Endomyocardial fibrosis: Progression to restricted ventricles and giant atria

Eloi Marijon MD^{1,2,3}, Dinesh Jani MD², Phalla Ou MD³

First described in 1948 by Davies (1), endomyocardial fibrosis (EMF) remains of unclear etiology. It predominates in tropical and subtropical areas, affects mainly children and young adults of low socioeconomic status, and is recognized as an important cause of heart disease in Africa, India and Brazil (2). EMF is a severe and progressively restrictive form of cardiomyopathy. Although eosinophilia is seen in most cases of EMF during the initial inflammatory process and histological endomyocardial lesions are similar to the hypereosinophilic syndrome observed in industrialized countries, eosinophil cell toxicity as a cause is not accepted by all authors (3). Echocardiography is now the most reliable tool for the assessment of the progress of disease.

At the onset, EMF is characterized by the formation of fibrous tissue in the endocardium, with patchy enhancement of endocardial echoes (Figure 1A). It may be present in both ventricles, but is usually predominant in the right ventricle, with apical obliteration (Figure 1B). Moreover, the initial inflammatory process may support endomyocardial thrombus formation. This first stage results in ventricular diastolic dysfunction of one or both ventricles, with valvular involvement. The totally fused aspect between the posterior papillary muscle, the posterior leaflet and the inferior left ventricular wall remains a useful specific aspect feature (Figure 1C) in distinguishing EMF from rheumatic heart disease.

Figure 1 A,B Apical (four-chambered) view, with fibrotic process (asterisk). C Parasternal (long-axis) view illustrating the characteristics of the mitral valve. D,E Visualization of auriculoventricular valve regurgitation in the apical four-chambered view. Ao Aorta; LA Left atrium; LV Left ventricle; RA Right atrium; RV Right ventricle

¹Service de Cardiologie, Hôpital Européen Georges Pompidou, Paris, France; ²Instituto do Coração, Maputo, Moçambique; ³Service de Cardiologie Pédiatrique, Hôpital Necker Enfants-Malades, Paris, France

Correspondence: Dr Eloi Marijon, Service de Cardiologie, Hôpital Européen Georges Pompidou, 20, rue Leblanc, 75908 Paris Cedex 15, France.

Telephone 33-1-56-09-37-31, fax 33-1-56-09-37-29, e-mail eloi_marijon@yahoo.fr

Received for publication December 20, 2005. Accepted December 28, 2005

Figure 2) **A** Modified parasternal (long-axis) view showing right-sided cavities and tricuspid valve. **B** Four chambers demonstrating the disorganized heart architecture and severe spontaneous contrast in the giant right atrium (RA). **C** Standard apical (four-chambered) view showing restricted ventricles with dilated atria. LA Left atrium; LV Left ventricle; PE Pericardial effusion; RV Right ventricle

The diastolic dysfunction, and tricuspid and/or mitral regurgitation (Figure 1D,E) result in an elevation of atrial pressures, with secondary marked atrial enlargement (Figure 2A). At a later stage, the right ventricular fibrosis leads to restriction of the ventricles, with cavity obliteration, and to reduction of the distance between the apex and tips of the tricuspid leaflets and the giant right atrium; this results in severe spontaneous contrast and totally disorganized architecture (Figure 2B). In the case of predominant left ventricular disease, an enormous left atrium with high pulmonary artery pressures are present (Figure 2C). Pericardial effusion is almost always present (Figure 2B). Chronic thromboembolic events

occurring from the right atrium may participate in the development of pulmonary hypertension and sudden death.

EMF remains a mysterious and misunderstood disease, with many extraordinary echocardiographic features.

REFERENCES

1. Davies JNP. Endomyocardial fibrosis. A heart disease of obscure aetiology in Africans. *East Afr Med J* 1948;25:10-4.
2. Kutty VR, Abraham S, Kartha CC. Geographical distribution of endomyocardial fibrosis in south Kerala. *Int J Epidemiol* 1996;25:1202-7.
3. Urhoghide GE, Falase AO. Degranulated eosinophils, eosinophil granule basic proteins and humoral factors in Nigerians with endomyocardial fibrosis. *Afr J Med Med Sci* 1987;16:133-9.