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Operative outcomes of infantile hypertrophic pyloric stenosis in patients with congenital heart disease

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ABSTRACT

Purpose: This study aims to compare the outcomes of pyloromyotomy for infantile hypertrophic pyloric stenosis (IHPS) in children with and without congenital heart disease (CHD).
Methods: A retrospective, single pediatric center, case–control, matched cohort study was performed over 10 years. A case of IHPS with CHD was paired with control patients of IHPS without CHD, matched by age and gender. Perioperative morbidity, 30-day mortality, length of hospital stay, and hospital cost were compared. Subgroups were analyzed based on the severity of CHD and the reason for admission.
Results: Twenty-six patients who underwent pyloromyotomy for IHPS with CHD (CHD group) were matched with 78 patients with IHPS without CHD (Non-CHD group). No 30-day mortality was identified in either group. Overall perioperative complications were not significantly different between groups (11.5% vs 5.2%, $p = 0.163$). However, postoperative length of stay was longer in CHD group (6 vs 1 days, $p < 0.001$) and any subgroups of CHD as compared to Non-CHD group. CHD group patients admitted only for IHPS had short postoperative LOS, whereas those who developed pyloric stenosis during a hospital admission stayed longer postoperatively (1.5 vs 26.5 days, $p < 0.001$).
Mean hospital costs in patients admitted for IHPS were $16,270 and $3591 for CHD group and Non-CHD group, respectively ($p < 0.001$).
Conclusions: IHPS patients with CHD have prolonged postpyloromyotomy course, especially when inpatients with CHD incidentally develop IHPS.

Infantile hypertrophic pyloric stenosis (IHPS) is a common pediatric condition, with an incidence reported of 1 to 8 per 1000 live births [1]. The current standard of care is operative pyloromyotomy. Though the route of access to the pylorus varies between techniques, the anesthetic and operative management are otherwise ritualized. Pyloromyotomy, longitudinal splitting of the hypertrophic pyloric muscle, was initially reported by Ramstedt in 1911 [2]. It is considered a relatively simple procedure with an excellent outcome. However, it is not risk free. Hulka et al. reported a 0.1% mortality rate, a 4% incidence of intraoperative complications, and a 6% incidence of postoperative complications [3]. These complications included bleeding, mucosal perforation and wound infection. Additionally, a study by Walker et al. showed that infants who had pyloromyotomy had significantly lower cognitive, receptive language, fine motor, and gross motor skills when compared to healthy control infants, suggesting potential harm associated with the surgical procedure or general anesthesia [4].

Congenital heart disease (CHD), defined as (a) defect(s) in the structure of the heart and great vessels which is (are) present at birth, may represent a population where standard surgical approaches are associated with even higher risk when CHD coincide with IHPS. Patients with CHD are known to have significantly higher perioperative risk of complications for noncardiac surgery in general [5-7]. Nevertheless, current practice remains the performance of pyloromyotomy, though little is known about the perioperative risks in this patient population. Children with CHD may be particularly susceptible to certain anesthetic complications, intraoperative and postoperative cardiac dysfunction, and may have poorer ability to tolerate laparoscopic or open abdominal operations.

Precedent exists for nonoperative management of IHPS. Atropine sulfate has been utilized as the primary treatment for IHPS in some Asian and European countries [8–19]. Based on the aggregated data from ten studies pooled in a metaanalysis, conservative therapy with atropine works in 88% of cases without recurrence [20]. Notably, there were no episodes of consequential impact upon cardiac function or any other significant complications.

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Table 1
Congenital Heart Disease Category.

Category	Diagnosis
1	AVCD, TOF, univentricular heart, transposition complex, truncus arteriosus, hypoplastic left heart syndrome
2	Aortic coarctation, Ebstein's anomaly, ASD, VSD, PDA
3	Unspecified defect of septal closure
4	Anomalies of the pulmonary artery/valve, mitral stenosis/insufficiency
5	Other unspecified anomalies of the heart/aorta

AVCD: Atrioventricular Canal Defect.
TOF: Tetralogy of Fallot.
ASD: Atrial Septal Defect.
VSD: Ventricular Septal Defect.
PDA: Patent Ductus Arteriosus.

The purpose of the current study was to assess the perioperative risks, length of stay (LOS), and cost in IHPS patients with CHD as compared to those of Non-CHD patients. We hypothesize that CHD children with IHPS will experience a significantly increased rate of surgical complications and/or longer LOS when compared to case-match controlled Non-CHD children with IHPS.

