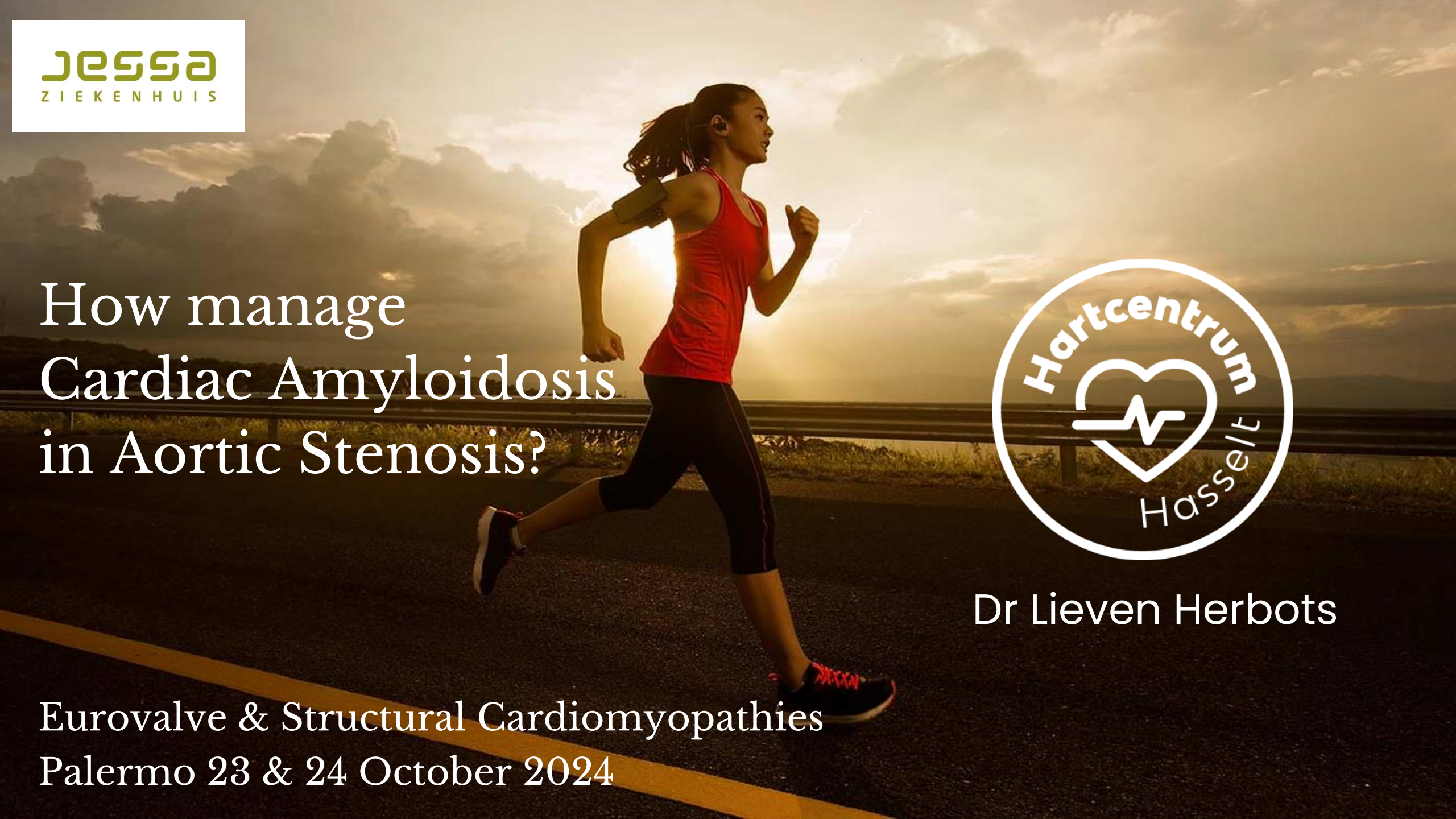


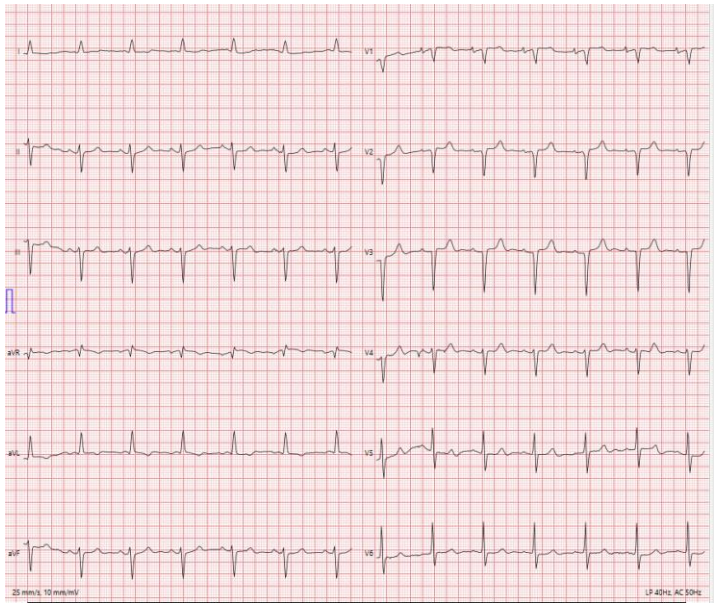
How manage Cardiac Amyloidosis in Aortic Stenosis?

