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hyperinsulinemic hypoglycaemia

Dr Veeranna Kotrashetti,Dr Vijay Sonawane,Dr Kapil Bainade,Dr Abir Lal Nath
Dr Abir Lal Nath,Dr Dy patil university,Dy patil medical college,nerul,sector 5,navi Mumbai:Country India

Abstract: *A 2 month male child born to 3RD degree consangious parents presented for convulsions 2 hour before admission.Preliminary investigations revealed low blood glucose.Further Investigations revealed hyperinsulinemichypoglycemia,Child was managed with intravenous glucose. Persistent hyperinsulinemic hypoglycemia of infancy (PHHI) represents the most common cause of hyperinsulinism in neonates.Severe recurrent hypoglycemia associated with an inappropriate elevation of serum insulin, C-peptide, and proinsulin levels defines congenital hyperinsulinism.If left untreated,congenitalhyperinsulinismcan lead to mortality and morbidity.*

Keywords: *PersistentHyperinsulinemic hypoglycemia(PHHI),Neonatal hypoglycaemia*

I. Introduction

PPHI is the most common cause of persistent hypoglycemia in infants and children, which if unrecognized may lead to development delays and permanent neurologic damage**.** In general, the high risk of brain damage appears to be due to delays in diagnosis and treatment rather than a consequence of the genetic defects and, thus, is potentially preventable[1].Most patients with PPHI present shortly after birth with symptoms of hypoglycemia (eg, hunger, jitteriness, lethargy, apnea, seizures).The presenceofjitteriness,lethargy and seizures along with decreased serum glucose, normal urinary ketones and with elevated insulin levels (>10 µU/mL) and normal levels of free fatty acid (FFA) suggested the possibility of hyperinsulinemic hypoglycemia.Among the nine known genetic causes of PPHI, mutations in the genes encoding the ATP-sensitive potassium channel represent the most common defect accounting for the majority of the cases [2].We describe a 2 month old male child who came with complain of convulsion,Investigations revealeddecreased serum glucose, normal urinary ketones and with elevated insulin levels (>10 µU/mL) and normal levels of free fatty acid.

II. Case Report

A 2 month old male child bought with convulsion.Routine investigations revealed hypoglycaemia.Physical examination revealed jitteriness,lethargy,apnea. Baby had similar episode in day 3 of life shifted to nicu for hypoglycaemia,managed by iv dextrose.At present baby was stabilized with iv dextrose,serum glucose and ketone levels were low.Glucagon(100micro gm/kg) was added to maintain Euglycaemia..Serum insulin was high,cortisol,free fatty acid were low and ammonia was high.Patient responded to diazoxide.MRI spectroscopy revealed hypoglycaemic insult.

III. Discussion

PPHI of infancy is a heterogeneous disorder of unregulated excessive secretion of insulin with variable age at onset from birth to childhood.Characterised by non-ketotic non-acidemic severe hypoglycemia with depressed fatty acid level consequent to its inhibitory action on counter regulatory hormones.Incidence of PHHI is high in areas with consanguinity.Aim of treatment is to maintain normoglycemia and prevent hypoglycemia related brain damage. Three treatment modalities were considered to maintain normoglycemia, namely opposing the insulin action via glucagon, hydrocortisone or preventing the secretion of insulin from beta-cells by diazoxide or octreotide and nifedipine or reducing the beta-cell mass by resection in failure of above two measures. Diazoxide is the main treatment of choice for PHHI(3). It prevents insulin secretion by acting on the pancreatic beta-cell KATP channel and has the effect of opening the channel.One third of these infants may still remain hypoglycemic requiring total pancreatic resection.The potential for preventing permanent brain damage caused by persistent hypoglycemia, makes it extremely important to identify and treat these children early.

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