Congenital Uro-Nephropathy: Beyond the Urinary Tract

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A 3-year-old female was admitted to the pediatric emergency department with vulvar pain and foul-smelling vaginal discharge for three days. Fever and urinary symptoms were denied. On inspection, vulvar erythema and vaginal introitus swelling were observed. Past medical history included a neonatal diagnosis of left ureteral duplication with ureterocele and nonfunctioning upper kidney pole segment. This congenital ureterovesical junction obstruction led her to a cystoscopy at 3 months with spontaneous complete ureterocele drainage. Following the procedure, persistent obstruction and upper kidney pole function exclusion, with a high risk of infection, led her to a left upper uretero-heminephrectomy at 6 months. After surgery, several urinary tract infections with concomitant abnormal vaginal discharge occurred, despite normal renal scintigraphy and isotope cystography. Family history included a cousin with a complex congenital uro-nephropathy. At the emergency department, a urinary tract infection was excluded. Pelvic ultrasound showed a bifid uterus and two vaginal canals, with a left cavity and ipsilateral vaginal canal distended by echogenic fluid (Fig. 1). The symptoms ended up spontaneously resolving in one day. For further detailed anatomic assessment, pelvic magnetic resonance imaging was performed, revealing a complete *bicornis bicollis* uterus, with uterine laps fusion and vaginal canal duplicity, which is compatible with a U3bC2V2 congenital gynecological malformation (Figs. 2 and 3).[1] Currently, she remains under follow-up and surgical approaches such as longitudinal septum section or hysterectomy of the less functional uterus cavity have been considered, which are to be performed by the time of puberty.

The interaction between mesonephric and paramesonephric ducts explains the known association between malformations of the urinary and reproductive tracts.[2-4] *Bicornis bicollis* uterus with vaginal canal duplicity is a rare congenital disorder that can be related to several urinary tract abnormalities.[3-5] It should be differentiated from a septate uterus, which has a normal outline and an internal indentation at the fundal midline resulting in two closely separated endometrial cavities. By contrast, *bicornis* uterus has an abnormal fundal outline with an external indentation at the fundal midline that results in two moderately separated endometrial cavities.[1] This congenital malformation is often asymptomatic before puberty and may cause cyclic pelvic pain starting upon menarche onset.[5] Complications include recurrent infections, pelvic adherences, and endometriosis.[4] The atypical presentation in this case allowed an early diagnosis and the chance of planning the best time for intervention.

Like some other authors, we recommend a genital tract ultrasound evaluation in all prepubertal girls with known urinary abnormalities to anticipate therapeutic measures in order to prevent complications later on.[4]

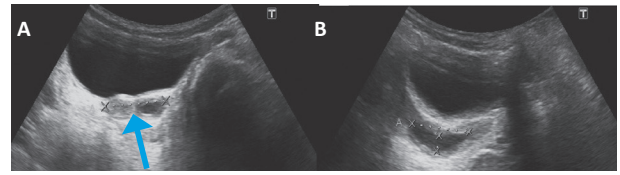

Figure 1. Transabdominal pelvic ultrasound. Transverse (A) and longitudinal (B) transabdominal pelvic ultrasound images showing a duplicated uterus, with two hemi-uteri separated by a thin fundal cleft (arrow). A slightly distended left side endometrial cavity with echogenic content (B).

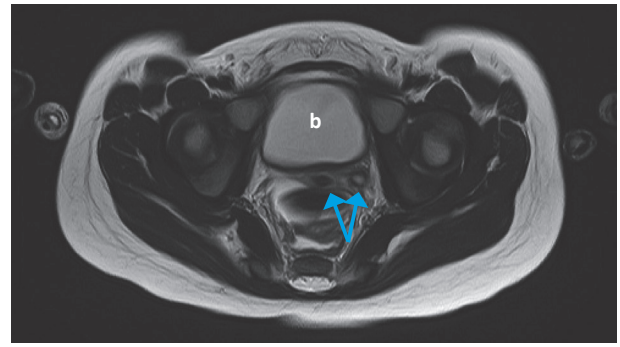

Figure 2. Pelvic magnetic resonance imaging. Axial T1-weighted MR images demonstrate behind the urinary bladder (b) two separate uteri with non-communicating endometrial cavities (arrows).

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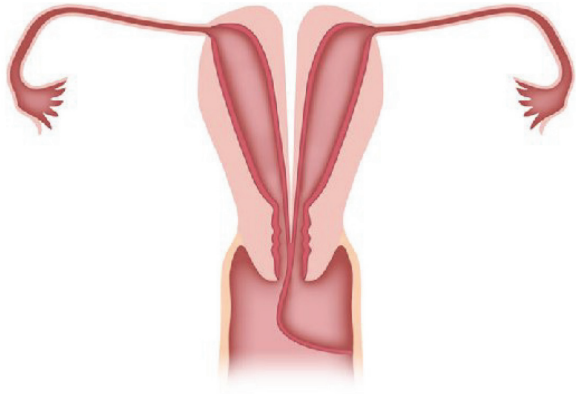

Figure 3. Schematic representation of U3bC2V2 congenital gynecological malformation.
From: Yoo R, Cho JY, Kim SY, Kim SH. A systematic approach to the magnetic resonance imaging-based differential diagnosis of congenital Mullerian duct anomalies and their mimics. Abdom Imaging 2014. DOI: 10.1007/s00261-014-0195-9.

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WHAT THIS REPORT ADDS

- A congenital uro-nephropathy must alert physicians to look for associated reproductive anomalies.
- A pelvic ultrasound should be performed in all prepubertal girls with known complex urinary tract abnormalities to rule out reproductive malformations.
- Early detection of a *bicornis bicollis* uterus allows for anticipating therapeutic measures to prevent recurrent symptoms during adolescence and avoid complications later on.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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