



## How I do it

## A novel approach in the intraoperative management of ovotesticular DSD

Xavier Delforge<sup>a,\*</sup>, Cécile Brachet<sup>b</sup>, Nasroola Damry<sup>c</sup>,  
Valérie Segers<sup>d</sup>, Stéphane Luyckx<sup>a</sup>, Claudine Heinrichs<sup>b</sup>,  
Karim Khelif<sup>a</sup>

<sup>a</sup>Department of Pediatric Urology, Hôpital Universitaire des Enfants Reine Fabiola, ULB, Brussels, Belgium

<sup>b</sup>Department of Pediatric Endocrinology, Hôpital Universitaire des Enfants Reine Fabiola, ULB, Brussels, Belgium

<sup>c</sup>Department of Radiology, Hôpital Universitaire des Enfants Reine Fabiola, ULB, Brussels, Belgium

<sup>d</sup>Department of Pathology, Hôpital Brugmann, Université Libre de Bruxelles, ULB, Brussels, Belgium

\* Corresponding author. Xavier Delforge, Department of Pediatric Surgery and Urology, Amiens University Hospital, Rond-point Professeur Cabrol, 80054 Amiens cedex 1, France. Tel.: +33 322087560; fax +33 322089741.  
[delforge.xavier@chu-amiens.fr](mailto:delforge.xavier@chu-amiens.fr), [xdelforge@gmail.com](mailto:xdelforge@gmail.com) (X. Delforge)

**Keywords**

Ovotestis; DSD; Minimal invasive surgery; Gonadal separation; Laparoscopy

**Abbreviations**

Ovotesticular Differences in Sex Development, OTD; Ultrasonography, US

Received 30 June 2020  
Revised 17 September 2020  
Accepted 6 October 2020  
Available online 15 October 2020

**Summary**

Ovotesticular disorder of sex development (OTD) management remains challenging. In OTD, cautious gonadal evaluation and separation of ovarian and testicular components might be required to avoid virilization of a patient with female identity. Herein we report our minimal invasive approach in this very rare condition. The gonads are externalized under

laparoscopic control through trocar openings.

Intraoperative ovotesticular ultrasonography (US) is used for clear identification of ovarian and testicular tissue which can then be safely separated. We strongly promote the use of a minimal invasive approach in the management of these patients undergoing long term treatment and often multiple procedures.

This report describes an innovative minimal-invasive approach of ovotesticular disorder of sex development (OTD), combined with intraoperative gonadal ultrasonography (US) for guiding testicular component removal.

OTD is characterized by the presence of both ovarian and testicular tissue in the same individual. Different histological types are described: bipolar ovotestis (11%) and mixed types (89%). The phenotype is highly variable. 46, XX is the most common karyotype [1].

Gender assignment and timing of surgery rely on a multidisciplinary management centered on the patient and his family [2]. In most OTD cases, the ovarian part of the gonad is functional, with reported live births, while the production of testosterone by the testicular part decreases after puberty, and spermatogenesis is impossible in case of XX karyotype [3]. While germ cell tumor can occur in OTD, preventive removal of the testis is not indicated [4]. To date, no germ cell tumor has been reported in OTD patients with XX karyotype. However, the testicular tissue induces virilization throughout mini-puberty and puberty, that needs to be prevented in case of female gender assignment. Among the treatment options, the injection of GnRH agonist has been suggested. This medical treatment puts a brake on both testicular and ovarian components, and is reversible. The long-term effects of such a medical castration

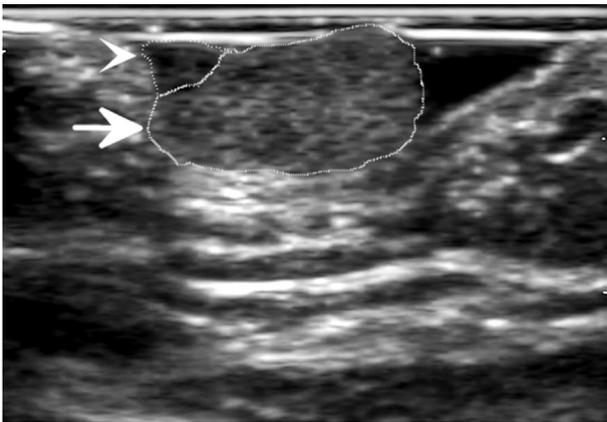
are barely known, and the patient will require hormonal treatment for pubertal induction. Another way to avoid the virilization is the surgical removal of the testicular component. This option is irreversible and needs to be carried out once the patient is able to participate in the shared-decision process. An intermediate solution consisting of a medical treatment during mini-puberty and a surgical treatment around puberty might be suggested.

Patients with DSD require long term follow-up and potentially repeated surgical procedures. Diagnostic laparoscopy in DSD is used since the 90's [3], while surgical treatment is often performed by laparotomy. We present here a mini-invasive approach, by externalizing the gonad through 5 mm trocar incisions (Fig. 1). While a complete intraabdominal procedure is possible [5], our approach offers the advantage of direct gonadal palpation coupled with US application [2].

