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# Risk evaluation in adult congenital heart surgery: analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database risk models on data from the European Congenital Heart Surgeons Association Congenital Database

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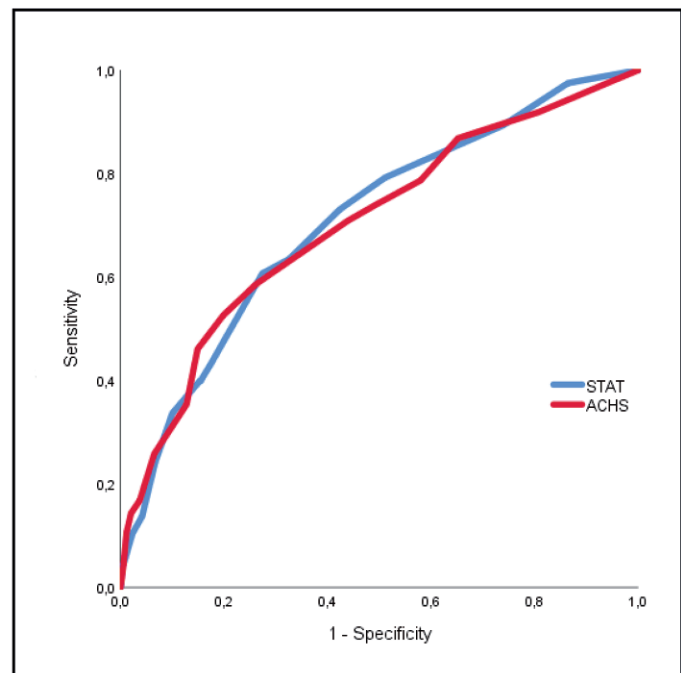
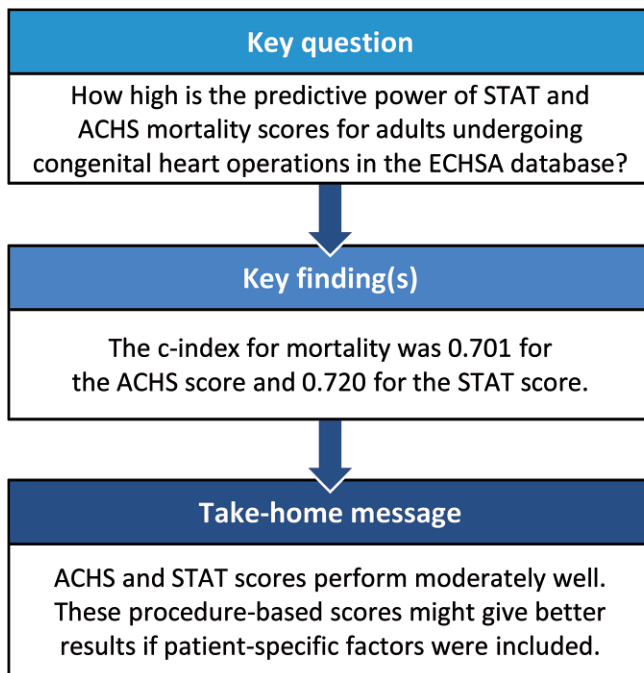
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## Abstract

**OBJECTIVES:** We sought to evaluate the predictive power of the Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery (STAT) mortality score and the adult congenital heart surgery (ACHS) mortality score for the adults undergoing congenital heart operations entered into the European Congenital Heart Surgeons Association (ECHSA) database.

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**METHODS:** The data set comprised 17 662 major operations performed between 1997 and 2019, on patients 18 years of age or older, in European centres participating in the ECHSA database. Each operation was assigned a STAT mortality score and category and an ACHS mortality score. Operative mortality was based on the 30-day status and on the status at hospital discharge. The discriminatory power of the STAT and ACHS scores was assessed using the area under the receiver operating characteristic curve (c-index).

**RESULTS:** A total of 17 214 (97.46%) operations were assigned ACHS scores. The 3 most frequent primary procedures were closure of the atrial septal defect (19.0%), aortic valve replacement (8.8%) and non-valve-sparing aortic root replacement (6.1%). Operative mortality for ACHS-coded operations was 2.07%. The procedures with the highest mortality were atrial septal defect creation/enlargement (19.0%), lung transplantation (18.8%) and heart transplantation (18.2%). A total of 17 638 (99.86%) operations were assigned a STAT score and category. The operative mortality for STAT-coded operations was 2.27%. The c-index for mortality was 0.720 for the STAT mortality score and 0.701 for the ACHS score.

**CONCLUSIONS:** The ACHS mortality score and the STAT mortality score reached similar, moderate predictive power in adult patients undergoing congenital heart surgery in ECHSA database.

**Keywords:** Grown-up congenital heart disease • Congenital heart surgery • Risk evaluation

#### ABBREVIATIONS

ACC	Aristotle comprehensive complexity
ACHD	Adults with congenital heart disease
ACHS	Adult congenital heart surgery
ASD	Atrial septal defect
CHD	Congenital heart disease
ECHSA	European Congenital Heart Surgeons Association
ECHSA-CHSD	European Congenital Heart Surgeons Association Congenital Heart Surgery Database
GUCH	Grown-Ups with Congenital Heart Disease
IQR	Interquartile range
STAT	Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery
STS	Society of Thoracic Surgeons
STS-CHSD	Society of Thoracic Surgeons Congenital Heart Surgery Database

## INTRODUCTION

Interdisciplinary improvements in the management of patients with congenital heart disease (CHD) have increased their survival to adulthood [1, 2]. In developed countries, more than 90% of patients with CHD have the prospect of reaching maturity [3, 4]. For more than 3 decades, there has been a noticeable increase in the prevalence of adults with congenital heart disease (ACHD) [5]. A significant fraction of these patients will need cardiovascular surgery at some point in their lives, either for a first corrective surgery or for reoperations to treat residual defects or long-term sequelae of CHD or previous treatment [6, 7]. The number of cardiac operations in ACHD is reported to reach 1.2–2.3 operations per 100 patient-years, depending on the age group of ACHD [8]. Early mortality following cardiac surgery in ACHD ranges from 1.8% to 3.6% [6, 7, 9–11]. Risk stratification models provide the possibility of predicting individual risk, help in counselling the patient and enable quality assessment of an ACHD centre on an interinstitutional level.

The Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery (STAT) score, the first evidence-based risk score for patients with CHD, was developed in 2009 using the combined resources of the Society of Thoracic Surgeons (STS) and the European Congenital Heart Surgeons Association (ECHSA)

databases [12]. The original STAT score stratified 148 congenital heart surgery procedures based on their relative risk of in-hospital mortality. The procedures were sorted by increasing estimated risk and partitioned into 5 homogeneous STAT categories. The STAT score and STAT categories reached good predictive power for estimating mortality (reported c-index 0.784 and 0.773, respectively) in paediatric patients and entire CHD populations [12, 13]. In ACHD, the predictive power of the STAT score was somewhat lower compared to that in the paediatric patients (reported c-index 0.60–0.777) [14, 15], however, it was superior to expert-based risk models such as Risk Adjustment for Congenital Heart Surgery categories or the Aristotle basic score [11, 16].

In 2015, the STS Congenital Heart Surgery Database (STS-CHSD) was tested on differences in procedure-specific risk of in-hospital mortality of adult and paediatric patients. Consequently, the evidence-based adult congenital heart surgery (ACHS) score was proposed for evaluating the mortality risk specifically in the ACHD [14]. The ACHS mortality score included 52 procedural groups, which comprised 94% of all operations in adults in the STS-CHSD [14]. The ACHS score reached a good predictive power in a preliminary validation sample of the STS-CHSD (c-index 0.809) [14] as well as in single-centre ACHD populations (c-index 0.760) [9].

We sought to evaluate the predictive power of STAT and ACHS mortality scores for the adults undergoing congenital heart operations entered into the ECHSA Congenital Heart Surgery Database (ECHSA-CHSD) by performing an external validation of the published models. The secondary aim of the study was to explore in more detail which operations are not coded in the ACHS score.

## PATIENTS AND METHODS

### Ethical statement

The study was carried out according to the policies of the ECHSA-CHSD (available at [www.echsacongenitaldb.org](http://www.echsacongenitaldb.org), paragraph 2). Since the individual patients were not identified, the database committee waived the need for patient consent. Patient consent is available for individual data submitting units if the respective institutional review board requires it for participating in the database.

### Inclusion and exclusion criteria

The ECHSA-CHSD [17] was scanned on 20 April 2020 for all operations performed on patients aged 18 years or more, between

1997 and 2019 in European centres. Minor surgical procedures, extracorporeal membrane oxygenation procedures, non-cardiac and unclassified surgical procedures were excluded from the analysis.

### Scoring

Each operation was assigned a STAT mortality score, a STAT mortality category and an ACHD mortality score as described in the corresponding original articles or their updated versions (STS-CHSD Version 3.3) [12, 14, 18]. For operations consisting of multiple concomitant procedures, the procedure with the highest STAT/ACHS score was set as the primary procedure and used for the analysis of each score. Procedures with no corresponding STAT or ACHS score were noted as missing and excluded from the score analysis.

Procedures with a valid STAT score were divided into 2 groups: the ACHS-coded group (operations to which an ACHS score could be assigned) and the non-ACHS-coded group (operations without an ACHS score).

### Outcome

Operative mortality was defined as death of any cause, occurring within 30 days after surgery in or out of the hospital, or after 30 days during the same hospitalization subsequent to the operation [19].

### Statistical analysis

Frequencies are given as absolute numbers and percentages. Continuous variables are expressed as the median and

