



Presents:



When Your Baby is in the Hospital NICU

From the series, "A Package of Hope"

PRADER-WILLI SYNDROME ASSOCIATION | USA WHEN YOUR BABY IS IN THE HOSPITAL NICU

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All pictures included in “When Your Baby is in the Hospital NICU” feature infants and toddlers who have Prader-Willi syndrome.

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Dedicated to all families living with Prader-Willi syndrome.

Your Baby Has Just Been Diagnosed with PWS... Now What?

Prader-Willi syndrome is a condition your baby has, not who your baby is. You are probably feeling shocked and scared and certainly worried about your baby with PWS. You may feel like your world has turned upside down and you are desperate for information about your infant's current care and future. What you may not appreciate right now, but will in the future, is that this new addition to your family is full of potential, personality, and love. Though it may be difficult to imagine now, your child with Prader-Willi syndrome will bring you overwhelming joy.

In the hospital, the focus may be on what's "different" about your child, but pause and take moments to enjoy and embrace the perfect beauty of YOUR baby. It will take time for you to process this diagnosis and its implications but there are things you can do now to help your baby.

- **Become informed, not overwhelmed.** Unfortunately much of the information regarding PWS on the internet is dated and misleading. However, Prader-Willi Syndrome Association | USA is an exception. One of the main missions of PWSA | USA is to bring families like yours the latest treatments, information, and support. Contact PWSA | USA to receive the **Package of Hope** with booklets on PWS infant feeding, growth hormone treatment, physical and other therapies, the child with PWS 0-3, and a grandparent booklet. To contact PWSA | USA, please call (941) 312-0400 or visit www.pwsausa.org. You can mail the Prader-Willi Syndrome Association | USA at 1032 E. Brandon Blvd, #4744, Brandon, FL, 33511. Encourage your family members to contact PWSA | USA with questions or for support. Your family and friends will want to help and PWSA | USA can teach them how.
- **Keep a notebook with your questions, and ask them!** Doctors spend a short time with you, so it is easy to forget the questions you want to ask as new information is being given to you. Most doctors appreciate the opportunity to answer questions especially when they are presented in an organized fashion.
- **Be a partner in the care of your baby.** Prader-Willi syndrome is a rare genetic disorder and as a result, many physicians have very limited experience with its treatment. Being able to provide some of the most recent literature to your child's physicians may be much appreciated. Your physicians are also encouraged to contact PWSA | USA with any questions or to request information.



While Your Baby is in the Hospital NICU

Is my child's blood sugar being monitored and treated?

In the recent study, "*Hypoglycemia in Prader-Willi Syndrome*," researchers noted that "the data raises concern that infants with PWS are predisposed to hypoglycemia from birth, a new finding." Hypoglycemia or a low blood sugar level is associated with brain developmental delays. Receiving inadequate oral feedings may contribute to hypoglycemia. ("*Hypoglycemia in Prader-Willi Syndrome*," *Rena A. Harrington, David A. Weinstein, and Jennifer L. Miller; Pediatrics, University of Florida, Gainesville, FL*).

Is my baby's oxygen level being monitored, especially during sleep and after feeding?

Oxygen is important for your infant to grow and develop. There is 21% oxygen in the air we breathe every day. Some infants may require supplemental or higher oxygen levels under specific circumstances. One of the ways that doctors can tell if your infant is getting enough oxygen is by using a pulse oximeter. This is a non-invasive monitoring device that is generally attached to the toe and monitors the oxygen saturation. Since problems can occur when oxygen levels are too high as well as too low, monitoring helps the medical team find the most appropriate level for your infant.

Infants, particularly those born early, may have irregular breathing patterns. If you notice this, bring it to the nursery staff's attention so it can be evaluated. Many infants with Prader-Willi syndrome have apnea due to low muscle tone during sleep when muscle tone is further decreased. This means that as their bodies relax during sleep, their airway may become temporarily blocked causing them to stop breathing for short periods of time. They are also prone to gastroesophageal reflux (see below) which can cause a similar phenomenon called hypopnea (episodes of overly shallow breathing or an abnormally low respiratory rate). If your infant does have apnea it can be treated. The type and severity of apnea will help your physician determine the best course of treatment. Once your child demonstrates a normal oxygen saturation while breathing room air, your baby should be able to go home without supplemental oxygen. If your baby does need to go home on oxygen, it will eventually be weaned by his/her physician.

How can I get more skin to skin contact with my baby and why is that important?

Studies have shown that skin to skin contact with your child has numerous benefits for you and your baby. As examples, babies stay warmer, they cry less, and they have decreased levels of stress hormones. They also breathe easier and are more stimulated to feed. The longer and more often skin to skin contact can occur during the days following birth, the greater the benefit. Skin to skin contact with the father has been shown to be just as beneficial to baby as it is when done with the mother. In spite of the fact your baby sleeps a lot and may be connected to many tubes, don't be afraid to ask for more time to hold your child. Skin to skin contact is great for your baby and allows all of you to bond, even while your baby sleeps. Some ways to get more skin to skin time include holding baby before, during, and after all feedings. If you are staying overnight at the hospital with your baby, nighttime is a great time to get long stretches of uninterrupted time with your baby.

