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## Genitourinary

# Contrast enhanced genitosonography (CEGS) of urogenital sinus: A case of improved conspicuity with image inversion

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### ABSTRACT

Imaging plays a pivotal role in evaluating the urogenital anatomy in children with ambiguous genitalia. Contrast enhanced genitosonography (CEGS) represents a low-cost and radiation-free alternative to the traditional techniques of fluoroscopic genitography and magnetic resonance imaging. We report a case of a child with ambiguous genitalia whose urogenital sinus anomaly is clearly demonstrated using CEGS and the postprocessing technique of image inversion. This case report shows the utility of CEGS in this patient population and the enhanced conspicuity with image inversion.

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## Introduction

Imaging plays an important role in evaluating urogenital anatomy in children with ambiguous genitalia. Fluoroscopic genitography and magnetic resonance (MR) imaging are both used to evaluate pelvic structures and their connections and help to define important markers for surgical management [1]. Conventional fluoroscopic genitography provides a negative image of the pelvic structures as the body cavities fill with contrast material and entails ionizing radiation. MR imaging has lower resolution compared to ultrasound (US) and commonly requires sedation or anesthesia at additional cost and risk. Contrast enhanced genitosonography

(CEGS) can detail anatomy including genitourinary communications at high resolution without sedation or radiation exposure.

US contrast material is composed of microbubbles—encapsulated, inert gas filled bubbles 1–5 μm in diameter—that enhance the reflection of sound. Intravenous or intracavitary administration of US contrast agents increases the sonographic contrast between the target and surrounding tissues. It has previously been shown that contrast enhanced voiding urosonography and genitosonography help delineate urogenital sinus (UGS), urethrovesical fistula, and internal duct structures [2]. In this report we describe using CEGS with post-processing image inversion to further delineate the detailed anatomy of a child with ambiguous genitalia.

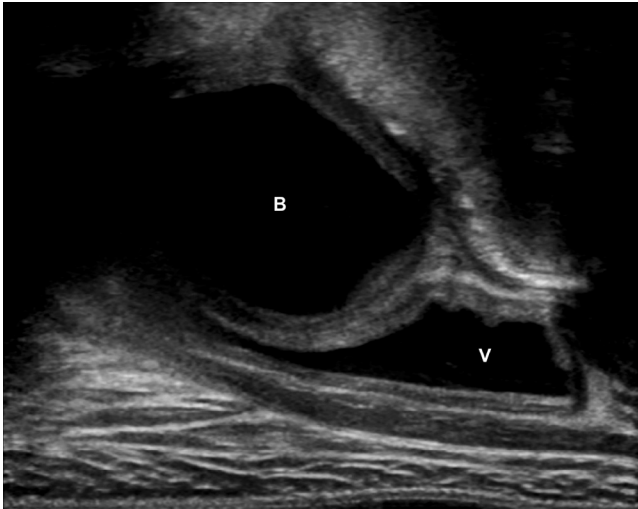
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**Fig. 1 – Sagittal transabdominal grayscale ultrasound of the pelvis demonstrates a partially distended urinary bladder and fluid within the vagina posteriorly. B, bladder; V, vagina.**

### Case report

A 5-week-old female born at 34 weeks gestation had ambiguous genitalia identified at birth. She was seen initially by our endocrinology service with laboratory findings consistent with congenital adrenal hyperplasia. Her initial pelvic US showed a normal prepubertal uterus and fluid distended vagina. Her ovaries contained multiple variable-sized follicles. A renal bladder US performed at 6 months of age demonstrated normal size kidneys with mild increased cortical echogenicity, and several punctate echogenic foci in the renal pyramids of each kidney. No adrenal abnormality was detected at the time of the renal bladder US.

CEGS was performed to better assess the suspected UGS. Transabdominal grayscale ultrasound of the pelvis demonstrated a partially distended urinary bladder and the tubular fluid-filled vagina (Fig. 1). Using sterile technique, a transurethral catheter was placed into the bladder, which was drained. Sixty milliliter of a 0.1% Lumason (Bracco Milan, Italy) normal saline solution was then instilled under sonographic observation during bladder filling and voiding. The patient tolerated the study without event.

