2019
ACOI-Internal Medicine Board Review
Valvular and Congenital Heart Disease

Robert Bender, DO, FACOI, FACC

Endocarditis Prophylaxis

AHA (2007) = antibiotic prophylaxis recommended only for patients with the highest risk:

* prosthetic valve
* previous endocarditis
* Congenital dz = repaired with residua, unrepaired/palliative repair, complete repair including catheter intervention (1^{st} – 6 mos)
* cardiac transplant pts with valve disease

Endocarditis Prophylaxis

- Routine antibiotic prophylaxis for patients with native valve disease and no prior history of endocarditis =

NOT RECOMMENDED !!

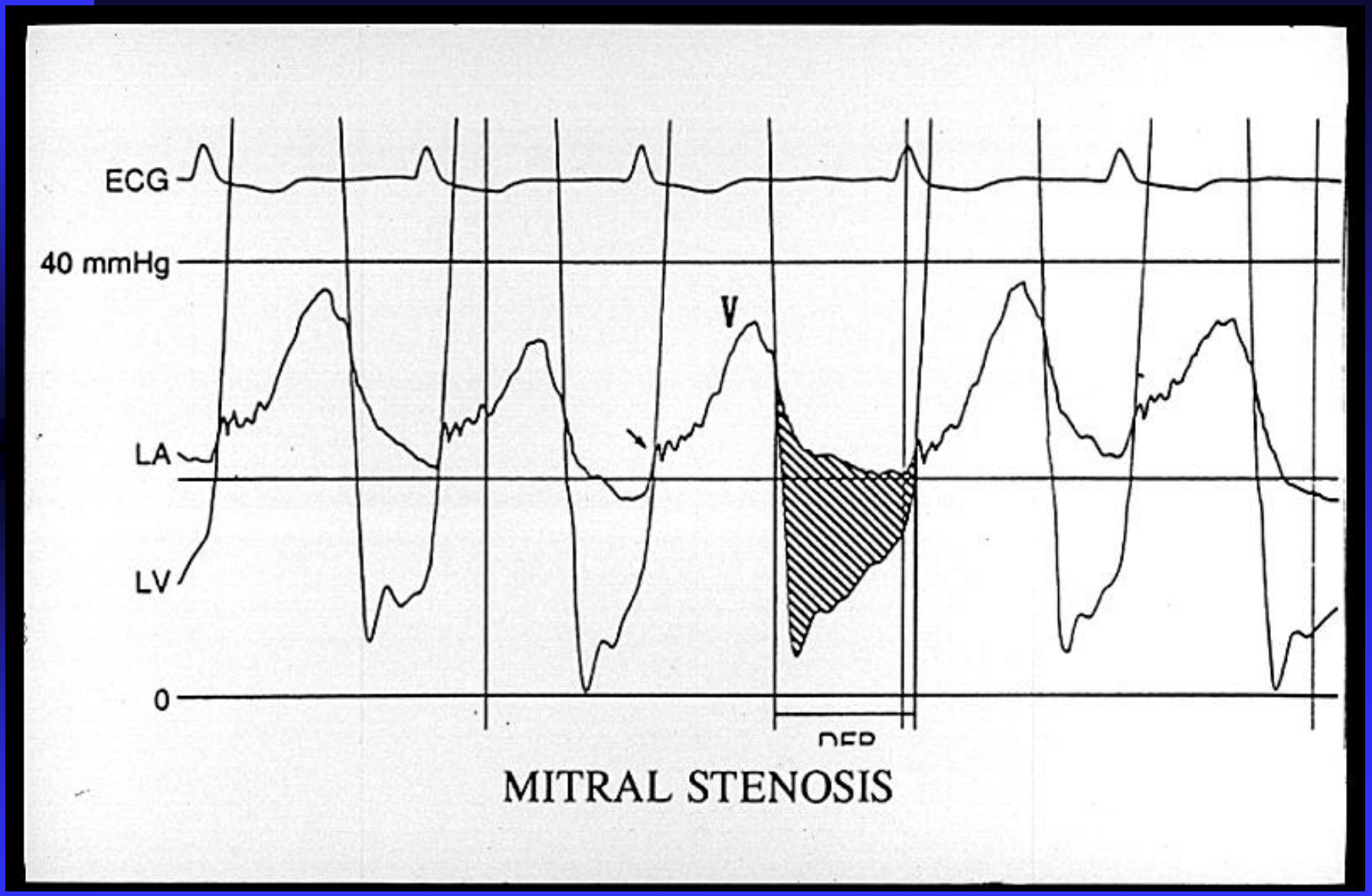
Mitral Stenosis (MS):

Etiology: Rheumatic Fever (20-40 yr. latency)

* Congenital
* Mimics MS = LA tumor, thrombus, cor triatriatum
* Lutembachers Syndrome = ASD and MS
* Severe MAC = elderly (difficult management)

MS - Hemodynamics:

- Mitral Gradient = flow dependent
- Mitral Valve Area =
  - Normal - 4 - 6 cm^2
  - Severe - </= 1.5 cm^2 (gradient > 10 mmHg)
  - Very severe - </= 1 cm^2



MITRAL STENOSIS

MS - Clinical:

- Sx = SOB/Heart Failure, Hemoptysis, CP
- Ortners Synd. = hoarseness d/t compression of left recurrent laryngeal nerve
- Pulses = small (d/t ↓ CO)
- Neck Veins = increased if right heart failure

MS - Clinical:

- Auscultation
  - Opening snap (OS) = early diastole, apex, high frequency
    - OS occurs earlier as MS worsens
    - OS absent = heavy Ca^{tt}

MS - Clinical:

- Auscultation
  - * Classic murmur = low pitch diastolic rumble at apex
  - * As MS worsens = murmur lengthens
  - * Pre-systolic accentuation = implies NSR
  - *↑Intensity = squatting, amyl nitrite, exercise
  - * ↓ Intensity = Valsalva

MS - Complications:

- Death = CHF, systemic embolism, PE
- Systemic Embolism = CVA, etc.
  - 80% AFib
  - < severe MS
  - Tx = anticoagulate (warfarin, not DOAC's)
  - ? indication for surgery.

MS - Non Invasive Testing:

- EKG = AFib (coarse), LA enlarge, RVH
- CXR =
  - LA enlargement = correlates poorly with severity
  - PA, RV, RA enlargement = severe MS
  - MAC, hemosiderosis, ossification

MS - Non Invasive Testing:

- Echo =
  - Thick, restricted leaflets
  - ↓ EF slope
  - Leaflet “doming” (diastole)
- Doppler =
  - Gradient
  - Valve area
  - Pulmonary artery pressure


A
P
LA

MS - Treatment:

- Medical = Anticoagulation, HR control, diuretic
- Surgical (balloon, commissurotomy, MVR)
  - Symptomatic: and MV area ≤ 1.5 cm2 (class I)
  - Asymptomatic:
    - Very severe MS (area </= 1.0 cm2) & valve favorable for balloon (IIa)
    - Severe MS (area </= 1.5 cm2) with new Afib & valve favorable for balloon (IIb)

Chronic Mitral Regurgitation (MR):

- Etiology = primary (degenerative) vs secondary (functional)
  - Mitral apparatus abnormalities:
    * leaflets, annulus, chordae, papillary muscle = eg: MVP, SBE, LV dil., MI
  - MVP = most common cause of isolated MR requiring MVR

MR - Pathophysiology:

- Volume Overload = Eccentric hypertrophy
  - LV mass/volume ratio = normal
- LV Ejection Fraction = increased
  - d/t ↓ afterload

MR - Clinical:

- SX = heart failure, may appear "late"
- Pulses = brisk (sharp upstroke, normal volume)
- Auscultation:
  - S1 = ↓, S2 = splitting
  - P_2↑ = (pulm. HTN)
  - S_3 = not necessarily LV failure

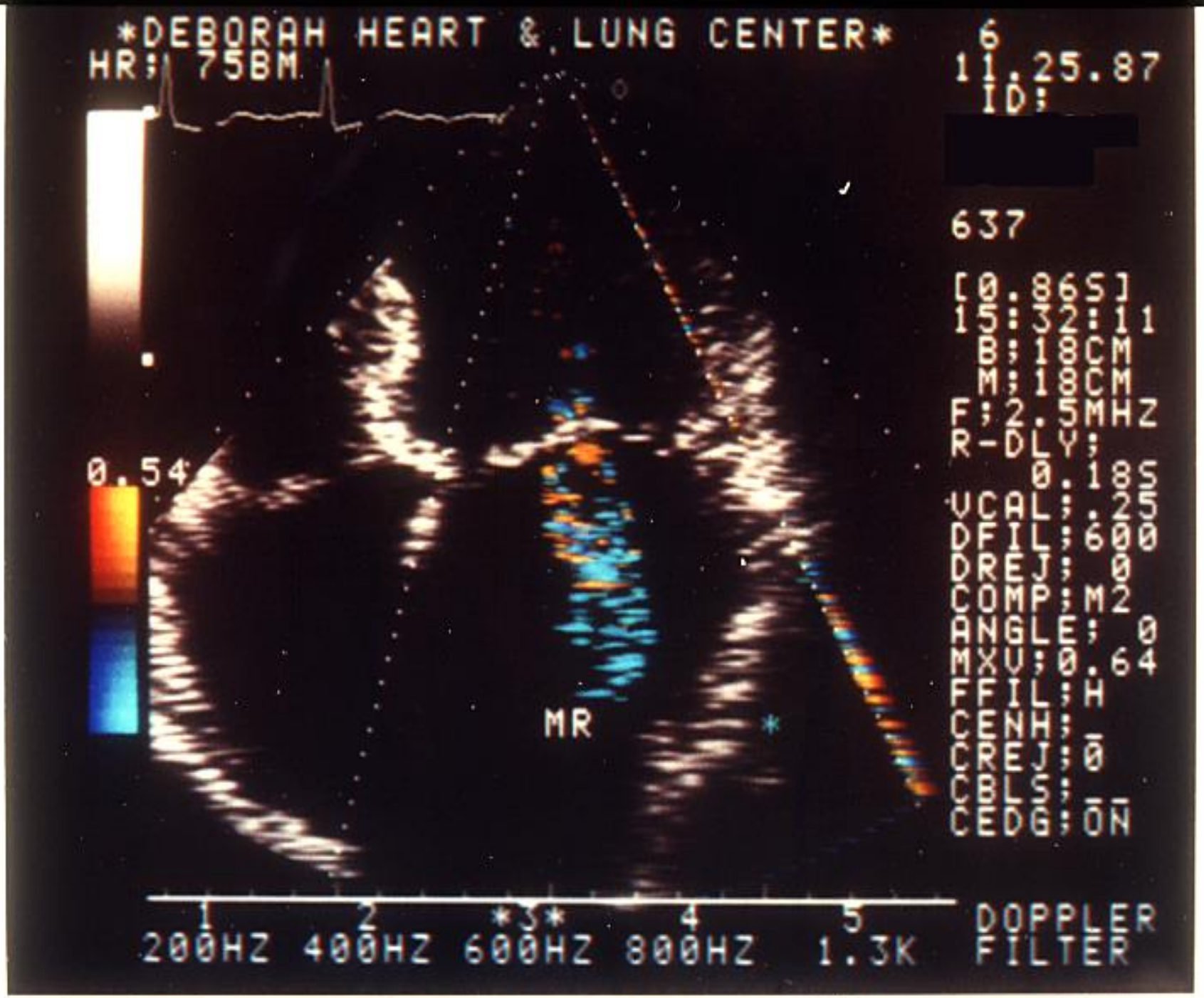
MR - Clinical:

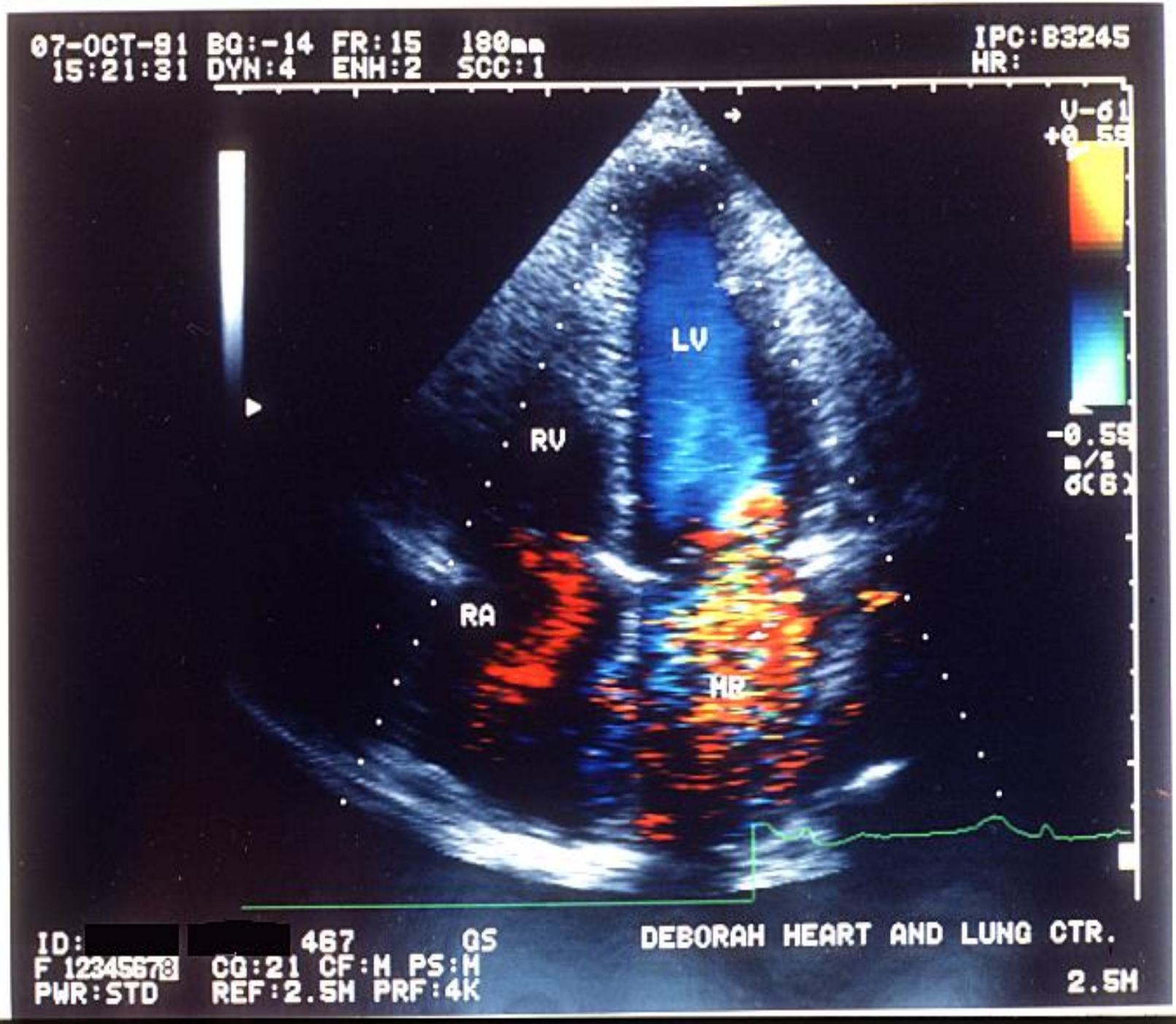
- Auscultation:
  - Murmur = holosystolic
    - apex to axilla (but not always)
    - intensity may not reflect severity
    - ↑ intensity = squatting, isometrics
    - ↓ intensity = Valsalva, amyl nitrite
    - Acute MR = atypical

