

# **Intensive Review of Internal Medicine**



***CAN'T MISS CARDIOLOGY  
DIAGNOSES, DECISIONS,  
AND EKGs***



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- Executive Vice Chair, Department of Medicine
- Kraft Family Distinguished Chair in Cardiovascular Medicine
- No disclosures

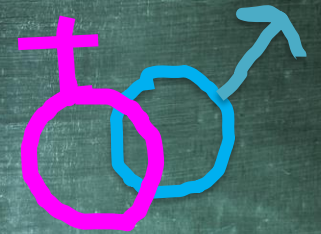


WELCOME!

simple



# Rules



1.

10-19 cases, depending on time.



2.

Nothing unusual

3.

Can be figured out with a history, good cardiac examination & EKG



4.

Think about epidemiology and physiology

5.



Coronary, valvular, myocardial, pericardial and vascular conditions



H<sub>2</sub>O



$$E=mc^2$$







## Case Study: 60 YO Woman

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Upper neck and back discomfort x 2 weeks (happened once when walking up stairs with briefcase)

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Usually occurs at night as she lies down in bed

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No SOB

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No Diaphoresis

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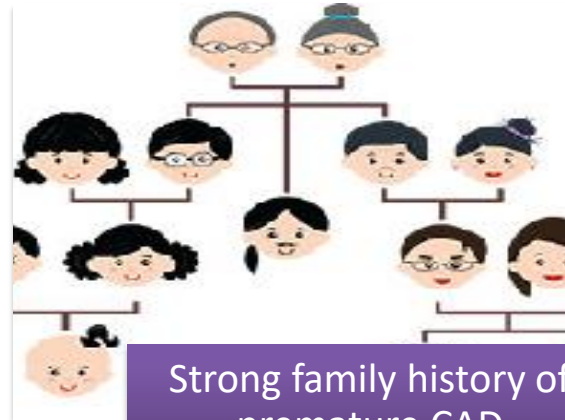


# Case Study: 60 YO Woman

## Medical History



Chronic left rotator cuff difficulties



Strong family history of premature CAD



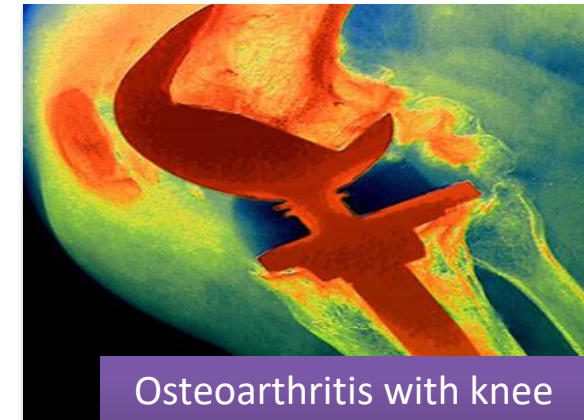
BMI 32



HTN, vigorously treated  
Type 2 DM, A1c 7.2%



Dyslipidemia treated with statin, TC 185 HDL 50 LDL 98  
TG 211



Osteoarthritis with knee replacement 2 years prior

# Case Study: 60 YO Woman - Medications





# Case Study 60 YO Woman



150/80 mmHg HR 100 bpm RR 14

JVP not elevated

Carotid upstrokes and volumes normal

Clear lungs

PMI non-displaced

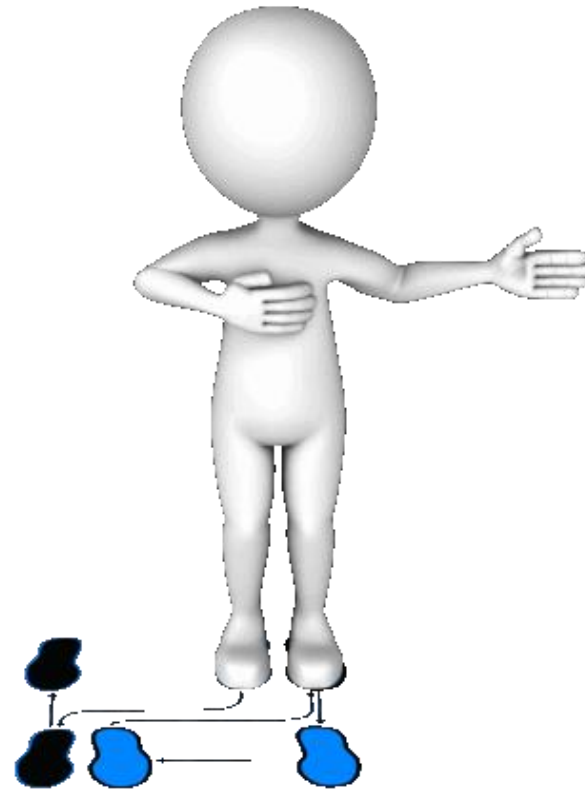
$S_1$  normal  $S_2$  Ø

2/6 systolic murmur, LSB, early peaking, decreases with Valsalva

Left arm hurts with rotation

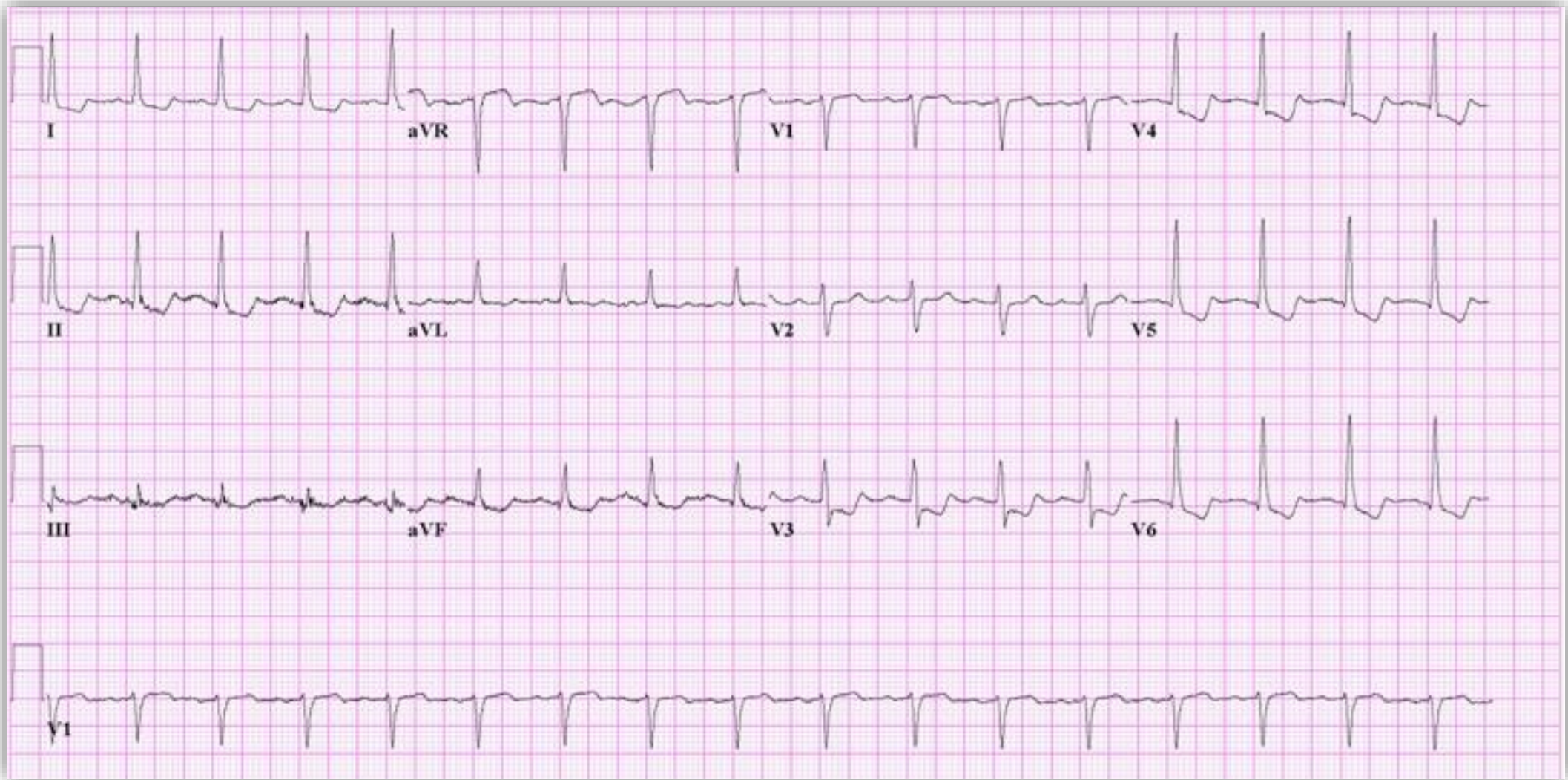
Has neck sensation during exam

# What are the Appropriate Next Steps?



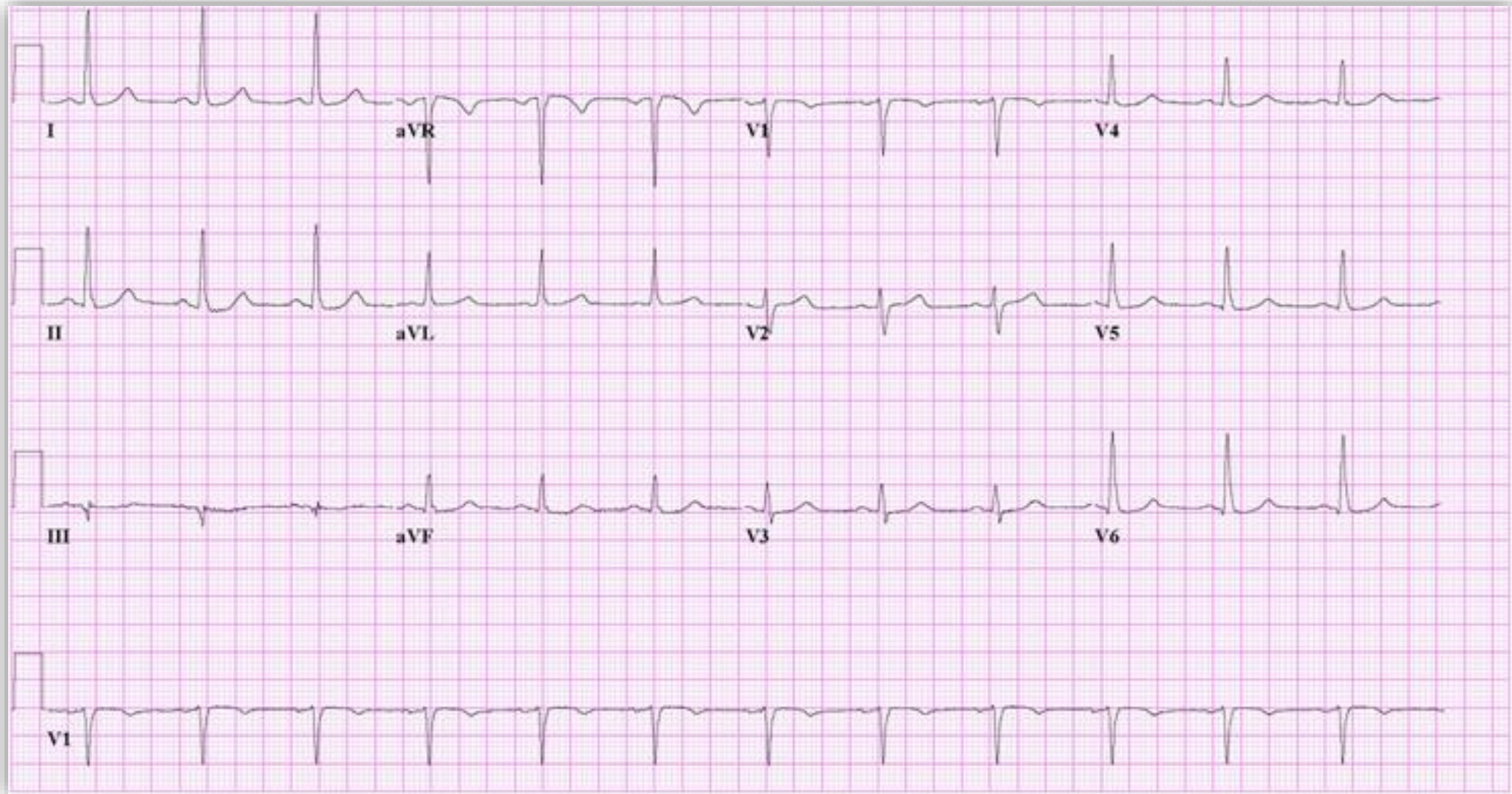


# Case #1: EKG-with discomfort



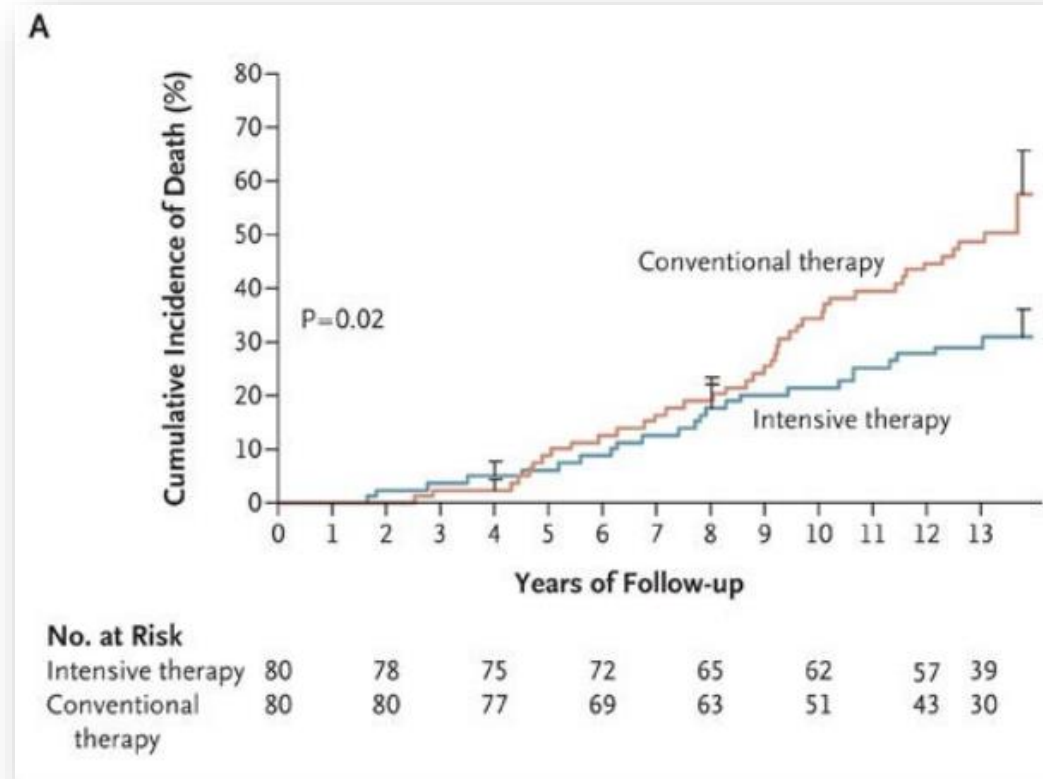


# Case #1: EKG-feeling better



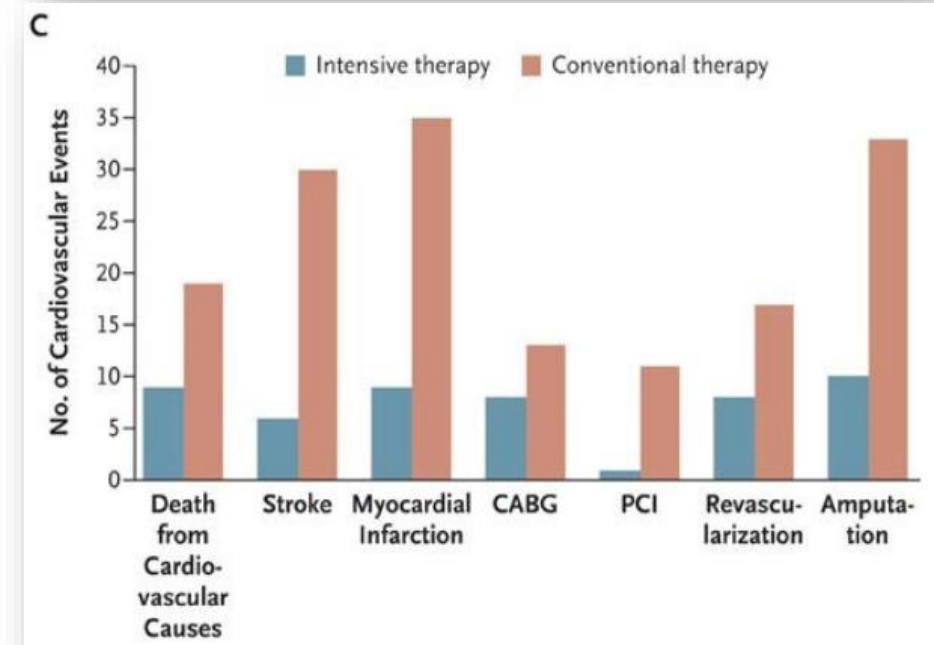
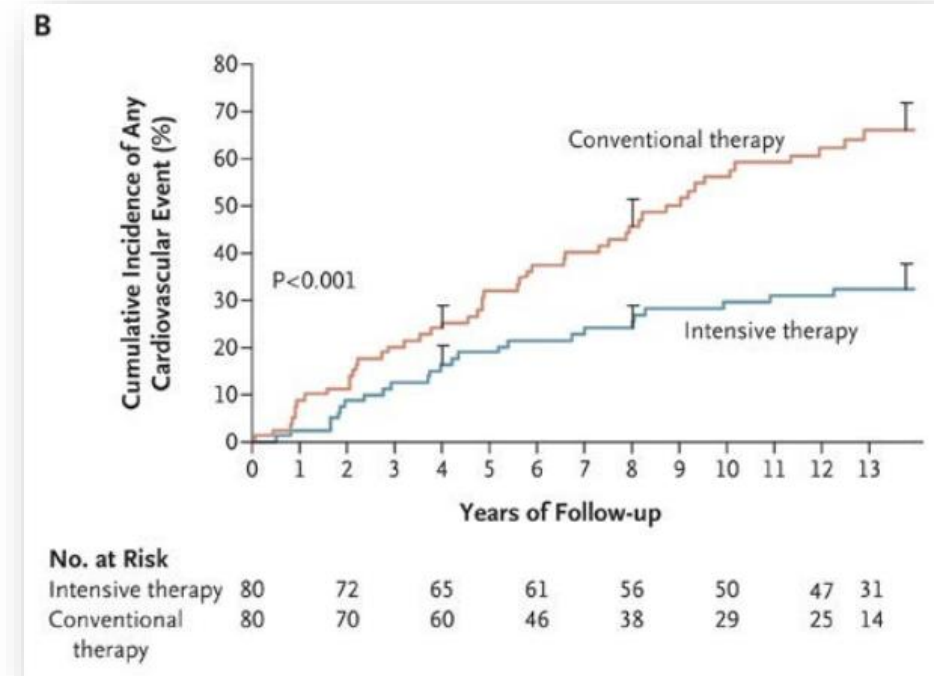


# Multifactorial intervention '08, BP, ace, statin, A<sub>1c</sub>, smoking cx

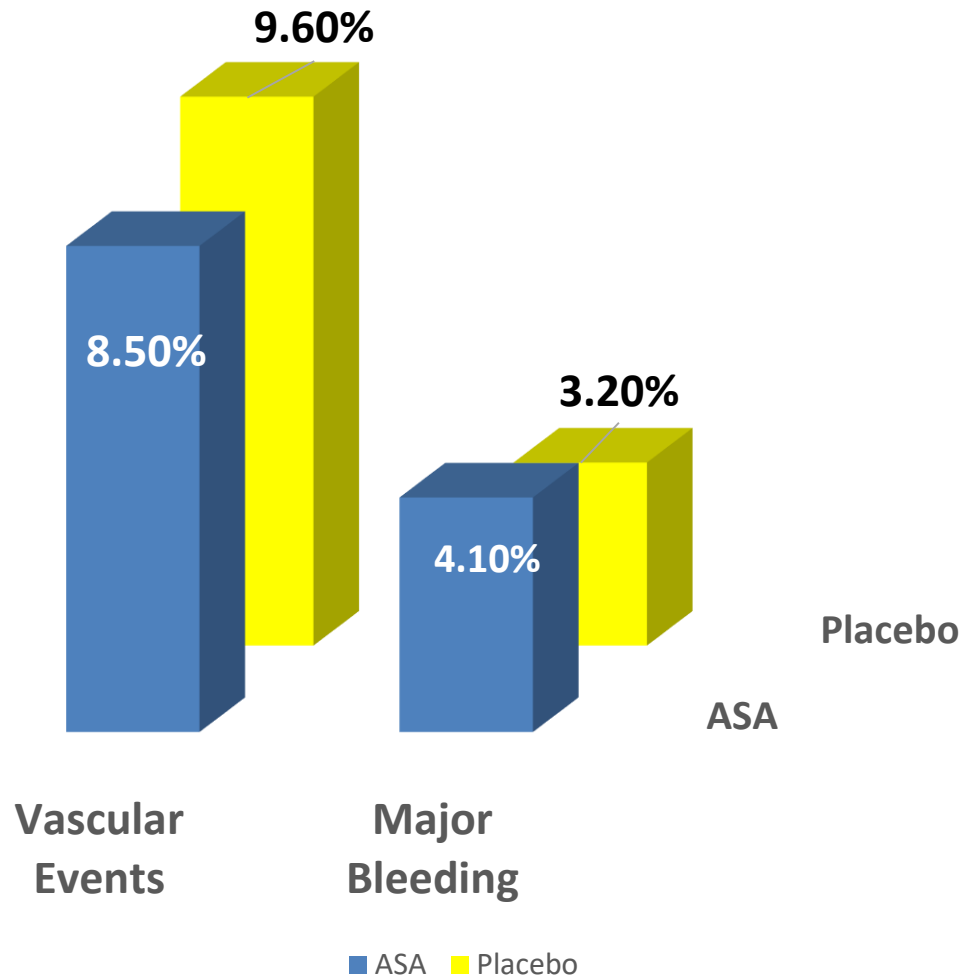


Gaede P, Lund-Anderson H, Parving HH, Pedersen O. Effect of a multifactorial intervention on mortality in type 2 diabetes. NEJM 2008; 358:580-91.

Multiple advancements since 2008



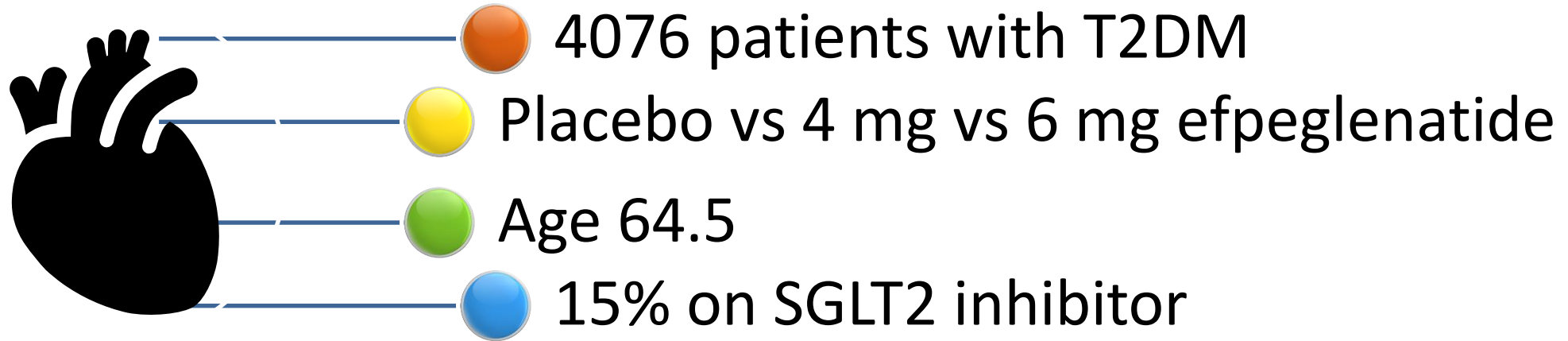
# ASCEND Study Results



	ASA	Placebo	HR
Vascular Events	8.5%	9.6%	0.88 (0.79-0.97)
Major Bleeding	4.1%	3.2%	1.29 (1.09-1.52)



## Effects of Efpeglenatide on CV Outcomes



### Outcomes:

- CV death, nonfatal MI, nonfatal stroke, ⊕Revasc ⊕Admission for unstable angina
- Combined renal: incident macroalbuminuria
  - >30% rise in UA Cr, EGFR ↓ by more than 40%, EGFR <15, renal replacement therapy



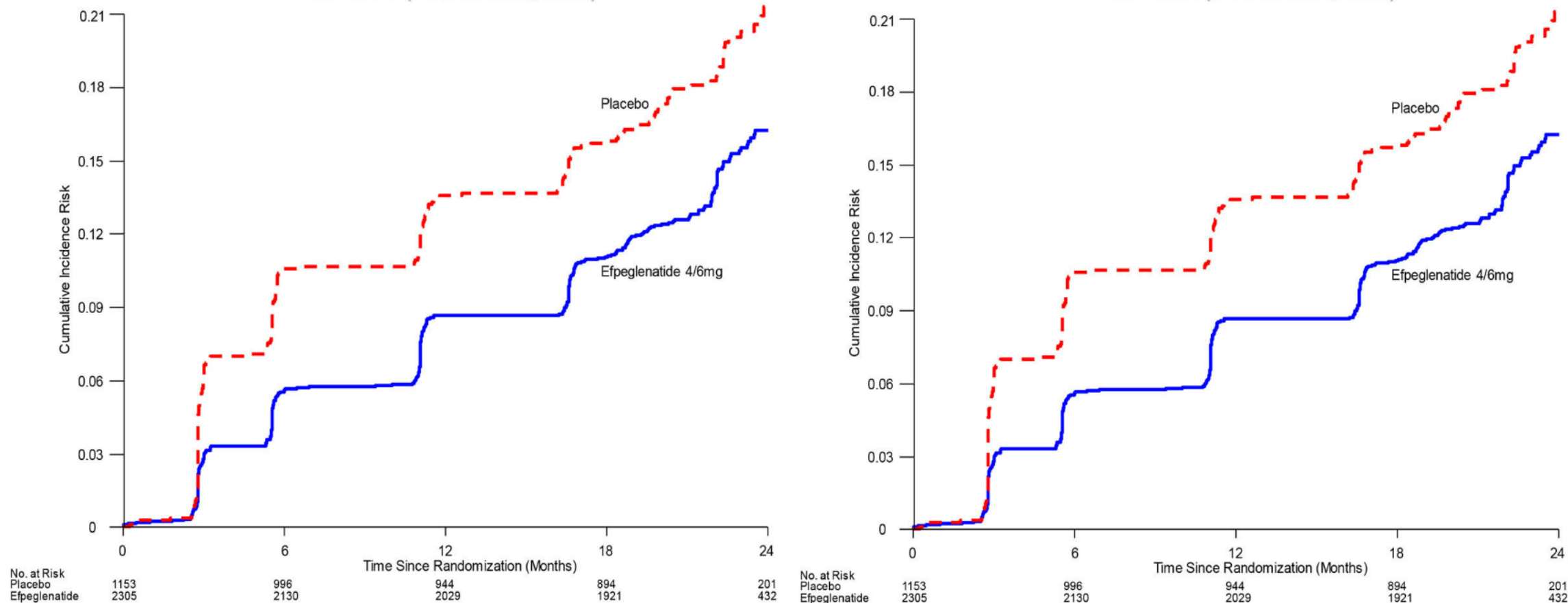
## Results at 1.8 Years as expected

CV Death, Nonfatal MI, Nonfatal Stroke	0.73	(0.58-0.92)
Additional Revasc, Unstable Angina	0.79	(0.65-0.91)
Renal Composite	0.68	(0.57-0.79)
Heart Failure Hospitalization	0.61	(0.38-0.98)

Learned that for the patients on SGLT2 inhibitor, outstanding effects on HF and renal outcomes; others, some benefit

**C****Renal Composite****No Baseline SGLT2 Inhibitor**  
HR 0.70 (95%CI 0.59, 0.83)

Interaction P = 0.38

**Baseline SGLT2 Inhibitor**  
HR 0.52 (95%CI 0.33, 0.83)



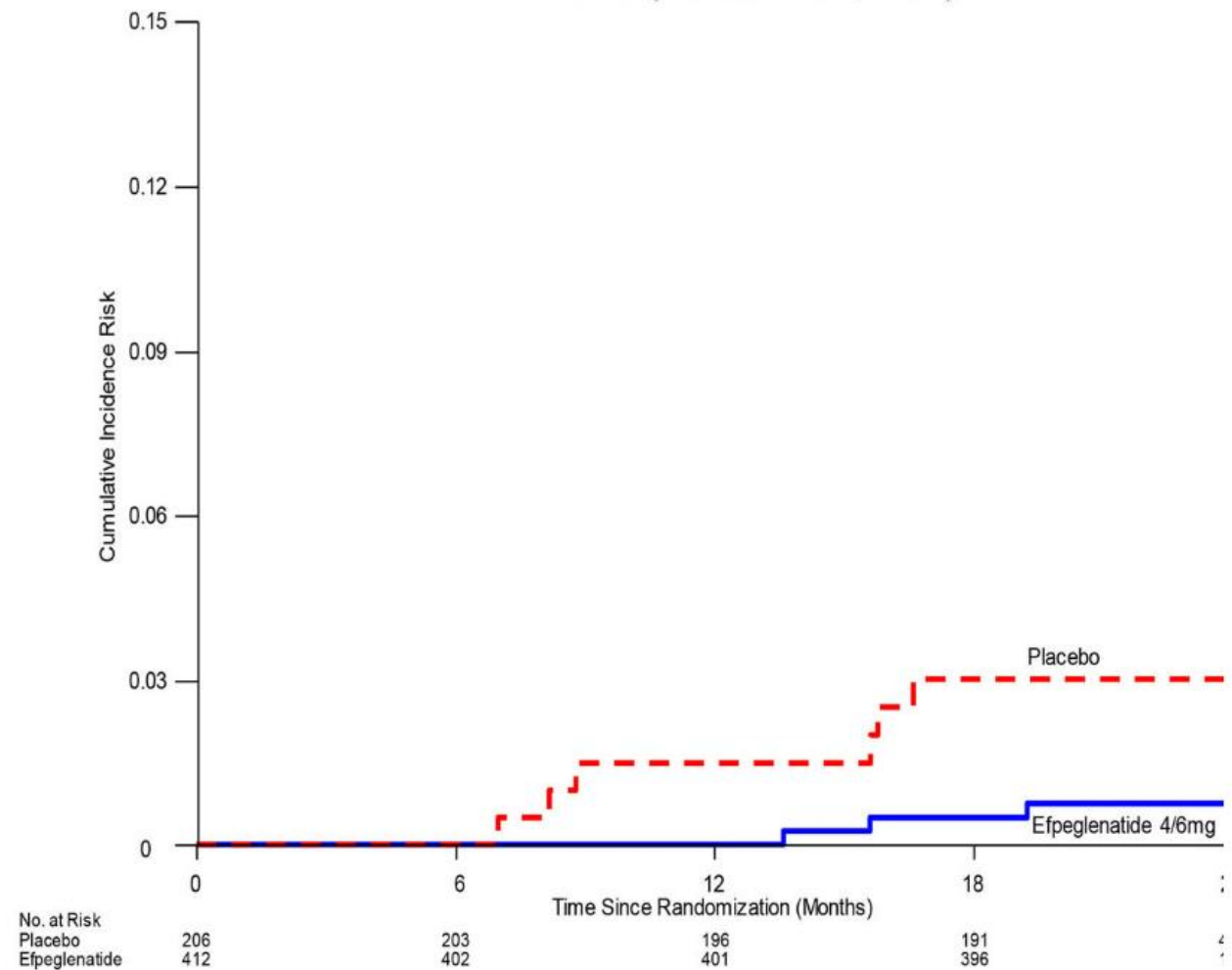
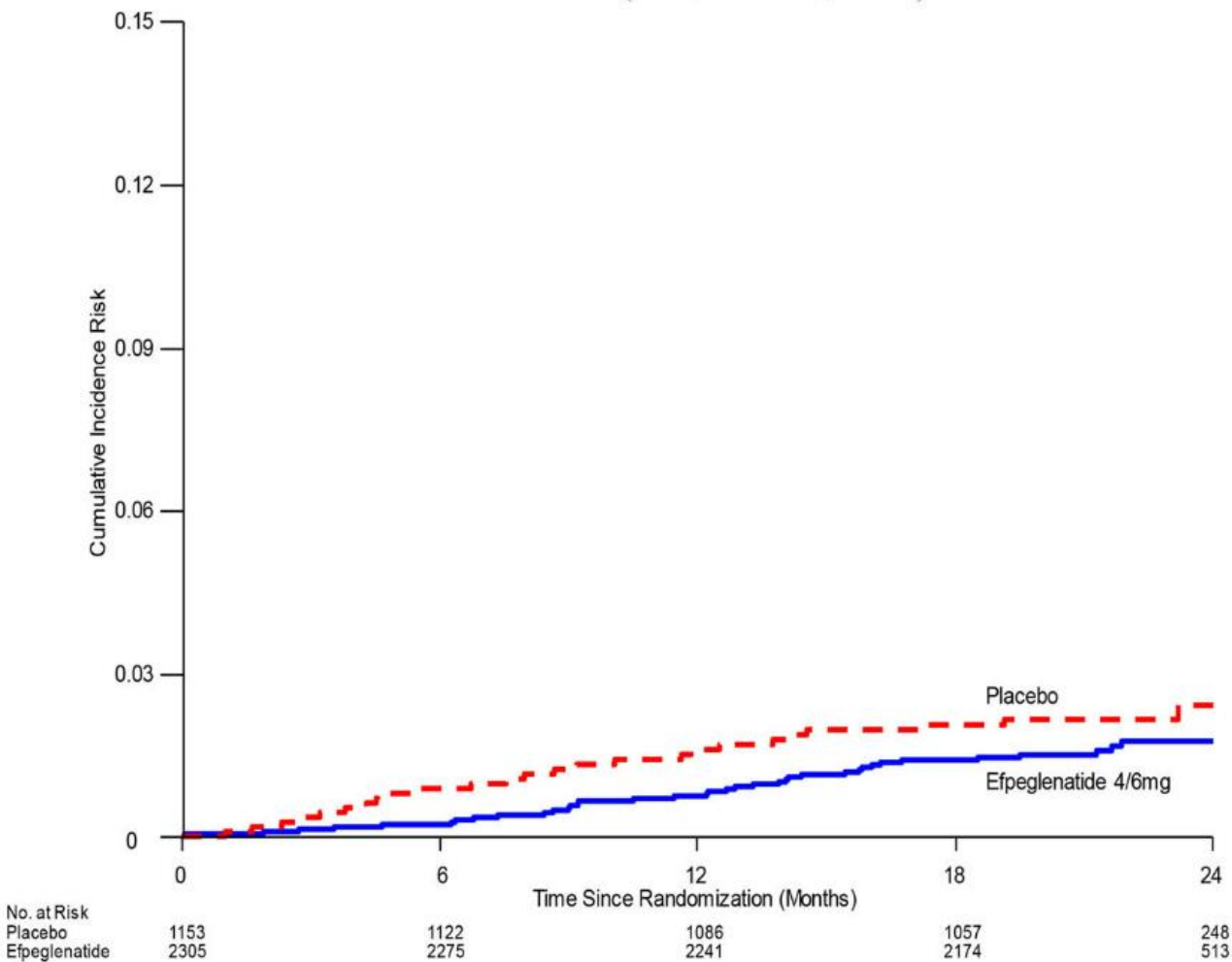
E

Heart Failure Hospitalization

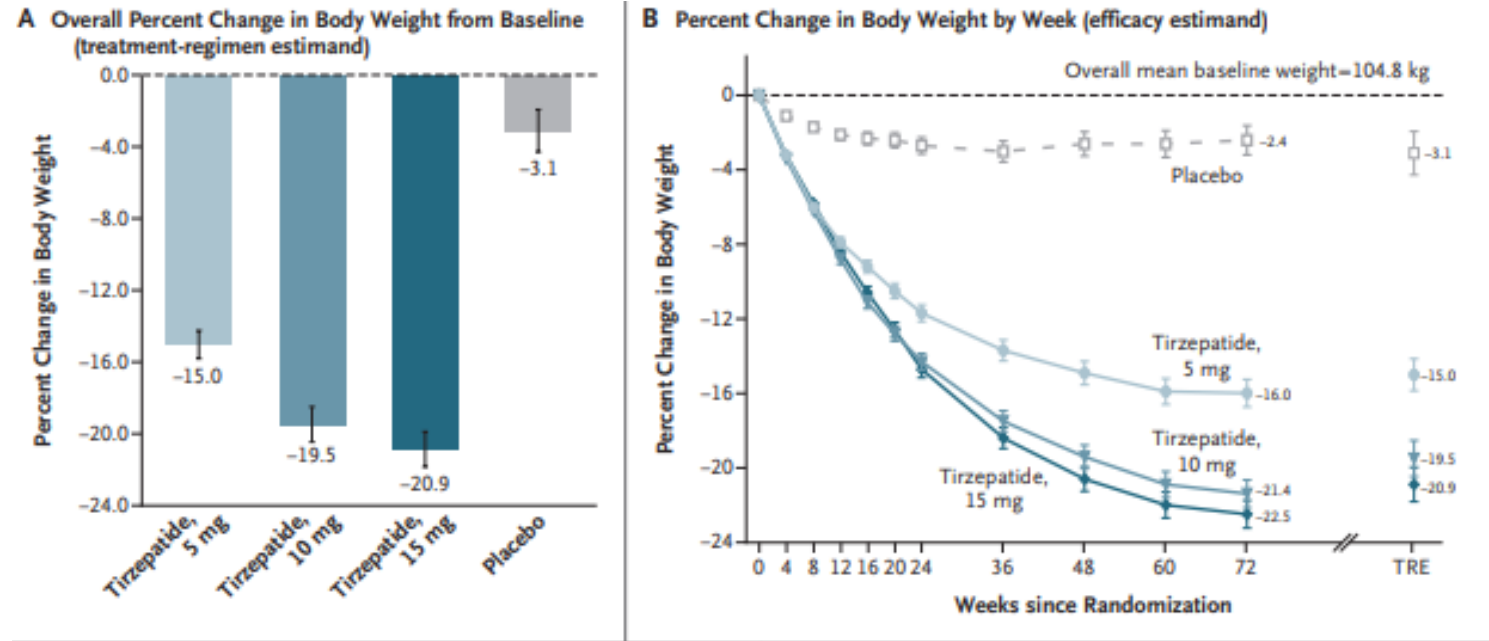
No Baseline SGLT2 Inhibitor  
HR 0.70 (95%CI 0.42, 1.17)

Interaction P = 0.35

Baseline SGLT2 Inhibitor  
HR 0.23 (95%CI 0.05, 0.97)



# Tirzepatide Study: SURMOUNT 1



## With 20 mg Dose

Weight reduction of >10%	83.5%
Weight reduction of >20%	56.7%
Weight reduction of >25%	36.0%

Improvement in physical activity, waist circumference, lipids, systolic and diastolic BP

Side effects – 3 fold > nausea, diarrhea, constipation, consistent with placebo and usually transient

No increase in pancreatitis, biliary or renal events

# Anatomy

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Normal left main

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95% LAD stenosis after  
first diagonal

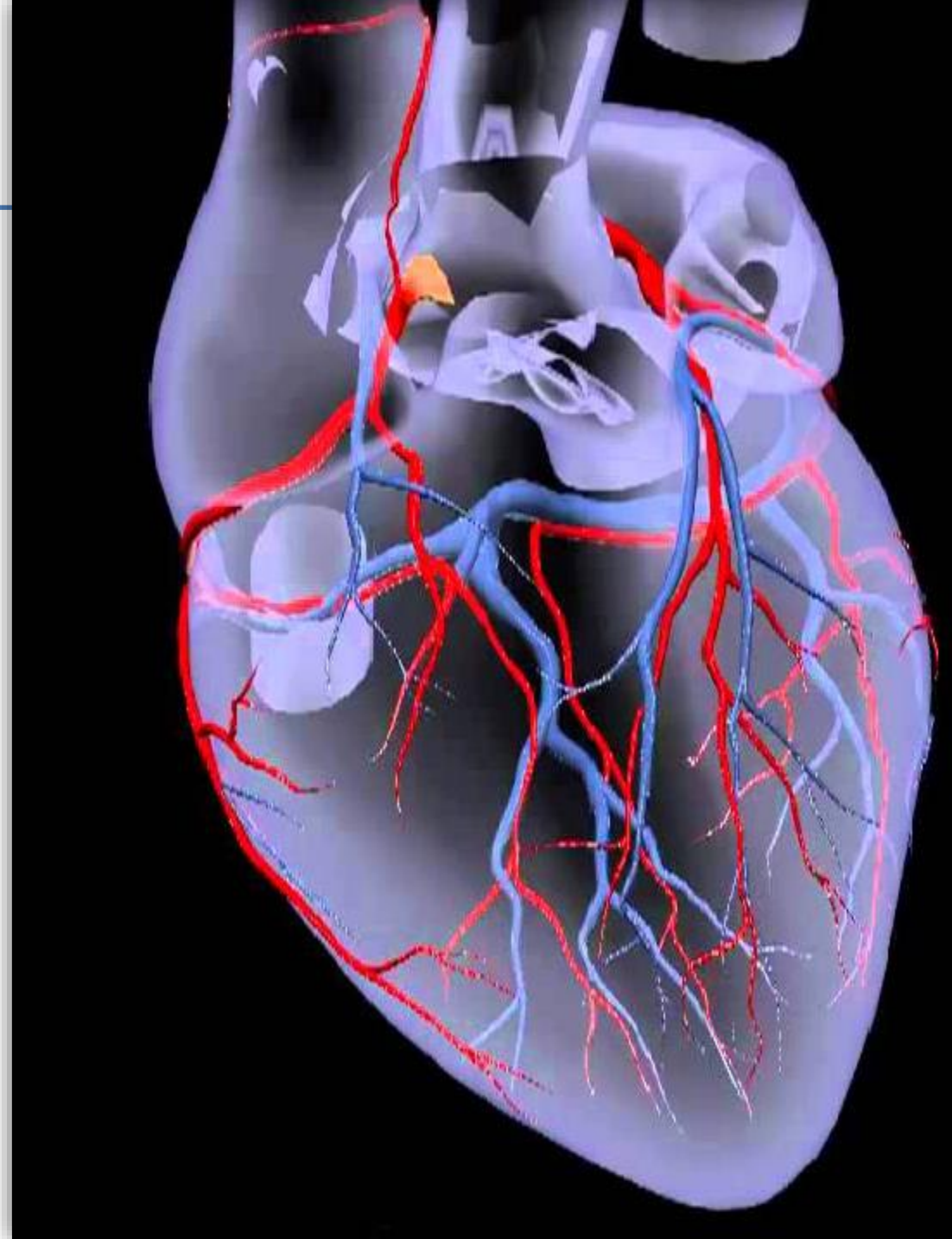
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Non dominant LCX  
without irregularities

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80% RCA stenosis after  
A.M. 1 60% R-PDA  
stenosis

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**a**



Revascularization  
for ACS is  
appropriate

**b**

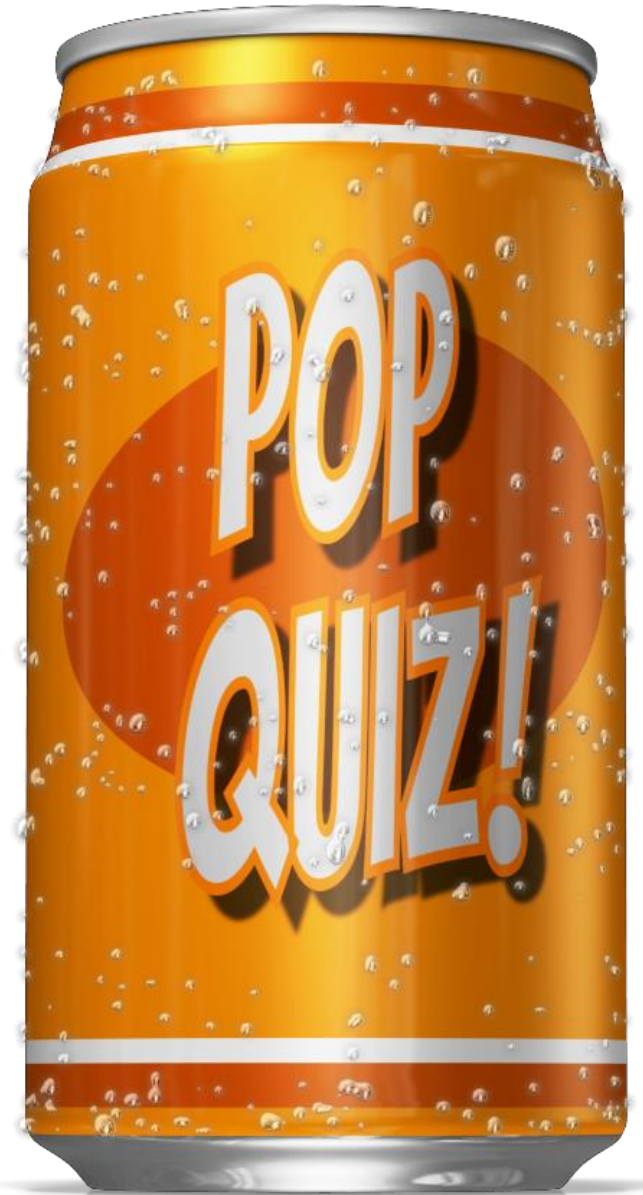


Stents or surgery  
associated with  
equal survival in non  
DM patients with  
multi-vessel disease  
and good LV function

**c**



For multi-vessel  
disease with  
non complex  
lesions, stents  
or surgery is  
equal in DM  
patients



