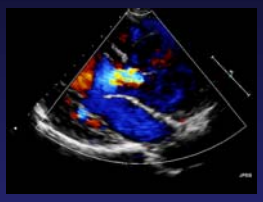
Case Studies: Morphology of Aortic Regurgitation, Root Involvement



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Medical Director, Echocardiography Laboratory
Northwestern Memorial Hospital
President, American Society of Echocardiography

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No Disclosures

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Introduction

- Aortic regurgitation may be caused by primary disease of the aortic valve leaflets and/or the wall of the aortic root.
- AR due to dilatation of the ascending aorta is now more common than primary valve disease in pts undergoing AVR for isolated AR.

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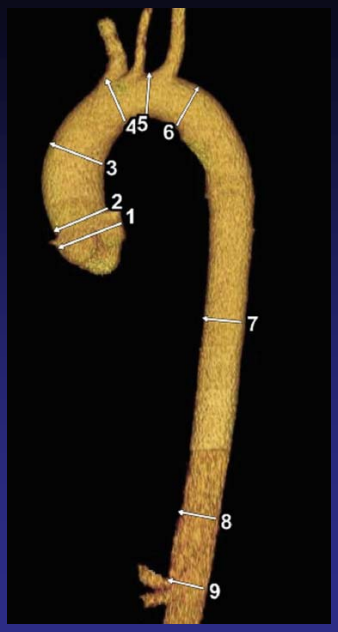

Anatomic Location	
1.	Aortic sinuses of Valsalva
2.	Sinotubular junction
3.	Mid ascending aorta (midpoint in length between Nos. 2 and 4)
4.	Proximal aortic arch (aorta at the origin of the innominate artery)
5.	Mid aortic arch (between left common carotid and subclavian arteries)
6.	Proximal descending thoracic aorta (begins at the isthmus, approximately 2 cm distal to left subclavian artery)
7.	Mid descending aorta (midpoint in length between Nos. 6 and 8)
8.	Aorta at diaphragm (2 cm above the celiac axis origin)
9.	Abdominal aorta at the celiac axis origin

AHA/ACC 2010 Guidelines for Thoracic Aortic Disease

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Gene Defects Associated with Familial Thoracic Aortic Aneurysm and Dissection

Defective Gene Leading to Familial Thoracic Aortic Aneurysms and Dissection	Contribution to Familial Thoracic Aortic Aneurysms and Dissection	Associated Clinical Features	Comments on Aortic Disease
TGFBR2 mutations	4%	Thin, translucent skin Arterial or aortic tortuosity Aneurysm of arteries	Multiple aortic dissections documented at aortic diameters <5.0 cm
MYH11 mutations	1%	Patent ductus arteriosus	Patient with documented dissection at 4.5 cm
ACTA2 mutations	14%	Livedo reticularis Iris flocculi Patent ductus arteriosus Bicuspid aortic valve	Two of 13 patients with documented dissections <5.0 cm

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Genetic syndromes associated with thoracic aortic aneurysm and dissecxtion

Genetic Syndrome	Common Clinical Features	Genetic Defect
Marfan syndrome	Skeletal features (see text) Ectopia lentis Dural ectasia	*FBN1* mutations*
Loeys-Dietz syndrome	Bifid uvula or cleft palate Arterial tortuosity Hypertelorism Skeletal features similar to MFS Craniosynostosis Aneurysms and dissections of other arteries	*TGFBR2* or *TGFBR1* mutations
Ehlers-Danlos syndrome, vascular form	Thin, translucent skin Gastrointestinal rupture Rupture of the gravid uterus Rupture of medium-sized to large arteries	*COL3A1* mutations
Turner syndrome	Short stature Primary amenorrhea Bicuspid aortic valve Aortic coarctation Webbed neck, low-set ears, low hairline, broad chest	45,X karyotype

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Risk factors for thoracic aortic dissection

Conditions Associated With Increased Aortic Wall Stress
Hypertension, particularly if uncontrolled
Pheochromocytoma
Cocaine or other stimulant use
Weight lifting or other Valsalva maneuver
Trauma
Deceleration or torsional injury (eg, motor vehicle crash, fall)
Coarctation of the aorta
Conditions Associated With Aortic Media Abnormalities
Genetic
Marfan syndrome
Ehlers-Danlos syndrome, vascular form
Bicuspid aortic valve (including prior aortic valve replacement)
Turner syndrome
Loeys-Dietz syndrome
Familial thoracic aortic aneurysm and dissection syndrome
Inflammatory vasculitides
Takayasu arteritis
Giant cell arteritis
Behçet arteritis
Other
Pregnancy
Polycystic kidney disease
Chronic corticosteroid or immunosuppression agent administration
Infections involving the aortic wall either from bacteremia or extension of adjacent infection

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Mechanisms of AR due to abnormalities of the ascending aorta

- Dilatation of the aortic annulus
  - **separation of the leaflets**
  - **Tension and bowing of the leaflets result in thickening and retraction**
- Aortic dissection

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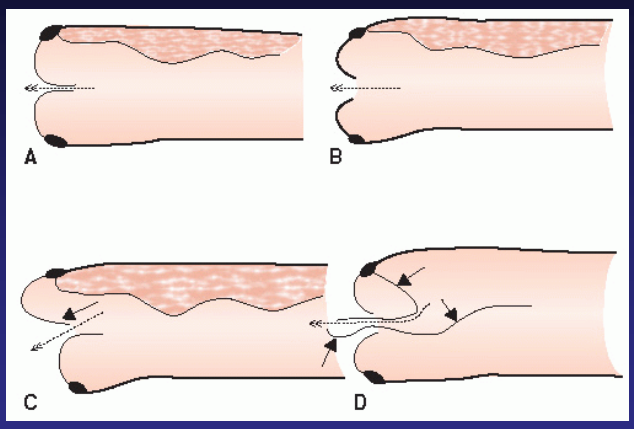
Mechanisms of AR in Aortic Dissection



Feigenbaum's Echocardiography, 7th ed. 2010

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History

- 29 yr old female with Marfan's syndrome
- Admitted with dyspnea and LE edema
- Diagnosed with an aortic aneurysm, chronic type A dissection and AR
- Presented to CT surgeon's office with c/o chest pain

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History

- PMH:
  - **Multiple sclerosis**
  - **Asthma**
  - **Scoliosis**
  - **Dislocated lens**
- FH
  - **Mother with Marfan's – died during aortic surgery**

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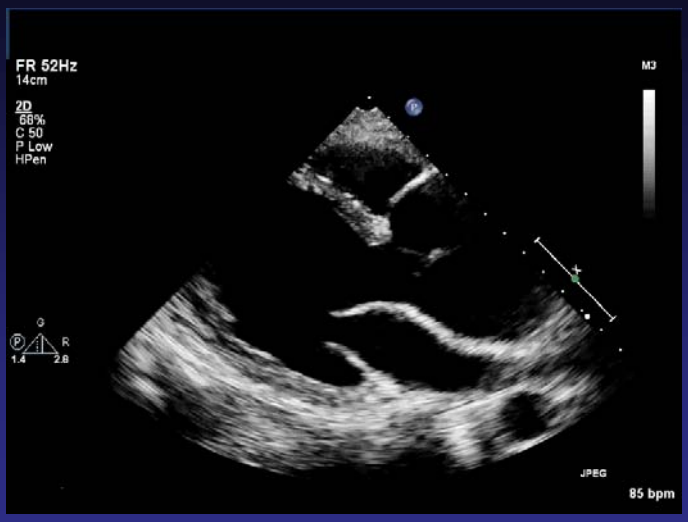

