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A Retrospective Study of Morphological Types, Surgical Outcome and Follow up of Total Anomalous Pulmonary Venous Connection, at Our Institute

Medical Science

KEYWORDS : Total Anomalous Pulmonary Venous Connection (TAPVC)

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ABSTRACT

Total Anomalous Pulmonary Venous Connection was first, described by Wilson in 1798, where there is no direct communication between pulmonary veins and the left atrium. Rather all pulmonary veins are connected to right atrium or any of its tributaries. Only 1.5 - 3 % of all the Congenital Heart Diseases have TAPVC's. Atrial Septal Defect is necessary for survival after birth. The site where the pulmonary veins connect the systemic veins is usually obstructed, resulting in severe pulmonary hypertension. Patients are often critically ill especially if obstruction is present. The median survival rate for Supra-cardiac and Cardiac TAPVC's are 2.5 and 3 months respectively and Infra-cardiac have only 3 weeks of median survival. Surgical mortality is very high ranging from 2-20% with early mortality 12%. Hereby, we present our experience with TAPVC from 2007 to 2015 with aims to evaluate the morphology and surgical outcome pertaining to the anatomy.

Introduction:

Total Anomalous Pulmonary Venous Connection was first, described by Wilson in 1798, where there is no direct communication between pulmonary veins and the left atrium. Rather all pulmonary veins are connected to right atrium or any of its tributaries through a common chamber and a vertical vein. Only 1.5 - 3 % of all the Congenital Heart Diseases have TAPVC's[1,2,3,4,5].

Four types of TAPVC's are seen (Darling): Supra-cardiac (45%), Cardiac (25%), Infra-cardiac (25%) and Mixed (5-10%)[4]. In Supra-cardiac variety the vertical vein connects to innominate vein most commonly and less commonly to SVC. (**fig.1**)

In Cardiac variety the pulmonary veins directly open into coronary sinus or direct to right atrium.

In Infra-cardiac variety the vertical vein connects to Portal vein in 65% of cases[6] (Duff & colleagues), Ductus venosus, Gastric vein L-R Hepatic vein and IVC. Rarely 2 vertical veins are seen.

In Mixed variety the vertical vein most commonly the left upper lobe pulmonary vein forms a vertical vein and all the other pulmonary veins open into the coronary sinus. 2nd most common combination is the right lung pulmonary veins connect to coronary sinus and left lung pulmonary veins form vertical vein (Chowdhury & group)[3].

Pulmonary venous obstruction is severe, resulting from stenosis of vertical vein usually at the connecting site. In some cases of Supra-cardiac variety the left vertical vein may pass posterior to left pulmonary artery and thus compressed between LPA and left main bronchus. In the Cardiac variety the obstruction is at the pulmonary veins and coronary sinus junction and rarely at the coronary sinus ostium.

Infracardiac variety shows obstruction at diaphragm or prtal vein or ductus venosus level. Resistance at portal sinusoids offers additional obstruction. All infra cardiac patients and those connecting to azygous vein have obstruction[5].

65% patients of those to superior vena cava and 40% of those to innominate vein have obstruction. 40% patients in mixed types and 20% of those in cardiac type which is at coronary sinus level though rarely seen[7].

Patients have tachypnea, mild cyanosis, and recurrent episodes of pulmonary venous congestion, failure to thrive or hepatomegaly. Chest X ray shows classical snowman or Figure of Eight sign. 2D-Echo, cardiac catheterization and cardiac CT is diagnostic and tells about the anatomy and the connection of the vertical vein.

For survival always a connection between systemic and pulmonary circulation is must to these patients. Almost all of must have ASD or patent foramen ovale

Only 50 % Neonates survive beyond 3 months. Infants have generally unfavourable prognosis with only 20% surviving the first year of life[11].

In Supra-cardiac/Cardiac TAPVC the median survival is 2.5 and 3 months respectively[11].

In Infra-cardiac the median survival is only 3 weeks[11].

Thus, the presence of TAPVC warrants urgent intervention in the form of Intra Cardiac Repair. But as the child is usually critically ill the Mortality rate is high and ranging from 2-20% with early mortality 12%.[12, 17, 14, 7, 15]

Hence, we have done this retrospective study of the patients presenting with TAPVC, in regards to their morphological types, the surgical out come from 2007 to 2015 at our institute.

