Assessing surgical risk for adults with congenital heart disease: Are pediatric scoring systems appropriate?

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Background: Patients with congenital heart disease are frequently surviving into adulthood, and many of them will require surgery. Currently, there is no validated risk scoring system for adult congenital heart surgery, and predicting outcomes in these patients is challenging. Our objective was to determine if commonly used pediatric congenital heart disease surgery risk scores are also applicable to adults.

Methods: Four hundred fifty-eight adult (age ≥ 18 years) operations involving cardiac surgery for congenital heart disease between 2000 and 2010 at a single institution were studied retrospectively. The pediatric scores evaluated were the Risk Adjustment for Congenital Heart Surgery (RACHS-1) score, the Aristotle Basic Score, and the Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery (STAT) Congenital Heart Surgery Mortality score. Receiver operating characteristic (ROC) curves were generated to assess the ability of the scoring systems to predict mortality, major adverse events (stroke, renal failure, prolonged ventilation, prolonged coma, deep sternal infection, reoperation, and operative mortality), and prolonged length of stay (>7 days).

Results: Of 458 operations, there were 16 (3%) deaths, 94 (21%) major adverse events, and 90 (20%) prolonged lengths of stay. Four hundred thirty (94%) of the operations were included in all 3 scoring systems and the ROC analysis. For mortality, areas under the ROC curve were 0.91, 0.91, and 0.65 for the Aristotle, STAT, and RACHS-1 scores, respectively. For major adverse event, areas under the ROC curves were 0.81, 0.76, and 0.61 for the Aristotle, STAT, and RACHS-1 scores, respectively. For prolonged length of stay, areas under the ROC curve were 0.82, 0.76, and 0.61 for the Aristotle, STAT, and RACHS-1 scores, respectively.

Conclusions: Pediatric risk scoring systems such as Aristotle, STAT, and RACHS-1 offer prognostic value in adults undergoing congenital heart surgery. The scores are predictive of mortality, major adverse events, and prolonged lengths of stay. The STAT and Aristotle systems fared best. (J Thorac Cardiovasc Surg 2014;147:666-71)

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Patients with congenital heart disease are frequently surviving into adulthood. In 2000, the number of adult patients with congenital heart disease in the United States was estimated to be between 650,000 and 1.3 million.¹⁻³ More importantly, the prevalence of severe congenital heart disease has increased by 85% in adults compared with 22% in children, consistent with the notion that the greatest survival trend has occurred in patients with more

severe disease.¹⁻⁴ As adults, many of these patients with complex conditions will require surgery.

Currently, there is no validated risk scoring system for adult congenital heart surgery, and predicting outcomes in these patients is challenging. In children, the Risk Adjustment for Congenital Heart Surgery (RACHS-1) scoring system, the Aristotle scoring system, and the Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery (STAT) Congenital Heart Surgery Mortality scoring system have been used to stratify risk and predict/compare outcomes.

The purpose of this study was to determine if commonly used pediatric congenital heart disease surgery risk scores were also applicable to adults. The prognostic value of the pediatric scoring systems with respect to mortality, major adverse events, and length of stay in adult was evaluated in patients with congenital heart disease.

METHODS

A retrospective cohort study of 458 consecutive operations on adult patients (age ≥ 18 years) with a previous history of congenital heart disease, who underwent cardiac surgery at a single, large, academic center between January 1, 2000, and December 31, 2010, was performed. For all operations, the surgical procedure was attributed to congenital cardiac pathology. Institutional Review Board approval was obtained for this retrospective study, and individual patient consent was waived.

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Abbreviations and Acronyms

EACTS	= European Association for Cardiothoracic Surgery
RACHS-1	= Risk Adjustment for Congenital Heart Surgery
ROC	= receiver operating characteristic
STAT	= Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery
STS	= Society of Thoracic Surgeons

Scoring Systems

The RACHS-1 scoring system, the Aristotle basic scoring system, and the STAT Congenital Heart Surgery Mortality scoring system were evaluated. Using tables from the landmark articles on the RACHS-1, Aristotle, and STAT scoring systems, we assigned each operation a score within each system.⁵⁻⁷ For the RACHS-1 system, the scale ranged from 1 to 6.⁵ For the Aristotle basic system, the scale ranged from 1.5 to 15, and a corresponding basic complexity level between 1 and 4 was assigned (level 1, 1.5-5.6; level 2, 6-7.8; level 3, 8-9.5; level 4, 10-15).⁶ For the STAT score, the scale ranged from 0.1 to 5.0, and a corresponding mortality category level between 1 and 5 was assigned (level 1, 0.1-0.3; level 2, 0.4-0.7; level 3, 0.8-1.2; level 4, 1.3-2.6; level 5, 2.9-5.0).⁷ In each scoring system, a higher score indicates a higher risk of mortality. For patients undergoing multiple procedures, the procedure with the highest level was scored.

