**INTRODUCTION**

Ectopia vesicae is a congenital abnormality having a spectrum of anomalies of the lower abdominal wall, bladder, anterior bony pelvis, and external genitalia. It develops at the third week of embryonic life. Its incidence is 1 in 30,000 to 50,000 people and the predisposition is greater for males in a ratio of 3:1. Women with bladder exstrophy are fertile and able to have children without this disease. Reconstruction can have significant effect on pregnancy and vice versa. This is the case of a pregnant female with reconstruction of urinary system, who experienced bad obstetric history.

**CASE REPORT**

A 23-year woman referred from Hafizabad, Punjab, presented in the Emergency Department of Jinnah Hospital, Lahore, because of high-risk pregnancy. She presented at 32 completed weeks of gestation with complaints of reduced fetal movements and mild lower abdominal pain for one day. This was her third spontaneous conception. The first two pregnancies had ended up in spontaneous miscarriage. She was born with ectopia vesicae. Findings of her examination were ectopic urethral opening just above the pubic symphysis measuring 2 - 3 cm with continuous urinary leakage from the opening with a normal bladder neck, and bladder capacity of 80 ml. The vaginal opening, clitoris and labia minora and labia majora were normal. Meatoplasty was attempted many times which had failed. Her ureterosigmoidostomy was done at the age of 9 years through multi-staged surgeries. Since then she was passing urine from the anal route along with fecal matter. She denied any urinary or fecal incontinence. Her renal function tests and serum electrolytes remained in the normal range in early years after surgery. She was prescribed sodium bicarbonate 900 mg, potassium chloride 20 MEq daily, and tablet septran life long. She was advised frequent voiding every 2 hours. Rest of the medical history was unremarkable. Her family history was not significant. Her BMI was 28 kg/m². Her general physical examination revealed mild poliar. On abdominal examination, there was a supra-pubic transverse scar of 8 cm. Her symphysio-fundal height was 30 cm with longitudinal lie and cephalic presentation; and the fetal heart rate was in the range of 146 - 158 bpm. She had mild palpable contractions for 10 seconds every 10 - 15 minutes. Cervix was closed on vaginal assessment. Instead of an opening at site of routine urethral sphincter, an old healed scar was found. Clitoris and vagina were normal. CTG was reactive at the time of admission. Laboratory evaluation revealed anemia (Hb=9 gm/dL) with raised TLC (19,500/Ml, P 81%, L 13%, M 6%) and normal platelet count and red blood cell count parameters. Her blood urea level (71 mg/dL) and creatinine was raised (1.7 mg/dL), Uric acid, sodium and postossium were normal. Chloride (110 mmol/l) was raised and HCO₃ was 19 mmol/l reduced. Obstetric ultrasound scan showed 32 weeks gestation with normal amniotic fluid index (AFI = 15 cm), fundo-posterior placenta and fetal weight of 2176 grams. Umbilical artery Doppler study was normal (S/D ratio = 2.65). Both maternal kidneys showed moderate hydrenephrosis and hydroureter with grade I echogenicity of renal parenchyma.

It was a high-risk pregnancy with ureterosigmoidostomy, preterm labour, reduced fetal movements, and mild anemia along with bilateral hydrenephrosis and hyperchloremic metabolic acidosis. Considering this, she was managed with multidisciplinary approach. Tocolysis was done with calcium channel blocker to stop contractions. Anemia was corrected with PCV transfusion. Steroid cover was given for fetal lung maturity.

**ABSTRACT**

The patient was born with bladder exstrophy and underwent multiple surgeries for its correction. Finally, she had ureterosigmoidostomy at the age of 9 years, which ultimately led her to live till reproductive age. The children born with this defect are capable of surviving till adult life. She was received by us with preterm labour, referred from Hafizabad. She also had hyperchloremic metabolic acidosis and mild hydrenephrosis. She was managed with multidisciplinary, modified care but the pregnancy ended up in an emergency cesarean section due to non-reactive cardiotocograph and persistently reduced fetal movements. She had fortunately successful pregnancy outcome. To the authors’ knowledge, this is the first reported case of pregnancy in a treated case of ectopia vesicae in Pakistan.

