to imagine an inclusive future for Maggie when she was just a toddler recently diagnosed with a devastating disorder that seemed to demand a highly medicalized, deficit-based approach. I am so grateful to Lou and to everyone in the Partners in Policymaking program for their courage and persistence, challenging parents to question our assumptions and beliefs, in order to create opportunities for our sons and daughters. Lou made me start asking the questions that led to my graduate study.

Finally, this work would not have been possible without the support and patience of my family. My husband, Rob Hickey, was always available with priceless logistical assistance as I juggled the demands of this project with our busy family life and many other commitments. My children, Maggie and Ella, patiently endured hours of mom buried in books, busy at the computer, or away at workshops. My mother, Sue Sheldon, provided long-distance encouragement and practical support for housecleaning and childcare so that I could finish this project. Without the love, support, encouragement, and patience of my family, I would never have been able to see this project to completion.

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Chapter 1: Introduction

Last fall, my 10-year old daughter Maggie typed her first decipherable word. Maggie has Angelman syndrome, a chromosome disorder that significantly affects her ability to access ordinary classroom instruction. For the past year, Maggie had been offered daily opportunities to write on any topic of her choice, often selecting a full QWERTY keyboard on her iPad screen as her tool to write with. Maggie's writing usually appears to be random scribbles but her writing portfolio documents her increased awareness of onset rimes, such as "su" and "sa" typed on pages of playing in the sand during the summer, and the appearance of r's, b's and e's typed on a page about the young civil rights leader Ruby Bridges. Maggie was not motivated by her writing task that day last fall, so when one of her best friends in her Grade 5 class arranged her hair in a particularly silly style and giggled with Maggie, Maggie's special educator quickly snapped a picture and offered the picture to Maggie as a topic to write about. Maggie independently typed "mailz," approximating her friend Maisy's name. Chaos erupted in the classroom as classmates and the educational team rushed to exclaim over this first typed word.

This project reflects our journey to that first word, as well as the path we are following with Maggie's educators to ensure many more words follow, along a route towards conventional literacy. This project is a compilation of the resources on which our family and Maggie's team of educators have relied to understand Maggie as potentially literate and as a full learner in her inclusive general education classroom. Meeting Maggie's needs as a student has been a complex and often stressful negotiation with the many professionals in Maggie's life, but we are confident that the research literature on teaching students with Maggie's specific learning characteristics has set her on a path towards conventional literacy. This project is an attempt to share that research base with other families and educators so that the learning needs of students with Angelman syndrome can be met, and these students can access the same educational opportunities as their peers without disabilities.

Our story

On August 14, 2003, a massive heat wave shut down the power grid across the northeast United States and much of the province of Ontario. I didn't even notice; I was in the final stages of the excruciating birth of my first child. My pregnancy with Maggie had been miserable but healthy. My husband and I were excited to become a family and meet our new baby. But the chaos caused by the blackout, combined with such a difficult birth, created a feeling that something was wrong. I felt like a massive upheaval was occurring, as though the blackout was a response to the seismic shift in our lives created by Maggie's birth.

Maggie's birth was complicated, and when she was finally born, she wouldn't cry. She was intubated at birth to protect her airway and was carefully monitored in the special hospital nursery. She didn't make any sound at all those first days. When she finally did cry, it was almost animalistic, a tortured sound wrenched from deep inside her body. I think we all recognized on some level that something was wrong, something was different, but Maggie was otherwise healthy. We went home a few days later, not realizing that her silence was the first sign that she had a profound communication disability.

In the weeks after her birth, it seemed like the drama of the blackout had been anti-climactic. Maggie was the kind of quiet baby that everyone considers "good." She settled in, making us a family, with her seeking only to nurse and sleep and cuddle into our chests. Maggie retreated into sleep at every opportunity; the more noise and stimulation there was, the deeper she appeared to sleep. I remember reading in my baby books about what to expect as she grew. But Maggie didn't make eye contact or babble or respond to her name or smile back at me or look at toys; she didn't do any of the things a newborn was supposed to do in those first weeks. I remember that it felt disloyal to compare my sweet baby girl to a textbook stereotype of what a baby should be doing. I remember slowly putting the baby books away. I remember deciding that Maggie would only be compared to herself, developing on her own time.

As Maggie approached her first birthday, we could no longer ignore that something was wrong. Her development at 12 months old was still that of a young infant. We saw a neurologist who predicted that, based on how she failed to respond to the sights and sounds around her, she was either blind and deaf or profoundly autistic. He asked me if Maggie knew I was her mother. I could only give one example (the fact that she didn't try to nurse with anyone else) as proof that she knew me. He said Maggie was severely developmentally delayed and likely mentally retarded. The neurologist wanted to run tests to rule out a host of horrible conditions. Those tests revealed that Maggie had significant visual, auditory, and sensory processing challenges, combined with a rare, random chromosome disorder called Angelman syndrome.

I remember googling Angelman syndrome from the public library nearest the doctor's office where we received the diagnosis. Angelman syndrome was one of the worst-case scenarios that the genetic tests were supposed to rule out. Among the host of genetic disorders they had tested for, I hadn't paid attention to Angelman as a possibility because Maggie didn't have many of the early signs. She had nursed well; she had neither the feeding difficulties nor the early reciprocal social smiling for which infants with Angelman are known. I remember sitting at a row of computers in that library, Maggie playing quietly at my feet, chewing her fingers and fists, as other library patrons retreated in discomfort while I sobbed as I read about the characteristics of Angelman syndrome.

I read in Angelman syndrome association websites that Maggie would have severe to profound mental retardation and would not speak more than one or two words over her lifetime. She would laugh uncontrollably for hours at a time, would have uncontrollable seizures, and would likely never learn even the most basic preschool skills of dressing herself or using the toilet. I read about children with Angelman who died in freak accidents like getting stuck in recliner chairs or drowning in shallow water. I read that children with Angelman who also have traits of autism (such as a lack of eye contact or joint attention) have more severe symptoms of Angelman; Maggie was already described as profoundly autistic, so she had a worse prognosis than other children with her syndrome. As I read about Angelman, all I could

envision for Maggie's future was a life spent sitting in the corner of a dark institutional room, laughing alone like a maniac. There was nothing I read that suggested she could ever go to school, have friends, make any form of contribution in the lives of others, or live a meaningful life.

The characteristics of Angelman syndrome described in the research literature gave me no greater hope for Maggie. Angelman syndrome is a neurogenetic disorder associated with significant intellectual disabilities, epilepsy, motor impairment, and expressive aphasia or lack of speech (Clayton-Smith & Laan, 2003; Williams et al., 2006). It is a rare disorder, affecting approximately 1 in 15,000-25,000 individuals (Dagli, Buiting, & Williams, 2012; Mertz et al., 2013). Angelman can be caused by one of several chromosome or single-gene errors that disrupt the expression of the UBE3A gene on the 15th chromosome; UBE3A is critical for synaptic development and neural plasticity in the brain. Angelman syndrome is known for its pronounced behavioural phenotype of an exuberantly cheerful demeanour with frequent laughter and smiling (Pelc, Chéron, & Dan, 2008). The defining characteristics of Angelman syndrome are the uniquely cheerful personality combined with a profound delay in expressive language ability, disproportionate to the individual's overall delayed developmental growth.

It felt like the massive power blackout that accompanied Maggie's birth had indeed heralded a seismic shift in everything we thought our life with Maggie would be like. This was not the child I had intended to parent, and this was not the family life I had ever imagined. I was powerless to help Maggie. I was powerless to help ourselves as her parents. I was powerless to change anything as absolute as a missing piece of chromosome. My husband and I read every medical journal article we could find; all of that literature reinforced the idea that Maggie's disability sentenced her (and us) to a life confined by the limitations of Angelman. Reading about Angelman in the published literature was like driving past a horrific car crash and realizing that it was our own family life that we were seeing in the wreckage.

In the years after Maggie's diagnosis, however, another shift occurred. We found families who had older children with Angelman. Many of these families and children seemed to be living lives full of love and hope and participation in ordinary childhood activities. We met girls with Angelman who had

friends and went to birthday parties. A few of these families explicitly rejected the idea that disability was inferior or unnatural to the human experience. I found a research article (Stainton & Besser, 1998) where families described the positive effects they experienced raising a child with intellectual disabilities; I was stunned but hopeful that families were able to find meaning in their child's disabilities. Stainton and Besser's article was the first piece of research that was helpful to envisioning what our family life could become.

I participated in a 10-month parent advocacy training called Partners in Policymaking. The Partners program introduced me to the movement for school and community inclusion for people with significant disabilities. I had believed that some people (such as those with the characteristics of Angelman syndrome) were too disabled to be meaningfully included, but in Partners, I heard that the people with the most severe disabilities needed inclusion the most. I came to understand that inclusion is not just a good thing to do, although it has many positive effects. Inclusion is fundamentally a problem-solving approach to the profound vulnerability created by significant disability and the lack of verbal speech. Abuses against people with disabilities are an abuse of power; this abuse occurs most easily in separate, private spaces with people who are paid to be in control of the person with disabilities, in the absence of unpaid competent witnesses. Inclusion is fundamental to personal safety for a person who cannot give recognizable testimony about her treatment by others. This explanation of inclusion as a tool for personal safety was so compelling that the pursuit of inclusive contexts became the foundation of our long-term planning for Maggie's school and recreational activities.

The Partners in Policymaking program provided advocacy training to parents alongside adults with intellectual and developmental disabilities. In this inclusive context, I had to confront my own bigotry towards people with intellectual disabilities. I ignored the contributions to our discussions from a woman with cerebral palsy because her speech was unintelligible. At some point, though, I emailed with this woman, mistaking her for someone else, and I enjoyed the insights and humour in her emails. When I later realized my mistake, I was shaken by my own prejudice. I was humbled and shamed to realize that I

could understand her speech after all, but only when I believed she had something valuable to say and only when I disciplined myself to listen carefully. I wondered how many people would ignore my own daughter's communication with the same callous assumption I had made that this woman had nothing valuable to share. My participation in the Partners program was the beginning of my realization that the limitations caused by disability are often a socially constructed barrier, not inherent to the biology of the disability itself. I had been powerless to confront Maggie's unusual biology, but bigotry and discrimination were something more tangible that I felt I could influence.

Discovering my own bigotry was particularly shameful because I was an impassioned social justice activist. In the decade before Maggie's birth, I was a full-time community and union organizer. My husband and I had met through our political activism. With exposure to movement leaders like Lou Brown and Kathie Snow, we began to understand intellectual disability through the lens of a social justice movement rather than as biologically determined fate. We began to piece together a vision of what Maggie's life could be like, despite her disabilities, even celebrating her disabilities. We looked to older families who were constructing inclusive lives for their children with Angelman that seemed remarkably ordinary and typical. We began articulating the unexpected but positive effects on our lives of having a child with developmental disabilities.

I was a professional organizer, so I began to apply the framework of organizing to the challenges created by Angelman syndrome. An organizer helps people re-imagine a problem they are experiencing in new ways such that they have the power to influence and address the problem. Organizing is about changing how people think about a problem, so they can change how they respond, to be more effective as they address the problem. Organizing is fundamentally a form of problem-solving that re-defines how a problem is understood in ways that create space for collective action to solve it. Organizing presumes that people are more powerful to address the problems in their lives than they think they are; they just need facilitation to re-orient them to the problem so that they can find their own power. I began to see how the problems associated with Angelman required an organizing approach. For me, this problem-solving

approach meant that we had to stop focusing on Maggie's biology as the problem, and instead focus on how we (the parents and professionals) responded to Maggie's differences. Maybe Angelman was not the problem; maybe the problem was our own inexperience and beliefs about the meaning of her disabilities.

I worked with other mothers of children with Angelman to address the very similar problems our children faced. The most common similarity among our children was often not their syndrome itself, but was, instead, the way that both families and professionals were unprepared to respond to the differences caused by the syndrome. We didn't know what to do differently as a result of our children's differences, but we knew our kids needed us to do something. We wanted to know how to support our children to participate in the same kinds of activities as other children, yet we were instead funnelled towards different activities in different, separate spaces. We needed information and strategies and support. We had a sense that we needed syndrome-specific information to help our children with Angelman grow and learn, yet the only syndrome-specific information was medical literature that was much better at measuring our children's limitations than prescribing effective new approaches. I met older children and adults who had abilities, such as conventional literacy skills, that belied everything I had read was possible according to the published Angelman literature. I decided that the problem was not Angelman syndrome. The problem was that we, the adults, were unprepared to respond to the challenges and differences presented by a child diagnosed with Angelman. As an organizer, I tried to reframe the problem by focusing on what we, the adults surrounding the child with Angelman, need to learn in order to be effective problem-solvers who can support the child's learning.

By this point, Maggie was a student in public school. When Maggie started Junior Kindergarten, it was quickly evident that there was a fundamental disconnect between how I understood the challenges Maggie experienced as a student in school, and how her team of educators understood these same challenges. While her classmates spent those first weeks transitioning to Junior Kindergarten, Maggie was on life support following respiratory failure. Maggie fought her way through a severe medical crisis caused by epilepsy and respiratory failure; she demonstrated she was a survivor. After her illness, Maggie

had to regain every skill; she couldn't even sit up unassisted. Maggie worked tirelessly during her recovery to regain her skills. Maggie never appeared to complain or ask "why me?" and I felt she was a role model of resilience and acceptance. I felt that her educators should be humbled by the challenges she had overcome. I felt that if there was a chance that Maggie's life would be shorter than that of her peers, then her time was precious and we could not afford to waste it. I felt her illness and her many challenges proved that addressing Maggie's needs was an urgent task.

To my dismay, Maggie's school staff focused exclusively on Maggie's fragility, not her resilience. Her educators were content with Maggie's presence but didn't expect her participation. They framed her presence in the classroom as an opportunity to extend their compassion, not as an opportunity to learn from Maggie's example about the traits of resilience and persistence. While Maggie's school team seemed eager to care for her, no one knew how to teach her. Her school work and art projects came home carefully printed and painstakingly coloured without a shred of evidence that Maggie had touched the work herself. I remember urging the educational assistant to take the perfectly assembled mother's day gift home to her own mother; her mother would appreciate to see her daughter's work, just as I would appreciate one day to see Maggie's work. Maggie's role in the classroom was an observer, not a learner. Her educators didn't know how to assess if she was learning. They did not fundamentally believe that she *could* learn. I had thought that we, as a team, would focus together on how to meet Maggie's learning needs. I thought we would explore, together, how Maggie could meaningfully engage in the learning in her classroom. Instead, I had to turn my organizing focus to the learning needs of Maggie's team of educators, to understand what they needed so that they could be prepared to teach Maggie.

As I talked to other parents, it was clear that this was a common problem for our children. Parents almost universally felt that their child's school team did not know what they needed to know in order to be effective teaching the child. Children were either placed in inclusive classrooms with little expectation that they could learn academic skills or participate in a meaningful way, or they were placed in self-contained classrooms with children with similarly severe levels of disability with educational programs

that emphasized basic self-care and functional skills. Our children's behaviour was usually understood as the problem that needed to be solved, rather than problematizing instead their need for meaningful learning and for developing effective communication skills. Everything I read as a parent trying to be informed and involved in Maggie's education suggested that our kids needed to be in print-rich, language-rich environments filled with genuine opportunities to learn how to communicate, read, and write about what mattered most to them. I did not see that occurring in either the inclusive or special classrooms where our children were being educated. I returned to school in the Master's of Education program at Queen's University to learn how to address the problems that our children with Angelman continued to encounter in their school systems and with their teams of educators.

At Queen's, I intended to study effective inclusive practices. I thought I could learn the *how* of inclusion so that I could share it with fellow parents and with Maggie's educators. However, I quickly concluded from the literature that inclusion is less about *how* than *why*. Effective inclusion occurs when the student with significant disabilities is understood as a learner with the need to fully participate in the educational endeavours of the classroom. If students are included for anything less than learning (such as inclusion for social reasons), then their placement is always at-risk and marginalized because they are exempt from the *work* of the classroom, the work of learning. A classroom is social in the context of the shared work of learning; a student who is not there to learn is what Kliever (1998) calls a squatter, a tourist on the margins of the classroom, there to observe the local culture but not to participate as a citizen. We could not move forward knowing how to educate Maggie in the inclusive classroom if we did not fundamentally agree on why she was present in the classroom in the first place. My own conclusion from the research was that we include students with Angelman syndrome in the general education classroom because it is the safest, most language-rich, text-rich context for learning language, literacy, and communication skills. The fundamental work of public education is to teach literacy to every student; thus learning to become literate is the work of school.

This project is the culmination of my attempt to apply the literature on inclusive education, emergent literacy, and communication development to the problem of Maggie's education. Chapter 2 is a review of this literature as it applies to the specific learning characteristics we would expect to see in students with Angelman syndrome. Chapter 3 is a booklet, *Angelman Syndrome for Educators*, that distills the literature review for an audience of educators and families. The booklet is intended to be used either as stand-alone fact sheets on specific topics or as one complete review document. Each section in *Angelman Syndrome for Educators* links to practical resources to help educators and families find the hands-on materials they need to meet their student's needs in the classroom. Chapter 4 is a summary of the experience of creating this project.

This project in its entirety is intended to disseminate what we have learned about teaching Maggie to other teams of families and educators. This project attempts to raise expectations for academic learning for students with Angelman syndrome by problematizing how we have traditionally responded to certain learning characteristics and imagined a "functional" curriculum. This project tries to provide the pedagogical content knowledge that will help educators and families meet these higher expectations for learning. This project will be a success if parents and educators find it useful as they problem-solve the challenges created by Angelman syndrome so that each student can access instruction and maximize his or her learning.

Chapter 2: The Effect of Angelman Syndrome on Children’s Educational Experiences

Students with Angelman syndrome experience significant disabilities. Significant disability refers to students labeled with moderate, severe, or profound intellectual impairment and complex communication challenges (Downing, 2005b). This population of students rarely uses symbolic communication or verbal speech in conventional ways (Calculator, 1999). It is common for students with significant disabilities to have physical or orthopaedic impairments that may require the use of wheelchairs or other mobility aids. Students with significant disabilities often have sensory disabilities such as visual and/or hearing impairments (the latter is not common with Angelman); may have chronic health needs that require management in the classroom (such as feeding tubes or epilepsy); and may be labeled with behavioural challenges. These are the characteristics of students who are the most likely to experience educational segregation (Smith & O’Brien, 2007) and to be deemed by experts and professionals as unable to profit from exposure to literacy instruction and a rigorous academic curriculum (Bentley, 2008; Downing, 2005b). Erickson and Koppenhaver (2007) describe six general areas of difference between students with disabilities and students without disabilities that can impact a student’s success or difficulty with learning: communication, cognition, physical abilities, senses (primarily vision and hearing), affect, and attention. Students with Angelman syndrome would be expected to experience each of these six areas of difference in learning, so that there is a need for strategies to assist educators to problem-solve and respond to each area of difference.

Communication

The most significant impact of Angelman syndrome on a student’s learning is a profound expressive communication disability (Williams et al., 2006) that is disproportionately severe in relation to the student’s overall development, cognitive abilities, and receptive language skills (Gentile et al., 2010;

Jolleff, Emmerson, Ryan, & McConachie, 2006). Individuals with Angelman syndrome have expressive communication skills that are significantly lower than their measurable level of understanding or cognition. Support for communication skill development is likely the single greatest educational need of these students (Calculator, 2013).

While as many as one third of individuals with Angelman syndrome develop some manual sign language, their use of sign language tends to be idiosyncratic (recognizable only to people who know them well within the contexts in which the signs are used) with very few reported to use conventional sign language (Calculator, 2013; Clayton-Smith, 1993). Apraxia (difficulty initiating motor movements) and fine motor disabilities likely contribute to the low level of signing ability in students with Angelman compared to students with comparable levels of cognitive disability (Alvares & Downing, 1998; Jolleff & Ryan, 1993). Parents can be taught to help shape their child's natural gestures into universally recognized gestures that even strangers can accurately interpret (Calculator, 2002).

Norm-referenced standardized communication assessments of individuals with Angelman syndrome rarely describe expressive language abilities above the 12-month age-equivalent or receptive understanding above the 24-month age-equivalent level (Gentile et al., 2010; Williams et al., 2006). However, receptive language assessments are necessarily dependent on the individual expressing her comprehension, and thus may under-report the individual's abilities. The communication disability in Angelman syndrome is exacerbated by intellectual disability and is complicated by co-existing ataxia (tremulous movements and low muscle tone), apraxia (difficulty initiating movement), and difficulty coordinating motor responses, known as dyspraxia (Gentile et al., 2010; Penner, Johnston, Faircloth, Irish, & Williams, 1993; Williams et al., 2006; Williams, Peters, & Calculator, 2009). Verbal speech is rare. Anecdotal reports of spontaneous full utterances are common among families, yet these same utterances cannot usually be performed on demand. This inability to produce or imitate a verbal sound on command is consistent with the nature of apraxia and with Penner et al.'s (1993) observation that individuals with Angelman syndrome cannot imitate even familiar gestures on command. Dyspraxia and

apraxia both become more pronounced when performance is stressed (Cousins & Smyth, 2005). Parents of children with Angelman syndrome complain that traditional norm-referenced or clinical assessment strategies underestimate their child's receptive language and cognitive abilities (Gentile et al., 2010).

