severe tricuspid stenosis, the European Society of Cardiology rates valvuloplasty or valve replacement in symptomatic patients, or in those undergoing left-sided valve intervention, as a class I indication (level C evidence).

Severe tricuspid regurgitation is associated with increased risks of morbidity and mortality. Diuretics are useful initially to improve edema. As the disease progresses, increasing doses are required, until eventually patients become unresponsive. The improvement in their edema also comes at the expense of more fatigue and dyspnea due to limited cardiac output. Tricuspid valve repair or replacement is the only proven method of interrupting this clinical progression. Severe symptomatic tricuspid regurgitation therefore carries a class I recommendation for surgery (level C evidence).

Prognosis

There are few data on the natural history of isolated unoperated tricuspid valve stenosis. Guideline recommendations for therapy are based on expert consensus, and individual patient management is largely based on clinical judgment given the infrequency of isolated stenosis.

A study performed at the Mayo Clinic examined 60 patients with severe tricuspid regurgitation treated over a 20-year period. Patients with unoperated tricuspid regurgitation had a 4.5% per year mortality rate, significantly higher than that for a matched U.S. population, supporting the class I recommendation for surgery.

PULMONARY VALVE DISEASE

Definition

Pulmonary valve stenosis, similar to stenosis of the aortic valve, results from incomplete opening of the three valve leaflets in systole and leads to a pressure gradient between right ventricle and the pulmonary artery. The severity of the resulting obstruction to flow determines the clinical sequelae. Isolated significant pulmonary valve regurgitation is uncommon and is well tolerated unless severe.

Pathology

Pulmonary valve stenosis is rare. It is caused predominantly by congenital heart disease and typically is identified in childhood. It is very common in certain congenital disorders, such as Noonan's syndrome. Rare acquired causes of the disease include carcinoid tumor and rheumatic heart disease, but other valves are typically involved as well in these conditions.

Moderate or severe pulmonary valve regurgitation can similarly result from congenital heart disease or, even more likely, from previous mechanical treatment of congenital pulmonary valve stenosis. Valvular causes include trauma, carcinoid, and endocarditis. RV outflow tract and pulmonary artery enlargement may also result in failed central leaflet coaptation with resulting regurgitation.

Clinical Presentation

Although the diagnosis of pulmonary valve stenosis is most often made in childhood, it is occasionally first identified in an adult. The RV pressure overload of severe stenosis results most commonly in symptoms of fatigue and dyspnea. Angina, exertional lightheadedness, or syncope can occur in more advanced stages of the disease.

Pulmonary regurgitation typically has a benign course unless it becomes severe. Even after that point, the volume overload of the right ventricle may be tolerated well for many years, similar to the overload of the left ventricle in aortic regurgitation. The right ventricle eventually enlarges, with some increased risk of arrhythmia. Ultimately, there is loss of RV systolic function, and patients develop signs and symptoms of right-sided heart failure, as previously described.

Diagnosis

On physical examination, the patient with severe pulmonary stenosis may have an RV lift as a result of the RV hypertrophy that develops. A prominent *a* wave may be identifiable on inspection of jugular venous pulsations. A crescendo-decrescendo systolic murmur is best heard at the left upper sternal border and may vary with respiration. S_2 may have relative fixed slitting, with the P_2 pulmonic component becoming soft or absent as the stenosis progresses. Chest radiography may demonstrate right heart and pulmonary artery enlargement with decreased vascular markings.

Similarly, the RV enlargement of pulmonary valve regurgitation may result in an RV lift. A diastolic decrescendo murmur may be present at the left sternal border and varies with respiration, but it may be inaudible if pulmonary pressures are normal. Chest radiography demonstrates right-sided heart enlargement.

TTE is sensitive and specific for diagnosis of both stenosis and regurgitation of the pulmonary valve if actively sought on the examination. Severe pulmonary valve stenosis is considered to be present when the peak gradient is greater than 64 mm Hg. Cardiac MRI may be useful in quantitating regurgitation severity or in estimating the gradient in stenosis if echocardiography is inconclusive.

Treatment

Percutaneous balloon valvuloplasty is an effective treatment for symptomatic severe pulmonary valve stenosis (class I indication, level C evidence). Surgical valvotomy or valve replacement is used only when anatomic features prevent balloon valvuloplasty or if there has been a poor result from previous attempts.

Valve replacement for severe pulmonary valve regurgitation is thought by most to be indicated when patients become symptomatic or RV systolic dysfunction develops, although there is no consensus on recommendations for the approach to this problem.

Prognosis

Studies have suggested that survival in pulmonary stenosis is related to the pressure gradient, with decreased survival in those with gradients greater than 50 mm Hg. Therefore, balloon valvuloplasty is a class I (level C evidence) recommendation for asymptomatic patients with peak pulmonary valve gradients at catheterization of at least 40 mm Hg and a class IIb recommendation for asymptomatic patients with a gradient of 30 to 39 mm Hg (level C evidence).