

18

Right-Ventricular Outflow Tract Obstruction

Antonio G. Cabrera, Athar M. Qureshi, Charles D. Fraser Jr.

Both pulmonary atresia with intact ventricular septum (PA-IVS) and isolated pulmonary stenosis (PS) represent variations within the spectrum of RVOT obstruction. These lesions vary significantly in terms of clinical and morphological characteristics, and thus management strategy. As such, PA-IVS and isolated PS will be discussed in separate sections of this chapter.

PA-IVS

PA-IVS is a lesion characterized by an atretic pulmonary valve, a muscularized and variably hypoplastic RV with no ventricular septal communication, and a variably hypoplastic and tethered tricuspid valve (TV). An ASD is obligatory for survival. This lesion is associated with coronary abnormalities which may make catheter and surgical procedures significantly risky.

PA-IVS is a highly heterogeneous lesion. As such, the decision-making process and management of these patients is quite complex and has been subject of much controversy. Many patients are able to achieve a biventricular circulation (through different stages) while others will benefit from single-ventricle palliation. In certain circumstances, a “1.5-V repair” can be achieved in patients that would otherwise not be candidates for a biventricular circulation.

Pathophysiology and Clinical Presentation

PA-IVS presents with variable degrees of TV and RV hypoplasia. TV annular dimension is moderately associated with the degree of RV hypoplasia. The TV leaflets may vary from normal to severely thickened with the subvalvar apparatus matted with indistinct chordae and fibrotic, and at times infarcted, papillary muscles. In a small number of cases (5-10%) there is significant TV dysplasia with apical displacement of

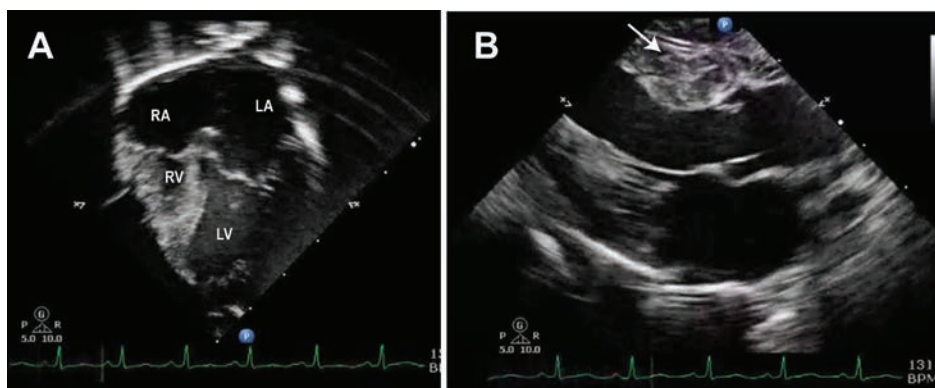


Figure 18-1. Echocardiography in PA-IVS. A) 4-chamber view showing a small, muscularized, and hypertrophied RV with a small TV. B) Parasternal long-axis view showing a normal-sized LV with a small and muscularized RV (arrow). Images courtesy of Dr. Josh Kailin, www.pedecho.org.

