Abstract
Cloacal malformation It is extremely rare, occurring in approximately 1 in 25,000 births. We describe here a 19 years old patient who conceived naturally with cloacal malformation after surgery of cloaca and proctoplastica (propter atresia ani et recti), after surgery of vaginal septum with agenesis of right tube and ovary, agenesis of coccygeus, dysplasia sacri, aneurysma septi interatrialis, hydronephrosis, chronical pyelonephritis, fecal incontinence. In childhood patient underwent many reconstructive surgeries. During pregnancy phlebothrombosis of v. femoralis communis, v. femoris profundi and v. iliaca externa occurred. A birth plan was discussed and approved together with paediatric surgeons and urologists. At 38 weeks of pregnancy, the patient was urgently hospitalized and underwent Caesarean section with cooperation with urologists. Newborn was delivered by cephalic presentation with birth weight of 2720 g and with an Apgar score of 9 and 9.

Introduction
Cloacal malformation is an anatomic defect occurring exclusively in females. It arises during the fifth to eight weeks of fetal development as a failure of the urorectal septum to form and divide the cloaca into rectum dorsally and urogenital sinus ventrally. This results in confluence of the rectum, vagina, and urethra into a single common channel. It is extremely rare, occurring in approximately 1 in 25,000 births. The etiology of persistent cloaca is unknown [1].

Persistent urogenital sinus (PUGS) is a pathological condition characterized by an abnormal communication between the urethra and vagina. It may be a part of a complex syndrome and can be more often associated with congenital malformations affecting the genitourinary tract system (33%) such as intersex, rectovaginal communication, bladder agenesis, absence of vagina, and hydrocolpos [2].

Cloacal malformation frequently has associated Mullerian anomalies with high incidence of gynecological problems that require surgical correction, such as hematometra due to cervical stenosis or vaginal stenosis, vaginal septum, and endometriosis [3].

20% of those patients suffering urinary incontinence and 40% fecal incontinence, sometimes-sexual dysfunction. About 80% of patients with cloacal malformation are born with a structural abnormality of the urinary tract, including renal dysplasia, ectopic kidney, solitary or duplex kidney, or ureteropelvic junction obstruction [4] and sacrum and spine anomalies [5].

We describe here a 19 years old patient who conceived naturally with cloacal malformation after surgery of cloaca and proctoplastica (propter atresia ani et recti), after surgery of vaginal septum with agenesis of right tube and ovary, agenesis of coccygeus, dysplasia sacri, aneurysma septi interatrialis, hydronephrosis, chronical pyelonephritis, fecal incontinence. During pregnancy phlebothrombosis of v. femoris communis, v. femoris profundi and v. iliaca externa occurred.
The aim of Pen’s surgery method – to draw the end of the large intestine to the abdominal wall and to form the anus with normal anal sphincter function [6].

At the age of 2 years OLD (22/09/1999), the patient was urgently hospitalized due to progressive bilateral hydronephrosis to the Pediatric Surgery. Cystogram showed grades IV-V right unilateral vesicoureteral reflux (VUR), hydroureter, and left nephroplasia due to the stricture at the intramural portion of the ureter and the pressure of constantly full bladder. The decision was made to perform vesiocystostomy as an alternative to epispadiostomy, however, after consultation with urologists from the USA and failure to perform cystoscopy, vesiocystostomy was postponed.

Following 3 months, cystoscopy was successfully performed, and the diagnosis of urogenital sinus was made. The patient underwent vesiocystostomy. Since the girl showed persistent dysfunction of the right kidney and bilateral nephroplasia, in July, 2003 (at the age of 6 years), ureteroileocutaneostomy (Bricker conduit) in the right lower quadrant was performed and vesiocystostomy closed. The aim of surgery – to form urinary reservoir using a portion of intestine. The urinary reservoir collects urine from both ureters and drains it via stoma created in the abdominal wall [7].

After her first menstruations, at age 12, the girl was hospitalized due to persistent urogenital sinus: two orifices of the vagina opened at the cervix of the urinary bladder. The patient again underwent surgery, during which urogenital sinus was mobilized, double vaginas disconnected, and after forming one channel, the distal vaginal portion was formed using patches from vagina tissue and urogenital sinus, and the urethra was tubularized. The vagina was dilated with Hegar dilator of size 16Ch and after 3 months to size 20Ch.

