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LAPAROSCOPIC REMOVAL OF MÜLLERIAN DUCT REMNANTS IN BOYS

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ABSTRACT

Purpose: Müllerian duct remnants (MDRs) are present in a male pseudohermaphroditic form characterized by failure of the müllerian duct to regress due to insufficient production or peripheral action of müllerian inhibiting substance. The MDR can be asymptomatic but it often results in infections, stones and voiding troubles. Furthermore, it may develop into a neoplasm. Therefore, surgery is mandatory for large MDRs and symptomatic patients. Laparoscopic removal is described.

Materials and Methods: Six males were treated from February 1998 to February 2003. Age at surgery was between 3 and 18 years (mean 8.6). All patients showed severe hypospadias and 2 had mixed gonadal dysgenesis with ambiguous genitalia. Three patients presented with urogenital infections and all had a large MDR. Laparoscopic procedures, which were preceded by cystoscopy, were performed using a 10 mm umbilical trocar for the camera and 3, 5 mm trocars for instruments placed in the suprapubic region and iliac fossa bilaterally. The remnants were ligated with endoscopic loops or an endoscopic GIA stapler and cut.

Results: Mean operative time was 2 hours. We noted no complications. In 2 cases there was deferential ectopia and in another of mixed gonadal dysgenesis bilateral gonadectomy was performed because of the risk of degeneration. Feeding started on postoperative day 1 and the patients were discharged home on day 5. After a followup of 8 months to 4 years all boys were healthy.

Conclusions: Multiple approaches are used in traditional surgery, often leading to complications. Laparoscopy improves the view, decreases surgical risk and operative time, avoids large scars and allows more rapid hospital discharge.

KEY WORDS: urethra, testis, mullerian ducts, laparoscopy, urogenital abnormalities

Müllerian duct remnants (MDRs) presenting as müllerian duct cysts (MDCs), enlarged prostatic utricles (EPUs) or vagina masculina (VM) can arise from the posterior and bulbar urethra due to failure of the fetal testis to suppress these female structures, presumably as a consequence of an inappropriate or delayed secretion of testosterone and müllerian regression factor.[1,2] These abnormalities of the genitourinary tract may be seen in normal boys rather than in those with hypospadias and intersex disorders (mixed gonadal dysgenesis and true hermaphroditism).[3] Although the embryogenetic difference between EPU and MDC is still controversial, from a clinical point of view these entities are well defined.[4] EPU is a tubular shaped structure communicating with the urethra that is frequently observed in younger patients in association with hypospadias, intersex disorders or other anomalies (prune-belly syndrome, posterior urethral valves, Downs syndrome and imperforate anus).[1,3,5] EPUs are commonly identified while performing voiding cystourethrography (VCU) or cystoscopy. When other müllerian structures (cervix, uterus or fallopian tubes) are present, this complex is better defined as vagina masculina. On the other hand, MDCs are round and located in the midline. Size is a few cm to huge masses occupying the whole pelvis. They are often associated with normal external genitalia and become symptomatic later in life. They usually do not communicate with the urethra and, thus, they are rarely discovered during VCU or urethroscopy.[4]

EPUs and MDCs are frequently symptom-free but they may also cause urinary problems, such as recurrent infections or epididymitis, irritative problems (frequency, dysuria and urgency), obstructive problems (hesitance, a decreased urinary flow rate, prolonged voiding, urinary retention and stone formation) or less frequently terminal hematuria, urethral discharge, perineal and abdominal pain, a palpable mass, constipation, impotence, azoospermia and infertility.[5] Furthermore, a small but significant number of reports describe the development of malignant carcinoma from MDRs.[6]

Several surgical techniques have been advocated to access the retroprostatic space and remove MDRs. In 1994 the laparoscopic approach to excise an MDC in a man was described, preserving continence and potency.[7] In 1998 we first successfully performed laparoscopic removal of VM in the pediatric age group.[8]

We present our series of 6 boys with MDR who underwent to laparoscopic ablation. Embryology, clinical presentation, diagnostic evaluation and surgical management are discussed.

MATERIALS AND METHODS

Six boys with MDR 3 to 18 years old (mean age 8 years 6 months) were admitted to our department from February 1998 through February 2003 (see table). All except patient 4 had severe hypospadias. Patients 3 and 6 also presented with mixed gonadal dysgenesis (MGD) and ambiguous genitalia. Patient 1 had monolateral cryptorchidism associated with "didymo-epididymal" dissociation and deferential ectopia, and patient 4 had anorchia.