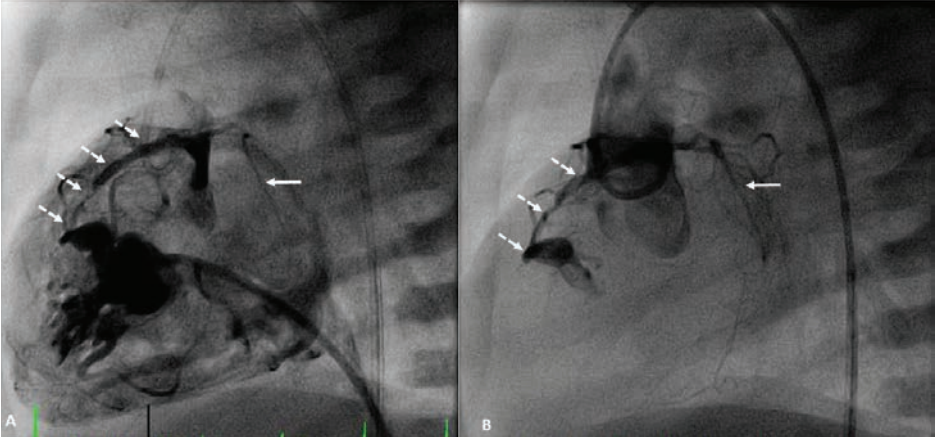


Figure 18-2. A) Right ventricular angiogram in a newborn with PA-IVS. Interruption of the mid left anterior descending coronary artery (dotted arrows) with a fistulous connection to the RV is seen. A small circumflex coronary artery (solid arrow) is also seen. B) Direct angiography in the left main coronary artery confirms interruption of the left anterior descending coronary artery (dotted arrows) and a small circumflex coronary artery (solid arrow).

the valve (Ebstein-like), which can lead to severe TR, massive RA dilation, a dilated RV, and concomitant LV dysfunction. Mortality in these patients may exceed 50-55%; heart transplantation should be considered.

In PA-IVS, since there is some inflow through the TV into the RV but no outflow, the RV becomes significantly pressurized. This phenomenon can lead to abnormal development of the coronary circulation and the presence of coronary sinusoids or fistulas between the RV and the coronary arteries. The flow dynamics of these fistulas can lead to the development of proximal coronary stenoses, making the coronary perfusion dependent on flow from the hypertensive RV through the fistulas and into the coronary arteries (RV-dependent coronary circulation, RVDCC). RVDCC is specifically defined as myocardial perfusion that is at least partially dependent on RV fistulas due to proximal epicardial coronary obstruction of two or more major coronary branches or ostial atresia. Intimal fibromuscular hyperplasia often occurs in the coronary arteries associated with fistulas and may be responsible for myocardial ischemia and infarction. The presence of RVDCC has significant implications not only for the management but the long-term prognosis of this lesion.

The factors that are mainly associated with long-term outcomes in PA-IVS are:

- Presence or absence of RVDCC
- Size and morphology of the RV
- Size and function of the TV

Patients with PA-IVS are dependent on a PDA and an ASD for early survival. If a patient has not been diagnosed prenatally and the duct is allowed to close, the patient will present with severe hypoxemia and acidosis. Once PGE has been started for ductal patency, the patient will have some cyanosis (due to the obligatory right-to-left shunt at

