



ORIGINAL ARTICLE

Comparison of outcomes following three surgical techniques for patients with severe jejunoileal atresia

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Abstract

Background: Severe jejunoileal atresia is associated with prolonged parenteral nutrition, higher mortality and secondary surgery. However, the ideal surgical management of this condition remains controversial. This study aimed to compare the outcomes of patients with severe jejunoileal atresia treated by three different procedures.

Methods: From January 2007 to December 2016, 105 neonates with severe jejunoileal atresia were retrospectively reviewed. Of these, 42 patients (40.0%) underwent the Bishop–Koop procedure (BK group), 49 (46.7%) underwent primary anastomosis (PA group) and 14 (13.3%) underwent Mikulicz double-barreled ileostomy (DB group). Demographics, treatment and outcomes including mortality, morbidity and nutrition status were reviewed and were compared among the three groups.

Results: The total mortality rate was 6.7%, showing no statistical difference among the three groups ($P = 0.164$). The BK group had the lowest post-operative complication rate (33.3% vs 65.3% for the PA group and 71.4% for the DB group, $P = 0.003$) and re-operation rate (4.8% vs 38.8% for the PA group and 14.3% for the DB group, $P < 0.001$). Compared with the BK group, the PA group showed a positive correlation with the complication rate and re-operation rate, with an odds ratio of 4.15 [95% confidence interval (CI): 1.57, 10.96] and 12.78 (95% CI: 2.58, 63.29), respectively. The DB group showed a positive correlation with the complication rate when compared with the BK group, with an odds ratio of 7.73 (95% CI: 1.67, 35.72). The weight-for-age Z-score at stoma closure was -1.22 (95% CI: $-1.91, -0.54$) in the BK group and -2.84 (95% CI: $-4.28, -1.40$) in the DB group ($P = 0.039$).

Conclusions: The Bishop–Koop procedure for severe jejunoileal atresia had a low complication rate and re-operation rate, and the nutrition status at stoma closure was superior to double-barreled enterostomy. The Bishop–Koop procedure seems to be an appropriate choice for severe jejunoileal atresia.

Key words: Jejunoileal atresia; Bishop–Koop procedure; double-barreled ileostomy; outcomes

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Introduction

Congenital jejunoileal atresia is the most common cause of neonatal bowel obstruction, with a reported morbidity ranging from 0.4 to 3.1/10,000 live births [1–4]. Although most patients with congenital jejunoileal atresia have excellent outcomes after initial surgical correction, some patients who suffer from progressive abdominal distension required long-term parenteral nutrition and those with anastomotic leakage or short-bowel syndrome required secondary surgery. Previous studies have revealed that luminal discrepancy, meconium peritonitis, and type IIIb and type IV intestinal atresia were associated with prolonged parenteral nutrition, higher mortality and secondary surgery [5, 6].

Improvement in surgical techniques that can avoid vital complications, such as anastomotic leak or short-bowel syndrome, and promote bowel function recovery has become a serious challenge for most surgeons. However, the ideal surgical procedures have been a matter of debate for a long time and the current treatments mainly depend on the surgeon's skills and experience [5]. Primary anastomosis, Mikulicz double-barreled ileostomy and the Bishop–Koop procedure are common surgical procedures widely used for the treatment of severe jejunoileal atresia. Each of these techniques has potential complications as well as varied success rates in the treatment of jejunoileal atresia [2, 6–8]. There are few comparative studies that have evaluated the outcomes of different surgical techniques on severe jejunoileal atresia. Hence, this study aimed to compare the mortality, morbidity and other clinical outcomes for patients with severe jejunoileal atresia who underwent primary anastomosis, Mikulicz double-barreled ileostomy or the Bishop–Koop procedure.

Patients and methods

Patients

Patients with severe jejunoileal atresia at Guangzhou Women and Children's Medical Center between January 2007 and December 2016 were retrospectively reviewed. Severe jejunoileal atresia was defined as follows: (i) proximal jejunal atresia (at a distance of less than 20 cm from the Treitz ligament), type IIIb intestinal atresia or type IV intestinal atresia accompanied by a ratio between the proximal intestinal diameter and the distal diameter of greater than 4:1 [6]; and (ii) intestinal atresia complicated by meconium peritonitis, which is characterized by one or more abnormalities (such as ascites, intestinal dilation and pseudocyst) under ultrasonic imaging in addition to intra-abdominal calcification. Patients who met any one of the above criteria were included in this study. Patients with major associated anomalies and sepsis before surgery were excluded.

