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Original Article

Management of the patients with persistent Müllerian duct syndrome: Is the ultimate goal testicular descent?

Persistan Mülleriyen kanal sendromlu hastaların yönetimi: Nihai hedef testislerin indirilmesi midir?

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ABSTRACT

Objective: Persistent Müllerian duct syndrome (PMDS) is a rare congenital disease characterized by the presence of rudimentary Müllerian structures within an intra-abdominal or hernial sac in a virilized male, often presenting as undescended testes. In this study, we aim to present a series of the PMDS patients who were managed by orchiopexy without removal of Müllerian remnants (MR).

Material and methods: Between May 2010 and June 2017, we treated six cases diagnosed as PMDS in our department. Laparoscopy and gonadal biopsy were performed in all patients, and vessel ligation was done in four patients for the first session of Stephen-Fowler orchiopexy. After initial diagnosis, genetic analyses and endocrine investigations were performed. Demographic and clinical features of the patients, operative methods and follow-up data were analyzed retrospectively.

Results: Mean age of the patients was 5.5 years. Three boys had undergone inguinal surgery due to hernia or undescended testis, while others were diagnosed during laparoscopic investigation of nonpalpable testis. As a definitive operation, testes and MR were completely removed in an adult patient, and the remaining patients were treated with laparoscopic or open orchiopexy with or without utero-cervical splitting and the MRs were left in situ. Two testes atrophied during follow-up period.

Conclusion: The goal of the approach in PMDS patients is to preserve testes, as well as carry them to their natural location. Leaving the MR in place is a suitable option for blood circulation of the testes but the long-term results are still unknown.

Keywords: Disorders of sex development; laparoscopic orchiopexy; Müllerian remnant; persistent Müllerian duct syndrome.

ÖZ

Amaç: Persistan Mülleriyen kanal sendromu (PMKS), virilize bir erkekte intraabdominal ya da fıtık kesesi içerisinde rudimenter Müllerian yapıların varlığı ile karakterize nadir görülen doğumsal bir hastalıktır ve sıklıkla inmemiş testis ile birliktedir. Bu çalışmada, Mülleriyen kalıntılar (MK) çıkarılmadan orşiyopeksi ile tedavi edilen olgularımızı sunmayı amaçladık.

Gereç ve yöntemler: Mayıs 2010-Haziran 2017 yılları arasında bölümümüzde PMKS tanısı alan altı olgu tedavi edildi. Tüm hastalarda laparoskopi ve gonadal biyopsi ve dört hastada Stephen-Fowler orşiyopeksi ilk seansı için damar ligasyonu yapıldı. İlk tanıdan sonra genetik analizler ve endokrin incelemeleri yapıldı. Olguların demografik ve klinik özellikleri, operatif yöntemler ve takip verileri retrospektif olarak incelendi.

Bulgular: Hastaların yaş ortalaması 5,5 yıl idi. Fıtık ya da inmemiş testis nedeniyle üç çocuk, kasık cerrahisi geçirmişti, diğerlerinde tanı, ele gelmeyen testis araştırılırken laparoskopi ile tanı konulmuştur. Kesin ameliyat olarak yetişkin bir hastada testisler ve MK tamamen çıkarıldı, geri kalan hastalar laparoskopik veya açık orşiyopeksi ile utero-servikal splitting yapılarak veya yapılmadan tedavi edildi ve MK yerinde bırakıldı. İzlem süresince iki testiste atrofi gelişti.

Sonuç: PMKS'li hastalarda yaklaşımın amacı, testisleri korumak ve doğal yerlerine taşımaktır. MK'yi yerinde bırakmak testislerin kan dolaşımı için uygun bir seçenektir ancak uzun vadede sonuçlar hala bilinmemektedir.

Anahtar Kelimeler: Cinsel gelişim bozukluğu; laparoskopik orşiyopeksi; Mülleriyen kalıntı; persistan Mülleriyen kanal sendromu.

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Introduction

Persistent Müllerian duct syndrome (PMDS) is a rare form of male 46 XY disorders of sex development (DSD). The pathophysiology of PMDS is mostly explained by failure of synthesis or release of Müllerian inhibiting substance (MIS) by immature Sertoli cells, or the failure of end organs to respond to MIS. Clinically, the patient with PMDS presents with non-palpable/undescended testes or an inguinal hernia with a palpable testis within the hernia sac. Although imaging techniques may help to investigate the intersex abnormalities, mostly preoperative diagnosis of PMDS is practically impossible, as the external male genitalia appears to be normal.[1-3]

A standard approach to PMDS has not been identified. Even so, when the Müllerian remnants (MRs) are found incidentally during surgery, generally, a gonadal biopsy is done, and MRs are left in the pelvis. After the genetic and hormonal analyses are carried out, the definitive surgery is done in another session.[4] Although a large number of cases and series have been published in the literature, no standard approach has been established for PMDS. We aimed to present our management of the PMDS patients, and to discuss the purpose of the management.

