

Echocardiographic Recognition of Chagas' Disease and Endomyocardial Fibrosis

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Chagas' heart disease and endomyocardial fibrosis are common medical conditions in Central and South America but are only rarely encountered in North America. In the small number of patients who have these conditions, recognition is frustrating because of a lack of familiarity with their characteristic echocardiographic pattern. In Chagas' heart disease a left ventricular apical aneurysm is characteristic, but in contrast to coronary artery disease, septal involvement is minimal. In endomyocardial fibrosis apical obliteration with a small inwardly moving left ventricular cavity, large atria, and atrioventricular valvular insufficiency are typical features. It is the aim of this article to present the characteristic echocardiographic findings with these conditions and thereby facilitate the recognition when they appear in nonendemic areas. (J AM SOC ECHO 1988;1:60-8.)

Although the number of patients with chronic Chagas' heart disease and endomyocardial fibrosis is very low in North America, the differentiation from other forms of cardiomyopathy is important. Migration of persons from countries where these diseases are more prevalent may pose diagnostic difficulties for the echocardiographer who must differentiate them from ischemic heart disease or other cardiomyopathies. The purpose of this article is to outline typical findings of each entity.

CHAGAS' DISEASE OF THE HEART

In South and Central America more than 20 million persons have been exposed to infection with *Trypanosoma cruzi*, and about 10 million have its sequela, Chagas' cardiomyopathy.¹ It is a continuing and formidable public health problem because the disease cannot be eradicated from its silvatic mammal reservoirs. Serologic tests afford highly sensitive and specific recognition of infected persons.² Of those seropositive results, about 75% are asymptomatic (group A), and most will not develop serious heart disease. The remaining 25% are symptomatic and

usually come to medical attention because of a dysrhythmia or an isolated embolic episode (group B), congestive heart failure (group C), or a combination of these. The clinical presentation of group C is often as a dilated cardiomyopathy. Increasing age and male sex are associated with more severe forms of the disease. Risk factors for the development and advancement of heart involvement is poorly understood; why some persons have more serious manifestations than others with similar living conditions is unknown.

Traditionally the diagnosis of chronic Chagas' heart disease rests on (1) coming from an endemic area, (2) positive serologic results, and (3) highly suggestive electrocardiographic findings that include right bundle branch block and left anterior hemiblock (either isolated or combined), sinus bradycardia, partial or advanced atrioventricular block, and polymorphic ventricular extrasystoles.³ Left bundle branch block is uncommon.

Over 50% of autopsy results from persons with Chagas' heart disease show a peculiar left ventricular apical aneurysm or thinning^{4,5} that is not caused by coronary heart disease. Therefore apical two-dimensional echocardiographic views are especially suited for the detection of this particular finding.⁶ We used the following criteria to classify and identify the apical wall motion abnormalities: we considered an aneurysm to be present if there were a discrete sacular area that exhibited systolic outward dyskinetic motion with clear hinge points and nonaneurysmal

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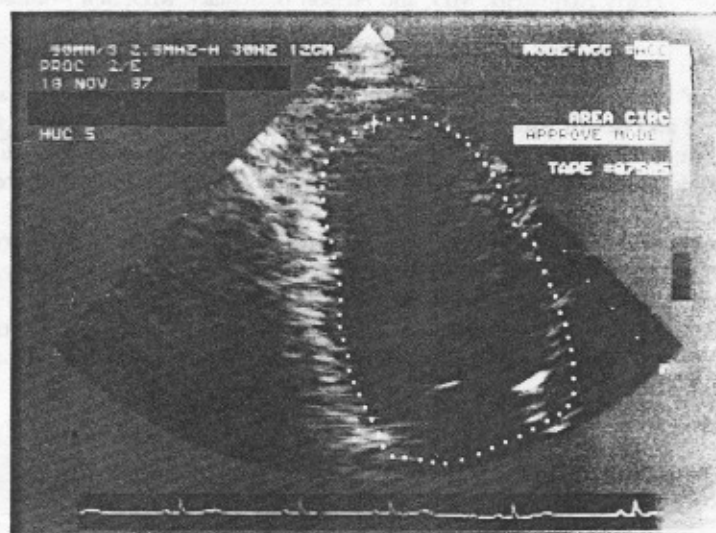