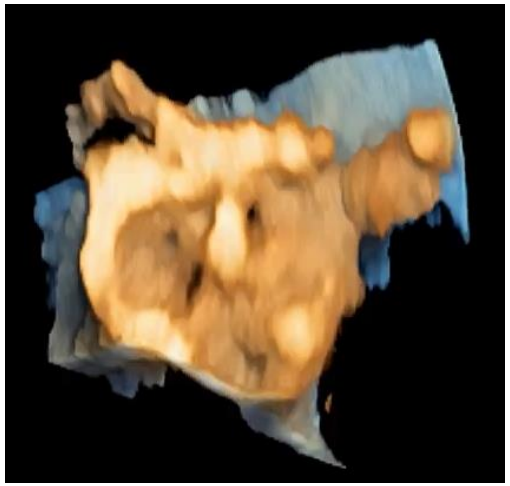
Dr Lieven Herbots

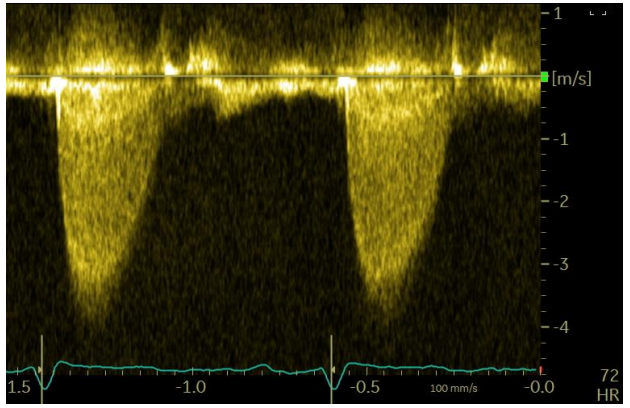
Eurovalve & Structural Cardiomyopathies
Palermo 23 & 24 October 2024

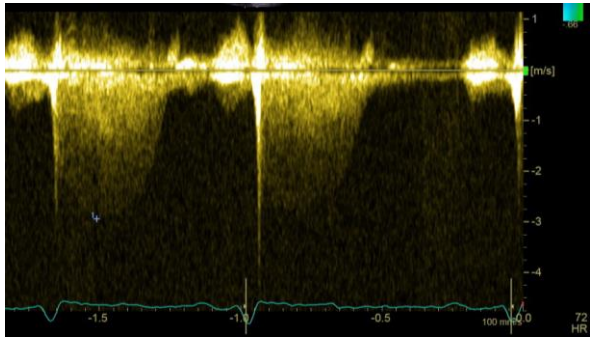
Case Male, 88y

1. History of CAD
   Ptca LAD/RAC
2. LFLG AS:
   PG 36 mmHg, MG 22 mmHg
   AVA 0,8 cm^2
3. Lower Backpains
4. Chronic Kidney Disease
5. Exercise-related runs of SVT
6. Left Ventricle Hypertrophy









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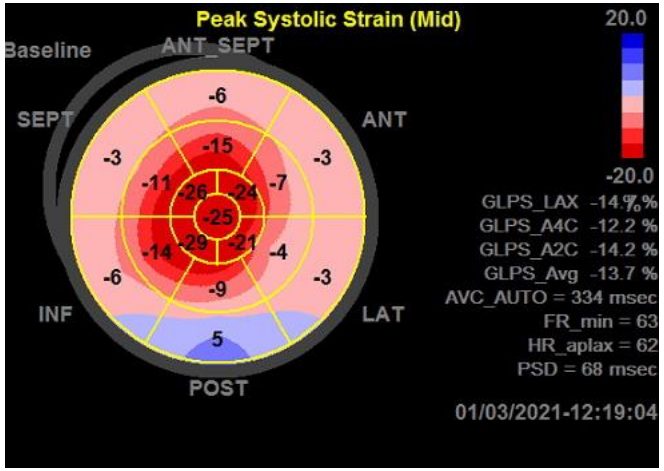
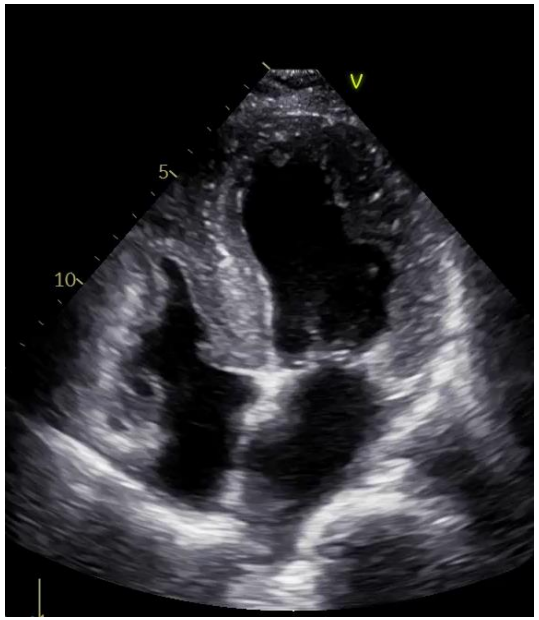
Case Male, 88y