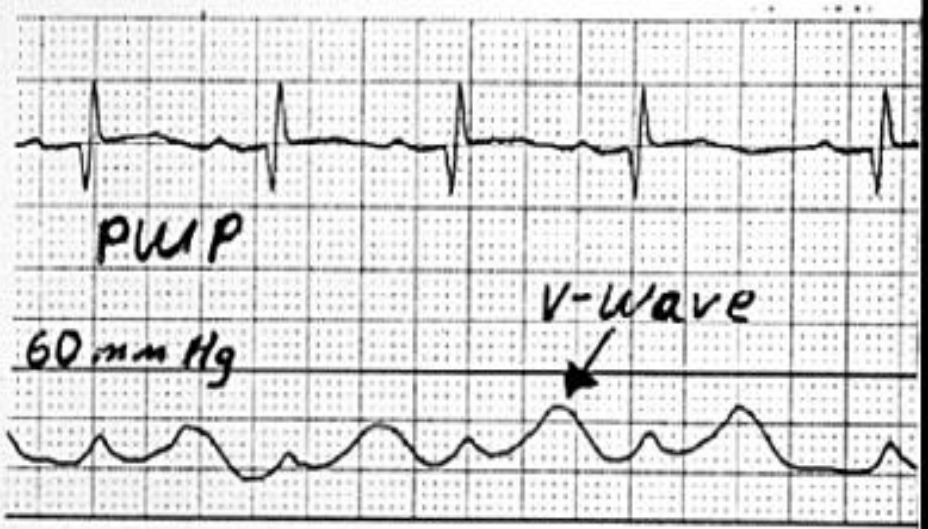
MR - Non Invasive Testing:

- EKG = LA enlarge., LVH
- CXR = LA, LV enlarge.
- Echo = chamber sizes, LV fxn., etiology
- Doppler = quantitate severity
  * TEE > TTE

- Cardiac MRI = discordant clinical vs echo


DEBORAH HEART & LUNG CENTER
HR; 75BM
6
11.25.87
ID;
637
[0.86S]
15:32:11
8;18CM
M;18CM
F;2.5MHZ
R-DLY;
0.185
VCAL;;250
VDFIL;;600
VDREJ;;0
VCOMP;;M2
VANGLE;;0
VMXVL;0.64
VFFIL;;H
VCENH;;;
VCREJ;;;0
VCBLS;;;
VCEDG;;ON
0.54
MR
1
2
3
4
5
200HZ 400HZ 600HZ 800HZ 1.3K
DOPPLER FILTER


07-OCT-91 BQ:-14 FR:15 180mm
15:21:31 DYN:4 ENH:2 SCC:1
IPC:B3245
HR:
V-61
+0.55
LV
RV
RA
MR
-0.55
m/s
ID:
467
QS
DEBORAH HEART AND LUNG CTR.
F 12345678
CQ:21 CF:M PS:M
PWR:STD
REF:2.5M PRF:4K
2.5M



MITRAL REGURGITATION
'V'- WAVES

MR - Treatment:

- Medical (acute) = afterload reduction, diuretics
- Surgical = mitral repair or replacement
- Primary MR:
  - Severe MR with sx (and LVEF > 30%) – class I
  - Severe MR without sx. but…
    - LV dysfunction (EF ≤ 60%, but > 30%) – class I

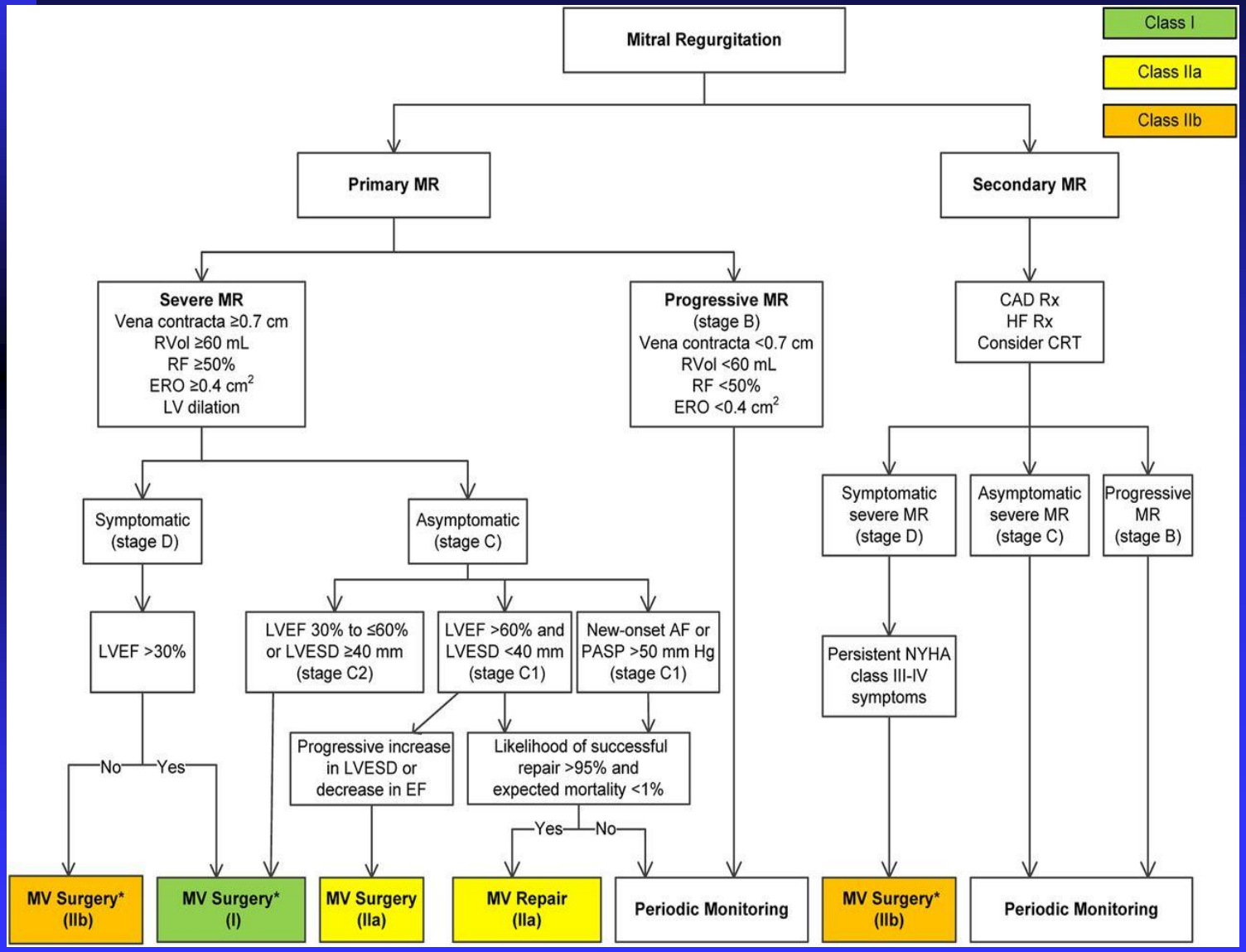
    or
    - End - Systolic dimension ≥ 40 mm – class I

    or
    - Pulm. HTN (systolic > 50 mmHg rest) – class IIa (repair)

    or
    - New onset AFib – class IIa (repair)

    or
    - High likelihood of repair & low surg risk – class IIa
  - EF< 30% = ? candidate for surgery (class IIb)
  - Transcatheter repair or replacement = ongoing investigation

ACC Guidelines (July 2017;Nishimura, et al)



Mitral Valve Prolapse (MVP)

- Prevalence = 5-10% of population
- Symptoms = asymptomatic, palps, CP
- Auscultation
  - mid-syst. click / late syst. murmur
  - earlier click/murmur = ↓ LV volume
    - Valsalva, standing
  - later click/murmur = ↑ LV volume
    - squatting, isometrics

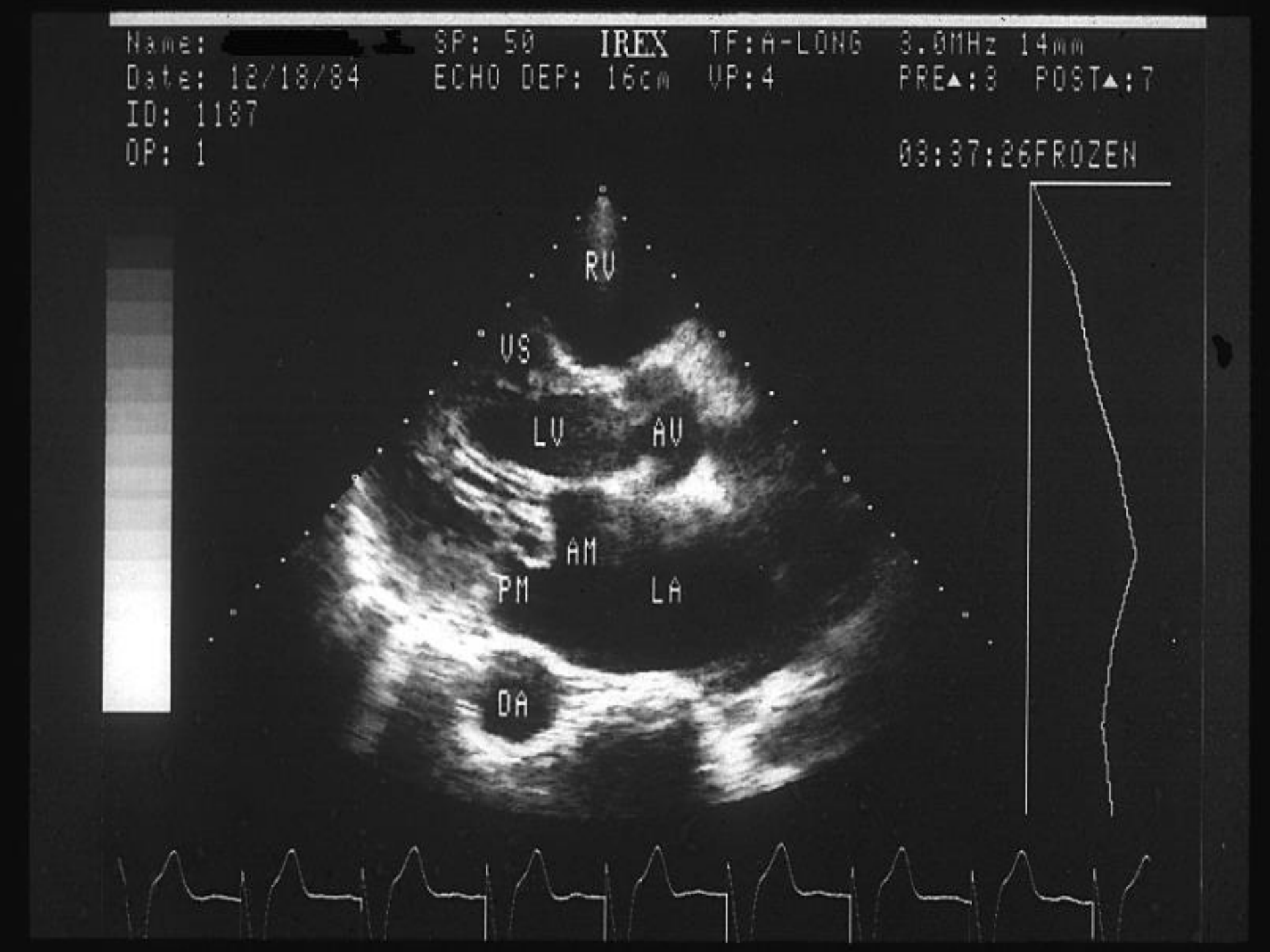
MVP - Non Invasive Testing:

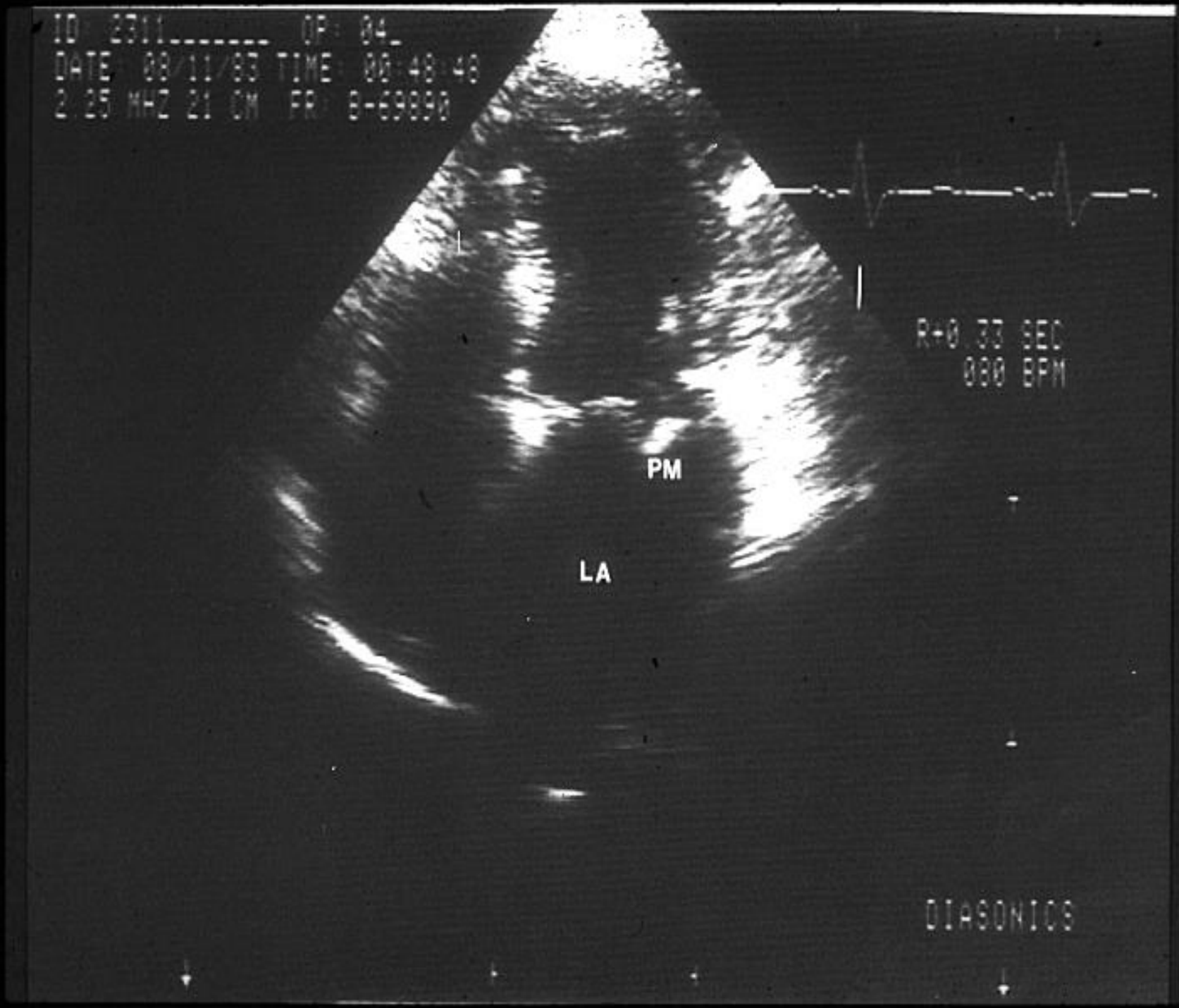
- EKG = usually normal
  - PSVT
  - ↑ incidence WPW
- CXR = unhelpful

