Early and Midterm Outcome of Surgical Repair of Total Anomalous Pulmonary Venous Connection

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Abstract

Background: The mortality rates following surgical correction of Total anomalous pulmonary venous connection (TAPVC) have shown improvement due to advancements in surgical techniques.

Objectives: This work aimed to analyze the early and midterm outcomes of TAPVC single-stage surgical repair and to identify the risk of morbidity and mortality.

Patients and methods: This prospective study was carried out on 27 TAPVC patients who underwent different single-stage surgical techniques. The extracardiac type was treated with side-to-side anastomosis and vertical vein ligation, while the cardiac type required cutting back to the coronary sinus, intraoperative data, postoperative course, mortality rate, and complications were collected and analyzed.

Results: 27 patients with ages ranged from 0.26 to 84 months, 51.85% were males while 48.15% were females. Mortality occurred in 9 (33.33%) patients due to cardiac arrest during anesthesia induction in 1 (3.7%) patient, sepsis in 2 (7.41%) patients, LCOP in 4 (14.81%) patients and pulmonary hypertensive crisis in 2 (7.41%) patients. Postoperative complications were pneumothorax in 2 (7.41%) patients, pulmonary edema in 1 (3.7%) patient, re-intubation in 2 (7.41%) patients, pneumonia in 1 (3.7%) patient, fits in 2 (7.41%) patients, thrombocytopenia in 1 (3.7%) patient, bleeding from ETT in 1 (3.7%) patient, LCOP in 5 (18.52%) patients, pulmonary hypertensive crisis in 3 (11.11%) patients, cardiac arrest in 1 (3.7%) patient, renal failure in 1 (3.7%) patient and no complication in 14 (51.85%) patients. Re-opening occurred in 1 (3.7%) patient. Sepsis occurred in 2 (7.41%) patients. The mean postoperative bleeding was 109.8 ±77.6 ml.

Conclusion: The high morbidity and mortality associated with TAPVC single-stage repair, emphasizing the need for careful preoperative evaluation and perioperative management.

Keywords: Early and Midterm Outcomes; Surgical Repair; TAPVC; Morbidity; Mortality.
DOI: 10.21608/SVUIJM.2024.270729.1809
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Received: 9 March,2024.
Revised: 13 April,2024.
Accepted: 14 April,2024.
Published: 4 October,2024.
Cite this article as: Sandy Adel, Karam Mosallam Eisa, Mohamed Adel Elgamal, Asmaa Gaber Rizk.(2024). Early and Midterm Outcome of Surgical Repair of Total Anomalous Pulmonary Venous Connection. *SVU-International Journal of Medical Sciences*. Vol.7, Issue 2, pp: 617-629.

Introduction

Total anomalous pulmonary venous connection (TAPVC) is referred to as aberrant pulmonary venous drainage, in which all the pulmonary veins drain into the right side of the heart either directly or indirectly forming a large left-to-right shunt. It is a rare condition which represents about 1-3% of all congenital heart diseases. The presence of isolated TAPVC is incompatible with life, and necessitates the presence of a shunt, usually at the atrial level, to allow blood to reach the left atrium **(Shaw and Chen, 2017)**.

The best prognosis is related to isolated TAPVC in a patient exhibiting biventricular architecture, in the absence of significant concurrent cardiac abnormalities. Newborns that possess single-ventricle physiology and newborns diagnosed with heterotaxia syndrome are particularly high-risk groups **(Hancock Friesen et al., 2005)**

TAPVC has a mortality rate of more than 78% in the first year if it is not surgically corrected **(Nasrolahzadeh et al., 2024)**.

The objective of surgical intervention is to change the course of pulmonary vein circulation towards the left atrium. The first surgical repair of TAPVC on cardiopulmonary bypass was conducted by Kirklin at Mayo Clinic **(Appelbaum et al., 1975)**.

The mortality rates following surgical correction of Total TAPVC have shown improvement due to advancements in surgical techniques and medical interventions. However, it is important to note that repairing TAPVC still presents a significant challenge, as described in many literature sources, with early fatality rates ranging from less than 10% to 50% **(Lenhoff, 2023)**.

The incidence of morbidity following surgery for TAPVC exhibits significant variability, encompassing complications such as arrhythmias, pulmonary hypertension, and persistent blockage of the pulmonary vein **(Lenhoff, 2023)**.

The aim of this work was to analyze the early and midterm outcome of TAPVC single-stage surgical repair and to identify the risk factors of morbidity and mortality.

