Repair of "Simple" Total Anomalous Pulmonary Venous Connection: A Review From the Pediatric Cardiac Care Consortium

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Background. **Outcomes for repair of total anomalous pulmonary venous connection (TAPVC) from individual institutions suggest a significant improvement in mortality over the past several decades. The purpose of this study is to review the outcomes after repair of TAPVC from a large multiinstitutional registry.**

Methods. **A retrospective review of the multiinstitutional database, the Pediatric Cardiac Care Consortium (PCCC), was used to identify patients with the diagnosis of TAPVC who underwent complete correction between 1982 and 2007. Data reviewed included age, decade of primary operation, anatomic type, presentation, and in-hospital mortality.**

Results. **Of the 118,084 surgical procedures submitted to the PCCC, 2,191 (1.9%) consisted of primary surgical correction of TAPVC. Sixty-one percent of the cohort was male, with 6.8% reported as premature. Overall in-hospital surgical mortality for simple TAPVC was 13%. Mortality was 20% from 1982 to 1989, 16% from 1990 to 1999, and 8% from 2000 to 2007. Obstruction to the anomalous pulmonary venous connection occurred in 29%, with a mortality of 26%.**

Conclusions. **Surgical outcomes from repair of congenital cardiac anomalies have significantly improved over the past several decades. Multiinstitutional large databases are needed to confirm results published from single-institution experiences. Although improvements in surgical repair of TAPVC have occurred over the past three decades, specific subtypes still experience significant mortality.**

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Outcomes after operative correction of congenital cardiac defects have generally improved over the past several decades. Total anomalous pulmonary venous connection (TAPVC) is a relatively rare congenital defect with reported outcomes paralleling less technically challenging procedures. Mortality is high when primary repair is not undertaken within the first year of life [1]. Preoperative patient characteristics have been considered to have a significant impact on overall surgical results [2]. These risk factors include patient age at the time of repair, morphologic connection, presence of pulmonary venous obstruction, and associated cardiac defects. Recent reviews have reported the neutralization of these risk factors over the past several decades [3, 4]. The majority of these data have been produced by single-center retrospective experiences [4, 5]. Using this multiinstitutional dataset, we sought to determine changes in mortality over the past several decades, as well as confirm which risk factors continue to have a significant impact on mortality after surgical correction.

Patients and Methods

Data used in this analysis were covered by the Data Use Agreement of the Pediatric Cardiac Care Consortium (PCCC) provided from individual member institutions. The study was approved by the Institutional Review Board at the University of Minnesota with an exception for obtaining individual patient consent.

Data Collection

A retrospective review involving the PCCC was used to identify patients admitted with the diagnosis of total anomalous pulmonary venous connection (TAPVC) who underwent primary repair. All patients assigned the diagnosis code for TAPVC and the procedure code for correction of TAPVC between 1982 and 2007 were reviewed. Data points were extracted from a dataset submitted by the member institution. Assignments of diagnosis and procedure codes were accomplished in a centralized fashion by the registry. The procedure and diagnosis codes were inclusive of additional anatomic and physiologic characteristics of the patients. Accuracy of the submitted data was confirmed by annual reviews of the individual institution's medical records confirming discharge and death information. Cases were defined as "simple" TAPVC if there were no associated cardiac anomalies (excluding patent ductus arteriosus and atrial septal defect). The overall analysis was conducted on all patients who underwent repair at less than 18 years of life. Patients were excluded from analysis if data points were missing (surgical weight, n = 12; anatomic connection, n = 39) or if they had undergone a previous surgical procedure (n = 322).

Mortality was defined as all in-hospital deaths after the primary repair of TAPVC. The periods studied for statistical

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analysis were 1982 to 1989, 1990 to 1999, and 2000 to 2007. Obstruction of pulmonary venous connection was determined by review of clinical and diagnostic information provided by the submitting institution.

Frequencies and proportions were computed for the overall group and by decade, age group, reported obstruction, and anatomic type. Categorical variables were defined by the investigators before the analysis. Age groups were defined as neonates (<30 days), infants (30–365 days), and children (>365 days). Timing of operation was defined as emergent (day 0 or 1 of life), urgent (day 2 of life), and elective (after day 2 of life). Low surgical weight was defined as less than 2.5 kg. Pearson χ^2 tests and Fisher exact tests were used to compare categorical variables. Multivariable logistic regression with generalized estimating equations was used to compute marginal effects while accounting for potential correlation within hospital. The binary outcome was defined as within-hospital mortality. An exchangeable correlation structure was specified.