18. RIGHT-VENTRICULAR OUTFLOW TRACT OBSTRUCTION

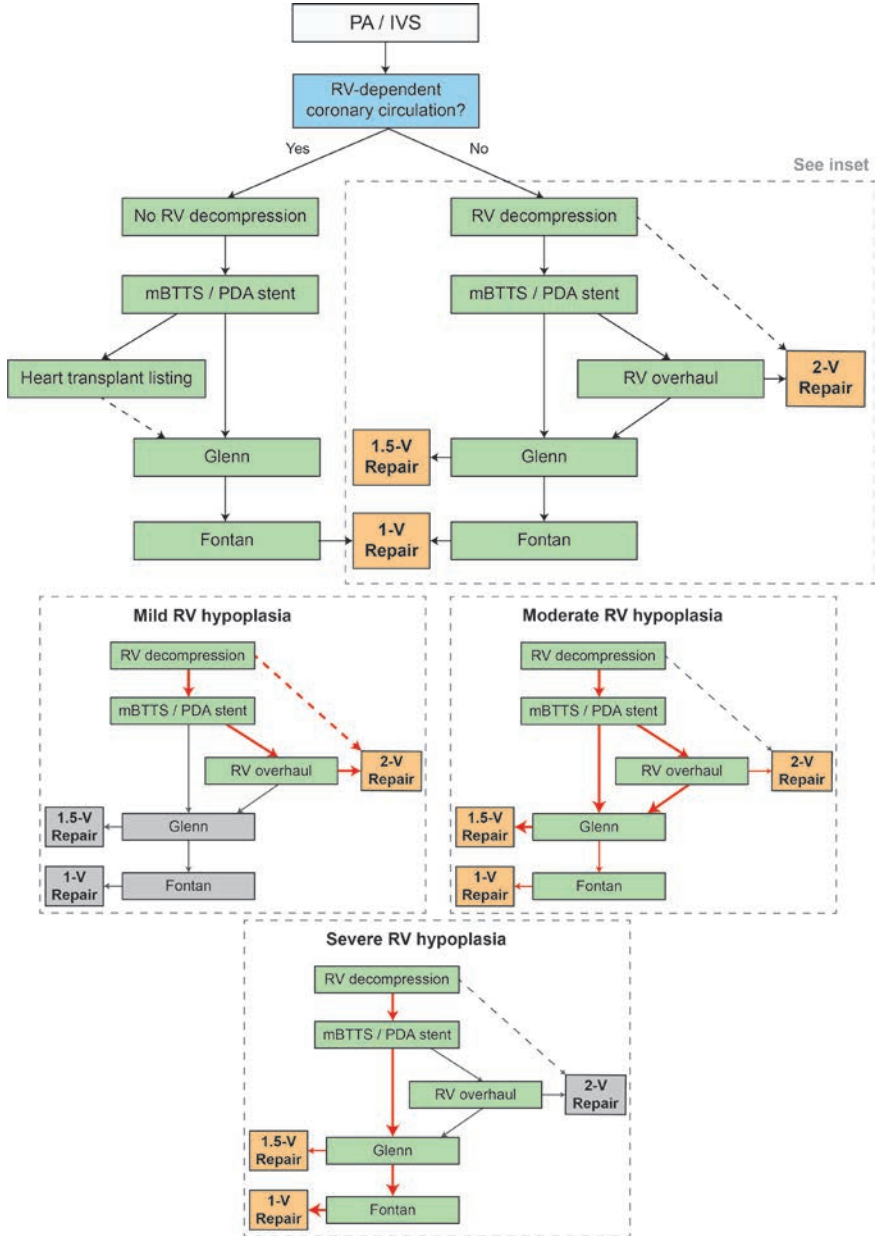


Figure 18-3. Management algorithm for patients with PA-IVS. Management depends on whether the patient has RVDCC and the degree of RV and TV hypoplasia. The treatment is individualized for each particular patient. Red solid arrows indicate the most common direction for each scenario, followed by less prominent red arrows, and then dashed red arrows. Grey arrows indicate very unlikely directions for that particular scenario.

the atrial level). As PVR decreases, patients will become more tachypneic and exhibit signs of overcirculation. On cardiac exam, there is usually a mid-precordial impulse. The second heart sound is single and sometimes there could be an S_3 gallop. When the TV is significantly hypoplastic and stenotic, a diastolic rumble may be heard. There is a harsh high-frequency holosystolic murmur at the right midsternal border when there is significant TV insufficiency. This could be misinterpreted as MV regurgitation or a VSD.

There is a wide spectrum on the clinical presentation of children with RVDCC. Some children may have overt ischemia with shock and poor ventricular function, others may have ventricular tachycardia/fibrillation, but the majority have subtle episodes of angina/inconsolability with transient ST-segment changes.

