

# Angelman Syndrome

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Angelman Syndrome is a genetic disorder affecting mental development. AS is believed to affect 1 in 25,000 children. A geneticist or neurologist can make the diagnosis - usually between the ages of three and seven years. About half of AS patients are missing part of chromosome 15. This discovery has aided the diagnosis of AS, helping doctors to identify more AS patients.

## Characteristics of AS

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AS patients develop slowly - particularly in the areas of speech and motor skills. Many AS sufferers cannot speak or will speak less than three words. The majority of AS patients (80%) will show signs of epilepsy or some other type of seizures.

In the early years, AS is also marked by feeding problems and abnormal sleep patterns. AS children are normally happy - in fact, they are often excessively jovial and easily provoked to laughter.

Short attention spans and hyperactivity are common in AS children. They tend to have difficulty concentrating and prefer physical activities. AS children usually enjoy social company, though they tend not to interact directly. Behavior modification can be effective in helping to curb problematic AS traits.

AS patients may have difficulty coughing or blowing their nose. There is a tendency for minor colds or infections to progress to pneumonia or other respiratory infections. AS patients are not overly prone to severe vision problems, but there are a few minor eye problems common to AS patients. AS children often have lighter pigmentation which can lead to squinting or roving eye movements.

The following is a list of common AS traits:

- Mental retardation, severe
- Absent Speech
- Movement or balance disorder (stiffness, jerkiness, unsteadiness, gait ataxia)
- Small head by age 3 years
- Happy, smiling behavior
- Seizures, abnormal EEG (around the age of two years)
- Flattened back of the head
- Protruding tongue with prominent jaw, wide mouth
- Drooling and excessive chewing and oral behaviors
- Small, wide-spaced teeth
- Strabismus (crossed eye)
- Hypopigmented skin; light hair and eye color
- Wide-based gait (feet far apart) with flat, out-turned feet
- Small hands and feet, short stature
- Excessive sweating; heat intolerance
- Feeding problems during infancy
- Uplifted arms (like a puppet)
- Crawling and sitting fail to appear at 6-12 months

## **Treatment**

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AS children can benefit from occupational, physical, and speech therapies as well as medical treatments.

Most medical treatment centers around seizure management. Close to 80% of AS patients suffer from seizures sometime during their lives. Administering medication to an AS patient can present problems in itself. A survey of parents of AS children revealed common difficulties with medication. Seizure-management medication is usually taken orally. If direct administration is unsuccessful, many parents mask the medicine in food somehow - though some parents pointed out this may "ruin" several foods upon discovery.

Occupational therapy can help AS patients develop sensory-motor and fine motor skills. This therapy can help develop skills such as independent eating. Physical therapists can help the child learn to walk or move around. The child may need help learning to shift positions or bear their weight best. Speech therapists would focus on communication skills - oral or alternate methods. Many AS students will not develop oral communication, but many patients are successful with modified forms of sign language. Other students may use pictures or symbols to communicate.

## **Classroom Consideration**

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A survey of AS parents showed Thirty-four percent of the children were in a special education classroom and integrated in regular education classes when appropriate, thirty-one percent were placed in full special education programs, and thirteen percent were fully integrated into regular classrooms. A smaller number of parents said their AS children were home schooled, sent in private schools, or placed in group homes.

AS students have movement, communication, and developmental problems. Depending on the severity of their condition, the classroom will have to be modified accordingly. Mental retardation is a common AS trait - though it does not affect all patients.

In severe cases of AS, the classroom teacher will work to augment the efforts of the student's therapists in an attempt to help the student eventually achieve some level of independence.

## **Sources of Information**

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Angelman Syndrome: A Parent's Guide  
[http://members.aol.com/miller566/as\\_paper.htm](http://members.aol.com/miller566/as_paper.htm)

Angelman Syndrome Information  
<http://shell.idt.net/~julhyman/angel.htm>