Neurobehavioral Approaches in Angelman syndrome: Part II

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What you will learn about today

- Talk is organized around:
  - Health Watch Table for Individuals with AS (AS-HWT)
  - On-line Behavior Modules
- Learn how these resources can be used to help individuals with AS receive appropriate care and support to maximize their physical, behavioral and mental health

About Us

- Jane Summers, PhD (psychologist), CAMH in Toronto
- Erick Sell, MD (neurologist), CHEO in Ottawa
- Mutual interest in seeing people with AS
- Neurology + behavior = Neurobehavioral approach
- Clinical aims:
  - Improve physical and emotional health, reduce occurrence of challenging behavior
  - Enable individual with AS to reach full potential, maximize quality of life

Health Watch Table — Angelman Syndrome (AS)

Forster-Gibson, Berg and Korossy 2015

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- Earlier today, Dr. Sell talked about health-related issues in AS with goal of optimizing physical health
- Current talk – optimize behavioral and mental health
- Refer to health watch table and behavioral modules

Health Watch Table — Angelman Syndrome (AS)
Forster-Gibson, Berg and Korossy 2015

AS Health Watch Table (AS-HWT)

- Developed in 2015 by Cynthia Forster-Gibson, Joseph Berg and Marika Korossy
- Purpose is to provide anticipatory guidance for health/mental health concerns, monitor health/mental health needs across all ages/stages
- Reviews physical and behavioural/mental health issues and provides recommendation or guidance for how to manage them

www.surreyplace.on.ca/documents/Primary%20Care/HWT-AS10Sep2015.pdf

Health Watch Table — Angelman Syndrome (AS)
Forster-Gibson, Berg and Korossy 2015

Areas covered in AS-HWT*

- Genetics
- Neurology
- Musculoskeletal
- HEENT (Head, Eyes, Ears, Nose, Throat)
- Gastrointestinal
- Dental
- Endocrine & Sexuality
- Mental health/behavioral

* *Toileting*
* Safety
* *Pain/discomfort*

* We have added our own information in italics

Mental Health/Behavioral

- Frequent laughter, apparently happy demeanor, easy excitability, hyperactivity, sleep disturbance, and aggressive behaviours such as grabbing and pulling, but not self-injury, are common

RECOMMENDATIONS: For hyperactivity, arrange consultation with occupational therapist for sensory issues, behavior therapist and then a psychiatrist to consider medication management for hyperactivity (atypical response to stimulant medication may occur). *Shape paying attention for longer periods of time; rearrange physical and teaching environment.*

Sleep Disturbance

High rates of sleep problems. Problems include decreased need for sleep, abnormal sleep-wake cycle, difficulties falling asleep or remaining asleep, reduced total sleep time, disruptive bedtime behaviors. Problems can have a significant impact on family functioning. Sleep is most affected in infancy and middle childhood and tends to improve in adolescence and adulthood.

RECOMMENDATIONS:

Implement consistent nighttime routine; *modify sleep environment;* consider trial of melatonin; possible referral to sleep specialist or clinic. *Consider referral to behavioral specialist to assess parent-child interactions. Sleep apnea may be an issue but can be difficult to diagnose and treat. Removal of tonsils/adenoids, weight loss may help.*

Sleep

Sleep and Biological Rhythms 2008; **6**: 95–101 doi:10.1111/j.1479-8425.2008.00339.x

ORIGINAL ARTICLE

Sleep disturbance in adults with Angelman syndrome

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John M SHNEERSON[2] and Graeme G LENNOX[3]

