

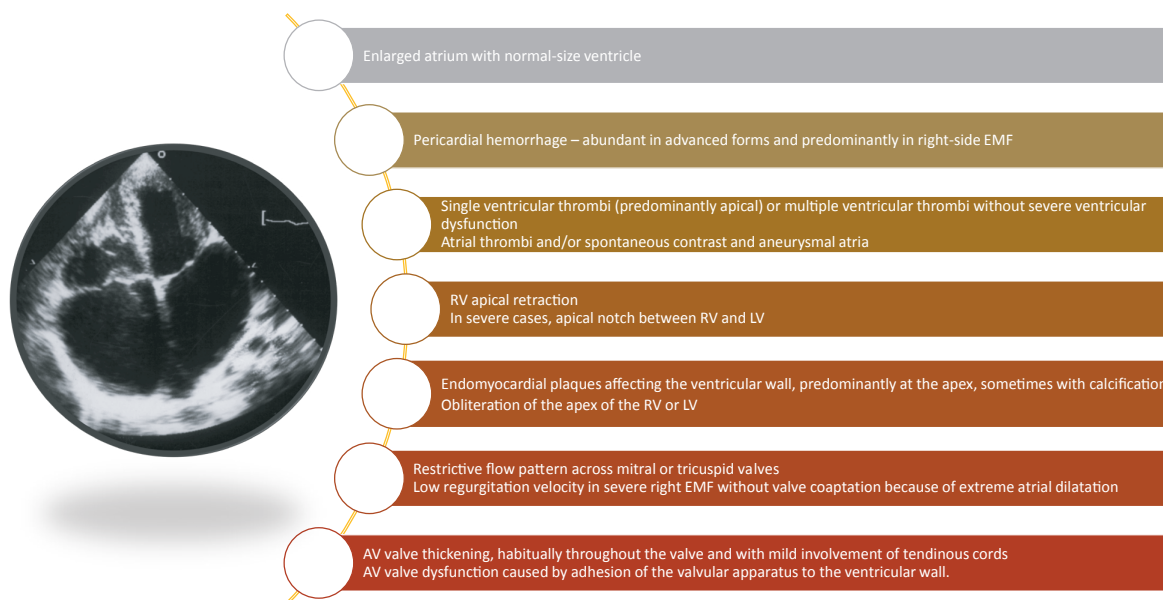
## How Do I Diagnose and Classify Endomyocardial Fibrosis in Under-Resourced Settings

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### Central Illustration: Echocardiographic Characteristics of EMF



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EMF: Endomyocardial fibrosis; RV: right ventricle; LV: left ventricle.

### Abstract

Endomyocardial fibrosis (EMF) is a disease of unclear etiology and pathogenesis that is characterized by fibrosis in the ventricular cavities and mainly affects children and adolescents from tropical regions of Africa, South America,

and Asia. It is classified as a restrictive cardiomyopathy, with pathophysiology of restriction to diastolic filling associated with valve abnormalities, both caused by endocardial fibrosis. Typical echocardiographic features of this condition are endocardial thickening, severe atrioventricular (AV) valve regurgitation, aneurysmal atria, and heart distortion.

### Keywords

Endomyocardial Fibrosis; Echocardiography; Diagnosis

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In endemic areas of Africa, in the absence of advanced imaging techniques, EMF diagnosis and management relies on transthoracic echocardiography for careful assessment of structural and hemodynamic abnormalities, aiming at planning medical or surgical management. This review highlights the key echocardiographic aspects of diagnosis and classification of EMF, as used in under-resourced settings in Africa. This is a standardized approach to screening populations with known occurrence of the disease and for informing medical and surgical management.

