indicated in adults with critical obstruction, i.e., with an aortic valve area <0.45 cm^2/m^2, with symptoms secondary to LV dysfunction or myocardial ischemia, or with hemodynamic evidence of LV dysfunction. In asymptomatic children or adolescents or young adults with critical aortic stenosis without valvular calcification or these features, aortic balloon valvuloplasty is often useful **(Chap. 296e)**. If surgery is contraindicated in older patients because of a complicating medical problem such as malignancy or renal or hepatic failure, balloon valvuloplasty may provide short-term improvement. This procedure may serve as a bridge to aortic valve replacement in patients with severe heart failure. Transcatheter aortic valve replacement is a potential alternative to surgery.

SUBAORTIC STENOSIS The discrete form of subaortic stenosis consists of a membranous diaphragm or fibromuscular ring encircling the LV outflow tract just beneath the base of the aortic valve. The jet impact from the subaortic stenotic jet on the underside of the aortic valve often begets progressive aortic valve fibrosis and valvular regurgitation. Echocardiography demonstrates the anatomy of the subaortic obstruction; Doppler studies show turbulence proximal to the aortic valve and can quantitate the pressure gradient and severity of aortic regurgitation. Treatment consists of complete excision of the membrane or fibromuscular ring.

SUPRAVALVULAR AORTIC STENOSIS This is a localized or diffuse narrowing of the ascending aorta originating just above the level of the coronary arteries at the superior margin of the sinuses of Valsalva. In contrast to other forms of aortic stenosis, the coronary arteries are subjected to elevated systolic pressures from the LV, are often dilated and tortuous, and are susceptible to premature atherosclerosis. The coronary ostia may also become obstructed by the aortic valve leaflets. In most patients, a genetic defect for the anomaly is located in the same chromosomal region as elastin on chromosome 7. Supravalvular aortic stenosis is the most commonly associated cardiac defect in Williams-Beuren syndrome, typically comprising the following: "elfin" facies, low nasal bridge, cheerful demeanor, mental retardation with retained language skills and love of music, supravalvular aortic stenosis, and transient hypercalcemia.

COARCTATION OF THE AORTA

Narrowing or constriction of the lumen of the aorta may occur anywhere along its length but is most common distal to the origin of the left subclavian artery near the insertion of the ligamentum arteriosum. Coarctation occurs in ~7% of patients with CHD, is more common in males than females, and is particularly frequent in patients with gonadal dysgenesis (e.g., Turner's syndrome). Clinical manifestations depend on the site and extent of obstruction and the presence of associated cardiac anomalies, most commonly a bicuspid aortic valve. Circle of Willis aneurysms may occur in up to 10%.

Most children and young adults with isolated, discrete coarctation are asymptomatic. Headache, epistaxis, chest pressure, and claudication with exercise may occur, and attention is usually directed to the cardiovascular system when a heart murmur or hypertension in the upper extremities and absence, marked diminution, or delayed pulsations in the femoral arteries are detected on physical examination. Enlarged and pulsatile collateral vessels may be palpated in the intercostal spaces anteriorly, in the axillae, or posteriorly in the interscapular area. The upper extremities and thorax may be more developed than the lower extremities. A midsystolic murmur over the left interscapular space may become continuous if the lumen is narrowed sufficiently to result in a high-velocity jet across the lesion throughout the cardiac cycle. Additional systolic and continuous murmurs over the lateral thoracic wall may reflect increased flow through dilated and tortuous collateral vessels. The ECG usually reveals LV hypertrophy. Chest x-ray may show a dilated left subclavian artery high on the left mediastinal border and a dilated ascending aorta. Indentation of the aorta at the site of coarctation and pre- and poststenotic dilatation (the "3" sign) along the left paramediastinal shadow are essentially pathognomonic. Notching of the third to ninth ribs, an important radiographic sign, is due to inferior rib erosion by dilated collateral vessels. Two-dimensional echocardiography from suprasternal windows identifies the site of coarctation; Doppler quantitates the pressure gradient. Transesophageal echocardiography and MRI or CT allow visualization of the length and severity of the obstruction and associated collateral arteries. In adults, cardiac catheterization is indicated primarily to evaluate the coronary arteries or to perform catheter-based intervention (angioplasty and stent of the coarctation).

The chief hazards of proximal aortic severe hypertension include cerebral aneurysms and hemorrhage, aortic dissection and rupture, premature coronary arteriosclerosis, aortic valve failure, and LV failure; infective endarteritis may occur on the coarctation site or endocarditis may settle on an associated bicuspid aortic valve, which is estimated to be present in 50% of patients.

TREATMENT COARCTATION OF THE AORTA

Treatment is surgical or involves percutaneous catheter balloon dilatation with stent placement; the details of selection of therapy are beyond this review; however, the use of transcatheter treatment techniques has increased dramatically, and many previously "surgical" cases are treated via percutaneous or hybrid techniques. Late postoperative systemic hypertension in the absence of residual coarctation is related partly to the duration of preoperative hypertension. Follow-up of rest and exercise blood pressures is important; many have systolic hypertension only during exercise, in part due to a diffuse vasculopathy and to noncompliance of the stented or surgically reconstructed region. All operated or stented coarctation patients deserve a high-quality MRI or CT procedure in follow-up.

PULMONARY STENOSIS WITH INTACT VENTRICULAR SEPTUM

Obstruction to RV outflow may be localized to the supravalvular, valvular, or subvalvular levels or occur at a combination of these sites. Multiple sites of narrowing of the peripheral pulmonary arteries are a feature of rubella embryopathy and may occur with both the familial and sporadic forms of supravalvular aortic stenosis. Valvular pulmonic stenosis (PS) is the most common form of isolated RV obstruction.

The severity of the obstructing lesion, rather than the site of narrowing, is the most important determinant of the clinical course. In the presence of a normal cardiac output, a peak systolic pressure gradient <30 mmHg indicates mild PS and >50 mmHg indicates severe PS; pressures between these limits are considered to indicate moderate stenosis. Patients with mild PS are generally asymptomatic and demonstrate little or no progression in the severity of obstruction with age. In patients with more significant stenosis, the severity may increase with time. Symptoms vary with the degree of obstruction. Fatigue, dyspnea, RV failure, and syncope may limit the activity of older patients, in whom moderate or severe obstruction may prevent an augmentation of cardiac output with exercise. In patients with severe obstruction, the systolic pressure in the RV may exceed that in the LV, because the ventricular septum is intact. RV ejection is prolonged with moderate or severe stenosis, and the sound of pulmonary valve closure is delayed and soft. RV hypertrophy reduces the compliance of that chamber, and a forceful RA contraction is necessary to augment RV filling. A fourth heart sound; prominent a waves in the jugular venous pulse; and, occasionally, presystolic pulsations of the liver reflect vigorous atrial contraction. The clinical diagnosis is supported by a left parasternal lift and harsh systolic crescendo-decrescendo murmur and thrill at the upper left sternal border, typically preceded by a systolic ejection sound if the obstruction is due to a mobile nondysplastic pulmonary valve. The holosystolic murmur of tricuspid regurgitation may accompany severe PS, especially in the presence of congestive heart failure. Cyanosis usually reflects right-to-left shunting through a patent foramen ovale or ASD. In patients with supravalvular or peripheral pulmonary arterial stenosis, the murmur is systolic or continuous and is best heard over the area of narrowing, with radiation to the peripheral lung fields.