Risk stratification in paediatric open-heart surgery

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Abstract

Objective: The aims of this study were to identify independent risk factors for mortality following paediatric open-heart surgery and to develop risk models for use in clinical audit based on identified risk factors. The study also tests the validity of the recently proposed Risk Adjustment in Congenital Heart Surgery (RACHS-1) method of risk stratification\cite{J Thorac Cardiovasc Surg 123 (2002) 110} as applied to open-heart operations.

Methods: A multiple logistic regression analysis was performed on all patients less than 18 years of age undergoing open-heart surgery at a single institution over a 3-year period. Preoperative and operative variables included for analysis were age at operation, weight, sex, American Society of Anaesthesiology (ASA) grade, RACHS-1 risk category, preoperative haemoglobin, bypass time, temperature, cross-clamp time, circulatory arrest time, blood transfusion on bypass and surgeon. The outcome measure was in-hospital death.

Results: 1085 consecutive open-heart cases were identified. There were 51 in-hospital deaths (4.7%). Variables identified as being independently significant risk factors for in-hospital death were age \((P = 0.0002)\), RACHS-1 risk category \((P < 0.0001)\) and bypass time \((P < 0.0001)\). Based on these three variables, a risk model was constructed to predict mortality. The area under the receiver-operating-characteristic (ROC) curve for this model was 0.86. A second model was constructed ignoring bypass time. In this model, the significance of the ‘preoperative’ risk factors was \(P = 0.0003\) for age and \(P < 0.0001\) for RACHS-1 risk category. The area under the ROC curve was 0.81 for the second model.

Conclusions: This study identifies age at operation, RACHS-1 risk category and bypass time as highly significant risk factors for mortality after paediatric open-heart surgery. It validates the RACHS-1 risk stratification method as applied to the subset of open-heart surgery, whilst accepting the limitations of such a system. The risk models formulated permit risk prediction and allow for analysis of surgical results. Such risk-adjustment is important when assessing performance and comparing outcomes amongst individuals or institutions.

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Keywords: Mortality; Risk factors

1. Introduction

Risk stratification in paediatric cardiac surgery is problematic owing to wide variations in casemix. Previous studies have documented differences in outcome between open and closed cases, and amongst different age groups\cite{2,3}. Other research has suggested factors such as institutional caseload and individual surgeon volume may affect results\cite{4,5}. Studies in neonates have demonstrated that preoperative risk factors such as age at admission, genetic diagnosis, Apgar scores and ventricular morphology may be important, as well as duration of circulatory arrest\cite{6}. Insurance patterns and access to medical care have also been shown to influence outcome\cite{7,8}.

Other efforts have been made to refine risk models by grouping many different congenital heart operations into a small number of categories ranked in order of perceived increasing risk\cite{1,9,10}. Jenkins et al. have recently proposed a ‘Risk Adjustment In Congenital Heart Surgery’ classification (RACHS-1), which has been shown to predict hospital mortality\cite{1,11}. This method, which is utilised in this study, places 79 individual open and closed operations into 6 risk categories (see Appendix B). The authors demonstrated that this grading system alone could be used for risk stratification, but that it could also be refined further by including age group, prematurity and major non-cardiac anomaly into the risk modelling.
The aim of this study was to perform a risk factor analysis for hospital mortality following paediatric open-heart surgery, and to test the validity of the RACHS-1 method as applied to open-heart cases. Risk models were then constructed based on identified risk factors to develop a risk-stratification method that could be applied for use in clinical audit.

2. Materials and methods

A 3-year retrospective cohort study (April 2000–March 2003) of paediatric open-heart surgery at a single institution was conducted. Prospectively collected data were retrieved from the hospital’s computerised cardiac surgery database. Data quality is validated by participation in the Central Cardiac Audit Database Project (CCAD), which is a national database funded by the Department of Health, overseen by paediatric cardiac surgeons and cardiologists and managed by the National Health Service Information Authority in the United Kingdom.

The dataset was checked for errors and omissions by cross-referencing with another similar but separate computerised hospital operating theatre database, and also by manual reference to perfusionists’ records, operation reports and departmental discharge summaries. RACHS-1 risk categories were assigned to each case retrospectively by strict adherence to the classification scheme as published in January 2002 [1].