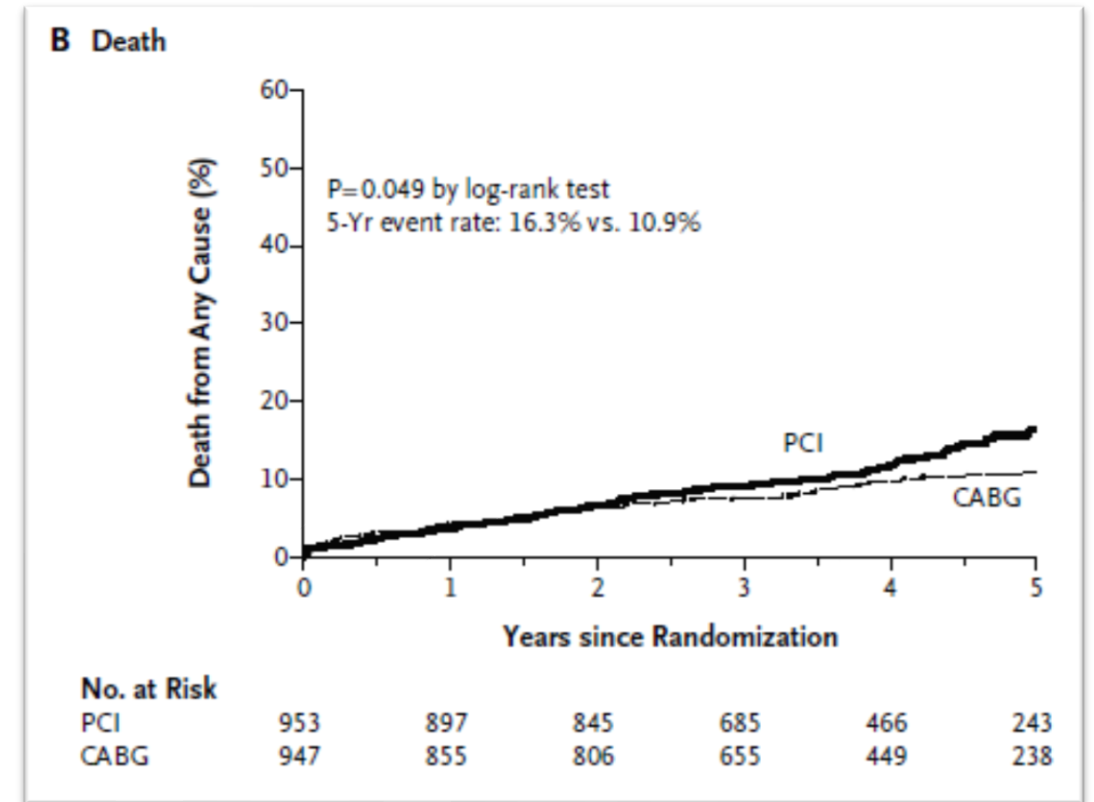
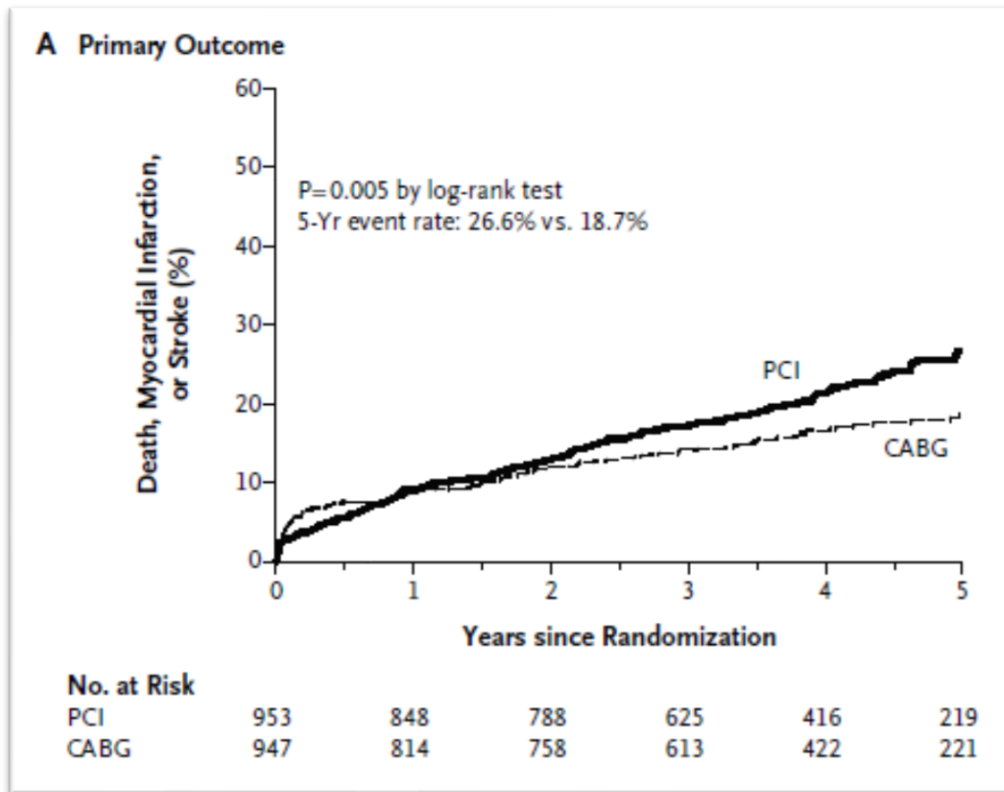
# Highest Syntax Scores

	Composite MACE			Cardiac Death			Repeat Revasc		
	CABG	PES	P	CABG	PES	P	CABG	PES	p
DM	12.2	32.4	0.003	5.4	24.3	0.001	4.1	13.5	0.004
Non DM	10.5	20.3	0.004	4.8	14.2	0.001	2.2	6.1	0.04

More complex anatomy, greater benefit to CABG

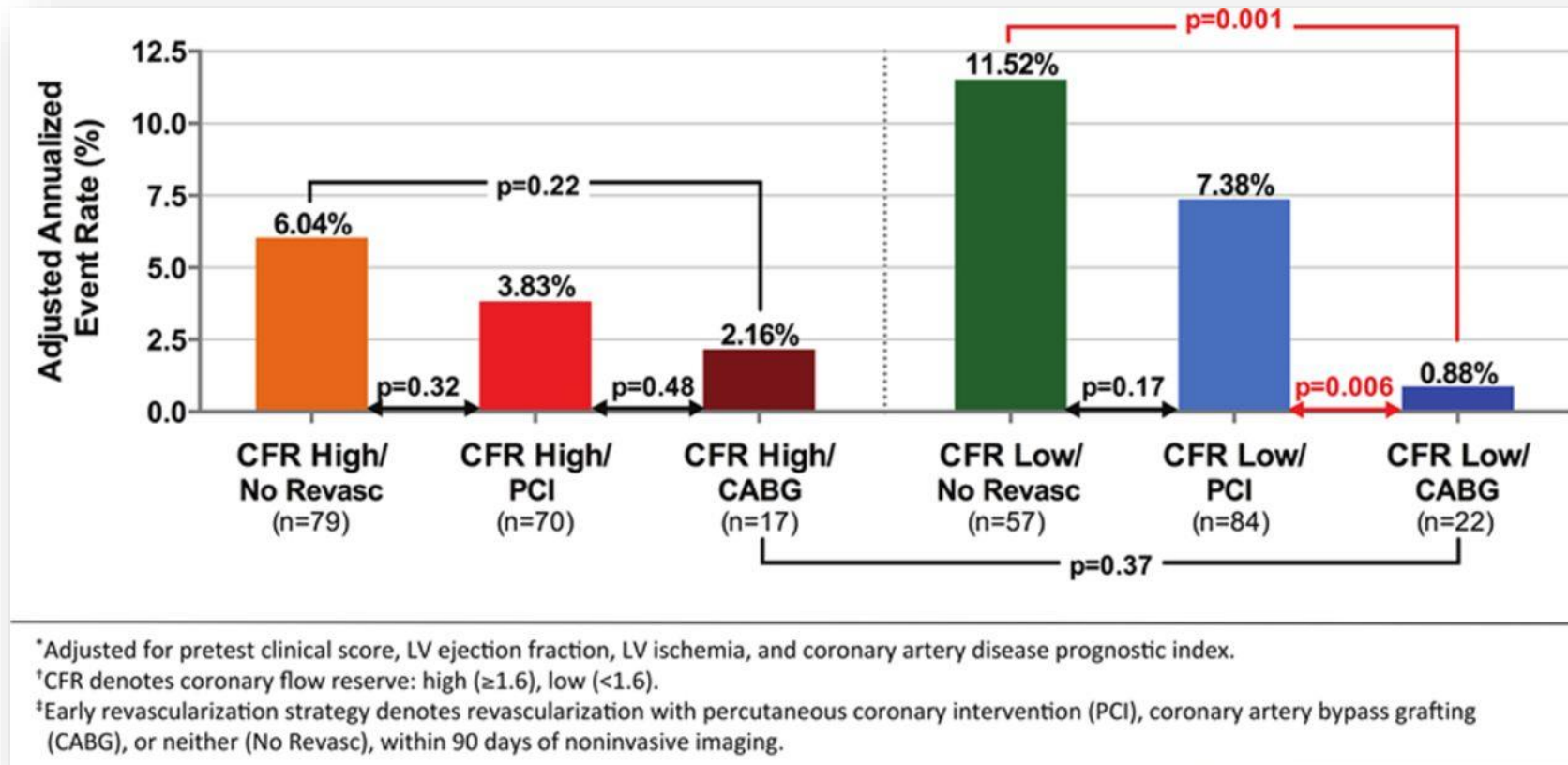
# FREEDOM Trial

**Future REvascularization Evaluation in Patients with Diabetes Mellitus: Optimal Management of Multivessel Disease**





Adjusted annualized rates of cardiovascular death and heart failure admission among patients referred for coronary angiography by coronary flow reserve (CFR) and early revascularization (Revasc) strategy (coronary artery bypass grafting [CABG], percutaneous coronary intervention [PCI], or neither).

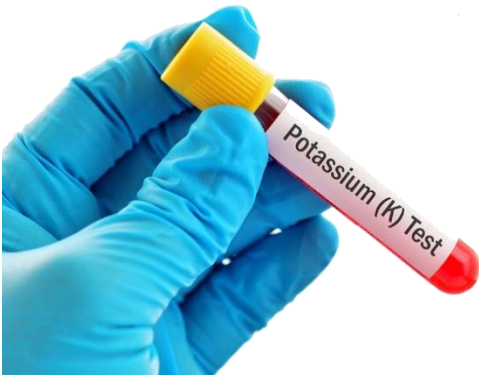


*Lower reserve needs more extensive/reliable revascularization*

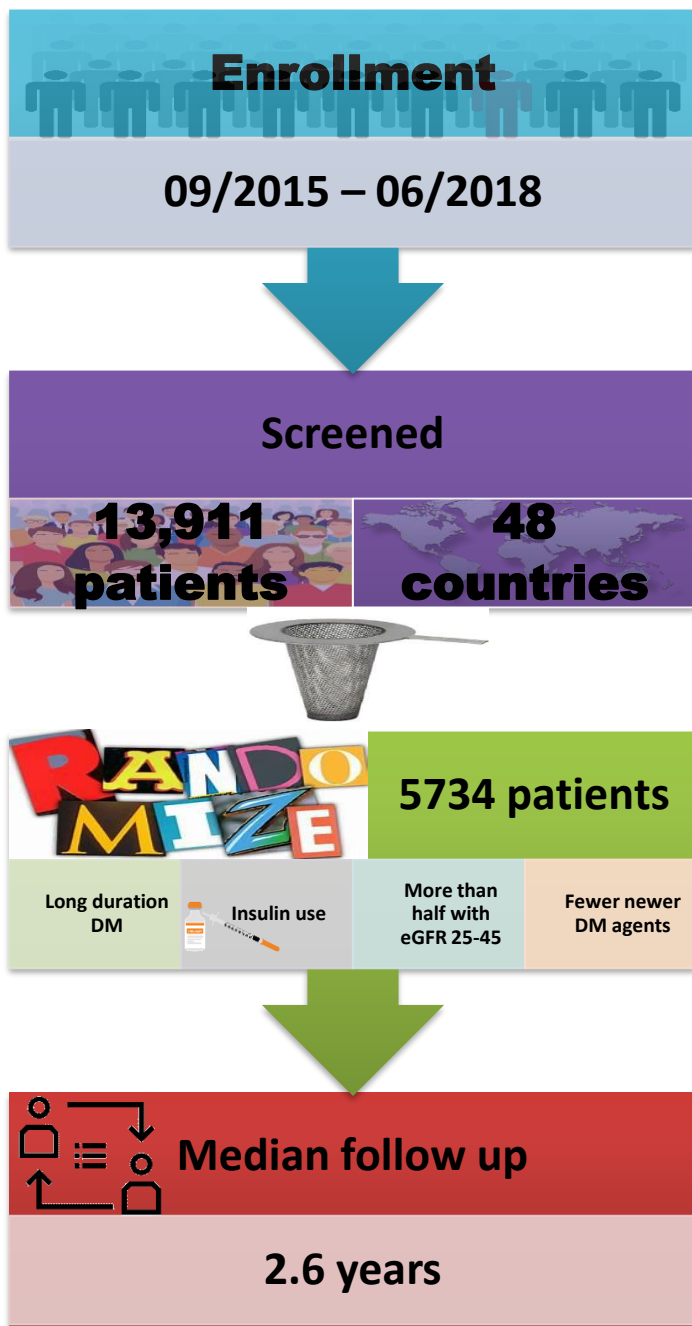
# **Finerenone**

## **Non-Steroidal Mineralocorticoid Receptor Antagonist**

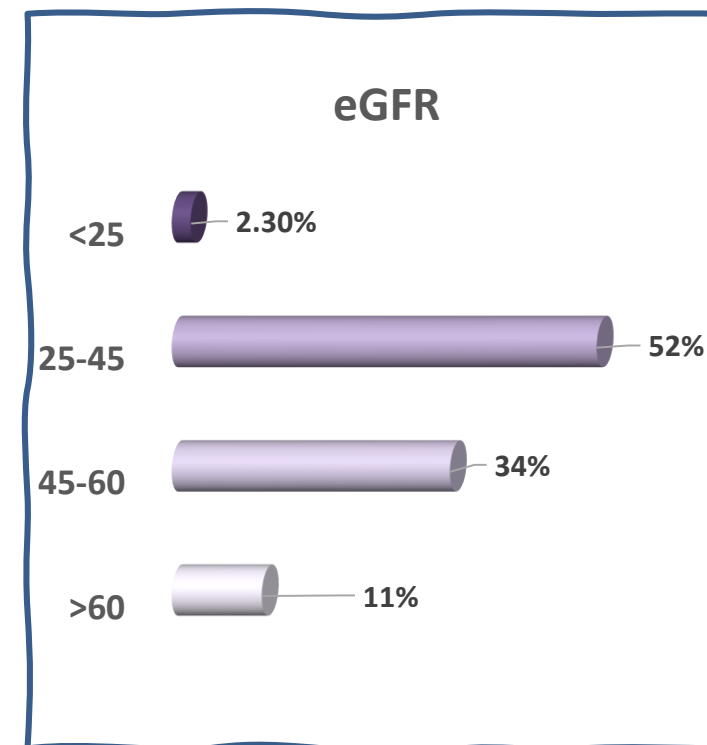
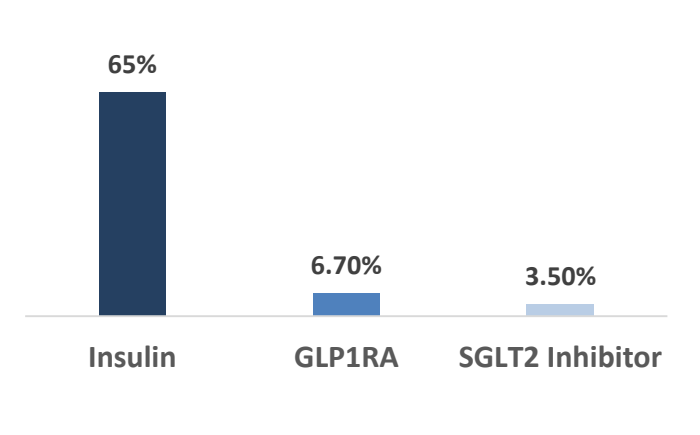
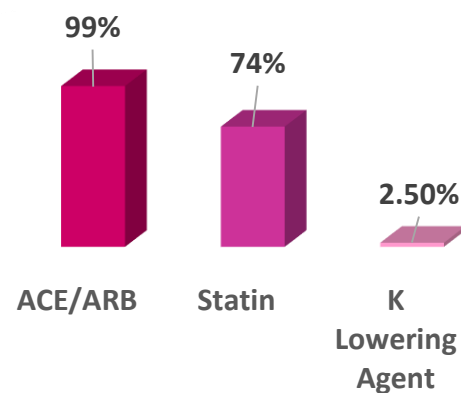
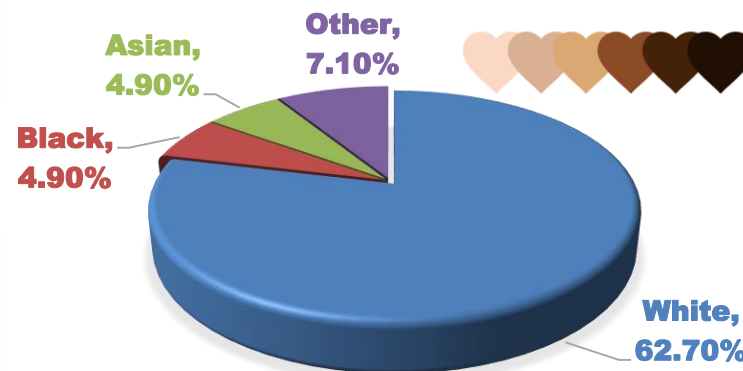
- Putative greater anti-inflammatory and anti-fibrotic effects than steroidal agents, spironolactone and eplerenone
- Less effect on potassium than spironolactone



# FIDELIO



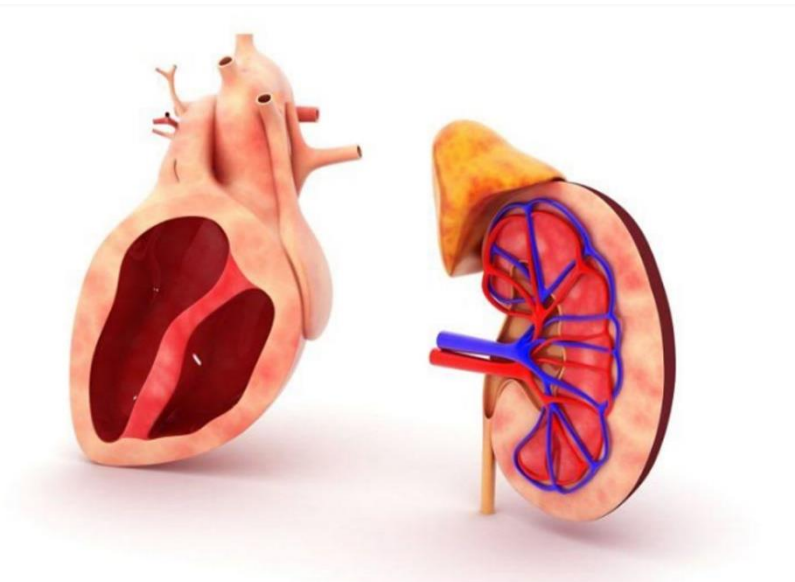
Age	65±8.9
Gender	69% male
DM Duration	16.6±8.8
A1c	7.7%
BP	138±14.3
Alb/Cr >300	87%



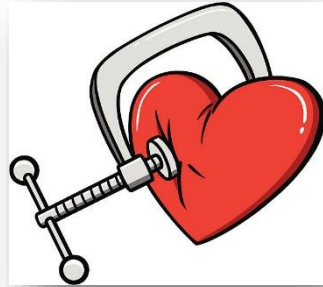


# FIDELIO-DKD

	Finerenone	Placebo	HR
Sustained >40% reduction in eGFR	17.8%	21.1%	0.82 (0.73-0.91)
CV Death, Non-Fatal MI, Stroke, Worsened HF	13%	14.8%	0.86 (0.75-0.99)
New onset AFIB/Flutter	3.2%	4.5%	0.71 (0.53-0.94)



# Case Study: MXXR - 75 Year Old Woman with Easily Precipitated Exertional Dyspnea



**Angina**  
>1 year



Crohns  
Disease:  
Stable



Impaired  
Glucose  
Tolerance

A1c 6.3%  
Treated with  
metformin



**Smoking**  
quit in 5<sup>th</sup> decade

Family History  
Father & paternal  
grandfather had MI's in 6<sup>th</sup>  
decade



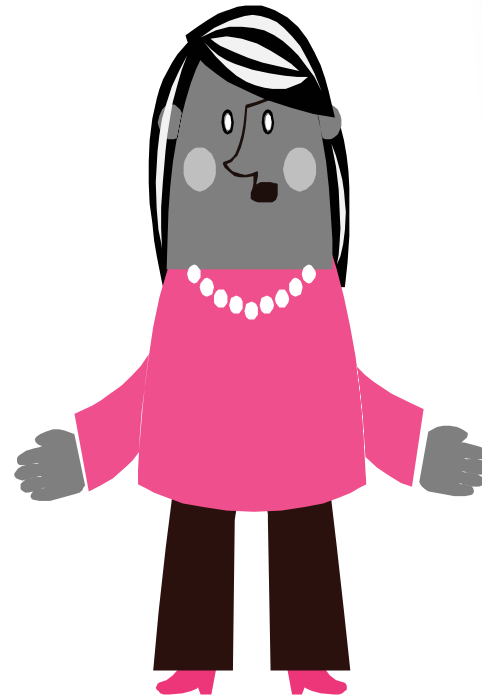
**Hypertension**  
Treated with Beta-blocker  
ACE Inhibitor



Cholesterol  
TC 180, HDL 70, LDL 92, TG 91  
Atorvastatin 40mg and  
ezetimibe 10 mg

## Exercise Study (outside institution)

- 7 minutes, 30 seconds
- Standard Bruce protocol
- Stops due to chest pain & SOB
- HR 118
- BP 164/70 mmHg
- 1.5mm horizontal ST segment depression (resolves after 3 minutes of recovery)
- Perfusion images normal
- EF 77%



## CTA (outside institution)

- 40-50% proximal LAD and LCx stenoses
- 30% mid RCA stenosis
- Mild cor calcifications

**Case Study: MXXR**



# Exam

127 pounds (trim)

BP 140/70 mmHg (both arms)

HR 58

RR 12

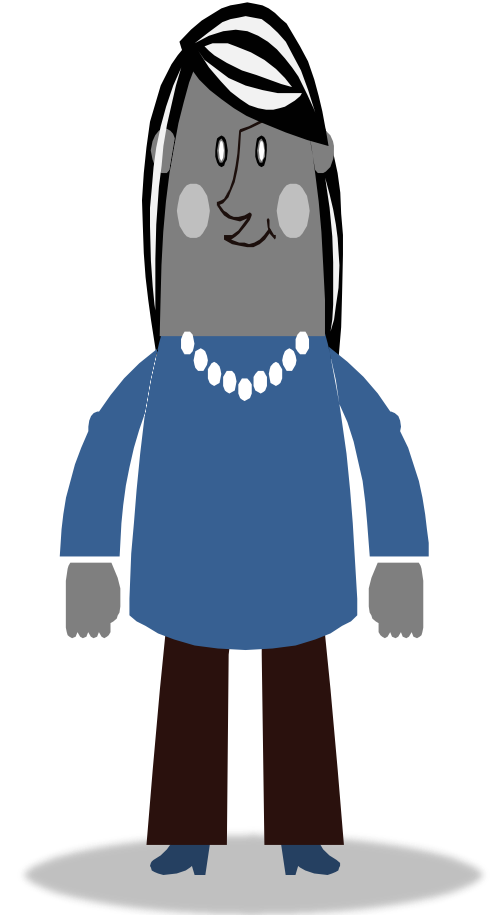
JVP not elevated (<8)

Carotid upstrokes & volumes normal

Clear lungs

PMI not displaced

Single  $S_1$ , physiologically split  $S_2$  with mid systolic click at apex that moves earlier in cardiac cycle with Valsalva



## Case Study MXXR

**Which of the statements are true?**



**a**

The story & studies are consistent with important epicardial coronary disease, and if symptoms cannot be managed, coronary angiography is indicated

**c**

The story & studies are consistent with microvascular angina with an outstanding prognosis and her medical regimen can be reduced.

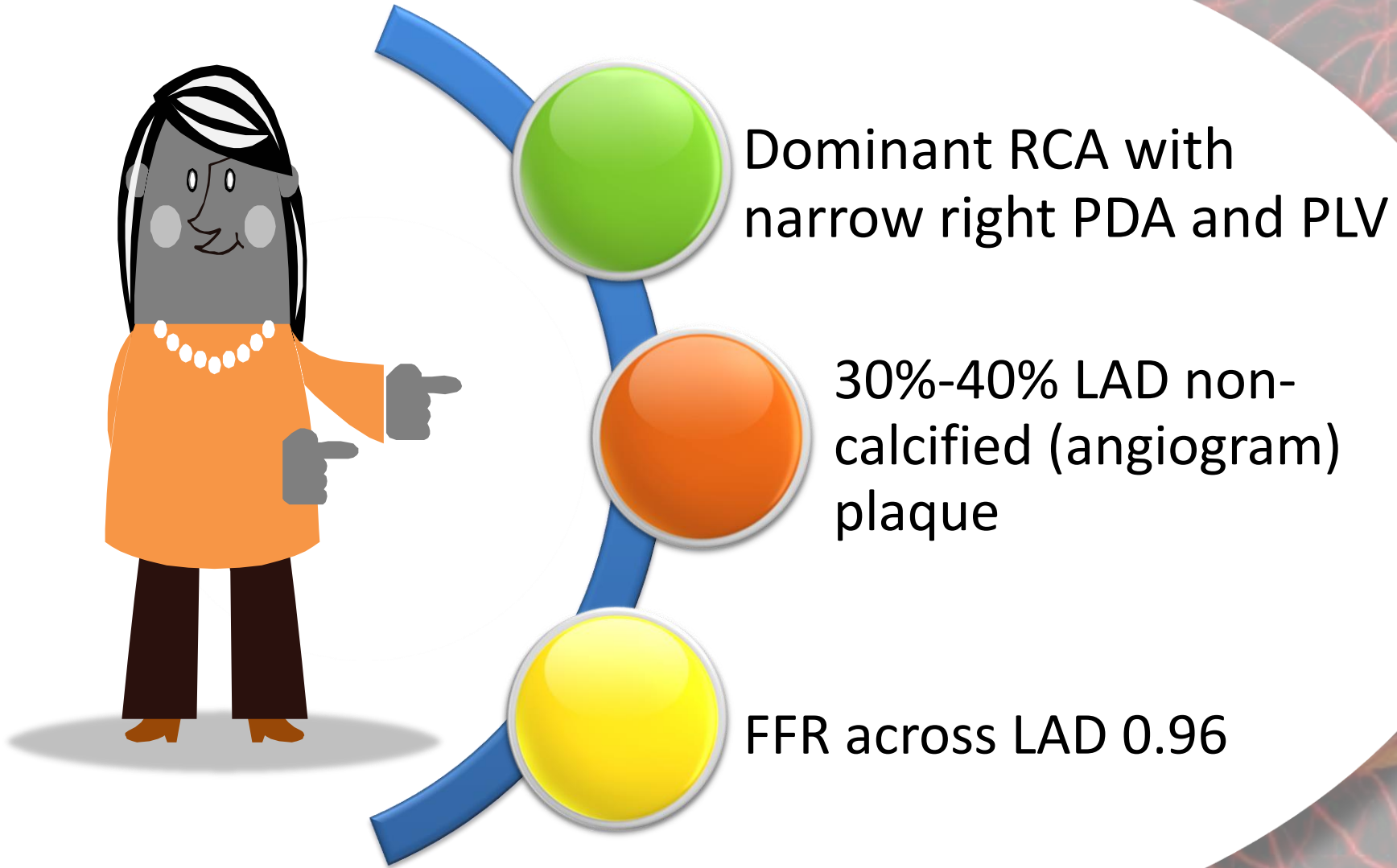
**b**

The story is consistent with mitral valve prolapse.

**d**

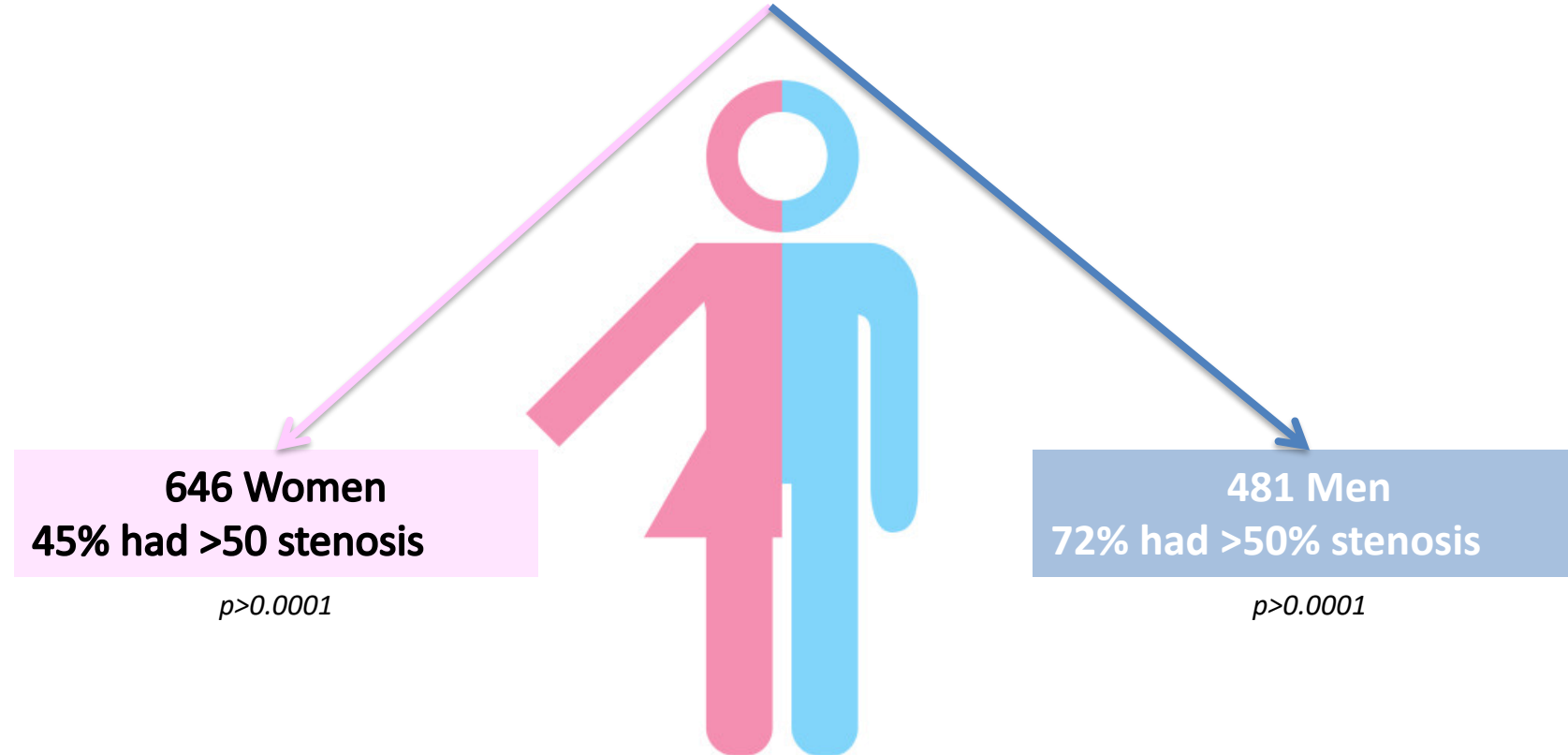
The story is consistent with the non-GI effects of Crohn's Disease.

**Case Study: MXXR**



# Sex Differences in CAD Outcomes

CONSECUTIVE CTA ON PATIENTS WITH ANGINA, SOB, NAUSEA, FATIGUE AND ABNORMAL EXERCISE TESTS

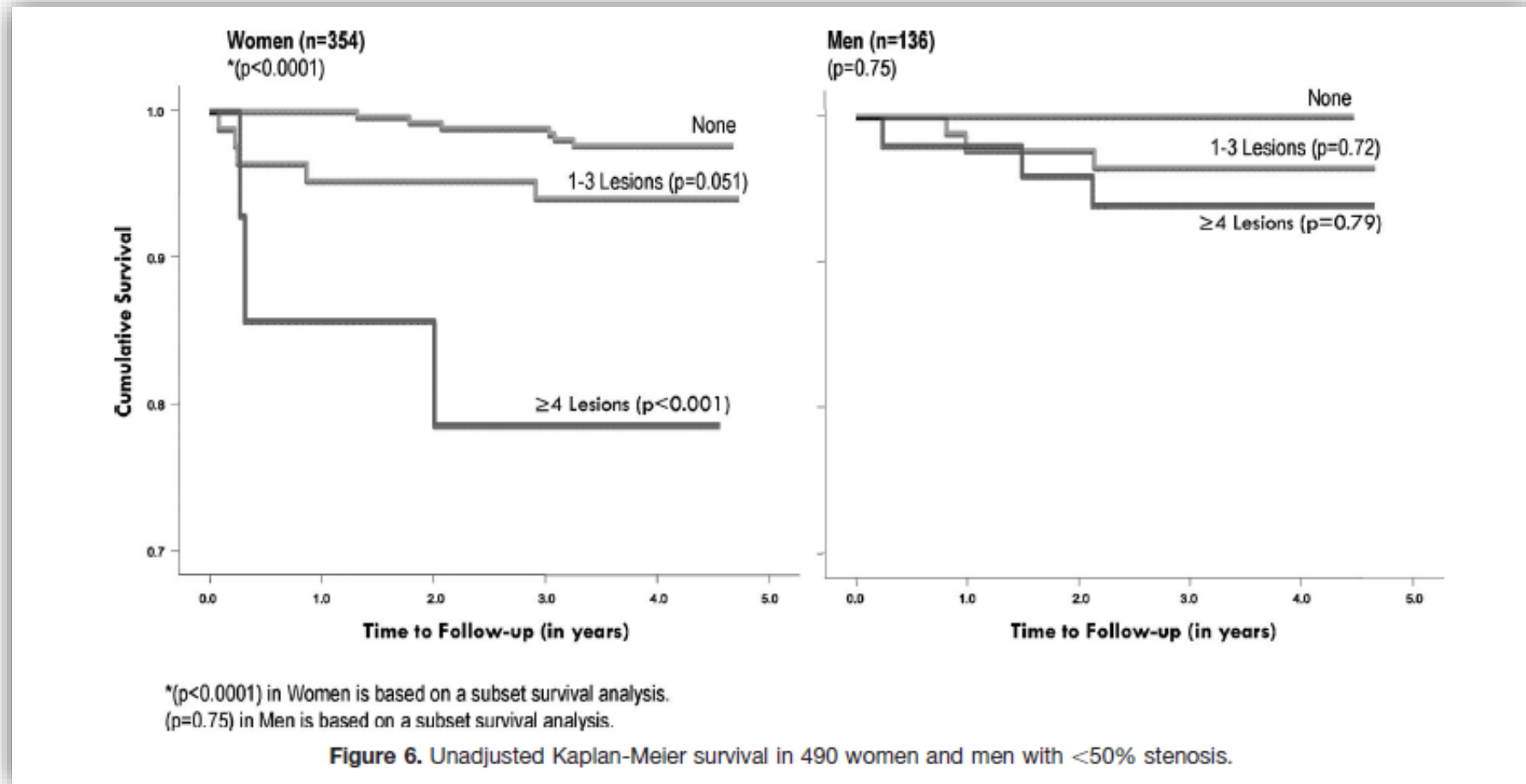


WOMEN WITH STABLE CHEST PAIN HAVE **56%** LOWER RISK ADJUSTED  
LIKELIHOOD OF MORE ADVANCED STENOSES, BUT

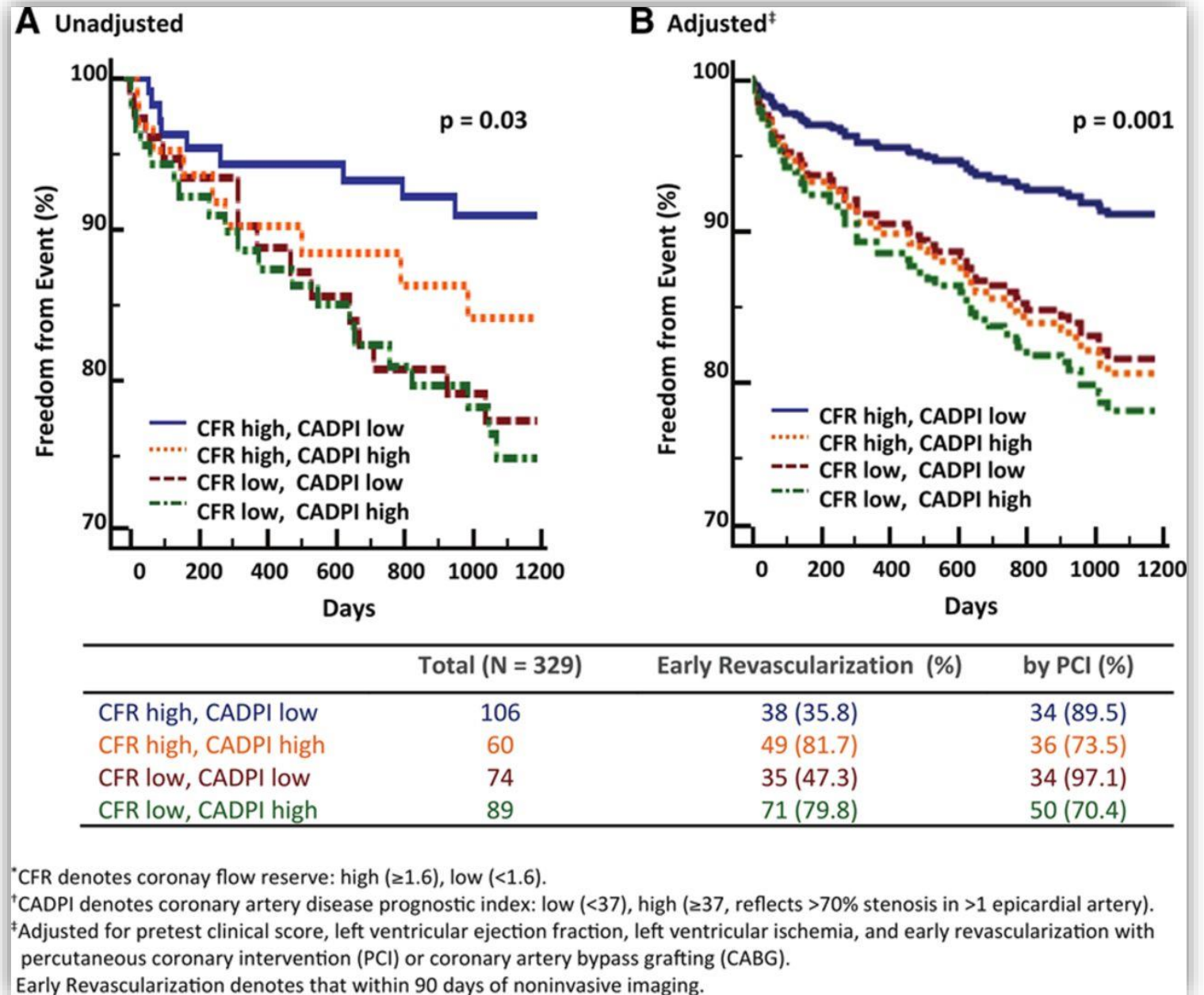


# Sex Differences in CAD Outcomes

Non-obstructive CAD warrants attention in all, and especially in women.



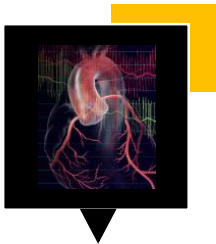
Freedom From Cardiovascular  
Death Or Heart Failure Admission  
**According** To Coronary Flow  
Reserve (CFR) And Angiographic  
Score (Coronary Artery Disease  
Prognostic Index [CADPI]).



# Evaluation of Coronary Artery Stenoses

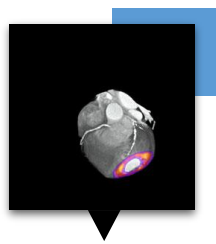
Anatomic stenoses is important, especially with severe stenoses, >90%

Otherwise, anatomic + functional parameters are helpful



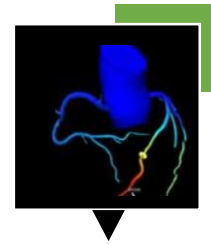
## FFR, IFR in Cath Lab

Associated with events (related to platelet aggregation)



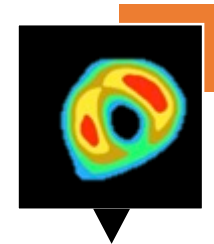
## PET Hyperemic Blood Flow and Flow Reserve

Hyperemic flow  $\div$  resting flow  
Related to overall reserve



## CT FFR

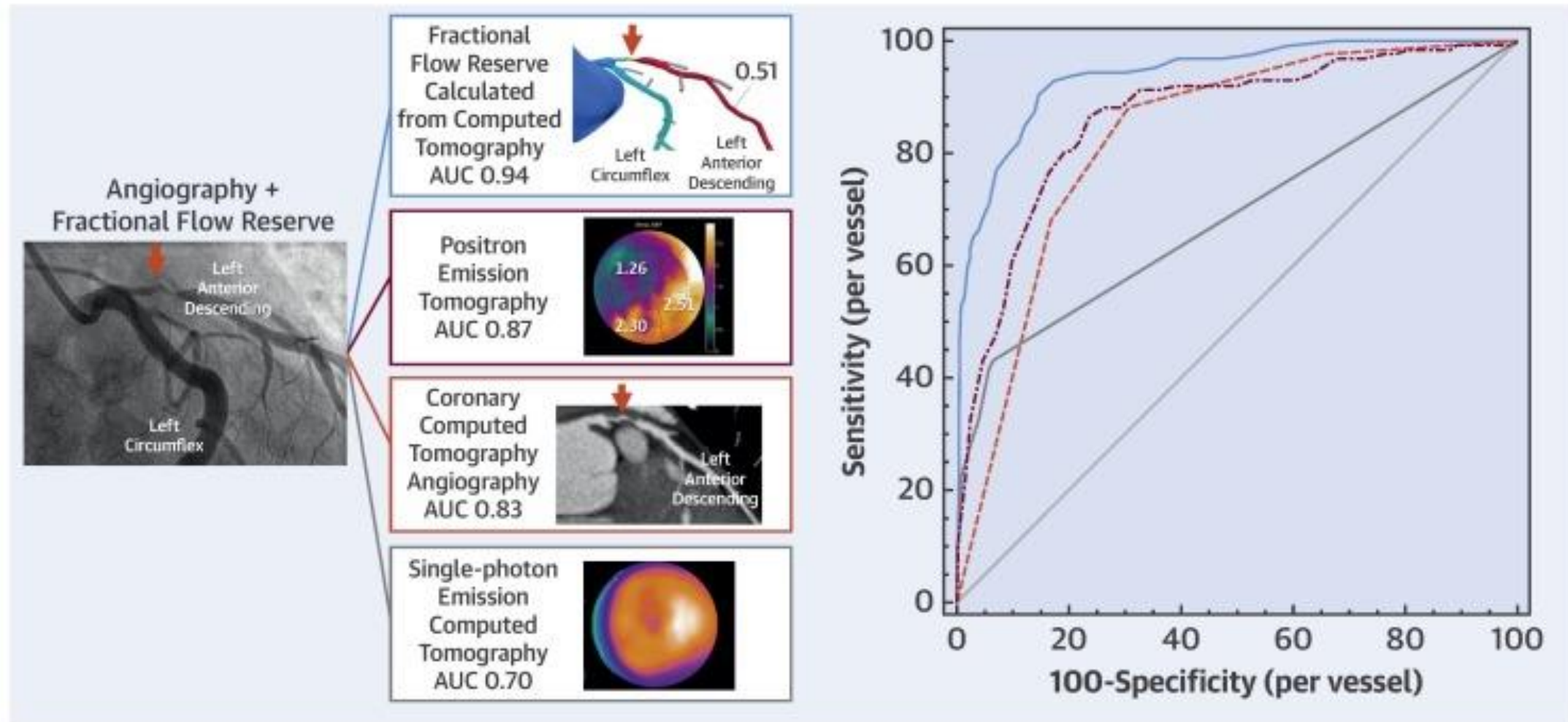
Similar to cath FFR, IFT in principle  
Can only do  $\sim$  75% of CTA  
Cannot do with stents  
Calcium score >500



## Perfusion Images

Wall motion with exercise

## CENTRAL ILLUSTRATION: Discriminative Ability of Imaging Modalities for the Detection of Per-Vessel Fractional Flow Reserve-Defined Ischemia



Driessen, R.S. et al. J Am Coll Cardiol. 2019;73(2):161-73.



## Case Study:

## 68 YO Woman

### Presentation

Few weeks of intermittent chest pain  
Can last a few hours, sometimes with exertion and radiates to left arm

### 4 years prior

Completed 6 cycles R-CHOP  
for Stage 1A large B-cell  
lymphoma (neck mass)



### Dyslipidemia

TC 325, HDL 82-113,  
LDL 163-191, TG 58-123  
and statin stopped due to ↑LFT's

### Family History



Father had MI in 7<sup>th</sup> decade.

