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AHA SCIENTIFIC STATEMENT

Perioperative Considerations for Pediatric Patients With Congenital Heart Disease Presenting for Noncardiac Procedures: A Scientific Statement From the American Heart Association

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ABSTRACT: Continuous advances in pediatric cardiology, surgery, and critical care have significantly improved survival rates for children and adults with congenital heart disease. Paradoxically, the resulting increase in longevity has expanded the prevalence of both repaired and unrepaired congenital heart disease and has escalated the need for diagnostic and interventional procedures. Because of this expansion in prevalence, anesthesiologists, pediatricians, and other health care professionals increasingly encounter patients with congenital heart disease or other pediatric cardiac diseases who are presenting for surgical treatment of unrelated, noncardiac disease. Patients with congenital heart disease are at high risk for mortality, complications, and reoperation after noncardiac procedures. Rigorous study of risk factors and outcomes has identified subsets of patients with minor, major, and severe congenital heart disease who may have higher-than-baseline risk when undergoing noncardiac procedures, and this has led to the development of risk prediction scores specific to this population. This scientific statement reviews contemporary data on risk from noncardiac procedures, focusing on pediatric patients with congenital heart disease and describing current knowledge on the subject. This scientific statement also addresses preoperative evaluation and testing, perioperative considerations, and postoperative care in this unique patient population and highlights relevant aspects of the pathophysiology of selected conditions that can influence perioperative care and patient management.

Key Words: AHA Scientific Statements ■ anesthesiologists ■ critical care ■ heart defects, congenital ■ perioperative care

Innovations and technical advancements in the past decades have led to multidisciplinary improvements in the treatment of congenital heart disease (HD). As a result, the natural history and survival rates of patients with congenital HD have evolved such that >90% of children born with congenital HD now survive into adulthood.[1] Improved longevity in these individuals has increased the need for diagnostic and interventional procedures, as well as for surgical treatments for unrelated, noncardiac disease processes.

Approximately 30% of children with congenital HD exhibit associated extracardiac congenital anomalies.[2] Data from 2004 to 2012 highlight that almost half of patients (41%) who underwent cardiac surgical procedures in the first year of life also underwent at least 1 noncardiac operative intervention by 5 years of age.[3–5] A query of the Pediatric Health Information System database confirmed a significant increase in the number of encounters for noncardiac procedures in patients with congenital HD (from 38212 in 2015 to 45993 in 2019) and significant increases in the numbers of both inpatients and outpatients.[6] Gastrointestinal and otolaryngological procedures are the most common noncardiac procedures. In addition, children with congenital HD are

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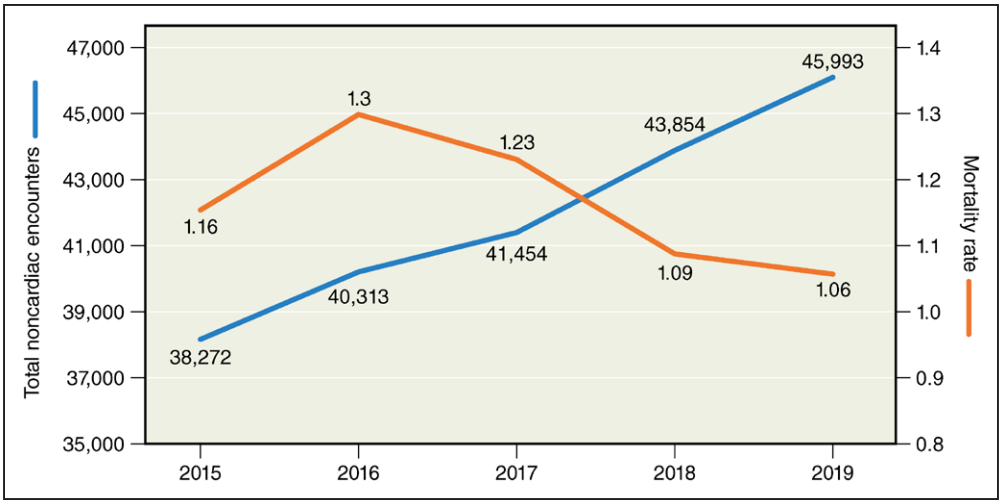

Figure 1. Total number of noncardiac encounters for patients with congenital heart disease and rate of mortality at freestanding children's hospitals from 2015 to 2019.[6]

at higher risk for mortality, complications, and reoperation after noncardiac procedures.[7] In 2019, the mortality rate in patients with congenital HD in this cohort was 1.06% compared with 0.12% in patients without congenital HD undergoing the same noncardiac procedures (Figure 1). Rigorous study of patient risk factors and outcomes has identified subsets of patients with congenital HD who have higher-than-baseline risk when undergoing noncardiac procedures, and this has led to the development of risk prediction scores specific to this population.[8–10]

This scientific statement reviews contemporary data on the risks associated with noncardiac procedures in pediatric patients with congenital HD and describes our current knowledge on the subject. Preoperative evaluation and testing, perioperative considerations, and postoperative care in this unique patient population are also addressed, and relevant aspects of the pathophysiology of selected conditions that can influence perioperative care and patient management are highlighted.

NONCARDIAC SURGERY RISKS IN PATIENTS WITH CONGENITAL HD

Perioperative Complications, Cardiac Arrest, and Mortality

It is generally accepted that patients with congenital HD undergoing noncardiac procedures have a higher risk for perioperative cardiac arrest, major complications, and mortality than patients without congenital HD.[3–7,11] A comprehensive analysis of data from the University Hospital Consortium identified 191261 pediatric noncardiac inpatient procedures between 1993 and 1996 and found that congenital HD was present in 5967 patients (3.1%).[3] Among these, the 30-day mortality rate was 6.0% compared with 3.8% in patients without congenital HD and 4.0% for the entire cohort (odds ratio [OR], 3.53 [95% CI, 3.15–3.95]).

The risk for complications from noncardiac surgery is high in younger individuals, particularly those <1 year of age; it plateaus thereafter and then increases again in late adolescence or early adulthood.[12,13] Miller and colleagues[12] reported a complication rate that was 9% overall but highest in infants, proportional to congenital HD severity. Complications included cardiac arrest, reintubation, infection, renal failure, neurological complication, thromboembolic complications, reoperation, 30-day unplanned revisit, 30-day prolonged hospital stay, and mortality. In older age groups, the presence of congenital HD was not independently associated with postoperative complications.[12]

Mortality is strongly influenced by age.[3] As with complications generally, the effect of age on operative mortality in children with and without congenital HD dwindles over time and is no longer apparent after 1 year of age. In infants <31 days of age undergoing noncardiac procedures, 30-day mortality rate was 13.3% for those with congenital HD versus 6.6% for those without congenital HD (OR, 2.18 [95% CI, 1.89–2.50]).[3] In children 31 days to 1 year of age, 30-day mortality rate was 3.1% for those with congenital HD versus 1.4% for those without congenital HD (OR, 2.23 [95% CI, 1.6–3.12]), and for children >1 year of age, the rate was similar at 1.5% versus 1.2% (OR, 1.19 [95% CI, 0.86–1.64]).

From 1994 to 2005, the Pediatric Perioperative Cardiac Arrest Registry collected data on 373 anesthesia-related cardiac arrests in children.[4] Among these, 127 patients (34%) had congenital or acquired HD. Mortality rate was 33% in patients with HD versus 23% in those without congenital or acquired HD. More than half (54%) of the cardiac arrests in patients with HD were reported from the general operating room compared with 26% from the cardiac operating room and 17%

Table 1. ACS-NSQIP Classification of Congenital HD Based on Residual Lesion Burden and Functional Status

Congenital HD classification	Definition and criteria
Minor	Cardiac condition with or without medication and maintenance (eg, atrial septal defect, small to moderate ventricular septal defect without symptoms) Repair of congenital HD with normal cardiovascular function and no medication
Major	Repair of congenital HD with residual hemodynamic abnormality with or without medications (eg, tetralogy of Fallot with free pulmonary regurgitation, hypoplastic left heart syndrome including stage 1 palliation)
Severe	Uncorrected cyanotic congenital HD Patients with documented pulmonary hypertension Patients with ventricular dysfunction requiring medication Listed for heart transplantation

ACS-NSQIP indicates the American College of Surgeons National Surgical Quality Improvement Program; and HD, heart disease.