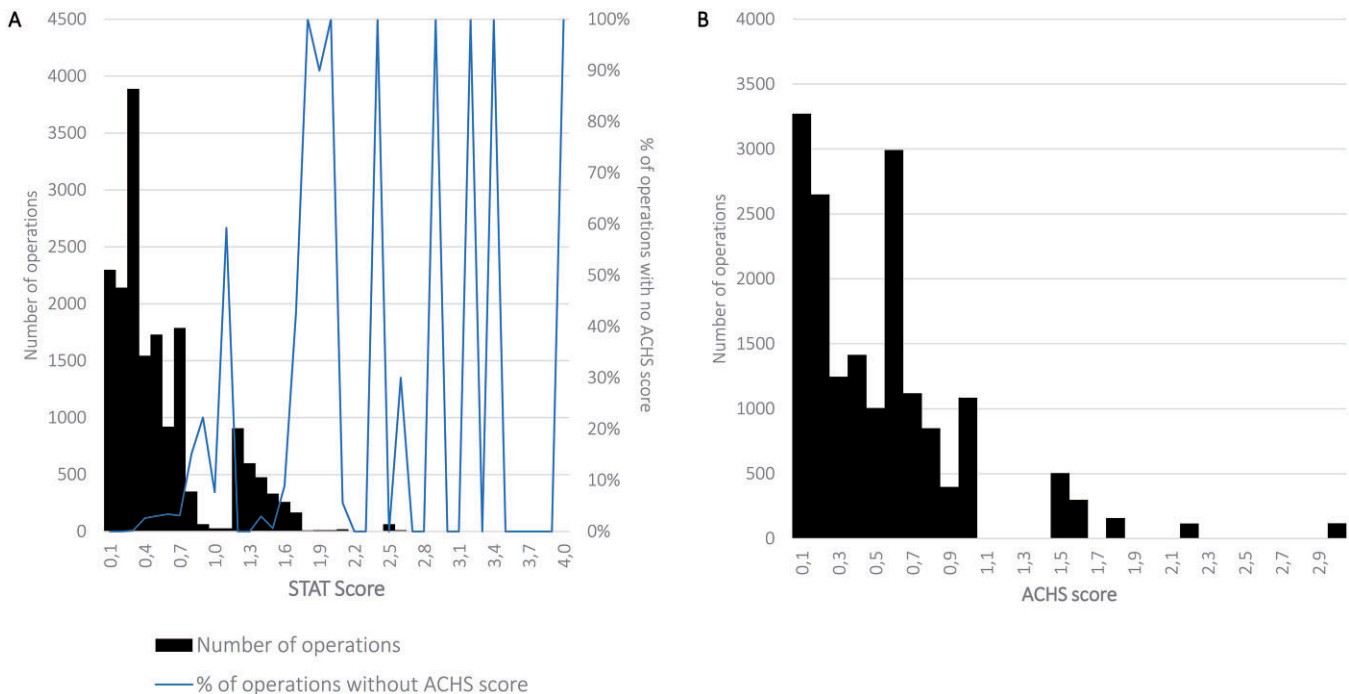
interquartile range (IQR; first-third quartile). Differences in the frequencies of the procedural groups and mortality within the procedural groups between the ECHSA-CHSD and the STS-CHSD data used for ACHS score development [14] were calculated using the two-tailed  $\chi^2$  test for each procedural group. The two-tailed  $\chi^2$  test was used to test for differences in mortality between individual STAT categories. The discriminatory powers of the STAT score and the ACHD score were assessed using the area under the receiver operating characteristic curve (c-index). The differences in the mortality and the frequency of each STAT category between the ACHS-coded and non-ACHS-coded groups were calculated using the two-tailed  $\chi^2$  test. The SPSS Median Test for 2 Independent Medians was used to compare the median STAT score between ACHS-coded and non-ACHS-coded groups. Analyses were performed with IBM SPSS Statistics for Windows, Version 26.0 (IBM SPSS Inc., Armonk, NY, USA).

### RESULTS

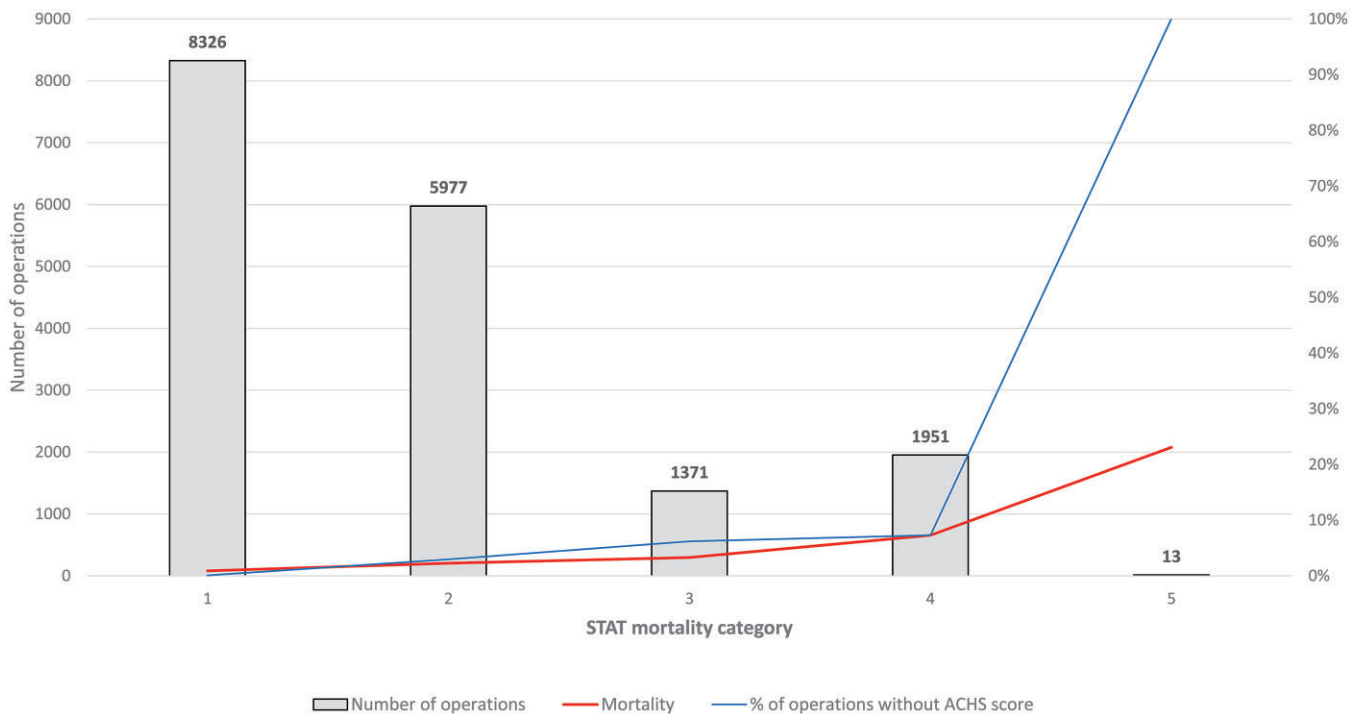
During the 23-year period, a total of 17 662 operations were performed on ACHD. The median age was 34.9 years (IQR 24.3–49.5 years). The mortality of the complete collective was 2.26%.

#### Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery mortality score and category

A total of 17 638 (99.86%) operations were assigned STAT scores and categories. The median STAT score was 0.4 (IQR 0.2–0.7). The distribution of the STAT score and the STAT categories is shown in Figs 1A and 2, respectively. Operative mortality for STAT-coded operations was 2.27%. Figure 2 illustrates the



**Figure 1:** (A) Distribution of the score values of the STAT mortality score. The percentage of operations to which no ACHS score could be assigned is depicted as a blue line on the secondary axis. (B) Distribution of the score values of the ACHS mortality score. ACHS: adult congenital heart surgery; STAT: Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery.



**Figure 2:** Distribution of the STAT mortality categories. The observed mortality of the entire cohort collective allocated to the STAT mortality categories is depicted as a red line on the secondary axis. The percentage of operations to which no adult congenital heart surgery score could be assigned is depicted as a blue line on the secondary axis. STAT: Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery.

observed operative mortality allocated to the STAT mortality categories. The mortality rate increased significantly from category 1 to 2 ( $P < 0.001$ ), 2 to 3 ( $P = 0.034$ ) and 3 to 4 ( $P < 0.001$ ).

### Adult congenital heart surgery score

A total of 17 214 (97.46%) operations were assigned ACHS scores. Table 1 shows a list of operations with absolute and relative frequency and operative mortality. The 3 most frequent primary procedures were closure of an atrial septal defect (ASD) (19.0%), aortic valve replacement (8.8%) and non-valve-sparing aortic root replacement (6.1%). The median ACHS score was 0.5 (IQR 0.2–0.7). The distribution of the ACHS scores is shown in Fig. 1B. The operative mortality for ACHS-coded operations was 2.07%. The procedures with the highest mortality were ASD creation/enlargement (19.0%), lung transplantation (18.8%) and heart transplantation (18.2%). It is important to note that the creation or enlargement of an ASD has a high ACHS score of 1.0. Therefore, in almost all operations comprising more than 1 procedure, the ASD creation/enlargement was coded as the primary operation. The exceptions were concomitant cardiac tumour resection, mitral valve replacement, coronary artery bypass, the Fontan procedure, heart transplantation, lung transplantation and Fontan revision, which have higher ACHS scores (1.5–3.0).

Significant differences occurred in the frequency of procedural groups and mortality within the procedural groups between the ECHSA-CHSD data and the STS-CHSD data used for ACHS score development [14] (Table 1). From the 52 procedural groups described in the ACHS score, operations assigned to 14 groups were performed significantly more frequently and operations from 25 procedural groups significantly less frequently in the ECHSA-CHSD compared to the STS-CHSD. Mortality was

significantly higher in 5 procedural groups in the ECHSA-CHSD compared to the STS-CHSD.

### The predictive power of Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery mortality score and adult congenital heart surgery mortality score

The receiver operating characteristic curves of the STAT score and ACHS score for operative mortality are depicted in Fig. 3. The c-index for mortality was 0.720 for the STAT mortality score and 0.701 for the ACHS score.

### Operations not coded in adult congenital heart surgery score

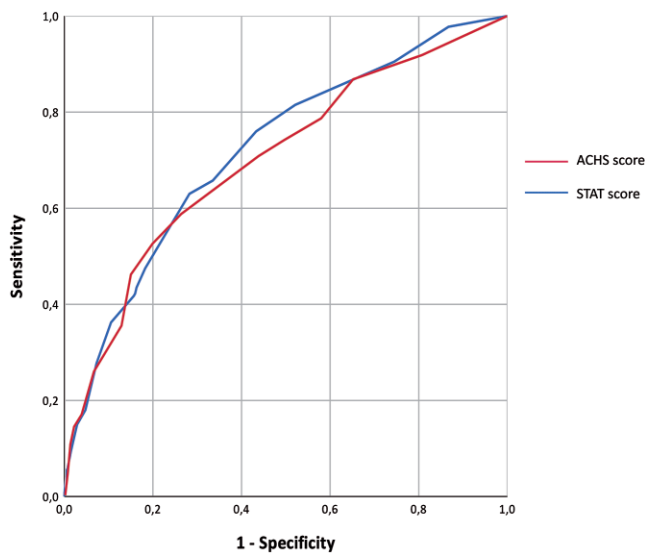
From the operations coded in the STAT score/category, 424 (2.4%) could not be assigned an ACHS score. These operations could be categorised in 54 different STAT procedures (Supplementary Material S1). The 5 most common procedures were aortic dissection repair ( $n = 44$ ), bidirectional cavopulmonary anastomosis ( $n = 37$ ), cor triatriatum repair ( $n = 36$ ), complete atrioventricular septal defect repair ( $n = 31$ ) and surgical closure of a patent ductus arteriosus ( $n = 25$ ). The percentage of operations to which no ACHS score could be assigned is depicted in Fig. 1 for each STAT score value and in Fig. 2 for each STAT category. The median STAT score in the non-ACHS-coded group of operations (0.4, IQR 0.2–0.7) was significantly higher than in the ACHS-coded group (0.8, IQR 0.6–1.7),  $P < 0.001$ . The proportion of non-ACHS-coded STAT operations increased significantly from category 1 to 2, 2 to 3 and 4 to 5 ( $P < 0.001$ ). Mortality in

**Table 1:** Comparison of frequencies and mortality of primary procedures on adults with congenital heart disease in the ECHSA-CHSD and the STS-CHSD [14]

