Perioperative Management of Infants with Congenital Hyperinsulinism (HI)

Introduction
Infants with the most common form of congenital hyperinsulinism, KATP hyperinsulinism, frequently require pancreatectomy for intractable hypoglycemia. The purpose of this presentation is to describe the perioperative management for these children at our institution which includes a standardized perioperative process, an electronic order set and a web-based clinical pathway.

Surgical Intervention for HI: Pancreatectomy
Focal vs. Diffuse
Focal HI- Partial pancreatectomy without G-tube placement and less likely to require insulin postoperatively. Pancreatic head resection may require a Roux-en-Y pancreaticojunostomy for adequate pancreatic duct drainage.

Diffuse HI- Near total (95-98%) pancreatectomy and G-tube placement and more likely to require insulin postoperatively.

Preoperative
Patients are transitioned to stock intravenous fluids (IVF) with the same dextrose concentration as HAL without potassium. This is critical to avoid hyperkalemia during pancreatic resection. Insulin is responsible for co-transporting glucose and potassium into cells which can lead to hyperglycemia and hyperkalemia.

Clinical Pathway

Order Set

Pharmacy IV Fluids

- Standard Patients (non-HI)
- 400 hours of age
- No PHP or on full feeds G12 = 2 L/kg
- Full feeds 1WHO = > 2 L/kg
- D10W + 5% Dextrose
- D4.25
- Lasix
- Gastrostomy (POD 0)
- Diet
- 10% Dextrose
- 0.9% NaCl
- ES}
- Bicarbonate 9% NaHCO3
- Heparin (5000 units)
- TPN
- Inotropic Support
- Albumin
- Blood Products
- Pain Management
- Fever
- DVT
- Hypothermia

Order Set

- dextrose 10%
- dextrose 15%
- dextrose 45%
- 0.45 NaCl
- 0.9% NaCl
- 0.05% NaHCO3
- 10% Dextrose
- 0.9% NaCl
- ES
- BWG
- Heparin
- Lasix
- 5000 units
- TPN
- Inotropic Support
- Albumin
- Blood Products
- Pain Management
- Fever
- DVT
- Hypothermia

Intraoperative
IVF is decreased to 1/3 to 1/2 maintenance. Blood sugars are obtained every 30 minutes.

At the end of the case, the maintenance IVF has a glucose infusion rate (GIR) 2:

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\text{GIR (mg/kg/min)} = \frac{\% \text{ Dextrose} \times \text{IV rate} \times 0.167}{\text{Weight in Kg}}
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Postoperative
POD 0
IVF D30W with a GIR 2 infused with 0.45 NaCl to give 80-100 mL/kg/day. Blood sugar checked hourly with goal range 80-180.

Blood sugar > 250 is tolerated for 6-8 hours postop before starting insulin. If blood sugar is > 250 during the 6-8 hour postop period, urine is sent for ketones.

If blood sugar remains > 250, an insulin infusion is started at 0.005 units/kg/hour. GIR is not weaned when insulin infusion is started.

POD 1
D30W infusion is increased to give a GIR 5. TPN is ordered to give a GIR 8.

GIR remains at 8 until feedings are started.

A sepsis evaluation is considered for persistent hyperglycemia or hypoglycemia past the acute postoperative period.

Consider continued disease for persistent hypoglycemia requiring GIR > 8.

Continue to check blood sugar every 1-2 hours. Do not wean GIR if infant requires an insulin infusion.

POD 2
Feeding Regimen
Feedings are restarted once bowel function returns, usually 5-7 days postop.

Infants with extensive duodenal manipulation or Roux-en-Y pancreaticojunostomy may have duodenal edema and are typically NPO for 1-2 weeks or longer.

Day 1- Start 1/3 volume feeds, decrease GIR to 5.
Day 2- Increase to 2/3 volume feeds, decrease GIR to 2.
Day 3- Advance to full volume feeds and discontinue TPN.

Infants tolerating full volume feedings who continue to require an insulin infusion can be changed to subQ NPH.

Surgical Outcomes Data 2004-2011

Objectives
The learner will be able to identify and differentiate between the two main types of congenital hyperinsulinism (HI) requiring surgical management.

The learner will be able to describe the perioperative management of patients with congenital hyperinsulinism (HI), which includes a standardized perioperative process, an electronic order set and a web-based clinical pathway.

The learner will be able to identify the postoperative management of patients with congenital hyperinsulinism (HI).