PRADER-WILLI SYNDROME
in the classroom

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Contents

FOREWORD

This handbook has been written for teachers, both primary and secondary, as well as giving guidelines for parents and caregivers. Information has been gathered from practical experience as well as taken from the Best Practice Guidelines for the Care of those with PWS, focussing on the holistic care, including education, of children and adults. These guidelines were compiled by an international group of professionals and cover the medical and physical characteristics of the syndrome as well as education and support needed, including transition from school.

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INTRODUCTION

Students with PWS are very receptive to learning and are keen to please. In general, they have good reading skills, but poor numerical skills and handwriting can be slow to develop. They show ability to learn computer skills and often have excellent fine motor skills, for instance many are particularly clever with jig-saw puzzles, threading beads, and many show an aptitude for fine handiwork including needlework and knitting. The IQ level generally falls in the just-below-normal category, but often shows 'islands of competence', in other words, they might be equal with their peers in some areas, but need support to reach potential in others.

Maths teaching needs to be conceptual, practical and often repeated several times before there is understanding. Teaching the use of a calculator immediately helps the level of understanding.
Once understanding has occurred the concepts generally remain.
Like all students, they thrive on praise.

Throughout integrated primary and secondary education it is important for the student to have teacher aide time. Although this is not always available for many students with special needs, it should be applied for on all levels. The need for teacher aides will not decrease as the student progresses.

This booklet addresses the unique needs of the student with PWS, giving a medical overview important to the understanding of the syndrome, as well as practical management strategies for teacher and student.

Prader-Willi Syndrome – Overview

PWS is a complex neuro-behavioural disorder resulting from an abnormality on the 15th chromosome. It was first described by Swiss doctors Prader, Labhart, and Willi, in 1956. The occurrence rate in live births is estimated to be between 1:12,000 – 1:22,000 affecting both males and females in all races. It is not inherited, although there are a few cases of more than one person with PWS in a family. This is because of an unusual genetic imprinting defect.

The syndrome is organic; there is a neurological flaw in the brain that affects cognition – understanding – emotions, and behaviours. Coupled with this, there is a presence of intellectual disability. However, it is equally important to recognise that people with PWS can be friendly, sociable, willing to help, and have some good and positive characteristics. It is also important to recognise that not every person with the syndrome will exhibit all the characteristics to the same extent.

Characteristics

Symptoms or characteristics of PWS may vary greatly in degree and severity from person to person, but at birth, the major characteristics present as follows:

- Decreased foetal movement
- Often abnormal position at delivery
- Baby is hypotonic (low muscle tone) and there is failure to thrive
- Weak cry
- Underdeveloped genitals

- Undescended testicles in males
- Delayed motor development.

As the child grows, further characteristics become apparent and these include:

- Hyperphagia (inability to stop eating)
- Obesity
- Speech delay/poor articulation
- Cognitive impairment/school problems
- Behaviour problems
- Skin-picking
- Abnormal pubertal development/hypogonadism

Among other things, the small deletion controls:

Appetite: There is no control mechanism to curb appetite and the person becomes obsessive and compulsive around food. Many behaviours stem from this.

Behaviour: Much behaviour is controlled from this small deleted area. Social niceties, ability to behave appropriately, behavioural outbursts and temper tantrums resulting in trashing, or prolonged crying, stealing, lying, stubbornness, manipulation, perseverance, and sometimes violence to possessions and people, are all common

Sexual development: Without sex hormone treatment, sexual development will be incomplete and will not develop normally.

Because there is no normal menstrual cycle, periods are inconsistent and often negligible. However, there have been a few international cases where a woman with PWS has conceived and given birth. Depending on the woman's diagnosis, she may have a normal infant, or one with Angelman's Syndrome. To date, there are no reports of men with PWS being able to father a child. A student with PWS taking part in classroom sex education may need some support in understanding the implications of PWS, but should need to know the physical repercussions of sexual engagement, stranger-danger, appropriate touch, and so on.

