Rheumatic Heart Disease

Katy Lonergan, MD
Associate Professor of Medicine
Medical Director, Echo Labs
UT Southwestern and Parkland Hospital
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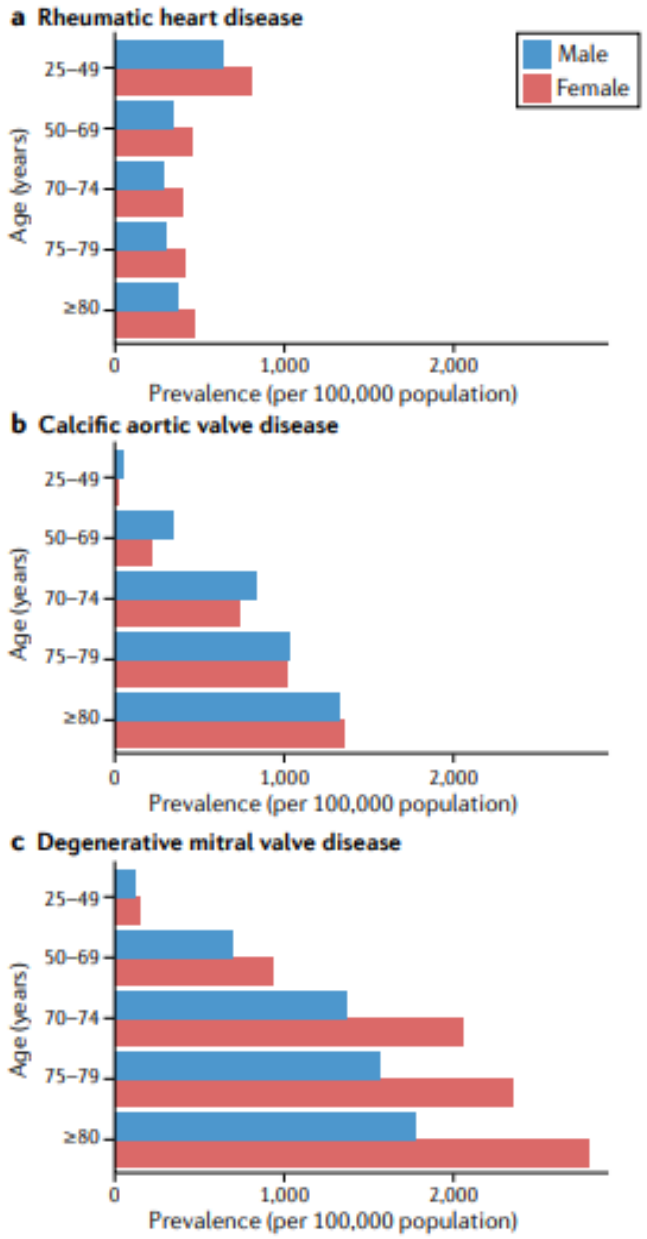
UT Southwestern Medical Center

Parkland

Aim to improve knowledge of:

- Rheumatic heart disease epidemiology and clinical features
- Cardiac valvular complications of rheumatic heart disease (RHD)
- Echo findings in RHD
- Management strategies for mitral stenosis



Coffey et al, *Nat Rev* 2021

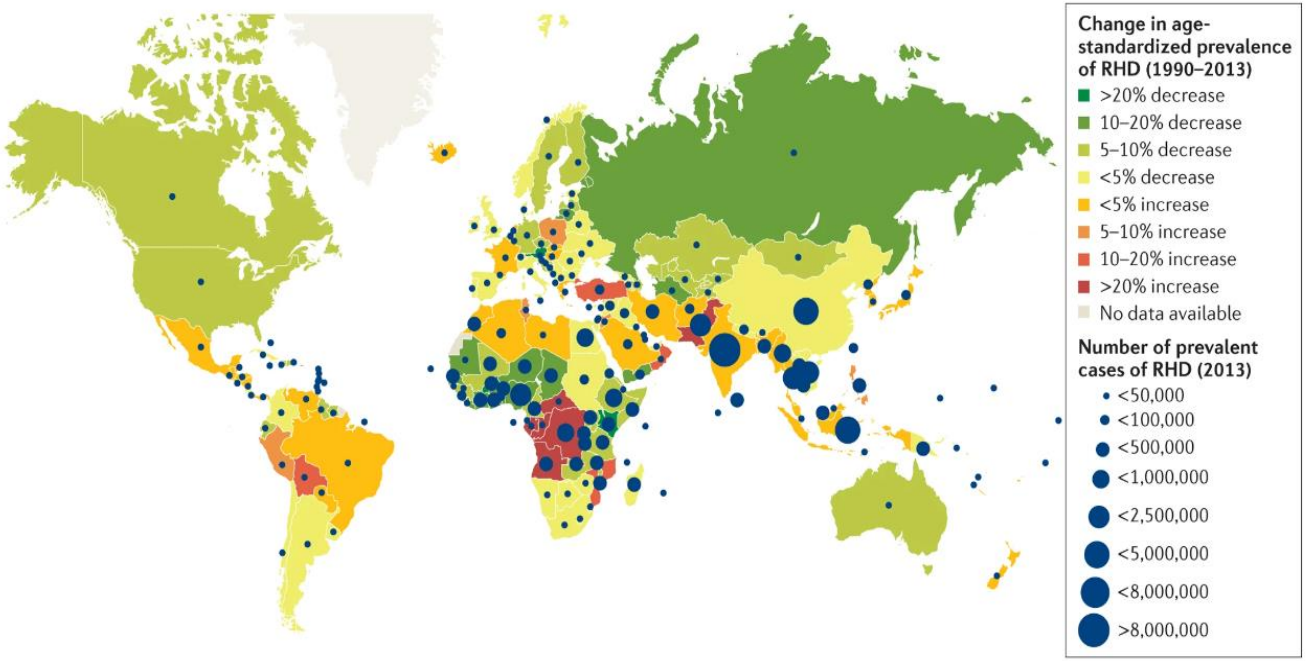
-Rheumatic heart disease remains common worldwide

-Unlike other etiologies which increase with age, RHD more common in younger patients, more women

-Global disparities: majority AS and MR in industrialized, higher income nations; majority RHD in nations with less health care resources

RHD Epidemiology

- Greater than 38 million cases and ~ 300,000 deaths globally in 2017
- Highest prevalence and mortality in Oceania, South Asia, and sub-Saharan Africa



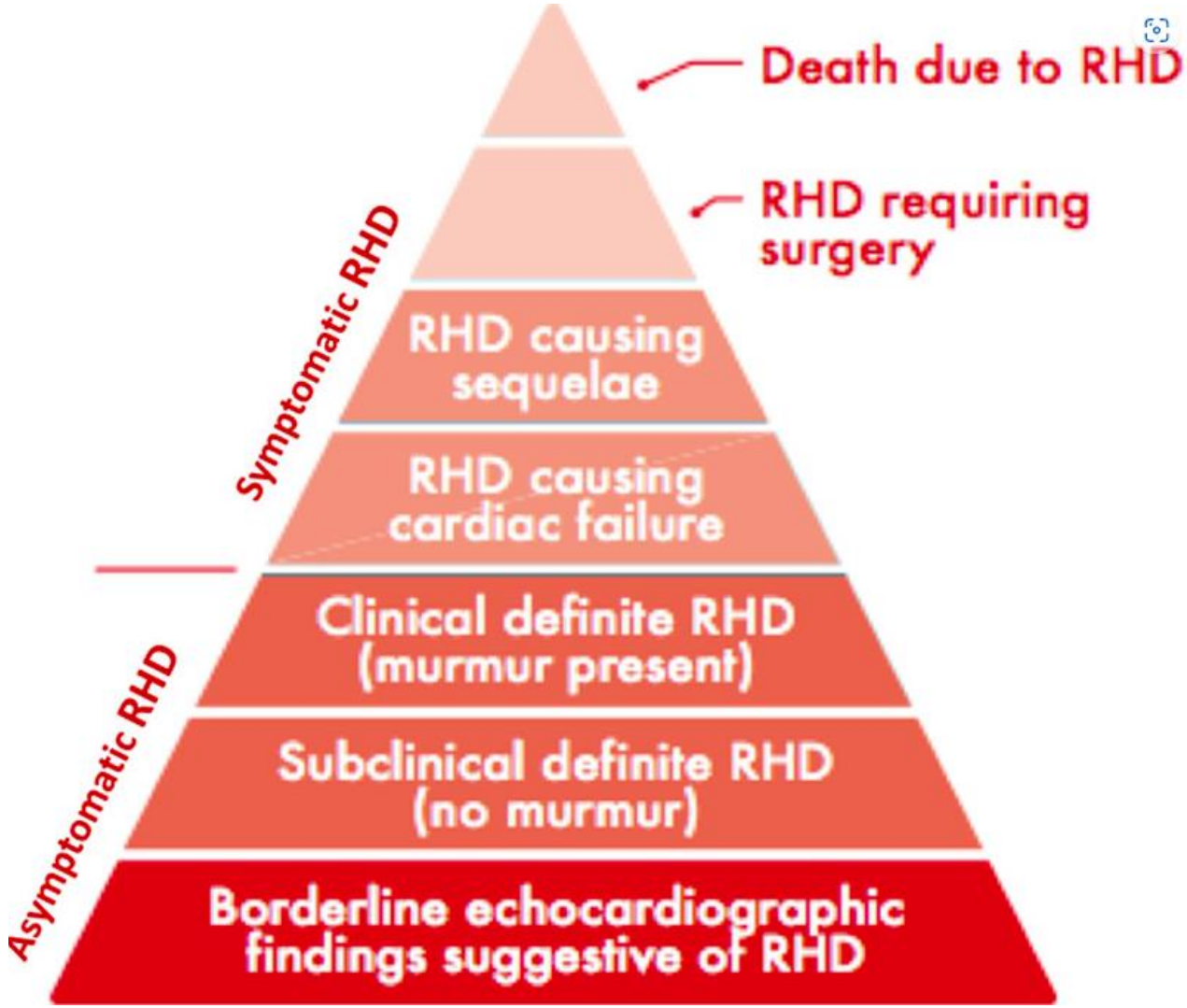
Kumar et al, *Circulation* 2020

Carapetis et al, *Nature Rev* 2016

RHD Epidemiology

- Sub-Saharan Africa: 0.5-3% school age children with RHD
- Zambia: study of school children screened with portable cardiac ultrasound resulted in RHD diagnosis 11.8 per 1000 children

