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Review of 5 patients with distal loopography

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ABSTRACT

Introduction: Anorectal malformation (ARM) is a wide spectrum of congenital malformations involving the distal rectum and anus as well as the urinary and/or gynaecological systems with an estimated incidence ranging between 1 in 2000 and 1 in 5000 live births. Types of fistulas are essential to diagnose to decide the management and surgical repair. We review several fistula types of anorectal malformation based on distal loopography examination.

Case Presentation: Review of 5 patients with distal loopography in Sanglah Hospital was performed. All patients were neonate with absence of the anus without any clues of fistulas. Plain abdominal x-ray and knee-chest position showed distended loops of bowel and blind-ending of distal rectum with various distance from anal dimple. Two cases were confirmed as high type of ARM with recto-vesica fistula, where the contrast filled rectum and the bladder which decreased significantly after micturition. The two other cases were confirmed as high type ARM with recto-urethrae fistula, and the last one as low type of ARM with recto-perineal fistula. All patients undergoing surgical management for fistula repair and anorectal reconstruction.

Discussion: Anorectal malformation forms a heterogeneous group of anomalies, most cases showed imperforated anus and blind-ending of distal enteric component (atresia) with or without fistula. Our cases were diagnosed by plain x-ray and contrast study. High or low type was evaluated from the distance rectal pouch to anal dimple. Distal loopogram was showed low type of ARM with recto-perineal fistula and high type of ARM with fistula extended anteriorly as recto-vesica fistula and recto-urethrae fistula, as The Krickenbeck classifications.

Conclusion: Distal loopography is an important diagnostic investigation to delineate the altered anatomy of anorectal malformations, the location fistula and the spectrum of associated fistula between the blind rectum and the bladder, urethra, perineum, scrotal and vagina. Radiology interpretation is indeed important for clinician to establish patient management.

Keywords: anorectal malformation, fistula, distal loopography.


INTRODUCTION

Anorectal malformations (ARMs) are congenital anomalies encountered in pediatric surgery, with an estimated incidence ranging between 1 in 2000 and 1 in 5000 live births. Anorectal malformations, also known as imperforate anus, are defects that occur during the fifth to seventh weeks of fetal development. With these defects, the anus (opening at the end of the large intestine through which stool passes) and the rectum (area of the large intestine just above the anus) do not develop properly.1,2

Anorectal malformations comprise a wide spectrum of diseases, which can affect boys and girls, and involve the distal anus and rectum as well as the urinary and genital tracts. Most cases are diagnosed in the early neonatal period. Defects range from the very minor and having simple management with an excellent functional prognosis, to those that are complex, difficult to manage, are often associated with other anomalies, and have a poor functional prognosis. Several abnormalities can occur, including the anal passage may be narrow, a membrane may be present over the anal opening, the rectum may not connect to the anus, the rectum may connect to part of the urinary tract or the reproductive system through a passage called a fistula.3,4

The etiology of such malformations remains unclear and is likely multifactorial. There appears to be a low rate of association in families, but some appear to have an autosomal dominant inheritance pattern with a high incidence, as much as 1 in 100.5,6

Approximately, ARMs are associated with other congenital anomalies in up to 70% of cases (2–5) (Table 1). The incidence of reported anomalies is variable, but most groups agree Urogenital abnormalities are the most frequently observed and appear in up to 60% of patients, with vesicoureteral reflux and hydronephrosis the most common findings, followed by cardiovascular (30–35%), spinal cord tethering (25–30%), gastrointestinal anomalies (5–10%), and VACTERL (4–9%) anomalies. The higher the rectal pouch ends, the higher are its chances to be associated with anomalies.1,3

At birth, a general examination in any newborn should include the perineum. The absence or abnormal location of the anus is generally apparent. In the male, besides the absent anus, a note...
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must be made of the anal pit. Clinical inspection of the buttocks is essential. A flat “bottom” or flat perineum, as evidenced by the lack of a midline gluteal fold and the absence of an anal dimple indicates that the patient has very poor muscles in the perineum. These findings are associated with a high malformation and therefore a colostomy should be performed.4,8

Perineal signs found in patients with low malformations include the presence of meconium at the perineum, a “bucket-handle” malformation (a prominent skin tag located at the anal dimple below which an instrument can be passed), and an anal membrane (through which one can see meconium). The fistula may be of small calibre; hence, it may take up to 24 hours for it to be apparent. The reason for this is that it takes this long for ingested gas to travel down the gut to the blind rectum. In the female child, the diagnosis rests on the appearance of the perineum.2,6