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from the cardiac catheterization laboratory.[4] Another study reviewed all perioperative cardiac arrests in children undergoing cardiac surgery, noncardiac surgery, or cardiac catheterization at the Mayo Clinic between 1988 and 2005.[5] Overall, 92881 anesthetics were administered; 26 patients undergoing noncardiac surgical procedures experienced cardiac arrest: 7 cardiac arrests (26.9%) occurred in children with congenital HD, and 6 (23.1%) were attributed to the anesthesia. The most common cause of perioperative cardiac arrest in noncardiac procedures was hypovolemia caused by hemorrhage or hyperkalemia resulting from massive blood transfusion, and the single most common cause of death in children undergoing noncardiac procedures was hemorrhage (6 of 14 deaths [42.9%]).[5]

In an analysis of 3010 patients with congenital HD presenting for noncardiac surgery at Boston Children's Hospital between 2008 and 2013, the incidence of cardiovascular events was 11.5% and the incidence of respiratory events was 4.7%.[14] Perioperative cardiovascular events were associated with various factors, including American Society of Anesthesiologists Physical Status (ASA PS) ≥3, emergency cases, major and severe congenital HD as defined by American College of Surgeons National Surgical Quality Improvement Program (ACS-NSQIP) criteria based on residual lesion burden and functional status across anatomic subtypes (Table 1), single-ventricle physiology (SVP), ventricular dysfunction as defined by preoperative echocardiography, and surgical type broadly defined by specialty (orthopedic surgery, general surgery, neurosurgery, and pulmonary procedures).[10,14] Continuous inotrope administration at the discretion of the staff anesthesiologist (either proactively or in response to hemodynamic compromise) was used to indicate a cardiovascular event; by this measure, 99% of cardiovascular events were associated with inotrope use. Intraoperative cardiac arrest occurred in 5 of the cases (0.16%). Respiratory events were associated with ASA PS ≥4 and otolaryngology, gastrointestinal, general surgery, and maxillofacial procedures. Of note, whereas cardiovascular events were independently associated with underlying cardiac status, respiratory events were not.

In a subsequent analysis of this same cohort, of the 1028 patients with congenital HD who underwent ambulatory surgery, 165 had major congenital HD and 25 had severe congenital HD.[15] Patients with major congenital HD were more likely to require hospital admission than patients with minor congenital HD (OR, 2.43 [95% CI, 1.08–5.48]). Patients who had an echocardiogram within the previous 6 months were more likely to require hospital admission than those who had not (OR, 4.47 [95% CI, 1.34–14.9]). The authors suggested that a patient with a low residual lesion burden and stable, preserved ventricular function would be less likely to have had an echocardiogram performed in the previous 6 months in their institution. Severe congenital HD was not identified as a risk factor for hospital admission, but this was likely related to the small number of patients in this category who were scheduled for outpatient procedures.

In a study using the ACS-NSQIP pediatric database, 4520 children with congenital HD (2805 minor, 1272 major, 417 severe) were propensity matched with control subjects for sex, age group, ASA PS classification, elective versus emergency surgery, and surgical complexity. Reintubation, 30-day mortality rate, and overall mortality rate were more likely in patients with severe congenital HD than in patients with minor congenital HD.[10] The likelihood of these adverse outcomes in patients with minor congenital HD was similar to that in individuals from the general population.

In a study using a similar propensity-matching strategy, children with major or severe congenital HD undergoing laparoscopic surgery had higher overall mortality rate than children with no or mild congenital HD (OR, 3.46 [95% CI, 1.49–8.06] and 12.31 [95% CI, 1.59–95.1], respectively).[16] Likewise, composite morbidity (defined as surgical site infection, pneumonia, urinary tract infection, central line–associated bloodstream infection, reintubation, readmission, renal insufficiency, venous thrombotic events, neurological events, graft failure, cardiac arrest, sepsis, transfusion, unplanned readmission, or unplanned reoperation) was higher in children with major (OR, 2.07 [95% CI, 1.65–2.61]) or severe (OR, 2.51 [95% CI, 1.57–4.01]) congenital HD.

Risk Assessment Tools

Several attempts to construct risk assessment tools for this patient population have been published.[8,10,17–23] In 2016, a risk classification system was developed on the basis of literature review and expert opinion gathered from pediatric cardiac anesthesiologists practicing in a single center.[22] Cardiac lesions were classified into

Table 2. Multivariate Risk Stratification to Predict Postoperative Mortality

Variable	B (SE)	OR	95% CI	*P* value	Risk score
Emergency procedure	0.50 (0.17)	1.66	1.19–2.31	0.003	1
Severe congenital HD	0.50 (0.19)	1.65	1.15–2.39	0.007	1
SVP	0.61 (0.26)	1.83	1.10–3.06	0.020	1
Surgery within 30 d	0.70 (0.18)	2.01	1.40–2.89	<0.001	1
Inotropic support	0.72 (0.19)	2.05	1.40–3.01	<0.001	1
Preoperative CPR	0.90 (0.32)	2.46	1.32–4.57	0.004	2
Acute or chronic kidney injury	1.48 (0.40)	4.42	2.00–9.75	<0.001	3
Mechanical ventilation	2.05 (0.18)	7.80	5.42–11.21	<0.001	4

CPR indicates cardiopulmonary resuscitation; HD, heart disease; OR, odds ratio; and SVP, single-ventricle physiology.

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low (eg, repaired atrial or ventricular septal defects and mild regurgitation or stenosis of a single valve), moderate (eg, Wolff-Parkinson-White syndrome, long QT syndrome, pacemaker dependency, unrepaired simple lesions, repaired complex lesions, previous single ventricle, or transplantation), and high (eg, unrepaired complex cardiac lesions, systemic-to-pulmonary arterial shunts, severe valvular disease, pulmonary hypertension with New York Heart Association class III or IV, ventricular assist device, Williams-Beuren syndrome, and hypertrophic obstructive cardiomyopathy) risk categories. The risk category for a given patient based on the cardiac lesion was upgraded to the next level if the patient was <1 year of age, had comorbidities significant enough to be classified as ASA PS ≥3, or was undergoing a high-risk surgical procedure (ie, one that could cause hemodynamic instability, large fluid shifts, or high blood loss).

This risk categorization system was not validated and was used primarily to allocate cases to 1 of 26 anesthesiologists, 9 of whom were considered cardiac anesthesiologists. All cases classified as high risk were allocated to 1 of these 9 individuals. Using this system in 100 consecutive patient encounters resulted in complication rates of 8% in the low- and moderate-risk group and 9% in the high-risk group. At best, it can be concluded that the use of this system might mitigate risk in the high-risk group but not in the low- and moderate-risk group.

A similar analysis of 117 cases in a single institution used another empirically derived, nonvalidated risk stratification scheme.[17] Patients were classified into the various risk categories according to their physiology (poorly or well compensated), cardiac lesion (simple versus complex), type of surgery (major versus minor), and other factors such as age, emergency nature of the intervention, length of preoperative hospital stay, and ASA PS. The study found that pediatric anesthesiologists can safely provide care to children with congenital HD in low-, intermediate-, and high-risk groups when communication between team members is clear and the requisite expertise is available.[17]

Using the ACS-NSQIP pediatric database and the previously described ACS-NSQIP definitions of cardiac disease, Faraoni and colleagues[8] developed a rigorously derived risk assessment model for in-hospital mortality rate after noncardiac surgical procedures in children with major or severe congenital HD. A derivation cohort of 4375 patients (ACS-NSQIP years 2012–2013) and a validation cohort of 2869 patients (ACS-NSQIP year 2014) were used. Multivariable analysis identified 8 preoperative predictors (Table 2) that were used to create a risk stratification score of 0 to 10, which showed good calibration and discrimination in the validation cohort (area under the curve, 0.831 [95% CI, 0.787–0.875]). Scores ≤3 were associated with low risk for mortality (OR, 1.54 [95% CI, 0.78–3.04]); scores of 4 to 6, with medium risk (OR, 4.19 [95% CI, 2.56–6.87]); and scores ≥7, with high risk (OR, 22.15 [95% CI, 15.06–32.59]).[8]

In another study, a cohort of 37568 children with congenital HD undergoing noncardiac procedures was analyzed.[18] The results suggested that in children with congenital HD, the intrinsic surgical risk is not a predictor of 30-day rate, as opposed to results from a broader pediatric population.[23] In fact, patient comorbidities and the severity of the cardiac lesion at the time of the noncardiac surgical procedure appear to be the overwhelming, predominant determinants of 30-day mortality rate (Figure 2).[18]

