Forward

The Prader Willi Network came together as a result of an Innovation Grant in 2005/06 from MCSS to the Central West Region. The Network had several goals, one of which was to create a handbook for providers that spoke to best practices designed to assist people living with this syndrome.

Our thanks to Jo Anne Nugent who pulled the Handbook together in record time and to all the agencies who shared experience, forms and information.

I’d like to thank the members of the Network for their dedication and financial assistance (both funding and in kind) throughout this project.

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To find out more about the PWS Network and Prader Willi Syndrome please visit our web site at www.pwsnetwork.ca

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INTRODUCTION

The first report of Prader Willi Syndrome (PWS) was published in 1952 by the authors Prader, Labhart, and Willi. (Alexander et al, 2000) Since then, there have been many books published about PWS. Some of these are comprehensive text books with a considerable amount of technical information. Other publications take the form of manuals that provide basic information and recommendations aimed at specific audiences such as parents or teachers. In addition, volunteer organizations have developed such as the Ontario Prader-Willi Syndrome Association for families or professionals in the field. These groups also make educational materials available. Furthermore, a growing number of families and agencies have been supporting people with PWS in our communities.

Therefore, we are now at the point in the history of PWS that a considerable amount of excellent ideas exist with respect to how to best provide these supports. It is time now to gather these ideas into one resource.

The purpose of this Handbook is to provide practical information to staff and organizations about best practices in supporting people with PWS.

Of course, this information could also be helpful for families and other professionals. But our primary focus is on those staff who work on a day to day basis with people with PWS.

The format of the book includes a basic introduction to PWS, some overall strategies for providing supports in categories related to food, environmental modifications, specific types of services across the lifespan, training, and the future of service provision.

The ideas in this book have come from interviews with dedicated staff from various agencies, as well as from written resources. The focus of the book is on ideas that work – suggestions that agencies can use whether they are currently providing services to individuals with PWS or are planning to do so in the future.

We sincerely hope that this Handbook is useful to you and the people with PWS that you work with.

Jo Anne Nugent
The Prader-Willi Network

April, 2007
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CHAPTER ONE:
AN OVERVIEW OF PRADER-WILLI SYNDROME

This Chapter is a basic introduction to Prader Willi Syndrome (PWS), focussing on:

- What PWS is
- What causes PWS
- Diagnosing PWS

What is Prader-Willi Syndrome?

A syndrome is a condition or disorder with a recognizable pattern of symptoms. In the case of PWS, the pattern of symptoms consists of altered growth and development that continues across the person’s lifespan. (Alexander et al, 2000)

Researchers believe that the main cause of the symptoms of PWS is a problem with the hypothalamus. (PWS Association USA, 2007) The hypothalamus is a part of the central brain which connects 2 critical body systems, the nervous system and the endocrine system. It regulates growth, sexual development, appetite, metabolism, body temperature, mood, and other functions. When we consider how important the hypothalamus is, we begin to understand how the malfunction of the hypothalamus which occurs in PWS can have a major impact on the person’s total functioning.

PWS is relatively rare. Estimates of its frequency vary, however it is usually estimated that PWS occurs in 1:10,000 to 1:25,000 newborns. PWS affects males and females equally. It also occurs in people of all races and in all geographic locations at the same approximate rate.

PWS cannot be “cured” and lasts a lifetime. However, there are now many successful strategies that can be used to minimize the symptoms of PWS. People with PWS can enjoy a typical lifespan with a good quality of life.

In spite of its low rate of occurrence, people with PWS do have significant needs and thus require many services and supports. Best practices help us to provide services in the most effective way possible to ensure quality of life for the individuals while stretching our resources to serve the most people.

What Causes Prader-Willi Syndrome?

PWS is a genetic disorder. This means that an error has occurred during conception which has affected the development of the fetus.
Understanding Genes and Chromosomes

The topic of genetics can be technical and somewhat intimidating for most of us! However, Linda Keder and Gerard Butler have provided a basic helpful explanation of the genetics of PWS which is summarized in this section. (Linda Keder & Butler, 2004)

Chromosomes are tiny packages of genes that are present in nearly all the cells of our bodies. Genes contain the instructions that direct the growth and development of the baby both within the mother’s womb and after birth. We inherit these genes from our parents during conception when the egg and sperm containing their genetic materials combine.

Each gene is located on a specific chromosome. Most cells in our bodies have 46 chromosomes which are structured as 23 pairs. 22 of these pairs are identical, half from the mother and half from the father. The 23rd pair determines the gender of the baby. The mother always supplies an X chromosome, while the father could supply either an X (girl baby) or Y chromosome (boy baby).

Changes or errors in genes and chromosomes are common when the egg and sperm cells are formed. Some of these genetic changes will have no effect on the baby at all; some will result in miscarriage, and some cause genetic disorders such as PWS, Down Syndrome and Williams Syndrome.

The Genetics of PWS

PWS is caused by an error on chromosome 15. (Butler et al 2006; Greenswag & Alexander, 2004) There are 3 different ways this can happen:

1. Paternal Deletion: in about 70% of PWS, part of the chromosome 15 which has been inherited from the father is missing.
2. Maternal Uniparental Disomy: In about 25% of PWS, the baby inherits both copies of chromosome 15 from the mother. When this happens, the father’s chromosome 15 is lost.
3. Imprinting Defect: In less than 5% of cases, the father’s genes on chromosome 15 that relate to PWS are present but they don’t work.

Please note that all 3 types of PWS occur through chance and the behaviour of the parents does NOT cause the chromosomal problems to occur.

Types 1 and 2 have a relatively small chance of reoccurrence of PWS within the family. In the 3rd type, there is a significant probability of more than 1 child in the family being born with PWS. In any case, it is strongly recommended that families who have a child with PWS seek genetic testing and counselling, no matter how their child’s PWS was caused.
**Diagnosing PWS**

The diagnosis of PWS is made in 2 steps.

Firstly, the family and/or medical personnel note the presence of specific “PWS-like” symptoms within the baby or young child. Secondly, genetic testing is conducted to investigate whether PWS is present. Sometimes, people may have some of these symptoms and NOT have PWS. Only the genetic testing can confirm if PWS is definitely present in the individual.

*The Diagnostic Criteria*

Diagnostic criteria are the signs or indicators of a condition that are used to make a diagnosis.

In 1993, an extensive list of diagnostic criteria for PWS was published. This list was found to be rather broad, somewhat cumbersome, and not always accurate.

Therefore, a revised list has been published which has the indicators which would prompt genetic testing. (McCandless & Cassidy, 2004)

**Table One: Proposed Revised Criteria to Prompt Diagnostic Testing for PWS**

<table>
<thead>
<tr>
<th>Age</th>
<th>Features Suggesting PWS</th>
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</thead>
<tbody>
<tr>
<td>Birth to 2 years</td>
<td>Hypotonia (low muscle tone, “floppy”)</td>
</tr>
<tr>
<td></td>
<td>Weak suck</td>
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<tr>
<td>Later childhood</td>
<td>Neonatal history of hypotonia and weak suck</td>
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<td></td>
<td>Global developmental delay</td>
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<td></td>
<td>Excessive appetite</td>
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<td></td>
<td>Lack of satiety (never feels full), also called hyperphagia</td>
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<tr>
<td></td>
<td>Obesity if food not controlled</td>
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<tr>
<td>Adult</td>
<td>Mild or borderline mental retardation</td>
</tr>
<tr>
<td></td>
<td>Excessive appetite</td>
</tr>
<tr>
<td></td>
<td>Lack of satiety (never feels full), also called hyperphagia</td>
</tr>
<tr>
<td></td>
<td>Obesity if food not controlled</td>
</tr>
<tr>
<td></td>
<td>Hypothalamic hypogonadism (under developed genitals)</td>
</tr>
<tr>
<td></td>
<td>Specific behaviours: obsessive-compulsive features, skin picking, temper tantrums</td>
</tr>
</tbody>
</table>

These are the most basic criteria for PWS.

Future Chapters will deal with criteria in more detail with reference to physical/health issues, food/nutrition, behavioural issues, and development across the life span.
CHAPTER TWO: CHARACTERISTICS OF PWS

An individual with PWS has said: “It is like a big black cloud that follows you wherever you go and every once in while, more often than not, it will rain on your parade.”

With understanding and effective supports, we can help to dispel that black cloud and people with PWS can have a good life.

This Chapter will present a brief overview of the most common characteristics of PWS. People with PWS are, of course, individuals with their own personalities. However, because they share a genetic problem, there are many symptoms that they have in common.

The topics covered in this Chapter are:

- Physical appearance
- Health issues
- Intellectual abilities
- Behaviour patterns
- Emotional/mental health

It is useful for service providers to be aware of these characteristics in order to provide supports that will lessen the severity of these issues.

Physical Appearance of People with PWS

Although there are individual differences, people with PWS tend to have:

- Facial characteristics of down turned mouth and almond shaped eyes
- Small hands and feet
- Short stature
- High fat to muscle ratio with body fat tending to be distributed in the trunk and thigh areas
- Fair colouring – depending on race

Health Issues

There is a discouraging long list of medical complications that accompany PWS. It is important to remember that with early intervention, appropriate supports, and consistent monitoring, many of these health issues can be dealt with very effectively.

It is also important for staff to be aware of these medical concerns because staff play a key role in monitoring the health of individuals with PWS.
The information for this section has been adapted mainly from the PWS Association of the USA website and Management of Prader-Willi Syndrome. (Butler et al, 2004, PWS Association USA, 2007)

**Hyperphagia**

The most serious health condition associated with PWS is hyperphagia or lack of satiety. The sensation of being full never reaches the brain of a person with PWS. Therefore, without help, s/he can eat constantly and would literally die from overeating. In addition to never feeling full, people with PWS have hormone deficiencies with the result that they gain weight on half the calories of others their age.

Therefore, the person with PWS is in a condition of “double jeopardy” regarding food – s/he never feels full and yet should actually eat half of what others eat to avoid serious weight gain.

**Gastrointestinal System**

The gastrointestinal system is used for eating and absorbing the nutrients in our food. People with PWS have:

- Lack of vomiting which can be an issue if unhealthy food items have been eaten since the body cannot do its job of getting rid of dangerous substances
- A higher number of severe gastric illnesses
- Weakness of the stomach
- Constipation
- Dental issues such as thick salvia and a tendency to cavities – anti bacterial toothpaste, mouth wash, or gum can help with this

It is also important to emphasize that people with PWS are more vulnerable to digestive system problems – what would be a minor problem for someone else is often a life threatening issue for someone with PWS.

**Obesity**

Without help, the person with PWS will eat to the point of life threatening obesity. Associated with obesity can be other problems such as:

- Heart failure
- Hypertension (high blood pressure)
- Type II diabetes
- Leg edema (an abnormal and serious build up of fluid between tissue cells) which makes the skin more prone to skin ulcers, thrombosis (blood clots), and cellulites
- Infections in deep fat folds

**Respiratory Disorders**

The most serious respiratory (breathing-related) disorder for people with PWS is breathing disruption during sleep. This can take 2 forms:

- The breathing process can be slowed down during sleep (central hypoventilation)
- Breathing can stop altogether during sleep (sleep apnea)

As well, pneumonia can occur due to poor muscle tone.

**Musculoskeletal Disorders**

Along with hypotonia (low muscle tone), people with PWS have higher rates of:

- Scoliosis
- Osteoporosis
- Hip dysplasia (abnormal growth)
- Fractured bones
- Lower limb abnormalities

**High Pain Threshold**

People with PWS often do not show the typical signs of pain and/or may not complain of pain. Therefore, parents and staff may not know that the person has an illness or injury until it reaches a serious stage. Careful observation is recommended to note minor changes that may indicate a health problem so that treatment can be given before a life threatening situation develops.

**Body Temperature Abnormalities**

Two types of abnormalities have been reported:

- Hyperthermia (abnormally high body temperature) which can occur during relatively minor illnesses or surgical procedures
- Hypothermia (dangerously low body temperature) which can result in lack of a fever even when there is an illness. In such a case, the illness could be undiscovered since the symptom of fever is missing.

Staff and parents should be aware of these issues and also ensure that doctors have been informed of these concerns.
Adverse Reactions to Some Medications and Anaesthesia

People with PWS have been documented as having unusual reactions to medications:

- Exaggerated and prolonged responses to sedating medications mean that extreme caution should be taken in the use of such medications
- Water intoxication can result when using medications with anti diuretic effects which cause a decreased flow of urine

Medical staff should be made aware of these issues since they can be life threatening.

Incomplete Sexual Development

Hypogonadism results in incomplete sexual development. This should be monitored by a doctor in case of medical complications. As well, staff and parents should understand that this can cause embarrassment for the individual if s/he is in a situation where other people are able to observe this condition such as being in a change room at a high school or a public swimming pool.

Tiring Easily

Individuals with PWS tend to tire easily. This is particularly noticeable later in the day. The tiredness can be accompanied by difficulty paying attention to tasks or actual falling asleep.

The Use of Growth Hormones

In PWS, the hypothalamus does not work properly. One result of this is that there is a deficiency in the hormone that regulates growth. Since 1985, synthetic growth hormones have been used with children and adults who have PWS in an effort to counteract the negative effects of their growth hormone deficiency. (Carrel et al, 2004)

There have been many positive results from Growth Hormone (GH) Therapy such as:

- Increase in height
- Growth of lean body mass and decrease in fat mass
- Increase in metabolism
- Increase in energy
- Increase in bone density
- Improvements in physical strength and function
• Reported increases in mental alertness and concentration – there is only informal anecdotal information on this point

GH therapy starts with children and continues until they reach full adult height. The decision is then made whether to transition the individual to adult GH therapy. It appears that the body composition benefits of GH therapy are reversed if the adult does not continue with it. Therefore, in most situations doctors will transfer the young adult to adult dosages of GH.

Overall, GH therapy appears to be relatively safe, although comprehensive research has not been done regarding the effects. However, there are medical complications that are known to be associated with GH therapy such as retention of salt and water, nausea, dizziness, decreased insulin sensitivity or headaches. (Ibid) Therefore careful medical monitoring is necessary.

**Intellectual Abilities**

*Developmental Delay*

Most people with PWS have a degree of developmental disability:

- Approximately 12% are within the “normal” range of intelligence
- Another 29% are in the “borderline” range
- The remaining individuals fall within the “moderate” range with a very small number being at the “profound” or “severe” levels

*Cognitive Strengths*

People with PWS tend to have specific strengths with regards to intellectual functioning (Levine et al, 1996):

- Long term memory
- Receptive language
- Ability to learn from visual materials such as videos, photos, etc.
- Reading
- Ability to learn from hands-on experience
- Writing

*Cognitive Issues*

Some areas of concern with respect to intellectual abilities are (Ibid):

- Short term memory
- Fine motor skills
- Mathematics
- Abstract thinking
Auditory processing (the ability to learn through verbal instructions or discussions)

Thus, in spite of potential intellectual delays, people with PWS have cognitive strengths that can be built upon both in school and in their adult lives.

