

Health Highlights For Caregivers of Adults with Prader-Willi Syndrome

STUDY GUIDE Participant Copy

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A SPECIAL THANK YOU AND ACKNOWLEDGEMENT

INTRODUCTION:

There are many factors that influence our health – lifestyle, habits, and family history are just a few examples.

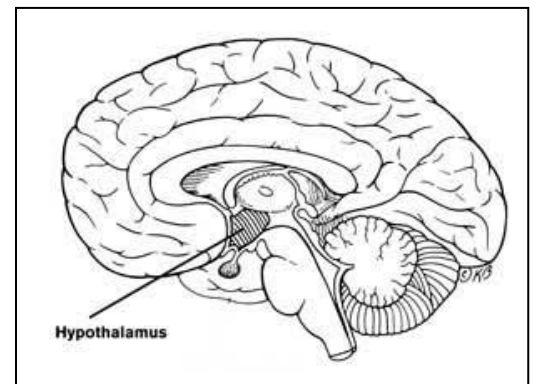
This presentation should not contradict recommendations of the health care professional. You are encouraged to consult and follow the advice of these professionals.

A DYSFUNCTION IN THE BRAIN

In many ways, the body of a person with PWS works _____ than yours and mine.

A problem in their brain affects their ability to:

- Control their appetite
- Regulate their body temperature
- Feel pain accurately
- Produce many hormones
- Have strong bones
- Swallow and digest their food properly
- Control their emotions
- Learn and understand things



CHALLENGES

Many of these special health issues often make it difficult to know when someone is sick or injured.

NOT FEELING FULL/LOW METABOLISM

Because of a dysfunction in an area of the brain (called the hypothalamus) – people with PWS always feel hungry. They will keep eating and eating.

When they eat – the message of _____ never reaches their brain.

In some people with PWS, this food drive is very intense. In others, it is not.

Not only do these individuals feel hunger all of the time, they also have a very _____ and lower calorie needs which makes gaining weight very easy.

Without your help using food security, they can overeat; become morbidly obese and even rupture their stomach.

People with PWS rarely vomit. If they do, it could be the sign of a _____.

Things You Can Do:

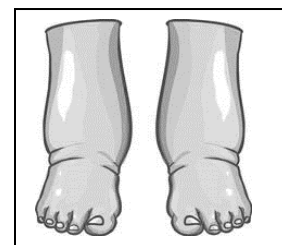
1. Monitor all sources of food – everywhere. Food security is _____.

- a. Locks and/or alarms
 - b. Precise planning for all activities where food could be present or impacted.
 - c. Plan, structure and prepare all meals. A low calorie diet is needed.
2. If anyone with PWS vomits, they should be evaluated by a health care professional.
 - a. Share the recommendation – a detailed plan for a workup is included in the PWSA (USA) Medical Alert booklet to help you advocate for what and why an evaluation is needed.

LIFE THREATENING OBESITY

Even though we have learned a lot about managing diet and food security, people with PWS may still gain a lot of weight.

Those who are very overweight may experience serious health issues including:



- Heart and circulation problems – high BP, heart failure, extreme swelling in their legs and feet.
- _____ that may require oxygen and other treatments.
- Diabetes which impacts the heart, eyes, kidneys, blood vessels and healing.

Things You Can Do:

1. Work with the health care professional to treat and follow the care recommendations for the health issues.
2. It is _____ to help him/her lose this weight.
 - a. Monitor weight daily.
 - b. Careful adherence to low calorie diet (or other prescribed diet).
 - c. Exercise as tolerated. Consult with Physical Therapist or Exercise Physiologist.
3. Avoid sitting for prolonged periods of time. Elevate feet and legs. Use circulation stockings as ordered.

GI ISSUES - THINGS YOU NEED TO KNOW

SWALLOWING PROBLEMS

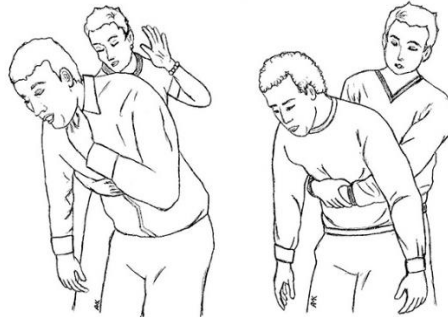
Recent research has shown that many people with PWS often have problems with swallowing.