Introduction

Endomyocardial fibrosis (EMF) is a progressive disease of unclear etiology and pathogenesis that, when established, is characterized by fibrosis in the ventricular cavities, particularly affecting the apex and the subvalvular regions. It is a restrictive cardiomyopathy that affects mainly children and adolescents, predominantly from certain rural settings in Africa.<sup>1</sup> The pathophysiology of EMF associates restricted diastolic filling with major changes in the valve apparatus caused by endocardial fibrosis, resulting in severe atrioventricular (AV) valve regurgitation, aneurysmal atria, and heart distortion. Advanced forms of EMF have high morbidity and mortality, with death occurring due to progression to chronic heart failure or as a result of arrhythmia and thromboembolism.<sup>2</sup> Although there is no specific treatment for EMF, surgery should be considered to correct some structural and functional abnormalities in symptomatic individuals. Surgical techniques have been progressing,<sup>3</sup> but outcomes are yet to be fully understood.<sup>4</sup>

In endemic areas of Africa, in the absence of advanced imaging techniques such as magnetic resonance imaging, EMF diagnosis and management often relies on detailed transthoracic echocardiography. Transesophageal and three-dimensional echocardiography are rarely used, magnetic resonance imaging is also rarely available, and catheterization is often contraindicated because the risks outweigh the benefit - due to presence of thrombi and because of the difficulty of obtaining endomyocardial tissue in advanced disease. Detailed echocardiographic characterization and classification of EMF has been shown to provide reliable information on the key structural and hemodynamic abnormalities that are confirmed in surgery.<sup>5</sup> This review highlights the key echocardiographic aspects of diagnosis and classification of EMF in under-resourced settings in Africa.

Echocardiographic Criteria for Diagnosis of EMF

The echocardiographic criteria for diagnosis of EMF were described in a highly endemic population from Mozambique.<sup>6</sup> Using these echocardiographic features, cardiac abnormalities in patients with EMF were divided into “major” or “minor”, according to their clinical relevance, uniqueness to the diagnosis, influence on management decision, and prognostic importance. (Table 1)

*Major criteria* are distinctive or pathognomonic features of EMF, which usually have a major impact on management. Each feature is attributed an individual score according to its relevance to determination of structural and hemodynamic changes. The following are considered major criteria for diagnosis of EMF: i) Endocardial plaque of at least 5mm in width or endocardial thickening greater than 1mm; ii) Patchy endocardial thickening in two or more ventricular walls; iii) Obliteration of ventricular apices or valve recesses; iv) Ventricular thrombi or spontaneous contrast in the absence of ventricular dysfunction; v) Reduction of the right ventricle (RV) cavity volume due to exclusion of the trabecular portion vi) Restriction of AV valve leaflet movements due to adherence to the ventricular walls.

For screening, endocardial plaques or endocardial thickening of more than 5mm in width and thicker than 1mm are considered

Table 1 – Criteria for Diagnosis and Assessment of the Severity of FEM\*6

| Criterion                                                                              | Score |
|----------------------------------------------------------------------------------------|-------|
| <b>Major criteria</b>                                                                  |       |
| Endomyocardial plaques > 2 mm in thickness                                             | 2     |
| Thin (≤ 1 mm) endomyocardial patches affecting more than one ventricular wall          | 3     |
| Obliteration of the right ventricular or left ventricular apex                         | 4     |
| Thrombi or spontaneous contrast without severe ventricular dysfunction                 | 4     |
| Retraction of the right ventricular apex (right ventricular apical notch)              | 4     |
| AV-valve dysfunction due to adhesion of the valvular apparatus to the ventricular wall | 1-4†  |
| <b>Minor criteria</b>                                                                  |       |
| Thin endomyocardial patches localized to one ventricular wall                          | 1     |
| Restrictive flow pattern across mitral or tricuspid valves                             | 2     |
| Pulmonary-valve diastolic opening                                                      | 2     |
| Diffuse thickening of the anterior mitral leaflet                                      | 1     |
| Enlarged atrium with normal-size ventricle                                             | 2     |
| M-movement of the interventricular septum and flat posterior wall‡                     | 1     |
| Enhanced density of the moderator or other intraventricular bands                      | 1     |

