Angelman Syndrome
for Educators
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Introduction

Students with Angelman syndrome (AS) are characterized as cheerful, inquisitive, hands-on learners in the classroom. The disabilities associated with this 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 (IEPs) to meet these student’s needs.

This booklet was created for school teams to assist with planning for students with AS.

- **General educators** will learn what differences they might expect in a student with AS 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 to access instruction and demonstrate learning.
- **Parents** will find information to help them participate in discussions about their child’s individual learning characteristics and planning their child’s placement and program.

This booklet is designed to:

- explain the general characteristics we expect to see in students with AS
- 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 a student, 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

Note: for simplicity and the purposes of this booklet, the term ‘AS’ refers to Angelman syndrome, and ‘students with AS’ or ‘students’ refers to students with Angelman syndrome.
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 AS are learners who require careful planning and accommodation in the classroom.

Communication
The most significant learning difference observed in nearly every student is a profound expressive language disability. Expressive communication is disproportionately impaired compared to overall comprehension, cognition, and global development. Students generally demonstrate strong motivation to communicate and interact with peers and adults. What a student can express should not be mistaken for what that student can understand.

The nature of the communication disorder in AS is fundamentally one of a struggle to access and express language. Students need intense support to learn how to communicate effectively to meet their social and academic needs. They 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 AS 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 AS 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 AS and often makes it difficult for them to perform on command. Students with dyspraxia may best demonstrate their knowledge in natural contexts.

**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 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 AS, but most of these students require support to integrate information received through their senses. All should be assessed for sensory integration dysfunction so that appropriate strategies can be implemented. Students 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 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

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*Literacy development is possible in students with AS and most 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.*
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 can express what they need to say when they need to say it.

**Attention**

Students have attention skills that are consistent with their overall development, particularly when engaged in preferred activities. The attention challenges 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.

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**Communication**

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

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

Students use a wide variety of expressive strategies in an attempt to meet their daily communication needs. They develop strong preferences for whatever system is the most efficient to convey the messages they are motivated to express. A minority of students develop some oral language or manual sign language. The majority, 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

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**Additional Resources:**

An Introduction to Angelman syndrome by ASSERT, UK
Facts About Angelman Syndrome, 7th Ed., 2009, by the Angelman Syndrome Foundation, USA
communication is inadequate to express abstract thoughts, such as messages related to the past or future (Beukelman & Mirenda, 2012). Students often rely more on unaided communication, which may reflect the instructional approaches generally used. According to findings by Dr Stephen N. Calculator (2013), only 5% of the instructional approaches used with students 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.

**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. Assessment for an AAC system takes into account how the student will access the system and how barriers to its use will be removed.

**Students with Angelman syndrome require aided communication support in order to meet their communication needs.**

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.

**AAC systems should provide access to enough language that the student can meet his or her daily communication needs, and so that communication partners can use the device to augment their communication with the student.**

Students with AS 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.
The Participation Model

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
  - supporting a family’s request for student placement in a language-rich regular classroom.

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.
Aided Language Input

Children acquire spoken language through constant immersion in their native language. They 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.”

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).

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. They 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.

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.

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).
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 he/she can access. 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. They 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)
PODD is 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. They are designed to meet the communication needs of both the child and his/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 also be programmed on dynamic display speech-generating devices.

The System for Augmenting Language
This form of aided language input, teaches 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, e.g. the iPad. 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.

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.
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.

Mouse models of Angelman syndrome suggest that the cognitive deficits of the syndrome may be specific to storing and retrieving information from memory. The mice demonstrate a disproportionate deficit in experience-dependent memory and 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, they 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 AS experience a similar memory deficit, educators may find it helpful to focus on supports for memory formation.

Students often crave multisensory 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. Therefore, multi-sensory learning experiences with a strong emotional component, form stronger schemes.

Alternative assessments that can separate motor skill performance tasks from cognitive tasks are often essential to capture what a student with AS understands.

Angelman syndrome 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 AS 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 their intellectual ability. 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 AS. 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 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.

Many students with Angelman syndrome 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. It 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.

The Bridge

One example of a validated portfolio assessment tool is 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.

Artefacts 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.