Patients and Methods

This prospective study was carried out on 27 patients, both sexes, with TAPVC who underwent surgical repair. The study was conducted from March 2022 to September 2023 after approval from the Ethical Committee Qena University Hospital approval code (SVU-MED-SUR011-1-22-3-357), Al Mansoura University Pediatric Hospital, Egypt. An informed written consent was obtained from relatives of the patients.

Exclusion criteria were patients with partial anomalous pulmonary venous connection (PAPVC), patients with heterotaxia syndrome and major extracardiac anomalies.

All patients were subjected to complete history taking, clinical examination, laboratory investigations, electrocardiogram (ECG), detailed pre-operative echocardiography and cardiac multi-slice computed tomography (MSCT). Anesthetic procedure, total operative time, aortic cross clamping time (ACC), total bypass time, intraoperative complications, need for blood transfusion for patients undergoing TAPVC repair are routinely collected.

Surgical technique

All cases were approached through median sternotomy using cardiopulmonary bypass (CPB), with moderate degree of hypothermia, about 24-28 ℃. Myocardial protection was achieved by the utilization of cross-clamping with cold crystalloid cardioplegia. The extracardiac type was addressed with standard side-to-side anastomosis and vertical vein ligation (**Hawkins et al., 1995**) (**Fig.1**), but surgical repair for the cardiac type involved cutting back to the coronary sinus and conducting a closure of the orifices of the coronary sinus (**Serraf et al., 1998**).

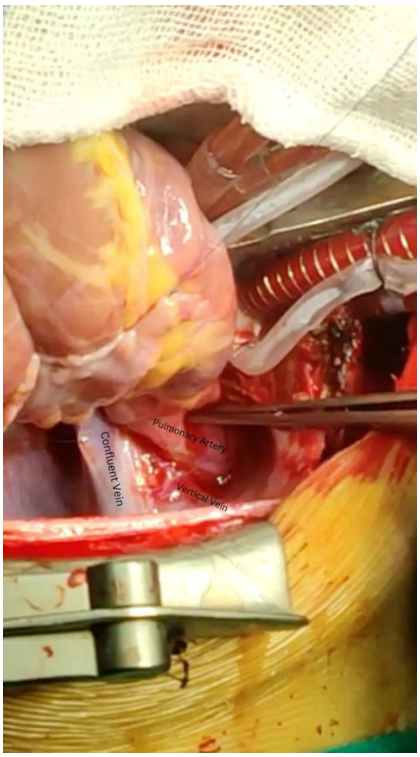

Fig.1. Intraoperative image of unobstructed TAPVC extracardiac type.

Postoperative data

Patients were transferred to Pediatric Intensive Care Unit (PICU) with monitoring of the vital signs, ventilation time, need for inotropic support, bleeding, blood transfusion, re-opening, arrhythmias, low COP syndrome, pulmonary hypertensive crisis, infection, ICU stay, total hospital stay. Also, post-operative follow-up of complications as pulmonary venous obstruction (PVO).

Follow-up

Patients were followed up in our cardiothoracic surgery outpatient clinic. Patients far to reach were contacted through their pediatrician, and the mean follow-up time was six months postoperatively. Follow-up echocardiography was done on patient's discharge from hospital and at three and six months postoperative.

Sample Size Calculation

Based on previously published data we assumed confidence level of 90% to detect 15% difference in hemodynamic variables. The sample size of 27 patients was calculated using (GraphPad Instat 3 developed by GraphPad Software, Inc).

Statistical analysis

Statistical analysis was done by SPSS v26 (IBM Inc., Chicago, IL, USA). Shapiro-Wilks test and histograms were used to evaluate the normality of the data distribution. Quantitative parametric data were presented as mean and standard deviation (SD) and were compared by paired T-test. A two-tailed P value < 0.05 was considered statistically significant.

Results

The mean age was 13.6 ± 22.77 months. The sex was male in 14 (51.85%) and female in 13 (48.15%) patients. The weight ranged from 2.5 to 19 kg. The mean height was 62.6 ±21.03 cm. The mean BSA was 0.3 ±0.16 m^2. Medical history was recurrent chest infection in 6 (22.22%) patients, severe respiratory distress in 20 (74.07%) patients, and dyspnea in 1 (3.7%) patient. The type of TAPVC was supra-cardiac in 21 (77.78%) patients, cardiac in 3 (11.11%) patients, mixed in 2 (7.41%) patients and infra-cardiac in 1 (3.7%) patient (**Table .1).**

Table 1. Demographic data, medical history, and type of TAPVC of the studied patients

Variables		N=27
Age (Months)		13.6 ± 22.77
Sex	Male	14 (51.85%)
	Female	13 (48.15%)
Weight (kg)		2.5 – 19
Hight (cm)		62.6 ± 21.03
BSA (m^2)		0.3 ± 0.16
Medical history	Recurrent chest infection	6 (22.22%)
	Severe respiratory distress	20 (74.07%)
	Dyspnea	1 (3.7%)
Type of TAPVC	Supracardiac	21 (77.78%)
	Cardiac	3 (11.11%)
	Mixed	2 (7.41%)
	Infracardiac	1 (3.7%)

Data are presented as mean± SD or frequency (%). BSA: Body surface area, TAPVC: Total anomalous pulmonary venous connection.

