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ORIGINAL ARTICLE

Clinical experience of complex jejunal atresia

Si Hak Lee · Yong Hoon Cho · Hae Young Kim · Jae Hong Park · Shin Yun Byun

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Abstract
Purpose Small intestinal atresia is relatively common anomaly that causes intestinal obstruction in neonates. Although surgical interventions are usually successful, critical problems could raise in certain cases. This study aimed to identify the distinct clinical characteristics of complex cases of jejunal atresia by retrospective analysis.
Methods Overall, 91 cases of small intestinal atresia, which occured in infants between 2001 and 2010 at Pusan National University Children's Hospital, were reviewed retrospectively. The clinical characteristics of complex jejunal atresia were analyzed.
Results Of the 91 small intestinal atresias, 11 cases of complex jejunal atresia were found: high jejunal atresia with distal deletion, 3; high jejunal atresia with distal multiple atresias, 4; jejunal atresia with distal apple peel appearance, 1; jejunal atresia with colonic atresia, 1; jejunoileal atresia with distal volvulus, 2. Short bowel syndrome was found in four patients and bowel-lengthening procedure was performed in all. Three patients presented with an adhesive intestinal obstruction during the early postoperative period. Postoperative mortality occurred in one patient with distal volvulus.
Conclusions From a surgical perspective, complex jejunal atresia can cause many critical problems after the correction operation. An aggressive and multidisciplinary approach is necessary for managing this condition.

Keywords Complex · Jejunal atresia · Critical

Introduction

Small intestinal atresia is a relatively common anomaly causing intestinal obstruction during the neonatal period. However, neonates with intestinal atresia have a low morbidity and a good long-term outcome in most cases, following early surgical management [1]. Otherwise, several problems could develop in certain cases after initial management. It may lead to delayed recovery of intestinal motility, a decreased nutritional absorption, and other related conditions. The atresias may occur evenly throughout the jejunum and the ileum, may be localized to a short segment when in continuity or may be missed with long segments when having mesenteric and intestinal interruptions [2, 3]. Jejunal atresias, in particular, may present as complex cases that exhibit the conditions listed above, or may be associated with meconium peritonitis due to perinatal perforation [4]. Unlike simple jejunal atresias, complex cases may also be associated with problematic management after initial surgical correction.

The present study was undertaken to evaluate the clinical characteristics and treatment outcomes related to complex jejunal atresia, managed at a single institution.

S. H. Lee
Medical Research Institute, Pusan National University Hospital, Busan, South Korea

S. H. Lee
Department of Surgery, Pusan National University Yangsan Hospital, Yangsan, Gyeongnam 626-770, South Korea

Y. H. Cho (✉) · H. Y. Kim
Department of Pediatric Surgery, Pusan National University Children's Hospital, Beomeo-ri, Mulgeum-eup, Yangsan, Gyeongnam 626-770, South Korea
e-mail: choyh70@pusan.ac.kr

J. H. Park · S. Y. Byun
Department of Pediatrics, Pusan National University Children's Hospital, Yangsan, Gyeongnam 626-770, South Korea

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Materials and methods

This study involved a retrospective review of patients with small intestinal atresia managed at a single center. Cases of duodenal atresia were excluded from the study. During the study period, between 2001 and 2010, 91 cases of intestinal atresia were managed at the Pusan National University Children's Hospital. Subjects included in this study were only those diagnosed as having jejunal atresia with distal deformities and a Type IIIb or IV, according to the classification of Martin and Zerella [5]; cases of simple jejunoileal atresia (Type I, II and IIIa) were excluded. This study was approved by the Institutional Review Board (IRB No. 05-2012-007) and the data have been managed with personal information protection.

The medical records for patients meeting the described criteria were reviewed and the data were collected regarding patient clinical characteristics (e.g., sex, gestational age, birth weight, location of atresia, state of the distal segment, type of procedure, and other associated gastrointestinal anomalies) and management outcomes. The examined postoperative results included postoperative complications, and age and body weight at discharge. Postoperative complications were also compared with those generally observed in cases of simple jejunoileal atresia.