\*A definite diagnosis of EMF was made in the presence of two major criteria or one major criterion associated with two minor criteria. A total score of less than 8 indicates mild EMF, 8 to 15 moderate disease, and more than 15 severe disease. † The score is assigned according to the severity of AV regurgitation. ‡ M-movement of the interventricular septum refers to a pattern of movement observed on M-mode echocardiography that is thought to be due to obliteration or restriction of the left ventricular apex combined with mitral regurgitation. EMF: Endomyocardial Fibrosis; AV: atrioventricular; LV: left ventricle

a major criterion for diagnosis of EMF. The term “large plaque” is used when endocardial thickening exceeds 10 mm in width.

*Minor criteria* are not specific for EMF but may suggest the disease. Although commonly found in patients with EMF, these features are not exclusive to this condition and do not define the condition when found in isolation. They are considered to have less influence on management and prognosis, when compared to the major criteria previously described. The features that are considered minor criteria for the diagnosis of EMF are: i) Patchy endomyocardial thickening localized to one ventricular wall; ii) Restrictive ventricular filling pattern; iii) Diffuse thickening of the AV valve leaflets; iv) Enhanced density of the moderator band or intraventricular trabeculae; v) Abnormal M-movement of the interventricular septum and/or posterior left ventricle (LV) wall; vi) Enlarged atrium with normal-sized homolateral ventricle; and vii) Presence of thickened “false tendon” of the LV.

In asymptomatic individuals, patchy endocardial thickening corresponds to small areas of endocardium with enhanced density, less than 1mm in depth and evenly distributed in the ventricular walls.

## Classification of EMF

The echocardiographic appearance of EMF varies both in terms of the distribution and of the severity of the structural lesions and hemodynamic abnormalities. The criteria described above are used to assess the presence of lesions in each side of the heart, and the scoring system is applied to each ventricle separately. According to the distribution of lesions (affecting exclusively or predominantly one or both sides of the heart) we classified the disease as Right EMF, Left EMF, or Bilateral EMF (Figure 1).

## Severity of EMF

The severity of EMF is assessed by quantifying structural and hemodynamic lesions. Four progressive degrees of severity are defined, supporting management decisions and informing on prognosis (Table 2):

- *Grade I (or Mild EMF)*: Endocardial plaque or patchy thickening associated with thickening of the AV valve leaflets, with no other structural or hemodynamic abnormalities.
- *Grade II (or Moderate EMF)*: Large endocardial plaques, apical/valve recess, ventricular obliteration, and mild to moderate AV valve regurgitation. There is mild to moderate atrial dilatation, mild ventricular cavity reduction, and preserved myocardial function.
- *Grade III (or Severe EMF)*: Large endocardial plaques, moderate reduction of ventricular cavity dimensions, marked atrial dilatation, and severe AV valve dysfunction. Myocardium is visible underneath the endocardial thickening and ventricular function is nearly normal.
- *Grade IV (or Advanced EMF)*: Presence of large endocardial plaques associated with severe reduction of ventricular size and compression of the contralateral cavities by the severely dilated atrium. Endocardial calcification, poor ventricular contractility, and large persistent effusions (pericardial, peritoneal, and/or pleural) are other features of advanced EMF.

The different stages of EMF are exemplified in Figure 2.

