Profiles in Congenital Heart Disease

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In 1938 the first successful surgical management of a congenital heart lesion, a patent ductus arteriosus (PDA), was reported by Gross, and this was followed several years later by aortic coarctation surgical repair. During the subsequent two decades, intracardiac repair of atrial septal defect (ASD), ventricular septal defect (VSD), valvar pulmonary and aortic stenoses, and more complex lesions such as tetralogy of Fallot (TOF) became commonplace. Cardiac catheterization techniques developed in parallel with these advances, and, indeed, preoperative catheterizations preceded essentially all operations and helped considerably in improving surgical outcomes. In 1966, Rashkind and Miller introduced the first widely used catheter-based interventional technique, namely balloon atrial septostomy (BAS) for transposition. In 1982 Kan et al. described static balloon pulmonary valvotomy for pulmonary stenosis, which heralded the current surge of interventional procedures in the management of congenital heart diseases. From a pediatric perspective, now more than a half century after the pioneering report of Gross and together with the remarkable advances in ultrasound technology and surgery, most of the complex lesions, particularly in neonates and infants, are repaired surgically without catheterization. The catheterization laboratory patient population now consists primarily of those with residual lesions or simple unoperated lesions who come for interventional management.

Table 34.1 compares the incidence of the more commonly studied lesions in adults (older than 21 years of age) catheterized at our hospital during the periods 1973–1978 and 1993–1998. This chapter attempts to reflect these changes. The techniques involved in each instance are described briefly and illustrated by case histories where appropriate. Some information from the increasing number of patients with Fontan-repaired single ventricles is included. More detailed descriptions of the interventional techniques used are presented in Chapter 28 and are also available elsewhere (1).

ATRIAL SEPTAL DEFECTS

In the past, all patients with an ASD were catheterized before cardiac surgical correction. It is now clear that virtually all such defects can be diagnosed by physical examination, electrocardiography (ECG), chest radiography, and, in particular, echocardiography (transthoracic, transesophageal, three-dimensional). Therefore, surgery is undertaken without catheterization at most institutions. However, improving success with an increasing variety of umbrella devices for transcatheter closure of centrally located defects of small or moderate size has resulted in an increase in the number of such patients being catheterized at selected institutions (2–5) (Table 34.1).

Anatomic Types

There are three main types of ASD; namely, ostium secundum, ostium primum, and sinus venosus defects. At our institution, the frequencies of these defects have been 68%, 18%, and 6%, respectively (6). Secundum defects are located in the fossa ovalis below the limbic band; they are usually single and central and are often amenable to device closure. Sinus venosus defects occur in the posterior part of the interatrial septum, near the entrance of the superior vena cava (SVC) or, rarely, the inferior vena cava (IVC) and at this time are not amenable to device closure, largely because of the proximity of nearby right pulmonary veins.

Physiology

The size and direction of shunting through an ASD depend on both the size of the hole and the relative compliances of the right and left ventricles. Factors that lead to right ventricular (RV) hypertrophy, thereby decreasing RV
compliance (e.g., pulmonary stenosis, pulmonary hypertension) induce a smaller left-to-right shunt or a larger right-to-left shunt, whereas factors that reduce left ventricular (LV) compliance (e.g., systemic hypertension, LV infarction) produce a larger left-to-right shunt. In infancy, the RV is normally hypertrophied as a result of intrauterine circulation, and there is little net shunt across an ASD. With increasing age, however, RV hypertrophy recedes, and a substantial left-to-right shunt develops, resulting in clinical detection of ASDs in childhood and early adulthood. The tendency for left-to-right shunting to increase throughout adult life (as systemic arterial blood pressure rises and LV compliance falls) has prompted many authors to advocate a policy of closing all ASDs at the time of diagnosis (6).

Catheterization Technique

As the catheter is advanced from the femoral vein, it frequently passes from right atrium (RA) to left atrium (LA). An alternate approach is to withdraw the catheter from the SVC to the RA with the tip positioned posteromedially, so that it drops beneath the limbic band into the fossa ovalis (as in a Brockenbrough transseptal puncture) and then to the LA. Primum defects are located more inferiorly, and sinus venosus defects are located more superiorly. Difficulty in crossing a known ASD usually implies the presence of the latter type of defect.

Crossing the atrial septum with a catheter does not confirm the presence of an ASD, because a probe-patent foramen ovale (in which the septum primum and septum secundum overlap but can be separated by a catheter or by a marked rise in RA pressure) is a normal finding in many children and in 10% to 15% of adults.

Oximetry Data

ASDs with left-to-right shunts are characterized by an increase, or step-up, in the oxygen content (or saturation) of blood in the RA (Fig. 34.1).

FIG. 34.1.

Cardiac catheterization findings in a 6-year-old girl with an atrial septal defect; note the left-to-right shunt at the atrial level and the small flow gradient across the pulmonary valve.

In children, an increase of 10% or more in oxygen saturation between the high SVC and the RA indicates an abnormal increase at the RA level (6). In adults (see Chapter 9), samples from both the SVC and IVC have been used to quantitate the degree of shunting. IVC blood is poorly mixed because of the streaming of renal vein blood, and multiple IVC samples have proved helpful in this regard.

A significant step-up at the RA level does not guarantee the presence of an ASD; patients who have a VSD and associated tricuspid regurgitation can have identical oximetric data, as can those with a so-called “LV-RA” VSD. Associated abnormalities such as partial anomalous pulmonary venous return can be difficult to diagnose from oximetry data alone, although they are suggested by saturations greater than 85% in the SVC or IVC. Oximetry data are used to calculate pulmonary blood flows and shunts, but because oximetry values can vary by 3% to 6% on a random basis, shunts in which the pulmonary-to-systemic flow ratio is less than 1.5:1 cannot be detected reliably by oximetry (7). Obviously, the absence of a measured shunt does not exclude an important ASD; with the development of pulmonary vascular disease and progressive RV hypertension, RV compliance may fall, with the left-to-right shunt becoming negligible. Further decreases in RV compliance may then result in right-to-left shunting and cyanosis.

Pressure Data

The hallmark of a moderate or large ASD is the equalization of atrial pressures; previous workers documented that right and left atrial pressures are equal within 1 to 2 mm Hg and that a and v wave peaks are within 3 to 4 mm Hg in a good-sized ASD (8). Obviously, the a and v waves tend to become similar in both atria with a large defect. We frequently use the pressure gradient across the atrial septum to differentiate a patent foramen ovale (PFO) from a moderate or large ASD. Conversely, the absence of a transatrial gradient does not make the diagnosis of an ASD;
pericardial tamponade, restrictive physiology, and diseases that raise RA pressures all tend to reduce the normal gradient across the atrial septum.