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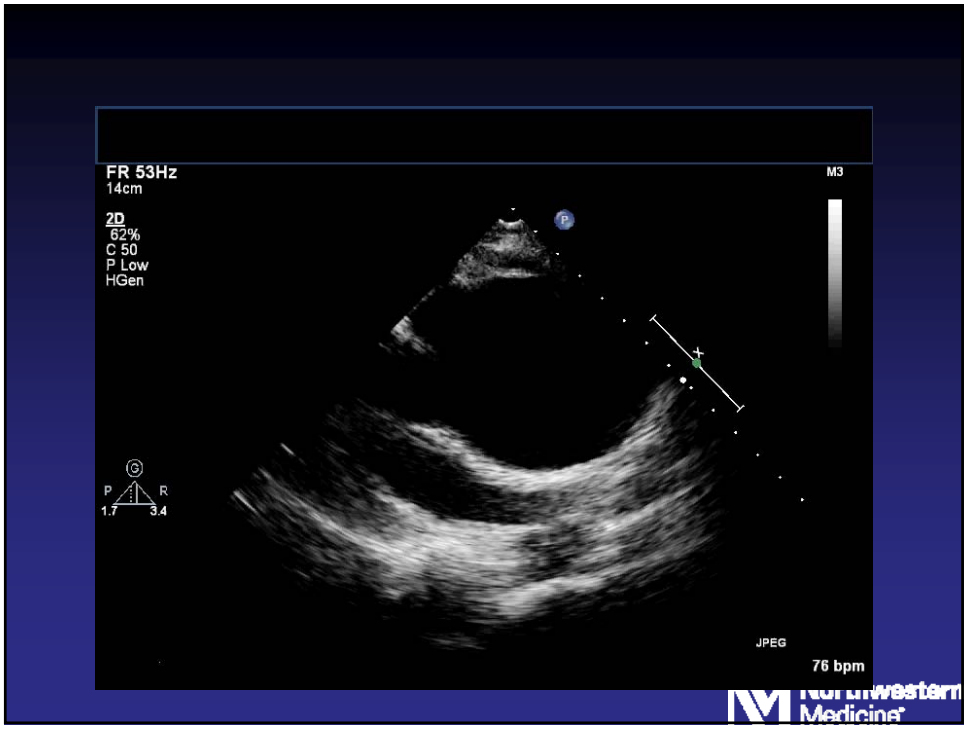
FR 53Hz
14cm
2D
62%
C 50
P Low
HGen
M3
P
R
1.7
3.4
JPEG
76 bpm
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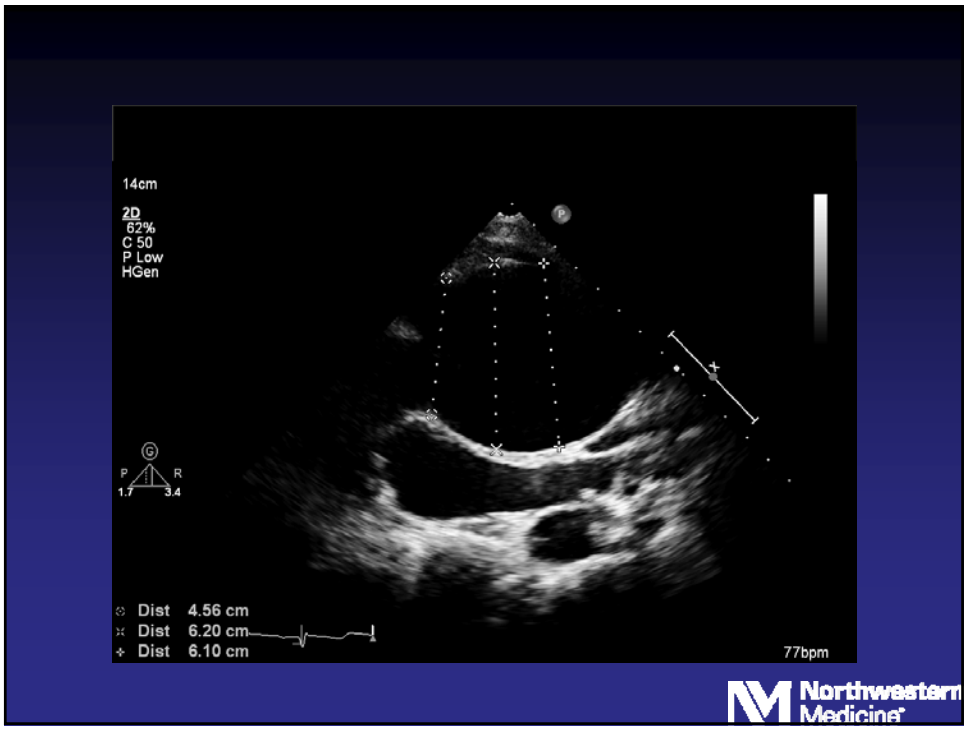
14cm
2D
62%
C 50
P Low
HGen
P
R
1.7
3.4
Dist 4.56 cm
Dist 6.20 cm
Dist 6.10 cm
77bpm
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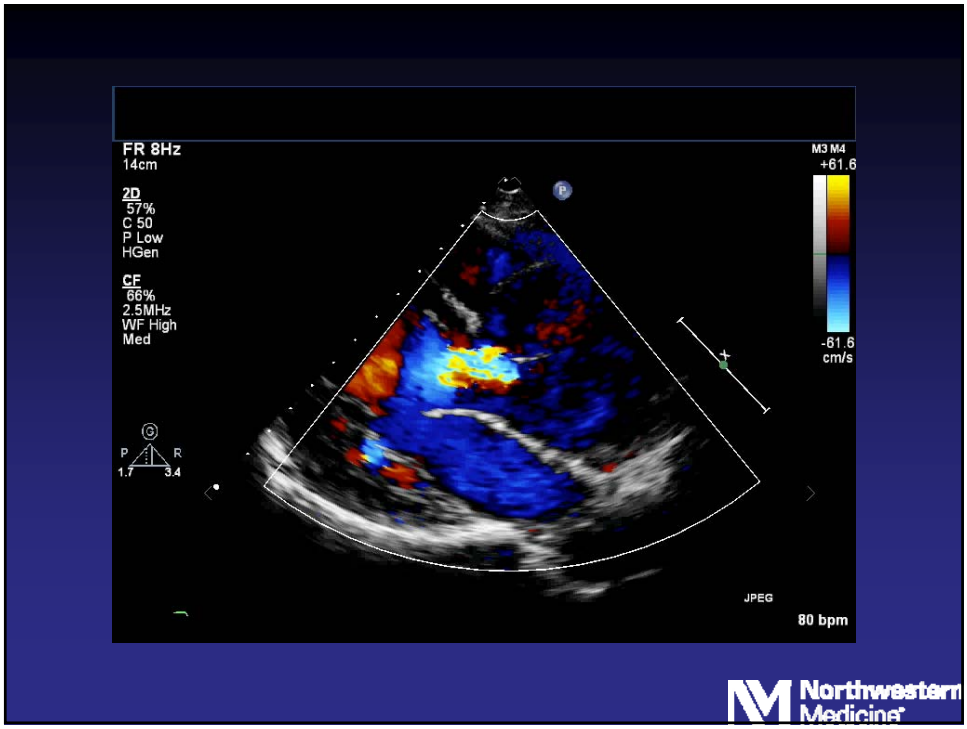
FR 8Hz
14cm
2D
57%
C 50
P Low
HGen
CF
66%
2.5MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
JPEG
80 bpm
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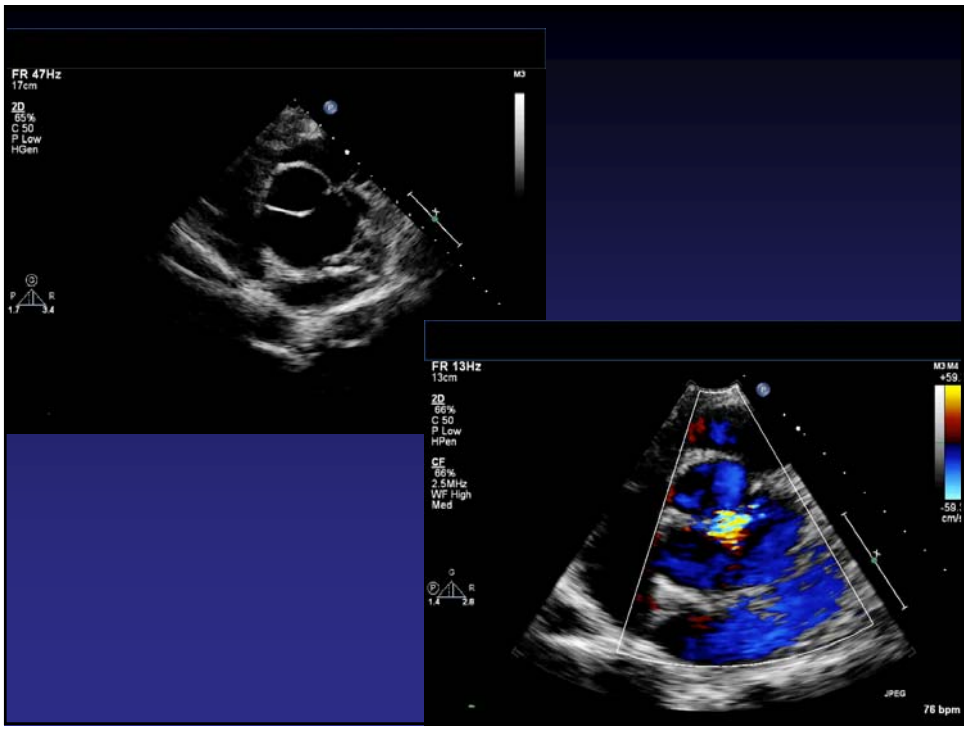
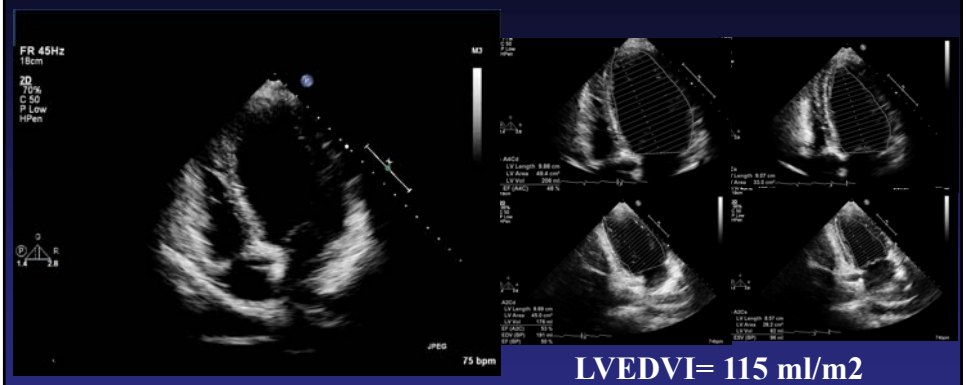
Medicine


