

## Partial Anomalous Pulmonary Venous Connection: Diagnosis by Transesophageal Echocardiography

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**Objectives.** This study sought to demonstrate that with proper technique, identification of the normal and abnormal pulmonary venous connection can be made with confidence using transesophageal echocardiography (TEE).

**Background.** Partial anomalous pulmonary venous connection (PAPVC) is an uncommon congenital anomaly whose diagnosis has classically been made using angiography.

**Methods.** We performed a retrospective review of all patients of all ages with PAPVC diagnosed at the Mayo Clinic who had undergone TEE because of either right ventricular volume overload or suspected intracardiac shunting by transthoracic echocardiography or intraoperatively.

**Results.** A total of 66 PAPVCs were detected in 43 patients (1.5/patient); in 2 additional patients, TEE suggested, but did not diagnose, PAPVCs. Shortness of breath was the most common presenting symptom (42.2%), followed by heart murmur and supraventricular tachycardia. Right-sided anomalous veins were identified in 35 patients (81.4%), left-sided in 7 (16.3%) and bilateral in 1 (2.3%). There was a single anomalous connecting vein in 23 patients (53.5%), two in 18 (41.9%), three in 1 (2.3%)

and four in 1 (2.3%). The connecting site was the superior vena cava (SVC) in 39 veins (59.1%), right atrial-SVC junction in 6 (9.1%), right atrium in 8 (12.1%), inferior vena cava in 1 (1.5%) and the coronary sinus in 2 (3.0%). Ten anomalous left pulmonary veins were connected by a vertical vein to the innominate vein (15.1%). Sinus venosus atrial septal defect (ASD) was the most common associated anomaly in 22 patients (49%), followed by ostium secundum ASD in 6 and patent foramen ovale in 4. Fifteen patients had an intact atrial septum. Thirty-one patients (68.8%) underwent surgical repair. PAPVC was confirmed in all patients, including the two whose TEE results were suggestive of PAPVC. All 49 PAPVCs detected by TEE preoperatively were confirmed at the time of operation.

**Conclusions.** TEE is highly diagnostic for PAPVC and can obviate angiography. Accurate anatomic diagnosis may influence the need for medical and surgical management. TEE should be performed in patients with right ventricular volume overload when the precordial examination is inconclusive.

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Partial anomalous pulmonary venous connection (PAPVC) was first described by Winslow in 1739 (1). This relatively uncommon congenital anomaly is found in 0.4% to 0.7% of autopsies (2,3) and represents persistence of embryonic anastomosis between the systemic and pulmonary vein plexus, resulting in one or more anomalously connecting pulmonary veins. Dotter et al. (4) first reported the diagnosis of this anomaly using cardiac catheterization in 1949. Since then, detection of PAPVC has been reported using contrast transthoracic echocardiography (TTE) (5), magnetic resonance imaging (MRI) (6), computed tomography (7), ventilation perfusion scan (8) and radionuclide angiography (9).

Although two-dimensional transesophageal echocardiogra-

phy (TEE) was first introduced in the late 1980s, to date there have been only isolated descriptions of the use of TEE for assessment of normal and anomalous pulmonary venous connection (10-17). TEE can provide diagnostic information regarding the normal and abnormal connections of the pulmonary veins (PVs) (13). The present study reviews a large experience in detecting PAPVC using TEE and describes the technique used for the systematic detection of the normal and abnormal pulmonary venous connections.

### Methods

**Patient selection and examination technique.** This is a retrospective review of patients of all ages with PAPVC who had a TEE at the Mayo Clinic between August 1988 and June 1995. Patients with complex congenital heart disease were excluded. TEE was performed with commercially available ultrasound instruments using either a biplane or multiplane scope. The TEE examination, including patient preparation, has been previously described (10,11,16,17).

