

WHAT IS CONGENITAL HYPERINSULINISM?

Congenital hyperinsulinism (HI) is the most frequent cause of severe, persistent hypoglycemia in newborn babies and children. Prolonged hypoglycemia is the most common cause of preventable irreversible brain damage.

About 60% of babies with HI develop hypoglycemia within the first month of life. Of the remaining, nearly all will develop hypoglycemia before 1 year of life.

HI babies need up to 5 times more sugar (glucose) than normal children.

There are a number of different causes of severe hyperinsulinism. Some forms will resolve and are considered transient. Others arise from genetic defects and can persist throughout life. Brain damage is a risk in both cases.

In most countries, HI has an incidence of 1:25,000-50,000 births.

For those with HI, the pancreas, which is responsible for insulin secretion, is blind to the blood glucose level and makes insulin even when blood sugar levels are not high, causing severe and often prolonged hypoglycemia.

#bemysugar
#stophelows

With prompt diagnosis, early treatment and aggressive prevention of hypoglycemia, brain damage and death can be prevented.

SIGNS & SYMPTOMS OF HYPOGLYCEMIA

Healthcare professionals caring for newborns should be alert to the possibility of **hypoglycemia** when witnessing the following signs and symptoms:

- Bluish-colored or pale skin
- Breathing problems, such as pauses in breathing (apnea), rapid breathing, or a grunting sound
- Irritability often followed by listlessness
- Loose or floppy muscles
- Poor feeding or vomiting
- Problems keeping the body warm
- Tremors, shakiness, sweating, or seizures

Key points for detecting **hypoglycemia**:

- Recurrent hypoglycemia in newborns, infants, and children is not normal. Healthy newborns who experience a blood sugar level drop, do not typically fall below a level of 50mg/dL(2.7mmol/L). Newborns and infants with blood sugars levels lower than 50 (2.7mmol/L) or those whose blood sugar levels drop past the usual transitional newborn nadir should be evaluated.
- At-risk infants suffering from low blood sugar levels must remain in an appropriate clinical setting with blood glucose levels maintained at normal levels until euglycemia is achieved and sustained with a normal age-appropriate feeding schedule. These babies must pass a fasting test or receive a diagnosis and management plan to treat the underlying causes of hypoglycemia before they are transitioned home.
- Newborns who show signs and symptoms of hypoglycemia even if not known to have a risk factor must be evaluated and managed for hypoglycemia to prevent brain damage and death.
- Newborn seizures present very differently from seizures in older babies, children and adults. Medical professionals caring for newborns should be familiar with the presentation of newborn seizures. Blood sugars levels should always be checked with new-onset seizure activity.