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Anesthesia-Related Cardiac Arrest in Children with Heart Disease: Data from the Pediatric Perioperative Cardiac Arrest (POCA) Registry

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BACKGROUND: From 1994 to 2005, the Pediatric Perioperative Cardiac Arrest Registry collected data on 373 anesthesia-related cardiac arrests (CAs) in children, 34% of whom had congenital or acquired heart disease (HD).
METHODS: Nearly 80 North American institutions that provide anesthesia for children voluntarily enrolled in the Pediatric Perioperative Cardiac Arrest Registry. A standardized data form for each perioperative CA in children 18 years old or younger was submitted anonymously. We analyzed causes of and outcomes from anesthesia-related CA in children with and without HD.
RESULTS: Compared with the 245 children without HD, the 127 children with HD who arrested were sicker (92% vs 62% ASA physical status III–V; $P < 0.01$) and more likely to arrest from cardiovascular causes (50% vs 38%; $P = 0.03$), although often the exact cardiovascular cause of arrest could not be determined. Mortality was higher in patients with HD (33%) than those without HD (23%, $P = 0.048$) but did not differ when adjusted for ASA physical status classification. More than half (54%) of the CA in patients with HD were reported from the general operating room compared with 26% from the cardiac operating room and 17% from the catheterization laboratory. The most common category of HD lesion in patients suffering CA was single ventricle ($n = 24$). At the time of CA, most patients with congenital HD were either unrepaired (59%) or palliated (26%). Arrests in patients with aortic stenosis and cardiomyopathy were associated with the highest mortality rates (62% and 50%, respectively), although statistical comparison was precluded by small sample size for some HD lesions.
CONCLUSIONS: Children with HD were sicker compared with those without HD at the time of anesthesia-related CA and had a higher mortality after arrest. These arrests were reported most frequently from the general operating room and were likely to be from cardiovascular causes. The identification of causes of and factors relating to anesthesia-related CA suggests possible strategies for prevention. (Anesth Analg 2010;110:1376–82)

The Pediatric Perioperative Cardiac Arrest (POCA) Registry was formed in 1994 to study the causes of and outcomes from perioperative cardiac arrests (CAs) in anesthetized children. The initial findings of the POCA Registry, published in 2000, analyzed data from 150 anesthesia-related arrests from 1994 to 1997.[1] A subsequent report in 2007 of 193 anesthesia-related CAs from 1998 to 2004 allowed comparison of the CA profile between the 2 time periods.[2] Of the 373 cases of anesthesia-related CA reported to the POCA Registry through 2005, 127 patients (34%) had congenital or acquired heart disease (HD). To our knowledge, this is the largest known collection of anesthesia-related CA in children with HD. We wished to identify factors associated with and outcomes from perioperative CA in this patient population, when compared with CA in children without HD.

METHODS

This study was approved by the University of Washington IRB, and the requirement for written informed consent was waived. The POCA Registry was formed in 1994; from that year through 2005, data were collected from voluntary enrollment of institutions in the United States and Canada that provide anesthetic care to children. During this time, an average of 68 (range 58–79) institutions enrolled in the Registry each year. Seventy-two percent of these institutions were university-affiliated hospitals, 16% were community hospitals, and 4% were government or military hospitals.

The data collection process has been described in detail.[1] Briefly, a designated representative from each participating institution submitted an anonymous standardized data form for all cases of CA (defined as the administration of chest compressions or as death) that occurred in children 18

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years or younger during administration of or recovery from anesthesia. Cardiac arrests in the pediatric and neonatal intensive care units or on the ward were excluded.

The standardized data form included patient demographics, prearrest status, surgical procedure, personnel involved in anesthetic care, anesthetic drugs, techniques and monitors, antecedent events, the immediate cause of CA, and details of the resuscitation. Possible causes for CA were previously defined.[1] The institutional representatives assessed the contribution of anesthesia, surgery, and underlying patient disease to the causation of CA as none, minor, major, or total. The institutional representatives also submitted a narrative summary specifying the sequence of events and causal relationships associated with the CA and providing any relevant information not included in the standardized data form.

Outcome was assessed using a modification of a 10-point severity of injury scale[3] applied 24 hours after the CA and at the last clinical evaluation if the patient survived >24 hours. No time limit was applied to any death, as long as the death was attributed to the anesthetic-related CA. Autopsy findings, if available, were also included.

A minimum of 3 members of the POCA Registry Steering Committee (CR, CMH, SMB, JPM) reviewed all data forms and categorized each CA as anesthesia-related, not anesthesia-related, or unknown according to previously published definitions.[1] A CA was designated as anesthesia-related if anesthesia personnel or the anesthesia process played at least some role (ranging from minor to total) in the genesis of the CA. Disagreements were resolved by discussion until consensus was reached. A similar approach was taken in assigning cause of CA. The institutional assignment of cause of CA was retained unless overruled by consensus of the Steering Committee. Cases of inability to wean from cardiopulmonary bypass were categorized as not related to anesthesia when there were no anesthesia-related problems in the prebypass period; such cases ($n = 74$) were not included in the data analysis.