Race and socioeconomic factors also have been found to affect outcomes in patients with congenital HD after noncardiac surgery. A 2021 propensity-matched analysis revealed that Black children with minor and major congenital HD experienced higher complication and postoperative mortality rates after noncardiac procedures than White children.[24] In fact, in children with congenital HD, poverty and low socioeconomic status have a disproportionately negative impact in the form of adverse postoperative outcomes. These include worse survival in patients with hypoplastic left heart syndrome; higher interstage and in-hospital mortality and greater resource use after orthotopic heart transplantation for single ventricle versus cardiomyopathy; higher mortality rate after congenital heart surgery; worse 1-year transplantation-free survival after stage 1 palliation in patients with single-ventricle congenital HD; unplanned readmission in the first 90 days after congenital heart surgery; and longer postoperative length of stay and higher resource use.[1]

Children and adults with congenital HD, especially those with unrepaired lesions or with a residual lesion burden and compromised cardiovascular status, require surgical care delivered by trained multidisciplinary teams and an individualized approach to anesthesia. Because congenital HD comprises many structural malformations,

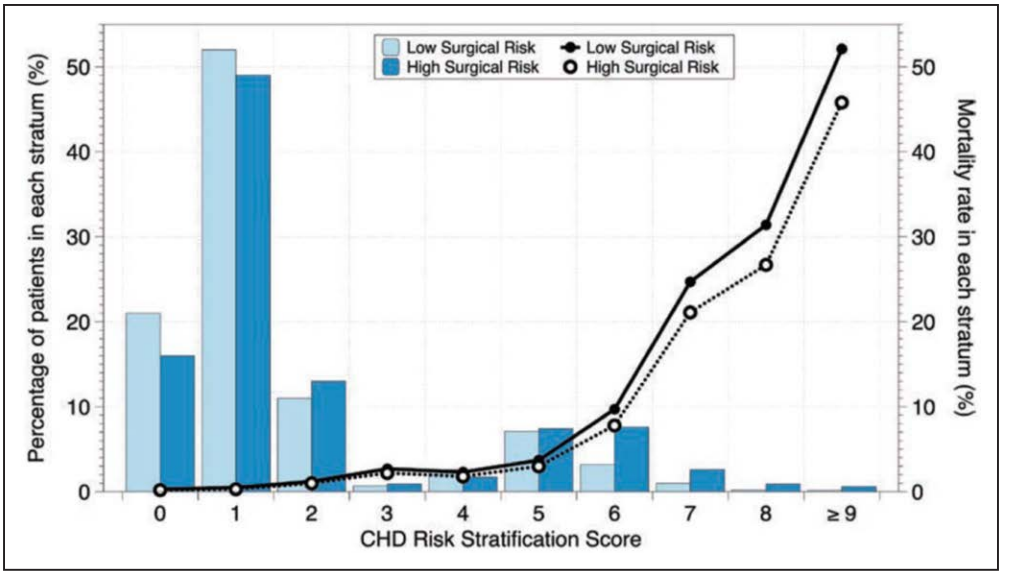

Figure 2. Observed 30-day mortality rate in patients with congenital heart disease undergoing low-risk vs high-risk surgical procedures.

This figure represents the distribution of the multivariable risk stratification score values in the derivation cohort in relation to the observed 30-day mortality rate for each score in patients with congenital heart disease (HD) undergoing low-surgical-risk or high-surgical-risk procedures. Reprinted from Faraoni et al[18] with permission. Copyright © 2020 Wolters Kluwer Health Inc.

each with specific physiological perturbations, hemodynamic consequences, and severity, perioperative management is challenging.[25] Practitioners, including cardiologists, intensivists, and primary care physicians who care for patients with congenital HD, must be able to identify the best location for procedures, specify a qualified team (eg, cardiac anesthesiologists, surgeons), and procure experts in noncardiac subspecialties (eg, nephrology, hematology, pediatricians). The American Heart Association guidelines for managing adults with congenital HD recommend a stepwise approach to preoperative cardiac assessment in this patient group.[26] Patient volume and the availability of specialized expertise have been found to influence overall clinical outcomes; consequently, it is recommended that these patients be treated in specialized congenital cardiac care centers to reduce risk.[26]

To date, no guidelines for comprehensive perioperative care of children with congenital HD undergoing noncardiac surgery have been established. Better delineation of the interplay among pathophysiology, preoperative evaluation, and perioperative and postoperative management in determining the unique risk profiles of patients in this population is definitely needed. Although there is no randomized controlled evidence that justifies the care of these children exclusively by a pediatric cardiac anesthesiologist for every anesthetic procedure, the anesthesiologist must have appropriate understanding of the anatomy and physiology coupled with experience to provide safe care. A suggested algorithm for risk stratification is presented in Figure 3.[6,8] It should be noted that the algorithm is based on the ACS-NSQIP classification and the congenital HD scoring published by Faraoni and colleagues,[8] neither of which was designed to assess the impact of the anesthesiologist on outcomes.

PREOPERATIVE EVALUATION BEFORE NONCARDIAC SURGERY

As previously discussed, children and adults with complex congenital HD who undergo noncardiac surgery have an increased risk for morbidity and mortality in the perioperative period.[6,10,20,27,28] Important determinants of this risk include the type of noncardiac procedure, the urgency of the procedure, and other underlying comorbidities.[14,16] Furthermore, although most heart defects occur in isolation, genetic and other extracardiac anomalies (eg, tracheoesophageal fistulas, anorectal anomalies, genitourinary and musculoskeletal pathologies) are present in about one-third of cases, and additional diagnostic and therapeutic procedures may be required to address these noncardiac ailments.[28–30] Thus, preoperative evaluation is critical for assessing potential risks from anesthesia related to the patient's clinical symptoms and current health status, the cardiac diagnosis, any altered physiology associated with noncardiac anomalies or illnesses, and the possible implications of the noncardiac procedure.

A comprehensive perioperative evaluation includes a review of presenting symptoms, medical and surgical history, and an assessment of cardiac status (eg, anatomy, pathophysiology, function, catheter-based or surgical interventions, present management). Clinical symptoms must be carefully evaluated, given that conditions such as intercurrent respiratory illnesses on pulmonary vascular resistance (PVR) can trigger adverse effects. Determining