No adjustments were made for multiple comparisons. All statistical analyses were completed using the R Project for Statistical Computing, version 2.14.1 [6].

Results

A total of 2,191 patients underwent primary surgical repair of TAPVC between 1982 and 2007. When evaluating patients with simple TAPVC (77%, n = 1,693), 91% (n = 1,543) were younger than 18 years, whereas 86% (n = 1,454) were younger than 1 year of life at the time of operation. This analysis will focus on those younger than 18 years of age at primary repair. Patient characteristics stratified by decade of primary repair are presented in Table 1. Sixty-two percent of this population was male. Approximately 5% of patients underwent repair with a weight less than 2.5 kg, with 7% (n = 101) considered premature (gestational age < 37 weeks). Fifty-nine percent of patients (n = 801) underwent primary repair during the neonatal period, with the frequency significantly increasing between each decade.

The overall unadjusted in-hospital mortality for patients with simple TAPVC was 13%. There was a significant decrease in mortality within each decade studied (Table 2). When comparing all patients who underwent repair as neonates and infants, we found a significantly lower mortality in the latter group (19.1% versus 4.9%; $p < 0.001$). There was a significant decrease in mortality when comparing 1990 to 1999 with 2000 to 2007 for the infant group (6.3% versus 1.7%, respectively; $p = 0.021$) and the neonatal group (23% versus 11.7%, respectively; $p < 0.001$).

The frequencies of anatomic connections for patients with simple TAPVC are presented in Table 1, with supracardiac connection occurring most often. A total of 401 patients (29%) presented with obstruction of the pulmonary venous connection. Mortality varied significantly when stratified to the type of pulmonary venous connection (Table 2). The overall mortality for the supracardiac group was 10.5%. Of this group, 27% presented with obstruction before the primary repair. The mortality associated with obstruction to the supracardiac connection was 22%, decreasing to 6% when obstruction was not present. Of the 273 patients with a cardiac connection, mortality was 3.7%. When obstruction was not present, mortality was 3%, but it increased to 14% with obstruction. One hundred eighty patients with an infracardiac connection pre-

Table 1. Patient Characteristics

Variable	Total	1982–1989	1990–1999	2000–2007	*p* Value
	1,366	186 (13.6%)	605 (44.3%)	575 (42.1%)	
Sex					
Female	521 (38.1%)	78 (41.9%)	228 (37.7%)	215 (37.4%)	0.516
Male	845 (61.9%)	108 (58.1%)	377 (62.3%)	360 (62.6%)	
Age group					
Neonates (<30 d)	801 (58.6%)	89 (47.8%)	343 (56.7%)	369 (64.2%)	<0.001
Infants (30–365 d)	493 (36.1%)	88 (47.3%)	224 (37.0%)	181 (31.5%)	
Children (>365 d)	72 (5.3%)	9 (4.8%)	38 (6.3%)	25 (4.3%)	
Low surgical weight					
No (≥2.5 kg)	1,299 (95.1%)	178 (95.7%)	573 (94.7%)	548 (95.3%)	0.822
Yes (<2.5 kg)	67 (4.9%)	8 (4.3%)	32 (5.3%)	27 (4.7%)	
Anatomic subtype					
Supracardiac	626 (45.8%)	73 (39.2%)	283 (46.8%)	270 (47.0%)	0.043
Cardiac	273 (20.0%)	35 (18.8%)	125 (20.7%)	113 (19.7%)	
Infracardiac	312 (22.8%)	43 (23.1%)	136 (22.5%)	133 (23.1%)	
Mixed	155 (11.3%)	35 (18.8%)	61 (10.1%)	59 (10.3%)	
Reported obstruction					
Yes	401 (29.4%)	57 (30.6%)	181 (29.9%)	163 (28.3%)	0.770
No	965 (70.6%)	129 (69.4%)	424 (70.1%)	412 (71.7%)	
Timing of operation					
Emergent (<2 d)	206 (15.1%)	20 (10.8%)	86 (14.2%)	100 (17.4%)	0.077
Urgent (2d)	93 (6.8%)	8 (4.3%)	46 (7.6%)	39 (6.8%)	
Elective (>2 d)	1067 (78.1%)	158 (84.9%)	473 (78.2%)	436 (75.8%)	