Heart disease lesions were grouped based on underlying pathophysiology into the following categories: single ventricle; Tetralogy of Fallot; truncus arteriosus; left-to-right shunt (atrial or ventricular septal defect, patent ductus arteriosus, and atrioventricular canal); obstructive lesion (aortic stenosis, pulmonic stenosis, and coarctation of the aorta); cardiomyopathy; and other. Patients with multiple defects were placed in the category that best defined their primary physiologic abnormality. Patients with cardiac lesions of trivial or no hemodynamic significance were grouped with the non-HD patients, as were patients with a history of patent ductus arteriosus that resolved without surgical correction. Lesions were classified as congenital if present at birth and as acquired if they developed postpartum. Lesions were also classified as unrepaired (no surgery), palliated (partial surgical repair with residual anatomic defects), or repaired (surgical repair with no residual anatomic defects). Lesions were only categorized as repaired if the procedure left the patient with no anatomic or physiologic deficit (e.g., atrial septal defect or isolated coarctation of the aorta). Single ventricle patients included those pre- and post-Stage I repair (i.e., Norwood-type procedure), post-Stage II repair (i.e., superior cavopulmonary anastomosis), and post-Fontan completion (i.e., addition of inferior cavopulmonary anastomosis). Postoperative patients with single ventricle lesions, even after Fontan completion, were placed in the palliated category. The phase of care when CA occurred included presurgical (preinduction and induction), surgical (maintenance), and postsurgical (emergence, transport, and recovery).

Table 1. Demographics of Patients With and Without Heart Disease

	Patients with HD ($n = 127$)	Without HD ($n = 245$)	P
Age			0.071
0–6 mo	59 (47%)	94 (39%)	
7 mo to 12 mo	8 (6%)	24 (10%)	
>1–2 y	22 (17%)	35 (14%)	
>2–5 y	13 (10%)	31 (13%)	
6–10 y	6 (5%)	28 (11%)	
11–18 y	18 (14%)	32 (13%)	
Emergency	18 (14%)	59 (24%)	0.030
ASA physical status			<0.001
I–II	10 (8%)	92 (38%)	
III–IV	117 (92%)	153 (62%)	

P values by Fisher exact test.
HD = heart disease.

Comparisons between patients with and without HD were performed with Pearson χ^2 test, Fisher exact test, Student t test, and the z test for proportions between the HD and non-HD groups. Statistical significance was defined as values of $P < 0.05$.

RESULTS

From 1994 to 2005, 373 cases of anesthesia-related CA were reported to the POCA Registry and 372 could be classified according to their HD status. One patient was excluded because of ambiguous HD status. One hundred twenty-seven CAs (34%) occurred in patients with HD. A comparison of the demographic profile of the patients with and without ($n = 245$) HD is shown in Table 1. Of the anesthesia care providers, 82% had fellowship training in pediatric anesthesiology and 95% were certified by the American Board of Anesthesiologists (or the equivalent). There was no difference in training and board certification between providers for patients with and without HD. Nearly half (47%) of all CAs in children with HD occurred at the age of 6 months or younger, and 70% occurred in those 2 years or younger; the age distribution for children without HD was similar ($P = 0.071$). Emergency surgery was less common in children with HD compared with children without HD ($P = 0.030$). Nearly all (92%) children with HD were ASA physical status III–V compared with 62% in children without HD ($P < 0.001$). Ten HD patients were classified as ASA physical status I–II by the referring institution, including patients with left-to-right shunt lesions, Ebstein's anomaly, pulmonary hypertension, sick sinus syndrome, heart block, Wolff-Parkinson-White syndrome, prolonged QT syndrome, and cardiomyopathy. The Pediatric Perioperative Cardiac Arrest Steering Committee reviewers did not alter physical status classifications, even when inaccuracy was suspected.