The classification of ARMs is mainly based on the position of the rectal pouch relative to the puborectal sling and the presence or absence of fistulas. Imaging studies play a key role in the initial evaluation of ARMs. In neonates, an accurate diagnosis of the type of ARM, the presence or absence of fistulas and their locations, and identification of associated anomalies are essential in deciding about immediate therapy. However, the real utility and the most appropriate sequence of performance of the different imaging diagnostic methods in the first days of life remain very important.5,8

The Krickenbeck classification distinguishes the types of fistulas: rectoperineal, rectovestibular, recto–urethral bulbar, recto–urethral prostatic, and rectovesical. Cloacal malformations, the absence of fistulas, anal stenosis, and rare regional variants complete this classification. The extremely rare rectovaginal fistula is considered a variant of cloacal anomaly. The Wingspread and Krickenbeck classifications for males and females are compared in Tables 1 and 2, respectively. The different types of stulas in males and in females are shown in Figures 5 and 6, respectively4

The radiologic evaluation of a newborn with imperforate anus includes an abdominal ultrasound to evaluate for urologic anomalies. In the case of persistent cloaca, a distended vagina (hydrocolpos) can be identified. Plain radiographs of the spine can show spinal anomalies such as spina bifida and spinal hemivertebrae. Plain radiographs of the sacrum in the anterior-posterior and lateral projections can demonstrate sacral defects such as a hemisacrum and sacral hemivertebrae.9,10

Plain abdominal radiograph can be variable depending on the site of atresia (i.e. high or low), level of meconium impaction and physiological effects such as straining may show multiple

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<th>Table 1 Comparison of Wingspread and Krickenbeck Classifications of ARM in male patients</th>
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<th>Table 2 Comparison of Wingspread and Krickenbeck classifications of ARMs in female patients</th>
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Figure 1 The prone lateral view is performed to demonstrate the position of the most distal portion of the large bowel and therefore determine whether it is a high or low.
dilated bowel loops with an absence of rectal gas. A cross-table lateral view or knee-chest position may help determine if it’s high or low in the anorectal area. Prone cross-table lateral radiograph of a pelvis demonstrating the pubococcygeal line. The fistula is considered low (below the levator ani plane) if it is below the pubococcygeal line (PCL) and high if above it.\textsuperscript{2,10}

Distal colostography, also called distal cologrophy or loopography, is an essential step in the reparative management of anorectal malformations (ARMs) with imperforate anus. It serves to identify/confirm the type of ARM, presence/absence of fistulae, leakage from anastomoses, or patency of the distal colon. High-pressure distal colostography is the most useful imaging technique for demonstrating fistulas. It consists of manual injection of water-soluble contrast material through a Foley catheter inserted into the distal colostomy, which is sealed by means of gentle traction of the inserted balloon. To increase the possibility of detecting a rectourinary fistula, the injection of contrast material should be continued until the patient voids the bladder. In most cases, the technique also provides accurate information about the position of the rectal pouch. However, sometimes the pressure required to demonstrate the fistula causes a slight depression of the distal rectum; this depression may reduce the real distance between the rectal pouch and the anus.\textsuperscript{1,3}

Types of fistulas are very important to diagnose to decide the management and surgical repair. We review several fistula types of anorectal malformation based on distal loopography examination.

**CASE PRESENTATION**

Review of 5 patients with distal loopography in Sanglah Hospital was performed. All patients were neonate with absence of the anus without any clues of fistulas. Plain abdominal x-ray showed distended loops of bowel with a lack of rectal gas and blind-ending of distal rectum with various distance from anal dimple.

Prone cross-table lateral radiograph or knee-chest position is to try to determine if the lesion is ‘low’ and can therefore have a primary definitive procedure without a covering stoma. The position of the anus should be marked with a radio-opaque material, and the distance from the skin to the rectal gas measured (Figure 1).

