## Persistant Hypoglycemia in Children: Hyperinsulinemia

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## **Abstract**

Congenital hyperinsulinemia occurs due to inappropriate insulin secretion from beta cells of the pancreas. It is the most common cause of resistant and recurrent hypoglycemia in neonates and infants and the threatment is very difficult. Although the incidence is reported as 1/50000, it is seen more frequently in consanguineous countries. Patients may present with lethargy, nourishment, irritability and seizures. If it is not treated, severe hypoglycemia may result in severe neurological damage. High glucose requirement in the diagnosis of congenital hyperinsulinism, detectable insulin level and ketone negativity are the key findings during hypoglisemia. Approximately 45-55% of the patients have an underlying genetic etiology. The most common mutations are ABCC8, KCNJ11, GLUD1 HADH, GCK, SLC16A1 hepatocyte nuclear factor 4 alpha and 1 alpha. In the treatment of drugs such as diazoxide, octreotide, nifedipine may be used, while some patients may require surgical treatment.

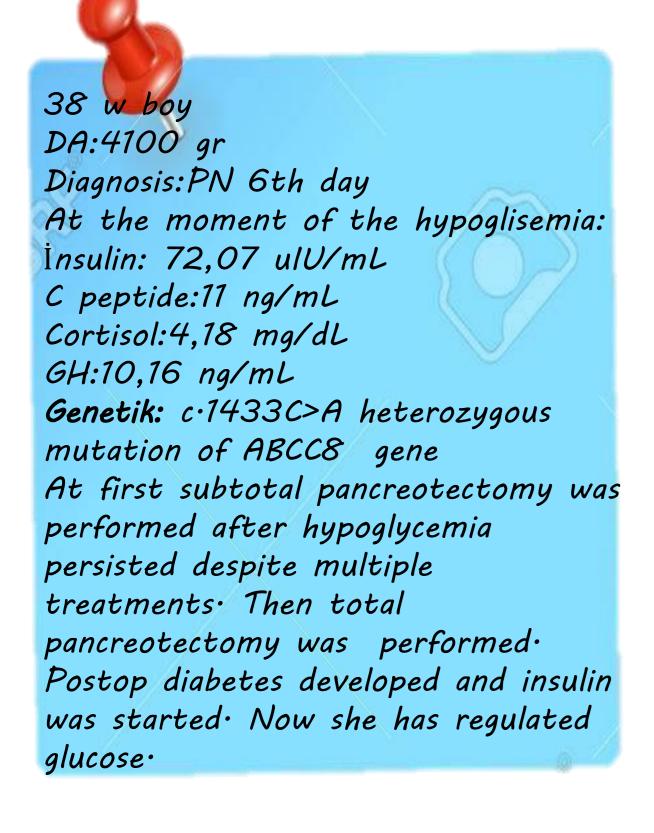
Here, the aim of this study was to evaluate the clinical, genetic and therapeutic responses of hyperinsulinemia patients.

37 w girl BW:4100 gr Diagnosis: first 1 week At the moment of the hypoglisemia: İnsulin:59,6ulU/mL C peptide: 9,93 ng/mL Cortisol:18,6 Genetic: c.2113C>T/c.4017G>A heterozygous mutation of ABCC8 gene Subtotal and total pancreotectomy was performed after hypoglycemia persisted despite multiple treatments. Postop diabetes developed and insulin was started. Now she has regulated glucose.

38 w boy BW:3750 gr Diagnosis: first 1 week At the moment of the hypoglisemia: Insulin: 26,65 uIU/mL C peptide:4,87 ng/mL Cortisol:42,90 mg/dL GH:9,59 ng/mL Genetic: c.2113C>T/c.4017G>A heterozygous mutation of ABCC8 gene Subtotal and total pancreotectomy was performed after hypoglycemia persisted despite multiple treatments. Postop diabetes developed and insulin was started. Now he has regulated glucose.

38 w kiz BW:4490 gr Diagnosis: İlk 1 hafta At the moment of the hypoglisemia: İnsulin: 17,13 ulU/mL C peptide: 9,54 ng/mL Cortisol:63,44 mg/dL GH:9,22 ng/mL Genetic: c.2113C>T/c.4017G>A heterozygous mutation of ABCC8 gene Diazoxide and octreotide treatments were started, but the patient died due to severe sepsis induced intestinal perforation

38 w boy
BW:4500 gr
Diagnosis:PN 5th day convulsiyon
At the moment of the hypoglisemia:
İnslulin: 8,34 uIU/mL
C peptide: ng/mL
Cortisol:5,1 mg/dL
GH:18,9 ng/mL
Genetic Chr11:g·17426198C>A
homozygous mutation of ABCC8 gene
Chr4:g·108911107C>T heterozygous
mutation of HADH mutasyon
Diazoxide and octreotide treatments
was started, now he has regulated
glucose·



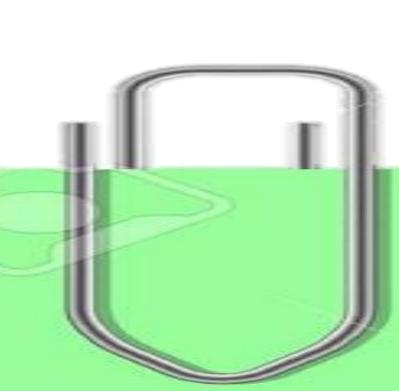
40 w boy
DA:2160 gr, 1' cousin marriage
Diagnosis:1,5 month
At the moment of the
hypoglisemia:
İnsulin: 9,9 uIU/mL
C peptide:3,93 ng/mL
Cortisol:17,4 mg/dL
GH:12 ng/mL
Genetik:There were no
mutations of ABCC8, GCK,
KCNJ11 genes
Regulate blood sugars with
diazoxide treatment.

34 w girl DA:2575 gr Diagnosis: PN 9 th day At the moment of the hypoglisemia: İnsulin: 25,92 ulU/mL C peptide:6,11 ng/mL Cortisol:1,49 mg/dL GH:18,1 ng/mL Genetic: c·148 C>T p.Arg50Trp heterozygous mutation of KCNJ11 gene Octreotide was started in the patient who developed pulmonary hypertension with diazoxide. Now she has regulated glucose.

Premature birth? boy
BW:?, he has otism
Diagnosis:8 year 5 month
At the moment of the
hypoglisemia:
İnsulin: 19,93 uIU/mL
C peptide:3,31 ng/mL
Cortisol:10,74 mg/dL
GH:0,47 ng/mL
Genetic: c·271 G>C p·V91L
mutation of GCK gene
Hypoglycemia persisted despite
multiple treatments·The patient
was suggested pancreotectomy
but the family did not accept.







## **Result:**

Hyperinsulinemia is more common in countries such as our country where the rate of consanguineous marriage is high. Early detection and treatment of hypoglycemia is very important for the prevention of neurological sequelae. Here, we wanted to draw attention to this group of diseases that are quite difficult to manage.





