



The Arc Answers: Prader-Willi syndrome

How many people have Prader-Willi syndrome?

It is estimated that between 1 in 10,000-25,000 people are born with Prader-Willi syndrome; approximately 17,000-22,000 Americans have Prader-Willi syndrome. It occurs in all races and affects males and females equally.

What causes Prader-Willi syndrome?

Prader-Willi syndrome is caused by one of several possible genetic anomalies on chromosome 15, including the deletion or mutation of genetic material. These are random events that happen during embryonic development.

Angelman syndrome also occurs due to anomalies on chromosome 15. What's the difference?

Prader-Willi syndrome is caused by either a random deletion on the chromosome 15 inherited from the father, or the inheritance of two copies of the maternal chromosome 15 (none from his or her father). Angelman syndrome is caused when the maternal copy of chromosome 15 is either partially or entirely missing.

What are the primary characteristics of Prader-Willi syndrome?

Physical characteristics include: short stature, small hands and feet, low muscle tone, temperature control problems, almond-shaped eyes, narrow face diameter, down-turned mouth, lack of eye coordination, and underdeveloped sex organs in males and females.

Behavioral characteristics include tantrums, stubbornness, mood swings, anxiety, and repetitive skin picking. Perhaps the most recognized trait is the child's relationship with food. Infants may be diagnosed with failure to thrive, because their low muscle tone prevents them from sucking. However, between ages one and six a child develops an uncontrollable appetite due to a malfunction in their brain. He or she feels hungry all of the time, and will sneak, hoard food and overeat if portions are not controlled. These behaviors continue into adulthood. People with Prader-Willi syndrome may have mild to moderate intellectual disabilities, but often display relative strengths in reading, long-term memory, and visual spatial skills.

What are common medical concerns?

Prader-Willi syndrome is the most common known cause of childhood obesity. This risk for obesity continues into adulthood, so medical concerns mainly include complications of obesity: diabetes, heart disease, high blood pressure, depression, skin problems, sleep apnea, stroke, etc. Less commonly, some children with Prader-Willi syndrome develop scoliosis, a curvature of the spine.

What interventions have been proven helpful for people with Prader-Willi syndrome?

Because people with Prader-Willi syndrome have a low metabolism and need less calories, even though they have insatiable hunger, a balanced, low-calorie diet in conjunction with regular weigh-ins and daily exercise are highly recommended. Doctors may recommend growth hormone therapy for some individuals. In addition, people with Prader-Willi syndrome may benefit from an individualized combination of educational supports, speech therapy, occupational therapy, and/or physical therapy.

Where can I find other resources?

Foundation for Prader-Willi Research: <http://fpwr.org/>

National Library of Medicine: <http://www.nlm.nih.gov/medlineplus/praderwillisyndrome.html>

International Prader-Willi Syndrome Organization: <http://www.ipwso.org/>

Prader-Willi Syndrome Association: <http://www.pwsausa.org/>

Resources in Spanish: <http://www.pwsausa.org/Spanish/>

In North Carolina:

Prader-Willi Association of the Carolinas: <http://health.groups.yahoo.com/group/pwsa-carolina/>

UNC-Chapel Hill Prader-Willi syndrome clinic brochure: <http://www.cdl.unc.edu/psychology/pwsbrochure.pdf>

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