

# Pulmonary Vein Anomalies

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This chapter encompasses 2 different lesions of the pulmonary veins: total anomalous pulmonary venous return (TAPVR) and isolated pulmonary vein stenosis. Partial anomalous pulmonary venous return (PAPVR) is covered in Chapter 9.

## TAPVR

TAPVR is a lesion characterized by the anomalous drainage of all pulmonary veins into the RA or a systemic venous structure, instead of into the LA. Embryologically, TAPVR is due to a failure of fusion of the pulmonary venous lung buds with the posterior outpouching of the LA. As a consequence, there is no connection between the pulmonary veins and the LA, but the veins usually maintain a connection to the systemic veins through an ascending or descending “vertical vein”.

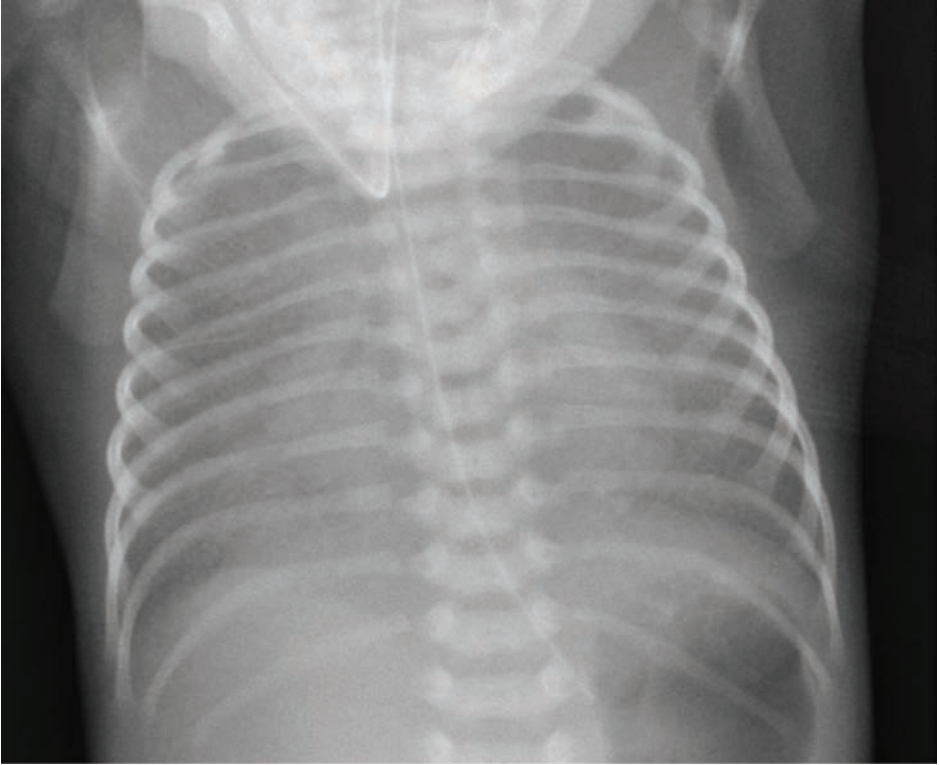
## Anatomical Considerations

In most cases of TAPVR, pulmonary veins drain into a pulmonary venous confluence that then drains into either the RA or a vertical vein. TAPVR is classified into 4 categories depending on the site of drainage:

- **Supracardiac (45%).** There is usually a horizontal pulmonary venous confluence behind the LA that drains into a vertical vein that ascends and connects to either the SVC or more commonly, the innominate vein. The vertical vein may travel anterior to the left PA or between the left PA and the left bronchus, where it may become compressed by these structures. Compression at this level may lead to obstructed physiology and pulmonary hypertension, which in turns dilates the PA and causes of further obstruction (hemodynamic “vise”).
- **Cardiac (25%).** The anomalous veins drain directly into the RA or via a dilated coronary sinus. In general, cardiac TAPVR is unobstructed and often presents later in infancy.
- **Infracardiac (25%).** The veins usually drain into a vertical pulmonary venous confluence that drains into a vertical vein that travels down through the diaphragm and drains into the IVC or into the portal venous system or hepatic veins. The vast majority of these patients have obstruction of the pulmonary venous return due to the high resistance of the hepatic sinusoids, compression at the level of the diaphragm, and/or stenosis at the entrapment into the IVC and as such, require emergent intervention.
- **Mixed (5%).** Different pulmonary veins have different types of drainage.

## Pathophysiology and Clinical Presentation

The clinical presentation of patients with TAPVR depends whether there is significant obstruction of the pulmonary veins or not. Since pulmonary veins normally drain into the LA through a large and unobstructed confluence, having all pulmonary veins drain into a smaller vertical vein is not optimal. As such, it is safe to assume that every patient

