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PICTORIAL ESSAY

Complex cloacal malformations: use of rotational fluoroscopy and 3-D reconstruction in diagnosis and surgical planning

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Abstract A cloacal malformation is a congenital malformation in which the urinary tract, gynecological system and distal rectum fail to separate and form a common channel with a single perineal opening. Precise anatomical information is required to plan surgery and predict prognosis for children with this abnormality. Conventional fluoroscopic studies provide limited information, primarily due to the overlap of structures and inability to make accurate measurements. Rotational fluoroscopy and 3-D reconstruction help clarify overlapping structures and allow for precise measurement of the common channel, thereby helping to predict the complexity of the surgical case as well as the long-term prognosis regarding bowel, bladder and sexual function.

Keywords Cloacal malformations · Rotational fluoroscopy · 3-D imaging · 3-D reconstructions · Children

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Introduction

A cloacal malformation is a rare congenital abnormality in which the urinary tract, gynecological system and the distal rectum form a confluent common channel. Diagnostic imaging of patients with complex cloacal malformations can be challenging; however, accurate anatomical visualization is essential for surgical planning. We examine the diagnostic applicability and utility in surgical planning of rotational fluoroscopy and 3-D reconstruction in children with complex cloacal malformations.

Background

A cloacal malformation is caused by failure of the urorectal septum to form or fuse properly during development [1]. Growth of this septum may be hindered at any stage of development; therefore, variable anatomy of the genital, urinary, rectal and perineal structures is seen in children with these malformations [1–3]. Cloacal malformations occur exclusively in females with an incidence of 1 in 40,000 to 50,000 newborns [1–3], although due to misdiagnoses, this incidence may be underestimated [4].

Children with cloacas typically require a diverting colostomy immediately after birth to prevent fecal contamination of the urinary tract and subsequent renal damage [3]. Children with hydronephrosis usually due to hydrocolpos may also require placement of a vaginostomy

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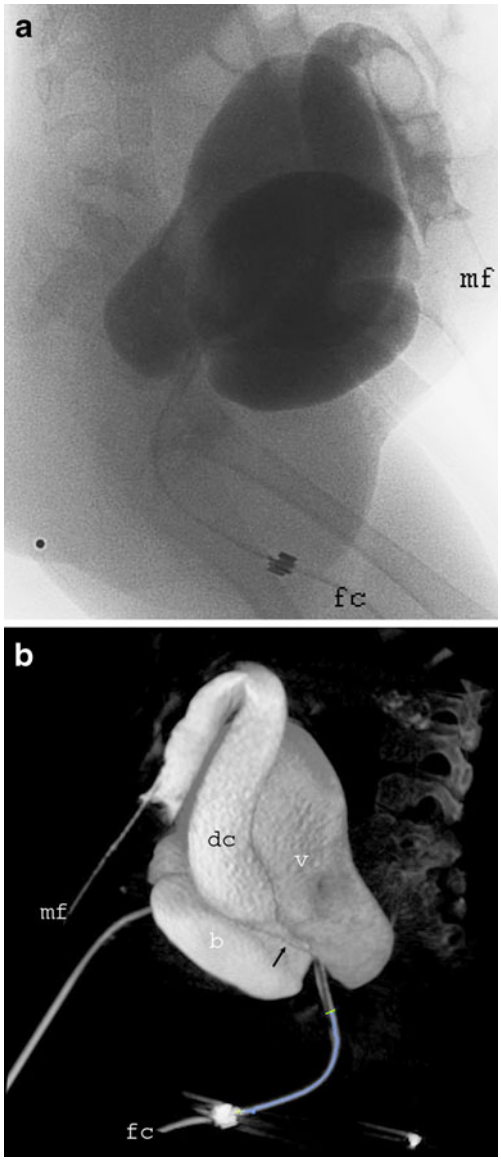

Fig. 1 This 9-month-old had a diverting colostomy and creation of a vesicostomy after birth. Conventional lateral fluoroscopic image (**a**) from a cloacagram demonstrates multiple overlapping structures making it difficult to distinguish the distal colon, bladder and vagina. Left sagittal 3-D reconstructed image from the cloacagram (**b**) clearly demonstrates the distal colon (*dc*), rectal fistula (*arrow*), bladder (*b*) and vagina (*v*). (*mf*=Foley catheter placed through mucous fistula; *fc*= Foley catheter placed through common channel.) The child has a small bladder and a markedly enlarged right hemi-vagina that wraps posteriorly and to the right of the distal colon and rectal fistula. The common channel (blue line as drawn by the workstation by selecting the proximal and distal aspects of the common channel) measures 5 cm. Because of the findings identified on 3-D cloacagram, it was determined that the child would require a laparotomy in addition to the posterior sagittal anorectalplasty (PSARP). The common channel was left untouched to function as the child's urethra. She also underwent a vaginal switch procedure. (Note the radiopaque BB at the expected location of the rectum and the external marker on the Foley catheter through the common channel at the level of the perineum.)

tube [3]. Rarely, a vesicostomy is also needed. Definitive surgical repair of cloacal malformations is technically challenging and requires an accurate understanding of the abnormal anatomy. Goals of definitive surgical repair are to achieve bowel and bladder control and normal sexual function, although in many patients, only one or two of these goals are achieved [4]. The likelihood of achieving these goals is dependent upon a number of anatomical factors, such as the sacral ratio [5], length of the common channel/level of the cloaca, characteristics of the vagina(s) and appearance of the bladder [5].

