



Contents lists available at ScienceDirect

Journal of Cardiothoracic and Vascular Anesthesia

journal homepage: www.jcvaonline.com

Original Article

Exploring Noncardiac Surgical Needs From Infancy to Adulthood in Patients With Congenital Heart Disease



A. Rebecca L. Hamilton, MD, MSc^{*,†,‡,1},
Kirsten C. Odegard, MD, MBA^{*,†}, Koichi Yuki, MD^{*,†}

^{*}Division of Cardiac Anesthesia, Department of Anesthesiology, Critical Care and Pain Medicine, Boston Children's Hospital, Boston, MA

[†]Department of Anaesthesia, Harvard Medical School, Boston, MA

[‡]Department of Cell and Molecular Biology, Karolinska Institute, Stockholm, Sweden

Objectives: As life expectancy for patients born with congenital heart disease (CHD) continues to rise, these patients will present increasingly for noncardiac surgery during childhood and adolescence. This study aimed to map the lifespan of noncardiac surgical needs among patients with CHD and explore how these needs may change over time.

Design: All patients with CHD presenting for noncardiac surgery between 2008 and 2014 were selected for review.

Setting: The study was conducted at a single urban academic tertiary pediatric hospital.

Participants: All patients with CHD presenting for noncardiac surgery during the study period were included and grouped by cardiac diagnosis.

Interventions: Descriptive analysis included patient demographics, CHD diagnosis, procedures performed, and clinical data, including baseline saturation and underlying cardiac function.

Measurements and Main Results: A total of 3,011 noncardiac surgical procedures were performed on patients with CHD during the study period. The most common CHD diagnoses were patent ductus arteriosus (27.6%), ventricular septal defects (24.7%), and patent foramen ovale (24.3%). The median age was 4 years, 87% of all the patients were ≤ 10 years, and 41% had associated syndromes. Of the patients, 76% underwent a pre-operative echocardiogram, and 10% had depressed cardiac function at the time of surgery. The most common procedures performed were ear, nose, and throat (20%), general surgery (14%), and radiology (11%). Intraoperative events were reported in 488 out of 3,010 encounters (16.2%), with the highest rates reported in patients with single-ventricle physiology (55/179; 30.7%).

Conclusions: These findings suggested a greater burden of noncardiac surgery in lower age groups, with ear, nose, and throat and general surgery most common in young children and orthopedic and dental procedures increasing in adolescence.

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Key Words: congenital heart disease; surgical outcomes; surgical planning; single ventricles; noncardiac surgery; pediatric surgery

CONGENITAL HEART DISEASE (CHD) is the most common group of birth anomalies, with a prevalence of 4-to-10 per 1,000 live births.¹ Although complex forms of CHD historically were considered fatal, >90% of patients now survive to adulthood, and, as a result, this patient population is presenting increasingly for noncardiac surgery during childhood and

adolescence.^{2,3} Previous studies have established that the presence of CHD is associated with a higher risk of morbidity and mortality in patients undergoing noncardiac surgery compared to patients without CHD.^{4,5} Considering this elevated risk, a number of recommendations on how to safely anesthetize patients with specific types of CHD undergoing noncardiac surgery have been proposed, emphasizing the importance of comprehensive perioperative and multidisciplinary planning for reducing morbidity and mortality in patients with CHD.^{6,7} Long-term planning for the surgical needs of these complex patients could benefit the individual patient, as well as improve

¹Address correspondence to A. Rebecca L. Hamilton, MD, MSc, Department of Anesthesiology, Critical Care and Pain Medicine, Cardiac Anesthesia Division, Boston Children's Hospital, 300 Longwood Avenue, Boston, MA 02115.

E-mail address: rebecca.hamilton@ki.se (A.R.L. Hamilton).

healthcare processes and overall healthcare expenditure. However, the timing and need of different types for noncardiac surgical procedures associated with specific cardiac lesions have not yet been explored fully. This study aimed to delineate any association between various CHD lesions and noncardiac surgical procedures to facilitate surgical planning and access to care for this vulnerable patient population.

