Neonatal Management

4

4.1 Introduction

In dealing with most congenital anomalies, errors that occur during the first few hours or days of life may have serious consequences and sequelae. Early accurate diagnoses, as well as adequate therapeutic decisions, require a high index of suspicion from pediatricians, neonatologists, pediatric surgeons, pediatric urologists, nurses, and other physicians and surgeons who take care of neonatal babies with congenital anomalies.

> Our eyes only see what our mind suspects.

It is hard to believe that in this twenty-first century in developed countries, we still hear of patients born with imperforate anus that were sent home as "normal babies." Subsequently, it was the mother who made the diagnosis or the babies suffer from bowel perforation and some of them die [1–9]. We cannot overemphasize the importance of the anorectal examination during the first physical examination of a neonate. In addition, it is also true what is written in the very old textbooks of pediatric surgery; it is not enough by looking at the external appearance of the anus, but it is rather necessary to introduce a little catheter or thermometer to be sure that the anus is patent. There is one specific type of defect in which the babies have a normal-looking anus externally, a normal anal canal, approximately 1–2 cm deep, and then an atresia. That type of defect is the one that can only be diagnosed by trying to pass a thermometer, catheter, or an instrument through the anus. That particular malformation only occurs in 1 % of all patients born with anorectal malformations [10].

4.2 Most Common Scenario

As pediatric surgeons, we are called to see a baby that is just born and has no anus. There are two very important questions to be answered.

1. Does the baby have a serious associated defect that may kill him/her within the next few hours or days of life?
2. Does the baby need some sort of primary anal repair or a colostomy?

These questions should be answered in the same order that they are presented here. In other words, the surgeon must refrain from jumping into trying to find the answer of the second question but rather concentrate during the first 24 h of life in trying to answer the first question.

4.3 Answering the Two Most Important Questions

At this point, the neonatologist and the surgeon should remember that about 30 % of all babies with anorectal malformation have some sort of cardiovascular condition [11]. However, only one third of that 30 % have serious hemodynamic repercussions that mandate to implement some sort of medical or surgical treatment [11]. The most common cardiac congenital anomalies seen in these cases

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are patent ductus arteriosus, atrial septal defect, ventricular septal defect, and tetralogy of Fallot, and then other more serious conditions are more rarely seen. The presence of cyanosis or respiratory distress should alert the clinician to investigate for the presence of these conditions. At the present time, most neonatal centers have a pediatric cardiologist and an echocardiogram machine which allows ruling out most of these conditions.

Eight percent of babies with anorectal malformations are born with esophageal atresia [12]. Part of the physical examination of the neonate includes the passing of a nasogastric no. 8 feeding tube through the nostril to confirm that the esophagus is patent. When the clinician feels resistance in the passing of the tube about 8–10 cm from the nostril, it is possible that the baby has an esophageal atresia (Fig. 4.1). In addition, babies with esophageal atresia cannot swallow their saliva; this represents a risk of aspiration. The saliva accumulates in their mouth as foam.

Approximately 3 % of babies with anorectal malformations suffer from duodenal atresia [12]. A simple abdominal film shows a classic "double bubble" image (Fig. 4.2). The "double bubble" represents the stomach and the duodenum full of gas: in addition, there is a conspicuous absence of gas in the rest of the abdomen. Approximately 50 % of all patients with anorectal malformations are born with an associated urologic condition [13]. The most serious of these are hydronephrosis, vesicoureteral reflux, absent kidney, and megaureters. One specific group of patients has a very high risk of having kidney damage; those are patients born with a single kidney, hydronephrosis, vesicoureteral reflux, and megaureter. In those cases, we must remember that the single kidney has been suffering in utero, and therefore, we must expect a serious functional limitation. Every effort should be done to protect the damaged kidney. For this, it is mandatory to do a kidney and bladder ultrasound in all babies with anorectal malformation. If the ultrasound shows abnormalities such as hydronephrosis, then the baby will need a full urologic evaluation including, of course, a voiding cystourethrogram. If the kidney ultrasound is normal and the patient is passing urine without difficulty, we must assume

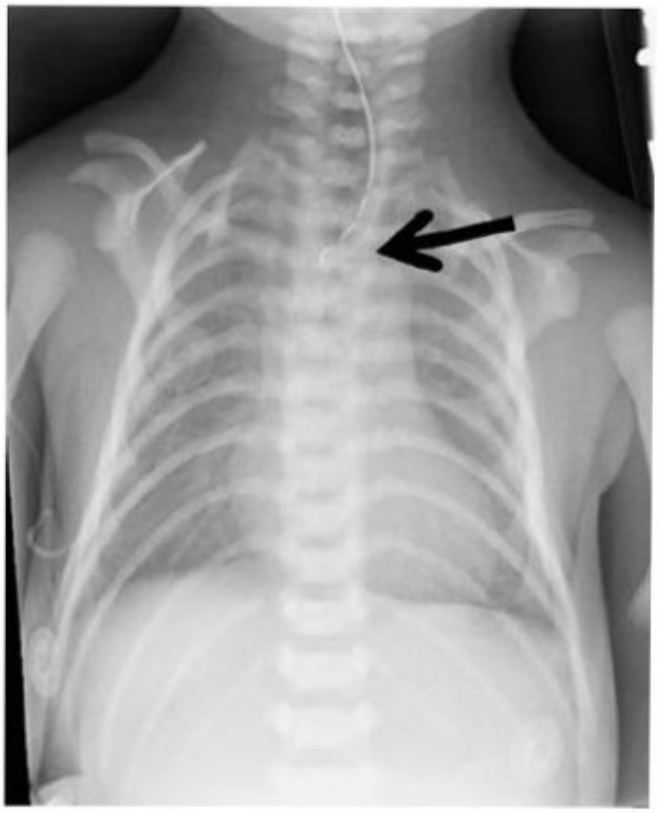

Fig. 4.1 Radiologic image of a baby with esophageal atresia. *Arrow* shows the blind end of the esophagus

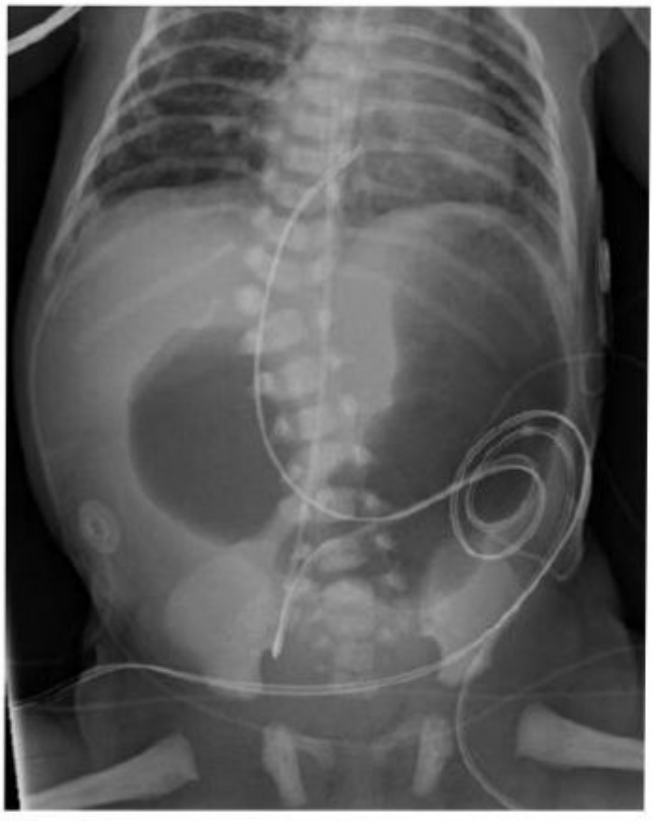

Fig. 4.2 Characteristic radiologic image known as "double bubble" in a baby with duodenal atresia