Mother underwent carotid endarterectomy in 7<sup>th</sup> decade

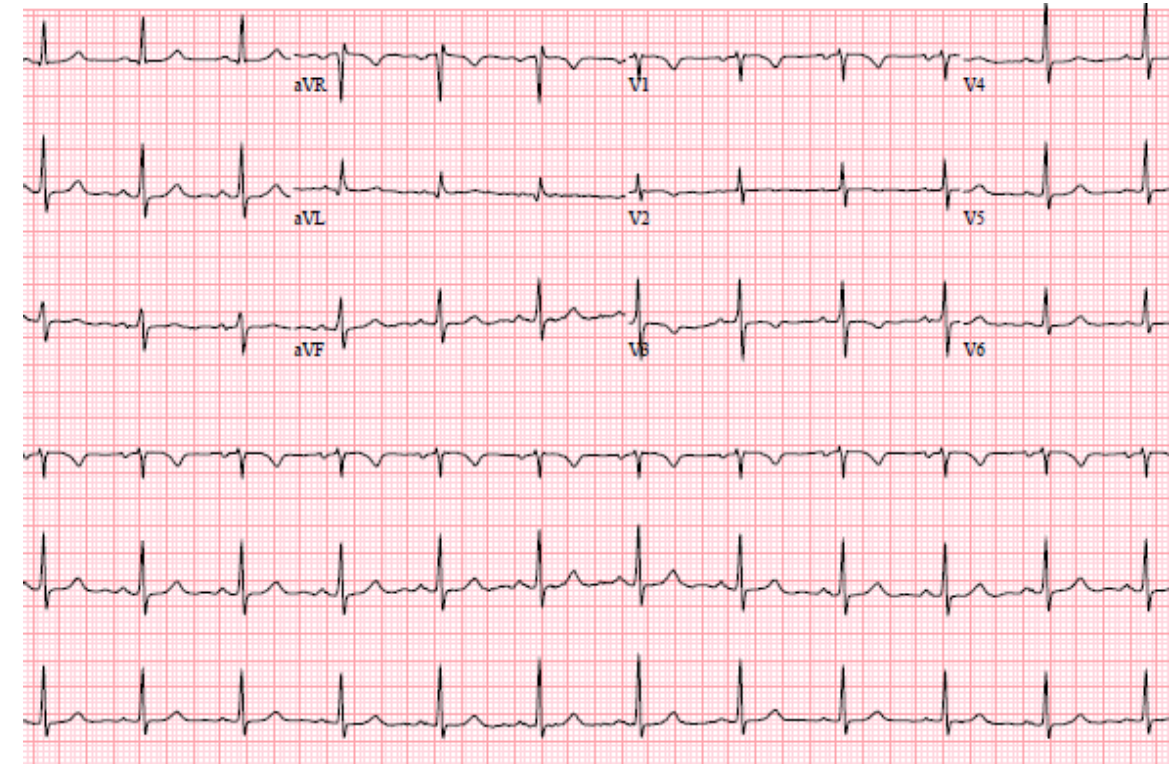
### Cardiac Markers

Tr-hs 21-22 (female <9 mg/dl)

### Chest CT

When staged for lymphoma, Chest CT showed mild LAD calcification

# Case Study: 68 YO Woman- EKGs





# Case Study: 68 YO Woman

Resting perfusion study: normal  
but due to chest pain, not  
exercised

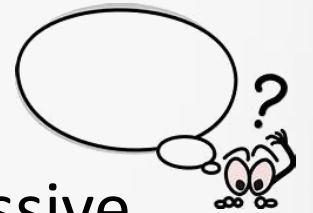
CT angiography shows 3 vessel,  
partly calcified plaques 1-24%, no  
severe stenoses





## Which of the Following Statements are True

- 1 She can be reassured. Not in midst of a large MI.
- 2 She can be told that her symptoms are not ischemic and are unrelated to coronary artery disease
- 3 Since she is 68 yo and has had only modest plaques and statins were associated with LFT abnormalities, there is no reason to be aggressive with her lipids
- 4 Her chest pain, Troponin elevations, and EKG changes need no more investigation and fall under broad category MINOCA



**nope**



# Myocardial Infarction with Non-Obstructive Disease: MI Symptoms



R/O

- sepsis, pulmonary embolism, cardiac contusion, HT, HF, tachycardia, renal failure, vasospastic agents, overlooked obstruction of a distal vessel or branch, overt myocarditis, overt takotsubo, lesions with FFR <0.8



Wonder about

- coronary embolism
- dissection
- spasm
- takotsubos
- thrombophilia
- myocarditis
- microvascular disease
- and if there are any coronary artery irregularities, plaque rupture or erosion (demonstrated with OCT)

Important to ascertain etiology because 5-year mortality rate may approach 11%, MACE ~25%

# MINOCA Diagnosed in 5-15% of Acute MI Cases



**204** consecutive patients  
Tr positive ACS  
Unobstructed coronaries  
Unclear diagnosis



Younger than usual  
acute MI population

**Age 56±17**

Higher % of ♀ than  
usual, also fewer CAD  
risks

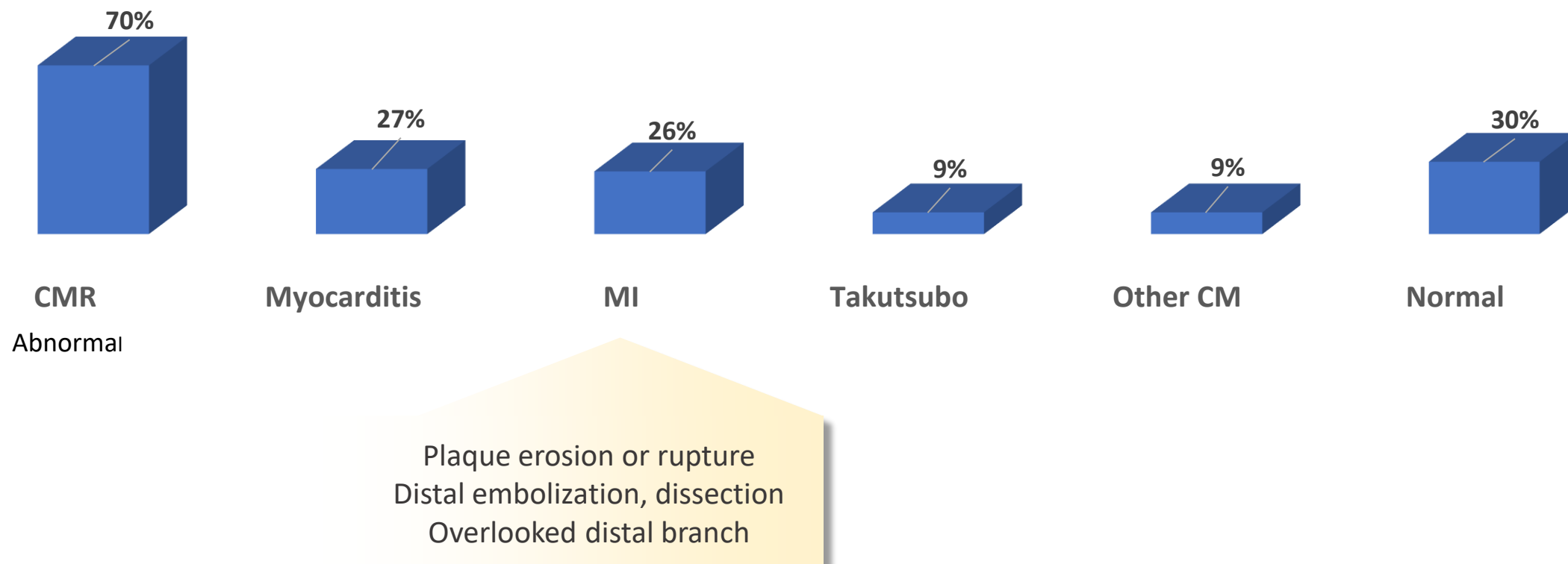
**51%**



**19% ST  
Segment  
Elevation**

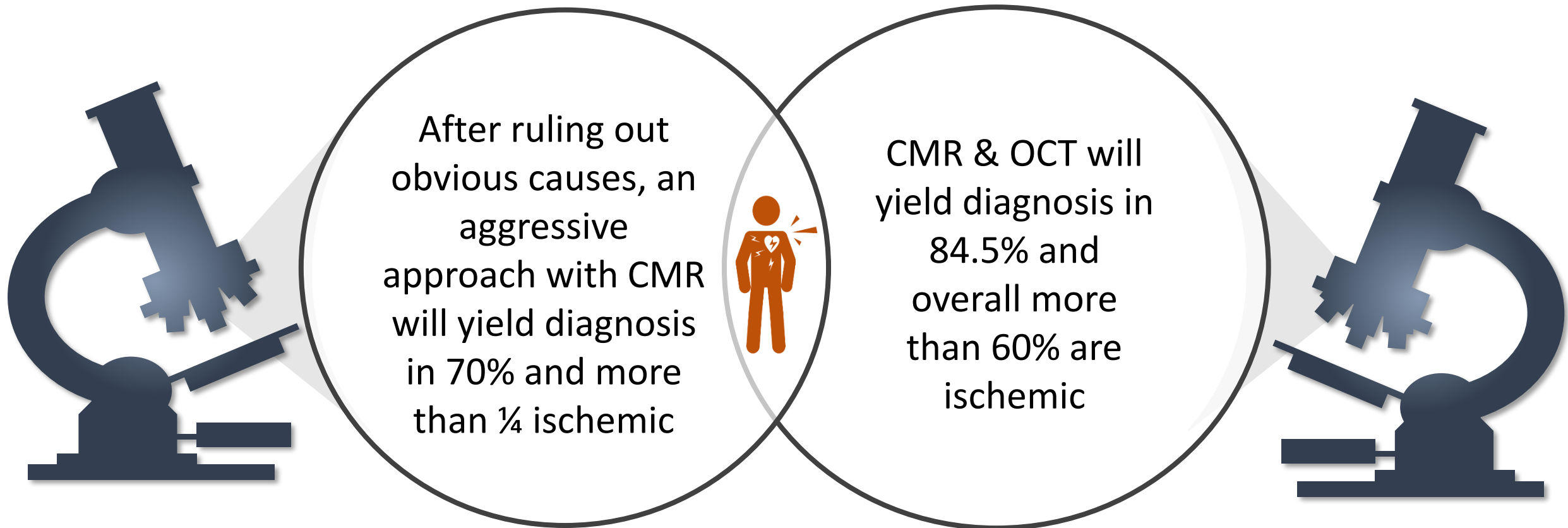
**Tr-hs 640  
ng/l (nl<14)**

# MINOCA: CMR Diagnosis in 143/204



Diagnostic Yield	84% for MRI < 2 weeks; better for myocarditis, takotsubo 57% for MRI > 2weeks
Change in Mgmt	41%

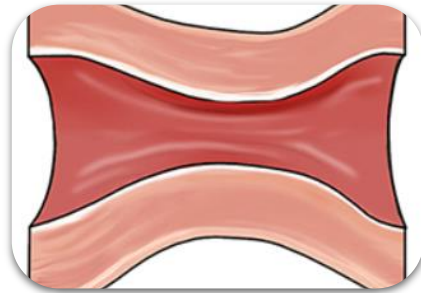
# Myocardial Infarction with Non-Obstructive (<50%) Coronary Arteries



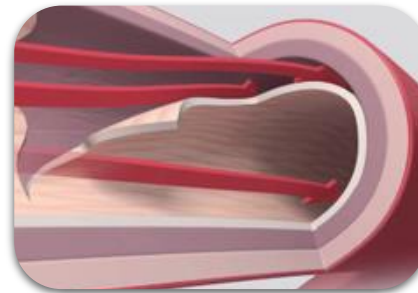


# Once a Minoca, always a Minoca? (Myocardial Infarction with non-Obstructive CAD)

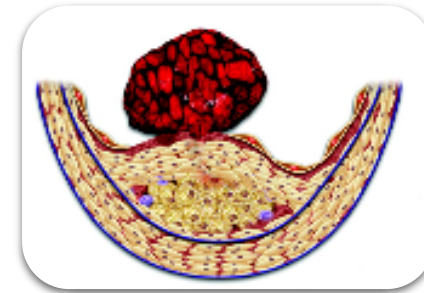
What is coronary anatomy in patients who have MINOCA, then a recurrent infarction?



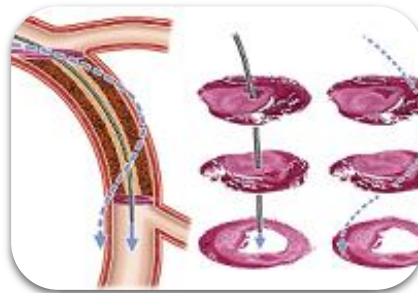
Spasm



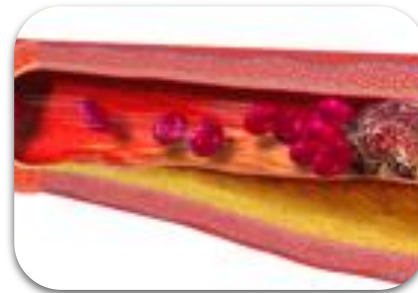
Dissection



Plaque Erosion



Recanalization



Embolism



Microvascular  
Disease

### 9,092 unique MINOCA patients

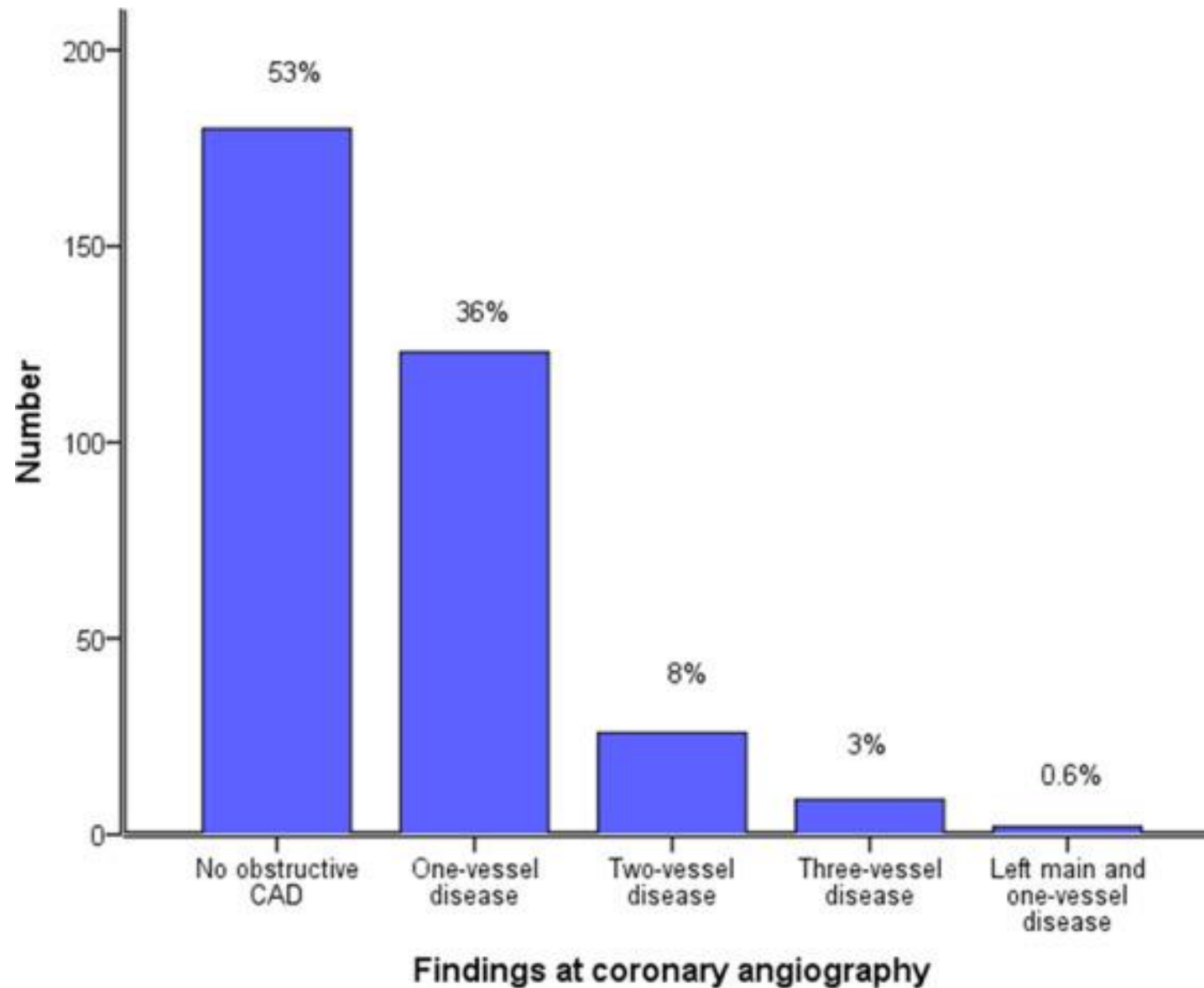
- 199,163 acute MI admissions
- 2003-2013
- 570 patients of original MINOCA (6.3%) readmitted with infarct
  - Median time 17 months (5-39 IQR)

### Re Infarct Patients

- Slightly older 67 vs 65 (p 0.001)
- More DM 16 vs 10 (p < .001)
- Slightly worse renal function
- Slightly worse EF
- On similar meds

*“Surprise” 340/570 underwent Cath.  
47% had obstructive disease.*

- Disease means > 50% stenoses.
- 340/570 cathed for re infarction post prior minoca.
- **NOTE- 230/570 PTS NOT CATHED**



# Case Study: OX-500



**61 year old man**



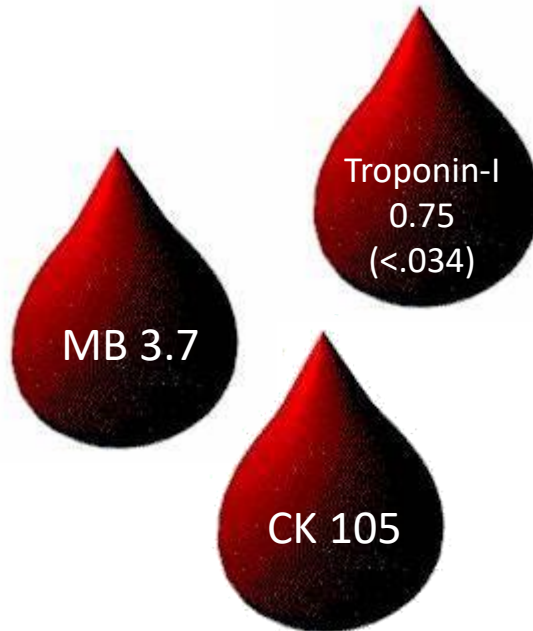
**Sudden onset SOB**

**3 days post left total knee replacement**



## History

- ✓ Hypertension
- ✓ Dyslipidemia
- ✓ Previous radical prostatectomy
- ✓ Traumatic splenectomy



- Anxious & diaphoretic
- BP 108/60 mmHg
- HR 98
- RR 20
- Pulse Ox 79%



- ⚡ JVP Carotid upstrokes & volumes normal
- ⚡ Crackles at left lung base
- ⚡ PMI not displaced
- ⚡ No sternal lift
- ⚡ Normal S1, physiologically split S2, P2 is not increased
- ⚡ Left leg swollen, above & below knee not elevated

# Case Study: OX-500 - EKG





# Case Study: OX-500



**A**

This man, with multiple CAD risk factors, is most likely having an acute coronary syndrome.

***He should be heparinized and he should have coronary angiography.***

**B**

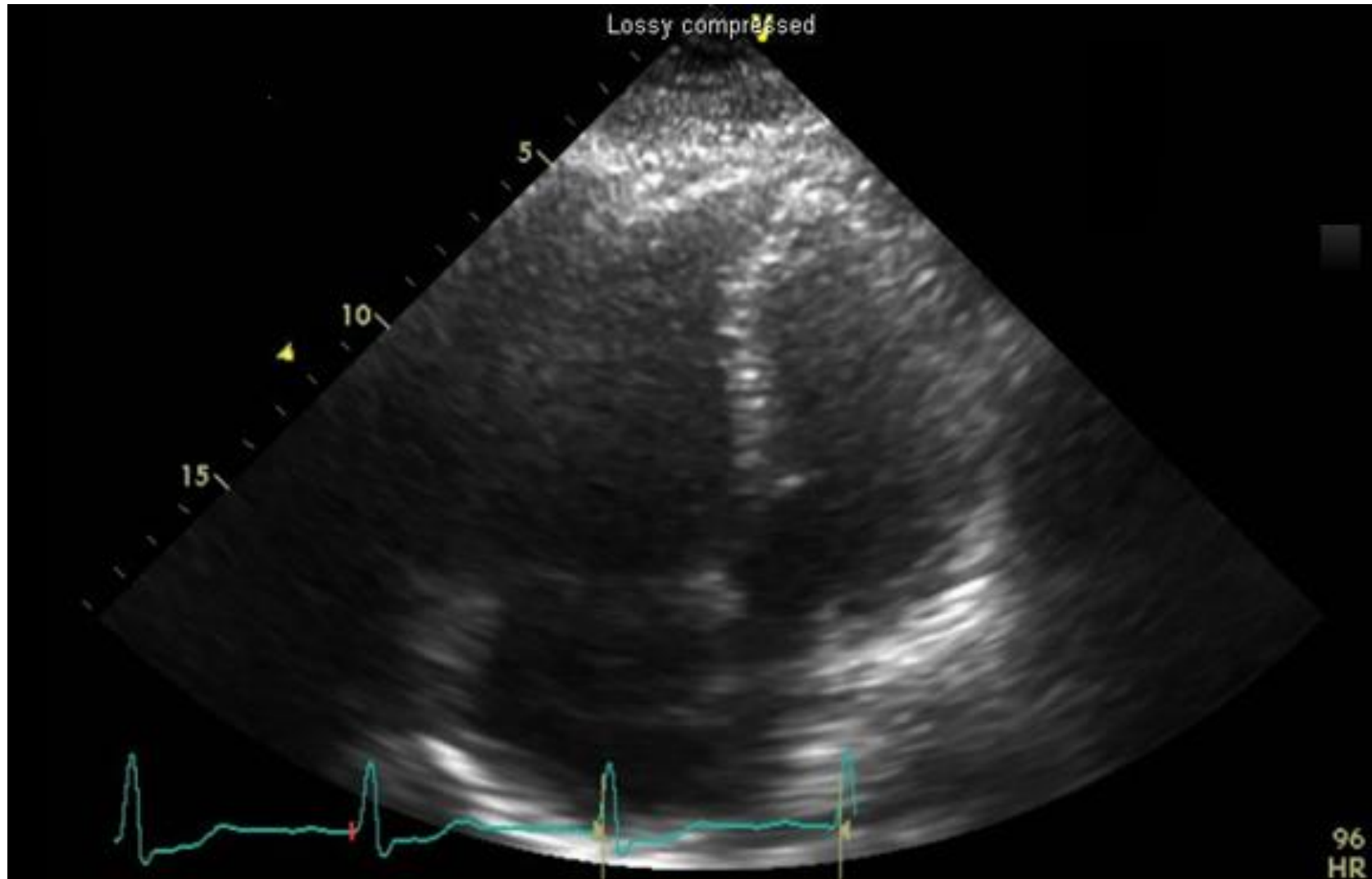
This man, s/p orthopedic surgery, is most likely struggling with a pulmonary embolism.

***He should be heparinized and have an echocardiogram.***

**C**

***This man should receive lytic therapy immediately which will be helpful in either circumstance***

# Case Study: OX500 - Echo



Dilated RV with paradoxical septum

# Case Study: OX500 - CT



# Case Study: OX500

## THE NEXT STEP



**IVC Filter &  
Catheter or  
Surgical  
Embolectomy  
and ongoing  
heparinization**



**IVC Filter and  
ongoing  
heparinization**



**IVC Filter and  
Lytic Agents**



**Lytic Agents  
given size of  
PE**

# Case Study: Ox500 - Outcomes

Before
HR 115
BP 100/70
O2 Sat: 94%, 100% non-rebreather
PA: 50/20
Angiogram: Like CT

## Procedure

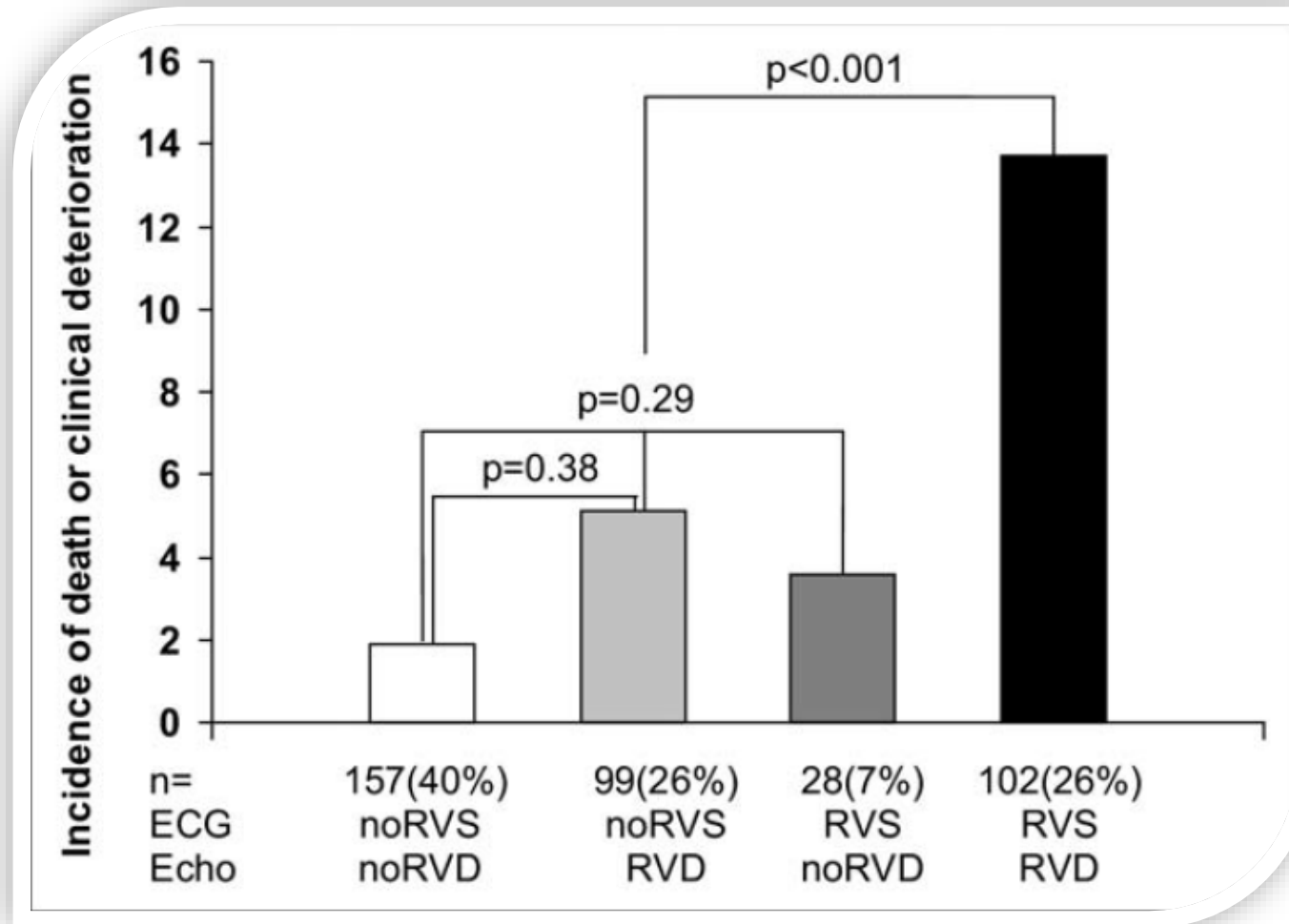


Suction embolectomy on right side; then placement of retrievable IVC filter

After
HR 87
BP 130/80
O2 Sat: 98% on 40% face mask
PA 40/



# ECG & Pulmonary Embolism Prognosis



# Use of ECMO and Surgical Embolectomy

*Patients with massive and sub-massive PE*

## MASSIVE

- **BP < 90 for > 15 minutes or use of pressors**
- **AND**
- **RV dysfunction (RV/LV > 0.9 on TTE or elevated NTProBNP or elevated Tr)**

## Sub-massive: RV dysfunction with hemodynamic stability

### Intermediate High Risk

- RV dysfunction
- Elevated Tr or
- NTProBNP, and tachycardia or mild hypotension

### Intermediate Low Risk

- RV dysfunction without increased Tr or NTProBNP
- And without increased HR or mildly decreased BP

---

Almost all the  
SMPE Underwent  
Embolectomy

---

40% of Massive  
started with  
ECMO

	All (N = 136)	SMPE (n = 92 [67.6%])	MPE (n = 44 [32.4%])	p Value
Primary embolectomy	117 (86.0)	91 (98.9)	26 (59.1)	<0.0001
ECMO after embolectomy	3 (2.2)	0 (0.0)	3 (6.8)	0.01
ECMO days	4 (2–5)	0	4 (2–5)	<0.001
Primary ECMO	19 (14.0)	1 (1.1)	18 (40.9)	<0.001
ECMO days	5 (1–10)	7	5 (1–10)	0.77
Embolectomy after ECMO	3 (2.2)	0 (0.0)	3 (17.6)	0.011

# RESULTS of Overall Surgical (ECMO/Embolectomy)

Mortality

6/136 = 4.4%

Ventilation  
> 72 Hours

18%

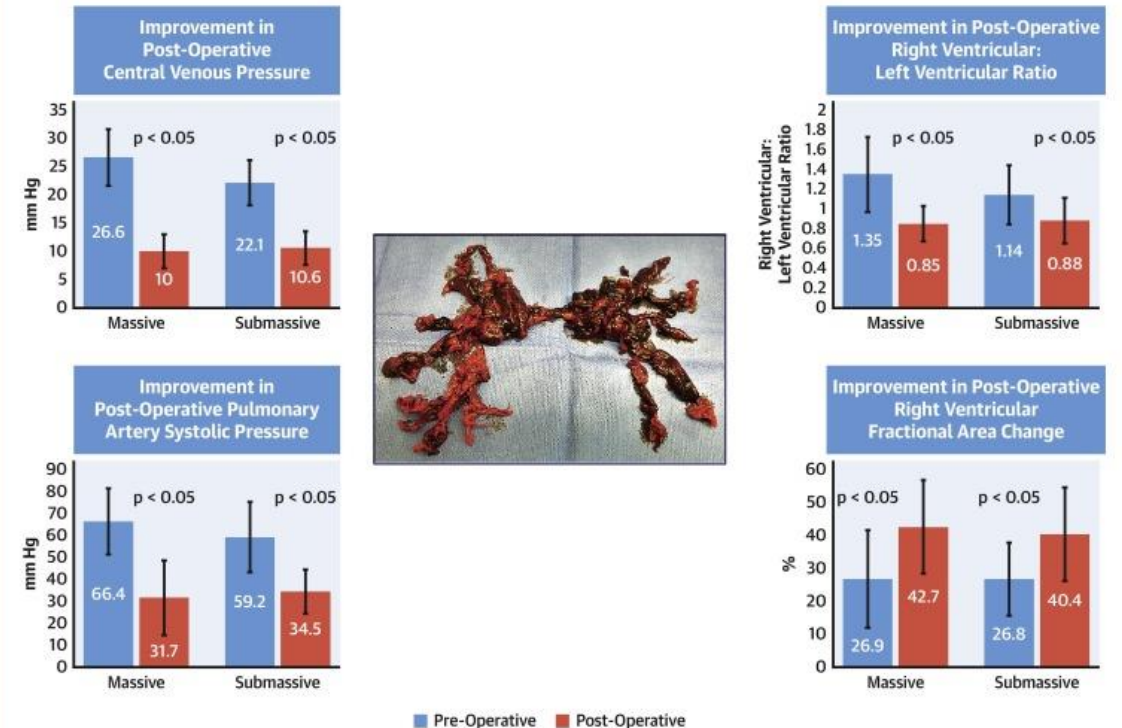
HIT

15%

LOS

13 (4-92)

## CENTRAL ILLUSTRATION: Improvement in Right Ventricular Function After Surgical Management of Acute Pulmonary Embolism

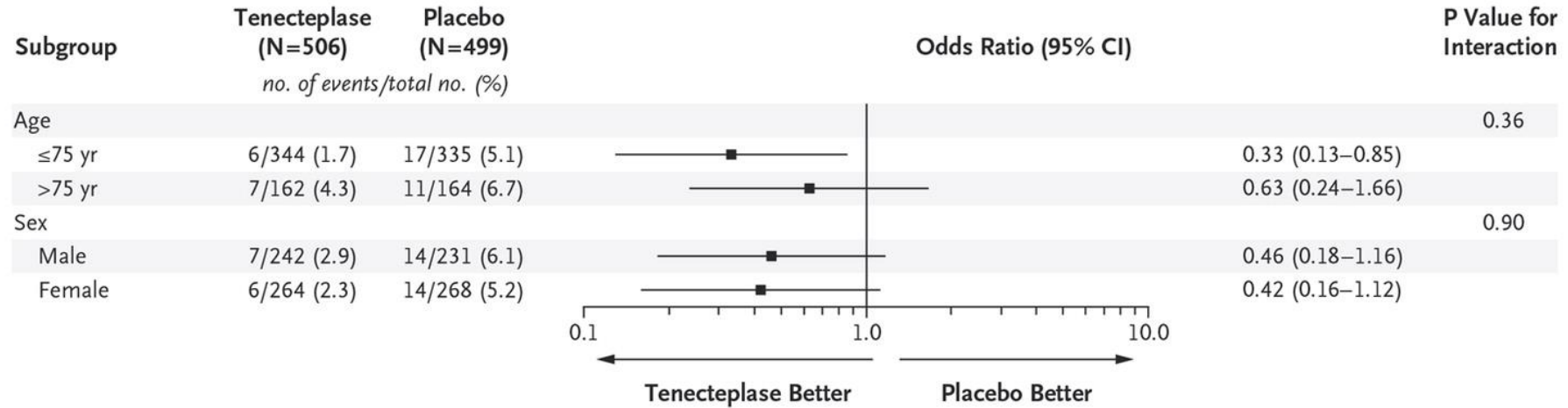


Goldberg, J.B. et al. J Am Coll Cardiol. 2020;76(8):903-11.

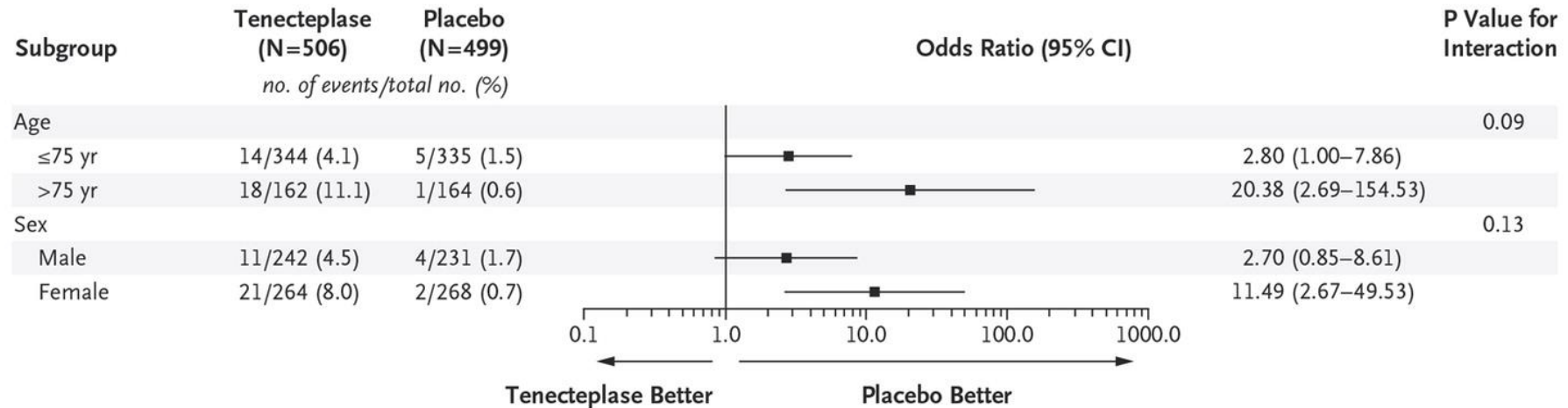


# Pictorial View of PEITHO Results

## A Death or Hemodynamic Decompensation

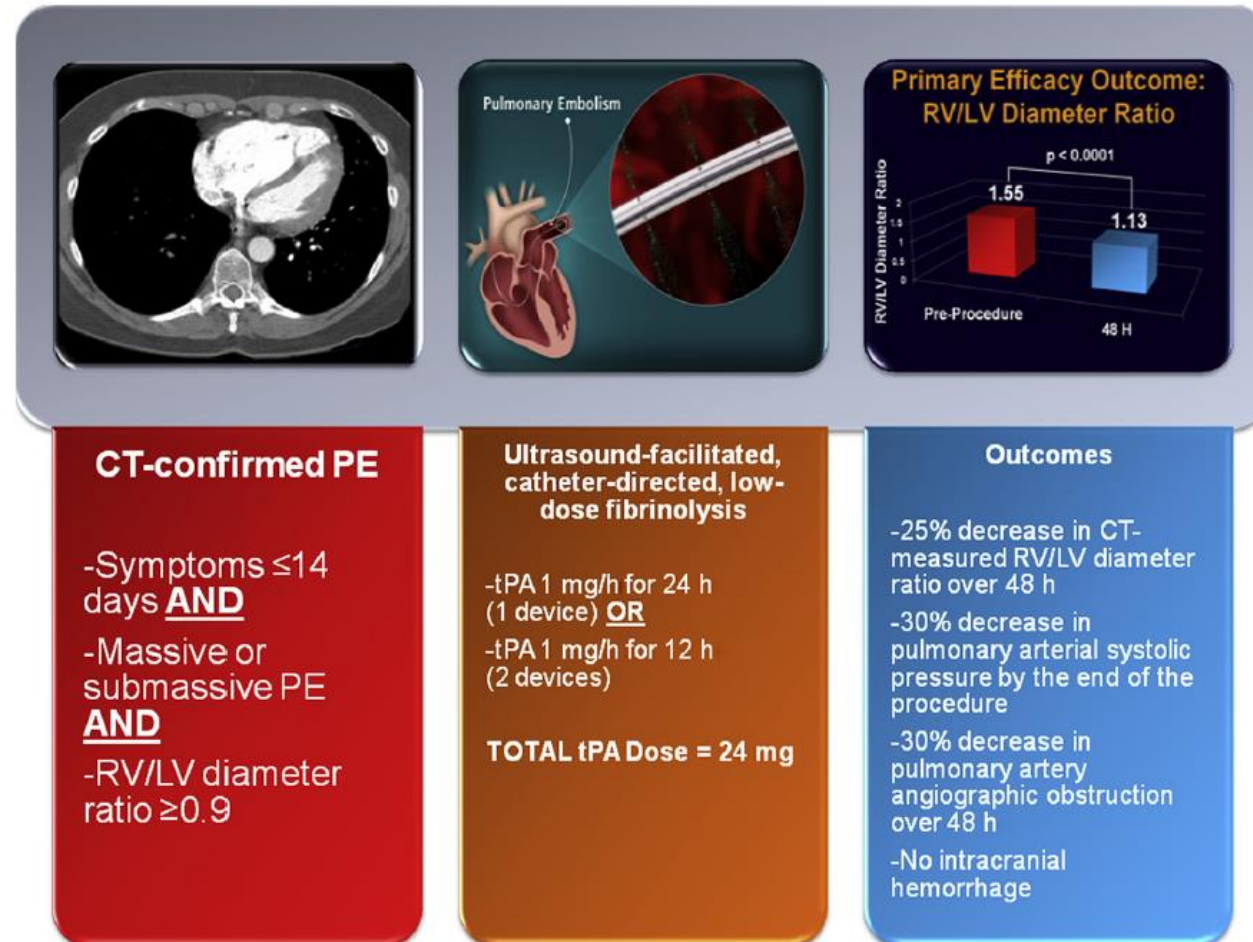


## B Major Extracranial Bleeding



# Seattle II Study

**CENTRAL ILLUSTRATION** Ultrasound-Facilitated, Catheter-Directed, Low-Dose Fibrinolysis for Acute Massive and Submassive Pulmonary Embolism



150  
Patients

Age 59

RV/LV 1.55

PA 51  
mmHg

# Case Study: JD47

47 y.o. man  
with several  
weeks of  
increasing  
dyspnea

Age 40

Episode of chest pain & SOB  
No further treatment

Age 43

for progressive shortness of breath, had  
CT-PE, and told of large clot in lungs →  
Anticoagulated

Reported HTN,  
dyslipidemia, family  
history of "clots"

## Medications

Diltiazem  
Furosemide  
Atorvastatin  
Warfarin

# Case Study – JD47



## Vitals:

120/82 mmHg HR 72 RR 14 94%- RA



## Carotids:

JVP 8-9 cm, prominent “a” wave



## Lungs:

Clear

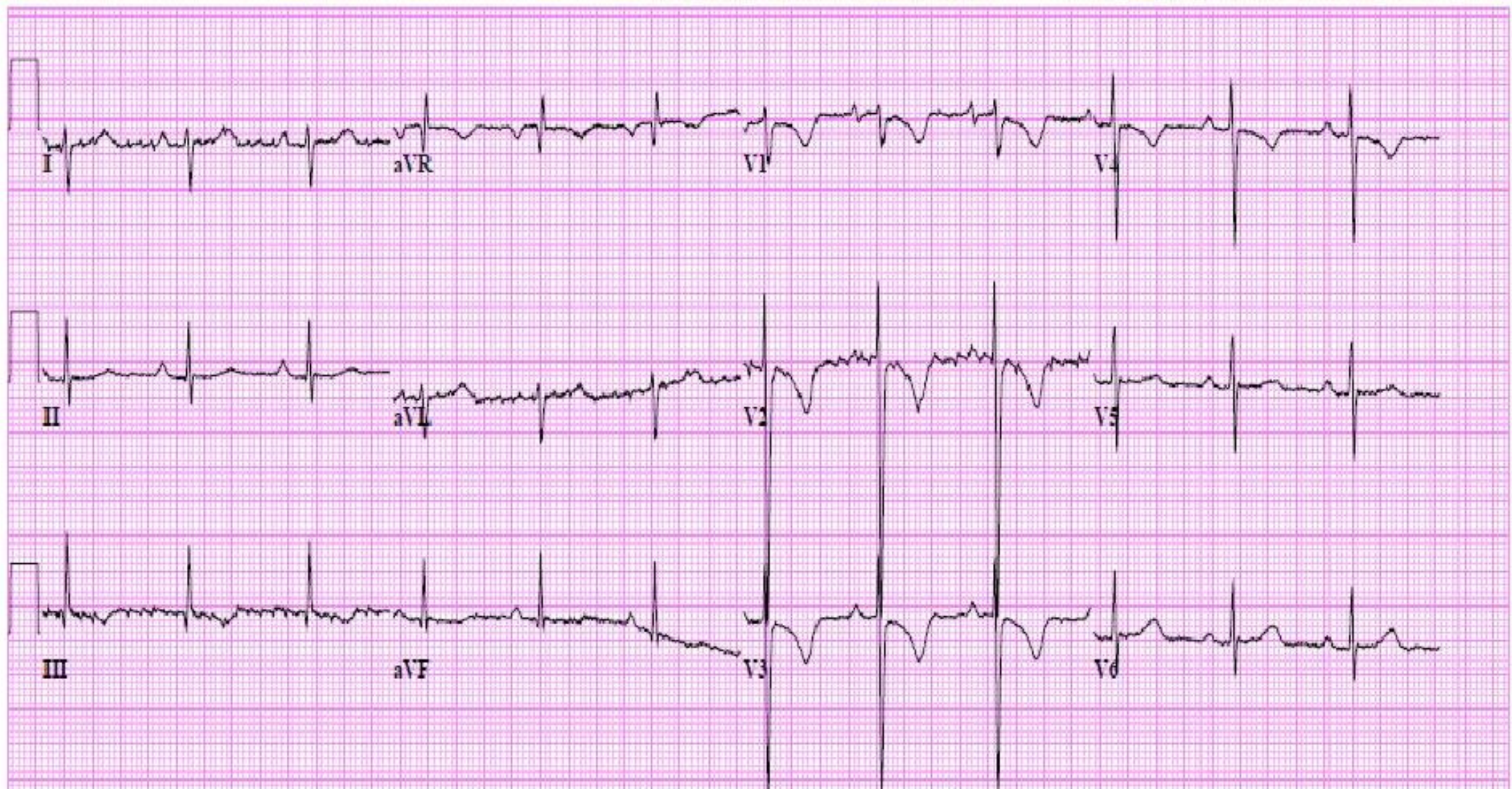


## Heart Sounds:

Left parasternal lift. PMI displaced laterally.  
Normal sounding S1, physiologically split S2,  
with P2 heard at apex, as well as in pulmonic  
and left lower sternal border regions



# Case Study: JUD47 - EKG





# Case Study – JD47

Tr-T below  
assay.

LABS

EKG

HR 72  
Intervals 0.19/0.08/0.42  
Axis 100 degree  
Right atrial abnormality,  
Right precordial T wave  
inversions

CT-PE shows occlusive  
right main PA  
thrombus and left  
lower lobe segmental  
thrombus. RV dilated.

ED Reports



# Case Study: JD47 - CT



# Case Study: JD47

WHAT'S  
**NEXT**



**1**

Lytic agents for acute large PE, with EKG, RV changes and risk of early decompensation.