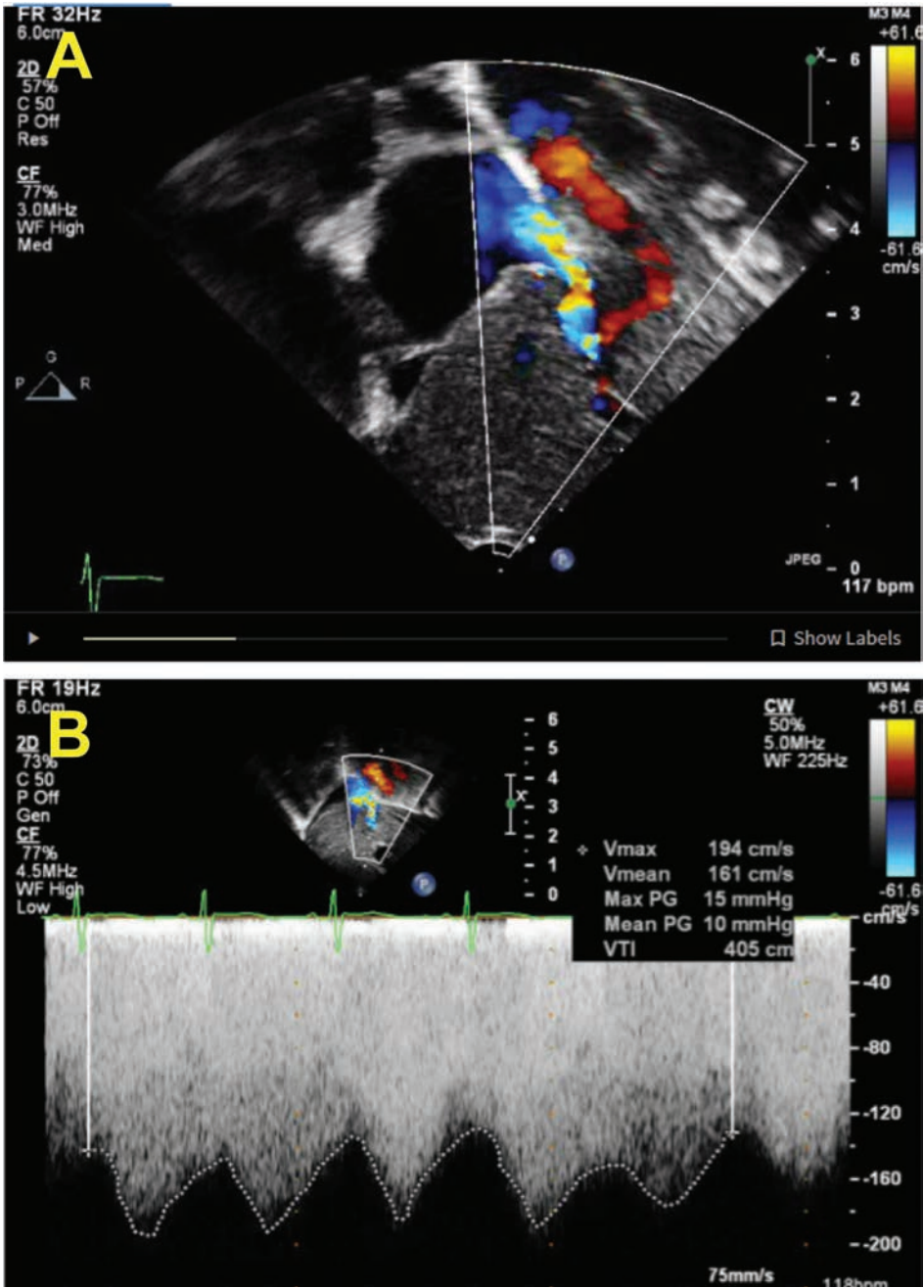


**Figure 10-1.** CXR of a patient with obstructed TAPVR showing severe pulmonary edema.

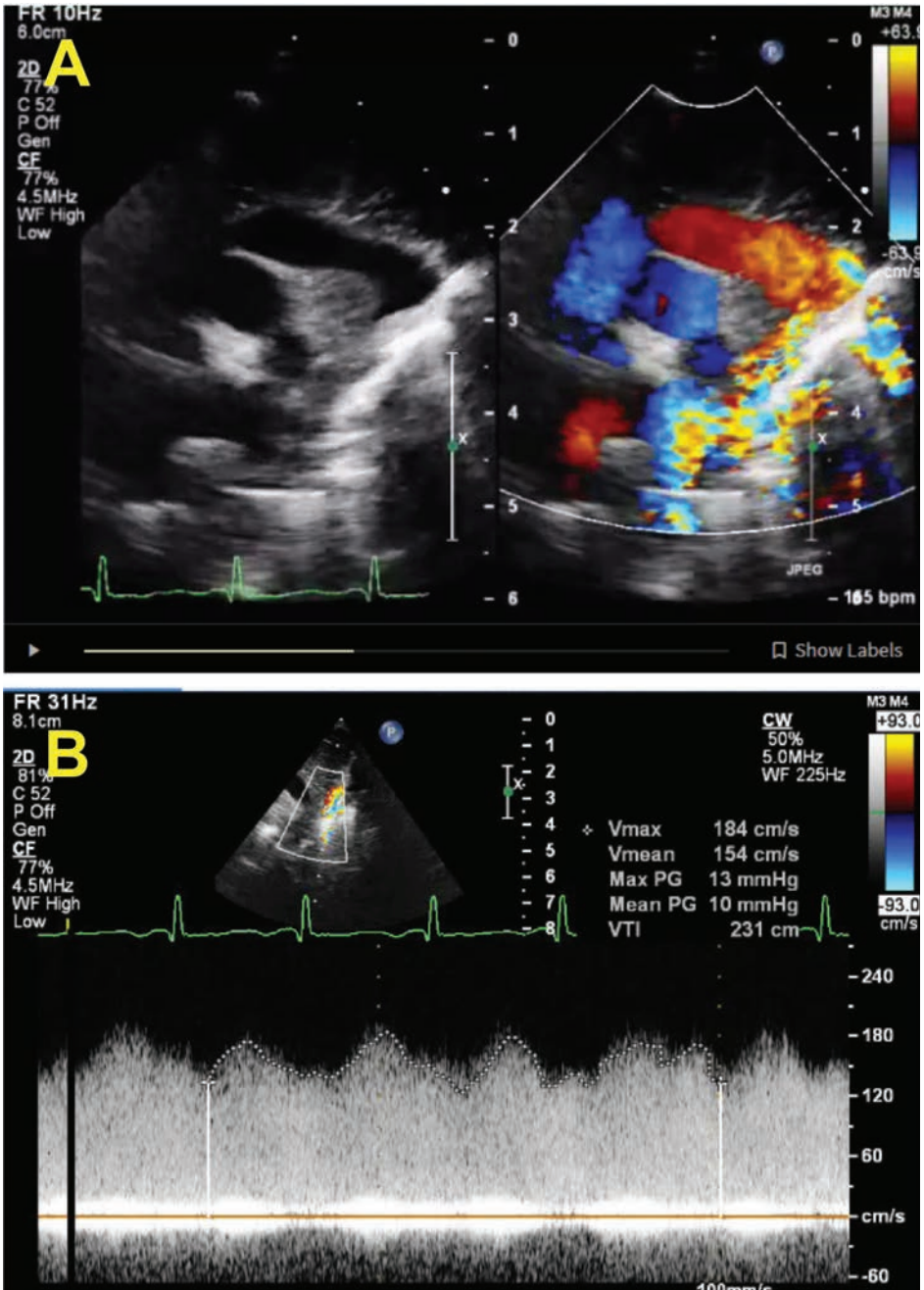
with TAPVR has some degree of obstruction, even if the obstruction is not clinically significant.