Table 2. Mortality

Variable	Total	1982–1989	1990–1999	2000–2007	*p* Value
	178/1,366 (13.0%)	38/186 (20.4%)	94/605 (15.5%)	46/575 (8.0%)	<0.001
Sex					
Female	58/521 (11.1%)	14/78 (17.9%)	28/228 (12.3%)	16/215 (7.4%)	0.032
Male	120/845 (14.2%)	24/108 (22.2%)	66/377 (17.5%)	30/360 (8.3%)	<0.001
Age group					
Neonates (<30 d)	153/801 (19.1%)	31/89 (34.8%)	79/343 (23.0%)	43/369 (11.7%)	<0.001
Infants (30–365 d)	24/493 (4.9%)	7/88 (8.0%)	14/224 (6.3%)	3/181 (1.7%)	0.021
Children (>365 d)	1/72 (1.4%)	0/9 (0.0%)	1/38 (2.6%)	0/25 (0.0%)	
Low surgical weight					
No (≥2.5 kg)	156/1299 (12.0%)	33/178 (18.5%)	82/573 (14.3%)	41/548 (7.5%)	<0.001
Yes (<2.5 kg)	22/67 (32.8%)	5/8 (62.5%)	12/32 (37.5%)	5/27 (18.5%)	0.046
Anatomic subtype					
Supracardiac	66/626 (10.5%)	14/73 (19.2%)	35/283 (12.4%)	17/270 (6.3%)	0.003
Cardiac	10/273 (3.7%)	0/35 (0.0%)	8/125 (6.4%)	2/113 (1.8%)	0.110
Infracardiac	72/312 (23.1%)	12/43 (27.9%)	40/136 (29.4%)	20/133 (15.0%)	0.013
Mixed	30/155 (19.4%)	12/35 (34.3%)	11/61 (18.0%)	7/59 (11.9%)	0.032
Reported obstruction					
Yes	103/401 (25.7%)	18/57 (31.6%)	54/181 (29.8%)	31/163 (19.0%)	0.038
No	75/965 (7.8%)	20/129 (15.5%)	40/424 (9.4%)	15/412 (3.6%)	<0.001
Timing of surgery					
Emergent (<2 d)	60/206 (29.1%)	13/20 (65.0%)	31/86 (36.0%)	16/100 (16.0%)	<0.001
Urgent (2 d)	19/93 (20.4%)	3/8 (37.5%)	9/46 (19.6%)	7/39 (17.9%)	0.470
Elective (>2 d)	99/1067 (9.3%)	22/158 (13.9%)	54/473 (11.4%)	23/436 (5.3%)	<0.001

sented with obstruction (57.6%), with a mortality of 28%. Mortality of patients who did not present with obstruction was 16%. The risk of obstruction at presentation was statistically greater in the infracardiac group compared with the other anatomic types ($p = 0.0001$). The mixed type (n = 155) had an overall mortality of 19%. The majority of these patients presented without signs of obstruction and had a mortality of 15%. The patients who presented with obstruction had a mortality of 32%. Mortality for each anatomic group, with the exception of the cardiac connection, improved over the periods studied.

For the cohort of patients who presented with simple TAPVC, 15% of patients underwent emergent repair, 7% urgent repair, and 78% elective repair. The frequency of emergent repairs increased from 10% of all cases during the first period (1982–1989) to 17% during the last period (2000–2007) (Table 1). Mortality also changed over the period of data collection based on the time to operative repair, significantly decreasing from the earliest to most recent decade for both the emergent (65% versus 16%; $p < 0.001$) and elective periods (13.9% versus 5.3%; $p < 0.001$) (Table 2).

The time to operative repair for the entire TAPVC cohort varied significantly ($p < 0.0001$) based on the anatomic connection of the anomalous pulmonary venous connection (infracardiac median = 4 days, mixed median = 30 days, supracardiac median = 32 days, and cardiac median = 46 days). The infracardiac type had the highest percentage of emergent and urgent repairs when compared with the other anatomic types ($p < 0.0001$). The infracardiac group was stratified into obstruction versus nonobstruction for the entire collection period. For this cohort, 21% of patients without obstruction and 37% with obstruction underwent emergent repair. During the last period (2000–2007), when the percentage of emergent primary repairs increased significantly compared with the earlier 2 periods, the percentage of patients with obstructed infracardiac connection who underwent emergent repair was 45% (n = 45), compared with 25% (n = 24) of patients without obstruction. The mortality associated with emergent repair of nonobstructed infracardiac anatomy was 42%, but decreased to 21% when the operation was undertaken on an elective basis ($p = 0.0631$).