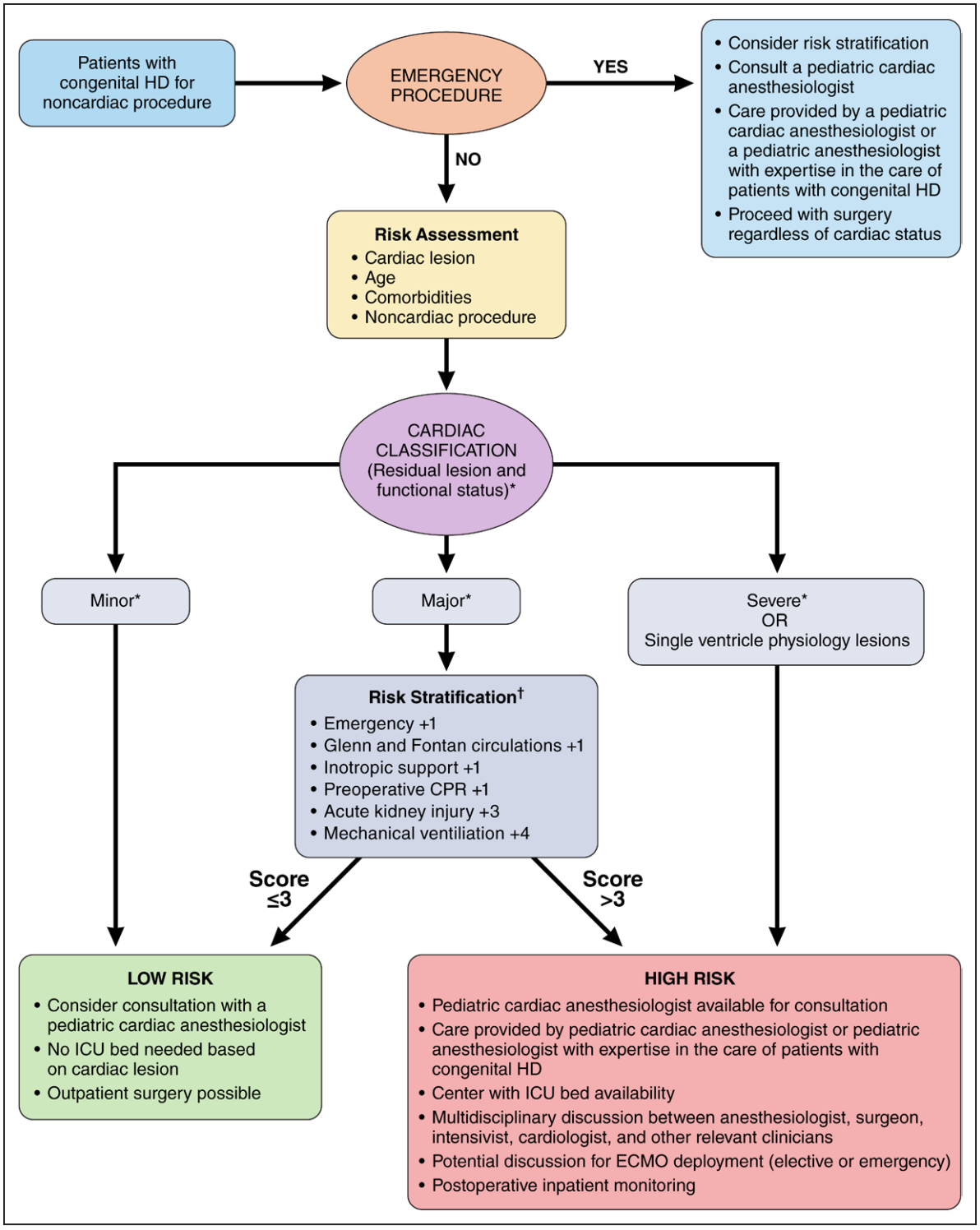

Figure 3. Systematic approach to patients with congenital heart disease presenting for noncardiac procedures.
*Minor, major, and severe disease is based on the American College of Surgeons National Surgical Quality Improvement Program definitions presented in Table 1.[6] †Please refer to Table 2 to calculate the score. Scores ≤3 were associated with low risk of mortality (odds ratio [OR], 1.54 [95% CI, 0.78–3.04]); scores of 4 to 6, with medium risk (OR, 4.19 [95% CI, 2.56–6.87]), and scores ≥7, with high risk (OR, 22.15 [95% CI, 15.06–32.59]).[8] CPR indicates cardiopulmonary resuscitation; ECMO, extracorporeal membrane oxygenation; HD, heart disease; and ICU, intensive care unit.

hydration status is particularly relevant in patients with cyanotic lesions and other defects for whom ventricular preload is important for maintenance of cardiac output. Fasting times should be carefully planned; in some cases, preoperative intravenous fluids may need to be initiated to prevent depletion of intravascular volume.

Pertinent surgical history and anesthesia records related to previous cardiac and noncardiac procedures

must be evaluated. Specifically, the type of anesthesia used, history of difficult or prolonged intubation, any postprocedural complications requiring intensive care monitoring, and intravenous access or monitoring difficulties must be assessed to anticipate problems that could require adjustment of the anesthesia plan. The expected impact of the noncardiac procedure on the underlying hemodynamics or pathophysiology also should be carefully assessed. The need for endocarditis prophylaxis should be considered.[31]

Functional status is determined primarily on the basis of clinical data. The physical examination includes assessment of vital signs, weight, and height. Blood pressure recordings in >1 extremity and quality of the arterial pulses can be useful in patients with underlying or treated aortic arch obstruction or systemic-to-pulmonary artery shunts. Baseline arterial saturation should be recorded by pulse oximetry. The examination should explore suitable sites for vascular access (venous and arterial) and identify potential difficulties. Airway abnormalities should be considered, given their likelihood in premature infants who have congenital HD, in patients <10 kg in weight, and in those who have cardiac lesions with an associated genetic syndrome (eg, Down, 22q11 deletion, or CHARGE [coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities] syndrome).[32] Identifying patients at high risk for airway abnormalities facilitates preparation and planning for perioperative airway management. Ultimately, emphasis should be given to the respiratory and cardiovascular systems during the examination.[32,33] Noncardiac conditions or pathologies that may affect anesthesia care also should be noted.

All recent preoperative cardiac imaging studies—ECGs, echocardiograms, any diagnostic or interventional catheterization data, cardiac magnetic resonance images, and chest computed tomography images—should be reviewed. Particular attention should be paid to the presence of ventricular systolic or diastolic dysfunction, lesions such as moderate or severe atrioventricular and semilunar valve dysfunction or aortopulmonary shunt compromise, severe systemic outflow tract obstruction, and associated end organ–related comorbidities such as liver, renal, or pulmonary disease. When available, reports tracking potential changes in those parameters over time are important to understand the course of the status and hemodynamic conditions of the heart. If a patient has developed new symptoms since the last imaging study, then it may be advisable to request a new echocardiographic examination. When cyanosis is present, low cardiac output syndrome must be distinguished from intrapulmonary gas exchange abnormalities (ie, ventilation-perfusion mismatch, intrapulmonary shunt) and insufficient pulmonary blood flow attributable to shunt stenosis or dysfunction, pulmonary artery stenosis, or collateral decompression of cavopulmonary connections.

Depending on the type of noncardiac surgical procedure that is planned, a chest radiograph may be helpful for assessing heart position and size, vascular markings, lung parenchyma, vascular access placement, and hemidiaphragm position. A chest radiograph can also be helpful in patients with implanted rhythm devices to determine type if not known, lead patency, and position. For the patient with an implanted pacemaker or automated internal cardiac defibrillator, the device manufacturer, model, modes, and settings; underlying reason for implantation (eg, congenital, acquired, or iatrogenic lesions); baseline cardiac rhythm; ventricular function; symptomatology; and extent of pacemaker dependency should be determined. These devices should be interrogated before the procedure and reprogrammed if necessary. After the procedure, they should again be interrogated and reprogrammed as indicated in consultation with a cardiologist.

In addition, conduction disturbances and arrhythmias can occur intraoperatively and postoperatively. Prevention strategies include avoidance of atrial and ventricular overdistention, acute hypoxemia, electrolyte abnormalities, and hyperthermia. Post-Fontan palliation and certain heart defects such as congenitally corrected transposition of the great arteries and Ebstein anomaly of the tricuspid valve have an enhanced substrate for conduction abnormalities and rhythm disturbances. In patients with SVP or ventricular impairment, sinus node dysfunction or the loss of sinus rhythm alone may be sufficient to drastically affect cardiac output. Hence, preoperative preparation with access to medications and devices to perform pacing, cardioversion, and defibrillation should be available in case a hemodynamically significant arrhythmia occurs.

Preoperative laboratory analyses should be obtained on the basis of clinical symptoms, medication use, and the noncardiac procedure to be performed. A chemistry panel that includes electrolytes may be indicated in patients taking diuretics or antihypertensive agents or patients with known arrhythmias triggered by electrolyte disturbances. A comprehensive metabolic profile that assesses acid-base balance, renal and hepatic function, and blood glucose and blood protein concentrations may be more appropriate in some cases. A complete blood panel with differential can provide markers for potential infection, anemia, or erythrocytosis; preoperative anemia has been identified as a risk factor for in-hospital mortality during noncardiac surgery in children with congenital HD.[34] In some cases, thyroid function testing, brain natriuretic peptide levels, or pregnancy testing may be needed.

Certain patient groups at risk for bleeding such as those on anticoagulant therapy and those with cyanotic congenital HD may benefit from coagulation testing. It is important to note that even relatively simple, elective procedures such as circumcision have been associated with a significant risk for bleeding.[35] Patients with Fontan

physiology are known to be at risk for bleeding because of their chronically elevated central venous pressures, associated liver disease, and known coagulation abnormalities.[36] At the same time, children with cyanotic HD, single-ventricle palliation, or implanted ventricular assist devices are known to be at risk for thrombotic complications.[37] Patients with GORE-TEX shunts, especially modified Blalock-Thomas-Taussig shunts, are at risk for shunt thrombosis when anticoagulation is stopped and the inflammatory response, hemodynamic changes, and fluid shifts related to surgery are encountered.[37a] For patients on anticoagulation therapy, prothrombin time, international normalization ratio, or anti–factor Xa levels may be obtained. Discontinuation of anticoagulation and the timing thereof, which depends on the cardiac lesion and expected bleeding from the surgical procedures, should be discussed with the cardiologist and the surgeon. Blood typing and cross-matching should be performed if the need for blood administration is anticipated.