Fetal diagnosis of TAPVR is often difficult as the blood is shunted away from the lungs and oxygenated through the placenta. There is therefore little flow through the pulmonary veins and thus they are difficult to delineate. At times, anomalous pulmonary veins will be diagnosed in utero when there are other cardiac defects such as in patients with heterotaxy syndrome. Due to the difficulty of prenatal diagnosis, patients with TAPVR often present postnatally.

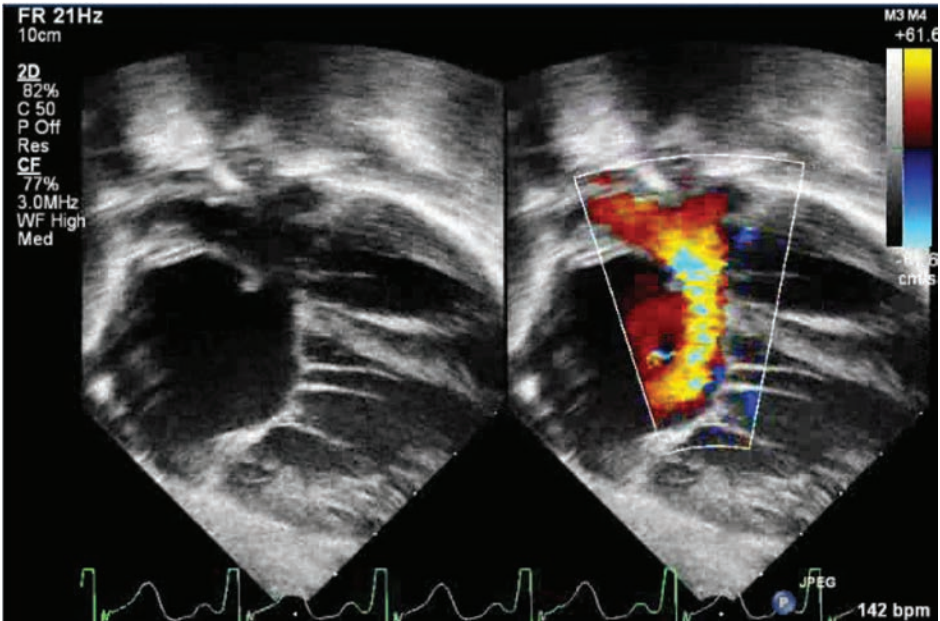
Patients with significant obstruction will present immediately after birth with significant pulmonary edema, severe and progressive cyanosis, pulmonary hypertension, metabolic acidosis, and frequently cardiogenic shock. Often, these neonates are treated as having persistent pulmonary hypertension of the newborn (PPHN) and are found to have TAPVR when an echocardiogram is performed as part of their workup. If the pulmonary veins are not obstructed, these children present in the first few months of life with signs of heart failure due to pulmonary overcirculation (increased left-to-right shunt) and cyanosis due to intracardiac mixing.



**Figure 10-2.** Echocardiogram of a patient with infracardiac TAPVR. The descending vein is shown coursing through the diaphragm and turning into the hepatic venous system and then into the right atrium (A). Spectral Doppler pattern of stenotic connection from descending vertical vein to hepatic venous system (B). Images courtesy of Dr. Josh Kailin, [www.pedecho.org](http://www.pedecho.org).



**Figure 10-3.** Echocardiogram of a patient with supracardiac TAPVR. The ascending vein is noted entering into the left innominate vein, which in turn drains into the SVC (A). There is obstruction between the left PA and left bronchus, as noted in the Doppler pattern (B). Images courtesy of Dr. Josh Kailin, [www.pedecho.org](http://www.pedecho.org).



**Figure 10-4.** The classic “whale’s tail” seen in anomalous pulmonary veins draining into the coronary sinus with flow into the RA.

### Diagnosis

- **CXR.** Patients with significant obstruction will show severe pulmonary edema (Figure 10-1) whereas patients with no obstruction will demonstrate evidence of pulmonary overcirculation (cardiomegaly and increased pulmonary vascular markings). Older patients with supracardiac TAPVR to the innominate vein may show what has been described as the “snowman sign” with the upper silhouette formed by the vertical vein on the left and the dilated SVC on the right.
- **Echocardiography.** Diagnosis usually relies on the initial echocardiogram. A sign that there is TAPVR is purely right-to-left shunting across the atrial level communication (PFO or ASD). There needs to be delineation of whether the TAPVR is supracardiac, infracardiac, cardiac (to the coronary sinus or to the right atrium directly), or mixed. Each pulmonary vein should be imaged, the insertion into the confluence or ascending/descending vein described, and the course of the ascending or descending vein should be traced to localize areas of obstruction. By definition, patients with infracardiac TAPVR (Figure 10-2) are obstructed due to the long course to return to the RA and multiple sites of potential obstruction. Supracardiac TAPVR (Figure 10-3) can be obstructed, most frequently where the ascending vein courses between the bronchus and left PA. The image on echocardiography of cardiac TAPVR to the coronary sinus (Figure 10-4) is termed a “whale’s tail” due to the similarity of appearance. In supra- and infracardiac TAPVR, the ascending or descending vein is noted by echocardiography as a venous structure coursing away from the heart. In

supracardiac TAPVR often the ascending vein is noted coursing into the innominate vein (flow toward the transducer in suprasternal notch views) and in infracardiac TAPVR a vein is noted descending through the diaphragm rather than the expected venous course toward the heart (the sine qua non is “red” venous flow through the diaphragm when imaging in subcostal windows). Echocardiography should also focus on delineating other intracardiac lesions as TAPVR is frequently associated with heterotaxy syndrome and single-ventricle cardiac disease. In isolated TAPVR, RV pressure can be evaluated and the size of the LV, LVOT, and aortic arch should be assessed as there can be associated hypoplasia of left-heart structures due to limitations of in utero flow through the left side of the heart.