Figure 1 Left ventricular apical dyskinesia. Four-chamber apical view of asymptomatic 28-year-old man (group A). Condition detected because on blood donation positive Chagas' serologic results were found. Left ventricular size and ejection fraction were normal. Note normal electrocardiogram. Apical systolic bulging "without neck" is present. Dotted outline represents end-diastolic endocardial border. Lesion is found in most asymptomatic persons. It is necessary to manipulate transducer in multiple positions to properly detect subtle systolic abnormality.

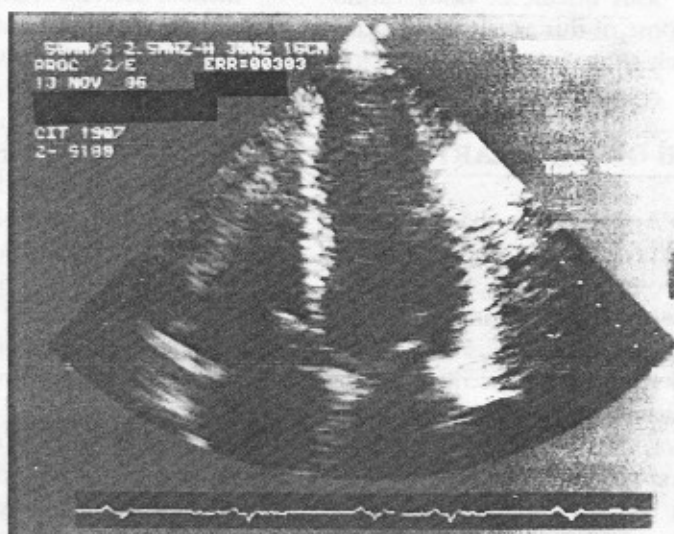


Figure 2 Typical "small neck" apical aneurysm. Man aged 47 years had frequent bouts of ventricular tachycardia (group B) but was otherwise able to perform vigorous exercise. Although adjacent ventricular walls were normal, careful scrutiny of posterior wall detected extension of apical dyskinesia. More than half of symptomatic patients may have either apical dyskinesia or aneurysm. We have not encountered "small neck" apical aneurysm in coronary heart disease.

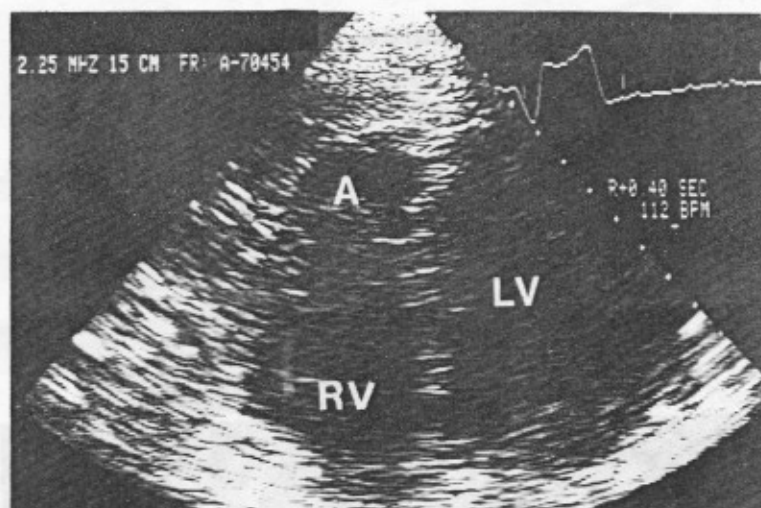


Figure 3 Right ventricular apical aneurysms. Patient aged 43 years complained of dyspnea on moderate exertion and had frequent premature ventricular contractions (group B). Prominent right ventricular (RV) apical systolic bulging (A) was detected on apical four-chamber view, lesion observed in about 10% of symptomatic patients. RV ejection fraction was also moderately diminished. No thrombi were observed. LV, Left ventricle.