Material and methods

Six consecutive patients with PMDS who managed in our clinic between May 2010 and June 2017 were retrospectively analyzed. Demographics, clinical presentation, diagnostic tests, operative data, and clinical outcomes were reviewed. Since our study was based on retrospective data, and we performed routine procedures, we did not receive approval from the ethics committee because our management did not pose an additional risk for the patients. Nevertheless, retrospective study was conducted in accordance with the Helsinki declaration.

The steps involved in our approach to the patients with PMDS are summarized as follows: Laparoscopy was performed in all cases with non-palpable testes, even if an MR was incidentally detected during an open inguinal exploration. On laparoscopy, the internal genital structures were inspected, bilateral testicular biopsies were performed if possible, and then the lengths of the gonadal vessels were assessed. After obtaining informed consent from the parent during surgery, if it was not long enough to perform orchiopexy, first session Stephen-Fowler's (SF) operation was done using the clips, and laparoscopy was terminated by leaving the MR in place. Each child was consulted with a pediatric endocrinologist and geneticist for hormonal and genetic evaluation to establish a definitive PMDS diagnosis and appropriate treatment plan. To relieve the concerns of the parents about the gender identity of their child, they were informed about the disease and the major risks associated with their surgeries. Another informed consent was obtained for the elective surgery. Following collaborative decision, 6 months later, open or laparoscopic orchiopexy was performed with/without utero-cervical splitting (UCS), but MRs were not removed in all patients because of the risk of gonadal vessel or cord injury. Patients were followed-up every three months in the first year postoperatively to assess the location and consistency of the testes, and additionally, MRs were controlled annually by ultrasonography (US).

Results

Clinical features, operative and follow-up data of six PMDS patients are presented in Table 1. Median age of the patients was 5.5 years (range 1.5-15). Three patients had previously undergone surgery for inguinal hernia, undescended testis, and acute scrotum. When abnormal cord structures were encountered during inguinal examination, the patients were referred to our clinic for further investigation. US revealed an intra-abdominally located testis in three testicular units, but no internal genital structures were demonstrated. Laparoscopy revealed the presence of intra-abdominal gonads suspended from persistent MR (Figure 1). In the last case, testicular fusion which is a part of transverse or crossed testicular ectopia (TTE) was detected (Figure 2). In the seven testicular units of 4 patients, testicular vessel ligations were done for the first session of SF operation for subsequent orchiopexy. Chromosome analysis revealed the patients to be a normal male of karyotype 46 XY. Serum pituitary and gonadal hormone levels were within the normal ranges. Histopathologically, testicular biopsies taken from ten units in 6 patients showed normal testicular structures.

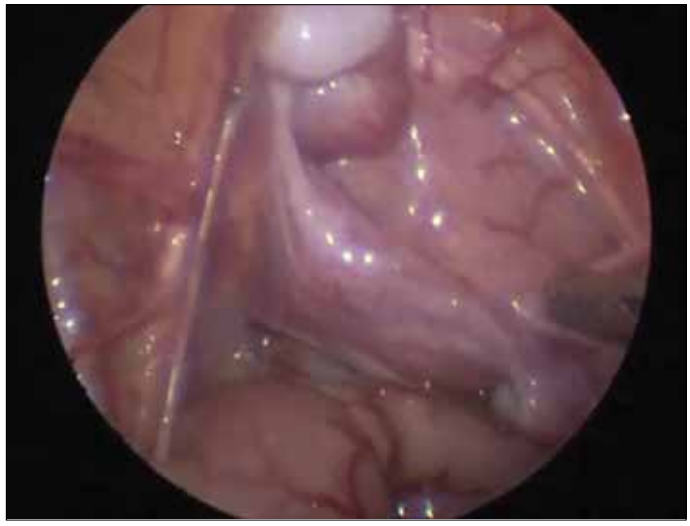

Figure 1. Bilateral intra-abdominal testes and Müllerian remnants in a patient who underwent bilateral vascular clipping at first operation

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Table 1. Demographic and clinical features of the patients

