Total Anomalous Pulmonary Venous Connection Requiring Emergency Surgery: What Comes First?

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Research article

Keywords: Total anomalous pulmonary venous connection, Surgery, Outcome

Posted Date: February 9th, 2021

DOI: <https://doi.org/10.21203/rs.3.rs-191703/v1>

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Version of Record: A version of this preprint was published at Journal of Cardiothoracic Surgery on August 2nd, 2021. See the published version at <https://doi.org/10.1186/s13019-021-01559-y>.

Abstract

Background

To examine two different methods for treating patients with a total anomalous pulmonary venous connection (TAPVC) who need emergency surgery and to summarize the effects of the two strategies by retrospectively reviewing the data of 54 patients.

Methods

A retrospective review of 54 patients with TAPVC who underwent emergency operations between December 2010 and July 2019 at a single institution was conducted. All patients exhibited respiratory and hemodynamic instability that required mechanical ventilation and inotropic support. Forty-four patients received emergency operations within 24 to 72 hours due to stabilization of the patient's condition. Stable hemodynamics were achieved, and a constant milieu interne was maintained before the operation. These patients comprised the subemergency operation (SEO) group. Rather than being subjected to efforts to obtain stable hemodynamics and maintain a constant milieu interne, ten patients received emergency operations immediately within 24 hours of diagnosis or an emergency operation is performed immediately due to uncorrectable acidosis or progressive cardiovascular collapse. These patients comprised the emergency operation (EO) group. The hospital course, operative data, and outpatient records were reviewed.

Results

The median weight, median age at surgery, mean cardiopulmonary bypass (CPB) duration and mean aortic cross-clamp (ACC) duration were not significantly different between the two groups. The median durations of ventilator support were 8.1 ± 4.6 (2-13) days in the SEO group and 4.9 ± 2.1 (2-18) days in the EO group, resulting in a significant difference ($p=0.008$). There were 12 operative mortalities (27.3%) in the SEO group and 2 operative mortalities (20%) in the EO group, resulting in no significant difference in mortality ($p=0.636$). Postoperative complications, such as low cardiac output and arrhythmia, were not significantly different between the two groups. The actuarial survival rates in the EO and SEO groups at 5 years were 87.5% and 89.9%, respectively. There was no difference in actuarial survival between the two groups at the latest follow-up (SEO group 89.9% versus EO group 87.5%, $p=0.8115$).

Conclusion

Rather than making efforts to achieve stable hemodynamics and maintain a constant milieu interne, an emergency operation should be performed immediately, and some efforts to address metabolic acidosis do not reduce mortality.

Introduction

The outcomes of the surgical repair of a total anomalous pulmonary venous connection (TAPVC) have historically improved, with the mortality rates reported in the literature ranging from 10–20%^{1,2} or being below 7%^{3,4}. In contrast, some patients have a risk of progressive pulmonary hypertension (PH) due to pulmonary venous obstruction (PVO), and other patients present with increasing cyanosis, tachypnea, hepatomegaly, hypoxia, gasping, or poor feeding⁵ due to a delayed diagnosis. All these patients have a risk of progressive PH, suffer from severe respiratory distress and metabolic acidosis and require immediate intubation, nitric oxide and inotropic support; more importantly, an emergency operation is needed. However, there are two viewpoints pertaining to the encounter of this situation. In some patients, the specialist would make efforts to obtain stable hemodynamics, maintain a constant milieu interne, and then perform an emergency operation. This process can take several days. In other patients, an emergency operation is performed immediately instead of trying to obtain stable hemodynamics and maintain a constant milieu interne, what's more, preoperative medical stabilization is impossible in such cases, even with unstable hemodynamics, an emergency operation is performed. Because of the rarity of reports on these patients who must undergo immediate emergency operations, we conducted a retrospective study to assess the outcomes of such patients. We aimed to determine whether pharmacological interventions to obtain stable hemodynamics and maintain a constant milieu interne are essential.