Surgical treatment

Patients were divided into three groups based on the surgical procedures, which included primary resection and anastomosis (PA group, Figure 1A); construction of a Mikulicz double-barreled ileostomy (DB group), where the bowel loop was completely divided and the two ends are brought together as the end stomas with resection of the large dilated loop (Figure 1B); and the Bishop–Koop procedure (BK group), where a Roux-en-Y anastomosis and ileostomy were performed for removing the maximally distended proximal segment and establishing a 'safety valve' (Figure 1C). The choice of surgical procedure was made by the attending surgeon based on the patient's hemodynamic state and the condition of the bowel during laparotomy. The stoma closure is an elective surgery performed 6 weeks after the operation when the patients had gained ideal weight. A written informed consent form was obtained from all participants' parents or guardians. The research protocols were approved by the Ethics Committee of Guangzhou Women and Children's Medical Center.

Data collection

Patients' characteristics, including sex, age at operation, birth-weight, birth gestational age and type of atresia, were collected. Surgical data, including surgical approach, operative time and intra-operative blood loss, were also collected. Clinical outcomes such as duration of hospital stay, duration of total parenteral nutrition (TPN), time to initial enteral nutrition after operation, post-operative complications, patients requiring re-operation and body weight during stoma closure were documented. Complications included anastomotic leakage, high-output stoma [output at the stoma >20 mL/(kg·day) with severe water–electrolyte imbalance requiring intravenous fluid therapy [8]], intestinal obstruction, necrotizing enterocolitis (NEC) and cholestasis. Premature birth was defined as gestational age >37 weeks. Low birth weight was defined as birth weight <2500 grams. The weight-for-age Z-score (WAZ) was calculated according to the World Health Organization (WHO) growth standards of children <2 years of age and patients with a WAZ of less than -2 were defined as malnourished [9].

Statistical analysis

Continuous data were expressed as mean with standard deviation or median with interquartile range (IQR). The data were compared by using one-way analysis of variance or t-test for normally distributed data and Kruskal–Wallis *H* test or Mann–Whitney *U* test for non-normally distributed data. Categorical data were presented as numbers with percentages and were compared using the χ^2 test or Fisher's exact test. Multivariate logistic regression was used to evaluate the association between surgical procedures and post-operative complications and re-

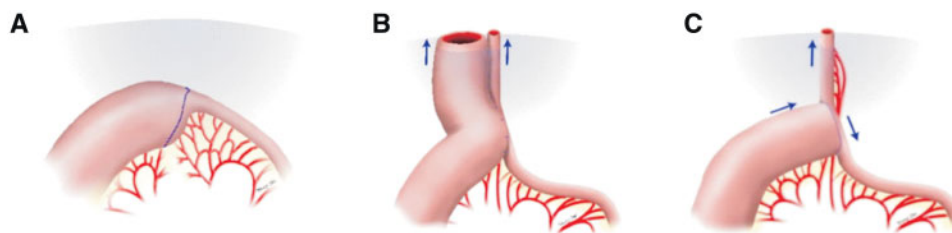


Figure 1. Schematic drawings of various surgical approaches for jejunoileal atresia. (A) Primary anastomosis. (B) Double-barreled Mikulicz ileostomy. (C) Bishop–Koop procedure.

Table 1. Characteristics of patients with severe jejunoileal atresia

Variable	BK group (n = 42)	PA group (n = 49)	DB group (n = 14)	P-value
Age at surgery, days	2 [1–3.25]	3 [1–5]	2.5 [1–8.5]	0.323
Male	20 (47.6)	29 (59.2)	9 (64.3)	0.415
Premature	22 (52.4)	23 (46.9)	2 (14.3)	0.042
Low birthweight	13 (31.0)	18 (36.7)	3 (21.4)	0.541
Location of atresia				
Ileum	25 (59.5)	31 (63.3)	13 (92.9)	0.066
Jejunum	17 (40.5)	18 (36.7)	1 (7.1)	
Type of atresia				
I–IIIa	24 (57.1)	32 (65.3)	13 (92.9)	0.051
IIIb–IV	18 (42.9)	17 (34.7)	1 (7.1)	
Meconium peritonitis	26 (61.9)	21 (42.9)	5 (35.7)	0.105

Data are presented as n (%) or median [interquartile range].

