**SESSION 3 – SPEAKER: BARB DORN RN, BSN**

**General Information for Patients in the Emergency Department: Things to Know & Advocacy Tips**

The following are some of the basic facts about the health issue, general information for patients in the Emergency Department (ED), as well as some advocacy tips that parents and caregivers can utilize as they educate and advocate for the person with PWS.

**HEALTH TOPIC: General Information for Patients in the Emergency Department**

**THINGS TO KNOW:**

* Trips to Emergency Department or Urgent Care – very stressful.
* If you can go to your PCP or an Urgent Care – do it.
* Most common reasons people utilize ED for persons with PWS:
  + GI issues
  + Choking
  + Bone or joint injuries
  + Psychiatric emergencies
* Goal of ED – complete a medical screening to facilitate diagnosis and treatment for people experiencing urgent or life-threatening health situations.
* Truths: People with PWS can be a diagnostic challenge for all clinicians. Their bodies work different and parents and guardians must often be the ones to make sure that the health care professionals understand this.
* Challenges seen in persons with PWS:
  + Increased pain tolerance
  + Respiratory vulnerability – especially if overweight
  + Poor muscle tone
  + Poor temperature regulation
  + Lack of vomiting – NORMAL (Must not allow them to think it’s just a stomach virus)
  + Sensitivity to medications – Always warn them of this.
  + Biggest challenge will be to make the health care professional believe what you are saying.
* Things to Keep in Mind:
  + Don’t expect the ED staff to be familiar with PWS.
  + Be prepared to be the teacher. Be patient. Wait time is often very long. You may be teaching to many different clinicians over and over again.
  + ALL GI ISSUES MUST BE TAKEN SERIOUSLY
    - Binge eating is biggest risk factor for gastric rupture.
  + BONE / JOINT AND SOFT TISSUE SWELLING – REQUIRE AN XRAY

**ADVOCACY TIPS:**

* + Come Prepared to educate and advocate:
    - Medical Alert booklet
    - GI chart
    - List of current medications
    - Contact information for Primary Care Provider, Psychiatrist or other applicable specialists
    - Phone number to PWSA l USA to contact Family Support staff **- (941) 312-0400**
    - Up to Date website very helpful to Health Care Provider
  + Collaborate – be a part of the team –YOU are a valuable asset.
    - Share information – It’s okay to generate a sense of urgency.
      * Give a quick overview of PWS focusing on the issue you are there for.
      * Share fears
    - Stay calm and respectful – aggressive but not confrontational
  + Negotiate – No one goes home with nausea, vomiting or extreme pain
    - Consider consultation
      * If willing provide with PWSA l USA phone number to consult with a PWS expert
    - Ask to speak to a social worker – may be able to support and strengthen your advocacy efforts.
    - If needed – request transfer (especially in pediatric cases to a Children’s Hospital).
    - If at impasse – request to talk to someone higher (especially true if dealing with a medical student, resident, Nurse Practitioner or Physician Assistant)

**Anesthesia and Postoperative Monitoring: Things to Know and Advocacy Tips**

The following are some basic facts about health issues for the person with PWS in a health care setting including anesthesia and postoperative monitoring, including some advocacy tips that parents and caregivers can utilize as they educate and advocate on behalf of the person with PWS.