- Food was caught in their throats and they were NOT able to tell.
- Most have a low production of saliva which contributes to swallowing problems.
- They often eat very fast which also puts them at high risk for choking.

Things You Can Do:

1. “Chase and Pace” – a strategy that _____ the rate that a person eats and helps to moisten, lubricate and push food down the esophagus.
 - a. Each bite of food is followed by a sip of water or other liquid.

2. *Try other measures to slow them down – putting eating utensil down between bites.*
3. *Serve smaller portions (if needed give seconds).*
4. *Provide food with moisture – soups, sauces.*
5. *Make sure you are trained in the Heimlich maneuver.*



SLOW STOMACH EMPTYING (GASTROPARESIS)

Many people with PWS experience slow stomach emptying.

Normally, when people eat, the stomach breaks down food and propels it into the small intestine.

When people have a condition called GASTROPARESIS, the stomach is unable to contract normally and food is not crushed nor pushed into the small intestine properly.

Stomach contents build up causing bloating and distention. Normal digestion stops. If not addressed immediately, the stomach can _____resulting in death.

SYMPTOMS YOU MAY SEE:

- Bloating or distention of the stomach (this can be very difficult to see)
- Vomiting
 - People with PWS rarely –if ever- vomit.
 - **THIS COULD BE A SIGN OF A SERIOUS PROBLEM!**
- Stomach “discomfort”
 - People with PWS do not experience or report pain accurately so they _____complain of stomach pain.

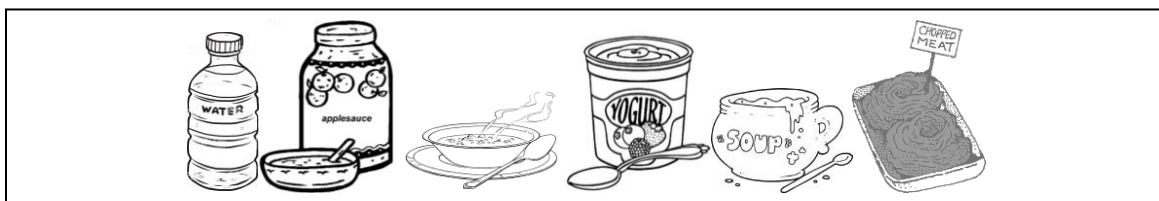
THINGS THAT COULD MAKE THIS PROBLEM WORSE:

- Overeating and/or Binge eating – ingesting too much food at one time which could result in a person _____.
- High Fiber foods and fatty foods – are harder for the stomach to digest and push these foods through the stomach.

Many people with PWS who have this problem require hospitalization until their stomach starts working properly again.

Things You Can Do:

1. If a person with PWS overeats, vomits and/or complains of abdominal pain or discomfort:
 - SEEK MEDICAL ATTENTION
 - BRING a COPY OF THE PWSA (USA) _____ which includes a chart that guides a health care professional on what is recommended in diagnosing and treating this health emergency. (It can also be found on the PWSA (USA) app or website – www.pwsausa.org)
2. Serve LOWER (not NO) fiber foods
 - _____ foods
 - Cooked vegetables
 - Fruits in natural juices or sauces
 - Ground meats
 - Increase _____ in cooking – liquids require less work to digest
 - Soups
3. **AVOID** high fiber foods – salads, raw vegetables and fruits
4. Smaller quantities of food at one time. Avoid “large meals”.
5. Include probiotics – yogurt, kefir
6. Keep the person upright after eating; go for a walk.
7. Make sure to provide fluids and adequate hydration. (Liquids pass through the stomach quickly with minimal work.)
8. In severe cases, a pureed diet may be needed.



TOO MANY LIQUIDS (WATER INTOXICATION)

Did you know that drinking too much liquids – especially at one time – can make you sick?

When anyone drinks large quantities of liquids (especially water), it disrupts the balance of _____ in our body and can cause – confusion, dizziness, muscle weakness, headache, stomach upset and could even cause seizures.

- Because of an altered pain response, people with PWS may not complain of a headache or stomach upset.

Things You Can Do:

1. Do NOT allow the person with PWS to purchase and drink very large quantities of drinks – even if they have no calories.
2. DO offer water or other liquids in 6-8 ounces every 2-3 hours.
3. If you suspect a problem, seek medical care.

CONSTIPATION

Constipation is a common problem in persons with PWS. It takes longer for food and waste to pass through their intestines.