Table 2 lists the cardiac lesions in children with HD and the number of reports of CA for each. The most common

Table 2. Cardiac Lesions in Children with Heart Disease

Lesion	*n* (% of 127)
Single ventricle	24 (19%)
Hypoplastic left heart syndrome	9
Double outlet right ventricle	5
Unbalanced AV canal	4
Tricuspid atresia	3
Pulmonary atresia	2
Double inlet left ventricle	1
Left-to-right shunt	23 (18%)
Ventricular septal defect	9
Patent ductus arteriosus	5
Atrioventricular canal	4
Combined lesions (ASD, VSD, PDA)	5
Obstructive lesions	20 (16%)
Aortic stenosis	13[a]
Coarctation of the aorta	6
Aortic obstruction	1
Cardiomyopathy	16 (13%)
Dilated	4
Hypertrophic	2
Restrictive	1
Disease specific	
Duchenne muscular dystrophy	4
Renal disease	2
AIDS	1
Unspecified	2
Tetralogy of Fallot	15 (12%)
Truncus arteriosus	6 (5%)
Miscellaneous	23 (18%)
Pulmonary hypertension	4
Status post-heart transplant	3
Heart block	3
Wolff-Parkinson-White	2
Other[b]	11

AV = atrioventricular; ASD = atrial septal defect; VSD = ventricular septal defect; PDA = patent ductus arteriosus; AIDS = acquired immunodeficiency syndrome.

[a] Two with Williams syndrome and 4 with pulmonary stenosis.

[b] Other includes anomalous pulmonary veins, coronary artery disease, Ebstein's anomaly, interrupted aortic arch, left ventricular hypertrophy, myocarditis, prolonged QT syndrome, sick sinus syndrome, systemic hypertension, transposition of the great vessels, and unspecified (1 each).

type of defect reported was single ventricle (n = 24; 19%). Seventeen CAs in this category occurred in patients before superior cavopulmonary anastomosis, 2 occurred in patients after this procedure, and 5 occurred in patients after Fontan completion. The next most common lesions were those resulting in left-to-right shunt (n = 23; 18%) and in obstruction to ventricular outflow (n = 20; 16%). Aortic stenosis was the most common obstructive lesion (n = 13). Cardiomyopathy was the most common of the acquired defects (n = 16; 13%). Most patients had defects that were either unrepaired (59%) or palliated (26%). Only 13% had lesions that were fully repaired at the time of CA.

Location of Arrests

Cardiac arrests in children with HD were reported most frequently from the general operating room (OR) (n = 69; 54%). The most frequent surgical procedures were gastrointestinal procedures (e.g., fundoplication, gastrostomy tube placement, esophagogastroduodenoscopy, and colostomy; n = 17). These procedures were followed in frequency by ear, nose, and throat procedures (myringotomy with tubes, bronchoscopy, choanal atresia repair, and tracheostomy; n = 16) and by placement of permanent central venous catheters (Broviac catheter or Port-A-Cath; n = 13). Urologic, orthopedic, ophthalmologic, plastics, dental, and thoracic procedures each accounted for 5 cases or less. There were 2 laparoscopic procedures and no thoracoscopic procedures.

There were 33 CAs (26%) reported from the cardiac OR, 21 (17%) from the cardiac catheterization laboratory, and 4 (3%) from other imaging suites (1 from the pulmonary laboratory and 3 from the magnetic resonance imaging suite). The patients reported from the cardiac OR were younger than those from other locations: 76% of CAs from the cardiac OR were in patients younger than 6 months, compared with 38% from the general OR and 35% from the catheterization laboratory (P = 0.001).

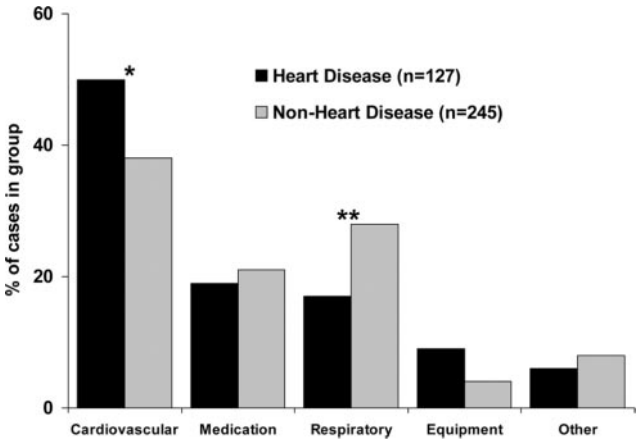

Figure 1. Causes of anesthesia-related cardiac arrest associated with heart disease (n = 127) versus nonheart disease (n = 245). *P = 0.03, **P = 0.01.

Phase of Anesthesia During Arrest

Cardiac arrest in children with HD occurred most commonly (48%) during the surgical (maintenance) phase of the anesthetic, compared with 36% and 16% during the presurgical and postsurgical periods, respectively. This distribution was statistically the same as for those patients without HD. There was no statistical association between location of CA and phase of care when the CA occurred in patients with HD.

