Laparoscopic orchidopexy in persistent Müllerian duct syndrome

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Purpose **When faced with the diagnosis of PMDS, the question still remains as to what is the best approach to Müllerian structures. The aim of this study was to describe a novel operative approach considering the vascular anatomy and malignant potential of these structures.**

Methods **Two boys with PMDS underwent orchidopexy by means of en-masse descent of Müllerian-gonad complex onto the scrotum. The gonads were biopsied and fixed in the scrota and the Müllerian structures occupied the region of the median raphe.**

Results **The operating time was the same as that taken for orchidopexy. The children were discharged on the day after surgery and were well at 6-month follow-up.**

Conclusion **The preservation of Müllerian structures and placement in the scrotum allow for easy palpation and recognition of changes in size and consistency heralding malignancy. The risk of operative injury to the vas and vessels is minimized.** ***Ann Pediatr Surg*** **11:222–225 © 2015 Annals of Pediatric Surgery.**

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Keywords: laparoscopic orchidopexy, persistent Müllerian duct syndrome, undescended testis

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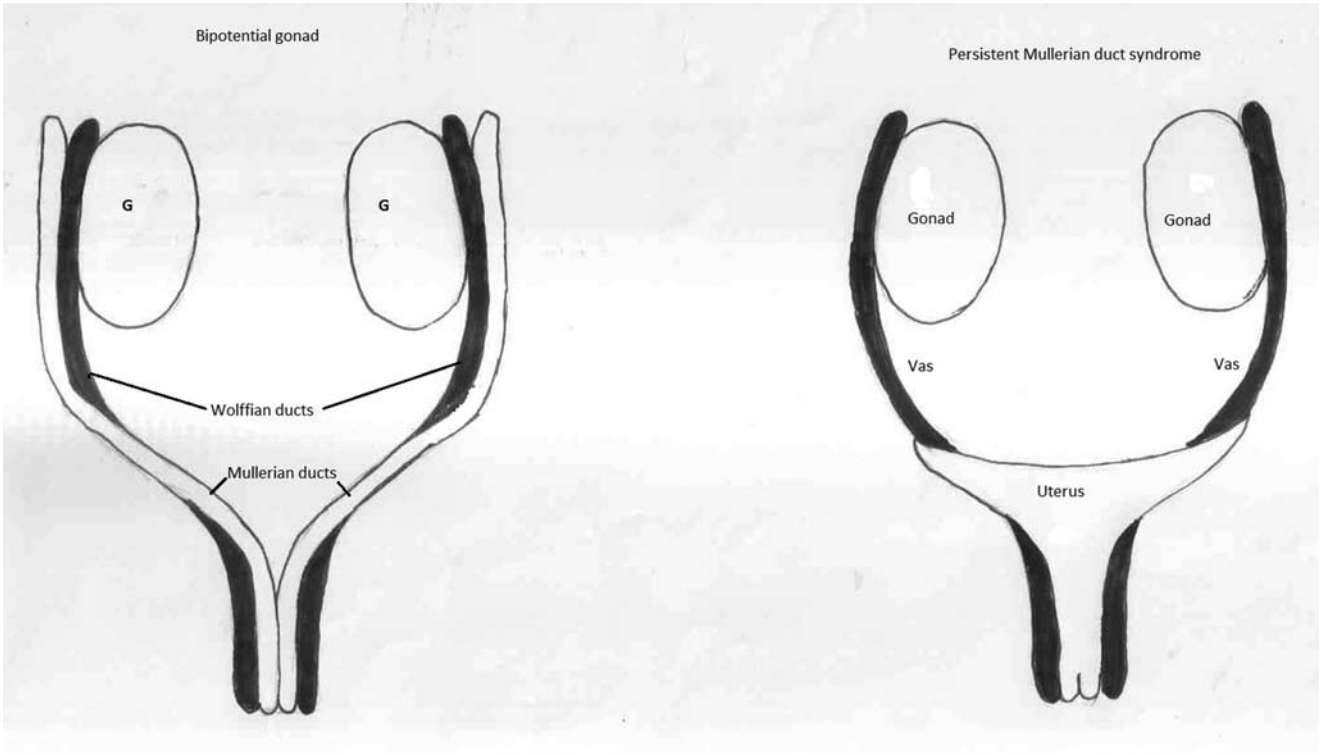
Introduction