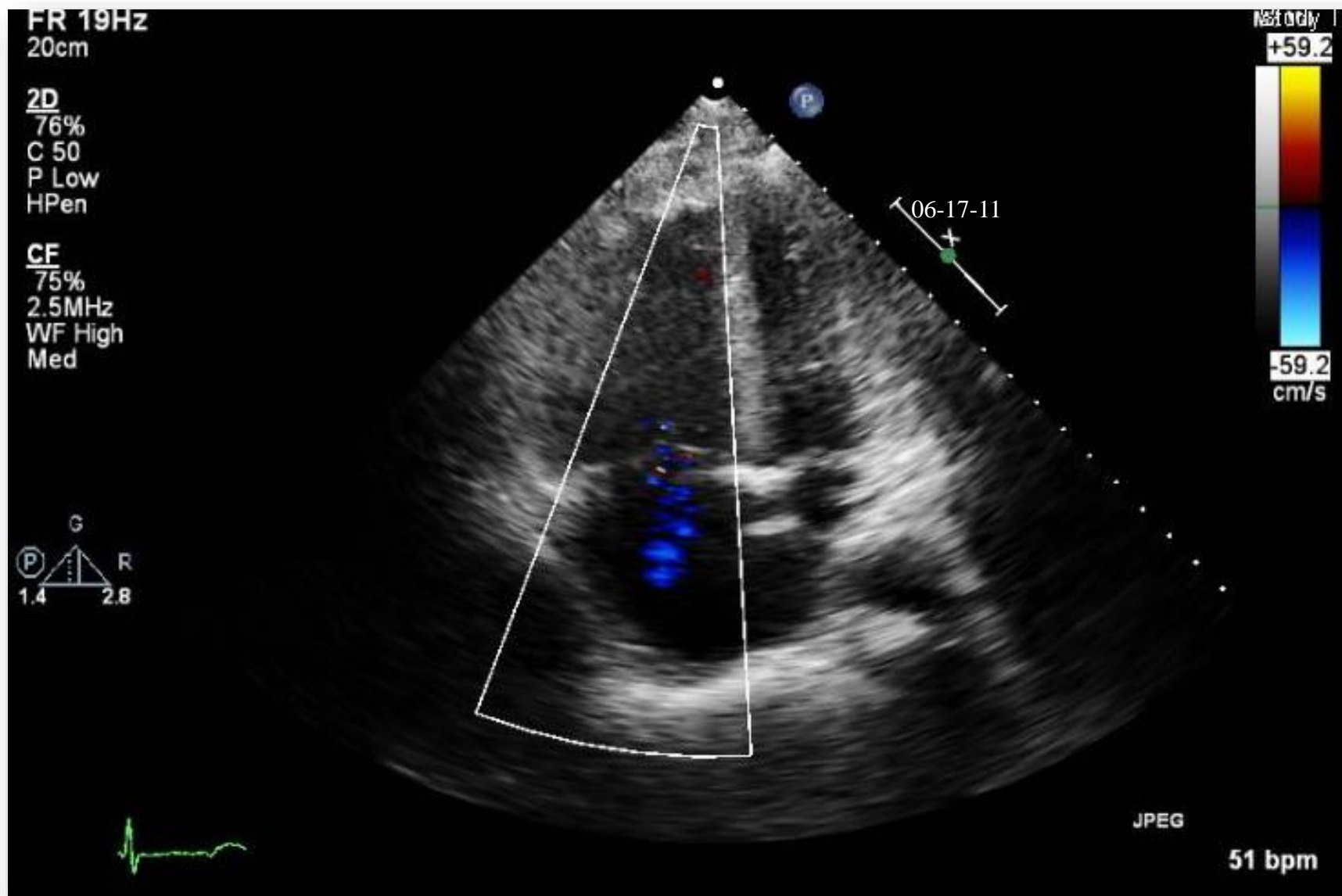
**2**

Heparinization and change to LMWH for small PE atop chronic thromboembolic pulmonary HT.

**3**

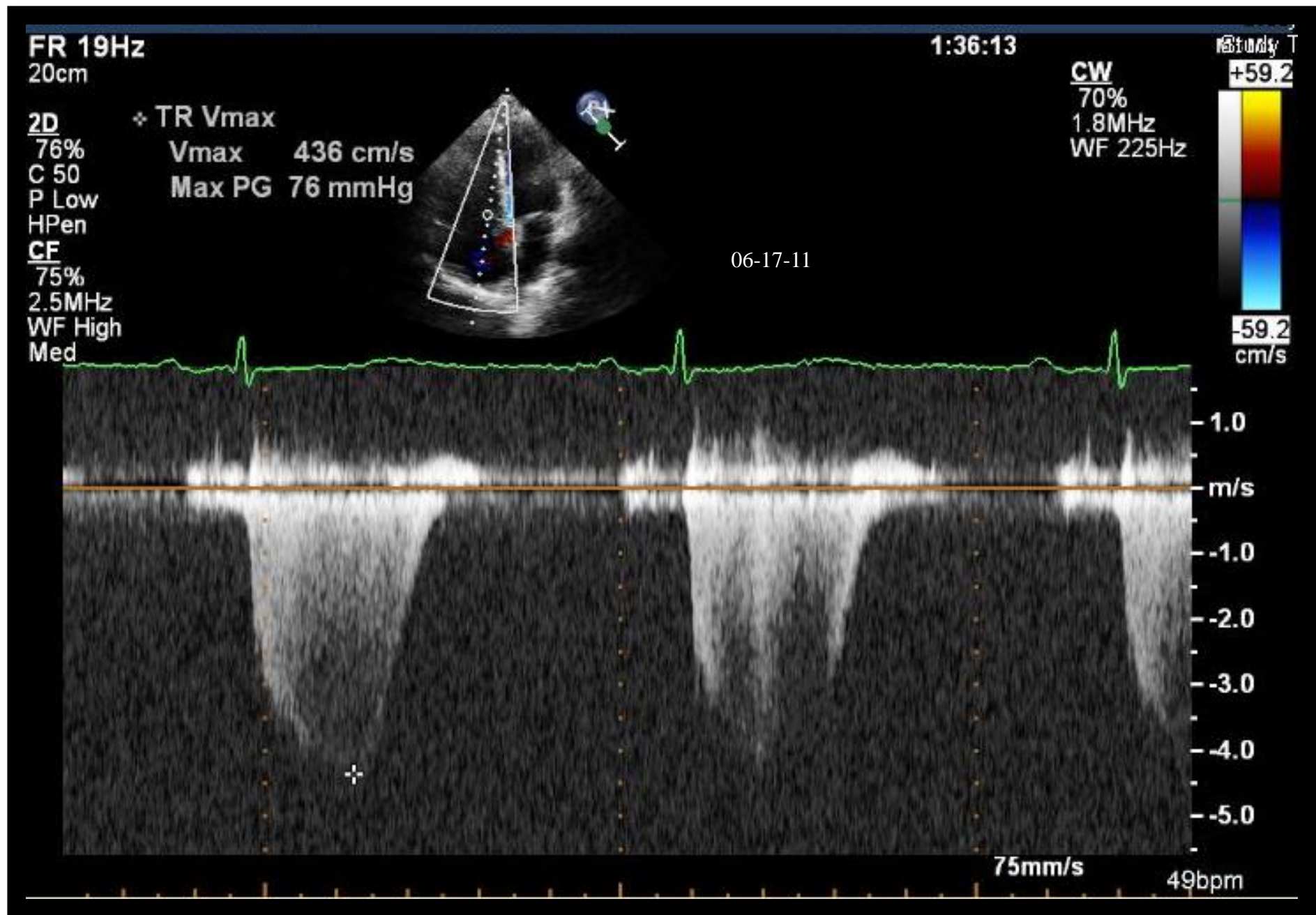
Echocardiogram, and if significant pulmonary HT, based on TR is PA catheter confirmed, elective surgical RX for CTEPH.

# Case Study: JD47 - Echo





# Case Study: JD47 - Echo



# Chronic Thromboembolic Pulmonary Hypertension

## Macroscopic

- Non resolved fibro-thrombotic obstruction in large and distal pulmonary arteries
- Resolve with surgery, balloon angioplasty

## Microvascular

- Appearance in the micro-circulation that resembles what is seen with pulmonary arterial hypertension
- Requires medical Rx
- ↑ Pressures in lungs: areas obstructed, blood moves with ↑ pressure to other segments
- May be broncho-pulmonary vessel venous shunting

# Medical Treatment, proof of principle. Surgery preferred

Riociguat soluble guanylate cyclase (sGC) stimulant

Macitentan: dual endothelial receptor antagonist – long acting

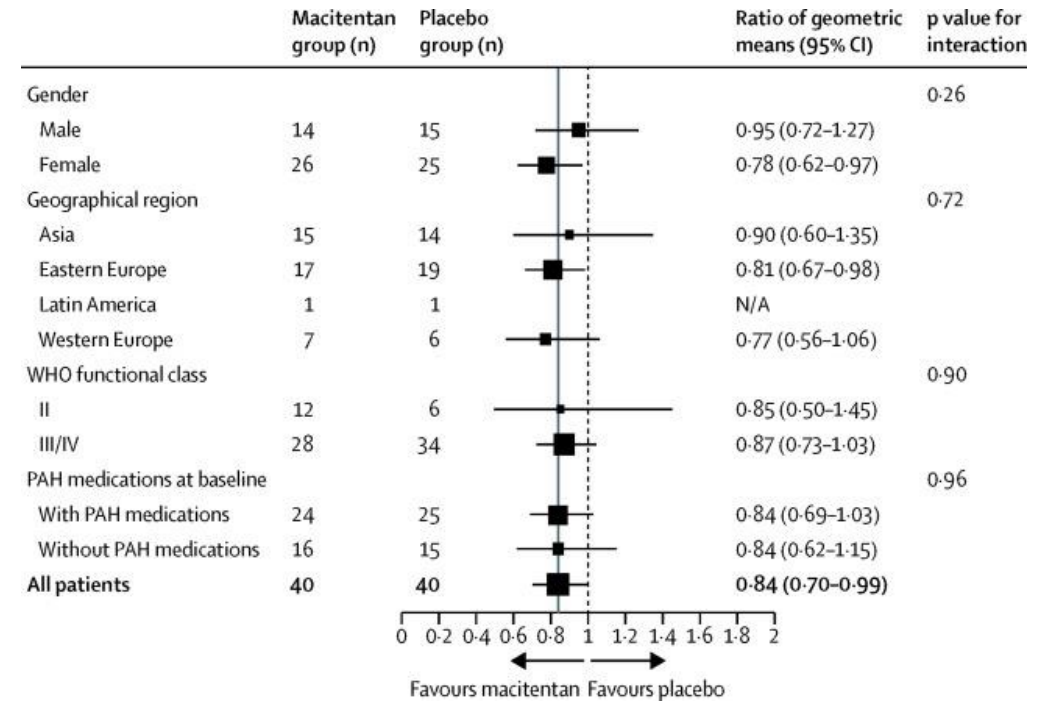
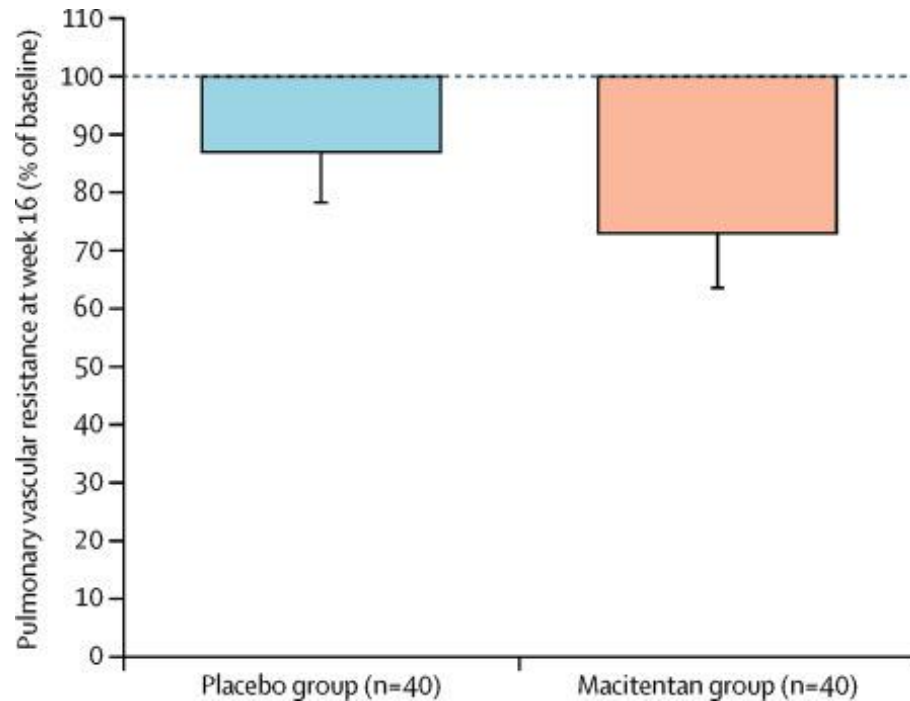
## Example

### MERIT 1 with Macitentan

- CTEPH – no surgery for various reasons
- Walk distance 150-450m
- Class II – IV
- PVR >400 dyn·s/cm<sup>5</sup>
- Mean PA ≥ 25, Mean PCW ≤ 15
- Stable doses of PDE5 inhibitor or inhaled prostanoid or Ca<sup>+</sup> channel antagonist

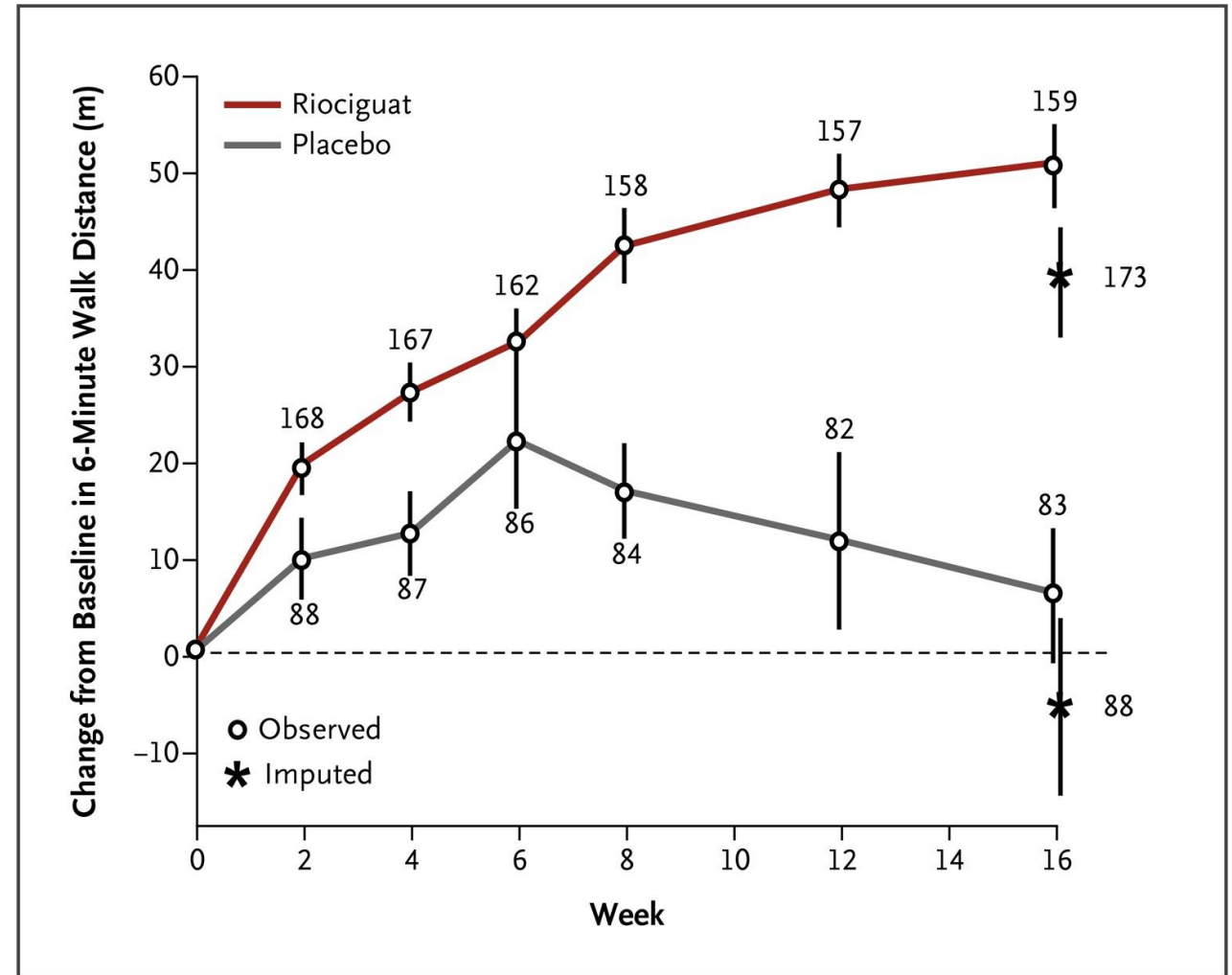
# MERIT: Macintentan, Endothelin Receptor Inhibitor in Operable CTEPH Patients

- Endpoints: PVR at week 16 (↓27%)
- Also assesses 6 minute walk at week 24 (↑35 m)
- NTProBNP at week 24 (↓28%)
- All significant differences
- \*Dyspnea index: no different







# Riociguat CHEST 1

- 261 patients
- Randomized 2:1
- 6-minute walk test





# **Race and Epidemiology of Venous Thromboembolism**

-  Incidence of VTE is 30-60% higher in blacks than whites.
-  Incidence of pulmonary embolism is higher in blacks, and percentage of black VTE patients who have PE is higher.
-  Incidence of fatal PE is higher in blacks, 3.73 vs. 1.15 deaths/100,000 people per year.
-  Idiopathic PE is seen in 18% of black, vs. 10% of whites. Sick cell trait, factor VIII, VWF ↓ protein C may play roles.

# Cardiovascular Risk

Enhanced in patients with post acute Covid sequelae (symptoms 5-12 weeks later)

Case control study of 13,435 US adults with post acute Covid sequelae; 26,870 matched adults with Covid

	PASC	No Covid	RR
Mortality	2.8%	1.2%	
Cardiac Arrhythmia			2.35 (2.26 – 2.45)
Pulmonary Embolism			3.64 (3.23 – 3.92)
Ischemic Stroke			2.17 (1.98 – 2.52)
CAD			1.78 (1.7 – 1.88)
Heart Failure			1.97 (1.84 – 2.10)
COPD			1.94 (1.88 – 2.0)
Asthma			1.95 (1.86 – 2.03)

# Outcomes

## Overall

- Double risk for lung, vascular, rhythm issues and mortality in patients with PACS

## Overall

- Always worthwhile to have vaccines, boosters, masks, ventilation

# Delta vs Omicron BA1, BA2 Wave

---

## 274 Patients

---

## No history of Covid or Vaccine

---

## 30-65 years old

	Delta	Omicron	Total
Developed CoVID	27 17%	123 83%	62%
Asymptomatic	0	8 6%	
Post Acute (>5 weeks)	13 48%	26 21%	26% post acute

## Conclusions and Implications

- Carefully followed cohort from mid 2021 to early 2022
- No vaccine/never had Covid (from early 2020 – mid 2021)
- 61% had Covid
- 26% had post acute syndrome
- But Omicron patients have much lower risk of post acute (↓ by 56%)  
**and**
- Much lower risk of needing health care assistance (↓ 79%)



# Case Study: LX

**58 yo woman brought from dialysis center with left arm and chest discomfort & “not feeling well”**

Bipolar disorder and in problematic marriage with alcohol dependent husband

ESRD secondary to prior lithium toxicity

Impaired glucose tolerance

Dyslipidemia

Catheterization 15 months prior for CP and reported NSTEMI:  
PA 45/10 mmHg  
No gradient across outflow tract or aorta  
EF 65%  
30% LAD and LCx stenoses.



# Case Study: LX - Exam



## Vitals:

BP 126/80

HR 92

RR 14

Pulse ox: 97% on 2L.

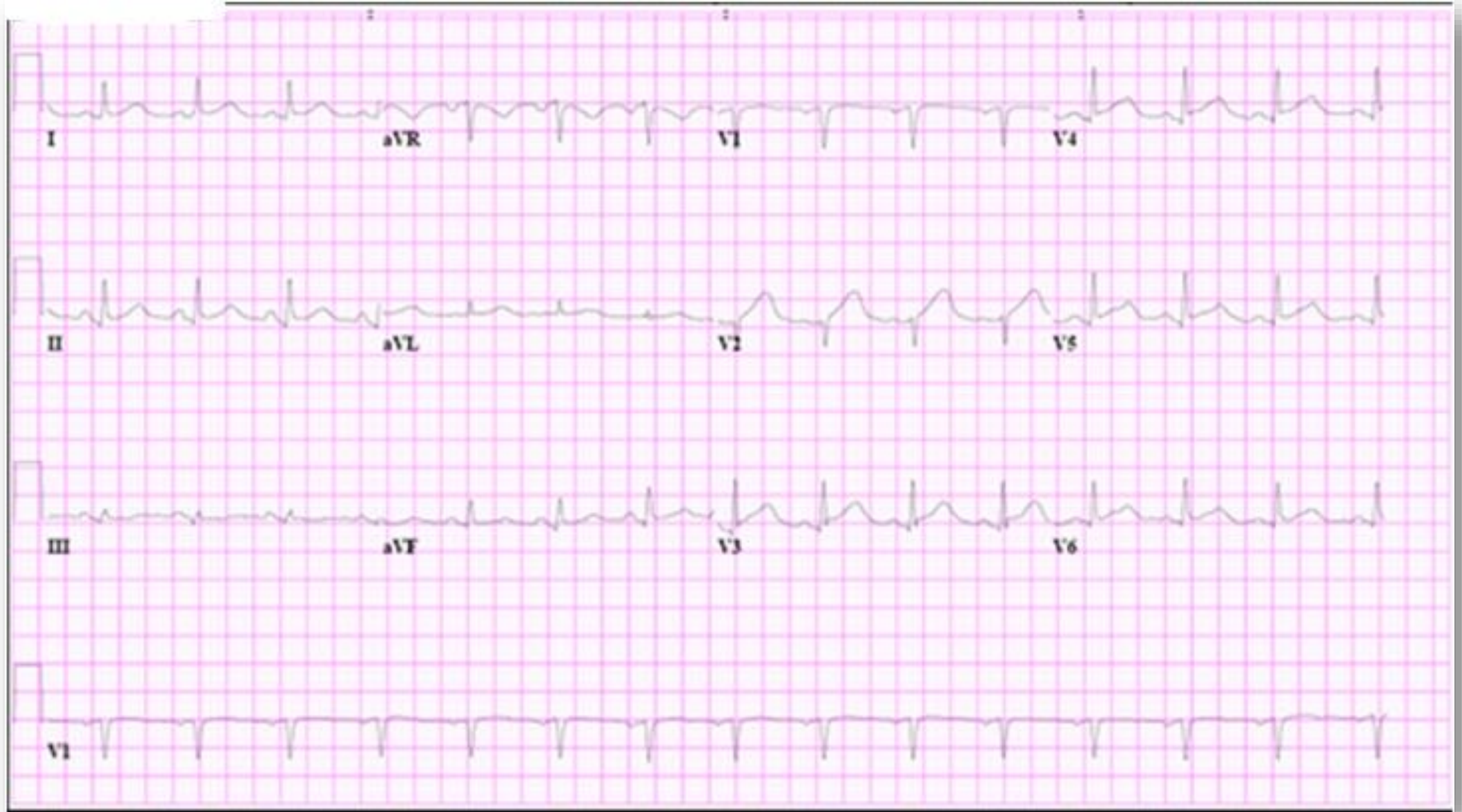
- ✓ JVP not elevated.
- ✓ Carotid upstrokes brisk, volumes normal.
- ✓ Right base crackles.
- ✓ Non-displaced PMI.
- ✓ Normal, non-muffled  $S_1$   $S_2$   $\Phi$

## Labs:

Tr I 0.29  
(.034)

HCT 30

**Case Study: LX - EKG at end of dialysis**



# Which Of Following Statements are True:

**A**

Presentation is consistent with coronary spasm, stress induced cardiomyopathy, or plaque rupture. Echo will distinguish between these entities.

---

**B**

No matter what echo shows, she should undergo angiography.

---

**C**

No matter what echo shows, there is no need for angiography.

---

**D**

She may have had stress induced cardiomyopathy last time she had catheterization, and likelihood of a recurrence is very, very low.

---

**E**

a and d.

---

# Case Study: LX

## Echo:

- Extensive anteroapical, inferoapical, apicolateral akinetic region
- Vigorous contraction of LV outflow tract region
- Mitral regurgitation

## Cath:

- No coronary artery irregularities
- Single coronary could not explain wall motion abnormality

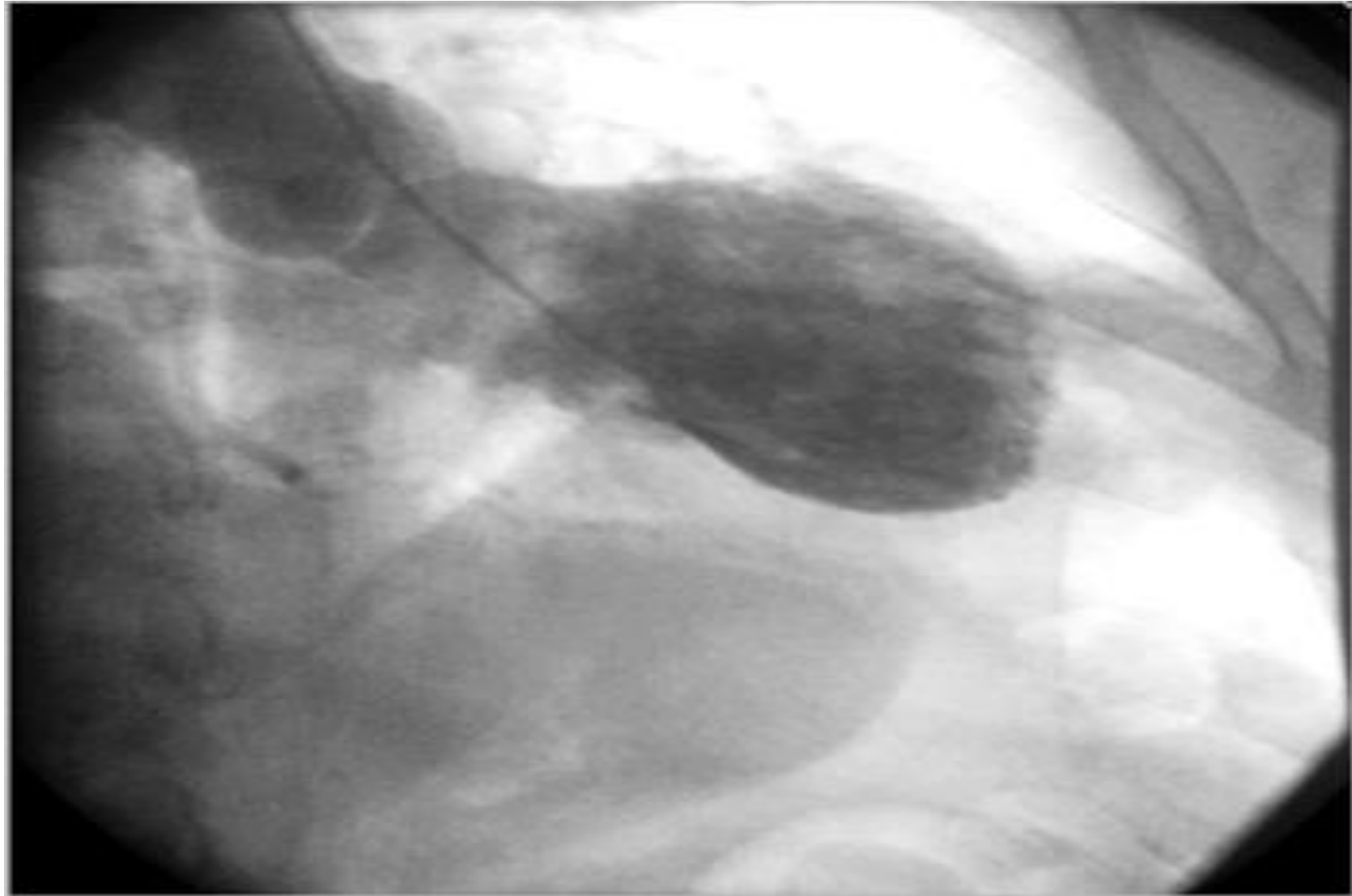
## V-gram:

- Like echo



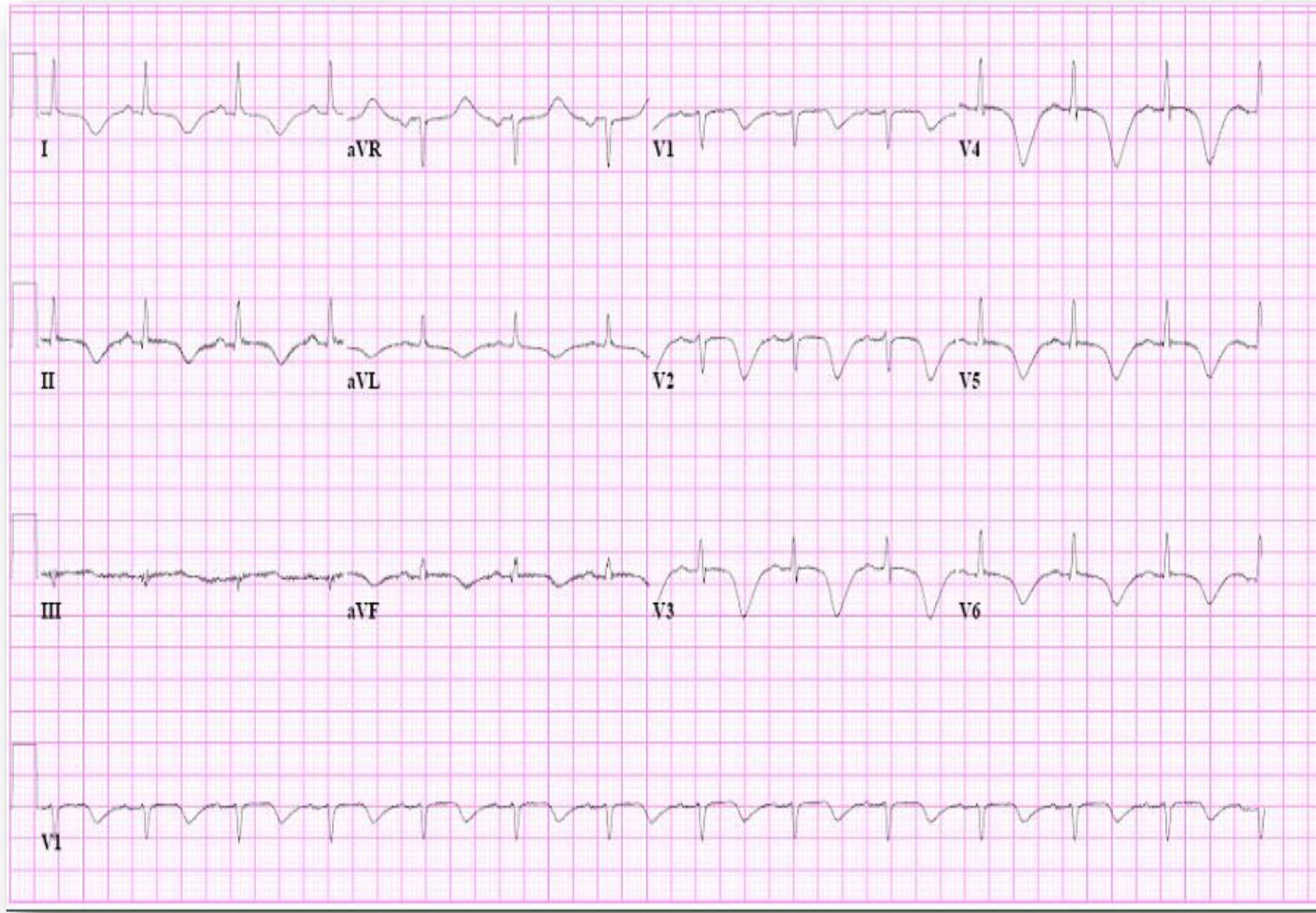
# **TAKOTSUBO**

**Extensive Mid Anterior Apical And Infero Apical Akinesis**





**Case Study: LX – EKG 48 Hours after Admission**





**Diagnosis:  
Takotsubo's  
or  
Stress Induced  
Cardiomyopathy**

**Seen in  
association  
with:**

Emotional or physiologic stress, including sepsis, cerebral events, pheochromocytoma.

Characterized by sympathetic stimulation

Can have full range of ST-T wave abnormalities.

Can be any region of myocardium, can have fixed perfusion defects, and can have myocardial edema (not late gadolinium enhancement).

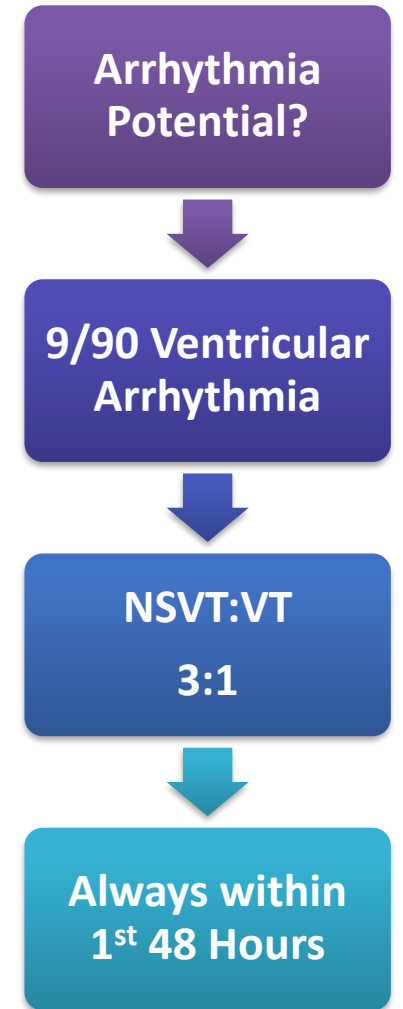
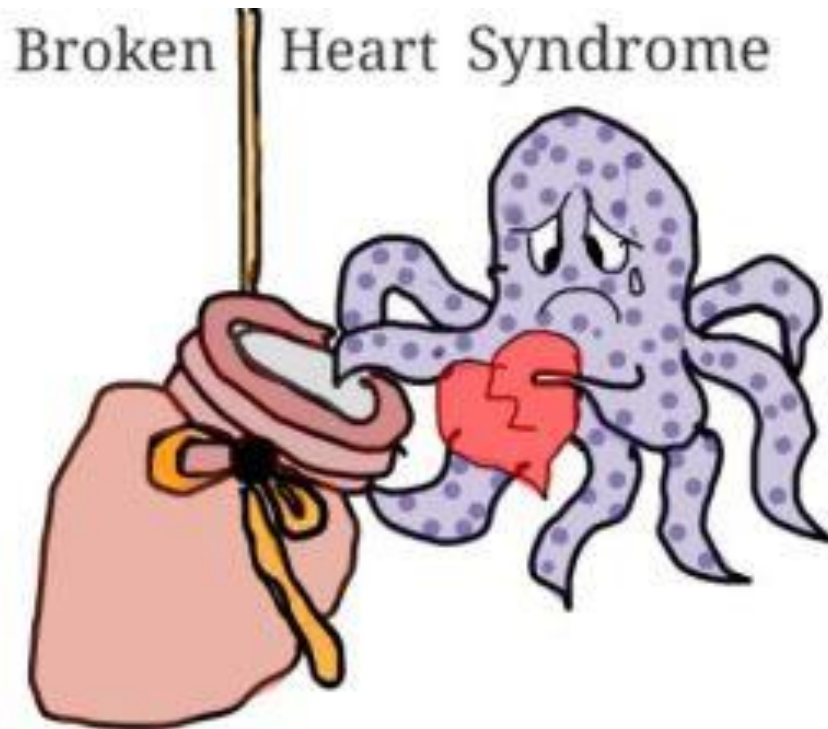
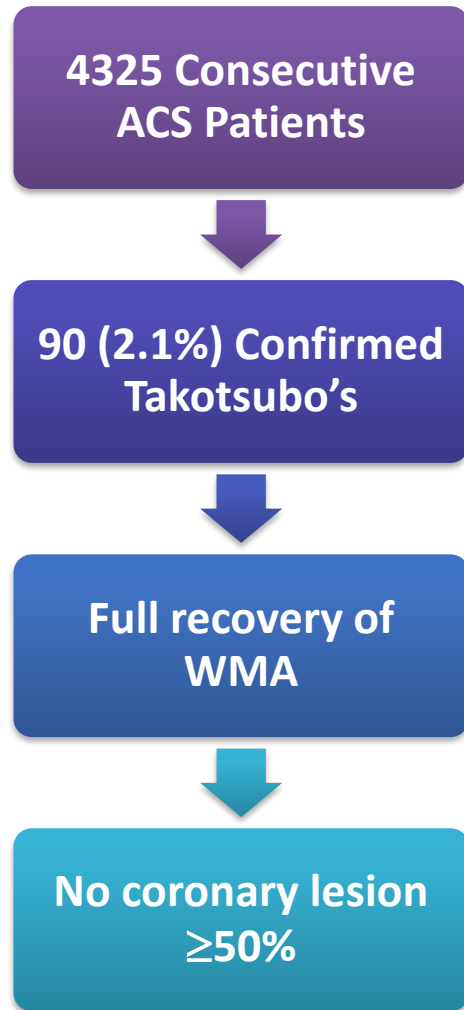
Can be associated with transient outflow tract obstruction and MR.

Rare cases of VF or rupture.

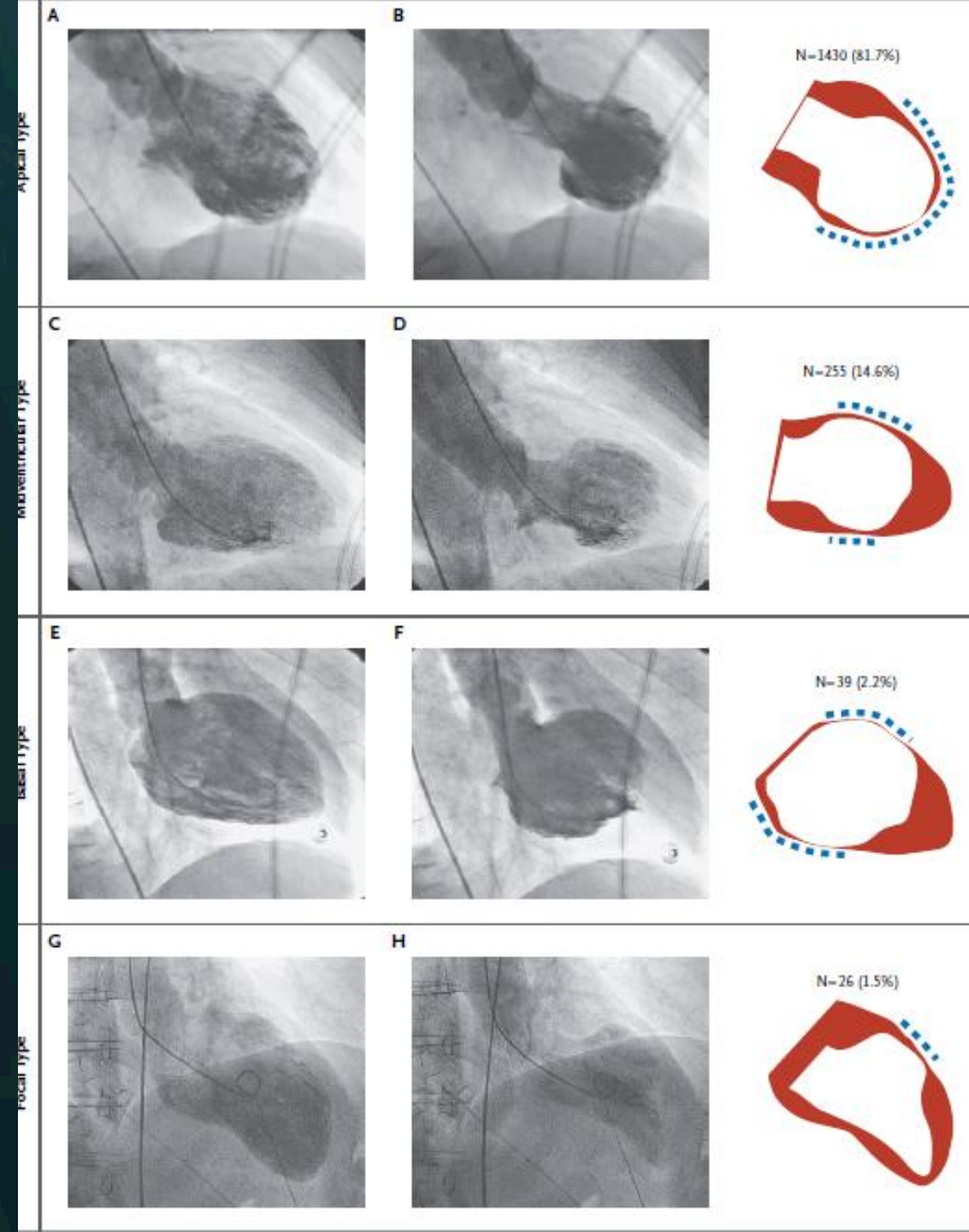
Usually, LV gets entirely better, with variable time course.

Can recur in 10 – 15% of patients.

# Takotsubo's Frequency



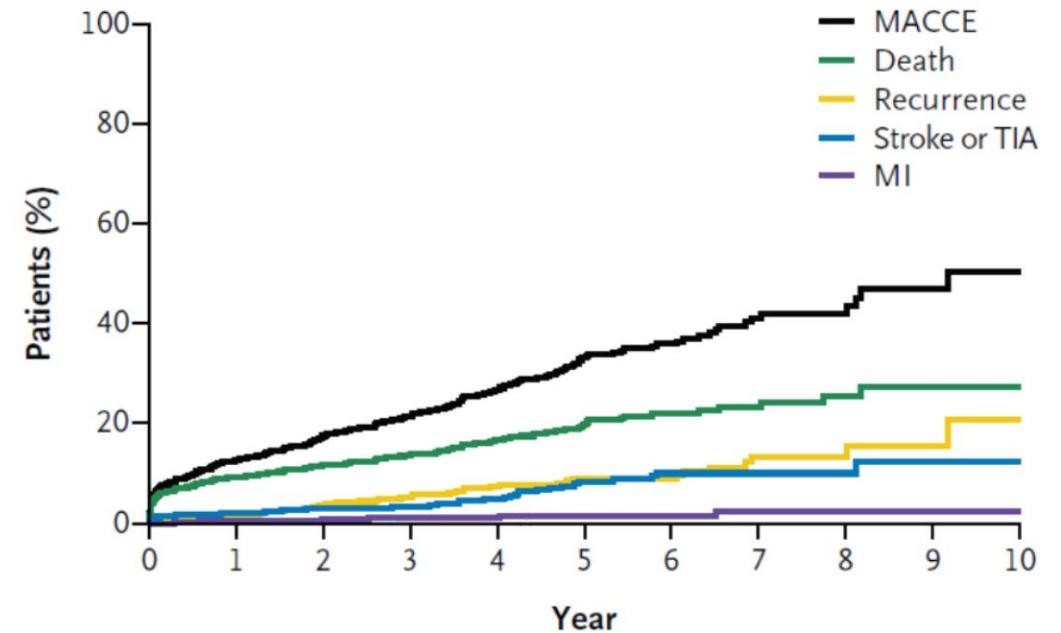




# Takotsubo.

## Does not have to be apical.

# Takotsubo's is NOT Benign



No. of Patients 1750 786 570 431 300 191 126 71 38 17 9

**Figure 3. Kaplan–Meier Estimates of 10-Year Outcome Events.**

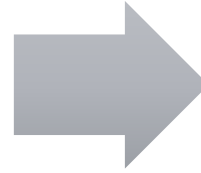
Shown are the proportions of patients with any major adverse cardiac and cerebrovascular event (MACCE), which was a composite of death from any cause, recurrence of takotsubo cardiomyopathy, stroke or transient ischemic attack (TIA), or myocardial infarction (MI).

Though most get better, and recurrence rate is ~ 15%,  
there can be stroke, arrhythmia, death...over 30 days.

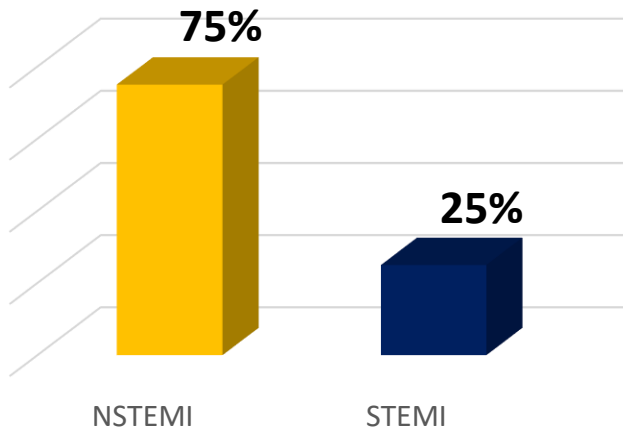


# Spontaneous Coronary Artery Dissection

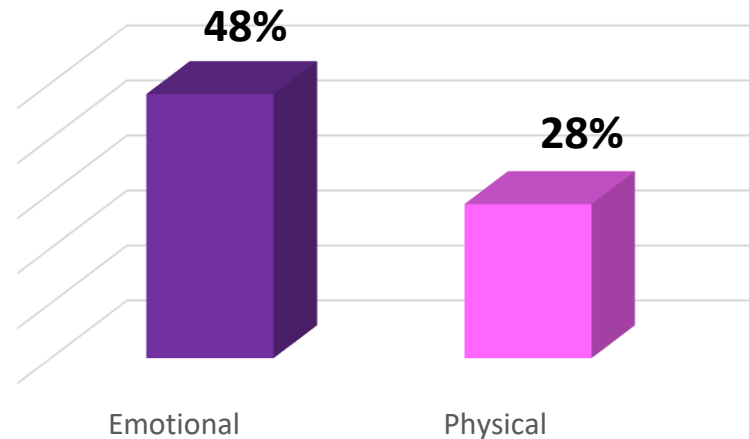
337 patients



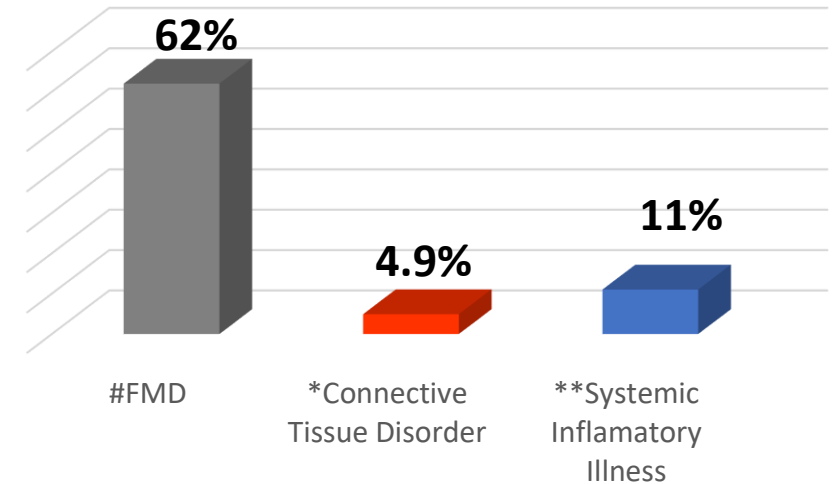
All presented with ACS, generally young, fewer risks



3.1 year follow up



10% recurrent SCAD



- #Carotids, Renal/Iliac screen – string of beads
- \*Marfans, Loeys Dietz, Ehlers Danlos T4, Polycystic KD, ±post partem
- \*\*RA, SLE, Crohns, UC, Celiac

# **Case Study: RP53 - Background**

## **83 Year Old Man**

**With easily precipitated exertional angina (after a meal, has angina if he walks across street).  
Duration 1 year.**

## **Hypertension and CKD**

**Cr 1.7 mg/dl.  
Treated with Lisinopril & Metoprolol.**

## **Dyslipidemia**

**Remote TC 340 mg/dl. Presently, TC 174,  
HDL 81, LDL 71, TG 71.  
Treated with atorva and icosapent**

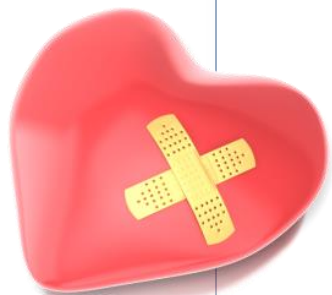
## **Type II Diabetes**

**A1c 7.4%  
Treated with sulfonyl urea, should be  
consideration for GLP1 RA.**

# Case Study: RP53 - History



Works everyday as a hands-on manager in a family business.



