

PRADER-WILLI SYNDROME

TEACHER PACK

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DISCLAIMER: While the information in this pack is believed to be accurate at the time of publication, it is not intended to be a substitute for seeking professional advice.

A BRIEF OVERVIEW OF PWS

GENETICS

PWS was first described in 1956 by Swiss Professors Prader, Labhart and Willi. It was called Prader-Labhart-Willi syndrome, but is usually known as Prader-Willi syndrome. It occurs randomly in approximately 1 in 16:000 births, equally affecting males, females and all ethnicities. It was not until 1981 that a genetic cause was discovered.

Scientists do not know why PWS randomly occurs at conception. A segment of genes in chromosome 15 are either deleted or unexpressed and cause the characteristics of PWS. There are 3 main different ways in which these genes are prevented from working: Chromosomal deletion (approx. 70% of cases), Uniparental Disomy (UPD) (approx. 30% of cases) and Imprinting Defect (approx 1-3% of cases.) Differences between these genetic subtypes are subtle and there is no feature that is exclusively found in one group.

MAIN CHARACTERISTICS

The main physical characteristics which may have led doctors to test for PWS are:

- Hypotonia (low muscle tone) – floppy as newborns with poor suck reflex, weak cry
- Distinct facial features – almond shape eyes, narrowing of the head at the temples, a turned down mouth and thin upper lip (more likely in those with a deletion)
- Fair colouring compared to other members of the family (also more likely in those with a deletion)
- Hypogonadism (underdeveloped sex organs)
- Excessive sleepiness and showing little response to stimulation
- Small hands and feet
- 'Failure to thrive' – low birth weight followed by slow weight gain / growth
- Possible lack of eye coordination (strabismus / squint)
- Possible congenital hip dislocation

The main characteristics of PWS which become evident in older children are:

- Delayed motor and speech development
- Hyperphagia (increased appetite) and preoccupation with food
- Short stature – with the availability of growth hormone treatment today, natural expected height can be reached (in addition to an improvement in muscle tone)
- Borderline / mild intellectual disability is most common, but some children score in the moderate intellectual disability range, whereas others score within the borderline to low-average ranges of normal intelligence in IQ tests
- Specific learning disabilities
- Behaviour problems linked to anxiety, rigidity in thinking and emotional control
- Incomplete puberty and infertility (hormone replacement treatments, such as testosterone, may be given to help develop the characteristics of puberty)

It is important to note that PWS is a complex genetic disorder with a spectrum of symptoms. A child can have some or all symptoms, each in varying degrees.

Most of the symptoms associated with PWS are likely to be caused by hypothalamic dysfunction. The hypothalamus is a small endocrine organ within the brain which links the endocrine and nervous systems via the pituitary gland. It is responsible for producing many of the body's hormones and controls endocrine functions such as metabolism and growth. It also plays a role in governing functions such as temperature regulation, thirst, hunger, the sleep/wake cycle, mood, the expression of emotions and the feeling of pain.

EARLY DEVELOPMENT

Most infants with PWS are initially fed via nasogastric tube. They often sleep a lot and have to be woken for feeding and stimulation. An infant with PWS can be slow to reach physical milestones such as rolling, sitting, crawling and walking, with the delay in reaching these varying greatly in individuals.

An early intervention team will work with the family to develop a programme that helps develop motor skills, motor planning, balance, strength, sensory integration processes and early speech. The team may include physiotherapists, occupational therapists, neuro-developmental therapists and speech and language therapists. Children with PWS may also have lax joints and need supportive insoles or boots from an orthotist.

Language is often delayed in PWS. A speech and language therapist should offer advice on modelling speech and encouraging communication. Many families use a simple form of sign language to supplement communication until words are more comprehensible. Delayed speech is mainly associated with hypotonia, but articulation problems can be ongoing. Dry mouth (due to insufficient saliva production), hypernasality and pitch variations can also interfere with clear speech. Symptoms of the speech disorder Apraxia of Speech are often present and may require separate evaluation.

HEALTH ISSUES

Once respiratory strength is gained, children with PWS can remain slim and healthy with growth hormone treatment and consistent dietary management strategies. However, there is an increased risk for diabetes which increases with weight gain. There may also be a reduced ability to fight illness, so the free influenza vaccination is recommended.

Temperature regulation and pain tolerance are affected by the disorder of the hypothalamus, making illness hard to detect. Normal body temperature may be low so a slight increase raises concern, or a sick child may not present with fever at all. Temperature instability also causes children to feel too hot or cold and wear inappropriate clothing. A high pain threshold can lead to a serious illness, infection or broken limb going undetected.

Poor vomit reflex is often observed and adds to difficulties in detecting illness. As vomiting is a self-defence mechanism, a weak vomiting reflex can be cause for concern at times. Conversely, vomiting in a child who rarely or never vomits can also be cause for concern.

Vision problems commonly include constant or intermittent strabismus (squint). Typically, one eye may turn inwards. An ophthalmologist will treat promptly if needed as the visual system develops rapidly in children. Strabismus may only require monitoring because it sometimes gets better as muscle tone improves, or it may only be present when tired. Other vision problems in PWS involve focusing difficulties, long sightedness (highly prevalent but usually mild) and short sightedness (fairly common by teenage years).

Abnormal sleep patterns occur because sleep cycles are controlled by the hypothalamus. Some children have sleep problems, the most common being night waking and EDS (Excessive Daytime Sleepiness). EDS may be improved by changes in diet, using dietary supplements or prescribed medication (in severe cases). Sleep studies monitor sleep patterns for efficiency and check for apnoea or respiratory issues. Narcoleptic-like symptoms are typical, but occasionally there may be a diagnosis of narcolepsy or cataplexy.