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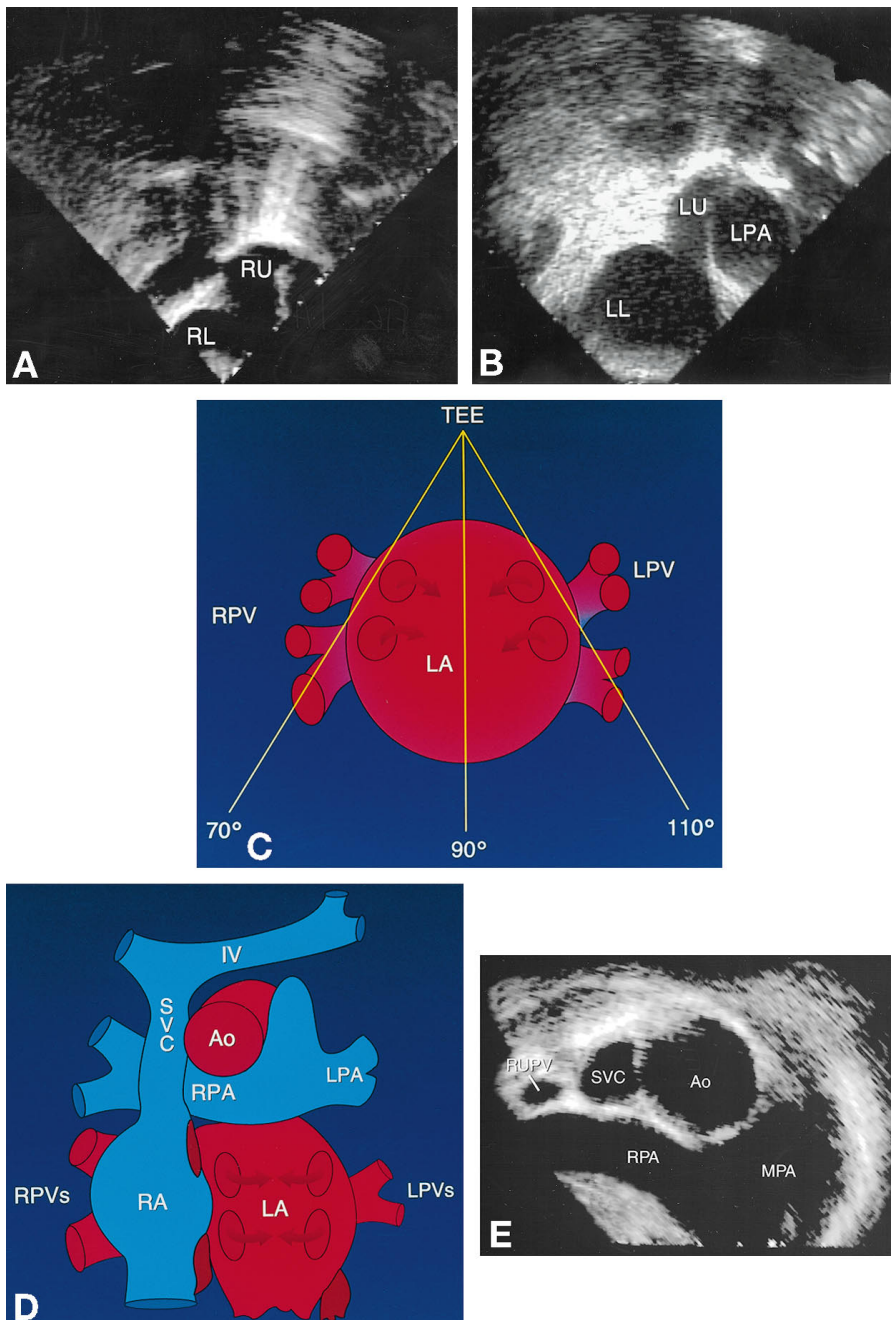
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**Abbreviations and Acronyms**

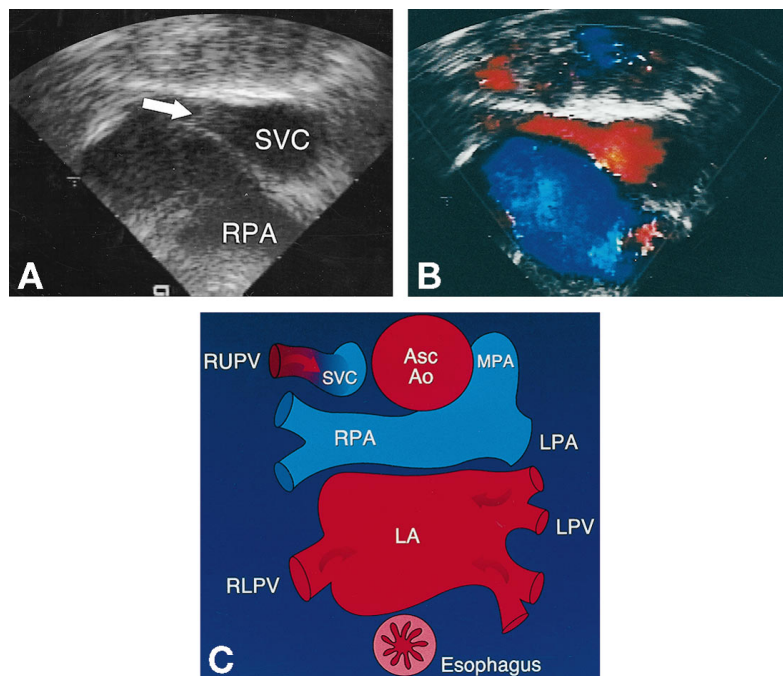
ASD	= atrial septal defect
LA	= left atrium
LAA	= left atrial appendage
MRI	= magnetic resonance imaging
PAPVC	= partial anomalous pulmonary venous connection
PV	= pulmonary vein
RA	= right atrium
SVC	= superior vena cava
TEE	= transesophageal echocardiography
TTE	= transthoracic echocardiography

