

# Perioperative Echocardiographic Features of Total Anomalous Pulmonary Venous Connection

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## ABSTRACT

Total anomalous pulmonary venous connection (TAPVC) refers to when all four pulmonary veins connect anomalously to the right atrium, directly or indirectly. Perioperative echocardiography, both transthoracic and transesophageal (TEE) with color flow imaging and Doppler interrogation, can identify the venous confluence, its connections, obstructions if any in the pathway, and any interatrial communication. They supplement each other in delineating the anatomy of such anomalous pulmonary venous connections. Perioperative TEE evaluation of patients with repaired TAPVC confirms the adequacy of repair, leaks or stenosis of the venous baffle. We summarize the role of perioperative echocardiography in understanding cases of TAPVC.

**Keywords:** Perioperative transesophageal echocardiography, Pulmonary veins, Total anomalous pulmonary venous connection, Transesophageal echocardiography, Transthoracic echocardiography.

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## INTRODUCTION

Total anomalous pulmonary venous connection (TAPVC) is a type of pulmonary venous anomaly in which all pulmonary veins (PVs) have connection to the systemic venous circulation. The TAPVC occurred in 1.5% cases with cardiovascular abnormalities in the Baltimore Washington Infant Study (1981–1987).<sup>1</sup> The TAPVC can be classified as type I, anomalous connection at the supracardiac level; type II, anomalous connection at the cardiac level [to the coronary sinus (CS)]; type III, anomalous connection at

the infracardiac level; and type IV, anomalous connection at two or more of the above levels.<sup>2</sup> These patients cannot survive without an atrial septal defect (ASD) or patent foramen ovale; therefore, an interatrial communication is considered part of the complex, and flow across the ASD is usually right-to-left shunt. When imaging, the presence of a pure right to left shunt across an ASD should alert one to the possibility of TAPVC.

In small children, cardiac catheterization has an inherent risk ranging from death, cerebral infarction, and cardiac injury to rhythm disturbances<sup>3</sup> and can be avoided with the use of echocardiography to diagnose TAPVC. Transthoracic echocardiography (TTE) can accurately diagnose TAPVC, and the precise pattern of venous drainage can be delineated.<sup>4,5</sup> Intraoperative transesophageal echocardiography (TEE) complements TTE and can be used to confirm the preoperative diagnosis in the operating room. The TEE can inform about the adequacy of repair in the immediate postbypass period and avoid reoperations.<sup>6</sup> In infants with TAPVC, however, the TEE probe can compress the pulmonary venous confluence and result in hemodynamic instability and thus, in these patients, the TEE probe insertion is safer after sternotomy.<sup>7,8</sup>

In this article, we discuss the various transthoracic views that are helpful in diagnosing TAPVC, and TEE examination of TAPVC in the perioperative period will also be discussed.

## Normal Pulmonary Venous Drainage

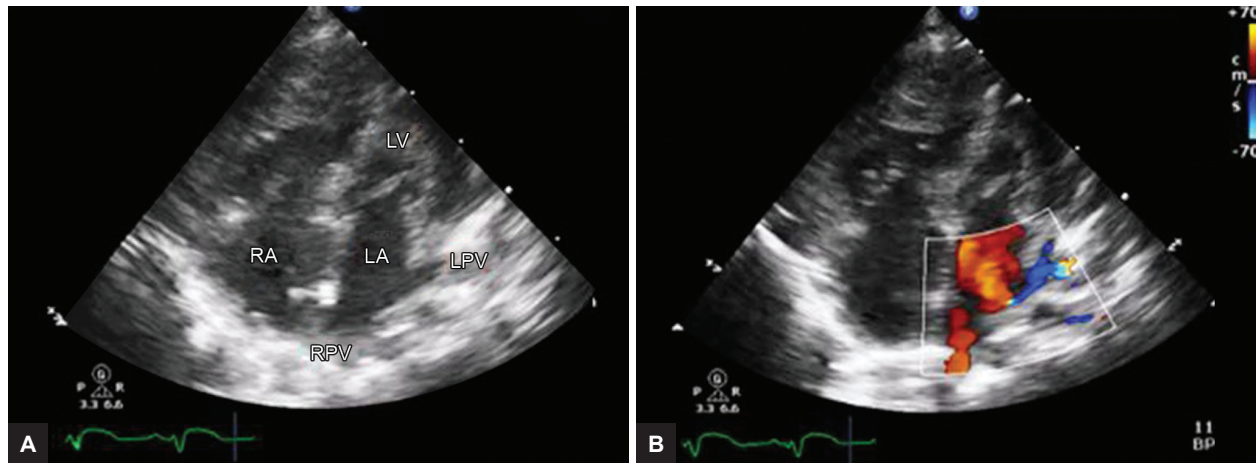
Normally, all PVs enter the left atrium (LA) along the superior portion of the posterior wall. The number of PVs is variable, but typically three PVs return from the right lung, and two from the left. The TTE examination of normal pulmonary venous drainage consists of suprasternal, parasternal, and subcostal views. From the apical 4-chamber view, one can see the entrance of one or two PVs into LA, most typically, the left lower PV, and either the right middle or right upper PV on the right (Figs 1A and B). Color flow mapping should be used to identify the PVs. Pulmonary venous flow pattern can be recorded by placing a 2- to 3-mm sample volume 2 cm into the left lower PV. Three distinct waves are demonstrated: An antegrade wave during systole (PVs) and diastole