2.1. Patients and variables

1085 open-heart cases in patients less than 18 years of age were identified. Open-heart surgery was defined as any operation employing cardiopulmonary bypass [3]. The peri-operative variables examined were age at operation, weight, sex, American Society of Anaesthesiology (ASA) grade (see Appendix C), RACHS-1 risk category, preoperative haemoglobin, total bypass time, lowest core temperature on bypass, cross-clamp time, circulatory arrest time, total red cell transfusion during bypass and surgeon. The outcome variable was in-hospital death. In cases where a death occurred following more than one open-heart operation during the same hospital admission, the death was only counted once and attributed to the primary operation.

2.2. Statistical analysis

Multiple logistic regression analysis was used to develop two separate prediction equations for in-hospital death, one restricted to factors available pre-operatively (the pre-operative model) and the other using all available data (the peri-operative model). Fractional polynomials of weight and age were used to optimise their association with the outcome (with 1 day added to age). A substantial minority (8%) of cases were operations not defined by the RACHS-1 grading system (see Table 3). The mortality of this group of patients approximated that of RACHS-1 risk category 3. The predictive equations were developed with (a) the cases omitted, and (b) the cases treated as RACHS-1 risk category 3.

Two alternative effects of bypass time on mortality were investigated: a linear effect, the log odds increasing linearly with bypass time, and a threshold effect, the risk assumed constant for bypass times below a given threshold, and then constant but higher at or beyond the threshold. The optimal

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value*</th>
<th>Odds ratio</th>
<th>Mortality</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (months)</td>
<td>11.6 (0–198)</td>
<td>1.32*</td>
<td>0.0001*</td>
<td></td>
</tr>
<tr>
<td>Weight (kg)</td>
<td>8.1 (1.8–81)</td>
<td>1.13b</td>
<td>0.0001b</td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>55.2%</td>
<td>4.8%</td>
<td>0.8</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>44.8%</td>
<td>4.5%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>RACHS-1 risk category</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>7.7%</td>
<td>0%</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>42.2%</td>
<td>1.2%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>38.1%</td>
<td>5.0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>8.2%</td>
<td>11.0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>–</td>
<td>–</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>3.7%</td>
<td>35.1%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unable to be classified</td>
<td>8.0%</td>
<td>5.7%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ASA grade</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>2.5%</td>
<td>0%</td>
<td>0.002</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>14.5%</td>
<td>2.0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>62.9%</td>
<td>3.2%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>18.9%</td>
<td>10.7%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>1.3%</td>
<td>0%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Preoperative haemoglobin (g/dl)</td>
<td>12.2 (6.3–22.5)</td>
<td>0.93</td>
<td>0.2</td>
<td></td>
</tr>
<tr>
<td>Total bypass time (min)</td>
<td>103 (9–930)</td>
<td>1.01</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>Total cross–clamp time (min)</td>
<td>44 (0–543)</td>
<td>1.006</td>
<td>0.004</td>
<td></td>
</tr>
<tr>
<td>Circulatory arrest</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>86.4%</td>
<td>3.4%</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>13.6%</td>
<td>12.8%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Circulatory arrest time (min)</td>
<td>20 (2–83)</td>
<td>1.04</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>when used</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Red cell transfusion during bypass (ml/kg)</td>
<td>40 (0–188)</td>
<td>1.02</td>
<td>&lt;0.0001</td>
<td></td>
</tr>
<tr>
<td>Surgeon</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>A</td>
<td>16.5%</td>
<td>3.4%</td>
<td>0.2</td>
<td></td>
</tr>
<tr>
<td>B</td>
<td>14.1%</td>
<td>2.6%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>26.8%</td>
<td>6.9%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>D</td>
<td>36.2%</td>
<td>4.9%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>E</td>
<td>6.3%</td>
<td>2.9%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Continuous variables: median (range); Categorical variables: %, ASA, American Society of Anesthesiology; RACHS-1 Risk Adjustment in Congenital Heart Surgery [1].

1 transformed age = \sqrt[365]{(\text{age in days} + 1)}.

1 transformed weight = 100/(\text{weight in kg}^2).
threshold was chosen to give the most significant effect on mortality.

There were no deaths among RACHS-1 risk category 1 patients, which meant that separate estimates of mortality by RACHS-1 risk category could not be estimated (logistic regression requires group mean mortality rates to exclude 0% and 100%). Instead the RACHS-1 risk category was treated as a continuous variable in the analysis, assuming a linear relationship between risk category and mortality (on the log odds scale). This implied a small but non-zero mortality rate for risk category 1.