Oftentimes, people with PWS have a bowel movement every day but it is small and there is incomplete emptying of their colon. That means they can be having BM's and be constipated.

According to the American College of Gastroenterology, constipation is defined as unsatisfactory defecation (pooping) with either infrequent stools, difficult stool passage or both. Any recent change in bowel habits, if persistent, may be cause for concern.

COMMON CAUSES IN PERSONS WITH PWS:

- Poor muscle tone and _____ contraction of the muscles in the intestinal system
- Low physical activity
- Poor water/fluid intake
- Medications
- Not enough fruits and vegetables

Things You Can Do:

1. Monitor bowel movements
 - a. Are they hard, dry and do they strain to push them out?
 - b. Use the Bristol Stool Chart as a guide (provided).
2. Offer water / fluids every 2-3 hours.
3. Provide well-balanced, low-calorie diet which includes softer, lower fiber fruits and vegetables.
4. Administer laxatives and follow a plan for managing constipation as prescribed by a health care professional.
5. Encourage activity, especially _____. Walks – even short ones - can help.
6. Try a “Squatty Potty”. A stool device designed to help position your colon into a squat position to help a person have a BM.



BODY TEMPERATURE REGULATION PROBLEMS

The area of the brain, called the hypothalamus, helps us regulate our body temperature.

This area does not always work properly in people with PWS.

Most have a _____ than normal body temperature.

Many can also overheat easily.

They may not have a _____ – even when they are sick.



Things You Can Do:

1. Know the baseline temperature of the people you support. Check it twice a month.
2. If illness is suspected, even if fever is _____ – have evaluated by health care professional.
 - a. May need to educate on this aspect of health in persons with PWS.
 - b. May need to advocate for throat culture or blood test to rule out an illness.
3. People may feel cold at temperatures that are not real cold for those without PWS.
 - a. May need _____ clothes in cooler temperatures.
 - b. May need guidance in dressing appropriately for the weather.
 - c. Keep thermostat in a home to a reasonable temperature.
 - i. They may not need air conditioners set at a very cold temperature.
4. In warm temperatures:
 - a. Limit time and activities in warmer environments.
 - b. Make sure they receive adequate, cool liquids.
 - c. Encourage to dress appropriate for temperatures. Don't _____. If needed, may need to make sure they are in cooler environments if insisting on wearing warmer clothes than appropriate.
5. In very cold temperatures:
 - a. Limit time and activities outside.
 - b. Make sure that when a resident leaves the home for any outings during colder weather, they are dressed appropriately to face those conditions. The individual should wear a hat, gloves, and a coat as well as being dressed in winter clothing.
 - c. If an individual refuses to wear appropriate clothing - **avoid a** _____. Staff should take items along.
 - d. In the event of snow accumulation, icy conditions or extreme cold, staff should err on the side of caution and reschedule plans.

ALTERED PAIN RESPONSE

People with PWS do not always _____ accurately.

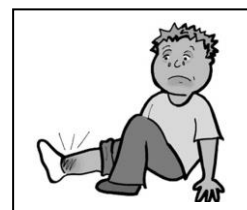
They can be injured, have wounds and painful conditions that don't hurt them very much (and sometimes not at all.)

- There have been reports of persons with PWS having a ruptured appendix who have not complained of abdominal pain.
- They may not report of a headache, sore throat or other symptoms when they have a medical problem.

This makes knowing when someone is sick or injured very challenging.

Things You Can Do:

1. Take complaints of pain _____. If any complaints persist, have them evaluated by a health care professional.



2. When a person falls or is injured:
 - a. Closely evaluate him/her at time of injury and every 4 hours for (at least) 24 hours. Watch for swelling, pain (of any kind), and an inability to use the affected area.
 - b. Seek medical care **IMMEDIATELY** if a person:
 - i. Complains of moderate to severe pain
 - ii. Has an area that looks deformed ("it just doesn't look right").
 - iii. Person is not using or moving their hand/arm/foot/knee or leg in a normal way.
 - c. Seek medical care **LATER** if:
 - i. Complaints of pain or discomfort persist.
3. Utilize _____ measures if needed – ice, elevation and rest.
4. Bring a PWSA (USA) Medical Alert booklet or use the PWSA (USA) phone app to help you educate and advocate about this aspect of PWS.
 - a. Diagnosing any medical problem or injury without using pain as a criteria can be very challenging.
 - b. Don't be afraid to ask for an x-ray or other testing to eliminate a more serious problem.