FR 47Hz
17cm
2D
65%
C 50
P Low
HGen
M3
FR 13Hz
13cm
2D
66%
C 50
P Low
HPen
CF
66%
2.5MHz
WF High
Med
JPEG
76 bpm



LVEDVI= 115 ml/m2
LVESVI= 58 ml/m2
LVEF=50%

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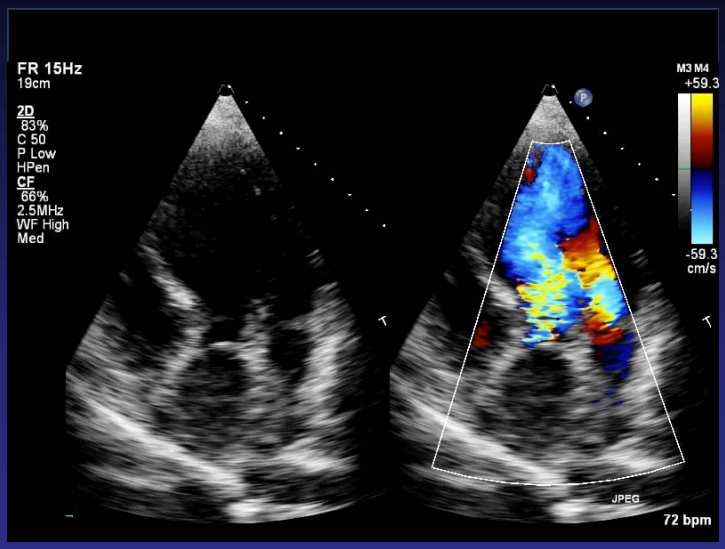

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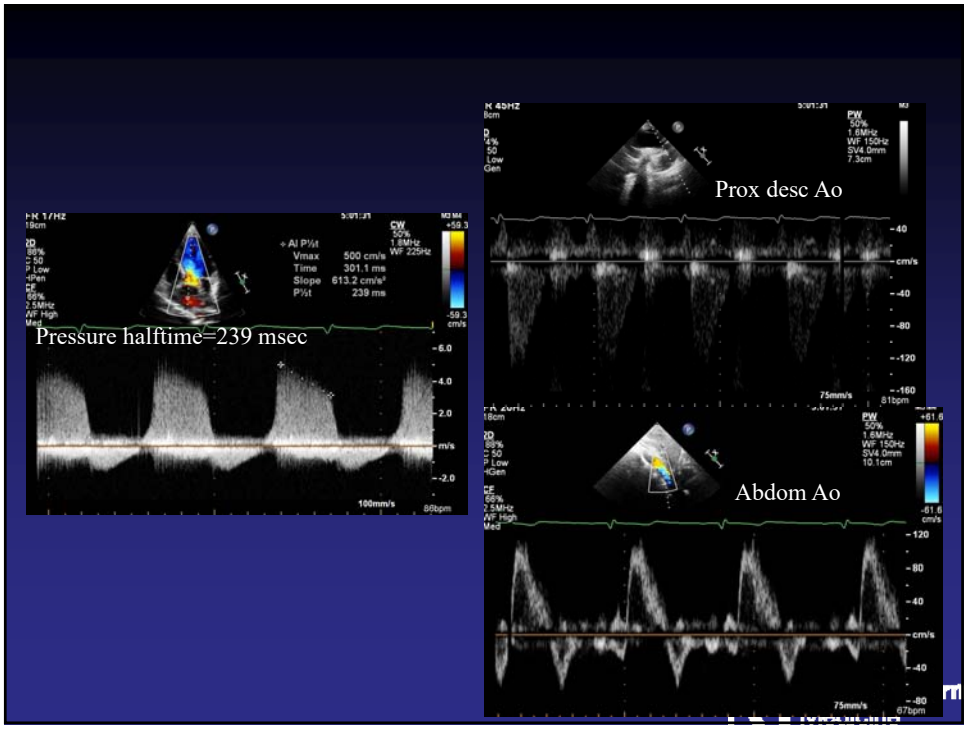
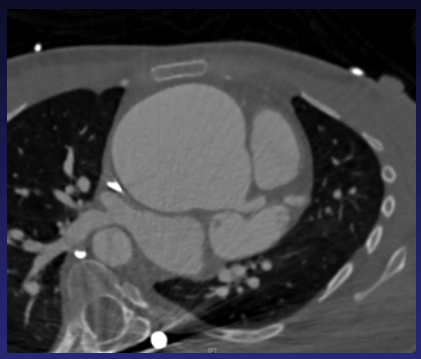


