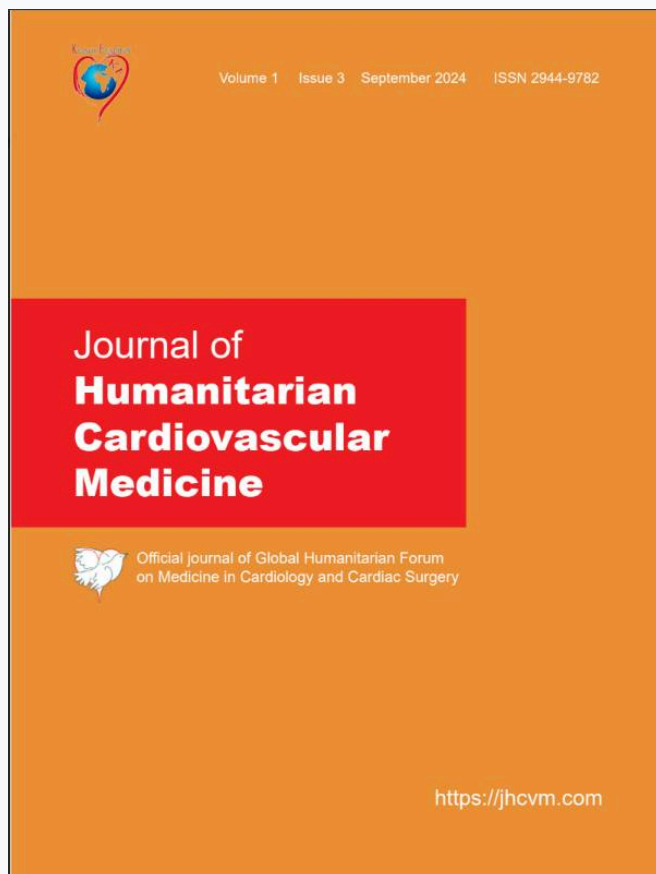


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# Total Anomalous Pulmonary Venous Return in Sulaimani, Iraq: A Retrospective Analysis of Pediatric Cases and Outcomes

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## Keywords

pulmonary, congenital, heart, anomalous, surgery

## ABSTRACT

**Background:** Total anomalous pulmonary venous return is a rare cyanotic congenital heart disease with bad outcomes without appropriate intervention. In which all four pulmonary veins drain incorrectly to the right atrium.

**Aim of study:** To determine the main morphological features and patient outcomes of isolated total anomalous pulmonary vein in pediatric heart diseases in Sulaimani Center.

**Patients and method:** A retrospective study including 46 patients aged from 15 days to 18 years has been performed in Department of Cardiology / Dr. Jamal Ahmad Rashid Teaching Hospital, Anwar Shekha hospital /HQ hospitals. The data were collected from the recorded files of the patients examined in that unit during the period from 2010 to 2022. Collected data include age, sex, family history, consanguinity, echo finding and outcome of the cases. Stoical analysis was done by SPSS21and P value of < 0.05 was regarded as significant.

**Results:** there are (45.7%) supra-cardiac, (45.7%) intra-cardiac and (8.6%) infra-cardiac. Male to female ratio is 1:1. (58.7%) of patients are associated with ASD, and (6.5%) have mixed ASD and VSD. Pulmonary hypertension is detected detects in (30.4%) of cases. Overall, (63.9%) of cases deceased and only (36.1%) are still alive. Median age of diagnosis for alive cases are 6.5 months (21 days to 21 months) and median age at repair are 15.6 months (one month to 10 years). Operation was conducted for (92.3%) of alive cases.

**Conclusion:** supra-cardiac and intra-cardiac are detected equally, infra-cardiac is the least common type and mixed type was not detected. ASD is the most common congenital heart defect associated with TAPVR. Only a third of cases survive, operation has been performed for most of them.