The ability to identify and separate gonadal tissues plays a major part in the surgical decision-making process. Classically, partial resection is guided by frozen sections [3]. Multiple gonadal biopsies are required with sometimes loss of a considerable amount of valuable tissue. Our strategy is believed to be tissue sparing. With the help of intra operative US using a high frequency linear probe, performed by an experienced senior radiologist, testicular and ovarian parts of the ovotestis



**Fig. 1** Ovotestis externalized through the suprapubic trocar opening (intraoperative view). The arrow indicates the head (H).



**Fig. 2** Intra operative US showing ovarian tissue with follicles (arrowhead) and testicular tissue (arrow).

are easily identified (Fig. 2). Hence, gonadal separation is guided by careful US margin delineation and confirmed by frozen section (Fig. 3). Compared to the classical management, our approach is promising regarding tissue sparing, operative time and hospital stay reductions. Although studying series is unlikely in this rare condition, further cases are needed to confirm our results. Our patient had spontaneous puberty and began menstruations at the age of 12.3 years. Long term follow up is mandatory to monitor the gonadal growth and the occurrence of tumor.

**Funding**

None.

**Conflicts of interest**

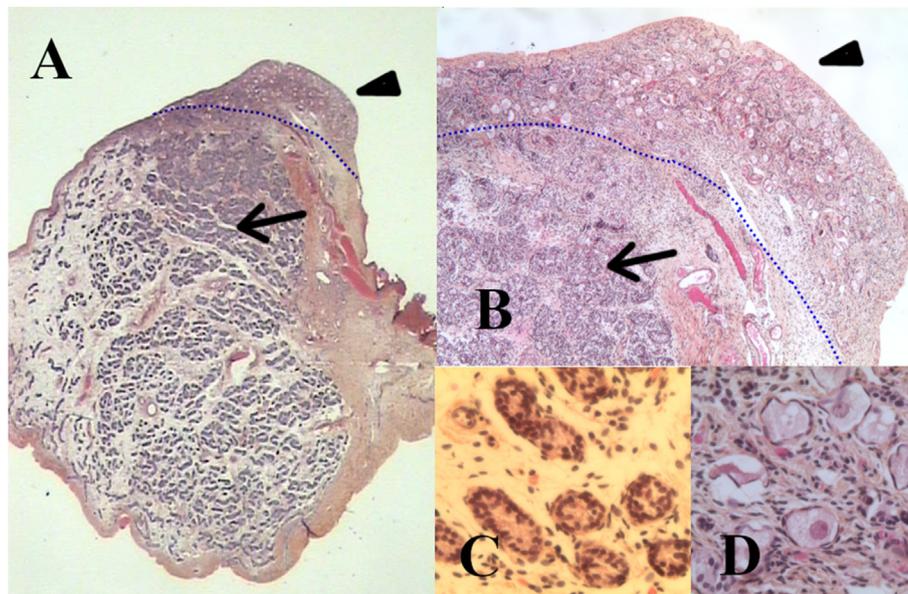
None.

**References**

[1] Berger-Zaslav A-L, Mehta L, Jacob J, Mercado T, Gadi I, Tepperberg JH, et al. Ovotesticular disorder of sexual development (true hermaphroditism). *Urology* 2009;73:293–6. <https://doi.org/10.1016/j.urology.2008.08.428>.

[2] Lee PA, Houk CP, Ahmed SF, Hughes IA. In collaboration with the participants in the international consensus conference on intersex organized by the Lawson Wilkins pediatric endocrine society and the European society for paediatric Endocrinology. Consensus statement on management of intersex disorders. *Pediatrics* 2006; 118:e488–500. <https://doi.org/10.1542/peds.2006-0738>.

[3] Verkauskas G, Jaubert F, Lortat-Jacob S, Malan V, Thibaud E, Nihoul-Fékété C. The long-term followup of 33 cases of true hermaphroditism: a 40-year experience with conservative gonadal surgery. *J Urol* 2007;177:726–31. <https://doi.org/10.1016/j.juro.2006.10.003>.



**Fig. 3** Histological aspect with Hematoxylin-Eosin stain of the ovotestis, showing both ovarian (arrowhead) and testicular (arrow) tissues, at magnification x1.6 (A) and x6 (B). Ovarian follicles (C), and seminiferous tubules with paucity of Leydig cells (D), at x40.

- [4] Looijenga LHJ, Hersmus R, de Leeuw BHCGM, Stoop H, Cools M, Oosterhuis JW, et al. Gonadal tumours and DSD. *Best Pract Res Clin Endocrinol Metab* 2010;24:291–310. <https://doi.org/10.1016/j.beem.2009.10.002>.
- [5] Moriya K, Morita K, Mitsui T, Kitta T, Nakamura M, Kon M, et al. Impact of laparoscopy for diagnosis and treatment in patients with disorders of sex development. *J Pediatr Urol* 2014;10: 955–61. <https://doi.org/10.1016/j.jpuro.2014.03.006>.