To identify predictors of mortality, a multivariable regression analysis was performed using variables listed in Table 1. Weight at operative intervention, decade the procedure was performed, neonate status, reported obstruction, and the need for emergent repair were all considered positive predictors for mortality in patients with TAPVC (Table 3).

For the entire cohort (n = 2,248), 148 patients (7%) had a diagnosis of a major "2-ventricle" cardiac anomaly. The most commonly reported anomaly was a ventricular septal defect, with double-outlet right ventricle, tetralogy of Fallot, or coarctation of the aorta occurring less commonly (Table 4). Eighty-four percent of these patients with 2-ventricle anomalies underwent correction of the associated defect at the same operation as the primary repair of TAPVC. Four hundred seventy patients presented with an associated cardiac anomaly that would have precluded the creation of 2-ventricle physiology. Of these patients, 295 (63%) were classified as having heterotaxy syndrome.

Table 3. Association of Characteristics with Mortality

Variable	Odds Ratio	95% Confidence Interval	*p* Value
Sex			
Females versus males	0.87	0.63–1.20	0.400
Surgical weight			
< 2.5 kg versus ≥ 2.5 kg	2.88	1.60–5.20	<0.001
Decade			
1982–1989 versus 2000–2007	3.71	1.88–7.32	<0.001
1990–1999 versus 2000–2007	2.19	1.48–3.24	<0.001
1982–1989 versus 1990–1999	1.51	0.83–2.74	0.175
Age groups			
Infants versus neonates	0.33	0.19–0.56	<0.001
Children versus neonates	0.11	0.01–0.86	0.036
Children versus infants	0.34	0.03–3.29	0.351
Reported Obstruction			
Yes versus no	2.51	1.81–3.49	<0.001
Anatomic subtype			
Cardiac versus supracardiac	0.62	0.31–1.22	0.165
Infracardiac versus supracardiac	1.41	0.99–2.02	0.059
Mixed versus supracardiac	2.45	1.54–3.92	0.000
Timing of operation			
Emergent versus elective	1.76	1.22–2.55	0.003
Urgent versus elective	1.41	0.75–2.65	0.282

Comment

Using a large multiinstitutional surgical registry, we analyzed changes in mortality and predictors of mortality in patients who underwent primary repair of TAPVC over an extended period. Several single-center retrospective studies have shown that survival in repair of TAPVC has generally improved over the past several decades [7–9]. Our goal was to confirm these findings using a large cohort of patients.

The overall mortality for patients with simple TAPVC undergoing primary repair was 13%. These results improved over time, with a significant decrease (8%) during the most recent period studied. This observation corroborates several small, single-center experiences showing a period-dependent improvement in survival [2, 5, 10, 11].

Our study identified low weight at operation, operative intervention within the first 30 days on life, and reported obstruction to the anomalous pulmonary venous return as risk factors for mortality. Operative correction during the neonatal period has been shown to improve survival for several congenital cardiac anomalies. In a recent study by Yong and colleagues [5], 112 patients who underwent primary correction of TAPVC during the neonatal period were reviewed and showed an overall in-hospital mortality of 10.4%, although there was no change in mortality over the duration of the review. Our study revealed a somewhat different pattern, with a significant improvement in survival for each decade studied. This may reflect individual institutional bias not to operate on neonates during the previous decades, with a transition to offering operative repair to complex lesions in the current era. Survival from repair of the nonneonatal population was significantly greater than that seen in the neonates, even within the most recent period. This difference is certainly multifactorial and may represent ongoing limitations in the ability to operate on the sickest neonates. Although our data analysis did not allow for identification of specific factors that accounted for these differences, recent publications have delineated several patient-specific factors that were associated with lower survival in this group [2, 12].

This study presents a larger number of patients, which defined the frequency of the anatomic presentation and the outcomes associated with this anatomy. The presence of obstruction at the time of presentation had been considered a significant risk factor for increased mortality [2]. Our data agree with this, as mortality was higher for both the overall group and within each of the anatomic connection groups. Several other recent studies have disagreed with these findings, concluding that the anatomic type and the presence of obstruction have been neutralized as a risk factor. Bando and associates [13] concluded that these anatomic characteristics have been neutralized in recent years. Others have also concluded that connection type was not related to outcomes [14]. The discrepancies between our study and the others may be due to size: these other studies evaluated approximately 100 patients per study.

