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Delivery in a Female Bladder Exstrophy Patient With Ileal W Pouch Continent Cutaneous Stoma

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Abstract

BACKGROUND: To deliver a case report of female bladder exstrophy patients who experienced pregnancy until the birth process.

CASE PRESENTATION: A 28-year-old woman with a history of bladder exstrophy, which received inadequate treatment during childhood, had bladder shrinkage and uterine prolapse. The patient underwent reconstruction of continent cutaneous stoma and sacrocolpopexy with mesh in her adulthood. She achieved complete continence by intermittent self-catheterization and improved her quality of life. She later got married and became pregnant. Pregnancy monitoring was done to evaluate the patient's condition and her fetus. A collaborative team was built to discuss the best option for delivering the procedure. At 38–39 weeks of gestational age, an elective cesarean section was performed to have the baby. Despite having a greater risk of complications, a woman with a history of bladder exstrophy could undergo pregnancy and give birth. The choice of delivery, either vaginal or cesarean section, is feasible for delivery.

CONCLUSIONS: An adult female with bladder exstrophy underwent pregnancy and gave birth. Prior surgery and increased risk of obstetrical complications must be considered.

Introduction

Bladder exstrophy is a rare congenital malformation of the genitourinary system. The incidence is estimated to be at 1/50,000 live births. It is a part of the exstrophy-epispadias complex (EEC), with a higher male predominance than females. Midline abdominal congenital disabilities in patients with EEC cause wide separation of the pubic symphysis, an abdominal wall defect, and an anterior-positioned empty bladder and urethra. The anomaly may be repaired during childhood while preserving the bladder and improving outcomes in terms of renal function [1].

Patients with a history of bladder exstrophy have a higher rate of infertility, ranging from 8% to 32%. Pregnant women with bladder exstrophy are scarce, accounting for only 252 cases since this abnormality was first discovered in 1922. Pregnant patients with bladder exstrophy may have a higher risk of obstetrical complications, such as preeclampsia, abortion, and premature delivery. Distortion of pelvic anatomy presenting with such patients may cause much more difficult delivery (by cesarean section) through possible multiple adhesions showing in the abdomen associated with the malformation and prior surgery [2]. We gave our experience with an adult pregnant patient with a history of bladder exstrophy.

Case Report

A 21-year-old woman presented with a history of EEC. During childhood, she had repeated inadequate management of bladder closure attempts, which resulted in an empty contracted bladder (Figure 1). She also had a uterine prolapse due to uncorrected pubic diastasis. Because of concern of small and fibrotic bladder, the potency of spite, and the inability of the patient to control routinely, it was decided to do cystectomy and substitute by reconstruction of serous lined ileal W pouch continent cutaneous stoma as described by Abol-Enein and Ghoneim [3]. Uterine prolapse was corrected by sacrocolpopexy that the posterior vaginal apex fixed to promontories using polypropylene mesh. All of these procedures were performed in Ciplo Mangunkusumo

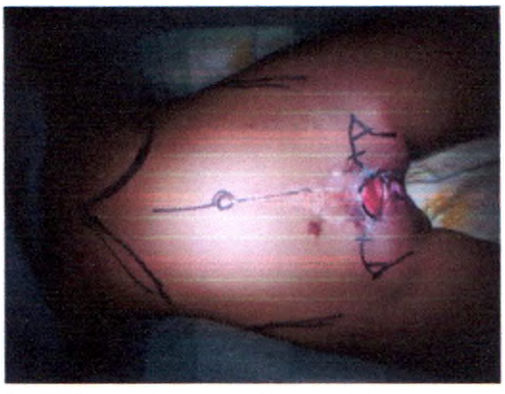

Figure 1 The clinical appearance of the patient before reconstruction was done. The scars at the lower midline abdomen due to multiple prior procedures during childhood.

Hospital, Jakarta. A long-distance pubic diastasis (15cm) was not corrected because bony fusion had occurred.

After reconstruction, the patient got completely dry and improved her personal and social life. The patient had not reported any other complaint other than a bulging pouch when it was completed. She got married and got pregnant 7 years later.

The patient was visited for antenatal evaluation at M Djamil Hospital, Padang. The patient's vital signs were within normal limits (Figure 2a). Creatinine serum was normal with slight right hydronephrosis was found from the ultrasound. There were no symptomatic urinary tract infections (UTI) and difficulties emptying the pouch through the stoma channel with a 16 Fr catheter. On obstetric examination at 34–35 weeks of gestational age (Figure 2b), it was revealed that the singleton fetus was in a transverse lie in which the vertex was located on the right dorso-superior side (Figure 2c). At the lower midline of the abdomen, suprapubic scars from previous surgeries and third-grade uterine prolapse were visible.