Hyperphagia (abnormally increased appetite) varies in age of onset and intensity. A lack of normal satiety cues puts people at risk of dangerously overeating, leading to stomach necrosis or rupture. A preoccupation with food usually exists, but significantly reduced food intake is vital to prevent rapid weight gain due to the altered body composition (low tone).

Gastroparesis is delayed stomach emptying and it is thought to be fairly likely that a significant number of people with PWS have this to some degree. Gastroparesis becomes a problem when the volume of accumulated food in the stomach becomes too much - watch for an overly distended stomach, nausea, vomiting or pain. The bowel intestinal tract can also empty too slowly causing constipation and retained stool in the colon.

Bruising easily is typical in PWS, although the exact cause of this is unknown.

Scoliosis is when curvature of the spine develops. Prevalence estimates vary, but range from 30% in children, increasing with age. Early detection is important and may reduce need for surgery. With a mild curve, physio exercises will be required to help control the curve. An orthopaedic spine surgeon may also treat with bracing or casting.

Hypothyroidism occurs in a small number of children and will be tested. It is easily treated.

CAI (Central Adrenal Insufficiency) causes ineffective cortisol responses at times of stress (illness). Although prevalence is unknown, it is thought to be fairly low. The adrenal glands stress response to a pituitary hormone called ACTH will be routinely tested.

EDUCATIONAL SETTINGS AND FUNDING

Nearly all children with PWS are able to attend mainstream primary schools. Some pupils go on to attend mainstream secondary schools, but as peer gaps widen, sometimes a special unit within the college is more appropriate for part or all of the time. A few children attend special schools and several pupils have benefitted from a temporary enrolment at the residential special schools of Salisbury or Halswell.

Pupils with PWS require some level of support. In early childhood education, an Education Support Worker (ESW) is needed and an Early Intervention Teacher will apply for this support prior to a pupil starting at a centre. During the school years, Teacher Aide support is essential, with additional support required whilst transitioning. If adequately supported, pupils with PWS can thrive in the school environment. They usually have a passion for learning and enjoy responsibility.

TRANSITION TO SCHOOL

Due to initial developmental delay, some parents decide to delay school starting age. Alternatively, other families choose the option of initial part-time attendance. Pupils may tire easily at first and require frequent rest breaks. Sometimes it can be of benefit to reclassify a pupil, allowing them to repeat a school year early on.

Prior to starting school, additional visits are recommended. It is important that the class teacher and other staff are knowledgeable about the pupil and about PWS to enable a smooth transition. Adopting a whole school approach to managing the underlying anxiety and stress sensitivity in PWS is recommended because pupils with PWS prefer consistency. The PWSA(NZ) is able to visit during a staff meeting to facilitate training if required.

TRANSITION TO SECONDARY SCHOOL

Transition to secondary school is a significant change that pupils with PWS may find more challenging and requires careful consideration and planning.

The larger school site and timetabled classroom changes will create organisational and physical challenges. Pupils with PWS tend to move more slowly and may have balance and motor planning difficulties, so they may take longer than others to move from one classroom to another. They also have difficulty in task switching and may find it hard to quickly pack up, move on and start something new when a bell rings. The impairment of executive functioning skills also means they may need help with planning and organising ahead what they might need. There may also be sensory challenges to overcome – many children with PWS do not like crowded or noisy spaces and may prefer to wait until corridors have cleared.

This larger school environment also means an increase in possible food sources. In addition to increased health risks, the potential availability of food will cause additional anxiety for the pupil and their thoughts may become focused on this. Potential food sources will need to be identified as part of a risk management plan. (Please see the section on managing the food environment.)

When they leave primary school, the pupil also loses the security of a single classroom teacher who knew and understood them well. Suddenly they have multiple new teachers, each with their own teaching style and different expectations which can be very confusing. Good pupil / staff relationships and a consistent approach to staff interactions with the pupil are essential for success, so it is important that home / school collaborations that build understanding and establish relationships start early during transition and are ongoing.

Another consideration is that peer groups who had perhaps been together since starting school will also be dispersed when moving to secondary school and these pupils may have grasped a good understanding of PWS over the years and been part of a support system for the pupil. Due to social skill difficulties, pupils with PWS are likely to find it hard to form new friendships and differences will become more apparent as pupils become teenagers and the social gap widens. Their experiences outside of school are also likely to differ because pupils with PWS are unable to have the same level of independence as their peers.

The adolescent years themselves bring about additional struggles for pupils with PWS, their families and teachers. As hormonal changes occur, it is quite typical for challenging behaviour to become more intense and also more frequent if they are experiencing greater stress. Intense behavioural episodes may increase into early adulthood. It is also typical for food seeking desire and / or ability to increase during adolescence. Puberty itself can be a sensitive issue for pupils with PWS because they do not develop typically and will be aware of these differences (please see our website for details.) Pupils with PWS are also likely to be aware of their peers starting relationships and may want to do the same, but additional guidance will be required for understanding appropriate friendly and affectionate behaviours and relationship etiquette. A somewhat naive approach to relationships can also make individuals with PWS vulnerable to bullying or abuse. Furthermore, there is an increased susceptibility for mental health problems in PWS during adolescence, particularly amongst those who have PWS by Uniparental Disomy.