Etiology of Arrest

Causes of CA in patients with and without HD are shown in Figure 1. Cardiovascular causes of CA were most common in both groups but occurred more frequently in patients with HD than in those without (50% vs 38%; P = 0.03). Respiratory causes of CA were more common in patients without HD (28% vs 17%; P = 0.01). A breakdown of cardiovascular, medication, respiratory, and equipment causes of CA in patients with HD is shown in Table 3. In about half of the patients (33 of 63) who had a cardiovascular cause of CA, the exact etiology of the CA could not be determined. In 9 of these cases, cardiovascular depression from 1 or more anesthetic drugs was a likely contributory factor: 6 from halothane and 1 each from propofol, sevoflurane, and the combination of fentanyl and midazolam. In 7

Table 3. Causes of Cardiac Arrest in Patients with Heart Disease

Cause of arrest	*n* (% of 127)
Cardiovascular	63 (50%)
Myocardial ischemia	5 (4%)
Hyperkalemia	3 (2%)
"Tet" spell	3 (2%)
Hypovolemia: preexisting	3 (2%)
Sudden arrhythmia	3 (2%)
Hypovolemia-blood loss	2 (2%)
Other miscellaneous CV cause[a]	11 (9%)
Presumed CV: unclear etiology	33 (26%)
Medication	25 (20%)
Inhaled anesthetic CV depression	
Halothane	8 (6%)
Sevoflurane	6 (5%)
Isoflurane	1 (1%)
Intravenous drug CV depression	
Propofol	1 (1%)
Narcotics	1 (1%)
Wrong dose	3 (2%)
Medication combinations	2 (2%)
Other[b]	3 (2%)
Respiratory	21 (17%)
Laryngospasm	6 (5%)
Inadequate oxygenation	6 (5%)
Difficult intubation	2 (2%)
Airway obstruction	2 (2%)
Other miscellaneous respiratory cause[c]	5 (4%)
Equipment	11 (9%)
Central line complications	9 (7%)
Breathing circuit obstruction	1 (1%)
Endotracheal tube obstruction	1 (1%)
Multiple events	3 (2%)
Unknown cause	4 (3%)

CV = cardiovascular.

[a] One case each: air embolism, hypovolemia from surgical retraction, left ventricular outflow obstruction, pacemaker failure, right-to-left shunt, severe valvular dysfunction, vagal response, acidosis, pulmonary hypertensive crisis, myocardial dysfunction, and severe coronary artery disease.

[b] One case each: epinephrine-induced ventricular fibrillation, prostacyclin effect, and intravascular injection of local anesthetic.

[c] One case each: esophageal intubation, premature extubation, pneumothorax, endobronchial intubation, presumed respiratory, and cause unclear.

cases, myocardial ischemia was likely but not certain. The most common known cardiovascular cause of CA was myocardial ischemia ($n = 5$).

An imbalance between pulmonary and systemic vascular resistance and flow was implicated in 7 CAs. In the single ventricle category, 4 CAs resulted or probably resulted from inadequate systemic (and therefore coronary artery) flow due to low pulmonary vascular resistance and pulmonary overcirculation. In the left-to-right shunt category, 3 CAs resulted from systemic desaturation due to pulmonary hypertension and right-to-left shunting.

Medication-related CAs in HD patients were most commonly due to cardiovascular depression from inhalation anesthetics (Table 3). Cardiac arrests related to halothane administration occurred before 2000; most CAs related to propofol and sevoflurane administration were reported after 2000. Frequently, CAs due to the cardiovascular depressant effects of propofol and sevoflurane were reported in patients with compromised ventricular function from a wide variety of causes.

Of the 21 CAs due to respiratory causes, laryngospasm resulting in hypoxemia and bradycardia ($n = 6$) and other causes of inadequate oxygenation ($n = 6$) were most common. In these patients, baseline low saturations (i.e., those on the steep portion of the oxyhemoglobin dissociation curve) led to a rapid deterioration in oxygen delivery after the critical event.

Eleven CAs were caused by equipment problems, including 9 that resulted from complications of placement of central venous pressure catheters. Seven of the 9 occurred in newborns (<30 days old) as a result of needle injury to the airway, lung, or a vascular structure.

There was no statistical difference in causes of CA when the combined locations of general OR and imaging suite were compared with the combined locations of the cardiac OR and catheterization laboratory ($P = 0.09$). The only exception was a trend toward more equipment-related CAs (i.e., central venous catheter complications) in the cardiac OR/catheterization laboratory.

Of the CAs reported from the magnetic resonance imaging suite, one may have been prevented had the anesthetic been given in an OR location: a sevoflurane-related CA in a 7 year old with Down Syndrome status post-atrioventricular canal repair. The quality of the electrocardiogram was poor, resulting in delayed diagnosis of asystole.

