

# Perioperative Outcomes of Major Noncardiac Surgery in Adults with Congenital Heart Disease

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## ABSTRACT

**Background:** An increasing number of patients with congenital heart disease are surviving to adulthood. Consensus guidelines and expert opinion suggest that noncardiac surgery is a high-risk event, but few data describe perioperative outcomes in this population.

**Methods:** By using the Nationwide Inpatient Sample database (years 2002 through 2009), the authors compared patients with adult congenital heart disease (ACHD) who underwent noncardiac surgery with a non-ACHD comparison cohort matched on age, sex, race, year, elective or urgent or emergency procedure, van Walraven comorbidity score, and primary procedure code. Mortality and morbidity were compared between the two cohorts.

**Results:** A study cohort consisting of 10,004 ACHD patients was compared with a matched comparison cohort of 37,581 patients. Inpatient mortality was greater in the ACHD cohort (407 of 10,004 [4.1%] *vs.* 1,355 of 37,581 [3.6%]; unadjusted odds ratio, 1.13;  $P = 0.031$ ; adjusted odds ratio, 1.29;

### What We Already Know about This Topic

- The prevalence of congenital heart disease in adults is increasing, but the risk for these patients having noncardiac surgery has not been well described

### What This Article Tells Us That Is New

- In an administrative database of over 10,000 adults with congenital heart disease undergoing major noncardiac surgery, in-hospital mortality was increased compared with a well-matched comparison cohort
- Adult congenital heart disease is an independent predictor of increased perioperative mortality

$P < 0.001$ ). The composite endpoint of perioperative morbidity was also more commonly observed in the ACHD cohort (2,145 of 10,004 [21.4%] *vs.* 6,003 of 37,581 [16.0%]; odds ratio, 1.44;  $P < 0.001$ ). ACHD patients comprised an increasing proportion of all noncardiac surgical admissions over the study period ( $P$  value for trend is  $<0.001$ ), and noncardiac surgery represented an increasing proportion of all ACHD admissions ( $P$  value for trend is  $<0.001$ ).

**Conclusions:** Compared with a matched control cohort, ACHD patients undergoing noncardiac surgery experienced increased perioperative morbidity and mortality. Within the limitations of a retrospective analysis of a large administrative dataset, this finding demonstrates that this is a vulnerable population and suggests that better efforts are needed to understand and improve the perioperative care they receive.

**T**HE prevalence of adult congenital heart disease (ACHD) has increased dramatically over the past 3 decades, due to large advances in medical and surgical management and resulting improvements in the likelihood of

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◆ This article is accompanied by an Editorial View. Please see: Cannesson M, Earing M: Adult congenital heart disease patients undergoing noncardiac surgery and the role of anesthesiologists as perioperative physicians. ANESTHESIOLOGY 2013; 119:747-8.

survival to adulthood.<sup>1</sup> Compared with the general population, ACHD patients have higher rates of healthcare utilization, including hospitalization and surgery.<sup>2</sup> National consensus guidelines for noncardiac surgery<sup>3</sup> and the care of ACHD patients<sup>4</sup> as well as expert reviews<sup>5–7</sup> posit that ACHD patients are at increased risk of perioperative complications when they undergo noncardiac surgery. However, although some series have demonstrated an increased perioperative risk in children with congenital heart disease having noncardiac surgery,<sup>8</sup> few clinical outcome data are available in adults.

We used the Nationwide Inpatient Sample (NIS) to examine perioperative outcomes among ACHD patients undergoing major noncardiac surgery and assess the hypothesis that ACHD patients experience increased perioperative morbidity and mortality.