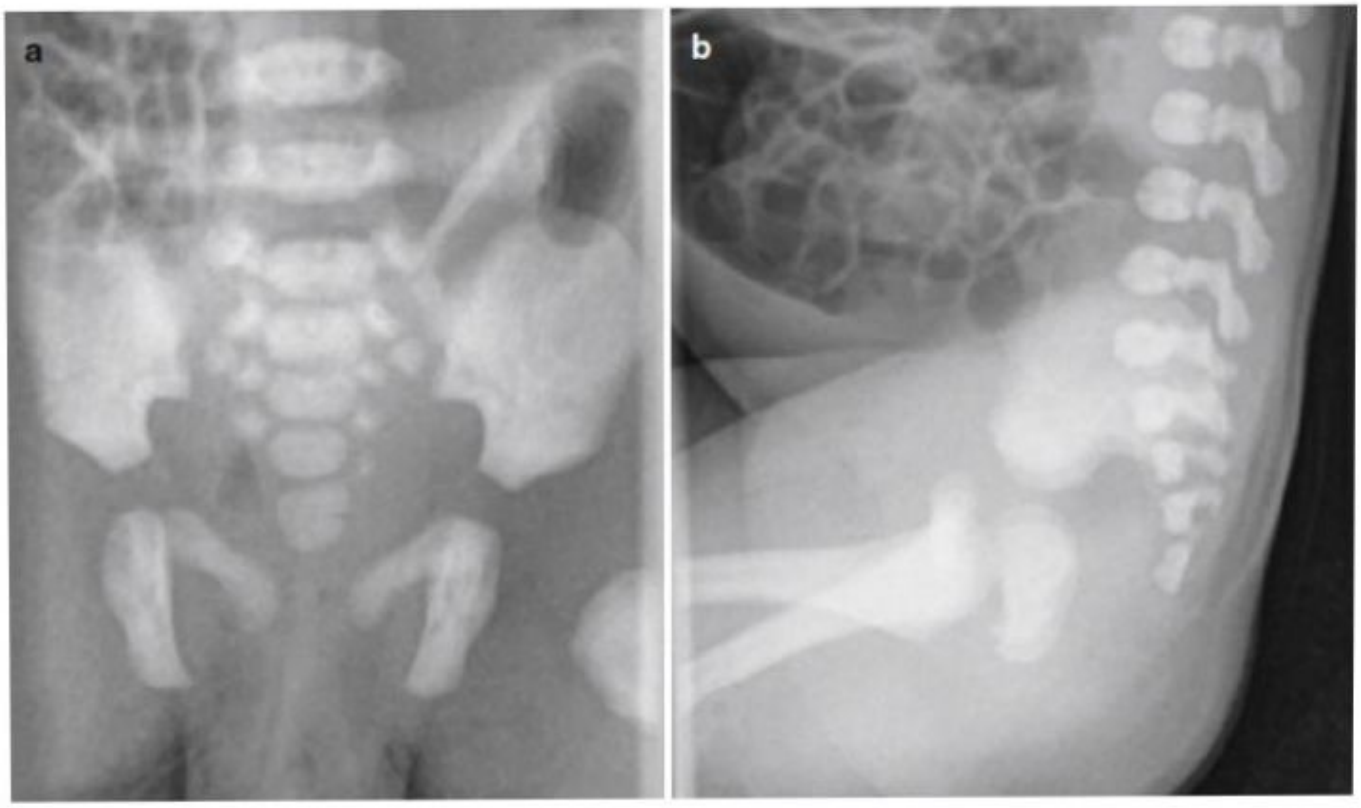

Fig. 4.3 X-ray films of a normal sacrum in a baby with an anorectal malformation. (a) AP view. (b) Lateral view

that most likely his urinary tract is otherwise healthy.

Taking a baby to the operating room to repair an anorectal malformation or to open a colostomy only to find that the baby gets very sick during the operation because he has a serious cardiac, esophageal, or kidney problem is an undesirable and preventable experience.

All babies with anorectal malformations must have an abdominal x-ray film that shows the degree of bowel dilatation, the characteristics of the spine, and the characteristics of the sacrum. The presence of intraluminal calcifications in the rectosigmoid most likely is due to the passing of urine to the bowel. Urine mixed with meconium may produce calcifications [14, 15]. The sacral films must include AP and lateral views (Figs. 4.3 and 4.4). The characteristics of the sacrum are extremely important in order to determine the future functional prognosis for bowel control, urinary control, and sexual function. Traditionally, we evaluated the sacrum by counting the number of vertebrae. We found this to be a rather limited nonuseful way, and therefore, we created what we call the sacral ratio (Figs. 4.5 and 4.6) (see Chap. 6).

Taking x-ray films of the sacrum is mandatory also because we must rule out the presence of a hemisacrum, which means that the patient has a presacral mass; this finding has important therapeutic and prognostic implications.

An ultrasound of the lower sacrolumbar spine is extremely useful and must be done during the first hours of life to determine whether or not the baby has tethered cord (Fig. 4.7). This is also important to determine the functional prognosis for bowel and urinary control.

All these studies can be done during the first 24 h of life. Babies with anorectal malformations are usually not born with a distended abdomen. It takes a few hours for the abdomen to start becoming distended. It is after 24 h of life that the abdominal distention becomes critical, and a management decision must be taken.

4.4 Physical Examination

4.4.1 Male Patients

During our first contact with the baby with anorectal malformation, we should dedicate a special time for a meticulous detailed examination of the baby's perineum. Babies with anorectal

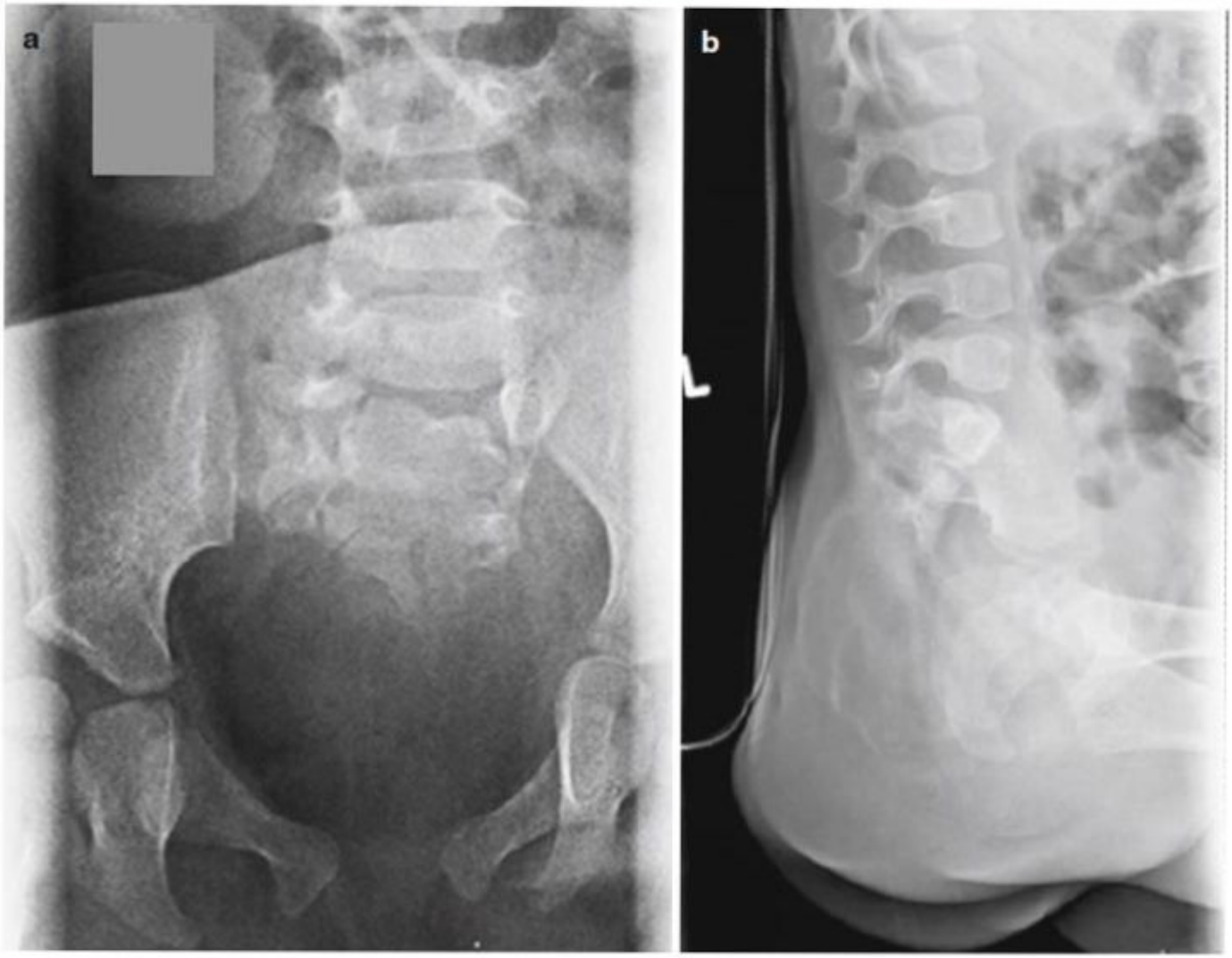

Fig. 4.4 X-ray films of an abnormal sacrum in a baby with an anorectal malformation. (a) AP view. (b) Lateral view

malformations have different external appearances of their perineum, and they have very important clinical significance.

The presence of a flat bottom, meaning absence of the normal midline groove that all human beings have between both buttocks, is usually associated with malformations with bad prognosis and very highly located rectum (Fig. 4.8). In addition, most patients with anorectal malformations have an anal dimple that represents the point in the perineum where the patients have the largest concentration of sphincter fibers (Fig. 4.9). The more prominent the anal dimple, the better the quality of the sphincter and therefore the prognosis. The absence of an anal dimple is a very bad sign, usually present in poor prognosis type of defects. The location of the anal dimple varies from patient to patient. The closer the anal dimple to the scrotum, the worse the prognosis (Fig. 4.10). The longer the distance between the tip of the coccyx and the anal dimple, the poorer the prognosis, either because the anal dimple is located too anteriorly or because the sacrum is very short or both. We call it a "good-looking perineum" when the baby has a well-formed midline groove and a well-located anal dimple (Fig. 4.9a). Even by touching that area, one can see the contraction of the sphincter of the anal dimple.

The most "benign" of all anorectal malformations is called "perineal fistula." The rectum opens into the perineum, anterior to the sphincter in a rather narrow orifice. This malformation is also known as a "low defect."

