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Malrotation is not associated with adverse outcomes after cardiac surgery in patients with heterotaxy syndrome☆

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ABSTRACT

Background: Patients with heterotaxy syndrome (HS) often have asymptomatic malrotation. There is a lack of consensus regarding the management of these patients, particularly in patients with complex congenital heart disease (CHD). We sought to describe the prevalence of malrotation and incidence of volvulus in a population of patients with complex CHD and to identify the impact of malrotation on morbidity and mortality following cardiac surgery.

Methods: We performed a retrospective review of all patients with HS and complex CHD who required cardiac surgery in the first year of life at a single center between October 1995 and September 2015. Malrotation was diagnosed by abdominal imaging or by direct inspection during abdominal surgery. Demographic data was collected along with details of hospitalization following cardiac and GI surgeries. Descriptive analysis along with appropriate hypothesis testing was conducted to evaluate the results.

Results: We identified 49 patients with HS, 42 with single ventricle anatomy and 7 with biventricular anatomy. Of the 49 patients, 29 (59%) were diagnosed with malrotation, 6 (12%) had normal intestinal rotation, and 14 (29%) had no evaluation of intestinal rotation. The prevalence of malrotation in the population who underwent abdominal imaging was 29 out of 35 (83%). There was no difference in survival following cardiac surgery between patients with malrotation and those with unknown or normal intestinal anatomy. Comparing patients with malrotation and patients with normal or unknown intestinal rotation, there was also no difference in surrogate markers of morbidity. Of the 29 patients with known malrotation, only 2 patients (7%) underwent therapeutic Ladd procedures and 19 (65%) underwent prophylactic Ladd procedures.

Conclusions: We conclude that the outcomes following cardiac surgery for patients with HS are not impacted by the presence of malrotation. Furthermore, we also found that the incidence of volvulus in the studied group is low. Given these findings, and the understanding that patients with HS and significant CHD are frequently tenuous and high risk surgical candidates, we do not believe performing prophylactic Ladd procedures is warranted.

Level of evidence: III.

1. Background

Heterotaxy syndrome (HS) has been defined as a pattern of anatomical organization of the thoracic and abdominal organs which is not the expected or normal arrangement, known as situs solitus [1]. These anatomic abnormalities often include congenital heart disease, ranging from simple septal defects to complex venous and intracardiac anomalies necessitating single ventricle palliation. Intestinal rotational anomalies (IRA) such as malrotation have been reported in 40–90% of patients with HS [2–4]. The feared complication of malrotation is midgut volvulus leading to bowel ischemia. The management of patients with malrotation who go on to have symptoms suggestive of volvulus is widely agreed upon. The Ladd procedure is the accepted treatment for symptomatic malrotation, which includes detorsion of the bowel in the presence of volvulus, division of fibrous bands, broadening the mesentery to reduce future risk of volvulus, placing the bowel in nonrotation, and removal of the appendix [5].

Due to the unknown incidence of volvulus in the HS population there are differing opinions regarding the need to screen patients with HS for malrotation and the management of asymptomatic patients

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with malrotation. Furthermore, the high incidence of complex congenital heart disease in patients with HS can complicate the prophylactic Ladd procedure. It is known that the majority of patients with volvulus will present within the first year of life corresponding to the period of greatest hemodynamic instability of the patient with complex congenital heart disease, especially those with shunt dependency [6]. Yu et al. described the outcomes of the Ladd procedure in patients with HS versus non-HS patients and found that patients with HS had a higher overall mortality than non-HS patients, primarily related to cardiac disease [7]. While cardiac disease may impact the Ladd procedure outcomes it is unknown how malrotation affects patients in the postoperative period following cardiac surgery.

We sought to describe the prevalence of malrotation and incidence of volvulus in a population of patients with heterotaxy syndrome and congenital heart disease, and to describe the factors associated with poor outcomes in this population. We also attempted to investigate the effect that malrotation has on the in-hospital postoperative cardiac surgery period of patients with single ventricle physiology.

2. Methods

2.1. Study design and patient population

We performed a retrospective review of all patients with HS and congenital heart disease who required cardiac surgery in the first year of life at a single center between October of 1995 and September of 2015. Patients with significant congenital heart disease were excluded if they did not undergo cardiac surgery prior to 1 year of age.

