

Pregnancy in a woman after eight reconstructive urological operations due to bladder extrophy: case report

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Summary

The progress of reconstructive surgical treatment of congenital malformations is associated with an increasing number of patients requiring special care in pregnancy. Although there have been some communications concerning the management of pregnancy after one or two reconstructions of bladder extrophy, a case with a successful course of pregnancy after eight urological operations of this disorder is presented.

Key words: Pregnancy complications; Bladder extrophy.

Introduction

The progress of reconstructive surgical treatment of congenital malformations is associated with an increasing number of patients requiring special care in pregnancy. Although there have been some communications concerning the management of pregnancy after one or two reconstructions of bladder extrophy, a case with a successful course of pregnancy after eight urological operations of this disorder is presented.

Case Report

AA, age 23, was born with bladder extrophy. In the second week of life the remnant bladder was very small, about 5 cm square with hypertrophic mucosae. At the age of 18 months, a rectosigmoid bladder procedure using the Duhamel technique was performed (2 procedures) [1]. In the next, step she underwent extrophic bladder excision and abdominal wall reconstruction. However after six months an ileal conduit urinary diversion was formed due to bilateral obstructed megaureters and hydronephrosis. After improvement of the previously obstructed upper urinary tract and normalization of kidney function, a Mainz I pouch was constructed. The ileal conduit was incorporated into this pouch [2]. Intermittent self-catheterization was necessary and trimetoprim or nitrofurantoin was introduced as prophylactic medication. Dynamic renography was normal and no other deterioration in renal function was detected. At 17, scarred and nonhairy skin from the midline mons pubis was excised. The flaps of hair-bearing skin, including all the subcutaneous fat, were mobilized and rotated to fill the defect. At the same session correction of the labia and introitus appearance was performed. The two halves of the clitoris were to unite and the anterior ends of the labia were to fuse to make a fourchette. Next, episiotomy was performed due to a very narrow introitus and the defect was covered using perineal, full thickness skin. At the age of 19, she required suspension due to uterine and vaginal prolapse. Uterine fixation was performed to the anterior abdominal wall. The following year, she

became sexually active and reported having orgasms and a satisfactory sexual life.

Prior to pregnancy renal function was diagnosed by scintigraphy [left kidney marker (Tc99m-GL) excretion was about 50% worse than the right one]. There was no deterioration in renal function as a consequence of the pregnancy, as assessed by serum urea, creatinine levels and clearance of creatinine. During the second trimester she received furagin as a prophylactic for urinary tract infection and later we found asymptomatic bacteriuria (*E. Coli* 10⁵/ml), which required targeted antibiotic therapy. From the beginning of the pregnancy mild dilatation of the urinary tract was detected, which resolved after delivery. Ultrasound examination and biochemical [PAPP-A (*pregnancy associated plasma protein-A*), AFP (*α-fetoprotein*), β-HCG (*β-human choriongonadotropin*) and others routinely performed in pregnancy] tests revealed no abnormalities. Pregnancy was uneventful until 33 weeks of gestation when she was treated up to 35 weeks, because of premature labor (0.5 mg/min fenoterol (Pliva Kraków) in 500 ml 0.9% sterile saline solution and subsequently oral fenoterol 4 x 5 mg daily). A routine ultrasound examination revealed normal intrauterine growth (approximately the 50th percentile of the normal growth curve), AFI (*amniotic fluid index*), normal umbilical and middle cerebral artery blood flow (CPR - *cerebroplacental ratio* at 28 weeks was 1.7, and at 30 was 2.04) and breech presentation. At 33 weeks uterine prolapse was noted, but it did not impact on the course of pregnancy, which was terminated at 37 weeks. Elective cesarean section was performed because of breech presentation, scars after reconstruction of mons pubis, vestibulum of the vagina and previous successful surgical repair of the prolapsed uterus. A normal female, weighing 3,130 g (Apgar score: 9, 10) was delivered, and both mother and daughter, after an uneventful postpartum course, were discharged from hospital in very good condition. Examination at six weeks and one year after delivery showed no complications in either mother or baby.

