Invited Review Article

3D Fluoroscopic imaging facilitates reconstruction of common urogenital sinus and cloacal anomalies

Yi Li, Laurence S. Baskin

Department of Urology, University of California San Francisco, San Francisco, CA, USA

Keywords:
Urogenital sinus, persistent cloaca, surgical planning, three dimensional imaging, three dimensional rotational fluoroscopy

Abstract

Cloacal anomalies and common urogenital sinus are rare structural abnormalities in hindgut and urogenital development. Surgical correction in childhood is often indicated to create normal external genital anatomy, allow for adequate bladder and vaginal drainage, and create the appropriate anorectal opening in patients with persistent cloaca.

Understanding the anatomy and relationships between the pelvic organs is critical as there is a drastic variation in potential surgical approaches to the repair. Traditional imaging modalities such as pelvic ultrasound, magnetic resonance imaging, and two-dimensional fluoroscopic imaging have been utilized to delineate the pelvic anatomy for facilitation of surgical planning. Limitations to these modalities include the inability to adequately dilate structures and the difficulty in identifying the common confluence, or where the structures ultimately coalesce within the pelvis.

In this article we describe the utilization of three-dimensional rotational fluoroscopy in combination with examination under anesthesia to provide optimal clarity of anatomy. Examination under anesthesia, specifically cystoscopy and vaginoscopy, helps the surgeon to visualize the anatomy and to place catheters in the correct lumens. Contrast material can then be injected into the catheters to dilate the bladder, vagina, and mucous fistula for fluoroscopic imaging. The rotational images can then be reconstructed in three dimensions to create a roadmap for the surgeons, providing accurate description of the location of the confluence, distance to the introitus and other critical measurements.

We believe that three-dimensional rotation fluoroscopy is an underutilized diagnostic modality in the evaluation and surgical planning in patients with urogenital sinus and cloacal anomalies and should be considered by surgeons prior to proceeding with corrective surgery.

Insight Urol 2022;43(2):159-66. doi: 10.52786/isu.a.62

Corresponding author: Laurence S. Baskin

Address: Department of Urology, University of California San Francisco, 400 Parnassus Avenue A610, San Francisco, CA 04143, USA

E-mail: Laurence.Baskin@ucsf.edu

Manuscript received: October 13, 2022

Revision received: November 25, 2022

Accepted after revision: December 4, 2022

Introduction

Cloacal anomalies and common urogenital sinus (UGS) are structural abnormalities of hindgut and urogenital development. UGS abnormalities result in abnormalities of the urethra and vagina whereas cloacal also include abnormalities of the anus and rectum.

Persistent cloaca

Cloacal anomalies are anorectal malformations that result in an incomplete separation between the urogenital and digestive tracts during embryonic development. A persistent cloaca is the most severe form of anorectal malformation found in females, with an external perineal opening common for the genital, urinary and digestive systems. In males, a high anorectal malformation with rectal atresia and congenital recto-urethral fistula is the anatomical equivalent.[1] Unlike common UGS, persistent cloaca is not a disorder of sexual differentiation due to androgen excess, but rather a result of incomplete formation of the urorectal septum.[2]

Persistent urogenital sinus

Common UGS occurs due to persistent communication between the urethra and vagina resulting in a single perineal opening of the urinary and Müllerian systems. The single opening, termed the urogenital sinus, is a transient feature of normal fetal development. The anatomical abnormality occurs when development of two separate urethral and vaginal openings is incomplete.

Common UGS may be an isolated malformation or may be associated with complex syndromes.[3] The formation of a single opening of the urethral and genital system may be attributed to the virilization of the external genitalia,[4] and therefore is often associated with disorders of sex differentiation and conditions resulting from androgen excess during fetal development, most commonly congenital adrenal hyperplasia (CAH).[5] Common UGS can also be associated with other signs of virilization of the external genitalia, such as clitoral hypertrophy, and scrotalization of the labia majora with a large variation of phenotype, from mild clitoromegaly without a common UGS to complete Prader V virilization with a long length of UGS.[1,6]

