



guide for insurers

- 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.
- Our team is among the best-equipped anywhere to diagnose and treat HI, resulting in shorter hospital stays and more cost-effective treatment.

WHY REFER TO CHOP?

Congenital hyperinsulinism is rare; most children's hospitals encounter 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. 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.

Among the highlights of our Center:

- We are dedicated to providing the full spectrum of care, from diagnosis through treatment, including education, psychosocial support and long-term follow-up.
- Our Center draws on the expertise of specialists from throughout Children's 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.

WHEN TO CALL US

Once a patient fails a trial of diazoxide therapy, it is usually an appropriate time to consider a transfer to Children's Hospital. We also invite providers to contact us at any point in the 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 his local institution, and when the condition requires a visit to CHOP.

WHAT HAPPENS AFTER REFERRAL?

After a child has failed treatment with diazoxide, we 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, while focal HI requires resection of the affected part of the pancreas only.

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Conventional preoperative radiological studies — such as CT scan, MRI or ultrasound — can't differentiate between focal and diffuse HI. Only functional studies of insulin secretion can distinguish between the two forms. ¹⁸F-DOPA PET scanning is the latest, most accurate testing method to determine the nature of the disease. Our Center performs this imaging, for appropriate cases, under an FDA-approved protocol. The scan helps locate abnormal tissue and pinpoint focal lesions, sparing healthy cells, reducing diabetes risk and potentially leading to a cure for the child. Without this technique, children at other centers may undergo unnecessary near-total pancreatectomies.

From before referral and transport through the treatment period, Children's Hospital works with referring physicians and maintains close communication and collaboration after patients return home.

HOW TO 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.
- Physicians must initiate precertification from the insurance company (allow a minimum of 72 hours to complete). We can advise and assist them as needed.



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