

Partial Anomalous Pulmonary Venous Return

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Partial anomalous pulmonary venous return (PAPVR) is a heterogeneous group of lesions where at least one, but not all, of the pulmonary veins drain into the right heart or systemic veins. Similar to total anomalous pulmonary venous return (TAPVR), PAPVR can be associated with a wide variety of intracardiac lesions, particularly when associated with heterotaxy syndrome or anomalies of atrial and abdominal arrangement. Individualized multimodality imaging and surgical strategies are often required as a result of the broad anatomical variants of PAPVR.

The most common PAPVR variant is drainage of the right upper pulmonary vein into either the RA, the right atrial-SVC junction, or into the SVC. Other variants include drainage of some of the left pulmonary veins into the innominate vein or all right pulmonary veins into the right atrial-IVC junction via a descending vertical vein (“scimitar” syndrome).

The majority of patients with PAPVR will have an associated ASD, usually of the sinus venosus type. These ASDs are close to the right/posterior wall of the septum with no posterior rim. PAPVR of the right upper pulmonary veins to the SVC tends to be associated with a superior sinus venosus ASD while PAPVR of the right veins into the IVC tends to present with an inferiorly located sinus venosus ASD.

Pathophysiology and Clinical Presentation

The pathophysiology of PAPVR is related to the left-to-right shunt and/or, very rarely, obstruction to pulmonary venous drainage from the lungs. Mechanisms of pulmonary venous obstruction, if present, are similar to TAPVR and due to: 1) stenosis at the connection of the anomalous veins to the RA or systemic vein, 2) mechanical compression from the PAs, bronchi, or diaphragm, and/or 3) intrinsic long segment vessel hypoplasia. Similar to other supraventricular left-to-right shunts, PAPVR results in cardiomegaly with right-heart dilation. The degree of right-heart dilation is proportional to the number of pulmonary venous segments involved and can be additive to any other associated shunts (e.g., ASD). Chronic RA and RV dilation over years or decades can increase the risk for arrhythmias and RV systolic and diastolic dysfunction.

Children with isolated, unobstructed PAPVR are usually asymptomatic. These patients tend to be evaluated due to idiopathic right-heart dilation. Some patients may present with recurrent respiratory infections. On physical exam, patients will have a systolic ejection murmur on the left superior sternal border (related to increased pulmonary valve flow) and a widely split S₂.

Diagnosis

- **ECG.** Patients may have an incomplete right bundle branch block, possible RVH, and possible RA enlargement.
- **CXR.** Will show cardiomegaly and increased pulmonary vascular markings.