Schwaninger et al et al, *BMC Cardiovasc Disord* 2018



Kumar et al, *Circulation* 2020

RHD Pathophysiology

Group A Strep (*Streptococcus pyogenes*) pharyngitis

↓

Acute Rheumatic Fever

↓

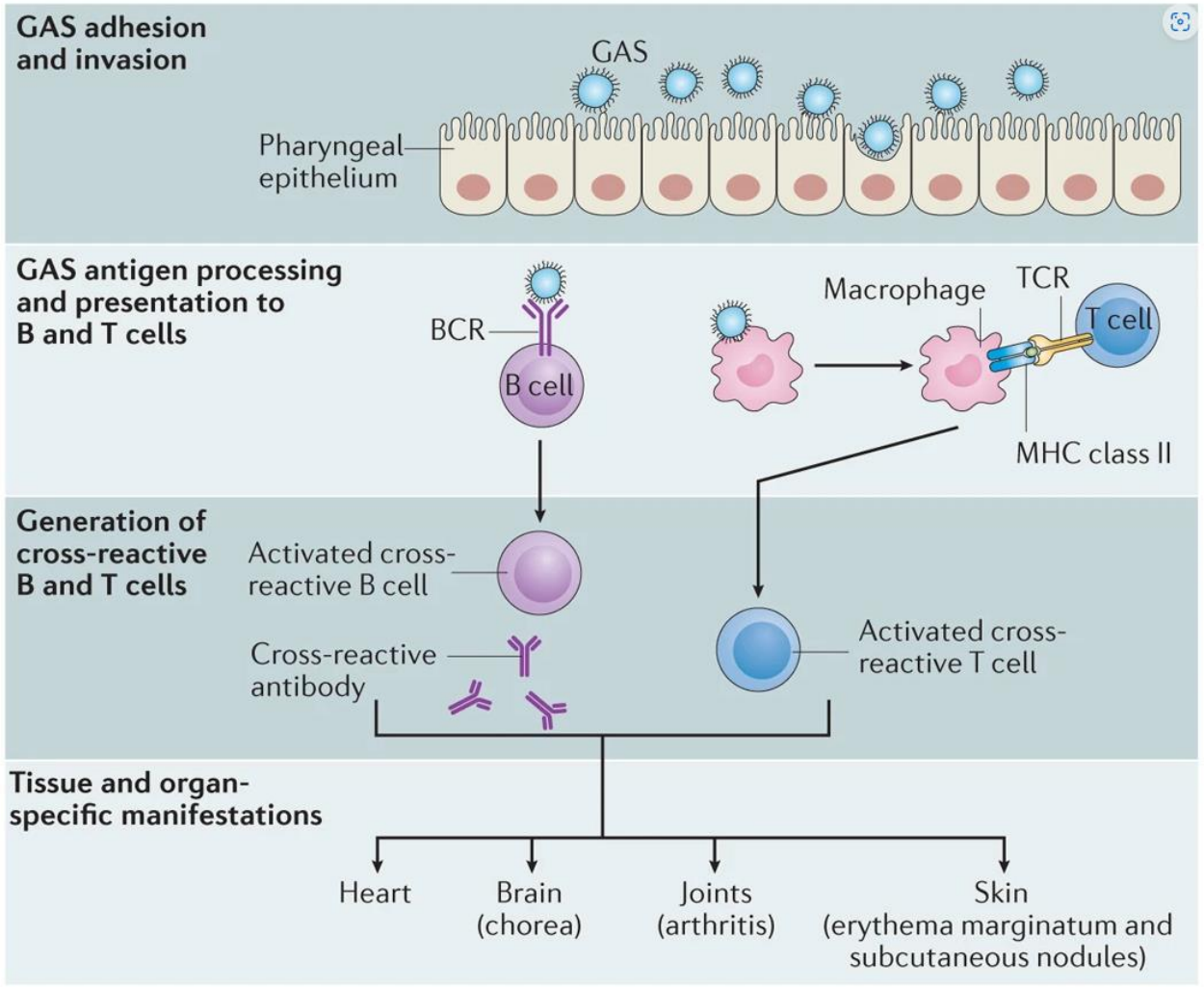
Immune/inflammatory Response

↓

Recurrent episodes

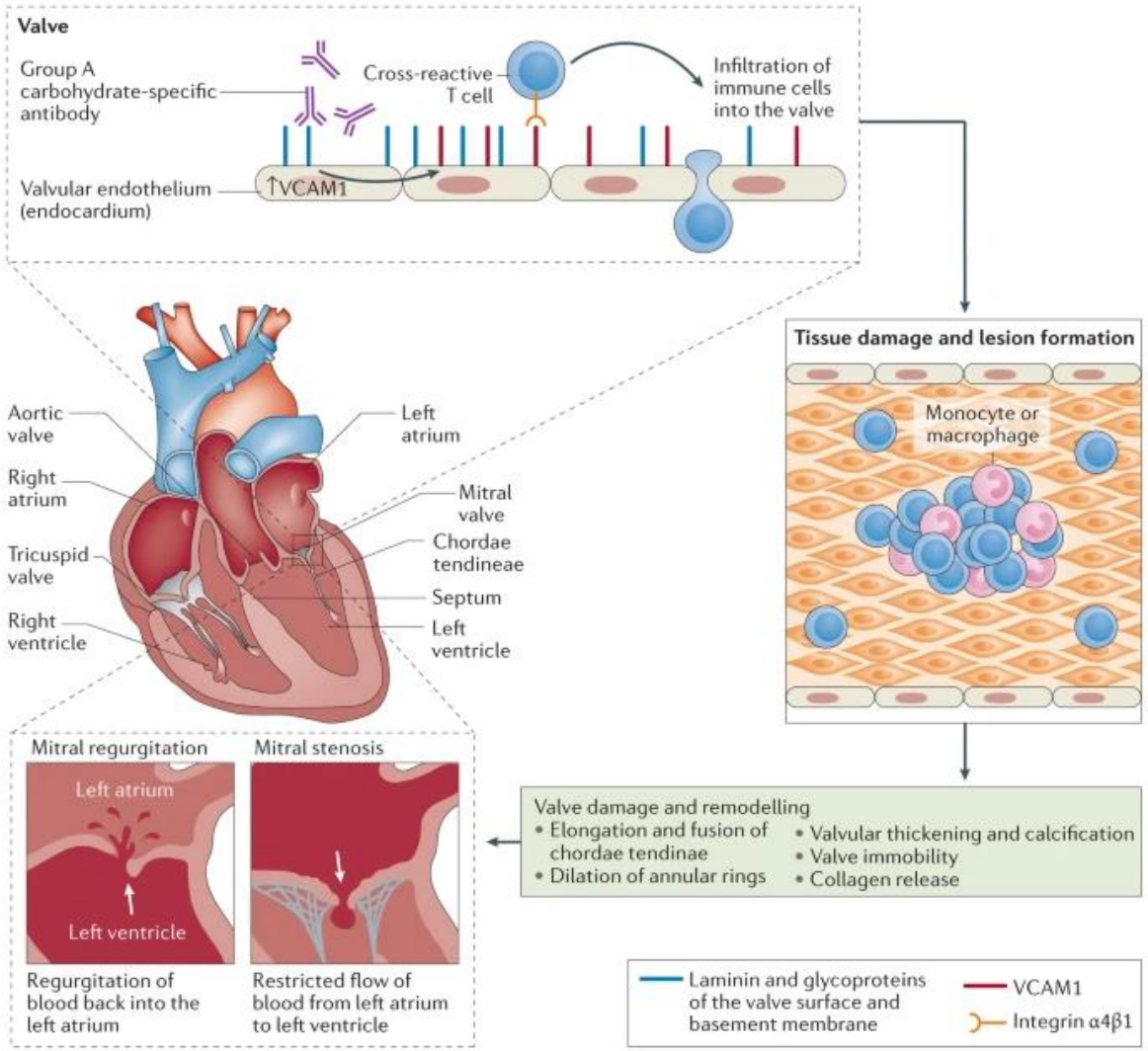
↓

Worsening inflammatory repose and progressive valve damage

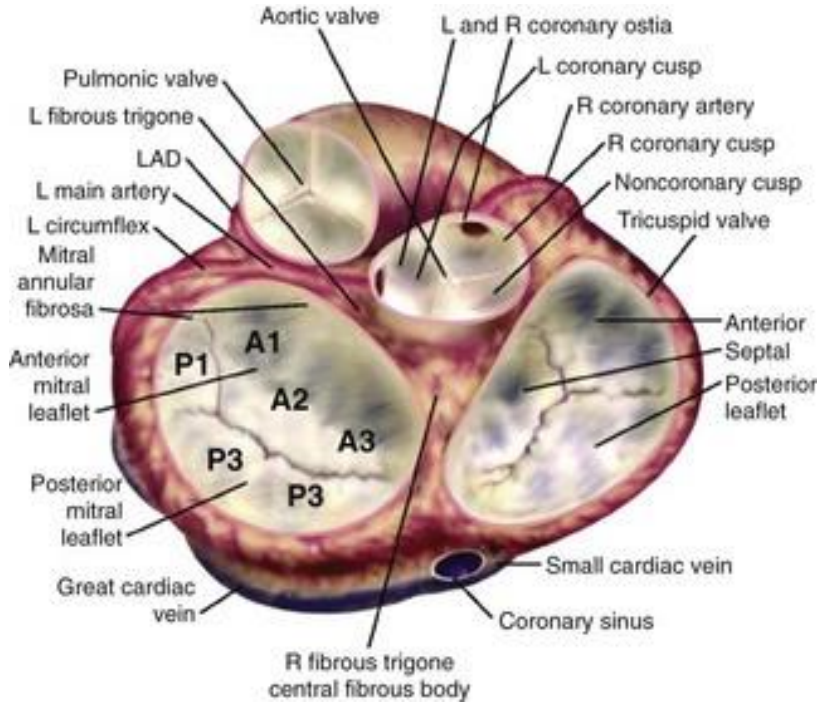


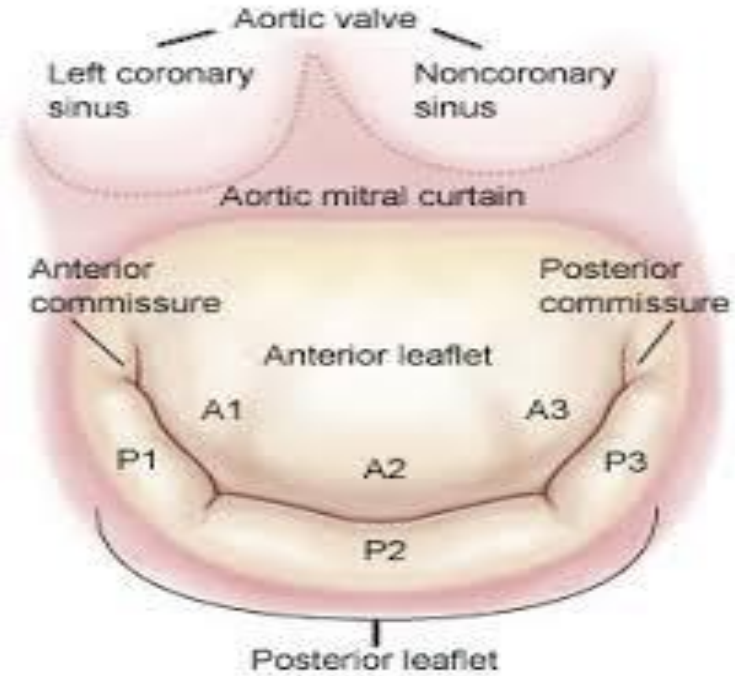
Carapetis et al, *Nature Rev* 2016

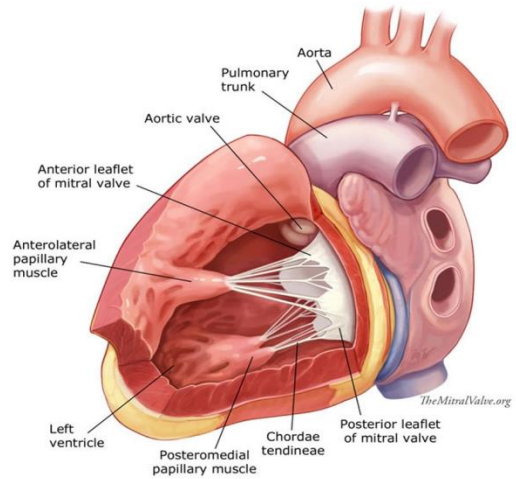
RHD Pathophysiology

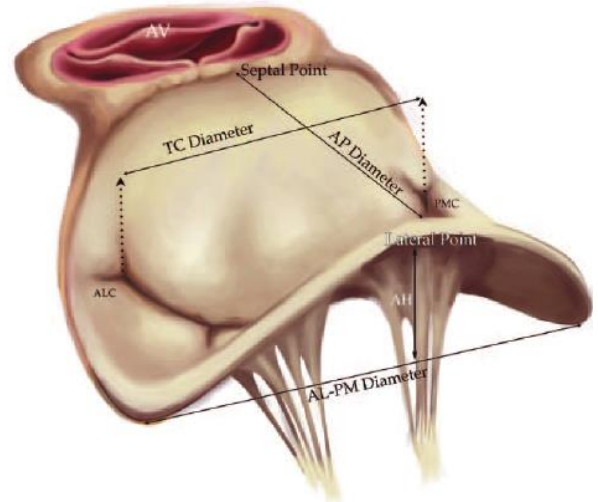


Carapetis et al, *Nature Rev* 2016

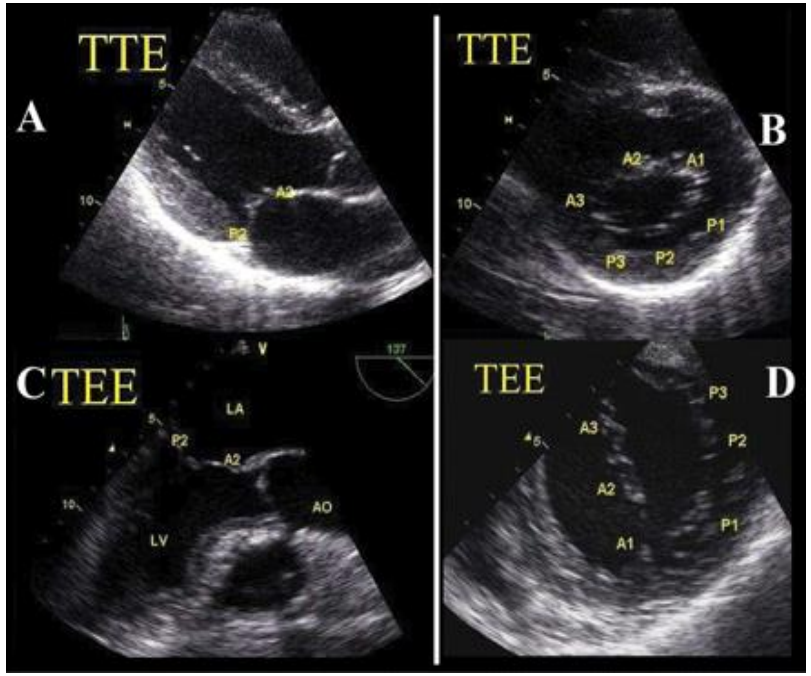

Aortic valve
L and R coronary ostia
L coronary cusp
Pulmonic valve
L fibrous trigone
R coronary artery
R coronary cusp
LAD
L main artery
Noncoronary cusp
L circumflex
Tricuspid valve
Mitral annular fibrosa
Anterior Septal
Posterior leaflet
A1
P1
Anterior mitral leaflet
A2
A3
P3
P3
Posterior mitral leaflet
Small cardiac vein
Great cardiac vein
Coronary sinus
R fibrous trigone
central fibrous body

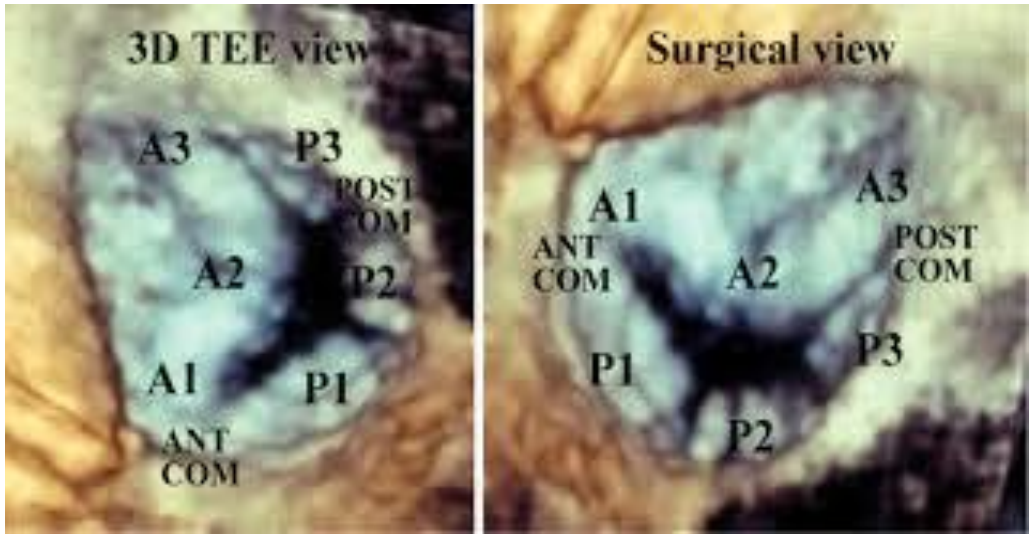

Aortic valve
Left coronary sinus
Noncoronary sinus
Aortic mitral curtain
Anterior commissure
Posterior commissure
Anterior leaflet
A1
A3
P1
A2
P3
P2
Posterior leaflet


Aorta
Pulmonary trunk
Aortic valve
Anterior leaflet of mitral valve
Anterolateral papillary muscle
TheMitralValve.org
Left ventricle
Posteromedial papillary muscle
Chordae tendineae
Posterior leaflet of mitral valve


AV
Septal Point
TC Diameter
AP Diameter
PMC
Lateral Point
ALC
AL
AL-PM Diameter

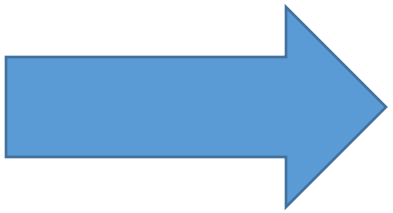
Mitral Valve Imaging





RHD Pathophysiology

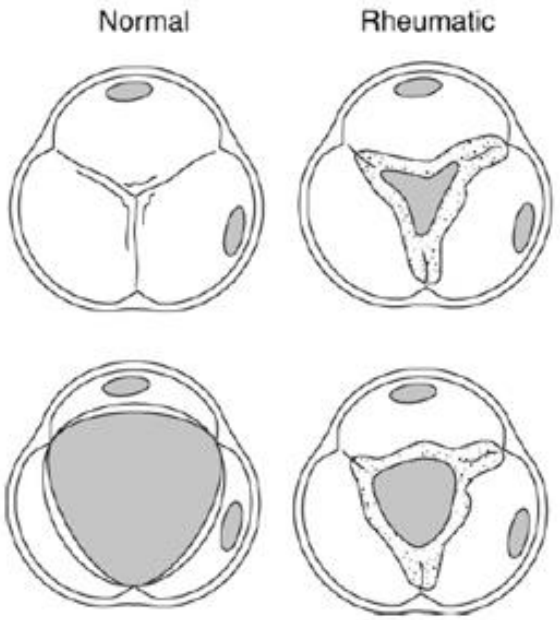
- Childhood:
  - Acute mitral valvulitis
  - Annular dilatation
  - Elongation of the chordae
  - Tethering of posterior leaflet
  - Relative prolapse of anterior leaflet
  - Predominantly mitral regurgitation

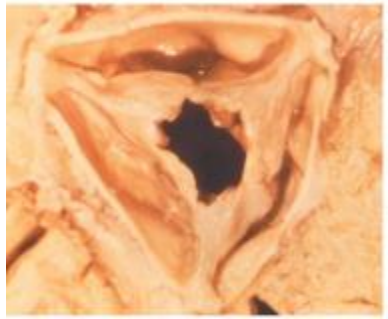


- Adulthood:
  - Mitral leaflet and chordal thickening/fibrosis
  - Commissural fusion
  - Leaflet doming, restricted leaflet opening, reduced mitral valve orifice area
  - +/- associated mitral regurgitation
  - +/- aortic valve commissural fusion-> aortic regurgitation

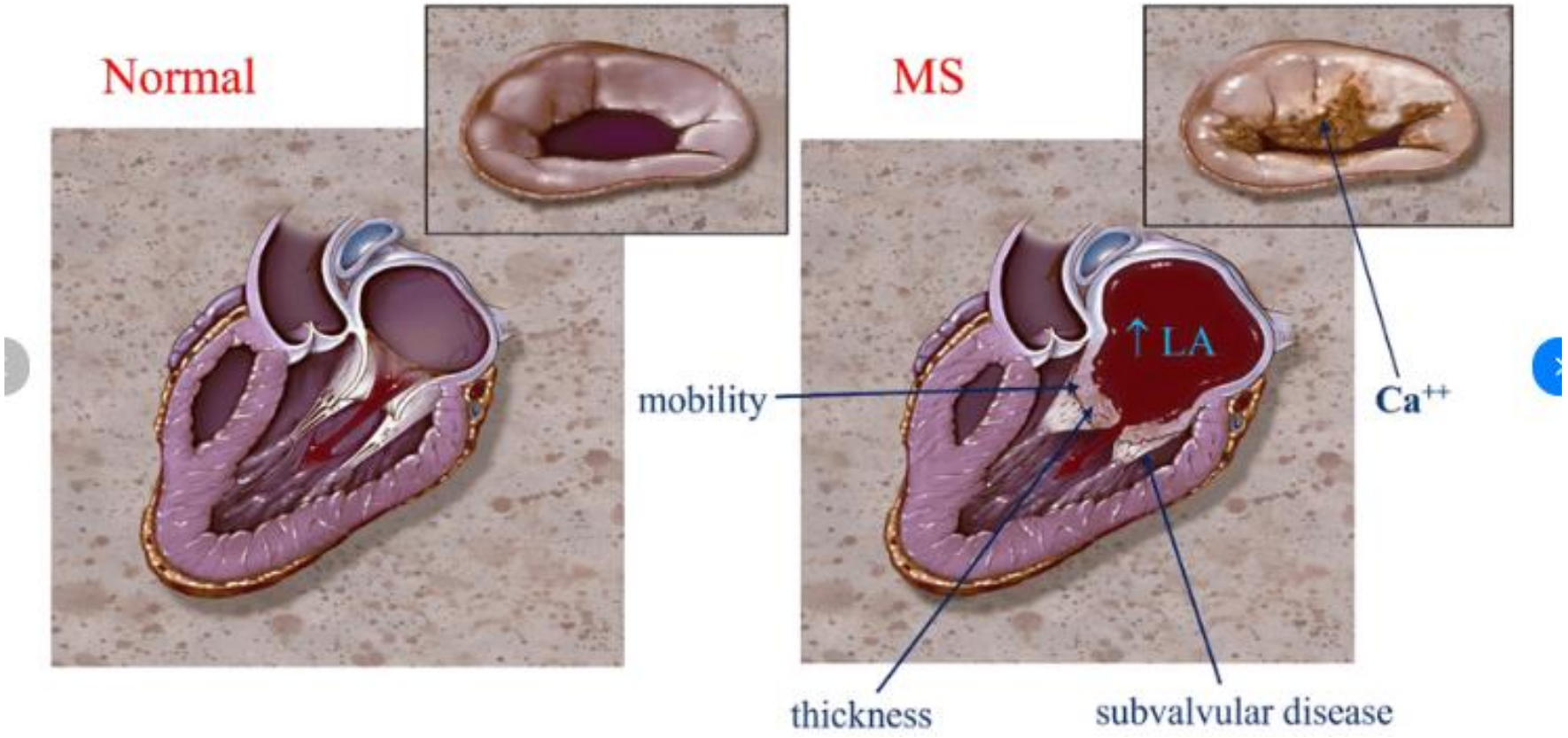
Auala et al, *Pathogens* 2022

RHD- Aortic Valve (more rare than mitral)