Aims and objectives:

1. To evaluate the morphological types of TAPVC's presented at our institute.

2. To evaluate the surgical outcome at our institute, a municipal based set up.
3. To evaluate the late complications after Intra-cardiac repair.

Material and methods:
Retrospective data of all the patients of TAPVC's presented and operated at our institute from the period of 2007 to 2015 were studied. The morphological type and the surgical approach were noted. All the patients intra-operative and the post operative findings were recorded including the CPB data and immediate post operative ICU vitals.

LA pressure monitoring was done for first 48 hours in all patients. Patients were ventilated for at least 12 hrs with PCO2 kept low 25-30 mmHg by hyperventilation and acidosis was treated vigorously. Nitric oxide was not used as we don't have the facility. Every patient was started on adrenaline, milrinone, calcium and Lasix drip.

All the patients were evaluated post operatively using 2D-Echo and if required CT.

Results:
Out of 130 cases of TAPVC, 112(86.15%) cases were pediatric and 18(13.85%) cases were adult age group (> 12 yrs) ranging from 1.5 months to 37 yrs. 47(36.15%) patients were below 6 months. 84 cases were male and the 46 were female indicating the male dominance. The mean age was 45 months and the mean weight was 10.35 kgs.

The various anatomical types seen were[4]: (**chart 1**)

Supra-cardiac TAPVC – 69 (53.08%)

Cardiac TAPVC- 34 (26.15%)

Mixed TAPVC- 23 (17.69%)

Infra-cardiac TAPVC- 4 (3.07%) all being Infra-diaphragmatic variety

Out of the total 112 pediatric patients, 74 (66.08% of pediatric age) were males and 38 (33.92% of pediatric age) females with mean age of 15.12 months and the mean weight of 6.6 kgs. 47 (36.15%) patients were below 6 months with the mean weight of 3.4 kgs. 9 (7%) children were weighing less than 3 kgs.

The various anatomic types were as follows: (**chart 2**)

Supra-cardiac -56 (50% of pediatric),

Cardiac -29 (25.90% of pediatric)

Mixed - 23 (20.53% of pediatric)

Infra-cardiac - 4 (3.57% of pediatric)

Out of 18 adults, 10 were male and 2 were female patients. Supra-cardiac TAPVC were 13 and Cardiac TAPVC were 5 with mean age of 19.3 yrs and mean weight of 35.6 kgs. (**Chart 3**)

Out of the 69 cases of supra-cardiac TAPVC's, 52 (75.36%) cases had Vertical Vein opening into the Innominate vein and 14 (20.28%) cases into the SVC passing posterior to the LPA and 3 (4.34%) cases into the Azygous vein.

Table 1:

Supra-cardiac TAPVC (69 cases)		Cardiac TAPVC (34 cases)		Mixed TAPVC (23 cases)	
Innominate vein	52 (75.36%)	Coronary sinus	15 (44.11%)	Supra and cardiac variety	19 (82.63%)
SVC	14 (20.28%)	RA	6 (17.64%)	Supra and infra cardiac variety	4 (17.39%)
Azygous vein	3 (4.34%)	RA-SVC junction	5 (14.70%)		
		LA roof	3 (8.82%)		

Out of 34 cases of cardiac TAPVC's, 15 (44.11%) cases had pulmonary veins opening into the coronary sinus, 6 (17.64%) into the Right Atrium directly, 5 (14.70%) at the junction of RA and SVC, 3 (8.82%) opening into the LA roof directly.

Out of the 23 mixed variety, 19 patients had supra-cardiac and cardiac type with 10 patients having left pulmonary veins draining into innominate through vertical vein and right sided veins into the RA. Were as 9 patients had supra cardiac and infra diaphragmatic variety which is the rare finding with left into portal vein and right sided into RA and RA- SVC junction.

Associated anomalies:
15% cases (20 cases) had associated PDA. 2 (1.53%) patients had uni-ventricular physiology were Bidirectional Glenn shunt was done. PAPVC was seen in almost 15 (11.53%) cases. 2 (1.53%) patients had common atrium. 1(0.76%) patient had associated TOF. Left SVC was seen in almost 25% i.e 33 cases.

