Long Acting Somatostatin Analogues: Early Experience in the Treatment of 5 Patients with Congenital Hyperinsulinism

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Objectives

• To identify standard medical therapy for patients with congenital hyperinsulinism:
  – Diazoxide
  – Octreotide
• To describe the use of long acting analogues (Lanreotide) in the treatment of congenital hyperinsulinism (CHI):
  – Introduction
  – Pharmacokinetics
  – Blood glucoses
  – Improvements
  – Fasting durations
• To describe new evidence based practices and education implication for nurses taking care of patients with congenital hyperinsulinism:
  – Study findings
  – Nursing implications

Congenital Hyperinsulinism

• Affects one in every 30-50,000 infants
• Rare disease
• Different genetic types
• Causes severe hypoglycemia
  – Which can lead to devastating neurologic consequences if not diagnosed and treated properly
• Two main drug therapies used in the treatment of CHI:
  – Diazoxide: first line treatment
  – Octreotide
• Surgery
Current Treatment for CHI

Diazoxide:
- K+ channel opener
- Oral
- Given BID

Octreotide:
- Synthetic analogue of somatostatin
- Believed to act on the Ca+ channel
- Reduces insulin secretion
- Short acting
- Given 2-4 times daily SQ

Long acting somatostatin analogs

Lanreotide
- Similar mechanism of action as Octreotide
- Longer half life
- Administered every 28 days deep SQ
- Single prefilled syringes: 60, 90, and 120 mg
- 20 mm needle
- Deep SQ in the superior external quadrant of the buttock
- Monitor- Growth factors, TFT, gallbladder sludge

Patients

- Five patients who were pre-treated with daily Octreotide injections who attended Cook Children’s Hyperinsulinism Center were enrolled into the study.
  - 4 had diffuse disease and 1 had focal disease
  - 3 had no pancreatectomy
    - (2 diffuse and 1 focal)
  - 2 had a 98% pancreatectomy for diffuse KATP HI
  - 4 were on overnight continuous feeds of D20%
Method

- Blood glucose monitoring
  - 1 month
  - 4 times a day
- Hospital admission
- Fasting study duration
- Started on lanreotide
- Daily Octreotide injections weaned
- Do not wean if blood glucose is less than 70 mg/dL.
- Blood glucose comparison - 1 month
  - Pre-lanreotide
  - Post-lanreotide
- Hospital readmission
  - Fasting study - determine duration and glucose infusion rate (GIR)

Blood Glucose Results

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<th>Blood Glucose Averages</th>
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Averages

Blood Glucose

Octreotide therapy
Somatuline therapy

Fasting Duration Results

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<th>Patients</th>
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Hours
Overnight Infusion Results

Conclusion

- All patients demonstrated less frequent blood glucose in the 50-60 mg/dL and 60-70 mg/dL range
- Majority of the blood glucose ranged > 70 mg/dL
- 5 patients showed a longer fasting duration post lanreotide treatment
- 3 out of 4 patients were able to eliminate their overnight infusion
  - 4th patient decreased the overnight GIR by 65%
- Families reported
  - Increase in freedom
  - Less worry about brain damage
  - Conveniences

Questions

Cook Children’s Hyperinsulinism Center Team
References