**Angiography**

Until recently, angiography was an unimportant part of catheterization in a patient with an ASD; pressure equalization across the atria, catheter course, and a step-up at the atrial level together make a compelling diagnosis. Many cardiologists do a pulmonary artery (PA) angiogram to visualize pulmonary venous return. With the advent of transcatheter closure of ASDs, both angiography and balloon sizing of atrial defects are carried out to assess whether transcatheter closure is feasible (see Chapter 28); this is nowadays the main reason for catheterizing these patients. Whenever we suspect the presence of an ostium primum ASD in a patient undergoing cardiac catheterization, we perform an LV angiogram in a so-called hepatoclavicular view (40° cranial, 40° left anterior oblique [LAO]) to outline the integrity of the most posterior part of the interventricular septum (9).

**Interventional Catheterization**

Although sinus venosus and ostium primum defects are not amenable to transcatheter double-umbrella closure, about 50% to 60% of patients with secundum ASDs appear to be good candidates (2),(10). In those lesions suitable for closure (less than 24 mm stretched diameter), balloon sizing of the defect with a soft, deformable balloon and angiography precede device placement (see Chapter 28). All other ASDs are surgically closed, with a ministernotomy technique for most secundum defects.

**Case History 1**

A 6-year-old asymptomatic girl was evaluated for a heart murmur. A grade 2/6 systolic ejection murmur was detected at the left upper sternal border, and a diastolic flow rumble of similar intensity was noted inferiorly. Her second heart sound was widely split and fixed. On ECG, right axis deviation and some RV hypertrophy were present, and echocardiographically a central 12-mm ASD secundum was identified. At catheterization (Fig. 34.1), an ASD of moderate size was confirmed, with a small transatrial mean gradient of 2 mm Hg and a pulmonary-to-systemic flow ratio of 2.8. The ASD was then closed with a 33-mm double umbrella device.

**VENTRICULAR SEPTAL DEFECTS**

It is increasingly rare for patients with VSDs to be referred for initial diagnosis during adulthood. Loud murmurs permit diagnosis of the patient with a VSD at an early age; large defects produce symptoms early in life and demand early closure; and small defects tend to get smaller (and often close completely) with advancing age. Pediatric cardiologists tend to “fish or cut bait” in patients with VSDs by 2 years of age, or certainly by age 5; defects unclosed by that age will probably never require surgical attention. Although today most single, clinically significant VSDs are repaired surgically without catheterization, particularly in infancy, there are patients with congenital, postoperative, posttraumatic, or postinfarction VSD who require diagnostic or interventional cardiac catheterization, or both.

**Anatomic Types**

Most congenital VSDs are defects in or about the membranous septum (“perimembranous”) and are located just underneath the aortic valve. Atrioventricular canal–type VSDs are less common, are developmentally related to ostium primum defects, and occur in the posterior interventricular septum adjacent to the atrioventricular valves. Like ostium primum defects, they tend to be associated with a counterclockwise superior QRS in the ECG frontal plane and are seen more commonly in patients with Down syndrome. Subpulmonary VSDs result from deficiency of the conal septum, occur more commonly in Asian patients, and are frequently associated with prolapse of the right coronary cusp and aortic regurgitation. In Caucasian patients aortic regurgitation also occurs, but it is more commonly associated with membranous defects and often with prolapse of the noncoronary cusp alone or in conjunction with the right cusp (11).
Muscular VSDs can occur anywhere in the interventricular septum. Most are “midmuscular,” located just below the moderator band in the RV, although defects can occur in the apical, anterior, or posterior septum. Finally, some defects, termed Swiss-cheese VSDs, have large openings on the LV side of the septum but then are divided into a myriad of channels by muscle bundles on the RV side.

**Physiology**

Because shunting through a VSD occurs primarily in systole, the size and direction of shunting are determined mostly by the afterload that each ventricle faces. Therefore, factors that increase LV afterload (e.g., hypertension, coarctation) or decrease RV afterload (e.g., the fall in pulmonary resistance that occurs normally in early infancy) increase the left-to-right shunt, and factors that decrease LV afterload (e.g., vasodilator therapy) or increase RV afterload (e.g., the development of pulmonary stenosis or pulmonary vascular disease) decrease the left-to-right shunt or even produce a right-to-left shunt and cyanosis.

There is a strong natural tendency for VSDs to close with advancing age; most muscular defects close or become small by 5 years of age, and perimembranous defects may close by aneurysm formation or by adherence of the septal leaflet of the tricuspid valve to the edges of the defect. Therefore, medical and not surgical management is generally advised initially for any restrictive VSD in an asymptomatic patient.

**Catheterization Technique**

Until recently, catheter passage through a VSD was avoided for the most part, because it was unnecessary in making the diagnosis or estimating the size of the shunt. With increasingly successful efforts to close certain VSDs using a transcatheter umbrella approach (see Chapter 28), it is important to emphasize the catheter courses in these defects. For perimembranous defects near the tricuspid valve, the catheter is advanced into the RV and turned posteriorly (clockwise) to cross the VSD into the LV outflow tract. Midmuscular and apical VSDs are most easily crossed with balloon flotation catheters from the LV, whereas anterior muscular VSDs are best crossed from RV to LV with precurved stiff catheters and soft torque-control wires. Right-sided heart pressures and oximetry measurements from the wedge position, distal PAs, main PA, RV, RA, and SVC define the pulmonary vascular resistance and the shunt size (Fig. 34.2).

**FIG. 34.2.**

Cardiac catheterization findings in 2-year-old patient with large, perimembranous ventricular septal defect; note the left-to-right shunt at the ventricular level, pulmonary artery mean pressure at two thirds of the systemic level with mildly elevated pulmonary vascular resistance, and elevated left ventricular filling pressure.

In patients with elevated PA pressure and pulmonary vascular resistance, previous authors have advanced the notion of a “reactive” pulmonary vascular bed; if the pulmonary vascular resistance falls in response to a vasodilator agent (nitric oxide), it is more likely to fall after corrective surgery (12).

**Pressure Data**

Patients with small VSDs have a large pressure gradient across the interventricular septum. Equalization of ventricular pressures always occurs with a large VSD but does not prove the presence of a large defect; small defects with associated pulmonary hypertension may mimic the pressure findings in a large defect. Therefore, the size and location of a VSD must be determined by an imaging technique.

**Angiography**

Optimal angiography of VSDs utilizes the long axial view technique introduced by Bargeron et al. (13). Because the interventricular septum may be regarded as having the surface of a cone that points from right back to left front, any
Left ventriculogram in the long axial oblique view, demonstrating a conoventricular ventricular septal defect in tetralogy of Fallot.

