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Early versus Delayed Surgical Correction of Malrotation in Children with Critical Congenital Heart Disease

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Abstract

Purpose—The purpose of this study was to compare outcomes between early and delayed surgical correction of malrotation in children with critical congenital heart disease (CHD).

Methods—Patients with CHD who underwent cardiac surgery by 1 year of age and had malrotation diagnosed during their initial admission at 34 hospitals contributing to the Pediatric Health Information System in 2004–2009 were included. Ladd's procedures performed during the first admission were considered early correction, and those at a subsequent admission were considered delayed. Inter-hospital variability in the proportion of patients undergoing delayed correction was assessed, and outcomes were compared between the groups.

Results—Of the 324 patients identified, 85.2% underwent early correction. Significant variability existed in the proportion of patients undergoing delayed correction across hospitals ($p < 0.0001$). Baseline characteristics, including severity of CHD, were similar between the groups. In the delayed group, 27% of patients underwent a Ladd's procedure during an urgent or emergent admission, but none had volvulus or underwent intestinal resection. Rates of mortality and readmission within 1 year of malrotation diagnosis were similar in both groups. Chart validation confirmed 100% accuracy of diagnosis and treatment group assignment.

Conclusions—In patients with critical CHD, delayed operative intervention for malrotation without volvulus may be a reasonable alternative.

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Keywords

malrotation; congenital heart disease; Ladd's procedure; midgut volvulus

Introduction

Intestinal malrotation is an associated anomaly in patients with congenital heart disease (CHD). In this high risk group of children, current practice has traditionally included urgent abdominal exploration for any neonate with malrotation. However, some investigators have suggested that early exploration may not be necessary in all patients.(1, 2) In particular, neonates with critical CHD may be at increased risk for peri-operative complications that may outweigh the potential benefit of a Ladd's procedure to prevent intestinal volvulus during their index admission for cardiac repair.(3-5)

Given the relative rarity of these coexisting conditions, studies in this population consist of only a handful of single-center series, and many of these focus on children with heterotaxy. (6-8) The objectives of this study are to: 1) create, validate, and describe a multi-institutional cohort of patients with both congenital heart disease and malrotation who underwent repair of both anomalies, 2) determine if practice variability occurs in the timing of the correction of malrotation across hospitals, and 3) identify any differences in outcomes in patients undergoing early versus delayed correction of malrotation

Methods

Data Source and Cohort Development

The Pediatric Health Information System (PHIS) is an administrative database for 44 free-standing children's hospitals managed by the Children's Hospital Association. It contains demographic information, International Classification of Diseases, 9th Revision – Clinical Modification (ICD-9-CM) diagnosis codes, and date-stamped codes for procedures, radiology and laboratory tests, and medications.

A cohort of patients with critical CHD as defined by the Agency for Healthcare Research and Quality was developed using a combination of the diagnosis code for congenital heart disease and a procedure code for a cardiac surgical procedure by 1 year of age.(9) Patients were excluded if they underwent cardiac transplantation or if they underwent isolated patent ductus arteriosus ligation. Patients born between January 2004 and December 2009 who were treated at one of the 34 PHIS hospitals that contributed inpatient data to PHIS during the entire study period were included (Figure 1). We then identified patients who had a malrotation diagnosis code (ICD-9-CM 751.4) at the admission at which they first had a CHD diagnosis, and who also had a procedure code for a Ladd's procedure (ICD-9-CM 46.80, 46.81, 46.82, 54.95) by the end of 2012. Because the code 54.95 is also a code for revision of a ventriculo-peritoneal (VP) shunt, we excluded any patients who had a VP shunt placed before this code occurred. The index admission was defined as the first admission where the diagnosis of malrotation was made (i.e. the diagnosis code for intestinal malrotation was associated with the encounter). Patients with early correction of malrotation

were defined as those who had the Ladd's procedure at the index admission. Patients with delayed correction were those who had the Ladd's procedure at a subsequent admission.

Medical record chart review validation of all patients treated at four of the 34 included PHIS hospitals (Nationwide Children's Hospital, Columbus, OH; Children's Hospital of Atlanta, Atlanta, GA; Children's Hospital Boston, Boston, MA; and Monroe Carell Jr Children's Hospital, Nashville, TN) was performed to estimate the rates of misclassification of variables in the PHIS. The validation cohort represents 12.7% of the entire cohort. The institutional review boards of each institution approved this study.

