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Persistant Hyperinsulinemic Hypoglycaemia Of Infancy And Neonates : Surgical Management And Intermediate Term Outcomes

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ABSTRACT

Persistent hyperinsulinemic hypoglycaemia of infancy, also called nesidioblastosis, or congenital hyperinsulinism, is a disease characterized by hyperinsulinemia even with hypoglycaemia, which may result in brain dysfunction as the brain thrives on glucose exclusively. The other causes of this hypoglycaemia are ruled out before aggressive medical treatment. Finally, surgical treatment is advised, where the use of early surgery, which is major, involving near complete removal of the pancreas, is undertaken. Here, we try to address the management with these four cases.

1. INTRODUCTION

: Nesidioblastosis is a rare but important cause of hypoglycaemia in neonates and infants, which is now called persistent hyperinsulinemic hypoglycaemia of infancy(PHHI), which occurs due to inappropriate high secretion of insulin by the pancreas, leading to severe hypoglycaemia, irreversible effects on the brain leading to irreparable neurological dysfunction, and even mortality. Here we present our experience with four such cases and their aggressive surgical treatment.

2. AIMS:

To study the neonates with persistent hyperinsulinemic hypoglycaemia of infancy[PHHI] with respect to their surgical management and the outcome

3. METHODS:

A study of neonates was conducted from January 2009 to December 2018, with inclusion criteria of unresponsiveness to IV dextrose & drugs at the highest possible doses at a tertiary care centre with a NICU. Three neonates and an infant who presented with episodes of seizures with hypoglycaemia were studied. Transient causes and other common causes of hypoglycaemia were excluded. These neonates and infants underwent complete medical treatment and then were subjected to surgery after no response and persistent hypoglycaemia.

4. RESULTS:

The three newborns and an infant diagnosed with persistent hypoglycaemia had increased insulin levels, normal cortisol, ammonia & ketone levels. Insulin levels were increased in all babies. Treatment initially was conservative, medical line which included IV dextrose >15mg/kg/min continuous infusion, hydrocortisone, hydrochlorthiazide, oral diazoxide started with 10 mg/kg/day, increased to 20mg/kg/day, and even octreotide was started, which did not affect hypoglycaemia. USG & CECT scans were done to look for any lesions in the pancreas. After no response to medicines, near total pancreatectomy was done in 4 pts with the dissection starting from the tail, body, till a part of the head of the pancreas was resected, with preservation of spleen & splenic vessels.(Fig 1,2)

Images

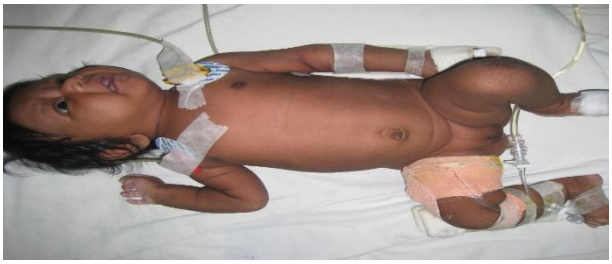


FIG 1. BABY WITH HYPOGLCAEMIA.

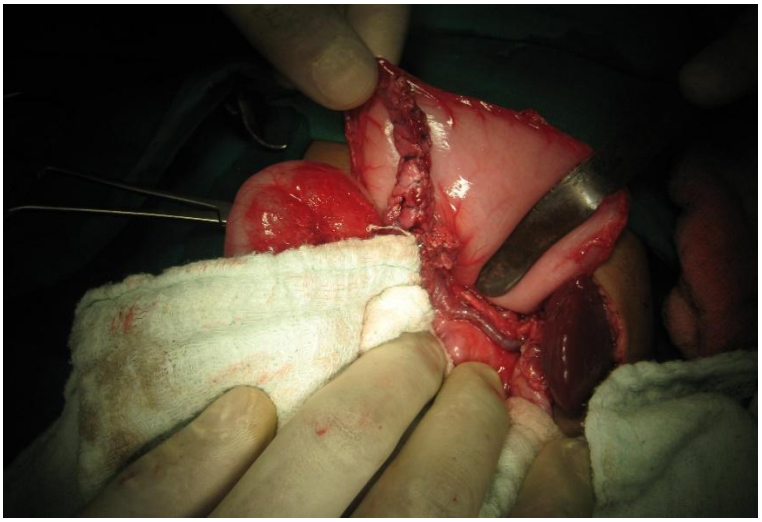
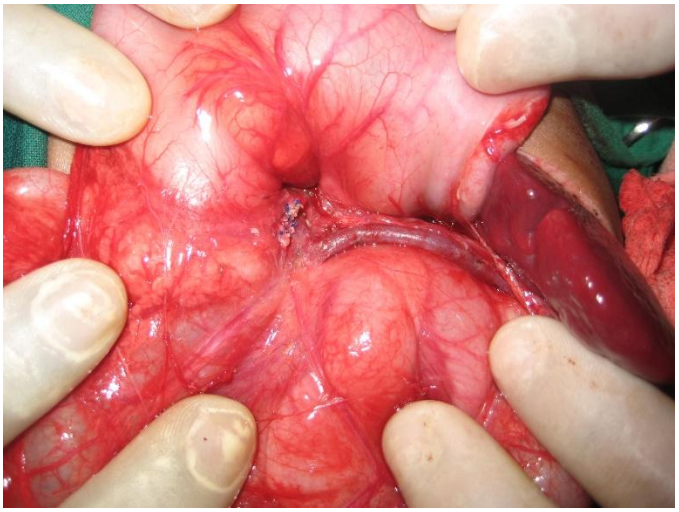


