Sitting in with the heart valve team: One last hurrah

"Nobody takes a proper history anymore!" Dr Harry Lever

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No Disclosures

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29 year old male with 1 year of low back pain and 1 month of progressive dyspnea on exertion

Present to ED for an episode of worsening chest pain and low grade fever

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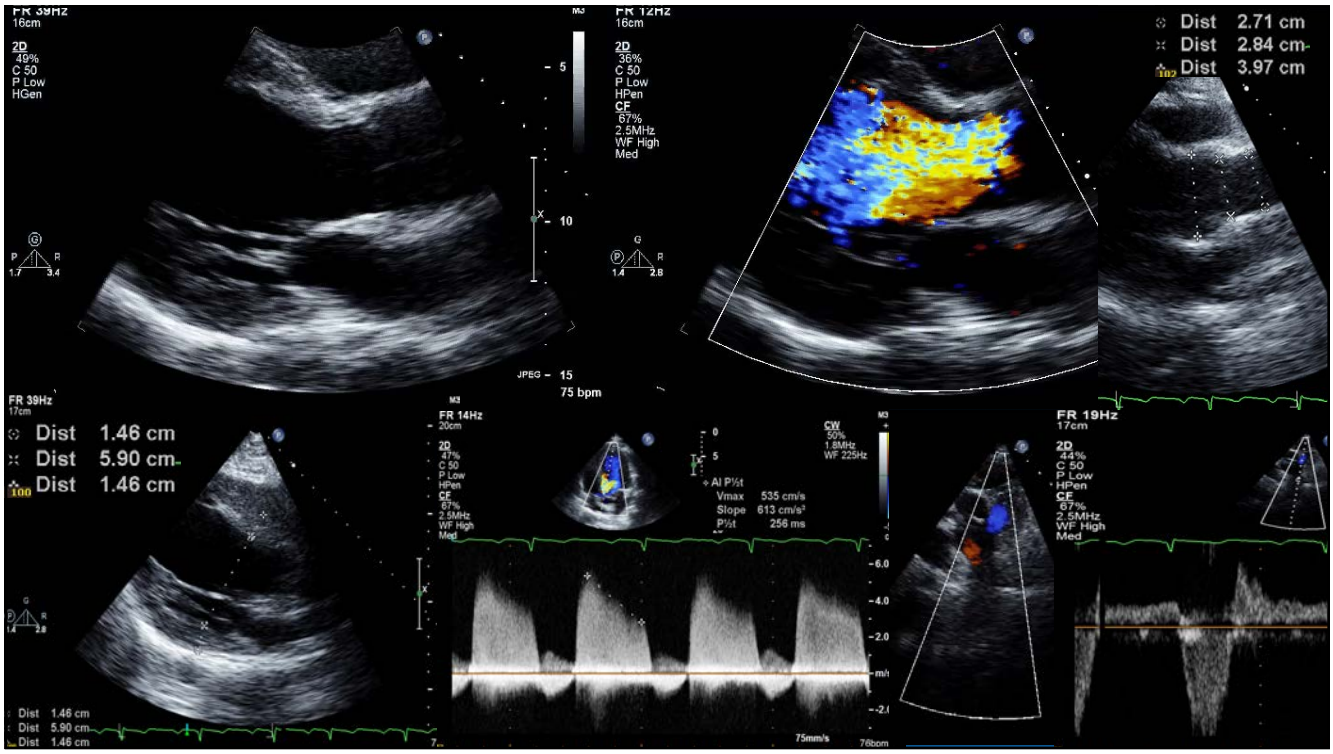

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ASE American Society of Echocardiography

OPERATION: Aortic valve replacement with #25 St. Jude mechanical prosthesis.

ANESTHESIA: General endotracheal.

PREOPERATIVE DIAGNOSES: Severe aortic insufficiency and dilated aortic root.

POSTOPERATIVE DIAGNOSES: Acute aortitis with aortic root inflammation, and severe aortic insufficiency.

OPERATIVE INDICATIONS: The patient is a 29-year-old gentleman who has had progressive exertional dyspnea. He is found to have severe aortic insufficiency and mild-to-moderate dilatation of the aortic sinuses. Preoperatively, I felt that this may be aortic insufficiency secondary to aortic sinus dilatation. However, on inspection, we found that there was impressive aortic root inflammation and edema with a white layer of scar that covered the sinus of Valsalva, the sinotubular junction, but not much beyond and covered most of the aortic valves. There also was some inflammation that extended into the LV outflow tract. The coronary ostia were free of any significant disease.