Two cases were confirmed as high type of ARM with recto-vesica fistula. The investigation involves the injection of water-soluble contrast into distal limb of colostomy using balloon catheter. Watersoluble injected under constant gentle pressure until a fistula is filled, and visualised with the aid of fluoroscopy (Figure 2 A-D). A radio-opaque marker is placed over the anal dimple or expected position of the anus and true lateral images are obtained. Lateral or oblique position and evaluated after micturition are needed to be done to confirm...
the fistula. The loopogram of our cases was showed contrast filled rectum and the bladder which decreased significantly after micturition. The first cases were showed tract fistule from rectum to posterior urethra continuously to posterior an anterior urethra and external meatus urethra, contrast also back flow to filled the bladder. The other case was showed continuation contrast from rectum to urethra posterior which also makes another fistula tract to scrotum (Figure 3 A-C). The obstruction of anterior urethra could be the cause of the fistula tract apparent to scrotum. The two other cases were confirmed as high type ARM with recto-urethrae fistula. The last one case was confirmed as low type of ARM with recto-perineal fistula. Distal loopogram was showed fistula tract from rectum to perineum (Figure 4 A & B). All patients are undergoing surgical management for fistula repair and anorectal reconstruction.

**DISCUSSION**

Anorectal malformation forms a complex group of anomalies, most cases showed imperforated anus and blind-ending of distal enteric component (atresia) with or without fistula. Our cases were diagnosed by plain x-ray and contrast study. High or low type were evaluated from the distance rectal pouch to anal dimple. Distal loopogram was showed low type of ARM with recto-perineal fistula and high type of ARM with fistula extended anteriorly as recto-vesica fistula and recto-urethrae fistula, as The Wingspread and Krickenbeck classifications.\(^1\)

The Krickenbeck classification is descriptive and emphasises the preoperative identification and anatomic evaluation of the rectal pouch and fistula. This information is vital for the surgeon as it allows anticipation of the extent of mobilisation in relation to the urinary tract as well as the most appropriate surgical approach for each case.

Children who have an anorectal malformation will need a surgery called an anorectoplasty (PSARP) to correct the defect. This involves moving the anus to the appropriate location within the muscles (anal sphincter) that are responsible for bowel control. When possible, the surgeon can use minimally invasive laparoscopic or robot-assisted surgical techniques. This can mean less pain, less blood loss, faster healing and shorter hospital stays, compared to what patients may experience with an “open” or traditional surgery. Male newborns with recto-perineal fistula do not need a colostomy. They can undergo a posterior sagittal anoplasty whereas male babies with evidence of a recto-urinary tract communication should undergo faecal diversion with a colostomy.\(^2,8\)

A definitive repair in the newborn period avoids a colostomy but there is considerable risk to the urinary tract with this practice because the surgeon does not know the precise anorectal defect. The only way to definitively determine the patient’s anorectal defect is to perform a distal colostogram, which of course requires the presence of a colostomy. Distal colostogram is the most accurate way of determining the anatomy of the distal rectal pouch and fistula in boys. The investigation is ideally done at 2–3 months of age in ARM patients who have undergone a diverting colostomy; it should be done just prior to the definitive repair. Without this information an operation in the newborn period is essentially a blind perineal exploration. The surgeon may not be able to find the rectum and may find and damage other, unexpected, structures, such as the posterior urethra, seminal vesicles, vas deferens, and ectopic ureters during the search for the...
rectum. Finally, without fecal diversion, there is the risk of dehiscence and infection. These complications may compromise the ultimate functional prognosis.\textsuperscript{2,3}

The prognosis and quality of life for children with ARMs will depend, to a large extent, on the presence and gravity of these associated anomalies.

**CONCLUSION**

ARMs are a complex group of congenital anomalies involving the distal anus and rectum. They result from abnormal development of the urorectal septum in prenatal life. The Krickenbeck Conference of 2005 established a new classification of ARMs based not only on the level of the rectal pouch but also the presence or absence of fistulas and their description, factors helpful in determining the most appropriate surgical approach.

Imaging plays a key role in evaluation of ARMs. In the first days of life, clinical and imaging findings facilitate early classification of ARMs and allow a decision about whether to perform an immediate colostomy. This information helps orient the medical and surgical teams as to the postoperative prognosis for continence.

Distal loopography is an important diagnostic investigation to delineate the altered anatomy of anorectal malformations, the location fistula and the spectrum of associated fistula between the blind rectum and the bladder, urethra, perineum, scrotal and vagina. Radiology interpretation is indeed important for clinician to establish patient management.

**CONFLICT OF INTEREST**

The author declares there is no conflict of interest regarding publication of current case report

**ETHICAL STATEMENT**

All of the patients have received signed informed consent regarding their respective radiology data to be presented in journal article.

**REFERENCES**


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