intermediate in non-Met30 ATTR, and minimal in Met30-related ATTR. Vascular deposition is far more frequent in AL than in ATTR.

1160-78 Pacemakers and Defibrillators in Patients With Cardiac Sarcoidosis

Allen J. Solomon, Henry Yeager, Jr., Cynthia M. Tracy, Georgetown University Hospital, Washington, DC

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 51 ± 11 years) with biopsy-proven systemic sarcoidosis and presumed sarcoid of the heart. All patients 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 received a PM, 3 patients received 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 80 months. For ICD patients, the defibrillation threshold at implant was 16.5 ± 6.6 joules. One ICD patient received multiple appropriate ICD shocks for VT, while the other three patients have 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 PM's and ICD's function appropriately in this population, despite the infiltrative cardiomyopathy. (3) VT patients often have preserved systolic function. (4) The clinical course frequently stabilizes over time.

1160-79 Impact of Microangiopathy on Left Ventricular Remodeling and Clinical Outcome in Cardiac Sarcoidosis Patients Treated With Corticosteroids: A Long-Term Echocardiographic Follow-Up Study

Yoshikazu Yazaki, Kamiyoshi Yuichi, Kaoru Kogashi, Osamu Yokoseki, Hiroshi Imamura, Hiroaki Takenaka, Mafumi Owa, Minoru Hongo, Keishi Kubo, Morie Sekiguchi, Shinshu University, Matsumoto, Japan

Background: Microangiopathy presenting as basal lamina layering around the microvasculature (BLL) 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 ultra structural observations of heart biopsy specimens were performed. Sarcoidosis 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 (39 ± 13% vs. 59 ± 23%, p<0.05). Percent 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 6 group A patients (67%) and 1 group B patient (14%) during the mean follow-up period of 66 months (p<0.05), and 5 group A patients (56%) died. Despite corticosteroid treatment, group A patients showed a significant increase in LV volume index (78 ± 21ml/m2 to 103 ± 23ml/m2, p<0.01) and decrease in wall thickness of the interventricular septum (12 ± 4mm to 9 ± 3mm, p<0.01) over 5-year follow-up, although there was no significant change in LV ejection fraction and ventricular mass. No group B patient died, and echocardiographic parameters were unchanged in this group. Conclusion: Microangiopathy may be related to ventricular remodeling, disease progression, and poor clinical outcome in cases of CS.

1160-80 Myocardial Infiltration in Sarcoidosis Demonstrated With Delayed Contrast Enhanced Magnetic Resonance Imaging

Robert J. van Geuns, Jan P. Smedema, Peter Pattynama, Piotr A. Wielopolski, Pim J. de Feyter, Erasmus MC, Rotterdam, The Netherlands

Background:
Cardiac involvement in sarcoidosis is clinically diagnosed in 5% of patients, although it can be demonstrated at autopsy in 20 to 30 per cent 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 larger contrast between normal and injured myocardium in the setting of acute myocardial infarction. This technique may be also more sensitive to demonstrated cardiac infiltration in sarcoidosis before left ventricular dysfunction appears.
Methods:
Nineteen patients with recently diagnosed extracardiac sarcoidosis were studied with a T1-sensitive (inversion recovery) gradient echo sequence (TE = 3.4 msec, TR = RR interval, inversion time 250 ms) before and after the administration after a T1 shorting contrast agent (0.1 mmol/kg Gd-DTPA, Magnevist, Shering, Berlin). Additionally 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.

1160-81 Elevated Serum Troponin Levels Predict Short-Term Mortality in Patients With Systemic Amyloidosis

Wayne L. Miller, Angela Dispenzieri, Allan S. Jaffe, Krishnaswamy Chandrasekaran, Joseph P. McConnell, Mary F. Burritt, Mayo Clinic, Rochester, MN

Background: The development of heart failure symptoms in systemic amyloidosis suggests advanced disease and a poor prognosis. Absent overt heart failure, echo parameters have often been used to determine prognosis. We hypothesized that the cardiac troponin T and I (cTnT, cTnI) 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, 266 patients with a tissue diagnosis of AL amyloidosis who had blood 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 63% and cTnI (Dade) in 88% of patients. Median survival was 6 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.

POSTER SESSION

1161 Exercise Testing: Estimating Prognosis

Tuesday, April 01, 2003, 9:00 a.m.-11:00 a.m.
McCormick Place, Hall A
Presentation Hour: 10:00 a.m.-11:00 a.m.

1161-59 The Relationship Between Diabetes Mellitus and the Chronotropic Index in the Prediction of Cardiac Death

Babak Azarbal, Sean W. Hayes, Rory Hachamovitch, Daniel S. Berman, Cedars-Sinai Medical Center, Los Angeles, CA

Background: Diabetes mellitus (DM) and impaired heart rate response to exercise, measured by chronotropic index (CI), are predictors of cardiac death (CD). Also, pts with DM often have autonomic dysfunction. We studied the relationship between DM and CI in pts undergoing exercise myocardial perfusion SPECT (MPS).
Methods: 10,830 consecutive pts without valvular disease and not onβ blockers underwent symptom-limited exercise MPS; 609 pts were censored for early revascularization (<60 d). CI = (peak HR - rest HR)/(220-age-rest HR), with CI<0.8 defined as low. Survival modeling was performed.
Results: 93 CD occurred during a follow-up of 719 ± 252 days (98% complete). Of 10,021 pts, 1085 (11%) had DM and 2956 (30%) had low CI. Pts with DM were more likely to have a low CI (46% vs. 27%, p<0.001).After adjusting for age, sex, family history of CAD, exercise duration, and MPS defect severity, CI was predictive and interacted significantly with DM (p=0.048). Adjusted survival by of CI and DM is shown in the Fig: A: pts with CI≥0.8/no DM; B: CI≥0.8/DM; C: CI<0.8/no DM; D: CI<0.8/DM.
In pts with CI≥0.8, DM did not increase risk compared to non-DM (Hazard Ratio=1.0, 0.4-2.7). In pts with CI<0.8, risk was doubled in non-DM (HazR=2.0, 1.2-3.1) and was even higher in DM (HazR=3.4, 1.9-6.4).
Conclusion: CI and DM interact significantly and diabetics frequently have low CI.