## Introduction

Total anomalous pulmonary venous return is a cardiac anomaly where the pulmonary veins do not return to the left atrium, but instead drain into the right atrium or its venous branches. There are four types of defects based on where the pulmonary veins drain: supra-cardiac, intro-cardiac, infra-cardiac, and mixed type. An atrial septal defect is necessary for survival, as it allows for the mixing of pulmonary and systemic venous blood in the right atrium. Patients with obstruction in the pulmonary venous return may experience systemic arterial desaturation. The level of systemic arterial oxygen saturation is dependent on the amount of pulmonary blood flow, with obstruction to pulmonary venous return leading to severe cyanosis.<sup>1,2</sup>

A study in 2012 using data from birth defects tracking systems across the United States, Researchers in United States expected that about 400 babies with total anomalous pulmonary venous return are delivered each year. In other words, about 1 in every 10,000 babies born in the United States are delivered with total anomalous pulmonary venous return.<sup>3</sup>

The latest information available on Total Anomalous Pulmonary Venous Return (TAPVR) suggests that about 504 babies are born with TAPVR each year in the United States, according to a 2019 study. This indicates that approximately 1 in every 7,809 babies born in the U.S. each year are affected by TAPVR.<sup>4</sup>

## Aim of the Study

To determine the main morphological features and patient outcomes of isolated total anomalous pulmonary vein in pediatric heart diseases in Sulaimani center Patient and method:

## Methodology

This is a retrospective study including 46 patients aged from 15 days to 18 years has been performed in cardiac unit / Dr. Jamal Ahmad Rashid teaching hospital, Anwar Shekha hospital /HQ hospital. The data were collected from the recorded files of the patients examined in that unit during the period from 2010 to 2022.

The study data included age, sex, family history, consanguinity, and echocardiographic morphological classification, which included (1- Supra-cardiac all drained to Superior Vena Cava, 2- Intra-cardiac directly drained to right atrium or coronary sinus and 3- Infra-cardiac).

The data also included the associated congenital cardiac defects with TAPVR, which were diagnosed by echocardiography. These defects included ASD, VSD, and PDA. As well as pulmonary hypertension, tricuspid incompetence, and small left side of heart in all patients were evaluated.

All cases included in this study were inquired by the same ECHO machine using Two dimensional, and Doppler (spectral and color) with 3V2C and 7V3C MHz transducers (adjusted according to examinations were obtained for each patient chest wall thickness) using a SIUI Apogee 3800 made in China, by the same examiner (pediatric cardiologist) and confirmed by either surgical team or visiting team. Transthoracic cross-sectional echocardiography always lets obvious demonstration of all intra cardiac anatomy. Echo was accomplished according to standard echo view from sub costal, apical, parasternal, and suprasternal recommended by the American Society of Echocardiography.<sup>5</sup>

you can use this version: Data such as **patient outcomes and follow-up**, operated and non-operated cases, median age of diagnosis (minimal; maximum), median age of death (minimal; maximum), median age at repair (minimal; maximum), and median current age of survival cases (minimal; maximum) were **obtained** through direct contact with patients or by phone call. Information was not **obtained from 10 patients** because they lost their **follow-up**.

Surgical repair was achieved on cardiopulmonary bypass with bicaval cannulation. In intra-cardiac TAPVR the coronary sinus was unroofed in the conventional repair but if the pulmonary veins connected with orifice stenosis the suture less repair (single- or 2-sided atriopericardial connecting) was helpful. For patients with supra-cardiac and infra-cardiac TAPVR, the suture less repair was functional. In this case, the incisions were created in the venous junction and then extend-

ed into both upper and lower pulmonary veins independently if there was unobstructed flow.

However, in small pulmonary venous confluences (i.e., smaller than the normalized mitral valve size diameter), such as those seen in infracardiac TAPVR, were more likely to need incisions in the separate pulmonary veins out toward the pleural pericardial reflection. The atrial septal defect or patent foramen oval was closed or partially closed.

All patient data **was** entered using computerized statistical software; , Statistical Package for Social Science (SPSS) Version 23 for **Windows**.

Chi-square test was used for categorical variables, P value of < 0.05 was regarded as significant.