Outcomes and Exposures

Outcomes were evaluated up to one year following the Ladd's procedure. The primary outcomes for comparison between early and delayed treatment groups were the rates of readmission, major abdominal operations, volvulus, and in-hospital mortality. In the delayed group, outcomes also included events that occurred after diagnosis of malrotation and discharge from the index admission, but prior to the Ladd's procedure, which was presumably not performed right away because the goal of treatment was either a delayed elective Ladd's procedure or watchful waiting. The pre-Ladd's outcomes, which were specific to the delayed group, included (1) an emergency admission for their Ladd's procedure, defined as admission through the ED, urgent or emergent priority of admission, or a primary diagnosis of intestinal perforation or volvulus (ICD-9-CM 569.83, 560.2); (2) a diagnosis of volvulus (ICD-9-CM 560.2) at any admission after the malrotation diagnosis, including the admission at which Ladd's was performed; and (3) any intestinal resection up to and including the date of the Ladd's procedure. It should be noted that patients in the early group were unable to be classified as having an urgent or emergent admission by the first two criteria (ED admission and urgent/emergent priority of admission) and were considered as have undergone elective surgery.

Baseline characteristics included demographic and clinical characteristics at the index admission where congenital heart disease and malrotation were first diagnosed. Characteristics examined included age, gender, race, insurance status, birth weight, gestational age, prematurity, congenital anomalies, cardiac operations, and the maximum Risk Adjustment for Congenital Heart Surgery (RACHS-1) category at any admission up to age 3.⁽¹⁰⁾ The total number of critical CHD patients treated at each hospital during the study period was also evaluated.

Statistical Analysis

Baseline characteristics were compared between treatment groups using two sample t-tests or Wilcoxon rank sum tests for continuous variables and Pearson chi square or Fisher's exact tests for categorical variables. Outcomes were compared between groups using logistic or linear mixed effects models or, for rare binary outcomes, exact logistic regression models adjusting for exposures that differed between treatment groups at $p < 0.05$ in bivariate analyses. Inter-hospital variability in the timing of the Ladd's procedure was assessed using a logistic mixed effects model that included random hospital intercepts, hospital volume of critical CHD patients, and any patient-level characteristics that differed between groups at

$p < 0.20$ in bivariate analyses. All analyses were performed using SAS v9.3 (SAS, Cary, NC). All tests were 2-tailed with $p < 0.05$ considered statistically significant.

Results

Cohort Development, Validation, and Inter-Hospital Variability

Of the 324 patients who met inclusion criteria at 34 freestanding children's hospitals, 276 (85.2%) were identified as having early correction of their malrotation and 48 (14.8%) met criteria for delayed correction (Figure 1). Chart review validation of all patients treated at four PHIS hospitals ($n = 41$, 12.7% of total cohort) demonstrated no misclassification in the PHIS for the diagnosis of critical CHD or malrotation, for treatment group assignment, and for outcomes, including reoperation, emergency department visits related to malrotation, and in-hospital mortality. Comorbidities, based on ICD-9-CM diagnosis codes present during the index admission, had an average rate of misclassification of 10.5%.

Significant variability exists in the percentage of patients undergoing delayed Ladd's procedures at each hospital both before and after adjustment for case-mix and hospitals' volume of critical CHD patients (Figure 2). The percentage of patients undergoing delayed Ladd's ranged from 0-50% before and 2%-40% after adjustment.

Comparisons of Early vs. Delayed Groups: Baseline Characteristics and Outcomes

Baseline characteristics are shown in Table 1. Patients in the delayed group were older at the time of their Ladd's procedure (median (inter-quartile range): 200 (106, 318) days vs. 21 (8, 39) days; $p < 0.001$) and were more likely to have a cardiac surgical procedure prior to their Ladd's (95.8% vs. 58.3%, $p < 0.001$). The median time between the date of discharge and the Ladd's procedure in the delayed group was 127 (41, 231) days. Of note, there was no significant difference between groups in patients' maximum RACHS-1 scores up to 3 years of age. The most common types of CHD diagnosed at the index admission (prevalent at $>10\%$ in either group) were similar in both groups with the exception of higher frequencies of endocardial cushion defects ($p=0.03$), malposition of the heart ($p=0.001$), and pulmonary artery anomalies ($p=0.008$) in the delayed group (Table 2). The most frequent additional procedures performed at the time of the Ladd's procedure in each group included: Early group: gastrostomy tube 29%, small bowel anastomosis 8%, and fundoplication 8%; Delayed group: gastrostomy tube 23%, fundoplication 13%, myringotomy tube insertion 6%, and circumcision 6%.