Thinking Patterns

It has been noted that people with PWS have specific thinking patterns:

- Rigid thought processes: People with PWS receive and store information in an orderly manner. They prefer routine and consistency to help them learn.
- Perseverative or obsessive thinking: This is a tendency to get “stuck” on one topic or thought, often accompanied by anxiety. The person may repeat a question many times or talk about the same thing repetitively.

Thus, people with PWS see the world in a structured way and they need reassurance that their day will take a consistent, familiar pattern. This familiarity is important to them and helps decrease their anxiety levels.

Behaviour Patterns

A number of consistent challenging behaviours are displayed by people with PWS.

Food Obsessive Behaviour

Perhaps the most obvious behaviour issue is what is referred to as food obsessive behaviour (Linder, 2002). Food obsessive behaviour (FOB) is a range of behaviours that relate to the individual’s constant need to eat:

- Over eating, to the point of endangering health and life
- Foraging: searching everywhere for food or food related products, including garbage cans, or other people’s rooms, lunch bags, desks, etc.
- Pica: eating non edible items, particularly products such as dish detergent, spoiled food, garbage, etc.
- Stealing: items which are stolen relate to food or obtaining food. Therefore, the person may steal food from others, or money with which to buy food, or keys to a locked food storage area.
- Hoarding: gathering and hiding food or food related products such as shampoo, even when there is evidence that the person has large stores of these items.
- Lying: dishonesty regarding having eaten a meal, how much was eaten, stealing food, hoarding, or stealing money.
Self Injury

Self injury can take many forms:

- Skin picking: this can lead to serious harm. The behaviour is somewhat puzzling because it typically occurs when the individual is calm so does not seem to relate to the person being in emotional distress. It could be caused by boredom or anxiety, but this has not been proven.
- Self injury or self mutilation: this is a sudden onset of self injury which may or may not accompany an emotional outburst.
- Rectal self injury: this behaviour is not well understood, but can result in injuries such as bleeding, infection, or fecal incontinence.

Many of these behaviours have the potential for grave consequences such as scarring, maiming, illness, or death, if not dealt with.

Compulsions

There may be other repetitive and seemingly non functional behaviours that the person engages in. S/he may play with a toy or manipulate an object over and over. This behaviour could be harmless, merely interfering with the person’s routines, or it could be dangerous.

Difficulty making transitions

The need for consistency results in a tendency to react more strongly to changes or transitions in ways such as:

- Physical aggression
- Verbal aggression
- Property destruction
- Emotional outbursts or temper tantrums with a variety of associated behaviours
- Stubborn resistance
- Elopement (running away)
- Skin picking

These behaviours are usually related to obvious triggers such as a change in routine or transitions. Note that even when the change is a pleasant one – for example when staff suggest substituting a favourite activity for a chore – the person may react negatively.
**Emotional/Mental Health**

*Emotional Health*

People with PWS have what is referred to as “tenuous emotional control.” This means that they react rather quickly and strongly to events or stressors that most people can cope with. It is difficult for them to control their emotions.

It seems that these people are living with high levels of anxiety, which interferes with their ability to cope and requires them to have extreme consistency.

There is no research evidence which states the level of mental illness in people with Prader-Willi Syndrome. People with developmental disabilities in general have an approximate rate of 33% mental illness. (Nugent, 2005) It is reasonable to assume that the same rate probably applies to individuals with PWS, but this is just an assumption. In any case, it is fair to state that individuals with PWS have been documented with a full range of mental illnesses such as mood disorders, anxiety disorders, and personality disorders. Many of them will need mental health supports such as a psychiatrist.

**In Conclusion**

This Chapter has focused on the problems associated with PWS. Reading through a long list like this can become discouraging for parents and staff alike.

However, we can also consider that having all this information about PWS provides us with the opportunity to intervene at a young age and help people with PWS to deal more effectively with these concerns.
CHAPTER THREE:
SERVICE DELIVERY:
PHILOSOPHY and GENERAL GUIDELINES FOR PRACTICE

Having provided an overview of PWS, the Handbook will now move on to a discussion of service delivery.

This Chapter will introduce this topic, including a discussion of the philosophy of service delivery and some recommended general principles for all types of services.

**Philosophy**

Since the majority of individuals with PWS have a developmental delay, it is likely that they will be receiving supports from the developmental services system or related systems such as early intervention programmes and school boards.

Therefore, it is crucial that the issue of philosophy be addressed since supporting people with PWS requires a revision of the commonly accepted values in that system. The debate is between the individual’s right to make choices versus the agency’s responsibility to exercise due care.

The question agencies must answer is: **Do people with PWS have the right to eat what they want?**

**Human Rights**

On the one hand, the current developmental services system is strongly committed to a vision in which “all persons live in a state of dignity, share in all elements of living in the community, and have the opportunity to participate effectively.” (Community Living Ontario, 2007)

Essential to this vision is the concept of human rights and equity. Agencies now focus on “empowerment” in which people who are receiving services have the right to make informed decisions about all elements of their lives. Such decisions include what medical care they receive, where they live, where they work, who their friends are, and what services they participate in. Empowerment is the basis for the “person-centred” approach to service delivery through which all service planning is individualized and controlled by the person’s own goals for his/her life.

Another component of this vision is “the least restrictive alternative”. As applied to people with developmental disabilities, this concept states that people with developmental disabilities should have the least intrusive service or treatment that can effectively and safely address their needs and stated preferences. Intrusive means invasive or meddling. Therefore, agencies delivering services
according to the least restrictive alternative try to avoid interfering in people’s lives.

It follows from this philosophy that a person with PWS who is supported by an agency would be making all the decisions about his/her life including what to eat.

Health and Safety

On the other hand, agencies delivering services have the ethical and legal responsibility to provide care for all the individuals they support. Of course, this should be done in a manner which empowers these individuals. However, ultimately, the agency must ensure that they are maximizing the health and safety of their clients.

Can people with PWS make a rational, informed decision about their eating? The answer is “No”. Unfortunately, people with PWS experience an insatiable hunger that distorts their thinking and impairs their ability to make sound judgements. They would literally eat themselves to death in the sense that they would eat to the point of obesity with health complications that would result in death.

The agency is forced to manage the person’s diet and eating related behaviours in the individual’s best interests.

The Philosophy of Service for People with PWS

Service providers are required to modify their typical philosophy when serving people with PWS. Therefore, in answer to our previous question, people with PWS do NOT have the right to eat what they want. Given the life threatening consequences of the behaviours associated with PWS, agencies must exercise an extreme amount of control over food and access to it.

This is a significant departure from our usual philosophy, but it is necessary both legally, to fulfil obligations of due care, and ethically. However, within these strict guidelines, staff also strive to maximize choices and lessen restrictions whenever possible.

Substitute Decision Making

When discussing rights, it is also important to raise the issue of decision making on the part of adults with PWS who have developmental disabilities or cognitive limitations.
In general:

- Adults with PWS have the legal right to make their own decisions
- This right can only be removed from them by an official legal process
- This official legal process will differ depending on the legal system where the person lives
- The process always involves an assessment of the capacity of the person to make a decision based on his/her understanding of the implications of that decision
- An official representative of the government, such as a judge, will make the decision
- If a person is assessed as incapable of making his/her own decisions, a substitute decision maker is chosen
- The substitute decision maker is usually a relative, but can be a government body such as a Public Trustee if that is in the best interests of the individual
- The only individual or organization that can LEGALLY make a decision on behalf of the client is one that has been legally appointed to do this

This is important because there can be misunderstandings about who can make decisions on the part of the individual with PWS. **Families are NOT automatically the decision maker on behalf of their adult family member with PWS.** This can be a contentious issue between service providers and families.

If there is a valid concern about the person’s capacity to make decisions, then the agency or family can pursue a competency evaluation. Once again, the conflict between the typical service philosophy and the realities of supporting people with PWS comes to the surface. Under normal circumstances, the agency strives **not** to have the person’s rights removed. However, with PWS, this may be a necessity if the individual does not consent to life saving strategies.

Unless the adult has been legally declared incapable of decision making, that person has the right to make decisions about his/her care.

**General Guidelines for Practice**

While conducting research for this Handbook, the author was fortunate to have met with staff with several agencies that provide services to people of all ages with PWS. These service guidelines are based primarily on the useful and honest information provided by these staff.

The purpose of practice guidelines is to offer broad concepts that can act as the foundation for delivery of services to people with PWS of all ages and ability levels.
We should remember that these are general rules. There will be individual differences, based upon the person’s level of developmental disability, ability to control his/her own behaviour, age, and ability to tolerate changes.

Subsequent Chapters will illustrate how these guidelines are utilized in a variety of service settings.

**A Brief Summary of PWS**

Before discussing the service guidelines, it is useful to briefly recap the essential information from previous Chapters since this is the basis for these guidelines.

The key points about PWS are:

- A malfunction of the hypothalamus resulting in lack of control over key aspects of the person’s functioning including mood, appetite, growth, and behaviour
- An inability to feel full, combined with a drive to eat
- A need for consistency, structure, and predictability
- High levels of anxiety associated with change and uncertainty
- A level of developmental disability or restricted thinking patterns
- Behaviours such as compulsions, perseverations, food obsessive behaviours, and tantrums when drives are thwarted or change is required
- PWS is a life long condition and while people can achieve many goals, they will always need controlled environments

Therefore, people with PWS have an intense need for consistency, lower levels of self control, and a great deal of anxiety when their needs aren’t met. We must not underestimate the stress that people with PWS feel on a regular basis. Providing control and structure enables these individuals to increase their sense of self control and lowers their stress levels.

The following service guidelines flow logically from these significant characteristics.

**Service Guidelines**

*Guideline One: Plan Ahead*

Spontaneity is not an option for people with PWS. Planning ahead is essential.

- **All aspects** of the individual’s life must be pre planned. This includes daily routines, food, special events, and transitions. As staff, it is difficult to imagine how all encompassing this is. Everything should be controlled – this is something we are not used to and it takes time for staff to become proficient in doing this.
Individuals should be informed about what is going to happen and when.
- They should also be informed about changes in advance and the reason for changes should be explained. There is a question regarding how far ahead to inform people about change. This depends upon the individual and staff will realistically only learn this through experience. It is helpful to give some people a lot of notice when change is going to occur so they can get used to the idea. Other people get very upset and perseverate about the change, asking repetitive questions about it. For them, it is better to wait until quite close to the change before informing them.
- Changes in one area can impact upon all other areas.
- The purpose of this planning is to minimize anxiety on the part of the clients and the staff.

Guideline Two: Rules are a Requirement

Along with planning ahead, it is necessary to create a structure for activities based upon rules.

- Rules must be very specifically stated. They cannot be subtle. It cannot be assumed that everyone “knows”, based upon “common sense”, what must be done.
- It is helpful to express these rules in different ways. Along with verbally stating the rules, visual reminders are ideal, in particular with people who are more developmentally delayed. Visual reminders can include pictures, calendars, charts, etc.
- There should be clear consequences if rules aren’t followed and the consequences should be used all the time by everyone involved.
- Rules must be consistently applied. Allowing someone to break a rule without applying the consequences will have the result that the person will think s/he can break this rule in the future. The person will persist in wanting to break the rule for a very long time.
- Rules are required for even the most basic activities and for areas where we would not typically think to have them, such as where all the clients are going to sit in the van on an outing.
- Depending on the individual, put into place more sophisticated ways of involving him/her such as contracts or written forms.
- A lapse in rules and consistency in one area can affect other areas. For example, if a staff member allows an individual to have an extra food treat, the individual will start to question the rules for self care, hoarding, etc. It seems that people with PWS need to have total consistency and if this breaks down in one area, they need to test if it is breaking down in other areas.
- Use of rules can help to avoid power struggles. Rather than arguing, staff can calmly refer back to the rules and utilize the consequences. No discussion is required and no debate should take place. There should be no blaming, just a neutral application of consequences.
These rules are in place to decrease anxiety for everyone, minimize “bugging” behaviour, and increase a sense of control for the clients.

**Guideline Three: “See” the Environment with “Different Eyes” and Structure it to Decrease Problems**

Because people with PWS have a constant drive for food, and associated behaviours, staff need to learn to view the environment differently.

- All environments should be scanned for the presence of food products and related products such as food garbage, as well as non food products that the person might eat, such as cleaning or craft items. Try to see the environment from the perspective of a person with PWS.
- As well, all stimuli that might trigger an idea about food should be noted such as smells, pictures, conversations, etc.
- Before entering a new environment with an individual, try to visit it or at least imagine what food stimuli might be present there.
- Environments should be controlled so that exposure to food or food related stimuli are eliminated, or at least minimized. An example is covering a vending machine with a sheet at a day program site.
- Some environments might not be advisable for these individuals. For example, visiting a community restaurant might be just too difficult for them.
- Other environments can be modified so that people with PWS can be in them. Such modifications may require creativity on the part of staff.
- The other aspect of an environment that should be evaluated is its capacity to be consistent. For example, some environments are quite easily structured and can be comfortable for the person with PWS. Other environments are too chaotic and aren’t recommended since it is impossible to predict what might happen.

The purpose of environmental control is prevention of problems.

**Guideline Four: Support People to Increase their Ability to Control their Lives**

In spite of the overwhelming need to impose controls upon people with PWS, it is still possible to help them to achieve a measure of self control.

- Try to provide acceptable choices, for example “Would you like to plan to have an apple or applesauce for dinner on Tuesday?”
- Assist people to problem solve. Ask them to think ahead – “How can you stay within your calorie limit at your sister’s wedding?”
- Offer help. Ask what you as a staff person can do that would make it easier for them in a potentially difficult situation.
Allow for negotiations within clearly stated boundaries. “If you choose to have a small piece of cake at your sister’s wedding, you can give up your snack the next day.”

**In Conclusion**

It is crucial to emphasize how **structured** and **consistent** staff must be to work effectively with individuals who have PWS.

At first, this is not easy! New staff might feel a high level of stress since they are always “on their toes”, knowing that they have to follow the rules all the time and constantly be aware of the environment.

However, staff who have more experience in this specialized area have said that they have gotten used to working in this way. After a while, staff learn how to be consistent and manage the environment.

In fact, over time, this extremely structured approach actually lessens staff anxiety.

As well, staff report that they feel a great sense of satisfaction when they see the successes that the individuals experience with respect to weight loss, decrease in challenging behaviours, increase in energy, and a much more fulfilling life.
CHAPTER FOUR:
WEIGHT CONTROL: FOOD and EXERCISE

As we have discussed, weight control is a major focus of life for people with PWS. Excess weight is the cause of potentially lethal health conditions and challenging behaviours.

Therefore, it is important to devote a Handbook Chapter to the issue of controlling food and increasing exercise levels with people who have PWS.

The General Approach to Food Management

Food management is absolutely essential for people with PWS. This can be a challenge for staff and parents, but it is certainly a goal that can be achieved.

To emphasize the importance of food management, we must remember that a person with PWS needs only about 60% of the normal energy requirements (calories) of other people. (Balko, 2006) For example:

- A 20 year old woman without PWS needs 2200 calories a day
- A 20 year old woman with PWS needs 1300 calories a day for weight maintenance and 1000 – 1200 calories a day to lose weight
- If an individual with PWS eats an extra 500 calories per day, this could result in a weight gain of 48 pounds in a year!