## Materials and Methods

The Stanford University Institutional Review Board granted an exemption from review because this research uses de-identified data. Administrative records were extracted from discharge datasets for the years 2002–2009 from the NIS, Healthcare Cost and Utilization Project, Agency for Healthcare Research and Quality. The NIS is the largest, publicly available, all-payer database for inpatient care in the United States. Each dataset year includes records on 7–8 millions of admissions from approximately 1,000 hospitals in 37 states, which reflect a 20% stratified sample of all U.S. nonfederal, nonrehabilitation hospitals.<sup>9</sup> It contains discharge sample weights (based on the stratified sampling methodology) to facilitate nationally representative estimates based on the sampling design. Although it contains limited administrative data on each inpatient encounter, its size and sampling frame convey the advantage of facilitating the analysis of comparatively rare clinical events at a national level.

Study procedures and reporting were carried out in accordance with the Strengthening the Reporting of Observational Studies in Epidemiology guidelines.||

### Cohort Generation and Matching

Healthcare Cost and Utilization Project–supplied Clinical Classifications Software for International Classification of Diseases, Ninth Revision, Clinical Modification was used to generate diagnostic, comorbidity, and procedural classification codes. A composite comorbidity point score was calculated to facilitate adjustment for coexisting medical conditions based on the van Walraven modification of the Elixhauser comorbidity measure,<sup>10</sup> which has been demonstrated to outperform other comorbidity schema (*e.g.*, Charlson Index) in administrative databases.<sup>11–13</sup> The van Walraven modification provides a validated method for calculating a composite comorbidity score that can be used for

matching<sup>14</sup> from the individual Elixhauser comorbidity variables (which are supplied by dedicated variables in the NIS).

Using the Healthcare Cost and Utilization Project procedure codes for diagnostic and therapeutic procedures, discharge records in which a major therapeutic intervention was performed (PCLASS 4) were retained for further examination. Records for patients under 18 yr of age and records in which a major cardiovascular procedure was performed (L2PCCS1 codes 7.1 through 7.9) were excluded from analysis. We also excluded obstetric procedures (L2PCCS1 codes 13.1 through 13.9), as this subset of procedures represents a different population and setting which has been analyzed previously using NIS data.<sup>15,16</sup> Previous analyses also suggest that a diagnosis code-based case definition for ACHD may be less reliable in obstetric patients because of the erroneous inclusion of congenital heart disease diagnosis codes in the records of healthy parturients who are carrying a fetus with a prenatally diagnosed congenital cardiac lesion.<sup>17</sup>

The remaining records comprised an adult cohort of hospital admissions during which at least one major noncardiac, nonobstetric therapeutic procedure was performed. Discharge sample weights were used to produce national estimates for the analysis of the relative frequencies of ACHD and noncardiac surgery.

ACHD was established based on the presence of International Classification of Diseases-9 codes (745.x, 746.x, and 747.1–4). With one exception, this is equivalent to the case definition strategy used in previous ACHD analyses of the NIS,<sup>15,18</sup> which have used the Clinical Classifications Software code 213 with exclusions for diagnoses codes 747.5 (absence of the umbilical artery), 747.6 (peripheral vascular anomalies), and 747.8 (cerebrovascular anomalies). The sole difference is that we also excluded code 747.9 (unspecified congenital anomaly of circulatory system), as it may not reliably identify true ACHD diagnoses.

ACHD cases within the subset of records containing a major noncardiac, nonobstetric therapeutic procedure comprised the study cohort. Matching was performed using a previously described SAS greedy caliper matching macro<sup>19</sup> (SAS Institute, Cary, NC) on the following variables: age ( $\pm 2$  yr), sex, race, year (exact match), elective or urgent or emergency procedure, van Walraven comorbidity score ( $\pm 1$  point), and primary procedure code (PRCCS1; exact match) to create a comparison cohort of up to 4:1 matched controls for each ACHD case. Records with missing data for the matching variables were excluded. We selected a 4:1 matching ratio to optimize statistical power within reasonable limits of computational efficiency.