- **CTA.** If the pulmonary veins cannot be adequately delineated by echocardiography, they can be imaged using cardiac CTA, provided that the patient is stable. Surgical intervention should not be delayed in a patient in extremis in order to obtain a CTA unless there is a high suspicion of complete pulmonary venous atresia (uncommon). Timing of contrast administration must account for the fact that flow through the pulmonary veins may be significantly delayed if severely obstructed.

### Preoperative Management

Preoperative management of obstructed TAPVR involves hemodynamic stabilization and expedited progression to the OR for repair. Patients are often acidotic and can have hemodynamic compromise due to severe cyanosis. Prolonging preoperative medical management can lead to worsening of end-organ dysfunction. Significant ventilation and treatment of pulmonary hypertension can even worsen the clinical picture by worsening pulmonary edema. Inotropic support is indicated to maintain end-organ perfusion. Preoperative management of unobstructed TAPVR involves diuresis due to the increasing left-to-right shunt as neonatal PVR decreases.

### Indications / Timing of Intervention

The diagnosis of TAPVR is an indication for surgical intervention. Patients with obstructed TAPVR require emergent intervention. Patients with a diagnosis of unobstructed TAPVR undergo semielective repair prior to initial discharge of the hospital (if diagnosed in utero or in the neonatal period) or shortly after diagnosis, if diagnosed as an outpatient.

### Anesthetic Considerations

The anesthetic management of TAPVR is very dependent on patient anatomy and on the degree to which the anomalous pulmonary veins are obstructed.

### Preoperative

Obstructed TAPVR is a true neonatal emergency. The preoperative evaluation should focus on the patient’s anatomy, intravenous and intraarterial access, and the degree of pulmonary venous obstruction and resulting pulmonary edema. Medications should be reviewed, as many of these patients may require infusions of epinephrine, dopamine, vasopressin, and PGE. The echocardiogram should be reviewed for associated anomalies, as the anatomy will dictate whether the patient may need a systemic-to-pulmonary artery shunt, a PA band, arch advancement, or other procedure in addition to the repair

of the TAPVR. The CXR should be reviewed with special attention to the appearance of the lungs and the position of the umbilical arterial and venous lines. A recent arterial blood gas should be reviewed and the degree of hypoxemia and/or acidosis should be noted, as well as the lactate. Ventilator settings may also yield important information regarding the condition of the lungs.

### Intraoperative

A TEE is usually avoided during TAPVR repair because it can cause obstruction of the pulmonary venous confluence and make the operation more challenging. In the pre-CPB period in patients with obstructed TAPVR, oxygenation can become increasingly difficult and inhaled nitric oxide may be necessary to bring the  $PO_2$  to minimally acceptable values. Post-CPB, pulmonary hypertension and poor pulmonary mechanics may be encountered. iNO, milrinone, and epinephrine may be required to augment RV function and decrease PA pressures.

### Surgical Repair

The goal of surgical repair of TAPVR is to create a large anastomosis between the left atrial structures and the pulmonary venous confluence. The procedure is performed through a median sternotomy. Aorto-bicaval cannulation is preferred.

Circulatory arrest is uncommonly required, but cooling after initiation of CPB can be helpful if a short period of circulatory arrest is anticipated. While cooling, the pulmonary venous confluence is dissected from behind the pericardium and the pulmonary veins are identified. It is useful to place fine marking stitches on both the pulmonary venous confluence and the posterior aspect of the atrium while slightly filling the heart in order to define the optimal position of the anastomosis.

Once the goal temperature is reached, the heart is arrested. A right atriotomy is performed to visualize the atrial anatomy. The heart is usually retracted rightward in order to allow for visualization. Using intermittent DHCA, the vertical vein is ligated, an incision is performed on the anterior aspect of the pulmonary venous confluence, and a corresponding incision is created on the posterior aspect of the LA, sometimes extending into the LA appendage. An anastomosis is carefully created between the LA and the pulmonary venous confluence with running fine Prolene suture. Partial or full CPB flows may be intermittently used during this portion of the procedure. After the anastomosis is created, full CPB is reinitiated and the patient is rewarmed. The ASD is closed either primarily or with a small pericardial patch. An LA line may be placed through the LA appendage (if not used for the anastomosis) or through the RA and the sutureline of the ASD closure. The cross-clamp is removed and the right atriotomy closed.

It is important to note that the left heart in patients with TAPVR tends to be relatively small and noncompliant. As such, administration of even small amounts of fluid can significantly increase LAP and push the left heart beyond its Starling curve. An LA line allows judicious management of intracardiac volume. As such, CPB is carefully weaned making sure that LAP is kept low, even if that means tolerating a relatively lower BP. It is common for the left heart to slowly improve its output as one usually sees significant improvements in BP at the same intracardiac volume over the first 30-60 minutes off CPB.

## Postoperative Management

Much of the postoperative management of patients with obstructed TAPVR is dealing with the residual end-organ dysfunction caused by preoperative instability. Patients with obstructed TAPVR have elevated PVR and very reactive pulmonary vascular beds. In addition, they often require significant preoperative ventilation and may have suffered barotrauma, making ventilation more difficult. Some of these patients may require treatment of pulmonary hypertension with oxygenation, iNO, and sildenafil. Patients initially remain sedated and at times require neuromuscular blockade in the initial postoperative period until the reactive component of their elevated PVR improves.

Patients with TAPVR have relatively small left hearts due to limitations of fetal blood flow. Patients are managed with relative hypotension initially to not add undue strain to the LV. As mentioned above, patients are intolerant to volume administration, which can significantly increase LAP due to a noncompliant LV. This can reflect back onto the pulmonary venous pressure and lead to pulmonary hypertensive crises. Patients are monitored with an LA line to guide intravascular volume and hemodynamic/inotropic management. Patients are also managed with peritoneal dialysis for volume and cytokine removal. Monitoring for neurologic sequelae of preoperative instability is also important in the postoperative management.

