



RESEARCH REPORT

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RESEARCH SUMMARY

Adaptive Skills of Individuals with Angelman Syndrome Assessed Using the Vineland Adaptive Behavior Scales, 2nd Edition

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WHAT WAS THE RESEARCH ABOUT?

Past research shows that children with Angelman syndrome (AS) have significant delays in adaptive and developmental skills, but relatively little is known about how adaptive skills change over time. Adaptive functioning, which is typically assessed via caregiver report, includes conceptual, social, and practical skills that an individual uses in their daily life (e.g., language, play, dressing, toileting, feeding, motor skills). In this study, we looked at adaptive skills and trajectories over time of children with AS using the comprehensive interview version of the Vineland Adaptive Behavior Scales, 2nd Edition (Vineland-II).

WHAT DID THE RESEARCH TEAM DO?

We analyzed Vineland-II data obtained from caregivers of children between the ages of one and 13 years who were evaluated in the AS

Natural History Study. The Vineland-II was designed to assess adaptive skills and behavior in the domains of communication, socialization, daily living skills, and motor skills, and is commonly used in studies of individuals with AS. Communication was assessed in terms of receptive and expressive skills. The Daily Living Skills domain has three subdomains: Personal, Domestic, and Community. The Socialization subdomains are Interpersonal Relationships, Play and Leisure Time, and Coping Skills. Finally, the Motor Skills domain assesses gross and fine motor skills.

We examined how adaptive skills in these ten adaptive behavior subdomains developed over time in children with AS. We also looked at differences in these adaptive domains among four molecular subtypes of AS (i.e., class I deletion, class II deletion, *UBE3A* mutation, and

Vineland-II

Communication

Daily Living Skills

Socialization

Motor Skills

Receptive

Expressive

Personal

Domestic

Community

Interpersonal Relationships

Play and Leisure Time

Coping Skills

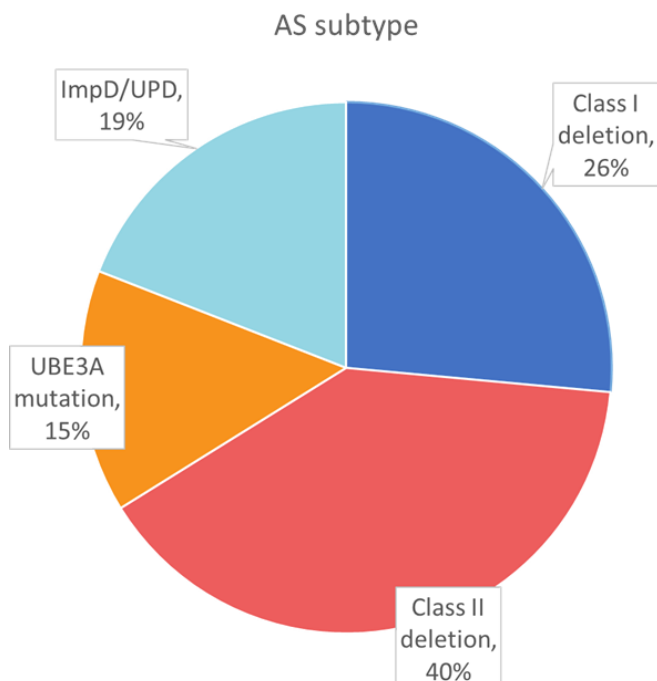
Gross Motor

Fine Motor

ImpD/UPD) and described patterns of strengths and weaknesses for children with AS based on these molecular subtypes. Documenting how adaptive skills vary by molecular subtype is important for future studies that are focused on the needs and prognoses of these children, as well as for future treatment studies that may potentially improve skills in one or more domains.

WHO WAS IN THE STUDY?

Participants were 257 individuals with a molecular diagnosis of AS between the ages of one and 13 years. The average age at the first study visit was 4.3 years. There were an equal number of male and female participants in the sample. On average, children had approximately three study visits.



STUDY PARTICIPANTS

257 participants (12 months - 13 years)

66%
Deletion

34%
Non-deletion

128
Males

129
Females

WHAT DID THE RESEARCH TEAM LEARN?

As expected, caregivers of individuals with non-deletion subtypes (i.e., *UBE3A* mutation and ImpD/UPD) reported higher levels of skills in all subdomains relative to caregivers of individuals with class I and class II deletion subtypes. At age six years, individuals with AS were demonstrating adaptive skills ranging from approximately nine months of age (Expressive Communication subdomain, class I and class II deletion groups) to 38 months of age (Domestic subdomain, ImpD/UPD group). Apart from the Expressive Communication subdomain scores (where scores ranged from approximately nine months in the class I deletion group up to approximately 14 months in the ImpD/UPD group), children in the class I

and class II deletion groups had age equivalent scores between approximately 12 months and 18 months at age six years, whereas children in the ImpD/UPD and *UBE3A* mutation groups had age equivalent scores between approximately 18 months to 38 months at age six years in the other domains. Overall, individuals with AS demonstrated growth over time in each domain, with faster growth early in life that slowed as individuals got older.

Communication

Children had higher scores in the receptive communication subdomain compared to the expressive communication subdomain. At six years of age, age equivalent scores ranged from about 14 to 27 months for receptive skills and nine to 14 months for expressive skills, well below the age equivalent scores from the normed typically developing sample. This gap between the current sample of individuals with AS and the normed typically developing sample became more pronounced with age.