Procedural group	ACHS mortality score	ECHSA-CHSD			STS-CHSD <sup>a</sup>			Differences in	
		N	% of 17 214	Mortality	N	% of 11 824	Mortality	Frequency	Mortality
ASD	0.1	3272	19.01	0.89	817	6.91	0.00	<0.001	0.004
Aortic stenosis, subvalvar	0.2	268	1.56	0.37	244	2.06	0.00	0.001	1
Conduit reoperation	0.2	229	1.33	1.75	232	1.96	0.00	<0.001	0.060
Pacemaker procedure	0.2	780	4.53	0.77	888	7.51	0.20	<0.001	0.16
PAPVC repair	0.2	711	4.13	0.42	299	2.53	0.00	<0.001	0.56
PV replacement	0.2	661	3.84	0.61	1110	9.39	0.50	<0.001	1
AICD implantation	0.3	109	0.63	0.92	267	2.26	0.00	<0.001	0.29
AICD procedure	0.3	95	0.55	1.05	88	0.74	0.00	0.049	1
Anomalous coronary from aorta repair	0.3	124	0.72	0.81	115	0.97	0.00	0.020	1
AV repair	0.3	239	1.39	1.26	163	1.38	0.60	0.96	0.65
Ebstein's repair	0.3	174	1.01	6.90	152	1.29	0.70	0.031	0.004
Explantation of pacing system	0.3	34	0.20	0.00	105	0.89	0.00	<0.001	NA
Pacemaker implantation, permanent	0.3	405	2.35	2.22	631	5.34	0.80	<0.001	0.059
Vascular ring repair	0.3	66	0.38	3.03	63	0.53	0.00	0.071	0.50
Anomalous coronary artery from pulmonary artery	0.4	42	0.24	0.00	41	0.35	0.00	0.12	NA
Aortic stenosis, supra-valvar	0.4	38	0.22	2.63	33	0.28	0.00	0.34	1
DCRV	0.4	50	0.29	0.00	33	0.28	0.00	0.91	NA
Konno procedure	0.4	32	0.19	3.13	54	0.46	0.00	<0.001	0.37
MV repair	0.4	566	3.29	1.24	244	2.06	0.80	<0.001	0.73
PA reconstruction	0.4	181	1.05	2.21	464	3.92	1.10	<0.001	0.28
PAPVC scimitar	0.4	44	0.26	0.00	34	0.29	0.00	0.65	NA
Pulmonic valvuloplasty	0.4	59	0.34	0.00	43	0.36	0.00	0.77	NA
RV aneurysm	0.4	9	0.05	0.00	33	0.28	0.00	<0.001	NA
RVOT repair	0.4	165	0.96	1.82	610	5.16	1.10	<0.001	0.45
Sinus of Valsalva aneurysm	0.4	23	0.13	0.00	40	0.34	0.00	<0.001	NA
Valve-sparing aortic root replacement	0.4	205	1.19	0.00	183	1.55	1.10	0.011	0.22
Aortic arch repair	0.5	46	0.27	2.17	79	0.67	1.30	<0.001	1
Common AV canal repair (incomplete)	0.5	389	2.26	0.26	147	1.24	1.40	<0.001	0.18
VSD	0.5	569	3.31	1.76	230	1.95	1.30	<0.001	0.77
Aortic aneurysm	0.6	503	2.92	1.59	288	2.44	1.70	0.013	1
AV replacement	0.6	1513	8.79	1.32	482	4.08	1.70	<0.001	0.66
Systemic venous stenosis repair	0.6	17	0.10	0.00	60	0.51	1.70	<0.001	1
TOF repair	0.6	166	0.96	2.41	58	0.49	1.70	<0.001	1
TV repair	0.6	793	4.61	1.39	432	3.65	1.90	<0.001	0.63
Arrhythmia, surgical ablation, ventricular	0.7	47	0.27	4.26	48	0.41	2.10	0.059	0.62
Conduit RV-PA	0.7	856	4.97	2.22	366	3.10	2.20	<0.001	1
Ross procedure	0.7	215	1.25	0.47	147	1.24	0.90	1	1
Arrhythmia, surgical ablation, atrial	0.8	438	2.54	3.42	457	3.87	2.40	<0.001	0.43
Coarctation repair	0.8	284	1.65	1.76	112	0.95	2.70	<0.001	0.69
Pericardial drainage	0.8	127	0.74	2.36	34	0.29	2.90	<0.001	1
Conduit placement, other	0.9	26	0.15	11.54	44	0.37	2.30	<0.001	0.14
Shunt, systemic to pulmonary	0.9	50	0.29	8.00	32	0.27	3.10	0.82	0.64
TV replacement	0.9	320	1.86	9.69	245	2.07	2.90	0.21	0.001
Aortic root replacement, non-valve-sparing	1.0	1043	6.06	2.49	291	2.46	3.10	<0.001	0.54
ASD creation or enlargement	1.0	42	0.24	19.0	214	1.81	3.30	<0.001	0.001
Cardiac tumour resection	1.5	33	0.19	0.00	54	0.46	5.50	<0.001	0.29
MV replacement	1.5	469	2.72	6.82	332	2.81	4.80	0.69	0.29
Coronary artery bypass	1.6	297	1.73	3.03	184	1.56	5.40	0.28	0.23
Fontan procedure	1.8	158	0.92	8.23	59	0.50	6.80	<0.001	1
Heart transplantation	2.2	99	0.58	18.2	227	1.92	7.50	<0.001	0.006
Lung transplantation	2.2	16	0.09	18.8	61	0.52	8.20	<0.001	0.35
Fontan revision	3.0	117	0.68	14.53	155	1.31	10.30	<0.001	0.35

<sup>a</sup>Data from Fuller *et al.* [14].

ACHS: adult congenital heart surgery; AICD: automatic implantable cardioverter-defibrillator; ASD: atrial septal defect; AV: aortic valve; AV canal: atrioventricular canal; DCRV: double-chambered right ventricle; ECHSA-CHSD: European Congenital Heart Surgeons Association Congenital Heart Surgery Database; NA: not available; MV: mitral valve; PA: pulmonary artery; PAPVC: partial anomalous pulmonary venous connection; PV: pulmonary valve; RV: right ventricle; RVOT: right ventricular outflow tract; STS-CHSD: Society of Thoracic Surgeons Congenital Heart Surgery Database; TOF: tetralogy of Fallot; TV: tricuspid valve; VSD: ventricular septal defect.