Table 2:

Associated anomaly	No of cases
Left SVC	33 (25%)
PDA	20 (15%)
PAPVC	15 (11.53%)
Common Atrium	2 (1.53%)
Uni-Ventricular	2 (1.53%)
TOF	1 (0.76%)

Intra-operatively:
The mean CPB time was 2.8 hrs with mean cross clamp time of 55 min under moderate hypothermia usually cooled upto 24^0c and 3 neonates required circulatory arrest. Earlier we used St Thomas 2 cardioplegia solution but since 2012 we started using del-Nido cardioplegia solution thus helping us in reducing the clamp time and repeated cardioplegia. Modified ultrafilteration was done in all pediatric patients.

Almost all the Supracardiac variety were operated through the retro-cardiac approach and only 10 cases were done through the Superior/LA roof approach, but was later not used due to limited exposure. 10 cases of the cardiac variety were operated by Van Praagh method. Mixed TAPVC's were dealt as per the anatomical findings with RA and retro-cardiac approach. 2 patients required Warden's procedure. All infra-cardiac cases were approached through retro-cardiac route. 2 needed BDG, 1 needed ICR for TOF. PDA was ligated in all patients who had.

ASD was closed using pericardial patch in all the patients. All the pediatric patients and the 10 adults were LA pressures and PA pressure were high the vertical vein was kept open.8 adult pa-

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tients were pa pressures were less than half of systemic pressures were ligated.

Post operatively:
10 (7.65%) patients succumbed to death immediate post operatively.

2 were cases of Infra-diaphragmatic TAPVC almost about 50% cases of the total Infra-diaphragmatic variety, 3 cases of mixed consisting of Infra-diaphragmatic variety mainly and 3 cases of supra-cardiac variety.

8 (6.1%) were of pediatric age group, 5 (62.5% of pediatric deaths) of which were < 3 kgs and 3 were > 3 kgs. All the deaths were due to repeated episode of Pulmonary Hypertensive crises.

2 were of adult age group both being supra-cardiac variety. Post mortem revealed irreversible (grade IV) changes of pulmonary artery hypertension.

Patients were assessed immediately after 48 hrs and at the time of discharge around 10-15th day with 2D-Echo and thereafter 6 monthly. Were assessed for
- LA size
- Pulmonary pressures
- Pulmonary venous and LA anastomotic site
- Pulmonary veins
- LV and RV function.

Almost all patients had mild or normal pulmonary pressures. LA size was normal or increased in size mostly due to the incorporation of pulmonary venous sinus or the coronary sinus. LV was normal or slightly increased in dimensions due to the improved LVEF, which was secondary to the improvement in the RV geometry after the ICR. PA pressure normalizes

8 patients mainly of supra-cardiac variety had anastomotic stenosis at the 6 months follow up and were advised re-operation. Of which only 2 patients turned out for surgery with fatal outcome.

25 patients came for regular follow ups for 3 years and most of the remaining patients were lost to follow up.

Discussion:
For survival always a connection between systemic and pulmonary circulation is must to these patients. Almost all of must have ASD or patent foramen ovale. Most patients have no associated anomalies or have small / large PDA in 15% cases. Some patients may have associated VSD, TOF, DORV or interrupted Aortic arch.

If SpO2 is <80% then Qp/Qs is <1.4 and pulmonary resistance is > 10u.m2[20].

Hypoplasia of small pulmonary artery has been recently identified in obstructive TAPVC (M1). Left atrial volume is 53% of normal predicted[8]. (Matthew and colleagues)

Infants have marked pulmonary hypertension with structural changes in lungs and pulmonary arteries and veins[9] are often critically ill especially if obstruction is present.

Risk factors for death :
1. Age and weight at time of surgery
2. Infra-diaphragmatic
3. Pulmonary venous obstruction
4. Poor preoperative physiologic state [3, 16, 17, 18, 15.]
5. Morphology of heart/anatomy/associated anomalies (single ventricle)
6. Severe acidosis
7. In older patients, pulmonary vascular resistance increases the risk

Mortality usually ranges from 2-20% [12, 17, 14, 7, 15] with early mortality of 12% this is comparable with our study which shows mortality rate of 7.65% which are mainly due to PH crisis.

