Case 1

A 30 year-old man presented with dyspnea and palpitations. The electrocardiogram showed 2:1 atroventricular block (AVB) that alternated with complete AVB with a heart rate of 35 beats per minute. Echocardiography revealed the presence of dilated cardiomyopathy with severe left ventricular dysfunction and ventricular septal thickening. Coronary angiography showed absence of lesions in the coronary arteries. Endomyocardial biopsy (EMB) documented sarcoid-like granulomas with epithelioid cells and Langhans giant cells. Other granulomatous diseases such as tuberculosis and deep mycosis were ruled out. Chest computed tomography and spirometry tests were normal. A definite pacemaker was implanted and high doses of corticosteroids were indicated. Three months after the initial diagnosis, the echocardiogram showed progressive ventricular dilation. The patient was hospitalized several times due to heart failure despite therapy with beta blockers, angiotensin-converting enzyme inhibitors (ACEIs) and spironolactone. He died 10 months after the diagnosis of CS due to sustained ventricular tachycardia unresponsive to electrical cardioversion.

Case 2

A 45 year-old man with palpitations and a history of
pulmonary sarcoidosis and neurosarcoidosis was evaluated with 24-hour Holter monitoring which evidenced first degree AV block with right bundle branch block alternating with complete AV block and pauses longer than 2 seconds. The initial echocardiogram was normal. A definite pacemaker was implanted. An echocardiogram performed 3 months later showed dilated cardiomyopathy with severe left ventricular dysfunction and severe functional mitral regurgitation. Coronary angiography showed absence of lesions in the coronary arteries. Therapy with ACEIs, beta blockers and high doses of corticosteroids was initiated. Twelve months later ventricular diameters had reduced and systolic function improved. He has not been rehospitalized and remains in good functional class 15 months after the diagnosis of CS.

Case 3
A 24 year-old woman with a history of pericarditis presented with syncope due to sustained ventricular tachycardia. Baseline electrocardiogram showed first degree AVB with abnormal Q waves and ST elevation in the inferior leads. Coronary angiography showed absence of lesions in the coronary arteries. The echocardiogram revealed severe biventricular dysfunction and hypokinesia particularly in the inferior wall. The EMB revealed the presence of interstitial and subendocardial granulomas with giant cells surrounded by fibrosis, suggestive of CS (Figure 1). She was treated with ACEIs, amiodarone and high doses of corticosteroids. After 18 months the echocardiogram revealed improvement of the systolic function in both ventricles. The patient has not been rehospitalized and remains free of symptoms 16 years after the diagnosis.

DISCUSSION
CS may present with a variety of symptoms; the most frequent presentations are severe conduction abnormalities in young patients, complex ventricular arrhythmias and dilated cardiomyopathy, as in the cases here presented. The diagnostic yield of EMB has been reported to be as low as 20% because of the patchy distribution of the disease, and granulomas and fibrotic scars are not mainly located in the right ventricular where the samples are taken. (5) In the present series we have documented lesions suggestive of CS in two patients with consistent clinical signs. According to The Japanese Ministry of Health and Welfare guidelines from 1993, the diagnosis of CS can be made using histological and clinical criteria (Table 1). (6) Although spontaneous resolution of the disease is observed in 2 / 3 of patients in the first 3 years after diagnosis, CS, as other forms of extrapulmonary disease, is associated with adverse outcomes. Mean survival is two years in patients who are receiving treatment with corticosteroids, and five-year survival is 40% to 60% in those under treatment. Low functional status, the presence of dilated cardiomyopathy and sustained ventricular arrhythmias are predictors of mortality. Cardiac involvement is the main cause of death after ventilatory failure, and the incidence of sudden death due to complete AV block or ventricular arrhythmia is high. (1, 2, 7)

Despite treatment with corticosteroids has not been evaluated in large randomized controlled studies, several series recommend their indication as first line therapy in order to prevent progression of inflammation and to improve the prognosis of the disease, particularly when it is initiated in
early stages, as in cases 2 and 3. (8) Early implant of devices is recommended in cases of CS with conduction disturbances or ventricular arrhythmias. The use of these devices has reduced the incidence of sudden death reported in previous series, and heart failure is currently the most frequent cause of death in patients with CS. (9)

Heart transplantation should be considered in patients with dilated cardiomyopathy and/or refractory ventricular arrhythmias. CS should not discourage the indication as recurrences are infrequent. (10)

Despite advances in the understanding of the pathophysiology and the use of high-tech in the diagnosis and treatment of CS, there are no conclusive data in the Latin American population. In this sense, prospective studies and updated consensus guidelines are necessary to assist the cardiologist in diagnostic and therapeutic decision-making in clinical practice.

RESUMEN
Sarcoidosis cardíaca: descripción de tres casos

La sarcoidosis es una enfermedad multisistémica de etiología desconocida que puede afectar cualquier órgano. Presenta considables diferencias en su prevalencia según los distintos países y razas. Diversas comunicaciones sugieren que el compromiso cardíaco determina mal pronóstico. Existen pocas series clínicas de sarcoidosis cardíaca en Latinoamérica y no hay consenso claro acerca de los aspectos diagnósticos y terapéuticos de esta patología. En esta presentación se describen tres casos de sarcoidosis cardíaca, sus características clínicas, electrocardiográficas, métodos de imágenes y, en dos de ellos, la anatomía patológica del material obtenido mediante biopsia endomiocárdica. Se refere, asimismo, la evolución final de los pacientes durante el seguimiento.

Palabras clave > Sarcoidosis - Granuloma - Arritmia cardíaca - Insuficiencia cardíaca

BIBLIOGRAPHY