Methods

After Institutional Review Board approval (BCH M10-08-0407), all encounters for noncardiac surgical procedures in patients with CHD performed between 2008 and 2014 at the authors' institution were selected for review. Data collection ended in 2014 due to a change in the anesthesia record system no longer allowing tracking of data the same way as before. Patients were categorized into cohorts based on cardiac diagnosis. The analysis included patient demographics, underlying CHD lesion, and the type of noncardiac surgical procedures performed. Data were collected on clinical factors such as the availability of preoperative echocardiogram within 6 months of the procedure, cardiac dysfunction (defined as an abnormal ventricular function or compromised cardiac output), baseline preoperative saturation levels, postoperative saturation, and any intraoperative events. Intraoperative events were defined as an occurrence of any of the following: cardiovascular events, including cardiac arrest, significant arrhythmias, significant hypotension requiring inotropic support, pulmonary hypertensive crisis, difficult intubation, loss of airway requiring reintubation, bronchospasm, laryngospasm, desaturation, stroke, seizure, hemorrhage requiring transfusion, and difficult access. Patients with underlying single-ventricle physiology were divided into cohorts based on their staging along the palliative pathway for further sub-group analysis.

The categorical variables were presented as number and percentage; the continuous variables were presented as mean and SD for normally distributed variables or median and IQR for variables with skewed distribution. Normality was examined using the Shapiro-Wilk normality test. All data were analyzed using 2021 Excel for Mac version 16.54 (Microsoft, Corp, Redmond, WA).

Results

A total of 3,010 surgical encounters between 2008 to 2014 were reviewed. The median age was 4 years (range 0–45 years), and the median weight 15.45 kg (range 0.82–143 kg). Most patients were categorized as an American Society of Anesthesiologists physical status classification system III; 70 out of 3,010 (2.3%) patients were neonates, and 1,234 out of 3,010 (41.0%) were reported to have an associated syndrome. Just over half of all patients (1,623/3,010; 53.9%) had received some cardiac intervention prior to their noncardiac surgery, whereas 1,387 out of 3,010 (46.1%) patients presented with unrepaired cardiac defects. The patient demographics are presented in [Table 1](#).

Table 1
Demographics of All Patients

Demographics (N = 3,010)	
Age, median (range), mo	48 (0.03–540)
Weight, median (range), kg	15.45 (0.82–143)
Median ASA, range	3 (1–5)
Neonates, n/N (%)	70/3010 (2.3)
Associated syndrome, n (%)	1234 (41.0)
History of cardiac surgery, n (%)	1623 (53.9)
Single ventricle physiology, n (%)	196 (6.5)
Preop echocardiogram available, n (%)	2284 (75.9)
Cardiac consult prior to noncardiac surgery, n (%)	536 (17.8)
Cardiac dysfunction, n (%)	228/2284 (10.0)
Preop saturation, median (range), %	100 (65–100)
Postop saturation, median (range), %	100 (59–100)
Intraoperative events, n (%)	488 (16.2)

Abbreviation: ASA, American Society of Anesthesiologists.

A total of 2,284 out of 3,010 (75.9%) patients presented with a preoperative echocardiogram obtained within 6 months of their surgical procedure. Of these 2,284 echocardiograms, 228 (10%) reported findings consistent with underlying cardiac dysfunction. The preoperative and postoperative ranges of oxygen saturation varied in the authors' patients according to cyanotic or noncyanotic cardiac disease.

Intraoperative events were defined as cardiovascular events (eg, arrhythmias, arrest, profound decrease in cardiac output), cerebrovascular events (eg, stroke, seizures), airway complications (eg, difficult intubation, reintubation, bronchospasm), surgical events (eg, massive hemorrhage, difficult access) and adverse events. A total of 488 out of 3,010 encounters (16.2%) reported at least 1 intraoperative event. The intraoperative event rate was 74 out of 229 (22.5%) in patients with a preoperative echocardiogram demonstrating abnormal ventricular function, 328 out of 2,067 (15.9%) in patients with a preoperative echocardiogram demonstrating a normal biventricular function, and 86 out of 715 (12.0%) in patients without a preoperative echocardiogram. The average age of patients with intraoperative events was higher than the overall cohort (82.56 v 71.52 months, CI [3.827–18.26], $p = 0.0027$). Events were more common in patients who had received some form of cardiac surgery intervention prior to their noncardiac surgery compared to patients with unrepaired cardiac defects (19.1% v 12.8%). Moreover, adverse events occurred more frequently in the adult population (52/167; 31.1%) compared to patients <18 years of age (436/2,843; 15.3%).

