2017
ACOI-Internal Medicine Board Review
Valvular and Congenital Heart Disease

Asif Serajian, DO, FACC

No disclosures relevant to this talk

Endocarditis Prophylaxis

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

* prosthetic valve
* previous endocarditis
* Congenital dz = repaired with residual, 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 !!

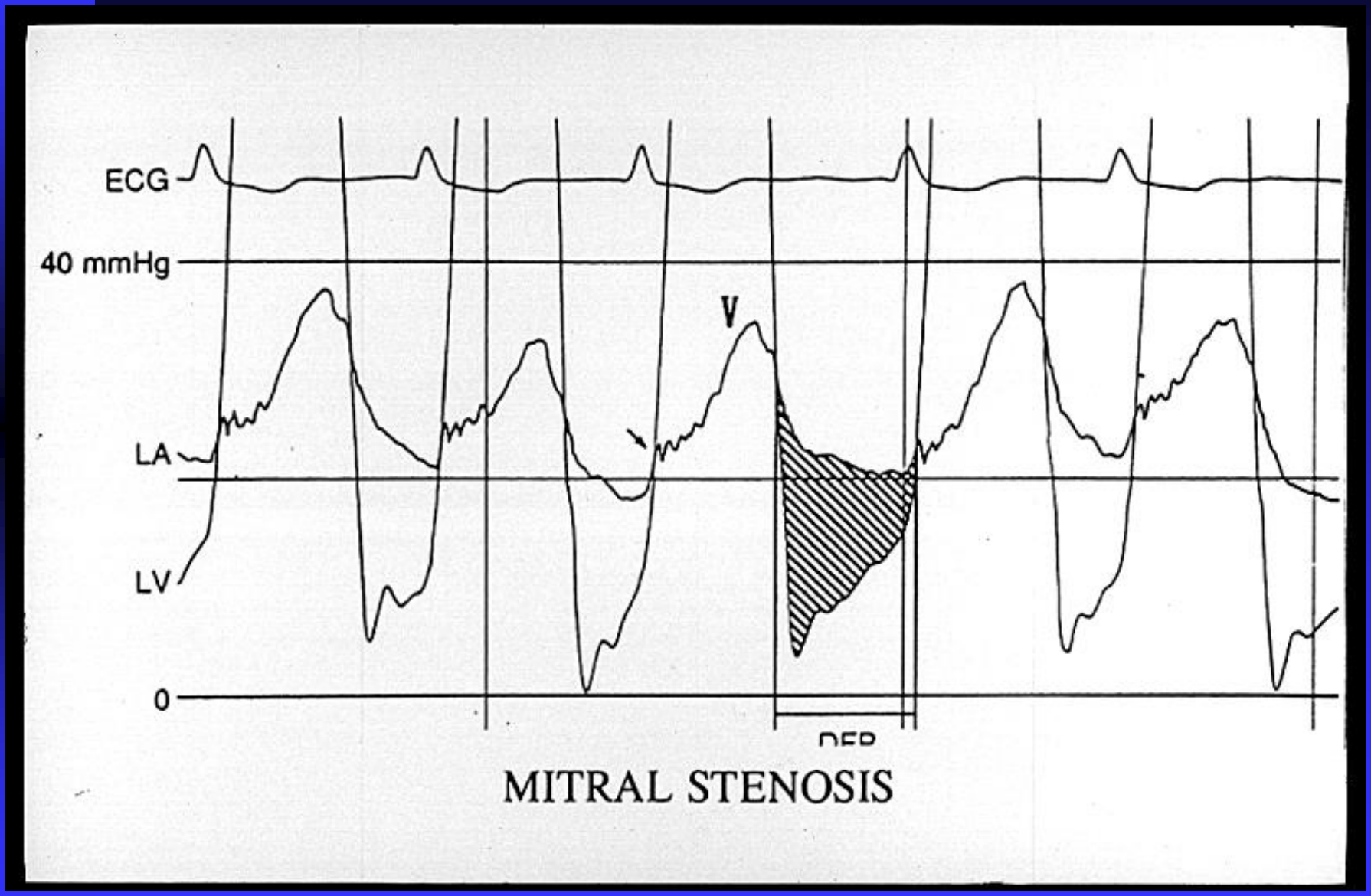
Mitral Stenosis (MS):

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

- Mimics MS = LA tumor, thrombus, cor triatriatum
- Lutembachers Syndrome = ASD and Rheumatic MS
- MAC = elderly

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 = 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(increase preload), amyl nitrite, exercise
  * □ Intensity = Valsalva (increases afterload then decreases preload)

MS - Complications:

- Death = CHF, systemic embolism, PE
- Systemic Embolism = CVA, etc.
  - 80% AFib
  - < severe MS
  - Tx = anticoagulate (warfarin, not NOAC'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)
  * MV area $\leq$ 1.5 and :
    Symptoms

or ... PASP > 50 mmHg (rest) or > 60 mm Hg (exercise)
or …Recurrent systemic embolism, new AFib (?)
or …very severe MS and valve favorable for balloon

Chronic Mitral Regurgitation (MR):

- Etiology = primary vs secondary
  - 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

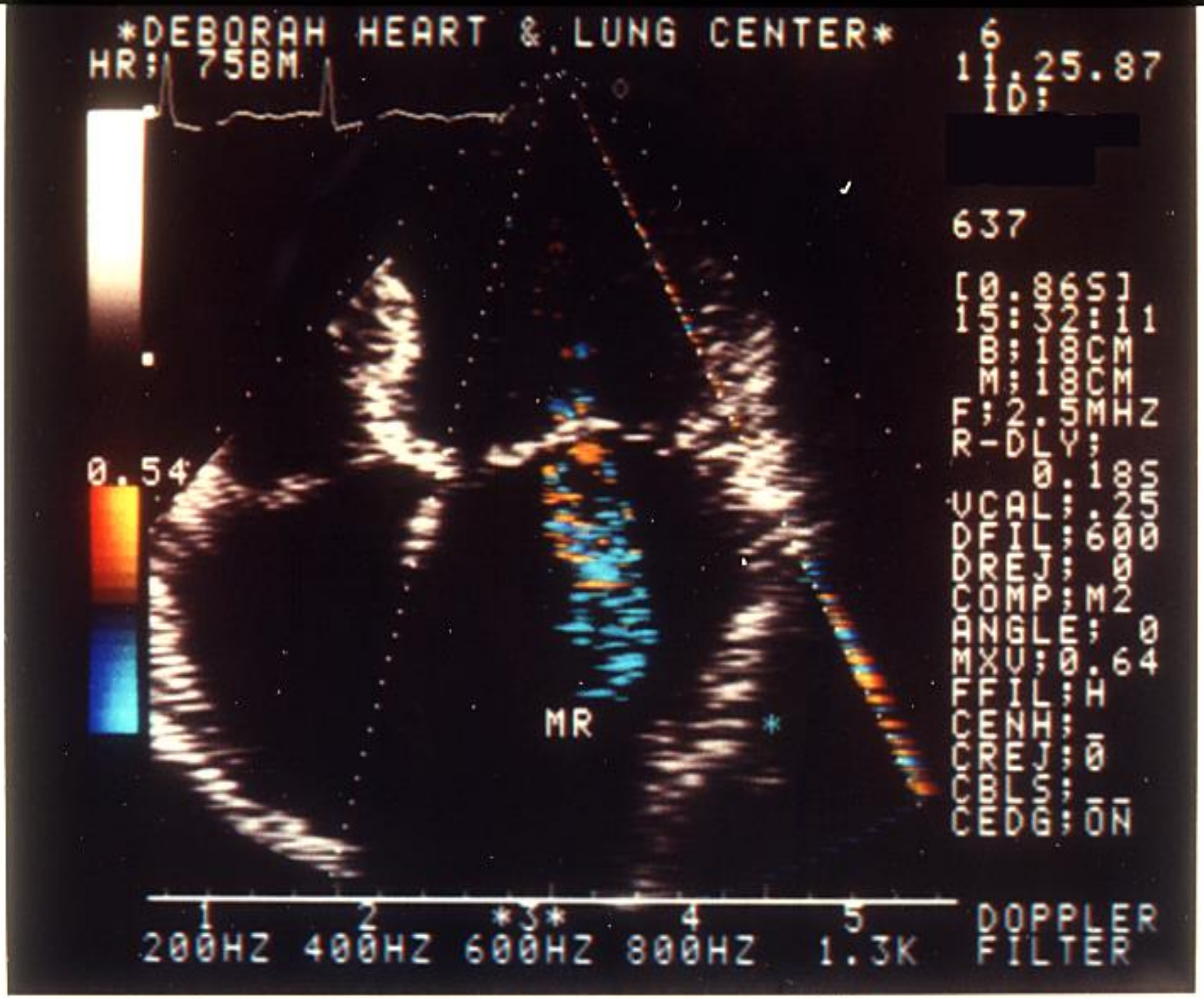
MR - Clinical:

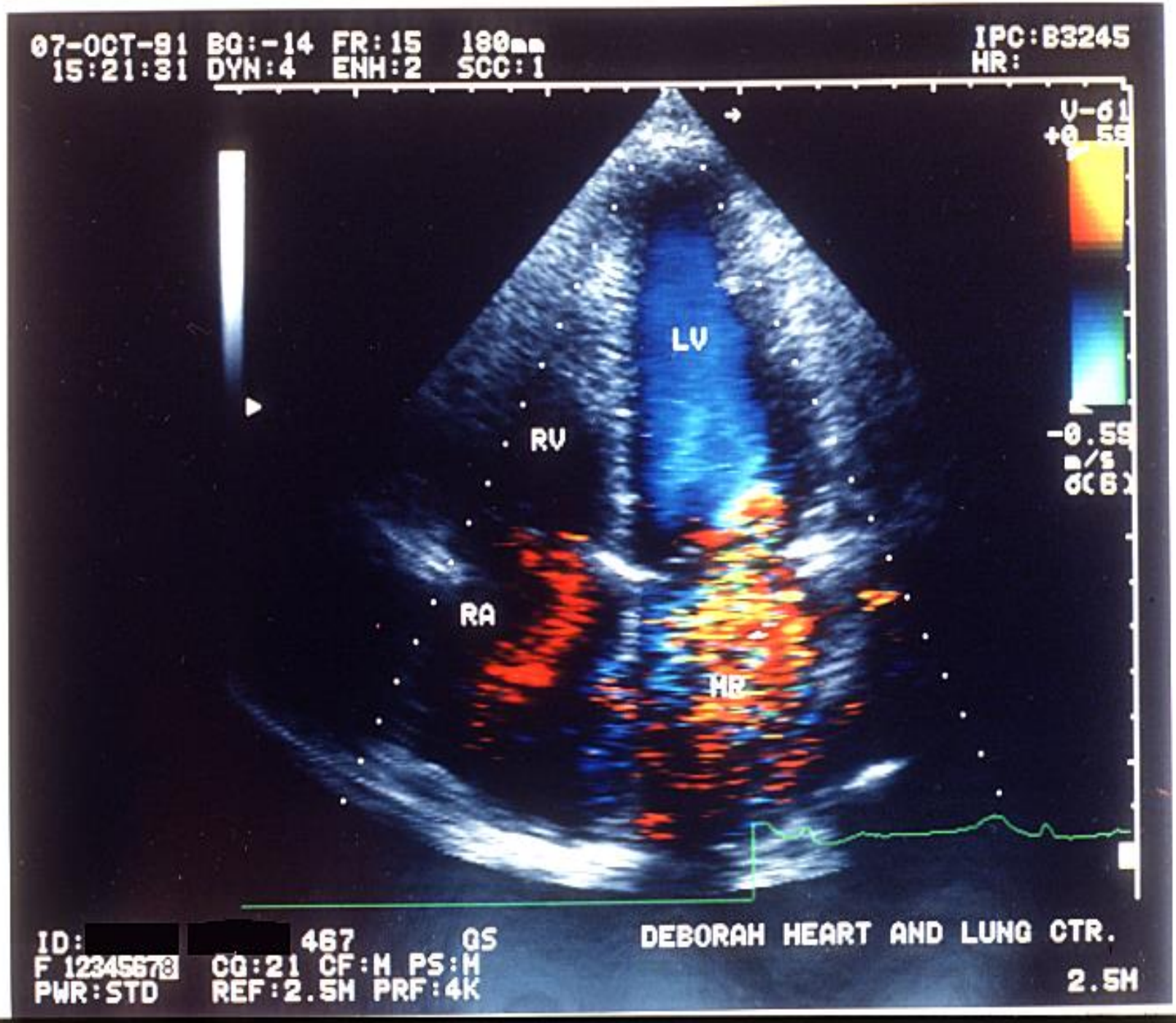
- Auscultation:
  - Murmur = holosystolic
    - apex to axilla (but not always)
    - intensity may not reflect severity
    - □ intensity = squatting (also increases afterload), isometrics (inc afterload)
    - □ intensity = Valsalva, amyl nitrite
    - Acute MR = atypical

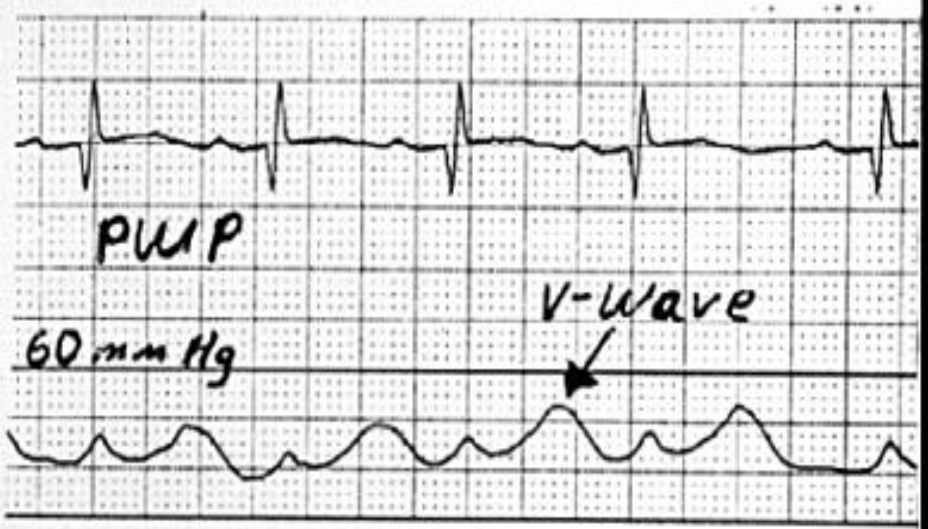
MR - Non Invasive Testing:

- 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
[0.865]
15:32:11
8;18CM
M;18CM
F;2.5MHZ
R-DLY;
0.185
VCAL;;250
VDFIL;;600
DREJ;;0
COMP;M2
ANGLE;;0
MXVL;0.64
FFILH;;
CENH;;
CREJ;;0
CBLS;;
CEDG;;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
+0.59
-0.59
m/s
dC6
LV
RV
RA
MR
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.
  - Severe MR without sx. but… (60 50 40 rule-EF PAP ESD)
    - EF ≤ 60%

    or
    - End - Syst. dimension ≥ 40 mm

    or
    - Pulm. HTN (> 50 mmHg rest or > 60 mm Hg w/ex.)

    or
    - New onset AFib

    or * High likelihood of repair
  - EF< 30% = ? candidate for surgery

- Mitral valve replacement is class III if < 50% posterior leaflet involved
- Recommend repair

Echocardiogram findings
Mild: VC < 0.3 volume < 30 mL RF < 30% ERO < 0.2 cm2
Severe: VC > 0.7 mm volume > 60 mL RF < 30 ERO >0.4

VC=vena contracts
RF=regurgitant fraction
ERO=effective regurgitant orifice

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

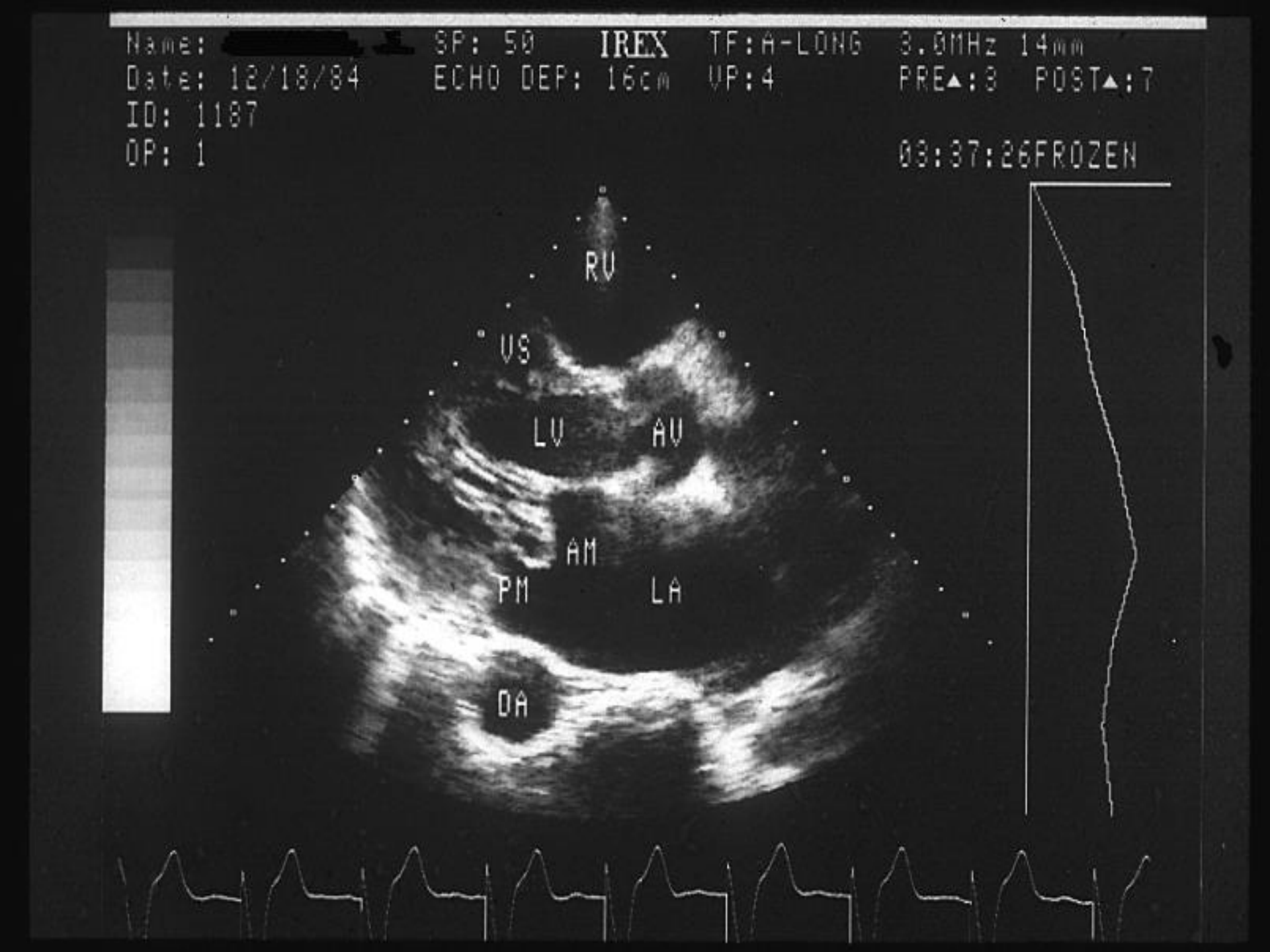
MVP - Non Invasive Testing:

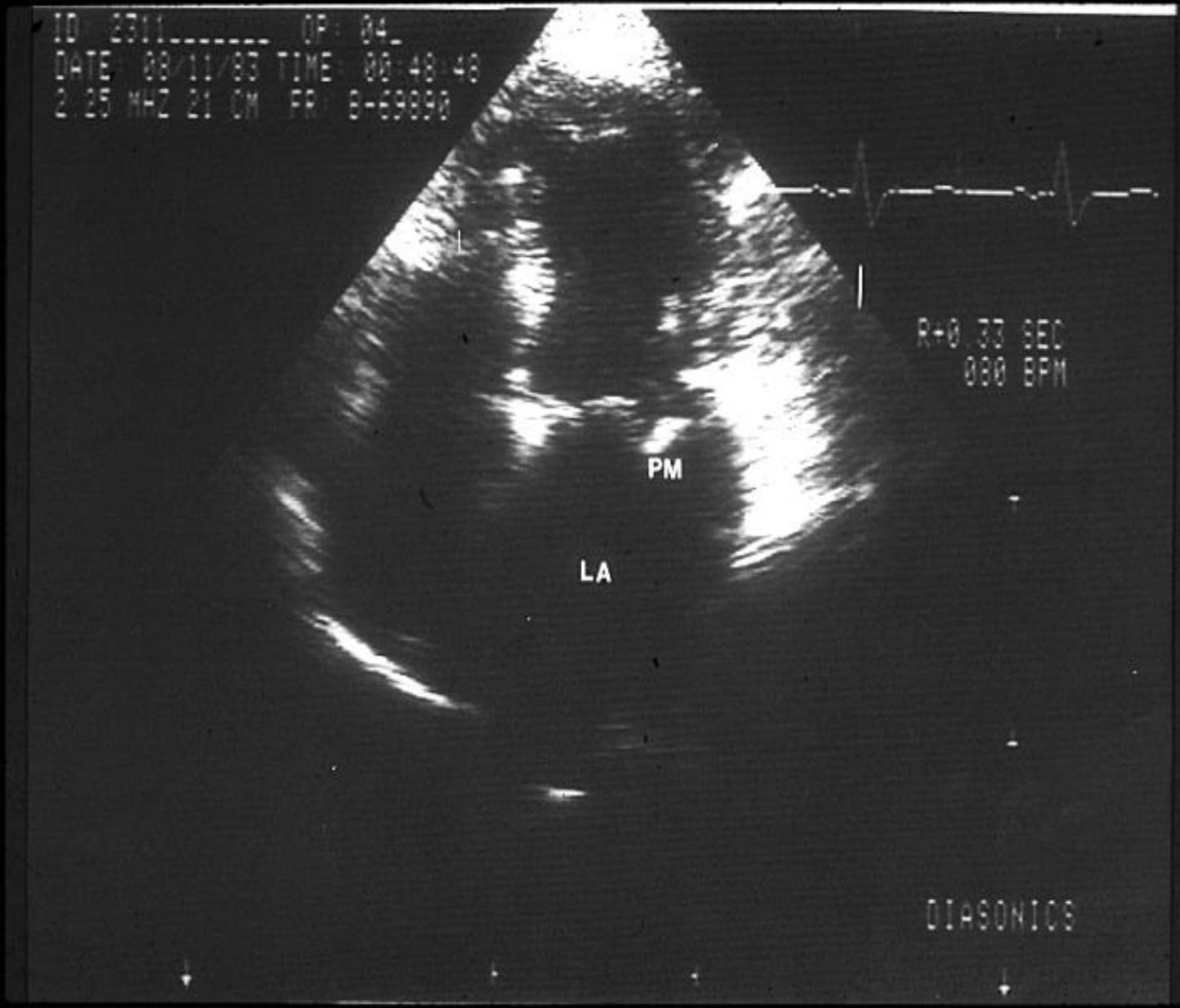
- EKG = usually normal
  - PSVT
  - ↑ incidence WPW
- CXR = unhelpful

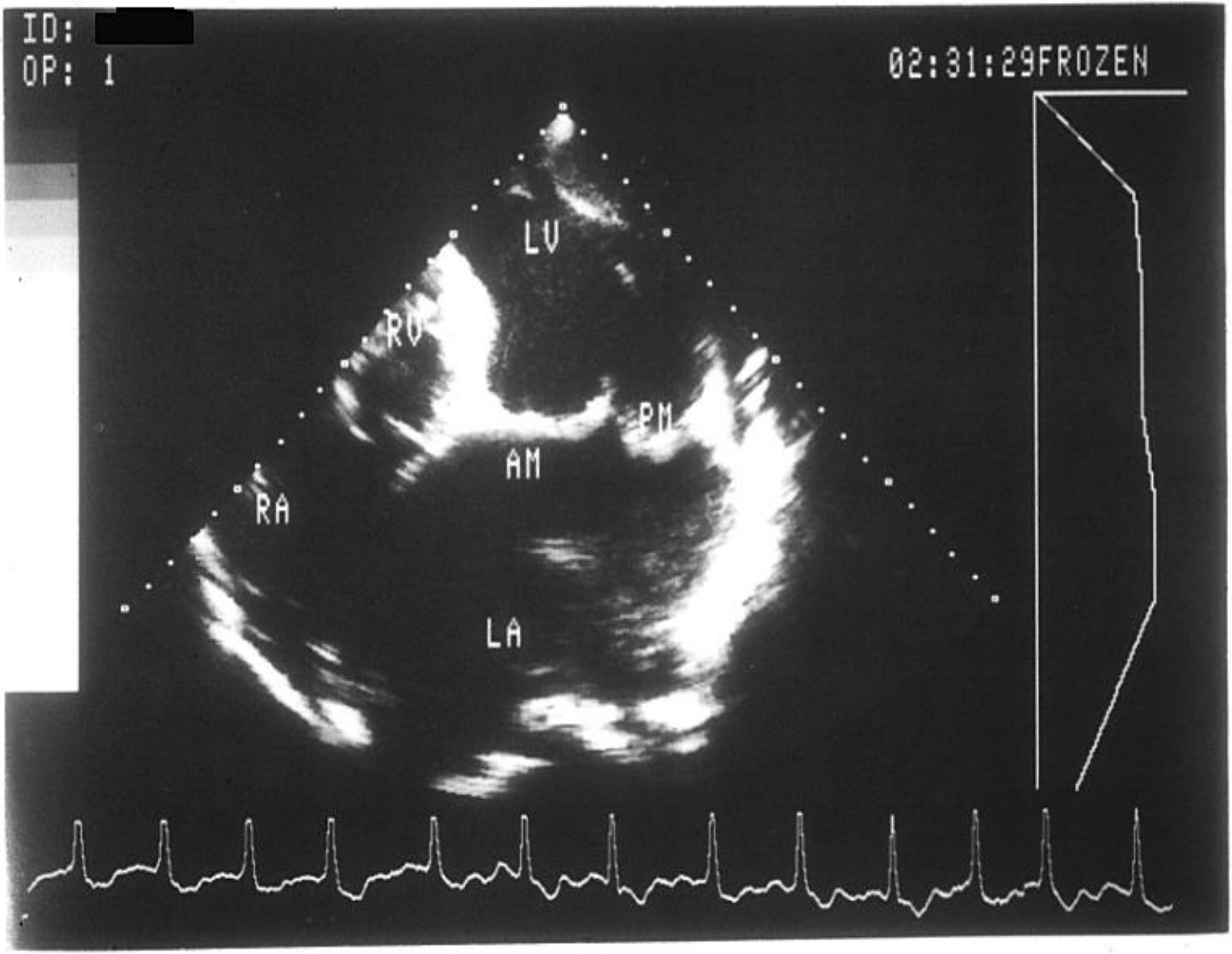
MVP - Non Invasive Testing:

- 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:

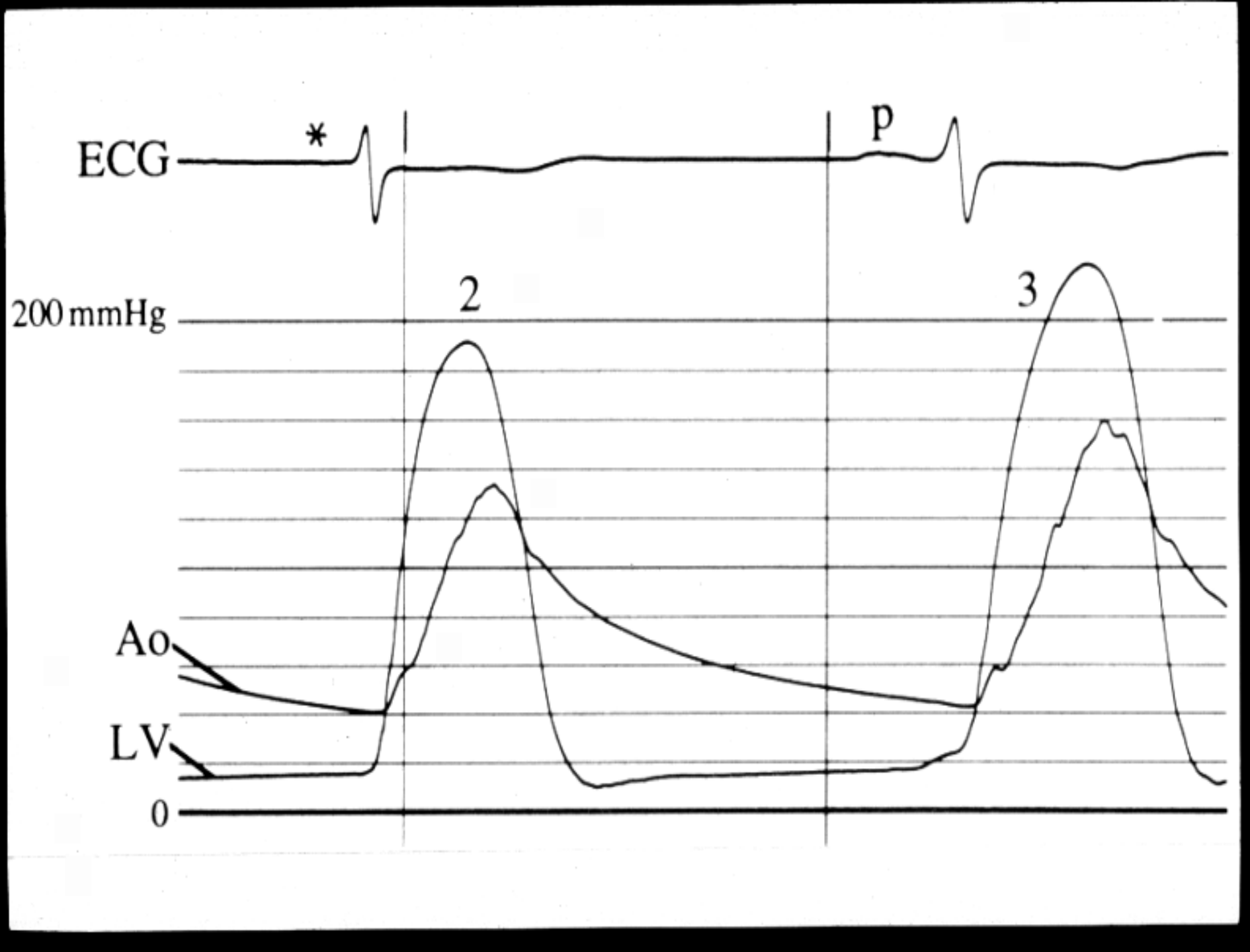
- MR = as previously reviewed
  * May develop acute severe MR due to chordal rupture !!