Students with Angelman syndrome use a wide variety of communication strategies and, like others with complex communication needs, appear to develop strong preferences for whatever system is the most efficient to convey the messages they are most motivated to express (Calculator, 2013). Expressive communication may be supported by expecting communicative responses in natural contexts, in the course of typical and motivating experiences, rather than expecting students to perform in clinical or unfamiliar settings (Mirenda, 2008). A growing number of parents report anecdotal success with naturalistic uses of aided language systems such as Pragmatic Organization Dynamic Display books (Porter & Cafiero, 2009) but no published research on aided language strategies with students with Angelman syndrome currently exists.

The clinical presentation of an individual with Angelman syndrome is that of a person with strong motivation to engage in communicative interactions but restricted in her ability to clearly and articulately demonstrate or perform her comprehension (Pelc et al., 2008). What a student with Angelman syndrome can communicate should thus be presumed to be the floor, not the ceiling, of her comprehension. Descriptive assessment tools, such as The Pragmatics Profile of Everyday Communication Skills in Children (Dewart & Summers, 1988), can provide educators and families with tools to capture the communication abilities the child has currently mastered and target intervention at areas of communication that are lacking (Downing, 2005b).

Angelman syndrome and autism

The expressive language deficits in Angelman are too severe to be explained by intellectual disability alone and are similar in nature to the disproportionate delays in social interaction skills and language deficits seen in autism (Bonati et al., 2007; Peters, Beaudet, Madduri, & Bacino, 2004;

Trillingsgaard & Østergaard, 2004). There is speculation that Angelman can be understood as a form of autism spectrum disorder, due to the disproportionate severity of the communication disorder combined with marked difficulty or inability to imitate or to engage in joint attention (Penner et al., 1993; Steffenburg, Gillberg, Steffenburg, & Kyllerman, 1996). However, what distinguishes the communication disability in Angelman syndrome from autism spectrum disorders is that individuals with Angelman usually demonstrate strong interest in communicating; their receptive understanding, motivation to communicate, social gregariousness, and interest in reciprocal social interaction is frequently cited as a relative strength even when these traits are reduced due to the presence of autism (Clayton-Smith, 2001; Clayton-Smith & Laan, 2003; Pelc et al., 2008; Williams et al., 2006). Studies attempting to use standardized assessments for autism on students with Angelman have yielded inconsistent results because of questions about whether symptoms of autism reflect very low levels of development versus the pathology of autism itself (Bonati et al., 2007; Peters, Horowitz, Barbieri-Welge, Taylor, & Hundley, 2012). Sahoo and colleagues (2006) compared the size of the genetic error causing the symptoms of Angelman syndrome in 22 children with chromosome deletions to the children's overall clinical presentation. They found that 11 of 22 children with Angelman in their study met the diagnostic criteria for autism; a larger chromosome deletion was a significant risk factor for autistic symptoms, as well as an overall more severe level of cognitive and communication disability and more severe epilepsy (Sahoo et al., 2006). Trillingsgaard and Østergaard (2004) found that 13 of 16 children with Angelman in their study met the criteria for classic autism, while the remaining 3 were described within a broader autism spectrum disorder. Bonati et al. (2007) found the co-morbid presence of autism or the broader autism spectrum disorder in 14 of 23 Angelman patients but questioned whether the patients who met the criteria for autism had adequate communication or cognitive development for the autism diagnosis to be valid. Peters et al. (2004) found a somewhat lower rate of autism in their study with 8 of 19 children with Angelman meeting the diagnostic criteria for classic autism.

In contrast to their peers with Angelman who did not meet the criteria for autism, children who met the criteria for autism were not observed to direct vocalizations at others, to initiate social overtures, to respond to their names being called, or to exhibit as much shared enjoyment in their interactions with others. These children were more focused on the repetitive use of objects and had more impaired play skills. These differences were noted when the children with both Angelman and autism were compared to peers with Angelman only who were matched for cognitive ability, thus suggesting that autistic traits in children with Angelman syndrome are independent of overall developmental growth. The 11 children with Angelman in Peters et al.'s (2004) study who did not meet the criteria for autism had an overall higher level of development, directed their vocalizations effectively at others, and used non-verbal gestures to enhance their communication. Students with Angelman syndrome who did not have autism displayed more evidence of communicative intent, such as establishing joint attention before gesturing or vocalizing, and escalating their communicative signals to achieve a goal. While all students with Angelman syndrome may make unusual use of direct eye gaze, students with autism may appear more aloof and avoid direct eye gaze, while students without autism may have unusually intense eye gaze and make unusually frequent social approaches (Peters, Horowitz, Barbieri-Welge, Taylor, & Hundley, 2012). In Peters et al.'s later longitudinal study (2012), the presence of autism was independent of cognitive ability and global development, such that traits of autism persisted even as a student's global development progressed. Together, these studies suggest that it is likely that a subset of individuals with Angelman syndrome have co-morbid autism and therefore experience more substantial challenges with key aspects of communication such as joint attention, initiating gestures or social overtures, and reciprocal interaction.

The possibility of autism compounding the effects of Angelman syndrome creates additional communication challenges for students with this co-morbid presentation. The literature on autism in Angelman offers few practical suggestions for educational planning for these students, other than a call to provide traditional autism services to these students (Gentile et al., 2010). Applied behaviour analysis

approaches may have a modest effect on cognitive and adaptive skill measures for students with Angelman (Summers, 2012; Summers & Hall, 2008; Summers & Szatmari, 2009).

While several traits of autism are clearly common in students with Angelman, it may be more helpful to understand which autistic traits are *not* reported in Angelman. For example, repetitive sensory motor behaviours are infrequently reported in students with Angelman (Walz, 2007) and appear to be a function of the student's global developmental level more than a symptom of autism (Bonati et al., 2007). These behaviours usually include: hand-flapping when excited or stimulated (often accompanied with smiles and laughter in response to social overtures); mouthing of non-edible objects; and ceaseless movement or pacing. These behaviours tend to decrease over time as cognitive growth occurs (Pelc et al., 2008). In marked contrast to their peers with autism only, students with both autism and Angelman syndrome do not appear to be preoccupied with movement (such as finger/hand flicking or spinning objects), predictability (such as a need for sameness in the environment or in routines), or with order (such as lining up objects); these students demonstrate stronger social reciprocity, such as expressing enjoyment in shared activities and responding to praise and gestures of affection (Walz, 2007). Students with both autism and Angelman syndrome develop cognitively at nearly the same rate as their peers with Angelman syndrome only, but are more severely impacted overall in their cognitive, social, and communicative skills (Peters et al., 2004; Peters et al., 2012). Unlike students with autism only, students with both Angelman and autism score very low on measures of irritability (Peters et al., 2012), suggesting that the unusually cheerful demeanour found in Angelman syndrome is not impacted by the co-morbid presence of autism.

Mirenda (2008) may offer insight into how educators and families might respond to the presence of autistic traits in a student with Angelman. Mirenda suggested that it is time for practitioners in the field of autism and communication disorders to rethink the challenges created by autism by emphasizing autism as primarily a motor disorder of apraxia (difficulty initiating movement) and dyspraxia (difficulty coordinating movement). Drawing on a review of the literature (Dawson & Watling, 2000) demonstrating

that motor planning is substantially disrupted in children with autism (and that greater deficits in motor planning are associated with more severe presentations of autism), Mirenda argued that children with autism who present with the greatest deficits in imitation, eye gaze, ceaseless or uncontrolled movement, lack of spoken language, and difficulty with manual sign language, may be struggling most with an underlying dyspraxia. Interestingly, the traits that Mirenda identifies as possible motor coordination issues in students with autism are many of the same traits that differentiate students with autism and Angelman from their peers with Angelman only. The idea that the communication disabilities in disorders like autism and Angelman syndrome may have a motor basis is further supported by neurological studies that confirm that the hands and mouth are coordinated by the same motor control system (Gentilucci & Volta, 2008). Penner and colleagues (1993) observed significant deficits in both vocal motor planning and in overall motor planning in adults with Angelman syndrome, including the absence of imitation skills and the lack of ability to execute even familiar oral acts on command. Penner et al. (1993) described these deficits as a widespread dyspraxia with particularly severe oral motor impacts; they proposed that a widespread developmental dyspraxia should be understood as a characteristic of Angelman. Extending Mirenda's (2008) argument - that these classically autistic traits might be better explained as a motor planning disorder - to Angelman syndrome may be useful to explore new, possibly more effective, approaches.

Understanding autism as a motor planning disorder opens up opportunity to address these deficits through what Mirenda calls "back-door" approaches that emphasize constant modeling and practice of the motor patterns involved in spontaneous communication, such as through naturalistic modeling of aided language, rather than more traditional performance-focused massed trials. Mirenda argued that the research to date (Drager et al., 2006; Goossens', Crain, & Elder, 1992; Ronski & Sevcik, 1996; Ronski et al., 2010) on various forms of aided language input or modeling in natural contexts provides preliminary evidence that these may be viable and effective language interventions for students with autism with the greatest language deficits. Extending Mirenda's argument to Angelman, the evidence around language

modeling with visual symbols could be an important avenue to explore for families and educators working with students with both Angelman and autism, or Angelman only.

Aided language input

Aided language input relies on the use of symbols to supplement communication with children with complex communication needs. Aided language input strategies build on what we know about language acquisition in typically developing children. Children without disabilities learn complex communication skills through immersion in language-rich contexts, surrounded by fluent language users communicating for a variety of authentic purposes. The average child in a working class home in North America hears approximately 1,250 words spoken per hour, or 6 million words per year (B. Hart & Risley, 1995). Children without disabilities are surrounded by models of oral speech and very slowly begin to experiment with producing the sounds of oral language and participating in verbal exchanges. Typically developing children generally hear this level of language for approximately one year before they produce their first word, but can speak about 1,500 words by the time they turn 3. Thus children without disabilities experience rich immersion in models of language they can observe, comprehend, imitate, and finally express. If children who can speak require deep immersion in oral language to master speech for fluent communication, then aided language input presumes that children who cannot speak require this deep immersion in an accessible visual language. Aided language input attempts to maximize exposure to an accessible visual symbol-based model of communication.

Children with disabilities are frequently exposed to less spoken language than their nondisabled peers (Light & Drager, 2007; Sutton, Soto, & Blockberger, 2002). Children who cannot produce speech experience minimal exposure to the use of visual symbols to communicate; unlike children who can speak, children with complex communication needs are rarely immersed in modes of communication they can observe, comprehend, imitate, and finally express (Drager et al., 2006; C. Goossens' & Kraat, 1985).

The oral language received by children who cannot generate speech does not serve as a useful model on which the children can experiment, imitate, and respond (Goossens' et al., 1992).

Systems of aided language input all share a commitment to problem-solving how children who cannot produce oral speech can still access models of communication that the children can observe, imitate, approximate, and then express. Systems of aided language input include aided language stimulation (Elder & Goossens', 1994; Goossens', 1989), the System for Augmenting Language (Romski & Sevcik, 1996), Natural Aided Language (Cafiero, 2001), and aided language modeling (Drager et al., 2006). What these systems have in common is the presumption that, by observing visual symbols as they are used by communication partners during motivating activities, a child can begin to establish a schematic representation for how visual symbols can be combined and recombined to generate communicative messages (Beukelman & Mirenda, 2012). Communication partners provide aided language input by highlighting symbols on the child's communication display concurrently as they interact with the child verbally; highlighting the symbol supports the child to map the verbal utterance to a concrete symbolic referent (Elder & Goossens', 1994). These aided language stimulation strategies occur in natural settings in the course of ordinary conversation, but the communication partner provides a visual referent for key words as he or she engages with the child who is learning to use the visual symbols. As a result of aided language input interventions, children with significant communication disabilities have increased both their receptive and expressive vocabularies (Dada & Alant, 2009; Harris & Reichle, 2004), have increased their overall symbol use, and have generated more complex messages. These interventions increase the student's participation in group activities and are also effective with teens and adults with significant disabilities (Beck, Stoner, & Dennis, 2009). Aided language modeling in the course of ordinary activities is an intervention that can be effectively implemented by parents (Binger, Kent-Walsh, Berens, Del Campo, & Rivera, 2008; Jonsson, Kristofferson, Ferm, & Thunberg, 2011; Kent-Walsh, Binger, & Hasham, 2010) and school educational assistants (Binger, Kent-Walsh, Ewing, & Taylor, 2010).

Longitudinal studies suggest that aided language modeling approaches may have a significant long-term effect on language development for students with complex communication needs. For example, Ronski and Sevcik (1996) designed a 2-year longitudinal intervention with over a dozen physically mobile school children with moderate to severe intellectual disabilities and little or no verbal speech. The children were provided with devices that activated speech output by touching visual symbols. Parents and teachers were trained to operate the devices and match their verbal utterances with the use of the speech-generating device. Every student in the intervention made significant gains in basic expressive communication skills, and the majority learned to generate multi-word utterances (Ronski & Sevcik, 1996). The gains in communication were still present in a 5-year follow-up study (Ronski, Sevcik, Adamson, & Bakeman, 2005) and significantly surpassed the expressive communication skills of an ability-matched control group. These results were replicated in an experimental study (Ronski et al., 2010) with 62 small children with complex communication needs; the children who received aided language input and had access to aided language output (a speech-generating device) developed significantly more expressive and receptive use of symbolic language than children who received only oral language input.

There is no published research on the use of aided language modeling for students with Angelman syndrome, but the positive results seen in children with autism and other complex communication needs provides a starting point for educators and families to consider how a student's educational program and home environment can expose the student to models of using visual symbols to communicate a range of motivating messages.

Augmentative and alternative communication

Students with Angelman syndrome have complex communication needs, disabilities that prevent them from meeting their daily communication needs through natural modes such as speech, gestures, or

handwriting (Beukelman & Mirenda, 2012). Individuals with complex communication needs require adaptive assistance for speaking and/or writing.

The strategies, services, and technologies that provide this assistance for speaking and writing are provided from within the field of speech language pathology and are known as augmentative and alternative communication (AAC). AAC references both the devices and tools that individuals with complex communication needs might use to communicate, as well as the services intended to support these individuals to learn how to effectively use these tools. AAC technologies and approaches include both *unaided* systems (such as manual signs and gestures) that rely solely on the individual's body to produce a communicative message, and *aided* systems that involve external equipment, such as symbol displays, speech generating devices, and picture boards (Drager, Light, & McNaughton, 2010). Unaided communication strategies are often effective at expressing concrete messages with an obvious referent (such as pointing at food and then pointing at oneself to indicate hunger, or rolling one's eyes to indicate exasperation) but, in the absence of a formal unaided language (such as American Sign Language), unaided systems are inadequate to express abstract thoughts, such as messages related to the past or the future (Beukelman & Mirenda, 2012).

AAC technologies span a range of products from the most sophisticated high-tech dynamic display devices to simple light-tech single message devices. Dynamic display refers to an electronic presentation of symbols that changes in response to user input, such as touchscreen devices that help a user refine her message. A dynamic display device might, for example, organize language in categories, such as a page for "activities" that can link to a page for "toys," within which the individual can find the desired item about which to communicate. Dynamic display speech-generating devices provide linked electronic pages of symbols; pressing the symbols might generate a recorded or text-to-speech electronic message, or might take the user to another page with more options. Pressing a symbol for "go" might, for example: play a pre-determined message like "I want to go"; open a page of possible destinations; or provide possible related messages, such as "I want to go" or "go away." Many dynamic display devices

can play media files (such as selected videos or music) or can link directly to social media or text messaging applications. Dynamic display speech output devices can provide access to thousands of words in a single device. Dynamic display speech-generating devices can also be used by communication partners to provide aided language modeling (Romski & Sevcik, 1996; Romski et al., 2010).

Electronic AAC options can also have a static display of symbols combined with the electronic voice output. A static display of symbols does not change in response to user input, such as a single printed page of symbols or photos. The simplest of these electronic static display AAC tools are devices like the BIGmack, a large button that plays a single pre-recorded message when the symbol or button is activated. Calculator (2013) studied the AAC use of 182 students with Angelman syndrome and found that 16% had never had access to any communication device more sophisticated than a single message BIGmack. Another 30% had never had access to a system that could express more than a handful of recorded messages, such as a Go Talk device, which uses a static display to offer between 4 and 32 pre-recorded messages at a time. Static display electronic AAC options can be used to provide very limited aided language modeling if the key words used in conversation with the child with complex communication needs are among the relatively small number of words that can appear on a static symbol display.

Light-tech AAC options are generally static symbol displays without any form of voice output. Light-tech AAC options include: activity-specific symbol displays (such as a single page with symbols specific to a theme such as play toys or meal options); binders of photos or symbols organized thematically (such as PECS books or photo albums for choice-making); and full books of carefully organized language, such as the Pragmatic Organization Dynamic Display book (PODD; Porter & Cafiero, 2009). Some light-tech AAC options require the communication partner to engage in joint attention with the student as they peruse the symbols and construct messages together, while others are designed with pull-off symbols that the user can remove from the book and take to a communication partner to convey a message, such as delivering a symbol for a glass of water to request a drink. Only the

PODD book is designed to offer as wide a range of vocabulary options as a dynamic display speech-generating device. The static-display nature of the PODD book may lend itself more readily to teaching communication partners and AAC users how to locate symbols in a hard-copy form than a dynamic display speech-generating option (Porter & Cafiero, 2009). The PODD book can be used for extensive aided language modeling and can supplement a dynamic display speech-generating device with similarly organized language.

Despite a wide range of AAC options that are available to support individuals with complex communication needs, most students with Angelman syndrome combine a variety of prelinguistic, nonsymbolic, unaided forms of expressive communication, including vocalizations, eye gaze, idiosyncratic manual signs, facial expressions, and contact gestures (Didden, Korzilius, Duker, & Curfs, 2004; Jolleff et al., 2006; Penner et al., 1993). Some students with Angelman syndrome use photographs or individual symbols (such as PECS cards) to enhance their communication. Only a minority use sophisticated speech-generating devices that allow a student to construct and play open-ended auditory messages with a speech-generating device (Calculator, 2013). However, Calculator (2013) observed that parents of students with Angelman reported that their children were more engaged by dynamic display speech-generating devices than with light-tech options such as symbol boards or PECS books; Calculator reported higher rates of abandonment of light-tech AAC supports than high-tech dynamic display devices.

Calculator (2013) assessed 182 parent reports of their child's exposure to various AAC interventions; his most significant finding was that students with Angelman syndrome, in general, have received neither the technological tools nor the instructional approaches that are considered evidence-based within the field of speech language pathology to be effective at teaching students to use and adopt AAC. When Calculator compared the interventions provided to students with Angelman syndrome to the practices most proven to result in long-term use and acceptance of AAC (Johnson, Inglebret, Jones, & Ray, 2006), only 5% of the practices reportedly used with students with Angelman syndrome were consistent with what could be termed evidence-based approaches. Calculator concluded that students with

Angelman syndrome are experiencing success with AAC tools, “despite marginal exposure to practices thought to foster their use” (p. 155). We cannot draw conclusions about what is possible for students with Angelman syndrome to learn regarding the use of AAC until we have at least exposed these students to effective instructional practices (Calculator, 2013).

Cognition

In the United States and Canada, approximately 1% of students are identified as having an intellectual disability (Beukelman & Mirenda, 2012). Schalock and colleagues (2007) characterize intellectual disability as “significant limitations both in intellectual functioning and adaptive behavior as expressed in conceptual, social, and practical adaptive skills” that originates in childhood (p. 118). Intellectual disabilities occur in different degrees that affect an individual’s rate of learning and the acquisition of adaptive skills: the social and practical skills the person needs to meet the demands of everyday living. Students with Angelman syndrome are described with intellectual disability in the functionally severe range (IQ level of 20-35; Gentile et al., 2010; Williams et al., 2006). Tests of cognitive functioning in patients with Angelman syndrome have commonly relied on standardized measures of infant development, based on a belief that children with developmental disabilities should be assessed using instruments tailored to their developmental age, not their chronological age (Gentile et al., 2010; Peters et al., 2012). Standardized psychometric testing for young and school-aged children with Angelman has assigned these children a developmental equivalent as high as 24 to 30 months, with most younger children with Angelman syndrome closer to a 12-month developmental equivalent (Gentile et al., 2010). A longitudinal study comparing improvement in cognition found that school-aged children with Angelman syndrome progressed the equivalent of approximately 6 weeks of typical infant development over the course of a chronological year (Peters et al., 2004) with all students improving significantly in their cognitive abilities over time.

Conventional, norm-referenced standardized measures of cognition presume a single, linear progression of development (Bagnato, 2005). Children with significant disabilities may, however, follow unusual developmental trajectories as they learn to compensate and strategize for the effects of limited mobility, sensory differences such as visual impairment, ataxia, and more (Downing, 2005). Conventional norm-referenced tests can miss this growth or characterize adaptations to disability as deviance rather than development (Bagnato, 2005).

