Emergency surgical treatment of total anomalous pulmonary venous connection

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Abstract

Background: This study explores the strategy and effect of emergency surgical treatment for total anomalous pulmonary venous connection (TAPVC). Methods: From March 2009 to February 2020, 78 patients with TAPVC underwent emergency surgical correction. There were 51 males and 27 females. The median age was 39.5 days, and the median weight was 4.0 kg. The preoperative percutaneous oxygen saturation was 80.8±4.5%. Results: Of the cases investigated, seven died during the perioperative period, 16 had delayed chest closure, 19 had early pulmonary vein obstruction, two had secondary tracheal intubation, one had a brain complication, and one had third-degree atrioventricular block. Low weight, younger age, cardiopulmonary bypass time, and aortic cross-clamp time were identified as risk factors for early mortality. During the follow-up from four to 137 months, 12 cases did not respond to follow up. Ten patients died within one to six months after discharge. One patient underwent reoperation due to pulmonary vein obstruction. The longer hospital stays after operation and intensive care unit time were identified as risk factors for late mortality. Conclusions: Emergency surgery for severe TAPVC patients after admission had achieved good results in the near future. Prenatal diagnosis should be strengthened to save more patients. The higher late mortality rate indicates that such patients should strengthen post-discharge management to reduce the occurrence of post-discharge deaths.

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4.0 kg. The preoperative percutaneous oxygen saturation was 80.8±4.5%.

Results: Of the cases investigated, seven died during the perioperative period, 16 had delayed chest closure, 19 had early pulmonary vein obstruction, two had secondary tracheal intubation, one had a brain complication, and one had third-degree atrioventricular block. Low weight, younger age, cardiopulmonary bypass time, and aortic cross-clamp time were identified as risk factors for early mortality. During the follow-up from four to 137 months, 12 cases did not respond to follow up. Ten patients died within one to six months after discharge. One patient underwent reoperation due to pulmonary vein obstruction. The longer hospital stays after operation and intensive care unit time were identified as risk factors for late mortality.

Conclusions: Emergency surgery for severe TAPVC patients after admission had achieved good results in the near future. Prenatal diagnosis should be strengthened to save more patients. The higher late mortality rate indicates that such patients should strengthen post-discharge management to reduce the occurrence of post-discharge deaths.

Total anomalous pulmonary venous connection (TAPVC) represents 1%–3% of all congenital heart diseases (CHD) [1-4]. TAPVC is one of the few congenital heart diseases requiring emergency surgery, with poor natural prognosis and high mortality. In this study, we reviewed and analyzed TAPVC emergency surgery for more than 10 years to assess the risk factors of early and late mortality.

Emergency operation refers to operation that occurs within 24 hours of diagnosis. We have determined that emergency operation should be performed in two situations: (1) critical illness, difficulty breathing, or need for tracheal intubation due to aspiration; and (2) patients with preoperative pulmonary venous obstruction (PVO) (pulmonary vein velocity $>$ 120 cm/s) or restrictive atrial septal defect (ASD) before operation. In order to strictly focus on the risk factors of critical TAPVC emergency operations.Two situations were excluded from this analysis. 1) Patients with TAPVC who were operated on within 24 hours but had no serious symptoms; 2) Patients with TAPVC had preoperative obstruction without serious symptoms but did not have an operation within 24 hours.

