CASE REPORT

Pregnancy in a woman with treated bladder extrophy, split pelvis and hypoplasia of ischial bones. Case Report.

Mariusz Zimmer[1], Kireki Omanwa[1],Wojciech Kolaczyk[2], Ewa Milnerowicz-Nabzdyk[1], Michal Pomorski[1], Artur Klosek[1], and Joanna Tlolka[1]

1. Department of Gynecology Obstetrics and Neonatology, Wroclaw Medical University;
2. Department of Urology, Specialist Hospital Wroclaw; Poland.

Correspondence to: Assoc. Professor Mariusz Zimmer, MD, PhD
Department of Gynecology, Obstetrics and Neonatology, Wroclaw Medical University
ul. Dyrekcyjna No. 5/7, 50-528 Wroclaw, POLAND
PHONE: + 48717331400, + 48665866660; FAX: + 48717331409
E-MAIL: mzimmer@op.pl

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Abstract

We present a case of a pregnancy in a 24 year old woman who was born with ectopia vesicae, split pelvis and hypoplasia of ischial bones. From childhood to adulthood she had undergone reconstructive surgeries of the abdomen and perineum, as well as urine diversion surgery.
During pregnancy she experienced recurrent urinary tract infections which were treated with antibiotics. In spite of tocolysis, she delivered by cesarean section a premature baby boy at 35th week of gestation due to premature uterine contractions. The postoperative period was uneventful and they were discharged from the hospital in a good general condition.

Abbreviations:
CTG - Cardiotocography
UTI - Urinary tract infection

INTRODUCTION

Bladder extrophy is usually associated with other congenital anomalies due to the fact that the embryogenesis of the urinary tract and genitalia is closely connected. These anomalies may include the absence of the uterus, a malformed uterus, vaginal septum, a fistula or even the possibility of renal agenesis (Mantel *et al*. 2001). Most of these anomalies can be corrected by surgery with urinary reconstruction carried out before or during the reproductive years.

Pregnancy in women after urinary reconstruction is rare and should be considered as high risk. These pregnancies are often complicated by: miscarriages, premature births, fetal malpresentation and frequent urinary tract infections, which must be treated with antibiotics. Another common complication is pyelonephritis. Although rare, ileal prolapse and secondary urinary incontinence may further complicate these cases and should be borne in mind (Mantel *et al*. 2001).

CASE

A 24 year old primigravida was admitted to our department at 33 weeks of gestation, based on the last monthly period, due to intermittent lower abdominal pains which had started a few days before.

Her medical history revealed that she had been born with a bladder extrophy (III degree according to Champneys), a split pelvis, as well as hypoplasia of the ischial bones and had undergone several reconstructive surgeries in the past.

At 17 months of life she underwent a surgery and had an ileal loop conduit as well as cutaneous ureterostomy formation, as described by Bricker.

At the age of three she had a reconstructive surgery of the lower abdomen where remnants of the bladder mucosa were excised and the abdominal wall defect was closed. At the age of five a myorraphy of the perineal muscle was performed.

At eighteen a hymenectomy and a widening of the vaginal introitus were done. At nineteen, because of subsequent incontinence of the urinary diversion, exploration was conducted and a partial resection of the ileal loop and a reimplantation of the ureters were carried out. Because of chronic pyelonephritis and recurrent urinary tract infections the antibiotic therapy was initiated. Serum creatinine and urea concentrations were within normal range. Serial ultrasound scans showed only mild bilateral hydronephrosis. On admission her pulse rate, blood pressure, and body temperature were within normal range, her BMI 24.2.

Until hospital admission, the course of pregnancy was generally uneventful, apart from the outpatient treatment of urinary tract infections (Amoxicillin/clavulanic acid and furazidin). The standard fetal ultrasonographic assessment had been carried out (Czuba *et al*. 2007).

On abdominal examination fundal height was consistent with 33 weeks of gestation and vertex presentation. A speculum examination revealed a cervical length of about 2.5cm; the external cervical os closed. A CTG trace was found to be normal.

A transabdominal ultrasound scan revealed a viable fetus with biometry consistent with 33 weeks of gestation and estimated fetal weight of 2370 g, with no visible morphological anomalies. The amniotic fluid index was 10. A transvaginal ultrasound revealed the cervical length of 29 mm and a closed internal cervical os. Serial ultrasound scans performed during pregnancy showed no increase in bilateral hydronephrosis. The position of the pouch in relation to the placenta and intestinal tract was easily visualized by ultrasound.

Renal function was stable with urea nitrogen of 2.9 mmol/l and serum creatinine of 69 mmol/l. Other routine laboratory results were within normal range.

A urinary culture and sensitivity test showed a urinary tract infection with Escherichia coli and Klebsiella pneumoniae. Amoxicillin/clavulanic acid was administered intravenously, 3 × 1.2 g daily.

Due to intermittent uterine contractions, accompanied by lower abdominal pain, the patient was administered oral tocolysis with beta2-agonists.

A follow-up transabdominal ultrasound was carried out every 7 days, while CTG traces daily, UTI treatment was continued and intravenous tocolysis (beta2-agonists and magnesium sulphate) to address premature uterine contractions was initiated several times. In spite of the above, at 35 weeks of gestation regular prolonged uterine contractions were observed. After a careful case analysis and consultation with a urologist, an emergency cesarean section was performed.

Laparatomy was undertaken through a long midline incision. The dissection was fairly easy, no adhesions in the abdominal cavity were noticed. The uterus was opened by a low transverse section. A newborn premature baby boy weighing 2880 g with no obvious abnormalities was delivered with Apgar scores of 8 and 10 at 1st and 5th minute of life.