Within the delayed group, 27.1% of the patients were admitted to the hospital emergently but none had a diagnosis of volvulus leading up to and including the Ladd's procedure admission. The most common primary diagnoses for these emergent admissions in which the Ladd's procedure was performed were malrotation ($N=5$, 38%) and malposition of the heart ($N=2$, 15%). Compared to patients undergoing early correction, patients undergoing delayed correction less frequently received post-operative mechanical ventilation, total parenteral nutrition (TPN), and blood products, and had a shorter post-operative length of stay after their Ladd's procedure (Table 3). Four patients (1.4%) in the early group had a diagnosis of volvulus at an encounter within 1 year of the index admission compared to none

in the delayed group. Of note, for early repair, this represents the rate of volvulus in the 1 year after the Ladd's procedure was performed, and for delayed Repair, this reflects the rate of volvulus both prior to and after the Ladd's procedure, which in most of these patients was performed within 6 months of the index admission.

Discussion

Utilizing the PHIS database, we accurately identified a multi-institutional cohort of patients with critical CHD and malrotation who underwent Ladd's procedure either at the same admission as the diagnosis of malrotation was made or at a later admission. After accounting for differences in patient and hospital characteristics, there was significant variability between hospitals in the percentage of these patients treated with a delayed Ladd's procedure. Amongst the group that underwent delayed correction, 27% had their Ladd's procedure performed during an urgent/emergent admission; however, none had volvulus or required intestinal resection. Overall, there were no differences between the groups in readmissions or mortality following the Ladd's procedure.

The significant variability in the percentage of CHD patients with malrotation treated with delayed correction between PHIS hospitals reflects the lack of consensus in the management of these patients. Determining the appropriate time to perform a non-emergent non-cardiac procedure in patients with critical CHD depends on many factors including the risks and benefits of the specific procedure, the type and severity of the patient's CHD, whether any palliative or reparative cardiac procedures have been performed, and the presence of additional non-cardiac comorbidities. In addition, other considerations may include the distance of the patient's home from adequate medical care and the parental comfort level with monitoring for signs and symptoms of volvulus.

In the case of Ladd's procedure for intestinal malrotation, the benefit of surgical correction to decrease the risk of developing midgut volvulus is well established.(6, 7, 11) The controversy exists over the timing of surgery to optimally balance the risks of surgery with the risks of volvulus in this vulnerable population.(12-15) The current study demonstrates that most surgeons at PHIS hospitals will surgically correct malrotation in patients with critical CHD during the admission in which it is first diagnosed with only 15% of patients undergoing a delayed correction. However, in the group of patients undergoing a delayed repaired, no patients presented with volvulus or underwent bowel resection at the time of their Ladd's procedure. In addition, following their Ladd's procedure, patients in the delayed group were treated less frequently with mechanical ventilation, total parenteral nutrition (TPN), and blood products, and had a shorter post-operative length of stay. Of note, more patients in the early correction group had cardiac surgical procedures following the Ladd's procedure, which may partially account for their higher readmission rate. On the other hand, more patients in the delayed group had cardiac surgery prior to their Ladd's procedure potentially indicating that these patients had a more severe type of CHD that is not captured when using RACHS scores as the method to control for severity of CHD.

Most surgeons agree that the benefits of performing a Ladd's procedure outweigh the peri-operative risks; however, there are some groups who advocate waiting until symptoms

develop to surgically correct malrotation in patients with heterotaxy citing the low risks associated with waiting for symptoms prior to intervention.(1, 2, 12, 15) Our results support a low risk of volvulus in a more diverse group of patients with CHD undergoing delayed correction; however, 27% of patients undergoing delayed correction in our study had their procedure performed during an urgent/emergent admission. This suggests that symptoms attributable to malrotation may develop more frequently in the larger more diverse group of patients with CHD and malrotation. It should be noted that we are unable to determine from this dataset whether the patients in the delayed group were planned to have an elective delayed Ladd's procedure or if the treatment decision was for observation only and they subsequently developed symptoms that prompted surgical intervention. Heterotaxy could not be specifically examined in this study because there is currently no accurate way of identifying it using ICD-9 diagnosis codes.

