

Review Article

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Apple-Peel intestinal atresia: A systematic review

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KEYWORDS

Apple peel atresia, Intervention, Primary resection, Complications, Neonate

ABSTRACT

Background: Apple-peel bowel atresia (APA) is rare and thus lacks proper management guidelines. This systematic review analyzes various interventions found in the literature for this type of atresia and highlights the outcomes of each intervention with their complications.

Methods: The relevant literature on PubMed, Scopus, Embase, and Cochrane was reviewed from November 2020 to January 2021. Articles published in English, French, Italian, or Spanish between 1990 and 2020, focusing on APA, were included. Data on demography, clinical profile, management provided, time to achieve full feed, length of hospital stay, complications, etc., reviewed.

Results: A total of 2495 articles were found, of which only 48 met the inclusion criteria. Among these, 125 patients were treated, with 15 deaths. The most frequently employed intervention was primary resection, with or without tapering, while ostomy creation was less commonly used. The most frequent complication across all techniques appeared to be cholestasis, followed by stenosis of the anastomosis and sepsis. Additionally, there were a total of 7 cases of short bowel syndrome (SBS).

Conclusion: The management of apple-peel atresia seems variable in various centers of different countries. The review failed to assess which interventions could lead to faster full enteral feeding. The distal part of the anastomosis must be considered as an active part in the resumption of bowel function.

INTRODUCTION

Apple-peel bowel atresia (APA) is a rare form of intestinal atresia characterized by a unique anatomical anomaly where the superior mesenteric artery is absent. The inferior mesenteric artery or the right ileocolic branch supplies the distal segment, resulting in a distinctive helical configuration known as the "apple-peel" [1]. To our knowledge, the current literature does not compare surgical techniques specifically for managing Apple-peel bowel atresia (APA); consequently, surgeons who have not previously encountered this situation may face uncertainty regarding the most appropriate course of action.

Recognizing the potential ramifications of different surgical approaches prompted us to undertake this study: the primary aim of our systematic review was to compile all interventions reported in the literature for Apple-peel bowel atresia (APA) and illuminate the outcomes associated with each. Our secondary objective was to compare interventions based on their potential to expedite full enteral feeding and explore whether specific anatomical considerations favor one surgical approach over another.

METHODS

The review analyzed the literature published on APA on PubMed, Scopus, Embase, and Cochrane from November 2020 to January 2021. The search strategy involved entering "congenital intestinal atresia" and 'Intestine, Small/abnormalities' in the search fields [all fields or MeSH Major Topic]. The complete search strategy, including keywords and the full scope of the search, can be found in the appendices section (Appendix 1).

Using the EndNote software (Clarivate EndNote V.20, available at https://www.myendnoteweb.com/EndNoteWeb.html),

the articles were organized, and duplicates were removed. Subsequently, a list of unique articles was

extracted from EndNote and evenly distributed among the four authors responsible for article selection. A comprehensive list of all articles was shared with all authors via a shared Google document.

Once the comprehensive list of articles was available, the authors collectively identified a set of relevant variables to consider, including:

- Sex of the child
- Birth weight
- Antenatal diagnosis
- Presence of associated abdominal malformations (such as biliary tract atresia and gastroschisis)
- Age at surgery
- Type of surgery
- Length of hospitalization
- Any complications encountered
- Duration of parenteral nutrition, if applicable
- Time taken to achieve full enteral feeding (FEFTA), defined as the number of postoperative days until patients were consuming full caloric intake via oral feeding
- Length of follow-up

All the data were put in the form of an Excel spreadsheet and shared among authors through Google Drive.

Articles were chosen according to the following criteria:

Inclusion criteria:

- Articles that dealt with Apple Peel/type III b bowel atresia
- Articles included in the period from January 1990 to November 2020
- Articles in English, French, Italian, and Spanish.

Exclusion criteria:

- Grey literature
- -Abstract or full text not available
- -Veterinary articles
- Reviews, book chapters
- Presence of associated malformations such as biliary tract atresia or gastroschisis
- Failure to meet completeness in the parameters sought and considered indispensable as the type of intervention proposed and the outcome.

Four Authors (FL, CB, OO, TM) selected articles considered eligible. In the case of disputes concerning the included and non-included articles, a final decision was arbitrated by the fifth author (MLPM).

The PRISMA algorithm for systematic reviews was used to describe it (Fig. 1).