Conventional tests of global development and psychoeducational functioning are designed for children who are typically developing. When applied to students with significant disabilities, these tests can only diagnose degree of disability, or degree of difference from the norm (Bagnato, 2005). These tests are inherently designed to measure difference and deficits for purposes of diagnosis, not to measure strengths and learning for purposes of educational planning (Downing, 2005).

Conventional norm-referenced tests used in many studies of students with Angelman syndrome compare a school-aged child with multiple disabilities to an infant norm, often conducted in an unfamiliar clinical setting. It is common for children with disabilities to fail to demonstrate a skill in an unfamiliar clinical environment despite mastery of the skill in their usual contexts (Downing, 2005). It is similarly common for children with disabilities to demonstrate an isolated skill in clinical environments but fail to demonstrate that skill in naturally-occurring contexts (Collins, 2012). “Misrepresenting children through mismeasuring them denies children their rights to beneficial opportunities and expectations” (Neisworth & Bagnato, 2004, p. 198). Growing recognition of the limits of cognitive measures such as intelligence quotients has resulted in the development of alternative measures of cognition and growth. These alternative measures are designed to more accurately capture an individual’s comprehension and learning for purposes of planning and providing supports for learning and living. They reflect new constructs of disability that emphasize the interaction between the individual and the environment as a source of disability or as a response to disability, rather than examining only the decontextualized individual.

The construct of disability has evolved from a person-centered trait or characteristic (often referred to as a “deficit”) to a human phenomenon with its genesis in organic and/or social factors. These organic and social factors give rise to functional limitations that reflect an inability or constraint in both personal functioning and performing roles and tasks expected of an individual within a social environment. (Schalock et al., 2007, p. 117)

Our understanding of the appropriate response to significant disabilities in learning, such as those seen in students with Angelman syndrome, has therefore shifted to a problem-solving approach that focuses on what supports or environments ameliorate the effects of the disability.

Government bodies across North America (such as Developmental Services Ontario) have implemented this shift by replacing measurements such as IQ scores with assessment tools such as the Supports Intensity Scale to assess the practical support requirements for individuals with intellectual disabilities (Weiss, Lunskey, Tassé, & Durbin, 2009). The Supports Intensity Scale is prescriptive in that it can be used for planning purposes to describe exactly what supports a person needs to perform a task. The Supports Intensity Scale has been validated as an accurate assessment of the frequency, type, and duration of supports that persons with intellectual disabilities need to perform activities successfully and meet their needs across multiple contexts (Schalock et al., 2007; Weiss et al., 2009).

In educational environments, the Student Environment Task Tools (SETT) Framework is intended to replicate this kind of prescriptive description of a student in the classroom (Zabala, 2005). The SETT Framework examines the inter-relationships among the individual characteristics of the student and her learning needs, abilities, and interests; the demands of the environment in which she must function; the tasks she must accomplish within that environment; and the tools and supports available to both herself and to the individuals who support her. Analyzing the interaction of the student, environment, tools, supports, and tasks is the basis of planning how to then meet the student’s needs (Zabala, 2004). For example, a SETT analysis might indicate that one student can effectively engage in self-selected reading when she receives regular prompts from peers in a cooperative group; another can engage best when she reads an adapted text alongside a peer in a corner of the class; and a third is most successful

when she can use an E-reader with text-to-speech software while wearing headphones. The SETT Framework illustrates that, from a pedagogical perspective, it is more useful for an educator to understand what a student can do when provided with specific tools and supports, as opposed to knowing only what a student cannot do, or does not do, independently. Use of the SETT Framework has been widely adopted by assistive technology professional associations as a best practice for purposes of planning to support learning for students with intellectual disabilities (Wojcik, 2011).

Students with Angelman syndrome make incremental progress in learning over long periods of time and may struggle to perform their learning on command. Students with this profile of learning benefit from formative assessment strategies, such as portfolio assessment (Browder, Spooner, Wakeman, Trela, & Baker, 2006; Kearns, Burdge, Clayton, Denham, & Kleinert, 2006). Portfolio assessment strategies are an effective, validated tool to measure and track learning over time (Browder et al., 2003). These strategies use multiple forms of evidence to construct a picture of a student's growth in learning. Evidence collected in a portfolio might include video footage of student performance, artifacts of student work (with notes, as needed on the context and support required), anecdotal reports, and performance data collected on IEP goals. Portfolio assessment permits educators to capture performance of knowledge that is demonstrated in anecdotal events, in context, rather than requiring performance on demand outside of specific contexts. Portfolios have the potential to help guide and improve instruction (Kampfer, Horvath, Kleinert, & Kearns, 2001) by actively involving students in selecting the work samples to include, engaging students in determining their own goals for learning, and allowing students to help select how they can best demonstrate their learning. Portfolios can be incorporated into daily instruction by including materials about the student's communication system, the student's daily or weekly schedule, and materials developed as part of projects or investigations. Teachers find portfolio assessment useful and viable when data collection is embedded in daily instructional activities, and students are actively involved in the development of the portfolio (Kampfer et al., 2001).

Portfolio assessment may be particularly appropriate to students with Angelman syndrome because of the ease with which visual, concrete materials can be gathered as data demonstrating student learning. Students with Angelman often demonstrate strong interest in visual materials and relative strengths in visual skills (Williams et al., 2010). Families share anecdotal reports of individuals with Angelman that suggest strong visual memories, such as children who appear to have memorized every photo and video in their iPad camera roll; who clearly recognize streets and highway exits and accurately associate them with destinations they may visit only occasionally; or who demonstrate unusual engagement with photos and videos. Educators and families can build on this strong motivation for visual supports by reviewing photos to activate background knowledge on specific topics; supplementing writing and reading tasks with related photos and videos; and creating adapted texts with enhanced photo features. Video modeling and video self-modeling may be an effective strategy to support learning in many students with Angelman syndrome.

Supporting students with Angelman syndrome to effectively form and retrieve memories may need to be a particularly important focus of their educational planning. A mouse model of Angelman syndrome has been used for laboratory research into the syndrome for nearly 20 years; it has proven validity as a model of the cognitive and motor deficits in Angelman (Huang et al., 2013; Jana, 2012). Experimental laboratory research on the mouse model suggests that the cognitive deficits are largely the result of disrupted synaptic plasticity during memory formation; the cognitive disability in Angelman syndrome is largely a memory deficit (Daily et al., 2011; Jiang et al., 2010). Mice with Angelman syndrome demonstrate a disproportionate deficit in experience-dependent memory; in mice, this deficit manifests as a very slow rate of changing behaviour based on experience in water maze and rotarod experiments (Daily et al., 2011; Jiang et al., 2010). While mice with Angelman can learn from repeated experience, they require substantially greater repetitions of the experience at much greater levels of intensity before they demonstrate the ability to retrieve information previously learned. Extrapolating this memory deficit to humans with Angelman syndrome, it is possible that these students need substantial

support for memory formation and memory retrieval with unusually high levels of repetition of particularly intense and engaging learning experiences. One way that educators and families can support memory formation and retrieval is by providing visual supports, such as reviewing pictures of past experiences, video-recording activities as they occur, activating background knowledge with video or photos, and creating access to digital or hard-copy photo albums and video records. The ubiquitous nature of mobile digital technologies makes it possible to create photo and video records of virtually any experience.

Literacy Instruction

The individualized educational programs of students with Angelman syndrome emphasize functional skill development, such as self-care and simple communication skills like requesting preferred items and communicating basic needs (Calculator, 2009). The specific content of a student's "functional curriculum" has traditionally been left open for individual teachers, families, and IEP teams to define on a case-by-case basis (Joseph & Seery, 2004; Zascavage & Keefe, 2004). Parents of students with Angelman syndrome prioritize the development of social relationships, communication skills, and independence above other school outcomes (Leyser & Kirk, 2011). In Leyser and Kirk's survey of 68 parents of children with Angelman, no parent described literacy or academic learning as a goal for schooling; literacy instruction was instead perceived as drawing attention from higher priorities like communication and social skills. However, this concern that literacy instruction competes with social skill development and communication is not supported by the literature (Erickson, Hanser, Hatch, & Sanders, 2009). Instead, for students with the characteristics of Angelman syndrome, comprehensive, systematic literacy instruction may be the most effective way to support language development and the ability to actively and independently participate in social experiences with other people (Browder et al., 2009; Downing, 2005a; Drager et al., 2010; Erickson, Clendon, Abraham, Roy, & Van de Carr, 2005; Ryndak, Ward, Alper, Storch, & Montgomery, 2010).

Comprehensive literacy instruction directly impacts language development. Language and literacy development maintain a reciprocal, mutualistic relationship, where increased understanding in one contributes to cognitive growth and learning in the other (Koppenhaver, Coleman, Kalman, & Yoder, 1991; Teale & Sulzby, 1986). Literacy is an inherently communicative, social practice. Strengthening even the most emergent literacy skills allows students with Angelman syndrome to engage more meaningfully in sharing their own personal experiences and relating to the experiences of others (Browder et al., 2009; Kliewer, 2008). Kaderavek and Rabidoux (2004) suggest that emergent literacy interventions for students with significant communication and intellectual impairments should emphasize the function of literacy (social interaction, such as joint attention to engaging literacy materials and responding to the literacy interactions), over the conventional forms of literacy, such as alphabet knowledge.

Kliewer (2008) describes five “currents” of early or emergent literacy development that demonstrate this relationship among communication, literacy, and social interaction for students with Angelman. Literacy development engages students in: 1) making sense of the stories of others through participation in sharing books, conversation, social interaction, and imaginative play; 2) finding and expressing meaning in one’s own experience, through understanding that one’s own experiences, ideas, and emotions are worthy of expression and can be shared with others; 3) communicating thoughts with graphic symbols to make shared communication more clear and efficient; 4) interpreting the meaning of graphic symbols used for expression by others; and 5) deriving joy from critical, reflective engagement with printed language and other graphic symbol systems, which creates the motivation to develop more sophisticated use of graphic symbols (Kliewer, 2008). Each of these currents of literacy illustrates the symbiotic relationship among social interaction, communication, and literacy.

Conventional literacy is the ability to read and write, the cognitive processes of comprehending and composing meaning in written texts (Erickson et al., 2009). Emergent literacy is the behaviours, understandings, and exploration of literacy materials that precede and develop into conventional literacy skills. Most students with the learning characteristics of Angelman syndrome have only early levels of

literacy awareness (Erickson et al., 2009). Emergent literacy behaviours include: observing and imitating the functional use of print, such as when a child pretends to read stories to her dolls or writes a scribbled list of items to get at the grocery store; browsing through books while observing the conventions of print, such as left to right directionality of words and pages; “drawing words” and “writing pictures” to re-tell a story (Kliewer, 2008); and learning that text carries meaning, and that print represents words that we use while speaking and listening (Erickson et al., 2009).

A handful of students with Angelman syndrome are conventional readers and writers, but most students with Angelman syndrome have emergent literacy skills; they may explore books and magazines from back to front or upside down. They may look at the pictures on the page rather than attend to the written text. Some of these students may not appear to attend to, or comprehend, shared story-reading. The marks that most of these students make with a pencil on paper may not appear to have a relationship to the alphabet or even to drawing pictures. Emergent literacy behaviours evolve as a result of opportunity and experience: “Children’s demonstration of emergent literacy seemed much less dependent on cognitive capability than on learning opportunity, modeling of possible uses of print and communication symbols, and access to supportive texts and technologies,” (Koppenhaver, 2000, p. 273). These emergent literacy behaviours can be assessed and scaffolded into more sophisticated understandings of literacy through comprehensive instruction. The approaches that most benefit the emergent literacy of typically developing young children have a substantially similar effect for students with significant intellectual disabilities, particularly when specific supports such as modelled language and augmentative communication devices for students with communication disabilities are integrated into daily literacy experiences such as shared reading and journal writing (Erickson et al., 2009).

Literacy instruction for students with Angelman syndrome should include all the components that are necessary for students without disabilities to learn to read and write, with the purpose to “build core skills and understandings across reading, writing, and communication so that students can move towards conventional uses of literacy to convey meaning to others” (Erickson & Clendon, 2009, pp. 195-196).

Conventional literacy skills provide access to the graphic symbol set of the alphabet, the most efficient, universal, and comprehensive symbol system with which a person can exercise control and autonomy over his or her life (Erickson et al., 2009). Even if conventional literacy skills are not achieved during the school years, evidence-based comprehensive literacy instruction will support the development of communication, social skills, and independence for these students. Literacy skills for students with Angelman syndrome should be functional in that these skills develop the capability of reading and writing at a level proficient enough to conduct one's daily affairs (Hatch, 2009). A functional approach to literacy for students with Angelman syndrome would support these students to 1) make sense of the stories of others through sharing books and social interaction; 2) share and express their own experiences, ideas, and emotions; 3) communicate their thoughts with graphic symbols; 4) interpret the meaning of graphic symbols used for expression by others; and 5) derive joy from their engagement with printed language and other graphic symbol systems (Kliewer, 2008). This functional literacy approach is consistent with the goals that parents of students with Angelman syndrome describe for their son's or daughter's education, while providing sufficient guidance to outline the framework of a comprehensive literacy program for these students.

Comprehensive instruction develops and integrates the skills of oral language and vocabulary, phonological and phonemic awareness, automatic word recognition, fluency, comprehension, writing, and reading independently for a variety of purposes (Allor, Mathes, Roberts, Jones, & Champlin, 2010; Erickson & Clendon, 2009; National Institute of Child Health and Human Development, 2000). This instruction includes frequent opportunities for students to engage in self-directed reading of a wide variety of texts and numerous opportunities to engage in writing for meaningful purposes. Browder and colleagues (2006) found in their review of the research that students with significant disabilities receive instruction largely in word identification (automatic sight word recognition) to the exclusion of other areas of literacy instruction. Yet numerous case study reports demonstrate that students with significant disabilities can make significant improvements in their abilities to read, write, and communicate when

provided with comprehensive literacy instruction (Erickson, Koppenhaver, Yoder, & Nance, 1997; Ryndak, Morrison, & Sommerstein, 1999).

While all aspects of a comprehensive program are important, an emphasis on writing may be particularly useful for students with Angelman syndrome to support expressive communication. For students who cannot use oral speech, the cognitive, memory, and physical demands of using an AAC device directly parallel the cognitive, memory, and physical demands of nondisabled students as they develop into writers (Clendon, 2006). Beginning AAC users demonstrate many of the same challenges experienced by beginning writers as they attempt to use tools to translate and encode their ideas into symbolic text. For students with Angelman syndrome, writing is a process of functional expressive communication. Clendon (2006) suggests that, when planning access to efficient and specific language for students with complex communication needs, educators should prioritize the language that beginning writers use to write, rather than the much larger oral vocabulary that same-age students use to speak. The process of writing helps students learn to read (Cunningham & Allington, 2006). Writing may also play an important role in assessment for students with significant intellectual disabilities; emergent writing may be the best indicator of developing literacy skills in students who do not speak (Erickson et al., 1997; Koppenhaver & Erickson, 2003; Wolf & Hogan, 2002).

Students with the learning characteristics associated with Angelman syndrome have not typically been considered candidates for the comprehensive literacy instruction that is the norm in general education classrooms (Fossett, Smith, & Mirenda, 2002; Kliewer et al., 2004). There is a convergence of evidence from qualitative (Koppenhaver & Erickson, 2003; Skotko, Koppenhaver, & Erickson, 2004), quantitative (Allor et al., 2010; Browder, Ahlgrim-Delzell, Courtade, Gibbs, & Flowers, 2008; Browder, Lee, & Mims, 2011; Erickson et al., 2005; Hanser & Erickson, 2007; Hedrick, Katims, & Carr, 1999), and descriptive case study (Erickson et al., 1997; Light, McNaughton, Weyer, & Karg, 2008; Ryndak et al., 1999) research that students with severe and profound intellectual and communication disabilities can show demonstrable gains in literacy skills when they receive the comprehensive and systematic literacy

instruction that is considered necessary and evidence-based in general education classrooms. Students with disabilities as severe as Angelman syndrome who received long-term (12-24 months) systematic instruction in comprehensive literacy programs that reflected all the major components of effective literacy instruction for students without disabilities (NICHD, 2000) made significantly more progress in literacy skills than control groups who received only traditional sight-word instruction (Allor et al., 2010; Browder, Ahlgrim-Delzell, Flowers, & Baker, 2012). Longitudinal research demonstrates benefits for literacy acquisition from education in an inclusive context (Erickson et al., 1997; Ryndak et al., 1999). Reviews of the literature have summarized the most effective practices to develop literacy in students with severe disabilities (Browder, Wakeman, Spooner, Ahlgrim-Delzell, & Algozzine, 2006; Koppenhaver, Hendrix, & Williams, 2007), students who rely on augmented or alternative communication (Bailey, Angell, & Stoner, 2011), and students with significant disabilities in general education contexts (Erickson et al., 2009).

Students with disabilities as significant as Angelman syndrome appear to learn literacy skills in a similar manner as their non-disabled peers but need more systematic, intensive instruction over longer periods of time to maximize their experiences engaging in literate acts for meaningful purposes (Allor et al., 2010; Browder et al., 2012; Erickson & Koppenhaver, 1995; Kliewer & Landis, 1999; Mims, Hudson, & Browder, 2012). Koppenhaver and Erickson (1998) explained that:

If we reduced our research to a single conclusion, it would be, “Good instruction is good instruction.” The greatest difference we find to date is that children with disabilities, particularly severe or multiple disabilities, require more conscious and careful consideration than children without disabilities in thinking through how to make good instruction accessible given the nature of their individual differences in communication, cognition, attention, behavior, sensory, or physical abilities. (p. 1)

For students with Angelman syndrome, this “good instruction” needs to be paired with strategically designed assistive technologies (including aided language supports and AAC) and instructional accommodations for their specific learning differences (Erickson & Clendon, 2009; Erickson, Hatch, & Clendon, 2010). Students with developmental and communication disabilities require more literacy

instruction and more time engaged in literacy activities yet instead receive significantly less literacy instruction and have access to considerably fewer literacy experiences than their typically developing peers (Browder, Wakeman, et al., 2006; Kliewer & Biklen, 2001; Light & Smith, 1993). The lack of language and literacy development in children with significant disabilities may reflect their failure to access effective literacy instruction and experience meaningful literacy experiences (Browder et al., 2009; Erickson et al., 2009; Kliewer, Biklen, & Kasa-Hendrickson, 2006; Mirenda, 2003). Browder and colleagues (2009) suggested that the only way to determine which students can learn to read is through teaching reading skills. Extending Browder's argument to Angelman syndrome, the only way to learn the literacy potential of students with Angelman syndrome is by providing these students with systematic, comprehensive literacy instruction.

Literacy development is positively affected when the student is immersed in genuine opportunities to speak, listen, read, and write in contexts that are real, of high interest, and literacy-need provoking for that individual student (Koppenhaver & Yoder, 1993). Students are most motivated to learn to read when they perceive literacy as a necessary tool to support their own activities. Students with significant disabilities benefit from approaches that: 1) deliver instruction within activities that are functional to the individual student; 2) provide instruction in contexts with nondisabled peers; 3) provide the student with adapted materials and individualized instruction; and 4) blend functional learning goals with general education content (Ryndak et al., 1999). The general curriculum in reading and literacy already addresses the components of a comprehensive literacy program. Erickson et al. (2009) concluded that "If the ability to read, write, and communicate is the ultimate goal, then we must learn how to maximize access to the entire general curriculum in literacy and reading while providing comprehensive instruction that addresses the individual needs of each student with significant intellectual disabilities" (p. 128).

Parents of students with Angelman syndrome may need explicit support to engage their child in home literacy experiences. Families of children with significant disabilities engage their children in fewer

literacy experiences and provide fewer literacy supports than typically developing children, such as less exploration of age-appropriate accessible books and shared storybook reading (Light & Smith, 1993). Caregivers of children with Angelman syndrome may simply lack leisure time to engage in literacy experiences due to their child's increased health care and caregiving needs. Speech language pathologists can help prepare aided language displays or simple voice output devices that provide students with a way to comment on and direct shared storybook reading. Occupational therapists can help families make books physically accessible with adaptations like page "fluffers" and can help students access electronic texts through touchscreen technology or switch access. Special educators can help families find age-appropriate engaging adapted texts and help write personal experience stories with repeated lines and predictable text.

The educators of students with Angelman syndrome may also require specific supports to develop the literacy potential of these students. Educators may lack expertise in assessing emergent literacy behaviours and in designing systematic, comprehensive literacy interventions for those students who do not use oral language. These educators may require professional development to access the applied professional literature on comprehensive literacy approaches for students with the most emergent literacy behaviours.

There is no published research on literacy development in students with Angelman syndrome. At the very least, it is premature to conclude that students with Angelman syndrome cannot achieve progress towards conventional literacy. At the least, comprehensive literacy instruction for a student with Angelman may improve the student's ability to engage in communicative and social interactions around print and print-related materials. At its best, such instruction may result in the development of conventional literacy skills, strong interests in reading and writing activities for recreation, and more effective means for independent communication (Beukelman & Mirenda, 2012).

