

## Total Anomalous Pulmonary Venous Connection (TAPVC) - A Case Report

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### ABSTRACT

A one year and three months male child having Total Anomalous Pulmonary Venous Connection (TAPVC) presented at Ibrahim Cardiac Hospital and Research Institute with repeated cough, tachypnoea, fever, failure to thrive & diaphoresis particularly during feeding with a mild degree of cyanosis without any evidence of heart failure. He underwent successful rerouting of pulmonary veins to left atrium (LA). The procedure consisted of side to side wide anastomosis between confluence of pulmonary veins (CPV) and left atrium with the division and ligation of vertical vein at its opening to left innominate vein and closure of atrial septal defect (ASD). The patient is doing well without any symptoms at followup.

**Key words:** TAPVC, CPV, Vertical vein

### Introduction

Total Anomalous Pulmonary Venous Connection (TAPVC) is a rare congenital anomaly in which all four pulmonary veins have no direct communication with the left atrium. Instead they drain abnormally to the right atrium or to one of the systemic veins by way of an anomalous connection.<sup>1,2</sup> TAPVC accounts for 1% of all congenital heart defects and supracardiac variety accounts for 50% of all TAPVC patients.<sup>1-3</sup>

Developmentally, TAPVC occurs when the evaginated common pulmonary vein from the posterior surface of left atrium fails to fuse with the pulmonary venous plexus surrounding the lung buds.<sup>1,3</sup> Depending on the drainage site of the pulmonary veins, the defects may be divided into (i) Supracardiac (50%)-when pulmonary veins open into left innominate vein and superior vena cava (SVC) (ii) Cardiac (28%)-when they open into coronary sinus (iii) Infracardiac (14%)-when they open into portal vein and (iv) Mixed (7%) variety.<sup>1-3</sup> For sustaining life, TAPVC requires some connection between right and left heart through PFO/ASD.<sup>3</sup>

**Two clinical patterns:** (a) The obstructive type, characterized by an obstruction to pulmonary venous (PVO) return to the right atrium, leading to cyanosis with early pulmonary oedema and congestive cardiac failure. Death ensues within 3 months of life.<sup>4</sup> (b) The non-obstructive type (non-PVO)-characterized by delayed

manifestation of symptoms with failure to thrive, growth retardation and recurrent respiratory tract infections. Death occurs in 70% of cases within a year.<sup>4</sup> PVO occurs most often with the infracardiac type and less often in the intracardiac type of TAPVC. With elevated pulmonary arterial pressure (PAP) an intracardiac shunt should be of right to left direction to induce more cyanosis.<sup>4</sup> Advent of 2-dimensional (2-D) color Doppler echocardiography,<sup>5</sup> CT angiography<sup>6</sup> and magnetic resonance imaging (MRI) have made TAPVC diagnosis easier.<sup>7</sup>

### Clinical presentation

A one year and three months old male child was admitted to Ibrahim Cardiac Hospital and Research Institute for elective operation of TAPVC with past history of repeated respiratory tract infections, failure to thrive with mild cyanosis since birth. Cardiac auscultation revealed soft systolic murmur at 2<sup>nd</sup> intercostal space in left parasternal area, S<sub>2</sub> is widely split and fixed with loud P<sub>2</sub>. The Electrocardiogram showed right atrio-ventricular hypertrophy. The Chest X-Ray demonstrated cardiomegaly with figure of eight appearance and increased pulmonary vascular markings (Fig. 1). Haematological parameters were within normal limits.

Echocardiography demonstrated dilated right atrium, right ventricle and main pulmonary artery and hugely dilated SVC, high up secundum atrial septal defect with

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right to left shunt and severe pulmonary hypertension. Pulmonary veins from both lungs made a confluence of pulmonary veins (CPV) posterior to left atrium that was smallish. A vertical vein went up from CPV to left innominate vein and subsequently into SVC that drained

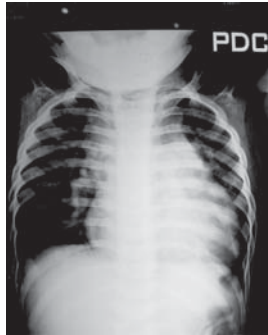


Fig. 1: X-Ray Chest P/A view showing cardiac silhouette as figure of eight.

into right atrium without any obstruction. CT pulmonary angiogram showed supracardiac type of TAPVC (Fig. 2 and 3) with ASD, dilated RA and RV.

