MANAGEMENT AND EARLY OUTCOMES OF MECONIUM ILEUS ASSOCIATED WITH CYSTIC FIBROSIS IN THE UNITED KINGDOM AND IRELAND; A PROSPECTIVE POPULATION COHORT STUDY

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Aim of the study: The aim of this study was to assess the incidence, current management strategies and early outcomes of Meconium Ileus (MI) in association with Cystic Fibrosis (CF), in the UK.

Methods: This was a prospective, multicentre population cohort study utilising an established reporting system to identify new cases of MI associated with CF presenting between October 2012 and September 2014. Data were collected on antenatal and birth history, presentation, management and early outcomes of all reported cases.

Main Results: 72 cases were reported, of which 51 (71%) have had the diagnosis of CF confirmed. Analysis of management and outcomes was undertaken on these 51 infants. 12 cases (24%) were considered ‘high risk’ for CF on antenatal screening. Additional congenital anomalies were found in 4 patients (8%).

A therapeutic contrast enema was performed in 36 infants (71%). 52 enemas were performed in total, (1-4 per patient). Two patients, (6%) had complications of enema treatment; one had a perforation detected the day after the procedure, another developed hyponatraemia following each of two enemas.

40 infants underwent laparotomy (79%). In 13(26%), this was a primary procedure; 27 were undertaken after contrast enema. At laparotomy, meconium obstruction was seen in 51%, perforation in 22%, atresia in 14% and 30% had evidence of volvulus. 35% underwent an enterotomy and irrigation, 10% had a resection with primary anastomosis and 24% had a bowel resection and enterostomy.

Two deaths were recorded within the cohort (4%). One had a contrast but no surgery and the other died several weeks post operatively.

Conclusions: These data provide a contemporary picture of the incidence of MI in association with CF in the UK, its clinical features and early management. The majority of patients required laparotomy despite the widespread and sometimes repeated use of therapeutic contrast enema (Fig 1).

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Cases n=51

No intervention required 2(4%)

Lower GI Contrast 36(71%)

Surgery as primary Rx 13(26%)

Laprotomy not required n=9

No Surgery 11(22%)

Laprotomy required n=27

Surgery 40 (78%)
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