Case report:

A case of pregnancy in patient with reconstructed bladder extrophy

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Abstract
Bladder extrophy (BE) is a rare anterior midline defect resulting into complex genitourinary malformations which require complex surgical treatment having incidence of 1:30000-50000. The reconstruction surgeries for BE and their outcomes have improved with time resulting in higher survival rates and near-normal life in patients undergoing these operations. With the history of multi-staged bladder and pelvic reconstruction operations in these patients, obstetric management presents a big challenge which needs to be addressed by Obstetrician and Urosurgeon. Preconceptional renal evaluation with regular follow up in antenatal period should be mandatory. Planned Caesarean delivery at term is appropriate mode of delivery which allows for timely planning and expert opinion. We presented a 20 year old Primigravida with 6 month amenorrhoea, married since one year, conceived spontaneously, had undergone 4-staged bladder and pelvic reconstruction operations at the age of 7, presented with pain in abdomen and had preterm vaginal delivery.

Key-words - Bladder Exstrophy, Pregnancy

Introduction
Bladder extrophy (BE) is a rare anterior midline defect resulting into complex genitourinary malformations which require complex surgical treatment. Incidence of Bladder extrophy is 1:30000-50000 with predisposition more to the male gender with ratio of 3:1[1]. The reconstruction surgeries for BE and their outcomes have improved with time resulting in higher survival rates and near-normal life in patients undergoing these operations. With the history of multi-staged bladder and pelvic reconstruction operations in these patients, obstetric management presents a big challenge which needs to be addressed by both Obstetrician and Urosurgeon. We present a case of a 20 year old pregnant female who had undergone 4-staged bladder and pelvic reconstruction surgeries in her childhood.

Case report
A 20 year old Primigravida with 6 month amenorrhoea came with pain in abdomen to our centre. She was married since one year, conceived spontaneously and was registered with private practitioner for ANC check-ups. She had undergone 4-staged reconstruction operations in childhood, for bladder reconstruction with Colonocystoplasty augmentation, Mitrofinoff's procedure for urinary diversion and continence alongwith posterior osteotomies and repair of pubic diastasis in our institution at the age of 7. Patient did not follow up since last 8-9 years during which her Mitrofinoff's
stoma was blocked and she had developed the paraurethral fistulae which rendered her incontinent.

Her Antenatal sonography confirmed 24 weeks gestation with a healthy and non-anomalous baby. On examination there was a linear midline laparotomy scar over lower abdomen 7-8 cms in length, with bifid clitoris and widely separated labia. Catheter could not be passed through urethral meatus or from the paraurethral fistulae. Perspeculum examination revealed no discharge or bleed. The cervical os was open and membranes were present. Patient was treated with progesterone and bed rest in order to prevent preterm delivery with a prompt surgical opinion to decide on management options if required. Our goal was to conserve the pregnancy and perform elective caesarean delivery at term with surgical team at hand. However, preterm labour progressed and delivered a male child of 800 grams before emergency cervical encirclage could be tried. Baby was admitted in NICU and died in 48 hours due to prematurity. Patient had a stable postnatal course but the dribbling of urine continued for which she was referred to Urology department.

Discussion

BE is rare malformation involving development failure of cloacal membranes with open bladder and pelvic and genital malformations. Its clinical characteristics vary from simple to most complex form such as cloacal extrophy. Bifid genitals are frequent and BE is frequently associated with other congenital malformations. Its incidence is 1:30000 with male to female ratio about 3:1[1]. Its treatment is clearly surgical and is carried out in first years of life, mainstay of which is to provide continent bladder. Treatment consists of staged reconstruction of bladder exstrophy (complete primary repair).

Staged reconstruction consists of closure of bladder plate and posterior urethra and bladder neck reconstruction along with osteotomies to repair pelvic deformities [2]. Bladder augmentation is surgery to increase bladder size and involves tissue grafts (anastomosis) from a section of the small intestine (ileum), stomach, colon or other substitutes that are attached to the urinary bladder by sewing or stapling [3].

Mitrofinoff's procedure constructs a catheterizable stoma between the skin and bladder /urinary reservoir with help of appendix, this stoma needs to be self-cathetered and renders patient continent.

The quality of life of individuals affected with BE is considerably affected by the disease, but the advances in reconstruction surgery while leading to increased survival rate have also contributed more and more patients to remain continent, lead a near normal life and thus get pregnant and bear children. Due to complexity of the disease and its surgical reconstruction involving changes in pelvic anatomy and increased adhesions around uterus, the obstetric management of such patients becomes a great challenge.

Patients with BE reconstruction are generally self conscious and have limited sexual activity because of cosmetic appearance of genitals, scars of multi-staged operations, in addition these patients have higher rates of infertility. According to Espinosa-Chavez et al, "first pregnancy in a woman with ureterosigmoidostomy was published in 1922 and since then only 252 cases of pregnancy after urinary diversion have been reported" [4]. The leading cause of infertility is tubal, expected due to large number of adhesions which affect tubal function; rates of infertility have varied from 8% to 32% in different studies. There is a high rate of acute pelvic
floor prolapse during pregnancy and high rate of miscarriages and preeclampsia complicate the gestation in these patients [3], these social and physical factors make these pregnancies high risk ones as well as precious in nature and thus require monitoring by both obstetric and urological units. Preconceptional renal evaluation with regular follow-up in antenatal period should be mandatory. Planned Caesarean delivery at term is appropriate mode of delivery [5]. 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 caesarean deliveries and complications like ureteric transection, fistula formation occur frequently owing to the distorted pelvic anatomy. Major neonatal risk involved is prematurity, premature delivery rate upto 29% having been recorded [3]. Greenwell et al were of opinion that vaginal delivery should only be considered where pregnancy is uncomplicated and in presence of senior obstetrician. Reproductive outcomes as discussed by Deans R et al in 2012 suggest 66% of their sample size had conceived and of the reported pregnancies 35% miscarried and 7% were stillborn. Major complications including transection of ureter in 4%, fistula formation in 4% and postpartum haemorrhage in 8% have been reported [3]. While in another study by AM Giron et al in 2011, 50% patients had genital prolapse [6].

Conclusion
The patients with Bladder exstrophy have high rates of infertility and social stigma due to previous scars and appearance of genitalia. Rates of miscarriages, preeclampsia with Urinary tract infection and genital prolapse are high which altogether make these pregnancies very high risk. Planned caesarean delivery is preferable and should be conducted by expert obstetrician along with urosurgery aid to avoid surgical morbidity.

References