The most common intraoperative events were cardiovascular events requiring inotropic support (70.5%; 344/488). Other relatively common events included bronchospasm and/or laryngospasm (12.7%; 62/488), difficult intubation (8.8%; 43/488), reintubation and/or failed extubation (6.1%; 30/488), and difficult intravenous access (7.0%; 34/488). Rare events included cardiac arrest (1.2%; 6/488), drug error (0.8%; 4/488), failed sedation (0.6%; 3/488), cerebrovascular events (0.4%; 2/488), and pulmonary hypertensive crisis (0.2%; 1/488).

The CHD lesions most frequently encountered in this cohort are presented in [Table 2](#). A total of 832 out of 3,010 (27.6%)

Table 2
Noncardiac Surgeries by Cardiac Diagnosis

Cardiac Diagnosis (N = 3,010), n (%)	No. of Procedures Performed
Pulmonary hypertension	90 (3)
Patent foramen ovale	732 (24.3)
Patent ductus arteriosus	832 (27.6)
Atrial septal defects	797 (26.4)
Ventricle septal defects	745 (24.8)
Atrioventricular canal defects	256 (8.5)
Hypoplastic left heart syndrome	94 (3.1)
Tricuspid atresia	33 (1.1)
Tetralogy of Fallot	293 (9.7)
Transposition of the great arteries	88 (2.9)
Coarctation	205 (6.8)
Truncus	13 (0.4)
Double outlet RV/LV	91 (3.0)
Pulmonary stenosis	209 (6.9)
Aortic valve stenosis	148 (4.9)

Abbreviations: AS, aortic valve stenosis; ASD, atrial septal defects; AVCD, atrioventricular canal defects; CoA, coarctation; DORV/DOLV, double-outlet right ventricle/left ventricle; HLHS, hypoplastic left heart syndrome; PDA, patent ductus arteriosus; PFO, patent foramen ovale; PHTN, pulmonary hypertension; PS, pulmonary stenosis; TA, tricuspid atresia TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VSD, ventricular septal defects.

patients presented with a history of patent ductus arteriosus; 732 out of 3,010 (24.3%) with patent foramen ovale (PFO); 794 out of 3,010 (26.4%) with atrial septal defects; and 745 out of 3,010 (24.8%) with ventricular septal defects (VSD). The PFO group included patients presenting with PFOs in isolation (179/732; 24.5%) and patients with PFOs presenting in combination with other cardiac defects (553/732; 75.5%).

A total of 3,381 noncardiac surgical procedures were performed >3,010 encounters, which are listed in Table 3. The most common types of procedures were ear, nose, and throat (ENT) procedures such as tonsillectomies and myringotomy tube placements (677/3,381; 20.0%); followed by general surgery procedures, such as exploratory laparotomy, hernia

Table 3
Types of Noncardiac Surgical Procedures Performed

Surgical Procedure (N = 3,381), n (%)	No. of Procedures Performed
ENT	677 (20.0)
GI	259 (7.7)
Direct laryngoscopy/bronchoscopy	278 (8.2)
General surgery	475 (14.0)
Orthopedic surgery	221 (6.5)
Spine surgery	123 (3.6)
Urology	181 (5.3)
Radiology	383 (11.3)
Dental/OMFS	156 (4.6)
Neurosurgery	81 (2.4)
Ophthalmology	88 (2.6)
Line placement	197 (5.8)
Bone marrow biopsy/lumbar puncture	77 (2.3)
Others	185 (5.5)

Abbreviations: DLB, direct laryngoscopy/bronchoscopy; ENT, ear, nose, and throat; GI, gastrointestinal; OMFS, oral and maxillofacial surgery.

repairs, and appendectomies (475/3,381; 14.0%); radiology procedures (383/3,381; 11.3%); airway procedures, including direct laryngoscopy and bronchoscopy (278/3,381; 8.2%); and gastrointestinal (GI) procedures (259/3,381; 7.7%).