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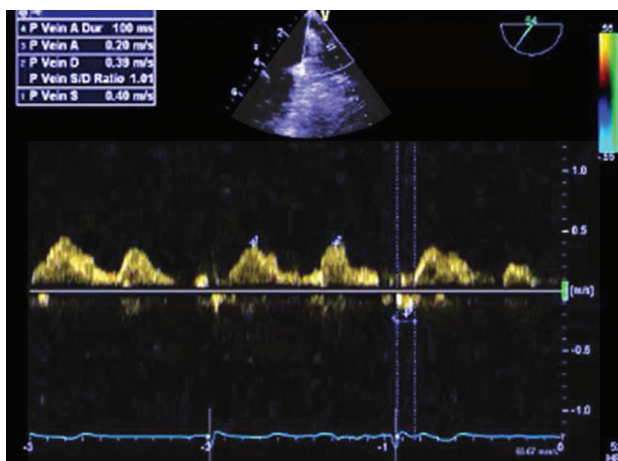
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**Figs 1A and B:** Transthoracic echocardiography apical 4-chamber with RPV and LPVs draining into LA



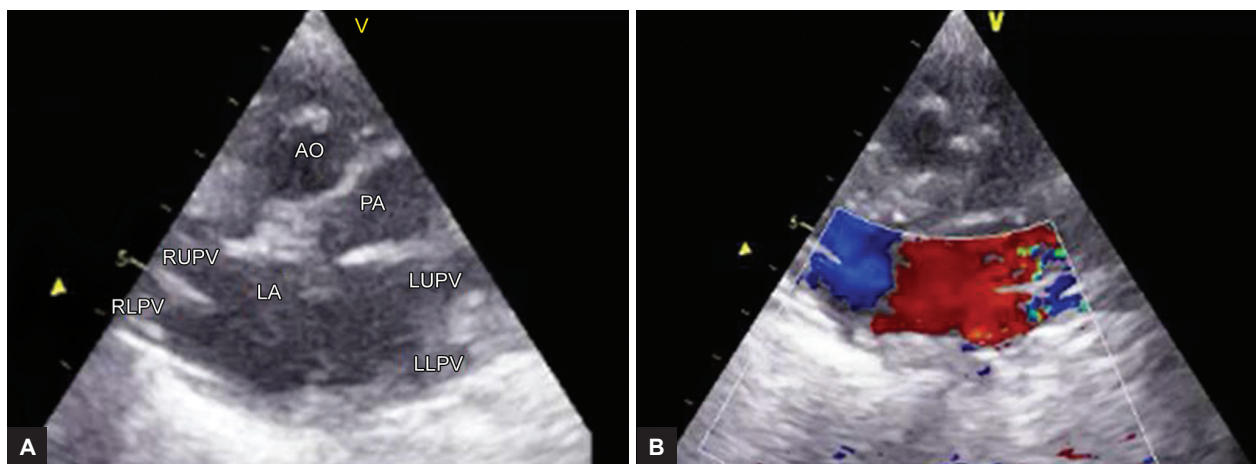
**Fig. 2:** Normal pulmonary vein pulsed wave Doppler

(PVd) and a retrograde wave coincident with atrial systole (PVa) (Fig. 2). All the four PVs draining into LA can be visualized in high parasternal short axis view angled inferiorly toward the LA or from the suprasternal notch with posterior angulation (crab view). Most commonly, in the crab view, all, but the right upper PV is seen well, although it is possible to see all PVs simultaneously (Fig. 3).

The TEE can record all the PVs entering LA. For left PVs, from midesophageal 4-chamber view probe is rotated toward far left (Figs 4A and B). The left atrial appendage can be used as a landmark to identify the left upper PV (LUPV). The probe is then gradually advanced to see the left lower PV (LLPV). The LLPV enters the LA (LA) just below the LUPV. For visualization of the left PVs, typically the angle is 120°, and one angulates toward the left until they reach the descending aorta, then turn slightly back to pick up the LPVs. In order to visualize the right PVs, set the angle at 30 to 45 in midesophageal 4-chamber view and rotate the probe clockwise to the patient's far right (Fig. 5).

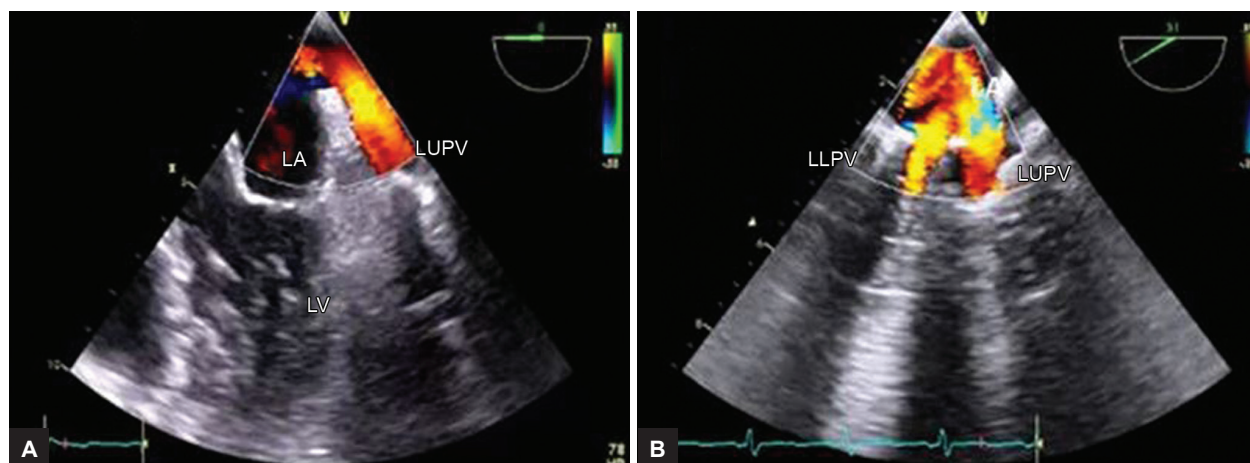
### SUPRACARDIAC TAPVC

In supracardiac TAPVC, all PVs form a common chamber. The common chamber usually drains via a left vertical vein into the left brachiocephalic vein, and less often, common chamber drains directly to the superior vena cava (SVC), usually at its junction with the right atrium (RA), and rarely to the azygos vein. The confluence of the anomalous PVs lies behind the upper aspect of the LA, generally directly below the pulmonary arteries (Video 1).