This study has several limitations. First, despite rigorous quality control measures, misclassification of data within the PHIS may occur.(16) For this reason, we validated the accuracy of our diagnoses and treatment group assignments using a multi-institutional chart review, which included 12.7% of the patients of the total cohort. In addition, to minimize misclassification, our cohort definition included undergoing a Ladd's procedure which excluded any patients who were managed with observation alone; this likely underestimated the number of patients in the delayed group and overestimated the rate of delayed patients with an urgent/emergent admission. Second, diagnosis codes within an encounter in the PHIS are not date-stamped making it difficult to determine the timing of some diagnoses in relation to the Ladd's procedure. To minimize this, we focused on outcomes that are based on procedure codes which include date-stamps that allow for determining their temporal relationship to the Ladd's procedure. Third, patient encounters at institutions that do not contribute data to the PHIS are not captured in this analysis. However, patients of this complexity will often receive lifelong care at a single tertiary care center.(17-19) Fourth, despite accounting for characteristics that may drive variability in our hospital variability analyses, unmeasured differences in case mix between hospitals may still account for the observed variability. Fifth, differences in outcomes between the treatment groups may not be directly attributable to the timing of surgical correction; the small sample size of delayed patients limited our ability to perform risk-adjusted comparisons that can account for differences in exposures between the groups. The small sample size also reduced the likelihood of the study to detect a rare event, such as volvulus. Lastly, we were unable to determine if Ladd's procedures performed in the early group were done in an elective manner or not because individual procedures in the PHIS database are not classified as urgent, emergent, or elective; this limits our ability to directly compare outcomes after emergent or urgent Ladd's procedures between the two groups.

In conclusion, this study demonstrates significant variability in practice patterns in the timing of surgical correction of malrotation in patients with CHD across PHIS hospitals. Patients undergoing delayed surgical correction of malrotation in this study did not develop volvulus or undergo bowel resection while waiting for their Ladd's procedure and did not have an increase in readmissions or mortality. These findings suggest that delaying surgical correction of malrotation in selected patients with critical CHD may be a reasonable

alternative; however, larger studies are warranted to determine the safety of delaying correction of malrotation.

Biography

ERIC SCAIFE (Salt Lake City, UT): I wonder if you might consider going one step further. I'm not saying delay but abandon screening. With zero volvulus and a significant mortality both in your delayed and your early group, one might actually argue that these patients would be safer if they had no Ladd's procedure at all.

JASON SULKOWSKI: Thank you for that comment. That is actually something that has been reported in the literature, the idea of just sort of watchful waiting for patients in this situation. That is something that given the constraints as I had previously mentioned of the administrative database – one of the ways that we are able to better ensure that we're looking at the patients that we think we're looking at, is by combining both diagnosis and procedure codes so a patient who has malrotation is one who has a diagnosis of malrotation and also has a Ladd's procedure. If we just relied on the diagnosis code, we might not necessarily have as good a chance of identifying the correct patients. Something like that would not necessarily be as amenable to this type of study but in terms of other prospective clinical analysis would certainly be a great thing to look at.

CHARLES STOLAR (Santa Barbara, CA): I would just suggest that if we're going to do nothing about this be aware that there are at least anecdotes of children re-emerging with a volvulus and death even though you may not have uncovered some in this database. There are reports of volvulus and death associated with errors or rotation and congenital heart disease, number one. Number two, the pediatricians and the pediatric cardiologists often will do upper GI series as a routine and uncover errors in rotation. As I have said before, once the word malrotation enters the medical record, it puts us at some liability exposure.

SALEEM ISLAM (Gainesville, FL): As a member of the outcomes committee which actually presented this data yesterday, it would have been nice to have included this in it. However, I think that your ...

CHARLES STOLAR (Santa Barbara, CA): Are you getting testy with our presenter?

