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# Successful Surgical Intervention of Total Anomalous Pulmonary Venous Drainage in the Third Decade of Life

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## Abstract

Total anomalous pulmonary venous drainage (TAPVD) accounts for approximately 1.5% of all congenital heart diseases. It is usually diagnosed in the neonatal period and is rarely seen in adults. We report an unusual case of a patient with TAPVD who was successfully treated at the age of 28 years. We believe that this is the oldest person in the South Asian literature to undergo surgical correction of TAPVD.

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## Introduction

Total anomalous pulmonary venous drainage (TAPVD) accounts for approximately 1.5% of all congenital heart diseases,<sup>1</sup> and the occurrence of atrial septal defect (ASD) is usually associated. It is usually diagnosed in the neonatal period and is rarely seen in adults.<sup>2</sup>

A classification of TAPVD has been proposed by Darling *et al.*,<sup>3</sup> who has divided it into four types: supra-cardiac (45%); infra-cardiac (25%); intra-acardiac (25%) and mixed (5%).

TAPVD requires surgical correction for survival, without which, 80% babies die before their first birthday.<sup>4</sup> We report an adult patient with supra-cardiac TAPVD who underwent successful surgical correction.

## Case Summary

A 28 year old man, weighing 57.5 kg, and a banker by profession, presented to our hospital with complaints of shortness of breath on exertion for the past 4–5 months. Classified as New York Heart Association (NYHA) Class II worsening towards Class III.

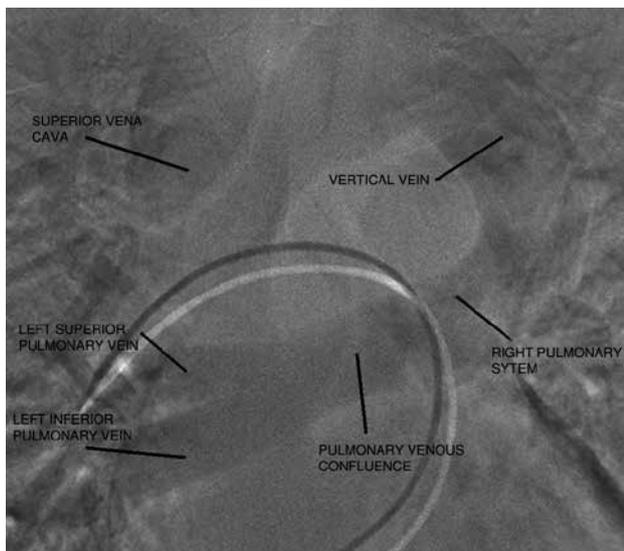
He was diagnosed with ASD in childhood but remained asymptomatic until recently when he started developing dyspnea. Following this, he was advised to undergo cardiac catheterization (Cath) and referred to our hospital. His past medical and surgical history was unremarkable and he was not currently using any medications.

On examination, he was a young man of medium size build in good general status. Peripheral examination of the limbs revealed mild cyanosis with markedly clubbed fingers; other examinations were unremarkable. His O<sub>2</sub> saturation was 96–98% at rest with haemoglobin and a haematocrit of 15.6 g/dl and 46.7%, respectively. Echocardiogram revealed a 13 mm Ostium Secundum ASD with bi-directional flow favouring a left to right shunt along with TAPVD to the superior vena cava (SVC) with a mild gradient across the pulmonary valve (PG) of 35–40 mmHg. His Cath revealed a left ventricular (LV) pressure of 130/13 mmHg, right ventricular pressure

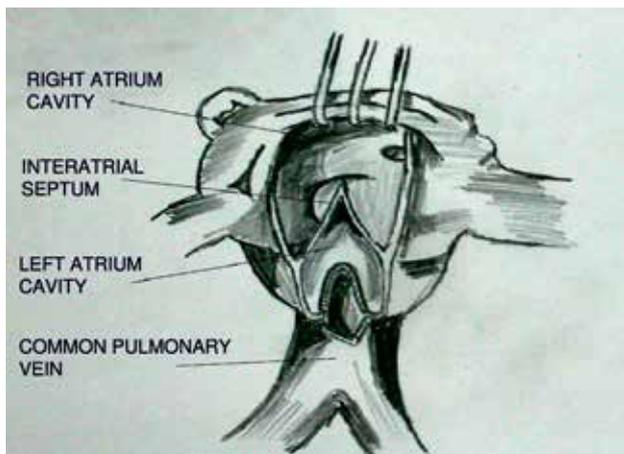
(RV) of 60/12 mmHg, pulmonary artery pressure (PA) of 50/18 mmHg with reversible moderate pulmonary arterial hypertension (PAHTN) confirming the findings of echocardiogram (Figure 1). He was advised to undertake elective corrective surgery.