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Pathology

AORTA

MILDLY ACTIVE AND MOSTLY HEALED AORTITIS

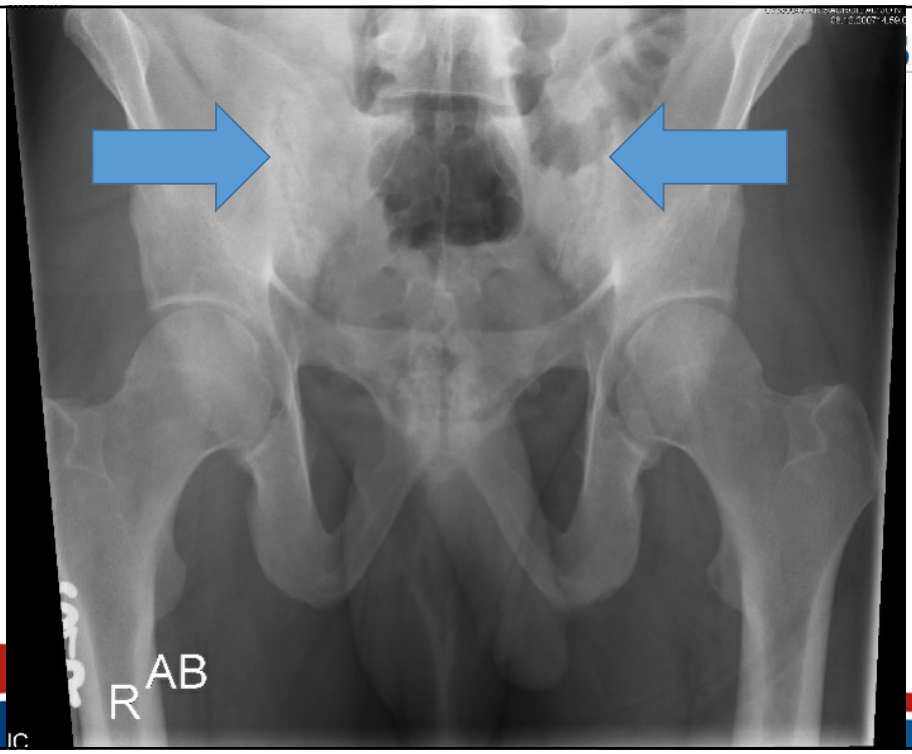
AORTIC VALVE

HEALED VALVULITIS AND MODERATE FIBROSIS

Thoughts on Diagnosis?

5

HLA-B27 POSITIVE



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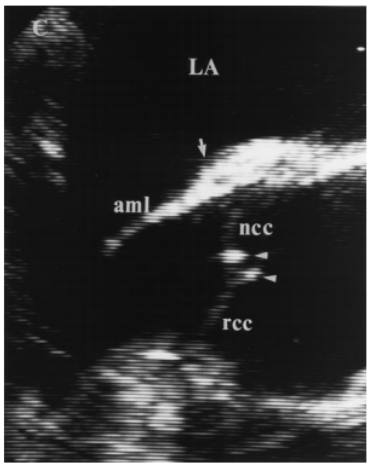
Aortic Root Disease and Valve Disease Associated With Ankylosing Spondylitis

American Society of Echocardiography

Table 3. Frequency of Aortic Root and Valve Abnormalities on Transesophageal Echocardiography

	Patients (n = 44)		Controls (n = 30)		
Abnormality	n	%	n	%	p Value
Aortic root abnormalities					
Thickening	27	61	2	7	<0.001
Dilatation	11	25	2	7	0.06
Abnormal Ep or stiffness	27	61	3	10	<0.001
Valve abnormalities					
Thickening	21	48	3	10	<0.001
Aortic	18	41	3	10	0.04
Mitral	15	34	1	3	<0.001
Subaortic bump	11	25	0	0	0.002
Regurgitation	20	45	1	3	<0.001
Aortic	7	16	0	0	0.02
Mitral	14	32	1	3	0.003
Tricuspid	1	2	0	0	
Any	36	82	8	27	<0.001

Ep = Peterson's pressure–strain elastic modulus.



Subaortic "hump"

Roldan et al., JACC, 1998;32

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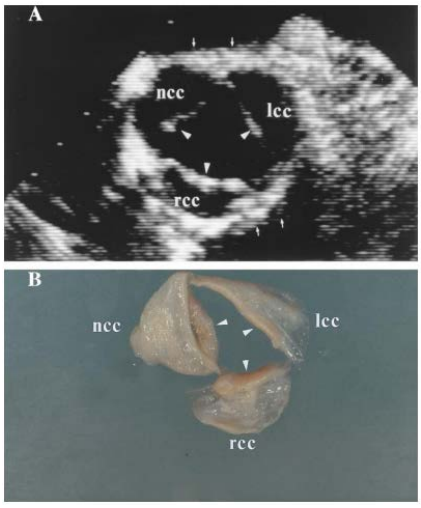