Diagnosis

- **CXR.** Ranges from normal-sized cardiac silhouette to cardiomegaly with pulmonary edema and hyperinflation.
- **ECG.** RA enlargement. It may vary between nonspecific ST-T wave changes and significant ST-depression/elevation if there is significant coronary ischemia.
- **Echocardiogram (Figure 18-1).** Important features to assess include the type of pulmonary atresia (membranous/plate like or muscular obliteration), the characteristics of the TV (size, mobility, function), and the size and morphology of the RV. The size and character of the tricuspid valve mechanism are important, although in most cases, the actual size of the tricuspid orifice may be very difficult to determine in the setting of the very hypertensive RV. Much has been said of determining whether the RV is “tripartite” (inlet, trabecular, and infundibular portions), but this is probably overstated. The most important determinant in predicting an effective RV in the long term is whether or not there is a well-formed RV infundibulum in the newborn period. It is important to assess the function of both ventricles and the presence of any segmental wall motion abnormalities. Coronary artery anatomy should also be assessed. To-and-fro flow through the coronary arteries demonstrated by color Doppler suggests coronary-RV fistulas although it does not establish whether there is RVDCC or not. A subcostal/coronal view will demonstrate the presence of an atrial communication with obligatory right-to-left shunting. The atrial septum usually appears elongated and aneurysmatic.
- **Cardiac CTA.** Rarely necessary unless there are particular questions that arise from the echocardiogram. CTA does not identify the presence or absence of RVDCC.
- **Cardiac catheterization.** It is important to perform a cardiac catheterization in all patients with PA-IVS shortly after birth in order to define the presence or absence of RVDCC, evaluate the RV chamber size, and potentially establish a source of pulmonary blood flow. Cardiac catheterization is not only diagnostic but plays a significant role in the management strategy of these patients. Cardiac catheterization with an RV injection is critical to determine the presence and character of RV to coronary communications. Many patients will have RV to coronary sinusoids and, as such, when the RV is injected, one may observe extensive filling of the epicardial coronary arteries (Figure 18-2A). It is also critical to perform antegrade coronary angiography (ideally with direct ostial injection) to critically assess the nature of the RV communication, to ascertain whether or not there is ostial atresia, to determine

whether there are interrupted segments of the epicardial coronary arteries (and whether the interruptions/stenosis are located in the proximal or distal coronary arteries), and to answer the critical question of whether or not there are RVDCC (Figure 18-2B). The presence of extensive RV-to-coronary communications alone (in the absence of ostial atresia and/or interrupted segments of the epicardial coronaries) *does not* equate with the diagnosis of RVDCC. Furthermore, it must be noted that if one leaves the RV without decompression over the long term, these sinusoidal communications, which are by definition hypertensive relative to normal coronary artery pressure, will often lead to distortion of otherwise normal coronary arteries and may ultimately lead to an acquired RVDCC.

Indications / Timing for Intervention

All patients should undergo a diagnostic cardiac catheterization after stabilization. In addition, all patients will require a stable source of pulmonary blood flow in the newborn period. This can be accomplished by RVOT perforation in the cath lab, placement of a PDA stent, a modified Blalock-Taussig-Thomas shunt (mBTTS), creation of a transannular RVOT patch, or a combination of these techniques (see below).

Management

The management of patients with PA-IVS is complex due to the heterogeneity of the disease. Figure 18-3 shows the algorithm used for decision-making in this disease.

RVDCC

In babies with true RVDCC, the RV cannot be decompressed as this will lead to critical ischemia. Unfortunately, many of these babies are at risk for progressive stenoses of not only the connections, but the epicardial coronaries themselves. As such, critical ischemia and sudden cardiac death are important ongoing risks. In these babies, the next step in management is to establish a stable source of pulmonary blood flow. This is typically achieved by either placing a PDA stent or creating an mBTTS (see Chapter 38). Placement of a PDA stent often requires discontinuation of the PGE to allow for ductal constriction prior to placement of the stent. Whether one places a PDA stent or surgically creates a shunt, it is very important to avoid “over-shunting” the patients, which may exacerbate the propensity for ischemia. An advantage that ductal stenting may offer is the option of performing a balloon atrial septostomy which may be an important adjunct to management in babies with marginal atrial-level communications. It is critically important to note that in the setting of RVDCC, an attempt to decompress the RV may be a fatal proposition.