CT of the aorta



Annulus: 2.5 x 3.3 cm
Sinus of Valsalva: 5.3 x 5.2 x 5.3 cm
Sino-tubular junction: 6 x 6 x 6.2 cm
Mid ascending aorta: 6.7 x 6.4 cm
Proximal aortic arch: 3.1 x 3.1 cm
Distal aortic arch: 2.5 x 2.4 cm
Lower descending thoracic aorta: 1.8 x 1.7 cm

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Surgery

- Aortic root replacement with 27 mm Mechanical valve-graft
- Coronary reimplantation
- Resection and replacement of ascending aorta and total transverse arch using 24 mm dacron graft with reimplantation of inominate artery
- Hypothermic circulatory arrest with antegrade cerebral perfusion via right axillary artery cannulation

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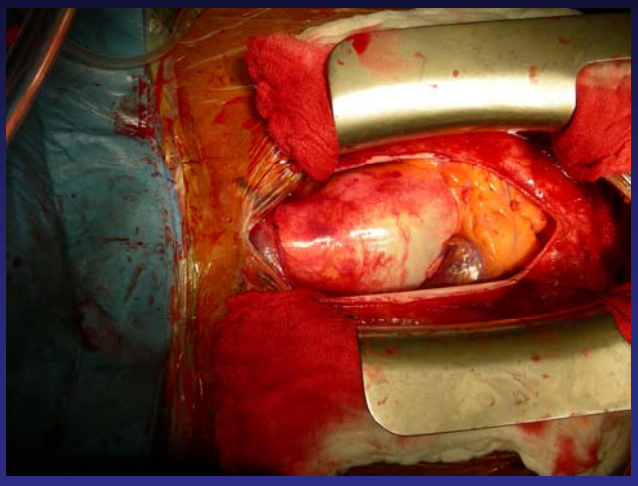

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History

- 65 yr old Asian female who presents to the ER with chest pain
- Patient does not speak English and relays her complaint by pointing to her chest and moaning
- Patient initially thought to have ACS
- Echo ordered when murmur was heard

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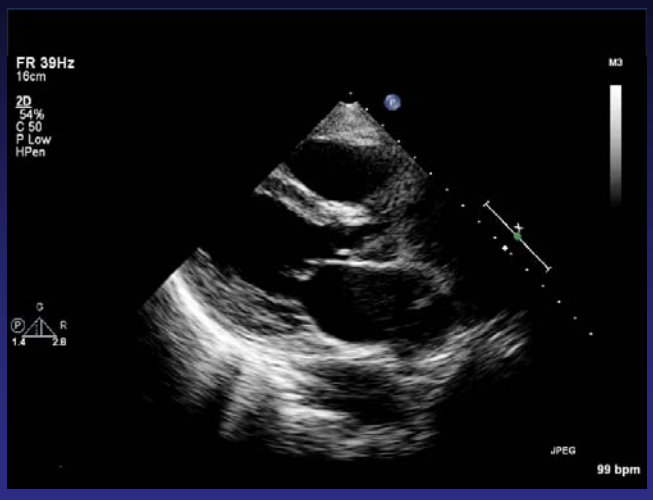

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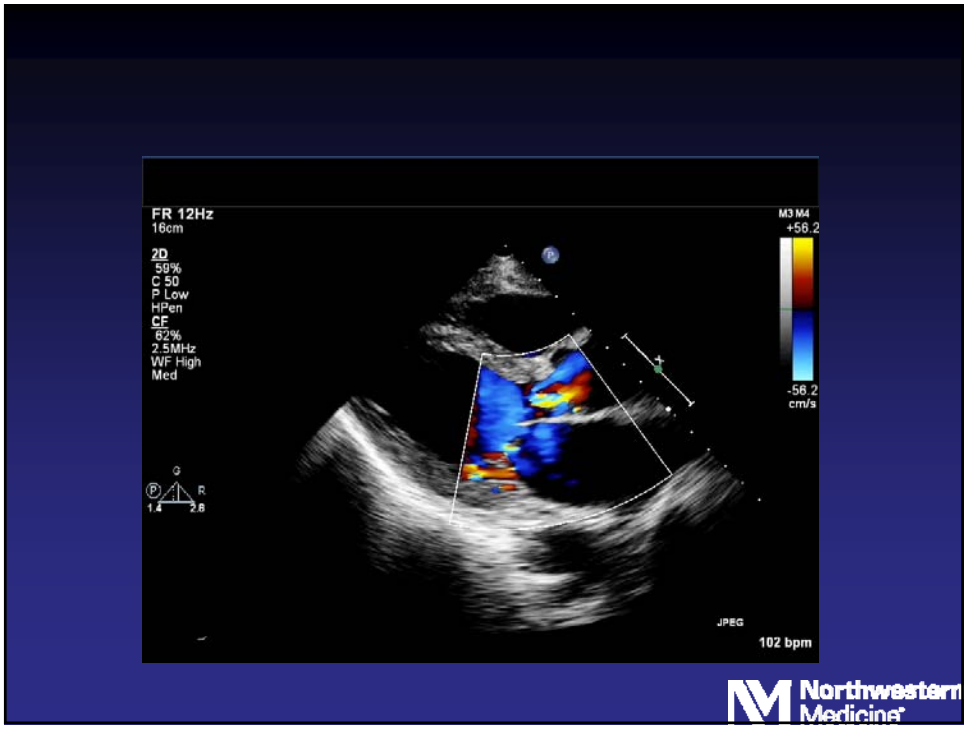
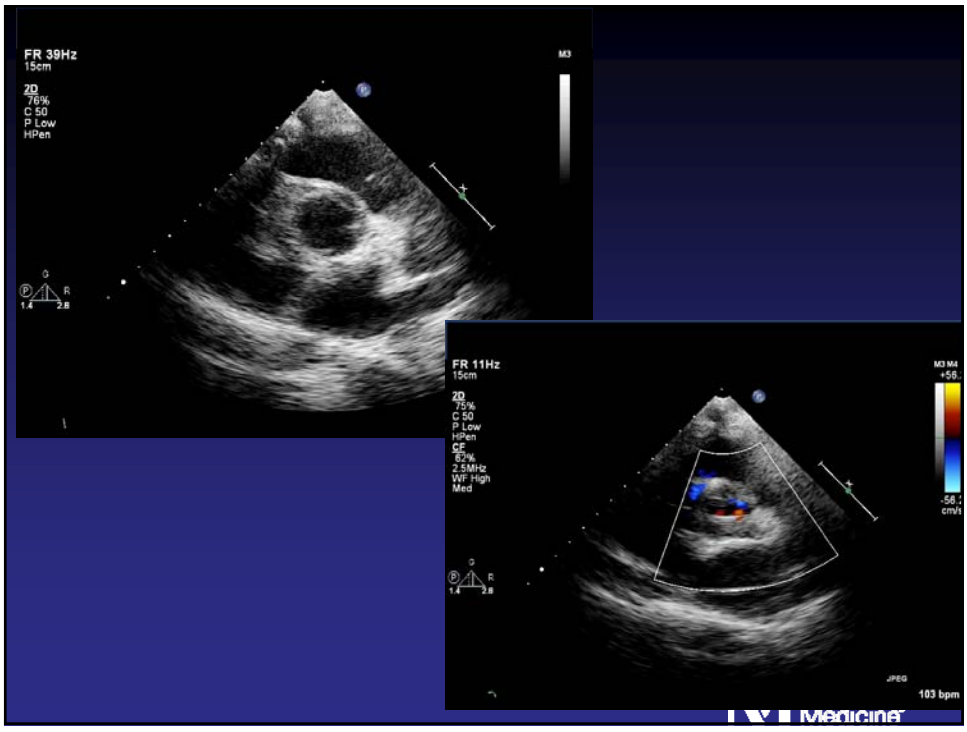
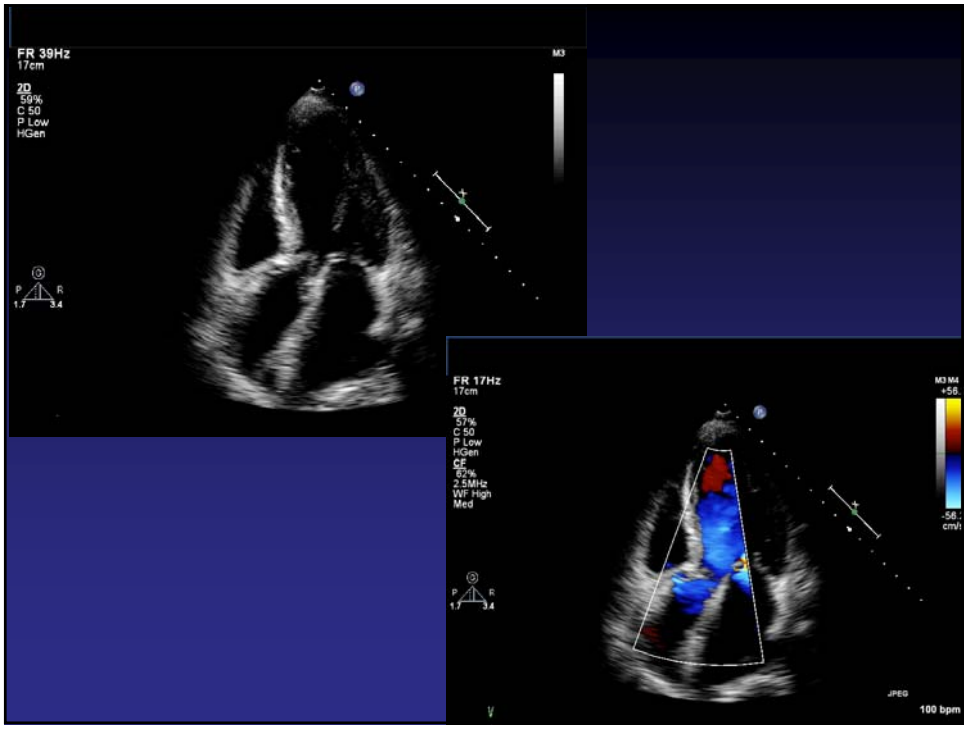

