TWO PERMANENT NEONATAL DIABETES MELLITUS CASES DUE TO MUTATION IN ABCC8 GENES IN VIET NAM: CLINICAL FEATURES AND LONG – TERM OUTCOME IN TREATING BY SULFONYLUREA (2008-2014)

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Background: Neonatal diabetes mellitus (NDM) is a rare insulin-requiring form of diabetes, diagnosed in the first six months of life. Unlike type 1 diabetes mellitus, it is caused by the mutation genes involved in the development and secretory function of the pancreas. ABCC8 gene mutation, resulting in both transient and permanent NDM, increases the sensitivity to the stimulatory actions of ADP, so it remain the potassium channel open and prevent insulin release. Sulfonylurea acts by an ATP-independent mechanism to close these channels and insulin release. It is therefore the best choice for treating permanent neonatal diabetes mellitus due to ABCC8 and KCNJ11 gene mutation.
Objective and hypotheses: We report two cases of NDM due to ABCC8 gene mutation about clinical features and good long-term outcome in management by Sulfonylurea.
Method: We collect data base on the interview their parents and the medical record about diagnosed-age, signs and symptoms, family history and laboratory test (Glucose, HbA1c, Anti-GAD, ICA, C-peptide, Insulin), Insulin regimen (dose, type, duration), Sulfonyurea (dose), side effects and complication at the beginning and after 3 months.
Results: Two girls, diagnosed diabetes mellitus before 6 months of age, one was incidentally discovered amd one was hospitalized in ceton acidosis. They were healthy before and there was no family history. The blood glucose and HbA1c were very high, C-peptide were 0,282 and 0,278 ng/ml respectively, ICA and Anti-GAD were negative.They were treated with subcutaneous injection Insulin but the blood glucose is in wide variance, and there were several hypoglycemic times. After gene analysis, with ABCC8 gene mutation, they have been transfered to Sulfonylurea and we rapidly stopped using Insulin after 2 weeks. The treament is more simplex, the blood glucose is stable in the long term, HbA1c dropped from 8,3% and 6,8% before Sulfonylurea to 5,8% and 6% respectively and there are no side effects up to now.
Conclusion: We should do gene analyze in all diabetes children diagnosed before 6 months of age. The changing management from Insulin to Sulfonylurea is much effective and safe in ABBC8 and KCNJ11 mutation NDM.

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