Anatomic implications for surgical management

Diagnosis of cloacal anomalies are typically made at birth when a single perineal orifice is observed. Imaging of the newborn is necessary to identify complicating factors such as hydrometrocolpos, or severe hydroureternephrosis due to obstruction from a dilated urine filled vagina.[7]

At birth, a life saving diverting colostomy is necessary to direct stool away from the malformation and allow the child to thrive clinically. Postnatally, bladder function will need to be assessed specifically with regard to bladder emptying and the absence of vaginal voiding and possible hydroureteronephrosis as a consequence of an obstructing urine filled vagina. If this is the case clean intermittent catheterization of the single perineal opening into the urine filled vagina can facilitate decompression of the urinary tract until reconstructive surgery can be carried out.

Following the initial colostomy and prior to the definitive repair, assessment of adequate urinary drainage, radiological studies and endoscopy are recommended to help delineate the anatomy in order to plan for surgery. Accurate assessment of the pelvic anatomy in these patients is critical, as the distance of the common channel from the perineum heavily influences the surgical approach.[8] A short distance of less than 1cm of common channel to the external opening often does not require significant mobilization of the urogenital sinus. Therefore, a posterior sagittal approach to separate the rectum from the vaginal structures, create a new perineal body and perform anoplasty is typically selected for repair.[8] Cloacae with a common channel between 1 and 3 cm can also be approached in this fashion, with the additional maneuver of total urogenital sinus mobilization (TUM) in order to create a separate urethral and vaginal opening in the perineum.[8] Cloacae with a common channel greater than 3 cm may require a laparotomy in order to perform a transabdominal urogenital mobilization successfully. In very high cloacae, this may need to be the initial approach if the confluence is found to be at the level of the bladder neck. For high cloacae with a more distal confluence, transabdominal urogenital mobilization may be performed after a posterior sagittal dissection and TUM, which at the time of reconstruction would require whole body skin preparation for intraoperative repositioning.

Similar to the cloacal patient, the length of the urogenital sinus and the distance from the confluence of the structures to the perineal skin greatly impacts the surgical approach in patients with common urogenital sinus.[1,6] In patients with a low confluence, vaginoplasty utilizing a perineal skin flap to the spatulated posterior urethra may be adequate to bring both orifices to the level of the skin.[9] Total urogenital sinus mobilization (TUM) may be necessary for patients with high confluence, the UGS being dissected circumferentially as a single unit in order to bring the confluence anteriorly to the level of the perineal skin.[9,10] Due to concerns with regard to urinary incontinence, this technique is typically reserved for the most severe cases. Partial UGS mobilization (PUM) spares the pubourethral ligament and is often adequate for most cases of common UGS repair.[9,11]

Traditional Imaging Modalities in Evaluation of Cloaca and Common Urogenital Sinus

Prenatal imaging

With the increase in routine use of prenatal ultrasound, cloacal anomalies and persistent UGS can often now be diagnosed prenatally. Prenatal cases are diagnosed by identification of hydrometrocolpos as an oblong, anechoic lesion located behind the fetal bladder (Figure 1A).[12] Septation can be visualized in some cases crossing the cystic mass, and this is thought to be the urogenital septum separating the vagina and the bladder.[13] Duplicated Mullerian structures can also be visualized as a vertical septation separating dilated hemivaginas (Figure 1C). Specific to cloacal anomalies, the rectum can also be dilated, and the absence of a "target sign" correlating with an anus can be indicative. However, these findings can be challenging to identify in the setting of significant vaginal and/or ureteral dilation, and are not diagnostic.[14]

If severe, hydrometrocolpos can result in urinary obstruction either to the lower or upper urinary tract, and compression of the fetal bladder (Figure 1B) and hydronephrosis can be seen. If the intra-abdominal pressure is sufficiently elevated, this can result in pulmonary hypoplasia in severe cases. Fetal ultrasound findings will often suggest this abnormality prior to birth and assist with prompt diagnosis and management postnatally.[15]

Postnatal ultrasound

In cases of cloacal anomaly, postnatal ultrasound prior to diverting colostomy is recommended to rule out hydrometrocolpos and upper tract urinary obstruction.[7] Contrast enhanced ultrasound has also been utilized in order to delineate the location of the rectovaginal fistula, length of the common channel, distance from bladder neck to common channel, and from urethra to perineum.[16]

