Acquired disease of the right-sided cardiac valves is much less common than disease of the left-sided counterparts, possibly because of the relatively lower pressures and hemodynamic stress to which the right-sided valves are subjected. Indeed, right-sided valvular dysfunction is usually seen when morphologically normal valves are subjected to abnormal hemodynamic stresses, such as pulmonary hypertension. Tricuspid and pulmonic valvular abnormalities are also part of numerous congenital syndromes (discussed in section VIII). This chapter focuses on acquired abnormalities of the right-sided cardiac valves, and, because it is frequently diagnosed in adults, pulmonic stenosis.

**TRICUSPID STENOSIS**

Tricuspid stenosis is uncommon. Most cases are due to rheumatic heart disease. When rheumatic tricuspid stenosis is present, it is generally associated with mitral stenosis, which usually accounts for most of the presenting signs and symptoms. Carcinoid heart disease may also cause tricuspid stenosis, and the signs and symptoms may be mimicked by tumors (myxoma or metastasis) or vegetations that obstruct RV inflow.

The symptoms of tricuspid stenosis are primarily from increased systemic venous pressure (Fig. 31-1). Peripheral edema, ascites, hepatic enlargement, and right upper quadrant discomfort may develop with chronic tricuspid stenosis or regurgitation. Decreased cardiac output may cause pronounced fatigue. Jugular venous pressure is increased, and there is a prominent a wave from impaired RV filling during atrial systole. The murmur of tricuspid stenosis is a low-pitched diastolic murmur at the lower left sternal edge. However, this is often obscured by or difficult to differentiate from the usually associated mitral stenosis murmur. The physical examination may demonstrate the presence of tricuspid stenosis in patients with mitral stenosis, including when there is accentuation of the diastolic murmur during inspiration (as is the case for most right-sided murmurs); and/or the presence of a prominent a wave in the jugular venous pulse.

Useful diagnostic studies include chest radiography, ECG, and echocardiography with Doppler evaluation. Right atrial enlargement is frequently evident on radiographs and is manifest on the electrocardiogram as a large peaked P wave in lead II (Fig. 31-2). Because of the increased right atrial pressure, atrial fibrillation is often present.

Echocardiography typically reveals thickened tricuspid leaflets, decreased mobility, scarred chordae, and sometimes doming, if the tricuspid valve leaflets remain pliable. Carcinoid heart disease is associated with a distinctive morphology of a thickened tricuspid valve that is narrowed and fixed in the open position. Doppler evaluation allows estimation of the diastolic pressure gradient by the modified Bernoulli equation. Cardiac catheterization is generally not necessary for the diagnosis of tricuspid stenosis, but when performed it calls for separate, simultaneous catheters in the right atrium and ventricle. If cardiac output is low, tricuspid gradients may also be low and are not adequately evaluated with use of a catheter pullback. Clinically significant tricuspid stenosis is usually associated with a valve area 1.5 cm² or less.

Treatment of tricuspid stenosis includes diuretics and nitrates to relieve venous congestion. Refractory patients can undergo tricuspid valve replacement, but concomitant mitral valve disease primarily determines the indication and timing for surgery. A surgical approach may also be indicated for debulking of obstructive tumors or myxoma. The early experience with percutaneous balloon valvuloplasty for tricuspid stenosis is encouraging.
Tricuspid Stenosis and/or Insufficiency

- Engorged, pulsating neck veins (presystolic in stenosis, systolic in insufficiency)
- “Cogwheel” respiration in insufficiency
- Pulmonary congestion of mitral stenosis decreased by tricuspid disease
- Cyanosis
- II Sounds
  - Soft, blowing systolic murmur; tricuspid insufficiency
  - Coarse diastolic murmur; tricuspid stenosis
- R. atrium enlarged; peaked P wave in lead I; atrial fibrillation common
- Diastolic obstruction in stenosis
- Systolic regurgitation in insufficiency
- R. ventricle may hypertrophy or fail because of associated mitral disease and tricuspid regurgitation
- 3rd L. interspace increased on inspiration
- Portal hypertension
- Ascites
- Liver enlarged, pulsating in insufficiency; slight jaundice possible
- Exaggerated if R-sided heart failure develops
- Moderate edema may be present
- Digesive disorders
- Multivalvular disease (mitral, aortic, tricuspid):
  Enlargement of all chambers and of pulmonary artery

