2018
ACOI-Internal Medicine Board Review
Valvular and Congenital Heart Disease

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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 (1st – 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):

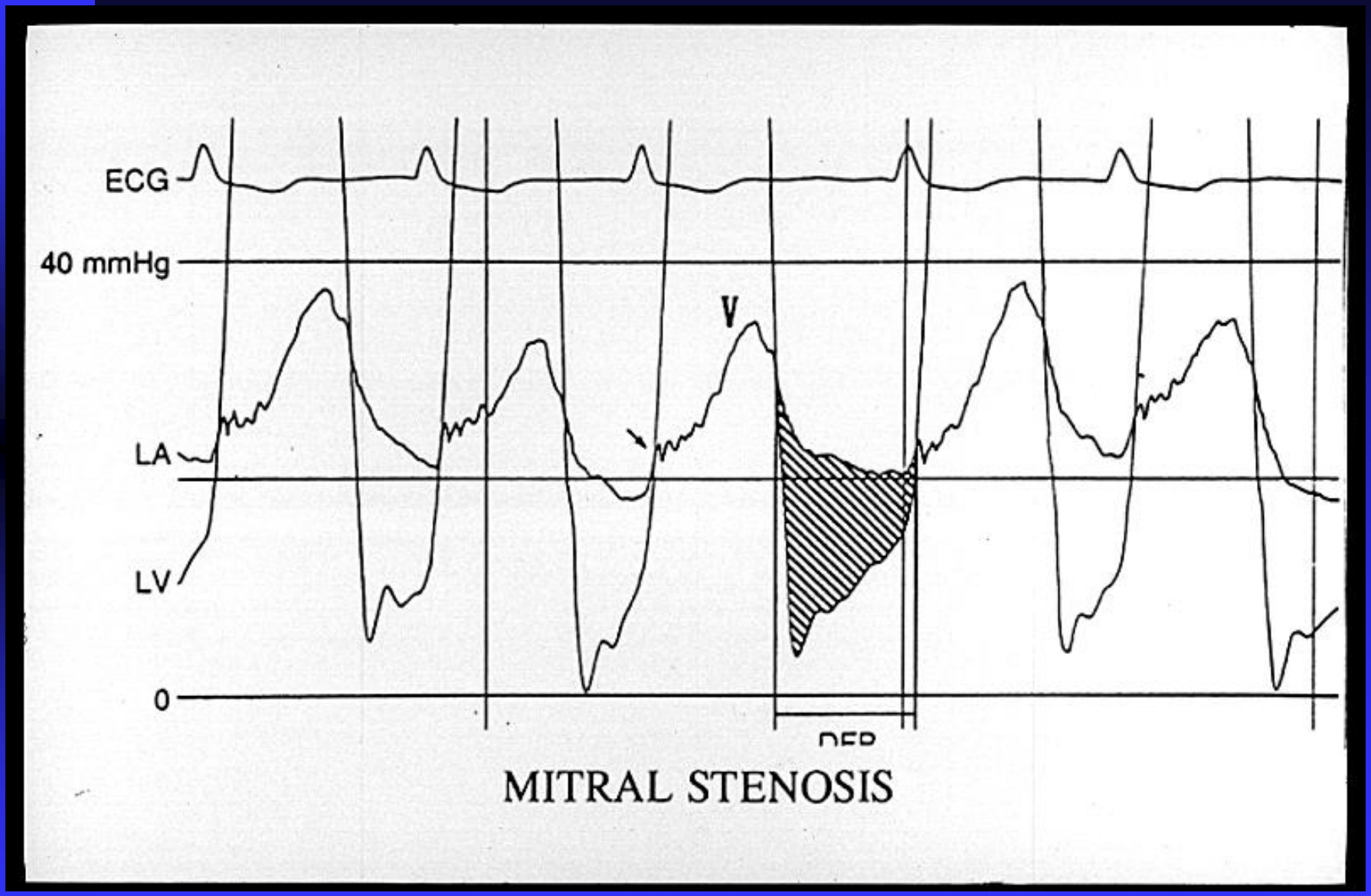
Etiology: Rheumatic Fever (20-40 yr. latency)

- 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, amyl nitrite, exercise
  - * ↓ Intensity = Valsalva

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 cm2 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:

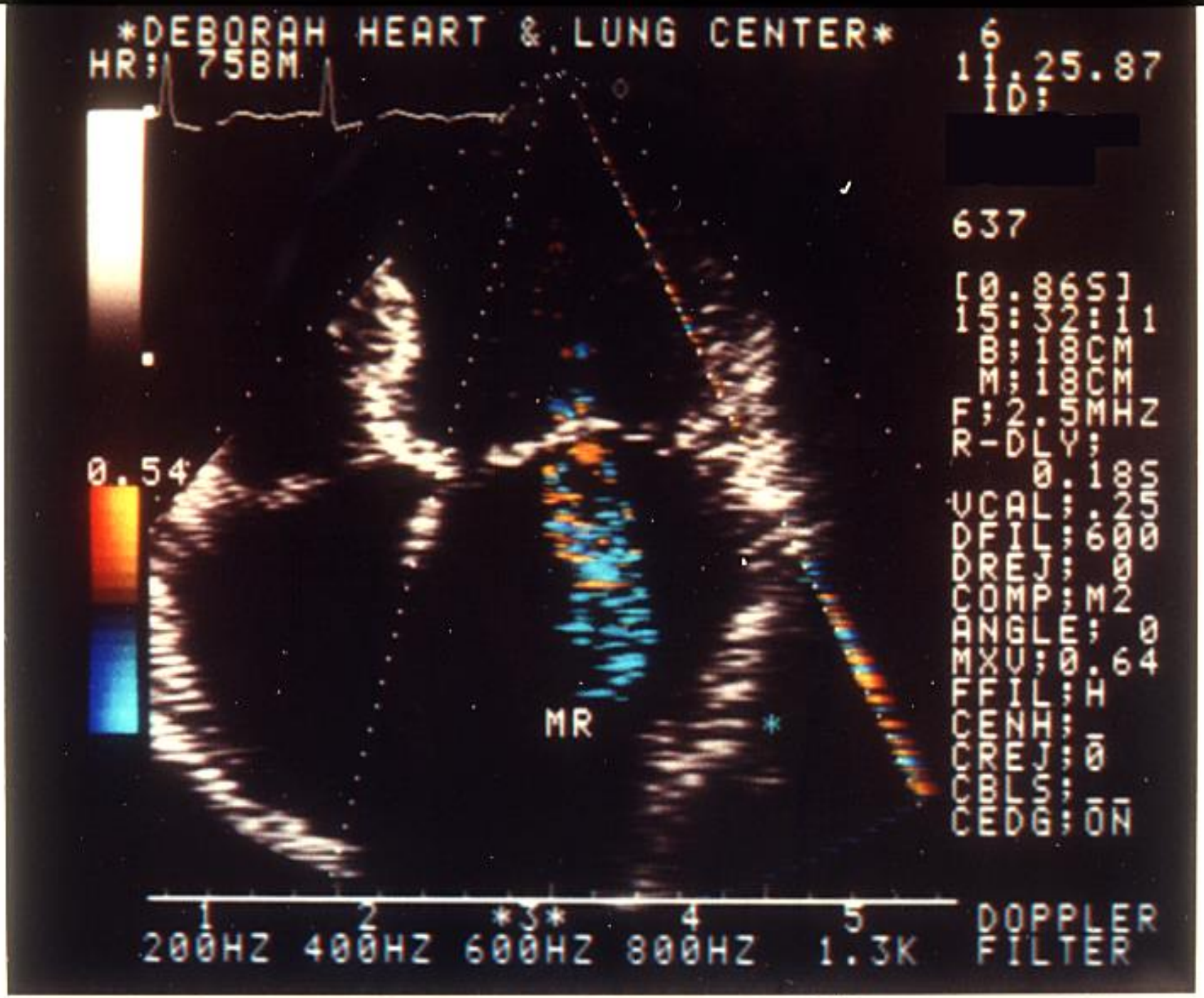
- Auscultation:
  - Murmur = holosystolic
    - apex to axilla (but not always)
    - intensity may not reflect severity
    - ↑ intensity = squatting, isometrics
    - ↓ intensity = Valsalva, amyl nitrite
    - Acute MR = atypical

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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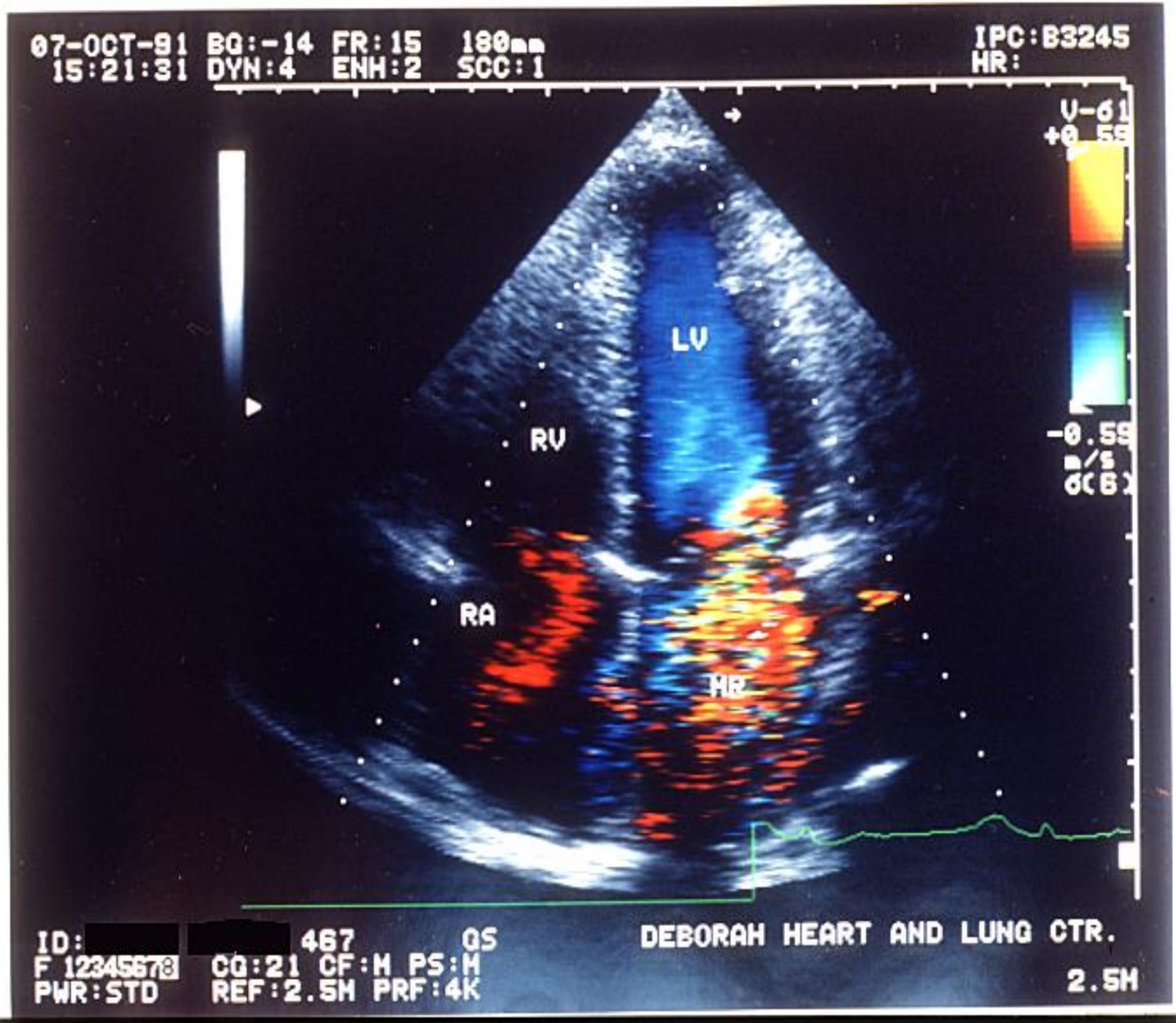
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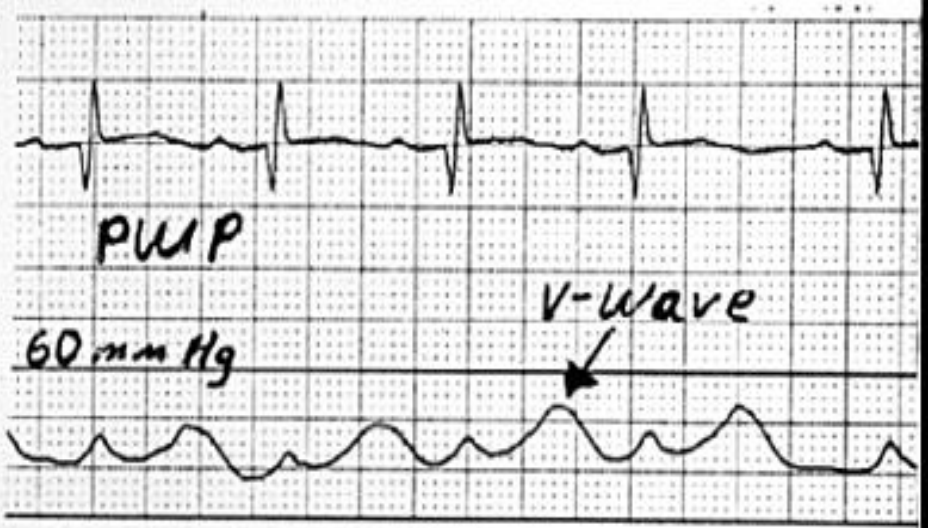


