

## Puzzling cases to learn from: Read with the experts

### A Bad combination!

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**No Disclosures**



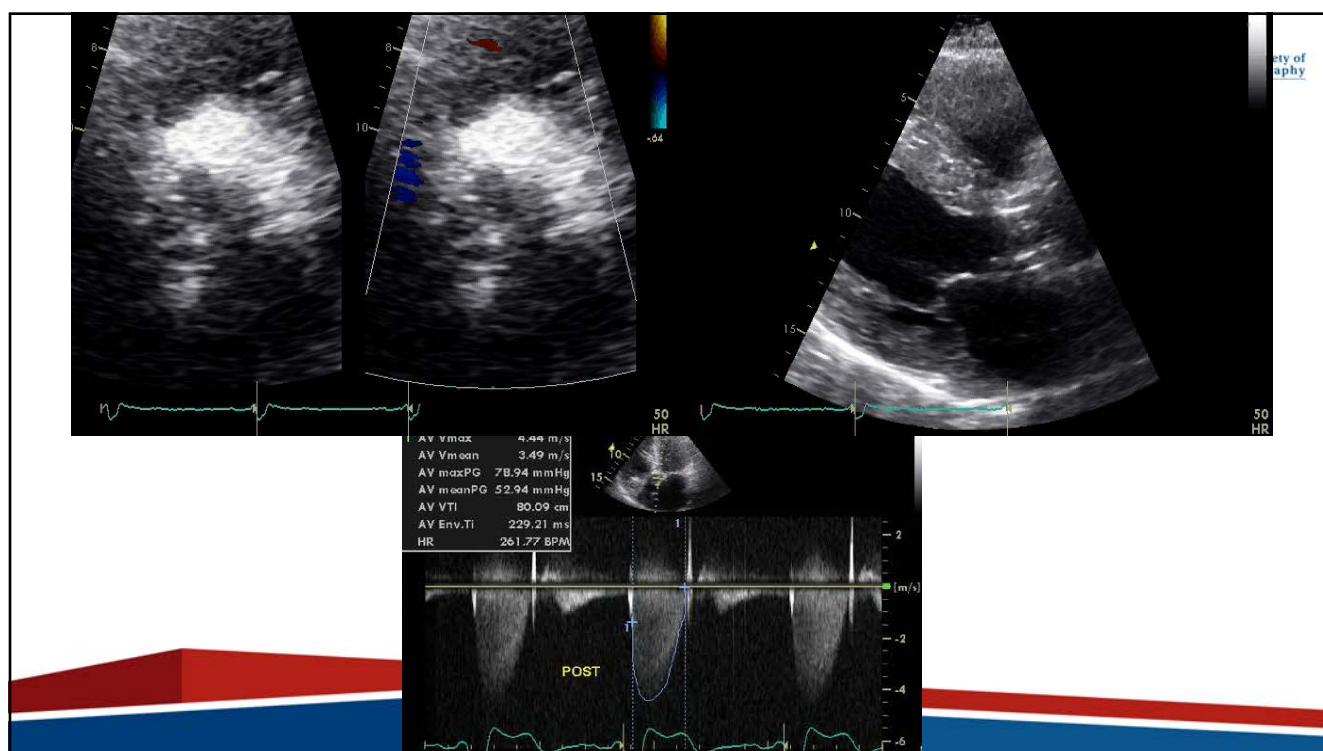
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## Case 1