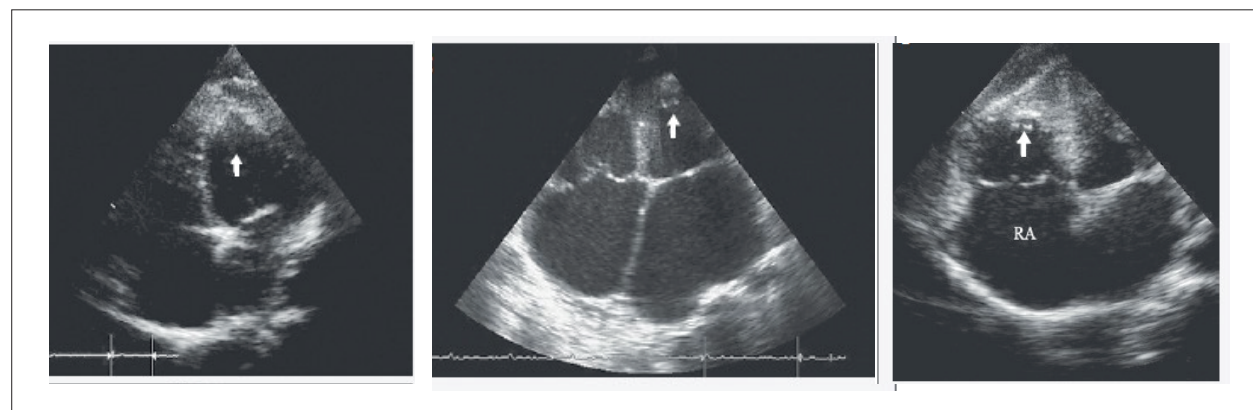
## Staging and Mode of Progression

Staging and characterization of the mode of progression of EMF aims to distinguish active from latent disease, particularly in clinical settings, and is used to support management and to indicate prognosis. In a patient with an echocardiographic diagnosis of EMF, activity was defined as a finding of two or more of the following signs: i) unexplained fever, recurrent facial edema, urticaria or asthma-like episodes; ii) severe hypereosinophilia (absolute eosinophil count  $> 1.5 \times 10^9/L$ ); iii) ventricular thrombi not attributable to severe myocardial dysfunction; iv) evidence of pancarditis with acute heart failure; and, v) increased C-Reactive Protein and/or erythrocyte sedimentation rate. The persistence of these signs and the speed of progression of structural and/or functional abnormalities in the six months following the diagnosis are used to define three distinct stages of the disease: active with remission, active persistent, and rapidly progressive.

*Active EMF with remission* refers to patients who had signs of activity at diagnosis that regressed during the first six months of follow up, while *Active persistent disease* signified persistence of signs of activity for 6 months or more. Finally, we use the expression *Active rapidly progressive* to define patients with persistence of signs of activity after the initial episode of heart failure, who progress to a higher degree of severity or death in less than six months. Depending on the occurrence of disease recrudescence during the first six months of follow up, patients are classified as having quiescent or recurrent disease. Patients who develop recrudescence of active disease in the course of follow up, going back to a quiescent stage thereafter, are considered to have chronic recurrent disease.

Signs of activity are frequent in patients from the clinical registries, who present laboratory signs of active disease, hypereosinophilia, and facial edema. Rapidly progressive disease and death may be associated with intraventricular thrombi and myocardial dysfunction.

Patients are also classified according to the duration of symptoms; if these are present for less than 6 months, they are considered to have acute disease. Chronic disease is defined by the presence of signs and symptoms attributable to EMF for more than six months with echocardiographic features of established EMF.



**Figure 1** – Progression of EMF from occupation of the apex with thrombi (left), to ventricular cavity reduction and atrial dilatation (centre); and large plaque of fibrosis and ventricular retraction (right).

### Experience from Community- and Hospital-Based Studies

The information used to develop the criteria results from careful evaluation of transthoracic echocardiography and follow-up of 1534 individuals (1063 individuals of all ages randomly selected from the community, 296 school children, and 175 patients with established disease cared for in

hospitals). On clinical grounds, the clinical features, biological profile, findings during open heart surgery, and pathological features of tissue obtained *in vivo* were used to complement the studies.<sup>6,7</sup> Of the 413 individuals with EMF (mean age at diagnosis 15 years, with no gender differences), 52 patients (12,1%) recalled having any complaints before diagnosis, confirming the high occurrence of clinical-echocardiographic dissociation in EMF.<sup>8</sup> The most common features seen on echocardiography are summarized in Table 3.