For sensitivity analyses, weight was expressed as a z-score using post-natal age and infants under 3 months with a z-score below 2 were rated as premature. This variable was added to models containing weight and/or age to see how the results were affected. The analyses were repeated with different values for the age cut-off (default 3 months) and z-score (default -2) cut-off.

3. Results

Of the 1085 cases, 51 died in hospital (4.7%). Table 1 summarises the univariate analysis of the risk factors studied, showing that several of the variables were significantly related to mortality. Table 2 summarises the results of the multiple logistic regression analysis, listing the independently significant risk factors for both the ‘pre-operative’ and ‘peri-operative’ models.

The associations of age and weight with mortality are rather complex. This is because the relationships between age and mortality, and weight and mortality, are not linear. Mortality is only increased in the smallest, lightest and youngest infants. To model this effectively, weight was transformed to $100/\text{weight}^2$ ($P < 0.0001$ on univariate testing) and age to $365/(\text{age in days} + 1)$ ($P < 0.0001$). After adjusting for transformed weight and transformed age simultaneously, weight was no longer significant ($P = 0.2$) while age remained highly significant ($P < 0.0001$). So the best indicator of raised mortality in young infants was age rather than weight.

Adding prematurity to the models, in addition to age and/or weight, made no appreciable difference to the results, and in particular it did not alter the conclusion that age was more important than weight.

In addition to age, the RACHS-1 risk category was the only other significant variable in the pre-operative model (odds ratio 1.89, 95% confidence interval 1.4–2.5, $P < 0.0001$). This indicates that the odds of death increased by 1.89 times per category. Fig. 1 shows the linear trend in mortality versus RACHS-1 risk category (on a log odds scale). Note that the zero mortality for risk category 1 is at minus infinity on the log odds scale. Fig. 2 shows the non-linear effect of age after adjusting for RACHS-1 risk category.

For the peri-operative model, total bypass time was also highly significant, whether modelled as a linear or a threshold effect. The univariate relationship between bypass time and mortality is displayed in Fig. 3. But in the regression model the threshold effect, with the threshold set at 200 min, was considerably more significant than the linear effect. The risk of death in

<table>
<thead>
<tr>
<th>Variable</th>
<th>Regression coefficient</th>
<th>Odds ratio (95% CI)</th>
<th>$P$-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Pre-operative model (n = 998)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transformed age</td>
<td>0.163</td>
<td>1.18 (1.08–1.29)</td>
<td>0.0003</td>
</tr>
<tr>
<td>RACHS-1 grade</td>
<td>0.636</td>
<td>1.89 (1.44–2.49)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td><strong>Peri-operative model (n = 991)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Transformed age</td>
<td>0.147</td>
<td>1.16 (1.05–1.27)</td>
<td>0.002</td>
</tr>
<tr>
<td>RACHS-1 grade</td>
<td>0.527</td>
<td>1.98 (1.46–2.68)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Total bypass time 200 + min</td>
<td>2.69</td>
<td>14.70 (6.5–33.1)</td>
<td>&lt;0.0001</td>
</tr>
</tbody>
</table>

Table 2

Results of multiple logistic regression analyses (see text for details)

RACHS-1 Risk Adjustment in Congenital Heart Surgery [1].

* transformed age = $\sqrt{365/(\text{age in days} + 1)}$. 

Fig. 1. RACHS-1 risk category versus log odds of death. Note that there were no cases performed in risk category 5 and that there were no deaths in risk category 1 (log odds $= -\infty$).
operations lasting 200 or more minutes was almost 15 times greater than in shorter operations.

RACHS-1 risk categories could not be assigned in 87 cases (8.0%) as listed in Table 3. The mortality for this group was 5.7%, similar to patients in RACHS-1 risk category 3.

The area under the ROC curve (AUC) for the pre-operative model using the risk factors age and RACHS-1 risk category was 0.81. For the peri-operative model with the three factors age, RACHS-1 risk category and total bypass time, the AUC was 0.86.

4. Discussion

Risk models for adult cardiac surgery have been well established and refined over the last 15 years [12,13]. However congenital heart surgery has lagged behind in the development of systems of risk stratification. As public awareness and government scrutiny of health care outcomes has escalated, the need for validated risk models is becoming paramount in the interpretation of surgical results [14–16].