Preoperative obstruction was present in 6 (22.22%) patients. Associated cardiac anomalies were secundum ASD in 20 (74.07%) patients, complete AVSD in 4 (14.81%) patients, DORV in 7 (25.93%) patients, right isomerism in 4 (14.81%) patients, pulmonary valve atresia in 1 (3.7%) patient, IVC draining into the LA in 1 (3.7%) patient, malposed GA in 1 (3.7%) patient, TGA in 2 (7.41%) patients, dilated hypertrophied RV in 1 (3.7%) patient, PFO in 1 (3.7%) patient and PDA in 2 (7.41%) patients. Associated extra cardiac anomalies were present in 5 (18.52%) patients. The preoperative oxygen delivery method was room air in 25 (92.59%) patients, mechanical ventilation in 1 (3.7%) patient and nasal oxygen in 1 (3.7%) patient. The mean temperature was 36.8 ±0.23 °c. Chest radiography revealed cardiomegaly in 23 (85.19%) patients, bilateral prominent bronchovascular markings in 14 (51.85%) patients and widened mediastinum in 6 (22.22%) patients. ECG revealed sinus rhythm in all patients. All patients did not have any preoperative inotropic support. Preoperative mechanical ventilation was done in 1 (3.7%) patient (**Table.2)**.

Table 2. Preoperative obstruction, associated cardiac, extra cardiac anomalies, oxygen delivery method, temperature, chest radiography, ECG and preoperative inotropic support of the studied patients.

Variables		N=27
Preoperative obstruction		6 (22.22%)
Associated cardiac anomalies	Secundum ASD	20 (74.07%)
	Complete AVSD	4 (14.81%)
	DORV	7 (25.93%)
	Right isomerism	4 (14.81%)
	Pulmonary valve atresia	1 (3.7%)
	IVC draining into the LA	1 (3.7%)
	Malposed GA	1 (3.7%)
	TGA	2 (7.41%)
	Dilated hypertrophied RV	1 (3.7%)
	PFO	1 (3.7%)
	PDA	2 (7.41%)
Associated extra cardiac anomalies		5 (18.52%)
Preoperative oxygen	Room air	25 (92.59%)

delivery method	**Mechanical ventilation**	1 (3.7%)
	Nasal oxygen	1 (3.7%)
Temperature (°c)		36.8 ± 0.23
Chest radiography	**Cardiomegaly**	23 (85.19%)
	Bilateral prominent bronchovascular markings	14 (51.85%)
	Widened mediastinum	6 (22.22%)
ECG	**Sinus rhythm**	27 (100%)
	Arrhythmia	0 (0%)
Preoperative inotropic support		0 (0%)

Data are presented as mean± SD or frequency (%).ASD: Atrial septal defect, AVSD: Atrioventricular septal defect, DORV: Double outlet right ventricle, IVC: Inferior vena cava, LA: Left atrium, GA: Great arteries, TGA: Transposition of the great arteries, RV: Right ventricle, PFO: patent foramen ovale, PDA: Patent ductus arteriosus, ECG: Electrocardiogram, ACC: Acceleration, CPB: Cardiopulmonary bypass.

Preoperative echocardiography revealed mild pulmonary hypertension in 22 (81.48%) patients, moderate pulmonary hypertension in 5 (18.52%) patients. Tricuspid valve regurgitation was mild in 13 (48.15%) patients, moderate in 11 (40.74%) patients, and severe in 3 (11.11%) patients. All patients had good left ventricular ejection fraction. Preoperative cardiac MSCT revealed that the pulmonary venous confluence drained into the left innominate vein in 6 (22.22%) patients, into SVC in 15 (55.56%) patients, into the coronary sinus in 3 (11.11%) patients, IVC supradiaphragmatic part in 1 (3.7%) patient, and mixed sites in 2 (7.41%) patients **(Table.3)**.

Table 3. Preoperative echocardiography and cardiac MSCT data of the studied patients.

