Factors Associated With Mortality and Reoperation in 377 Children With Total Anomalous Pulmonary Venous Connection

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Background—We sought to determine era-specific changes in the incidence of mortality and reoperation in children with total anomalous pulmonary venous connection.

Methods and Results—We reviewed the records of 377 children presenting from 1946 to 2005 with total anomalous pulmonary venous connection. Multivariable parametric regression models determined the incidence and risk factors for death and reoperation after repair. Pulmonary venous connection was supracardiac in 44%, infracardiac in 26%, cardiac in 21%, and mixed in 9%. Pulmonary venous obstruction was present in 48% at presentation, most frequently with infracardiac connection type ($P<0.001$). In total, 327 patients were repaired (median age, 1.7 months). Overall survival from repair was 65% at 14 years, with a current survival of 97%. Significant ($P<0.01$) incremental risk factors for postrepair death were cardiac connection type, earlier operation year, younger age at repair, use of epinephrine postoperatively, and postoperative pulmonary venous obstruction. More recent operation year was associated with younger age at repair ($P<0.001$), decreased use of deep hypothermic circulatory arrest ($P<0.001$), and use of specific drugs postoperatively ($P<0.001$). Risk-adjusted estimated 1-year survival for a patient repaired at birth with unfavorable morphology in 2005 is 37% (95% CI, 8 to 80) compared with 96% (95% CI, 91 to 99) for a patient with favorable morphology repaired at 1 year of age. Freedom from reoperation was 82% at 11 years after repair, with increased risk associated with mixed connection type ($P=0.04$) and postoperative pulmonary venous obstruction ($P<0.001$).

Conclusions—Mortality after total anomalous pulmonary venous connection repair has decreased but remains highest in young patients and in those with cardiac connection type or pulmonary venous obstruction. Unfavorable anatomic characteristics remain important determinants of postrepair survival despite improved perioperative care. (Circulation. 2007;115:1591-1598.)

Key Words: congenital defects • pulmonary veins • risk factors • surgery • survival

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital heart defect, making up ≈1% to 3% of all cardiovascular abnormalities.1 Controversy exists regarding those factors affecting short- and long-term outcomes. Recent reports2–4 have suggested that neutralization of patient characteristics such as younger age, morphological type, and preoperative pulmonary venous obstruction (PVO) underlies the improved outcomes over time after repair of TAPVC.

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We sought to determine, using all subjects presenting to a single institution that circumscribed a broad historical period, whether the benefit conferred by the contemporary era was related to identifiable changes in management over time, neutralization (ie, previously described risk factors becoming noninfluential regarding a specific outcome) of potential risk factors, or inherent changes in the subjects studied.

Methods

Patients

The present study was approved by the institutional Research Ethics Board. All patients presenting with a diagnosis of TAPVC at the Hospital for Sick Children from 1946 to 2005 were identified from computerized databases from the Divisions of Cardiology and Cardiovacular Surgery. Those with minor associated defects such as atrial septal defect and/or a patent ductus arteriosus were included. However, patients with important concomitant cardiac defects,
including single ventricle, atrial isomerism, transposition of the great vessels, and/or hypoplastic left ventricle, were excluded, leaving a final number of 377 study subjects. Demographic and morphological characteristics at initial presentation are shown in Table 1. Data were abstracted by review of clinical records and diagnostic reports obtained at the time of initial admission, before any intervention, and at the last follow-up available. Median follow-up time was 5.2 years (minimum, 11 days; maximum 21.3 years) from repair, with 207 having >1 year of follow-up, and was available for 218 survivors.

Echocardiography/Catheterization

Patient records were examined for both echocardiographic and catheterization findings at presentation and before and after major events such as repair and reoperation. When both echocardiogram and catheterization were performed in a given period, echocardiographic values were used. All echocardiographic and catheterization values, including semiquantitative grades such as those for valve regurgitation, were extracted from diagnostic reports. PVO was considered present if Doppler examination measured a pulmonary venous flow velocity >2 m/s.