The RACHS-1 method represents one such means of risk stratification that might potentially be useful for comparing inter-institutional outcomes, and its recent publication [1] was the impetus for initiating our study. It is a procedure-based risk scheme, rather than a diagnosis-based method. This study in part examines its validity and attempts to identify its strengths and weaknesses. Another such consensus-based model currently being developed by the European Association of Cardiothoracic Surgery, the Society for Thoracic Surgeons, the Congenital Heart Surgeons Society and the European Congenital Heart Surgeons Foundation is the so-called ‘Aristotle score’ [10]. This differs significantly from the RACHS-1 method in being a complexity score, rather than a risk grade. The scoring system gives weight to a number of factors for any

<table>
<thead>
<tr>
<th>Operation</th>
<th>Number of cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart transplant</td>
<td>51</td>
</tr>
<tr>
<td>Heart-lung transplant</td>
<td>5</td>
</tr>
<tr>
<td>Bilateral lung transplant using CPB</td>
<td>3</td>
</tr>
<tr>
<td>Tracheal reconstruction using CPB</td>
<td>11</td>
</tr>
<tr>
<td>Repair of pulmonary vein stenosis</td>
<td>7</td>
</tr>
<tr>
<td>Right ventricular remodelling</td>
<td>3</td>
</tr>
<tr>
<td>Coronary arterioplasty or CABG</td>
<td>2</td>
</tr>
<tr>
<td>Resection of intra-abdominal tumour using CPB</td>
<td>2</td>
</tr>
<tr>
<td>Other</td>
<td>3</td>
</tr>
</tbody>
</table>

CABG, coronary artery bypass grafting; CPB, cardiopulmonary bypass.
individual case, including estimated risk of mortality and morbidity, relative technical difficulty, diagnosis-dependent factors (such as presence of an intramural coronary artery in transposition) and diagnosis-independent factors (such as weight and age).

Our study identified three independent risk factors for paediatric open-heart surgery: age at operation, RACHS-1 risk category and total bypass time. The study therefore validates the RACHS-1 method as it applies to the subset of open-heart surgery, and demonstrates an extremely strong relationship between RACHS-1 risk category and risk of postoperative death. Moreover, the relationship was shown to be linear (on the log odds scale) in this study, a point which was not observed in the paper published by Jenkins et al. [1] but which adds strength to their proposed grading system.

However we noted a number of deficiencies in the RACHS-1 method. Firstly, a small but important minority of cases (8%) in our series could not be classified using the RACHS-1 method. Secondly, none of our 1085 cases fell into RACHS-1 risk category 5 (see Appendix B), rendering it meaningless, as noted by Jenkins et al. themselves. We suggest that this category be revised and if necessary removed altogether. Thirdly, some of the procedures listed are rather vague or ambiguous (e.g. 'repair of unspecified septal defect') or open to variable interpretation (e.g. 'repair of double outlet right ventricle'). This therefore leaves the system open to errors in coding owing to inadequate definition of nomenclature. We also have concerns about the classification of combination procedures, which are assigned according to the category of highest risk. For example, we are unsure how to classify the 'problem' in our series of a zero mortality in RACHS-1 risk category 1 by making certain statistical assumptions (as detailed in Section 2).

Our evaluation of the RACHS-1 method is that it is statistically useful when applied to clinical audit of large numbers of patients. By its nature however, it lacks precision when estimating risk for individual patients and should not be used for this purpose (as its authors correctly indicate). This is because many different and varied operations are lumped together in each group. Even for the same operation, different anatomical features may imply greater operative risk. Clinicians understand that, for example, the presence of an intramural coronary artery in an arterial switch operation, or a Fontan operation in a patient with elevated pulmonary vascular resistance, represents an increased risk. The advantage of the RACHS-1 method is in overcoming the lack of

statistical power when trying to demonstrate differences amongst patients with so many different variables. The disadvantage is that it fails to account for subtle but important clinical characteristics and is therefore a rather blunt tool in assigning risk. It awaits to be seen whether a complexity-based scoring system such as the Aristotle score [10] can better allow estimation of risk in individual cases as well as patient populations.

We should point out that in our evaluation of the RACHS-1 method, we have excluded all 'closed heart' (non-bypass) cases. This is because our study was primarily aimed at identifying risk factors related to open-heart surgery (such as bypass time, cross clamp time, etc.). Therefore we are unable to pass judgement on the entire RACHS-1 classification system. Furthermore, the survival rates achieved by our institution for the various RACHS-1 categories are not directly comparable to those quoted by others because of these exclusions.