Variables		**N=27**
PASP (mmHg)	**< 20**	0 (0%)
	20 - 35	22 (81.48%)
	36 - 55	5 (18.52%)
	> 55	0 (0%)
Degree of TR	**Mild**	13 (48.15%)
	Moderate	11 (40.74%)
	Severe	3 (11.11%)
EF (%)	**< 55**	0 (0%)
	≥ 55	27 (100%)
Site of drainage of the pulmonary venous confluence	**Left innominate vein**	6 (22.22%)
	SVC	15 (55.56%)
	Coronary sinus	3 (11.11%)
	IVC supradiaphragmatic part	1 (3.7%)
	Mixed	2 (7.41%)

PASP: pulmonary artery systolic pressure, TR: tricuspid regurgitation, EF: ejection fraction, SVC: superior vena cava, IVC: inferior vena cava

The mean total operative time was 127 ±49.01 min. The mean ACC time was 63.7 ±25.72 min. The mean CPB time was 92.2 ±41.06 min. The mean CPB temperature was 25.8 ±5.23°c. Intraoperative complications were cardiac arrest on anaesthesia induction in 1 (3.7%) patient, open chest in 3 (11.11%) patients, LCOP in 5 (18.52%) patients, failed weaning from CPB in 6 (22.22%) patients which were admitted to ICU on pacemaker, lung congestion in 2 (7.41%) patients demonstrated by serosanguinous secretions in 2 (7.41%) patients, leaving the vertical vein open in 4 (14.81%) patients, myocardial stunning in 2 (7.41%) patients, pulmonary hypertensive crisis in 3 (11.11%) patients and haematuria 1 (3.7%) patient (**Table.4)**

Table 4. Total operative, ACC, CPB time, CPB temperature, intraoperative complications.

Variables		N=27
Total operative time (min)		127 ± 49.01
ACC time (min)		63.7 ± 25.72
CPB time (min)		92.2 ± 41.06
CPB temperature (°c)		25.8 ± 5.23
Intraoperative Complications	**Cardiac arrest on anesthesia induction**	1 (3.7%)
	Open chest	3 (11.11%)
	LCOP	5 (18.52%)
	Failed weaning from CPB (admitted to ICU on pacemaker)	6 (22.22%)
	Lung congestion	2 (7.41%)
	Leaving the vertical vein open	4 (14.81%)
	Myocardial stunning	2 (7.41%)
	Pulmonary hypertensive crisis	3 (11.11%)
	Hematuria	1 (3.7%)

Data are presented as mean± SD or frequency (%). ACC: Aortic Cross clamp, CPB: Cardiopulmonary bypass, LCOP: Low cardiac output, CPB: Cardiopulmonary bypass, ICU: Intensive care unit.

Mean arterial pressure (MAP) and respiratory rate were significantly lower postoperative than in preoperative (P value <0.05) while heart rate and oxygen saturation level were insignificantly different between preoperative and postoperative (**Fig.2**).