Persistent Müllerian duct syndrome (PMDS) describes a phenotypic male with unilateral or bilateral impalpable testis and persistent Müllerian ducts (Fig. 1). The currently acceptable surgical management involves either removal of the Müllerian structures with or without microvascular autotransplantation of the testes in the scrotum or splitting the uterus in the midline to facilitate descent of the gonads with or without mucosal stripping [1–3]. These require open surgery or advanced laparoscopy skills and are time consuming. Furthermore, separating the Müllerian structures can damage the vas

Fig. 1



Schematic diagram of bipotential gonad and persistent Müllerian duct syndrome.

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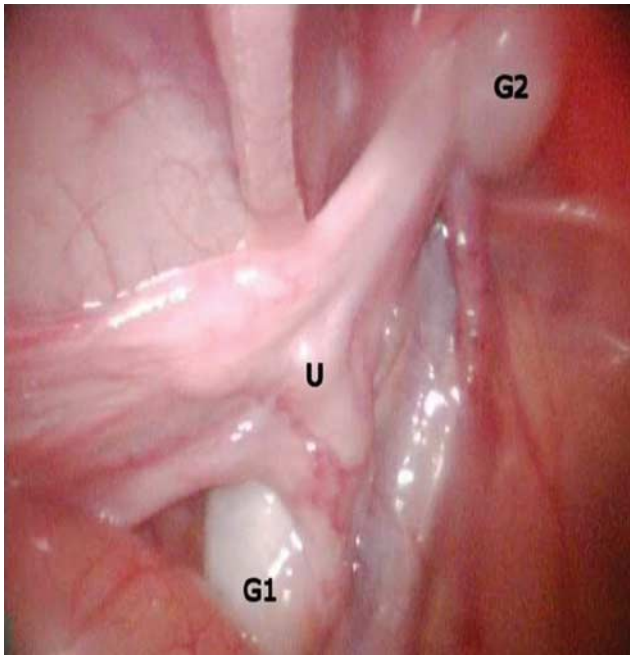
and vascular supply to the gonad. We approached two patients with PMDS using a novel technique that is safe and straightforward and renders the Müllerian structures accessible for palpation. The risk of surgical injury to the vas and vessels is also minimized.

Methods

The diagnosis of PMDS was established during diagnostic laparoscopy for two boys (Fig. 2). The clinical findings are summarized in Table 1.

Our operative policy in PMDS had been to excise or split the uterus to facilitate testicular fixation in the scrotum. However, on reviewing the current literature as regards the risks and benefits of retaining the Müllerian structures, a new operative strategy evolved and was performed on the two children. A standard laparoscopic orchidopexy was being undertaken when the diagnosis of PMDS was encountered. The internal spermatic vessels were surprisingly lax due to the associated transverse testicular ectopia (Figs 3 and 4). The peritoneal and gubernacular attachments on one side were divided. The entire complex of gonads and Müllerian structures was brought through the opposite side using the modified Prentiss manoeuvre without tension on the vessels (Fig. 5) [4]. Biopsies were taken, and the contralateral gonad was passed through the scrotal median raphe into the opposite hemiscrotum (Fig. 6). This positioned the Müllerian structures in midline of the scrotum between the testes (Fig. 7). The importance of self-examination and the implications of increase in size and consistency of the external genitalia and the occurrence of hematuria or pain were explained to the caregiver. Both children were