RHD – Mitral stenosis

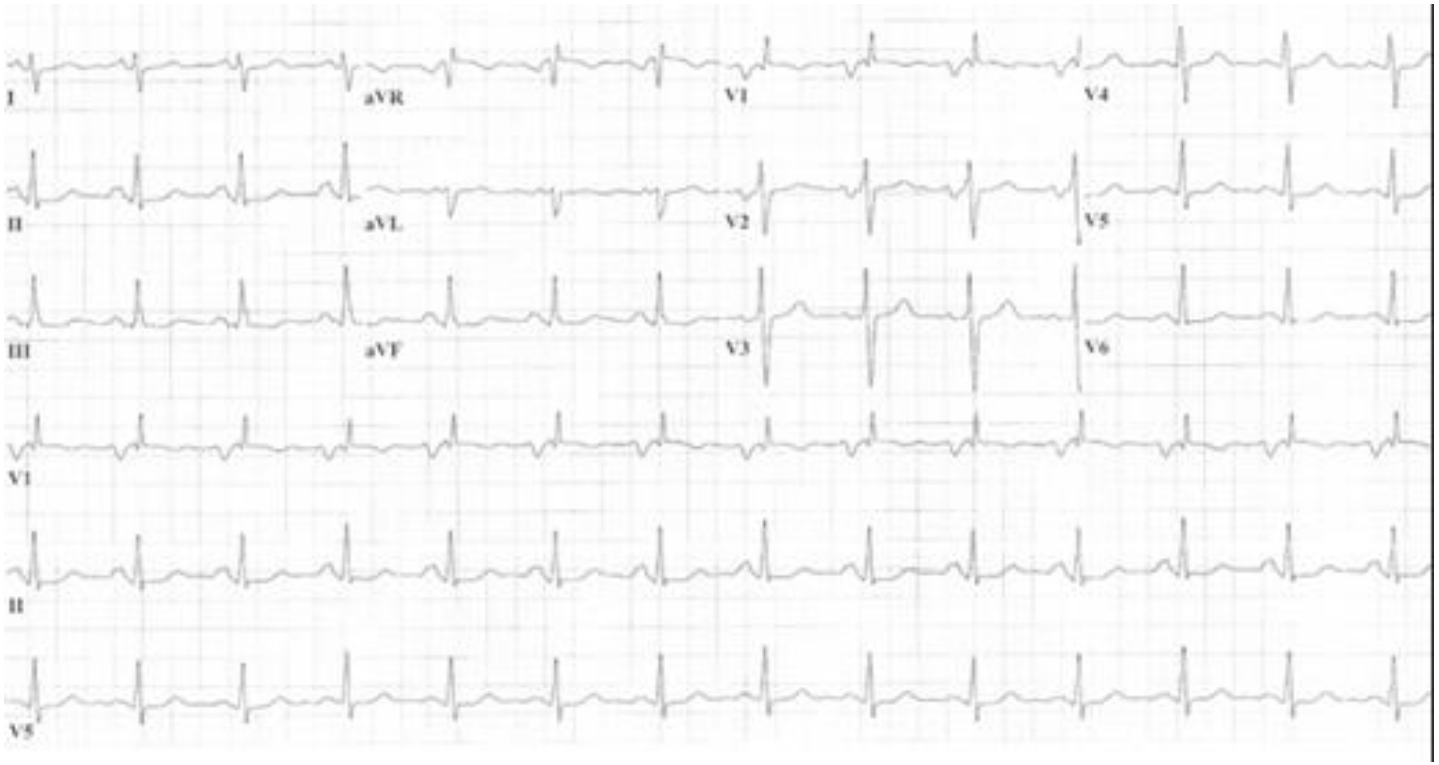


Zuri, *Circulation* 2004

RHD - Mitral Stenosis

- Increased left atrial pressure, increased pulmonary capillary wedge pressure
- Clinical features:
  - Slowly progressive (decades)
  - Dyspnea, increased with exertion, ⇧HR/CO
  - Atrial fibrillation -> embolic events, stroke
  - Pulmonary hypertension->Right sided heart failure
  - Tricuspid regurgitation secondary to PH
  - Advanced: pulmonary edema, blood-tinged/hemoptysis

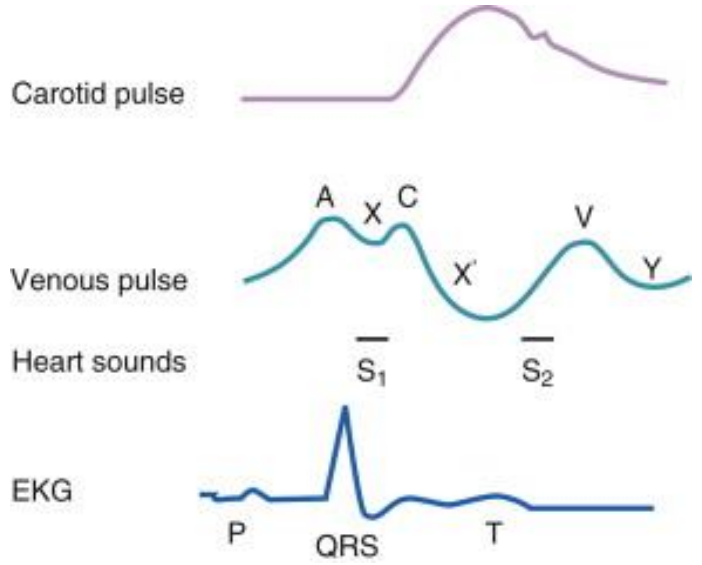
RHD - Mitral Stenosis

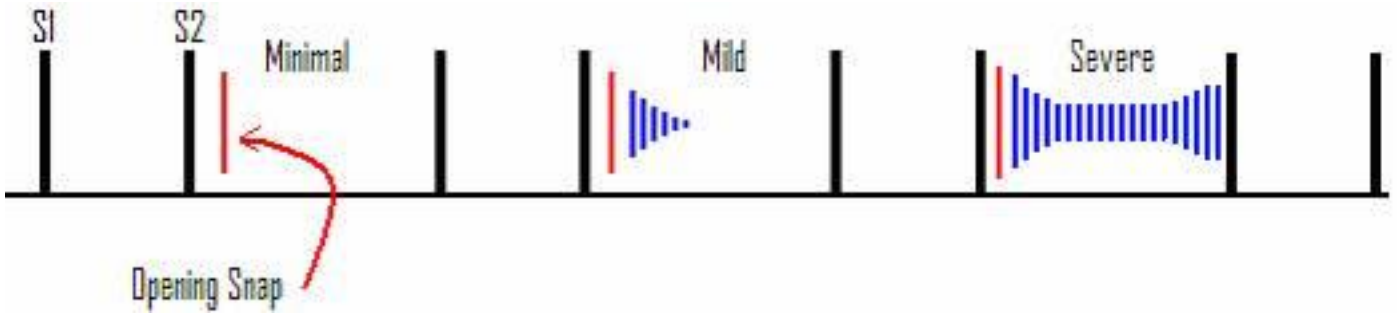


Source: J.E. Tintinalli, J.S. Stapczynski, O.J. Ma, D.M. Yealy, G.D. Meckler, D.M. Cline: Tintinalli's Emergency Medicine: A Comprehensive Study Guide, 8th Edition
www.accessmedicine.com

RHD- Mitral Stenosis

- Physical Exam:
  - JVD: Prominent “a” wave (if in NSR), no “a” wave if AF
  - Low pitched diastolic rumble
  - Loud S1 (stiff leaflets close)
  - P2 louder when PH present
  - Opening snap after S2 : this interval becomes shortened as MS worsens : less than 70 msec = severe MS

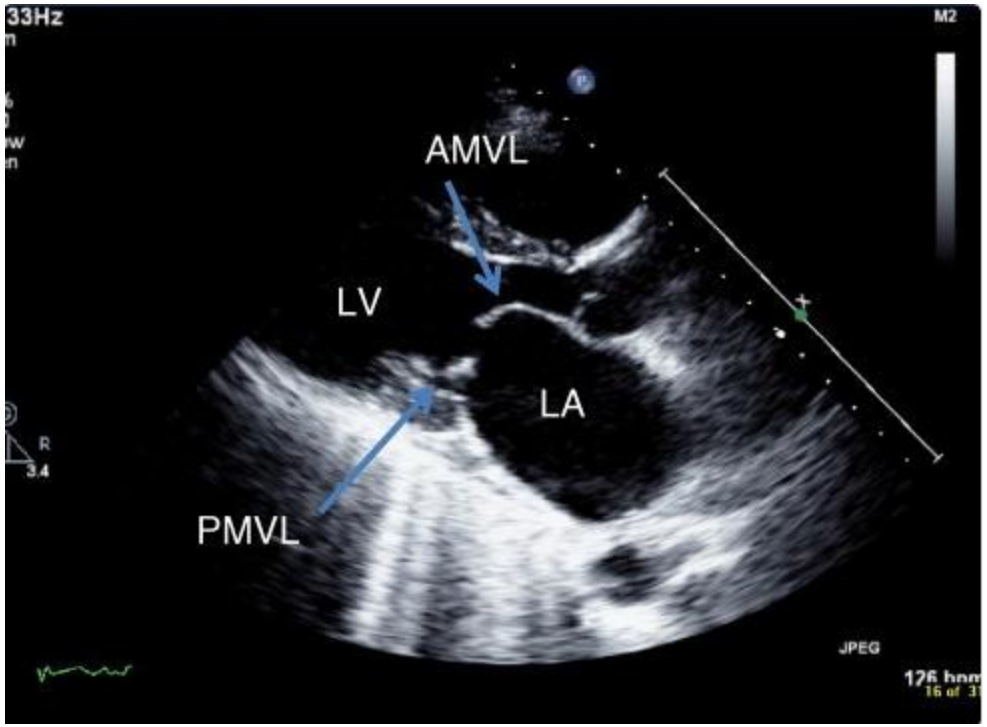


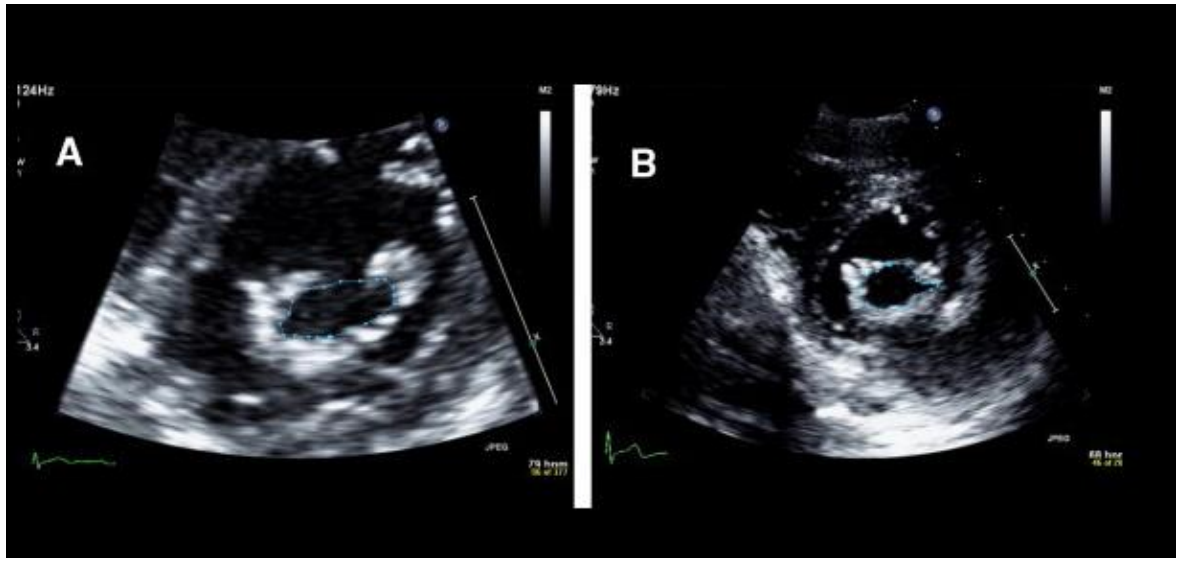


RHD - Mitral Stenosis

- Echo findings:
  - Thickened leaflets, doming, commissural fusion
  - Reduced mitral valve area on planimetry/Doppler
  - Increased transmitral gradient (at HR = x)
  - Dilated LA; normal LV size
  - Pulmonary HTN – increased RVSP, RA, RVH

