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Case Report: A Rare Presentation of Fowler's Syndrome in Pregnancy with Mitrofanoff Procedure

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Abstract : Introduction: Fowler's syndrome, first described by Clare Fowler in 1985, is a rare urological condition characterized by difficulty in urination due to the abnormal function of the urethral sphincter. It predominantly affects young women and leads to chronic urinary retention. The main concern is to ensure regular bladder emptying. The Mitrofanoff procedure, a surgical creation of a continent urinary diversion, is often performed in patients with Fowler's syndrome who require long-term catheterization. This procedure involves creating a conduit (from the appendix or a segment of the small intestine) between the bladder and the skin, allowing for intermittent self-catheterization to manage urinary retention. Study: A 39-year-old woman with a history of Fowler's syndrome and a Mitrofanoff procedure was booked in our obstetrics department at 8 weeks of gestation. The patient was gravida 3, para 0+2, (both first-trimester miscarriages) with a BMI of 40 and type I diabetes managed with insulin. Fowler's syndrome was diagnosed at 23 years of age. A sacral nerve stimulator device was initially placed but stopped working after one year due to trauma so it was removed and the suprapubic catheter was inserted. The Mitrofanoff procedure was performed at the age of 29, where the bowl was used to create—a continent-catheterized stoma, which has been successfully used for intermittent self-catheterization. During her current pregnancy, some vaginal bleeding was experienced at 10 weeks and was started on progesterone pessaries to support the pregnancy, with advice to discontinue them at 16 weeks. A dating scan at 13 weeks was normal. At the beginning of the second trimester, at 14+4 weeks, the patient was admitted with signs and symptoms of a Urinary tract infection and was treated with antibiotics. Despite these challenges, she continued self-catheterization through the Mitrofanoff stoma to prevent urine retention. The patient was seen jointly in the diabetic clinic and had a fetal anomaly scan at 20 weeks, which was normal. Her current gestational age is 21 weeks and the plan was made for a joint diabetic clinic every 4 weeks, serial growth scans from 28 weeks to assess fetal growth, and to start low molecular weight heparin (LMWH) from 28 weeks due to the intermediate risk of venous thromboembolism (VTE). Regular monitoring was arranged by a multidisciplinary team, including an obstetrician and urologist, with admission from 30 weeks and planned cesarean birth in a tertiary center at 34 weeks. Conclusion: Pregnancy in women with Fowler's syndrome who have undergone a Mitrofanoff procedure is rare, and management requires careful planning and a multidisciplinary approach. This case highlights the importance of individualized care plans and close monitoring of the mother and fetus. The patient with a risk of recurrent UTIs, coupled with her diabetes and high BMI, necessitated coordinated care across specialties to ensure the best possible outcomes. The Mitrofanoff procedure proved effective in managing her urinary retention, allowing her to maintain self-catheterization in pregnancy. The multidisciplinary team approach was crucial in addressing her complex medical needs, involving obstetrics, urology, and endocrinology. This case adds valuable information to the limited literature on pregnancy management in patients with Fowler's syndrome with mitrofanoff procedure highlighting the need for comprehensive, individualized care and the involvement of a multidisciplinary team to achieve the best results.

Keywords : fowlers syndrome, mitofanoff procedure, pregnancy, chronic urine retension
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