A perineal fistula in a male patient may have different external manifestations. A common one is the presence of a malformation called "bucket handle" (Fig. 4.11) which is a prominent skin band under which we can pass a mosquito clamp. Another external manifestation of a perineal

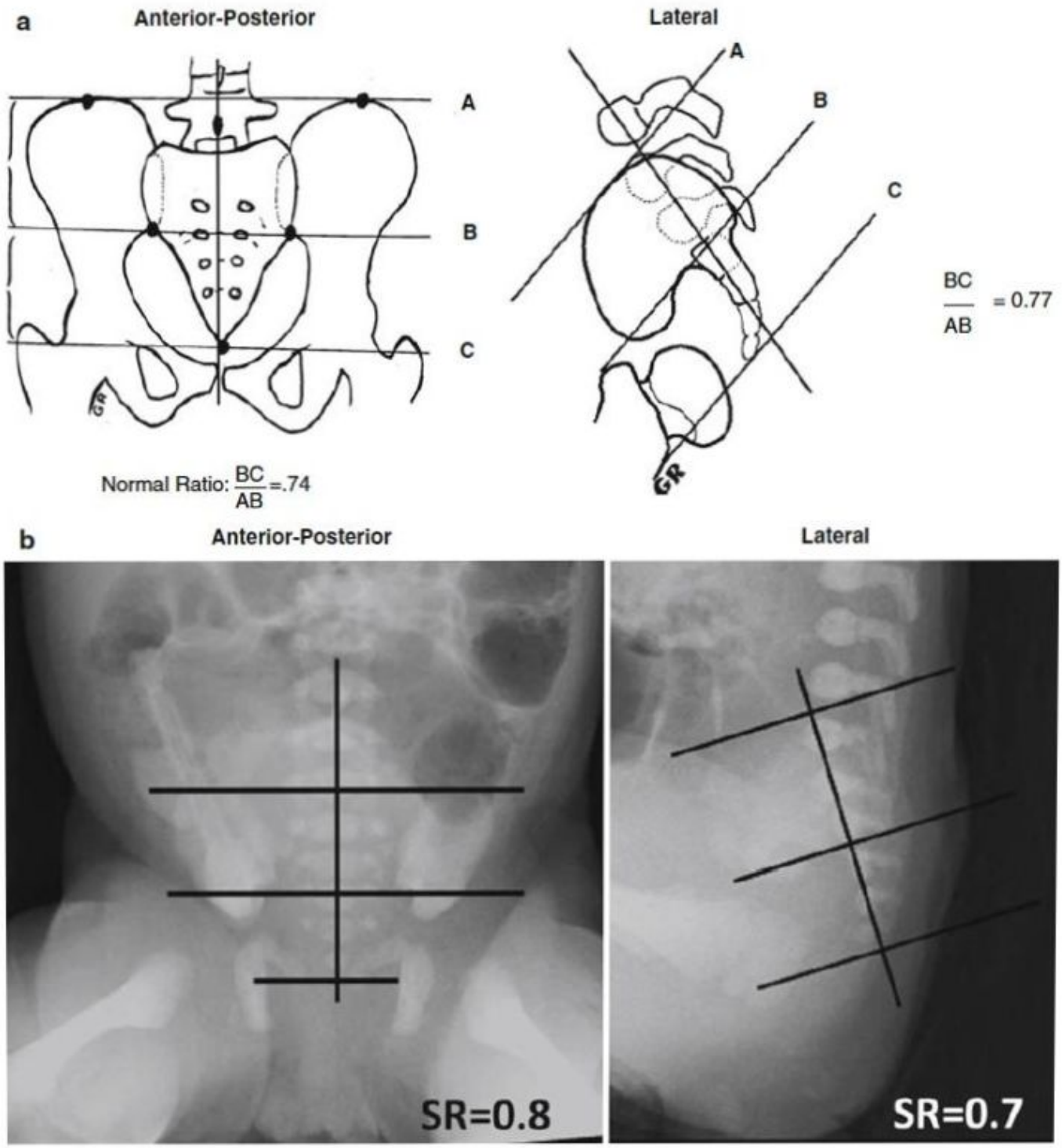

Fig. 4.5 Normal sacral ratio. (a) Diagram. (b) Radiograph

fistula can be a subepithelial fistula (Fig. 4.12). The fistula tract may be full of meconium, giving the appearance of a "black ribbon." Other times, it may be full of white mucous material (Fig. 4.13). The black or white subepithelial tract may extend toward the scrotum in the midline or even to the base of the penis.

A perineal fistula can be repaired with an anoplasty during the neonatal period without a colostomy and has an excellent functional prognosis. The exception would be the group of patients with perineal fistulas associated to an abnormal sacrum and a presacral mass. Ironically, the presence of a presacral mass and hemisacrum seems to be more frequently associated to perineal fistulas than to other anorectal defects.

Occasionally, one may see a patient with a subepithelial fistula or a "bucket handle" malformation; we try to repair the defect only to find that the patient has actually a very long narrow

