# TRP Session:
# Adult Congenital Heart Disease

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# Agenda

- Useful study resources
- Three cases with questions to answer; requiring participation
  - Raise your hand to be called upon
- “Rapid Fire” questions, time permitting

# Canadian CCS Guidelines on ACHD

- 2009
- 2020 (due soon!)

www.ccs.ca

Canadian Cardiovascular Society

# International Guidelines

www.escardio.org

# Useful Websites

www.cachnet.org

CANADIAN ADULT CONGENITAL HEART NETWORK

www.cchaforlife.org

CANADIAN CONGENITAL HEART ALLIANCE

www.isachd.org

International Society for Adult Congenital Heart Disease

# Useful Websites

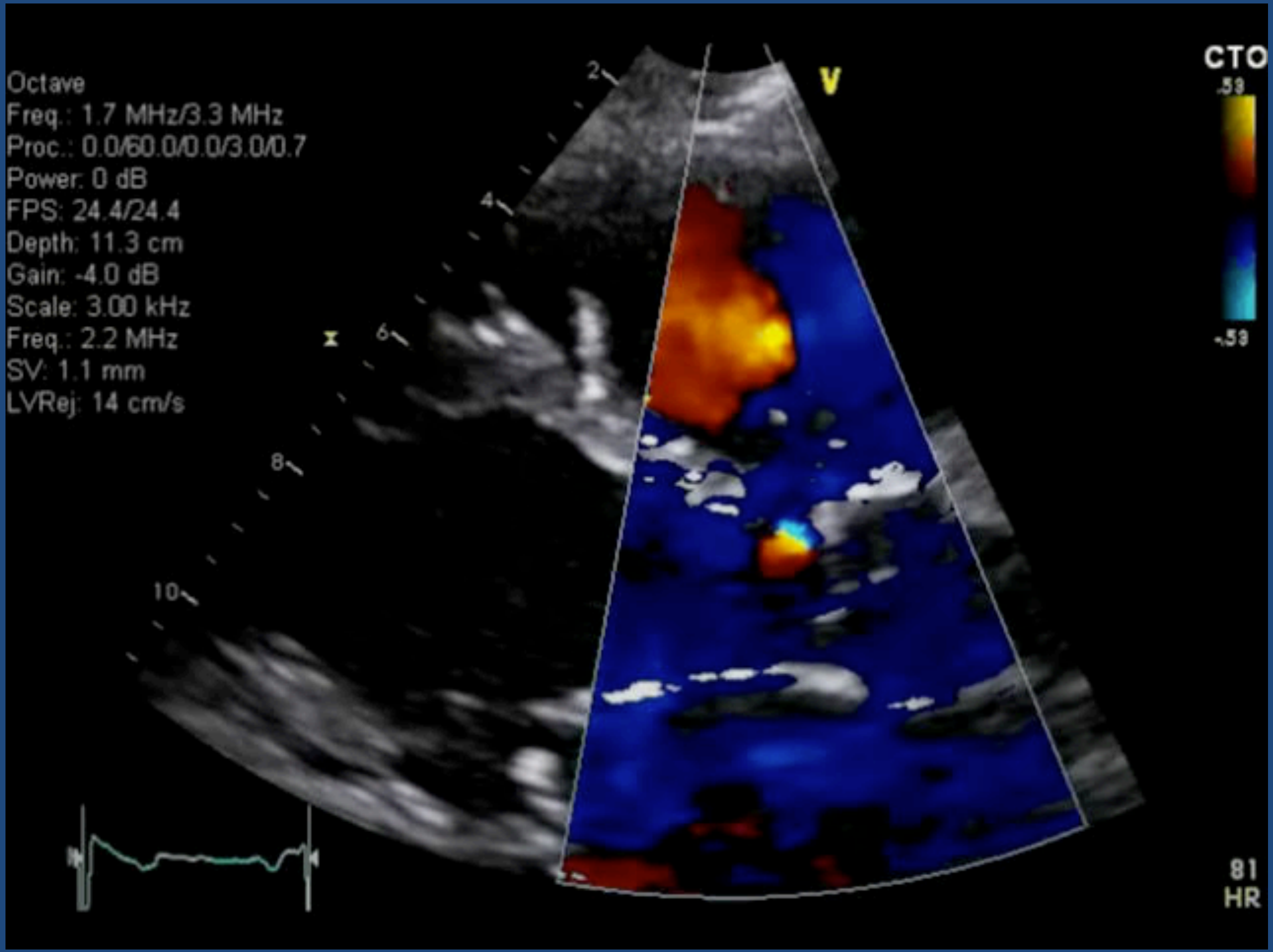
HEART DISEASE
& PREGNANCY

www.heartdiseaseandpregnancy.com

# Case #1

- 32 G2P1, 36 weeks pregnant
- What is the congenital abnormality?
- What requires urgent attention?

- There are 8 echo clips to view.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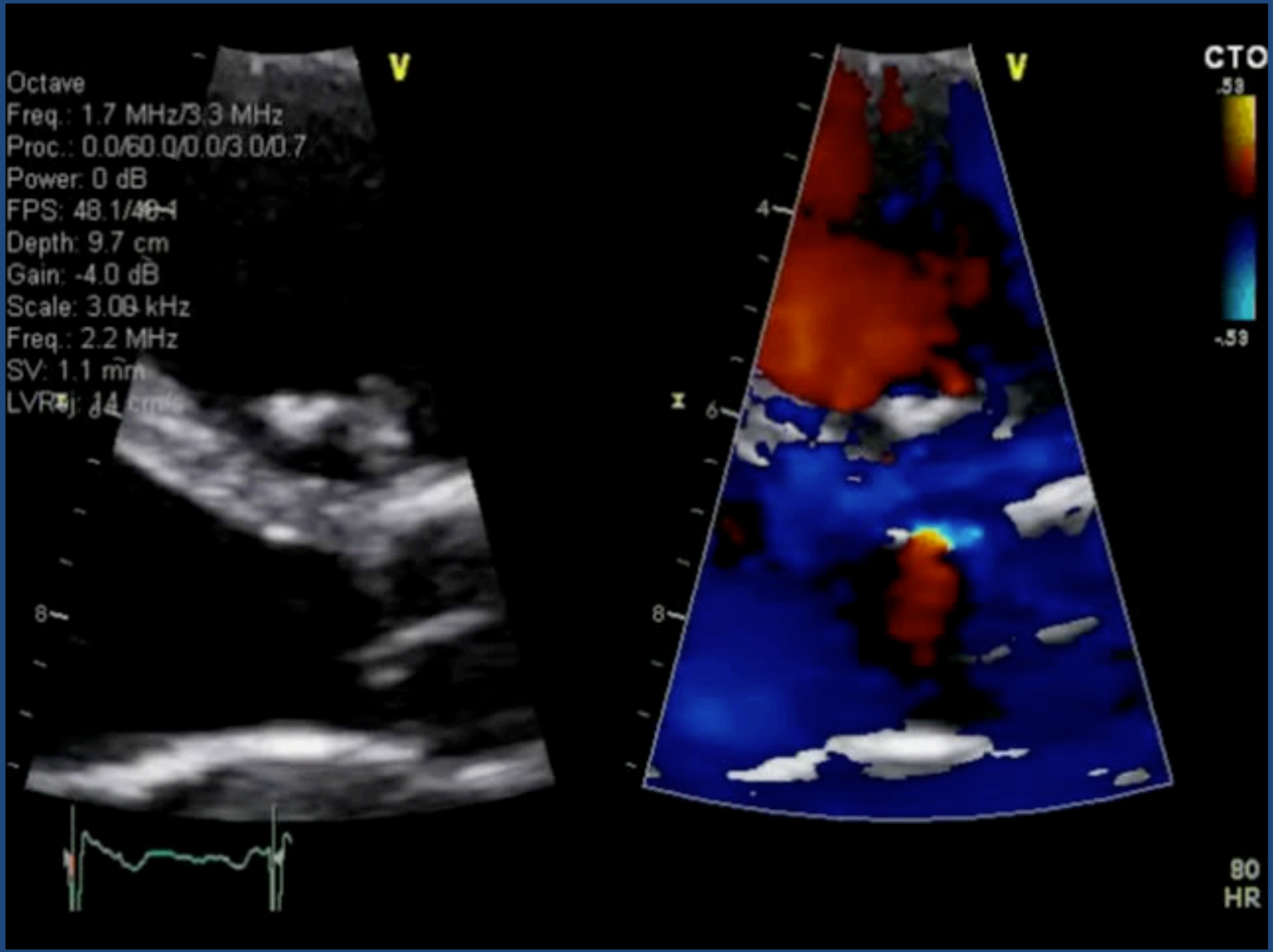
LVRej: 14 cm/s
V
CTO
.59
-.59
81
HR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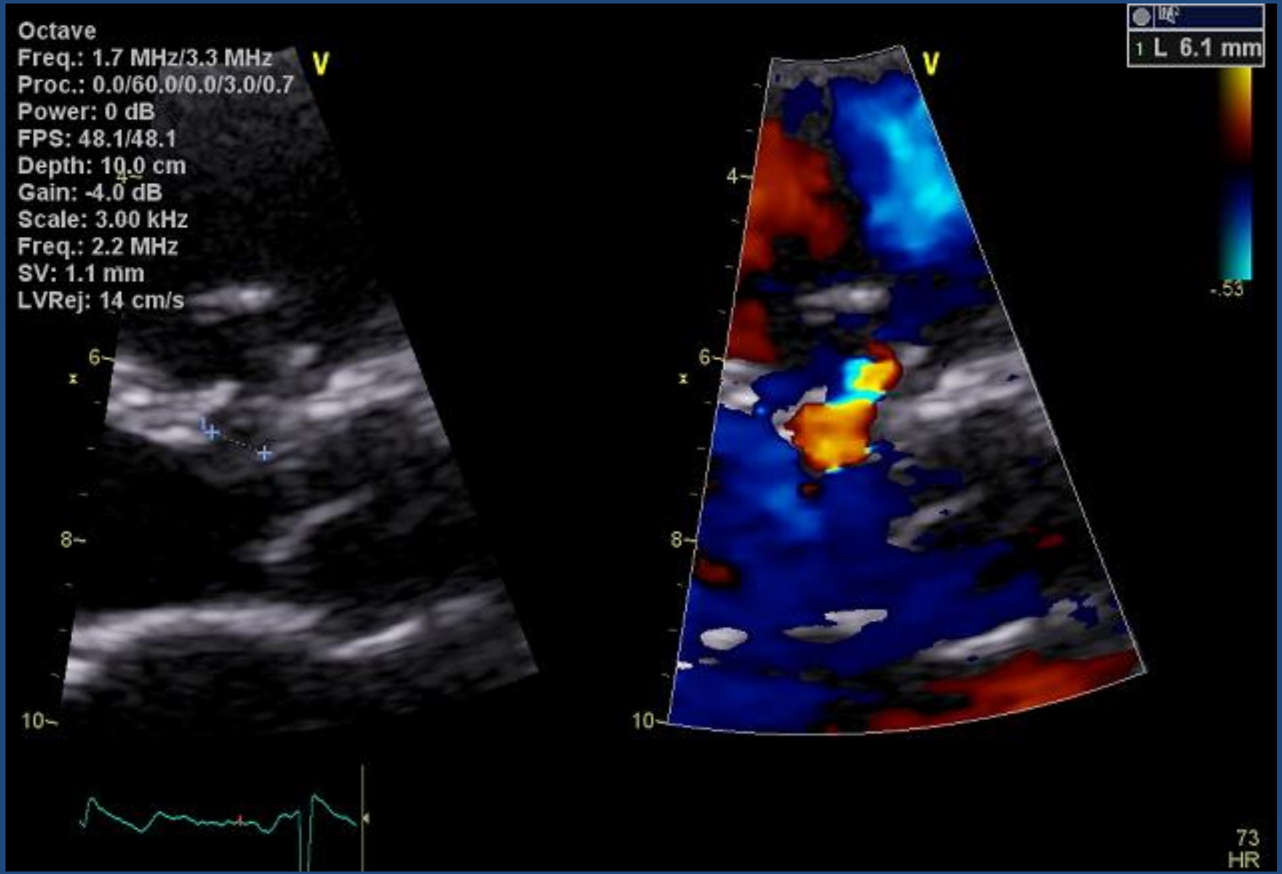
V
V
CTO
.53
-.53
80
HR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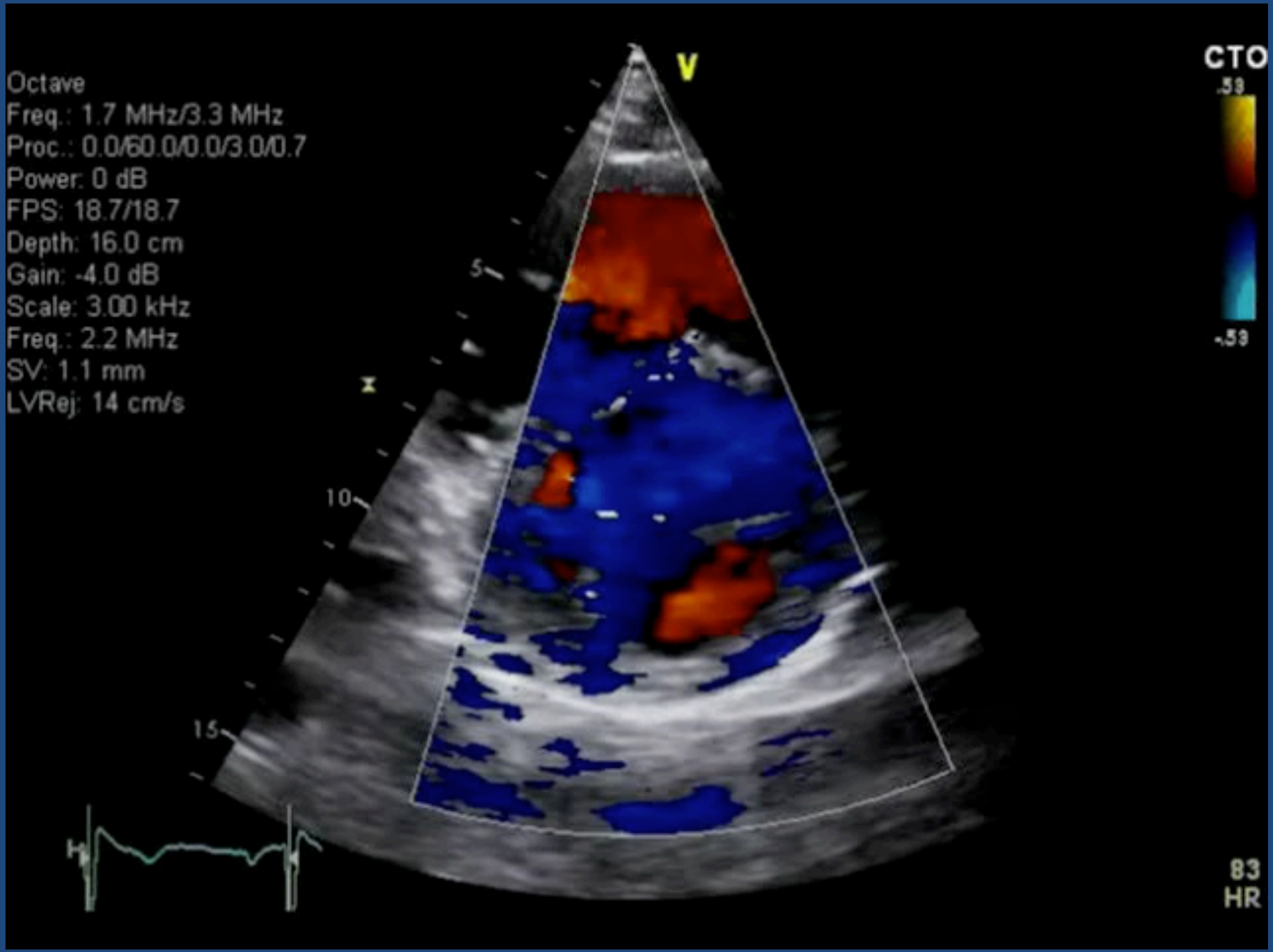
Gain: -4.0 dB
Scale: 3.00 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 14 cm/s
V
V
1 L 6.1 mm
-.53
4
6
8
10
73
HR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Gain: -4.0 dB
Scale: 3.00 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 14 cm/s
V
CTO
.59
-.59
5
10
15
83
HR

