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Multidisciplinary Management of Pregnancy in Bladder Exstrophy: A Case Report

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 33-year-old
Final Diagnosis: Control of high-risk pregnancy • pregnancy and delivery in a patient with bladder exstrophy
Symptoms: Bladder dysfunction • intrauterine growth retardation • pregnancy • recurrent urinary tract infections
Clinical Procedure: Cesarean section • laparotomy
Specialty: Obstetrics and Gynecology

Objective: Rare disease

Background: Classic bladder exstrophy (CBE) is a rare congenital condition affecting multiple organs, primarily the urinary and musculoskeletal systems. Management involves multiple reconstructive surgical procedures and aims to maintain renal function, achieve urinary continence, and support normal reproductive health. Pregnancies in women with CBE can be complicated by recurrent urinary tract infections, pelvic prolapse, and a higher risk of preterm birth.

Case Report: We report the case of a 33-year-old woman with a history of CBE, admitted at 32 weeks of pregnancy with fever and symptoms of a urinary tract infection. Her medical history included multiple surgeries, including formation of neovesica from the cecum with appendicovesicostomy to the abdominal wall. The pregnancy was complicated by recurrent urinary tract infections, anemia, gestational diabetes, and stage IV uterine prolapse. A multidisciplinary team managed her care, with imaging revealing altered abdominal anatomy and breech fetal presentation. At 37 weeks, due to concerns about fetal growth restriction, breech position, and potential prolapse exacerbation, an elective cesarean delivery with median relaparotomy and vertical uterine fundal incision was performed to avoid emergency intervention. Both the delivery and postpartum recovery were uneventful despite the complexity of the case.

Conclusions: Managing pregnancy in patients with CBE requires close, multidisciplinary collaboration to address potential complications. Detailed delivery planning and vigilant monitoring are crucial to ensure maternal and fetal safety.

Keywords: Bladder Exstrophy • Cesarean Section • Congenital Abnormalities • Fetal Growth Retardation • Urinary Tract Infections

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Introduction

CBE, or classic bladder exstrophy, is a rare congenital condition, with its incidence ranging from 2.1 to 4.0 per 100 000 live births. It is 3-4 times more common in males than in females [1]. Incidence varies according to the geographic region and socioeconomic status [2]. CBE is one of the exstrophy-epispadias complex (EEC) anomalies, which is the most severe type of midline abdominal malformation. This condition primarily affects multiple organs within the body, including the urinary system, musculoskeletal system, pelvis, pelvic floor, abdominal wall, genitalia, and in some cases, the spine, and anus [3,4].

Managing patients with CBE poses a significant challenge. Beyond achieving a satisfactory closure of the abdominal wall, the treatment goals encompass kidney preservation, achieving urinary continence, and preventing complications [5]. There are 2 primary treatment options for CBE: the Modern Staged Repair of Exstrophy (MSRE) and the Complete Primary Repair of Exstrophy (CPRE). The MSRE involves initial closure of the bladder plate and posterior urethra, followed by subsequent steps of epispadias repair and bladder neck reconstruction. In contrast, CPRE achieves closure of the bladder, urethra, and urethral meatus in a single procedure. It is also worth noting that ureterosigmoidostomy has become less common in contemporary practice, especially in 2024, due to advancements in other reconstructive techniques [5].

In females, the anatomy and function of the reproductive organs usually is unaffected. However, the cervix, in most instances, attaches low on the superior vaginal wall near the introitus. Combined with the absence of cardinal ligaments, there is an increased risk of experiencing vaginal or uterine prolapse, with up to 50% of cases being affected [4].

Most females with CBE conceive spontaneously, but 20-40% of pregnancies end with a spontaneous abortion [6]. Pregnancy is often complicated with recurring urinary tract infections (UTIs). Usually, delivery is performed via planned cesarean delivery (CD) to reduce the risk of complications [1].

Case Report

This case study presents a 33-year-old woman with a complex medical history, primarily related to CBE, who presented to our obstetric unit at 32 weeks of pregnancy.