## Complications and Long-Term Follow-Up

The development of postoperative complications in these patients is directly related to their preoperative status. Patients with obstructed TAPVR are at a higher risk of end-organ dysfunction as a consequence of their preoperative course. As mentioned above, pulmonary hypertension episodes are not unusual in patients with preoperative obstructed TAPVR. However, pulmonary vascular reactivity tends to improve over the first few days after repair.

One of the most significant and vexing long-term complications after TAPVR repair is the development of pulmonary venous stenosis. This complication occurs in approximately 10% of patients after TAPVR repair (Morales et al. 2006) and usually presents within the first 6 months after repair. Patients with heterotaxy syndrome who present with obstructed TAPVR are at a higher risk of developing postrepair pulmonary vein stenosis (Spigel et al. 2020). Even though the stenosis can occur at the site of the

### TAPVR repairs at TCH (1995-2019) (Spigel et al. 2020)

Number of patients: 336

Patients with heterotaxy: 118 (35%)

- Single ventricle patients: 106/118 (90%)
- Obstructed TAPVR: 48/118 (41%)
- TAPVR repair: 94/118 (80%)

Patients without heterotaxy: 218 (65%)

- Single ventricle: 14/218 (6%)
- Obstructed TAPVR: 87/218 (40%)
- TAPVR repair: 213/218 (98%)

Median follow-up time: 6.6 years

Mean number of pulmonary vein interventions:

- Heterotaxy, obstructed TAPVR: 2.5
- Heterotaxy, unobstructed TAPVR: 1.3
- No heterotaxy, obstructed TAPVR: 1.3
- No heterotaxy, unobstructed TAPVR: 1.3

30-day survival: 97% (95-99%)

5-year survival: 86% (83-91%)



anastomosis, some patients present with more diffuse fibrotic involvement of the upstream pulmonary veins. Initial intervention for postrepair pulmonary venous stenosis is usually surgical and entails a “sutureless” repair in which the affected pulmonary veins are opened into the pericardial well and the atrium is sutured to the pericardium around the pulmonary veins. Recurrence of pulmonary venous stenosis is a challenging problem and pulmonary venous stenting in the cardiac catheterization lab may be required.

### Isolated Pulmonary-Vein Stenosis

Patients with pulmonary vein stenosis comprise one of the most critically ill and challenging groups of patients we treat. Pulmonary vein stenosis may be broadly divided into 2 categories: primary (congenital) or secondary (e.g., post-TAPVR repair) pulmonary vein stenosis. Primary (congenital) pulmonary vein stenosis carries a worse outcome. In either category, survival for patients with progressive pulmonary vein stenosis is approximately 50% at 1 year.

#### Pathophysiology and Clinical Presentation

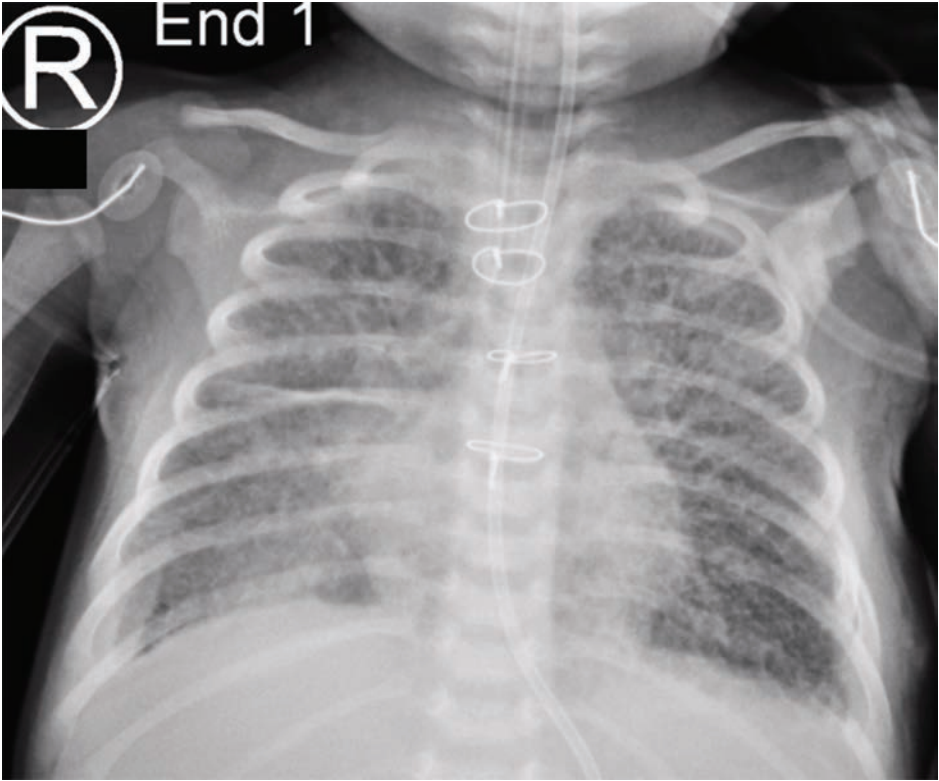
The origin of primary stenosis of the pulmonary veins relates to incomplete incorporation of the common pulmonary vein into the posterior wall of the LA. Pulmonary vein stenosis may be localized or diffuse. Diffuse stenosis of individual pulmonary veins has been seen in children with pulmonary atresia or hypoplastic left heart syndrome and TAPVR in the presence of heterotaxy.

Patients usually present with tachypnea or recurrent pneumonia. If present, right-heart failure in infants will manifest as vomiting and abdominal distention from pulmonary hypertension and hepatomegaly. Hemoptysis is less frequently observed in infants but it tends to be present in older children with individual pulmonary vein stenosis. Due to the combination of pulmonary venous desaturation and right-heart failure, patients can be cyanotic or desaturated.

