

Other medical treatment as indicated including antiarrhythmic therapy, device placement if appropriate, standard heart failure management

Active cardiac sarcoidosis

- Ventricular tachyarrhythmias, high-grade heart block or new bifascicular block, left or right ventricular dysfunction, and
- Active FDG uptake on cardiac FDG-PET

Treat in cases of active inflammation with symptoms based on the following stratification (see Figure 2)

- Definite: Should treat
- Highly probable: Should treat
- Probable: Individualized decision-making

Initiate corticosteroids

- Prednisone 30–40 mg/day with taper specific to patient presentation and comorbidities
- Consider concomitant initial therapy with one of the following steroid-sparing agents: methotrexate, mycophenolate, azathioprine, or leflunomide

Reassessment at 3–6 months

- Cardiac FDG PET

Decreased or resolved inflammation

- Continue prednisone at 10–20 mg/day or taper prednisone further (if on a steroid sparing agent or if clinical manifestations are improved)

Reassessment at 3–6 months

- Cardiac FDG PET

Resolved inflammation

- Taper immunosuppression to off after 12 months of treatment from time of initiation
- Additional decision-making for monitoring of active CS should be guided by clinical course

Decreased but residual inflammation

- Individualize decision-making for immunosuppression based on clinical picture (eg change in arrhythmia burden or cardiac function)
- Additional decision-making for monitoring of active CS should be guided by clinical course

Unchanged or worsening inflammation

Unchanged or worsening inflammation

- Raise prednisone to the lowest previous effective dose
- Add a previously unused steroid-sparing agent: methotrexate, mycophenolate, azathioprine, or leflunomide

Reassessment at 3–6 months

- Cardiac FDG PET

Minimal residual or resolved inflammation

- Wean prednisone to 10–20 mg/day
- Continue steroid sparing agent

Reassessment at 6 months

- Cardiac FDG PET

Resolved inflammation

- Wean prednisone to off
- Continue steroid sparing agent
- Additional decision-making for weaning immunosuppression should be guided by clinical course

Unchanged or worsening inflammation

In cases of no response, evaluate for an alternative diagnosis and/or false positive PET scan especially if diagnosis of CS was not histologically confirmed

If significant drug side effects or contraindications to a steroid sparing agent, start a biologic

Unchanged or worsening inflammation

- Raise prednisone to the lowest previous effective dose
- Add a biologic agent (infliximab or adalimumab)
- Additional decision-making for weaning immunosuppression should be guided by clinical course

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