Case Report

Bladder Mucosal Graft Vaginoplasty: A Case Report

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A B S T R A C T

Background: Female vaginoplasty reconstruction, by choice, is usually performed with adjacent tissue. However in some clinical conditions such as high urogenital confluence sinus, cloacal malformation with extreme vaginal hypoplasia, local tissue may not be available. When vaginal replacement is performed in pediatric patients intestinal segments is preferred to non-operative procedures that require continuative dilations. However mucus production, malignant transformation risk and diversion colitis are important side effects.
Technique: We present a nouvel technique for vaginoplasty in a female child presenting with an isolated urogenital sinus malformation without virilization. The patient at 20 months underwent vaginoplasty using tubularized bladder mucosal graft.
Results: Surgical procedure was devoid of complications. Pubertal development occurred at age of 15. She underwent regular follow up until 18 years of age. At this age we performed clinical evaluation: absence of vaginal introitus stenosis and good cosmetic results were observed. Then she underwent vaginoscopy with multiple biopsies. Pathology examination of the bladder mucosal graft evidenced a normal structure of the mucosa, with a stratified squamous epithelium.
Discussion: Different techniques are taken into account for vaginal reconstruction according to the severity and to the type of malformation. We describe the use of bladder mucosal graft with favorable results after long term follow-up.
Key Words: Vaginoplasty, Bladder mucosa, DSD, Urogenital sinus, Graft

Vaginal malformations (VM) have conventionally been studied as a part of more complex malformations, such as mullerian and uterine anomalies. Many classifications have been proposed correlated to embryological aspect and surgical treatment.[1]

Persistent Urogenital Sinus (UGS) is a complex malformation where urethra and vagina fuse together to form a common channel. It represents the developmental arrest of paramesonephric ducts at about 9 weeks of gestation. It is usually associated with congenital adrenal hyperplasia (CAH) or other disorders of sexual development (DSD) in which external genitalia are virilized to a variable degree.[2]

It is rare to find a UGS isolated, without any external genitalia anomaly and DSD. Treatment ranges from urogenital sinus mobilization to intestinal replacement, pedunculated skin graft, amnion, buccal or peritoneum graft.[1,3–5]

Most frequently the vaginoplasty is performed using flap of local tissue when urethro-vaginal confluence in the UGS is close to perineal floor, while creation of neovagina with vascularized intestinal graft in high confluence is used.

We report long term results of a free bladder mucosal graft for vaginoplasty: the procedure was used to treat an infant with high confluence UGS without virilization of external genitalia.

Methods

We obtained informed consent from our patient to anonymously publish the medical history.

A female newborn was referred to our department with prenatal US diagnosis of an anechoic formation in pelvic area. There were no risk factors, and no previous cases of DSD within the family. Clinical examination of genitalia did not show virilization, but a single urogenital orifice opening in the vulva was present (Figure 1). No inguinal or labial hernias were observed. A distended abdomen due to a pelvic mass, caused a mild respiratory distress. At postnatal ultrasound a large anechoic image was confirmed, suggesting the presence of hydrometrocolpos, while the bladder was normal. Passing through urogenital sinus a 6 Fr. catheter was inserted into the bladder and an 8 Fr. catheter was inserted into the dilated uterus in order to decompress fluid collection. Antibiotic therapy was started. At 6 days of life X-ray genitography was carried out, with evidence of high confluence urogenital sinus and no evidence of VUR (Figure 2A).

Neonatal screening for associated pathologies or chromosomic alterations was negative. Karyotype was 46, XX. Hormonal profile was within normal ranges: 17-Hydroxyprogesterone, Androstenedione, LH and FSH were included in the study. Genetic studies for all principal forms of CAH were performed (CYP 21A2, CYP11) and resulted negative, as well as FISH study for the presence of SRY.

At 20 months of age vaginoplasty was performed. Endoscopy of UGS was carried out prior to surgical procedure. The location of the vaginal confluence in relation to the bladder neck and the perineal meatus is determined

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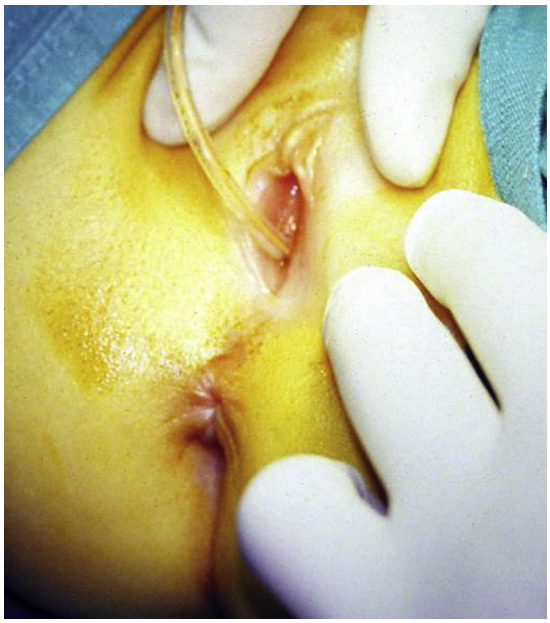

Fig. 1. Preoperative external genitalia.

endoscopically. The cystoscope and a calibraterd catheter are advanced into the bladder, through the perineal meatus. Using the centimeter marks, the distance from the bladder neck to the confluence is measured. The procedure was repeated to measure vaginal length and the distance between UGS confluence and the perineum. The hypoplasic vagina was about 10 mm between cervix and the confluence, without evidence of septum and urethra 15 mm from bladder neck to the confluence, while common channel was measured about 40 mm (Figure 2B). Uterine cervix was visualized and appeared normal. A Fogarty catheter was placed in the vagina, a Foley catheter in the bladder, and a probe in the rectum.