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:
    Valve area > 1.5 cm^2 = mild AS
    Valve area 1 - 1.5 cm^2 = moderate AS
    * AV Gradient 25 – 40 mm Hg (mean)
    Valve area < 1.0 = severe AS
    - AV Gradient >/= 40 mm Hg (mean)
    - Peak flow vel. >/= 4.0 m/s
  - 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:
    * “parvus et tardus” (↓ amplitude with delayed upstroke)
    * pulsus alternans = ↓ CO
  - Apical impulse = sustained, left shift
  - Thrill = base, supra-sternal notch

AS - Auscultation:

- S_2 = paradoxical split, □ intensity (A_2)
- Systolic ejection click = young, congenital
- Systolic ejection murmur
  - base to carotids
  - base to apex = Gallavardin phenomenon
  - severe AS = longer, louder, peaks later

AS - Non Invasive Testing:

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

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

Aortic stenosis

Severe

Mg >40 max velocity >4 area <1

or indexed < 0.6cm2/m2

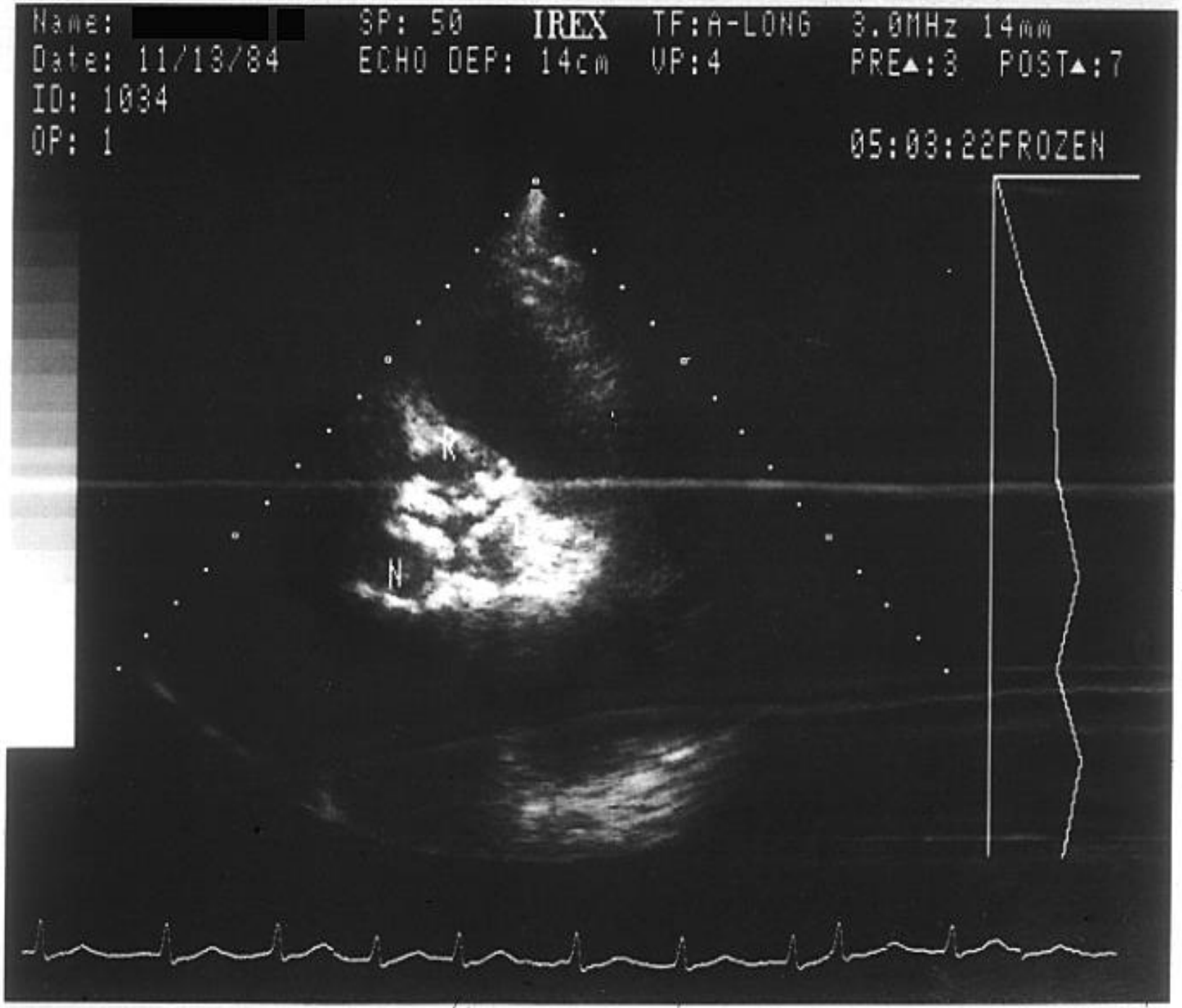
Very severe

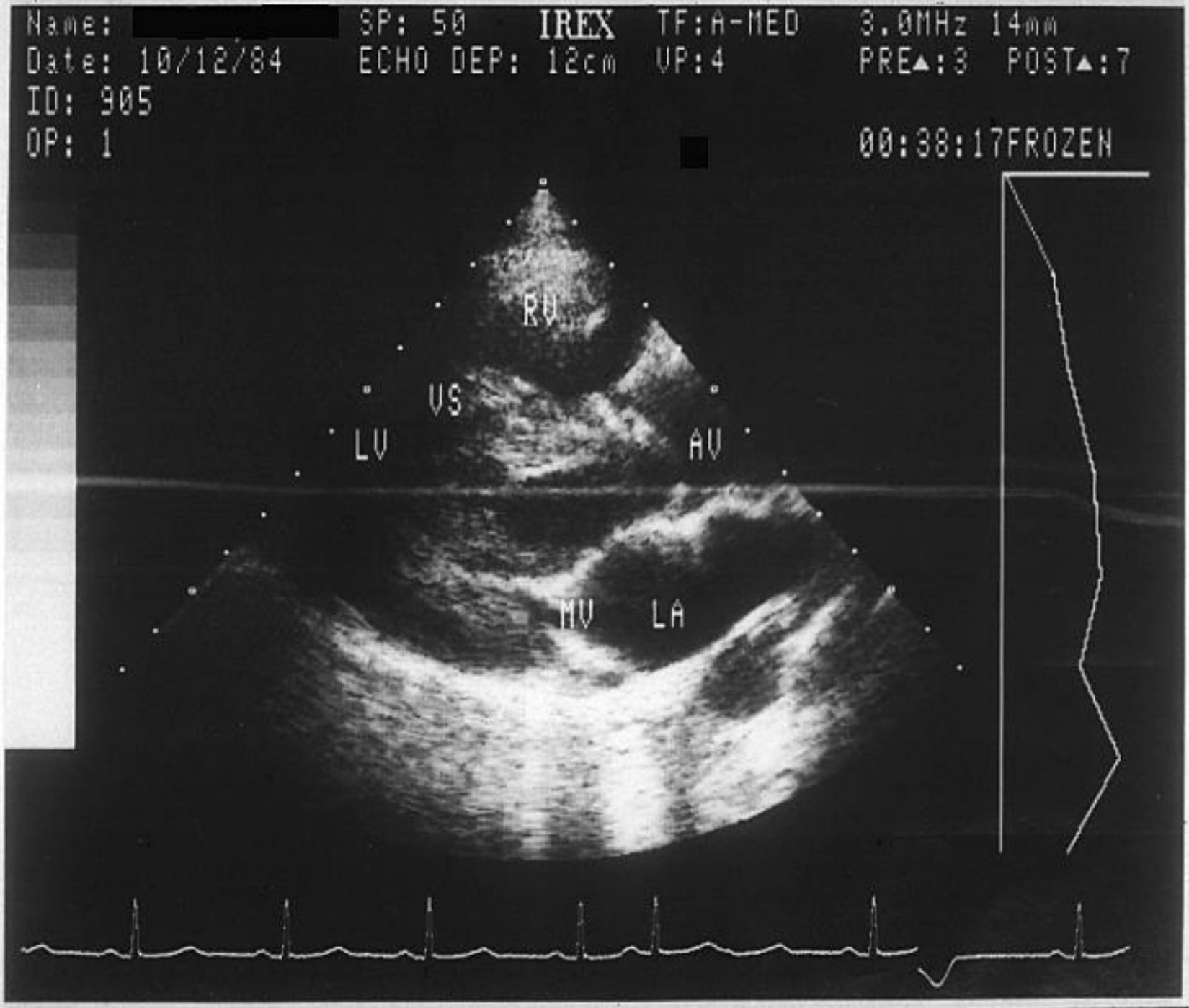
Mg >50 max velocity >5 area < 0.6

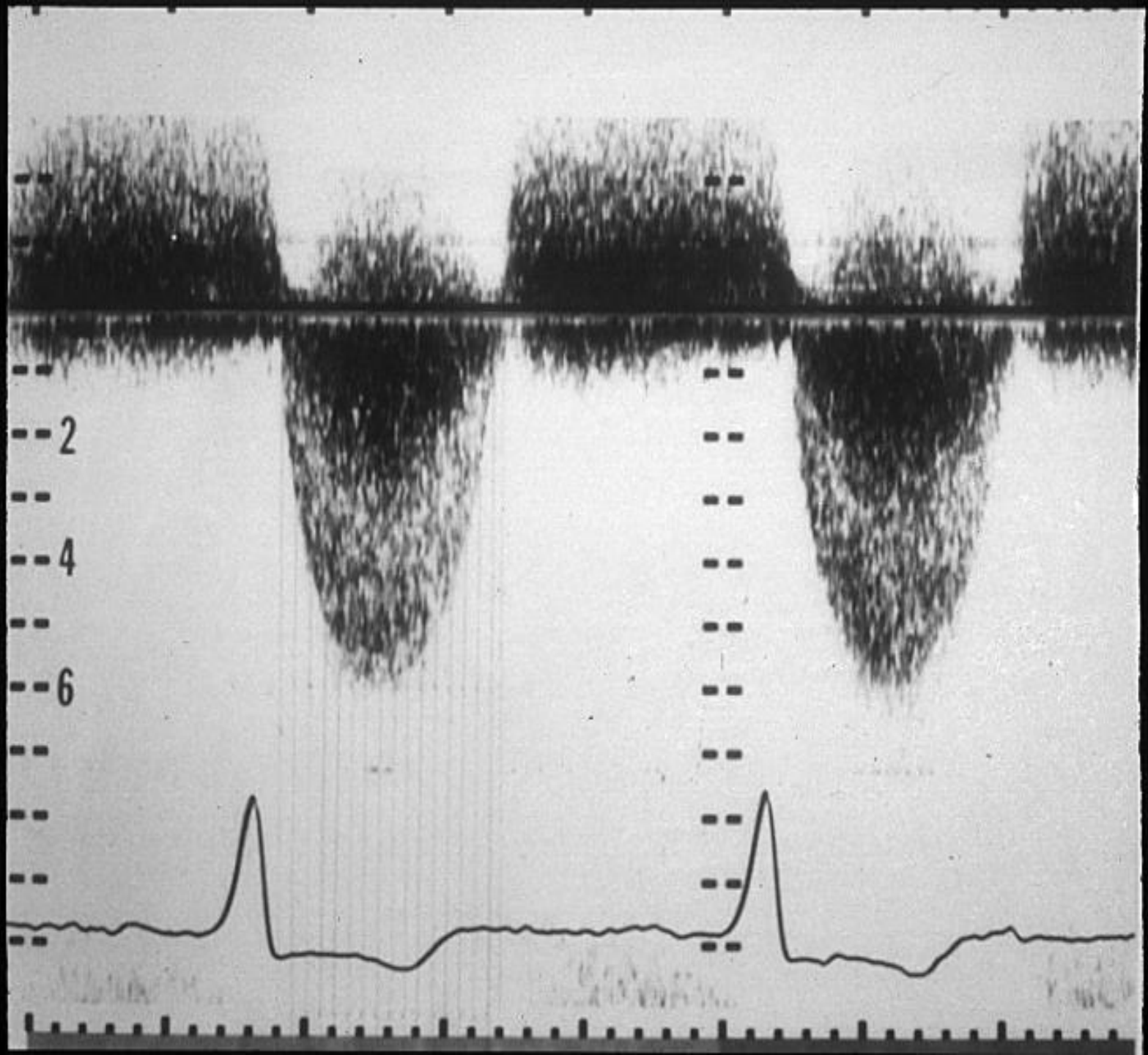
Surgery if severe + sx or ef < 50 or gxt with hypotension or sx

DSE, subjects with low-gradient AS who manifest an increase in peak

pseudo-AS, dobutamine caused a considerable increase in valve area (≥


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
R
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
  - Do statins reduce progression of degenerative AS = ??
  - The decision for surgery is based primarily on presence of symptoms

AS:

- "Surgical" therapy = valve replacement

  * 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
    or.. Abnormal ETT
    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 = > 0.3 m/s per year increase in flow vel.

* NB: **TAVR** (percutaneous) = effective tx, in high risk patients

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
- Dobutamine stress echo = may be helpful for patient with reduced LVEF

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:

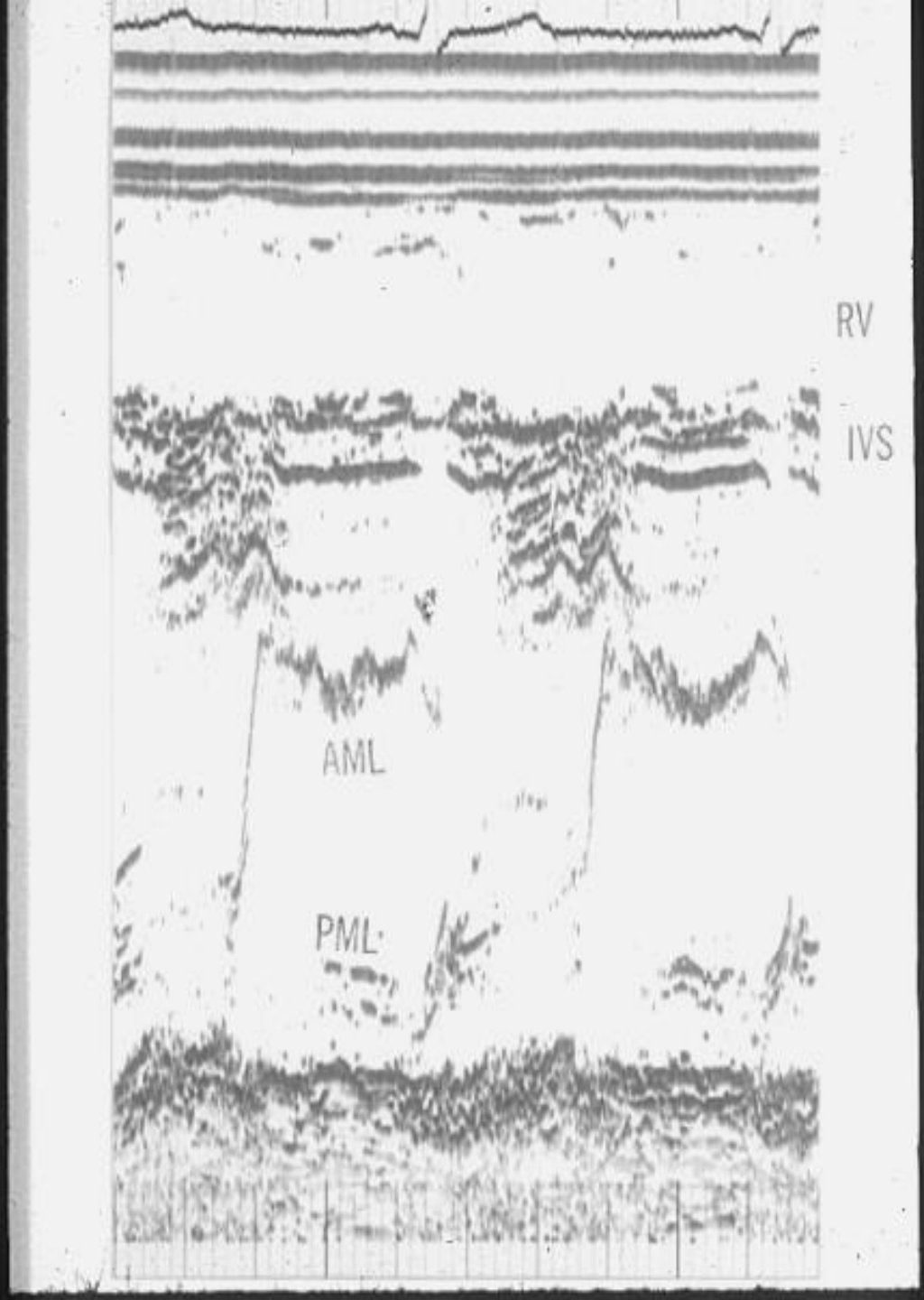
- Dynamic Auscultation

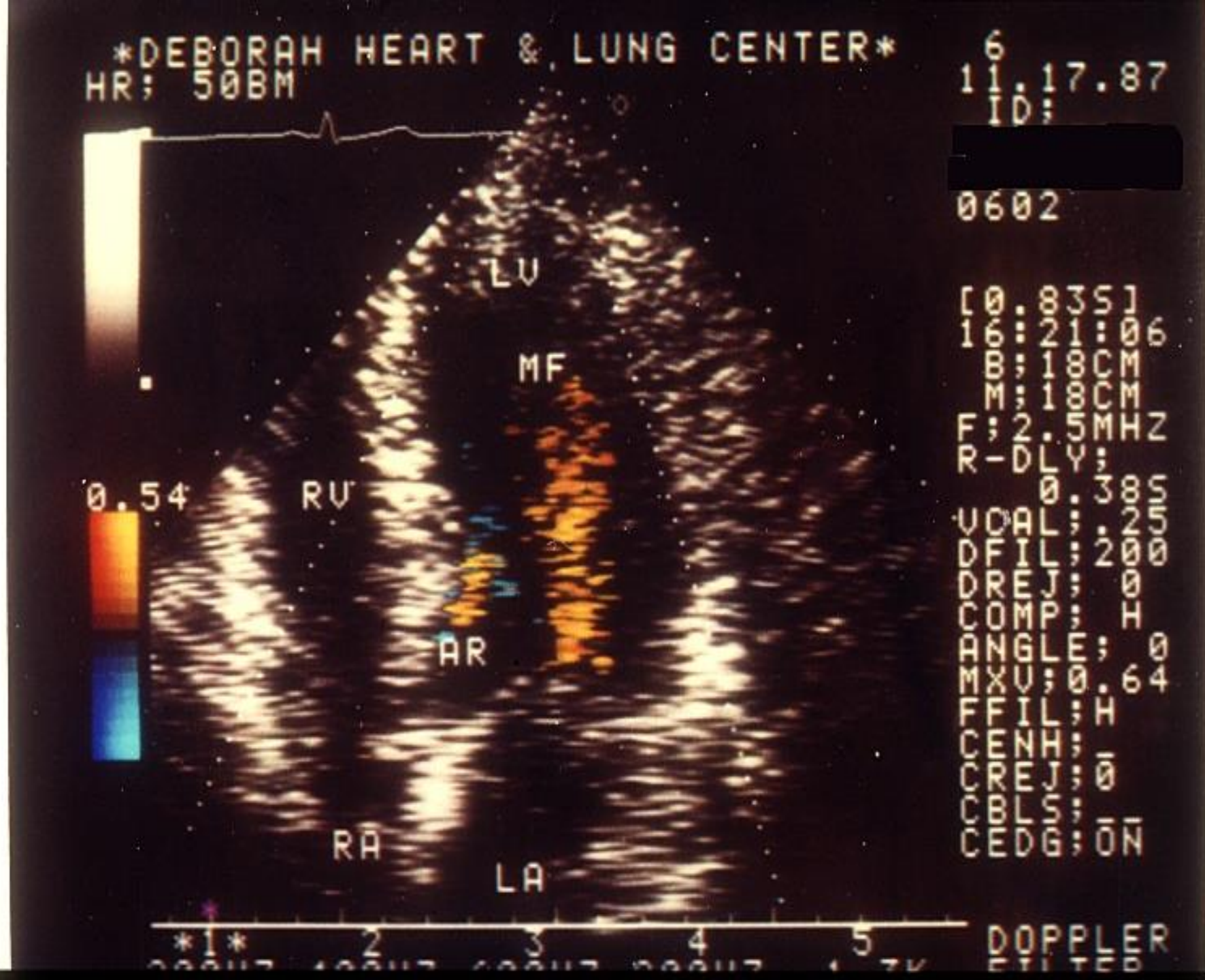
  * ↑ intensity = pressors, squatting, isometrics

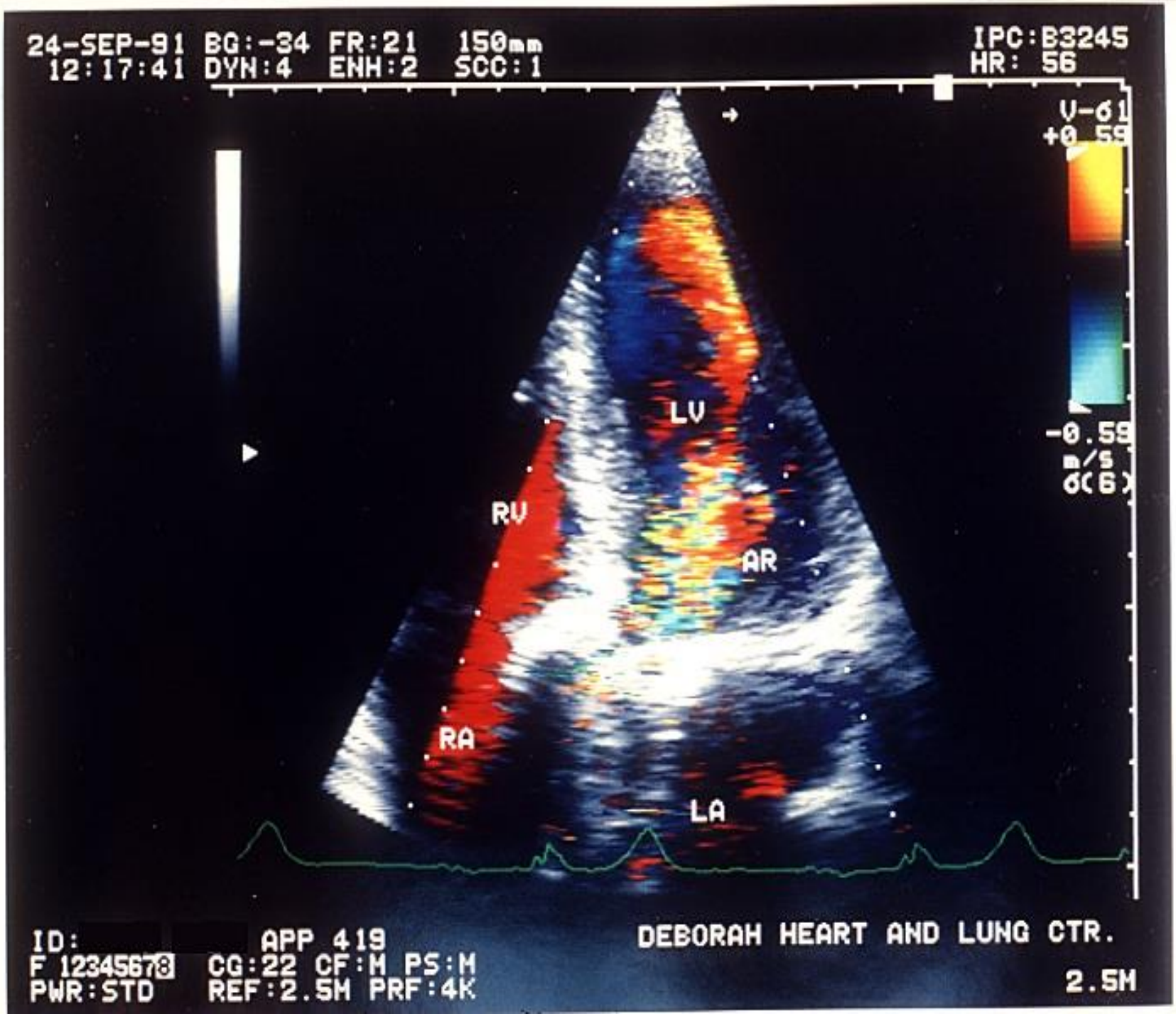
  * ↓ intensity = amyl nitrite, Valsalva

AR - Non Invasive Testing:

- EKG = LVH
- CXR = cardiomegaly, dilated aorta
- Echo = etiology, LV size and function
- Doppler (color flow) = semi-quantitate severity


RV
IVS
AML
PML


DEBORAH HEART & LUNG CENTER
HR; 50BM
6
11.17.87
ID;
0602
LV
MF
RV
0.54