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Abstract

Angelman syndrome is a neurodevelopmental disorder characterized by severe learning difficulties, epilepsy, and a typical behavioral phenotype. The diagnostic criteria state that 20–80% of individuals have decreased sleep need and abnormal sleep–wake cycles. A wide variety of sleep problems have been reported, including reduced total sleep time, frequent night awakenings and nocturnal enuresis. Most previous reports have used sleep questionnaires to assess the frequency of various sleep disorders. Most patients studied have been children or adolescents and only one previous study has used complex sleep studies (polysomnography). We report three adult sisters with Angelman's syndrome who have been assessed with sleep diaries, actigraphy and, in one case, overnight polysomnography. Despite sleep diaries showing prolonged sleep with a mean of 9 h a night with few nocturnal arousals, the actigraphy in all three patients showed increased sleep fragmentation and in one the polysomnography was strikingly abnormal, with a greatly reduced total sleep time and very frequent, predominantly obstructive sleep apneas fragmenting night sleep (desaturation index 63.3 of total sleep time). Despite over 10 h in bed, she had only 69.5 min of actual sleep. There was no evidence of a circadian rhythm disorder. This is the first report of polysomnography in an adult with Angelman syndrome and it highlights the need to look for the presence of sleep apnea in this group, which may be an under-recognized cause of nocturnal sleep disturbance.

Pain and Discomfort

Pain and discomfort can present as distress, sleep disturbance or behavioral changes, such as an increase in aggression or irritability. Facial expression and vocalizations can change.

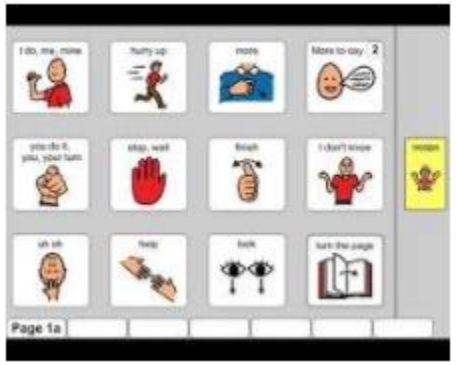
RECOMMENDATIONS:

Investigate and treat possible physical cause (GERD, headache, toothache, sore foot, uncomfortable clothing, etc.). Look for and address environmental factors (too loud, too hot/cold, too crowded).

Non-Communicating Children's Pain Checklist

(NCCPC; Breau et al., 2002)

- **Vocal** (moaning, whining, whimpering, crying, screaming)
- **Eating/sleeping** (eating less, increase or decrease in sleep)
- **Social** (cranky, irritable, less interaction, seeking comfort or contact)
- **Facial** (furrowed brow, not smiling, lips tight or puckered, clenching or grinding teeth, chewing)
- **Activity** (not moving, less active, agitated, fidgety)
- **Body/limb** (floppy, stiff, gesture to part of body that hurts, flinch or move body part away)



Mental Health/Behavioral

(continued)

- Language development is variably, though markedly impaired – majority do not develop speech; receptive language skills are always more advanced than expressive language skills and continue to improve even in adulthood.

RECOMMENDATIONS: Early and ongoing intervention by speech-language therapist is essential and should focus on nonverbal forms of communication. Use of augmentative and alternative communication aids, such as *Pragmatic Organization Dynamic Display*, picture cards, communication boards, *speech generating devices*, should be encouraged.

Aggressive behavior

- behavior that can result in harm to oneself, others or the environment
- common forms of aggression in AS: hair pulling, grabbing, pinching, kicking, slapping
- in the absence of verbal and motor skills, aggression may be a way to "tell" people:
  - to pay more attention or stay connected
  - want something
  - don't like/don't want something
  - not feeling well or unhappy

Mental Health/Behavioural

(continued)

- Interventions based on applied behaviour analysis (ABA) are being used to teach adaptive and communication skills to improve individuals' functioning and address behaviours that challenge people and services.

RECOMMENDATIONS: Visit www.angelmanbehaviors.org.

If behaviour changes/occur, evaluate for medical cause. *Consider emotional needs and possible mental health issues*.