**Figure 3:** Receiver operating characteristic curves of the STAT score (blue line) and ACHS score (red line) for operative mortality. ACHS: adult congenital heart surgery; STAT: Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery.

the non-ACHS-coded group of operations [10.38% (44/424)] was significantly higher than in the ACHS-coded group [2.07% (356/17 214)],  $P < 0.001$ .

## DISCUSSION

This study evaluates the predictive power of the STS-CHSD risk models on data from the ECHSA Congenital Database. The ACHS mortality score and the STAT mortality score reached similar, moderate power in predicting mortality in ACHD operated on in European centres that participate in the ECHSA-CHSD. To understand this result, the data from the ECHSA-CHSD were compared to the STS-CHSD data used for ACHS score development [14]. The study also analysed which operations are not coded in the ACHS score compared to the STAT score. Patients undergoing the extremely rare operations that are not included in the ACHS score have significantly higher operative mortality than the rest of the cohort.

### Moderate performance of adult congenital heart surgery score

The ACHS score was the first evidence-based score designed specifically for estimating mortality in ACHD undergoing cardiac surgery. Its development included 11 824 procedures from the STS-CHSD and was based on the specific mortality of the adult patients, which in some procedural groups differed significantly from the mortality in the paediatric age [14]. The ACHS score reached a good predictive power in the validation sample of the STS-CHSD, which included 1603 operations [14] as well as in combined ACHD populations from 2 European centres with 1639 operations [9]. In this study with 17 662 operations, ACHD did not perform as well as previously reported.

To establish the reasons for this result, the frequency and the mortality of a specific procedure in the original STS-CHSD data used in developing the ACHS (original STS-CHSD) were

compared to the ECHSA-CHSD data. Although the median ACHS score was similar in both cohorts [original STS-CHSD 0.4 (IQR 0.2–0.7) vs ECHSA-CHSD 0.5 (IQR 0.2–0.7)], there were significant differences in frequency of 39 from 52 procedural groups as well as in mortality of 5 from 52 procedural groups.

The 3 most commonly performed procedures in the original STS-CHSD were pulmonary valve replacement, pacemaker procedure and ASD closure. These procedures were in the eighth, sixth and first place on the frequency list of the ECHSA-CHSD, respectively. The 3 most common procedures in the ECHSA-CHSD—ASD closure, aortic valve replacement and non-valve-sparing root replacement—were positioned in 3rd, 6th and 13th place on the frequency list of the original STS-CHSD, respectively. The operations on the aortic root and the aortic valve most likely reflected the older age of the ACHD population in the ECHSA-CHSD (median 34.9 vs 26.1 years in the original STS-CHSD) [14, 20–22].

Another potential reason for the different performance of the ACHS score are the different definitions of mortality used in the 2 studies [9]. Whereas the primary outcome in the original ACHS publication was defined as in-hospital mortality [14], the current study used the definition of operative mortality [19] that includes the in-hospital mortality but also the deaths of patients released from the hospital who died within 30 days following the operation.

The reported mortality was significantly higher in the ECHSA-CHSD compared to that in the STS-CHSD [14] (2.07% vs 1.6%;  $P < 0.001$ ). In 5 procedural groups, the mortality was significantly higher in the ECHSA-CHSD than in the original STS-CHSD: ASD closure, Ebstein's repair, tricuspid valve replacement, ASD creation/enlargement and heart transplantation. In order to explain the possible causes for the difference in the mortality, STS and ECHSA congenital databases would have to be analysed for primary diagnoses, percentages of patients with a single ventricle and/or a right systemic ventricle, pulmonary hypertension, the function of the systemic ventricle, etc., all of which exceeds the scope of this study.

### Adult congenital heart surgery score did not perform better than Society of Thoracic Surgeons–European Association for Cardio-Thoracic Surgery score and category

To further explore the reasons for the moderate performance of the ACHS score, the performance of the STAT score from which the ACHS score originated was evaluated. In this study, the STAT score reached a predictive power similar to that of the ACHS score. Since the time of its initial development, several important changes were made in the STAT scoring system [18]. Approximately 50 additional procedure codes were added to the original 148 procedures [23]. The end point changed from the initial 'discharge mortality' into the current definition of operative mortality [19]. These 2 points might clarify the somewhat better performance of the 'current STAT score' in the adult population.

In the development of the ACHS score, extremely rare procedures were excluded from the analysis [14] and therefore lack the corresponding score. Failure to capture all operations performed on ACHD patients was pointed out by Fuller *et al.* [14] to be probably the largest limitation of the ACHS score. The procedures that lacked the ACHS score in our cohort were rarely performed: each had a frequency of  $<0.25\%$  of the STAT-coded

operations. They had a significantly higher median STAT score and were more often assigned a higher STAT mortality category than the ACHS-coded operations. Operative mortality was significantly higher for the operations that could not be assigned an ACHS score (10.38%) than for the operations with a valid ACHS score (2.07%). As already suggested by Fuller *et al.* [14] at the time the ACHS score was developed, incorporation of data from the ECHSA-CHSD would likely have refined the mortality score, especially for the rare procedures.

### Adults with congenital heart disease mortality risk model: a need for including patient characteristics

The risk scores for patients with acquired heart disease often focus on a single procedure and determine the risk of an individual patient based on comorbidities and clinical factors. The European System for Cardiac Operative Risk Evaluation (EuroSCORE) II [24], one of the most widely used risk indexes for cardiac surgery, concentrates primarily on patient characteristics and can be applied to practically any cardiosurgical procedure performed. It is reported to have excellent predictive power in many international reports on patients with acquired heart disease [24]. However, this procedure non-specific score does not perform well in the ACHD population [10, 25] and tends to overestimate operative mortality. Therefore, it is clear that the type of procedure in congenital heart surgery plays a significant role in the outcome of ACHD [7].