Fig. 2



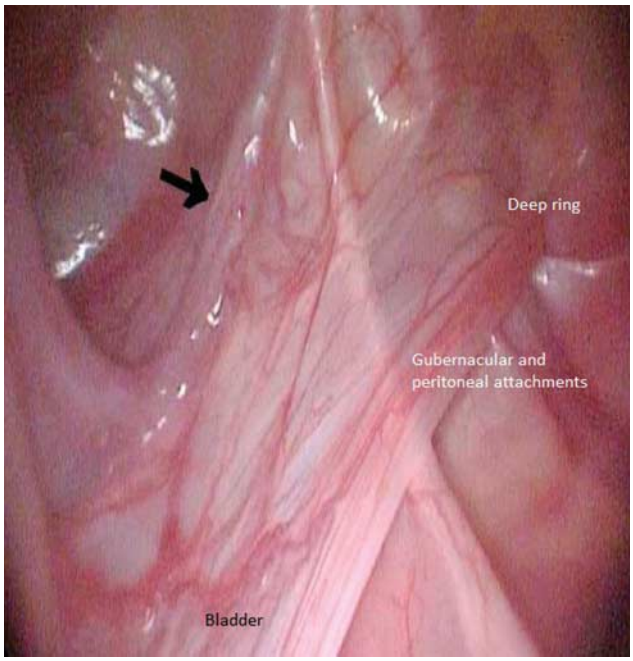
Persistent Müllerian duct syndrome with two gonads (G1, G2) and Müllerian remnants (U) in a phenotypic male.

Table 1 Clinical details of the two children with persistent Müllerian duct syndrome

Variables	Case 1	Case 2
Age at presentation (years)	1	1
Nonpalpable UDT	Left	Bilateral
Hernia	Absent	Present
Transverse ectopia	Present	Absent
Cystic change in gonads	Present	Present
Karyotype	46XY	46XY
Gonadal biopsy	Testis	Testis

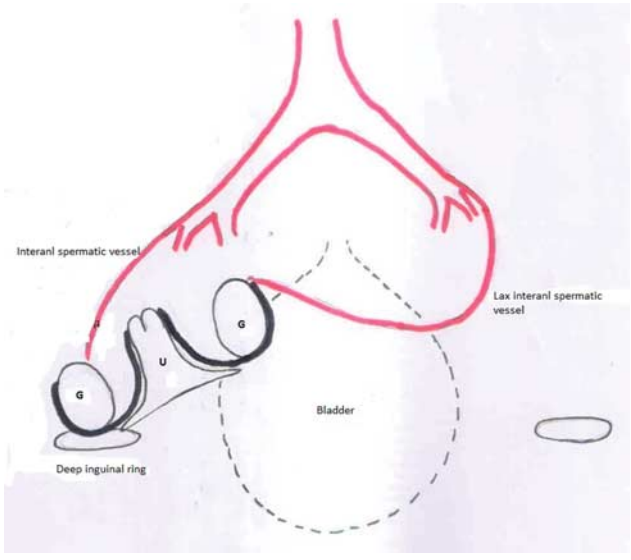
UDT, undescended testis.

Fig. 3



Lax left internal spermatic vessels (black arrow) and gubernacular and peritoneal attachments that were divided.

Fig. 4



Schematic diagram of the internal view showing the laxity of internal spermatic vessels and close association of the vas to the uterus.

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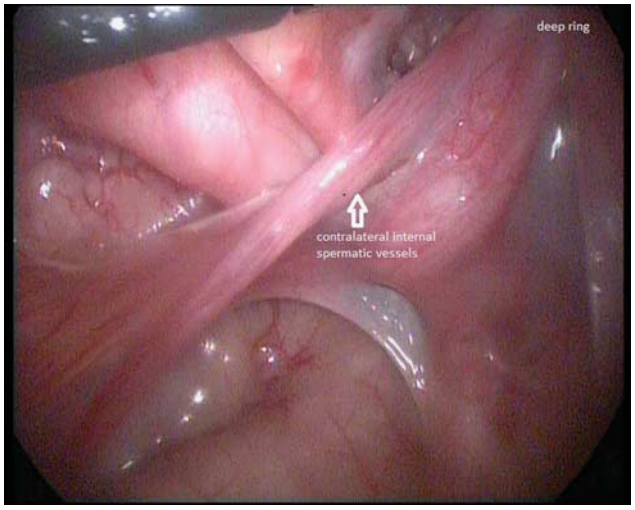
Fig. 5



Lack of tension on vessels after gonads placed in the scrotum with M**ü**llerian structures.

