WHAT'S IMPORTANT TO KEEP IN MIND WHEN VALVES GO NARROWED AND LEAKY?

Insights from the ASE Recommendations for Rheumatic Heart Disease

Contributed by

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This article serves as a practical primer to the recently published "Recommendations for the Use of Echocardiography in the Evaluation of Rheumatic Heart Disease."[1] It summarizes key points presented in this document and aims to support active clinical translation of these guidelines by emphasizing practical aspects pertaining to the evaluation of patients with rheumatic valve disorders.

Back to the Basics: What is ARF & RHD?

Acute rheumatic fever (ARF) is the consequence of an immune-mediated response to pharyngitis caused by Streptococcus pyogenes. Prolonged or multiple recurrent episodes of the illness results in long-term damage to the heart and cardiac valves, referred to as rheumatic heart disease (RHD). While RHD remains the single most common cardiovascular disorder among children and young adults in resource-constrained global settings, it has evolved to impose significant worldwide burden with widespread emigration in recent years.

Why are Recommendations for Echocardiographic Evaluation of RHD necessary?

Echocardiography plays a key role for accurate detection of rheumatic valvulitis and is extensively utilized in worldwide screening programs to assess RHD prevalence. While minimum Doppler echocardiographic diagnostic criteria have been published previously,[2] a guide for comprehensive non-invasive evaluation of rheumatic cardiac lesions is needed. The aim of the recently published recommendations is to provide clinicians and sonographers with guidelines for the use of echocardiography in screening, diagnosis, classification, and risk assessment of RHD.

Let's Get Technical! Instrumentation and Image Optimization

The assessment of a rheumatic valve entails optimization of equipment settings to provide high-resolution diagnostic images. Tissue harmonics should be turned off to avoid overestimating valve thickness, gain settings and dynamic range are to be adjusted to enhance leaflet border delineation, and focus set at the level of the valve being interrogated. A magnified view of the valve is always recommended. Heart rate, rhythm, and blood pressure should be recorded prior to each study. Multiple views of the valve are to be captured for reliable jet characterization. In the setting of atrial fibrillation (AF), at least 5 representative cycles are to be taken into consideration during measurements.

When 3D echocardiography (3DE) is employed, spatial and temporal resolution should be optimized, gain settings set to around 50 dB to avoid image dropout, and compression balanced for detailed valve characterization. Supine bicycle is the preferred stress modality when hemodynamic alterations are studied under exercise. However, if treadmill stress is utilized, images are to be acquired at baseline and within two minutes post exercise. Although Dobutamine stress is less physiological, low-dose Dobutamine may be employed in symptomatic patients when exercise is not feasible.

It's getting hot in here! Screening in Acute Rheumatic Fever

Echocardiographic evidence of valvulitis is a major criterion in the diagnosis of subclinical carditis. Careful inspection of mitral and aortic valve morphology in addition to identification of pathological valvular regurgitation is integral to assessment. Changes in valve morphology may be absent during early ARF presentation. When present, thickening of the free edges of the valve along with nodularity along the leaflet length can be seen. Mitral valve thickness <3 mm in children and <3.5 mm in adults is considered normal. In the absence of non-rheumatic causes, greater than trace MR or AR may be considered pathological if a pansystolic (in MR) or pandiastolic (in AR) jet is seen in multiple views using CW Doppler with peak velocity > 3 m/sec in at-risk populations.

Rigid and Stiff! Evaluation of Rheumatic Mitral Stenosis

A comprehensive approach to the evaluation of mitral stenosis includes a) inspection of mitral cuspal, chordal and commissural morphology, b) accurate determination of mitral valve area (MVA) at the leaflet tips, c) Doppler assessment of pressure half-time, mean mitral gradient and associated pulmonary hypertension, and d) evaluation of associated findings such as left atrial enlargement, presence of thrombi, right ventricular size and function.

