

# An uncommon picture of endomyocardial fibrosis: no embolism yet

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## ABSTRACT

We present here a review of the various gradings of endomyocardial fibrosis from autopsy (Shaper's types). Echocardiography accurately delineates the extent of fibrosis of either the right or left ventricle and we have illustrated a typical classical case. We have images of the same patient from 2010, 2011 and 2012 and so we use this to illustrate the echocardiographic gradings.

## INTRODUCTION

Endomyocardial fibrosis (EMF) is a disease of unknown aetiology with a short natural history.<sup>1</sup> But recent reports have described longer survival.<sup>2–4</sup> We have recently reported that warfarin prolongs survival in patients with EMF.<sup>2</sup> A report has even called it a vanishing entity.<sup>5</sup> A Geochemical basis was thought to be the cause of EMF.<sup>6–8</sup> It was hypothesised that a reduced magnesium level in the soil leads to more absorption of cerium which leads to EMF. This was studied in plant tissue culture which showed that there was an increase in cerium entering the tubers that are grown on magnesium deficient medium.<sup>9</sup>

## Clinical case

A 38-year-old woman who developed recurrent swelling of the abdomen has been on our follow-up for 4 years. On ascitic tap, she was found to have leucocytosis and was treated with intravenous metronidazole and cefotaxime. On follow-up, she developed atrial fibrillation (AF) and a fast ventricular rate and was put on digoxin. Subsequently, she developed AF with complete heart block and so digoxin was stopped. As she had a large right atrium (RA) thrombus she was put on long term warfarin. She was also put on diuretics, including metolazone, furosemide and spironolactone.

Throughout the period of follow-up, she had serial two dimensional and Doppler echocardiograms. Her initial echocardiogram showed obliteration of the entire right ventricle and massive right atrial dilatation. The left atrium was dilated but the left ventricle appeared normal (figure 1). Her subsequent echocardiograms showed the presence of a thrombus in the right atrium which remained relatively unchanged on follow-up (figures 2 and 3). The massive cardiomegaly (figure 4) and myocardial calcification were visible on the chest x-ray. Her videos show her heart in 2010, her right atrial thrombus in 2011 and another view on 2012 (see online supplementary videos 1–3).

## DISCUSSION

Various authors have graded the angiographic stages of EMF.<sup>10</sup> To summarise their findings, right ventricle (RV) EMF has been graded by angiography as grade I: minimal involvement of the RV chamber in the form of alterations in the trabecular pattern of the apex and along the septal border with small filling defects; grade II: obliteration of the RV apex and body but not extending up to the tricuspid annulus; and grade III: obliteration of the RV endocardium, including the area near the tricuspid annulus but sparing the RV outflow.

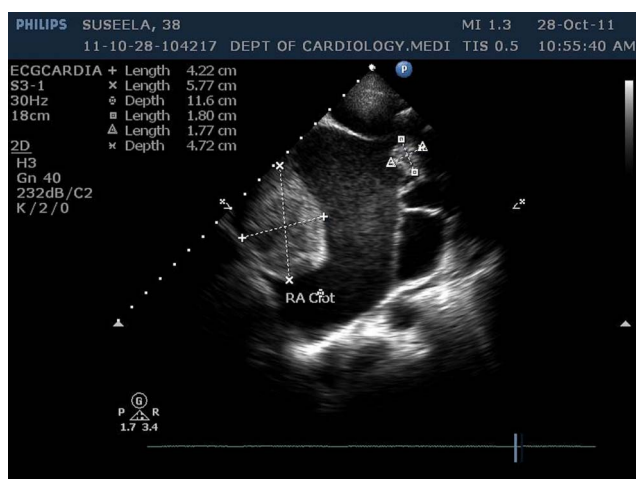
Various workers have described the echocardiographic picture of EMF.<sup>11 12</sup> However, an echocardiographic grading is not commonly known.<sup>12</sup> Dr Vijayraghavan<sup>13</sup> is one of the first authors to describe the echocardiographic features of RV and left ventricle (LV) EMF. In RV EMF, classically the RA was massively dilated. The interatrial septum tended to bulge to the left. In addition, the tricuspid annulus is in a normal position (unlike Ebstein's anomaly). The tricuspid valve appeared tethered to the endocardium in many patients and thick fibrosed endocardium is usually seen in the inflow region. EMF classically involves the inflow and trabecular regions of the RV or LV and the outflow regions are classically spared. Usually, a dimple is seen at the right ventricular apex. More recently, the merlon sign has been described in RV EMF.<sup>11</sup> This is a hyperactive, hypercontractile base of the heart with no movement of the apex due to obliteration.