2.2. Clinical variables of interest

Malrotation was diagnosed by abdominal imaging (upper gastrointestinal fluoroscopy, CT of the abdomen, or MRI of the abdomen) or by direct inspection during abdominal surgery. Splenic anatomy was determined by abdominal ultrasound and was designated as asplenia, polysplenia, or single spleen. Demographic data was collected along with cardiac diagnoses, cardiac surgery, diagnosis of malrotation, gastrointestinal (GI) surgery, cardiac and splenic anatomy at time of GI surgery, intensive care unit (ICU) length of stay, ventilator days, hospital length of stay, and disposition at the time of discharge. Primary outcome of interest was mortality following cardiac surgery, though secondary analysis of ICU and ventilator days was used as surrogates for morbidity.

2.3. Statistical analyses

Continuous data was tested for normality and appropriate hypothesis testing was performed by the use of t-tests. Results are displayed as means with standard deviation. Categorical data was compared using chi-squared analysis and Fisher's Exact test where appropriate. Survival analysis was performed via Kaplan–Meier with time to event (death) being displayed in days. Due to previous data suggesting that risk of volvulus from malrotation is highest in the first year of life, patients were either labeled as succumbing to the event or were censored to the last available date of known follow-up within the first year of life. Curves were compared via log-rank analysis. All analysis was performed via SPSS (version 22.0, Armonk, NY). The Institutional Review Board at the University of Virginia approved this study.

3. Results

3.1. Patient characteristics

There were 49 patients identified with heterotaxy syndrome and congenital heart disease requiring cardiac surgery in the first year of life. Of those, 42 (86%) of patients had single ventricle anatomy and 7 (14%) underwent biventricular repair. A total of 29 patients were found to have malrotation, 6 had no malrotation on imaging, and 14 had unknown intestinal anatomy. The prevalence of malrotation in the population who underwent abdominal imaging was 29 out of 35 (83%). Table 1 describes the characteristics of our HS population. Of the patients with TAPVR, 28% were obstructed with the majority being in the asplenic subgroup.

3.2. Description of symptomatic patients

There were two patients during the study period who had symptoms concerning for volvulus and underwent therapeutic Ladd procedure. The first patient had known malrotation and presented with abdominal distention in week 7 of life and prior to any cardiac surgery. The patient was found to have volvulus with intestinal perforation and underwent a therapeutic Ladd procedure with ileostomy. The second patient had a fluoroscopy study in infancy that was not suggestive of malrotation. He underwent the three staged palliation for single ventricle and presented at age 5 with emesis. A CT scan at that time demonstrated malrotation without volvulus and he underwent a therapeutic Ladd procedure given his gastrointestinal symptoms. No volvulus was found by direct inspection in the operating room.

3.3. Institutional experience with malrotation

Our patient experience is depicted in Fig. 1 with in-hospital mortality following cardiac surgery shown based on intestinal anatomy and operative intervention of malrotation. Of the 29 patients with known malrotation, 19 (66%) underwent a prophylactic Ladd procedure and two (7%) patients had a therapeutic Ladd procedure. 4 (20%) of patients who underwent prophylactic Ladd procedure had a gastrostomy tube placed during same operation. Of the 19 patients who underwent prophylactic Ladd, 10 (52%) had the Ladd procedure following cardiac surgery, 5 of these patients had a Blalock-Taussig shunt at the time of prophylactic Ladd. One patient who underwent prophylactic Ladd procedure suffered respiratory arrest following surgery necessitating ECMO for 4 days but ultimately survived to hospital discharge. At a median follow up time of 2.4 years, 47/49 (96%) of patients were volvulus-free.

3.4. Comparison of patient demographics and outcomes

Table 2 describes the mean gestational age, mean birth weight, and mean age at cardiac surgery of our patients who underwent cardiac surgery and differences in outcomes based on each variable. There was a significant difference in the mean birth weight between those patients who survived to hospital after cardiac surgery and those who did not, 3.10 versus 2.39 kg ($p = 0.02$). There was no difference in the gestational age or age at cardiac surgery between the two groups.

Overall mortality after cardiac surgery in patients with single ventricle anatomy was 6/41 (14%) compared with 0/8 (0%) in biventricular patients which was not statistically different ($p = 0.571$). Mortality

Table 1
Characteristics of patients with heterotaxy syndrome.

Patient characteristic	N (%) or mean
Male	24 (49%)
Gestational age (weeks)	38
Age at cardiac surgery (months)	0.58
Birth weight (kg)	3.01
TAPVR	18 (37%)
- Obstructed	5 (28%)
Pulmonary atresia	10 (20%)
Single ventricle	42 (86%)
Splenic anatomy	
- Asplenia	28 (57%)
- Polysplenia	16 (32%)
- Single spleen	4 (8%)

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