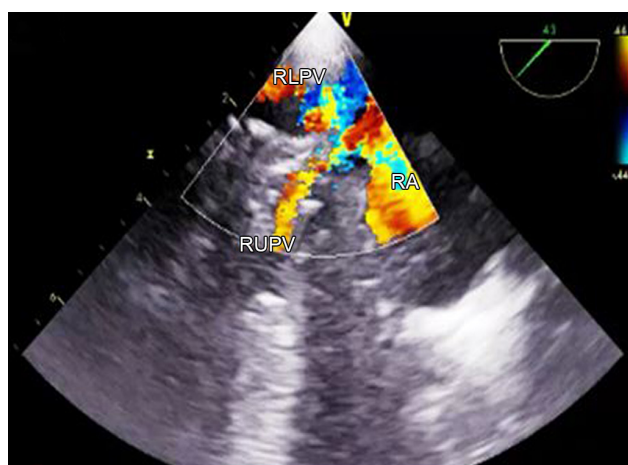


**Figs 3A and B:** Transthoracic echocardiography high parasternal short axis view (crab view) showing normal drainage of pulmonary veins into LA

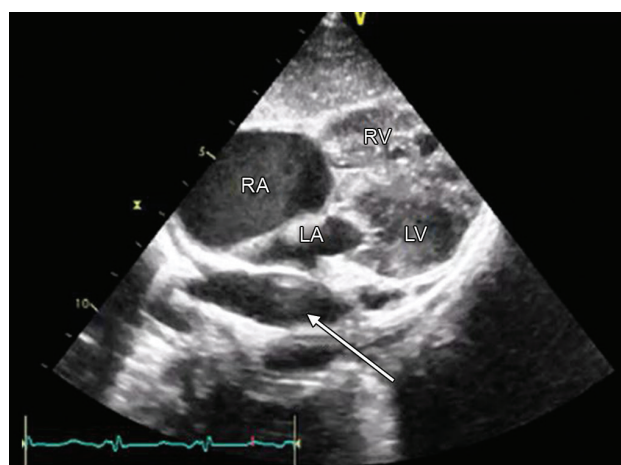




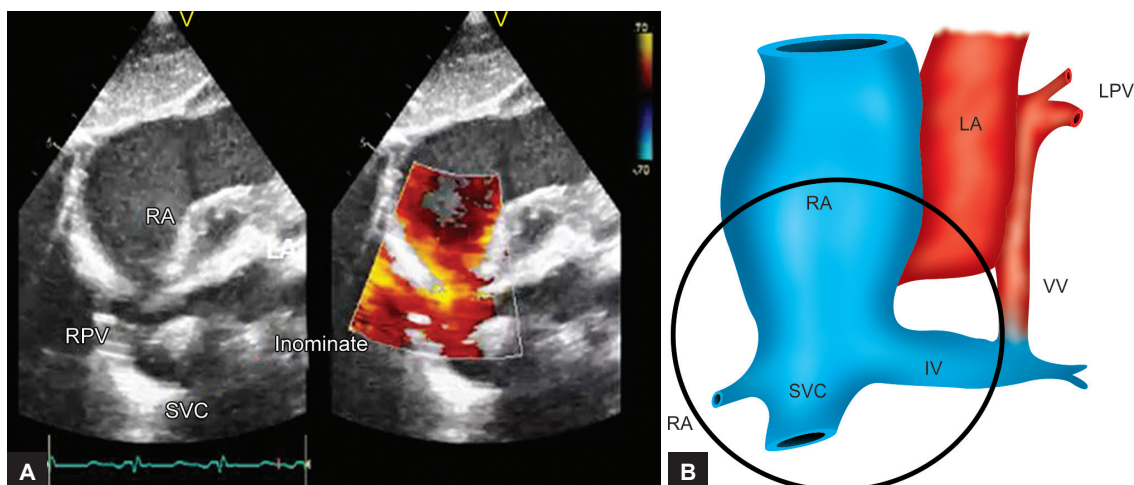
**Figs 4A and B:** Transthoracic echocardiography ME 4-chamber view rotated to left showing focused left-sided PV view



**Fig. 5:** Transthoracic echocardiography ME 4-chamber view rotated to right showing focused right-sided PV draining into RA



**Fig. 6:** Transthoracic echocardiography subcostal long axis view showing venous confluence behind LA (arrow) (this image is upside down)



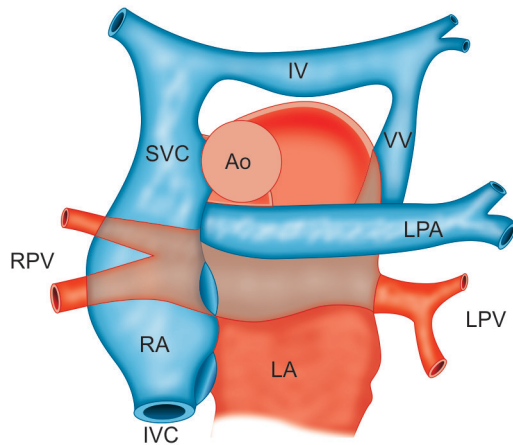
**Figs 7A and B:** Transthoracic echocardiography subcostal long axis view showing innominate vein connection (this image is upside down)

By TTE imaging from the subcostal long axis window, one can see the CS in the inferior posterior aspect of the LA as it runs in the atrioventricular groove. By moving the scan head from medial to lateral position, the presence of venous confluence can be seen (Fig. 6).