In patients 2 and 5 MDRs were represented by an EPU (grade II according to Ikoma et al[9]), whereas in the others VM with a hypoplastic uterus was present. In all cases the MDR ended in the prostatic urethra. In patient 1 2 small fallopian tubes and a vaginal septum were found in the surgical specimen.

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Müllerian duct remnant treated by laparoscopic removal

Pt No.	Clinical Features	Previous Surgery	Symptoms	Diagnostic Evaluations	Age at Surgery (yrs)	Surgical Procedure
1	VM, vaginal septum, scrotal hypospadias, lt cryptorchidism, D-E dissociation, deferential ectopia	Urethroplasty, orchiopexy	Recurrent epididymitis	Neg US, pos VUCG	15	Laparoscopic removal
2	EPU, hypoplastic, lt fallopian tube, scrotal hypospadias, deferential ectopia + atresia	None	None	Neg US, pos VUCG	3	Laparoscopic removal
3	VM, MGD, 46XY-45XO mosaicism, scrotal hypospadias, D-E dissociation, rt testis, lt gonadal streak	Rt testis biopsy, lt streak removal done elsewhere, urethroplasty	None	Neg US, pos VUCG + neg excretory urography done elsewhere, pos magnetic resonance imaging	18	Laparoscopic removal, lt streak gonadectomy
4	VM, rt cryptorchidism, lt inguinal hernia	Diagnostic laparoscopy (blind ending), lt herniorrhaphy	Lt epididymitis	Neg US, pos VUCG	4	Laparoscopic removal
5	EPU, scrotal hypospadias, post-urethroplasty stenosis + fistula	Urethroplasty, urethral dilations, urethroplasty revision	Recurrent urinary infections	Pos US, neg VUCG	8	Laparoscopic removal
6	VM, MGD, scrotal hypospadias, rt cryptorchidism, lt gonadal streak, post-urethroplasty fistula	Diagnostic laparoscopy (VM, hypoplastic rt testis + lt streak on diagnostic laparoscopy	None	Pos US, neg VUCG	4	Laparoscopic removal, bilat gonadectomy, testicular tissue cryopreservation

Disease-free outcome in all patients.

History was positive in 3 cases. Patients 1 and 4 had had many episodes of monolateral epididymitis. Patient 5 with a huge EPU presented with recurrent urinary infections unresponsive to antibiotic therapy. In these 3 cases the diagnosis of MDR was made by VCU, laparoscopy performed for unilateral cryptorchidism and ultrasound (US), respectively. In the latter patient VCU provided a false-negative result because of the narrow passage of the MDR into the urethra.

Three cases were asymptomatic. In patients 2 and 3 the diagnosis was made after screening VCU performed to detect hidden associated uropathies in patients with scrotal hypospadias (fig. 1). In patient 6 routine US at birth showed a hypoechogenic cystic-like structure located in the retroprostatic space, which was correctly interpreted as an MDR. VCU was negative. Briefly, US was diagnostic in 2 of the 6 cases (66.6% false-negative findings) and VCU was diagnostic in 4.

Peripheral blood lymphocyte karyotyping confirmed a normal male 46XY configuration in all except patient 3, who had MGD and incomplete masculinization with 46XY-45XO mosaicism. This patient also underwent magnetic resonance imaging to provide better anatomical definition of the internal genitalia and plan the correct surgical strategy. Treatment was laparoscopic removal in all boys.

Preliminary cystoscopy and MDR endoscopy were always performed to evaluate the shape, size and exact location of the communication between the MDR and the urethra. MDR endoscopy was often repeated during laparoscopic removal to use the endoscope light as a guide in the dissection of the retroprostatic space. It was particularly useful for EPUs in which the dome did not extend out from the peritoneal reflection (fig. 2).

Four trocar ports were used. A 10 mm trocar was placed for the 0-degree camera just below the umbilical scar. An additional 3, 5 mm trocars were inserted under direct vision in the suprapubic region, and in the left and right iliac fossae, respectively.

After pneumoperitoneum was achieved (11 per minute and 10 mm Hg) and the MDR was localized the peritoneal reflection coating the fundus of the EPU or surrounding the body of the hypoplastic uterus was incised. The MDR was grasped with forceps introduced by the suprapubic trocar and carefully dissected free of the surrounding tissues of the retrovesical space with another 2 instruments introduced through the iliac fossa trocars. Particular attention was given to avoid injury to the bladder neck, urethra, rectum, ureters, vas deferens, prostate and seminal vesicles. The use