Anatomical considerations in the assessment of mitral stenosis have important therapeutic implications on choice of percutaneous or surgical intervention. Diastolic doming of the anterior leaflet is best appreciated in the parasternal long-axis view, and the 'fish mouth' appearance characteristic of commissural fusion leading to a narrowed mitral orifice in the short-axis view. Chordal thickening, shortening and fusion are best appreciated in parasternal and apical views. Planimetry is the preferred method for determination of anatomic MVA. Using 2D echocardiography (2DE), overestimation of MVA can be avoided by scanning carefully from the sub-valvular plane in the short axis view to the level of the mitral leaflet tips. MVA is assessed using a magnified view and gain settings should be optimized to avoid signal dropout (low gain) or MVA underestimation (high gain). 3D echocardiography is more accurate and reproducible than 2DE, offers stronger commissural definition in addition to perspectives from both left atrium (LA) and the left ventricle (LV) (Figure 1).

Hemodynamic assessment includes estimation of MVA using the pressure half-time (PHT) method, evaluation of transmitral pressure gradients and corresponding pulmonary pressures. Cardiac rhythm and heart rate should be documented as part of Doppler assessment. Transmitral gradient

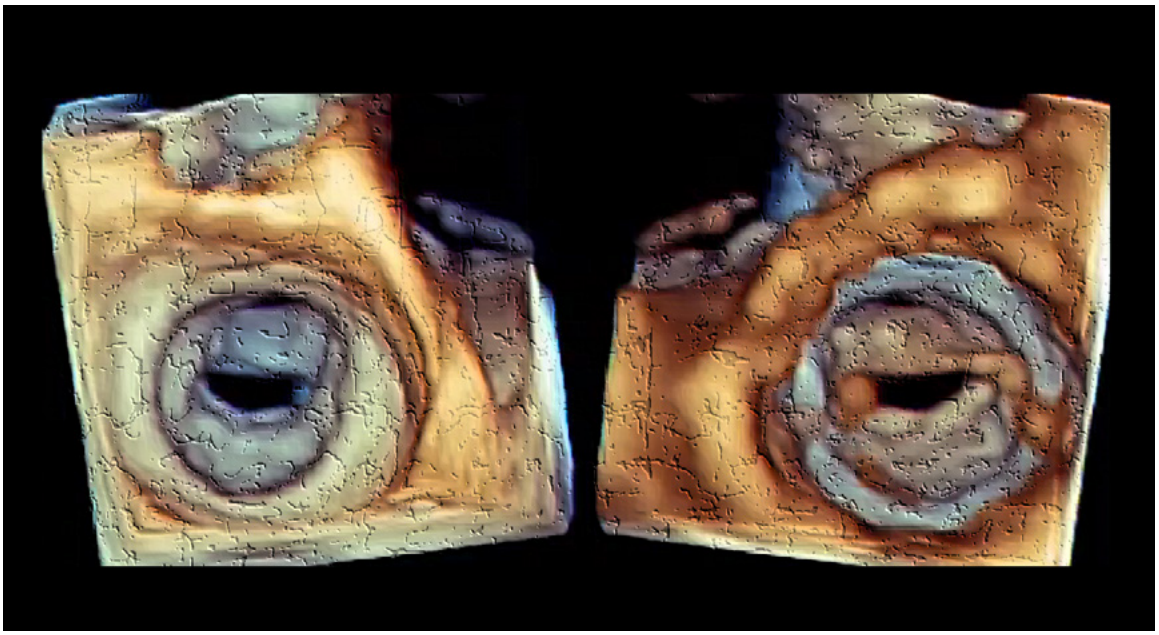

FIGURE 1. Mitral stenosis visualized using 3D real-time imaging providing simultaneous LA (left) and LV (right) perspective

is measured using CW Doppler, aligning the signal coaxially with mitral inflow. In the setting of a bimodal spectral flow pattern, deceleration time should be assessed considering the mid-diastolic slope rather than the early steep decline. Rheumatic MS is defined by a transmitral mean gradient > 4 mmHg in the setting of morphology suggestive of rheumatic aetiology. Severe MS is defined by MVA ≤ 1.5 cm^2, PHT ≥ 150 msec and transmitral mean gradient ≥ 10mmHg.