FR 12Hz
16cm
2D
59%
C 50
P Low
HPen
CF
82%
2.5MHz
WF High
Med
M3 M4
+56.2
-56.2
cm/s
JPEG
102 bpm
Northwestern
Medicine


FR 39Hz
15cm
2D
76%
C 50
P Low
HPen
M3
FR 11Hz
15cm
2D
75%
C 50
P Low
HPen
CF
82%
2.5MHz
WF High
Med
M3 M4
JPEG
103 bpm
Medicine



CT scan of the aorta

Stanford type A aortic dissection extending from the aortic valve to just beyond the origin of the left subclavian artery. The dissection also extends into the brachiocephalic artery.

Aneurysmal dilatation of the ascending aorta with a maximal dimension of 4.4 cm (non-orthogonal measurement).

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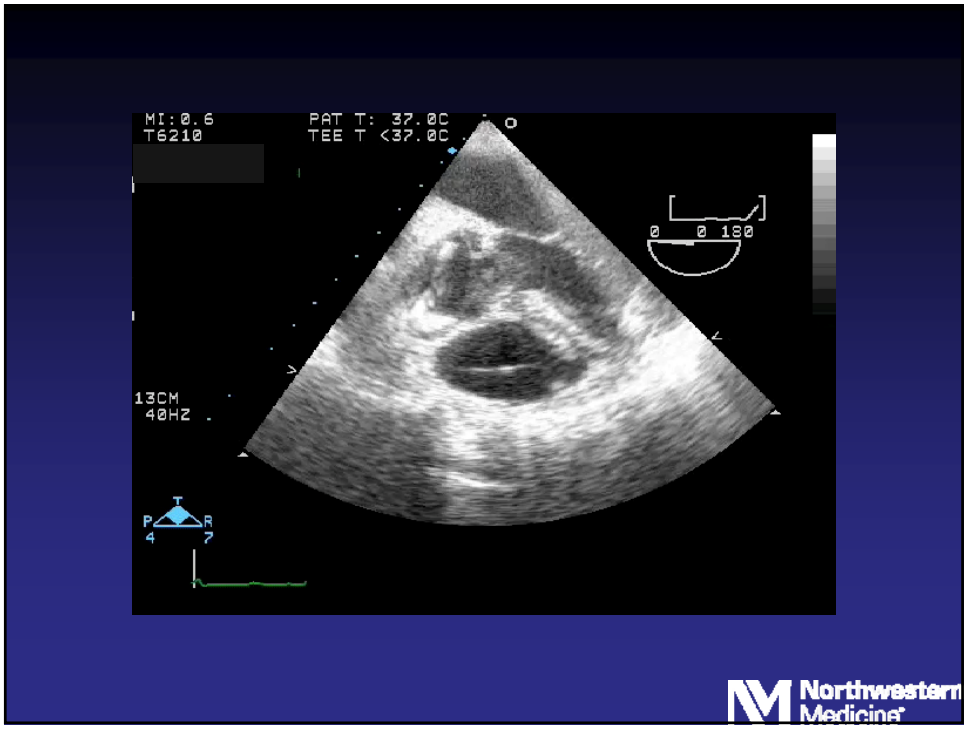
MI:0.6
T6210
PAT T: 37.0C
TEE T <37.0C
0 0 180
13CM
40HZ
T
P R
4 7
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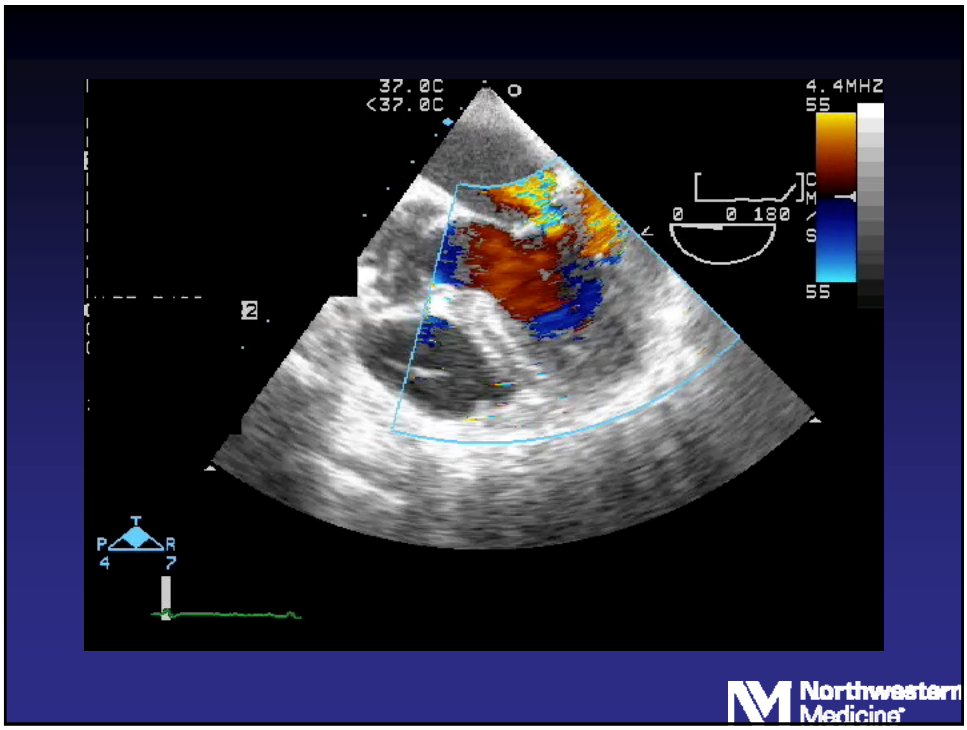
37.0C
<37.0C
4.4MHZ
55
55
0 0 180
T
P R
4 7
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Surgery