Diagnostic imaging is essential to provide precise anatomical definition for accurate surgical planning and appropriate patient counseling regarding probability of fecal and urinary continence. Traditionally, a high-pressure distal colostogram using conventional 2-D fluoroscopy has been the imaging method of choice for patients with cloacal malformations [2, 3]. After injection of water-soluble contrast material into the mucous fistula, frontal, lateral and, occasionally, oblique images are obtained to visualize the relationships among the rectum, vagina(s) and bladder. However, there is significant overlap of structures on the 2-D images, which often obscures anatomical detail (Fig. 1). In addition, accurate measurement of the length of the common channel is

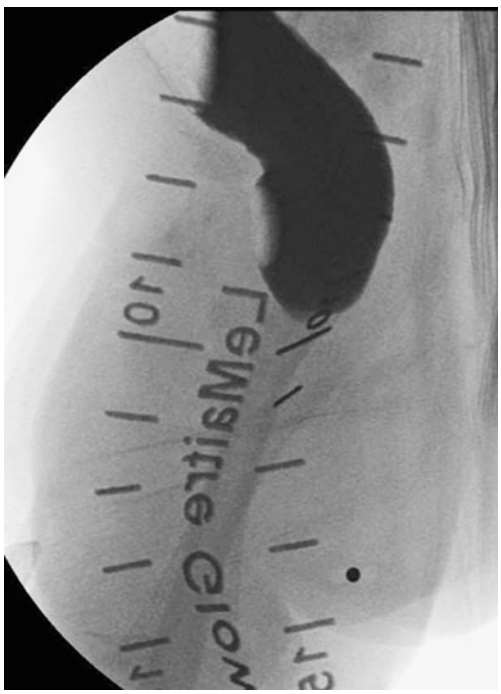

Fig. 2 Lateral fluoroscopic image from a child with an imperforate anus. A ruler was placed externally to measure the distance from the rectum to the perineum (BB marker). Precise measurement is difficult as there is magnification and foreshortening of the ruler

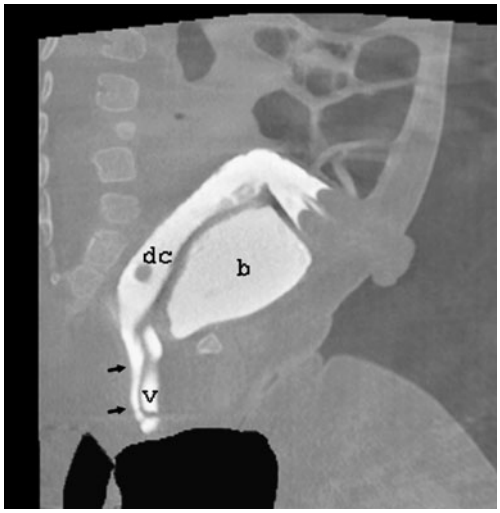

Fig. 3 Sagittal CT-like image from a cloacagram nicely demonstrates the relationships among the sacrum, distal colon (*dc*), rectal fistula (*arrows*), vagina (*v*) and bladder (*b*)

problematic because of the difficulties associated with the 2-D nature of fluoroscopic imaging, including foreshortening, no internal reference for measurement and magnification errors associated with using skin surface measuring devices (Fig. 2).

Rotational fluoroscopy with 3-D reconstruction is a C-arm cone-beam CT technique that provides 3-D isotropic soft-tissue volumes reconstructed from rotational acquisitions. This 3-D data set can be viewed and manipulated both in the control room and tableside in the procedure room. The 3-D images help clarify overlapping structures and allow precise measurement of the common channel in these patients.