BK, Bishop-Koop; PA, primary anastomosis; DB, Mikulicz double-barreled ileostomy.

Table 2. Perioperative data of patients with severe jejunoileal atresia

Variable	BK group (n = 42)	PA group (n = 49)	DB group (n = 14)	P-value
Operative time, hours	2.17 [1.67–2.85]	1.83 [1.42–2.32]	2.16 [1.63–2.44]	0.337
Intra-operative blood loss, mL	5 [2–10]	5 [2–10]	7.5 [4.25–10]	0.460
Transfusion	11 (26.2)	23 (46.9)	7 (50.0)	0.086
Duration of TPN, days	13 [9–21]	19 [11–29.5]	14 [8.75–22.75]	0.078
Time to enteral nutrition, days	11 [7–13.75]	11.5 [7–20.5]	6 [5–9.5]	0.069
Hospital stay, days	21.5 [16–36.25]	25 [16–38]	18.5 [13.75–33.5]	0.682
Weight at discharge, kg	2.78 ± 0.56	2.62 ± 0.51	2.89 ± 0.47	0.164
Post-operative complication	14 (33.3)	32 (65.3)	10 (71.4)	0.003
Cholestasis	8 (19.0)	6 (12.2)	0 (0.0)	0.184
High-output stoma	1 (2.4)	–	9 (64.3)	<0.001
Intestinal obstruction	3 (7.1)	12 (24.5)	1 (7.1)	0.048
Anastomotic leak	3 (7.1)	11 (22.4)	–	0.044
Necrotizing enterocolitis	0 (0.0)	4 (8.2)	0 (0.0)	0.184
Perioperative death	3 (7.1)	4 (8.2)	0 (0.0)	0.164
Re-operation	2 (4.8)	19 (38.8)	2 (14.3)	<0.001
Age at stoma closure, months	6 [4–7.88]	–	5.62 [4.61–6.14]	0.932
WAZ at stoma closure	−0.99 ± 1.66	–	−2.52 ± 2.16	0.021

Data are presented as n (%), mean ± standard deviation or median [interquartile range].

BK, Bishop-Koop; PA, primary anastomosis; DB, Mikulicz double-barreled ileostomy; TPN, total parenteral nutrition; WAZ, weight-for-age Z-score.

operation. Data were expressed as adjusted odds ratios (ORs) and 95% confidence intervals (CIs). Analysis of covariance was used to assess the association of surgical procedures and WAZ at colostomy closure with adjusted mean and 95% CI. A two-sided *P*-value of <0.05 was considered to be statistically significant. The statistical analysis was performed by using R version 3.4.1 and GraphPad Prism Version 5.0 (La Jolla, CA, USA).

Results

Participant characteristics

A total of 105 patients were included in the study. Of these, 58 patients were males and 47 were females. Forty-two (40.0%) patients were included in the BK group, with a median age at intestinal surgery of 2 days (IQR: 1–3.25); 49 (46.7%) patients were included in the PA group, with a median age at intestinal surgery of 3 days (IQR: 1–5); and 14 (13.3%) patients were included in the DB group, with a median age at intestinal surgery of 2.5 days (IQR: 1–8.5). Patient characteristics are summarized in Table 1. The DB group had the lowest ratio of premature births

(14.3%). No significant differences in type and location of atresia among the three groups were observed.

