How you can help ...

TAKE THE TIME TO UNDERSTAND THE CHALLENGES OF THIS COMPLEX SYNDROME

Follow dietary plans in place and be guided by people living with PWS, their parents or carers in keeping anxiety low and responding to behaviours.

Learn more about PWS via our website. Help us to share information about PWS – awareness leads to understanding and acceptance.

Membership of PWSA(NZ) is free (with a subscription option available for organisations). Please see our website for membership options.

People living with PWS are friendly, sociable, caring and unique - please take the time to get to know and love them.

Most services offered to people living with PWS and their families are provided free of charge. We value donations that help us support families and we also fundraise to help eliminate the challenges of PWS through research. Donations to:

PWSA: **www.givealittle.co.nz/org/pwsasupport**

Research: **www.givealittle.co.nz/org/pwsresearch**

How we can help ...

THE PWSA(NZ) SUPPORTS PEOPLE LIVING WITH PWS, THEIR FAMILIES & CAREGIVERS

ADVOCACY

- Providing advocacy services on behalf of people living with PWS and campaigning on relevant issues.

EDUCATION

- Accessible information via our website and our range of free publications and resources
- Training workshops offered to schools and residential care providers
- Increasing awareness and understanding of PWS
- Circulating research to medical professionals as needed
- Keeping members informed of the latest news, ideas and research through our newsletters
- Links to the international PWS community

SUPPORT

- Offering information, diagnosis and crisis support to people living with PWS, parents, whānau and caregivers
- Facilitating connection via our parent support network
- Hosting events, family meetings and support weekends

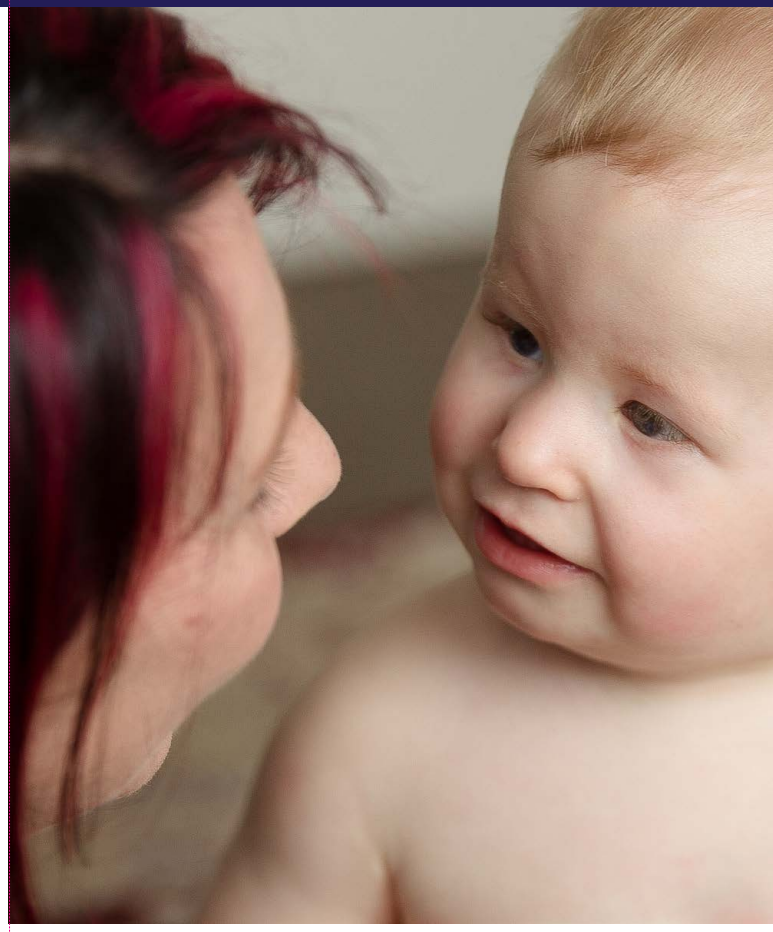
PLEASE CONTACT US

Prader-Willi Syndrome Association (NZ) Incorporated

PWS
PRADER-WILLI SYNDROME
ASSOCIATION NEW ZEALAND

0800 4 PWS HELP
(0800 4 797 4357)
enquiries@pws.org.nz
www.pws.org.nz

An introduction to Prader-Willi Syndrome for Whānau, Friends & Community



ADVOCACY | EDUCATION | SUPPORT

PWS PRADER-WILLI SYNDROME ASSOCIATION NEW ZEALAND

What is Prader-Willi Syndrome?

PWS is a rare and complex neurodevelopmental disorder resulting from an abnormality on the 15th chromosome. First described by Swiss doctors Prader, Labhart and Willi in 1956, PWS randomly occurs in approximately 1 in every 16,000 births.

PWS is associated with a wide range of symptoms, including impacts on cognition, emotional regulation, growth, muscle development, metabolism and appetite. Many symptoms are thought to be caused by dysfunction of the hypothalamus in the brain.