Physical Abilities

Angelman syndrome is associated with significant physical disabilities (Clayton-Smith, 2010; Williams et al., 2006), such as jerky uncoordinated movements, disrupted muscle tone, ataxia, poor fine motor skills, and a balance disorder that is frequently referenced but poorly explored in the Angelman literature. All of these physical disabilities impact how students with Angelman syndrome engage in learning experiences, but families and educators can problem-solve to ameliorate the effects of many of these physical impairments.

Young children with Angelman have delayed motor skills. Achievement of independent sitting is usually reached soon after the first birthday and independent walking generally occurs between age 3 and 6 (Clayton-Smith, 2001; Williams et al., 2006). By the time children with Angelman reach school age, most can walk independently, usually with a distinctive, broad-based gait with upraised arms and flexed wrists. However, there is significant variability in when children with Angelman syndrome reach these gross motor milestones; 10-15% of individuals with Angelman never walk independently.

The physical impairment observed in Angelman syndrome is often unrelated to the child's cognitive and global development. While children with higher cognitive skills frequently have the mildest physical disabilities (Gentile et al., 2010), children with the greatest physical disabilities often have greater ataxia, more disrupted muscle tone, and a more significant balance disorder. Milder motor control impairment may permit some students to develop stronger motor skills and perform better on cognitive tasks than their peers with Angelman who experience more severe motor control impairment. The presence of severe physical disabilities observed in an individual with Angelman syndrome should not be presumed to reflect more severe cognitive disability.

The characteristic gait associated with Angelman is an adaptive response to the ataxia, motor planning, and balance disorder associated with the syndrome; it is a strategic response to stabilize posture while walking (Dan & Chéron, 2005). Children with Angelman organize their motor movements in a distinctively different pattern from typically developing toddlers. Typically developing toddlers learn to

walk using highly stereotyped but flexible movements intended to maximize efficient energy expenditure, while easily adapting to the constraints of their environment and pursuing their interest in various activities. Walking in typical toddlers is characterized by automaticity such that, once mastered, the child does not need to exert conscious effort to stay upright and moving. In contrast, children with Angelman syndrome develop independent walking while preoccupied with maintaining postural stability in the face of ataxia, disrupted motor planning, and abnormal muscle tone (Dan & Chéron, 2005). Children with Angelman syndrome may be much slower to develop automaticity in their gross motor skills as a result; some may always need to exert intense conscious effort to maintain stability while walking. Walking, therefore, may be much more physically and mentally taxing for these children; the greater effort required for walking will impact their stamina and the development of more advanced gross motor skills such as running or climbing stairs.

Children with Angelman syndrome demonstrate a “central dyscoordination resulting in difficulties in positioning the body and interacting with the environment” with particular problems in sensory-motor integration (Beckung & Kyllerman, 2005, p. 143). Dan and Chéron (2004) investigated this dysfunction further by using electrophysiological recording to study the postural impairment in 14 school-aged children with Angelman compared to 18 typically developing controls (Dan & Chéron, 2004). All patients with Angelman syndrome (and none of the controls) demonstrated brief high amplitude rhythmic bursts of activity indicative of cerebellar dysfunction related to motor control, particularly the processing of motor commands. These findings are consistent with Penner et al.’s (1993) characterization of Angelman syndrome as a widespread developmental dyspraxia. Thus the physical disabilities associated with Angelman syndrome should be understood as the result of disrupted motor planning combined with a poorly defined balance disorder.

Muscle tone is frequently disrupted in Angelman syndrome but presents in different clusters of symptoms. Half of all infants with Angelman have reduced muscle tone and about one-quarter continue to experience moderate to severe global hypotonia throughout childhood (Thibert, Larson, Hsieh, Raby, &

Thiele, 2013). These students have reduced muscle tone and will need supportive classroom seating to reduce fatigue and maximize postural stability. Depending on the severity of their individual balance disorder, these children are likely to be the slowest to gain gross motor skills such as independent sitting and walking, and may require the greatest supports for independent mobility. Helping these children achieve autonomy over their bodies may need to emphasize alternatives to walking, such as giving these students the tools to direct others where to take them, rather than waiting until these children have sufficient muscle tone to independently navigate to desired locations. Motorized wheelchairs and scooters should be considered as potentially emancipatory tools to help these students achieve independence and autonomy over their bodies.

About one-third of students with Angelman syndrome present with a mixed muscle tone disorder, with truncal hypotonia and hypertonic spastic lower limbs, resulting in a stiff gait and jerky arm movements (Clayton-Smith, 2010; Thibert et al., 2013). These students may also require supportive classroom seating to counter fatigue and to help coordinate motor movements, so that the children aren't simultaneously trying to stabilize their trunk while managing fine motor tasks. Children with mixed muscle tone or globally high muscle tone often appear physically restless, as though finding a comfortable sitting posture is challenging for them. Various adaptive seats, particularly those with high-quality supportive foam, may support these students to sit comfortably so they can better attend to classroom activities. Seating options that could allow them to relax their bodies and focus on fine motor tasks might include dense foam seats, posture-supporting seats, hammocks, rocking chairs, and beanbag chairs. Occupational therapists, parents, and educators can trial each of these seating options to see if any particular option conveys a benefit on the performance or participation of specific classroom activities.

Children with Angelman syndrome frequently demonstrate an intention tremor (Dan & Chéron, 2005; Williams et al., 2006). This tremor appears when the child is concentrating on a motor task. The tremor appears to be strongly correlated to the bursts of electrical activity noted by Dan and Chéron (2005) when children with Angelman syndrome are attempting to control motor movements. This

intention tremor is not of epileptic origin. However, these bursts of electrical activity are not typically seen when the muscle is at rest. If a student with Angelman syndrome presents with a constant or resting tremor, this condition should be investigated as potentially epileptic activity (Dan & Chéron, 2005).

Students with Angelman syndrome generally demonstrate weaker fine motor skills than gross motor (Williams, Driscoll, & Dagli, 2010). The fine motor disabilities associated with Angelman syndrome are generally significant. Jerky or spastic arm movements and large, sweeping motions of the hands are common in children with Angelman syndrome, who may struggle to control and target the use of their fingers and hands (Clayton-Smith, 2001). Poor fine motor skills, such as uncoordinated movements of the hands, may co-exist with sensory integration deficits such as tactile defensiveness. This difficulty can result in the presentation of students who resist tasks that involve grasping, such as using writing tools. These students may benefit from adaptations that remove the need for grasping, or contact with the palm of the hand, to execute tasks. Light-tech eye-gaze boards can be used for students who cannot, or prefer not, to touch tactile surfaces; these eye-gaze boards can be used for classroom activities as diverse as making simple choices or as complex as composing a piece of writing (Hanser, 2010). High-tech supports for these students may include touchscreen computers and handheld tablets in lieu of traditional writing instruments.

The motor disabilities associated with Angelman frequently cause educational teams to prioritize the development of functional motor skills, such as walking, over academic learning (Calculator & Black, 2010; Leyser & Kirk, 2011). However, the motor disabilities in Angelman should not be mistaken as a purely developmental delay. Students with significant motor disabilities may have relative strengths in cognitive skills, attention, and communication. Their impaired mobility can have the side effect of making the student more available for seated academic work over longer instructional periods. Instruction in communication and language development should not be delayed until the student achieves a certain baseline of gross or fine motor skills, as this “readiness” approach may have the unintended consequence of denying students with greater dyspraxia access to academic instruction.

Dyspraxia, complex motor planning difficulties, can profoundly inhibit the initiation and continuance of what would normally be volitional movements (Donnellan, Hill, & Leary, 2013; Donnellan, Leary, & Robledo, 2006). Students who experience these kinesthetic dilemmas (Bauman & Kemper, 1994) may benefit from well-established special education strategies such as backward chaining, system of least to most prompts, and naturalistic predictable scripted sequences so that the final step that requires initiation can be part of a flow of activity rather than a sudden performance demand (Collins, 2012; Donnellan et al., 2013; Kliever, 2008).

Senses

Sensory impairment, such as hearing loss or visual impairment, is not reported in the medical literature as a consistent feature of Angelman syndrome. However, sensory processing disorders may be under-diagnosed and directly impact a student's ability to access instruction in the classroom. Angelman syndrome is primarily a neurological disorder; most sensory differences observed in children with Angelman are neurological in origin. These differences relate to how the brain processes and integrates information received through the senses rather than with dysfunction in the anatomical organs that perceive sensory stimuli. Examination of students with Angelman syndrome in the medical literature has focused on whether the children have normal scores on the anatomical organs of their eyes and ears (Williams et al., 2006), not whether these students are effective at integrating and using the information they see and hear. The neurological rather than physical basis for these differences in perceiving sensory stimuli does not change the significant effect these differences can have on the ability to participate in classroom learning experiences.

Angelman syndrome does not appear to increase the risk of hearing loss (Williams et al., 2006). Children with a history of frequent ear infections should be monitored by the appropriate medical professionals for complications that could lead to hearing loss. Students who have symptoms of hearing loss but have normal scores on measures of hearing may have an auditory processing disorder, a

neurological cause of impaired hearing (Millet et al., 2012). Peters et al. (2004) described unusual or impaired auditory attention, such as not responding to their name being called, as a symptom of autism in students with Angelman syndrome; these differences in attention to auditory information might also be explained by auditory processing challenges and should be investigated as such (Millet et al., 2012). Auditory processing disorders can significantly affect the development of communication skills if affected children struggle to distinguish the sounds of oral speech from background noise. Targeted interventions can accommodate these auditory processing challenges. Distractibility and inattention can be improved by increasing the auditory signal to noise ratio, such as through using FM systems, acoustic modifications, and preferential seating to amplify the teacher's voice. Visual supports and "chunking" instructions can support memory and improve listening comprehension.

Visual impairment is more commonly reported in Angelman. Strabismus (poor alignment between the eyes) affects as many as 80% of individuals with Angelman and may require surgical correction (Michieletto, Bonanni, & Pensiero, 2011). Strabismus can reduce visual acuity, the accuracy with which the physical organ of the eye can perceive visual information. Strabismus can be caused by physical conditions (such as muscle weakness while coordinating eye movement) or can be a neurological symptom of the broader motor coordination dysfunction associated with Angelman syndrome. Low vision is not commonly reported in Angelman, but many children require regular monitoring of strabismus. Most children with Angelman have astigmatism and far-sightedness significant enough to require corrective lenses (Michieletto et al., 2011), while a subset of children have cortical vision impairment.

Cortical visual impairment (CVI) is the result of disturbed or reduced vision due to disrupted neurological functioning. CVI is distinct from disturbed or reduced vision caused by problems with the mechanical features of the eye as a physical organ, such as with astigmatism (Roman et al., 2010). There is no research examining the incidence of CVI in children with Angelman syndrome, but the characteristic behaviours associated with CVI are commonly reported in children with the most severe presentations of Angelman. CVI is more common in children with intellectual disability than in those

without, and may be under-diagnosed in children with significant disabilities due to the challenge inherent in assessing visual acuity and visual processing in students who cannot report on what they see, or whose use of their vision appears to be a function of their overall low levels of development (Nielsen, Skov, & Jensen, 2007). As many as 20% of children with significant disabilities have CVI; major risk factors include severe intellectual disability, epilepsy, and underlying neurological dysfunction (Swaminathan, 2011).

CVI is diagnosed based on three criteria: the clinical finding that visual acuity alone is insufficient to explain the functional limits of the student's vision; a history or co-existence of a neurological problem; and specific visual behaviours and delays in a student's visual development consistent with CVI. Angelman syndrome is associated with a constellation of disorders that elevates the risk of CVI. The symptoms of CVI can be mistaken as a function of overall cognitive delay or as autistic features, but the presence of specific visual behaviours should trigger an assessment by a vision expert.

These specific visual behaviours include:

- unusual use of eye gaze, such as avoiding eye contact, appearing to rely on peripheral vision, or appearing to “look past” whatever object or person the child is interacting with;
- unusual visual responses to motion, such as attending only to moving versus stationary objects;
- apparent day-to-day variation in visual functioning;
- signs that visual information interferes with or does not appear to support movement, such as when a student appears not to look at objects while reaching for them or does not look at objects on the ground while navigating around them;
- poor depth perception;
- unusual sensitivity or attention to light; and
- unusual sensitivity to auditory stimuli, including the need for auditory cues to recognize visual stimuli, such as a child who does not appear to recognize familiar people until she hears their voice (Roman-Lantzy & Lantzy, 2010; Roman et al., 2010).

CVI affects the presentation of co-morbid disabilities and can slow development in mobility, communication, social interaction, and literacy (Roman-Lantzy & Lantzy, 2010). Managing the symptoms of CVI without accommodation may require a child to utilize all her cognitive resources just to make sense of visual stimulation; this increased attention may leave her unavailable to concentrate on other stimuli and can result in an apparent inability to engage in joint attention. Cortical vision impairment combined with auditory processing dysfunction can mimic the presentation of autism but may require a substantially different therapeutic response (Millet et al., 2012). A vision specialist can recommend adaptations and accommodations that will enhance the student's ability to process visual information (Roman et al., 2010).

Angelman syndrome impacts how affected individuals receive and process input from multiple senses. Sensory integration or sensory processing refers to “the neurological process that organizes sensation from one's own body and from the environment and makes it possible to use the body effectively within the environment” (Ayres, 1972, p. 11). Sensory integration includes how the brain processes information from the classically recognized senses of vision, hearing, touch, smell, and taste, as well as the less conscious senses of proprioception (awareness of one's body in space) and the vestibular sense (one's sense of balance and movement) (Ayres, 1972). Disruptions in sensory processing can cause an individual to under- or over-perceive sensory information, which can result in maladaptive behaviours (Walz & Baranek, 2006). Occupational therapists support students with Angelman syndrome by screening for sensory processing disorders and developing effective therapeutic responses.

There is limited research on sensory integration in Angelman syndrome, but many of the diagnostic characteristics commonly observed in these students suggest an underlying global disruption in how sensory input is perceived (Walz & Baranek, 2006). Disruption in sensory processing (particularly of vestibular and proprioceptive information) is consistent with challenges with motor planning and presents as dyspraxia (Ayres, 1985), a feature of Angelman (Penner et al., 1993). Students with Angelman syndrome exhibit a high rate of behaviours such as excessive chewing and mouthing of non-food objects,

increased sensitivity to heat, ceaseless movement, and attraction to objects with properties that provide high levels of sensory input, such as crinkly plastics, mirrors, and water (Didden, Korzilius, Sturmey, Lancioni, & Curfs, 2008; Williams et al., 2006).

Walz and Baranek (2006) studied 340 individuals with Angelman syndrome by analyzing standardized caregiver questionnaires about their child's sensory behaviours. The researchers concluded that individuals with Angelman exhibit a high rate of symptoms of sensory dysfunction, with a particularly high rate of hypo-responsiveness or under-response to sensory stimuli, suggesting these individuals need more frequent and more intense sensory input to adequately process and respond to their environments. Individuals with Angelman demonstrated abnormal sensory responses to the senses of touch, hearing, vision, smell, and the sensation of movement and awareness of the body in space. Some of these symptoms suggested hyper-responsiveness to specific stimuli, such as aggression or distress when touched by others, aversion to touching certain textures or to tasting specific textures or flavours in food, gagging in response to smells, and a pronounced startle to unexpected sounds. Many symptoms of sensory dysfunction can be disruptive to classroom learning, such as gagging, mouthing objects, and ceaseless movement. The authors conjectured that, while some of the sensory dysfunction reflects a global sensory processing deficit, some of these sensory seeking behaviours are symptoms of the physical disabilities associated with Angelman: physical impairments combined with ataxia may contribute to limited physical activity and leave these individuals seeking greater sensory input.

Occupational therapists assist school teams to assess the sensory needs of students with Angelman syndrome to provide adequate supports (Walz & Baranek, 2006). Planning around students' sensory needs can help these students be successful in the classroom by minimizing the sensory experiences that may be perceived as distracting or aversive, and maximizing opportunities to engage in appropriate sensory exploration. School teams can plan how to maximize movement exploration and outdoor play, incorporate materials with sensory-rich properties into school assignments, and support the development of healthy self-regulation. Walz and Baranek (2006) suggest that, at the least, awareness of

the student's sensory needs contributes to a positive reframing process whereby educators and parents can better understand why an individual engages in a specific sensory behaviour. This positive reframing can position parents and educators to better support students to shape their sensory seeking into more appropriate activities.

Affect

One of the most unique and consistent features of Angelman syndrome is the behavioural phenotype. Individuals with Angelman syndrome are characteristically described with a "happy" or cheerful personality with easily provoked laughter (Clayton-Smith, 2010; Williams et al., 2006). Pelc et al. (2008) describe these individuals as socially gregarious, with profuse smiling, and prone to proactive social contact and general exuberance. Laughter and smiling in children with Angelman syndrome is most prevalent in highly social contexts (Oliver, Demetriades, & Hall, 2002; Oliver et al., 2007). Individuals with Angelman score very low on standardized measures of irritability, lethargy, and withdrawal compared to children with similarly severe levels of disability or autism (Clarke & Marston, 2000; Peters et al., 2012; Summers & Feldman, 1999). Compared to children with other genetic syndromes, children with Angelman demonstrate resilient good humour and are less likely to argue, cry, express hurt feelings, or be overly sensitive to criticism (Walz & Benson, 2002). Most children with Angelman syndrome demonstrate strong behavioural flexibility and are better able to adapt to changes in environment, personnel, and scheduling than children with similar levels of intellectual disability or autism (Didden, Sigafoos, et al., 2008). Clinicians and parents of children with Angelman describe the children as curious, interested, and engaged (Clayton-Smith & Laan, 2003; Gentile et al., 2010; Williams et al., 2010). The overall affect of students with Angelman syndrome is thus likely to be a cheerful personality, prone to over-excitement, with high motivation for the social aspects of learning.

Children with Angelman generally demonstrate strong motivation to secure adult attention and to elicit social responses from others (Adams, Horsler, Mount, & Oliver, 2010; Moss et al., 2010; Oliver et

al., 2007). Mount and colleagues (2011) observed that many children with Angelman syndrome engage in high levels of approach behaviours (smiling, laughing, reaching) in order to achieve and maintain eye contact and social response from both caregivers and unfamiliar adults. Children with Angelman are particularly skilled at eliciting laughter and smiling from social partners (Moss et al., 2010; Mount, Oliver, Berg, & Horsler, 2011). This high interest in social interaction can become problematic if it is too intense and frequent; many children with Angelman are described as “needy” or attention-seeking (Mount et al., 2011). Children who demonstrate an intense drive for social interaction can be taught to recognize and respond to consistent environmental cues regarding whether an adult is available or unavailable for social interaction; the children can learn to resist the impulse to approach even when the child’s interest in social interaction does not appear to abate (Heald, Allen, Villa, & Oliver, 2013). Educators and families should know that adult eye contact, laughter, and smiling have particularly high value as reinforcement for many children with Angelman syndrome; careful planning around the use of adult attention can facilitate classroom management (Heald et al., 2013; Mount et al., 2011; Williams et al., 2010). Although there is no published research on social interest with same-age peers, anecdotal parent reports consistently describe high interest in peers on par with the heightened interest in adults. Peer models and cooperative work groups may thus be particularly motivating for some students with Angelman.

Certain aggressive behaviours, such as grabbing, are commonly observed in students with Angelman (Strachan et al., 2009; Summers, Allison, Lynch, & Sandier, 1995). These aggressive acts are frequently part of a strategy of escalating social approach behaviours to secure and maintain attention from others; if attempts to achieve social interaction with non-aggressive means such as eye gaze and vocalizations are unsuccessful, children with Angelman may escalate to touching, grabbing, and then pulling hair or scratching in an effort to secure the person’s attention (Allen et al., 2010; Radstaake, Didden, Oliver, Allen, & Curfs, 2012) . Over time, children may simply default to the more extreme aggressive behaviour (of scratching or hair-pulling) unless the early, non-aggressive bids for social attention are consistently ratified and reinforced with a reciprocal social response. Parents and educators

can work together to provide contingent positive reinforcement for appropriate bids for social attention (Radstaake et al., 2013). In addition, educators and families can prioritize approaches to teach communication skills that emphasize using AAC to request social interaction. When children with Angelman were taught to use an AAC device to request social interaction, aggression was significantly reduced (Allen et al., 2010; Radstaake et al., 2012). Clayton-Smith (2001) found that, amongst 28 adults with Angelman, stronger communication skills directly related to less aggression; the researcher conjectured that aggression could be the result of an inability to get one's needs met.

Aggression is common in Angelman but appears to have specific antecedents and serve specific purposes in addition to communication. Arron et al. (2011) found that individuals with Angelman syndrome who were the most impulsive and overactive were also the most prone to aggression, suggesting that aggression may be as much about controlling impulses as communication. Some children with Angelman use aggression as a strategy to escape from a demand (such as a non-preferred activity) or to achieve access to tangibles (Heald et al., 2013). Prioritizing these communicative functions (such as teaching a child to use AAC to ask for a break, to request that an activity stop, or to say "no") may be an effective approach to addressing aggressive behaviours.