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 AS 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.
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 AS can walk independently by the time they reach school age. They 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 will always need to exert intense conscious effort to maintain stability while walking. Students with AS demonstrate a “central dyssynchrony 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
- are slow to respond to verbal instructions

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

Some students with AS will never walk independently. Those 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.

Students with AS 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.

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.

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

‘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.

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.
Senses

Sensory disabilities such as vision loss or hearing impairment are not commonly reported in Angelman syndrome; however, AS is primarily a neurological disorder. Sensory challenges observed in students 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 these students.

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 AS include strabismus and low acuity, both of which can be treated by an ophthalmologist. The characteristics of AS are associated with heightened risk for cortical vision impairment (CVI). 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
- 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/processing refers to how the brain organizes sensation received through the body in order to function in the environment. Students with AS have a high rate of global sensory dysfunction and hypo-responsiveness to sensory stimuli. **This 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 may also 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 in order 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.

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
- provide verbal or auditory cues in lieu of visual or physical cues such as body language

Combined hearing and visual impairment
Some students with AS 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. These students 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; e.g. 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, curl up amongst pillows or in a small enclosed space
- dimmer switches on lights to reduce visual input;
- headphones 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

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.
Attention

Students with Angelman syndrome generally have stronger attention skills than students without AS who present with the same level of cognitive ability. They are usually physically active and inquisitive and are often prone to excess activity and hypermotoric behaviours that require management in the classroom. Students often demonstrate the greatest attention when engaged in personally meaningful and enjoyable activities. Linking instruction to their 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 can usually engage in joint attention, a shared focus on both an object and another individual. Those 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.

Dyspraxia is common in AS 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.

Some students with AS demonstrate an intense need for physical movement. They tend to be constantly moving, may frequently invade the space of others, 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.

Distractibility

Attention challenges in students 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 extraneous input. Many students 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
- in the gym or on the playground with 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 in students with AS. 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.

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 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.

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. However, a laminated display of symbols might be too distracting because of the sensory properties of its shiny texture so the student can’t also attend to the symbols on the page. A matte laminate or durable rip-proof paper can be used to reduce the stimuli of the laminate. Some students may attend better to visual symbols with tear-off Velcro backing; others find the Velcro itself too enticing.

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. The sensory properties of materials can be planned to ensure that they contribute, rather than detract, from the maintenance of attention.
Remnant Books

All students benefit from instruction that is directly relevant to their own lived experience. Students with AS 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.

Make a remnant book

Remnant books are a fun and easy way to create a visual/tactual resource that can help students record important events. These events can then be used as a starting point for communication, writing and literacy topics.

Remnant books can easily be created using inexpensive files, scrapbooks, folders or mini photo albums. Each time the student participates in some type of event, they collect artefacts and pictures from that event. The student should be involved in deciding what goes in their remnant book.

Examples of remnants: Entry ticket stubs, brochures, menus, party hat, balloon, napkins with logo or containers from fast food restaurants, pictures from parties or holidays, pamphlet or business card from doctor’s visit...anything that will serve as a reminder of the event and that is important to the student.

Add brief notes with the remnants - the date and key details of the event in first person language, e.g. “I visited the aquarium”
Affect

Students with Angelman syndrome are generally described as cheerful, prone to over-excitement, and highly motivated by the social aspects of learning. Many are exuberant and socially gregarious. They often deploy smiles and laughter as a strategy to attract and maintain attention from others. This strategy is very 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 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 AS. Educators of some students may want to strategically plan the use of adult attention and eye contact to facilitate classroom management.

Challenging behaviours

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 behaviours that are observed in students with AS. Behaviours such as grabbing are common. 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 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 use aggressive behaviours to bring a halt to whatever demands are being placed on them or as a result of feeling unwell or being in pain. It is important to analyse these behaviours from a functional perspective so that educators understand what the student is trying to achieve or trying to communicate.

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 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. In general, students 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.
Cheerful affect
Some students use smiles and laughter as a strategy to distract adults from making a demand on them. Many laugh to express a range of emotional states, including shyness, anxiety, discomfort, and pain. Because of the limited repertoire of communication skills and the struggle to organize planned motor responses, smiles and laughter become the default expression of many complex emotional states.

