# Screening for asymptomatic rheumatic heart disease: Understanding the mechanisms key to the diagnostic criteria

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## ABSTRACT

**This focussed review describes important problems experienced in the world of echocardiographic screening for asymptomatic rheumatic heart disease (RHD). It offers a critical appraisal of the screening criteria and their application and explores some of the fundamental principles underpinning the shortcomings of individual criteria. The author illustrates important mechanisms that underlie the morphological changes seen in RHD that must be accounted for if these criteria are to be rationalised and improved upon.** SAHeart 2015;12:134-144

The burden of rheumatic heart disease (RHD) remains unacceptably high in the third world, including Sub-Saharan Africa.[1-4] Screening for asymptomatic RHD (with a view to instituting secondary prophylaxis in affected cases) has received much press and is being actively studied as a potential strategy for tackling the RHD problem. This strategy relies entirely on case detection and echocardiography has been identified as a superior modality (compared to clinical screening with auscultation) to deliver these cases.[5-15] However, this strategy has its problems. This focused review will critically evaluate the shortcomings of the current criteria used for the screening of asymptomatic RHD. The review provides an in depth look at the mechanisms underpinning these shortcomings in order to find potential solutions.

Significant variation in the criteria used in the echocardiographic screening studies quoted above has made it difficult to compare studies directly. In an effort to address this, the World Heart Federation (WHF) published a set of guidelines that has been widely adopted as the diagnostic criteria of choice for diagnosing subclinical or asymptomatic RHD.[16] The WHF criteria are a marked improvement on the original WHO Doppler based criteria for specificity. The latter were derived from criteria designed to diagnose acute rheumatic carditis during episodes of acute rheumatic fever (ARF) by differentiating functional (normal spectrum) from pathological regurgitation. This understandably ignored morphological valvular changes that characterise more chronic rheumatic cardiac involvement outside that of the acute episodes.[17-19] Marijon, et al. exposed the lack of sensitivity and specificity of these criteria, during screening for subclinical disease, by adding important morphological criteria of chronic rheumatic valve involvement and degrading the importance of differentiating functional from pathological valvular regurgitation, although not removing this functional requirement completely (Marijon Combined Criteria).[19] The modified WHO criteria, and new WHF criteria, are broadly similar in terms of requiring both a significant functional and morphological deficit in order to make a definite diagnosis of RHD echocardiographically when dealing with the most common lesion, mitral regurgitation.[16,20] The WHF criteria have raised the bar in terms of diagnostic requirements for RHD with the aim of improving specificity of these criteria. Concern has however now been raised that this is increasing specificity at the expense of sensitivity which is particularly problematic from a screening perspective.[14,21] Further concerns relate to the complexity of these criteria as screening criteria to be used in the field by moderately skilled personnel. This is an important goal if population based screening is to be implemented on a large scale (Table A). It remains very difficult, even for experienced echocardiography operators, to differentiate some cases of mild rheumatic cardiac involvement from the normal spectrum or alternative pathologies. This complexity, as well as differences in the application of the guidelines, technical echocardiography pitfalls and fundamental concerns about some of the screening criteria and methodology all have the potential to lead to non-uniformity in assessment and degradation of screening accuracy. This must be kept in mind when comparing different studies, even if identical criteria have been used.

The WHF criteria broadly classify rheumatic involvement into "definite RHD" and "borderline RHD" categories. In principle, for the majority of cases, they rely on the identification of both significant morphological and functional deficits, typical of rheumatic involvement, to diagnose a case as "definite RHD".

**TABLE A: Screening criteria for asymptomatic RHD**

**A: World Heart Federation (WHF) Guideline 2012 for echocardiographic diagnosis of Rheumatic heart disease (RHD) in individuals younger than 20 years, abridged[16]**

**Diagnostic requirements for Definite RHD (either A, B, C, or D)**

A: Pathological MR and at least 2 morphological features of RHD of the MV

B: MS mean gradient ≥4mmHg

C: Pathological AR and at least 2 morphological features of RHD of the AV

D: Borderline disease of both the AV and MV

**Diagnostic requirements for Borderline RHD (either A, B, or C)**

A: At least 2 morphological features of RHD of the MV without pathological MR or MS

B: Pathological MR

C: Pathological AR

**B: WHF guideline 2012: Echocardiographic criteria for pathological regurgitation**

**Diagnostic requirements for pathological mitral regurgitation (MR)**

(All 4 Doppler echocardiographic criteria must be met)

- Seen in 2 views
- In at least 1 view, jet length ≥2cm
- Velocity ≥3m/s for 1 complete envelope
- Pan-systolic jet in at least 1 envelope

**Diagnostic requirements for pathological aortic regurgitation (AR)**

(All 4 Doppler echocardiographic criteria must be met)

- Seen in 2 views
- In at least 1 view, jet length ≥1cm
- Velocity ≥3m/s in early diastole
- Pan-diastolic jet in at least one envelope

**C: WHF Guideline 2012: Morphological features of RHD on echocardiography**

**Features in the MV**

- AMVL thickening ≥3mm (≥4mm if aged over 20yrs, ≥5mm if aged over 40yrs)
- Chordal thickening
- Restricted leaflet motion
- Excessive leaflet tip motion during systole

**Features in the AV**

- Irregular or focal thickening
- Coaptation defect
- Restricted leaflet motion
- Prolapse

Cases with either a typical morphological or functional abnormality (principally regurgitation in the asymptomatic screening population), but not both, are designated "borderline RHD". It is not difficult to predict that this latter group could be large and diverse and include cases varying from the normal spectrum to those that undoubtedly have RHD. The reason for this is partly due to the fact that the main form of valvular dysfunction seen, mitral regurgitation (MR), is quite non-specific and occurs secondary to a variety of pathologies. Seen in its mild form it is commonly identified as part of the normal spectrum. Moreover, differentiation of true pathological from functional (normal spectrum) regurgitation is not as simple as measuring a jet length and attributing excess length to RHD. It is important to consider the potential mechanisms at play. Valvular clefts, milder forms of myxomatous degeneration and small fenestrations are potential alternative causes that need to be considered on the pathology side of the spectrum. In addition, it is now becoming clear to us that normal (non-rheumatic) spectrum valves that leak through prominent posterior leaflet inter-scallop separations/clefts (part of the normal spectrum of mitral valves) can cause regurgitation in the pathological range, as judged by the WHF criteria. This muddies the waters of functional versus pathological regurgitation assessment and underscores the importance of identifying the mechanistic cause of regurgitation in every case of regurgitation, whether pathological (but mild) or not. Identifying whether the regurgitation you have identified has a normal spectrum variant morphological counterpart that can be demonstrated on the echocardiogram to be the cause of the MR goes a long way to reducing the bulk of normal cases that land up in the "borderline" bin. This highlights the danger of divorcing valve dysfunction from a mechanistic cause based on morphology and leaflet motion and illustrates one of the problems related to weighting the functional assessment too heavily in the criteria. The other side of this coin is that a heavy handed reliance on a functional valvular deficit leaves cases of isolated, but typical, morphological valve changes of RHD, stuck in this "borderline RHD" group. This would appear to protect specificity in some instances, but at the cost of sensitivity. Getting the balance right will require reassessment of the main elements of the current diagnostic criteria.