Common UGS is most commonly diagnosed by the presence of atypical genitalia at birth. However, in unclear cases, pelvic ultrasound can be useful in diagnosis. Neonatal Mullerian

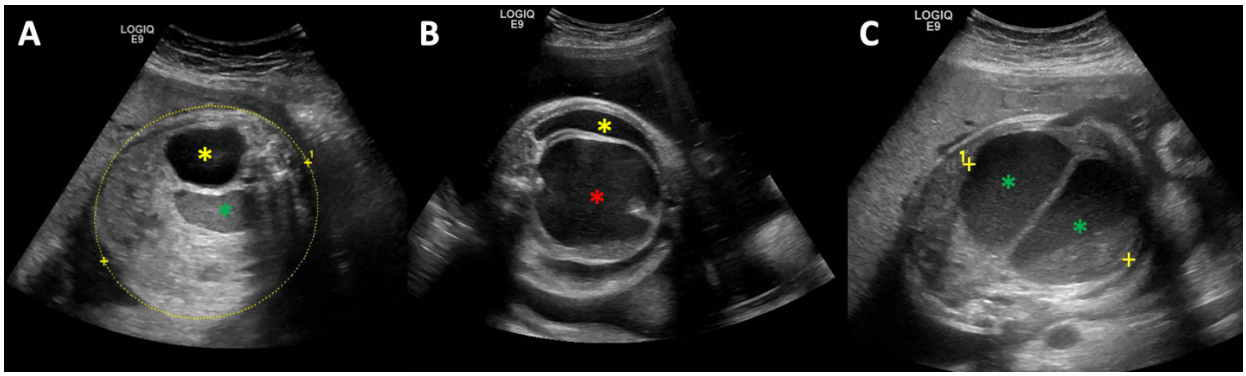

Figure 1. Prenatal ultrasound images of fetuses with cloacal and urogenital sinus anomalies. (A) shows a transverse image of the fetal pelvis of a fetus with common UGS with the distended fetal bladder marked by the yellow asterisk and the dilated fetal vagina marked by a green asterisk. Notably, the dilated vagina has more echogenic material within, representing hydrometrocolpos. (B) is a transverse ultrasound image of a fetal pelvis in a patient with a cloacal anomaly. The urinary bladder (yellow asterisk) is compressed posteriorly by a large and dilated cloaca (red asterisk). (C) is a transverse ultrasound image of a fetal pelvis in a patient with common UGS and duplicated vagina. The two hemivaginas are marked by green asterisks and separated by a vaginal septum in between. There is evidence of debris layering in the more inferior hemivagina.

structures are stimulated up to the time of birth due to the effects of maternal estrogen, and can be identified with ultrasound in the post-natal period.[17] Identification of hydrometrocolpos and hydronephrosis due to urinary obstruction are important first-line ultrasound findings. However, ultrasound is typically unable to identify the location of the confluence, and therefore follow up imaging is necessary (Figure 1A).

Voiding cystourethrogram (VCUG)/genitogram and colostogram

Voiding cystourethrogram (VCUG)/genitogram is performed by placing contrast medium through a bladder catheter and obtaining radiograph images. VCUG is an efficient and cost-effective modality justifying the exposure to radiation in these cases as it provides important anatomic information pertinent to identification of the confluence. During the voiding phase of imaging, after the bladder catheter has been removed, the contrast medium may flow through the confluence and lower urinary tract/vagina, and/or enteric fistula enhancing delineation of the anatomy.[3]

The challenges of VCUG in these cases are due to the abnormal anatomy of the patient. Placement of a catheter into the bladder may not be straightforward, and inadvertent placement of the catheter into the vagina or enteric fistula is possible. In difficult cases, cystotomy or cutaneous vesicostomy has been utilized to gain access into the bladder for both decompression and drainage as well as diagnostic studies.[18]

In cases of cloacal anomaly, an augmented pressure colostogram through the previously created colostomy/mucus fistula can be utilized to help identify the location of the fistula.[19] (Figure 2A) Prior studies have reported an accuracy of 66% for colostogram and 58% for VCUG for identifying the level of the fistula.[20]