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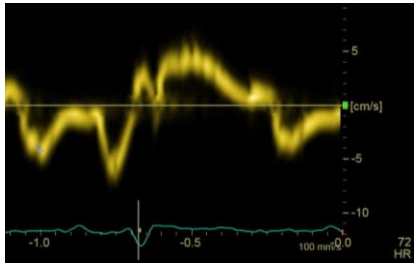

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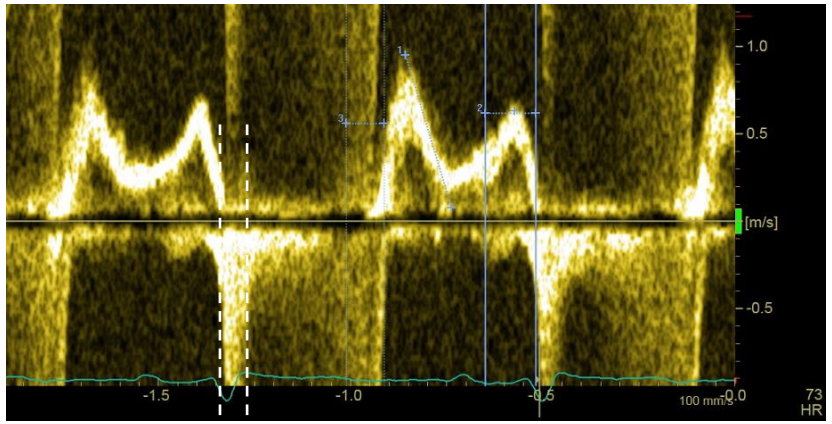

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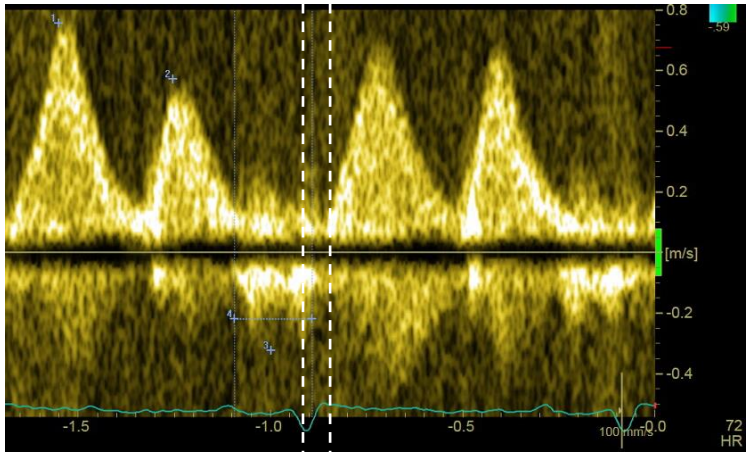

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Case Male, 88y, Evolution

1. All pre-TAVI examinations performed: eligable
2. Coronary Angiography: Ptca LAD
3. Bone Scan: Positive for ATTR Amyloidosis
4. The patient was denied TAVI because of CA and restricted nr/year
5. He was treated medically
6. He passed away last August, age 94y

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Questions

1. Did we make the right decision to wait?
2. Do we have treatment options for ATTR and LC-CA?
3. When is LVH inappropriate? Other clues/red flags?
4. Is Bone-scintigraphy sufficient or biopsy?
5. Prevalence Cardiac Amyloid CMP?
6. Does AVR alone or with CA therapy change the prognosis?

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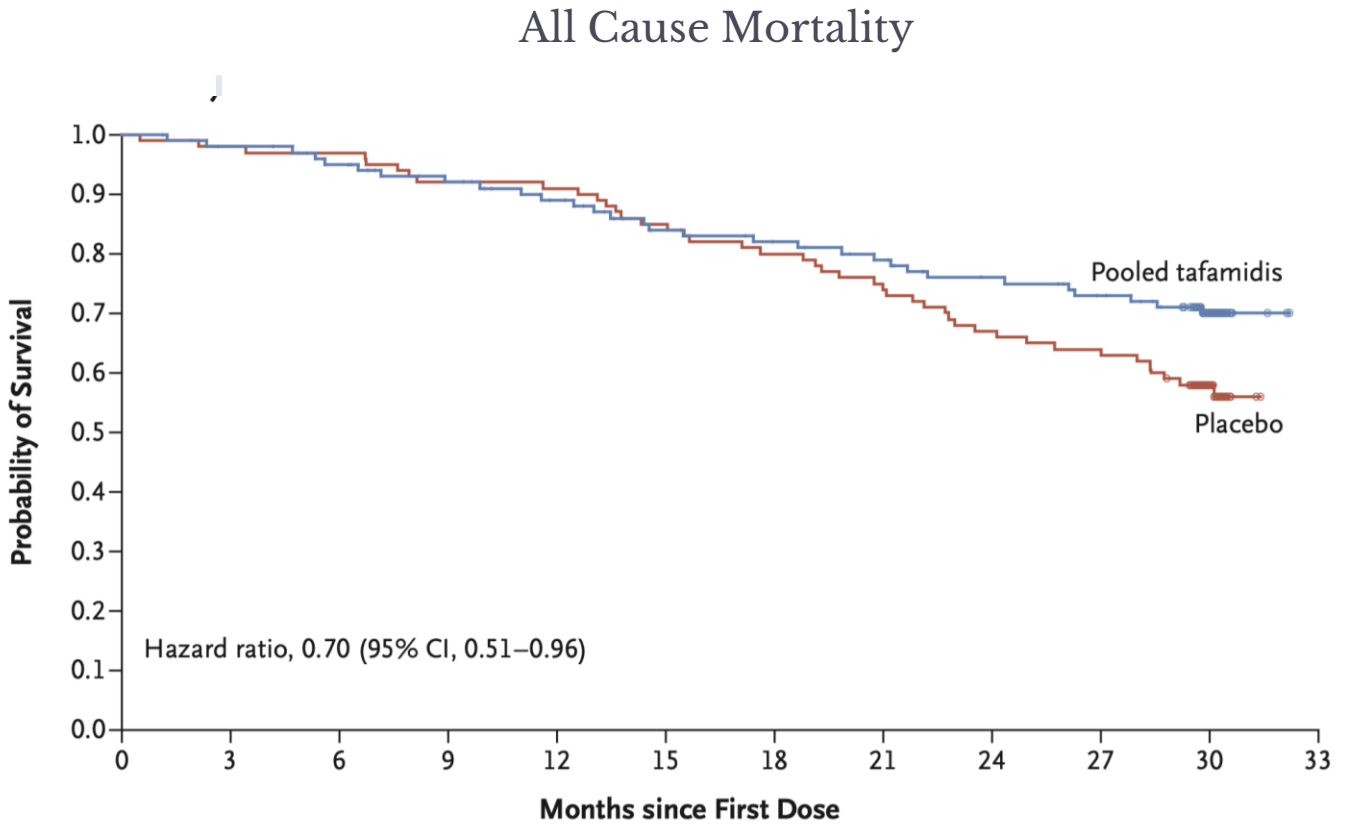
Tafamidis for TTR Amyloid CMP Pts

