DOI: https://doi.org/10.36811/gju.2022.110014 **GJU: September-2022: Page No: 01-06**

Global Journal of Urology

Research Article **Open Access**

What is to be done, how to proceed with PMDS?

Benslimane Hammou*

Department Pediatric Urology, Oran, Algeria

***Corresponding Author:** Benslimane Hammou, Department Pediatric Urology, Oran, Algeria, Tel: 213558239596; Email: benslimanehammou2019@gmail.com

Received Date: *Sep 06, 2022 /* ***Accepted Date:*** *Sep 21, 2022/* ***Published Date:*** *Sep 23, 2022*

Abstract

Aim: Disorders of sex development (DSD) affecting 1 birth in 10,000. Current management is very heterogeneous due to the small volume of patients on this subject. The series reported are short, often retrospective, and non-consensual support.

Material and Methods: Our study concerns the management of DSD patients admitted to the pediatric urology department since October 2015 until july2018. Paraclinical characteristics of the patients are recorded. All the patients included in the study underwent an ultrasound and laparoscopic exploration. The surgical management of Mullerian duct is done with laparoscopy procedure. Literature pubmed is reviewed. Data analyzed with spss software version 20.

Result and Discussion: 31 one patients, were identified from October 2015 to July 2018,The ultrasound and laparoscopy were succefull identified Mullerian duct in 5 cases (17%), in 16 cases (51%) they confirm the absence of Mullerian duct, in 10 cases there was a difference between the ultrasound and the laparoscopy , the ultrasound haven't identify the Mullerian duct in 10 cases (32%) , and streak gonads was not detected against the diagnostic laparoscopy is a excellent tool which allowed us all the identified mixed type or it was necessary to do a biopsy as reported by Yu and al. It is believed that the anatomical and histological data of diagnostic laparoscopy can be useful for the understanding the decision-making of parents. Recently, routine resection of Mullerian duct for patients without any symptoms is not carried out because the literature rarely describes the malignancy of the Mullerian duct. We proceeded to the division of the mullerian duct (uterus to the cervix) for the realization of the orchidopexy, and avoided damage the vas. Conclusion: all the clinical situation mentioned above, means that the surgeon must be familiar with the diagnosis of PMDS. Laparoscopy provide diagnosis of PMDS syndrome, and management without disturb blood supply of the testes, and the vas.

Keywords: Laparoscopy; Mullerian Duct; Mullerian Division

Cite this article as: Benslimane Hammou. 2022. What is to be done, how to proceed with PMDS?. Glob J Urol. 4: 01-06.

Introduction

Disorders of sex development (DSD) affecting 1 birth in 10,000 [1]. Current management is very heterogeneous due to the small volume of patients on this subject. The series reported are short, often retrospective, and non-consensual support.

DOI: https://doi.org/10.36811/gju.2022.110014 GJU: September-2022: Page No: 01-06

*Persistence of Mullerian duct syndrome (PMDS), is a rare form of DSD. It's can be seen either [2]:

- Intraoperative diagnosis in case of hernia repair in a boy normally with the finding of a fallopian tube or uterus when the hernial sac is opened.
- During a laparoscopy for non-palpable testis.

*During sexual differentiation, AMH is secreted by sertoli cells, causing the regression of the Müller's paramesonephrotic channels, between 8 and 10 GESTATIONAL WEEKS. This regression may be incomplete and remnants of these channels may persist: prostatic utricle, uterus, and/or fallopian tubes as well as 2/3 of the upper vagina.

*Various genetic studies have shown that this situation is due to mutations in the AMH hormone gene or mutations in its primary receptors [3]. This syndrome is transmitted in an autosomal recessive manner.

*This syndrome can be associated with other DSD: gonadal dysgenesis; 46, XY DSd with Mullerian duct or retro-vesical cavities. It is not recommended to have this cavity removed if the child is not symptomatic. These children may have febrile reccurent urinary tract infections, dysuria may indicate the removal of these cavity.

The laparoscopy surgery combined with urinary endoscopy is the best indication in the excision of these Mullerian cavities.

*The treatment is surgical and some authors recommend a dissection making it possible to separate the vas from the mullerian residues and to bring down the testis intra-scrotal and remove the residues [4,5]. This treatment is done by laparoscopy and will consist of a longitudinal section of the uterus and the cervix to allow to lower each testicle in the corresponding scrotum by an internal channel medial to the umbilical artery and preserving the vascularization of testis and the referent [6,7]. This is our plan towards PMDS syndrome.

Material and Methods

Study protocol:

*Type of study: This is a prospective descriptive study,
*Place of study: Pediatric urology department, Pediatric Hospital of Oran/Algeria.
*Study period: Our study concerns the management of DSD patients admitted to the service since October 2015 until July 2018.