## Results

Forty-six cases of total anomalous pulmonary

venous return, 21 patients (45.7%) were supracardiac, 21 patients (45.7%) were intracardiac and 4 patients (8.6%) were infracardiac. It's mean that both supra-cardiac and intra-cardiac were equal, infra-cardiac was the least common type of TAPVR and mixed type did not exist in the current study.

The most common associated congenital cardiac defects in patients with all types of TAPVR were ASD, in 46 cases, 27 cases had isolated ASD and 3 cases had mixed ADS and VSD. These details and other associated cardiac defects arranged in table 2

Pulmonary hypertension happened in 14 cases (30.4%). Tricuspid incompetence was found in 7 cases (15.2%) and 2 cases (4.3%) were associated with hypoplastic left side of heart as shown below.

**Table 1.** Descriptive feature regarding the morphological classification of TAPVR.

Variables	No.	drainage	NO.
Supracardiac	21 (45.7%)	Superior venacava	21
Intracardiac	21(45.7%)	Right atrium	8
		Coronary sinus	13
Infrocardiac	4(8.7%)		
Total	46 (100%)		

**Table 2.** Associated other congenital cardiac defects with TAPVR.

Variables	ASD	PFO	Mixed (ASD &VSD)	PDA
Supracardiac	14 (66.7%)	1 (4.8%)	--	1 (4.8)
Intracardiac	11 (52.5%)	--	2 (9.5%)	1 (4.8)
Infrocardiac	2 (50.0%)	--	1 (25.0%)	--
Total	27 (58.7%)	1 (2.2%)	3 (6.5%)	2 (4.3%)

**Table 3.** Frequency of pulmonary hypertension, tricuspid incompetence and small left side of heart in all types of TAPVR.

Variables	Pulmonary hypertension	Tripcuspid incompetance	Hypoplastic left side of heart
Supracardiac	5 (23.8%)	3 (14.3%)	--
Intracardiac	7 (33.3%)	3 (14.3%)	2 (9.5%)
Infrocardiac	2 (50.0%)	1(25.0%)	--
Total	14 (30.4%)	7 (15.2%)	2 (4.3%)

**Table 4.** Association between family history, consanguinity and TAPVR.

Variables	Supracardiac	Intracardiac	Infrocardiac	Total	P value	
Family history	Positive	6 37.5%	5 29.4%	1 33.3%	12 (36) 33.3%	0.0161
	Negative	10 62.5%	12 70.6%	2 66.7%	24 (36) 66.7%	
Consanguinity	Positive	8 50.0%	10 58.8%	1 33.3%	19 (36) 52.8%	0.0212
	Negative	8 50.0%	7 41.2%	2 66.7%	17 (36) 47.2%	

Note: Information could not be obtained from other 10 cases because they lost their follow up.

Ten cases have lost on **follow-up in Table 4** because information could not be obtained from there. In the 35 remainder cases, family history is positive in 11 cases (33.3%) and negative in 24 cases (66.7%). **Consanguinity is positive in 19 cases (52.8%) and negative in 16 cases (47.2%).**

Table 5 shows male to female ratio in all variables of TAPVR 1:1, 23 patients (50.0%) are males, and 23 patients (50.0%) are females.

Table (7) shows that 13 cases are still alive. Operation has been done for 12 cases (92.3%) and the other case is still alive without intervention. Median age of diagnosis is 6.5 months, median age during operation is 15.6 months and median current age is 55.6 months.

Table (8) demonstrates 23 cases deceased, surgery has not been done for 21 cases (91.3%) and the other two cases (8.7%) have been operated. Median age of diagnosis was 4.9 months and median age of death was 8.2 months.

**Table 5.** Male to female ratio.

Types	Female	Male
Supracardiac	10	11
Intracardiac	11	10
Infrocardiac	2	2
Total	23	23
Percentage	50.0%	50.0%
P value	0.0044	

In 36 patients, 23 patients (63.9%) died, and 13 patients (36.1%) are still alive.