Medical History

Born outside Slovenia, she underwent her initial surgery at the age of 2 for bladder exstrophy repair. Subsequent procedures included bladder neck closure at age 13, vaginal and vulvar reconstructive surgery, and ileocystoplasty with Mitrofanoff appendicovesicostomy, performed at a tertiary medical center. These interventions enabled her to manage urination effectively through self-catheterization, reaching the neovesica via a catheter inserted through the anterior abdominal wall, up to 6 times daily. At 22 years of age, she underwent sacrofixation to treat stage IV uterine prolapse, requiring a blood transfusion. Three years later, she was diagnosed with stage 3 chronic renal failure due to obstructive uropathy, with renal scintigraphy showing a small left kidney with 67% functionality and a normal-sized right kidney functioning at 33%. Reno-parenchymal arterial hypertension developed as a complication, necessitating antihypertensive treatment with bisoprolol. At 30 years of age, she underwent a LLETZ procedure due to atypical squamous cells in her PAP smear.

Prenatal Course

During her first pregnancy, at age 33, she was admitted at 32 weeks of gestation with a high fever due to a recurrent UTIs. Her perinatal history was notable for anemia, insulin-dependent gestational diabetes mellitus, and recurrent stage IV uterine prolapse.

Management of the UTI involved intravenous ceftriaxone, while anemia was treated with oral iron (iron(II) sulfate). Given the complexity of her medical history and previous surgical interventions, a multidisciplinary team promptly developed a delivery plan. The fetus presented in breech position, further complicating the case. The team included obstetricians, urogynecologists, urologists, nephrologists, diabetologists, and radiologists to ensure comprehensive care. A urinary tract ultrasound was performed to rule out any underlying pathology contributing to the recurring UTIs. Subsequently, an abdominal MRI revealed significant anatomical changes: extensive adhesions, absence of pelvic bones and bilaterally dilated ureters, with the left one running horizontally between the uterine isthmus and the anterior abdominal wall, before reaching the neovesica in the lower right abdominal quadrant (**Figure 1**). The neovesica was present in the right hypogastrium, originating from the cecum, with the appending functioning as a duct from the neovesica to the abdominal wall (**Figure 2**).

Delivery

At 37 weeks, due to concerns regarding fetal growth restriction (FGR), breech presentation, and the potential exacerbation of uterine prolapse, an elective CD was performed to avoid emergency intervention and to minimize associated risks. The procedure was conducted under combined spinal and epidural anesthesia via a midline relaparotomy, initiated approximately 4 cm below the umbilicus (**Figure 3**). During the surgery, adhesions were encountered between the uterus and the anterior abdominal wall at the lower portion of a prior

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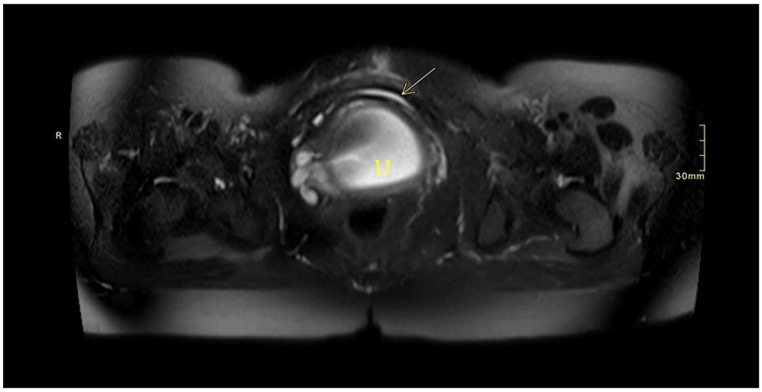

Figure 1. MRI image of the abdomen of the woman at 34 weeks of gestation, showing a transversal cross-section in the level of the uterine isthmus (U). In front of the uterus the arrow is pointing at the left ureter.