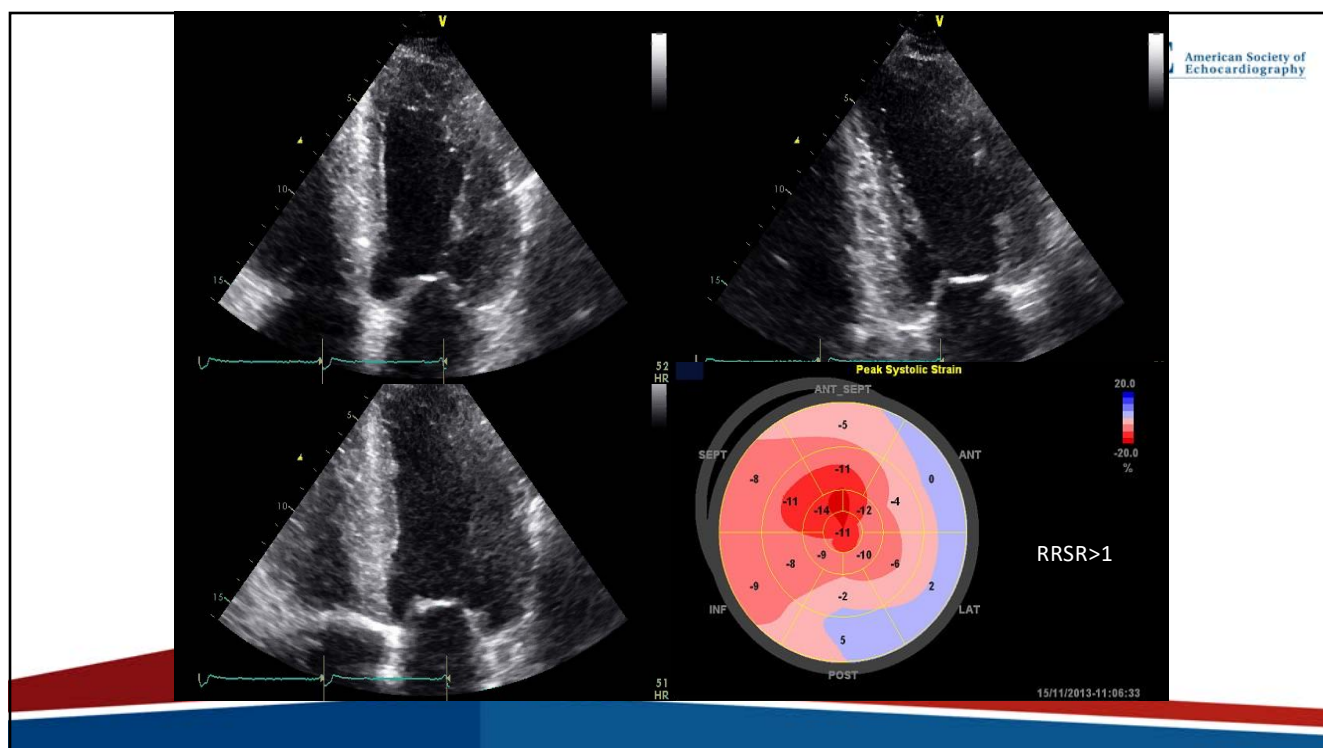
**54 year old male presents with recurrent hospital admissions for congestive cardiac failure.**

**History of congenital AV disease s/p commissurotomy at age 18 and AVR (#21 bioprosthetic valve) in 2007 for severe AS, HTN and Stage III CKD.**

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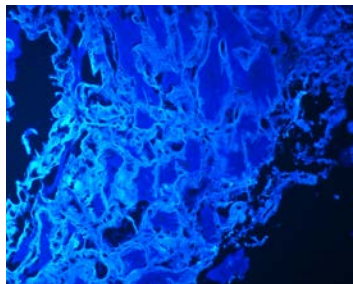
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## Cardiac biopsy



Interstitial infiltrates positive in the **Thioflavin S stain** and identified as **amyloid**.

Identification of the subtype of amyloid by immunohistochemistry shows Transthyretin positivity.