Paraclinical characteristics of the population study:

Abdominal ultrasound: Ultrasound was indicated in all patients. The uterus and/or the fallopian tubes were not seen in 27 patients' vs. 24 patients (Figure 1).

DOI: https://doi.org/10.36811/gju.2022.110014 GJU: September-2022: Page No: 01-06

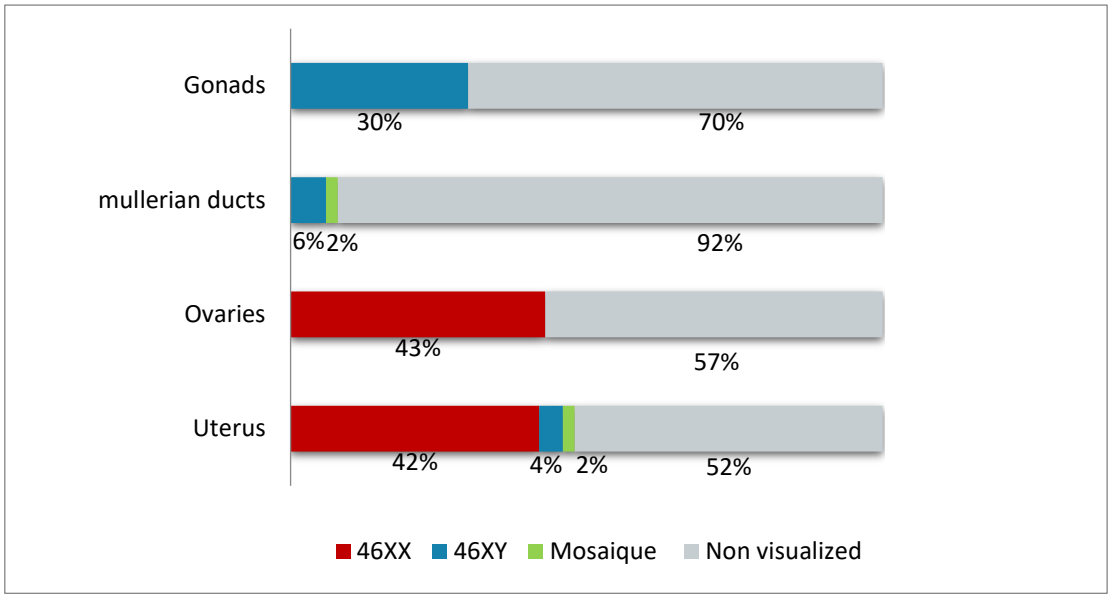

Figure 1: Distribution of organs visualized during abdominal ultrasound according to the karyotype of patients

-The ovaries were visualized in 22 patients (43%) of all the patients included in the series, all these patients are of karyotype 46 XX.
-The ultrasound has revealed Mullerian duct in 4 patients: 3 patients with karyotype 46 XX, 1 case with mosaic karyotype
-In 22 patients with karyotype 46 XX, an enlarged adrenal gland was demonstrated in 4 patients.
-Ultrasound allowed the visualization of the gonads in 15 patients, all of 46 XY karyotype.

Laparoscopy

-Laparoscopy was indicated in 30 patients (58.8%),
-The average age was 30 months +/- 21 months with extremes [1-72 months]
-The diagnostic laparoscopy involves the evaluation of the pelvic structures and the gonadal biopsy was indicated in all patients 46, XY and the mosaic karyotypes and was indicated in a case of DSD 46 XX with late diagnosis (Figure 2).

The presence of Mullerian duct: (Photo 1). The presence of Mullerian duct was diagnosed in 14 patients (46.7%)

DOI: https://doi.org/10.36811/gju.2022.110014 GJU: September-2022: Page No: 01-06

