

# A Retrospective Study of the Clinical Outcomes Following Surgical Correction of Supra Cardiac Total Anomalous Pulmonary Venous Connection

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**Abstract:** *Total anomalous pulmonary venous connection is a rare (1% of children born with congenital heart diseases), life threatening congenital cardiac anomaly. In this study we review and discuss the presentation, clinical features, investigations and factors, which influence the surgical outcome in patients with total anomalous pulmonary venous connection who underwent surgery at Madras Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019.*

**Keywords:** Total anomalous, pulmonary, congenital

## 1. Introduction

Total anomalous pulmonary venous connection is a rare (1% of children born with congenital heart diseases), life threatening congenital cardiac anomaly. It was first described by Wilson (1) in 1798. Friedlowsky in 1886 described it as a pathologic entity. But it was not until 1950 (2) that the first case was reported in which the clinical diagnosis was made, and this was accomplished through the use of cardiac catheterization. Muller (3) in 1951 was the first to repair it successfully. Since then there has been progressive improvement in operative results. However, the surgical mortality varies from 8% to 20% in most series (4, 5, 6). The mortality rate is higher in infants. The causes for the higher surgical mortality in this group may include pulmonary congestion, elevated pulmonary vascular resistance, and the critical condition of many infants at the time of operation. Thus early accurate diagnosis, prompt surgical intervention and careful preoperative and postoperative care are the key to success.

In this study we review and discuss the presentation, clinical features, investigations and factors, which influence the surgical outcome in patients with total anomalous pulmonary venous connection.

## 2. Aims and Objectives

To retrospectively evaluate presentation, clinical features, investigations and factors which influence the surgical outcome in patients with total anomalous pulmonary venous connection who underwent surgery at Madras Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019.

## 3. Methods and Materials

### Patient Population

22 patients who underwent surgery for supracardiac total anomalous pulmonary venous connection at Madras

Medical College and Rajiv Gandhi Govt General Hospital, between January 2009 and January 2019, formed the basis of this study. There were 13 patients below the age of 1 year. There were 18 males and 4 females. Patient weight at operation varied from 4kg to 57kg. In the patient group below 1-year weight at operation varied from 4kg to 7kg (mean, 4.76kg), while in the other group weight at operation varied from 15kg to 57kg (mean, 35.56kg).

A retrospective review of the clinical, surgical, echocardiographic and cardiac catheterization was done. Preoperative diagnoses were established by echocardiography in 15 patients and by cardiac catheterization in 4 patients and both were used in 3 patients.

### Surgical Technique

Median sternotomy was used in all patients. The thymus was excised partly or in whole to facilitate exposure during cannulation. Surgery was performed on cardio pulmonary bypass using bicaval cannulation and cold blood cardioplegia administered antegrade through the root. Moderate hypothermia was used in 9 patients and deep hypothermic circulatory arrest was used in 13 patients. Modified Schumaker and King technique was used in all.

The mean bypass time in the moderate hypothermic group was 103.11min, while in the group, which underwent deep hypothermic arrest was 93.92min. Mean duration of circulatory arrest was 46min. All patients were cooled to 18°C. All patients were weaned of cardiopulmonary bypass with inotropic support. The mean age at surgery was 100.59 months. Mean BSA was 0.64sq.m.

### Post Operative Management

This included prolonged elective ventilation. We used only pressure control mode of ventilation. We maintained partial pressure of carbon dioxide below 30mmHg. The duration of ventilation ranged between 1 and 8 days with a mean of 2.18 days.

**Weaning Protocol**

All patients were electively ventilated for a minimum of 24hours. If patients were haemodynamically stable weaning was started. Frusemide was given approximately 3 hours before extubation. They also received one dose of steroids prior to extubation.

Mean ICU stay was 3.45 days. The mean ICU stay among those who survived was 3.54 days. Survivors were discharged from the hospital after a mean interval of 9.23 days (range, 6 to 15 days).

**Echocardiographic and Cardiac Catheterisation Studies**

Echocardiography was done for 18 patients while cardiac catheterization was done for 6 patients. Most of our diagnosis was echo based.