Doppler-based assessments are, however, not reliable as sole estimates of MS severity. High output states and significant mitral regurgitation result in a disproportionate rise in flow velocity. Reductions in left ventricular compliance in the setting of LV hypertrophy or with concomitant aortic regurgitation (AR) result in a lowered transmitral gradient. When imaging findings and patient symptoms are not congruent, exercise stress testing may help identify patients that benefit from intervention. A transmitral mean gradient > 15 mmHg with exercise, ≥ 18 mmHg with dobutamine infusion, or pulmonary artery systolic pressure > 60 mmHg may be considered as hemodynamically significant.

Achy, Leaky Heart! Evaluation of Rheumatic Mitral Regurgitation

The multi-parametric approach to assessing rheumatic MR includes both semi-quantitative and quantitative evaluation of regurgitant flow as per current recommendations. Anatomic considerations include establishing at least two morphological features of rheumatic MV, classifying MR as primary (attributable to valve abnormality) or secondary (attributable to LV/LA remodelling), and jet orientation as central or eccentric. 3DE with real-time volumetric imaging or when combined with color Doppler provides stronger characterization of the jet origin, extent and trajectory. Quantifiable measures of MR severity are recommended whenever feasible, particularly when jet severity is estimated as more than moderate. Semi-quantitative evaluation of MR severity by vena contracta (VC) using 2DE, or vena contracta area using 3DE are relevant in the setting of eccentric regurgitant jets common to RHD. Quantitative indices include effective orifice regurgitant area (EROA) by PISA, regurgitant volume and regurgitant fraction. Severe rheumatic MR is indicated by a VC ≥ 0.7 cm, VC area ≥ 0.40 cm^2, EROA ≥ 0.4 cm^2, regurgitant volume ≥ 60 ml.

Evaluation of the Rheumatic Aortic Valve

The aortic valve should be evaluated using the Zoom function in the parasternal long- and short-axis views, paying attention to commissural fusion, fibrotic thickening and retraction of the leaflet edges suggestive of rheumatic etiology. Superimposed calcification during early stages generally starts from the leaflet free edges rather than the base. Multiplanar and real-time 3D imaging are useful for detailed characterization of the valve en-face.

When evaluating AS, a multiparametric approach is recommended taking into consideration anatomic aortic valve area (AVA) obtained using planimetery, effective AVA employing the continuity relationship, AVA indexed to BSA, mean pressure gradient, transvalvular peak velocity, Doppler velocity index (DVI) and acceleration time. When employing the continuity equation, 3D multiplanar imaging offers a more accurate measurement of cross-sectional area of the left ventricular outflow tract (LVOT), which may be underestimated using 2DE. Other potential sources of measurement error include malposition of the PW Doppler sample volume, malignment of the CW Doppler signal and subsequent misrepresentation of peak aortic velocity. Blood pressure, BSA, high and low flow states should always be considered. Severe AS is suspected if transaortic flow Vmax ≥ 40 mmHg, AVA < 1 cm^2, indexed AVA < 0.6 cm^2/m^2, and DVI < 0.25.

Assessment of the severity of rheumatic AR takes into consideration qualitative, semi-quantitative and quantitative Doppler measures in addition to LV size and function in keeping with current recommendations. Severe AR is indicated by a dense CW Doppler spectrum, PHT < 200 msec, VC > 0.6 cm, jet width/LVOT width ≥ 65%, regurgitant volume ≥ 60 ml, regurgitation fraction ≥ 50% and EROA ≥ 0.3 cm^2. Flow reversal in the descending aorta and LV enlargement are supportive findings.