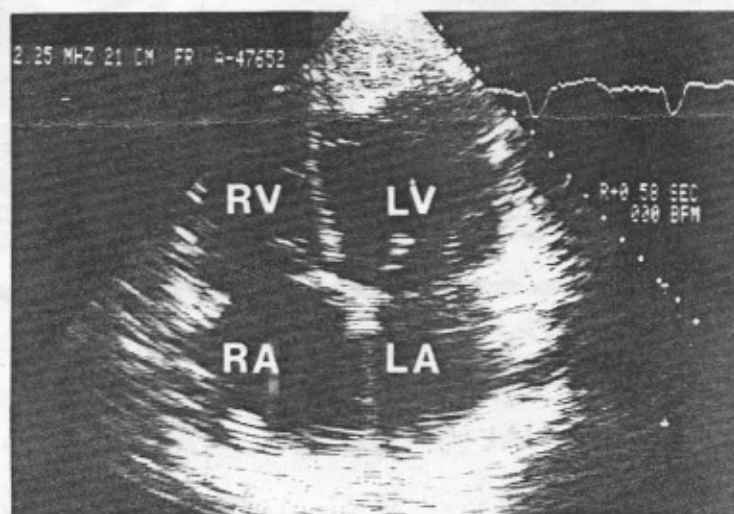


Figure 4 Congestive heart failure and systemic embolism. Apical four-chamber view from 54-year-old patient with biventricular congestive heart failure (group C) demonstrating biventricular severe, diffuse hypokinesis. Large, somewhat mobile thrombus is seen fixed to "large neck" LV apical aneurysm. Size of aneurysm is similar to those encountered in ischemic heart disease. Both atria (RA and LA) are also dilated. Patient had embolic event resulting in right-sided hemiplegia. Electrocardiogram showed typical combination of right bundle branch block and left anterior hemiblock.

apical dyskinesia if the wall motion abnormality was subtle without apical remodeling. We found in asymptomatic persons that about 10% to 15% may have apical dyskinesia (Figure 1), whereas about 50% to 60% of symptomatic persons may have left ventricular apical dyskinesia or aneurysm (Figures 2 and 3). Right ventricular aneurysm is found in about

10% (Figure 4). Laminar fixed or mobile thrombi are also found at the apical lesion and are the source of systemic embolism. We have not yet observed an apical aneurysm in an asymptomatic person with seropositive results. Many theories have been proposed to explain the apical lesion including ischemia caused by small vessel disease, apical muscle verticil-