### Statistical Analysis

Demographic, preoperative, and perioperative outcome variables were compared between the two cohorts. The primary outcome measure was all-cause in-hospital mortality, as recorded in the NIS dataset (variable DIED). The secondary outcome measure was a composite endpoint

|| STROBE Statement. Available at: <http://www.strobe-statement.org/index.php?id=available-checklists>. Accessed March 6, 2013.

of major nonfatal morbidity constructed from multilevel Clinical Classifications Software diagnosis codes including pneumonia (8.1.1), acute respiratory failure (8.6.1), acute renal failure (10.1.2), deep venous thrombosis or pulmonary embolism (7.5.1), stroke (four diagnosis codes: 6.3.1 ["hemiplegia"], 6.6 ["coma, stupor, or brain damage"], 7.3.1.1 [intracranial hemorrhage], 7.3.1.2 [occlusion of cerebral arteries]), myocardial infarction (7.2.3), and cardiac arrest (7.2.10). For outcomes that could overlap with chronic comorbid conditions, no acute complication was recorded for patients with the corresponding chronic comorbid measure present (variable CM\_NEURO or CM\_PARA for stroke; variable CM\_RENLFAIL for acute renal failure). This modification sacrificed the undercounting of some complications (*e.g.*, acute or chronic renal failure) in order to reduce the likelihood of erroneously classifying a chronic condition as an acute perioperative complication. Length of stay (variable length of stay) and total hospital charges (variable TOTCHG) also were compared between groups.

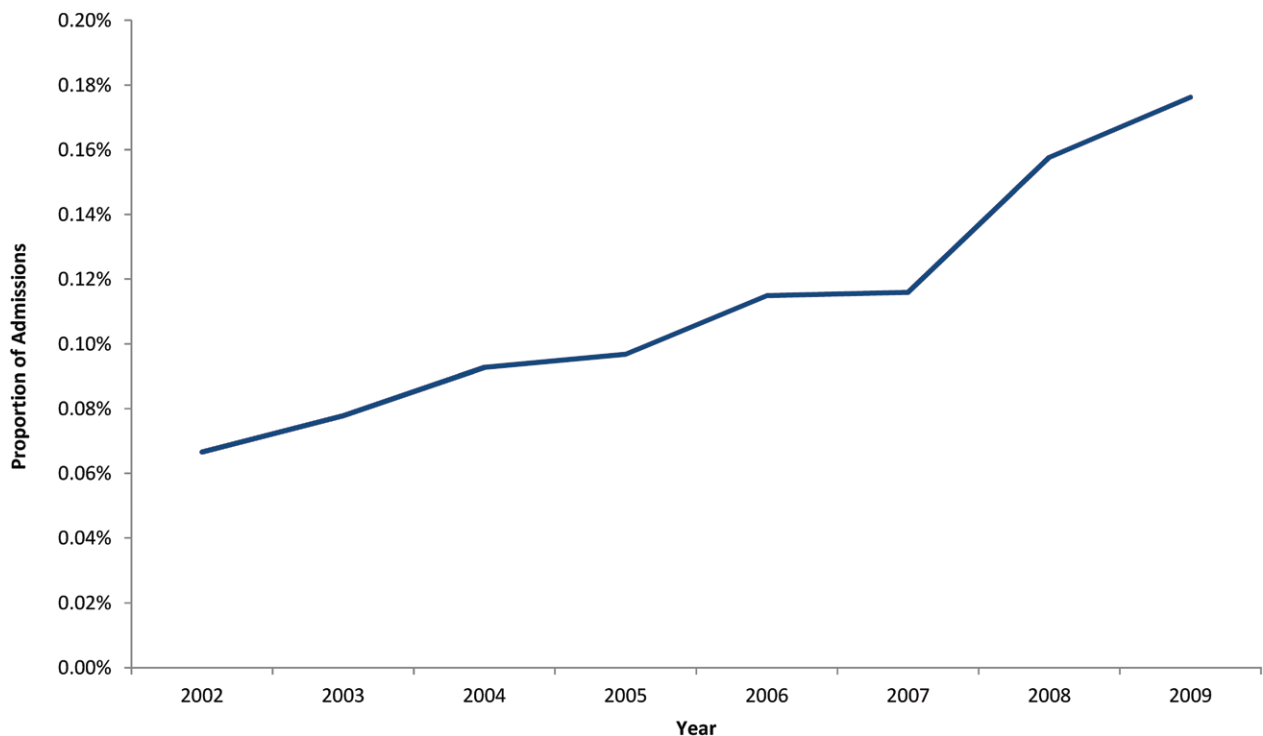
A conditional multivariate logistic regression model (using matching pairs as strata for conditional logistic regression) was constructed to further evaluate the comparative effect of an ACHD diagnosis on the primary outcome (in-hospital mortality) while adjusting for demographic and procedural differences. All matching variables except for exact procedure code (age, sex, race, year, elective or non-elective admission, and van Walraven comorbidity score) were included in the regression model to account for any