MRI has also been used to provide 3-D detail of the cloacal anatomy and to define spatial relationships [6]. MR imaging, however, cannot be performed in real time, whereas fluoroscopy provides dynamic real-time anatomical information as various structures are filled with contrast. Real-time fluoroscopy also provides an endpoint for when optimal distension and opacification of the various struc-

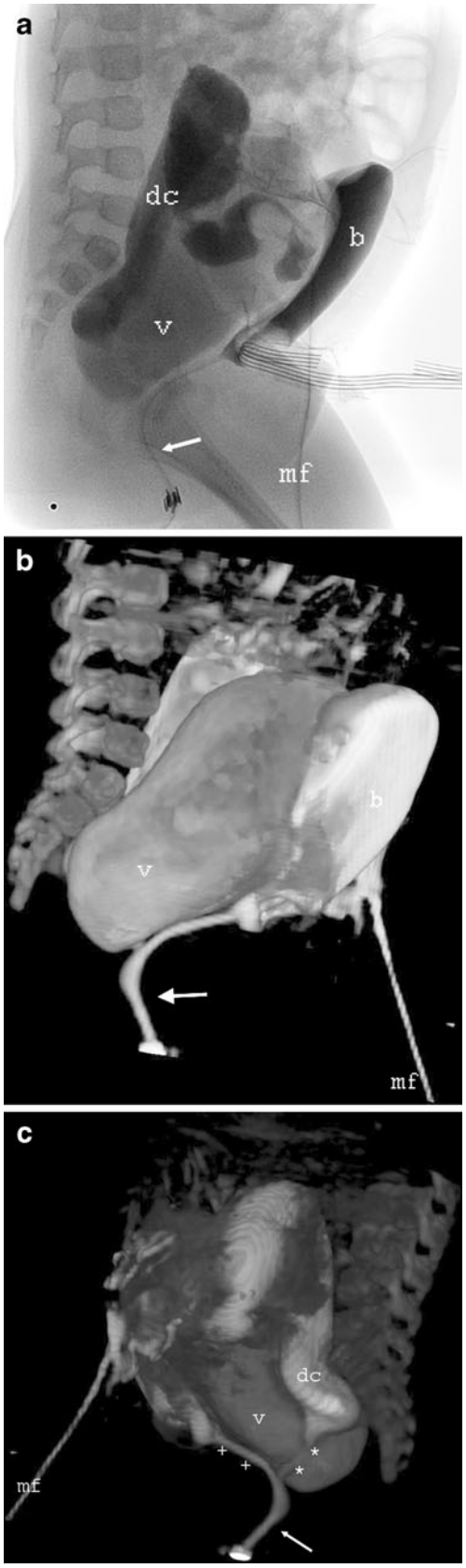

Fig. 4 This 8-month-old had a diverting colostomy at birth. Conventional lateral fluoroscopy image from the cloacagram (**a**) does not show the rectal fistula or its insertion to the common channel (*white arrow* indicates Foley catheter in common channel). The reconstructed 3-D images as viewed from the right hemi-pelvis (**b**) and left hemi-pelvis (**c**) show a large midline vagina (*v*) extending into the right hemi pelvis and resulting in right-sided displacement of the bladder (*b*). The insertion of the rectal fistula into the common channel (*white arrow*) is seen on the left hemi-pelvis image. The common channel measures 3 cm. Because of the 3-D cloacagram findings, it was determined that the child would require a PSARP without laparotomy. (*b* = bladder, *dc* = distal colon, + = urethra, *mf* = Foley catheter placed through mucous fistula)

tures (rectum, bladder, vagina[s], common channel) are achieved. Subsequent rotational fluoroscopy and 3-D reconstructions provide 3-D anatomical detail.

Since 2005, we have performed 87 rotational fluoroscopy studies with 3-D reconstructions for children with cloacal malformations. The mean age of the children was 11.6 months with a range of 1 to 52 months. The imaging technique and the clinical utility of these studies will be described.

Technique

The child is placed under general anesthesia to avoid movement during the procedure and an antibiotic (a second-generation cephalosporin) is administered. A metal BB marker is placed on the perineum at the expected location of the anus. Frontal and lateral fluoroscopic images of the sacrum are then obtained to calculate the sacral ratio [5]. An 8-French end-hole Foley catheter (Folysil Pediatric Foley; Coloplast, Minneapolis, MN) is placed through the child's mucous fistula and the balloon inflated. While the child is supine, a small amount of water-soluble contrast (Cysto-Conray; Mallinckrodt Inc., St. Louis, MO) is injected to verify the location of the ostomy and length of distal colon and rectum. The child is then placed in the lateral position and a high-pressure distal colostogram is performed to fully distend the rectum. Occasionally, the vagina, bladder and common channel are all successfully opacified from this injection, and rotational fluoroscopy and 3-D reconstructions are then performed. However, the bladder usually does not opacify from the initial injection. Since we now usually perform the imaging studies immediately following cystoscopy and vaginoscopy, during which the surgeons place a Foley catheter into the bladder via the common channel, we administer contrast through the Foley catheter to opacify the bladder. If the child does not have a Foley catheter, suprapubic catheter or vesicostomy tube in place, she is placed in the supine position