Magnetic resonance imaging (MRI)

In cases where ultrasound and VCUG have not been adequate for the delineation of the pelvic anatomy, magnetic resonance imaging (MRI) has been a useful modality in providing clarity. MRI provides excellent soft tissue and spatial resolution, allowing for clear visualization of any malformation (Figure 2B, 3B). Accuracy in identifying the confluence in cloacal anomalies with MRI has been reported to be similar to cystoscopic and fluoroscopic findings.[19] MRI can be particularly useful in patients with severe anomalies.[21] Other structures such as gonads, kidneys, adrenal glands, and the presence or absence of internal and external genital organs may all be evaluated with this modality. The presence or absence of spinal abnormalities can also be assessed with MRI. The major drawback for MRI is the need for anesthesia in cases where the young patient is no longer small enough to tolerate the "feed and swaddle technique".[3,22] Additionally, the inability to artificially dilate the bladder or vagina can make it difficult to identify key structures, and the confluence is not always easily visible in this modality (Figure 2B).

Examination Under Anesthesia and 3D Rotational Fluoroscopy

Three-dimensional (3D) rotational fluoroscopy is an imaging technique performed in con junction with the pediatric urologist/surgeon and interventional radiology (IR). This imaging technique paired with examination under anesthesia (EUA) allows for careful delineation and understanding of the anatomy with manual dilation of the structures. The location of the confluence and length of the common channel can be assessed. Any vaginal abnormalities such as duplication or absence, as well as any vesicoureteral reflux or bladder abnormalities are able to be evaluated as well to optimize surgical planning. Utilization of this technique has been well described in preoperative investigation of cloacal anomalies,[23] and thus easily translates into evaluation of common UG sinus as well. One of the main advantages over EUA alone is that the 3D rotational fluoroscopy provides a permanent image of the deciphered anatomy that becomes an easily accessible part of the medical record available to all providers.

At our institution, we advocate 3D Rotational Fluoroscopy for patients with cloacal anomalies and more severe UGS anomalies when surgical reconstruction is being considered, typically at approximately 6 months of age. This includes a physical examination, cystoscopy, vaginoscopy, and endoscopy with the patient under general anesthesia in the IR suite with the 3D fluoroscopy scanner. The examination is performed in the frog leg position. It is important to take careful note of the location of the confluence in relation

