

6.1 Introduction

Advances in imaging technology have been extremely beneficial for the management of patients with anorectal malformations. Every day, we learn about technologic innovations that frequently surpass our imagination. Concurrently with these, the surgeons have to speculate less and less, like in the past [1], and have the privilege of making very precise anatomic diagnosis that allows the planning of a complex reconstruction in a very accurate manner.

In dealing with the spectrum of anorectal malformations, we depend very much on the images obtained by traditional x-rays, ultrasound, CAT scan, and magnetic resonance imaging (MRI). We use all of these images not only in the early stages of our management but actually through the entire life of the patient, since each stage of life of the patient brings new therapeutic challenges that require an accurate diagnosis.

6.2 Prenatal Diagnosis

Every year, more and more, the pediatric surgeon is asked to participate in the diagnostic discussions of babies in utero that have congenital malformations. The reader is invited to read Chap. 3.

Electronic supplementary material Supplementary material is available in the online version of this chapter at [10.1007/978-3-319-14989-9_6](https://doi.org/10.1007/978-3-319-14989-9_6).

6.3 Neonatal Imaging

The first 24 h of life, before making the decision to open a colostomy or to perform a primary repair, represents a window of opportunity to diagnose potential associated defects. The chest film taken during the first 24 h of life also allows us to see the integrity of the thoracic vertebrae and ribs. In addition, it helps us in the diagnosis of esophageal atresia and potential cardiac malformations (Fig. 6.1).

The abdominal x-ray film allows us to see and rule out the possibility of hemivertebrae (Fig. 6.2). The early detection of these types of



Fig. 6.1 Chest film of a child with anorectal malformation associated to thoracic hemivertebrae and esophageal atresia



Fig. 6.2 Abdominal film showing a hemivertebrae

malformations is relevant to establish the functional prognosis in these babies. The abdominal film must include an AP view of the sacrum (Fig. 6.3). It is also important to take a lateral abdominal film that allows a more accurate measurement of the sacral ratio (Figs. 6.3 and 6.4).

Traditionally, the number of sacral vertebrae is counted to evaluate the quality of the sacrum. Most pediatric surgeons agree that when a patient has less than three sacral vertebrae, the prognosis for bowel and urinary control is not good. Many others while discussing the subject of the sacrum use rather nondescriptive terms such as “dysplastic” or “hypoplastic.” There is no question that the presence of the sacrum as well as its integrity is crucial to determine the functional prognosis of the patient.

We found the terms “dysplastic” or “hypoplastic” very inaccurate. In addition, we found that there are cases with five sacral vertebrae, yet very abnormal ones, which results in an extremely short sacrum associated to fecal and sometimes urinary incontinence. We thought that it was necessary to create a more objective way to evaluate

the sacrum, in an effort to determine the functional prognosis of the baby. The result was the creation of the “sacral ratio” (Fig. 4.5 in Chap. 4), [2, 3].

The sacral ratio results from comparing the vertical length of the sacrum with the size of the pelvis of the same patient.

- A. A line is drawn between the most upper portions of the iliac bone in an AP film of the sacrum.
- B. Another line is drawn between both the inferior and posterior iliac spines (Fig. 4.5 in Chap. 4).
- C. A third line is drawn, parallel to the first two lines, touching the lowest radiologically visible point of the sacrum or coccyx.

The distance between lines A and B is measured, as well as the distance between lines B and C. A ratio is created: AB/BC .

We measured this ratio in 100 normal children who had abdominal films taken for other reasons and found that the average ratio in anterior-posterior films was 0.76 and 0.77 in lateral films [2]. Patients with anorectal malformations frequently suffer from different degrees and types of sacral abnormalities. The sacral abnormalities present like a spectrum with ratios similar to normal children, in what we call the good side of the spectrum. However, in the “bad” extreme of the spectrum, we see patients with a sacral ratio of 0.

We found that it is extremely unusual for a patient with anorectal malformation and with a ratio lower than 0.4 to have bowel control. To have a normal sacral ratio is a good prognostic sign, but it does not mean that the patient will necessary have bowel control, since there are other factors that influence the final functional results.

One of the relatively common associated defects in children with anorectal malformations are defects of the radial bone (Fig. 6.5). Sometimes, the defect in the forearms and the hands is very obvious (Fig. 6.5a). Other times, one can see only a slight radial deviation of the hand that would make us suspect this defect.

An ultrasound study is also part of the group of imaging studies that must be done during the first 24 h of life before the baby suffers from abdominal distention. The kidney ultrasound is perhaps the most important part of the evaluation of this baby since about 50 % of them, globally, have

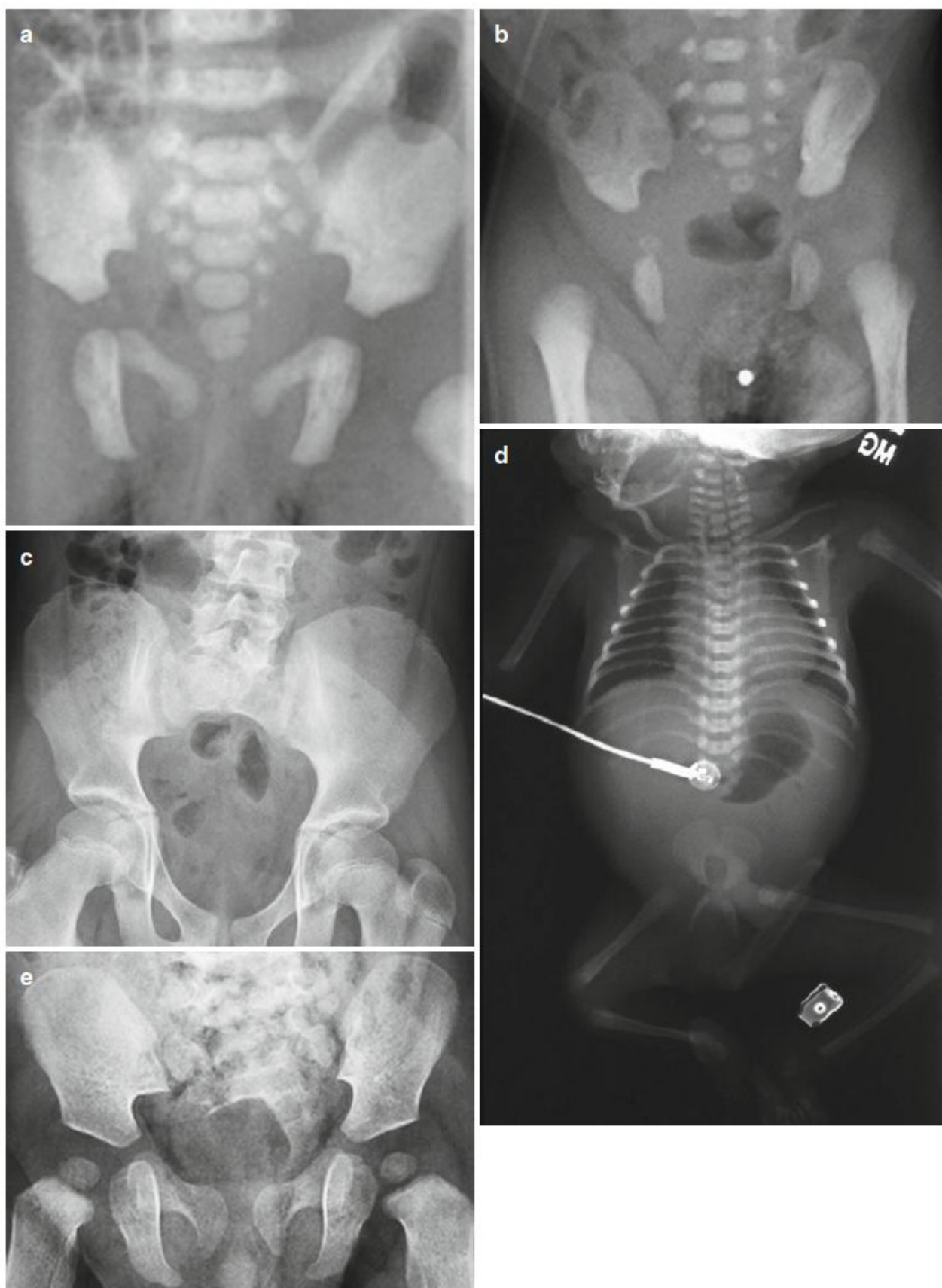


Fig. 6.3 AP film of sacrum in a child with anorectal malformation. (a) Normal sacrum. (b) Short sacrum. (c) Severely deficient. (d) Caudal regression. (e) Hemisacrum

some sort of urologic-associated condition. We specifically look for hydronephrosis (Fig. 6.6). One of the most common urologic anatomic



Fig. 6.4 Lateral film of sacrum

defects is absent or multicystic kidney (Fig. 6.7). The ultrasound must include the rest of the abdomen looking for the presence of megaureters and the bladder. This is particularly useful and important in female babies with a single perineal orifice (cloaca). In these babies, we will specifically look for the presence of a cystic structure located behind the bladder (hydrocolpos). Frequently, this is a double cystic structure since about 30 % of the babies with cloaca have two hemivaginas (Fig. 6.8).