1. Materials and methods

This is a retrospective, single pediatric center, matched case–control study to evaluate the perioperative morbidity and mortality in patients with CHD and IHPS who underwent laparoscopic or open pyloromyotomy during the study period 2005–2014. Patients with both IHPS and CHD were identified by ICD-9 codes within our patient care database systems. A case of IHPS with CHD was randomly paired with three control patients of IHPS without CHD, matched by postnatal age and gender. Fourteen days of difference (younger and older) was allowed for age matching. Any intraoperative morbidities (e.g. dysrhythmias, hypotension, hypoxemia, vasopressor/inotrope requirements, need for blood product transfusions, cardiopulmonary resuscitation, abortion of the procedure), postoperative morbidities (e.g. bleeding, wound infection, incomplete pyloromyotomy, mucosal perforation, pneumonia, stroke), length of mechanical ventilation, length of postoperative/overall hospital stay, 30-day mortality, and hospital cost in IHPS with CHD were evaluated and compared against controls. Hospital cost was obtained from our institutional billing department database, and was defined as total cost associated with direct patient care (i.e. labor and direct material).

CHD is defined as (a) defect(s) in the structure of the heart and great vessels which is (are) present at birth. Such structural defects include hypoplastic left/right heart syndrome, transposition of the great arteries, double outlet right ventricle, tetralogy of Fallot, pulmonary atresia, atrioventricular canal defect, aortic stenosis, tricuspid atresia, pulmonary stenosis, truncus arteriosus, Ebstein's anomaly, coarctation of the aorta, atrial septal defect, ventricular septal defect, and patent ductus arteriosus.

Categories of CHD as proposed by Marelli et al. were used to select patients [21]. Among five categories proposed, the two most severe forms of CHD (Categories 1 and 2) were included in the study (Table 1).

For purposes of subgroup analyses, Category 1 and Category 2 CHD were individually evaluated against control using 1:3 case–control matching method. Furthermore, CHD group was also divided into two groups based upon the reason for admission; children who were admitted for IHPS, or those who were already admitted for other reasons when they developed IHPS.

Descriptive statistics were calculated for all variables. The p values for categorical variables were derived from Pearson's chi-square test or Fisher's exact test. For continuous variables Mann–Whitney U test or independent t test was deployed, as appropriate. A p value <0.05 was considered significant. Values are reported as mean $\pm$ standard deviation (SD) for continuous variables with normal distribution, as median with interquartile range for continuous variables without normal distribution, and as percentages for categorical variables. IBM SPSS Statistics version 22 was used for database management and analyses (SPSS Inc., Chicago, IL).

2. Results

Twenty-six patients who underwent pyloromyotomy for IHPS in patients with CHD were identified (CHD group). As a result of 1:3 matching, 78 patients with IHPS without CHD (Non-CHD group) were paired with each study patient (Fig. 1).

The clinical characteristics between CHD and Non-CHD groups are shown in Table 2. No 30-day mortality or intraoperative morbidity was identified in either group (Table 3). While mucosal perforation was more common in CHD group (11.5% vs 0%, $p = 0.014$), the difference in overall perioperative complications between groups did not reach statistical significance (11.5% vs 5.2%, $p = 0.163$). Median LOS was significantly longer in CHD group (overall 11.5 vs 3 days, and postoperative 6 vs 1 days, $p < 0.001$). The results were similar when CHD were divided into Category 1 and Category 2 (Tables 4a and 4b). CHD patients admitted for IHPS had brief postoperative LOS, whereas those who developed pyloric stenosis during a hospital admission stayed longer postoperatively (1.5 vs 26.5 days, $p < 0.001$, Table 5). Hospital costs were higher in CHD groups, regardless of the CHD category.

3. Discussion

Although our results showed that CHD patients have slightly higher risk of mucosal perforation, the difference between CHD and Non-CHD groups in overall perioperative risk did not reach statistical significance. However, LOS was longer in CHD groups regardless of CHD category. Notably, when inpatients with CHD incidentally developed IHPS, their postoperative LOS from pyloromyotomy was significantly longer. While the longer LOS and higher cost in these patients are almost certainly attributable to their baseline cardiac conditions and/or the

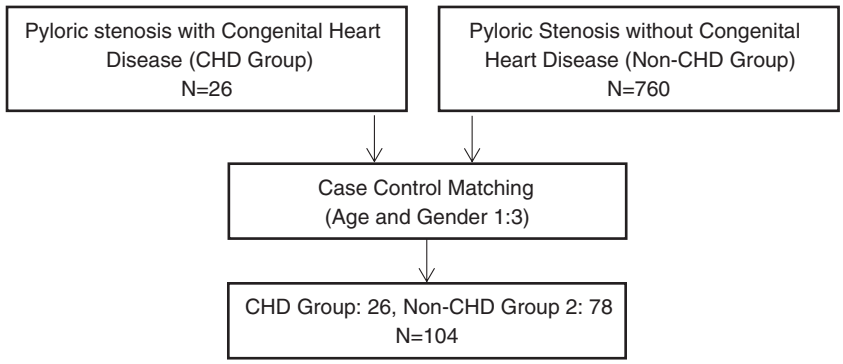

Fig. 1. Patient Selection.