**Normal pulmonary venous connections.** *Technique 1.* The most consistent and systematic examination of normal pulmonary venous connections begins with the tip of the TEE probe posterior to the left atrium (LA). *The right PVs* are imaged in the longitudinal plane by 1) flexing the biplane TEE tip medially or rotating the multiplane array to  $\sim 70^\circ$  (a foreshortened short-axis view of the left ventricular outflow tract is obtained); and 2) rotating the scope shaft to the patient's right off the medial wall of the LA. A Y-appearing image of the normal upper and lower PVs entering the LA is thus obtained (Fig. 1, A and C). *The left PVs* are imaged by 1) flexing the biplane TEE tip laterally or rotating the multiplane array to



**Figure 1.** Normal pulmonary venous connections. **A**, Normal right pulmonary venous connections are most consistently imaged from the transesophageal approach by orienting the array to  $\sim 70^\circ$  or medial flexion of the scope tip and then rotating the scope shaft rightward (medially). The right upper (RU) and right lower (RL) PVs form a typical Y configuration as they enter the LA. **B**, Normal left pulmonary venous connections are most consistently imaged from the transesophageal approach by orienting the array to  $\sim 110^\circ$  or lateral flexion of the scope tip and then rotating the scope shaft leftward (laterally). The left upper (LU) and left lower (LL) PVs form a typical Y configuration as they enter the LA. The left upper PV lies adjacent to the main left pulmonary artery (LPA). **C**, Omniplane transesophageal probe orientation. **D**, Gross anatomic arrangements of the PVs in relation to the surrounding cardiovascular structures. **E**, Computer composite wide-field view. With the transesophageal array in the horizontal orientation ( $0^\circ$ ) and withdrawn to the level of the right pulmonary artery (RPA), the right pulmonary artery is cut in its long axis. Anterior to the right pulmonary artery are three ovoid-appearing vessels—furthest rightward is the distal right upper PV (RUPV) lying immediately adjacent to the SVC. A typical anomalous pulmonary venous connection is for the right upper PV to join the SVC at this level. The conjoined structures would then form a “teardrop” appearance (see Fig. 2). The ascending aorta (Ao) is immediately leftward from the SVC and medial to the main pulmonary artery (MPA). IV = innominate vein; LPV = left PV; RPV = right PV.

**Figure 2.** Anomalous right pulmonary venous connection to the SVC. **A**, Short-axis transesophageal view of the SVC at the level of the right pulmonary artery (RPA). The SVC has a “teardrop” appearance caused by the entry of a PV (**arrow**) into the medial aspect of the SVC. **B**, Color flow Doppler demonstrating the flow disturbance at the medial aspect of the SVC where the anomalous venous connection is located. **C**, General anatomy of the view used to diagnose anomalous venous connection at the SVC. Note that the esophagus and transducer are posteriorly located. The LA and right pulmonary artery (RPA) are anterior to the esophagus. The SVC lies medial to the ascending aorta (Asc Ao) and anterior to the right pulmonary artery. The transesophageal transducer is withdrawn to the level of the right pulmonary artery while in a horizontal orientation. The typical anomalous pulmonary venous connection of the right upper PVs (RUPV) occurs in the medial SVC (**arrow**) and gives the SVC a “teardrop” appearance. RLPV = right lower PV; other abbreviations as in Figure 1.



~110° (a foreshortened long-axis view of the left ventricular outflow tract is obtained); and 2) rotating the scope shaft to the patient's left off the free wall of the LA. A Y-appearing image of the normal upper and lower PVs entering the LA is thus obtained (Fig. 1, B and C).

**Technique 2.** A second maneuver to image normal pulmonary venous connections (Fig. 1D) utilizes short-axis views at the base of the heart with the transverse plane of a biplane scope or 0° of a multiplane transducer. *The left PVs* are imaged by 1) rotating the transducer shaft to the patient's left; 2) placing the left atrial appendage (LAA) in the center of the image (adjacent to the LAA is the left upper PV); and 3) advancing the transducer into the esophagus, whereby the left lower PV is imaged closest to the transducer and courses almost directly laterally. *The right PVs* are imaged by 1) placing the right atrium (RA) in the center of the image; 2) rotating the transducer shaft medially to the patient's right; 3) withdrawing the transducer to the level of the right pulmonary artery (projected in its long axis; medial to the superior vena cava [SVC], the right upper PV is viewed in the short-axis view) (Fig. 1E); 4) slowly advancing the transducer into the esophagus while observing the course of the right upper PV, which enters the medial aspect of the LA; and 5) advancing the transducer further to observe the right lower PV close to the transducer coursing almost directly medially.

If normal pulmonary venous connections are not visualized with these maneuvers, then anomalous connection of PV must be suspected (13).