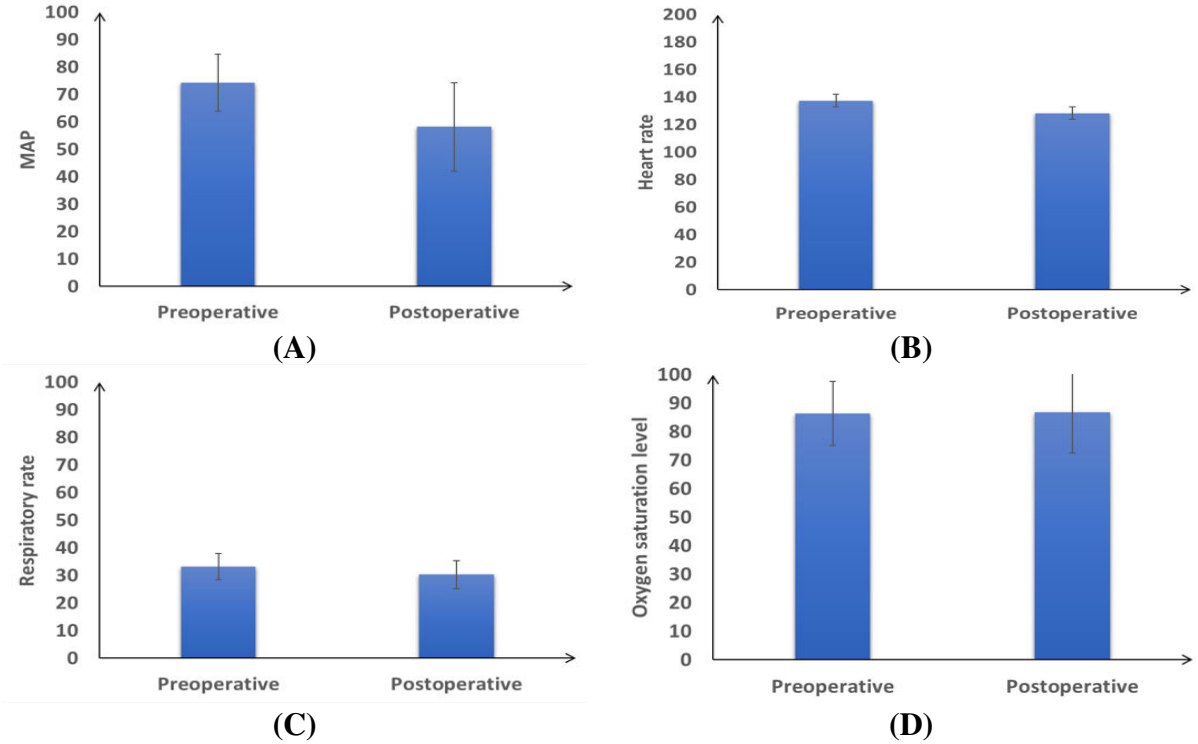

Fig.2. (A) MAP, (B) heart rate, (C) respiratory rate and (D) oxygen saturation level of the studied patients

The post operative oxygen delivery method was mechanical ventilation in 8 (29.63%) patients, and 18 (66.67%) patients did not need postoperative oxygen therapy. Postoperative pacemaker was present in 6 (22.22%) patients. The post operative ventilation time ranged from 2 to 333 hr. The mean length of ino-pressor drugs was 76.7 ± 98.84 hr. 26 (96.3%) patients had adrenaline. 4 (14.81%)

patients had noradrenaline. 8 (29.63%) patients had milrinone. 3 (11.11%) patients had dopamine. 17 (62.96%) patients had dobutamine. Postoperative complications were pneumothorax in 2 (7.41%) patients, pulmonary edema in 1 (3.7%) patient, re-intubation in 2 (7.41%) patients, pneumonia in 1 (3.7%) patient, fits in 2 (7.41%) patients, thrombocytopenia in 1 (3.7%) patient, bleeding from ETT in 1 (3.7%) patient, LCOP in 5 (18.52%) patients, pulmonary hypertensive crisis in 3 (11.11%) patients, cardiac arrest in 1 (3.7%) patient, renal failure in 1 (3.7%) patient and no complication in 14 (51.85%) patients. Re-opening occurred in 1 (3.7%) patient. Sepsis occurred in 2 (7.41%) patients. Neither re-operation, arrhythmias, nor surgical site infection occurred in any patients. The mean postoperative bleeding was 109.8 ±77.6 ml. The mean ICU stay was 4.4 ±4.04 days. The mean total hospital stay was 6.8 ±4.53 days. Mortality occurred in 9 (33.33%) patients. Causes of mortality were cardiac arrest in 1 (3.7%) patient, sepsis in 2 (7.41%) patients, LCOP in 4 (14.81%) patients and pulmonary hypertensive crisis in 2 (7.41%) patients (**Table.5)**

Table 5. Postoperative outcomes, mortality and causes of mortality of the studied patients

Variables		**N=26**
Postoperative oxygen delivery method	**Room air**	18 (66.67%)
	Mechanical ventilation	8 (29.63%)
Postoperative pacemaker		6 (22.22%)
Postoperative ventilation time (hr)		53.2 ± 73.22
Length of ino-pressor drugs (hr)		76.7 ± 98.84
Postoperative inotropic adrenaline		26 (96.3%)
Postoperative inotropic noradrenaline		4 (14.81%)
Postoperative inotropic milrinone		8 (29.63%)
Postoperative inotropic dopamine		3 (11.11%)
Postoperative inotropic dobutamine		17 (62.96%)
Postoperative complications	**Pneumothorax**	2 (7.41%)
	Pulmonary edema	1 (3.7%)
	Re-intubation	2 (7.41%)
	Pneumonia	1 (3.7%)
	Fits	2 (7.41%)
	Thrombocytopenia	1 (3.7%)
	Bleeding from ETT	1 (3.7%)
	LCOP	5 (18.52%)
	Pulmonary hypertensive crisis	3 (11.11%)
	Cardiac arrest	1 (3.7%)
	Renal failure	1 (3.7%)
	Re-opening	1 (3.7%)
	Sepsis	2 (7.41%)
	Arrhythmias	0 (0%)
	Surgical site infection	0 (0%)
	Re-operation	0 (0%)
Postoperative bleeding (ml)		109.8 ± 77.6
ICU stay (days)		4.4 ± 4.04
Total hospital stays (days)		6.8 ± 4.53
Mortality and causes of mortality (n=27)		
Mortality		9 (33.33%)
Causes of mortality	**Cardiac arrest**	1 (3.7%)

	Sepsis	2 (7.41%)
	LCOP	4 (14.81%)
	Pulmonary hypertensive crisis	2 (7.41%)

Data are presented as mean± SD or frequency (%). LCOP: Low cardiac output, ETT: Endotracheal Tube, ICU: Intensive care unit.