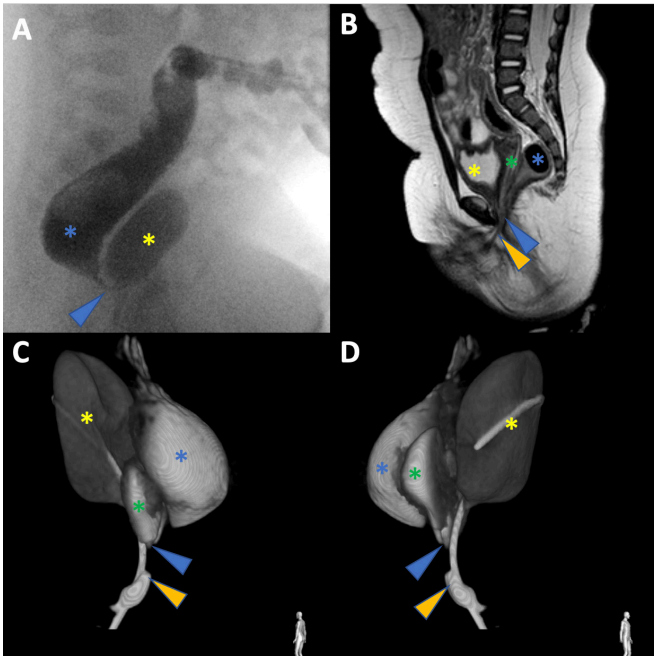

Figure 2. Images from radiology of a single patient with a congenital urogenital sinus with a high confluence. (A) shows an ultrasound image in the longitudinal plane showing the bladder (yellow asterisk) and vagina (green asterisk). The confluence and external opening are not visible. (B) shows an MRI image in the sagittal plane. The bladder (yellow asterisk), dilated vagina (green asterisk) and external opening (orange arrow) are visible. (C) shows a fluoroscopic section in the sagittal plane with contrast material in the vagina (green asterisk) and layering in the bladder (yellow asterisk). The confluence is visible (blue arrow) as well as the external opening (orange arrow). (D) shows the three-dimensional reconstruction of image (C) with the bladder visible (yellow asterisk) as well as the confluence (blue arrow) and external opening (orange arrow). (E) and (F) are rotated images of the same reconstruction with the bony structures removed digitally. Bladder (yellow asterisk), vagina (green asterisk), confluence (blue arrow), and external opening (orange arrow) are all easily identified.

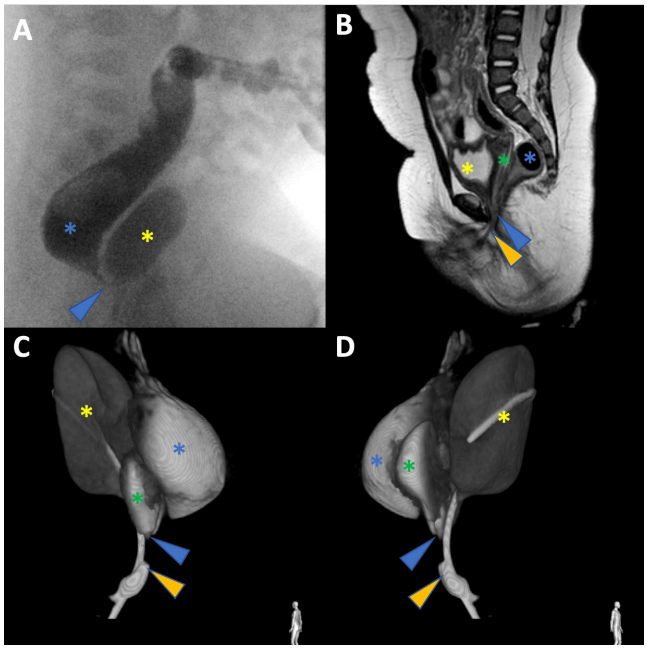

Figure 3. Images from radiology of a patient with persistent cloaca. (A) shows a colostogram image with contrast being injected through the colostomy and dilating the colon (blue asterisk). The fistulous connection to the common cloaca is identified (blue arrow) and contrast is also filling the bladder (yellow asterisk). (B) shows a sagittal plane MRI image of the same patient with the bladder (yellow asterisk), vagina (green asterisk), and colon (blue asterisk) visible. The confluence is identified (blue arrow), as is the common opening in the perineum (yellow arrow). (C) and (D) are both rotational fluoroscopic images after 3D reconstruction which show the bladder (yellow asterisk), vagina (green asterisk), and colon (blue asterisk) visible. The confluence is identified (blue arrow) as well as the common opening in the perineum (yellow arrow). Of note, this patient has a duplicated vagina, which is visible in the 3D fluoroscopic images but not identified in the colostogram or MRI.