FIG. 1. MDR detected by routine VCUG

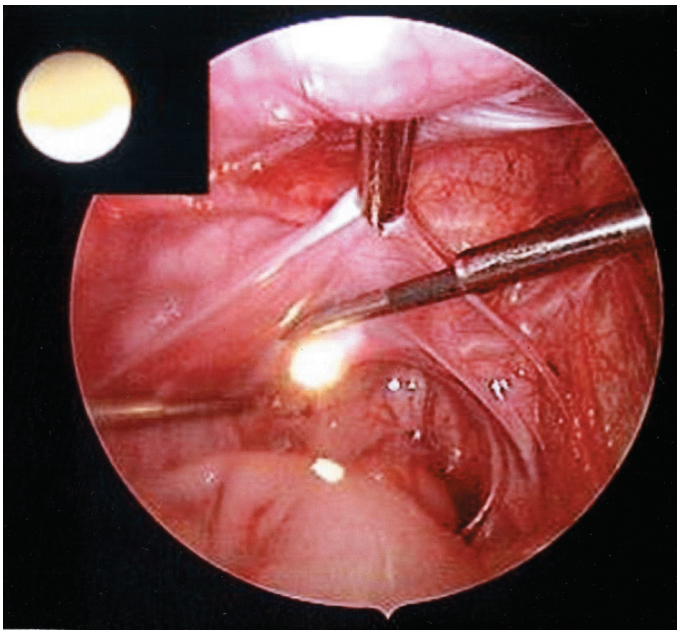

FIG. 2. Laparoscopic view of MDR under light of inserted cystoscope.

of a monopolar hook, bipolar coagulation and harmonic scalpel allowed completely bloodless dissection of all adhesions without clips and ligatures. After it was completely dissected the MDR neck was secured with 2 endoscopic loops or an endoscopic GIA stapler in older patients and resected just above its junction with the urethra (fig. 3). The specimen was removed through the 10 mm port.

A bladder Foley catheter was positioned before the end of the procedure.

RESULTS

In our experience mean operative time was 1 hour 53 minutes (range 1 hour 12 minutes to 3 hours 54 minutes). No major complications were recorded.

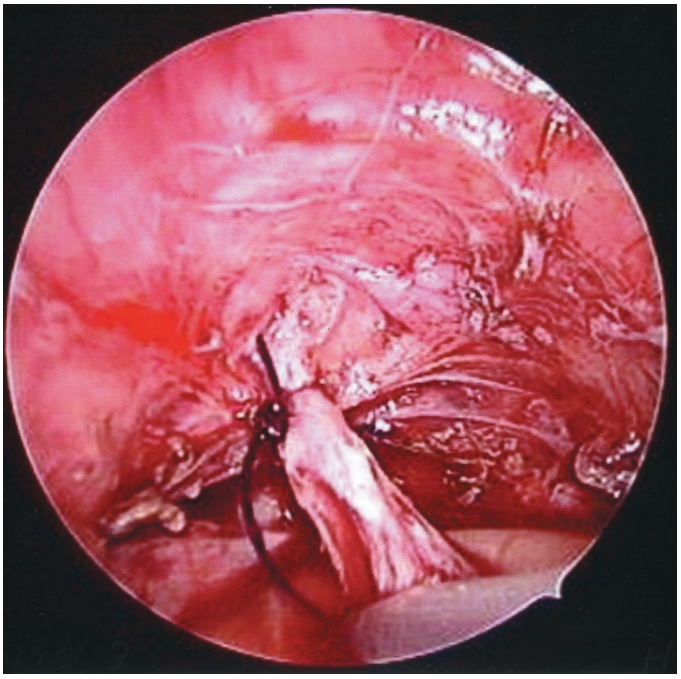

FIG. 3. Position of endo-loop down to MDR neck

Deferential ectopia, in which the vas is joined with the vaginal pouch or hypoplastic uterus, was found in patients 1 and 2, and associated with didymo-epididymal dissociation and histologically confirmed deferential atresia, respectively. In each case the vasa was transected. Patients 3 and 6 with MGD and ambiguous external genitalia had VM with a hypoplastic uterus and fallopian tubes connected with the posterior urethra. Moreover, a dystrophic testis dissociated from the epidydimis was found on 1 side and a gonadal streak was noted on the other side. In patient 3 the testis was normally located in the ipsilateral hemiscrotum. In this patient parents refused consent for right orchiectomy despite the risk of neoplastic evolution. Close long-term clinical and ultrasonographic followup was planned. In contrast, in patient 6 with MGD bilateral gonadectomy was performed. To provide the child with a chance of reproduction using new assisted reproductive techniques cryoconservation of testicular tissue was done.

Histological examination of the removed testis showed a poor germinal component with focal signs of spermatogenesis and diffused peritubular fibrosis.