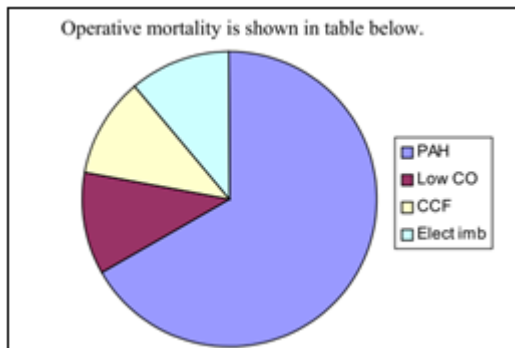
**4. Data Analysis**

The SPSS version 11 statistical program was used to analyze results. Chi square test was used for discrete variables.

**5. Results**

The operative mortality rate for isolated TAPVC was 40.9% (9 of 22 patients).

The major cause of death was pulmonary hypertensive crisis 67% (6 of 9 patients). The other causes of death were low cardiac output, congestive cardiac failure and electrolyte imbalance.



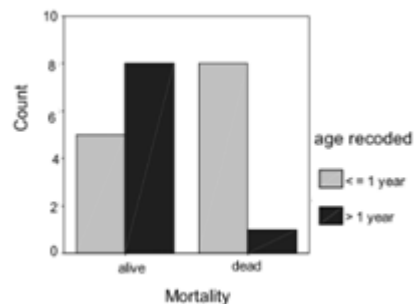
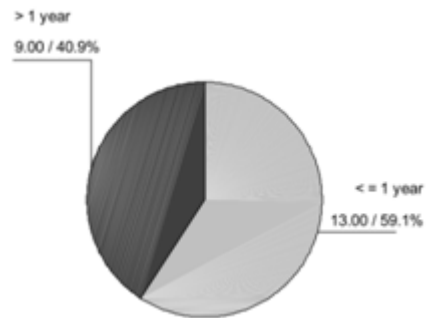
Cause of death	Number	Percentage
PAH	6	66.67
Low CO	1	11.11
CCF	1	11.11
Elect imbalance	1	11.11

Risk factors for early death were analyzed by univariate analysis. Age less than 12 months, weight less than 10kg, size of ASD less than 10mm, and preoperative failure were significant risk factors for postoperative mortality. Of the 22 patients, 13 patients were below the age of 1 year and 9 were above 1 year. Also there were 8 patients who were below the age of 12 months among the 9 patients who died. This was statistically significant (p=0.009).

Ranks

	Mortality	N	Mean Rank	Sum of Risks
Age in months	.00	13	14.42	187.50
	1.00	9	7.28	65.50
	Total	22		

p=0.009

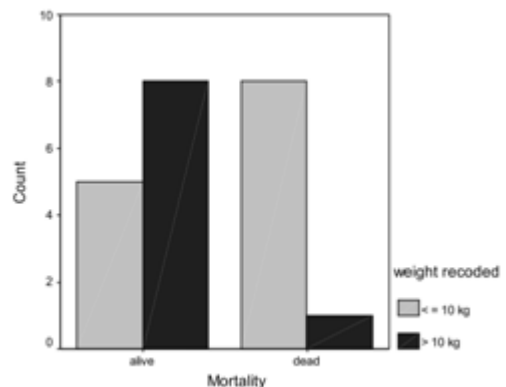


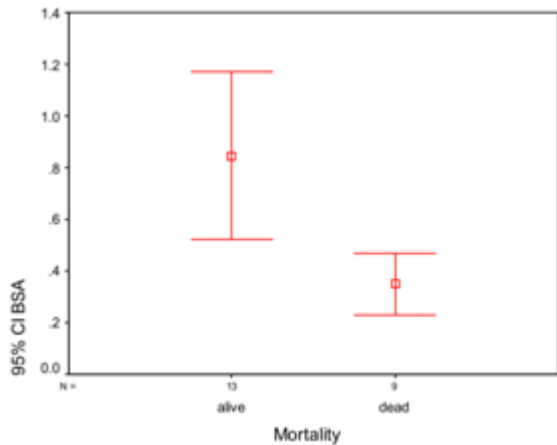
Among the 22 patients, 13 patients weighed less than 10kgs. Of the 9 patients who died, 8 weighed less than 10kg, which was statistically significant (p=0.021).