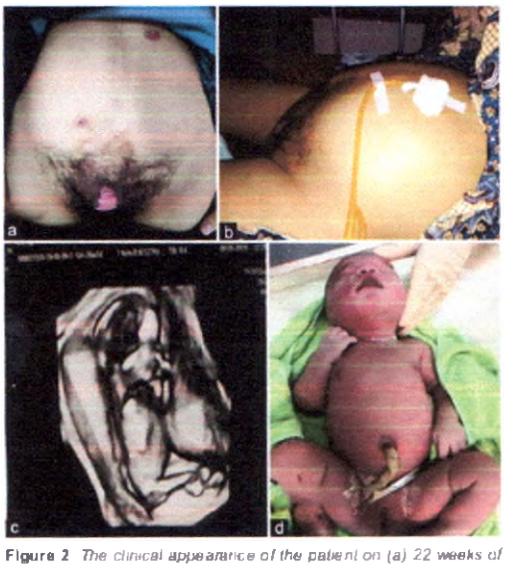

Figure 2 *The clinical appearance of the patient on (a) 22 weeks of gestational age and (b) 35 weeks of gestational age and her baby on (c) ultrasound at 35 weeks of gestational age and (d) after delivery.*

An elective cesarian section was performed on 38–39 weeks of gestational age. The incision was made in the lower midline, and the female infant was delivered with 2,600 g in weight and 48 cm in length (Figure 2d).

Discussion

Most bladder exstrophy is diagnosed during the neonatal period: only approximately 13% were diagnosed prenatally. The bladder plate may protrude just beneath the umbilical cord with a divergent rectus muscle on either side of the bladder, leading to the separated pubic bones. Risk factors for the development of bladder exstrophy are yet unclear. Possible genetic links may be found, the findings, however, were limited only to case reports [4].

Due to a relatively higher risk of inguinal hernia and urinary continence issues in infants with bladder exstrophy, early repair surgery is typically warranted. Bladder closure may be achieved by initially closing the pelvic ring. If closure was not completed within the first 72 h of life, an osteotomy might be required to complete the pubic bones. Bladder neck reconstruction is required by age 4–5 when the child is old enough to cooperate with toilet training [5]. Bladder exstrophy may be associated with other congenital disabilities, such as abdominal wall defects [6]. In turn, abdominal wall defects and subsequent weakening of abdominal muscles and pelvic ligaments may increase the risk of pelvic organ prolapse (POP); approximately 18%–30% of women were diagnosed with POP [7]. In our case, the bladder exstrophy had been treated (with the previous history of surgery at age 3, 4, and 14) inadequately; this resulted in shrinkage of the bladder and prolapse of the uterus, which requires reconstruction of continent cutaneous stoma and sacrocolpopexy during adulthood. This correction made the patient gain completed continence through self-catheterization.

Pregnant patients with previous bladder exstrophy are at higher risk of obstetrical complications. UTI is the most common complication in such patients. In our case, no symptomatic UTI related to obstetrics was present during pregnancy. This could be a result of regular self-catheterization. The patient had no self-catheterization difficulty even in the third semester of the pregnancy because of the high location of the stoma outlet at the left-hand side of the upper abdomen.

The construction of the serous line technique as an anti-reflux mechanism as described by Abol-Enein and Ghoneim [3] was made in this case, and it could contribute to protection from symptomatic ascending UTI and maintain kidney function, which is also mentioned in the study by Mensah *et al.* [8].

One of the obstetrical complications associated with vaginal or cesarean section delivery is uterine prolapse [9], [10]. Sathishkumar *et al.* had noted in a case report that sacrocolpopexy with posterior fixation of the vaginal apex and anterior fixation of the cervical-uterine unit might be utilized in the management of prolapse in a woman with classical bladder exstrophy [11]. In our case, sacrocolpopexy using a polypropylene mesh that fixed the vaginal apex posteriorly to promontories was performed concomitantly during the reconstruction of the continent cutaneous stoma. This fixation effectively supported the uterus to retain the fetus to reach a full-term period. However, pelvic osteotomy was not performed in this case, and a long gap in pubic diastasis was not corrected. Consequently, any degree of POP would still potentially reoccur.

A study by Dy *et al.* had noted that 14 out of 20 patients with bladder exstrophy (70%) had successful deliveries with live births, the rest of the patients, however, had their pregnancy terminated or had experienced spontaneous abortion [12]. Management of the delivery on patients with bladder exstrophy may depend on the state of the birth canal. Although vaginal delivery may be chosen and may bring relatively favorable outcomes on both the mother and the infant, there is a subset of patients who had associated birth canal defects or malpresentation that requires cesarean section.

A multidisciplinary team is required to manage of pregnant patients with bladder exstrophy. Although the ultimate choice of delivery shall be decided by the obstetrics department, urologists should be involved in the discussion. In our case, the option to perform a cesarean section was based on the consideration that the fetus was in a non-vertex position, which made a vaginal delivery difficult; furthermore, uterine contraction and vaginal delivery would provoke POP. Nevertheless, a meticulous incision and prompt dissection should be done to anticipate multiple adhesion resulting from prior surgeries and to hinder the injury of the ileal pouch. Since the position of the ileal pocket in this type of continent stoma was relatively high and not fixed to the pelvic floor or any organ in the extraperitoneal area, a lower midline incision was preferable as it was considered safe.

