Diagnostic laparoscopy to exclude malrotation following inconclusive upper gastrointestinal contrast study in infants

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**Abstract**

Purpose:

We report the use of diagnostic laparoscopy as an alternative to laparotomy in the investigation of infants with clinical features concerning for malrotation and inconclusive upper gastrointestinal contrast study.

Method:

Casenote review of all infants in whom laparoscopy was performed during 2016-2020 to investigate for possible malrotation.

Results:

Eight infants were identified. All presented with acute clinical features of malrotation (bilious vomit) without evidence of an alternate explanatory diagnosis. All underwent upper gastrointestinal contrast study, with three also undergoing abdominal ultrasound. The radiological examinations could not exclude malrotation and all proceeded to laparoscopy.

At laparoscopy the small intestine was run to exclude the presence of midgut volvulus. In six cases normal rotation was confirmed and no abnormal pathology found. Two proceeded to laparotomy and underwent correction of malrotation. All infants recovered without complication.

Conclusion:

Laparoscopy is an excellent modality for further investigation of infants presenting acutely in whom intestinal malrotation cannot be formally excluded radiologically. The positive identification of the DJ flexure and caecum in correct anatomical sites, both fixed to the posterior abdominal wall, provides adequate reassurance of low risk of volvulus and avoids a full laparotomy. We recommend diagnostic laparoscopy in cases of inconclusive upper gastrointestinal contrast study.

## Introduction

Midgut malrotation, with its associated risk of midgut volvulus, is a congenital rotational anomaly resulting in abnormal orientation and fixation of the midgut within the abdominal cavity [1]. The incidence of malrotation is estimated at 1 in 500 births. The importance of this anomaly is the significant morbidity and mortality that may result from midgut volvulus including need for emergency surgery, intestinal resection, short bowel syndrome and in some cases death.

The most common mode of presentation of intestinal malrotation is an infant presenting with bilious vomiting (95%), abdominal distention with associated tachycardia and shock are late signs [2] and should warrant urgent surgical exploration. The gold standard investigation for an infant suspected to have malrotation in the early stages, when bile vomiting is potentially the only indication of the underlying pathology is an upper gastrointestinal (UGI) contrast study. UGI contrast study is considered abnormal when the duodenojejunal (DJ) flexure is abnormally positioned in the midline or to the right of the left-sided pedicles of the vertebral body, or below the transpyloric plane [3]. It is estimated that 70% of cases of malrotation have this characteristic finding. The sensitivity of the UGI contrast study at detecting malrotation is reported as high as 96% [4]. The presence of a normally sited DJ flexure is therefore reassuring that the infant does not have malrotation, whereas in the presence of an abnormal study most surgeons would undertake a laparotomy.

Controversy arises when the UGI contrast study is equivocal. The paediatric surgeon seeks certainty in such a situation, wishing to be sure that the infant categorically does not have malrotation, yet the gold standard diagnostic test has not provided this security. The use of ultrasonography (US) in experienced hands has been described to aid in the investigation and diagnosis of malrotation [5 - 7]. Malrotation is suspected when the orientation of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) is abnormal, with the greatest sensitivity in cases of volvulus when the SMV is coiling round the SMA (so-called whirlpool sign). However, if vessel orientation is normal one still cannot rule out malrotation. Whilst contrast enema may be helpful in assessing placement of the caecum, again a normally sited caecum does not exclude malrotation.

Historically, infants with an equivocal or inconclusive UGI contrast study in the presence of bile vomiting and no other cause confirmed would proceed to diagnostic laparotomy to achieve that desired certainty. In the interest of achieving this diagnostic certainty through a less invasive approach we have used diagnostic laparoscopy in this group of infants and here report our experience.

## Method:

We performed a retrospective case note review at a single tertiary neonatal surgical referral centre from 2016 to 2020. All infants (<1 year) who underwent a UGI contrast for investigation of possible malrotation after presenting acutely with bile stained vomiting were identified from hospital radiology and neonatal databases. Our standard protocol for investigation of an infant with bile stained vomiting comprises assessment through history, examination, abdominal radiograph and assessment for other conditions which may present with bile stained vomit. If no causative pathology is found and malrotation with or without volvulus is considered a possibility we proceed an UGI contrast study. Only infants who underwent diagnostic laparoscopy following an inconclusive UGI contrast study were included. Any infant who had elective investigation of possible malrotation (for example during work-up for poor feed tolerance , failure to thrive or isomerism) were excluded. Similarly infants who presented with bile stained vomiting with associated acute abdomen and were clinically unstable were excluded since these patients proceeded to emergency surgery. Demographic, clinical and outcome data were recorded and are reported descriptively as median (range). This study was registered and approved in our institution as a service evaluation.