residual differences after matching. In addition, payer type, surgical service, hospital size, and teaching hospital status were added to the model to adjust for any confounding by these parameters. Two- and three-way interactions between predictive variables were included for initial evaluation but retained in the final model only if statistically significant. The area under the receiver operating characteristic curve, deviance, and Pearson goodness-of-fit statistics were calculated for calibration of the model.

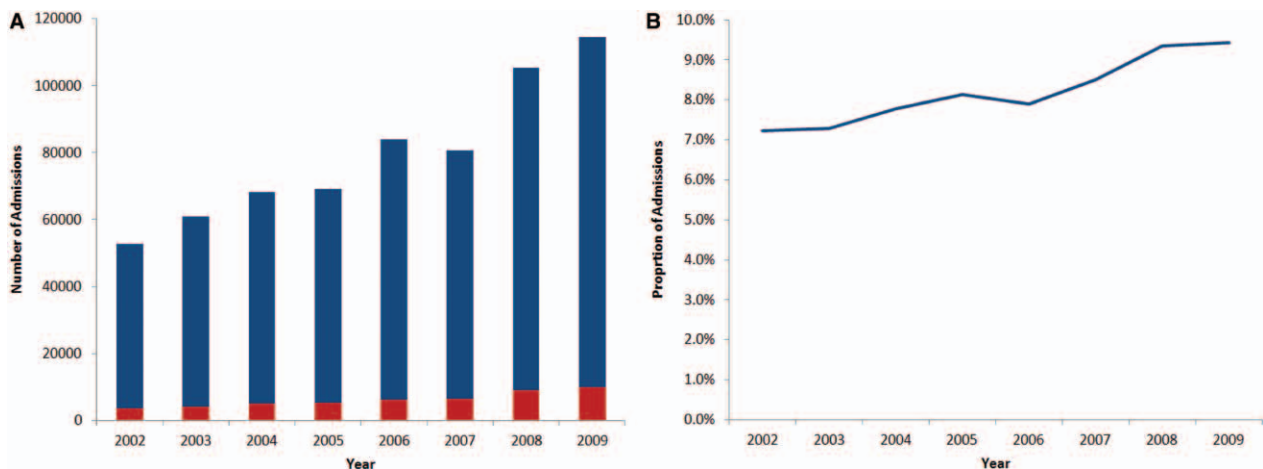
Continuous variables were compared using the Wilcoxon test. Discrete variables were compared using Fisher exact test or Pearson chi-square test, as appropriate. For outcome variables, odds ratios with 95% CIs also were calculated. Trends over time were examined using a Mann–Kendall test for trend (a nonparametric test to determine the presence and direction of a trend over time).<sup>20</sup> A predetermined  $\alpha$  of 0.05 was used as the threshold of statistical significance for the primary outcome and the composite secondary outcome. For the purposes of evaluating the six individual outcome contributors to the composite endpoint, a Bonferroni-adjusted significance level of 0.0083 was used to account for the increased possibility of type-I error. Analyses were performed using SAS (SAS 9.3; SAS Institute).

## Results

ACHD patients represent 0.11% of admissions involving noncardiac surgery, which is consistent with previous estimates that 0.1% of all NIS admissions represent ACHD patients.<sup>18</sup>



**Fig. 1.** Adult congenital heart disease admission records with noncardiac surgery as a percentage of all admissions with noncardiac surgery, by year (2002–2009). Discharge sample weights were used to produce national estimates. Trend is increasing over time ( $P$  value for trend is  $<0.001$ ).



