



## Gastrointestinal surgery in cystic fibrosis: A 20-year review

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### ABSTRACT

**Objectives:** The purpose of this study was to evaluate outcomes of the surgical management for meconium ileus (MI) and Distal Intestinal Obstruction Syndrome (DIOS) in Cystic Fibrosis (CF).

**Methods:** Children born between 1990 and 2010 were identified using a regional CF database. Retrospective case note analysis was performed. Outcome measures for MI were mortality, relaparotomy rate, length of stay (LOS), time on parental nutrition (TP), and time to full feeds (TFF). Outcome measures for DIOS were: age of onset, number of episodes, and need for laparotomy.

**Results:** Seventy-five of 376 neonates presented with MI. Fifty-four (92%) required laparotomy. Contrast enema decompression was attempted in nineteen. There were no post-operative deaths. Thirty-nine (72%) neonates with MI were managed with stomas. LOS was longer in those managed with stomas ( $p = 0.001$ ) and in complex MI ( $p = 0.002$ ). Thirty-five patients were treated for DIOS. Twenty-five patients were managed with gastrografin. Ten patients underwent surgical management of DIOS. Overall, MI did not predispose to later development of DIOS. There was a significantly greater incidence of laparotomy for DIOS in children who had MI.

**Conclusion:** The proportion of neonates with complex meconium ileus was high (49%) and may explain the infrequent utilisation of radiological decompression. Complex MI or management with stomas both significantly increase LOS. Re-laparotomy rate is high (22%) in MI irrespective of the type of management. DIOS is not a benign condition, particularly when the child has had previous abdominal surgery. Early referral to a surgical team is essential in these children.

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Meconium ileus (MI) may be the presenting feature in 10%–15% of patients with cystic fibrosis (CF) [1,2] and can be classified as simple or complex. Simple MI is an uncomplicated intraluminal distal small bowel obstruction due to abnormally viscid meconium. Complex MI additionally can be characterized by complications such as meconium peritonitis, pseudocyst formation, volvulus or intestinal atresia.

A variety of operative strategies have been utilized in the management of both simple and complicated MI [1–4,6], the ultimate aim being to decompress the bowel and relieve obstruction by clearing the abnormal meconium. Strategies can be broadly classified as those employing the formation of a stoma (e.g. divided, Bishop–Koop or Santulli), and those where bowel continuity is maintained (e.g. enterotomy and washout, with or without intestinal resection and primary anastomosis). There is no consensus in the current literature as to what constitutes the preferred surgical strategy for the management of MI.

Partial or complete small bowel obstruction attributed to CF has also been described since 1945 in older children [8]. This was initially termed

meconium ileus equivalent (MIE) but more recently the term distal intestinal obstruction syndrome (DIOS) has become commonplace. The term DIOS came to encompass a variety of intestinal symptoms and signs, possibly including recalcitrant constipation. The varied definitions have made it difficult to estimate the true incidence and morbidity associated with DIOS [5,7–9]. However, in 2010 the ESPGHAN CF Working group introduced a clear and specific definition for the term DIOS [8] (Table 1). Since then there has been very little contemporary literature on the morbidity and surgical management of DIOS [7].

Our aim was to evaluate the success of managing meconium ileus and DIOS using different surgical strategies over a 20-year period at a single institution. We were specifically interested as to whether infants with MI managed with stomas were comparable to those where intestinal continuity was maintained. The utilization and outcome of radiological decompression in MI were also investigated. For the group with DIOS the principle aim was to test the hypothesis that MI predisposes to the later development of DIOS overall and for DIOS requiring surgery.

### 1. Methods

Patients were retrospectively identified using the regional CF database. All patients under the care of the Alder Hey Children's NHS

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**Table 1**  
ESPGHAN CF Working Group definition for DIOS in cystic fibrosis [8].

Criteria	Explanation
1	Complete intestinal obstruction as evidenced by vomiting of bilious material and/or fluid levels in small intestine on an abdominal radiography
2	Faecal mass in ileo-caecum
3	Abdominal pain and/or distension
Complete DIOS: 1 and 2 and 3	
Incomplete/Impending DIOS: 2 and 3 only	

Foundation Trust with CF who were born between 1990 and 2010 were eligible for inclusion.

Patients requiring management of MI or DIOS were identified from the CF database and their hospital case notes retrieved.

Data were collected for each patient with MI with regard to; conservative versus operative management and stoma versus other operative procedure. Outcome measures included mortality, re-laparotomy (excluding restoration of continuity), time to restoration of continuity, length of stay (LOS), time on parenteral nutrition (TP) and time to full feeds (TFF).

DIOS was defined using ESPGHAN criteria of abdominal pain +/- distension and ileocaecal mass with evidence of obstruction (Table 1) [8]. Case notes and radiological investigations were rigorously reviewed and children were only included when they met all the criteria for the contemporary definition of "complete DIOS". Data for this group included age of onset, number of episodes, history and type of preceding MI, and need for previous surgery.

