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Typical and Atypical Malrotation and the Need For Elective Ladd's Procedure In Patients With Heterotaxy Syndrome

Study Purpose and Rationale:

Heterotaxy syndrome is a complex congenital heart disease characterized by the disorder of abdominal and thoracic organs that are abnormally arranged across the left-right-axis of the body [1,7]. Because of the involvement of multiple organ systems, many extracardiac abnormalities coincide, particularly intestinal malrotation that can potentially have severe complications including midgut volvulus [2]. Based on literature, patients who have abdominal symptoms such as bilious vomiting, feeding intolerance, or abdominal distention are worrisome for midgut volvulus and should undergo emergent surgical treatment [3]. However, controversy arises when asymptomatic patients are found to have intestinal malrotation incidentally, and then undergo elective prophylactic operations, specifically the Ladd's procedure.

There have been numerous methods in classifying and diagnosing intestinal malrotation. Because the anatomic variations can present in a wide spectrum, however, it has been difficult to classify. Although the typical malrotation has been documented well, atypical malrotation is still not yet clearly defined. In a study looking at the classification of typical and atypical malrotation, McVay et al described classifying malrotation based on upper gastrointestinal contrast studies and particularly looking to the location of the Ligament of Treitz and the cecum [6]. Typical was defined as the Ligament of Treitz (LOT) located to the right of midline or non-existent, where as atypical malrotation was defined as the LOT located at or to the left of the midline and below the level of the pylorus. This specific subset of patients showing atypical malrotation is at lower risk of severe complications such as midgut volvulus and may not necessarily need a surgical intervention such as Ladd's procedure [2].

The Ladd's procedure involves lysis of peritoneal bands coursing through the right upper quadrant down to the cecum, reduction of any midgut volvulus, broadening the mesentery and rearrangement of the small and large bowels, and an appendectomy [4]. It has been documented that there is significant post-operative complication risk in asymptomatic patients who undergo an elective Ladd's procedure: 14% risk of adhesive small bowel obstruction [5] and 9.7% in-hospital mortality due to post-operative complications [3]. The question then arises does performing a Ladd's procedure for asymptomatic malrotation with heterotaxia benefit the patient in the setting of their complicated heart disease.

Hypothesis:

Our hypothesis is that heterotaxy patients who undergo positive screening for malrotation by an upper gastrointestinal series and classified as atypical malrotation based on anatomic criteria may have a higher post-operative complication rate from an elective Ladd's procedure when compared to typical malrotation.

Methods:

Study Design: This will be a retrospective review looking at a 10-year period between 2001-2010. Patients with heterotaxy syndrome will be identified by cross referencing two separate databases from the cardiology and pediatric surgery department and identified further as those who were positively screened for intestinal malrotation by upper GI series. At our institution, the practice is to perform the Ladd's procedure on all patients positively screened regardless of symptomology. Therefore, two cohorts will be formed based on reviewing the upper GI series by our pediatric radiologists to locate the Ligament of Treitz (LOT) and categorize the malrotation as either typical or atypical. The first will be patients who's LOT is located right of the midline or non-existent defined as typical and the second will be those who's LOT is at or left of the midline defined as atypical. The location of the cecum will be documented, however will not be used to classify in this study. Hospital medical records, operative reports, echocardiogram reports, discharge summaries, and follow up outpatient clinic notes will be reviewed to discuss the type of cardiac disease, mortality rate, and post-operative complications follow the Ladd's procedure in a 1-year period.

Inclusion/Exclusion Criteria: To be included in the study, patients must have the diagnosis of heterotaxy syndrome and have one or more radiographic findings of intestinal malrotation and subsequently undergone a Ladd's procedure regardless of presentation of symptoms prior to the procedure during the time period of interest. Patients will be excluded if incomplete records are found, or if intestinal malrotation was found concomitant with other conditions predisposing malrotation (i.e. congenital diaphragmatic hernia, gastroschisis, or omphalocele).

Statistical Analysis: Statistical Analysis will be performed using software such as SPSS and Microsoft Excel to measure incidence, demographics, phenotype, outcome, and post-operative complications. A chi-squared test will be used to compare the complication rate with typical and atypical malrotation. A Kaplan-Meier analysis will be used to compare the each type of intestinal malrotation and overall survival rate of heterotaxy patients.

Sample Size: In a study to classify the post-operative complication of Ladd's procedure between typical and atypical malrotation (heterotaxy patients were excluded), it was found that typical had a 16% complication rate and atypical had 27% [2]. To compare the increase in post-operative complication rate within each individual group (typical and atypical) with the overall complication rate of heterotaxy patients (~65%) [1], using a chi-square test to achieve an appropriate sample size, we would need between 42-150 patients in each group for statistical significance.

Miscellaneous:

Recruitment of Subjects: Not applicable.

Study Drugs: Not applicable.

Medical Device: Not applicable.

Study Questionnaires: Not applicable.

Confidentiality of Subject Data:

All patient data and medical records will be kept confidential and maintained on password-

protected computers and secure storage media. Patient records will be assigned a unique identifying number and all identifying data will be securely discarded once study is over.

Potential Conflict of Interest: Not applicable.

Location of Study: New York Presbyterian Hospital Columbia University

Potential Risks: Not applicable.

Potential Benefits: No direct benefit to patients being studied.

Alternative Therapies: Not applicable

Compensation to Subjects: Not applicable

Costs to the Subjects: Not applicable

Minors as Research Subjects: Not applicable

Radiation or Radioactive Substances: Not applicable

References:

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6: Mehall JR, Chandler JC, Mehall RL, Jackson RJ, Wagner CW, Smith SD. Management of typical and atypical intestinal malrotation. *J Pediatr Surg*. 2002 Aug;37(8):1169-72. PubMed PMID: 12149695.

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