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Review Articles

Asymptomatic malrotation: Diagnosis and surgical management An American Pediatric Surgical Association outcomes and evidence based practice committee systematic review

Kathleen Graziano [a,*], Saleem Islam [b], Roshni Dasgupta [c], Monica E. Lopez [d], Mary Austin [e], Li Ern Chen [f], Adam Goldin [g], Cynthia D. Downard [h], Elizabeth Renaud [i], Fizan Abdullah [j]

[a] Division of Pediatric General Surgery, Phoenix Children's Hospital, Phoenix, AZ
[b] Division of Pediatric Surgery, University of Florida, Gainesville, FL
[c] Division of Pediatric General and Thoracic Surgery, Cincinnati Children's Hospital, Cincinnati, OH
[d] Michael E. DeBakey Department of Surgery, Baylor College of Medicine, Houston, TX
[e] Department of Surgical Oncology, University of Texas M.D. Anderson Cancer Center; Department of Pediatric Surgery, University of Texas Medical School, Houston, TX
[f] Department of Surgery, Baylor University Medical Center, Dallas, TX
[g] Division of Pediatric General and Thoracic Surgery, Seattle Children's Hospital; Department of Surgery, University of Washington, Seattle, WA
[h] Division of Pediatric Surgery, Hiram C. Polk, Jr, MD; Department of Surgery, University of Louisville, Louisville, KY
[i] Division of Pediatric Surgery, Department of Surgery, Albany Medical Center, Albany, NY
[j] Department of Surgery, Johns Hopkins University School of Medicine, Baltimore, MD

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ABSTRACT

Objective: Patients with malrotation, or an intestinal rotation abnormality (IRA), can experience serious adverse events. Increasingly, asymptomatic patients are being diagnosed with malrotation incidentally. Patients with symptomatic malrotation require surgery in an urgent or semiurgent manner to address their symptoms. The treatment of asymptomatic or incidentally discovered malrotation remains controversial.
Methods: Data were compiled from a broad search of Medline, Cochrane, Embase and Web of Science from January 1980 through January 2013 for five questions regarding asymptomatic malrotation.
Results: There is minimal evidence to support screening asymptomatic patients. Consideration may be given to operate on asymptomatic patients who are younger in age, while observation may be appropriate in the older patient. If reliably diagnosed, atypical malrotation with a broad-based mesentery and malposition of the duodenum can be observed. Regarding diagnostic imaging, the standard of care for diagnosis remains the upper gastrointestinal contrast study (UGI), ultrasound may be useful for screening. A laparoscopic approach is safe for diagnosis and treatment of rotational abnormalities. Laparoscopy can aid in determining whether a patient has true malrotation with a narrow mesenteric stalk, has nonrotation and minimal risk for volvulus, or has atypical anatomy with malposition of the duodenum. It is reasonable to delay Ladd procedures until after palliation on patients with severe congenital heart disease. Observation can be considered with extensive education for family and caregivers and close clinical follow-up.
Conclusions: There is a lack of quality data to guide the management of patients with asymptomatic malrotation. Multicenter and prospective data should be collected to better assess the risk profile for this complex group of patients. A multidisciplinary approach involving surgery, cardiology, critical care and the patient's caregivers can help guide a watchful waiting management plan in individual cases.

Malrotation, also known as intestinal rotation abnormality (IRA), can pose a great risk to the pediatric patient owing to the potential for a catastrophic event such as midgut volvulus, ischemic bowel, and possible short bowel syndrome or death [1]. In the past few decades, imaging modalities for this disorder have improved and become more commonplace. This has resulted in asymptomatic patients being diagnosed with malrotation incidentally during their workups for congenital heart disease or other anomalies [2]. The Centers for Disease Control reports the prevalence of malrotation to be 3.9 per 10,000 live births, while others estimate a prevalence as high as 1 in 500 live births [3]. While management recommendations for neonates and infants with symptomatic malrotation are well established, the treatment of asymptomatic or incidentally discovered malrotation remains controversial. In the present review, the APSA Outcomes and Evidence Based Practice Committee summarizes the available literature regarding the management of asymptomatic malrotation. For the purposes of this review we have divided the anatomical definitions into three categories:

* Corresponding author at: Division of Pediatric Surgery, Phoenix Children's Hospital, 1920 E Cambridge AVe, Ste 201, Phoenix, AZ 85048.
E-mail address: kgraziano@surgery4children.com (K. Graziano).

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1. True malrotation with a narrow mesenteric stalk
2. Nonrotation with a broad mesentery
3. Atypical malrotation, defined as malposition of the ligament of Treitz or duodenal malposition.

1. Methods

The APSA Outcomes and Evidence Based Practice Committee proposed five broadly applicable questions regarding asymptomatic malrotation as the focus for this systematic review:

1. Which patients should undergo screening for malrotation?
2. Do all patients with a diagnosis of malrotation need to undergo a Ladd procedure?
3. What is the best imaging study to diagnose malrotation?
4. What is the optimal surgical technique to address malrotation? Laparotomy or laparoscopy?
5. What is the best approach to treat "asymptomatic" malrotation in patients with severe congenital heart disease?