Evaluation of the Rheumatic Tricuspid Valve

Tricuspid stenosis (TS) is most often associated with rheumatic mitral or aortic valve disorders. Anatomical characteristics include leaflet thickening, doming and subvalvular shortening seen best in the RV-focused apical view in addition to assessment of right atrial (RA) size and to rule out thrombi. 3DE is useful

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to characterize commissural fusion in real-time and to measure tricuspid valve area (TVA) using multiplane imaging. Hemodynamic assessment includes evaluation of mean TV pressure gradient, TVA and associated RA and pulmonary artery (PA) pressure. TS is hemodynamically significant if mean gradient ≥ 5 mmHg, inflow VTI ≥ 60 cm, PHT ≥ 190 ms and TVA ≤ 1 cm^2. Rheumatic tricuspid regurgitation (TR) is classified as primary if attributable to valvulitis, or secondary due to right heart enlargement attributable to left-sided valvular disease. Primary TR is characterized by thickening and doming of the tricuspid valve, in the presence or absence of stenosis. Severe TR is associated with a color Doppler regurgitant area > 10 cm^2 at a Nyquist limit > 50 cm/sec, VC ≥ 0.7 cm in the apical 4-chamber view, VC area > 0.4 cm^2, PISA radius ≥ 0.9 cm, EROA ≥ 0.4 cm^2 and regurgitant volume ≥ 45 ml. Supportive findings include flow reversal in a hepatic vein, tricuspid inflow E-wave velocity > 1.0 cm and a dense spectral regurgitant waveform. In the setting of rheumatic MR and AR, TR may be accentuated owing to elevated pulmonary venous pressure. Both anatomic and hemodynamic considerations are to be integrated to arrive at an accurate diagnosis.

Evaluation of the Rheumatic Pulmonic Valve

Rheumatic pulmonary valve (PV) stenosis is uncommon and always associated with mitral or aortic valve disease. Pulmonary stenosis (PS) is best appreciated in the parasternal short axis view with limited leaflet mobility, thickening and doming suggestive of a narrowed orifice. 3DE offers additional value in capturing an en-face view of all 3 leaflets. Severe PS is suspected when peak velocity > 4 m/sec and mean gradient > 35 mmHg.

Significant isolated pulmonary regurgitation (PR) has not been reported in large RHD cohorts. Severe PR is identified as having a jet width/annular diameter ≥ 70%, PHT < 100 msec and deceleration time < 260 msec in keeping with published recommendations.

Assessment of Secondary Pulmonary Hypertension

Pulmonary hypertension is common in the setting of rheumatic mitral valve disorders and left ventricular dysfunction associated with aortic valve disease. Echocardiographic evaluation of PA systolic pressure in addition to right heart size and function should be included during assessment. The dense, well-defined spectral envelope that reflects modal velocities is to be considered when measuring TR gradient. Extra systolic beats are to be avoided. RA pressure is estimated using IVC size and collapsibility in keeping with current recommendations.

Echocardiographic Guidance for Percutaneous and Surgical Intervention

Selection for percutaneous balloon mitral valvuloplasty (PBMV) includes assessment of degree of thickening and calcification of leaflets, extent of commissural fusion, severity of subvalvular disease, grade of MR and the presence/absence of thrombi. Immediately post procedure, transmitral gradient and MVA should be measured using planimetry. The PHT method is not recommended within 24 hours of PBMV owing to acute alterations to left heart chamber compliance. Additionally, complications such as an overtly large iatrogenic septal defect, significant MR and tamponade should be ruled out.

In summary, comprehensive echocardiographic evaluation of rheumatic heart disease is invaluable for accurate diagnosis, risk assessment and therapy selection. Sole reliance on isolated measurements is strictly not advisable and a multi-parametric approach including both qualitative and quantitative measures is recommended. Finally, an echocardiographic assessment is incomplete in the absence of a larger clinical context that includes medical history, symptoms, physical evaluation and vital signs.

REFERENCES

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