**Key Words:** Pregnancy. Ectopia vesicae. Ureterosigmoidostomy. Hyperchloremic metabolic acidosis. Preterm labour.

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Close monitoring of her renal functions, electrolytes and arterial blood gases was started along with antibiotic prophylaxis for prevention of UTI. She was put on oral sodium bicarbonate and potassium chloride along with calcium supplement for correction of hyperchloremic metabolic acidosis. For moderate hydronephrosis and hydroureter, no intervention was planned on opinion of urologist.

After consent for surgery and publication, cesarean section was done under spinal anaesthesia at 33+6 weeks due to persistently reduced FHR variability on CTG trace. Abdomen was opened through the previous scar; there were flimsy adhesions between the visceral and parietal peritoneum anterior to lower uterine segment. There was no bladder in anterior relation to the lower uterus consistent with the history of cystectomy. Peritoneum was separated by blunt and sharp dissection. It was difficult to find a landmark of lower uterine segment for uterine incision because of the absent bladder. Routine procedure for cesarean section was done. Ureteric diversion was visible in sigmoid colon between flimsy bowel adhesions.

A male baby of 1.8 kg was delivered and observed in NICU for prematurity and LBW, and was later on discharged in good condition. Postoperative period was uneventful. Patient was discharged from the hospital with her baby in satisfactory condition and was advised to continue medications for hyperchloremic metabolic acidosis and continue follow-up to the urologist.

**DISCUSSION**

To the authors’ knowledge, this is the first reported case of pregnancy in a treated case of ectopia vesicae from Pakistan. Bladder exstrophy is a rare anterior midline defect that causes a series of genitourinary and muscular malformations. Genetic mutation in gene P63 DNA and Delta N p 63 (anti-apoptotic factor) is the causative factor for this defect.5 When a female is born with this defect then urethra is absent, labia minora are rudimentary, and usually clitoris is bifid.6 Labia majora are normal and anus tends to be forward facing and vagina may be obscured by a ventrally displaced perineum.5,6 There is a fibrous band between the widely divergent and separated pubic rami. The mons veris is absent. Internal genitalia may be normal.6,7

At puberty, genital and reproductive functions are important issues; in addition these patients have higher rates of infertility.8 Psychosocial and psychosexual outcomes depend on long-term multidisciplinary care to improve quality of life.9 Due to advances in reconstructive surgery, more patients lead a near normal life.

Few successful pregnancies after urinary diversion with an ileal conduit or ureterosigmoidostomy have been reported. There is a significant increase in the risk of urinary tract infections and hydronephrosis during pregnancy in these patients, especially when the expanding uterus compresses the conduit.9 It is advisable to put all these women on antibiotic prophylaxis throughout the pregnancy. There is risk of acute pelvic floor prolapse during pregnancy and also risk of miscarriage and pre-eclampsia. These social and physical factors make these pregnancies high risk as well as precious, and thus they need monitoring by both obstetric and urological units. Pre-conception renal evaluation with regular follow-up in antenatal period is mandatory.8

Planned cesarean delivery at term is the appropriate mode of delivery.6 It allows for timely planning to ensure that delivery is carried out by an experienced obstetrician with the help of expert surgeon, which helps reducing surgical morbidity in an already complicated procedure. Multiple pelvic adhesions are commonly encountered during cesarean deliveries and complications like ureteric transection and fistula formation occur frequently owing to the distorted pelvic anatomy.8 Reconstructive surgeon should be available at the time of elective delivery to better facilitate the coordinated care of both mother and fetus.6 Major neonatal risk involved is prematurity; premature delivery rate upto 29% have been reported.8 Bowel is used in urologic surgery as a substitute for the ureter, bladder and for bladder augmentation and genital reconstruction.7 Since intestine is not naturally meant to function as a conduit or a storage vehicle for urine, so numerous complications may occur including disordered electrolyte metabolism, altered hepatic metabolism, abnormal drug metabolism, infection, stone formation, nutritional disturbances, growth retardation and rarely colonic cancer.7 Metabolic complications are quiet common, and about 80% of patients with urinary diversion have hyperchloremic metabolic acidosis. Other electrolyte abnormalities are low K+, Ca+, Mg, elevated ammonia, blood urea and creatinine. Hyperchloremic metabolic acidosis is a common complication due to urinary diversion in intestine.2 For its treatment, usually alkalinising agents or chloride channel blockers are prescribed. Oral sodium bicarbonate is very effective in restoring normal acid-base balance.2

The present case had majority of the above complications, including previous two pregnancy losses. She was treated with multidisciplinary care with good feto-maternal outcome.

**REFERENCES**