In those children with true RVDCC, the next step in the management algorithm is to consider cardiac transplantation. It must be noted (and thereby shared with the patient’s parents), that the prospect for neonatal transplantation in terms of suitable donor organ availability is a statistically improbable consideration. This does *not* mean that newborns cannot be successfully transplanted; it does mean that listing does not equate with successful transplantation. As such, we have recommended urgent listing of appropriate candidates at the earliest appropriate time. While candidates are waiting, we have for the most part required the patients to stay in the hospital in an advanced care unit. While unheralded decompensations do occur (acute shunt occlusion, dysrhythmia), in

TCH experience with PA-IVS (1995-2017)

Number of patients: 89

Median age at first intervention: 4 days (0-64 days)

Median weight at first intervention: 3 kg (1-5 kg)

Median TV Z-score at presentation: -2.5 (-5.3 – -2.2)

Presence of RV-coronary fistulas: 42 (55%)

RVDCC diagnosed at presentation: 20 (22%)

Final strategy for patients (median follow-up 8 years):

- 1-V Repair: 37%
- 1.5-V Repair: 17%
- 2-V Repair: 24%
- Transplant: 3%
- Death: 20%

10-year survival: 82%

most patients there are subtle warning signs that may signal impending decompensation (declining NIRS, declining SaO₂, poor weight gain, new ECG changes, etc.) that are potentially reversible if rapidly addressed. As the children get older, they may become eligible for second- (bidirectional Glenn shunt) or even third- (Fontan) stage palliation. We have had a number of children with RVDCC who have been successfully progressed through all stages

of palliation. As an important intraoperative management point, children with RVDCC typically cannot tolerate complete RA (and thereby RV) decompression on CPB. As such, perfusion techniques designed to maintain RV filling (partial CPB or SVC-to-RA shunt with an oxygenator) are important along with very careful intraoperative assessment for ischemia, primarily as demonstrated by important ECG changes. It is also important to recognize that even after palliation with Glenn and Fontan circulations, these patients are at risk for coronary events and even sudden death, due to progressive distortion (or progression of stenoses/occlusions) of the coronary arteries. As such, routine surveillance and testing as an outpatient over time with a particular emphasis on the coronary arteries is mandatory.

RV Decompression

In patients without evidence of RVDCC, we have moved forward with efforts to open the pulmonary valve to promote prograde RV ejection, even in the setting of RV sinusoids. The first step is typically to perform a radiofrequency perforation (or perforation using chronic total occlusion guidewires) of the atretic pulmonary valve in the cardiac catheterization lab followed by serial balloon dilation. In some cases, this may be all that is needed and the patients may eventually be weaned from PGE with adequate prograde pulmonary blood flow provided by the RV. In patients in whom the RV cannot generate adequate blood flow, either a PDA stent or mBTTS is constructed. It has generally been our approach to allow the PDA to constrict and accept slightly low saturations in some patients in whom RV remodeling (with resultant improvement in saturations) can take months. If it is evident that a patient will require another source of pulmonary blood flow, this is usually performed 1-2 weeks later after a trial off PGE. However, in some cases, it is obvious at the initial cardiac catheterization that another source of pulmonary blood flow will be needed. In those instances, a PDA stent may be placed at the time of the initial procedure, or an mBTTS created soon afterward in the OR. If percutaneous pulmonary valvotomy is not successful, our management algorithm is

to then perform an open pulmonary valvotomy with at most a minimal transannular incision. While we may then perform an RVOT resection, we do not favor a transmural infundibulotomy (RVOT incision) as some centers do. At the primary operation, we will usually perform an atrial septectomy and inspect the tricuspid valve, but do not attempt tricuspid valvuloplasty in most cases at this time.

RV “Overhaul”

In the presence of a well-formed infundibulum, even very diminutive RVs are amenable to promotion and may grow significantly. In shunted patients, the RV and PA morphology is carefully followed by echocardiography and catheterization or CTA. If the RV is muscle bound and not progressing in size, an intermediate operation is often necessary; typically around 3-6 months of life. This operation was initially described as an RV “overhaul” and consists of tricuspid valvuloplasty, RV endocardial and trabecular resection, and RVOT resection. This operation is done on CPB and the systemic-to-PA shunt is temporarily occluded. The patient is then left with a partially shunt-dependent circulation and RV progress is carefully monitored.