Patients resumed free oral fluid intake on the day of the laparoscopic procedure and they were able to ingest food on postoperative day 1. Postoperative discomfort, which was generally limited to days 1 and 2, was easily controlled by the administration of nonsteroidal analgesics on demand. Convalescence was uneventful and patients were discharged home on postoperative day 5 after Foley catheter removal and the exclusion of micturition problems.

All patients are well and free from urogenital tract infections, voiding dysfunctions or urinary incontinence at a followup of from 6 months to 4 years. Because symptoms were completely absent, no further x-rays were performed.

DISCUSSION

During normal embryogenesis at about 6 weeks of development the paramesonephric or müllerian ducts appear bilaterally beside the wolffian duct and grow caudal, fusing together in the midline to form the uterovaginal cord. By month 4 of gestation they join the urogenital sinus in association with epithelial proliferation, called the müllerian tubercle.

In the female embryo these primordial structures form the tubes, uterus and the distal 4/5 of the vagina, which at its caudal end originates from the urogenital sinus.

In the male embryo the müllerian ducts regress under the effect of müllerian regression factor, a nonsteroidal hormone produced by Sertoli's cells in the testis during a short critical period at the end of the undifferentiated stage (weeks 9 to 10). After complete male sexual development only the cephalic and caudal ends of the müllerian ducts persist, as the appendix testis and a part of the prostatic utricle, respectively.[1,9]

Differentiation of the urogenital sinus and external genitalia is a complex phenomenon that develops from weeks 9 to 16 of gestation and depends on circulating levels of testosterone and its derivative dihydrotestosterone.[1]

Glenister reported that the prostatic utricle has clearly a mixed origin.[10] Its cranial portion is from the müllerian duct, while the caudal portion arises from the urogenital sinus. Because of this mixed origin, a hormonal imbalance in müllerian regression factor and testosterone secretion or the MRF responsiveness of target tissues during the short critical period could influence the formation of the prostatic utricle. Therefore, anomalies can result from incomplete regression of the müllerian ducts or incomplete androgen mediated closure of the urogenital sinus.[1,2]

US may demonstrate cystic or elongated formation located in the retrovesical space between the urethra, bladder neck and rectum. To our knowledge no experience with transrectal

US has been reported in the pediatric age group. VCU can show the EPU and its location, size and communication with the urethra. Magnetic resonance imaging or computerized tomography could be helpful in cases of persistent diagnostic problems after US and VCU.

In our experience the management of MDR has been based on clinical presentation, size, relationship with adjacent structures and radiological classification. Conservative treatment was adopted in grades 0 and 1 uncomplicated EPU[9] in the absence of other müllerian structures (ie a vagina or uterus) or lower urinary tract anomalies. In these patients we consider that careful followup is advisable based on periodic clinical examinations and US, if necessary integrated by VCU and urethroscopy. The development of persistent clinical symptoms, progressive MDR enlargement or other complications indicate a change from medical to surgical treatment. In contrast, surgery is mandatory in grade 2 or 3 EPU, which is unresponsive to medical treatment, and in all cases of VM because it may cause psychological disturbances in older patients. Sometimes MDR can hamper bladder catheterization by providing a preferential route to the urinary catheter. Thus, a further indication to remove MDR surgically is the difficulty of catheterizing a child with neurologic bladder who requires clean intermittent catheterization.[11]

Several surgical techniques have been proposed to treat MDR, including a perineal, retropubic or suprapubic extravesical, transvesical transtrigonal, transperitoneal, posterior sagittal transanorectal, anterior sagittal transanorectal and posterior perirectal or pararectal approaches.

The perineal approach[12] may be technically extremely difficult, especially in young children, since the small size of the perineum may not permit adequate exposure of the rectoprostatic space. When the EPU is large, it can endanger the rectum, external sphincter and branches of the pelvic plexus innervating the corpora cavernosa and rectum. For this reason it has been abandoned by most surgeons.