Resuscitation

Resuscitation details were available for 68 HD patients from 1998 to 2005, including 49 survivors and 19 nonsurvivors. Nonsurvivors had a longer total duration of resuscitation (53 minutes vs 6 minutes; $P = 0.001$), a larger number of drugs (3.4 vs 2.5; $P = 0.046$), and more rounds of drugs (3.6 vs 2.2; $P = 0.038$) compared with survivors. There was no statistical difference between nonsurvivors and survivors in the delay until start of resuscitation (<1 minute) or in the use of defibrillation (26% vs 14%, $P = 0.294$). Epinephrine alone was used in 23 survivors (47%) and 6 nonsurvivors (32%); in both groups, epinephrine was the intervention most often associated with a return of circulation. Cardiopulmonary bypass ($n = 4$) and extracorporeal membrane oxygenation (ECMO; $n = 3$) were used occasionally when standard resuscitative measures were unsuccessful. One patient survived from each group. The longest resuscitation resulting in an intact survivor occurred in a patient placed on ECMO 35 minutes after CA.

Outcome After Arrest

The overall mortality rate for patients with HD was higher than that for patients without HD (33% vs 23%; $P = 0.048$). However, when mortality rates for ASA physical status III–V patients from both groups were compared, there was no difference (34% for both). Within the HD groups, multiple possible factors predictive of mortality were examined. There was no association with age, phase of care, cause of CA, or year of CA. Mortality rates for ASA physical status III–V patients and ASA physical status I–II patients (34% vs 20%) were not statistically different. Mortality rates after CA in the cardiac OR, general OR, and catheterization laboratory were 45%, 26%, and 33%, respectively; P = NS).

Mortality rates from the various HD categories, including the surgical repair status, are shown in Table 4. Although statistical comparison of mortality by category or lesion was precluded by small sample size in some categories, the highest mortality rates were seen in patients with

Table 4. Lesion Surgical Status and Mortality

	No. of cases	Lesion surgical status[a]			Mortality
		Unrepaired	Palliated	Repaired	Died
Single ventricle	24	5 (21%)	19 (79%)	0 (0%)	6 (25%)
Left to right shunts (ASD, VSD)	23	14 (61%)	0 (0%)	9 (39%)	4 (17%)
Obstructive (coarctation/AS/PS)	20	15 (75%)	3 (15%)	2 (10%)	9 (45%)
Aortic stenosis	13	10 (77%)	2 (15%)	1 (8%)	8 (62%)
Cardiomyopathy	16	15 (94%)	1 (6%)	0 (0%)	8 (50%)
Tetralogy of fallot	15	5 (36%)	7 (50%)	2 (14%)	3 (20%)
Truncus arteriosus	6	5 (83%)	0 (0%)	1 (17%)	2 (33%)

Percentages based on number of cases with this lesion (row total).
ASD = atrial septal defect; VSD = ventricular septal defect; coarctation = coarctation of the aorta; AS = aortic stenosis; PS = pulmonary stenosis.
[a] Cases with unknown status excluded.

aortic stenosis (62%) and cardiomyopathy (50%). Overall mortality for patients with single ventricle was 25%. However, single-ventricle patients had a bimodal distribution of CA according to surgical stage: 17 were either pre-stage 1 ($n = 5$) or post-stage 1 ($n = 12$) palliation; the combined mortality for this group was 35%. Another 5 patients with single ventricle had undergone the Fontan procedure (complete palliation); mortality for these patients and for the 2 patients who had superior cavopulmonary anastomosis was zero. The lowest mortality (17%) was seen in patients with left-to-right shunt lesions. Eleven of the 23 CAs in this category were due to respiratory events (e.g., laryngospasm, difficult intubation, and esophageal intubation) with successful resuscitation in all but 1 case.

Surgical repair status had a statistical association with mortality rate. Patients with unrepaired lesions had a higher mortality rate (43%) compared with those with palliated (27%) or completely repaired (6%) lesions ($P = 0.006$).

Seven (6%) of the 127 children with HD survived CA but suffered significant central nervous system (CNS) injuries. These included new onset of seizure activity without other deficits ($n = 2$), parietal and occipital infarcts ($n = 1$), global cerebral ischemia, ($n = 1$), grade IV intraventricular hemorrhage ($n = 1$), left-sided paresis ($n = 1$), and 1 unspecified CNS injury. Non-HD survivors had a similar incidence of residual CNS injuries (5%).

DISCUSSION

The 127 cases of anesthesia-related CA in children with congenital and acquired HD submitted to the POCA Registry from 1994 to 2005 are the largest known collection of such events. Compared with patients without HD submitted to the Registry over the same period, patients with HD were sicker and were less likely to survive the CA, even though they were more likely to be having elective surgery. Patients with HD were more likely to have CA from cardiovascular causes, although in many the exact cause of CA could not be determined.