The uterine scar was closed by a single continuous suture. The consulting urologist present during surgery in order to check the ileal loop conduit found it to be continent. No complications were encountered during and after the surgery. Blood loss was estimated to be about 400 ml. The course of the puerperium was complicated by an infection of the urinary tract with Klebsiella pneumoniae.Cefuroxime sodium was administered intravenously 1.5 g every 12 hours. On the 23rd postoperative day, the mother and the child were both discharged in good condition.

DISCUSSION

Bladder extrophy is a rare and most distressing malformation, in which the anterior bladder wall is absent and the posterior wall protrudes as a pink tender fleshy mass in the lower abdomen. As a result of this urine flows away from the ureteric orifices, making the sufferer constantly wet. It occurs in 2-3.3/100,000 deliveries with a male to female ratio of 3-4 to 1 (Mantel *et al*. 2001). It is often associated with the anomalies of other organs, for example: the genitalia, urinary tract and bones. This deformity has to be treated in childhood and a subsequent follow – up is necessary. A pregnancy in a woman with urinary diversion treatment is a rare event (Volkmer *et al*. 2002).

Champneys describes the following four degrees of bladder extrophy:

I -The lowest or slightest degree of deformity tending to extroversion, where the pubic bones are separated but there is no fissure in the abdominal wall; the weak linea alba forms a hernial pouch containing the bladder .

II -The bladder is perfect but protrudes through a fissure in the abdominal wall.

III -The full deformity, where both the bladder and the abdominal walls are cleft.

IV -In the highest or greatest degree of deformity the extroverted bladder is separated into two halves by an opening of the intestine.

Even in the first degree ectopia vesicae the pubic symphysis is split and the pubic bones are often widely separated: a fibrous inter pubic band or ligament often replaces the missing bone and unites the pubic rami: the lower ends of the rectus abdominis are divaricated and inserted into the separated bones, thereby forming a

triangular window enclosing the malformed bladder (Clemetson, 1958).

Although women with bladder extrophy are able to conceive, many other reasons make it difficult for them to become pregnant. They suffer from the appearance of their external genitalia (deformed pubis with an unusual pilosity), their multiple abdominal scars and often urinary incontinence (Mantel *et al*. 2001).

The first record of pregnancy in a woman with ectopia vesicae was reported by Mr John Bonnet in the transactions of the Royal Society, 1724. He successfully delivered her infant by dividing the perineum between the forchette and the anus (Clemetson, 1958).

Women after bladder extrophy treatment are prone to complications during pregnancy and delivery. The most common complication during pregnancy is recurrent UTI. Pyelonephritis usually occurs in the second trimester and can be a cause of premature labor (Kennedy II *et al*. 1993). Regular follow-up of urine sterility, renal function, as well as maternal renal ultrasound examination is necessary (Mantel *et al*. 2001). In case of premature uterine contractions the use of tocolysis together with antibiotic therapy should be considered in order to maximize its effect (Bobrzynska & Reron, 2004). All the published cases of pregnancy in women with a history of bladder extrophy report a high frequency of premature delivery ranging from 20.9- 40 % versus 7-11% in the general population (Kennedy II *et al*. 1993; Monga & Creasy, 1995; Bibby & Stewart, 2004; Carson, 2004).

Many causes can lead to a premature delivery, including uterine malformations, frequent urinary infections that easily worsens to a pyelonephritis with hydronephrosis. A failure of the supporting structures of the genital tract and uterine prolapse are another possible explanations (Mantel *et al*. 2001).

Malpresentation of the fetus is quite common, partly because of the frequency of associated uterine anomalies but also because the widely split pelvis ails while trying to control the presenting part, especially when the fetus is small and premature, which is often the case. Clemetson (1958) reported that fetal malpresentation concerns 25% of the pregnancies.

Uterine prolapse is another major problem in these women. This is due to several factors, such as the large transverse pelvic diameter, low and weakened pelvic floor, the short anterior vaginal wall and the weakness of the cardinal ligaments supporting the uterus. Most authors only take this complication into consideration at the time of pregnancy or during post partum (Mantel *et al*. 2001).

Ileal prolapse is a possible complication in women with an external urinary diversion. Even though it is a rare complication one has to be aware of it (Krisiloff *et al*. 1978).

Urinary incontinence, subsequent to pregnancy and delivery, after bladder extrophy treatment is a serious complication. Several cases occurring at different stages have been described in literature (Mariona & Evans, 1982; Burbridghe *et al*. 1986). It is difficult to estimate the incidence of this complication because the reported cases do not always clearly mention how continent their patients were.

A literature review reveals a controversy as to the method of delivery (cesarean section versus vaginal delivery) in such women. Several observations have proven that a cesarean section does not always help to avoid the prolapse, when vaginal delivery is rather easy due to pelvic anatomy conditions (Krisiloff *et al*. 1978).

In our opinion it is better to avoid perineal lesions in these patients who have undergone reconstructive surgery, and whose ano - vulva distance is short. Obstetric indications as well as the rate of fetal malpresentation should be taken into account.

Furthermore, the obstetrician must be aware of the possible urological complications, especially concerning the risk of subsequent incontinence. Preserving continence, which in most cases has been difficult to achieve, is of primary importance. That is why a planned cesarean section appears to be the best solution in the majority of cases, particularly for women who have undergone bladder reconstruction or diversion with a continent sphincter (Mantel *et al*. 2001).

Planning a cesarean section before the onset of labor may help to avoid the necessity of operating on women with a history of multiple abdominal operations in urgent and difficult conditions.

Our experience is in agreement with other authors such as Mariona and Evans (1982). We suggest that a urologist familiar with various forms of urinary diversion should be present at the time of surgery.

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