Outcomes

Operative mortality, presence of a major adverse event, and length of hospital stay greater than 7 days were chosen as the 3 primary outcomes. Operative mortality was defined as a death occurring during the surgical hospital stay or within 30 days of surgery. Because of the infrequency of individual complications, a composite adverse event outcome was used. The major adverse event composite outcome variable was positive if the patient had any of the following major complications: stroke, renal failure, prolonged ventilation, deep sternal infection, reoperation, and operative mortality. All patients were followed until the time of discharge to determine the occurrence of a primary outcome.

The Society of Thoracic Surgeons (STS) database was used to define the list of major adverse events.⁸ Stroke was defined as a neurologic deficit caused by a disturbance in cerebral perfusion that did not resolve within 24 hours. Renal failure was defined by a serum creatinine level greater than 2.0 and twice the preoperative level, or by a new requirement for dialysis. Prolonged ventilation was defined as any requirement for mechanical ventilation more than 24 hours postoperatively. Deep sternal infection was defined as a sternal infection within 30 days of surgery, requiring all of the following conditions: (1) wound or mediastinal exploration with excision of tissue, (2) positive wound culture, and (3) treatment with antibiotics. Reoperation was defined as surgery performed for any reason within 30 days of surgery. Operative mortality included (1) all deaths occurring during the hospital stay when the operation was performed and (2) those deaths occurring after discharge from the hospital, but within 30 days of the procedure.

Analysis/Statistics

The occurrences of mortality, major adverse events, and prolonged lengths of stay were summarized for the entire cohort. For those patient operations with a score from each scoring system, logistic regression was used to generate receiver operating characteristic (ROC) curves to assess the ability of each scoring system to predict each of the 3 primary

TABLE 1. Frequency of operations performed (in descending order)

Procedure name	n
Pulmonary valve repair	142
Tricuspid valve repair	29
Conduit reoperation	28
Atrial septal defect repair, patch	25
Aortic root replacement, mechanical or bioprosthetic	24
Valvuloplasty, tricuspid	19
Atrial septal defect repair, patch + partial anomalous pulmonary venous connection repair	16
Fontan revision or conversion (redo Fontan procedure)	16
Mitral valve repair	15
Transplantation, heart	14
Valvuloplasty, mitral	14
Aortic valve replacement, mechanical or bioprosthetic	11
Pulmonary artery, reconstruction (plasty), branch, central (within the hilar bifurcation)	10
Aortic valve closure (aortic valve septal defect) repair, partial (incomplete)	8
Double chamber right ventricle repair	8
Ventricular septal defect repair, patch	8
Coronary artery bypass	7
Ross procedure	7
Coarctation repair, end-end	5
Valvuloplasty, aortic	5
Aortic stenosis, supraaortic, repair	4
Cardiac tumor resection	4
Anomalous origin of coronary artery repair	3
Aortic aneurysm repair	3
Arrhythmia surgery, atrial, surgical ablation	3
Coarctation repair, interposition graft	3
Partial anomalous pulmonary venous connection, scimitar, repair	3
Anomalous systemic venous connection repair	2
Aortic arch repair	2
Aortic stenosis, subvalvular, repair	2
Bidirectional cavopulmonary anastomosis (bidirectional Glenn procedure)	2
Ventricular assist device	2
Aortic arch repair + ventricular septal defect repair	1
Aortic dissection repair	1
Aortic root replacement, homograft	1
Aortic root replacement, valve sparing	1
Atrial septal defect partial closure	1
Konno procedure	1
Left atrial appendage excision	1
Patent ductal arteriosus closure, surgical	1
Pericardiectomy	1
Right ventricular outflow tract procedure	1
Septal myectomy	1
Shunt, systemic to pulmonary artery, modified Blalock-Taussig shunt	1
Sinus of Valsalva, aneurysm repair	1
Transplantation, lung(s)	1
Total	458

outcomes. For the Aristotle and STAT systems, the raw scores were used. Higher area under the ROC curve indicated better discriminatory

FIGURE 1. RACHS-1 scoring system distribution. *RACHS*, Risk Adjustment for Congenital Heart Surgery.

performance of the scoring system. ROC curves were compared using a test of joint equality of ROC curve areas.⁹ All statistics were performed using SAS 9.3 (SAS Institute, Inc, Cary, NC).