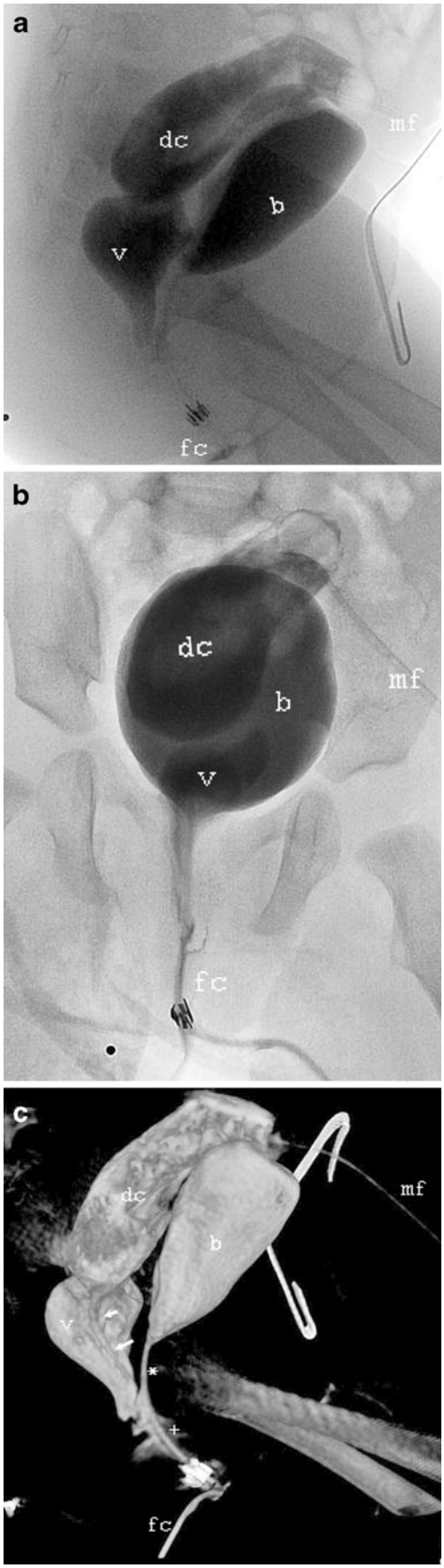

Fig. 5 This 6-month-old had diverting colostomy at birth. Conventional radiographs (**a, b**) from a cloacagram demonstrate a short segment of distal colon (*dc*) and a left hemi-vagina (*v*). The right hemi-vagina does not opacify, suggesting that it is rudimentary, although occasionally a vaginal obstruction may have a similar appearance. The rectal fistula is not seen inserting into the common channel. The 3-D reconstructed image (**c**) shows the long narrow rectal fistula (*arrow*) anterior to the left hemi-vagina. The common channel (+) measures 2 cm. Because of the long, narrow rectal fistula identified on the 3-D cloacagram, the surgeon was prepared to perform a laparotomy for rectal dissection. (* = urethra, *b* = bladder, *mf* = Foley catheter placed through mucous fistula, *fc* = Foley catheter placed through common channel)

and an attempt is made at placing a 6- or 8-French Coude catheter (Coloplast Self-Cath; Coloplast USA, Minneapolis, MN) through the perineal opening into the common channel and bladder. An external radiopaque marker (sticker) is placed on the urinary catheter at the level of perineal opening of the common channel.

Immediately after the rectum, vagina, bladder and common channel are opacified, the patient is placed in a lateral position for the rotational fluoroscopy portion of the study. Imaging is performed on an Integris Allura angio/interventional system (FD20 Philips, Best, The Netherlands) by selecting an image acquisition protocol originally developed for 3-D rotational angiography. During rotational fluoroscopy, 120 images are acquired as the c-arm rotates 180° around the child; this takes 8 sec. A 3-D reconstruction is immediately available on a dedicated 3-D workstation. Images can be evaluated in both a 3-D rendering mode or as "CT-like" slices in any obliquity (Fig. 3). Following this portion of the study, the residual contrast is drained and catheter(s) are removed.

Clinical utility: answering key anatomical questions

Length of common channel/level of cloaca

Children with cloacal malformations are generally separated into two major subgroups: those with a common channel less than 3 cm in length and those with a common channel greater than 3 cm in length [7, 8]. Children with a shorter common channel have a relatively good prognosis and can usually undergo posterior sagittal anorectalplasty (PSARP) with total urogenital mobilization [4, 5, 7] (Fig. 4). These are usually shorter procedures that require only a posterior sagittal approach and have a shorter operative time and postoperative hospital course. Children with shorter common channels have a higher incidence of voluntary bowel movements and urinary continence compared to children with a common channel greater than 3 cm. On occasion, cloacas with a common channel of less than 3 cm with a long, narrow rectal fistula may require a laparotomy for rectal dissection (Figs. 5 and 6). The rectal fistula is usually not well seen in these cases on conventional cloacagram.