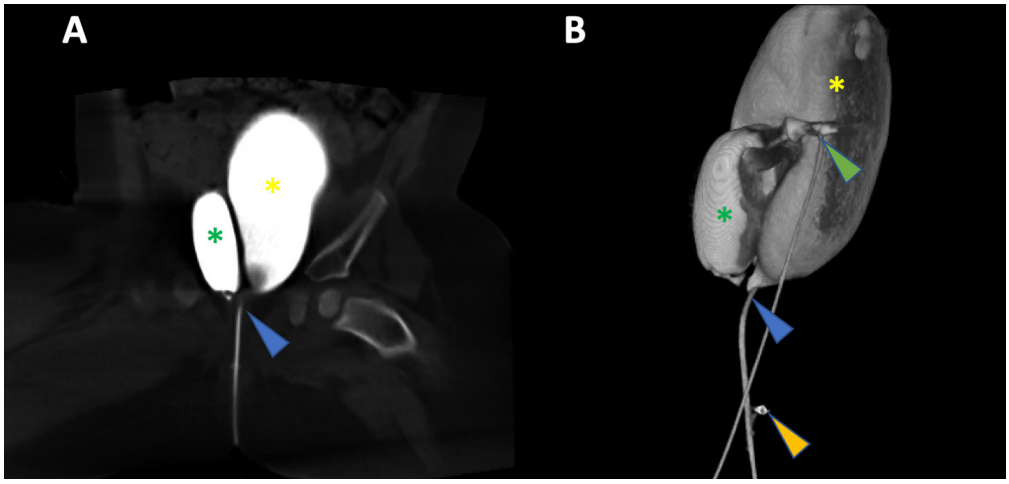

Figure 4. Images from 3D rotational fluoroscopy of a patient with high confluence common UGS and a prior vaginostomy. (A) shows a coronal section of the fluoroscopic imaging at the level of the confluence. (B) shows the 3D reconstruction of the vagina and bladder structures as viewed with the patient oriented in a similar supine position. The vagina (green asterisk) has been distorted to the patient left and brought anterior to the bladder due to the vaginostomy (green arrow). The bladder (yellow asterisk) is distended. The confluence is notably very high and close to the bladder neck (blue arrow) and the introitus is tagged at the perineal skin with a button (yellow arrow).

to meatus as well as distance from confluence to bladder neck. After completion of the EUA, 3-5 French open ended catheters can be placed into the bladder, the vagina/each hemivagina and the enteric fistula (if cloacal) under direct vision with care being taken to tag each catheter for easy identification. A metal BB is also placed at the perineal skin to define the location of the perineal opening. If a vesicostomy, vaginostomy, or colostomy is present, this can also be intubated with a Foley catheter for contrast injection and fluoroscopic imaging (Figure 4).

The IR technician will take a scout image to determine adequate positioning. The bladder is then filled with contrast solution via gravity until either calculated expected capacity is reached or drainage around the catheter is noted. Rotational fluoroscopy is then performed with the bladder filled after a scout image. The vaginal catheter (s) is (are) then slowly filled under gravity until there is evidence of drainage around the catheter. A second rotational image is then obtained. (Figure 2C, 4B, 5B). Finally, the rectal catheter is filled if the hindgut can be intubated or contrast is injected down the mucus fistula and a third rotational image can be obtained (Figure 3C, 3D). The catheters are then drained and removed. A urodynamics catheter can be placed if urodynamics are desired following the procedure. We have found this procedure to be well tolerated in patients ranging from six months to teenage years (Figure 5).

The 3D rotational imaging occurs as a 460-image series as the c-arm rotates 180 degrees around the patient. The imaging takes less than one minute and three-dimensional reconstruction occurs immediately on the IR 3-D workstation (Figure 2D, 3D-F, 4B, 5B). Images can be viewed as a 3-D render with rotation, or as slices/sections after reconstruction in any plane desired. Bony structures can be digitally removed from optimal viewing of the structures of interest (Figure 2E-F, 3C-D, 4B, 5B). There is some increase in radiation exposure in this method compared to 2D fluoroscopy, and prior studies have estimated that 1 minute of 3D rotational fluoroscopy time is equivalent to 1.5 minutes of 2D fluoroscopy.[24] At our institution, approximate effective dose of radation per rotational series ranges from 5-10 mSv, compared to the median effective dose of 31 mSv for a multi-phase abdomen/pelvis CT scan.[25]

Discussion

There are a few drawbacks to this technique. 3D fluoroscopy does require a slightly higher radiation dose than traditional 2D imaging.[23] Prior studies have shown that 1 minute of 3D fluoroscopy is equivalent to 1.5 minutes of 2D fluoroscopy with regards to dose area product. However, given the spatial understanding achieved by rotational fluoroscopy, the increase in the number of 2D images required to achieve similar understanding would make this difference negligible.[24] Additionally, anesthesia is required for this imaging protocol.

