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Delay in diagnosis of congenital anal stenosis

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ABSTRACT

Although a minor anorectal malformation the delay in diagnosis and treatment of anal stenosis may result in significant early or late complications. Early inspection of the perineum in the neonate to pick up and correct anorectal malformation improves long term outcome but this requires proper anal examination or it could be missed. We present and discuss a case of delayed diagnosis of congenital anal stenosis (a low anorectal anomaly) with an imminent colonic perforation. Severe anal stenosis will always require examination under anesthesia with graded Hegar's dilatation followed by postoperative maintenance. The Hegar dilator is thus both diagnostic and therapeutic in congenital anal stenosis.

Anorectal anomalies are complex congenital abnormalities of either the anus or rectum or both. They affect approximately 1 in 5000 live births, with a nearly equal distribution between males and females [1]. The high anomalies are more severe malformations which include rectal atresia with or without a fistula. They develop at a relatively early stage as a result of incomplete division of the cloaca by the urorectal septum that grows into the cloacal membrane and separates the urogenital system from the rectum at between 4 and 10 weeks gestation. On the surface, the ectodermal pit overlying the rectum becomes the lower part of the anal canal and the low anorectal anomalies which include ectopic anus, covered anus and anal stenosis develop at a later stage. Anal stenosis may be caused by incomplete involution of the cloacal membrane characterized by a narrow rigid fibrous ring. The covered anus probably results from overgrowth of the anal folds and the ectopic anus may result from incomplete migration of the anus back across the perineum (Fig. 1) [1,2]. Generally, delayed diagnosis of ARM is defined as a diagnosis made after the first 48 h [3]. Diagnostic and therapeutic delays in the management of ARM may lead to complications. The overall incidence of a delayed diagnosis has recently been reported to be as high as 21.2% [4]. Spontaneous perforation of the colon is estimated to occur in 2% of neonates with anorectal malformation, and the incidence rises to 9.5% when the diagnosis is delayed [5–8]. The longer the stenosis is untreated the more severely affected the rectum becomes by being grossly distended, insensitive and aperistaltic. This constitutes a severe disability, the treatment is prolonged and major surgery may be required [1,4,6]. The new international classification (Krikenbeck) of ARM has grouped all common anomalies into a major clinical group based on the presence or absence of the fistula in the perineum (Table 1) [9,10].

1. Case report

A 1-month-old baby presented as an emergency with a 1 week history of progressive abdominal distension and pain (crying) but no vomiting. He breast fed normally but later had decreased stool frequency. He had a normal vaginal delivery at term by a 39-yr-old mother and passed normal meconium and later soft stools. He was the second sibling, the first was a normal 3-yr-old male and there was no family history of congenital anorectal or other malformations. On examination, there was a crying baby with a grossly distended abdomen. The baby was dehydrated, small for age and weighed 3 kg. The vital signs were normal. There was visible peristalsis and a tympanic abdomen with 'tinkling' obstructive bowel sounds. Inspection of the perineum revealed a pin-point anus showing excoriation due to passage of 'toothpaste stools,' and with lack of suppleness of the canal on palpation. There was no fistulous connection to skin on the penile shaft or median raphe of the scrotum. The pin hole anus could just accept a rectal thermometer with some resistance. The rest of the physical examination was normal with no other obvious congenital anomaly. A plain abdominal radiograph with oral contrast showed grossly distended

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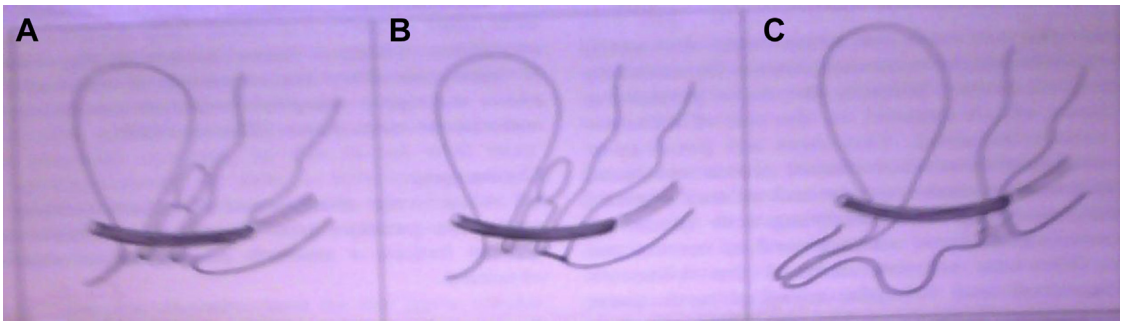

Fig. 1. Low anorectal malformations. A: vestibular ectopic anus, B: anal stenosis, C: covered anus.

colon with faeces but intestinal gas below the level of coccyx-pubis line confirming a low type anomaly (Fig. 2). A full blood count was normal. Examination under anesthesia revealed a pin-point anus that could not accept the little finger. It was dilated with the opening of a 'mosquito' artery forcep. Following the longitudinal division of a rigid fibrotic band, the dilated rectum was entered with the resulting gush of flatus and about 2 L of soft stool. The distended abdomen completely decompressed. The anal canal was dilated until it could accept the little finger and the anal mucosa sutured to the skin in a transverse plane with 3.0 vicryl to maintain correction. The anal tone was satisfactory. The child breast fed following recovery, had normal bowel motions and was discharged on the 2nd postoperative day. Twice daily anal dilation for 2 months was advised. At 2 months follow-up the anus could accommodate a size 12 Hegar. Follow-up is planned to ascertain normal bowel function, and a urogram to exclude any associated urinary tract anomaly, although common but with high-type anomalies.