During these first hours of life, it is extremely useful to take an ultrasound of the lumbosacral spine trying to see the conus medullaris to rule out the presence of tethered cord (Fig. 6.9). The presence of a tethered cord represents a negative factor in terms of prognosis for urinary control and to some degree, although not clear, for bowel control. It is a well-known fact that if the ultrasound of the spine is not done during the first 3 months of life, after that time, it is no longer a reliable study for the diagnosis of tethered cord, due to the ossification of the spine, and at that point, the diagnosis of tethered cord can only be done reliably with an

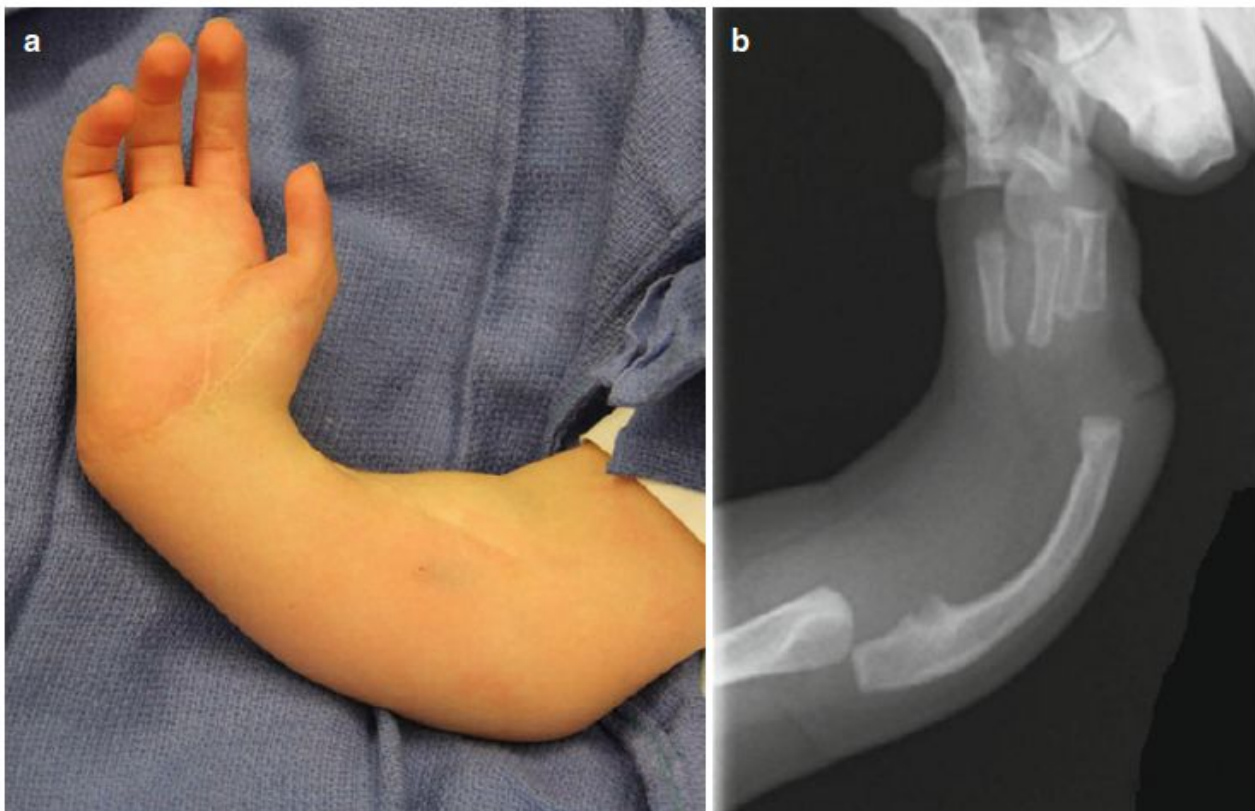


Fig. 6.5 Absent radial bone – a frequently associated defect. (a) External appearance. (b) Radiologic appearance

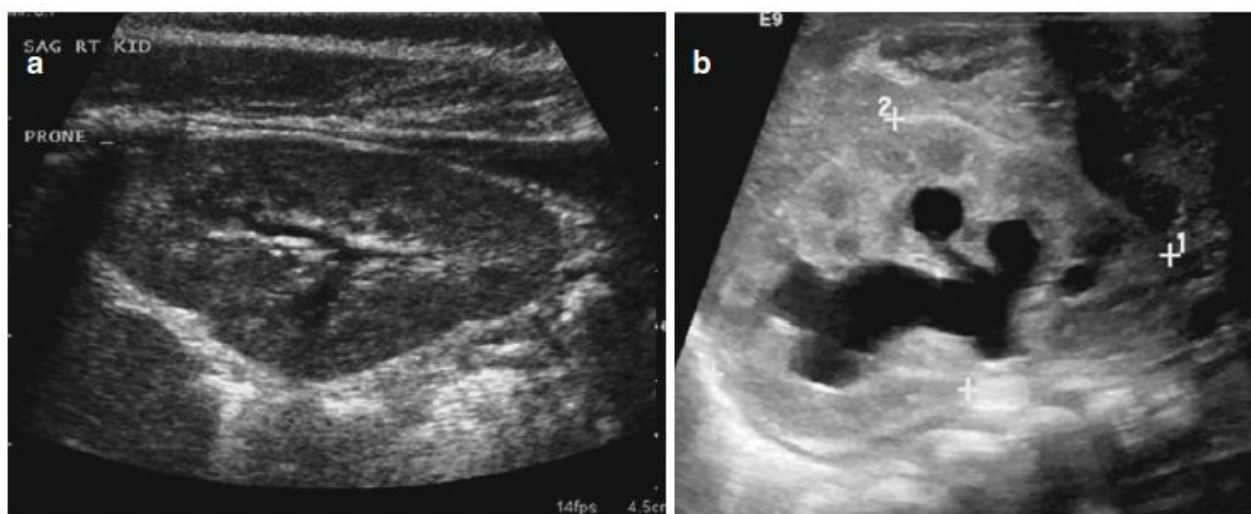


Fig. 6.6 Neonatal ultrasound. (a) Normal. (b) Hydronephrosis

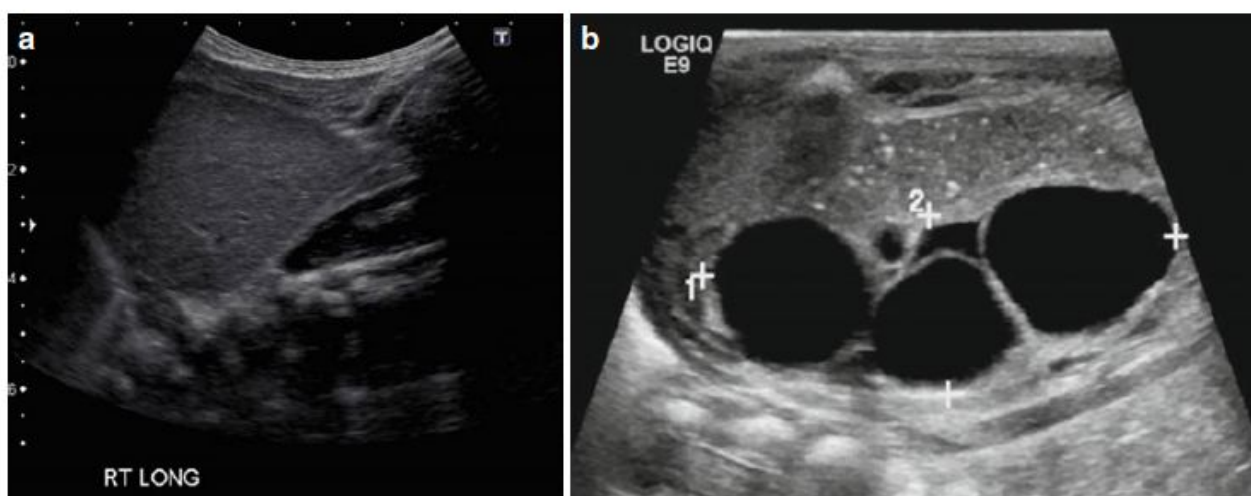


Fig. 6.7 Ultrasound. (a) Absent kidney. (b) Multicystic kidney

MRI study which requires heavy sedation or general anesthesia in babies (Fig. 6.9c).

Some surgeons routinely perform a voiding cystourethrogram in male babies with anorectal malformations. We do not believe this routine is necessary. When the baby has normal kidneys by ultrasound, no evidence of megaureters, and is passing urine normally, we do not see the relevance of the voiding cystourethrogram. Over 80 % of the male patients with anorectal malformations have a connection between the rectum and the urinary tract (fistula), and at the location of the fistula, sometimes there is a kink of the urethra that interferes with the passing of a catheter. Rough manipulations of this baby's urethra in the radiology

department might have negative consequences; sometimes they fall into urinary retention as a consequence of injuries provoked by a failed attempt to pass a catheter. When the baby has hydronephrosis and megaureter, that is when we consider an indication for a voiding cystourethrogram. Other surgeons believe that the voiding cystourethrogram would allow them to determine the size and location of the rectourethral fistula. We considered a voiding cystourethrogram a non-reliable study for the diagnosis of the fistula location. Most of the time that study does not show the fistula. Occasionally, one can see a kink of the urethra that "suggests" where the fistula is located but certainly is not considered a reliable study (see Fig. 6.10).