Patent LIMA-OMB, SVG-AM occluded, Sequential SVG-D1-LAD with 99% stenosis. Received SVG-A.M, SVG-LAD

**1988 Evolving Anterior MI**

**Quit Smoking in 3<sup>rd</sup> Decade**

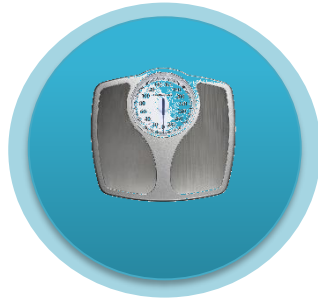


Received TPA, then underwent LIMA-OMB, SVG-AM, sequential SVG-D1-LAD.

**1999 Accelerating Angina**



# Case Study: RP53 - Exam



184 lbs  
extra 15 lbs. in abdomen



149/64 in both arms  
HR 60 regular  
RR 12



Terrific memory

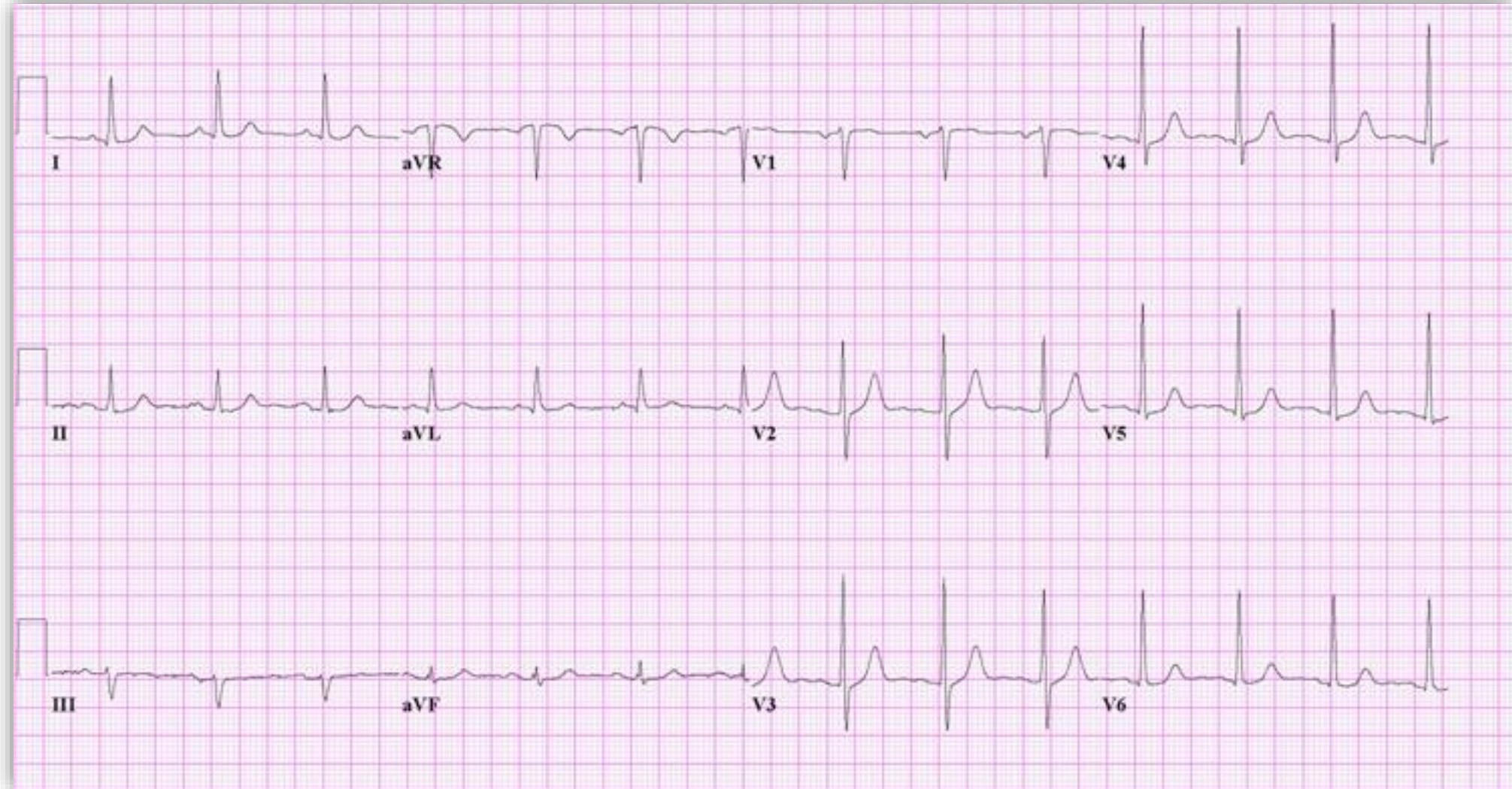
## Exam

- JVP < 8 cm
- Carotid upstrokes **with slight delay**
- Good volumes
- Transmitted murmur
- Clear lungs

## Cardiac Exam

- Normal S1.
- Single S2, **heard only over pulmonic region.**
- 3/6 late peaking, nearly honking systolic murmur heard from apex to base to neck and in back, that softens with Valsalva
- Normal distal pulses

# Case Study: RP53





## **Most likely reason (s) for this patient's easily precipitated angina (worse post prandial)**



Hiatal hernia  $\pm$  Para esophageal hernia without erosive esophagitis.



Aortic stenosis combined with or without native or graft coronary disease.



Mitral regurgitation and hypertensive/diabetic microvascular impairment.

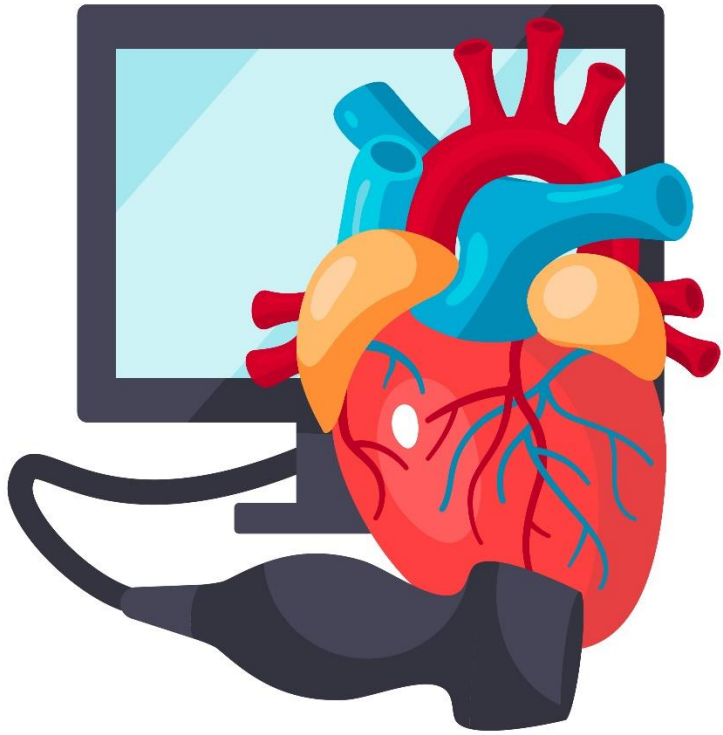


Worsening anemia and outflow tract obstruction.



Left subclavian stenosis jeopardizing flow to LIMA.

# Case Study: RP53 - Echo



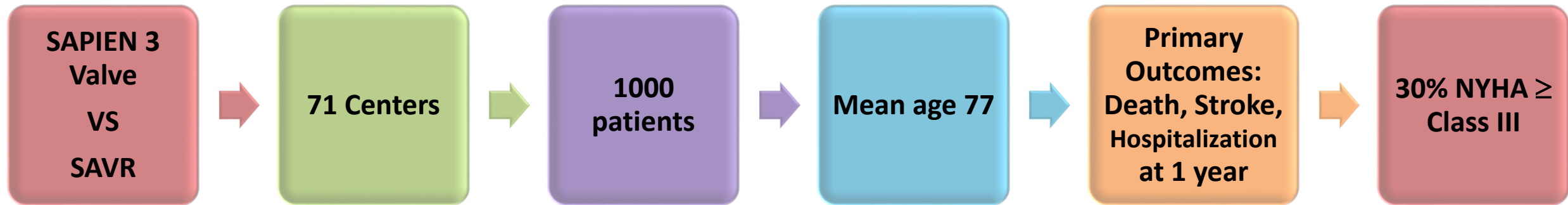
- EDD 5.6
- ESD 4.1
- Wall thickness 1.4cm
- EF 50%
- No wall motion abnormalities
- Heavily calcified aortic valve
- Mean gradient 44
- VMAX 4.4 m/sec
- TVI below the valve 23, above the valve 119
- AVA 0.7cm<sup>2</sup>

# Case Study: RP53 - Cath



- Patent grafts
- RCA occluded after acute marginal
- Good L→R collaterals.
- Acute marginal mid 95% stenosis
- Occluded left main
- Far distal LAD 80% stenosis
- Fully patent, large ileofemoral systems.

# TAVR has evolved, and now used in high, intermediate, and low Risk Patients



	12 Months Death Stroke reHosp	Death	Stroke	ReHosp	AFib	New LBBB
“SAPIEN” 3 - TAVR	8.5%	1.0%	1.2%	7.3%	5	23.7%
SAVR	15.1%	2.5%	3.1%	11%	39.5%	8.0%

Wary of: Bicuspid valve, calcified outflow tract, pacer incidence, perivalvular leak, early thrombosis

# Case Study: 39 YO Woman

Occasional  
orthostatic  
lightheadedness

Maternal  
Grandfather died at  
age 55

No history of heart  
murmur

**Feels Well**

Medications:

- estrogen/progesterone
- zolpidem



# Case Study: 39 YO Woman

## Vitals

68" tall

120 pounds

100/70 mmHg 62 bpm

90/60 mmHg 70 bpm

## Exam

Neck veins not elevated

Carotid upstrokes brisk, normal volumes

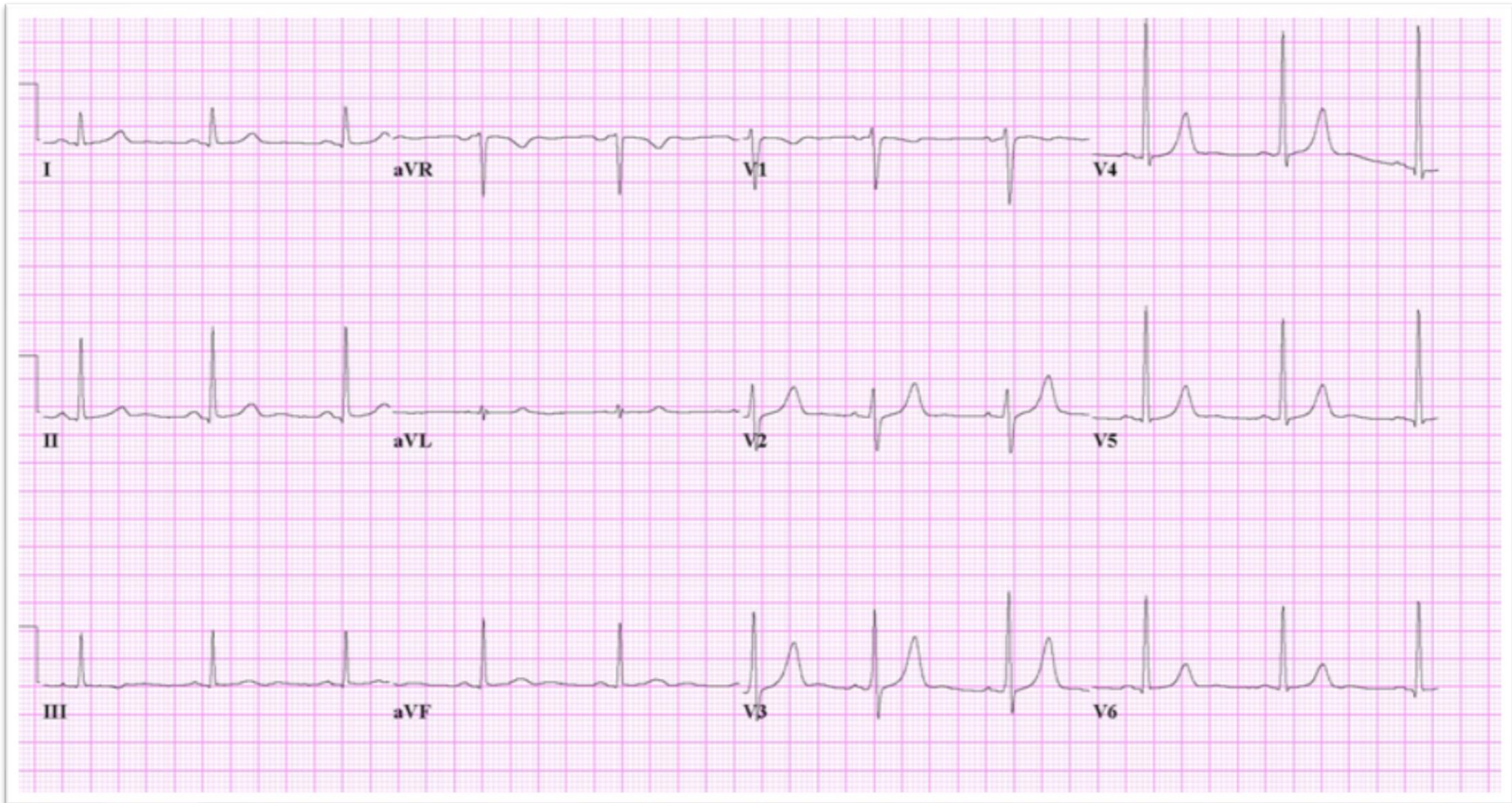
Clear lungs

Minimal pectus excavatum

## Cardiac

- PMI at 5<sup>th</sup> intercostal space, mid clavicular line
- No thrill
- Loud S1, S2 phys. split
- III/VI systolic murmur loudest just left of LLSB, becomes minimally louder with Valsalva and moves later with handgrip
- Extra sound after S1 with handgrip

# Case Study: S99 - EKG



# Case Study: S99 - Diagnosis

**A**

Autonomic insufficiency, unclear etiology.

**B**

Mitral stenosis.

**C**

Obstructive hypertrophic cardiomyopathy.

**D**

Mitral valve prolapse and mild mitral regurgitation.  
Can follow with yearly exams.

**E**

Posterior mitral valve prolapse and significant mitral regurgitation.

**F**

A and D.

# Movement of MV Clicks

MITRAL VALVE PROLAPSE/*Devereux et al.*

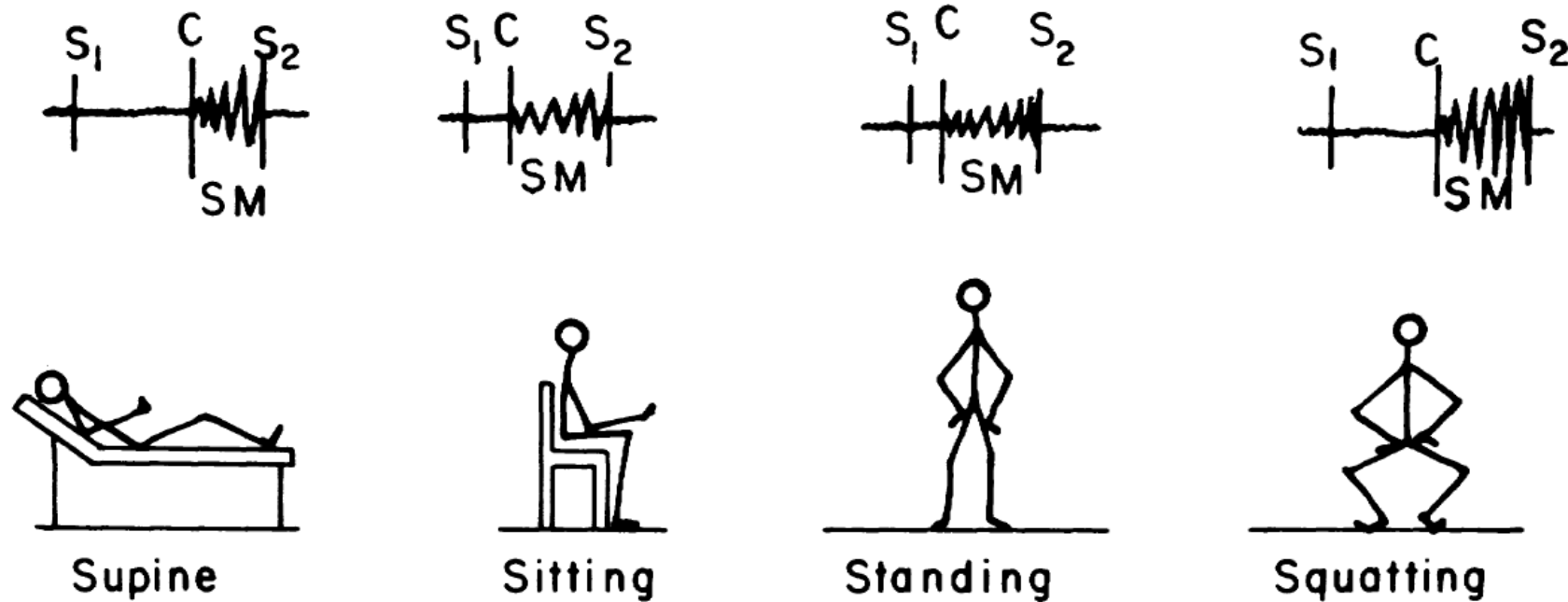


FIGURE 6. Postural changes affecting the auscultatory signs of mitral prolapse. On sitting and standing, the systolic click moves closer to  $S_1$  and the murmur is prolonged. On squatting the click moves toward  $S_2$  and the murmur becomes shorter. These auscultatory variations are related to changes in left ventricular volume and shape (see fig. 5).



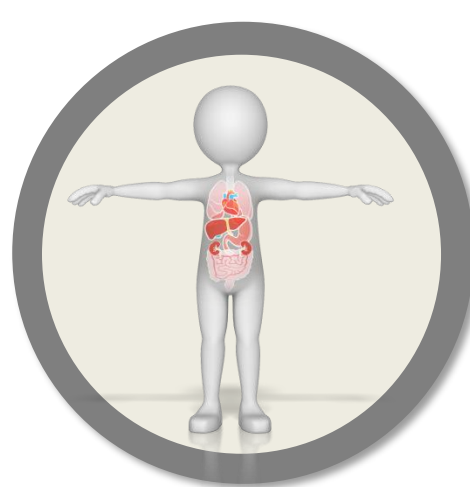


### **34-YEAR-OLD WOMAN**

Abdominal Pain

Nausea & Emesis for 9 hours

Suspect bowel obstruction



### **MEDICAL HISTORY**

Paraplegia in aftermath  
of MVA

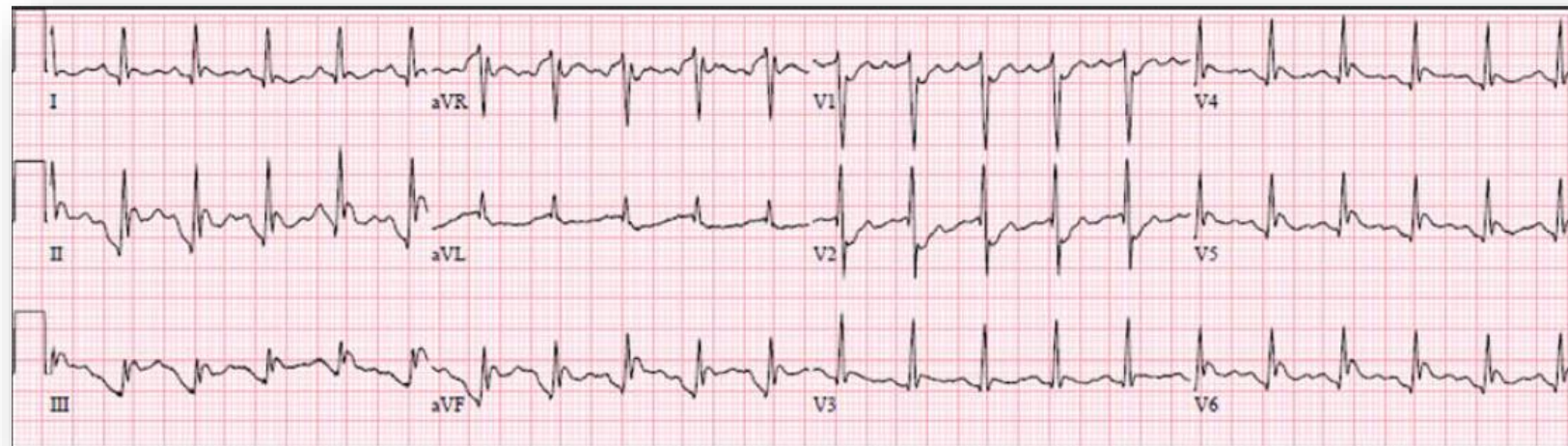
Colostomy



### **EXAM**

BP 124/84 mmHg

Abdominal distention

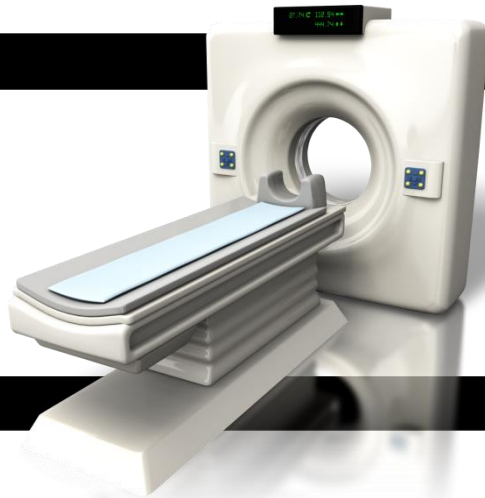




**Troponin T Negative**

**Echo**

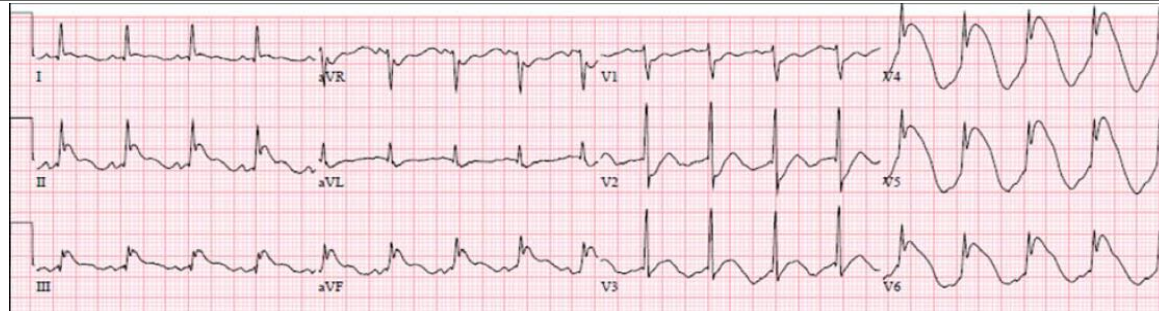
Ejection Fraction 65%  
No Wall Motion Abnormalities



**Chest CT**

No Pericardial Effusion  
No Mass

**EKG**





**Figure 2.** Admission computed tomography of abdomen and pelvis without contrast. The stomach (white asterisk) is markedly distended with fluid. Also present are dilated fluid-filled small-bowel loops (white arrowheads).

## Discussion

Represents Giant J Waves (Osborne Waves)

Sometimes seen with hypothermia

ST elevations

Due to gastric distention

Key is prominent J Waves that worsen, with normal echo (not ischemia/myocarditis/pericarditis)

# CASE STUDY S17

51 YO  
Woman



Abrupt onset pleuritic  
chest discomfort –  
worse supine



Hodgkin's Disease Stage IIA – 22  
years previously treated with Mantle  
RT and MOPP following splenectomy



Treated  
hypothyroidism



Soleal DVT 3  
years previously



Contrast  
allergy



# CASE STUDY S17

## Vitals

BP 110/80-10mmHg paradox  
HR 110 beats per minute  
RR 20

---



## Neck Veins

Mid neck level seated upright  
Collapse with inspiration

---

## Lungs

Clear  
No palpable PMI

---



## Carotids

Upstrokes brisk  
Volumes diminished

---

## Heart Sounds

Normal S1, single S2  
II/VI early peaking  
systolic murmur  
along LLSB

---

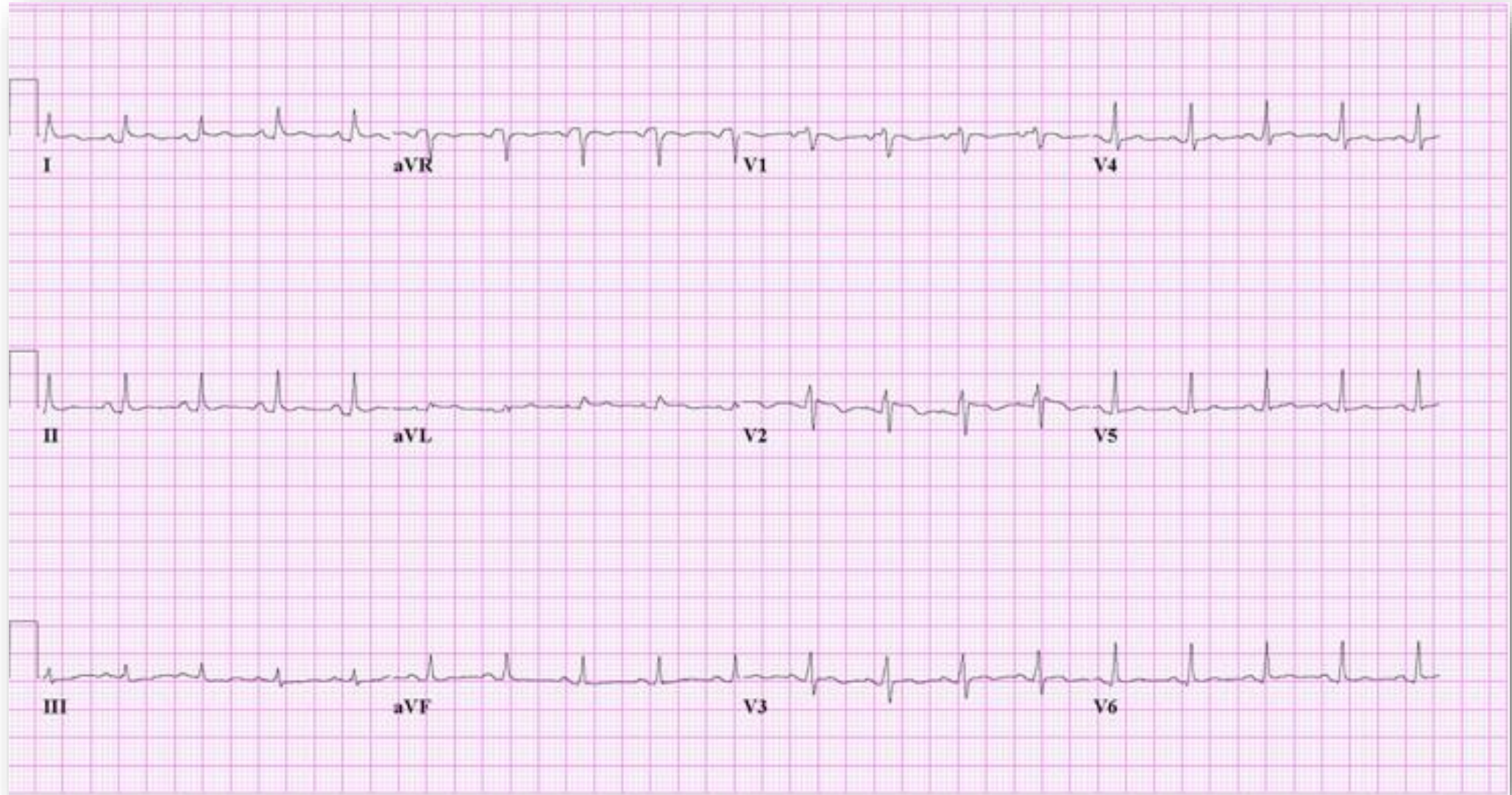


## No Peripheral Edema





# CASE STUDY S17





# Which of the Following are True?

A

Pleuritic discomfort against her background of DVT suggests pulmonary embolism as possible diagnosis

B

Radiation & symptoms and EKG suggest pericardial processes that could include effusive disease or effusive constrictive disease

C

Prior radiation & symptoms suggest chronic constrictive pericarditis

D

Appropriate testing would include ventilation perfusion study and surface echo-doppler

E

All except C

F

All except B

# Case Study S17: ECHO




Large circumferential  
pericardial effusion

No diastolic collapse  
of RV or RA

Transmitral early filling  
falls 35% with inspiration

Transtricuspid early  
filling increases 75% with  
inspiration

# Case Study S17: Hemodynamics



All diastolic pressures between 18-20 mmHg



Blunted “y” descent in RA



Blunted early filling in RV

# Case Study S17: Hemodynamics

**Pericardial Pressure =**

**A**

18-20  
mmHg

**B**

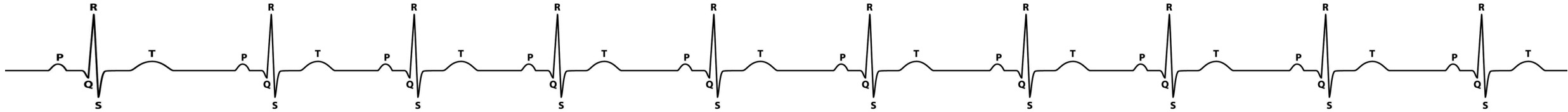
Zero

**C**

5-10  
mmHg

# EKG Changes in Acute Pericarditis

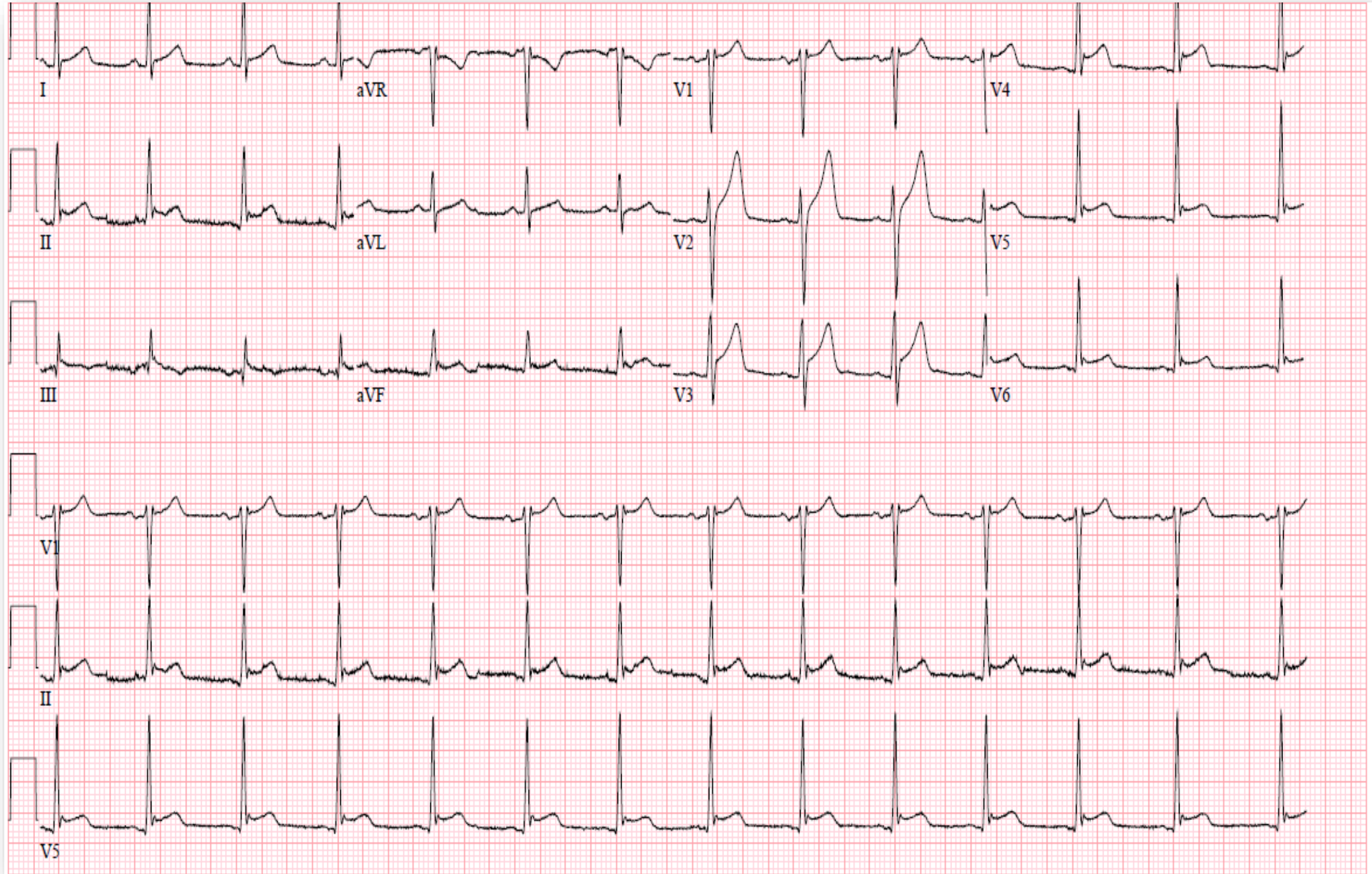
- Diffuse ST segment elevation with upright T-waves (lead aVR may have ↓ ST)
- PR segment depression, except in lead aVR, where there is elevation
- J point/T wave peak ratio often > 25%
- NO “reciprocal changes” except in R



- Resolution
- Normalization of PR segments and ST segments, followed by T wave inversions
- Ultimately resolution of T wave inversions
- Time course highly variable, effusions/myocarditis alter QRS amplitude
- Tamponade (especially from a complex effusion) may be accompanied by QRS electrical alternans



# Case 234



# Recurrent Pericarditis & Colchicine

- Little interest in glucocorticoids
- Interleukin-1 (↑ CRP Levels) can be attacked by interleukin-1 receptor antagonist anakinra
- Interleukin-1 $\alpha$  and especially Interleukin-1 $\beta$  can be attacked by rilonacept, a cytokine trap



COLCHICINE

# RHAPSODY: Safety

7% (2) rilonacept patients (had missed doses) had recurrence at 2 years

74% placebo had recurrence (50% by week 8)

All given bailout rilonacept

## Adverse Events

Generally mild  
Occurred early

Hypersensitivity site reaction

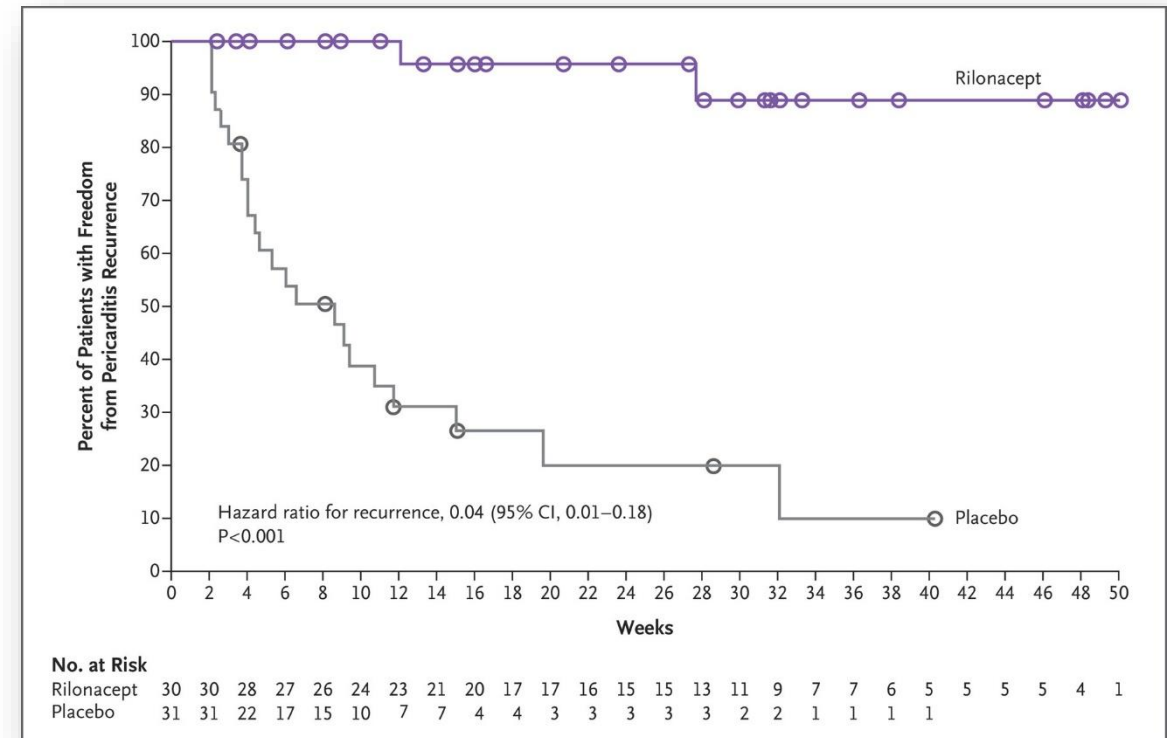
Allergic alveolitis

Alopecia

Only 4/81 needed to stop (during run-in phase)

LDL (↑ 13)

TG (↑ 10%)



# Case Study 322

## Patient History



21 year old man

3 days of intermittent flu-like symptoms

“Lung burning” – worse supine, non-pleuritic

MVP

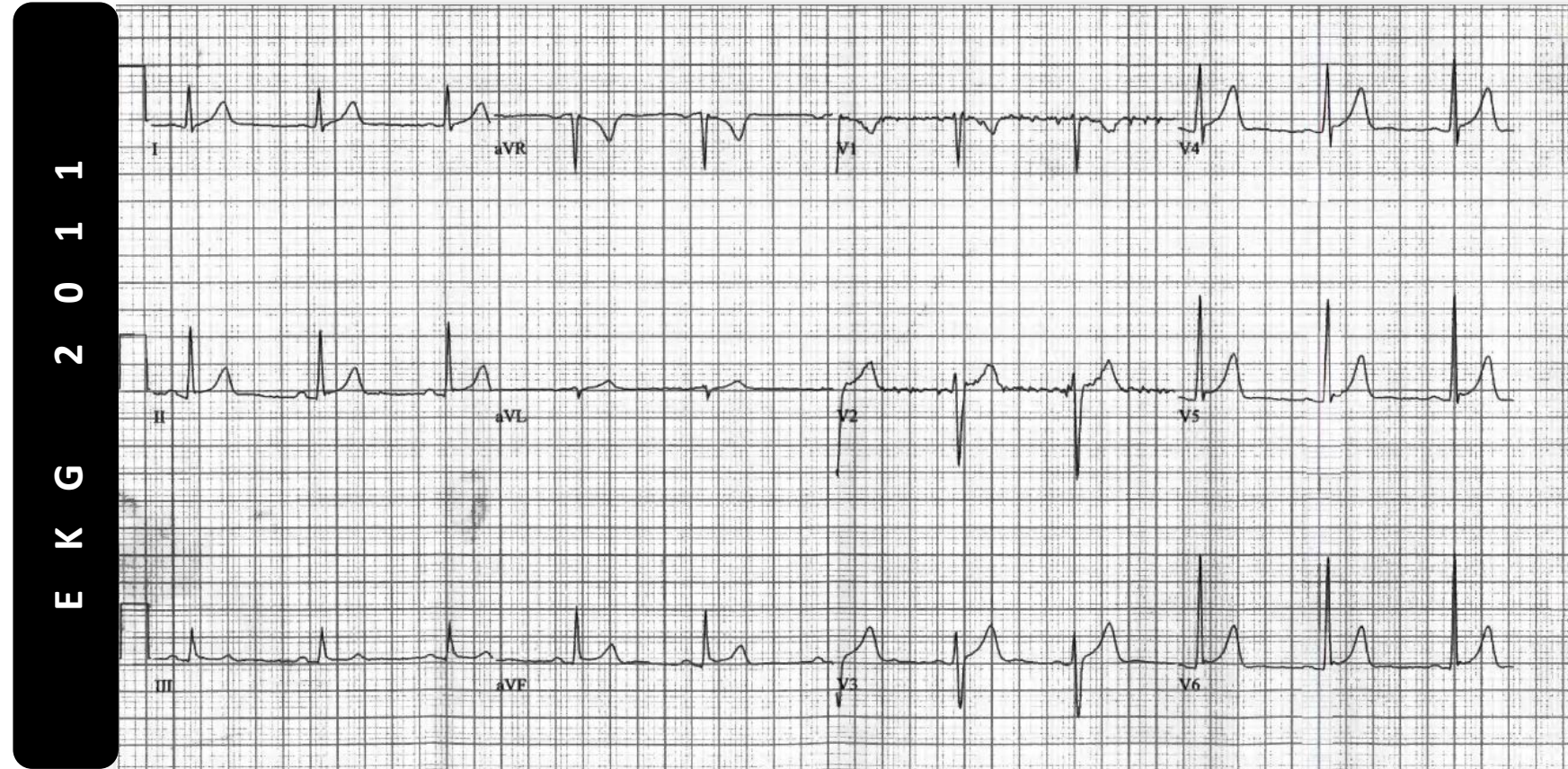
Father 7th decade, moderate coronary disease

## Exam

- BP 127/70 mmHg HR 88
- RR 12 Temp 99.3 degrees
- Pain free
- JVP not elevated
- Carotids normal
- Clear lungs
- Pectus excavatum
- Normal S1, click not heard
- Physiologically split S2
- TrT 1.41 -1.79 mg/dl (0.01)
- CK 1570-900
- Echo: IPL hypo, EF 55%, no effusion

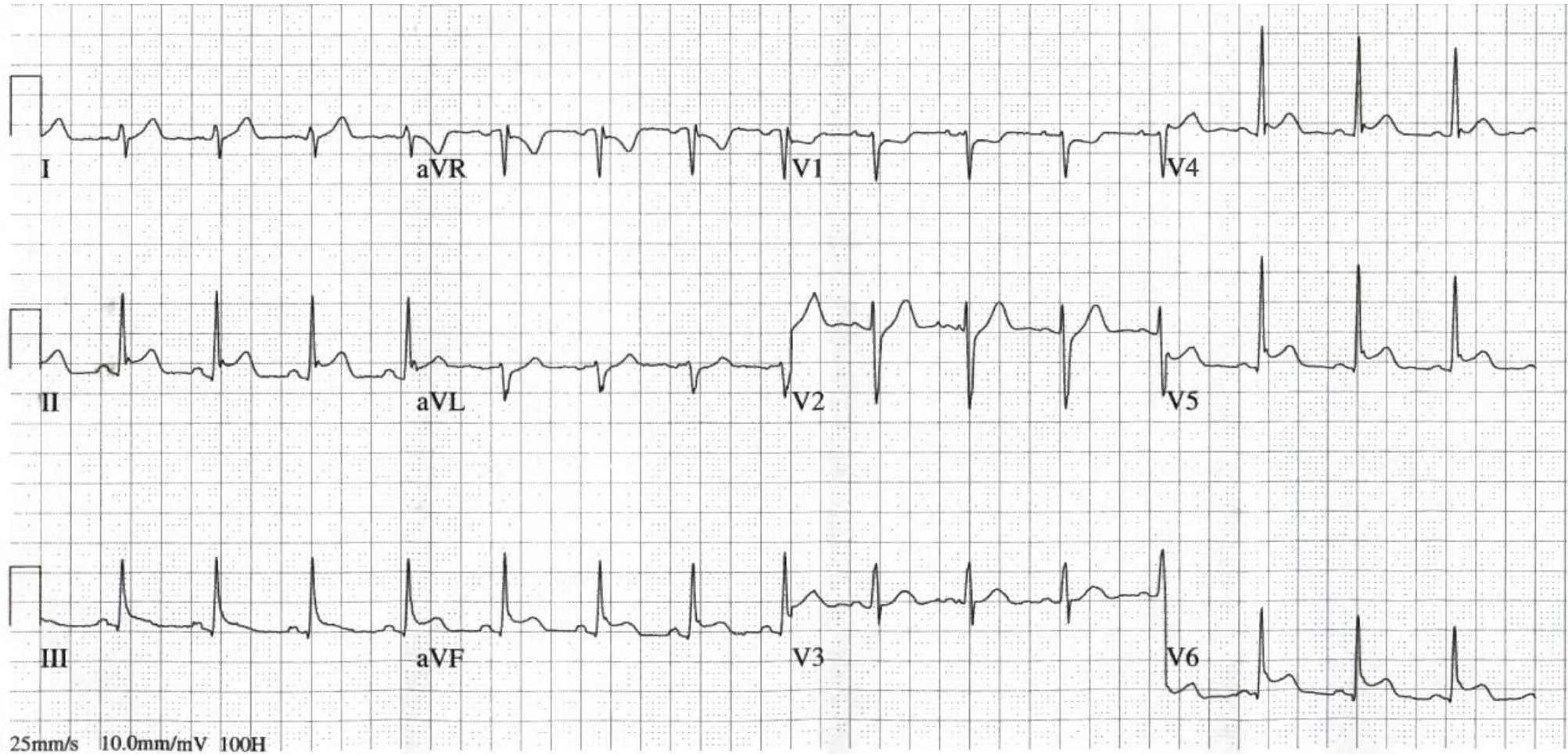


# Case Study 322, 3 years prior





# Case Study 322



# Next Steps For This 21 Yo Man True or False

A

Conservative mgmt with colchicine, nsoids, for acute pericarditis.  
No further work up needed.