Involvement of the mitral and tricuspid valves usually causes mild mitral regurgitation or tricuspid regurgitation. The Doppler of the tricuspid valve and mitral valve showed a restrictive pattern with a



**Figure 1** An echocardiogram in 2011 showing the presence of a large thrombus in the right atrium.

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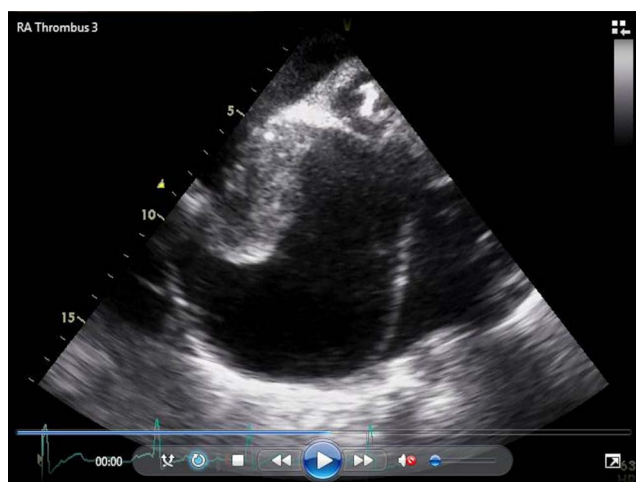


**Figure 2** The right atrial thrombus remains relatively unchanged in 2011.

markedly short deceleration time. The pulmonary veins showed a marked diastolic D wave and a broad reversal of A wave. The hepatic veins showed a deep diastolic forward wave and a marked reversal in inspiration.

Another classification is described as Shaper's types.<sup>14</sup> Under this classification, EMF is classified as follows: Type 1: involving only the apex of the ventricle, R1 for right and L1 for left; type 2: obliteration from apex to valve; type 3: valvular involvement only; type 4: separate involvement of the valve and apex with a free endocardial area in between; and type 5: patchy ventricular involvement. It is also notable that in Shaper's autopsy study, there was an increase in the heart weight due to left ventricular hypertrophy. Camara and colleagues<sup>12</sup> have graded EMF by echocardiography for the purpose of comparing patients evaluated by echo and by angiocardiology. For this classification, the heart was assessed in diastole: grade 1 or 1+ for obliteration of the apical region only, 2+ for those with less than 50% obliteration, and 3+ for those with greater or equal to 50% obliteration from apex to the atroventricular level. Based on this grading, our patient had a 3+ grade.

As can be seen in the online supplementary videos 1-3 and figure 2, our patient has Shaper (14), type 2 or R2 type of EMF.



**Figure 3** Follow-up echocardiography in 2012. The dilated right atrium shows a large right atrial thrombus that has not changed much over the years.



**Figure 4** A recent 2012 chest x-ray of the patient showing calcification within the enlarged cardiac shadow.

In figure 2, a large right atrial thrombus can be seen. Overall, the right atrium is massively dilated and the RV is fully obliterated. After being put on chronic warfarin treatment, she has not developed any embolism. In the study of Subair *et al*,<sup>2</sup> warfarin use was associated with better survival on multivariate analysis.

We provide some supplementary figures also (online supplementary figures S1–6) which all show the patient's images in 2012, 2011 or 2010. Online supplementary figures S3 and S4 show an additional small thrombus near the tricuspid valve in 2011. Online supplemental figures S7–S10 are recent MRI pictures of the patient with contrast.

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