# When Sugar Takes a Dive: The Curious Case of Idiopathic Ketotic Hypoglycemia

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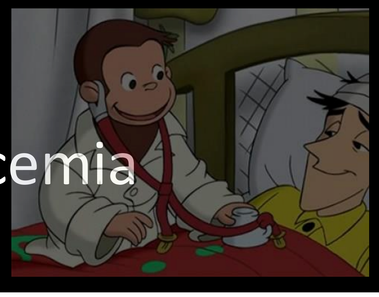

## Chief Complaint

Vomiting x3 days since starting antibiotics for Strep

## HPI

7 year old male with no significant PMH was brought to the ED by his dad due to 3 days of vomiting and diarrhea. He was diagnosed with Strep throat 6 days ago and was started on amoxicillin. Pt with 4-5 episodes of nonbloody emesis in the last 24 hours. Had decreased PO intake in the last 3 days. Dad attempted to give PO Zofran at home but patient was unable to keep it down. Tmax was 100F at home last night. Today patient is less playful and interactive.

## Physical Exam

Vitals: Temperature 98°F, Heart Rate 108 bpm, Respiratory Rate 20 breaths/min, Blood Pressure 96/70 mmHg.
General: Lethargic but arousable male, pale appearing
HEENT: No signs of trauma. Dry mucous membranes.
Abdomen: Soft, non-tender, no hepatosplenomegaly.
CV and Pulm: RRR, CTAB, no wheezes.
Skin: Cool, clammy skin.
Neurologic: Alert to voice, but slow to respond. No focal deficits.
**POC Glucose**: 42

## Pertinent Labwork

- **Initial Labs:**
  - **CBC:** Normal Hb and WBC
  - **CMP:**
    - Glucose: 44 mg/dL
    - pCO2: 14
    - Anion Gap: 26
    - BUN/creatinine: 46.2
- **COVID, flu A/B, RSV**: (-)
- **Peds Recommendation Results:**
  - **Urine**: Glucose (70 mg/dL), Ketones (>150 mg/dL), Protein (50 mg/dL)
  - β-hydroxybutyrate: ↑6.80 mmol/L
  - Ammonia, Lactate, Cortisol, GH, insulin: WNL

## References

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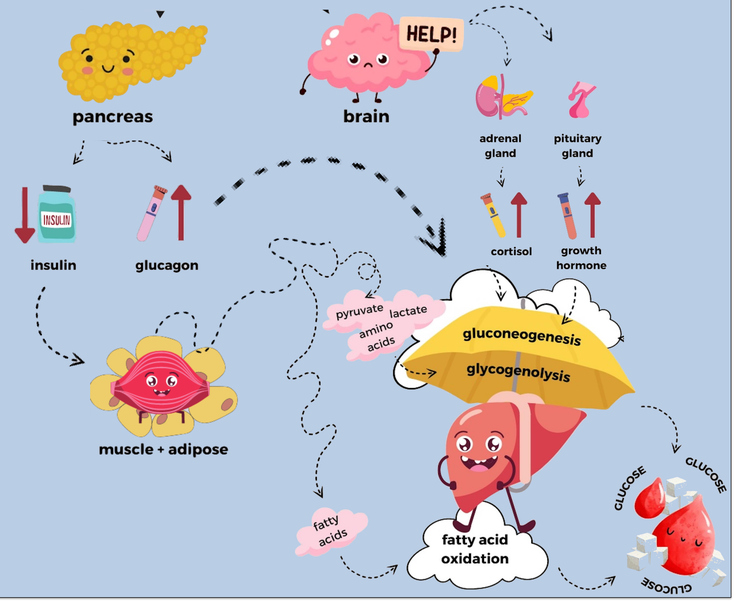
## DDx + Pathophysiology

| Class of Disorder | Principle Examples | Clues to Diagnosis |
|---|---|---|
| **Ketotic Hypoglycemia** | | |
| Idiopathic | Idiopathic Ketotic Hypoglycemia | - Normally 2/2 to illness<br>- Normal exam |
| Counter-regulatory Hormone Deficiencies | - Congenital Adrenal Hyperplasia<br>- Hypopituitarism<br>- Adrenal Insufficiency | - Ambiguous genitalia (CAH)<br>- Hyperpigmentation (CAH/AI)<br>- Small phallus, midline defects (hypopit) |
| Glycogen Storage Diseases | Types I, III, VI, IX, and rarely 0 | - Wakes at night to feed<br>- Hepatomegaly (except GSD 0)<br>- Elevated CKD (GSD III)<br>- Elevated lactate (GSD I)<br>- Elevated cholesterol (GSD III) or triglycerides (GSD I) |
| Gluconeogenic defects | Fructose 1,6-bisphosphatase deficiency | - High anion gap metabolic acidosis<br>- Lactic acidosis |
| Other inborn errors of metabolism | Galactosemia, Hereditary fructose intolerance, organic acidemias | - Liver failure<br>- Multi-system disease |

**DDx also includes:**
- Sepsis
- Hyperinsulinism
- Exogenous insulin use
- Fatty acid oxidation defects


## Case Discussion

- Idiopathic ketotic hypoglycemia (IKH) - Most common in children aged 6 months – 6 years. Often triggered by fasting or minor illness.
- Common **symptoms** - lethargy, confusion, and weakness, often following an illness with reduced food intake.
- Lab findings show hypoglycemia with elevated serum **ketones**.
- The pathophysiology involves **low glycogen stores** with an inability to efficiently produce glucose via gluconeogenesis during fasting.
- In this case, the patient presented with hypoglycemia, ketonuria, and symptoms consistent with IKH, triggered by fasting after a viral illness.
- **Immediate management** included the correction of hypoglycemia with **IVF and D5**.
- IKH is **self-limiting** and typically resolves as children age. However, it is crucial to educate families on prevention to avoid severe hypoglycemic episodes and potential neurological complications.

## Clinical Questions & Answers

**1. What are the differential diagnoses for hypoglycemia in children?**
Endocrine disorders (e.g., adrenal insufficiency, growth hormone deficiency)
Metabolic disorders (e.g., fatty acid oxidation disorders)
Inborn errors of metabolism
Sepsis or severe infections
Drug/toxin exposure
**2. How is idiopathic ketotic hypoglycemia diagnosed?**
Diagnosis of exclusion, supported by *hypoglycemia with elevated ketones*, normal growth, and absence of other metabolic disorders. Evaluate for other causes of ketotic hypoglycemia.
**3. What is the initial treatment of a child presenting with hypoglycemia?**
Immediate administration of D5 + IV fluids of choice. (If possible, obtain recommended labs prior to initiating treatment).

## Clinical Pearls

- **Early Recognition**: Immediately consider hypoglycemia in children with lethargy and recent illness.
- **Early Intervention**: Quickly consult Peds. **If possible**, obtain serum and urine labs (β-hydroxy, lactate, cortisol, GH, insulin, ketones) ***prior to starting treatment*** with dextrose and IV fluids.
- **Long-Term Outcomes**: Families should be educated on preventive measures. Though children typically outgrow the condition by the age of 8-9 years as their metabolic processes mature.