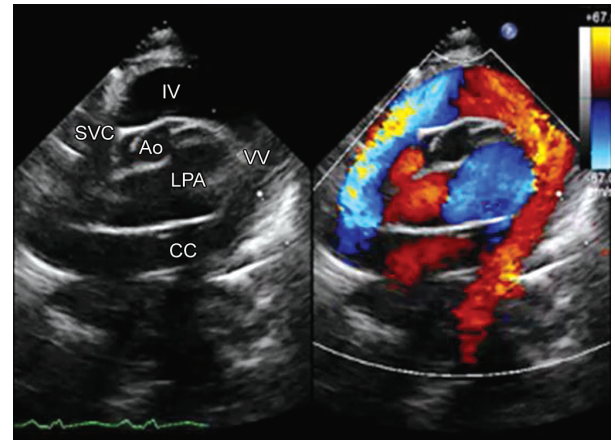
The subcostal long axis view demonstrates the connection of vertical vein to the innominate or the SVC

(Figs 7A and B). The vertical vein connects the venous confluence to innominate vein or directly to SVC. This vertical vein can pass anterior or posterior to the left pulmonary artery (Figs 8 and 9).

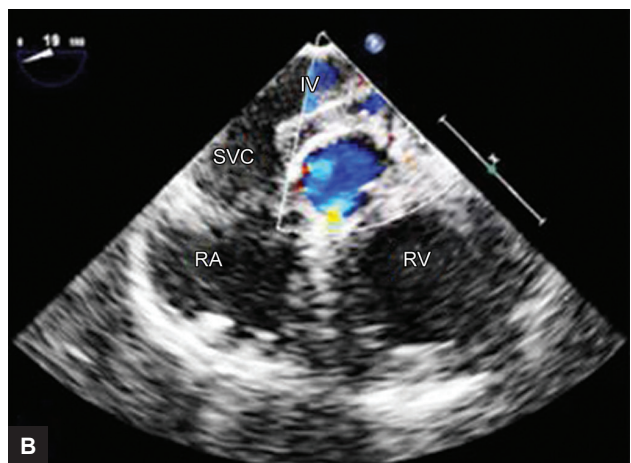
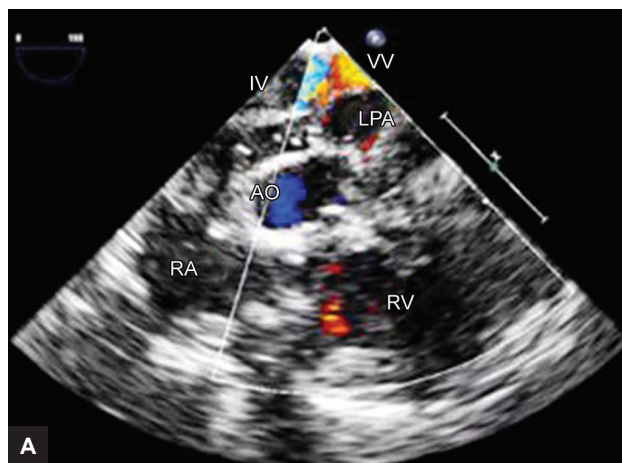
In a case of supracardiac TAPVC, midesophageal TEE views are most helpful for localizing the venous confluence behind the LA. From midesophageal



**Fig. 8:** Venous confluence behind LA

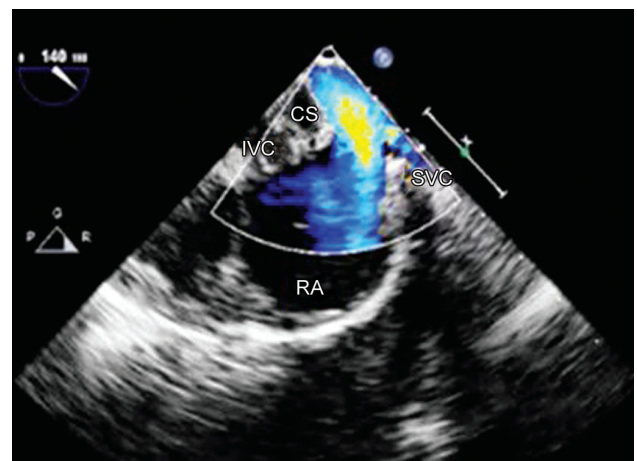


**Fig. 9:** The TTE suprasternal view showing venous confluence draining to vertical vein lateral to LA. Vertical vein draining into left innominate vein and vertical vein into superior vena cava



**Figs 10A and B:** (A) Transesophageal echocardiography ME aortic short axis view showing vertical vein to innominate vein anterior to LPA and aortic arch; and (B) probe rotated to right showing innominate vein into SVC