RESULTS

Of 458 patient operations, there were 16 (3%) deaths, 94 (21%) major adverse events, and 90 (20%) prolonged lengths of stay. A breakdown of the major adverse events showed the following: stroke (6, 1.3%), renal failure (36, 7.9%), prolonged ventilation (63, 13.8%), deep sternal infection (0), reoperation (58, 12.7%), and operative mortality (16, 3.5%).

A complete list of the operations and associated frequencies is shown in Table 1. Four hundred thirty (94%) operations were included in all 3 scoring systems and the ROC analysis. The scoring distributions among the RACHS-1, Aristotle, and STAT systems are shown in Figures 1 to 3. Some patients could not be scored within the RACHS-1 (24, 5.2%), Aristotle (2, 0.4%), and STAT (5, 1.1%) scoring systems.

Mortality

For mortality, areas under the ROC curve were 0.91, 0.91, and 0.65 for the Aristotle, STAT, and RACHS-1 scores,

respectively (Figure 4, A). Compared with the Aristotle score, there was no difference in the STAT score ($P = .97$), whereas the RACHS-1 score performed significantly worse ($P < .0001$).

Major Adverse Events

For major adverse events, the areas under the ROC curve were 0.81, 0.76, and 0.61 for the Aristotle, STAT, and RACHS-1 scores, respectively (Figure 4, B). The Aristotle score performed significantly better than either the STAT score ($P = .03$) or the RACHS-1 score ($P < .0001$).

Length of Stay

For prolonged length of stay, the areas under the ROC curve were 0.82, 0.76, and 0.61 for the Aristotle, STAT, and RACHS-1 scores, respectively (Figure 4, C). The Aristotle score performed significantly better than either the STAT score ($P = .01$) or the RACHS-1 score ($P < .0001$).

Comparison

For each scoring system, a comparison was also performed between mortality rates in this adult cohort and a similar pediatric cohort at our institution (Table 2). Although the trend is increasing mortality with increasing scores in both populations, the absolute mortality is generally higher in the adult cohort than the pediatric cohort throughout.

DISCUSSION

In this retrospective cohort study, the first of its kind to evaluate potential scoring systems in congenital heart surgery in an exclusively adult population, we found that scoring systems used in the pediatric population may have prognostic value in adults. Specifically, the Aristotle score fared particularly well in predicting outcomes pertaining to mortality, major adverse events, and length of stay in adults undergoing congenital heart surgery. The STAT score similarly performed well, although not as well as the

FIGURE 2. Aristotle scoring system distribution.

FIGURE 3. STAT scoring system distribution. *STAT*, Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery.

Aristotle score in the domains of adverse events and length of stay. The RACHS-1 score, although of some prognostic value, did not function as well as the other scoring systems.

Current Tools in Pediatric Congenital Heart Surgery

Over the last 2 decades, pediatric cardiac surgeons have recognized the need for reliable outcome reporting, as shown by the creation of several congenital heart surgery databases and several risk stratification scoring systems. Nonetheless, it remains challenging when the individual diagnoses and surgical procedures are numerous and the number of individual patients is small. Despite this, several tools have been developed to measure outcomes, compare performance, and provide a platform for quality improvement in pediatric congenital heart surgery.

RACHS-1 was first described in 2001. A panel of pediatric cardiologists and pediatric cardiac surgeons used

clinical judgment to place surgical procedures into 6 risk categories. The aim was to develop a consensus-based method of risk adjustment for in-hospital mortality among children younger than 18 years after surgery for congenital heart disease. In the initial report, mortality rates were 0.4% in category 1, 3.8% in category 2, 8.5% in category 3, 19.4% in category 4, and 47.7% in category 6. There were too few cases in category 5 to estimate mortality rates.⁵

The Aristotle scoring system was described in 2004. A panel of expert surgeons representing the European Association for Cardiothoracic Surgery (EACTS), STS, European Congenital Heart Surgeons Association, and Congenital Heart Surgeons Society, assigned each operation 2 scores. The first score was the Basic Score, which adjusted only for the complexity of the procedure. The second score was the Comprehensive Score, which took into account specific procedure-dependent and

FIGURE 4. ROC curves. A, Mortality ROC curves for comparison. B, Major adverse event ROC curves for comparison. C, Length of stay (*LOS*) ROC curves for comparison. *ROC*, Receiver operator characteristic; *MAE*, major adverse event; *STAT*, Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery; *RACHS*, Risk Adjustment for Congenital Heart Surgery.