On examination, there is an RV impulse with narrowing of S2, making it at times single with an accentuated P2. There might be a murmur of TR manifested as a holosystolic murmur that is high frequency (similar to MR) and localized in the midsternal border.

#### Diagnosis

- **ECG.** It is common to see RV hypertrophy with RA enlargement.
- **CXR (Figure 10-5).** May show significantly increased pulmonary vascular markings or pulmonary edema, with or without cardiomegaly. Pulmonary vascular markings may be asymmetric if there is regional/segmental obstruction.
- **Echocardiogram (Figure 10-6).** The echocardiogram should focus on detailing the individual distal portions of the pulmonary veins using high parasternal views or subcostal windows. Abnormal pulmonary venous Doppler will manifest as continuous, high-velocity turbulent flow with loss of phasic variation. The RV should be examined as it may be dilated and dysfunctional as a consequence of pulmonary hypertension. Indirect measures of pulmonary hypertension (e.g., septal configuration, TR jet) are important.
- **CTA (Figure 10-7).** CTA plays a significant role in diagnosis and management



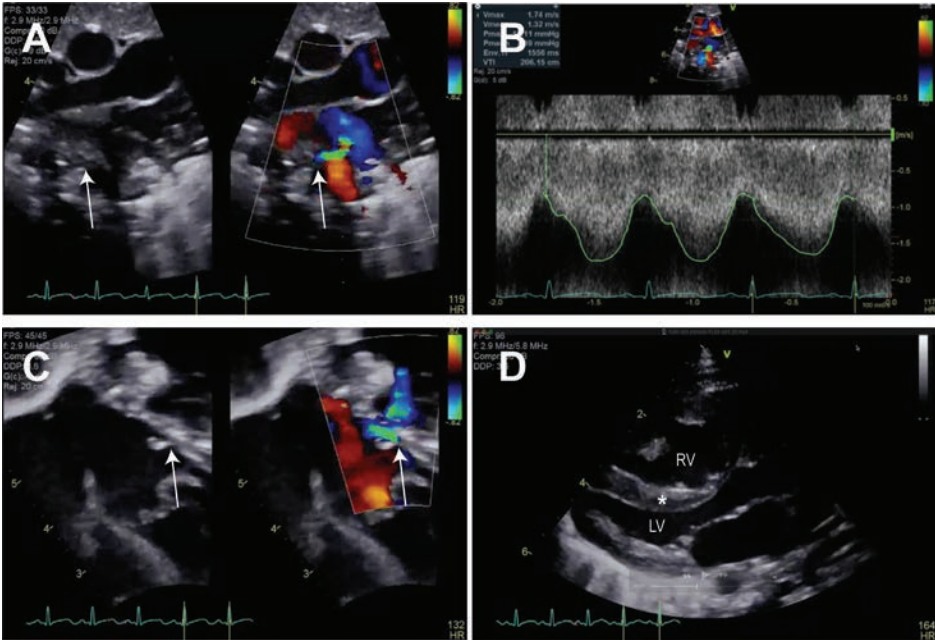
**Figure 10-5.** CXR of an infant with pulmonary vein stenosis (diffuse) after surgical correction for TAPVR. There is bilateral pulmonary edema with fluid in the fissure and bilateral pleural effusions (more right than left). Of note, there is no obvious cardiomegaly.

of patients with pulmonary vein stenosis. It is useful in assessing the degree and length of stenoses, the differential involvement of each pulmonary vein, and the morphology of the upstream segments. CTA not only helps to provide a roadmap for intervention but is also helpful for patient follow-up.

- **Cardiac catheterization.** Diagnostic and therapeutic cardiac catheterization play a critical role in the management of these patients (see below).

### Management

The management of patients with pulmonary vein stenosis requires coordination of care within the context of a multidisciplinary team. At TCH, a dedicated Pulmonary Vein Stenosis team consists of cardiothoracic surgeons, interventional cardiologists, noninvasive cardiologists, pulmonary hypertension specialists, intensive care specialists, anesthesiologists, nurses, and other specialists. Cases are reviewed and therapeutic plans, which integrate catheter-based and surgical interventions, are orchestrated with the expectation that patients will receive multiple interventions, aggressive surveillance, and medical management of pulmonary hypertension and remodeling of pulmonary arteriopathy.



**Figure 10-6.** Echocardiograms of patients with pulmonary vein stenosis. A) Side-by-side supra-sternal notch color-compare views of a stenotic left upper pulmonary vein (arrows). B) Pulmonary venous Doppler of the left upper pulmonary vein demonstrating turbulent flow with a continuous and abnormal increased velocity. C) Side-by-side 4-chamber color-compare views of a stenotic left lower pulmonary vein (arrows). D) Dilated and dysfunctional RV with a shifted interventricular septum (\*) due to pulmonary hypertension from pulmonary vein stenosis. Images courtesy of Dr. Josh Kailin, [www.pedecho.org](http://www.pedecho.org).

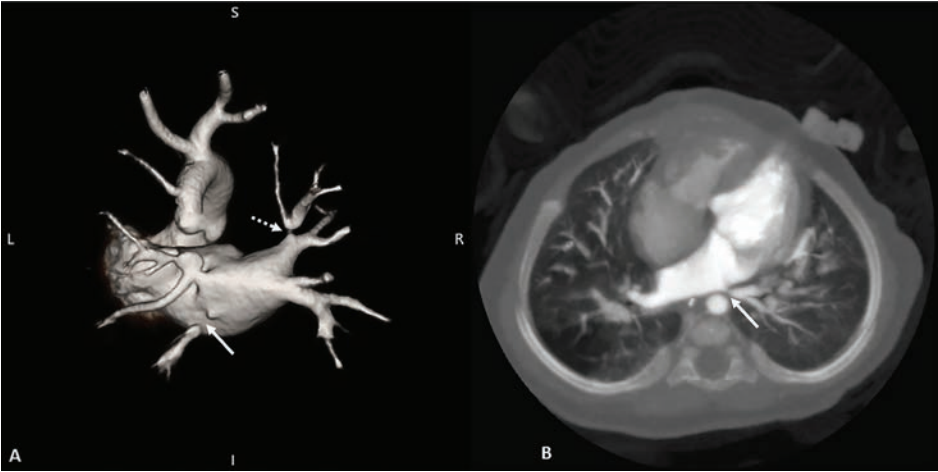
### Medical Management

Although the management of pulmonary vein stenosis is either catheter-based or surgical, diuretics can be used to mitigate the effects of pulmonary congestion on respiratory mechanics. However, preload changes should be carefully managed since RV dilation and restricted pulmonary blood flow will also impact LV preload and potentially reduce systemic output.