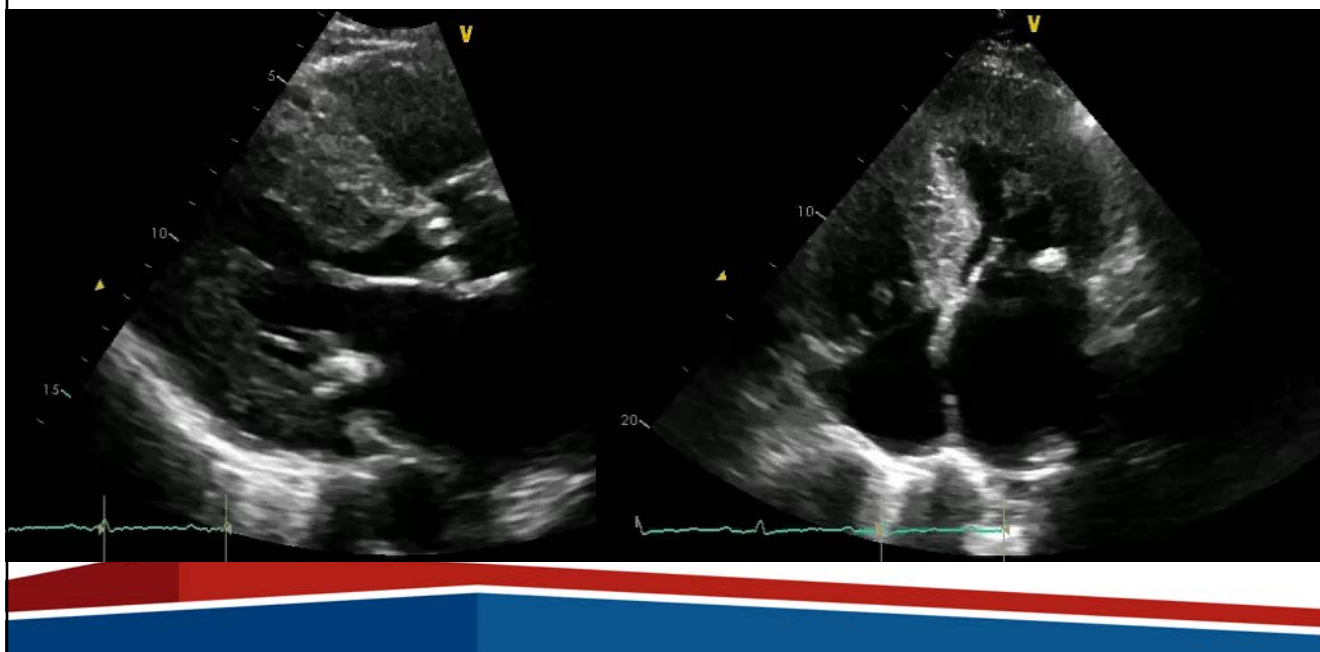
5

## Case 2



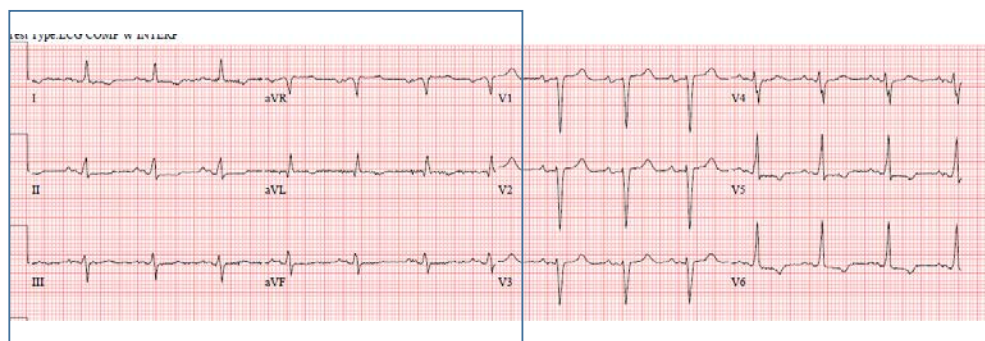
**77 year old African American female presents with recurrent hospital admissions for congestive cardiac failure.**