For the 18 patients discharged from hospital, postoperative echocardiography done on discharge revealed normal pulmonary pressure in 3 (16.67%) patients, and mild pulmonary hypertension in 15 (83.33%) patients. Tricuspid valve regurgitation was mild in 12 (66.66%) patients, and moderate in 6 (33.34%) patients. Pressure gradient across the anastomosis was within normal range in 16 (88.89%) patients, and mildly increased in 2 (11.11%) patients.

Follow up echocardiography was done at 3 months and 6 months postoperative. At 3 months, there was normal pulmonary pressure in 6 (35.29%) patients, and mild pulmonary hypertension in 11 (64.71%) patients. Tricuspid valve regurgitation was trivial in 5 (29.41%) patients, and mild in 12 (70.59%) patients. Pressure gradient across the anastomosis was within normal range in 16 (94.12%) patients, and mildly increased in 1 (5.88%) patient. One patient was lost to follow up. At 6 months, there was normal pulmonary pressure in 10 (58.82%) patients, and mild pulmonary hypertension in 7 (41.18%) patients. Tricuspid valve regurgitation was trivial in 6 (35.29%) patients, and mild in 11 (64.71%) patients. Pressure gradient across the anastomosis remained the same as the previous echocardiography. (**Table.6, Figs 3,4)**

Table 6. Post-operative echocardiography data of the studied patients.

Variables		**On discharge (n=18)**	**At 3 months postoperative (n=17)**	**At 6 months postoperative (n=17)**
PASP (mmHg)	**< 20**	3 (16.67%)	6 (35.29%)	10 (58.82%)
	20 – 35	15 (83.33%)	11 (64.71%)	7 (41.18%)
	36 – 55	0 (0%)	0 (0%)	0 (0%)
	> 55	0 (0%)	0 (0%)	0 (0%)
Degree of TR	**Trivial**	0 (0%)	5 (29.41%)	6 (35.29%)
	Mild	12 (66.66%)	12 (70.59%)	11 (64.71%)
	Moderate	6 (33.34%)	0 (0%)	0 (0%)
	Severe	0 (0%)	0 (0%)	0 (0%)
Pressure gradient across the anastomosis (mmHg)	**< 2.5**	16 (88.89%)	16 (94.12%)	16 (94.12%)
	2.5-5	2 (11.11%)	1 (5.88%)	1 (5.88%)
	6-10	0 (0%)	0 (0%)	0 (0%)
	>10	0 (0%)	0 (0%)	0 (0%)

PASP: pulmonary artery systolic pressure, TR: tricuspid regurgitation.

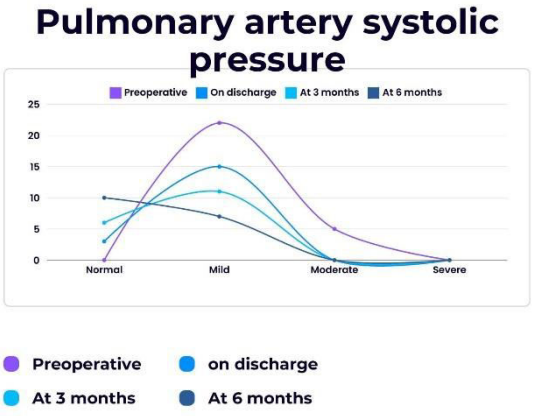

Fig.3. Comparison of pulmonary artery systolic pressure: preoperative and postoperative

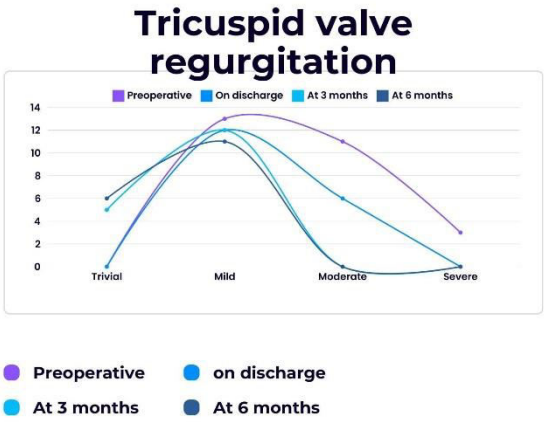

Fig.4. Comparison of the degree of tricuspid valve regurgitation: preoperative and postoperative

Discussion

TAPVC develops when the primordial PV fails to unite with the plexus of veins surrounding the lung buds. This cardiac malformation is classified according to the site of PV connection. TAPVC is supracardiac in about 45% of cases, cardiac in about 25%, infracardiac in about 25% and mixed in about 5% **(Seale et al., 2010)**.