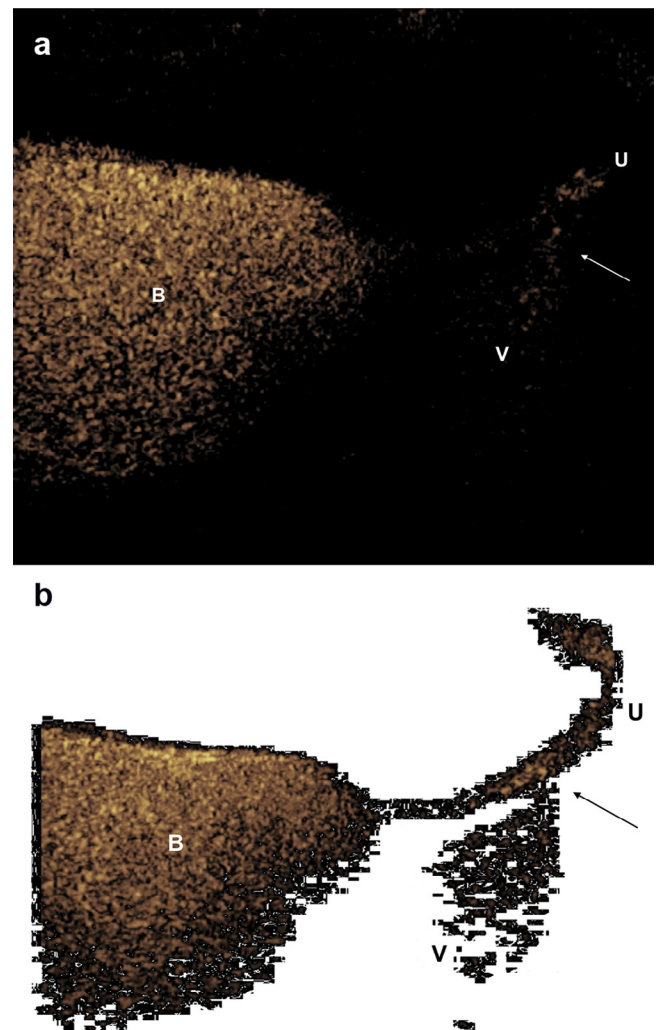
A high connection between the urethra and vagina was noted with dilute contrast appearing within the vagina (Fig. 2A). The urethra was relatively long and compatible with male urethral morphology. No vesicourethral reflux was visualized into the renal collecting system. The US images were transferred to the picture archiving and communication system and the images were inverted making the microbubbles and urovaginal fistula even more conspicuous (Fig. 2B). Immediately following the CEGS using the same transurethral catheter a fluoroscopic genitogram was performed for direct comparison. Both studies demonstrated the same findings (Fig. 3).

With the aforementioned findings, the patient underwent cystoscopy which showed there was about a 2.5 cm distance

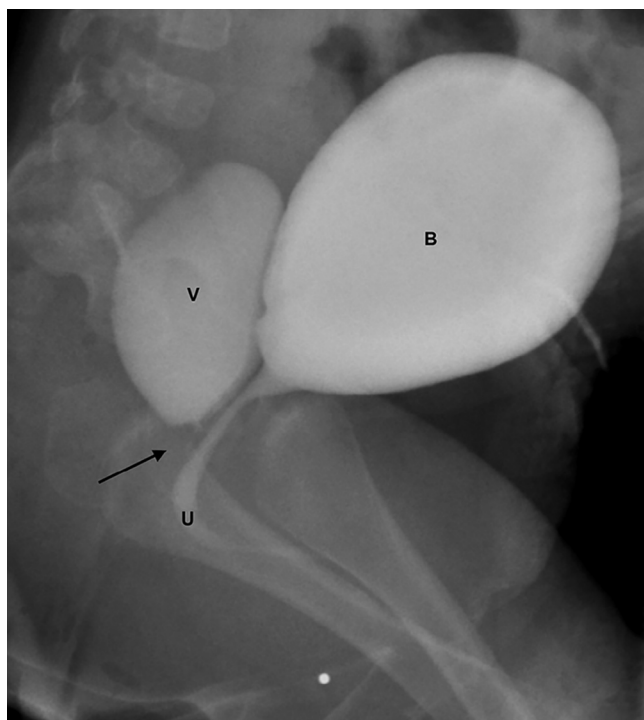
from the beginning of the common urogenital sinus to where the vaginal takeoff was and there was about a 1.5 cm distance between the takeoff of the vagina and the bladder neck. To repair the high UGS a modified anterior sagittal transrectal approach was performed without complication.