1. Reduced Mortality (30%)
2. Reduced CV Hosp (32%)
3. Improved Exercise Cap
4. Enhanced QUALI



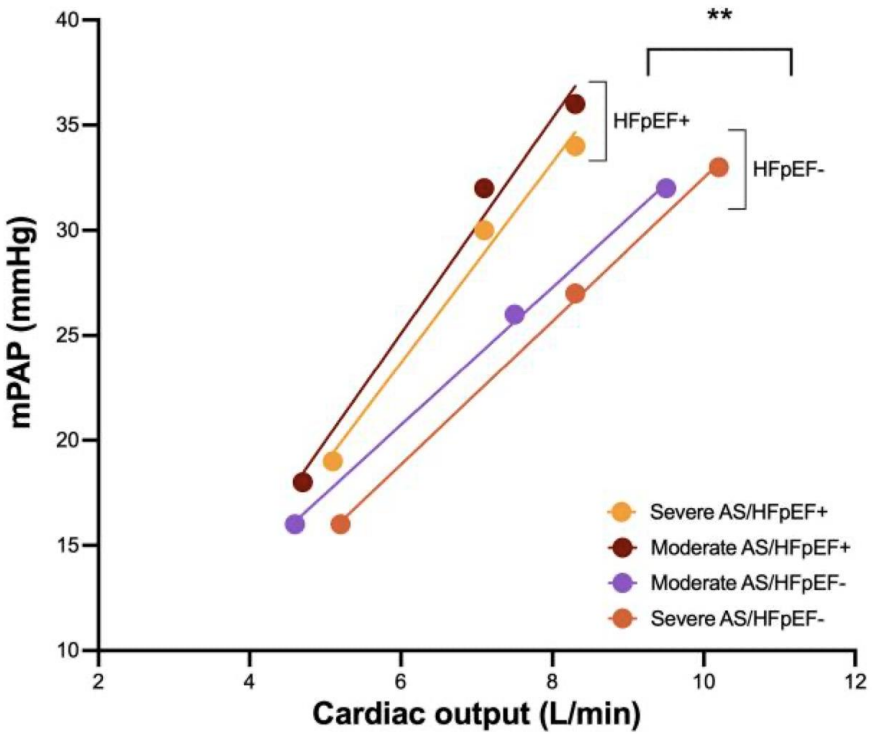
The NEW ENGLAND JOURNAL of MEDICINE

Mathew Maurer

13 September 2018

What drives adverse HD in AS/HFpEF?

1. HFpEF scores - determine cardiac dysfunction
2. mPAP/CO slopes determine resistance RH
3. Relate HFpEF and AS to HD burden
4. Cardiac Dysfunction has more impact than AS



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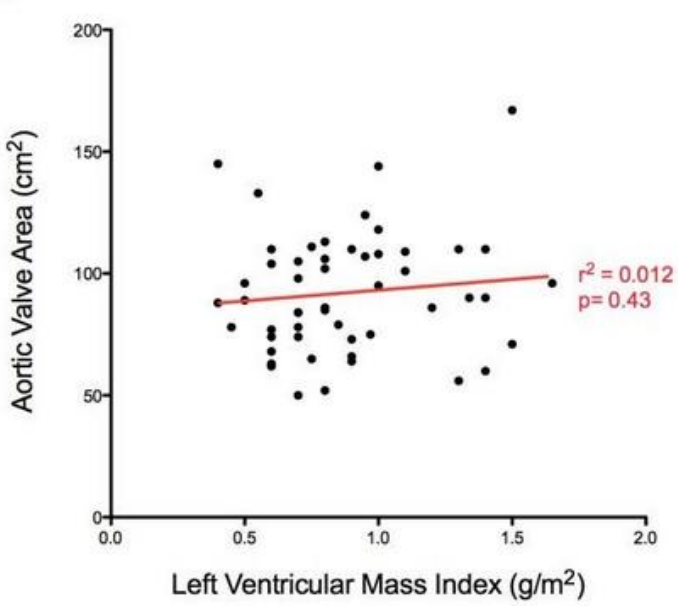
Sarah Hoedemakers. EHJ CVI 2023