The retropubic or suprapubic extravesical method requires incomplete or complicated excision often entailing the removal of 1 or the 2 seminal vesicles, the vasa and portions of the prostate. Recently Ikoma et al reported a success rate of 100% with this approach alone or associated with the perineal approach.[12]

The transvesical transtrigonal approach was proposed by Monfort due to excellent exposure of the retrovesical space after posterior splitting of the bladder wall.[13] The MDR can be easily identified as well as the vasa, which can be separated and even saved when it ends normally. The disadvantages of this technique are the need for prolonged urinary drainage to allow bladder healing and occasional transient vesicoureteral reflux in the postoperative period. Monfort also performed antireflux vasocystostomy in 7 patients with deferential ectopia to preserve testicular function due to the possibility of collecting spermatozoa by urine ultracentrifugation.[14] Ikoma et al proposed reimplantation of the severed vas deferens into the posterior urethra or, if the vas was too short, vasovasostomy on the contralateral vas.[12] The effectiveness of deferential preservation must be investigated, especially in cases of mixed gonadal dysgenesis since no proof of fertility has been reported in these patients to date.[4,15] In our 2 cases of an ectopic vasa entering the MDR we did not perform vesical or urethral reimplantation because of didymo-epdidymal dissociation and deferential atresia, respectively. At any rate the laparoscopic approach does not allow any kind of vasal reimplantation in any case.

The transperitoneal approach was proposed for large MDRs and especially for VM, in which a uterine structure emerges from the peritoneal fold. In all other cases of a small MDR this invasive approach does not allow easy dissection of the rectovesical space, causing a high risk of damage to the nerves, urethra, ureters and vasa. Moreover, opening the peritoneum can create adhesions and consequently bowel obstruction.[16]

The posterior sagittal transanorectal approach, which was proposed by Siegel et al,[17] provides extensive exposure of the rectourethral space by splitting the posterior and anterior rectal walls without interfering with fecal continence. Nevertheless, it requires meticulous bowel preparation and patients must fast for a week postoperatively. Moreover, although with current methods of bowel preparation and antibiotic therapy it is possible to avoid colostomy, when bowel preparation is unsatisfying, temporary colostomy may be required to decrease the risk of an infectious complication, which in this area could compromise fecal continence.

The anterior sagittal transanorectal approach[18] provides similar excellent surgical exposure of the posterior urethra and retrovesical region by splitting only the anterior rectal wall and saving the elevator anus, posterior part of the muscle complex and external sphincter. Temporary colostomy is indicated if bowel preparation is not excellent.

A posterior pararectal or perirectal approach with rectal retraction has been proposed to avoid colostomy.[19,20] Although Pinter et al observed that this procedure does not impair fecal continence,[19] we believe that rectal mobilization causes rectal denervation by destruction of the reflex arc between the rectum and striated muscle sphincters.[18] Moreover, in cases of a huge MDR, this approach is unsuitable because the anus limits the distal extent of incision.

All of these procedures are often technically challenging and require prolonged hospitalization. Furthermore, they carry the potential risk of complications, such as infection, incontinence and impotence.[5]

For these reasons endoscopic treatment was introduced by many groups with encouraging results but several limitations. Recently Husmann and Allen reported their experience with endoscopic electrofulguration to treat EPU or residual rectourethral fistulas with a good success rate and minimal perioperative morbidity.[11,15] Nevertheless, they stated that this procedure is not adequate for VM because it does not remove the MDR, rather causing its obliteration, while there is a risk of injuring the adjacent ejaculatory ducts by transmitted electrical hazard.[11]

Laparoscopy obviates these disadvantages because it provides an optimal view due to imaging magnification. It permits fine dissection of the MDR with excellent exposition of all anatomical structures with a minimal access to the peritoneal cavity and a low incidence of postoperative adhesions.

In 1994 McDougall et al described laparoscopic excision of a 7 cm MDC in a 48-year-old man with the preservation of continence and potency.[7] They completely fulgurated the remaining cystic internal mucosa, which was less than 1 cm, to destroy any residual epithelium and achieve complete obliteration of the cyst cavity.

We agree with Husmann and Allen that endoscopic electrofulguration does not completely remove the cystic wall or eliminate the chance of neoplastic degeneration from the scarified remnant.[11] Moreover, in young children and in the hands of a skilled endoscopist the endoscopic procedure is not easy and it is potentially risky. Therefore, when a few mm of residual tissue are left in place, we prefer to adopt a precise clinical, ultrasonographic and eventually radiological or endoscopic program of long-term surveillance rather than risking iatrogenic damage to the important structures of the retrourethral space.

In our experience laparoscopy allowed us to perform effective dissection of the MDR from the retrovesical space because it provides clear identification of the ureters, seminal vesicle, urethra and bladder neck which, thus, can be gently retracted and preserved.

On the other hand, laparoscopy does not resolve the problem of the abnormal connection between the vas deferens and

the MDR because today it is still impossible to perform laparoscopic vasal reimplantation in the bladder or in urethra.

In conclusion, when compared with other surgical approaches, in the hands of a careful, trained surgeon the laparoscopic approach provides minimally invasive access to the retrovesical space, improving the view, decreasing the incidence of complications and operative time, and avoiding large scars.

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