Genetics

Prader-Willi Syndrome is caused by a small piece of missing genetic material. This can happen in three different ways:
(1) The developing embryo fails to pick up a small piece of its father's genes, leaving a small "paternal deletion". This is the case in at least 70% of all diagnoses. (2) The developing embryo fails to pick up a small piece of its father's genes, but picks up two copies from its mother, giving a "maternal disomy" or UPD, which is the case in 25% diagnoses, or (3) there is a rare chance (less than 5%) of inheritance, or "genomic imprinting" when the fathers' genes are present, but are faulty and do not work.

Research is being undertaken to see what differences there may be in the three different genetic diagnoses. It is already known that those with the paternal deletion are clever with jig-saw puzzles, but may have a lower IQ, those with maternal deletion may show more of an

aptitude for learning, but can be more challenging, and those with genomic imprinting can show tendencies towards some autistic-like behaviours.

Growth Hormone Treatment

This treatment has now become an important medical tool to help control the likelihood of early onset of obesity and its inherent diseases. GHT is available from the age of 18 months until the child reaches mature bone growth which is somewhere between ages 14-16 years. With this type of medical intervention a child with PWS may not show the typical physical characteristics of the syndrome. The benefit of GH treatment improves muscle tone to the extent that the physique becomes as normal as possible. However, other characteristics of the syndrome, such as the drive to seek food, and behaviours associated with that, will still be present. Not all children will be able to access GH treatment. In this latter situation the physical characteristics of the syndrome will be more apparent. This may present problems of its own as the more 'normal' a child may look, the higher the expectations are that his or her behaviour will also be 'normal'.

Physical and Medical Characteristics

Without growth hormone intervention, physical characteristics will include:

- Almond-shaped eyes
- Triangular-shaped mouth with thin upper lip
- Small hands and feet
- Short stature (unless being treated with growth hormone)
- Scoliosis
- Narrow forehead

Medical characteristics may include:

- Scoliosis, or sideways curvature of the spine
- Osteoporosis in older age, creating brittle bones
- Dental caries
- Strabismus of the eye, requiring glasses
- Bruising easily
- Altered temperature regulation and inability to dress appropriately (ie too much clothing in summer, and not enough in winter)
- Lack of vomiting – this should be a 'red alert' when unknown foodstuffs have been ingested
- High pain threshold
- Incomplete onset of puberty
- Very low probability of reproduction
- Onset of diabetes Type II with weight gain.

Without environmental controls there is a great risk of extreme obesity. Coupled with poor muscle tone and an inability to exercise normally, inactivity, and a depressed metabolic rate where a person can gain weight on approximately one-half the calories required by a same-age peer, death may occur in young adulthood due to complications from obesity.

BEHAVIOURS UNIQUE TO PWS

As a result of the small deletion, IQ levels vary from just below average, to well below average, and because of the unique characteristics of PWS, behaviour problems including some of the following, will be seen:

- Hyperphagia and other food-related behaviours
- Self-injurious behaviour (skin-picking, biting…)
- Anxiety
- Obsessive thinking
- Compulsive behaviours
- Perseveration (asking the same question over and over again)
- Running away
- Concrete and rigid thinking
- Perceived unfairness
- Excessive sleepiness
- Lying and stealing (food, money, personal possessions)
- Tenuous emotional control

Links to Autistic Spectrum

It is now common to find there may be some link with the autistic spectrum, particularly in obsessive compulsive behaviours. Being aware of this will help solve some of the behavioural jig-saw and provide teachers with a wider range of behavioural tools.

Hyperphagia and other food-related behaviours

Those with PWS have a seemingly insatiable appetite. This is caused by the genetic deletion and dysfunction in the hypothalamus. It helps to look at this as a 'starvation syndrome' as this can shift the perspective of 'bad' behaviour to a need for support and boundaries. Regardless of whether the student has had growth hormone treatment and does not present as overweight, there is still a preoccupation with food, food-seeking or foraging, sneaking, hiding or hoarding. There have also been many cases of a person eating unusual food, or food-related products. You may also find, if your school provides breakfasts for students, that the person with PWS will be lining up for this as well. It would be unusual for this student not to have had breakfast already, and it would be wise to check with their home.