Daily Living Skills

At six years of age, children had scores ranging from 12 to 27 months in the

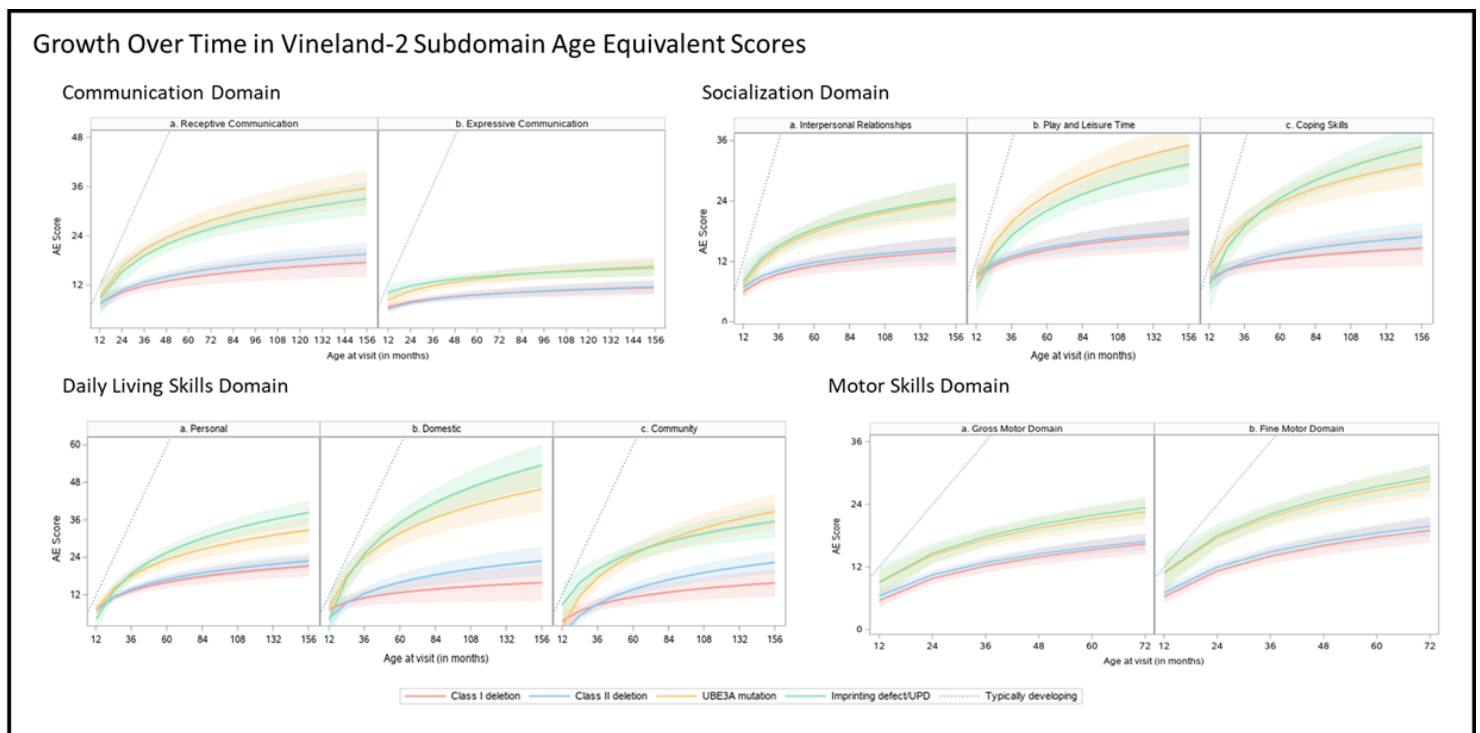
Community subdomain (functioning in the world outside the home, such as safety), 17 to 27 months in the Personal subdomain (skills such as eating, dressing, and hygiene), and 14 to 38 months in the Domestic subdomain (household tasks such as cleaning up and chores).

Socialization

At six years of age, age equivalent scores in the Interpersonal Relationships subdomain ranged from approximately 11 to 19 months, Coping skills ranged from approximately 13 to 26 months, and Play and Leisure Time skills ranged from 14 to 27 months.

Motor Skills

At three years of age, gross motor skills ranged from about 12 to 18 months, whereas fine motor skills ranged from approximately 14 to 22 months. Consistent with the other domains, those with a deletion subtype had lower age equivalent scores relative to those with a non-deletion subtype. By age six, there were no differences between scores in those with a deletion subtype compared to those with a non-deletion subtype.



WHAT DOES THIS MEAN FOR FAMILIES?

The results of the current study confirmed many previous reports suggesting that **adaptive skills vary by molecular subtype**, with individuals with non-deletion subtypes typically demonstrating a higher level of skills compared to those with deletion subtypes. Importantly, **individuals with AS are demonstrating growth in all adaptive domains through at least early adolescence** indicating that continued services, including speech-language, occupational, and physical therapies, as well as academic modifications, could continue to provide opportunities for sustained development.

Significant deficits in expressive communication in particular highlight the need for speech and language intervention that focuses on various modes of communication, including gestures, signs, and augmentative and alternative communication (AAC) systems, including both low-tech (e.g., picture symbols, recordable buttons) and high-tech (e.g., iPad apps, such as Proloquo2Go and GoTalk Now, and dedicated speech-generating devices) options. Utilizing these options early in the affected individual's development is critical for ensuring that the child can successfully learn the communication system and provides opportunities for communication partners to develop familiarity and proficiency with the AAC system.

Full article by Dr. Gwaltney and colleagues:

[Read here](#)

Gwaltney A, Potter SN, Peters SU, Barbieri-Welge RL, Horowitz LT, Noll LM, Hundley RJ, Bird LM, Tan WH, Sathwani A, Wheeler A. Adaptive Skills of Individuals with Angelman Syndrome Assessed Using the Vineland Adaptive Behavior Scales, 2nd Edition. J Autism Dev Disord. 2023 Aug 15. doi: 10.1007/s10803-023-06090-8. Epub ahead of print. PMID: 37581718.