Expert consensus recommendations on criteria for the diagnosis of cardiac sarcoidosis and Consensus recommendations for ICD implantation in patients diagnosed with cardiac sarcoidosis

Approximately 5% of patients with sarcoidosis will have clinically manifest cardiac involvement(Cardiac Sarcoidosis: CS) and another 20 – 25% have asymptomatic cardiac involvement (clinically silent disease). There is a growing understanding that CS can be the first manifestation of sarcoidosis in any organ. The extent of LV dysfunction seems to be the most important predictor of prognosis. In addition, the extent of LGE is emerging as an important prognostic factor. There is some controversy regarding outcomes for patients with clinically silent CS and larger studies are needed. Immunosuppression therapy (usually with corticosteroids) has been suggested for the treatment of clinically manifest CS despite modest data. Patients with clinically manifest disease often need device therapy, typically with implantable cardioverter defibrillators. There are still many unknowns in terms of best practices in diagnosing and managing CS patients and multi-centre research efforts are underway.

Expert consensus recommendations on criteria for the diagnosis of cardiac sarcoidosis

There are two pathways to a diagnosis of CS:

(1) Histological diagnosis from myocardial tissue

CS is diagnosed in the presence of non-caseating granuloma on histological examination of myocardial tissue with no alternative cause identified (including negative organismal stains if applicable).

(2) Clinical diagnosis from invasive and non-invasive studies

It is probable* that there is CS if:

(a) There is a histological diagnosis of extra-cardiac sarcoidosis

and

(b) One or more of following is present

- Steroid ± immunosuppressant responsive cardiomyopathy or heart block
- Unexplained left ventricular ejection fraction <40%
- Unexplained sustained (spontaneous or induced) ventricular tachycardia
- Mobitz type II second degree heart block or third degree heart block
- Patchy uptake on dedicated cardiac FDG-PET (in a pattern consistent with CS)
- Late gadolinium enhancement on CMR (in a pattern consistent with CS)
- Positive gallium uptake (in a pattern consistent with CS)

and

(c) Other causes for the cardiac manifestation(s) have been reasonably excluded

CS, cardiac sarcoidosis; FDG-PET, Fluorodeoxyglucose-Positron Emission Tomography; CMR, cardiac magnetic resonance.
*In general, 'probable involvement' is considered adequate to establish a clinical diagnosis of cardiac sarcoidosis.

Birnie DH, Sauer WH, Bogun F, Cooper JM, Culver DA, Duvernoy CS, Judson MA, Kron J, Mehta D, Cosedis NJ, Patel AR, Ohe T, Raatikainen P, Soejima K. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. Heart Rhythm 2014;11:1305 – 1323.

Consensus recommendations for implantable cardioverter defibrillator implantation in patients diagnosed with cardiac sarcoidosis