**FIG. 34.3.**

Left ventriculogram in long axial oblique view demonstrating apical muscular ventricular septal defect.

**FIG. 34.4.**

Left ventriculogram in long axial oblique view demonstrating apical muscular ventricular septal defect (arrow).

The anterior portion of the septum is best seen with a straight anteroposterior (or even a right anterior oblique) view, and the posterior portion of the septum (the location of atrioventricular canal defects and posterior muscular defects) is best seen with a four-chambered view (see Chapter 6). As usual, injection of contrast material into the high-pressure chamber (LV) is required; because anatomic definition is needed rather than extrasystole-free function evaluation, in children we inject nonionic contrast volumes (1 to 1.5 mL/kg) at rapid rates (less than 1-second injection times) to outline the defects.

**Interventional Cardiology**

Our earlier experience with transcatheter closure of selected VSDs using a double umbrella (14) has now been expanded to some 200 cases, and results continue to improve. Such transcatheter closure (see Chapter 28) is our treatment of choice for apical and anterior muscular defects and for most residual postoperative defects. Evaluation of this technique in the management of postinfarction defects continues. Considering that the aortic and tricuspid valves are close to the edges of most perimembranous VSDs, surgery currently remains the mainstay of management for these patients.

**Case History 2**

A 2-year-old patient presented with a large perimembranous VSD and continued to exhibit congestive heart failure and failure to thrive despite medical treatment. At catheterization (Fig. 34.2), a large left-to-right shunt at the ventricular level (pulmonary-to-systemic flow ratio, 3.1) was identified, with PA mean pressure at two thirds of the systemic level, mildly elevated pulmonary resistance (3.5 Wood units), and mildly elevated left ventricular end-diastolic pressure (LVEDP) at 14 mm Hg. Angiographically, a perimembranous VSD was identified. The defect was successfully closed surgically with a patch via a transatrial approach, with return of PA pressure to normal.

**Case History 3**

A 3-year-old patient who had had surgical repair of both a membranous VSD and coarctation of the aorta in infancy was catheterized because of an unsuspected additional VSD. A large, muscular apical VSD was identified and closed at that study with a 28-mm double-umbrella device.

**PATENT DUCTUS ARTERIOSUS**

One of the more common forms of congenital heart disease, PDA is usually diagnosed in childhood and corrected at the time of diagnosis. Patency of the ductus is maintained before birth by the production of prostaglandin E (PGE). Premature babies may have an incidence of PDA as high as 40%; these are readily closed by administration of the cyclooxygenase inhibitor indomethacin in most cases.

In the past, the diagnosis of a PDA was made on the basis of physical examination and echocardiography and then the
lesion was closed surgically. Catheterization was reserved for those patients with unusual findings or suspected pulmonary hypertension. The development of safe, reliable methods for transcatheter PDA occlusion (15–19) has reestablished the importance of catheter techniques in this lesion and brought many more patients, including adults with PDA (Table 34.1), to the catheterization laboratory. Alternatively, the PDA can be closed surgically by the video-assisted thoroscopy (VATS) technique.

PDAs are almost always located off the underside of the aortic arch just distal to the origin of the left subclavian artery, left of the trachea, and proximal to the left main stem bronchus. Most commonly, they have an hourglass shape with a prominent aortic diverticulum and a narrowing near the PA end. Some are cone shaped or arise from an anomalous left subclavian artery in a patient with a right aortic arch.

**Physiology**

PDAs are rarely large, except perhaps in patients with Down syndrome and those who live at high altitude. The usual “restrictive” PDA is characterized by a measurable step-up in PA blood oxygen saturation, perhaps some pulmonary hypertension, and no change in aortic or RV blood oxygen saturation (Fig. 34.5).

**FIG. 34.5.**

Cardiac catheterization findings in a 2-year-old child with a small patent ductus arteriosus; note the left-to-right shunt at the great vessel level.

The size of the shunt usually does not change much with the passage of time, although the elderly patient with a PDA may tolerate a relatively small shunt poorly. In our experience, patients with small PDAs remain asymptomatic throughout childhood and most of adulthood but are likely to become symptomatic in their sixties and seventies.

**Catheterization Technique**

The techniques of a standard right-sided heart catheter study estimate the hemodynamic effects of a PDA. Because in utero the ductus arteriosus is merely an extension of the main PA, any soft, straight catheter usually passes from the main PA into the descending aorta across the PDA or in the opposite direction from an aortic approach. Transcatheter closure is accomplished either from a venous approach for double-umbrella or coil occlusion or from a retrograde arterial route for coil occlusion.

**Oximetry Data**

Although it is seemingly straightforward, calculation of the shunt in a patient with a PDA is technically difficult. Because aortic blood crosses into the PA without passing through a mixing chamber, there is considerable streaming in the PAs. Blood sampled from the cephalad portion of the main PA is often fully saturated; left PA blood usually has a higher oxygen saturation than that from the right PA; and a “mixed” PA value cannot be defined accurately. Similarly, in the presence of a right-to-left shunt, descending aortic blood is bluer than ascending aortic blood. Therefore, shunt sizes are estimates at best and do not allow accurate calculation of resistances. If it is necessary to determine (in a patient with a hypertensive PDA) whether closing of the PDA will result in a fall (or a rise) in PA pressure, one must temporarily balloon-occlude the duct and remeasure saturations and pressures.

**Pressure Data**

Most PDAs do not alter right- or left-sided heart pressures unless they are large. In the face of any left-sided heart abnormalities (e.g., poor LV function, aortic stenosis), a PDA increases LV systolic pressure, LV diastolic pressure, or both.

**Angiography**
Needed to provide an accurate landmark for transcatheter closure, angiographic definition of PDA is best done in a straight lateral view with contrast material injected distal to the PDA so that it is outlined before the transverse aortic arch (Fig. 34.6).

**FIG. 34.6.**

Lateral aortogram showing a small (2 mm diameter) patent ductus arteriosus (PDA) (arrow) in a 2-year-old child. Contrast material has been injected distal to the PDA so that the PDA is outlined before the transverse aortic arch.

**Interventional Catheterization**

Although a plug and long-wire technique had been used to close PDAs in the catheterization laboratory for more than 20 years by Porstmann and colleagues (20), most workers use coils in small PDAs and a modification of the double-umbrella technique of Rashkind (21) or a Grifka coil and bag (19) in larger ones (see Chapter 28).

**Case History 4**

A symptomatic 2-year-old girl was seen for evaluation of a loud, continuous murmur in the left infraclavicular area. Her ECG and chest radiograph were normal, and a constrictive PDA was confirmed echocardiographically. At catheterization, a small left-to-right shunt and normal pulmonary resistance were measured (Fig. 34.5), and a small PDA was visualized angiographically (Fig. 34.6). Using a single Gianturco coil delivered in retrograde fashion, the PDA was closed completely with concomitant elimination of the murmur.