Octave
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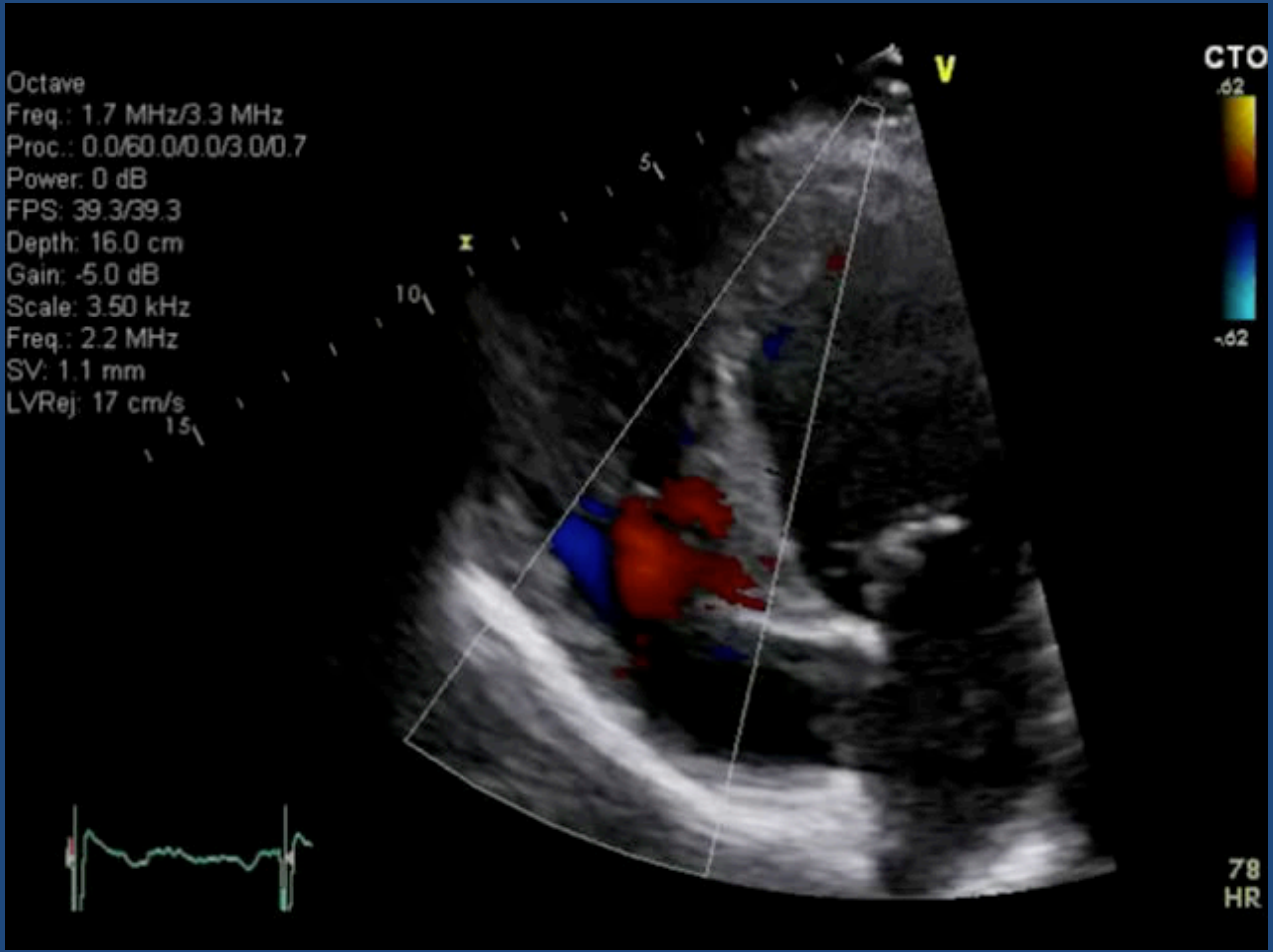
Proc.: 0.0/60.0/0.0/3.0/0.7
Power: 0 dB
FPS: 9.4/18.7
Depth: 13.0 cm
Gain: -4.0 dB
Scale: 3.00 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 14 cm/s
Scale: 893.0 cm/s
LVRej: 7.6 cm/s
Freq.: 2.0 MHz
V
1 v 708.92 cm/s
p 201 mmHg
-.53
8
6
4
2
[m/s]
-2.0
-1.5
-1.0
-0.5
0.0
100 mm/s
81
HR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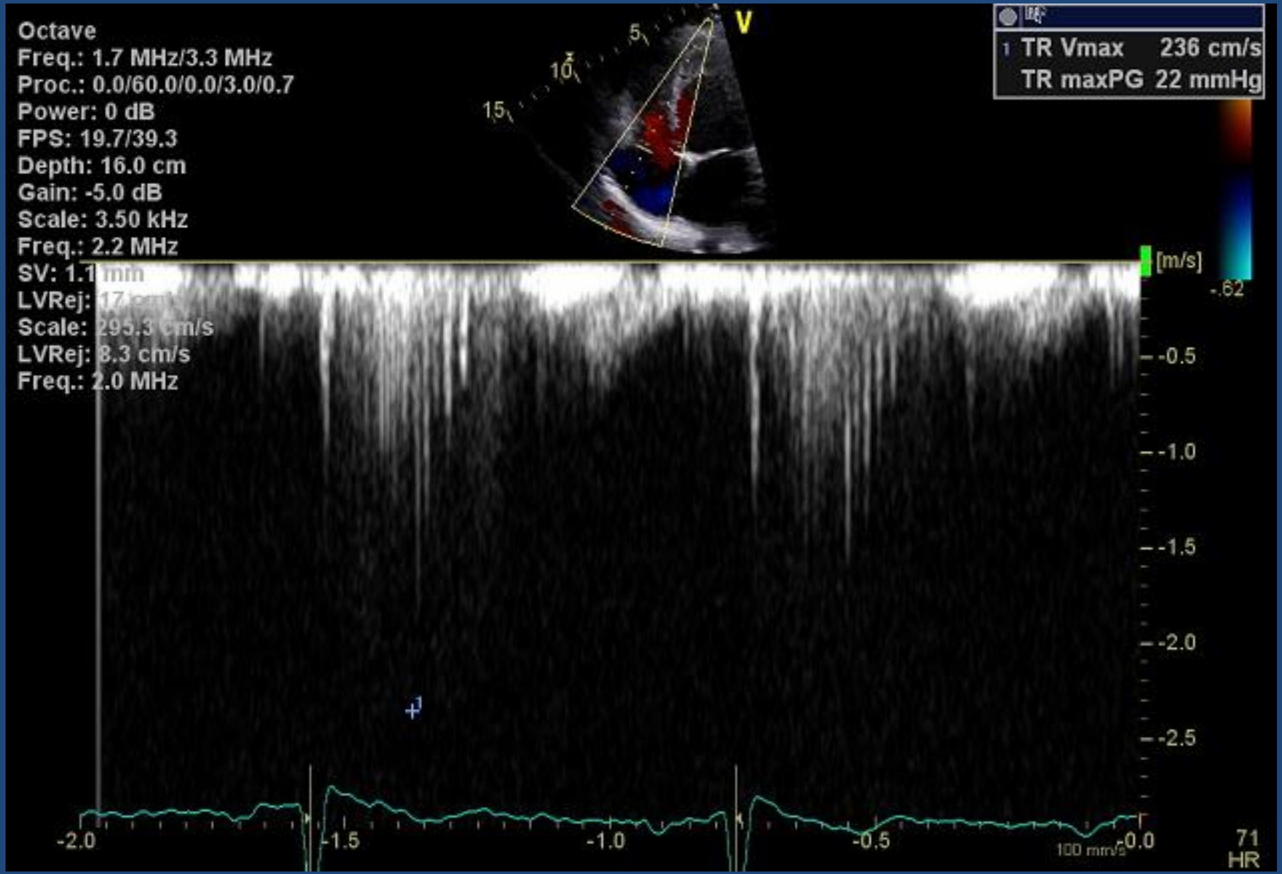
FPS: 39.3/39.3
Depth: 16.0 cm
Gain: -5.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 17 cm/s
CTO
.62
-.62
V
5
10
15
78
HR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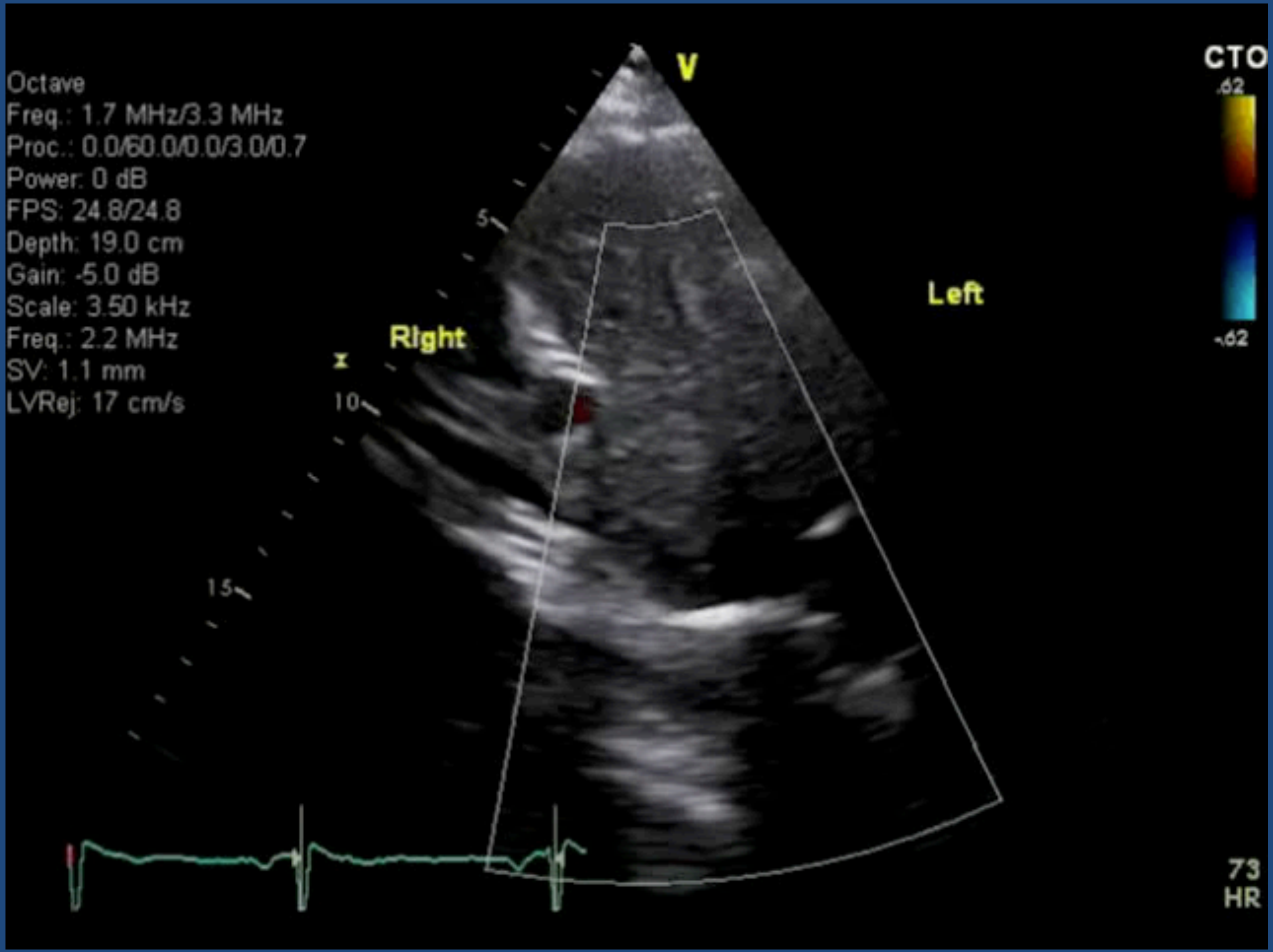
FPS: 19.7/39.3
Depth: 16.0 cm
Gain: -5.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 17 cm/s
Scale: 295.3 cm/s
LVRej: 8.3 cm/s
Freq.: 2.0 MHz
TR Vmax 236 cm/s
TR maxPG 22 mmHg
[m/s]
71
HR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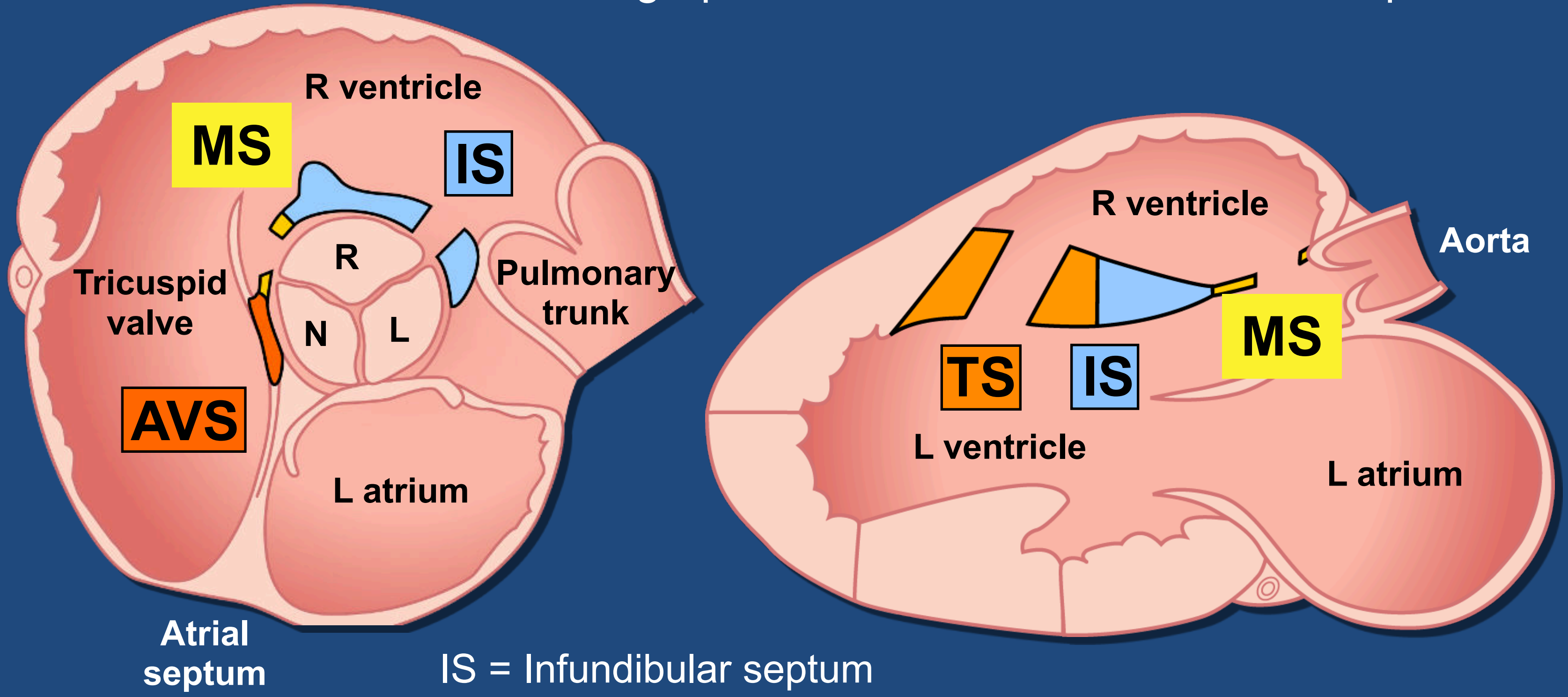
Depth: 19.0 cm
Gain: -5.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 17 cm/s
V
Right
Left
CTO
.62
-.62
5
10
15
73
HR

# What is the congenital abnormality?

# What is the congenital abnormality?

## Perimembranous VSD

Echocardiographic Examination of Ventricular Septal Defects




IS = Infundibular septum
MS = Membranous septum
AVS = Atrioventricular septum
TS = Trabecular/muscular septum

Ammash, N et al. *Ann Int Med* 2001; 135(9):818.