But also, make sure you are getting the rest you need as well. Ask for help from your baby's nurse to facilitate skin to skin contact with your baby as much possible throughout your hospital stay.



Are my baby's feeding issues being addressed?

The initial phase of PWS is characterized by failure to thrive (failure to adequately gain weight and grow). During this time it is important your baby receives the necessary nutrition for growth and brain development. Your baby's healthcare team will develop feeding and follow up plans prior to discharge from the NICU. You will also receive an *Infant and Nutrition Booklet* from PWSA | USA. If the nurses in the NICU also want that information, the contents can be emailed on request.

- A Registered Pediatric Dietician should be addressing your baby's nutritional needs in the NICU. They will monitor your baby's weight gain and growth, assess what your baby is eating, and make recommendations regarding feeding fortification to assure your baby's body gets what it needs. If your baby is not able to get enough nutrition orally, as is common in babies with PWS, your baby may require a feeding tube. The feeding tube is usually temporary and typically very short term (6-8 weeks). A nasogastric tube or NG tube (a small tube that is inserted into the nose down into the stomach) is typically recommended. NG tubes are easy to maintain/manage at home and promote a more timely transition from tube feeding to oral feeding. Gastric tube or G tube (a tube that is surgically placed through the abdomen into the stomach) is not typically recommended unless supplemental feeding will be needed more than 2 to 3 months.
- An Occupational Therapist (OT) and /or Speech Language Therapist (SLP) should be consulted if your baby is having ANY issues with oral feeding. Common PWS feeding issues include: fatigue, difficulty sucking, difficulty swallowing, and risk for aspiration (entrance of milk into baby's lungs or "going down the wrong pipe" because baby may not be able to protect airway). The therapist will assess and recommend feeding techniques/strategies, positioning, and any equipment that might assist your baby. They may also recommend a swallow study if one has not already been ordered.
- A Lactation Consultant may assist with breast feeding techniques if your baby is able. Since breast milk is considered the best nutrition for your baby, a lactation consultant can help you maintain your breast milk supply if you have to pump to feed with bottle or via NG tube.

- Most babies with PWS are at high risk for aspiration (fluid or food entering the lungs), which can cause very serious pneumonia. Their low muscle tone can result in difficulty protecting their airway, with or without a feeding tube present. Infants with PWS are also particularly prone to **Gastroesophageal Reflux (GER)**, which can lead to “silent aspiration” (stomach contents entering the lungs without showing signs at the time). Strict aspiration precautions should be observed until GER is well controlled and a videoflouroscopic swallow study can be performed and further recommendations made.

Aspiration precautions if gastroesophageal reflux is an issue:

- Baby’s head should be held upright during feedings 45-60 degrees
- The head of baby’s bed should be elevated to a 30 degree angle.
- Avoid lying baby flat when possible.
- GER can be treated with medication if the problem persists



Some infants with significant silent aspiration or severe reflux may warrant specialized consultation with a pediatric gastroenterologist with consideration of placement of a g-tube and in rare cases surgical procedure to address severe reflux.



For additional information on reflux management, go to the NASPGHAN public website: www.gikids.org.

Have the appropriate specialists been contacted to evaluate my baby in the hospital?

Your baby is likely being followed by the **NICU team** of doctors, who are specially trained pediatric physicians. Your baby has probably also been seen by a **geneticist** if you already have a PWS diagnosis. This is a doctor who specializes in diagnosing genetic disorders. If you have not already done so, it is important for you to follow up and see a medical geneticists and/or a genetic counselor to discuss the sub-type of PWS plus the expected course and management of the syndrome.

It is important that you educate yourself on **growth hormone therapy (GHT)** for PWS. You may want to consider a consultation with a **pediatric endocrinologist** while they are in the NICU, or arranging an appointment for after discharge, preferably with an endocrinologist that has experience in treating children with PWS. The FDA approval for growth hormone therapy in the US is specifically for poor growth, but studies have found other benefits from GH treatment in infants with PWS, such as improvement for muscle tone, so parents need to discuss the risks and benefits of starting GHT with the endocrinologist. GHT for the low muscle tone associated with PWS is now being initiated in some situations by PWS experts as soon as weeks after diagnosis in newborns, but there is no current published data supporting the benefits of GHT until the baby is between 4-6 months of age.