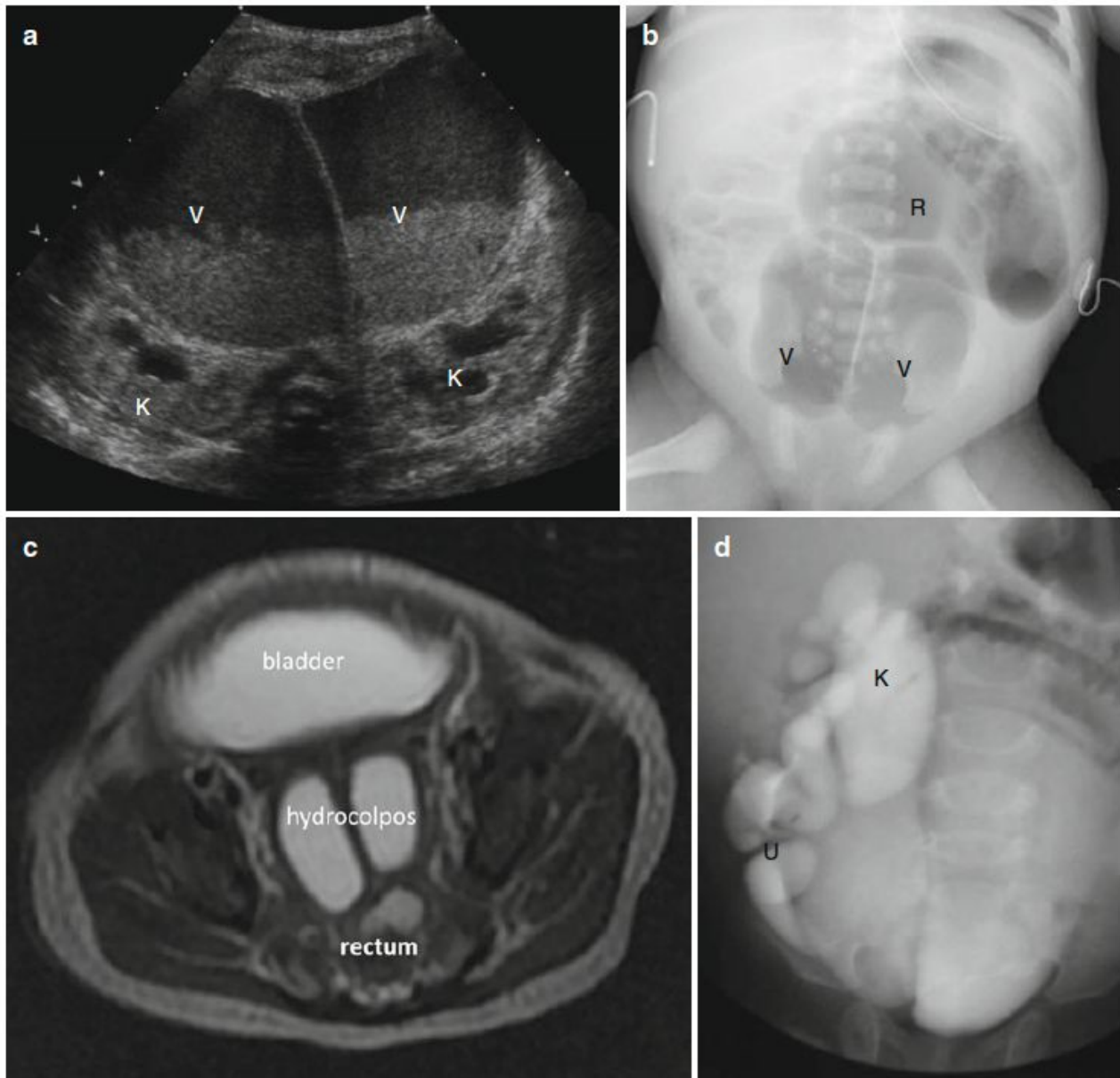


Fig. 6.8 Neonatal hydrocolpos in a newborn baby with hydronephrosis. (a) Ultrasound. *V* vagina, *K* kidney. (b) Abdominal film. *V* vagina, *R* rectum. (c) MRI – transverse section. (d) Abdominal film with contrast. *K* kidney, *U* ureter

6.4 Determination of the Fistula Location Prior to the Colostomy

6.4.1 Anatomic Facts and Timing

To understand the rationality of the imaging during the neonatal period to determine the location of the rectum and the fistula, it is extremely important for the clinician and the radiologist to understand the anatomy of the pelvis of babies with anorectal malformations, this is illustrated in Animation 6.1.

The sphincter mechanism in babies with anorectal malformations is represented by a

wide spectrum that goes from almost normal striated sphincter mechanism to almost absent sphincters.

The sphincter mechanism in normal individuals is represented by a funnel-like voluntary muscle structure, the upper limits of that funnel being the pubococcygeal line (Fig. 6.11). That funnel-like muscle mechanism is a continuum of a striated muscle that runs all the way down to the skin of the perineum. The upper part of the funnel-like mechanism inserts in the pubic bone and surrounds the rectum. The contraction of those fibers compresses the rectum from behind. During surgical explorations, there is no way to identify

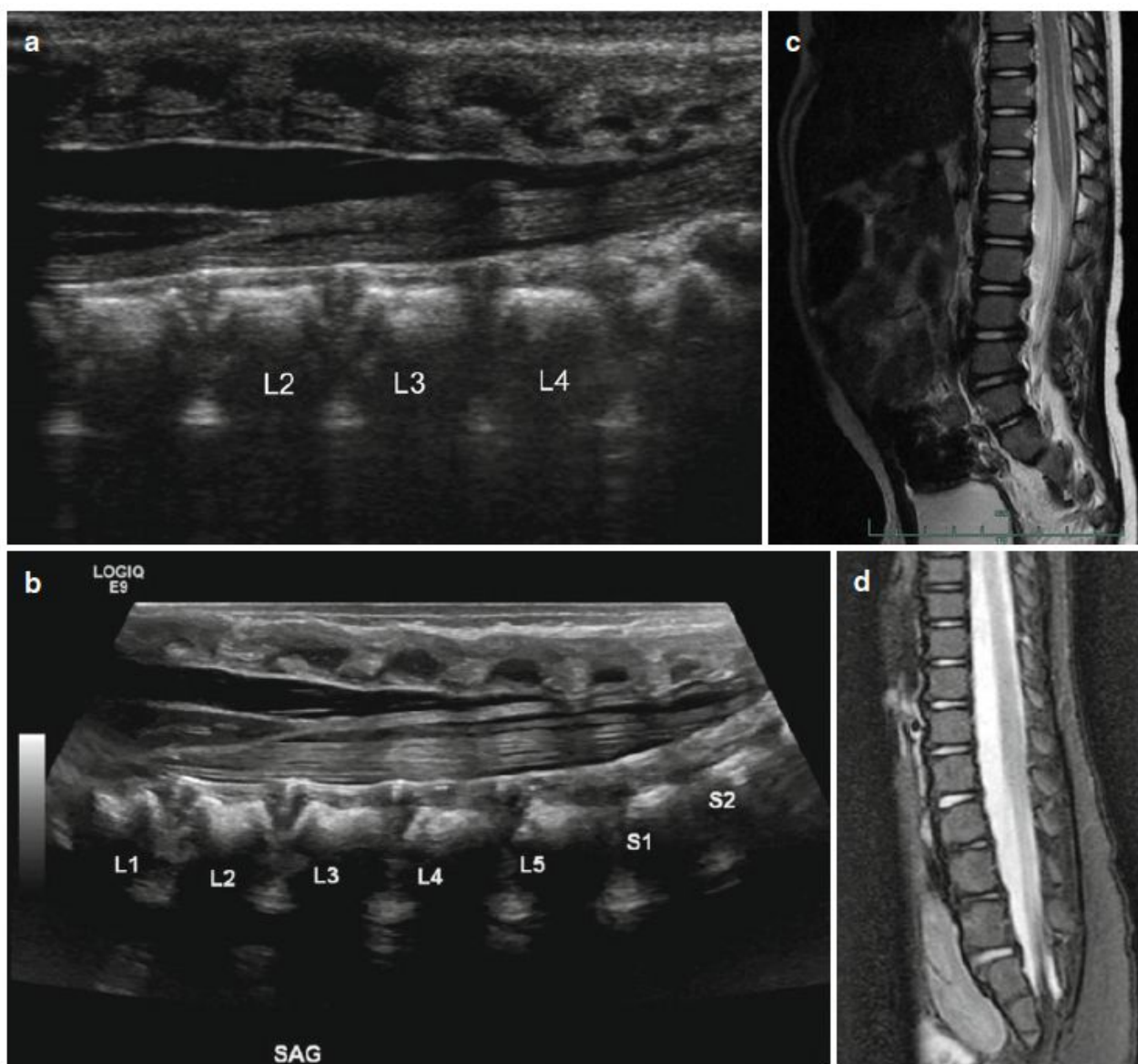


Fig. 6.9 Spinal ultrasound. (a) Normal location of the conus. (b) Tethered cord, ultrasound image. (c) Normal, MR image. (d) Tethered cord, MRI image



Fig. 6.10 VCUG showing a kink in the urethra suggesting the fistula location

separated portions of that muscle that has been referred to as “levator mechanism,” “puborectalis muscle,” “ischiococcygeal muscle,” “pubourethralis muscle”; one rather sees only a continuum of musculature.