AR
RA
LA
[0.835]
16::21::06
B;18CMM
M;18CMM
F;2.5MHZ
R-DLY;385
VCAL;;200
DFILJ;;0H
DREJP;;0
COMPE;;04
ANGLE;0.64
MXV;;H
FFIL;;
CENH;;0
CREJ;;
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
  - severe AR without symptoms with normal systolic function ?? (Class IIb rec.)
- Surgical = AV replacement
  - Severe AR with symptoms
  - Severe AR without symptoms:

* EF < 50%

or .. * ESD > 50 mm (echo measurement)

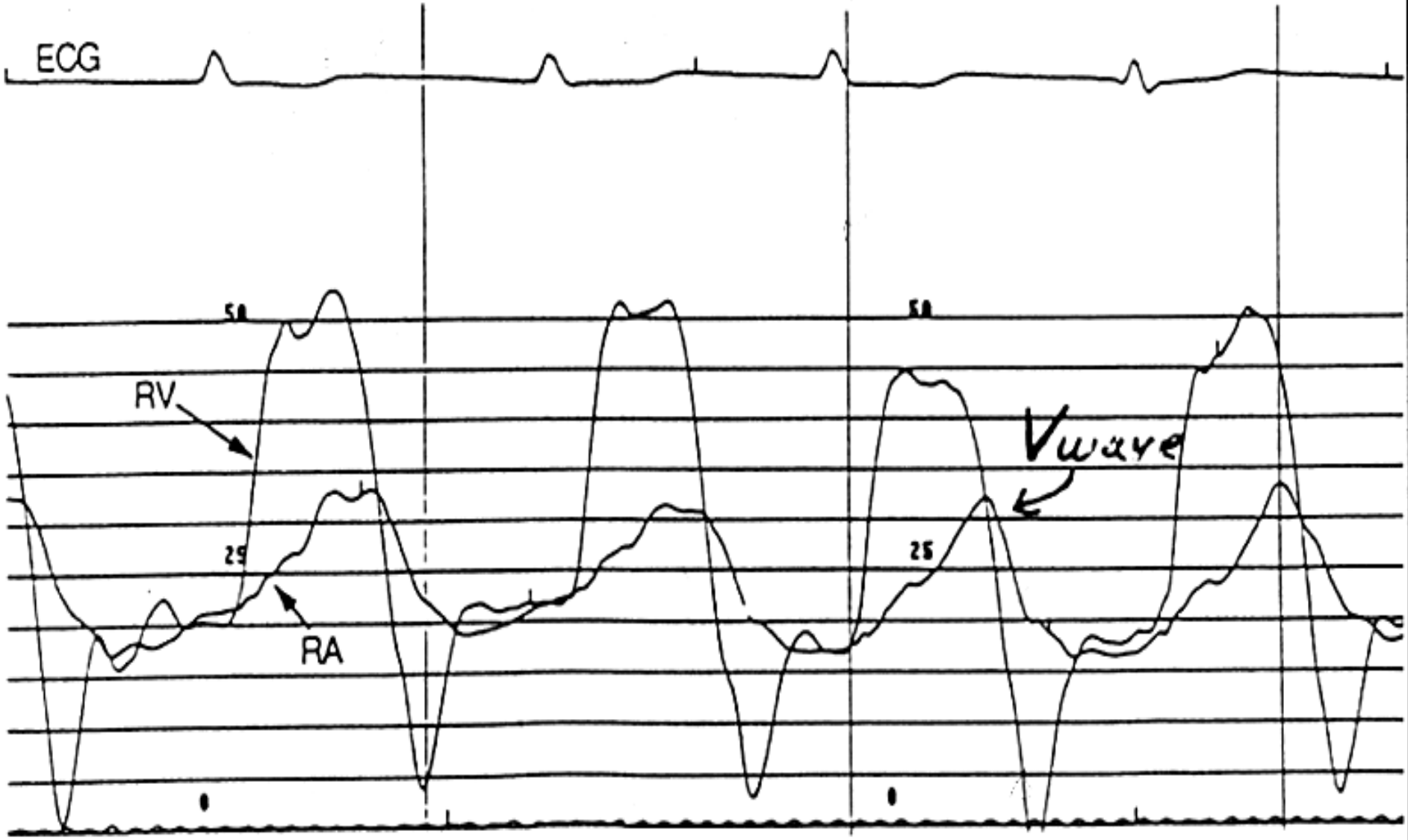
(50-50 pneumonic) (MR was 60 50 40)