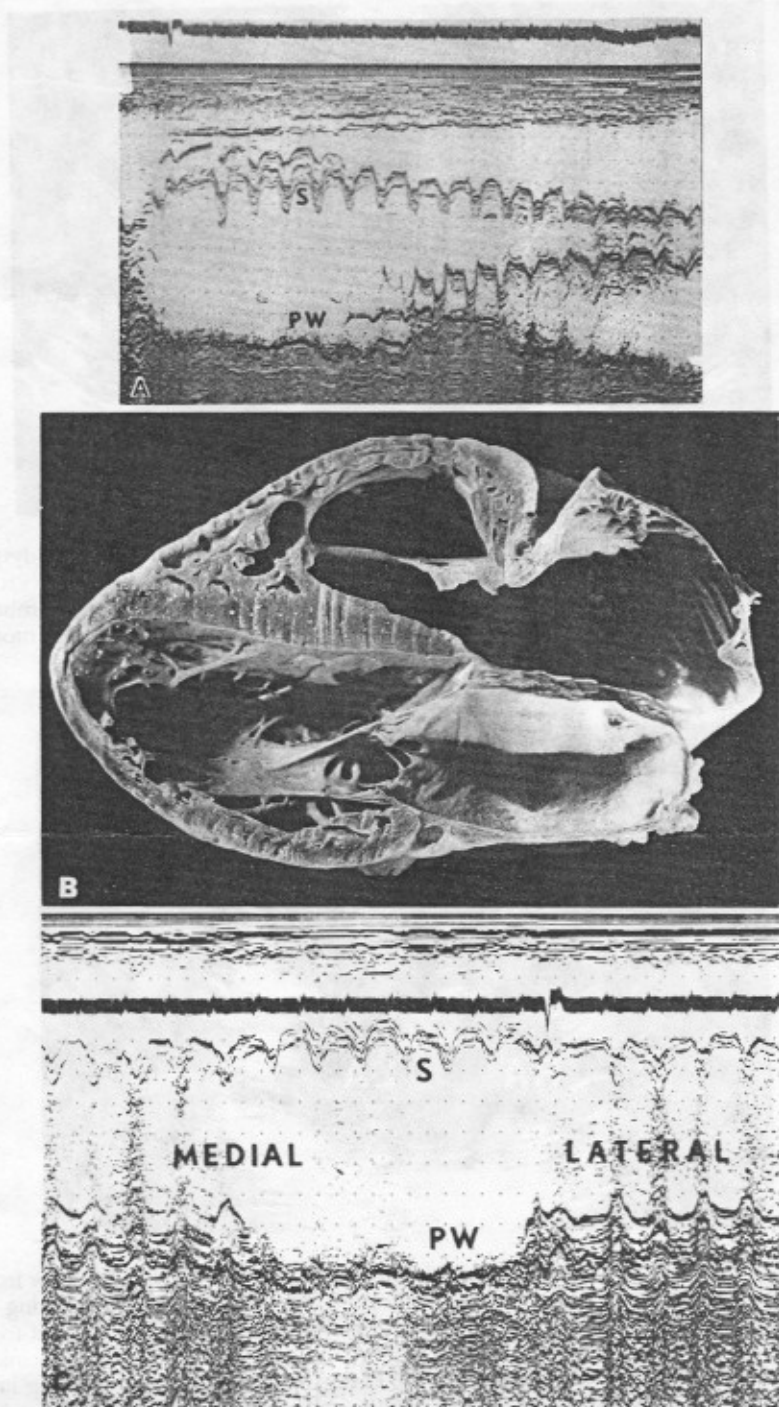


Figure 5 Postmortem specimen and echocardiograms from patient who had congestive heart failure caused by chronic Chagas' heart disease, showing echocardiographic pattern of segmental posteroapical hypokinesis observed in nearly half of our symptomatic patients. **A**, Long-axis, slow-sweep echocardiogram showing relatively preserved systolic septal (*S*) motion and thickening and thin, noncontractile posterior wall (*PW*). **B**, Long-axis autopsy section of heart. Left ventricular apical and posteroapical thinning and fibrosis with relative septal sparing. Coronary arteries were normal. **C**, Short-axis, M-mode T scan depicting lateral extent of posterior dyskinesia and thinning. Scanning technique enhances effectiveness of M-mode echocardiography by allowing exploration of extent of dyskinetic areas. (From Acquatella H, Schiller NB, Puigbo JJ, et al. *Circulation* 1980;62:790. Reprinted with permission of the American Heart Association.)