Discussion

Congenital bladder extrophy affects one in 125,000 to 250,000 females and frequently is associated with genital prolapse and recurrent renal infections. The goals of extrophy reconstruction are anatomical closure of the

bladder and abdominal wall, preservation of renal function as well as a functional and cosmetically satisfactory external genitalia. After failed reconstruction when the extrophic bladder is very small, continent urinary diversion is carried out [3]. A few centers treat bladder extrophy primarily by internal urinary diversion into the sigmoid colon [4]. There is a tendency to reconstruct the genitalia in infancy or childhood. Stein *et al.* suggest opening of the introitus and uniting the labia at three to four years of age. This allows easy drainage of secretions and, later menstrual flow [5]. Hohenfellner and colleagues advocate fixation of the uterus to the anterior abdominal wall in childhood, which is said to prevent prolapse but still allow future normal pregnancy [2]. Other authors prefer fixation to the sacral promontory using the Gore-Tex graft [6, 7].

Today surgical treatment of bladder extrophy with neonatal primary closure or osteotomies gives more promising functional and cosmetic results [8]. However, delivery and its effect on renal function as well as the status of the lower urogenital tract is still the challenging problem. Moreover, pregnancy could be complicated by recurrent urinary tract infections, preterm labor, mild proclivencia, and malpresentation [7, 8, 12]. The first report of pregnancy occurring in a woman with bladder extrophy was made by Bonet in 1724 [12]. After a period of casuistic information Clemetson [10] in 1958 on the basis of an original case report and review of the literature (64 pregnancies in 45 women) found that recurrent ascendant pyelonephritis, premature labor, malpresentation, vaginal stenosis and uterine prolapse were major problems. In patients with bladder extrophy Kennedy *et al.* [11] have reported four cases of successful pregnancies in four young adult females who had previously undergone a flap vaginoplasty as part of earlier management, and more recently a continent right colonic urinary reservoir with a perineal stoma (Indiana pouch). Pregnancy in each of these patients was characterized by several urinary tract infections, cervical prolapse, and maternal hydronephrosis, which resolved after delivery. All pregnancies were delivered by cesarean section, and infants were healthy and in good condition. The influence of pregnancy on uterine prolapse is still one of the most important problems in these patients. Rose *et al.* [7] reported the case of a young woman with surgically repaired bladder extrophy who developed genital prolapse. The uterus was suspended using a sacral colpopexy utilizing a Gore-Tex graft. Subsequently, the patient became pregnant and delivered a healthy male infant at 35 weeks' gestation by cesarean section (without recurrence of the genital prolapse postpartum). Mathews and co-workers [9] presented their 15 years experience in 83 women with extrophy-epispadias complex and eight of them had 13 pregnancies. High incidence rates of abortion and miscarriage (5 cases) were noted, and finally five women delivered eight normal healthy children: seven by cesarean section and one – uncomplicated vaginally.

The mode of delivery is still discussed. Clemetson in 1958 [10] suggested that cesarean section was necessary

to prevent prolapse developing later. Lately many physicians prefer to use cesarean section to avoid complications, especially in those patients who have had a successful functional reconstruction [14]. A high rate of breech presentation (57% vs 4% in the general population) [15] as well as other relative obstetric indications (previous cesarean section) might be the case for this preferable mode of delivery.

Greenwell *et al.* [15] concluded that pregnancy has no long-term effect on renal function and does not compromise reconstruction. Although there is a substantial complication rate and increased need for cesarean section, pregnancy in women with lower urinary tract reconstruction for congenital urological abnormalities is ultimately safe for both mother and baby. Interdisciplinary cooperation is desirable for a successful outcome.

In summary, based on a review of the literature and the follow-up of the described case, we can conclude that pregnancy after multiple reconstructive urological operations due to bladder extrophy can proceed safely. The obligatory condition for a successful course and outcome of pregnancy is tight monitoring of both mother and fetus. The crucial point in these pregnancies is to regularly estimate renal function (serum urea and creatinine levels or clearance of creatinine) as well as ultrasound examination of dilatation of the urinary tract, which may be a marker of obstruction and could be related to urinary tract infection. As a prophylaxis of urinary tract infection antimicrobial therapy should be considered. Fetal development should also be assessed using mainly biophysical methods. The decision on the time and mode of delivery depends on the condition of the mother and fetus, and should be made individually.

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