**Fig. 2.** Discharge sample-weighted national estimates for adult congenital heart disease (ACHD) admission records by year (2002–2009), partitioned by those with noncardiac surgery (A, red bars) and those without noncardiac surgery (A, blue bars). (B) Shows ACHD admissions with noncardiac surgery as a percentage of all ACHD admissions. Trend is increasing over time ( $P$  value for trend is  $<0.001$ ).

This proportion increased steadily over the study period, from 0.07% in 2002 to 0.18% in 2009 (fig. 1), which represents a significantly increasing trend over time ( $P < 0.001$ ).

Noncardiac surgery admissions represent 8.4% of admissions for ACHD patients. This fraction also increased steadily over the study period from 7.2% in 2002 to 9.4% in 2009 (fig. 2), which represents a significantly increasing trend over time ( $P < 0.001$ ). This trend was specific to the ACHD cohort, as there was no similar increase over time in the fraction of all NIS records with noncardiac surgery ( $P = 0.42$ ).

A study cohort of 10,004 admissions for ACHD patients undergoing noncardiac surgery was identified. The matched comparison cohort consisted of 37,581 admissions for non-ACHD patients undergoing noncardiac surgery. Of admissions in the ACHD cohort, all were successfully matched within the prespecified parameters to controls, with 8,102 (81%) matched to the maximum of four controls, 1,452 (15%) to three controls, 368 (4%) to two controls, and 81 (1%) to one control. Table 1 contains demographic and baseline characteristics of the ACHD and comparison cohorts.

Table 2 shows outcomes for the ACHD cohort and comparison cohort. Mortality was greater in the ACHD cohort (odds ratio, 1.13;  $P = 0.031$ ; 95% CI, 1.01–1.27), as was the composite endpoint for perioperative morbidity (odds ratio, 1.44;  $P < 0.001$ ; 95% CI, 1.36–1.52). Each contributor to the composite endpoint (acute renal failure, pneumonia, respiratory failure, deep venous thrombosis or pulmonary embolism, stroke, myocardial infarction, and cardiac arrest) was significantly more common in the ACHD cohort ( $P < 0.001$  for all, which is below the Bonferroni-adjusted significance level of 0.0083). In addition, length of stay and total hospital charges were greater in the ACHD cohort. Table 3 shows mortality rate by congenital cardiac lesion within the ACHD cohort. Complex lesions were associated with the highest mortality rate (7.3%).

Conditional multivariate analysis revealed independent predictors of mortality to include age, female sex, nonwhite race, nonelective surgery status, surgical service, payer, large hospital size, teaching hospital, van Walraven comorbidity score, and presence of ACHD (table 4). No two- or three-way interactions were retained in the final model. The area under the receiver operating characteristic curve was 0.818, and the model was well-calibrated ( $P = 0.99$  for both deviance and Pearson goodness-of-fit statistics).

## Discussion

The principal finding of this study is that ACHD patients undergoing major noncardiac surgery experienced greater in-hospital mortality compared with a well-matched comparison cohort. ACHD was an independent predictor of increased perioperative mortality in both univariate and multivariate analyses. Absolute mortality exceeded 4% in this group, which is consistent with the suggestion that these patients are especially vulnerable in the perioperative period. For comparison purposes, a recent series of unselected surgical patients in a different setting (498 hospitals in 28 European countries) documented a similar in-hospital mortality rate (4%).<sup>21</sup> By contrast, a review of the American College of Surgeons National Surgical Quality Improvement Project database (which captures a broad American surgical population with the exclusion of minor procedures) found an overall mortality rate of 1.76% and an in-hospital mortality rate of 1.35%.<sup>22</sup> Perioperative morbidity, length of stay, and hospital charges were also greater in the ACHD cohort.

