

Successful Treatment of Diabetic Ketoacidosis and Hyperglycemic Hyperosmolar Status in an Infant with *KCNJ11*-Related Neonatal Diabetes Mellitus via Continuous Renal Replacement Therapy

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Introduction

- Neonatal Diabetes Mellitus (NDM) is a rare monogenic disorder presenting as uncontrolled hyperglycemia during the first 6 months of life and which may be either permanent or transient.
- HHS (Hyperglycemic Hyperosmolar State) is a rare condition in NDM patients, only observed previously in 6q24-related transient NDM.
- *KCNJ11* gene mutation caused NDM patients hasn't been observed with HHS in previous researches.

Case Report

- A Chinese girl, 2 months old, 4.6Kg.
- Presented with dyspnea and lethargy
- Blood gas showed severe hyperosmotic Diabetic Ketoacidosis (DKA), the DKA and HHS exacerbated during the treatment
- The Gene analysis revealed a de novo mutation (c.602G>A(p.R201H)) of *KCNJ11* gene

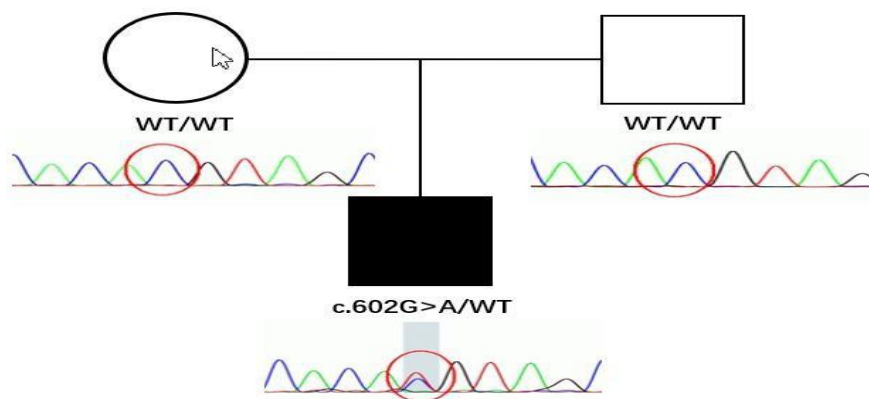


Figure 1: The family pedigree. The mutation status of *KCNJ11* c.602 G>A is indicated by the symbol for each subject. WT indicates wild type. Sanger sequencing of three members of the family in c.602 position is also indicated under the symbol of each subject. The red circles indicate the mutation points.

Case Report

- Routine fluid and insulin therapy was recommended in the guideline.
- CRRT (Continuous Renal Replacement Therapy) was used after the routine therapy failed
- DKA and HHS was totally corrected after 18 h treatment with CVVHDF (Continuous Venovenous Hemodiafiltration).

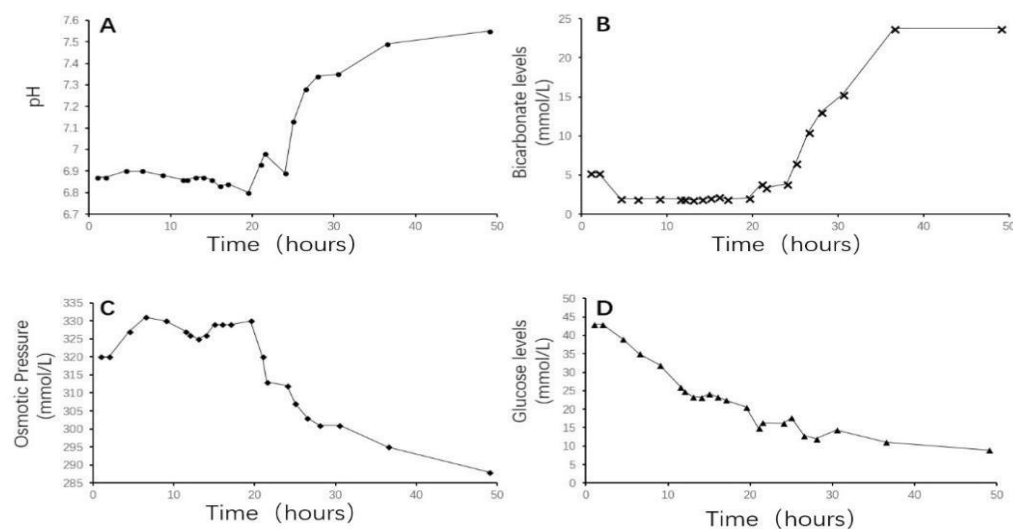


Figure 2: Blood gas analysis of the patient during the course of treatment. The pH value, bicarbonate level, osmotic pressure, and glucose levels over the whole course of treatment are shown in A, B, C, and D, respectively. The red vertical lines indicate the start of CVVHDF.

Conclusions

- CRRT has significant potential in saving critically ill children with severe DKA and/or HHS.
- Further studies are still required to assess the value of CRRT in more cases.

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Disclosures

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Compliance with Ethics Guidelines

The authors received informed consent from the patient's parents for this case report.

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