Data are quoted as median (range). The Mann-Whitney U-test and the Fisher exact 2-tailed test were used for statistical analysis. P values of <0.05 were considered significant.

## 2. Results

The regional database identified 376 children with CF born between Jan. 1990 and Dec. 2010. However complete data were only available for the 146 patients that were managed and followed-up at Alder Hey Children's NHS Foundation Trust. The remaining patients predominantly attended district general hospitals for the subsequent management of their CF.

Seventy-five (19.9%) of 376 children presented as infants with MI. Complete data sets were available for only 59 as 16 were subsequently transferred back to a more local specialist centre as infants or relocated later and were not followed up at Alder Hey hospital. These were excluded from this analysis. Thirty (51%) infants were regarded as having simple MI and 29 (49%) had complicated MI. Gastrografin® enemas were attempted initially in 19 (32%) infants but in only 5 (9%) was this successful and the remaining 54 (91%) required laparotomy.

Of the 54 patients requiring laparotomy 39 (72%) had divided stomas formed, 7 (13%) were managed with enterotomy and washout, and 8 (15%) with resection and primary anastomosis. The

time to closure for patients managed with stomas was 47 days (4–202).

All neonates with meconium ileus were given *N*-acetyl cysteine in the post-operative period. It is administered through the feeding tube or by lavage of the proximal stoma to aid resolution of obstructive symptoms. Once enteral feeding is established the *N*-acetyl cysteine is replaced by pancreatic enzyme replacement therapy. Enzyme replacement therapy is initiated once infants have reached a volume of 30 ml per feed.

There was a significant difference in LOS after MI between operative patients managed with and without stomas: 49 days (19–390) and 23 days (15–60) respectively.

Table 2 shows that those managed with stomas tended to take longer to reach full feeds compared with neonates where bowel continuity was maintained after laparotomy (25 days (7–148) versus 9 days (5–49)), however the difference failed to reach statistical significance ( $p = 0.067$ ). There was also a trend towards a reduced duration of parental nutrition in the group managed without stomas (18 days (5–104) versus 37 days (1–148)) that failed to reach statistical significance ( $p0.062$ ). Infants tended to stay on supplemental parenteral nutrition for a period of time after establishing appropriate full volume enteral feeds. Parenteral nutrition was discontinued once it was demonstrated that growth was adequate on serial weight measurements.

Infants with complex MI had a longer LOS than those with simple MI [26.5 (6–390) days versus 52 (15–151) days;  $P < 0.002$ ] (Table 2). There was no difference in the re-laparotomy rate between infants managed with or without stomas [8 of 36 (22%) versus 3 of 13 (23%);  $P = 1.00$ ]. Data regarding re-laparotomy were incomplete for 5 patients. Although surgical management with stomas increased length of stay [49 (19–390) days vs 23.5 (15–60) days;  $P = 0.001$ ], it did not increase re-laparotomy rates [22% versus 23%;  $P = 1.00$ ]. Median follow-up in this group was 118 (12–225) months. There were no deaths in the post-operative neonatal period. There were 3 late deaths due to non-gastrointestinal manifestations of CF.

The number of CF patients treated for DIOS was 35 (24%). The age of onset of the first episode of DIOS was 7.7 (0.9–16.7) years. Of these, 22 (62%) were managed with oral Gastrografin and 3 (9%) with Gastrografin enemas. The other 10 (29%) had DIOS requiring laparotomy. In total there were 80 separate episodes of "complete DIOS" in these patients.

Presentation with MI did not predispose to later development of DIOS [ $n = 20$  (29%) versus  $n = 15$  (19%);  $P = 0.24$ ] (Table 3). However, there was a greater incidence of surgical management of DIOS in children who had had MI [ $n = 10$  (50%) versus  $n = 0$  (0%);  $P = 0.002$ ]. Laparotomy was never required for the management of DIOS in children who had not previously had MI (Table 3). Subgroup analysis showed that neither complex MI ( $P = 1$ ) nor management with stomas ( $P = 0.35$ ) predisposed to surgical management of DIOS. Of the children that required laparotomy for DIOS, 5 (50%) had simple meconium ileus and 5 (50%) had complex meconium ileus (Table 3).

**Table 2**  
Outcomes in Meconium Ileus (MI).

	Type of Surgery			Type of MI		
	Stoma $n = 39$	No Stoma $n = 15$	P value	Simple MI $n = 30$	Complex MI $n = 29$	P value
LOS (days)	49 <sup>a</sup> (19–390)	23.5 (15–60)	0.001	26.5 (6–390)	52 (15–151)	0.002
TP (days)	37 (1–148)	18 (5–104)	0.062	19.5 (5–75)	32.5 (1–148)	0.14
TFF (days)	25 (7–148)	9 (5–49)	0.067	19 (5–75)	21 (5–148)	0.34

LOS: length of stay, TP: time on parenteral nutrition, TFF: time to full enteral feeds.

<sup>a</sup> All values are median (range)

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