Using a Menu System

All the successful PWS diets use a menu system. (Ontario Prader-Willi Syndrome Association, Prader-Willi Syndrome Association USA)

Menus allow everyone to know ahead of time what food/beverages will be provided and when this will happen. There are several important points to review:

- All menus should be prepared only after a doctor and nutritionist have been consulted. Each person’s health and caloric requirements are different.
- Menus should be prepared for a specific period of time. 3 weeks is a time period that is typically used.
- As much as possible, the individual with PWS should participate in menu planning.
- Choices are allowed within limits. For example, the person can choose between toast or oatmeal for breakfast on Wednesday next week.
- Menus cover all meals and snacks.
- Menus are posted for all to see.
- Once a menu is posted, it cannot be changed.
Negotiations for modifications of menu are done when the next set of new menus is being planned.

It works best to have one person in charge of food and menus. Clients should be directed to that person if they have a concern. This way, confusion is minimized and the person can’t play one staff member off against another.

Plan ahead – if there is an event coming up, build that into the menu. For example, if the individual is going to a movie, plan for his/her diet pop snack that day to be consumed at the movie theatre.

Menus must be maintained in all environments including group home, work place, parents’ home, community settings, and special events.

Just remember to keep to the menu. You are not being “a good guy” or “a kind person” by permitting someone to sneak in an extra treat or substitute one food for another at the table.

Instead, if you allow someone to convince you that one food change is okay, the person’s anxiety levels will increase. That person will bother you thereafter to make other food changes and s/he may start to pressure you to make changes in other routines and expectations that aren’t food related.

This menu system takes getting used to! Such rigid control over food is quite odd at first for staff and a little frightening.

Menu plans are included at the end of this Chapter. They indicate the structure required, but also demonstrate that individual variations are allowed.

However, staff report that after a while they become quite accustomed to a menu system. In fact, it has two key benefits:

- It eliminates arguing over food
- It lessens the anxiety over food that is felt by both staff and clients.

The Food

In our society, we actually consume quite a high number of calories every day and many of these are empty calories with little food value. Working with people who have PWS, you will soon notice how very controlled their food intake is.

- The menus prepared include all foods and beverages including condiments like ketchup and salad dressings, seasonings, and spices.
- Portions are tightly controlled – 1 tablespoon of ketchup means just that amount and no more.
- The brands are also controlled. For example, the brand of bread will be specified since each brand has a different caloric count.
There is no such thing as unlimited foods or beverages. The lack of limits creates too much anxiety. Even no-calorie foods or beverages are controlled.

For the most part, cooking using recipes is minimal unless the recipes can be made within the calorie restrictions.

Pre packaged foods can be a real asset. Buying yogurt in small containers allows staff to easily serve it that way, instead of measuring out yogurt from a large container. This leads to less chance of serving someone too much.

Grocery lists should very clearly specify what foods are to be purchased, including brand names and sizes of containers.

Be aware of how difficult it is to find healthy foods in the “outside world”. It is often better to take food along to activities.

Obviously, ensuring that people with PWS get the foods that they should in proper portion sizes is a challenge.

The Red Yellow Green Diet

The Red Yellow Green (RYG) Diet has proven to be an extremely successful approach for people who have PWS.

First developed in 1972 to be used with obese teens, the Red Yellow Green diet was adapted at the Children’s Institute in Pittsburgh for PWS. (The Children’s Institute, 2007) it has since been modified by North York General Hospital in Toronto. (Balko, 2006) Two excellent books on this diet are available:

- The New Red Yellow Green System for Weight Management. (Balko, 2007)
- The Weight Control Handbook. (Prader-Willi Syndrome Association USA, 2007)

This Chapter will provide a brief overview of the RYG diet, but staff and parents are advised to purchase a book on this topic for complete information.

Research by North York General Hospital indicates that parents who have used the RYG system reported good weight control, plus a reduction in the child’s request for food and temper tantrums concerning food. (Ibid)

The RYG system is effective because:

- It is structured
- It is written on a primary school level and is understandable for most clients
- It provides for choice, within the structure
- It uses everyday foods
- It ensures that all the food groups are represented – even people with PWS need some fat, protein, starches, etc.
- The parents or staff do not need to worry about setting up on their own a diet that is healthy. Following the RYG guidelines will ensure that the person’s diet is safe and lessen this daunting responsibility for them.
- It lends itself to visual presentation

The RYG system is built upon the “traffic light” concept:

- **RED – STOP.** These are high calorie foods that can only be eaten 4 times per month. Examples are fried foods, bacon, ice cream, cake, candy bars, etc. When the individual receives a red food, it is in a controlled portion.
- **YELLOW – CAUTION.** These are foods from all 5 food groups that are to be eaten every day. However, the serving sizes must be carefully monitored:
  - Meat – 75 calories per serving
  - Bread – 70 calories per serving
  - Fruit – 80 calories per serving
  - Milk – 100 calories per serving
  - Fat – 50 calories per serving
- **GREEN – GO.** These are foods with low or no calories. Larger portions are allowed and this raises the level of both physical and emotional satisfaction. Portion sizes are still controlled, to avoid the unlimited concept.
  - Carrots, tomatoes, tossed salad, asparagus – 1 cup
  - Tea, coffee, diet drinks – 1 cup
  - Water – 1 cup. Water is healthy and can be made more appealing by ensuring it is chilled, using a special cup/straw, or adding lemon/lime juice.
  - Be aware of the potential for water intoxication, a serious condition in which the person drinks too much water too quickly. The condition has symptoms that resemble being drunk, which can then move to symptoms of seizures, coma, and ultimately death. People should drink no more that 16 ounces (2 cups) of water at any one time.

Because of the low calorie intake in the RYG system, vitamins are usually recommended but this should be done in consultation with a doctor.

**The RYG diet is a life long approach to eating for people with PWS.** Once the individual has achieved a healthy weight, daily calorie intake can be modestly increased. However, the structure of the menu system has to be adhered to or weight gain will occur.
All in all, the RYG system provides the life long security and dietary controls that are necessary for people with PWS.

*Restructuring the Environment*

If the RYG diet is maintained, there should be very good results with respect to weight loss or maintenance. Not only is diet controlled using the RYG system, the environment is also controlled to minimize food related problems.

- Food is served on smaller plates, to give the illusion of sufficient amounts.
- All food is pre served on the individuals’ plates and brought to the table ready to be eaten.
- No serving dishes, pots, or pitchers are brought to the table.
- All condiments are also brought to the table in a pre served form. Ketchup is in a small cup, salad dressings can be in small packages such as used at fast food restaurants, sugar is in little envelopes, etc.
- Containers for condiments and seasonings are not brought to the table.
- All meals/snacks are served at the table
- Schedules are established for meals – when the time is done, the eating is done.
- Food is locked away. This includes the refrigerator, cupboards, pantries, freezers, kitchen door, staff lunch boxes, etc.
- Individuals’ rooms are locked to prevent scrounging.
- Access to money is restricted and all money is locked away. This includes petty cash and people’s purses or wallets.
- People who are in the environment are not allowed to bring in extra food or food related products such as a coffee, cough drops, a bottle of water – no Tim Horton’s or Starbucks for the staff!
- Lingering smells of food should be eliminated.
- Visual reminders of food should be eliminated – magazines, books, etc.
- Conversations should not focus on food – no discussions of the food at a recent party, restaurants, cooking a new recipe at home, etc.
- Try to choose environments where you can exercise some control – don’t go to a fall fair with food booths all around, go to a craft show where food is usually available in a few avoidable locations.
- **These controls are needed in all environments.**

Three photos of a typical meal are displayed at the end of this Chapter. Please note how very tightly controlled the food is, how small the actual portion of protein is, and how the “green” foods increase the volume and appeal of the meal. Also note that the individual is actually receiving a considerable amount of food for dinner. No one is “starving” and the food is very healthy.
A Change in Culture – De-emphasizing Food

Perhaps the most challenging aspect of this approach to food and eating is the fact that we must change the way that our culture approaches food. Food is an important element of so much that we do. For the sake of the individual with PWS, this has to be adapted:

- Meals must become very functional and the focus should be on eating.
- Socializing and fun should not occur during meals. Family discussions, planning future activities, and reviewing the day’s events should be done away from the table. This pattern can be started when the individual is a young child so that it is established by adulthood.
- Food should not be the main purpose of activities such as going to the movies. Talk about the other aspects of events, such as the plot of the movie you are going to see or whether other people have liked it. Point out other aspects of the movie theatre such as the people there or posters for other movies. Afterwards, review the movie and discuss people’s opinions of it. This process will help to broaden people’s perspectives beyond the food at the theatre.
- Food should also be downplayed as a focus of celebrations or events such as Christmas or birthdays. At birthdays, build in fun activities and don’t focus on the birthday treats. At Christmas, decorate a tree, go for a walk to look at the Christmas lights in the neighbourhood, or make presents for others.
- Cooking cannot be a hobby or topic of interest. Look to other sources of entertainment and pleasure.

This is not an easy issue. Staff and parents must learn to rethink their own lifestyles. As well, we need to enlist the support of others in this process such as family members, staff in other settings, and people in the community.

Exercise

Although restricting food intake is a key component of weight control, the other aspect to remember is increasing activity levels. Motivating the individual with PWS can be challenging. However, stretching, aerobics, and strength training should be part of the individual’s day. (Goldband & Pividor, in print)

Ensure that the individual is drinking enough water during exercise. Since people with PWS may not enjoy drinking water, they may not voluntarily drink during exercise. However, hydration (having the correct fluid balance in the body) is important so the strategies discussed earlier in this Chapter can be used to make the water more appetizing.
Establishing Routines for Exercise

Regular exercise must be a part of the person’s daily routine. This will assist with weight loss/maintenance and other issues such as muscle tone.

**Exercise should be done with the approval of a physician**, especially for people with PWS who may be obese and not be active. There are several medical issues associated with PWS that can affect the person’s participation in exercise such as lax muscle tone, flat feet, osteoporosis, water retention in the legs, or lethargy. The exercise program is often done in stages to accommodate the person’s weight loss and growing stamina.

Exercise should NOT be a punishment. It should be an expectation. Involve the individual in choosing activities that they like as much as possible. For example, one person may prefer a tread mill while another would rather walk briskly outside.

A variety of exercises can be considered:

- Walking
- Swimming
- Riding bikes
- Sports
- Exercise bikes
- Elliptical trainers
- Hippotherapy (horseback riding)
- Exercise videos
- Special Olympics

Create Environments and Activities that Increase Exercise

There are many creative ways that we can subtly add exercise to the person’s life:

- Create environments that require activity. For example, put the TV in the basement.
- Plan activities that require activity such as going to the gym or walking through a museum.
- Add in opportunities for extra exercise during the daily routines. Park further away from the store to increase walking, ask people to help with carrying the laundry downstairs, wash the van, or walk over to the library.
- Have interesting and well maintained exercise equipment where the person lives.

Once you start to think about this, there will be endless possibilities that you will create.
In Conclusion

In this Chapter, we have presented the key component to supporting people with PWS through food management and exercise.

Fortunately, there are many examples of how this can be done successfully for staff to use as a starting point.

Although this will seem very rigid to you at first, you will see results for the people you work with and this will be exciting and encouraging!
Sample Meals

Breakfast:

- Small portion of hot cereal, packet of stevia (sweetener)
- Toast with no butter, measured portion of jam (sugar free)
- ½ cup peaches
- Skim milk, water
Lunch:

- Small portion of fish (2 oz)
- Carrots, ½ cup corn
- Salad, packet of salad dressing (flax seed oil)
- Measured condiments (mustard, ketchup, butter substitute, salt, pepper)
- Juicy jello cup (no aspartame)
- Orange
- Skim milk, water
Supper:

- Small portion of chicken
- Carrots, salad with measured salad dressing
- Baked potato, with measured portion of butter substitute (Molly Mcbutter)
- Packet of ketchup, mustard, packet of stevia, salt, pepper
- Applesauce, Jello
- Skim milk, water
- Sometimes have: Mrs. Dash spice - 2 teaspoon portion
  Garlic – 2 teaspoon portion
**BREAKFAST: WEEK 1**

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*For CR and DN: Use only weight watchers Diet Bread*

**For others: Use a variety of whole grain breads-Stone Ground, Bran Bread**

***1 tbsp of ketchup for eggs and meat***

****DP receives decaf coffee with 1 sweetener @ breakfast M-F, CR and DN receive their coffee @ 9am at their Day Programs**
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# LUNCH AND SNACK: WEEK 1

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<td>CRYSTAL LITE</td>
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<td>TUNA (1/4 C) WITH</td>
<td>PEANUT BUTTER</td>
<td>EGG (1) SALAD</td>
<td>TURKEY (1 OZ)</td>
<td>HAM (1 OZ)</td>
<td>GRILLED SKIM</td>
<td>BEEF, BEAN AND</td>
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<tr>
<td>LETTUCE SANDWICH</td>
<td>SANDWICH</td>
<td>WITH LETTUCE</td>
<td>LETTUCE AND FAT</td>
<td>WITH LETTUCE AND</td>
<td>MILK CHEESE</td>
<td>VEGIE SOUP (1 C)</td>
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<td>WITH LETTUCE</td>
<td>FREE MAYO</td>
<td>MUSTARD</td>
<td>(1 SLICE)</td>
<td>COLD CUT</td>
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<tr>
<td>TOSSED SALAD 1C</td>
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<td>GRAPES (1/2 C)</td>
<td>APPLESAUCE (1/2 C)</td>
<td>PEAR (1)</td>
<td>APPLE (1)</td>
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<td>SNACK 3PM DP ONLY</td>
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<td>LITE POPCORN (1C)</td>
<td>ARROWROOT</td>
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<td>DP ONLY MINI RICE CAKES</td>
<td>DP ONLY GRAIN SALTINES</td>
<td>DP ONLY ARROWROOTS (2)</td>
<td>DP ONLY LOW FAT WHEAT THINS (8)</td>
<td>DP ONLY GRAHAM WAFERS (2)</td>
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*For CR and DN: Use only weight watchers Diet Bread*

