

Introduction

Background:

Heterotaxy Syndrome (HS) is a disorder that develops in utero characterized by abnormal positioning of the organs. As a result of this abnormal development, many patients with HS are born with severe congenital heart disease (CHD) with single ventricle physiology. In addition, most also have intestinal malrotation placing them at increased but undefined risk for catastrophic midgut volvulus. The timing and sequence of surgical intervention in this challenging group of patients remains undefined.

Significance:

Historical data suggests repair of asymptomatic intestinal malrotation in normal infants (NHS) at the time of diagnosis is safe and effective. However, this strategy has not been validated in infants with severe CHD who are at increased risk for procedural complications given their tenuous single ventricle physiology. It is likely that the procedural risks in infants with CHD and single ventricle physiology would lower following second stage cardiac surgical palliation.

Research Question:

This study sought to determine whether deferring the Ladd procedure in asymptomatic infants with intestinal malrotation and HS during the first six months of life is safe.

Methods

Medical records of patients with intestinal malrotation with or without HS at Phoenix Children's Hospital from 2006-2011 were reviewed. Severity of heart disease, diagnosis of intestinal malrotation, and timing of Ladd procedure if applicable were recorded. Incidence of volvulus, morbidity of volvulus, and morbidity of an elective Ladd procedure were studied in both the HS and Non-HS population.

Results

	Heterotaxy Syndrome (n = 31)	Non-Heterotaxy Syndrome (n = 109)
Ladd procedure <6 months of age, no volvulus	7	34
Ladd procedure <6 months of age, with volvulus	2	18
No Ladd procedure <6 months age	22	57
Total	31	109

Table 1: Results from data collection

Demographics

Characteristic	HS received elective Ladd procedure prior to six months	HS did not receive Ladd procedure prior to 6 months	NHS received elective Ladd procedure prior to six months
% Male	57% (4 males, 3 females)	27% (6 males, 16 females)	56% (19 males, 15 females)
Mean Birth weight (kilograms)	3.284	3.150	2.498
Mean Weight last visit before six months (kilograms)	4.984	6.151	4.953
Mean Age at last visit before six months (months)	4.250	4	4
Mean O2 sat at last visit before six months	87%	85%	98%

Table 2: Demographics of patients in the study

Statistics

Incidence of volvulus in HS No Ladd Procedure	8.3% (2/24)
Incidence of volvulus in NHS No Ladd Procedure	24.0% (18/75)
Relative Risk of volvulus NHS vs HS	3.47 (p-value = 0.089)

Table 3: Statistical analysis of results

Results

31 patients with Heterotaxy Syndrome and intestinal malrotation were identified. Of the 31, 9 had a Ladd procedure prior to six months of age. 2 out of the 9 were for volvulus and the other 7 were done either electively or for less severe GI symptoms that were not suggestive of volvulus. The other 22 did not have a Ladd procedure prior to six months of age. There was one death (1/22) from a non-gastrointestinal cause in a patient who had not undergone a Ladd procedure. There were no deaths in the 9 patients who underwent a Ladd procedure (0/9).

109 patients without Heterotaxy Syndrome but with intestinal malrotation were identified. 34 did not have a volvulus but rather had an elective Ladd procedure before 6 months of age. 18 had an emergent Ladd procedure before 6 months of age. 57 with known intestinal malrotation did not have a Ladd procedure before 6 months of age.

Discussion and Conclusions

One of the most surprising findings from this study was the decreased incidence of volvulus in HS patients as compared to NHS patients. This finding has been mentioned in the literature but has yet to be fully elucidated. Due to the small number of HS patients undergoing the elective Ladd procedure this study was unable to demonstrate any increase or decrease of mortality with elective surgery in the first 6 months of life.

However, given the low overall incidence of volvulus in HS, and with continued vigilance for obstructive symptoms, **this study suggests that delaying a Ladd procedure in asymptomatic patients with HS and CHD and intestinal malrotation is safe.** Watchful waiting may reduce the incidence of cardiac complications during the Ladd procedure by allowing for stabilizing cardiac surgical palliation prior to elective abdominal surgery.

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