After 5 months following the last operation, the patient had augmentation cystoplasty using a segment of ileum; the earlier ureteroileocutaneostomy was removed due to nutritional disorder, and a new ureterovesicocystostomy formed. Also, continent appendicovesicostomy was formed, and the girl was taught to use catheter via stoma every 2-4 hours using 12 Fr Nelaton catheter.

At the age of 15, following 3 years of vaginoplasty and the urethra formation, the patient was taught to use catheter via stoma due to insufficient drainage of the bladder and hydronephrosis progression.

The patient at 34 weeks of pregnancy presented to the Obstetrics department of the (HLUHS) Kaunas Clinics. Blood tests (full blood count (FBC), electrolytes, kidney function) were normal, and vesicovaginal fistulae were apparent. The abdomen was distended, double vaginas divided using a speculum and dully pushing away the formed urethra, and the rectum was proctoscoped. The incision length was 3-4 cm superior the umbilicus and 7 cm superior the pubic bone.

The abdominal cavity was incised by longitudinal section by passing the umbilicus from the left side. The incision length was 3-4 cm superior the umbilicus and 7 cm superior the pubic bone.

The augmented bladder was filled by methylene blue using Nelaton type catheter.

Adhesions between the uterus, peritoneum and omentum were divided using a speculum and dully pushing away the formed bladder to the left (Figure 3). A transverse uterine incision was made and female newborn was delivered by cephalic presentation with birth weight of 2720 g and with an Apgar score of 9 and 9.

The examination during surgery revealed absent of left adnexa. Nelaton catheter inserted via stoma was left in the augmented bladder after surgery. Also, two drains remained in the pouch of Douglas and in the vesicouterine excavation. Postoperative urine culture detected E.coli resistant to ampicillin, cefturoxime and nitrofurantoin. Cefuroxime was replaced by sulbactam 1.5 g 4 times per day.

During treatment, the primary healing of incision was observed, the patient was not febrile, a repeat urine test showed no changes, urine culture – sterile. The newborn was breast fed. On the 10 day the patient was discharged with a healthy newborn from the hospital home.


Discussion

Given the limited information on the preferred mode of delivery of patients with PUGS repair. There have been several case reports of vaginal deliveries in these patients [8,9].

At 38 weeks of pregnancy, with the persistence of diarrhea for two days, the patient was urgently hospitalized to the Obstetrics department of (HLUHS) Kaunas Clinics. Acute pyelonephritis was diagnosed and urine culture showed a significant level of E.coli. Coagulogram showed a mild hypercoagulation. HIV and syphilis were negative.

The patient complained of increasing backache and she did not experience uterine contractions (UC). Cardiotocography (CTG) showed UC are every 3-7 minutes. After 2 hours, when the patient started experiencing regular UC 3 times per 10 minutes, the decision was made to start CS earlier with participation of urologist using endotraheal anaesthesia.

Before operation, the bladder was filled with fluid by Foley catheter and examined using ultrasound: the urine bladder was detected in the left lower quadrant (Figure 1, 2).

Delivery After Cloaca Malformation Surgery.
Figure 1: Ultrasound before operation.

Figure 2: Patient before s/c operation. On the right side-appendicovesicostoma.

Figure 3: The beginning of operation-adhesiolysis. Žemiau ir on the left –gimda.Dešinėje- iš žarnos suformuotas šlapimo rezervuaras.
The patients who conceive after repairs invariably have had cesarean deliveries on the assumption that vaginal delivery would be difficult and/or dangerous. However, because they usually have undergone multiple previous abdominal procedures, CS delivery also may carry greater risks than in the general population [10].

Alon Shrim, Tiina Podymow et al. from Canada described a patient with cloacal malformation, solitary kidney, bilateral fallopian tube obstruction, and didelphic uterus who required in vitro fertilization to conceive and careful surveillance resulted in an excellent pregnancy outcome with term delivery. Careful surveillance during pregnancy and a planned abdominal delivery resulted in a successful outcome [11].

The patients with cloacal malformations must give birth at third level university hospitals, because they need multidisciplinary team for assistance.

References