## FUNCTIONAL EVALUATION FOR RHD: FOCUS ON REGURGITATION EVALUATION

The principle that pathological MR identified during screening in a high prevalence area for RHD, is more likely to represent RHD, is a well-accepted one and is the departure point for the next discussion. Whether the presence of MR adds fundamentally to the diagnosis of RHD, or is useful in screening because it alerts the screener of a potential valve problem that might turn out to be RHD, is another question altogether.

Despite the well known technical pitfalls of assessing regurgitant jets with colour Doppler, including the importance of colour scale settings, jet length measurement represents a very reasonable start to the assessment of regurgitation significance. The majority of guidelines on severity assessment of patients with moderate or more MR and AR (Aortic Regurgitation) have removed jet length assessment from suggested quantification assessment. However, the context here is different and relates to the assessment of mild, or very mild, MR. Guideline quantification focuses on, and performs best at differentiating moderate from severe MR but has a smaller role to play with milder degrees of MR. MR jet length assessment is simple and reasonably reproducible. The question asked here is whether this is more MR than expected in the normal population. The jet length requirement of >2cm seems to fare reasonably well in isolation, although the validation of this against a set of criteria requiring MR as part of the criteria is of course inherently flawed.[14,22] In addition, as one would predict, it has been shown to miss a number of cases with isolated morphological defects in a screening context.[14] The increase of jet length, from the 1cm used in previous guidelines to the 2cm (for MR) cut-off used in the WHF, appears to have addressed specificity problems quite well and in this context the question really is what added value any additional Doppler criteria offer to a simple colour flow assessment.

The requirement that a regurgitant jet should be visible in 2 different views tests nothing else than the operator's ability to visualise where the jet is originating from, and to section that plane in another view. This should always be achievable, even if it requires dynamic scanning, and this per se should say nothing about whether the MR is pathological or physiological. It could even be argued that eccentric or commissural jets, which are not central but probably have a higher likelihood of being pathological or related to RHD, are less likely to fulfil this requirement as they are less likely to be sectioned in the standard echo planes. This criterion is in serious need of retirement.

The requirement that an MR jet should achieve a maximum MR jet velocity of >3m/s to rule in for pathological MR again makes little sense. The MR jet velocity represents the instantaneous pressure difference between the left ventricle (LV) and left atrium (LA) and unlike CW jet density is not a measure of regurgitation severity. Very broadly speaking, the pressure difference between the systemic LV and low pressure LA is often in the range of 100mmHg (e.g. 110mmHg LV systolic pressure in a normotensive individual) – 10mmHg (normal LA pressure in an individual with low filling pressures). This would equate to an MR jet velocity of 5m/s via the simple Bernoulli equation ($\Delta P = 4V^2$). This velocity should not reasonably be expected to be below 3m/s (36mmHg) in anyone with an intact circulation. The argument here is undoubtedly that an incomplete MR CW Doppler envelope, related to milder degrees of regurgitation, is likely to have a lower velocity because the envelope is incomplete. If this is in fact the argument, the criterion can simply be restated to require a complete Doppler envelope to be present, a simple assessment to do. The misleading idea of a velocity assessment of 3m/s as a judgement of severity could then be abandoned. In addition, it represents duplication of the principle of assessment for an incomplete jet, which is better judged in other ways. Problems with Doppler jet alignment degrading Doppler jet velocities, irrespective of regurgitation severity, are sidestepped if the velocity focus of this criterion is removed. At the very least, abandoning this criterion would be an honest attempt to establish a logical mechanistic foundation for the assessment. In short, as it currently stands, it makes no logical sense.

The last requirement is that a pathological MR jet should be pansystolic (and pandiastolic for AR). Here it is important to differentiate the jet that is not occurring throughout systole from the jet that is periodically moving out of the Doppler beam due to cardiac translation in systole (or diastole), the latter represents a technical problem and would not indicate a lesser degree of regurgitation. Late systolic MR may hint at the underlying mechanism due to prolapse spectrum disease, which remains the one useful aspect of this evaluation. This must then be evaluated specifically. Colour m-mode is a very useful modality to add in this context, as it can be used to accurately define the time course of regurgitation after the fact, during post processing. This may be relevant if the Doppler envelope suggests MR is not pansystolic, and yet this does not correlate

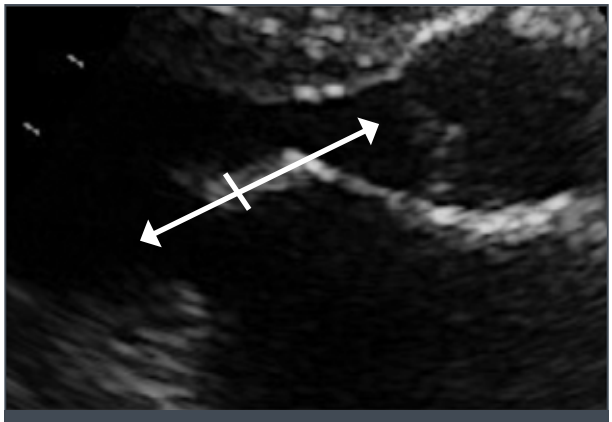

**FIGURE 1: Measuring AMVL thickness.**
The double headed arrow indicates the direction of leaflet orientation. The thickness measurement is done perpendicular to this from edge-to-edge at the thickest area of the leaflet. A measurement of >3mm done with harmonics off would indicate a thickened AMVL and score a morphological criterion for RHD based in the WHF criteria. PSLAX freeze frame at end diastole (zoom view).

with your visual assessment, essentially giving you a second chance to reassess this. It follows that, all else being equal, a pansystolic jet is likely to represent more severe degrees of MR when compared to a jet that occupies only part of the cardiac cycle. However, without a pathological mechanism demonstrated to be responsible for the regurgitation, it actually adds little. Conversely, if an abnormal mechanism of regurgitation is present, the pansystolic nature of the jet is not required to make the diagnosis of abnormality. As noted though, the identification of late systolic MR, and the possible clue that this gives of a prolapse spectrum mechanism, remains informative.