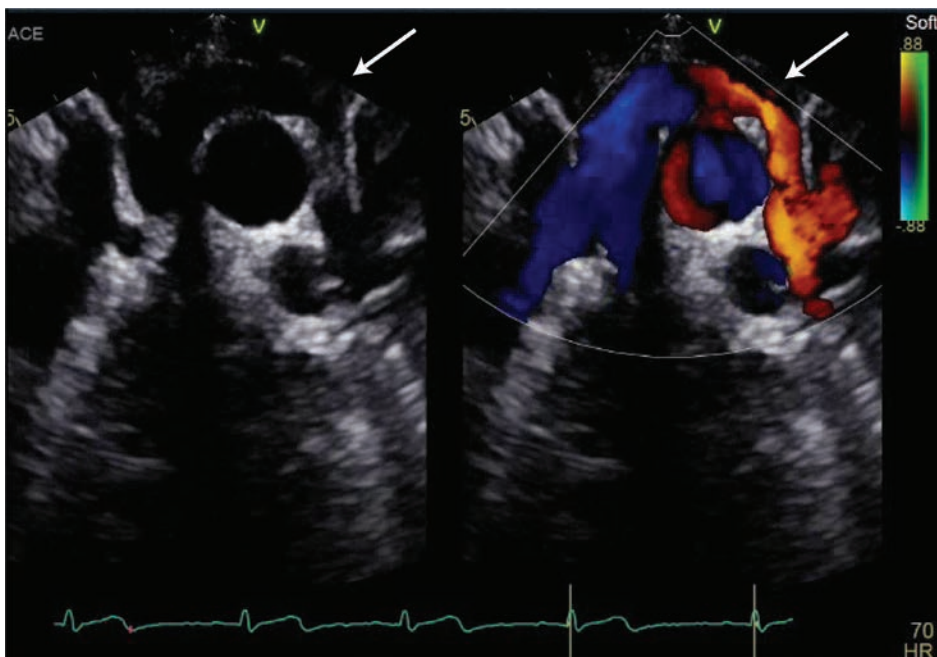


Figure 9-1. Echocardiographic suprasternal color-compare image of an anomalous left upper pulmonary vein draining into the left brachiocephalic vein (arrow). Image courtesy of Dr. Josh Kailin, www.pedecho.org.

- **Echocardiogram.** There is usually right atrial and ventricular dilatation. The defect may not be obvious on the standard parasternal views. Subcostal coronal views can demonstrate the entry point of the right upper pulmonary vein. If the defect is higher in the SVC, a high parasternal long axis may demonstrate the defect. Mild-to-moderate TR may be present. Pulmonary valve flow acceleration secondary to left-to-right shunting can mimic mild pulmonary valve stenosis (velocity no more than 2.5-3 m/s) but the valve should have normal appearance. Like in other atrial loading defects, there will be interventricular septal flattening during diastole. If there is additional septal flattening in systole, pulmonary hypertension should be suspected. Echocardiography can also show an anomalous left upper pulmonary vein draining into the left innominate vein (Figure 9-1).
- **CTA and MRI.** Cross-sectional imaging may be helpful to delineate the anatomy of the anomalous veins for surgical planning (Figure 9-2). It is particularly useful for surgical planning in patients with scimitar syndrome to define the location of the vein with respect to the right and left atria.
- **Cardiac catheterization.** Not needed in the vast majority of patients with PAPVR. Cardiac catheterization is indicated in patients with scimitar syndrome in order to embolize aortopulmonary collaterals (that are almost always present in these patients) prior to surgical intervention (see below).