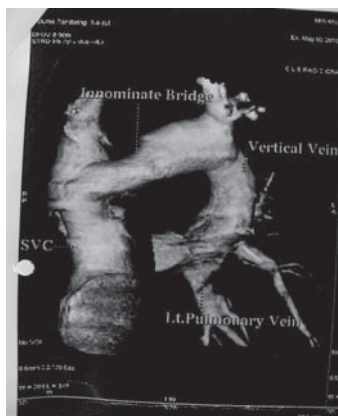


Fig. 2: CT angiogram demonstrates confluence of pulmonary veins, Left pulmonary vein, vertical vein, wide left innominate vein and a large SVC.

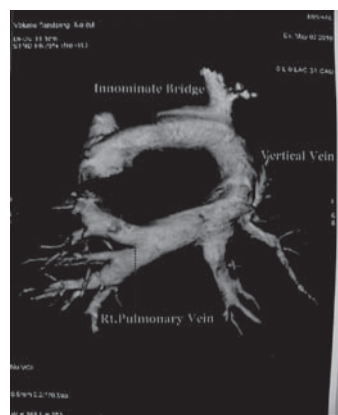


Fig. 3: CT angiogram showing lobar pulmonary veins, right & left pulmonary veins, common pulmonary veins, vertical vein, innominate vein.

Cardiopulmonary bypass (CPB) was established through aortic and bicaval cannulation. The CPV and the vertical vein were dissected from the surrounding structures. Under total circulatory arrest (TCA) at 22°C, the CPV and the LA were incised (from the interatrial septum to the base of left atrial appendage) transversely in equal length of 2.5 cm. and anastomosed side to side with each other to make them a single chamber. The Vertical Vein was ligated and divided from the Lt. innominate vein. The ASD was closed through a separate right atriotomy. Postoperative recovery was prolonged to some extent, due to left ventricular dysfunction that responded to ACE-inhibitor. Routine follow up at 1 month and 3 months interval showed that the condition of the patient was satisfactory.

## Discussion

The clinical symptoms and signs in TAPVC are variable depending on the pathological anatomy and the haemodynamics. Patients with obstructive type of TAPVC (with PVO) usually present with tachypnoea, tachycardia, difficulty feeding and cyanosis and even sudden death within the first 2-3 months after birth.<sup>4,8-9</sup> While those with non-obstructive type (without PVO) tend to present with failure to thrive, growth retardation, recurrent respiratory tract infections and heart failure later. Without proper surgery or treatment the prognosis of these patients is grim in the 1st year of life.<sup>4,10</sup>

TAPVC is either an isolated defect with small ASD or PFO or is a non isolated anomaly which is associated with various complex cardiac anomalies, such as Tetralogy of Fallot, common A-V canal defect, pulmonary stenosis or pulmonary atresia, mitral atresia, conotruncal anomalies, systemic venous anomalies or heterotaxy syndrome.<sup>2,4</sup> With the advent of 2-D and color Doppler echocardiography, TAPVC can readily be diagnosed without any difficulty and the procedure is noninvasive and safe. Now a days, it is the modality of choice for diagnosis of complex congenital cardiac diseases.<sup>4</sup> The sensitivity and specificity for diagnosis by echocardiography including cross-sectional and color Doppler flow mapping have been reported to be up to 97% and 99% respectively.<sup>11</sup>

Diagnostic Cardiac Catheterization and angiocardiology is the gold standard for diagnosis of TAPVC. But the procedure being invasive and time consuming, the technique should be kept reserved for the patients who have coexisting complex heart lesions or mixed connection of pulmonary veins.<sup>4</sup> Now a days, CT angiogram and cardiac MRI are detailed imaging producer to correctly detect anomalous pulmonary venous

channels in 98% of cases<sup>7</sup>. But these procedures are supplementary if echocardiography and angiography are inadequate.<sup>4</sup>

The natural course of obstructive type of TAPVC is unfavorable because of progressing pulmonary artery hypertension and heart failure.<sup>4</sup> Our case had the features of non-obstructive type of TAPVC with severe pulmonary hypertension at the age of one year and three months with mild cyanosis and stunted growth. The only option of treatment was open heart repair of the anomaly to prevent progressive pulmonary hypertension and heart failure. That is why ligation and division of vertical vein and wide anastomosis between CPV and left atrium was done.

The estimated first year survival rate in patients with TAPVC is only 25% without treatment; 50% death occurs before 3 months of age and 80% death occurs before 1 year of age.<sup>12</sup> The operative mortality in patients under 1-year of age decreased significantly from 50%<sup>13</sup> in 1970s to 10-20% after 1970s.<sup>14</sup> However, some studies still report a higher operative mortality in TAPVC patients with PVO where surgery was done urgently<sup>15</sup> but more recent reports have described an early mortality rate of less than 10%.<sup>2</sup> In TAPVC without PVO surgical redirection can be performed within first month of life.