## MORPHOLOGICAL ASSESSMENT OF RHD

### Leaflet thickness

The anterior mitral valve leaflet (AMVL) thickness is measured in the parasternal long axis view (PSLAX) towards the end of diastole with the MV maximally open and with the AMVL at maximal excursion. It is intuitive to imagine the leaflet taking on its thinnest configuration in this fully stretched out state, which supports assessing the leaflet thickness at this time in the cardiac cycle. In addition, it is suggested that the leaflet and chords are maximally separated at this point before a leaflet thickness measurement is done.[16] The AMVL thickness measurement is done as an edge-to-edge measurement (tissue blood interface to tissue blood interface) and perpendicular to the long axis of the leaflet at the point where the leaflet is deemed thickest (Figure 1). Another important stipulation is that the measurement be done with harmonics turned off (harmonic imaging will tend to cause blooming of leaflet tissue with an increase in the apparent leaflet thickness), an often noted omission when reviewing measurement technique (Figure 2).

The theoretical logic behind this methodology is sound as it firstly promotes accurate measurement of leaflet thickness by utilising axial resolution of ultrasound (when ultrasound hits the structure in question in a perpendicular fashion) in this view and timing in the cardiac cycle. Secondly, utilising this methodology the measurement is done in the near field, which also maximises resolution, minimises edge smearing and therefore tends to minimise over-measuring of leaflet thickness (Figure 3). However, these positives have to be weighed against the downside of this technique, namely, difficulty in separating chords from leaflet when the AMVL is maximally open. The maximally open leaflet position promotes positioning of the chords on top of the ventricular aspect of the open MV leaflet. It can be difficult, or impossible, to achieve separation of chord and leaflet in this position. Over-measurement easily occurs at the tips of the AMVL where chords tend to implant and where leaflet–chord separation can be especially difficult to achieve. This is of course the area of the leaflet that often thickens first when affected by the rheumatic process and it is critical to ensure that subvalvular tissue is not included in the measurement. To measure the true leaflet thickness (free from chordal inclusion) might mean doing the measurement in a different view and at a different time point in the cardiac cycle which often leads to a remarkably smaller maximum thickness measurement (leaflet measures thinner) when compared to that achieved using the standard, recommended methodology. The realisation is often that the true leaflet was just not visualised separate from the chords (Figures 4, 5 and 6).

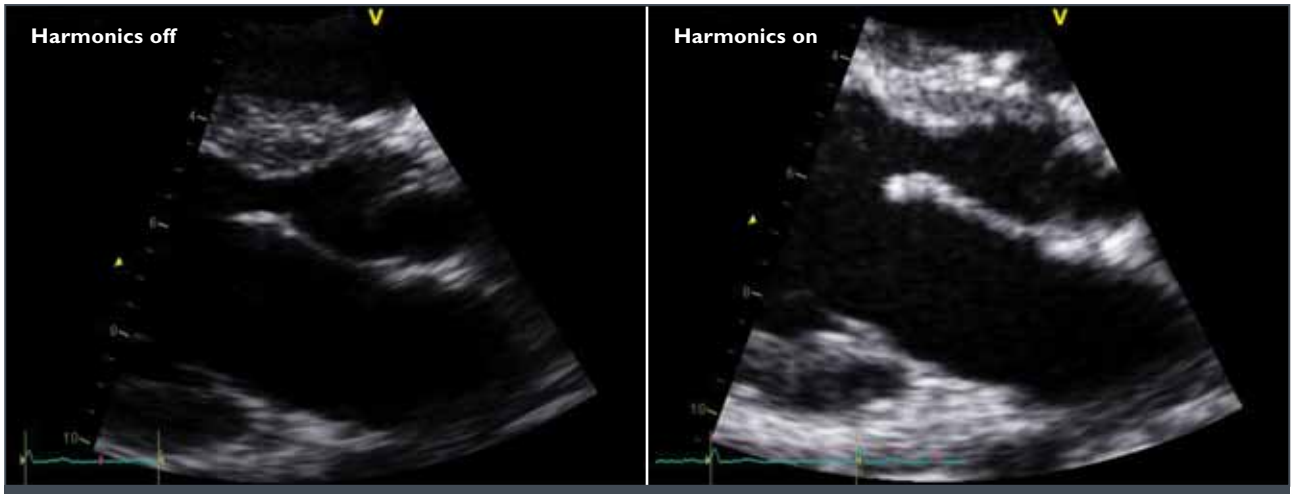


**FIGURE 2: Harmonic Imaging.**
Harmonic imaging improves overall image quality and has become almost the default setting in general scanning practice. However, it can make structures look significantly thicker than they actually are due to the increase in echo pulse length. This becomes critical when making measurements of a very small magnitude such as with leaflet thickness measurements. The normal values given for leaflet thickness are based on measurements derived with harmonic imaging set to off. These PSLAX freeze frames at end diastole in the same patient illustrates the point.