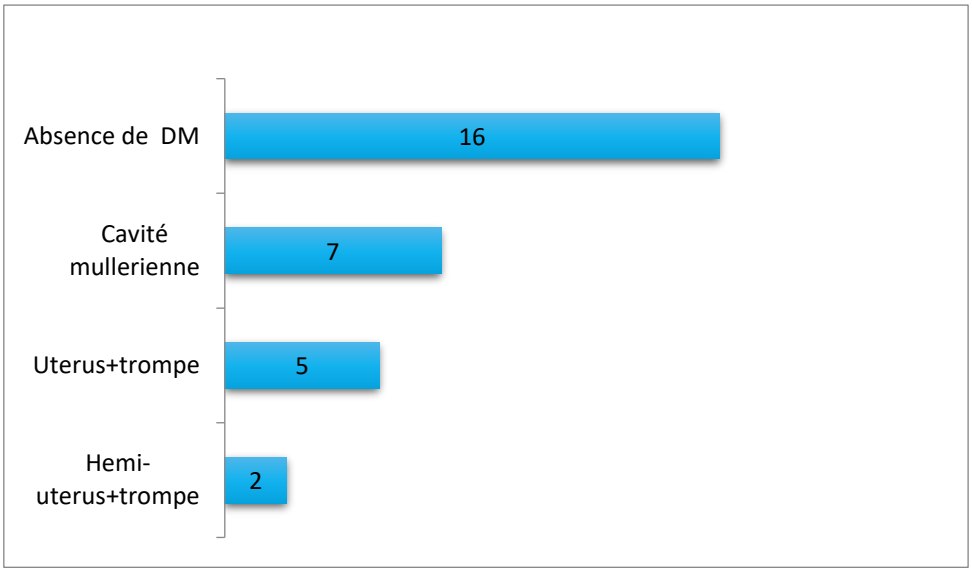

Figure 2: Result of diagnostic laparoscopy.

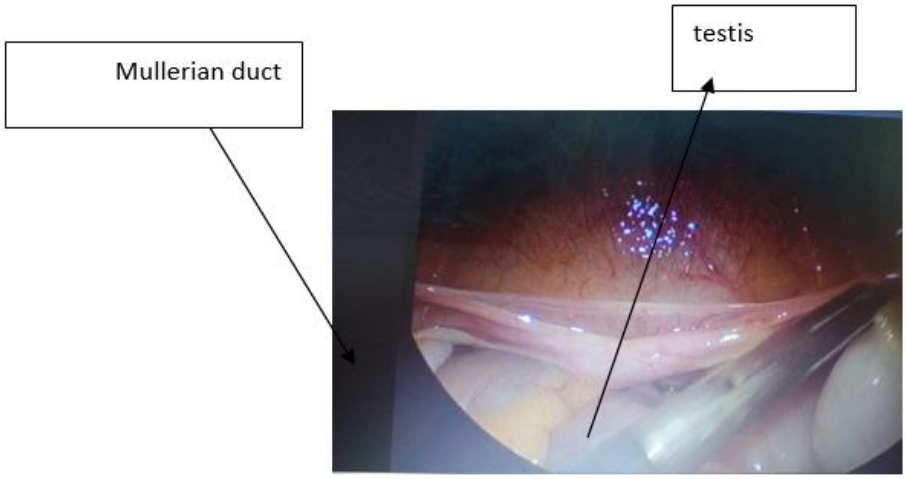

Photo 1: Mullerien Duct

Laparoscopy VS ultrasound in the evaluation of the internal genital organs

The aim of this study is to assess the degree of precision of laparoscopy VS ultrasound in the evaluation of Mullerianduct in the exploration of DSD patients. Using the SPSS software, we selected the patients who underwent at the same time an ultrasound and laparoscopic exploration, 31 patients were identified: 14 patients with androgen partial insensitivity syndrome, 2 cases of testicular dysgenesis, 4 cases of syndrome of persistence of Mullerian duct, 1 case of complete androgen insensitivity, 1 case of 5αreductase, hypogonadotropic hypogonadism in 3 cases, no diagnosis in 1 case; and 1 case of congenital adrenal hyperplasia, 2 cases of mixed gonadal dysgenesis with karyotype 45 xo / 46xy, and 1 case of mosaic 48xxyy / 46xy. A DSD ovotestis with 46xx / xy karyotype. All patients underwent ultrasound exploration prior to laparoscopic exploration. The two methods identified Mullerian duct in 5 cases (17%), in

DOI: https://doi.org/10.36811/gju.2022.110014 GJU: September-2022: Page No: 01-06

16 cases (51%) the confirm the absence of Mullerian duct, in 10 cases there was a difference between the ultrasound and the laparoscopy, the ultrasound haven't identify the Mullerian duct in 10 cases (32%)

Coelio OUI 15 NON 16
ECHO 5 26

With a significant 0.047 KHI-two test (less than 0.05).

1. The role of laparoscopy in the evaluation of the intra-abdominal gonads has been proven and the precision regarding the macroscopic aspect and the identification is clearly higher than ultrasound.
2. In our series, laparoscopy identified 26 intra-abdominal testes and, Vs 25 gonads on ultrasound without visualization of streak gonads.
3. Laparoscopy allows the visualization of 6 streak gonads including 1 ovotestis VS none on ultrasound
4. Division of Mullerian duct done in 5 patients.