CP1233104-1

# What requires urgent attention?

# What requires urgent attention?

## Severe Hypertension; R/O preeclampsia

- TR jet = RV Pressure - RA Pressure

22 mmHg = RV Pressure – 3 mmHg

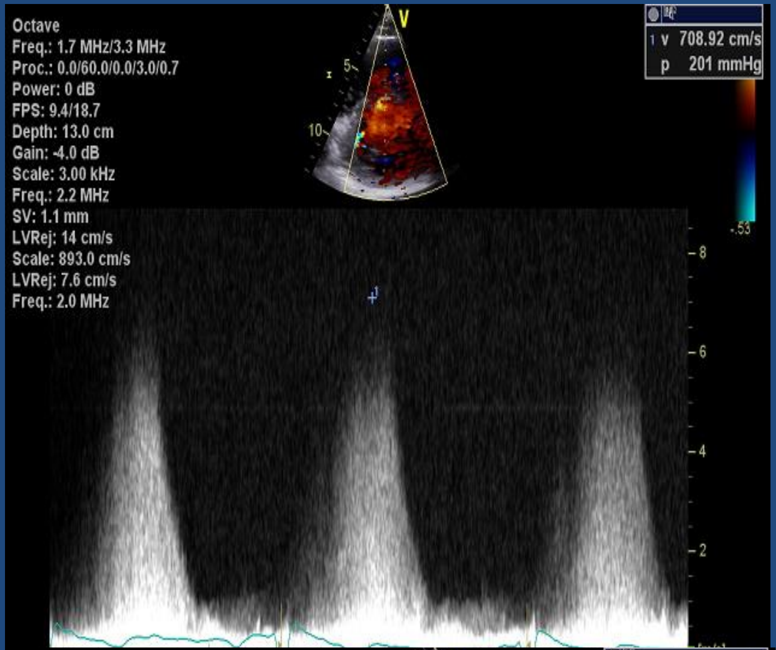
25 mmHg = RV Pressure

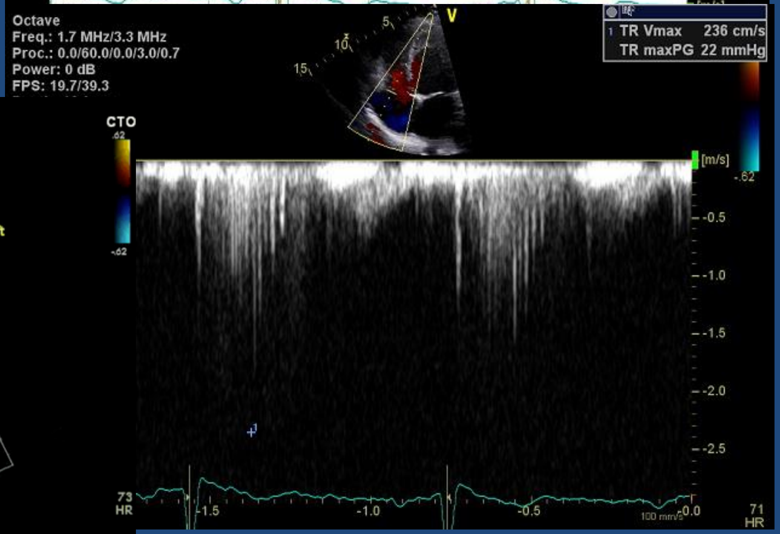
- VSD jet = LV Pressure – RV Pressure

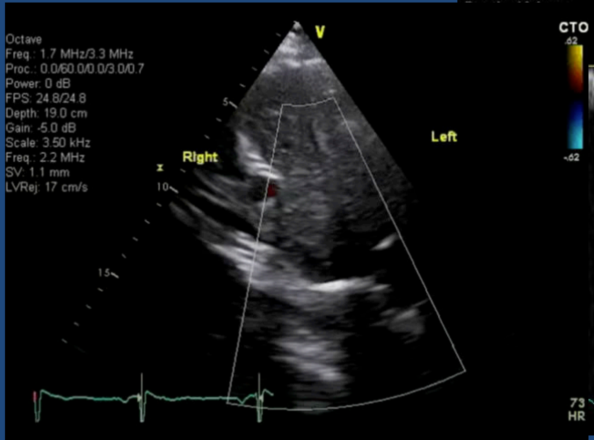
201 mmHg = LV Pressure – 25 mmHg

LV Pressure = 176 mmHg = SBP

**Patient has SEVERE HTN**

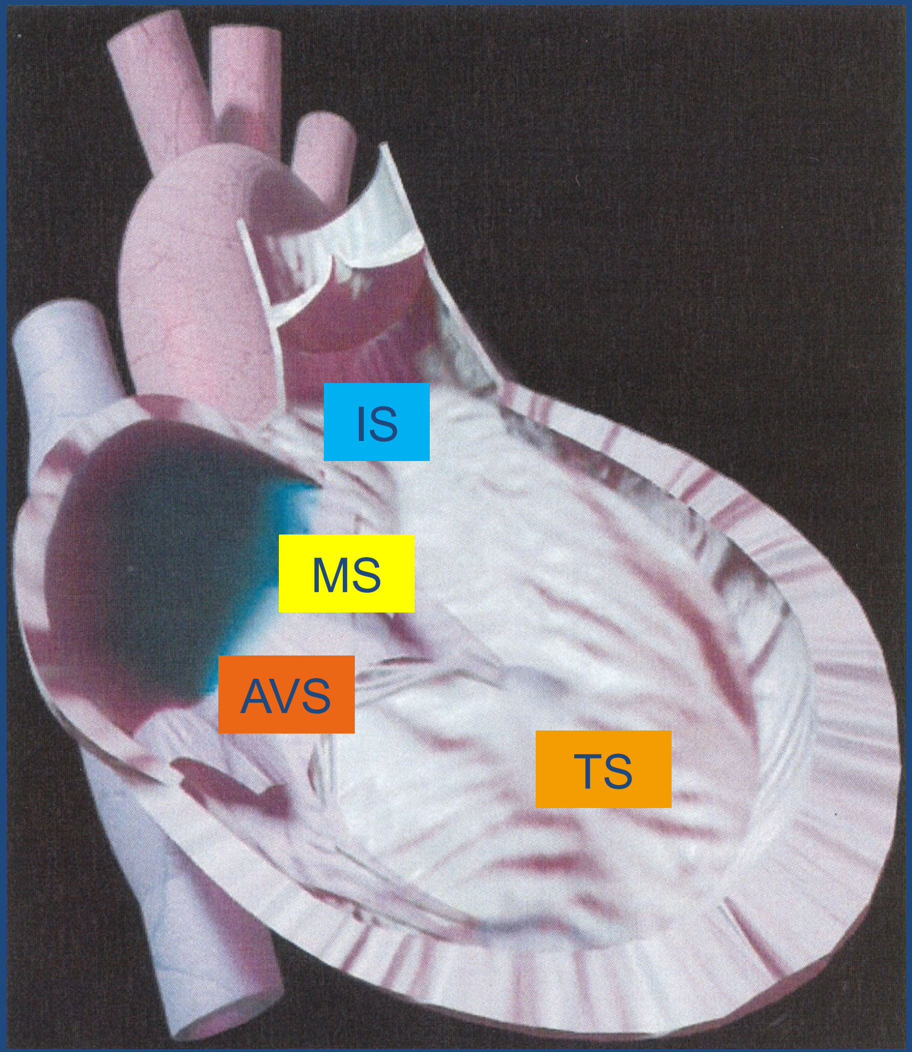


# Anatomy of the Ventricular Septum

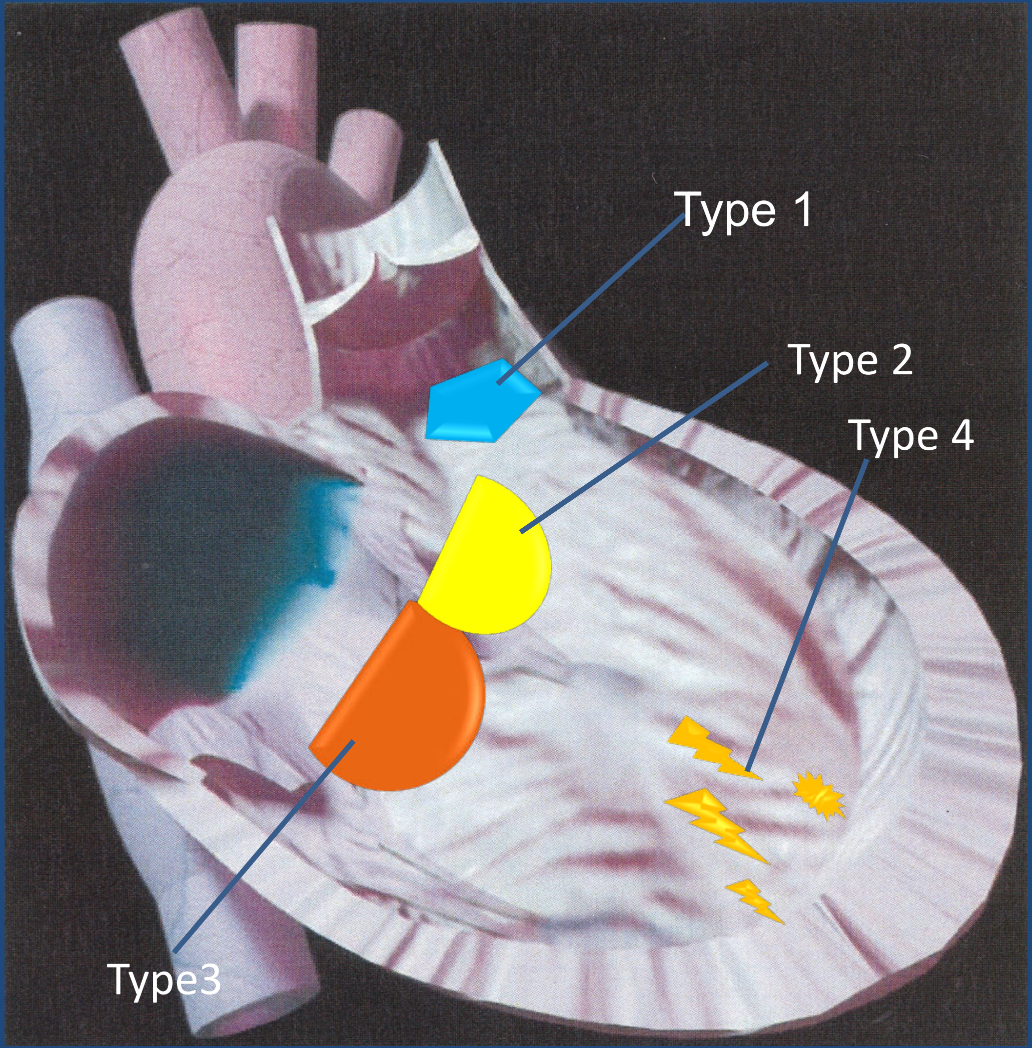
- Components:
  - Infundibular septum (IS)
  - Membranous septum (MS)
  - Atrioventricular/Inlet septum (AVS)
  - Trabecular/muscular septum (TS)




Jacobs, JP et al. Congenital Heart Disease Nomenclature and Database Project: Ventricular Septal Defect. *Ann Thorac Surg* 2000; 69: S25-35

# VSD Nomenclature

- **Type 1** **(6% non-Asian)** (33% Asian)
  - Subarterial
  - Supracristal
  - Subpulmonary
  - Infundibular
  - Conal
  - Outlet
  - Doubly committed
  - Juxta-arterial
- **Type 2** (80%)
  - Perimembranous
  - Paramembranous
  - Conoventricular
- **Type 3** (8%)
  - Inlet
  - AV canal type
- **Type 4** (5-20%)
  - Muscular




Modified from: Jacobs, JP et al. Congenital Heart Disease Nomenclature and Database Project: Ventricular Septal Defect. *Ann Thorac Surg* 2000; 69: S25-35

# Shunt Lesions: ASD, VSD, PDA, APVR

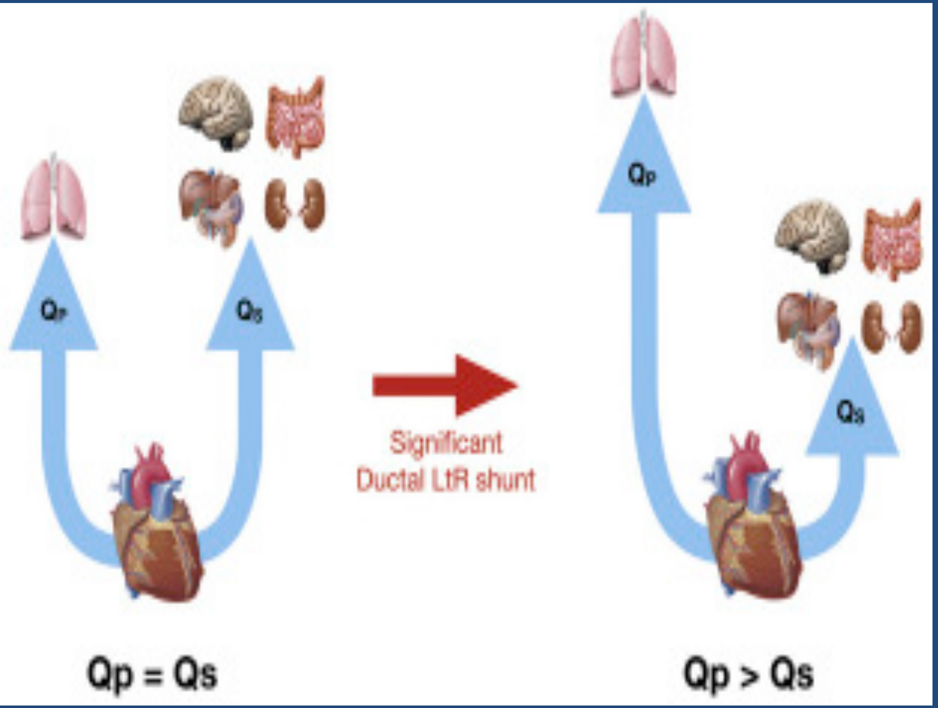
- Magnitude of (flow) the shunt related to:
  - Size of the shunt
  - Receiving cavity compliance
  - Pulmonary and systemic vascular resistances

- A shunt creates a volume burden on the receiving chamber, which if hemodynamically significant, will result in volume overload and chamber dilatation.
  - ASD: RA + RV dilatation
  - Membranous VSD: LA + LV dilatation
  - Muscular VSD: RV + LA + LV dilatation

# Shunt Lesions

- Restrictive shunt = not hemodynamically significant
  - QP:QS < 1.5: 1
  - No chamber enlargement
  - No arrhythmias
  - Normal exercise capacity
  - Normal pulmonary pressures

- Unrestrictive shunt = hemodynamically significant
  - QP:QS >1.5:1
  - Chamber dilatation → HF
  - Arrhythmias (stretch conduction system)
  - Reduced exercise capacity
  - Pulmonary HTN (develops by age 2 except with ASD)

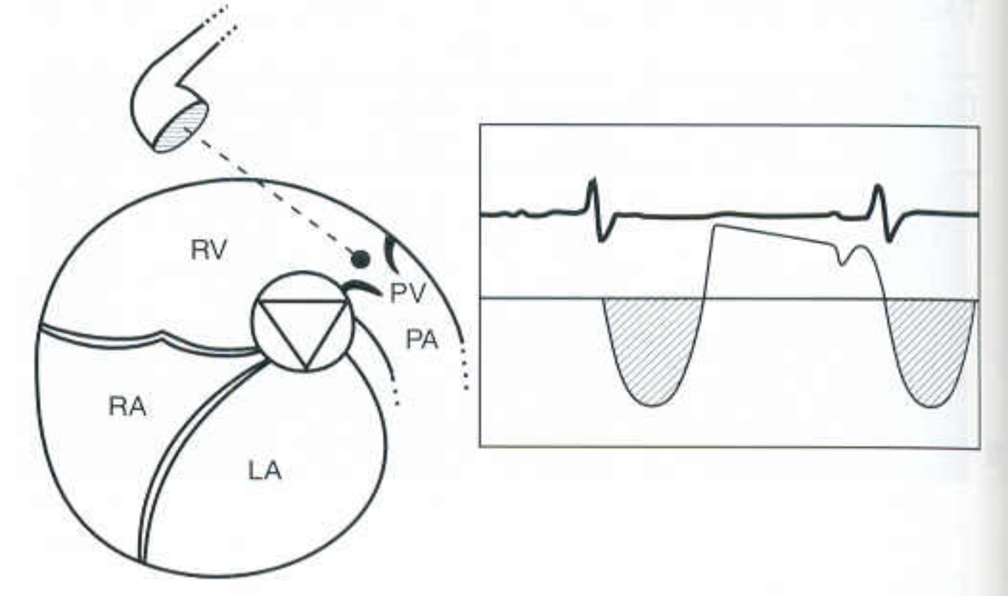


# Qp/Qs

- Qp = pulmonary flow
- Qs = systemic flow

# Qp/Qs by Echo

$$\frac{Qp}{Qs} = \frac{\text{RVOT CSA X RVOT TVI}}{\text{LVOT CSA X LVOT TVI}}$$



# Calculate Qp/Qs in Cath Lab

| | |
|---|---|
| High SVC | 67% |
| Low SVC | 67% |
| High RA | 69% |
| Mid RA | 68% |
| Low RA | 70% |
| High IVC | 71% |
| Low IVC | 71% |
| RV inflow | 80% |
| Mid RV | 92% |
| RV outflow | 88% |
| PA | 89% |
| Aorta | 99% |

- Where is the shunt?
- What is the Qp/Qs?

# Calculate Qp/Qs in Cath Lab

$$\frac{Qp}{Qs} = \frac{\%\ \text{sat Ao} - \%\ \text{sat MV}}{\%\ \text{sat PV} - \%\ \text{sat PA}} = \frac{98 - [(3 \times 67) + 71]\ /\ 4}{99 - 89}$$

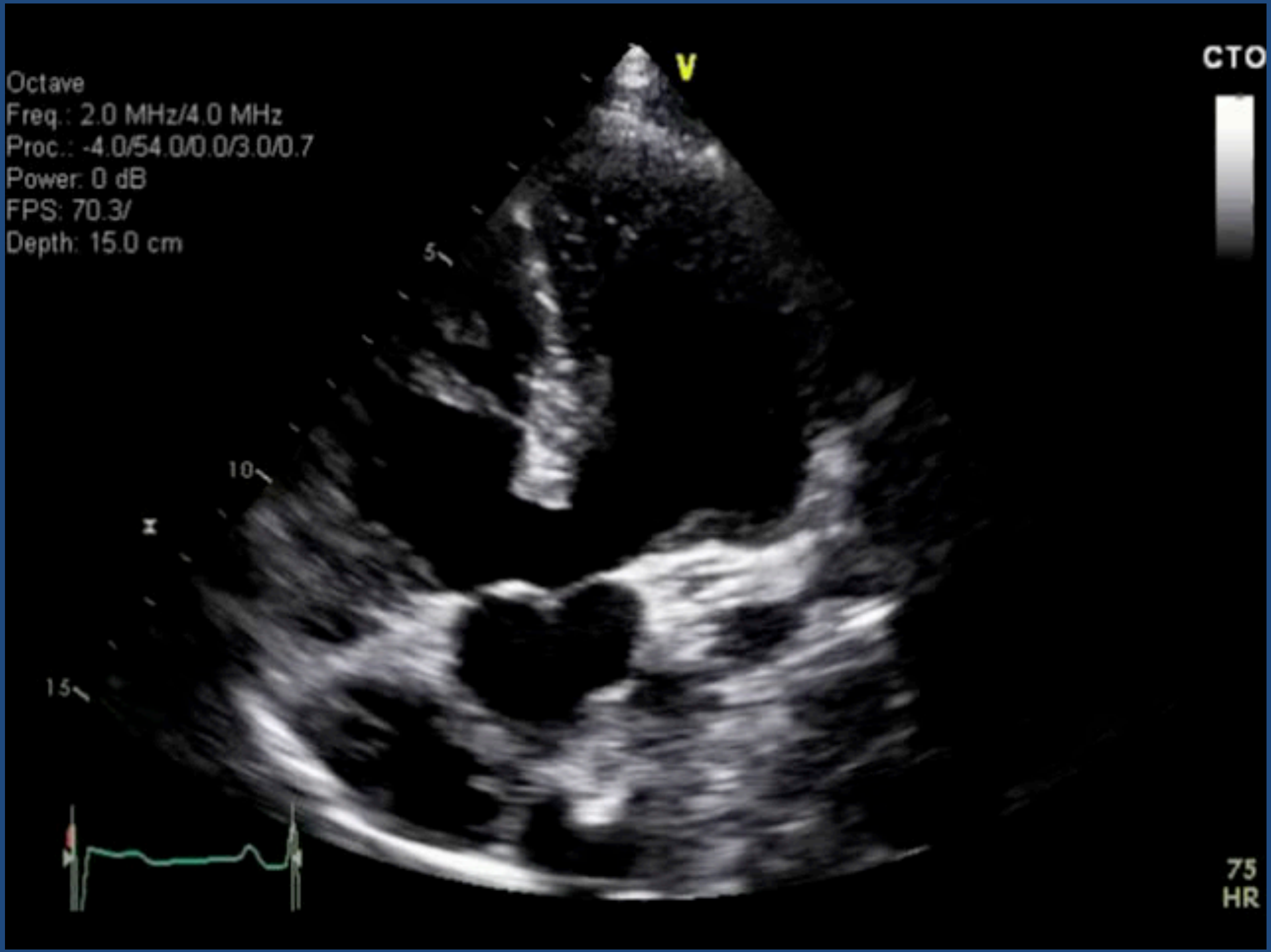
$$= 30/10$$

$$= 3/1$$

# ToF

# Case #2

- Look at this echo. There are 5 clips.
- What is the diagnosis?