**AORTIC STENOSIS**

Aortic stenosis, the great majority of which occurs at valvar level, remains a common form of congenital heart disease in both children and adults. The advent of Doppler echocardiography has markedly improved the noninvasive assessment of obstruction severity, and therefore the need for frequent diagnostic cardiac catheterizations in patients with this lesion has largely disappeared. In general, catheterization in the patient with valvar aortic stenosis is undertaken for balloon valvotomy, which we recommend whenever cardiac dysfunction appears in the neonate or whenever the peak-to-peak transvalvar gradient (at normal cardiac output) is higher than 50 mm Hg and associated with mild (at most) aortic regurgitation in older children. Rarely, patients have symptoms or ECG changes out of proportion to their estimated transvalvar gradient, prompting a diagnostic catheterization.

**Anatomic Types**

More than 75% of children with valvar aortic stenosis have a bicommissural valve (rarely, uncommissural) with leaflet fusion. In these cases, absence of the intercoronary commissure is more common than absence of that between the right and noncoronary cusps. Absence of the commissure between the left and noncoronary cusps is extremely rare. If only one commissure is affected, there is little fusion between the cusp edges, and the valve is not thickened, then there may be little gradient or murmur, with the only finding being a constant aortic ejection click at the cardiac apex. Progression of obstruction occurs in one third of those with valvar aortic stenosis, making careful follow-up mandatory (22).

In a small proportion of patients with congenital aortic stenosis, the obstruction is subvalvar owing to either a thin fibrous ridge or fibromuscular dysplasia of the LV outflow tract. These lesions are progressively obstructive in a large proportion of cases and are known to cause deterioration of the otherwise normal aortic valve, producing aortic regurgitation. Surgery is generally indicated for significant stenosis or symptoms in order to protect the aortic valve, but it is probably best delayed until after the first decade of life to reduce the risk of recurrence.

Finally, some children have an hourglass deformity above the aortic valve, so-called supravalvar aortic stenosis. Caused at least partly by thickening of the supracoronary ridge, supravalvar aortic stenosis is often seen in association with Williams syndrome and with branch PA stenosis.
Physiology

The clinical findings in all three lesions are similar in these patients with uncompromized LV function, except for the absence of an ejection click in subvalvar and supravalvar aortic stenosis. In addition, although patients with supravalvar aortic stenosis usually have large coronary vessels with unobstructed flow, a few have stenosis of a coronary ostium or even occlusion of the orifice by an adherent cusp, with evidence of ischemia.

Catheterization Technique

Most of what has been written about catheterization technique in adults with aortic stenosis (see Chapters 4 and 5) applies to children with congenital aortic stenosis as well. In addition, congenitally narrowed valves tend to have an opening in the posterior part of the valve, between the left and noncoronary cusps. As a result, once the catheter crosses the valve it tends to pass posterior among the chordae of the mitral valve. Because crossing congenitally stenotic aortic valves can be difficult, we sometimes use a side-arm arterial sheath that is one size larger than the catheter; simultaneous pressures are measured from the catheter and the sheath with the catheter in the ascending aorta to directly measure the amount of pulse delay and pulse amplification. Then, once the catheter is advanced across the valve, a simultaneous measurement of LV and femoral artery pressures allows accurate assessment of the transvalvar gradient and valve area. Beyond infancy, we usually place a second arterial catheter in the ascending aorta via the contralateral femoral artery.

Oximetry Data

There are no oxygen saturation changes in the left or right heart blood in these patients. Indeed, the normal variation in right-sided heart oximetry values was established primarily from children with mild valvar aortic or pulmonary stenosis.

Pressure Data

In addition to the simultaneous pressure tracings from the LV and aorta, a pullback tracing across the aortic valve always should be obtained as an internal control. Multihole pigtail catheters are used to enter the LV; a pullback tracing with these catheters may not localize the presence of subvalvar or supravalvar stenosis, making use of an end-hole catheter necessary for this purpose.

Angiography

Both aortography and left ventriculography should be obtained in patients with aortic stenosis. Aortography (usually in anteroposterior and lateral views) assesses the degree of aortic regurgitation, as well as anatomy and mobility of the leaflets and coronary arteries grossly. Ventriculography (LAO with cranial angulation) is used to measure the annulus diameter, to estimate ventricular function, to identify subvalvar pathology, and to further outline valvar and supravalvar anatomy. Multiple views may be required to best outline the subvalvar region, including a right anterior oblique view with caudal angulation.

Interventional Catheterization

Balloon valvotomy (see Chapter 28) has become the treatment of choice for valvar aortic stenosis at our institution. The results are roughly equivalent to those of surgical valvotomy, even in some of our older patients in the fourth decade of life. Although balloon valvotomy may reduce the gradient in some cases of membranous subaortic stenosis, it has been ineffective in most cases in our experience.

Case History 5

An 11-day-old infant was seen with critical valvar aortic stenosis. He was intubated, ventilated, and given PGE1 to
reopen his PDA and improve cardiac output. He was catheterized via an umbilical artery and femoral vein. The hemodynamic data included an elevated LVEDP of 25 mm Hg, a peak systolic ejection gradient of 90 mm Hg across the aortic valve, moderate mitral regurgitation, and a left-to-right atrial shunt. A balloon valvotomy was carried out via the umbilical artery; the peak transvalvular gradient reduced to 37 mm Hg and the LVEDP to 11 mm Hg. Some 5 years later, a second balloon valvotomy was carried out for recurrent obstruction, with reduction of the gradient from 80 to 30 mm Hg via a femoral artery approach with only mild aortic regurgitation resulting. Both the atrial shunt and mitral regurgitation had resolved spontaneously after the initial valvotomy.

**PULMONARY STENOSIS**

Obstructions to the RV outflow tract usually are seen in association with other congenital lesions, such as TOF, transposition of the great arteries, or single ventricle. Isolated valvar pulmonary stenosis is nonetheless common, although it is frequently an asymptomatic lesion beyond the neonatal period. Noninvasive diagnosis and severity assessment are now accurate in almost all circumstances, and catheterization is reserved for those who require balloon valvotomy, at any age.

**Anatomic Types**

Valvar obstruction accounts for more than 80% of isolated pulmonic stenosis. In typical valvar pulmonary stenosis the annulus is of normal size, the leaflets are thin, the commissures are fused, and there is marked poststenotic dilation. Dysplastic pulmonary valves exhibit a different pathology: the annulus is small, the leaflets are markedly thickened (usually thicker than they are long), they do not move during systole, and they are fused. In addition, the main PA is short and narrow. A spectrum of anatomic variants exists between these extremes, and all but the most dysplastic valves can be dilated successfully.