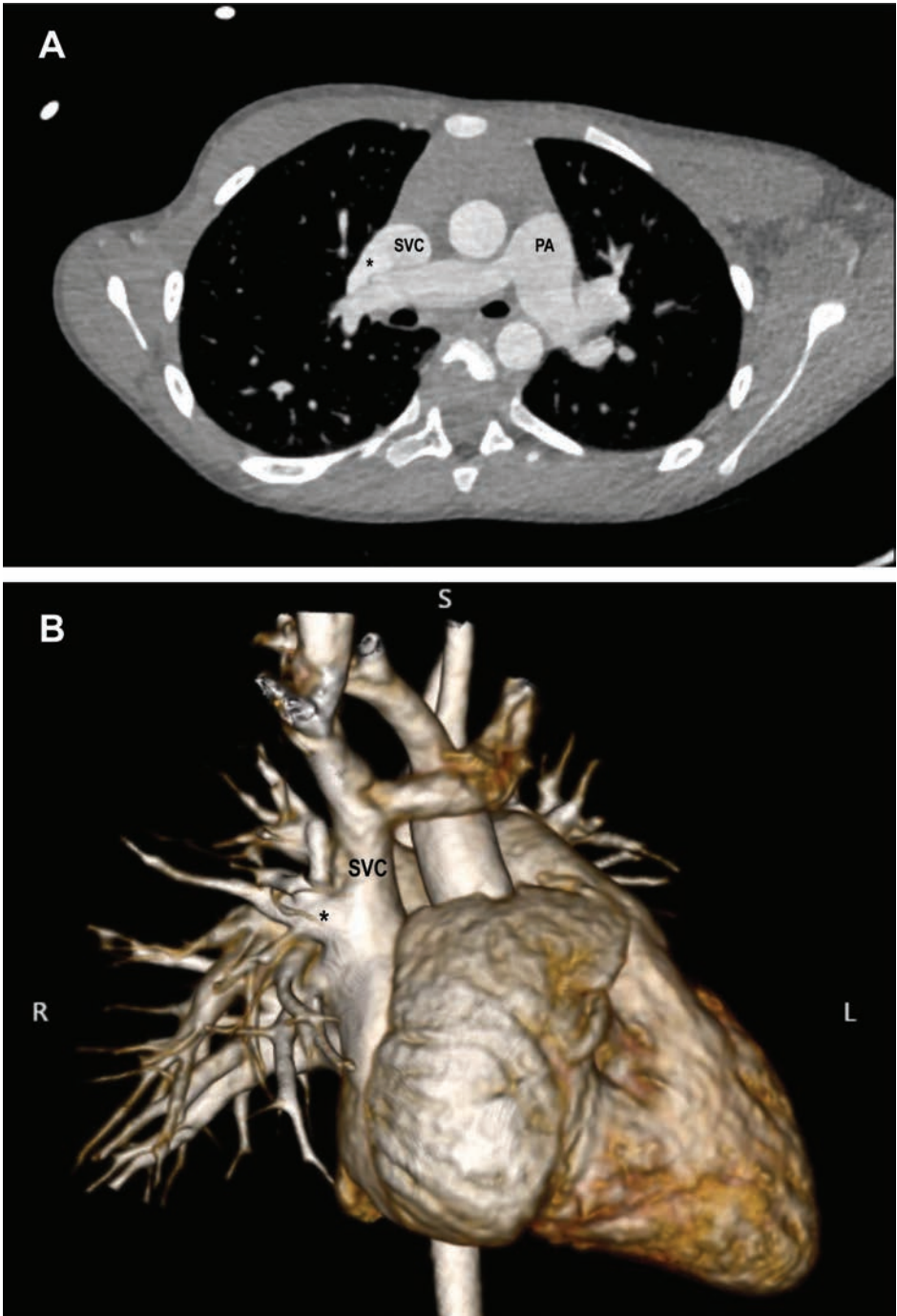


Figure 9-2. Axial (A) and 3D-reconstruction CTA images of a patient with PAPVR of the right upper pulmonary vein (*) to the SVC.

