Pacemakers and Defibrillators in Patients With Cardiac Sarcoidosis

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Background: The prognosis associated with cardiac sarcoidosis remains controversial. Additionally, the use of pacemakers (PM) and defibrillators (ICD) in this population has not been fully evaluated.

Methods: We describe 10 consecutive patients (9 women; 7 African-Americans; mean age 2.11 years) with biopsy-proven systemic sarcoidosis and presumed sarcoid of the heart. At admission they had a PM and/or an ICD. Results: These patients had a mean left ventricular ejection fraction (LVEF) of 0.58 ± 0.09 (0.58 ± 0.05 in the ventricular tachycardia [VT] population) and have carried a diagnosis of cardiac sarcoidosis for 80 ± 38 months. The indications for device implantation were complete heart block in 6 patients, sustained VT in 2 patients, and both complete heart block and VT in 2 patients. In addition, 4 patients had paroxysmal atrial flutter. As a result of these arrhythmias, 6 patients required a PM, 3 patients required an ICD, and one patient had both a PM and an ICD. For patients with a PM, the initial pacing and sensing thresholds were excellent and did not change significantly over a mean follow-up of 86 months. For ICD patients, the defibrillation threshold at implant was 16.5 ± 6.6 joules. One ICD patient went to multiple shocks for VT, while the other three patients had no recurrent VT. All patients are currently alive and asymptomatic from a cardiac standpoint.

Conclusions: (1) The prognosis for device patients with cardiac sarcoidosis is often very good and better than portrayed in the earlier literature. (2) Both PMs and ICDs can function safely in this population. Despite the invasive nature of the disease, (3) VT patients often have preserved systolic function. (4) The clinical course frequently stabilizes over time.

Impact of Microangiopathy on Left Ventricular Remodeling and Clinical Outcome in Cardiac Sarcoidosis Patients Treated With Corticosteroids. A Long-Term Echocardiographic Follow-Up Study

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Background. Microangiopathy presenting as basal laminar layering around the muscularis (SLL) detected by electron microscopy is often observed in various tissues affected by sarcoidosis, including the heart. Although microangiopathy may be related to disease process in sarcoidosis, the clinical significance is unknown in cardiac sarcoidosis (CS). Methods: To investigate the clinical significance of microangiopathy, we analyzed the clinicopathological findings and long-term outcome in consecutive 16 CS patients in whom invasive structural evaluation was performed. Symptomatic improvement or clinical stability was confirmed histologically in all patients from the lung, skin, lymphnode or heart. We also underwent serial echocardiographic assessment over 5 years. Results: Granulomas were confirmed in only 3 patients (20%) by endomyocardial biopsy. Multiple BLL (5 layers) was found in 9 patients (group A), but the other 7 patients showed a few or no BLL (group B). The group A patients showed a lower left ventricular (LV) ejection fraction compared to the group B patients (36 ± 15 vs. 59 ± 21%, p < 0.05). Presence area of interstitial fibrosis measured by morphometry in the group A patients was significantly greater than that in the group B patients (17 ± 10 vs. 8 ± 5%, p < 0.05). A cardiac event (worsening heart failure requiring hospitalization, sustained ventricular tachycardia, or cardiac death) occurred in group A patients (67%) and 7 group B patient (14%) during the mean follow-up period of 66 months (p < 0.05). Although in the group A patients, the left ventricular ejection fraction was significantly decreased compared to the group B patients (38 ± 19% vs. 54 ± 23%, p < 0.01), systolic function was unchanged in group B. Conclusion: Microangiopathy may be related to ventricular remodeling, disease progression, and poor clinical outcome in cases of CS.

Myocardial Infiltration in Sarcoidosis Demonstrated With Delayed Contrast Enhanced Magnetic Resonance Imaging


Background: Cardiac involvement in sarcoidosis is clinically diagnosed in 5% of patients, although it can be demonstrated at autopsy in 20 to 30% of cases. It can lead to significant morbidity and mortality, and early diagnosis may improve outcome. Echocardiography may demonstrate systolic and diastolic left ventricular dysfunction, but not demonstrate more subtle, early changes. Delayed contrast enhanced inversion recovery magnetic resonance imaging provides a clearer contrast between normal and involved myocardium in the setting of acute myocardial infarction. This technique may be more sensitive to demonstrated cardiac infiltration in sarcoidosis before left ventricular dysfunction appears.

Methods: Nineteen patients with recently diagnosed sarcoidosis were studied with a TI-sensitive (inversion recovery) gradient echo sequence (TE = 3.4 msec, TR = RR interval, inversion time = 800 msec) before and after the administration of a TI shortening contrast agent (0.1 mmol/kg Gd-DTPA, Magnevist, Schering, Berlin). Addionally cine gradient echo images in short- and long-axis orientations where obtained to evaluated left ventricular function.

Results: The 3 patients with active, untreated extracardiac sarcoidosis showed regions with patchy increased signal intensity due to local inflammation. In these regions systolic myocardial wall thickening was decreased. After treatment with steroids in 2 patients, follow-up studies showed a decrease of these patchy abnormalities. In the 16 patients who had received treatment with steroids, no regions with increased signal intensity where observed. These patients had a normal systolic LV function.

Conclusion: Contrast enhanced magnetic resonance imaging can demonstrate myocardial involvement in sarcoidosis, and be used to monitor the effect of initiated treatment.

Elevated Serum Troponin Levels Predict Short-Term Mortality in Patients With Systemic Amyloidosis

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Background: The development of heart failure symptoms in systemic amyloidosis suggests advanced disease and a poor prognosis. Abstent overt heart failure, echo parameters have often been used to determine prognosis. We hypothesized that the cardiac troponin I and T (cTnI, cTnT) which are sensitive and specific biomarkers of myocardial injury might predict early cardiac involvement and provide improved risk stratification.

Methods: Between April 1979 and November 2000, 268 patients with a tissue diagnosis of AL amyloidosis who had brad samples collected within 30 days of diagnosis were seen at the Mayo Clinic, Rochester, Mn. Troponin levels were determined and related to survival.

Results: cTnT (Roche) was elevated in 93% and cTnT (Dade) in 85% of patients. Mortal survival was 60 months in patients with elevated cTnT and 9 months with elevated cTnI. Patients with undetectable levels had median survival of 21 months. Multivariate analysis demonstrated cTnT was the most powerful predictor of mortality, providing a continuous distribution of risk. Prognostication improved still further when the troponin data were added to age and IV septal wall thickness.

Conclusion: Elevated serum troponin levels, particularly cTnT, detected early after the diagnosis of PSA predict cardiac involvement and a poor short-term prognosis. Their measurement should aid in determining prognosis and therefore, the appropriateness of therapeutic interventions.

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