RV muscle bundles occur as anomalously thickened bundles of muscle within the RV cavity and are usually associated with other lesions such as VSD or subvalvar aortic stenosis. The pulmonary valve and annulus are generally normal. Unlike other forms of pulmonary stenosis, muscle bundles frequently become progressively more obstructive.

The rarest form of pulmonary stenosis is branch PA stenosis. It is often associated with both supravalvar aortic stenosis and Williams syndrome.

**Physiology**

Except for anomalous muscle bundles, pulmonary stenosis rarely increases in severity after the first year of life. Severe forms of stenosis in the neonate generally lead to markedly reduced RV compliance with increasing RA pressure and then right-to-left shunting across a PFO. Therefore, critical valvar pulmonary stenosis commonly manifests with cyanosis and sometimes with heart failure. Moderate degrees of stenosis (e.g., gradients of 40 to 80 mm Hg) rarely cause symptoms. However, several studies have shown that patients with moderate pulmonary stenosis have decreased exercise performance at cardiac catheterization, even in childhood (23). On the basis of these studies and because balloon valvotomy is both safe and successful, most cardiologists now recommend dilation in any patient with more than mild stenosis (Fig. 34.7), on an elective basis, before 5 years of age.

**FIG. 34.7.**

Catheterization data in 5-year-old patient with moderate valvar pulmonary stenosis, with a peak systolic pressure gradient of 55 mm Hg.

**Catheterization Technique**

The foramen ovale is usually open, allowing catheter access to the left side of the heart from a femoral venous
approach. Arterial catheters usually are required only in ill neonates and those with severe or more obstruction. The opening in the pulmonary valve of an infant with severe pulmonary stenosis may be extremely small (less than 1 to 2 mm in diameter) (Fig. 34.8).

**FIG. 34.8.**

Lateral right ventricular angiogram in a neonate with critical valvar pulmonary stenosis showing contrast jet (arrow) through stenotic orifice and poststenotic dilation of main pulmonary artery.

Passage of a standard 5F or 6F catheter across such a valve essentially occludes the orifice, stops pulmonary blood flow, and may result in hemodynamic collapse in less than 1 minute. For this reason, we do not routinely measure PA pressure directly in infants with critical valvar pulmonary stenosis. Rather, we use preformed 4F catheters and a 0.018-inch torque wire to cross the valve, followed by rapid balloon dilation.

**Oximetry Data**

Most of these patients have neither a right-to-left nor a left-to-right shunt. In critical severe valvar pulmonary stenosis, however, a right-to-left shunt occurs at the atrial level, producing cyanosis. In these patients, the pressure gradient underestimates the severity of the stenosis (because less than a full cardiac output is crossing the pulmonary valve), and the pulmonary venous blood is fully saturated with oxygen (Fig. 34.7). Rarely, one encounters a patient with both mild pulmonary stenosis and an ASD. With a large left-to-right shunt at the atrial level, the gradient across the pulmonary valve is “falsely” elevated. Closing the ASD reduces the RV pressure as well as the gradient across the pulmonic valve.

**Pressure Data**

In mild pulmonary valve stenosis, the RV pressure is normal. As the degree of stenosis increases, the RA a wave increases. With severe pulmonary stenosis, RV systolic pressure approaches or exceeds LV pressure, main PA pressure falls, and the PA pulse pressure dampens. Marked hypertrophy of the RV may cause the infundibular os to close during late systole, resulting in further obstruction.

**Angiography**

Right ventriculography in a straight lateral view outlines both the pulmonary valve and the subvalvar region (Fig. 34.8).

Although a straight anteroposterior view with cranial angulation also may outline the pulmonary valve, poststenotic dilation of the main PA often obscures the PA branch origins. Following the contrast material temporally until it appears in the LA allows one to rule out an associated ASD. Further angiography is rarely required.

**Interventional Catheterization**

As noted in Chapters 26 and 28, balloon dilation, safe and effective at all ages, has become the treatment of choice for this lesion.

**COARCTATION OF THE AORTA**

Catheterization of the patient with isolated coarctation for diagnostic purposes is rarely required before surgery, because improved noninvasive imaging, including magnetic resonance imaging, allows precise anatomic definition of the lesion. However, the continued incidence of recurrent coarctation after surgical repair, the common association of coarctation with other forms of congenital heart disease (especially VSD, valvar or subvalvar aortic stenosis, and
mitral stenosis), and increasing use of balloon dilation in discrete nonoperated coarctation make this a common lesion studied in the laboratory.

Anatomic Types

Virtually all forms of coarctation occur at or just distal to the left subclavian artery, at or near the level of the old ductus arteriosus. Coarctations have discrete “curtains” of tissue indenting the posterior wall of the aorta, although they may be associated with hypoplasia of the transverse aortic arch. Rarely, a coarctation involves a long segment of thoracic or abdominal aorta.

Physiology

The gradient across a coarctation is influenced not only by the degree of obstruction but also by the degree of collateral flow around the obstruction. Collateral flow is carried most commonly by the internal mammary, the intercostal (whose enlargement may eventually produce rib notching), the scapular, and the epigastric arteries. Rarely, an obstruction in the transverse aortic arch produces a cerebral steal from the carotids, through the circle of Willis and down the left vertebral artery.

Upper-extremity hypertension, the primary sequela of a coarctation, usually resolves after surgical correction. In some children, however, hypertension persists despite adequate anatomic repair. This persistent hypertension is more common when repair occurs late in childhood, prompting the general recommendation that coarctation should be diagnosed and corrected before the child reaches 3 years of age.

Catheterization Technique

Crossing a coarctation is usually straightforward and not hazardous. Occasionally, when catheterizing an older patient with coarctation, one may enter an enlarged collateral vessel thinking it to be the aorta. Complex catheter manipulation in this confined space may prove difficult and even hazardous.

Oximetry Data

Patients with isolated coarctation have no abnormalities in their intracardiac oxygen saturations. Those with associated intracardiac defects (ASD, VSD) have the size of their left-to-right shunt augmented by the coarctation-induced increase in LV afterload.

Pressure Data

Even mild coarctations have a systolic pressure gradient across the coarctation site, although only a minimal diastolic gradient is found. As obstruction increases, a gradient is present throughout the cardiac cycle. The gradient measured by pullback in the catheterization laboratory is frequently smaller than the gradient measured by sphygmomanometer in clinic; although the normal pulse amplification persists in the arms in patients with coarctation, it is frequently absent in the legs, contributing further to the classic finding of diminished femoral pulses on physical examination.