Patients and Method s

Clinical Data

From March 2009 to February 2020, 78 of 400 TAPVC cases underwent an emergency operation. The studies involving human participants were reviewed and approved by the ethics committee of Anzhen Hospital.Of these cases, 36 were admitted to the intensive care unit before operation, and six were intubated or rescued before operation. There were 66 cases of preoperative PVO, including 51 cases of pulmonary venous reflux obstruction and 15 cases of restrictive ASD. Seven cases had confirmed prenatal diagnosis. The Darling's classifications were 39 cases of supracardiac, 21 cases of intracardiac, 16 cases of infracardiac and two cases of mixed type. There were 51 males and 27 females. In regard to age, 34 cases were a patient less than one month old, 26 cases were a patient from one month to three months old, and 18 cases were a patient more than three months old. The median age was 39.5 days, and the median weight was 4.0 kg. The preoperative percutaneous oxygen saturation was 80.8±4.5% (Table 1). Risks of early mortality were measured individually with patient parameters to assess their significance on the outcome of surgical repair. These factors included age and weight at the time, type of TAPVC, preoperative PVO, preoperative O_2 saturation, preoperative signs of heart failure, cardiopulmonary bypass (CPB) time, aortic cross-clamp (ACC) time, postoperative complications (pulmonary restenosis, delayed chest closure), total hospital stay, and duration of postoperative ventilation.

Table 1. Patient characteristics (n=78)

Sex		n	%
	Male	51	65.4%
	Female	27	34.6%
Median weight at surgery	Median weight at surgery	4.0 (2.4–12) kg	

Sex		n	%
Median age at surgery	Median age at surgery	39.5 (1–576) days	
Type of TAPVC	Type of TAPVC		
	Supracardiac	39	50.0%
	Cardiac	21	26.9%
	Infracardiac	16	20.5%
	Mixed	2	2.6%
Preoperative PVO	Preoperative PVO		
	Supracardiac	34	87.2%
	Cardiac	18	85.7%
	Infracardiac	12	75.0%
	Mixed	2	100.0%
Preoperative signs heart failure	Preoperative signs heart failure	36	46%
Preoperative O2 saturation	Preoperative O2 saturation	79.6±6.5	
Postoperative complications	Postoperative complications		
	Delayed chest closure	16	20.5%
	PVO	19	24.4%
	Secondary endotracheal intubation	2	2.6%
	Brain complications	1	1.3%
	III atrioventricular block	1	1.3%

TAPVC: Total anomalous pulmonary venous connection; PVO: pulmonary venous obstruction

Operative Data

All operations were performed through the median sternotomy under general anesthesia. Cardiopulmonary bypass was established via aorto-bicaval cannulation. Myocardial protection was carried on with HTK cardioplegia and systemic hypothermia to 28°C.

Supracardiac type. The vertical vein was ligated, and an incision was made in the pulmonary venous confluence and left atrium from the gap between the aorta and the superior vena cava. A wide anastomosis was made between the pulmonary vein confluence and the left atrium. The ASD was closed with a patch to enlarge the left atrium (LA).

Intracardiac type. The coronary sinus was unroofed to connect the coronary sinus with the LA. The ASD was closed using a patch to baffle the anomalous veins into the LA.

Infracardiac type. A vertical incision was made in the confluence of pulmonary veins and a matching incision was done on the posterior left atrial wall. Both incisions were then anastomosed side-to-side together. The ASD was closed with a patch to enlarge the LA.

Mixed type. A combination of the above techniques was used.

Statistical Analysis

Data were coded and entered into the statistical package SPSS (Statistical Package for the Social Sciences) Version 19. Data were summarized using mean ± standard deviation, median, minimum and maximum (range) in quantitative data as well as frequency (count) and relative frequency (percentage) for categorical data. Univariate analysis and the significance of the association between quantitative variables were done using the non-parametric Mann-Whitney test. For comparing categorical data, the Pearson chi-square test was performed unless the expected frequency was less than 5, in which case the Fisher exact test was used instead. Multivariate logistic regression analysis was done to detect independent predictors of death. P values less than 0.05 were considered statistically significant. Kaplan-Meier curves were used to determine the overall survival rate.

Results

In-Hospital Results

The average CPB time was 122.1±44.9 minutes. The average ACC time was 59.5±22.4 minutes, and the average postoperative hospitalization time was 17.4±9.4 days. The average intensive care unit (ICU) time was 11±6.8 days, and the average mechanical ventilation time was 4.0±2.1 days.