On-Line Behavior Modules

- Social and environmental influences on aggressive behavior
- Aggression as a communicative behavior
- Cognitive and sensory issues
- Mental health influences on aggressive behavior
- Neurologic and medical influences on aggressive behavior

www.angelmanbehaviors.org

Mental Health/Behavioral

(continued)

- Emotional needs are often neglected in severe disability combined with limited communication

RECOMMENDATIONS: Work with the individual and family to optimize opportunities for inclusion, participation and friendship. *Watch for changes in behavior that can occur in relation to stressors or major life events (moves, end of school, loss of family, friends, workers). Use tools that have been developed to identify possible signs of mental health problems in individuals with severe intellectual disability. Signs may include increased irritability and agitation or changes (increase/decrease) in sleep, energy, appetite and mood. Consider referral to psychologist or psychiatrist.*

Tools for assessing behavioral/emotional disturbance

Characteristics of Depression as Assessed by the *Diagnostic Assessment for the Severely Handicapped-II (DASH-II)*

Johnny L. Matson, Karena S. Rush, Martha Hamilton, Stephen J. Anderson, Jay W. Bamburg, and Christopher S. Baglio

Louisiana State University

Don Williams and Sharon Kirkpatrick–Sanchez

The Developmental Behavior Checklist: The Development and Validation of an Instrument to Assess Behavioral and Emotional Disturbance in Children and Adolescents with Mental Retardation[1]

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Monash University Centre for Developmental Psychiatry

Distinctive Pattern of Behavioral Functioning in Angelman Syndrome

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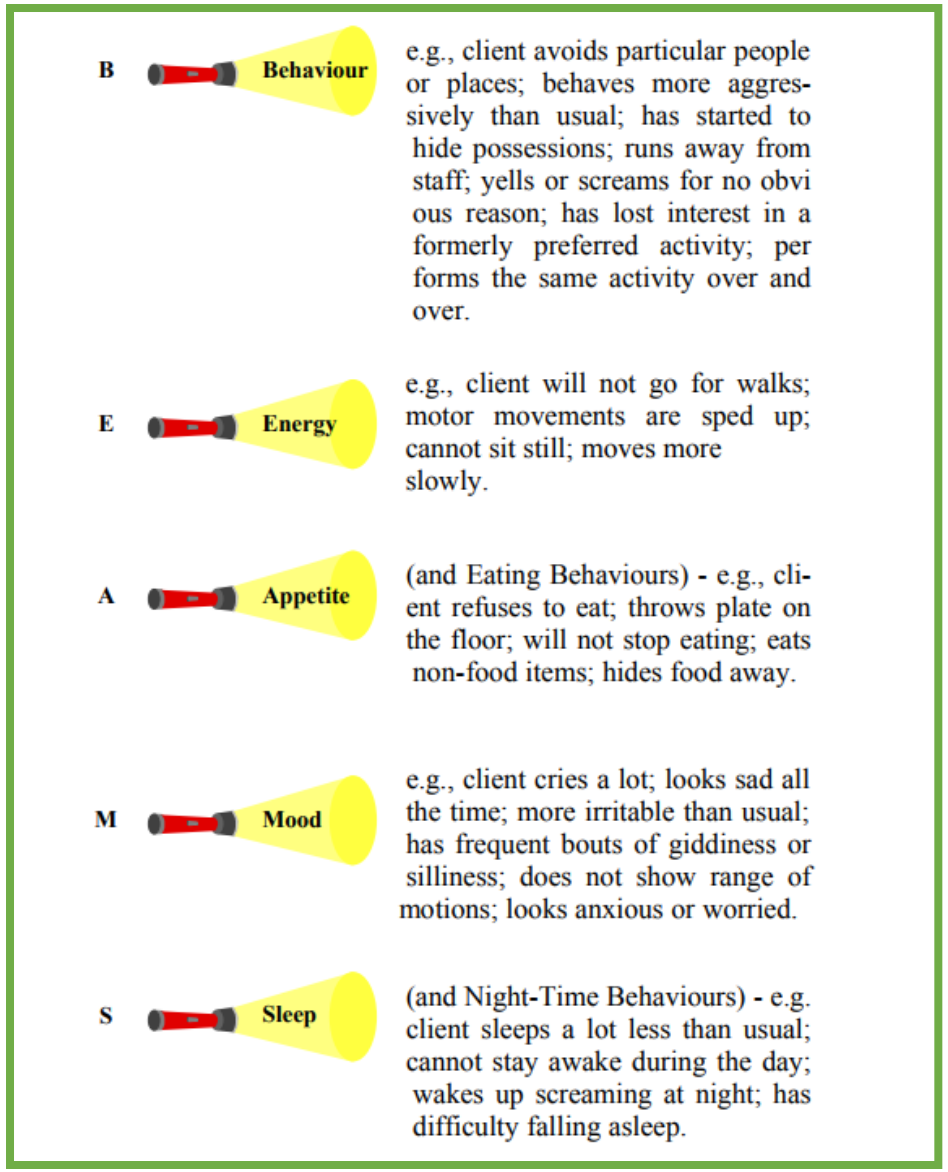
Hamilton Health Sciences Corporation (Hamilton, Ontario)