Review of current medications and medication history is an important aspect of the preoperative evaluation, given that patients taking medications such as β-blockers, diuretics, or angiotensin-converting enzyme inhibitors may be at risk during anesthesia for hemodynamic changes potentiated by hypovolemic states such as prolonged fasting. The anesthesiologist must be mindful of associated side effects of or drug interactions among certain cardiac and noncardiac medications in the patient with congenital HD. Typically, most cardiac medications are well tolerated, although some practitioners prefer to withhold diuretics if prolonged fasting is anticipated. Opinions differ about the administration of angiotensin-converting enzyme inhibitors or angiotensin receptor blockers before anesthesia because of reports of intractable vasodilation and hypotension during general anesthesia, particularly in the adult population.[38] The presurgical management of these drugs in children is not well characterized, with most centers advising discontinuation the day before surgery if possible.[39] Low-dose aspirin is often well tolerated during superficial surgical procedures but is held ≥5 days before major surgical procedures because of the potential for bleeding.[40] In contrast, patients on warfarin may need to be either transitioned to subcutaneous therapy in the outpatient setting or admitted for conversion to intravenous heparin.

Institutional practices related to asymptomatic nasopharyngeal viral panel screening before cardiac or noncardiac surgery may vary. This screening is often based on diagnosis, risk factors, and history of illness or sick contacts. However, limited evidence of improved outcomes supports this practice in the asymptomatic child.[41] Other surveillance includes methicillin-resistant *Staphylococcus aureus* colonization and coronavirus disease 2019 (COVID-19) symptom monitoring and testing.[42]

After the patient, the underlying cardiac and other coexisting conditions, and the noncardiac procedure to be performed are thoroughly assessed, the anesthesiologist must develop a plan, assess risk, and consider whether the institution is prepared to handle potential complications. If surgery is being performed in a nonspecialized center, preoperative consultation with local cardiac anesthesia teams may be warranted, particularly if a higher level of expertise becomes necessary after the procedure. Occasionally, the results of a preoperative evaluation dictate the need to delay or defer elective noncardiac surgery, to initiate other interventions, or to undertake additional diagnostic procedures.

PERIOPERATIVE CONSIDERATIONS FOR SPECIFIC HIGH-RISK CARDIAC LESIONS

An important component of preoperative risk stratification is functional status, as defined according to cardiac function, residual lesion burden, requirements for inotropic support, need for preoperative mechanical ventilation, and preoperative intensive care unit admission at the time of surgery. Beyond these factors, the type of cardiac lesion also has been shown to be a significant predictor of risk.[8,18] Residual lesions can be present throughout life in patients with certain genetic syndromes such as Williams-Beuren syndrome, in whom arteriopathy, especially coronary insufficiency, may not have been evaluated or fully addressed, and Noonan syndrome with incompletely treated outflow tract obstruction.[43,44] Furthermore, even in patients with a favorable intraoperative course, postprocedural observation is key to ensure early identification or prevention of acute decompensation in the postoperative period.

It is also important to consider the proposed surgical procedure in perioperative planning. During procedures associated with significant physiological responses prompted by direct surgical tissue injury, the mechanical deformation of organs, blood loss, core temperature variations, and fluid shifts, it is prudent to consider using more invasive monitoring such as intra-arterial and central venous catheters. During laparoscopic procedures, careful attention should be paid to the intra-abdominal insufflation pressure because excessive pressure can compromise cardiac output by impeding venous return and increasing mean airway pressure. Systemic carbon dioxide embolization is always a risk during these procedures but is of particular concern in patients with right-to-left anatomic shunts from intracardiac or collateral circulation sources.

Single Ventricle

Several series have shown that patients with a single ventricle are at increased risk for adverse events when undergoing noncardiac surgery.[4,20,21] In analyses of this type of risk, it is important to differentiate patients with SVP such as those pre– or post–stage 1 single-ventricle

palliation from patients at more advanced stages of single-ventricle palliation such as those with a superior cavopulmonary anastomosis (SCPA; also known as a bidirectional Glenn shunt) or total cavopulmonary anastomosis (Fontan palliation). SVP refers specifically to the physiology defined in the Single-Ventricle Physiology section. In a 2015 retrospective analysis by Brown and colleagues,[21] the incidence of intraoperative and early postoperative adverse events in children with SVP undergoing noncardiac surgery was as high as 11.8%. Of the 34% of children with congenital HD in the Pediatric Perioperative Cardiac Arrest Registry who had a cardiac arrest during noncardiac surgery, 13% were patients with a single ventricle.[4] An analysis of the Health Care Cost and Utilization Project's National Inpatient Sample found that the incidence of in-hospital mortality among children with a single ventricle undergoing noncardiac surgery varied between 17% and 22%.[19]

Although the risk associated with noncardiac surgery is known to be elevated in patients with single-ventricle palliation, the risks associated with each stage of palliation (SVP, SCPA, Fontan) are less well characterized. Physicians caring for patients with a single ventricle should understand the anatomy and specific physiological vulnerabilities associated with each stage of palliation.[45–47]

Single-Ventricle Physiology

In SVP, the complete mixing of pulmonary and systemic venous blood occurs at the atrial or ventricular level, and the ventricle(s) then will distribute output to both the systemic and pulmonary beds. As a result of this physiology, ventricular output is the sum of pulmonary blood flow and systemic blood flow; distribution of systemic and pulmonary blood flow depends on the relative resistances to flow (both intracardiac and extracardiac) into the 2 parallel circuits; and oxygen saturations are the same in the aorta and the pulmonary artery.

This physiology can exist in patients with 1 well-developed ventricle and 1 hypoplastic ventricle, as well as in patients with 2 well-formed ventricles when there is complete or near complete obstruction to outflow from 1 of the ventricles.[48] Supplemental Table 1 provides a list of the full spectrum of SVP lesions. Single-ventricle lesions commonly occur in conjunction with heterotaxy syndrome. Heterotaxy is characterized by failed embryonic development of normal left-to-right asymmetry. There is a high prevalence of airway ciliary dysfunction in patients with heterotaxy with congenital HD similar to that seen with primary ciliary dysfunction. Mucociliary clearance defects in the airway predispose these patients to high postsurgical morbidity and mortality associated with respiratory complications.

In the case of a single anatomic ventricle, the pathway to either the pulmonary artery or aorta may be obstructed as a result of variable degrees of obstruction to inflow, outflow, or both. In this circumstance, a downstream, unobstructed pathway to the great vessels is necessary to ensure postnatal survival. In some instances of a single anatomic ventricle, a direct connection between the aorta and the pulmonary artery through a patent ductus arteriosus is the sole source of systemic blood flow (eg, hypoplastic left heart syndrome) or of pulmonary blood flow (eg, pulmonary atresia with an intact ventricular septum). This is known as ductal-dependent circulation. In other instances of a single anatomic ventricle, intracardiac pathways provide both systemic and pulmonary blood flow without a patent ductus arteriosus. This is the case in tricuspid atresia with normally related great vessels, a nonrestrictive ventricular septal defect, and minimal or absent pulmonary stenosis.

Although patients with totally anomalous pulmonary venous return have complete mixing of pulmonary and systemic venous blood at the atrial level, they do not manifest the other features necessary to create SVP. This also holds true for lesions in which a common atrial or ventricular chamber exists attributable to bidirectional (both left-to-right and right-to-left) anatomic shunting across a large defect (atrial or ventricular septal communication) and where there is no obstruction to ventricular outflow.

Patients with SVP, most commonly those with stage 1 palliation or uncorrected hypoplastic left heart syndrome, often require noncardiac procedures within the first several months of life. The most common procedures include central line insertion, laparoscopy for gastric tube insertion, laparotomy for necrotizing enterocolitis or the Ladd procedure, and direct laryngoscopy and bronchoscopy for airway assessment. In addition to having vulnerabilities related to their underlying physiology, these neonates and young infants are at further increased risk for perioperative complications, even in the absence of congenital HD. Severe dysfunction of the single ventricle can occur during the first months of life, and these patients are often dependent on ventilatory support, inotropic support, or both at the time of their noncardiac surgery.

All patients with SVP who have severe hypoplasia of 1 ventricle will ultimately be staged down the single-ventricle pathway to the Fontan procedure. Patients with SVP and 2 well-formed ventricles may ultimately be able to undergo a 2-ventricle (biventricular) repair. With SVP, arterial oxygen saturation will be determined by the relative volumes and saturations of pulmonary and systemic venous blood flows that have mixed and reached the aorta.[49] Supplemental Figure 1A illustrates the anatomy, oxygen saturation, and blood flows in neonates with hypoplastic left heart syndrome palliated with Damus-Kaye-Stansel anastomosis, aortic arch reconstruction, coarctectomy, atrial septectomy, and an aortopulmonary shunt.[49]

It should be emphasized that patients with unrepaired SVP and palliated patients with SVP are at risk of

hemodynamic instability and cardiovascular collapse if a balanced pulmonary blood flow and systemic blood flow ratio is not maintained. Ventilatory interventions that can precipitously reduce PVR such as alveolar hypocarbia or hyperoxia should be avoided. Likewise, noxious stimuli or administration of medications that acutely increase systemic vascular resistance should be avoided. With this in mind, it is important for the perioperative team to understand the specifics of the palliation when caring for these patients. For example, there is an important difference between infants with a systemic-to-pulmonary shunt (modified Blalock-Thomas-Taussig) and those with a ventricle-to-pulmonary shunt. In the presence of a systemic-to-pulmonary shunt, aortic diastolic pressure is reduced because of delivery of blood into the low-resistance pulmonary circulation during both systole and diastole. Efforts should be made to maintain adequate diastolic pressure by manipulating PVR to minimize diastolic runoff into the pulmonary bed. In particular, the combination of tachycardia and diastolic hypotension places the volume-loaded single ventricle at risk for acute ischemic decompensation. In the presence of a ventricle-to-pulmonary shunt, the diastolic pressure is generally higher, but care should still be taken to balance pulmonary and systemic blood flow.