More CAs in HD patients were reported during noncardiac surgery in the general OR (54%) than during cardiac surgery (26%) and cardiac catheterization (17%) combined, perhaps because noncardiac surgery is more common than cardiac surgery in HD patients. Causes of CA in HD patients were not different comparing the general OR/imaging suite to the cardiac OR/catheterization laboratory, suggesting that underlying cardiac pathophysiology may be more important than location or type of surgery as a predictor of risk. It also suggests that any preventive measures should be directed toward patients in all locations.

Three-quarters of the CAs in patients with HD (and 63% in patients without HD) occurred in children younger than 2 years old. The association with young age was especially true in the cardiac OR, where 75% of the CAs occurred in infants younger than 6 months old, most often in those with unrepaired HD lesions.

Not surprisingly, resuscitation took less time and required fewer drugs in survivors compared with nonsurvivors. The success of epinephrine compared with other interventions in restoring circulation was a notable feature of resuscitative efforts in HD patients.

Optimally, assessment of risk of CA by cardiac lesion would require precise denominator data for calculation of incidence.[4] Because those denominators (e.g., number of anesthetics given to patients in each category) were not consistently provided to the POCA Registry, risk can only be assessed according to the number of CAs submitted and the mortality rate after arrest for each lesion. The most common heart lesions were in the single ventricle category. Higher than average mortality was seen in patients with aortic stenosis, cardiomyopathy, and single ventricle (before superior cavopulmonary anastomosis). These 3 groups accounted for >75% of all deaths reported to the POCA Registry in patients with HD. Across the spectrum of HD lesions, mortality was higher in those who were unrepaired at the time of CA compared with those who were palliated or completely repaired.

Patients with single ventricle have been described as high risk in other studies. The Risk Adjustment for Congenital Heart Surgery, created to understand differences in mortality among patients undergoing congenital heart surgery,[5] placed the stage 1 procedure for hypoplastic left heart syndrome in the highest risk category (expected mortality of 40%–60%, depending on institution[6]). Torres et al.[7] reported a 19% mortality rate for noncardiac surgery in patients younger than 2 years with hypoplastic left heart syndrome.

Patients with single ventricle and a failing Fontan also may be at increased risk. Five arrests reported to the POCA Registry in patients with single ventricle occurred in teenagers with Fontan palliation and myocardial dysfunction. Such patients are susceptible to myocardial depression when exposed to anesthetic drugs. Sevoflurane can reduce myocardial contractility[8] and cause hypotension in patients with HD and compromised ventricular function.[9] Similarly, propofol administration may result in sympatholysis,[10] a

reduction in systemic vascular resistance, and myocardial depression.[11,12]

Concerns over anesthetic medication-related myocardial depression are also appropriate in patients with cardiomyopathy. In this report, anesthetic drug-induced hypotension was partially responsible for CA in at least 3 patients with cardiomyopathy. Resuscitation in this subset of patients was unsuccessful in 50% of cases. Kipps et al.[13] reviewed outcomes after anesthesia and surgery in 26 children with cardiomyopathy, including 21 with severe myocardial dysfunction. Sixty-one percent of the patients became hypotensive and required inotropic support. Two patients required ECMO, and 1 patient died.

Patients with aortic stenosis also warrant concern, based on the number of cases of CA reported to the POCA Registry, and on the 62% mortality rate after arrest. The Risk Adjustment for Congenital Heart Surgery classification scheme lists aortic valvotomy or valvuloplasty in infants at risk category 4 of 6.[6] Of the 8 deaths after CA in patients with aortic stenosis, 5 had clinical evidence and 1 had autopsy evidence of myocardial ischemia. Aortic stenosis predisposes to myocardial ischemia for 2 reasons: (1) coronary artery anatomy may be abnormal, as in patients with Williams Syndrome[14] and (2) the hypertrophied and hypertensive ventricle may suffer inadequate oxygenation in the presence of reduced diastolic blood pressure, tachycardia, bradycardia, arrhythmias, hypoxemia, or anemia.

Recommendations concerning strategies to prevent CA, although speculative, follow from an understanding of causes of and factors relating to anesthesia-related CA in children with HD. For example, of the 24 CAs in patients with single ventricle reported to the POCA Registry, 17 CAs and 6 deaths occurred before superior cavopulmonary anastomosis. In the setting of parallel systemic and pulmonary circulations, a decrease in pulmonary vascular resistance increases pulmonary blood flow and reduces systemic blood pressure and flow; these changes may result in a decrease in coronary perfusion, resulting in myocardial ischemia. Preventive strategies for dealing with patients with parallel circulations must be focused on maintaining the balance between the pulmonary and systemic circulations in the perioperative period. Inspired oxygen concentrations must be limited and end-tidal carbon dioxide must be maintained at normal or slightly elevated levels to avoid potentially lethal reductions in pulmonary vascular resistance.