The importance of these perioperative outcomes is heightened by the additional observations that over the study period, ACHD patients comprised an increasing fraction of all noncardiac surgical admissions, and noncardiac surgery accounted for an increasing percentage of ACHD admissions. These trends suggest that the impact of greater



**Table 1.** Demographic and Baseline Characteristics of ACHD and Comparison Cohorts

	ACHD Cohort	Comparison Cohort	
	n = 10,004	n = 37,581	
	n (%)	n (%)	P Value
Age (yr, mean $\pm$ SE)	57.6 ( $\pm$ 0.3)	58.1 ( $\pm$ 0.2)	0.0192
Female	5,520 (55.2)	21,005 (56.0)	0.18
Race			0.06
White	6,140 (81.8)	23,374 (82.7)	
Black	534 (7.1)	1,885 (6.7)	
Hispanic	469 (6.3)	1,547 (5.5)	
Asian	136 (1.8)	559 (2.0)	
Native American	39 (0.5)	150 (0.5)	
Others	185 (2.5)	754 (2.7)	
Nonelective status	4,717 (47.2)	17,659 (46.8)	0.67
Payer			<0.001
Medicare	4,313 (43.2)	15,900 (42.4)	
Medicaid	781 (7.8)	2,988 (8.0)	
Private	4,296 (43.0)	15,527 (41.4)	
Self-Pay	254 (2.5)	1,384 (3.7)	
Others	342 (3.4)	1,728 (4.6)	
Operation by surgical service			<0.001
ENT	225 (2.3)	969 (2.6)	
General surgery	3,514 (35.1)	13,329 (35.5)	
Gynecologic	770 (7.7)	3,078 (7.5)	
Neurosurgery	1,201 (12.0)	4,881 (13.0)	
Orthopedic	2,991 (29.9)	12,061 (29.2)	
Thoracic	1,596 (13.9)	4,563 (12.1)	
Urologic	597 (6.0)	2,445 (6.5)	
Hospital size			<0.001
Small	6,812 (68.1)	24,916 (66.3)	
Medium	2,153 (21.5)	8,553 (22.8)	
Large	1,038 (10.4)	4,112 (10.9)	
Hospital teaching status			<0.001
Nonteaching	5,901 (58.9)	19,493 (51.9)	
Teaching	4,103 (41.1)	18,088 (48.1)	
van Walraven score (mean $\pm$ SE)	3.4 $\pm$ 0.8	3.1 ( $\pm$ 0.7)	0.68

Values are reported as number (percentage) unless otherwise denoted as mean  $\pm$  SE.

ACHD = adult congenital heart disease; ENT = ear, nose, throat; SE = standard error.

perioperative morbidity and mortality on this population may continue to increase as more ACHD patients undergo noncardiac surgery. As the majority of patients in this study were cared for in nonteaching hospitals, this is an issue that will confront perioperative providers in a broad range of settings, not just tertiary care centers.

The major limitations of this analysis are the well-established limitations of a retrospective administrative database analysis, principally that of classification error.<sup>23,24</sup> Misclassification may be a greater source of error in the current setting than in other analyses that have used the NIS. When ACHD patients undergo noncardiac operations, noncardiac surgeons and other inpatient providers (*e.g.*, hospitalists) may not be accurate or precise in their identification of the patient's congenital cardiac lesion. Factors contributing to

this inaccuracy include the time elapsed from initial repair, lack of familiarity with a rare condition, and an impression on the part of the patient and/or physician that their previous congenital heart lesion has been "fixed" and therefore no longer represents an active diagnosis. Care patterns may also contribute; the patient may not currently be under the care of a cardiologist, and current or past cardiology and cardiovascular surgical care may have occurred at a different institution, such that records are incomplete or unavailable to current providers. Overall, administrative data likely underestimate the frequency of comorbid congenital heart disease in adult patients.<sup>18</sup> Even when accurate codes for coexisting ACHD are present, this information does not reveal each patient's specific underlying lesion, number or nature of previous surgical repairs or palliative procedures, or