Assessment bias:

In all instances, we encountered case reports or series. Consequently, we utilized the bias assessment tool provided by the Joanna Briggs Institute [2], which comprises a set of eight standardized questions. These questions were applied uniformly to evaluate all articles included in the study.

RESULTS

Figure 1 shows the results obtained after article selection.

A total of 2495 articles were analyzed and only 48 were found to be eligible for our study (Appendix 2).

Table 1 presents the bias assessment for each article. It was observed that in most cases, crucial information such as the length of the remaining bowel, duration of parenteral nutrition, and FEFTA were inadequately reported. Additionally, data were scarce regarding preoperative conditions, antenatal diagnosis, or age at surgery, leading to an "Unclear" assessment for most articles evaluated. For instance, antenatal history was detailed in only 52 cases (41.6%), with "bowel obstruction" being the most frequently described sign. In five cases, no antenatal identified. Furthermore, anomalies were regarding parenteral nutrition and FEFTA were notably lacking, with only 8% of authors reporting this parameter. While there is some missing data regarding prematurity, it was noted that 55.2% of patients (n=69) were born prematurely, whereas 6.4% (n=8) were born at term.

The interventions suggested by the authors for this type of malformation can be categorized into four main approaches:

- 1. Resection of the dilated proximal loop and anastomosis, with or without tapering (RA)
- 2. Primary Anastomosis (PA)
- 3. Bowel tapering and anastomosis (TA)
- 4. Ostomy (such as Bishop-Koop type or other) with delayed anastomosis (ODA). The 48 articles have been categorized according to the type of intervention performed, encompassing descriptions of 125 patients. In instances where multiple interventions were detailed within a single article, it was represented across all relevant categories. Tables 2a and 2b provide an overview of the articles segmented by type of intervention along with complications and outcomes. Tables 3, 4, 5, and 6 offer a comprehensive summary of each specific procedure.

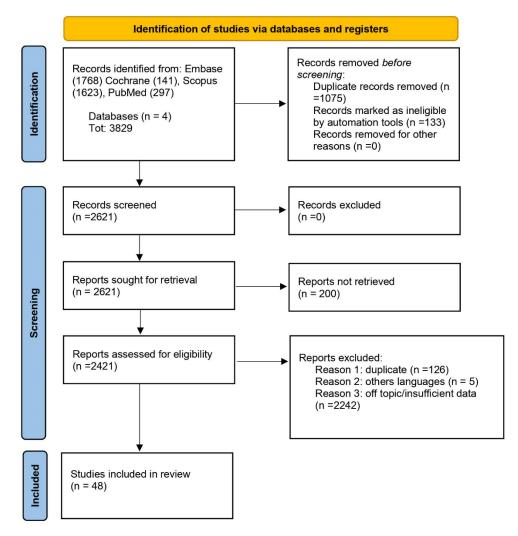


Figure 1: PRISMA flow diagram

Table 1: Assessment Bias

			0.010 111100					
	Item 1	Item 2	Item 3	Item 4	Item 5	Item 6	Item 7	Item 8
Yes	45	44	42	47	47	29	43	42
No	1	3	2	/	/	1	/	5
Unclear	/	/	1	/	/	16	4	/
Not applicable	1	/	2	/	/	1	/	/

Table 2a: Summary of articles RA= resection and anastomosis; PA= primary anastomosis; TA= tapering and anastomosis; ODA Ostomy and delayed anastomosis; SBS= short bowel syndrome

	Articles	Patients	Complications (n)	SBS (n)	Mortality (n)
RA	24	46	28,2% (13)	8,6% (4)	28,2% (13)
PA	15	19	31,5%(6)	5,2% (1)	10% (2)
TA	6	44	93 % (52)*	0	0
ODA	9	16	56,2% (9)	33,3 % (2)	0

^{* 52} complications occurred in 41 patients (93% of the cohort).

Resection and anastomosis, with or without Tapering (RA)

This involves resection of the dilated proximal loop and anastomosis to the distal loop with or without tapering of the proximal loop. This surgical approach was adopted by most authors, comprising 24 articles and a total of 46 patients. The rationale behind this treatment choice is rooted in the notion that the dilated proximal loop may contribute to bowel dysmotility to some extent.