Patients with cardiomyopathy or a failing Fontan may have severely compromised ventricular contractility. Such patients require careful titration of anesthetics, because standard doses or concentrations may not be tolerated. Infusions of vasoactive substances such as dopamine may be required to maintain hemodynamic stability during anesthetic administration. In addition, alternative anesthetic drugs may be useful in patients with compromised ventricular function. Ketamine supports sympathetic tone and when administered to children with HD results in stable hemodynamics.[15,16] When compared directly to propofol in children with congenital HD, ketamine caused fewer episodes of hypotension.[17] Etomidate has been shown to result in no significant changes in hemodynamics when administered to children in the catheterization laboratory[18] and therefore is potentially suitable for induction.

Myocardial ischemia is an important cause of CA in the current report and also has been emphasized in a report by Odegard et al.[19] Myocardial ischemia can result from either a decrease in oxygen supply or an increase in oxygen demand in the susceptible patient. Many of the factors influencing the myocardial oxygen supply/demand ratio are under the direct control of the anesthesiologist. Preventive strategies include maintaining adequate oxygenation, hematocrit, preload, and afterload while avoiding arrhythmias and extremes of heart rate.

Complications of central line placement also are potentially preventable. Damage to large or small airways, heart, or great vessels resulted from needle or wire penetration. Most of these events occurred in newborns or infants; providers used anatomic landmarks alone for guidance, suggesting that technical difficulties played a role. Use of ultrasound guidance has been shown to improve success rates and decrease the complication rate for central venous cannulation in children with congenital HD.[20]

For patients failing conventional resuscitative measures, the use of ECMO has improved survival in patients who would otherwise die. Flick et al.[21] from the Mayo Clinic reported a 54% survival rate in patients placed on ECMO after failing to wean from cardiopulmonary bypass. The Extracorporeal Life Support Organization has reported a 30% survival rate in children (with and without HD) who were placed on ECMO after failing conventional cardiopulmonary resuscitation.[22] In this report, the presence of HD was one of the pre-ECMO factors associated with an improved survival rate.

The POCA Registry has several methodological weaknesses, as previously described.[1] First, enrollment into the Registry and reporting of cases was purely voluntary and underreporting was likely. Selection bias was also likely, and highly sensitive cases may not have been reported. Self-reporting may also have led to inaccuracies in the data and underestimates of the anesthetic-related elements of a CA. Second, participation in the POCA Registry was skewed toward university-affiliated children's hospitals, whereas a large percentage of pediatric anesthesia services in North America are delivered in smaller community hospitals. Third, the inclusion criteria for the POCA Registry were initiation of chest compressions or death. The Registry did not include cases in which a CA was successfully treated with defibrillation but not chest compressions. Fourth, the data were gathered retrospectively, often by a person not directly involved in the anesthetic care; lack of concurrent first-hand information may have led to inaccuracies. Finally, we cannot provide incidence figures for perioperative pediatric CA, given the lack of adequate denominators, including total number of anesthetics provided in each participating institution in each HD category.

SUMMARY

One hundred twenty-seven anesthesia-related CAs in children with HD were reported to the POCA Registry. These children were sicker, more likely to arrest from cardiovascular causes, and less likely to survive than children without HD. More than half of all CAs in children with HD occurred in the general OR. Patients with single ventricle, aortic stenosis, and cardiomyopathy were notable because

of the report frequency of CA and the mortality rates after CA. Children with unrepaired HD lesions had poorer outcome than those who had been palliated or completely repaired. Implementation of preventive strategies based on causes and related factors for CA in children with HD may reduce the incidence of these adverse events.

DISCLAIMER

All opinions expressed are those of the authors and do not necessarily reflect those of the ASA.