1.5-Ventricle Repair

In some patients, the RV never progresses to the degree that it is capable of handling the entire cardiac output. Our experience suggests that often, the rate-limiting feature is the TV dimension. It is unwise to attempt to “force” more flow through the TV than it can accommodate. If the true tricuspid orifice (not the annular dimension, but the actual effective orifice) is 50% or less than what it should be for the patient’s BSA, one should strongly consider a 1.5-ventricle repair. This operation includes RV endocardial/trabecular resection, partial or complete ASD closure, takedown of the systemic-to-PA shunt, and creation of a bidirectional Glenn anastomosis. This can be a very effective circulation, which provides reasonable pulmonary blood flow (SaO₂ usually in the mid-to-high 90s even if there is a small ASD left), and low hepatic venous pressures. Of note, we have not typically placed a pulmonary valve in most cases.

Biventricular Repair

The decision to proceed with a septated circulation may be challenging in cases of a truncated RV or marginal TV. If the right-sided structures are believed to be adequate, the ASD is completely closed and CPB weaned while measuring RA pressures. If the measured CVP is <10-12 mmHg with good cardiac output, the chest is closed. If it is greater than this, CPB is reinstated and a small (4-5 mm) fenestration is created in the atrial septum. Later in life, test occlusion and even permanent closure of the ASD can be performed in the cardiac catheterization laboratory.

Fontan Circulation

Other than patients with RVDCC, there are some patients with a previously decompressed RV in whom a Fontan operation may ultimately be necessary. This occurs in cases of a very small TV, an RV that fails to progress, and inadequate pulmonary blood flow to maintain adequate oxygen saturations. In such cases, the additional flow provided by a total cavopulmonary connection Fontan is warranted. In the setting of PI, it may be

necessary to place a patch over the TV (carefully avoiding the AV node) to prevent a circular shunt after the Fontan connection.

Isolated Pulmonary Valvar and Supravalvar Stenosis

Isolated PS is the most common valvar abnormality that requires intervention in the neonatal period. Valvar PS has been associated with Allagille and DiGeorge syndromes, while supravalvular PS has been associated with Williams and Noonan syndromes.

Pathophysiology and Clinical Presentation

Overall, there are 3 distinct types of valvar PS: critical neonatal PS, typical PS, and dysplastic PS. *Critical PS* presents in the newborn period as cyanosis, and is defined as PS that is ductal-dependent for pulmonary blood flow. Cyanosis is the result of right-to-left shunting at the atrial level. These patients will present with decreased pulmonary blood flow and may have variable degrees of diastolic dysfunction from severe RV hypertrophy. Patients who have *typical PS* have leaflets that are fused and the main PA demonstrates poststenotic dilation. Patients with *dysplastic pulmonary valves* have thicker leaflets than those with typical PS, and little or no leaflet fusion. The main PA segment is small and frequently, the branch PAs are also small. This form of PS is seen in patients with Noonan syndrome, Williams syndrome, and other genetic conditions.

On clinical exam, there will be a prominent RV impulse with or without a thrill, a systolic ejection click, and a harsh systolic ejection murmur. The murmur frequency will vary depending on the degree of valvar stenosis and the PVR. At birth, the murmur will likely be low frequency, and as PVR decreases, the frequency of the murmur will increase. In cases of supravalvular PS, the exam will be similar, with the exception of the systolic ejection click, which will be absent. Depending on the degree of obstruction, there can be hepatomegaly secondary to a combination of high RV afterload and diastolic dysfunction.

