Journal of Pediatric Urology (2014) 10, 929—933

Journal of Pediatric urology

Excision of Mullerian duct remnant for persistent Mullerian duct syndrome provides favorable short- and mid-term outcomes

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Received 11 October 2013; accepted 17 January 2014
Available online 8 February 2014

KEYWORDS
Persistent Mullerian duct syndrome;
Undescended testis;
Mullerian duct anomaly;
Inguinal hernia

Abstract *Objective:* In dealing with persistent Mullerian duct syndrome (PMDS), excision of Mullerian duct remnant (MDR) has been rarely mentioned in the past, but recent discussions have taken place. This study aimed to evaluate the operative feasibility and outcomes.
Materials and methods: Three patients with PMDS operated on with excision of MDR between 2000 and 2009 were enrolled. Medical records were retrospectively collected and reviewed.
Results: Bilateral undescended testis was manifested in all cases. Two patients presented with incarcerated hernia, requiring emergency herniorrhaphy at the ages of 6 months and 10 days, respectively. Reconstruction comprising simultaneous MDR excision and orchiopexy was made at the age of 1 year. MDR was incidentally found in another patient during operation for undescended testis. Immediate reconstruction was accomplished. Follow-up periods were 12.0, 3.5, and 2.5 years, respectively. Worse outcomes were noted on the two testes with repeated operations for incarcerated hernias, whereas the outcomes on the other four testes with a single operation were favorable.
Conclusions: Excision of MDR is technically feasible, and provides favorable outcomes in cases of a single operation. For experienced surgeons, immediate reconstruction should be the priority when this abnormality is incidentally encountered at an age suitable for orchiopexy.

Introduction

Persistent Mullerian duct syndrome (PMDS) is a rare 46,XY disorder of sex development, characterized by the presence of internal female productive organs in genotypically normal males. The etiology is absence of either Mullerian inhibiting substance or anti-Mullerian hormone receptor, subsequently blocking Mullerian duct regression.

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1477-5131/$36
http://dx.doi.org/10.1016/j.jpurol.2014.01.012

PMDS is frequently associated with a variety of testicular abnormalities, mostly cryptorchidism [1,2]. There is agreement that orchiopexy is indicated to preserve spermatogenesis function and prevent testicular malignancy [3,4]. Excision of Mullerian duct remnant (MDR) is thought to be technically demanding and to put patients at risk of deferential injuries [4]. The main concern is that the retained Mullerian structure may become malignant [5–9]. Optimal surgical management of PMDS has not been standardized. Here, we review our experience of excision of MDR, and evaluate surgical feasibility and outcome.Table 1

Materials and methods

Excision of MDR was performed in three patients with PMDS at our institution between January 2000 and December 2009. Orchiopexy was performed simultaneously as bilateral undescended testis was present in all patients. With institutional review board approval, patient medical records were retrospectively reviewed; they included presentation, operative findings, and genetic and hormonal surveys, as well as the outcomes of testicular locations and development. The operative techniques and clinical courses of each case are described in the following sections.

Operative method

By entering the peritoneal cavity via a low transverse abdominal incision, MDR could be detected posterior to the urinary bladder. Vasa deferentia were tightly attached on both sides of the MDR. Biopsies were performed on both gonads. With the assistance of loupes magnification, vasa deferentia were cautiously separated from MDR (Figs. 1 and 2). For the purpose of protecting the vasa deferentia, it was imperative to dissect closely along the margin of the MDR. Generally, tutting into the adventitia was necessary. Dissection was carried out down to the confluence where vasa deferentia joined the vagina (Fig. 3). MDR was resected

Figure 1 Posterior view of Mullerian duct remnant (MDR). A Mullerian structure and normal male external genitalia were observed. A loose space between the MDR and the vas deferens could be inspected at the site of arrow; dissection began here toward the gonads.

(Fig. 4), while the stump was repaired. Vasa deferentia were detethered at this time and no longer restricted to orchiopexy. When a broad ligament was present, it was released to facilitate mobilization of the testes and spermatic cords. The original length of internal spermatic vessels was sufficient for orchiopexy; hence, the Fowler–Stephen procedure was generally dispensable. Thereafter, the testes were brought down through the inguinal canal and comfortably fixed to the bottom of the scrotum.

Patient 1

This male infant was born via in vitro fertilization with bilateral peeping-type undescended testes. An irreducible right inguinal hernia occurred at the age of 6 months.

Table 1 Summary of surgical management for persistent Mullerian duct syndrome.

Author	Year	Cases (n)	Presentation	Surgery	Duration of follow-up	Outcome
Loeff et al. [10]	1994	2	Undescended testis; inguinal hernia	MDR excision	NA	NA
Vandersteen et al. [4]	1997	1	Bilateral undescended testis; transverse ectopia	Left MDR in situ	NA	NA
De et al. [3]	2002	1	Unilateral undescended testis	Left MDR in situ	NA	NA
Shirasaki et al. [13]	2003	1	Bilateral undescended testis	Partial MDR excision	NA	NA
Parelkar et al. [12]	2009	1	Bilateral undescended testis	Split MDR in midline; laparoscopy	6 mo	NA
Chaabane et al. [14]	2010	1	Transverse testicular ectopia	Proximal salpingectomy; corporeal hysterectomy	18 mo	Normal
Farikullah et al. [9]	2012	8	Undescended testis	MDR excision; laparoscopy; preferred one-stage reconstruction	0.5–5.0 y	NA
This study	2014	3	Undescended testis; incarcerated hernia	MDR excision; preferred one-stage reconstruction	2.5–11.0 y	Favorable

Note. MDR = Mullerian duct remnant; NA = not available.