Data Analysis

Data are given as frequency, median with minimum and maximum value, or mean±SD as appropriate, with the number of nonmissing values indicated. Informative imputation, when possible, was used to determine missing patient variables such as age or weight from either nomograms or redundant information available in the medical record. Mean imputation was used otherwise, with missing value flags created and forced into all models in which the imputed variable was used. The following statistical techniques were used. First, multivariable factors associated with the binary response, PVO at presentation (n=111), were sought through logistic regression. This model included patient gender, age, and weight at admission; pulmonary venous connection type; and associated cardiac defects. Second, multivariable factors associated with more recent year of repair (n=327), considered a continuous variable, were sought through linear regression. This model included cardiopulmonary bypass time; perfusion strategy used (as dichotomous indicator variables); patient gender, age, and weight; inotropes used (as dichotomous variables); and pulmonary venous connection type. Third, mortality from admission (n=377), mortality after repair (n=327), and reoperation after repair (n=327) were modeled as time-dependent events by use of both Kaplan-Meier estimates and parametric methods, with the association with risk factors being explored in multivariable parametric analysis.6,7 Potential explanatory factors included patient factors (gender, age, weight), morphological factors (pulmonary venous connection type and associated cardiac defects), and management factors (cardiopulmonary bypass time, perfusion strategy, additional procedures at the time of repair, inotrope use). Briefly, parametric probability estimates of freedom from prerepair and postrepair mortality and reoperation were generated from models based on the decomposition of the hazard function into multiple, overlapping phases of risk (available for use with the SAS system at http://www.clevelandclinic.org/heartcenter/hazard). The HAZARD procedure uses the method of maximum likelihood to aid in resolving up to 3 phases of the distribution of times until an event (early decreasing or peaking hazard, constant hazard, and late increasing hazard). Although 3 phases are potentially present for any time-related event, model validity does not require that separate scaling parameters be specified for all 3 phases because, realistically, many events have only 1 or 2 discrete phases of risk.8 For model building and variable selection, with a probability value criterion for retention of variables in the model of 0.05, we used bootstrap aggregation (bagging) from automated analysis of 1000 bootstrap data sets. Variables appearing in ≥50% of the models were retained as risk factors (median rule). Parameter estimates for risk factors are expressed as regression coefficients and their standard errors, in part because transformation of these to equivalent hazard ratios for continuous variables and interactions

<table>
<thead>
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<th>Value</th>
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<td>PDA</td>
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Values are expressed as n (%) unless otherwise indicated. ASD indicates atrial septal defect; VSD, ventricular septal defect; and PDA, patent ductus. n=377.

*Lesions definitively identified by preoperative echocardiogram or cardiac catheterization. Other defects included anatomic vice (n=11), pulmonary vein atresia (n=3), pulmonary valve stenosis (n=3), aberrant subclavian artery (n=1), and calcified ventricles (n=1).

is a less interpretable expression and in part because the model is one of nonproportional hazards in which hazard ratios change across time. In addition, trends in the use of different management strategies over successive decades were sought in Mantel-
Operative intervention was highly correlated with more recent birth year (OR, 1.1/y; P<0.001).

**Infracardiac**

Of the 98 infracardiac TAPVC patients, 78 (80%) underwent TAPVC repair. Of these patients, the type of anastomosis was documented in 58 operations. The most common procedure (82%; n=47) was a side-to-side anastomosis between the pulmonary venous confluence and the descending vertical vein (ie, ligation of the descending vertical vein without concomitant vertical vein division). End-to-side anastomosis between the pulmonary venous confluence and the descending vertical vein (ie, with concomitant division, incision, and incorporation of the vertical vein in the suture line as described by Phillips et al9) was used in 10 patients (18%).

**Supracardiac**

Seventy-one repairs (91%) were performed in the cardiac TAPVC patients, with the type of repair documented in 41 of those surgeries. Of the infracardiac repairs, 36 (88%) involved unroofing of the coronary sinus, followed by baffle connection of the coronary sinus to the atrial septal defect, based on the technique described by Malm.9 Posterior displacement of the interatrial septum as described by Hiramatsu and colleagues10 was used in 3 repairs (7%).

**Mixed**

Thirty-two of the 35 patients (91%) with mixed TAPVC underwent surgical repair, although the type of procedure varied with the location of the anomalous connection. For patients with a single isolated anomalous connecting vein, no correction of the anomalous connection was undertaken in 44%.

**Mortality**

The crude mortality rate in the study subjects was 40%, with 152 deaths. Of these patients, 49 (32%) died without any surgical intervention, and the remaining 103 died after TAPVC repair (Figure 1). Cause of death among the nonoperated patients was cardiac in 31 (6 of whom died awaiting operation), sepsis in 3, and unknown in 15 patients.