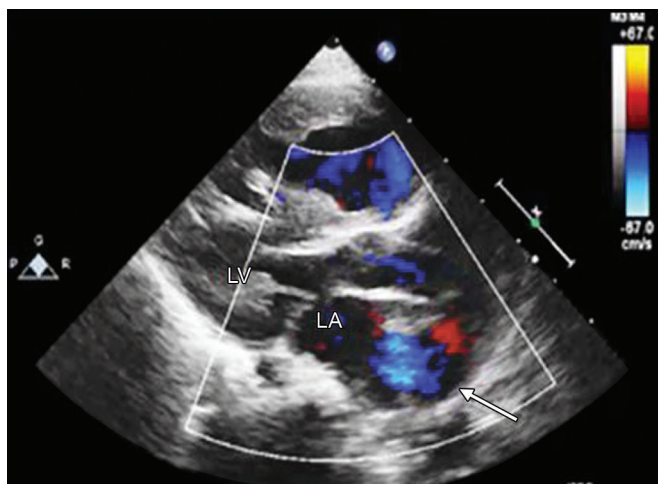
4-chamber view, retroflexion of the probe will show the venous confluence behind the LA. The vertical vein is seen as a vessel lateral to the LA (best viewed at 90°). The vertical vein appears similar to left SVC; however, the flow is venous flow directed away from the heart. The technique of visualizing the vertical vein to innominate vein connection has been described by Ammash et al.<sup>9</sup> Instead of the veins entering LA, the PVs enter a vessel lateral to the LA, which courses anterior to the left pulmonary artery. Visualize the proximal transverse aortic arch, with the longitudinal plane, in the short-axis view (30–45°). Rotate the scope shaft back and forth from right to left, and while in the longitudinal image plane, the long-axis view of the innominate vein will appear anterior to the upper thoracic aorta and transverse arch (Figs 10A and B). The innominate vein can be traced draining into SVC on gradual withdrawal of the probe and rotating it to right. While looking at the innominate vein in the long-axis view, slowly advance the transducer into the esophagus and reduce the angle slightly until the anomalous connecting PVs are visualized adjacent to



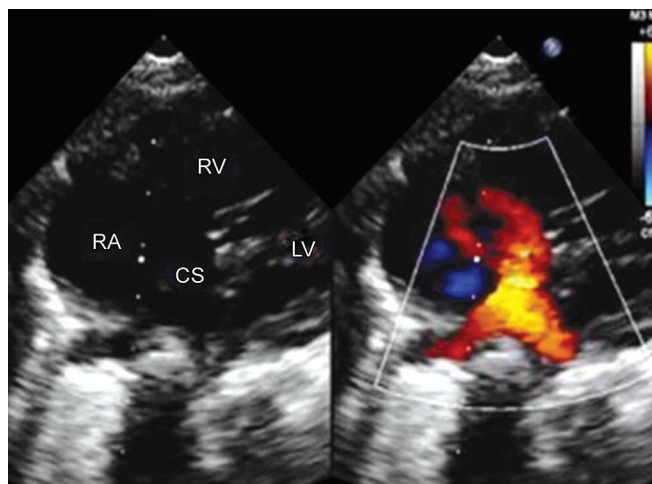
**Fig. 11:** Transesophageal echocardiography ME showing dilated SVC

the lateral wall of the LA. In contrast, rightward rotation of the scope and increasing the angle to 120 to 140° will result in a long-axis view of the SVC that will be dilated (Fig. 11). Rarely, there could be right-sided or bilateral vertical vein from the common chamber draining into right or left SVC or azygous vein.<sup>10,11</sup>

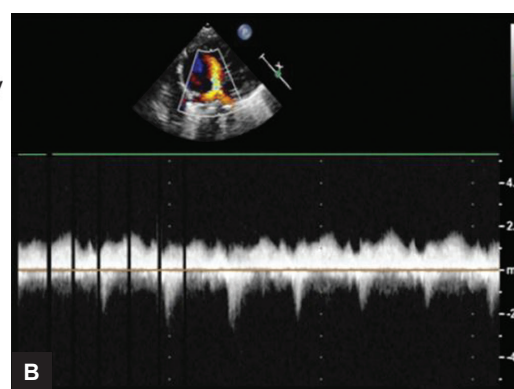
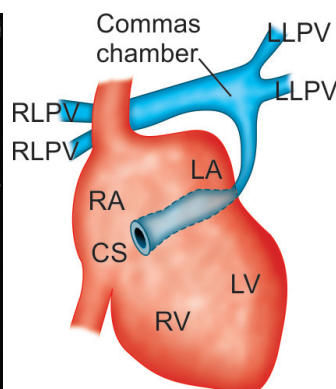
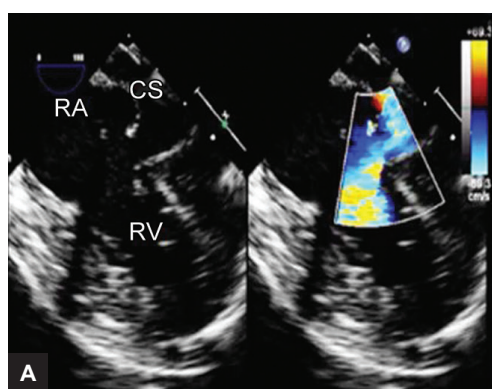




**Fig. 12:** Transthoracic echocardiography PLAX view showing venous confluence (arrow) behind LA in cardiac TAPVC



**Fig. 13:** Transthoracic echocardiography subcostal long axis view showing dilated coronary sinus opening into dilated RA



**Figs 14A and B:** (A) Transesophageal echocardiography ME 4 chamber showing dilated coronary sinus; and (B) A high flow PW is seen in CS in cardiac TAPVC

## CARDIAC TAPVC

Cardiac TAPVC drains usually to the CS or less often directly to the RA. In these cases, there is no confluence and the CS will be notably dilated due to the increase flow through it.