MITRAL REGURGITATION
'V'- WAVES

MR - Treatment:

- Medical (acute) = afterload reduction, diuretics
- Surgical = mitral repair or replacement
- Primary MR:
  - Severe MR with sx.
  - Severe MR without sx. but…
    - 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 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

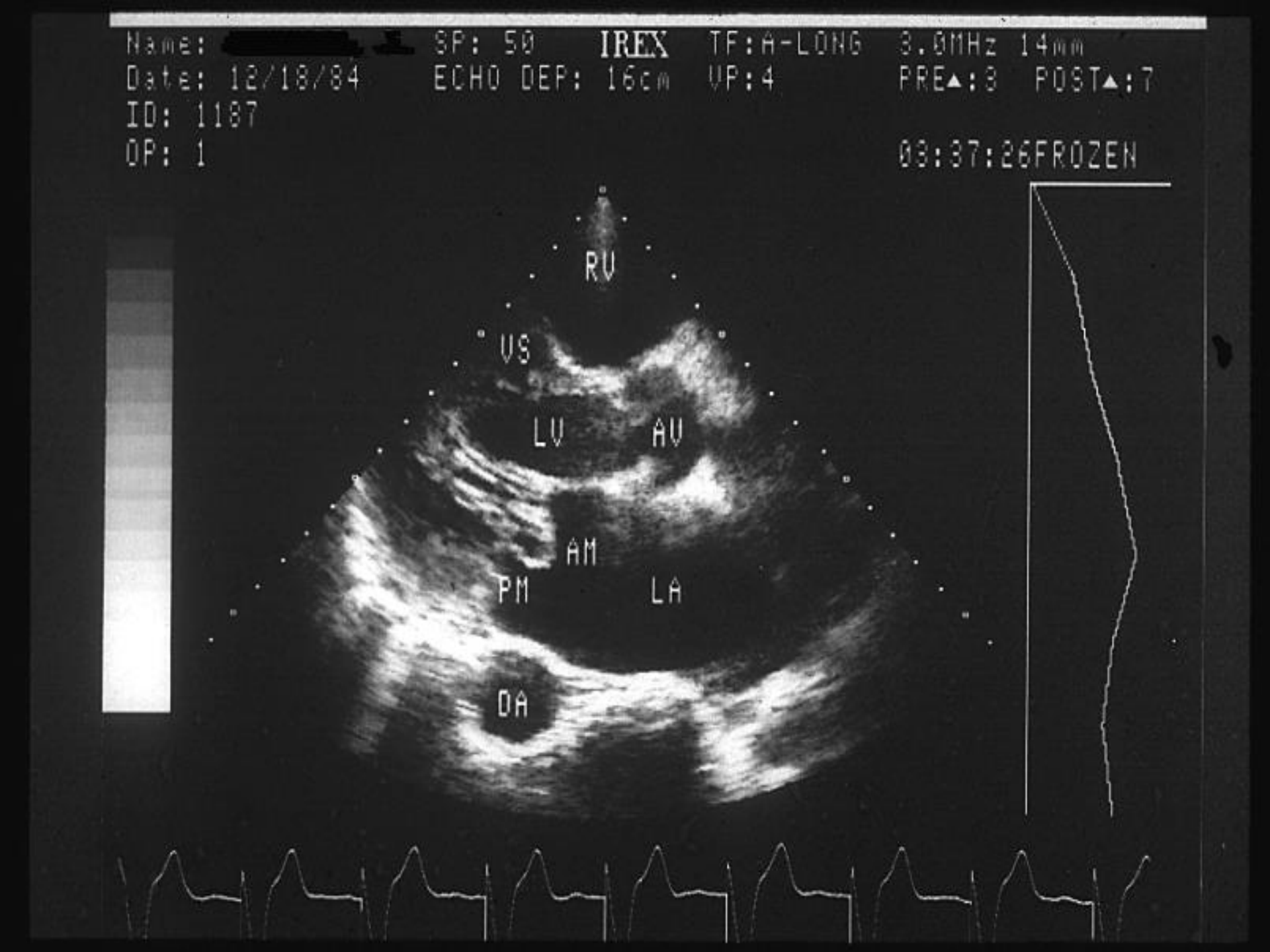
MVP - Non Invasive Testing:

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

MVP - Non Invasive Testing:

- Echo = leaflet abnormalities
- Doppler = quantitate MR

- Stress Testing = false positive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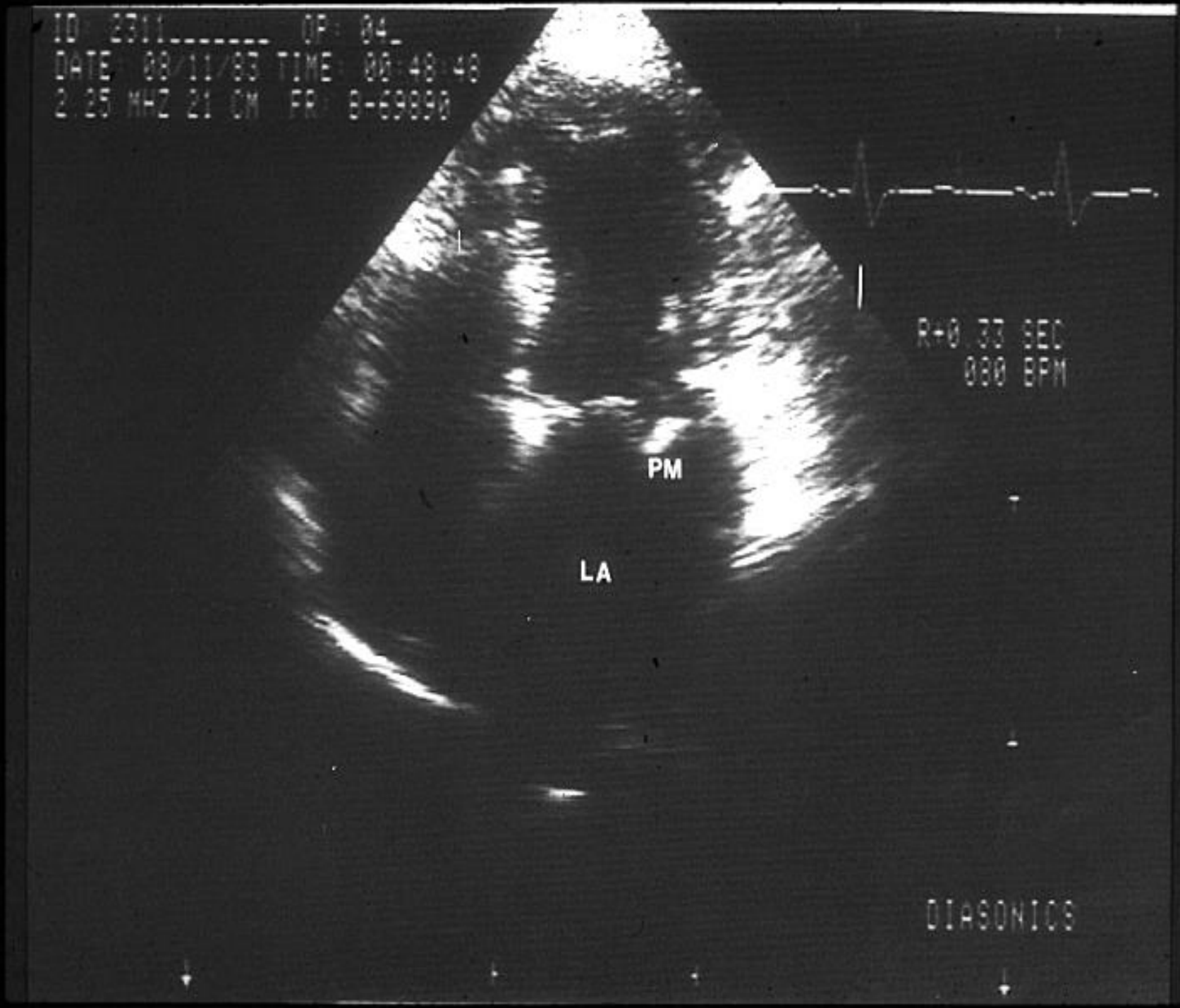
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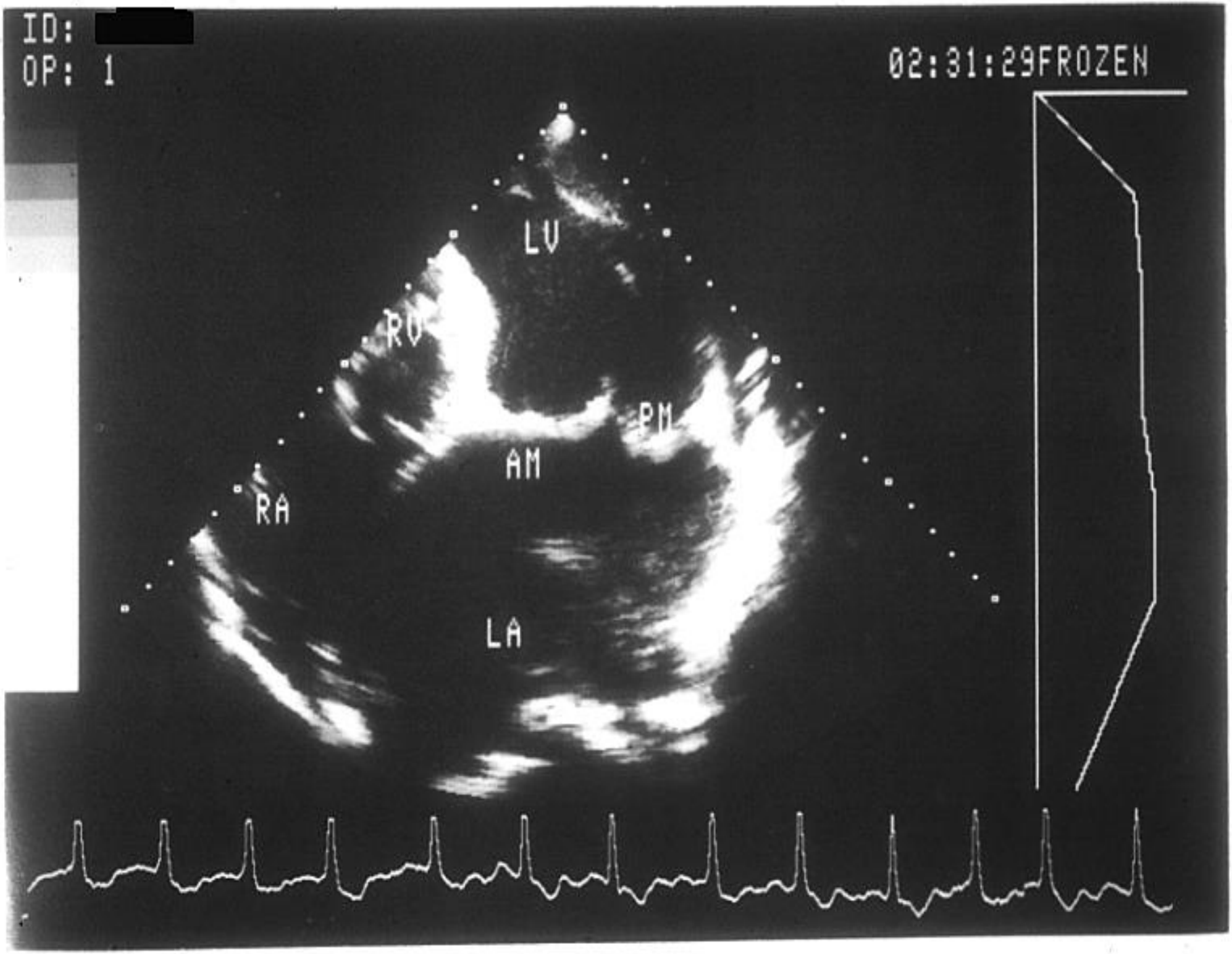
DIASONICS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LV
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MVP - Treatment:

- 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:

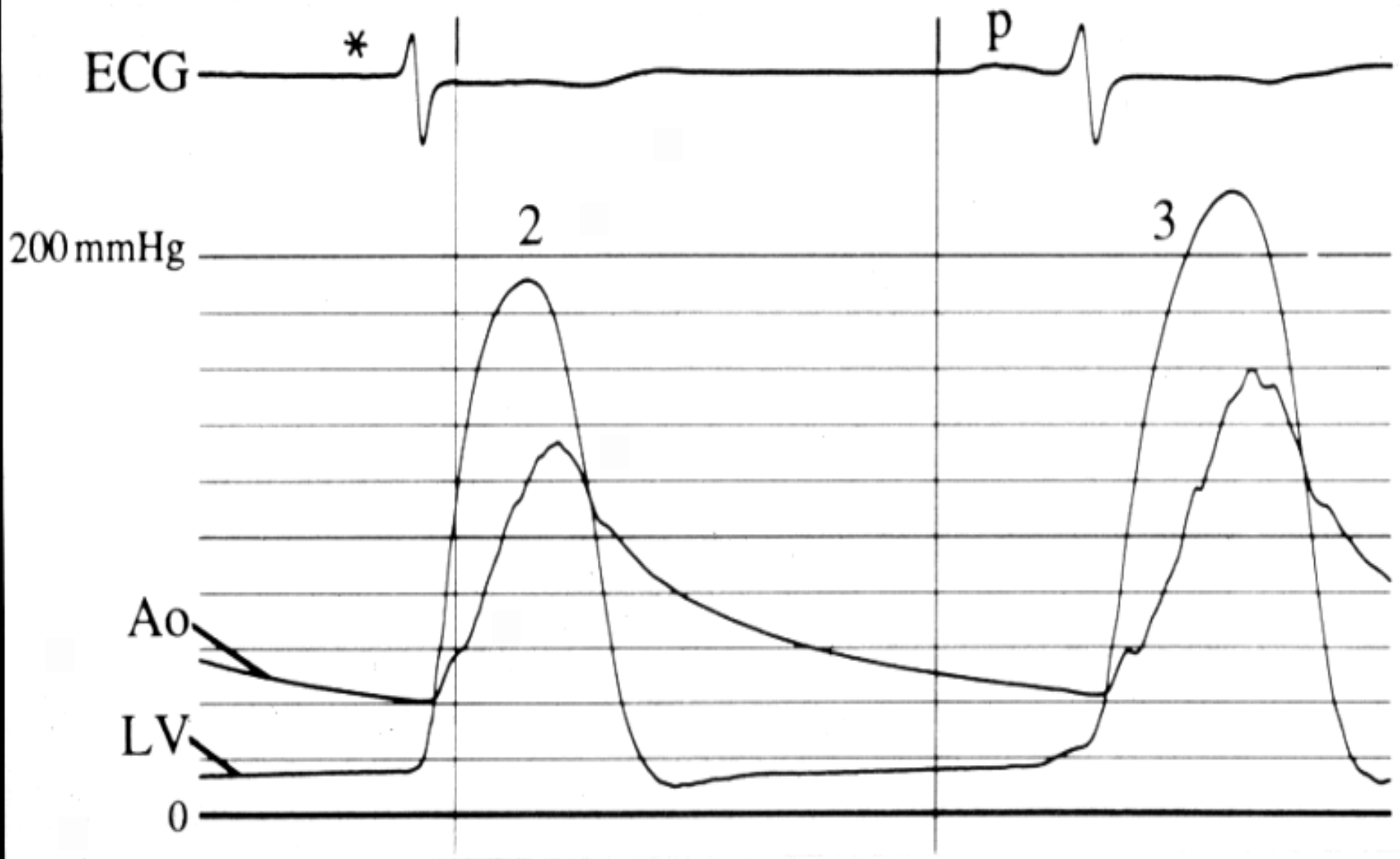
    Valve area > 1.5 cm^2 = mild AS

    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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3
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AS:

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

AS:

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

AS - Auscultation:

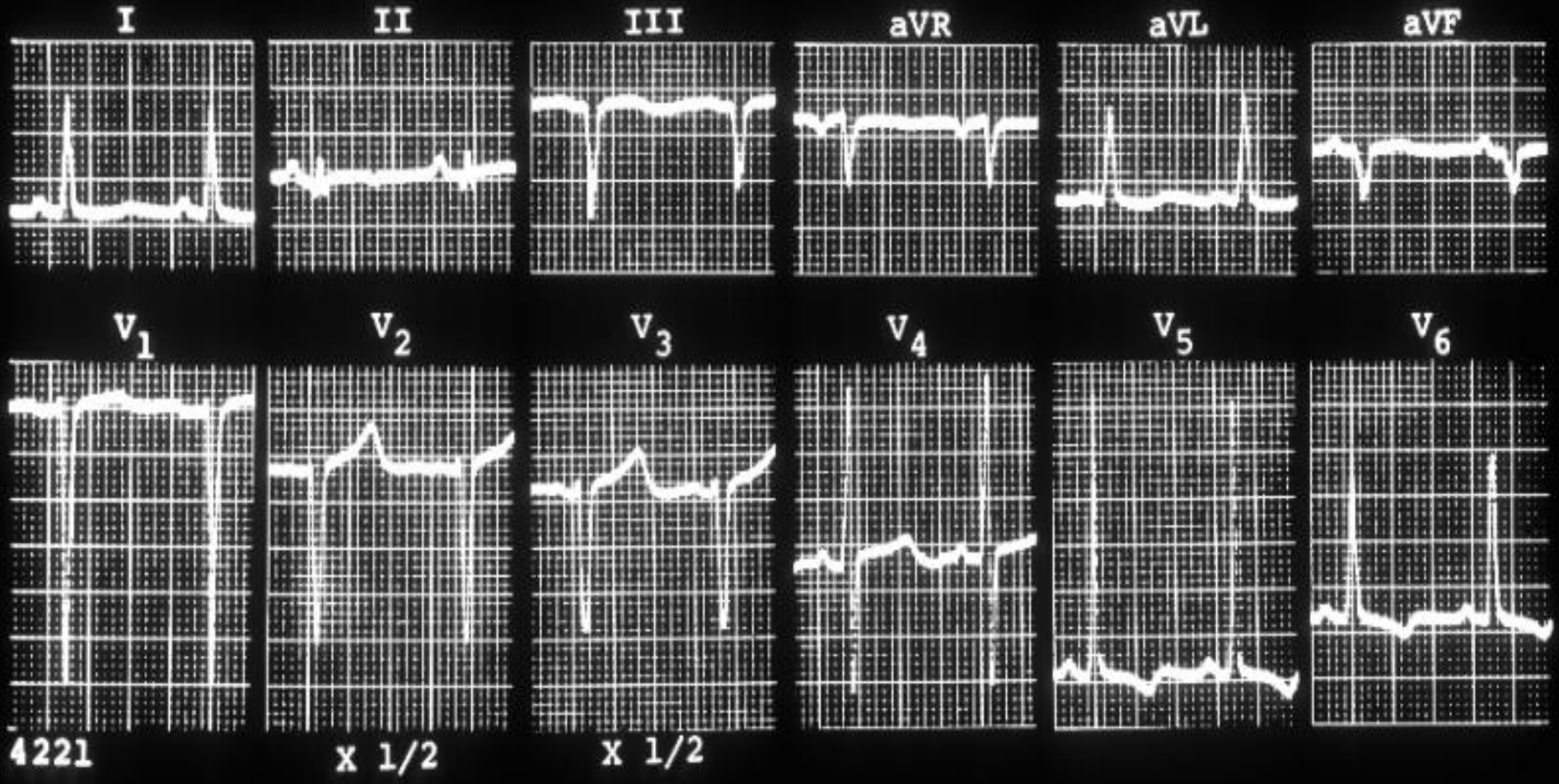
- S_2 = paradoxical split, ↓ intensity (A_2)
- Systolic ejection click = young, congenital
- Systolic ejection murmur
  - base to carotids
  - base to apex = Gallavardin phenomenon
  - severe AS = longer, louder, peaks later

AS:

- 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”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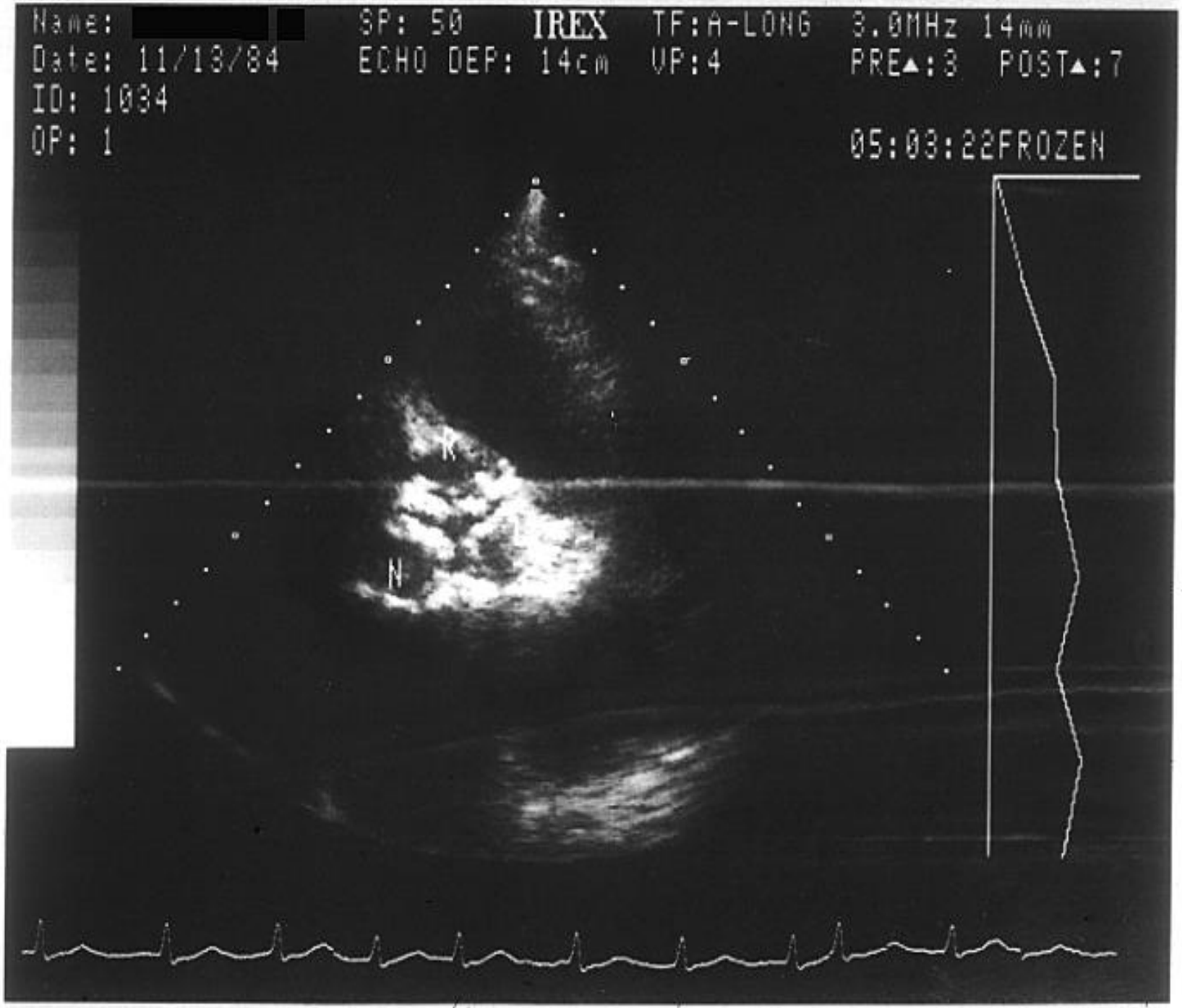
x 1/2