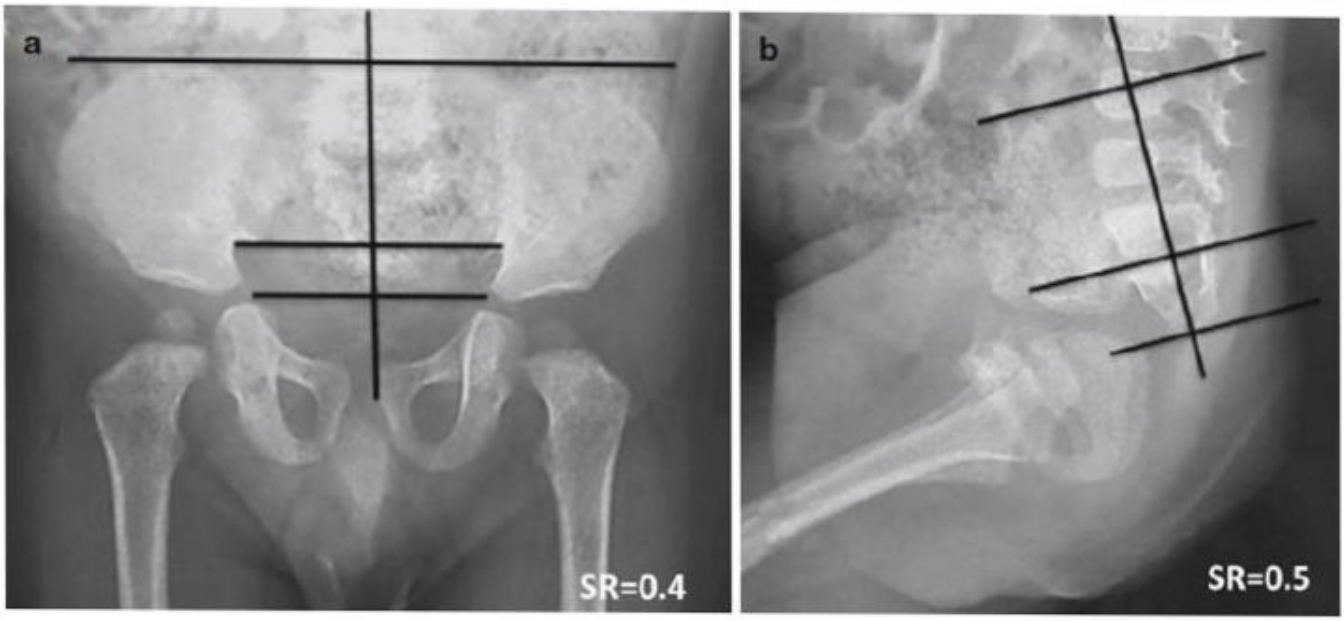

Fig. 4.6 Abnormal sacral ratio. (a) AP view. (b) Lateral view

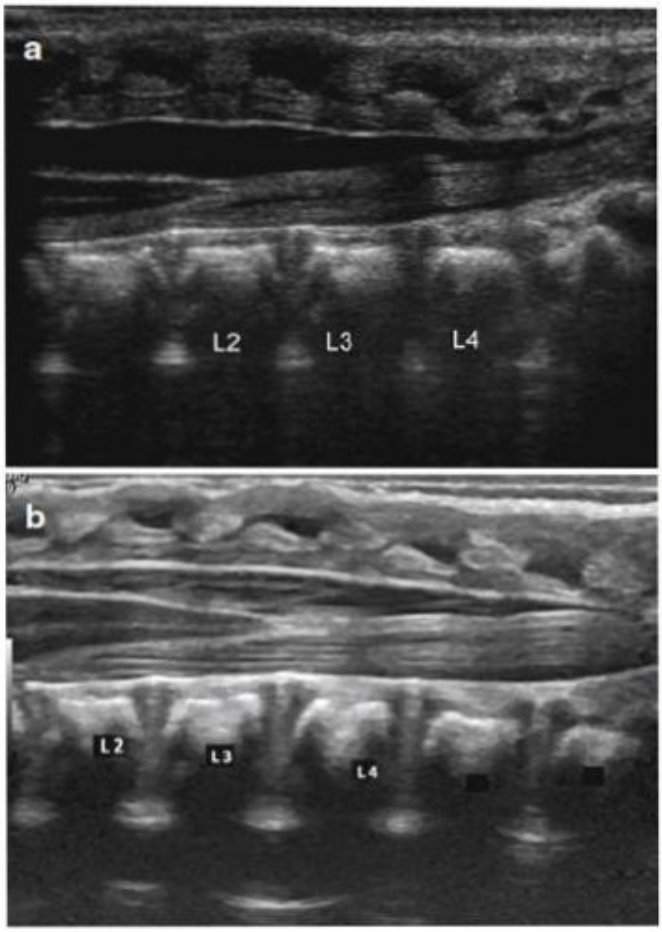

Fig. 4.7 Spinal ultrasound of a newborn baby. (a) Normal. (b) Tethered cord

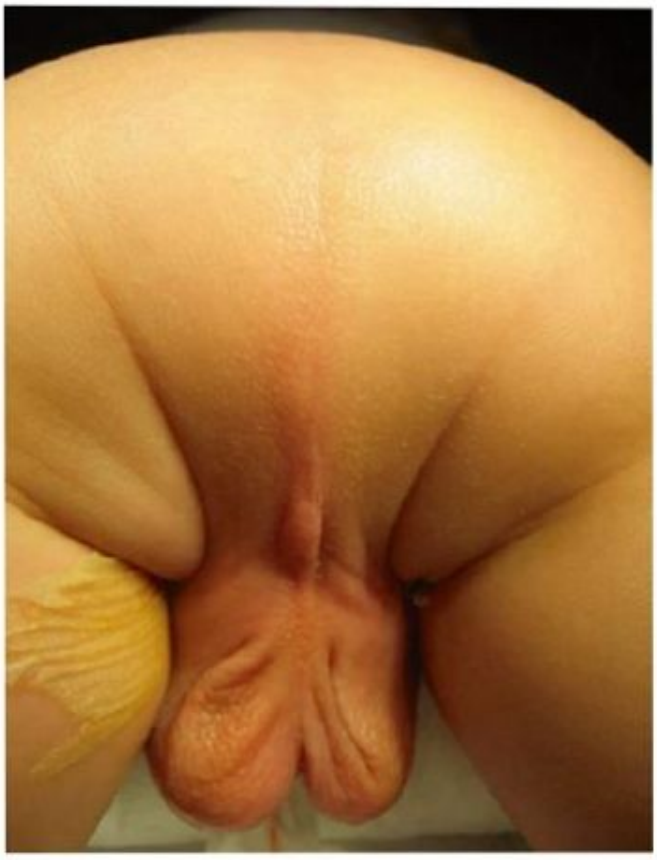

Fig. 4.8 "Flat bottom" in a baby with a recto-bladder neck fistula

fistula and the rectal pouch is located high in the pelvis. That is a real exceptional situation (Fig. 4.14). Under those circumstances, the surgeon has to decide to continue the operation to mobilize the rectum down or to abort the procedure and open a colostomy. That would depend on the degree of experience of the operator.

The presence of bifid scrotum (Fig. 4.15) is usually associated to a rather complex defect; most likely the rectum is located high in the pelvis, connecting to the urinary tract very high (bladder neck or prostatic fistula), although there

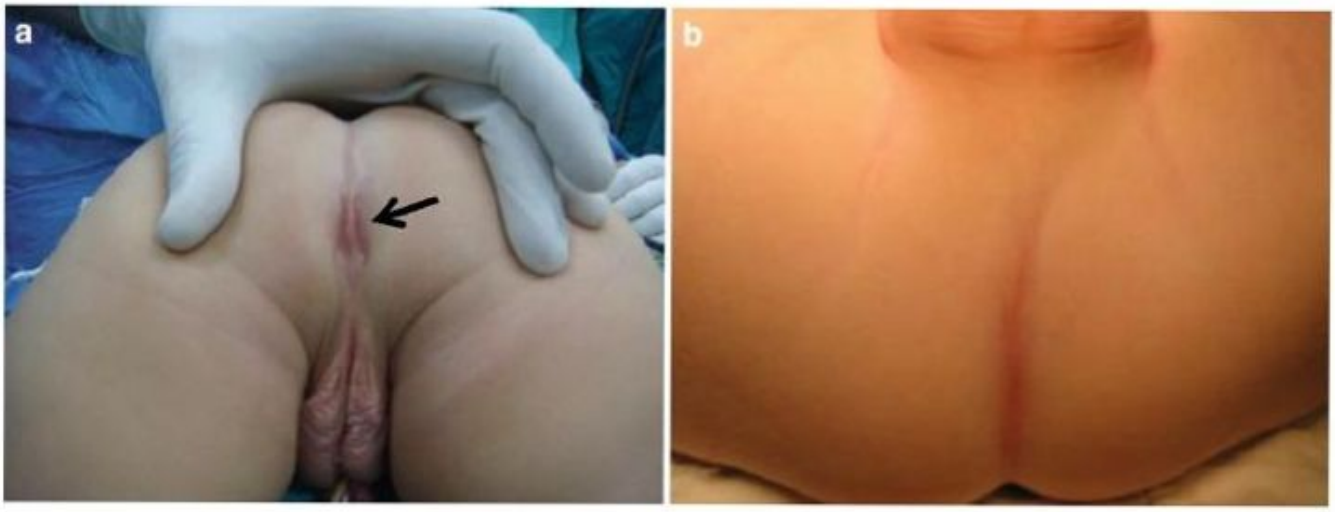

Fig. 4.9 Photograph of the perineum of a newborn baby with (a) good anal dimple. (b) Absent anal dimple. *Arrow* shows the anal dimple

are exceptions. The bifid scrotum is something that we usually repair at the same time that we repair the anorectal malformation (see Chap. 23, Sect. 23.5).

4.5 Female Babies

In female patients, the surgeon must be particularly careful in the examination of the baby's perineum and genitalia. One should put the baby in a convenient lithotomy position with somebody holding the baby's legs. We should have good illumination and magnifying glasses if necessary and take the time to clean the genitalia and to separate the labia to see if there is meconium and precisely where it comes from. Some fistulas are very narrow, and it takes several hours, sometimes up to 24 h, for the babies to pass meconium. The most common defect seen in babies is a malformation called vestibular fistula. As we separate the labia, we can see the urethral opening, the hymen, and the vaginal orifice, and immediately behind that, but still within the introitus of the baby, one can see another little orifice that we call vestibular fistula (Fig. 4.16).

The second most common defect that we see is what we call perineal fistula; the orifice is located somewhere between the normal location of the anus (anal dimple) and the vestibule of the genitalia (Fig. 4.17). Frequently, that orifice, vestibular or perineal, is too narrow to function adequately as an anus. One can identify the anal dimple very clearly located posterior to the perineal or the vestibular fistula.

Although most patients have a well-defined vestibular or perineal fistula, some babies are born with an orifice located right in between, in what the French authors call the "fourchette" of the genitalia (Fig. 4.18).

When the female baby has no anus and the external genitalia look rather small (Fig. 4.19), the surgeon should suspect that the baby has a cloaca. A cloaca is defined as a malformation in which the baby is born with a single perineal orifice. Early diagnosis by inspection of this defect is extremely important. The surgeon must remember that about 90 % of the patients with cloacas may have serious urologic problems. The surgeon must take the time and be meticulous enough to separate the little labia of those small-looking female genitalia and will be able to see a single perineal orifice, and by doing that, he/she already made a diagnosis of a cloaca. Some babies are born with genitalia that induce the doctor to make a diagnosis of intersex or a disorder of sexual development. That is because the patients have a structure that looks like a phallus (Fig. 4.20). They have a single perineal orifice and no testicles, and therefore, the doctors are incapable of saying whether the baby is male or female. In fact, about 60 of our patients with cloaca born in other institutions have been sent to us with a label or misdiagnosis of "intersex." These families were told that the baby had an

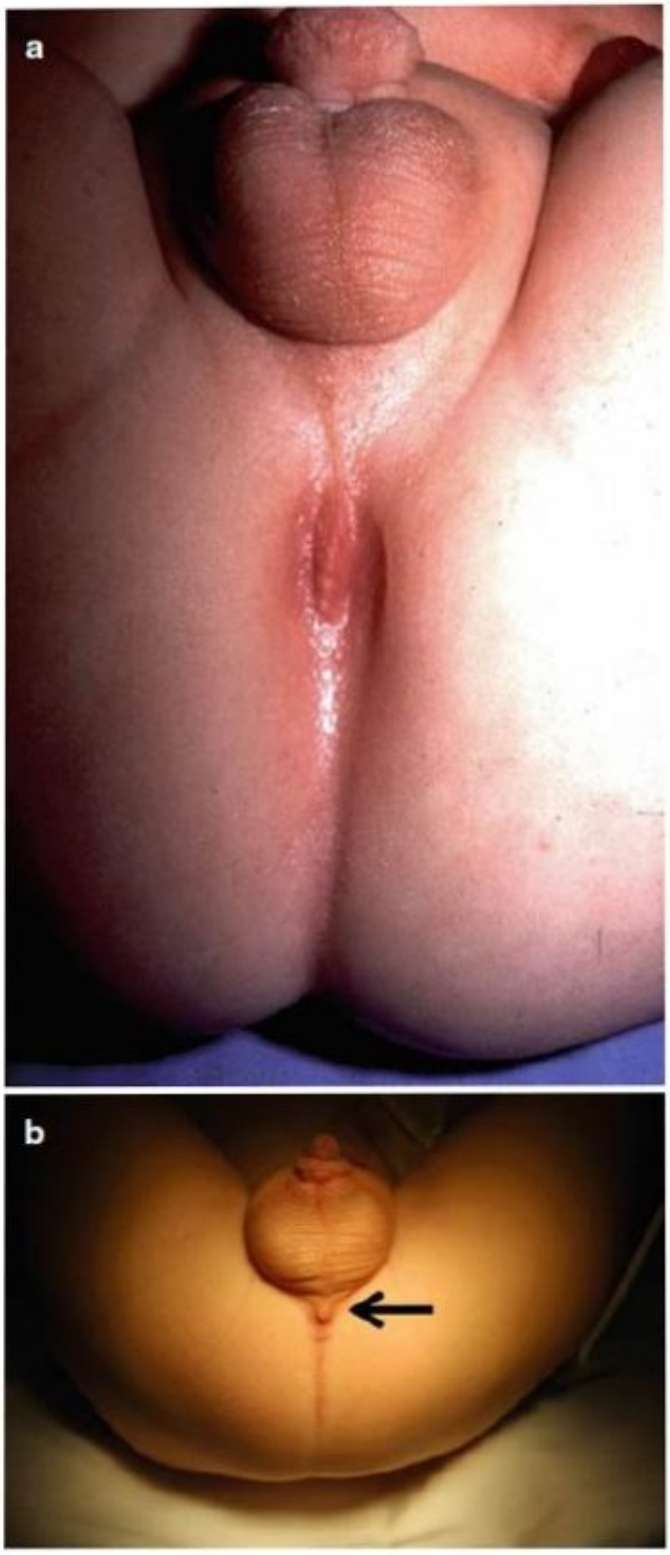

Fig. 4.10 Location of the anal dimple. (a) Normal distance between the scrotum and the anal dimple (malformation with good functional prognosis). (b) Anal dimple next to the scrotum (malformation with less than optimal functional prognosis). *Arrow* shows the anal dimple