In cases of anorectal malformations, the rectum is passing through this funnel-like muscle mechanism and stops at different heights. In cases of perineal fistulas, for example, most of the rectum is passing through this muscle mechanism and is only anteriorly deviated in the lowest portion (Fig. 6.12). In rectourethral fistulas, most of the rectum also passes through this funnel and ends into the upper part of the posterior urethra (prostatic fistula) or into the lowest portion of the posterior urethra (rectourethral bulbar fistula)

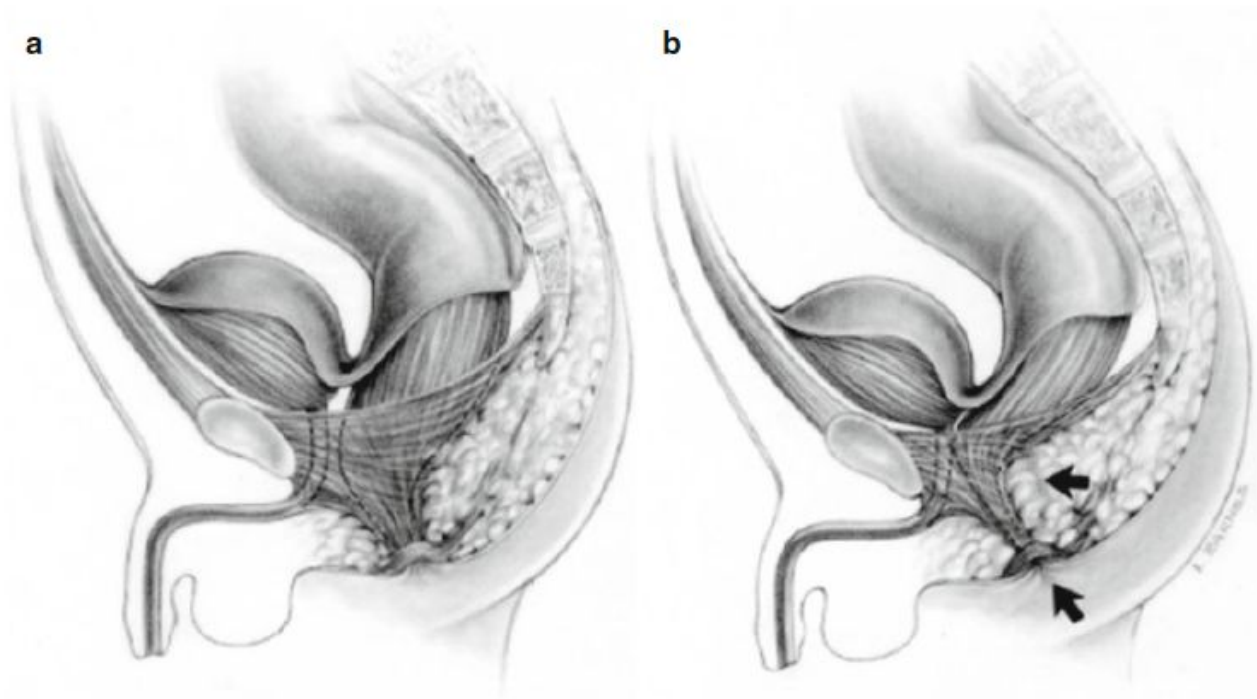


Fig. 6.11 Funnel-like normal sphincter mechanism. (a) Relaxed. (b) Contracted



Fig. 6.12 Diagram of a perineal fistula. Most of the rectum is surrounded by the funnel-like sphincter mechanism

(Fig. 6.13). In cases of recto-bladder neck fistula which represents the highest of all defects in male patients, the rectum opens in the bladder neck and is not surrounded by this sphincter

mechanism (Fig. 6.14). Strictly speaking and using the old terminology, this particular defect is the only one that we can call “supralelevator malformation.”

The funnel muscle mechanism has, as expected, a muscle tone that keeps the rectum collapsed. This muscle only relaxes in normal individuals during the evacuation of feces. If one takes an abdominal x-ray film of a normal newborn with no anorectal malformation, it would be easy to see that the gas in the rectum stops at the level of the pubococcygeal line (which is the upper limit of the funnel-like sphincter mechanism). From there down to the skin, the rectum remains collapsed due to the tone of the muscle that surrounds it. In cases of anorectal malformations, if one takes an abdominal x-ray film during the first few hours of life, we will never find the gas of the rectum located below the pubococcygeal line and certainly that does not mean that the baby has a “very high malformation” since most likely (90 % chance), the rectum is located below the pubococcygeal line but is compressed by the sphincter mechanism. Interestingly, in our literature review, we only found one author [4]

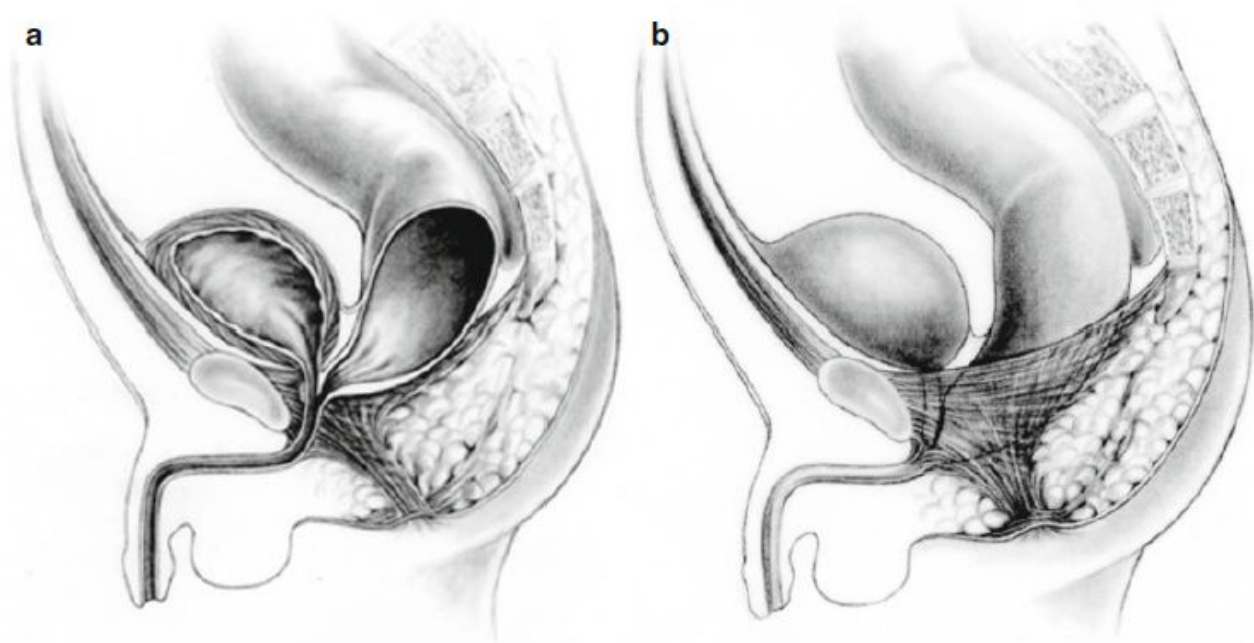


Fig. 6.13 Diagram showing a rectourethral fistula. (a) Prostatic. (b) Bulbar



Fig. 6.14 Diagram showing recto-bladder neck fistula. The bowel is not surrounded by sphincter muscle

who suggested that the contraction of the “puborectalis muscle” must be taken into consideration to interpret radiologic studies in the newborn.

We are convinced that:

Diagnostic Imaging Studies performed during the first few hours of life are not reliable to determine the real location of the rectum

The diagnostic challenge during the newborn stage, in patients with anorectal malformations prior to the opening of a colostomy, is not related so much to the quality and sophistication of the imaging technology used, but rather to the knowledge of the anatomy and physiology of the rectum and the surrounding sphincter in patients with anorectal malformations during the first few hours of life. That is the reason why we recommend not doing diagnostic studies trying to determine the location of the rectum during the first 24 h of life. We have learned that babies with anorectal malformations usually are not born with abdominal distention. They rather become distended after 20 or 24 h of life. This abdominal distention represents, as expected, an increase of the intraluminal pressure of the bowel, and at some point, that pressure overcomes the muscle tone of the funnel mechanism that surrounds the rectum, and then one can see the real location of the gas inside the rectum (Fig. 6.15).

We have been exposed to patients that are referred to us after failed attempted repairs. Some

the baby carefully, we actually need this kind of film (cross-table lateral film) only in less than 5 % of our patients. Most of the times, we obtain enough clinical information to make a good therapeutic decision without this study (see Chap. 4).

Some authors are very enthusiastic about the use of perineal ultrasound in neonates to determine the location of the rectum [9–14]. Others use to recommend the injection of contrast material through the perineum [15, 16] or through the perineal fistula [17]. We feel that these studies are very much dependent on the degree of experience of the radiologist; the images are not easy to interpret for us surgeons.

The CT scan has also been used to determine the location of the rectum, in order to plan the best possible surgical approach [18–21]. Unfortunately, those studies show only transverse section images, and the sagittal reconstructions show poor-quality images.

The MRI technology obviously represents a great advancement that contributes enormously to the anatomic diagnosis of multiple conditions. We use these kinds of studies to evaluate the anatomy in patients already operated on. Some authors suggest doing MRI studies in newborns with anorectal malformations [22, 23]. Even when the images are very good, we consider the study logistically demanding, expensive, sometimes risky for the baby (anesthesia), and not indispensable.

More important is the fact that none of the authors that we reviewed mentioned what we consider is the most important aspect of the neonatal diagnosis, which is the timing of the studies and its relationship with the anatomy and physiology of the rectum and surrounding sphincters in the newborn.

There are multiple papers that recommend other imaging studies (before the colostomy's opening) to try to determine the location of the rectum. None of them discuss the anatomic facts presented here. Some authors claim that an MRI is good enough [3]; others believe it is the CT scan [3]. Finally, some authors propose perineal ultrasound [5], and others prefer the injection of contrast material through the perineum [6]. If a specific doctor or hospital uses this kind of technology for this diagnosis, they should take in



Fig. 6.16 Long narrow fistula. Contrast injected through a perineal fistula. Gives the false impression of a “high” anorectal malformation

consideration the anatomic facts already discussed. Otherwise, those studies are not considered reliable.