**HEALTH TOPIC: Anesthesia and Postoperative Monitoring**

**THINGS TO KNOW:**

* People with PWS who are in need of any kind of anesthesia, surgery or any procedure that requires anesthesia must have close monitoring and needs special considerations.
* There are 2 ways a person may receive anesthesia that affects their consciousness – general anesthesia and conscious sedation.
  + **GENERAL ANESTHESIA** – Medication is administered by a specialized health care professional (anesthesiologist or nurse anesthetist) that causes the person receiving it to become unconscious. They typically need help with breathing and close monitoring of their heart, blood pressure and breathing.
  + **CONSCIOUS SEDATION** - Medication is also administered by a health care professional that has received special training. The person receiving this becomes very sleepy but may not become totally unconscious. They do not need help with breathing, but still require close monitoring of their heart, blood pressure and breathing. People who receive conscious sedation may be able to follow commands and move. Many people with PWS are more sensitive to this type of medication and need to be closely monitored. This often does not happen.
* Surgical and anesthesia risk factors.
  + **OBESITY** – This can cause many problems, especially with breathing and the balance of oxygen and other things that are needed for a safe surgical or treatment response. People who are obese do not breathe as deep as normal and may also have obstructive sleep apnea. Obesity causes more stress on their lungs and heart because of the extra weight. Many people have other experience other problems related to obesity including the development of blood clots, diabetes or poor healing. All people with PWS who are obese are at increased risk of experiencing complications with either type of anesthesia.
  + **HIGH PAIN THRESHOLD** – People with PWS do not typically respond to pain the same as others. The challenge is that pain can be a signal of a problem. If the person with PWS is experiencing a lot of postoperative or post-procedure pain, all possible problems should be investigated.
  + **TEMPERATURE INSTABILITY** – The hypothalamus does not work properly in persons with PWS and this may impact their ability to maintain the proper body temperature. Our body’s thermostat is located in the hypothalamus. It **does not** appear that people with PWS are at greater risk for a condition called malignant hyperthermia. This is a life-threatening reaction that occurs on administration of general anesthesia, which results in dangerously high body temperature, rigid muscles or spasms, and a rapid heart rate. Fever may not be present in case of infection.
  + **THICK SALIVA/ NARROW THROAT** – Both of these issues are common findings in persons with PWS. These can impact the ease for the health care professional to insert and remove the breathing tube.
  + **FOOD SEEKING BEHAVIORS** – It is important for people who are receiving anesthesia to not have any food in their stomach several hours before this is performed. Having no food in the stomach, prevents the risk of aspiration during a procedure. It’s very challenging for people with PWS to not eat or drink for an extended period of time. If they do sneak some food – they may not admit to doing so. Extra surveillance is needed during times of fasting.
  + **LOW MUSCLE TONE / RESPIRATORY CONCERNS** – Many infants, children and adults have poor muscle tone. The primary risk associated with this is that these individuals may not take as deep breaths or cough as strong as is needed to prevent respiratory problems. Obesity, spine curvatures (like scoliosis), sleep apnea and sedating medications also add to respiratory concerns.
  + **SKIN PICKING** – Many people with PWS have a significant problem with picking at their skin or any openings. They often pick at IV insertion sites, tubes and incisional wounds.
  + **DIFFICULTY GETTING IV STARTED** – It is often difficult for clinicians to find veins for inserting an IV or for obtaining a blood sample.
  + **BEHAVIOR CONCERNS** – Some people with PWS have more challenges with emotional outburst, anxiety and other psychiatric illnesses. The surgical experience, along with all of the preparation and changes in routine, can often increase these behaviors. The actions and effects of many of the medications may be impacted with anesthesia. Anything that impacts them having food will result in increased anxiety.
  + **RECOVERY** – It takes many people with PWS longer to wake up after receiving anesthesia. They appear to be more sensitive to medications that cause sedation. They should be closely monitored and have a pulse oximeter (a device placed on the finger to monitor their oxygen level) in place. You want to make sure that breathing and alertness returns to normal before considering discharge.
  + **GI MOVEMENT** – People with PWS are very sensitive to anything that can slow their gastrointestinal system. GI conditions or surgery, anesthesia, pain medication and decreased activity (walking) are often factors that cause things to slow down. Extra care is required when advancing food. It must be taken slowly after surgical or other procedures where these are impacted. If problems occur, x-rays may be indicated.

**ADVOCACY TIPS:**

* *Share the Anesthesia handout with all health care professionals. Make sure the anesthesiologist is made aware of their sensitivity and that the hypothalamus is involved in PWS.*
* *Patients with PWS who undergo deep sedation and general anesthesia should be recovered overnight in a monitored unit where their heart and breathing rate is closely monitored. Infants and children may require intensive care monitoring.*
* *Any dental problems should be evaluated and corrected before having a procedure.*
* *Strongly advocate that the person with PWS be one of the 1st cases or procedures in the day. Make sure extra supervision takes place to make sure the person with PWS does not eat or drink as advised.*
* *Strongly advocate for aggressive respiratory therapy and movement (walking) as soon as possible after surgery.*
* *Make sure IV’s, tubes & wounds are closely monitored and covered with extra dressings or clothing to prevent picking.*
* *Ask for the most skilled person to insert an IV or do a blood draw. (An anesthesiologist is often this person).* 
  + *For IV insertions and obtaining blood samples, have the person with PWS sit up and applying a warm blanket or warmth to the area to help veins come to the surface.*
  + *Ask for use of ultrasound if available.*
* *If possible, someone should remain with the person with PWS through the entire experience. They can serve as an advocate, lessen anxiety and help with food security.*
* *Be sure that all health care providers are aware of any mood-altering medications so that appropriate monitoring can take place.*
* *Make sure that the person with PWS is alert and their breathing is stable before offering food or being discharged. If they are very sleepy, they are greater risk of choking or aspirating. People with PWS require a longer period to wake up and recover from anesthesia. (Sometimes outpatient settings are in a hurry to discharge people).*
* *Share the GI chart with health care professionals if any sort of stomach distention, obstruction or problems with severe constipation occurs. If problems develop, x-rays and other testing may be needed.*

