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Management and Outcomes of Jejunoileal Atresia Within the United Kingdom

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ABSTRACT

Background: Jejunoileal atresia (JIA) is seen in 0.7 per 10,000 births and requires early surgical intervention to restore gastrointestinal continuity. Many intra-operative techniques exist to manage the atresia depending on anatomy encountered and proximal bowel dilatation. Existing studies are predominantly single centre experiences. This study aims to report contemporary management and outcomes of JIA in the United Kingdom (UK) and explore how operative technique is associated with outcome.

Methods: Multicentre UK based observational study over 5 years with follow-up to 1 year post surgery. Outcomes were time to full enteral feeds, length of stay, unplanned reoperation, number of general anaesthetics, development of short bowel syndrome and mortality. These outcomes were stratified by infant related, disease related and surgical technique related factors.

Results: There were 159 infants with JIA from seven tertiary paediatric surgical units. JIA was suspected antenatally in 92 (57.8 %) infants and associated congenital anomalies were seen in 47 (29.6 %) infants. Age at surgery was 2 (0–70) days and primary anastomosis was undertaken in 114 (71.7 %) procedures most commonly (87.9 %) via end-to-end anastomosis. Primary anastomosis was associated with shorter duration to full enteral feeds (21 vs 60 days, $p = 0.001$), shorter length of stay (28 vs 78 days, $p < 0.001$), fewer general anaesthetics (1 vs 3, $p < 0.001$) and lower mortality (0.9 vs 11.1 %, $p = 0.002$) than enterostomy formation. Overall, unplanned reoperation was required in 27 (17.0 %) infants primarily for intestinal obstruction ($n = 21/27$).

Conclusions: These data provide contemporary outcomes by atresia type and highlight obstructive complications are frequent in this cohort. These data support primary anastomosis in JIA.

1. Introduction

Jejunoileal atresia (JIA) is seen in 0.7 per 10,000 births in Europe and more recently the majority are suspected antenatally [1]. Various surgical options exist for JIA which are determined by infant and anatomical factors. After deciding between either primary anastomosis or enterostomy, further decisions are required regarding type of enterostomy or method of anastomosis. Methods

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of enterostomy formation include double barrelled, Bishop-Koop [2], Santulli [3] and Mikulicz [4] techniques. If a primary anastomosis is undertaken management of proximal to distal width discrepancy is often required which can involve tapering [5], plication [6] or "Fish-mouthing" (longitudinal enterotomy of distal bowel to increase width) and anastomoses can be end-to-end [7], end-to-side [8] or side-to-side. Decision making can be more challenging if multiple atresias are found.

Reports of which of these methods are used and outcomes of JIA related to these techniques are limited mainly to single centre studies [5,9–12]. One recent national study of JIA from Germany used administrative level data and reported significant post operative morbidity in 16.3 % of infants, a median length of stay of 66 days and death occurred in 5.1 % of infants [13]. It was not possible in this study to report these outcome stratified by anatomical type of JIA or surgical technique. This study aimed to report contemporary multicentre management of JIA within the United Kingdom (UK) and identify how patient characteristics, disease classification and surgical techniques are associated with important outcomes.

2. Methods

2.1. Case identification

Data were retrospectively collected from seven tertiary paediatric surgical units in the UK including all infants with a diagnosis at surgery of JIA who were born over a 5 year period from January 2016 to December 2020. Those with isolated duodenal or colonic atresia without JIA were excluded. The minimum follow-up duration was 1 year. Anatomical type of JIA was classified according to Grosfeld [14].

2.2. Outcomes

Outcomes of interest were defined in the study protocol and were time to full enteral feeds, duration of initial inpatient stay, unplanned reoperation within 1 year, number of general anaesthetics within 1 year for reasons related to JIA (i.e. laparotomy or central venous access for parenteral nutrition), development of short bowel syndrome and mortality at 1 year. Short bowel syndrome was defined as requirement for home parenteral nutrition. Data were analysed to identify infant related, disease related and surgical technique related associations with these outcomes.

2.3. Statistical analysis

Continuous data were compared using a Mann Whitney U test or a Kruskal–Wallis test as appropriate and reported as median with range. Categorical data were compared using a Chi squared test and reported as absolute number with percentage. Statistical analyses were undertaken using StataSE v18 (StataCorp LLC, Texas, USA).

2.4. Ethical considerations

This study was classified as a service evaluation as per Health Research Authority (HRA) guidance and was therefore registered as such at each participating centre.