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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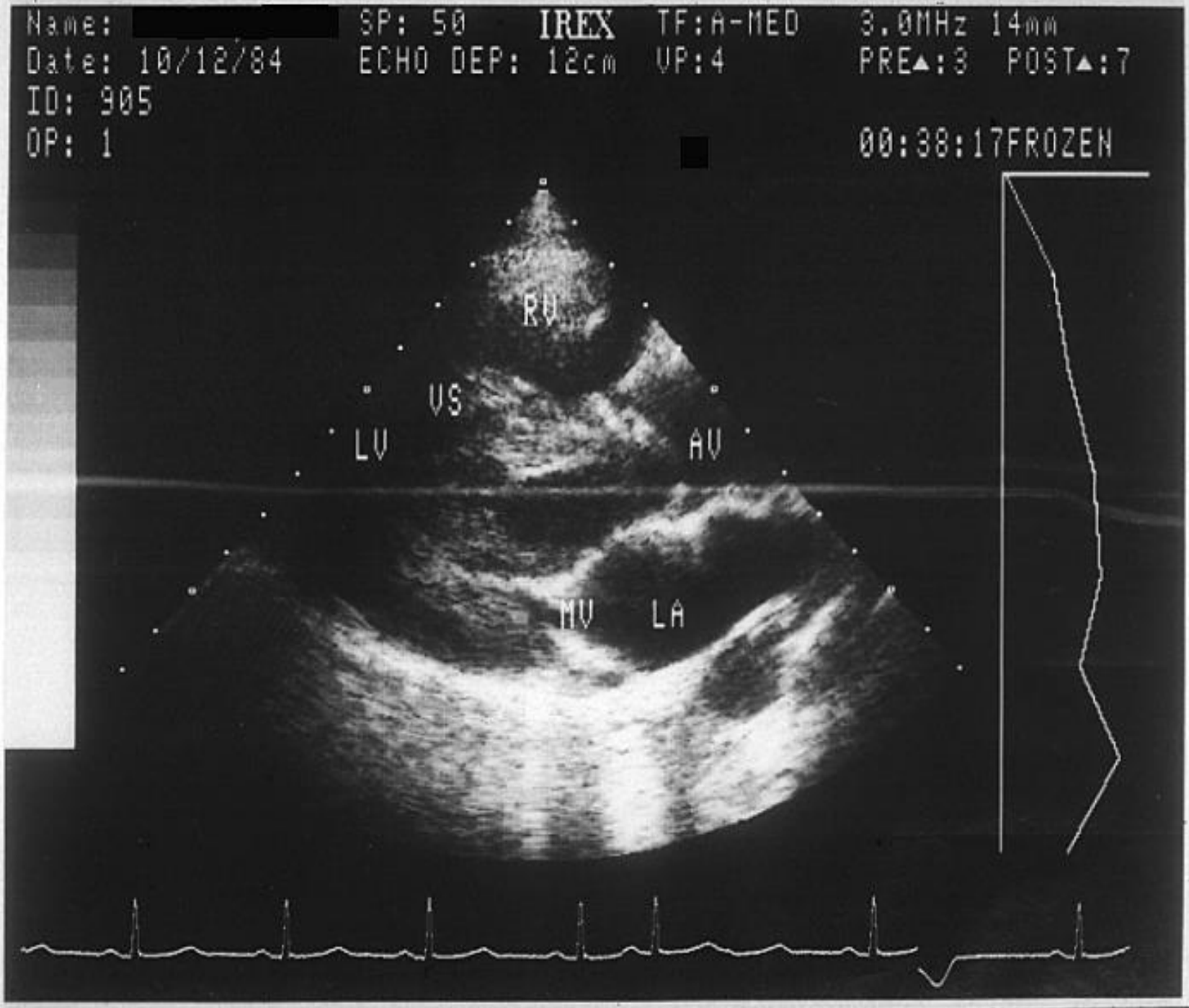
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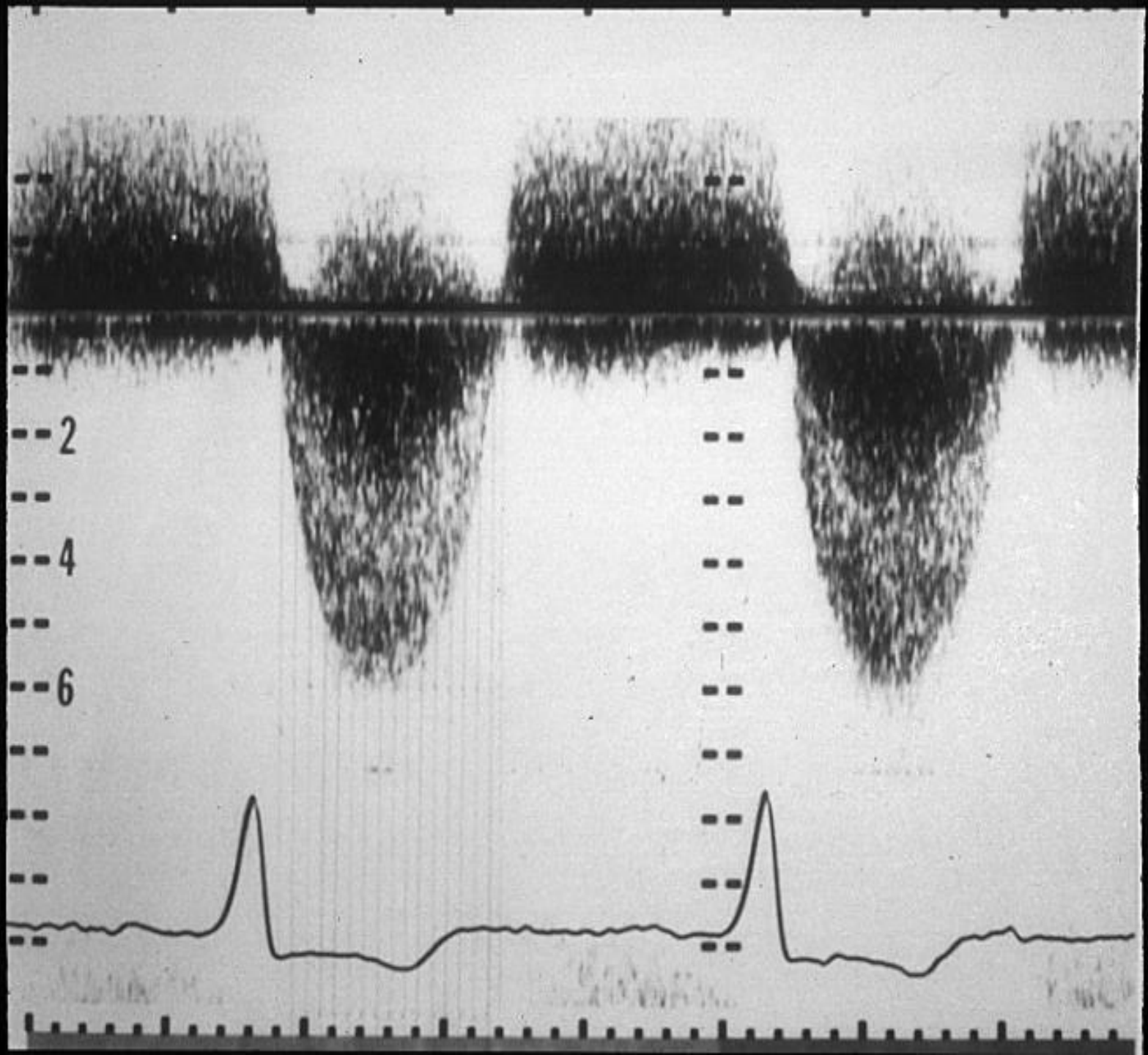
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6

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 (…but beware of the sedentary patient)

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 moderate to high risk patients, may soon be an option in low 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:

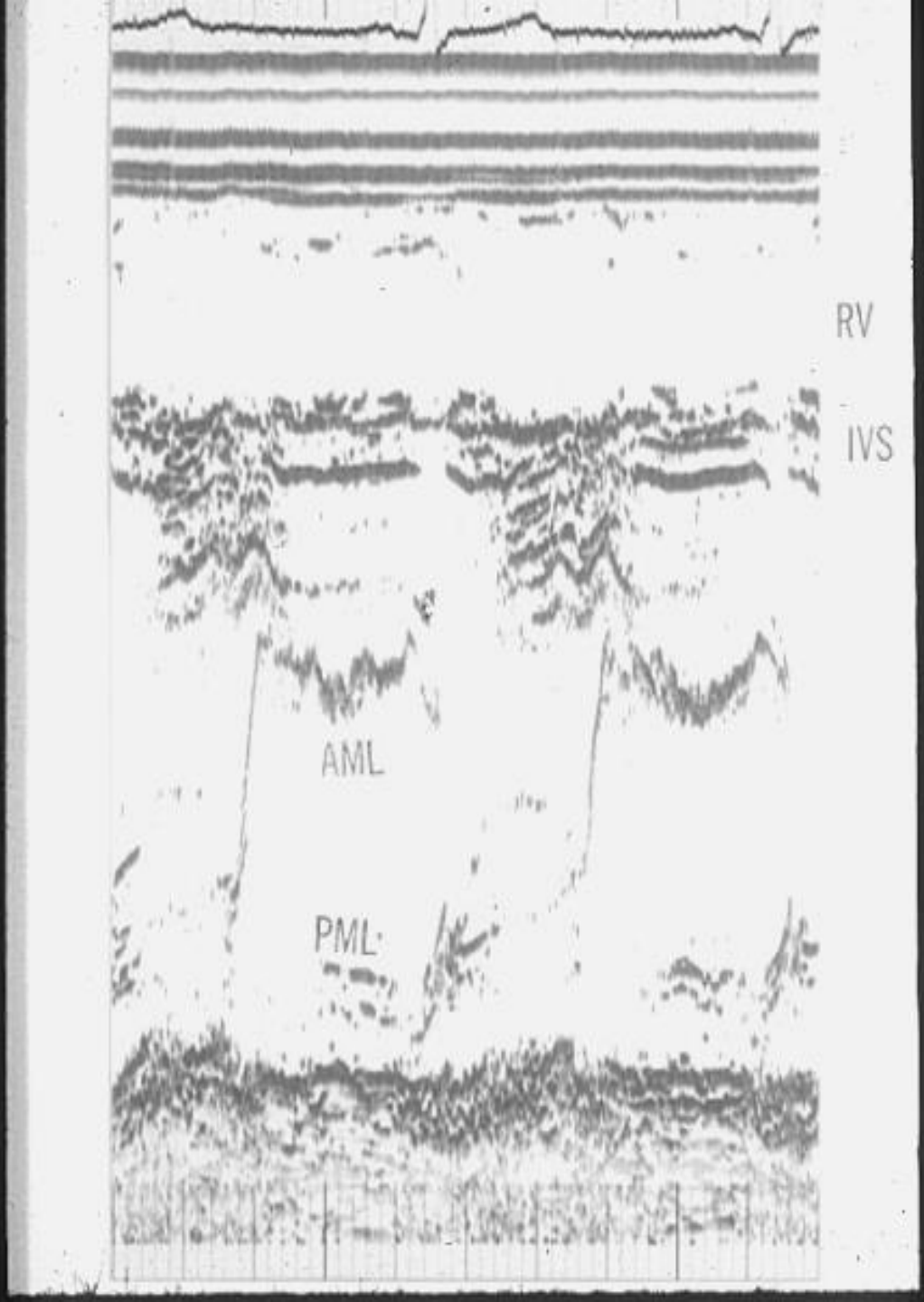
- Dynamic Auscultation

  * ↑ 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) = quantitate severity
- Cardiac MRA = good option if echo equivocal, but $$ and availability ?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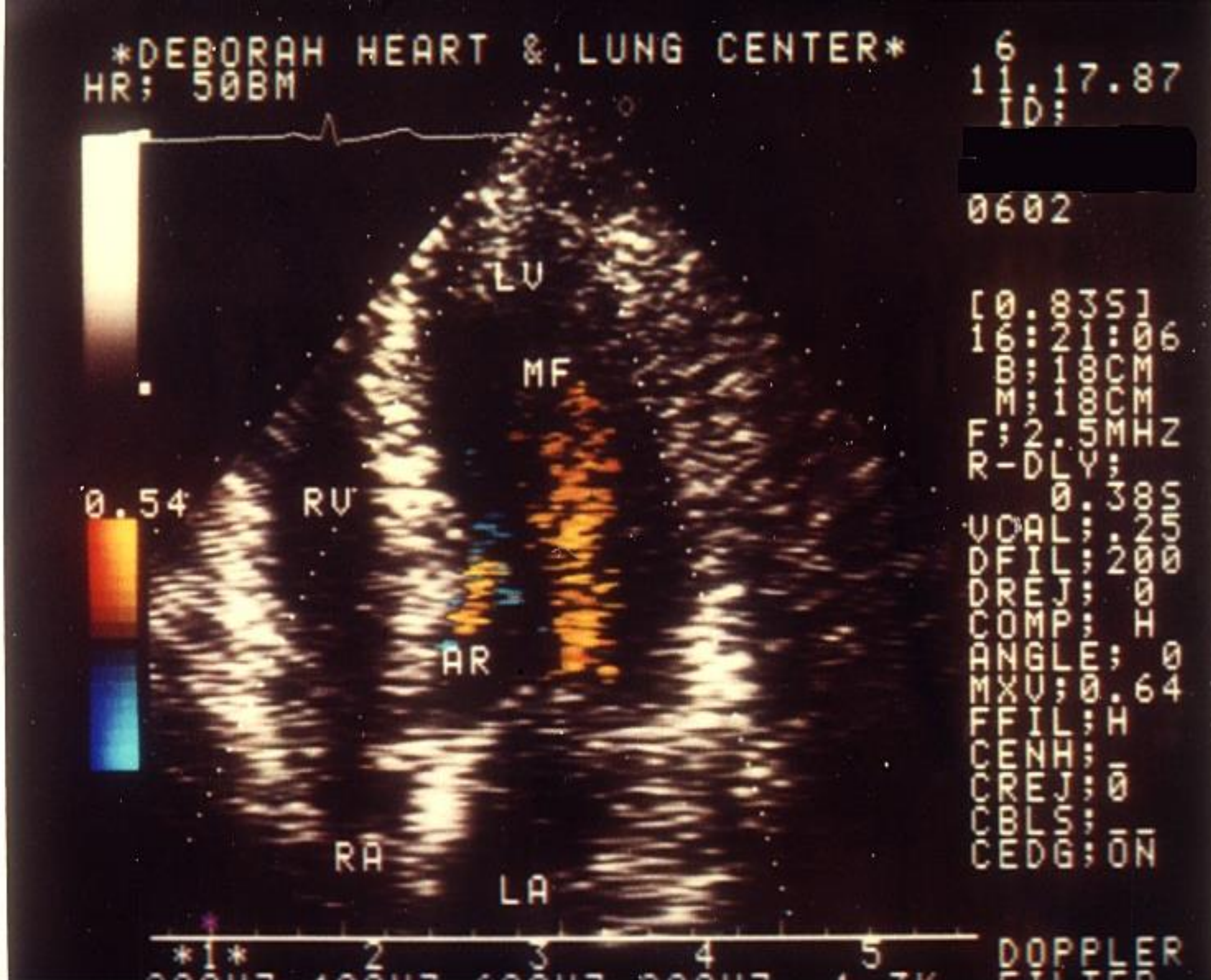
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ANGLE; 0
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PWR:STD REF:2.5M PRF:4K
DEBORAH HEART AND LUNG CTR.
2.5M

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%

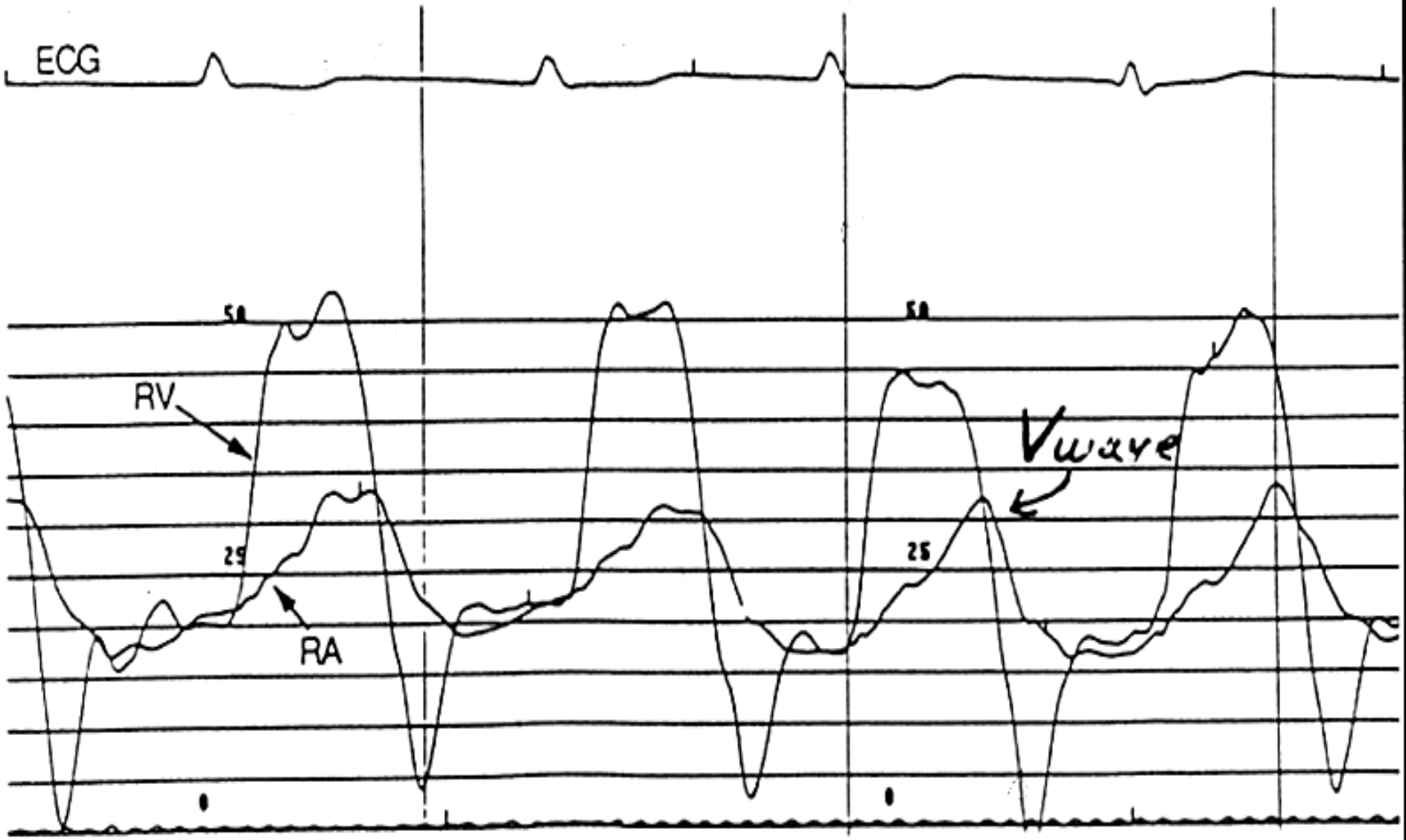
    or .. * ESD > 50 mm (echo measurement)

Tricuspid Stenosis:

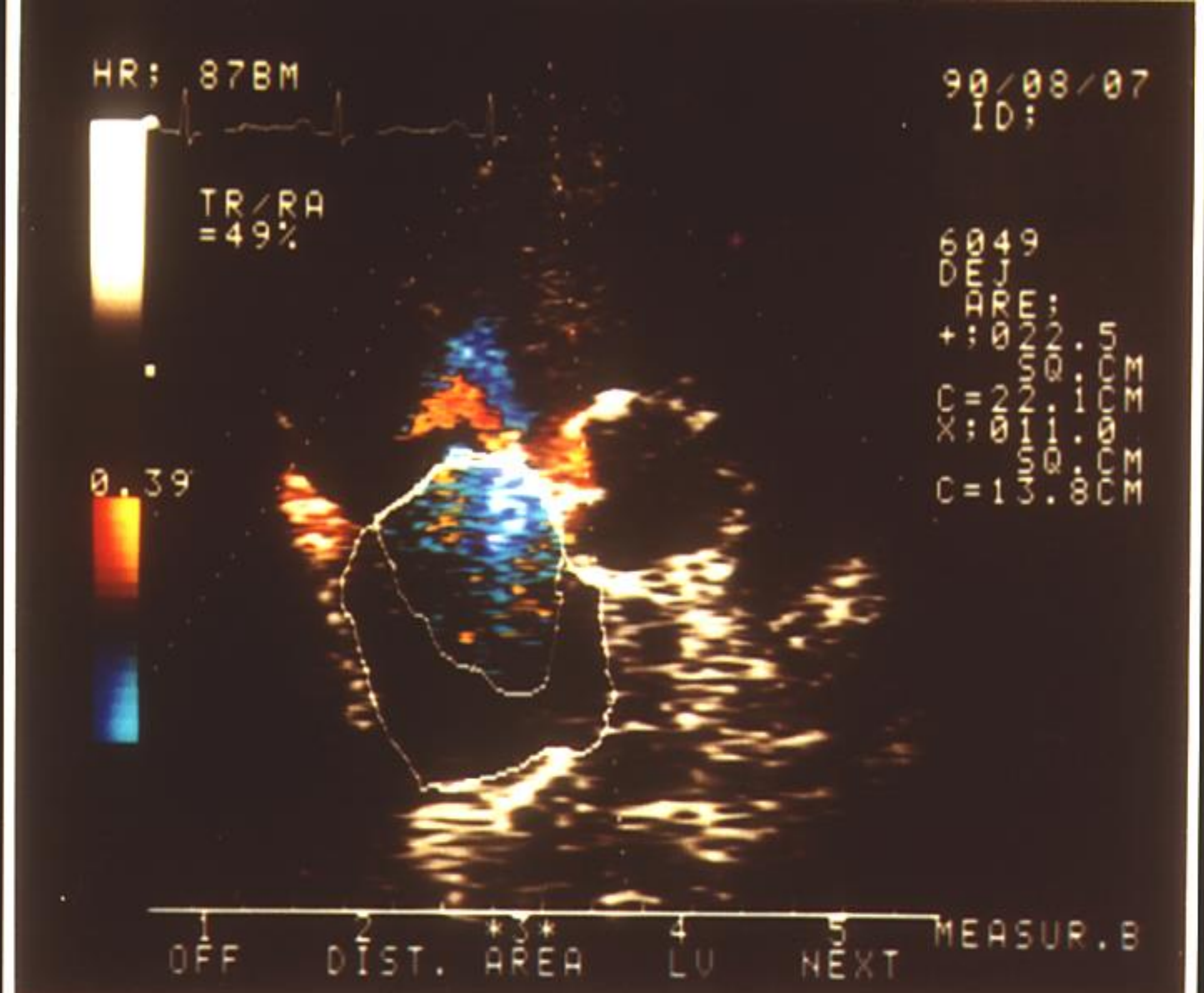
- Etiology = rheumatic, congenital, carcinoid

