

## Cardiac sarcoidosis – elusive diagnosis

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## Cardiac sarcoidosis, a clinical perspective

- | background on sarcoidosis
- | clinical features
- | role of  $^{18}\text{F}$ FDG PET in the diagnosis of cardiac sarcoidosis
- | The use of  $^{18}\text{F}$ FDG PET in the monitoring of CS activity

## Background: Sarcoidosis

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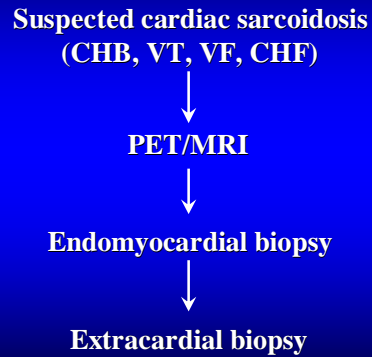
- Sarcoidosis is a multisystem, granulomatous disease of unknown etiology.
- Noncaseating granulomas
- Most commonly: pulmonary and lymph node involvement
- Heart, liver, spleen, skin, eyes, phalangeal bones, parotid glands, CNS

## Background: Sarcoidosis?

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- Highest reported incidence in scandinavia 50-60/100.000
- Cardiac sarcoidosis has a poor prognosis due to arrhythmias and hear failure
- Definition of cardiac sarcoidosis varies

## Cardiac sarcoidosis in HYKS



## Definition: Cardiac sarcoidosis

- **Cardiac sarcoidosis:**
  - granulomas in RV or LV biopsy
- or
- **typical clinical features**
  - histologically verified sarcoidosis in other organ
  - pulmonary sarcoidosis (radiology only, histology not necessary)

## Patient characteristics

| Diagnosis            | Cardiac bx  | Other bx     |
|----------------------|-------------|--------------|
| n                    | 26          | 16           |
| Age (average, range) | 50 (33-66)  | 50 (26-75)   |
| Sex (% women)        | 77% (20/26) | 50% (8/16)   |
| Other sarcoidosis    | 27% (7/26)  | 100% (16/16) |

HYKS 2000-2009

## Clinical features of Cardiac sarcoidosis

| Diagnosis                       | Cardiac bx         | Other organ bx     |
|---------------------------------|--------------------|--------------------|
| <b>Complete heart block</b>     | <b>54% (14/26)</b> | <b>44% (7/16)</b>  |
| <b>Ventricular tachycardia</b>  | <b>69% (19/26)</b> | <b>63% (10/16)</b> |
| <b>Ventricular fibrillation</b> | <b>15% (4/26)</b>  | <b>6% (1/16)</b>   |
| <b>Heart failure</b>            | <b>79% (19/26)</b> | <b>25% (4/16)</b>  |

## Treatment of Cardiac sarcoidosis

| Diagnosis  | Cardiac bx  | Other organ bx |
|------------|-------------|----------------|
| Pacemaker  | 38% (10/26) | 56% (9/16)     |
| ICD        | 46% (12/26) | 25% (4/16)     |
| Cardiac tx | 27% (7/26)  | 0%             |

## Prognosis in cardiac sarcoidosis

Death/ cardiac transplant/ VF (resuscitation & aborted sudden death)

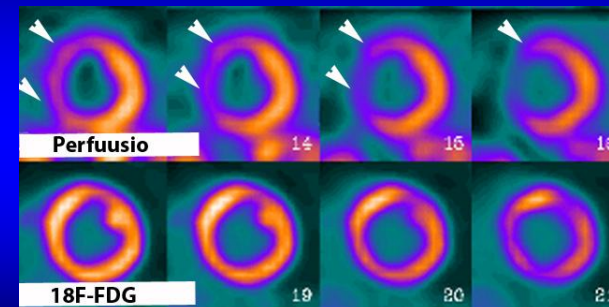
Myocardial bx pos 46% (12/26)

Other organ biopsy pos 25% (4/16)