When the baby has a tiny orifice in the perineum (perineal fistula), some surgeons pass a fine catheter through the fistula and inject contrast material. They find frequently a long narrow tract with a dilated rectum located up in the pelvis; based on that, they may think that the baby has a “high” malformation (Fig. 6.16). However, that is not a reliable study because what they consider a long narrow fistula may be just a consequence of the compression of a normal-caliber rectum given by the surrounding muscle mechanism, and the rectum is actually located very low in the pelvis (Fig. 6.16).

6.6 High-Pressure Distal Colostogram

After the colostomy has been opened, the surgeon must plan the best surgical strategy to repair the anorectal malformation. We have found through the years that this study is by far the most

important, valuable, and accurate diagnostic test that we can do in cases of anorectal malformation (3D Animations 6.2, 6.3, and 6.4 illustrate this study). Unfortunately, there are not many publications advocating this study [24–28], and many radiologists are not familiar with this study and the way to do it. We believe that we cannot over-emphasize the value of this diagnostic test and the technical details of its performance.

Some authors still believe that a voiding cystourethrogram is good enough to demonstrate the location of the rectourethral fistula [29]. We do not agree because we have seen many false negatives. Attempts have been made to combine distal colostogram with an MRI [30, 31]; there is no question that the images are good, but the study is expensive and logistically difficult.

6.7 Technique

This study is only feasible in patients that already have a colostomy. The baby is taken to the radiology suite and a no. 8 Foley catheter is introduced through the mucous fistula of the colostomy. Many babies, unfortunately, are subjected to a loop type of colostomy, and this makes it difficult, sometimes for the radiologist, to find in which direction the catheter should be introduced to be sure that the contrast material is injected in the distal bowel. The mother usually knows which one is the orifice that produces stool and the one that is just a mucous fistula. The catheter is inserted about 5 cm, and the balloon is inflated with 2 mL of air or water. Traction is applied on the catheter to be sure that the balloon impacts on the stoma serving as a plug that will prevent the contrast from leaking out, even after we apply hydrostatic pressure during the injection. We only use water-soluble contrast material. The injection of the contrast is done using a 60 mL syringe with a catheter tip connected to the Foley catheter and is done by hand. Under fluoroscopic control, with the baby in supine position, the injection starts and continues until the contrast seems to reach the most distal part of the bowel. At this point, we obtain very valuable information related with the length of bowel that the baby

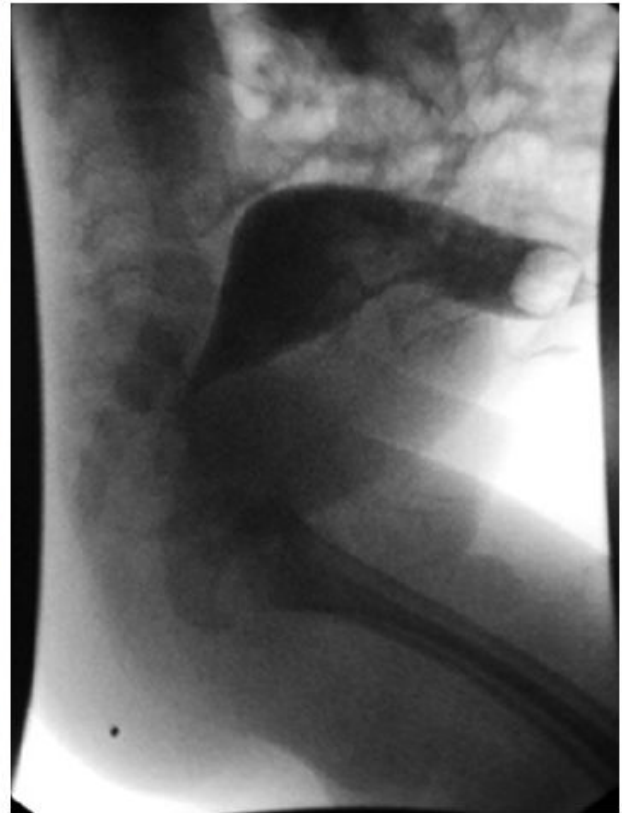


Fig. 6.17 Distal colostogram showing a very short piece of bowel distal to the colostomy pull-through is not feasible

has available for the pull-through, from the colostomy site to the most distal end or fistula site. This is extremely important, since the most common error that we have seen in the location of the colostomies is a too-distal colostomy, meaning that the surgeon left a really short piece of bowel beyond the colostomy site, which will certainly interfere with the pull-through. Trying to repair an anorectal malformation without this crucial information frequently ends in a disaster. Figure 6.17 shows a very short piece of bowel distal to a colostomy. Figure 6.18 shows a colostomy done in the descending colon leaving a loop of the sigmoid colon long enough to perform a comfortable pull-through. Once we obtained that information, the patient is then turned into meticulous lateral position. A lead marker is placed in the anal dimple. During the fluoroscopic study in lateral position, the images must include the sacrum, the coccyx, the lead marker of the anal dimple, and the entire lower pelvis. The radiologist must know that we are planning on a potential



Fig. 6.18 Distal colostogram showing a redundant piece of bowel distal to the colostomy pull-through is feasible

posterior sagittal approach, and therefore, we want to know the position of the rectum and fistula as related to the coccyx and the perineal skin (anal dimple). A common error from a radiologist is to show only the fistula location without the other points already mentioned that would orient the surgeon in terms of the location of the fistula. The injection continues, applying hydrostatic pressure on the syringe and pulling on the Foley catheter to avoid leakage of the contrast (Animation 6.2). The contrast material runs through the distal bowel and stops in a horizontal line that corresponds to the pubococcygeal line (Fig. 6.19). The fact that this line is horizontal and the fact that it is located in the pubococcygeal level indicate that it is not the end of the rectum. That image is given by the contraction of the “funnel-like” muscle mechanism. At that point,

the radiologist must exert enough hydrostatic pressure to overcome the muscle tone of the funnel mechanism to be able to distend the rectum and see the real location of the end of the bowel and the fistula site (Fig. 6.19). If the baby happens to have a recto-bladder neck fistula, the contrast goes into the bladder directly in a rather easy way with minimal hydrostatic pressure. On the other hand, if the rectum is connected to the prostatic urethra and even more in cases of rectourethral bulbar fistula, it requires a significant hydrostatic pressure in order to force the contrast material through the rectum surrounded by muscle tone and through a tiny orifice communicating with the urethra. Figure 6.20 shows characteristic images of a prostatic fistula and a rectourethral bulbar fistula.

Interestingly, in cases of rectal urethral fistula, the contrast material gets into the urethra and usually goes up toward the bladder rather than toward the penile urethra. We interpret this as a manifestation of an increased muscle tone of the so-called external urinary sphincter (Animation 6.2). The overwhelming majority of rectal urinary fistulas are located above this external sphincter, and we believe that is the reason why the contrast goes rather toward the bladder. The injection of contrast material must continue until the bladder is full. By doing this, we now have a cystogram, and if the baby has reflux, it will become then evident. The injection continues until the baby is forced to void. During the voiding, we have the best possible images of the anatomy of the rectum and the urinary tract (Fig. 6.20). Babies with suspected urinary tract problems, such as hydronephrosis and evidence of reflux, must receive prophylactic antibiotics prior to this study because we must keep in mind that we are pushing the contrast passing through a contaminated distal rectum.

The high-pressure distal colostogram, as most studies, has some potential risks. In over 1,000 male patients we have operated, we have evidence of two perforations of the bowel that occurred during the injection of contrast material. One of them had a transverse colostomy. We do not advocate the use of transverse colostomies in anorectal malformations, and one of the reasons

