Journal of Pediatric Surgery (2012) **47**, 720–726

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Spontaneous perforation of the colon and rectum complicating anorectal malformations in neonates

Venkatachalam Raveenthiran*

Rajah Muthiah Medical College, Annamalai University, Tamilnadu, India

Received 7 June 2011; revised 23 July 2011; accepted 25 July 2011

Key words:
Anorectal malformation;
Pneumoperitoneum;
Imperforate anus;
Persistent cloaca;
Neonatal surgery;
Colorectal perforation

Abstract

Background: Spontaneous perforation of the colon is a rare complication in neonates with anorectal malformations (ARMs). There are no detailed studies concerning this complication.

Materials and Methods: A retrospective review of hospital records between 1994 and 2010 revealed 8 cases of bowel perforation among 136 cases of ARM. Eighteen additional cases were culled from the literature by searching Pubmed, Indmed, Embase, and Google Scholar.

Results: Perforations occurred more commonly in males with ARM (85%). Low and high anomalies were equally affected. The median age at diagnosis was 48 hours. Pneumoscrotum and abdominal wall erythema were occasionally suggestive of perforation. In addition to the Rigler sign and collapsed bowel on plain radiographs, a newly described "rectal-tail sign" was useful in recognizing pneumoperitoneum in the lateral view invertogram. A lower midline incision offered optimal surgical access. Two distinct patterns of perforation were identified: type 1 (88%) occurred before surgical decompression of the obstructed colon, whereas type 2 (12%) occurred postoperatively. Type 1 cases were subdivided into cecal (type 1a, 16%), transverse colon (type 1b, 8%), rectosigmoid (type 1c, 60%), and miscellaneous (type 1d, 4%) perforations. Type 1a is best treated with cecostomy and distal colostomy; type 1b, with exteriorization of the perforation; and types 1c and 1d, with closure of the perforation and proximal colostomy. Dense fibrous adhesions caused by extravasated meconium posed technical difficulty during the definitive pull-through operation and was responsible for considerable morbidity. The overall mortality was 19%.

Conclusions: Colorectal perforation is associated with considerable morbidity and mortality in neonates with ARM. Radiographs rather than clinical examination should be relied on for diagnosis of bowel perforation in ARM. Treatment options are chosen according to the subtype of perforation. Because most perforations occurred more than 24 hours after birth, early referral and surgical decompression of the colon may avoid this complication.

Physical examination of the perineum is often sufficient to diagnose anorectal malformation (ARM) in neonates. Notwithstanding, delay in diagnosis is not uncommon, and even in developed countries, a diagnostic delay of 3 to 43 days has been reported in as many as 21% to 32% of newborns [1,2]. In developing countries, initiation of treatment is further delayed by social factors such as poverty, illiteracy, poor transport facilities, and scarcity of specialists [3]. Diagnostic and therapeutic delays in the management of ARM may lead to complications such as sepsis, aspiration,

* 200. Fifth Street, Viduthalai Nagar, Sunnambu Kolathur, Chennai 600117, Tamilnadu, India. Tel.: +91 44 22463882; Mobile: +91 9443310182.
E-mail address: vrthiran@yahoo.co.in.

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.07.025

abdominal distension, colonic perforation, respiratory embarrassment, electrolyte imbalance, and even death.

Spontaneous perforation of the colon is estimated to occur in 2% of neonates with ARM, and the incidence rises to 9.5% when the diagnosis is delayed [1]. Colon perforation accounts for 15% of pneumoperitoneum seen in neonatal age group [4]. Bowel perforation increases the neonatal mortality of ARM from 3% to 23% [4,5]. Literature on this topic is restricted to anecdotal information and isolated case reports [6-16]. This study appears to be the first detailed evaluation of etiopathology, diagnostic difficulties, optimal management, and outcome of colorectal perforations complicating ARM.

1. Methods and materials

Between 1994 and 2010, the author was involved in the care of 149 neonates with ARM (97 boys and 52 girls) at 3 different

Table 1 Summary of literature on colorectal perforations complicating ARM

Sr. no.	Author (year)	Sex	Age [a]	ARM type	Site of perforation	Peritoneal soiling	Management of perforation	Outcome	Remarks
1	Amundsen (1958)	Male	36 h	Low—no fistula	Sigmoid colon	Contained	Anoplasty + PSEC	Survived	In-hospital perforation, preterm, LBW
2	Hass (1958)	Male	1 d	Low (anal stenosis)	Transverse colon	Diffuse	PSEC	Died	
3	Khope (1989)	Male	24 h	Low with fistula	Cecum	–	PSEC	Survived	
4	Stephenson (1992)	Female	?	Cloaca	Vaginal wall	Diffuse	Proximal colostomy	Survived	Probably prenatal perforation with meconium peritonitis, Preterm
5	Digray (2001)	Male	40 h	Low—no fistula	Hepatic flexure	Contained	PSEC	Survived	Postoperative perforation, no pneumoperitoneum in x-ray
6	Digray (2001)	Male	36 h	High—no fistula	Rectum	Contained	CP + PC	Survived	Developed adhesive bowel obstruction, No pneumoperitoneum in x-ray
7	Sharma (2004)	Male	3 d	High—no fistula	Sigmoid colon	Contained	CP + PC	Survived	
8	Sharma (2004)	Male	4 d	High—no fistula	Transverse colon	Diffuse	PSEC	Survived	Parietal erythema present, preoperative peritoneal drainage
9	Sharma (2004)	Male	2 d	High	Cecum	Contained	RHC + ostomy	Survived	No pneumoperitoneum in x-ray
10	Sharma (2004)	Male	6 d	Low—no fistula	Sigmoid colon	–	PSEC	Survived	Postoperative perforation, no pneumoperitoneum in x-ray
11	Komuro (2005)	Male	2 d	Low with fistula	Rectum	–	?	Survived	
12	Eltayeb (2008)	Male	4 d	Low—no fistula	Rectum	–	CP + PC	Survived	
13	Eltayeb (2008)	Male	5 d	High—no fistula	Rectum	–	CP + PC	Survived	
14	Fares (2008)	Male	2 d	Low	Cecum	Contained	Cecostomy	Survived	Postoperative perforation
15	Fares (2008)	Male	3 d	High	Rectum	Contained	CP + PC	Died	Died of sepsis and DIC
16	Eltayeb (2010)	Male	4 d	Low—no fistula	Rectum	Diffuse	CP + PC	Died	Died of sepsis and DIC
17	Eltayeb (2010)	Male	14 d	High with fistula	?	–	CP + PC	Died	Died of sepsis and DIC
18	Sandlas (2011)	Male	24 h	High	Sigmoid colon	Diffuse	CP + PC	Survived	Preoperative peritoneal drainage, LBW

PSEC indicates perforation site exteriorized as colostomy; CP + PC, closure of perforation + proximal colostomy; DIC, disseminated intravascular coagulation; LBW, low birth weight; RHC, right hemicolectomy.

[a] Age indicates age at diagnosis of pneumoperitoneum or onset of perforation.