BACKGROUND

- Congenital Hyperinsulinism (CHI) is a common cause of recurrent and persistent hypoglycemia in the neonatal period. (1)
- Diazoxide is an FDA approved drug used in treating CHI. (2)
- It activates sulfonylurea receptor of ATP-sensitive potassium (KATP) channel on β-cell of pancreas leading to suppression of insulin secretion. It is widely used in management of CHI. (3, 4)

RESULTS

- Among 90 children (males= 69.4%) 72 were diagnosed in neonatal period.
- Only in 18.1% children diazoxide therapy was required for >2 yrs.
- On comparison of those required treatment of <2yrs & >2yrs it was noticed that c-peptide level were significantly high in children requiring longer duration therapy.
- Among those requiring therapy for > 2yrs, 23.1% were genetically positive as compared to 5.1% among those needing treatment <2 years.

AIM

- To report the clinical, laboratory, treatment and outcome profile of diazoxide responsive CHI from a tertiary pediatric endocrine unit

METHOD

- A retrospective study at a tertiary pediatric endocrine unit (from January 2012 to Nov 2019).
- Inclusion criteria- children diagnosed with CHI during the episode of hypoglycemia, elevated c-peptide level inappropriately suppressed ketones and free fatty acids who were responsive to diazoxide.
- Children switched to octreotide or lost to follow-up or transferred to other centers were excluded.
- Out of 103 children who were diazoxide responsive 13 were excluded from the study and data of 90 children was included and analysed.
- Detailed history, clinical findings, biochemical parameters, 2-D echocardiography, adverse events following diazoxide therapy, genetic analysis & long-term follow-up data were noted.
- All data were entered in MS excel 2016.
- Analysis was performed with SPSS v 21.
- P <0.05 was considered significant.

CONCLUSIONS

- We have reported children with diazoxide responsive CHI where the median duration of therapy is shorter in neonatal onset CHI compared to those with late onset.
- Also most of the neonatal onset CHI had risk factors present and diazoxide was eventually discontinued in majority of them.
- Genetics were negative in majority of the diazoxide responsive patients who had transient CHI.

REFERENCES


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Table 1. Profile of diazoxide responsive neonatal CHI (continuing)

Table 2. Profile of diazoxide responsive neonatal CHI (categorical variable)

Table 4. Positive Genetic mutation description of the study population