CHARACTERISTICS

The hallmark features of PWS include:

- failure to thrive as infants
- hypotonia (low muscle tone) } GHT improves body composition and normalises height
- slow growth rate
- some subtle, but distinct facial characteristics
- sleep disorders and excessive sleepiness
- delays in motor development and speech, with possible ongoing motor planning, balance, coordination or articulation difficulties
- hyperphagia - increased feelings of hunger, preoccupation with food and an enhanced sense of food reward
- specific learning disabilities, often mild intellectual disability
- behavioural challenges - anxiety, rigidity and emotional control
- hypogonadism causing incomplete puberty & usually infertility

Not every person living with PWS will be equally affected, and as PWS is a spectrum disorder, the presentation and severity of each symptom may vary. The presentation and severity of a symptom can also change during an individual's lifetime.

CHILDREN AND PWS

Infants are typically small, floppy, have a weak cry and usually require assisted feeding for the first few weeks or months. Parents may initially struggle to ensure adequate calorific intake for growth. Reflux and aspiration may cause additional difficulties. Children display global developmental delay but do achieve normal milestones in time.

Growth hormone therapy (GHT) is available for all children with PWS and in addition to building strength, it can help to improve motor development, speech and cognitive skills. Input from speech therapists, physio and occupational therapists, neuro-developmental therapists and orthotists can all be of benefit.

COGNITION AND EDUCATION

Most children have mild intellectual disabilities and can thrive in a mainstream school environment with appropriate supports. Abilities vary, with relative strengths often including good long-term memory skills, visual processing, reading accuracy, using IT, and determination.

People living with PWS are often attentive listeners but many experience auditory processing difficulties - allow extra processing time, give brief, clear instructions, and check understanding. PWS is associated with poor short-term memory and impaired executive functioning skills, particularly in the areas of working memory, flexible thinking and task switching. Repetition, breaking down tasks and the use of organisational aids can help. Abstract concepts can be difficult to grasp, but concrete examples and systematic approaches make learning easier. Other difficulties might include gross and fine motor skills, visual-motor integration, sensory processing and expressive language. People living with PWS are usually very sociable, but poor social skills can hinder ability to connect with peers. As children grow older, the social, emotional and ability gaps widen, and support is required to help navigate adolescence.

HEALTH ISSUES

The hypothalamus plays a role in regulating body functions such as thirst, hunger, the sleep/wake cycle, temperature, mood and the feeling of pain. An inability to regulate body temperature, a high pain threshold and a reduced vomit reflex, can make illness hard to detect or evaluate. The altered appetite and common digestive issues increase risk for choking or serious gastric illness, particularly if a binge eating episode occurs. Any abdominal pain, distension or vomiting should be evaluated.

People living with PWS have increased risk for developing mood disorders, particularly during adolescence and early adulthood. Other health issues may include scoliosis, osteoporosis, bruising easily, sticky, thick and reduced saliva contributing to dental problems, vision abnormalities, and self-injury (skin picking etc). PWS is the most common genetic cause of obesity, but with the advantages of early diagnosis and growth hormone treatment, in conjunction with appropriate diet and exercise, people living with PWS can remain slim and healthy.

BEHAVIOURAL MANAGEMENT

Increased anxiety is central to many behavioural challenges and can lead to perseveration (being stuck on a thought, question or behaviour), rigidity in thinking, stubbornness or temper outbursts. Sudden change, difficulties with interpreting the environment or understanding expectation can all raise anxiety.

People living with PWS can be extremely stress sensitive and 'hyper-reactive'. Many people who have PWS display behaviours on the autistic spectrum. Minimising anxiety and reducing stressors are key to managing behaviours. Forewarn of change, reassure to avoid uncertainties, check understanding and remember to use the 4Cs:

Be CALM, CONSISTENT, CLEAR + avoid CONFRONTATION

Remember that oppositional behaviour is often a 'flight or fight' stress response – don't 'give in' but provide empathy and extra time to process choices that allow a sense of control. Develop strategies for coping with stress, but if a person experiences a loss of control, remember that they are unable to self-monitor, avoid reasoning and reflect when calm. Behaviour fluctuates and is more likely to be challenging during adolescence and early adulthood.

DIETARY MANAGEMENT

Due to the altered body composition (high fat to muscle ratio) and abnormal appetite, weight gain can rapidly occur which is very difficult to lose, compounded by a diminished ability to exercise. Reduced energy expenditure means that calorific intake needs to be restricted (often 20-40%) from when weight gain begins (between 18 months - 3 years). Strict dietary control is required - a reduced carbohydrate diet and avoiding sweet foods are recommended.

Food has intensified significance from around 4.5 years old, with increased hunger and lack of satiety possible from around 8 years old. People living with PWS can be preoccupied with food and food seeking. This obsession with food is at the heart of many behavioural problems. Remove temptation by preventing access to food, whilst also reducing anxiety through the provision of clear boundaries about when and what food will be available. This method works well and allows a person living with PWS to focus on other things:

FOOD SECURITY = NO DOUBT + NO HOPE (or chance) = NO DISAPPOINTMENT (Pittsburgh Partnership, USA)

Please check with parents or carers before offering food.