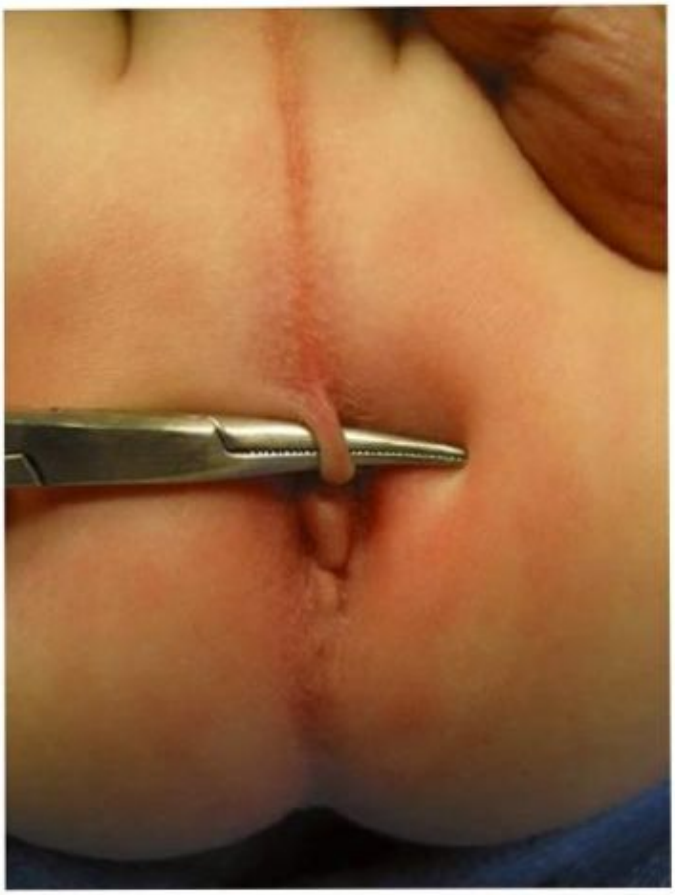

Fig. 4.11 "Buckle handle" malformation in a baby with a perineal fistula

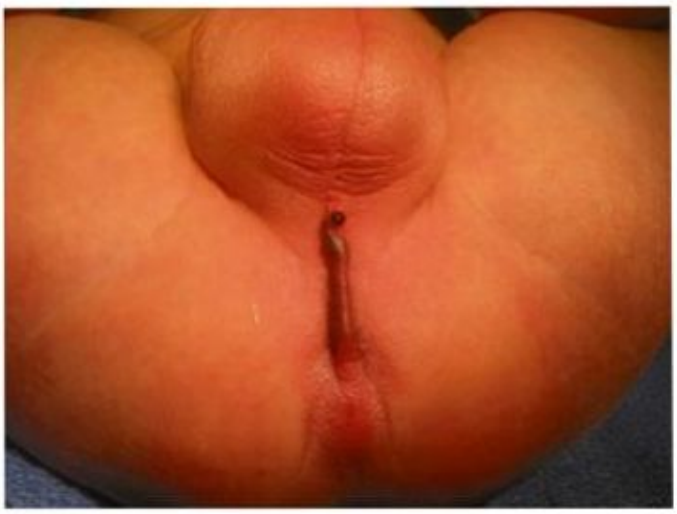

Fig. 4.12 "Black ribbon" appearance of a subepithelial fistula, external manifestation of a perineal fistula

undetermined gender and therefore needs a full evaluation by a geneticist, endocrinologist, and urologist. A series of tests were run, only to find out that the baby actually is XX and is otherwise a normal female, except for the cloaca malformation. In fact, we have never seen a baby with a cloaca with sexual developmental disorder. The key for the diagnosis in this case is to palpate that prominent structure that looks like a phallus. In a real phallic hypertrophy like in cases of adrenal hyperplasia, one can palpate the corpora inside that structure, whereas in babies with

Fig. 4.13 "White ribbon" appearance of a subepithelial fistula, external manifestation of a perineal fistula

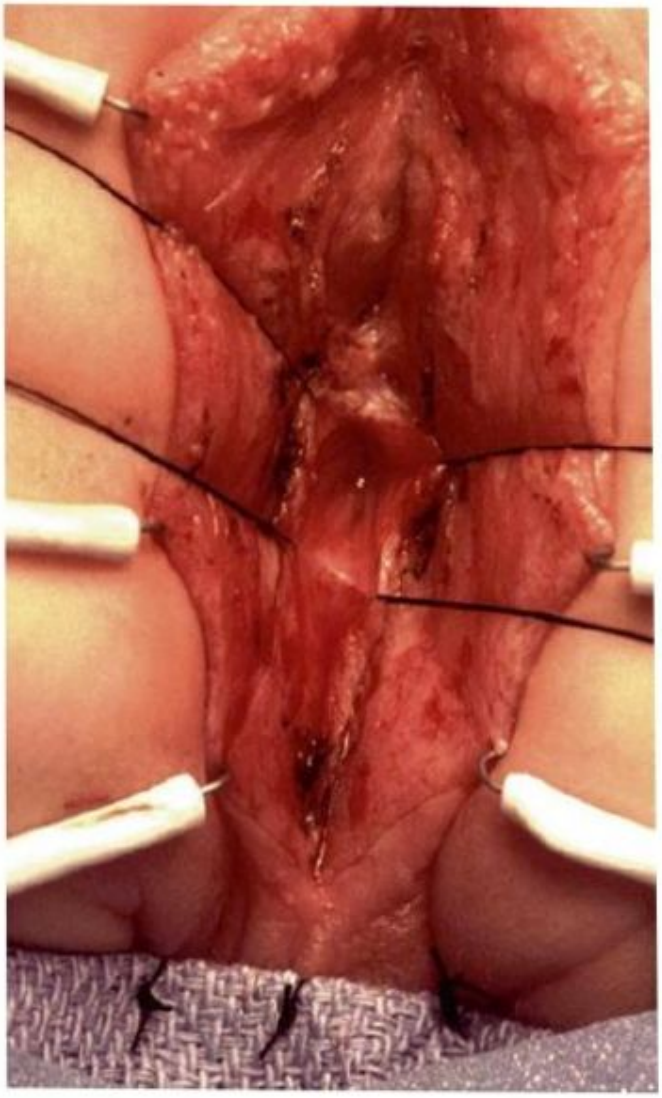

Fig. 4.14 Intraoperative aspect of a long narrow perineal fistula communicating with a very high rectum

cloacas, the palpation reveals that there is only folded prominent skin with no corpora and that makes the diagnosis of a cloaca with no need to rule out an intersex.

A baby with Down syndrome and absent anal orifice has over 90 % chances to have an imperforate anus with no fistula (see Chap. 12).

4.6 Neonatal Management

When we see the baby for the first time, we must make a series of management suggestions to our colleagues, neonatologists, or pediatricians. These include to start the administration of intravenous fluids, to maintain the baby with nothing by mouth, and to introduce a nasogastric tube to avoid vomiting. The nasogastric tube does not interfere with the development of abdominal distention that the baby will have in the following 24 h, but will avoid the risk of vomiting and aspiration. A urinalysis is ordered particularly in male babies, looking for the presence of meconium in the urine. We also prescribe intravenous antibiotics. If it is a female baby and has a fistula (vestibular or perineal), we might pass a little metallic dilator to facilitate the passing of meconium to determine whether or not the fistula is competent to decompress the abdomen and avoid abdominal distention. We must explain to the neonatologist that during the following 20–24 h, the baby should have the diagnostic studies that we already mentioned, including a chest film, an abdominal film, an echocardiogram, an ultrasound of the lumbosacral spine, an ultrasound of the kidneys, and an ultrasound of the pelvis. In babies with cloacas, we emphasize the need to do an ultrasound of the kidneys and also an ultrasound of the pelvis, looking specifically for the

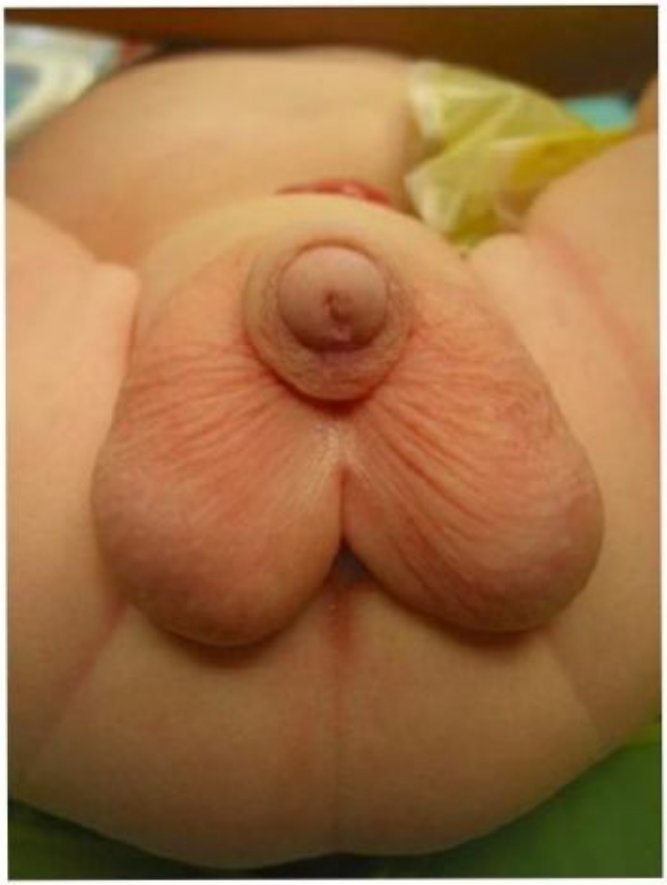

Fig. 4.15 Bifid scrotum, a defect frequently associated to a highly located rectum

presence of a hydrocolpos. We know, from our experience with cloacas, that approximately 60 % of them have an associated hydrocolpos that may be unilateral or bilateral. We also know that the hydrocolpos may compress the trigone of the bladder provoking an extrinsic ureterovesical obstruction with megaureter and hydronephrosis. It is extremely important to make this diagnosis prior to any kind of intervention to be done in the baby. The hydrocolpos must be drained as early as possible, during the first surgical intervention of the baby.

