Congenital Hyperinsulinism in a resource limited setting: Overcoming barriers towards a survival path

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INTRODUCTION

Congenital hyperinsulinism is genetic disorder characterized by dysregulation of insulin secretion

It is the most common cause of persistent hypoglycaemia in infancy

The incidence in individuals of northern European extraction is approximately 1:30,000 live births.

Published data on the diagnosis and management of congenital hyperinsulinsim in resource limited setting is scarce.

OBJECTIVES

To describe the clinical presentation, diagnosis and management of congenital hyperinsulinism in resource limited setting

METHODS

We report on an 18 months old boy who presented in the new-born period with history of poor suck and recurrent hypoglycaemic seizures.

He was born by spontaneous vaginal delivery weighing 2.6 kg. The full septic work for possible infection was unremarkable.

He was managed with recurrent oral feeds and intravenous dextrose infusion.

To maintain euglycaemia, he required intravenous dextrose solution at 10 mg/kg/min.

Critical blood samples taken during hypoglycaemic episode.

RESULTS

The results of the critical sample taken during a hypoglycaemic episode revealed;

- Blood Glucose 0.02 mmol/l (Range-3.5-7.0 mmol/l)
- Growth Hormone 22.2 mIU/l (Range-0.2-13 mIU/l)
- Insulin 22.8 mIU/l
- C-Peptide 1086 pmol/l (Range-364-1655)
- Cortical 6147 nmol/l(Range-55-304 nmol/l)
- Total Cholesterol 3.3mmol/l
- Triacylglycerides 0.78mmol/l
- High Density Lipoproteins 1.62mmol/l
- Low Density Lipoproteins 0.6 mmol/l
- No urine ketones.

In view of the inappropriately elevated insulin levels in the presence of very low blood glucose, the diagnosis of congenital hyperinsulinism was made.

He was commenced on oral diazoxide and frequent oral feeds with polycose.

He continued to develop recurrent hypoglycaemic episodes despite the treatment.

Following 3 weeks of oral diazoxide therapy with no response, he eventually underwent partial pancreatectomy.

The hypoglycaemic episodes resolved following that and he was discharged home.

CONCLUSIONS

The diagnosis and management of congenital hyperinsulinism is feasible in resource limited setting.

Congenital hyperinsulinism should be considered in the differential diagnosis of children presenting with recurrent hypoglycaemia at any age

References

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Diabetes

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