Ranks

	Mortality	N	Mean Rank	Sum of Risks
Weight in kgs	.00	13	14.12	183.50
	1.00	9	7.72	69.50
	Total	22		

P=0.021





From the graph depicting the 95% CI for BSA, those who died had a significantly lower BSA. The BSA for those who died ranged from 0.21 to 0.72 (mean 0.35), while for those who were alive it ranged from 0.22 to 1.65 (mean 0.84).

Sixteen patients were diagnosed to have clinical cyanosis. However there was an equal distribution between the two groups.

CYANOSIS\*Mortality Crosstabulation

		Mortality		Total
		.00	1.00	
CYANOSIS	0	5	1	6
	1	8	8	16
Total		13	9	22

p=0.157

CCF\*Mortality Crosstabulation

		Mortality		Total
		.00	1.00	
CCF	0	9	4	13
	1	84	5	9
Total		13	9	22

p=0.245

Nine patients were in CCF. Of these five patients died. The size of the liver was an estimate of the degree of the heart failure. The larger the size of the liver, the greater was the mortality. Statistically this was not significant. This could be attributed to the small sample size. However the trend was evident.

Liver\*Mortality Crosstabulation

		Mortality		Total
		.00	1.00	
Liver	0	8	3	11
	1	1		1
	2	1	1	2
	3	2	2	4
	4	1	2	3
	6		1	1
Total		13	9	22

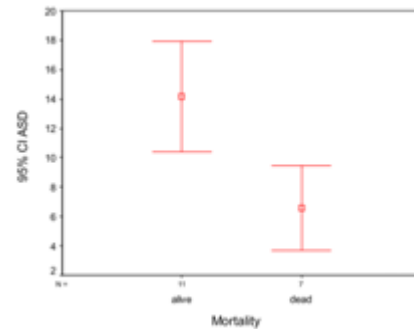
p=0.548

The size of the atrial septal defect had a significant bearing on the surgical outcome. Of the 9 patients who died, 7 had an ASD, which measured less than 10mm (p=0.004). The

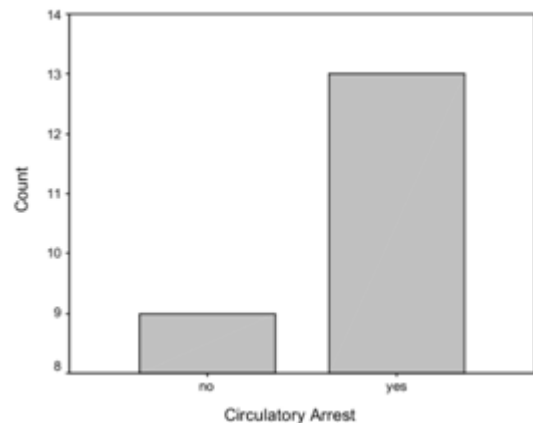
mean of the ASD among the mortality group was 5.21mm while that in the other group 12.23mm.

		N	Mean Rank	Sum of Ranks
ASD	.00	11	12.23	134.50
	1.00	7	5.21	366.50
Total		18		22

p=0.004



Thirteen patients were operated under deep hypothermic circulatory arrest of which six had an uneventful recovery. Nine were operated with moderate hypothermia of which seven were alive. There was no statistical significance between the two groups (p=0.138).



In the group, which underwent circulatory arrest, the total pump time ranged between 42min and 134min (mean 95.62min). The total cross clamp time ranged between 23min and 79min (mean 54.31min).

In the group, which was operated on with moderate hypothermia, the total pump time ranged between 60min and 156min (mean 100.67min). The total cross clamp time ranged between 41min and 53min (mean 49.67min).

All patients required postoperative inotropic support. The mean duration of ventilation was 2.18 days (range 1 to 8 days).

Follow-up ranged from 6 to 60 months. Postoperative follow up was not possible in 3 patients. Surviving patients had normal growth and were free of symptoms

## 6. Conclusion

Surgery for TAPVC still carries a high morality.

The conditions contributing to this are: 1. weight at surgery less than 10kgs 2. age at surgery less than 12 months 3. size of ASD less than 10mm 4. late recognition and referral for surgery. It is important therefore to identify patients with TAPVC early and subject them to a corrective procedure at the earliest.

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