The distribution of surgical procedures among the 10 most common CHD cohorts is presented in Figure 1. Radiology procedure, ENT surgery, and general surgery were common across all types of CHD.

The age distribution for different noncardiac surgical procedures in patients with CHD is presented in Figure 2. The median age at presentation in all combined cohorts was 4 years. Most patients were between 0 and 5 years (68%), followed by 6-to-10 years (19%), 11-to-15 years (10%), and >15 years (8%). This age distribution was similar in patients with VSD, atrioventricular septal defects, tetralogy of Fallot, and hypoplastic left heart syndrome. In the transposition of the great arteries (TGA) cohort, surgery was more common in the 6- to 10-year age group and weighted toward lumbar procedures (bone marrow biopsies [BMBs], lumbar punctures [LP], intrathecal chemotherapy), and line placements.

Patients with CHD resulting in single-ventricle physiology were selected for further analysis and separated based on palliative staging, as presented in Figure 3. During the study period, 179 patients presented with single-ventricle physiology at some point along their palliation pathway—17 patients presented after a Norwood (stage 1) procedure, 37 presented with a bidirectional Glenn (superior cavopulmonary connection), 92 presented following a Fontan procedure (total cavopulmonary connection), 6 following a biventricular repair, and 20 patients presented after an orthotopic heart transplant.

General surgery, ENT, and GI procedures were most common in this single-ventricle cohort. General surgery procedures were primarily gastrostomy tube placements in younger patients following the Norwood and bidirectional Glenn procedures, and Ladd's band takedowns for malrotation and hernia repairs in older patients following Fontan. Ophthalmology procedures were more common among premature patients in the Norwood cohort and in patients following biventricular repair. Transplant patients presented primarily for radiology procedures and, to a lesser extent, for surgical procedures compared with the other cohorts. Intraoperative events occurred in 55 out of 179 encounters (30.7%).

Discussion

The aim of this study was to evaluate noncardiac surgical needs in patients with CHD from early childhood into adulthood. The results suggested that the need for noncardiac surgery in patients with CHD is greater in the younger age groups, with ENT and general surgery procedures dominating in toddlers and young children, whereas orthopedic and dental procedures increase in adolescence and into adulthood. General surgery procedures were more common in the hypoplastic left heart syndrome and TGA cohorts compared to VSD, Tetralogy of Fallot, and atrioventricular canal defects. Urology procedures and BMBs and/or LPs frequently occurred in the TGA cohort, especially among the older age groups; however,