Material And Methods

Record Review and Definitions

In this retrospective analysis, we reviewed the clinical records of patients with isolated TAPVC between December 2010 and July 2019. Patients with single ventricles, atrial isomerism and other complex congenital heart diseases were excluded. Patient characteristics, such as weight and age, pulmonary venous anatomy, demographics, preoperative data, operative data, postoperative data and follow-up data, were recorded. A total of 234 patients who were diagnosed with TAPVC underwent surgical repair at the Children's Hospital of Chongqing Medical University. Fifty-four patients (23.1%) needed emergency operations for one or more of the following indications: (1) hemodynamic instability, (2) worsening metabolic acidosis, (3) severe progressive cyanosis, or (4) escalating intensive care support as described in other studies^{6,7}. We performed conventional hyperventilation and pharmacological interventions, such as treatment with prostaglandin E1, milrinone and dopamine, with the aim of controlling PH, maintaining stable hemodynamics and restoring homeostasis^{5,8}. Forty-four of these patients received emergency operations after their hemodynamics and milieu interne were stabilized, within 24 to 72 hours, and are referred to as the subemergency operation (SEO) group. Additionally, 10 patients received emergency operations within 24 hours after diagnosis due to uncorrectable acidosis or progressive cardiovascular collapse. We refer to these patients as the emergency operation (EO) group.

Echocardiography combined with computed tomography angiography (CTA) was performed in all patients in the SEO group. CTA is an accurate, noninvasive, economical diagnostic modality for the

preoperative evaluation of TAPVC^{3, 9, 10}. Echocardiography was performed in all of the patients in the EO group, and 7 patients underwent CTA. None of the patients in either group underwent cardiac catheterization, given the invasiveness of the procedure. PVO was defined if Doppler examination revealed a pulmonary venous flow velocity > 2 m/s^{3, 11, 12}. Hospital mortality was defined as mortality within 30 days of surgery or before discharge. All other deaths were considered late mortality^{3, 13}.

Long-term Follow-up

All surgical patients who were discharged alive from the hospital were required to return for outpatient follow-up visits after the initial operation and annually thereafter. Some patients were required to visit local hospitals for routine examinations. Any abnormal examination result or suspicious change in the cardiac condition prompted a return visit for further evaluation. During the follow-up period, echocardiography was performed routinely.

Surgical Technique

The surgery was performed using standard cardiopulmonary bypass (CPB). Ice slush was used for myocardial protection in all patients. A temporal reduction in bypass flow could be used to achieve satisfactory intraoperative visualization if needed. The operation was performed through a median sternotomy, there was minimal manipulation of the heart until CPB was established, and the ductus arteriosus was dissected and ligated following the bypass.

In patients with supracardiac TAPVC, a long incision was made at the pulmonary venous confluence, and then an incision was made at the corresponding position on the left atrium. Direct side-to-side anastomosis was performed between these two chambers from outside with a running suture to ensure precise geometric alignment and avoid tension, torsion, and rotation; the anastomosis was made to be as large as possible¹⁴. This approach was described by Tucker¹⁵. The atrial septal defect (ASD) was directly closed with a patch. In patients with cardiac TAPVC, the coronary sinus was unroofed, and a wide tunnel was created between the left atrium and the coronary sinus¹⁶. In patients with infracardiac TAPVC, the heart was lifted, and a wide anastomosis was generated between the pulmonary venous chamber and the left atrium with the same procedure as that used for the supracardiac form¹⁴.

Statistical Analysis

Quantitative data are expressed as the mean $\pm$ standard deviation as appropriate. All P values were two-sided, and $P < 0.05$ was considered statistically significant. Age, sex, and type were determined by using the chi-square test. Quantitative data (weight, bypass time, and clamp time) were analyzed by using a t test. Survival data are presented by means of the Kaplan-Meier method.

Results

There were 44 patients in the SEO group, of whom 31 were male and 13 were female. The median weight at surgery was 4.3 ± 1.4 kg (1.7–5.5 kg), and the median age at surgery was 72.4 ± 85.4 days (1–455 days). One patient was 15 months old and lived in a remote mountain area. He had respiratory and hemodynamic instability that required mechanical ventilation. Twenty patients were diagnosed with prerepair PVO; of these patients, 13 exhibited the supracardiac type, 6 exhibited the cardiac type, and 1 exhibited the mixed cardiac type. Ten patients were enrolled in the EO group, of whom 6 were male and 4 were female. The median weight at surgery was 4.3 ± 0.9 kg (2.8–4.9 kg), and the median age at surgery was 37.5 ± 25.5 days (6–90 days). Preoperative PVO occurred in 6 patients; of these patients, 2 exhibited the supracardiac type, 1 exhibited the cardiac type, and 3 exhibited the infracardiac type. Patient demographics are listed in Table 1.