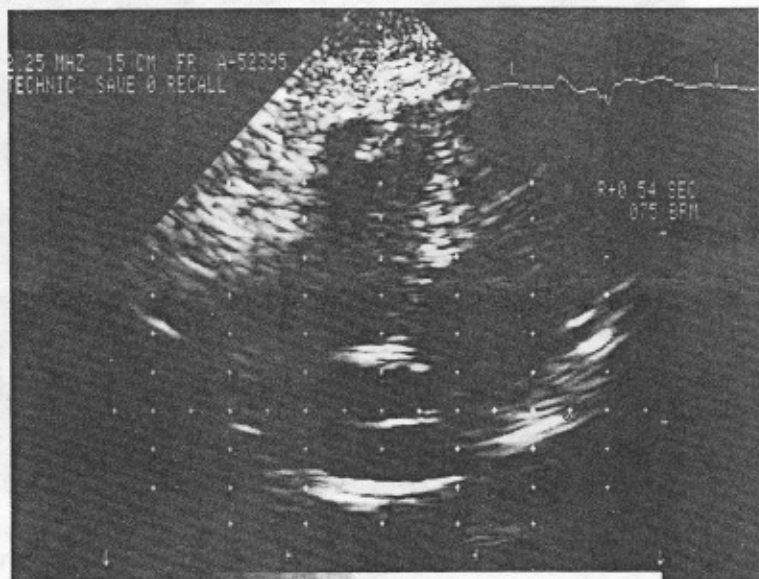


Figure 6 Right ventricular apical thrombus in acute eosinophilic myocarditis. Woman aged 42 years had an acute hypereosinophilic syndrome on September 1981 with biventricular heart failure caused by eosinophilic myocarditis. She also had pulmonary infiltrates and mononeuritis multiplex. She had dramatic response to steroid therapy, which improved her heart failure to New York Heart Association class I. Nevertheless, she developed right ventricular apical obliteration because of fibrosis 11 months later (see Fig. 7).

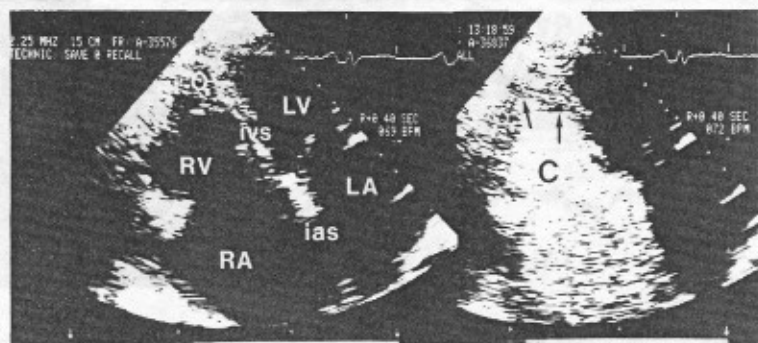


Figure 7 Apical four-chamber view, taken 11 months later, showing right ventricular apical obliteration in same patient as Fig. 6. Changes developed, despite excellent clinical response to oral prednisone. *Left panel* shows RV apical obliteration before and after (*right panel*) agitated intravenous saline solution contrast (C) delineated RV cavity and "horseshoe" apical lesion. IVS and LAS, Interventricular and interatrial septum. Other abbreviations are given in previous figures. (From Acquatella H, Schiller NB, Puigbo JJ, Gomez-Mancebo JR, Suarez C, Acquatella G. *Circulation* 1983;67:1223. Reprinted with permission of the American Heart Association.)

lated disarray, uncoordinated left ventricular contraction, and parasympathetic heart denervation. Other regional left ventricular motion abnormalities may be encountered in 15% to 20% of symptomatic persons at the posteroinferior wall and far less frequently in the lateral wall. In sharp contrast to ischemic heart disease, septal scar is very rarely found

(Figure 5). The reasons for this peculiar distribution of regional wall motion abnormalities are unknown. In those patients with advanced heart failure, global severe biventricular hypokinesia and dilation are found.