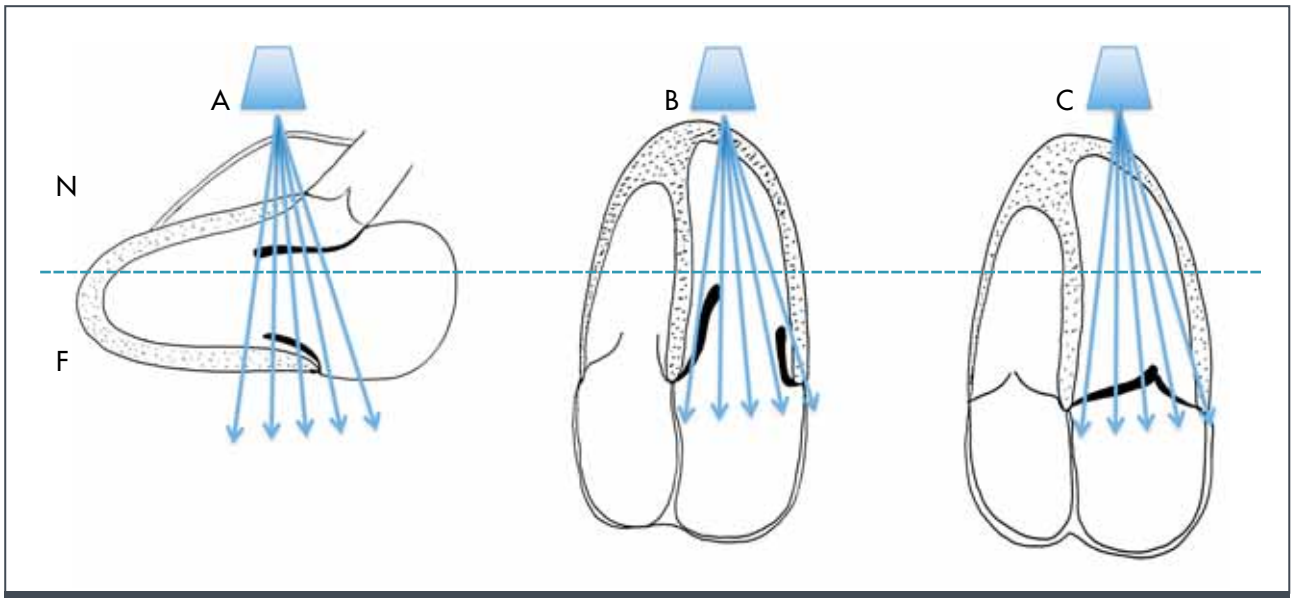


**FIGURE 3: Maximising Resolution.**
In the PSLAX view (A) the maximally open MV places the AMVL in the ultrasound near-field and perpendicular to the ultrasound (US) waves utilising axial resolution. This maximises resolution. In B and C (representing the Apical 4 chamber view [A4C]) the US waves run parallel to the open leaflet in B (utilising lateral resolution) and although perpendicular to the closed MV leaflet in this view the valve is in the far-field which decreases resolution due to the spread of the US beam in the farfield. N – Nearfield, F – Farfield, MV – mitral valve, AMVL – anterior mitral valve leaflet. The arrows represent the direction of the ultrasound beams. Arrow line density represents theoretical resolution.

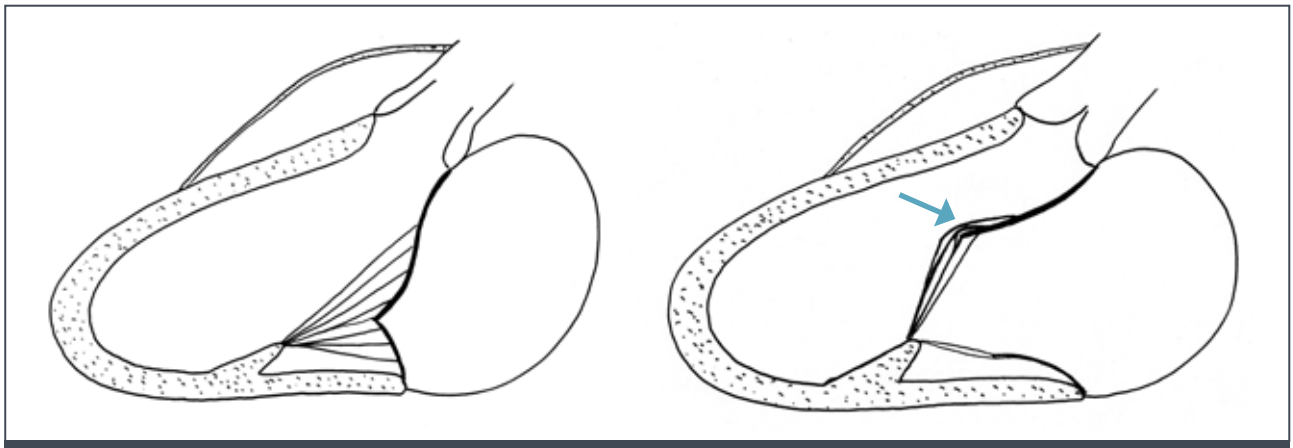

**FIGURE 4: Separating leaflet from chords.**
The image on the left demonstrates good separation of leaflet from chordal tissue with the mitral valve in the closed position. This is significantly more difficult to achieve with the valve open (image on the right) where chords attaching to the ventricular surface of the valve end up "on top" of the anterior mitral valve leaflet (Arrow). In this position chords are often very difficult to separate from the leaflet and are included in the thickness measurement. An intermediate position might provide a good compromise of separation and resolution in these cases.

Modification of this methodology might be one strategy of bringing normal AMVL thickness measurements by echocardiography (normal up to 3mm with harmonics off and possibly up to 4mm with harmonics on) more in line with leaflet thickness measurements quoted from pathology studies (around 1mm in the typical screening ages and up to 1.6mm in the older patient population).[16, 23, 24] The effect that ultrasound pulse length has on increasing apparent leaflet thickness is unlikely to account for a three-fold increase in this measurement on fundamental imaging and if relevant could be accounted for by using a leading-edge-to-leading-edge measurement convention.

When acquiring the PSLAX image, with a view to assessing leaflet thickness of the mitral valve, the sonographer should

attempt to section the most central portion of the AMVL rather than towards the commissures. A narrow central strip of the leaflet can often be identified that is relatively devoid of chordal tissue in the frame (Figure 7). Once the image is stopped and scrolled it is also useful to visually compare the basal aspect of the AMVL to the apparent thickness of the leaflet tip. If a uniform thickness can be seen to run through, from base to tip, it makes the presence of true thickening unlikely and aids the eye in separating leaflet from chord. The fact that RHD often affects the leaflet in a focal manner with the tips affected first, also presents the opportunity to do a relative leaflet thickness assessment, similar to that done for the assessment of aortic valve thickness.

Posterior mitral valve leaflet (PMVL) tip thickening is recorded much less frequently in the patient cohort with asymptomatic RHD, but may be more specific and is not as easily "over read". Sensitivities suffer if this is used in isolation though.

### Leaflet restriction

Leaflet restriction, and specifically the pattern of leaflet restriction, is arguably the most specific feature of "rheumatic morphology" quoted in the WHF criteria. The most common form of leaflet restriction in RHD is leaflet tip restriction due to commissural fusion (see explanation of mechanism below). This

**FIGURE 5: Chordal arrangement to the AMVL.**
The AMVL in this gross pathology specimen shows the ventricular aspect of the mitral valve. Primary chords attach closer to the edge of the leaflet and secondary strut chords (white arrow) to the body of the leaflet. This adds significant bulk to the otherwise thin leaflet.

