Outcome

- Sarcoid on EBUS biopsy
- Positive PET
- ICD and immunosuppression
Case 2 – CMR Referral

- Symptomatic VEs, but admitted from ETT with nasty rapid VT on exercise. Not typical outflow tract. Normal echo. Father had CAD (but was heavy smoker). No clear aetiology. Normal CT Thorax
Case 2 – Cines
RV Free Wall LGE?
Overall, the diagnosis is of a **mild dilated cardiomyopathy with a degree of non compaction**. The architectural abnormalities make it most likely to be familial in aetiology. However, no fibrosis is seen as a nidus for the VT which has been seen. Referral to the inherited heart disease team is suggested, particularly to potentially screen for a lamin mutation, although typically such patients have PR prolongation and septal fibrosis.
What happened

- ICD implant
- Presents a year on with repeated shocks
- And....
Amyloidosis
AL amyloidosis

- Plasma cell dyscrasia > Free light chains > Amyloid
- Most common type (<80 years old)
- Median survival 6 months if untreated
ATTR amyloidosis

- Liver > Transthyretin - Wild type form
  - Mutant form
- Exponential increase in new diagnosis
  - >80 yrs 25% amyloid
  - Specific ethnicities
Typical ECG
ECG in V122I ATTR amyloidosis: 25% meet LVH criteria

Dungu J Am Heart J 2012
Echo: structure and function
Echo: diastolic function
Echocardiogram

2009/05/21 13:56:40

GS = -10.3%

Peak Systolic Strain

Longitudinal Strain

SL 28.0
28.0%

AVC

Click here for tips
Other causes of interstitial expansion: amyloid deposition

Maceira et al. Circ 2005
Late gadolinium imaging - PSIR
3 patterns

Fontana M et al. Circulation 2015
LGE limitations

Need for quantitative method

track changes over time

Low eGFR

Contraindication to contrast
Intrinsic contrast of the myocardium
Native T1 in the hypertrophic phenotype

- Hypertension
- HCM
- AS

Sado DM et al. Circ Cardiovasc Imaging 2013
Spot the amyloid! There are 2 here.
What is T1 mapping?
Native T1 and ECV

*MOLLI at 1.5 T*
Why should we use T1 mapping

Martinez Naharro A YIA SCMR 2017
Grading $^{99m}$Tc-DPD uptake in the heart

- **Grade 1** = Mild cardiac uptake, less than bone; no attenuation of bone uptake
- **Grade 2** = Moderate cardiac uptake equal or greater than bone plus some bone attenuation
- **Grade 3** = Strong cardiac uptake with little or no bone signal
Non invasive diagnosis of ATTR amyloidosis

Heart failure, syncope, or bradyarrhythmia, with echocardiogram and/or cardiac magnetic resonance imaging (CMR) suggesting/indicating cardiac amyloid

Bone scintigraphy with $^{99m}$Tc-DPD/HMDP/PYP

Grade 0
Grade 1
Grade 2 to 3

Serum immunofixation + Urine immunofixation + serum free light chain assay (Freelite)
Monoclonal protein present?

No
Yes
No
Yes
No

Cardiac AL/ATTR amyloidosis unlikely
Review/request CMR
Need specialized assessment for Diagnosis: Histological confirmation and typing of amyloid
Cardiac ATTR amyloidosis

Cardiac amyloidosis (AL/ApoAI/ATTR/other)
Variant ATTR amyloidosis
Wild-Type ATTR amyloidosis

TTR genotyping
Untreated systemic amyloidosis is usually progressive and fatal

General treatment principals in amyloidosis

- Support or replace compromised organ function
- Reduce supply of the fibril precursor protein
Determining the amyloid fibril type

- Immunohistochemistry
  - Standard method in clinical practice
  - AA: sensitive and specific
  - Hereditary: mostly diagnostic
  - AL - false negatives in ~30%

- Mass Spectrometry
  - Important development. Tiny amounts of tissue required