Immediate coronary angiography, for STEMI, that includes ST segment elevation, elevated Tr, wall motion abnormality.

B

C

Given age and prodrome, CMR looking for evidence for myocarditis.

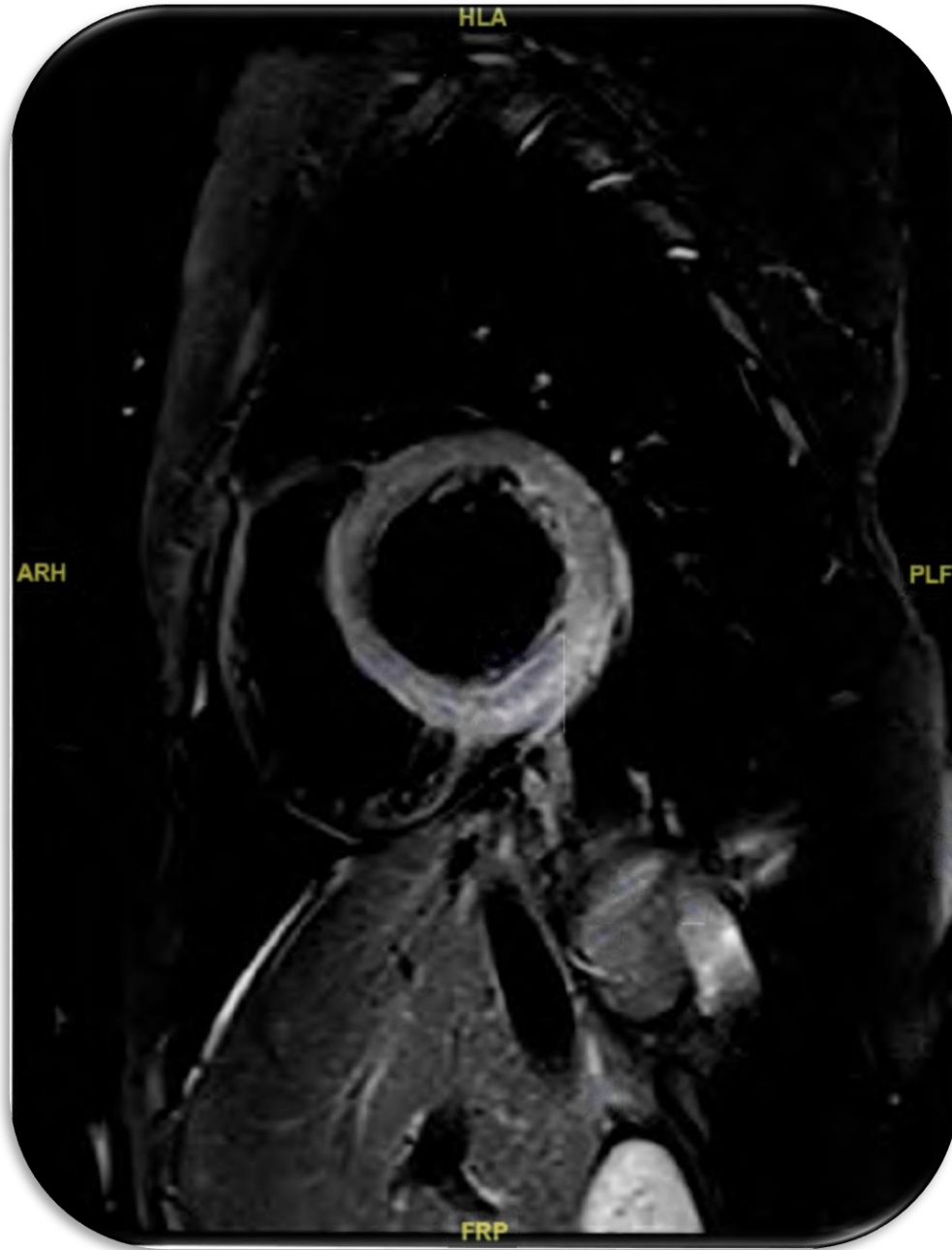
If CMR pattern suggests infarct or ischemia, CT angiography should be done.

D

E

Prognosis is poor, if myocarditis, given EF.

# Case Study 322



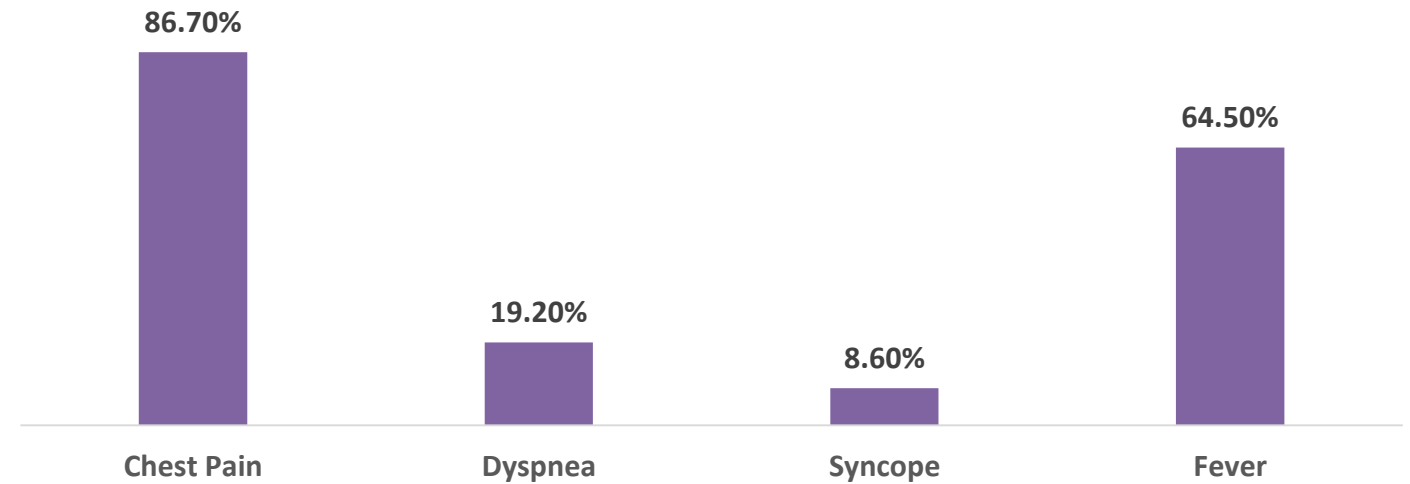
M R I 2 0 1 6

# Outcome of Acute Myocarditis, hinges on

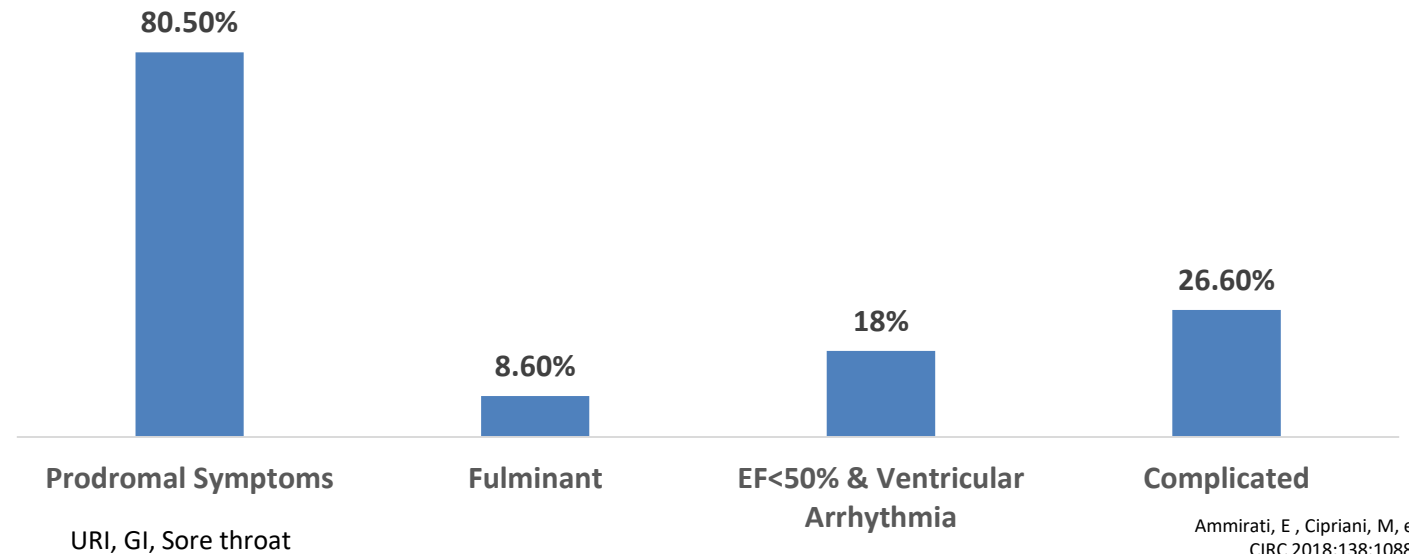
“Complicated” or not. Complicated =

EF < 50%, sustained VA, or

Fulminant: Requiring inotropes +/- mechanical circulatory support



443 Patients



# 5 Year

Mortality &  
Heart  
Transplants

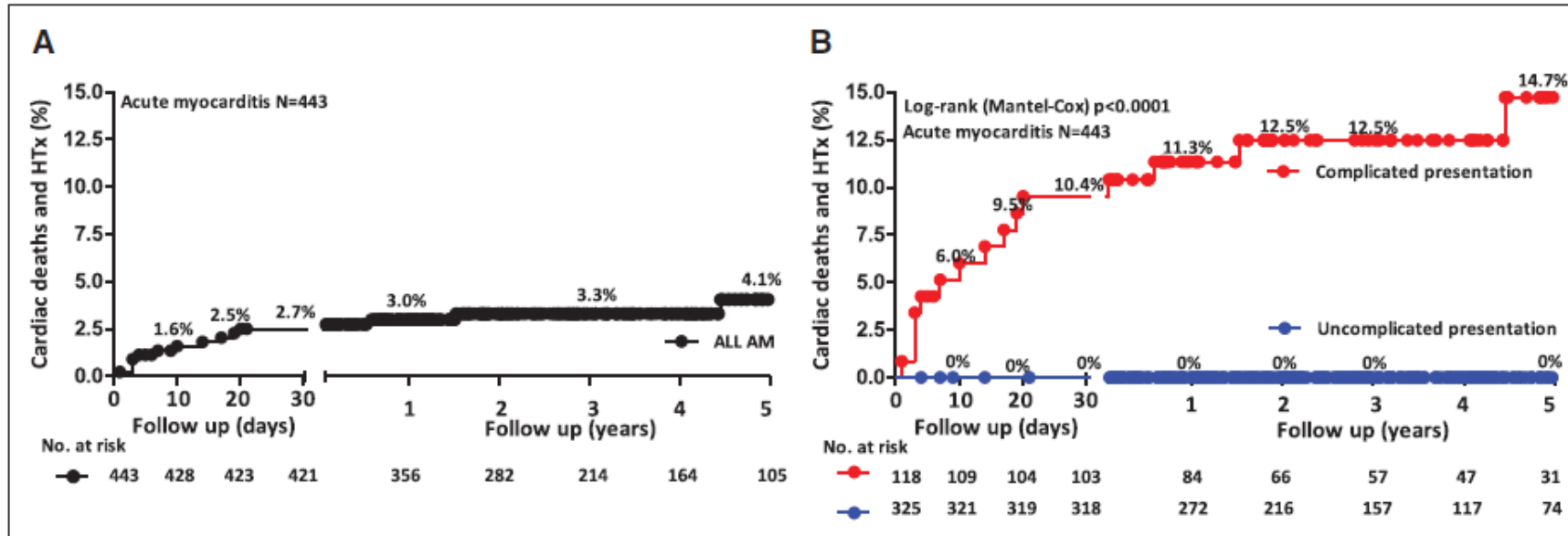
4.1%

Only seen in  
patients with  
complicated  
presentation

1 yr EJECTION  
FRACTION < 50%

1/145  
Uncomplicated

8/55  
Complicated  
(14.5%)

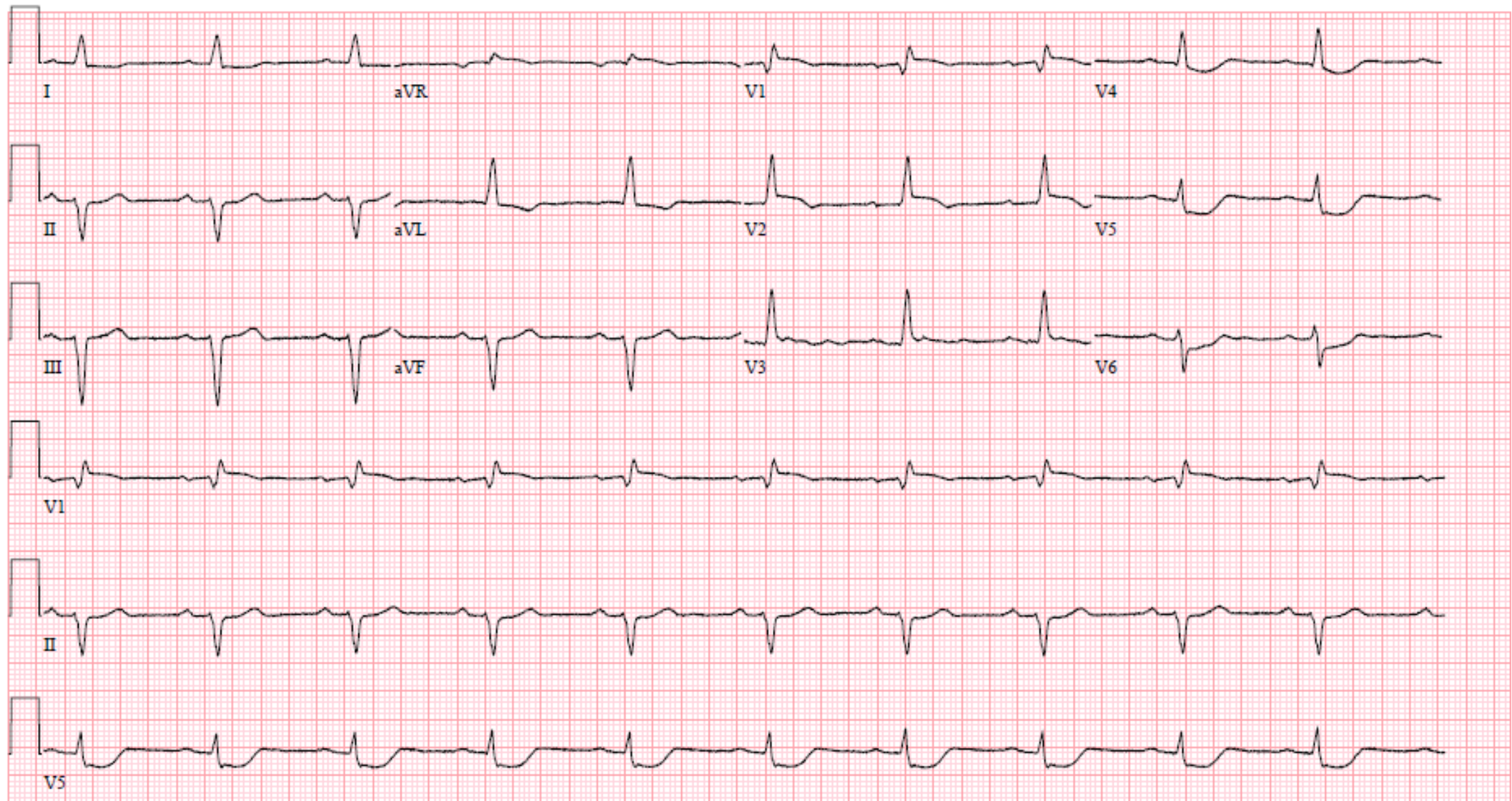


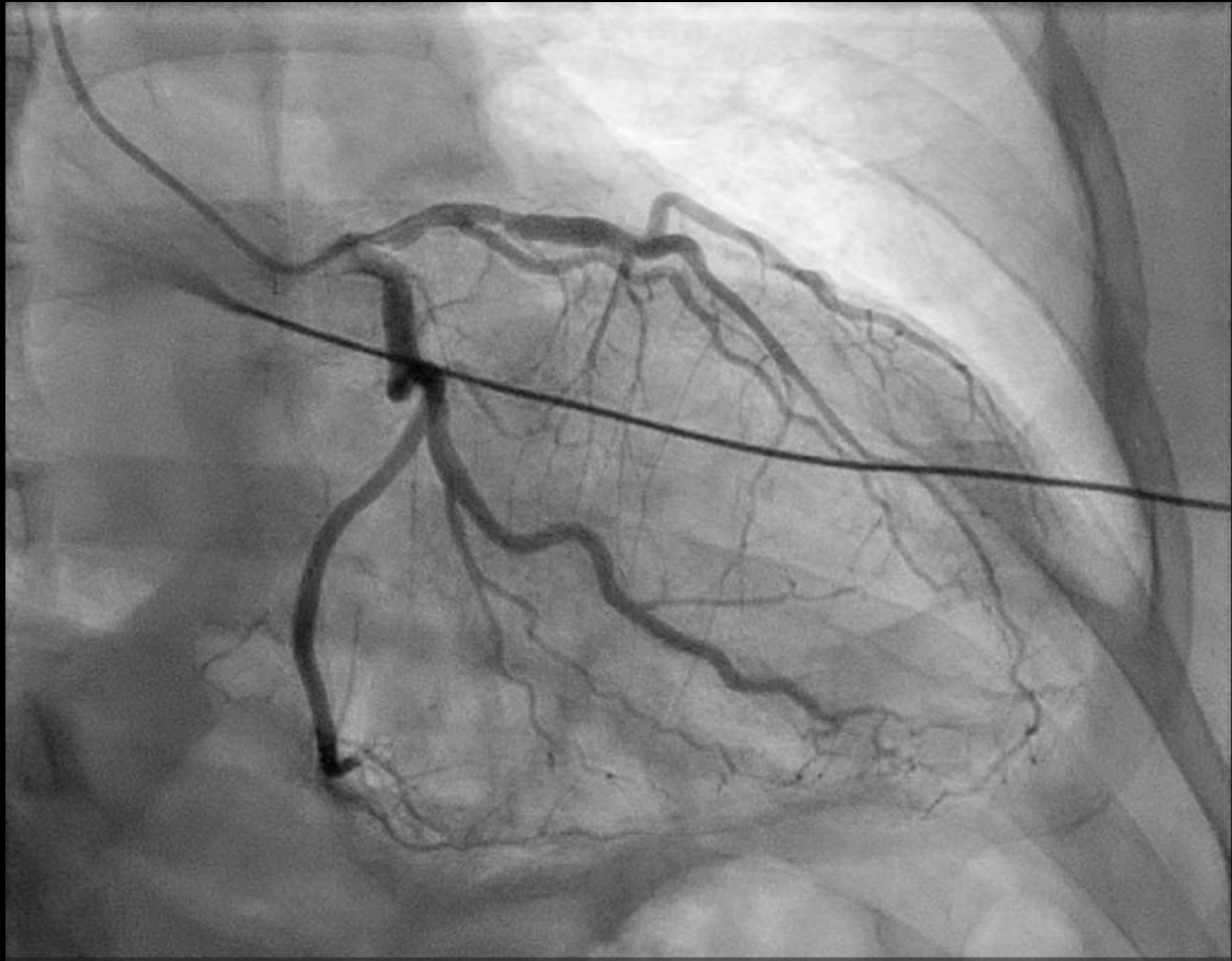
**Figure 2.** Kaplan-Meier estimates of 5-year cardiac mortality and heart transplantation (HTx).

**A**, Events in the whole study population with acute myocarditis (AM) and **(B)** events in patients with AM complicated at presentation by left ventricular ejection fraction <50%, sustained ventricular arrhythmias (VAs), or a low cardiac output syndrome compared with patients without such complications at presentation. Of total cardiac deaths and HTx, 16 patients initially presented with fulminant presentation and 1 with VAs. One cardiac death occurred after HTx and was omitted.



# 57 y.o. With Second Course Immune Check Point Inhibitor. Double Vision & Chest Pain

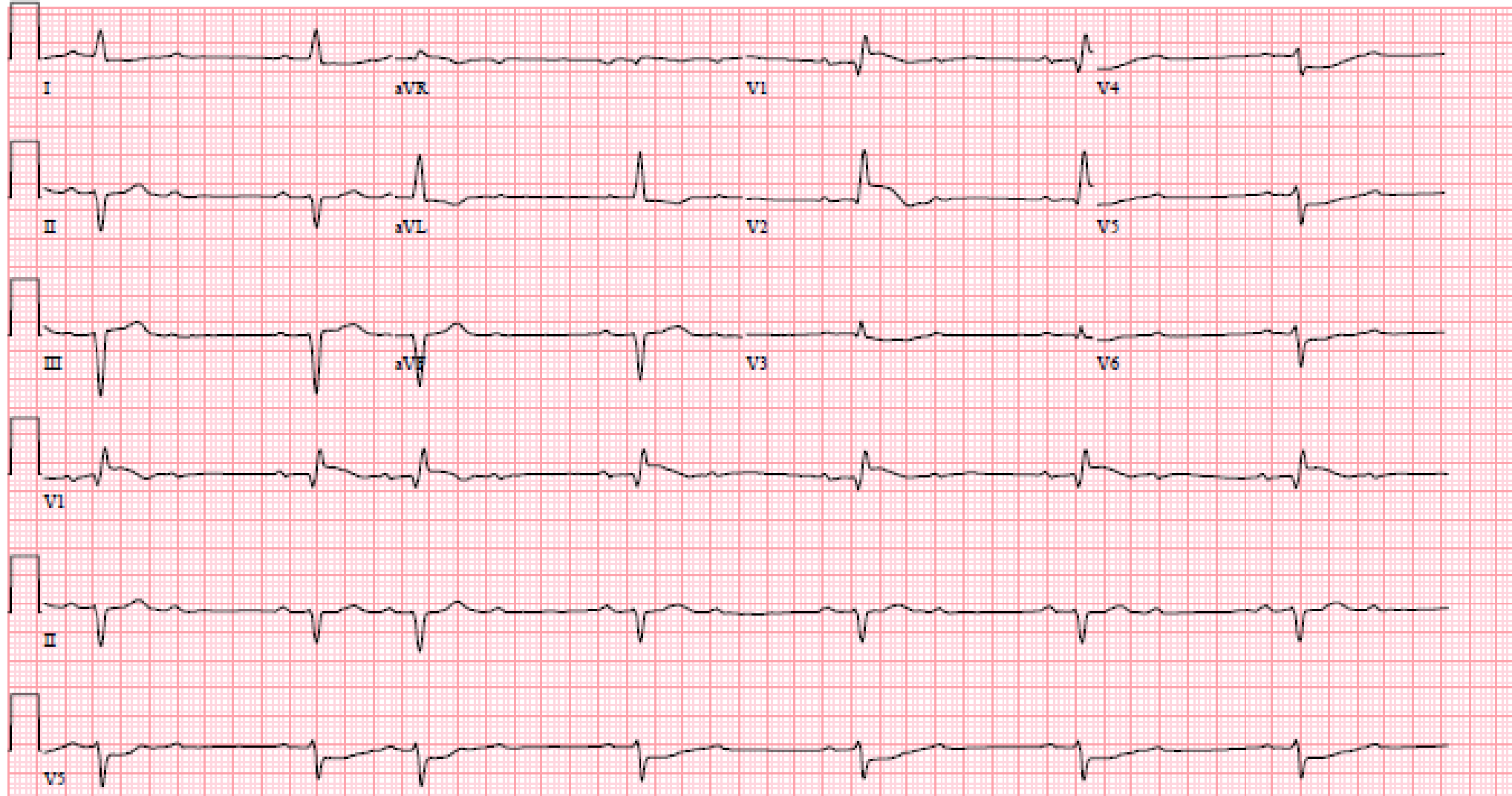




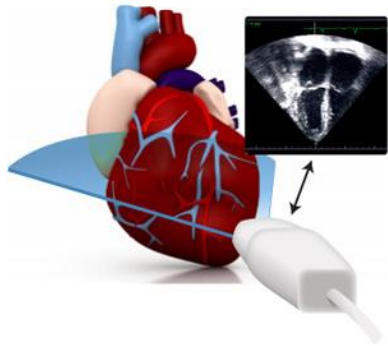
# **LAD Stent.**

## **ST Elevations & High Degree Heart Block.**

### **Diagnosis?**



# Case Study 085



Echo

- EF 65%
- Septal thickness 2.3 cm
- Posterior wall 1.3 cm

No history of hypertension

## Family History



SCD in 18 year old  
grand daughter of  
father



55

Year Old  
Woman

## Holter



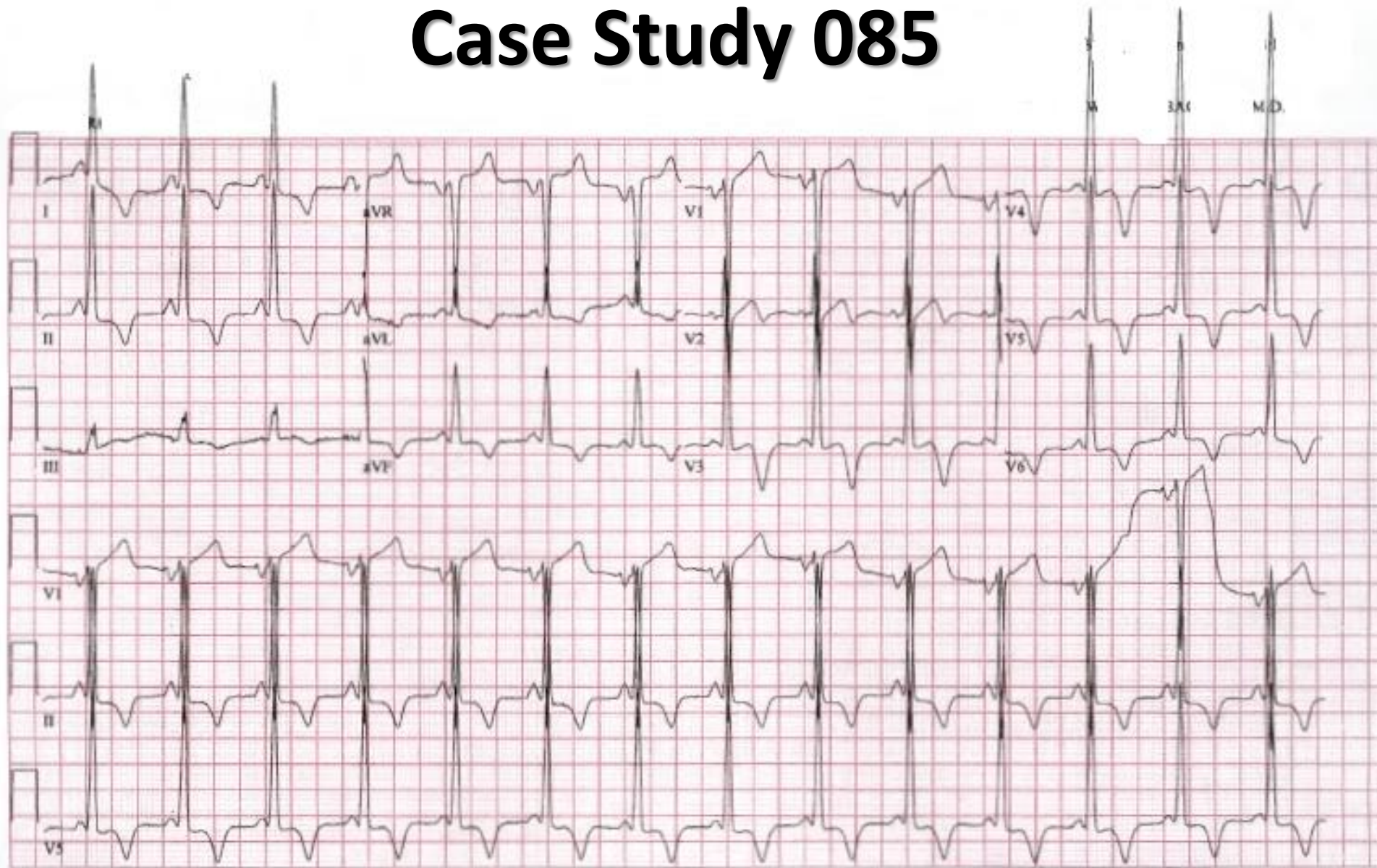
- 8-10 beat runs of ventricular tachycardia,
- rate 170,
- clear-cut AV dissociation
- RBBB
- Superior axis configuration



Exertional  
dyspnea and  
heart pounding



# Case Study 085





# Case Study 085



# **Case Study 085**

## **Which of the following is true?**

**A**

Ventricular ectopy with this EKG, good EF: can be managed with a Beta-blocker and reassurance

**B**

This patient has an infiltrative process and should undergo EMB, looking for sarcoid or amyloid.

**C**

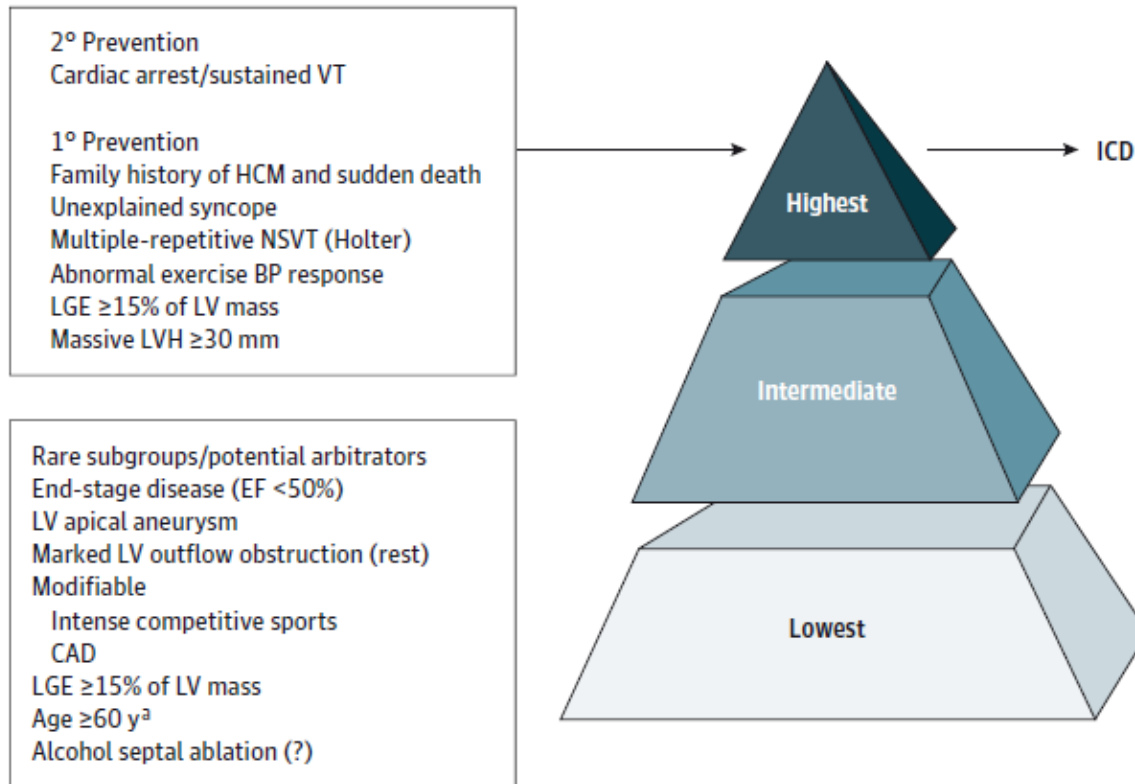
The combination of verapamil and Beta-blockers may be used to quiet the ectopy and she can be followed conservatively.

**D**

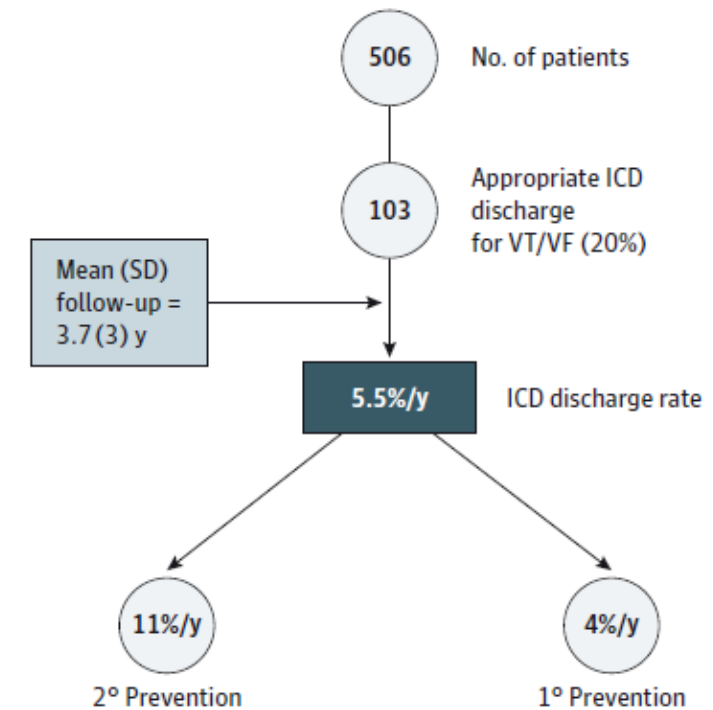
This patient has an excellent story for hypertrophic cardiomyopathy and an MRI might be helpful.

# Stratification of Hypertrophic Cardiomyopathy Patients

**A** Risk stratification model (ACC/AHA)



**C** Flow diagram of 506 high-risk patients treated with ICD



# Ontario: 95% of people > 45 y.o. with SCD undergo autopsy

## Cases of HCM with SCD

44  
Definite

3  
Probable

6  
Possible

Estimated annual incidence of SCD: 0.31-0.39/1000 HCM years

Estimate of HCM related SCD and aborted arrest, ICD discharge: 0.84/1000 HCM years

70% SCD in previously undiagnosed: 64.8% occurred during rest  
18.5% occurred during light activity

*Well below the discussed risk of 0.5-1% per year*

### Autopsy Findings:

Mean maximal cardiac wall thickness  $2.54 \pm 0.8$  cm

>3cm 31%

>2.5 cm 50%

57% asymmetric septal hypertrophy

# **Hypertrophic Cardiomyopathy with increased myocyte size, disarray and fibrosis. Alteration in 8 genes.**

- Usually, missense
- Resultant alteration in  $\text{Ca}^{++}$  cycling and  $\uparrow$  actin-myosin binding change in configuration from the binding activates TGF $\beta$  and  $\beta$  signaling and fibrosis
- Resultant hypertrophy and fibrosis and HCM phenotype



# Interventions

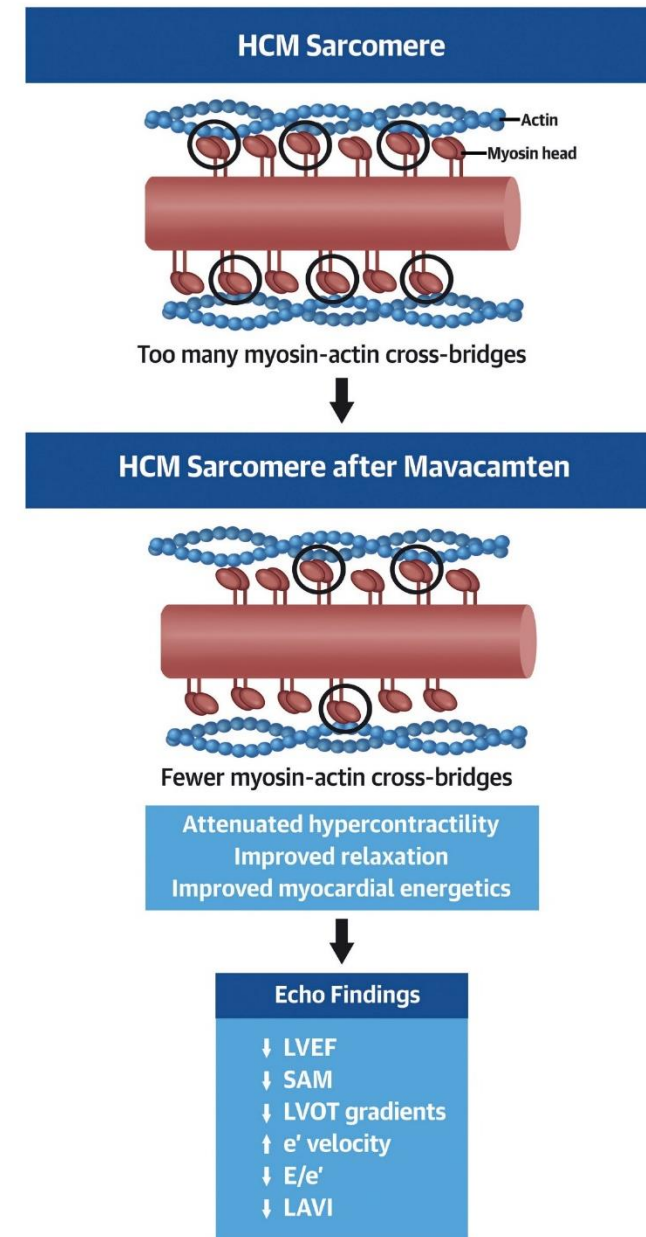
## Valsartan or Losartan to inhibit TGF $\beta$ signaling

- Patients with no phenotype evidence or minimal doppler abnormality

## Mavacamten

- Patient with LVH, SAM, diastolic abnormality
- Inhibit actin-myosin binding

## CENTRAL ILLUSTRATION: Mechanism of Action of Mavacamten and Observed Changes



# EXPLORER HCM – Study Rationale

Obstructive Hypertrophic Cardiomyopathy Class II or III



At end of 30-week treatment period

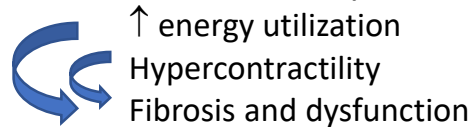
## Primary Endpoints

Peak VO<sub>2</sub> ↑ 1.5 ml/kg/min  
and  
↓ NYHA by 1 class

## Secondary Endpoints

Post exercise LVOT gradient  
NYHA  
Peak VO<sub>2</sub>  
KCCQ

- Gene mutations promote excessive myosin—actin binding



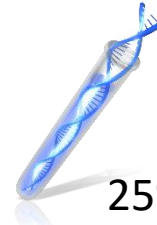
- Mavacamten inhibits cardiac myosin ATPase; restores proportion of super relaxed myosin, ↓ cross bridges, normalizes ATP consumption



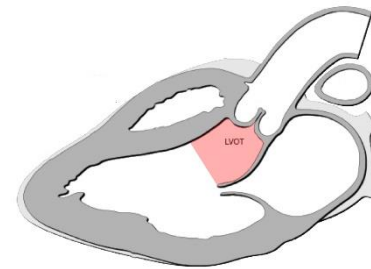
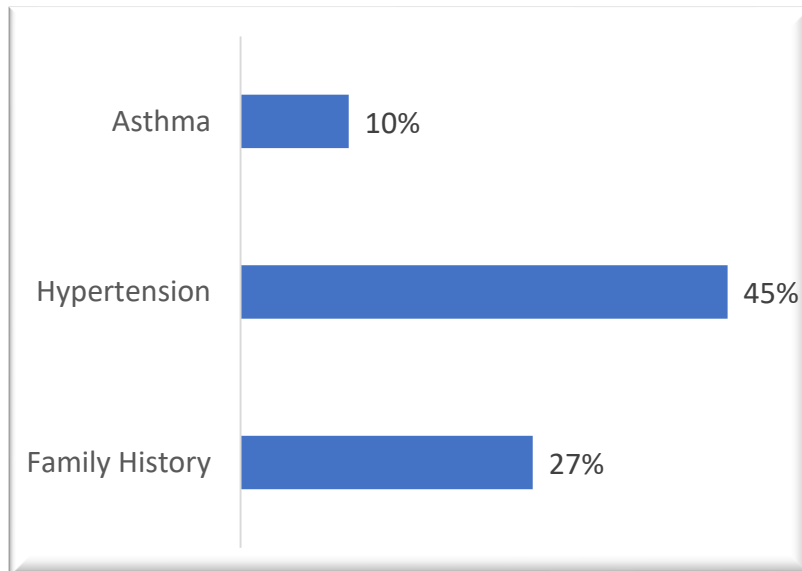
# EXPLORER-HCM



Age 58  
Caucasian 90%  
BMI 29



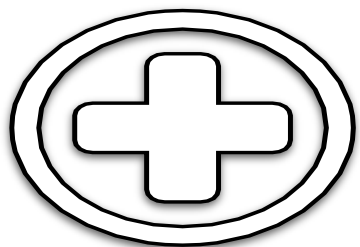
Genetic testing 73%  
25% of those tested had variant



22% ICD  
B-Blocker ~75%  
Resting LVOT 52  
Valsalva 72  
Post exercise 86



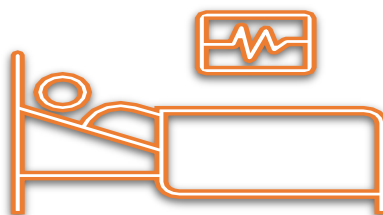
# EXPLORER-HCM



68 Centers



13 Countries



251 patients  
429 patients assessed

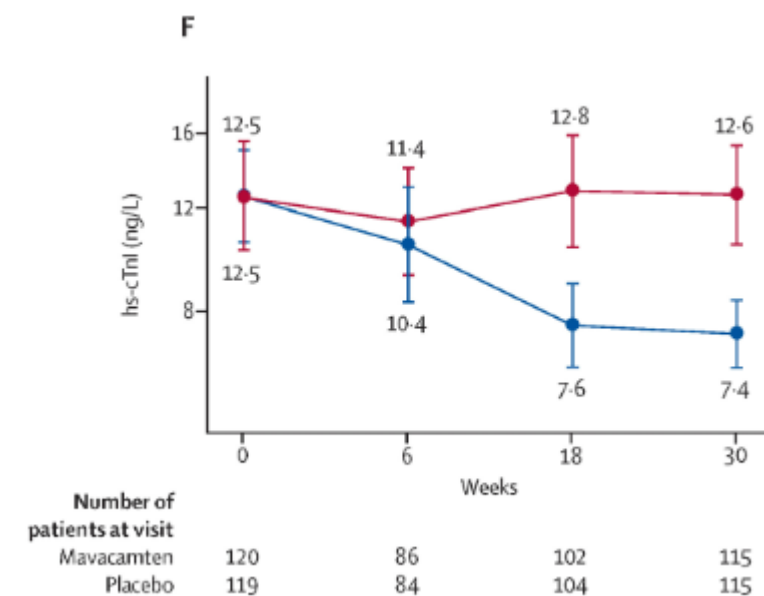
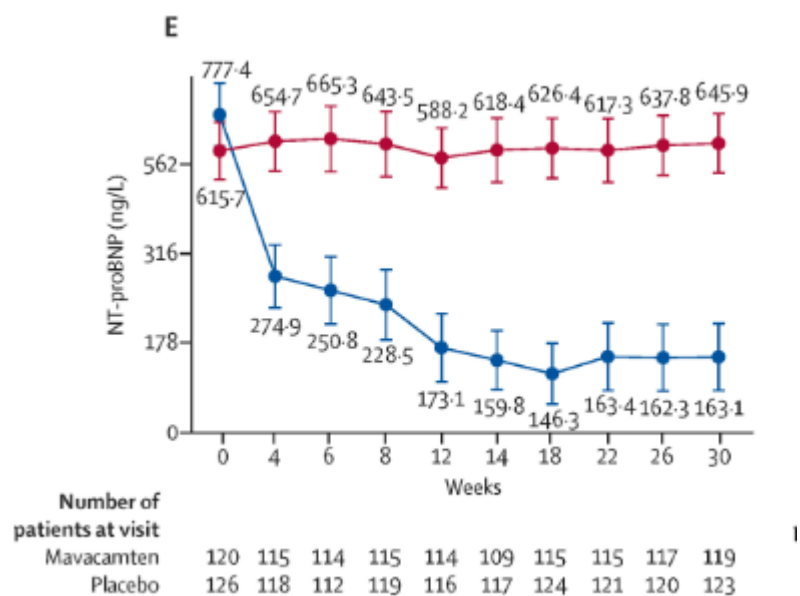
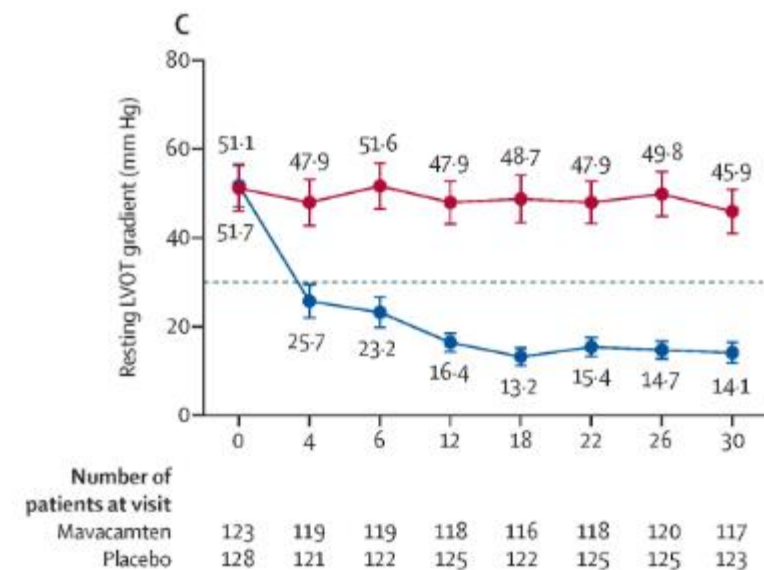
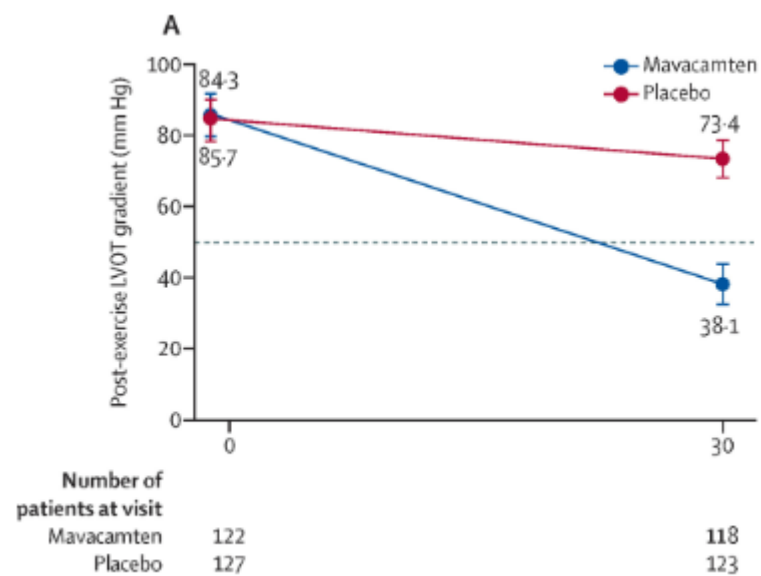