Angiography

A straight lateral aortogram usually provides excellent visualization of the coarctation. If the contrast injection also opacifies the head and neck vessels, an estimate of collateral flow can be obtained. Such estimates are not precise; if the coarctation site is balloon-occluded and pressures are measured in the descending aorta, a more precise estimate of collateral adequacy is available. After surgery has distorted the coarctation site, a lateral aortogram may not define the anatomy well, and various oblique views with cranial or caudal angulation may be needed to define the narrowest site.

Interventional Catheterization
Although balloon dilation frequently reduces the gradient across an unoperated coarctation site, the results (particularly in babies) are not generally as good as those seen with surgical management. However, in thin membranous obstructions beyond infancy, the use of this technique, with care, is increasing and appears effective. Balloon dilation of recurrent coarctation has, however, proved clinically invaluable with or without stent placement.

**TETRALOGY OF FALLOT**

The association of a malalignment VSD, infundibular and valvar pulmonary stenosis with resultant aortic overriding, and cyanosis is referred to as TOF complex. It remains a difficult and important surgical challenge. Catheterization of infants with this lesion, especially if they are cyanotic, may be hazardous. Therefore, if echocardiographic information is adequate in this age group, surgery is undertaken without catheterization. However, if anatomic details are uncertain, these babies may be studied with great care and with particular emphasis on not traversing the RV outflow tract. Beyond infancy, many patients with TOF are still catheterized preoperatively, particularly if pulmonary atresia with aortopulmonary collaterals is present.

**Anatomic Types**

In addition to the lesions already mentioned, common additional defects include (a) branch PA stenosis (5% to 10%), (b) pulmonary atresia with PDA-dependent pulmonary blood flow (5% to 10%), (c) additional muscular VSDs (5% to 10%), (d) aortopulmonary collateral arteries supplying blood flow to the lungs (5% to 10%), and (e) coronary arterial anomalies, especially the left anterior descending arising from the right coronary artery (1% to 2%). Each of these variables must be assessed accurately and considered before an attempt at operative repair is made.

**Physiology**

The combination of pulmonary obstruction and a VSD produces a right-to-left shunt at the ventricular level. The size of this shunt is unrelated to the size of the VSD, which in TOF is almost always large and unrestrictive. Rather, the right-to-left shunt and degree of cyanosis are determined primarily by the degree of pulmonary obstruction and less by the level of systemic vascular resistance. So called “tetrad spells” (characterized by hyperventilation, acidosis, extreme desaturation, and unconsciousness) or hypercyanotic episodes may be provoked by increased pulmonary obstruction or decreased systemic resistance, or both; they are best treated by sedation (which may decrease catecholamine tone and lower the level of muscular pulmonary obstruction), intravenous volume infusions, and increasing the systemic vascular resistance.

In general, the anatomic obstruction to pulmonary blood flow tends to increase with time in these patients, thereby increasing the degree of right-to-left shunting. Previously, aortopulmonary shunts such as the Blalock-Taussig shunt were created surgically to increase pulmonary blood flow. Currently, so-called complete cardiac correction, relieving the pulmonary stenosis and closing the VSD, is usually undertaken in infancy or early childhood.

**Catheterization Technique**

Because a PFO is the rule in these patients, one can enter the left side of the heart from the RA without difficulty. Similarly, one can generally pass the catheter from the RV both to the aorta and into the PA. Catheter passage into the PA, however, often provokes a hypercyanotic spell, and catheter passage from the RV to the aorta often produces transient heart block. Both are to be avoided.

**Oximetry Data**

Because the VSD is just below the aortic valve in patients with TOF, there are usually no abnormalities in oxygen saturation at the atrial level or the ventricular level. The right-to-left shunt is documented only in the aorta.

Small changes in the degree of RV outflow obstruction or level of systemic resistance have major effects on the
Right ventriculogram in tetralogy of Fallot demonstrates subpulmonary valvar stenosis, small main pulmonary artery, and early filling of the aorta through right-to-left shunting via the ventricular septal defect.

**Pressure Data**

Almost by definition, RV and LV pressures are equal in patients with TOF. Both atrial pressures are usually normal, as are LV and aortic values. PA pressures (when measured) are decreased in cyanotic patients without prior surgery; in the presence of the surgically created Waterston or Potts aortopulmonary shunts, the PA is often distorted and pressures are elevated even to the level of advanced pulmonary vascular disease, but this is rarely so after Blalock-Taussig shunts.

**Angiography**

Angiographic definition of anatomic detail (Fig. 34.9) is the key element in the cardiac catheterization of patients with TOF.

**FIG. 34.9.**

Right ventriculogram in tetralogy of Fallot demonstrates subpulmonary valvar stenosis (*open arrow*), small main pulmonary artery (*white arrow*), and early filling of the aorta through right-to-left shunting via the ventricular septal defect.

A biplane RV angiogram (with cranial angulation of the anteroposterior camera) establishes the diagnosis, defines the anatomy of the pulmonary valve and subpulmonary region, and identifies the main PA and proximal PA branches. A left ventriculogram in the so-called long axial oblique view but with less LAO, because the ventricular septum runs more straight right to left, outlines the VSD and may identify the coronary artery pattern. An ascending aortogram may be necessary to further delineate coronary artery anatomy, and in particular to identify the origin of the left anterior descending artery. Aortography (especially of the descending aorta) is also often necessary to see whether any aortopulmonary collaterals are present.

**Interventional Catheterization**

Interventional procedures are rarely required before definitive surgical correction in patients with uncomplicated TOF. However, in infants with TOF, pulmonary atresia, and diminutive PAs, dilation of hypoplastic PAs and coil embolization of aortopulmonary arteries make up an essential component of management. Similar interventional procedures together with PA stent placement are often required postoperatively.

**Case History 7**

A 36-year-old woman with TOF was seen for evaluation. She had undergone a classic left Blalock-Taussig shunt for cyanosis at 5 years of age and had been lost to cardiology follow-up since that time. She was married, had had three miscarriages but no live children, and worked full-time. On examination she had marked generalized cyanosis and clubbing, a grade 3/6 continuous murmur at the left upper sternal border, and a single second heart sound. Her ECG showed right axis deviation, and both RA and RV hypertrophy were evident. Her hematocrit was 70% and her platelet count 96,000/mm³. Echocardiographic information was limited because of patient size and poor windows, but a VSD was visualized with predominant right-to-left shunting.

She underwent repeated red cell phereses and then had cardiac catheterization (Fig. 34.10).

**FIG. 34.10.**
Catheterization findings in a 36-year-old woman with tetralogy of Fallot showing right-to-left shunting at the ventricular level (larger arrow), with aortic saturation lower than the left ventricular value due to streaming, equal ventricular pressures, a 93 mm Hg peak gradient across the pulmonary outflow tract and a left Blalock-Taussig shunt (smaller arrow).