3. Results

3.1. Patient population

There were 159 infants included in this study of which 84 (52.8 %) were male. Gestational age at birth was 36.3 (23.0–42.0) weeks and birthweight was 2.70 (0.57–4.55) kg. Associated congenital anomalies were present in 47 (29.6 %) infants and were gastrointestinal (n = 22 [13.8 %] – see Table S1 for further detail), cardiac (n = 21 [13.2 %]), cystic fibrosis (n = 9 [5.6 %]) and chromosomal (n = 7 [4.4 %]). Some infants had multiple associated anomalies. Infants with associated anomalies had a lower gestational age at birth, lower birthweight, less proximal to distal bowel width discrepancy and were more likely to have necrotic bowel than those without (Table S2).

3.2. Pre-operative management

There was antenatal suspicion of JIA in 92 (57.8 %) infants and diagnostic contrast radiological imaging was undertaken in 59 (37.1 %) infants. Modality of contrast study, where available was upper gastrointestinal (GI) tract (n = 19), lower GI tract (n = 37) and both upper and lower GI tract (n = 2).

3.3. Operative findings and management

Age at surgery was 2 (0–70) days. Meconium peritonitis was found in 19 (12.0 %) infants. Distance from the duodenal–jejunal junction to the atresia (first atresia if multiple) was 32 (1–120) cm and classification of JIA, where recorded, was type I (n = 31 [21.5 %]), type II (n = 16 [32.6 %]), type IIIa (n = 31 [21.5 %]), type IIIb (n = 25 [17.4 %]) and type IV (n = 41 [28.5 %]) (Table 1). There was proximal necrosis or perforation, due to significant dilatation, in 32 (20.1 %) infants and a median of 10 (2–44) cm of bowel required resection. A Ladd's procedure due to associated of midgut malrotation was undertaken in 17 (10.7 %) infants.

Primary anastomosis was undertaken in 114 (71.7 %) procedures, anatomical configuration of these were predominately end-to-end (n = 73) (Table S3) but side-to-side (n = 7), end-to-side (n = 2) and jejunoplasty (n = 1) were also used. The median bowel width discrepancy ratio, between proximal and distal anastomosis, was 4 [1–10] to 1. Recorded strategies to manage discrepancy were fish-mouthing (n = 34), tapering (n = 4) and plication (n = 1) whilst this wasn't required in 65 infants and in some, multiple techniques were used. Those that underwent one of these methods of discrepancy management had a higher proximal to distal

Table 1
Demographics and disease characteristics by atresia type.

	I (n = 31)	II (n = 16)	IIIa (n = 31)	IIIb (n = 25)	IV (n = 41)	p
Male, n	14 (45.2)	9 (56.3)	19 (61.3)	11 (44.0)	25 (61.0)	0.48
Gestational age at birth, weeks	36.6 (26.6–40.1)	36.3 (23.0–40.0)	37.0 (25.0–42.0)	36.0 (27.6–38.4)	36.0 (31.0–38.4)	**0.04**
Birthweight, kg	2.82 (0.67–3.95)	2.77 (0.59–4.00)	2.90 (0.57–4.16)	2.46 (0.61–3.32)	2.68 (0.94–4.55)	0.11
Associated anomalies, n	5 (16.1)	6 (37.5)	8 (25.8)	10 (40.0)	12 (29.3)	0.31
Distance from DJ, cm	50 (5–120)	48 (20–80)	49 (1–120)	23 (1–90)	20 (2–90)	0.12
Discrepancy ratio, n:1	4 (1–9)	4 (3–10)	4 (1.5–6)	4 (2–10)	5 (2–10)	0.36
Proximal necrosis or perforation, n	4 (12.9)	5 (31.3)	12 (38.7)	5 (20.0)	2 (4.9)	**0.004**

kg - kilograms, DJ – duodenal–jejunal junction, cm – centre metres. Data are n (%) or median (range). Statistically significant p values are in bold.

discrepancy ratio than those that didn't undergo one of the three techniques (5 [1.5–10]:1 *vs.* 4 [1–10]:1, p = 0.007). A trans-anastomotic tube was placed in 4 infants.

Enterostomy formation was undertaken in 45 (28.3 %) procedures and was utilised more often in infants with lower birthweight, lower gestational age, presence of associated anomalies and findings of necrotic proximal bowel at laparotomy (Table 2). Enterostomy type was unspecified enterostomy (n = 35), Mikulicz (n = 6) and tube (n = 4).

