

# Cardiac Sarcoidosis versus Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

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Patients with cardiac sarcoidosis may present with clinical and morphological features similar to ARVC/D or cardiomyopathy (Ott 2003). Sarcoidosis is an inflammatory granulomatous entity of unknown cause, characterized by multisystemic involvement. Practically no organ is immune to sarcoidosis; most commonly, in up to 90% of patients, it affects the lungs. (Hoitsma 2004). The most commonly involved organ in sarcoid related death has been reported to be the lung in western countries, while it was the heart in the Japanese autopsy series. (Iwai 1994).

The diagnosis of myocardial sarcoidosis is difficult and frustrating. Its clinical manifestations depend on the location and extent of granulomatous inflammation, and the symptoms and signs range among benign arrhythmias, heart block, intractable CHF, intense chest pain, to fatal VF. (Sharma 2003).

The ECG finding may be normal or may reflect every degree of block of the atrioventricular junction and bundle of His and every type of arrhythmia along with nonspecific ST-T-wave changes (Flemming, 1994).

Cardiac sarcoidosis should be considered in all young patients with unexplained conduction disorders, (Kollermann 2001) CHF or in cases of SCD (Lip 1996).

In extensive forms are frequently pseudo myocardial infarction patterns with pathological Q waves on ECG. (Shindo 1998).

MRI abnormalities, consisting of cardiac signal intensity and thickness, with the following three patterns:

- 1) Nodular;
- 2) Focal increase in signal on gadolinium diethylenetriaminepentaacetic acid-enhanced, T1-weighted images;
- 3) Focal increased signal on T2-weighted images without gadolinium uptake.

The improvement or stability of the MRI findings is correlated with clinical features.

With corticosteroid therapy, the MRI images improved either partially or completely, whereas.

The cardiac MRI may find its usefulness as a guide to obtaining EMB specimens and to monitoring the response of the disease to treatment.

The study is small and lacks a correlation of myocardial histology with MRI features. However, the study clearly calls for a large multicenter trial.

The most significant drawback of MRI is that the patient with a pacemaker and/or automatic ICD will not be able to take advantage of it. In such patients, <sup>201</sup>Tl scanning remains the test for assessing myocardial damage.

Cardiac PET using (18) F-FDG under fasting conditions (fasting (18) F-FDG PET) is a promising technique for identification of cardiac sarcoidosis and assessment of disease activity. The methodology can detect the early stage of cardiac sarcoidosis, in which fewer perfusion abnormalities and high

inflammatory activity are noted, before advanced myocardial impairment. The sensitivity of fasting (18) F-FDG PET in detecting cardiac sarcoidosis was 100%, significantly higher than that of (99m)Tc-MIBI SPECT (63.6%) or (67)Ga scintigraphy (36.3%). The accuracy of fasting (18) F-FDG PET was significantly higher than (67) Ga scintigraphy.(Okumura 2004.).

An EMB is preferable, but the procedure has sensitivity as low as 20% (Uemura, 1999). Others author referred sensitivity approximately of 50% thus, the search for a safe, reliable, and easily available diagnostic test for cardiac sarcoidosis continues. The pathological feature is the presence of noncaseating granulomas that eventually form fibrotic scars.

The table below shows the principal differences between the two entities:

	Cardiac Sarcoidosis	ARVC/D
Family history:	Absent.	Present in 30% to 50% of cases. When the disease is identified genetic screening should be performed among patient's family members.
Gender (M/F):	1 to 1.	2.9 to 1
Age at presentation:	Young or middle-aged adults.	Adolescents and young adults, perhaps There are rare references in childhood
Multisystemic involvement:	Yes.	No.
Chest pain:	Intense chest pain is referred.	No.
Clinical myocardial restrictive features:	Possible.	No.
Mitral regurgitation:	Is common.	Only in late stage with involvement of LV.
Pseudo myocardial infarction patterns on ECG:	Frequent in extensive forms.	No.
Chest roentgenogram:	Bilateral hilar lymphadenopathy.	Eventually RV cardiomegaly.
Lungs affectation:	In up to 90% of patients. Cor pulmonale is frequent.	No.
Pathological features:	Noncaseating granulomas that eventually form fibrotic scars.	Typical fibro-fatty replacement of the RV myocardium on dysplasia triangle.
More common cardiac sites involved:	LV free wall and interventricular septum.	RVOT, RVIT, and apex of RV.
Pericardial effusion:	Are not uncommon.	Absent.
Improved MRI images with corticosteroids:	Yes.	No.
Therapy with corticosteroids,	Sometimes are indicated. (Mitchell 1997).	No.

hydroxychloroquine, methotrexate or cyclophosphamide:	Immunosuppressive and anticytokine treatments can be effective in severe systemic sarcoidosis and should be considered in sight-threatening disease.	
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## References

1. Flemming, H, Bailey, S Cardiac sarcoidosis. James, DG eds. *Sarcoidosis and other granulomatous disorders (vol 73)*. 1994;323-334 Marcel Dekker (New York, NY)).
2. Hoitsma E, Faber CG, Drent M, Sharma OP. Neurosarcoidosis: a clinical dilemma. *Lancet Neurol*. 2004;3:397-407
3. Iwai K, Sekiguti M, Hosoda Y, Racial difference in cardiac sarcoidosis incidence observed at autopsy. *Sarcoidosis*. 1994;11:26-31.
4. Kollermann J, Roos G, Helpap B. Sudden cardiac death from unrecognized cardiac sarcoidosis *Pathologe*. 2001; 22:141-4.
5. Lip GY, Gupta J, Gill JS, et al. Sarcoid heart disease: a rare cause of chest pain and malignant cardiac arrhythmia in a young Asian man. A case report. *Angiology*. 1996; 47:905-10.
6. Mitchell DN, du Bois RM, Oldershaw PJ. Cardiac sarcoidosis. *BMJ*. 1997; 314:320-1.
7. Okumura W, Iwasaki T, Toyama T, Usefulness of Fasting 18F-FDG PET in Identification of Cardiac Sarcoidosis. *J Nucl Med*. 2004;45:1989-98.
8. Ott P, Marcus FI, Sobonya RE, et al. Cardiac sarcoidosis masquerading as right ventricular dysplasia. *Pacing Clin Electrophysiol*. 2003;26: 1498-1503.
9. Sharma OP, Diagnosis of Cardiac Sarcoidosis An Imperfect Science, A Hesitant *Art Chest*. 2003; 123:18-19.
10. Shindo T, Kurihara H, Ohishi N, et al. Images in cardiovascular medicine. Cardiac sarcoidosis. *Circulation*. 1998; 97:1306-7.
11. Uemura, A, Morimoto, S, Hiramitsu, S, et al *Histologic diagnostic rate of cardiac sarcoidosis: evaluation of endocardial biopsies. Am Heart J* 1999; 138,299-302.