Results

There were 11 cases of complex jejunal atresia, accounting for 12.1 % of the small intestinal atresias managed within the period of this study. These cases were categorized into three groups according to condition of the distal remained segment. The three cases with distal agenesis were classified as Group I. Group II consisted of four cases of high jejunal atresia with multiple distal atresias and cases of multiple atresias in distal segment are shown in Fig. 1. Group III consisted of one case of jejunal atresia with distal apple peel appearance (Fig. 2), one case of jejunal atresia with colonic atresia, and two cases of jejunal atresia with distal volvulus (Table 1).

In the 11 patients, the male to female ratio was 2.7:1; premature babies accounted for 5 (45.5 %) patients, with a median gestational age of 37 weeks; and low birth weight infants included 3 (27.3 %) patients, with a median birth weight of 2,750 g. Most of the subjects (8/11, 72.7 %) were born by cesarean section. The period of hospitalization was longer for group I infants with a median hospitalization period of 57 days. The median body weight at discharge was 3,830 g (Table 2).

In Group I, all patients had a high jejunal atresia with agenesis of the distal small intestine or a midgut deletion. The initial management procedure involved a segmental resection with primary anastomosis. Two of these patients had a loss of the ileocecal valve. The length of the remaining small intestine was, on average, 35 cm, resulting in a short bowel syndrome. These patients received a delayed serial transverse enteroplasty procedure (STEP).

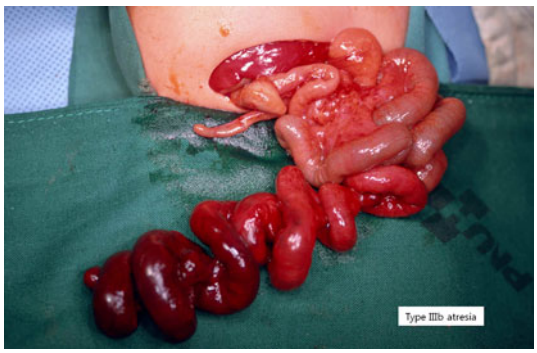

Fig. 2 This case was Type IIIb atresia

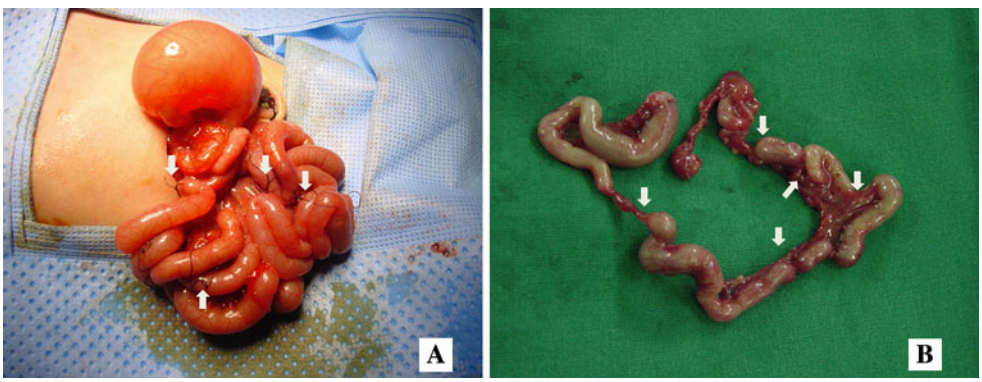

Fig. 1 *White arrows* mark a atretic point: multiple atretic points were located in distal segment (**a**) and resected segment of multiple atresia (**b**)

Table 1 Description of subgroup in complex jejunal atresia

Subgroup	Type	No. of case
Group I	High jejunal atresia with distal agenesis	3
Group II	High jejunal atresia with distal multiple atresias (Type IV)	4
Group III	Jejunal atresia with distal apple peel deformity (Type IIIb)	1
	Jejunal atresia with colonic atresia	1
	Jejunal atresia with distal volvulus	2

They were also managed for intestinal failure-associated liver disease and this resulted in protracted hospitalization. In Group II, all of the patients were premature and showed type IV atresia. The average length of the remaining small intestine in these patients was 80 cm. One patient received multiple laparotomies, also resulting in a short bowel syndrome and a delayed serial transverse enteroplasty. In Group III, each exhibited a distal apple peel deformity (Type IIIb atresia) and jejunal atresia associated with colonic atresia; especially in the latter, the colonic atresia was identified 19 days after the primary operation. In addition, there were two cases of jejunal atresia with a volvulus of the distal segment, which resulted in the death of one patient (Table 3).