Although bypass time had a significant effect on mortality in our risk model, it may in fact be acting as a surrogate of outcome. In other words, in cases that carry a high mortality, long bypass time may simply be reflecting difficulties during surgery (e.g. inability to wean from bypass, multiple bypass runs to revise the repair). This became evident statistically when observing the threshold effect of 200 min, beyond which the odds of death increased almost 15-fold. It might therefore be too simplistic to state that minimising time on bypass can reduce the risk of death. For this reason, different models were produced either including or excluding bypass time (see Table 2). Both models have significant values for area under the ROC curve, i.e. they are both highly accurate in their prediction of outcome.

The association between age and risk of mortality is also extremely strong, but non-linear. This is because in our study, the risk only starts to rise importantly once age falls into the neonatal period. Beyond the neonatal period, risk becomes almost constant within each RACHS-1 group (see Fig. 2). However the risk rises markedly and exponentially with each decreasing day of life in the neonatal period. Previous studies have also demonstrated higher risk in younger patients, but have typically measured risk by age groups (<30 days, 30 days to 1 year, >1 year) [1,3] Our results suggest that comparing results amongst age strata is in fact a fairly crude form of risk-stratification, and that risk models can be significantly refined when age is measured as a continuous variable, remembering the non-linear association with mortality.

It is important to emphasise that this relationship does not imply that neonatal surgery becomes safer if delayed. In many instances, such as simple transposition or hypoplastic left heart syndrome, there may well be a detrimental effect in postponing surgery. Our data have an inherent selection bias because the age at operation has been preselected in every individual case, based on our clinical judgement as to the optimal timing of surgery. All we can safely conclude is that babies who required bypass surgery at a very young age
had an importantly higher risk of death than older babies. Trying to prove scientifically the optimal age for surgery in any specific lesion (e.g. tetralogy of Fallot) is probably impossible, as this could only be determined in a prospectively controlled trial.

A fourth factor, weight, was shown to be important but not as significant as age. Although weight and age are closely correlated, it can be concluded that age is the more important determinant of outcome. However this has implications for prematurity, a factor that was not directly measured in our study. As a sensitivity analysis, a proxy measure for prematurity was constructed based on age and weight z-score, on the basis that premature infants tend to have low z-scores when young. Adding this variable to the models made no difference to the results, a finding different to that in the study by Jenkins et al. [1]. The explanation may be that few infants in our own series were premature, and/or that we tend to postpone bypass surgery where possible or offer palliation (closed surgery) in very premature babies. This represents a possible selection bias in our data and we therefore cannot completely discount prematurity as a potentially important risk factor.

Reassuringly, there were no significant differences in mortality amongst surgeons in this series. Interestingly, intraoperative variables such as cross-clamp time, circulatory arrest and transfusion were significant predictors of mortality on univariate testing, but became non-significant once adjusted for RACHS-1 risk category. In an attempt to include the importance of acute physiological status and comorbidity present at the time of surgery, we chose to use the ASA grade (see Appendix C). This widely used grading system is a global index of a patient’s immediate preoperative condition and can therefore be representative of a multitude of factors (e.g. ventilator-dependence, congestive heart failure, critical hypoxia). The ASA system, although highly subjective in nature [17], has been shown to be a valid measure of perioperative risk in other types of surgery [18,19]. Somewhat surprisingly, ASA grade was found to have no significant influence over postoperative mortality in multivariate analysis. It may be that age and RACHS-1 risk category already have implications for comorbidity. For example, a patient aged 2 months undergoing VSD closure is likely to have more important congestive heart failure than a patient having VSD closure aged 12 months. Likewise patients in higher RACHS-1 risk categories are usually (but not always) more acutely unwell and operated on more urgently than those in lower grades.

Chromosomal disorders, non-cardiac anomalies and syndromes were not analysed in this study. Since not all patients at our institution are screened for genetic disorders, and not all non-cardiac comorbidity is recorded in our database, these variables were excluded from the study. However as the understanding of genetic determinants of disease continues to advance and genetic screening becomes routine practice, genotype could well be found to play an important role in pre-determining outcome.

