**Viewpoints in Cardiac Sarcoidosis:**
Our adoption and modification of current guidelines

Dae-Won Sohn, M.D., Ph.D., FACC, FASE
Department of Internal Medicine, Seoul National University College of Medicine

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**Cardiac sarcoidosis**

1. Endomyocardial Bx

Cardiac + Sarcoidosis

2. Imaging diagnosis + Dx of syst sarcodiosis

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**EM Bx in Sarcoidosis**

- Histologic diagnostic rate of cardiac sarcoidosis: Evaluation of endomyocardial biopsies
- AHA/ACCF/ESC Scientific Statement: The Role of Endomyocardial Biopsy in the Management of Cardiomyopathy

5/26 pt (19.2%) ≈ 20-30%

SNUH: 2/25 pt (8%)

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**Diagnosis of cardiac sarcoidosis**

- 2008: Revised guideline by the Japanese Society of Sarcoidosis and Granulomatous Disorders

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**Cardiac sarcoidosis**

Cardiac + Sarcoidosis

2. Imaging diagnosis + Dx of syst sarcocidosis
Histologic Dx of cardiac sarcoidosis

- Prevalence of extracardiac sarcoidosis in cardiac sarcoidosis: isolated cardiac sarcoidosis: up to 2/3

Histologic Dx of extracardiac sarcoidosis

- 25 pts with final Dx of cardiac sarcoidosis
- enlarged hilar LN in chest CT in 10 pts

- 11 hilar EBUS bx
- 10 enlarged LN: 1 granuloma
- 8 not diagnostic
- 1 normal sized LN: pts – not diagnostic

Dx of cardiac sarcoidosis:

<table>
<thead>
<tr>
<th>Findings</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlarged hilar LN in chest CT</td>
<td>1</td>
</tr>
<tr>
<td>Enlarged LN bx</td>
<td>2</td>
</tr>
<tr>
<td>Findings suggesting systemic involvement but not amenable to biopsy</td>
<td>1</td>
</tr>
</tbody>
</table>

Steroid treatment: Is it effective?

- Composite of All (cause death $Y+C+D$)
- Cardiac death $B$
- Symptomatic arrhythmias $C$
- HF requiring admission $D$

| A Overall                          | 1     |
| B Steroid Rx                       | 1     |

Steroid treatment: How?

- Prednisone 0.5mg/kg/day or 0.5mg/kg/day (max 60 mg)
- Remit RT year after 1 month of treatment
- No abnormal cardiac ICG uptake or PIT
- Abnormal cardiac ICG uptake or PIT


Birnie DH et al. JACC 2016;68:411-21
**Steroid treatment: How?**

- Steroid therapy in patients with EF <35% and (+) FDG-PET. Initiate after baseline FDG-PET scan. 
  - Prednisone 0.5 mg/kg/day for 2 weeks, then 0.25 mg/kg/day to 3-4 mg/day.

- Re-evaluation of disease activity after 2 months, then every 3-6 months.

- FDG-PET scan 3 months after starting treatment.

- Evaluation of regional or global LV function with echocardiography every 3 months. 
  - No improvement in symptoms or function: continue treatment.

- Improvement in symptoms or function: taper treatment over 3 months.

**Dx of cardiac involvement: FDG-PET**

- Risk of ventricular fibrillation and sudden death.

- Conduction disturbance, VT, Sudden death.

- Pacemaker?:

  - Beta blocker?

- VT ablation

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**Conduction disturbance, VT, Sudden death**

**Study**

- No. of patients
- Risk factors

**Complete suppression of VT**

- VT detection

**Follow-up**

- VT recurrence

<table>
<thead>
<tr>
<th>Study</th>
<th>No. of Patients</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Kaplan et al</td>
<td>8</td>
<td>650 (15)</td>
<td>490 (44)</td>
<td>6</td>
<td></td>
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<tr>
<td>Morel et al</td>
<td>9</td>
<td>250 (30)</td>
<td>94 (29)</td>
<td>6</td>
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<tr>
<td>Schmitt et al</td>
<td>6</td>
<td>0.5 (0.4)</td>
<td>0.5 (0.4)</td>
<td>6</td>
<td></td>
<td></td>
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<tr>
<td>Nihal et al</td>
<td>14</td>
<td>450 (45)</td>
<td>150 (15)</td>
<td>20</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Number et al</td>
<td>21</td>
<td>150 (25)</td>
<td>15 (5)</td>
<td>24 (median)</td>
<td></td>
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</tr>
</tbody>
</table>

Note: VT indicates ventricular tachycardia.
Current HRS recommendations

Expert Consensus Recommendations for VT Abatement and Risk Stratification for Sudden Cardiac Death in CS

1. VT is recommended for prophylaxis when it occurs in the setting of myocardial dysfunction or wall motion abnormality.
2. VT should be assessed in patients with sustained ventricular arrhythmia in or near the 12-lead ECG and echocardiographic evidence of myocardial dysfunction.
3. VT ablation is recommended in patients with symptomatic ventricular arrhythmia and evidence of myocardial dysfunction.
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Diagnostic issue

VT with LBBB morphology

1) ARVC
2) Sarcoidosis
3) Idiopathic giant cell myocarditis

- Vasaiwala et al: 15 pts diagnosed as ARVC - EM Bx showed sarcoidosis in 3/15 (18%)
- Dechering et al: 5/8 patients (63%) with cardiac sarcoid - Fulfill Dx criteria of ARVC

Diagnostic issue

70 YO male with dyspnea

Diagnostic issue

42 YO male with complex Hx