SCPA and Fontan Physiology

The SCPA procedure (also known as the bidirectional Glenn shunt) directs systemic venous blood from the superior vena cava directly to the pulmonary circulation while the inferior vena cava blood drains to a common atrial chamber.[49] This anatomy and physiology are summarized in detail in Supplemental Figure 1B. In contrast, the Fontan operation (or total cavopulmonary connection) directs systemic venous blood from both the superior and inferior vena cavae directly to the pulmonary circulation and places the systemic and pulmonary circulations in series.[49] This anatomy and physiology are summarized in detail in Supplemental Figure 2A.[49] In both circulations, pulmonary blood flow is gradient driven, with energy ultimately provided by a single or common ventricular chamber. Increased systemic venous pressure is a necessity in Fontan physiology because the interposition of the cavopulmonary connection and pulmonary vascular bed between the systemic venous system and the pulmonary venous atrium increases the resistance to venous return to the systemic ventricular atrium. This physiology in summarized in detail in Supplemental Figure 2B.[49] These procedures provide a physiology that is much less prone to hemodynamic instability and cardiovascular collapse than that present in SVP.

The goal of SCPA is to maintain pulmonary blood flow while simultaneously reducing the volume load on the systemic ventricle. This volume reduction promotes the remodeling necessary for a Fontan procedure to be performed. This reduction in ventricular volume alone is not sufficient to improve atrioventricular valve dysfunction. After SCPA, most patients have preserved ventricular function with a stable source of pulmonary blood flow. Regardless of the mode of ventilation and in patients with a natural airway, moderate hypercapnia (partial pressure of carbon dioxide [$Paco_2$] 45–55 mm Hg) with a mild respiratory acidosis (pH 7.30–7.35) improves arterial oxygen saturation and overall oxygen transport. On balance, the slight increase in PVR seen with this strategy is offset by increased cerebral blood flow and increased superior vena cava drainage, which augments pulmonary blood flow. These patients are preload dependent and sensitive to significant increases in PVR requiring normal or near-normal atrioventricular valves and ventricular function. They are more affected by rhythm disturbances than are patients with a normal heart.

Whereas any of these issues can decrease cardiac output and induce cyanosis, SCPA provides resilient circulation because cardiac output can be temporarily maintained with little or no pulmonary blood flow; blood from the inferior vena cava is naturally routed to the systemic ventricle without crossing the pulmonary bed. If positive pressure ventilation is initiated in a patient with SCPA physiology, it is important that tidal volume, peak end-expiratory pressure, inspiratory:expiratory ratio, respiratory rate, and gas flow pattern be manipulated to provide the lowest possible mean airway pressure compatible with the desired tidal volume, minute ventilation, and gas exchange parameters and with the prevention of atelectasis. Of note, patients with SCPA, particularly those with reduced pulmonary blood flow, will have large areas of zone 1 lung and physiological dead space. Consequently, a wide end-tidal carbon dioxide–to–$Paco_2$ gradient should be anticipated. Because patients with SCPA are less likely than patients who have had a stage 1 procedure to exhibit hemodynamic decompensation with acute changes in systemic and PVR, it may be prudent to delay elective noncardiac surgery until the SCPA is completed.

The Fontan circulation is characterized by a preload-limited, high-ventricular-afterload state. Stroke volume is dependent on delivery of blood across the pulmonary vascular bed driven by the pressure gradient between the system venous circulation and the common atrium. Cardiac output can be significantly compromised by even small increases in PVR, reductions in systemic venous pressure, or elevations in common atrial pressure. In addition, administration of medication with venodilating properties is problematic because these patients are normally venoconstricted to maintain a large stressed venous volume (volume of venous blood that generates pressure). Using venodilation to convert stressed volume to unstressed volume (volume of venous blood that does not generate pressure) reduces systemic venous pressure and the pressure gradient needed to maintain cardiac output. This phenomenon can generally be

counteracted by administration of a fluid bolus (10–20 mL/kg) or a venoconstricting agent, with the caveat that these drugs might simultaneously elevate afterload.

If positive pressure ventilation is initiated in a patient with Fontan physiology, it is important that tidal volume, peak end-expiratory pressure, inspiratory:expiratory ratio, respiratory rate, and gas flow pattern be manipulated to provide the lowest possible mean airway pressure compatible with the desired tidal volume, minute ventilation, and gas exchange parameters and with the prevention of atelectasis. The potential deleterious effects of positive pressure ventilation may be more than offset by the detrimental effects of hypercarbia, hypoxemia, and reduced lung volumes on PVR and pulmonary blood flow that accompany inadequate spontaneous ventilation.

The long-term consequences of chronically elevated systemic venous pressure such as Fontan-associated liver disease, protein-losing enteropathy, plastic bronchitis, and renal dysfunction in these patients may substantially alter their risk profile. In addition, heart failure, arrythmias, and thromboembolic events present management challenges.[47]

Pulmonary Hypertension

Pulmonary hypertension, defined as a mean pulmonary artery pressure ≥20 mm Hg at rest, can occur as a result of congenital heart lesions.[50] The stimulus for the development of pulmonary hypertension in congenital HD is exposure of the pulmonary vascular system (both in utero and after birth) to abnormal pressure and flow patterns. The condition can be considered to have a precapillary cause (pulmonary arterial hypertension), a postcapillary cause (pulmonary venous hypertension), or a combination of both.

Precapillary pulmonary hypertension is defined as mean pulmonary artery pressure ≥20 mm Hg at rest, pulmonary artery wedge pressure ≤15 mm Hg, and a PVR index >3 Wood units/m^2 for biventricular circulation. After SCPA or Fontan palliation, the definition changes to a PVR index >3 Wood units/m^2 or a transpulmonary gradient >6 mm Hg even if the PVR index is <3 Wood units/m^2. Postcapillary pulmonary hypertension is defined as mean pulmonary artery pressure ≥20 mm Hg at rest in conjunction with pulmonary artery wedge pressure ≥15 mm Hg. Postcapillary causes are most commonly related to left-sided HD and are characteristically associated with interstitial pulmonary edema. Children with long-standing postcapillary pulmonary hypertension develop a component of precapillary pulmonary hypertension attributable to reactive pulmonary artery vasoconstriction.

The morphological changes associated with pulmonary arterial hypertension in patients with congenital HD have 3 components: increased muscularity of small pulmonary arteries; small-artery intimal hyperplasia, scarring, and thrombosis; and reduced numbers of intra-acinar arteries. These changes produce advancing obstruction to pulmonary blood flow and result in progressive and irreversible elevations in PVR and pulmonary artery pressure. In addition, the increased muscularity of the small pulmonary arteries enhances the response of the pulmonary vasculature to pulmonary vasoconstrictors.