TABLE 2. Comparison of adult and pediatric mortality

Scoring system	Adult cohort mortality (%)	Pediatric cohort mortality (%)
RACHS-1		
1	0	0
2	2.2	0
3	4.6	1.4
4	0	3.8
5-6	na	13.6
STAT		
1	0.3	0
2	5	0.6
3	6.6	1.7
4	9.7	4.2
5	na	12.5
Aristotle		
1	0	0
2	3.3	1.4
3	6.2	2
4	0	4.2

na, Not applicable; STAT, Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery; RACHS, Risk Adjustment for Congenital Heart Surgery.

procedure-independent patient characteristics. The Basic Score ranged from 1.5 to 15 and 4 levels of difficulty were defined.⁶

In 2010, a new scoring system (STS-EACTS, or STAT) was developed to create an objective index, as opposed to the RACHS-1 and Aristotle systems, which were predominantly subjectively derived. Mortality risk was estimated using data from operations entered into the EACTS Congenital Heart Surgery Database and the STS Congenital Heart Surgery Database. The objectives of this new system were to (1) estimate procedure-specific relative risks of in-hospital mortality, (2) convert these procedure-specific mortality estimates into a scale ranging from 0.1 to 5.0 (STAT mortality score), and (3) group procedures with similar estimated mortality into a small number of relatively homogeneous categories (STAT mortality category).⁷

Although these scoring systems were designed primarily to predict mortality, they also have prognostic value for

other outcomes. Studies have shown correlation with higher-risk procedures and postoperative infection, prolonged mechanical ventilation, feeding difficulty, acute renal failure requiring dialysis, and readmissions after pediatric congenital heart surgery.¹⁰⁻¹⁴

Current Tools in Adult Congenital Heart Surgery

The STS risk calculator and the RACHS-1, Aristotle, and STAT scoring systems have been instrumental in outcome reporting, performance comparison, and quality improvement initiatives in adult-acquired heart surgery and pediatric congenital heart surgery, respectively. Unfortunately, the available tools for evaluating risk and predicting outcome in adult congenital heart surgery are limited. The current version of the STS risk calculator does not allow for entry of most congenital operations (only coronary bypass operations, aortic or mitral valve operations, or some combination thereof), and it is unclear whether the established pediatric congenital scoring systems apply to adult patients with congenital heart disease. As a result, predicting outcomes in these patients is challenging.

The distribution of operations is not the same in the 2 patient populations. Using the currently available risk factor scoring systems, the distributions in our pediatric congenital cardiac operations and our adult congenital cardiac operations are different; the distribution is skewed toward the lower-risk procedures in adults (Figure 5). In our adult congenital population, few patients were in the higher-risk category (ie, Norwood operation, interrupted aortic arch, truncus arteriosus), whereas many patients were in the lower- and middle-risk categories (ie, pulmonary valve replacement, aortic valve replacement). In addition to the distribution of operations, the patient-specific characteristics and comorbidities that may influence outcomes are also different between the adult and pediatric patient populations. For example, based on the Aristotle Comprehensive Scoring system, factors that may influence outcome in the pediatric population include low birth weight, prematurity, genetic and chromosomal

FIGURE 5. Adult and pediatric case distribution within each scoring system. RACH, Risk Adjustment for Congenital Heart Surgery; STAT, Society of Thoracic Surgery–European Association for Cardio-Thoracic Surgery.

issues, and noncardiac congenital anomalies.⁶ Based on the STS risk calculator for acquired adult cardiac surgery, factors that may influence outcome in the adult population include diabetes mellitus, hypertension, peripheral vascular disease, and New York Heart Association class.⁸

With these differences in mind, our objective was to determine if commonly used pediatric congenital heart disease surgery risk scoring systems were also applicable to adult patients with congenital heart disease. From our study, it seems that despite the differences in the pediatric and adult populations with congenital heart disease, pediatric risk scoring systems can predict mortality in adult patients undergoing congenital surgery. Just as in the pediatric population, they also seem to have prognostic value with respect to other outcomes, such as major adverse events and prolonged length of stay. As shown, the Aristotle and STAT scoring systems are better than the RACHS-1 system in predicting these adverse outcomes. The Aristotle and STAT scoring systems likely fared better because of the wider distribution of adult operations for congenital heart conditions across the scoring spectrum compared with the RACHS-1 system.

Limitations

This study is limited by its observational nature and the inherent limitations of a retrospective database study. Also, these scoring systems, particularly the Aristotle system, take into account additional procedure- and patient-specific factors that were not included in the analysis.

CONCLUSIONS

Although there is currently no validated scoring system for congenital heart surgery in adults, the risk stratification systems for similar surgeries in children do have some prognostic value; the Aristotle and STAT scoring systems

fared the best. Future efforts are warranted to further optimize these or similar scoring systems to obtain even better prognostic value in the adult population with congenital heart disease.

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