On the other hand, due to the large number of different procedures, the risk assessment scores for congenital heart surgery are typically procedure-based. However, the individual characteristics and comorbidities of an adult patient can play an essential role in outcome following (congenital) heart surgery [2, 7, 8, 15, 26, 27]. This fact is reflected in the moderate performance of both ACHS and STAT scores when applied to ACHD.

The Aristotle comprehensive complexity (ACC) score [28] is a procedure-based score for congenital heart surgery, which includes additional procedure-dependent (anatomical factors, associated procedures) and patient-dependent (preoperative clinical and genetic factors, number of previous sternotomies, etc.) risk factors. Reports showed that the ACC score has excellent predictive power in the ACHD population (c-index 0.8) [11, 15]. Hörer *et al.* suggested using the STAT score in combination with the procedure-dependent and patient-dependent risk factors of the ACC score. In this way, the new Grown-Ups with Congenital Heart Disease (GUCH) score was created [11]. The GUCH score reached very good predictive power in the initial validation samples (c-index 0.81–0.84) [11, 29]. Since the ACC online calculator is a part of the past, no further validation or improvements to the GUCH score were reported.

The recently published STS-CHSD Mortality Risk Model [30] is a procedure-based risk model for congenital heart surgery that is applicable to ACHD. The model adjusts for additional procedure-dependent (each combination of primary procedure and age group) and patient-dependent (e.g. preoperative clinical and genetic factors, prior cardiothoracic operations) risk factors. This current STS-CHSD Mortality Risk Model reached a c-index of 0.875 in the development sample and 0.857 in the validation sample.

It is certain that with the increasing age of ACHD, a greater cumulative percentage will require an operation or reoperation. Risk modelling will become more important for this population.

However, the complexity of this patient group will also increase as these patients get older. Interestingly, the patients with more concomitant procedures during an operation had a significantly higher mortality rate in the ECHSA-CHSD ( $P < 0.001$ ). The late sequelae of abnormal physiology or previous treatments will reveal themselves, and the acquired heart disease will start mixing with the congenital heart defect. Finally, all ACHD may require non-cardiac operations. Anaesthesiologists and non-congenital heart surgeons are often not aware of the potentially high-risk profile of these patients and would profit from a patient-related component of a risk model for ACHD.

### Limitations

The data in the ECHSA-CHSD are compiled from multiple European centres, some with a small annual load. The relationship between the annual caseload of a centre and the outcome was outside the spectrum of this study. A thorough analysis of the STS and ECHSA congenital databases from this perspective might partially explain the different performance of the STAT and ACHS scores. The clinical data entered in the ECHSA-CHSD was verified in an external ECHSA audit for 4617 of 17638 operations (26.2%). The enrolled patients underwent surgery over a 23-year period, during which time the surgical outcome of surgery has improved. Often, more than 1 procedure was performed during the operation. It may be possible that procedures other than the primary one with the highest score influenced the outcome.

### CONCLUSION

The ACHS and STAT mortality scores have moderate predictive power when applied to adult patients undergoing congenital heart surgery in ECHSA-CHSD. Both scores are exclusively procedure-based, which might be inadequate for risk stratification in ACHD. The inclusion of patient-specific factors might improve the predictive power of the risk models. Furthermore, the development of such risk models with a specific focus on adults undergoing surgery for congenital heart disease is needed.

### SUPPLEMENTARY MATERIAL

Supplementary material is available at *EJCTS* online.

**Conflict of interest:** Georgios Sarris, Zdzislaw Tobota, Bohdan Maruszewski, Vladimiro L. Vida and Jürgen Hörer are the members of the database committee of the European Congenital Heart Surgeons Association. The other author declared no conflict of interest.

### Author contributions

**Jelena Pabst von Ohain:** Formal analysis; Methodology; Validation; Visualization; Writing—original draft. **Georgios Sarris:** Data curation; Writing—review & editing. **Zdzislaw Tobota:** Conceptualization; Data curation; Writing—review & editing. **Bohdan Maruszewski:** Conceptualization; Data curation; Writing—review & editing. **Vladimiro L. Vida:** Writing—review & editing. **Jürgen Hörer:** Conceptualization; Writing—review & editing.