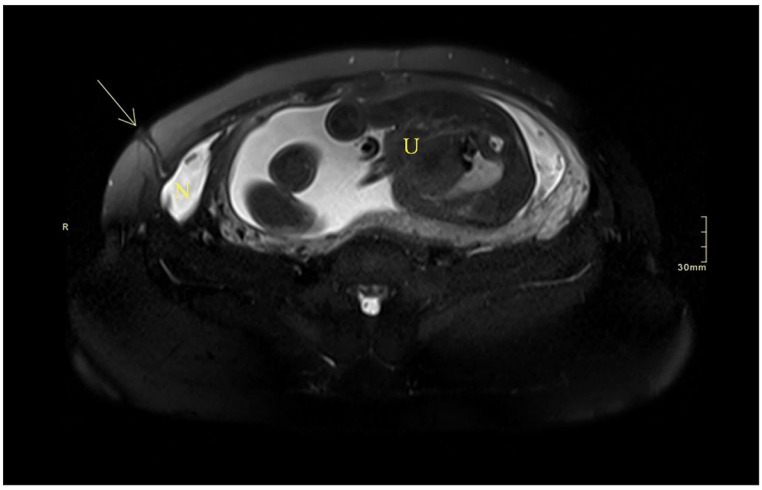

Figure 2. MRI image of the abdomen of the woman at 34 weeks of gestation, showing a transversal cross-section at the level of the neovesica (N), derived from the cecum and the appendicovesicostomy (arrow), with its ostium at the right lower abdominal wall. Left of the neovesica, taking up the majority of the abdominal space on this cross-section, is the pregnant uterus (U).

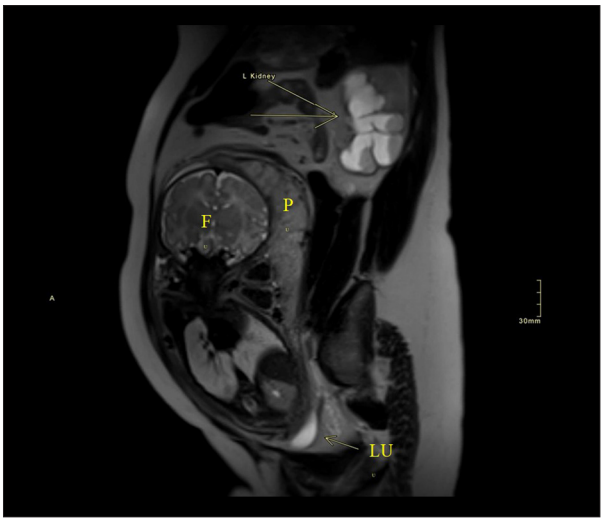

Figure 3. MRI image of the abdomen of the woman at 34 weeks of gestation, showing a sagittal cross section at the level of the left kidney with significant hydronephrosis. The uterus with the fetus (F) in breech position and the placenta on the posterior uterine wall (P) is seen anterior to the left kidney. At the uterine cervix, the arrow points to a hyperechoic structure, which is the left ureter (LU).

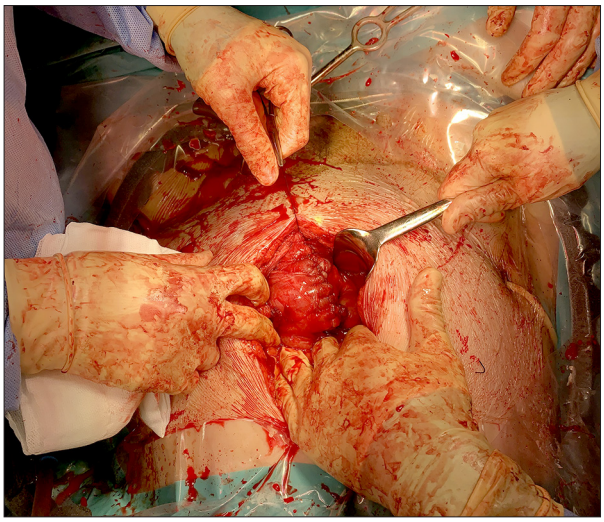

Figure 4. The cesarean delivery, showing the sutured uterus after the longitudinal incision at the fundus and the extraction of the fetus head-first. The photo was taken from above the patient's head.

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laparotomy incision. To minimize complications, these adhesions were left intact. The incision was extended superiorly above the umbilicus, where the abdominal wall was free, allowing improved surgical access. Extensive adhesions of the intestines to the anterior uterine wall were identified, necessitating meticulous dissection. Adhesions on the right side of the uterus were carefully resolved, enabling a controlled incision into the uterine fundus (**Figure 4**).

The fetus, positioned breech, was carefully extracted headfirst, with Apgar scores of 9 at 1 and 5 minutes. The prenatal FGR diagnosis was accurate, since the newborn's weight was 2040 grams (4 pounds 8 ounces). The placenta was delivered spontaneously with minimal traction. The uterine cavity was inspected and confirmed to be empty, and the uterine incision was sutured in 2 layers, with effective hemostasis achieved. To promote uterine contraction, 10 IU of intravenous oxytocin and intramuscular carboprost were administered.