Table 2b: Details of complications for each type of intervention

Complications	RA	PA	TA	ODA
Adhesion-obstruction	2	0	4	0
Sepsis	3	0	9	0
SBS	4	1	0	2
Leakage	1	1	0	0
Intestinal obstruction/ stricture	2	2	4	6
Dysmotility	0	2	0	0
Cholestasis	0	0	20	0
Dumping syndrome	0	0	0	1
Electrolyte imbalances	0	0	15	0
Bowel gangrene	1	0	0	0

Many articles fail to specify the length of the residual bowel, and it is consistently emphasized that the decision between interventions remains at the discretion of the operating surgeon.

Mortality was recorded in 13 cases (28.2%), predominantly among patients from low-income countries, and to our knowledge, all these patients were preterm. Complications associated with this technique included four cases of short bowel syndrome (SBS), three cases of sepsis, two cases of adhesion-related bowel obstruction, and only one case of anastomotic leakage.

Only four authors reported the FEFTA, which ranged between 24 and 40 days. Table 3 provides a comprehensive overview of the results of this technique.

Primary anastomosis (PA)

Primary anastomosis involves restoring bowel continuity without tapering or bowel resection. Fifteen authors opted for this technique, totaling 19 patients. Table 4 provides a summary of the characteristics of these articles. None of the authors provided details regarding the difference in caliber between the proximal and distal loops. However, they justified the choice of direct anastomosis by explaining that resections could increase the risk of short bowel syndrome (SBS) [3].

Among these articles, only one case of SBS was reported. The most described complications included anastomotic strictures (two cases) and dysmotility (two cases). In these instances of dysmotility, impaired bowel motility prompted the authors to perform secondary tapering as it had not been initially conducted. Additionally, one case of anastomotic leakage was identified.

Two deaths occurred in patients for whom parenteral nutrition was not feasible and in patients with late presentation coupled with severe prematurity. Three authors reported the FEFTA, which ranged from 23 to 38 days.

Ostomy-delayed anastomosis (ODA)

This technique, chosen by nine authors, ranks third in frequency. All 16 patients underwent an interruption of intestinal continuity, with various types of ostomies (such as lateral, "baguette," and Bishop-Koop type) considered within this category. In all cases except one, ostomy reversal was performed between 30 and 60 days after the initial intervention. Complications included six cases of anastomotic stenosis and two cases of short bowel syndrome (SBS).

Only two authors reported the FEFTA, which ranged from 31 to 40 days respectively. There were no reported mortalities in this group. The complication rate was 56.2%. Table 6 details the breakdown of all results.

Tapering (TA)

This technique aims to reduce the caliber discrepancy between the dilated proximal loop and the usually tiny distal loop. Tapering is performed to achieve more uniform segment sizes, facilitating anastomosis between segments of more adequate size. Six authors opted for this technique, involving a total of 44 patients.

Notably, this technique was employed in the largest case series reported in the literature, encompassing 39 children. The most frequently reported complications in this series were cholestasis and electrolyte imbalance, followed by sepsis, anastomotic stenosis, and postoperative adhesions. The authors of this significant review also described a laparoscopic approach for Apple-peel bowel atresia (APA). However, no details about FEFTA were provided in this group of articles. Among these articles, only three patients had no complications (6.8%), while 41 children (93%) experienced more than one complication.

DISCUSSION

Apple-peel intestinal atresia (Type IIIb) is considered one of the rarest forms of atresia, accounting for approximately 10% of all intestinal atresias [4]. The

exact cause of this malformation remains unclear, although the prevailing hypothesis suggests a vascular origin. Environmental and genetic etiological factors are also suggested [5-11].

What emerges from our research is that the predominant treatment approach for this type of malformation involves resection of the proximal loop, with or without tapering. Authors who employ this technique justify their choice by suggesting that resection may mitigate the risk of anastomotic dysfunction or dysmotility, which are considered potential short-term postoperative complications in Apple-peel bowel atresia (APA). Dewberry [12], in a study encompassing all types of bowel atresia, raises the question of which procedure yields better outcomes between resection with or without tapering (RA) and bowel loop tapering and anastomosis (TA). However, this question remains unanswered due to the scarcity of cases. Nonetheless, it is noteworthy that in Dewberry's study, all cases of APA were treated with resection.

Furthermore, Ozguner [13] elucidates the microanatomical abnormalities observed in the proximal loop of intestinal atresia, supporting the necessity for resection. Similarly, Saha [14] explains the differences in the representation of Cajal cells and muscle layers in various types of atresia, asserting that the discrepancy between the proximally dilated and distal parts in terms of motility justifies resection.

