



guide for parents

“The experience of being here is like no other hospital we’ve been to. They embrace a parent’s opinion about what they’re doing, which is unique.”

Rob Simpson, whose daughter Cyenna was successfully cured
— *Boston, Mass.*

The Congenital Hyperinsulinism Center at The Children’s Hospital of Philadelphia is dedicated to your child’s safety and well-being. Our goal is to help you take your child home as soon as possible, either cured of hyperinsulinism or on a safe and manageable treatment plan. We are committed to providing family-centered care and consider you an important member of our team. You will be intimately involved in making decisions about your child’s treatment throughout your entire hospital experience and during any follow-up care.

EVALUATION AND TREATMENT

We follow a careful program to evaluate and treat each child in our Center. Some of these steps may be put into motion or completed before you arrive at Children’s Hospital. The order of these steps may vary case by case.

- A diagnosis of hyperinsulinism (HI) is made and confirmed. This is based on your child’s glucose requirement, insulin and ketone levels, and a positive glucagon response (during low blood sugar).
- Medical therapy is attempted using diazoxide and octreotide.
- If your child continues to have low blood sugar with medical therapy, surgery may be needed to remove part of the pancreas.
- Before surgery, your child may undergo additional testing to determine if the HI is focal or diffuse. In focal HI, a localized area of the pancreas has defective beta cells that cause the disease. Since the rest of the pancreas has normal beta cells, the focal lesion can be removed through surgery, which may cure the disease without removing the majority of the pancreas.
- Unfortunately, focal lesions are often too small to be found through regular MRI, CT scan or ultrasound. Based on medical history alone, there is no way to determine if your child has diffuse or focal HI. The Congenital Hyperinsulinism Center is currently performing ¹⁸F-DOPA PET scanning under an FDA-approved research protocol (for appropriate cases). Our staff has the most experience with this preoperative testing procedure in the world. This innovative radiological imaging helps surgeons locate abnormal tissue and determine the exact locations of focal lesions, preventing unnecessary removal of healthy cells in the pancreas and significantly reducing the risks of diabetes in patients with focal HI. This type of precision will assist in pursuing a cure.

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guide for parents' *continued*

- The ^{18}F -DOPA PET scan takes approximately two hours and is performed at the CHOP PET facility under the care and supervision of pediatric anesthesiologists. Your child will receive a small dose of a radioactive isotope, which is picked up by insulin secreting cells in the pancreas. A bright spot on PET imaging indicates an area of focal disease. If this procedure is appropriate for your child, we will discuss it with you in greater detail.
- If your child requires surgery for HI, you should expect to stay at Children's Hospital for at least one month.
- Pancreatectomy surgery generally takes from two to four hours. There is a special waiting room for families of children in surgery where you will receive periodic updates.
- After surgery, your child should be ready to go home in two to three weeks.

Based on your child's age and condition, he will be admitted to either Children's Hospital's endocrine unit or the Harriet and Ronald Lassin Newborn/Infant Intensive Care Unit (N/IICU). Nurses on both units are experienced in dealing with HI. They routinely and expertly perform the follow-up tests your child needs. In addition, any infant who requires surgery will stay in the N/IICU for at least six days after surgery. Children older than 1 year may be treated in the Pediatric Intensive Care Unit (PICU) after surgery.

WHERE TO STAY

At Children's Hospital, parents and caregivers are permitted and encouraged to be with their children at all times. Upon admission, you will receive a wristband that allows you to enter and exit Children's Hospital easily, 24 hours a day. If your child is staying on the endocrine unit, you can sleep in a chair bed located at your child's bedside. If your infant is admitted to the N/IICU, you have access to special sleep rooms at Children's Hospital. If you wish to stay in a room outside the Hospital, choices include the Ronald McDonald House and several hotels in the area. You will need to make these arrangements before coming to the Hospital.



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