**Table 2 – Main features in EMF of different degrees of severity**

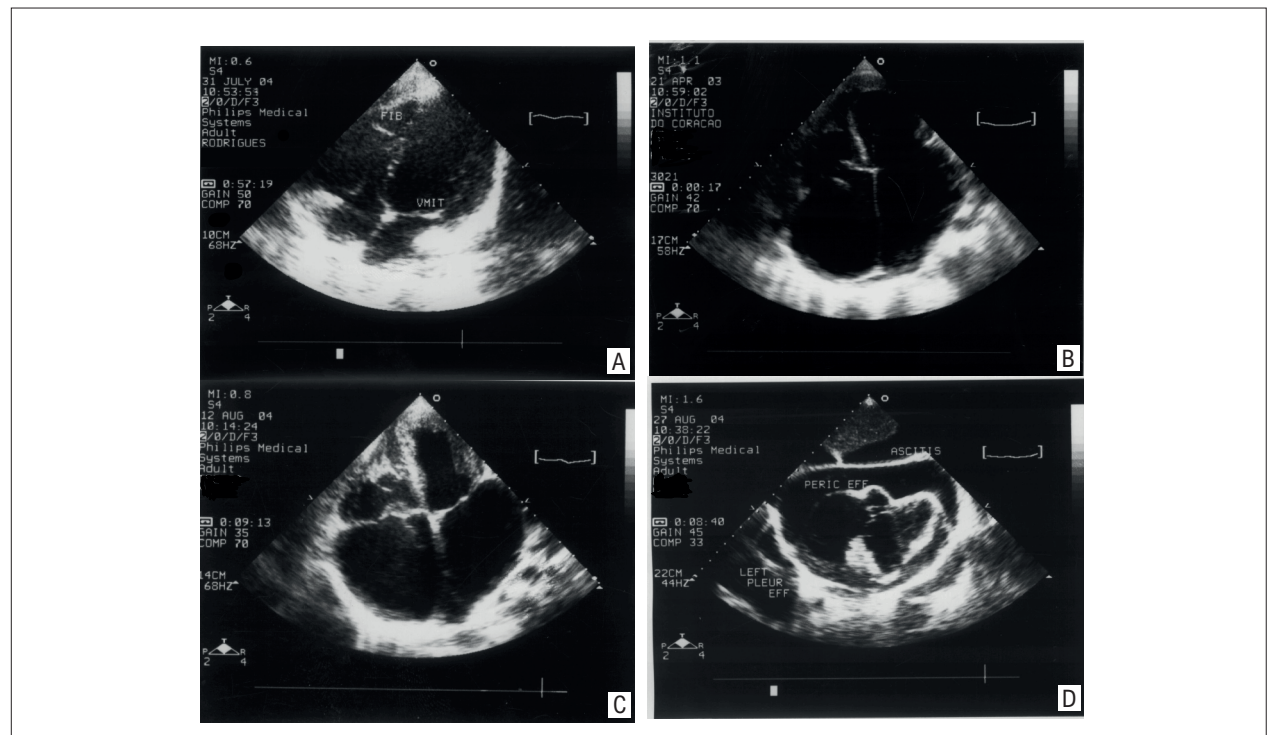
| Grade                  | FEATURES                                                                                                                                                                                                                      |
|------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>I<br/>MILD</b>      | Patchy fibrosis of any ventricle, diffuse thickening of the leaflets and restrictive pattern of ventricular filling, without any other structural changes                                                                     |
| <b>II<br/>MODERATE</b> | Hyperdense lesions on the mural and valvular endocardium, obliteration of the ventricular apex, ventricular thrombi, and/or AV valvular dysfunction, in the absence of cavity deformation/reduction or myocardial dysfunction |
| <b>III<br/>SEVERE</b>  | Extensive fibrosis and/or calcification with functioning myocardium visible underneath it, reduction of ventricular size, severe atrial dilatation, and AV valvular dysfunction                                               |
| <b>IV<br/>ADVANCED</b> | Heart distortion and cavity deformation due to extensive fibrosis or calcification that affects the contractility of the myocardium and leads to free AV regurgitation                                                        |

AV: atrioventricular.