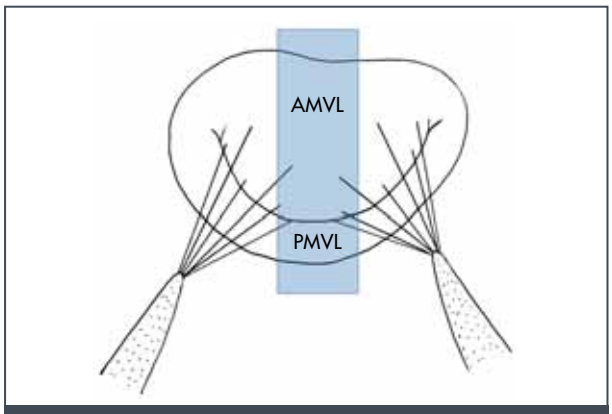


**FIGURE 7: Measuring leaflet thickness.**
The central portion of the AMVL (blue strip) is relatively devoid of strut chords that add bulk to leaflet thickness measurements. The leaflet should be sectioned in this central portion to try and minimise the effect that strut chords have on leaflet thickness assessment.

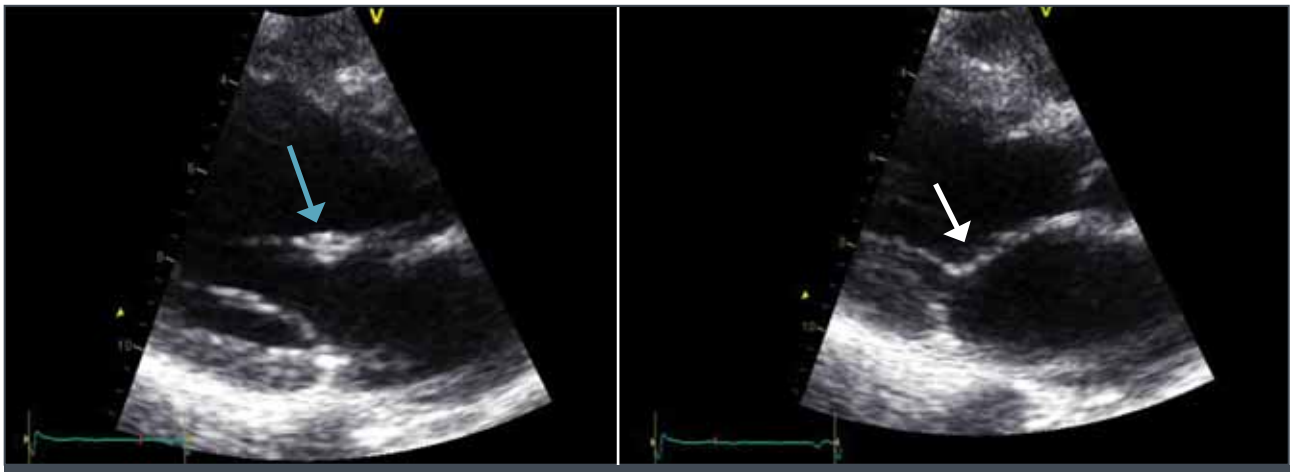

**FIGURE 6: Apparent AMVL tip thickening.**
These 2 parasternal long axis still frames of the mitral valve demonstrate apparent anterior mitral valve leaflet (AMVL) tip thickening (blue arrow) due to chordal interference left. Partially closing the MV solved the dilemma easily. The chordal overlap responsible for the thickening could then be identified as separate structures and eventually a separate and thin AMVL running through this (white arrow) was demonstrated with the valve almost completely closed (right).