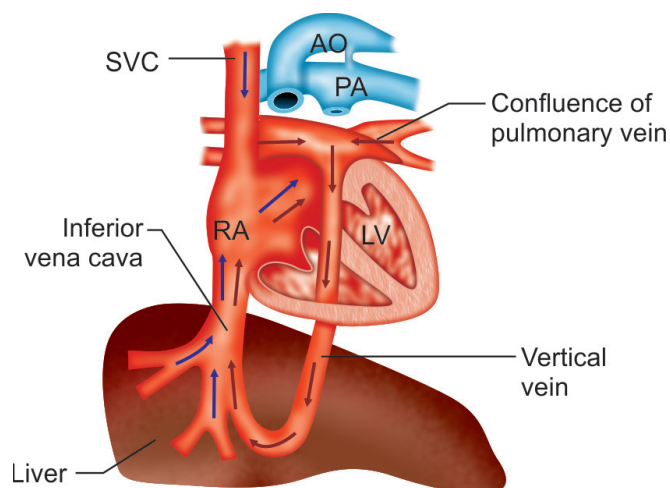
By TTE imaging, in the subcostal coronal window, parasternal long-axis view, the common venous confluence is seen as an echo-free space behind the LA (Fig. 12). In cardiac TAPVC draining to the CS, the CS is dilated and can be visualized from subcostal and parasternal views. The view of the PVs draining to the CS has been termed the “whale’s tail” due to a typical echocardiographic appearance (Fig. 13). Persistent left SVC and PAPVC can cause CS dilatation and should be excluded.

The TEE allows for clear imaging of the atria, atrial septum, PVs and the venae cavae from multiple imaging planes. A dilated coronary sinus is seen in cardiac type TAPVC to the CS midesophageal 4-chamber view focused for CS by slight retroflexion (Fig. 14A). A high flow pulsed wave (PW) is seen in CS in subcostal TTE whale’s tail view as described in above paragraph

(Fig. 14B). The pulmonary confluence can be seen behind the LA with all PVs draining into it. Both right and left PVs can be seen draining into common chamber and into CS thereof. Left PVs can be seen in midesophageal 4-chamber view focused on left PV (rotate probe to left and withdraw slightly), and then rotating the probe to right, wherein it can be seen draining into CS. Right PV can be seen draining into CS in modified bicaval view by rotating the probe from right to left and keeping RA in center (Video 2).

## INFRACARDIAC TAPVC

The pulmonary venous confluence is situated behind the LA. The confluence is usually oriented in a more vertical plane, with PVs attaching in a superior-to-inferior fashion, resulting in a more “Christmas tree” arrangement. A draining vertical vein travels through the esophageal hiatus, and then typically connects to the portal system (Fig. 15). The PVs draining into the pulmonary venous confluence can be visualized best by suprasternal windows. Subcostal imaging shows the size, course, and location of the draining vertical vein, which



**Fig. 15:** Intracardiac-type TAPVC

is draining venous flow away from the heart inferiorly. The connection of the vertical vein to the portal venous system, ductus venosus, hepatic vein, or inferior vena cava can be seen in subcostal views (Figs 16A and B). On TEE, opening of the vertical vein into the IVC can be seen in low bicaval view (90°). A dilated IVC and high flow on pulsed wave Doppler is seen. It is very difficult to delineate the entire path of the vertical vein from the pulmonary confluence to the IVC.

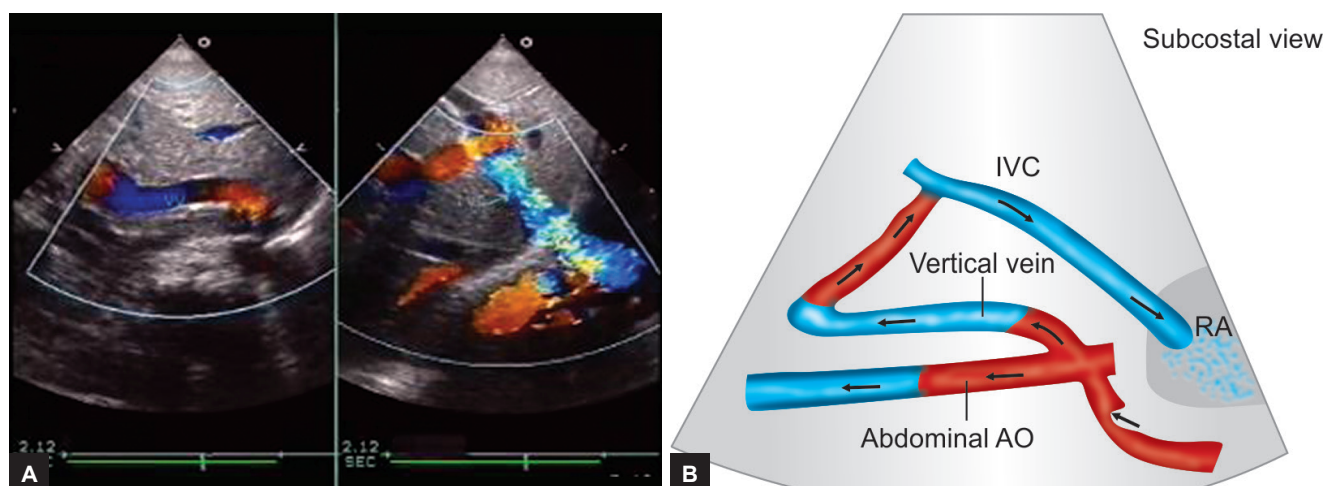
### PULMONARY VENOUS OBSTRUCTION IN TAPVC

The presence of pulmonary venous obstruction in TAPVC is a predictor of long-term outcome and determinant of surgical plan.<sup>12,13</sup> In supracardiac TAPVC, obstruction can occur at any point along the course of vertical vein, or at the junction of vertical vein to innominate or SVC. Color Doppler and pulse wave Doppler are used to demonstrate the obstruction. At the site of obstruction, a high-velocity, nonphasic flow profile is seen. Obstruction at the innominate vein–SVC junction can be visualized in transesophageal transgastric view, wherein, after