Most (7 infants) of the patients had other associated anomalies; the most prevalent was malrotation, which observed in five cases (Table 3). Complications developed in five patients during the early and late postoperative periods. Minor postoperative complications, wound infection and adhesive ileus occurred in three patients, whereas serious complications, short bowel syndrome and cholestasis occurred in four patients. One Group III patient died due to sepsis, followed by multiorgan failure during the early postoperative period (Table 3). Overall, complex jejunal atresia indicated a morbidity and mortality rates of 45.5 and 9.1 %, respectively. These rates were higher than those associated with simple jejunoileal atresia, but the difference was only significant for overall morbidity (Table 4).

Discussion

Intestinal atresia is a well-known cause of intestinal obstruction in the neonatal period. Intestinal atresia may occur with equal frequency in either the jejunum or the ileum, with single atresias being significantly more common (90 %) than cases of multiple atresias (10 %) [2, 6]. The condition is often associated with other complicated conditions, particularly in cases of jejunal atresia. However, adequate surgical management of infants with intestinal atresia has resulted in improved survival and overall outcomes. Multiple factors affect intestinal atresia outcomes, including the length of the remaining bowel and the presence of associated anomalies. A birth weight and the preservation of the ileocecal valve are also regarded as important indicators of patient prognosis [7, 8]. Additionally, an improved prognosis has also been associated with the occurrence of the atresia at more distal locations [9]. The reported mortality rate associated with intestinal

Table 2 Demographics of patients

Subgroup	Sex	Gestational age (weeks)	Type of delivery	Birth weight (g)	Hospital stay (days)	Weight at discharge (g)	Survival
Group I							
Case 1	M	37	C/S	2,660	479	4,420	Yes
Case 2	M	34	NSVD	2,090	319	7,150	Yes
Case 3	F	37	C/S	2,630	100	5,180	Yes
Group II							
Case 1	M	34	NSVD	2,300	63	2,810	Yes
Case 2	M	35	C/S	2,830	51	3,300	Yes
Case 3	M	34	C/S	2,220	130	4,430	Yes
Case 4	M	35	C/S	2,750	57	3,830	Yes
Group III							
Case 1	F	40	C/S	4,110	23	3,870	Yes
Case 2	F	38	C/S	2,830	36	2,870	Yes
Case 3	M	40	NSVD	3,110	25	3,650	Yes
Case 4	M	41	C/S	2,980	12	2,970	No
Median/mean		37/36.8		27,500/2,774	57/117.7	3,830/4,044	

C/S cesarean section, *NSVD* normal spontaneous vaginal delivery

Table 3 Clinical characteristics of patients

Subgroup	Atretic point (distal to the LOT)	Remained small intestinal segment	Ileocecal valve	Management	Associated anomaly	Complications
Group I						
Case 1	15 cm	20 cm jejunoileum	+	SR + PA STEP later	–	Adhesive ileus SBS
Case 2	50 cm	Behind transverse colon	–	SR + PA STEP later	PDA	Adhesive ileus SBS
Case 3	35 cm	Behind splenic flexure	_	SR + PA STEP later	Malrotation	SBS
Group II						
Case 1	30 cm	80 cm jejunoileum	+	SR + PA	–	Wound infection
Case 2	70 cm	100 cm jejunoileum	+	SR + PA	ASD; Malrotation	–
Case 3	20 cm	Type I atresia at distal 9 points	+	SR + PA Stricturoplasty STEP later	–	Adhesive ileus SBS
Case 4	15 cm	Type II atresia at distal 6 points	+	SR + PA	Malrotation	–
Group III						
Case 1	40 cm	80 cm jejunoileum	+	SR + PA	Malrotation	–
Case 2	50 cm	90 cm jejunoileum Lt. colon atresia (Type I)	+	SR + PA Coloplasty later	–	–
Case 3	35 cm	95 cm jejunoileum	+	SR + PA	Trigger thumb	–
Case 4	50 cm	70 cm jejunoileum	+	SR + ileostomy	Malrotation	Sepsis