- Replacement of aortic root and ascending aorta with a 32 mm Gelweave graft
- Repair of the aortic valve

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History

- 35 yr old male with a known heart valve problem since childhood
- He presented with a febrile illness 8 months prior. Blood cx positive for Strep
- Successfully treated with antibiotics
- Now feels well

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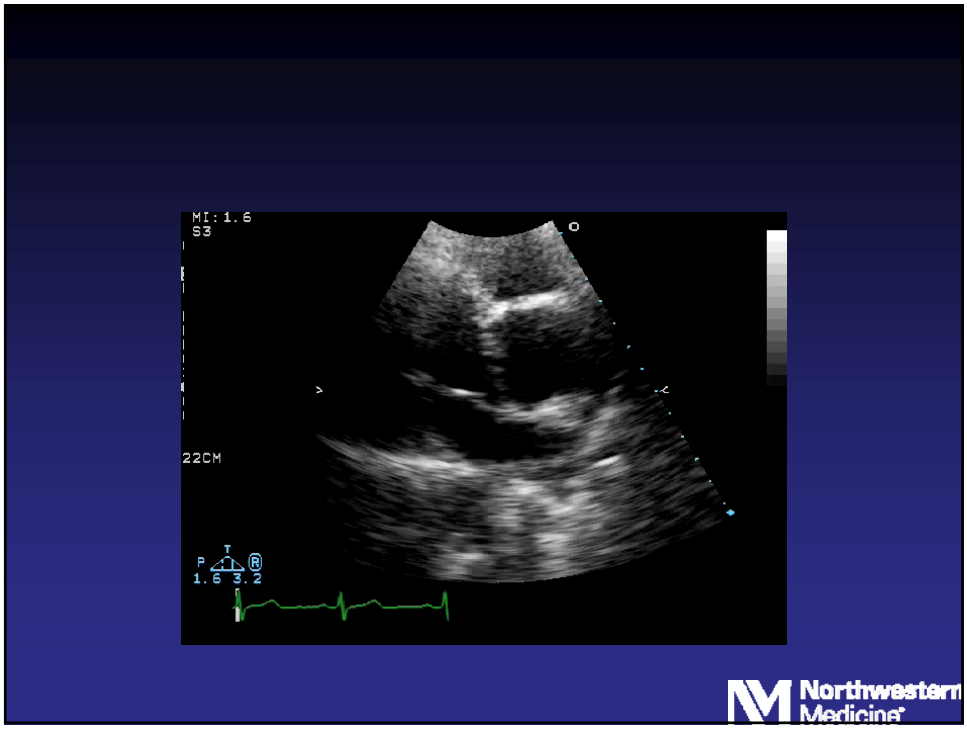
MI:1.6
S3
22CM
P T R
1.6 3.2
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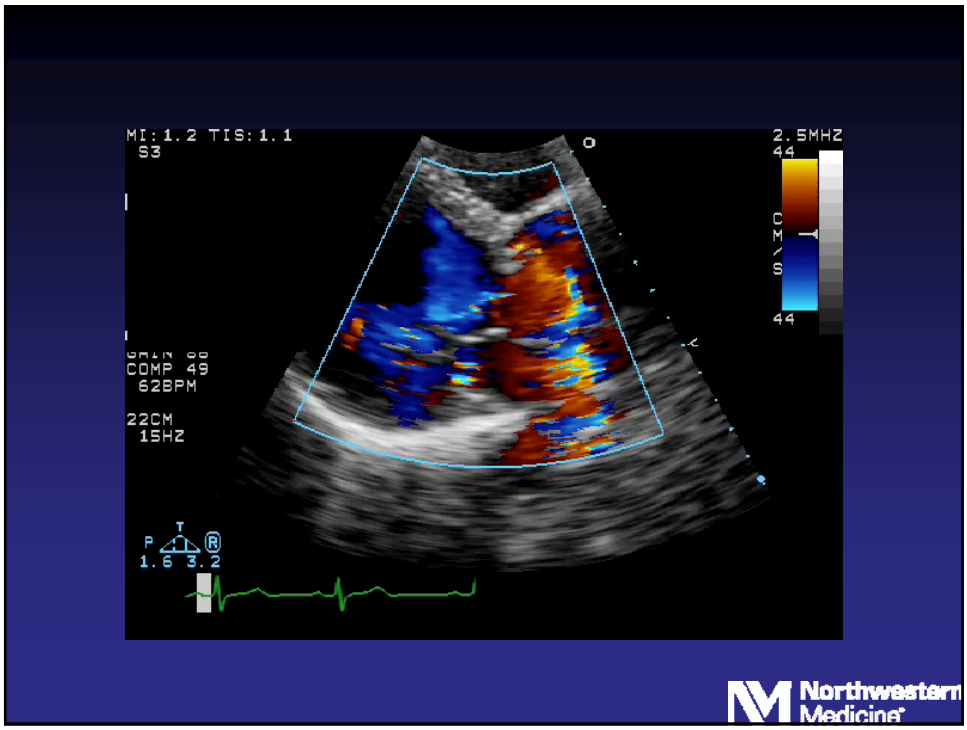
MI:1.2 TIS:1.1
S3
2.5MHZ
44
C
M
S