Inapropriate Hypertrophy in AS

1. LVH early in the course of AS
2. Marked variation in the magnitude of response
3. Degree of LVH weakly related to AS severity
4. Hypertrophic Response more related to
   Age, Male, Obesity, AHT, Metabolic Syndrome
5. Increased arterial stiffness
   Reduced systemic arterial compliance



Diagnostic Approach

1

Clinical Suspicion

Recognise red flags in AS patients

2

Non-Invasive Testing

Echo, ECG, CMR, nuclear imaging

3

Biomarker Analysis

NT-proBNP, Troponin, light chains

4

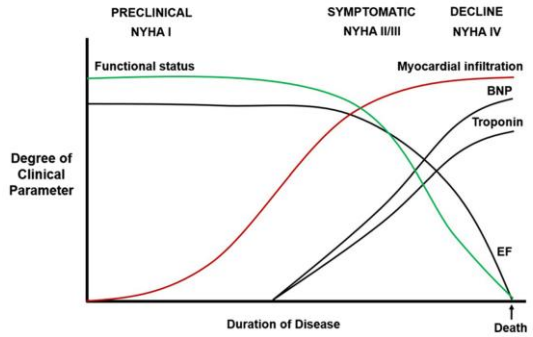
Tissue Biopsy

Endomyocardial or extra-cardiac confirmation

Clinical Manifestations

1 Heart Failure Symptoms

Disproportionate to AS severity



2 Orthostatic Hypotension

Due to autonomic neuropathy

3 Carpal Tunnel Syndrome

Bilateral involvement, often preceding cardiac symptoms

4 Periferal Neuropathy

Sensory disturbances in extremities

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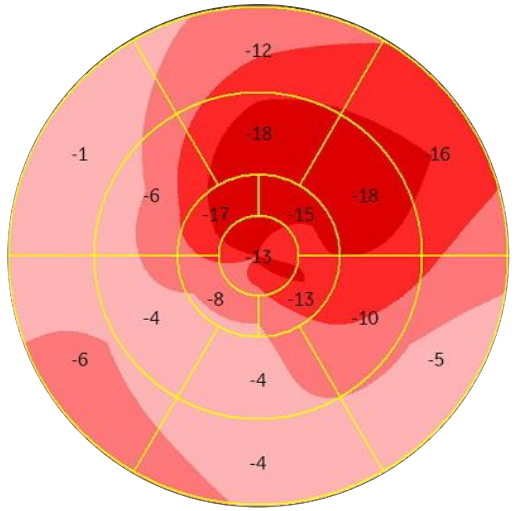
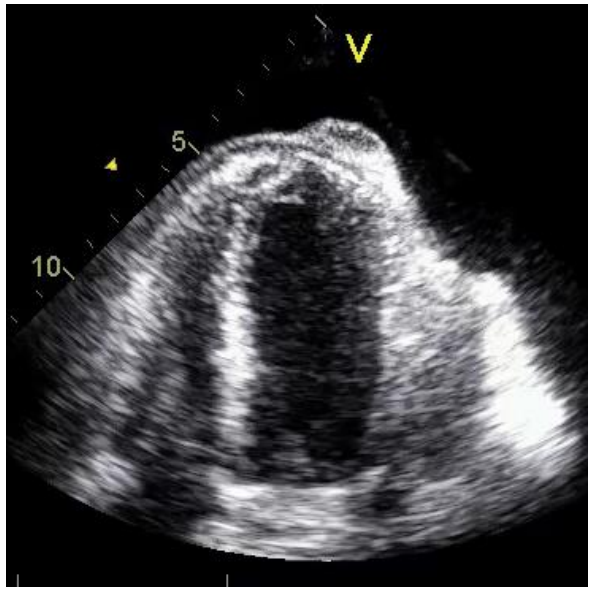
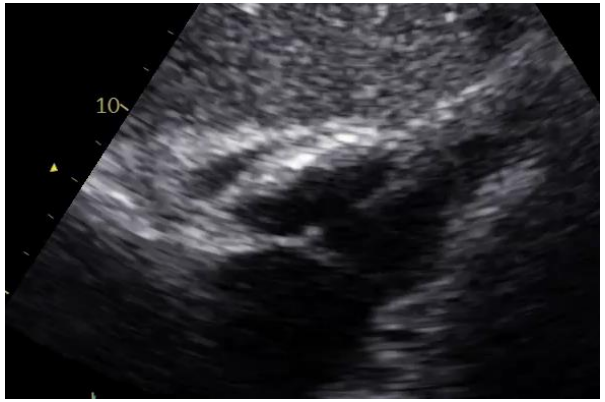
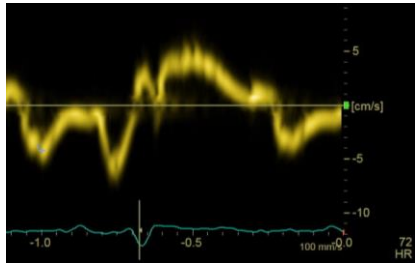
Echocardiographic Findings: Red Flags