Mortality Rate

In seven cases, the patient died in the hospital after operation. This included, by Darling's classifications, four cases of the supracardiac type, two cases of infracardiac type, and one case of mixed type. All the cases in which the patient died were accompanied with preoperative PVO, and in two cases patients were rescued before operation. In the all cases,16 had delayed chest closure, 19 had early pulmonary vein obstruction, two had secondary tracheal intubation, one had a brain complication, and one had third-degree atrioventricular block.

Upon univariate analysis to determine significant risks of early mortality, lower age and weight at time of surgery, as well as longer CBP and ACC times were found to be associated with increased mortality (Table 2). Significant variables in mortality were entered into a multivariate analysis to determine the independent predictors of in-hospital mortality. One independent predictor determined was CPB time (Table 3,4).

Table 2. Univariate analysis of early mortality of TAPVC in emergency surgery

Age, days			
Sex, n (%)			
	Male		
	Female		
Weight, kg	Weight, kg		
TAPVC type, n (%)	TAPVC type, n (%)	TAPVC type, n (%)	
	Supracardiac	Supracardiac	S
	Cardiac		
	Infracardiac	Infracardiac	I
	Mixed		
Preoperative PVO, n (%)	Preoperative PVO, n (%)	Preoperative PVO, n (%)	P
Preoperative O2 saturation	Preoperative O2 saturation	Preoperative O2 saturation	P
Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	P
CBP time	CBP time		
ACC time	ACC time		

TAPVC: Total anomalous pulmonary venous connection; PVO: pulmonary venous obstruction; CBP: cardiopulmonary bypass; ACC :aortic cross-clamp

Table 3. Univariate analysis of postoperative PVO of TAPVC in emergency surgery

Age, days		5
Sex, n (%)		
	Male	1
	Female	6
Weight, kg	Weight, kg	4
TAPVC type, n (%)	TAPVC type, n (%)	

	Supracardiac	Supracardiac	
	Cardiac		
	Infracardiac	Infracardiac	
	Mixed		
Preoperative PVO, n (%)	Preoperative PVO, n (%)	Preoperative PVO, n (%)	
Preoperative O2 saturation	Preoperative O2 saturation	Preoperative O2 saturation	
Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	
CBP time	CBP time		
ACC time	ACC time		

TAPVC: Total anomalous pulmonary venous connection; PVO: pulmonary venous obstruction; CBP: cardiopulmonary bypass; ACC :aortic cross-clamp

Table 4. Most significant predictors of early mortality on multivariate analysis

	B	SE	Wals	df	P	OR	95% CI	95% CI
							lower	upper
CBP time	.027	.009	8.353	1	.004	1.028	1.009	1.047
Constant	-6.264	1.589	15.533	1	.000	.002		

CBP: cardiopulmonary bypass

Follow-Up

Of the 71 patients who survived until discharge, 59 patients were followed up for a period of four to 137 months. There were 10 late deaths one to six months after discharge, including eight cases within three months and two cases between three and six months. Two cases died of asphyxia, one case died of pneumonia, five cases died of PVO after operation, and the other two cases died of unknown causes(fig 1). One case was re-operated on because of PVO, and survived normally after the operation. Upon univariate analysis to determine significant risks of late mortality, longer hospital stays after operation and ICU time were found to be associated with increased mortality. Significant variables in mortality were entered into a multivariate analysis to determine the independent predictors of late mortality. One independent predictor determined was ICU time (Table 5, 6).

