

## Prader-Willi Syndrome

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Prader-Willi syndrome is a genetic disorder which is caused by the lack of a contributing chromosome from the paternal inheritance.

Prader-Willi syndrome is characterised by hyperphagia (overeating) and is the leading known genetic cause of obesity.

There are two distinctive stages in early childhood development of children with Prader-Willi syndrome. The first is evident in the child's difficulty in sucking and failure to thrive during infancy and even before birth, infants present decreased fetal movements, abnormal fetal position and difficulty at the time of delivery. A dramatic change occurs during the 1-6 age period which preludes a lifelong pattern of overeating and thriving "too well."

Characteristic facial features may include a narrow face, almond shaped eyes, narrow nasal bridge, down-turned mouth with a thin upper lip. While subtle at birth these features may become more pronounced over time.

Many individuals who have Prader Willi syndrome also display a disposition for solving jigsaw puzzles.

Increased risks for developing compulsive behaviours have been noted in individuals with Prader—Willi syndrome in addition to other maladaptive features.

Individuals with Prader-Willi syndrome will be presented with unique challenges related to food management, choice making and self-determination.

(Dykens, Hodapp, Finucane: 2000:1)

### **Children may exhibit the following developmental characteristics:**

#### **Social and Emotional Development**

- May have extreme difficulties in controlling behaviours
- May be impulsive, disruptive and aggressive

### Physical Development

- Failure to thrive during infancy— difficulty sucking
- Poor weight gain during infancy
- Delayed motor milestones—average age for sitting 12 months & walking 24 months (Dykens, Hodapp, Finucane: 2000:174)
- Infantile lethargy—decreased arousal and weak cry
- Around 3-4 years children begin to overeat
- Food preoccupations common
- Low muscle tone—lack motivation to exercise
- Reproductive organs develop insufficiently
- Short stature
- Heart a & circulation problems that result from obesity
- Sleep disturbances & excessive daytime sleepiness
- Hypo pigmentation
- High pain threshold
- Temperature regulation problems
- Thick saliva which contributes to both tooth decay and articulation problems
- Joint problems

### Language Development/Communication

- No distinctive language profile in Prader-Willi syndrome
- Articulation problems due to thick saliva

### Cognitive Development

- Intellectual impairment varying in severity
- Interference from food related behaviour problems that interfere with cognitive development
- School age children sometimes show strengths in reading and weakness in arithmetic
- Strengths in visual processing as opposed to auditory processing
- Long term memory, spatial organisation, attention to visual detail and visual motor development are well developed
- Weakness evident in short term memory tasks

### Prader -Willi Syndrome Inclusion Strategies

Each child diagnosed with **Prader-Willi Syndrome** will be different and individual. It is important to gain information from the parents as to what characteristics of **Prader-Willi Syndrome** their child displays. It is important to work closely with the parents as well as any additional support specialists e.g. therapists who may be involved with the child. It is also important to gain an understanding from the parent as to what is the most important aspect of their child attending your service. What is it that parents hope to gain from using your service? The following inclusion strategies are just some examples which may be applied to support the inclusion process. This list is only the start and it is dependant on a variety of factors such as environment, length of time child is in care, child's interest, likes, dislikes and skills already achieved. The strategies are divided into developmental areas however some strategies overlap and assist in a variety of developmental areas.

#### **Social and Emotional Development**

- Recognise children may have difficulties controlling behaviour.
- Establish an environment that promotes emotional stability (remove things from the environment that may aggravate obsessive/compulsive behaviours).
- Provide opportunities for positive social interactions.
- Model appropriate behaviours.

#### **Physical Development**

- Encourage children to engage in physical activity.
- Activities that encourage children to run, jump, balance etc such as obstacle courses and playground challenges may be beneficial.

#### **Language Development/Communication**

- Provide opportunities for children to engage in activities to enhance language and articulation e.g. small group show and tell, making different noises, repetitive songs.

#### **Cognitive Development**

- Focus on some of the child's displayed cognitive strengths e.g. puzzles.
- Provide visual and perceptual clues for tasks (pictures or models).
- Provide opportunities for children to practice short-term memory tasks.
- Consider the impact that the physical environment may place on children's ability to learn and function.

### Reference

- Umansky, W. and Hooper, S. (1998) *Young Children with Special Needs* Third Edition New Jersey, USA:Prentice-Hall
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- Brannnelly K. "Ideas for Inclusion & Programming Tips" Carata 2000

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