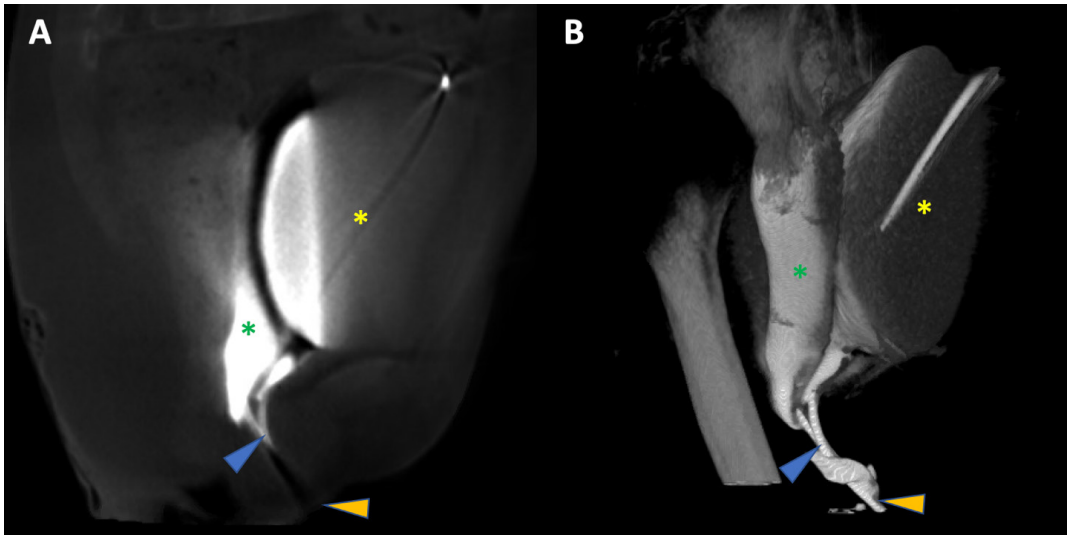

Figure 5. Images from 3D rotational fluoroscopy of a teenage patient with common UG sinus. (A) shows a sagittal section from the fluoroscopy with contrast material in the vagina (green asterisk) and layering in the bladder (yellow asterisk). The confluence (blue arrow) and introitus (yellow arrow) are both visible. (B) shows the reconstructed 3D image where the layering of contrast in the bladder (yellow asterisk) is clearly visible with the curl of the bladder catheter also visible.

The excess anesthesia exposure can be fully justified if the procedure is combined with planned cystoscopy, vaginoscopy and examination under anesthesia.

3D rotational imaging in conjunction with examination utilizing cystoscopy and vaginoscopy allows for detailed imaging with the bladder and vaginal structures distended, resulting in optimal understanding of the anatomy in these patients with a permanent radiographic record available for review at any time by the entire care team. Bladder capacity, morphology, reflux are evaluated. Vaginal size, morphology, and relative position to the bladder are evaluated. The length of the common channel, the length of the urethra from the bladder neck, and the location of the confluence are noted. Additional findings such as clitoral size, external virilization and other findings can also be recorded. All these findings are critical for optimal surgical planning prior to reconstruction. We believe that this is an underutilized diagnostic modality in the evaluation and surgical planning in patients with UGS and cloacal anomalies.

Conclusion

We believe that three-dimensional rotation fluoroscopy is an underutilized diagnostic modality in the evaluation and surgical planning of patients with urogenital sinus and cloacal anomalies This imaging technique should be considered by surgeons prior to proceeding with corrective surgery.

Conflict of Interest

The authors declare no conflict of interest.

References

1. Thomas D. The embryology of persistent cloaca and urogenital sinus malformations. Asian J Androl 2020;22:124-8.
2. Thomas DFM. The embryology of persistent cloaca and urogenital sinus malformations. Asian J Androl 2020;22:124-8.
3. Valentini AL, Giuliani M, Gui B, Laino ME, Zecchi V, Rodolfino E, et al. Persistent Urogenital Sinus: Diagnostic Imaging for Clinical ManagementWhat Does the Radiologist Need to Know? Am J Perinatol 2016;33:425-32.
4. Li Y, Sinclair A, Cao M, Shen J, Chuodhry S, Botta S, et al. Canalization of the urethral plate precedes fusion of the urethral folds during male penile urethral development: The double zipper hypothesis. J Urol 2015;193:1353-9.
5. Lottman H, Thomas DFM. Disorders of sex development. In: Thomas DFM, Duffy PG, Rickwood AMK (editors). Essentials of Paediatric Urology. 2nd ed. London: CRC Press; 2013. p. 289-308.
6. Jesus VM, Buriti F, Lessa R, Betania M, Toralles MB, Oliveira LB, Barroso Jr U. Total urogenital sinus mobilization for ambiguous genitalia. J Pediatr Surg 2018;53:808-12.