Case	Age	Complaint	Physical examination	US	Previous interventions	Definitive operation	Outcome Right/Left
1	15 y	Absence of bilateral testes	Bilateral nonpalpable testes	Right intra-abdominal testis, nonvisualized left testis	1. Diagnostic laparoscopy	Total excision	-/-
2	2 y	Right hernia and absence of left testis	Right inguinal hernia+ nonpalpable left testis	Right nonvisualized testis	1. Right orchiopexy 2. Diagnostic laparoscopy	UCS+left open single-stage SF orchiopexy	Good/Good
3	1.5 y	Absence of bilateral testes	Bilateral nonpalpable testes	Bilateral nonvisualized testes	1. Diagnostic laparoscopy and bilateral first- stage SF	UCS+bilateral laparoscopic second-stage SF orchiopexy	Good/Good
4	2.5 y	Absence of bilateral testes	Bilateral nonpalpable testes	Bilateral intra-abdominal testes	1. Diagnostic laparoscopy and bilateral first stage SF	UCS+ bilateral open second- stage SF orchiopexy	Good/Atrophic
5	7 y	Right testicular pain	Right testicular torsion+ nonpalpable left testis	Right testicular ischemia, nonvisualized left testis	1. Right detorsion and orchiopexy 2. Diagnostic laparoscopy and left first stage SF	UCS+unilateral laparoscopic second-stage SF orchiopexy	Atrophic/Good
6	5.5 y	Absence of bilateral testes	Bilateral nonpalpable testes	Bilateral nonvisualized testes	1. Right herniorraphy 2. Diagnostic laparascopy and bilateral first- stage SF	Open second- stage SF orchiopexy by passing through single site	Good/Good

US: ultrasonography; SF: Stephen-Fowler operation; UCS: utero-cervical splitting

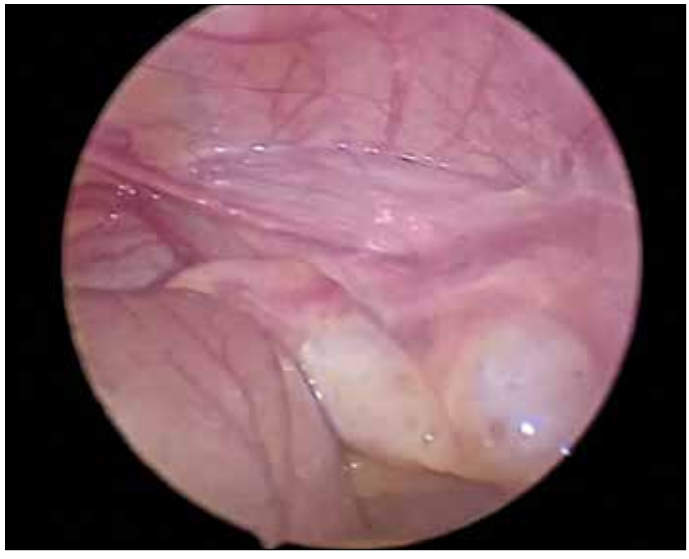

Figure 2. Photograph showing the intra-abdominal fused testes which is extremely rare form of transverse testicular ectopia in a patient with persistent Müllerian duct syndrome

As definitive surgery, in the fifteen year- old patient, the gonads and the MR were completely removed with the decision of our hospital council due to the concern of malignancy. Laparoscopic second stage SF orchiopexy with UCS was performed in two

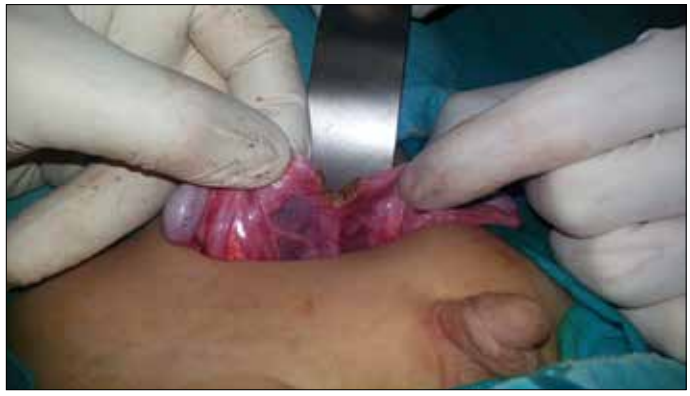

Figure 3. Utero-cervical splitting

patients whose vessels were ligated previously. One of these patients had previously undergone laparoscopic bilateral first stage SF, and the other one unilateral SF. Open second stage SF orchiopexy by Pfannenstiel incision was done in two patients who had their testicular vessels ligated laparoscopically. While UCS was applied to one of these patients (Figure 3), the testes with the TTE variant were lowered by passing through the same inguinal canal (right) by laparotomy and fixed without removing MR. One patient who had previously orchidopexy was treated with open single-session SF orchiopexy with UCS.

