

Careful Steps – Newborn Girl

First and foremost, CONGRATULATIONS on your new baby! While you may feel very overwhelmed right now, we want to assure you that it is all going to be O.K. Your child will amaze you in ways you may not feel are possible right now.

Below is a “To Do” list of things that should be done during this first year to make sure your child is off to the best start. Please don’t feel overwhelmed and remember, we are here to support you, your family and your child. We want to welcome you into our community and hope you feel comfortable in reaching out with any questions you may have.

Lisa Thornton – President of UPWSA

lisathornton@upwsa.org

Brooke Ramos – Support Care Coordinator

brookeramos@upwsa.org

Zeinab Bahsoun – Medical Care Coordinator

zeinab.bahsoun@hsc.utah.edu

Annabelle Wilcox–Genetics Physician Assistant

annabelle.wilcox@hsc.utah.edu

Doctors to See

✦ **Schedule First Visit at PWS Medical Clinic** – Please reach out to the Medical Care Coordinator to schedule a time to attend the clinic. At this clinic you will be seen by specialists who have experience with Prader Willi Syndrome. These specialists include a pediatric endocrinologist, pulmonologist/sleep specialist, geneticist, psychologist, psychiatrist, nutritionist, physical therapist, occupational therapist, and speech therapist. You will have follow up appointments with the endocrinologist and pulmonologist/sleep specialist. Please check with the Medical Care Coordinator to see if you need to schedule an appointment with these specialties prior to your child’s scheduled PWS Clinic appointment.

Your infant will attend the PWS Clinic twice in the first year, and then yearly after that.

Many specialists are hard to get into so schedule appointments as soon as possible.

✦ **Endocrinologist**

- **Check Infant for Hypothyroidism** –tested by a simple blood draw, with the Newborn screening.

- **Growth Hormone** – discuss the treatment of growth hormone. Human Growth Hormone is now considered standard of care for individuals with PWS in the United States. Your endocrinologist should be able to discuss the benefits and risks regarding this treatment.
- **Insurance Applications for Growth Hormone started** – this should be done at the first visit with the endocrinologist
- **Follow up visit with Endocrinologist** – Infant should be seen every 6 months for growth hormone dose adjustment and monitoring.



Pulmonologist/Sleep Specialist

- **Check Infant for Restrictive Lung Disease** – If diagnosed, be sure infant is receiving appropriate breathing treatments.
- **Monitor Oxygen Saturation Levels** – Oxygen levels need to be monitored consistently, especially during sleep, feedings and upper respiratory illness. If oxygen SATs drop below 93, oxygen is to be administered to bring oxygen levels up to at least 96, or as advised by a pulmonologist. It may be necessary for your child to remain on oxygen until the first sleep study is done. At that time the pulmonologist will be able to determine your child's oxygen needs.
- **Baseline Sleep Study** – This should be completed as early as possible but should not delay the start of growth hormone.

§ If Moderate to Severe Obstructive Sleep Apnea is detected in first study:

- Infant to be seen by ENT for tonsil and adenoid evaluation
- Infant has a GI evaluation for reflux (which can cause sleep apnea)
- Infant has had an evaluation by pulmonologist
- If CPAP is needed, infant should be successfully using CPAP as instructed by pulmonologist



Pediatrician

- **Consult with doctor on proper precautions to prevent illness** – Is your child more at risk for becoming sick than other children? Additional precautions may need to be taken, such as keeping the child home for a longer amount of time than typically developing infants.

- **Synagis shots** – for prevention of RSV during RSV season. The pulmonologist can assist in providing letters, stating the need if insurance denies coverage.

- **Check Infant for Esotropia (or Strabismus)** – This is a condition where one or both eyes cross inward looking “cross eyed”. This can be common in newborns. If the condition persists after 6 months of age, the infant will need to be seen by an ophthalmologist.

- **Discuss Supplements with Medical Provider**

 - § Iron supplement

 - § Vitamin supplementation

 - § Fish Oil

 - § CoQ10

 - § Levocarnitine

- **Infant current on vaccinations**

- **Regular Weight Checks** – height and weight should be charted at regular intervals using standard growth charts. It does not matter where your child falls on the chart but you are looking for a consistent growth curve just like any other child.

- **Calorie Enhancers** – may need to be added to breastmilk or formula if infant is not gaining adequate weight.

- **Head Circumference Measurements** – carefully monitored to detect craniosynostosis (premature closure of cranial sutures) and monitor brain growth.

- **NG or G-tube placed** – Your doctor should be able to help you decide if a feeding tube is needed for your child, and which type of feeding tube will be most appropriate. Once tube is in place, it is important that a feeding

therapist works with you to provide feeding strategies. There will be a plan set for how to wean off the tube.