Based on this evaluation and on the experience gained with the long-term follow-up of our patients, we can establish, fairly accurately, the future functional prognosis of the baby and have a long conversation with the parents. We usually tell the parents that when a baby is born with an anorectal malformation, the main concern of the parents as well as the clinicians is to determine what is going to be the quality of life of the baby for the following 80 years. More specifically, is the patient going to have bowel control? Is the

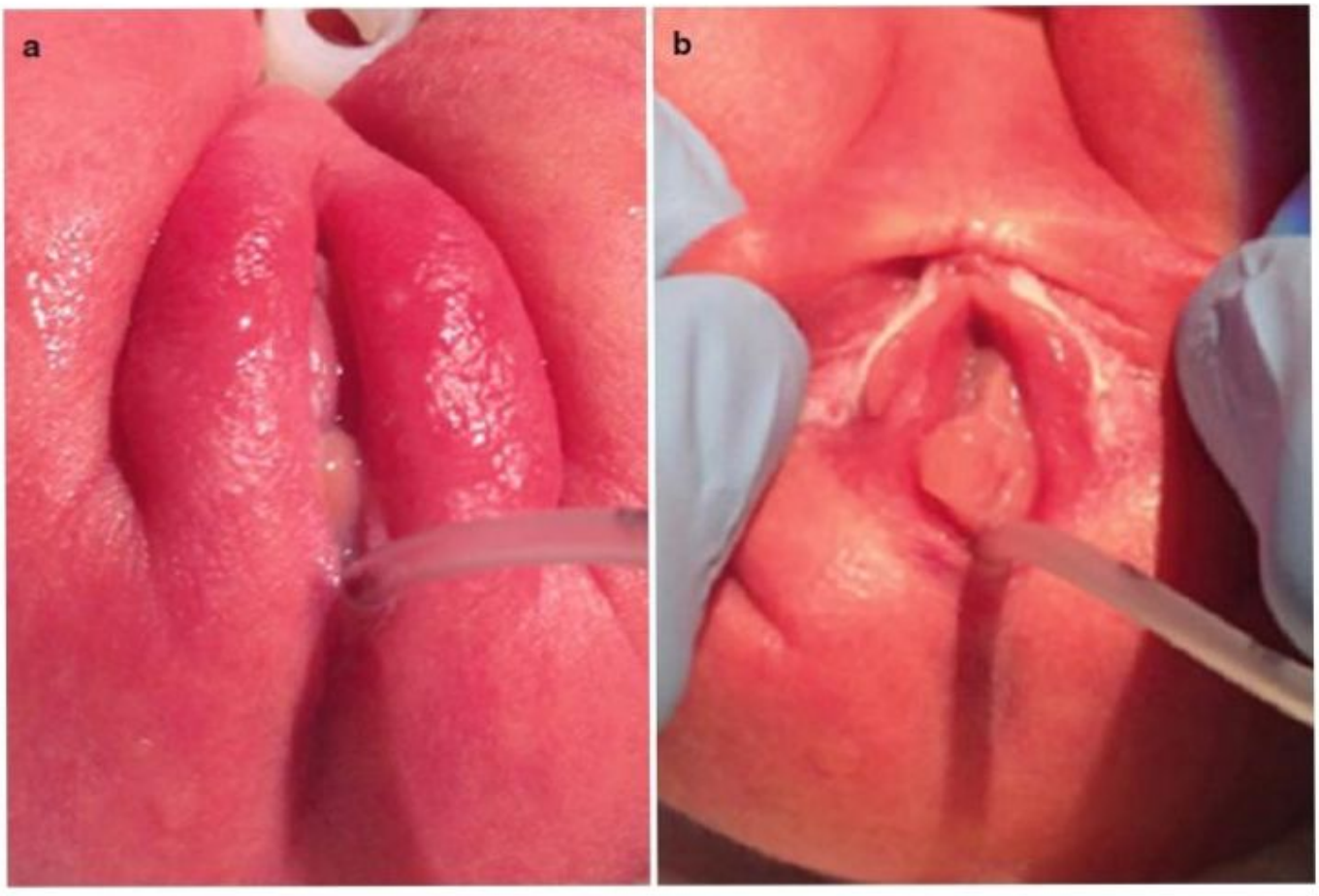

Fig. 4.16 Vestibular fistula in a newborn baby. (a) Without separating the labia. (b) Separating the labia

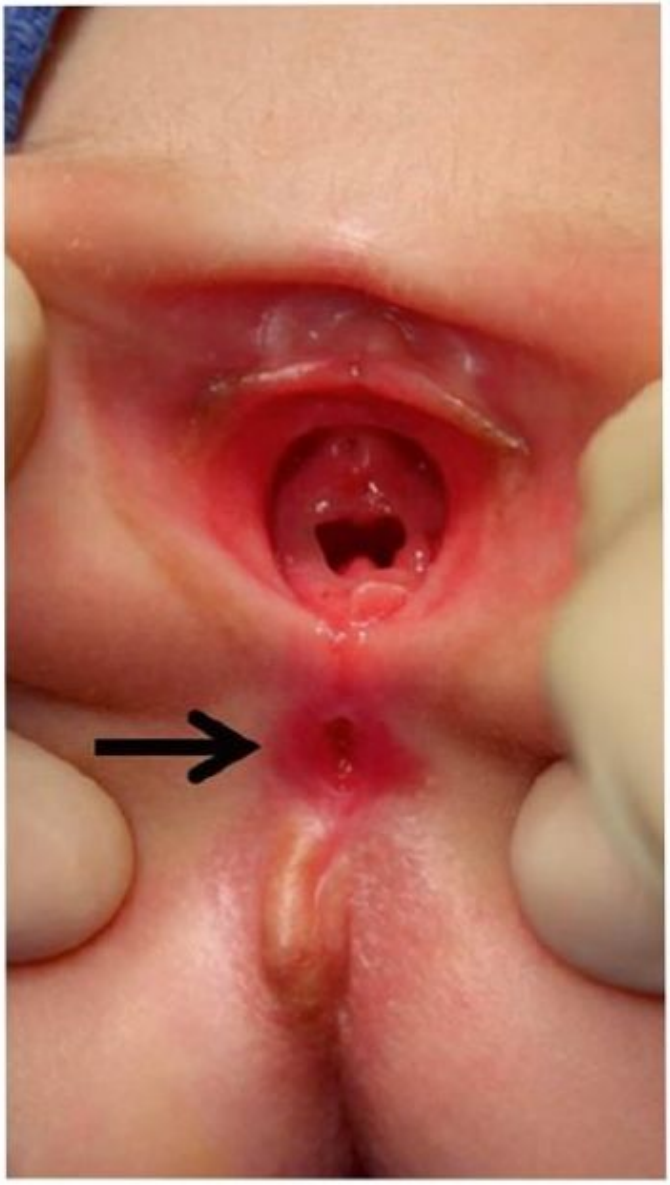

Fig. 4.17 Perineal fistula – *arrow* shows the fistula

baby going to have urinary control? Is the patient going to have sexual function and will be able to reproduce? All these with the specific purpose to avoid what we call the saga of parents of children with anorectal malformations.