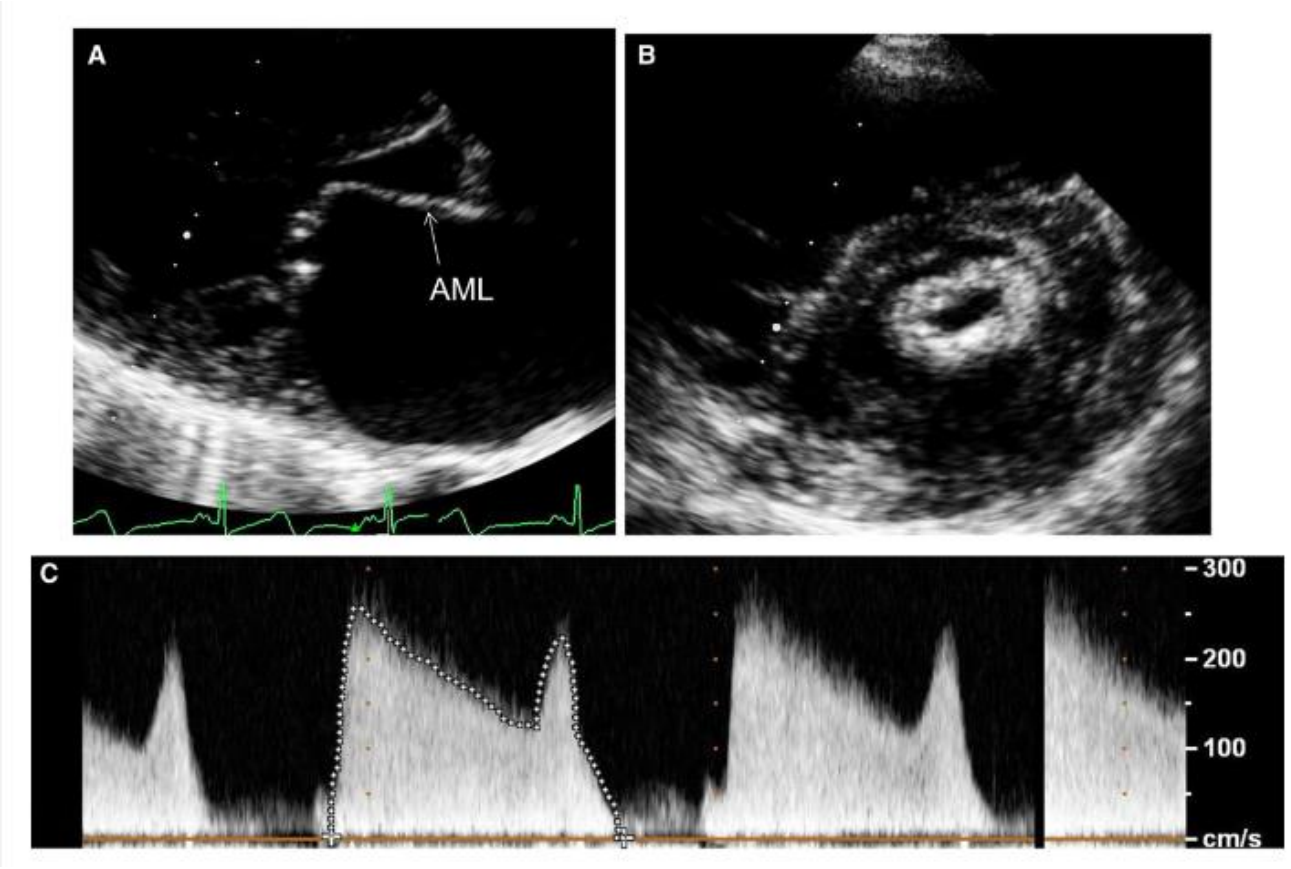
RHD – Mitral stenosis





Omran et al, *J Saudi Hear Assoc* 2011

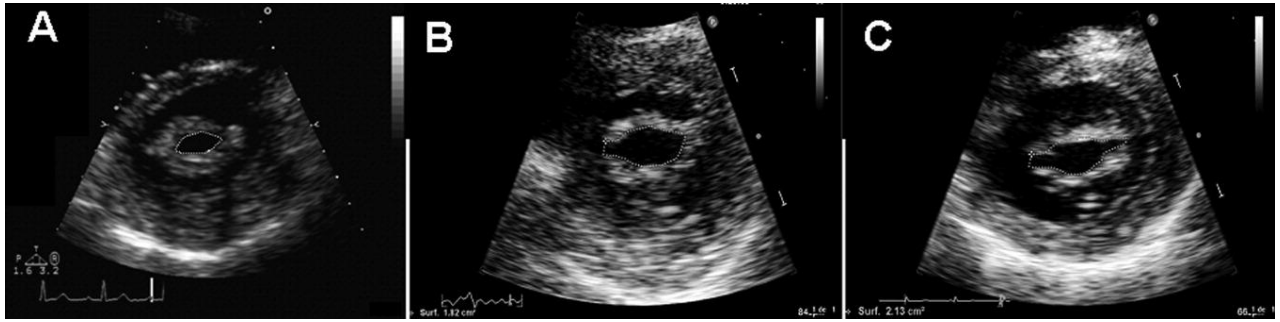
RHD – Mitral stenosis



Kumar et al, *Circulation* 2020

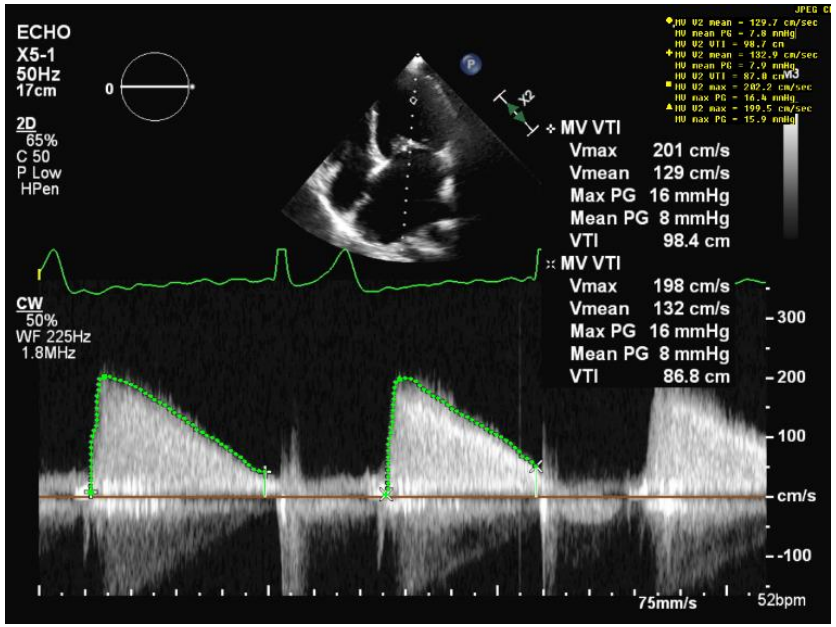
Echo Assessment of mitral stenosis

- Diastolic color flow acceleration
- Mean gradient – trace VTI of CW mitral inflow
- Always report HR; Average several cardiac cycles if afib
- MVA by pressure half time or planimetry
  - MVA=220 / PHT

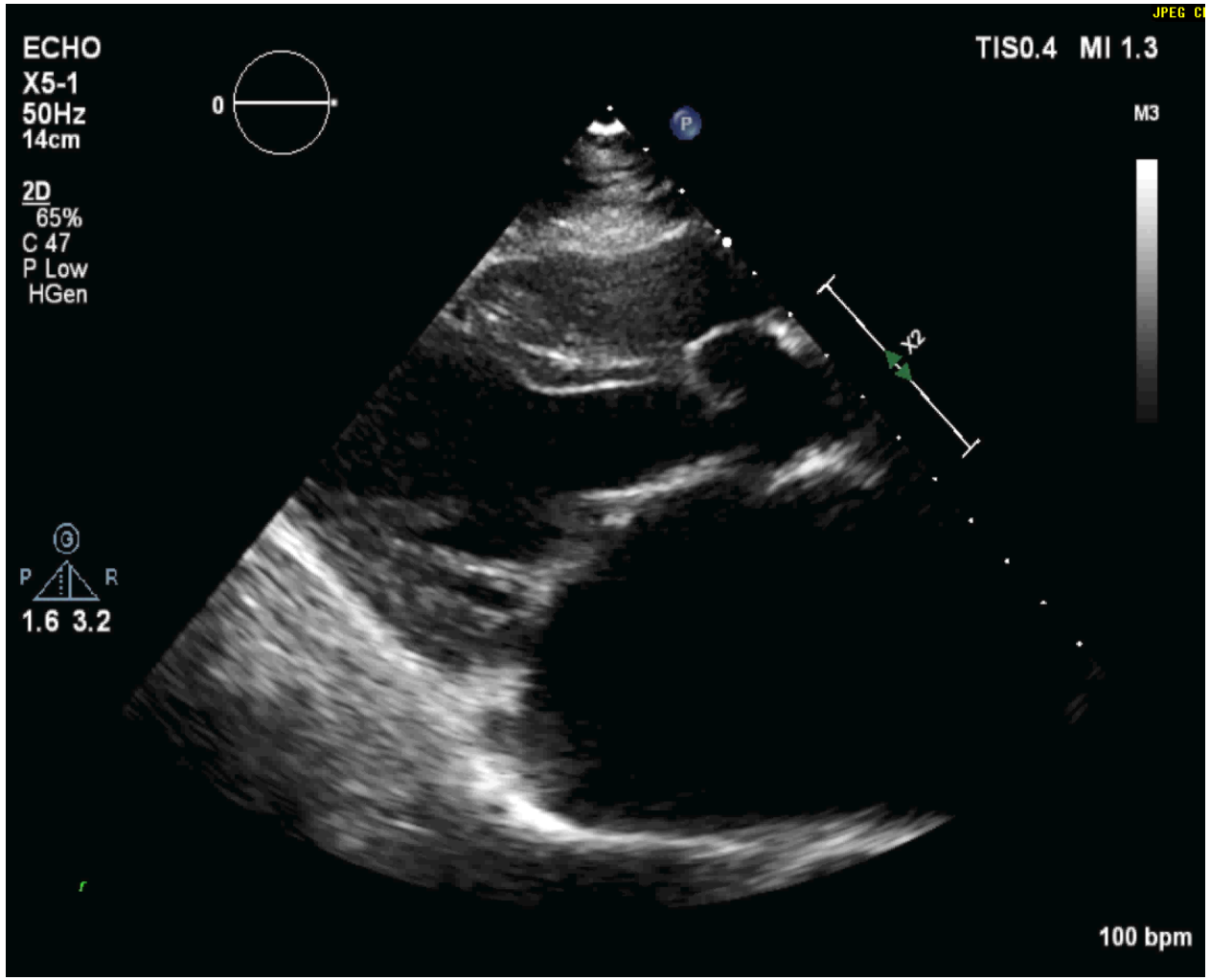


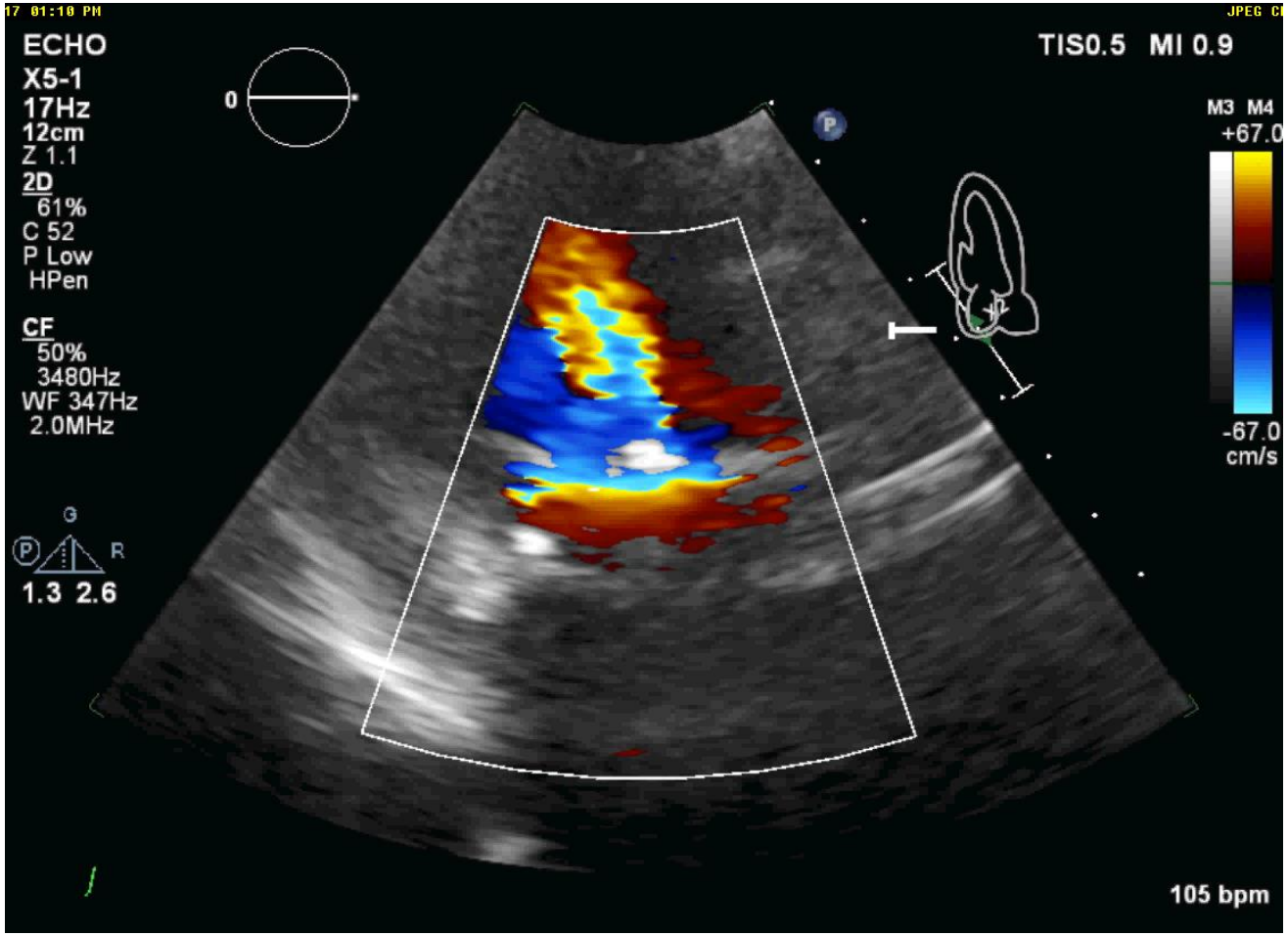
1.17 cm2 1.82cm2 2.13cm2

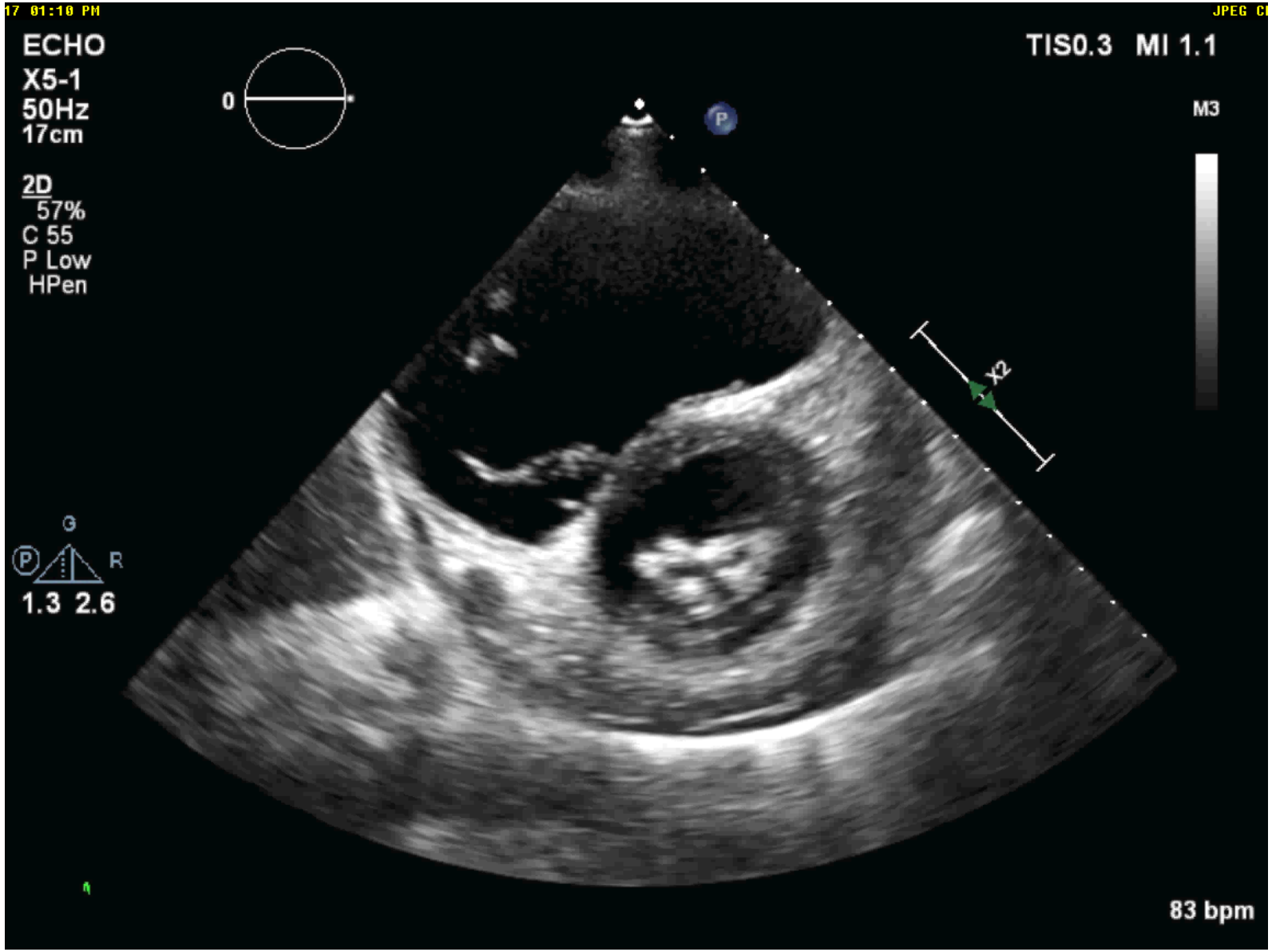
(Normal MVA > 4 cm2)