Maurice A. Feldman
Queen's University (Kingston, Ontario)

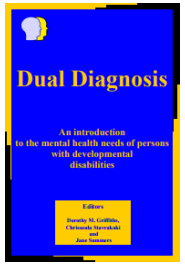
Angelman syndrome is a neurogenetic disorder that is characterized by impairments in neurological, motor, and intellectual functioning. This study compared 27 participants with Angelman syndrome to clinical and community participants with developmental disabilities of mixed etiology to determine whether Angelman syndrome is associated with a distinctive pattern of behavioral functioning. The groups with and without Angelman syndrome were matched on chronological age, gender, and level of intellectual functioning. The dependent measure was parent ratings of maladaptive behavior using the Aberrant Behavior Checklist. The Angelman syndrome group had significantly lower scores on measures of irritability and lethargy. Results contribute to the delineation of a behavior phenotype for the syndrome.

"BEAMS"

Duration
Breadth
Intensity



Reid, J., **Summers, J.**, Adamson, J., Habjan, B., Meister, C., & Gignac, V. (2002). Mental health issues in clients with severe communication impairments. In D. Griffiths, C. Stavrakaki, & J. Summers (Eds.), *An introduction to the mental health needs of persons with developmental disabilities* (pp. 535-559). Sudbury: Habilitative Mental Health Resource Network.



CASE EXAMPLE: DANIEL

Daniel is a 30-year-old gentleman who lives in a group home with five other adults with developmental disabilities. He lived at home until six months ago when he required urgent placement in a group home due to his elderly parents' declining health. He takes three different anticonvulsants for seizure control. Daniel has a history of choking and coughing at mealtimes and suffers from recurrent pneumonia. Sometimes he will go several days without having a bowel movement. He is able to walk short distances with staff assistance but is very unsteady on his feet and has fallen a number of times.

“DANIEL”

The staff don’t know a great deal about how Daniel communicates apart from reaching toward desired items, primarily other clients’ food, or pushing aside things that he doesn’t want such as his own food. They are expressing concern because Daniel has started to hit them at different times during the day, often without a clear reason why. He has also lost interest in some of the activities he used to enjoy, such as watching videos and listening to music. Sometimes Daniel starts to cry and makes distressed vocalizations

“DANIEL”

Daniel’s team found that he is more likely to engage in physical aggression or display signs of sadness (a) when he is given pureed foods to eat, (b) a short time after meals or when straining to have a bowel movement; (c) when his parents leave following a visit, and; (d) when he is asked to walk for any distance. After the behavior occurs, Daniel is often spoken to/comforted by staff, is given assistance, or is able to avoid something unpleasant.

What could Daniel be trying to say?

He seems to be “saying” he misses his family, wants or doesn’t want something, and feels pain or discomfort.

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Behavioral tracking sheet for Daniel

Behavioral Tracking Sheet Name of Individual: Daniel Problem Behaviors: Aggressive behavior and episodes of sadness
Description of Problem Behavior(s): 1. Physical aggression – hitting staff anywhere on their body with an open hand 2. Sadness – crying with tears accompanied by wailing sounds

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Frequency of behavior

Date	Behavioural Count		Total Daily Count
	Physical aggression	Sadness	
May 12/14	✓✓✓	✓	4
May 13/14	✓✓✓	✓✓✓	6
May 14/14			0
May 15/14	✓✓	✓✓✓✓	6
May 16/14	✓✓	✓✓	4
May 17/14			0
TOTALS	10	10	20