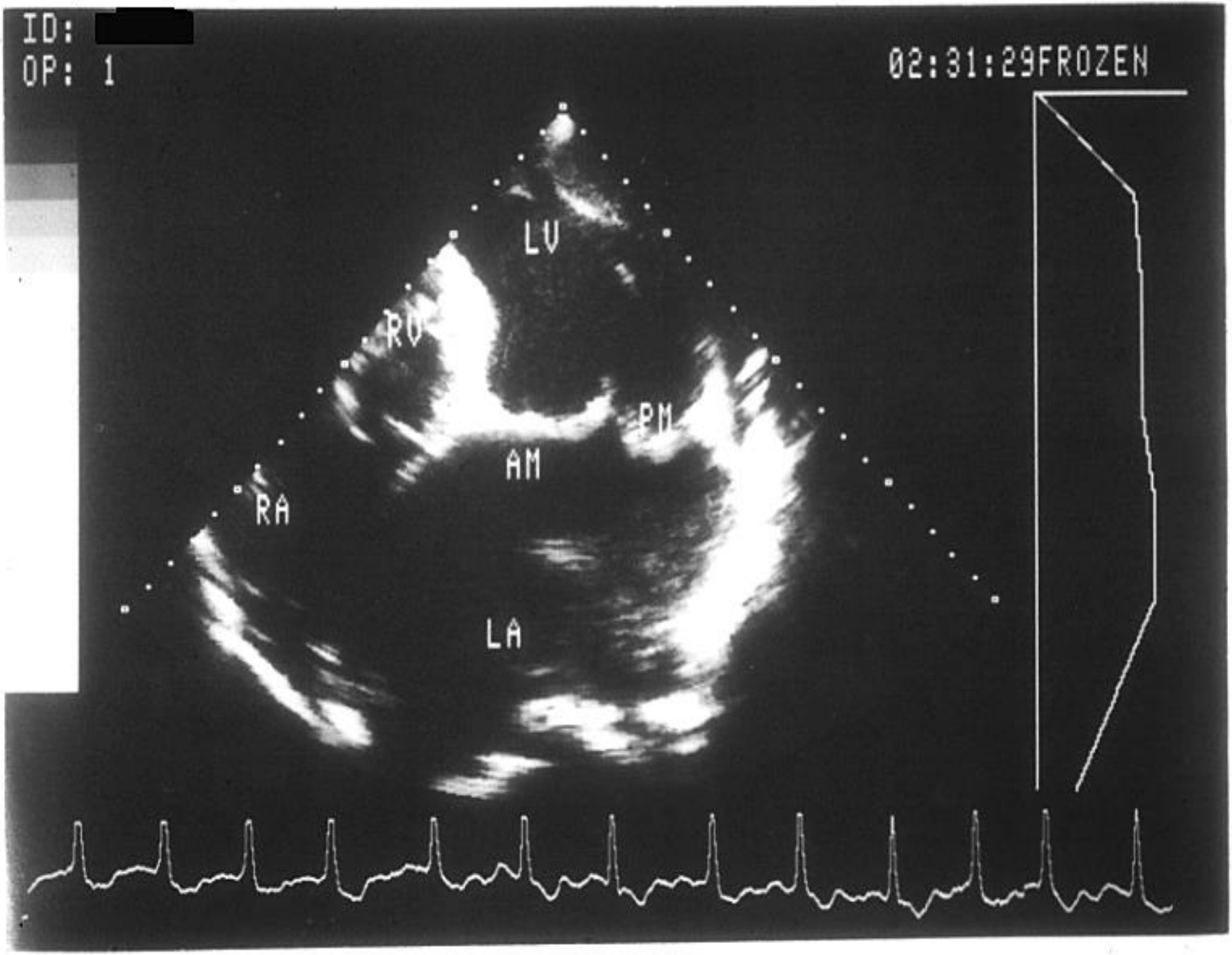
MVP - Non Invasive Testing:

- Echo = leaflet abnormalities
- Doppler = quantitate MR

- Stress Testing = false positive


Name: [redacted]
Date: 12/18/84
ID: 1187
OP: 1
SP: 50
ECHO DEP: 16cm
IREX
TF:A-LONG
VP:4
3.0MHz 14mm
PRE▲:3 POST▲:7
03:37:26FROZEN
RV
VS
LV
AV
AM
PM
LA
DA


ID 2311_____ OP: 04_
DATE 08/11/83 TIME 00:48:48
2.25 MHZ 21 CM FR: 8-69890
R+0.33 SEC
080 BPM
PM
LA
DIASONICS


ID: [redacted]
OP: 1
02:31:29FROZEN
LV
RV
PM
AM
RA
LA

MVP - Treatment:

- MR = as previously reviewed
  * May develop acute severe MR due to chordal rupture !!

Aortic Stenosis (AS):

- Etiology = Degenerative > congenital > rheumatic
  - Degenerative (senile calcific) = elderly/very elderly
  - Congenital = 1, 2 or 3 cusps (1-2% of population)
  - Rheumatic = rarely without mitral disease

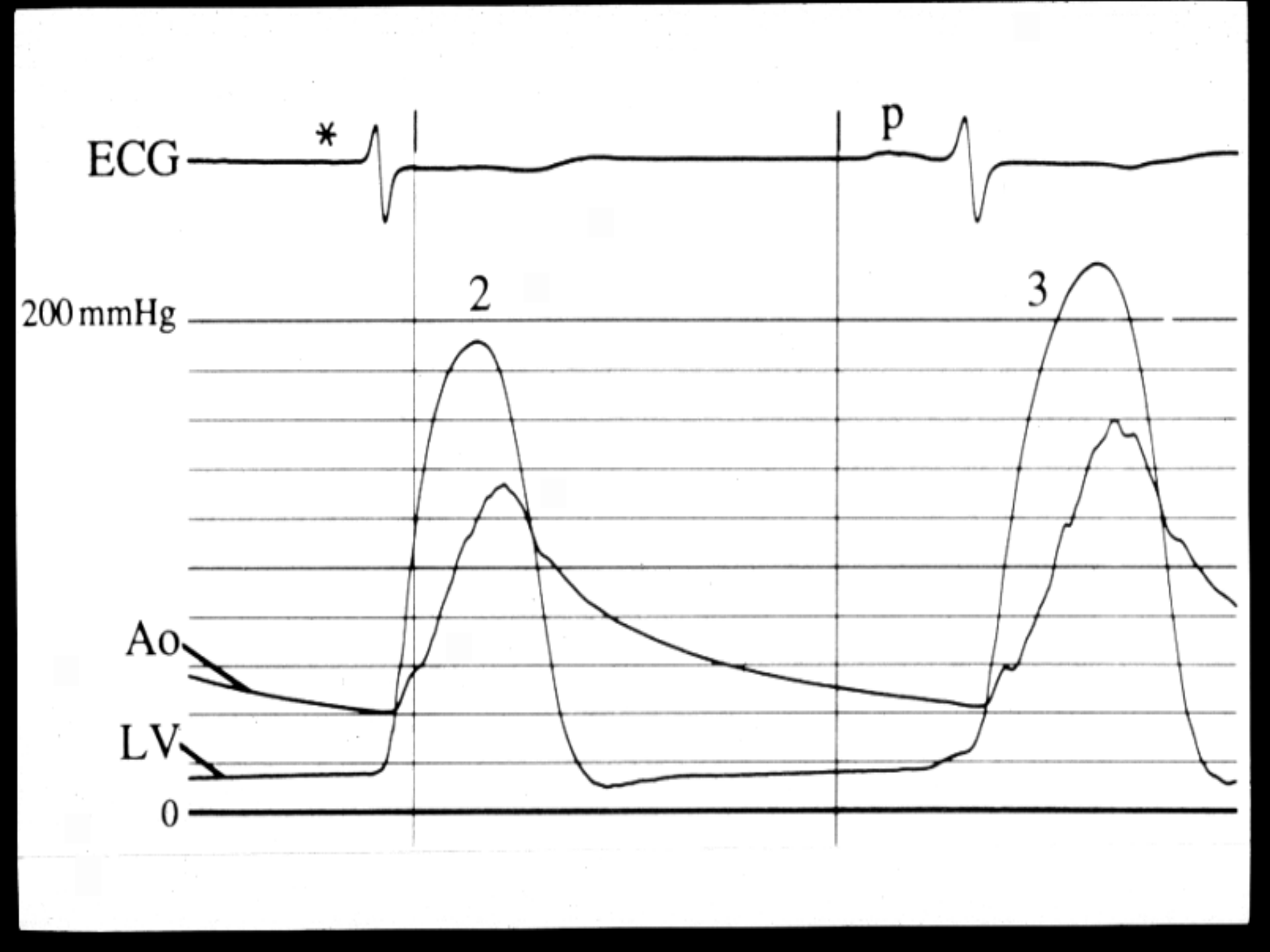
AS:

- Pathophysiology = pressure overload
  - Concentric LVH = mass/volume
  - Critical values: mean gradient or peak velocity

Mild AS = mean gradient < 20 mmHg, velocity 2.0-2.9 m/s
area > 1.5 cm2
Moderate AS = mean gradient 20-39 mmHg, velocity 3.0-3.9 m/s
area 1.0-1.5 cm2

SEVERE AS = gradient >/= 40 mmHg, velocity >/= 4.0 m/s
area </= 1.0 cm2 (area index </= 0.6 cm2/m2)

  - Rate of progression = variable


ECG
*
p
2
3
200 mmHg
Ao
LV
0

AS:

- Symptoms = average survival - 3 yrs after onset (untreated)
  - SOB - most common sx.
  - Angina, syncope, CHF
  - Colonic angiodysplasia = ↑ incidence

AS:

- Physical Exam
  - Pulses (carotid): may be insensitive in elderly
    * “parvus et tardus” (↓ amplitude with delayed upstroke)
    * pulsus alternans = ↓ CO
  - Apical impulse = sustained, left shift
  - Thrill = base, supra-sternal notch

AS - Auscultation:

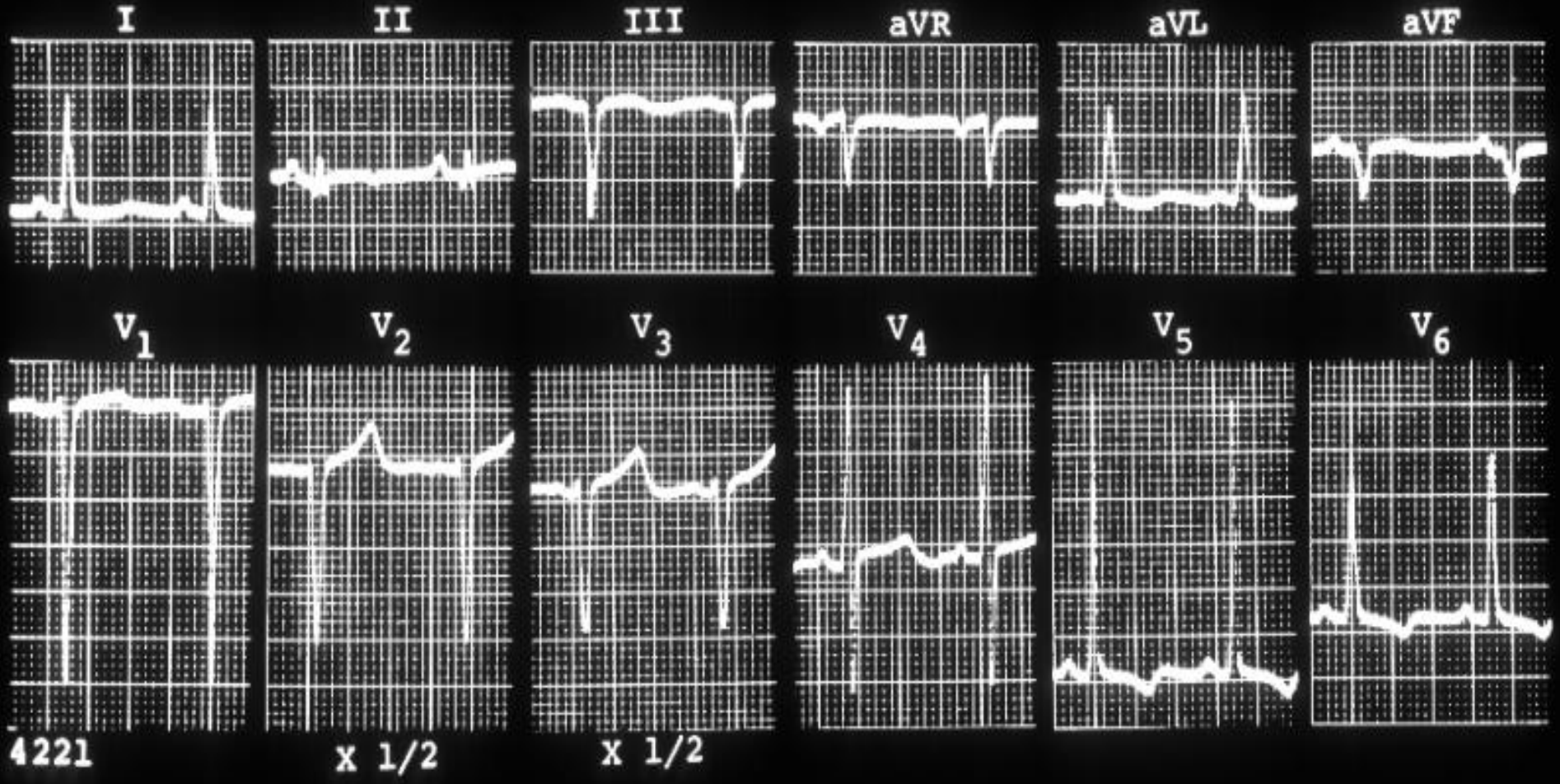
- S_2 = single or paradoxically split, decreased intensity
- Systolic ejection click = young, congenital
- Systolic ejection murmur
  - base to carotids
  - base to apex = Gallavardin phenomenon
  - severe AS = longer, louder, peaks later

AS:

- Dynamic auscultation:
  - * ↑ intensity = amyl nitrite, squatting
  - * ↓ intensity = valsalva

AS - Non Invasive Testing:

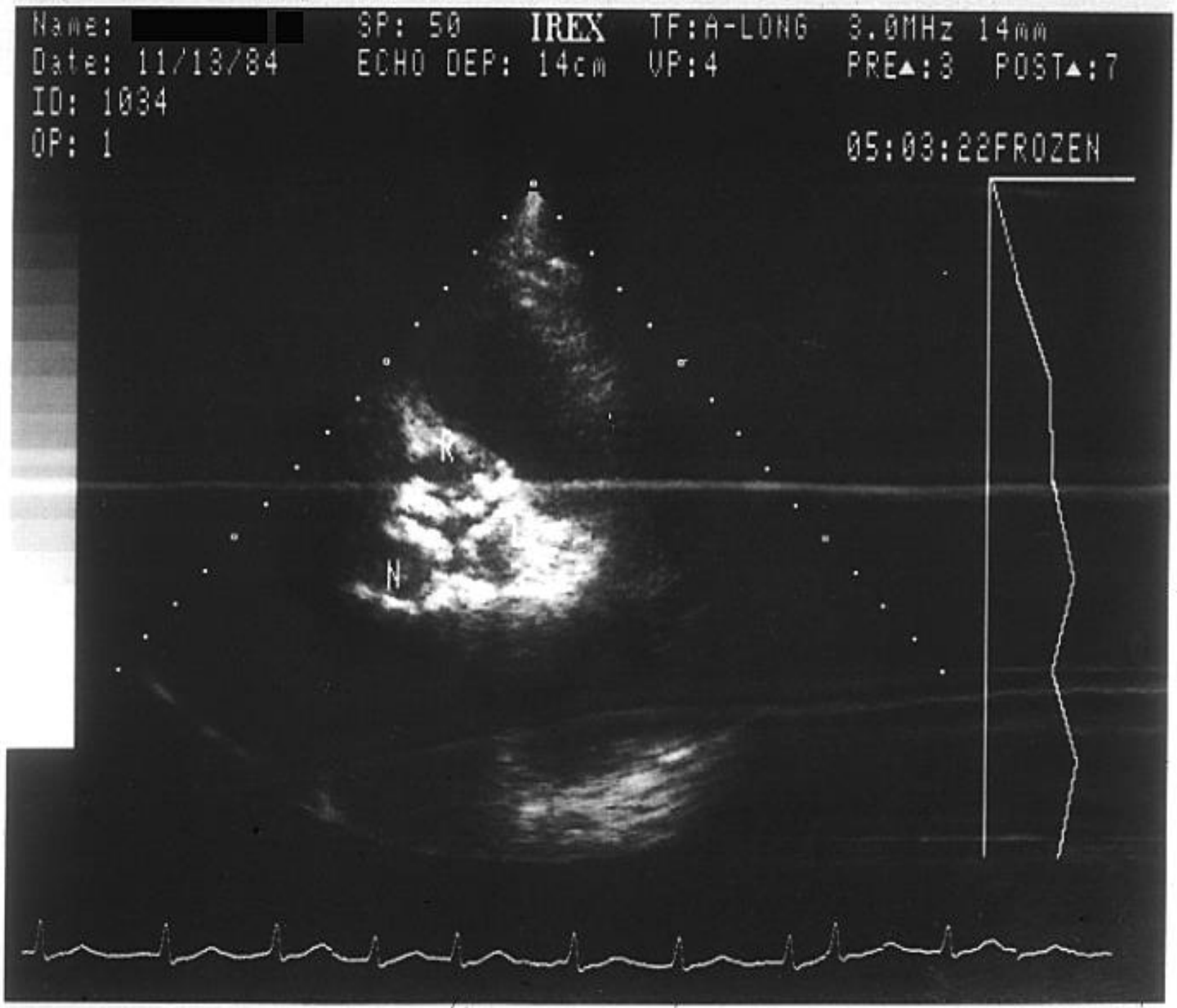
- EKG = LVH (80% with severe AS)
  LA enlargement
  AV block
- CXR = aortic dilatation (aortopathy)
  AV calcification
  may be “normal”

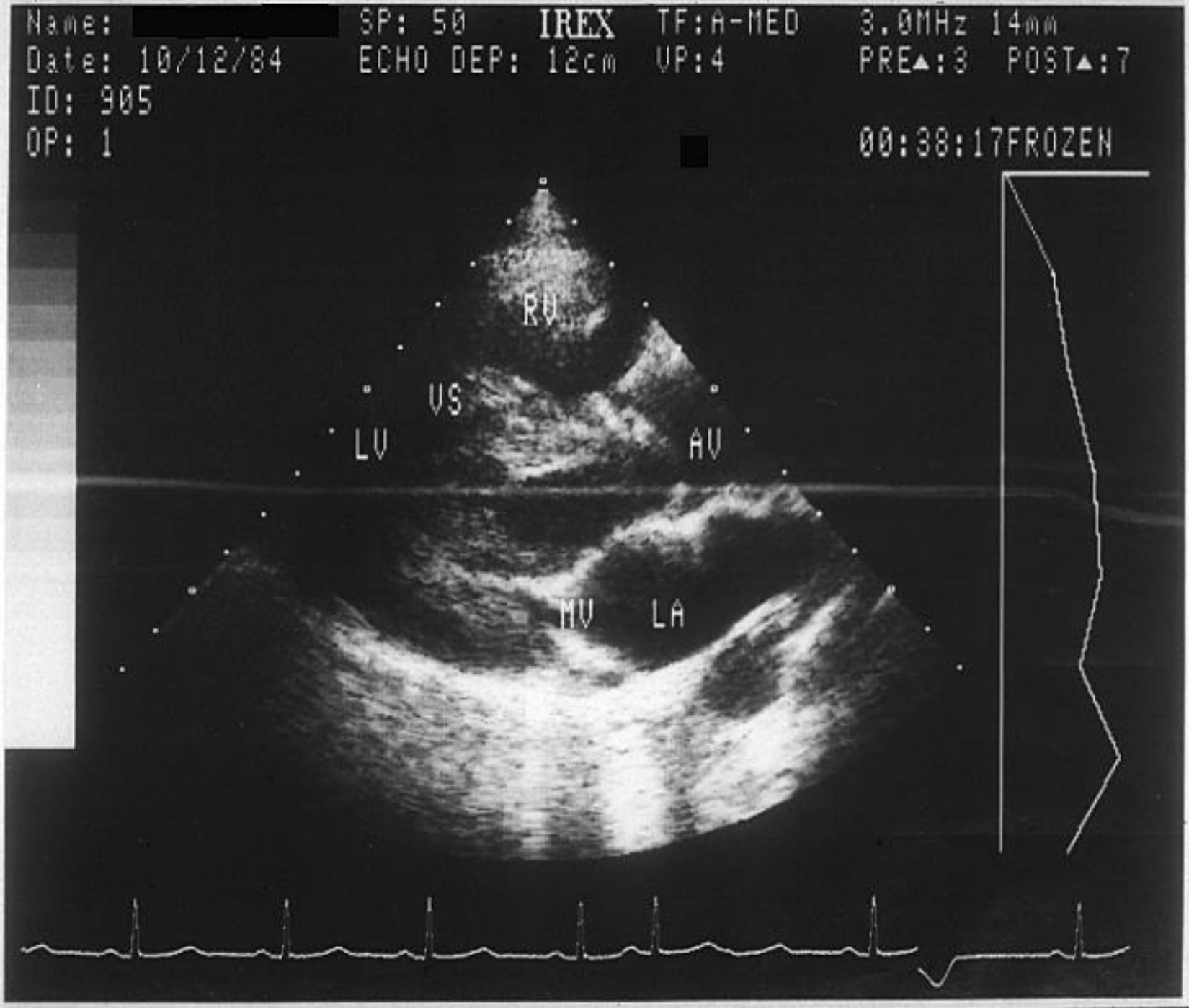

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
4221
x 1/2
x 1/2

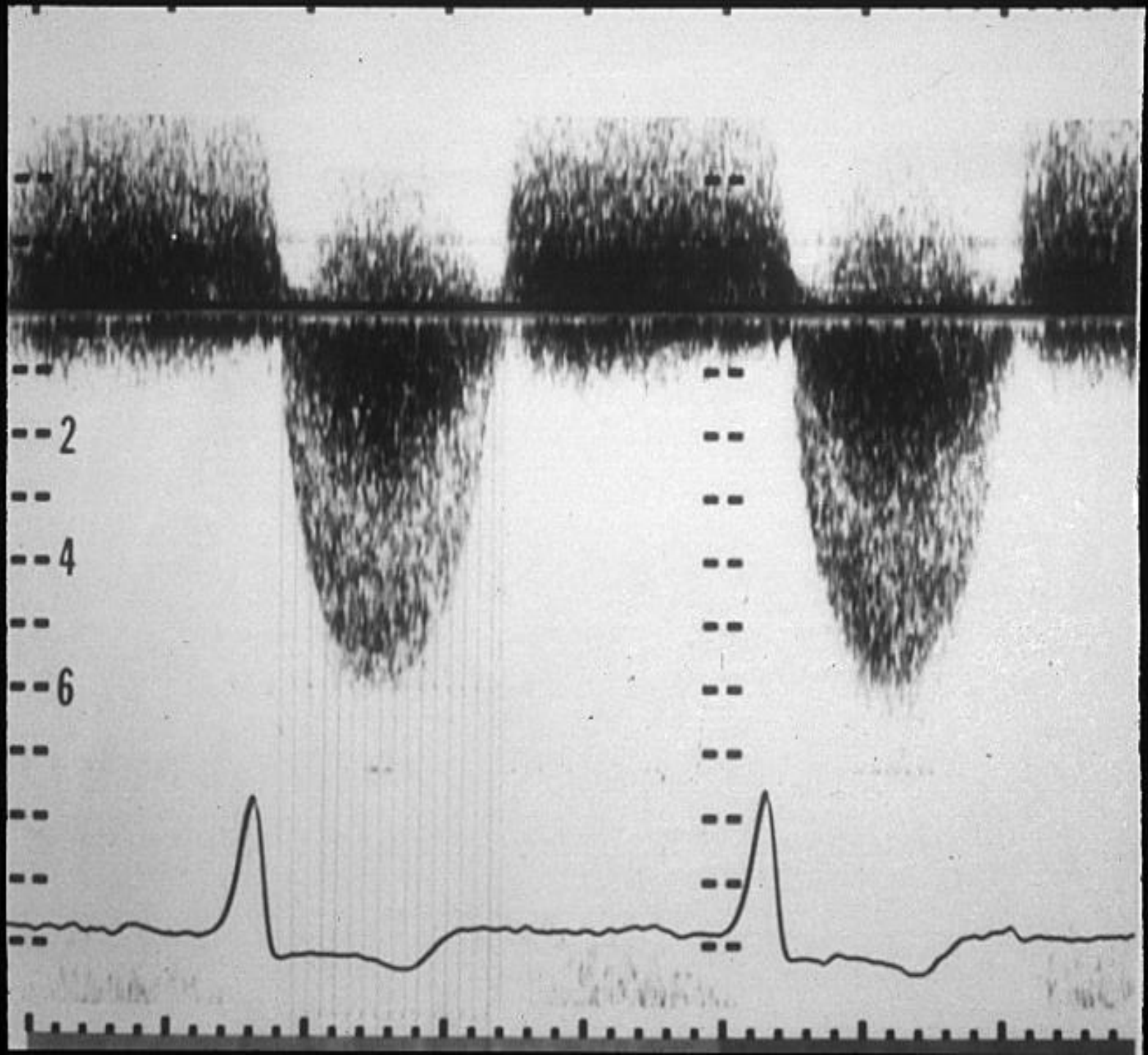
AS - Non Invasive Testing:

- Echo: Valve morphology
  LVH
  LV function
  Aorta (especially if bicuspid AV)

- Doppler: AV Gradient / flow velocity
  AV Area = may be discordant


Name: [redacted]
SP: 50
IREX
TF:A-LONG
3.0MHz 14mm
Date: 11/13/84
ECHO DEP: 14cm
VP:4
PRE▲:3
POST▲:7
ID: 1034
OP: 1
05:03:22FROZEN
K
N


Name: [redacted]
SP: 50
IREX
TF:A-MED
3.0MHz 14mm
Date: 10/12/84
ECHO DEP: 12cm
VP:4
PRE▲:3
POST▲:7
ID: 905
OP: 1
00:38:17FROZEN
RV
VS
LV
AV
MV
LA


2
4
6

AS:

- Treatment = this is a "surgical" disease.
  - Medical = caution w/ negative inotropes and preload/afterload reduction
    - * but…tx of HTN appropriate
  - The decision for surgery is based primarily on presence of symptoms (…but beware of the sedentary patient)

AS:

- Valve replacement = surgical or transcatheter (TAVR)

* Severe AS with symptoms
* Severe AS without symptoms:
  LVEF < 50 %
  or.. Undergoing OHS for other disease (eg:CAD) = mod-sev AS
  or..Very severe AS = mean grad >/= 60 mmHg, velocity >/= 5.0 m/s (class IIa)
  or.. Abnormal ETT (class IIa)
  or.. Bicuspid Aortic valve (regardless of severity) with dilated asc. aorta > 5.0 – 5.5 cm, or dia. increase >/= 0.5 cm/yr
  or ???.. Rapid progression of AS = increase in peak velocity > 0.3 m/s/year (class IIb)

AS: low flow / low gradient

- Discordant echo hemodynamics:
  * AVA < 1.0 cm2 … but…
  * flow vel 3-3.9 m/s
  * mean gradient 20-39 mmHg
- With normal or reduced LVEF
- Low-Dose Dobutamine stress echo = may be helpful for patient with reduced LVEF

(*goal: velocity >/= 4.0 m/s or mean gradient >/= 40 mmHg with area </=1.0 cm2 at any dobutamine dose).

Chronic Aortic Regurgitation (AR):

- Etiology = abnormality of leaflets or aortic root
- Pathophysiology = volume and pressure overload
  - * concentric and eccentric hypertrophy
- Acute AR = rapid LV failure
  - * absence of “classic” findings

AR:

- Symptoms = late appearance
  - SOB
  - LV failure
- LV may begin to fail before symptom onset
- Absence of symptoms does not preclude severe AR

AR - Physical Exam:

- Pulses = bounding, wide pulse pressure
  - Quinckes
  - Corrigans
  - Bisferiens
- Apex = diffuse, hyperdynamic, left shift

AR - Physical Exam:

- S3 = LV failure
- diastolic decrescendo murmur
    - high pitch, base
    - severity = duration ??
- Austin - Flint murmur = functional diastolic rumble mimics MS (but no opening snap)
- Systolic ejection murmur

AR - Physical Exam:

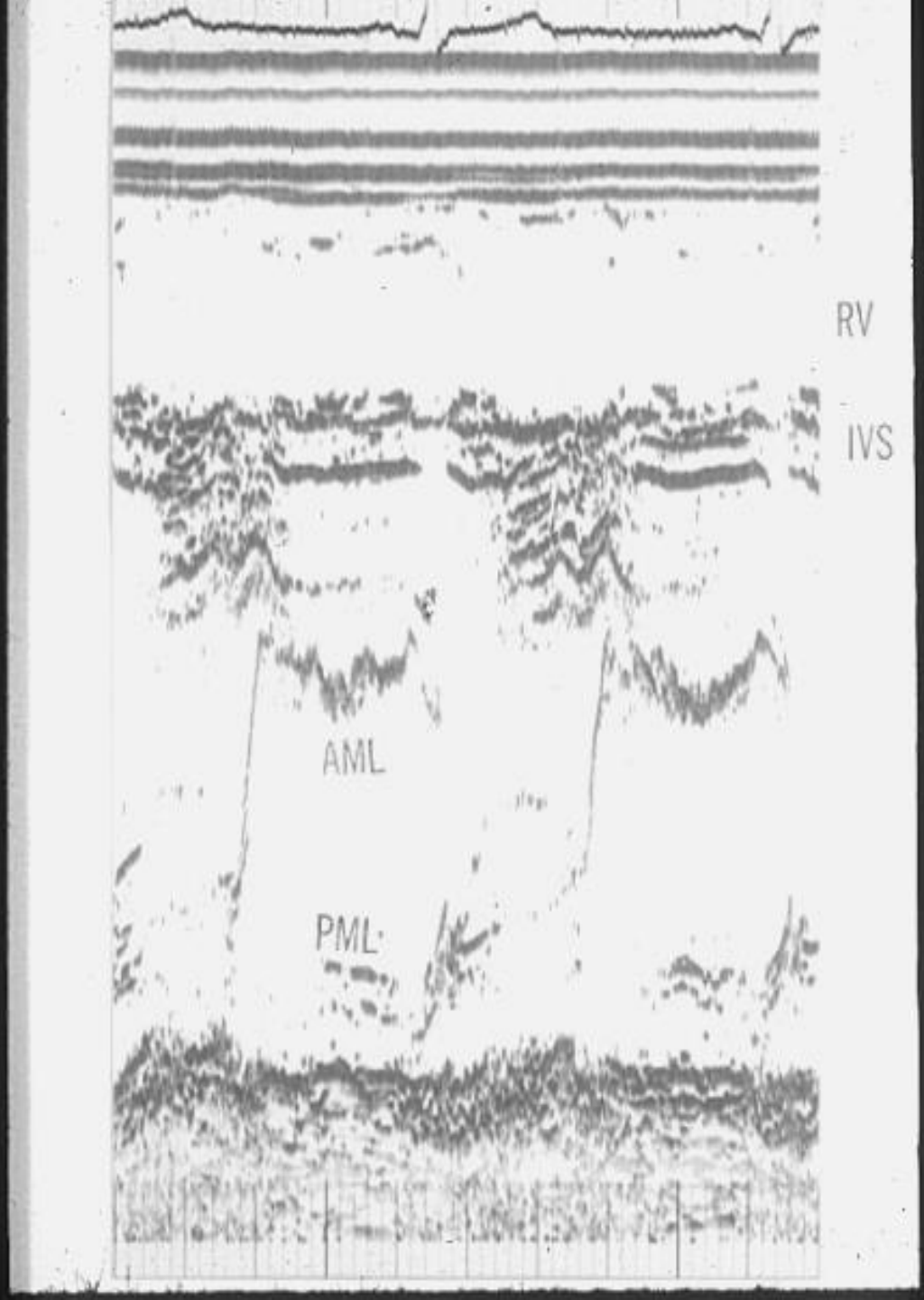
- Dynamic Auscultation

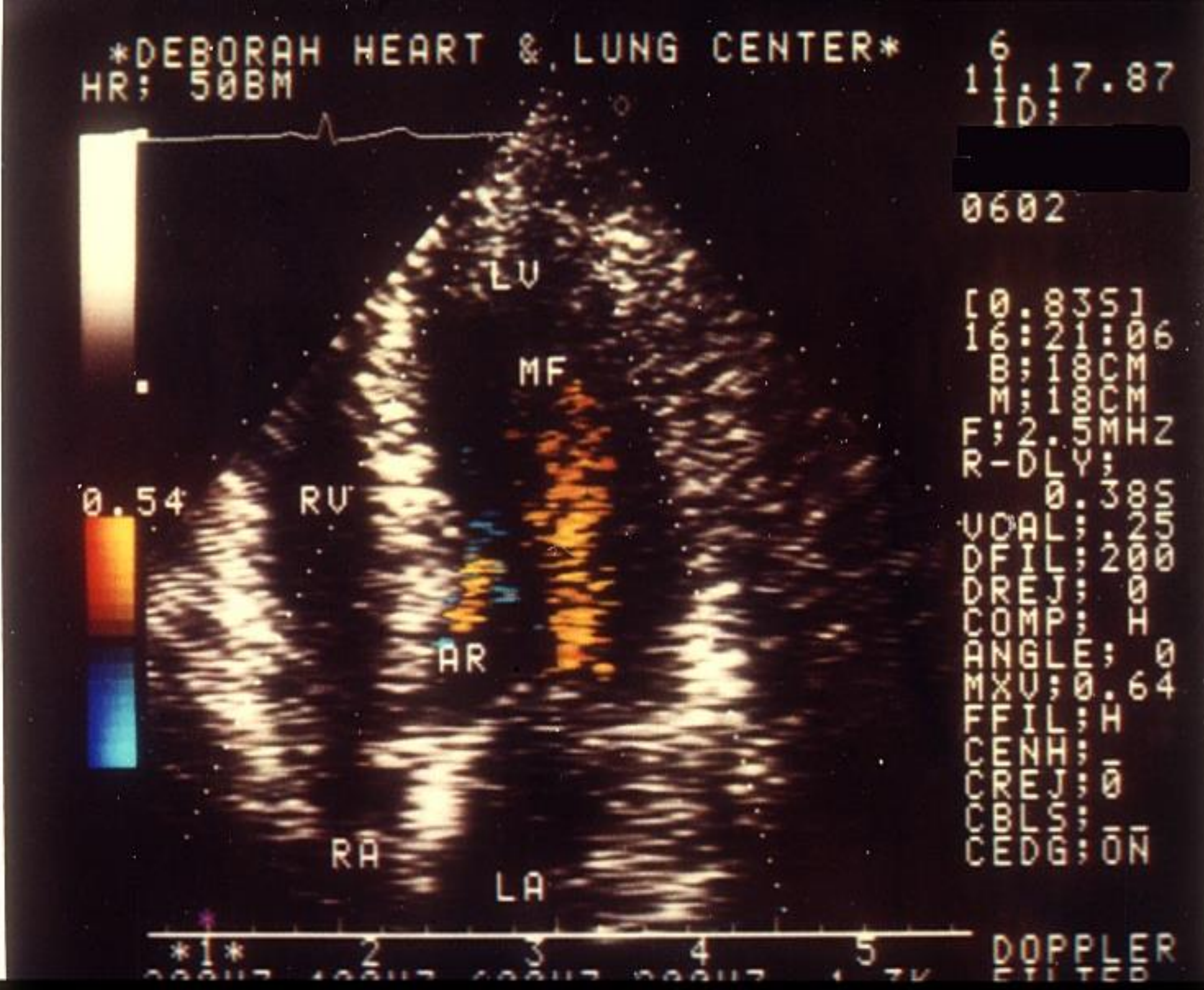
  * ↑ intensity = pressors, squatting, isometrics

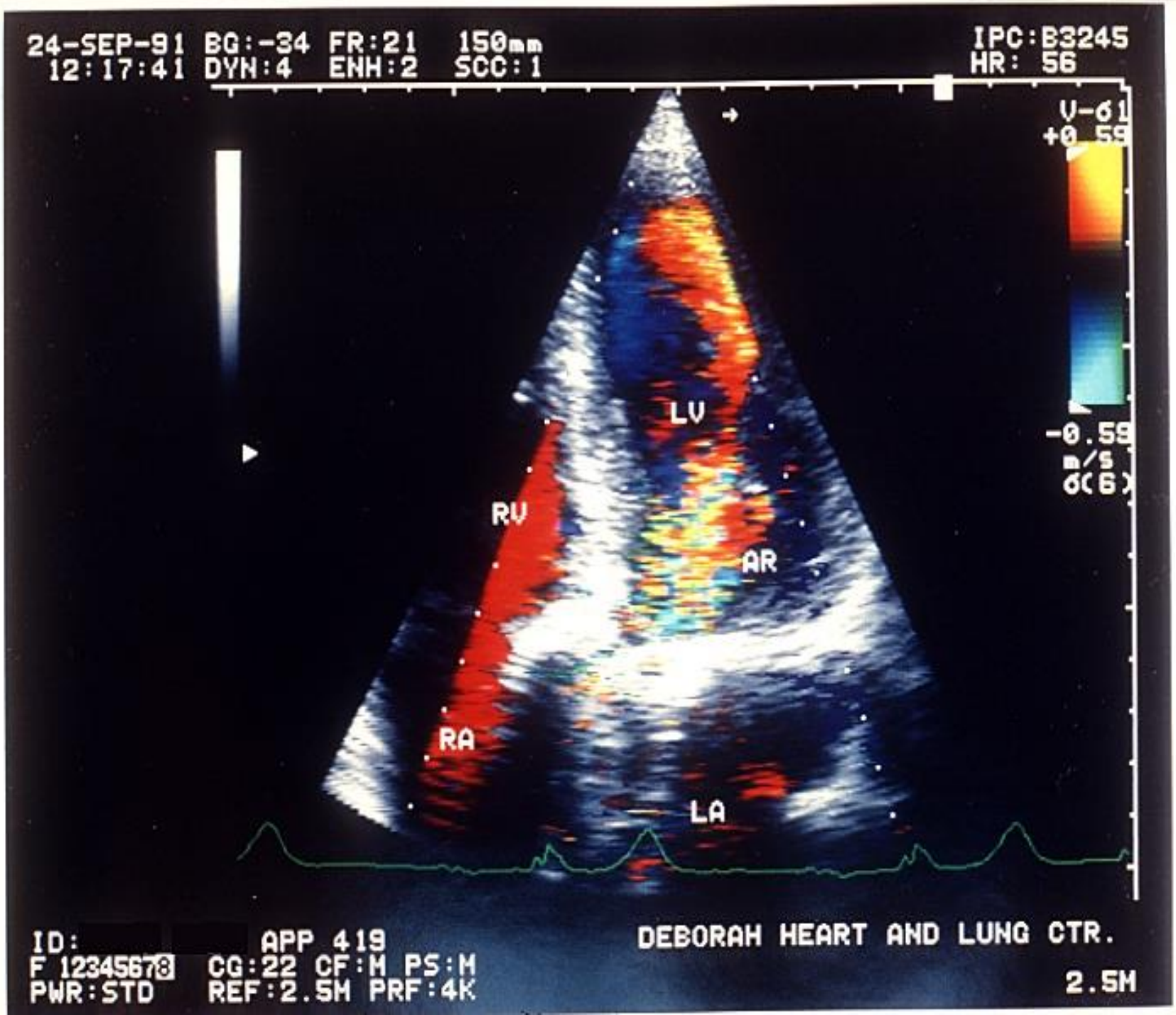
  * ↓ intensity = amyl nitrite, Valsalva

AR - Non Invasive Testing:

- EKG = LVH
- CXR = cardiomegaly, dilated aorta
- Echo = etiology, LV size and function
- Doppler (color flow) = quantitate severity
- Cardiac MRA = good option if echo equivocal, but $$ and availability ?


RV
IVS
AML
PML


DEBORAH HEART & LUNG CENTER
HR; 50BM
6
11.17.87
ID;
[redacted]
0602
LV
MF
RV
AR
RA
LA
0.54
[0.83S]
16:21:06
B;18CM
M;18CM
F;2.5MHZ
R-DLY;
0.38S
VCAL;.25
DFIL;200
DREJ; 0
COMP; H
ANGLE; 0
MXV;0.64
FFIL;H
CENH;_
CREJ;0
CBLS;_
CEDG;ON
1 2 3 4 5
DOPPLER


24-SEP-91 BG:-34 FR:21 150mm
12:17:41 DYN:4 ENH:2 SCC:1
IPC:B3245
HR: 56
V-61
+0.59
-0.59
m/s
dC(6)
LV
RV
AR
RA
LA
ID:
APP 419
F 12345678 CG:22 CF:H PS:M
PWR:STD REF:2.5M PRF:4K
DEBORAH HEART AND LUNG CTR.
2.5M

AR - Treatment:

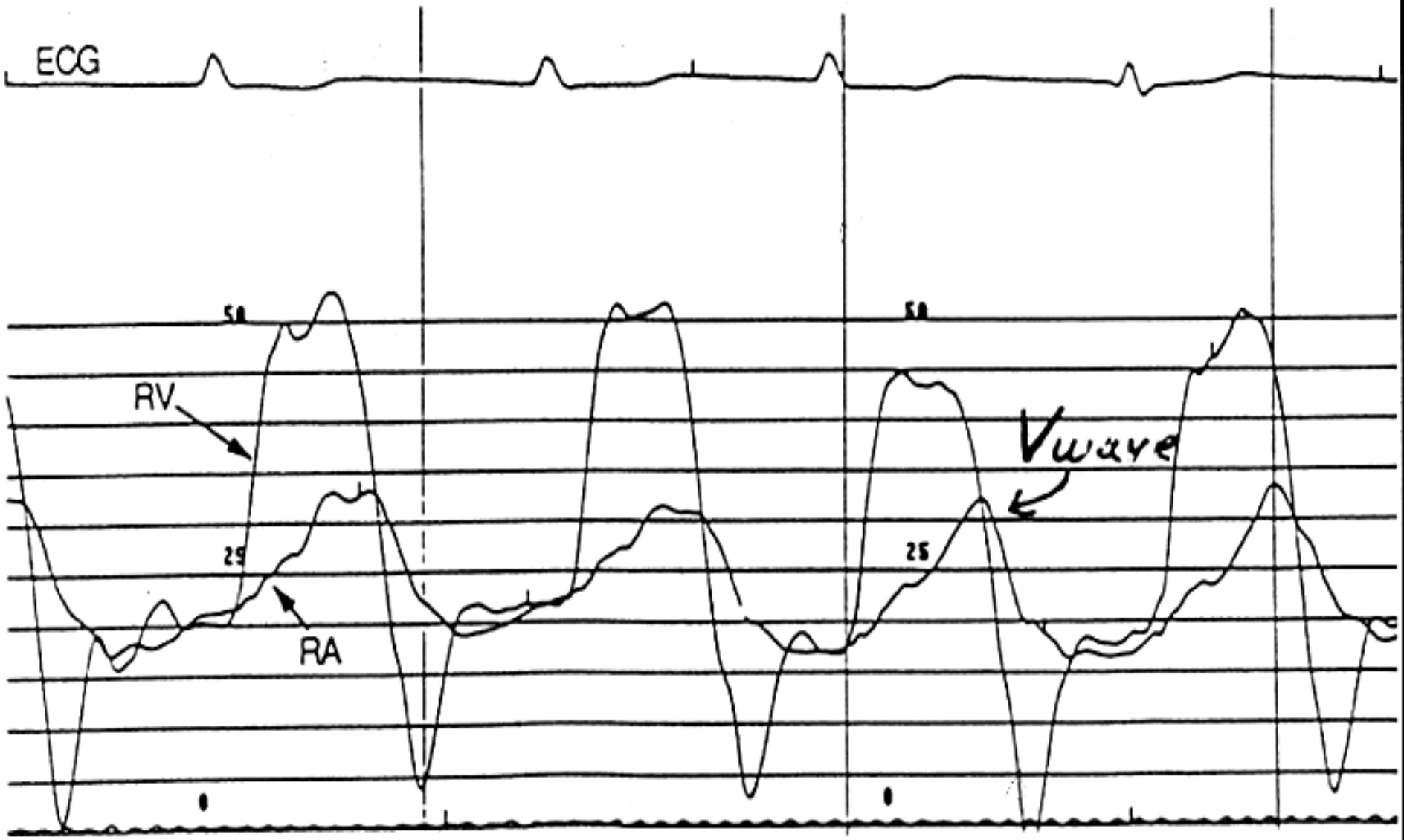
- Medical = afterload reduction for HTN.
  - * ACEi / ARB, dihydropyridine CCB
  - * Medical tx is NOT a substitute for AVR
- Surgical = AV replacement
  - Severe AR with symptoms
  - Severe AR without symptoms:
    - * EF < 50% (class I)
    - * or .. LV end-systolic dimension > 50 mm (class IIa)

Tricuspid Stenosis:

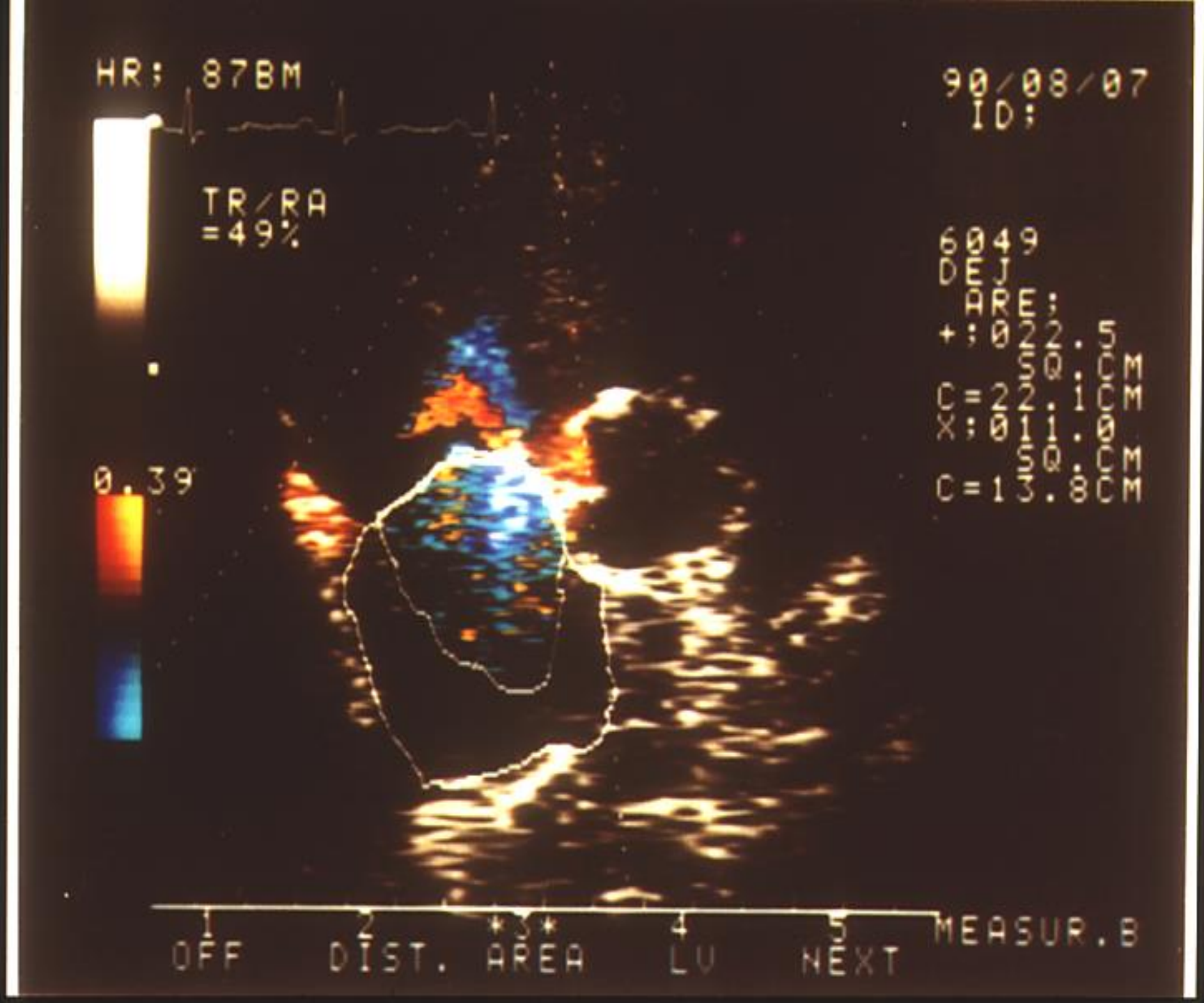
- Etiology = rheumatic, congenital, carcinoid

Tricuspid Regurgitation: 95% functional

- Etiology = RV dysfxn/dilatation, TVP, Ebstein's, Rheumatic fever, XRT, carcinoid, PPM or ICD
- Clinical =
  - Sx = right heart failure
  - PE = holosystolic murmur (LLSB) … may be inaudible
    - * Increases with inspiration = Carvallo's sign
    - * JVP = large "V" or "C-V" waves
    - * Hepatic pulsation (systolic)



TRICUSPID REGURGITATION
'V' - WAVES


HR: 87BM
90/08/07
ID:
TR/RA
=49%
6049
DEJ
ARE;
+:022.5
SQ.1 MM
C=22.1
X:011.0CC
SQ.CC MM
C=13.8CC
0.39
1
2
3
4
5
OFF
DIST.
AREA
LV
NEXT
MEASUR.B

Pulmonic Regurgitation =

- Graham - Steel murmur = PR 2° to pulm. HTN

Pulmonic Stenosis =

- Etiology = congenital, carcinoid

Congenital Heart Disease:

- Incidence = 0.8% of births (excluding Bicuspid AV)

VSD = 30%	Coarctation Aorta = 7%
ASD = 10%	AS = 7%
PDA = 10%	Tetralogy of Fallot = 6%
PS = 7%	Transposition = 4%

* Bicuspid aortic valve = 1-2 % of gen population

Congenital - Syndrome/Association:

- Noonan = PS
- Holt - Oram = ASD
- Kartageners = dextrocardia, sinusitis, bronchi-ectasis
- Muscular Dystrophy = cardiomyopathy
- Downs Syndrome = ASD, VSD, AV-valve regurg.
- Williams Synd. = supravalvular AS
- Turners Synd. = coarctation of aorta, bicuspid AV

Bicuspid Aortic Valve (BAV)

- 1-2% population
- AS &/or AR
- Screen 1st degree relatives = ~ 25% incidence of bicuspid valve or aortopathy
- Associated with aortopathy = aneurysm, coarct, dissection
- Can have severe ascending aorta dilatation without signif. valve dysfunction (either level of the sinuses or tubular asc. aorta)

  * Surgery:

    if: asc. aorta dia > 5.0 – 5.5 cm

    if: asc. aorta dia > 4.5 cm (if AVR required for sev. valve dz)

  * Monitor (echo, MRA, CTA):

    q 1 yr: if ascending aorta >/= 4.5 cm

Congenital:

Coarctation of the aorta = narrowing of aorta in region of ligamentum arteriosum adjacent to left subclavian artery origin

*Clinical = HTN, delayed lower extrem. pulses (brachio-femoral delay)

*Associations = bicuspid AV, congenital aneurysm of Circle of Willis, sub-aortic stenosis, VSD, mitral abn.

*CXR = rib notching

*complication = HTN, aortic dissection, rupture

Congenital

- Coarctation (cont'd):

  Pre- and post- repair concerns:

  * HTN
  * accelerated CAD
  * CHF
  * dissection
  * CVA, intracerebral hemorrhage
  * Aneurysm

* These patients MUST be monitored lifelong following repair (with intermittent imaging of the aorta)

Coarctation of Aorta

Atrial Septal Defect (ASD):
Secundum (75%), Primum(15%), Sinus Venosus, Coronary Sinus

Secundum ASD = most common

*30 - 40% of congenital heart disease in adults > 40 yo

* Mid-septal defect

* Increased incidence MVP

ASD - Pathophysiology:

- Shunt = left to right
  - *right heart volume overload
  - *Increased pulm. blood flow
- Clinical = may be asympt. for decades
  - * pulmonic systolic ejection murmur
  - * right sided diastolic rumble
  - * fixed widely split S2

ASD (cont'd):

- Natural History = dependent on size of shunt
  - Right heart failure
  - Atrial arrhythmias
  - Pulm arterial HTN
  - Paradoxical embolism

ASD - Diagnostics:

- EKG = RAD, RAE, RVH, inc. RBBB (secundum)
- CXR = RA, RV, PA enlarge, pulm. vascular markings
- Echo (TTE, TEE):

  paradoxical septal motion
  diastolic ventricular septal flattening
  RAE, RV dilatation

  > d/t RV vol. overload

  "Bubble" test - shunt visualized
  Color Doppler - shunt visualized

*MRI = may be useful if echo findings ?

*Cath = O2 "step up" in RA (>/= 7% vs vena cavae)

ASD - Treatment

Indications for Closure (surgical or percutaneous):

* Right heart enlargement without severe pulm HTN
* Hx of paradoxical embolus = ?
* Orthodeoxia – platypnea = ?

Patent Ductus Arteriosus (PDA):

- Anatomy = connects pulm. art. and descending aorta
- Assoc. lesions = ASD, VSD
- PE = continuous "machinery" murmur (left infraclavicular area)
- Clinical course = dep. on size of shunt

  *LV vol. overload, sev. PAH, Eisenmenger's (differential cyanosis and clubbing)
- Treatment (in adult) = device or surgical closure

  * Left heart enlarge with net left to right shunt without severe pulm HTN

Ventricular Septal Defect (VSD):

- Most common defect at birth
- Seldom seen in adults unless small
- Holosystolic murmur LLSB
- Spontaneous closure frequent = if small
- L $\rightarrow$ R shunt = size dictates sequelae

  *LV vol. overload, pulm HTN
- Severe pulm. HTN = shunt reversal (Eisenmengers Synd.)

Ventricular Septal Defect (cont'd)

- Echo = test of choice
- Surgical closure:

  * Evidence of LV volume overload and Pulm /Systemic flow ratio >/= 1.5 without severe Pulm HTN = class I

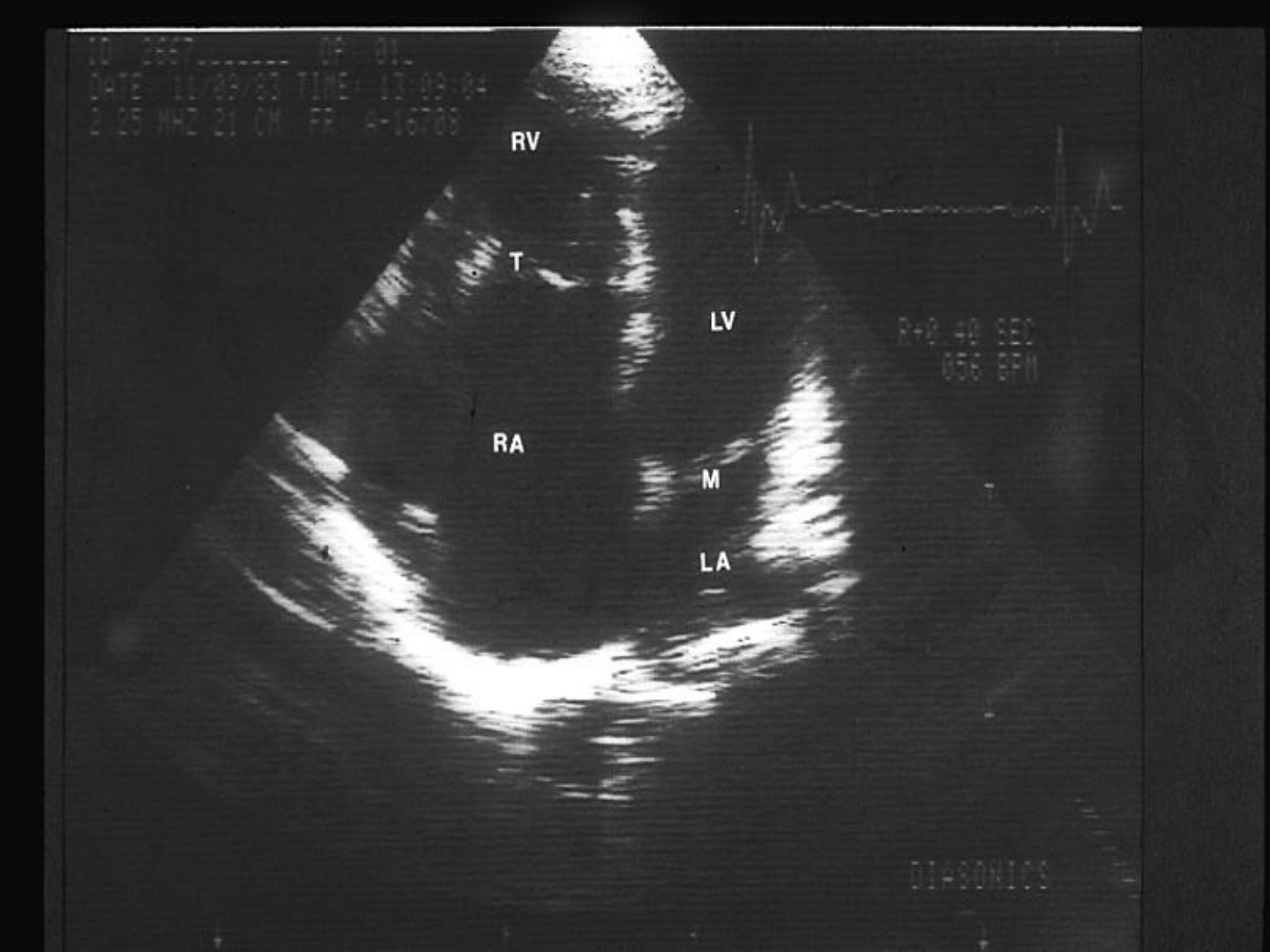
Tetralogy of Fallot (TOF):

- Tetrad = VSD, PS, RVH, over-riding aorta

  * Hemodynamic sequelae d/t size of VSD and degree of RV outflow obstruction
- Squatting = relief of hypoxic episode
- Occasional survivor to adulthood
- Most common anomaly resulting in cyanosis after one y.o.

Ebstein's Anomaly:

- Congenital TR
  "Atrialized" right ventricle
- Associated anomalies = ASD, VSD, PS, WPW (Wolf-Parkinson-White)
- Adult presentation =
  * Right heart failure
  * Arrhythmias


RV
T
LV
RA
M
LA

Transposition of Great Arteries (TGA):

D - Transposition = 2 separate circulations

*Aorta arises from RV

*Pulm. artery arises from LV

*Need shunt to survive

> AV concordance, ventriculo-arterial discordance

TGA:

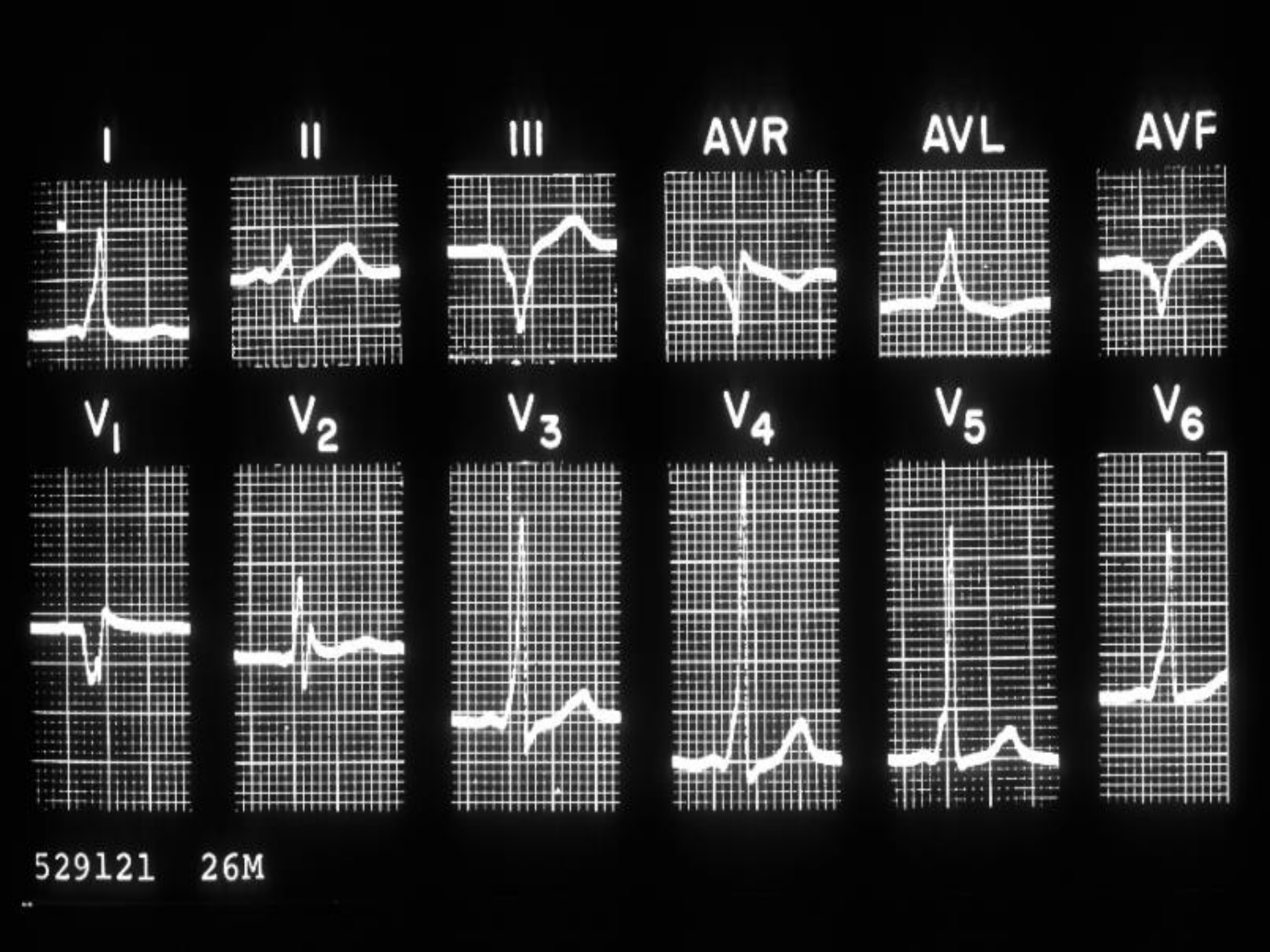
- L – Transposition (congenitally corrected):
  * AV discordance and ventriculo-arterial discordance
  * Morphologic RV = systemic ventricle
  * Morphologic LV = venous ventricle
  * Function = blood follows normal course
  * Survival into adulthood
  *Problems = systemic A-V valve regurg and systemic ventricular failure

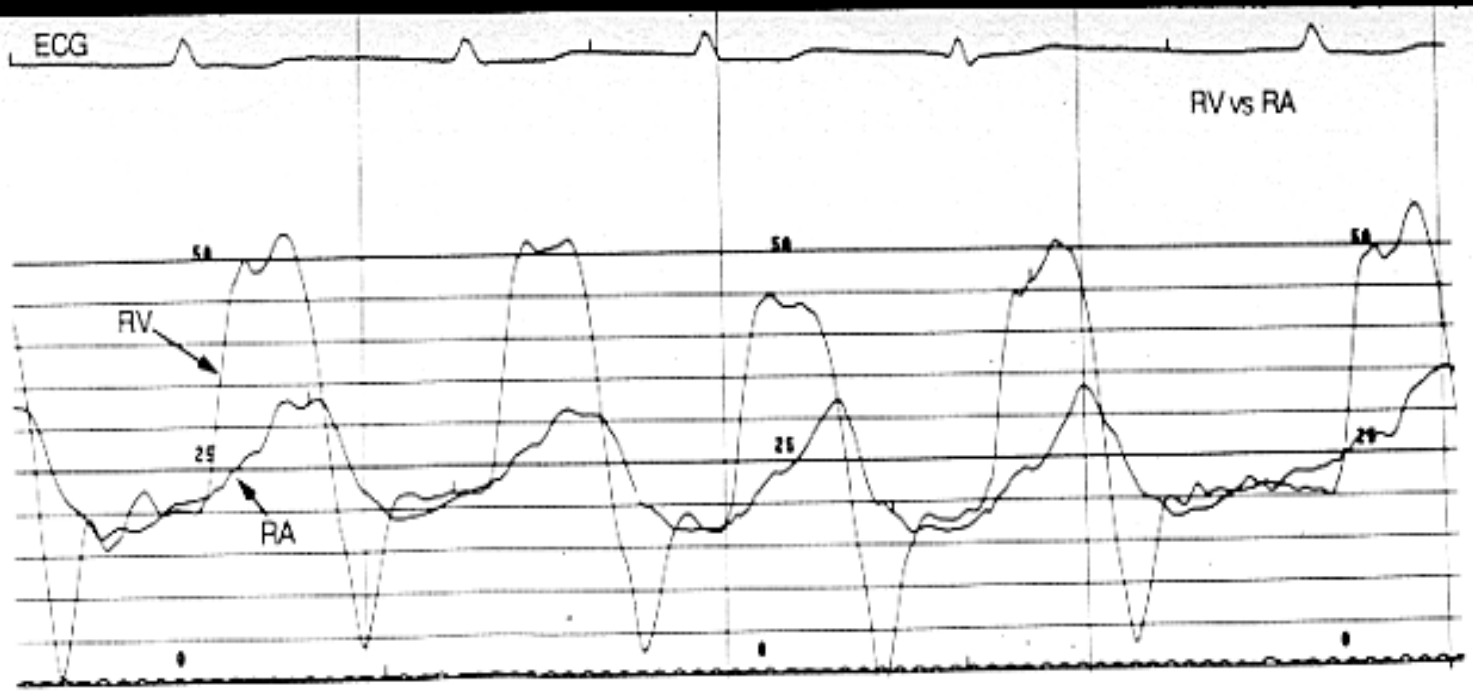
Congenital Disease - Summary:

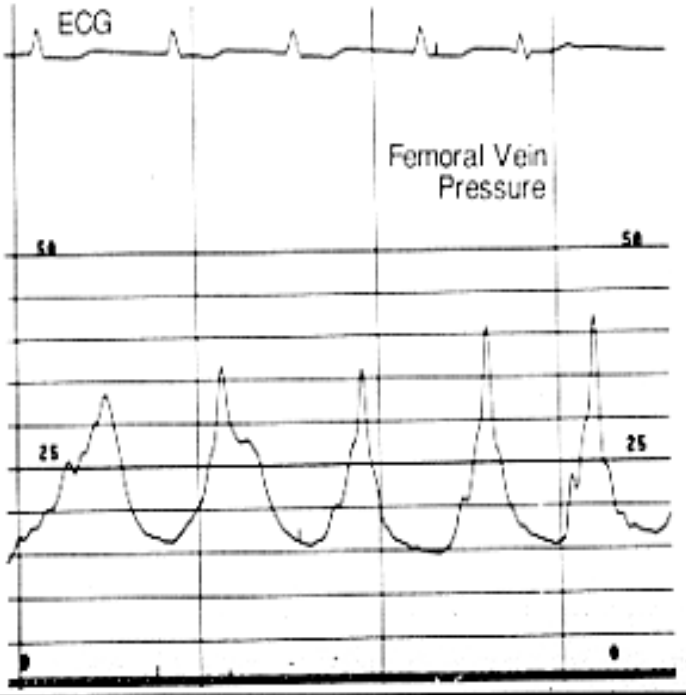
- L $\rightarrow$ R shunt = non-cyanotic
  - * ASD, VSD, PDA, Persistent truncus
- R $\rightarrow$ L shunt = cyanotic
  - * TOF ($\pm$ cyanosis)
  - * Tricuspid atresia
  - * Complete transposition (“D”)
  - * Double outlet RV

Congenital Disease - Summary:

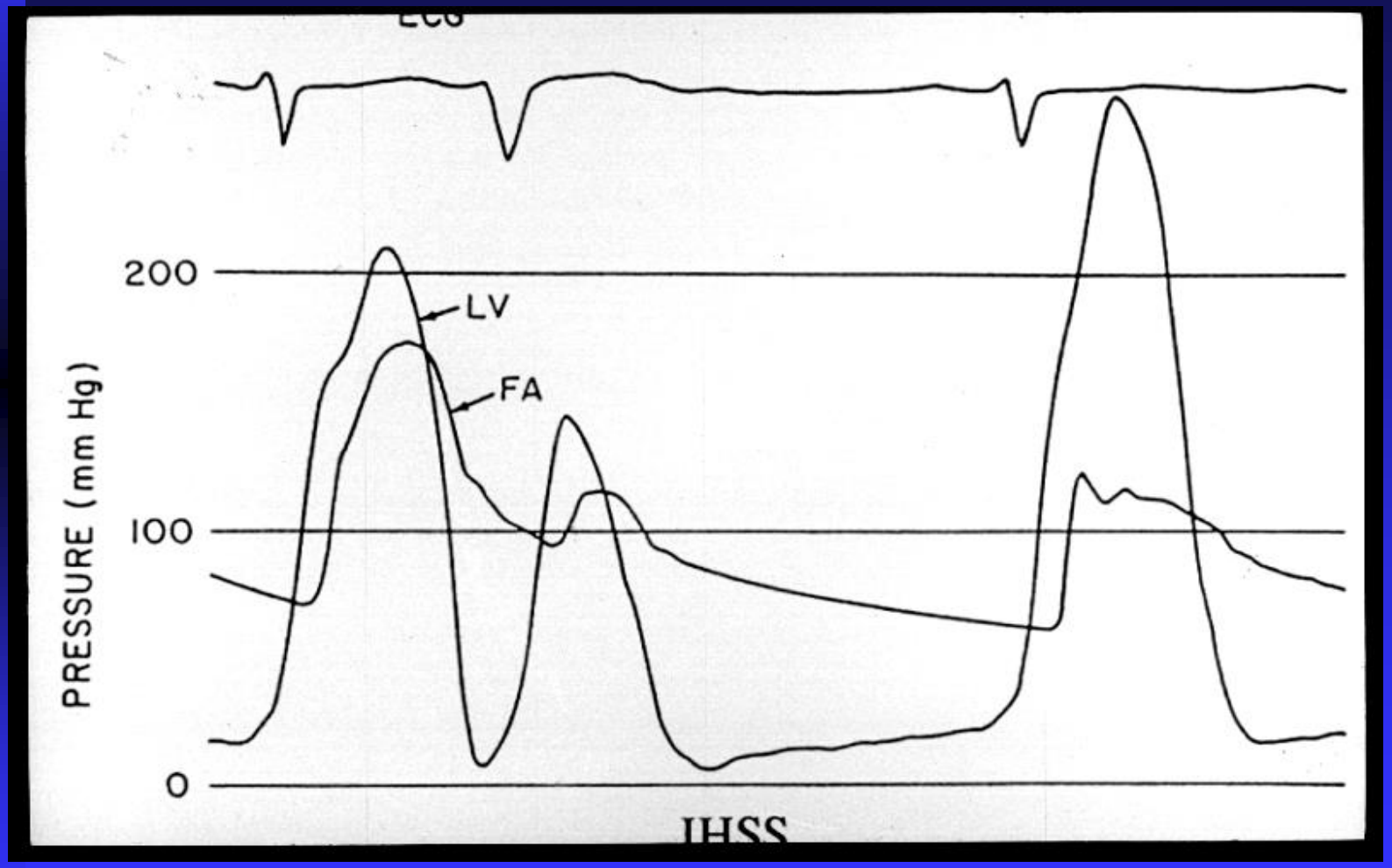
- Survival to adulthood:
  - * Bicuspid aortic valve
  - * Coarctation of aorta
  - * Pulmonic stenosis
  - * Secundum ASD
  - * PDA

