



FIGURE 381-2 Transthoracic echocardiographic image from a 5-year-old boy with chronic rheumatic heart disease. This diastolic image demonstrates leaflet thickening, restriction of the anterior mitral valve leaflet tip and doming of the body of the leaflet toward the interventricular septum. This appearance (marked by the *arrowhead*) is commonly described as a “hockey stick” or an “elbow” deformity. AV, aortic valve; LA, left atrium; LV, left ventricle; MV, mitral valve; RV, right ventricle. (Courtesy of Dr. Bo Remenyi, Department of Paediatric and Congenital Cardiac Services, Starship Children’s Hospital, Auckland, New Zealand.)

affected, sometimes together with the aortic valve; isolated aortic valve involvement is rare. Damage to the pulmonary or tricuspid valves is usually secondary to increased pulmonary pressures resulting from left-sided valvular disease. Early valvular damage leads to regurgitation. Over ensuing years, usually as a result of recurrent episodes, leaflet thickening, scarring, calcification, and valvular stenosis may develop (Fig. 381-2). See Videos 381-1 and 381-2 on the DVD. Therefore, the characteristic manifestation of carditis in previously unaffected individuals is mitral regurgitation, sometimes accompanied by aortic regurgitation. Myocardial inflammation may affect electrical conduction pathways, leading to P-R interval prolongation (first-degree atrioventricular block or rarely higher level block) and softening of the first heart sound.

People with RHD are often asymptomatic for many years before their valvular disease progresses to cause cardiac failure. Moreover, particularly in resource-poor settings, the diagnosis of ARF is often not made, so children, adolescents, and young adults may have RHD but not know it. These cases can be diagnosed using echocardiography; auscultation is poorly sensitive and specific for RHD diagnosis in asymptomatic patients. Echocardiographic screening of school-aged children in populations with high rates of RHD is becoming more widespread and has been facilitated by improving technologies in portable echocardiography and the availability of consensus guidelines for the diagnosis of RHD on echocardiography (Table 381-1). Although a diagnosis of definite RHD on screening echocardiography should lead to commencement of secondary prophylaxis, the clinical significance of borderline RHD has yet to be determined.

JOINT INVOLVEMENT

The most common form of joint involvement in ARF is arthritis, i.e., objective evidence of inflammation, with hot, swollen, red, and/or tender joints, and involvement of more than one joint (i.e., polyarthritis). Polyarthritis is typically migratory, moving from one joint to another over a period of hours. ARF almost always affects the large joints—most commonly the knees, ankles, hips, and elbows—and is asymmetric. The pain is severe and usually disabling until anti-inflammatory medication is commenced.

TABLE 381-1 WORLD HEART FEDERATION CRITERIA FOR ECHOCARDIOGRAPHIC DIAGNOSIS OF RHEUMATIC HEART DISEASE (RHD) IN INDIVIDUALS <20 YEARS OF AGE*

Definite RHD (either A, B, C, or D):
(A) Pathologic MR and at least two morphologic features of RHD of the mitral valve
(B) MS mean gradient ≥ 4 mmHg (note: congenital MV anomalies must be excluded)
(C) Pathologic AR and at least two morphologic features of RHD of the AV (note: bicuspid AV and dilated aortic root must be excluded)
(D) Borderline disease of both the MV and AV
Borderline RHD (either A, B, or C):
(A) At least two morphologic features of RHD of the MV without pathologic MR or MS
(B) Pathologic MR
(C) Pathologic AR
Normal Echocardiographic Findings (all of A, B, C, and D):
(A) MR that does not meet all four Doppler criteria (physiologic MR)
(B) AR that does not meet all four Doppler criteria (physiologic AR)
(C) An isolated morphologic feature of RHD of the MV (e.g., valvular thickening), without any associated pathologic stenosis or regurgitation
(D) Morphologic feature of RHD of the AV (e.g., valvular thickening), without any associated pathologic stenosis or regurgitation
Definitions of Pathologic Regurgitation and Morphologic Features of RHD:
Pathologic MR: All of the following: seen in two views; in at least one view, jet length 2 cm; peak velocity ≥ 3 m/s; pansystolic jet in at least one envelope
Pathologic AR: All of the following: seen in two views; in at least one view, jet length ≥ 1 cm; peak velocity ≥ 3 m/s; pandsystolic jet in at least one envelope
Morphologic features of RHD in MV: anterior MV leaflet thickening ≥ 3 mm (age specific); chordal thickening; restricted leaflet motion; excessive leaflet tip motion during systole
Morphologic features of RHD in AV: irregular or focal thickening; coaptation defect; restricted leaflet motion; prolapse

*For criteria in individuals >20 years of age, see source document.

Abbreviations: AR, aortic regurgitation; AV, aortic valve; MR, mitral regurgitation; MS, mitral stenosis; MV, mitral valve.

Source: Adapted from Remenyi B et al: World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease—an evidence-based guideline. *Nat Rev Cardiol* 9:297–309, 2012.

Less severe joint involvement is also relatively common and has been recognized as a potential major manifestation in high-risk populations in diagnostic guidelines from Australia, but at the time of writing, this was not reflected in the Jones criteria. Arthralgia without objective joint inflammation usually affects large joints in the same migratory pattern as polyarthritis. In some populations, aseptic monoarthritis may be a presenting feature of ARF, which may, in turn, result from early commencement of anti-inflammatory medication before the typical migratory pattern is established.

The joint manifestations of ARF are highly responsive to salicylates and other nonsteroidal anti-inflammatory drugs (NSAIDs). Indeed, joint involvement that persists for more than 1 or 2 days after starting salicylates is unlikely to be due to ARF.

CHOREA

Sydenham’s chorea commonly occurs in the absence of other manifestations, follows a prolonged latent period after group A streptococcal infection, and is found mainly in females. The choreiform movements affect particularly the head (causing characteristic darting movements of the tongue) and the upper limbs (Chap. 448). They may be generalized or restricted to one side of the body (hemi-chorea). In mild cases, chorea may be evident only on careful examination, whereas in the most severe cases, the affected individuals are unable to perform activities of daily living. There is often associated emotional lability or obsessive-compulsive traits,