About 20% of these symptomatic patients with seropositive results for Chagas' disease may have

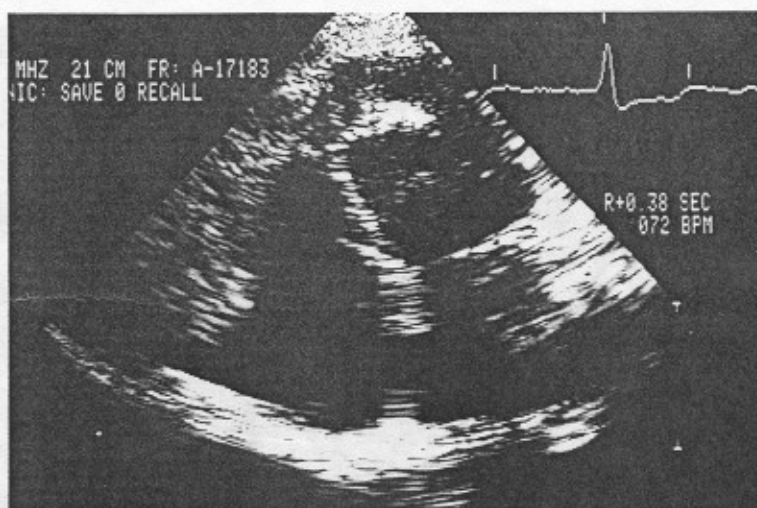


Figure 8 Left ventricular apical obliteration in endomyocardial fibrosis. Bright echoes are present, delineating apical lesion in 47-year-old man. Left ventricular ejection fraction was normal. Combination of normal or slightly increased ventricular dimensions with dilated atria caused by filling restriction and atrioventricular valve regurgitation gives typical echographic appearance.

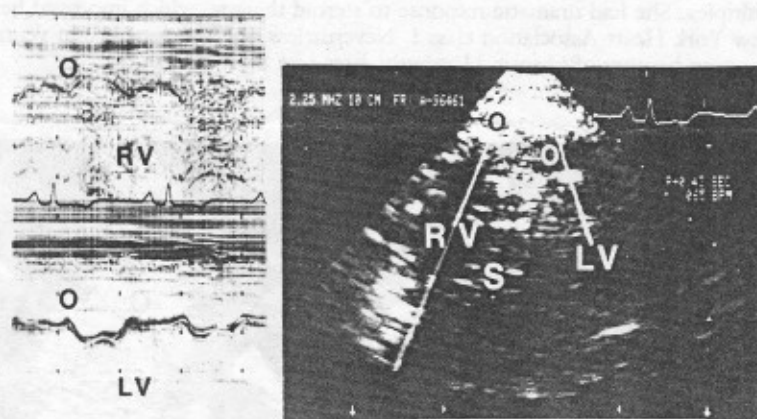


Figure 9 Dual apical M-mode sector scan of both ventricles in endomyocardial fibrosis. Right panel shows four-chamber apical view of patient with biventricular endomyocardial fibrosis, showing bright echoes of biapical obliteration (O). Within each sector M-mode cursor was directed to produce M-mode tracings at left. Left upper panel is from RV, and left lower panel, LV apex. With electrocardiography one can appreciate paradoxical systolic bulging of RV apex, whereas LV apex shows preserved inward systolic motion. All our patients had preserved, if not hypercontractile LV inward systolic motion, whereas RV apical motion was either normal or dyskinetic.

atypical chest pain. In older subjects, differentiating the pain that originates from Chagas' disease from that of coronary heart disease may warrant coronary angiography. Similarly, patients with signs and symptoms of congestive heart failure and an echocardiographic appearance that is indistinguishable from idiopathic congestive cardiomyopathy also pose diagnostic difficulties.

ENDOMYOCARDIAL FIBROSIS

It is now recognized that Löffler's parietal fibroplastic endocarditis with eosinophilia of temperate countries and endomyocardial fibrosis from tropical countries belong to the same disease process.⁷ The echocardiographic appearance will depend on how advanced the disease is when the patient is studied. Initially,