In our study, mortality occurred in 9 (33.33%) patients. Causes of mortality were cardiac arrest in 1 (3.7%) patient, sepsis in 2 (7.41%) patients, LCOP in 4 (14.81%) patients and pulmonary hypertensive crisis in 2 (7.41%) patients. Preoperative ventilation was done in 1 (3.7%) patient. In accordance, Gholampour et al., reported that overall mortality was 33.8% in their series **(Gholampour et al., 2008)**. This is markedly higher than most reports, except the report of Korbmacher et al. presented the long-term results of TAPVC repair of 52 patients in 2001. Early mortality rate in their study was 34.6%. This may be due to presence of associated cardiac anomalies in our study. **(Korbmacher et al., 2001)**.

However, in cases where significant cardiac anomalies, such as SV heart physiology, are present, the mortality rate for TAPVC patients can reach as high as 41–52%. Patients with heterotaxy have a 79% five-year survival rate **(Zhao et al., 2015)**. In patients with SV heart physiology, Sugano et al., report a 5-year survival rate of 58%, and Nakayama et al., report a 10-year survival rate of 51% **(Nakayama et al., 2012; Sugano et al., 2019)**.

In our study, preoperative obstruction was present in 6 (22.22%) patients. Kogon et al., reported that risk factors for surgical mortality in mixed TAPVC included obstructive TAPVC **(Kogon et al., 2017)**. Seale et al., can speculate as to why preoperative obstruction has an influence on postoperative outcome **(Seale et al., 2010).**

In our study, the type of TAPVC was supracardiac in 21 (77.78%) patients, cardiac in 3 (11.11%) patients, mixed in 2 (7.41%) patients and infracardiac in 1 (3.7%) patient.

Jaworski et al., observed that the supracardiac subtype of TAPVC was related to worse outcomes than the other types of TAPVC **(Jaworski et al., 2022)**. However, as reported by Xiang et al., the results of the mixed type of TAPVC patient corrections are good, with a hospital mortality rate of 7.7% and a five-year survival rate of 991% **(Xiang et al., 2020)**.

In our study, the mean total operative was 127 ±49.01 min. The mean ACC time was 63.7 ±25.72 min. The mean CPB time was 92.2 ±41.06 min. The mean

CPB temperature was 25.8 ±5.23 °c. Prolonged ACC clamp time (> 60 minutes) had been associated with increased operative mortality **(Karamlou et al., 2007; Seale et al., 2010)**. Kogon et al., reported that risk factors for surgical mortality in mixed TAPVC included prolonged cardiopulmonary bypass time **(Kogon et al., 2017)**.

In our study, chest radiography revealed cardiomegaly in 23 (85.19%) patients, bilateral prominent bronchovascular markings in 14 (51.85%) patients and widened mediastinum in 6 (22.22%) patients. ECG revealed sinus rhythm in all patients. All patients did not have any preoperative inotropic support. Preoperative mechanical ventilation was needed in 1 (3.7%) patient. In our study, younger age below 2 months and lower body weight below 6 kg at time of surgery, presence of preoperative pulmonary venous obstruction, prolonged total operative time and CPB time were all associated with higher incidence of postoperative pulmonary hypertensive crisis. Hung et al., reported body weight below 6 kg, pneumonia, tachycardia, hepatomegaly, preoperative pulmonary congestion on chest x-ray, preoperative elevated mean PAP, preoperative PVO, emergency surgery and prolonged aortic cross-clamping time were significant associated with postoperative pulmonary hypertensive crisis **(Hung et al., 2021)**.