Diagnostic laparoscopy was performed using a 5mm umbilical port for insertion of a 5mm 30 degree laparoscope and two 3 mm stab incisions for 3mm bowel grasping laparoscopic instruments. Pneumoperitoneum was established at 6-8mmHg. Under laparoscopic vision the caecum was identified and its position and degree of fixation noted. The small intestine was run proximally to the DJ flexure at the ligament of Treitz. The position of this was noted in relation to infant’s midline by applying pressure externally in the midline in the epigastric region and visually inspecting the position of the resulting bulge that appeared on the internal surface of the anterior abdominal wall. The position of the proximal duodenum passing behind the root of the mesentery in a fixed retroperitoneal position was noted and an overall assessment was made of the width of the mesentery between these two points (DJ flexure and caecum). The rotation was considered to be normal or not in need of intervention if the DJ flexure was to the left of the infant’s midline and in a fixed position with the proximal duodenum passing behind the root of the mesentery, the caecum fixed in the right lower quadrant, and the mesenteric root wide. In the presence of a mobile caecum but the other two features being within acceptable limits, particular assessment was made of the width of the root of the mesentery and a judgement made as to whether the situation would be improved (and hence the risk of volvulus reduced) by performing a Ladd’s procedure.

## Results

*Patient characteristics*

A total of 8 infants underwent a diagnostic laparoscopy. Mean gestational age at birth was 40 weeks (range 38-42weeks) and birth weight 3965kg (3030-4910kg). Six presented with a bilious vomit, unremarkable clinical examination and normal blood work-up during the first few days of life; one presented with bile vomiting in association with a distended abdomen at a month of age and one infant presented with a bile stained vomit and dehydration on a background of more chronic non-bilious vomiting and failure to thrive at 70 days of age. Following plain abdominal radiography all proceeded to UGI contrast study. In all cases the contrast study was equivocal (Table 1) and a diagnosis of intestinal malrotation could not be excluded by an experienced consultant paediatric radiologist (Figure 1). Three subsequently underwent abdominal ultrasound to assess the orientation of the superior mesenteric vessels. Ultrasound revealed normal SMA/SMV orientation in two and abnormal in one. In all infants the UGI contrast study with or without US could not conclusively exclude a diagnosis of malrotation. Upon discussion with the consultant surgeon and radiologist it was felt safest to proceed to diagnostic laparoscopy to provide diagnostic certainty.

*Surgery*

The procedure was performed as described in the methods in all 8 cases. There was no evidence of intestinal malrotation in 6 cases. In these 6 cases the DJ flexure was positively identified in the left upper quadrant to the left of the midline with the proximal duodenum passing behind the root of the mesentery in a fixed retroperitoneal position. The caecum was positively identified on the right and found to be fixed to the posterior/lateral peritoneal wall in all cases although in 2 cases this was midway down the right side of the abdomen rather than truly in the right iliac fossa. In these 6 infants, none were found to have Ladd’s bands, nor any other cause of extrinsic duodenal compression and no other causative pathology was noted during laparoscopy. All were found to have a wide based mesentery. Infants 7 and 8 had confirmed malrotation on laparoscopy; one with volvulus and one without. Both cases were converted to laparotomy for correction of malrotation performed (Ladd’s procedure) as per surgeon preference.

*Postoperative course*

In the 6 infants without malrotation, median postoperative hospital stay was 2 days (range 2-3 days). These 6 infants were allowed to feed upon recovery from anaesthesia and rapidly established on full enteral feeds with no infant receiving parenteral nutrition. All 6 infants recovered without complication and were discharged home. None has experienced a recurrence of symptoms requiring further investigation nor operative management at median follow-up 27.5 months (range 9 - 46 months). Infant 7 required a longer hospital stay due to post-operative ileus, need to re-establish feeds and demonstrate weight gain. Infant 8 progressed well postoperatively and was discharged on day 5.