### Abnormalities of the Left Side of the Heart

Early left EMF is often detected in asymptomatic individuals in the community and may be characterized by fibrosis of the *false tendons*, thickening of the mitral leaflets (Figure 2a), apical thrombus, and/or obliteration of the apex or the recess between the posterior leaflet and the posterior wall. Thrombi in the subvalvular apparatus involve the free edges of both papillary muscles. Flow across the mitral valve shows early diastolic filling.

In established left EMF, thickening of the endocardium is prominent in the apex and posterior wall behind the recess of the posterior leaflet of the mitral valve. The ventricle assumes a spherical shape (Figure 2b), being hypercontractile in its basal portion. At this stage, most cases may have a competent mitral valve, but in some cases the movement of the posterior leaflet is restricted, leading to an eccentric mitral regurgitation and passive pulmonary hypertension. A common finding in left EMF is the so-called M-movement of the interventricular septum on M-mode.



**Figure 2 – The classification of EMF captures the diversity in location and severity of structural abnormalities detected on echocardiography.**

A) Grade I predominantly right; B) Grade II bilateral; C) Grade III bilateral with obliteration of the right ventricle; D) Grade IV with severe retraction of the right ventricle, heart distortion and pericardial effusion.

**Table 3 – Frequency of echocardiographic features on the left side of the heart in 411 EMF patients in clinical and community studies performed in Mozambique**

| Echocardiographic features                | Community Studies | Clinical Registry |
|-------------------------------------------|-------------------|-------------------|
| Thickening of the AML                     | 121 (51.3%)       | 148 (84.5%)       |
| Thickening of the PML                     | 117 (49.6%)       | 102 (58.3%)       |
| Endocardial thickening of the septal wall | 106 (44.9%)       | 140 (80.0%)       |
| Restrictive filling pattern               | 81 (34.3%)        | 57 (24.2%)        |
| Enhanced density of the papillary muscles | 78 (33.1%)        | 130 (74.3%)       |
| Apical endocardial thickening             | 55 (23.3%)        | 57 (32.6%)        |
| Thickening of the false tendon            | 47 (19.9%)        | 11 (6.29%)        |
| Obliteration of the recess behind the PML | 27 (11.4%)        | 30 (17.1%)        |
| Mitral regurgitation                      | 26 (11.0%)        | 107 (61.1%)       |
| Severe dilatation of the left atrium      | 13 (5.5%)         | 87 (49.7%)        |
| Apical thrombus/obliteration              | 4 (1.7%)          | 13 (7.4%)         |
| Non-apical intraventricular thrombus      | 1 (0.4%)          | 5 (2.9%)          |
| Left atrial thrombus                      | 0 (0%)            | 7 (4.0%)          |
| <b>Total</b>                              | <b>236</b>        | <b>175</b>        |

PML: posterior mitral leaflet; AML: anterior mitral leaflet

The most characteristic features of advanced left EMF are septal and apical fibrosis, severe eccentric mitral regurgitation, due to fusion of posterior leaflet to the wall, disproportionately small LV, aneurysmal left atrium, and severe pulmonary hypertension (Table 4). Even in patients with extensive endocardial thickening, retraction of the LV apex does not occur.(Figure 2c)

### Abnormalities of the Right Side of the Heart

In early right EMF disease, the longitudinal view of the RV (using the short axis view of the LV at the level of the aorta) shows a stretched moderator band separating the inflow and outflow tracts from the trabecular portion. (Figure 2a) There may be turbulent blood flow inside the trabecular portion of the ventricle. Right ventricular obliteration (disappearance of the trabecular portion of the RV) is usually associated with mild to moderate tricuspid regurgitation caused by restriction to the movement of the tricuspid anterior and septal leaflets. (Figure 2b) The leaflets may be attached to the wall leading to an echocardiographic picture similar to the “Ebstein Malformation”, with dilatation of the tricuspid annulus and tricuspid regurgitation jet originating inside the RV.