The cheerful affect in Angelman syndrome should not be presumed to mean the student is always happy.

Health factors
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 sometimes seen in Angelman syndrome.

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 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 AS 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. They may be affected by the sleep disorder or seizure disorder that is common in Angelman syndrome. Seizure treatments themselves may be interfering with the student’s impulse control. Physician support to manage the sleep and epilepsy may be helpful. Over-activity is also 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 AS can also strategically engage in challenging behaviours. These behaviours serve a functional purpose; for example, to 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**

Functional behaviour assessment uncovers the purpose behind the behaviour by carefully analysing 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.

**Epilepsy**

Most students with AS have epilepsy (approx. 80%). Some types of seizures that are common in AS 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 and an appropriate emergency plan. 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
- absence spells, staring or chewing during periods of very reduced or absent consciousness

Epilepsy is usually managed with medications. These 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.
Non-Convulsive Status Epilepticus (NCSE)

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’. NCSE 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.

NCSE 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 NCSE. It can also 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 disrupts learning, heightens the risk for more emergent seizures and usually requires medical management to help students return to their baseline. Low-carbohydrate, high-fat dietary remedies for epilepsy have the lowest rate of side effects and are often very effective.

Sleep disorder

Many children with Angelman have a significant sleep disorder and medical management may be considered. 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, functioning and behaviour.

Pain

Students with AS 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.

They have risk factors for pain from gastroesophageal reflux, constipation, and headache. Some students express pain or anxiety with intense episodes of laughter.

Obesity

Students are at increased risk of obesity and loss of independent mobility as they age. Some 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 therefore 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, and can alert parents to the signs and symptoms they observe. Because students spend a substantial portion of their day in the classroom, documentation collected at school can help families and physicians make treatment decisions.

Epilepsy

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. 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.

Common seizure triggers include illness, fever, infection, exhaustion, over-stimulation, constipation, and sleep deprivation.
Dietary treatments

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 preventative treatments 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 charts

Each student has a unique set of behaviours and gestures that communicate anxiety, anger, pain, or the onset of seizures. School teams become intimately familiar with their students and are important sources of information about how the student expresses a range of feelings and emotions. 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 and helpful way to record what a person communicates with his/her behaviour and gestures.

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.

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 are common in both disorders. Studies measuring the prevalence of autism in students with AS have yielded inconsistent results.

A substantial number of students with AS have autistic traits. Compared to their peers with AS 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
- may not respond to their names being called
Students with AS 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. They 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 AS make unusual use of their eye gaze, students with AS only, may demonstrate unusually intense eye gaze while students with AS and autism, may appear aloof and avoid eye gaze. These differences between students with AS 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 AS and autism, and students diagnosed with autism only. Students with AS 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 AS 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).

Educators and clinicians can decide on an individual basis whether or not it is helpful to understand a student with AS, 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 the 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.

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*Students with 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.*
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 AS 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
- struggle to discriminate between visual symbols
- unable to attend to blocks of written text

Students with CVI who have poor contrast sensitivity need high-contrast visual stimulation. 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. They:
- will struggle to perceive visual information in low-contrast contexts
- may see only a blank visual slate in contexts such as being outdoors on a cloudy winter day or if their desk faces a bank of windows in the school classroom
- 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 the child’s 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
- frequent breaks to rest vision
Literacy Instruction

The individualized education programs (IEPs) 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 AS 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 AS. 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 AS most need: receptive vocabulary development and expressive communication.

Educators can ensure that literacy experiences are enriching for students with AS 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.

The emergent literacy stage

Most students with AS 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. Students with AS require the same comprehensive literacy instruction as their peers without disabilities. Emergent literacy behaviours:

- precede and lead to conventional literacy skills such as reading and writing
- include observing and imitating the functional use of print, browsing through books while observing the conventions of print, 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 nondisabled peers.

Students with significant disabilities 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

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 AS 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
- 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**

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 AS need access to writing activities for meaningful purposes. They 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.

**Augmentative and Alternative Communication (AAC)**

Students 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, students with Angelman syndrome require opportunities to engage in literacy experiences for personally meaningful experiences.*

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Acknowledgments

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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