Anorectal Anomaly in two Cases

*Chowdhury Mokbul-E-Khoda,[1] M A Rouf,[2] Nayeem Ullah,[3]
Liaquat Ali,[4] Musfiq Anjum[5]

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ABSTRACT

Anorectal malformations (ARMs) are the most common surgically treated congenital anomalies in newborns. Early diagnosis and timely surgical intervention affords the best opportunity for correction without complications. We report two cases of ano-rectal malformation. First baby was six days old male while second baby was three days old male child admitted in the department of paediatrics, North Bengal Medical College Hospital, Sirajganj with the complaints of not passing meconium since birth with gradual distention of abdomen and absence of anal opening in the perineum. Mothers also complained of vomiting for several times for last two days. For confirmation of the diagnosis, we performed invertogram for both babies. We observed that first baby has anorectal anomaly (imperforate anus -low variety) while the second baby has high variety anorectal anomaly. First baby was surgically corrected by cutaneous anoplasty, while the second baby was initially corrected by colostomy.

1. *Associate Professor, Department of Paediatrics, North Bengal Medical College, Sirajganj*
2. *Assistant Professor, Department of Surgery, North Bengal Medical College, Sirajganj*
3. *Assistant Professor, Department of Radiology and Imaging, North Bengal Medical College, Sirajganj*
4. *Assistant Professor, Department of Paediatrics, North Bengal Medical College, Sirajganj*
5. *Assistant Registrar, Department of Paediatrics, North Bengal Medical College, Sirajganj*

***Corresponding author:** ✉ abdurroufdr@gmail.com.*

INTRODUCTION

Imperforate anus' has been a well-known condition since antiquity.[1] Now-a-days, broadly termed as anorectal malformations (ARMs). No specific cause has been described. The average incidence worldwide is 1 in 5000 live births.[2] When compared to older children, neonates have a wide divergence in physiology, anatomy, immunity and response to stress and when they have surgical illness they are further compromised by the condition itself which predisposes anaesthesia and surgery.[3-5] Anorectal malformations are the major surgical emergencies in neonatal intestinal obstruction (NIO). Defects range from the very minor easily treated 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. Many neonates with

imperforate anus have an abnormal sacral progression, as well as a fistula between the rectum and the genitourinary tract, demonstrated by finding meconium in the urine. NIO presented with a triad of bile stained vomiting, abdominal distension and failure to pass meconium.[6] Failure to pass meconium (48% of normal neonates pass meconium in the first 24 hours of life) is symptomatic of lower intestinal obstruction. Normally it takes between 3 and 6 hours for the gastric contents to reach the small bowel. The intestinal contents reach the Caecum in a liquid state. It then takes about 20 to 24 hours for that faecal material to reach the rectum and become formed stool with the absorption of water that occurs in the colon. The rectum and sigmoid colon acts as a reservoir and holds the faecal material for a variable period of time. First the clinician must perform a thorough perineal inspection, which usually provides the most important clues about the type of malformation that the patient has. It is important to not make a decision about a colostomy or a primary operation before 20 to 24 hours of age. Radiologic evaluations do not show the real anatomy before 24 hours because the rectum is collapsed by the muscle tone of the sphincters that surround its lower part. Therefore radiologic evaluations done too early (before 24 hours) will likely reveal a "very high rectum" and thereby yielding a false diagnosis. After 24 hours, if there is no meconium on the perineum, we recommend obtaining a crosstable lateral radiograph with the patient in the prone position. Historically, an invertogram was used to identify whether the anomaly is high or low. When a low anomaly (perineal fistula) is diagnosed, the fistulous track to the perineum is always located anterior to the sphincter mechanism. A limited posterior sagittal anorectoplasty can be performed in the newborn period. If air in the rectum is seen distal to the coccyx and the patient is in good condition with no significant associated defects, one may consider performing a posterior sagittal operation without a protective colostomy. A more conservative alternative would be to perform the posterior sagittal repair and a protective colostomy at the same stage. Conversely, if the rectal gas does not extend beyond the coccyx, or the patient has meconium in the urine, an abnormal sacrum, or a flat bottom, we recommend a colostomy. With emerging advancements in perinatology and prenatal ultrasound techniques, anorectal malformations are more commonly being suspected.[7] Sonographic findings such as a dilated rectum or hydrocolpos or demonstration of an associated anomaly such as an absent kidney, a vertebral anomaly such as a hemisacrum, or an orthopaedic problem such as a missing radius can make the perinatologist suspicious that the neonate may in fact have an anorectal malformation. With improving technology, it is likely that this area of diagnosis and even neonate intervention for, perhaps, a massive hydrocolpos will continue to advance.[8,9] We reported two cases of anorectal malformations.

The Cases

We reported two patients having ano-rectal malformation. First one was six days old male baby (Figure 1) while second patient was 3 days old male term baby, admitted in the department of paediatrics in North Bengal Medical College Hospital, Sirajganj with the complaints of not passing meconium since birth with gradual distension of abdomen. Mothers also complained of vomiting for several times for last two days. On clinical examination, we found the first baby was mildly icteric, activity and tone was normal, respiratory rate was 36 breaths/minute, heart rate was 120 beats/minute, Temperature-99^0 F,

mildly anaemic, abdomen was hugely distended, hard in consistency, bowel sound was absent, ascites was also absent, there was no organomegaly. Genitourinary system was normal, there was no fistulous communication with urethra but a small opening present in the perineum. Other systems examination revealed normal findings.