TRANSITION FROM SCHOOL

This process can start from age 14 and pupils who have ORS funding will be able to access a funded MSD Transition Service for one year prior to leaving school (after age 16). Non-ORS funded pupils may still be able to access a service but there are limited places and possible waitlists. It is important that the specific needs of pupils with PWS are considered in the transition process, in particular, the openness of programme providers to training and

management of the food environment. Thorough compatibility checks need to be made so that the pupil does not end up in a no school and no placement situation if transition doesn't go well. Opportunities for social skill training through school years and during transition can help pupils cope with different social situations and solve problems at school, but they are also very helpful for preparing pupils for real life situations in the future.

FUNDING

Approximately 60% of pupils with PWS in New Zealand currently qualify for ORS funding. These pupils mainly receive funding in the high needs category under criteria 5, 8 or 9. Of the pupils who do not receive ORS funding, some are able to access School High Health Needs Funding (SHHNF). An application for SHHNF is often successful due to the need to keep a pupil safe around food, but sometimes more Teacher Aide hours are obtained via SHHNF than the ORS fund, although this fund is not designed to provide learning support. SHHNF also has other limitations in that it is reviewed annually, pupil need is expected to diminish with age and it does not extend beyond 19 years, as ORS currently does.

Some tips for SHHNF applications are to include the following health needs:

- fatigue and the need for rest breaks to re-engage/focus or avoid sensory overload
- poor balance, coordination and motor skills creating a need for safety monitoring in crowds, on stairs, with tools etc.
- hypotonia and the need for physio / OT exercises to develop strength / motor skills
- hyperphagia and the need for supervision around food
- lack of thirst awareness and the need for supervision of water intake
- repetitive self-harming / habit behaviours and the need for monitoring, prevention strategies and ensuring of hygiene
- anxiety and the need to monitor, use support strategies and reassure
- poor temperature regulation and the need to ensure appropriate clothing is worn
- the need for support with toileting (for younger pupils)

ABILITIES AND LEARNING

As with all children, pupils with PWS are individuals whose abilities will vary. This section only serves as a guide to observed patterns of learning strengths and weaknesses in PWS.

Most pupils with PWS will have some degree of intellectual disability. Cognitive test scores mostly fall in the mild to moderate intellectual disability range, but some individuals have scores in the borderline to low average range, with a range of learning disabilities still present. Learning strengths and weaknesses may not concur with overall cognitive ability as pupils often present with islands of competence as well as specific learning difficulties.

Social and emotional immaturity also impacts on learning and performance in addition to the many links to ASD in PWS. (The majority of pupils will present with persistent habits, a preference for predictability, rigid thinking and particular ways of doing things, compulsive tendencies, social-communication impairments, sensory issues and anxiety.) A recent study found 26.7% of participants met the full criteria for a dual diagnosis of PWS and ASD.

	STRENGTHS	WEAKNESSES
Learning styles	Pupils with PWS tend to be visual learners . They have a tendency to do well in visual tasks such as wordsearches and jigsaw puzzles (using a visual matching approach which uses memory of static features rather than a spatial approach of rotating pieces.) Pictures, videos, diagrams and demonstrations aid learning. Pupils learn better by doing than observation.	Visual-motor integration skills (Pupils were found to score higher in separate tests for visual perception and motor coordination than in the combined test. Separation of tasks into pure visual or pure motor improved learning.) Difficulties in reproducing a sequence by observation.
Memory	Good long term memory - if information can be transferred from short term into long term memory by repetition, recall will be good.	Poor short term memory skills. Poor auditory memory - difficulty following verbal instructions, especially if too many. Visual cues / aids are helpful. Poor working memory – notably multi-tasking
Language	Receptive language is usually better than expressive language although there are often language processing difficulties.	Expressive language is often delayed and difficulties with speech can continue. Speech problems may include Apraxia of speech (difficulty sequencing motor movements to execute speech) and articulation difficulties related to hypotonia. Communication difficulties lead to frustration. Poor auditory processing including difficulty with non-literal meanings, idioms, detecting sarcasm and inferences. Using clear, specific language aids understanding. Need extra time.
Reading	Reading accuracy - visual and long term memory skills aid the learning of sight words.	Comprehension skills involving inference and deduction.
Writing	Spelling can be learned using long term memory and visual skills. Non-narrative writing following a set format.	Weak hand strength and impaired motor skills impact on handwriting and typing ability . Creative writing is improved when using personal experience, idea webs, word banks etc

Maths	Learn by seeing concrete examples and using clear systematic methods.	Difficulties with abstract mathematical concepts and multi-step problems .
IT skills	Using computers and technology.	
Physical skills	Fine motor skills tend to be better than gross motor skills although weak hand strength and motor planning ability can cause difficulties with using scissors and drawing. Fine motor skills can be improved with practise and by activities such as beading, using dough or sand.	Gross motor skills such as catching, throwing, jumping, climbing. Sensorimotor balance issue. Symptoms of dyspraxia are common; some pupils may have a dual diagnosis. Reduced stamina due to low muscle tone – pupils often prefer to be sedentary so extra praise and motivation are required because exercise builds necessary muscle strength.
Thinking skills	Visual processing – visual aids benefit learning. Generally, pupils are concrete thinkers who learn by experiencing discrete objects, seeing concrete examples and who like to use a systematic approach to problem solving using specific 'how to' steps.	Auditory processing – delay (3-5 seconds) Sequential processing deficit – difficulties with order, timing, reasoning if...then..., multi-step tasks, summarising and restating events. May need extra time for tests / tasks. Advance notice of questions can allow processing time. Abstract thinking – difficulties with reasoning, abstract concepts, summarising, generalising or transferring learning from one environment to another. Rigid 'black and white' thinking – once something is learned, that can be the only 'right' way and it is hard to relearn. A tendency to keep to literal meanings and difficulty interpreting ambiguous content.
Social skills	Friendly, outgoing, sociable and caring. Although they may not appear to, pupils usually value relationships and have a real need to connect with others. Like to be helpful.	Young children tend to parallel play rather than engage in play with others. Difficulties with cooperative play and sharing. Pupils often find group work difficult. Poor conversational skills such as interrupting, not standing an appropriate distance, speaking too loudly and poor listening. (<i>APD / working memory / speech.</i>) Interactions with peers may be minimal so learning to identify the emotions of others and role playing situations can help.