Children with a common channel longer than 3 cm have a more complex defect, usually require laparotomy in addition to the posterior sagittal approach and are more likely to have long-term sequelae [4, 5, 7] (Fig. 1). Laparotomy allows for transabdominal urogenital mobilization, which provides extra length for pull-through [7]. Common channels greater than 5 cm may be left untouched to function as the urethra (Fig. 1 and 7), although these children usually require intermittent catheterization.

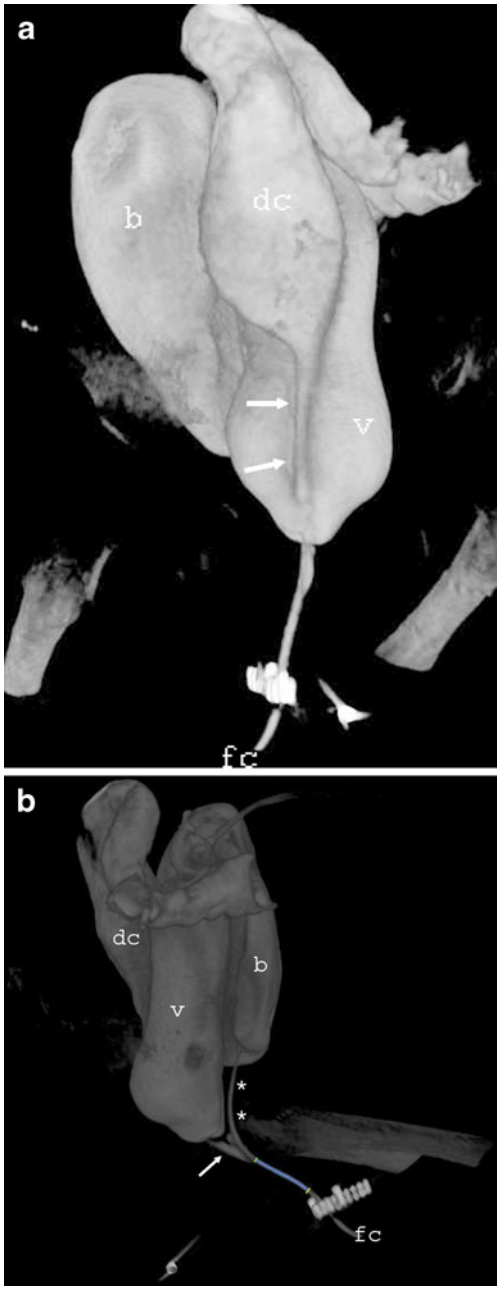

Fig. 6 This 6-month-old had a diverting colostomy and placement of a vaginostomy tube at birth. Selected posterior (**a**) and right sagittal (**b**) 3-D reconstructed images from a cloacagram show a long narrow rectal fistula (*arrows*) inserting at the base of the vagina (*v*) into the common channel (*blue line*). The common channel measures 2.5 cm. There is an adequate amount of bowel present to reach the expected location of the anus (BB marker). Again, the long, narrow nature of the rectal fistula suggested the possible need for laparotomy for rectal dissection, despite a short common channel. (*dc* = distal colon, *b* = bladder, * = urethra, *fc* = Foley catheter placed through common channel)

Accurate measurement of the common channel is difficult using 2-D fluoroscopy because of magnification and foreshortening; however, it can be performed relatively easily from the 3-D reconstruction on a dedicated 3-D workstation. The proximal portion of the common channel (the confluence of the rectal fistula, urethra and vaginas) and the distal portion of the common channel (the level of the perineal opening) are selected and the measurement is automatically made from the 3-D data set (Figs. 1 and 6).

Vaginal anatomy

The separation of the vagina from the urinary tract during definitive repair of the cloacal malformation is a tedious and delicate maneuver [8]. Preoperative definition of the vaginal characteristics, such as size, location and presence of hemi-vaginas, is important in determining the best approach to the repair of the genital component of the malformation [8]. If a child has a large vagina, the surgeon can more easily mobilize it and has more alternatives for the vaginal repair [4] (Fig. 4). If the child has a small vagina or a vagina located high in the pelvis, it is usually necessary to perform a vaginal switch, in which one of the hemi-vaginas is attached to the other to reach the perineum (Fig. 8), and/or vaginal replacement (Figs. 7 and 9) using rectum, colon or small bowel [7]. Rectum is the preferred donor for vaginal reconstruction, followed by descending colon, sigmoid colon and small bowel.

Children with cloacal malformations may also have varying degrees of vaginal and uterine septation, duplication or atresia; these abnormalities may lead to future retrograde menstruation [4, 9]. Atretic or rudimentary vaginas may be suggested by the location of the opacified vagina (Fig. 5).