FRAGILE BONES (OSTEOPENIA / OSTEOPOROSIS)

Because of hormone deficiencies, people with PWS may have fragile bones.

- Many people take medications and/or hormones to help their bones get stronger. (Calcium, Vitamin D, Growth Hormone, estrogen, testosterone)

This problem has been found in people with PWS of all ages – even children.

This puts them at _____ of spine problems, curvatures and undiagnosed broken bones.

- As people with PWS are aging, we are seeing more problems with spine curvatures.
- Many are at higher risk for falling and experience problems with walking.
- Some require use of a walker or cane.

Low bone density (fragile bones) also makes bones break very easily.



Things You Can Do:

1. Exercise can help bones become and stay strong. Offer and encourage exercise daily.
2. Administer calcium, vitamin D and other medications as ordered by a health care professional.
3. All falls and injuries should be _____ and monitored.
4. Physical therapy, braces and daily exercises may be prescribed for spine problems.

EMOTIONAL, BEHAVIORAL AND MENTAL HEALTH CONCERNS

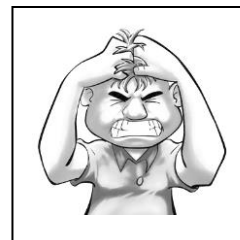
Many people with PWS face emotional and behavioral challenges.

They thrive on structure and _____. They do not handle changes well.

Food-related issues and topics often cause _____ and emotional upset.

Some may have extreme behavior outbursts resulting in destruction of property or assault of others.

There are some that also may have a diagnosis of a mental health problem – depression, bipolar disorder.



Things You Can Do:

1. *Provide structure and consistency. They do well when their day is predictable.*
2. *Don't argue with them. Talk in a _____ voice and style.*
3. *Prepare and discuss changes – especially when related to food issues. Be prepared for the person to display anxiety.*
4. *Teach, encourage and praise when they use words when frustrated and display appropriate behavior.*
5. *Putting things in writing may be helpful.*
6. *Maintain food security.*

SKIN PICKING

Many people with PWS pick at their skin and many openings of their body (nose, rectum, nails...).

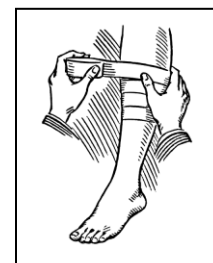
Preventing, treating and managing this problem can be very _____.

Pain is often not a factor.

Wounds can become chronic and severe. They can also be confused as a sign of abuse.

Things You Can Do:

1. *Keep hands _____. Provide stress balls or other activities that keep their hands busy.*
2. *Limit time alone.*
3. *Keep nails short. Apply lotion to keep skin hydrated.*
4. *May require supervision when going to the bathroom*
5. *Cover with bandage, clothing (or both).*
6. *In severe cases, medication may be tried.*
7. *Monitor for signs of infection – redness, warmth and/or drainage.*



MEDICATION SENSITIVITIES

Many people with PWS can be extra sensitive to many medications.

Some of the medications that can cause more problems include:

- Pain medications – especially narcotics or strong pain medications.
(Can cause extreme sedation).
- Medications used to treat diarrhea (May cause intestines to _____)
- Sedative medications (May cause over-sedation). *These are often used in procedures used to diagnose and treat problems – colonoscopy, dental procedures, and surgical procedures.
- Anesthesia (May be slow to wake up and prolonged sedation)
- Medications used to treat mood and behavior (May experience more _____ and side effects.)



Things You Can Do:

1. *Know the medications you are giving. If you think someone is having a problem or is becoming over-sedated, contact a _____ or health care provider.*
2. *Don't be afraid to help to educate health care professionals about this sensitivity.*
 - a. *Remember – you may be the primary educator to these individuals.*
 - b. *Share handouts that are available through PWSA (USA).*
3. *Encourage to start dosage low – and go slow with increases.*
4. *Limit use of medications used to treat diarrhea. They can cause intestines to totally stop functioning.*

BREATHING CONCERNS

Many people with PWS suffer from a number of different breathing problems.