EXECUTIVE FUNCTIONING

An area of weakness not mentioned above is Executive Functioning. This is an umbrella term for the management of cognitive processes including working memory, mental flexibility, problem solving and self-control. Studies have found that PWS is associated with a global impairment of EF, with particular difficulties in the areas of task switching and cognitive estimation. Pupils with PWS may also have difficulties with the planning and execution of tasks, attention span, resisting impulses, shifting thoughts and adapting ideas. Executive Functioning can be improved with:

- A step-by-step approach to work and tasks broken down into chunks
- Visual organisational aids, visual schedules, checklists and tools for organising time
- Instructions in multiple formats
- Planning for transition times and a change in activities

TEACHER TIPS FOR TEACHING AND LEARNING

- ✓ Support strengths in visual learning with the use of **visual aids**.
- ✓ **Repetition of learning** is necessary due to poor short term memory.
- ✓ Provide **brief and clear verbal instructions**.
- ✓ **Check tasks are understood** by asking them to repeat an instruction back.
- ✓ Use **clear, unambiguous language** and **allow additional processing time** (3-5 secs).
- ✓ Explain and **teach non-literal meanings** (including idioms, metaphors, slang) to aid linguistic understanding and increase social competence.
- ✓ Give advance warning of questions where possible. Allow extra time for tests.
- ✓ **Break tasks into smaller parts**. Consider multi-tasking difficulties, i.e. note taking.
- ✓ Allow pupils to **achieve in small steps** - progressing slowly avoids anxiety, upset and refusal when tasks become too hard.
- ✓ **Use writing frames** / graphic organisers and language supports.
- ✓ Use **real-life or hands-on, concrete examples** to aid understanding.
- ✓ Multi-step problems can be overwhelming - **repeat systematic approaches** to problem solving.
- ✓ Use **computers as a learning tool** or to demonstrate new concepts. **Use assistive technology** when beneficial, i.e. speech to text for handwriting / typing difficulty.
- ✓ **Develop organisational skills** with the use of schedules, checklists, timers etc.
- ✓ Understand that pupils with PWS **tire easily** and may be more receptive to new learning earlier in the day.
- ✓ Provide **changes in activity levels** and rest breaks. **Minimise distraction**.
- ✓ Discuss **whether homework is appropriate** with parents. The pupil may wish to complete homework on a voluntary basis at primary school.
- ✓ Avoid setting lengthy assignments – **short assignments** are best or longer assignments separated into manageable parts.
- ✓ **Adapt the PE curriculum** to focus on developing gross motor skills and building strength. Praise efforts.
- ✓ Consider that **group work can be difficult** for pupils with PWS. Small groups work best with verbal cues and guidance to promote appropriate interactions.
- ✓ **Teach social skills** through role-play and social stories.
- ✓ Encourage pupils to participate in **activities that foster creative play and social connection**. Encourage them to **create and maintain supportive relationships**.
- ✓ Support clarity of speech with the **practise of pronunciation for difficult new words** (multi-syllabic words are often the most tricky) and provide **extra time for communicating needs or speaking in class**.

BEHAVIOUR MANAGEMENT

It has been found that individuals with PWS do not learn from negative behavioural consequences or respond well to consequences that might work well for other pupils. However, they do respond well to positive behaviour management.