Our findings indicate that the most common complications in Apple-peel bowel atresia (APA), irrespective of the surgical technique employed, appear to be cholestasis, followed by sepsis and stenosis of the anastomosis (Table 2b). This observation is corroborated by Festen et al. [15]. Additionally, our analysis suggests that these complications may be more prevalent in cases treated with the bowel tapering and anastomosis (TA) technique. However, we acknowledge the inherent heterogeneity and variability among the cases reviewed, which precludes the establishment of statistical significance for this observation.

Intriguingly, some authors have suggested a higher incidence of anastomotic leak in Apple-peel bowel atresia (APA) compared to other types of intestinal atresia (14% vs. 4%), hypothesizing that inadequate blood supply at the anastomotic site, with its retrograded single artery blood supply, could be the underlying cause [16]. However, our findings do not align with these results, as anastomotic leakage does not appear to be a frequent complication in our review (occurring in only two cases treated with resection and primary anastomosis). Conversely, anastomotic strictures or stenosis were observed more frequently, with six cases in ostomy with delayed anastomosis

(ODA), four cases in loop bowel tapering and anastomosis (TA), two in primary anastomosis (PA,), and two in resection with or without tapering (RA).

Due to the lack of comprehensive information on the duration of full enteral feeding time (FEFTA), no definitive conclusions can be drawn in this regard. Similarly, complete data are absent regarding the length of hospital stay, further complicating the analysis.

The mortality rates associated with Apple-peel bowel atresia (APA) have demonstrated significant improvement over time, decreasing from 70% before 1970 to 26% after 1970, and further dropping to 14% in the last two decades [15]. However, in cases complicated by volvulus, mortality rates can soar as high as 90% [17].

In our review, we identified 15 cases of mortality, accounting for 12% of the entire series. Notably, most of these fatalities were observed in the resection with or without tapering (RA) cohort, and the patients predominantly originated from low-income countries or were reported in articles published in the early 1990s.

Zhu et al. [18] elucidate that when Apple-peel bowel atresia (APA) is compounded with prematurity and other malformations, the prognosis tends to be bleak. This finding is echoed by other studies [19], including our present review. From our series, it can be inferred that prematurity emerges as a significant risk factor for mortality, particularly when coupled with limited access to high-quality neonatal intensive care.

Expanding on this, Festen's work explicates how although the utilization of total parenteral nutrition (TPN) improves survival rates, especially in premature infants, it also carries certain risks. TPN can exacerbate the risk of mortality due to sepsis, primarily associated with central venous catheter infections, and increase the likelihood of developing Parenteral Nutrition-Associated Liver Disease (PNALD) when short bowel syndrome (SBS) is present.

In our review, we identified 12 cases of sepsis, although it is not confirmed whether it was related to central venous catheter infections. Additionally, short bowel syndrome (SBS) was described in seven cases. It appears that Apple-peel bowel atresia (APA) may be more closely associated with the development of SBS (74%), although there is no consensus among authors [3]

Interestingly, while it might be expected that resection with or without tapering (RA) would expose patients more to this complication, our review suggests that ostomy with delayed anastomosis (ODA) appears to be more associated with the development of SBS (33.3% of cases). However, it is important to note that there is

insufficient data in the available series to establish a statistical correlation between resection and SBS. Furthermore, it is essential to remember that the definition of SBS encompasses not only the length of the remaining bowel but also the functionality of the bowel itself [20].

Our review encountered several limitations. Firstly, we noted significant variability in outcomes across the reviewed articles. This variability can be attributed to considerable technical advancements over 30 years and the diverse standards of care across different geographic regions. Secondly, most of the articles lacked critical details, contributing to incomplete data. Thirdly, the majority of the articles consisted of case reports or case series, wherein the choice of intervention remained operator-dependent.

In this context, Hillyer [21] highlights a potential correlation between the surgeon's choice and the child's baseline condition. Specifically, the Apgar score appears to influence decision-making. Hillyer also suggests that ostomy could lead to inferior outcomes.

Due to these limitations, we can only consider the achievement of the first objective of the review. Consequently, our work cannot conclude the preferred technique or precise complications related to each type of intervention.

