What is Prader Willi Syndrome?

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Prader Will Syndrome is a genetic disorder resulting from an abnormality of chromosome 15 such as a loss of active genes. In most cases (70%) the paternal copy is missing and in some cases (25%), will exhibit two maternal copies of Chromosome 15. The genetic disorder was initially described by John Langdon Down and was named after Drs. Andrea Prader, Heimrich Willi and Alexis Labhart in 1956 and is found in 1 in 20,000 births affecting both sexes. It is also the most common recognized genetic form of obesity. During childhood, individuals diagnosed with Prader-Willi Syndrome tend to eat constantly leading to obesity and for some, type 2 diabetes will develop. This complex disorder affects appetite, growth, metabolism, cognitive functioning and behavior.

Signs and Symptoms

People with Prader-Willi Syndrome (PWS) tend to never feel full (hyperphagia) which leads to constant eating. Signs in infants include, problems with strength, coordination and balance. Often there are feeding problems at birth, delayed speech and gross motor development. Children may be born with almond-shaped eyes and undeveloped sexual organs. Cognitive disabilities and developmental delays may also be present.

As children began to grow, constant craving for food often leads to behavior challenges including hoarding food, eating frozen food and food left in the garbage causing controlling or manipulative behavior.

Medical Issues

Medical concerns may include the following:

- Sleep Apnea
- Respiratory/Breathing
- High pain tolerance
- Severe stomach illness
- Difficulty with vomiting reflex
- Excessive appetite
- Binge eating
- Eye problems
- Choking
- Hypothermia
- Leg Swelling
- Consuming unsafe items
- Negative reactions to medications

Teaching Strategies

Most people diagnosed with Prader Willi Syndrome fall between the moderate and mild levels of an intellectual disability meaning there may be challenges in the area of reasoning, problem-solving, planning, judgment, abstract thinking and learning. A child or student functioning at the moderate level may lag behind their peers in the area of language and pre-academic skills. Adults may function at an elementary school level and will require support in both work and daily living skills. For children and students functioning at the mild level, there may be difficulties in the area of reading, writing, math and money management. as children grow into adults, there may be a need for support in abstract thinking, executive functioning (planning, prioritizing and flexibility) as well as short-term memory and money management. Teaching strategies should focus on the following:

- Aggression management
- Anger management skills
- Anxiety management
- Emotional regulation
- Personal safety
- Social skills

Keep in mind that many children and adults diagnosed with Prader-Willi Syndrome may have additional challenges in learning due to medication. Some people take medication such as a growth hormone therapy which can cause fatigue. The following teaching strategies may also be useful when teaching a student diagnosed with Prader Willi Syndrome:

- Use a multi-sensory approach. This involves a teaching style that includes auditory, visual, tactile, spatial, and kinesthetic (hands on activities)
- Break learning into small steps. Check for understanding by asking the student to repeat back to you.
- Teach a skill at least 2-3 times a day. This will help the student retain information.
- Managing perseveration. Set up a rule where the student can a question no more than 3 times. After the third answer. Ask the student to repeat the response back to you.

Adult Day Program/Residential Setting

Most people with Prader Willi Syndrome due to their cognitive level, will be provided services in either a day habilitation program or live in a community providing residential services. Once a person becomes an adult, it becomes a little bit tricky on maintaining issues especially behavioral. For instance, while living at home, a parent has the right to lock the refrigerator which is often suggested by experts. However, this becomes a violations of a person's rights once they reach adulthood. Typically, committees meet to

help make the right decisions along with family members and the adult diagnosed with Prader Willi Syndrome. Here are some suggestions.

- Allow the person to have control of what is important to them. Have discussions on nutrition and staying healthy. Check to see if this may be an appropriate topic the person may want to improve by adding to their person-centered plan. Hold discussion groups in both day programs as well as in residential to discuss various topics on health and nutrition including holding classes on mindfulness and meditation.
- Trips to shopping malls can be very tricky. Try to avoid mall's eatery and plan if it is a group trip to have people bring their own lunches.
- When teaching, allow time before giving additional prompts
- Give praise as much as you can when it is appropriate.
- Use visuals as much as you can including graphics and pictures.

Staff Training

Staff training on Prader-Willi Syndrome should include the following topics:

- Overview of Prader-Willi Syndrome including, causes, symptoms, characteristics, nutrition, and self-regulation.
- Impact on the family including the stresses families experience.
- Teaching techniques including problem-solving, forward shaping and role-modeling.
- Individual rights
- Managing behavior and crisis intervention
- Community inclusion trips and activities

Resources
Foundation for Prader-Willi Research
Prader-Willi Syndrome Association (USA)
Prader-Willi Syndrome (Mayo Clinic)
Reference
Prader-Willi Syndrome Association