A Synthesis of Current Research in Augmentative and Alternative Communication Practices for Individuals with Angelman Syndrome

Kaitlyn A. Clarke & Janice C. Light

Introduction

- Angelman Syndrome (AS) is a genetic disorder that occurs in approximately 1 in 15,000 births (Angelman Syndrome Foundation, n.d.).
- The primary cause of AS is the deletion of bands 11-13 on the long arm of the maternally inherited 15th chromosome (Clayton-Smith & Laan, 2003) , while other cases of AS are caused by specific defects in imprinting of the UBE3A gene, the replacement of the maternal copy of UBE3A by a second paternal copy, or other unknown origins (Clayton-Smith & Laan, 2003; Jolleff et al, 2006).
- AS is characterized by global developmental delays, little or absent speech, motor disorder—such as ataxia, intellectual disability, hyperactivity, and seizures (Jolleff et al, 2006).
- Individuals with AS' expressive language skills are characterized by limited speech, typically with fewer than five words (Weltman & Weiman, 2016).
- Individuals with AS display stronger comprehension of spoken language than their production (Alvares & Downing, 1998; Weltman & Weiman, 2016).
- Given their phenotypical and developmental profiles, individuals with AS can be characterized as having complex communication needs (CCN). They often require the use of augmentative and alternative communication (AAC).
- There is limited research on the use of AAC with individuals with AS to improve communication skills.

Purpose

- The primary goals of this synthesis are:
  (a) review and synthesize current research in the field of AAC as it relates to AS;
  (b) identify strengths and gaps in the current research;
  (c) determine clinical implications for the use of AAC with individuals with AS.

Methods

SEARCH

Databases:
- EBSCO (Academic Search Complete), ERIC, PsychInfo, PubMed, Linguistics and Language Behavior Abstracts (LLBA), and Pennsylvania State University library

Search term categories
- "Angelman syndrome", "AAC", "augmentative communicat*"

Author and Ancestry Review

INCLUSION CRITERIA

- Peer-reviewed scholarly journal
- Research participants diagnosed with AS
- Study outcomes were related to AAC use
- Experimental studies, qualitative reports from caregivers, or review of clinical data

DATA EXTRACTION AND CODING

- Study authors and design
- Participant age/gender
- Context
- Type of AAC
- Intervention/training
- Target measure/DV
- Gain score
- PND/TauU
- Certainty of evidence

Results

The systematic search identified 3 quantitative and 9 qualitative studies (11 studies total)

Quantitative Studies

PARTICIPANTS

- Participants ranged in age from 21 months to 10 years
- Three males, two females (n=5).
  - 40% two years and under (n=2)
  - 60% nine years and older (n=3).

TYPES OF AAC

- Unaided AAC:
  - Gestures
  - enhanced natural gestures
  - sign
- Aided AAC:
  - graphic symbols
  - vocal output device
  - speech generating devices
  - PECS, picture boards, and object

INTERVENTION CONTEXT

- 20% in the home (n=1)
- 40% in a preschool setting (n=2)
- 60% in the school setting (n=3)

INTERVENTION CHARACTERISTICS

- Prompting
- Modeling
- Aided and unaided AAC
- Structured enhanced natural gesture instructional protocol.

DEPENDENT VARIABLES

- increased requests for preferred items
- increased initiations with clear intentions
- increased spontaneous use of ENG.

Qualitative Studies

PARTICIPANTS

- 654 parent/provider respondents
  - 3 professionals (.46%)
  - 651 including parents (99.54)
  - Individuals with AS that were included ranged from birth to adulthood (age 66), with a total sample of 954.

IDENTIFIED THEMES

- **Acceptability and usefulness-** perceived acceptability and long-term use
- **Effectiveness/success-** perceived effectiveness, effectiveness when used consistently, changes in previously used gestures, and ease of teaching others
- **Willingness-** willingness to utilize and change daily schedules, as well as reasonableness of the intervention
- **Priorities-** summarizes parent perceptions and priorities of AAC use
- **Disadvantages-** perceived disadvantages, disruptiveness, negative side effects, amount of use, and reason for rejection

COLLECTION METHODS

- Likert scale questionnaires
- Web surveys
- Communication Inventories.
- Parent and professional reports

Paper	Acceptability	Effectiveness/ Success	Usefulness	Willingness	Exposure to electronic SGD	Disadvantages	Priorities
Calculator (2002)	X	X				X	
Calculator & Black (2010)							X
Calculator & Diaz-Caneja Sela (2010)	X						
Calculator (2013a)	X				X		
Calculator (2013b)	X					X	X
Calculator (2014)	X	X	X				
Calculator (2015)		X		X			

Discussion and Implications

- Individuals with AS are reported to benefit from AAC supports, evidenced by both the quantitative and qualitative studies.
- The specific AAC interventions and supports that best meet the needs of individuals with AS is still unclear; however, guidance from parent perspectives can be beneficial.
- Acceptance of a device for individuals' with AS was based on increased success in interactions and independence; while parent willingness and perceived usefulness of device was based on the individual with AS' success with a device.
- The use of systems that were non-symbolic for individuals with AS received higher ratings in the themes of usefulness and importance, followed by electronic devices (i.e. aided AAC), and finally enhanced natural gestures (i.e. unaided AAC).
- Across all included studies, all individuals with AS made gains when using AAC—including both aided and unaided AAC; however, outcomes for younger individuals tended to be larger and reported to be more important by caregivers/family members.
- Individuals with AS should be provided with effective, evidence-based AAC supports that are functionally equivalent to their current idiosyncratic means of communicating.
- When providing AAC systems for individuals with AS, access should be quick and reliable; providing meaningful and beneficial outcomes for meeting wants and needs, as well as social closeness.

References

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Contact: Kaitlyn Clarke, KAC529@psu.edu