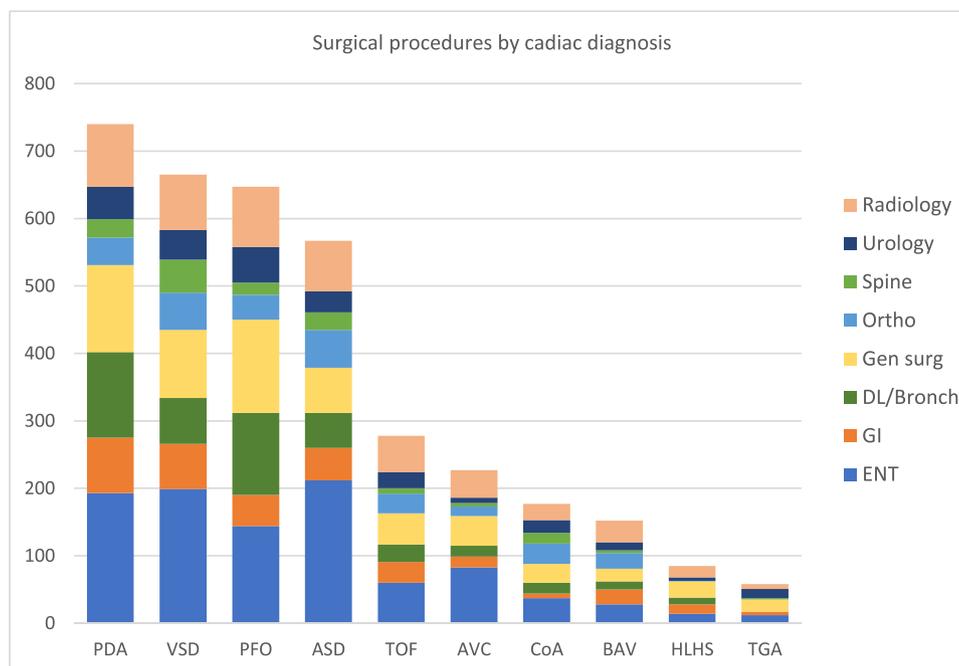


Fig 1. Distribution of surgical procedures in the most common cardiac patient groups. ASD, atrial septal defects; AVC, atrioventricular canal; BAV, Bicuspid aortic valve; CoA, coarctation; DLB, direct laryngoscopy/bronchoscopy; ENT, ear, nose, and throat; GI, gastrointestinal; HLHS, hypoplastic left heart syndrome; PDA, patent ductus arteriosus; PFO, Patent foramen ovale; TOF, tetralogy of Fallot; TGA, transposition of the great arteries; VSD, ventricular septal defects;

a further subgroup analysis revealed that many of the BMBs and LPs stemmed from 1 patient with an underlying hematologic condition that skewed some of the data in this particular patient group.

This study offered a novel perspective on surgical needs over time among patients with CHD. Prior studies on noncardiac surgical needs in patients with CHD largely have been limited to younger age groups. In 2014, Sulkowski et al. looked at patients with CHD across multiple institutions up to the age of 5, and found that 40% of all patients would undergo noncardiac procedures within 5 years of life, whereas general surgery and ENT accounted for 60% of all procedures performed.⁸ The wider cohort of patients presented here followed a similar trend, with ENT, GI, and general surgical procedures dominating amongst younger patients, with a noticeable shift toward orthopedic and dental procedures in early adolescence. Although most patients (68%) were ≤ 5 years, patients in the TGA cohort tended to be older than the average, with more than half of procedures performed after 5 years of age.

Congenital heart disease is a known risk factor for intraoperative events during noncardiac surgery, and patients with single-ventricle physiology are associated with an even higher incidence of complications.⁹⁻¹⁴ When looking at all patients with CHD together, the intraoperative event rate in the authors' overall study cohort was 16.2% (488/3,010). Although the authors' definition of intraoperative events was purposely left broad to include all forms of cardiovascular events, airway complications, failed sedation, and other adverse events, the incidence of intraoperative events occurred at a markedly higher rate than typically reported in the general pediatric literature.^{10,15-19} Interestingly, intraoperative events were more common in patients who had received some form of cardiac

intervention prior to their noncardiac surgery, likely reflecting a higher cardiac risk category amongst patients receiving early interventions.^{4,20} Adverse events also occurred more frequently in the adult patient population compared to the pediatric population, although this may have been somewhat skewed by self-selection and a smaller proportion of adult patients included in the overall study cohort. The rate of intraoperative events was higher in the cohort of patients with known ventricular dysfunction leading up to surgery compared to patients with a preoperative echocardiogram and normal biventricular function, and lowest in the group presenting for surgery without a preoperative echocardiogram, likely reflecting a good baseline function with a low reason for perioperative concerns. As expected, patients with single-ventricle physiology demonstrated the highest rate of intraoperative events.

Ambulatory surgical services is a rapidly expanding field in both pediatric and adult settings, and with >90% of children born with CHD today surviving to adulthood, it is expected that the number of patients with CHD presenting for ambulatory surgery will only continue to increase.²¹ Although the rate for unexpected hospital admission is generally low in the pediatric population, typically cited at around 1%,²² the admission rates for patients with CHD undergoing noncardiac surgery have been found to be significantly higher at 2.7%.²³ As a result, preoperative planning strategies become of uttermost importance for this vulnerable patient population in order to reduce perioperative complications. The authors' results demonstrated that the types of surgical needs vary across age groups and types of underlying CHD lesions and are not sufficiently categorized by the severity of underlying disease as suggested by functional status, American Society of Anesthesiology classification, and other significant comorbidities.