Tricuspid Stenosis:

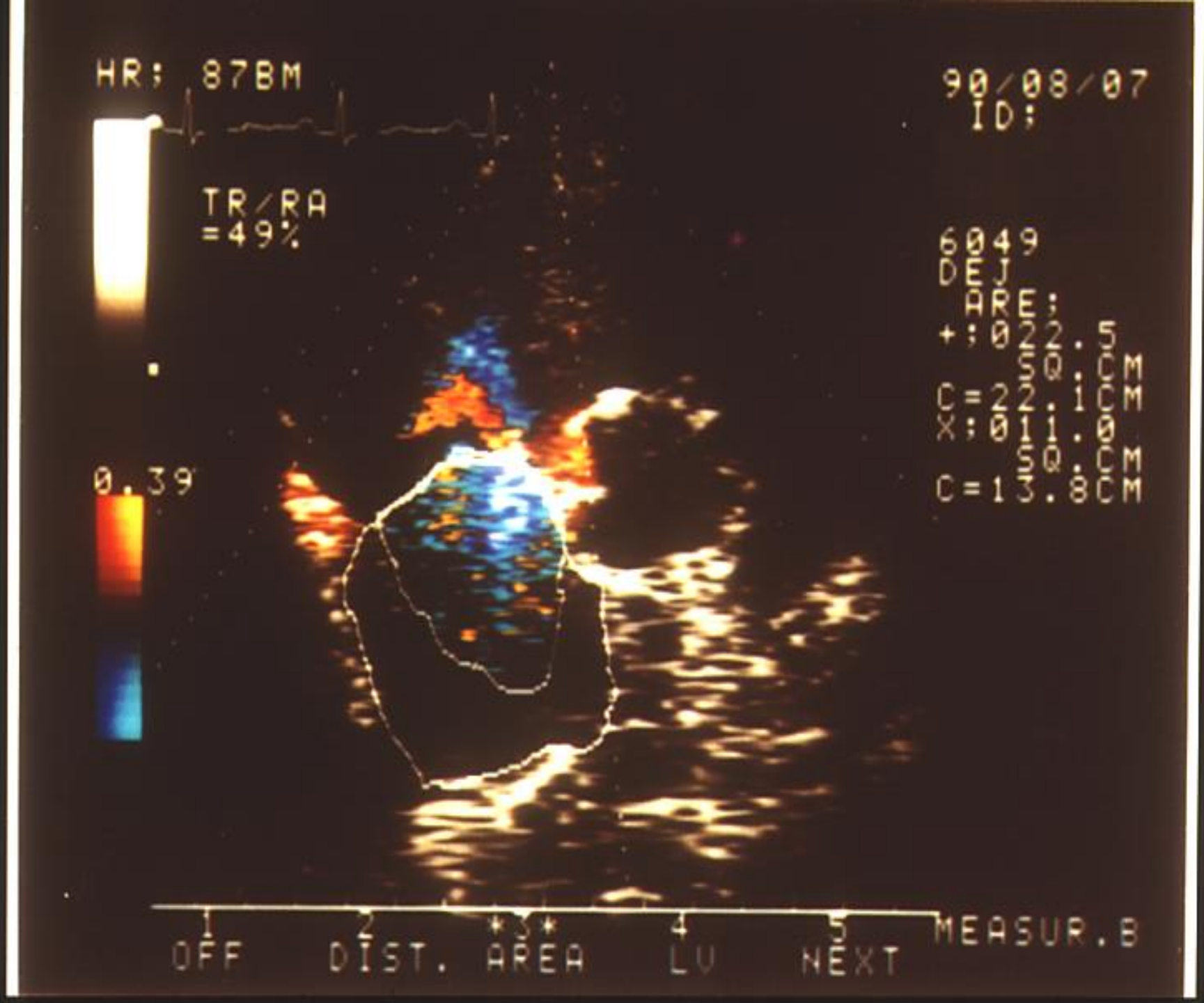
- Etiology = rheumatic, congenital, carcinoid

Tricuspid Regurgitation:

- Etiology = RV dysfxn/dilatation, TVP, Ebstein's, Rheumatic fever, XRT, carcinoid
- 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=13.8
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 BAV)

VSD = 30%
ASD = 10%
PDA = 10%
PS = 7%

Coarctation Aorta = 7%
AS = 7%
Tetralogy of Fallot = 6%
Transposition = 4%

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. = coarct., bicuspid AV

Bicuspid Aortic Valve (BAV)

- 1-2% population
- AS &/or AR
- Screen 1st degree relatives = esp. if hx aortopathy
- Associated with aortopathy = aneurysm, coarct, dissection
- Can have severe ascending aorta dilatation without signif. valve dysfunction

  * 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 asc. 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:
  * residual HTN
  * accelerated CAD
  * CHF
  * dissection
  * CVA, intracerebral hemorrhage

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

Coarctation of Aorta


1.5T MR2MR2
Ex: 8747
Post FT 3D MRA
C: Y
Se: 18/2
Im: 39/128
Sag: L29.7
S
Maine Medical Center
GENDRON DEREK
1980 Sep 16 M 04394626
Acc: 10520936
2008 Oct 27
Acq Tm: 13:15:19
288 x 128
A
P
ET: 1
TR: 6.4
TE: 1.9
8CARDIAC
4.2thk/-2.1sp
Lin:DCM / Lin:DCM / Id:ID
W:3196 L:1598
I
DFOV: 34.0 x 34.0cm

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
  *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(secondum), 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 (with or without sx) = class I
* Hx of paradoxical embolus = class IIa
* Orthodeoxia – platypnea = class IIa

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, PAH

  * Net left to right shunt

  * Hx of endarteritis

Ventricular Septal Defect (VSD):

- Most common defect at birth
- Seldom seen in adults unless small
- Holosystolic murmur LLSB
- Spontaneous closure frequent = if small
- L □ 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:

  * Pulm / Systemic flow ratio >/= 2.0 and evidence of LV vol. overload = class I

  *Hx endocarditis = class I

  * Pulm / systemic flow > 1.5 (in absence of severe pulm HTN) = class IIa

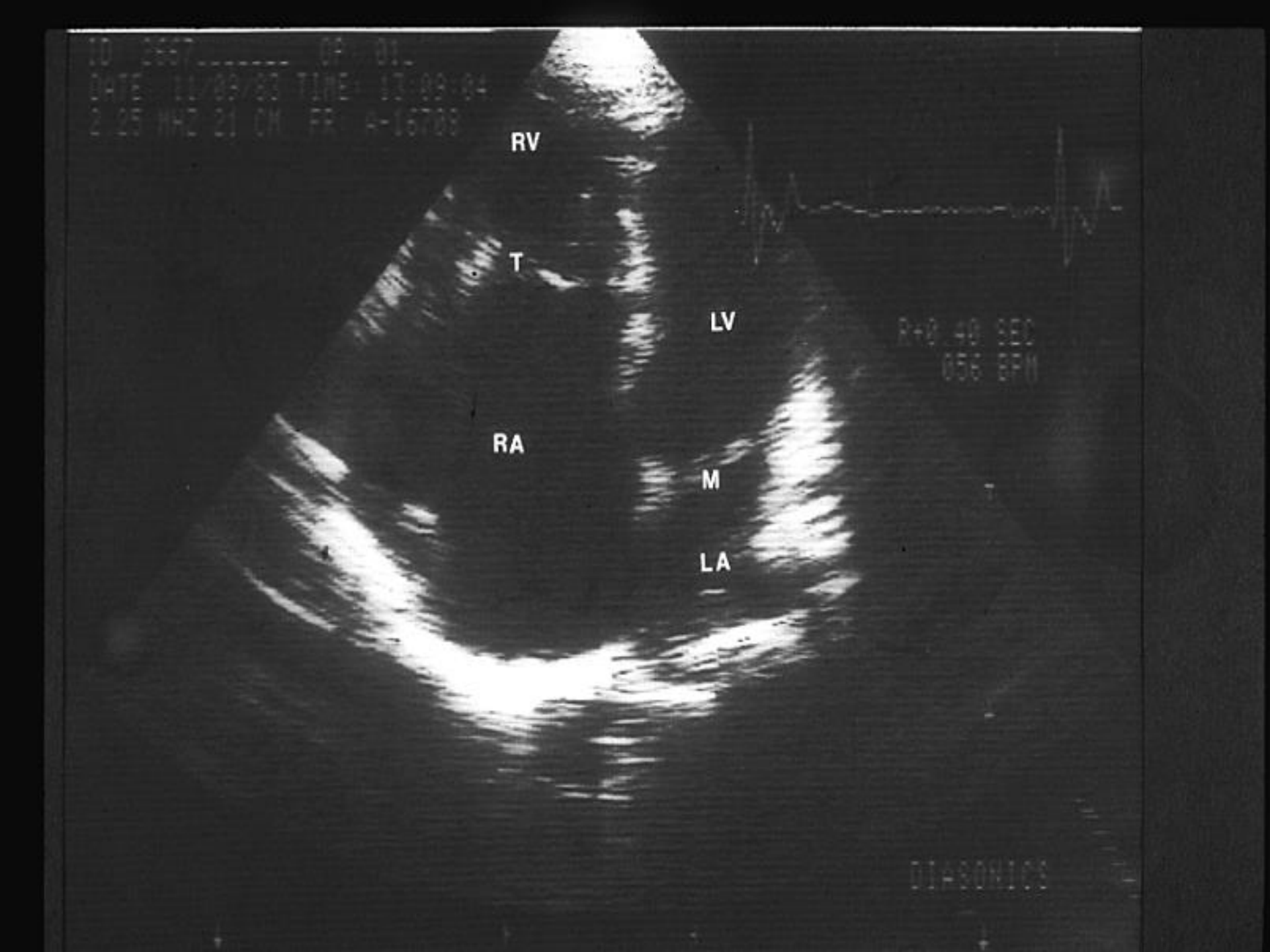
Tetralogy of Fallot (TOF):