BEHAVIOUR	MANAGEMENT STRATEGIES
<p><u>High preference for routine and predictability</u> is typical and therefore pupils often thrive in the school environment where activities are usually timetabled. However, their need for consistency also means that they may not cope well with changes in expectation (theirs or yours).</p>	<ul style="list-style-type: none"> • Clear visual schedules and outlines of expectation are helpful and reassuring. • Provide forewarning of possible changes; avoid sudden changes and rushing. If sudden change can't be helped, use 'spin' giving a positive first before advising of change. • Allow time to process and accept changes. • Avoid making promises that can be broken. • Try to avoid vague, ambiguous answers such as 'perhaps' or 'we'll see' which increase anxiety. • Avoid open-ended questions or too much choice. • Reward flexibility.
<p><u>Task switching</u> is a common area of difficulty with pupils feeling compelled to complete tasks. Support is required during task switching to reduce anxiety which may manifest in frustration.</p>	<ul style="list-style-type: none"> • Pupils may respond well to rituals and visual cues or timetables. • Use verbal and visual countdowns. • Modify tasks so that there is less to complete. • Timetable activities that are typically difficult to stop before a motivating activity, i.e. lunch. • Use incentives but hurrying a pupil can make matters worse.
<p><u>Obsessive thinking or perseveration</u> Pupils with PWS can think or talk obsessively about a topic of interest. Their thoughts can become stuck on a subject or issue. Perseveration or repetitive questioning can be a sign of stress.</p> <p>It may be useful at times to channel obsessive behaviour into positive activities.</p>	<ul style="list-style-type: none"> • Bring a topic to a resolution. • Set limits for the amount of time they can talk on a topic. • Do not provide more information than is necessary and avoid grey areas which can raise anxiety. Avoid providing information too far in advance. • Answer questions specifically (not with maybe / later), check understanding by asking them to repeat answer, limit the number of times a question can be asked. • Offer a range of appealing alternative activities or a variety of people to meet / spend time with. • Use diversionary tactics.
<p><u>Rigidity in thinking</u> is common. Rigid thought processes mean that information is stored in an orderly manner and there is a strong need for routine and consistency. Once something is learned it can be seen as the only 'correct' answer or method and it is difficult to have a change of mind or relearn.</p> <p>Rigidity can lead to stubbornness and argumentativeness.</p>	<ul style="list-style-type: none"> • Activities which encourage more flexible thinking from a young age may help. • Praise flexible thinking. • Encourage an understanding that there may be differing opinions, perspectives, answers or ways of doing things. • Pupils with PWS value authority figures but when they 'authorise' information, it can become set in stone and if incorrect, it can be difficult to convince a pupil with PWS of this. On the other hand, their response to authority figures can be useful.
<p><u>Non-compliance</u> An oppositional response can be an automatic 'flight or fight' response when a pupil is feeling anxious. Individuals with PWS are known for their ability to dig their heels in and be extremely stubborn. Sometimes stubbornness may be a coping device when there are genuine reasons for not wanting to do something, but they cannot find a way to tell you. They may not be consciously aware of their reasoning. It may be that something</p>	<ul style="list-style-type: none"> • Provide limited preferential choices to allow a sense of control over decision making. • Create clear rules, boundaries, consistency and a calm environment. • Avoid just saying 'No' and confrontation. Negative reactions raise anxiety further. • Resolve issues with compromise, finding a new solution together. Avoid ultimatums. • Offer empathy but repeat your expectation, then ignore unwanted escalation behaviour as much as possible.

<p>has gone wrong earlier in the day which is a precursor to the avoidance behaviour. Fortunately, pupils with PWS are eager to please, enjoy consistency and following rules. Routines and rules reduce anxiety.</p> <p>Occasionally, manipulation, lying or confabulation can be additional problems. Lying is an abstract concept and pupils with PWS may have difficulty in recognising what they say as lying.</p>	<ul style="list-style-type: none"> • Allow processing time for 'coming around' to a new way of thinking or an agreement. • Set a limited number of clear, positive goals for expectations of compliant behaviour and reward success. (<i>Incentives: computer time, colouring, music, reading...</i>) • In the event of extreme stubbornness that puts a person in danger, help them 'save face' by saying you need their help or try to move them on by offering a more pleasurable activity. • Focus on blame or recrimination fosters an atmosphere where manipulation or lying can flourish. • Ensure good team communication so that lying is identified and a behavioural pattern is prevented.
<p><u>Tenuous emotional control</u> is due to the impaired hypothalamus having poor control over emotional responses. Pupils can be volatile, becoming easily upset or frustrated.</p> <p><u>'Meltdown'</u> can be caused by sudden changes in routine or expectation, increased anxiety, pressurised tasks, confusion, sensory overload, communication difficulties, teasing, frustration, use of their possessions or other triggers.</p>	<ul style="list-style-type: none"> • Avoid false expectation and disappointment. • Acknowledge feelings and encourage communication about anxieties. Use tools such as feelings cards or the feelings thermometer. • Develop pupil understanding of how situations affect their emotions, 1. I can handle this. 2. This makes me uncomfortable. 3. This makes me nervous. 4. This can make me mad. 5. This can make me lose control. • Help them to identify their feelings and teach ways of coping with stress, i.e. using stress balls, listening to music, taking a break, relaxation. • Praise efforts to control emotions, especially in difficult situations. • Have a 'calm down' plan where the pupil may remove themselves to a safe area to calm down. The plan may include the availability of prepared calming activities. • Monitor for an impending meltdown, learn to recognise individual triggers, signs and take preventative steps. • Stay calm and try distraction, perhaps humour. • Avoid discussion and reasoning during a meltdown; reflect afterwards.
<p><u>Lack of impulse control</u> means that pupils will not self-monitor their behaviour well. They are more likely to engage in risky or dangerous behaviour.</p>	<ul style="list-style-type: none"> • Discuss situations and environments beforehand, what your expectations will be and what will happen if a lack of self-control occurs. • Encourage self-monitoring and evaluation. Offer reminders of their evaluations so they can focus on what they need to control / improve i.e. controlling voice volume.
<p><u>Aggressive behaviour</u> is rare but occasionally emotional outbursts can escalate to physical aggression.</p>	<ul style="list-style-type: none"> • List strategies a pupil can use when they are feeling anger, such as going for a walk, counting or rehearsed breathing exercises. • Have a plan for keeping the pupil and others safe in the event of an aggressive behavioural episode. • Predict times when anxiety will be higher and emotional control is likely to be reduced, such as proximity to meal times, when tired, if teased.
<p><u>Sensory processing difficulties</u> Pupils with PWS may be over-sensitive to certain stimuli or under-sensitive sensory seekers, or both. Sensory processing difficulties often improve with age. They do not respond well to chaotic environments with too much stimuli or too little structure.</p>	<ul style="list-style-type: none"> • Consider seating arrangements for comfort and limiting noise distraction. • Teacher transmitters and headphones enable the ability to block out other sounds and focus. • Avoid talking too much and information overload. • Hand fidget tools may improve concentration.