Table 1
Demographics and clinical characteristics of the study cohort

Demographics		SEO (n = 44)	EO (n = 10)	t/ χ^2	p
Sex	Male	31	6	0.413	0.521
	Female	13	4		
Age at presentation, mean $\pm$ SD (d)		72.4 $\pm$ 85.4	37.5 $\pm$ 25.5	1.273	0.209
Weight at presentation, mean $\pm$ SD (kg)		4.3 $\pm$ 1.4	4.3 $\pm$ 0.9	0.066	0.948
PVO		45.5% (20/44)	60% (6/10)	0.927	0.485
TAPVC type	Supracardiac	23	3	4.123	0.248
	Cardiac	15	4		
	Infracardiac	4	3		
	Mixed	2	0		

The mean CPB duration was 134.9 ± 42.9 (58–303) minutes in the SEO group and 133.0 ± 41.0 (66–193) minutes in the EO group ($p = 0.901$). The mean aortic cross-clamp (ACC) duration was 68.2 ± 25.1 (36–135) minutes in the SEO group and 54.7 ± 21.5 (range, 27–93) minutes in the EO group ($p = 0.123$). The median duration of ventilator support was 8.1 ± 4.6 (2–13) days in the SEO group and 4.9 ± 2.1 (2–18) days in the EO group ($p = 0.008$). Patient demographics are listed in Table 2.

Table 2
Surgical data

	SEO (n = 44)	EO (n = 10)	t/ χ^2	p
Mean aortic cross-clamp (ACC) duration (min)	68.2 $\pm$ 25.1	54.7 $\pm$ 21.5	1.569	0.123
Mean cardiopulmonary bypass duration (min)	134.9 $\pm$ 42.9	133.0 $\pm$ 41.0	0.125	0.901
Duration of ventilation (d) ^a	8.1 $\pm$ 4.6	4.9 $\pm$ 2.1	2.892	0.008
Mortality (n/%)	12 (27.3%)	2 (20%)	0.224	0.636
^a : deaths were excluded				

Table 3 shows the comparison of means and χ^2 test results for the complications encountered postoperatively in both groups. Delayed sternal closure was performed in 5 patients and 1 patient in the SEO group and EO group, respectively. Low cardiac output occurred in 9 patients and 2 patients. Arrhythmia occurred in 6 patients and 1 patient. One patient in the SEO group and no patient in the EO group developed PVO. Diaphragmatic paralysis was found in 1 patient in the SEO group and none in the EO group; peritoneal dialysis was performed in 8 patients in the SEO group and 3 patients in the EO group. There were no statistically significant differences between the two groups regarding the complications encountered postoperatively.

Table 3
Postoperative complications in the two groups

	SEO (n = 44)	EO (n = 10)	t/ χ^2	p
Low cardiac output	9 (20.5%)	2 (20%)	0.001	0.974
Peritoneal dialysis	8 (18.2%)	3 (30%)	0.702	0.402
Arrhythmia	6 (13.6%)	1 (10%)	0.095	0.757
Postoperative PVO	1 (2.3%)	0	0.232	0.630
Diaphragmatic paralysis	1 (2.3%)	0	0.232	0.630
Delayed sternal closure	5 (11.4%)	1 (10%)	0.015	0.901

There were 12 (27.3%) and 2 operative mortalities (20%) in the SEO group and EO group, respectively, but the difference was not significant ($p = 0.636$). Among the 14 patients who died within 30 days of surgery, the main causes of death were as follows: low output failure ($n = 8$), pulmonary hypertensive crisis ($n = 3$), multiple organ dysfunction syndrome ($n = 1$) and failure to wean from ventilation ($n = 1$). All patients were weaned off CPB with inotropic support, except for 1 patient in the SEO group.