Figure 1: Clinical picture of patient 1 showing perineal fistula with well-developed buttock (Arrow).

During clinical examination of second case, we found the baby was mildly pale, anicteric, vital signs were normal, abdomen was hugely distended and hard in consistency, bowel sound was absent, ascites was also absent, there was no organomegaly, other systems examination was normal. For confirmation of the diagnosis we performed invertogram of both babies. By the invertogram, we observed that first baby has imperforate anus (low variety) as shown in Figure 2 and 3, but second baby has high variety anorectal anomaly (Figure 4 & 5). Both children were treated by surgical correction. First baby was surgically corrected by cutaneous anoplasty while second baby was initially corrected by colostomy.

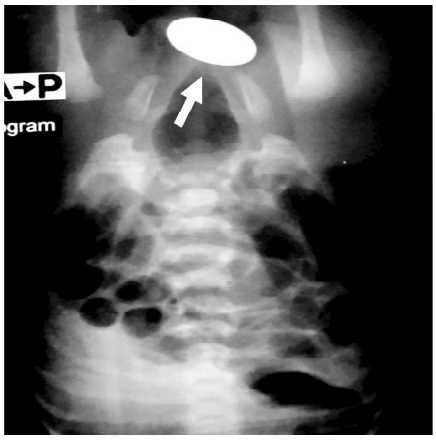

Figure 2: The invertogram of patient 1 showing low variety (AP view) (Arrow).

Figure 3: Invertogram of patient 1 showing low variety (lateral view) (Arrow).

Figure 4: Invertogram of patient 2 showing high variety (AP view) (Arrow).

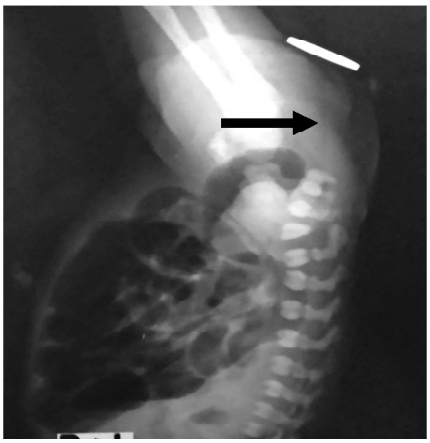

Figure 5: Invertogram of patient 2 showing high variety (lateral view) (Arrow).

DISCUSSION

Anorectal malformations represent a wide spectrum of defects. The terms *'low,' 'intermediate,'* and *'high'* are arbitrary and not useful in current therapeutic or prognostic terminology. A therapeutic and prognostically oriented classification is depicted.[10] In the case of boys, the perineal body, the median raphe, scrotum, and penis should be carefully inspected.[11] Meconium being expressed from the urethral meatus is indicative of a fistulous connection between the Rectum and the urinary tract. The finding of "meconuria" is typically a trigger for a diverting colostomy with staged reconstruction.

In girls, it is vitally important to determine the number and location of perineal orifices. However, the presence of 3 orifices does not exclude an Anorectal Malformation (ARM). The fistulous opening can be located either in the perineal body, as seen in an anterior anus, or more commonly in the vestibule of the vaginal vault outside the hymen (rectovestibular fistula). Hence, determining the location of all orifices is as important as accounting for them. The presence of two orifices is indicative of a urogenital sinus. In this case, the genitourinary tract and reproductive tract share a common channel, and the gastrointestinal tract has a separate opening in the normal location. Female hypospadias can sometimes be confused with this, and is considered a milder form of urogenital sinus. Note that all boys by definition have a urogenital sinus. A single orifice is consistent with a cloacal anomaly. In this anomaly, the reproductive, urinary, and gastrointestinal tracts share a common orifice. This is the most complex form of female ARM and requires extensive evaluation and staged corrective surgery.

On occasion, fistulous connections may be difficult to identify immediately. A waiting period of approximately 24 hours allows the meconium to descend, and subsequently facilitates visualization. In addition to determining the number and location of the orifices, additional physical findings that should be observed include the "flatness" of the bottom, presence of an anal dimple, midline groove, and whether there is a presacral mass. Patients with relatively flat bottoms are thought to have had poor development of the pelvic musculature, and subsequently have a higher chance of incontinence. A presacral mass with ARM and sacral anomaly are present in infants with the Currarino triad. The presence of an ARM should prompt an evaluation for VACTERL (*v*ertebral anomalies, *a*nal atresia, *c*ardiac defects, *t*racheoesophageal fistula and/or *e*sophageal atresia, *r*enal and radial anomalies, and *l*imb defects) anomalies. Transthoracic echocardiography, renal ultrasound, and plain film radiography of the chest and abdomen are necessary in the initial evaluation for VACTERL. The presence and extent of associated anomalies may affect the choice of the surgical approach for the ARM, as well as the timing for definitive reconstructive surgery. Once an anorectal anomaly is suspected, a paediatric surgical consultation is recommended.

The vast majority of ARMs do not require emergent surgical intervention. The timing of surgery is typically after the first 24 to 48 hours after birth. During the first 24 hours, gas and meconium descend through the rectum to either the lowest level of the pouch or into the fistula. This time allows for a more accurate assessment of the extent of the anomaly. For further management of baby, it is described that two main questions must be explored during the initial postnatal evaluation which is there a need for a staged approach with colostomy or primary repair, and other is there an associated anomaly that requires more urgent intervention?[11-13]

Conflict of Interest: None

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