**For others: Use a variety of whole grain breads-Stone Ground, Bran Bread**

***1 tbsp of fat free salad dressing, ketchup, relish, Kraft Ultra Mayo and salsa.***
<table>
<thead>
<tr>
<th></th>
<th>MONDAY</th>
<th>TUESDAY</th>
<th>WEDNESDAY</th>
<th>THURSDAY</th>
<th>FRIDAY</th>
<th>SATURDAY</th>
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<tbody>
<tr>
<td>10 AM SNACK</td>
<td>SKIM MILK CHEESE SLICE OR YOGURT</td>
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<td>LUNCH 12:00 PM</td>
<td>SUGAR-FREE CRYSTAL LITE (8OZ)</td>
<td>SUGAR-FREE CRYSTAL LITE (8OZ)</td>
<td>SUGAR-FREE CRYSTAL LITE (8OZ)</td>
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<tr>
<td>CR/DN: TOSSED SALAD 1C W/ 1TBS PAT FFF DRESSING</td>
<td>CR/DN: TOSSED SALAD 1C W/ 1TBS PAT FFF DRESSING</td>
<td>CR/DN: TOSSED SALAD 1C W/ 1TBS PAT FFF DRESSING</td>
<td>CR/DN: TOSSED SALAD 1C W/ 1TBS PAT FFF DRESSING</td>
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<td>CR/DN: TOSSED SALAD 1C W/ 1TBS PAT FFF DRESSING</td>
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<tr>
<td>GRAPE (1/2 C)</td>
<td>PEAR (1)</td>
<td>PLUM (1)</td>
<td>(1/2 C) APPLESAUCE</td>
<td>ORANGE (1)</td>
<td>BANANA (1)</td>
<td>APPLE (1)</td>
<td>BANANA (1)</td>
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<td>SNACK 2 PM</td>
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<tr>
<td>DP ONLY</td>
<td>MINI RICE CAKES (6)</td>
<td>DP ONLY</td>
<td>GRAIN SALTINES (6)</td>
<td>DP ONLY</td>
<td>ARROWROOTS (2)</td>
<td>LOW FAT WHEAT THINS (8)</td>
<td>DP ONLY</td>
</tr>
</tbody>
</table>

*For CR and DN: Use only weight watchers Diet Bread*

**For others: Use a variety of whole grain breads-Stone Ground, Bran Bread**

***1 tbsp of fat free salad dressing, ketchup, relish, Kraft Ultra Mayo and salsa.***

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33
# DINNER: WEEK 1

<table>
<thead>
<tr>
<th>MONDAY</th>
<th>TUESDAY</th>
<th>WEDNESDAY</th>
<th>THURSDAY</th>
<th>FRIDAY</th>
<th>SATURDAY</th>
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<tbody>
<tr>
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<tr>
<td>CRYSTAL LITE (8 OZ)</td>
<td>DECAF OR HERBAL TEA (8 OZ)</td>
<td>CRYSTAL LITE (8 OZ)</td>
<td>DECAF OR HERBAL TEA (8 OZ)</td>
<td>CAFFEINE FREE DIET POP (8 OZ)</td>
<td>CRYSTAL LITE (8 OZ)</td>
<td>CRYSTAL LITE (8 OZ)</td>
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<tr>
<td><strong>DINNER 5:30PM</strong></td>
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</tr>
<tr>
<td>BBQ BAKED CHICKEN BREAST (3OZ)</td>
<td>SEASONED POTATO WEDGES (1 C)</td>
<td>HERBED RICE (½ C)</td>
<td>HERBED RICE (½ C)</td>
<td>HAMBURGER PATTY (1-3OZ)</td>
<td>(3OZ) PORK LOIN W/ APPLESAUCE FAT FREE GRAVY (1 TBSP)</td>
<td>MASHED POTATOES (½ C W/ NO FAT)</td>
</tr>
<tr>
<td><strong>SEASONED POTATO WEDGES (1 C)</strong></td>
<td><strong>HERBED RICE (½ C)</strong></td>
<td>(1 MED) POTATOE WITH FAT FREE SOUR CREAM &amp; SALSA (1 TBSPX2)</td>
<td>BOILED POTATOES WITH PARSLEY (1 MED)</td>
<td>HAMBURGER BUNS (1)</td>
<td>(STARCH IS IN NOODLE)</td>
<td>GREEN BEANS (1/2 CUPS)</td>
</tr>
<tr>
<td><strong>GREEN PEAS (1/2C)</strong></td>
<td><strong>GREEN BEANS W/ MUSHROOMS (1/2C)</strong></td>
<td><strong>DICED BUTTERNUT SQUASH (1/2C)</strong></td>
<td><strong>SLICED TOMATO, ONION W/ KETCHUP, MUSTARD, RELISH</strong></td>
<td><strong>BROCCOLI (1/2 C)</strong></td>
<td><strong>GREEN BEANS (1/2 CUPS)</strong></td>
<td><strong>GREEN BEANS (1/2 CUPS)</strong></td>
</tr>
<tr>
<td><strong>COLE SLAW W/ FAT FREE MAYO (1C)</strong></td>
<td><strong>(1 C) ROMAINE SALAD W/ FAT FREE DRESSING</strong></td>
<td><strong>SPINACH SALAD (1C) W/ FAT FREE DRESSING</strong></td>
<td><strong>COLE SLAW W/ DICED APPLE &amp; FAT FREE MAYO (1C)</strong></td>
<td><strong>TOSSED SALAD (1C) W/ FAT FREE DRESSING</strong></td>
<td><strong>CEASAR SALAD (1C) W/ FAT FREE DRESSING</strong></td>
<td><strong>TOSSED SALAD (1C) W/ FAT FREE DRESSING</strong></td>
</tr>
<tr>
<td><strong>SKIM MILK (250 ML)</strong></td>
<td><strong>SKIM MILK (250 ML)</strong></td>
<td><strong>SKIM MILK (250 ML)</strong></td>
<td><strong>SKIM MILK (250 ML)</strong></td>
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<td><strong>BEDTIME SNACK</strong></td>
<td><strong>BEDTIME SNACK</strong></td>
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<tr>
<td><strong>JELLIED FRUIT COCKTAIL (3/4 C)</strong></td>
<td><strong>LITE VANILLA PUDDING W/ SPRINKLES (1/2 C)</strong></td>
<td><strong>LITE STRAWBERRY MOUSSE (1/2 C)</strong></td>
<td><strong>LITE JELLO (1/2 C)</strong></td>
<td><strong>LITE JELLO (1/2 C)</strong></td>
<td><strong>FAT FREE ASTRO YOGURT 175g</strong></td>
<td><strong>FAT FREE FROZEN CHAMPIONS RAINBOW SORBET (1/2 C)</strong></td>
</tr>
</tbody>
</table>

*Three (3) ounces (OZ) of meat*

**1 tbsp of low fat salad dressing, ketchup, relish, Kraft Ultra Mayo**

***Light low fat popcorn***
CHAPTER FIVE: BEHAVIOURAL STRATEGIES

Unfortunately, PWS is associated with a number of ongoing challenging behaviours. The most common of these behaviours are (Linder, 2002):

- Food-obsessive behaviour
- Temper tantrums
- Non-compliance (not following through on requests or expectations)
- Attention seeking behaviours

Many behavioural strategies have been used successfully with PWS to reduce these challenging behaviours and increase appropriate behaviours.

This Chapter will review functional behaviour assessment, an all-purpose behavioural approach that is very practical and helpful. Then, there will be a discussion of general behavioural strategies to use with people who have PWS. The Chapter will conclude by listing some specific techniques that are extremely helpful with PWS.

**Functional Behaviour Assessment: A Review**

Functional Behaviour Assessment is a method of behavioural programming which assists us to understand why behaviour is occurring and to use this knowledge to help people to change their behaviours. (Cooper et al, 2007; Nugent, 2006a)

*The ABC’s of Behaviour*

Understanding of behaviour starts with observation. We observe the various situations in which the behaviour happens, keeping clear records of the events surrounding the behaviour. In behavioural language, we observe the sequence of events. This sequence is divided into 3 components, the “ABC’s”.

The **Antecedents**: all the events and stimuli that occur **before** the behaviour

The **Behaviour itself**: the specific behaviour we wish to change

The **Consequences**: what happens **after** the behaviour and is a result of it

By identifying the sequence, the ABC’s, we can start to develop some ideas about why this behaviour is happening. These ideas are then used to create strategies or plans to help the person to change his/her behaviour.

**The Antecedents**

Sometimes, by working with antecedents we can eliminate or drastically decrease the behaviour.
The antecedents occur **before** the behaviour and basically cause it to occur. They cover a wide range of stimuli and their effect will differ depending on the individual.

**What are Antecedents?**

Just about anything can be an antecedent.

- Antecedents can be stimuli or events outside the individual. This includes how other people behave, the events that are going on, the schedules, the routines, or the actual physical environment. Therefore, the temperature in the room, the actions of other students, and the fact that there is an exam going on can all influence the behaviour of a student in a classroom.
- Antecedents can be stimuli or conditions inside the individual. This includes physical health, mental health, skills (or lack of skills), and personality.

**When do Antecedents Occur?**

- Antecedents can occur just before the behaviour happens and can set the behaviour off. This type of antecedent is referred to as a **trigger**, since it actually triggers the behaviour.
- Antecedents can also occur minutes, hours, days, weeks, months or even years before the behaviour. This type of antecedent is called a **setting condition** since it sets up the probability that the behaviour could occur. It can also be called a **contributory condition** since it adds to the probability that the behaviour could occur. Setting or contributory conditions increase the person’s vulnerability to challenging behaviour.

**Antecedents and PWS**

There are a vast number of antecedents associated with PWS. The contributory or setting conditions have been listed in previous chapters, but the following chart reminds us of a summary of the key factors.

<table>
<thead>
<tr>
<th>Physical Health Factors</th>
<th>Mental Health Factors</th>
<th>Cognitive Skills/Patterns</th>
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</thead>
<tbody>
<tr>
<td>Insatiable hunger</td>
<td>Anxiety</td>
<td>Developmental delays</td>
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<tr>
<td>Sleep apnea</td>
<td>Obsessions</td>
<td>Poor short term memory</td>
</tr>
<tr>
<td>Poor muscle tone</td>
<td>Emotional extremes</td>
<td>Difficulties with abstract thinking</td>
</tr>
<tr>
<td>Dental problems</td>
<td>Loss of control</td>
<td>Difficulties with auditory processing</td>
</tr>
<tr>
<td>Stomach problems</td>
<td>“Acting out” behaviours</td>
<td>Poor math skills</td>
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<tr>
<td>High pain threshold</td>
<td></td>
<td>Rigid thought processes</td>
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<tr>
<td>Tendency to tire easily</td>
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<td></td>
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<tr>
<td>Lack of sexual development</td>
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<td>Scoliosis</td>
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As well, the individual with PWS is very sensitive to the environment:

- Exceedingly aware of and easily disturbed by food related stimuli – seeing food or pictures related to food, smelling food, and hearing conversations about food are examples of this.
- Highly responsive to and upset by change – transitions, alterations in menu, changes in routines or people, etc.

We must remember that an individual with PWS carries these personal antecedents with him/her all the time. They influence how s/he is thinking, feeling, and reacting to the environment throughout his/her life.

Working with Antecedents

Basically, there are 3 ways that we can work on antecedents:

- **Eliminate the antecedent**: In some cases, it is helpful and acceptable to remove the antecedent. Examples of this are removing food smells, stopping abuse by staff, or providing medications to eliminate pain.
- **Modify the antecedent so the person can cope with it**: In these situations, the antecedent can be adapted so the person is no longer as upset by it. Examples in this case are changing the schedule so the person has time for a nap in the afternoon and can then cope with exercising or using humour when asking the person to do a chore.
- **Help the person to cope with the antecedent**: In some cases, the antecedent can not be eliminated; it is a fact of life. We try to help the person to cope better. Strategies in this category include warning the person ahead of time when a transition is coming or teaching the person a skill that will increase his/her ability to carry out a chore.

With PWS, a great deal can be accomplished by dealing with antecedent factors.

The Consequences

Consequences occur after the behaviour occurs. They determine whether the behaviour will be repeated. If the person likes the consequence, s/he will be more likely to repeat the behaviour.

There are two types of consequences, reinforcement and punishment.

**Reinforcement**

Consequences that make the behaviour more likely to be repeated are called reinforcement.
Key points about reinforcement are:

- Reinforcement is highly individualized. What you may like, I may not like. We can’t assume that something will be reinforcing for a client. The only way we can tell if something is reinforcing is by observing what happens to the person’s behaviour after we use that consequence.
- Reinforcement can be an object (like food), an activity (like watching a preferred TV show), or a person (spending time with a favourite staff).
- Reinforcement can consist of giving someone something they want. This is called positive reinforcement.
- Reinforcement can also consist of taking away something the person doesn’t like. This is called negative reinforcement. Examples of this are allowing the person to avoid doing a chore or removing an unpleasant room mate.

Punishment

*The use of punishment is controversial.* When discussing behavioural approaches in this Handbook, it is important to include punishment because:

- Punishment does exist – we can’t ignore it
- Punishment is used by some services so we should understand what it is and why it isn’t the best choice

*We want to stress that this Handbook is not recommending that punishment be used with people who have PWS.* We have included it in this Chapter for educational purposes.

The decision to use punishment should be made by a team with proper clinical supervision and based upon comprehensive data.

Punishment is defined as a consequence that stops the behaviour or makes it less likely to repeat. Please note that the word punishment has different meanings in the English language. It carries a sense of being bad and something dreadful happening to the person. In behavioural terms, it is simply a consequence that decreases the probability that the behaviour will repeat.

The same points apply for punishment as for reinforcement:

- Punishment is also highly individualized. What you may not like, I may like. We can’t assume that something will be punishing for a client. The only way we can tell if something is punishing is by observing what happens to the person’s behaviour after we use that consequence.
- Punishment can be an object (like a hated food), an activity (like exercising), or a person (a peer who teases)
Punishment can consist of giving someone something they don’t want. This is called **positive punishment**. A slap or a chore can be a punishment. The word “positive” does not mean that the punishment is a good thing; it just means that something is given to or added to the situation.

Punishment can also consist of taking away something the person likes. This is called **negative punishment**. Examples of this are grounding a teenager or removing someone’s allowance. Similarly, the word “negative” does not mean that the punishment is a bad thing; it means that something is taken away from the person.

*Using Consequences to Change Behaviour: Reinforcement as the Preferred Strategy*

Changing consequences is another way to change behaviours, along with the previously discussed strategy of changing antecedents. Research indicates that using reinforcement is a more effective way to work with behaviours (Griffiths, Gardner, & Nugent, 1998):

**Punishment may work in the short term, but in the long run reinforcement is more effective.** Reinforcement teaches people what behaviours we want by reinforcing these. Punishment teaches people what not to do, but doesn’t help them to learn how to cope. Punishment is also very unpleasant and often unethical. Using punishment can ruin the relationship between the individual and staff.

Consequences must be interpreted in light of the person’s subsequent behaviour. **If you use a consequence and the behaviour repeats, then that consequence is reinforcing.**

This can be confusing. Sometimes, we think we are punishing a person but the behaviour repeats. Therefore, in spite of our intentions, we were actually reinforcing the behaviour.

A good example of this is “time out”. If we are **continually** putting a person in time out because the challenging behaviour is being repeated, that person is not being punished! That person is actually being reinforced or s/he would not repeat the behaviour. Observe the situation carefully and you may find that the person likes time out because it lets him/her escape an unpleasant environment, an annoying person, over stimulation, or a disliked activity.

**Developing a Behaviour Plan**

A behaviour plan is individualized. Sometimes a group, like a classroom of students or all the residents in a group home, may share a plan. However, a group plan would only make sense when the functions of the target behaviour are
the same for each member of the group. Sometimes, a portion of a plan may be
shared in an instance when there are common issues and the rest of the plan is
individualized. In any case, plans should be reviewed on an individual basis to
make sure that each plan is appropriate for all the individuals involved.