THE STUDENT WITH PWS

Students with PWS have varied aptitudes, abilities and interests; and a representative learning profile should be developed. Regardless of IQ, most students have learning disabilities and benefit from teaching strategies utilised for students with similar difficulties.

Relative Learning strengths:

- Good long-term memory skills
- Receptive language
- Good at puzzles
- Visual processing (relative to auditory processing)
- Basic maths skills (especially with calculator)
- Reading skills
- Good computer skills (some)
- Social and friendly

Learning weaknesses:

- Poor short-term memory skills (visual-motor and auditory-visual)
- Expressive language
- Poor fine and gross motor skills
- Sequential processing deficit
- Difficulty with abstract concepts
- Short attention span
- Communication difficulties (may end in temper outburst)

Identifying the challenges

It is important that the behaviours or issues that challenge are identified. Some will result in having to manage the actual environment with school lunches being kept out of reach, money being held by teachers, and some will call for new management skills.

TEACHING STRATEGIES:

Give positive instructions

Students with PWS tend to have a fairly rigid, or concrete way of thinking and tend to work best to a set routine and positive timetable. They can accept change if prepared for it beforehand, but a sudden unexpected change may result an uncooperative student. It is sensible preparation to warn beforehand if something is to be postponed or cancelled. If the student doesn't clearly understand the instructions, you may find resistance to change. Do make sure the student knows that this will not change lunch time! A common indicator of this might be persistent or repetitive questioning from the student in spite of having heard the answer. Be patient; use visual as well as verbal instructions.

Perseveration

Constant persistent questioning is a common characteristic of the syndrome. Answer questions up to two times, then write it down if necessary. Check with the student to see they clearly understand your answer. Ask them to tell you what they are going to do, or what the answer is; even ask them to write it down for you.
Clear communication is the key here.

Ignore-Redirect-Praise

Simple behaviour modification techniques such as this work well. It may require removal from a situation that appears to be heating up, and redirection to another task until the person has calmed down. Redirection may include taking the student to a quiet room where he or she may be able to talk to you about their concerns. If talking is not an option, quiet reading, or music, or just time away from the classroom activities may allow the student to calm down. Giving them a small job to do will enhance their self-esteem and enable them to move gracefully back into the classroom.

Use visual schedules

The student may have difficulty in finishing a task on time, or may display perseveration (repetitive questioning), both of which are characteristics common to the syndrome. Perseveration indicates an inability to process information coherently or sequentially. Not finishing a task on time can indicate some obsessive compulsive behaviour. In both cases, using both a verbal reminder combined with a visual schedule, or prompt, will assist the student. This may be anything from a simple egg-timer, to a daily pictorial class schedule, to help with time-keeping.

Always have a Plan A and a Plan B

It is sensible to have a fall-back plan in the likelihood that your Plan A does not eventuate. This will prevent disappointment, frustration and anger, and keep the student in the loop. Working out probabilities and possibilities beforehand, including extra staffing, is a precaution that is essential. Occasions will arise when the student refuses to go on an outing, or to participate in a class activity. Rather than have this spoil the day, Plan B can be brought into play.

No more than two-step directions

Generally, there is an inability to process more than two things at once, so make life simple and make choices "either/or" and directions simple.

Break tasks down into manageable parts

Giving the student a task to complete may seem simple to you, but almost incomprehensible to the student. "Make this bed", for instance, is a simple instruction, but unless you teach the method and process of making the bed, it will not get done. Make sure the student really understands how to do something, before expecting the task to be completed. Check, and check again. Have them repeat instructions back to you. If need be, write down the stages, or ask the student to write them down. If the task is not finished in time, but the process has been followed, you will have a clear idea of the student's capabilities and be able to match the task to the instructions in future.

Shortened assignments

For the same reason as tasks need to be broken down into manageable parts, assignments need to be short, to the point, and

within the student's capabilities. You may find some students become obsessive over doing assignments, spending hours of time on the topic. Although this may show dedication and a desire to please on the part of the student, it may lead to their compromising other areas of work including sleep hours! If this is the case, re-set the assignment, but not necessarily with time limits as this may also arouse frustration sometimes to the point of work being ripped up and destroyed.