**Table 2.** Outcomes

Outcome	ACHD Cohort	Comparison Cohort		
	n = 10,004	n = 37,581	P Value	OR (95% CI)
	n (%)	n (%)		
Death	407 (4.1)	1,355 (3.6)	0.031	1.13 (1.01–1.27)
LOS (median [IQR])	4.8 (2.4–10.4)	2.9 (1.5–5.6)	<0.001	
Total charges (median [IQR])	\$42,171 (\$22,918–\$93,847)	\$26,982 (\$15,814–\$46,784)	<0.001	
ARF	620 (6.2)	1,826 (4.9)	<0.001	1.29 (1.18–1.42)
Pneumonia	942 (9.4)	2,998 (8.0)	<0.001	1.20 (1.11–1.29)
Respiratory failure	916 (9.2)	2,933 (7.8)	<0.001	1.19 (1.10–1.29)
DVT/PE	405 (4.1)	773 (2.1)	<0.001	2.01 (1.78–2.27)
Stroke	607 (6.1)	1,168 (3.1)	<0.001	2.01 (1.82–2.23)
MI/cardiac arrest	431 (4.3)	1,307 (3.5)	<0.001	1.25 (1.12–1.40)
Composite	2,145 (21.4)	6,003 (16.0)	<0.001	1.44 (1.36–1.52)

Values are reported as number (percentage) unless otherwise denoted as median (IQR). Composite = ARF, pneumonia, respiratory failure, DVT/PE, stroke, MI, and cardiac arrest.

ACHD = adult congenital heart disease; ARF = acute renal failure; DVT = deep venous thrombosis; IQR = interquartile range; LOS = length of stay; MI = myocardial infarction; OR = odds ratio; PE = pulmonary embolus.

detailed measures of hemodynamic status at the time of the noncardiac operation.

The range of variables used for matching was selected to create a comparison cohort that would represent a face value “peer group” for the study cohort, but matching is likely an imperfect strategy for risk adjustment. For instance, previous studies have demonstrated that ACHD patients may have substantial impairments in functional status and renal function that are not clinically apparent<sup>25,26</sup> and that patients themselves may not appreciate.<sup>27</sup>

**Table 3.** Mortality by Lesion Type within the ACHD Cohort

	n	Died
		n (%)
Atrial septal defect	4,068	155 (3.8)
Congenital aortic stenosis/aortic insufficiency	1,789	53 (3.0)
Congenital mitral stenosis/regurgitation	85	3 (3.5)
Congenital conduction defect*	469	10 (2.1)
Congenital coronary anomaly	248	9 (3.6)
Pulmonic stenosis	239	13 (5.4)
Tetralogy of Fallot	121	7 (5.8)
Ventricular septal defect	831	52 (6.3)
Ebstein anomaly	65	4 (6.2)
Others	1,745	76 (4.4)
Combined complex†	344	25 (7.3)

Values are reported as number (percentage).

\* Congenital heart block, Brugada syndrome. † Tricuspid atresia or stenosis, pulmonary atresia, Common ventricle, hypoplastic left heart syndrome, cor biloculare, and truncus arteriosus.

ACHD = adult congenital heart disease.

A 4:1 matching ratio and caliper matching parameters (age  $\pm 2$  yr, van Walraven score  $\pm 1$  point) were selected to balance goals of optimizing match quality while avoiding unmatched cases. Although it remains a limitation of a multiple-matching algorithm that cases with less than the maximum number of matched controls may exhibit different characteristics than cases fully matched, this seems unlikely to have dramatically changed these results, given the small number of cases with less than a maximum of four matched controls.

Outcome information is similarly limited in an administrative database. Nonfatal complications are based on diagnosis codes, which do not include information on outcome severity. Moreover, nonfatal complications are probably undercounted in this analysis, both because of limited reliability of diagnostic codes for perioperative complications and because of our exclusion of patients with an existing comorbidity indicator for the same type of condition as the complication being measured. For instance, patients who had both preexisting renal failure as indicated by the CM\_RENLFAIL data variable, and a diagnosis code for acute renal failure were not considered to have acute renal failure as a perioperative complication, even though these patients may be among those at highest risk of this type of complication (e.g., acute or chronic renal failure). For these reasons, we used mortality as the primary outcome, as we felt it represented a more reliable endpoint.