Developing a behaviour plan is a basic process:

**Step One:** Choose the “target behaviour” which is the behaviour you want to
change. The plan could be designed to increase the behaviour, like doing
homework, or decrease the behaviour, like tantrums. The target behaviour should
be one which is in the individual’s best interests.

**Step Two:** Conduct the ABC observations. Notice what happens before and after
the behaviour. Keep thorough records of what you see. Remember to include
antecedents that are ongoing, such as health issues, as well as the triggers.

**Step Three:** Decide what factors (antecedents and consequences) appear to be
contributing to the behaviours.

**Step Four:** Develop a plan that describes how antecedents and consequences will
be changed to influence the behaviour. The plan should be very specific and
include who will do what by when. For example, if you decide to decrease the
noise in the environment, indicate which staff member is responsible for carrying
out this idea and the deadline for finishing this.

**Step Five:** Record what happens to the behaviour after you put your plan into
effect.

**Step Six:** Evaluate the success of your plan. Review what has happened to the
behaviour and decide if you will continue with your current strategies or modify
them.

This is a very brief overview of the strategy. More comprehensive information is
available in other sources. (Cooper, 2007; Nugent, 2006a)

Basically, a behaviour plan will work on changing the antecedents and
consequences that are influencing the behaviours.

**Working with Antecedents in a PWS Behaviour Plan**

**Specific Elements in a PWS Behaviour Plan**

When working with people who have PWS, there are consistent
antecedent/consequence elements that must be included.
Deal with Physical Health Issues

Chapter Two reviewed the many health problems that people with PWS encounter throughout their lives. Medical issues are very powerful antecedents since they affect the person’s ability to deal with stresses and other environmental factors.

For example, a man may cope quite well with doing a chore on a day-to-day basis. However, if he is constipated, he may be in pain and just generally feel unhappy. This may be enough to erode his coping abilities to the point where he gets upset and becomes aggressive when he is asked to do his chore.

Please keep in mind that health issues also include medications. Any drug has side effects. Some side effects are minimal, but others can be quite negative and have an impact on behaviours. For example, anti-inflammatory medications can upset the digestive system of some people. We should carefully monitor for side effects of medications. Also, remember that over-the-counter medications can have side effects just as much as prescribed medications can.

We should do all that we can to lessen the impact of health problems. We may not be able to make them disappear, but we can help by carefully monitoring health, getting medical assistance, ensuring that regular medical exams are conducted, watching for side effects of medications, providing pain relief, etc.

Dealing with medical issues is the ethical thing to do. It is also practical, since it helps to decrease challenging behaviours.

Deal with Mental Health Issues

Some people with PWS will have mental illnesses. It is important that we obtain supports for them since mental illness also affects the person’s ability to cope with life.

As well, specific mental illnesses are accompanied by symptoms such as aggression, self-injury, or excessive restlessness. (Nugent, 2005) It is not within the scope of this Handbook to discuss in detail how to support people with dual diagnosis. In general, it is recommended that (Ibid):

- Careful observations be conducted to note any potential symptoms of mental illnesses
- Appropriate assessments and services be obtained
- The impact of mental illness be considered in any behaviour planning
**Have Structure for all Aspects of the Person’s Life**

The best way to eliminate arguments and tantrums is to have rules for all aspects of the individual’s life. This may seem oppressive, but it decreases anxiety and supports consistency. Rules should include:

- What behaviour is expected under which circumstances
- Time limits for activities
- Consequences of behaviours

**Control the Environment with respect to Food**

As was discussed in Chapter Four, there are many ways to control the environment to lessen stress and decrease food-obsessive behaviours. The main focus should be to remove anything that could remind clients of food and to lock up food and money.

**Always have Staff Present when Individuals are Eating**

One non negotiable rule is to always have a staff person present when clients are eating.

This system should be adhered to whether they are having a meal or a snack. Staff can make sure that eating is done in a calm setting without fighting or stealing.

**Working with Consequences with a PWS Behaviour Plan**

There are some important elements to review with respect to consequences and PWS.

**Should Food be Used as Reinforcement?**

This is a controversial question!

On the one hand, people have stated that food is such a significant issue for PWS, that using food as reinforcement makes no sense. It focuses even more attention on food.

On the other hand, for some people food is a very powerful motivator.

It seems that the best answer to this question is: “It depends”. For some people, food is a valuable reinforcement. We need to know the individual well to know whether that person will respond positively to food reinforcement. We may even need to test out food as reinforcement before we are sure about its effectiveness.
In any case, if food is reinforcement, only low or no calorie items such as diet pop or sugarless candies are used. The calories should be included in the daily allocation and the amount of food reinforcement should be strictly controlled. A person might be able to gain 1 candy per day as reinforcement for doing chores, but s/he should not be allowed to get many candies per day each time s/he completes 1 chore. This would be too many “snacks” per day and would eventually disrupt the concept of a structured, limited number of times in a day to consume food or drinks.

*Be Consistent*

Because people with PWS require structure to lessen their anxieties, consequences should be applied consistently according to the already established agreement. The nature of the consequences should not be changed. If someone is to receive a stick of sugarless gum as reinforcement, this should not be replaced with a diet pop.

**General Behavioural Strategies**

General behavioural strategies are ideas that staff and parents can use in any situations when supporting people with PWS. These strategies will be helpful with all the behaviour problems that are usually associated with PWS and any other behaviour that individuals may display.

*Strategy One: Be Consistent*

Consistency is the key! The individual staff person must behave the same way all the time. Have the identical expectations of the person, use identical consequences for specific behaviours, and coordinate with others to ensure that they are synchronized with you.

*Strategy Two: Keep Records of Everything*

Services that have been successful with people who have PWS have emphasized the need to record everything. Forms should be developed for staff to track food consumption, weight, health issues, shopping purchases, clients’ routines, safety procedures, etc. As well, data with respect to specific behaviours should be kept.

Having records provides a sense of stability and consistency. Recording behavioural data is necessary for proper monitoring of behavioural programming.
Strategy Three: Be a Positive Person

Staff should try to be positive people who are naturally reinforcing and warm. All human beings like to be reinforced. Your attention and support will be more meaningful if you are pleasant.

Use of humour can make things go more smoothly. Joke about doing a chore or see the positive side in situations. It may take a while, but you will see that clients will respond to your positive approaches.

Strategy Four: Control your Emotions

A person with PWS can be infuriating. S/he may perseverate by asking a question many, many times. S/he may “test” a staff to find out if that staff member can be persuaded to change a routine. These behaviours are understandably upsetting for staff.

A recommended approach is to try to be calm. Remain pleasant and neutral. Don’t blame the person, don’t get angry, and don’t allow the person to see that you are getting upset. Also, watch your body language. It is necessary to control not only your words, but also your facial expression, your tone of voice, and your body.

If a “mistake” is made and the person does not behave appropriately, calmly explain what went wrong and apply the consequences. Outline the steps needed to make things go right the next time. Avoid blaming.

Strategy Five: Get Help if you are Becoming Upset

We are all human. In spite of our best intentions, we get upset. If you find you are getting angry or losing control, it is perfectly acceptable to ask other staff to take over for you for a while. This gives you a break and allows you to return to the person with renewed energy and self control.

Strategy Six: Minimize Discussion, Don’t Negotiate, Don’t Give In, Avoid Power Struggles

The most successful approach for a staff person is to stick with the rules and plans that have been developed. Try not to be enticed into discussions about the expectations, just calmly repeat what people are required to do. NEVER give in, even if the person is persistently asking you to change the rules. Once you give in, the person will continually ask for more concessions.

There is no reason to engage in a power struggle with the individual – just stay with the plan.
**Strategy Seven: Ensure that People Understand the Rules and Expectations**

If we expect clients to follow the rules, we should make sure that they have all the information about what is required of them. List all the rules and consequences in simple language and post the list where it is clearly visible. If it is helpful, use pictures to explain the rules as well.

It is important to provide the opportunity for people to have choices when the rules are being developed. However, once the plan is finalized, no changes are made.

Ask people to repeat the rules back to you so you are sure that they understand them.

**Strategy Eight: Teach People what to do, Rather than Telling Them what not to do**

This is a somewhat complicated concept but it is important:

- Usually, we focus on telling people what NOT to do
- This is not always effective because it leaves them at a loss – they don’t know what else to do instead

It is more helpful to tell them what to do, i.e. what behaviour is expected of them. This allows the individual to learn a new skill and have a clear idea of what the requirements are. For example, instead of telling someone to “stop eating so quickly” we can ask the person to “take 1 spoonful, count 30, and take another spoonful”. This makes it easier for the person to learn the new behaviours.

**Strategy Nine: Teach Replacement Behaviours**

Not only do we want to tell people what to do, it is very useful to teach them new behaviours that will actually take the place of the challenging behaviours. These are referred to as replacement behaviours. Replacement behaviours should have the same general purpose as the challenging behaviour.

For example, we may have a person who picks at the skin on her hands when she is bored. If we teach this woman to knit or crochet, she will be doing something else with her hands, she won’t be bored, and she won’t be able to pick at her skin.

The skin picking behaviour has been replaced by a socially acceptable and safe skill that meets the need for something to do.

**Strategy Ten: Provide Opportunities for People to Practice Behaviours**

Since people with PWS have limited cognitive skills, we cannot rely on verbal instruction alone when teaching behaviours.
Ensure that the individual has really understood what is expected:

- Describe the required behaviour
- Demonstrate the behaviour
- Ask the person to demonstrate the behaviour
- Don’t expect the person to be perfect on his/her first try – reinforce his/her efforts and help him to gradually become more accurate with the behaviour
- Post reminders of how the behaviour is done, including visual cues if that is helpful

An example of this is relaxation training. Relaxation training can include physical techniques such as deep breathing. It can include mental techniques such as imagining a pleasant situation. It can include environmental modifications such as leaving the room or putting on favourite music. Thus, relaxation training is complex. Staff should ensure that the person understands what is involved and practices the components.

**Dealing with Crisis Situations**

A crisis is a situation in which the person’s behaviour is a serious threat to self or others. Examples are:

- Aggression towards others
- Self injury
- Property destruction such as putting a fist through the wall or throwing furniture

Implied in the word crisis is the concept of loss of control by the person over his/her behaviour.

This Handbook will not provide a complete procedure for dealing with crisis. Staff working with people who have PWS should have in depth training in crisis management from an approved source such as Safe Management or Non Violent Crisis Intervention.

However, there are some general strategies that can be recommended:

- **Try to intervene when the behaviour starts to escalate:** Know the person well enough to recognize the signs that s/he is starting to lose control. Distract the person, help him to leave the situation, support him to use relaxation techniques, etc. It is always preferable to prevent the crisis.
- **If the crisis occurs, try to keep everyone safe:** Remove other people from the room, take objects out of the person’s way, and try to minimize the self harm.
Don’t touch the person unless you have been trained in how to do this safely: Touching a person in crisis can lead to injury to both the individual and the staff. Don’t do this unless you have been trained to do so. Avoid restraining, trying to move, or giving emergency medication (a PRN) to the individual.

Crisis intervention strategies are designed to be used in the short run. In the long run, comprehensive behavioural strategies should be developed that prevent the behaviour.

**PWS Specific Techniques**

When working with people who have PWS, there are several specific techniques which staff have used successfully to decrease challenging behaviours.

*Technique One: Use the “Preferred and non Preferred Activities” System (Linder, 2005)*

Preferred activities are ones that are enjoyed and looked forward to by the individual such as eating and watching a TV show. Non preferred activities must be done but aren’t particularly enjoyed. All regular activities should be categorized as either preferred or non preferred. Then, the following strategies can be used.

*Technique Two: Schedule non Preferred Activities before Preferred Activities*

A successful technique is to schedule activities that someone likes after the non preferred activities. This will motivate the person to complete the non preferred activity and act as a reward for having completed the non preferred activity.

*Technique Three: Make non Preferred Activities as Pleasant as Possible*

Even though someone may not like an activity, staff can try to make it as positive as possible. For example, if someone needs to do a chore, play music that the person likes during the chore or provide some company while the chore is being done. We all have activities that we must do that we don’t like! Often, we use this technique ourselves.

*Technique Four: Wait it out/Broken Record*

This approach can be used for non preferred activities. Quietly wait for the person to complete the activity. Occasionally prompt the person, but don’t nag. When the person starts the activity or completes a part of the activity, give immediate praise.
Another aspect of this procedure is called “Broken Record”. Politely repeat the prompt at specific time intervals. Don’t engage in other conversation or negotiations.

Be prepared for a stand off when you first use this technique. The person may test you to see how long you will wait before you give up on making him/her complete the activity. You should decide beforehand your time limit.

**Technique Five: Time Sensitive**

This is used with preferred activities to help people to stick with their schedules. Quietly remind the person that the preferred activity is going to start in a few minutes. The start time is non negotiable. If the person is not ready at the start time, s/he does not get the opportunity to take part in that activity.

**Please note**: if the missed activity is a meal or snack, this should be recorded. Ensure that all staff and supervisors are made aware when food is not eaten since the daily calorie intake is so limited already.

**Technique Six: The 3 Question Rule**

Sometimes, a person with PWS will be anxious about something and will repeatedly ask the same question. While this is the person’s way of expressing his/her anxiety, it can be very annoying for others and is not a socially acceptable behaviour.

The following method can be used:

- Answer the question the first time it is asked.
- The second time the question is asked in a short period of time, politely reply but indicate that this is the second time you have answered it.
- Refer the person back to the information that has the answer to their question such as the posted menu or schedule.
- The third time the question is asked, answer it but state that this is the third time you have answered that question and that you will not answer it again.
- If the question is asked again, ignore the question. Turn your head and change the topic. Do not ignore the person altogether. This will increase his/her anxiety and lead to other behaviours.

In another scenario, the person is very anxious and asks a great number of different questions about the same topic. In this situation, politely inform the person that you will answer only 3 questions on that issue and ask the person to choose which 3 s/he wants answered. This latter technique only works with someone who has the cognitive skills to understand the process.
Technique Seven: Manage Transitions

Changes and transitions are difficult for people with PWS. Give multiple warnings ahead of time when transitions are going to occur. Warn at 15 minutes before changes, 5 minutes before, and 2 minutes before. Warnings are more effective when they are provided through multiple means such as auditory (a timer) or visual (a flashing light, cue cards, or a hand signal) that can bolster the verbal warnings. It is also helpful to have specific tasks to engage in while moving from one activity to another. Once an activity is over, it is over. Staff should no longer participate in the activity once it is supposed to be finished.

Technique Eight: Group Programs

On occasion, a group of individuals may share a challenging behaviour and a group program makes sense. In this situation, ensure that all group members understand what the expectations are. Reinforcement is given when all group members behave as required. Consistency is absolutely necessary in this type of program – all members must meet requirements.

An example of this is a group program for doing chores in the house.

Technique Nine: Using Contracts

One technique which increases the individual’s control over his/her behaviour program is the use of contracts. As the name implies, the person and the staff develop a contract regarding the behaviours that are required. The consequences of performing that behaviour and the consequences of NOT performing that behaviour are clearly indicated. Staff and the individual sign the contract.