FIG 2. DISSECTION OF PANCREAS



.FIG 3. AFTER PANCREATECTOMY

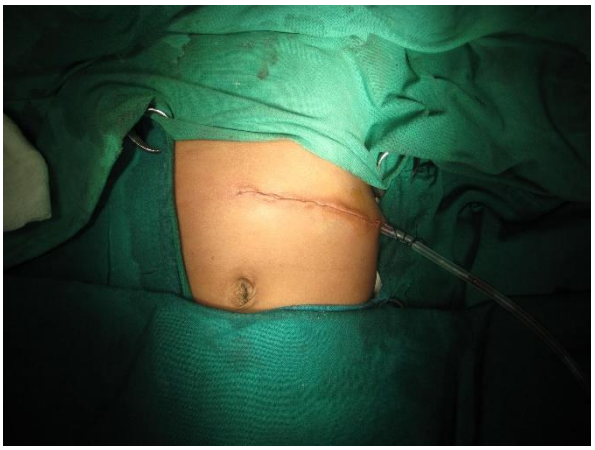


FIG 4. POSTOP IMAGE

Postoperatively, blood sugars became normal, and drugs were tapered off completely. One patient had a minor visual acuity problem, which was resolved with conservative treatment by an ophthalmologist. One patient required prolonged diazoxide, and another one required redo surgery for the removal of residual pancreas for persistent hypoglycaemia with demonstrably high insulin levels. One patient was lost to follow-up. Histological examination revealed

diffuse hyperplasia of pancreatic cells. Two babies are doing well clinically and neurologically at follow-up, with a minimum of 2 years of follow-up.

5. DISCUSSION:

Nesidioblastosis, the term coined by Laidlaw in 1938, also called PHHI, is a very rare condition where there is hypersecretion of insulin irrespective of glucose levels in the blood. The incidence of hypoglycaemia is 4 in 1000 term neonates, and that of PHHI is 1 in 50,000 live births. The manifestations vary by age and severity of the hypoglycemia.1 Hypoglycemia in early infancy can cause jitteriness, lethargy, cyanosis & seizures.1,2Severe forms may cause in-utero macrosomia. Children who have prolonged or recurrent hyperinsulinemic hypoglycemia in infancy can have intellectual retardation, developmental delay, and even death.2,3 The different types of hypoglycaemia include transient neonatal hyperinsulinism, focal hyperinsulinism, which has either a paternal SUR1 mutation or a paternal Kir6.2 mutation, and the other type: diffuse hyperinsulinism, which has either an autosomal recessive form.4 Investigations include detectable amounts of insulin during hypoglycemia, inappropriately low levels of FFA, ketone bodies, and a high requirement for

IV glucose to maintain adequate blood glucose levels. And a GIR above 10 mg/kg/minute in infancy suggests hyperinsulinism.[2]USG, CT & MRI have been used to search for a focal mass in the pancreas, but have low yield. The PET scan uses a novel isotope, [18-F]-L-DOPA, for which neuroendocrine cells have a high affinity, is better and is 96% accurate for diagnosing focal or diffuse disease.[5] The medical line treatment starts shorter feeding interval; every 2 hours, increased calorie density of formula, continuous feeding through an NG tube, continuous intravenous dextrose, hydrocortisone, diazoxide, octreotide by subcutaneous injection or infusion, glucagon by continuous i.v infusion, and calcium channel blockers. Pancreatectomy is usually the treatment of last resort and includes removal of about 95% of the pancreas, but the decision of early surgery is proven to be beneficial in many of these babies.[5,6] Some do not recover which is as high as 50%, and need second procedures to remove even the last remnants. Hypoglycaemia may resolve months or years later. The development of diabetes mellitus is in 14%, with very few reports have shown an incidence of 30 %, the mean time of development of which is around 9.6 years.[4,6]

6. CONCLUSIONS:

Early onset forms of PHHI are usually resistant to medical therapy, and the neurological sequelae are related to delay in the diagnosis and surgery. Hence lies the importance of early diagnosis and aggressive treatment with surgery with near total pancreatectomy, which is sometimes even less, for hypoglycemia secondary to hyperinsulinemia of infancy is life-saving procedure and is better than manageable diabetes mellitus. However long-term studies are needed in our subset of population to prove the same

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