Brief (or no) homework assignments

You need to identify whether the student is capable of carrying out homework assignments. It is a good idea to check with the parent to see whether it is creating any extra stress at home before assigning too much work.

Ask – don't demand

As with any student, more will be obtained with careful asking rather than a direct demand or instruction. Although this may take a little longer, you will gain the trust and respect of the student and bank valuable points for future use!

Controlled access to food and money

It practically goes without saying that, for the student with PWS, it is paramount to keep the temptation of food and money strictly limited.

MANAGING THE ENVIRONMENT

If the student finds an easy access to food, they will take it without a second thought. It does helps to think of PWS as a 'starvation syndrome' because the food-seeking mechanism in the brain is permanently turned on and the brain is constantly telling the stomach that it's hungry and needs to seek food; if not for now, then for later. This is fundamental to the syndrome. It will mean that all access to food is controlled.

Identify areas of accessibility:

- o School lunches
- o School bags
- o Food 'rewards' used by teachers
- o Cooking/manual training classrooms
- o Staff rooms
- o Nearby dairies or shops

Work out ways to control them, for example:

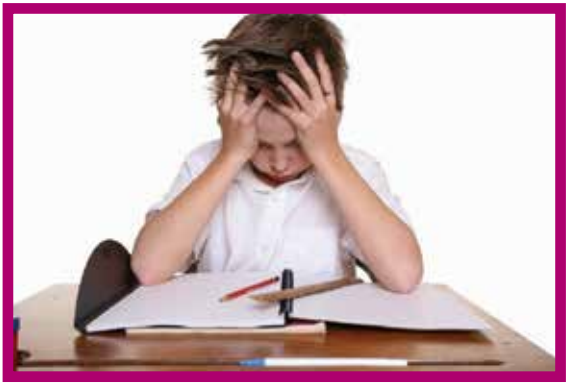
- o Lunches distributed by teacher
- o School bags kept closed and inaccessible (locked away)
- o Any food treats or sweets kept in teacher's possession at all times
- o Cooking rooms locked
- o Staff rooms made 'food-safe' or 'food free'
- o Shops and dairies made out-of-bounds or told of the problem
- o Do not put on 'rubbish-bin' duty!

People with PWS have anxieties that are increased around food. They are not as capable as you are of controlling these anxieties.

If you know the student has had access to food, do not accuse them of this as it will result in denial and a "yes you did" "no I didn't" situation. Make sure to the best of your ability that the situation won't occur again. By telling the student what you are doing in such a way that does not lay blame, for example, "I am taking these sweets away to be locked up as I know they are too much of a temptation/I am locking this kitchen door so that you are safe from temptation" you are respecting the innate condition of the syndrome while, at the same time, taking control.

If the student has a diary of messages that go home with them, make sure you let the parent/caregiver know of excess food that has been eaten during the day. However, if the student is capable of erasing the message, use another system such as phone, email.

Teacher aide hours during lunch-time and other breaks where food is accessible, is vital.



WORKING WITH CHALLENGING BEHAVIOURS

The most effective way to deal with challenging behaviours is to prevent them from happening in the first place. However, this can't always happen and hindsight is a wonderful thing. The most common identifiable thing that leads to challenging behaviour is anxiety.

For any person with an intellectual disability, anxiety is about the biggest threat to their wellbeing.

Many things can make a person anxious but one of the most important is not being able to make themselves clearly understood, or understand what is expected of them. The other major issue is knowing what the rules are around food, when they might expect it, at what time, and that there is enough. Other issues include not letting them know something is about to be changed; having boundaries shifted; new rules that they did not know about; new staff member (will they understand me?), change in staff.

Anxiety will display itself in many different ways. Some of the symptoms which may present:

Non-compliance

Usually non-compliance occurs when the person does not understand what they have to do, or something has gone wrong earlier in the day and anxiety levels are beginning to rise; or they simply do not want to do the task now, but may do it later; or do not feel well, or do not feel able to do the job.