Some points about contracts:

- The person must be able to understand the content and process of the contract. Not all clients have the cognitive abilities to use a contract system.
- Pictures can be added to the written content to increase comprehension
- Contracts must be scrupulously adhered to by all participants
- Contracts can be a successful way to avoid nagging and complaining.
**Strategies for Specific PWS Related Behaviours**

The following suggestions are adapted from Greenswag & Alexander, 1995.

<table>
<thead>
<tr>
<th>Rigid thought process and inflexibility</th>
<th>Provide weekly and daily schedules</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perseverative or obsessive thinking</td>
<td>Restate request or what you said, use visuals, less is best, i.e., one direction at a time. Think of perseveration as an “on/off” button for people with PWS; it sometimes gets stuck in the “on” position.</td>
</tr>
<tr>
<td>Tenuous emotional control, shows frustration or intolerance</td>
<td>Avoid over stimulation, stick to schedule, set daily (hourly) behavioural expectations</td>
</tr>
<tr>
<td>Poor stamina (falling asleep during day)</td>
<td>Get person up and walking, send on errand, diversion, schedule high energy activities after lunch, schedule rest periods if required</td>
</tr>
<tr>
<td>Scratching or skin picking</td>
<td>Diversion, short nails, use lotions, supervision and reward for not picking or alternative behaviour, cover area with bandage, consult medical advice for serious cases</td>
</tr>
<tr>
<td>Rectal picking</td>
<td>Check for constipation or hemorrhoids, use a body suit</td>
</tr>
<tr>
<td>Difficulty with peer and social interaction</td>
<td>Keep social groups small, many do better alone; pre-plan outings, limit or avoid surprises or schedule. Involve person in planning. Provide social skills program training that emphasizes sharing and taking turns</td>
</tr>
</tbody>
</table>

**In Conclusion**

This Chapter has a great deal of information about behavioural approaches. These ideas have been used with a multitude of people with developmental disabilities in general and PWS in particular.

Although the content may appear to be overwhelming, you will find that the strategies are helpful and well worth using.
CHAPTER SIX: EARLY INTERVENTION

With this Chapter, we will start to investigate the provision of specific types of services.

Considering the life span of the individual, it is logical to begin by looking at Infant and Child Development services. (ICDS)

**What is Early Intervention?**

Infant and Child Development Services provide a range of prevention and early intervention services and supports for families whose infants are “at risk.” This includes infants with genetic anomalies, medical issues, traumas, or problems in their environments.

ICDS include:

- Assessment
- Consultation and support in creating strategies to help infants to develop skills
- Teaching parents and caregivers how to carry out strategies, including demonstrating and providing equipment
- Nourishing of the parent/infant relationship
- Education and information
- Referral
- Advocacy
- Parent support
- Support to other agencies working with the child

Infants with Prader-Willi Syndrome are definitely at risk and can benefit from ICDS.

**The 2 Stages of PWS in Preschoolers**

There are 2 distinct stages of PWS in preschoolers.

*Stage One: Failure to Thrive*

The infant with PWS up to 2 years of age presents as a child that has failure to thrive. Key traits are:

- Hypotonia (poor muscle tone)
- Poor suck
- Difficulty feeding
- Hypogonadism (incomplete sexual development)
- Obvious delay in motor milestones
At this point, it is very difficult to believe that this weak, slow to respond child is going to become a demanding child who is fixated on eating and is very emotional.

**Stage Two: PWS Behaviour Begins**

By the age of approximately 2 years of age, the young child displays a noticeable change in behaviour. The child becomes more active and the “typical” PWS characteristics start to emerge. These include:

- A decrease in hypotonia as muscle development occurs
- Hyperphagia (lack of feeling full)
- Increasing obesity if hyperphagia is not controlled
- Cognitive limitations
- Emotional instability
- Temper tantrums
- Stubbornness or difficulty switching attention

This shift to this more “typical” PWS characteristics makes extremely different demands on both the family and the ICDS staff.

**Infant and Child Development Services**

*Adapting the ICDS Philosophy of Working with Families*

In Chapter Three there was a discussion about the necessity of modifying our typical philosophy of service delivery because of PWS. The rights approach is changed to one of more control by staff for the health of the individual.

A similar modification of the philosophy of service delivery occurs for ICDS staff when they work with families whose infants have PWS.

ICDS has a strong commitment to the family-centred approach. (Nugent, 2006b) This approach states that the family is the primary decision maker in the child’s life and the ICDS service providers should support the family’s ability to provide the best quality of life for the young child. Families choose the priorities for their children and thus the goals that they want to work on.

A key element of the family-centred approach is that the staff take their cues from the family with respect to next steps. Each family moves at its own pace. Some families are ready to accept the child’s diagnosis and work on strategies much sooner than others. Some families are operating on a day-to-day basis and can’t consider the longer term future.

However, with PWS, the ICDS staff must modify this approach.
It is critical to educate families regarding the change that will occur when the child is approximately 2 years old and to prepare them to deal with the strikingly different strategies that they will be using as the changes occur. In this way, the family will be able to more effectively help the child to avoid the serious implications of PWS. It is also important for the family to be involved with a paediatrician and dietician who are both knowledgeable in PWS.

The practical implications of this philosophical shift for ICDS staff are significant:

- ICDS staff need to be more assertive with the family than they ordinarily would with respect to the goals that are chosen for service plans.
- Education about PWS and its implications for the future become a powerful focus, along with discussion about preventative measures the family might strongly consider.
- They emphasize a focus on preparing for the future of the child, even if the family would not normally be ready for this type of information.

Such a shift in philosophy can be difficult for ICDS staff. They try to maximize the family-centred approach while ensuring that the family provides the best possible supports for their child. This includes taking a longer range view with these families and addressing this longer range view more often than ICDS staff typically would. Being more directive goes against all their training, but ICDS staff have learned to come to terms with this approach given the needs of the child with PWS.

**ICDS Service Focus for the Young Infant**

With the first stage of PWS, the ICDS staff are helping the family with feeding and muscle strengthening strategies. Understandably, the family is very concerned about the fact that the child is not eating and there are very realistic failure to thrive issues.

However, at this stage, ICDS staff also introduce information regarding the future of the PWS. Understandably, the family may find this information almost unbelievable. After all, how could this child who has such great difficulty eating become a child with food obsessions? ICDS staff may be required to do a lot of supportive education before families can accept the reality of their child’s future.

ICDS staff highlight the fact that families should start early to become educated about the future strategies that they will be using with their child.

ICDS staff use all available opportunities to assist families to become educated about their child’s future. One excellent opportunity, for example, is when meeting with families to develop service plans – this opens the door to introducing these future oriented issues. Within the structure of discussing goals, the ICDS staff can talk about what will happen when their child is older.
ICDS Service Focus for the Child Whose Behaviour is Changing

When the typical PWS behaviours emerge, the focus of ICDS strategies will alter. ICDS staff help the families to deal with the food obsessions, the problems with transitions, and the emotional volatility of their child.

The major focus will be on teaching the family to develop new ways of dealing with food in the family and the other PWS behaviours. This includes:

- Establishing a limited caloric intake per day for the child under the supervision of the paediatrician and nutritionist (this is particularly difficult given that the child wasn’t eating previously)
- Developing a strict routine for eating which dictates the only times when food/snacks are eaten
- Encouraging exercise and activity
- Developing rules and realistic expectations for other behaviours
- Helping the child to cope with transitions
- Learning how to respond when the child is obsessional or temperamental
- Carrying out consequences if the child engages in particular behaviours
- Minimizing their child’s anxieties

The family’s culture must be taken into account. In some ethnic cultures, food play a crucial role in socializing, celebrations, and major milestones. Food can also be the primary way that love is demonstrated.

Families also have their own individual styles. In some families, meal times are regular, food is only eaten at the table, and snacks aren’t allowed. In other families, meals are not organized and people eat at different times. Snacks are often eaten and meals are eaten in different rooms of the house. For the first type of family, it is relatively easy to introduce the controls that the PWS child will need with respect to food. For the second type of family, a complete rethinking of their family patterns is necessary.

Extended family is also involved. Grandparents and other relatives may have a difficult time understanding why they can’t just feed the child treats all the time.

Some ideas which have been successful are:

- Making mealtimes very practical and focussed on eating
- Having family discussions, meetings, and fun times away from the dinner table
- Re orienting family activities and celebrations away from food
- Increasing the level of exercise that the family as a whole and the child participate in
As well, it is critical to ensure that families understand that PWS is a **lifelong condition**. Rules and restrictions will be required forever with their child. Even when the child’s weight is stabilized and behaviours are minimized, the structure cannot be relaxed. This is a problematic message to deliver to families. It should be accompanied by information about how successful and enjoyable the lives of people with PWS can be.

Infant Child and Development staff use a somewhat modified family centred approach as they support families to adapt in the best interests of their child. They can also consult with other agencies such as day care centres that work with the child.

**Early Intervention: The Earlier the Better**

The younger the child is when the PWS diagnosis is made, the more effective the ICDS can be.

In particular, families who learn about their child’s diagnosis at a very young age are better able to begin understanding the diagnosis and building a network of support. They can introduce the structure into their child’s everyday life that is the best for the child. They have the opportunity to be more effective at establishing the routines around food and using behaviour management strategies that will stand the child in good stead for his/her entire life.

Therefore, early identification helps the family, the child, and the ICDS staff to work together more successfully.
CHAPTER SEVEN: 
THE SCHOOL YEARS

After participating in Infant and Child Development Services and preschool programs, children with PWS will move on to the school system.

School may be the first setting where the child and family are forced to deal with the impact of PWS in the more demanding expectations of the academic environment. The family has considerable control over its own environment and the strategies that are used to help their child. Pre school settings such as child care centres or nursery schools are relatively small and very structured with respect to routines and food availability.

Schools are larger and include many different environments such as classrooms, bathrooms, gyms, lunchrooms, school yards, music rooms, libraries, and hallways. The child with PWS will be dealing with many different staff plus students of all ages. Control is not as tightly exercised. For example, older children go to the washroom on their own, children eat snacks outside at recess, and schedules may be modified during holidays or changes in the weather.

All this adds up to a new set of challenges for parents and the child with PWS.

This Chapter will discuss the school years from two perspectives: the school staff and the parents.

**Educating the Child with PWS: Strategies for School Staff**

All school staff must be aware of the needs of the child with PWS and support the strategies that are required. The teacher will be responsible for the majority of the supports provided to the student. However, educational assistants, administrators, bus drivers, lunch room supervisors, office staff, recess supervisors, and special education consultants will all be involved in ensuring that the child with PWS gets the structure that is needed. The principal has the ultimate responsibility to guarantee the health of the student with PWS is maintained.

**Education about PWS**

When the child is enrolled, the school must be made aware that the child has PWS. The school usually arranges a meeting, or case conference, which may include the school administrator, resource staff, the parents, and, ideally, the teacher. In addition to talking about the child’s educational plan, consideration should be given to developing a plan to educate all staff, children, and parents about the new routines that are required in the school.
A brief information sheet can be prepared which informs all school staff about the specific characteristics of PWS, the importance of food control, and the necessity of maintaining as much consistency as possible in routines.

The teachers can also develop a simplified way to explain PWS to the children in the school. A brief letter to parents of other children in the school, signed by the principal and the teacher, can be prepared as well. This letter can ask for the cooperation of the parents and list the specific actions that are requested of them.

It is helpful to present the food issue as being similar to a child who has a life threatening allergy to peanuts. This is an example that many people are familiar with and emphasizes that PWS is a non contagious medically related problem that the child is born with. This approach removes the sense of blame away from the child while stressing the importance of the food restrictions.

*Environmental Modifications: Food Stimuli*

The first priority is to modify the school and its grounds with respect to food stimuli:

- Be aware of the child’s menus and stick to them
- If the child says s/he has missed a meal or snack, check this out with the parent before giving the child food
- In the classroom, put all lunches and snacks in a locked cupboard
- Avoid keeping treats on the teacher’s desk
- Purse and other sources of money such as petty cash should be locked away
- Adults should not enter the classroom with beverages
- Monitor the child’s behaviour with non edibles such as glue, cleaning products, etc. If the child tries to eat these items, lock them up
- Ensure that other students and their parents understand that there are strict rules around food: don’t have food sitting in the classroom, don’t “share” food with the child who has PWS, don’t send unexpected treats in for the children in the class, inform the teacher if treats are being sent in for a special occasion so that plans can be made to accommodate the child with PWS.
- If snacks are eaten in the classroom, clear away all leftovers promptly
- Remove food garbage from the classroom
- Control visual stimuli related to food – don’t post pictures of food, use food to represent alphabet letters, have magazines/books with food, or use food in craft projects.
- Control smells related to food as well
- Avoid using food in lesson plans when teaching concepts such as measurements, colours, etc.
- If there are any pets in the classroom, put their food away
- During holidays, steer class conversations away from food and concentrate on activities
Plan how to deal with Hallowe’en, a particularly food oriented occasion – children might be given non edibles such as small toys or healthy foods such as sugarless candies for treats.

Monitor food garbage in the lunchroom and on the playground.

Before going on outings or field trips, plan how the food issue will be dealt with.

If the child rides the school bus, his/her lunch should be given to the driver for safekeeping and passed to a teacher upon arrival.

If a mistake occurs and the child does overeat, don’t blame him/her. Just return to the basic routines. Inform the parents if this has happened.

These types of environmental modifications will help reduce the behaviours of the child while reducing his/her anxiety levels. This will lead to a much calmer and more pleasant classroom.

**Routines**

Due to the rigid thinking processes and difficulties with change, it is highly recommended that strict routines be maintained in the classroom. This is critical to reduce the anxiety levels of the child.

- At the beginning of the day, review the day’s routines with the child.
- Post schedules, using visuals to help remind the students of the routines.
- Give the child warnings before transitions occur – use different modalities such as a verbal warning, flashing the lights in the classroom, ringing a bell, etc.
- Schedule preferred activities after non preferred activities.
- If a change in the routine must happen, inform the student ahead of time and explain why the change is occurring.
- Involve the child in planning how s/he will cope with changes, field trips, etc.
- Offer the child a limited number of appropriate choices – should we read the book about the puppy or the book about the airplane?

Consistency is **essential** for a child with PWS.

**Behavioural Strategies**

The key to encouraging appropriate behaviours is to develop and adhere to rules and expectations.

- Involve the children in developing classroom rules where appropriate.
- Make sure that the child with PWS understands the classroom rules and consequences – use visuals to help explain the rules.
● Post rules for all children to see
● Be consistent in enforcing the rules
● Avoid negotiations and power struggles – calmly remind the child of the rules and stick to them
● Use logical consequences and apply them as soon as possible after the behaviour is displayed
● If the child persists in asking the same question, use the 3 question rule as described in Chapter Five
● Use large amounts of reinforcement when the child behaves in a socially acceptable way
● Have a quiet area in the classroom where the child can go if s/he is stressed
● Avoid using food as a reinforcement
● If specific situations or locations are problematic, help the child to avoid them – hallways, pep rallies, etc.
● Include physical activities in the daily routines.
● Schedule activities in pairs or small groups to help the child improve his/her social skills

*Teens in High School*

The major issue that has to be dealt with as the child becomes a teenager is the decreased level of supervision that occurs in the school setting. At high school, students are out on the school grounds without supervision and are also allowed to leave the school grounds for lunch, etc.

