Prader-Willi
SYNDROME ASSOCIATION | USA
SAVING AND TRANSFORMING LIVES

Anesthesia and Prader-Willi Syndrome

by James Loker, MD Laurence Rosenfield, MD

Obesity

Obese individuals are more prone to obstructive apnea, pulmonary compromise, hypoventilation, and diabetes. Each of these should be taken into account when preparing for anesthesia. The individual may have altered blood oxygen or blood carbon dioxide levels that will change their response to medications including oxygen. Pulmonary hypertension, right-heart failure, and edema may necessitate evaluation by a cardiologist or pulmonologist prior to surgery. An ECG or echo to detect right ventricular hypertrophy may be beneficial to assess pulmonary hypertension. Frequently obese individuals with PWS may have significant body edema (extra fluid) that is not fully appreciated due to obesity. This should be carefully evaluated, and if necessary, diuretics used before and after the anesthesia. Airway management can be a particular problem when conscious sedation is used and appropriate safeguards should be in place.

High Pain Threshold

Individuals with PWS may not respond to pain in the same manner as others. While this may be helpful in post-operative management, it may also mask underlying problems. Pain is the body's way of alerting us to problems. After surgery, pain that is out of proportion to the procedure should alert the physician that something else may be wrong. Other possible signs of underlying problems should be monitored.

Temperature Instability

Because PWS is a disorder involving the hypothalamus, individuals may be either hypo- or hyperthermic. The parent or caregiver can be helpful in letting the anesthesiologist know what the individual's usual temperature is. Although there is no indication of a predisposition to malignant hyperthermia in PWS, depolarizing muscle relaxants (i.e., succinylcholine) should be avoided unless absolutely necessary.

Thick Saliva

A common problem in PWS is unusually thick saliva. This can complicate airway management, especially in cases of conscious sedation or during extubation (when a breathing tube is removed). Thick saliva also predisposes an individual to dental caries (cavities) and loose teeth. Oral hygiene should be evaluated prior to anesthesia.

Food-Seeking Behaviors

It is vitally important that any individual undergoing general anesthesia or conscious sedation have an empty stomach. This reduces the risk of aspiration of the stomach contents into the lungs. Individuals with PWS generally have an excessive appetite and may not tell the truth if they have eaten just prior surgery.

Any individual with PWS should be assumed to have food in the stomach unless it is verified by the caregiver that they have not eaten. A tube may need to be placed in the stomach to assure no food

is present prior to attempting to place the breathing tube. Some individuals with PWS may ruminate (regurgitate some of their food) and are at higher risk of aspiration.

Hypotonia
The majority of infants with PWS are significantly hypotonic. This usually improves by 2-4 years of age. The majority, however, continue to have lower muscle tone than normal individuals. This may be a problem in the ability to cough effectively and clear the airways after use of a breathing tube.

Skin Picking
Habitual skin picking can be a significant problem in PWS. This can complicate healing of IV sites and incisional wounds. Usually if these remain well covered, they will be left alone. Depending on the individual's cognitive impairment, restraints or thick gloves may be needed to protect surgical wounds during healing.

Hypothyroidism
Although the incidence of hypothyroidism in PWS is not known, low levels of thyroid hormone could occur due to lack of thyroid stimulating hormone or thyroid releasing factor, not necessarily due to problems of the thyroid gland itself. If not already done, a check of thyroid hormone levels may be beneficial in the preoperative evaluation.

Central Adrenal Insufficiency
There are conflicting studies as to the incidence of CAI in PWS. Early papers reported a 60% incidence but more recent papers show a much lower rate (5-15%). In any patient with PWS that is having problems post-surgery, a cortisol level should be drawn and if appropriate, a stress dose of steroids given.

Difficult Access
Due to several problems including obesity and lack of muscle mass, individuals with PWS may pose difficulties with insertion of an intravenous line. Individuals with PWS may have smaller airways and veins than would be expected for their body size. A stable IV line should be present in any individual undergoing anesthesia.

Behavior Problems
Individuals with PWS are more prone to emotional outbursts, obsessive-compulsive behaviors, and psychosis. They may be on extensive psychotropic medication, and the possible interaction of these medicines with anesthesia should be appreciated.

Recovery Post Anesthesia
Drowsiness after anesthesia may be due to the underlying somnolence and a component of central apnea or hypoventilation. For typical outpatient procedures, consideration should be given to an overnight observation.

GI Motility
Individuals with PWS tend to have decreased GI motility and are prone to severe gastric distension, obstruction, and constipation that can be life threatening. Care must be taken when starting feeding after anesthesia. A GI algorithm is available from PWSA to help with assessing gastric distension in PWS.

Summary

In individuals with Prader-Willi Syndrome there are health issues that can alter the course of anesthesia. The majority of complications do not appear to come from general anesthesia, which is always closely monitored, but from poorly monitored conscious sedation. Any individual with PWS undergoing either general anesthesia, or conscious sedation should be considered a high - risk patient and appropriate safeguards put in place.

Edited 2/2