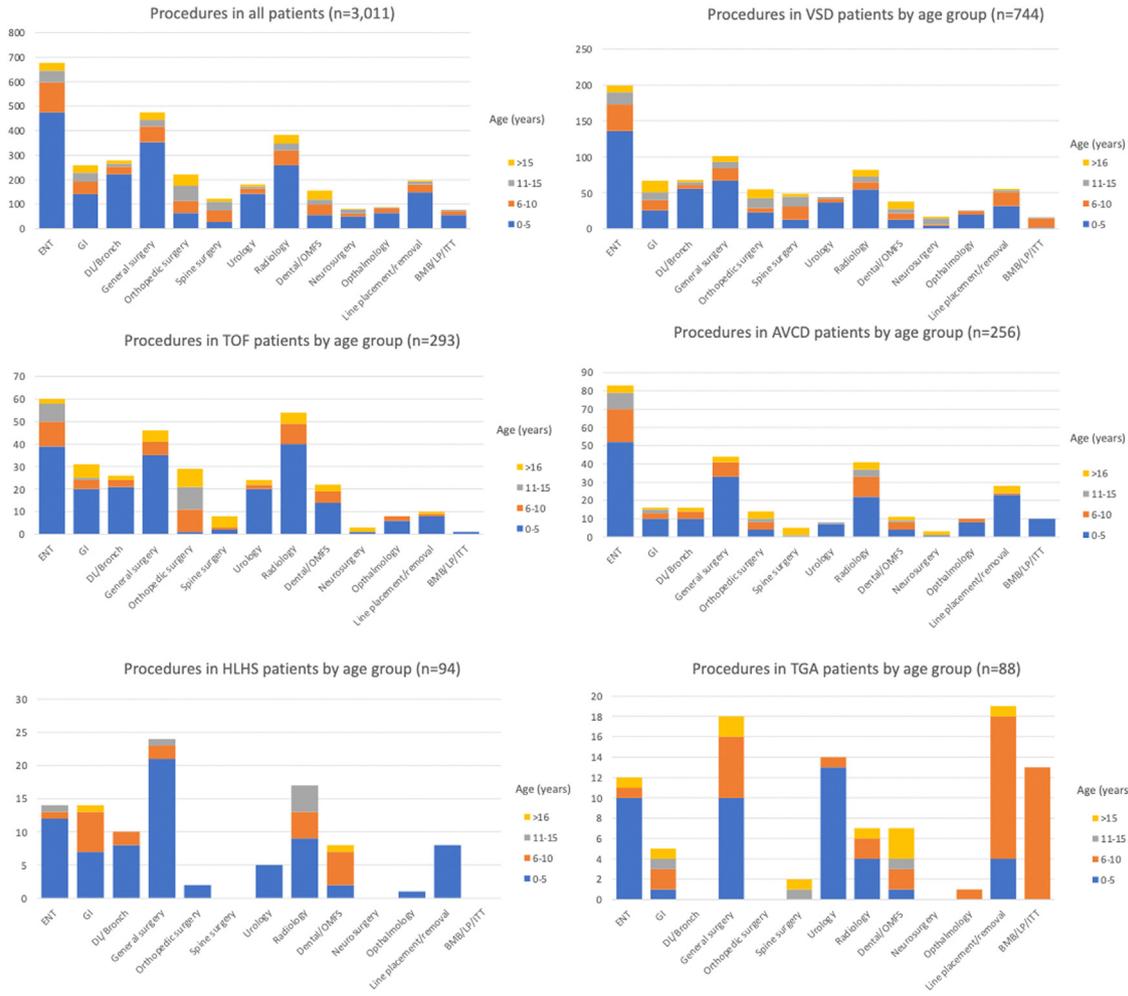


Fig 2. Age distribution of surgical procedures in different congenital heart disease cohorts. AVCD, atrioventricular canal defects; ENT, ear, nose, and throat; GI, gastrointestinal; HLHS, hypoplastic left heart syndrome; VSD, ventricle septal defects; TOD, TGA, transposition of the great arteries.

