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Introduction

Neonatal Diabetes Mellitus (NDM) is a rare form of diabetes

- Onset within the first 6 months of age
- Incidence of 1/90.000 live births
- Sub-classified in transient (50%) and permanent neonatal diabetes mellitus (PNDM)

Case History

We present the case of a female patient with positive family history of **PNDM**, admitted to the Neonatal Unit. Father's diabetes was due to activating mutation of **potassium channels** (KCNJ11-V59M mutation) and ,in view of the genetic risk, a glucose screening was performed.

In the first week of life, sporadic hyperglycemic events were spotted and **continuous glucose monitoring (CGM)** was started.

- On day 11, because of persistent hyperglycemia (233-239 mg/dl) we started **glibenclamide** at the dosage of 0,33 mg/kg/day, which was doubled during the following days up to 0,65 mg/kg/day.
- On day 13, considering the poor glycemic control and the weight loss, IV **insulin** was added (0,6 IU/kg/day), achieving a better control and weight gain.
- On day 17 molecular testing confirmed the same mutation of her father, so we attempted a **switching from insulin to glibenclamide**, doubling the dose of sulfonylurea and halving the insulin dose according to the CGM profile.

Insulin was successfully **stopped** in 4 days and the patient **was discharged on glibenclamide** (0,75 mg/kg/day administrated every 8 hours).



Conclusions

Approximately 90% of patients with activating mutations in KCNJ11 can be switched from insulin to sulfonylurea.

In this case, starting glibenclamide in the first weeks of life didn't allow to reach a satisfying glycemic control and gaining weight, though we did not reach during the first attempt the weaning dose of 0.75 mg7kg/day. For those reasons, should insulin therapy always be considered as first approach?

The debate is still open...

References

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 Minimal incidence of neonatal/infancy onset diabetes in Italy is 1:90,000 live births.