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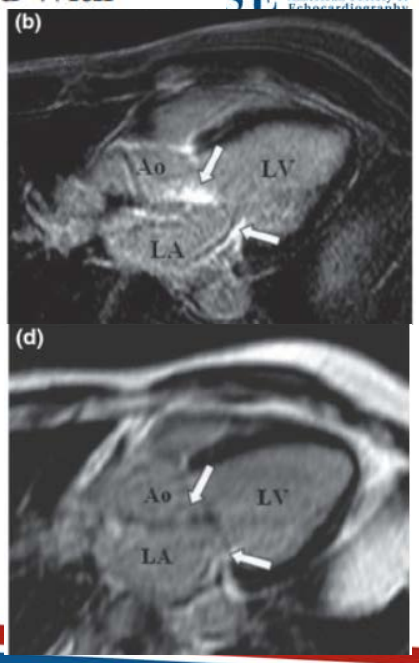

Follow up of 25 patients: 24% with new aortic root or valve abnormalities, 12% with worsening valuvular regurgitation, 20% with resolution of abnormalities

Roldan et al., JACC, 1998;32

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Management

- **Immunosuppression**
- **Surveillance of aortitis/aneurysm and valve disease with periodic history/exam and imaging**
- **Valve replacement according to AHA/ACC guidelines**

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Follow up

Started on methotrexate and prednisone 1 week post-operatively, eventually transitioned to adalimumab (Humira)

Serial chest imaging:

- 12/2011 Pre-op CTA Sinus 4.2cm
- 1/2016 cMRI Sinus 4.9cm
- 9/2017 cMRI Sinus 4.9cm
- 11/2018 CTA Sinus 5.5cm

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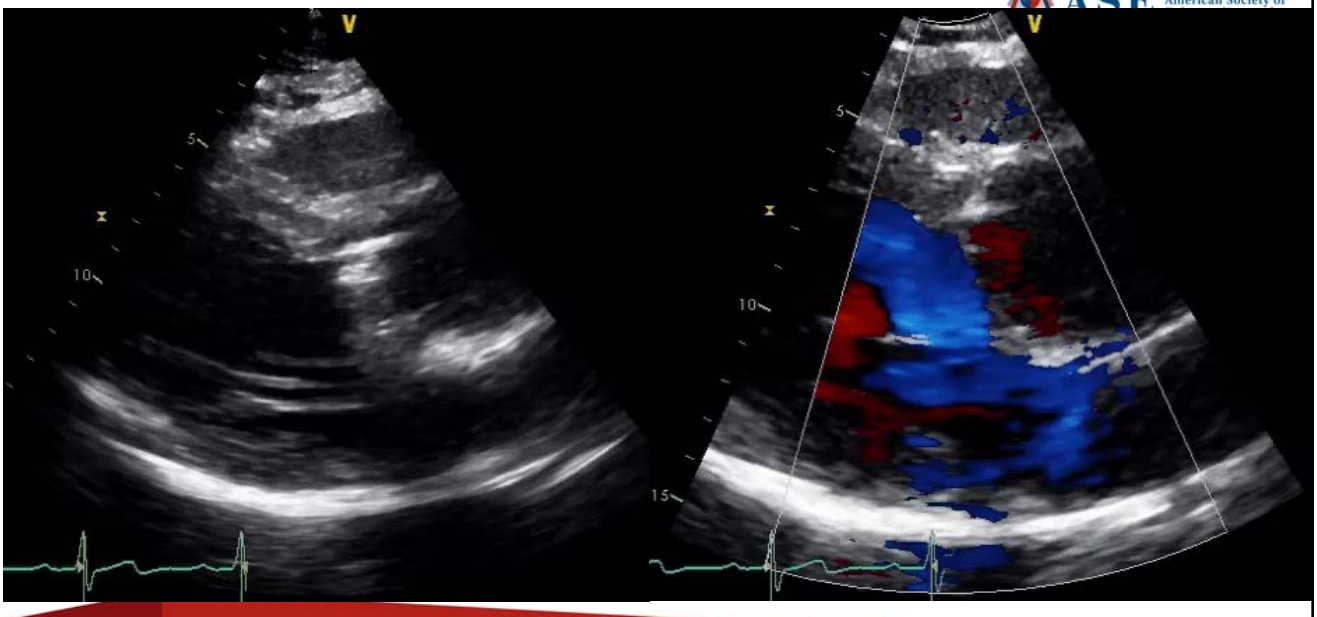

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Surgical Management of Aortic Regurgitation Associated With Takayasu Arteritis and Other Forms of Aortitis

