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Treatment of congenital nephrogenic diabetes insipidus in pregnancy

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Congenital nephrogenic diabetes insipidus (NDI) is a hereditary renal disorder characterized by the failure to concentrate urine in response to antidiuretic hormone (ADH). No data was found concerning the influence of NDI on the chances of a successful pregnancy. We present the case of a woman patient diagnosed with NDI in childhood and report on the influence of NDI on the course of two of her pregnancies and the treatment of her fluid and electrolyte disturbances during these pregnancies. On the basis of the described clinical vignettes, we believe that careful monitoring of a patient's fluid and electrolyte balance is essential as the symptoms of NDI tend to become aggravated during pregnancy. Also, our report shows that the administration of thiazyde diuretics during NDI manifestations in the pregnant woman was safe and effective.

Congenital nephrogenic diabetes insipidus (NDI) is a hereditary renal disorder characterized by the body's failure to concentrate urine in response to the antidiuretic hormone (ADH). Ninety percent of cases are caused by X-linked mutations of the ADH V2 receptor and 10% by autosomal mutations of the aquaporine2 water channel (AQP2). No data was found concerning the overall prevalence of females with NDI and their chances for successful pregnancies. We present the case of a woman patient diagnosed with NDI in childhood and report on the influence of NDI on the course of two of her pregnancies and the treatment of her fluid and electrolyte disturbances during these pregnancies. A 23-year-old primigravida at 15 weeks gestation was referred to the hospital with dysuria and abdominal pain. The patient had polydipsia and polyuria and at admission her urine volume reached 17 L/day. Anamnesis revealed the diagnosis of NDI in childhood and hydrochlorotihiazyde treatment until the age of 18. The patient claimed that she has ingested large quantities of fluid for as long as she remembers and that during adulthood the NDI symptoms didn't impact her quality of life. In an early pregnancy, she observed gradual aggravation of the symptoms which were excessive thirst (up to 18 L/day) and increased urine output. Upon examination, the patient revealed no definite evidence of dehydration. Upon examination she was found to have the following: blood pressure of 100/80 mmHg, serum sodium 132 mmol/L, urine specific gravity 1.003, serum creatinine 44.2 µmol/L, 276 mOsm/L plasma osmolality, 78 mOsm/L urine osmolality, culture urine positive (E. coli 10^7), USG detected bilateral pelvicalyceal dilatation and an enlarged bladder. The treatment strategy was equalization of electrolyte disturbances; reduction of diuresis and urine retention; treatment of urinary tract infection; and treatment to prevent recurrent urinary infections; thus lowering the risk of miscarriage or preterm delivery. Hydrochlorothiazyde was used in the treatment of polyuria in incrementally increasing doses up to 75 mg/day which reduced the diuresis to 7 L/day. The optimal dose of hydrochlorothiazyde was determined to be 37.5 mg/day (Fig. 1).

Potassium and magnesium oral supplementation was provided under control in serum; and there was 24h urine collection. During therapy the patient developed gestational diabetes and was placed on a well-controlled diet.

Regular monitoring of the fetus was provided. Elective C-section in the 38th week of gestation was performed, and a female neonate was delivered with an Apgar score of 10. The woman and her infant were discharged from hospital on the 8th day of the neonate's life. Genetic tests were performed in which two different autosomal AQP2 recessive

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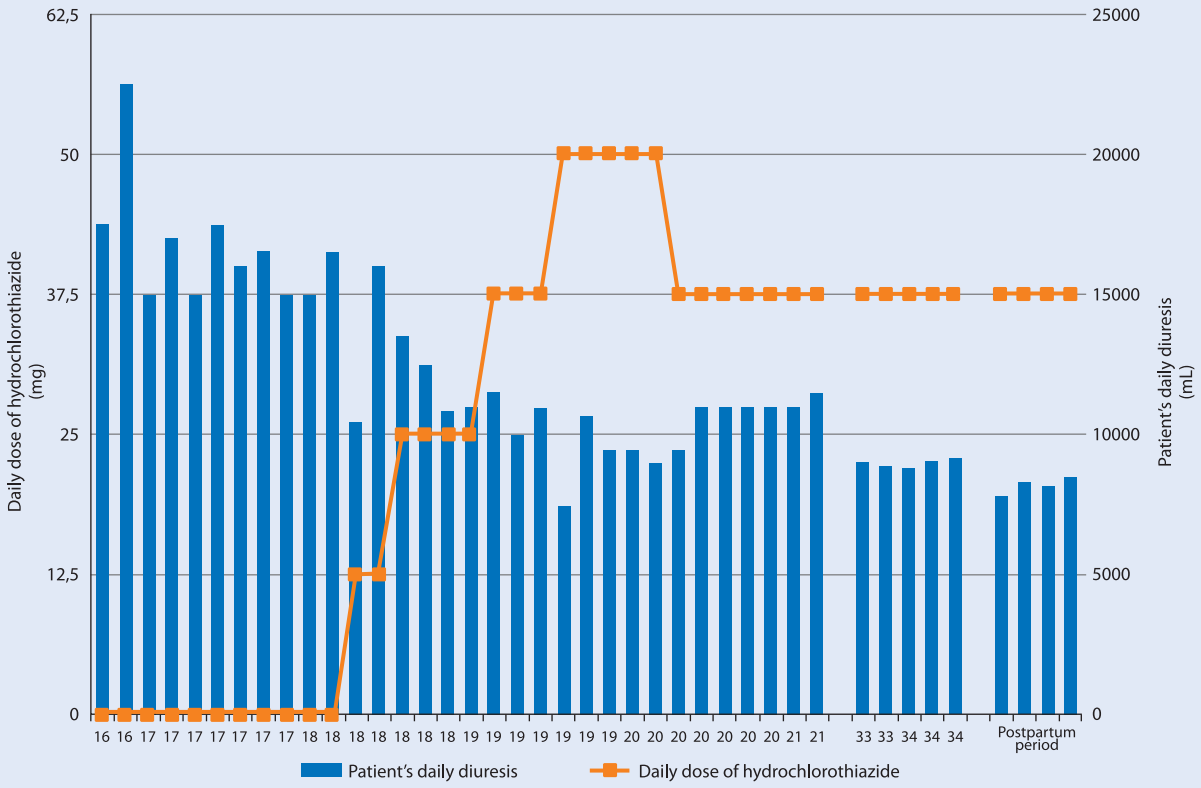

Figure 1. The patient's daily diuresis depending on the daily dose of hydrochlorothiazide

mutations were detected in the patient. Additionally, the same treatment strategy was performed during the patient's 2nd pregnancy resulting in another successful delivery.

Based on the described clinical vignettes, we believe that careful monitoring of the patient's fluid and electrolyte balance is essential as the symptoms of NDI tend to become aggravated in pregnancy. Although thiazyde diuretics belong to the FDA pregnancy B category (risk of fetus thrombocytopenia, hemolytic anemia, electrolyte imbalances), during NDI manifestations, they can decrease diuresis and, in the case presented, the benefits outweigh the potential risks.