<p>Common sensitivities are loud noises, smells, movement, balance, touch and clothing.</p>	<ul style="list-style-type: none"> • Earplugs can reduce hypersensitivity to noise. • Pre-warn of high stimulus environments, i.e. fire drill • Provide space in crowded situations, around desk / locker • Hypersensitivity to fluorescent lighting may exist. • Water is often calming and stimulating. • A sensory diet, including sensory gyms and sensory boxes are useful developmental and management tools. • A quiet, sensory smart area is helpful. • New therapies target spacial awareness with vestibular-visual-auditory approaches involving listening programmes.
<p>Poor social skills Pupils with PWS are friendly and need friends, although they often lack the age appropriate social skills to form and maintain healthy friendships.</p>	<ul style="list-style-type: none"> • Identify what is confusing about conversations, i.e. eye contact, turn taking, listening, expressing thoughts and responding to humour etc. • Teach, model, role-play conversational skills such as listening to what others say, thinking about what they say, putting thoughts into words. • Practise conversations with topics of interest. • Use social stories for situations such as making friends. • Pupils could engage with a social skill group in activities that are of interest to them. • Support pupils during morning tea /lunch or planned school activities with peers. • Clear rules may be needed about social appropriateness and in particular for interactions with the opposite sex.
<p>Self-harming or repetitive behaviour can take the form of nail biting, skin picking, teeth grinding, teeth, hair or eyelash pulling and varies greatly in severity. It is usually a repetitive habit which worsens with boredom / disengagement and may be a form of self-stimulation. Less frequently, it may be the result of anxiety or emotional distress. Sometimes it can even be to provoke a reaction or seek attention. It is hard to stop due to difficulties in thought switching and lack of impulse control. Impaired neurological signalling also means pain or disgust may not be felt as intensely.</p>	<ul style="list-style-type: none"> • Use distraction and redirection. • Keep engaged and / or their hands busy. • Various lotions can moisturise wounds making them less tempting to pick. • Dress wounds and keep nails short. • Avoid giving attention to the behaviour – positive or negative attention. • If the behaviour is sensory seeking, provide more socially appropriate forms of stimulation, although avoid linking to the behaviour in case it rewards it. • Although it appears to be a compulsive behaviour, medications targeting OCD or anxiety often prove unhelpful. • A natural supplement called PharmaNAC (available in the USA) has proven beneficial.
<p>Increased risk for mental health issues Anxiety disorder is very common. Compulsive behaviours such as skin picking, hoarding and concerns with exactness are often seen in PWS, but some individuals will have a dual diagnosis of OCD. Oppositional defiance and problems with aggression can also be present. Occasionally, extreme impulsivity or attention deficit can lead to a separate diagnosis of ADHD. There are also increased rates of 'thought' problems in PWS (seeing things / hearing voices / strange ideas). In rare cases, behavioural episodes can become more extreme and evaluation for psychotic disorders may be required.</p>	<ul style="list-style-type: none"> • Possible interventions for psychiatric illness in PWS may involve supplements, medication and psychological strategies. • Little is known about the early phase of illness and risk factors that predict the emergence of psychosis in PWS. Research is ongoing although it is known that earlier intervention leads to improved health outcomes. <p>(NB. The management of food related anxiety and food seeking behaviour is covered in a separate section.)</p>

MANAGING THE FOOD ENVIRONMENT

UNDERSTANDING THE NUTRITIONAL PHASES OF PWS

In the past, PWS was described as a 2 stage syndrome - failure to thrive followed by hyperphagia. It is now known that the changes in appetite and weight gain which occur in PWS are more gradual and complex.

<u>Phase</u>	<u>Ages</u>	<u>Characteristics</u>
0	prenatal-birth	Decreased fetal movements and lower birth rate than siblings
1a	0-9 months	Hypotonia with difficulty feeding and decreased appetite
1b	9-25 months	Improved feeding and appetite; growing appropriately
2a	2.1-4.5 years	Weight increasing without appetite increase or excess calories
2b	4.5 - 8 years	Increased appetite and interest in food, but can feel full
3	8 years - adulthood	Hyperphagic; rarely feels full

Parents of children diagnosed in infancy have the opportunity to instil diet modifications and healthy eating habits well before the child's appetite or interest in food increases. As a result, when phase 3 begins it is often less severe in families who have implemented early intervention measures.

From age four, a child may become preoccupied with food, more anxious about it and have an increased appetite. At this stage they are likely to eat more than a typical child if allowed and may take food left around them if unattended. However, they can usually feel full and are able to be redirected or stop eating voluntarily.

The age at which phase 3 begins varies, but at approximately 8 years, an intense preoccupation with food can develop accompanied by food related anxiety. A child may rarely feel full; they may seek or steal food, eat from unsavoury sources or in rare cases, eat inedible items. A child can rapidly gain weight over a short period of time if not supervised. At this time, parents typically lock food cupboards to prevent food related anxiety and keep their child safe. However, the intensity of hyperphagia symptoms varies in individuals.

WHY DIETARY MANAGEMENT IS ESSENTIAL IN PWS

It is important to understand the **high risk of obesity in pupils with PWS due to the high fat to muscle ratio causing reduced energy expenditure** which means dietary intake has to be restricted to **approximately 50-70% of RDA**. Weight is gained very easily and a **reduced exercise capacity** makes it very difficult to lose. Imposing a restrictive diet on a child who can be obsessive about food and is likely to be feeling hungrier than their peers is difficult. Therefore, it is essential to support the strategies families have in place, especially as their efforts are additionally compounded by the behavioural challenges of PWS.