**Table 6.** Outcome of cases.

Variables	Deceased cases	Percentage (%)	A live case	Percentage (%)
Supracardiac	9 (15)	60.0%	6 (15)	40.0%
Intracardiac	12 (18)	66.7%	6 (18)	33.3%
Infrocardiac	2 (3)	66.7%	1 (3)	33.3%
Total	23 (36)	63.9%	13 (36)	36.1%

Note: Ten cases have lost on follow up.

**Table 7.** Illustrates median age of diagnosis, median age during and median current age in operated and non-operated cases.

Variable	NO. of cases	Operated cases	Non operated cases	Median age of diagnosis (month)	Median age during operation (month)	Median of current age (month)
Supracardiac	6	5	1	3.4	4	42
Intracardiac	6	6	0	6.6	24.3	66.5
Infracardiac	1	1	0	21	21	60
Total	13	12 (92.3%)	1 (7.7%)	6.5	15.6	55.8
Note	P value=0.0000045			Between Current age and age of operation		
	P value=0.00223			Between age of diagnosis and age of operation		

**Table 8.** Median age of diagnosis and median age of death in operated and non-operated cases.

Variable	NO. of cases	Operated cases	Non operated cases	Median age of diagnosis (month)	Median age of death (month)
Supracardiac	9	1	8	8.7	10.8
Intracardiac	12	1	11	2.6	7.2
Infracardiac	2	0	2	1.35	2.5
Total	23	2(8.7%)	21(91.3%)	4.9	8.2
Note:	P value = 0.005 between the age of diagnosis and the age of death				

## Discussion

In the current study, the most common morphological types are supra-cardiac and intra-cardiac, both types occurred equally. There are 46 cases of TAPVR, 21 cases (45.7%) are supra-cardiac, 21 cases (45.7%) are intra-cardiac and 4 cases (8.6%) are infra-cardia. According to our data infra-cardiac is the least common type and the mixed type was not present. In supra-cardiac type, pulmonary veins connect to Superior Vena Cava in all cases. In intra-cardiac type, pulmonary veins drain directly to right atrium in 8 cases (38.1%) or drain to coronary sinus in other 13 cases (61.9%). According to the studies have been done in Taiwan and Toronto, the supra-cardiac was (42.3%, 54%), intra-cardiac was (39.8%, 26%), infra-cardiac was (12.8%, 19%) respectively<sup>6,7</sup>. In these two studies, the supra-cardiac was the most common types of TAPVR, but in this study, both supra-cardiac and intra-cardiac detected equally.

Regarding other Congenital heart defect associated with TAPVR patients, in 46 cases 27 cases (58.7%) have ASD, one case (2.2%) has PFO, mixed ASD and VSD are found in 3 cases (6.5%) and PDA is detected in 2 cases (4.3%). In the current study, overall, 33 cases (71.7%) are associated with CHD. The most common type of CHD is ASD, 27 cases (82%) have ASD alone and 3 cases (9%) have mixed ASD with VSD. In contrast, Ali et al. reported that in 47 patients only 15 patients (32%) had associated congenital heart defects, all of whom (100%) had a ventricular septal defect<sup>8</sup>. The most common CHD associated with TAPVR patients were ASD (38.2%) and PFO 13 (38.2%) as reported by AS et al. in Bangladesh.

In this study, 14 patients (30.4%) progressed pulmonary hypertension, 7 patients (15.2%) have tricuspid incompetence, and hypoplastic left side of heart is associated with 2 cases (4.3%). In contrast to this study, the researchers in Bangladesh detected Pulmonary hypertension developed in (91.2%) of cases<sup>9</sup>.

Regarding male to female ratio is 1:1 in the current study, 23 patients (50.0%) are male, and 23 patients (50.0%) are female. A study has been done in Boston, in 123 cases, 72 cases (59%)

were male, and 51 cases were female (41%)<sup>10</sup>. Min Fu et al. reported that in 78 cases, 41 cases (52%) were male, and 37 cases (48%) were female<sup>6</sup>.