1. Granular Sparkling of Myocardium
2. Pericardial Effusion
3. Low-Flow Low-Gradient Phenotype
4. Apical Sparing
5. Mitral Anulus S' < 6 cm/s
6. RV Involvement









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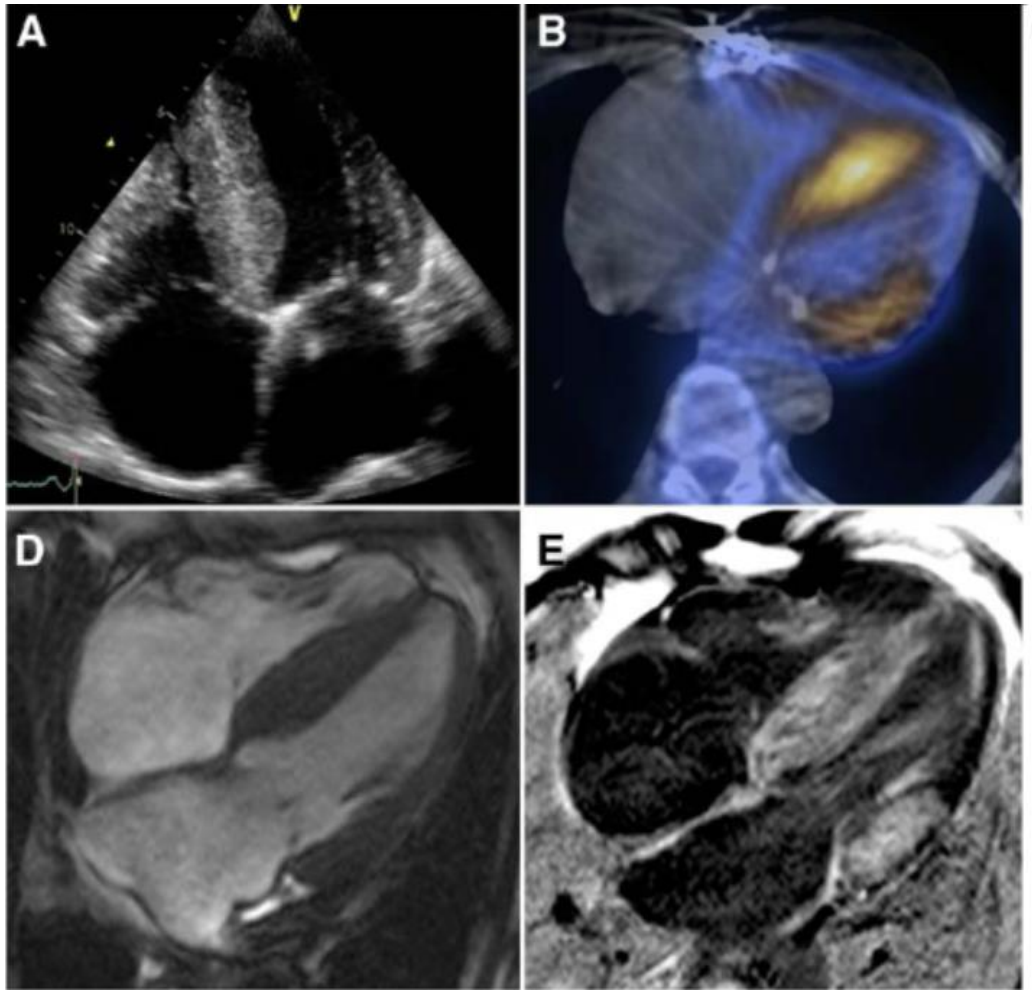
Laboratory Findings

Biomarker	Expected Finding
NT-proBNP	Markedly elevated
Troponin	Persistently elevated
Serum Free Light Chains	Abnormal ratio
Proteinuria	Present in AL amyloidosis