obtaining RV basal view by anteflexing, the angle of the probe is increased to 50 to 60° (Fig. 17). In a variety of obstructed supracardiac TAPVCs, the vertical vein can get externally compressed in between two vascular structures or between mainstem bronchus and any other vascular structure, specially, left pulmonary artery. In cardiac type of TAPVC, obstruction can occur at the level of entry of individual PVs to the pulmonary confluence or at the level of pulmonary confluence to CS (Fig. 18). Infracardiac TAPVC is associated with obstruction in majority of cases either at the point where the connecting vein joins the portal vein or ductus venosus, or it may be compressed where it penetrates the diaphragm. Obstruction most often occurs at the junction between the vertical vein and the portal vein, ductus venosus or hepatic vein, and Doppler tracings are characterized by an increase in absolute venous velocities and a loss of phasicity. Loss of phasic flow in the PVs is an indication of downstream obstruction.<sup>14</sup>

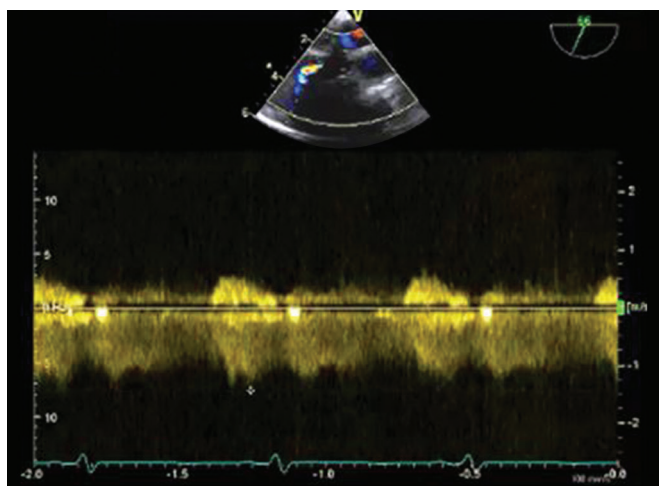
### POSTOPERATIVE ASSESSMENT

The goals of postcardiopulmonary bypass imaging include evaluation of the repair, exclusion of residual lesions, and functional assessment including the atrio-ventricular valves, right heart pressures by Doppler estimate, and ventricular function. In TAPVC, the surgical anastomosis is directly anterior to the esophagus and too close to the TEE probe to allow complete interrogation. One should look for any obstruction at the level of pulmonary confluence to left atrial anastomosis. Following repair of TAPVC, obstructions of both the pulmonary venous pathway and the SVC have been described.<sup>15</sup> In repaired anomalous pulmonary venous drainage, echocardiographic follow-up evaluation should include assessment for any stenosis of the venous baffle to the LA, residual leaks, and presence of any stenosis of the systemic vein, particularly, SVC (Figs 19A and B).

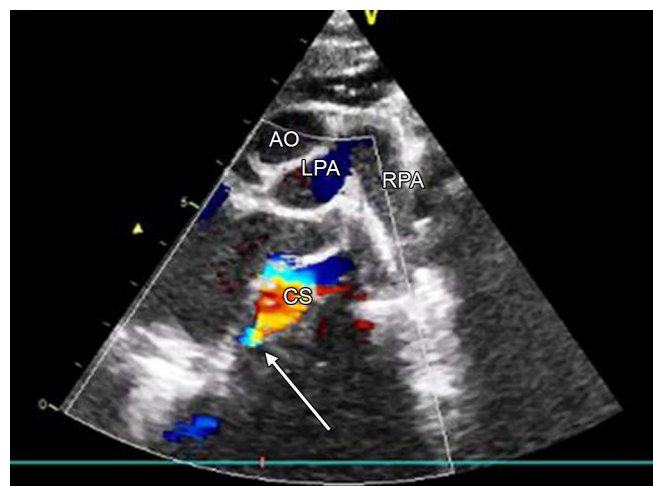


**Figs 16A and B:** Transthoracic echocardiography subcostal view showing vertical vein to IVC

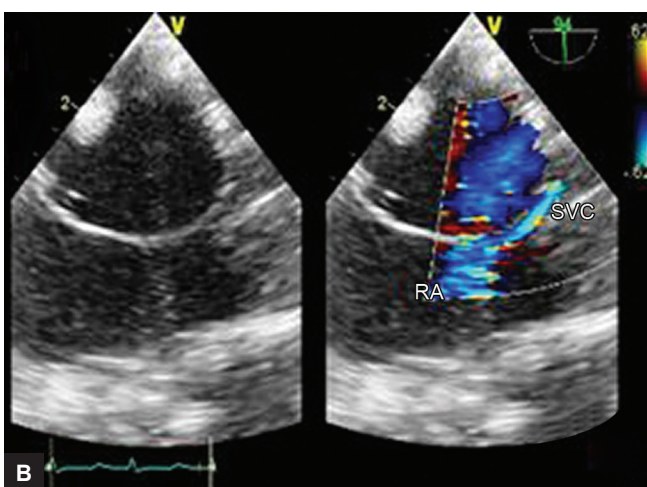
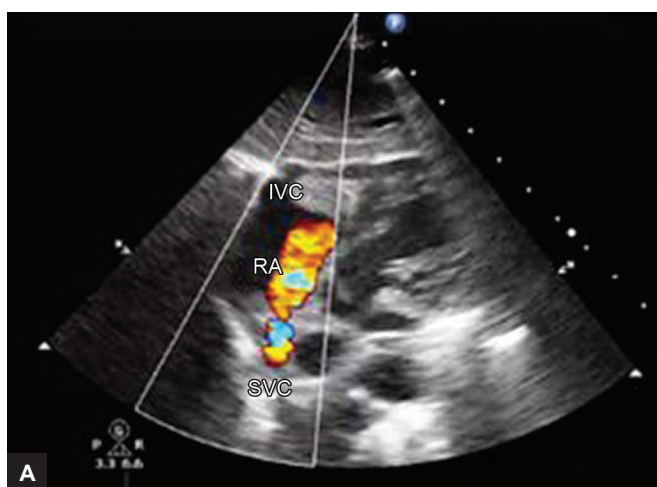