At a median follow-up of 2.5 years (range 6 months-6 years), all of testes were still in the scrotum, but unilateral testicular atrophy developed in two patients. Close follow-up of all patients still continues in our clinic.

Discussion

Three anatomic types of PMDS have been described.[5] The female type is bilateral intra-abdominal testes in a position analogous to ovaries. The male type, also called hernia uteri inguinale, is characterized by one testis along with the uterus and tubes in a hernia sac or scrotum. The other type is the least frequently encountered. Both the testes are located in the same hernial sac along with the Müllerian structures (TTE).[6] Three patients of this series were of female, and two of them were of male type. Our last patient was a rare form of TTE ie. a testicular fusion.

Although the diagnosis of PMDS is usually made by the appearance of abnormal internal genitalia during undescended testis or inguinal hernia surgery, it can also be diagnosed during imaging studies performed for the localization of the testis in the patients with non-palpable testis. Pelvic US is routinely used to describe the internal anatomy in patients with non-palpable testes. On the other hand, Steven et al.[7] found the sensitivity and specificity of US in localizing Mullerian structures as 54% and 50%, respectively, while US could not visualize Mullerian structures in the pelvis in two of five patients. In our series, in two patients US demonstrated that three testes were located intra-abdominally, but the MR could not reveal these intraabdominal testes. Gonads and PMDS could only be detected during laparoscopy. In fact, it is known that laparoscopy is very effective in identifying gonadal and pelvic structures in patients with PMDS.[7-9] We also think that laparoscopy is a better diagnostic method which demonstrates all internal genital structures especially in children with non-palpable testis.

When the disease is diagnosed definitely by experienced surgeons, the definitive procedures can be performed during the same anesthetic session after taking informed consent of the parents or the patients. But, generally, the procedures are postponed after taking intra-operative details (or photos) with or without gonadal biopsies.[10] In the majority of our cases with PMDS, the procedures, such as first- stage SF, biopsies, etc, were performed after the parents informed, but none of the procedures were completed in the same session.

The need of testicular biopsy to exclude mixed gonadal dysgenesis or carcinoma in situ is under debate. Since the biopsy may cause the antisperm antibodies to transgress the blood-testis barrier, it is stated that testis biopsy is unnecessary as long as the testis is placed in a palpable location and the testis can be followed-up with the US.[5] On the other hand, Patel et al.[11] have proposed that prepubertal testis biopsy does not induce production of antisperm antibodies. Testicular biopsy of the intra-abdominal testes was routinely performed in our series, and histopathologic examinations revealed the presence of testicular tissue without dysgenesis or malignancy.

After establishment of diagnosis, the aim of the treatment of PMDS patients is the prevention of the main complications as infertility and cancer. Patients with PMDS may have impaired gonadal function with normal testosterone release, however infertility is common. Whether infertility is caused by primary gonadal dysfunction or by undescended, and impaired testis is controversial. Since the fertility is rarely reported in these patients, it is recommended that every attempt should be made to place the testes into the scrotum.[4] In this series, after establishment of diagnosis, the goal of our management was to lower the intra-abdominal testes into the scrotum without disturbing their blood supply as much as possible leaving the MR in place, to protect the fertility potential, and to relieve the concerns of the parents about the gender identity of their child. This approach was attempted in all but one patient who underwent a staged orchiopexy, and all of his internal genitalia were totally removed for malignancy concern. Various open or laparoscopic procedures have been described to lower testes into scrotum, including staged SF, UCS in the midline, and microvascular auto-transplantation of the testis.[2,8,9] In our series, the first session of staged SF procedure was done in five patients. In the second session, as definitive surgery orchiopexy with UCS was conducted laparoscopically (n=2) or surgically (n=2). The UCS in the midline ensured the testicles to maintain blood supply, as well as allowing scrotal access into the testicles.