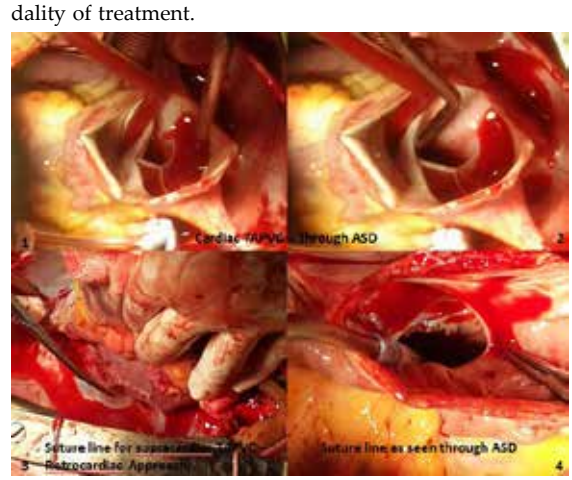
The mortality in Infants with normal weight appears to be 9.9% [19] were as our study showed 6.1% mortality in pediatric age group of which almost 62.5% infants were below 3 kgs as compared to the standard other studies were Infants with weight <2.5 Kg was 29% [19].

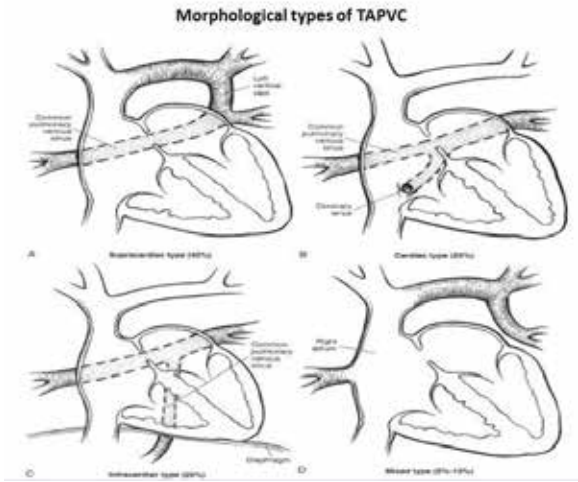
1 month survival was usually 90 % and 3 years survival was 87%, which was also seen in our study. Chowdhury demonstrated 4.3 % mortality in Mixed TAPVC with 42 months follow up. Delayed development of anastomotic stenosis (half of the cases) and pulmonary vein stenosis (one third cases) or due to both (remaining one third) requiring re-operation[14, 7.]

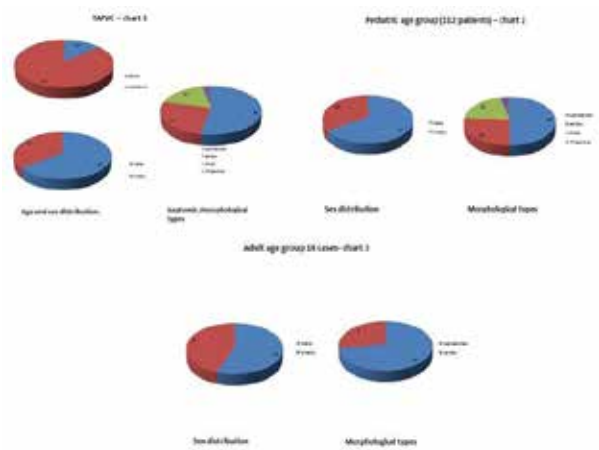
Pulmonary vein stenosis a Lethal complication (less common)[20] arises due to diffuse fibrosis and thickening of the vein wall and often to localized narrowing at vein – left atrial junction (suture less technique (Lacour Gayet 1996)[15,21,22,23,24.]

So, even though the mortality were comparably high due to the critical pre operative state of the patients the surgery results are excellent. Thus, increasing the survival rate almost upto 90% at 1 month and 87% at 3 years respectively. Surgery for TAPVC offers an excellent survival rate and we highly recommend early surgery in all the patients as the median survival rates are very low if not operated.

Conclusion:
To conclude with, our study showed the usual trends as seen in the standard western studies with the mortality rates as comparable. The need of expertise and well equipped ICU care is mandatory in all the patients. Even though the need of Nitric Oxide treatment is boosted, claiming increased effectiveness and increased survival rate by reducing the pulmonary hypertensive crisis in immediate post operative period, we could get the excellent results. Infants below 3 kgs still have a high mortality as they are critically ill pre-operatively and these patients may need nitric oxide treatment for the reduction in their mortality. Hence, we are looking forward for acquiring this modality of treatment.







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