At this point in time, the student must have developed some understanding of the need to stick to the rules or the new freedoms can be disastrous.

Realistically, the teen with PWS may have to move into a more segregated environment such as a special education class. This environment offers more supervision and controls. For the child who was able to be integrated into regular classes during elementary school, this can be a difficult change.

*In Conclusion*

For children with PWS in the school, the keys to success are controlling food related behaviours and having structured routines. At first, the food issues will seem overwhelming, but the classroom can settle into a “PWS-friendly” system fairly quickly. The focus on structure can be helpful for all the students in the classroom.

With early identification, most children with PWS are entering school with an understanding of rules and routines already firmly established. School staff can then build upon this foundation to continue the child’s success in coping with PWS.
Your Child and the School: Strategies for Parents

Every parent experiences feelings of anxiety and apprehension when his/her child starts school. There is a sense of losing control as your child comes under the supervision of other adults for a significant part of his/her day. For a child with special needs, the parents’ worries are quite understandably magnified.

When the child has PWS, the anxiety and apprehension are even greater because of the potentially life threatening outcomes if the controls around food aren’t maintained.

Advocacy

Parents will certainly be required to advocate on behalf of their child. This may sound somewhat frightening for parents, but that is because there is a misunderstanding about what advocacy is. Most people think of advocacy as extreme measures that involve fighting with others and being labelled as a trouble maker.

In fact, advocacy refers to providing support to or promoting the best interests of an individual. Advocacy does not need to be an unpleasant process. The most effective advocates try to maintain a positive relationship with others. There is truth in the old saying that “you can catch more flies with honey than vinegar”.

In the case of a child with PWS, parents’ advocacy will concentrate on educating the school. The best service for the child will occur when the parents and school work together cooperatively. The parents’ focus should be on helping the school to support their child.

Educating the School

Initially, the school is not likely to know anything about PWS since it is a rare condition. Therefore, parents will be required to basically start at the beginning with respect to educating the school:

- Provide basic information about PWS – what it is, why it happens, and the characteristics.
- Emphasize that PWS is a genetic problem – it is not the child’s fault nor is it contagious.
- Ensure that the school knows about the health issues such as atypical reactions to medications, atypical body temperature regulation, and high pain threshold.
- Explain the food obsessive behaviours and how the child is at serious risk of health problems if food intake is not controlled.
Discuss typical behaviours and the importance of rules and consequences. Emphasize that PWS is a life long condition and that the controls will be needed the entire time that the child is in the school.

This information may be quite overwhelming for the school, but schools are now used to working with many students with a broad range of special needs. The school will be more successful if the parent can make the situation as easy as possible for the school:

- Provide written copies of the menu plan.
- Show the school copies of routines or behavioural strategies that you have used with your child at home or in other settings such as day care.
- Offer to give the school copies of any plans or assessments that you have from other professionals such as nutritionists or physiotherapists.
- Let the teacher know what your child’s strengths are academically so that these can be incorporated into his/her individual educational plan.
- Get to know the system in your child’s class so you can offer suggestions that are compatible with that setting.
- Offer information about strategies that have worked with other children in other schools – written information is best. Books, pamphlets, and web site addresses are all useful.
- If you know of other schools that have supported children with PWS, give the school information about how they can contact these other schools.
- Offer to speak to school staff about working successfully with children who have PWS.
- Be prepared to repeat all your hard work every time your child changes teachers, educational assistants, or principals.

Relating Positively to the School

Schools are as different as the children who go to the school. We all know that some schools and teachers will be open to suggestions, committed to serving all children, and willing to “go the extra mile” for a student with special needs. Other schools may not.

However, the parent should be as positive as possible in relating to the school:

- Don’t expect the school to be perfect. At first, there will be mistakes made, even in a school that is trying very hard to support the child with PWS.
- When mistakes are made, remain calm and discuss how the situation could be better dealt with in the future.
- Appreciate and reinforce what the school has done well.
- Try to figure out why the school is not following the required structure. Once you understand the reasons, you will be in a better position to deal with the situation.
If Things Aren’t Going Well

In spite of all your positive efforts, you may encounter a school or teacher that just isn’t supporting your child safely and consistently. In such a situation, advocacy will indeed become more adversarial to obtain better service for your child:

- Find out what the process is in your school board for obtaining services for a child with special needs.
- Find out what your rights are as parents – the information you are entitled to, meetings that you should be invited to, whether you can bring support people with you to meetings, etc.
- Contact your local PWS family association and/or other parents whose children have PWS to get support.
- Keep copies of all reports, letters, communication books, etc.
- Keep a diary of all events that occur, conversations with school staff, observations you have made, etc.
- Be prepared to state specifically what you are asking for. It is not useful just to say that you aren’t happy with what is happening or that you want something done. You need to clearly state what you want for your child and by what date.

In Conclusion

The vast majority of children and teens with PWS are successfully attending school. The key is to communicate with the school and keep in mind that everyone shares the same goal – the student’s well being.
CHAPTER EIGHT:  
RESIDENTIAL SERVICES

Once adults with PWS leave their family’s home, they typically move into a residential program that is offered by an agency in the developmental services system. The residential program becomes the prime support for the individual.

Although a small number of individuals with PWS may live successfully in individual settings, most people with PWS will live in group homes. Therefore, the focus of this Chapter is on the group home model.

There are a great many aspects involved with establishing a successful residential program that will be reviewed in this Chapter. The ideas introduced in previous Chapters dealing with Food and Behaviour will be illustrated in this Chapter. Topics include:

- Starting a new residential program
- The physical set up of the home
- The individuals in the home
- Dealing with food related issues
- Health factors
- Behaviour programs
- Other activities outside the home
- Relationships with families
- Staffing

The information in this Chapter is based upon interviews with residential service providers in developmental services agencies.

Starting a New Residential Program

A considerable amount of planning is required before opening a residential setting.

This planning is to be done before the individuals move into the home. Staff should be hired ahead of time and have the opportunity to participate in the planning.

The first issue to be discussed is coming to terms with the level of control that staff will be exerting in this home. As was reviewed extensively in Chapter Three, this is a change in the usual service philosophy and staff need to be given the opportunity to understand and accept this change.

Training in PWS is necessary. Staff recommend that, if at all possible, training include a trip to the Children’s Institute in Pittsburgh which has been acknowledged as the service that currently has the most expertise.
The structure for the home, including physical plant, rules, and routines, is then developed.

When individuals move into the home, everything is already established and there is a comforting consistency that welcomes them to their new home.

**The People Living in the Home**

Most homes vary from 3 to 5 people. This appears to the optimal size range.

*Should it be an All PWS Home?*

One issue which is often discussed is whether the home should only have people with PWS or a mix of people with different diagnoses living in it.

Both arrangements can work if the mix of people is carefully screened.

If only people with PWS live in the home, it can be easier to maintain the rules. There is no jealousy about the food that other people are eating nor is there resentment of the rigid structure by the people who don’t have PWS.

On the other hand, having a mix of people can lessen the stress in the home because the other clients may not have the same levels of anxiety and need for constant control. This can be a benefit for both the clients and the staff.

Either option is fine.

The one caution is to ensure that the people without PWS can behave appropriately with the people who have PWS. For example, in one home a man without PWS teased the individuals with PWS about the fact that he could eat what he wanted and he made a point of telling them what he had eaten that day at work. This man was moved to another home. The woman who replaced him did not have PWS but she was very cooperative and understanding of the needs of the people with PWS. She has successfully stayed in the home.

**The Physical Set Up of the Home**

Homes are located in regular neighbourhoods and from the outside they look like any other home in the community.

Inside the home, the impact of PWS on the set up of the home are evident.

*Security*

All group homes for people with PWS need excellent security to maintain control over their potentially dangerous behaviours.
The home should have alarms on all the doors to the outside – only one home did not have this feature, but that home is very small and it is easy for staff to hear when people leave.

Alarms can be equipped to play chimes during the day and give off a more piercing sound at night. This makes the alarm less annoying to people living in the home, staff, and neighbours during the daytime when the alarms tend to go off more often because people are coming and going.

Individuals’ bedrooms are locked. In some homes, the individuals have keys to their rooms, while in other homes, only staff have keys to bedrooms. This decision is based upon the needs and behaviours of the individuals in that home.

Other rooms that are related to food or consumable items are locked such as kitchens, storage rooms, pantries, and offices.

In one home, strict rules are enforced regarding individuals’ entry into the staff offices. Individuals are only allowed to enter the office to a certain point. All other areas of the office are “off limits” and are where staff keep medications, personal records, petty cash, purses/wallets, cleaning products, etc.

Security checks are carried out at regularly scheduled intervals and checked off on a recording form. Security checks include testing alarms and all locks.

**Bedrooms**

Individuals have their own bedrooms in which they have décor according to their personal taste and all their personal belongings. Having room mates is not recommended. It is very important for people to have their own space for privacy and calming down.

**Bathrooms**

Since some people with PWS will attempt to eat “non edible” items like soap and cleaning products, most homes keep the bare minimum of products in bathrooms. Usually there is only soap and toilet paper in the bathrooms. All other supplies are locked up by staff or in bedrooms, if this is safe for the individual.

**Kitchens**

As might be expected, very specific controls are put into place for the kitchens:

- Kitchens are locked
- Cupboards, pantries, and refrigerators are locked
In one home, a one-way window has been installed which allows staff in the kitchen to watch the dining room area but does not allow clients to see into the kitchen.

Several homes have installed equipment to vent smells out of the kitchen.

**Other Rooms**

Homes should have an exercise room with appropriate exercise equipment.

All public rooms should be monitored so that pictures, magazines, books etc. do not refer to food.

**Food**

The majority of the rules and procedures in the home apply to food.

The residential programs visited all used the Red Yellow Green diet. Staff are very enthusiastic about RYG. Clients find the RYG easy to understand and adhere to.

Menus are set up 3 weeks at a time. The menu plans include 3 meals and 1 or 2 snacks. Individuals participate in developing their own menu plans and there can be some individual differences in what is eaten. This is the time when negotiations can be made for special events, outings, etc. A menu plan for a home with several people is at the end of Chapter Four. It demonstrates how the general group menu can be individualized.

If a mistake is made and someone gets more food or different food, no comment is made. A note is made for future reference to determine how to fix the problem but no one is blamed for what happened. The client does not lose food from other meals as a punishment. In one agency, clients are required to check their food before starting to eat to ensure that everything is there. They must tell staff that something is missing **before** they start eating. The rule is: once you start eating, you have indicated that you have everything you need and no changes or additions to the food/beverages will be made.

Changes are not allowed in the menu once they have been printed for the 3 week period. Menus are always posted in the kitchen. In some agencies, menus are usually posted in a public area so that everyone knows what is food is being served and when. However in one home, the menus were only posted in the staff office since the clients got anxious when they saw what the other residents were eating. In another home, menus are not posted in public areas. Instead, to maintain privacy, each client receives a copy of his/her menu.

Individuals do not participate in food preparation. Food is brought to the table already served and dishes are cleared when the meal time is over. No serving
dishes or containers are ever brought to the table. Everyone eats at the table. In some homes, staff eat with the clients but this doesn’t happen in other homes.

Schedules are established for all meals and snacks. At the end of the time period, the meal is over and food is removed. In some homes, people who are late for a meal receive a less appetizing meal. In other homes, people who are late miss the meal or snack. Typically, there is a time period within which people can come for a meal and be considered “on time”. For example, lunch may be from 12:00 noon to 1:30pm and people can come to eat during that time period. However, they must finish by 1:30.

Food garbage is removed promptly from garbage cans.

There are some differences in how meals are prepared. In one home, the meals/snacks are made in another location and brought in daily for the client with PWS. In other homes, staff prepare the meals in the kitchen at the home.

Grocery shopping and cooking are strictly controlled, with clear rules regarding both processes. Grocery shopping is not discussed with clients present. Two homes had proscribed methods for bringing groceries into the home so that clients could not see the food being brought in. The van was locked if it was left with food or groceries in it.

Staff recommend that a system be established so that all food related issues are handled through one person, typically the home’s supervisor or manager. As well, that person will schedule the timing for discussions about menus. Food negotiations are not allowed with anyone else or at any other time. In this way, ongoing arguments are eliminated and all the staff do not have to deal with food issues all the time.

Finally, it is necessary to ensure that all diets are approved and monitored by doctors and nutritionists.

**Health Factors**

There is no doubt that group homes have had tremendous success in helping people with their health issues:

- All individuals have lost impressive amounts of weight
- One individual had surgery to remove the flaps of skin left when he lost a huge number of pounds
- All individuals participate in regular exercise
- General activity levels are increased
- Individuals are sleeping less and have more energy
In spite of all these improvements, the individuals still have a large number of medical appointments. Staff appear to be constantly scheduling appointments, taking people to appointments, recording data for appointments, etc.

There is a lack of medical and other professionals with expertise in PWS. Staff report that they find they are often educating professionals about the implications of PWS.

**Behaviour Programs**

Behavioural approaches were discussed in considerable detail in Chapter Five. This Chapter will expand on this by offering ideas that have been suggested by residential staff.

**Using a Behavioural Consultant**

All residential programs recommend having a behaviour consultant assigned to the group home. The consultant will help the home to develop the rules and structures that were discussed in detail in Chapter Five. As well, after a while, staff may lose their consistency or reinforcers might be less effective. A consultant can act as a neutral, objective observer who will notice these changes and provide ideas to get everyone back on track.

**The Structured Approach**

All the activities and routines in the home were strictly scheduled and monitored. Rules and data collection were in place for all behaviours. The group home staff had several comments to make about the use of strict rules and keeping records:

- Each person’s day is scheduled from waking up to going to bed at night
- Anxiety is decreased for both clients and staff
- A “no discussion” approach is used – staff state the expectations in a matter of fact manner, no processing is allowed, no arguments ensue, power struggles are avoided
- Consistency is mandatory
- Choices are limited
- Writing everything down allows staff to evaluate how things are going and helps communication
- Changes are kept to a minimum
- Clients should be prepared for changes or transitions
- If an issue arises or a mistake occurs, it is dealt with in a straightforward manner and then everyone moves on. There is no nagging.
- Clients are encouraged to write down their concerns for discussion later
- Regular meetings are held to discuss concerns – a weekly schedule for meetings is recommended
Teaching Skills

The behavioural approach also includes a component of teaching skills. Staff worked diligently to include teaching in the clients’ lives. Some of the key skills included relaxation, problem solving, and social skills.

Rectal Picking

One home had an ingenious method of helping a woman who engaged in harmful rectal picking. They purchased a dance leotard and put a zipper in the back and she wore that during the daytime under her regular clothes. At night, she slept in a one piece pyjama. She was not able to get her hands into these types of clothing to engage in the rectal picking. However, this clothing can become loose over time so staff must monitor to ensure that the fit is still too tight to allow the client to pick.

In Conclusion

Overall, staff were insistent that a rigorous behavioural approach is an absolute necessity.