3.4. Outcomes

Time to achieve full enteral feeds was 27 (1–397) days and was significantly slower in those with type IV JIA (Table 3), those with enterostomy (Table 4) and those with associated anomalies (Table 4). Length of stay was 31 (6–399) days and was greater in those with type IV JIA (Table 3), those with enterostomy (Table 4), those with an end-to-end versus other type of anastomosis (Table 4) and those with associated anomalies (Table 4). Unplanned reoperation was required in 27 (17.0 %) infants for intestinal obstruction (n = 21), stoma complications (n = 2), bowel necrosis (n = 2), bowel lengthening (n = 1) and intra-abdominal abscess (n = 1). Where further detail was provided (12/21), intestinal obstruction was due to either anastomotic stricture (n = 8) or intra-abdominal adhesions (n = 4) Those that underwent unplanned reoperation achieved full enteral feeds later (61 vs 21 days, p < 0.001), had a longer length of stay (116 vs 29 days, p < 0.001) and more frequently developed short bowel syndrome (37 vs 5 %, p < 0.001) than those without a reoperation. Furthermore, 13/27 (48 %) infants went on to have a further unplanned reoperation.

The total number of general anaesthetics was 1 [1–13]. Significantly more general anaesthetics were required in those with type IIIb or IV JIA (Table 3), those with enterostomy (Table 4), those with an anastomosis which wasn't end-to-end (Table 4) and those with associated anomalies (Table 4). Short bowel syndrome was seen in 17 (10.7 %) infants and occurred more frequently in those with a proximal to distal bowel discrepancy ratio of more than 5:1 (Table S4 and Table 4) and with presence of associated anomalies (Table 4). In those that were alive at 1 year of age, 146/154 (94.8 %) had achieved full enteral autonomy. Death occurred in 6 (3.8 %) infants at 97 (6–629) days of age. Death occurred more frequently in those with an enterostomy versus primary anastomosis (Table 4).

Table 2
Demographics and disease characteristics comparing primary anastomosis versus enterostomy.

		Primary anastomosis (n = 114)	Enterostomy (n = 45)	p
Male, n		59 (51.8)	25 (55.6)	0.67
Gestational age at birth, weeks		36.8 (27.6–42.0)	35.0 (23.0–40.0)	**0.001**
Birthweight, kg		2.78 (0.61–4.55)	2.28 (0.57–3.80)	**<0.001**
Associated anomalies, n		21 (18.4)	26 (57.8)	**<0.001**
Atresia type	**I**	23 (22.1)	8 (20.0)	0.66
	II	9 (8.7)	7 (17.5)	
	IIIa	23 (22.1)	8 (20.0)	
	IIIb	18 (17.3)	7 (17.5)	
	IV	31 (29.8)	10 (25.0)	
Distance from DJ, cm		28 (1–120)	50 (1–120)	**0.02**
Discrepancy ratio, n:1		4 (1–10)	4 (2–8)	0.43
Proximal necrosis or perforation, n		14 (12.3)	18 (40.0)	**<0.001**

kg - kilograms, DJ – duodenal–jejunal junction, cm – centre metres. Data are n (%) or median (range). Statistically significant p values are in bold.

Table 3
Outcomes by atresia type.

	I (n = 31)	II (n = 16)	IIIa (n = 31)	IIIb (n = 25)	IV (n = 41)	p
Time to full enteral feeds, days	16 (4–210)	23 (1–130)	16 (5–74)	29 (7–397)	42 (6–265)	**0.002**
Length of stay, days	27 (7–377)	28 (6–383)	24 (9–257)	34 (6–399)	50 (9–280)	**0.02**
Reoperation required, n	4 (12.9)	3 (18.8)	1 (3.2)	3 (12.0)	11 (26.8)	0.09
General anaesthetics, total	1 (1–6)	1 (1–6)	1 (1–4)	2 (1–8)	2 (1–13)	**0.04**
Short bowel syndrome, n	0 (0)	1 (6.3)	3 (9.7)	3 (12.0)	7 (17.1)	0.18
Death, n	1 (3.2)	1 (6.3)	1 (3.2)	1 (4.0)	0 (0)	0.72

Data are n (%) or median (range). Statistically significant p values are in bold.

Table 4
Outcomes comparing those with and without associated anomalies, primary anastomosis versus enterostomy, anastomosis type and proximal to distal bowel width ratio at anastomosis.

	Associated anomalies			Primary anastomosis			Anastomosis type[a]			Proximal to distal bowel width[b]		
	Present (n = 47)	Absent (n = 112)	p	Yes (n = 114)	No (n = 45)	p	End-to-end (n = 73)	Other (n = 10)	p	Ratio <5:1 (n = 47)	Ratio >5:1 (n = 35)	p
Time to full enteral feeds, days	51 (7–397)	21 (1–211)	**<0.001**	21 (1–397)	60 (5–210)	**0.001**	28 (1–397)	14 (5–211)	0.11	18 (1–210)	28 (3–136)	0.24
Length of stay, days	94 (9–399)	28 (6–215)	**<0.001**	28 (6–399)	78 (6–383)	**<0.001**	32 (7–399)	12 (9–120)	**0.01**	22 (7–210)	31 (6–320)	0.15
Reoperation required, n	12 (25.5)	15 (13.4)	0.06	20 (17.5)	7 (15.6)	0.76	15 (20.6)	4 (40.0)	0.17	6 (10.6)	9 (25.7)	0.07
General anaesthetics, total	3 (1–13)	1 (1–5)	**<0.001**	1 (1–13)	3 (1–7)	**<0.001**	1 (1–13)	2 (1–8)	**0.04**	1 (1–6)	1 (1–8)	0.44
Short bowel syndrome, n	9 (19.2)	8 (7.1)	**0.03**	12 (10.5)	5 (11.1)	0.91	7 (9.6)	3 (30.0)	0.06	0 (0)	4 (11.4)	**0.02**
Death, n	3 (6.4)	3 (2.7)	0.26	1 (0.9)	5 (11.1)	**0.002**	1 (1.4)	0 (0.0)	0.71	1 (2.1)	1 (2.9)	0.83

