

## 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



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## 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



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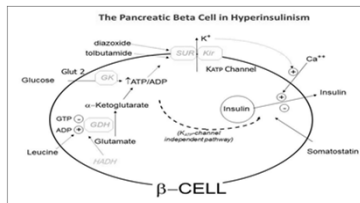
## Current Treatment for CHI

**Diazoxide:**

- K<sup>+</sup> channel opener
- Oral
- Given BID

**Octreotide:**

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



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## 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



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## 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%



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## 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)



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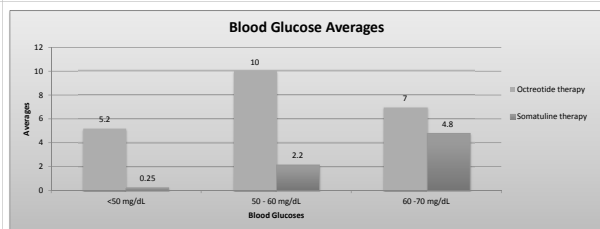
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## Blood Glucose Results



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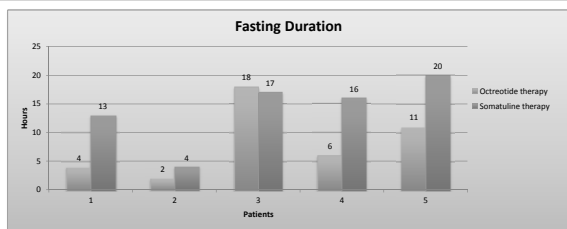
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## Fasting Duration Results



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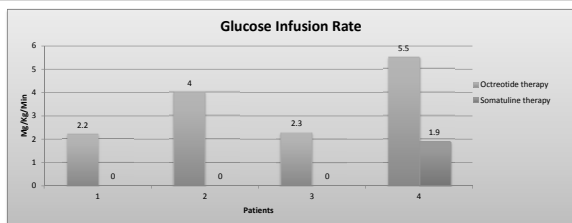
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## Overnight Infusion Results



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## Conclusion

- All patients demonstrated less frequent blood glucoses 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
  - 4<sup>th</sup> patient decreased the overnight GIR by 65%
- Families reported
  - Increase in freedom
  - Less worry about brain damage
  - Conveniences



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## Questions

Cook Children's Hyperinsulinism Center Team



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## References

- Palladion, Andrew., Stanley, Charles., (2011) A specialized team approach to diagnosis and medical versus surgical treatment of infants with congenital Hyperinsulinism. *Seminars in Pediatric Surgery* . 20, 32-37.
- Petersen, Holger., Bizec, Jean-Claude., Schuetz, Helmut., and Deloporte M Marie-Laure. (2011) Pharmacokinetic and technical comparison of Sandostatin LAR and other Formulations of long acting octreotide. "BMC research Notes" 4:344. <http://www.biomedcentral.com/1756-0500-4-344>

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