When right-heart failure is present, inotropic support with catecholamines may be required. Several pharmacological strategies have been used to reduce proliferation of the intimal layers of the pulmonary veins. The addition of sirolimus or everolimus to the treatment regimen of these challenging patients is still under investigation.

### Catheter-Based Intervention

Indications for intervention include severe anatomic pulmonary vein stenosis (especially multivessel involvement) with significant hemodynamic compromise due to pulmonary hypertension, cyanosis in the presence of a shunt, right-heart failure, respiratory symptoms (e.g., tachypnea with need for respiratory support), recurrent pneumonias, hemoptysis, and failure to thrive. For high-risk patients, planning consists of appropriate



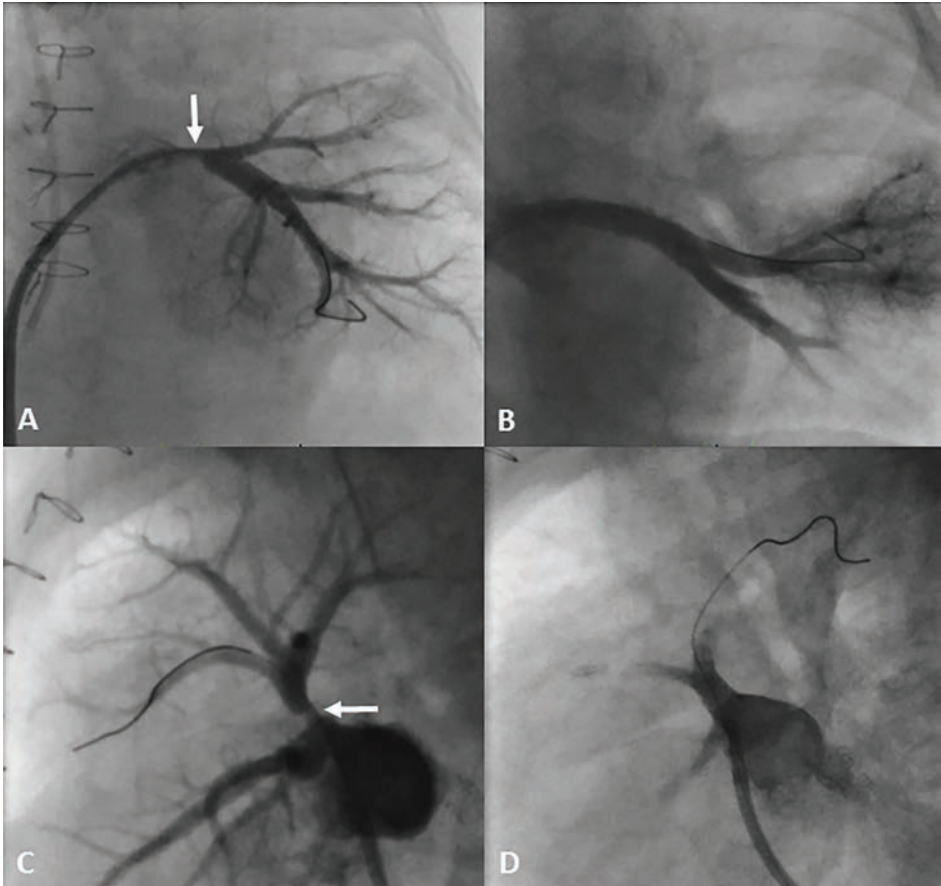
**Figure 10-7.** CT scan in a 4-month-old, ex-premature infant with a chromosomal abnormality, postsurgical VSD closure and newly discovered severe pulmonary vein stenosis with pulmonary hypertension. A) 3D imaging shows severe left lower lobe pulmonary vein stenosis (solid arrow) and severe right upper pulmonary vein stenosis (dotted arrow). The left upper pulmonary vein and its subsegments can also be seen to be stenotic. B) Axial imaging demonstrates severe left lower pulmonary vein stenosis (arrow).

surgical backup and the potential use of ECMO or other forms of mechanical support if a patient's condition deteriorates in the cardiac catheterization laboratory. In some instances, the procedure may have to be performed with the patient already on ECMO support.

Cardiac catheterization is performed with general anesthesia and biplane fluoroscopy. Arterial access is obtained for pressure monitoring. Femoral venous access is the preferred route for intervention.

In critically ill children considered to be at high risk for an impending cardiac arrest, the first portion of the procedure should be the creation of an ASD to improve hemodynamics. The ASD is created with a transeptal needle in standard fashion or by using radiofrequency energy/electrocautery. Transeptal puncture should be carried out using biplane fluoroscopy. However, echocardiography (transthoracic or transesophageal) may be necessary for difficult transeptal punctures, such as those in patients with prior surgical patches or challenging anatomy because of leftward bowing of the atrial septum from severe pulmonary hypertension.