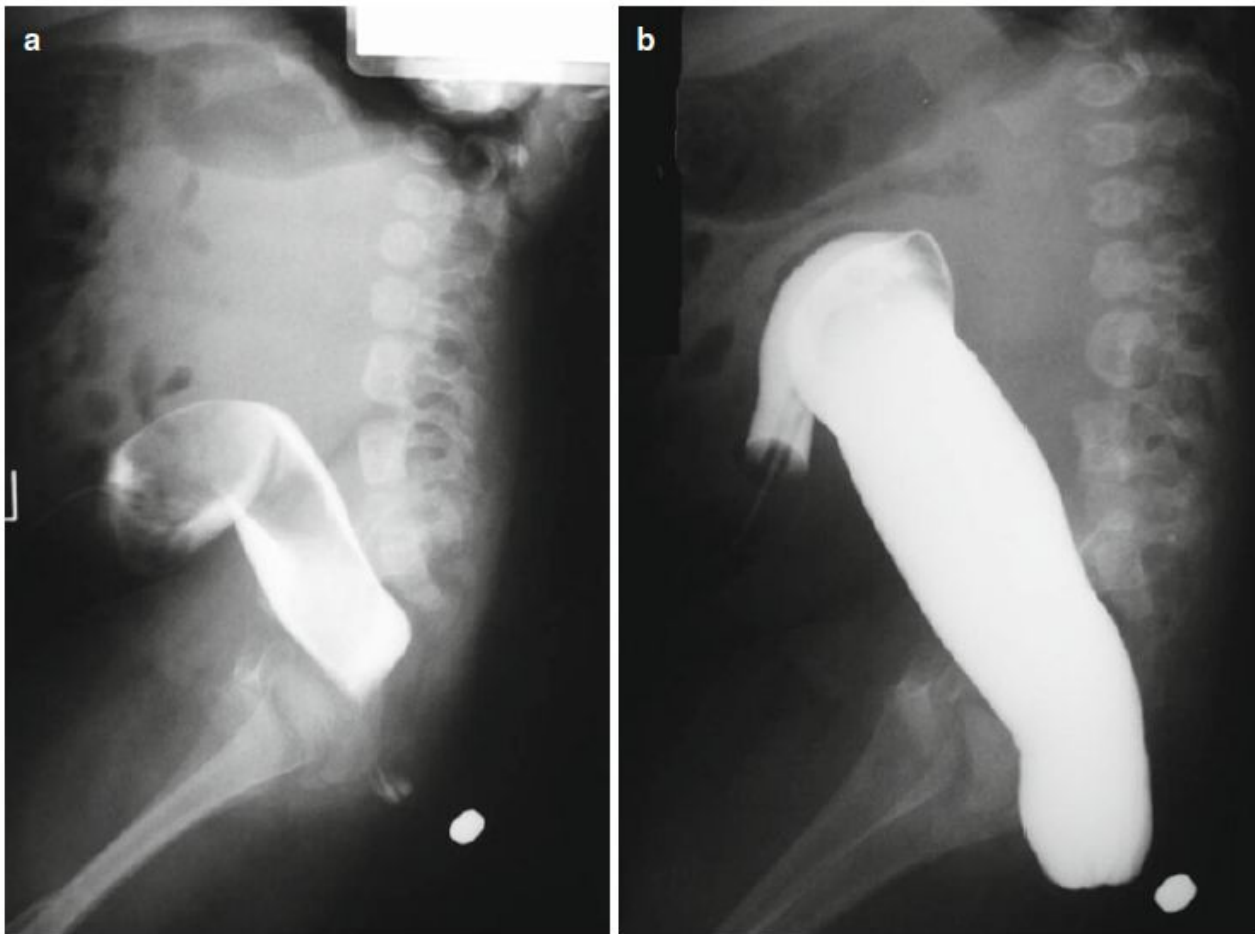


Fig. 6.19 Distal colostogram showing (a) the contrast material ending in a horizontal line at the level of the pubococcygeal line, giving the wrong impression of a

“high malformation.” (b) Same study after applying more hydrostatic pressure

is because it is very difficult to generate enough hydrostatic pressure through a transverse colostomy to be able to achieve our goals in trying to determine the anatomy of the recto-urinary fistula. The colostomy in cases with transverse colostomies requires much higher pressure and perhaps that is the reason why that patient had a rupture of the colon during the injection of contrast material. Since we were dealing with a defunctionalized portion of the bowel, we thought that what all the baby would require would be intravenous fluids, antibiotics, and observation. However, to our surprise and alarm, that baby went into severe hypovolemic shock that required immediate resuscitation followed by a laparotomy to clean the peritoneal cavity and close the perforation of the colon. We attribute this severe reaction to the fact that the contrast is very hyperosmolar. The second case was a patient that had a

very abnormal type of bowel with very abnormal blood supply, and we believe that could be a predisposing factor to explain the perforation, although we do not have evidence of that.

6.8 Most Common Errors

Some patients come to us with a distal colostogram that has been done in another institution. We have seen many errors including passing the Foley catheter too far (Fig. 6.21). This may give the false impression that the patient has a very short piece of bowel distal to the colostomy. That is why we emphasized that the catheter should be introduced only 5 cm and then pulled back to ensure that the balloon is impacted against the abdominal wall.

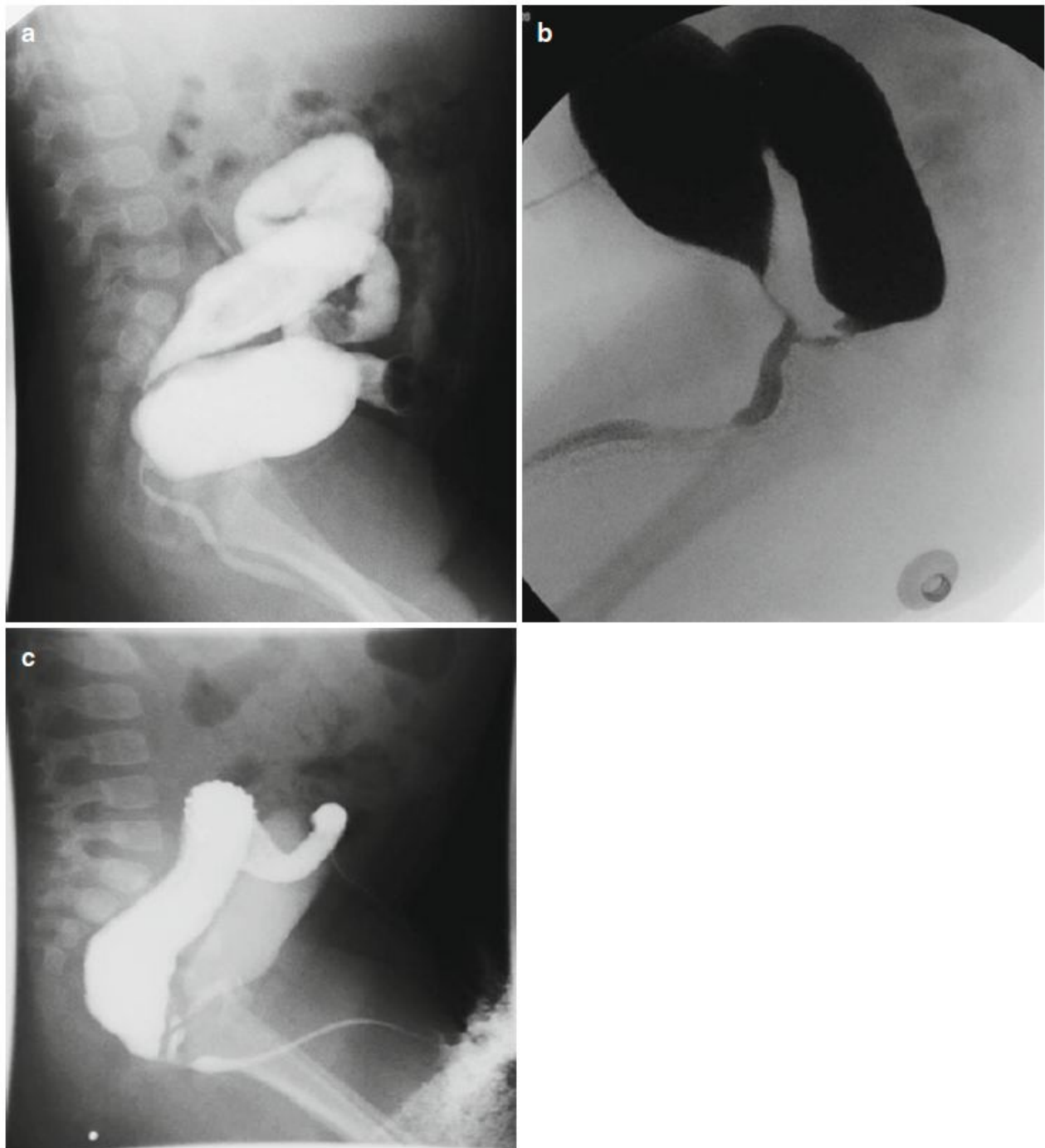


Fig. 6.20 Distal colostogram showing a (a) recto-bladder neck fistula, (b) rectoprostatic fistula, (c) rectourethral bulb-bar fistula

By far, the most common error that we have seen is the lack of hydrostatic pressure that induced the surgeon to believe that the patient had a “high malformation” when actually he was looking at a characteristic image of the contrast material stopping at the level of the pubococcygeal line.

6.9 Not Showing the Coccyx and the Sacrum During the Fluoroscopy Studies

The radiologist must remember that this study is done with the specific purpose to determine how reachable the rectum is through a posterior

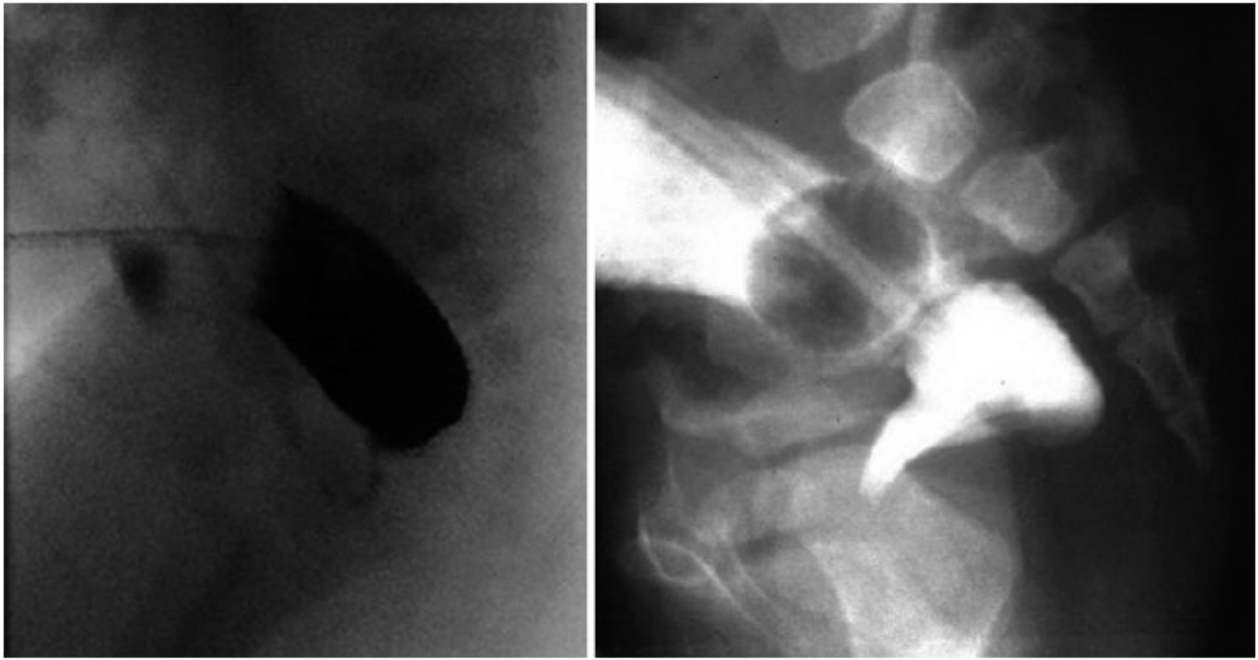


Fig. 6.21 Inadequate distal colostogram. The Foley catheter was introduced too deep, giving a false impression of a short bowel distal to the colostomy