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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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
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Acc: 10520936
2008 Oct 27
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ET: 1
TR: 6.4
TE: 1.9
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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
  - *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 (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 $\rightarrow$ 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:

  * 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

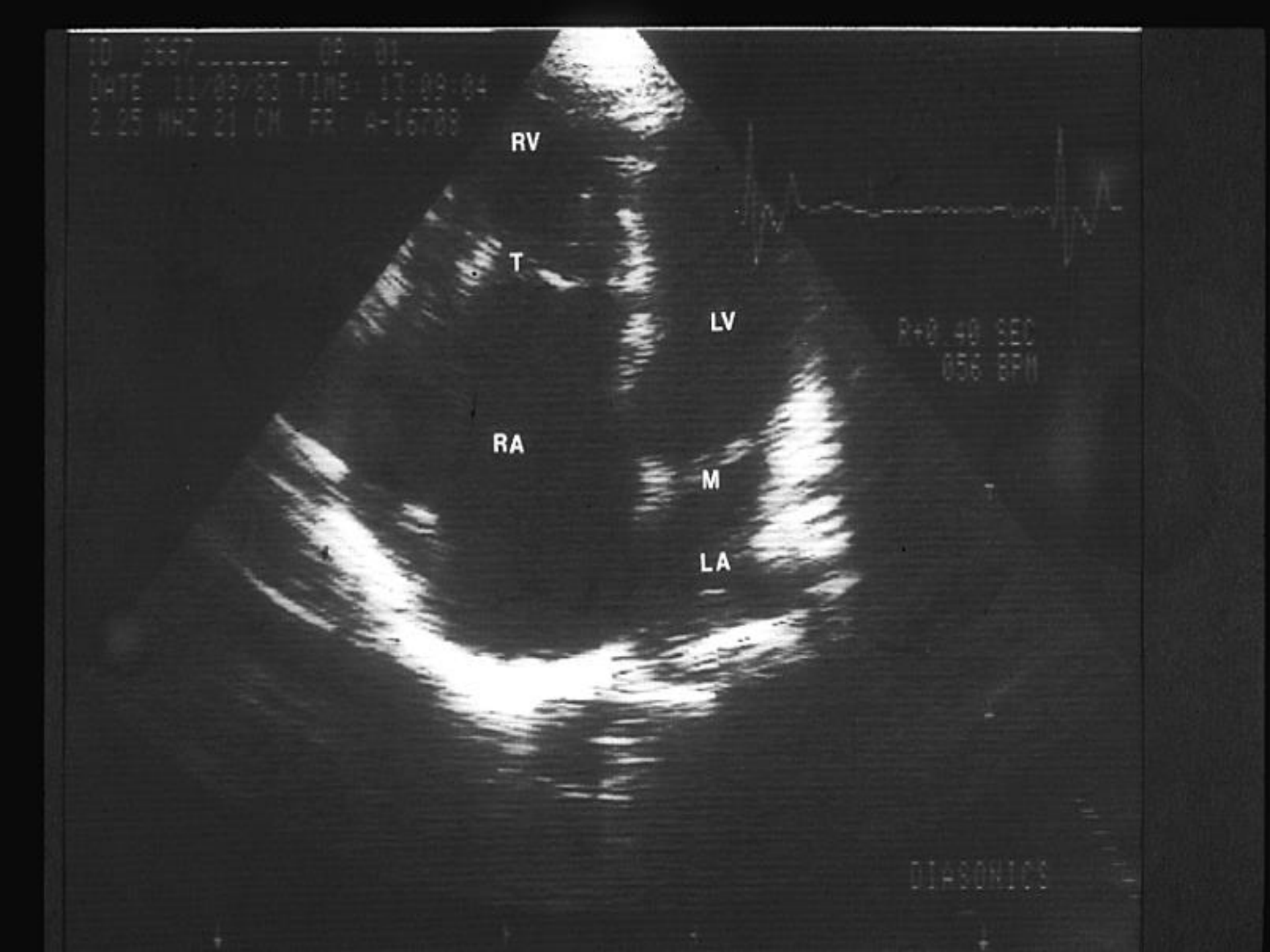
Tetralogy of Fallot (TOF):

- 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:

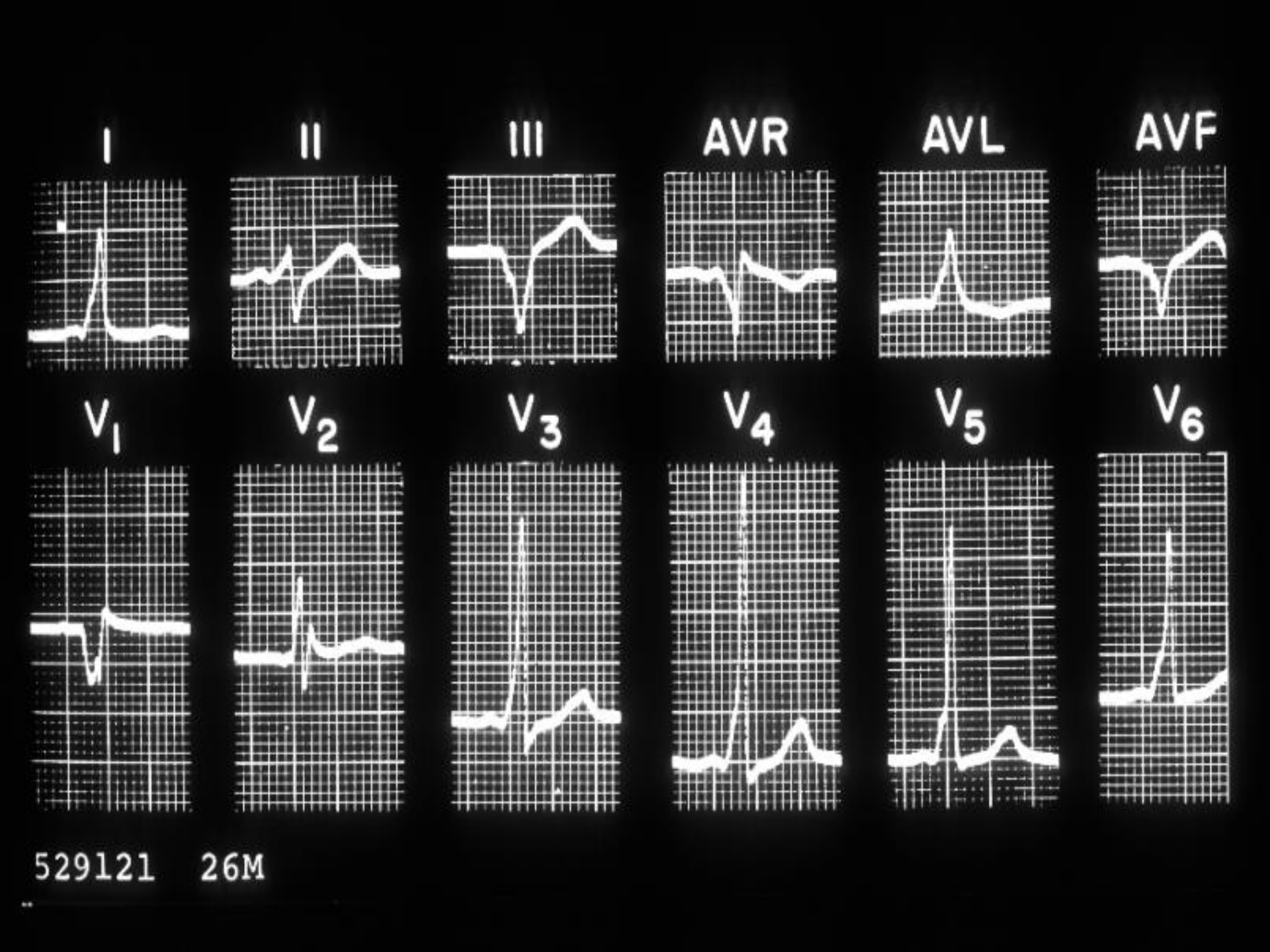
- L – Transposition (congenitally corrected):
  - * AV discordance and ventriculo-arterial discordance
  - * Morphologic RV = systemic ventricle
  - * Morphologic LV = venous ventricle
  - * Function = blood follows normal course
  - * Survival into adulthood
  - *Problems = systemic A-V valve regurg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

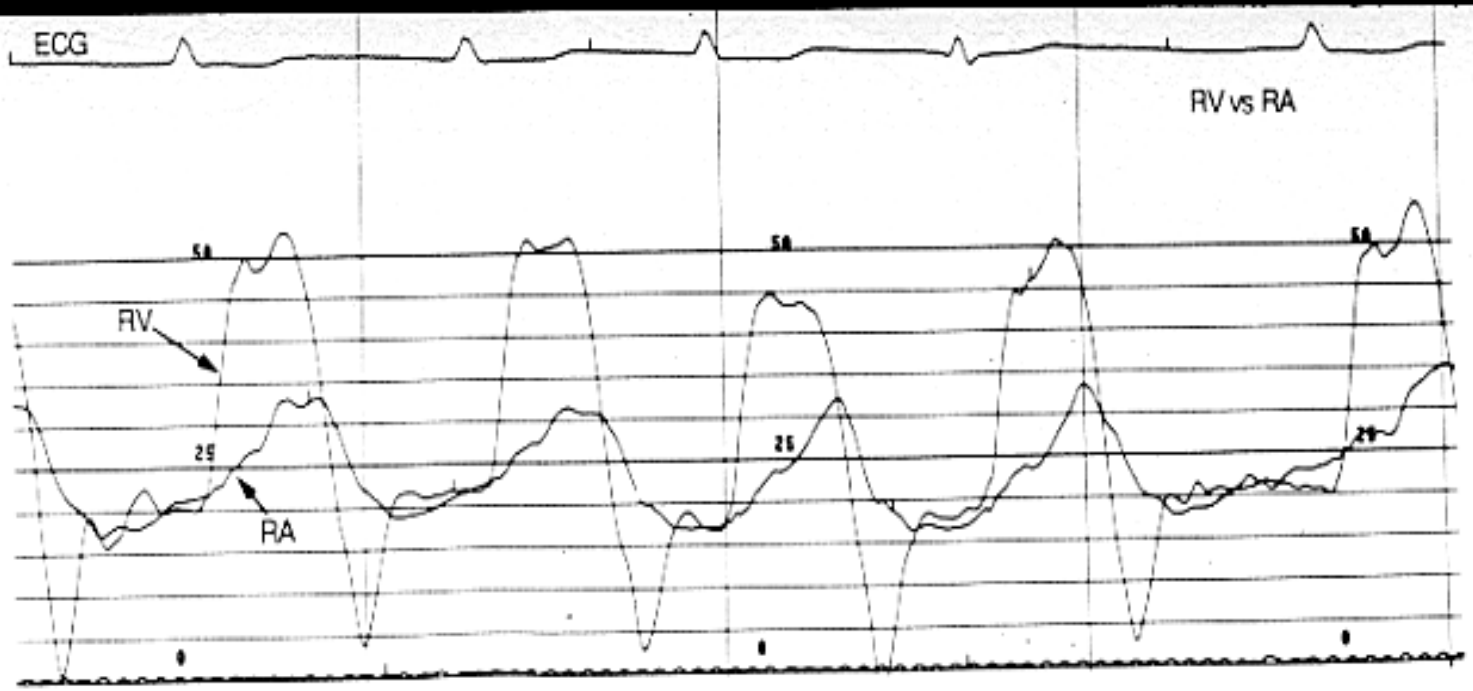
Congenital Disease - Summary:

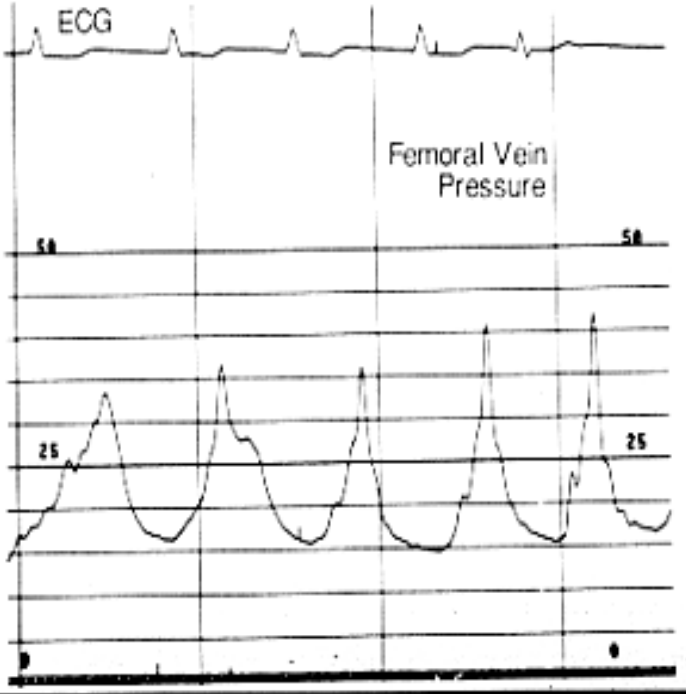
- L → R shunt = non-cyanotic
  - * ASD, VSD, PDA, Persistent truncus
- R → L shunt = cyanotic
  - * TOF (± cyanosis)
  - * Tricuspid atresia
  - * Complete transposition (“D”)
  - * Double outlet RV

Congenital Disease - Summary:

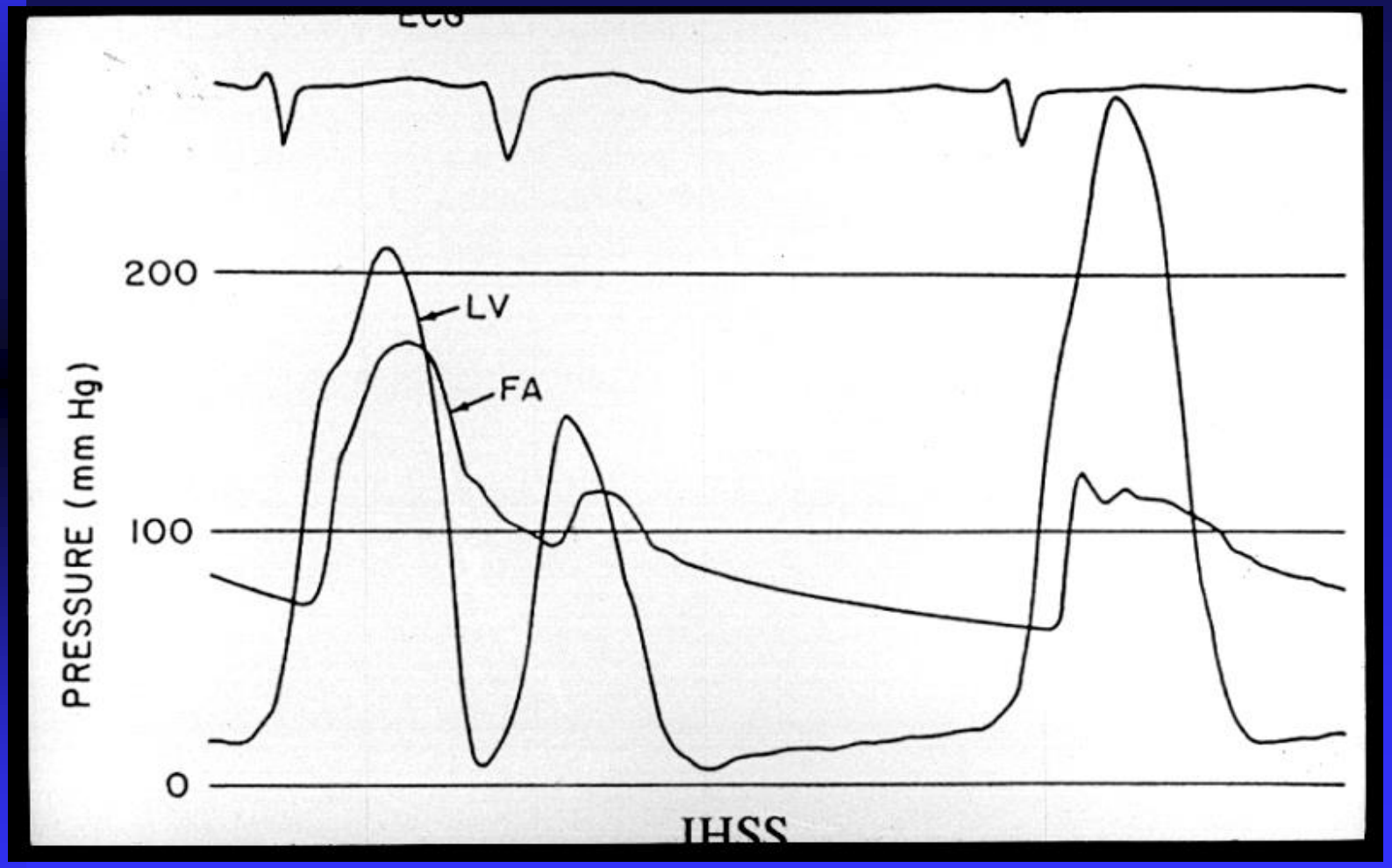
- Survival to adulthood:
  - * Bicuspid aortic valve
  - * Coarctation of aorta
  - * Pulmonic stenosis
  - * Secundum ASD
  - * PDA