Strachan et al. (2009) found that, while some children used aggression to escape from unpreferred activities, others used smiles and laughter in an apparent attempt to distract adults from the unpreferred activity or to express a variety of emotional states and feelings, such as discomfort. Just as some adults laugh nervously when anxious, certain children with Angelman laugh uncontrollably in response to pain, anxiety, or stress (Horsler & Oliver, 2006). Families and educators can collaborate to identify the types of laughter that seem to indicate true enjoyment versus other emotional states.

Attention

Students with Angelman syndrome tend to be physically active, inquisitive, and engaged (Clayton-Smith & Laan, 2003; Williams et al., 2006). Excessive activity and hypermotoric behaviours are

common and require active management (Gentile et al., 2010; Williams et al., 2010). However, compared to children with similar levels of intellectual ability, children with Angelman syndrome have strong attention skills (Barry, Leitner, Clarke, & Einfeld, 2005), particularly when engaged in preferred activities, suggesting that the attention challenges inherent in Angelman are most likely a function of overall delayed development rather than specific to the syndrome.

Attention may be affected differently in certain subsets of students with Angelman syndrome. Summers and Impey (2011) found, in a sample of four school-aged students with Angelman, that all four had joint attention skills, including the student with autistic traits. Students with Angelman who do not demonstrate joint attention may benefit from evaluation for other conditions that can inhibit the development or performance of joint attention, such as cortical vision impairment (Roman et al., 2010) or central auditory processing disorder (Millet et al., 2012). Another subset of students with Angelman have significant global low muscle tone and do not achieve independent mobility. While data on the abilities of students with Angelman have not been reported in such a way that we can compare the association among gross motor, fine motor, communication, and cognition, parents of these students anecdotally report that many demonstrate greater attention to a variety of stimuli and often have strong fine motor skills and communication skills, suggesting these children are able to channel their energy into success with more stationary activities.

Students with Angelman syndrome tend to be highly motivated by social experiences and will closely attend to a social partner in engaging interactions; educators and families should strategize about how to use social attention as a reinforcer for engagement. Direct instruction, peer supports, peer partners, and cooperative learning groups may all be engineered to intentionally use social attention to maintain engagement (Heald et al., 2013; Mount et al., 2011; Williams et al., 2010).

Sensory dysfunction may play a strong role in the maintenance of attention for some students with Angelman syndrome (Walz & Baranek, 2006). The ceaseless need for movement described in these students (Williams et al., 2010) is consistent with sensory processing dysfunction, such as hyposensitivity

to vestibular and proprioceptive sensory input (Ayres, 1972). Ceaseless or purposeless movement is also common in students with dyspraxic deficits in motor planning (Mirenda, 2008). Students who demonstrate this intense frequent need for movement may benefit from investigation of underlying sensory needs or dyspraxia in order to help make these students more available for prolonged attention. Occupational therapists can support educators and families to strategically plan how to meet a student's sensory needs in ways that maximize the student's availability to attend to classroom learning. Common sensory integration strategies include building regular physical activity into the day, increasing proprioceptive input with weighted aids (such as weighted vests, pressure vests, or lap weights), and providing extra vestibular input with activities such as swinging and rocking (Walz & Baranek, 2006).

Students with Angelman are described as curious and engaged (Williams et al., 2006); their attention deficits are likely more a function of easy distractibility than lack of interest. In any learning experience, a student's short-term memory is constantly bombarded with incoming stimuli, far more than the student can consciously accommodate. Students with Angelman syndrome may find it particularly difficult to identify the most essential aspects of incoming stimuli that are worthy of their attention. Students with Angelman tend to be particularly drawn to items that are colourful, plastic, shiny, and crinkly (Williams et al., 2006). Educators and parents should be strategic about identifying and deploying materials with properties such as these that a student finds most appealing to draw the student's attention to the materials to which the student needs to attend.

Students with Angelman syndrome frequently demonstrate an interest in exploring materials in a hands-on manner (Gentile et al., 2010). This tendency may simply reflect their global developmental delay but can also be symptomatic of underlying visual impairment (Downing, 2008), a common issue in Angelman. Hypermotoric activities combined with impulsivity lead many students to grab classroom materials to hold and explore them with their hands and even their mouths (Gentile et al., 2010; Williams et al., 2006). Parents and educators can plan for this need for multi-sensory exploration by providing access to safe, tangible materials to explore during classroom activities. Students with Angelman

syndrome may need accommodations such as additional time to explore tangible materials; one strategy that may be effective is to permit the student with Angelman to explore a tangible instructional item in advance of when it will be shown to all students, so that his or her curiosity is satiated beforehand. Providing the student with her own adapted version of classroom materials (such as an adapted version of the book the teacher is reading in a shared reading lesson) may also be effective to meet this need for hands-on interaction while supporting the student to attend (Downing, 2008).

The hands-on interest in learning common in students with Angelman can be capitalized upon to support memory formation and learning. Mouse models of Angelman syndrome suggest that the cognitive deficits in the syndrome are largely the result of disrupted memory formation (Daily et al., 2011; Jiang et al., 2010). Mice with Angelman require more intense and more frequent experiences to develop and access long-term experience-dependent memory. Our long-term memory is tasked both with storing all of the information we learn, and with making that information available for future recall on demand (Brinkerhoff & Keefe, 2007). As we process incoming stimuli from each of our senses, neurons in our brain form complex spider web-like patterns of informational nodes throughout the brain, organizing the memory of these stimuli into schemes. These schemes include information about everything from the physical properties of an object, to its possible taste and smell, to the feelings we associate with the experience. The links within each scheme keep expanding as we add to the stored information associated with these stimuli. The multi-sensory learning experiences that students with Angelman syndrome seem to crave may support the creation of more complex, richer schemes of information. Enriched multi-sensory learning experiences may be essential for students with Angelman syndrome as a response to the deficits these students experience in forming these schemes and accessing the information as needed. Providing students with Angelman syndrome with multi-sensory learning materials that can be explored with touch, taste, vision, hearing, and movement support the development of more effective schemes on which to build future learning.

The more informational links a student has in each memory scheme, the easier it is for the student to access stored memories (Brinkerhoff & Keefe, 2007). Instructional approaches that incorporate multiple senses, such as providing both visual and tactile information in addition to auditory stimuli, will help the student use stimuli received from the senses of sight, sound, movement, and touch to form more robust schemes. Strategies that activate prior knowledge (such as reviewing photos about a museum field trip before discussing a related historical topic) are very helpful (Downing, 2008). Connecting to a student's prior learning helps build more complex schemes by linking new information to pre-existing schemes, rather than rote memorization tasks that occur in isolation and require the student to begin construction of a new scheme (Brinkerhoff & Keefe, 2007). Students with intellectual disabilities may need greater support activating prior knowledge and associating new learning with old, so teaching approaches should explicitly make these connections for the student (Downing, 2008).

Downing (2008) suggested that families and educators should supplement classroom instruction with tangible materials related to the theme of study: these materials might include a collection of seasonally appropriate items during discussions of the seasons, like leaves in fall or real snow in winter, or lifelike models of birch bark canoes and log cabins during a unit on pioneer society. Tangible materials can be included in remnant books or experience books for the student to continue to explore throughout the course of the unit or school year (Beukelman & Mirenda, 2012). Remnants are tangible objects that represent a personal experience; a movie ticket, photograph, wristband worn at a concert, art project, and school assignment are all possible remnants. Remnants have a high degree of iconicity, meaning they carry stronger associations with a specific event than more symbolic, intentionally more generic forms of communication, such as words or visual symbols (Ho, Weiss, Garrett, & Lloyd, 2005). While remnant books have historically been used to support conversation about personally meaningful topics, the concept can be adapted to support attention during classroom learning (Beukelman & Mirenda, 2012; Cumley & Swanson, 1999). Remnant books are a helpful way to build vocabulary comprehension by combining concrete objects with the visual symbols that represent them. Individuals with cognitive disabilities and

aphasia demonstrate increased initiation of communicative interactions and improve their attention to conversation when partners support the regular use of remnant books, particularly when the remnants have strong personal associations (Ho et al., 2005).

Video-based instruction can “anchor” otherwise abstract instruction by providing a narrative with a protagonist to whom students can relate, while introducing targeted instructional concepts in a style that students with developmental disabilities often find more accessible (Ayres & Langone, 2008). Video of a student’s own personal experiences reinforces memories of her own experiences; reviewing these videos can activate background knowledge and enhance attention. Handheld touchscreen tablet computers with built-in cameras and internet access make video modeling and video viewing much more efficient and accessible than in the past. Research is emerging about the effectiveness of iPad-based video modeling for everything from functional communication development to academic instruction (Hart & Whalon, 2012). Students with characteristics of autism (Mechling, 2005) and students with visual impairment are commonly attracted to video and electronic media; students with Angelman who demonstrate these characteristics may benefit from instructional approaches that utilize these tools (Micheletto et al., 2011; Peters et al., 2012). The appeal of video may be its intensity and movement: we pay more attention to stimuli that are louder, faster, or more colourful than competing stimuli, and movement attracts our attention (Brinkerhoff & Keefe, 2007).

Educators and families can capitalize on this attraction to digital media with the use of video modeling and computer-mediated instruction (Ayres & Langone, 2008). Video models can be created in multiple ways: traditional video modeling (where others act as models of the target behaviour), video self-modeling (where the student is filmed performing behaviours; then the video is edited to only show the student performing the best behaviours), and point-of-view modeling (where scenarios are filmed from the physical point-of-view of the target student so that students can explore features of their environment and their routines through a video context). Each of these models supports learning for students who struggle with imitation, with generalizing routines to unfamiliar contexts, or with following instructions

(Shukla-Mehta, Miller, & Callahan, 2010). Video modeling of social and communication skills results in quicker acquisition of targeted skills than live, in vivo, modeling (Charlop-Christy, Le, & Freeman, 2000), likely because of the easier access to more frequent repetition.

Students with auditory processing or visual processing impairment (such as many students with Angelman) may benefit from video-supported instructional approaches because they provide easy opportunity for repetition of the otherwise fleeting sounds of oral language and the visual referents to speech (Mechling, 2005; Thiemann & Goldstein, 2001). Video of aided language modeling may have potential as a format to increase the volume of symbol-supported communication modeling available to students; families and educators can videotape engaging conversations modeling the use of visual symbols, for the student to review at her leisure. Video modeling is associated with significant gains for students with autism in areas such as spontaneous communication, commenting, and requesting (Mechling, 2005). These video modeling strategies can be combined with other highly engaging activities, such as videotaping a communicative interaction between the student and her communication partner as they review the student's remnant book about a favourite past event. Digital remnant books could combine photos and video of past events in a multi-media format that provides prompts and contextual cues to support conversation.

Mobile tablet technologies facilitate access to interactive video instruction, such as video prompting (Mechling, 2005). Multiple iPad apps, for example, permit the educator to play a video demonstration, and then require the student to engage in some action correctly before more video is played. These applications can be used in social scripting, where an AAC user is prompted to activate her touchscreen to share a narrative by showing video of her own activities; parents or educators can record interactive elements for potential communication partners (such as prompts like "Guess what happened next?") as part of creating the social script. The video prompts can support students to complete steps in a sequence, maintain engagement in multi-step activities, and engage in more reciprocal interactions.

Health conditions that affect learning

Angelman syndrome is associated with specific health concerns that may affect attention and learning and often require management in the classroom. The most common of these health concerns is epilepsy, which impacts over 80% of individuals with Angelman (Pelc et al., 2008; Thibert et al., 2009) and can directly impact learning, behaviour, affect, and attention. Additionally, certain other health conditions associated with Angelman can cause an atypical expression of pain of which educators should be aware.

Epilepsy emerges in the first year of life for half of all children with Angelman (Thibert et al., 2013). About one-third of children experience their first seizure with a high fever, and the large majority of children with Angelman will have experienced multiple epileptic seizures by the time they reach school-age. The seizure disorder in Angelman syndrome is complex and mixed. Students may experience multiple versions of various seizure types. The common presentations of seizures include:

- Brief atonic head drops: the child's head or entire body will suddenly lose tone and drop;
- Brief myoclonic jerks, like a shock or startle, usually of the entire trunk;
- Absence staring spells with momentary loss of awareness;
- Atypical absence staring spells with longer periods of loss of awareness, often accompanied with unusual chewing or smacking movements of the mouth, enlarged pupils, and/or eyes deviated to the side;
- Myoclonic absence spells, where the student has reduced or absent awareness combined with a tremor or subtle jerking movements;
- Classic tonic-clonic convulsive seizures;
- Myoclonic status or cortical myoclonus, a prolonged episodic disabling tremor.

- Non-convulsive absence status, frequently characterized as reduced awareness, usually combined with regression in previously acquired skills or behaviours and/or marked change in personality and affect (Pelc et al., 2008; Thibert et al., 2009; Thibert et al., 2013).

Families can work with educators to develop an effective safety plan for the management of epilepsy in the classroom. Angelman syndrome is not associated with developmental regression (Thibert et al., 2013); students with Angelman syndrome continue to make significant progress in their learning and development across the lifespan (Peters et al., 2012). Developmental regression is a common symptom of poor seizure control and should be reported to families for medical follow-up. In addition, nonconvulsive status epilepticus is a form of seizure that affects as many as 90% of individuals with Angelman at some point over the lifespan, with episodes frequently recurrent (Thibert et al., 2013). Nonconvulsive status is caused by increased epileptic spikes of activity in the brain and can cause altered levels of awareness, regression in motor skills and development, and a marked change in personality. Educational teams should document and report to families whenever students with Angelman syndrome demonstrate a change in personality or affect (such as the appearance of lethargy or aggression), loss of skills, or reduced awareness.

The effects of the treatment of epilepsy can also manifest in the classroom. Seizure medications and treatments can cause lethargy, aggression, change in appetite, reduced attention, reduced muscle tone, and a paradoxical increase in seizures (Pelc et al., 2009; Thibert et al., 2009). Treatment of epilepsy involves a balancing act, where the possible benefits of aggressive seizure treatment (such as increased cognitive functioning, improved quality of life, and improved safety) are weighed against the potential side effects of various medications and their possible drug interactions. Treating seizures reduces the risk of long-term cognitive damage and sudden unexpected death in epilepsy (Thibert et al., 2013). At least half of all students with Angelman syndrome require multiple daily medications to manage their epilepsy. Educators can help families document the side effects and efficacy of seizure medications by noting changes that could be related to medications and reporting this information to families. Well-documented

information can help medical professionals make effective changes in the student's seizure treatment plan.

Some children with Angelman syndrome will present in the classroom with specific dietary needs caused by seizure treatment. Various mainstream medical dietary therapies for epilepsy include the ketogenic diet (Thiele, 2003), the modified Atkins diet, and the Low Glycemic Index Treatment (Thibert et al., 2013). Dietary remedies for epilepsy attempt to replicate the long-observed positive effects that fasting has on preventing seizures; these physician-prescribed diets are generally high in fat, reduced in protein, and very restricted in carbohydrates (Thibert et al., 2012). These dietary treatments are often more effective than conventional medication and can positively influence attention and sleep, but require strict maintenance with their respective dietary protocols to continue the positive effects on seizure prevention. Educational teams can learn how to support students on these diets by understanding the importance of the dietary restrictions for seizure control, and alerting families before classroom treat days so that the student on the restricted diet can bring in a special treat from home.

Epilepsy in Angelman syndrome is associated with the sleep disorder that is common in Angelman. Children with Angelman frequently have a severe sleep disorder and often have difficulty falling asleep, staying asleep, and sleeping past the early hours of the morning (Bruni et al., 2004). Approximately one in four children with Angelman sleeps less than 6 hours per night (Walz, Beebe, & Byars, 2005). There is debate about whether or not children with Angelman syndrome somehow require less sleep than their same-aged peers (Bruni et al., 2004; Didden, Korzilius, Smits, Curfs, & Dykens, 2004; Thibert et al., 2013; Williams & Battaglia, 2013), but there is no research demonstrating that students with Angelman syndrome perform better with less total sleep than is considered necessary for the rest of the population. Sleep deprivation can worsen epileptic seizures, while epileptic activity disrupts sleep. If a student with Angelman syndrome demonstrates significant changes in her sleep patterns and/or cognitive regression, medical professionals should investigate for prolonged or persistent seizure activity as a possible cause (Thibert et al., 2013). Sleep deprivation results in deficits in attention, impulse control,

learning, memory, emotional reactivity, and response times to various stimuli (McCoy & Strecker, 2011). Children with Angelman syndrome who demonstrate both the symptoms of a sleep disorder and the symptoms of sleep deprivation should be evaluated by a medical professional for support with management of sleep. The sleep disorder can be effectively treated with intensive behavioural interventions (Allen, Kuhn, DeHaai, & Wallace, 2013).

Gastroesophageal reflux and constipation are both commonly experienced by children with Angelman syndrome (Williams et al., 2010). As well, epilepsy and its treatment are both associated with an increased risk of headache and migraine (Bauer et al., 2013). The student with Angelman syndrome is thus at increased risk of experiencing and expressing pain in the classroom. The expression of both pain and anxiety may be unusual in students with Angelman syndrome and may include aggression or laughter (Oliver et al., 2007; Thibert et al., 2013). Self-injury in students with Angelman syndrome is rare but is usually associated with the experience of pain (Oliver et al., 2007); self-injury can be a strategic response to the experience of pain by attempting to substitute a lesser pain in the presence of a more severe one (Kugelmann, 1997). The dyspraxia and sensory disintegration experienced by many students with Angelman syndrome can disrupt pain signals and cause the student to experience agitation without expressing obvious signs of pain. Children with Angelman are reported to have an unusually high tolerance for pain and may not indicate pain until it's quite severe (Williams et al., 2006). Educators should be alert for a change in behaviour or personality that could indicate pain and report these instances to parents.

Conclusion

The profile of learning characteristics of students with Angelman syndrome suggests these students approach learning with curiosity, a positive attitude, and a strong drive to engage socially with others (Clayton-Smith & Laan, 2003; Williams et al., 2010). While few students with Angelman syndrome make effective use of augmented or alternative communication strategies (Alvares & Downing,

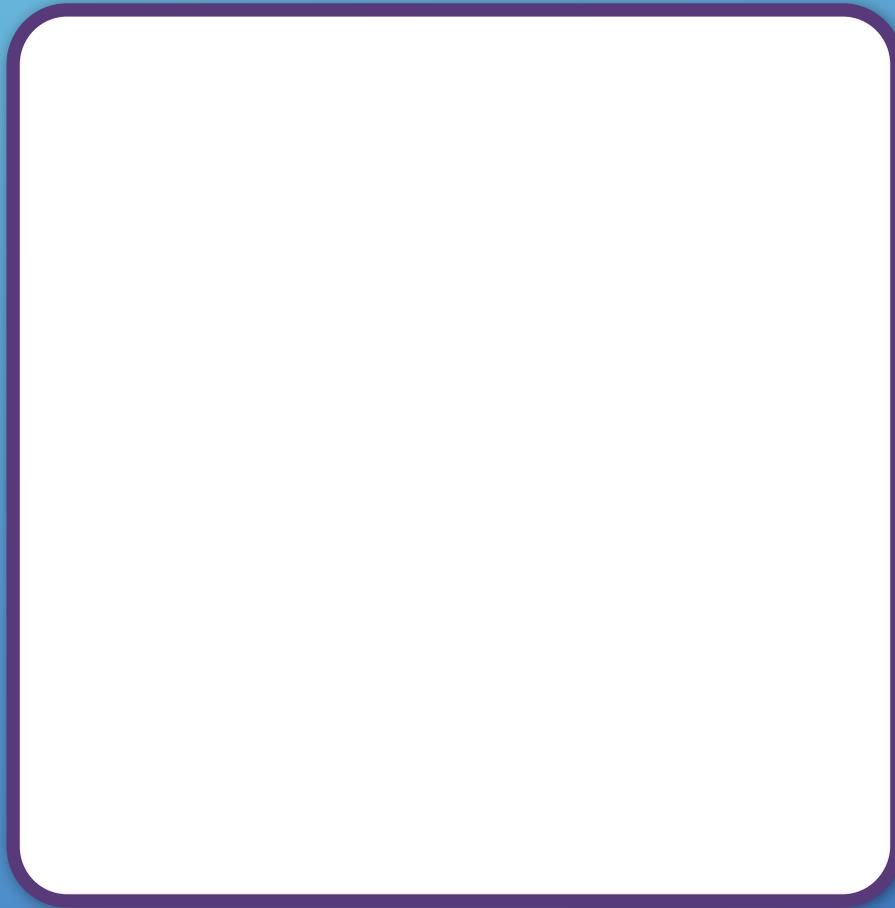
1998; Calculator, 2002), aided language input (Beukelman & Mirenda, 2012) and the opportunity to participate in rich, frequent communicative exchanges is essential in order to foster autonomous communication. Students with Angelman syndrome may require explicit support to build and retrieve memories (Daily et al., 2011; Jiang et al., 2010); these students likely benefit from multi-sensory learning experiences with a strong social element. Additional prompts to support communication and the development of memories may include the use of remnant books (Beukelman & Mirenda, 2012) and other tangible, hands-on educational materials. Students with Angelman syndrome make slow but significant growth in learning throughout their school-age years (Peters et al., 2012); dynamic or portfolio assessment approaches may be particularly appropriate for compensating for performance deficits and capturing this incremental growth (Kearns et al., 2006).