There was right-to-left shunting at the ventricular level via the VSD and a small left-to-right shunt at the PA level due to the Blalock-Taussig shunt. The RV pressure was at the systemic level, a peak gradient of 93 mm Hg was measured across the stenotic RV infundibulum and pulmonary valve, and the PA pressure was low. The Blalock-Taussig shunt was dilated with an 8-mm balloon, resulting in a 5% rise in aortic saturation. TOF was confirmed angiographically, with a single VSD, marked infundibular and valvar pulmonary stenosis, adequate PA size, and normal coronary arteries. Two months later, the TOF was repaired surgically, and at follow-up 1 year later the patient was asymptomatic. There had been a dramatic improvement in her activity level, cyanosis was absent, and a substantial reduction in the degree of digit clubbing had occurred.

TRANSPOSITION OF THE GREAT ARTERIES

In transposition of the great arteries (TGA), the great arteries arise from the wrong ventricles (i.e., the aorta from the RV and the PA from the LV). There are two main types. By far the more common variety is known as dextro-TGA (DTGA); the ventricle position is normal (i.e., the RV is right-sided and the LV left-sided, with the RV giving rise to a right-sided anterior aorta and the LV to a left-sided posterior PA). In the most common form of DTGA, there are no other major anomalies; a smaller number of patients have a VSD in addition, and in the least common variation subpulmonary stenosis is present in addition to the VSD. BAS (24), PGE1, and, in particular, the introduction of the remarkable arterial switch operation in the first few days of life have made DTGA, with or without VSD, a correctable lesion and no longer a lethal one. The other type of TGA, referred to as levo-TGA (LTGA) or corrected transposition, is much less common. In this anomaly the ventricles are inverted (the so-called L-loop), so that the LV is right-sided and the RV left-sided, with the LV giving rise to a right-sided PA and the RV to a left-sided aorta. This type of transposition is almost always accompanied by a VSD and subpulmonary stenosis, is often by tricuspid regurgitation and atioventricular conduction abnormalities, and is a very difficult lesion to deal with. The following brief comments are confined to the more common DTGA variety.

Physiology

Unlike the normal circulation, in which blood travels to the lungs and then to the body in sequential fashion, the pulmonary and systemic circulations in TGA run in parallel: Red blood coming back from the lungs returns to the lungs, and blue blood coming back from the body returns to the body. Without a defect in the circulation (e.g., ASD, PDA, VSD) to allow mixing between the two circuits, the patient would die a few minutes after birth. Therefore, the early goal of therapy is to promote mixing between the circulations by making a hole in the atrial septum (BAS) or between the great arteries (using prostaglandins to open the PDA). These measures may stabilize the patient and relieve acidosis, but they relieve the severe associated cyanosis only partially and must be followed within days by an arterial switch operation, together with VSD closure if the VSD is more than tiny in size.

Catheterization Technique

Because echocardiographic definition is so precise, the only reasons to catheterize such neonates are to do a BAS and, in a few, to outline coronary artery anatomy. The umbilical vessels provide the necessary access routes in the majority. The almost invariable presence of an ASD in TGA makes the entire heart accessible to a venous catheter. Nonetheless, catheter entry into the PA, usually unnecessary in the newborn, may be difficult; after the catheter enters the LV from the LA, it must take a 180° turn, over a short distance, to enter the PA. The use of a tip deflector wire to bend the catheter at the apex of the LV reliably allows PA cannulation.

Oximetry Data
As noted previously, the well-being of the infant before surgical repair is determined primarily by the degree of mixing that occurs between the two parallel circuits. Although the mixing can take place at the atrial, ventricular, or great artery level, its net effect is reflected in the aortic and PA blood oxygen saturations. With perfect mixing, the aortic and PA blood oxygen saturations are equal (at perhaps 82%). As mixing becomes progressively less adequate, the aortic blood oxygen saturation falls and the PA saturation rises (Fig. 34.11).

**FIG. 34.11.**

Catheterization findings in an infant with transposition of the great arteries and a small atrial septal defect. There is bidirectional shunting at the atrial level (arrows), arterial desaturation, poor mixing, and a small systolic pressure gradient across the left ventricular outflow tract.

**Pressure Data**

Because the aorta arises from the RV, the RV systolic pressure is always at least at aortic level. In the early neonatal period, a persistent PDA in many (especially if PGE1 is being administered) causes the PA pressure (and hence LV pressure) to be elevated. Over the first few weeks of life, both PA and LV pressures fall as the PDA closes, unless there is an associated VSD or pulmonary stenosis. Of note, pulmonary stenosis may be masked if a PDA is present, with a pressure gradient developing only when the PDA closes. LA pressure varies with the degree of LV pressure elevation, the amount of pulmonary blood flow returning to the LA, and the size of the ASD; it commonly exceeds that the RA pressure when the ASD is very small.

**Angiography**

RV angiography in standard anteroposterior and lateral views establishes the diagnosis, assesses RV function, determines the presence of a VSD or a PDA, and assesses tricuspid regurgitation. An LV angiogram in the long axial oblique view demonstrates the presence and nature of LV outflow tract lesions and demonstrates the location of a VSD.

With the neonatal arterial switch operation now the surgical procedure of choice, the precise definition of coronary arterial anatomy in TGA is mandatory. We use the “laid-back” balloon occlusion aortogram (25) as the angiographic procedure that best outlines coronary arterial anatomy. With the 45° of caudal angulation inherent in this view, the coronary anatomy is displayed in a transverse section, identifying both the cusp origins and branching patterns of these vessels.

**Interventional Catheterization**

Even though an arterial switch operation is performed electively in the first 10 days of life, we continue to perform a BAS in the early neonatal period (see Chapter 6). Creation of an ASD remains the optimal method for stabilization of these cyanotic infants during the days before surgical repair. Postoperatively, the majority have at most trivial obstruction in the region of the RV outflow–main PA anastomosis, but in some this may be significant together with right PA–and left PA–origin stenosis, this being amenable to balloon dilation/stent placement in some. Significant lesions involving the left heart outflow or coronary arteries are very rare in our experience.