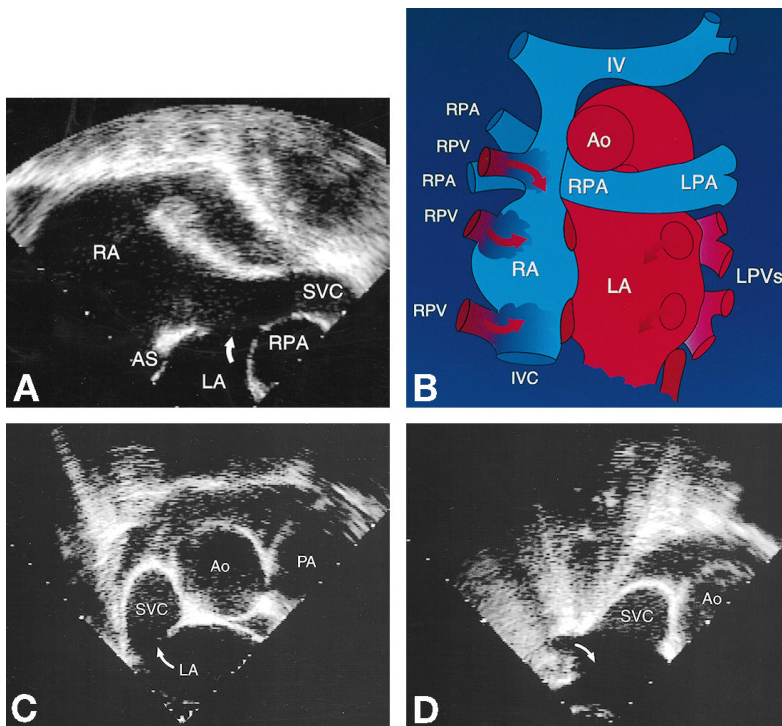
**Anomalous pulmonary venous connections. Right veins.** Anomalous pulmonary venous connections to the SVC were visualized by technique 2, using a short-axis view of the SVC at the level of the right pulmonary artery. The right upper PVs enter the free wall of the SVC, resulting in a teardrop

appearance instead of the normally round-appearing SVC (Fig. 2, A to C).

Connections to the RA-SVC and free wall of the right atrium are best visualized by slowly advancing the transducer into the esophagus while in the same short-axis plane. In each instance, color flow Doppler will help identify the anomalous pulmonary venous flow. Connection to the inferior vena cava is often difficult to visualize, but in our isolated experience, the short- and long-axis views were diagnostic of an anomalously connecting PV. The longitudinal plane of a biplane TEE scope or 90° on a multiplane scope can be used to visualize both sinus venosus atrial septal defect (ASD) commonly associated with PAPVC (Fig. 3A) and anomalous venous connection to the SVC or RA (Fig. 3B). At 0°, rotate the scope shaft rightward beyond the free wall of the SVC and RA. This will visualize sinus venosus ASD in the short-axis view (Fig. 3C). The anomalous connecting PVs will appear adjacent to the SVC or free wall of the RA (Fig. 3D).

**Left veins.** The most common anomalous connection is for one or more left PVs to enter a vertical vein that drains superiorly into the innominate vein (Fig. 4, B and E). The innominate vein empties into the right SVC, which may be dilated secondary to the shunted blood volume. The preferred means of visualizing this anomaly is technique 1. Instead of the veins entering LA, the PVs enter a vessel lateral to the LA, which courses anterior to the left pulmonary artery (Fig. 4, C and F). The vertical vein has the same appearance as a persistent left SVC. A modification of technique 1 is to view the mid-thoracic aorta in a longitudinal projection. Slowly withdraw the TEE transducer past the left bronchus and visualize the proximal transverse aortic arch. With the longitudinal plane, the transverse arch is viewed in the short-axis view. As the scope shaft is rotated back and forth from right to





**Figure 3.** Sinus venosus ASD with anomalous pulmonary venous connection to the RA. **A**, Diagnostic transesophageal long-axis view of a sinus venosus ASD (arrow). The view is a long-axis projection of the SVC. The defect, which allows communication of the LA and RA, is located in the superior fatty limbus of the atrial septum (AS) (arrow). The defect is just inferior to the right pulmonary artery (RPA). **B**, Anatomy of the anomalous pulmonary venous connections (arrows) typically seen in association with the sinus venosus defect. Connections are most common to the SVC (see Fig. 2C) and to the free wall of the RA. The left PVs (LPVs) are usually normally connected to the LA. **C**, Short-axis view at the level of a sinus venosus ASD (arrow). This is a view near the superior dome of the RA at the SVC-atrial junction. The SVC lies adjacent to the ascending aorta (Ao). The main pulmonary artery (PA) lies leftward to the aorta. **D**, Near the caval-atrial junction and the free wall of the RA, an anomalous pulmonary venous connection (arrow) is visualized entering the free wall of the RA. IVC = inferior vena cava; other abbreviations as in Figure 1.

left 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 (Fig. 4A). This is often the most sensitive maneuver to also visualize the vertical vein. While looking at the innominate vein in the long-axis view, slowly advance the transducer into the esophagus until the anomalous connecting PVs are visualized adjacent to the lateral wall of the LA (Fig. 4, C and F). In contrast, rightward rotation of the scope will result in a long-axis view of the SVC that will be dilated (Fig. 4D).