The impact of the timing of operative intervention on survival was investigated. The time of operative intervention from birth was categorized into emergent, urgent, and elective, based on previously published literature. Several authors have reported that the need for emergent/urgent operative intervention was no longer a risk factor for increased in-hospital mortality [4, 15]. This is in contrast to a recent study by Karamlou and colleagues [2] that found that a younger age at time of repair was a significant risk factor for mortality. Our study revealed a similar finding in patients undergoing emergent repair; they had a significantly increased mortality compared with those who underwent procedures within 48 hours in the most recent era. The reason for this is certainly multi-

Table 4. Major Cardiac 2-Ventricle Anomalies (n = 146)

Anomaly	n (% of all patients)
VSD	61 (2.71)
DORV	14 (0.62)
TOF	13 (0.58)
Coarctation of aorta	12 (0.53)
Pulmonary stenosis	8 (0.36)
Partial AVC	5 (0.22)
Truncus arteriosus	4 (0.18)
Complete AVC	3 (0.13)
D-TGA	2 (0.09)
D-TGA + VSD	2 (0.09)
Mitral ring	2 (0.09)
Vascular ring	2 (0.09)
Bicuspid aortic valve	1 (0.04%)
Miscellaneous	17 (0.76%)

AVC = atrioventricular canal; DORV = double-outlet right ventricle; D-TGA = transposition of great arteries; TOF = tetralogy of Fallot; VSD = ventricular septal defect.

factorial and likely reflects the general overall physiologic status of the patient before operative repair, although our dataset did not allow us to investigate specific physiologic risk factors. Karamlou and associates [2] suggested that mortality was higher in patients requiring emergent intervention because the presence and severity of obstruction to the pulmonary venous return was worse than in patients not requiring emergent repair. Our overall findings are in agreement with this statement.

Limitations

Several limitations exist within this dataset. The most important is that this is a historic registry with information recorded from various sources over an extended period. Mortality was recorded only until discharge; therefore information on 30-day and longer term outcomes were not available. The determination of obstruction to the anomalous pulmonary venous return was made at the time of submission to the registry. Objective criteria may not have always been available, and this determination may have been made based on subjective statements within the narratives provided by the submitting institutions.

Conclusions

Overall, mortality for primary repair of TAPVC has decreased for the past several decades. The most significant improvement in outcomes after repair of TAPVC occurred in the neonatal population. Certain risk factors for increased mortality have persisted over time, including operative intervention in small infants, presence of obstruction at the time of repair, and repair during the neonatal period.

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References

1. Bourroughs JT, Edwards JE. Total anomalous pulmonary venous connection. Am Heart J 1960;59:913–31.
2. Karamlou T, Gurofsky R, Al Sukhni E, et al. Factors associated with mortality and reoperation in 377 children with total anomalous pulmonary venous connection. Circulation 2007;115:1591–8.
3. Nakata T, Fujimoto Y, Hirose K, et al. Functional single ventricle with extracardiac total anomalous pulmonary venous connection. Eur J Cardiothorac Surg 2009;36:49–56.
4. Kirshbom PM, Flynn TB, Clancy RR, et al. Late neurodevelopmental outcome after repair of total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 2005;129:1091–7.
5. Yong MS, d'Udekem Y, Robertson T, et al. Outcomes of surgery for simple total anomalous pulmonary venous drainage in neonates. Ann Thorac Surg 2011;91:1921–7.
6. R Development Core Team. R: a language and environment for statistical computing. Available at: http://www.R-project.org/. Accessed: August 25, 2011.
7. van de Wal HCJM, Hamilton DI, Godman MJ, Harinck E, Lacquet LK, van Oort A. Pulmonary venous obstruction following correction for total anomalous pulmonary venous drainage: a challenge. Eur J Cardiothorac Surg 1992;6:545–9.
8. Caldarone CA, Najm HK, Kadletz M, et al. Surgical management of total anomalous pulmonary venous drainage: impact of coexisting cardiac anomalies. Ann Thorac Surg 1998;66:1521–6.
9. Hancock Friesen CL, Zurakowski D, Thiagarajan RR, et al. Total anomalous pulmonary venous connection: an analysis of current management strategies in a single institution. Ann Thorac Surg 2005;79:596–606.
10. Chowdury UK, Airan B, Malhotra A, et al. Mixed total anomalous pulmonary venous connection: anatomic variations, surgical approach, techniques, and results. J Thorac Cardiovasc Surg 2008;135:106–16.
11. Kelle AM, Backer CL, Gossett JG, Kaushal S, Mavroudis C. Total anomalous pulmonary venous connection: results of surgical repair of 100 patients at a single institution. J Thorac Cardiovasc Surg 2010;139:1387–94.
12. Boger AJ, Baak R, Lee PC, Boersma E, Meijboom FJ, Witsenburg M. Early results and long-term follow-up after corrective surgery for total anomalous pulmonary venous return. Eur J Cardiothorac Surg 1999;16:296–9.
13. Bando K, Turrentine MW, Ensing GJ, et al. Surgical management of total anomalous pulmonary venous connection. Thirty-year trends. Circ 1996;94(9 suppl):II12–6.
14. Hyde JAJ, Stumper O, Barth MJ, et al. Total anomalous pulmonary venous connection: outcome of surgical correction and management of recurrent venous obstruction. Eur J Cardiothorac Surg 1999;15:735–41.
15. Serraf A, Bruniaux J, Lacour-Gayet F, et al. Obstructed total anomalous venous return. Toward neutralization of a major risk factor. J Thorac Cardiovasc Surg 1991;101:601–6.