For standard-risk patients, complete right-sided hemodynamics are performed (and left-sided, if indicated) and all PA wedge positions are entered. After hemodynamics are measured, PA wedge angiography is performed to delineate the pulmonary veins on levophase (both upstream and ostial pulmonary veins) and provide anatomic diagnostic information and a roadmap for the intended interventions on stenotic/occluded pulmonary veins. In addition to delineating stenotic pulmonary veins, PA wedge angiography may identify a small channel of a pulmonary vein that was felt to be occluded by noninvasive imaging. If indeed the pulmonary vein is occluded, PA



**Figure 10-8.** Angiography in the same patient described in Figure 10-7 shows severe stenosis of the left lower pulmonary vein at 4 months of age in a left anterior oblique/cranial projection (arrow) (A). A follow-up catheterization at 11 months of age shows an unobstructed left lower pulmonary vein after stenting with a drug-eluting stent and dilation to 5 mm (B). The right upper pulmonary vein is also severely stenotic at 4 months of age (C) on the lateral projection (arrow). An unobstructed right upper pulmonary vein is seen at 7 months of age after implantation of a 4 mm drug-eluting stent (D).

wedge angiography still provides an anatomic target for recanalization, if feasible. In some instances, a more focused diagnostic cardiac catheterization may be performed based on findings from a CTA (Figure 10-7).

After transseptal puncture, the patient is heparinized to maintain an ACT  $\geq 250$  seconds for the duration of the procedure. Once the pulmonary veins are entered, balloon angioplasty is performed (standard or high-pressure angioplasty). A cutting balloon may be used, if needed, to treat resistant lesions. In general, an appropriately sized stent is implanted. It is preferable to insert a large diameter stent (bare metal stent), if possible. If the vessel is small, which is frequently the case in pediatric pulmonary vein stenosis,

a coronary stent is implanted (usually 4-5 mm, but smaller in some instances). These stents can be redilated later and intentionally fractured to accommodate implantation of a larger-diameter stent. “Kissing” stents or intentional vessel jailing with dilation/stenting through stent side cells may be needed to treat pulmonary veins in close proximity. While some centers perform balloon angioplasty (standard, high-pressure, and cutting-balloon angioplasty) for pulmonary vein stenosis, we prefer to implant stents whenever possible. This may be associated with a higher likelihood of maintaining vessel patency and achieving the largest lumen diameter possible in follow-up. We prefer to implant drug-eluting stents (Figure 10-8) in small-diameter pulmonary veins. Over a 24-year period at TCH, drug-eluting stents (implanted in 105 lesions) have been found to result in a significantly less rate of lumen loss than bare metal stents (implanted in 58 lesions) (Khan et al. 2019).

An ASD is usually created with a large balloon to facilitate subsequent interventions and decrease procedure times (in addition to providing a pop-off mechanism). Meticulous attention to central venous access is important. Due to repeated procedures, transhepatic venous access and recanalization of femoral/internal jugular veins may be needed for future interventions.

Operators should have the ability to treat infrequent, but significant complications (e.g., cardiac perforation, pulmonary vein tears, thrombotic events, stent dislodgement) with appropriate surgical backup.

### Surgical Intervention

Surgical therapy for established pulmonary vein stenosis should be performed within the context of a multidisciplinary program to manage a highly lethal condition, which frequently recurs despite adequate decompression in the OR. The concept of a “one-and-done” operation that cures the problem should not be expected. With aggressive surveillance and a low threshold for repetitive reintervention, the state of the art suggests that the prognosis can be improved.

The anatomic profile of disease in an individual patient has a strong influence on the likelihood of a durable surgical result. The group with the most favorable prognosis includes patients with stenosis at the site of anatomic repair after correction of TAPVR, where the stenosis is largely related to stricture at the site of the anastomosis between the atrium and the pulmonary veins. This complication occurs in approximately 10-15% of TAPVR repairs. The most likely etiology is a failure to create a sufficiently large anastomosis at the TAPVR repair or a pursestring effect due to inappropriate tension during construction of the anastomosis. Both problems may be avoided using a sutureless repair technique when repairing TAPVR. This type of pulmonary vein stenosis is termed post-repair pulmonary vein stenosis (PR-PVS).

When evaluating a patient with PR-PVS, a key feature to evaluate is the size of the upstream pulmonary veins. Typically, patients with PR-PVS with early detection will have dilated upstream pulmonary veins and are amenable to surgical decompression with a sutureless approach. When the upstream pulmonary veins are small, however, the likelihood of recurrent stenosis within the smaller pulmonary veins is high and aggressive surveillance is indicated. The sutureless approach can be used to reestablish

continuity with atretic pulmonary veins, but the size of the upstream pulmonary veins is also an important predictor of postoperative restenosis.

Another important subset of patients are patients with primary (congenital) pulmonary vein stenosis. In these patients, the size of the upstream pulmonary veins is also an important predictor of need for future reintervention and ultimate survival.

Patients who are brought to the OR after stent-based therapy can undergo resection of the stents as part of a sutureless repair or intraoperative stent dilation in those with either small upstream pulmonary veins or stents that extend significantly into the upstream segments.

### **Long-Term Follow-Up**

Patients are followed after catheterization or surgical therapy by the Pulmonary Vein Stenosis team. Surgical patients undergo a surveillance CT scan within a month of repair. Catheterization-based interventions are typically followed with a repeat catheterization after 3 months. In both tracks, there is a low threshold for subsequent reinterventions.

Pulmonary vasodilators and pulmonary vascular remodeling therapy may be considered for patients in the interim period between surgical or catheterization-based decompression and the expected recurrence of pulmonary vein stenosis.

For patients with relentless progression and/or poor upstream pulmonary vein development, adjunctive medical therapy with rapamycin (found to be effective in a very small clinical experience) or losartan (efficacy demonstrated in an animal model) may be considered. Efficacy of these and other medications in large clinical cohorts of patients with pulmonary vein stenosis is lacking.

### **Suggested Readings**

Khan A, Qureshi AM, Justino H. Comparison of drug eluting stents versus bare metal stents for pulmonary vein stenosis in childhood. *Catheter Cardiovasc Interv* 2019;94:233-242.

Morales DLS, Braud BE, Booth JH, et al. Heterotaxy patients with total anomalous pulmonary venous return: improving surgical results. *Ann Thorac Surg* 2006;82:1621-1628.

Spigel ZA, Edmunds EE, Binsalamah ZM, et al. Heterotaxy syndrome and obstructed pulmonary veins. Accepted for presentation at AATS 2020. Manuscript in preparation.