There is also an **elevated risk of diabetes** in PWS and parents are advised to avoid foods with a high glycaemic load – foods high on the Glycaemic Index cause spikes in blood glucose and insulin. Food also poses a serious health risk if a pupil with PWS were to gain unsupervised access to a significant quantity of food. A person with PWS **could quickly consume a large quantity of food resulting in choking or a perforated stomach lining**. Unfortunately, those who do not understand the reasons for dietary management in PWS often see a slim, healthy child and cannot understand why parents are so strict about food.

DIETARY MANAGEMENT STRATEGIES FOR SCHOOLS

Treats and rewards

Current medical advice for PWS is to follow a low carbohydrate diet and avoid sweet foods. It is known that continual blood sugar spikes increase diabetes risk, but they also do little for satiety. Brain scan imaging in PWS has revealed that people with PWS already experience a higher 'reward' from food so this is exacerbated by consuming sweet foods which are high reward foods and addictive.

Some families decide to avoid sweet treats altogether and only use non-food treats. Others decide to make or buy only healthy foods as treats. If a family does allow occasional food treats, it is important these are scheduled so not too many are given and anxiety is not raised by uncertainty about whether they will be received. Be prepared for the expectation that any treats given will be received again, and an excellent long term memory means they will not forget! **Ask the family for their rule about treats and support them in their plan.**

Do not provide any unplanned extras.

It is preferred if teachers do not use food as a reward because the anticipation of the reward raises anxiety. Any changes in diet or food rules can also raise expectation that these will happen again - consistency minimises future battles over food.

Celebrations and events involving food

Considering the needs of pupils with PWS at class or school events involving food is no different to catering for pupils with food allergies or intolerances. Most parents will not mind their child's health needs being shared with other parents and the PWSA can help with providing information if needed.

Make a plan with the child's family for celebrations and events involving food. If parents are forewarned that there will be shared food, they have the option to either provide an alternative, avoid the event, or to make allowance by adapting dietary intake before or after (they will want to place a limit on how much is allowed and will need to know how much was eaten.) The availability of food at school can be relentless with class birthdays, shared lunches, fundraising bake sales and sausage sizzles etc. It would provide great relief to families if schools were to **consider alternative food free celebrations**. Many alternative ideas for celebrating class birthdays can be found online and with a general population shift toward reducing sugar in our diet, **new ideas for food free and healthy fundraising are emerging**. A whole school policy on food at school would be of benefit to all pupils.

Food security

Food security is not just about keeping food out of reach – it is also about keeping food out of sight and mind. **Removing temptation to take food also removes the thought of food** which is preoccupying all thought. By taking that thought away, a pupil is able to focus on play and learning. Many older children with PWS actually ask for food to be locked away because it relieves the pressure of resisting urges and they want to follow their diet.

Keeping food out of mind requires more than just storing it out of sight. Food security is achieved by ensuring there is **no uncertainty about what and when food will be available** and there is no hope of any extras outside the schedule. No doubt and no hope leads to no disappointments - this method is taught by Drs Forster and Gourash at The Pittsburgh Partnership who coined the phrase 'food security' for PWS.

NO DOUBT

There is a predictable routine in which food is scheduled. (Focus on the sequence rather than the time of meals.) The person with PWS is able to relax and think less about food. Predictability of the menu also reduces anxiety.



NO HOPE (or chance)

There will be no unplanned extras outside the schedule and no opportunities for gaining access to food. Chances to obtain food cause stress. Food acquisition capability varies and can fluctuate, so modify the measures that need to be taken.

NO DISAPPOINTMENT

The person only has expectations that will be reliably carried out. No other expectations have been raised so there is no disappointment when these are not realised or fulfilled.

Tips for achieving food security in schools

- ✓ The pupil is aware of the schedule and where eating food fits within the schedule.
- ✓ The pupil has separate food for morning tea and lunch.
- ✓ The pupil is forewarned and reassured if the schedule is disrupted for any reason.
- ✓ There are no suggestions or promises of extras, and no surprises.
- ✓ Other pupils and staff know not to offer unplanned food to the pupil with PWS.
- ✓ Treats are not used as incentives or unplanned rewards.
- ✓ Food is not used as a consequence, i.e. the withdrawing or delaying of food.
- ✓ The pupil may require supervision during breaks and transitions.
- ✓ Opportunities for access to food are eliminated. This may require an inaccessible or lockable area for lunchboxes or bags or any other food sources.
- ✓ The pupil cannot gain access to money and opportunity to buy unpermitted food.
- ✓ Plans are in place for any special occasions and celebrations involving food and the pupil is aware of the plan in advance.
- ✓ The pupil is supported with making appropriate food choices when this is allowed.
- ✓ There are no consequences if food is stolen – encourage sharing of what was eaten.

[An A4 profile can be shared with all staff. Please ask for an electronic copy if you would like to edit this example template.]