7. Winkler NS, Kennedy AM, Woodward PJ. Cloacal Malformations. J Ultrasound Med 2022;31:1843-55.
8. Bischoff A. The surgical treatment of cloaca. Semin Pediatr Surg 2016;25:102-7.
9. Leslie JA, Cain MP, Rink RC. Feminizing genital reconstruction in congenital adrenal hyperplasia. Indian J Urol 2009;25:17-26.
10. Peña A. Total urogenital mobilization - An easier way to repair cloacas. J Pediatr Surg 1997;33:263-8.
11. Rink RC, Metcalfe PD, Kaefer MA, Casale AJ, Meldrum KK, Cain MP. Partial urogenital mobilization: a limited proximal dissection. J Pediatr Urol 2006;2:351-6.
12. Taori K, Krishnan V, Sharbidre KG, Andhare A, Kulkarni BR, Bopche S, et al. Prenatal sonographic diagnosis of fetal persistent urogenital sinus with congenital hydrocolpos. Ultrasound Obstet Gynecol 2010;36:641-3.
13. Geifman-Holtzman O, Crane SS, Winderl L, Holmes M. Persistent urogenital sinus: prenatal diagnosis and pregnancy complications. Am J Obstet Gynecol 1997;176:709-11.
14. Rohrer L, Vial Y, Hanquinet S, Tenisch E, Alamo L. Imaging of anorectal malformations in utero. Eur J Radiol 2020;125:108859.
15. Gul A, Yildirim G, Gedikbasi A, Gungorduk K, Ceylan Y. Prenatal ultrasonographic features of persistent urogenital sinus with hydrometrocolpos and ascites. Arch Gynecol Obstet 2008;278:493-6.
16. Lin XY, Xu XH, Yang YP, Wu J, Xian X, Chen X. Preoperative evaluation of persistent cloaca using contrast-enhanced ultrasound in an infant. Med Ultrason 2020;22:250-2.
17. Chavhan GB, Parra DA, Oudjhane K, Miller SF, Babyn PS, Salle FLP. Imaging of ambiguous genitalia: classification and diagnostic approach. Radiographics 2008;28:1891-904.
18. Kitta T, Kakizaki H, Iwami D, Tanda K. Successful bladder management for a pure urogenital sinus anomaly. Int J Urol 2004;11:340-2.
19. Kraus SJ, Levitt MA, Peña A. Augmented-pressure distal colostogram: the most important diagnostic tool for planning definitive surgical repair of anorectal malformations in boys. Pediatr Radiol 2018;48:258-69.
20. Tofft L, Salö M, Arnbjörnsson E, Sternstron P. Accuracy of pre-operative fistula diagnostics in anorectal malformations. BMC Pediatr 2021;21:283.
21. Ryu JA, Kim B. MR imaging of the male and female urethra. Radiographics 2001;21:1169-85.
22. Antonov NK, Ruzal-Shapiro CB, Morel KD, Millar WS, Kashyap S, Lauren CT, et al. Feed and Wrap MRI Technique in Infants. Clin Pediatr (Phila) 2017; 56:1095-103.
23. Reck-Burneo CA, Lane V, Bates DG, Hogan M, Thompson B, Gasior A, et al. The use of rotational fluoroscopy and 3-D reconstruction in the diagnosis and surgical planning for complex cloacal malformations. J Pediatr Surg 2019;54:1590-4.
24. Patel MN. Use of rotational fluoroscopy and 3-D reconstruction for pre-operative imaging of complex cloacal malformations. Semin Pediatr Surg 2016;25:96-101.
25. Smith-Bindman R, Lipson J, Marcus R, Kim KP, Mahesh M, Gould R, et al. Radiation Dose Associated with Common Computed Tomography Examinations and the Associated Lifetime Attributable Risk of Cancer. Arch. Intern. Med. 2009;169:2078-86.