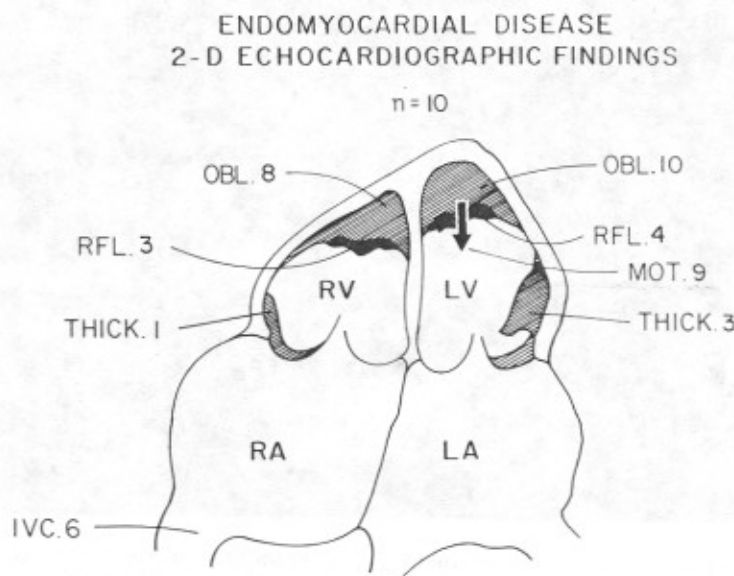


Figure 10 Summary of findings from four-chamber apical views in patients with endomyocardial fibrosis. Numbers along right and left indicate number of patients with each finding. IVC, Inferior vena cava, dilated; MOT, preserved or increased inward motion of oblative process; OBL, obliteration; RFL, increased reflectance of oblative surface; THICK, increased thickening of basal inflow tract papillary muscle and/or posterior atrioventricular valve. Other abbreviations are given in preceding figures. (From Acquatella H, Schiller NB, Puigbo JJ, Gomez-Mancebo JR, Suarez C, Acquatella G. *Circulation* 1983;67:1225. Reprinted with permission of the American Heart Association.)

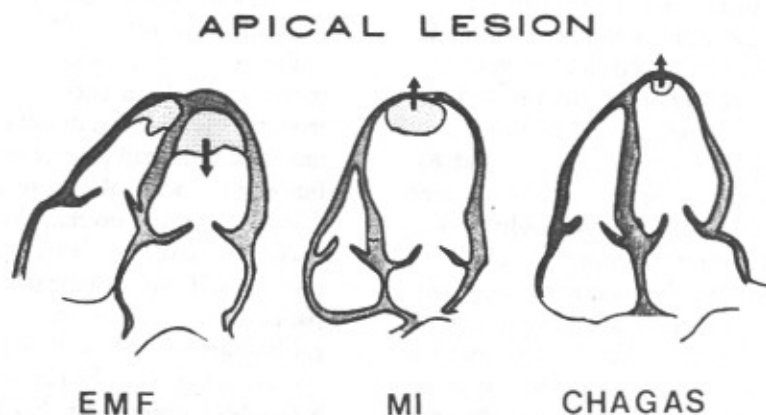


Figure 11 Differences in the apical lesion between endomyocardial fibrosis (EMF), myocardial infarction (MI), and Chagas' heart disease. Both apices can be affected in EMF, but typically LV function is preserved or hypercontractile, and LV obliteration moves inward. In apical MI, dyskinetic apex frequently is combined with septal or anterior wall motion abnormalities, depending on extent of disease of left anterior descending coronary artery. "Neck" of dyskinetic area tends to be large. In chronic Chagas' disease, although apical aneurysm can be as large as in ischemic heart disease, several patients may have typical "small" neck aneurysm. When apical dyskinesia without aneurysm is found, its appearance cannot be used to differentiate between ischemic or Chagas' disease. Chagas' disease with isolated apical aneurysm typically spares all but most apical motion of septum. Finally, in Chagas' disease, RV apical dyskinesia or aneurysm may also be present.