6



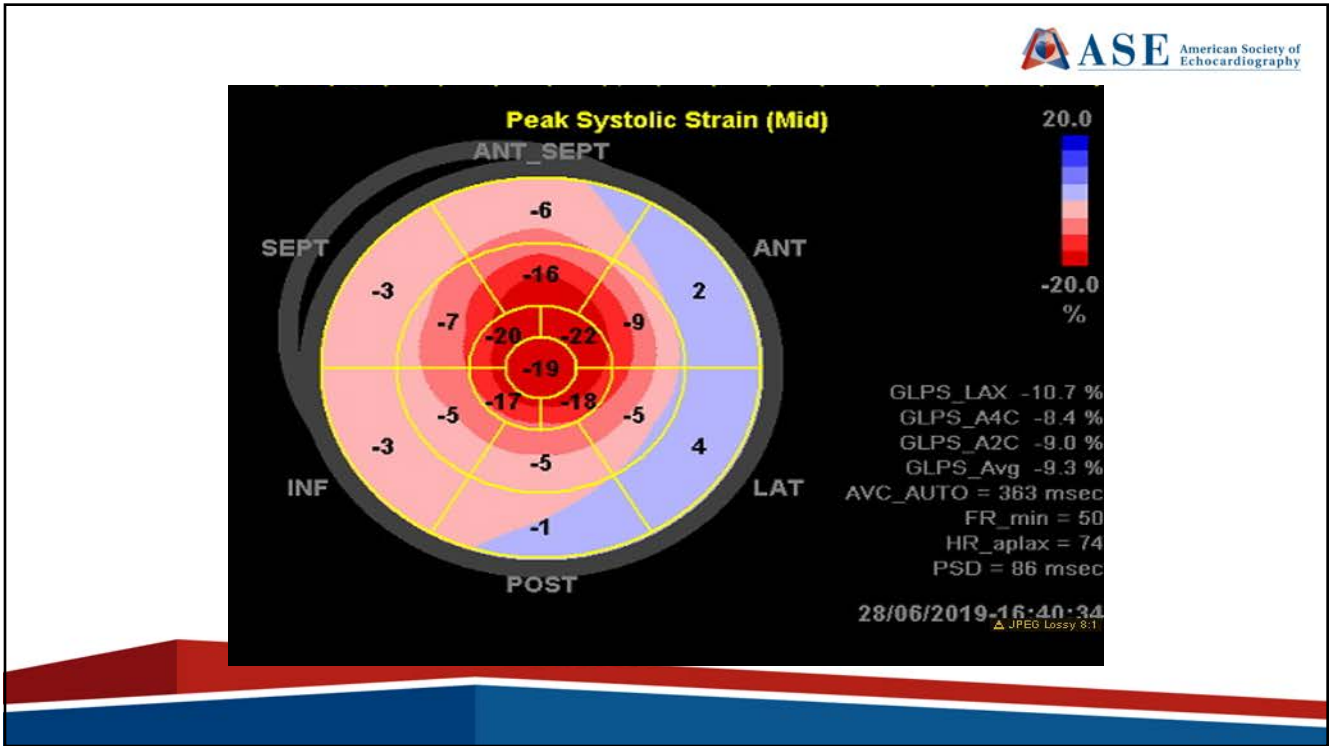
7

## 12 lead ECG

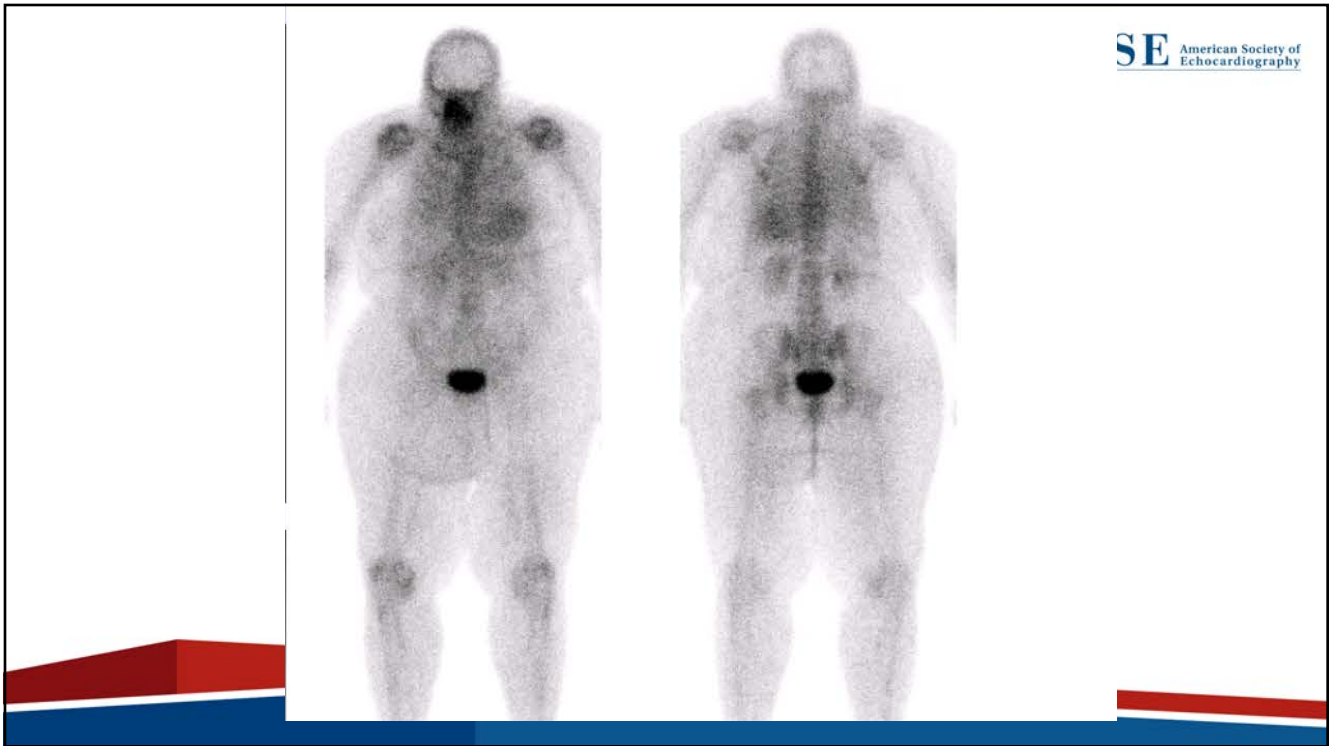


AND..... → 78 yr old African American ♀

8

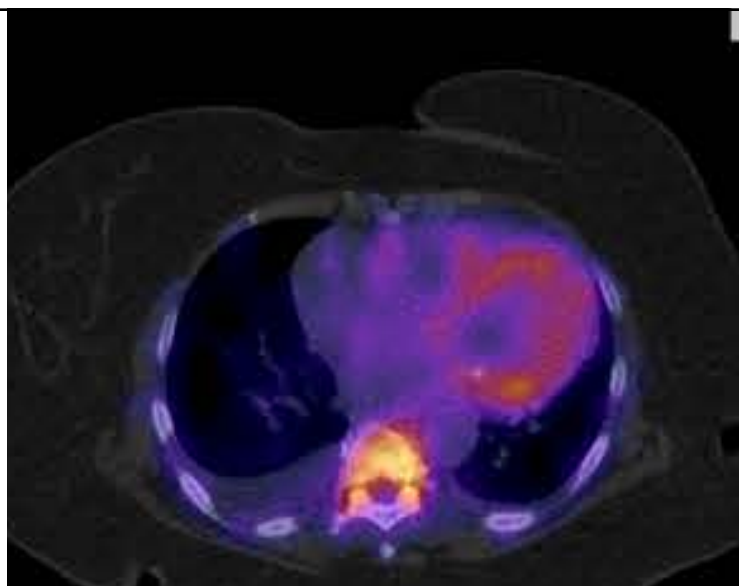


9



10





Genetic testing positive for V122I mutation

11

## Aortic Stenosis and Transthyretin Cardiac Amyloid

- What is the prevalence of amyloid heart disease in elderly patients with severe aortic stenosis?
- Do they benefit from replacing the aortic valve?

12



ACC.16

ASE American Society of  
Echocardiography1535  
JACC April 5, 2016  
Volume 67, Issue 13

Heart Failure and Cardiomyopathies

TRANSTHYRETIN CARDIAC AMYLOIDOSIS IS PREVALENT AMONG MEN WITH SEVERE AORTIC STENOSIS UNDERGOING TRANSCATHETER AORTIC VALVE REPLACEMENT

- 75 patients with severe AS underwent 99mTc-PYP planar cardiac imaging within 30 days of TAVR.
- 99mTc-PYP uptake was found in 16%.
- 80 % in a phenotype of severe concentric left ventricular hypertrophy and low flow AS.

Costano *et al* JACC 2016

13

## Aortic Stenosis and Transthyretin Cardiac Amyloid - Prognosis

ASE American Society of  
Echocardiography

**A retrospective analysis was performed of 171 consecutive patients at Cleveland Clinic with cardiac amyloidosis of ATTR type.**

- Patients with moderate or severe aortic stenosis were included in the analysis.
- Low flow low gradient aortic stenosis was considered severe if the aortic valve area was  $\leq 1.0 \text{ cm}^2$  with a dimensionless index  $\leq 0.25$  and either ejection fraction  $\leq 50\%$  or LVOT SVI  $\leq 35 \text{ mL/m}^2$ .

