Diagnostic Criteria in Rheumatic Heart Disease

Over a decade of echocardiography-based studies have reinforced the perception that rheumatic heart disease (RHD) is not an ignorable health problem in the developing countries [1]. Socioeconomic improvement is considered the most important determinant leading to decrease in the prevalence of RHD. Differences in streptococcal infection and carriage rates across different populations have been best explained as microbiological correlates of socioeconomic improvement.

RHD is a result of autoimmunity triggered by recurrent streptococcal infections. Passive detection of RHD is with low yield because such cases are often asymptomatic and do not seek medical attention. However, active case detection, by universal echocardiographic screening of a population (e.g., school children), even if attempted, is potentially limited by nonexclusivity of echocardiographic features such as leaflet thickening and significant regurgitation of mitral and/or aortic valve due to other etiologies such as myxomatous degeneration, thyrotoxicosis, infective endocarditis, to name a few [2,3].

We attempted to identify the entire spectrum of RHD in a northwestern Indian population in a 3-stage study. In the first stage, 2-dimensional transthoracic echocardiography (2DTTE) was used for universal screening ($n = 1,059$) of asymptomatic school children. Significant regurgitation of mitral and/or aortic valves was taken as the sole criterion for the diagnosis of RHD (after excluding other causes of significant valvular regurgitation such as congenital heart defects or mitral valve prolapse), and 54 children thus identified with RHD were placed on penicillin prophylaxis [3]. In the second stage, conducted 2 years later, these 54 children along with 100 children declared normal by echocardiography were reassessed by 2DTTE. A notable finding of this second stage of study was the high negative predictive value of the normal echocardiography study in the first stage of the study [4]. In the third stage, 2DTTE and real-time 3-dimensional transthoracic echocardiography (3DTTE) were used to evaluate a new, independent set of 31 symptomatic cases in a tertiary care hospital in order to track the spectrum of symptomatic RHD using specific cutoffs as mentioned by World Heart Federation (WHF) criteria for the diagnosis of "borderline/definite RHD" [1].

After the second stage of study, it was evident that the problem of borderline RHD defies opportunistic screening protocols based on passive case detection using specific diagnostic criteria. Thus, we inferred that significant regurgitation of mitral and/or aortic valves forms an essential pre-requisite for identification of borderline RHD. The problem of borderline RHD could be appreciated better as a continuum ranging from earliest evidence such as a 1- to 2-cm regurgitant jet beyond the mitral or aortic valve, which is later accompanied by leaflet thickening. Co-existing mitral and aortic regurgitation, severe regurgitation of the mitral valve with characteristic morphologic abnormalities such as valve leaflet thickening, subvalvular thickening, leaflet prolapse, and mitral stenosis, with or without mitral regurgitation, occur late in the natural history of the disease process when the diagnosis of definite RHD is not in doubt.

We were able to identify a couple potential limitations of 2DTTE in detecting some echocardiographic features potentially diagnostic of borderline and definite RHD:

1. Real-time 3DTTE shows that the thickening of the anterior mitral leaflet is irregular across its span (Figure 1A). This can lead to a false negative result by 2DTTE, which images a random thin section of an irregularly thickened leaflet. This finding is of importance as WHF criteria mandate the presence of morphologic valve abnormalities for diagnosis of definite RHD.
2. Real-time 3DTTE shows that prolapse of the anterior mitral leaflet is pathognomonic of RHD and is characterized by the following features: (a) normally situated coaptation line in the left ventricle (Figure 1B); (b) absence of leaflet redundancy); and (c) absence of chordal lengthening. These features differentiate it from prolapse due to myxomatous degeneration.

We employed a quantitative diagnostic score for borderline RHD and a consensus cutoff for definite RHD including some additional echocardiographic features (Table 1). A cumulative score of ≥ 2 enables diagnosis of "definite RHD" and a score of 1 to <2 identifies "borderline RHD."

LIMITATIONS

Real-time 3DTTE assessment of length and thickness of the subvalvular mitral apparatus is predominantly qualitative. Although it forms a valuable criterion to differentiate myxomatous and rheumatic pathologies, it would be difficult to use this criterion to screen for RHD.

By the surgeon's view of anterior mitral leaflet, irregular thickening across the span leading to poor apposition appears to be the mechanism of regurgitation for RHD cases. However, the role of leaflet retraction due to fibrosis, which could hypothetically lead to regurgitation, could not be appreciated.