Clinical outcomes

The clinical outcomes are summarized in Table 2. Seven patients (6.7%) died during the perioperative period. Among these, three died due to uncontrolled sepsis, two due to liver failure with infection and two due to malnutrition owing to treatment abandonment by their parents. The BK group experienced the lowest post-operative complication rate (33.3%, *P* = 0.003) and re-operation rate (4.8%, *P* < 0.001). The most common complications in the PA group were intestinal obstruction (24.5%) and anastomotic leakage (22.4%), whereas that in the DB group was high-output stoma (64.3%). There was a significant decline in these complications in the BK group. The TPN duration in the BK group was shorter than that in the PA group, whereas the time to enteral nutrition was similar between the BK and PA groups. The length of hospital stay in the PA group was the longest when compared with the BK and DB groups, but the difference was not statistically significant (*P* = 0.682). The

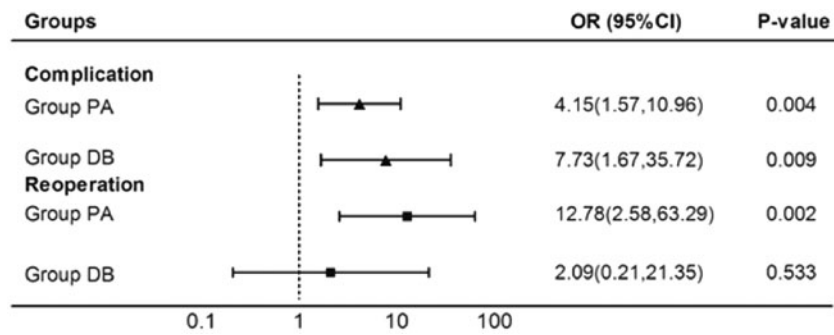


Figure 2. Forest plot of adjusted odd ratios with 95% confidence intervals for the association between three surgical techniques and post-operative complication and re-operation (compared to the BK group). Adjusting for age, sex, full-term and low birthweight, location of atresia, type of atresia and meconium peritonitis. x-axis: conversion of Base-10 logarithm.

mean WAZ score during stoma closure in the BK group was higher than that in the DB group (-0.99 vs -2.52 , $P = 0.021$).

Associations between surgical technique and clinical outcomes

Multivariate logistic regression was performed to compare the BK group with the PA group. The results showed a positive correlation with regard to the complication rate (OR = 4.15, 95% CI: 1.57, 10.96) and the re-operation rate between the two groups (OR = 12.78, 95% CI: 2.58, 63.29). The DB group also demonstrated a positive correlation with the complication rate (OR = 7.73, 95% CI: 1.67, 35.72). But the re-operation rate between the DB and BK groups showed no significant difference (Figure 2). The results of the analysis of covariance after adjusting for full-term delivery showed that low birthweight, location of atresia, type of atresia and meconium peritonitis, and the mean WAZ in the BK group were higher than in the DB group at stoma closure [-1.22 (95% CI: $-1.91, -0.54$) vs -2.84 (95% CI: $-4.28, -1.40$), $P = 0.039$].

Discussion

With modern advances in medical care, including parenteral nutrition and neonatal intensive care, the survival rate of neonates with jejunoileal atresia has improved dramatically in the twentieth century [2, 10]. However, complication rate of severe jejunoileal atresia still remains high due to the great disparity in the proximal and distal bowel, ineffective peristalsis, multiplicity of lesions and associated peritonitis. So, the appropriate treatment for severe jejunoileal atresia remains controversial. The common techniques include primary anastomosis, double-barrel enterostomy and the Bishop-Koop procedure. In this retrospective study, we initially compared the outcomes of the three surgical techniques for severe jejunoileal atresia.

The total mortality of severe jejunoileal atresia in this study was 6.7% (7/105), which was similar to the figures described in other studies [2, 6]. Among the three surgical procedures, primary anastomosis had the highest mortality (8.2%), followed by the Bishop-Koop procedure (7.1%), and no patient died in the DB group. The differences showed no statistical significance ($P = 0.164$).