DISCUSSION

DR JORGE SALAZAR (Jackson, MS): Thank you for the opportunity to discuss this paper. I appreciate the excellent presentation and also the opportunity to review your manuscript ahead of time.

These are very interesting results you present looking at the management and outcomes of total anomalous pulmonary venous connection since 1980, divided into 3 time periods. The specialty of thoracic surgery has advanced by its tenacity and nonacceptance of poor outcomes and as demonstrated in your paper, we see that the outcomes for TAPVC continue to improve. These improvements are based on advances in preoperative, intraoperative, and postoperative management. One cannot help but acknowledge that the results being presented still leave room for improvement for all of us, and as you discuss in your paper as well as in your presentation, incredible opportunities still exist for accomplishing better outcomes. I have 2 questions.

One, to what would you attribute primarily the advances our specialty has achieved in managing these difficult children? Second, recognizing what a challenging technical and physiologic situation that is presented to us in managing these children, how would you focus our efforts for the future to further improve these outcomes and get closer and closer to 100% success? The things that come to mind for me that might be possibilities would be technical modifications, changes in anesthetic management, perfusion strategies and circuit design, pre- and postoperative management, et cetera. Thank you very much.

DR ST. LOUIS: Thank you. For the first question, I would agree. I think that although this dataset when taken superficially, which is reviewing sort of the limited amount of information that we are able to obtain from a center without mining the data itself, doesn't give us the

opportunity to delve into it, but, as you know, several publications have looked at timing of surgery, the degree of acidosis, the bypass times, and have shown, in limited smaller number of data sets, to also improve. We are undertaking more specific assessment of each 1 of these parameters by mining the data, looking at the operative notes (over 2,000), looking at the autopsy reports in some cases, to help us better define these parameters.

DR SALAZAR: My questions are obviously rhetorical and I acknowledge that, but I appreciate your responses. I believe, as you suggest, all of these different factors are extremely important. Additionally, another opportunity to be addressed is the management of team dynamics to maximize these outcomes. I do look forward to our specialty having improved outcomes for this population.

Thank you.

DR JEFFREY JACOBS (St. Petersburg, FL): I just rose to congratulate you for an excellent presentation using data from really 1 of the largest registries of data about patients with congenital heart disease in the world. You and me and John Mayer have discussed in the past performing similar analyses for other diagnostic and procedural cohorts using these data from the Pediatric Cardiac Care Consortium, and I was wondering if you could comment a little bit about some of the plans with the consortium for the future using these data.

DR ST. LOUIS: There is no question, James Moller is the founder and father of this dataset and it has flourished over the years sort of with multiple competing datasets that are a little bit more poignant into what is needed today for quality outcomes. The PCCC is going to cease to take in patients as of December 31st. So we are presently looking at different retooling of this incredible amount of information. One of the possible mechanisms that we have come up with is linking to the STS and actually downloading and transferring these well over 300,000 pieces of information to the STS dataset so that it is available for everyone to utilize in this retrospective analysis, as well as to be able to try to crosslink those patients that are operated on with whether those patients are re-presenting in the STS. There is a lot going on and I think this is an exciting time.