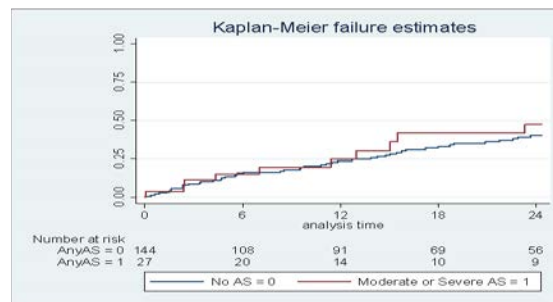
Sperry *et al* JACC imaging April 2016

14

## Aortic Stenosis and Transthyretin Cardiac Amyloid - Prognosis



**Mortality was 37% in patients with aortic stenosis and 33% in those without aortic stenosis (HR 1.22 [CI 0.62-2.42],  $p = 0.566$ ).**



**Mortality in patients with both diseases is more likely driven by ATTR amyloidosis as opposed to a stenotic aortic valve or its treatment**

*Sperry et al JACC imaging April 2016*

15

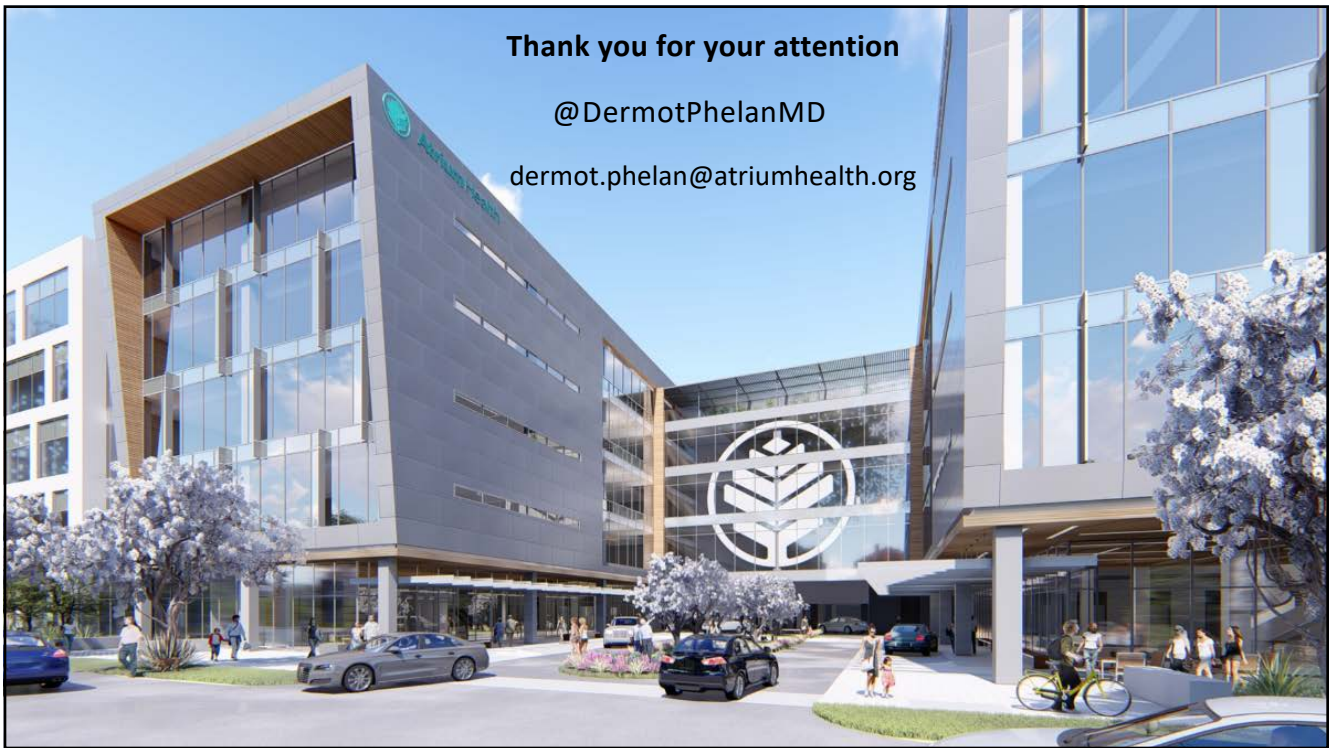
## Follow up



- **Case 1 TAVR 2018 – Alive but still has HF symptoms**
- **Case 2 TAVR Jan 2019 – RIP 6 months later from HF**

16





17