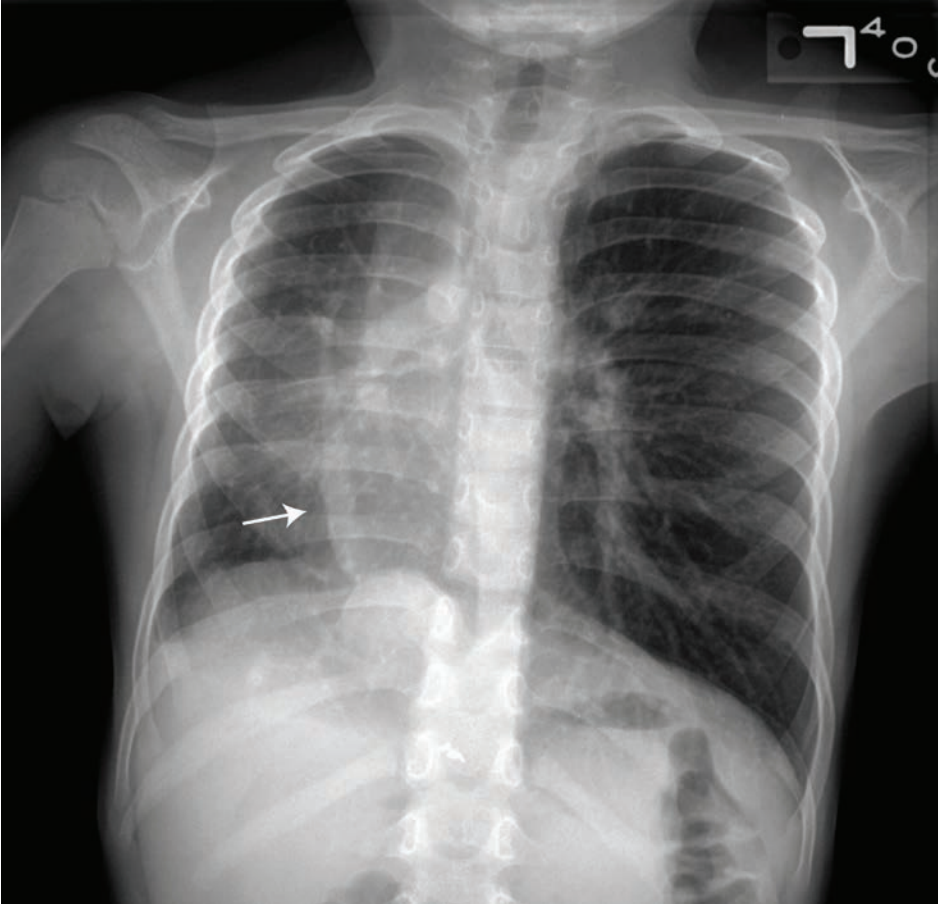


Figure 9-3. CXR of a patient with scimitar syndrome showing right lung hypoplasia and the shadow of the anomalous right upper pulmonary vein descending towards the IVC (arrow).

Indications / Timing of Intervention

Indications for surgical intervention are dependent on the degree of pulmonary overcirculation. Isolated single anomalous pulmonary veins may not result in enough right-heart dilation to warrant intervention. In borderline cases, MRI estimates of Qp:Qs can be helpful in determining indications for intervention. Patients with low-volume pulmonary overcirculation (Qp:Qs <1.5:1) likely do not require surgical correction and long-term cardiology follow-up is needed to evaluate for sequelae of pulmonary overcirculation. On the contrary, little is known regarding the long-term effect of right-heart volume overload. In such borderline cases, it is important to have a balanced view of the natural history of the condition against the surgical risks.

Elective surgical intervention for isolated PAPVR is optimally performed later in life (school age). In patients that require neonatal surgery for other intracardiac lesions,



Figure 9-4. 3D-reconstruction CTA image of a patient with scimitar syndrome showing the anomalous right pulmonary vein (blue) descending and inserting into the IVC.