The patient was operated through a median sternotomy and was put on cardiopulmonary bypass. Operative findings revealed a supra-cardiac TAPVD with a large vertical vein draining into the right brachiocephalic vein through into the SVC; along with a large secondary 2° ASD (Figure 2). Both right and left pulmonary vein (PV) were draining into the confluence. Pre-operative aortic and PA pressures were 77/56 mmHg and 28/13 mmHg, respectively. The procedure involved redirection of PV to the left atrium (LA), closure of ASD with autologous pericardial patch, and ligation of vertical vein. Post-operative aortic and PA pressures were 90/40 mmHg and 22/10 mmHg, respectively. The patient was transferred to the ICU intubated, ventilated, and on minimal inotropic support.

Post-operative echocardiogram revealed all 4 PV were entering into LA with no obstruction appreciated at the entry site. The inter-atrial septum was found to be intact. There was no pulmonary stenosis and the pulmonary gradient (PG) was found to be 8 mmHg. The post-operative stay was stable and



**Fig 1** Angiogram of the patient revealing right and left pulmonary venous system draining into a venous confluence. The venous confluence drains into the vertical vein which is connected to the superior vena cava through innominate vein.



**Fig 2** Trans-atrial trans-septal incision, opening the left atrium, a corresponding incision in the common pulmonary venous confluence and anastomosis.

the patient was discharged on the 6th post-operative day. Six months after the surgical repair the patient has been persistently improving and is now in NYHA Class I.

## Discussion

In TAPVD, the common PV either does not connect with the pulmonary venous system or does not form at all. Right atrium (RA) and RV enlargement occurs because all pulmonary venous return connects to the systemic venous system. If neither pulmonary nor systemic connection is available to the lungs, then death usually occurs during the neonatal period.<sup>5</sup> Patients with TAPVD usually present in the early neonatal period, often with marked cyanosis and shock, and almost always require a surgical intervention on an emergency basis.<sup>6</sup>

An ASD/patent foramen ovale serves a vital function in keeping these patients alive allowing the LV to receive mixed oxygenated and de-oxygenated blood. The size of the communication determines the volume of blood that able to cross to the left side, eventually determining the cardiac output and systemic oxygenation. This is one of the structural factors that determines the patient's clinical status. The other is an obstruction in the path of PV drainage from the lungs to the systemic venous system. 75% of patients with TAPVD have a widely patent connecting vein with no obstruction of venous return.<sup>5</sup> The consequences of obstruction are pulmonary venous congestion, decreased oxygenation, and elevation of PA pressures that lead to pulmonary hypertension and eventually heart failure.<sup>7</sup>

Survival until the third decade is unusual, and increasing age is known to be an unfavorable factor for successful surgical outcome. The mortality of which, when surgical repair is not performed, is estimated at 80% within the first year.<sup>8</sup> Few cases have been reported of a successful surgical correction in the second decade<sup>8</sup> and the sixth decade<sup>7</sup> but none in the Asian literature. The major factors that contributed to the long-term survival of our patient was the supra-cardiac type of TAPVD, unobstructed pulmonary venous drainage, a large inter-atrial communication, and maintenance of vascular resistance and pulmonary pressure closer to normal limits.

Some authors suggest that the vertical vein should not be ligated when the left atrium is small and, therefore, poorly compliant to accommodate blood flow after surgical repair.<sup>9</sup> Others<sup>10</sup> believe that, although this technique may be associated with smoother postoperative course, it has the disadvantage of requiring re-intervention to eliminate the left-to-right shunt that may persist in the late post-operative phase. Generally, long-term prognosis after successful repair of TAPVD is favorable. Approximately 10% to 15% of patients have evidence of late pulmonary vein obstruction, which tends to be recurrent and progressive.<sup>7</sup> For this reason, long-term surveillance is important, and further studies are warranted to monitor the outcomes and long-term course of such patients.

## Conclusion

In conclusion, this report describes a case of an unobstructed supra-cardiac TAPVD in a 28-year-old man who was successfully treated through surgery. It is clearly evident, that surgery should not be excluded as an option in such cases. However, careful and thorough pre- and peri-operative assessment should be done to exclude other abnormalities; and a uniform management plan should be drawn for the management of such cases.

### *Ethical approval*

Consent was obtained.

### *Conflicts of interest*

No conflicts of interest have been declared by the author.

### *Author contributions*

AA – Conception and design of study, acquisition and analysis of data, initial draft of the article.

NS – Draft and revising article for critical intellectual content.

MK, MA, MMA – Interpretation of data, draft and revising article for critical intellectual content.

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