Figure 1. Kaplan-Meier curves for all survival

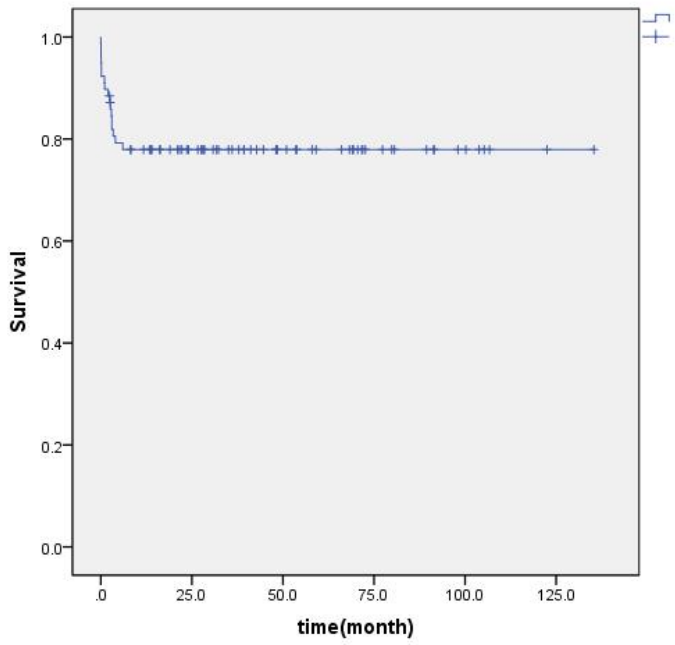

Table 5. Univariate analysis of later mortality of TAPVC in emergency surgery

Age, days	Age, days		
Sex, n (%)	Sex, n (%)		
	Male	Male	
	Female	Female	
Weight, kg	Weight, kg		
TAPVC type, n (%)	TAPVC type, n (%)	TAPVC type, n (%)	
	Supracardiac	Supracardiac	S
	Cardiac	Cardiac	
	Infracardiac	Infracardiac	I
	Mixed	Mixed	
Preoperative PVO, n (%)	Preoperative PVO, n (%)	Preoperative PVO, n (%)	
Preoperative O2 saturation	Preoperative O2 saturation	Preoperative O2 saturation	P
Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	Preoperative signs heart failure, n (%)	P
CBP time	CBP time		
ACC time	ACC time		
Postoperative hospitalization time	Postoperative hospitalization time	Postoperative hospitalization time	P
ICU time	ICU time		
Mechanical ventilation time	Mechanical ventilation time	Mechanical ventilation time	M
Postoperative PVO	Postoperative PVO	Postoperative PVO	P
Delayed chest closure	Delayed chest closure	Delayed chest closure	D

TAPVC: Total anomalous pulmonary venous connection; PVO: pulmonary venous obstruction; CBP: cardiopulmonary bypass; ACC :aortic cross-clamp; ICU: intensive care unit

Table 6. Most significant predictors of later mortality on multivariate analysis

	B	SE	Wals	df	P	OR	95% CI	95% CI
							Lower	Upper
ICU time	.111	.051	4.679	1	.031	1.117	1.010	1.235
Constant	-3.175	.748	18.008	1	.000	.042		

ICU: intensive care unit

Comment

There have been many descriptions of emergency operations for various CHDs. However, with the clinical application of prostaglandin E1, TAPVC has become one of the few CHDs that really necessitates emergency operation [5-7].

Non-obstructive TAPVC will generally cause progressive right ventricular dilatation and pulmonary hypertension, which directly leads to heart failure. Additionally, irreversible pulmonary vascular obstruction will occur if not treated. With the growth of some children, unrestricted ASD gradually develops into restrictive ASD, and serious symptoms appear, which is life-threatening. There were 12 patients with non-obstructive TAPVC, all of whom had shortness of breath or difficulty breathing, pulmonary congestion, and severe pulmonary hypertension before operation, and one of them had an emergency endotracheal intubation. Among 15 patients with TAPVC with restrictive ASD, 11 patients did not reach the degree of restrictive ASD in newborns, but they had restrictive ASD at the time of visit and had symptoms of different degrees. Thus, we cannot ignore the patients with non-obstructive TAPVC, which should be operated on as soon as possible because serious consequences can occur if not treated in time.

Obstructive TAPVC causes increased pulmonary artery pressure in the early postnatal period, which leads to a large number of right-to-left shunts, and decreased pulmonary blood flow. Pulmonary edema caused by pulmonary vein obstruction will quickly lead to progressive hypoxemia, and hemodynamics will fail. Severe respiratory and heart failure may occur before operation. If not treated in time, most patients die within two months of birth. In this group, 66 children had preoperative PVO, including 34 supracardiac, 12 infracardiac, two mixed and 18 intracardiac type who had increased flow velocity into the right atrium or restrictive ASD. In these 66 children, five of them were rescued before operation.