Distal colon length (length of bowel from mucous fistula to distal rectum)

Ideally, the length of the distal colon is adequate for the pull-through and even for possible vaginal reconstruction. Inadequate length may necessitate a laparotomy and/or takedown of the mucous fistula. This is generally a subjective assessment based on the location of the colostomy and length of distal colon present on the cloacagram. To make this determination, a BB marker is

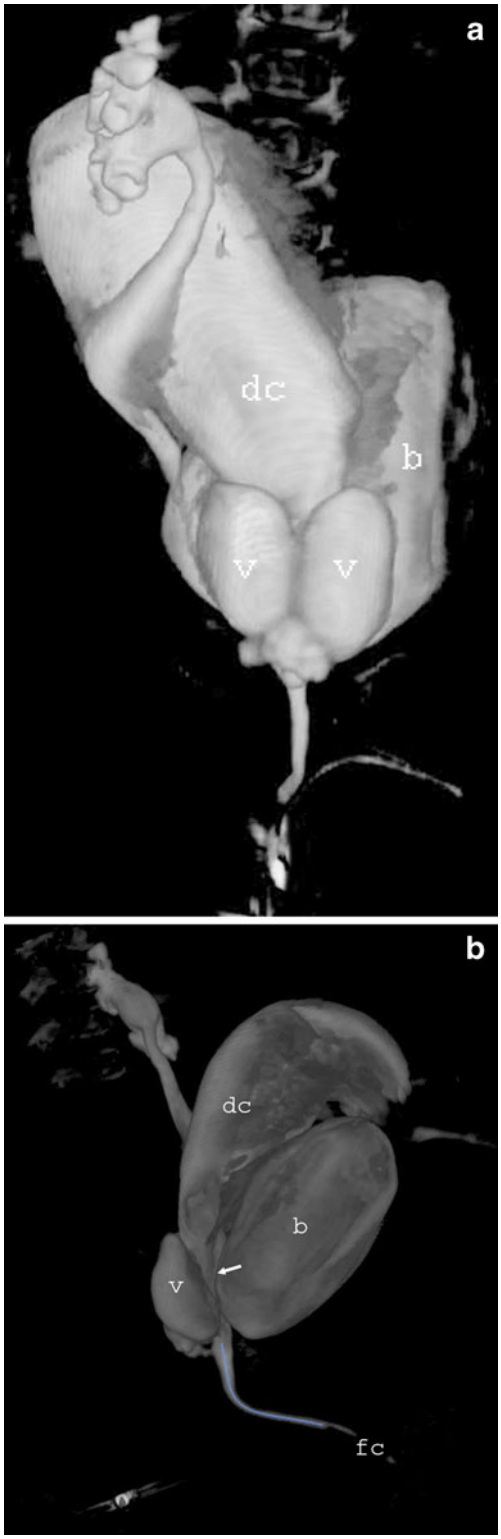

Fig. 7 This 2-month-old had a diverting colostomy and placement of a suprapubic catheter at birth. Posterior (**a**) and right sagittal (**b**) reconstructed 3-D images reveal that the child has a short segment of distal colon that would likely not reach the expected location of the rectum (BB marker). The rectal fistula (*arrow*) inserts anteriorly and in the center of the hemi-vaginas (*v*). Left-sided vesicoureteral reflux is present (note the presence of contrast in the ureter and collecting system). The common channel (*blue line*) measures 5.3 cm. Due to the short segment of distal colon and long common channel, the child was predicted to require a laparotomy in addition to a PSARP. Surgically, the common channel was left untouched and the distal colon was used for partial vaginal replacement. The colostomy site was pulled through to the neoanus and the child then had a new colostomy created. (*fc* = Foley catheter placed through the common channel)