Anomalous connection of left PVs to the coronary sinus (Fig. 5D) are also visualized by technique 1. In the longitudinal plane, either start with the dilated coronary sinus (Fig. 5A) and rotate the transducer shaft to the patient's left, or, conversely, start with the veins and rotate the longitudinal plane rightward to the coronary sinus (Fig. 5, B and C). Short-axis views are less helpful for confident visualization of anomalous left PVs.

## Results

Forty-five patients (2 to 75 years old, mean age 41; 21 male, 24 female) had PAPVC and a TEE examination. TEE was performed as part of a diagnostic evaluation for suspected left to right shunt or unexplained pulmonary hypertension in 31 patients (69%) (Group I). The remaining 14 patients (31%) had the diagnosis of PAPVC previously made by cardiac catheterization ( $n = 6$ ), TTE ( $n = 4$ ) or MRI ( $n = 4$ ), and TEE was performed intraoperatively with operator's knowledge of the diagnosis (Group II).

Shortness of breath was the most common symptom associated with PAPVC (19 [42.2%] of 45 patients), followed by

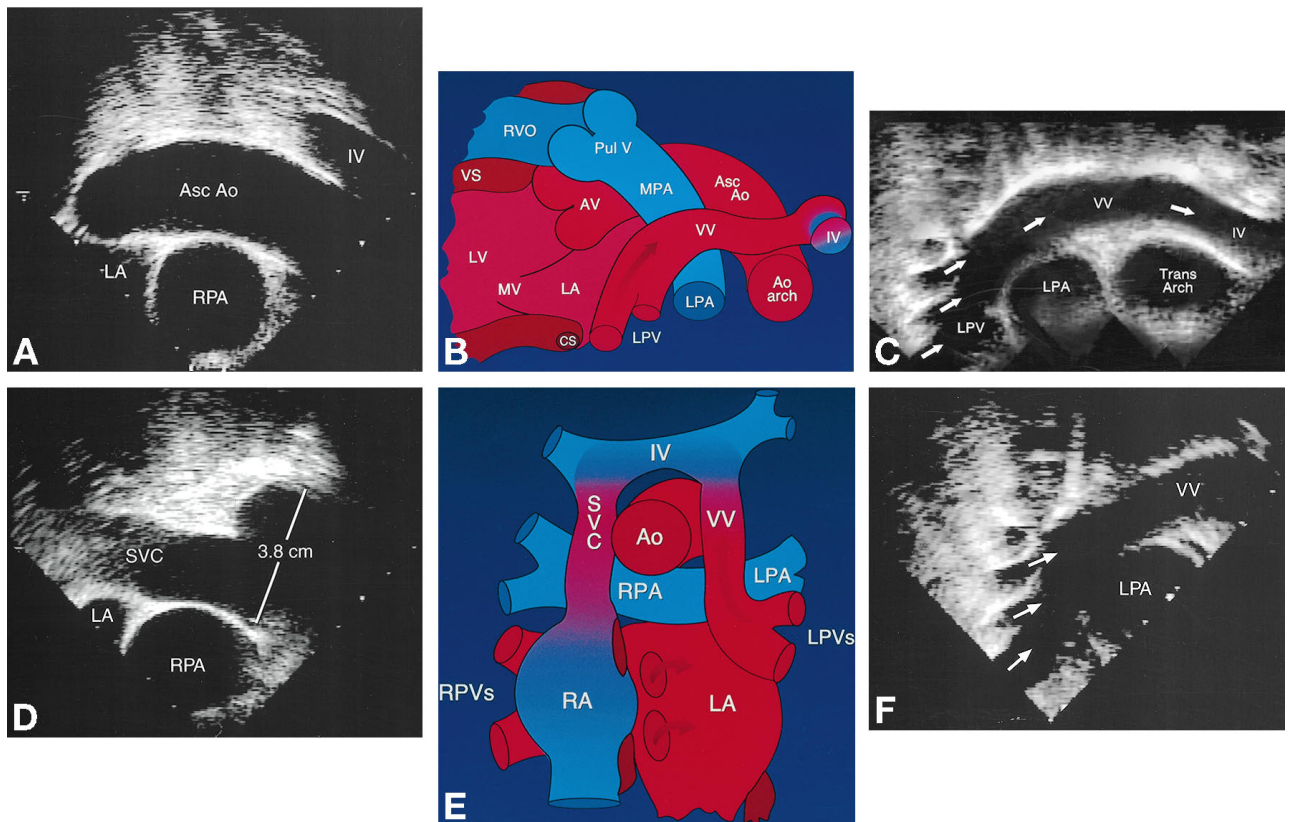
heart murmur (11 patients [24.5%]), supraventricular arrhythmia (9 patients [20%]) and decreased exercise tolerance (3 patients [6.7%]).