Mavacamten  
vs  
Placebo

Compared with placebo,  
Post exercise LVOT -36 mmHg  $p=0.0001$   
PVO2 +1.4 mmHg  $p<0.0001$   
KCCQ + 9.1  $p=0.0006$

## Primary Endpoint

37% vs 17%  $p=0.005$   
2/3 vs 1/3 improved by 1 NYHB  
Heavier, already on b-blockers had less improvement





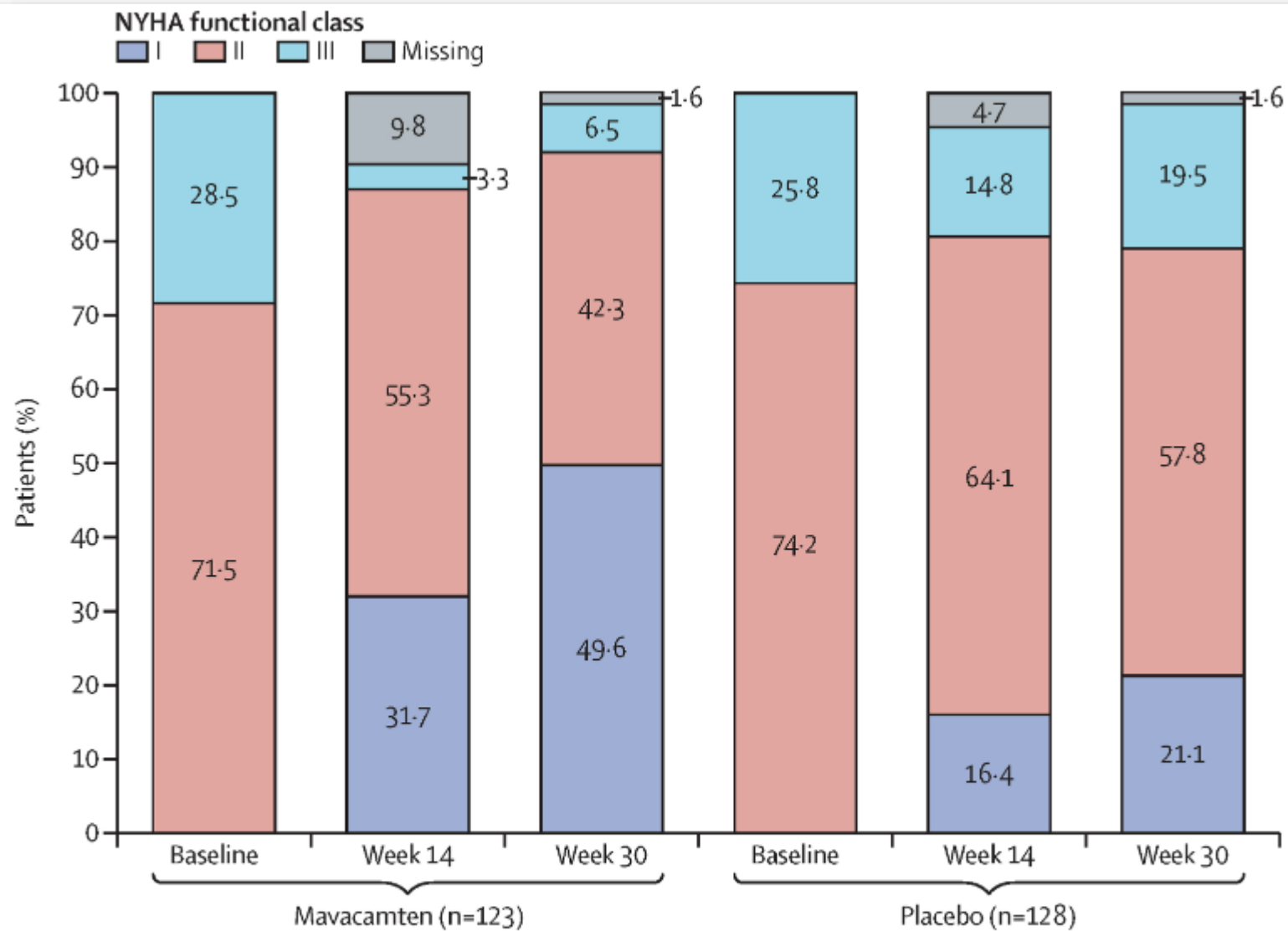


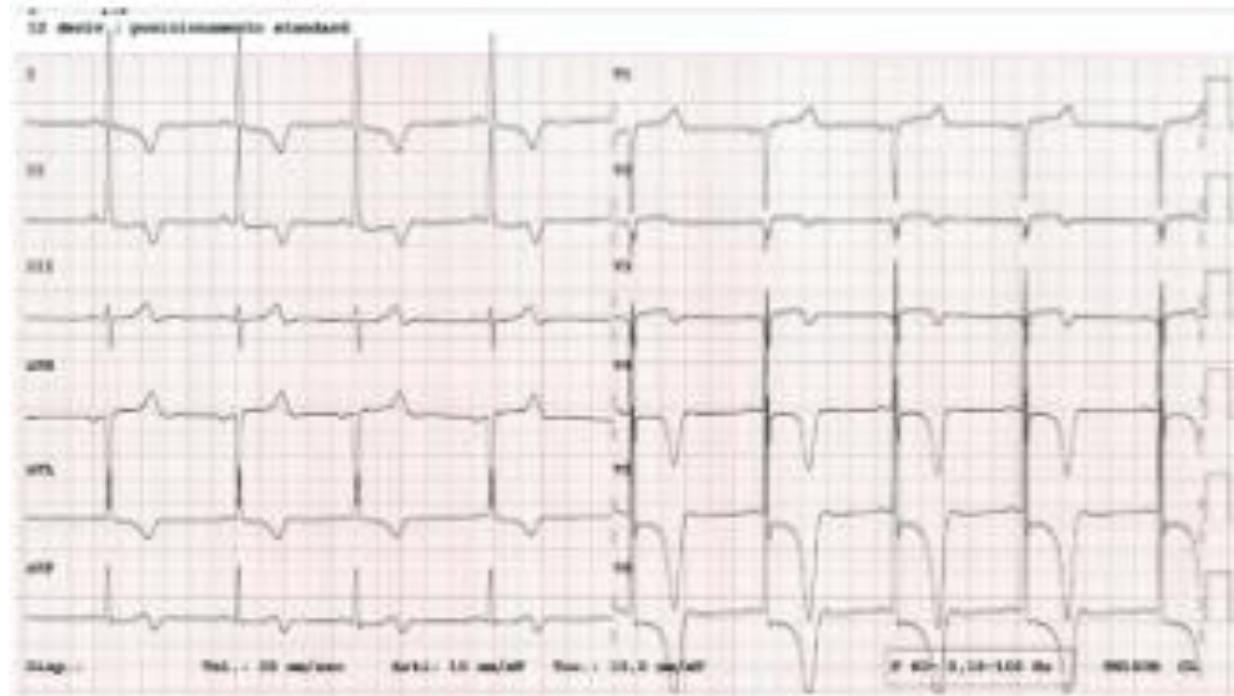
Figure 2

NYHA functional class

Percentage of patients who had NYHA class I, II, or III at baseline, after 14 weeks and 30 weeks of treatment, for the mavacamten and placebo groups. NYHA=New York Heart Association.

# Can look Like HCM

So we have to think of “it”



# Fabry's Disease

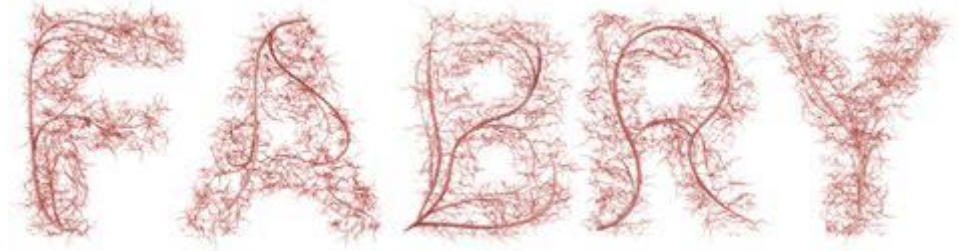
Mimic of hypertrophic cardiomyopathy

May represent 0.93% of HCM



Pan ethnic, x-linked lysosomal storage disease

1/8800 newborn



GLA gene defects  
Reduced  $\alpha$ -galactosidase ceramide  
(Gb3) and globotriasosylsphingosine (lyso-Gb3)

# Heart, Kidney, CNS

- Later onset if there is some production of  $\alpha$ -Gal and may pr... heart

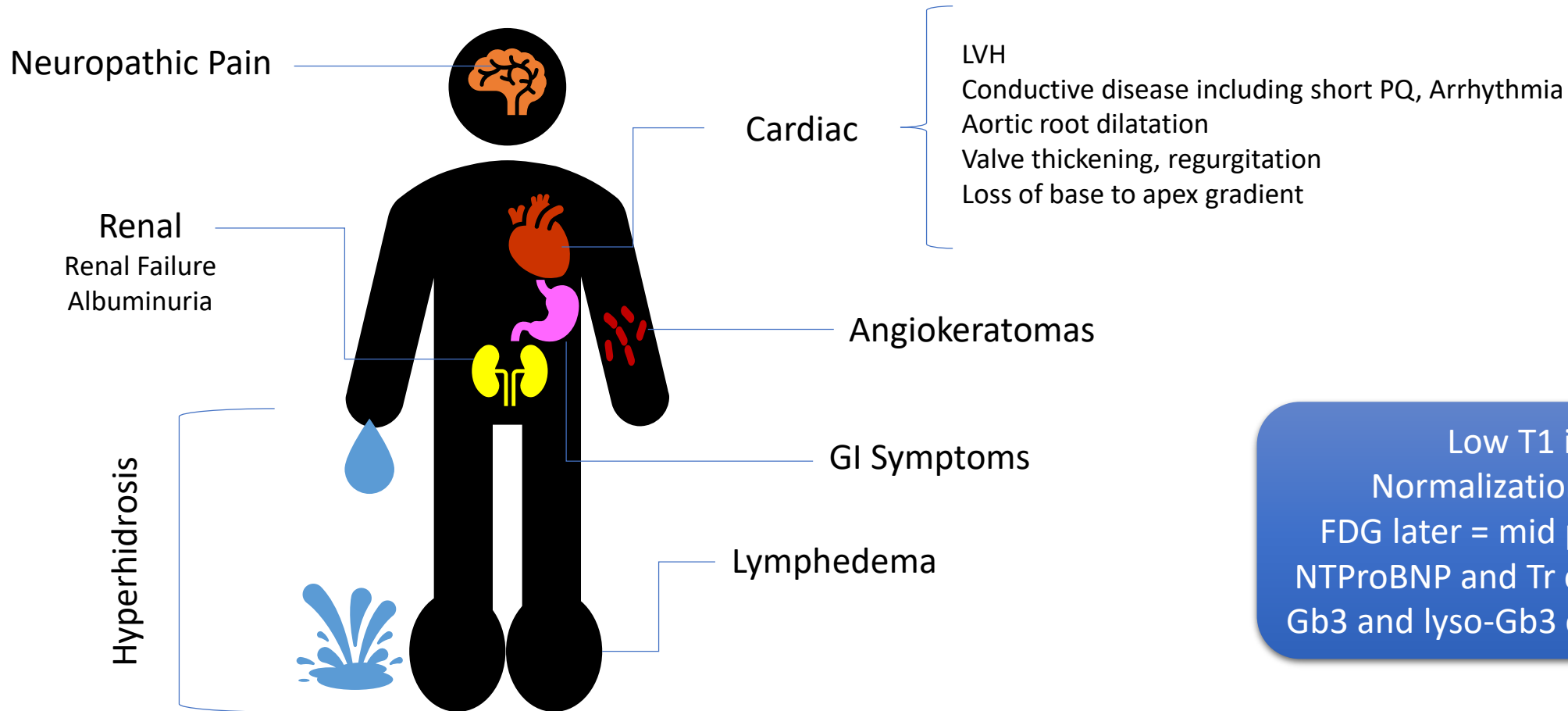
N215S:	F113L:	IVS4+919 G>A:
<ul style="list-style-type: none"><li>• North America</li><li>• Europe</li></ul>	<ul style="list-style-type: none"><li>• Portugal</li></ul>	<ul style="list-style-type: none"><li>• Taiwan</li></ul>

- Woman xx, so some cells under express, some by .....

Diagnosis

- Males, low  $\alpha$  Gal, still want genetics
- Females, may have normal levels, need genetics

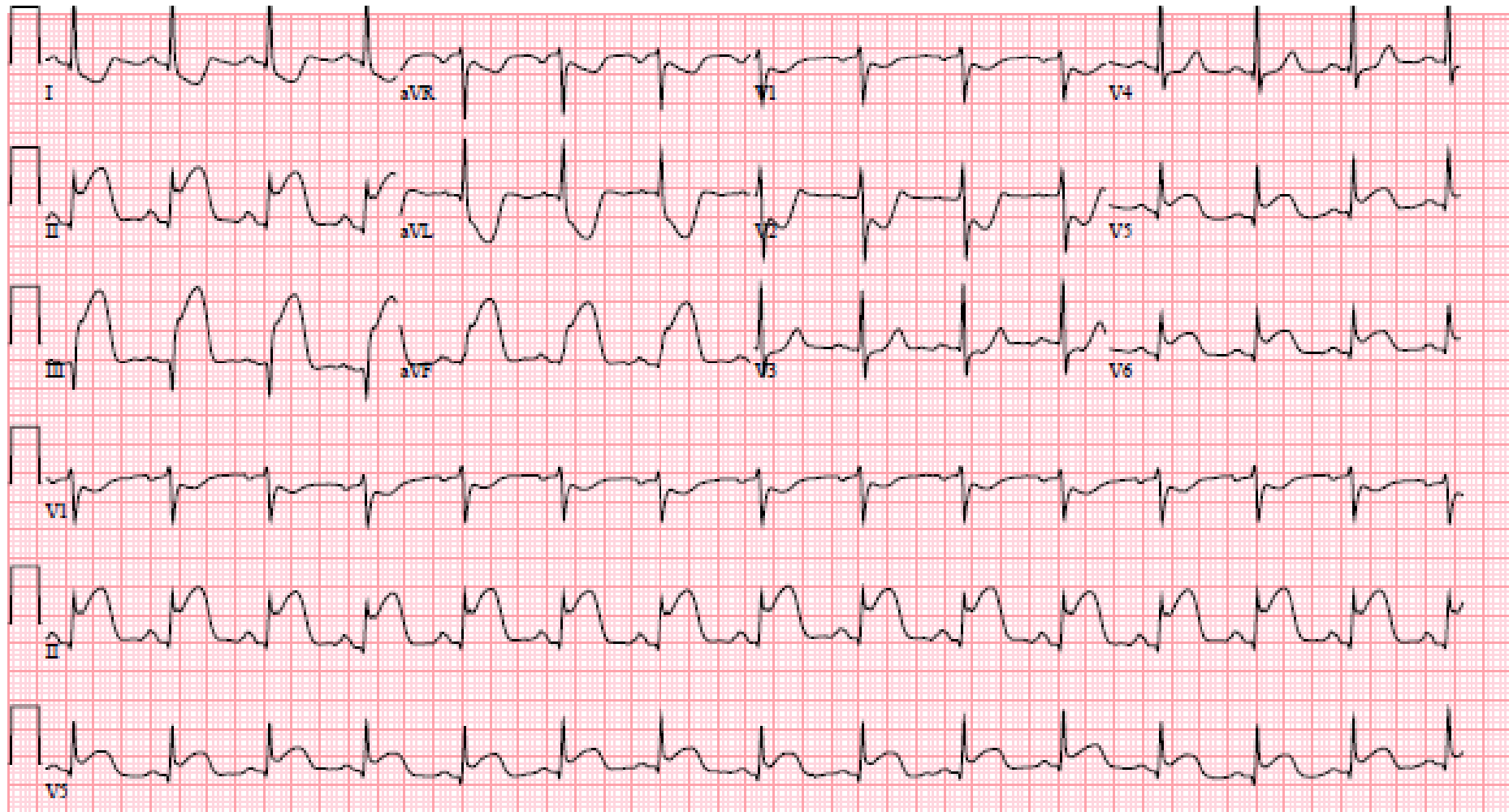
# Fabry Disease Red Flags Cerebrovascular and Cardiac



Low T1 initially  
Normalization of T1 later  
FDG later = mid post lateral LGE  
NTProBNP and Tr elevation with T2  
Gb3 and lyso-Gb3 cause inflammation

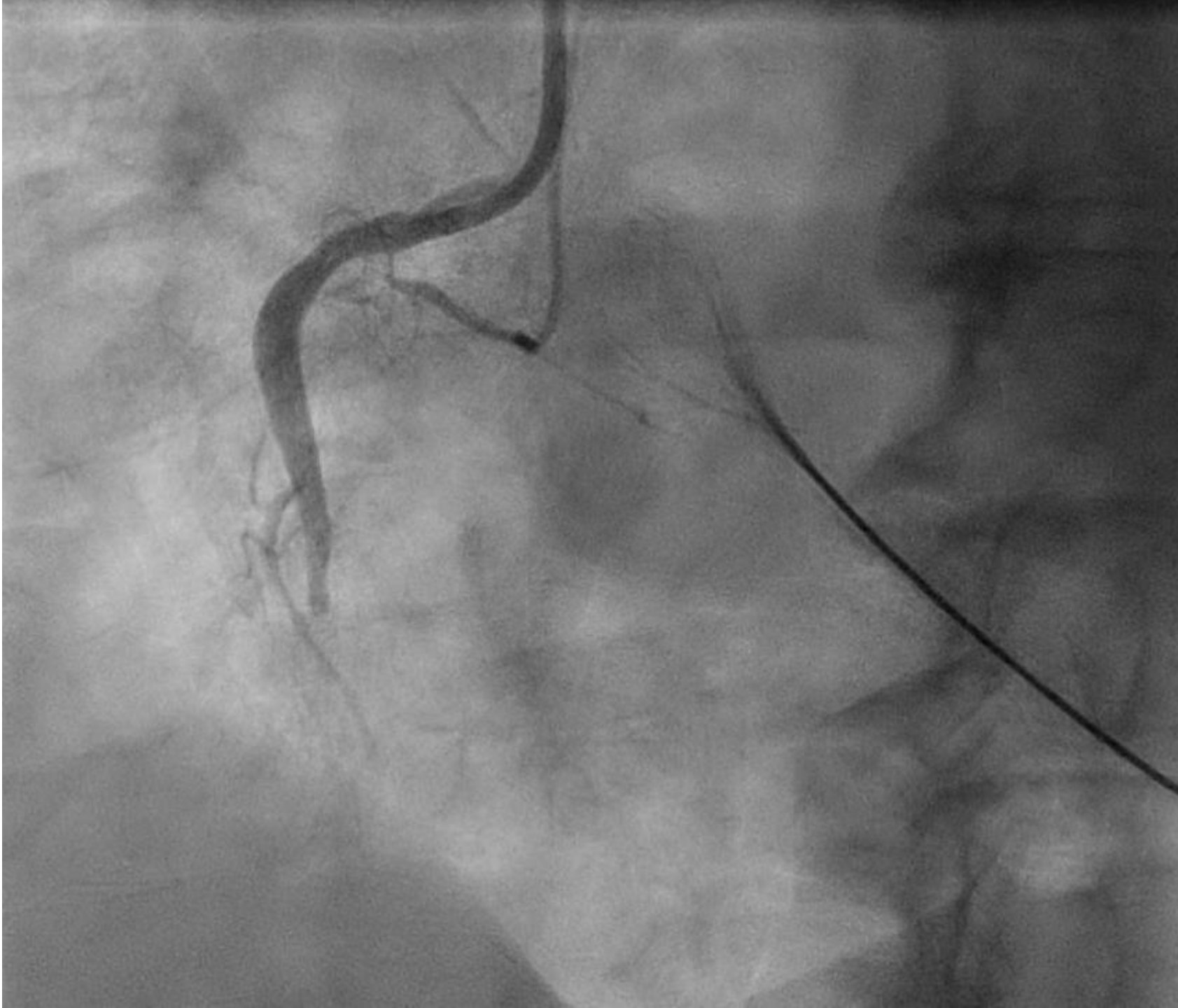


## 76 Yo Woman with One Hour of Chest Pain.



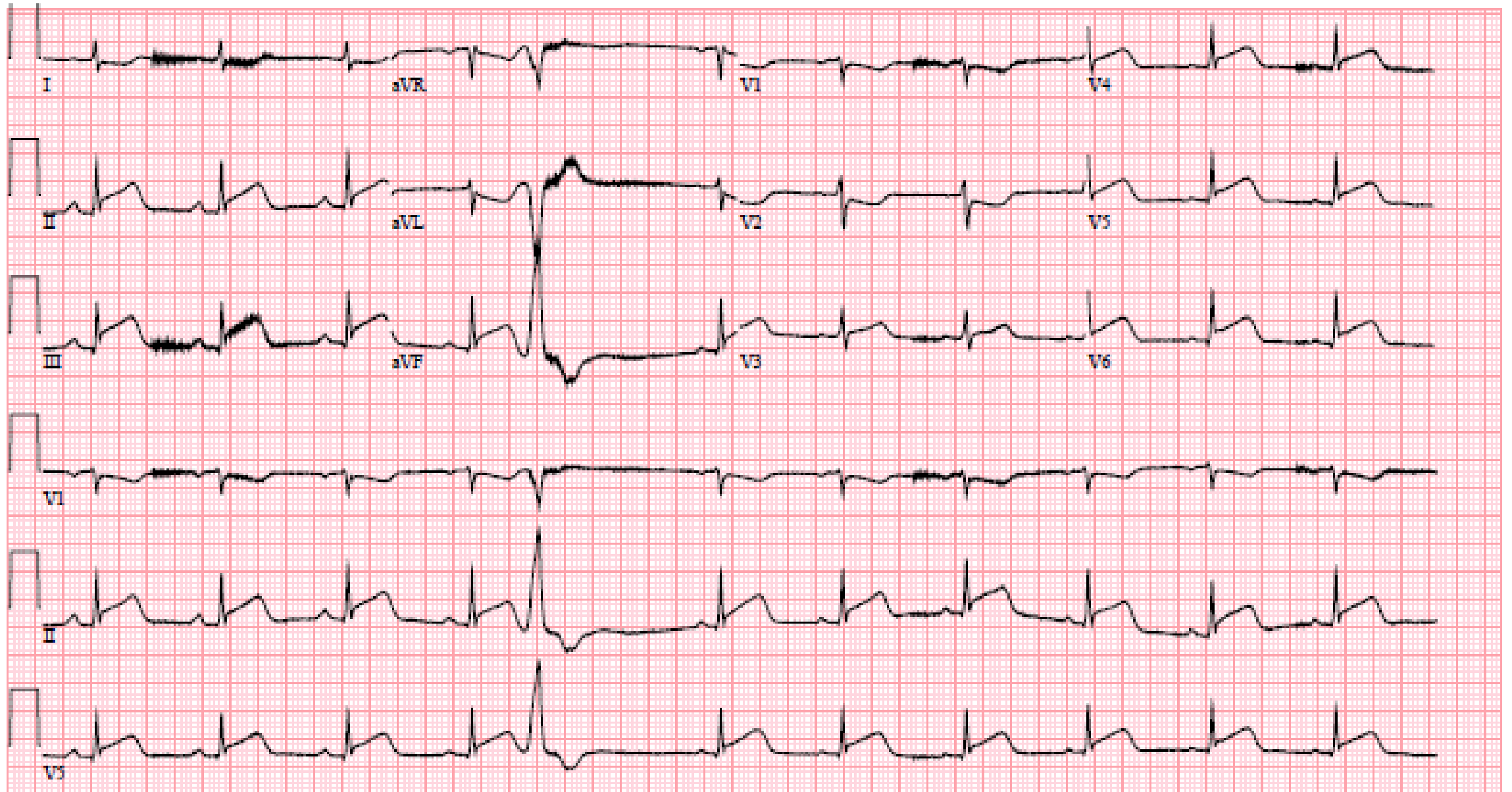
**Where Is The Lesion?**

**76 y.o.  
Woman  
with 1  
Hour of  
Chest  
Pain:**



**The lesion is  
after the RV  
branches.  
Thus right  
precordial  
leads ST  
segments are  
not being  
pulled up.**

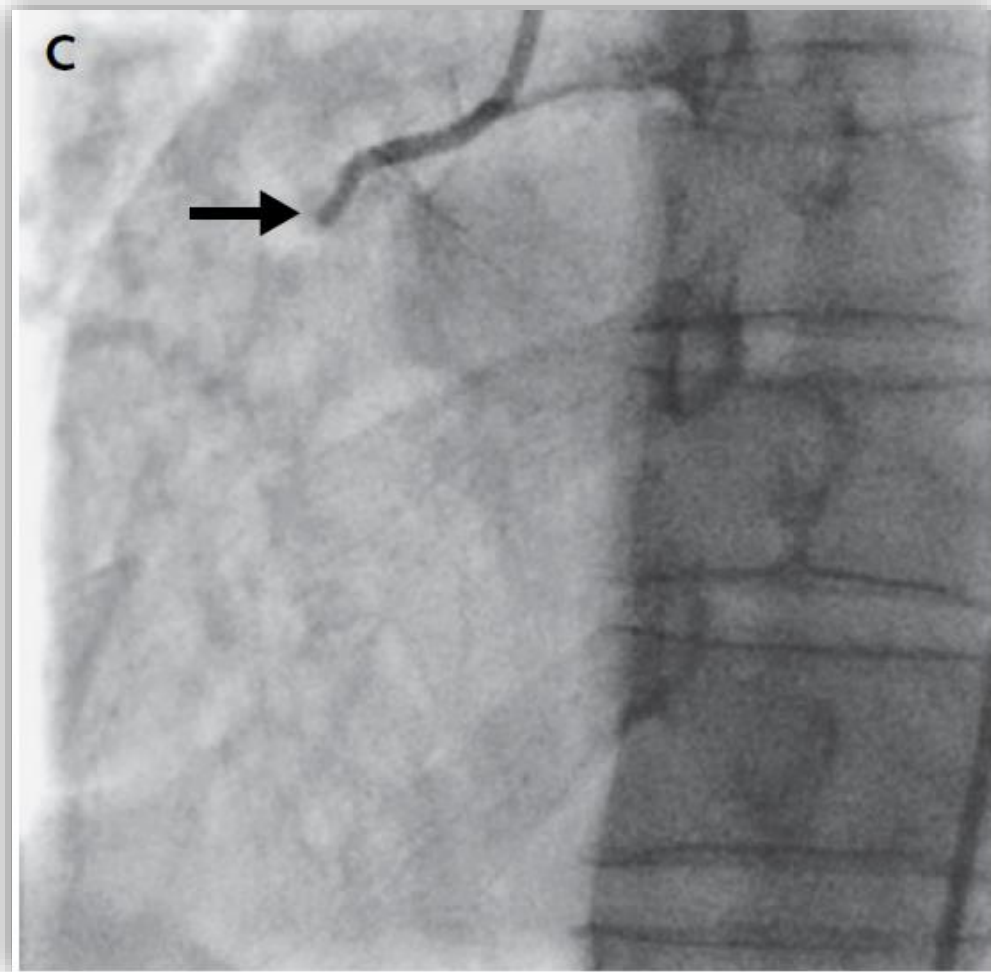
**65 yo Woman with 1 Hour Of Chest Pain,  
BP 95/70, Neck Veins Elevated. Where Is Lesion? Does The VPB Help?**



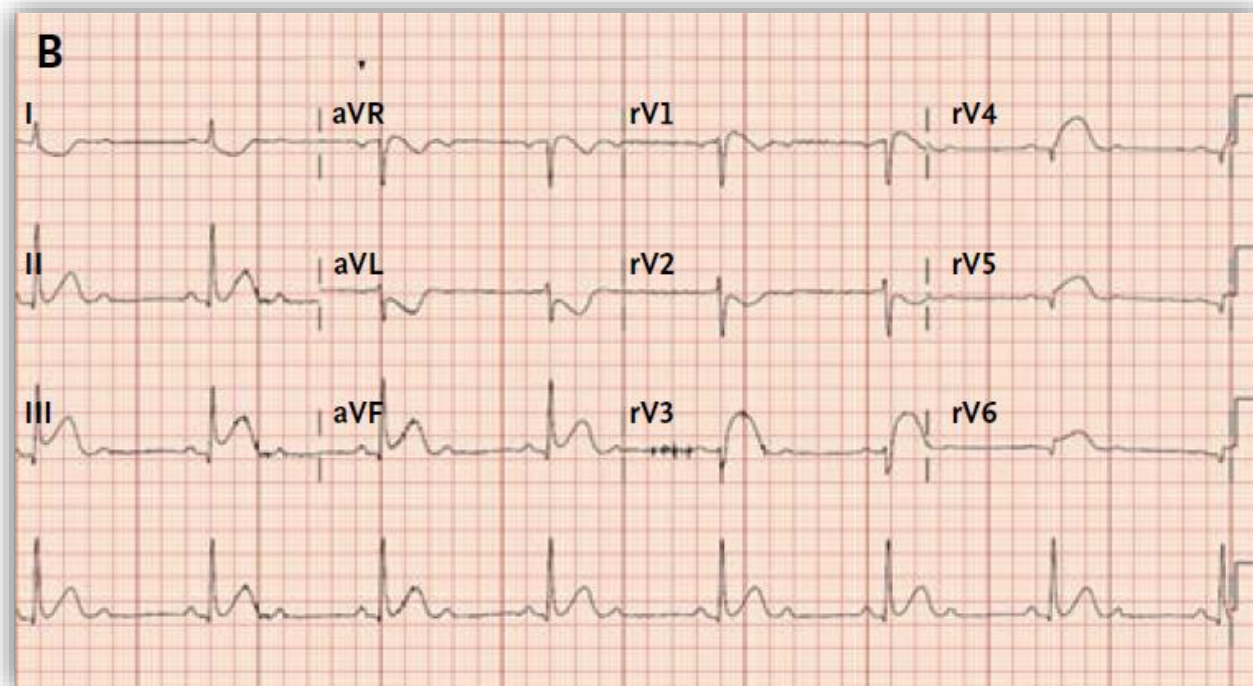
**65 yo  
Woman  
with 1  
hour of  
Chest  
Pain:**



low BP,  
more  
inferior ST  
elevation  
than  
posterior ST  
depression  
RV supplied  
by acute  
marginal  
branches is  
ischemic



**Case Study JD-61-what  
would V2-V3 look like?**





# Case Study

## 61 Year Old Man



- develops severe chest – back – abdominal pain and profound diaphoresis, then feels slightly better

## Current Medical Hx



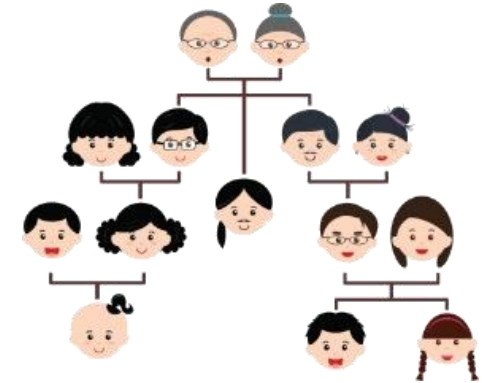
- Longstanding HR in 50's
- ↑LDL on atorvastatin 10 mg daily
- History of hypertension on lisinopril 10mg each day

## Other Medical Hx



5 years ago, while involved in a heavy weight-lifting program, developed a right rotator cuff tear and a bicipital tendon rupture

## Family History



- Father underwent CABG in 7th decade and lived to 9th decade
- Mother well in 9th decade
- 1st cousin underwent CABG in 6th decade
- 4 children all well
- No family history of aortic disease

# Case Study

---



- 71" tall, 171 pounds
- 167/93 mmHg (both arms no paradox)
- HR 55 bpm, RR 18
- Unable to get comfortable
- Diaphoretic

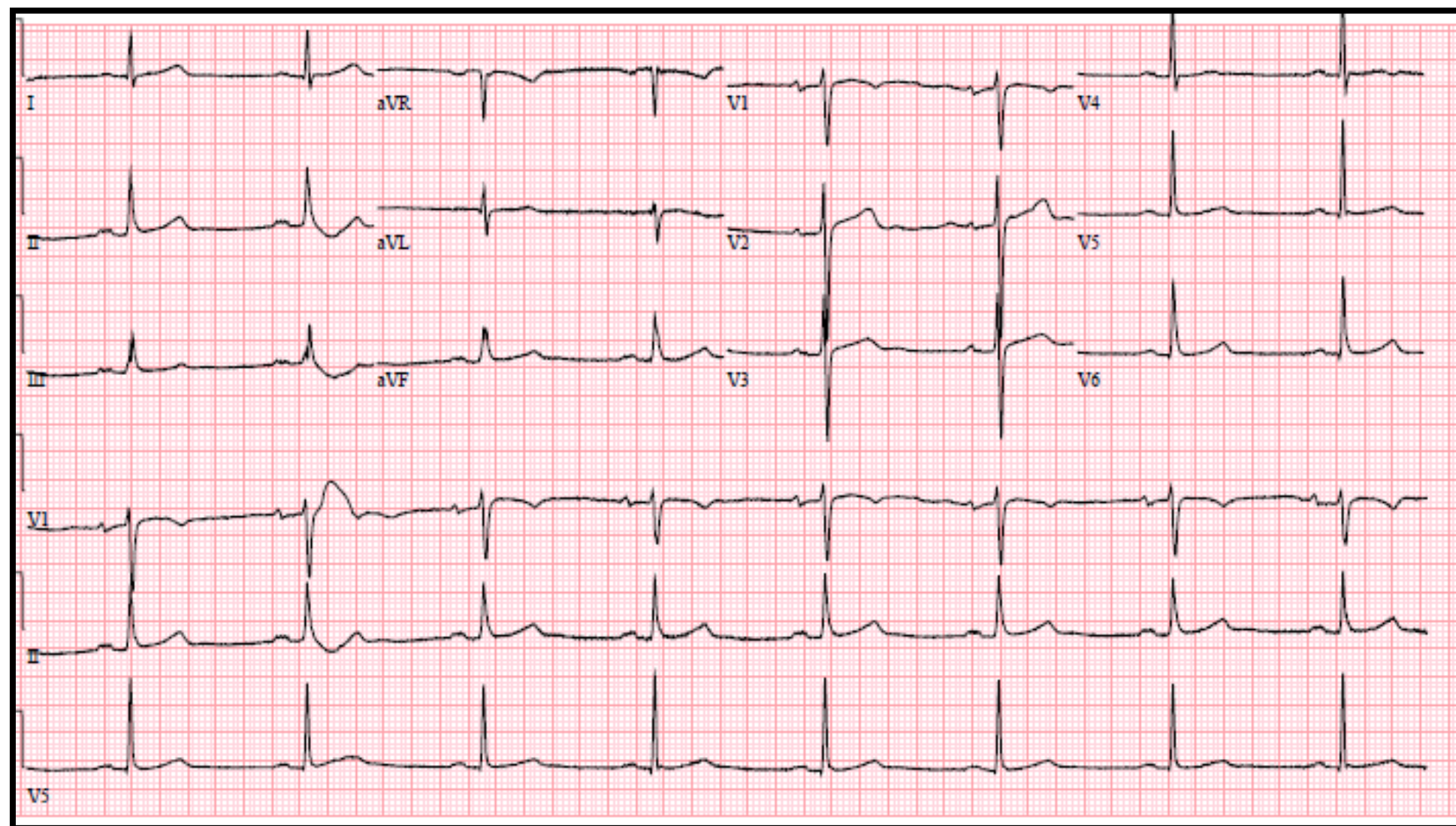


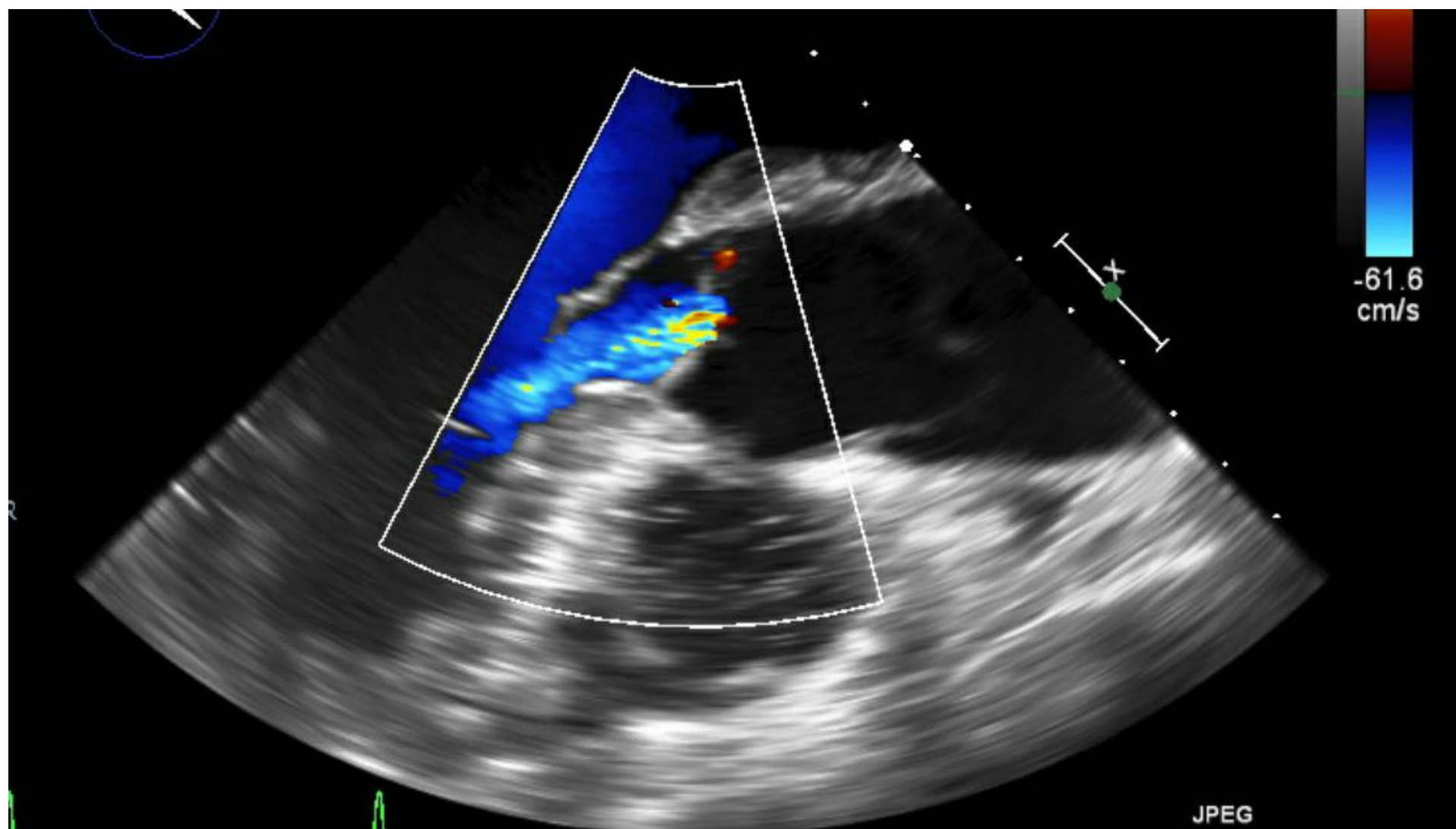
- Normal carotid upstrokes and volumes
- Flat neck veins
- Clear lungs

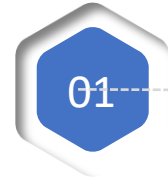


- Normal S1, S2 physiologically split, not muffled
- No aortic insufficiency
- Abdomen is soft
- Normal upper extremities and distal LE pulses

# EKG







Posterior leads, given back pain.  
This could be a Lcx event.



Right sided leads, given flat  
neck veins that could be sign  
of RV MI

### Next Steps

Include, in addition to IV  
TNG to lower BP



CT Angio, with concern for  
aortic dissection



Give antacids, for GERD



Study  
Study T  
SCAN  
AcqT



0.5 Soft

# Clinical Presentations and Signs of Type A Dissection

	<u>No previous cardiac surgery</u>	<u>Previous cardiac surgery</u>	
Chest pain or back pain	88%	68%	p <0.05
Abrupt onset pain	92%	84%	p <0.05
Any pulse deficit	28%	30%	NS
Pericardial effusion	48%	26%	p <0.05
Tamponade	16%	3%	NS
*Widest diameter of ascending Ao	5.28 (1.34)	5.40 (1.57)	NS

# Heritable Presentation of Thoracic Aortic Aneurysm and Dissection (HTAAD)

10-23% at most of Type A dissection have a genetic basis that we understand  
53 Gene Panel for aortic dissection

Many are variants of unknown significance and some have little evidence, thus only create concern



- 37 autosomal dominant
- 11 autosomal recessive
- 4 x-linked recessive
- 1 x-linked dominant



2 genes COL3A1 and MYLK are associated with dissection but not with aneurysmal dilation



# Genes in the Definitive and Strong Categories:



Genes in moderate?  
Newer genes and category D?