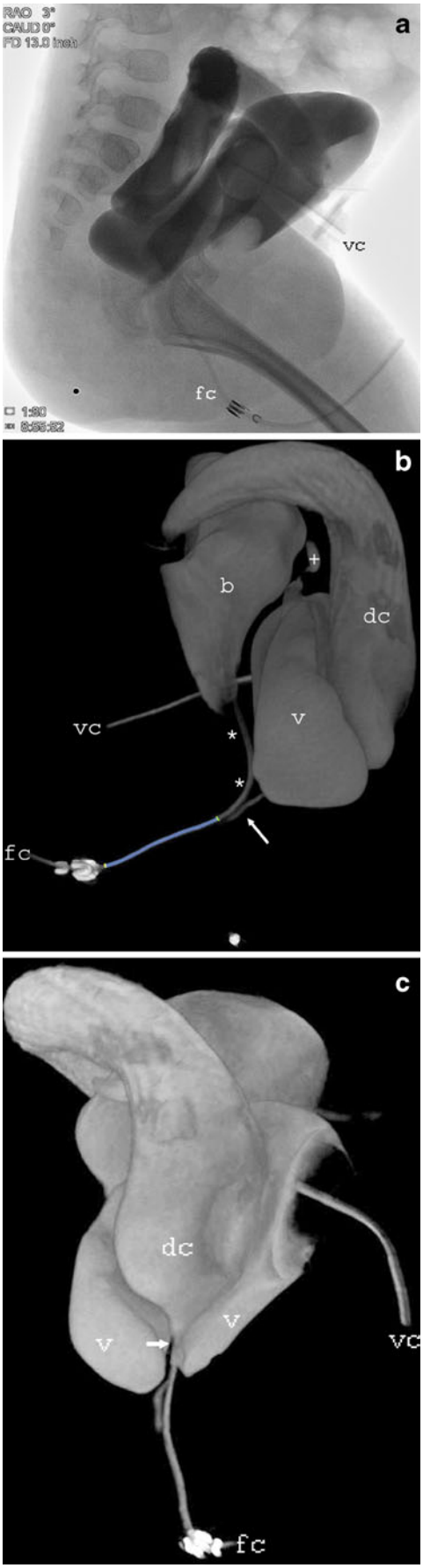

Fig. 8 This 15-month-old had a diverting colostomy and placement of a vaginostomy tube after birth. Conventional lateral image (**a**) from the cloacagram demonstrates multiple overlapping structures. It is difficult to decipher the vagina, distal colon and bladder. A Foley catheter is present in the bladder through the child's common perineal opening (*fc*); another Foley catheter is present in the right hemi-vagina through the vaginostomy site (*vc*). Both catheters are difficult to visualize on the 2-D image. The bladder and hemi-vagina are both relatively high within the pelvis. Three-dimensional reconstructed left sagittal (**b**) and posterior (**c**) images from the cloacagram demonstrate the rectal fistula (*arrow*) inserting at the base of the hemi-vaginas (*v*) into the common channel (*blue line* as drawn from the work station). The hemi-vaginas are moderate in size but somewhat high within the pelvis. Reflux of contrast into the left uterine cavity (+) is noted on the reconstructed sagittal image. The Foley catheters are now more easily visualized. The common channel measures 4.5 cm. Due to the length of the common channel and location of the hemi-vaginas, it was determined that the girl would likely need a laparotomy. Surgically, she had a PSARP for total urogenital mobilization and then a laparotomy to complete the separation of the vaginas from the bladder after the urogenital complex was delivered up into the abdomen. She also required a vaginal switch procedure and carving of the pubic bone for final repair. (* = Foley catheter through the urethra)

placed on the expected perineal location of the anus and good frontal and lateral images are obtained during the original injection of the mucous fistula. Often, there is a redundancy in the sigmoid; rotational fluoroscopy and 3-D reconstruction help to more accurately estimate the length of distal colon, particularly when there is redundancy in this bowel segment. A colostomy in the very proximal sigmoid is the ideal location [7].

Presence of reflux and/or ectopic ureter

Occasionally, vesicoureteral reflux (Fig. 7) and ectopic insertion of the ureter (Fig. 9) are noted on the cloacagram. This may necessitate additional surgical interventions. In cases of ectopic insertion of the ureter, cutaneous ureterostomy may be created or the child may require a ureteral reimplantation.

Clinical utility in practice

In our experience, the information obtained from rotational fluoroscopy and 3-D reconstructions enabled the surgeons to predict the complexity of the surgical case, including length of operating room time, operating room equipment needs and postoperative length of stay. They were better able to anticipate the size and location of the vagina(s) and therefore better anticipate the need for vaginal replacement. They were able to more accurately define the height and location of the rectum, which helped them anticipate the need for bowel preparation. The 3-D rotational imaging also provided visualization of ectopic ureters, which facilitated surgical planning (reimplantation, cutaneous