Break task into manageable components.

Do not push the student until they are ready to move on. You may notice other avoidance signals such as putting head on desk and saying they are tired, complaining about a pain somewhere, spending time looking for another ruler, piece of paper, pencil, etc.

Use your judgement as to whether the task is necessary; whether it will disrupt the class or routine if not done immediately… it doesn't really matter whether the student completes the task. What matters is that s/he doesn't disrupt the rest of the class and the behaviour doesn't lead to a blow-out

Frustration

Frustration only happens because the student is unable to understand how to make a thing work, how to get their message across to you, or what is expected of them. Generally this has occurred because of mis-communication.
People with PWS think very concretely. They value authority figures. If someone 'authorises' something, then that almost becomes a rule, even if the information is wrong, or slightly off the mark, it won't matter. You will have to work very hard to convince the person with PWS that their information is incorrect.

Take time to back-track with the student to find the root cause of the frustration. Ask simple closed questions which are easier for them to answer (yes/no) as they may be unable at this stage to put into words the cause of their frustration.

Make sure the communication is clear – and check with whoever else needs to be in the loop.

Provide good, logical details and reasons (invoking a higher authority if need be), such as you would use yourself. If a higher authority is invoked (principal, school board, police, doctor's surgery, etc), make very sure of your facts as you may need to verify this by phone.

Argumentative

Take a break. and ask the student if they would like to be on their own for a while.

Sit with the student and listen actively – keep asking 'what else?' until they literally run out of argument.

Muddledness

This is another sign that the message isn't getting through: try to redirect to one task at a time – if there are too many tasks, there will be anxiety.

Accentuated compulsive behaviour

This usually shows up as insistence on completing the task in hand (whether it is a set task, or something the student has decided s/he 'needs' to do)

Sometimes it is better to let the student carry on and complete the task regardless of whether it is within the timeframe, or help them complete it and move onto something else.

The student may need to learn from his/her mistakes.

Reasoning at this stage may be impossible. But you will be able to talk about it later and point out the inappropriateness once the student has moved on (this could be an hour later, or a day later)

Denial

Usually the first form of defence. Unless you catch the person red-handed (ie rummaging around the rubbish bin, or in someone else's property) there is no point whatsoever in accusing them of doing something.

It may be better to note your suspicions and pass them onto the relevant person.

Choose your battles in order to win the war (ie if it's not the end of the world, don't make an issue of it).

You may be able to readdress this later on and ask the student if s/he needs help around that area, *"Let's try locking up the sweetie jar so you won't be tempted."*

Be careful about using closed questions (yes/no), you may not get an accurate answer.

You may be able to ignore the situation, or redirect.

A person with PWS will not reason coherently when in a state of denial. Do not try to win the argument.

Perseveration

Take time to answer their question as fully as possible. Check to see if they have understood. Check again *"Can you tell me what I've just said, so that I know you've understood me?"*

Be prepared for them to repeat the question, so answer it in exactly the same way, and check it. If the person continues to perseverate despite your clear communication, be aware that this may be a type of compulsive disorder and that they are becoming anxious.

Try breaking down the question, or task, into very small portions. It is simply no good saying, *"Do the work on page 22"*. Break it down into manageable components.

Use visual aids. Make a check list with two or three items on it – never give more than one choice, *"this, or that"*, if it looks as though the person is becoming confused.

Drop in the level of communication or concentration

Try taking a short break, giving the person something else to do, running a message (accompanied).

What can you do?

In working with a person who is unable to control anxiety, it is absolutely vital that you ask yourself the following:

- What is fair to expect?
- What is set to fail?
- What needs to be rewarded?
- Are rewards needed?
- What rewards will work? *(Do not use food as a reinforcer – you will set up an automatic expectation)*
- Are lines of communication still open?