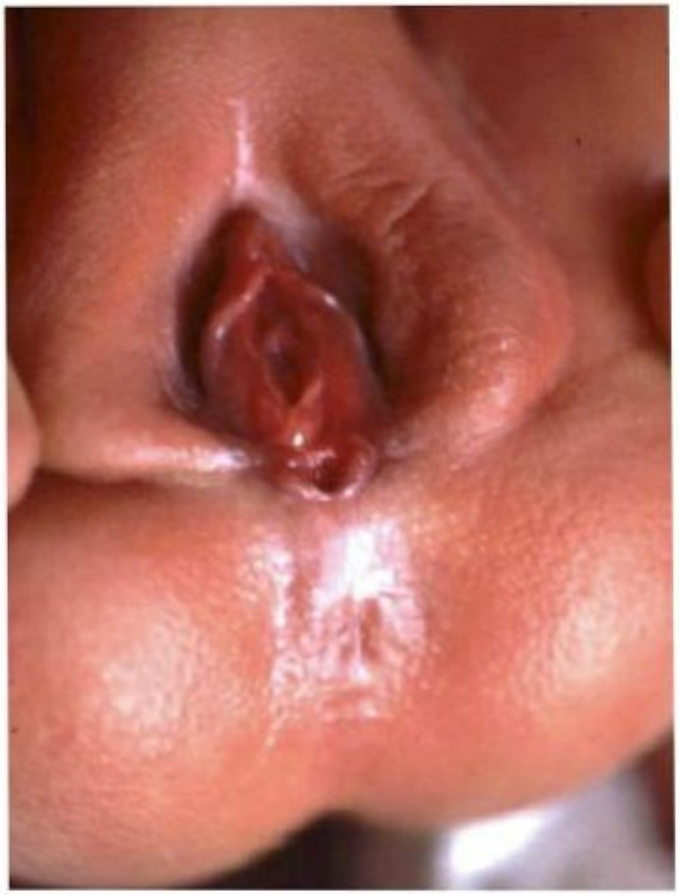

Fig. 4.18 "Fourchette fistula" – fistula located between the vestibule and the perineum

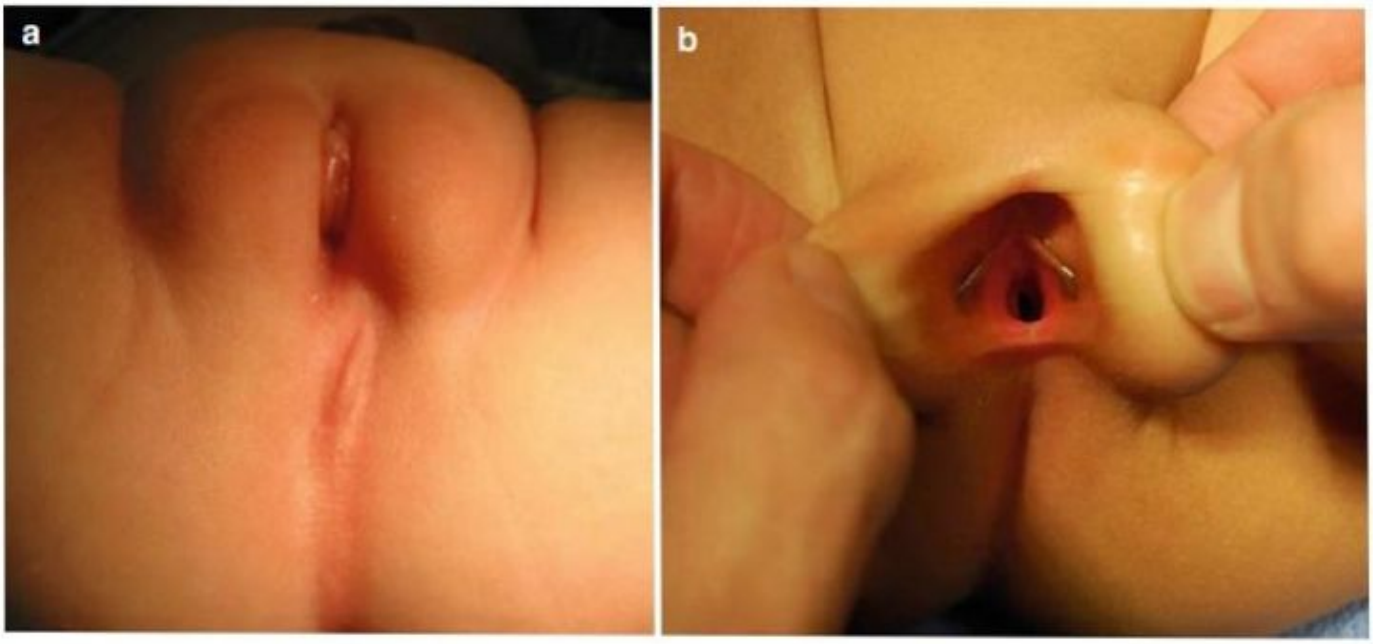

Fig. 4.19 Single perineal orifice. (**a**) External view. (**b**) Separating the labia

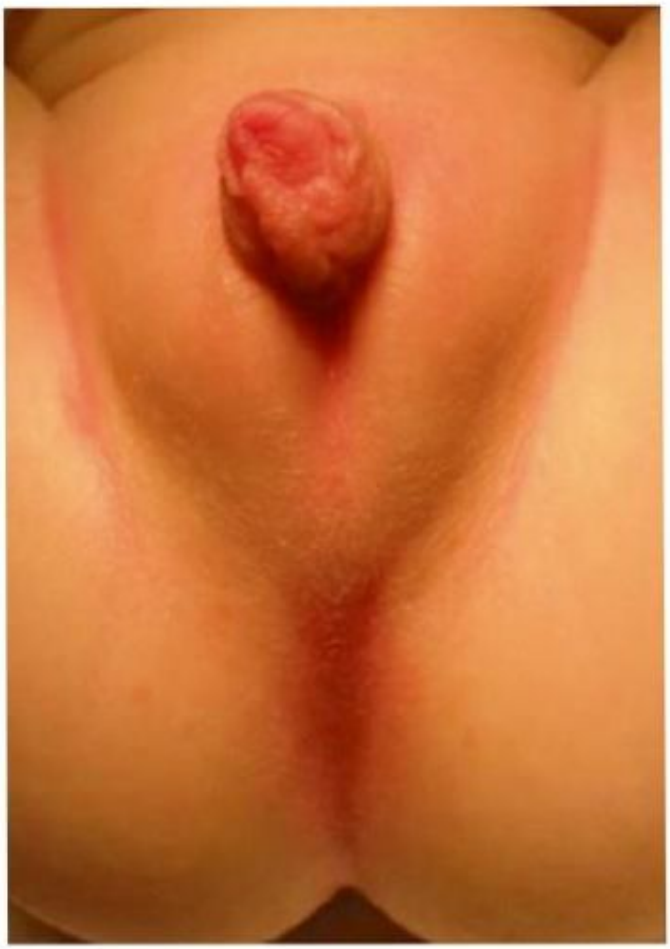

Fig. 4.20 "Phallus-like" prominent skin in a patient with a cloaca. Frequently confused with "intersex"

Many parents of patients born with anorectal malformations describe their unfortunate experience of having a newborn baby with an anorectal malformation. They mention that they were a young happy couple with great expectations and hopes about having a baby. After the delivery of the baby, a doctor appeared in the mother's room and said that the baby had "no anus." Most parents never heard of this malformation because it is not the type of malformation that people like to talk about. As a consequence, most parents never heard of the existence of this malformation. Then the doctors tell the family that the baby is going to have an operation, and subsequently, they come back to say that the operation was "successful." The parents feel happy and take the baby home, but most of the time nobody discusses the future of the baby. At that time, the baby is either on diapers or has a colostomy. Eventually, the baby undergoes the main repair of the malformation followed by a colostomy closure. Since the baby is usually still wearing diapers, the parents do not perceive the difference of their baby's bowel habits when compared with babies without anorectal malformations. However, when the baby reaches the age of bowel control, the parents start making observations, comparing the bowel habits of their baby with other normal children and start worrying. It is then when they start going from doctor to doctor only to find out, sometimes years later, that the baby actually was born with malformation and with a bad functional prognosis and therefore will never have bowel control. We believe that even if this is extremely painful and difficult for us, we are morally obligated to try to establish the functional prognosis as early as possible to adjust the parents' expectations and avoid future painful revelations. As can be seen in Chap. 20, when dealing with families of children born with poor functional prognosis type of defects, we offer them a comprehensive bowel management program to be started at age 3, and we commit ourselves to keep these patients artificially clean in the underwear, in order to be socially accepted, attend school, and avoid psychological sequelae. On the other hand, if the baby was born with a good prognosis type of defect, we have the pleasure to tell the parents the good news.

Once we have all of the information that we already described and we are certain that the baby does not have a serious associated malformation that requires urgent care, we are ready to answer the second question, related to the possibility of opening a colostomy or doing some sort of primary operation to create an anal opening. Most of the time, with the meticulous examination of the perineum, the result of the urinalysis, or the obvious presence of meconium in the urine, as well as the results of the ultrasound and the radiology studies, with a good index of suspicion, we have enough information to make a decision. Occasionally, after 24 h, in spite of all the studies and examinations, we still do not have a clear idea of what we are going to do (primary repair or colostomy). In the old times, it was customary to take an upside-down film [16] in lateral position and with a marker in the anal dimple, with the specific purpose of measuring the distance between the skin of the perineum and the

blind end of the rectum full of gas. At some point, it became obvious that the same image obtained with the upside-down film could be obtained with the baby in a prone position, with the pelvis elevated [17] (Fig. 4.21). In addition, it was also risky to put the baby upside down, for the risk of vomiting and aspiration.

The cross-table lateral film renders a reliable image when it is taken 24 h post birth. When the rectal bubble is located well below the coccyx, the surgeon knows where to expect to find the rectum (Fig. 4.21).

Babies with a flat bottom, poor sacrum, and tethered cord need a colostomy most of the time. Also, in babies that are passing meconium with the urine, we suggest to open a colostomy.

Patients with "bucket handle" malformations, subepithelial fistula, or an obvious perineal fistula opening can be repaired primarily during the newborn period.

There is one particular physiologic event worth discussing, because of its diagnostic and therapeutic implications. At birth, most babies with anorectal malformations do not have a distended bowel and abdomen. It takes 18/24 h for the rectosigmoid to become distended. In addition, the rectum is surrounded by a striated funnel-like sphincter mechanism (see Chap. 2) with a significant tone. The muscle tone keeps the most distal part of the rectum collapsed, until the intraluminal pressure is high enough to overcome the muscle tone. This occurs usually after 24 h; therefore, all imaging diagnostic tests aimed to detect the location of the blind rectum are inaccurate when performed before 24 h of life. This important fact is rarely mentioned. This is the reason why we emphasize to spend the first 24 h of the baby's life in trying to rule out serious associated conditions and try to determine the location of the rectum to decide the surgical approach after a 24 h period.