Children with pulmonary hypertension have significantly elevated risk for perioperative morbidity and mortality; the incidence of perioperative cardiac arrest ranges from 0% to 5%, and the incidence of perioperative death is as high as 1.5%.[51–56] For patients at risk, preventive therapy should include avoidance of hypothermia, stress, pain, acidosis, hypercarbia, and hypoxia.[52] A pulmonary hypertensive crisis is characterized by an acute elevation in mean arterial pulmonary pressure above the mean systolic arterial pressure meditated by abrupt elevations in PVR leading to acute right ventricular dysfunction, systemic hypotension, and inadequate cardiac output. It may also lead to systematic arterial desaturations in the presence of shunts. These crises are an important cause of morbidity and mortality in these patients. Treatment may require the use of additional sedation and paralytics, select pulmonary vasodilators (eg, inhaled nitric oxide, phosphodiesterase inhibitors), and inotropic therapy to support right ventricular function.[57]

Left Ventricular Outflow Tract Obstruction

Patients with left ventricular outflow tract obstruction from subvalvular, valvular, or supravalvular aortic stenosis can develop concentric left ventricular hypertrophy. These patients are at high risk for developing supply-demand subendocardial ischemia with hypotension, particularly in combination with tachycardia. Supravalvular aortic stenosis is seen in patients with elastin arteriopathy, the majority of whom have Williams-Beuren syndrome. All patients with left ventricular outflow tract obstruction, particularly those with elastin arteriopathy, are known to be at high risk for complications during and after general anesthesia and sedation.[58–61]

Patients with elastin arteriopathy also may have central and peripheral pulmonary artery stenosis leading to severe bilateral outflow tract obstruction with biventricular hypertrophy. Severe biventricular outflow tract obstruction is uniformly recognized as a risk factor for sudden cardiovascular collapse under anesthesia or sedation. There are believed to be multiple sources of potential coronary blood compromise in these patients, which alone or in combination can lead to myocardial ischemia and cardiovascular collapse. Both ostial and diffuse left and right coronary artery stenoses may be present; adhesion of the right or left aortic leaflet edge to the narrowed sinotubular junction can restrict coronary blood flow into the sinus of Valsalva; and loss of aortic distensibility reduces the diastolic component of phasic coronary blood flow.[62]

For patients with elastin arteriopathy, comprehensive risk stratification, preoperative intravenous hydration, intravenous induction, and early use of continuous vasoactive medications may improve hemodynamic stability during anesthesia induction and maintenance (2% incidence of adverse events).[63–66] Although the risks from general anesthesia in patients with Williams-Beuren syndrome are high, when patients are appropriately screened and anesthesiologists are well versed in hemodynamic goals, anesthesia for noncardiac procedures can proceed safely with minimal morbidity and mortality.[66]

In patients with left ventricular outflow obstruction who develop left ventricular hypertrophy, an elevated left atrial pressure is necessary to ensure an adequate left ventricular end-diastolic volume; prolonged periods of fasting should be avoided, or arrangements should be made to provide intravenous fluid supplementation before a procedure.

Cardiomyopathy

It is generally acknowledged that cardiomyopathies increase the risk for perioperative adverse events. However, information with which to define the risk is limited. Hypertrophic cardiomyopathy is characterized by left ventricular hypertrophy, diastolic dysfunction, reduced left ventricular compliance, left atrial enlargement, and increased left atrial pressure. Left ventricular contractility is generally preserved or hyperdynamic. Systolic anterior motion of the mitral valve causes dynamic left ventricular outflow tract obstruction and is associated with an inferolaterally directed mitral regurgitant jet. Left ventricular hypertrophy may lead to subendocardial ischemia and subsequent hemodynamic compromise from myocardial oxygen supply-demand imbalance. The combination of hypotension and tachycardia is particularly threatening to subendocardial perfusion.[67,68]

Dilated cardiomyopathy is often idiopathic but can be secondary to genetic disorders, infection, drugs, congenital HD, or other medical conditions. Dilated cardiomyopathy is characterized by ventricular dilation, impaired ventricular systolic function, loss of preload recruitable stroke work, and exaggerated depression of stroke volume in the setting of elevated afterload. Restrictive cardiomyopathy is a rare form (<5% of pediatric cardiomyopathies) characterized by biatrial enlargement and severely restrictive biventricular physiology but normal systolic function and size. Patients with restrictive cardiomyopathy are at risk of hemodynamic collapse with acute decreases in preload and systemic vascular resistance, given the inability to increase stroke volume. The need for higher atrial pressures to ensure adequate preload to the restrictive ventricles must be balanced against the consequences of pulmonary and systemic venous congestion that may result. Patients with restrictive cardiomyopathy are at risk of postcapillary pulmonary hypertension and development of secondary precapillary pulmonary hypertension, predisposing them to pulmonary hypertensive crises. In general, patients with cardiomyopathy benefit from invasive blood pressure and central venous pressure monitoring for dynamic assessment of the hemodynamic consequences of alterations in preload, afterload, and heart rate.

POSTOPERATIVE CARE

Postoperative care involves many of the same principles as intraoperative management. The level of postoperative recovery (eg, outpatient ambulatory versus inpatient) and need for invasive monitoring will depend on the type of noncardiac procedure, the severity of the congenital heart lesion with underlying hemodynamics, other coexisting medical conditions, and the clinical condition after surgery.

Observation and physical examination can provide a great deal of information on respiratory status, cardiac function, and systemic perfusion. Hemodynamic changes may occur when the patient is emerging from anesthesia. Airway protection is vital for maintaining adequate oxygenation and ventilation so as to avoid hypoventilation and the negative effects on pulmonary vascular tone. Knowledge of preexisting extrinsic compression or intrinsic airway abnormalities (eg, spinal deformities, bronchomalacia) is essential for postoperative airway management. The need for reintubation after a noncardiac procedure is a potential risk and has been reported to be higher in children with major or severe types of congenital HD than in children with other defects and control subjects.[10] Thus, monitoring for signs of respiratory distress or depression is imperative in case reintubation is needed. Effective pain management can help restore normal respiratory status in the initial postoperative period.

Hypotension can occur after surgery and is multifactorial. It can result from hypovolemia from prolonged fasting or procedure-related blood loss, arrhythmias, postoperative sedation and analgesia, or myocardial dysfunction. Management should focus on the causative factor. In the patient with hypovolemia, intravascular volume can be restored with a fluid challenge to re-establish adequate perfusion and blood pressure. In addition, a vasopressor may be needed in the acute setting until a more definitive therapy is instituted. Depending on the patient's cardiac defect, extent of the surgical procedure, blood loss, or insensible volume loss, monitoring hemoglobin or hematocrit values, electrolytes, glucose, and calcium may be necessary in the postoperative period. Replacement with dextrose-containing fluids may be required in neonates and small infants. In the patient with cyanosis, it is imperative that adequate postoperative hydration is maintained to avoid venous stasis and risk for thrombotic events. In the child with 22q11 deletion syndrome, ionized calcium

levels should be examined to monitor for hypocalcemia, which is commonly seen in children affected by this syndrome.

The timing for resumption of anticoagulation and antiplatelet therapies after noncardiac surgery is based on the relative risk for bleeding versus thrombosis and must be deemed safe by the surgical team. The risk for thrombosis is elevated in patients with cyanotic HD and highest in patients with a single ventricle.[37,47] Depending on the indication for anticoagulation (eg, mechanical mitral or aortic valve), intravenous heparin may be required until oral anticoagulation can be reinitiated.

In addition, conduction disturbances and arrhythmias can occur postoperatively as described in the Preoperative Evaluation Before Noncardiac Surgery section.

NONCARDIAC CONDITIONS

As mentioned, whereas congenital HD and other syndromic cardiovascular conditions are often considered in isolation, most patients with congenital HD have additional associated noncardiac comorbidities. However, many of these noncardiac conditions may be underappreciated or go unevaluated until symptoms that affect the patient's quality of life are reported.[68,69] Elective procedures allow time for additional evaluation of noncardiac systems to better define their severity and impact on the noncardiac surgery risk, whereas an emergency procedure may not allow sufficient time to fully investigate this multisystem involvement. Consequently, risk assessment based on available information may substantially underestimate the actual risk. Figure 4 details the most common extracardiac conditions encountered in patients with congenital HD.

An analysis of adolescents with congenital HD found significant noncardiac comorbidities: pulmonary (16%–34%), infectious disease (17%–22%), gastrointestinal (10%–28%), musculoskeletal (10%–32%), non–congenital HD birth defects (12%–23%), and mental health disorder (9%–30%).[13] Neidenbach and colleagues[70] also evaluated noncardiac comorbidities. In a cohort of 821 adults with congenital HD, most (89%) were classified as being in New York Heart Association class I or II, and 53% were classified as having moderate congenital HD according to the American College of Cardiology severity system. Noncardiac comorbidities were assessed across 16 systems; 95% of patients had a significant noncardiac comorbidity, and many patients had >1 noncardiac comorbidity. In addition, adolescent and adult patients with congenital HD are more likely to have acquired diseases such as obesity and diabetes and additive habits such as long-term use of tobacco, vaping, alcohol, or opioids.[71]

Age and the type of comorbidities affect which noncardiac procedure will be performed. Neonates and infants are likely to undergo procedures related to the underlying syndrome or congenital conditions and gastrointestinal procedures such as gastrostomy tube placement to treat feeding disorders. For adults, necessary procedures are more likely to be associated with acquired conditions such as cholecystitis, trauma, pregnancy, or cancer. Many of these procedures may be needed on an urgent or emergency basis, which will amplify the associated risk.