## Discussion

In cases with clinical features concerning for malrotation the paediatric surgeon seeks diagnostic certainty. If radiological investigations cannot provide such certainty then historically most infants would undergo a laparotomy to examine the orientation and position of the midgut within the abdominal cavity. It is well recognised that open abdominal surgery in children has a long-term morbidity with an overall readmission rate with small bowel obstruction of 5.3% [8], rising to 8.3% following neonatal laparotomy [9] and 10-15% specifically for malrotation[9, 10]. Additionally short term recovery has been reported to be swifter following minimally invasive compared to open surgery [11]. In an attempt to reduce short and long term morbidity associated with diagnostic surgery we have performed diagnostic laparoscopy in cases concerning for malrotation with equivocal imaging despite the involvement of an experienced paediatric radiologist.

Using this approach we have been able to examine the orientation and position of the midgut within the abdominal cavity to provide the certainty we seek that the rotation is normal whilst minimising the morbidity of the procedure through the use of laparoscopy rather than laparotomy. In two cases of malrotation, the laparoscopic findings were clear cut and we proceeded to laparotomy. In none of the remaining cases did we identify any additional anatomical findings requiring further surgical intervention. Although our experience is limited we believe there are key steps to the diagnostic laparoscopy that will help avoid false negative examinations. It is paramount to have clear visualisation and confirm key anatomical structures including (i) the DJ flexure to the left of the midline fixed by a ligament of Treitz and sited in the left upper quadrant; (ii) the proximal duodenum passing behind the root of the mesentery in a fixed retroperitoneal position; (iii) the caecum on the right and fixed to the posterolateral peritoneal wall; (iv) no Ladd’s bands from the caecum or right colon crossing the duodenum; (v) a wide-based mesentery and no other causative pathology. Using this structured approach we have achieved diagnostic certainty, avoided open laparotomy and importantly not missed any cases of malrotation (although we acknowledge relatively short follow-up). If there were to be uncertainty regarding the laparoscopic findings we recommend a low threshold for conversion to open procedure.

Whilst the lack of a comparison group precludes firm conclusions we believe there are benefits to diagnostic laparoscopy over an open diagnostic procedure including improved cosmesis, rapid return to full feeds, short post-operative hospital stay and reduced long-term risk of adhesional complications. We acknowledge other minimal incision surgery via transumbilical incision has being reported [20], however only a limited number for malrotation. Although cosmetically appealing, such an approach would still require bowel handing with greater possibility of post-operative ileus and adhesional bowel obstruction than laparoscopy.

We acknowledge that some may challenge the use of surgical intervention in these infants at all citing high sensitivity and specificity from radiological investigations including in particular advanced ultrasound technologies. Whilst the literature does indeed support high sensitivity and specificity of upper GI contrast study and ultrasound, particularly when used in combination, it is well recognised that these investigations may be user dependent and simply do not provide diagnostic certainty 100% of the time in all infants. Thus when in the rare situation that the radiologist cannot provide the certainty that is sought, the surgeon is obliged to continue the process of investigation. Some may also suggest that a repeat delayed UGI contrast study may be appropriate rather than proceeding to diagnostic laparoscopy. It has been our institutional preference to achieve diagnostic certainty as soon as possible in cases of possible malrotation since there are potentially catastrophic outcomes associated with delay in the event that an infant has intestinal malrotation with volvulus. Furthermore repeat delayed UGI contrast study does not always improve the diagnostic certainty. Our opinion is that this minimally invasive approach is therefore justified.

We also acknowledge that many others have proposed the use of therapeutic laparoscopy in the management of patients with proven rotational abnormality including non- and malrotation [13-16] and that there are a small number of previous reports of diagnostic laparoscopy in cases of suspected malrotation in older children [17, 18]. To our knowledge there is just one previous report of diagnostic laparoscopy for this indication in infants [19]. We recognise that neonatal laparoscopy is an advanced skill set, therefore only recommend this procedure be performed by surgical and anaesthetic teams with appropriate experience and expertise.

In summary we recommend the use of diagnostic laparoscopy as we have described as an adjunct investigation for this small group of infants with clinical concern for malrotation but in whom radiological investigations are inconclusive.

**Compliance with Ethical Standards:**

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Conflict of interest: Authors K Sloan, A Alzamrooni, FE Stedman, O Ron, NJ Hall declare no conflict of

interest.

Ethical approval: This study was registered in our institution as a service evaluation in accordance with local requirements for ethical approval.

 Informed consent: Informed consent not applicable for this retrospective case note review.

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