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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 in managing this condition is ensuring regular bladder emptying. Clam cystoplasty is a bladder augmentation surgery in which the bladder is clam-shelled open, and a segment of the intestine is used to increase the bladder's capacity and reduce bladder pressure. 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: This case study examines a 39-year-old gravida 3, para 0+2 woman with a BMI of 40, Fowler's syndrome, type I diabetes, and post-traumatic stress disorder (PTSD), presenting at Dumfries and Galloway Royal Infirmary at 8 weeks of gestation. Diagnosed with Fowler's syndrome at 23, . A sacral nerve stimulator (SNS) device was initially placed but was subsequently removed after one year due to malfunction caused by trauma, subsequently she had undergone clam cystoplasty and the Mitrofanoff procedure for bladder management. Her pregnancy was complicated by vaginal bleeding at 10 weeks, treated with progesterone pessaries, and a urinary tract infection at 14 weeks, managed with antibiotics. Despite these challenges, she continued self-catheterization through the Mitrofanoff stoma and was placed on prophylactic antibiotics. Her diabetes was well-controlled on insulin, and a 20-week fetal anomaly scan was normal. The multidisciplinary team, including an obstetrician and a urologist, planned for serial growth scans and the initiation of low molecular weight heparin (LMWH) from 28 weeks due to the intermediate risk of venous thromboembolism (VTE) and to continue six weeks after delivery. A planned cesarean delivery at 37 weeks was arranged, with an MRI scan scheduled later in the pregnancy to assist in surgical planning, ensuring the preservation of the Mitrofanoff stoma's function. The surgery will occur in an elective setting and include a consultant urologist. Conclusion: Pregnancy in women with Fowler's syndrome who have undergone Clam cystoplasty and the 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 both mother and fetus. The patient's 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 during 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 who have undergone the Mitrofanoff procedure, highlighting the need for comprehensive, individualized care and the involvement of a multidisciplinary team to achieve the best results.

Keywords : fowler's syndrome, clam cystoplasty, mitrofanoff procedure, pregnancy
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