Fig. 6

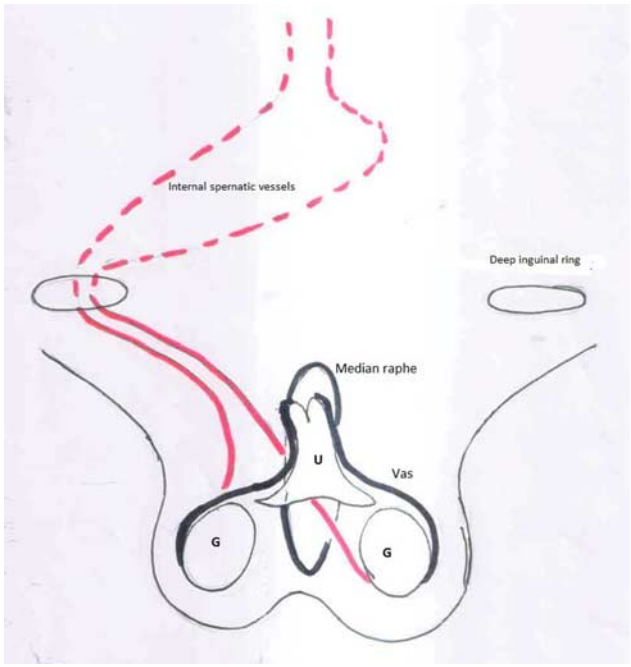
Both gonads and Müllerian structures in the scrotum.

discharged the next day. At 6-month follow-up, both testes were palpable and the children were asymptomatic. We plan a systematic follow-up of these children through puberty to educate the patients themselves on self-examination.

Discussion

PMDS is a rare autosomal recessive disorder that is caused by a defect in anti-Müllerian hormone or its receptor [3]. Two types of PMDS have been described clinically [5]. The male type comprises 60–70% of cases and consists of a unilateral palpable gonad. The female type is less common with bilateral impalpable gonads. The diagnosis of PMDS must be suspected when there is

Fig. 7



Schematic diagram of the final position of the gonads and Müllerian structures in the scrotum.

a palpable undescended testis with hernia on one side with nonpalpable undescended testis on the other side. The diagnosis is usually made during orchidopexy or inguinal herniotomy [3].

Approximately 200 cases of PMDS have been reported in the literature [3]. When encountered with the diagnosis of PMDS, the question arises on the fate of the Müllerian structures. The aim of surgical correction of PMDS was to reduce risk and facilitate early detection of malignancy and maximize reproductive potential. The incidence of malignancy reported in the testes in PMDS is 18% – similar to the rate of abdominal testes in normal men [6]. Thus, the scrotal position facilitates early detection. Eleven cases of malignancy have been reported in the Müllerian structures in PMDS [3]. This has prompted the removal of the Müllerian structures [1–3]. The vascular anatomy of the vas deferens is complex and closely relates to the uterus and fallopian tubes. Removal of the Müllerian structures poses a risk to the viability of the testis and may damage the vas. Microvascular autotransplantation of the testis after complete excision of the Müllerian structures has been described but may not be always feasible [1]. Stripping of the endometrium after division of the uterus has also been tried [2]. Retaining the Müllerian structures could also cause cyclical hematuria and urinary obstruction. However, these concerns are not borne out in the literature in the benign setting, and the presence of these symptoms heralds malignant transformation [3,5].

Our technique is a safe and straightforward alternative to the management of Müllerian structures in PMDS. The operating time is the same as that taken for orchidopexy by the particular surgeon. The procedure is single-staged and can be performed in day care. The internal spermatic vessels are inherently lax and the proximity of the testis

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and uterus in the pelvis is an added advantage. This is especially true in cases with transverse testicular ectopia in which one gonad is palpable in the inguinal canal. Furthermore, avoiding excision of the Müllerian structures minimizes the possibility of vascular compromise or injury to the vas, which is closely applied to the uterus. It places the Müllerian structures within the scrotum with the testes, where changes in size and consistency heralding malignancy can be easily palpated. This avoids the usage of radiological investigations for follow-up, and clinical examination can be used for long-term follow-up, which is required after orchidopexy in PMDS.

Conclusion

As more cases of PMDS are being diagnosed, the operative strategy requires a rethink. Our approach of laparoscopically bringing both testes and Müllerian structures into the ipsilateral scrotum using the modified Prentiss manoeuvre is safe, straightforward and single-staged with no risk to the vascularity of the gonads or vas.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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