Octave
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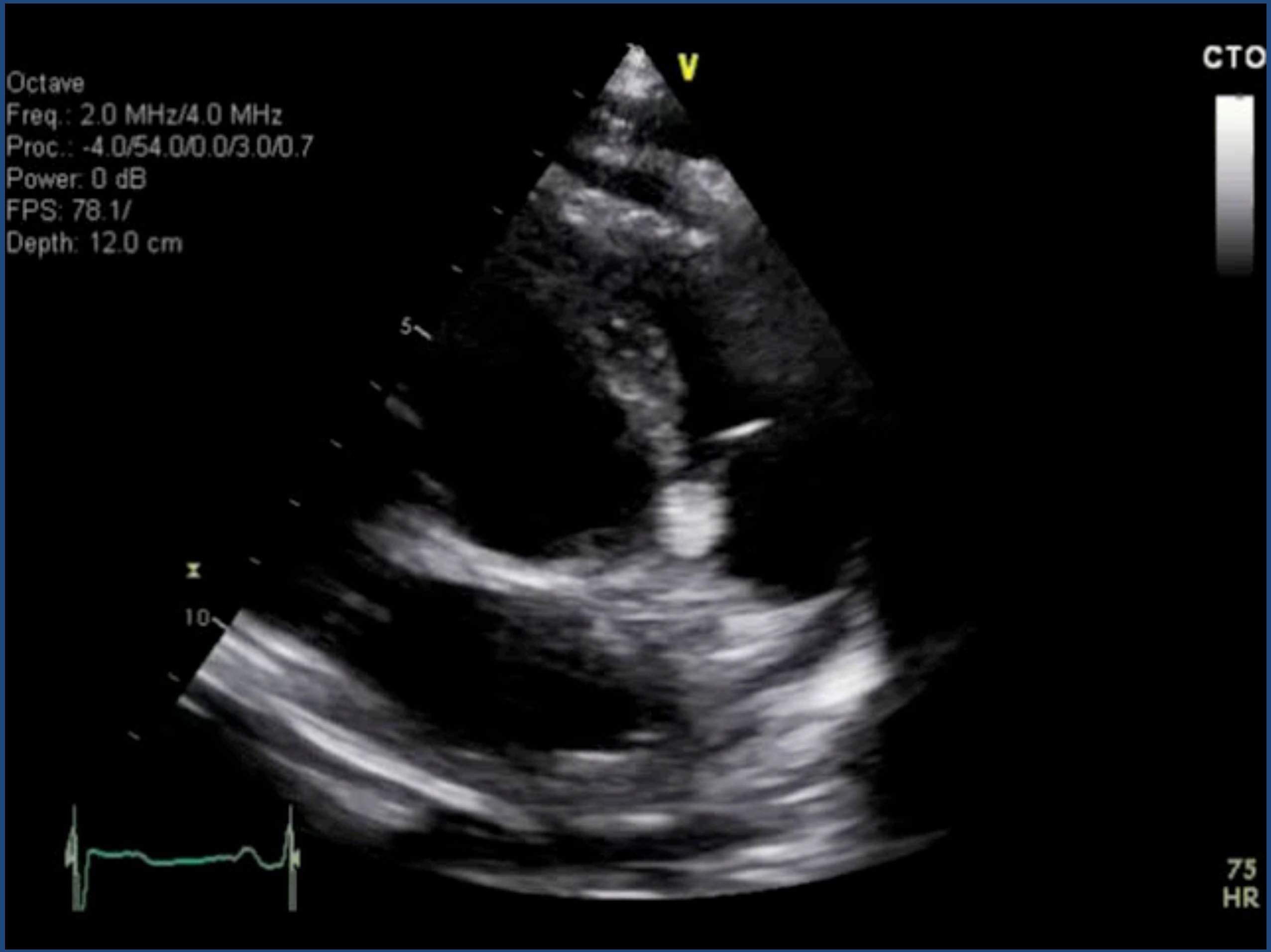
Power: 0 dB
FPS: 70.3/
Depth: 15.0 cm
V
CTO
5
10
15
75
HR



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Proc.: -4.0/54.0/0.0/3.0/0.7
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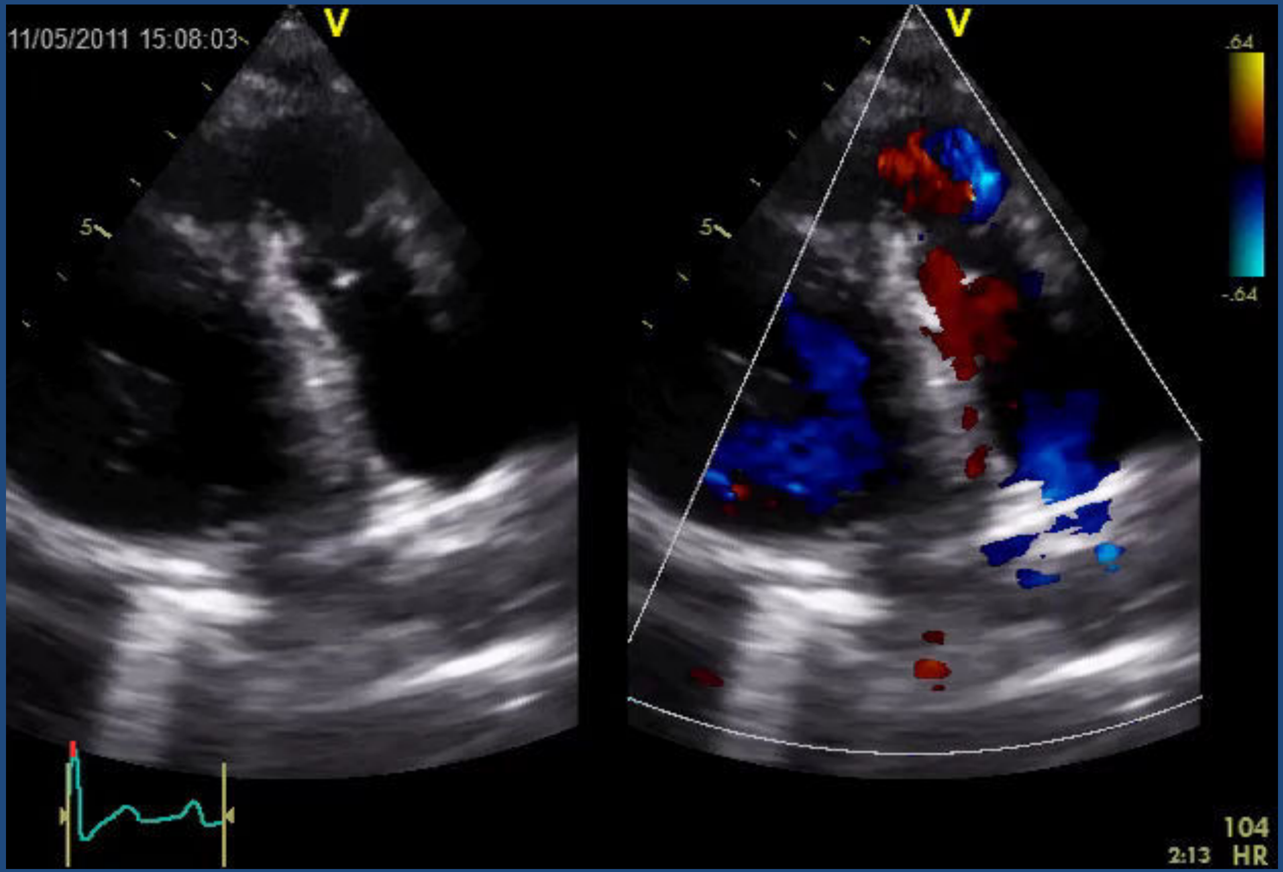
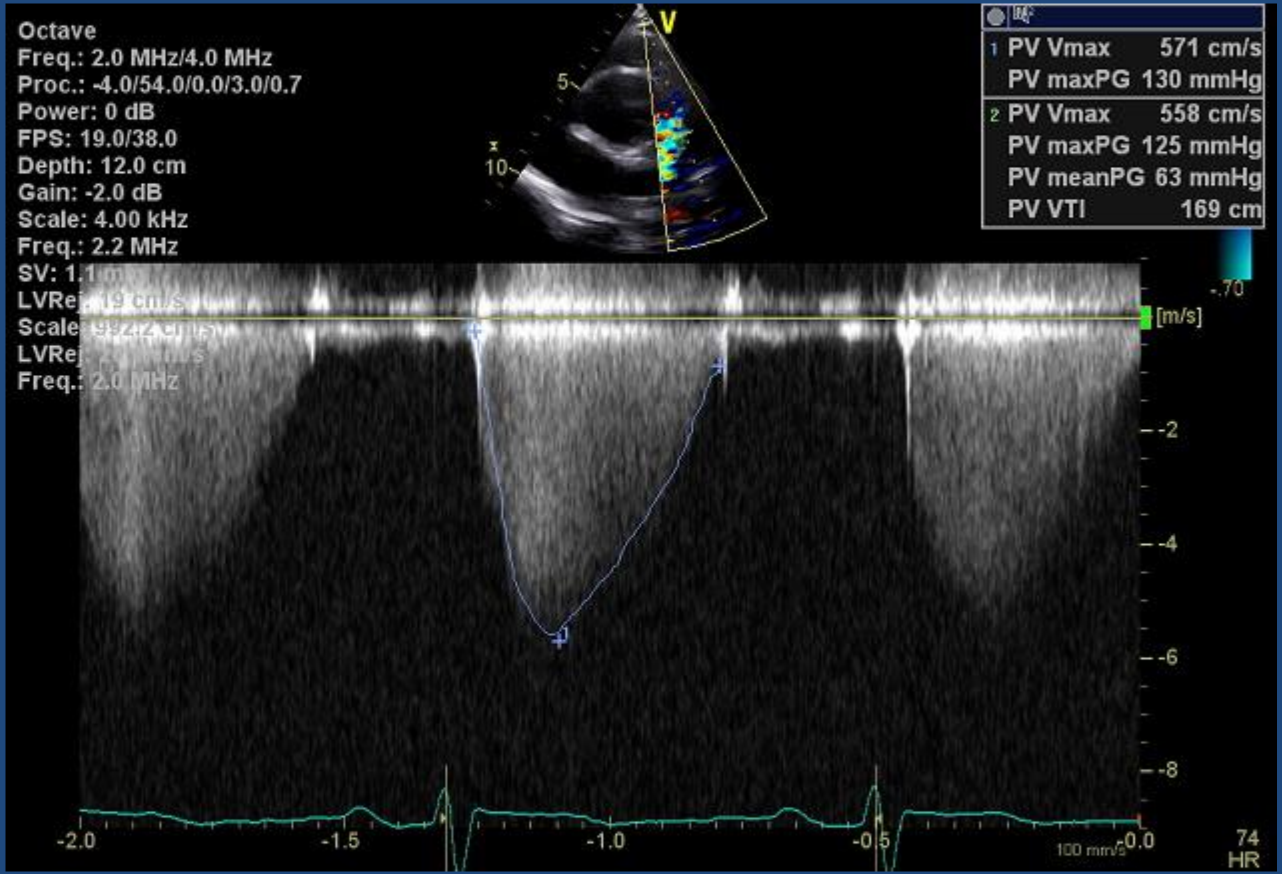
FPS: 78.1/
Depth: 12.0 cm
V
CTO
5
10
75
HR


11/05/2011 15:08:03
V
5
V
5
.64
-.64
104
2:13
HR



Octave
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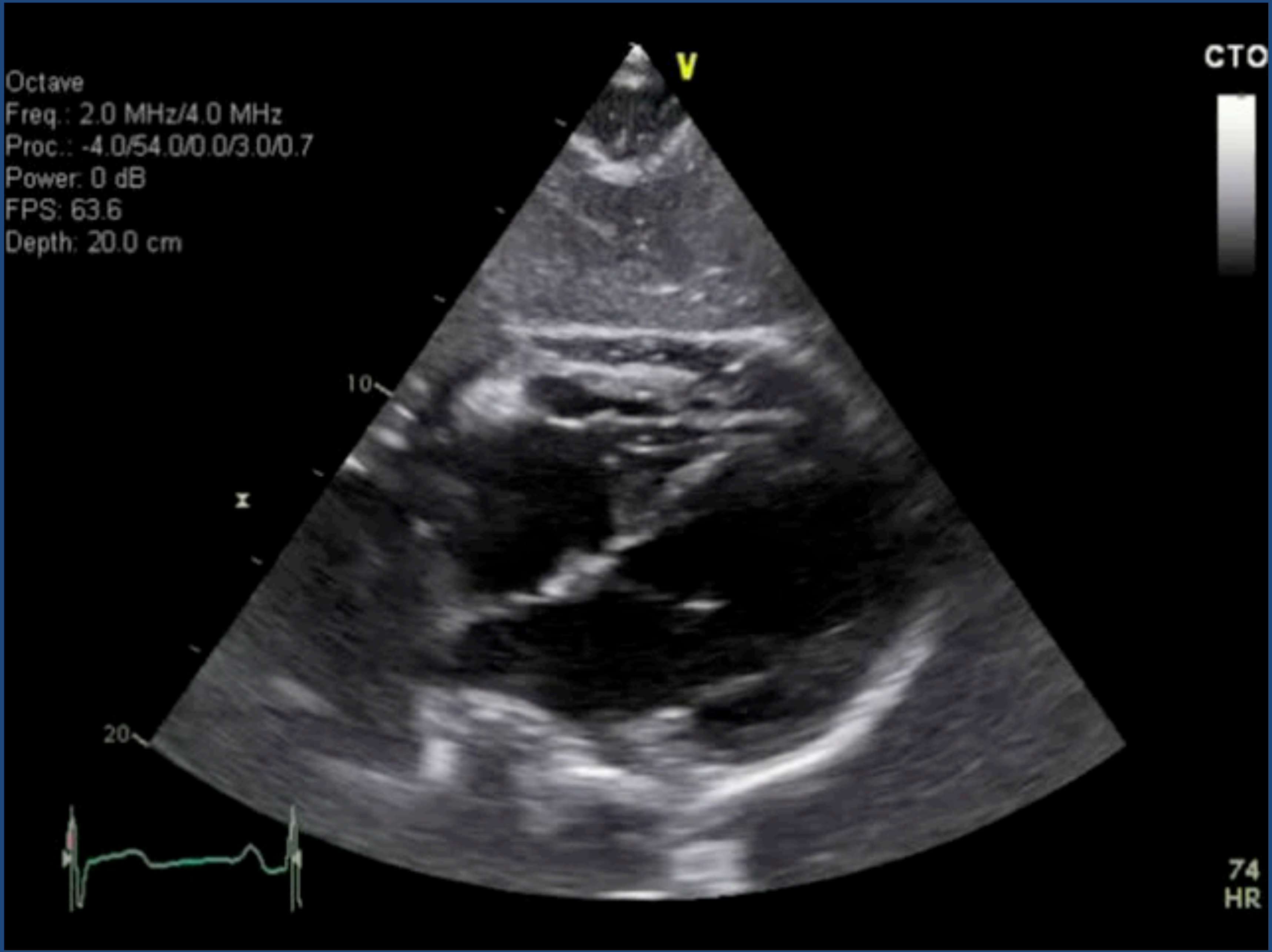
Proc.: -4.0/54.0/0.0/3.0/0.7
Power: 0 dB
FPS: 19.0/38.0
Depth: 12.0 cm
Gain: -2.0 dB
Scale: 4.00 kHz
Freq.: 2.2 MHz
1 PV Vmax 571 cm/s
PV maxPG 130 mmHg
2 PV Vmax 558 cm/s
PV maxPG 125 mmHg
PV meanPG 63 mmHg
PV VTI 169 cm
[m/s]
74
HR


Octave
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Proc.: -4.0/54.0/0.0/3.0/0.7
Power: 0 dB
FPS: 63.6
Depth: 20.0 cm
V
CTO
10
20
74
HR

# What is the diagnosis?

Tetralogy of Fallot

- VSD
- Overriding aorta
- RVOT obstruction (sub PS +/- PS)
- RVH

What is Eisenmenger Syndrome?

Would you expect this 32 y.o. patient with unrepaired ToF to have Eisenmenger syndrome? Explain.