In early years, prenatal diagnosis was less in China. Most patients were older when they went to see a doctor. Liufu R reported 198 cases of children treated with TAPVC, of which only 21 were newborns. Of the 768 patients with TAPVC reported by Shi G, 112 were newborns[8][9]. In this group, only 7 cases were diagnosed in fetus. The rest went to see a docter when they had symptoms or for other diseases. Sometimes they could not be diagnosed or treated in time in other hospitals.

For obstructive TAPVC, it is necessary to give targeted cardiac strengthening, diuresis, oxygen inhalation and limited intake, before operation for preoperative adjustment. In such situations, once progressive hypoxia, systemic hypoperfusion, and progressive hemodynamic deterioration occur, preoperative intubation and ventilator-assisted ventilation should be given immediately. There were 36 cases with severe symptoms in this group, among which six patients were treated by surgery after rescue. All six patients were given necessary treatment before rescue, and there were no obvious signs prior to rescue. Among them, two patients died due to internal environment disorder before rescue and after operation.

Young age, low surgical weight, preoperative PVO, surgical method (wireless suture), longer cardiopulmonary bypass and aortic occlusion time may be important risk factors for death or postoperative PVO [3][8-14]. The risk of postoperative death occurred in the first 18 months after discharge. Because of the critical condition, all of the children in this group underwent emergency operation, which has many influencing factors. However, younger age, lower operation weight, and longer CPB and ACC time are still related to

early death, but not to postoperative PVO. Postoperative death occurred within six months after discharge, which was related to postoperative hospital stay, ICU time, and mechanical ventilation time.

Emergency TAPVC surgery has a great influence on the results because of the combined effects. Some studies had excluded emergency surgery[15]. In this study, the result of emergency TAPVC operation is just slightly worse than that of overall TAPVC operation. The overall early mortality was 5.75%, the early postoperative PVO was 16.5%, and the one-year survival rate was 88%.

The most commonly used definition of PVO in the literature is that the pulmonary vein velocity measured by echocardiography is 2 m/s or more, and that the gradient measured by catheter is 4 mm Hg or more. Other standards have been used, including 1.2 m/s or higher, 1.5 m/s or higher, and 1.8 m/s or higher [16][17][18]. We define mild obstruction as pulmonary vein velocity greater than or equal to 1.2 m/s measured by echocardiography. In this group, preoperative PVO (66 cases) had nothing to do with postoperative PVO (19 cases). As most of these cases were preoperative PVO cases, univariate analysis had limitations.

Supracardiac TAPVC had the highest proportion of PVO. In this group, there were 34 cases of supracardiac TAPVC with PVO. The incidence rate of PVO of intracardiac TAPVC was low. In this group, only eight cases had preoperative PVO. Almost all infracardiac TAPVC had PVO. In this group, there were 16 cases of infracardiac TAPVC, and 12 cases showed increased flow velocity by echocardiography. The other four cases did not show increased flow velocity due to intrahepatic obstruction. There was one case of mixed type TAPVC (supracardiac and infracardiac mixed) with PVO.

Limitations

Due to the limitation of regional medical level and the lack of prenatal diagnosis, many patients in this group were older. Due to the limitation of retrospective study, the understanding of emergency surgery was inconsistent, and some patients who need emergency surgery did not have timely surgery, which needs further understanding.

Conclusion

Through this retrospective analysis, emergency surgery for severe TAPVC patients after admission had achieved good results in the near future, although the condition was critical and the deformity was complex. Emergency surgery is needed for such patients, not necessary to the other patients. Among the severe TAPVC patients the proportion of preoperative PVO of newborns was high, indicating that more newborns with TAPVC need emergency surgery, and prenatal diagnosis should be strengthened to save more patients. The higher late mortality rate indicates that such patients should strengthen post-discharge management to reduce the occurrence of post-discharge deaths.

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