Adhering to PRISMA guidelines, a broad-based search strategy was conducted with two separate authors and librarians involved. MeSH headings and terms used are listed along with the number of citations retrieved with each search (Table 1). Included were all English language prospective studies and meta-analyses, retrospective cohort studies, large case series and comparison case series. Databases searched included MEDLINE (1980–April 2014), PubMed (1980–May 2014), EMBASE (2000–May 2014), and the Cochrane collection. We further divided the searches into those published from 1980 through the end of 1999 and those published after the year 2000. We retrieved 2322 citations that were cross-referenced and screened. Abstracts were then reviewed and relevant articles selected by two of the authors (KG and SI), who also then performed independent manual searches of the reference lists of these articles, bringing the total reviewed to 2545. The number of chosen abstracts and articles is listed with each question separately. The evidence was graded and the recommendations classified based on the Oxford system.

1.1. Question 1: Which patients should undergo screening for malrotation?

A total of 224 abstracts were found in a broad search strategy and screened. No prospective studies were found. Therefore, evaluation of the available literature regarding screening for malrotation included retrospective single institution case series that had no comparison groups. Historical bias, selection bias, and institutional bias were noted in most studies.

Table 1
Systemic review search strategy: MeSH headings.

MeSH combinations	Number of articles 1980–2000	Number of articles after 2000
Children/malrotation	226	364
Pediatric/malrotation	79	289
Asymptomatic/malrotation	15	62
Ultrasound/malrotation	85	177
UGI/malrotation	27	44
Ladd procedure/malrotation	12	48
Malrotation/heterotaxy	4	44
Malrotation/cardiac	70	122
Volvulus/heterotaxy	0	21
Risk/intestinal malrotation	26	78
Anomalies intestinal rotation	109	135
Malrotation/radiology diagnosis	235	393
False negative/malrotation	3	8
Laparoscopy/Ladd	6	19

The World Health Organization published guidelines for what constitutes effective population screening tests in 1968 [4] (see Table 2). These guidelines can be applied to the population of patients with asymptomatic malrotation. Screening for malrotation is important in the context of the dire consequences that can result from a missed diagnosis. Imaging modalities used to diagnose malrotation carry minimal risk. The natural history of malrotation is not completely understood and can vary widely. The ability to recommend a procedure or treatment is controversial in this group of patients. Issues of cost of screening have not been worked out formally as there are no prospective studies. The WHO guidelines, when broadly applied, suggest that patients with a higher prevalence of malrotation are an appropriate group to screen. This target population consists of patients with congenital heart disease (in particular heterotaxy syndrome (HS)), and pediatric patients with "known" intestinal rotation anomalies such as congenital diaphragmatic hernia (CDH), and abdominal wall defects.

In cases where there is a known malrotation, such as congenital diaphragmatic hernia (CDH) or abdominal wall defects, it is unusual to perform a formal Ladd procedure as part of the initial repair of the congenital anomaly. The rate of complications from malrotation in patients with gastroschisis, omphalocele or CDH is not available in the literature. Sinha et al. published a review of their 18-year experience of omphalocele and malrotation, in which they looked at 42 cases of patients with abdominal wall defects [5]. They noted a 45% incidence of malrotation in the "major" anomaly (abdominal wall defect >5 cm) and 31% in the "minor" one (defect <5 cm). Malrotation was defined operatively if the duodenal-jejunal flexure was to the right of the midline, with or without a narrow mesenteric base. They reported one case of midgut volvulus, and one complication of a Ladd procedure in their series. The authors recommended screening during the initial surgery or postoperatively with UGI, although no supporting evidence was presented to support for this recommendation. Levin et al. found that in 24 patients with CDH who had an upper gastrointestinal contrast study (UGI), all had some rotational anomaly, but the right-sided CDH had more "obvious" ones. There were no cases that had volvulus, which the authors attributed to adhesions [6]. The anatomical definitions in the studies did not clearly distinguish between true malrotation, nonrotation with a broad mesenteric base and atypical malrotation and therefore it is difficult to support routine Ladd procedures in these patients. Most patients with CDH or abdominal wall defects have a form of nonrotation and are therefore not likely to be at risk for midgut volvulus.

Congenital heart disease (CHD), and in particular HS, are associated with malrotation in 40–90% of cases. The concern is for the patient with HS who may have an episode of volvulus and have a poor outcome from intestinal ischemia. Choi et al. from Toronto looked at their experience with 177 patients with HS over 34 years [7]. Of these patients, 152 were described as asymptomatic. Nine of these asymptomatic patients underwent a screening UGI, with 6 abnormal results. There was not a distinct definition of what constituted abnormal anatomy. The

Table 2
World Health Organization Principles of Screening (Wilson's Criteria) [4].

1. The condition should be an important health problem.
2. There should be a treatment for the condition.
3. Facilities for diagnosis and treatment should be available.
4. There should be a latent stage of the disease.
5. There should be a test or examination for the condition.
6. The test should be acceptable to the population.
7. The natural history of the disease should be adequately understood.
8. There should be an agreed policy on whom to treat.
9. The cost of finding a case should be economically balanced in relation to medical expenditure.
10. Case-finding should be a continuous process, not just a "once and for all" project.