### Discussion

Disorders of Sexual Development (DSD) encompass a large group of congenital conditions in which there is abnormal development of chromosomal, gonadal, or anatomic sex [3]. When the external genitalia do not have normal anatomic male or



**Fig. 2 – (A) Sagittal transabdominal contrast enhanced genitosonography. The contrast enhanced image demonstrates accumulation of dilute contrast solution within the vagina which is less concentrated compared with the contrast solution filling the urinary bladder. The arrow depicts communication between the urethra and the vagina. (B) Image inversion performed at the picture archiving and communication system workstation. This depicts a more conspicuous connection (arrow) between the vagina and urethra. B, bladder; U, urethra; V, vagina.**



**Fig. 3 – Left lateral decubitus fluoroscopic image from conventional genitography confirms a urogenital sinus with high communication (arrow) of the vagina to the proximal urethra. Radiopaque marker is present on the perineum. B, bladder; U, urethra; V, vagina.**

female appearance, the condition is known as ambiguous genitalia. The evaluation and initial management of the neonate with ambiguous genitalia must be regarded as a medical and psychosocial emergency and be handled with great sensitivity towards the family. The goal of this initial evaluation is to make a precise diagnosis of the disorder and, with the involvement of the parents, to assign a sex of rearing based on the diagnosis, the status of the child's anatomy, and the functional potential of the genitalia and reproductive tract [4].

UGS abnormalities are most often seen in DSD, commonly in association with congenital adrenal hyperplasia, with an incidence as high as 1 in 500 in the nonclassic, mild form [5]. Critical details of the anatomy that must be defined include the length of the common UGS, location of the vaginal confluence and its proximity to the bladder neck, the size and number of vaginas, the presence of a cervix, and bladder and urethral anatomy [4]. The ability to delineate these anatomical

characteristics is important for surgical planning. Low UGS can be successfully treated with a simple flap vaginoplasty whereas a high UGS can be challenging [6].

Traditionally, fluoroscopic genitography has been used to demonstrate the exact urethral configuration and any fistulous communication in patients with DSD. However, the procedure of conventional fluoroscopic genitography exposes the patient to ionizing radiation and is only able to visualize the inner contour of the genital tract. Contrast enhanced genitosonography represents a promising alternative to conventional fluoroscopic genitography. Although CEGS also requires placement of a bladder catheter, there is sonographic visualization of structures in proximity to the genital tract and it is free of ionizing radiation. To our knowledge there has only been 1 case in the literature describing the use of contrast enhanced genitosonography in the evaluation of ambiguous genitalia. The current report further builds on these findings and introduces the use of a postprocessing technique, image inversion, to enhance conspicuity of the ultrasound findings. We have since performed CEGS on a patient with UGS anomaly with similar success.

#### REFERENCES

- [1] Chavhan GB, Parra DA, Oudjhane K, Miller SF, Babyn PS, Pippi Salle FL. Imaging of ambiguous genitalia: classification and diagnostic approach. *Radiographics* 2008;28(7):1891–904. doi:10.1148/rg.287085034.
- [2] Kopac M, Riccabona M, Haim M. Contrast-enhanced voiding urosonography and genitography in a baby with ambiguous genitalia and urogenital sinus. *Ultraschall Med* 2009;30(3):299–300. doi:10.1055/s-0028-1109353.
- [3] McNamara ER, Swartz JM, Diamond DA. Initial management of disorders of sex development in Newborns. *Urology* 2017;101:1–8. doi:10.1016/j.urology.2016.08.010.
- [4] Rink R. Surgical Management of Disorders of Sex Development and Cloacal and Anorectal Malformations. In: *Campbell-Walsh Urology*. 11th ed. Elsevier; 2015. <<https://www.clinicalkey.com/#!/content/book/3-s2.0-B9781455775675001515?scrollTo=%23hl0000379>>; [accessed August 18, 2017].
- [5] Hughes IA. Congenital adrenal hyperplasia—a continuum of disorders. *Lancet* 1998;352(9130):752–4. doi:10.1016/S0140-6736(98)22037-X.
- [6] Pippi Salle JL, Lorenzo AJ, Jesus LE, Leslie B, AlSaid A, Macedo FN, et al. Surgical treatment of high urogenital sinuses using the anterior sagittal transrectal approach: a useful strategy to optimize exposure and outcomes. *J Urol* 2012;187(3):1024–31. doi:10.1016/j.juro.2011.10.162.