Multimodality Imaging



Echocardiography
LVH - AS

A

Single Foton Emission Tomography

B

Bone Scan
Perugin Grd 2 Uptake

C

Cardiac Magnetic Resonance
LVH – Impaired Syst Function

D

Cardiac Magnetic Resonance
Transmural GDE

E

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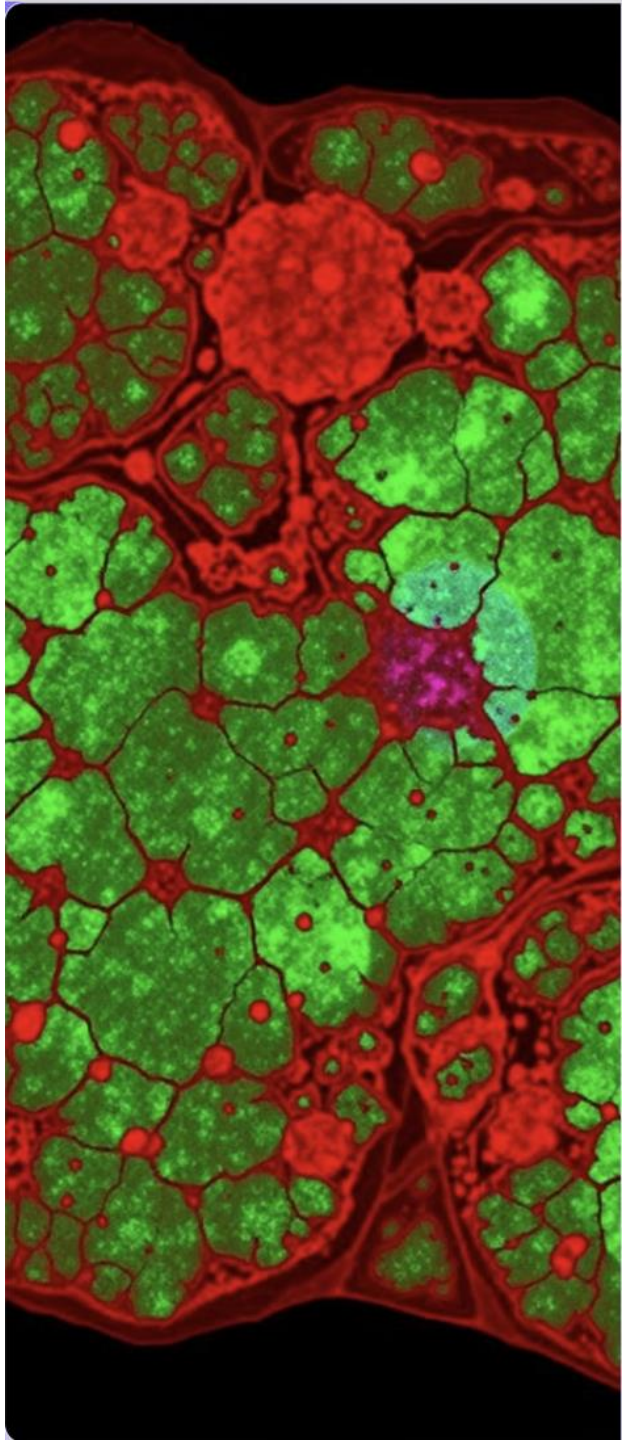
Is a Biopsy Required?

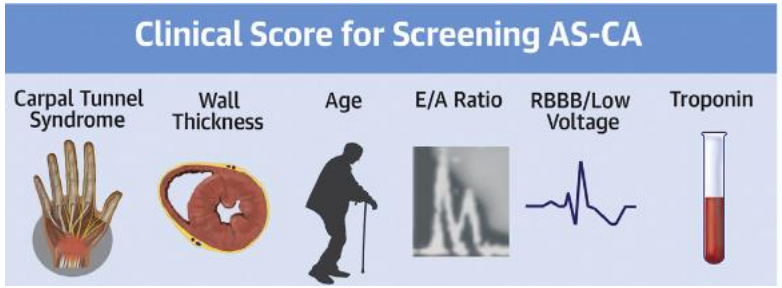
1. Technetium-labelled tracers bind to amyloid deposits.
2. They are a non-invasive alternative to biopsy
3. 1217 pts – bone scan, biochemical and biospies
4. Diagnostic accuracy?
5. Sensitivity: 99%, Specificity 86%, False Pos in AL
   Combine with monoclonal protein
6. Bone scintigraphy is a reliable diagnosis tool in ATTR CA



Julian Gillmore. Circulation 2016

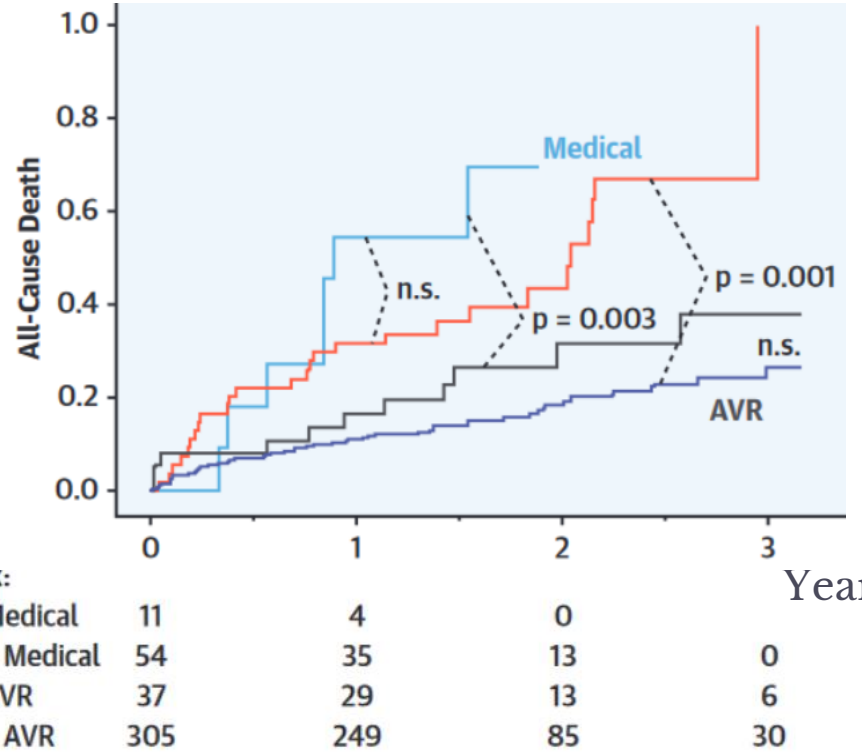
Prevalence & Prognosis

1. 408 pts for TAVI, 48 CA
   - Prevalence 4-25%
2. Raise Score > 2
   - Discrimination lone AS vs AS/CA
3.