Most students have a discrepant profile of physical abilities. Many children with Angelman are physically active and seek movement and hands-on exploration. In contrast, students with Angelman may be at heightened risk for problems with sensory integration (Beckung & Kyllerman, 2005; Walz & Baranek, 2006), visual and auditory processing dysfunction, and/or dyspraxia of motor planning (Penner et al., 1993), all of which conditions may benefit from referral and support from outside experts. Most students with Angelman experience epilepsy, which may require monitoring and management in the classroom (Thibert et al., 2013).

The available research on Angelman syndrome supports the presumption of competence, the presumption that the student can learn communication and literacy skills if provided literacy-rich opportunities and quality instruction. For example, Calculator (2013) found that, despite minimal exposure to the most effective approaches for teaching use of AAC, students with Angelman syndrome were learning to use these devices to meet their communication needs. Donnellan (1984) described the presumption of competence as the “least dangerous assumption,” suggesting that, in the absence of conclusive proof about a student’s potential for learning, presuming competence has less dangerous consequences for the student than presuming incompetence and denying a student the opportunity for

instruction. The presumption of competence ensures that educators and families provide the language-rich, text-rich stimulation that all students need to maximize their literacy and communication potential. Mirenda (2008) suggests that the handful of individuals with autism who were believed to have profound intellectual impairment - who have nonetheless been able to demonstrate strong intelligence through augmentative and alternative communication systems - should create “cognitive dissonance” for educators about what we think we can know about the potential learning growth of all students. The least dangerous assumption is that every student with Angelman syndrome is motivated to communicate and learn but may need carefully individualized supports to access rich learning experiences.

Chapter 3: Angelman Syndrome for Educators



Angelman Syndrome

for Educators

By Erin Sheldon, M. Ed.

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Introduction

Students with Angelman syndrome are characterized as cheerful, inquisitive, hands-on learners in the classroom. The disabilities associated with Angelman syndrome present educators with specific dilemmas in how to support these students to actively participate and to demonstrate their learning. The research literature on Angelman syndrome details the developmental disabilities seen in students with the syndrome but provides minimal direction to support school teams in planning and implementing individual education programs to meet these student's needs.

This booklet was created for school teams to assist with planning for students with Angelman syndrome. General educators will learn what differences they might expect in a student with Angelman and how these differences might be accommodated in an inclusive classroom. Special educators will find information on the nature of the learning characteristics of these students so that instructional approaches can meet their specific learning needs. Allied professionals such as speech language pathologists, physical therapists, and occupational therapists will see how essential they are to supporting the student with Angelman to access instruction and demonstrate learning. Parents will find information to help them participate in discussions about their student's individual learning characteristics and planning their student's placement and program.

This booklet is designed to:

- explain the general characteristics we expect to see in students with Angelman syndrome
- demonstrate the complexity of the spectrum of abilities and disabilities observed in the syndrome
- assist IEP teams as they plan specific accommodations and set learning goals
- support school teams with ideas and resources as they problem-solve common challenges
- answer common questions that educators and families have about specific topics.

Each section of this booklet explains one of the learning characteristics that educators might expect to observe in the student with Angelman in the areas of:

- **communication, including strategies for aided language input**
- **cognition**
- **physical abilities**
- **senses**
- **attention**
- **affect**
- **associated health concerns**

Additional sections explore specific topics, including:

- **autism in Angelman syndrome**
- **cortical vision impairment**
- **literacy instruction**

Each of these sections was summarized from a comprehensive review of the published literature. The complete literature review is available by contacting the author.

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General Learning Characteristics

Angelman syndrome is a neurological disorder that results from disruption to a specific gene on the 15th chromosome. The syndrome is associated with significant disabilities that affect how these students learn and how they can demonstrate their knowledge. Students with Angelman syndrome are learners who require careful planning and accommodation in the classroom.

Communication

The most significant learning difference observed in nearly every student with Angelman syndrome is a profound expressive language disability. Expressive communication is disproportionately impaired in students with Angelman Syndrome compared to overall comprehension, cognition, and global development.

Students with Angelman syndrome generally demonstrate strong motivation to communicate and interact with peers and adults. What a student with Angelman syndrome can express should not be mistaken for what that student can understand. The nature of the communication disorder in Angelman is fundamentally one of a struggle to access and express language. Students with Angelman syndrome need intense support to learn how to communicate effectively to meet their social and academic needs. These students require instructional approaches that scaffold access to tools for

symbolic communication while respecting and refining non-symbolic communication, such as their use of body language and natural gestures to express meaning.

Students with Angelman syndrome have complex communication needs. These students can benefit from aided language input, an approach to developing communication skills based on what we know about how all children acquire language. Typically developing children acquire language through constant exposure to spoken language in meaningful contexts. Aided language input recognizes that children who cannot produce oral speech still require this deep immersion in a model of language they can access. The aided language approach teaches communication partners to use symbol displays to immerse the student in a model of symbolic language that is accessible to students who do not have speech.

Cognition

Angelman syndrome is associated with significant intellectual disability. The cognitive disabilities documented in Angelman syndrome may be specifically related to challenges in memory formation. Understanding these cognitive disabilities is complicated by the profound communication impairment and by physical disabilities that disrupt muscle tone and impair motor control and motor planning. Educators and speech language pathologists should emphasize access to augmentative and alternative communication (AAC) so that these students can express their understanding.

Angelman syndrome is not associated with cognitive regression; these students continue to make progress in their learning throughout their school career. Regression or lack of progress should not be attributed to the cognitive aspects of the syndrome.

Global dyspraxia (difficulty coordinating motor movements) is common in students with Angelman and often makes it difficult for these students to perform on command. Students with dyspraxia may best demonstrate their knowledge in natural contexts.



Literacy development is possible in students with Angelman syndrome. Most students with Angelman have emergent literacy skills that can be fostered with comprehensive literacy instruction in print-rich classrooms with daily opportunities to read, write, and communicate about personally meaningful topics.

Physical abilities

Angelman syndrome can affect physical abilities in multiple ways, impairing both gross and fine motor skills and often causing ataxia, dyspraxia, a balance disorder, and disrupted muscle tone. Students with Angelman should be assessed by an occupational therapist to ensure they have optimal postural support to engage in learning activities. For some students, these physical impairments may prevent independent walking. Tremors should be investigated as a form of epilepsy that may benefit from medical treatment.

Senses

Sensory impairment (such as vision or hearing loss) is not commonly reported in Angelman syndrome, but most of these students require support to integrate information received through their senses. All students with Angelman syndrome should be assessed for sensory integration dysfunction so that appropriate strategies can be implemented to ensure the student is available for learning.

Students with Angelman syndrome have many risk factors for cortical vision impairment and central auditory processing disorders. Educators should be aware of the symptoms of visual and auditory processing disorders so that appropriate referrals and accommodations can be made if necessary.

Affect

Students with Angelman syndrome are often described with a cheerful, curious affect. As a group, they demonstrate strong behavioural flexibility and can be prone to over-excitement. Educators can capitalize on this characteristic through social learning activities such as peer-mediated learning, cooperative learning

approaches, and opportunities to learn in print-rich regular classrooms alongside typically developing peers.

Challenging behaviours are common and are often related to unmet communication needs. Functional behaviour assessment can reveal the underlying purpose behind the student's behaviour so that a more appropriate but equally effective behaviour can be substituted. Individuals who use the most effective communication systems have the least challenging behaviours (Clayton-Smith, 2001); it is essential that intensive communication interventions are in place to ensure that students with Angelman syndrome can express what they need to say when they need to say it.

Attention

Students with Angelman syndrome have attention skills that are consistent with their overall development, particularly when engaged in preferred activities. The attention challenges in Angelman may be an issue of easy distractibility; many of these students struggle to filter out competing stimuli. Video-based instruction and video-modelling provides both more stimuli and more opportunity for repetition; video-based approaches may help many of these students focus and maintain their attention in the face of competing stimuli. Many students benefit from hands-on learning activities such as the use of manipulatives, tangible objects, and materials with appealing sensory properties. Poor attention may reflect an underlying challenge with sensory integration, visual or auditory processing, or epilepsy management.

Additional Resources

[An Introduction to Angelman syndrome by ASSERT](#), the UK Angelman syndrome association

[Facts About Angelman Syndrome](#), 7th Ed., 2009, by the Angelman Syndrome Foundation

Communication

Students with Angelman syndrome have complex communication needs. The expressive communication disability in Angelman is more severe than the student's overall development, cognitive abilities, and receptive language skills. The communication disorder in Angelman syndrome is symptomatic of a global dyspraxia that prevents many students with Angelman from even imitating familiar gestures on command (Penner et al., 1993). The communication disorder in Angelman is thus partly a motor disorder.

Students with Angelman syndrome are motivated to communicate. The essential communication challenge for educators is that these students require enormous support to express themselves. What a student with Angelman syndrome can express should be presumed to be the floor, not the ceiling, of his or her comprehension. Students with Angelman syndrome cannot meet their daily communication needs without adaptive assistance for communication and writing. Support for communication development is the greatest need of students with Angelman syndrome.

Students with Angelman syndrome use a wide variety of expressive strategies in an attempt to meet their daily communication needs. These students develop strong preferences for whatever system is the most efficient to convey the messages the student is motivated to express. A minority of students with Angelman syndrome develop some oral language or manual sign language. The majority of students with Angelman syndrome use prelinguistic, non-symbolic communication strategies such as vocalizations, natural gestures (such as reaching and touching), and facial expressions.

Unaided communication relies solely on a person's body to produce a message (such as gestures or manual sign language). Unaided communication strategies can effectively express a concrete message, such as requesting or refusing a tangible object. However, in the absence of a formal unaided language (such as American Sign Language), unaided communication is

inadequate to express abstract thoughts, such as messages related to the past or future (Beukelman & Mirenda, 2012). Students with Angelman require aided communication support in order to meet their communication needs.

Students with Angelman syndrome often rely more on unaided communication than on aided communication with assistive technology. This reliance on unaided communication may reflect the instructional approaches used with many of these students. Calculator (2013) found that only 5% of the instructional approaches used with students with Angelman syndrome could be considered evidence-based as effective for teaching students to use and adopt aided, augmentative communication. Aided language modelling or stimulation is an evidence-based approach that fosters the adoption of AAC systems.



Aided communication relies on external tools (such as symbol displays, voice-output devices, or picture exchange) to generate or augment a message. Aided or augmentative communication tools include light-tech (paper-based) symbol displays and high-tech speech generating devices that electronically change the symbol display. Speech language pathologists trained in augmentative or alternative communication can help select the most appropriate aided communication tools to ensure the student can meet his or her daily communication needs.

Communication Strategies

Students with Angelman syndrome require intensive support to learn to use alternative and augmentative communication (AAC) systems to meet their daily communication needs.

Access to a comprehensive AAC system is essential for students with Angelman syndrome because, without assistive technology, these students cannot otherwise meet their daily communication needs. Assessment for an AAC system takes into account how the student will access the AAC system and how barriers to its use will be removed. AAC systems should provide access to enough language that the student can meet his or her communication needs, and so that communication

partners can use the device to augment their communication with the student.

Students with Angelman syndrome are motivated to communicate about personally meaningful topics. Educators can expand opportunities for communication by incorporating the students' personal experiences into the classroom. Remnant books or conversation books include tangible objects from a child's life (such as movie stubs, photographs, and found objects) paired with concise text to help spark conversation with peers and enhance sharing. Communication circles train peer tutors to model and support aided communication with the supervision of a supportive adult.

Beukelman and Mirenda (2012) proposed the **Participation Model** as a framework for speech language pathologists, educators, and families to understand a student's daily communication needs and to plan interventions.

The Participation Model first identifies the participation requirements of a peer without disabilities of the same chronological age. Next, it inventories the target student's current levels of participation compared to the peer. Participation is understood in degrees of independence. For example, a student may be able to read hard-copy books when provided with full physical assistance for each step of the task. But the same student might be able to select and navigate electronic books on the computer independently if an instructor has prepared the activity. The Participation Model suggests that planning should maximize independence.

Barriers to the student's greater participation are then examined. These barriers are understood as either access barriers or opportunity barriers. **Access barriers** are intrinsic to the student's disability, such as the student's inability to speak or the presence of fine motor disabilities that prevent directly touching a speech generating device or handling a hard-copy book. Access barriers can be problem-solved with assistive technology.

Opportunity barriers are distinct from access barriers because they are imposed by others as a

response to the student's disability. Opportunity barriers come in many forms:

- **Policy and practice barriers** include practices such as congregating all students with similar functional skills in the same classroom rather than educating all students in neighbourhood schools.
- **Skill and knowledge barriers** are those such as educators not being trained to use and integrate a student's assistive technology, or when educational teams lack experience including students with disabilities in regular classrooms.
- **Attitudinal barriers** are those such as educational teams reducing their expectations for the student with disabilities, or not considering inclusive placements due to beliefs about the nature of the student's disability.

When a student fails to make measurable progress in communication development, it is essential to accurately identify the nature of barriers to the student's communication development so these barriers can be dismantled. Educators and families can address opportunity barriers by: partnering to request additional training in approaches such as aided language modelling; insisting that planning time for educators be written into the IEP; pursuing peer-mediated supports such as Communication Circles; or supporting a family's request for student placement in a language-rich regular classroom.

Aided language input

Children acquire spoken language through constant immersion in their native language. Children hear about 1,250 words spoken per hour, or 6 million words per year. Infants and toddlers are immersed in oral language; over time, they observe, comprehend, imitate, and slowly begin to express words. Through this rich exposure to oral language, infants experiment with producing the sounds of the language they hear; over the course of many years, they develop their ability to participate in verbal exchanges.

Aided language input is an attempt to replicate this language acquisition experience for children who cannot speak. Children who cannot speak also require immersion in a language they can observe, comprehend, imitate, and eventually express.

Through aided language input, families and educators supplement their oral language with a visual symbolic language. Communication partners highlight symbols on a symbol-based communication display as they interact with the child verbally. Highlighting the symbol assists the child to “map” the verbal utterance to the visual symbol. By observing communication partners as they use visual symbols during motivating activities, the child begins to establish a schematic representation for how visual symbols can be combined and recombined to generate communicative messages ([Beukelman & Mirenda, 2012](#)).

Families and educators highlight these visual symbols on some form of communication display, either a light-tech symbol display (such as a laminated book of carefully arranged symbols) or a speech-generating device where visual symbols are matched with the sound of the word. There are multiple systems designed to provide aided language input. All of these systems share a commitment to problem-solving how children who cannot produce oral speech can access a visual symbolic language for communication.

Aided language input is provided in natural contexts in the course of ordinary conversation. It is often emphasized during activities that are particularly motivating to the child. As the child begins to indicate symbols herself, these first utterances are ascribed meaning, shaped, and reinforced, just as an infant’s first attempts to say “da-da” are shaped and reinforced to result in a first word like “daddy.”

Aided language input is an effective intervention to develop access to symbolic communication. Children who receive aided language input benefit from these visual supports for their receptive understanding of language; these children begin to associate visual symbols with the spoken sound and meaning of words. Children exposed to aided language input learn to expressively communicate using new symbols.

As a result of aided language interventions, children learn to combine symbols, to express more complex messages, and to increase their participation in communicative exchanges and in group activities ([Romski & Sevcik, 1996, 2005, 2010](#)). Developing language through aided language stimulation supports generalization because it occurs in natural contexts. The gains seen as a result of aided language input persist long-term.



Methods of aided language input

There are no prerequisites for aided language input. Just as infants begin the process of acquiring language at birth, so can every child who cannot access oral language benefit from the modelling of a visual language that she can access. Rather than waiting for the child to demonstrate readiness to use a symbolic communication system, adults use the symbol displays to supplement their own communication with the child. It should be expected that, just as typically developing children observe language long before they attempt to express it, many children will need a prolonged period of time to observe the modelled language before they begin to use it themselves. Multiple tools and approaches are available to provide aided language input. All of these systems are rooted in language acquisition theory. All of them share a commitment to immersing children who cannot speak in a symbolic language that they can observe, imitate, approximate, and finally express.

Pragmatically Organized Dynamic Display (PODD) books are the most commonly used light-tech aided language system for children with Angelman syndrome. PODD instructors teach communication partners to use carefully organized light-tech displays called PODD books. PODD books are a full language system organized in a book or binder format. PODD books are designed to meet the communication needs of both the child and her communication partners, at all times, in all settings.



PODD books organize symbolic language pragmatically. Symbols do not move around, but can instead always be accessed using the same navigational pathways so that communication becomes automatic and efficient. The PODD system relies on the communication partner to be a “smart partner” to scaffold the student to greater understanding and expression of language. PODD books generally start as light-tech books but can be programmed on dynamic display speech-generating devices.

Other forms of aided language input, such as The System for Augmenting Language, teach communication partners to model the use of dynamic display speech generating devices. Communication partners highlight symbols on the electronic display and model how to navigate the dynamic display. Dynamic display devices have the advantage of voice output; a recorded voice or text-to-speech software speaks the message out loud. Dynamic display systems are often available in more compact digital formats (such as the iPad) than light-tech systems. However, emergent communicators may struggle with the rapidly changing electronic screen and the lack of a “smart partner” to help them navigate back to a home page if unintentional movements change the symbol display. A speech language pathologist familiar with aided language input can help select the most appropriate system.



Cognition

Angelman syndrome is associated in the medical literature with significant intellectual disability, according to standardized psychological assessments designed for typically developing children. However, norm-referenced assessments presume a single, linear progression of development; this presumption fails to account for how students with significant disabilities may follow unusual developmental trajectories as they learn to strategize and compensate for the specific effects of their own disabilities. Alternative assessments that can separate motor skill performance tasks from cognitive tasks are often essential to capture what a student with Angelman syndrome understands.

Mouse models of Angelman syndrome suggest that the cognitive deficits of Angelman syndrome may be specific to storing and retrieving information from memory. Mice with Angelman syndrome demonstrate a disproportionate deficit in experience-dependent memory; they require substantially greater repetitions of an experience at greater levels of intensity before they demonstrate the ability to retrieve and apply information learned from these experiences. For example, mice with

Angelman syndrome approach familiar water maze experiments with patterns of exploration that suggest they have no memory of their previous experiences in the maze. If students with Angelman syndrome experience a similar memory deficit, then educators may find it helpful to focus on supports for memory formation.

Students with Angelman syndrome often crave multi-sensory learning experiences and intense stimulation. This attraction to sensory stimulation may be, in part, a strategy to support stronger memory formation. During a learning experience, as we process information from each of our senses, neurons in our brains form complex spider web-like patterns of informational nodes that form schematic representations of our experience. Each of these nodes store information associated with the experience: the touch, taste, smell, sound, sight, and emotional feelings. The more senses involved in a learning experience, the more links we form in these schemes; the more links, the easier it is to access the stored information. Multi-sensory learning experiences with a strong emotional component form stronger schemes.

Angelman is a complex neurological disorder, and multiple factors influence the ability of affected students to demonstrate their intelligence. Educators should understand the factors that may contribute to the presentation of intellectual disability so that the student's cognitive potential can be isolated and understood separately from these other factors.

Expressive communication in Angelman syndrome is affected disproportionately to overall development and cognitive ability. This profound communication impairment inherently affects the ability of these students to express what they understand. Cognitive assessments that rely on expressive communication are likely to underestimate the intellectual ability of students with Angelman. Strong support and

instruction in augmentative and alternative communication is essential to enable these students to express their learning.

Co-existing physical disabilities make it difficult for many students to perform motor tasks to demonstrate their comprehension. Abnormal muscle tone, tremors and unintentional movements, apraxia, and dyspraxia are all common in students with Angelman syndrome. Cognitive assessments that rely on instructions to perform motor activities are likely to underestimate the intelligence of students affected by these movement disorders. Physical and occupational therapists can help assess the presence of movement disorders so that appropriate accommodations can be made.

Approaches to support cognition

Students with Angelman syndrome make cognitive gains throughout their school careers. Educators can support cognitive development in students with Angelman syndrome by intentionally supporting memory development with the formation of robust schemes.

Educators can scaffold learning by ensuring that students are building on pre-existing schemes rather than constructing new schemes for each new experience. Building on these prior schemes requires careful attention to activating background knowledge before introducing new information. Activating background knowledge is most effective when it taps into multiple senses associated with the same scheme, such as using objects that students can touch and explore, visual supports (photos and symbols), and multi-media supports (video footage).

Supporting memory storage and retrieval includes frequent reviews of learned information. Repetition with variation ensures that students have multiple opportunities to engage information, or use a skill, with many different people across many different contexts. Multi-media formats such as video modelling are particularly helpful for students who attend best to repetition in video format.

Reading and writing tasks can be supplemented with related photos, tangible objects, and videos to tap into auditory, visual, and tactile memory. Written and oral instruction should be paired with visual supports. Remnant books or memory books can involve the family in recording personal experiences that can be activated as related background information.