Diagnoses	Case Number	Orchdopexy	Gonadectomy	Division of mullerian duct	Trocard Number
PIAS	6	7	1	1	3
MGD	2	1	3		3
PMDS	4	5	3	4	3
OVOTESTIS	2		2		
Partial Gonadal dysgenesic	1		2		3
Hypogonadisme hypogonadotrope	2	4			3

Discussion

In our study regarding the mullerian duct, the ultrasound did not succeed in identifying mullerian duct in 10 cases, and streak gonads was not detected against the diagnostic laparoscopy is a excellent tool which allowed us all the identified mixed type or it was necessary to do a biopsy as reported by Yu et al. [8]. The impact of diagnostic laparoscopy: DSD being a complex abnormality, it is very hard for parents to understand the anomalies and to make decisions for sex assignment, especially situation. From this point of view, it is believed that the anatomical and histological data of diagnostic laparoscopy can be useful for the understanding the decision-making of parents, as well as the information provided by genetic and endocrinological exam. According to the recommendations of the Consensus [9] and the ESPU / SPU point of view on the surgical management of sexual development disorders [10] stated that in patients with a symptomatic utricle, resection is preferably done by laparoscopy and that these Mullerian remnants can also be removed in boys if they cause a urinary tract infection, dysuria or gynecological (menstruation)", Recently, routine resection of Mullerian duct for patients without any symptoms is not carried out because the literature rarely describes the malignancy of derivatives of the Mullerian canal [11]. Consequently, in the previous cases, we proceeded to the division of the mullerian duct (uterus to the cervix) for the realization of the orchidopexy, and avoided damage the vas, and all the laparoscopic surgeries were completed without conversion to laparotomy, without significant intraoperative complications.

Conclusion

All the clinical situation mentioned above, means that the surgeon must be familiar with the diagnosis of PMDS. Laparoscopy provide diagnosis of PMDS syndrome, and management without disturb blood supply of the testes, and the vas.

DOI: https://doi.org/10.36811/gju.2022.110014 GJU: September-2022: Page No: 01-06

References

1. Woodward M. 2010. Disorders of sex development. Surg Oxf. 28: 396-401. Ref.: https://pubmed.ncbi.nlm.nih.gov/10690872/ DOI: https://doi.org/10.1210/jcem.85.2.6337
2. SF Ahmed, Cheng A, Dovey L, et al. 2000. Phenotypic Features, Androgen Receptor Binding, and Mutational Analysis in 278 Clinical Cases Reported as Androgen Insensitivity Syndrome. J Clin Endocrinol Metab. 85: 658-665. Ref.: https://pubmed.ncbi.nlm.nih.gov/10690872/ DOI: https://doi.org/10.1210/jcem.85.2.6337
3. Moore K, Persaud T. 2008. The developing humen, clinicalorientated embryology. 263-269.
4. Wongprasert, Somanunt S, De Filippo R. 2013. Novel mutation of anti-mullerian gene in persistant mullerian duct, presented with bilateral cryptorchidisme. A case report. J Ped Urology. 9: 147-149. Ref.: https://pubmed.ncbi.nlm.nih.gov/23611722/ DOI: https://doi.org/10.1016/j.jpurol.2013.03.004
5. Warmann Y, Vogel M. 2006. Giant mullerian duct cyst with malignant transformation in 15 years boy. Urology. 67: 424-426. Ref.: https://pubmed.ncbi.nlm.nih.gov/16461114/ DOI: https://doi.org/10.1016/j.urology.2005.09.009
6. Shinmura Y, Yokoi T. 2002. A case of clear cell carcinoma in persistant mullerian duct. Am J Surg Pathol. 73: 81-83. Ref.: https://pubmed.ncbi.nlm.nih.gov/12218580/ DOI: https://doi.org/10.1097/00000478-200209000-00014
7. Al Gohary Ma. 2003. Laparoscopy management of persistant mullerian duct syndrome. Pediatric Surg Int. 19: 533-536. Ref.: https://pubmed.ncbi.nlm.nih.gov/13680290/ DOI: https://doi.org/10.1007/s00383-003-0984-7
8. Raval C, Chantot B. 2000. Aspect moléculaire du déterminisme sexuel régulation génétique et pathologie. Encyclopédie Médico-chirurgicale de Gynécologie obstétrique et fertilité. 32 : 584-594. Ref.: https://pubmed.ncbi.nlm.nih.gov/15450256/ DOI: https://doi.org/10.1016/j.gyobfe.2004.06.003
9. Kallen B, Finnstorm O. 2002. Congenital malformation in infant born after in vitro fertilization in Sweden. Cinical Mol Teratol. 88: 137-143. Ref.: https://pubmed.ncbi.nlm.nih.gov/20063307/ DOI: https://doi.org/10.1002/bdra.20645
10. Charmichael SL, Shaw GL. Maternal progestin intake and risk of hypospadias. Archive pediatric. 159: 957. Ref.: https://pubmed.ncbi.nlm.nih.gov/16203941/ DOI: https://doi.org/10.1001/archpedi.159.10.957
11. Steven M, O'Toole S, Lam JP, et al. 2012. Laparoscopy versus ultrasonography for the evaluation of Mullerian structures in children with complex disorders of sex development. Pediatr Surg Int. 28: 1161. Ref.: https://pubmed.ncbi.nlm.nih.gov/23064803/ DOI: https://doi.org/10.1007/s00383-012-3178-3