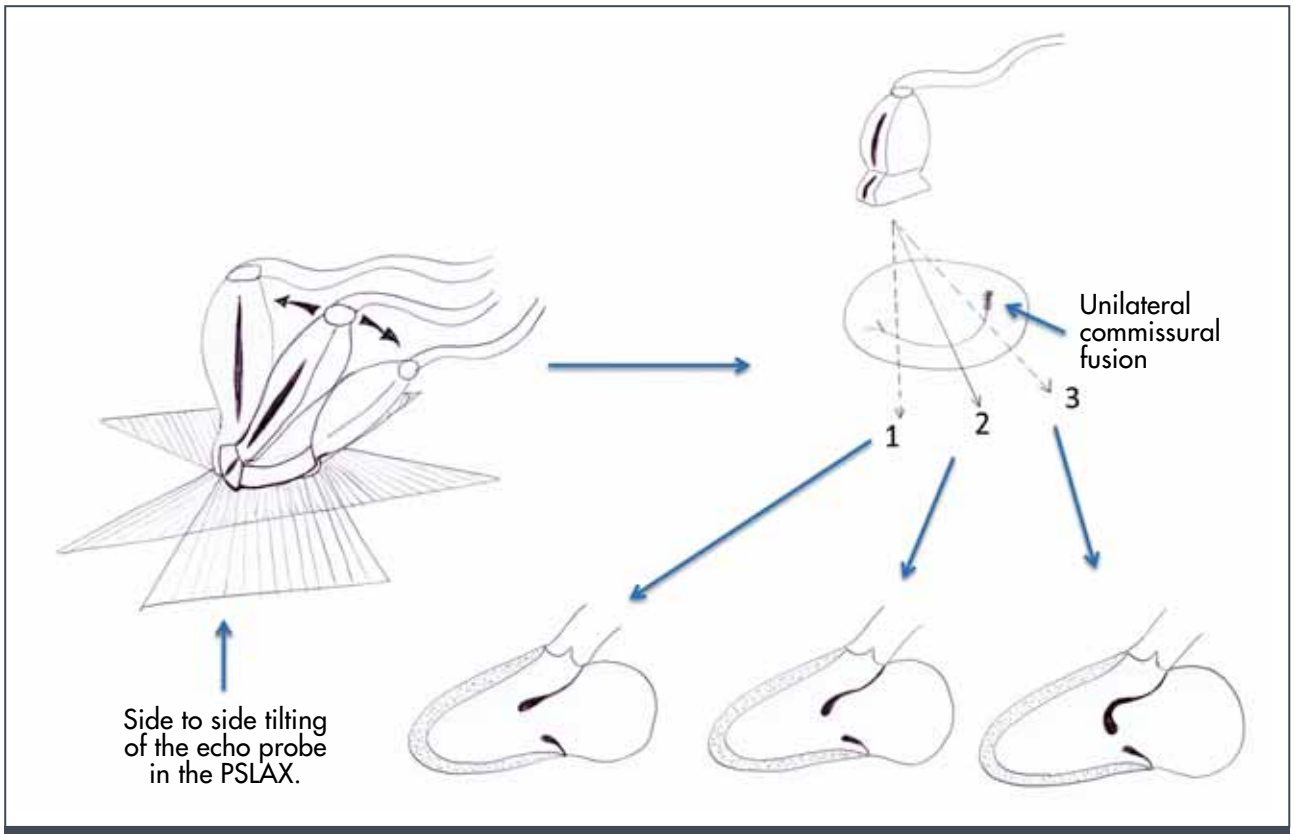


**FIGURE 8: PSLAX sweep.**
See text for full descriptive legend.

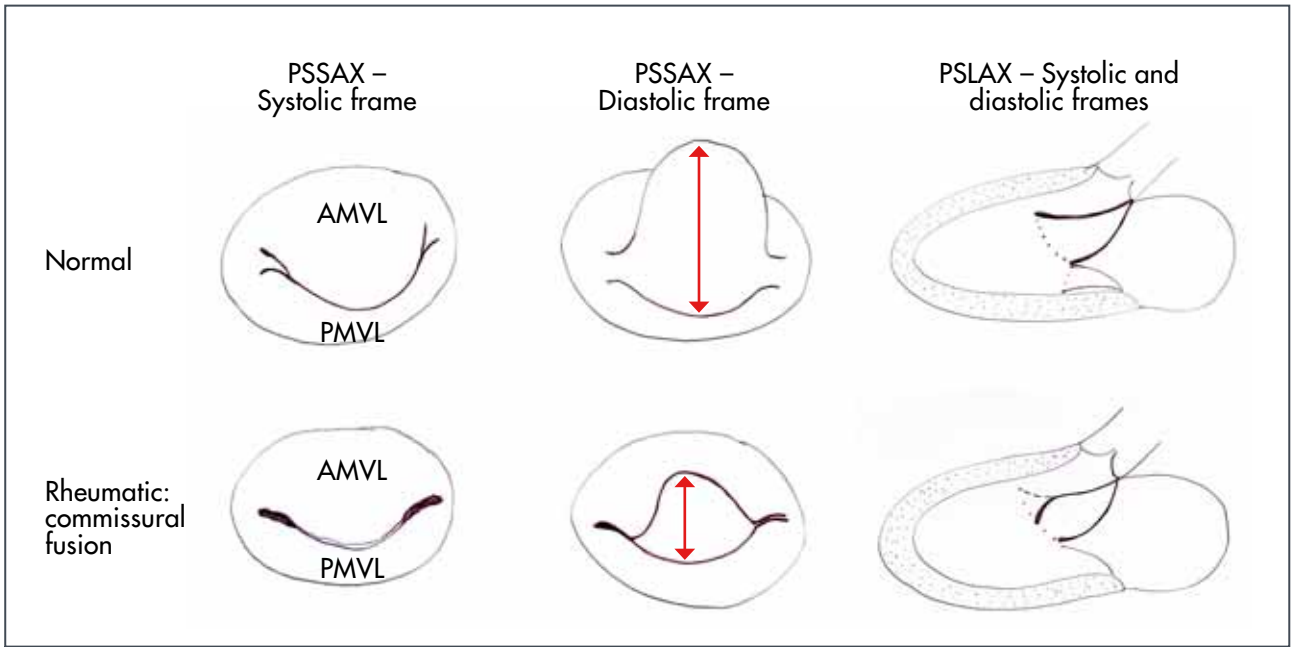


**FIGURE 9: Mechanism of bowing from commissural fusion: Mitral Valve.**
Commissural fusion limits vertical leaflet edge separation in diastole (see red arrow above). If AMVL length remains unchanged, this must translate into bowing of the mobile leaflet body as seen in the PSLAX frames. The PSLAX images illustrate the fact that the leaflet tip motion is halted along the normal arc of motion but the body and or base continues to move forward leading to bowing. PSSAX – parasternal short axis, PSLAX – parasternal long axis.

leads to diastolic bowing of the leaflet, identical to that seen in patients with Mitral stenosis (MS), but often to a lesser degree in the subclinical RHD population. Bowing of the AMVL is assessed primarily in the PSLAX. Freeze the PSLAX image optimised to show the AMVL and carefully scroll to show the AMVL when the leaflet reaches its maximal diastolic excursion. At this point bowing can be defined as the tip of the AMVL pointing posteriorly towards the posterior LV wall rather than parallel or away from this wall. Assessing the presence and distribution of bowing of the AMVL is a critical step in the evaluation of early restriction of a valve. As is often seen in more advanced disease with unilateral commissural fusion, and also in the post mitral valvuloplasty patient after unilateral commissural cleaving, the leaflet bowing is maximal on the side of maximal commissural fusion. Sweeping from commissure to commissure (tilting the echo probe from side to side in the PSLAX) is therefore an important manoeuvre to do since unilateral bowing of the leaflet may be prominent on one side of the valve only, ipsilateral to the fusion (Figure 8). It should be noted that bowing of the AMVL is also seen in congenital clefts of this leaflet and AV bowing is typical of bicuspid aortic valve disease, so that the presense of bowing should always spark a search for these pathologies. Unfortunately the complexity does not stop here as it would appear that bowing of the medial aspect of the AMVL is also seen quite frequently in the normal population and must be differentiated from that associated with RHD. This also explains why bowing is often overcalled when evaluated in the apical 4 chamber view (and why this view should not be used to evaluate bowing) where the sonographer is evaluating the posteromedial aspect of the AMVL (A2/A3). The exact mechanism of this observation requires to be fully elucidated. More advanced leaflet restriction can be seen as a fixed leaflet, a configuration often seen of the PMVL in more advanced cases. It is informative to understand why bowing occurs in rheumatic valves, as it is such an important identifier of rheumatic valvular involvement (Figures 9 and 10).

When acquiring the PSLAX, and scrolling from commissure to commissure, the sonographer should attempt to acquire the most central portion of the AMVL for assessment of evidence of rheumatic bowing. The relatively narrow central strip of the leaflet is identified as an area relatively devoid of chordal tissue. A judgment should also be made as to whether bowing appears to be isolated to the medial aspect of a valve other-