In order to determine the position of the rectal pouch, different authors suggest using a perineal ultrasound [18–21]. Others are enthusiastic about the perineal injection of contrast material [22–25]. More advance and sophisticated imaging technology has been used, including CT scan [26] and MRI [27–29].

None of the publications related with the optimal imaging diagnostic studies to determine the

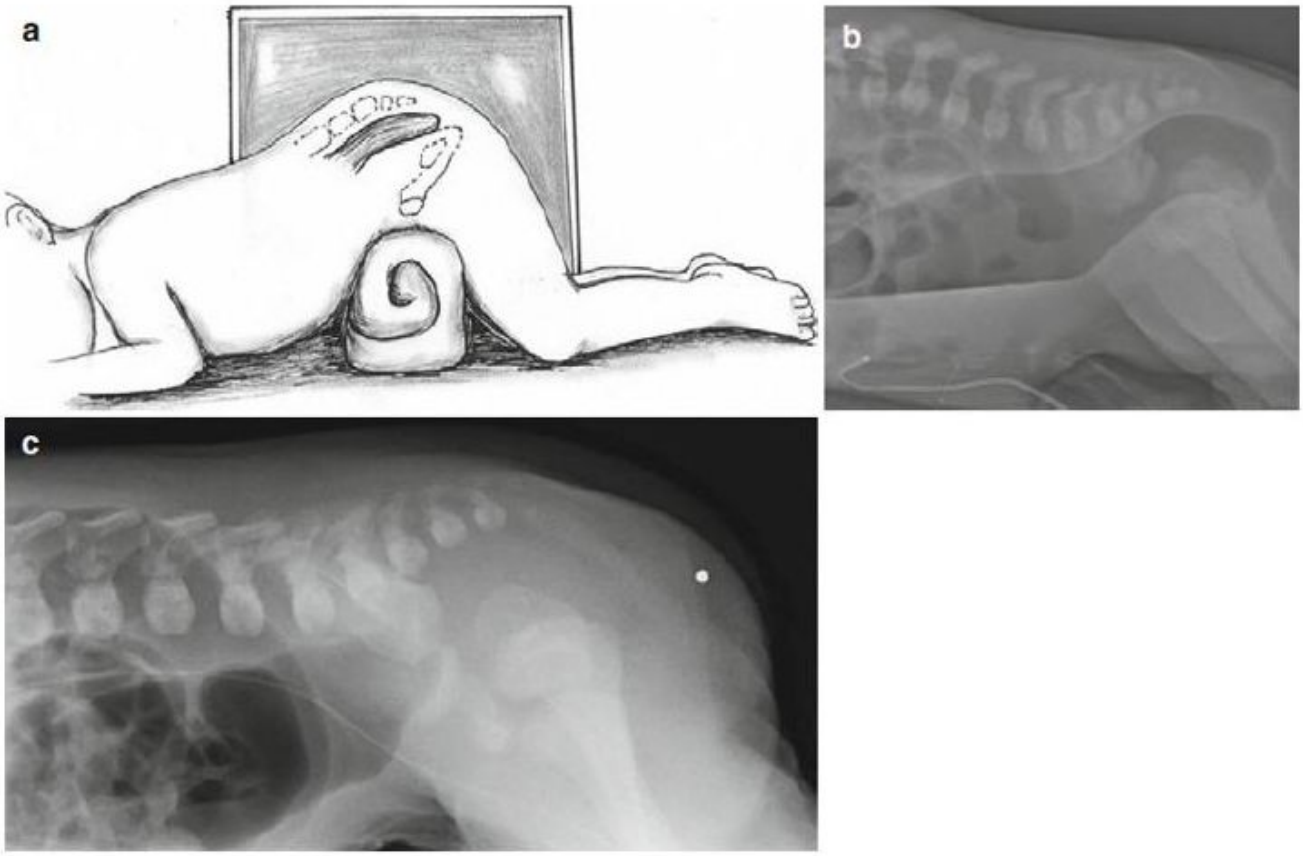

Fig. 4.21 Cross-table lateral film (rarely used study). (a) Baby's position. (b) Image of a reachable rectum. (c) Image of a non-reachable rectum

position of the rectal pouch mentioned the most important key factor which is the timing of those studies. It does not matter how sophisticated the imaging technology employed is, if one does not take into consideration the fact that the distal rectum is surrounded by a striated muscle. Studies done before the rectum becomes distended will fail to make an accurate diagnosis.

In general, all over the world, the pediatric surgical community is moving toward the primarily repair of anorectal malformations, in an effort to avoid the significant morbidity of two important operations: colostomy opening and colostomy closure (see Chap. 5) [30–37]. We agree that we must try to move in that direction; however, we must keep a very critical attitude to be able to balance the desire of notoriety with the benefit of the patient. We should always ask ourselves what we would do if the patient was our son or daughter.

When making these kinds of decisions, the surgeon must take into consideration his specific surrounding circumstances, as well as his experience. One example could be the case of a newborn baby that has a perineal fistula but is extremely sick either because he/she is premature, has respiratory distress, and may have a cardiac condition or other aggravating factors. In such cases, we can simply dilate the fistula. If that is not enough, we can make a cutback procedure on temporary basis, in order to decompress the abdomen and help the baby to recovery.

In general, we consider contraindicated performing abdominal perineal, pull-throughs open or laparoscopic, as well as posterior sagittal anorectoplasties in neonatal babies. As can be seen in the chapter of reoperations (Chap. 22), we have seen multiple cases of patients approached during the neonatal period, without the necessary anatomic information, trying to repair an anorectal malformation; many of those patients suffered very serious damage of important pelvic structures.

In addition, in newborn babies, we cannot do the most valuable radiologic study in the management of anorectal malformation which is called high-pressure distal colostogram (Chap. 6), which shows us the precise location of the most distal part of the rectum, as well as the location of the recto-urinary fistula; all of this represents crucial information that allows us to make a well-defined, precise surgical plan. Primary repairs during the newborn period frequently become authentic misadventures that expose the baby to serious consequences and sequelae.

When the decision is reached to open a colostomy, the patient is taken to the operating room, and the surgeon should follow the principles described in Chap. 5. The anoplasty that we use for the treatment of perineal fistulas is described in Chap. 8.

4.7 Cloacal Exstrophy

There is a specific chapter dedicated to this defect (Chap. 17); here, we will only mention the special neonatal care that these patients require. This is the most serious of all congenital anorectal and urogenital malformations. These babies are born with an omphalocele, an exstrophic bladder with two hemibladders and in the middle of both of them, a portion of intestine protruding in what is called an "elephant trunk" (Fig. 4.22). The pubic bones are widely separated. There is no anus, and

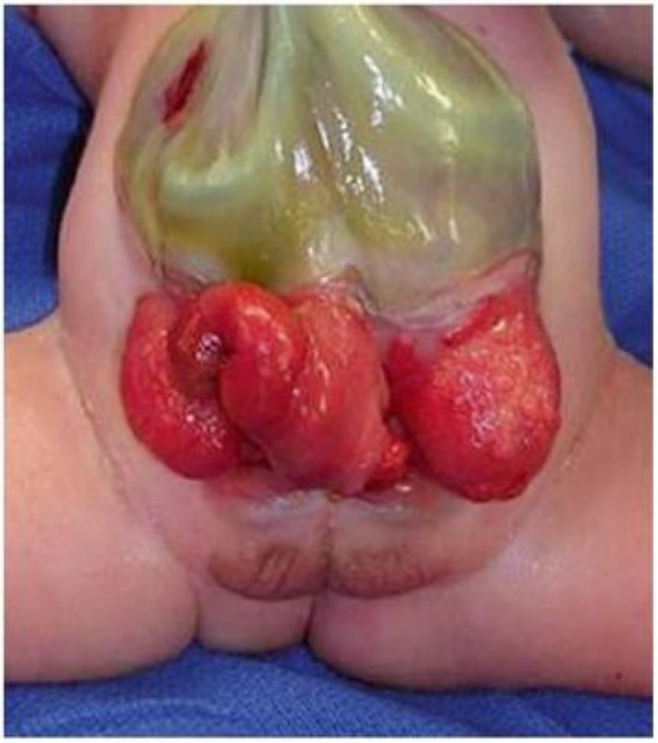

Fig. 4.22 Cloacal exstrophy

meconium may be seen coming out through the "elephant trunk." It is very common to see an associated meningocele and serious spinal defects that may affect the motion of the lower extremities.

The management of these patients requires a multidisciplinary team of experts that include a pediatric surgeon, pediatric urologist, orthopedic surgeon, neurosurgeon, and of course neonatologists. From the time of delivery, nurses and doctors must try to protect the pelvic structures. The mucosa of the bladder and bowel are both exposed and must be covered and handled with care. The omphalocele is a delicate structure that must be handled with special care to avoid its rupture. These structures must be protected with humid and/or lubricated sterile towels, and the baby should be placed in a special care room, putting special interest in preserving the temperature of the body, as well as his/her metabolic concerns. The baby should also receive all the studies that we mentioned to rule out associated important defects that may put the baby's life at risk. We must confirm that the baby is passing meconium and is not becoming more distended. The baby will also receive antibiotics and a nasogastric tube and will be on NPO. Twenty-four hours later (sometimes more), the patient is usually taken to the operating room. The surgical management is described in Chap. 17.

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