

Warning – Medication combinations may fatally impair breathing in PWS!

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We want to remind parents that impaired breathing is a leading cause of death in people with PWS. After the recent unexpected death at home of a young person with PWS, we want to alert parents and professionals to the potential risk factors with combinations of medications, especially after surgery. PWSA | USA continues to collect and analyze valuable data from families who generously offer information after a death has occurred.

Communicate about medications with every health care provider

Bring a detailed list of every supplement and medication by name and dose to every health care visit, and be sure this is updated at each office, every visit. It is also helpful to bring all current medications to each visit. This includes vitamins, dietary supplements (such as MCT oil) or over-the-counter supplements (such as coenzyme Q 10 or melatonin); allergy medications (Flonase, Zyrtec). Some over-the-counter products can play a part in creating serious drug interactions (such as Benadryl). Well tolerated prescription drugs (such as clonidine) can cause increased sleepiness and decreased breathing after surgery or in combination with a new pain medication.

Medication issues unique to PWS

We know that people with PWS often absorb and keep medications in their system longer than the average person. The medications of greatest concern include prescription pain medications, anesthetics, and psychotropic (behavior) medications, all of which have the potential to depress breathing in any person. Combinations of several new medications create the most worrisome situation and is most likely to occur after surgery.

Anesthesia: While your child will be monitored closely during the procedure and in the recovery area, remember that it often takes longer for the effects of anesthesia to wear off in individuals with PWS. This is a greater concern in out-patient surgeries, where the goal is to send the patient home as soon as possible. We believe it is important for persons with PWS to be monitored longer (with many people with PWS staying overnight) to be sure that breathing and alertness have returned to normal.

DO NOT allow your child to eat after surgery until they are fully awake and alert. Insist that the progression from liquids to solid food take longer than the surgical team usually suggests. Choking is a significant risk in PWS and it increases with sedation.

Gastroparesis (stomach stops sending food to the intestines) can occur after anesthesia which can create a blockage, which is a medical crisis. This can be avoided if the digestive system is given more time to “wake up”.

Pain medication: People with PWS are less sensitive to pain than typical people and lower doses are recommended. The combination of anesthetics and pain medication can lead to excessive sedation and decreased consciousness. Since both of these substances may take longer to clear in individuals with PWS, great caution is needed.

What are the warning signs?

A valuable and early sign is a change in your child's personality, alertness, liveliness, talkativeness or sense of humor. After a surgical procedure, do not take a child home from the hospital on pain medication who "looks medicated." Let the professionals monitor your child in a safe place until they are fully recovered.

Breathing issues are always an emergency. Sleepiness may be a sign of slowed breathing (too out of breath to stay awake). Lip color should be monitored if sleepiness occurs – bluish lips are an emergency. The person who is overweight is always at a higher risk for breathing problems.

Pain is also a warning sign. Most pain medications and anesthesia slow down digestion, and abdominal pain can occur. Take abdominal pain or a distended abdomen very seriously and head to the ER when this occurs.

PWSA|USA has additional handouts explaining the gastrointestinal, anesthesia and surgery considerations for persons with PWS on our PWSA app and our website – www.pwsausa.org.

Reviewed by Elizabeth Roof, MA, as well as members of the PWSA | USA Scientific Advisory Board (Merlin G. Butler, MD, PhD) and Clinical Advisory Board (Janice Forster, MD, Linda Gourash, MD, and Daniel J. Driscoll, MD, PhD).

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