We believe the paucity of outcome data in the ACHD population justifies some tolerance of these limitations. Although pediatric patients with congenital heart disease comprise a closely followed group that receives care predominantly in a relatively small number of highly specialized centers, ACHD patients often receive care in community settings without subspecialist follow-up. Only

**Table 4.** Results of Conditional Multivariate Logistic Regression Modeling Mortality

	P Value	OR/Point Estimate	95% CI
Age	<0.001	1.021	
Sex	0.001		
Male		[1]	
Female		1.16	1.09–1.23
Race			
White	[1]	[1]	
Black	<0.001	1.38	1.24–1.54
Hispanic	0.026	1.20	1.05–1.37
Asian/Pacific Islander	0.041	1.29	1.05–1.60
Native American	<0.001	2.51	1.80–3.48
Others	0.08	1.15	0.94–1.40
Surgery status	<0.001		
Elective		[1]	
Nonelective		2.13	1.99–2.28
Payer			
Medicare	[1]	[1]	
Medicaid	<0.001	1.35	1.20–1.52
Private including HMO	<0.001	0.89	0.82–0.97
Self-pay	<0.001	1.38	1.17–1.62
Others	0.13	0.72	0.60–1.04
Surgery type			
ENT	0.32	1.04	0.70–1.58
General surgery	[1]	[1]	
Gynecologic	<0.001	0.40	0.26–0.62
Neurosurgery	<0.001	5.29	4.65–6.02
Orthopedic	<0.001	3.68	3.33–4.07
Thoracic (noncardiac)	<0.001	8.80	7.90–9.80
Urologic	0.12	1.05	0.81–1.35
Hospital size			
Small	[1]	[1]	
Medium	0.49	1.11	0.94–1.25
Large	<0.001	1.23	1.10–1.38
Hospital teaching status			
Nonteaching	[1]	[1]	
Teaching	<0.001	1.46	1.37–1.55
van Walraven score	<0.001	1.082	
ACHD	<0.001		
No		[1]	
Yes		1.29	1.07–1.43

ACHD = adult congenital heart disease; ENT = ear, nose, throat; HMO = health maintenance organization; OR = odds ratio.

a small fraction of the noncardiac operations that ACHD patients undergo later in life occur in the same institution as the original cardiac operation. For instance, in a recent retrospective analysis, investigators at the Mayo Clinic reviewed noncardiac surgery in all patients who previously had undergone a Fontan operation ( $n = 1,133$ ), and identified only 39 general anesthetics in 31 patients.<sup>28</sup> In a similar analysis at the University of Michigan, of 73 patients with hypoplastic left heart syndrome who underwent a

subsequent noncardiac operation in their health care system, none were aged more than 18 yr.<sup>29</sup> The lack of adult patients was not due to a pediatric-only database; if adult patients existed, they should have been captured by the database that was used (Robert Christensen, M.D., Clinical Lecturer, Department of Anesthesiology, University of Michigan, written communication, December 3, 2012.) In a previous era, this might have reflected a lack of significant numbers of patients surviving to adulthood, but we know this not to be the case.<sup>7</sup> Even if, young survivors of hypoplastic left heart syndrome only infrequently require noncardiac operations.

These patterns make it unlikely that single-institution case series will provide adequate outcome information. In the absence of a national or multicenter registry that follows large numbers of these patients after they “graduate” from receiving centralized, coordinated care at pediatric cardiac centers, administrative data are the best currently available tool to assess the perioperative outcomes and risks in this population.

Our analysis suggests vulnerability of the growing ACHD population to increase morbidity and mortality during the perioperative period, and highlights gaps in current knowledge. Large prospective databases are needed to gain more detailed information about risk factors within the ACHD population, and to determine which aspects of their perioperative risk are modifiable in order to improve patient outcome.

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