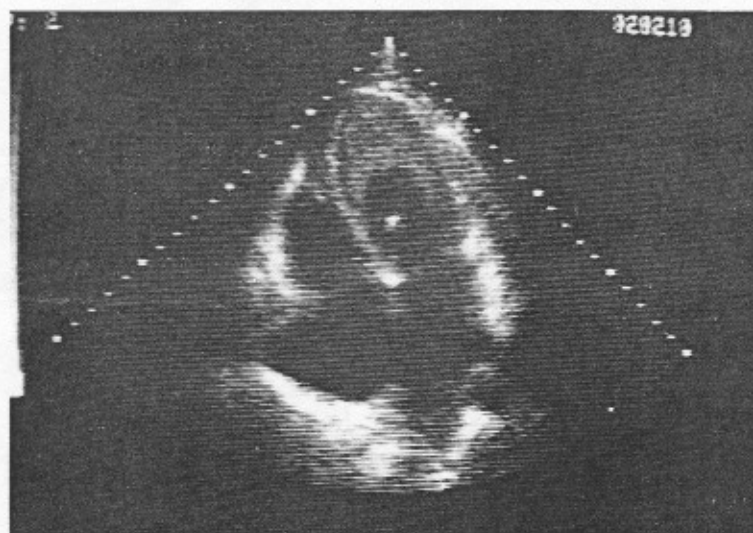


Figure 12 Endomyocardial fibrosis may occur spontaneously in North America. Images are from 56-year-old San Francisco Bay man who had never been outside North America. He had subacute onset of biventricular failure and eosinophilia. Echocardiogram presented typical picture of LV apical obliteration. Contrast injection (agitated saline solution) showed right ventricular apical obliteration also. Doppler examinations showed moderate mitral regurgitation, severe tricuspid regurgitation, and pulmonary hypertension. Surgical resection was attempted but failed. Patient was referred by Drs. Eugene Shafton, Brian Strunk, Kent Gershengorn, and Mark Wexman, San Francisco.

there is an acute eosinophilic myocarditis (necrotic phase) that lasts a few weeks.⁸ This is followed by thrombus formation, usually at the apex (thrombotic phase), which lasts a few months (Figure 6). Finally, a healing and scarring process ensues for years (fibrotic phase), which typically obliterates one or both ventricular apices and extends basally to the inflow portion of the ventricular wall (Figures 7 and 8). Left ventricular function is preserved, and the area of apical obliteration typically shows bright echoes from the endocardial surface, with preserved inward systolic motion (Figure 9). Frequently the posterior atrioventricular valves and their subvalvular apparatuses also become involved,^{9,10} which leads to massive mitral and/or tricuspid regurgitation. The combination of the above findings with huge dilated atria and normal or slightly enlarged hyperdynamic ventricles leads to a typical two-dimensional echocardiographic appearance (Figure 10). At this stage the patient has the clinical and hemodynamic picture of restrictive cardiomyopathy. The presence in the peripheral blood of degranulated eosinophils¹¹ or myocardial eosinophilic infiltrates on biopsy is important, since they suggest the potential for the use of chemotherapy with antineoplastic agents or steroids to

retard or halt the evolution of the process.¹² In the fibrotic phase apical endocardectomy with atrioventricular valve replacement may substantially improve the patient's condition.¹³

Figure 11 depicts a summary of the apical findings presented by both entities and their differentiation from the apical lesion of ischemic heart disease. Chagas' disease probably never occurs in North America, but endomyocardial fibrosis does occur on rare occasions. At the University of California, San Francisco, we have seen three individuals with endomyocardial fibrosis in the past 10 years. Two of these patients came to us in the past 12 months. The echocardiogram of one of these, a 56-year-old man, is shown in Fig. 12. The patient had always worked as a ferryboat captain on San Francisco Bay and had never traveled outside the continental United States. He went to a local cardiology group with the subacute onset of biventricular failure and eosinophilia. His echocardiogram demonstrated the typical picture of biventricular apical obliteration and considerable biatrial dilation. Because of his rapidly deteriorating clinical condition and a lack of active infiltration on endocardial biopsy, it was elected to refer the patient for surgery. Apical decortication proved to be im-

possible because of the density and extent of the apical scar, and the patient died from severe congestive heart failure a few weeks after surgery.

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