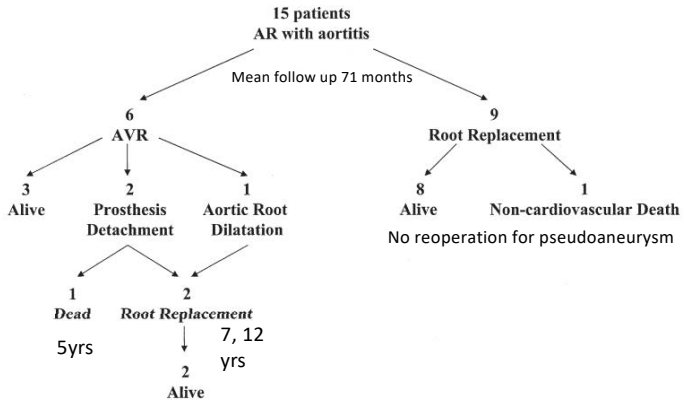

Fig 1. Surgical outcomes for 15 patients with aortic regurgitation (AR) associated with aortitis. (AVR = aortic valve replacement.)

Adachi et al., Ann Thorac Surg, 2007;84

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Follow up

- Underwent redo aortic root replacement and AVR composite graft with 29mm On-X mechanical valve and 32mm Gelweave Valsalva conduit
- Continued on adalimumab

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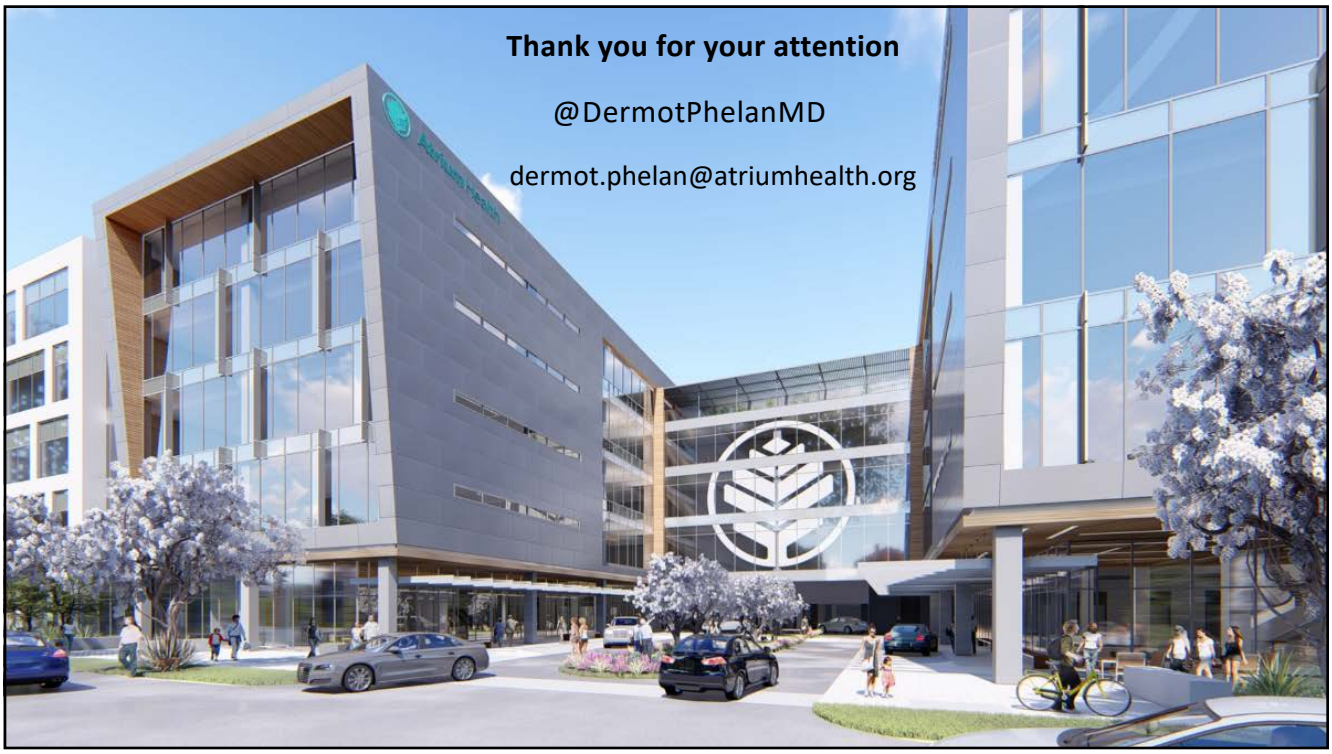
Thank you for your attention
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