Pupil Profile: **INSERT NAME**

Diagnosis: **Prader-Willi Syndrome**

Overview of PWS

PWS is a genetic disorder caused by a segment of genes on chromosome 15 either being deleted or unexpressed. The main characteristics are:

- **low muscle tone**
- **poor motor skills** (symptoms of dyspraxia)
- **excessive sleepiness or becoming easily fatigued**
- **a preoccupation with food and impaired satiety signalling**
- **incomplete puberty and infertility**
- **specific learning disabilities usually coupled with mild intellectual disability** (some pupils score in the borderline to low average range in cognitive testing, whereas a few pupils score in the moderate intellectual disability range)
- **speech delay, frequently with ongoing difficulties in expressive speech** (apraxia is common)
- **behavioural difficulties linked to anxiety, rigidity in thinking, sensory processing and emotional control** (many similarities to characteristics of ASD- strategies that work for ASD, work well for PWS)
- **impaired social skills** (reading social situations, picking up others' perspectives, conversational skills)

Insert photo

HEALTH AND SAFETY ALERTS

Individuals living with PWS may rapidly consume a dangerous amount of food if unsupervised access to food is gained. This could result in choking, serious GI issues, life-threatening stomach necrosis or rupture.

A special diet needs to be maintained - **consult with parents about activities involving food.**
A significantly reduced food intake is necessary due to an altered body composition causing reduced energy expenditure - **do not allow others to share food with them. Monitor food environments.**

Minor pain, vomiting, bloating, or slight temperature may indicate an emergency and requires evaluation.

Poor impulse control in PWS may result in unsafe behaviour, such as leaving the school grounds.

INSERT NAME's Learning Strengths

Visual learning – make use of visual aids.

I.T. – use computers and assistive technology.

Long term memory – excellent recall, especially of visual information. **Reading accuracy. Spelling.**

Non-narrative writing when using writing frames / graphic organisers and language banks.

Friendly / social – support to connect.

INSERT NAME's Learning Support Needs

Auditory processing – allow an additional 3 to 5 seconds for processing verbal information, use clear, literal language and check understanding.

Poor short term memory - use repetition to transfer learning from short term to long term.

Poor working memory – avoid lengthy, multi-step instructions, break down tasks, use organisational aids.

Reading comprehension. Creative writing – use personal experience and idea webs / supports.

Difficulty with abstract concepts – use concrete examples, systematic steps, hands-on maths aids.

Poor motor skills & stamina – adapt PE (exercise is vital to build strength), handwriting, typing tasks.

Expressive speech – allow extra time for speaking, model correct pronunciation of new words.

INSERT NAME's Behaviour Management Tips

Group work can be difficult – provide cues and guidance to encourage appropriate peer interactions.

Easily fatigued and distracted – provide activity level changes & breaks, consider positioning in class, homework?

High anxiety – small achievement steps, ensure tasks not too hard and expectations understood, routines and predictability are reassuring – avoid sudden changes.

Difficulty with switching task or topic (anxiety causes obsessive thinking / rigidity and looks like becoming 'stuck' – use visual cues / reminders / schedules, shorten tasks, use preferential activity incentives, set a number or time limits for questions, answer avoiding grey areas / ambiguity.

Tenuous emotional control (raised anxiety, stress sensitivity, poor coping skills can all lead to outbursts) – learn triggers, predict, plan, prevent. Typical triggers: confusion, frustration, sensory issues, unexpected changes. Minimise disappointment. Use 4C's. Acknowledge feelings, encourage use of coping strategies, ignore escalation, allow processing or calm down time. Reflect after.

Non-compliance (when anxiety has caused 'stuck' thoughts and refusal to accept change) – allow extra time to change thinking or accept change, empathise, offer choices, try distraction / redirection. Create consistency.

Skin picking / repetitive behaviour – use distraction and redirection, keep engaged / busy, avoid giving attention to it.

USEFUL RESOURCES

The following short video resource by Elizabeth E. Roof, M.A. (Senior Research Specialist at the Vanderbilt Kennedy Centre, USA) is recommended for teachers:

Tips for Teachers: <https://www.youtube.com/watch?v=Y5LhSePDvqk>

This lengthier video by a special education teacher and produced by PWSA(VIC) is also designed for teachers and includes an overview of the syndrome:

Learning with Prader-Willi Syndrome: https://www.youtube.com/watch?v=Y6_mivc-h1U

The PWSA(NZ) also has the following DVDs available on free loan from our library:

'Understanding the Student with Prader-Willi Syndrome: Strategies for Success' created by the PWSA(USA) and PWCF.

'Food, Behaviour and Beyond' by Drs Linda Gourash and Janice Forster MD from the Pittsburgh Partnership, USA. A developmental paediatrician and a neuropsychiatrist, together they have extensive experience and expertise in treating many patients with PWS.

For those working with younger children, the following short video by Janice Argawal (CNDT-PT) suggests useful therapeutic interventions for PWS:

Therapeutic Interventions for the Young Child with PWS:

<https://www.youtube.com/watch?v=-KtRnkoZdTw>

The following website provides very helpful strategies to help in different areas of **executive functioning**: <https://globalgenes.org/raredaily/leading-iep-champion-strategies-to-improve-executive-functioning-ef/>

and this website provides age related guides to activities for enhancing **executive function and self-regulation skills**: <http://developingchild.harvard.edu/resources/activities-guide-enhancing-and-practicing-executive-function-skills-with-children-from-infancy-to-adolescence/>

The following is useful information on how to help in the event of a **sensory meltdown**:

<http://asensorylife.com/sensory-meltdowns.html>

Many useful resources by other PWS Associations exist online, including IPWSO's FamCare advice on topics such as meltdowns, change, boundaries: <http://www.ipwso.org/famcare>

The PWSA(NZ) is able to **facilitate staff training meetings**. Please contact us for further details, or if **further resources or an electronic version of this pack** are required. If you are interested in connecting with other NZ teachers who are working with pupils who have PWS, please let us know as we are keen to facilitate the sharing of professional experience.