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A-B-C chart for Daniel

Date, Time, Person Recording	Setting Events (factors or events that may "set the stage" for the behavior to occur)	A - Antecedents (immediate trigger for the behavior)	B – Behavior (what the behavior looked like)	C – Consequence (what happened immediately after the behavior occurred)	Possible function(s) of the behavior (likely reason why the behavior happened)
May 18/14 12:00 pm Mrs. Buchanan	Daniel was sitting at the table, waiting for lunch	Clients were given their lunch to eat	Hit staff on the arm	Daniel was told to stop hitting and eat his lunch	Daniel doesn't like his pureed food
May 18/14 12:45 pm Mrs. Price	Daniel had finished eating lunch 30 minutes earlier	Nothing identified	Crying and wailing sounds	Staff took him to the toilet and he strained to have a bowel movement	Daniel was in pain from constipation
May 20/14 4:30 pm Ms. Bolton	Clients were getting ready to go on outing	Staff tried to get Daniel up to walk to the van	Hit staff on the arm	Staff had Daniel sit in the wheelchair and pushed him to the van	Daniel didn't want to walk to the van
May 22/14 7:00 pm Ms. Roberts	Daniel's family came to visit	Family said goodbye and left the group home	Crying and wailing sounds	Staff tried to comfort Daniel	Daniel misses his family

Medical, physical or emotional factors that may be linked to challenging behavior for Daniel

Factor	Yes/No		Additional information
	Yes	No	
Illness	☑		Ear infections
Pain or discomfort	☑		When he is constipated
Seizures		☑	
Hunger	☑		Pureed diet isn't appealing
Thirst		☑	
Fatigue		☑	
Change in medication		☑	
Change in diet	☑		Switched to pureed diet recently
Angry/bad mood		☑	
Sad/unhappy mood	☑		After parents leave from a visit
Unusually happy mood		☑	

Team Approach

- Multidisciplinary team to address Daniel's aggressive behavior and bouts of sadness
- Behavior therapist to assist staff to collect and analyze data about patterns of behavior
- Speech-language pathologist to assess Daniel's functional communication needs
- Dietician to find out if there are ways to make his pureed diet more appealing

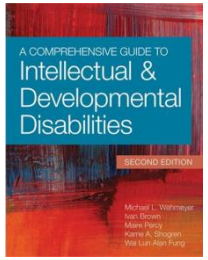
Team Approach (continued)

- Primary care physician for possible pain-related issues such as constipation
- Mental health professional (psychiatrist or psychologist) for possible depression following a major life event of moving to a group home, leaving family behind
- Physiotherapist for mobility limitations

Treatment Principles*

Treatment principles and approaches	Comments & examples
Treat underlying physical and medical conditions that contribute to behavior and mental health problems	• Treat medical conditions that may cause pain and discomfort • Treat vision and hearing problems and neurological conditions that may impair functioning
Treat psychiatric disorders using medication and non-medication approaches according to evidence based practice for specific disorder	• When medication is used, avoid using multiple medications when possible and monitor closely for side effects • Adapt psychological treatment approaches to align with cognitive and communication abilities • Develop strategies to promote mental well-being
Modify the environment and provide needed supports to meet daily life and emotional needs	• Reduce levels of lighting and noise • Decrease crowding • Increase/decrease opportunities for activity and stimulation • Increase access to preferred items, people and activities • Offer choices • Enhance opportunities for inclusion and meaningful relationships and participation