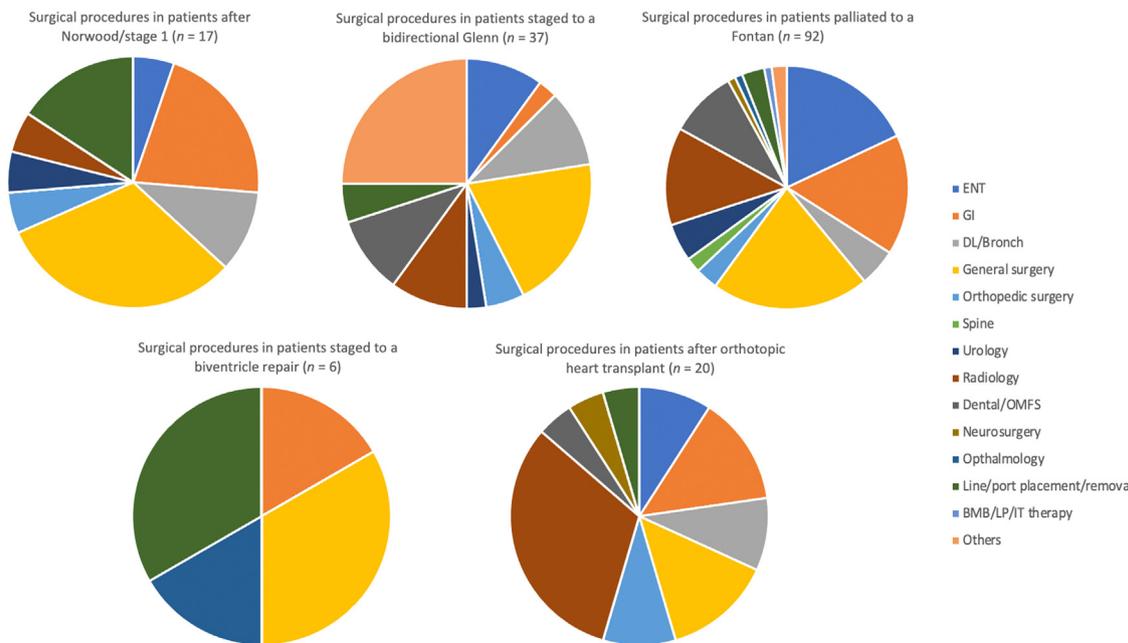


Fig 3. Types of noncardiac surgical procedures in patients with single-ventricle physiology following surgical palliation. BMB, bone marrow biopsies; DLB, direct laryngoscopy/bronchoscopy; ENT, ear, nose, and throat; GI, gastrointestinal; IT, LP, lumbar punctures; OMFS, oral and maxillofacial surgery;

Patients with CHD presenting to the authors' institution for noncardiac surgery typically are managed in the main operating room by the general pediatric anesthesia team. Pediatric cardiac anesthesia is available as a consulting service, and, in the authors' cohort, 17.8% (536/3010) of patients had been reviewed by a pediatric cardiac anesthesiologist prior to their noncardiac procedure in an effort to optimize preoperative planning and reduce the risk of intraoperative events. A multidisciplinary preoperative evaluation of cardiac function may further assist in identifying patients at increased risk for intraoperative events and stratify patients with abnormal ventricular function to nonambulatory settings with adequate perioperative support structures to manage unanticipated complications.^{13,24} As the age expectancy for patients living with CHD continues to increase, these considerations will become equally relevant for the adult population undergoing noncardiac surgery.²⁵

Although this study was limited to observational data from a single institution requiring careful interpretation without extrapolation to a larger population prior to further analysis, the authors believe that the findings presented here may be of value to medical providers caring for patients with CHD in similar perioperative settings. As careful surgical planning and multidisciplinary coordination are imperative when caring for patients with CHD, a better understanding of the surgical needs based on age groups and diagnosis may help inform some of the management choices at hand. Furthermore, the authors hope that these results may provide some additional knowledge on the different surgical needs along the palliative pathway for patients with single-ventricle physiology and a greater understanding of the specific risks associated with these patients as they present for noncardiac surgery in both the pediatric and adult setting. As the demand for noncardiac surgery in patients with CHD continues to increase, anticipating the specific needs related to different types of common CHD lesions may facilitate surgical planning, improve the quality and access to care, and reduce healthcare costs for this vulnerable and complex patient population.

Conflict of Interest

None.

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