

Managing and Treating HI

Safety Fasts and Cure Fasts

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NHS

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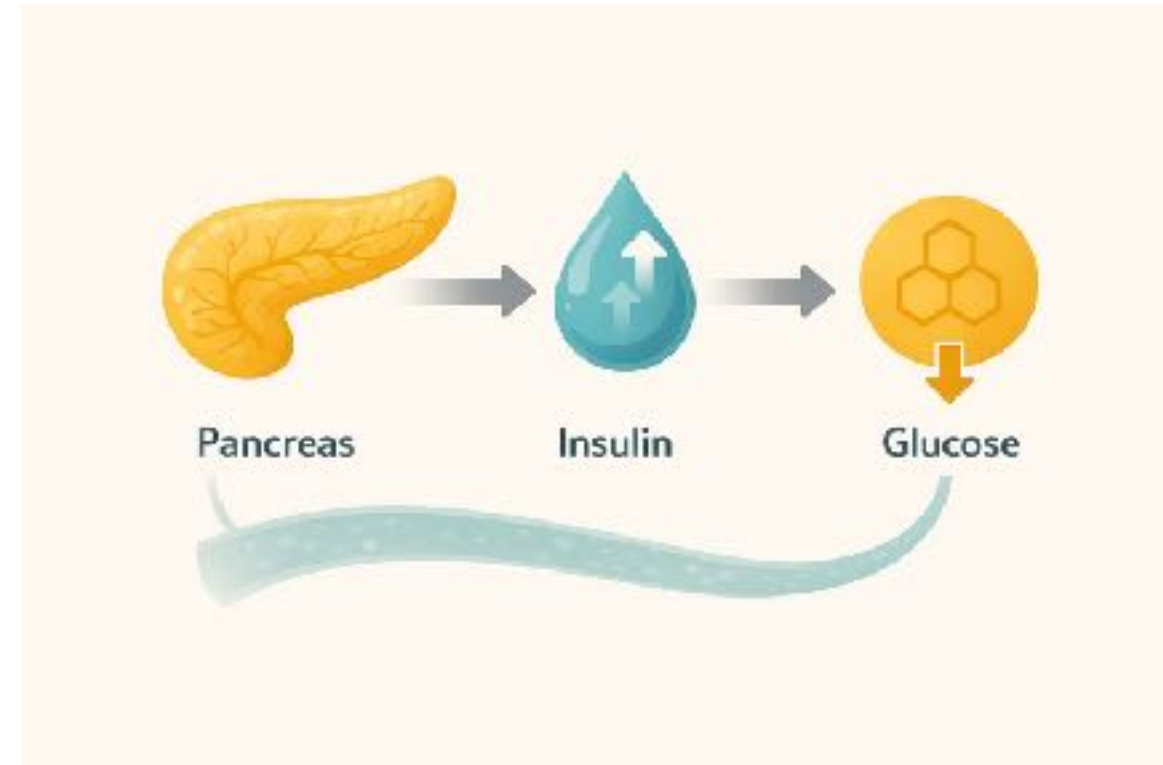
**No conflict of
interest**

DISCLOSURES

I am PI in studies of Zealand, Rezolute, Hanmi

What is HI?

- ✓ Hyperinsulinism (HI) = pancreas makes too much insulin
- ✓ Insulin lowers blood glucose levels
- ✓ Low blood glucose levels can affect the brain
- ✓ Goal of treatment: keep glucose safe so children grow and develop normally
- ✓ HI is the most common cause of persistent low blood glucose in babies



Why Blood Glucose Matters?

- ✓ Babies need steady glucose for healthy brain development
- ✓ Keeping glucose safe is our top priority
- ✓ With the right treatment, we can protect the brain and support normal development



Forms of HI

✓ Transient HI

- Related with perinatal stress

✓ Congenital HI

- Rare disease
- Forms: focal, diffuse, atypical
- 50% caused by a genetic defect
- Some improve over time

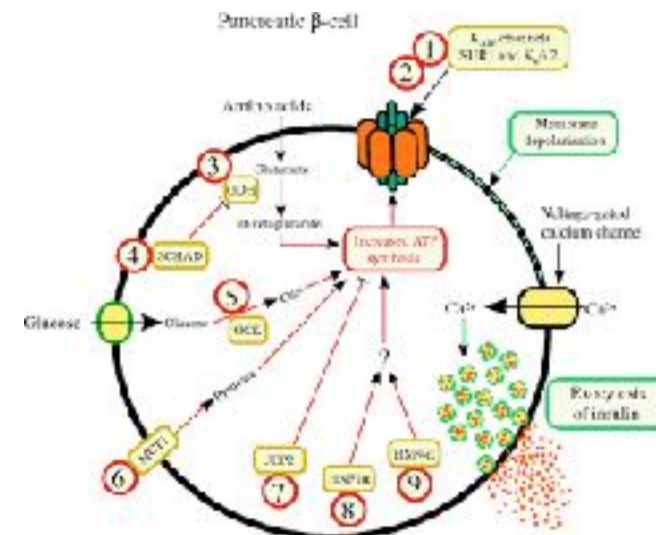
- ✓ Impact: Recurrent hypoglycaemia in neonates & infants causing neurological damage if untreated