sagittal incision. Therefore, our main point of reference is the coccyx. Sometimes the radiologist, trying to avoid unnecessary radiation to the baby, uses diaphragms or cones that only show the fistula site, but do not show the points of reference that are the pubis, coccyx, and sacrum as well as the lead marker in the anal dimple. Another common mistake is not to take the film on perfect lateral position. Some radiologists were trained mainly in adults; in those cases, to study the urethra, they recommend an oblique view. This is highly inconvenient in patients with anorectal malformation because of the reasons already explained.

When the babies were subjected to a transverse colostomy and did not have the main repair for a long period of time, the distal colostogram shows a very characteristic image of a non-used distal colon (microcolon) followed by an extremely dilated rectosigmoid (Fig. 6.22). That is another reason why we do not like transverse colostomies. After the transverse colostomy has been opened, the colon continues having peristalsis, passing mucus and desquamation cells, and pushing all that material distally. All these produce a characteristic megacolon as the one seen in Fig. 6.22. In addition, the transverse



Fig. 6.22 Distal colostogram done through a transverse colostomy. It shows a characteristic image of a narrow, nonused colon, followed distally by a very dilated rectum

colostomy does not allow cleaning the distal colon during the newborn period, and the meconium is left there. We have evidence that the severity of megarectosigmoid correlates with



Fig. 6.23 Distal colostogram in a patient that previously had an attempted failed repair

the degree of constipation that the patient will suffer later in life.

Some patients come to us after a failed attempted repair. They suffered from catastrophes and misadventures during an attempted operation. They still have the colostomy, and we had to do a distal colostogram without knowing what we are going to find. We may find that the rectum is completely atretic (Fig. 6.23). We may find also that the patient has also persistence or recurrent rectourethral fistula (Fig. 6.24).

6.10 Distal Colostogram in Female Patients

In the past, we used distal colostogram also in female patients with vestibular fistulas and sometimes with perineal fistulas. We do not recommend doing that kind of study because we consider that it is not useful for the diagnosis and treatment of the patient. The injection of contrast material through the distal stoma in that kind of patient would show an image similar to a long narrow fistula, and even if we apply more hydrostatic pressure, we never are able to distend the



Fig. 6.24 Voiding cystogram showing a recurrent rectourethral fistula

distal rectum because the contrast material escapes through the fistula site and one cannot generate enough pressure to show the real size of the rectum (Fig. 6.25). These may give a false impression of a long narrow fistula when actually the rectum is distended all the way down to reach a point a few millimeters from the skin. A real long narrow fistula is an extremely unusual condition.

6.11 Distal Colostogram in Cloacas

In patients born with cloacas, the distal colostogram is still a very valuable study, but this is true particularly if it is combined with a three-dimensional rotational scan.

Depending on the specific anatomy of the cloaca, the distal colostogram may show only the rectum followed by the common channel (Fig. 6.26) or may show also the one vagina or two vaginas (Fig. 6.27). It is extremely unusual to be able to fill up the urethra and bladder through a distal colostogram. The study in cloacas can be



Fig. 6.25 Distal colostogram in a patient with rectoves-tibular fistula, giving a false impression of a long narrow fistula



Fig. 6.27 Distal colostogram in a patient with a cloaca showing the rectum and the vaginas



Fig. 6.26 Distal colostogram in a patient with a cloaca showing only the rectum

done in combination with the passing of a catheter through the common channel, which may go into the urethra and bladder and have an image of those three structures (bladder, vagina, and rectum) (Fig. 6.28).

During the last few years, with the very valuable collaboration of our interventional radiologists, we have been using a 3D rotational scan for the evaluation of cloacas, particularly those that have a complex anatomy. This study is done under anesthesia, and we take advantage of that in order to do a vaginoscopy and cystoscopy. This study is usually done 1–2 days before the main repair. During the vaginoscopy and cystoscopy, we can also insert catheters in a selective manner into the urethra or vaginas to give the radiologist more alternatives for contrast injection. Some of those patients come, already, with vesicostomies or suprapubic cystostomy tubes, vaginotomies, and colostomies. We insert catheters in every single orifice that we can see in the pelvis, and



Fig. 6.28 Distal colostogram combined with injection from below showing the rectum, urethra, bladder, and vagina

then the contrast material is injected sequentially through each one of those catheters, and the scan is done rotationally which gives extraordinary useful images (see videos 6.1, 6.2 and 6.3 showing the rotational studies). This imaging represents the ultimate technology. Cloacas can be repaired without the help of this study, but certainly this type of study allows us to be more precise in our preoperative diagnosis and to plan our procedures in a more accurate way.

6.12 Monitoring Constipation

The most common sequelae in terms of bowel function in patients with anorectal malformation is constipation. Constipation produces dilatation of the rectosigmoid and sometimes the entire colon. When constipation is not treated properly, it produces fecal impaction, and that interferes with bowel control in those patients that are born with potential for fecal continence. Those patients may behave like the child with severe idiopathic

constipation and encopresis. To prevent this, we try very actively to monitor the dilatation of the bowel and the capacity to empty every day. This is with the specific purpose to prevent irreversible dilatation of the colon that would make the management of these patients much more difficult. After the colostomy is closed, the babies have usually a period of frequent bowel movements and tendency to diarrhea. We believe this is a consequence of passing stool through a colon that has never been used. In addition, the use of strong antibiotics administered during the colostomy closure we believe may contribute to this phenomenon.

However, the clinician must be aware of the fact that after a few days or sometimes weeks, the bowel movement pattern changes and must be prepared to diagnose and treat aggressively enough the problem of constipation. We have learned through the years that the fact that the baby passes stool every day does not mean that it is not constipated since the main problem is not the number of bowel movements but the capacity to empty completely the rectosigmoid during the bowel movement. In fact, some of the babies that have multiple bowel movements in a day are the most severely constipated, and eventually those tiny bowel movements become episodes of soiling. After years of speculating and guessing as to whether or not the baby was constipated, we learned that the only objective way to know if the baby is emptying the colon is radiologically.

After the colostomy is closed, we recommend taking routine abdominal x-ray films 2 weeks after surgery, then 1, 3, and 6 months and a year. In addition, every time the mother suspects the child is constipated, we suggest taking an abdominal x-ray film. Every patient needs a different amount of laxative, and to determine whether or not we are using the right amount, we used abdominal x-ray films. In other words, by trial and error, we prescribe a specific amount of laxative and then, after a few days, we take an abdominal film to be sure that the amount of laxative that we are using is the correct amount. Many patients come to us after having an anorectal malformation repair either by us or by