**Transesophageal echocardiography.** Sixty-six anomalous pulmonary venous connections were demonstrated by TEE in 43 patients (1.5/patient), including 46 in Group I and 20 in Group II. PAPVC could not be completely defined but was suspected from TEE in two patients. There was a single anomalous connecting PV in 23 patients (53.5%), two veins in 18 (41.9%), three veins in 1 (2.3%) and four veins in 1 (2.3%).

Anomalous pulmonary venous connections were right-sided in 35 patients (81.4%), left-sided in 7 (16.3%) and bilateral in 1 (2.3%). The connecting site, as shown in Table 1, was the SVC in 39 PVs (59.1%), RA-SVC in 6 (9.1%), RA in 8 (12.1%), inferior vena cava in 1 (1.5%) and coronary sinus in 2 (3.0%). In addition, there were 10 anomalous left PVs (15.1%) that connected to the innominate vein by means of a vertical vein before communicating with the SVC (Table 1).

The most common associated congenital heart defect was sinus venosus ASD in 22 patients (49%). Among those, 20 patients (91%) had a right upper PV anomalously connected to the SVC. Of the remaining 23 patients, 6 had secundum ASD (13.3%); 4 had a patent foramen ovale that was isolated in 2 (4.4%); and 15 had an intact atrial septum (33.3%).

**Surgical repair.** Thirty-one patients (68.8%) underwent surgical repair. PAPVC was confirmed in all patients (100%), including the two whose TEE was suggestive, but not diagnostic, of PAPVC. Five patients of this surgical group (16%) had six additional anomalous right PVs communicating with the RA that had not been described at TEE. Of the 14 patients not operated on, 6 had significant pulmonary hypertension. Four



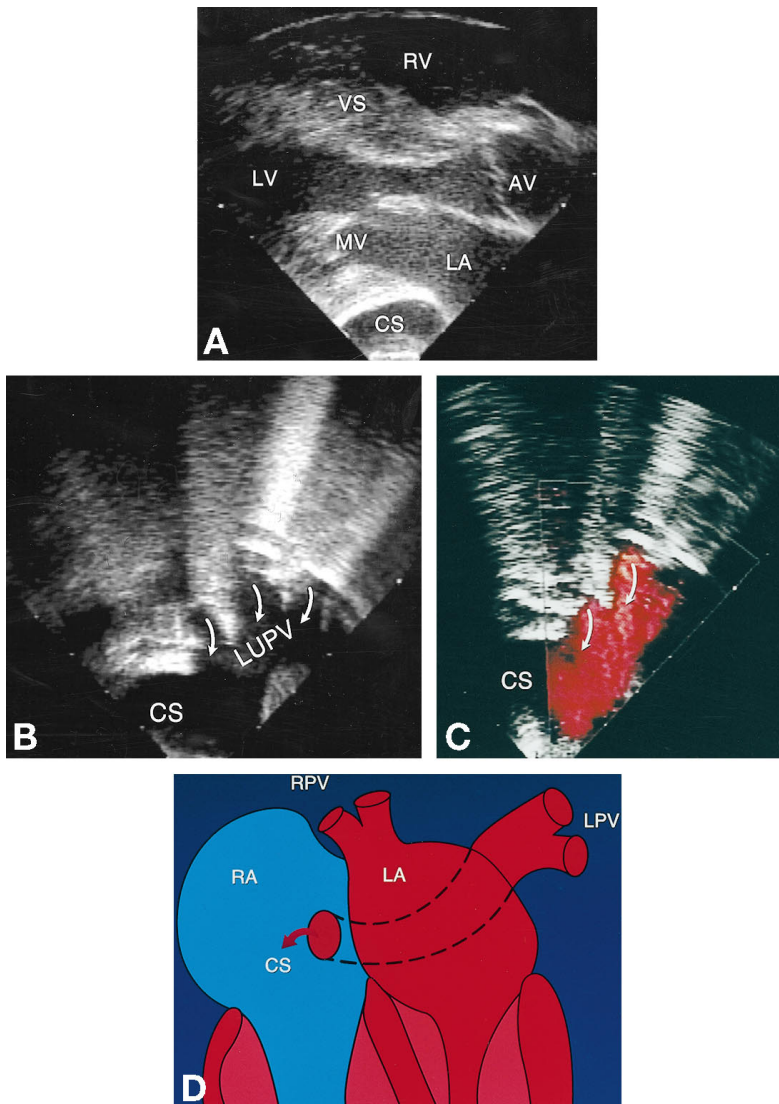
of these patients had a left-sided PAPVC, and three had an intact atrial septum.