Students with Angelman syndrome have a strong memory for social interactions. Learning experiences with a strong emotional or social component support the formation of more robust cognitive schemes.

Cognitive support technology helps students take

more responsibility for their own executive functioning to increase their independence. For example, students can learn to use a visual schedule to plan what activity is coming next and what materials they may need to prepare.

Many students with Angelman make incremental gains that are best measured over the long-term rather than the short-term. Portfolio assessment may be particularly appropriate as a form of measuring this progress, especially for students who struggle to perform their knowledge on demand. Portfolio assessment strategies gather multiple forms of evidence (often including performance data, video footage, samples of student work, and descriptive anecdotal reports of specific learning moments) to create a picture of the student's learning over time. The triangulation of these diverse forms of evidence serves as proof of the student's emerging comprehension. This evidence is collected in the course of ordinary classroom learning experiences, in the familiar and deeply contextualized environments where children are most likely to enact their learning.

One example of a validated portfolio assessment tool is the [The Bridge](#), an observation-based assessment tool that provides a framework to assess and plan a student's emergent literacy development. Educators partner with families to gather evidence of the student's early literacy behaviours in the areas of: Foundations of Reading, Foundations of Writing, Alphabet Knowledge, Phonological Awareness, and Literacy-Related Language. Artifacts of student work and anecdotal reports (such as, "Maggie touched the letter M when I asked her to write her name?") are recorded, dated, and filed under the appropriate area in the student's portfolio. Portfolio assessment allows progress to be measured longitudinally so that comprehension can be documented as it emerges. Portfolio assessment is most feasible when it is incorporated in regular instruction, such as providing students with daily opportunities for journal writing.

Physical Abilities

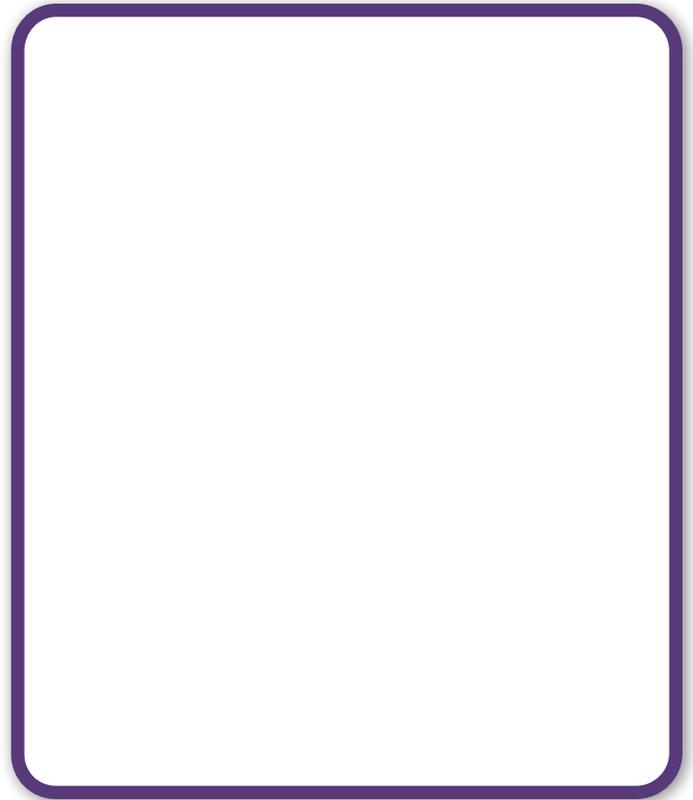
Angelman syndrome is usually associated with physical disabilities that require accommodation in the classroom. Understanding these physical disabilities is particularly important for supporting these students as learners, because many of these physical disabilities make it difficult for students to perform motor tasks to demonstrate their comprehension.

Most students with Angelman syndrome can walk independently by the time they reach school age. Students with Angelman generally walk with a distinctive broad-based gait with upraised arms and flexed wrists. This gait is a strategic, adaptive response to stabilize posture. To be independently mobile, these students must overcome ataxia, disrupted muscle tone, impaired motor planning, and a balance disorder. Many students with Angelman syndrome will always need to exert intense conscious effort to maintain stability while walking.

Students with Angelman syndrome demonstrate a “central dyscoordination resulting in difficulties in positioning the body and interacting with the environment,” with particular problems with sensory-motor integration ([Beckung & Kyllerman, 2005, p. 143](#)). [Penner et al.](#) (1993) described this dyscoordination as a widespread developmental dyspraxia. Dyspraxia is a complex motor planning disorder that inhibits the initiation and continuance of what would normally be volitional movements. Dyspraxia can be observed when students:

- are unable to imitate familiar motor movements;
- have high levels of purposeless and inefficient motor activity and excessive extraneous movement;
- struggle to coordinate movement, such as controlling head posture while visually scanning and attending;
- are unusually sensitive to environmental stimuli such as noise or light; or,
- are slow to respond to verbal instructions.

Physical and occupational therapists can help educators accommodate the effects of dyspraxia in the classroom.



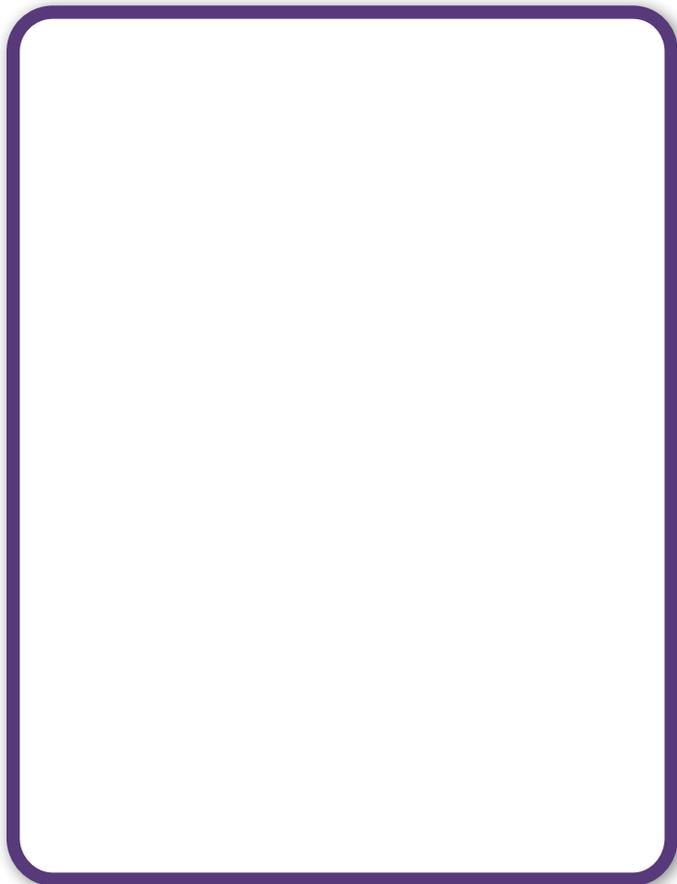
Some students with Angelman syndrome have more significant physical disabilities. These students usually have greater ataxia, global low muscle tone, and a more significant balance disorder. This greater physical disability is often not related to the student’s overall global development or cognitive growth. More severe physical disabilities create more challenges for these students to demonstrate their comprehension. Students with the most severe physical disabilities require strong postural support and pervasive support from assistive technology in order to access learning and demonstrate their knowledge.

Many students with Angelman present with a mixed muscle tone disorder with truncal hypotonia, high extensor tone, and hypertonic spastic limbs, resulting in a stiff gait and jerky arm movements. These students may require support from an occupational therapist to find the right supportive classroom seating to counter fatigue and position their bodies so they are available for learning.

Strategies: Physical Abilities

Controlled, fine motor movements tend to be more difficult for many children with Angelman than larger gross motor movements. Combined with sensory integration challenges such as tactile defensiveness, many of these students may resist hand-over-hand assistance or tasks that require grasping tools. Assistive technology support can help these students access classroom work.

Students with Angelman syndrome often use adapted chairs or wheelchairs and are known for broad, poorly targeted hand and arm movements. These students require adaptations and assistive technology to help them access instruction and demonstrate their learning.



[Kangas](#) (2008) emphasizes the importance of occupational therapists supporting students to maintain a posture in the classroom that helps students organize and coordinate their vision and hands. She describes this posture as an “alerting” task performance posture, an

active posture that prepares the body to activate and grade the movement of the hands, head, and trunk. This posture is achieved with the head and shoulders in front of the pelvis, feet on the floor and weight bearing (not necessarily symmetrical), knees flexed, with freedom to rotate the trunk and pelvis. Students in wheelchairs will need the most support to achieve this task performance posture.

Students who experience dyspraxia may benefit from approaches such as backward chaining, the system of least to most prompts, and naturalistic, predictable sequences; these strategies may prepare the student to initiate a motor task as part of a natural flow of activity rather than trying to initiate the task in response to a sudden performance demand.

Students who demonstrate symptoms of sensory dysfunction (such as tactile defensiveness) should be assessed by an occupational therapist. These students may require assistive technology for tasks like writing. “Alternative pencils” ([Hanser, 2010](#)) are tools such as light-tech eye-gaze boards, alphabet flip charts with partner-assisted scanning, touchscreen technology, or computer keyboards. These alternative pencils can remove the sensory and fine motor demands from the task of writing, freeing up the student to engage in the cognitive processes of writing, such as generating ideas and expressing them.

Some students with Angelman syndrome will never walk independently. Students with the most severe physical disabilities require support to achieve autonomy over their bodies. These students should not have to wait until they have sufficient muscle tone and motor control to independently get to where they want to go. Achieving autonomy over their bodies may involve alternatives to walking, such as learning to direct others where to move them or instruction in using motorized wheelchairs. Communication interventions should teach these children to request to be moved, to stay in one place, or to invite or call others to join them.

Senses

Sensory disabilities such as vision loss or hearing impairment are not commonly reported in Angelman syndrome. However, Angelman is primarily a neurological disorder. Sensory challenges observed in students with Angelman are primarily neurological in origin and affect how the brain processes and integrates information received through the senses. Educators should be aware of the symptoms of sensory processing disorders that can impact learning for students with Angelman.

Hearing

Central auditory processing disorder results in unusual or impaired auditory attention. These students may struggle to distinguish the sounds of oral speech from background noise in the classroom; they may, therefore, not respond appropriately or effectively to spoken language. Poor auditory processing may be suggested in students who: do not respond to the sound of their name, avoid eye contact, do not appear to attend to someone who is speaking to them, or appear over- or under-sensitive to sounds in the classroom. Students who are suspected to have hearing loss but have normal hearing screens should be investigated for central auditory processing disorder.

Vision

Visual impairments common in students with Angelman syndrome include strabismus and low acuity, both of which can be treated by an ophthalmologist. The characteristics of Angelman syndrome are associated with heightened risk for cortical vision impairment. Symptoms of cortical vision impairment include:

- unusual use of eye gaze, such as avoiding eye contact, appearing to “look past” people or objects, or appearing to prefer to watch things with peripheral vision;
- unusual attention to objects that are in motion versus stationary objects;
- poor depth perception, suggested when students appear to perceive a change in floor surface that is not

there, such as mistaking a shadow or line on the floor for a possible step or hole in the ground;

- unusual sensitivity or attention to light;
- unusual sensitivity to auditory stimuli, including the need for auditory cues to recognize visual stimuli, such as not showing recognition of a person until the person speaks.

Students who demonstrate these symptoms should be referred for functional vision assessment.

Sensory integration

Angelman syndrome can affect how all the senses receive and process information. Sensory integration or sensory processing refers to how the brain organizes sensation received through the body in order to function in the environment. Students with Angelman syndrome have a high rate of global sensory dysfunction, with a particularly high rate of hypo-responsiveness to sensory stimuli. This hypo-responsiveness suggests that most students need more frequent and more intense sensory input to adequately process and respond to information in their environment. A need for greater sensory input is indicated by behaviours such as mouthing or chewing, ceaseless movement, and attraction to materials that provide high levels of sensory input, such as water. Hypersensitivity to sensory stimuli can include symptoms such as sensitivity to heat, smells, or light; gagging or vomiting in response to specific stimuli; aversion to specific textures or touch; and pronounced startles to unexpected sounds. All of these symptoms can be addressed with a clear understanding of the student’s sensory integration needs.

Students with Angelman syndrome may be hyposensitive to the stimulation received through the vestibular and proprioceptive systems. Vestibular input is received through the middle ear and controls balance. Proprioceptive input is received through the joints in activities that are weight-bearing. Students who are not mobile need support to maximize the time they are weight-bearing to maximize proprioceptive feedback.

Strategies to address senses

Hearing

Central auditory processing disorders are difficult to assess in children with significant disabilities. However, strategies to support improved auditory processing may improve overall comprehension and can be simple to implement. A classroom amplification system improves the signal-to-noise ratio, allowing students to better isolate the teacher's voice and oral language from background noise. Voice amplification systems have value for all students and protect teachers from vocal strain.

Enhanced visual input can maximize comprehension; photos, audio, and videos are rich sources of visual input, permit frequent repetition, and provide context to help students derive word meaning from otherwise fleeting oral language. Verbal instructions can be “chunked” to improve comprehension and supplemented with visual cues and supports. Aided language input has a double benefit by providing visual cues to supplement oral language while simultaneously modelling the use of symbols for expressive communication.

Vision

The biggest effect of visual impairment on learning is that affected students are limited in their ability to learn incidentally from their environment. The majority of student learning is through visual cues; students with limited vision require direct intervention to compensate for the loss of these visual experiences. A teacher for students with visual impairment can assist with planning to:

- control lighting and prevent glare;
- remove visual clutter from the walls;
- adjust contrast and the use of colours, such as employing high-contrast visual symbols;
 - magnify or enlarge visual symbols or text; and
 - provide verbal or auditory cues in lieu of visual or physical cues such as body language.

Combined hearing and visual impairment

Some students with Angelman syndrome may experience both auditory processing and visual processing challenges. Specialists who support students who are deafblind can make appropriate recommendations. These students may require more support to use physical touch, more time to explore tangible objects, and greater response times. Students with combined auditory processing and visual processing challenges can present in ways that are very similar to students with autism.

Sensory integration

An occupational therapist can help determine the extent to which a student is over- or under-sensitive to various stimuli. Some strategies that benefit students with sensory integration challenges include:

- choices for seating, such as chairs with arms, rocking chairs, ball chairs, beanbags, large pillows, and work stations where children can stand rather than sit;
- break stations where a student can sit in dim lighting and curl up amongst pillows or in a small enclosed space to take a break from sensory input;
- dimmer switches on lights to reduce visual input;
- headphones for students to block out auditory input;
- tactile fidget toys such as stress balls, available at all times but particularly during periods of waiting;
- heavy work (such as carrying objects or wheelbarrow walking), weighted lap blankets, weight-bearing, the use of standers for students who need support to be weight-bearing, and therabands on chair legs to have something to kick, all of which provide proprioceptive input;
- options for oral motor stimulation, such as the use of straws, snacks with hard and soft textures, and chewing gum;
- swinging, rocking, and movement to provide vestibular input.

Attention

Students with Angelman syndrome generally have stronger attention skills than students without Angelman who present with the same level of cognitive ability. They are usually physically active and inquisitive. They are often prone to excess activity and hypermotoric behaviours that require management in the classroom.

Students with Angelman syndrome often demonstrate the greatest attention when engaged in personally meaningful and enjoyable activities. Tying instruction to the student's personal experiences builds more robust cognitive schemes, facilitates memory formation, and helps maintain attention. Visual supports such as photos, symbols, and video can help students make connections between their own lived experience and the topic of instruction.

Students with Angelman syndrome can usually engage in joint attention, a shared focus on both an object and another individual. Students who do not demonstrate joint attention may benefit from assessment of their visual or auditory processing. Students with unmet visual or auditory processing needs may focus on just one sense at a time (hearing *or* vision, not both) or may make unusual use of their peripheral vision, such as appearing to look away while listening or not looking at materials they are simultaneously touching.

Many students demonstrate a hands-on approach to learning, such as a need to touch and taste classroom materials. This need to touch may reflect global developmental delay but is also common in students with poor visual processing or low vision, and in students with unmet sensory integration needs.

Assisting students to meet their sensory integration needs can help them become more available for instruction. Many students are hyposensitive to sensory input; they require more intense vestibular and proprioceptive input to organize their bodies. Vestibular input provides information about our bodies in space as we move and maintain our balance. Proprioceptive input

is received through the joints as we engage in weight-bearing activities or receive deep pressure. Both these senses provide stimulation that is important for organizing and maintaining attention.

Some students with Angelman syndrome demonstrate an intense need for physical movement. These students tend to be constantly moving, may frequently invade the space of others, may demonstrate a need to touch everything, often demonstrate poor inhibition or impulse control, and may become fixated on visual stimulation (such as video). These characteristics are common in students with dyspraxia. This ceaseless need for movement should be problem-solved so that the student's underlying needs can be met, and the student can become available for learning.

Dyspraxia is common in Angelman syndrome and frequently accompanies sensory processing disorders. Dyspraxia is a movement planning disorder. It creates problems with: generating new ideas for a motor action, sequencing the steps to perform a motor action, and executing the motor action. Tasks that appear as simple as coordinating the head and eyes to work together to visually attend can be challenging for students who have dyspraxia. Occupational therapists can support the school team to understand the dyspraxic behaviours and help meet the student's underlying needs.

Attention challenges in students with Angelman syndrome may be related to easy distractibility. Our short-term memories are constantly bombarded with incoming stimuli. Attention is a function of knowing what external stimuli to attend to while filtering out the extraneous input. Many students with Angelman syndrome struggle to screen out excess sensory input. It may be helpful to observe what the student is paying attention to and then examine what properties or attributes attract the student's attention. These same properties can then be deployed strategically to either attract and maintain the student's attention, or avoided in order to reduce potential distractors.

Strategies to support attention

Students who demonstrate a ceaseless need for movement should be assessed for an underlying sensory processing disorder. Many of these students are hyposensitive to vestibular and proprioceptive input. Greater vestibular input can be provided in the classroom with rocking chairs, and in the gym or on the playground with movement activities such as jumping, rocking, and swinging (including swinging side-by-side as well as front-to-back). Proprioceptive input can be enhanced with pressure vests, weighted aids (such as weighted lap blankets or vests), and by performing heavy work, such as carrying weighted objects or wheelbarrow walking. An occupational therapist can assess the student's sensory needs and suggest accommodations.

Ceaseless movement is a common symptom of the dyspraxia that is common in students with Angelman syndrome. These students benefit from explicit supports to imagine and then plan their physical actions. Educators can use prompts such as “what should we do next?”, “how can we do this?”, and “should we do this, or that?” Sequenced photos, visual schedules, video modelling, or visual symbols can all help these students plan and implement the steps in an activity. For some students, physical touch on the body part that needs to move will help the student organize the related movement, such as touching the ankle before putting on shoes, or touching the shoulder before writing.

Attention can be supported by reducing distractions. Students who are sensitive to sound may be willing to wear noise-cancelling headphones. Minimal posters and dim lighting reduce excess visual stimuli. Some students benefit from sturdy partitions on their desks or creative desk and table arrangements that help block excess stimuli during activities that require the most concentration.

Our attention tends to be naturally drawn to movement and novelty. Some students with Angelman syndrome may be particularly drawn to highly-visually stimulating media such as video. Video-mediated

instruction and video modelling may be particularly helpful for these students.

All students benefit from instruction that is directly relevant to their own lived experience. Students with Angelman syndrome may benefit from supports such as remnant books. Remnant books collect tangible objects the student encounters and collects these objects in a book or binder, paired with concise supporting text. These books support memory recall for the student while providing educators with information about the student's background knowledge.

Finally, positioning and seating in the classroom directly impact attention and the ability to organize the body to plan for work tasks. Students are best positioned to attend and organize their movements when they are in an alerting task performance posture, similar to the posture we adopt as we are about to rise out of a chair. Students with low truncal tone who are reclined in strollers or wheelchairs will require seating that supports weight-bearing through the pelvis to adopt the postures that support attention and organize voluntary movement.

Some students will better attend to materials that have enticing sensory properties, such as crinkly textures. At times, an educator may elicit stronger attention by enhancing learning materials with enticing sensory properties. Other times, the educator may consider whether or not these properties are distractors. For example, a laminated display of symbols may be too enticing because of the sensory properties of its shiny texture; the student can't also attend to the symbols on the page. A matte laminate or durable rip-proof paper can replace the distracting stimuli of the laminate. Some students may attend better to visual symbols with tear-off Velcro backing; other students find the Velcro itself too enticing. The sensory properties of materials can be planned to ensure that they contribute, rather than detract, from the maintenance of attention.

Affect

Students with Angelman syndrome are generally described as cheerful, prone to over-excitement, and highly motivated by the social aspects of learning. Many of these students are exuberant and socially gregarious. Students with Angelman syndrome often deploy smiles and laughter as a strategy to attract and maintain attention from others. This strategy is effective; even when, for research purposes, adults in the environment are firmly instructed not to smile and laugh back, these adults report that it was nearly impossible not to respond to the smiling overtures.