**Results**

**Pulmonary Venous Anatomy**

Pulmonary venous connection was classified as supracardiac in 44%, infracardiac in 26%, cardiac in 21%, or mixed-type drainage in 9% of patients (Table 1).

PVO at presentation was found in 53 patients with infracardiac TAPVC, 8 patients with mixed TAPVC, 42 patients with supracardiac TAPVC, and 8 patients with cardiac TAPVC. PVO at presentation was significantly associated with infracardiac anatomic type (odds ratio [OR], 6.1; 95% CI, 2.1 to 17.5; P<0.001).

**Initial Surgical Procedures**

**Overall**

A total of 327 patients (87%) underwent TAPVC repair at a median age of 1.6 months (minimum, birth; maximum, 14 years) and at a median weight of 3.6 kg (minimum, 1.8 kg; maximum, 39.3 kg) (Table 2). Of the remaining patients, 49 died without any surgical intervention, and 1 patient was lost to follow-up. Initial surgery at or beyond 5 years of age was undertaken in 17 patients, 3 of whom subsequently died.
1 year. Incremental risk factors for death after repair included earlier year of repair (P<0.001), younger age at repair (P<0.001), presence of postoperative PVO (P<0.001), use of epinephrine postoperatively (P=0.002), and cardiac connection type (P=0.007; Table 3).

The favorable influence of more recent year of repair on survival after repair is shown in Figure 2a and b. Figure 2a represents 3 specific solutions to the multivariable equation for death after repair and demonstrates that the benefit conferred by more recent year of repair occurs within the immediate postoperative period. However, Figure 2b shows the marked disparity in risk-adjusted 1-year survival estimates between 2 patients undergoing repair in the current era based solely on differences in their anatomic and demographic characteristics. Thus, unfavorable anatomic characteristics have not been completely neutralized as important determinants of postrepair survival despite improvements in perioperative care.

Figure 3 is a risk-adjusted nomogram wherein survival is expressed as a function of increasing age at complete repair. The nonlinear and rapid increase in survival with repair beyond the neonatal period is a proxy for nonemergent repair for patients presenting without PVO.

**Factors Associated With More Recent Era of Operation**

Because the more recent era was significantly associated with increased survival after repair and because the present study circumscribed a broad historical period, we further investigated whether the improvement was related to identifiable changes in management over time or whether it could be explained by changes in the study subjects treated at our institution.

Deep hypothermic circulatory arrest (DHCA) without cerebral perfusion was used in 153 patients (82%), whereas continuous hypothermic low-flow perfusion was used in 32 (17%). Perfusion strategies varied over time, with uninterrupted DHCA used less frequently with subsequent repair decades (χ², P<0.001). Death after repair was less common for patients without DHCA (3%) compared with patients who had DHCA (26%; P=0.002).

Linear regression analysis indicated that more recent year of repair (as a continuous interval response variable) was associated with younger age at surgery (P<0.001), decreased use of DHCA (P<0.001), infracardiac-type TAPVC connection (P=0.04), and the use of the following specific drugs postoperatively: milrinone (P<0.001), epinephrine (P<0.001), and nitroprusside (P=0.04). An important point is that improved postrepair survival with successive repair decades was found despite the increase in less favorable
morphological connection (e.g., infracardiac connection) and the decrease in age at repair that also characterized the more recent era.

Reoperation
Thirty children (9%) underwent reoperation after repair: 24 children had 1, 5 children had 2, and 1 child had 3 reoperations. Initial reoperation was performed at a median age of 4 months (range, 7 days to 11 years) and at a median interval of 3.3 months (range, 1 day to 11.3 years) from initial repair. Indication for reoperation was PVO in 19 patients, operative revision of anatomic defects without documented PVO in 4, closure of residual atrial septal defect in 1 patient, and unknown in 6 patients. Reoperation was characterized by an early hazard phase with no constant or late phase, with 88% and 82% freedom from reoperation at 1 and 11 year(s) after initial repair, respectively. Incremental risk factors associated with reoperation included younger age at repair \( (P < 0.001) \) and mixed anatomic type \( (P = 0.04) \). There was a trend toward an increased risk of reoperation in patients presenting with PVO and in those undergoing more recent repair, but statistical significance was not reached (Table 3).

Follow-Up Data After Repair
Most recent clinical follow-up (obtained at cardiology clinic visit) after repair, available in 157 survivors (70%), occurred at a median interval of 8.4 years (minimum, 11 days; maximum, 21.3 years). Cardiovascular symptoms were present in 17 (11%), and 10 patients (6%) reported other noncardiac symptoms. Only 5 patients reported important activity restrictions or limitations.