### Discussion

Previously published autopsy reports (2,18) indicated that the most common form of PAPVC is a right-sided PV to the SVC or to the RA, followed by a left-sided PV to the vertical-innominate vein. Anomalous connection of the left-sided PVs are much less common than anomalous right-sided veins (2,18,19). In our series, 54 (81.8%) of the 66 PAPVCs originated from the right lung and 12 (18.2%) from the left lung, with the most common PAPVC involving the right upper PVs to SVC in 32 (48.5%). There are reports of rare PAPVCs to the left subclavian vein (18), brachiocephalic vein (20), azygos vein (21), portal vein (22) and coronary sinus (13,23). In our series, one patient had a PAPVC to the coronary sinus. The most common variant, reported to occur in up to 24% of autopsy studies of patients with PAPVC, is the presence of a single PV draining the entire right or left lung (2). Four (9%) of our patients had a common left-sided anomalous PV (Table 1).

**Associated congenital anomalies.** PAPVC is frequently associated with other congenital heart disease, most commonly ASD, in >75% of patients (19,24). It is estimated that 10% to 15% of patients with ASD and up to 85% of patients with sinus

**Figure 4.** Anomalous connection of the left PVs to a vertical vein. **A**, With the transesophageal array in the longitudinal plane (90°) and the shaft of the scope rotated rightward to view the long axis of the ascending aorta (Asc Ao), the dilated innominate vein (IV) is viewed anteriorly. The innominate vein is usually not seen in this view and is only evident because of its dilation. Posterior to the ascending aorta is the dilated volume-overloaded right pulmonary artery (RPA) and dome of the LA. **B**, Lateral view looking toward the left surface of the heart. The left PVs (LPV) are shown to be connected to a vertical vein (VV) coursing anterior to the left pulmonary artery (LPA) and ultimately communicating with the innominate vein. The vertical and innominate veins lie anterior and superior to the transverse aortic arch (Ao arch). **C**, Wide-field computer composite. These views were obtained by progressive withdrawal of the transducer up the esophagus from the transesophageal projection seen in the **right upper panel**. Shown is the complete course of the vertical vein (VV), which received the left PVs and empties into the innominate vein (IV) anterior to the transverse aortic arch (Trans arch). **D**, Further rightward rotation of the scope shaft results in a long-axis view of the SVC. Note that the superior portion of the SVC is markedly dilated (3.8 cm) because of volume overload where the innominate vein enters the SVC. **E**, Frontal view of the heart. The left PVs connect to a vertical vein (VV) that enters the innominate vein (IV). The innominate vein enters the normal SVC and delivers the left pulmonary venous blood to the RA. **F**, With the transesophageal array in the longitudinal plane at 110° angulation (see Fig. 1) and the shaft of the scope rotated far laterally, the anomalously connecting left PVs (LPV) (**arrows**) communicate with a vertical vein (VV) coursing toward the patient's head anterior to the left pulmonary artery (LPA). Ao = transverse ascending aorta; AV = aortic valve; CS = coronary sinus; LV = left ventricle; MV = mitral valve; PULV = pulmonary valve; RVO = right ventricular outflow; VS = ventricular septum; other abbreviations as in Figure 1.



**Figure 5.** Anomalous left pulmonary venous connection to the coronary sinus. **A**, Transesophageal long-axis view of the mitral (MV) inflow. Note the presence of a dilated coronary sinus (CS) in the mitral atrioventricular groove. Anterior to the coronary sinus is the LA. The other normal structures visualized are the left ventricle (LV), ventricular septum (VS), aortic valve (AV) and right ventricle (RV). **B**, The shaft of the transesophageal scope is rotated laterally while continuously visualizing the coronary sinus (CS). The PVs (arrows) are visualized entering the coronary sinus. **C**, Color flow Doppler demonstrating flow (arrows) toward the esophageal transducer from the PVs (arrows) to the coronary sinus (CS). **D**, Anatomic arrangement of PAPVC connection to the coronary sinus (CS). The left PVs (LPV) are shown connected to the coronary sinus, whereas the right pulmonary veins (RPV) connect normally to the LA. LUPV = left upper PVs.

venous ASD have PAPVC (19,24,25). Exclusive of ASD, other more complex cardiac malformations occur in ~20% of patients (26). PAPVC can also be an isolated defect with intact atrial septum (19,27). In our study of uncomplicated PAPVC, the most common associated anomaly was an ASD in 30

patients (66.7%); 22 having sinus venosus ASD (49%). Fifteen patients (33.3%) had an intact atrial septum.