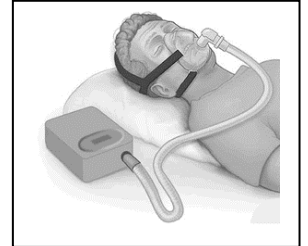
- Central hypoventilation is a disorder of decreased breathing rate or depth, particularly during sleep.
 - Causes problems with daytime sleepiness and if significant can cause problems with elevated blood pressure in the lungs.
 - Individuals with Prader-Willi syndrome may be at increased risk for this due to decreased muscle tone and mass, excessive obesity, and possibly decreased neural drive for breathing.
- Central apnea means the complete cessation of breathing during sleep.
 - There are several studies that show an alteration in the response of some individuals with Prader-Willi syndrome to chemicals that would normally increase breathing.
- Obstructive sleep apnea is well known to occur in Prader-Willi syndrome as well as in other syndromes with hypotonia (poor muscle tone) such as Down syndrome.
 - This results when the individual is trying to breathe while asleep, but due to obstruction in the airway _____ enters the lungs.

- Many of these individuals have loud breathing and snoring associated with periods of quiet where no air movement is noted.
- Untreated obstructive apnea can have serious _____ including death.

Because of weak chest muscles, they often are at greater risk of developing pneumonia when they get an upper respiratory infection.

Things You Can Do:

- *If sleep apnea is suspected, a sleep specialist should be consulted.*
- *The use of Continuous Positive Airway Pressure (CPAP) or BiPAP are often prescribed to help keep a person's airway open while sleeping. It can be challenging to get the person with PWS to use and comply with this therapy.*
 - *Use incentives to reward the person for use.*
 - *Start _____ – have the person wear it for short periods before sleeping to get used to the feel of it.*
 - *Gradually have them increase their time using it.*
- *Make sure to _____ CPAP/BiPAP equipment as instructed and order new supplies as directed.*
- *A persistent upper respiratory infection (a cold) should be evaluated by a health care professional.*



HOSPITALIZATION – SPECIAL CONSIDERATIONS

When a person with PWS is hospitalized, they will require extra support, advocacy and education of the hospital staff.

Remember, many Health Care professionals do not know about PWS and you will be the _____ and advocate for the person you support.

It is not uncommon for persons with PWS to _____ a great deal of weight while hospitalized.

Many people with PWS have cognitive or learning limitations and may not be able to provide accurate information about their condition or medical history.

Many diagnostic tests and/or procedures (including surgery) require a person to not have anything to eat or drink for a prolonged period of time. This can be very difficult for the person with PWS and they may seek out food or drink.

Some higher need areas include:

- Food security and ordering of food – most meals ordered in a hospital are ordered “restaurant style” where the person with PWS may order an extraordinary amount of food.
- IV access – many have challenges with blood testing and inserting IV’s.



- Medication sensitivity – many are sensitive to medications often utilized in a hospital setting – pain medications, sedatives, and anesthesia.

Things You Can Do:

- Visit the PWSA (USA) website (www.pwsausa.org) to access written handouts on the following topics: (These can be found under the “Medical tab”)
 - Hospitalization – PWS Alert for Inpatient Caregivers
 - Psychiatric Admission – Guidelines for Inpatient Staff
 - Anesthesia and PWS
 - Warning – Medication combinations may fatally impair breathing in PWS!
 - Postoperative Monitoring of Patients with Prader-Willi Syndrome Treatment Recommendations
- Talk about the above concerns with nurses, doctors, specialists, blood drawers and whoever you think needs to learn about PWS.
- You need to make sure there is _____ of the person to make sure they are not eating or drinking when they should be fasting.
- Insist on an experienced person to do blood draws and/or inserting IV’s.

RESOURCES – HELP FOR YOU

We know remembering all of these health issues can be very challenging. There are _____ you can access and use **to help you** and the health care professionals caring for the person with PWS.

- PWSA (USA) phone app
- PWSA (USA) website – www.pwsausa.org
 - Many health handouts
 - PWS Medical Alert Booklet (available for download under “Medical” section of the PWSA (USA) website or on the PWSA (USA) app)
- Physician Consultations – Physician to physician consultation
 - Contact PWSA (USA) at 800-926-4797

SUMMARY

People with PWS face some unique health challenges. In many ways, their body works different.

YOU are a very important part of the team that this person needs to safely access health care promptly. You are an educator and advocate for this person.

Additional Handouts:

PWSA Medical Alert

Bristol Stool Chart

Hospitalization – PWS Alert for Inpatient Caregivers