Data are n (%) or median (range). Statistically significant p values are in bold.
[a] Only those that underwent primary anastomosis and anastomosis type available.
[b] Only those that underwent primary anastomosis and anastomosis discrepancy ratio available.

Cause of death was sepsis (n = 3), redirection of care due to short bowel syndrome (n = 1), multiple comorbidities (n = 1) and complications of liver and bowel transplant (n = 1).

4. Discussion

This is the first multicentre UK based study to report management and outcomes of JIA. We found that most commonly an end-to-end primary anastomosis is undertaken and the most common method for managing bowel width discrepancy is fish-mouthing of the distal bowel. Unplanned reoperation, excluding enterostomy reversal, was seen in just under 1 in 5 infants with JIA. This was as high as 26.8 % in those with multiple atresias highlighting this group as particularly high risk.

Primary anastomosis, rather than enterostomy formation, was associated with reduced time to full enteral feeds and shorter length of stay without an increase in unplanned reoperation. There are however situations including presence of significant necrotic bowel or very short bowel where enterostomy formation is more appropriate. A recent study from the United States found that the most common reason for enterostomy formation in JIA was significant discrepancy between proximal and distal bowel [15]. We found that those who underwent enterostomy formation were younger, smaller, and more frequently had associated anomalies and necrotic bowel than those where primary anastomosis was undertaken. The primary anastomosis rate of 72 % in this study is similar to that reported by others [12,16]. Interestingly, the length of bowel from duodenal-jejunal flexure to the atresia was shorter in those that underwent primary anastomosis suggesting that surgeons balance the risk of an anastomotic complication versus the risk of a high output enterostomy during intra-operative decision making. Additionally, it is known from previous study that stoma related complications are seen in up to 41 % of infants and are known to be associated with poor growth [17,18].

When anastomosis was undertaken this was most commonly in an end-to-end configuration. Bowel width discrepancy was most frequently managed with fish-mouthing of the distal bowel. Knowledge and experience of the various techniques to manage this are important and should be tailored to the individual anatomical configuration encountered in each infant with JIA.

The requirement for unplanned reoperation was moderately high in this cohort but similar rates have been reported by others [7,9,10,19]. This rate is also higher than more proximal atresias as an UK population based study found that only 6 % of those with congenital duodenal obstruction required unplanned reoperation within the same length of follow-up [20]. Most of the unplanned reoperations were required for intestinal obstruction, either due to adhesions or anastomotic stricture. Whilst addressing the former cause is challenging, the latter highlights the importance of effective proximal to distal discrepancy management either at primary anastomosis or enterostomy reversal. This is an important area for future research as it is possible that some configurations of anastomosis are more prone to obstruction than others.

This study was limited by its retrospective nature however provides a multicentre perspective with detailed information regarding intra-operative findings and management strategies. Infants with JIA represent a heterogenous population hence other factors may influence the reported outcomes not measured in this study and we were unable to utilise multivariable regression in analyses due to modest group size of each JIA type. Outcomes are however stratified by anatomical classification, presence of associated anomalies and intra-operative management strategy to allow understanding of expected outcomes in future infants and potential for surgeons to compare their outcomes to a contemporary, multicentre cohort. Outcomes in this study are reported to one year however this study will have missed morbidity that happens beyond infancy. A previous study that did measure morbidity up to 19 years age found that quality of life was generally good [19]. Those with associated gastrointestinal anomalies did however have lower gastrointestinal symptom scores and almost 40 % of those with follow-up reported embarrassment associated with their surgical scar [19].

This multicentre observational study has reported contemporary management and outcomes of JIA in the UK across several centres. These data support primary anastomosis in JIA if the anatomical configuration encountered allows this however there will always be infants where enterostomy formation is required. Expected outcomes stratified by JIA type, presence of associated anomalies and common management strategies may be useful to both surgeons and families of future infants with JIA.

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Conflict of interest

None declared.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2025.162334.

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