Strong relation has been found between positive family history of other congenital heart disease and consanguinity with occurrence of TAPVR. family history of other congenital heart disease is positive in 12 out of (36) cases (33.3%) and negative in 24 out of (36) cases (66.7%). This is mean one-third of cases have positive family history of other congenital heart disease. Consanguinity is positive in 19 (36) cases which (52.8%) and negative in 16 (36) cases which is (47.2%). Moreover, more than half of cases have positive consanguinity between parents and most of them are first cousins. Other (10) cases, information could not be obtained from them. Nazari et al. in Iran reported the incidence of congenital heart disease among family pedigrees of patients with CHD (11.1%) and (48.7%) of patients' parents with CHD had positive consanguinity<sup>11</sup>.

Regarding the outcome of TAPVR cases, mortality is 23 out of (36) cases, (63.9%), 13 out of (36) cases (36.1%) are still alive, and 10 cases have not been followed up. Operation has been performed for 14 cases, 12 cases (85.7%) are alive, and 2 cases (14.3%) are deceased. M. Kelle et al mentioned only (21%) of cases were passed away<sup>12</sup>. Also, in 78 cases, three cases died before operation, and mortality rate was (9%) only in other 75 cases who went under operation as a study reported in Taiwan<sup>6</sup>.

As mentioned above, 13 cases (36.1%) are still alive, operation has been conducted for 12 cases (92.3%). One of the cases has been operated when she was 10 years old, and now she is 18 years old. Only one case (7.7%) lived without any intervention, now he is 18 years old. Median diagnostic age was 6.5 months range (21 days-21months), median age at repair was 15.6 months range (1months-10 years), and median current age 55.8 months range (8 months-18 years). 23 cases (63.9%) deceased, operation has been conducted only for two the patients (8.7%), and the other 21 patients deceased before operation. Median diagnostic age was 4.9

months range (15 days-26 months), and median age of death 8.2 months range (21 days-36 months). M. Kelle et al. researched in Chicago, that median age at repair was 14.6 days, and survival rate was (79%)<sup>12</sup>. Karamlou et al. has mentioned that median age during operation was 1.7 months and survival rate from repair was (70%) at 14 years<sup>13</sup>. Delay intervention and unavailability of surgical skills are the main causes of death in our hospital.

In the current study, due to delay in the diagnosis, there is not TAPVR associated with pulmonary venous obstruction. Therefore, the risky patients with obstruction deceased before the diagnosis was made. Also, that is the reason for the absence of the mixed type and small numbers of infra-cardiac type which are mostly associated with obstruction. Naturally, supra-cardiac is the most common type of TAPVR, and sometimes associated with PV obstruction; therefore, supra-cardiac and intra-cardiac equally occurred in the present study. Husain et al. researched in Texas, that (25.5%) of patients with TAPVR had PVO while (69.2%) cases were anatomically infra-cardiac, and (23%) of cases with obstruction were supra-cardiac<sup>14</sup>.

Regarding the 12 alive cases who have been operated, pulmonary venous obstruction has not been recorded in early and late post operative echocardiographic follow ups. Seale et al. in UK found out that (17.5%) postoperative cases had PVO, (83%) of patients was made diagnosis within 6 months<sup>15</sup> and a researcher has found out in Australia that the reoperation for PVO after primary repair was performed in (11.9%) of patients<sup>16</sup>.

### Conclusion and recommendation

- Supra-cardiac and intra-cardiac TAPVR occur with equal frequency, while infra-cardiac TAPVR is the least common, and mixed type TAPVR was not observed.
- Parental consanguinity (first cousins) is a significant risk factor, present in over half of the cases, and a family history of congenital heart defects is also prevalent.
- Atrial septal defect (ASD) is the most com-

mon co-occurring congenital heart defect in TAPVR patients.

- The majority of TAPVR cases (almost two-thirds) result in mortality, primarily in those who did not undergo surgical intervention.
- Surgical intervention significantly improves survival, with one-third of operated cases remaining alive.
- we recommend a further work up for early diagnosis and detection of TAPVR and more research to evaluate the prevalence of TAPVR in our community.
- we recommend early intervention and close follow ups in every case with TAPVR.

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### Conflict of Interest

The authors declare that they have no conflict of interest.

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