Congenital Hyperinsulinism International
Collaborative Research Network

Essential Medical Care, Medication, Supplies and Services for

People with Congenital Hyperinsulinism (HI)

Congenital hyperinsulinism (HI) is the most common cause of persistent hypoglycemia (low blood sugar or low plasma glucose) in infants and children. Low plasma glucose is extremely dangerous, if not treated promptly and appropriately, hypoglycemia will lead to brain damage, developmental delays and even death.

People born with HI require the following from their national and local health care and social systems:

- Attention to family member/caregiver concerns of their newborn
- Continual appropriate management of hypoglycemia from the time of its initial onset
- Timely medical diagnosis of HI through critical sample blood tests
- Upon diagnosis, local physicians should consult a CHI Center of Excellence (COE) and continue communication throughout diagnostic process and when necessary for continuing care. Transfer to COE may be considered.
- Diazoxide must be readily available as long as needed when prescribed by child's doctor.
- Octreotide/Lanreotide must be readily available as long as needed when prescribed by child's doctor.
- Glucagon must be readily available as long as needed when prescribed by child's doctor for emergency~~.~~
- Glucose (Dextrose), feeding pumps, and feeding bags must be available for those individuals who need glucose (dextrose) therapy at home as prescribed by their doctor.
  Gastrostomy tube placement and supplies must be available when prescribed by their doctor.
- Targeted HI genetic testing may be necessary to identify specific mutations that aid in ~~the patient's~~ future treatment decisions.
- Syndromes associated with HI must be identified and managed, e.g., Beckwith-Wiedemann Syndrome.
- An 18 F-DOPA PET scan must be performed on each infant/child suspected of having focal disease (corroborated by medical presentation and genetic testing).
- A surgical team experienced in focal pancreatectomy surgeries along with a specialty trained histopathologist must be available to perform surgery on each infant/child with suspected focal disease.
- A surgical team in subtotal pancreatectomy surgeries must be available to perform surgery on an infant/child with diffuse disease that cannot be treated medically.
- Once discharged from the initial hospitalization, the infant/child must be able to obtain blood glucose testing supplies including test strips and glucometers. Continuous glucose monitoring may also be considered.
- Consultation with HI expert physician about relevant clinical trials when existing therapies are not effective or are suboptimal.
- Child developmental support (evaluation and therapies)
- Psychological support for caregivers and children
- Medical care for potential secondary medical issues resulting from HI such as diabetes, seizures, and pancreatic insufficiency
- Home nursing – Where available and for the most severe cases.
- Appropriate medical day care for infants and Children
- Proper school modifications
- Caregiver leave to attend medical appointments
- Transition services to Adult Care
- Adult care for adults with HI, when necessary