The most common complications included intestinal obstruction and anastomotic leakage in the PA group, causing the highest re-operation rate. Anastomotic leakage is a serious complication after the repair of intestinal atresia [11]. The high incidence of anastomotic leakage in apple-peel atresia (14%) when compared with the other types of intestinal atresia (4%) is caused due to inadequate blood supply at the anastomotic site,

as it involves a single-artery retrograde blood supply [12, 13]. Approximately one-half of the sepsis cases are due to anastomotic leakage and therefore functional anastomosis remains a key prognostic factor for the early survival of these children [13]. In the current study, we noticed that the PA group had a 22.4% anastomotic leakage and 24.5% intestinal obstruction owing to the great disparity in the proximal and distal gut, and ineffective peristalsis. Also, the main reasons for the highest re-operation rate were anastomotic leakage and intestinal obstruction. Due to the high complication and re-operation rates, primary anastomosis was not recommended for the management of severe jejunoileal atresia.

High-output stoma was the most prominent problem in the DB group. Construction of a certain temporary ostomy for the discharge of fecal contents can protect a fragile anastomosis and alleviate symptoms [14]. However, stoma creation may provoke post-operative complications, with an estimated incidence of 20%–60% [15, 16]. These complications included stoma stenosis, retraction, necrosis, small-bowel obstruction, skin excoriation, and fluid and electrolyte abnormalities. An issue of excessive output from the stoma and its relation to electrolyte abnormalities has raised interest in research [17]. Some studies have identified this complication as a precursor to dehydration and renal dysfunction, with an estimated incidence of 1%–17% [18, 19], while some believe it to be the reason for 4%–43% of hospital readmissions [20]. In our study, a high-output stoma of as high as 71.4% was observed in the DB group. So, a double-barreled ileostomy was considered to be safe, but it also carries some additional morbidity related to stoma management such as fluid and electrolyte abnormalities, malnutrition and subsequent surgical closure. If double-barreled ileostomy is chosen for the treatment of severe jejunoileal atresia, it is necessary to closely monitor the volume of output from the stoma during the short-term follow-up period.

The total complication rate remained the lowest in the BK group when compared with the other two groups. The Bishop-Koop procedure has the advantages of the early restoration of bowel continuity, favorable nutritional management and an easy extraperitoneal approach [21, 22]. The presence of a stoma just proximal to the anastomosis acts as a vent, partially decompressing the anastomosis and reducing the chances of anastomotic leakage. Also, 7.1% anastomotic leakage was observed in the BK group, but was much lower than that of the PA group (22.4%).

The nutritional status at stoma closure in the BK group was better than that of the DB group. Ng *et al.* [23] reported that patients with an ileostomy tended to have low body weight, body mass index and lean body mass. A high-output stoma was associated with poor weight gain [16]. Crealey *et al.* [8] reported

that the weight gain or weight loss was closely related to the consistency and volume of the stoma output after the construction of ileostomy in newborns. On the other hand, the Bishop-Koop procedure can maintain the continuity of the gut, ensuring the early utilization of the distal small bowel and colon, promoting the early recovery of bowel function and decompressing the anastomotic pressure. However, due to the lack of follow-up nutritional data regarding patients in the PA group, the long-term nutritional status between the BK and PA groups could not be compared.

However, the results of the current study have certain limitations. First, it is a retrospective study that is conducted using historical controls, and only information that has been previously recorded and is available as per our hospital's medical records. Second, the sample size of the DB group is small and data were also lacking regarding the nutritional status of the PA group. Finally, the gestational age among the three groups did not reach statistical consistency in the baseline characteristics due to the small sample size.

In conclusion, despite these limitations, the results presented some evidence that primary anastomosis was not a good choice for severe jejunoileal atresia due to its high complication and re-operation rates. Double-barreled enterostomy is considered to be the safest procedure, but also carried some additional morbidity related to stoma management such as high-output stoma and malnutrition. The Bishop-Koop procedure demonstrated the lowest complication and re-operation rates, and the nutrition status at stoma closure was superior over double-barreled enterostomy. The Bishop-Koop procedure seems to be an appropriate choice for severe jejunoileal atresia due to fewer post-operative complications and better nutrition status. A prospective trial is still warranted to confirm these results and improve the outcomes of severe jejunoileal atresia in the future.

Authors' contribution

All authors designed the study and interpreted the results; H.Z. collected the data; H.Q.Z. provided statistical advice. Y.F.P. and H.Q.Z. shared the first authorship for this work. All authors read and confirmed the final version of this paper.

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None.

Conflicts of interest

None declared.

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