Management of bladder exstrophy in recent times has been improved to the point that continence and the satisfactory functional outcome may be achieved [13]. Beyond the concern about urinary continence and normal kidney function, other personal (including sexual life and maternity) and social processes can be improved by multidisciplinary management and seamless transition from childhood to adulthood care.

Conclusions

An adult female with bladder exstrophy underwent pregnancy and gave birth. Prior surgery and increased risk of obstetrical complications must be considered. The choice of delivery, either vaginal or cesarean section, is feasible for delivery. The involvement of urologists in a collaborative team was deemed necessary to give the best management in this complicated case. Further research series on bladder exstrophy should involve multicenter urology education in Indonesia.

References

1 Massanyi EZ, Gearhart JP, Kost-Byerly S. Perioperative management of classic bladder exstrophy. Res Rep Urol. 2013;5:67-75. https://doi.org/10.2147/rru.s29087
PMid:24400236

2 Amin KV, Tiwari N, Goel A, Chauhan AR. Pregnancy in case of exstrophy of the bladder. Int J Reprod Contracept Obstet Gynecol. 2017;6:1676-8. https://doi.org/10.18203/2320-1770.ijrcog20171453

3 Abol-Enein H, Ghoneim MA. Serous lined extramural ileal valve: A new continent urinary outlet. J Urol. 1999;161(3):786-91. https://doi.org/10.1016/s0022-5347(01)61769-5
PMid:10022685

4 Messelink EJ, Aronson DC, Knuist M, Heij HA, Vos A. Four cases of bladder exstrophy in two families. J Med Genet. 1994;31(6):490-2. https://doi.org/10.1136/jmg.31.6.490
PMid:8071977

5 Ben-Chaim J, Docimo SG, Jeffs RD, Gearhart JP. Bladder exstrophy from childhood into adult life. J R Soc Med. 1996;89(1):39P-46P.
PMid:8709084

6 Orioli IM, Amar E, Arteaga-Vazquez J, Bianca S, Botto LD, Clementi M, *et al.* Sirenomelia: An epidemiologic study in a large dataset from the international clearinghouse of birth defects surveillance and research, and literature review. Am J Med Genet C Semin Med Genet. 2011;157C(4):358-73. https://doi.org/10.1002/ajmg.c.30324
PMid:22002878

7 Pacquee S, Conrad DH, Saar TD, Rosen D, Cano G, Chou D, *et al.* Surgical management of complete procidentia in a female patient with bladder exstrophy-epispadias complex: Case report and literature review. Gynecol Minim Invasive Ther. 2018;7(3):127-9. https://doi.org/10.4103/gmit.gmit_14_18
PMid:30254955

8 Mensah JE, Asante EK, Mensah WA, Glover-Addy H. Continent cutaneous diversion for bladder exstrophy in adults. Afr J Urol. 2013;19(2):94-8. https://doi.org/10.1016/j.afju.2013.01.002

9 Wu S, Sun J, Lv H, Zhang Y, Shang H, Zhang H, *et al.* Pregnancy in a woman with untreated bladder exstrophy: A case report. Case Case Rep Womens Health. 2018;17:11-3. https://doi.org/10.1016/j.crwh.2018.02.002
PMid:29594008

10 Liu S, Qu X, Song L, Li N, Xu A. Repeated successful vaginal delivery in a pregnant woman with unrepaired ectopia vesicae

and split pelvis: A case study. BMC Pregnancy Childbirth. 2020;20(1):290. https://doi.org/10.1186/s12884-020-02931-x
PMid:32397972

11. Raamya SM, Mavuduru RS, Agarwal MM. Is sacrocolpopexy adequate for cervico-uterine prolapse in patients with classical bladder exstrophy? J Obstet Gynaecol India. 2014;64(Suppl 1):83-4. https://doi.org/10.1007/s13224-012-0321-y
PMid:25404822

12. Dy GW, Willihnganz-Lawson KH, Shnorhavorian M, Delaney SS, Ames Oelschlager AM, Merguerian PA, *et al.* Successful pregnancy in patients with exstrophy-epispadias complex: A University of Washington experience. J Pediatr Urol. 2015;11(4):213.e1-6. https://doi.org/10.1016/j.jpurol.2015.04.019
PMid:26092092

13. Alsowayan O, Capolicchio JP, Jednak R, El-Sherbiny M. Long-term functional outcomes after bladder exstrophy repair: A single, low-volume centre experience. Can Urol Assoc J. 2016;10(3-4):E94-8. https://doi.org/10.5489/cuaj.3104
PMid:27330586

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