



## Letter to the Editor

### Re: Perineal hypospadias repair with preservation of a coincidental vagina or perineal utricle in boys with disorders of sex development



gonad showed a dysgenetic testis with a mixed pattern of seminiferous tubules and fragmentary areas of undifferentiated gonadal tissue. Evaluation of the left gonad clearly showed the required components for the diagnosis of an ovary, namely, primordial follicles, confirming the diagnosis of ovotesticular disorder of sex development (DSD) in our case.

We thank the authors for their interest and comments about our article on hypospadias repair while retaining an accidental perineal utricle or vagina.

We agree that the embryology of enlarged utricles and variations thereof is complex. The appropriate nomenclature for these often asymptomatic entities has indeed long been debated. However, given the current social developments, in particular the broad acceptance of the concept of gender fluidity, we are not in favor of the use of dichotomous nomenclature such as 'male vagina.' We proposed to distinguish the perineal utricle and vagina; of which, only the latter has a connection with the cervix and/or uterus. As outlined in the discussion section, the distinction between the enlarged utricle and vagina is especially important in the event of symptoms because of the difference in the surgical approach.

Regarding your comment on the diagnosis of our fourth patient, we agree that the evaluation of dysgenetic gonads by the pathologist can be difficult. An experienced pathologist and the application of immunohistochemistry, in particular germ cell markers (e.g., OCT3/4, TSPY, DDX4), Sertoli cell (SOX9), and granulosa cell markers (FOXL2), are important for an accurate diagnosis [1,2]. In our fourth case with a complex mosaic karyotype (45,XY in blood and 45,X/46,XX/46,XY in gonadal tissue), biopsy of the right

## References

- [1] Wolffenbuttel KP, Hersmus R, Stoop H, Biermann K, Hoebcke P, Cools M, et al. Gonadal dysgenesis in disorders of sex development: diagnosis and surgical management. *J Pediatr Urol* 2016;12:411–6.
- [2] Spoor JA, Oosterhuis JW, Hersmus R, Biermann K, Wolffenbuttel KP, Cools M, et al. Histological assessment of gonads in DSD: relevance for clinical management. *Sex Dev* 2018; 12:106–22.

Katja Wolffenbuttel\*  
*Erasmus MC-Sophia Children's Hospital,  
Department of Urology and Pediatric Urology,  
University Medical Center, Rotterdam, the  
Netherlands*

Gundela Holmdahl  
*Queen Silvia's Children's Hospital,  
Department of Pediatric Surgery, Sahlgrenska  
Academy Women's and Children's Health,  
Gothenburg, Sweden*

\*Corresponding author.  
E-mail address: [k.wolffenbuttel@erasmusmc.nl](mailto:k.wolffenbuttel@erasmusmc.nl) (K. Wolffenbuttel)

13 January 2020  
Available online 22 January 2020