



guide for referring physicians

- Congenital hyperinsulinism is a disorder that causes severe and persistent hypoglycemia in neonates and infants.
- Children who develop severe hypoglycemia are at risk for death, brain damage, seizures, mental retardation, blindness or cerebral palsy.
- About half of patients present in the newborn period; the other half present within the first year of life. Only a few cases appear later in life.

WHY REFER TO CHOP?

Congenital hyperinsulinism is rare; most children's hospitals encounter only one or two cases a year. Since October 1998, the Congenital Hyperinsulinism Center at The Children's Hospital of Philadelphia has evaluated and treated more than 400 patients — making it the largest program of its kind in the world. Among the highlights:

- We are dedicated to providing the full spectrum of care, from diagnosis through treatment, including education, psychosocial support and long-term follow-up. Our team is among the best-equipped anywhere to diagnose and treat HI, resulting in shorter hospital stays and more cost-effective treatment.
- Our Center draws on the expertise of specialists from throughout the Hospital, all uniquely suited to caring for the patient and family. We offer support from a wide variety of subspecialties, including a nationally top-ranked neonatal intensive care unit, allowing us to provide seamless care for children with complex conditions.
- Conventional preoperative radiological studies — such as CT scan, MRI or ultrasound — can't differentiate between focal and diffuse HI. Our Center performs ¹⁸F-DOPA PET scanning under an FDA-approved research protocol (for appropriate cases). This innovative imaging technique helps surgeons pinpoint abnormal tissue and focal lesions, sparing healthy cells in the pancreas, significantly reducing the risk of diabetes in patients with focal HI, and potentially leading to a cure. Without ¹⁸F-DOPA imaging, many children at other centers have unnecessary near-total pancreatectomies.

WHEN SHOULD I REFER?

We invite you to contact us at any point in your patient evaluation process to discuss options or to seek advice on patient management. We serve as a resource for physicians and families, providing information to gauge when a child can be diagnosed and managed at their local institution, and when the condition requires a visit to CHOP.

Once a patient fails a trial of diazoxide therapy, it is usually an appropriate time to consider a transfer to Children's Hospital. (See referral chart on reverse for more detail.)

over ►



The Children's Hospital of Philadelphia®

CONGENITAL
HYPERINSULINISM CENTER

referrals ■ appointments ■ information
215.590.7682 ■ hyperinsulinism.chop.edu

WHAT HAPPENS AFTER I REFER?

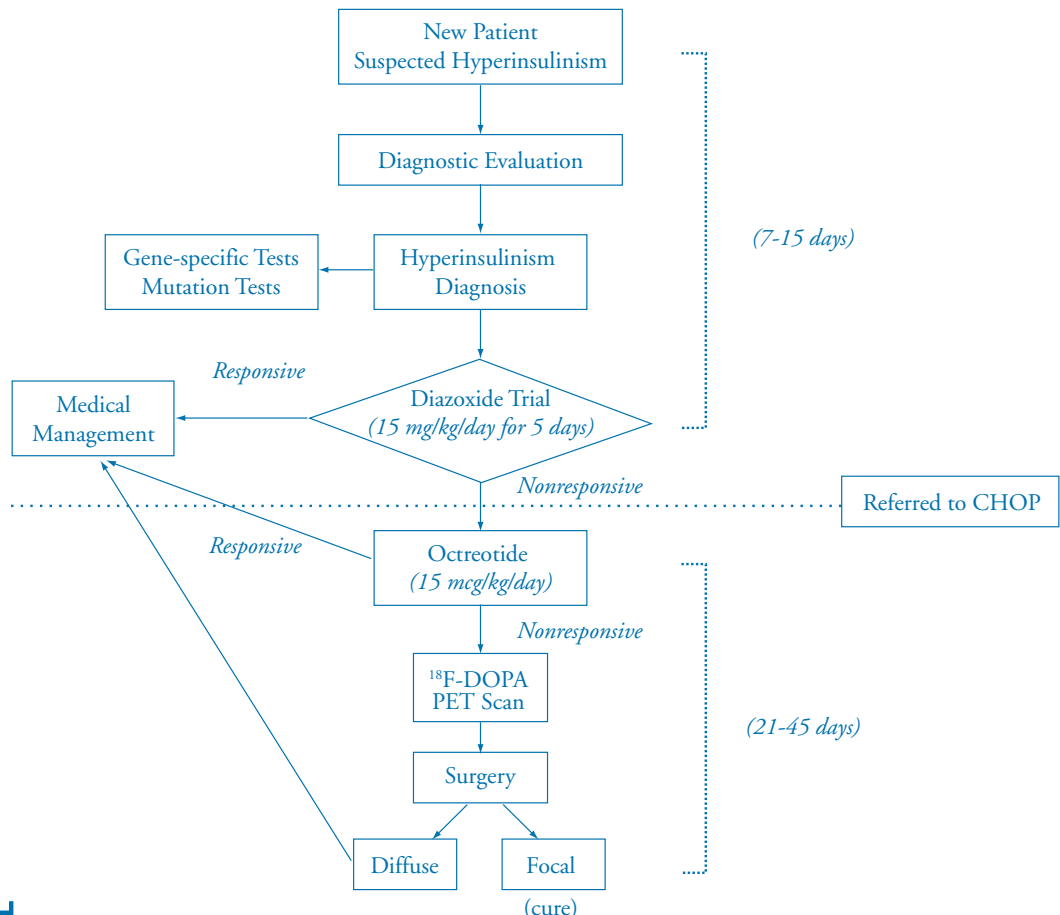
After a child has failed treatment with diazoxide, we usually start octreotide. If the child continues to have low blood sugar with medical therapy, we consider surgery to remove part of the pancreas. First, the child may undergo additional testing to determine if the HI is focal or diffuse, as surgical management depends on the patient's type of hyperinsulinism: diffuse HI requires a 98 percent pancreatectomy, and focal HI requires a resection of the affected part of the pancreas only. ¹⁸F-DOPA imaging allows us to determine the nature of the disease.

Children's Hospital works with referring physicians before transport and maintains close communication and collaboration after the patients return home.

HOW DO I REFER?

- Call the Congenital Hyperinsulinism Center at 215-590-7682, or e-mail us at hyperinsulin@email.chop.edu.
- If the patient may need surgery, it is useful to send a specimen to a specialty genetic reference lab for mutation analysis as early as possible, since results may take more than two weeks.
- Initiate precertification from the insurance company (allow a minimum of 72 hours to complete). We can advise and assist you as needed.

HI Diagnosis and Treatment



The Children's Hospital of Philadelphia®

CONGENITAL
HYPERINSULINISM CENTER

referrals ■ appointments ■ information
215.590.7682 ■ hyperinsulinism.chop.edu