TIMING OF NONCARDIAC PROCEDURES

As previously noted, the complexity of the noncardiac surgical procedure is an important risk factor. Nonetheless, 1 study suggests that the degree of functional impairment and the type of cardiac lesion are more important than the surgical procedure itself.[18] Hence, in patients with complex HD and severely impaired heart function, the need for general anesthesia even for short and minimally invasive procedures remains an important risk factor. Noninvasive imaging procedures (magnetic resonance imaging, computed tomography scanning) or minimally invasive interventional radiology procedures are often performed in clinical environments in which invasive monitoring is not routine and personnel may not have extensive experience managing patients with significant cardiac disease.

When possible, noncardiac surgery should be deferred until the cardiac lesion has been treated and the patient's functional status has been optimized. However, this approach is not always feasible, and many patients require noncardiac procedures before their definitive or palliative intervention or before their functional status can be improved. The risk should always be weighed against the benefits of the procedure, in consultation with a multidisciplinary care team. Preoperative assessment and optimization should include consideration of procedure location and team composition. Recovery should take place in an environment with personnel experienced in the care of patients with cardiac conditions.

LOCATION OF CARE FOR NONCARDIAC PROCEDURES

Maxwell and colleagues[72] analyzed data from a large statewide database of outpatient surgery in the United States to determine the proportion of children and adults with congenital HD undergoing noncardiac surgery, both outside and within congenital HD centers of expertise. The authors determined that children are more likely than adults (57% of children versus only 26% of adults) to undergo noncardiac surgery at a congenital HD center. Both children and adults undergoing a procedure at a non–congenital HD center lived farther away from a congenital HD center: Pediatric patients with congenital HD traveled an average of 18 miles to their surgical facility (which was 17 miles closer than the nearest congenital

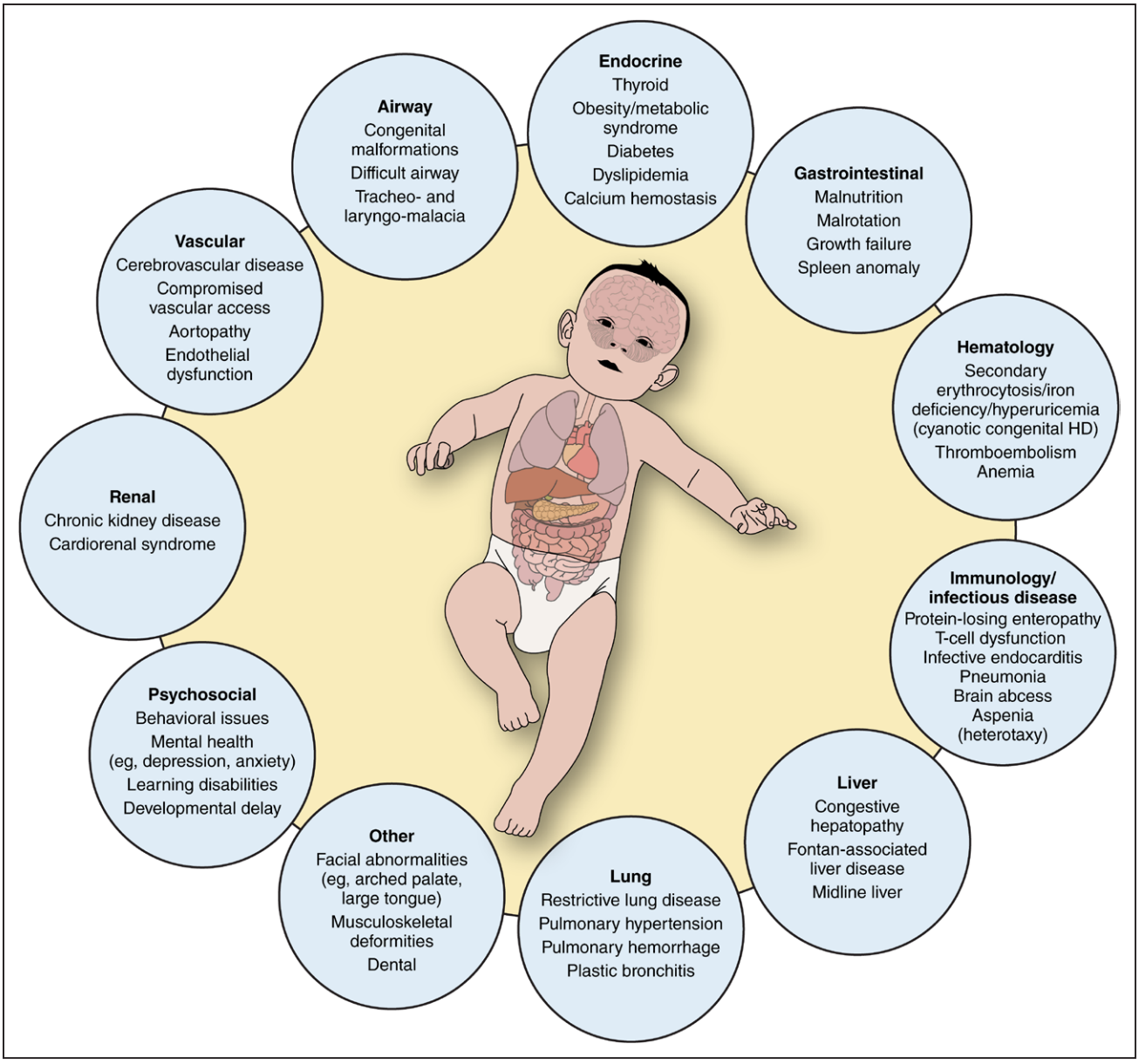

Figure 4. Noncardiac conditions in patients with congenital heart disease.
HD indicates heart disease.

HD center), and adult patients with congenital HD traveled an average of 12 miles to their surgical facility (which was 26 miles closer than the nearest congenital HD center). This is not surprising, given that surveys of parental preferences have revealed that in hypothetical scenarios involving the choice between surgery at a local center and a referral hospital, more families will choose the local center as the distance to the referral hospital increases, even if informed that the local center had a higher surgical mortality rate.[73] This situation is potentially problematic because, although the proceduralist at a non–congenital HD center may have the expertise to perform the procedure, anesthesiologists and other consultants well versed in the care of patients with congenital HD may not be available there.

In 2017, the American Heart Association published a scientific statement titled "Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease."[74] Many of the principles discussed therein apply to the discussion here. In particular, it is optimal to perform the preoperative evaluation, surgical procedure, and postoperative recovery for patients with congenital HD at a regional center with health care professionals who appreciate not only the implications of the congenital HD but also the multisystem age-related factors that can contribute to morbidity and mortality. Recently, a retrospective cohort analysis of pediatric inpatient data from the Center for Healthcare Information and Analysis of the Commonwealth of Massachusetts, Texas Healthcare Information Collection, and Health Care Cost and

Utilization Project State Inpatient Database has shown that patients with congenital HD are more likely to travel to a hospital with a cardiac surgical program for noncardiac procedures than to a hospital without a cardiac surgical program, especially patients with single-ventricle disease, other complex congenital HD, and ≥6 chronic conditions.[75]

EVIDENCE GAPS AND FUTURE DIRECTIONS

There are significant challenges to developing evidenced-based care paradigms for patients with congenital HD undergoing noncardiac surgery. The heterogeneity of the myriad combinations of congenital lesions, comorbidities, and specific surgical and nonsurgical procedures yields small numbers of specific patient types from which to derive guidelines or establish strong recommendations. Comprehensive multicenter databases with extremely granular data are needed to have adequate numbers of patients to address clinical questions during the perioperative period and to characterize the optimal location of care.

Although data suggest that patients with complex congenital HD undergoing noncardiac surgery have improved survival when cared for at a specialized center, many questions remain. For example, it is not clear how these centers should be configured or where they should be located so that patients have reasonable access to the expert care they need. Further work in the field is needed to determine factors that may affect the care of congenital HD patient subgroups during noncardiac surgery and that will facilitate their care while optimizing clinical outcomes.

ARTICLE INFORMATION

The American Heart Association makes every effort to avoid any actual or potential conflicts of interest that may arise as a result of an outside relationship or a personal, professional, or business interest of a member of the writing panel. Specifically, all members of the writing group are required to complete and submit a Disclosure Questionnaire showing all such relationships that might be perceived as real or potential conflicts of interest.

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Disclosures

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*Modest.

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