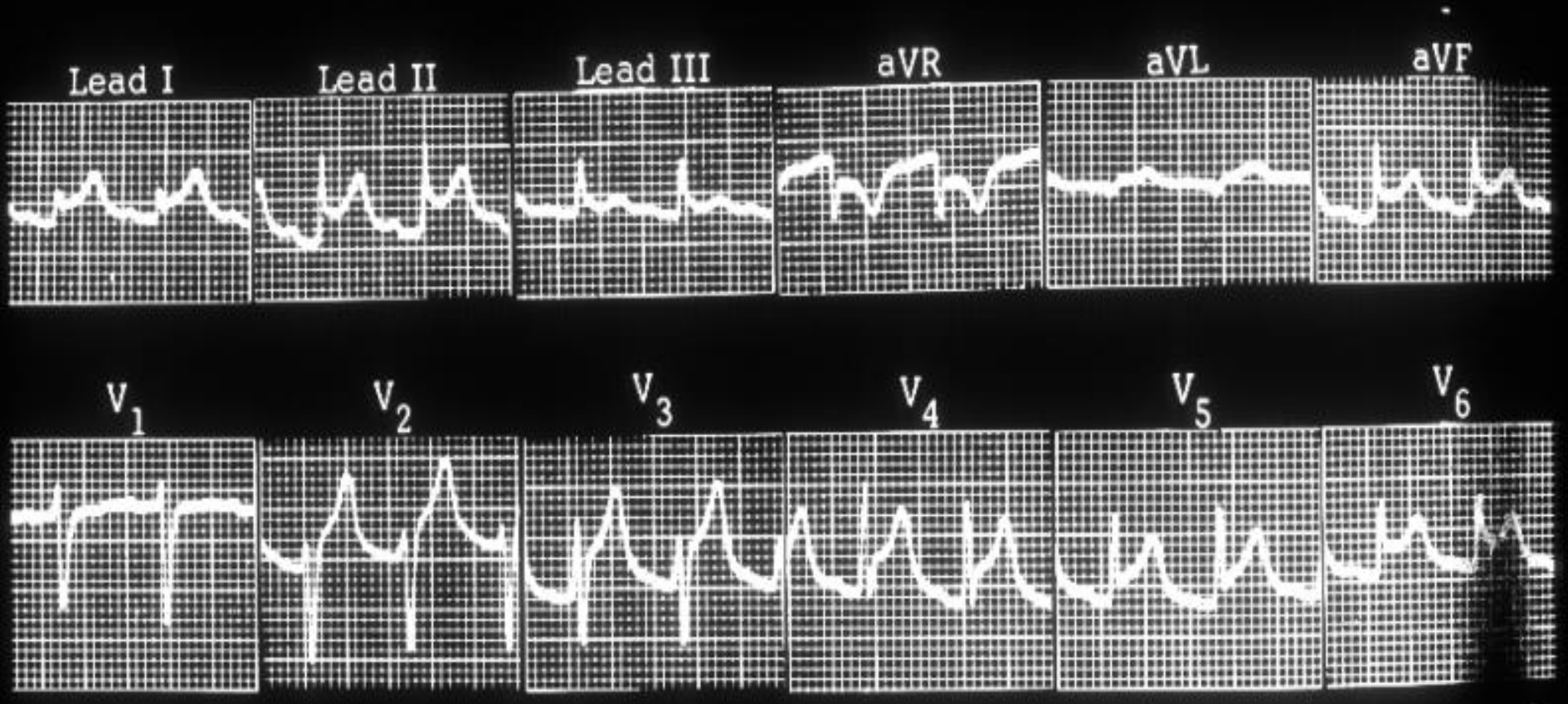

I
II
III
AVR
AVL
AVF
V1
V2
V3
V4
V5
V6
529121 26M

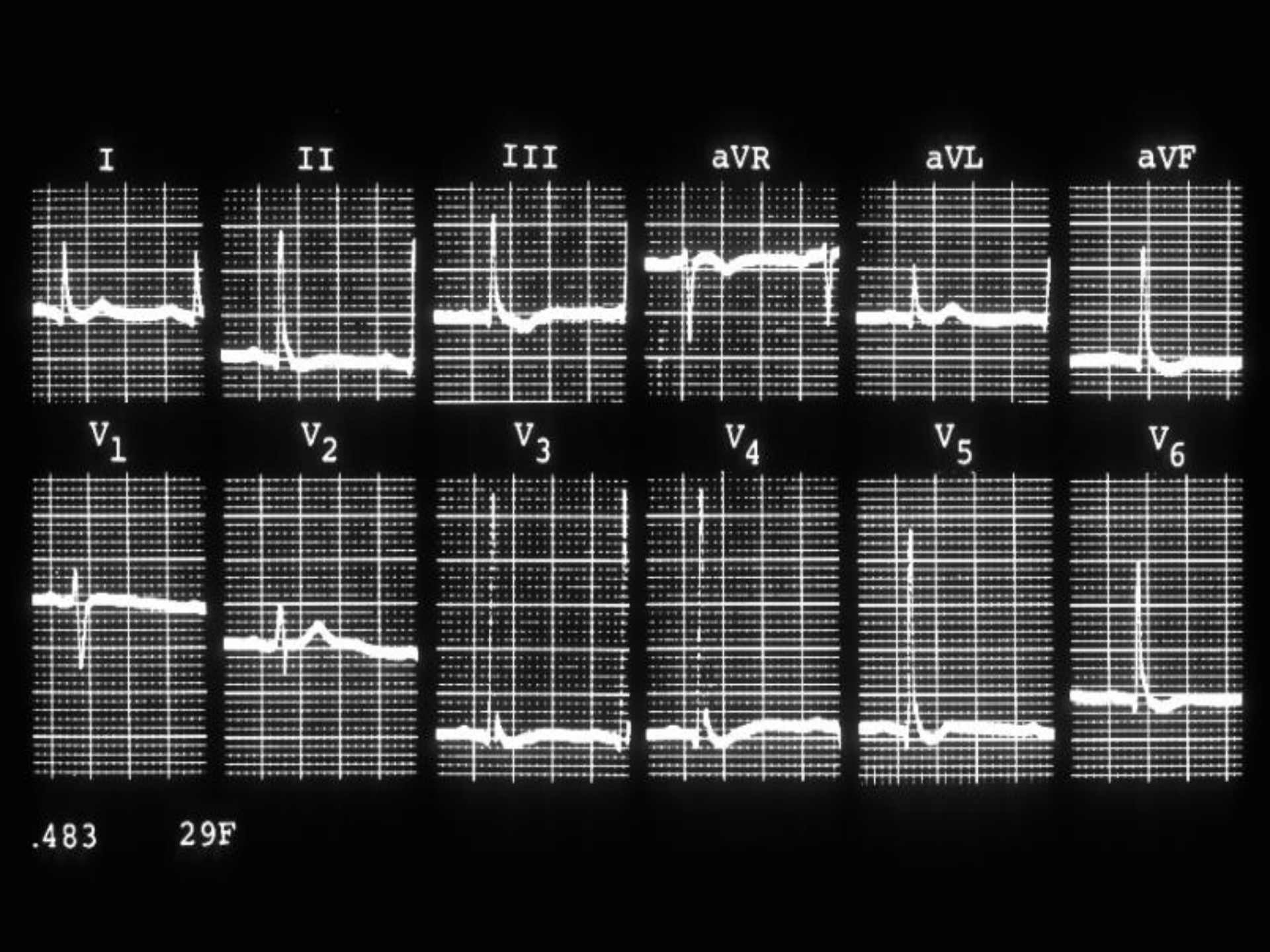

ECG
RV vs RA
RV
RA
50
25
0

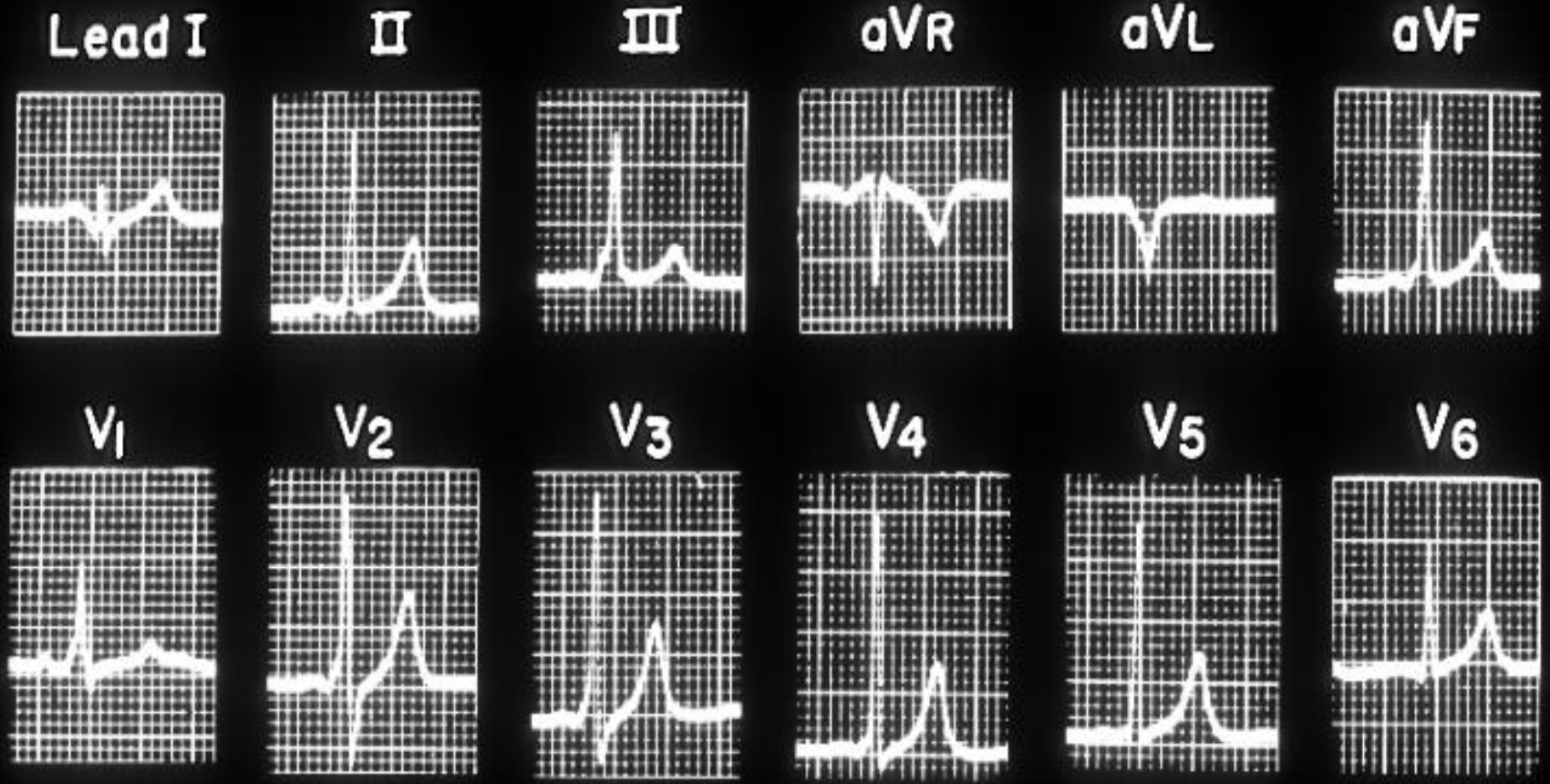

ECG
Femoral Vein
Pressure
50
25
0

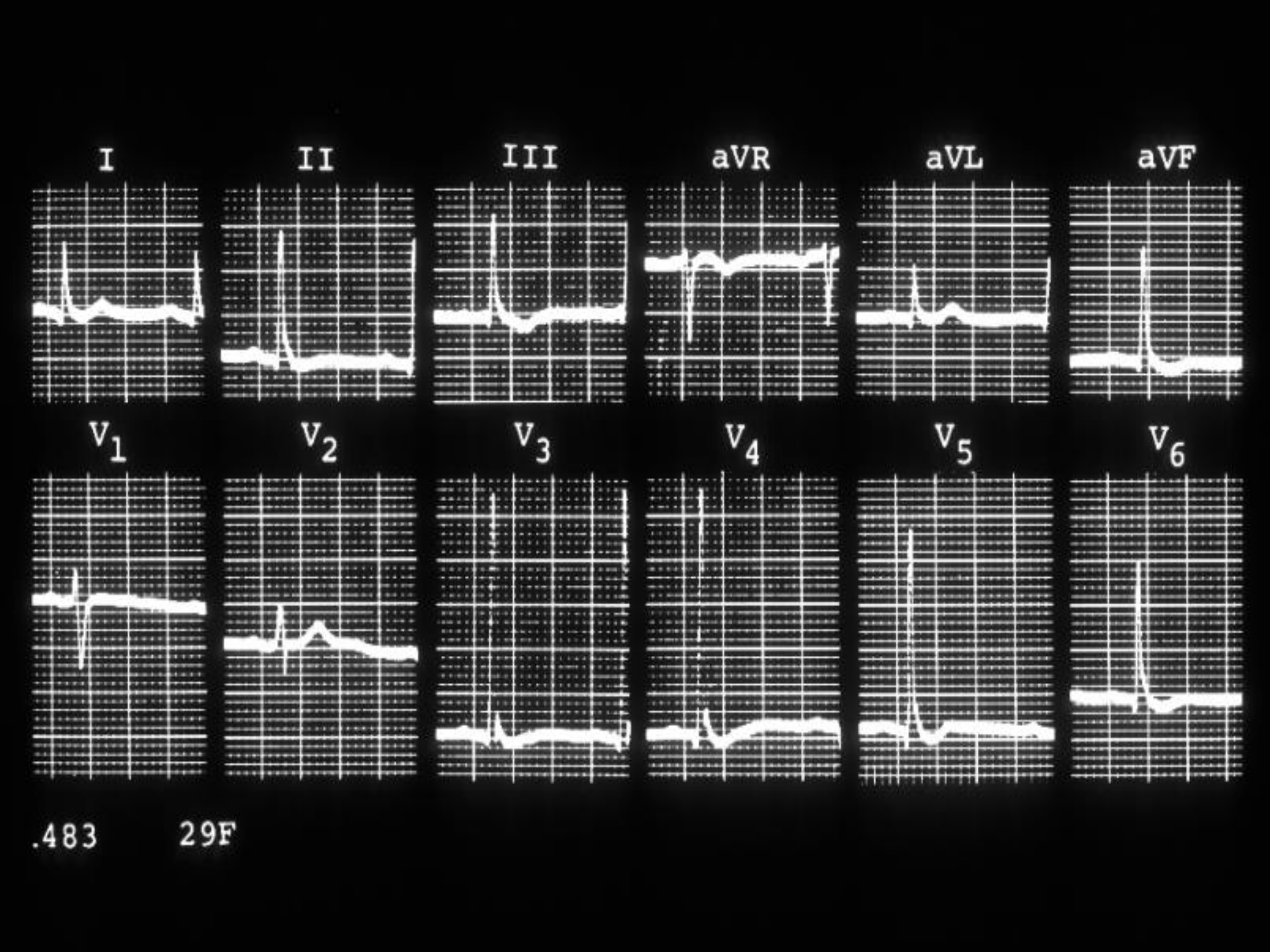

I
II
III
V1
V3
V5
5102

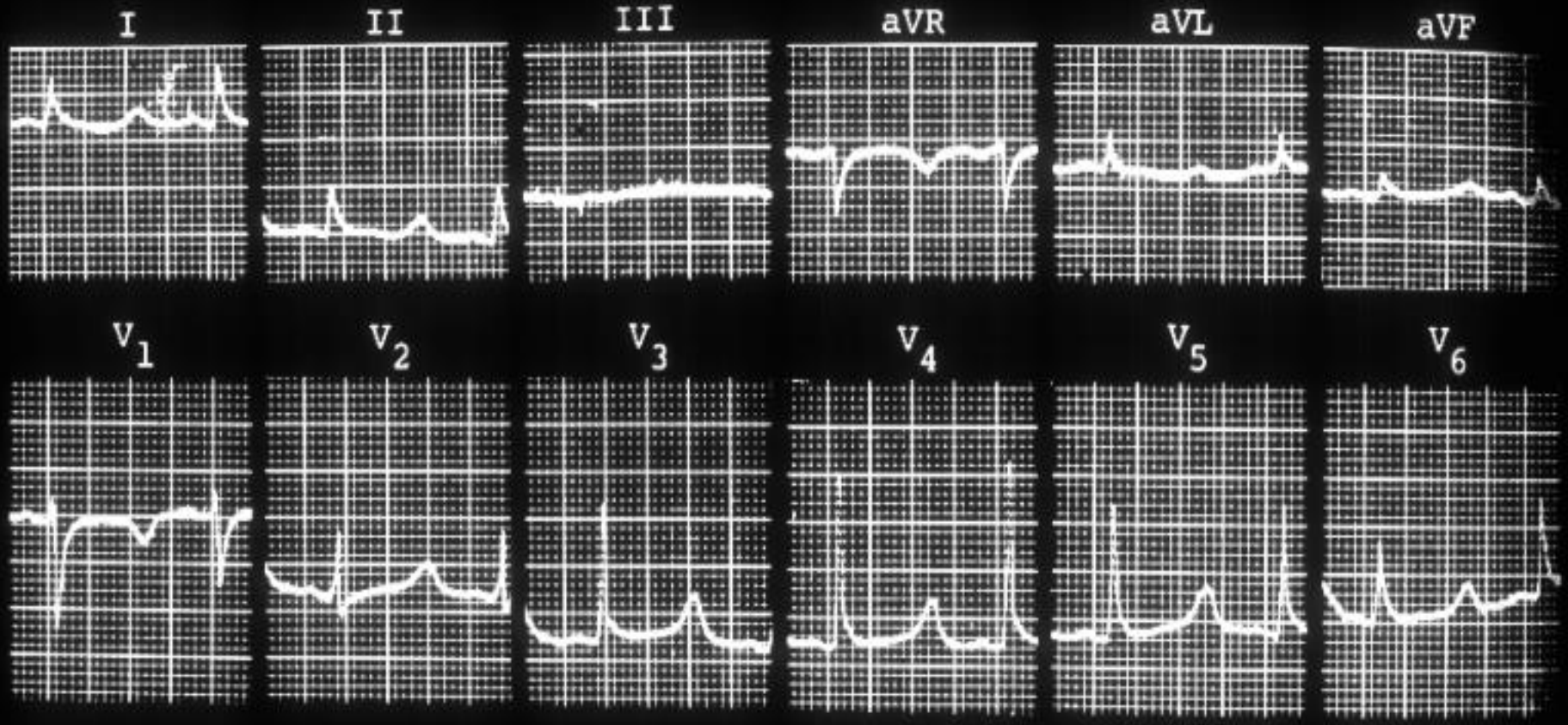

ECG
200
100
0
LV
FA
PRESSURE (mm Hg)
IHSS


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