# What is Eisenmenger Syndrome?

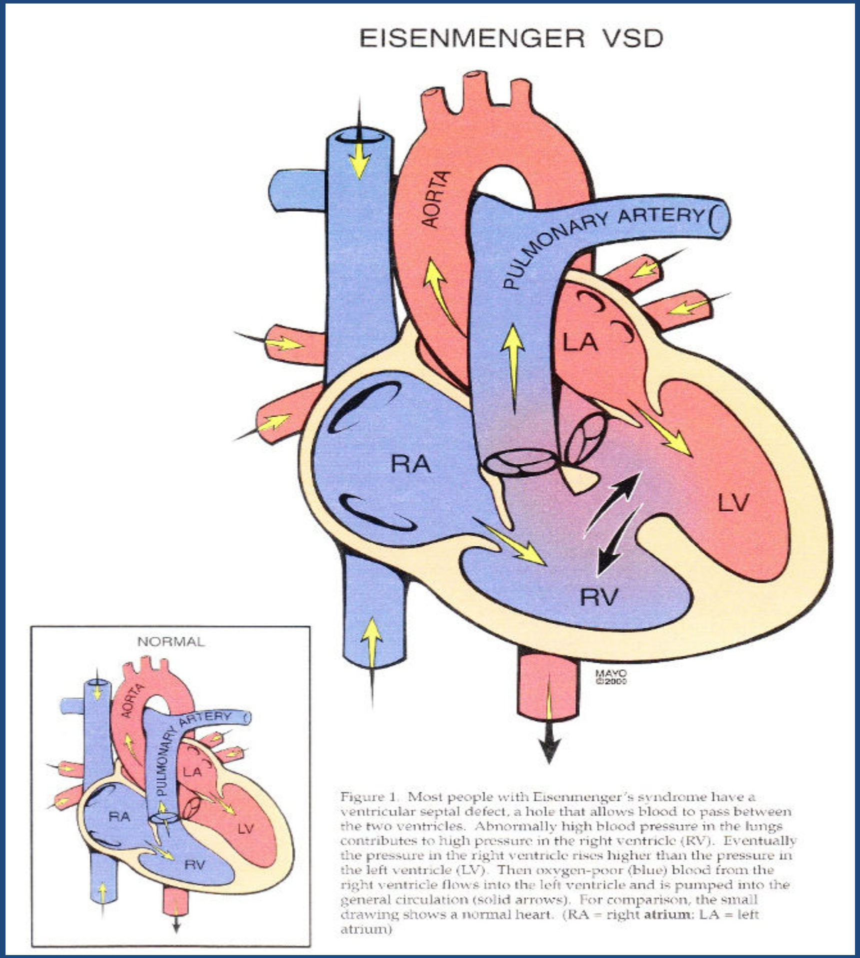



Figure 1. Most people with Eisenmenger's syndrome have a ventricular septal defect, a hole that allows blood to pass between the two ventricles. Abnormally high blood pressure in the lungs contributes to high pressure in the right ventricle (RV). Eventually the pressure in the right ventricle rises higher than the pressure in the left ventricle (LV). Then oxygen-poor (blue) blood from the right ventricle flows into the left ventricle and is pumped into the general circulation (solid arrows). For comparison, the small drawing shows a normal heart. (RA = right **atrium**; LA = left atrium)

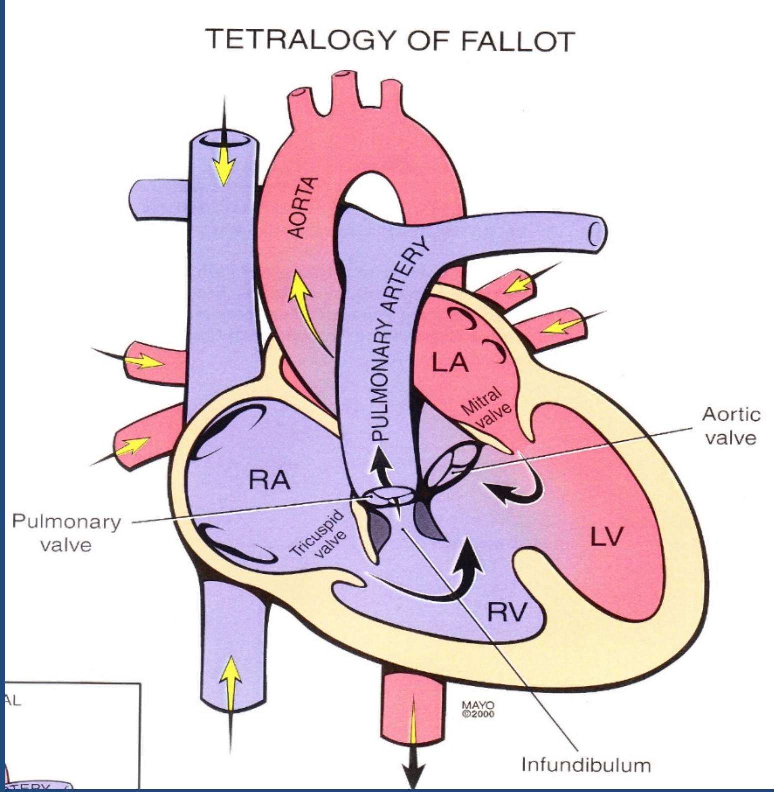
Mayo Foundation for Medical Education and Research (MFMER). Eisenmenger VSD from
Mayo Clinic Patient Education Booklet on Eisenmenger's Syndrome
Adult Congenital Heart Disease. Page 2, 2000.

*Eisenmenger Syndrome*:

Pulmonary vascular obstructive disease that develops as a result of a LARGE, uncorrected, longstanding acyanotic L→R shunt that causes irreversible remodeling of the pulmonary arteriolar vascular bed (pulmonary arteriolar media hypertrophy and intimal proliferation) such that the pulmonary arterial pressure (PAP) and pulmonary vascular resistance (PVR) become ≥ 2/3 systemic arterial pressure (SAP) and ≥ 2/3 systemic vascular resistance (SVR) which causes the shunt to become bidirectional and then reverse (R→L) resulting in cyanosis.

# Would you expect this 32 y.o. patient with unrepaired ToF to have Eisenmenger physiology? Explain.

- NO. The RVOT obstruction protects against pulmonary overcirculation. Therefore, PHTN does not occur in patients with native ToF.




Mayo Foundation for Medical Education and Research (MFMER). Tetralogy of Fallot from Mayo Clinic Patient Education Booklet on Tetralogy of Fallot Adult Congenital Heart Disease. Page 2, 2000.

# Name three physical exam findings for this patient with unrepaired ToF?

- Cyanosis
- Clubbing
- Low O2 saturation
- Sustained RV
- Systolic murmur due to RVOT obstruction

# ToF Surgical Repair

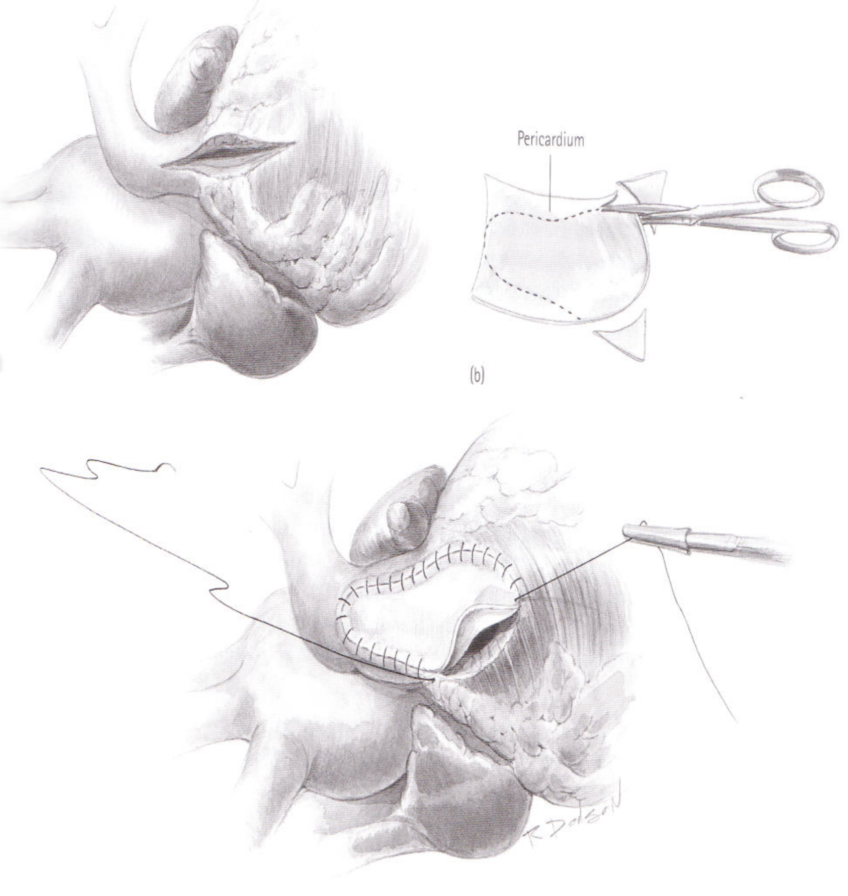
- ToF repair usual done by 6-12 months

- Repair involves:
  - VSD closure
  - Relieve the RVOT obstruction
  - Repair associated defects

# ToF Surgical Repair

## Before 1990's

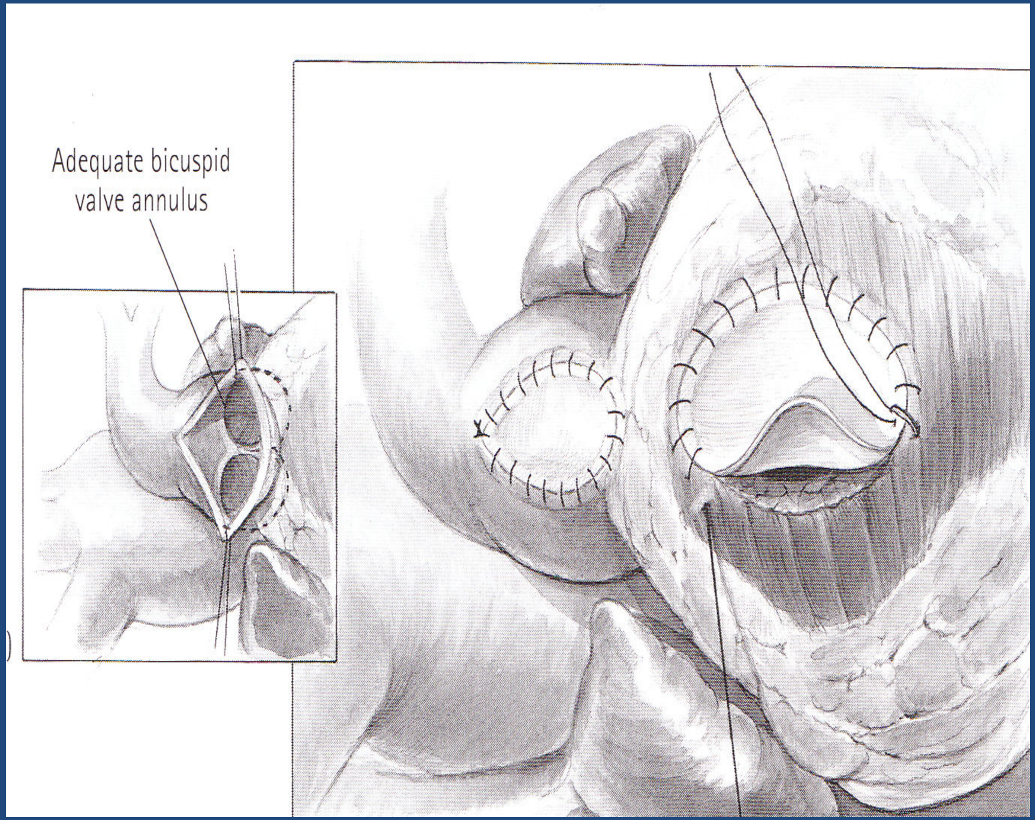
- Right ventriculotomy approach (↑ incision)
- Generous transannular patch augmentation
- Severe free PR: complications in adults
  - RV dilatation
  - ↓RV function
  - Arrhythmias
  - SCD




Jonas, R.A. (2004) . *Comprehensive Surgical Management of Congenital Heart Disease*. London : Arnold.

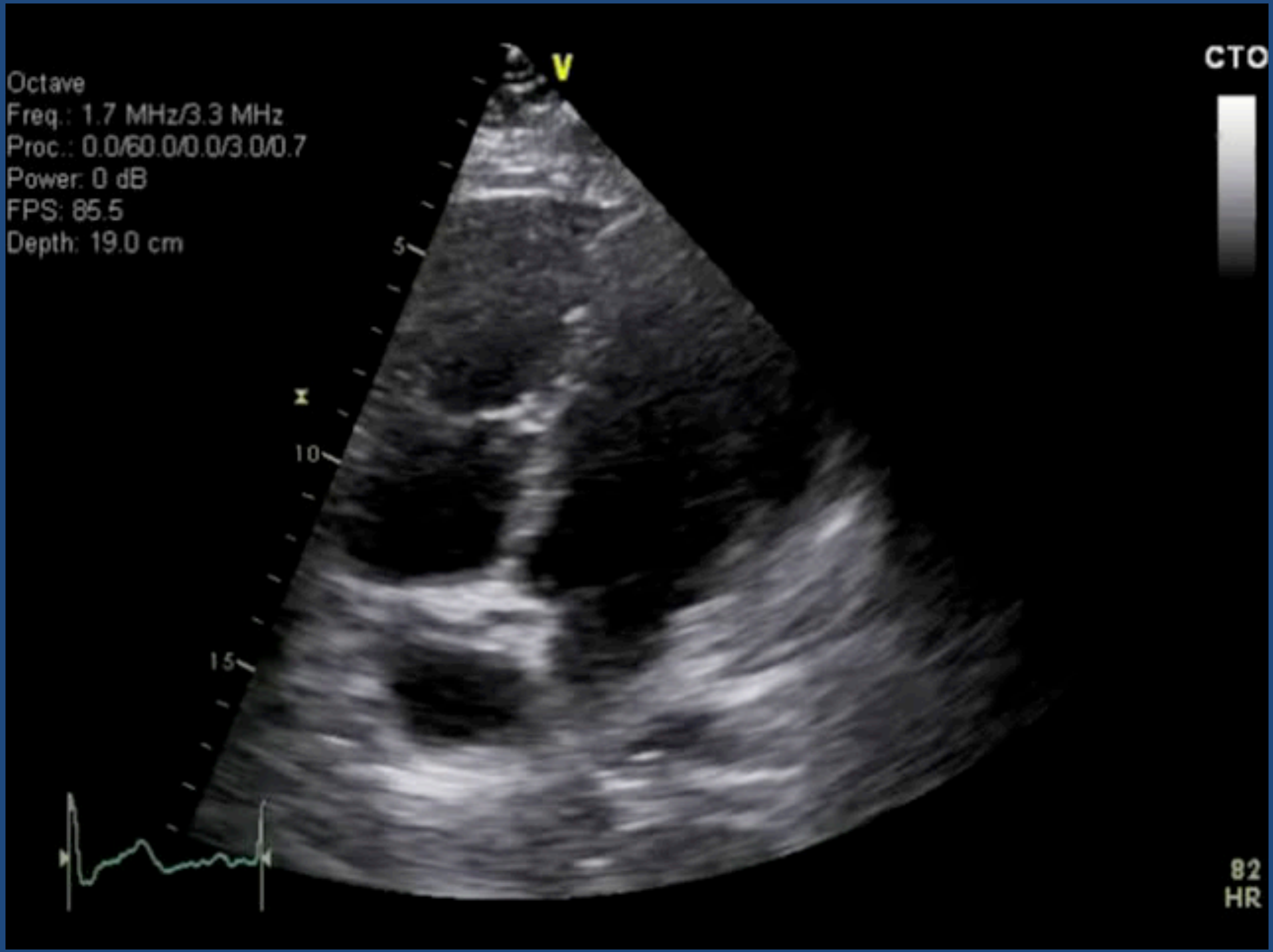
# ToF Surgical Repair
## Today

- Transatrial/transpulmonary approach
- Limited RV incision for patch augmentation of the RVOT +/- PV annulus
- Goal: Avoid PR at the expense of some residual PS




Jonas, R.A. (2004) . *Comprehensive Surgical Management of Congenital Heart Disease*. London : Arnold.

# TOF S/P TAP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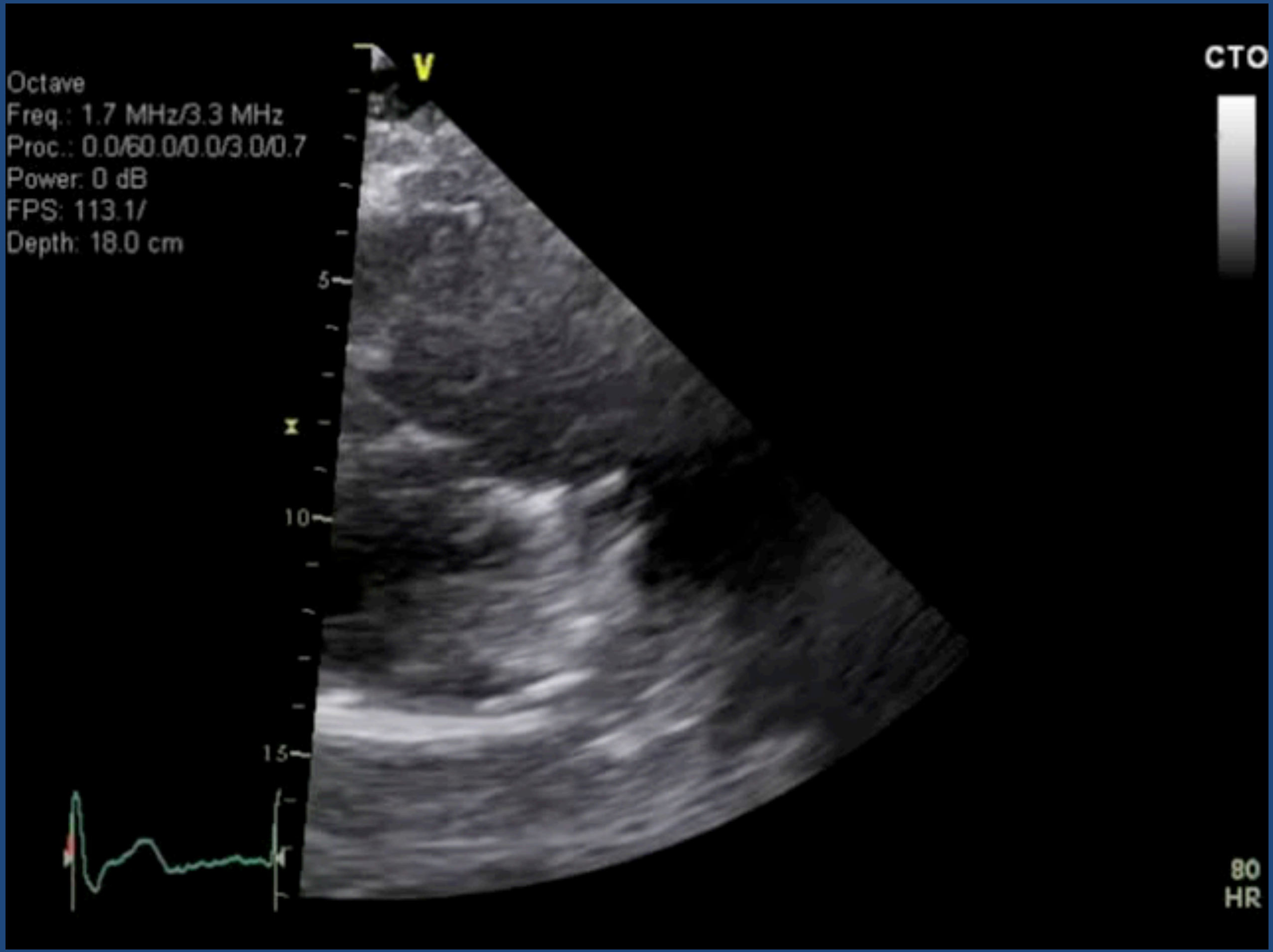
FPS: 85.5
Depth: 19.0 cm
V
CTO
5
10
15
82
HR