### 4.1. Limitations of this study

As with other statistical risk-stratification models, there are a number of limitations and therefore precautions that should be taken into account when applying this model to clinical audit. Firstly, the data analysed are by definition retrospective and therefore automatically historical. This problem is well recognised in older cardiac surgery risk models such as the Parsonnet score [20], where outcomes by today’s standards would be expected to be superior to those at the time the model was published. Secondly, all models fall short of being 100% accurate in predictive ability. Despite a high level of discrimination in our model (which compares favourably with those commonly used in adult cardiac surgery [12,13]), there are finite limitations of statistical methods to measure biological outcomes. We have attempted to address some of the deficiencies of the RACHS-1 risk stratification system in the preceding discussion. Finally, future research may uncover new factors (e.g. genetic) that are subsequently shown to have important effects on outcome. This underscores the need for risk-stratification models to be able to adapt to surgical science and practice that continues to evolve.

### 4.2. Conclusion

This study identifies independent risk factors for mortality after paediatric open-heart surgery. It validates the recently proposed RACHS-1 method as applied to open-heart surgery, whilst accepting the shortcomings of such a system. The data provide a risk model that can be used to predict outcome and allow comparison of results amongst appropriately risk-stratified groups of patients, provided the necessary safeguards are applied and that the limitations and caveats associated with the model are understood.

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### References

Appendix A. Conference discussion

**Dr C. Tchervenkov (Montreal, Canada):** Can you expand a little bit on the issue of age? Did you analyze this for the overall cohort or per lesion? Some lesions have a higher mortality with younger age, perhaps tetralogy of Fallot, while others have a greater risk of death with increasing age, such as the arterial switch operation for transposition or the Norwood operation for hypoplastic left heart syndrome. Can you elaborate this point, since depending on the lesion the impact of age may have an opposite effect.

My second point is regarding category 5 of Jenkins. It doesn’t seem to be very practical or relevant, since in her latest paper and your paper, category 5 has almost no patients. So perhaps it ought to be eliminated and either moved up or down to the next category.

**Dr Kang:** I think you’ve raised a couple of good points. One of the limitations of our study is that there is a selection bias involved. In other words, we selected what age to operate on what patients. For example, in our institution, most but not all tetralodies would have palliation first with a shunt and then have a repair operation several months down the track. So already we have a selection bias in our risk model. What we can say is that age, based on our data, is independently predictive of death; that means independent of the lesion or independent of the risk category.

I agree completely with you about risk category 5 in the Jenkins system. We had none of about 1000 patients in that category, and I think in the original paper, very few of nearly 10,000 patients were in that risk category. So it sounds like that risk category should be discarded altogether.

**Dr Tchervenkov:** I really think that the issue of age has to be further studied in the future because the wrong inference may be made based on this paper as to when to operate on some patients.

**Dr Kang:** Exactly. We’re not saying that we should delay surgery in neonates for as long as possible for the obvious reasons you’ve made, and that’s why there is a bias in our study, a selection bias. All we’re saying is that when you need to operate at a very young age, in the first week of life or perhaps the first 10 days of life, that the risk was high, and that’s the only conclusion we wish to make to the audience.

**Dr M. Jacobs (Philadelphia, PA, USA):** My question goes to your conclusion that length of cardiopulmonary bypass time is a surrogate for bad outcome. I think it has been observed in now countless studies in congenital surgery, in adult surgery and whatnot that there is a statistical correlation between bypass time and mortality, and I really wondered what part of your analysis led to your conclusion that it was a surrogate. If one looked at your slide, certainly on the right upper quadrant, patients with 5 and 6 h of bypass made every last effort to salvage and that duration for those patients may very well be a surrogate, but I think if you limited your slide to the left lower quadrant with bypass times out to 120 or 150 min, there probably still is a statistical correlation, and I wondered what part of the analysis led you to conclude that this was a surrogate and not a contributing factor in mortality.

**Dr Kang:** Well, you might be absolutely right. We drew that conclusion because we didn’t want to send a message saying that we should try to reduce our bypass time for every operation by 10 min by going a little bit faster. What we thought was that those cases that had high mortality were ones that, for example, needed a second run on bypass to revise a repair, or where things were just going badly; perhaps we needed to bridge onto mechanical support, and therefore we had a prolonged bypass time. Our statistician told us that around about 220 min is when there was a more critical increase in the risk. So most operations that I can think of where things are going well should take under about 220 min to complete on bypass.
Appendix B. RACHS-1 classification system [1]

**Risk category 1**
Atrial septal defect surgery (including atrial septal defect secundum, sinus venosus atrial septal defect, patent foramen ovale closure)
Aortopexy
Patent ductus arteriosus surgery at age > 30 days
Coarctation repair at age > 30 days
Partially anomalous pulmonary venous connection surgery