Managing anger outbursts

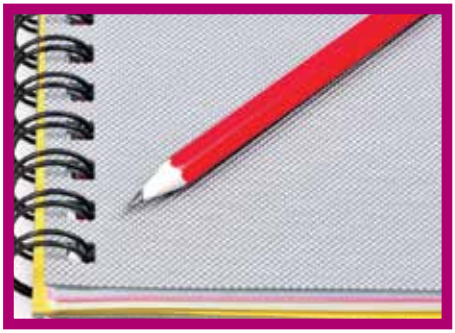
In spite of your best efforts, the student may have anger control problems and, when especially anxious, will yell and scream, destroy property and may hit others (usually the adults). Your job is to find ways to prevent these outbursts to begin with and to have a plan of action for when they do occur. Once the student's behavioural level starts to rise, never, ever, under any circumstances attempt to win an argument or to persuade the student with PWS to take a different viewpoint – it simply does not work, and your logical and rational approach will be wasted. Persistence on your part will encourage resistance on their part. Keep your voice low, and your actions slow.

You may choose to remove the student from the classroom, or, if that is not possible, remove other students. Use a time-out method, listen quietly to the student's concerns and try to involve the student with any problem-solving. Offer them something practical to do, or say, the next time they feel in the same threatened situation.

Think of PWS as an anxiety disorder and a starvation syndrome, rather than an over-eating and 'bad behaviour' syndrome, but at the same time, never forget their powerful drive to seek food. Recognise that the individuals often feel out-of-control so sticking to what they know or believe to be true, will help alleviate their anxiety. They do not have adequate coping skills to manage their many stressors.

The more knowledge you have about PWS, and the more you can think like a person with PWS, the better you will understand and be able to support their needs. Get to know the student really well – get family input to help you. The more you collaborate with the family, the more you will understand the big picture – you need their knowledge of their child's life with PWS, and they need your expertise. Recognise that this syndrome can cause a greater amount of family and personal stress than almost any other syndrome.

Set up the environment and schedules to be as predictable as possible while anticipating and planning for changes. Make small changes one at a time and remember that "consequences" don't work in the long run.



SERVICES AND SUPPORT

A network of support to help the student through school might include any or all of the following:

- Teacher aide
- Speech therapy
- Occupational therapy
- Physical therapy
- Behaviour management
- Medical management
- Dietary consultation
- Adaptive physical education
- Advocacy
- Staff training programmes offered by the PWS Association

Some of this will already be in place when the student comes to school, some of it the school can provide.

Transition

When looking at transitioning to another school, or out into the community, there are many things a school can do to work in with the new systems and help this go smoothly:

- Case management
- Vocational services
- Residential services
- Recreation and leisure services
- Transportation services

The Future

What will this mean for the student leaving your school? There are several 'givens' that must be included in any planning; here are three of the non-negotiables:

1. Individuals with PWS must be supervised day and night (whether by home support or paid staff) to ensure appropriate weight and health status

2. The drive for food is so strong that a person with PWS will knowingly put themselves in dangerous situations to obtain food or money

3. Without adequate supervision the individual eventually becomes morbidly obese and dies.

Specialist Schools

New Zealand has two special residential schools, Salisbury School for Girls, in Nelson, and Halswell College for Boys, in Christchurch, both of which were established by the Ministry of Education for students with learning and social difficulties, and both schools have acquired a high level of knowledge and understanding of PWS over the years. You can learn more from their websites:
www.salisbury.school.nz and
www.halswellcollege.com

Employment

Many individuals with PWS are able to manage paid employment with natural supports and a job coach. Most common jobs are assembly, clerical, cleaning, and some childcare and animal care. Many individuals with PWS are motivated by money and job responsibility. There is also a desire to be the same as their working peers. Self-esteem and pride in their work comes naturally to them. However, the most common reasons for leaving a job are food or behaviour-related. It is often more difficult to provide one-on-one job support for an adult, but supported employment is a good start.

Conclusion

Most students with PWS work well at school with the right support. They usually enjoy schoolwork and have a desire to please. A representative learning profile within the syllabus, and with support, will work best for the student. This may carry through to their final years of school with more time spent on life-skills and work experience as they transition from school into the community.

For further help, contact the PWS Association.
Freephone 0800 4 PWS HELP

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