

PREGNANCY IN A WOMAN WITH FOWLER’S SYNDROME

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ABSTRACT

Fowler’s syndrome- Is a rare cause of urinary retention in young woman which can be quite debilitating and challenging to manage, especially when they are pregnant. In most cases, there is no other neurological or anatomical abnormality to explain their inability to pass urine. The abnormality lies in the urethral sphincter’s failure to relax, hence leading to retention. We present a successful outcome of a pregnancy in a 31-year-old pregnant woman with underlying Fowler’s syndrome managed with a Mitrofanoff conduit and neuromodulator.

CASE

- Mrs X was diagnosed with the condition 20 years prior to the index pregnancy, due to recurrent urinary tract infections associated with symptoms of urinary retention, voiding dysfunction and pelvic pain. Although she was initially managed with intermittent self-catheterisation, she finally had a Mitrofanoff conduit and neuromodulator (connecting the dome of the bladder with umbilicus) for urinary incontinence and an ileostomy for bowel management.
- Mrs X’s first presentation to our obstetric department was at 6 weeks gestation with symptoms of urinary tract infection. She responded well to the antibiotic treatment and was discharged to be followed up in Antenatal clinic under consultant led care. At booking, she described being Asthmatic, Epileptic and hypothyroidism along with Fowler’s syndrome. She was started on low-dose-maintenance regimen of antibiotic to cover the remainder of her pregnancy along with aspirin and her regular medications(levetiracetam and levothyroxine). She had a normal dating scan and her first trimester bloods were also normal.
- Mrs X had several hospital admissions along the course of her pregnancy for parenteral antibiotic, due to recurrent urinary tract infections associated with significant pelvic pain. A multidisciplinary team approach involving the microbiologist, urologist and obstetric team was adopted to manage her symptoms as an in-patient.
- During the third trimester, her urinary symptoms worsened resulting in more frequent and prolonged hospital admissions. US kidney showed severe/ moderate hydronephrosis on both sides, however she was clinically improving with parenteral antibiotics. At 35 weeks gestation she was administered a course of dexamethasone to promote fetal lung maturation. Serial ultrasound growth scans demonstrated normality of fetal growth, liquor volume and dopplers.
- After several discussions, it was agreed that vaginal delivery would be the safest for Mrs X and would be in her and her baby’s best interest, but a detailed plan for delivery was put in place after discussion with the urologists and surgeons in case she needed an emergency caesarean section.
- At 39 weeks, Mrs X was induced and went on to deliver a healthy female infant with the help of Neville-Barnes forceps weighing 3.01kg. Post-natal recovery was uneventful on the ward

DISCUSSION

Fowler's syndrome was first identified in 1985 as painless urinary retention in young women¹ and is associated with the abnormality that lies in the urethral sphincter’s failure to relax, hence leading to retention.

Professor Paul Mitrofanoff created the Mitrofanoff procedure in 1976². The Mitrofanoff was introduced to permit bladder drainage where voiding or urethral self-catheterisation is not possible. The purpose of this operation is to form a channel between the bladder and the wall of the abdomen to allow intermittent self-catheterisation.

Embarking on pregnancy creates several complications in an already complex clinical scenario. As in Mrs X's case the main concern was recurrent urinary tract infections and pelvic pain requiring hospital admissions for parenteral antibiotics and analgesia.

Meta-analyses of studies evaluating bacteriuria in pregnancy have concluded that there are confirmed associations with preterm delivery and low birth weight³. In addition, there are increased risks of pre-eclampsia, anaemia, chorioamnionitis and postpartum endometritis³. Fetal risks include fetal growth restriction, stillbirth, perinatal mortality, mental retardation and developmental delay³.

There is no evidence in literature to determine the ideal mode of delivery specifically for a case of Fowler's syndrome.

With the lack of evidence to guide the ideal mode of delivery and no clear reference, we concluded vaginal delivery would be the safest course of action. This decision was made in conjunction with the urologists, surgeons, microbiologists and other obstetricians, keeping in mind the chronic and incurable nature of the underlying disease and to prevent any possible further damage to the implanted bladder.

CT image showing
Neuromodulator



Anomaly scan – no
abnormalities



LEARNING POINTS

- A pregnant patient with Fowler’s syndrome requires multi-disciplinary care for an optimal outcome.
- Recurrent urinary tract infection should be anticipated and managed aggressively.
- There is no evidence in literature to determine the ideal mode of delivery specifically for a case of Fowler's syndrome and decision should be made based on obstetric indicators/ additional medical co-morbidities and will require individualized management plan by adopting a multi-disciplinary team approach.

References

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3. Fowlers Syndrome and Pregnancy A T Khan, R B Kinder, R G Hayman <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3028175/>