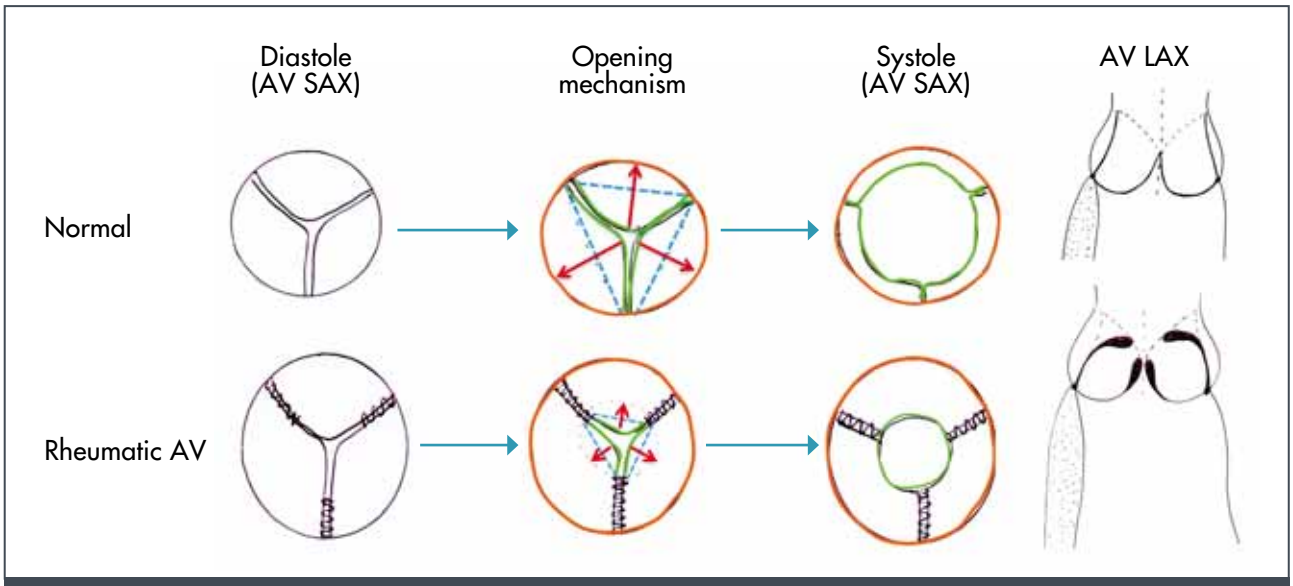


**FIGURE 10: Bowing from Commissural fusion: Aortic valve (AV).**
This top image row shows how normal AV opening occurs around a fulcrum with its two corners in the apexes of adjacent AV commissures (blue interrupted lines). Open commissures allow the free luminal edges of the 3 semilunar cusps (green lines) to end up almost flush against the aortic wall in systole (orange lines). The bottom image row represent a rheumatic aortic valve with fusion of the basal half of each commissure. The fulcrum around which each semilunar cusp can now hinge (blue interrupted line) has been moved closer to the tip of each semilunar cusp and therefore towards the center of the aortic lumen because of the commissural fusion. This prevents the aortic leaflet edges (green) from ending up flush with the aortic wall at maximum systole. This leads to stenosis in severe cases, but also underpins the mechanism of bowing in less severe cases. Commissural fusion limits leaflet edge separation in systole and therefore limits the opening area of the AV in systole (green area). If AV leaflet length remains unchanged, this must translate into bowing of the mobile leaflet body as seen in a long axis view. The long axis (LAX) images illustrate the fact that the leaflet tip motion is halted along the normal arc of motion, but the body and or base continues to move forward leading to bowing. Bowing is a very telling sign of commissural fusion seen at any valve from whatever pathology. This AV description underlines the fact that chordal involvement is not a prerequisite for bowing to occur.

wise free of rheumatic features which, as stated before, is a frequently seen normal pattern.

A more subtle finding of leaflet restriction, often seen in the presence of bowing, is that the AMVL loses its free fluttering of the tips (ossilation around the horizontal when the leaflet is maximally open) as the AMVL leaflet becomes tethered to the PMVL in the commissures. Identifying this free fluttering tip motion is reassuring and argues against commissural fusion being present. The absence of fluttering should spark the search for leaflet tip bowing, more advanced forms of leaflet restriction and more direct evidence of commissural fusion.

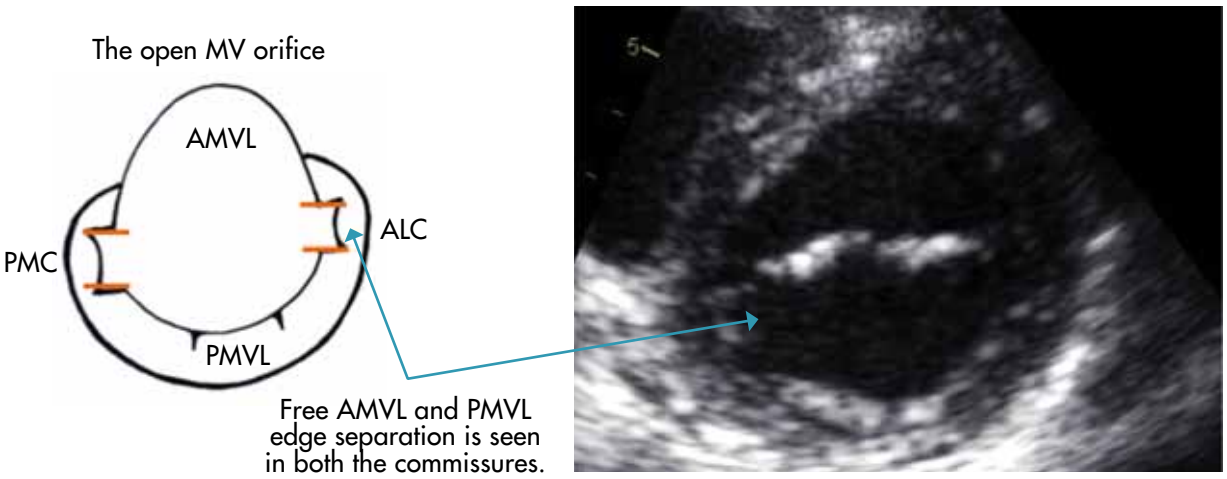


**FIGURE 11: The normal mitral commissure.**
The normal mitral commissure allows for free separation of the AMVL and the PMVL in the commissure. A commissural scallop often facilitates this separation. This normal "unhinging" of the AMVL and PMVL is critical for ensuring a large MV orifice in diastole. Commissural fusion causes fusion of the AMVL and PMVL edge in the commissure resulting in a reduced MV orifice opening area. Fluttering is a more subtle feature of the normal, freely mobile commissure.