The median follow-up time for survivors was 5.9 years (0.4 to 9.2 years). In the EO group, there was one late mortality of a patient with infracardiac TAPVC 9 months after the operation because of PVO. The

actuarial survival at 5 years was 87.5% (Fig. 1). In the SEO group, 2 patients were lost to follow-up. Three late deaths occurred after the operation, and all occurred during follow-up at 7 months, 9 months and 1.2 years due to PVO. The actuarial survival at 5 years was 89.9% in the SEO group (Fig. 1). There was no difference in actuarial survival between the two groups at the latest follow-up (SEO group 89.9% versus EO group 87.5%, $p = 0.8115$).

Discussion

TAPVC is a frequently encountered problem in some heart centers. Some patients have PVO or a delayed diagnosis, and the condition can suddenly worsen, leading to critical illness with respiratory and hemodynamic instability, requiring mechanical ventilation, volume substitution, and inotropic support. Briefly, an emergency operation should be performed in such patients. In this case, someone would make efforts to obtain stable hemodynamics and maintain a constant milieu interne, and then, an emergency operation would be performed. In other cases, an emergency operation would be performed immediately. However, few studies have focused on these patients, and the more appropriate scheme for these patients remains controversial.

Treatment of these patients has been a topic of interest and has attracted much attention, but there has been no consensus reached regarding the best strategy. Some reports show that an emergency operation should be performed immediately after a diagnosis is made. Medical efforts are ineffective at managing the hemodynamic and metabolic problems that subsequently occur. Mechanical ventilation, volume substitution and inotropic support do little to help maintain stable hemodynamics and a constant milieu interne. A megadose of vasoactive medications can even worsen cardiac function. Previous studies concluded that immediate emergency surgery was the best strategy, regardless of the high operative risk^{5, 17, 18}.

In contrast, stabilizing the patient's condition before surgery is considered important for improving the surgical outcome. Some studies have reported the use of vasoactive medications such as dopamine, prostaglandin E1 and milrinone to stabilize hemodynamics and correct metabolic acidosis, and when the patient's condition was stable, an urgent operation was performed. Although one study reported a reduction in mortality, there was insufficient evidence to support this conclusion¹⁷. In addition, other palliative surgical techniques, such as balloon dilatation, stenting and atrial septectomy, are alternative approaches to relieve cyanosis and improve the patient's clinical state. These palliative techniques have been shown to be somewhat successful for decompressing pulmonary venous pressure with restricted ASD or PVO. These techniques are associated with some inherent complications, such as the need for reintervention (due to occlusion of the stent or the recurrence of PVO) and the need for anticoagulation. Research has shown that the mortality rate is 38% at 1 year and that the reintervention rate is 58% at 1 year^{2, 5, 19}. Moreover, not all patients could tolerate these palliative measures, and patients needed to undergo surgery to correct the anomalous pulmonary veins⁵. Extracorporeal membrane oxygenation

(ECMO) is also an alternative approach, but its cost and serious complications restrict its popularization^{2, 20}.

A finding of our review is that the mortality rate was higher in the SEO group than in the EO group, though without significance. The reasons might be as follows. First, according to some clinical reports, there is evidence that the long-term application of cardiovascular drugs does not improve myocardial function or survival^{21–23}. Indeed, some researchers have shown that the use of these drugs results in myocardial injury, the destruction of myocardial function via myocardial cellular apoptosis and a decrease in heart contractility, which substantially affects myocardial function²¹. Second, due to severe dyspnea and aggravated metabolic acidosis, these patients' hearts contracted weakly, and their oxygenation continued to worsen unless the malformation was corrected. Additionally, their cardiac function worsened during the preoperative preparation period¹.

Another more important and reasonable finding is that the duration of ventilation was significantly longer in the SEO group. Longer postoperative mechanical ventilation results in a higher risk of ventilator-associated pneumonia and prolongs the length of stay. Additionally, studies have shown that prolonged postoperative mechanical ventilation is the most significant predictor of early mortality²⁴. Long postoperative mechanical ventilation is usually attributed to the presence of chest infection or a worsened condition of heart failure. Therefore, the longer postoperative mechanical ventilation period in the SEO group might be explained as follows. First, an infusion of excess fluid, aiming at stabilizing hemodynamics and correcting metabolic acidosis before the operation, has been demonstrated to worsen mechanical ventilation-related complications and aggravate pneumonia in critically ill patients^{6, 24}. Second, an infusion of excess fluid and long-term application of cardiovascular drugs can also weaken myocardial functioning and prolong mechanical ventilation, as discussed above.