LOT ligament of Treitz, *SR* segmental resection, *PA* primary anastomosis, *STEP* serial transverse enteroplasty procedure, *SBS* short bowel syndrome, *PDA* patent ductus arteriosus, *ASD* atrial septal defect

Table 4 Comparison of morbidity and mortality between simple and complex small intestinal atresia

	Simple atresia ($n = 80$)	Complex atresia ($n = 11$)	p
Morbidity (%)	7.5 (6/80)	45.5 (5/11)	0.0032*
Mortality (%)	0	9.1 (1/11)	0.1209

* Statistically significant value

atresia has been 30–50 % in the past [2, 6, 9, 10], but has recently been reported to be approximately 10 % for small intestinal atresia [1, 11–13]. Although improvements in the survival of patients with intestinal atresia have been reported, critical problems still remain in certain cases, such as those exhibiting complex jejunal atresia.

The proportion of complicated or multiple atresias is not clearly known, but is usually reported at between 10 and 24 % [2, 6, 14]. Miller reported an unusually high preponderance (88 %) of complex and multiple atresias [4]. Our 10-year experience showed the prevalence of complex jejunal atresia at 12.1 %, similar to the majority of the previous reports.

All cases in the current study had critical problems in the distal remaining segment and 5 patients (45.5 %) showed other associated gastrointestinal anomaly, especially malrotation. This observation suggests that complex jejunal atresia may result from mesenteric vascular accidents, which have been considered to be the major cause of jejunoileal atresia [15–17]. However, all patients with high jejunal atresia with multiple distal atresia (Type IV) and 1 patient with high jejunal atresia with distal agenesis were premature baby, implying that other malformative processes may be involved in the development of complex cases, contrary to classical theory [1, 16, 18].

Operative management of the present cases was based on morphologic findings and involved primary anastomosis that attempted to preserve as much bowel length as possible. Of these patients, 4 (36.4 %) inevitably experienced short bowel syndrome, three of the patients were Group I patients and one was a Group II patient. Patients, such as these, require management with long-term total parenteral nutrition (TPN) and later followed by a serial transverse enteroplasty (STEP). As these patients were subjected to multiple procedures, overall morbidity was affected. For example, postoperative adhesions and wound infections

were more common in these patients and, as a result, they were hospitalized longer and experienced a delay in enteral feedings. Although mortalities were not observed in the present cases with short bowel syndrome, they were all managed for chronic cholestasis and other conditions related to their parenteral nutrition. These management techniques are the most critical problems associated with complex jejunal atresia and also affect later outcomes [1, 18].

Some studies have suggested that low birth weights and associated anomalies appear to negatively impact patient on outcomes [8, 9, 19]. In this study, premature babies and low birth weight infants comprised 45.5 and 27.3 % of patients, respectively, and were not associated with the observed severity of disease. Fortunately, these infants did not manifest any severe associated anomalies that affected outcomes; therefore, such correlations could not be identified in this study. However, the numbers of cases in this study were so small that it is difficult to determine any such correlations with any accuracy.

There is little recent literature focusing on complex cases of jejunal atresia, but a few studies have reported a survival rate of 64–86 % in these cases [2, 4]. Morbidity in these complex cases is generally much higher than that associated with simple cases (7.5:45.5 %, $p = 0.0032$), but this may result from the nature of the disease itself. The overall mortality observed in the present study was 9.1 %, involving only a single case in jejunal atresia with distal volvulus. This result, somewhat lower than the expected mortality, appears to be the result of improvements in parenteral nutritional support and in the surgical procedures used to manage the short bowel syndrome. A STEP was performed in 4 patients exhibiting short bowel syndrome that developed early. However, they all inevitably experienced chronic cholestasis and other TPN (totally parenteral nutrition) related problems, which may also affect long-term outcomes.

Complex jejunal atresia is a rare disease with higher morbidity and mortality than that associated with less complex cases. In spite of proper initial management, this complex disease may yield critical problems that require long-term management or that result in the death of the patient. TPN and additional operative procedure also seem to be essential components of the management of any associated short bowel syndrome outcomes. Taken together, these observations indicate that, particularly due to the infrequent occurrence of this condition, appropriate management of complex jejunal atresia requires pediatric surgeons to treat it with an aggressive and multidisciplinary approach.

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