- **Check for Gastroesophageal Reflux** – Treat for reflux until infant is ambulatory. Lying the infant flat on their back should be avoided, in case the infant has reflux. A 30 degree angle is a safer sleeping position for these infants.
- **Examined for Constipation** – Likely, there will be no obvious signs of constipation. If needed an oral laxative can be administered.
- **Monitor for Bladder Infections** – Again, typical signs of bladder infection (such as crying in pain or fever) may not be present.

Testing to be Done

‡ **Methylation Test** – ordered to confirm PWS diagnosis. Further genetic testing may be necessary and will be ordered by the Geneticist as needed.

‡ **Swallow Study** – Silent aspiration has been known to occur among infants with Prader Willi Syndrome due to dysphagia and low muscle tone. A swallow study will be done to ensure that your child is swallowing safely and effectively. The results of the swallow study will tell you if your child will require their bottles to be thickened and to what consistency (e.g. nectar, honey, or pudding consistency) and what thickening product to use (such as “simply thick”). The swallow study is not performed until the infant has appropriate feeding cues. The feeding therapist and your pediatrician will work together to determine this. Please contact the Medical Care Coordinator with any concerns.

‡ **Baseline X-ray for scoliosis** – This should be done as soon as the infant is able to sit solidly and independently

- **If scoliosis is detected:**

§ Growth Hormone is not generally stopped

§ If there is a degree of curvature 25-30 degrees or greater, the child is to be seen by an orthopedic physician and bracing will be discussed.

‡ **Oral Hygiene Strategies** – use soft foam toothbrushes and wetting solutions to care for gums and

teeth as soon as they start appearing.

Therapy to Enroll In

‡ **Enroll in your county's Early Intervention Program** – An order for the Early Intervention Program should be ordered prior to your hospital discharge, or through your pediatrician. An assessment will be done in your home, to determine your child's needs and write up an Individualized Family Service Plan (or IFSP). This IFSP should include physical, occupational, vision, and feeding/speech services.

‡ **Cognitive Therapy** (If available)

‡ **Physical Therapy** – through Early Intervention, an outpatient program, or both

‡ **Occupational Therapy** – through Early Intervention, an outpatient program, or both

‡ **Speech Therapy** – through Early Intervention, an outpatient program, or both

- Feeding Issues Addressed – access to feeding therapy if needed

Applications to Complete

‡ **Pfizer Bridge Program** – this program helps cover the cost of growth hormone if insurance approval is delayed for any reason. Your endocrinologist should be able to help you through the application process for this program.

‡ **Copay Assist Program** – Through Pfizer, will help cover up to \$5,000 of deductible for growth hormone prescription (genotropin)

‡ **Home and Community Supports Waiver**– In Utah this program is known as “DSPD” (Division of Services for People with Disabilities) and is administered through the Utah Department of Human Services. This program is not income based and provides a Medicaid card, as well as monetary support, based on the need of the child.
<https://dspd.utah.gov/>

‡ **Institutional Medicaid**: After discharged from hospital or even while baby is still in hospital, contact the finance office at the hospital and ask about applying for Institutional Medicaid, whatever your income. Provides a second insurance and potentially pays out of pocket for hospital stay. Receiving this makes getting Medically Complex Waiver quicker and easier.

‡ **Medically Complex Waiver** – Apply for this waiver if your baby qualifies. It is NOT income based. It provides a Medicaid waiver and monetary support:
<https://medicaid.utah.gov/ltc-2/mccw/>

‡ **Medicaid Waiver** – This program IS income based:
<https://medicaid.utah.gov/apply-medicaid/>

- Know when your yearly medicaid review is and be prepared for that (have appropriate forms completed- can contact Integrated Services for help)

‡ **Supplemental Security Income** – Income based for children under 18

‡ **National Parks Pass** – Ask your pediatrician for an application and letter. This pass allows free admission into any national park for the vehicle carrying the person with disability.

‡ **Handicapped Parking Pass** – Received from your pediatrician, or the Medical Care Coordinator at your visit with the Utah PWS Multidisciplinary Medical Clinic

‡ **Create a Special Needs Trust** – Lisa Thornton, our president, is an attorney and can help get you started

Sources of Support to Connect With

‡ **Receive a copy of the booklet “Nutrition Care for Children with PWS, Infants and Toddlers”** – Can be found on PWSA(USA) website and is part of the package of hope.

‡ **Become a member of PWS(USA)**

- Receive all publications helpful for infant stage of development
- Access the parent-to-parent 0-5 list through pwsausa.org and know how to submit questions for the parent board to answer
- Receive a Parent Mentor
- Please visit this website link: <https://www.pwsausa.org/new-diagnosis/>

‡ **Become a member of Utah Prader Willi Syndrome Association**

‡ **Become a member of the Facebook group “UPWSA – Parent’s Only”**

‡ **Become a member of the Facebook group “PWSA | USA Birth to Three”**

If you have questions or get “stuck” at any point in this checklist, please don’t be afraid to reach out. We are here to help!

This list is not to be considered medical advice; consult the appropriate physician for appropriate treatment