44
COMP 49
62BPM
22CM
15HZ
P T R
1.6 3.2
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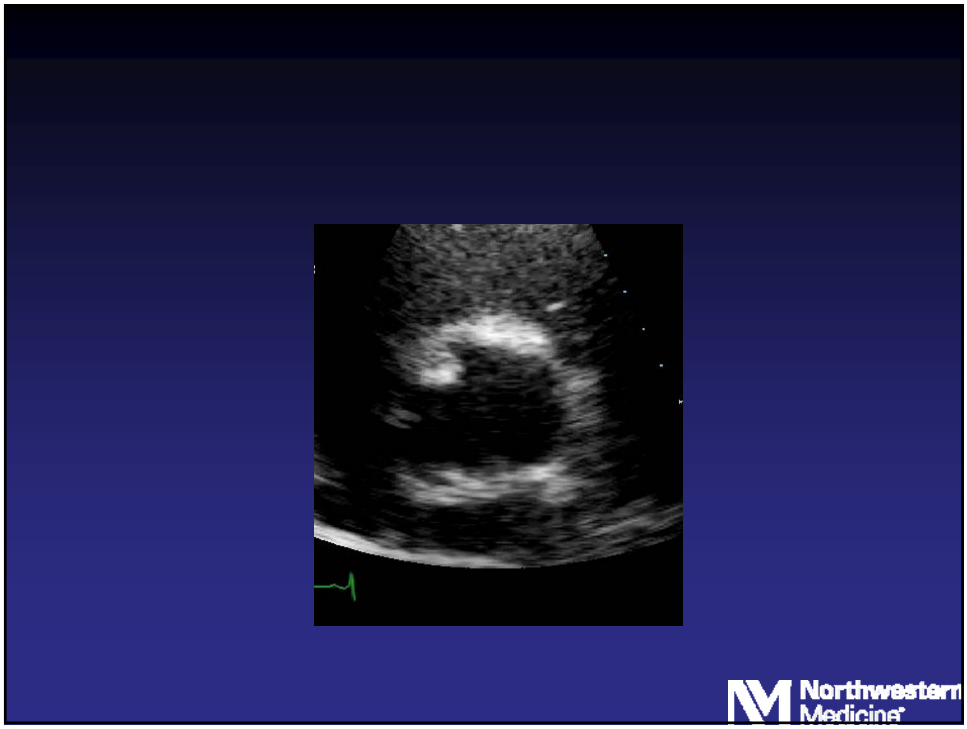
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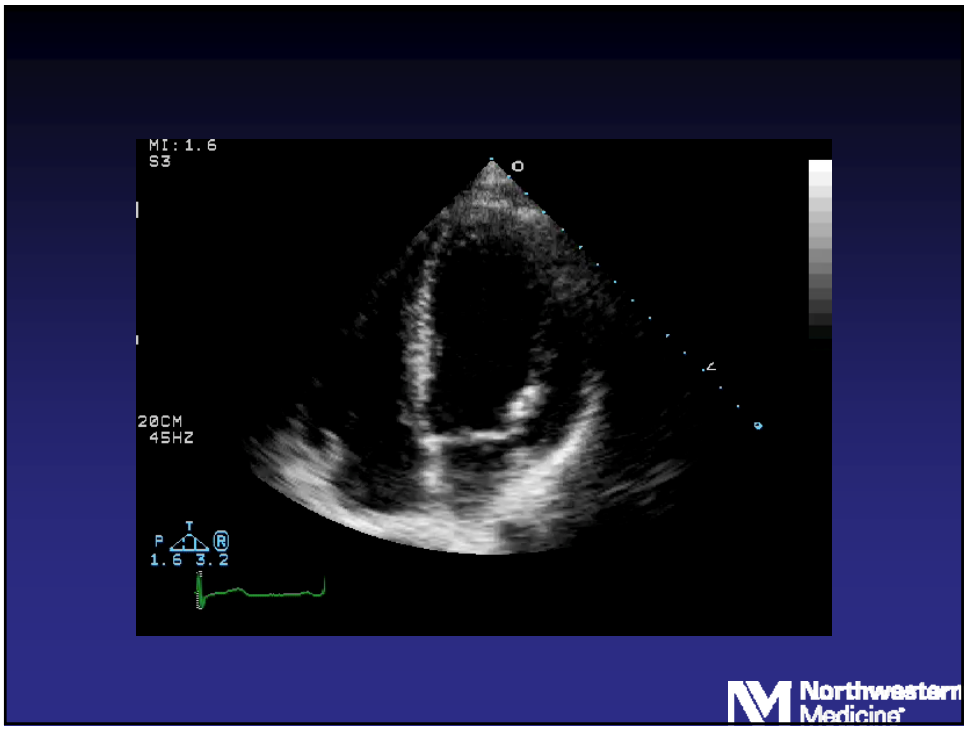
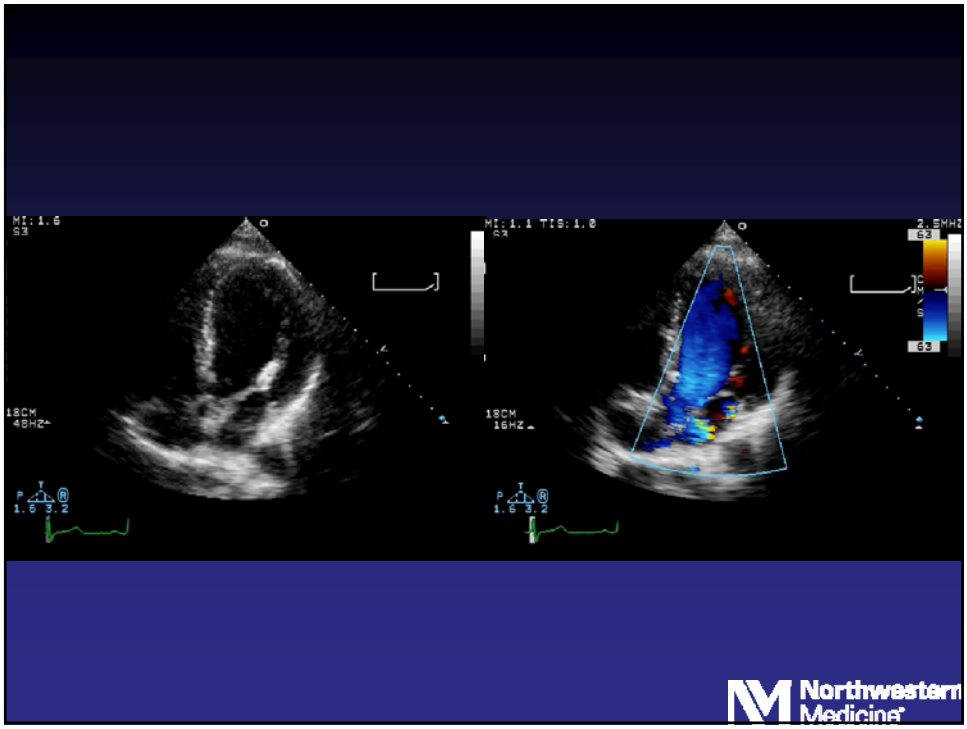
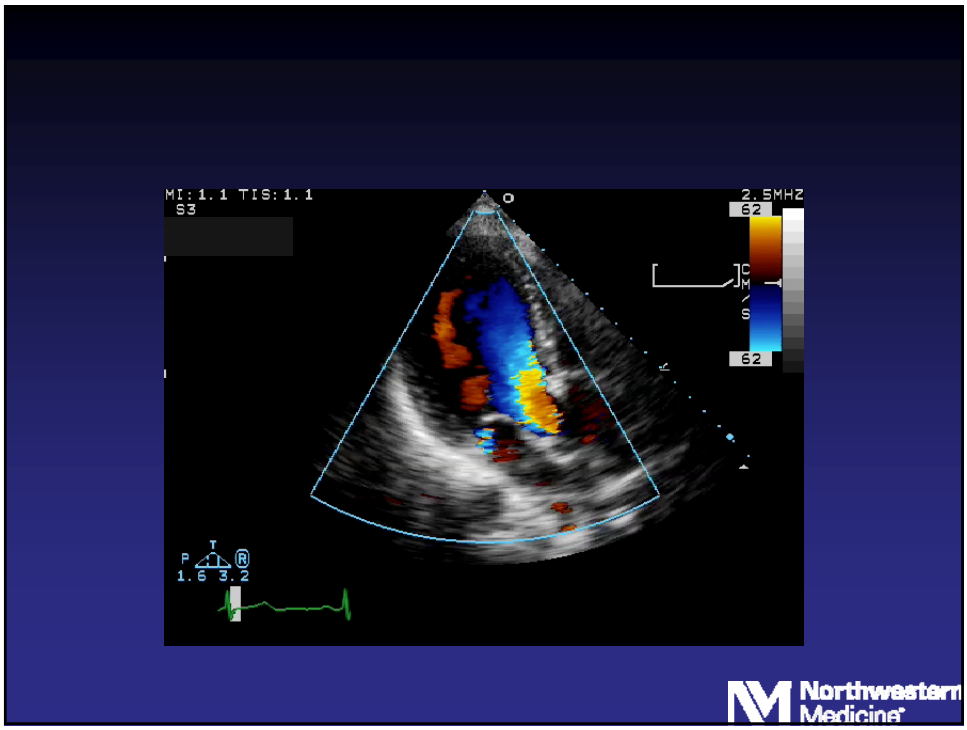
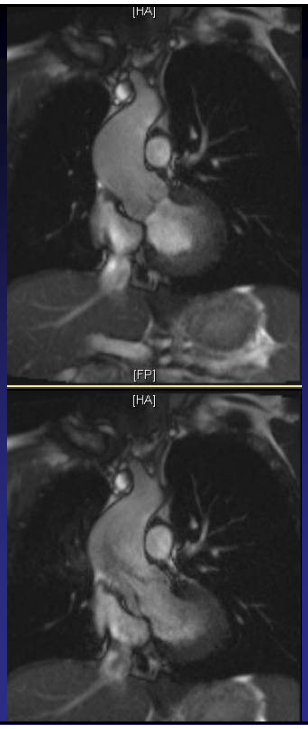

MI:1.6
S3
20CM
45HZ
P
1.6 3.2
Northwestern
Medicine


MI:1.6
S3
18CM
48HZ
MI:1.1 TIS:1.8
S3
2.5MHZ
63
63
18CM
16HZ
Northwestern Medicine


MI:1.1 TIS:1.1
S3
2.5MHZ
62
62
Northwestern Medicine



MRA of the Aorta

There is aneurysmal dilatation of the mid ascending aorta.