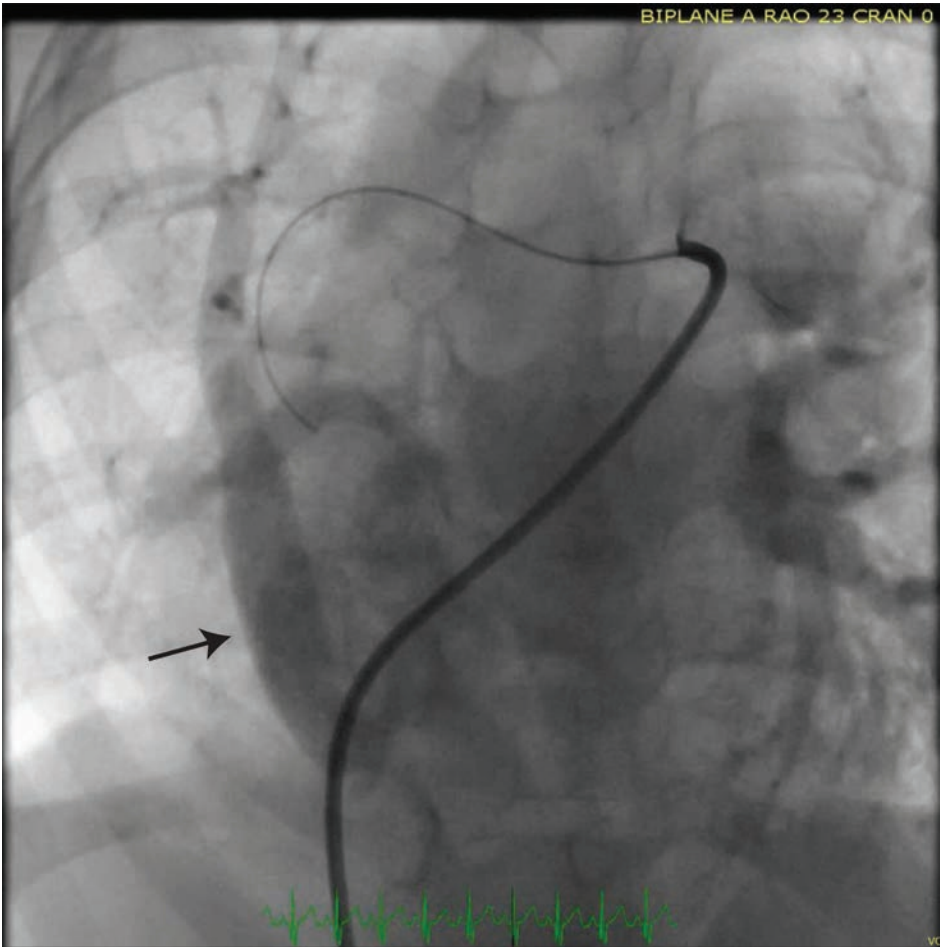


Figure 9-5. Cardiac catheterization showing a prominent scimitar vein (arrow).

PAPVR of a single isolated pulmonary vein may be left alone in order to optimize surgical results of manipulation, redirection, and/or reimplantation of the anomalous vein.

Surgical Repair

The specifics of the surgical repair depend on the particular type of PAPVR. Most procedures are performed via median sternotomy and under cardiopulmonary bypass with cardioplegic arrest.

Anomalous Left Pulmonary Vein Into The Innominate Vein

The anomalous vein may be left in situ, ligated at its entrypoint into the innominate vein, and a longitudinal anastomosis performed between the anterior surface of the pulmonary vein and the left atrial appendage, which is laid on top of the vertical vein.

Alternatively, the anomalous vein may be detached and anastomosed directly to the LA appendage with care not to torque these structures.

Anomalous Right Upper Pulmonary Vein

For patients with anomalous right pulmonary veins into the RA, an intracardiac baffle is created with autologous pericardium to redirect the flow through the sinus venosus ASD (if present) or through a created defect (if no ASD present) toward the LA. If the anomalous veins drain into the SVC and creating a baffle would potentially obstruct the SVC, there are 2 different surgical options: a Warden procedure and a two-patch repair. For the Warden procedure, the SVC is divided just above the entrypoint of the pulmonary veins and sutured close (with or without a patch). The SVC orifice is baffled through the atrial septum towards the LA and the distal SVC is reimplanted into the right atrial appendage to provide drainage of the SVC. If there is significant tension due to a short SVC, a posterior anastomosis may be created and an anterior patch placed on the anastomosis to relieve tension. In the two-patch repair, a baffle is created between the pulmonary veins and the LA, and a second patch is placed to enlarge the SVC and prevent obstruction. This repair may be associated with a higher incidence of sinus node injury (or injury to the blood supply of the sinus node), especially in older patients.

Complications

Potential complications of surgical repair of PAPVR include:

- **Arrhythmias and sinus node dysfunction.** Sinus node dysfunction may complicate PAPVR repair. Long-term freedom from dysfunction appears to be similar when comparing both surgical techniques.
- **Postoperative stenosis.** Patients undergoing repair of PAPVR of the right upper pulmonary veins can develop postoperative stenosis in the baffle between the anomalous pulmonary veins and the LA. Additionally, the connection between the SVC and the RA in a Warden procedure is also vulnerable to developing postoperative stenosis. Both potential complications should be evaluated with routine postoperative echocardiograms.
- **Reintervention.** Smaller size and younger age are the dominant predictors of reintervention after repair of PAPVR for both, the two-patch technique and the Warden procedure.