# TOF S/P TAP


Octave
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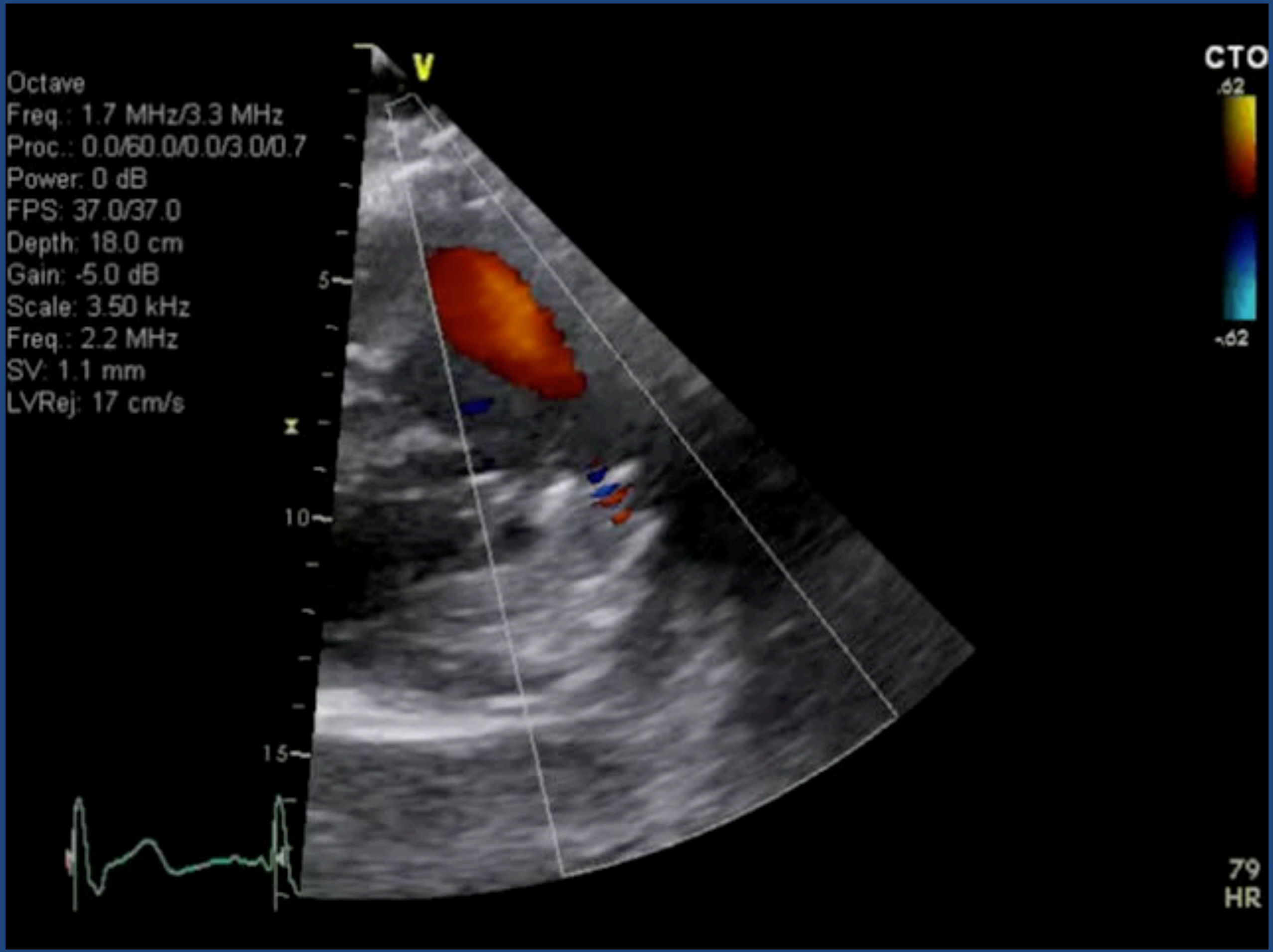
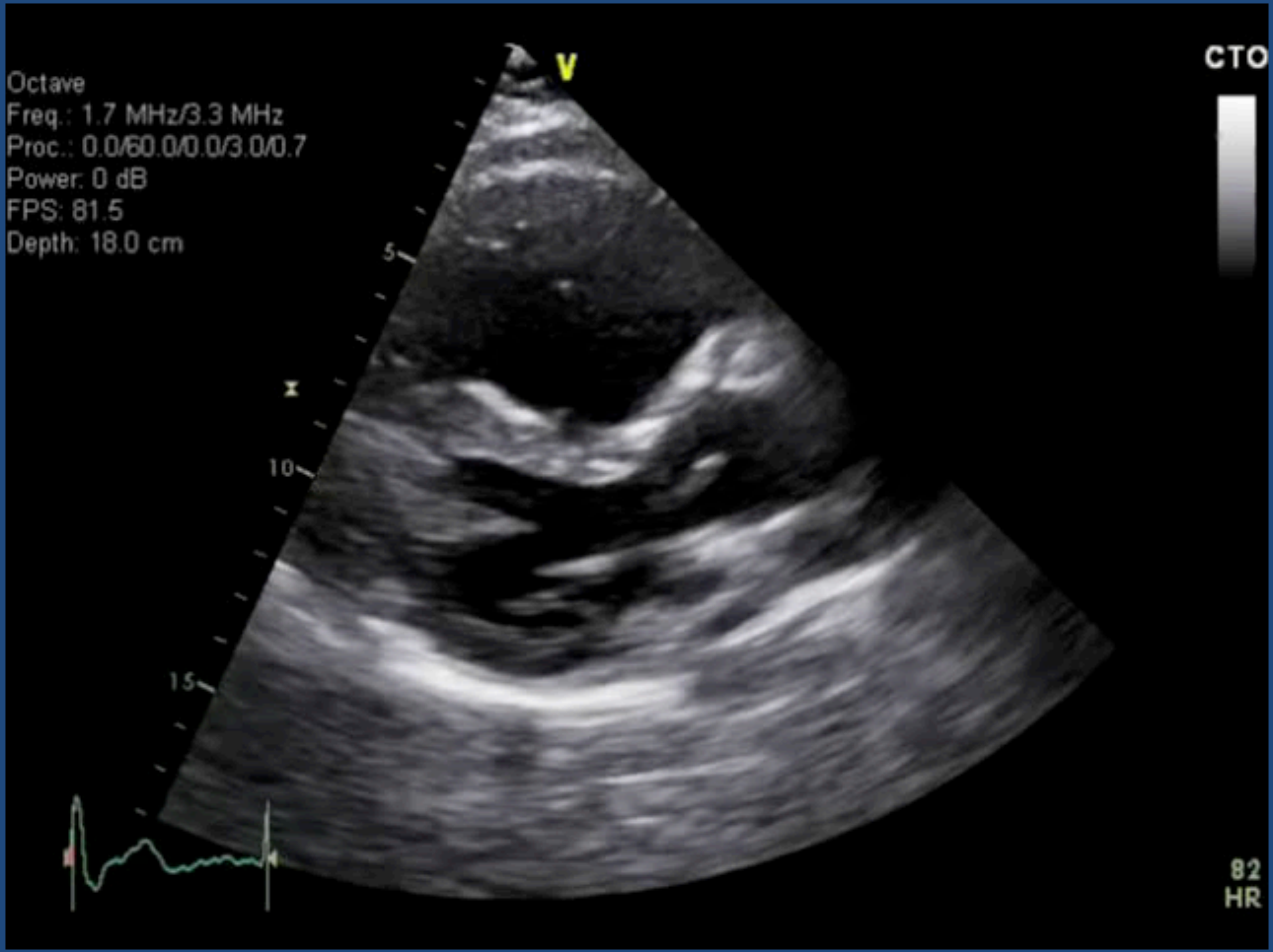
Proc.: 0.0/60.0/0.0/3.0/0.7
Power: 0 dB
FPS: 113.1/
Depth: 18.0 cm
V
CTO
5
10
15
80
HR

# TOF S/P TAP

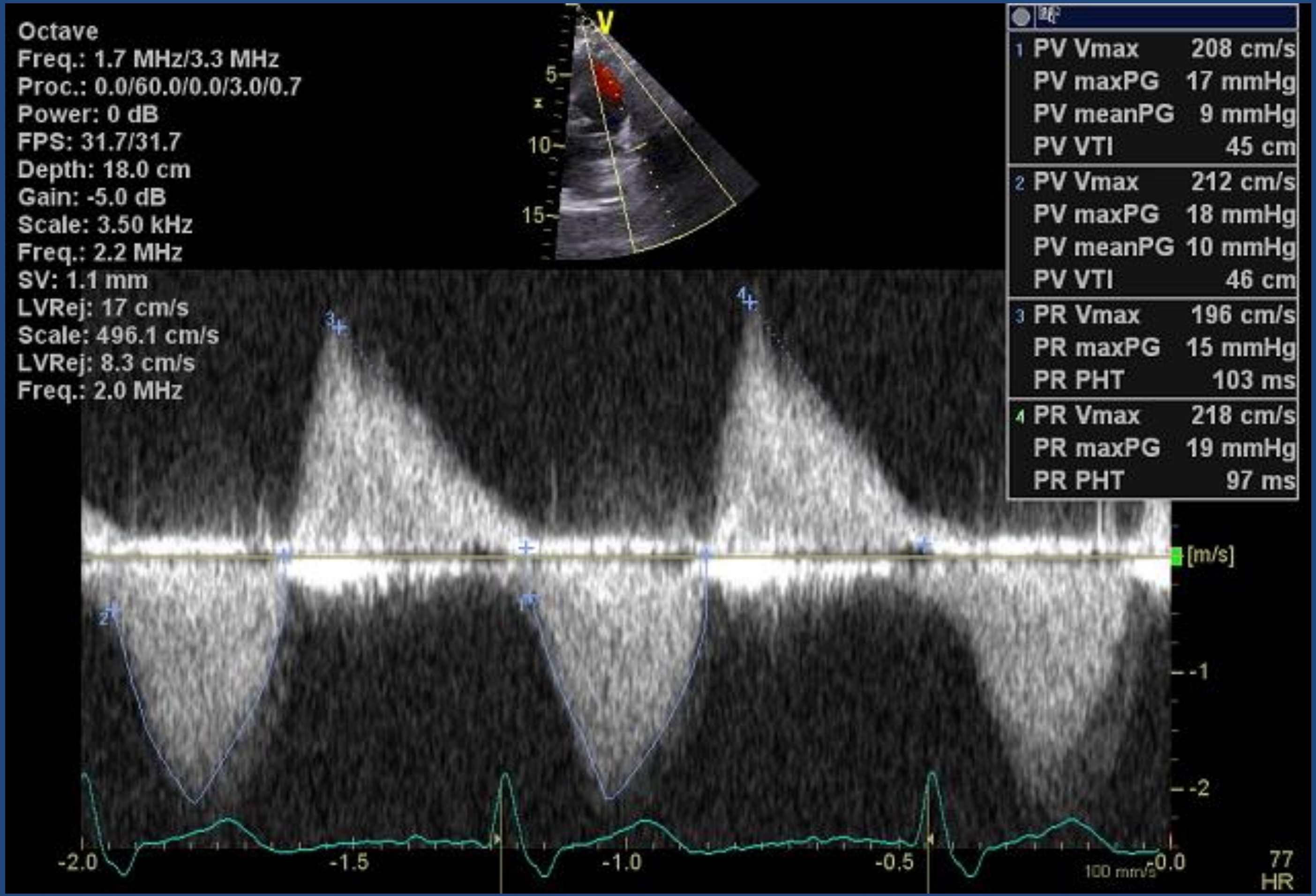

Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: 0.0/60.0/0.0/3.0/0.7
Power: 0 dB
FPS: 37.0/37.0
Depth: 18.0 cm
Gain: -5.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 17 cm/s
V
CTO
.62
-.62
5
10
15
79
HR

# TOF S/P TAP



Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: 0.0/60.0/0.0/3.0/0.7
Power: 0 dB
FPS: 81.5
Depth: 18.0 cm
V
CTO
5
10
15
82
HR

# TOF S/P TAP


Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: 0.0/60.0/0.0/3.0/0.7
Power: 0 dB
FPS: 31.7/31.7
Depth: 18.0 cm
Gain: -5.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 17 cm/s
Scale: 496.1 cm/s
LVRej: 8.3 cm/s
Freq.: 2.0 MHz
1 PV Vmax 208 cm/s
PV maxPG 17 mmHg
PV meanPG 9 mmHg
PV VTI 45 cm
2 PV Vmax 212 cm/s
PV maxPG 18 mmHg
PV meanPG 10 mmHg
PV VTI 46 cm
3 PR Vmax 196 cm/s
PR maxPG 15 mmHg
PR PHT 103 ms
4 PR Vmax 218 cm/s
PR maxPG 19 mmHg
PR PHT 97 ms
[m/s]
-1
-2
-2.0
-1.5
-1.0
-0.5
0.0
100 mm/s
77
HR

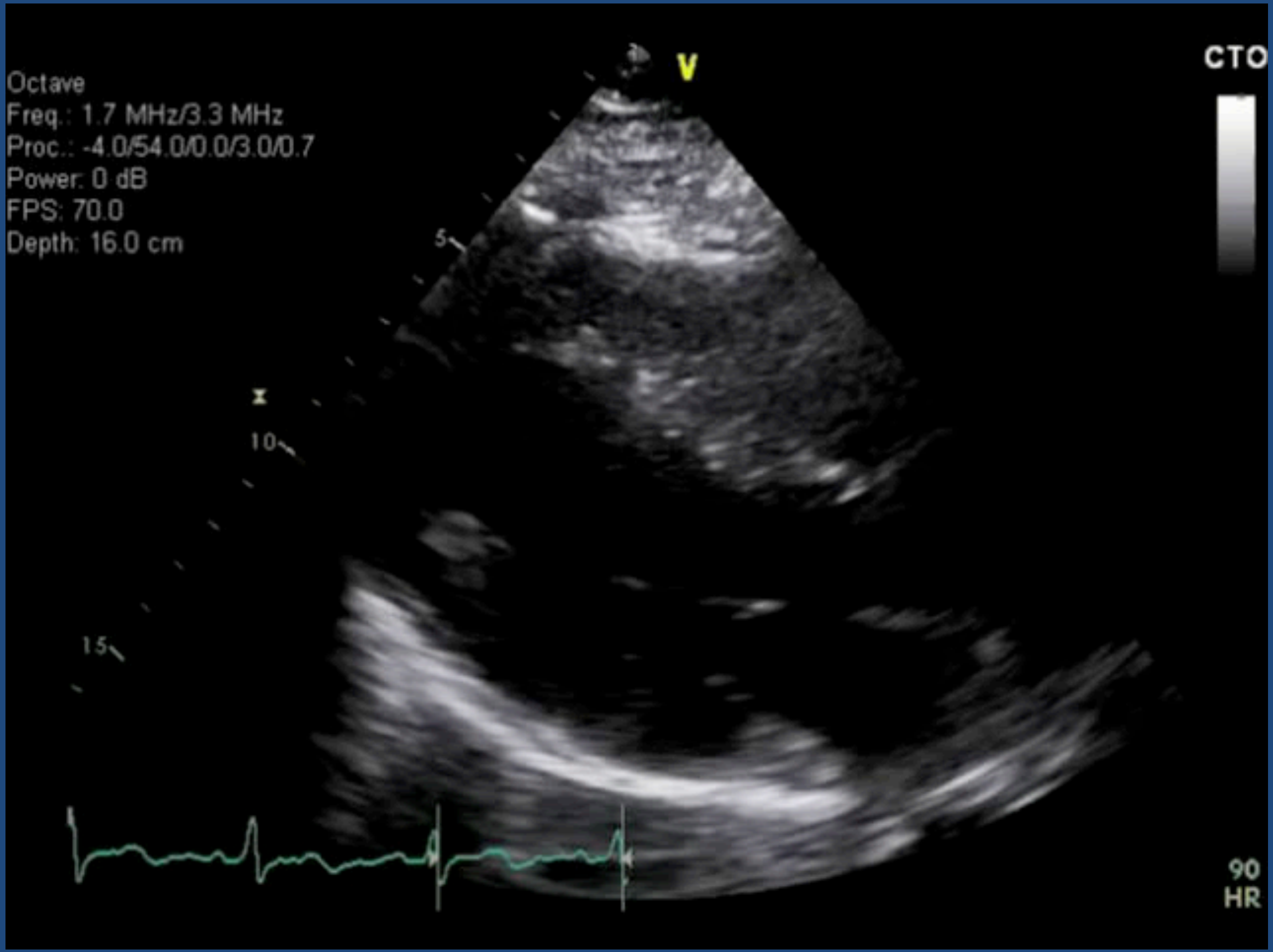
# List five complications in an adult patient following a transannular patch repair in infancy for ToF.