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REFERENCES

1. Morray JP, Geiduschek, JM, Ramamoorthy C, Haberkern CM, Hackel A, Caplan RA, Domino KB, Posner K, Cheney FW. Anesthesia-related cardiac arrest in children: initial findings of the Pediatric Perioperative Cardiac Arrest (POCA) Registry. Anesthesiology 2000;93:6–14
2. Bhananker SM, Ramamoorthy C, Geiduschek JM, Posner KL, Domino KB, Haberkern CM, Campos JS, Morray JP. Anesthesia-related cardiac arrest in children: update from the Pediatric Perioperative Cardiac Arrest Registry. Anesth Analg 2007;105:344–50
3. Brunner EA. Analysis of anesthetic mishaps. The National Association of Insurance Commissioners' closed claims study. Int Anesthesiol Clin 1984;22:17–30
4. Flick RP, Sprung J, Harrison TE, Gleich SJ, Schroeder DR, Hanson AC, Buenvenida SL, Warner DO. Perioperative cardiac arrests in children between 1988 and 2005 at a tertiary referral center: a study of 92,881 patients. Anesthesiology 2007;106: 226–37
5. Jenkins KJ, Gauvreau K, Newberger JW, Spray T, Moller JH, Iezzoni L. Consensus-based method for risk-adjustment for surgery for congenital heart disease. J Thorac Cardiovasc Surg 2002;123:110–8
6. Jenkins KJ, Gauvreau K. Center-specific differences in mortality: preliminary analyses using the Risk Adjustment in Congenital Heart Surgery (RACHS-1) method. J Thorac Cardiovasc Surg. 2002;124:97–104
7. Torres A, DiLiberti J, Pearl RH, Wohrley J, Raff GW, Bysani K, Bond LM, Geiss DM. Noncardiac surgery in children with hypoplastic left heart syndrome. J Pediatr Surg 2002;37: 1399–1403
8. Wodey E, Pladys P, Copin C, Lucas MM, Chaumont A, Carre P, Lelong B, Azzis O, Ecoffey C. Comparative hemodynamic depression of sevoflurane versus halothane in infants: an echocardiographic study. Anesthesiology 1997;87:795–800
9. Russell IA, Miller-Hance WC, Gregory G, Balea MC, Cassorla L, DeSilva A, Hickey RF, Reynolds LM, Rouine-Rapp K, Hanley FL, Reddy VM, Cahalan M. The safety and efficacy of sevoflurane anesthesia in infants and children with congenital heart disease. Anesth Analg 2001;92:1152–8
10. Ebert TJ, Muzi M, Berens R, Goff D, Kampine JP. Sympathetic responses to induction of anesthesia in humans with propofol or etomidate. Anesthesiology 1992;76:725–33
11. Williams GD, Jones TK, Hanson KA, Morray JP. The hemodynamic effects of propofol in children with congenital heart disease. Anesth Analg 1999;89:1411–6
12. Stowe DF, Bosnjak ZJ, Kampine JP. Comparison of etomidate, ketamine, midazolam, propofol and thiopental on function and metabolism in isolated hearts. Anesth Analg 1992;74:547–58
13. Kipps AK, Ramamoorthy C, Rosenthal DN, Williams GD. Children with cardiomyopathy: complications after noncardiac procedures with general anesthesia. Pediatric Anesthesia 2007;17:775–81
14. Burch TM, McGowan FX, Kussman BD, Powell AJ, DiNardo JA. Congenital supravalvular aortic stenosis and sudden death associated with anesthesia: what's the mystery? Anesth Analg 2008;107:1848–54
15. Morray JP, Lynn AM, Herndon PS, Kawabori I, Stevenson JG. Hemodynamic effects of ketamine in children with congenital heart disease. Anesth Analg 1984;63:895–9
16. Williams GD, Philip BM, Chu LF, Boltz MG, Kamra K, Terwey H, Hammer GB, Perry SB, Feinstein JA, Ramamoorthy C. Ketamine does not increase pulmonary vascular resistance in children with pulmonary hypertension undergoing sevoflurane anesthesia and spontaneous ventilation. Anesth Analg 2007;105:1578–84
17. Lebovic S, Reich DL, Steinberg LG, Vela FP, Silvay G. Comparison of propofol versus ketamine for anesthesia in pediatric patients undergoing cardiac catheterization. Anesth Analg 1992;74:490–4
18. Sarkar M, Laussen PC, Zurakowski D, Shukla A, Kussman B, Odegard KC. Hemodynamic responses to etomidate on induction of anesthesia in pediatric patients. Anesth Analg 2005;101:645–50
19. Odegard KC, DiNardo JA, Kussman BD, Shukla A, Harrington J, Casta A, McGowan FX, Hickey PR, Bacha EA, Thiagarajan RR, Laussen PC. The frequency of anesthesia-related cardiac arrests in patients with congenital heart disease undergoing cardiac surgery. Anesth Analg 2007;105:335–43
20. Verghese ST, McGill WA, Patel RI, Sell JE, Midgley FM, Ruttimann UE. Ultrasound-guided internal jugular venous cannulation in infants. Anesthesiology 1999;91:71–7
21. Flick RP, Sprung J, Gleich SJ, Barnes RD, Warner DO, Dearani JA, Scott PD, Hanson AC, Schroeder DR, Schears GJ. Intraoperative extracorporeal membrane oxygenation and survival of pediatric patients undergoing repair of congenital heart disease. Paediatr Anaesth 2008;18:757–66
22. Thiagarajan RR, Laussen PC, Rycus PT, Bartlett RH, Bratton SL. Extracorporeal membrane oxygenation to aid cardiopulmonary resuscitation in infants and children. Circulation 2007;116: 1693–700