Scimitar Syndrome

Scimitar syndrome is rare variant of PAPVR with a wide variety of presenting scenarios. It is characterized by right pulmonary veins draining anomalously into the IVC and associated variable degrees of right-lung hypoplasia. Additional findings can include pulmonary hypertension and/or pulmonary sequestration with an aortopulmonary collateral from the descending aorta. An inferior sinus venosus or secundum ASD is often present. Some patients present early in life or infancy with pulmonary hypertension and symptoms out of proportion to the PAPVR. Some patients are incidentally diagnosed in adolescence or adulthood, often when a CXR identifies the characteristic scimitar vein, which appears like the 8th century Turkish sword of the same name (Figure 9-3).

The pathophysiology of scimitar syndrome is secondary to a combination of the left-to-right shunting (PAPVR, ASD, aortopulmonary collateral) and pulmonary vascular disease, which is likely secondary to a combination of the pulmonary overcirculation, pulmonary hypoplasia, and pulmonary venous obstruction.

Diagnosis and procedural planning depend on a multimodality imaging strategy. TTE usually makes the diagnosis of scimitar syndrome and estimates right heart pressures. Cardiac MRI or CTA is needed to characterize the PAPVR, particularly the insertion into the IVC and the relationship with the RA or LA (Figure 9-4). Cardiac catheterization can measure PA pressures directly and confirm the anatomy of the PAPVR (Figure 9-5). However, cardiac catheterization is best used for coiling of the aortopulmonary collaterals that are commonly present from the descending aorta to the right lung.

Indications for surgical intervention can be controversial, particularly in asymptomatic older patients with no significant pulmonary overcirculation ($Q_p:Q_s < 1.5:1$). Younger presentation is typically associated with more severe disease and neonatal presentation tends to be associated with poor outcome, with or without surgical intervention. Timing of intervention can also be controversial, weighing the consequences of ongoing pulmonary hypertension and left-to-right shunting versus the benefits of somatic growth and improved surgical outcomes.

Surgical intervention is individualized for each particular patient. Options include creation of a long baffle between the entrypoint of the scimitar vein into the LA (through an existing or created ASD), reimplantation of the scimitar vein higher into the RA followed by creation of a shorter baffle, creation of a side-to-side anastomosis between the vein and the RA with creation of a baffle, or reimplantation of the anomalous vein directly into the LA. Baffle stenosis is a long-term consideration after these surgical repairs, especially if a long baffle is created.

Suggested Readings

Binsalamah ZM, Ibarra C, Edmunds EE, et al. Younger age at operation is associated with reinterventions following the Warden procedure. Presented at AATS 2019. Submitted for publication, 2020.

Bonilla-Ramirez C, Saliccioli KB, Adachi I, et al. Smaller right pulmonary artery is associated with longer survival time without scimitar vein repair. Presented at CHSS 2019. Submitted for publication, 2020.

Supracardiac PAPVR repairs (1996-2019)

(Binsalamah et al. 2020)

Number of patients: 158

- Warden procedure: 122 (78%)
- Single-patch repair: 26 (22%)

Median age at operation: 6.3 years

Median length of stay: 4 days

Perioperative mortality: 1 (0.6%)

Long-term outcomes (median follow-up 3.8 yrs):

- Cath-based interventions: 10 (6%)
- Reoperation (patch dehiscence): 1 (0.6%)

Scimitar syndrome diagnoses (1995-2019)

(Bonilla-Ramirez C, Saliccioli KB, et al. 2020)

Number of patients: 61

Median age at diagnosis: 1.3 years

Surgical repair: 23 (38%) patients:

- Intracardiac baffle: 13
- Reimplantation: 9
- Other: 1

Postoperative 5-year freedom from stenosis: 70%

Postoperative 5-year survival: 95%