A urogynecology specialist was consulted to evaluate the uterine prolapse. The fixation to the promontory was deemed appropriate; however, extensive adhesions precluded a comprehensive assessment of the neovesica. The total estimated blood loss was approximately 500 mL.

Postpartum Course

Postoperative recovery was uneventful. The patient continued intravenous ceftriaxone to manage her UTI and received an iron infusion to address anemia. Follow-up laboratory results demonstrated normal blood and urine parameters. The surgical wound healed well, and uterine involution progressed appropriately. The patient successfully initiated breastfeeding and was discharged in stable condition 1 week after delivery, with instructions for ongoing care.

Discussion

Prenatal course

This case was particularly challenging due to 2 main factors. First, the patient presented to the obstetric unit relatively late in her pregnancy, at 32 weeks, which limited the time available for optimal prenatal care and proper planning of delivery. Late referrals in high-risk pregnancies often complicate decision-making and can impact the management of both maternal and fetal well-being. Second, there were significant gaps in the patient's medical history. As the patient had received prior surgical care abroad, we faced challenges in obtaining detailed information about her previous treatments and any complications associated with them. This lack of complete medical records created difficulties in accurately assessing her current health status and tailoring an appropriate management plan. The combination of these factors made the case particularly complex, requiring a multidisciplinary approach to ensure the best possible outcome.

Additionally, individuals with CBE face lifelong risks of infections, urinary incontinence, and bladder cancer [1]. In this patient, prior bladder augmentation and urinary diversion led to recurrent UTIs due to urinary stasis, requiring long-term antibiotic prophylaxis and regular urological follow-up, with challenges like antibiotic resistance and patient discomfort.

While reconstructive surgeries preserved fertility, they resulted in extensive adhesions, complicating subsequent procedures and diagnostics. In adulthood, the patient developed uterine prolapse, initially managed with sacrofixation. However, the prolapse worsened after pregnancy, despite an elective cesarean delivery to minimize pelvic floor strain. This case highlights the persistent challenges of pelvic stability post-reconstruction and the need for individualized, long-term follow-up. Some evidence suggests bed rest in the third trimester can reduce prolapse risk, but its effectiveness remains unclear [5,7].

Delivery

Despite the patient's complex medical condition, including recurrent (UTIs), chronic renal failure, and arterial hypertension, we successfully managed the pregnancy to term. The multidisciplinary care approach aimed to prolong the pregnancy as much as possible to allow the fetus to mature and reduce the potential adverse effects of prematurity. Premature delivery is a concern that arises from the observed high incidence of premature births associated with urogenital anomalies and recurrent UTIs [1]. According to the literature, up to 21% of patients who have undergone urinary diversion experience premature births due to UTIs during pregnancy [1]. In our patient's case, recurrent UTIs posed a substantial challenge throughout pregnancy, necessitating continuous monitoring and antibiotic management.

Considering the patient's overall condition and the growth-restricted fetus, an elective CD was performed at 37 weeks to balance the risks of prematurity and maternal complications. Vaginal delivery was not a viable option due to the patient's history of uterine prolapse, breech presentation, and FGR. Furthermore, prolonging the pregnancy was considered too risky, as it could have required an emergency CD, which would have been challenging given the patient's anatomical complexities. The delivery was meticulously planned and executed by a specialized team, highlighting the importance of advanced planning in high-risk cases.

Fetal malpresentation is frequently observed in patients with CBE, often due to altered uterine and pelvic anatomy resulting from previous surgeries [1]. The lack of consensus regarding

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the optimal mode of delivery for pregnant patients with CBE reflects the variability in individual presentations. In this case, the decision for elective CD was driven by the presence of uterine prolapse, FGR, and the patient's surgical history. The involvement of a multidisciplinary team, including radiologists, urogynecologists, and urologists, was critical in managing the unique anatomical challenges. The surgery was performed by 2 experienced obstetricians with the support of urologists and a urogynecologist, ensuring that any potential complications could be promptly addressed. Fortunately, no additional interventions were required during the procedure.

Conclusions

Managing pregnancy in patients with a history of CBE is a complex process that requires thorough coordination and close monitoring by a multidisciplinary team, including obstetricians, urologists, and other healthcare specialists. While this case highlights the importance of individualized care and proactive planning, further studies are needed to better understand the challenges and optimize management strategies for similar patients in the future.

Declaration about informed consent

We would like to thank the patient for providing oral and written consent and permission to publish this case report.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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