2. Discussion

The congenital abnormalities of the anus are easily detected but the diagnosis is sometimes missed with serious consequences such as sepsis, aspiration, abdominal distension, colonic perforation, respiratory embarrassment, electrolyte imbalance, and even death or may present later in life with chronic constipation [3–5]. The anus is easily amenable to examination in the newborn. The normal passage of meconium and stools is not a reliable guide to the state of the anus, as a stenosed (narrowed) anus would often allow meconium and soft stool of the newborn to escape as in this case. A rectal thermometer can also usually be easily inserted into the rectum. However as the stool becomes more solid the effect of the stenosis would manifest as bowel obstruction and its sequelae [3–6]. The low type of anorectal malformation should be treated within 48 h of birth if the bowel is obstructed. Anal stenosis although classed as a minor abnormality, if neglected, becomes a major abnormality as the rectum and colon becomes greatly distended, insensitive and apparently aperistaltic (acquired megacolon) which can be difficult to treat. This may be due to bowel wall thickening and loss of the enteric nerve plexus development and coordination from the central autonomic control center in the brain [3] The sooner the diagnosis is made and effective treatment instituted, the better the functional outcome and prognosis [1–6]. Kiely et al. reported unsatisfactory functional results when diagnosis was made at age of presentation greater than 4 months [4].

The technique of anal examination in the newborn entails first inspection and then palpation. During the palpation the two points noted are firstly, the absolute size of the anus, and secondly the anus should be appropriately located within the anal sphincter complex and confirmed by stimulation (cutaneous anal reflex) or electromyography (EMG) if in doubt. A rigid anal canal is an abnormal one. The normal anus will stretch following a slow and gentle insertion with pad first of a well-lubricated little finger and, in most cases a little finger can enter into the rectum. However the examiner's finger may be too big or, as in the case of a very small baby, it is obviously unwise to over dilate and injure the anus. With practice all degrees of anal stenosis can be diagnosed during palpation, without the finger tip entering the rectum as in this case. With gradual increase in size a newborn anus should accommodate a 12 Hegar dilator (Fig. 3) [11]. The result of radiology in distinguishing between high and low anomalies from the position of gas in the distal bowel in relation to the puborectalis on a lateral X-ray taken at the level of the greater trochanter must be interpreted with caution. The X-ray can be misleading if done too early in the

Table 1
Krikenbeck classifications of ARM.

Major clinical groups	Rare regional variants
Perineal (cutaneous) fistula	Pouch colon atresia/stenosis
Rectourethral fistula/atresia/stenosis	Rectal atresia/stenosis
Bulbar fistula	Rectovaginal fistula
Prostatic fistula	H-type fistula
Rectovesical fistula	Others
Vestibular fistula	
Cloaca	
ARM's with no fistula	
Anal stenosis	

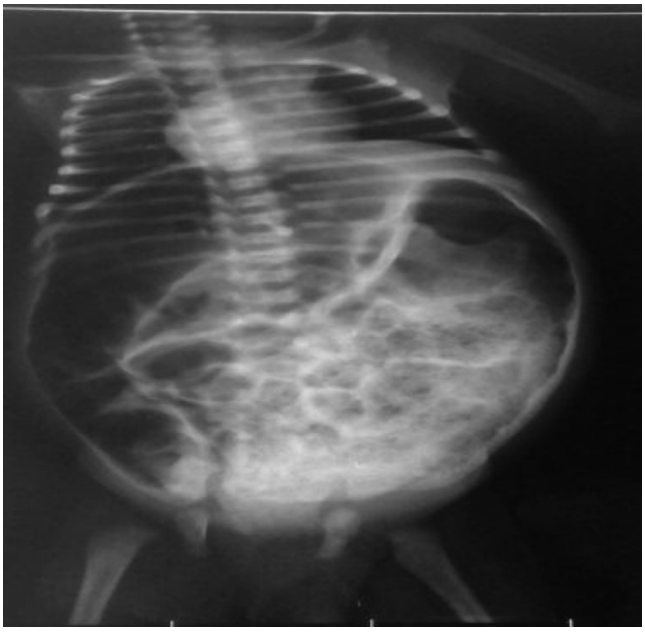

Fig. 2. Supine abdominal X-ray (with oral contrast) showing grossly distended colon.