Hancock *et al*<sup>16</sup> reported that re-operation of post operative pulmonary veins stenosis in univentricular heart was 18% which was significantly high compared with 8% in the biventricular group. Improvement in surgical and suture techniques have brought improvement to anastomotic site stenosis but histologic anomaly in the form of an obliterative intimal fibrous hyperplasia near to the CPV and away from the anastomotic site of an individual pulmonary vein considerably increases the risk of postoperative individual vein stenosis.<sup>17</sup> The size of the pulmonary veins is an important determinant of outcome in patients with TAPVC.<sup>2</sup>

### Conclusion

There is marked improvement in surgical results for isolated TAPVC with the improvement of preoperative diagnosis, treatment and surgical technique. However, surgical correction of TAPVC combined with univentricular heart still carries a high risk with mortality. In particular, concomitant palliation of the pulmonary artery at early age and low weight at the time of surgery are significant risk factors.

### References

1. Sabiston DC. Jr, Spencer FC. Total anomalous pulmonary venous connection. In: Surgery of the Chest, edited by Edward HW Jr 5th Edition, 1990, Vol-II, Published by W.B. Saunders Company, Harcourt Brace Jovonovich, Inc., Philadelphia. PA19106.3399 p-1278-89
2. Castaneda AR, Jonas RA, Mayer JM, Hanley FL. Anomalies of the Pulmonary Veins In: Paediatric cardiac surgery of the neonate and infant. Philadelphia :WB Saunders co, 1994 p-
3. Myung KP. Total anomalous pulmonary venous return. In: Paediatric Cardiology for Practitioners. 4th Edition, 2002, Mosby Inc., An imprint of Elsevier science, 11830 Westline Industrial drive, St. Louis, Missouri 63146, p-202-2
4. Ping YW, Be TH, Jen HL et al. Significance of Pulmonary Venous Obstruction in TAPVR. J Chin Med Assoc 2004; 67: 331-335
5. Smallhorn JF, Sutherland GR, Tommasini G, Hunter S, Anderson RH, Macartney FJ. Assessment of total anomalous pulmonary venous connection by two-dimensional echocardiography. Br Heart J 1981; 46:613.
6. Chen YC, Hsieh KS, Chi CS. Total anomalous pulmonary venous return: a clinical observation of 36 cases. J Chin Med Assoc 1994; 54:44-50.
7. Chang YC, Li YW, Liu HM, Wang TC. Finding of anomalous pulmonary venous return using MRI. J Formos Med Assoc 1994; 93:462-8.
8. Gathman GE, Nadas AS. Total anomalous pulmonary venous connection: clinical and physiologic observations of 75 pediatric patients. Circulation 1970; 42:143-54.
9. Byard RW, Moore L. Total anomalous pulmonary venous drainage and sudden death in infancy. Forensic Science International 1991; 51:197-202.
10. Lucas RV Jr, Adams P Jr, Anderson RC. Total anomalous pulmonary venous connection to the portal venous system: a case of pulmonary venous obstruction. Am J Roentgenol 1961; 86:561-75.
11. Huhta JC, Gutgesell HP, Nihill MR. Cross section echocardiographic diagnosis of total anomalous pulmonary venous connection. Br Heart J 1985; 53:525-34.
12. Keith JD, Rowe RD, Vlad P. Complete anomalous pulmonary venous drainage. Am J Med 1954; 16:23-38.
13. Behrendt DM, Aberteen E, Waterson DJ et al. TAPVD in infants. Clinical and haemodynamic findings, method and results of operation in 37 cases. Circulation 1972; 46: 347
14. Bonchek LI, Anderson RP, Wood JA, Chapman RD, Starr A. Intracardiac surgery with extracorporeal circulation in infants. Ann Thorac Surg 1974; 17:280-95.
15. Galloway AC, Campbell DN, Clarke DR. The value of early repair for total pulmonary venous drainage. Pediatr Cardiol 1985; 6:77-82.
16. Hancock Friesen CI, Zurakowski D, Thiagarajan RR et al. TAPVC- an analysis of current management strategies in a single Institution. Am Thorac Surg 2005;79:606
17. Yamaki S, Tsumemoto M, Shimada M et al. Quantitative analysis of pulmonary vascular disease in TAPVC in 60 infants. J Thorac Cardiovasc Surg 1992; 104: 728-35.