SALEEM ISLAM: Not at all, not at all. (laughter) The primary person is actually a member of the outcomes committee on this as well. I think that your conclusions are in agreement with what we said broadly, with the lack of any good evidence, that observing these patients based on two large studies was appropriate and seemed to be in the right direction. I think it behooves us all to probably look at our data more closely and have better outcomes clinical database so we can actually answer these questions better. Thank you for your presentation.

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Figure 1.
Development of cohort and treatment groups from the PHIS database.

Figure 2.

Inter-Hospital Variability in Timing of Ladd Procedure.

Open circles and dotted lines = Unadjusted estimates with 95% confidence intervals (CI) of the percentage of patients with a delayed Ladd procedure at each hospital; solid circles and lines = Adjusted estimates with 95% CI of the percentage of patients with a delayed Ladd procedure at each hospital. Adjustment factors included age at the index admission at which malrotation was diagnosed, insurance, prematurity, any major non-cardiac structural anomaly, palate anomaly, Downs syndrome, having at least one cardiac surgery procedure before the Ladd procedure, having at least one cardiac surgery procedure after the Ladd procedure, and the maximum RACHS-1 category at any admission up to age 3 years. An asterisk denotes a hospital with an adjusted estimate significantly different from the average as shown by the reference line.

Table 1

Baseline characteristics of study cohort.

Variable	Total Cohort (N=324)	Early Repair (N=276)	Delayed Repair (N=48)	P
Age at first cardiac surgery (days)	16 (5, 76.5)	16 (5, 75)	16 (4, 85.5)	0.64
Age at admission when malrotation first diagnosed (days)	0 (0, 3)	0 (0, 2)	1 (0, 26.5)	0.01
Age at Ladd procedure (days)	25.5 (10.5, 59.5)	21 (8, 39)	200 (106, 317.5)	<.001
Male	160 (49.4)	139 (50.4)	21 (43.8)	0.40
Birth Weight (g) ^a	2940 (2450, 3370)	2950 (2410, 3400)	2914 (2615, 3165)	0.82
Gestational age (weeks) ^b	38 (36, 39)	38 (36, 39)	39 (37, 40)	0.10
Race				
White	203 (62.7)	171 (62.0)	32 (66.7)	
Black	40 (12.3)	34 (12.3)	6 (12.5)	0.76
Other/ Unknown	81 (25.0)	71 (25.7)	10 (20.8)	
Premature	60 (18.5)	55 (19.9)	5 (10.4)	0.12
Any major non-cardiac structural anomaly (not including malrotation)	122 (37.7)	109 (39.5)	13 (27.1)	0.10
Other GI anomaly besides Malrotation	94 (29.0)	80 (29.0)	14 (29.2)	0.98
Neurologic anomaly	31 (9.6)	28 (10.1)	3 (6.3)	0.59
Head & Neck anomaly	2 (0.6)	2 (0.7)	0 (0.0)	1.0
Respiratory anomaly	51 (15.7)	44 (15.9)	7 (14.6)	0.81
Palate anomaly	9 (2.8)	6 (2.2)	3 (6.3)	0.13
Renal anomaly	50 (15.4)	45 (16.3)	5 (10.4)	0.30
Genital anomaly	15 (4.6)	14 (5.1)	1 (2.1)	0.71
Downs Syndrome	30 (9.3)	29 (10.5)	1 (2.1)	0.10
Other chromosomal anomaly	25 (7.7)	19 (6.9)	6 (12.5)	0.23
<i>Cardiac Surgical Procedure Details</i>				
At least one cardiac surgery procedure before Ladd procedure	207 (63.9)	161 (58.3)	46 (95.8)	<.001
At least one cardiac surgery procedure after Ladd procedure and before age 3	229 (70.7)	208 (75.4)	21 (43.8)	<.001
Maximum RACHS-1 category at any admission up to age 3				
1	7 (2.2)	5 (1.8)	2 (4.2)	0.10 ^c
2	60 (18.5)	52 (18.8)	8 (16.7)	
3	129 (39.8)	107 (38.8)	22 (45.8)	
4	66 (20.4)	53 (19.2)	13 (27.1)	
5/6	26 (8.0)	24 (8.7)	2 (4.2)	
Unclassifiable	36 (11.1)	35 (12.7)	1 (2.1)	

Data are presented as frequency and percent or median and interquartile range.