- 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
- 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:

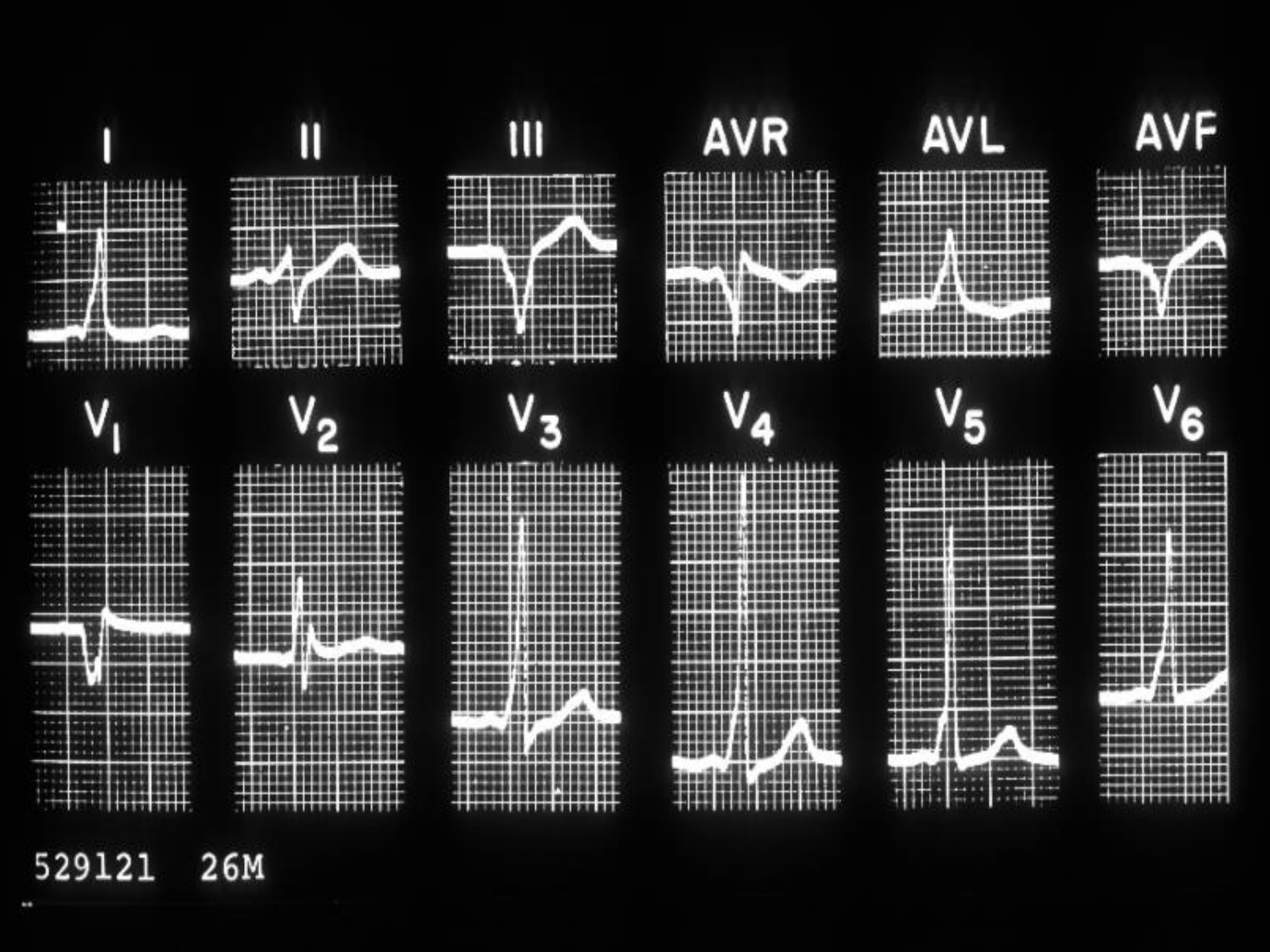
- L – Transposition (congenitally corrected):
  * AV discordance and ventriculo-arterial discordance
  * Morphologic RV = systemic ventricle
  * Morphologic LV = venous ventricle
  * Function = blood follows normal course
  * Survival into adulthood
  *Problems = systemic A-V valve regurg and systemic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

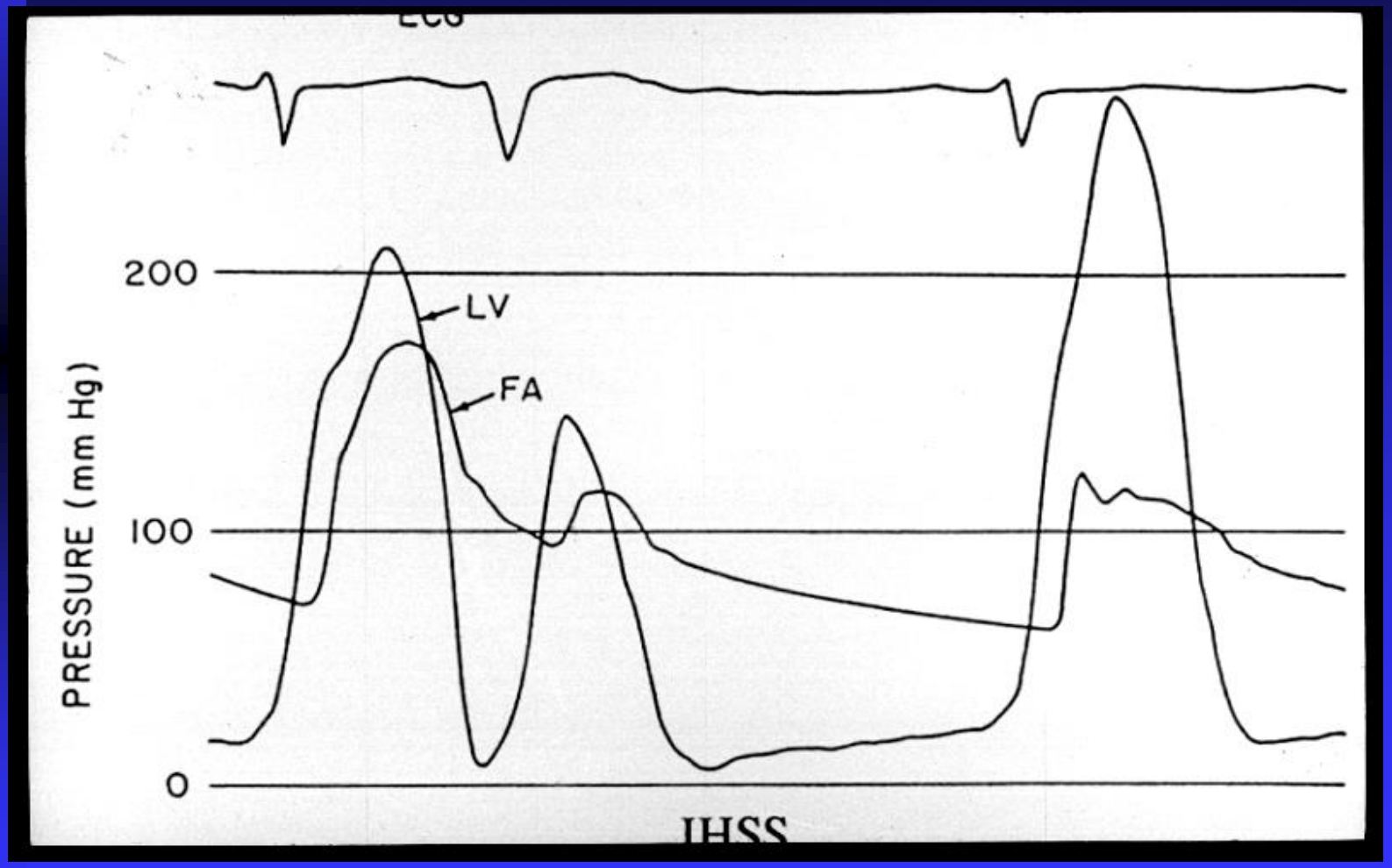
Congenital Disease - Summary:

- L □ R shunt = non-cyanotic
  * ASD, VSD, PDA, Persistent truncus
- R □ L shunt = cyanotic
  * TOF (± 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
200
100
0
LV
FA
PRESSURE (mm Hg)
IHSS