DR PETER MANNING (Cincinnati, OH): When I read your abstract, the thing that really jumped out at me was the 13% mortality for unobstructed total veins. I thought that was lousy, and I was trying to look through things in the abstract and your presentation to try to understand why. One thing that struck me was that your infracardiac total veins, you had about a 50/50 split, obstructed/nonobstructed, which raises the question, how are people really defining obstruction, because in my mind, infracardiac is obstructed until proven otherwise. They almost always are, and so I wonder whether your dataset is contaminated. Another way of looking at it, which I think you tried to analyze, is the urgency of operation which should also equate with obstruction versus nonobstruction. In our practice, unobstructed total veins is an elective operation that's done at 6 to 6 weeks of age. I noticed you did a little bit of analysis, but your total percentages of the urgency only added up to about 75%, so there is something goofy with that data, too.

DR ST. LOUIS: The biggest thing I looked at was how do you define obstruction, because that formed sort of a pivot point for this study. The way it has been defined is at the institution—well, not by the institution but when data was submitted to the dataset itself over the last 25 plus years. I now code for the PCCC in working our way through this, and we really try to look for echocardiography data, but there wasn't a lot of that in the '80s or '90s, so a lot of it was what the member institution said. A lot of the data didn't come through us. You couldn't actually read some of the datasets, the echo reports that came on. So we looked for a definition of greater than 5 mm Hg on Doppler flow when we diagnosed an individual patient when it comes into the dataset.

DR MANNING: I would argue that obstructed total veins is not an echo diagnosis. It is a diagnosis made by oxygen saturations.

DR ADIL HUSAIN (San Antonio, TX): I really like these types of presentations because I think it affords us a strong piece of clinical work which can be cited in the challenging discussions we have with parents and families preoperatively. Families commonly inquire about outcomes and this comprehensive data is very helpful to share with them during that challenging time of counseling and obtaining informed consent.

My question was really along the lines of Dr Manning's, and I am wondering if you could somehow clean your data analysis by looking at a correlation between timing of operation and the diagnosis of obstruction. Did you go back and look to see if the urgency versus emergent operations were within the obstructed patients or did you have some patients that were diagnosed as being obstructed who perhaps didn't get their operation in the emergent time period? This would be a manner of cross-correlating those data points to clean the data a little bit better.

DR ST. LOUIS: I did, and I took it out because of timing, but clearly the majority of patients that underwent emergent repair were obstructed infracardiac. The thing that I found interesting was that the outcomes from whether they were obstructed infracardiac or nonobstructed infracardiac in that emergent/urgent time period didn't change.

DR CARL BACKER (Chicago, IL): Jim, that was a great presentation. It confirms a number of important points. We analyzed our results at Children's Memorial Hospital with 100 patients with total anomalous pulmonary venous connection. The factor that we found most predictive of mortality was whether or not the patient had a functionally univentricular heart. If the patient had the diagnosis of "functionally univentricular heart" with total anomalous pulmonary venous connection, the mortality rate was 50%. If the patient had a biventricular circulation with unobstructed total anomalous pulmonary venous connection the mortality rate was 5%. What I would recommend is try to tease out whether it is total anomalous pulmonary venous connection "obstruction," which as you noted is a "soft call" or "functionally univentricular heart" that was the deciding factor that really elevated the mortality. We were surprised at how high the mortality was in this group, but if you look at the literature, that is the mortality for this group. I think the only exception was the paper that was presented from Texas Children's, which had the best outcomes with heterotaxy that I have seen reported lately. They similarly showed a nice improvement in results over time. Again, this was a very nice presentation. Thanks for the opportunity to discuss.

DR JOHN EDMUND MAYER (Boston, MA): Following up on the same sort of thought process that Carl articulated, what we have here is a potential for highly linked variables and 1 variable being a surrogate for others, and I think the constellation that we all recognize that is the highest risk is heterotaxy, obstructed veins, pulmonary atresia, single ventricle, and when you do this univariant, obviously you have that potential obfuscation or 1 variable being a surrogate for multiple others. So I guess my only suggestion would be that you really have to go look at that and see how highly linked these variables or several of them might be, and I think that will give us the clues that we need to understand how to improve.