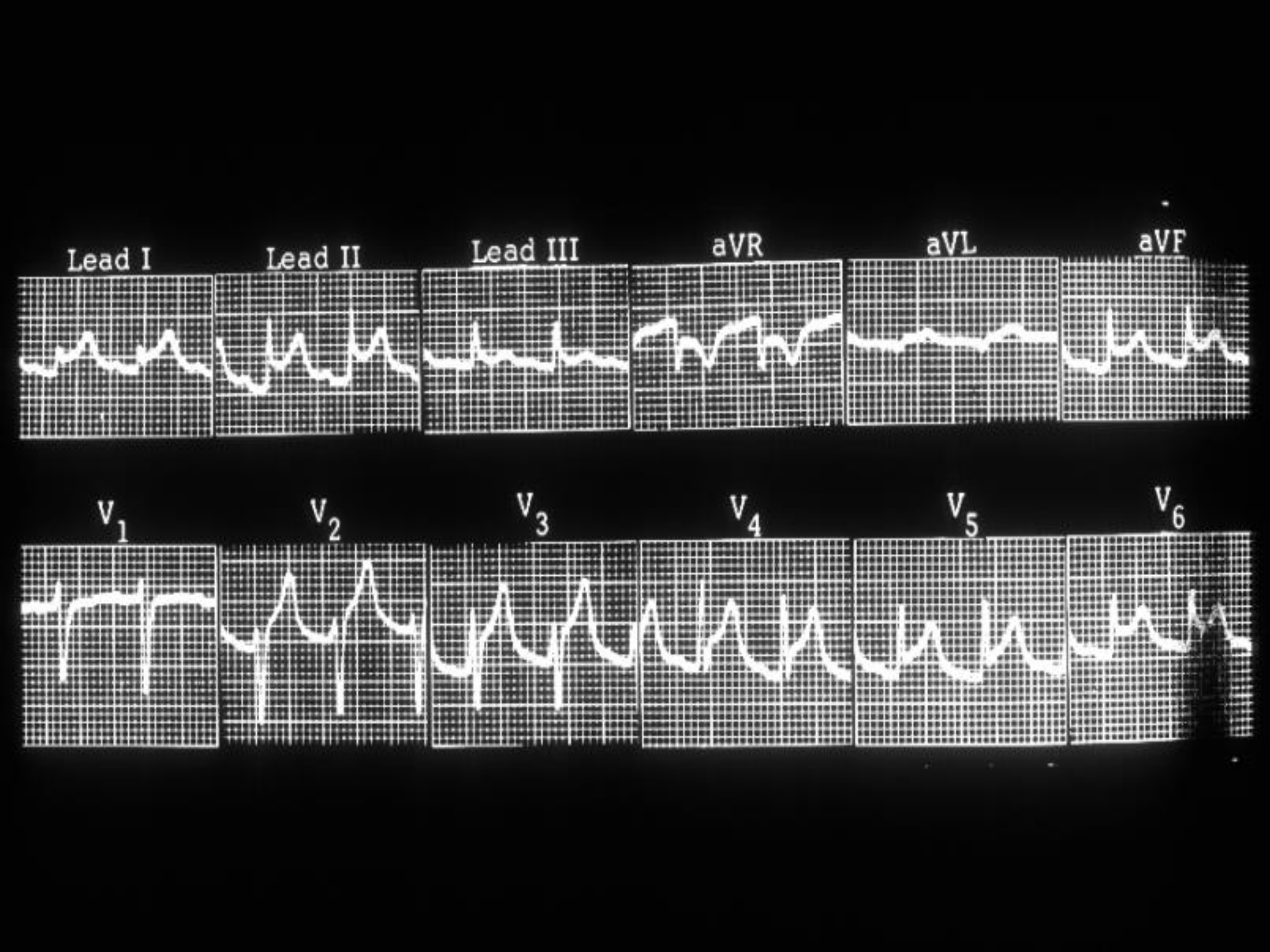

I
II
III
AVR
AVL
AVF
V1
V2
V3
V4
V5
V6
529121 26M

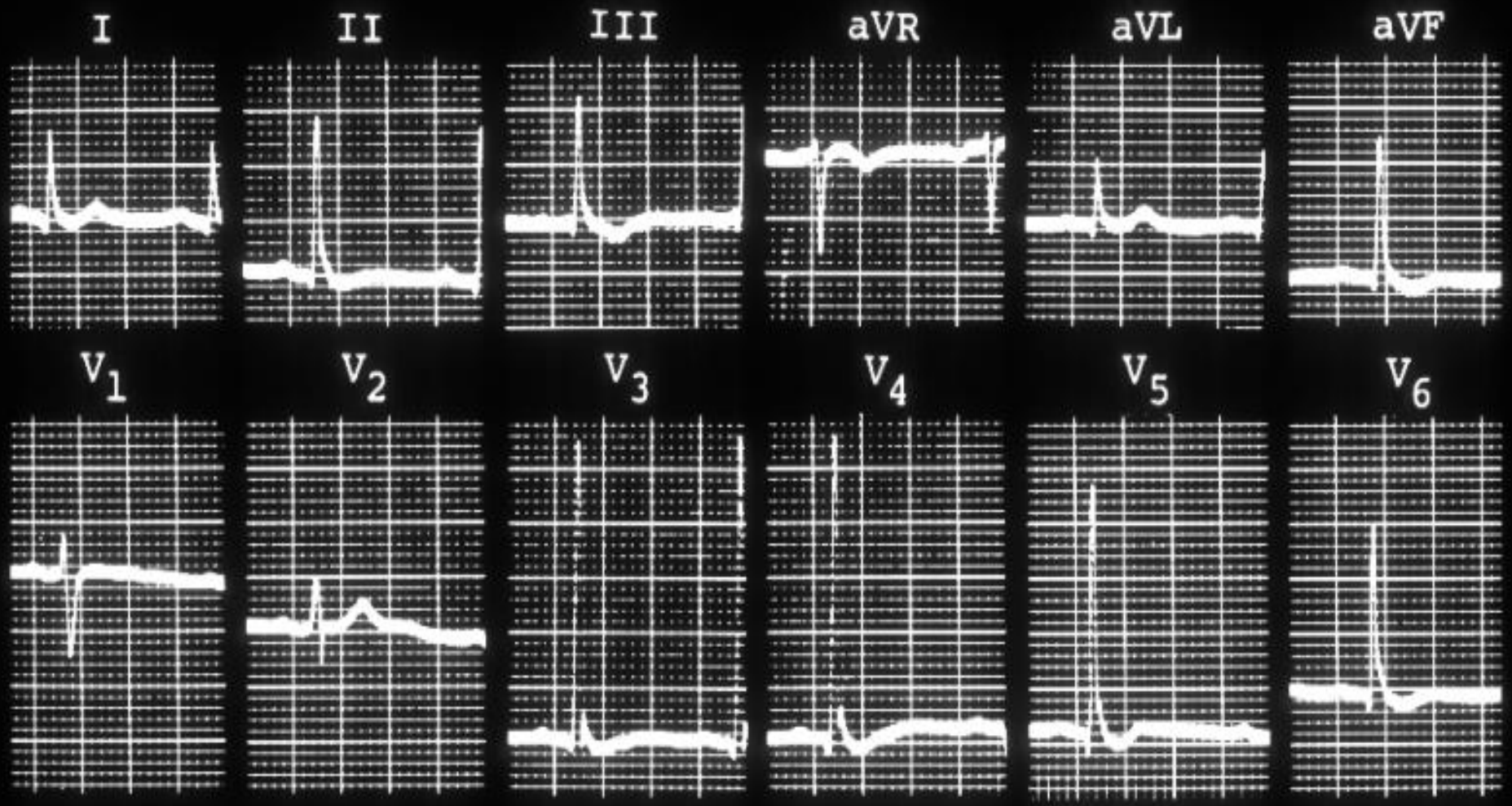

ECG
RV vs RA
RV
RA
50
25
0

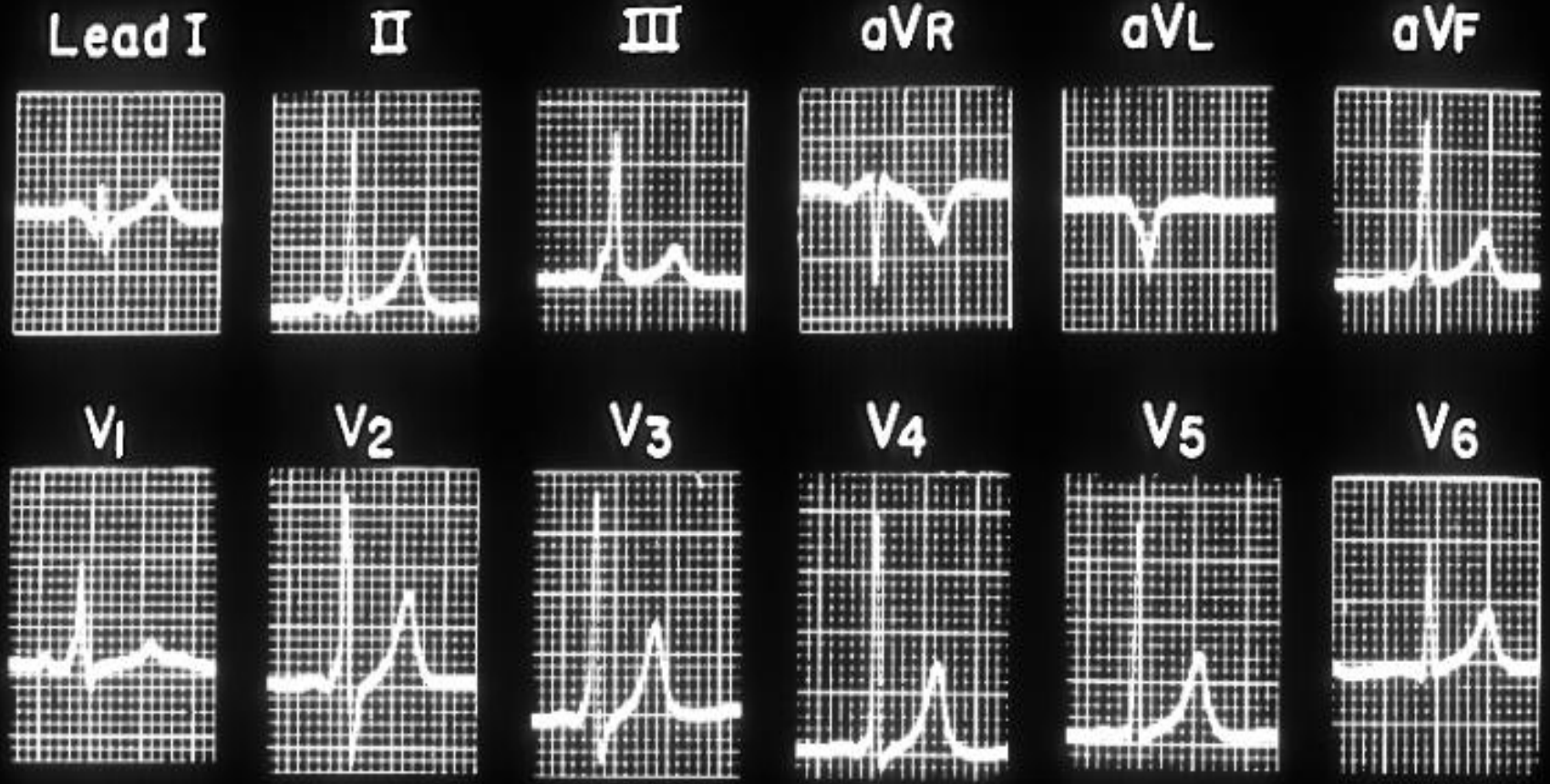

ECG
Femoral Vein
Pressure
50
25
0

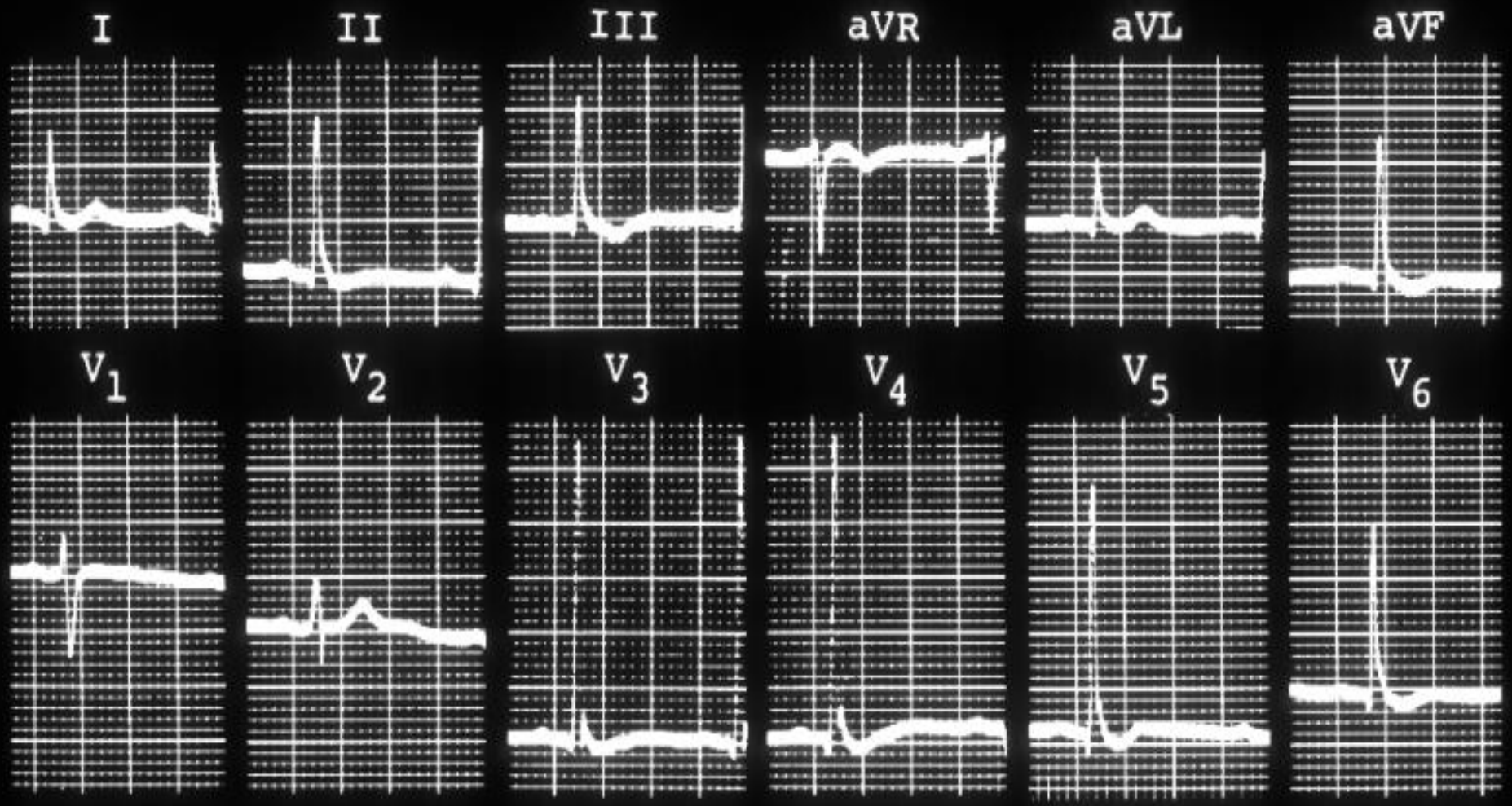

I
II
III
V1
V3
V5
5102

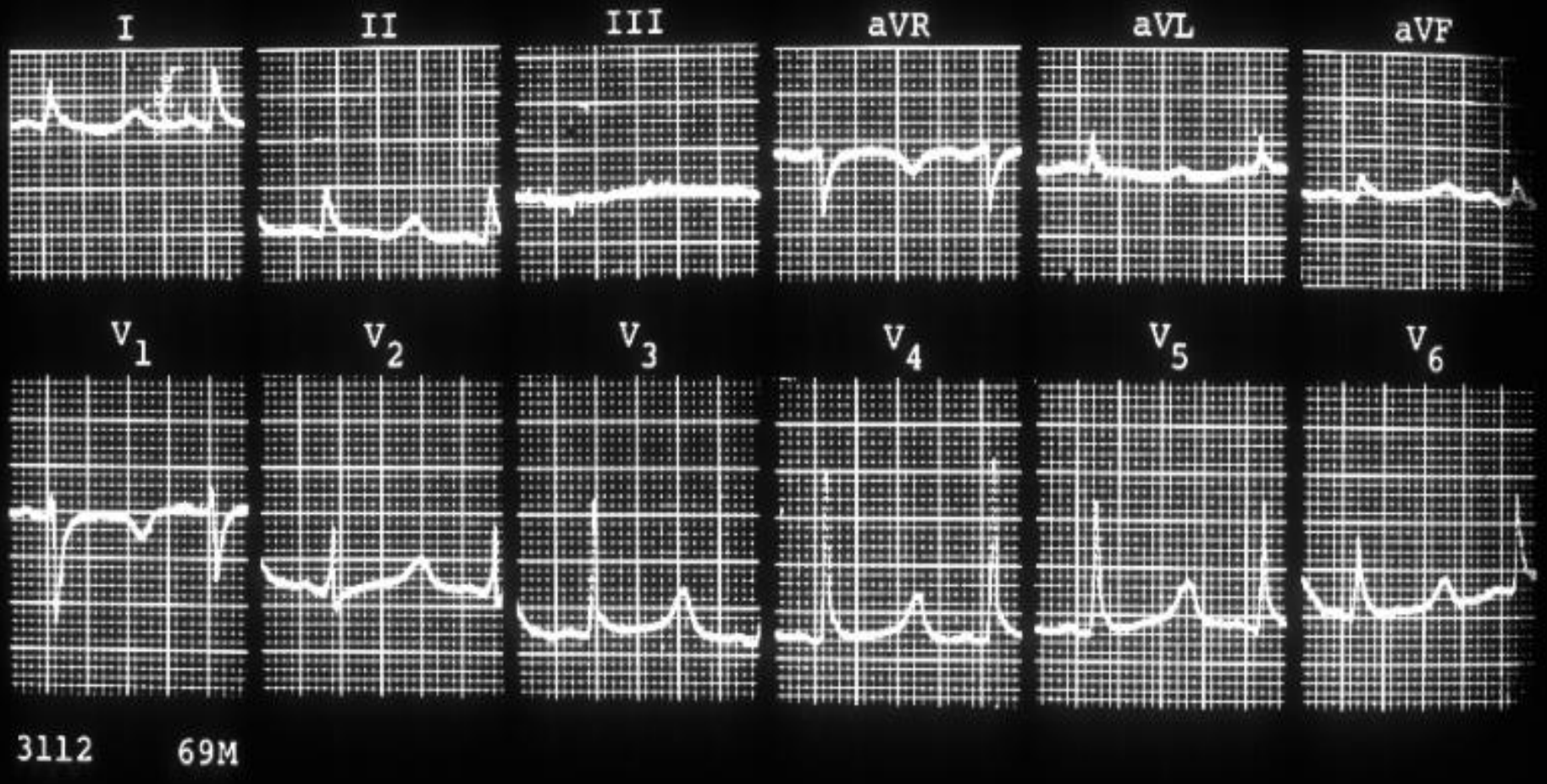

ECG
200
100
0
LV
FA
PRESSURE (mm Hg)
IHSS


Lead I
Lead II
Lead III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
.483
29F


Lead I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
.483
29F


I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6
3112
69M