Figure 31-1

TRICUSPID AND PULMONIC VALVE DISEASE

VALVULAR HEART DISEASE
TRICUSPID REGURGITATION

Tricuspid regurgitation may be due to primary disease of the valve apparatus or diseases causing pulmonary hypertension with secondary annular dilatation. Secondary tricuspid regurgitation is seen in any condition associated with increased pulmonary artery pressures, and is the predominant cause of tricuspid regurgitation. The most common secondary causes are LV failure, mitral regurgitation, mitral stenosis, primary pulmonary disease, and primary pulmonary hypertension. The rare causes of primary tricuspid regurgitation include rheumatic heart disease, myxomatous disease (prolapse), infective endocarditis, carcinoid heart disease, and trauma.

Symptoms are often due to associated left-sided heart disease or pulmonary disease. Prominent signs and symptoms of right-sided heart failure suggest tricuspid regurgitation as a component. Endocarditis or carcinoid syndrome may be associated with characteristic systemic symptoms.

Jugular venous pressure is usually increased, and there is a prominent cv wave produced by regurgitant flow into the right atrium. The typical murmur is holosystolic and located at the left sternal edge. Augmentation of the murmur with inspiration helps distinguish tricuspid from mitral regurgitation.

Chest radiography often reveals RV enlargement manifested as filling of the retrosternal space. Dilation of the right ventricle often causes incomplete or complete right bundle branch block, seen on the ECG.

Doppler echocardiography is helpful in evaluating tricuspid regurgitation. Two-dimensional echocardiography evaluates the structure of the valvular apparatus and size of the right atrium and ventricle. Pulse-wave or color flow Doppler reveals the presence, direction, and magnitude of the regurgitant jet. Finally, continuous wave Doppler and the modified Bernoulli equation can be used to estimate the RV and pulmonary artery systolic pressures. In tricuspid regurgitation, the gradient between the right ventricle and the right atrium during systole equals four times the square of the velocity. This gradient is then added to the estimated right atrial pressure (the jugular venous pressure) to estimate RV systolic pressure. In the absence of pulmonic stenosis,
this also equals pulmonary systolic pressure. Note that the calculation estimates the severity of the pulmonary hypertension, not the severity of the tricuspid regurgitation.

The mainstay of therapy for tricuspid regurgitation is treatment of the condition causing pulmonary hypertension. Diuretics may be useful for refractory fluid retention. Tricuspid valve replacement or repair is appropriate for patients refractory to medical therapy or sometimes at the time of surgery for coexistent mitral valve disease. Often a prosthetic ring is used for annuloplasty. If valve replacement is necessary, bioprostheses are favored because the tricuspid valve may be relatively prone to thrombosis.

**PULMONIC STENOSIS**

Right ventricular outflow obstruction may be subvalvular, valvular, or supravalvular. Both the subvalvular and the supravalvular forms are usually associated with other congenital heart disease, as discussed in section VIII. True valvular pulmonic stenosis, however, usually occurs as an isolated congenital defect. Rarely, pulmonic stenosis is due to rheumatic disease, endocarditis, or carcinoid syndrome (Fig. 31-3).

Patients with pulmonic stenosis are often asymptomatic. Patients may reach the fourth through sixth decades of life with significant pressure gradients across the pulmonic valve, but with no symptoms and no evidence of right-sided heart failure. If right-sided heart failure does develop, abdominal swelling, peripheral edema, abdominal discomfort, and fatigue may be present. Patients seldom present with chest pain or exertional syncope.

The physical examination typically reveals a mid systolic crescendo–decrescendo murmur at the left sternal edge. Often, an associated ejection click, which usually decreases with inspiration, is present. P₂ is soft and delayed, producing a widely split S₂, but one that does narrow with appropriate physiologic changes (unlike the fixed, widely split S₂ present in patients with an atrial septal defect). An RV lift may also be present. If RV failure is present, there may be peripheral edema, hepatomegaly, abdominal swelling, and jugular venous distention with a prominent a wave.