The operation started with a perineal inverted U incision above the anal verge. Dissection was carried inward and posteriorly to the confluence of the urogenital sinus, the vagina was separated from the urethra, and the posterior wall of the urogenital sinus was sutured with interrupted stitches. The UGS was dedicated to form the neourethra.

After that, Pfannenstiel skin incision was performed followed by a vertical fascia incision. The bladder, previously filled with air, was exposed. The detrusor muscle was incised vertically for approximately 5 cm: once performed careful dissection between the mucosa-muscolaris plane, the mucosa bulged through the muscolaris forming the classical blue dome cyst, then bladder mucosa graft was harvested: stay sutures were placed to define the limits of the graft, that was isolated by blunt forceps. At the end of the harvesting procedure the mucosal gap was sutured with running suture, as well as muscular layer and abdominal wall. After the graft was removed it was moistened with saline solution and wrapped in cylindrical shape using a 24 Fr. silicone tube as mold. The mucosal surface was directed toward the lumen and sutured with running 6.0 PDS suture. Interrupted sutures were used to reinforce the closure. Running suture did not traverse the entire graft, but ended 2 centimeters before the end of the cylinder, then interrupted stitches were placed in order to tailor the graft to the perineal bed. The tubularized graft was anastomized proximally to the vaginal stump and distally to the perineal skin adapting the length (Figure 3). At the end of the procedure the silicone tube was left in the neovagina through the anastomoses for 10 days. An 8 Fr. Foley catheter was left in the bladder passing through the urethra. III generation Cephalosporine for 10 days was given to the patient as protocol for major surgeries. Post-operative course was devoid of complication. We decided to avoid performing periodic dilation in order to minimize trauma to the child.[6]

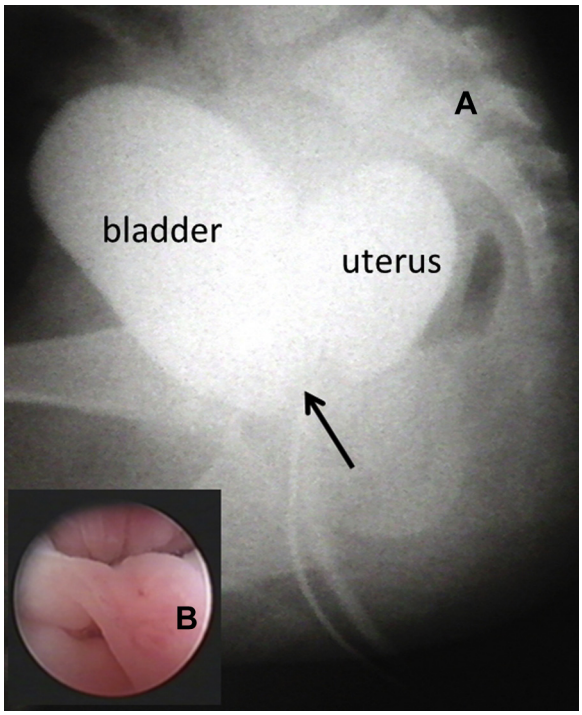

Fig. 2. X-ray genitography showing dilated uterus, bladder and urogenital sinus high confluence (arrow) (A). Endoscopic appearance at confluence (B).

Results

The patient underwent yearly clinical evaluation until 18 years of age. Bladder function was regular, with normal

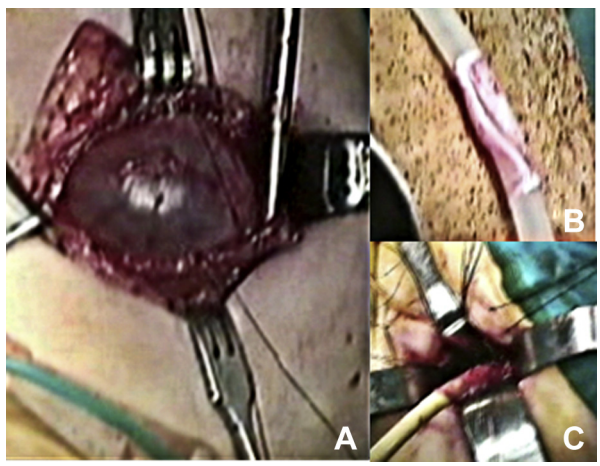

Fig. 3. Blue dome bladder mucosa (A). Tubularized bladder mucosa (B). Graft in situ after proximal anastomosis (C).