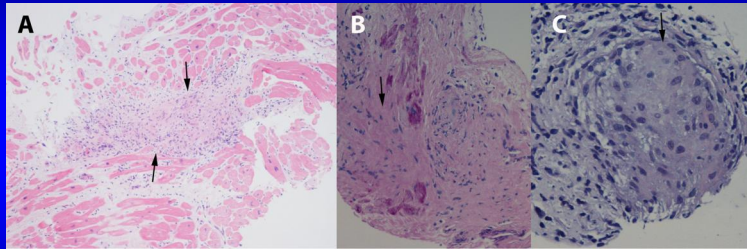
## Diagnostic imaging in cardiac sarcoidosis

| Diagnosis |   | Cardiac bx  | Other organ bx |
|-----------|---|-------------|----------------|
| TTE       | Septal abnormality                                | 69% (18/26) | 73% (19/26)    |
|           | EF <50%   | 31% (5/16)  | 25% (4/16)     |
| PET       | <sup>18</sup> F-DG enhancement & perfusion defect | 75% (12/16) | 50% (3/6)      |
|           | MRI   |             |                |
|           | Late enhancement                                  | 73% (8/11)  | 60% (3/5)      |

## Cardiac <sup>18</sup>F-FDG PET



## Sarcoidosis is a tissue diagnosis



- | granulomas
- | giant epithelioid cells



## PET positive but biopsy negative

Suspected cardiac sarcoidosis  
(CHB, VT + heart failure)

PET/MRI

Endomyocardial biopsy

Extracardial biopsy

|     |   | Biopsy |    |
|-----|---|--------|----|
|     |   | -      | +  |
| PET | + | 31     | 29 |
|     | - | 27     | 3  |

## Follow-up of inflammatory activity

- Cardiac sarcoidosis can be progressive myocardial disease – how to select these patients for more aggressive and long-term immunosuppressive therapy?
- Anecdotal data – <sup>18</sup>F FDG PET? (Yamagishi et al. J Nucl Med 2003; 44:1030–1036).

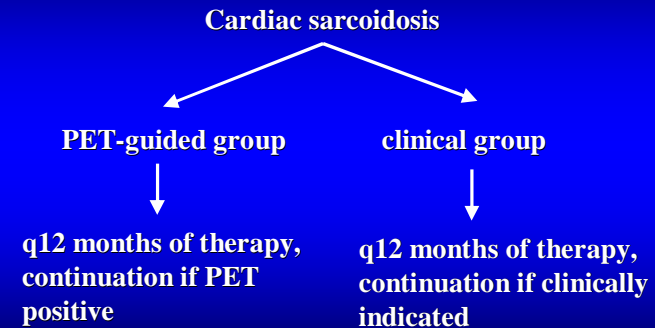
## Issues with imaging

- No information about the sensitivity or specificity of PET <sup>18</sup>F FDG imaging exists
- Prognostic value of PET findings is not established
- The use of <sup>18</sup>F FDG PET to follow-up the disease activity is not established

***PET-guided immunosuppressive therapy in cardiac sarcoidosis. A prospective, randomized trial .***

**Hypothesis:  $^{18}\text{F}$ -FDG PET can be used to measure inflammatory activity and immunosuppressive therapy can be administered based on the actual disease activity rather than a less sensitive surrogate of disease activity such as left ventricular dysfunction.**

**$^{18}\text{F}$ FDG PET in sarcoidosis: Design**



**$^{18}\text{F}$ FDG PET in sarcoidosis: endpoints**

- 1) death & aborted sudden death & cardiac transplant.
- 2) worsening LV function, hospitalization for CHF, ventricular tachycardias
- 3) clinical manifestations of corticosteroid side-effects: cataracts, osteoporosis, infections

**Conclusions**

- Cardiac sarcoidosis afflicts most commonly 30-50 y/o females and has poor prognosis due to progressive heart failure and fatal ventricular arrhythmias
- $^{18}\text{F}$  FDG PET appears to be useful in the diagnosis of cardiac sarcoidosis, however, normal  $^{18}\text{F}$ FDG PET does not rule out cardiac sarcoidosis
- The use of  $^{18}\text{F}$  FDG PET in the monitoring of the inflammatory activity needs to be validated

## Sensitivity of 18 FDG-PET in the diagnosis of cardiac sarcoidosis

- Sensitivity: no systematic data
- Based on our experience sensitivity of pet < 100%
  
- Specificity: no systematic data
- A number of PET positive, biopsy negative.  
How to treat these patients?