## CENTRAL ILLUSTRATION Evaluation of the Clinical Validity of Genes for Heritable Thoracic Aortic Aneurysms and Dissections (HTAAD)

Category A	Category B	Category D
<u>DEFINITIVE</u> COL3A1 FBN1 SMAD3 TGFB2 TGFB1 TGFB2 } A1  ACTA2 MYH11 MYLK } A2  <u>STRONG</u> LOX PRKG1 } A2	<u>MODERATE</u> EFEMP2  <u>LIMITED</u> ELN FBN2 FLNA NOTCH1 SLC2A10 SMAD4 SKI	<u>NO EVIDENCE</u> ACVRL1 ADAMTS10 B3GAT3 COL1A1 COL1A2 COL4A1 COL5A1 COL5A2 COL9A1 COL9A2 COL11A1 COL18A1 EMILIN1 ENG GATA5 GJA1 JAG1 MED12 PLOD1 PLOD3 SMAD6 UPF3B VCAN
Recent genes	Category C	
<u>UNCERTAIN</u> BGN FOXE3 HCN4 MAT2A MFAP5 SMAD2 TGFB3	<u>LIMITED</u> CBS COL4A5 PKD1 PKD2	

# Incidence and Outcomes of Acute Aortic Dissection: BP Matters!

2002-2015

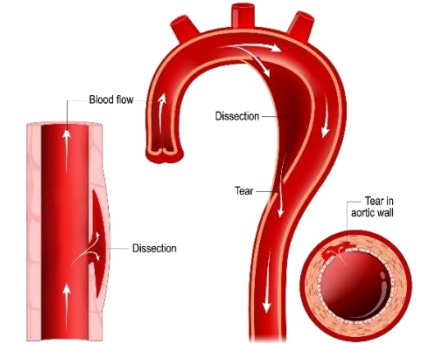
**92,728 people** in Oxfordshire UK

**155 patients** with 174 acute aortic events

**54 patients** had 59 thoracoabdominal aortic dissections

52 incident events – 6/100,000

Aortic dissection



## 31 Type A

46.6% died before hospitalization  
of those who arrived at hospital 47%  
died by 30 days

## 15 Type B

13% died by 30 days

Correlate with the immediately fatal events:

Pre-morbid BP  $151.2 \pm 19.3$  vs  $137.9 \pm$  p<0.001



# Case Study: 62-Year-Old Woman

Comes to ED  
for worsening of what was considered chronic chest  
pain,  
a few hours after an outpatient visit for venography pre  
AV fistula (Cr 2.1-2.4) Tr-hs 72

Lives alone, former postal worker, on  
disability related to depression and PTSD  
after witnessing a horrific 2009 MVA

## Childhood

Thymectomy via left  
chest

## 10 yrs PTA

Right renal cell  
carcinoma  
Partial nephrectomy

## 3 years PTA

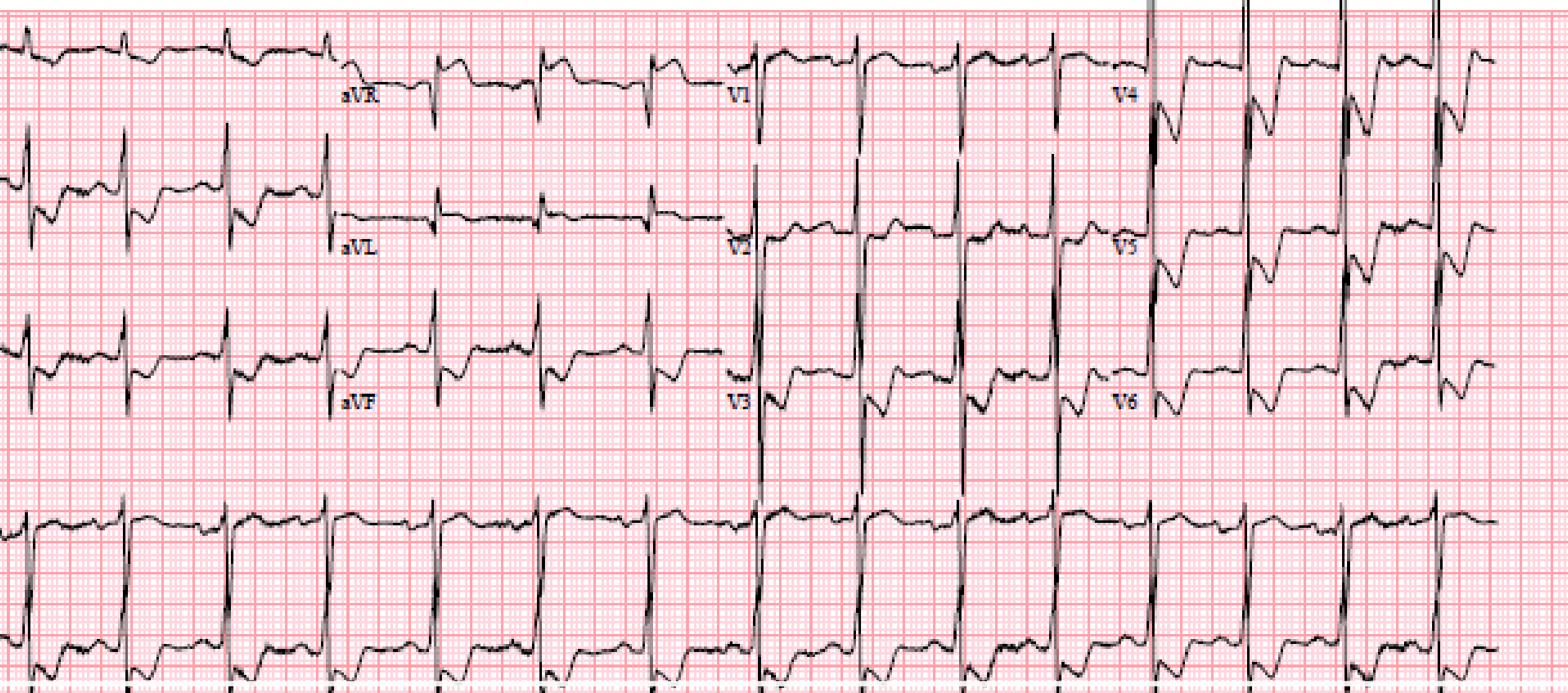
Right renal cell  
carcinoma  
Robotic excision

## 1 year PTA

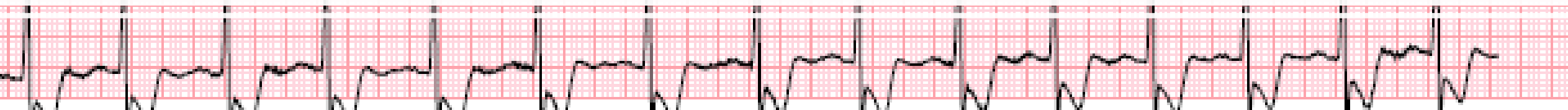
EGD with  
esophagitis

## Stature, Risks

66" tall  
177 pounds  
DM A1c 7.6%  
HTN  
Former smoker  
LAD stent 7 months prior



**Case Study: 62 yo Woman - 11/2020 admission**



Study Date:  
IM Ti  
Zoom



Case Study:

62 yo Woman with chest pain, global ST segment changes, 7 months after attention to LAD, conservatively managed, 50% left main

# Case Study: 62-Year-Old Woman - Cath

80% LM



Patent LAD stent



Occluded RCA



70% MI



6.0 mm left main stent, excellent result, IVUS  
area 23 mm



Stable hemodynamics. Though prepped for impella, with pigtail  
placed in LV, did not need circulatory support.



Recovers quickly

# Case Study: 55-year-old Man

## BACKGROUND

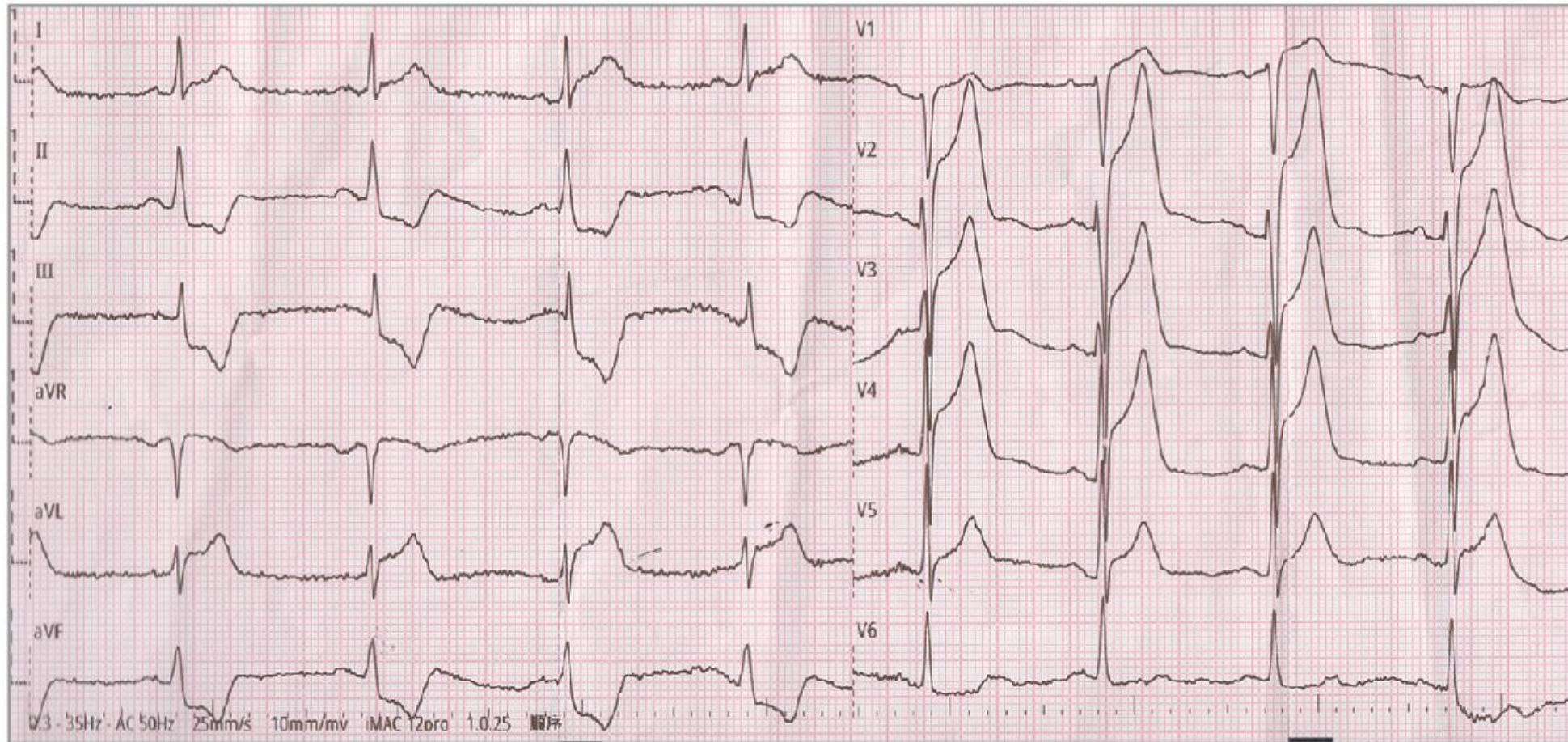
- 20 minutes of sustained chest pain, radiating to shoulders and back, associated with shortness of breath and diaphoresis
- Hypertensive
- Smoker

- Unable to stay still
- BP right arm 185/104; left arm 113/98
- HR 58, rr 24, O2 sat 98% on room air
- Neck veins 8 cm H2O
- Carotids, brisk on right
- Clear lungs, nondisplaced PMI, barely palpable, soft S1, physiologically split S2, faint diastolic murmur along left sternal border

## EXAM



# Case Study: 55-year-old Man



- Anterior ST  $\uparrow$ , I, L ST  $\uparrow$
- “Reciprocal” inferior ST depression
- Subtle ST  $\uparrow$ , aVR, thus not summing lateral leads.

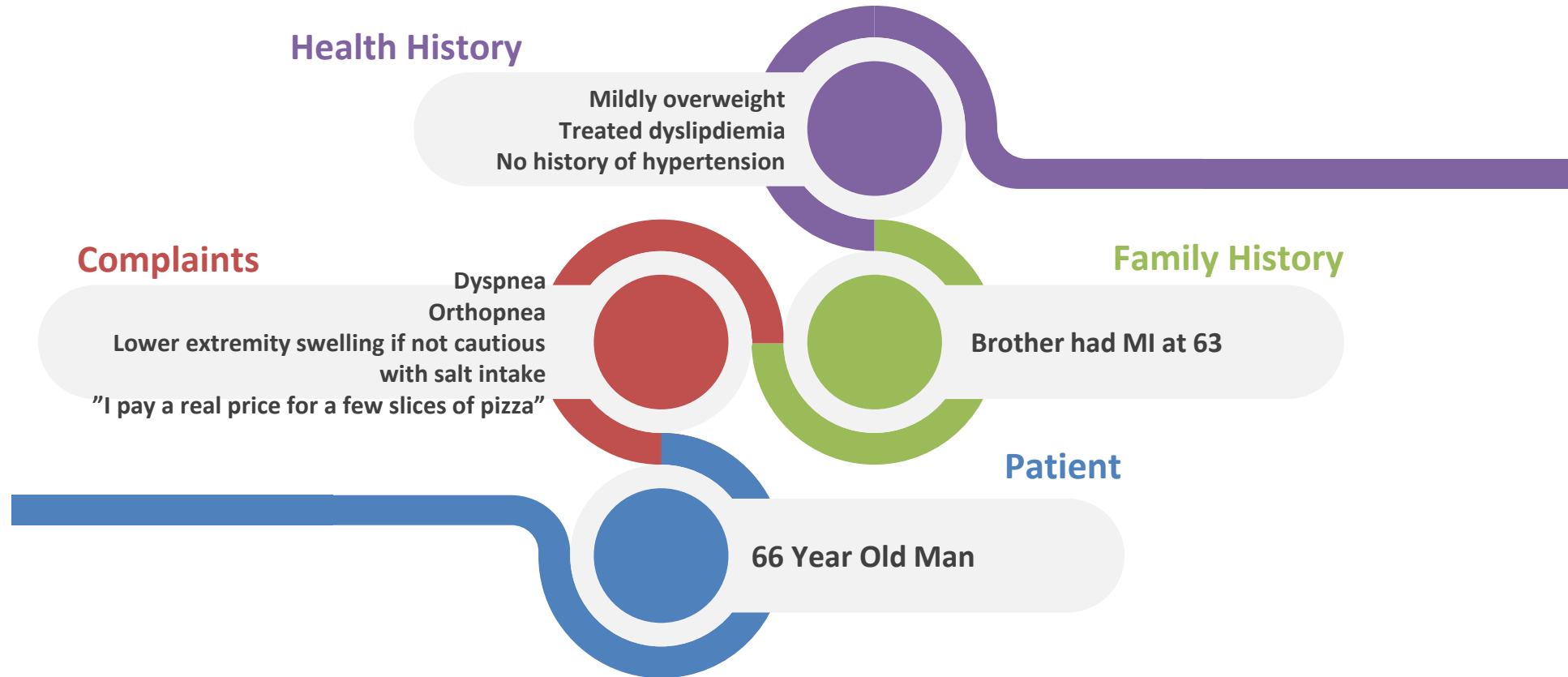
# Case Study: 55-year-old Man

- More than 8 leads, left main should be considered
- Lead AVR should sum V4, V5, V6 and be reciprocal (↓). When it is not, think ostial LAD, pre septal perforator, or left main
- V1 elevation, possible posterior ST depression, possible RV involvement, ostial LAD or left main or ...

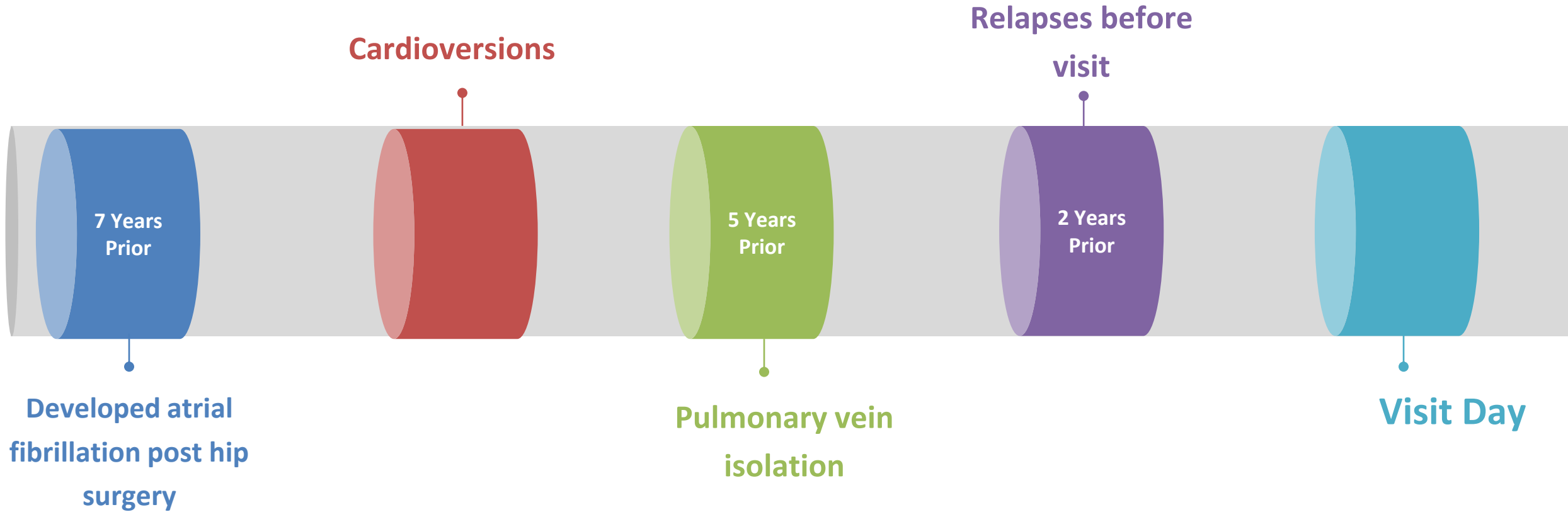
# Case Study: 55-year-old Man

- Unable to sit still-more often aortic dissection, than coronary issue
- BP differential between left and right
- Diastole murmur
- EKG
- Aortic dissection, involving ostium of left main, and ischemia so severe, even proximal supplied area, upper septum, reflected in avR, is ischemic. Different than prior case, where proximal area still has some blood flow, and more distal regions suffer more.

# Case Study: D365a-a



# Case Study: D365a-a





# Case Study: D365a-a

## Meds

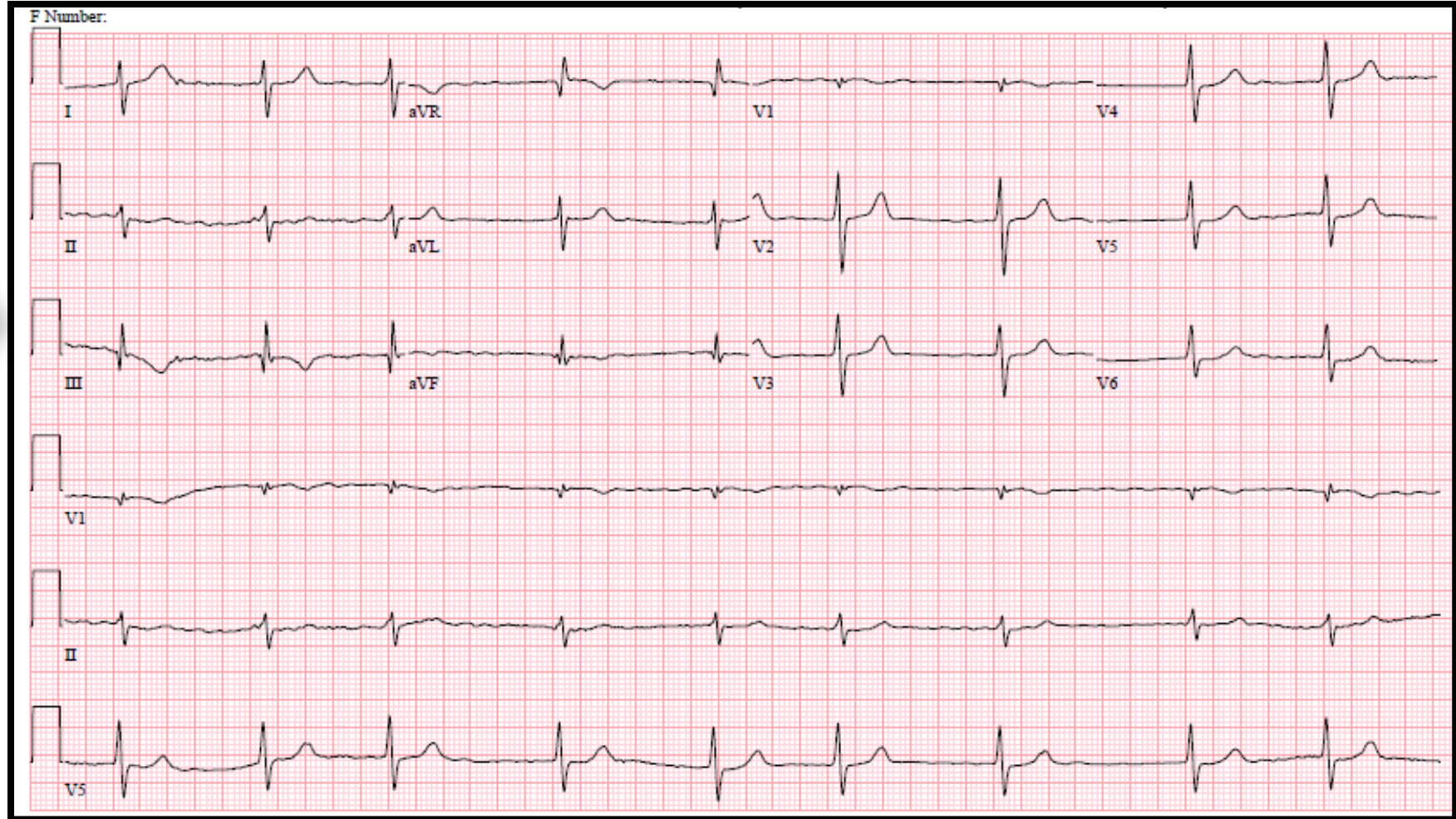
Apixaban  
Carvedilol  
Lisinopril  
Diltiazem  
Furosemide  
Pravastatin

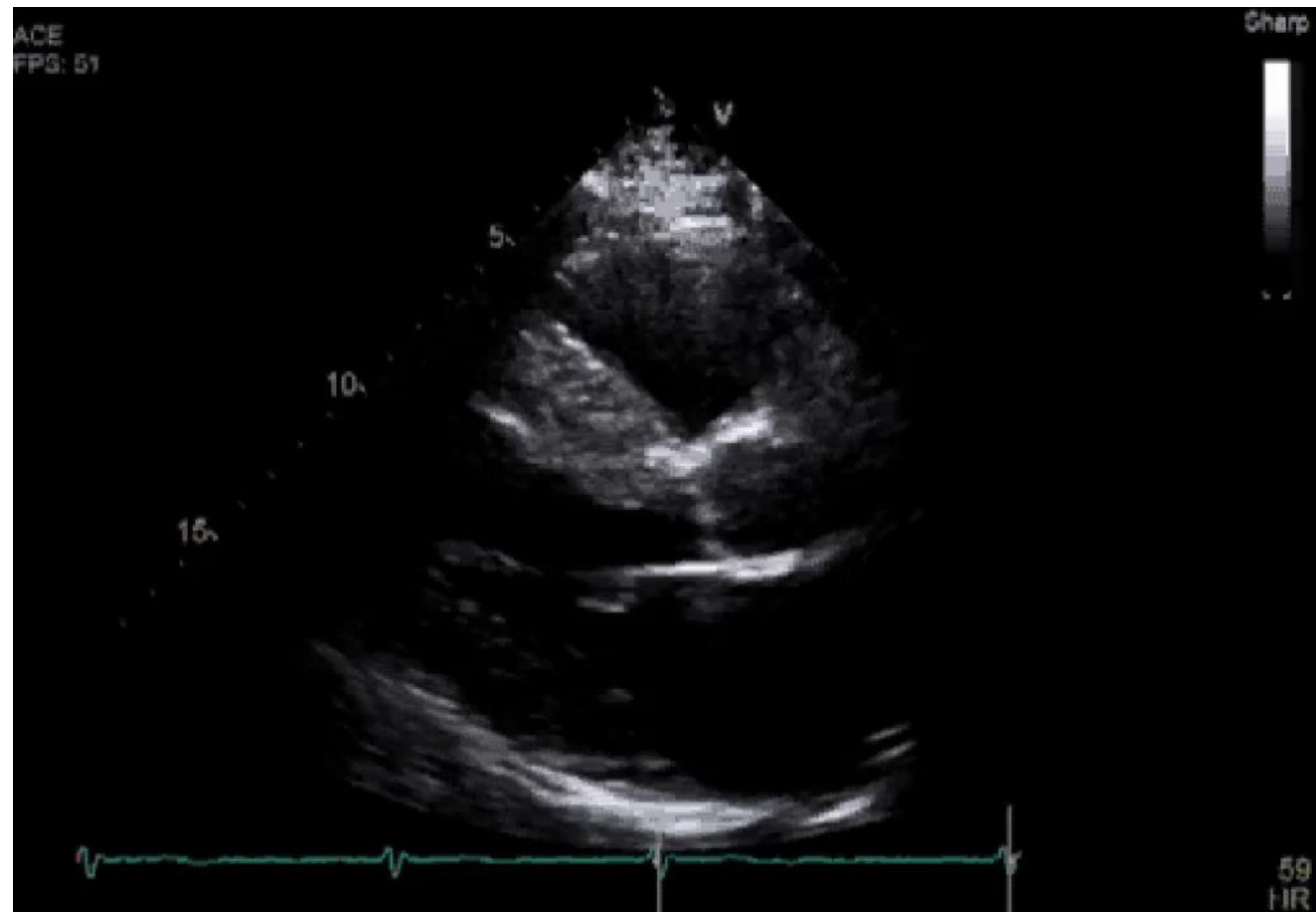
## Exam

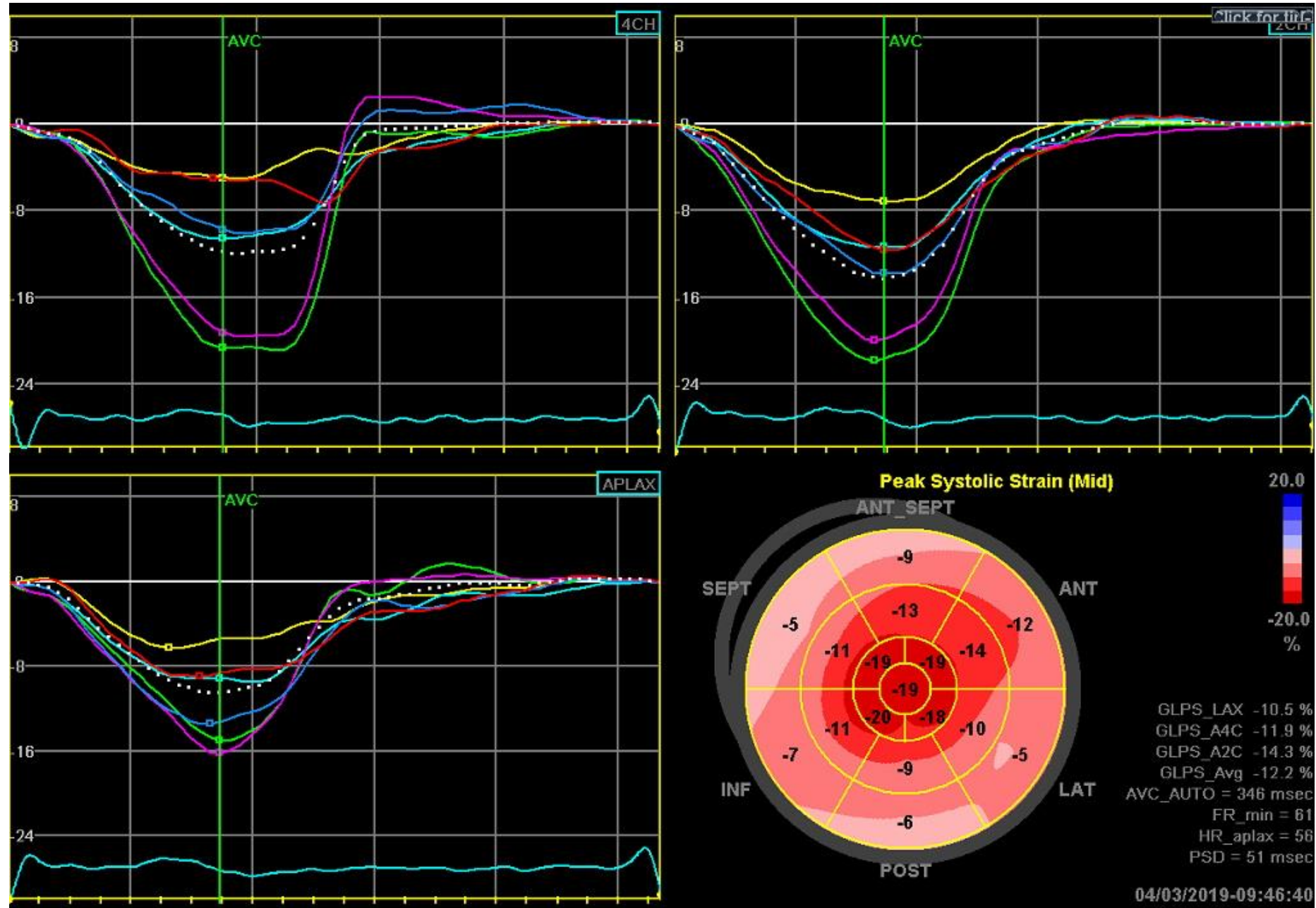
120/70 mmHg seated and standing  
HR ~ 50-60 bpm  
Neck veins < 7cm H<sub>2</sub>O  
Normal carotid upstrokes and volumes  
Variable S<sub>1</sub>,  
Physiologically split S<sub>2</sub>  
P<sub>2</sub> not increased

# Case Study: D365a-a

**ECG**







GLS abnormal -14, and base (worse) to apex gradient

# Next Steps

**Wonder about ischemia that was missed?**

- Suggest exercise echo

**Suspect infiltrative disease, based on recurrent atrial fibrillation issues, poor tolerance of atrial fibrillation, and LVH.**

- Order SPEP, UPEP, Serum free light chains looking for amyloid; if all negative, technetium pyrophosphate scan looking for TTR amyloid

**Voltage is not low, nuclear perfusion study showed only a fixed defect and no ischemia.**

- Just manage the patient's volume

**Thinking about infiltrative disease does not exclude CAD.**


- Look for both



# Course-D365



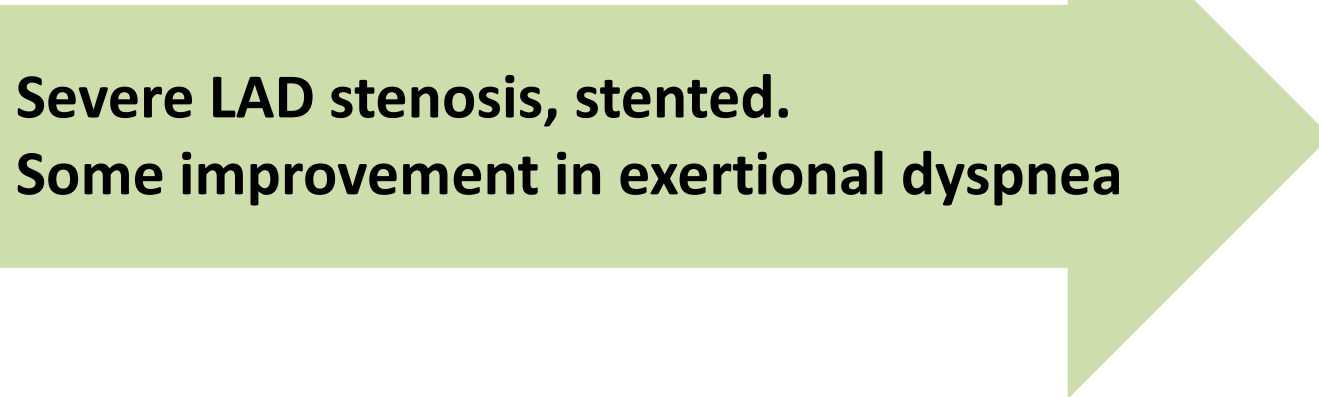
SPEP, UPEP and SFLC unremarkable.  
99.2% of time at least one of these will  
be abnormal with AL



Technetium pyrophosphate with  
marked uptake, highly suggestive of TTR  
amyloid. Low voltage seen less than half  
the time. TTR gene unremarkable, thus  
wild type.



Exercise echo positive anteriorly



Severe LAD stenosis, stented.  
Some improvement in exertional dyspnea



Embarking on course of tafamidis

# Suspect AL Amyloid Amongst HF Population

## Serum free light chain abnormalities

Normal K:  $\lambda$  0.26-1.74

Ratio markedly elevated or depressed especially when  $K > 140$ ;  $\lambda > 60$

May precede clinical diagnosis by many years

↑ NTProBNP – especially when really ↑

Posterior wall thickness > 1.3 cm

## Other helpful ECG findings:

Any infarct ~ 50%

Low voltage ~17%

RVSP > 50

Global longitudinal strain -14 ↑

Base worse

Then apex

# Case Study-67 yo woman with chest pain



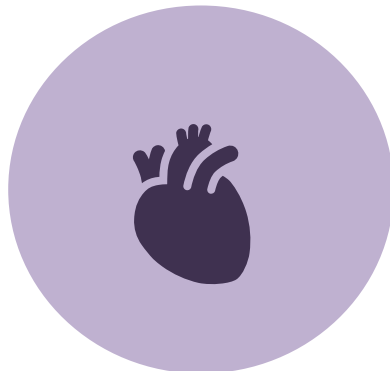
Family History

Premature  
Vascular  
Disease



Age 47

Right  
Cerebral  
Event – soon  
after initiating  
estrogen



Age 55

Told of  
LBBB



Age 67

Treated  
dyslipidemia  
hypertension

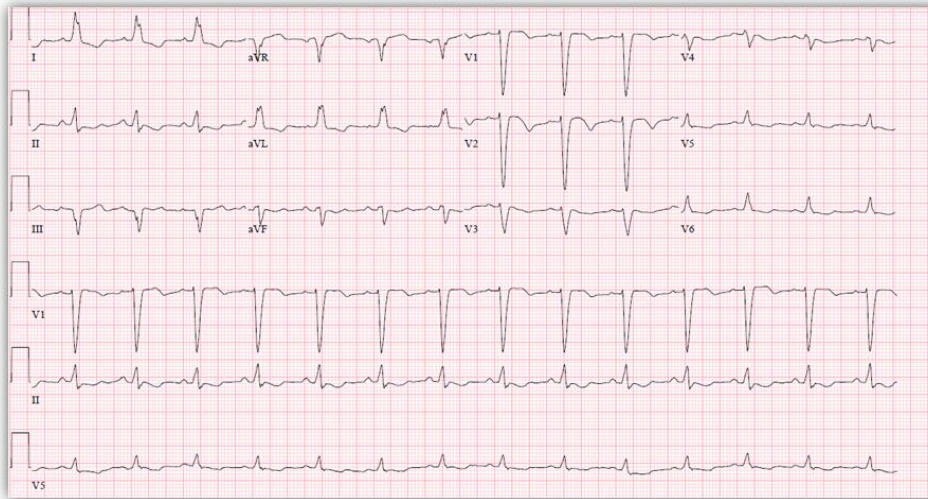


Present Cardiac  
History

Chest Pain –  
following  
mechanical fall  
related left-hip  
fracture and open  
reduction and  
internal fixation

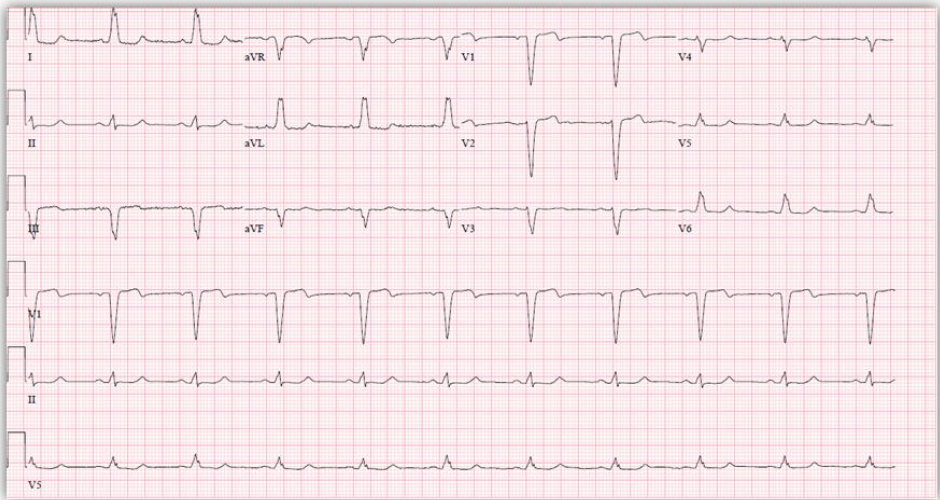
# EKG Comparison

Acute



Concordant T waves V2,V3

old

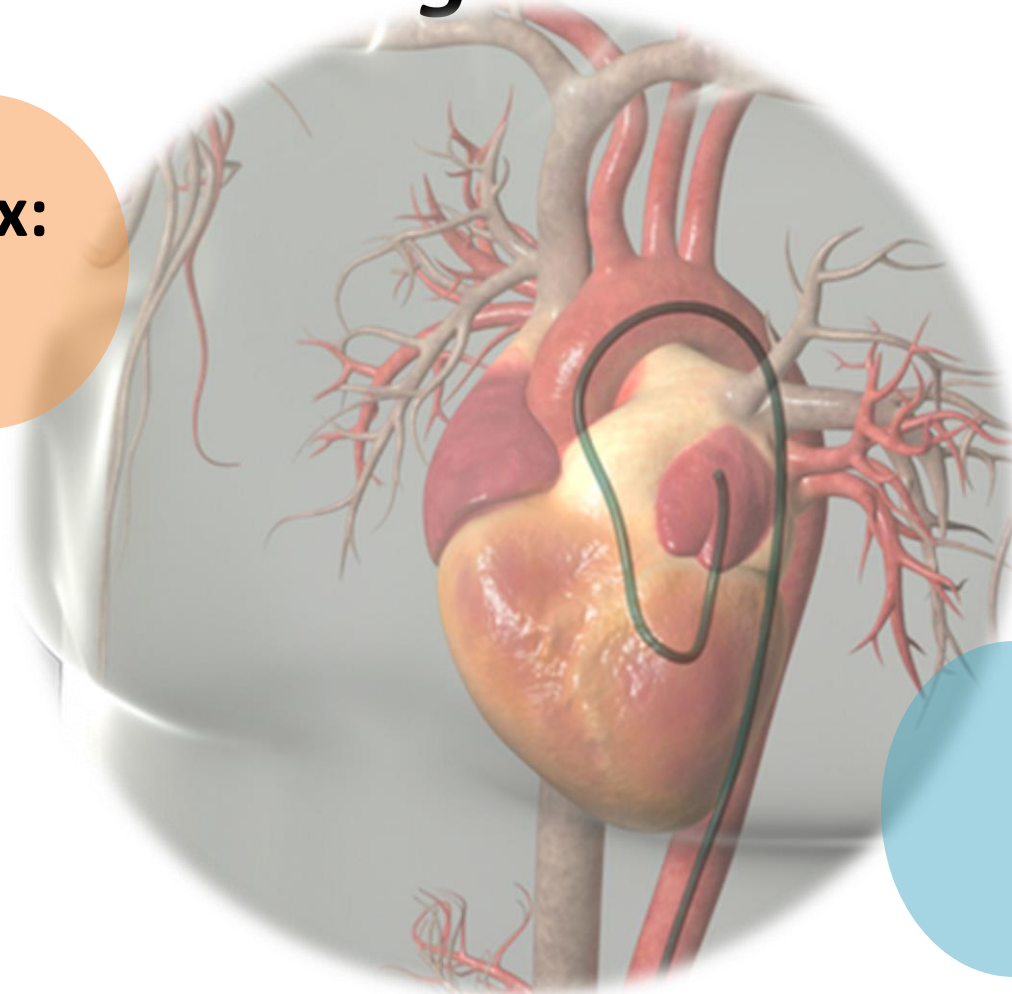


Concordant T waves laterally

# Outcome

*Underwent Emergent Catheterization*

Occluded left circumflex:  
stented



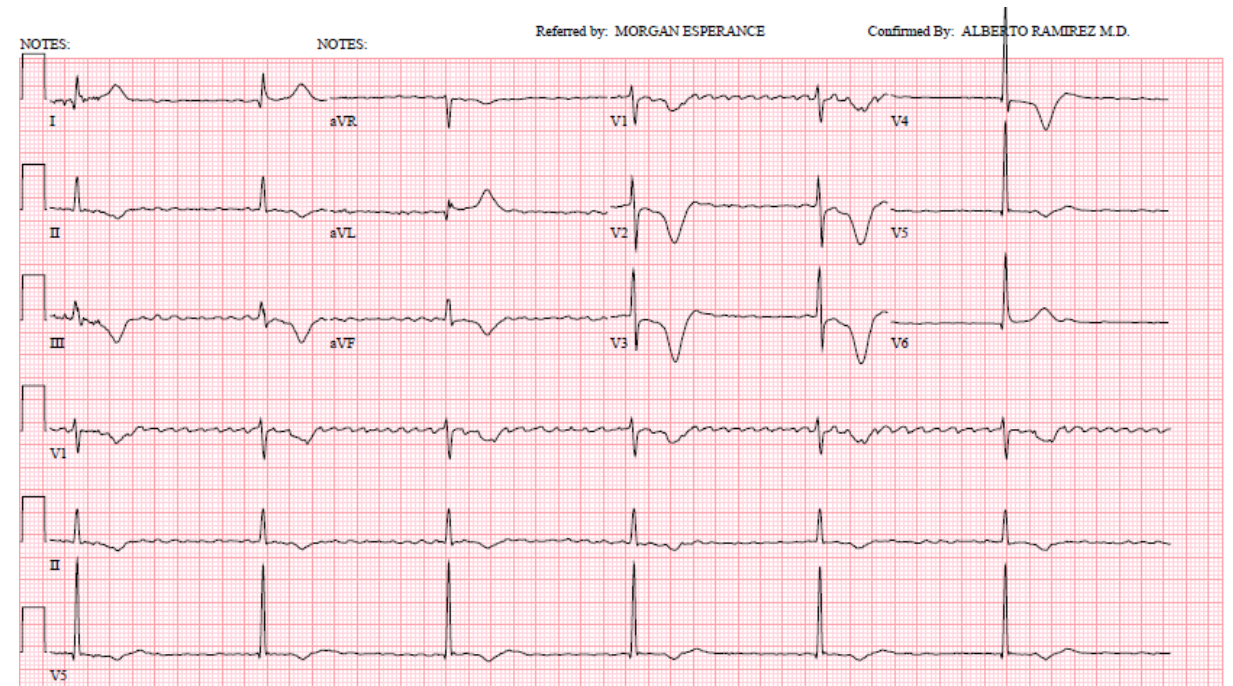
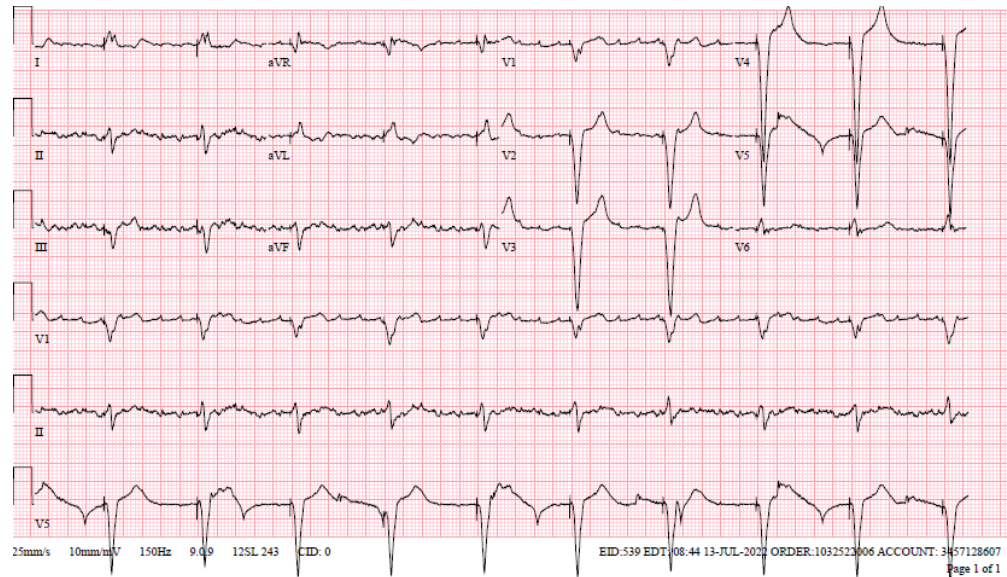
Occluded LAD:  
collateralized



# Case Study-Last One!!

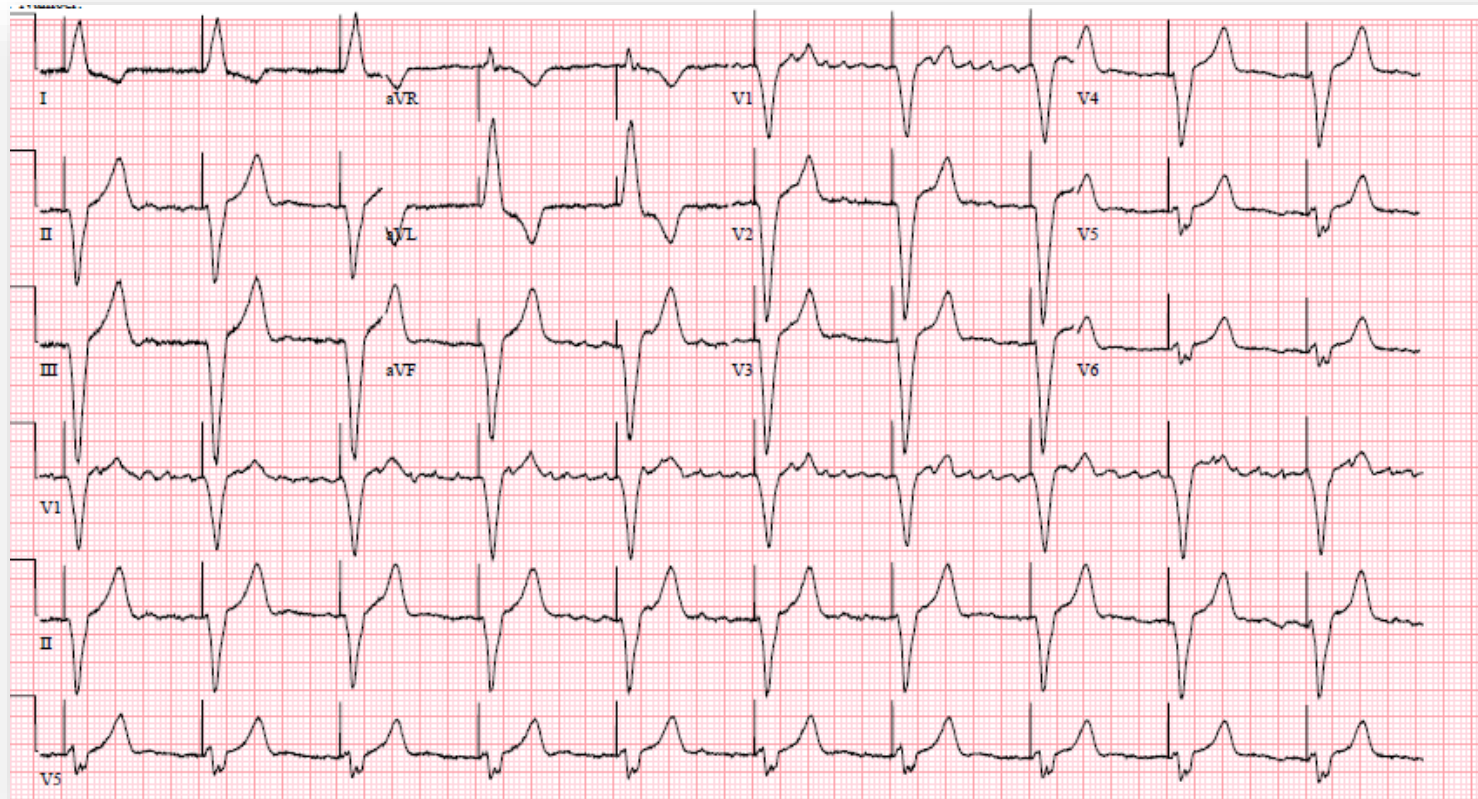
- 63 yo man with intermittent total body weakness episode
- C5-6 quadriplegia related to 1992 MVA
- Permanent atrial fibrillation
- 2018 received Micra Pacemaker for periods of bradycardia.

# Initially Fine, Then Profoundly Weak



# Which of the Following is True?

1. He is having pacer failure with bradycardia
2. He is having anterolateral ischemic issues causing weakness
3. He is having pacer failure with post pacing repolarization abnormality



**Thank You For Your Attention**

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