4. CA is a disease modifier in AS
   Intervention not withheld

All-Cause Mortality



Nitsche Christian. JACC 2021

Management Implications

1

Early Detection

Crucial for timely intervention and prognosis

2

Targeted Therapy

Amyloid-specific treatments

3

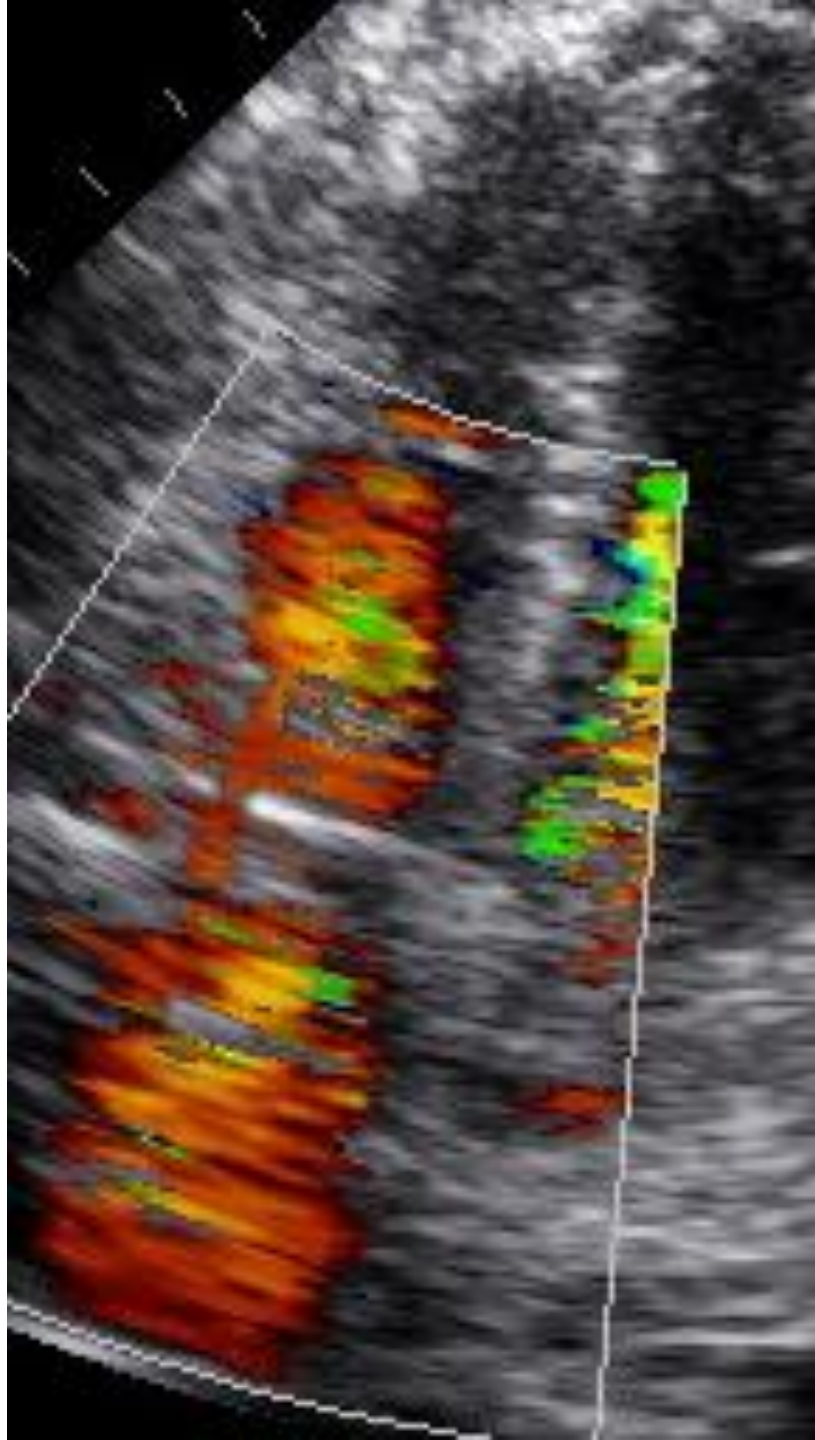
Surgical Considerations

Higher risk for TAVR/SAVR in amyloidosis

4

Multidisciplinary Approach

Collaboration between cardiology, hematology and genetics




SYSTEM OVERVIEW


AI


AI

Conclusions

Patient Group	Mortality (1+ year follow-up)
AS-CA	33%
AS Alone	22%

1. CA 4-25% prevalence (age groups studied)
   AS-CA older, male, carpal tunnel
2. Clinical Impact: higher mortality and HF burden
3. AS/CA altered treatment response: AVR and CA therapy
   TAVI benefits both AS-CA and AS alone pts
4. Need for improved screening strategies for early detection
   The RAISE score can effectively predict AS-CA
5. Multidisciplinary approach
   Card – CAH – Rad – Nuc – Gen – Haem
6. Optimising timing and therapy and improve outcome
   AS-CA: TAVI and ATTR-CA specific therapy

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