**SINGLE VENTRICLE**

The term “single ventricle” refers to a family of lesions, often complex and with highly variable anatomy, in which there is only one functional ventricular chamber. All these lesions have the features of a single effective pumping chamber with one or two atrioventricular or semilunar valves. Since the introduction of the Fontan procedure and other preceding palliative procedures such as the Norwood stage 1 and bidirectional Glenn operations, it is clear that this patient population with its evolving problems is rapidly increasing (Table 34.1), as are vascular anomalies as a consequence of these operations.
Anatomic Types

The most common form of single ventricle is the highly lethal hypoplastic left heart syndrome (HLHS), which is usually caused by aortic or mitral atresia. In this syndrome, the LV is diminutive, much too small to sustain life. Although in the past neonates with HLHS uniformly died when the PDA closed, the pioneering efforts of Norwood et al. (26) to create a new aorta surgically, using the RV as a single ventricle, have improved the survival of these children dramatically. A less common but similar condition is tricuspid atresia, in which the right side of the heart is hypoplastic and blood flow to the lungs occurs through a VSD into a small RV. Patients with asplenia and polysplenia syndromes also frequently have single ventricles. Nowadays, after a Norwood stage 1 procedure in babies with HLHS and others palliated with a shunt, a second-stage bidirectional Glenn shunt surgical procedure is performed in the latter half of the first year of life. Subsequently, usually 1 or 2 years later, the Fontan procedure is completed, often leaving a fenestration in the atrial baffle. A year or so later this fenestration, if still patent, is closed with the use of a clamshell double umbrella (27). Therefore, most of these patients are catheterized at least twice electively—between the stage 1 and bidirectional Glenn procedures and between the latter and the Fontan operation—and many a third time after the Fontan surgery. In 1971, Fontan demonstrated that atrial pressure alone was enough to maintain a full cardiac output through the lungs, allowing separation of the systemic and pulmonary circulations in children with single ventricle, which eliminates both the volume load on the heart and the deleterious effects of longstanding cyanosis. The resulting RA pressures are elevated in patients so treated, to 12 to 18 mm Hg, and are generally well tolerated.

Catheterization Techniques

Between the Stage 1 and Bidirectional Glenn Procedures

These patients have in common a pulmonary blood flow supplied by an aortopulmonary shunt, usually a Blalock-Taussig shunt, and the mandatory catheterization information consists of measurement of PA pressure and resistance, intracardiac and aortic pressures, and PA and systemic venous anatomic details. Measurement of PA pressure through the shunt is best accomplished by modifying the tip of a pigtail catheter to a 180° bend. This catheter is advanced from the descending aorta into the mouth of the (usually right) subclavian artery. The soft end of an 0.018-inch torque guidewire is passed from the tip of the pigtail to the distal PA via the shunt; once it is securely in the PA, the catheter is advanced over the guidewire to allow both pressure measurement and angiography. An alternative method of PA pressure estimation is to measure the pulmonary venous wedge value, but this is a reliable approximation only when the PA mean is less than 20 mm Hg. Additional angiography is necessary in the left innominate vein to exclude a persistent left SVC-to-coronary sinus which, if present, must be coil-occluded. This vessel, if present and left patent, will enlarge after a bidirectional Glenn procedure because of the increased SVC pressure, resulting in significant right-to-left flow with subsequent further decrease in systemic arterial saturation.

After the Bidirectional Glenn Procedure and Just Before the Modified Fontan Procedure

Because the SVC has been disconnected from the RA and anastomosed to the PA, another venous line from the left subclavian vein is required in the catheterization of these patients, in addition to femoral venous and arterial lines. As before, the basic information required includes, from all sites, measurement of saturations and pressures together with PA resistance and anatomy. In addition, because SVC pressure has been elevated for some time, venovenous channels may have appeared in many cases, such as left innominate to left and/or right pulmonary vein (Fig. 34.12).

FIG. 34.12.

Angiographic demonstration of venous channels from left innominate vein (black arrow) to right and left pulmonary veins (white arrows) resulting in right-to-left shunting, after a bidirectional Glenn procedure.

FIG. 34.13.
Angiographic demonstration of diffuse punctate arteriovenous fistulas in right lung after a bidirectional Glenn procedure, resulting in dramatic decreases in right pulmonary venous and systemic arterial oxygen saturations.

There are many variations in the types of such vessels encountered (28), and all require occlusion (usually by coils) when identified. The development of pulmonary arteriovenous fistulas in these patients is increasingly recognized (29) and is considered to be a consequence of the exclusion of hepatic venous blood from the pulmonary circulation by the bidirectional Glenn procedure (Fig. 34.13).

These are angiographically recognized by their punctate appearance and by the rapid transit (less than three cardiac cycles) of contrast agent through the affected lung to the left PA; they are verified by direct sampling of blood from the appropriate pulmonary veins (29). These lesions are usually too diffuse to attempt coil occlusion, and many appear to regress with time after surgical incorporation of hepatic flow into the pulmonary circuit.

**After the Fontan Procedure**

Many patients after a Fontan procedure who have not had a fenestration placed in the baffle at the time of surgery and who are doing well are not electively catheterized. There are, however, a significant number who had a fenestration placed who later come to have this communication electively closed with a double-umbrella device (27). There are also others who have cyanosis for some other reason or evidence of congestive heart failure/venous hypertension who require catheterization for diagnostic and interventional purposes. Many of the latter have had earlier types of baffling procedures, such as direct RA-to-PA anastomosis or interposition of valved conduits between RA and RV or RA and PA, these being done before the more recent intraatrial lateral tunnel procedures. In those with a simple fenestration, the studies are straightforward and basically require hemodynamic evaluation (saturation, pressure, and cardiac output measurements) with and without balloon occlusion of the fenestration. If such temporary occlusion is hemodynamically tolerated, the fenestration is occluded with a double-umbrella device. In the others, the catheterizations tend to be very long and complex, and meticulous evaluation of a large and growing list of possible abnormalities with appropriate interventional procedures is necessary. For example, in those with cyanosis, sites of right-to-left shunting may be encountered that result from an ASD, a baffle leak, or a coronary sinus-to-LA shunt, alone or in combination. In those with congestive heart failure/systemic venous hypertension, obstruction may be present in a conduit (Fig. 34.14) or in a branch PA (a 2-mm gradient is very significant), and the latter requires balloon dilation and stent therapy.

**FIG. 34.14.**

Simultaneous right atrial (a)=nright ventricular (v) pressure tracings across a valved RA-RV conduit placed as part of modified Fontan procedure for tricuspid atresia. Significant obstruction (shaded area) is present across conduit.

In those with the earlier RA-RV or RA-PA anastomoses, the RA may be of enormous size and may compress the right pulmonary veins (30). This is best visualized by magnetic resonance imaging and may require surgical conversion to an atrial tunnel channel. Excessive aortopulmonary collaterals may be present; they may lead to ventricular failure and require coil occlusion. Aortic arch obstructions similarly cause ventricular failure and require dilation and stenting even if gradients are small (e.g., 10 mm Hg). In addition, a few patients have significant subaortic stenosis, a complication that is best managed surgically.