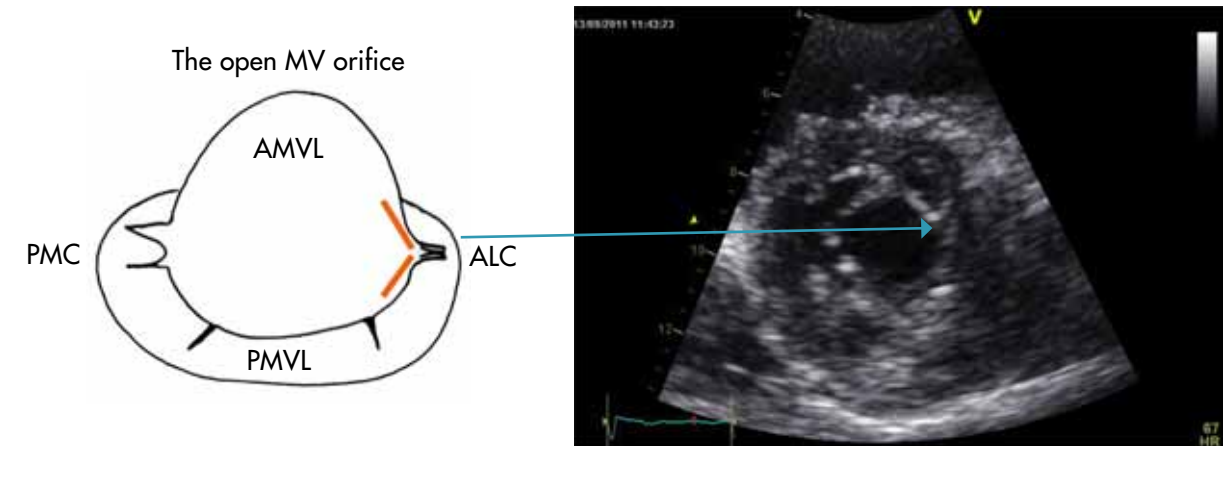


**FIGURE 12: The rheumatic mitral commissure.**
Partial fusion at the apex of the postero-medial commissure (PMC) involving the lower edge of the commissural scallop and complete fusion at the apex of the antero-lateral commissure (ALC) is illustrated in the line drawing. The leaflet angle running into the commissure often becomes more angulated once fusion occurs. This freeze-frame illustrates a lateral commissure that does not separate freely due to fusion seen right at the edge of the commissure. AMVL bowing was prominent in this case.

## COMMISSURAL FUSION

Commissural fusion is a very specific marker of rheumatic mitral valve involvement. It is well known to be the mechanism for the development of MS in advanced RHD. The normal mitral valve area is 4-6cm$^2$ and it stands to reason that mild degrees of commissural fusion will initially reduce the mitral valve area only slightly, and not into the MS range. The specificity of this process is what makes it such an appealing marker of rheumatic valve involvement. As noted above, rheumatic bowing of the mitral and aortic valves lead to bowing through commissural fusion as the predominant mechanism, but commissural fusion should also be sought directly. Both commissures must be carefully inspected in the parasternal short axis view (PSSAX) for evidence of early fusion. The normal mitral valve is seen to allow free separation of the AMVL from the PMVL in the commissure. This is often fascilitated by a commissural scallop and allows for almost parallell separation of the 2 leaflets in the commissure. The PSSAX view must be modified, with an acquisition done slightly more basally, to optimise the view of the commissures and in some cases the two commissures must be acquired using two separate views (angulated differently for each commissure) in order to optimise each commissure for assessment. Unfortunately, the learning curve for this assessment is quite steep and minor fusion can be difficult to visualise on 2D echocardiography which significantly degrades the sensitivity of this technique as an isolated feature. In some cases, however, it simplifies an otherwise difficult assessment significantly (Figures 11, 12).

## SUB VALVULAR INVOLVEMENT

The rate of identifiable sub valvular involvement in the asymptomatic RHD population appears to be quite low. It is important to screen both chordal systems carefully for areas of thickening. The most difficult area to assess is the area of chord just below the AMVL tips. It can be difficult to accurtely judge where the leaflet ends and the chords begin in the PSLAX view with the MV maximally open. Sidelobe artifact can cause "lateral smearing" of linear structures (such as the chords which will be running horizonatlly in the PSLAX view) which often make this assassment very difficult. For the same reasons chordal thickening is often over read in the PSLAX view and if suspected from this view, must be confirmed by apical scanning before calling it. The modified 4C, 2C and 3C views from the apex are very useful in assessing the whole length of the chords for areas of abnormal thickening.

## EXCESSIVE LEAFLET TIP MOTION DURING SYSTOLE / LEAFLET TIP PROLAPSE (MV)

The central mechanism for the development of MR in RHD is so-called pseudo-prolapse. As suggested by this terminology there is no true prolapse at play here. Rather, the mechanism of MR in these cases is PMVL restriction which causes the AMVL tip to move past the relatively fixed PMVL during systole, leading to a coaptation defect and MR. True prolapse is rarely seen. It is accepted that chordal rupture, with subsequent prolapse or flail, rarely complicates acute rheumtic fever and excessive leaflet motion can, in these selected cases, be a cause for pathological MR. Excessive leaflet motion is, however, in our experience not a feature of rheumatic MR or AR outside of the acute rheumatic fever phase of the condition, and only rarely in this situation. The inclusion of excessive leaflet motion as defined by "excessive leaflet tip motion during systole/tip prolapse" conspires to achieve 2 things: Firstly, it risks inclusion of prolapse spectrum disease into the rheumatic popultion being screened and secondly it risks double scoring for true RHD cases with pseudo prolapse. In the latter cases the candidate will be scored for both "tip prolapse" and PMVL restriction which represents the same thing in these cases. Identifying PMVL restriction is an important component of identifying the mechanism of pseudo-prolapse seen in RHD. This mechanism should not be implied by focusing on tip movement without identifying the restriction component. The recurring theme of true rheumatic diastolic leaflet restriction appears to be a central ingredient in the diagnostic process towards asymptomatic RHD. Note: Similar arguments can be made for the aortic valve in terms of the "prolapse" criterium.

## COAPTATION DEFECT OF THE AORTIC VALVE (AV)

Visualising a coaptation defect in AR is typically associated with significant AR, but is in no way specific for rheumatic involvement. The inclusion of this criterion amongst the morphological critera of rheumatic AV involvement is interesting, but either inaccurate or out of place, belonging perhaps amongst morphological markers of severity rather than suggesting it represents rheumatic AR specifically.

It is important that we are critical when looking at the criteria used to diagnose RHD so that we can improve on them and thus take the field forward. Some of the individual criteria of the WHF tasked with identifying pathological regurgitation appear to be redundant. Conversely, some of the criteria that form the basis of a morphological diagnosis of the condition appear to deviate from what we see in daily clinical practice. This will have to be reconciled if we wish to move forward with our

hope of large-scale population based screening. Weaknesses in the original WHO Doppler criteria has been perpetuated by "incorporation" of these criteria into the WHF criteria and the matter of form versus function, and where the focus should lie, still needs to be addressed. Further progress in development of the criteria must remain an important goal. This will necessitate ongoing and critical scrutiny of the criteria to understand where and why certain aspects are weak and where they could be improved upon. A deeper understanding of the mechanisms involved, and credible fundamentals underlying all proposed criteria, are paramount to rationalising and improving the criteria.

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