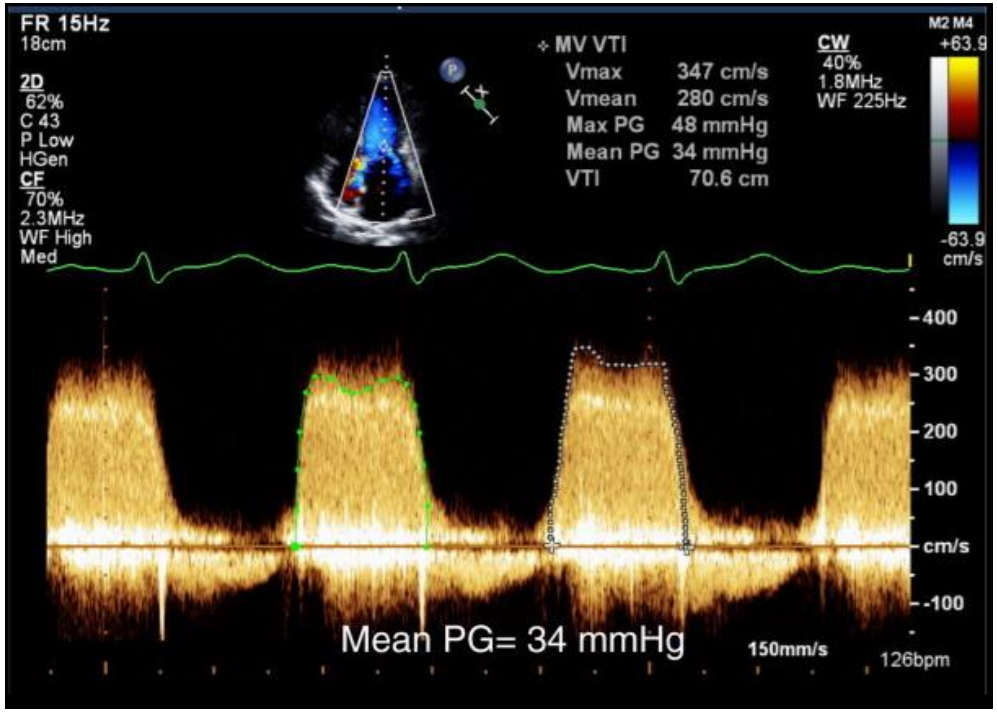
Baumgartner et al, *ASE* 2009

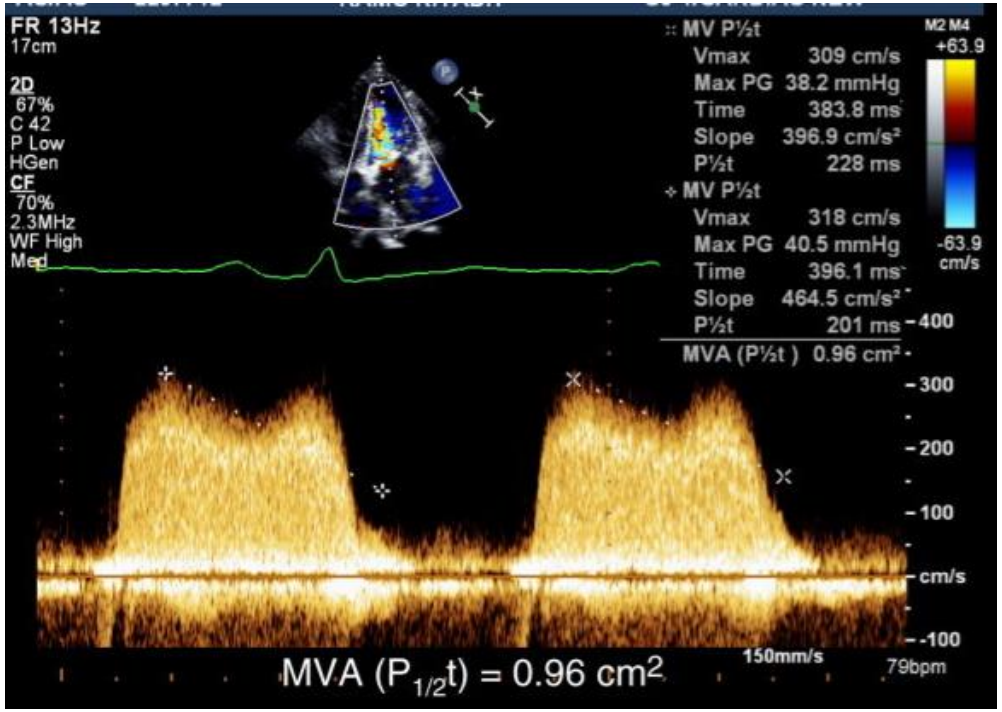

JPEG C
ECHO
X5-1
50Hz
14cm
2D
65%
C 47
P Low
HGen
TIS0.4 MI 1.3
M3
0
P
X2
P
R
1.6 3.2
100 bpm


17 01:10 PM
JPEG C
ECHO
X5-1
17Hz
12cm
Z 1.1
2D
61%
C 52
P Low
HPen
CF
50%
3480Hz
WF 347Hz
2.0MHz
G
P
R
1.3 2.6
0
TIS0.5 MI 0.9
M3 M4
+67.0
-67.0
cm/s
105 bpm

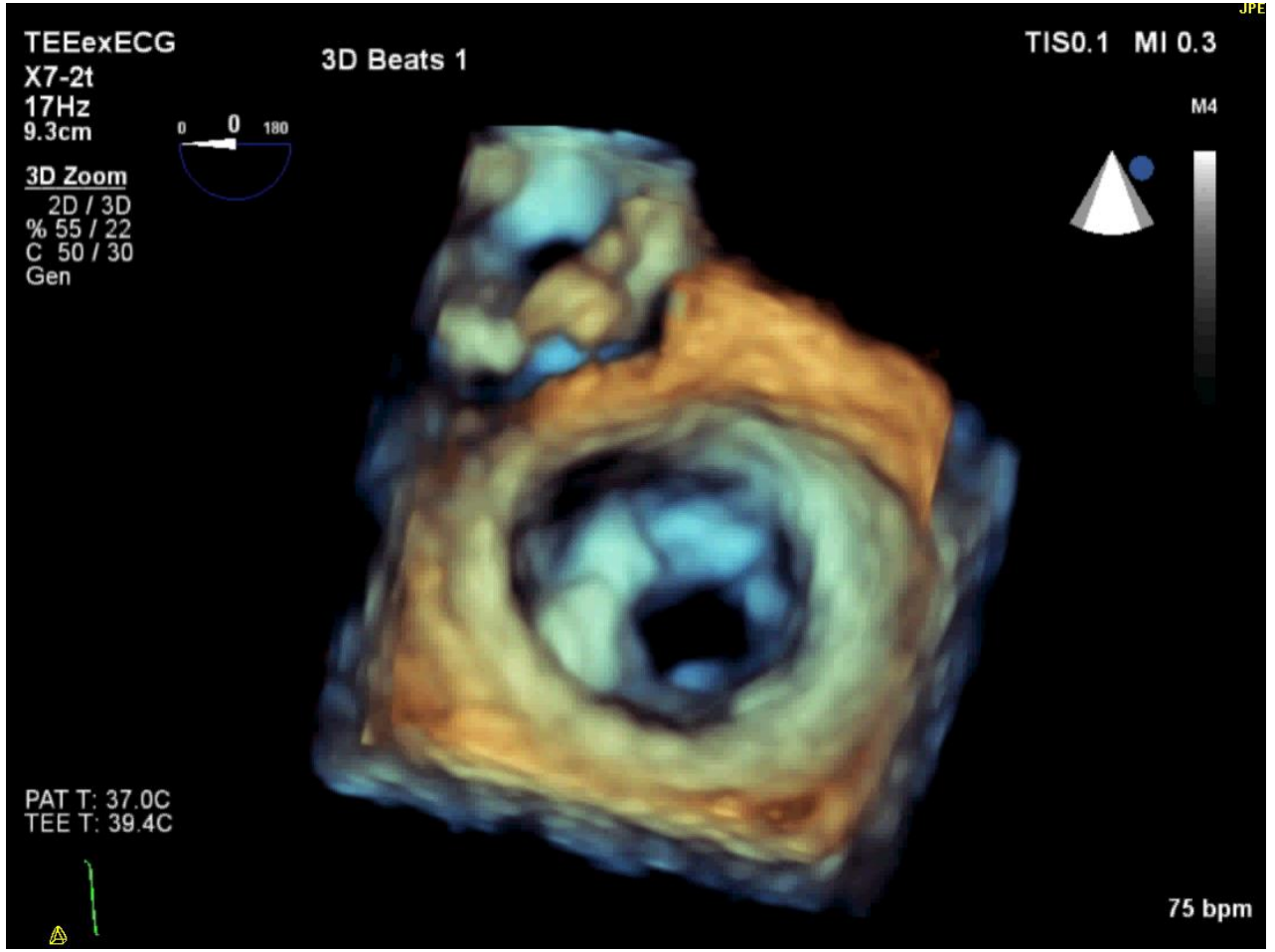

17 01:10 PM
JPEG C
ECHO
X5-1
50Hz
17cm
2D
57%
C 55
P Low
HPen
0
TIS0.3 MI 1.1
M3
X2
G
P
R
1.3 2.6
83 bpm

RHD – Mitral stenosis



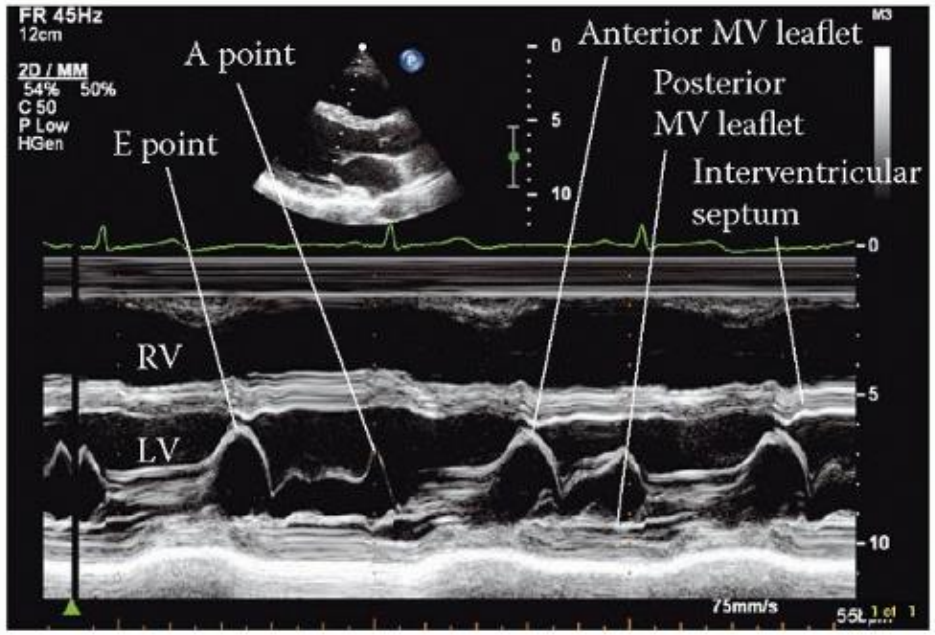


Omran et al, *J Saudi Hear Assoc* 2011

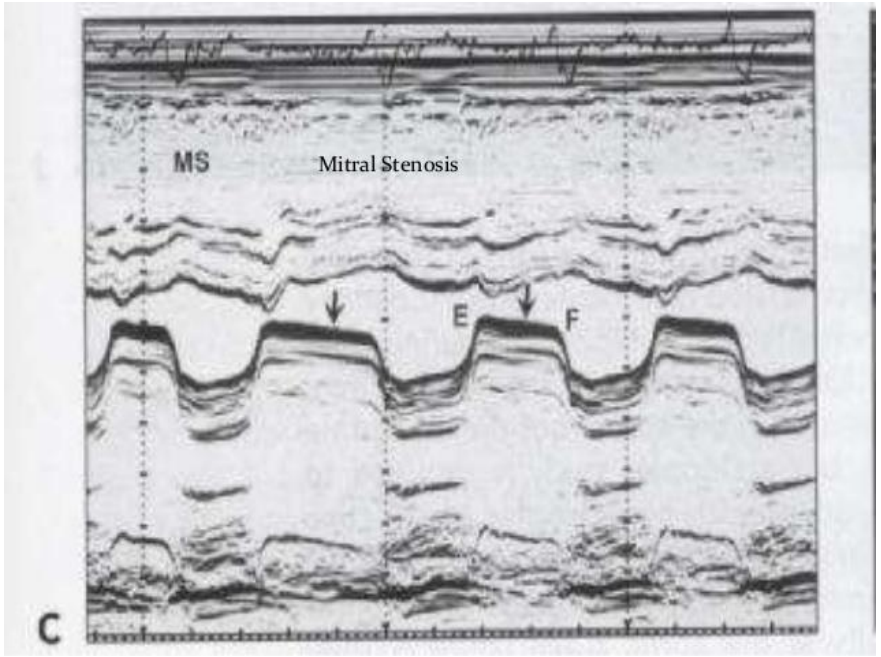


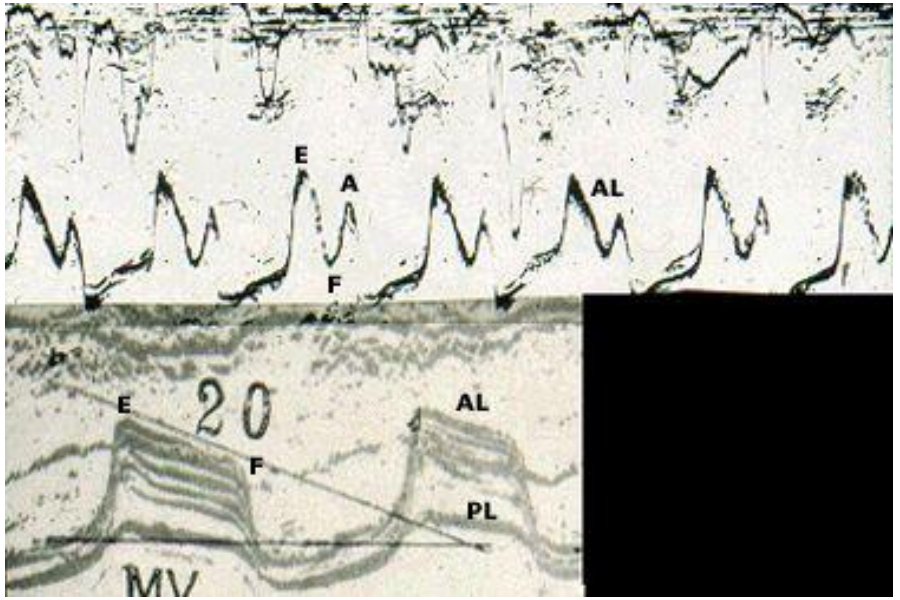
3D planimetry is a good way to ensure smallest mitral valve area is obtained

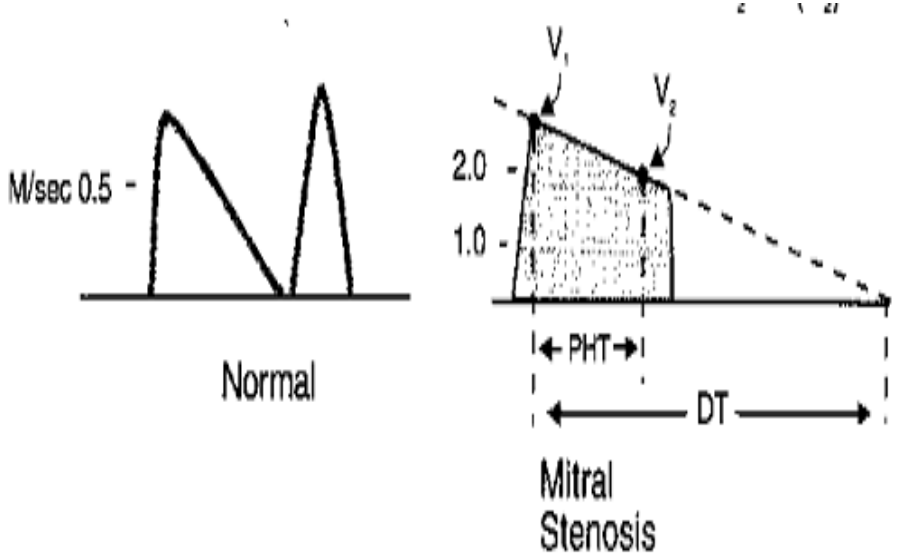
M-Mode pearls



View	Parasternal long axis
Modality	M-mode







PHT = time interval (msec) Between Max gradient and the time where the gradient is half the initial value ($P=4v^2$)

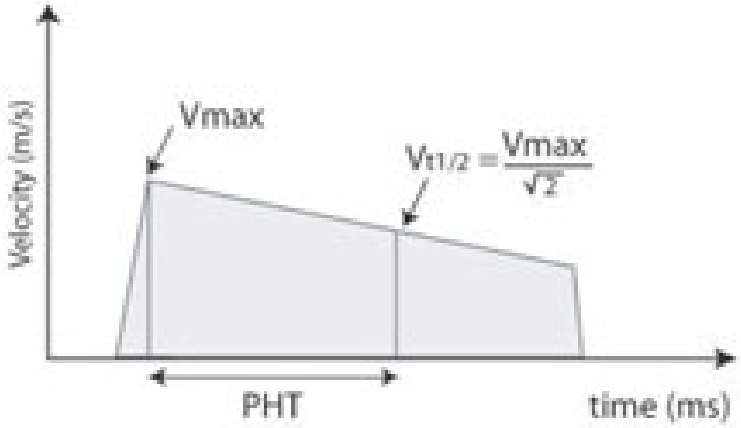
DT= time from Vmax to zero

MVA by pressure halftime:

MVA=220 / PHT

PHT = DT x 0.29

MVA = 759/DT



Mitral Stenosis Severity – ACC/AHA 2020

	MVA	
Progressive ***(Stage B)***	>1.5 cm2; PHT<150 msec, Mildly dilated LA, normal PASP	
Severe Asymptomatic ***(Stage C)***	<1.5 cm2; PHT>150 msec, Sev. dilated LA, PASP>50 mmHg	
Severe Symptomatic ***(Stage D)***	<1.5 cm2; PHT >150 msec, Sev. dilated LA, PASP>50 mmHg	++ DOE

* Very Severe = MVA <1.0 cm2

** Mean gradient >5-10 mmHg accompanies severe mitral stenosis, varies by HR/clinical conditions

nature reviews cardiology https://doi.org/10.1038/s41569-023-00940-9

Evidence-based guidelines Check for updates

2023 World Heart Federation guidelines for the echocardiographic diagnosis of rheumatic heart disease

Joselyn Rwebembera[1,38], James Marangou[2,3,4,38], Julius Chacha Mwita[5], Ana Olga Mocumbi[6], Cleonice Mota[7,8], Emmy Okello[1], Bruno Nascimento[9,10], Lene Thorup[11], Andrea Beaton[12,13], Joseph Kado[14,15], Alexander Kaethner[2,16], Raman Krishna Kumar[17], John Lawrenson[18,19], Eloi Marijon[20], Mariana Mirabel[21], Maria Carmo Pereira Nunes[9,10], Daniel Piñeiro[22], Fausto Pinto[23], Kate Ralston[24], Craig Sable[25], Amy Sanyahumbi[26], Anita Saxena[27], Karen Sliwa[28], Andrew Steer[29,30,31], Satupaitea Viali[32], Gavin Wheaton[33], Nigel Wilson[34], Liesl Zühlke[35,36] & Bo Reményi[2,16,37]

Screening for patients <20 yo in high risk areas:

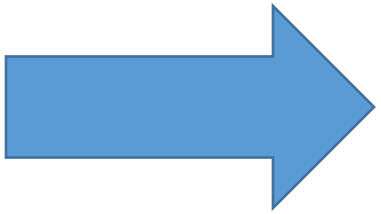
Mitral regurgitation :

-jet length >1.5 cm in pts <30 kg

-jet length >2 cm in pts >30 kg

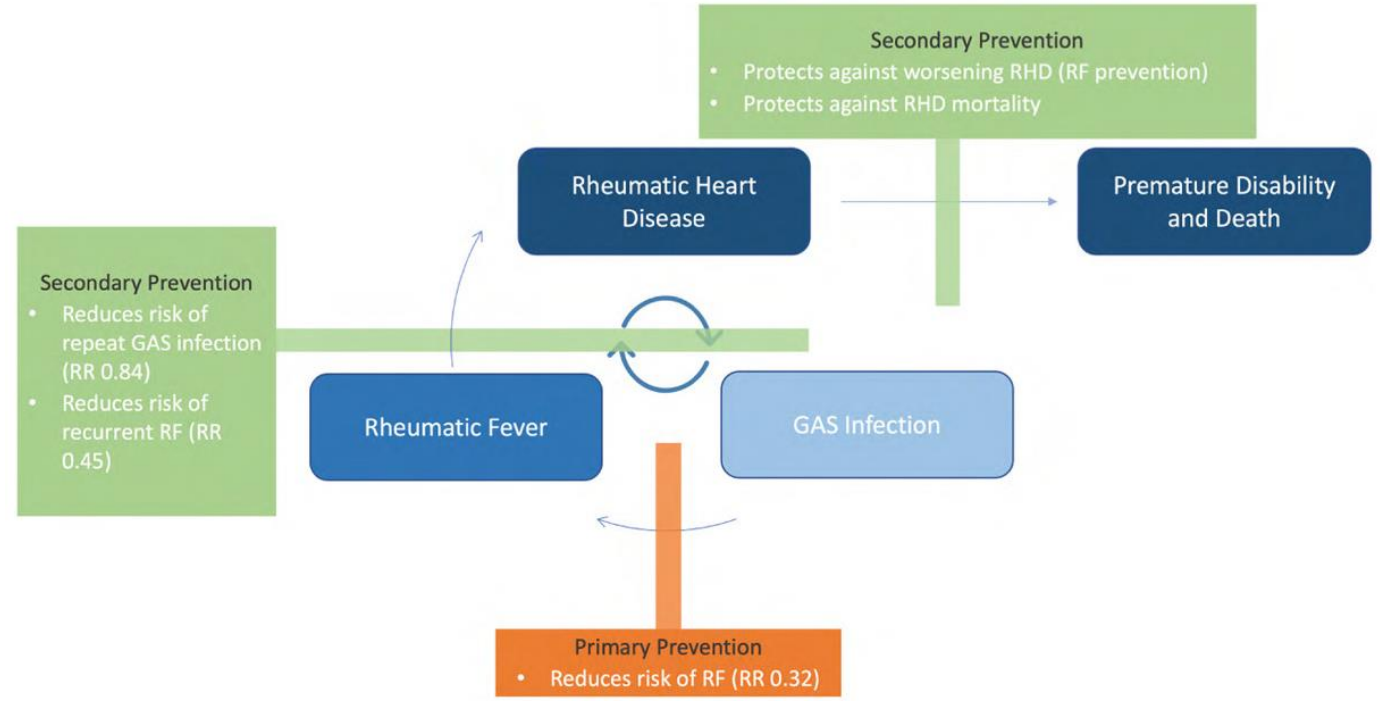
Aortic regurgitation

Restricted mitral leaflet mobility



If positive screening imaging, referral for echocardiography, more formal Doppler assessment

Mitral stenosis : mean gradient > 4 mmHg



Secondary antibiotic prophylaxis:

-Recurrent acute rheumatic fever is a risk until age 25-30 and worsens disease progression

-IM Benzathine Penicillin G every 4 weeks for 10 years or until age 40 (by AHA guidelines)

-WHO guidelines:

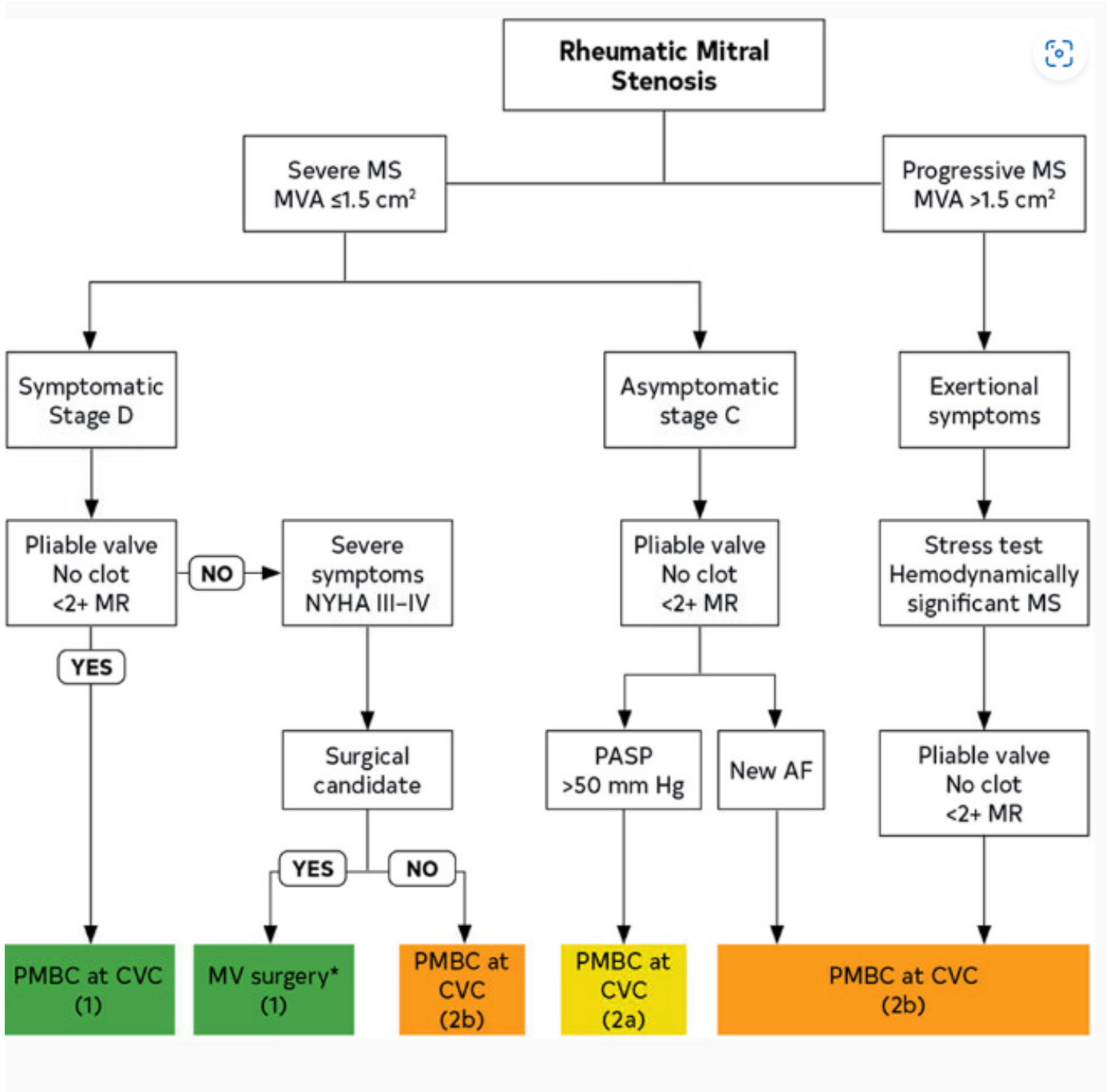
-10 years or until age 25

-lifelong if severe valve disease

Kumar et al, *Circulation* 2020

Mitral Stenosis: Important Questions

- Severity ?
- Symptomatic vs. Asymptomatic?
- Clinical Consequences:
  - LAA thrombus / Atrial Fib / Pulmonary HTN
  - Is mitral regurgitation also present ?
- Treatment?
  - Medical (rate control, anticoagulation)
  - Invasive
    - Balloon Valvuloplasty vs. Surgery


Rheumatic Mitral Stenosis
Severe MS MVA ≤1.5 cm²
Progressive MS MVA >1.5 cm²
Symptomatic Stage D
Asymptomatic stage C
Exertional symptoms
Pliable valve No clot <2+ MR
NO
Severe symptoms NYHA III–IV
Pliable valve No clot <2+ MR
Stress test Hemodynamically significant MS
YES
Surgical candidate
PASP >50 mm Hg
New AF
Pliable valve No clot <2+ MR
YES
NO
PMBC at CVC (1)
MV surgery* (1)
PMBC at CVC (2b)
PMBC at CVC (2a)
PMBC at CVC (2b)

Mitral Stenosis - Wilkins Score

Grade	Mobility	Subvalvular Thickening	Valvular Thickening	Calcification
1	Highly mobile valve with only leaflet tips restricted.	Minimal thickening just below the mitral leaflets.	Leaflets nearly normal in thickness (4-5 mm).	Single area of increased echo brightness.
2	Leaflet mid and basal portions normally mobile.	Thickening of chordal structures extending up to one third of chordal length.	Mid leaflets normal, considerable thickening of margins (5-8 mm).	Scattered areas of brightness confined to leaflet margins.
3	Valve continues to move forward in diastole, mainly from base.	Thickening extending to the distal third of chords.	Thickening extending through the entire leaflet (5-8 m m).	Brightness extending into midportion of the leaflets.
4	No or minimal forward movement of the leaflets in diastole	Extensive thickening and shortening of all chordal structures extending down to papillary muscles.	Considerable thickening of all leaflet tissue (>8-10 mm).	Extensive brightness throughout much of the leaflet tissue.

*Score ranges from 4-16

*8 or less is favorable for balloon valvuloplasty

Mitral Stenosis – Class 1 recommendations

- Percutaneous mitral balloon valvuloplasty/ commissurotomy (PMBC)
  - Severe, symptomatic (Stage D) MS: favorable anatomy with pliable valve, less than 2+ MR, no LA thrombus
- Mitral valve surgery (commissurotomy/replacement)
  - Severe, symptomatic (Stage D, NYHA III-IV) MS: non-pliable valve, 2+ or more MR

Mitral Stenosis – Diagnostic Testing

- Exercise Testing – Class I recommendation when symptoms out of proportion to MS severity by resting echo
  - Stress echo (treadmill, supine bike echo) or invasive cath
  - Assess MV gradients, PA pressure with stress

- 45 yo with rheumatic mitral stenosis and shortness of breath comes for cath. PA pressure 90/45 mmHg (mean 60), LVEDP 5 mmHg, C.O. 3L/min. Mitral valve area 1.0 cm2. Echo reveals fused commissures, mild thickening of mitral valve leaflets and chordal apparatus, focal calcification of anterior mitral leaflet, and mild MR. What is best next step in her care:

a) Beta Blocker and warfarin

b) IV epoprostenol

c) Surgical mitral valve replacement

d) Percutaneous balloon mitral valvuluplasty

- 45 yo with rheumatic mitral stenosis and shortness of breath comes for cath. PA pressure 90/45 mmHg (mean 60), LVEDP 5 mmHg, C.O. 3L/min. Echo reveals fused commissures, mild thickening of mitral valve leaflets and chordal apparatus, focal calcification of anterior mitral leaflet, mild MR, mitral valve area 1.0 cm2. What is best next step in her care:

a) Beta blocker and warfarin

b) IV epoprostenol

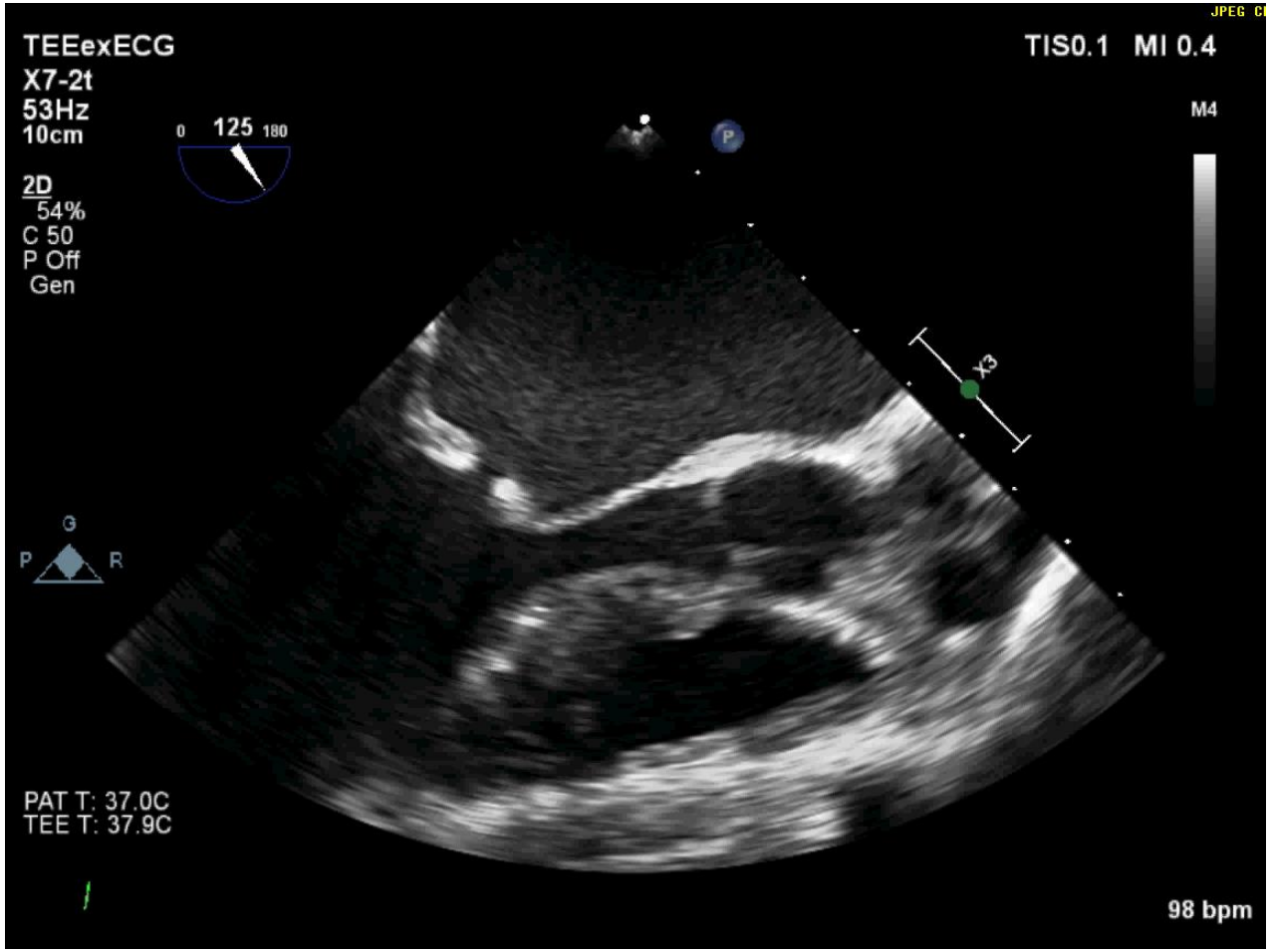
c) Surgical mitral valve replacement

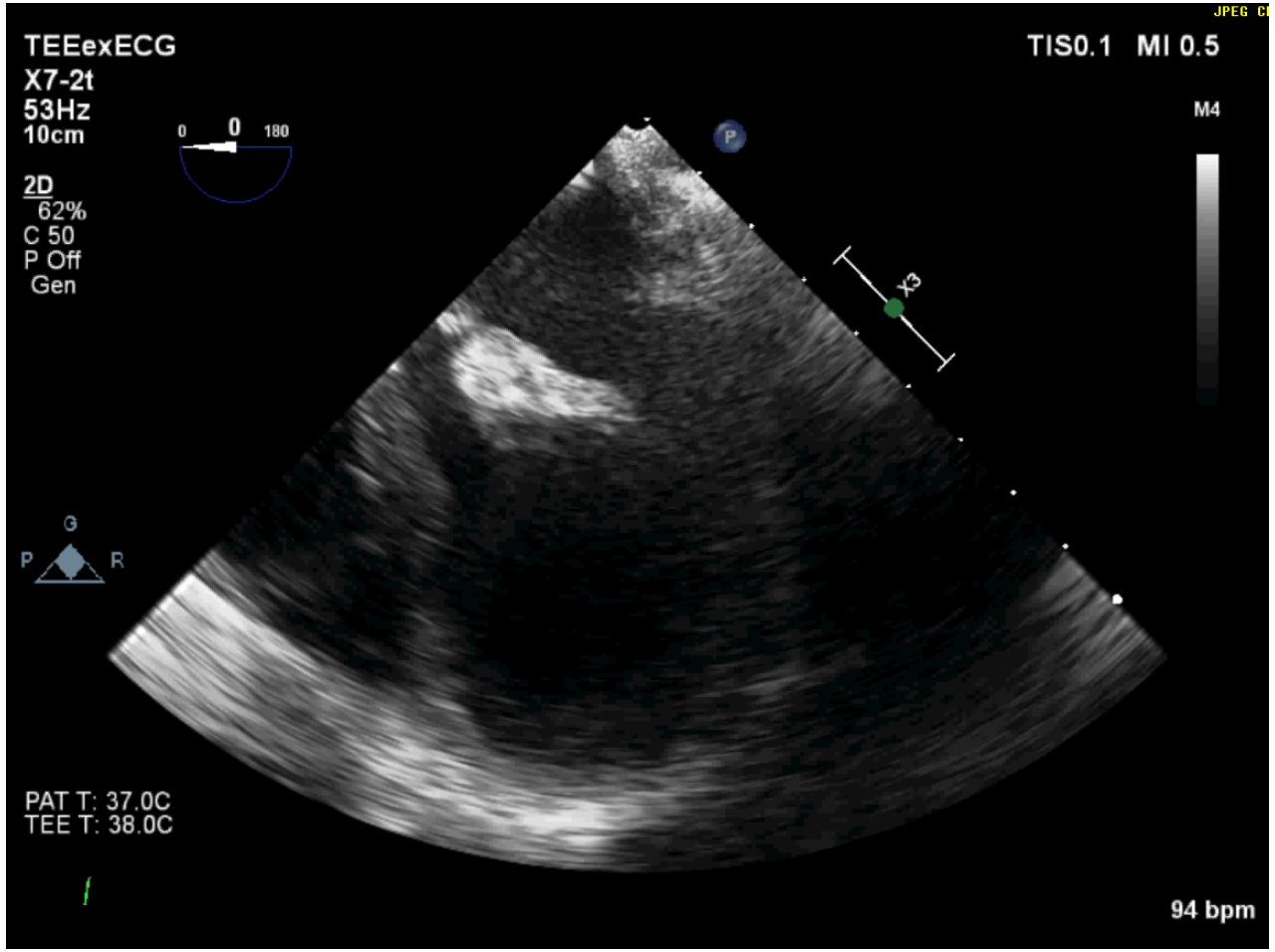
d) **Percutaneous balloon mitral valvuluplasty**

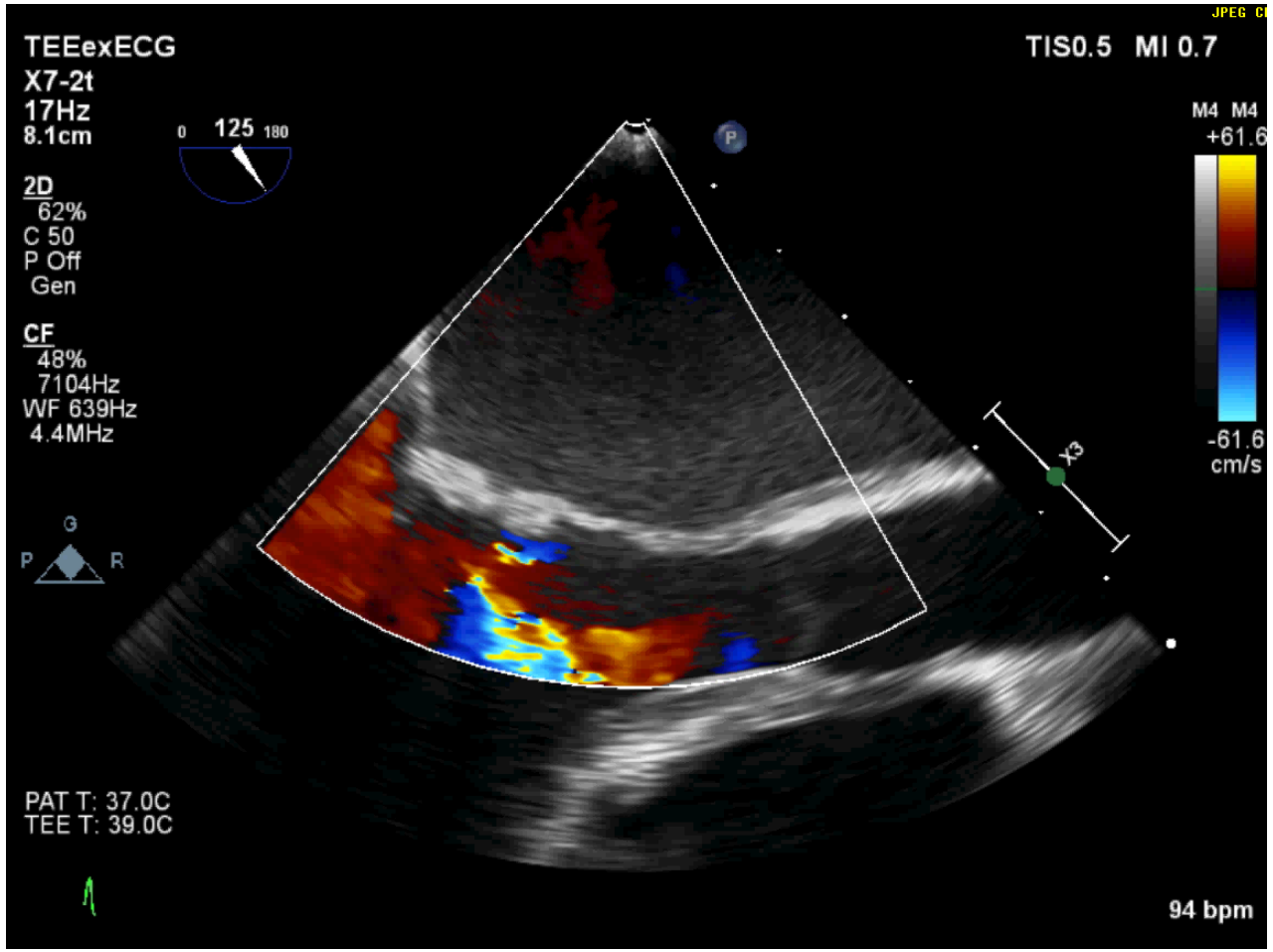
Mitral Stenosis

- Percutaneous mitral balloon valvuloplasty is indicated for severe, symptomatic MS and favorable anatomy, no LA thrombus

Mitral stenosis: pre-valvuloplasty




TEEexECG
X7-2t
53Hz
10cm
2D
62%
C 50
P Off
Gen
0
0
180
TIS0.1 MI 0.5
M4
X3
G
P
R
PAT T: 37.0C
TEE T: 38.0C
94 bpm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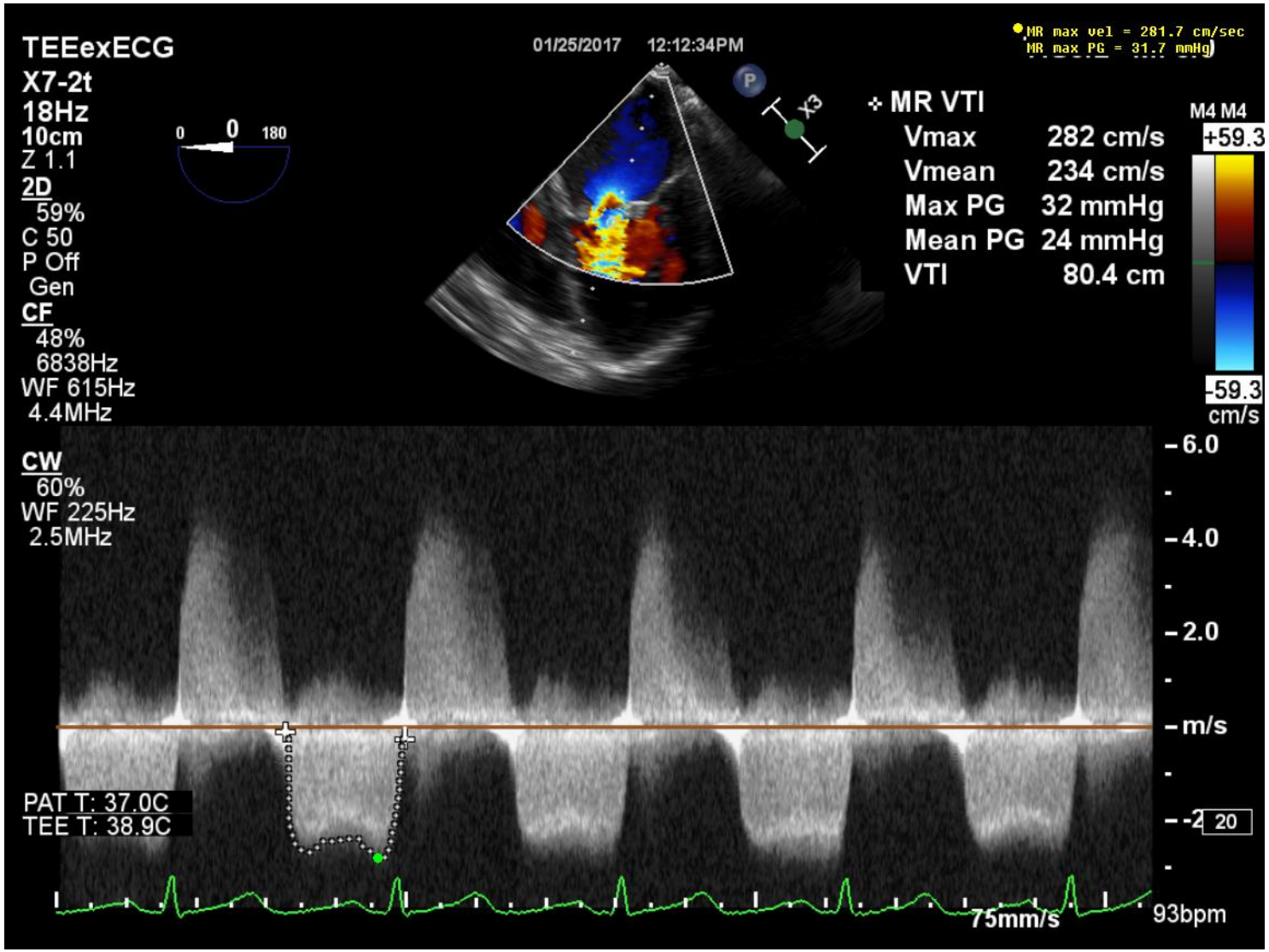
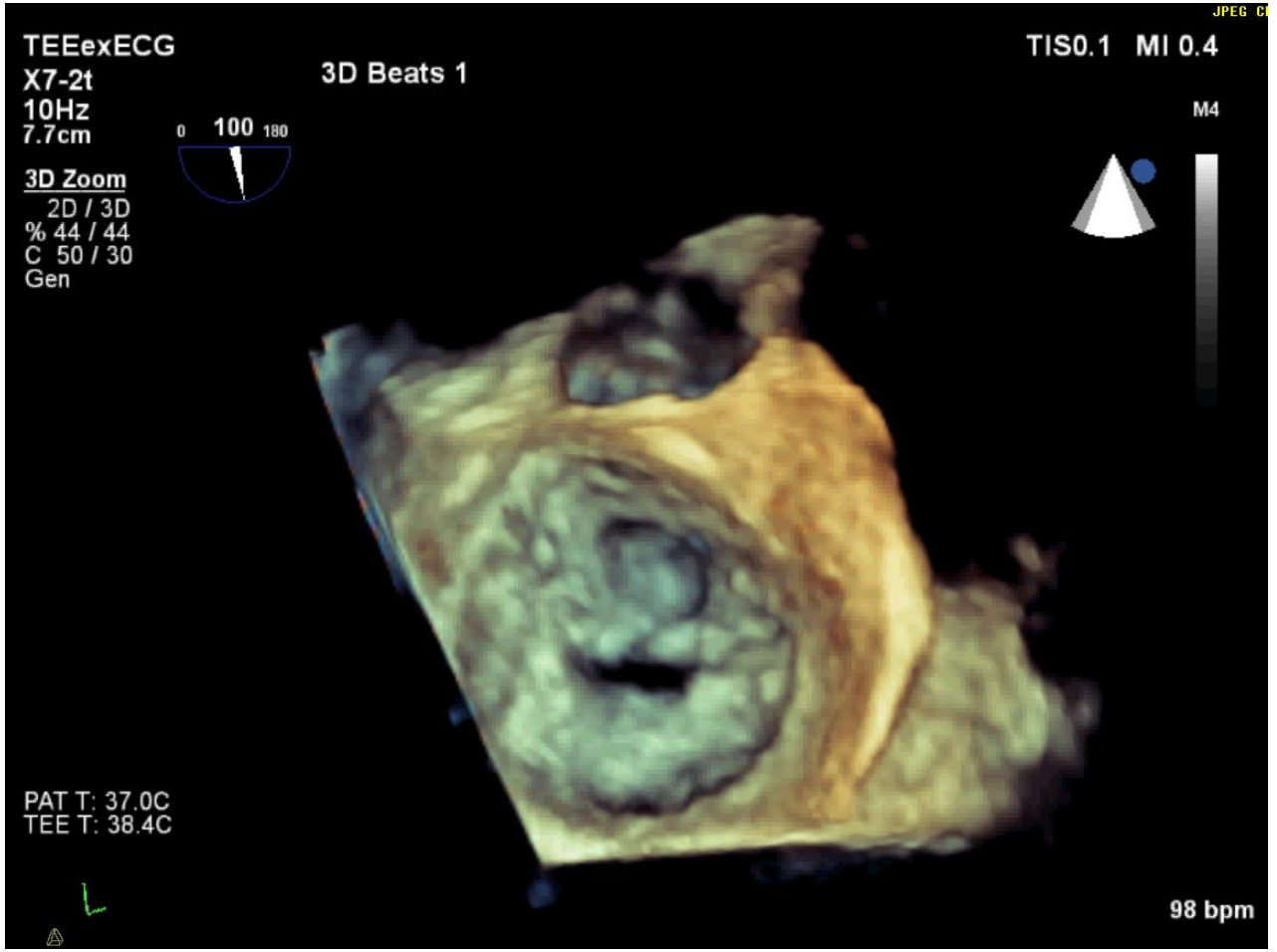
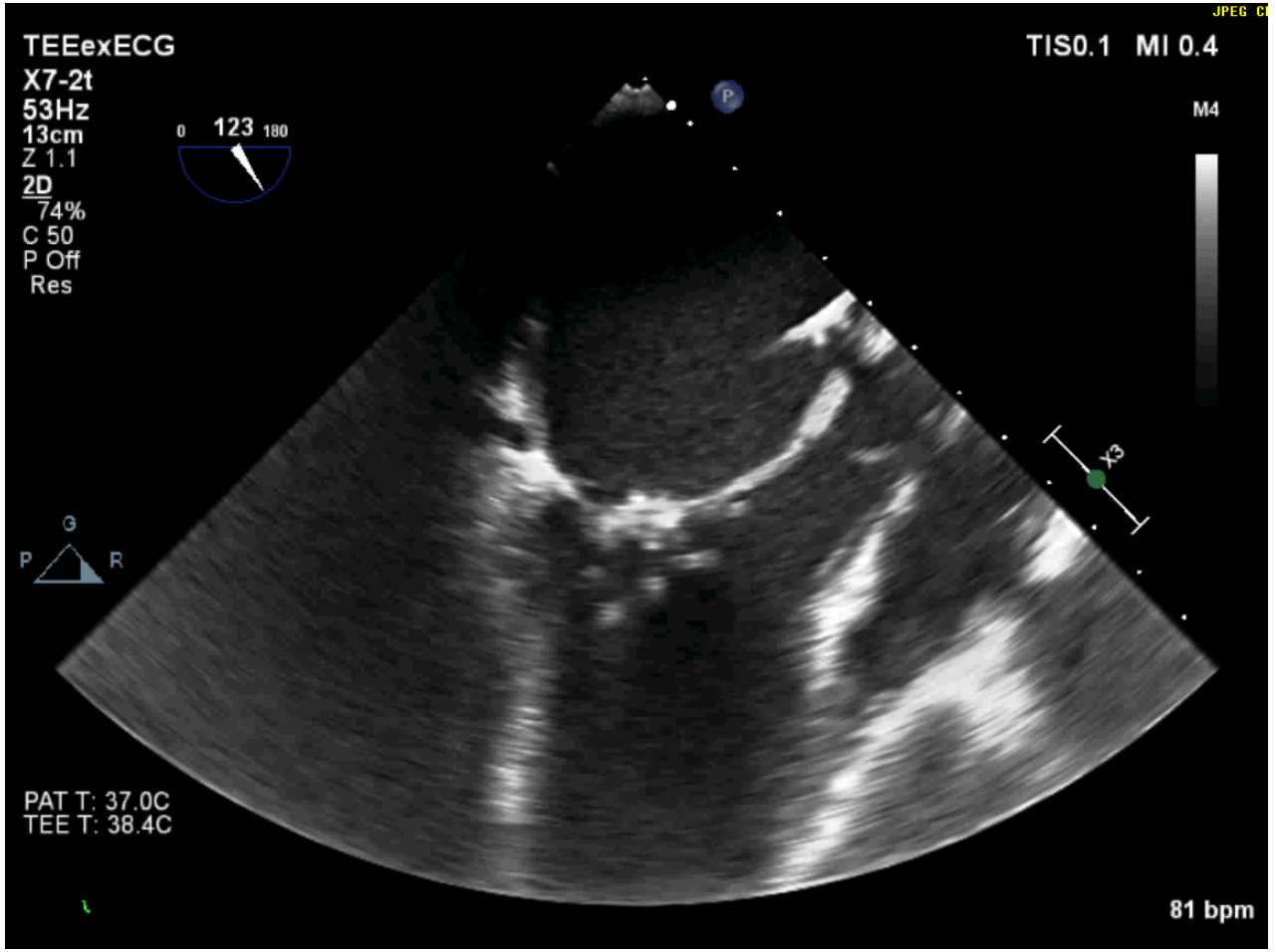
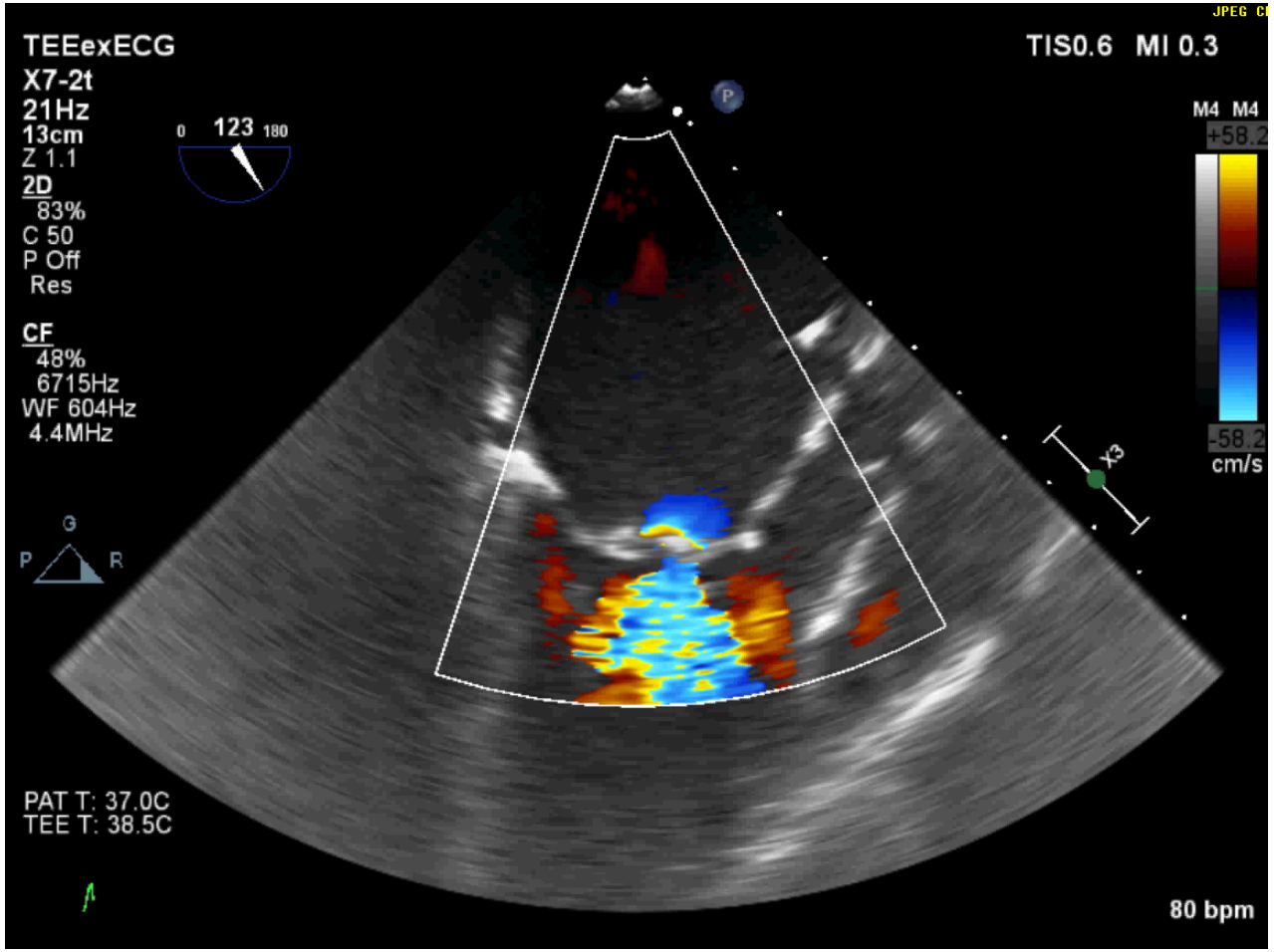
X7-2t
17Hz
8.1cm
0
125
180
2D
62%
C 50
P Off
Gen
CF
48%
7104Hz
WF 639Hz
4.4MHz
G
P
R
TIS0.5 MI 0.7
M4 M4
+61.6
-61.6
cm/s
X3
PAT T: 37.0C
TEE T: 39.0C
94 bpm