The following orthogonal measurements of the thoracic aorta were obtained:
Annulus: 3.2 by 3.3 cm
Sinus of Valsalva: 4.1 x 4.8 x 4.0 cm. The largest dimension is between the left coronary cusp and the non-coronary cusp.
Sino-tubular junction: 4.1 x 3.9 cm
Mid ascending aorta: 5.0 x 5.1 cm
Proximal aortic arch: 2.9 x 2.9 cm
Distal aortic arch: 2.3 x 2.5 cm
Lower descending thoracic aorta: 2.1 x 2.2 cm

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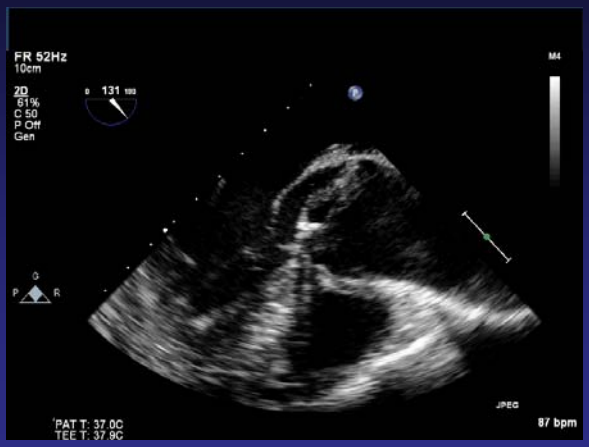

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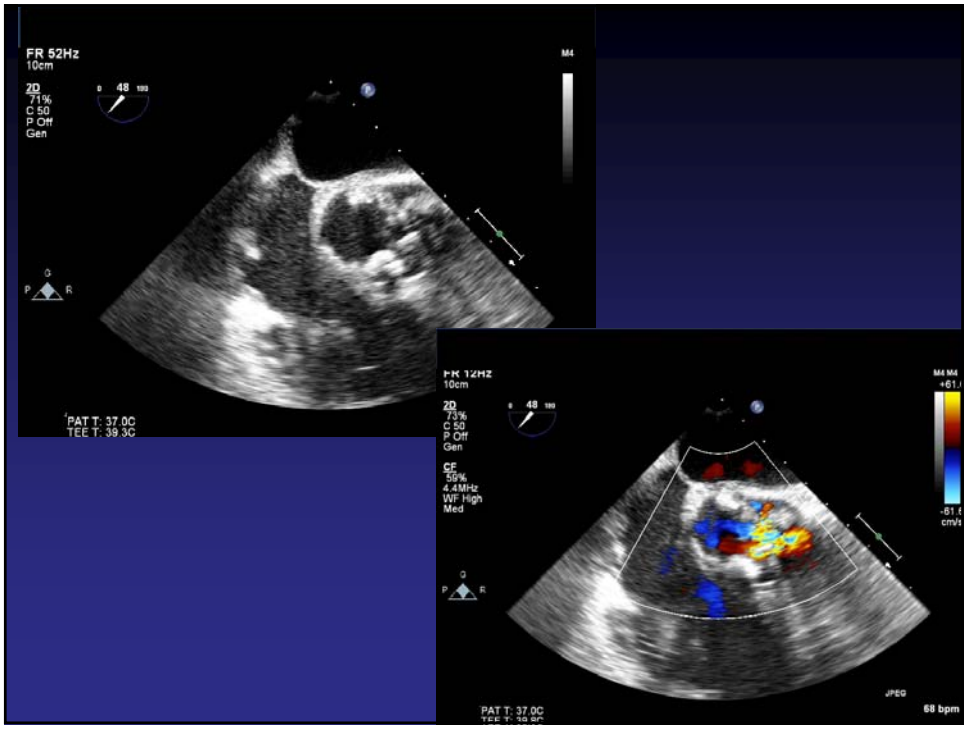
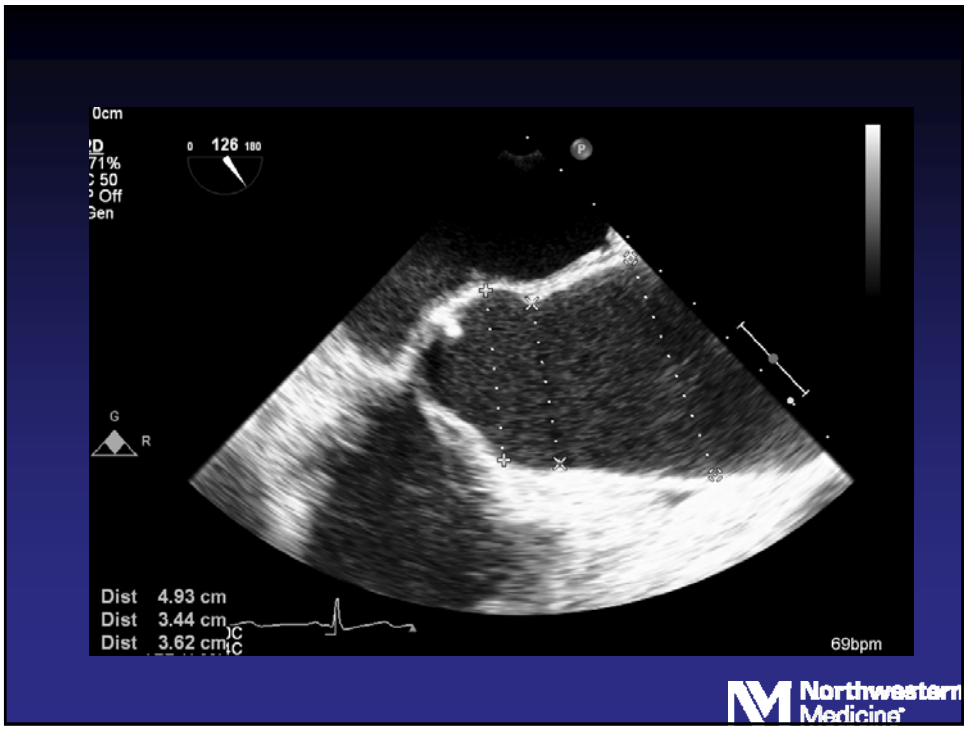

FR 52Hz
10cm
2D
71%
C 50
P Off
Gen
PAT T: 37.0C
TEE T: 39.3C
FR 12Hz
10cm
2D
73%
C 50
P Off
Gen
CF
59%
4.4MHz
WF High
Med
JPEG
68 bpm


0cm
126
180
Dist 4.93 cm
Dist 3.44 cm
Dist 3.62 cm
69bpm
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Thank You

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