Discussion
We have determined outcomes in a large single-institution series of children with TAPVC. We described era-dependent

![Risk-adjusted survival from repair improved significantly with increasing year of operative repair, indicating a strong era effect.](image)

![Risk-adjusted nomograms show 1-year survival after repair expressed as a function of increasing year of operation for 2 different patients. The top line (A) shows the multivariable solution for a patient with favorable anatomic characteristics (noncardiac connection without PVO) undergoing repair at 1 year of age; the bottom line (B) shows the solution for a patient with unfavorable characteristics (cardiac connection with PVO) undergoing operation at birth. The nomograms show that more recent era has improved survival among all patients, especially within the last decade. However, unfavorable anatomic characteristics have not been neutralized as important determinants of postrepair survival despite improvements in perioperative care. Solid lines are continuous parametric estimates enclosed by dashed 95% confidence limits. Numbers in parentheses represent parametric estimates of median survival at 1 year after repair in 2005.](image)
changes and have shown, contrary to previous reports, that survival after repair remains highly dependent on patient factors even in the modern era. Thus, risk factors such as emergent repair in those with obstructed pulmonary venous drainage (surrogated by younger age at repair) and unfavorable connection type have not been neutralized as important determinants of postrepair survival despite improvements in perioperative care.

Mortality
Overall survival at 14 years from admission in the present study was 56%, and survival at a similar interval from repair was 65%. Our results improved significantly over time, with the majority of deaths occurring early in our experience and only 2 deaths within the last 5 years. This agrees with previously published series in which operative mortality was nearly 50% for infants repaired before 1 year of age in the 1970s but decreased to 5% in the contemporary era. Higher mortality in our series is related to the broad time period circumscribed by our cohort (>50 years) and the predominance of less favorable infracardiac connection type at all time points.

Influence of Era
A number of changes in practice have occurred over the past 4 decades that may account for decreased mortality, all of which cannot be delineated in a single analysis. We have, however, identified both patient-specific factors and intraoperative and postoperative management factors that are associated with the benefit of the more recent era. First, there was a shift in perfusion strategy away from DHCA in the more contemporary era that was significantly associated with improved survival after repair. Bogen and colleagues similarly found decreased operative mortality with hypothermic (18°C to 22°C) continuous perfusion with cardioplegic arrest compared with DHCA. However, others, including Lincoln et al and Kirshbom et al, have not found the use of DHCA to be a risk factor for mortality despite a favorable era effect and a coincident change in perfusion strategy.

We also found that use of specific drugs in the immediate postoperative period, including milrinone, epinephrine, and nitroprusside, increased over time. Low cardiac output syndrome was reported as the main cause of early postoperative death in older series, highlighting the underlying rationale for increased use of inotropic agents.

It is important to note that improved survival with successive birth cohorts was found despite the increase in less favorable substrate (eg, infracardiac connection type) and the decrease in age at repair that also characterized the more recent era.

Influence of Patient Characteristics
Infracardiac and cardiac connection type and PVO at presentation in the present study were significantly associated with an increased risk of death after adjustment for other factors. Infracardiac connection type was correlated with preoperative PVO, potentially explaining the comparative disadvantage of this subgroup, but PVO at presentation was not a defining characteristic of cardiac connection type. Although our multivariable analysis contained a comprehensive variable set, missing values, specifically regarding perioperative variables, and limitations on event-rate-to-variable ratio certainly could have influenced our findings. It also is possible that the cumulative effect of many small differences between groups (ie, those that are not statistically significant) could have contributed some degree of bias. Interestingly, children presenting with mixed anatomic type were not at increased risk of death after repair and no identifiable predilection to the development of postoperative PVO, in contradistinction to previous reports by Kirshbom et al and Delius and colleagues. Potential reasons for the lower prevalence of postoperative PVO in patients with mixed connection type in our series could be related to a lower prevalence of infracardiac connection within the “mixed” phenotype, differences in surgical repair technique, or incorrect assignment of connection type.

The impact of connection type on outcomes has been debated partly because results are confounded by the multicollinearity between preoperative PVO and certain connection types (eg, infracardiac and supracardiac) that often precludes discrimination of both as potential risk factors within a single multivariable model. Bando and colleagues made the controversial statement that both preoperative PVO and anatomic type had been neutralized as potential risk factors beyond calendar year 1991. Hyde et al similarly reported that connection type was not related to outcome.