In our study, associated cardiac anomalies were secundum ASD in 20 (74.07%) patients, complete AVSD in 4 (14.81%) patients, DORV in 7 (25.93%) patients, right isomerism in 4 (14.81%) patients, pulmonary valve atresia in 1 (3.7%) patient, IVC draining into the LA in 1 (3.7%) patient, malposed GA in 1 (3.7%) patient, TGA in 2 (7.41%) patients, dilated hypertrophied RV in 1 (3.7%) patient, PFO in 1 (3.7%) patient and PDA in 2 (7.41%) patients. Associated extra cardiac anomalies were present in 5 (18.52%) patients. Padalino et al., reported that the presence of major cardiac anomalies has been confirmed to be a highly significant risk factor for mortality and morbidity in the postoperative period, which corresponds to our results, as out of mortality of nine patients (33.33%), four patients (14.81%) had associated significant cardiac anomalies such as DORV and TGA **(Padalino et al., 2014).**

In our study, intraoperative complications were cardiac arrest on anesthesia induction in 1 (3.7%) patient, open chest in 3 (11.11%) patients, LCOP in 5 (18.52%) patients, failed weaning from CPB in 6 (22.22%) patients which were admitted to ICU on pacemaker, lung congestion in 2 (7.41%) patients demonstrated by serosanguinous secretions, leaving the vertical vein open in 4 (14.81%) patients, myocardial stunning in 2 (7.41%) patients, pulmonary hypertensive crisis in 3 (11.11%) patients and hematuria 1 (3.7%) patient. Karaci et al., could not demonstrate any statistically significant difference between patients with a closed or opened vertical vein **(Karaci et al., 2012)**.

In our study, re-operation did not occur in any patients, while Jaworski et al., reported that four of their patients' required reoperations for recurrent PVO (5.7%) **(Jaworski et al., 2022)**.

Other studies showed that the need for reoperation due to recurrent PVO ranges between 4% and 17.5% **(Feng et al., 2020; Harada et al., 2019)**. Seale et al., reported that the rate of reoperation for isolated TAPVC is 5%–18% **(Seale et al., 2010).**

In our study, Arrhythmias did not occur in any patients. On the other hand, few reports describe cardiac rhythm in patients after TAPVC operations, in which small groups of patients are presented, but arrhythmia is found in almost half of them **(Emmel & Sreeram, 2004; Korbmacher et al., 2001)**. Jaworski et al., reported that our 40 patients monitored with a 24-h ECG, such an arrhythmia was found only in 13 children, who did not require therapy **(Jaworski et al., 2022).**

In our study, postoperative complications were pneumothorax in 2 (7.41%) patients, pulmonary edema in 1 (3.7%) patient, re-intubation in 2 (7.41%)

patients, pneumonia in 1 (3.7%) patient, fits in 2 (7.41%) patients, thrombocytopenia in 1 (3.7%) patient, bleeding from ETT in 1 (3.7%) patient, LCOP in 5 (18.52%) patients, pulmonary hypertensive crisis in 3 (11.11%) patients, cardiac arrest in 1 (3.7%) patient, renal failure in 1 (3.7%) patient and no complication in 14 (51.85%) patients. Re-opening occurred in 1 (3.7%) patient. Surgical site infection did not occur in any patients. Sepsis occurred in 2 (7.41%) patients. This concludes that in our study, the most common postoperative complications were LCOP and pulmonary hypertensive crisis. Jaswal et al., reported that the most common postoperative complications seen were pulmonary hypertensive crisis (28.6%) and sepsis (28.6%) **(Jaswal et al., 2021)**.

As the literature, the incidence of postoperative pulmonary hypertensive crisis varied depending on the specific population, estimated between 4% and 35% and the early mortality rate directly caused by pulmonary hypertensive crisis after surgery could be up to 16.7% **(Halpern et al., 1990)**. Hung et al., documented the incidence of postoperative pulmonary hypertensive crisis was 27.59% **(Hung et al., 2021).**

Limitations of this study included that relatively small sample size of 27 patients, limiting the generalizability of the findings. The study was conducted at a single institution, which may introduce institutional bias and restrict the variability of results. The study population comprised patients from a specific geographic area and may not represent the overall population with TAPVC. The study did not provide information on certain important variables, such as long-term follow-up outcomes and specific details of surgical techniques employed was relatively small.

Conclusion

The single-stage surgical repair of TAPVC showed a significant mortality rate of 33.33%. Causes of mortality included cardiac arrest, sepsis, low cardiac output syndrome (LCOP), and pulmonary hypertensive crisis. Intraoperative complications such as cardiac arrest on anesthesia induction, LCOP, failed weaning from CPB, and pulmonary hypertensive crisis were encountered. Postoperative complications included pneumothorax, pulmonary edema, re-intubation, pneumonia, fits, and renal failure. Re-operation and arrhythmias did not occur. The study highlights the high morbidity and mortality associated with TAPVC single-stage repair, emphasizing the need for careful preoperative evaluation and perioperative management.

Financial support and sponsorship: Nil

Conflict of Interest: Nil

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