**Pulmonary hypertension.** The physiologic disturbance caused by PAPVC depends on the number of anomalous pulmonary venous connections involved, site of the connec-

**Table 1.** Site of Anomalous Pulmonary Vein Connection

Origin	Connecting Site						Total
	SVC	RA-SVC	RA	RA/IVC	VV/INV	CS	
RUL	32	—	2	1	—	—	35
RML	7	6	4	—	—	—	17
RLL	—	—	2	—	—	—	2
LUL	—	—	—	—	4	—	4
LUL + LLL	—	—	—	—	3 + 3	1 + 1	8
Total	39	6	8	1	10	2	66

CS = coronary sinus; LLL = left lower lobe; LUL = left upper lobe; RA = right atrium; RA/IVC = right atrium/inferior vena cava; RA-SVC = right atrial-superior vena cava junction; RLL = right lower lobe; RML = right middle lobe; RUL = right upper lobe; SVC = superior vena cava; VV/INV = vertical vein/innominate vein.



tion, status of the pulmonary venous bed and the presence of ASD or other congenital heart disease (26,28). Each normally connecting PV contributes an average of 25% of the total pulmonary blood flow. However, when a PV connects anomalously to the RA or SVC, blood is preferentially shunted to this anomalous vein because of the lower RA pressure, compared with LA pressure, producing significant volume overload. This is especially true in the presence of systemic hypertension, mitral valve disease or left ventricular dysfunction, which increases LA pressures. Isolated PAPVC with intact atrial septum may also be associated with pulmonary hypertension (29). Those patients with right ventricular volume overload will present with dyspnea, new murmur and supraventricular arrhythmia very similar to patients with ASD.

**Diagnostic modalities.** Before the mid-1980s, the diagnosis of PAPVC was made by cardiac catheterization, angiography in combination with dye dilution curve or contrast TTE (5,30,31). MRI and computed tomographic scan have also been reported in small series to be diagnostic in patients with PAPVC (6,7,32,33). When MRI was performed in older patients, the detection rate of the PV was low (34).

Two-dimensional TTE, combined with color flow Doppler, has been reported (35,36) as useful in diagnosing PAPVC, particularly in the pediatric age group. However, with adults, image acquisition is more difficult. As a result, TEE has gained widespread acceptance as an excellent diagnostic technique in congenital heart disease, especially for evaluation of the atrial septum and pulmonary venous connections (12-14,16,17,37-40). Nonvisualization of normal pulmonary venous commitment to the LA is a strong suggestion that the nonvisualized vein is anomalously connected (13). However, anatomic variation of the normal venoatrial connection does occur (17). Therefore, the demonstration of four PVs entering the LA does not always exclude the presence of PAPVC because more than four PVs may be found due to separate incorporation of segmental branches into the LA (2,17,19,38). In our own experience, anatomic variants have not been a major problem in clinical diagnosis. In 1989, Sutherland reported that defining normal PVs by TEE was 100% for left upper PV, 62% for left lower PV, 90% for right upper PV and 23% for right lower PV (39). Stumper et al. (13) demonstrated all four pulmonary venous connections in 91% of patients in his series. With current instrumentation, identification of normal pulmonary venous connection (as described in Methods) is virtually 100%.

TEE detected 66 anomalous PVs in 43 patients, and PAPVC was suspected in the 2 remaining patients. When TEE was done for diagnostic purposes (Group I), it demonstrated at least one PAPVC in 29 (93.5%) of 31 patients. Furthermore, in the subgroup of patients who subsequently underwent surgical repair (n = 31), the diagnosis of PAPVC was confirmed in all 29 patients whose TEE was diagnostic (93.5%) and in the 2 whose TEE suggested this anomaly (6.5%). All 49 PAPVCs detected by TEE preoperatively were confirmed at the time of operation (100%).

This large retrospective study is not intended to study the frequency of normal or abnormal pulmonary venous connec-

tion. However, it does demonstrate that with proper technique, identification of the normal and abnormal pulmonary venous connections, especially in patients with unexplained right ventricular volume overload, can be made with confidence. TEE made or suspected the diagnosis of PAPVC in all patients. TEE should probably be considered before transcatheter ASD closure, especially if right ventricular volume overload is out of proportion to the size of the defect. This also has important surgical implications in terms of surgical planning. When a sinus venosus ASD or a right-sided PAPVC is identified before operation, it is the standard practice in our center to open the pleura anteriorly on the right and directly inspect the entire pulmonary venous return from the right lung. A different surgical approach is undertaken when a left-sided PAPVC is identified. Equally important is the confident identification of normal venous connections. Accurate preoperative or intraoperative identification of PVs simplifies and shortens the operative procedure.

**Conclusions.** TEE has expanded the diagnostic anatomic, functional and hemodynamic role of echocardiography in congenital heart disease. The diagnosis of PAPVC has always been challenging, and invasive angiography is expensive and inappropriate for routine assessment. This large review describes the assessment of normal and abnormal pulmonary venous connections using TEE and demonstrates that TEE is a sensitive method for the detection of PAPVC and can obviate angiography.

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