Diagnosis

- **ECG.** Significant RVH and possible RA enlargement.
- **CXR.** Nonspecific. In severe cases, it will show decreased pulmonary blood flow (oligemic lungs).
- **Echocardiogram (Figure 18-4).** There is typically fusion of the valvar commissures that prevents complete opening or “doming” during systole, leaving a small orifice for ventricular stroke volume ejection. There are variable degrees of RV hypertrophy and/or hypoplasia. It is important to perform a careful assessment of the size and z-score of the branch PAs, the presence or absence of collaterals, and the presence of a PDA.

Indications / Timing for Intervention

Patients with severe PS (and some with moderate PS) require intervention to relieve the obstruction. Indications for intervention include patients with critical PS and patients with a peak gradient >40 mmHg by TTE or a peak-to-peak systolic gradient >40 mmHg in the cardiac catheterization laboratory. Lesser gradients may be an indication to intervene in the presence of RV dysfunction or poor cardiac output.

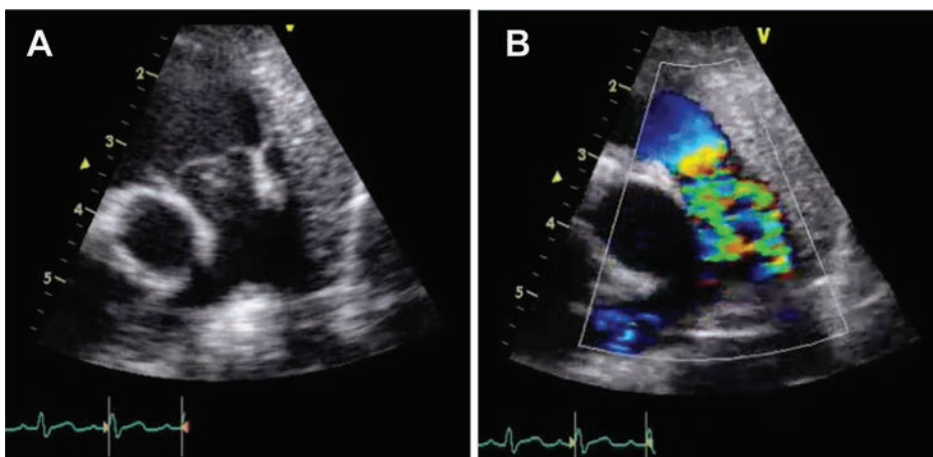


Figure 18-4. Parasternal short-axis 2D (A) and color-Doppler (B) echocardiographic views showing a doming pulmonary valve with PS.

Catheter-Based Intervention

Percutaneous balloon valvuloplasty is the treatment of choice for isolated valvar PS. Neonates with critical PS may require intervention to be able to stop PGE administration and improve oxygen saturations. In our experience, roughly 20-30% of patients with critical PS will need reintervention after balloon valvuloplasty (either an alternate source of pulmonary blood supply, or repeat balloon pulmonary valvuloplasty or RVOT surgery) in the first 4 months of life. A small minority of patients will not exhibit adequate RV growth and may require a Glenn or an RV overhaul operation later on.

The fused valves in patients with typical PS are very amenable to balloon valvuloplasty with nearly all procedures being successful, as the areas of fusion can be easily torn with a balloon. These patients typically present a large main PA due to poststenotic dilation (Figure 18-5A).

For patients with dysplastic pulmonary valves (Figure 18-5B), the success rate for balloon valvuloplasty is less (approximately 50%). These patients may require more than one catheter procedure, larger balloons, and inflations at higher pressure to relieve the obstruction. Those who do not respond with these catheter-based interventions require surgical relief of the obstruction.