Many students with Angelman syndrome demonstrate an intense drive to maintain eye contact with social partners. Eye contact, laughter, and smiling have particularly high value as positive reinforcement for many children with Angelman. Educators of some students with Angelman syndrome may want to strategically plan the use of adult attention and eye contact to facilitate classroom management.

The combination of a strong desire to connect socially with others and the lack of access to oral language may be the root of some aggressive behaviours that are observed in Angelman syndrome. Behaviours such as grabbing are common. These behaviours are often observed as part of a strategy of escalating social approach behaviours; when attempts to achieve social interaction or eye contact through methods such as eye gaze and vocalizations are unsuccessful, students with Angelman may escalate to behaviours that are harder to ignore, such as touching, grabbing, pulling hair, or scratching. Over time, students may simply default to these more aggressive behaviours unless parents and educators work together to consistently acknowledge and reinforce the socially appropriate bids for attention, such as vocalizing and eye gaze.

Students with Angelman syndrome commonly use challenging behaviours to either request something that is desired (such as social contact or a tangible object) or escape from a non-desired context. Some students use

aggressive behaviours to bring a halt to whatever demands are being placed on them. It is important to analyze these challenging behaviours from a functional perspective so that educators understand what the student is trying to achieve with the behaviour. If the student is using a challenging behaviour to say “no,” then the student needs a more specific, more appropriate, and more effective way to express “no” and have that refusal be respected. For example, the student may need to be re-directed to learn that they can say “no” on their communication system.

Some students with Angelman syndrome have intense challenging behaviours and require support from a functional behaviour therapist. Challenging behaviours are directly related to both the communication needs and the impulse control of these students.

Students with Angelman syndrome, in general, demonstrate unusually strong behavioural flexibility (such as cooperative responses to unexpected changes in the environment, personnel, or routine) compared to students with other forms of intellectual disability.

The cheerful affect in Angelman syndrome should not be presumed to mean the student is always happy. Some children use smiles and laughter as a strategy to distract adults from making a demand on them. Many students laugh to express a range of emotional states, including shyness, anxiety, discomfort, and pain. Many students with Angelman have a limited repertoire of communication skills and struggle to organize planned motor responses; smiles and laughter become the default expression of many complex emotional states.

Educators and families should observe changes in affect or personality with concern. Aggression (particularly self-injury) can indicate the student is feeling pain or feels unsafe in specific contexts. A change in personality, sudden aggression or anxiety, or a sudden loss of impulse control can also be a symptom of a form of epilepsy that is common in Angelman.

Strategies: Affect

Students with Angelman syndrome frequently crave high levels of social contact. This need can often be met by infusing social interaction into classroom learning with strategies such as peer tutoring and cooperative learning. Typical classrooms provide strong modelling of appropriate social interactions. Classmates can be taught to acknowledge the student's social bids and prompt use of the student's communication system to maintain social interaction.

Students with Angelman syndrome require strong support to meet their daily communication needs. In the absence of a comprehensive communication system, these students are left with only their smiles, laughter, and the strength of their hands and bodies to try to initiate social interaction and communicate what is important to them. When students with Angelman syndrome are taught to use communication devices to request social interaction, aggressive behaviours are significantly reduced. Strong communication skills are directly correlated with less aggression in these students.



Students with Angelman syndrome who are the most impulsive and overactive are also the most prone to aggression. These students benefit from careful attention to the root of their over-activity and impulsivity. These students may be affected by the sleep disorder or seizure disorder that is common in

Angelman. Seizure treatments themselves may be interfering with the student's impulse control. Physician support to manage the sleep and epilepsy may be helpful. Overactivity is a common symptom of unmet sensory integration needs; an occupational therapist may be able to design interventions to help the student meet his or her sensory needs so that the student is more available for learning.

Students with Angelman syndrome strategically engage in challenging behaviours; these behaviours serve a functional purpose. Challenging behaviours may be, for example, inadvertently increasing the amount of adult attention. How adults respond to challenging behaviour may be precisely what is reinforcing the behaviour.

For some students, challenging behaviours can be an expression of autonomy, of the student seeking more control over his or her learning environment.

Functional behaviour assessment uncovers the purpose behind the behaviour by carefully analyzing what was happening before, during, and after the challenging behaviour occurred. These assessments are a problem-solving approach to examine why behaviours occur, so that school teams can create interventions that meet the student's needs while substituting a more appropriate replacement behaviour.

[Beukelman and Mirenda](#) (2012) caution that providing the student with access to a communication system to substitute for the challenging behaviour only works if the communication system is more effective than the challenging behaviour at achieving the purpose the student was pursuing. Communication interventions should prioritize the communicative functions that the student is trying to achieve with challenging behaviour. These communicative functions might include requesting a break, requesting for an activity or demand on the student to stop, or saying "no."

Associated Health Concerns

Angelman syndrome is associated with specific health concerns that can impact learning in the classroom. Educators can partner with families to ensure students are healthy, safe, and available for learning.

Most students with Angelman syndrome have epilepsy. Some types of seizures that are common in Angelman are difficult to recognize and are rarely seen in the general paediatric epilepsy population; educators and families need to be aware of these seizure types to ensure proper management. Subtle seizure types that might occur in the classroom include:

- atonic head drops (sudden loss of tone in the neck or body so the student's head or body suddenly drops);
- sudden myoclonic jerks, like the student has just been shocked or startled;
- myoclonic tremors, a tremor or shaking of the body that may frighten or distress the student and may reduce her level of consciousness; and
- absence spells, staring or chewing during periods of very reduced or absent consciousness.

Non-convulsive status is one of the most challenging forms of epilepsy reported in Angelman syndrome; it is accompanied by changes that are easy to presume are “behaviours.” Non-convulsive status is low-level constant seizure activity that bombards the student's brain with too much electricity to function at his or her normal level, but not enough electricity to cause visible convulsions or other classic epilepsy symptoms.

Non-convulsive status should be suspected whenever a student with Angelman syndrome demonstrates regression in skills or cognition, or has a shift in personality or affect (such as suddenly becoming lethargic, manic, or aggressive). Disrupted sleep and an increased level of more visible seizures are both common when a student is in non-convulsive status. Non-convulsive status can emerge in waves or episodes that occur in between periods of normal functioning.

This form of epileptic status is not a medical emergency, but it heightens the risk for more emergent seizures and usually requires medical management to help students return to their baseline.

Epilepsy is usually managed with medications. These medications may cause side effects such as lethargy, poor concentration, or aggression. Educators can help document changes in a student's learning or affect that can help families and physicians find the best possible treatment. Low-carbohydrate, high-fat dietary remedies for epilepsy have the lowest rate of side effects and are often very effective.

Many children with Angelman have a significant sleep disorder. Symptoms of sleep deprivation include poor attention, poor impulse control, difficulty with memory recall, and slower response times. Families can partner with educators to document the impact of the sleep disorder on the student's learning and functioning. Medical management of poor sleep may be considered.

Students with Angelman syndrome have an increased risk of experiencing pain, combined with a communication disorder that restricts their ability to express that pain. Aggression and self-injury are both common expressions of pain in students with Angelman. Students with Angelman syndrome have risk factors for pain from gastroesophageal reflux, constipation, and headache. Some students express pain or anxiety with intense episodes of laughter.

Students with Angelman syndrome are at increased risk of obesity and loss of independent mobility as they age. Some students with Angelman develop eating disorders such as excessive appetite and food obsessions; these eating disorders can be as severe as those found in students with Prader-Willi syndrome. Parents and educators should exercise caution using food as a motivator to influence behaviour or motor skill development.

Strategies: Health Concerns

Educators can support the health care needs of students with Angelman syndrome primarily through awareness of the risk factors these students face. Educators can alert parents to the signs and symptoms they observe. Students with Angelman syndrome spend a substantial portion of their day in the classroom; documentation collected at school can help families and physicians make treatment decisions.



Each student with epilepsy has his or her own risk factors to experience seizures. Each student can have a unique presentation of seizures. Educators can learn about and help document the individual student's epilepsy profile. Common seizure triggers include illness, fever, infection, exhaustion, over-stimulation, constipation, and sleep deprivation.

Many students demonstrate specific early symptoms of seizures before they occur. The close supervision and observation that is common in the classroom makes educators and teaching assistants unusually well-positioned to observe and document these early signs.

Dietary treatments for epilepsy are mainstream and medically supervised. These dietary remedies (such as the ketogenic diet, modified Atkins diet, and low glycemic index treatment) are some of the most

effective seizure preventatives and are associated with the least cognitive side effects of all seizure treatment options. However, these diets are difficult for families to enact unless everyone around the student is able to support compliance with the diet. Educators can learn about the specific dietary rules for each student who is receiving one of these treatments. Just a single carbohydrate-rich treat can shift a student's body chemistry enough to lose seizure control. Students who are using dietary remedies need support in the classroom so they are not tempted by treats they cannot afford to enjoy.

Communication dictionaries are a tool that enables those who are closest to the student to document how the student communicates a range of issues and concerns. School teams become intimately familiar with their students with Angelman and are important sources of information about how the student expresses a range of feelings and emotions. Each student likely has a unique set of behaviours that communicate anxiety, anger, pain, or the onset of seizures.

Educators and families can pool their collective knowledge of the student and record it in a [communication dictionary](#) or chart. [Communication charts](#) are a simple way to record what a person communicates with his or her behaviour.

Additional Resources

[Management of Angelman Syndrome: A Clinical Guideline](#)

[Growing Up with Epilepsy](#), Massachusetts General Hospital

[Teaching Students with Epilepsy: Strategies for Educators](#)

[Sample seizure diary](#)

Autism in Angelman syndrome

There are many similarities in characteristics between students diagnosed with autism and some students diagnosed with Angelman syndrome. The most common similarity is the disproportionate delay in language and some social interaction skills that is common in both disorders. Studies measuring the prevalence of autism in students with Angelman syndrome have yielded inconsistent results.

A substantial number of students with Angelman syndrome have autistic traits. Compared to their peers with Angelman only, these students who have symptoms of autism:

- are not observed to direct their vocalizations at others;
- initiate fewer social overtures;
- have greater delays in play skills and joint attention;
- are more focused on the repetitive use of objects;
- show less shared enjoyment in their interactions with others; and
- may not respond to their names being called.

Students with Angelman syndrome who do not have traits of autism have an overall faster rate of development, direct their vocalizations effectively at others, and use more non-verbal gestures to enhance their communication. Students with Angelman who do not have traits of autism display more evidence of communicative intent, such as establishing joint attention before gesturing or vocalizing, and escalating their communicative signals to achieve a goal. While most students with Angelman make unusual use of their eye gaze, students with Angelman only may demonstrate unusually intense eye gaze while students with both Angelman and autism may appear aloof and avoid eye gaze. These differences between students with Angelman syndrome only versus those who also have traits of autism persist even as both groups' cognitive development grows over time.

Autism characteristics may compound the effects of Angelman syndrome on affected students, particularly in

the realm of communication. However, there are important differences between students diagnosed with both Angelman syndrome and autism, and students diagnosed with autism only.

Students with both Angelman and autism are less likely to have repetitive sensory motor behaviours; these behaviours tend to be limited to mouthing, ceaseless movement, or hand-flapping when excited, and these behaviours decline as cognitive growth occurs.

Compared to their peers with autism only, students diagnosed with both Angelman and autism are unlikely to be preoccupied with movement (such as finger-flicking or spinning objects), with predictability (such as need for sameness in the environment and in routines), and with order (such as lining up objects).

Students with both Angelman syndrome and autism demonstrate stronger social reciprocity than students with autism only, such as expressing enjoyment in shared activities and showing a positive response to praise and gestures of affection.

Educators and clinicians can decide on an individual basis whether or not it is helpful to understand a student with Angelman syndrome as also having autism. Some students may benefit from autism-specific services. However, it may also be useful to assess individual students who demonstrate symptoms of autism for signs of dyspraxia, poor sensory integration, and visual and auditory processing challenges. The symptoms of autism that are observed in these students may be explained by these other disorders. Students with autism frequently share these same challenges with dyspraxia and sensory modulation; however, isolating the effects of motor planning and sensory modulation disorders from traits understood as autistic might help explain whether the core deficit observed in the student is one of social interaction, versus one of how the student receives and processes sensory information and engages in motor planning.

Cortical Vision Impairment

Students with Angelman syndrome have many risk factors for cortical vision impairment (CVI), such as intellectual disability, a neurological disorder, and epilepsy. Students with CVI will make unusual use of their eye gaze and vision. Educators and parents of students with Angelman syndrome should be aware of the characteristic behaviours associated with CVI so that they can seek assistance from a vision specialist to support the child's learning if these behaviours are observed.

Children with CVI may be unable to attend to models of sign language, may struggle to discriminate between visual symbols, or may be unable to attend to blocks of written text. CVI can have a direct effect on the student's ability to learn to use symbolic communication systems if the student's visual needs are not met.

A student with CVI who has poor contrast sensitivity needs high-contrast visual stimulation. S/he will struggle to perceive visual information in low-contrast contexts: s/he may see only a blank visual slate in contexts such as being outdoors on a cloudy winter day or if her desk faces a bank of windows in the school classroom. S/he may struggle to make sense of facial expressions. Facial expressions are an example of low-

contrast shadows in motion ([Roman-Lantzy, 2007](#)); the face can appear to be a motionless flat surface to students with CVI.



For a child who demonstrates a low level of social responsiveness, adults may be able to support greater responsiveness by enhancing her access to visual information through closer proximity.

Students with Angelman syndrome who have CVI may need vision supports such as:

- high-contrast visual aids and adapted communication symbols so that they can visually attend to and discriminate between visual symbols;
- reduced visual clutter in their classroom and at their work stations;
- a reduced visual field, such as displays with fewer objects or symbols to view at a time;
- attention to lighting to prevent glare and shadows from appearing in their workspace;
- matte rather than glossy laminate, to reduce glare, or waterproof/tear-proof paper to avoid laminate altogether;
- visual objects presented on a black background;
- visual symbols and text presented on a yellow background;
- a daily schedule that structures a student's educational program so that the most visually challenging tasks occur in the morning before the student's vision is fatigued, and
- frequent breaks to rest vision.



Literacy Instruction

The individualized education programs of students with Angelman syndrome tend to emphasize functional skill development, such as independent self-care and communicating personal wants and needs. Parents of students with Angelman syndrome often prioritize the development of social skills, communication, and independence over academic instruction in areas such as literacy ([Leyser & Kirk, 2011](#)).

The development of literacy skills are functional for students with Angelman syndrome. Research demonstrates that comprehensive literacy instruction may be the most effective means to support language development and the ability to actively and independently participate in shared social experiences. Comprehensive literacy instruction targets some of the skills that students with Angelman syndrome most need: receptive vocabulary development and expressive communication.

Educators can ensure that literacy experiences are enriching for students with Angelman syndrome by emphasizing the communication and social aspects of literacy experiences. Literacy development is inherently social and communicative. Literacy skills include learning to enjoy words, stories, and books as they are read aloud. It includes telling our own stories and enjoying and relating to the stories of others. Reading is an essential way to learn more about the world around us.

Conventional literacy skills provide access to the alphabet, the most specific and universal graphic symbol set for communicating with others and exercising control over one's own life. Even just basic knowledge of the alphabet can permit a student to use letters to enhance his or her communication.

Most students with Angelman syndrome are at the emergent stage of literacy skill development. Emergent literacy awareness includes the understanding that sounds and words can be represented by letters of the

alphabet, that books are read from left to right and top to bottom, and that text carries meaning. Emergent literacy behaviours precede and lead to conventional literacy skills such as reading and writing. Emergent literacy behaviours include observing and imitating the functional use of print, browsing through books while observing the conventions of print, and drawing and writing (including scribbling) to share stories or represent meaning.



Emergent literacy skills help develop the ability to communicate using a symbol system, to enjoy shared storybook reading, and to understand and share in conversation and story-telling.

Students with significant disabilities appear to learn literacy skills in a manner that is similar to their non-disabled peers. Students with Angelman require the same comprehensive literacy instruction as their peers without disabilities. These students require more intensive instruction over longer periods of time, paired with careful attention to how to make quality instruction accessible given the nature of their differences in communication, cognition, attention, behaviour, and sensory and physical differences ([Erickson & Koppenhaver, 2007](#)).

Comprehensive literacy programs include phonological and phonemic awareness, vocabulary development and automatic word identification, reading and listening comprehension, writing, and reading independently for a variety of purposes.



Most students with Angelman syndrome have emergent literacy skills. Educators can measure and document progress in emergent literacy behaviours using tools such as the Bridge, an observation-based assessment portfolio-rating scale of the earliest literacy skills. The Bridge organizes the earliest emergent literacy skills into categories that include foundations of reading (such as awareness of books and print), foundations of writing, alphabet knowledge, phonological and phonemic awareness, and oral language. These categories assist educators to identify specific areas of strength and need in the student's overall literacy learning. The portfolio rating scale provides a useful structure for developing individual learning goals and discussing literacy with family members.

Emergent writing is likely the best way to assess literacy development in students with significant disabilities. Writing is the cognitive act of translating thoughts from your head into a symbolic form that others can comprehend. The cognitive process of translating thoughts into symbolic form is the same whether a student is translating thoughts into symbols for communication or letters for writing. Emergent writing activities give students access to the entire

alphabet to generate messages and explore how letters can be combined and re-combined to express different meanings. Students with Angelman syndrome need access to writing activities for meaningful purposes.

Students with Angelman syndrome require strategically planned assistance to access quality literacy instruction. These students have fine motor and gross motor disabilities that may prevent them from physically accessing books and reading materials. They may require adapted texts to support their cognitive and attention needs. Many of these students may be most attentive to stories that are directly related to their own personal experiences; sharing texts about their own experiences and interests has the added benefit of supporting friendship development and communication. Students with Angelman require AAC in order to share in communication about their literacy experiences, such as commenting on texts, requesting a text to be read, or making connections with the text. Most importantly, these students require opportunities to engage in literacy experiences for personally meaningful experiences.

Additional Resources

[The Bridge: An authentic literacy assessment strategy for individualizing and informing practice with young children with disabilities](#)

[Resources for educators to use the Bridge](#)

[PowerPoint book templates](#) for accessible electronic books

Sample [IEP goals for emergent literacy](#)

[Toward Positive Literacy Outcomes for Students with Significant Developmental Disabilities](#)

[Tar Heel Reader](#): free online library of accessible texts for beginning readers of all ages

[“Alternative pencils”](#) accessible writing for all students.

Chapter 4: Summary

Before attempting this project, I was frustrated by the lack of useful information found in the research literature on Angelman syndrome. It seemed to me that our children living with the syndrome (and even ourselves as their mothers and fathers) were just research subjects whose abnormalities and differences had been painstakingly documented but not necessarily for our own benefit. I felt that much of the research literature was a self-fulfilling prophecy constraining the opportunities that might be extended to students affected by the syndrome. My experience with the research literature was that it provided minimal direction for a teacher or parent to craft an appropriate response to our children's learning needs: the literature could tell you what our children and students cannot do but not how they learn and how we need to teach them. There was such a paucity of research on students with Angelman syndrome as learners in the classroom that I never expected to find enough material to develop this project.

However, once I encountered Erickson and Koppenhaver's (2007) description of the six areas of difference observed in students with disabilities, I found a framework to organize the information that researchers had gathered about our children and students with Angelman in a way that could be potentially useful. I am satisfied that this review of the literature is comprehensive, and that the research has been thoroughly mined for what it has to offer parents and educators as we plan for our children and students with Angelman. I am satisfied that the Angelman Syndrome for Educators document will assist educators to find some of the resources and information that will help them teach their learners with Angelman.

The research on students with Angelman syndrome as learners in the classroom is still painfully lacking. We don't know what our students with Angelman know, because we lack strategies and tools to adequately assess them and support them to demonstrate their knowledge. This project exposes the glaring lack of a comprehensive, cohesive curriculum to guide learning for our students with Angelman,

particularly in the areas of literacy and communication. The biggest lesson I learned as I pursued this project was that the information our educators need to teach our children and students with Angelman is scattered and piecemeal, in pockets of best practices and expertise, rather than widely accessible and effectively disseminated through our teacher's colleges and professional development initiatives. My conclusion is that only the most highly motivated and engaged educators and parents will currently find the information that can help them maximize learning for our students. I hope this project helps to make this process easier.

Throughout Maggie's school career, I have struggled with knowing when the label of Angelman syndrome is helpful versus damaging. My biggest concern developing this project was that the materials contained within it would serve to further define and limit students with Angelman by their medical diagnosis, rather than describing their similarities to other students and emphasizing their learning potential. It remains to be seen whether or not this project will do more good than harm. I've concluded, however, that most educators want to teach their students with Angelman but feel they lack the specific tools and strategies to adapt their existing practices and knowledge to our children and students. I hope this project will fill some of that gap so that educators can recognize students with Angelman as familiar and as potentially literate. For now, given the low expectations for learning generally expressed about students with Angelman, and the lack of materials for educators and parents of these students, I feel this project is long-overdue and on the whole helpful. I hope that this project will be a useful tool to raise expectations for learning. I truly hope it will provide a road-map for educators and parents searching for routes to maximize learning opportunities for their students with Angelman.

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