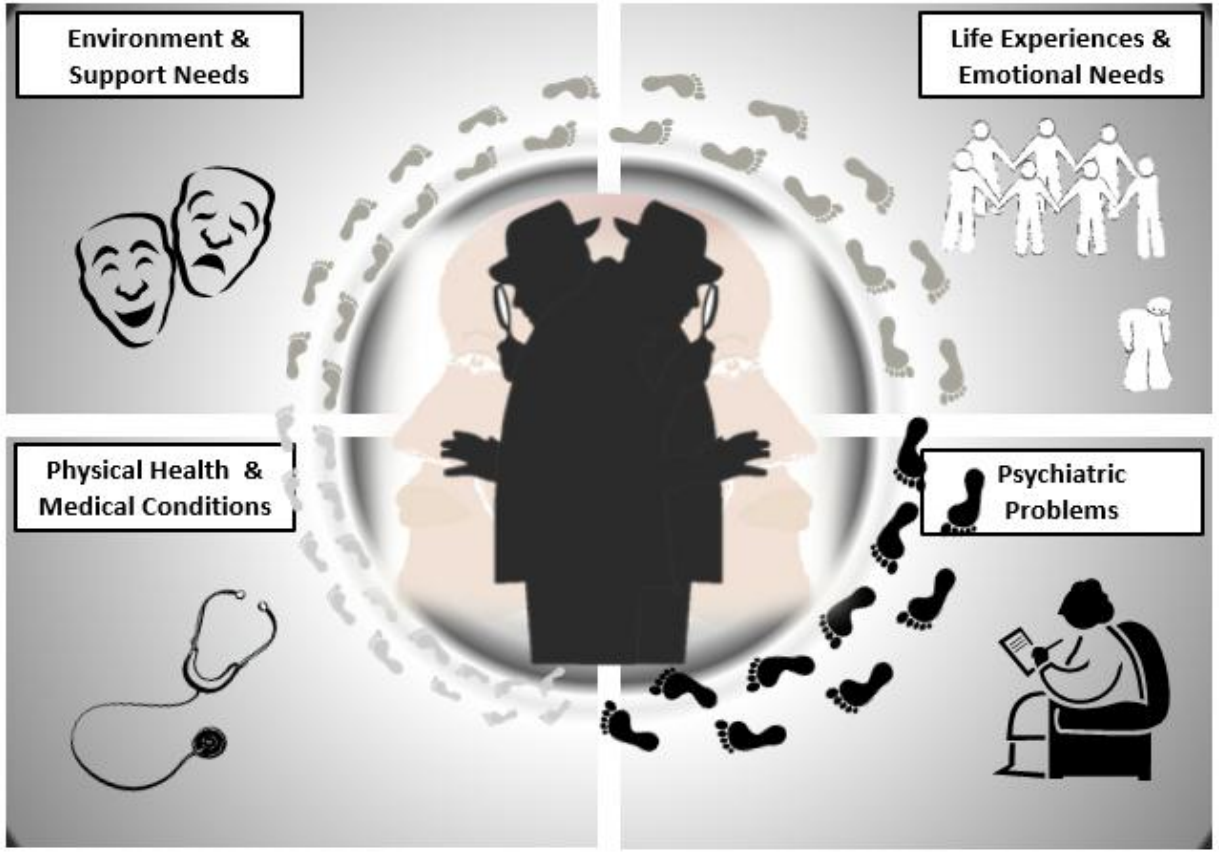
*Summers, J., Fletcher, R., Bradley, E. People with intellectual disabilities and mental health needs (pp. 679-694).



Treatment Principles (continued)

Decrease stress	• **Protect from physical, emotional, financial and sexual harm** • **Realign expectations to match abilities** • **Provide appropriate levels of support**
Increase coping and skills	• (Increase/improve) communication, social, self-help, play and leisure skills • Provide opportunities to develop coping skills • Enhance autonomy and self-esteem
Develop a coordinated system of supports	• Ensure coordinated care (e.g., when moving from child to adult system, inpatient to outpatient care) • Identify primary and specialist supports (e.g., family physician, medical specialists, case manager, behavioral specialist) and how and when to obtain these • Develop proactive crisis management protocols
Management versus treatment of behavioral problems	• Focus management on immediate safety of the individual and care providers; psychotropic medication as a short-term intervention may be used • Focus treatment on the underlying cause of the behavior problem; psychotropic medication is used to treat specific psychiatric disorder

Bringing it Together



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SYNDROME DU SYNDROME
SOCIETY D'ANGELMAN

Thanks

- Canadian Angelman Syndrome Society
- Angelman Syndrome Foundation
- Parents and families from Ottawa Clinic

Questions and Comments

- Are we missing any behavioral and mental health issues?
- What are your priorities?