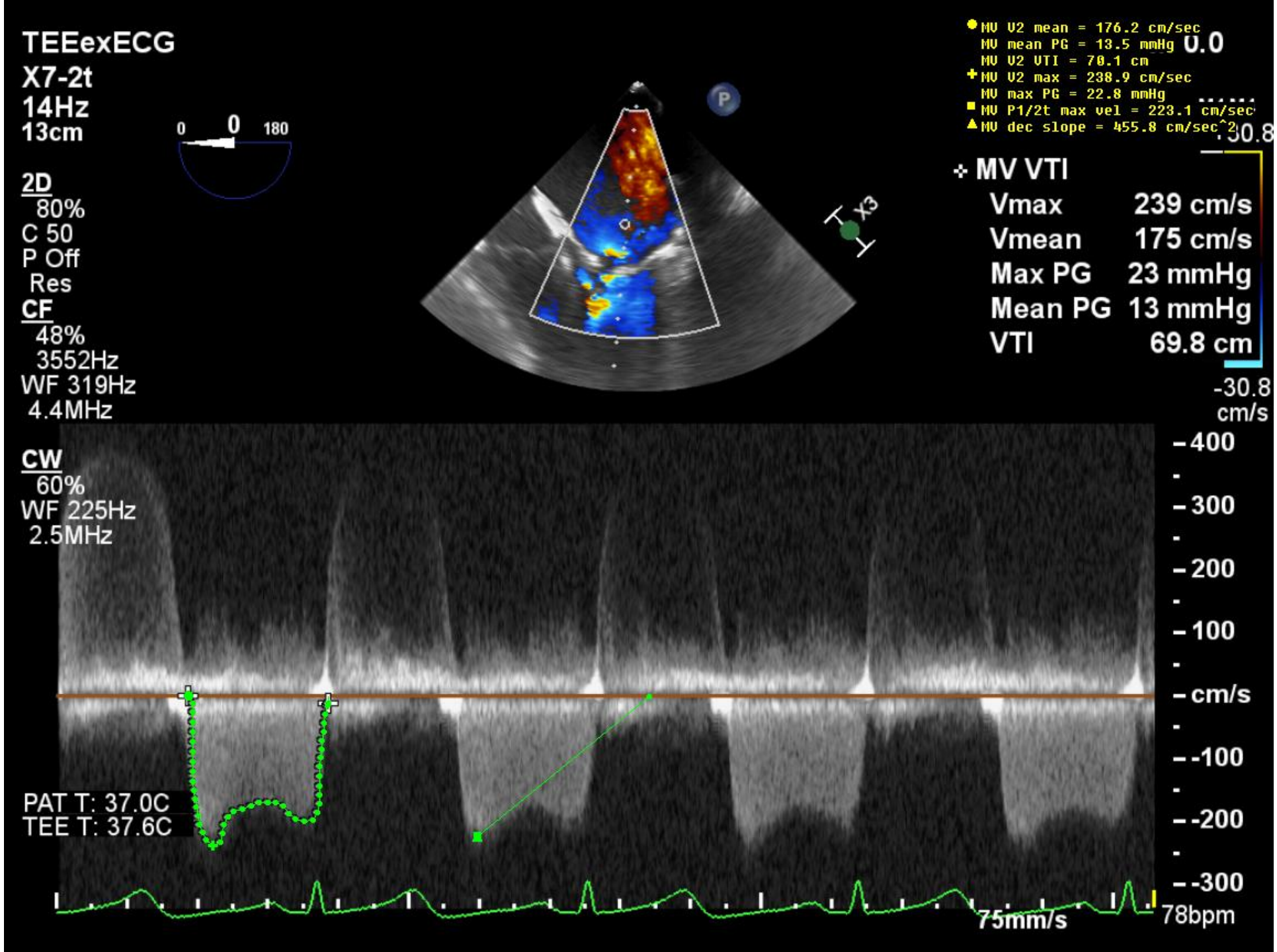

TEEexECG
X7-2t
18Hz
10cm
Z 1.1
2D
59%
C 50
P Off
Gen
CF
48%
6838Hz
WF 615Hz
4.4MHz
01/25/2017 12:12:34PM
MR max vel = 281.7 cm/sec
MR max PG = 31.7 mmHg
MR VTI
Vmax 282 cm/s
Vmean 234 cm/s
Max PG 32 mmHg
Mean PG 24 mmHg
VTI 80.4 cm
M4 M4
+59.3
-59.3
cm/s
CW
60%
WF 225Hz
2.5MHz
6.0
4.0
2.0
m/s
20
PAT T: 37.0C
TEE T: 38.9C
75mm/s
93bpm


TEEexECG
X7-2t
10Hz
7.7cm
3D Beats 1
TIS0.1 MI 0.4
M4
0 100 180
3D Zoom
2D / 3D
% 44 / 44
C 50 / 30
Gen
PAT T: 37.0C
TEE T: 38.4C
98 bpm

Post-valvuloplasty




TEEexECG
X7-2t
21Hz
13cm
Z 1.1
2D
83%
C 50
P Off
Res
CF
48%
6715Hz
WF 604Hz
4.4MHz
0
123
180
TIS0.6 MI 0.3
M4 M4
+58.2
-58.2
cm/s
X3
G
P
R
PAT T: 37.0C
TEE T: 38.5C
80 bpm


TEEexECG
X7-2t
14Hz
13cm
0 0 180
2D
80%
C 50
P Off
Res
CF
48%
3552Hz
WF 319Hz
4.4MHz
CW
60%
WF 225Hz
2.5MHz
MV V2 mean = 176.2 cm/sec
MV mean PG = 13.5 mmHg
MV V2 VTI = 70.1 cm
MV V2 max = 238.9 cm/sec
MV max PG = 22.8 mmHg
MV P1/2t max vel = 223.1 cm/sec
MV dec slope = 455.8 cm/sec^2
0.0
-30.8
MV VTI
Vmax 239 cm/s
Vmean 175 cm/s
Max PG 23 mmHg
Mean PG 13 mmHg
VTI 69.8 cm
-30.8
cm/s
400
300
200
100
cm/s
-100
-200
-300
PAT T: 37.0C
TEE T: 37.6C
75mm/s
78bpm

- A 55 yo woman w/ mitral stenosis s/p successful mitral valvuloplasty 2 years ago comes for follow up complaining of progressive dyspnea. Previously able to do Zumba classes, now walking her dog is difficult. She has gained weight. Exam notable for soft, early diastolic rumble preceded by a crisp opening snap. There is a wide separation of A2 opening snap interval. Her echo shows mean transmitral gradient 4 mmHg, MVA by echo 2.0 cm2. HR is 60 bpm. You recommend:

a) Supine bike stress echo

b) Start Metoprolol and Lasix

c) TEE

d) Cardiac MRI

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c) TEE

d) Cardiac MRI

- Stress Testing – recommendation when symptoms out of proportion to echo estimated MS severity
  - Stress Echo (treadmill, supine bike echo, or cath)
  - Assess MV gradients, PA pressure with stress
  - Abnormal: Mean gradient >15 mmHg, PASP > 60 mmHg , PCWP >25

You are consulting on a 30 year old woman with symptomatic severe mitral stenosis presenting for mitral balloon valvuloplasty. She is in NSR. TEE prior to the procedure reveals a Wilkins score of 4, mild MR, and left atrial thrombus. She currently takes asa 81, estrogen-containing OCP, Lasix, and metoprolol. You recommend:

a) Discontinue birth control pill
b) Begin Xarelto 20 mg daily.
c) Start LMWH bridge to warfarin with periodic INR monitoring
d) Referral for urgent mitral valve replacement surgery

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d) Referral for urgent mitral valve replacement surgery

- Class I indication for Vitamin K antagonist for mitral stenosis and:
  - left atrial thrombus
  - atrial fibrillation
  - prior embolic event

  - Patients with mitral stenosis and mechanical valve replacements were excluded in DOAC trials. DOACs have been studied in observational studies but randomized studies are few and inconclusive for changing practice.

- A patient is referred in her 28th week of 1st pregnancy with murmur. She reports murmur as a child. A few weeks ago, developed dyspnea and orthopnea. On exam, HR is 112 bpm and regular, BP 125/72 mmHg, minimal bibasilar rales, 2/4 diastolic murmur heard at the apex. JVP is mildly elevated, no edema. ECG shows sinus tachycardia. Echo shows rheumatic deformity of mitral valve with mean gradient 8 mmHg. Which is best medical therapy to help with symptoms?

a) Digoxin
b) Amiodarone
c) Lisinopril
d) Metoprolol

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c) Lisinopril

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- Pregnancy can uncover symptoms of mitral stenosis due to increased HRs and plasma volumes (peaks at end of second trimester). Medical therapy with metoprolol can slow the HR, increase diastolic filling time, and improve symptoms.
- Balloon valvuloplasty with shielding can be considered if patients fail medical therapy, safest after 1st trimester.
- Mitral valve surgery during pregnancy has a high mortality rate and should be a last resort

Thank you from Dallas, TX!

UT Southwestern
Medical Center

Parkland