This review represents the first systematic exploration of Apple-peel intestinal atresia. After examining all proposed interventions, the authors argue that the literature primarily focuses on the condition of the proximal loop, overlooking the significance of the distal portion. The distal portion, with its atypical vascularization, warrants maximum attention. Moreover, as explained in several articles on fluid dynamics in anastomoses [20,21], the distal part plays a crucial role in bolus progression, emphasizing the importance of preserving vascularity. Interrupting bowel continuity with an ostomy may pose greater risks of complications due to the exclusion of the distal part, which does not contribute to bolus progression [22-23]..

CONCLUSION

This type of atresia is treated in the literature most widely with a resection. The review failed to assess which interventions could lead to faster full enteral feeding. The distal part of the anastomosis must be considered as an active part in the resumption of bowel function.

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Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs/material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Author Contributions: Author(s) declared to fulfil authorship criteria as devised by ICMJE and approved the final version.

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Table 3. Resection and Anastomosis (RA).

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35	NR			28			34		,	ა ა		NR			38		NR		31		34		Prematurity (WoP)
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66	available	Not		NR			20			NR		29			15				NA				TPN (days)
NR	achieved	V ot		40			24			NR		NR			39				Not achieved	31			FEFTA
No	No			No			No			No		Yes			No				No				Tapering
60		NR			NR			130		C+	ì		NR			NR					NR		RBL
adhesion- related bowel obstruction	fistula aspiration	Enterocutaneous		/			_		ħ	Septicemia		obstruction	1 Adhosin		/		loss weight	bowel, SBS	gangrenous distal	atresia	Misdiagnosed colic		Complications
Doing well	Died			Doing well			Doing well			Died		Doing well			Doing well		Died				Outcome		

27	26	25	24	23	22	21	19	17	16	14
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	NR	NA	0	NR	2	NR	NR	0	1	
	NR	NR	NR	NR	NR	NR	NR	8d	NR	210 32
	NR	NR	NR	NR	NR	32	NR	No	NR	
	No	No	No	No	No	No	No	No	No	
	NR	NR	6	NR	NR	NR	90 95	NR 80	12	35 120
/	Anastomotic leakage	/	sepsis	/	Intestinal and respiratory Infections, sepsis	/	/	Pneumonia	SBS	/
doing well doing well	death due to aspiration	Doing Well	Died	Doing well	death (SPINK5 mutation)	Doing Well	Doing Well	Death pneumoni a day 8	SBS	'

30 31 31 33 33 46 38-45 37 36 35 29 Sasa Montenegro 2016 CR Turnock UK 1991 CS Slee
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CR Shakya Nepal 2010 CS Vilella Spain 2014 CR NR NR NR ⋈ ⋈ Ħ ᅿ cystic intra-abdominal mass dilated stomach and small bowel Small bowel dilatation Not done Normal NA NA NRNR 33 32 31 34 29 NRNRNA 0 IJ 2 2 NRNRNR NRNRNR 18 NRNRNRNR NRNRNR Yes No No No N_0 No No NRNRNRNRNRNR10 2: intestinal obstruction and failure to feed died died death, cardioresp iratory failure Doing well Doing well Doing well Doing well 2 SBS2 died

WoP= weeks of pregnancy, RBL= Residual bowel length

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Yada Japan 2018 CR	Herman USA 2008 CR	Bellini Italy 2002 CR	2008 CS	Baglaj UK	Arbell Israel 2006 CR	CS	Akinola Nigeria 2011	Alnosair Saudi Arabia 2014 CR	Ahmad India 2009 CR	Authors Country Year Type of article
д	M	M		NR	M		ਸ	ħ	М	Sex
Dilatation of stomach and duodenum	Amniotic fluid stained	Bowel dilatation		NR	No screening		Double bubble sign	Polyhydramnios and double bubble sign	Polyhydramnios	Antenatal findings
31	36	37		NR	32		34	31	34	Prematurity (WoP)
4	NR	NR		NR	ĸ	,	4	NR	00	Days at surgery
NR	NR	NR		NR	21		No availab le	NR	NR	TPN (days)
NR	NR	NR		NR	21		NR	NR	NR	FEFTA
End to end	NR	End to end		NR	End to end		End to side	End to end	End to end	Type of anastomosis
Reverse anastomosis, re-laparotomy	/	/	/	Anastomotic strictures	/		/	/	Lost to follow up	Complications
Full enteral feeding	Normal oral feeding	Uneventful		Normal oral feeding	Normal feeding	rectal atresia	Death sepsis and associated	Normal feeding	Dead	Outcome