Severe right EMF is characterized by retraction of the RV (Figure 2c, 2d), severe tricuspid regurgitation with almost no turbulence, restriction of leaflet motion caused by involvement of the papillary muscles in the fibrotic process, and dilatation of the tricuspid annulus, related to severe right atrial dilatation.

Spontaneous contrast images are common inside the right atrium, extending both to the right ventricular inflow tract and to the inferior vena cava and dilated supra-hepatic veins; these do not usually show the normal respiratory changes, indicating increased systemic venous pressure.

In patients with advanced right EMF, the reduction of the right ventricular cavity size is partially compensated for by dilation of the outflow tract and free tricuspid regurgitation occurs due to an aneurysmal right atrium. There are usually dynamic intracavitary echoes or large thrombi. There may be equalization of pressure between the atrium, the ventricle, and the pulmonary artery, as well as diastolic opening of the pulmonary valve. The aneurysmal right atrium causes heart distortion and compression of the left cavities, hampering evaluation of the mitral valve. Abundant pericardial effusion is common in advanced right EMF (Table 4).

### Bilateral EMF

In established bilateral EMF, the heart has a peculiar aspect of enlarged atria with small ventricles - the “Mickey mouse” heart in the four chambers view - coexistence of mitral and tricuspid regurgitation (Figures 2b & 2c). The reduced right ventricular output and pulmonary perfusion partially reduces the effects of the pulmonary venous hypertension caused by left ventricular disease, favoring a better outcome compared to that of pure left EMF.

## Discussion

The echocardiographic criteria for diagnosis of EMF can be employed for screening, follow-up, and management of EMF in endemic areas. We hypothesize that serial evaluations of early EMF using these criteria would provide better understanding of its natural history. However, epidemiological studies to validate these criteria in follow up studies are difficult to implement in endemic areas.<sup>7</sup> The classification has been used to assess geographic variation in clinical presentations of EMF in India.<sup>9</sup>

The hallmark of established EMF is endocardial thickening in large plaques or patchy distribution, interfering with diastolic function and reducing cardiac output. Ventricular thrombosis or “spontaneous contrast” are frequently seen: in left EMF, ventricular thrombi are mostly apical and may involve the subvalvular apparatus, while the trabecular portion is more affected in right EMF. In this classification, thrombi are attributed a high score because they pose an enormous risk to life and are a major determinant of management and prognosis.<sup>10</sup>

Heart distortion is a characteristic feature of EMF due to the aneurysmal atria and partial or complete exclusion of the right ventricular trabecular portion from the circulation, in cases of ventricular retraction with the characteristic “apical notch” – a distinctive feature of advanced right EMF (Figure 2d).

The criteria discussed in this paper do not include the conventional measures for assessment of left ventricular systolic function because these are difficult to apply in severe and advanced EMF due to marked heart cavity distortion. Evaluation of myocardial function is hampered by the presence

**Table 4 – Echocardiographic characteristics of EMF that allow classification as left, right and bilateral**

| RIGHT EMF                                                                                 | LEFT EMF                                                                                   |
|-------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------|
| • Enhancement of endocardial echo                                                         | • Enhancement of endocardial echo                                                          |
| • Tricuspid Valve apparatus adherent to the mural fibrosis                                | • Plastered down post mitral leaflet                                                       |
| • Obliteration or retraction of RV trabecular portion; apical notch or shrunken RV cavity | • Obliterated LV apex, reduction of the longitudinal diameter, with oval or globular shape |
| • Features of Tricuspid Regurgitation                                                     | • Features of Mitral Regurgitation                                                         |
| • Endocardial calcification on the RV                                                     | • Calcificação endocárdica no VE                                                           |
| • Dilated and hyperdynamic RV outflow tract; paradoxical movement of the IVS              | • M movement of IVS/PW; dilated and hyperdynamic basal portion of the LV                   |
| • Restrictive pattern of RV filling                                                       | • Restrictive pattern of LV filling                                                        |
| • Dilated right atrium                                                                    | • Dilatation of the left atrium                                                            |
| • Variable pericardial effusion                                                           | • Features of Pulmonary Hypertension                                                       |
| • Dynamic intracavitary echoes or thrombi                                                 | • Dynamic intracavitary echoes or thrombi                                                  |
| • Dilatation of the IVC and IHV                                                           |                                                                                            |
| • Pulmonary Valve diastolic opening                                                       |                                                                                            |