**Fig. 17:** Transesophageal echocardiography transgastric view. Nonphasic pulsed wave due to obstruction of venous drainage



**Fig. 18:** Transesophageal echocardiography high parasternal view depicting obstruction at junction of pulmonary confluence opening into coronary sinus in CS TAPVC (arrow)



**Figs 19A and B:** (A) Superior vena cava obstruction postrepair TTE subcostal; and (B) transesophageal echocardiography ME bicaval view

Certain varieties of total anomalous pulmonary venous return could not be visualized completely, in particular drainage to the RA by individual veins. It appears that mixed drainage may present difficulties, but then the same problem does not occur with angiocardiology or magnetic resonance imaging. Such advanced imaging studies are helpful, particularly, in the presence of a small LA and large right ventricle, but by itself difficulties still arise between total anomalous pulmonary venous connection and persistent fetal circulation. When basing a surgical decision upon echocardiography, the risks of clearly identifying the pulmonary venous system, which is absolute in every detail, must be weighed up against the risks of invasive procedures. In TAPVC, noninvasive studies enable a correct diagnosis with some potential deficiencies in the mixed types and in visualizing the descending channel beneath the diaphragm. Thus, with two-dimensional echocardiography using a high-frequency system, the diagnosis can readily be made and, in the majority of cases, the exact mode of drainage identified.

## REFERENCES

1. Correa-Villaseñor A, Ferencz C, Boughman JA, Neill CA. Total anomalous pulmonary venous return: familial and environmental factors. The Baltimore-Washington Infant Study Group. *Teratology* 1991 Oct;44(4):415-428.
2. Craig JM, Darling RC, Rothney WB. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. *Lab Invest* 1957 Jan-Feb;6(1):44-64.
3. Krasemann T. Complications of cardiac catheterisation in children. *Heart* 2015 Jun;101(12):915.
4. 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 Dec;46(6):613-623.
5. Lee ML, Wu MH, Wang JK, Lue HC. Echocardiographic assessment of total anomalous pulmonary venous connections in pediatric patients. *J Formos Med Assoc* 2001 Nov;100(11):729-735.
6. Mishra A, Madhavan J, Ghuman BPS, Raj R, Kumar A, Dutta V, Negi S, Tandon Y, Kumar A, Arya VK, et al. impact and cost effectiveness of routine intraoperative transthoracic and transesophageal echocardiography on surgical decision

- making in pediatric cardiac surgery. *J Perioper Echocardiogr* 2014 Jan;2(1):3-9.
7. Chang YY, Chang CI, Wang MJ, Lin SM, Chen YS, Tsai SK, Lue HC. The safe use of intraoperative transesophageal echocardiography in the management of total anomalous pulmonary venous connection in newborns and infants: a case series. *Paediatr Anaesth* 2005 Nov;15(11):939-943.
8. Frommelt PC, Stutz EA. Transesophageal echocardiographic in total anomalous pulmonary venous drainage: hypotension caused by compression of the pulmonary venous confluence during probe passage. *J Am Soc Echocardiogr* 1994 Nov-Dec;7(6):652-654.
9. Ammash NM, Seward JB, Warnes CA, Connolly HM, O'leary PW, Danielson GK. Partial anomalous pulmonary venous connection: diagnosis by transesophageal echocardiography. *J Am Coll Cardiol* 1997 May;29(6):1351-1358.
10. Mahla H, Kavya Mallikarjun K, Sastry UMKR, Mahimarangaiah J, Manjunath CN. Supracardiac total anomalous pulmonary venous connection with right ascending vertical vein. *Cardiovasc Med* 2014;17(9):273-274.
11. Grosse-Wortmann L, Friedberg MK. Images in cardiovascular medicine. Bilateral vertical veins from a common confluence in supracardiac total anomalous pulmonary venous connection. *Circulation* 2008 Aug;118(6):e103-e104.
12. Seale AN, Uemura H, Webber SA, Partridge J, Roughton M, Ho SY, McCarthy KP, Jones S, Shaughnessy L, Sunnegardh J, et al. Total anomalous pulmonary venous connection: morphology and outcome from an international population-based study. *Circulation* 2010 Dec;122(25):2718-2726.
13. Yun TJ, Coles JG, Konstantinov IE, Al-Radi OO, Wald RM, Guerra V, de Oliveira NC, Van Arsdell GS, Williams WG, Smallhorn J, et al. Conventional and sutureless techniques for management of the pulmonary veins: evolution of indications from postrepair pulmonary vein stenosis to primary pulmonary vein anomalies. *J Thorac Cardiovasc Surg* 2005 Jan;129(1):167-174.
14. Grosse-Wortmann L, Al-Otay A, Goo HW, Macgowan CK, Coles JG, Benson LN, Redington AN, Yoo SJ. Anatomical and functional evaluation of pulmonary veins in children by magnetic resonance imaging. *J Am Coll Cardiol* 2007 Mar;49(9):993-1002.
15. Breckenridge IM, de Leval M, Stark J, Waterston DJ. Correction of total anomalous pulmonary venous drainage in infancy. *J Thorac Cardiovasc Surg* 1973 Sep;66(3):447-453.