Measurement of leaflet thickness by real-time 3DTTE appears different from what is visualized by 2DTTE. So, use of leaflet thickness as a quantitative criterion for diagnosis of borderline RHD is questionable.

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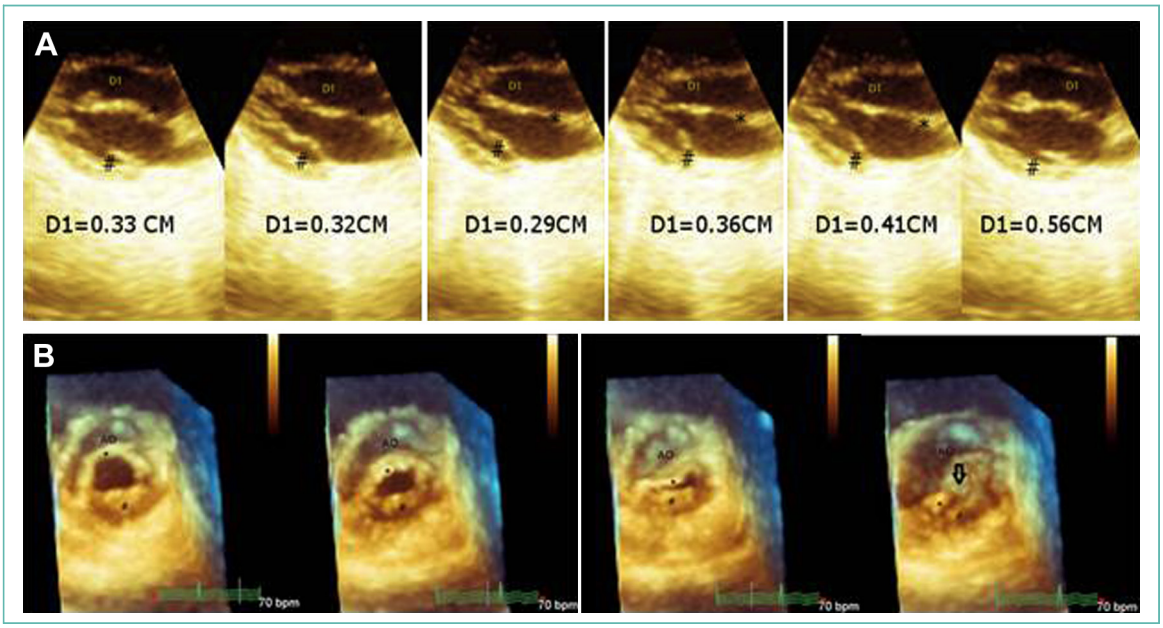

FIGURE 1. (A) Six parallel thin slices mimicking parasternal long-axis view by 2-dimensional transthoracic echocardiography showing variation in the maximum thickness of the anterior mitral leaflet measured by the multiplanar resetting mode of live/real-time 3-dimensional transthoracic echocardiography. *The anterior mitral leaflet; #the posterior mitral leaflet. (B) The en-face view of the mitral valve from the left ventricular side by live/real-time transthoracic echocardiography seen in various stages of cardiac cycle. The *arrow* depicts the "reverse doming"/prolapse of the middle (A2) and lateral (A1) scallops of the anterior mitral leaflet as seen from the ventricular side (Video 1) and left atrial side (Video 2). *The anterior mitral leaflet; #the posterior mitral leaflet.

TABLE 1. Diagnostic score for "definite" and "borderline" rheumatic heart disease

	Score
Functional deficits	
Pathological MR	1
Pathologic AR	0.5
Mitral stenosis with mean gradient during diastole ≥4 mm Hg	2
Morphological abnormalities	
Thickening of AML and/or PML	0.5
Thickened chordae	0.5
Restricted movement of AML and/or PML	0.5
Excessive leaflet tip movement of AML and/or PML	0.5
Doming of AML during diastole and/or reverse doming (prolapse) in systole	0.5/0.5
Aortic valve leaflet thickening	0.25
Prolapse of aortic leaflets	0.25
Restricted movement of aortic leaflets	0.25

AML, anterior mitral leaflet; AR, aortic regurgitation; MR, mitral regurgitation; PML, posterior mitral leaflet.

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