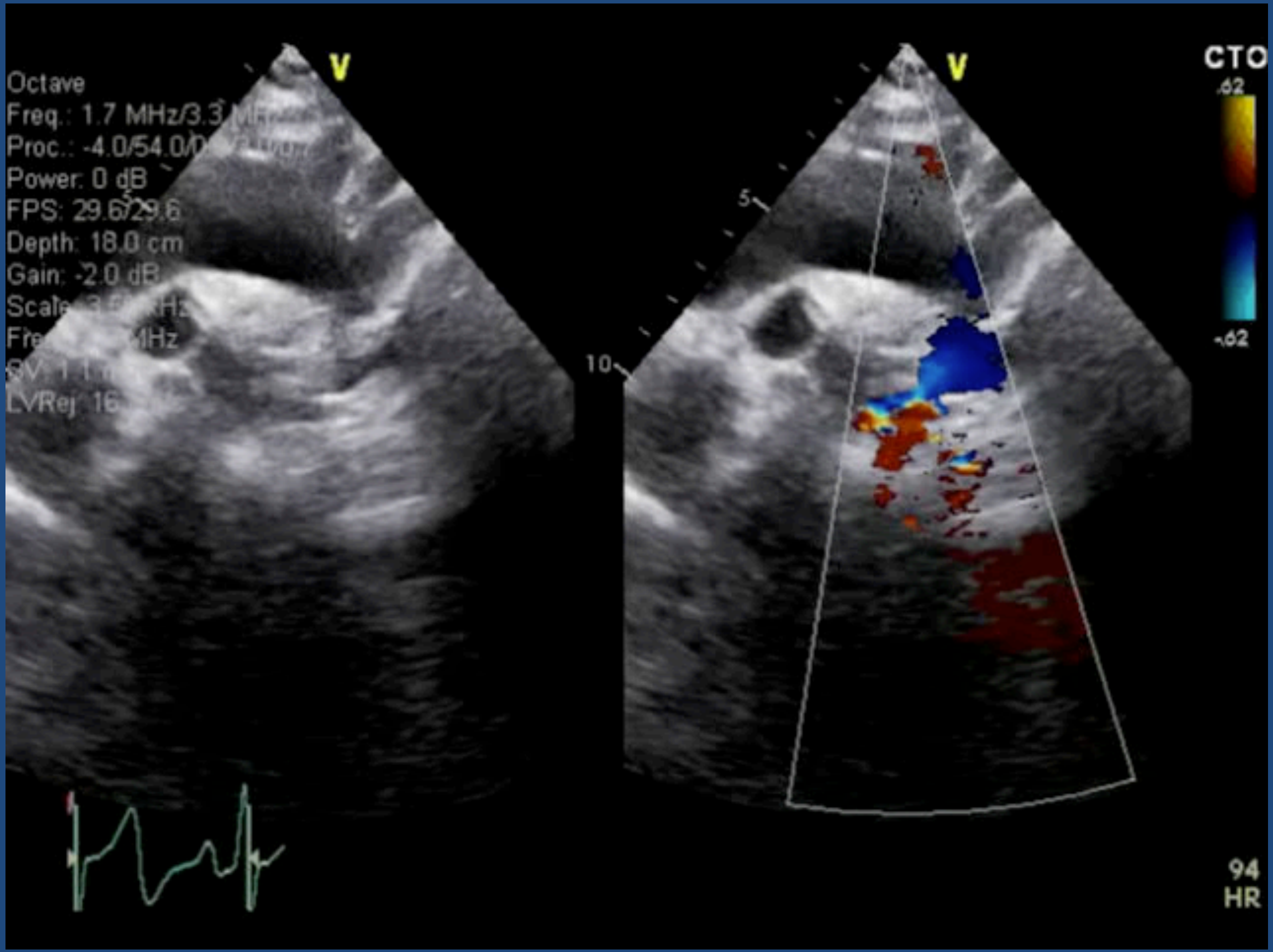
- Right ventricular dilatation
- Right heart failure
- Severe free pulmonary valve regurgitation
- Atrial arrhythmias
- Ventricular arrhythmias
- Sudden cardiac death
- Aortic dilatation
- Aortic valve regurgitation
- Endocarditis
- Septic pulmonary emboli

# Case #3

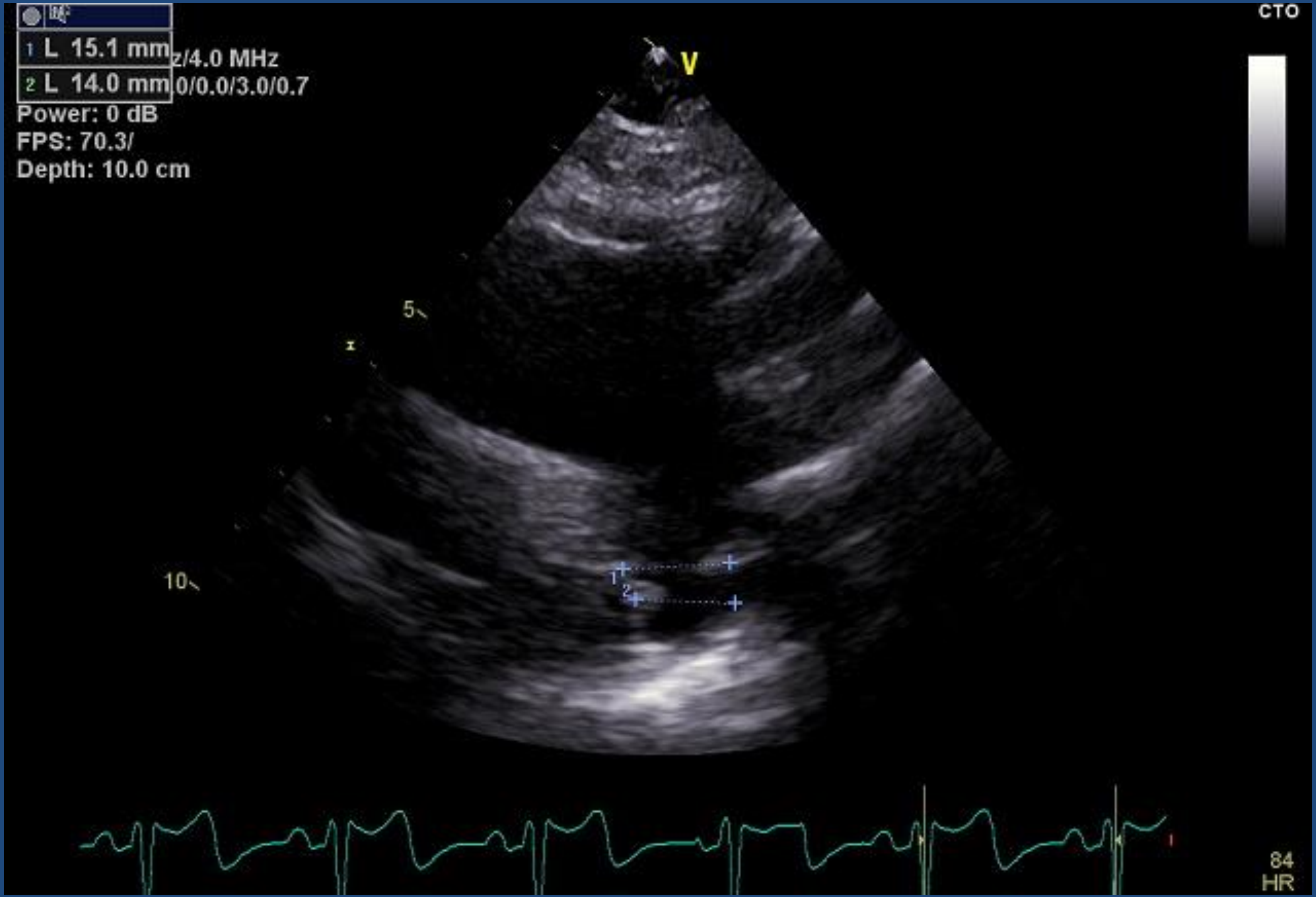
- 24 F, referred for severe HTN (220/110 mmHg)
- Frequent headaches
- On enalapril 10 mg PO BID and atenolol 100 mg PO die since 18 y.o.
- Would like to become pregnant

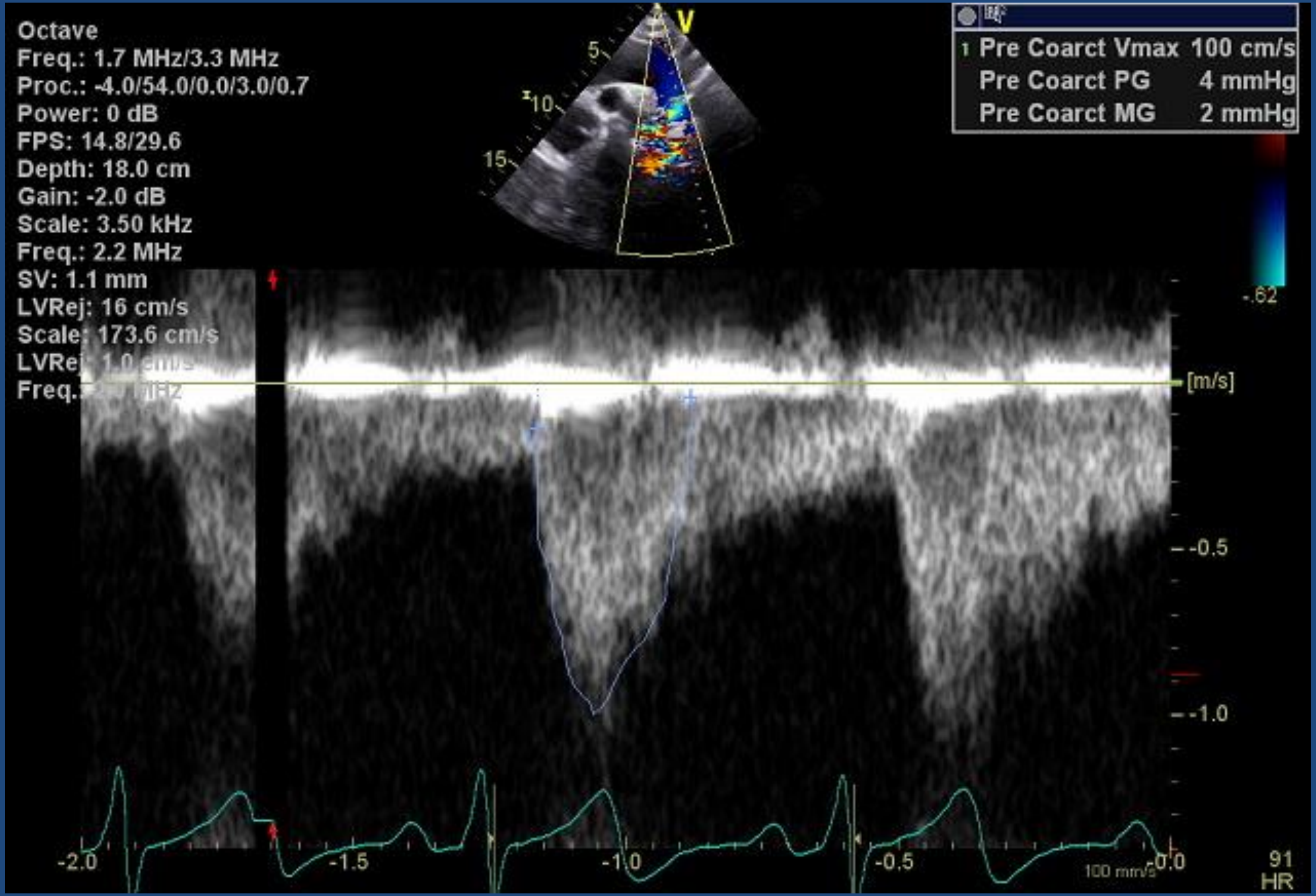
There are 5 echo clips to view followed by questions


Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: -4.0/54.0/0.0/3.0/0.7
Power: 0 dB
FPS: 70.0
Depth: 16.0 cm
V
CTO
5
10
15
90
HR

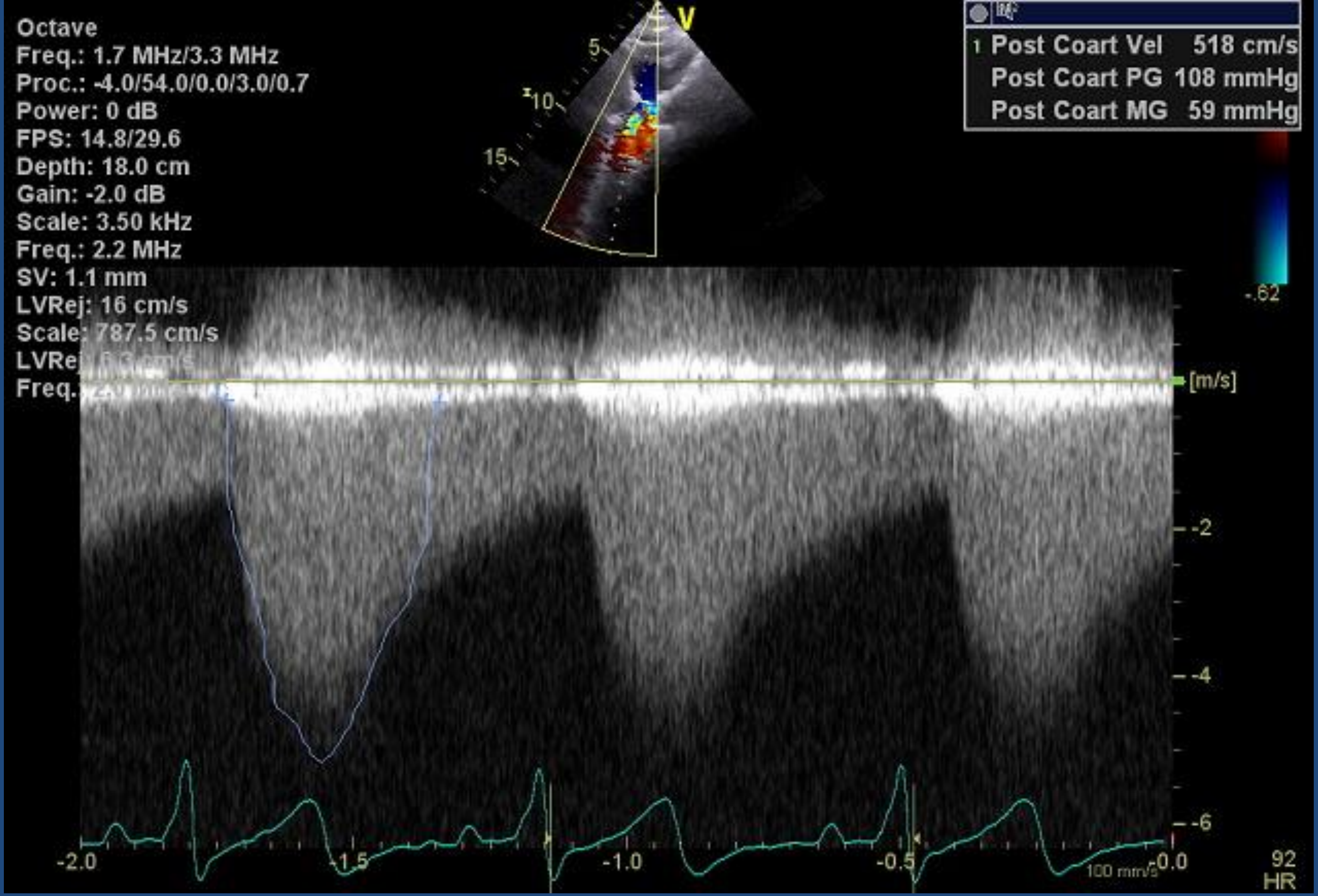


Octave
Power: 0 dB
FPS: 29.6/29.6
Depth: 18.0 cm
V
V
CTO
.62
-.62
5
10
94
HR

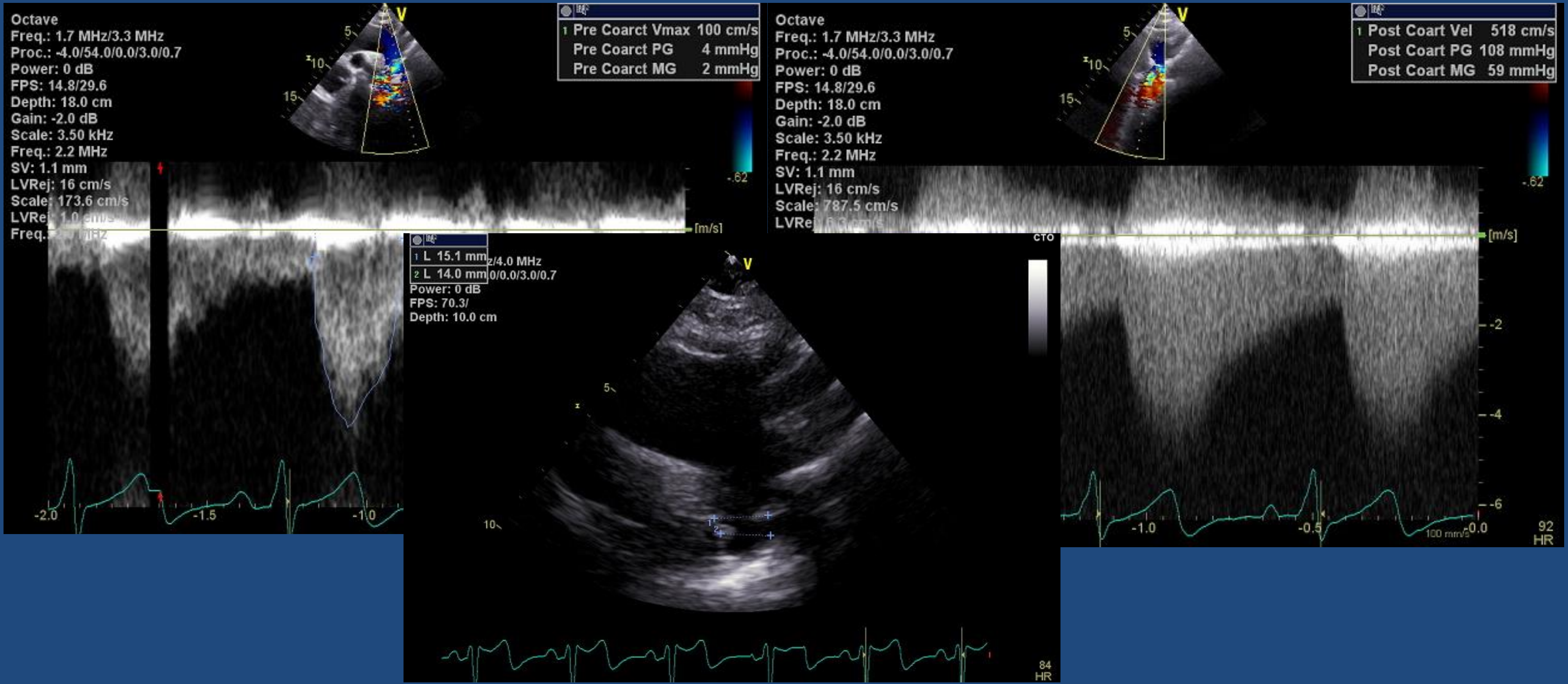

1 L 15.1 mm
2 L 14.0 mm
z/4.0 MHz
0/0.0/3.0/0.7
Power: 0 dB
FPS: 70.3/
Depth: 10.0 cm
CTO
V
5
10
84
HR



Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: -4.0/54.0/0.0/3.0/0.7
Power: 0 dB
FPS: 14.8/29.6
Depth: 18.0 cm
Gain: -2.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 16 cm/s
Scale: 173.6 cm/s
1 Pre Coarct Vmax 100 cm/s
Pre Coarct PG 4 mmHg
Pre Coarct MG 2 mmHg
[m/s]
-0.5
-1.0
-2.0
-1.5
-1.0
-0.5
0.0
100 mm/s
91
HR