^aTotal cohort N=269, Early cohort N=239, Delayed Cohort N=30.

^bTotal cohort N=201, Early cohort N=174, Delayed Cohort N=27.

^cIf patients with unclassifiable RACHS-1 scores are excluded, p=0.49. GI, gastrointestinal; RACHS-1, Risk Adjustment for Congenital Heart Surgery

Table 2

Most frequent types of Congenital Heart Disease diagnosed at the index admission in each group

Early Repair		Delayed Repair	
Diagnosis	Number of patients (%)	Diagnosis	Number of patients (%)
Endocardial cushion defect NEC	109 (39.5)	Endocardial cushion defect NEC	26 (54.2)
Ostium secundum type atrial septal defect	92 (33.3)	Malposition of the heart	20 (41.7)
Double outlet right ventricle	84 (30.4)	Pulmonary artery anomaly	18 (37.5)
Ventricular septal defect	69 (25)	Ventricular septal defect	15 (31.3)
Pulmonary artery anomaly	60 (21.7)	Ostium secundum type atrial septal defect	15 (31.3)
Total anomalous pulmonary venous return	58 (21)	Total anomalous pulmonary venous return	14 (29.2)
Malposition of the heart	54 (19.6)	Double outlet right ventricle	11 (22.9)
Common ventricle	45 (16.3)	Common ventricle	9 (18.8)
Coarctation of aorta	40 (14.5)	Great vein anomaly NEC	8 (16.7)
Hypoplastic left heart syndrome	37 (13.4)	Congenital pulmonary valve stenosis	7 (14.6)
Other congenital heart anomaly	37 (13.4)	Other congenital heart anomaly	7 (14.6)
Congenital pulmonary valve stenosis	32 (11.6)	Complete transposition of great vessels	6 (12.5)

NEC, not elsewhere classified

Table 3

Outcomes following Ladd's procedure.

Variable, n (%)	Total Cohort (N=324)	Early Repair (N=276)	Delayed Repair (N=48)	P
Readmitted within 30 days ^a	71 (21.9)	66 (23.9)	5 (10.4)	0.56
Readmitted within 1 year ^a	228 (70.4)	201 (72.8)	27 (56.3)	0.34
Mechanical ventilation post-op ^b	230 (71.0)	211 (76.4)	19 (39.6)	0.03
ECMO post-op ^b	12 (3.7)	12 (4.3)	0 (0.0)	0.60
TPN post-op ^b	223 (68.8)	210 (76.1)	13 (27.1)	0.01
Blood product post-op ^b	52 (16.0)	50 (18.1)	2 (4.2)	0.29
Post-op length of stay (days)	19 (8, 42.5)	22 (11, 47.5)	6 (4, 10.5)	<.001
Any malrotation-related procedure within 1 year of Ladd's	22 (6.8)	20 (7.3)	2 (4.2)	0.55
Volvulus at any admission within 1 year of discharge from admission in which malrotation diagnosis was made ^c	4 (1.2)	4 (1.4)	0 (0.0)	0.1
Died in-hospital within 1 year of malrotation diagnosis	43 (13.3)	41 (14.9)	2 (4.2)	0.08
Died at Ladd's admission	32 (9.9)	31 (11.2)	1 (2.1)	0.55

^aFrom the date of discharge from the admission at which the Ladd's procedure was performed.^bBased on charges incurred and/or procedures performed only on a date after the date that the Ladd's procedure was performed but during that same admission.^cFor Early repair, this reflects the rate of volvulus in the 1 year after discharge from the admission in which the Ladd's was performed; for Delayed Repair, this reflects the rate of volvulus in the 1 year after discharge from the index admission; in most cases the Ladd's was performed by 6 months into this year. For all variables except post-op length of stay and volvulus, p-values are from logistic mixed effects models. For volvulus, an exact logistic regression model was used. For post-op length of stay, p-values are from a linear mixed effects model after natural log transformation. All models included treatment group, age at admission when malrotation was first diagnosed, occurrence of at least one cardiac surgery procedure before Ladd's procedure, and age at Ladd's procedure (not included in the model for volvulus). ECMO, extra-corporeal membrane oxygenation; TPN, total parenteral nutrition