The reason for a consult while in the NICU is that it may take 1-2 months to get an outpatient appointment with an endocrinologist. Additionally, testing required prior to growth hormone initiation may take weeks. Approval from your insurance company will be required which could result in further delays. Seeing a pediatric endocrinologist to discuss growth hormone for your baby while in the hospital (or if this is not practical, set up the appointment while your baby's in the NICU for 1-2 months after discharge) will help ensure that your baby may be started on growth hormone by the age of 4-6 months if not sooner. If the endocrinologist is not an expert with PWS, please refer to the article referenced below that is an international consensus statement published in 2013 by *The Endocrine Society's Journal of Clinical Endocrinology & Metabolism* with your baby's endocrinologist and the growth hormone booklet provided by PWSA | USA to new parents.

Often, infants with PWS have sleep apnea due to low muscle tone. A **pediatric pulmonologist** (lung doctor) will typically be the specialist to follow your baby for this. A sleep study should be completed prior to starting growth hormone therapy to obtain a baseline. However, an abnormal sleep study is NOT necessarily a reason to withhold growth hormone therapy. In fact, growth hormone therapy started in infancy will often improve obstructive sleep apnea due to low muscle tone (JCEM 2006 Feb; 91(2):413-7 *Short-Term Effects of Growth Hormone on Sleep Abnormalities in Prader-Willi Syndrome*). Infants with PWS at ages preceding the development of any obesity already demonstrate some degree of sleep-disordered breathing. They are two types of apnea a child with PWS may have – central and obstructive.

It is very common for newborn baby boys with PWS to have undescended testicles. A **pediatric urologist** should be consulted and a plan created to address this issue. Typically, a “wait and see” approach is taken to see if the testicles will descend on their own. If they do not, surgery may be required at a later date, but surgery is not recommended until the child is at least six months old, so this consult is not urgent and can typically wait until after discharge if you are feeling overwhelmed by specialists.

Your baby's discharge from the hospital will require planning and coordination of resources. The hospital based social worker should be able to streamline this process for you. Be sure to ask for help with any of your concerns for your baby's care at home.

Has my baby been checked for strabismus (cross eyedness) or have plans been made to evaluate for this after discharge?

Strabismus (cross eyedness) is a fairly common finding in children with PWS and will need to be assessed and treated by an ophthalmologist. It is appropriate to wait and schedule this appointment after hospital discharge. Typically, babies do not develop conjugate gaze until the first few months of life so cannot be evaluated for strabismus until then. Recommendations from the American Academy of Pediatrics (Clinical Report – Health Supervision for Children with Prader-Willi Syndrome, Pediatrics Jan 2011 127(1):195-204) are to screen for strabismus during well baby visits in the first year of life and refer to a pediatric ophthalmologist if strabismus is detected or suspected.

Should my baby get the Synagis vaccination for RSV (Respiratory Syncytial Virus)?

Acute infections due to a respiratory virus, known as Respiratory Syncytial Virus or RSV are common in childhood. In the United States, the peak activity for this virus is typically between November and March, but this can vary from year to year and region to region. In fact, most children have had the infection during the first year of life.

Due to their low muscle tone and the potential for adrenal insufficiency, infants with Prader-Willi syndrome are at risk for serious complications with RSV including respiratory failure. Synagis [si-nahjis] is an FDA-approved prescription injection of antibodies that is given monthly to help protect highrisk infants from severe RSV disease throughout the RSV season. Annually, recommendations come from the CDC regarding who qualifies for Synagis. PWS has not traditionally been thought of as a qualifying condition, and many Synagis programs will not accept patients with PWS, and some insurance plans will not cover this expensive treatment.

While there is no consensus about administering Synagis routinely to infants with Prader-Willi syndrome, the Red Book® states that “*immunoprophylaxis may be considered for infants who have either congenital abnormalities of the airway or a neuromuscular condition that compromises handling of respiratory secretions.*” This could be considered applicable to PWS. Speak with your doctor to see if Synagis can benefit your infant. (Reference: American Academy of Pediatrics. Red Book: 2012 Report of the Committee on Infectious Diseases. Pickering LK, ed. 29th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2012.) It is up to your baby’s pediatrician or PCP to request the continuation of synergist administration during RSV season, even if synergist is initiated while the baby is in the NICU.

There are other preventative measures that parents can take to decrease the risk of RSV infection. These include: avoidance of exposure to tobacco smoke, crowded areas and situations where exposure to infected individuals may occur. Attention to good hand hygiene is always important but is especially so during RSV season. Breast feeding, which is felt to be protective against many viral illnesses has not been confirmed to be protective against RSV infection.

Taking care of yourself is taking care of your baby.

As they say on the airplane, “*Put your oxygen mask on first before helping others.*” Good physical and mental health of the parents is central to the long-term care of their baby. This means ample sleep and regular meals for mom and dad, as much as this is possible with a newborn. Communicate to others what they can do to help you. Also know that you are not alone, PWSA | USA is here to help and to connect you with other families who truly understand.



The pictures above feature Nick Joncas who has PWS. Many thanks to Nick's parents, Colette and Eric Joncas, the primary authors of *When Your Baby is in the Hospital NICU*.

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