			65				64			63	61-				60				59				58			57	56
CR	2020	Bosnia	Zvizdic	CR	1999	Oman	Weber	CS	1997	USA	Waldhausen	CR	2007	Netherand	Van bever	CR	2007	Japan	Tatekawa	CR	1991	USA	Smith	CS	1993	Norway	Stromme
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polyhydramnios	the gut and	dilatation of	proximal				Double bubble				NR				Double bubble			duodenal atresia	Suspicion of			in twins	Abdominal mass				Normal
			34				36				NR				34				36				31			36	35
			2				ω				NR				2				NR				NR				NR
			39				7		16.3	+/-	31.4				NR				NR				NR				NR
			38				23				NR				NR				NR				NR				NR
			End d to end				End to end				NR				Side to side				Side to side				End to end				End to end
			_				/	dysmotility	leakage, 2	obstruction,	Anastomotic				_				/				/			/	SBS
			Doing well		syndrome	thrive, Down	Mild failure to				Doing well				Doing well		bowel)	cm of distal	Doing well (25		bowel)	cm of distal	Doing well (35				SBS

Table 5. Tapering and Anastomosis (TA)

109	108	107	106	105	66- 104	Pt. #
Kirtane India CR	Peetsold Netherlands 2004 CR	Rich USA 2013 CR	Onofre Brazil 2013 CR	Harper France 2009 CS	Zhu China CS	Authors Country Year Type of article
m	ਸ	ਸ	ਸ	ਸ	25F	Sex
Polyhydramnio s, 2 Apple peel in previous pregnancies	NR	Dilated bowel	NR	NR	Dilated bowel (20)	Antenatal findings
36	NR	37	35	35	30 preterm	Prematurity (WoP)
NR	NR	NR	3 m	66	37 d	TPN (days)
NR	NR	NR	NR	NR	NR	FEFTA
NR	NR	NR	20	60	76+/- 5	RBL
Bowel obstruction, Sepsis	Vomiting, late diagnosis of duodenal web	1	/	/	cholestasis (n = 20) electrolyte imbalances (n = 15), anemia (n = 10), sepsis (n = 8), adhesive obstruction (n = 4) and anastomotic obstruction (n = 3).	Complications
Doing well	Doing well	Doing well	Doing well	Doing well	Doing well	Outcome

Table 6. Ostomy and Delayed Anastomosis (ODA)

,	120	119	118	117	116	115	114		113	<u>.</u>	112			111			110			Pt. #	
		1997 CS	Waldhausen USA					CS	1990	Turnock			CR	Italy 2019	Digilio	CR	USA 2013	Bhalla	Type of article	Year	Authors
		į	N R						NR				NR			F				Sex	
		;	NA						NR				dilatation	nios, bowel	Polyhydram		Bowel obstruction			findings	
		;	NR						NR					35			37			(WoP)	D
		į	NR						NR					2			2			surgery	
		days	50						NR				NR			NR				(days)	MON
		ļ	NR						NR						NR			NR			FEFTA
Bishop Koop stoma	Bishop Koop stoma	Ostomy		Ostomy	Ostomy	Koop stoma	Bishon	double barrel	barrel	double	double		jejunostomy			jejunostomy				stoma	
		;	N R				•		NR						NR		TEP	89, 2S			RBL
		!	2 m						NR						1m			2 m		SCOTIO	Closure
/			occlusions			\		/	functioning bowel	SBS, resection of poorly	poorly functioning bowel	SBS, resection of		/			Dumping, syndrome			Complication	
		0	Doing well				•		2 SBS					Doing well			Doing well			Outcome	

125	124	123	122	121
Federici Italy 2003 CR	Miraglia Italy 2010 CR	Filipa Portugal 2019 CR	Broekaert Germany 2014 CR	Harper France 2009 CS
×	ੰਸ	দ	ਸ	M
Upper small bowel obstruction	NR	dilated stomach and small bowel	Polyhydram nios	NA
33	34	36	38	35
NR	2	30	NR	NR
49	on going	40	NR	127
NR	NR	40	NR	31
double	NR	double jejunostomy	jejunostomy	double enterostom y
80	23	NR	110	100
53d	NR	NR	6 w	2 m
	Anastomotic stenosis		Cystic fibrosis	Anastomotic stenosis
Doing well	Doing well	Doing well	Doing well	Doing well