**Risk category 2**
Aortic valvotomy or valvuloplasty at age > 30 days
Subaortic stenosis resection
Pulmonary valvotomy or valvuloplasty
Pulmonary valve replacement
Right ventricular infundibulectomy
Pulmonary outflow tract augmentation
Repair of coronary artery fistula
Atrial septal defect and ventricular septal defect repair
Atrial septal defect primum repair
Ventricular septal defect repair
Ventricular septal defect closure and pulmonary valvotomy or infundibular resection
Ventricular septal defect closure and pulmonary artery band removal
Repair of unspecified septal defect
Total repair of tetralogy of Fallot
Repair of total anomalous pulmonary veins at age > 30 d
Glenn shunt
Vascular ring surgery
Repair of aorta-pulmonary window
Coarctation repair at age ≤ 30 d
Repair of pulmonary artery stenosis
Transsection of pulmonary artery
Common atrium closure
Left ventricular to right atrial shunt repair

**Risk category 3**
Aortic valve replacement
Ross procedure
Left ventricular outflow tract patch
Ventriculotomy
Aortoplasty
Mitral valvotomy or valvuloplasty
Mitral valve replacement
Valvectomy of tricuspid valve
Tricuspid valvotomy or valvuloplasty
Tricuspid valve replacement
Tricuspid valve repositioning for Ebstein anomaly at age > 30 days
Repair of anomalous coronary artery without intrapulmonary tunnel
Repair of anomalous coronary artery with intrapulmonary tunnel (Takeuchi)
Closure of semilunar valve, aortic or pulmonary
Right ventricular to pulmonary artery conduit
Left ventricular to pulmonary artery conduit
Repair of double-outlet right ventricle with or without repair of right ventricular obstruction
Fontan procedure
Repair of transitional or complete atrioventricular canal with or without valve replacement
Pulmonary artery banding
Repair of tetralogy of Fallot with pulmonary atresia
Repair of cor triatriatum
Systemic to pulmonary artery shunt
Atrial switch operation
Arterial switch operation
Reimplantation of anomalous pulmonary artery
Annuloplasty
Repair of coarctation and ventricular septal defect closure
Excision of intracardiac tumor

**Risk category 4**
Aortic valvotomy or valvuloplasty at age ≤ 30 d
Konno procedure
Repair of complex anomaly (single ventricle) by ventricular septal defect enlargement
Repair of total anomalous pulmonary veins at age ≤ 30 d
Atrial septectomy
Repair of transposition, ventricular septal defect, and subpulmonary stenosis (Rastelli)
Atrial switch operation with ventricular septal defect closure
Atrial switch operation with repair of subpulmonary stenosis
Arterial switch operation with pulmonary artery band removal
Arterial switch operation with ventricular septal defect closure
Arterial switch operation with repair of subpulmonary stenosis
Repair of truncus arteriosus
Repair of hypoplastic or interrupted arch without ventricular septal defect closure
Repair of hypoplastic or interrupted aortic arch with ventricular septal defect closure
Transverse arch graft
Unifocalisation for tetralogy of Fallot and pulmonary atresia
Double switch

**Risk category 5**
Tricuspid valve repositioning for neonatal Ebstein anomaly at age ≤ 30 d
Repair of truncus arteriosus and interrupted arch

**Risk category 6**
Stage 1 repair of hypoplastic left heart syndrome (Norwood operation)
Stage 1 repair of nonhypoplastic left heart syndrome conditions
Damus–Kaye–Stansel procedure
### Appendix C. American Society of Anesthesiology (ASA) Physical Status Classification scheme

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>A normal healthy patient</td>
</tr>
<tr>
<td>P2</td>
<td>A patient with mild systemic disease</td>
</tr>
<tr>
<td>P3</td>
<td>A patient with severe systemic disease</td>
</tr>
<tr>
<td>P4</td>
<td>A patient with severe systemic disease that is a constant threat to life</td>
</tr>
<tr>
<td>P5</td>
<td>A moribund patient who is not expected to survive without the operation</td>
</tr>
<tr>
<td>P6</td>
<td>A declared brain-dead patient whose organs are being removed for donor purposes</td>
</tr>
</tbody>
</table>

Source: [http://www.asahq.org/clinical/physicalstatus.htm](http://www.asahq.org/clinical/physicalstatus.htm)