Electrocardiography usually reveals RV hypertrophy, right axis deviation, and right atrial enlargement. A complete or incomplete right bundle branch block is sometimes present. Chest radiography reveals poststenotic dilatation of the pulmonic artery but diminished peripheral pulmonary vascular markings. RV hypertrophy and enlargement are highly variable.

Echocardiography with Doppler evaluation is useful for establishing the diagnosis and assessing therapy. Morphologic assessment is best performed with the parasternal short axis view and the subcostal view. Transesophageal echocardiography is not usually necessary but can be performed if a transthoracic study fails to provide an adequate assessment. The right ventricle may be normal, particularly in childhood, but stenosis of long duration, greater severity, or both may be associated with RV hypertrophy and enlargement. Paradoxical motion of the interventricular septum is often apparent. Continuous wave Doppler evaluation is highly reliable in establishing the gradient across the pulmonic valve. Cardiac catheterization is usually not necessary but may be performed if Doppler studies are suboptimal or if balloon valvuloplasty will be performed.

Adult patients with mild-to-moderate pulmonic stenosis generally do well and require no intervention. When symptoms develop, it is often in the fourth decade of life. Balloon valvuloplasty is highly effective and indicated in symptomatic patients and possibly in patients with severe stenosis even in the absence of symptoms (see chapter 33). The most recent ACC/AHA task force has recommended balloon valvuloplasty as a class I indication in asymptomatic adolescents and young adults with transvalvular gradients greater than 50 mm Hg. A gradient of 40 to 49 mm Hg was considered a class IIa indication (meaning that this consideration should be based on individual patient circumstances) based on these recent guidelines.

**PULMONIC REGURGITATION**

Pulmonic valve regurgitation is usually secondary to severe pulmonary hypertension, pulmonic artery dilatation, or both. Rarely, it is secondary to endocarditis, carcinoid syndrome, rheumatic heart disease, trauma, or congenital valvular abnormalities. Accordingly, the dominant symptoms in pulmonic regurgitation are
Pulmonary Valvular Stenosis and Atresia

Pulmonary valvular stenosis with intact septum:
Hypertrophy of right ventricle

Stenotic pulmonary valve viewed from above: Poststenotic dilatation of pulmonary trunk

Complete atresia of pulmonary valve

Bicuspid pulmonary valve
usually those of the underlying disease process. Patients without severe underlying disease are often asymptomatic. However, patients with severe pulmonic regurgitation may ultimately have typical symptoms and signs of right-sided heart failure.

The characteristic physical finding is a decrescendo diastolic murmur, loudest at the left third and fourth intercostal spaces, which increases with inspiration. S2 is usually widely split with an accentuated pulmonic component. There is often an associated systolic murmur from increased flow across the valve. Jugular venous distention and signs of right-sided heart failure may be apparent.

Right ventricular hypertrophy and dilatation may be evident by chest radiography and ECG. Echocardiography with Doppler can identify and grossly quantitate pulmonic regurgitation and assess the size and contractility of the right ventricle.

Treatment is generally directed at the underlying disease. If regurgitation is severe, valve surgery may be necessary. Patients should receive endocarditis prophylaxis for dental or other non-sterile procedures.

FUTURE DIRECTIONS

The treatment of pulmonic and tricuspid valve disease will continue to benefit from the steady evolution of percutaneous techniques. Pulmonic valvuloplasty was introduced in the early 1980s. Follow-up studies confirm the continued long-term effectiveness of a percutaneous approach. It is clear that most patients have a subsequent further decrease in the RV outflow gradient due in part to resolution of infundibular hypertrophy. This success has led to a generally lower threshold for intervention, as reflected in the most recent ACC/AHA guidelines discussed previously and in chapter 33. With respect to tricuspid stenosis, valvuloplasty techniques that use multiple balloons or the newer Inoue balloon appear promising.

REFERENCES