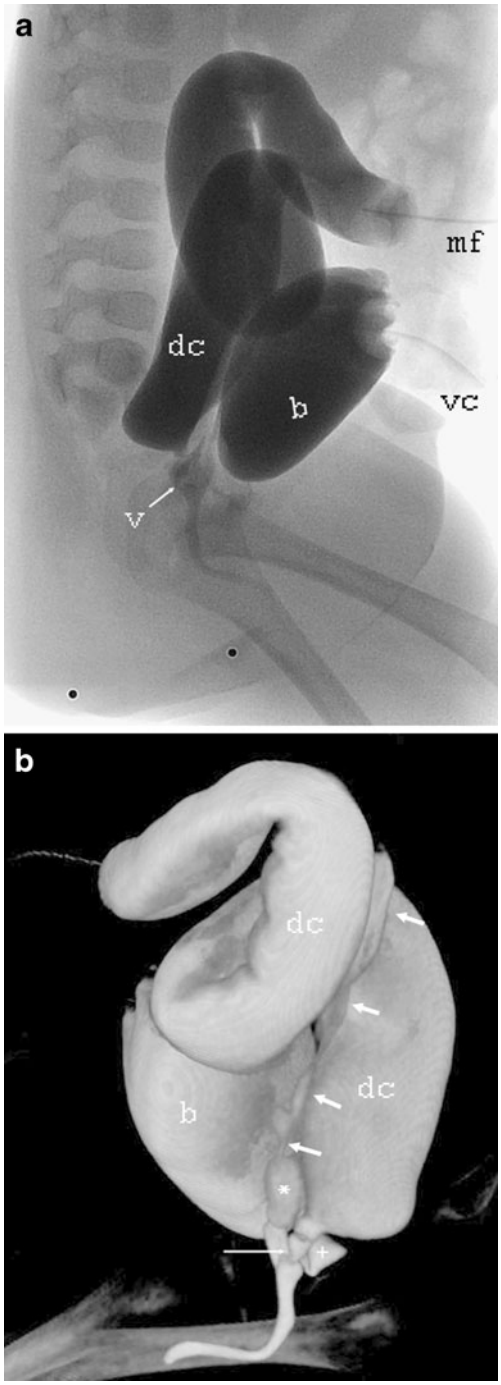

Fig. 9 This 6-month-old had a diverting colostomy with creation of a vesicostomy after birth. Lateral image from a conventional cloacagram (**a**) Lateral image from a conventional cloacagram shows the bladder (*b*) and distal colon (*dc*). There is suggestion of small vaginal structures (*v*). (**b**) Reconstructed left sagittal 3-D image from the cloacagram shows two small hemi-vaginas (* = left, + = right). An ectopically inserting left ureter (*thick arrows*) on the left hemi-vagina is suggested. The reconstructed image also shows the insertion of the rectal fistula (*thin arrow*) on the common channel. The common channel measures 4 cm. From this study, it was determined that the child needed a laparotomy. Surgically, the left ectopic ureter was confirmed and removed due to an associated dysplastic left kidney. A neovagina was created using terminal ileum and the rectum was able to be pulled through. The common channel was left to function as the urethra. (*mf* = Foley catheter placed through mucous fistula, *vc* = Foley catheter placed through vesicostomy)

ureterostomy, etc.). It also allowed for more accurate predictions of long-term prognoses.

A disadvantage to performing rotational fluoroscopy and 3-D reconstructions is that the child must be placed under general anesthesia for the examination; however, we now combine the surgical endoscopy and the 3-D fluoroscopy imaging so that they are both done in the same room and require only one episode of anesthesia. At this point, the surgeons continue to feel that the endoscopy is vital, especially for visualizing how far back the vagina lies on the common channel, and its width and depth at that location; this spatial understanding is key for planning whether a total urogenital mobilization would work, vs. leaving the common channel to become the neourethra and separating the vagina from the urinary tract. However, as the surgeons become more comfortable with the accuracy and reliability of the 3-D imaging, they may eventually be able to discontinue performing preoperative endoscopy.

The technique of rotational fluoroscopy to acquire 3-D images may expose the child to more radiation than 2-D fluoroscopy alone. From internal data from Philips Healthcare, 2-D fluoroscopy at the lowest commercially available setting (which is typically the setting we use for cloaca studies) produces 12 mGy/minute, while a rotational scan of 120 images (typically what we choose for cloaca studies) produces 18 mGy. In other words, one rotational acquisition is equivalent to approximately 1.5 min of fluoroscopy at the lowest commercially available setting (internal unpublished data, personal communication, principal clinical scientist, Philips Healthcare). We reviewed the radiation dose for the 34 children who underwent rotational fluoroscopy and 3-D reconstructions for evaluation of a cloacal malformation during the past two years (July 2009 to July 2011). The mean 2-D fluoroscopy DAP was 2,258 mGy/cm^2 (range 242–10,196). The mean rotational fluoroscopy DAP was 1,245 mGy/cm^2 (range 103–5,496). Therefore, the radiation exposure from rotational fluoroscopy was, on average, 36% of the total dose required for the study, while 64% was from 2-D fluoroscopy. In addition, the spatial understanding that was achieved through rotational fluoroscopy and 3-D reconstruction allowed us to decrease the amount of 2-D fluoroscopy that would have been required to approach a similar anatomical understanding.

This technique also incurs additional expense and the technology is not yet available at all pediatric institutions. We, however, feel that the clinical utility far outweighs the disadvantages.

Conclusion

Rotational fluoroscopy and 3-D reconstructions provide the surgeon with precise anatomical information to facilitate accurate surgical planning of a cloacal malformation and to define prognoses regarding long-term bowel, bladder and sexual function.

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