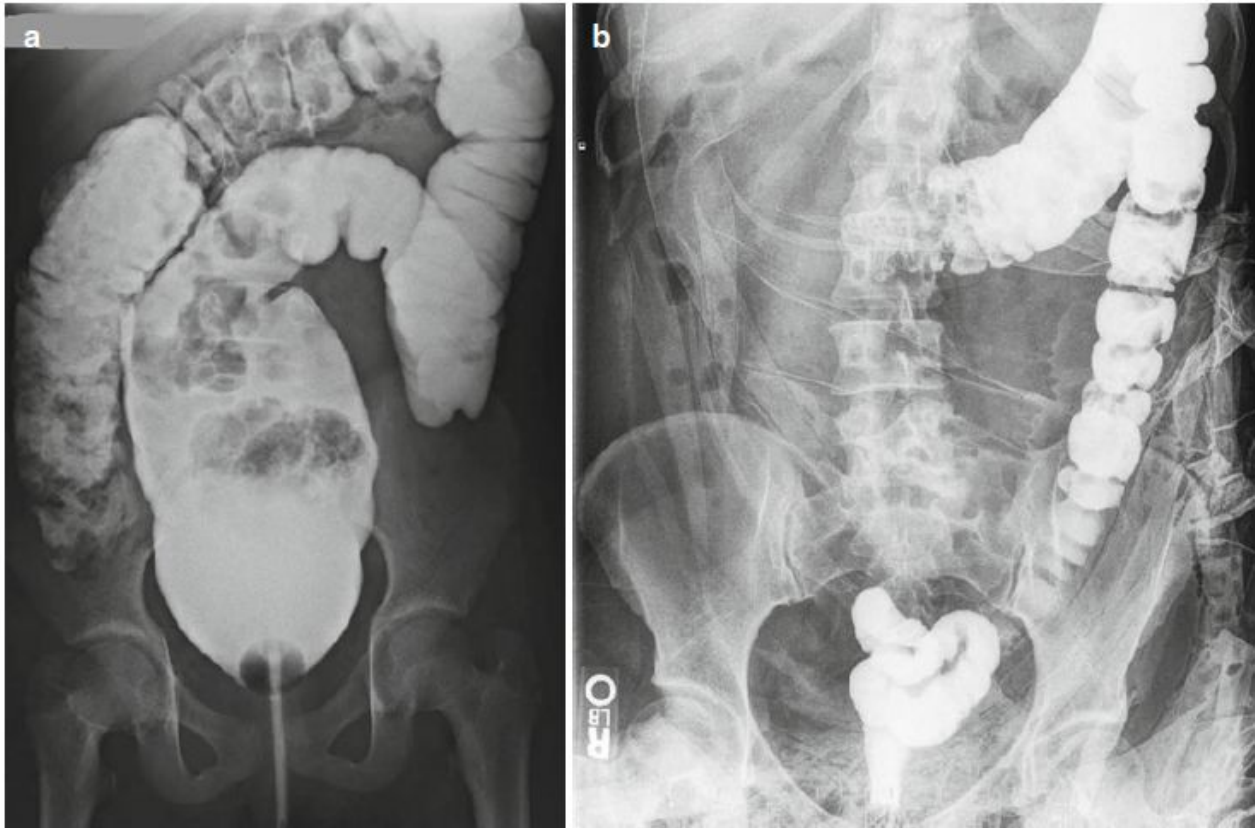


Fig. 6.29 Contrast enema done to evaluate colonic motility. (a) Hypomotility. (b) Hypermotility

others at other institutions, and they were not subjected to any kind of monitoring through years. We suspect clinically that the patients are constipated, and to evaluate the degree of constipation that they suffer from, we order a contrast enema with water-soluble material and without colon preparation. The contrast material must fill up the entire colon, and we request to show us post-evacuation films which give us an idea of the degree of hypomotility that the patient suffers from (Fig. 6.30).

6.13 Radiology During the Bowel Management Program

In Chap. 20, the reader may find a description of our bowel management program, created to help patients who suffer from fecal incontinence. Basically, over a period of 1 week, we determine the enema that is capable of emptying at least all of the left side of the colon, to keep the patient

completely clean in the underwear for 24 h. To do this, we take an abdominal x-ray film every day over a period of 1 week. In addition, also daily we hear the clinical information from the parents and adjust the type of enema accordingly.

6.14 Monitoring the Urinary Tract

As previously mentioned, 50 % of the patients with anorectal malformations have an associated urologic condition. The most common anatomic problem is absent kidney followed by hydronephrosis, and the most common functional problem is vesicoureteral reflux. Many times the babies are born with significant kidney damage that occurred in utero. All patients require a close monitoring of the urinary tract, since we have seen many patients who have a tendency to deteriorate with time. Even under normal circumstances in babies with good anorectal malformations, we always like to follow them through life. In a patient that has normal kidneys

by ultrasound, normal functional bladder, no urinary tract infections, and urinary control, we still like to take a kidney ultrasound at 3 months, 6 months, 1 year, 3 years, and 5 years later. If the patient has no urologic symptoms and the ultrasounds remain normal, we believe that the kidney

ultrasound is enough as a monitoring of the urinary tract. On the other hand, if the baby has problems with anatomic urologic defects previously diagnosed, he/she may require a much closer and sophisticated monitoring of the urinary tract that may include kidney ultrasound,

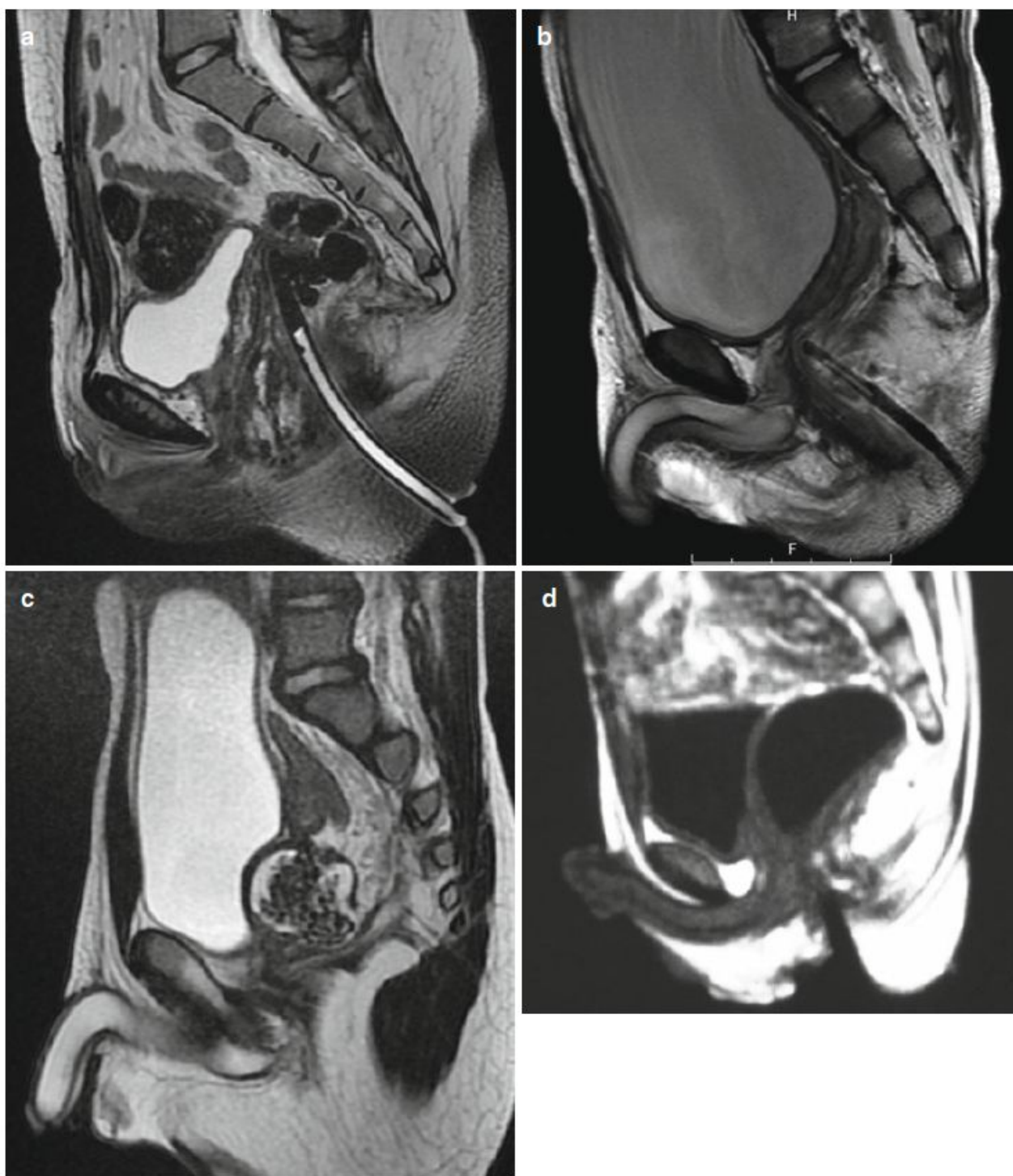


Fig. 6.30 MRI (Peña/Patel technique). (a) Well-located rectum. (b) Mislocated rectum. (c) Posterior urethral diverticulum. (d) Giant posterior urethral diverticulum

cystograms, and urodynamic studies, as well as a frequent nephrologic monitoring. Patients with neurologic deficits including abnormal sacrum, tethered cord, or complex cloacas are, by definition, urologic patients that require very close follow-up monitoring of the urinary function. Many of the complex cloacas eventually will require urinary reconstructions including bladder augmentations and Mitrofanoff.

For patients that have been operated on at other institutions and come to us, for instance, for bowel management due to fecal and/or urinary incontinence, we evaluate the entire colorectal anatomy, pelvic anatomy, and urinary tract. We specifically order a magnetic resonance imaging (MRI) study with a specific technique that includes the placing of a big Foley catheter in the rectum (Fig. 6.30a and b). This will allow us to see the location of the rectum as related to the sphincter mechanism. In addition, it is an excellent study to make a diagnosis of a posterior urethral diverticulum which is a complication that we have seen in the past in patients that were operated abdominoperineally, having a rectourethral bulbar fistula; the surgeons found it difficult to reach the lower end of the bowel, amputated the bowel, and left a piece of rectum attached to the urethra (Figs. 6.30c and d). That piece of rectum becomes a diverticulum that produces mucus, forms stones, and produces pseudourinary incontinence, and in one specific case, the patient developed an adenocarcinoma. With the advent of laparoscopic procedures, we are seeing again this problem of posterior urethral diverticulum, when the surgeons try to approach laparoscopically a rectourethral bulbar fistula, finding difficult to reach the lower end of the rectum Animation 5 (Posterior urethral Diverticulum in a patient with a recto-urethral bulbar fistula approached laparoscopically). The voiding cystourethrogram may or may not show the posterior urethral diverticulum and therefore is not the ideal method of diagnosis. The diagnosis of this entity is confirmed cystoscopically, and the repair is described in Chap. 22.

When the babies come to us for the first time with anorectal malformations and did not have spinal ultrasound during the first 3 months of life, the only way to know the location of the conus

terminalis is by an MRI study that is the most accurate way to determine whether or not the patient has tethered cord (Fig. 6.9c).

Imaging plays a very important role in the diagnosis, follow-up, and management of patients already operated (see Chap. 19).

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