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CASE REPORT

Persistent Mullerian duct syndrome with bilateral abdominal testis: Surgical approach and review of the literature

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Abstract *Objective:* We present two cases of persistent Mullerian duct syndrome (PMDS) with bilateral intra-abdominal testes and review the world's literature with special attention to diagnosis and surgical management.
Patients and methods: Two consecutive cases of PMDS with bilateral intra-abdominal testes from our institution are presented with detailed descriptions of the presentation, physical examination, laboratory profiles, surgical findings, and treatment undertaken. Follow-up at 1 year postoperatively is included.
Results: Bilateral orchiopexy was accomplished in both the cases. In one case this was possible after division of the persistent Mullerian structures in the midline to achieve testicular mobility. In a subsequent case, splitting of the Mullerian complex did not provide adequate mobilization and microvascular autotransplantation was performed with an excellent surgical outcome.
Conclusions: Bilateral intra-abdominal testes in the setting of persistent Mullerian duct syndrome are a rare entity and controversy remains as to the ideal surgical treatment. Our two cases represent the first reported examples of open single-stage bilateral orchiopexy with division of the Mullerian complex and preservation of the vas deferens (1-year-old boy) and microvascular autotransplantation (5-year-old boy).
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Introduction

Persistent Mullerian duct syndrome (PMDS) is characterized by normal virilization and XY genotype with failure of Mullerian duct regression. It was first described by Nilson [1] in 1939 and given the name hernia uteri inguinale. Since then, nearly 240 cases of this rare condition have been described. PMDS is caused by either lack of Mullerian inhibitory substance (MIS) or dysfunction of the MIS receptor, with the former slightly more common. In 16% of PMDS subjects no genetic mutation is detectable [2]. The most common presentation is a child with a normally appearing phallus and one scrotal and one inguinal testis. Less commonly, transverse testicular ectopia or bilateral cryptorchidism may be seen [3–5].

Bilateral cryptorchidism with intra-abdominal testis is the least common presentation and represents an interesting surgical challenge. Numerous methods of treatment for abdominal testis have been described in the literature. PMDS with bilateral intra-abdominal testes can pose additional challenges for patients. We report two cases of PMDS with bilateral abdominal testis and discuss the unique management dilemmas for this condition. We attempt to review the world's literature with special attention to diagnosis and surgical management.

Case reports

Case 1

A 1-year-old boy presented for elective orchiopexy for bilateral undescended testis discovered at birth. MIS testing was offered but refused by the parents. Laparoscopy was performed which revealed bilateral low abdominal testis. A pale yellow fibrous structure was noted originating deep in the pelvis, extending across to both gonads and tethering them towards the midline. Standard bilateral laparoscopic orchiopexy was performed (Fig. 1). To test whether leaving the uterine structures in the midline would adversely compress the bladder, we filled the bladder with normal saline and the testicles were pulled back up into the abdomen. With the midline structure impinging on the bladder, we considered dividing the midline structure in the vertical midline through a laparoscopic approach. However, from a posterior vantage point we did not have adequate visualization of the vas deferens to allow safe midline division without potential injury to the Wolffian structures. We therefore converted to an open approach via a Pfannenstiel incision. Biopsy of the gonads and midline structures confirmed testicular and Mullerian histology. The persistent midline Mullerian structure was divided in the midline after conversion to an open procedure. Great care was taken to divide the uterus–vagina in the midline so as not to injure the vas deferens on either side (Fig. 2). Orchiopexy was successfully performed (Fig. 3) and the testicles were noted to be sitting comfortably in the dependent scrotum 1 year following the procedure. Subsequent retrograde urethrography failed to demonstrate any connection between the vaginal remnant and the prostatic urethra.

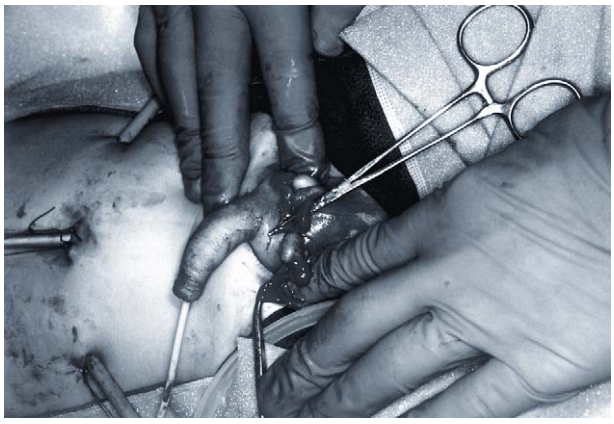

Figure 1 Testicles sitting comfortably in scrotum following bilateral laparoscopic orchiopexy. Upon filling the bladder through a Foley catheter, tension was put on the intact Mullerian structures and the testicles ascended into the abdomen.

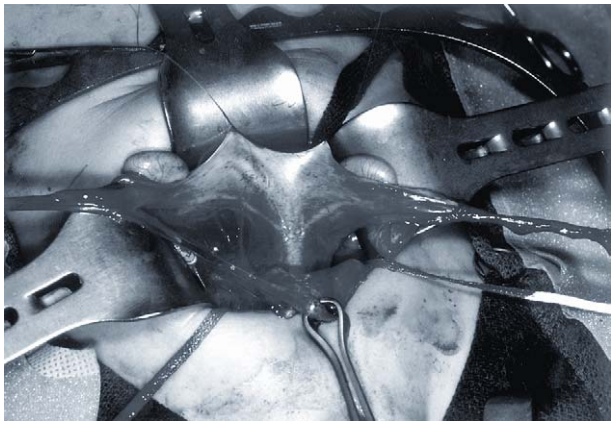

Figure 2 Exposure of the Mullerian structures and testicles through an open surgical approach. The vas deferens can be seen running under the uterine horns–fallopian tubes bilaterally.

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