Younger age at repair also was identified in the present study as an important risk factor for mortality after adjustment for era. It is likely, however, that younger age at repair is a proxy for emergent repair in neonates presenting with obstructed pulmonary venous drainage because these factors are highly correlated. Suppression of “young age at repair” in our overall model for mortality uncovered PVO at presentation as a risk factor. The “effect” of young age is also relative given that the median age at repair decreased monotonically over time from 2 years in the first decade to 18 days in the current decade.

Our results suggest that patient characteristics, including connection type, PVO, and young age, are still important risk factors for adverse outcomes even after adjustment for era. We therefore disagree with the statement by Lupinetti et al that “it is unlikely that identification of preoperative risk factors can be used to predict the results after operative repair.” Although results have improved over time concomitant with novel surgical techniques and an evolution in perioperative strategies specific for pulmonary hypertension, mortality continues to be higher in young patients with unfavorable anatomy.

Short-term palliation strategies such as balloon angioplasty or atrial septostomy, described by Ramakrishnan and Kothari and Bu’Lock and colleagues, to forestall emergent operation in the youngest patients may provide alternative solutions.
Influence of Repair Type

We did not find that the type of operative procedure influenced either the risk of death or reoperation. Certainly, the study was not powered to discriminate among every possible reparative procedure because some were present in few patients. Additionally, the repair type could be considered more appropriately as an outcome rather than a potential explanatory factor because it is determined by more influential patient characteristics such as connection type.

Reoperation

The overall incidence of reoperation at 11 years after repair (9%) was not negligible in our series and was not associated with repair type. Parallel observations have been made in previous reports,\textsuperscript{1,20} with reoperation rates ranging from 9% to 13%. Our finding that later birth cohort increased the reoperative risk is consistent with the younger age at repair (and hence longer at-risk period) for those patients treated in more recent eras. It also is probable that reoperation rates have increased commensurate with the increase in the number and level of complex repairs undertaken in the recent era and a concomitant decrease in the threshold for reintervention. Sutureless techniques for recurrent PVO as described by our group\textsuperscript{23,24} and others\textsuperscript{25} have demonstrated superior outcomes over conventional surgical (eg, pulmonary venous endarterectomy or patch enlargement) and catheter-based methods.\textsuperscript{25,26} A recent study from our institution\textsuperscript{24} reported acceptable freedom from death and reoperation with extension of the sutureless technique to patients with primary pulmonary venous anomalies. Further studies are needed to determine the utility of novel techniques in the heterogeneous spectrum of TAPVC.

Conclusions

We have determined outcomes and associated risk factors in a large single-institution series of children with TAPVC. Mortality after repair of TAPVC has decreased in the modern era, but specific patient demographics and anatomic characteristics remain important determinants of postrepair survival despite improved perioperative care.

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Disclosures

None.

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**CLINICAL PERSPECTIVE**

Total anomalous pulmonary venous connection is a congenital cardiovascular defect characterized by failure of the pulmonary venous confluence to be absorbed into the posterior wall of the left atrium. As a result, a variety of anomalous pathways for drainage of the confluence develop. The type of anomalous connection can influence the propensity for the pulmonary venous drainage to be obstructed and can impose technical challenges in achieving anastomosis to the left atrium during surgical repair. We show that despite overall improvements in the more recent era in surgical outcomes, the presence of cardiac connection type, usually characterized by anomalous drainage to the coronary sinus, was independently associated with increased mortality after repair. We also note that mixed connection type, with the left and right pulmonary veins draining by different anomalous pathways, was independently associated with an increased risk of reoperation. Pulmonary venous obstruction remains a challenge, with obstruction present postoperatively associated with increased mortality and obstruction present preoperatively associated with an increased risk of reoperation. Thus, specific anomalous connection type and the presence of pulmonary venous obstruction are important risk factors yet to be completely addressed with better perioperative management and modern surgical techniques. Trends were noted in the more recent era toward earlier repair, avoidance of deep hypothermic circulatory arrest, and increased prevalence of postoperative use of inotropic and vasoactive agents, concomitant with reduced mortality and risk of reoperation after repair. The challenges of managing total anomalous pulmonary venous connection with specific connection types or pulmonary venous obstruction remain of concern.
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