**Medication Sensitivity: Things to Know and Advocacy Tips**

The following are some of the basic facts about medication sensitivity, as well as some advocacy tips that parents and caregivers can utilize as they educate and advocate on behalf of the person with PWS.

**HEALTH TOPIC: Medication Sensitivity**

**THINGS TO KNOW:**

* People with PWS are extra sensitive to many medications – especially ones that can cause sedation. Medicines often stay in their system longer than others.
* Anesthesia, pain medications and medications used to treat mood or behavior problems are some of the most common medications that can cause an exaggerated response.
* A slow, conservative approach to the use of these medications is often needed. “Start low and go slow” is a good approach to use.
* Medications that impact the functioning or motility of the stomach and gastrointestinal tract can also cause a more severe response. These medications are similar to those causing sedation but medications used to treat diarrhea can also cause problems.

**ADVOCACY TIPS:**

* *Share a list of all medications (including vitamins and supplements) the person with PWS is taking with the health care professional.*
* *Share the Medical Alert booklet and highlight the section that shares information about medication sensitivity.*
* *Use extreme caution in using medications to treat diarrhea. Monitor closely for abdominal distention and/or severe constipation*

**Food Security in a Health Care Setting: Things to Know and Advocacy Tips**

The following are some of the basic facts about the health issue, food security in the health care setting, as well as some advocacy tips that parents and caregivers can utilize as they educate and advocate on behalf of the person with PWS.

**HEALTH TOPIC: Food Security in the Health Care Setting**

**THINGS TO KNOW:**

* Managing food security while in a health care setting can be very challenging. There are many variables and many people that become a part of the treatment team, that may not have a clear understanding of the intensity of food seeking that can be present in a person with PWS.
* There are numerous opportunities for the person with PWS to access food sources – roommate’s tray, open kitchen on the nursing unit, staff break room, trays that have been picked up from other patient rooms, and the ease in ordering food (like carryout) at almost any time.
* There is a strong need for oversight when the person with PWS should not eat or drink. This can be very challenging.
  + If a tray is accidentally served when the person should be fasting – they will eat it.
  + They may drink water from a sink in the room and in some cases, water from a toilet.
* There have been cases of binge episodes of both food and beverages, as well as extraordinary weight gain, while the person with PWS is hospitalized or in a rehabilitation facility.
  + It’s important that a calorie restricted diet is maintained.
  + Limit quantity of fluids that are provided to prevent “water intoxication.” This is a condition when too many liquids (especially water) disrupt the chemical balance of sodium in the body and affects how the kidneys get rid of fluids. Individuals can headaches, fatigue, frequent urination, nausea, vomiting and confusion. In severe cases, there can be loss of consciousness.
  + Check weight daily. An unexpected weight gain should be considered a warning sign.

**ADVOCACY:**

* *The person with PWS needs supervision and advocacy while they are hospitalized.*
* *Request a nutrition /dietary consultation. Make sure everyone becomes informed of dietary and food seeking needs.*
* *If possible, the person with PWS should be in a private room (to limit easy access to another person’s food or tray).*
* *Make sure there is constant monitoring of food carts that may either be delivering food or storing dirty trays. Promptly remove food trays. People with PWS will eat food from plate where there’s food is remaining.*
* *Put a block on food being ordered on the phone. Daily visits by dietary staff for meal and snack preferences/choices should be utilized.*
* *Limit access to the kitchen on the nursing unit. Check waiting room areas. There may be beverage machines or other food items for families and visitors.*
* *If possible, ask that one staff person be assigned responsibility for giving the person all food and beverages. This will prevent the individual from receiving repeated items from different people.*
* *The person with PWS should not go off of the unit without 1:1 supervision. They may get access to vending machines or gift shops.*
* *Keep break room doors closed to prevent access to food or purses. (Purses have money and money buys food).*