Catheterization Procedure

During the cardiac catheterization procedure, careful pullback measurements are made to discern the exact level of obstruction and to ensure there are no other obstructions present in the branch PAs or RVOT. Angiography with delineation of the anatomy and pulmonary valve annulus is made. Generally, a balloon is initially chosen that is 1.2-1.3 times the pulmonary valve annulus. With a properly selected balloon (exception being dysplastic pulmonary valves), a waist is seen at the narrow segment within the leaflets, that yields with inflation. After inflation, pressure measurements are made to determine whether further intervention is warranted. The goal of the procedure is

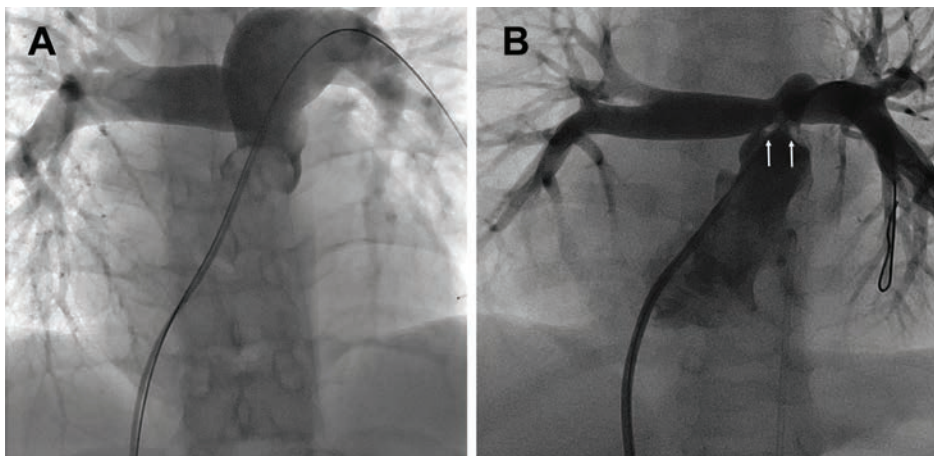


Figure 18-5. A) Main PA angiogram in a 15-year-old with typical valvar PS. Note the dilated main PA segment. B) Main PA angiogram in an infant with a dysplastic pulmonary valve. The small main PA can be seen, as can the very thick dysplastic pulmonary valve leaflets (arrows).

not to completely abolish the gradient, as more than desired PI (requiring treatment later in life) may develop. Mild-to-moderate PS is tolerated well for many years, if not for one's entire life.

Postcatheterization Management

PGE may be maintained in the immediate post-balloon-valvuloplasty period. Cyanosis and desaturations may still be present secondary to significant RV diastolic dysfunction and right-to-left shunting through the PFO. Diastolic dysfunction, especially when there is RV hypoplasia, tends to improve after several days and more than one attempt at weaning PGE.

Postcatheterization Follow-Up

Repeat balloon valvuloplasty may be needed in some patients, particularly younger patients or patients with dysplastic pulmonary valves. PI is common in follow-up and a small subset of patients may require pulmonary valve replacement later in life. For patients with critical PS, PGE can be stopped after the procedure, and patients should be monitored for the need for another source of pulmonary blood flow or reintervention on the RVOT. In these patients, remodeling of the RV may take months, and some degree of cyanosis is expected during this timeframe.

Surgical Intervention

Surgical intervention is reserved for patients who fail percutaneous balloon valvuloplasty or those in which the annulus of the pulmonary valve is significantly hypoplastic. Patients with isolated valvar PS and a reasonable annulus may benefit from surgical valvotomy (carefully incising into each of the commissures to enlarge the effective orifice of the valve) and/or debridement of thickened and dysplastic pulmonary valve leaflets. Patients with a significantly hypoplastic pulmonary annulus may require a

transannular pulmonary patch to enlarge the RVOT. In cases of supraavalvar PS, patients may undergo patch enlargement of the supraavalvar area with either a single patch or a pantaloony patch (analogous to supraavalvar aortic stenosis repair, see Chapter 24). Patients with critical PS may require placement of an aortopulmonary shunt to supplement pulmonary blood flow (see Chapter 38).

In patients with long-standing PS, secondary muscular infundibular obstruction develops over time and may result in residual obstruction after open valvotomy. Our practice has been to carry out a careful infundibular resection through the pulmonary valve if there appears to be the potential for residual muscular obstruction.