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## REFERENCES

- [1] Khairy P, Ionescu-Iltu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. *J Am Coll Cardiol* 2010;56:1149–57.
- [2] Diller GP, Kempny A, Alonso-Gonzalez R, Swan L, Uebing A, Li W *et al.* Survival prospects and circumstances of death in contemporary adult congenital heart disease patients under follow-up at a large tertiary centre. *Circulation* 2015;132:2118–25.
- [3] Moons P, Bovijn L, Budts W, Belmans A, Gewillig M. Temporal trends in survival to adulthood among patients born with congenital heart disease from 1970 to 1992 in Belgium. *Circulation* 2010;122:2264–72.
- [4] Warnes CA. The adult with congenital heart disease: born to be bad? *J Am Coll Cardiol* 2005;46:1–8.
- [5] Marelli AJ, Ionescu-Iltu R, Mackie AS, Guo L, Dendukuri N, Kaouache M. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. *Circulation* 2014;130:749–56.
- [6] Vida VL, Berggren H, Brawn WJ, Daenen W, Di Carlo D, Di Donato R *et al.* Risk of surgery for congenital heart disease in the adult: a multicentered European study. *Ann Thorac Surg* 2007;83:161–8.
- [7] Horer J. Current spectrum, challenges and new developments in the surgical care of adults with congenital heart disease. *Cardiovasc Diagn Ther* 2018;8:754–64.
- [8] Tutarel O, Kempny A, Alonso-Gonzalez R, Jabbour R, Li W, Uebing A *et al.* Congenital heart disease beyond the age of 60: emergence of a new population with high resource utilization, high morbidity, and high mortality. *Eur Heart J* 2014;35:725–32.
- [9] Hörer J, Belli E, Roussin R, LeBret E, Ly M, Abdullah J *et al.* Evaluation of the adult congenital heart surgery mortality score at two European centers. *Ann Thorac Surg* 2018;105:1441–6.
- [10] Beurtheret S, Tutarel O, Diller GP, West C, Ntalarizou E, Resseguier N *et al.* Contemporary cardiac surgery for adults with congenital heart disease. *Heart* 2017;103:1194–202.
- [11] Horer J, Kasnar-Samprec J, Cleuziou J, Strbad M, Wottke M, Kaemmerer H *et al.* Mortality following congenital heart surgery in adults can be predicted accurately by combining expert-based and evidence-based pediatric risk scores. *World J Pediatr Congenit Heart Surg* 2016;7:425–35.
- [12] O'Brien SM, Clarke DR, Jacobs JP, Jacobs ML, Lacour-Gayet FG, Pizarro C *et al.* An empirically based tool for analyzing mortality associated with congenital heart surgery. *J Thorac Cardiovasc Surg* 2009;138:1139–53.
- [13] Jacobs JP, Jacobs ML, Maruszewski B, Lacour-Gayet FG, Tchervenkov CI, Tobota Z *et al.* Initial application in the EACTS and STS Congenital Heart Surgery Databases of an empirically derived methodology of complexity adjustment to evaluate surgical case mix and results. *Eur J Cardiothorac Surg* 2012;42:775–9.
- [14] Fuller SM, He X, Jacobs JP, Pasquali SK, Gaynor JW, Mascio CE *et al.* Estimating mortality risk for adult congenital heart surgery: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. *Ann Thorac Surg* 2015;100:1728–35.
- [15] van Gameren M, Putman LM, Takkenberg JJ, Bogers AJ. Risk stratification for adult congenital heart surgery. *Eur J Cardiothorac Surg* 2011;39:490–4.
- [16] Kogon B, Oster M. Assessing surgical risk for adults with congenital heart disease: are pediatric scoring systems appropriate? *J Thorac Cardiovasc Surg* 2014;147:666–71.
- [17] European Congenital Heart Surgeons Association. 2020. <https://www.echsacongenitaldb.org/> (20 April 2020, date last accessed).
- [18] Society of Thoracic Surgeons. 2016. <https://www.sts.org/registries-research-center/sts-national-database/congenital-heart-surgery-database/data-collection> (20 April 2020, date last accessed).
- [19] Jacobs JP, Mavroudis C, Jacobs ML, Maruszewski B, Tchervenkov CI, Lacour-Gayet FG *et al.* What is operative mortality? Defining death in a surgical registry database: a report of the STS Congenital Database Taskforce and the Joint EACTS–STS Congenital Database Committee. *Ann Thorac Surg* 2006;81:1937–41.
- [20] Niwa K. Aortic dilatation in complex congenital heart disease. *Cardiovasc Diagn Ther* 2018;8:725–38.
- [21] Horer J, Kasnar-Samprec J, Charitos E, Stierle U, Bogers AJ, Hemmer W *et al.* Patient age at the Ross operation in children influences aortic root dimensions and aortic regurgitation. *World J Pediatr Congenit Heart Surg* 2013;4:245–52.
- [22] Lange R, Cleuziou J, Horer J, Holper K, Vogt M, Tassani-Prell P *et al.* Risk factors for aortic insufficiency and aortic valve replacement after the arterial switch operation. *Eur J Cardiothorac Surg* 2008;34:711–17.
- [23] Society of Thoracic Surgeons. 2019. <https://www.sts.org/sites/default/files/102519%20835.%20Jacobs.%20Update%20to%20STAT%20Scores.pdf> (1 May 2020, date last accessed).
- [24] Nashef SA, Roques F, Sharples LD, Nilsson J, Smith C, Goldstone AR *et al.* EuroSCORE II. *Eur J Cardiothorac Surg* 2012;41:734–44.
- [25] Putman LM, van Gameren M, Meijboom FJ, de Jong PL, Roos-Hesselink JW, Witsenburg M *et al.* Seventeen years of adult congenital heart surgery: a single centre experience. *Eur J Cardiothorac Surg* 2009;36:96–104.
- [26] O'Brien SM, Jacobs JP, Pasquali SK, Gaynor JW, Karamlou T, Welke KF *et al.* The Society of Thoracic Surgeons Congenital Heart Surgery Database mortality risk model: part 1—statistical methodology. *Ann Thorac Surg* 2015;100:1054–62.
- [27] Jacobs JP, O'Brien SM, Pasquali SK, Kim S, Gaynor JW, Tchervenkov CI *et al.* The importance of patient-specific preoperative factors: an analysis of the Society of Thoracic Surgeons Congenital Heart Surgery Database. *Ann Thorac Surg* 2014;98:1653–8.
- [28] Lacour-Gayet F, Clarke D, Jacobs J, Comas J, Daebritz S, Daenen W *et al.* The Aristotle score: a complexity-adjusted method to evaluate surgical results. *Eur J Cardiothorac Surg* 2004;25:911–24.
- [29] Hörer J, Roussin R, LeBret E, Ly M, Abdullah J, Marzullo R *et al.* Validation of the grown-ups with congenital heart disease score. *Heart* 2018;104:1019–25.
- [30] Jacobs JP, O'Brien SM, Hill KD, Kumar SR, Austin EH, Gaynor JW *et al.* Refining the Society of Thoracic Surgeons Congenital Heart Surgery Database mortality risk model with enhanced risk adjustment for chromosomal abnormalities, syndromes, and noncardiac congenital anatomic abnormalities. *Ann Thorac Surg* 2019;108:558–66.