EMF: Endomyocardial fibrosis; LV: left ventricle; RV: right ventricle; IVC: inferior vena cava; IHV: intra-hepatic veins; IVS: interventricular septum; PW: posterior wall

of endocardial fibrosis restricting the underlying myocardial contractility. Visual semi-quantitative scales are used to assess systolic ventricular function.<sup>11</sup> Regarding right ventricular assessment, the ventricular outflow tract shortening fraction (a simple and noninvasive measure of systolic function)<sup>12</sup> does not seem adequate, due to the presence of compensatory dilatation and hypercontractility of the outflow tract, as a result of trabecular cavity obliteration or retraction.

Due to the geometric changes associated with EMF, the sphericity index has been suggested as a good tool to describe abnormalities in ventricular shape and volume. This index is used to assess changes occurring in non-ischemic cardiomyopathy and seems to be a good tool to quantify the abnormal geometric changes that accompany heart failure in dilated failing LVs,<sup>13</sup> but it needs to be validated.

Importantly, the standardized criteria for diagnosis and classification of EMF identify patients who can benefit from surgery and allow for risk stratification, defining patients with mild EMF who do not benefit from surgery and those who are unsuitable for surgery. Extensive endocardial thickening with disappearance of the AV valve apparatus, thick endocardium without visible myocardium underneath it, extensive calcification, and/or severe myocardial dysfunction are signs used to define contraindication to surgery. This classification also allows stratification of operative risk, planning of tailored surgical techniques, and post-operative follow up.

Limitations

We acknowledge the limitations of transthoracic echocardiography to accurately assess structural and hemodynamic abnormalities in adult patients with advanced EMF, particularly when calcification or major distortion of the heart cavities are present. Three-dimensional echocardiography

allows better characterization of EMF, but is not widely used in endemic settings. Magnetic resonance imaging and myocardial contrast echocardiography provide more detailed anatomical and functional information, including the location and size of endocardial fibrosis and thrombi and are the techniques of choice for pre- and post-surgery evaluation, particularly for detecting recurrence.<sup>14-16</sup>

In areas endemic for both EMF and rheumatic heart disease, differentiation between left EMF and rheumatic heart valve disease may be challenging; while diffuse irregular AV valve leaflet thickening with thin chordae is suggestive of EMF, prominent thickening of the free borders of the leaflets extending to the chordae is typical of rheumatic valve disease.<sup>17</sup>

Conclusions

The use of echocardiography allows a confident non-invasive diagnosis of EMF, is essential for indication of surgery and choice of operative techniques, and has the potential to be used to define prognosis. EMF presents great phenotypic variability with lesions varying from patchy endocardial thickening without any hemodynamic changes, to extensive mural and AV valve endocardial fibrosis with resulting structural and hemodynamic changes.

Because the assumptions made to calculate parameters for assessment of systolic and diastolic function of both ventricles are not present in a considerable number of individuals with moderate, severe, and advanced EMF, non-conventional measurements and indices have been used to evaluate ventricular function. The standardized criteria for diagnosis and classification of EMF may support better understanding of its pathogenesis and natural history, and allow comparison of affected individuals in different endemic areas.

### Author Contributions

Conception and design of the research, acquisition of data, analysis and interpretation of the data, writing of the manuscript, critical revision of the manuscript for intellectual content: Mocumbi A.

### Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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### Study Association

This study is not associated with any thesis or dissertation work.

### Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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