Activities Outside the Home

All of the group homes ensured that individuals had opportunities to engage in a variety of activities outside the home.

The main focus was ensuring that the controls that the clients needed could be maintained during the activities. In particular, environments were evaluated for the presence of food or food related stimuli and access to money. Depending on the nature of the activity, staff tried to talk to the people supervising it ahead of time to modify food. For example, when clients were going to a dance at the day program, the usual refreshments were changed to include diet pop, sugar free candies, pop corn, and fruit.

All activities were part of the written schedules that were posted on a regular basis. Most schedules were developed on a weekly basis. Changes in schedules very rarely occurred since they caused so many anxieties for the individuals.

Activities were individualized if staffing could be arranged so that clients could follow their own interests.

Relationships with Families

The variation in families is as broad as the variation in the individuals.
Some families were very cooperative with the requirements of the group home system. They followed all the rules when visiting their children or having them home for visits.

Other families did not truly understand how important the structure was and did not follow the rules about food and other behaviours. In these cases, families were encouraged to visit their family members in the group home or take them on outings rather than having the client visit the family’s home for days at a time. This minimized the negative impact of the family’s behaviours.

Some residential services have used a contract system with families. For example, the contract will state the conditions under which the individual will go for a visit to the family home. Such conditions might include the family maintaining the diet and exercise routines.

In one agency, the frequency and length of visits with families are decided upon by the manager or director. Clients know that the staff are not making these decisions. This prevents the situation in which clients challenge staff to change how often or how long their family visits are.

Also, some clients would feel jealous if others had more positive relationships with their families. This could lead to challenging behaviours. Staff tried to downplay any comparisons of families among clients.

In general, staff tried to assume the major responsibilities for the individuals’ programming and let families have more relaxed relationships with their children.

**Staffing**

The staff are the key to the smooth functioning of the home. Recruiting, training, and scheduling staff are important elements of the supervisor’s job.

**Staff Schedules**

People with PWS need 24 hour staff support. Depending on the number of people living in the home, 2 or 3 staff will work the daytime and evening shifts. All group homes have overnight staff. In most homes, the overnight staff are awake. However, in one small setting with a very capable group of residents, the overnight staff slept in the living room in the way of access to the front door.

It is best to avoid rotating staff across shifts. This lack of consistency will cause anxiety and confusion for the individuals.
Training

All staff who work in PWS residential services should have a comprehensive training program. This should include information about PWS, visiting the setting, and performing a trial shift. The final Chapter will provide a detailed overview of a PWS training curriculum.

Communication

Ongoing communication is a necessity. Weekly staff meetings for all staff are very important. As well, when staff conscientiously keep all records, these are an excellent source of communication as well.

As well, all staff should meet regularly with the behaviour consultant to work on issues.

Choosing the “Right” Staff

Working with people who have PWS is not for everyone!

The most successful staff are people who like structure and can be consistent. They must be comfortable with having more control than usual over the clients’ lives but they can’t abuse their power.

As well, as one staff person says, “You can’t take it personally”. In other words, staff have to be able to understand that the client’s behaviour is not aimed at any individual, but is the result of their PWS and related anxieties.

It also helps to have staff who are easy going and not easily upset. Working with people who have perseverative or dangerous behaviours can be very annoying. A volatile staff member will not stay long in a setting with PWS.

Interestingly enough, supervisors have said that the clients will let them know which staff “follow the program” and which staff are inconsistent with the rules and diets! This illustrates that the clients themselves feel more anxious with staff who don’t provide them with reliable structure.

Avoiding Staff Burnout

Since consistency is so important for people with PWS, it is ideal if staff turn over can be limited.

Staff usually leave for one of two reasons: burn out or moving on to other opportunities. The latter is to be expected and can’t really be stopped. However, burn out can be decreased if:
Staff have proper ongoing training
- Staff are placed in jobs that are compatible with their personalities
- Staff are empowered – supervisors refer issues back to staff to solve rather than making decisions with clients without staff input
- Appropriate professionals such as behaviour consultants, nutritionists, physical therapists, and doctors are available to help staff
- Staff have the opportunity to vent – talking about the stress of working with such a demanding group of individuals.

In Conclusion

The right staff in the right setting with the right structure will result in a group home that provides excellent service for people with PWS.

According to one staff: “Working here sounds like a nightmare if you don’t understand Prader-Willi Syndrome. Once you understand it and get to know the people, it’s a great job!”

View from Dining Room of One Way Mirror Window into the Kitchen:
Clients in the Dining Room Can Not See into the Kitchen
View of Dining Room from the Kitchen through the One Way Mirror Window. Staff working in the kitchen can see clients in the dining room/living room area.
CHAPTER NINE:
VOCATIONAL PROGRAMS AND DAY ACTIVITIES

We now move on to the last service related Chapter in the Handbook, looking at the topic of developing appropriate day time activities for adults after the school years are completed.

Most individuals with PWS will finish high school, or its equivalent, at approximately the age of 21 years, depending on geographical locale. Realistically, most of these individuals will not attend a college or university.

Therefore, the next step will be to pursue day activities that are enjoyable, meaningful, and can support the structure that these individuals require:

- Supervised access to food according to detailed menus
- Controlled access to other substances that could be eaten such as garbage, cleaning products, or craft products
- Strict routines for daily schedules
- No access to money
- Clear rules that have logical consequences that are consistently enforced

When we consider this list, we can understand that the day activities could be limited in choice. However, it is important to pursue the most fulfilling activities as possible.

Many individuals will have a diverse schedule which combines many options. The key is to ensure that the week is organized and consistent so that the individual knows what s/he is doing each day, thus decreasing anxieties.

Day Program Options

Day programs are developed on an individual basis, keeping in mind the person’s developmental disability and behavioural needs.

Work or Volunteering

Some individuals with PWS are able to work in a competitive environment or volunteer in situations in which they are fulfilling legitimate functions.

Some strategies that have proved to be successful in the work and volunteer arenas are (Saporito, 1995):

- The employer and other staff are educated about PWS and its implications for the work site
- Access to food is restricted at all times in lunch rooms, break rooms, washrooms, kitchens, outside eating areas, etc.
- The individual is supervised whenever s/he is eating
- Off limits areas are clearly identified and marked with signs
- One supervisor is identified to whom the individual can take all work-related concerns, questions, and problems
- A place is designated outside the main area where the individual can vent or calm down
- Accommodations are made for the developmental disability since the individual might take longer to learn and understand
- Behavioural strategies that are used in other environments such as “broken record” and “the 3 question rule” can be used at work
- Use visual models, demonstrations, schedules, check lists, and signs to assist with job performance

It is helpful to seek out work places that might have some sensitivity to special needs already. For example, one woman is working part time as a secretary at the head office of the agency which supports her residentially. She does legitimate work, but in an environment where people understand her needs.

Other choices are jobs which don’t offer many potential problems. This same lady also does part time house cleaning in the company of a support staff person. At another group home, some individuals share a paper route which they complete under the supervision of a residential staff person. Both of these options are controlled, with minimum exposure to food or anxiety producing stimuli.

Some individuals might not be able to work or volunteer for a full five day week, but could cope with a limited number of hours of these activities each week.

**Sheltered Work Environments**

Sheltered work options can be somewhat diverse:

- A workshop in which the individual is performing work for pay
- A workshop in which the individual is engaged in life skills and recreational activities – this is typically appropriate for individuals with more significant developmental disabilities and/or behaviours
- A work crew from the workshop which goes out of the workshop setting to perform work in the community such as yard work, bus shelter maintenance, or office cleaning
- A work enclave in which a work crew from the workshop goes to a community work site to perform specific jobs while under the supervision of workshop staff

An individual might have a schedule with a combination of these options.
As with employment settings, sheltered workshops also need consistent, structured environments. Once again, it is necessary to ensure that staff at the workshop are well aware of the rules that must be enforced for the individuals with PWS.

Clients must be informed too, but some judgement must be used with respect to how this is done. Given that the clients will have some intellectual delays, a complicated explanation of PWS might not make sense. It may be easier just to say that the individual has a health problem and no food sharing is allowed.

Some strategies which have been helpful for individuals who work in sheltered work environments:

- The individual gets escorted into the workshop through a separate entrance to avoid potentially upsetting stimuli such as the kitchen.
- If there is a contract involving food that is being completed at the workshop, this is done in the main work area while the individual moves to a small room to work on a different contract with a small number of people.
- Before a new job begins, the individual is prepared for it and a contract is established so the individual knows what is expected of him/her in this new situation.

Sheltered workshops offer individuals the opportunity to learn work skills, work for pay, and socialize with others while in more controlled settings.

*Non Vocational Day Activities*

Many adults with PWS who live in residential services are engaged in a structured weekly schedule of activities.

The goal is to develop a schedule which offers variety, activity, community participation, and fun. Examples of activities are:

- Sports
- Fitness clubs
- Mall walks
- Academics and literacy
- Therapies such as music therapy, art therapy, and communication therapy
- Attending activities at another group home
- Visiting friends

As with all plans for people with PWS, the day activities schedule should be clear and consistent. An example of such a schedule is located at the end of this Chapter.
Back Up Activities

In keeping with the required strict structure of daytime activities, agencies have found it helpful to have organized “back up” activities. If the person misses out on a regularly scheduled activity, then a back up activity is completed. This back up activity is included in the pre-established routines and is clearly communicated to the client. For example, if a woman is late in the morning and misses her ride to a volunteer at the library, she does chores at her home.

Having a roster of back up activities maintains the structure which is so helpful to people with PWS.

In Conclusion

There are no magical solutions to the issue of employment for adults with PWS.

We must acknowledge that some people with PWS will find employment to be too anxiety provoking. Better options for them will be sheltered employment or a roster of well-chosen daily activities.

The priority is choosing day program options that maintain the physical and mental health of the individual.
### Weekly Activity Schedule Checklist

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<th>TIME</th>
<th>MONDAY</th>
<th>TUESDAY</th>
<th>WEDNESDAY</th>
<th>THURSDAY</th>
<th>FRIDAY</th>
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<td>9:00 AM</td>
<td>Activity Picked: __________________________</td>
<td>BOWLING @ BURLINGTON BOWL (MEET UP WITH ROTARY)</td>
<td>Activity Picked: __________________________</td>
<td>MUSIC THERAPY @ HEADON HOUSE (ROTARY HOUSE TO JOIN)</td>
<td>SWIMMING @ TANSLEY WOODS (MEET UP WITH ROTARY)</td>
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<tr>
<td>10:00 AM</td>
<td>Completed: _____________________________</td>
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<td>DH: LITERACY CLASS @ MAINWAY (LM @ BERKSHIRE)</td>
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<td>SWIMMING @ TANSLEY WOODS (MEET UP WITH ROTARY)</td>
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CHAPTER TEN: 
STAFF TRAINING

This Handbook has been devoted to helping staff understand how to support people with PWS in a variety of programs and services. Staff working with individuals with PWS must complete extensive training before they can do this job well.

This Chapter will recommend a general format for staff training, highlighting some key components.

Training Overview

A successful PWS training curriculum will incorporate specific elements:

- Teaching basic PWS information
- Observation of the potential work setting
- Completion of at least one shift to demonstrate skills and knowledge.

At each stage, accountability should be built in through a system of feedback either through discussions, completion of exercises, tests, and/or observation notes.

Teaching PWS Information

The curriculum for the first phase of PWS training for new staff should include basic information similar to the first five Chapters of this Handbook. Essential content would include:

- What is PWS and why does it occur
- Physical and Health Features
- Service Delivery: Philosophy and General Guidelines
- Weight Control
- Behavioural Strategies

Teaching is more effective when different modalities are used. Instructors can present information visually, orally, and with videos.

It is critical to build in opportunities for new staff to use the information during their training. Case scenarios, exercises, and problem solving activities are recommended.

The service philosophy issue is a particularly challenging one since it represents such a shift in perspective. Participants should have lots of time to deal with this topic:
Allow for discussion and self reflection.
Provide many examples that illustrate this philosophy in practice
Present scenarios and have staff answer how they would deal with these situations.
Provide specific feedback to staff regarding their answers
There should also be the opportunity for staff to develop behavioural plans based upon scenarios

It is important for trainers to “test” new staff to ensure that they have learned the fundamental information.

An individual who cannot demonstrate an understanding of this philosophy will have a very difficult time adhering to the rigid requirements of the lifestyle of a person with PWS.

Observation of the Potential Work Setting

After staff have knowledge regarding PWS, the next step is for them to observe current staff on the job.

Observations can include all the different elements of the job:

- Different times of the day
- Eating meals
- Preparing meals
- Exercising
- Day time activities

Typically, observations during new staff training are unstructured and there is no formal feedback mechanism. We are recommending the opposite.

Observation periods should be structured. Before they go to observe, new staff can be given recording sheets to complete in order to track what they have seen. Conduct formal debriefings after the observations during which the new staff have the responsibility to report what elements of the academic training has been demonstrated at the work site – for example, the use of rules and structure.

Accountability is essential during the observation activities.

Completing a Shift

After learning information and observing current staff on the job, new staff will complete at least one shift on the job.

Once again, accountability is critical. One current staff, preferably a supervisor, should be assigned to monitor the performance of the trainee during the shift.
This supervisor should have a report form that s/he uses to evaluate the performance of the new staff during the shift. This report can be reviewed with the new staff at the end of his/her shift by the supervisor at the work site or it can be done by the trainer.

The purpose of this process is to provide the new staff with concrete data about his/her performance, as well as helping the agency to decide if that staff is appropriate for working with PWS.

**In Conclusion**

Many agencies have excellent PWS staff training packages. The purpose of this Chapter is to remind agencies of the information and processes that have been demonstrated to have maximum training effectiveness.
CHAPTER ELEVEN:
WHAT DOES THE FUTURE HOLD?

This Handbook was written for staff who are working with people with PWS.

While there has been a review of the most current information on PWS, the emphasis has been on suggestions from agencies that are successfully supporting these individuals. Their ideas are practical and “road tested”.

Many staff are now at the point where they have helped people with PWS to accomplish the goals of weight loss, food control, regular exercise, and a structured lifestyle. This has improved both the physical and mental health of the clients.

These staff are now wondering what the future holds for these individuals.

After such significant accomplishments:

- Can we lessen the structure in their lives?
- Can we ease off the food restrictions?
- Can we allow these individuals to have more freedom?
- Are we failures as staff if the clients don’t become more independent?

The answer to these four questions is an emphatic “NO!”

**People with PWS must have lifelong controls placed on them.** They just don’t have the self control to live independently within the structure that they need to live a healthy life.

However, this doesn’t mean that there can’t be change. Just remember that any change must occur within structure.

As staff, it is important to keep an open mind and try to be creative. Rather than saying no to a new idea or request, we should problem solve. If someone wants a job, how can we make that happen in a healthy way? If another person would like to make friends, what strategies can we develop?

Services for people with PWS are becoming better all the time. Early identification means that systems can be established at a young age so that people with PWS will avoid the serious weight gain and health complications that seemed inevitable for people with PWS even 20 years ago.

The future does hold a great deal of promise for individuals with PWS.
BIBLIOGRAPHY