The primary operative consideration in PMDS patients is performing orchiopexy for especially cancer surveillance and preservation of potential fertility, or removing the testes if orchiopexy is not possible. Actually, the risk of testicular malignancy is accepted to be comparable to an undescended testis, and in addition to seminomas, mixed cell tumors and choriocarcinomas have been reported in the testes of PMDS patients.[12,13] On the other hand, it was seen that the overall incidence of malignancy in the testes of the patients with PMDS is 5-18%, which is similar to the incidence rate detected in otherwise healthy patients with intra-abdominal testes.[10]

Another issue concerning the PMDS patients is the removal of the MR. Although not absolutely essential; it is recommended that the MR can be removed if appropriate.[14] Salehi et al.[15] partially removed MR in only 2 out of 8 patients, and stated that these patients had a satisfactory overall prognosis; but their fertility potential appeared to be decreased, and risk of testicular malignancy due to cryptorchidism as well as a small

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risk of malignancy due to retained MR existed. Saleem et al.[16] didn't prefer bilateral orchidopexy with retention of MR or removal of MR along with testes and vas. Although their patients whose MRs were removed had not been followed-up in terms of testicular atrophy, to balance the chance of malignancy in the remnants and potential of fertility, removal of Mullerian structures and bilateral orchidopexy were preferred. But, the removal of the remnants without compromising the testes is often difficult. Vandersteen et al.[17] suggested that MR have no risk of malignancy because PMDS patients have no functioning ovarian tissue or do not produce cyclic estrogen. However, we have noticed that a large number of malignancies arising from MR have been reported with the review of the literature over the last fifteen years. Romero et al.[18] reported an adenocarcinoma arising from MR and suggested that this was only the second case until 2005. Manjunath et al.[5] have reviewed literature in 2010, and they found three cases (age range 14-67 years) of malignancies arising from the retained MR. They thought that these malignancies arised from the mucosa of MR, and proposed the destruction of the mucosal lining to reduce the risk of malignancy. A more comprehensive investigation concerning the current evidence on malignancy of MR was published by Farikullah et al.[10] in 2012. They reviewed approximately 200 reported PMDS cases over the last 50 years. Although the authors have not detected any malignancy arising from MR in their 8 patients, they found 11 cases aged between 4 and 68 years who had been diagnosed as clear cell carcinoma, squamous cell carcinoma and different types of adenocarcinoma arising from MR over the last 40 years. In four of these cases, diagnosis of PMDS had been overlooked at previous urological procedures, a patient with metastatic spread had been accepted as inoperable tumor and died, no recurrence was detected in four patients whose MR tumors had been resected during the follow-up period, and metastatic spread had occurred after removal of MRs in other cases. They assumed that cases of cancer are binomially distributed, and found that the risk of developing Müllerian malignancy ranged between 3.1% and 8.4% of males with PMDS. This increased malignancy rate in patients with intraabdominal residual MR cannot be ignored. However, it is not obvious whether this situation represents occurrence of the neoplasm arising from MR or an independent second neoplasm. In this situation, there is a need to develop new strategies to remove MR without compromising the testes. While it is still unclear what the management of MR should be, in our practice we left MR *in situ* during orchiopexy in 4 patients. Thus it is became uncertain how to approach to the patients who had been previously undergone orchiopexy with leaving MR *in situ*. Current management of the PMDS patients must consider the risks of malignancy and impairment of testicular blood supply. All of our patients are closely monitored by physical examination and US findings in terms of the risks of testicular and MR malignancy.

There are some limitations to our study. There is lack of long-term follow-up in our small series. All of the patients enrolled into the study were PMDS patients who underwent orchiopexy without removal of MR, so we need more studies involving case series with removed MRs so as to compare the results. Since this requires follow-up of 50-60 years to evaluate our management of PMDS, the 2.5 year follow-up in this small series does not provide additional information on this topic.

In conclusion, our main goal in approaching to the patients with PMDS is to descend the testes into the scrotum after establishment of diagnosis so as to ensure fertility potential and exclude risk of malignancy. Removal of MR may not be done in order to avoid the risk of vascular damage of the testes. However, new management strategies have to be developed as malignities developed from these remnants have been reported in recent years.

Ethics Committee Approval: Authors declared that the research was conducted according to the principles of the World Medical Association Declaration of Helsinki "Ethical Principles for Medical Research Involving Human Subjects", (amended in October 2013).

Informed Consent: Written informed consent was obtained from patients' parents who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - S.S., E.S., M.K.; Design - S.S., E.S., M.K.; Supervision - M.K.; Resources - S.S., E.S., M.K.; Materials - S.S.; Data Collection and/or Processing - S.S., M.K.; Analysis and/or Interpretation - S.S., E.S., M.K.; Literature Search - S.S., E.S., M.K.; Writing Manuscript - S.S., M.K.; Critical Review - E.S.

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Etik Komite Onayı: Yazarlar çalışmanın World Medical Association Declaration of Helsinki "Ethical Principles for Medical Research Involving Human Subjects", (amended in October 2013) prensiplerine uygun olarak yapıldığını beyan etmişlerdir.

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