Focal Hyperinsulinism



Diffuse Hyperinsulinism



Goals of HI Treatment



Immediate

Keep blood glucose level safe



Mid-term

Support feeding & find the right treatment



Long-term

Protect the brain & support normal development

HI Treatment options



Medical management

Medicines to reduce insulin

Feeding strategies (frequent feeds, overnight feeds, emergency plans)



Surgical management

Most children do not need surgery

Only for specific types of Congenital HI

Sometimes curative, sometimes supportive



Diazoxide

- 1st treatment option
- Reduces insulin release
- Oral medicine, 2–3 times daily
- Some genetic types of HI don't respond



Somatostatin Analogues

- Used when diazoxide doesn't work
- Short-acting injections (Octreotide)
- Long-acting monthly injections (Lanreotide, Sandostatin LAR)



Focal Hyperinsulinism



Cure in 91.4-95%



Diffuse Hyperinsulinism



May help but not cure

Pancreatic Surgery for Congenital HI

- Surgery is considered for certain types of Congenital HI that can be cured or when we are confident it will help

Assessment of response to HI treatment

- Confirms medications working

- Post-surgery for focal form confirms cure
- Monitors disease remission and supports decision when to stop medications

Safety Fast and Cure Fast in HI

- To check if your child can stay safe overnight
- To see whether treatment is working
- To know if HI is improving or cured

- Stable blood glucose
- Rising ketones (a sign the body is coping)

- Helps us decide feeding schedules
- Helps us plan glucose monitoring routine and emergency strategies
- Helps us know when medicines can be reduced and stopped

What is a Fast?

- ✓ A fast is a planned test in hospital
- ✓ It checks if your child can safely go without feeds
- ✓ Usually done before going home or changing treatment

We are checking:

- ✓ Does blood glucose stay safe?
- ✓ Does the body start making ketones (a normal backup fuel)?

It helps us make sure your child is safe between feeds, especially overnight

What Happens During a Fast?



Start

- Normal feed
- Fast begins
- Check glucose + ketone



First few hours

- Child rests
- No interventions



Monitoring Phase

- Blood glucose checked hourly
- Monitor closely
- If blood glucose drops → stop



End of Fast

- Check glucose + ketone
- Take bloods
- Baby is fed straight away

Your child is monitored closely the entire time

What Do the Results Mean for You at Home?

If the fast is successful:

- Glucose stays above 3.5 mmol/L
- Ketones rise well

This means:

- ✓ Your child can go longer between feeds
- ✓ We may reduce feeds or medication
- ✓ You can feel more confident at home

If the fast is not successful:

- Glucose drops
- Ketones don't rise

This means:

- ✓ Your child still needs more support
- ✓ We adjust feeds or medication
- ✓ We keep things safe and controlled

The fast helps us personalize your child's care and keep them safe at home

International Guidelines for the Diagnosis and Management of Hyperinsulinism

Prior to discharge from the hospital, a fasting study to determine the control of HI is suggested for all children, regardless of whether they are on medical therapy or have undergone pancreatic resection. The fasting duration should be predetermined for each patient to ensure that they will be safe in their home environment and to guide the child's sleeping, feeding, and glucose monitoring regimen.

Safety/cure fasting test

1. Have blood drawing IV line in place
2. Check glucose (POC meter) and beta-hydroxybutyrate every 2–3 h until glucose <70 mg/dL; then every 2 h until <60 mg/dL; then hourly until <50 mg/dL. When glucose <60 mg/dL (by POC meter), send specimen for laboratory confirmation of plasma glucose
 - a. Terminate fast when
 1. Plasma BOHB >2 mmol/L on two separate samples 1 h apart
 2. Plasma glucose <50 mg/dL
 3. Duration of fasting >18 h in <1 year old or >36 h in children 1–10 years old or 72 h in >10 years old

Standardised practices in the networked management of congenital hyperinsulinism: a UK national collaborative consensus

Prior to discharge the infant should undergo a safety fast to ensure a reasonable fasting interval at home. The duration of this fast is intended to replicate a reasonable overnight food free interval that fits into parental routines of care. An age-appropriate safety fast should be performed in all patients with a diagnosis of CHI, including those infants managed with additional CHO alone and those with transient CHI.

Assessing Remission in HI

- ✓ **If glucose control is stable on minimal treatment, consider a supervised trial of treatment withdrawal**
- ✓ **Remission criteria:**
 - **Completion of an age-appropriate safety fast**
 - **Suppressed insulin with ketones >1.5 mmol/L at end of fast**
- ✓ **In milder or shorter-duration cases, a consistently normal home glucose profile may be enough without hospital admission or a full fast**

Take home message

A vertical orange line is positioned to the left of four stacked, empty rounded rectangular boxes. These boxes are intended for taking notes or providing a take-home message.

Thank you



- Patients and their families
- NHS Highly Specialized Services Commissioning team
- GOSH CHI Team
- EXETER Genetic Lab
- NIHR Research Facility
- Children's Hyperinsulinism Charity & CHI International



CHI Centers of Excellence (COEs)