Our report indicates that early mortality among these critically ill patients (in both the SEO group and the EO group) is higher than the overall TAPVC mortality reported by others. It seems that those who undergo emergency operations have a higher overall mortality rate². Clearly, patients with respiratory distress and metabolic acidosis or PVO are the most severely ill patients, and these factors have been shown to be risk factors for mortality in many studies. The urgency of surgery itself is also a risk factor for mortality¹, and therefore, such patients are expected to have a higher mortality rate^{1, 4, 11}. Moreover, in some patients, the hemodynamic status was still unstable after all of the measures mentioned above had been implemented, and an emergency operation became the only choice when the palliative techniques were abandoned, considering the aforementioned defects. Regarding the emergency operations themselves, the early mortality is similar to the result of other reports^{1, 3, 24}.

Postoperative complications, such as low cardiac output and arrhythmia, were not significantly different between the two groups. Long-term survival also did not differ between the two groups. Operative survivors in the two groups (among critically ill patients) appear to have similar long-term outcomes, such as overall actuarial survival. Zhao K et al. also reported a similar result¹. Postoperative patients may have

late pulmonary vein obstruction, as seen in other patients with TAPVC; the proportion of patients is similar to that with overall TAPVC postoperative PVO³. In other words, patients undergoing an emergency operation are unlikely to be more predisposed to stenosis. The statistical analyses in our series showed a good outcome, with freedom from medication and normal activity.

Considerable research has been performed to identify the risk factors for mortality among patients with TAPVC, while little research has been devoted to the subset of patients undergoing emergency surgery. Our study showed that the mortality rate was higher in the SEO group than in the EO group, although without significance. Moreover, the duration of ventilation was significantly longer in the SEO group than in the EO group, which was not previously reported. Therefore, emergency operations should be performed immediately to correct cardiac malformations rather than taking efforts to obtain stable hemodynamics and maintain a constant milieu interne. Medical treatments and other processes do not reduce mortality.

There are multiple limitations of this study. This was a retrospective single-center study, and the sample size was small; thus, the results may not be applicable to the population as a whole. TAPVC often combined with various malformations. Although cases combined with malformations of high risk for death were excluded, other concurrent abnormalities could still affect survival outcomes.

Conclusion

Taken together, our experience with these critically ill patients is as follows. First, given the lower mortality rate (though without significance compared to the SEO group) and shorter duration of ventilation in the EO group, emergency operations should be performed without any delay. The evaluation of the clinical state before the operation should be accurate and rapid, and focus should not be placed on maintaining stable hemodynamics and a constant milieu interne. Second, for patients with respiratory and hemodynamic instability, mechanical ventilation and inotropic support should be applied quickly.

Abbreviations

TAPVC: total anomalous pulmonary venous connection; PVO: pulmonary venous obstruction; subemergency operation (SEO); emergency operation (EO); cardiopulmonary bypass (CPB) duration and mean aortic cross-clamp (ACC); pulmonary hypertension (PH) because of pulmonary venous obstruction (PVO); computed tomography angiography (CTA); atrial septal defect (ASD); extracorporeal membrane oxygenation (ECMO)

Declarations

Ethics approval and consent to participate: This research received ethical approval and consent. It did not require the approval of an ethics committee.

Consent for publication: All patient information contained in this manuscript has been published with the consent of their parents.

Availability of data and materials: The data and materials in the manuscript are available, and the original data for the relevant results are owned by myself; I can be contacted if needed.

Competing interests: There are no conflicts of interest or legal economic disputes in this study.

Funding: Not applicable.

Authors' contributions: Linyun Xi, the first author, designed the work, acquired, analyzed, and interpreted the data, drafted the work and substantively revised it. Chun Wu, the second author, substantively revised it. Zhengxia Pan, the third author, provided some suggestions for this work. Ming Xiang, the corresponding author, designed the work, drafted the work, substantively revised it, and approved the submitted version.

Acknowledgements: I would like to thank all the doctors and nurses of the Cardiothoracic Surgery Department of the Children's Hospital of Chongqing Medical University for giving me so much advice and help. I appreciate my family for their companionship, support and care.

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Figures

Figure 1

Actuarial survival at 5 years in the two groups.