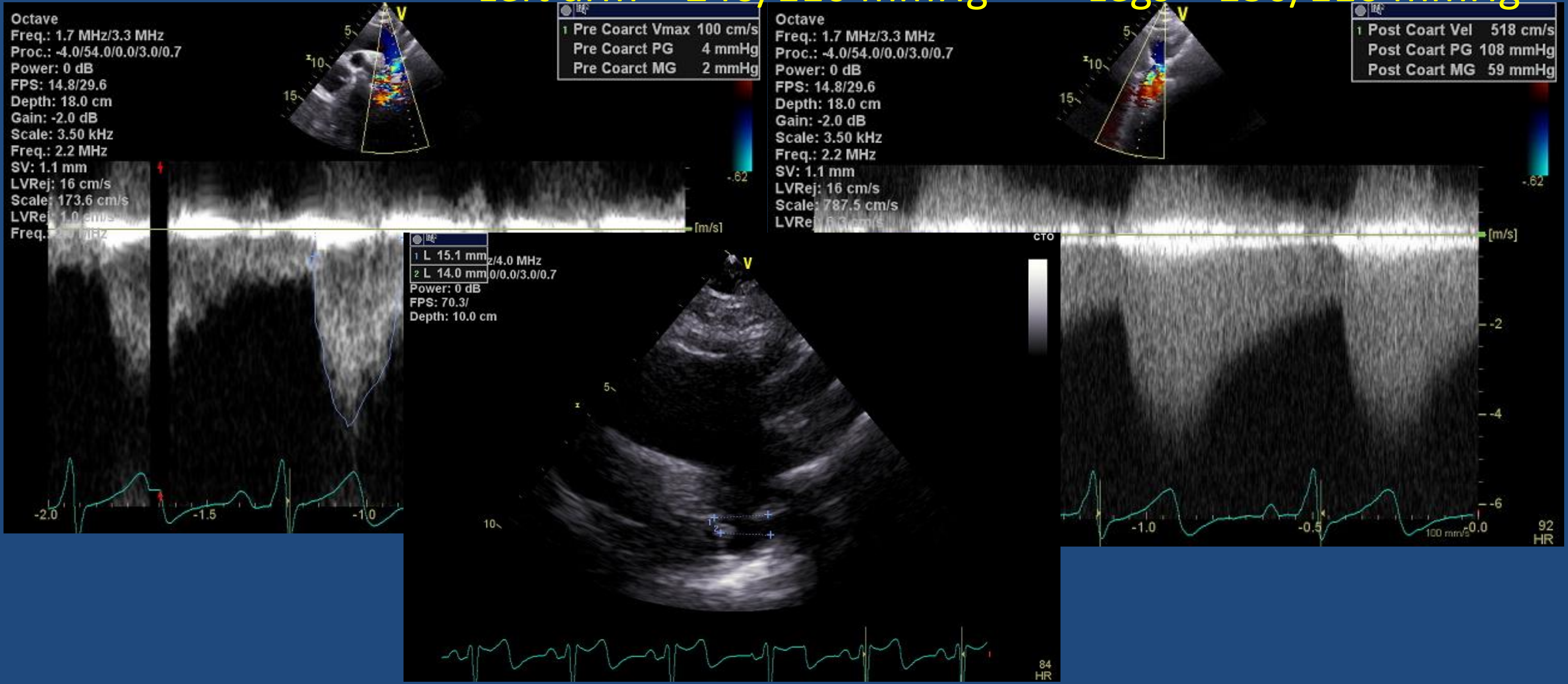

Octave
Freq.: 1.7 MHz/3.3 MHz
Proc.: -4.0/54.0/0.0/3.0/0.7
Power: 0 dB
FPS: 14.8/29.6
Depth: 18.0 cm
Gain: -2.0 dB
Scale: 3.50 kHz
Freq.: 2.2 MHz
SV: 1.1 mm
LVRej: 16 cm/s
Scale: 787.5 cm/s
1 Post Coart Vel 518 cm/s
Post Coart PG 108 mmHg
Post Coart MG 59 mmHg
[m/s]
-2
-4
-6
-2.0
-1.5
-1.0
-0.5
0.0
100 mm/s
92
HR

# The BP in the right arm is 240/110 mmHg.
# What is the blood pressure in the left arm and in the legs?



# The BP in the right arm is 240/110 mmHg.
# What is the blood pressure in the left arm and in the legs?

Left arm = 240/110 mmHg        Legs = 136/110 mmHg

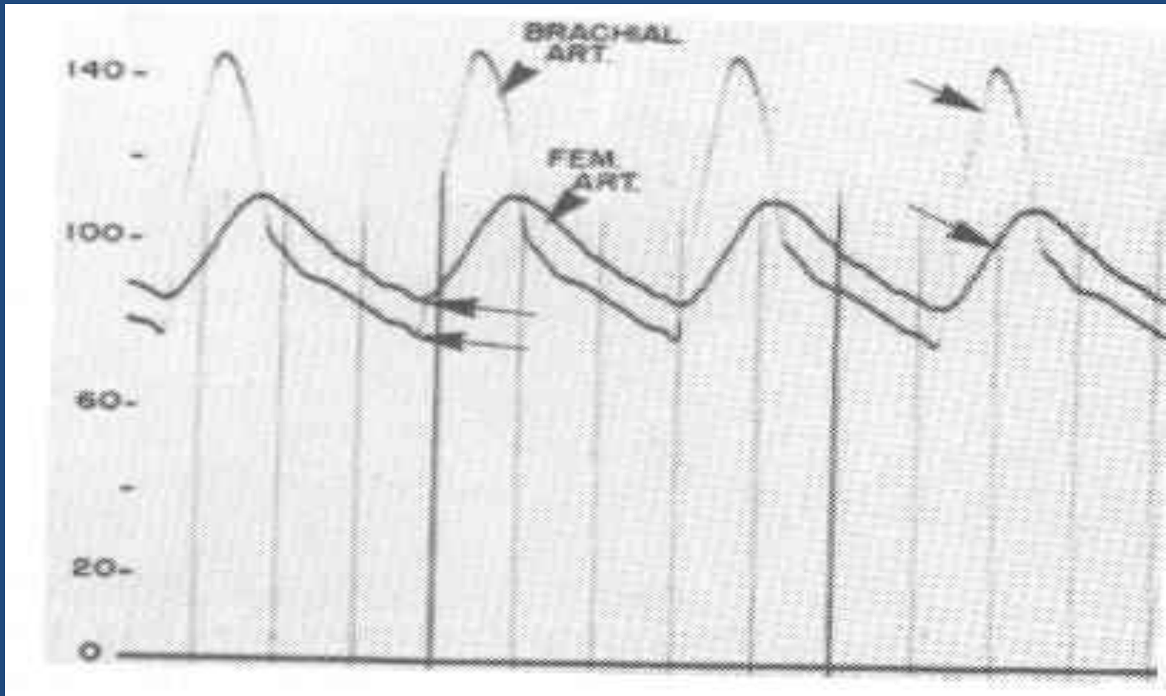


# Define significant coarctation?

# List supportive findings on physical exam (no collaterals)

- Proximal HTN with echocardiographic or angiographic evidence of coarctation
- Peak transcoarctation gradient >20 mmHg across the coarctation site at angiography

- Upper extremity HTN
- Arm>leg SBP difference ≥20 mmHg
- Radial-femoral pulse delay
- ↓ pulses in the lower extremities



# List two <u>non-cardiac</u> conditions associated with coarctation.

- Turner's syndrome (10%)
- Intracranial aneurysms (circle of Willis) (3-5%)
- Corkscrew retinopathy
- Acquired intercostal artery aneurysms

# List three cardiovascular sequelae of coarctation that may be present AFTER a coarctation repair.

- Hypertension (difficult to control)
  - Diffuse arteriopathy persists after coarctation repair
- Exaggerated blood pressure response to exercise
- Recoarctation
- Aneurysm/pseudoaneurysm at the repair site
- Bicuspid aortic valve (+/-AS, AR) (70-80%)
- Intracranial aneurysms (circle of Willis) (3-5%)

# What do you advise this patient wrt pregnancy?

- Pregnancy is contraindicated: mWHO IV recommendation

# Pregnancy Risk Assessment

- Recommended to perform a risk assessment in all women with cardiac diseases of childbearing age <u>before </u>conception (or early in pregnancy). *mWHO classification of maternal risk (ESC; IC)*

# mWHO Classification

- Most accurate system of risk assessment*
- Best score performance in women with CHD
- More appropriate for developed countries
- Indications for intervention similar for women desiring pregnancy and non-pregnant women
  - Except if
    - ≥ moderate MS
    - Aortopathy
- Fertility treatments contraindicated mWHO IV

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ESC
European Society of Cardiology

European Heart Journal (2018) **39**, 3165–3241
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**ESC GUIDELINES**

# 2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

## The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

## Endorsed by: the International Society of Gender Medicine (IGM), the German Institute of Gender in Medicine (DGesGM), the European Society of Anaesthesiology (ESA), and the European Society of Gynecology (ESG)

**Authors/Task Force Members: Vera Regitz-Zagrosek* (Chairperson) (Germany), Jolien W. Roos-Hesselink* (Co-Chairperson) (The Netherlands), Johann Bauersachs (Germany), Carina Blomström-Lundqvist (Sweden), Renata Cífková (Czech Republic), Michele De Bonis (Italy), Bernard Iung (France), Mark Richard Johnson (UK), Ulrich Kintscher (Germany), Peter Kranke[1] (Germany), Irene Marthe Lang (Austria), Joao Morais (Portugal), Petronella G. Pieper (The Netherlands), Patrizia Presbitero (Italy), Susanna Price (UK), Giuseppe M. C. Rosano (UK/Italy), Ute Seeland (Germany), Tommaso Simoncini[2] (Italy), Lorna Swan (UK), Carole A. Warnes (USA)**

# mWHO Classification

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**Table 3** Modified World Health Organization classification of maternal cardiovascular risk

| | mWHO I | mWHO II | mWHO II–III | mWHO III | mWHO IV |
|---|---|---|---|---|---|
| **Diagnosis (if otherwise well and uncomplicated)** | Small or mild<br>– pulmonary stenosis<br>– patent ductus arteriosus<br>– mitral valve prolapse<br>Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage)<br>Atrial or ventricular ectopic beats, isolated | Unoperated atrial or ventricular septal defect<br>Repaired tetralogy of Fallot<br>Most arrhythmias (supraventricular arrhythmias)<br>Turner syndrome without aortic dilatation | Mild left ventricular impairment (EF >45%)<br>Hypertrophic cardiomyopathy<br>Native or tissue valve disease not considered WHO I or IV (mild mitral stenosis, moderate aortic stenosis)<br>Marfan or other HTAD syndrome without aortic dilatation<br>Aorta <45 mm in bicuspid aortic valve pathology<br>Repaired coarctation<br>Atrioventricular septal defect | Moderate left ventricular impairment (EF 30–45%)<br>Previous peripartum cardiomyopathy without any residual left ventricular impairment<br>Mechanical valve<br>Systemic right ventricle with good or mildly decreased ventricular function<br>Fontan circulation. If otherwise the patient is well and the cardiac condition uncomplicated<br>Unrepaired cyanotic heart disease<br>Other complex heart disease<br>Moderate mitral stenosis<br>Severe asymptomatic aortic stenosis<br>Moderate aortic dilatation (40–45 mm in Marfan syndrome or other HTAD; 45–50 mm in bicuspid aortic valve, Turner syndrome ASI 20–25 mm/m$^2$, tetralogy of Fallot <50 mm)<br>Ventricular tachycardia | Pulmonary arterial hypertension<br>Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV)<br>Previous peripartum cardiomyopathy with any residual left ventricular impairment<br>Severe mitral stenosis<br>Severe symptomatic aortic stenosis<br>Systemic right ventricle with moderate or severely decreased ventricular function<br>Severe aortic dilatation (>45 mm in Marfan syndrome or other HTAD, >50 mm in bicuspid aortic valve, Turner syndrome ASI >25 mm/m$^2$, tetralogy of Fallot >50 mm)<br>Vascular Ehlers–Danlos<br>Severe (re)coarctation<br>Fontan with any complication |
| **Risk** | No detectable increased risk of maternal mortality and no/mild increased risk in morbidity | Small increased risk of maternal mortality or moderate increase in morbidity | Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity | Significantly increased risk of maternal mortality or severe morbidity | Extremely high risk of maternal mortality or severe morbidity |
| **Maternal cardiac event rate** | 2.5–5% | 5.7–10.5% | 10–19% | 19–27% | 40–100% |
| **Counselling** | Yes | Yes | Yes | Yes: expert counselling required | Yes: pregnancy contraindicated: if pregnancy occurs, termination should be discussed |
| **Care during pregnancy** | Local hospital | Local hospital | Referral hospital | Expert centre for pregnancy and cardiac disease | Expert centre for pregnancy and cardiac disease |
| **Minimal follow-up visits during pregnancy** | Once or twice | Once per trimester | Bimonthly | Monthly or bimonthly | Monthly |
| **Location of delivery** | Local hospital | Local hospital | Referral hospital | Expert centre for pregnancy and cardiac disease | Expert centre for pregnancy and cardiac disease |



ASI = aortic size index; EF = ejection fraction; HTAD = heritable thoracic aortic disease; mWHO = modified World Health Organization classification; NYHA = New York Heart Association; WHO = World Health Organization.

| Maternal cardiac event rate | 2.5–5% | 5.7–10.5% | 10–19% | 19–27% | 40–100% |
|---|---|---|---|---|---|
| | **mWHO I** | **mWHO II** | **mWHO II–III** | **mWHO III** | **mWHO IV** |
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# Other Risk Scores

- CARPREG Score (Siu SC et al. Circ 2001)
  - PHTN and aortopathy ≠ predictors of outcome
  - Included pts with congenital and acquired heart dz
  - Overestimates risk in RHD?
- CARPREG 2 (Silversides C et al. Circ 2018)
  - Combines lesion specific and general factors to improve predictive accuracy
- Zahara Risk Score (Drenthen W et al. EHJ 2010)
  - PHTN and aortopathy ≠ predictors of outcome
  - Only included women with CHD
  - Valve dz and mechanical prosthesis used in model

# CARPREG

- Score calculated by giving 1 point for:
  - Cardiac events prior to pregnancy
    - Arrhythmia
    - Congestive heart failure
    - Stroke/transient ischemic attack
  - Baseline NYHA class >II or cyanosis
  - LV dysfunction (LVEF <40%)
  - Left heart obstruction
    - MVA <2 cm^2, AVA < 1.5 cm^2, LVOT PG >30 mmHg on echo

- Very few patients were included with:
  - Pulmonary HTN (not included in score but known high risk)
  - Aortopathy (not included in score so not predictor of poor outcome)

- Higher scores are associated with worse outcomes

Siu SC et al. *Circulation*. 2001; 104: 515-521.

# Assessing Pregnancy Risk

**TABLE 4. Accuracy of Risk Index**

| No. of Predictors | Estimated Risk, % | Rate of Primary Cardiac Events | | | Rate of Primary or Secondary Cardiac Events, Revised Index | |
|---|---|---|---|---|---|---|
| | | Derivation Group, Revised Index | Validation Group | | Derivation Group | Validation Group |
| | | | Revised Index | Original Index | | |
| 0 | 5 | 7/249 (3%) | 5/137 (4%) | 5/136 (4%) | 10/249 (4%) | 6/137 (4%) |
| 1 | 27 | 27/111 (24%) | 17/64 (27%) | 16/61 (26%) | 35/111 (31%) | 20/64 (31%) |
| >1 | 75 | 16/25 (64%) | 8/13 (62%) | 9/17 (53%) | 17/25 (68%) | 9/13 (69%) |
| C statistic (95% CI) | | 0.83 (0.77–0.89) | 0.80 (0.72–0.88) | 0.79 (0.71–0.87) | 0.82 (0.76–0.88) | 0.81 (0.74–0.88) |

CI indicates confidence interval.

Siu SC et al. *Circulation*. 2001; 104: 515-521.

# CARPREG II

Table 1: CARPREG II Risk Predictors

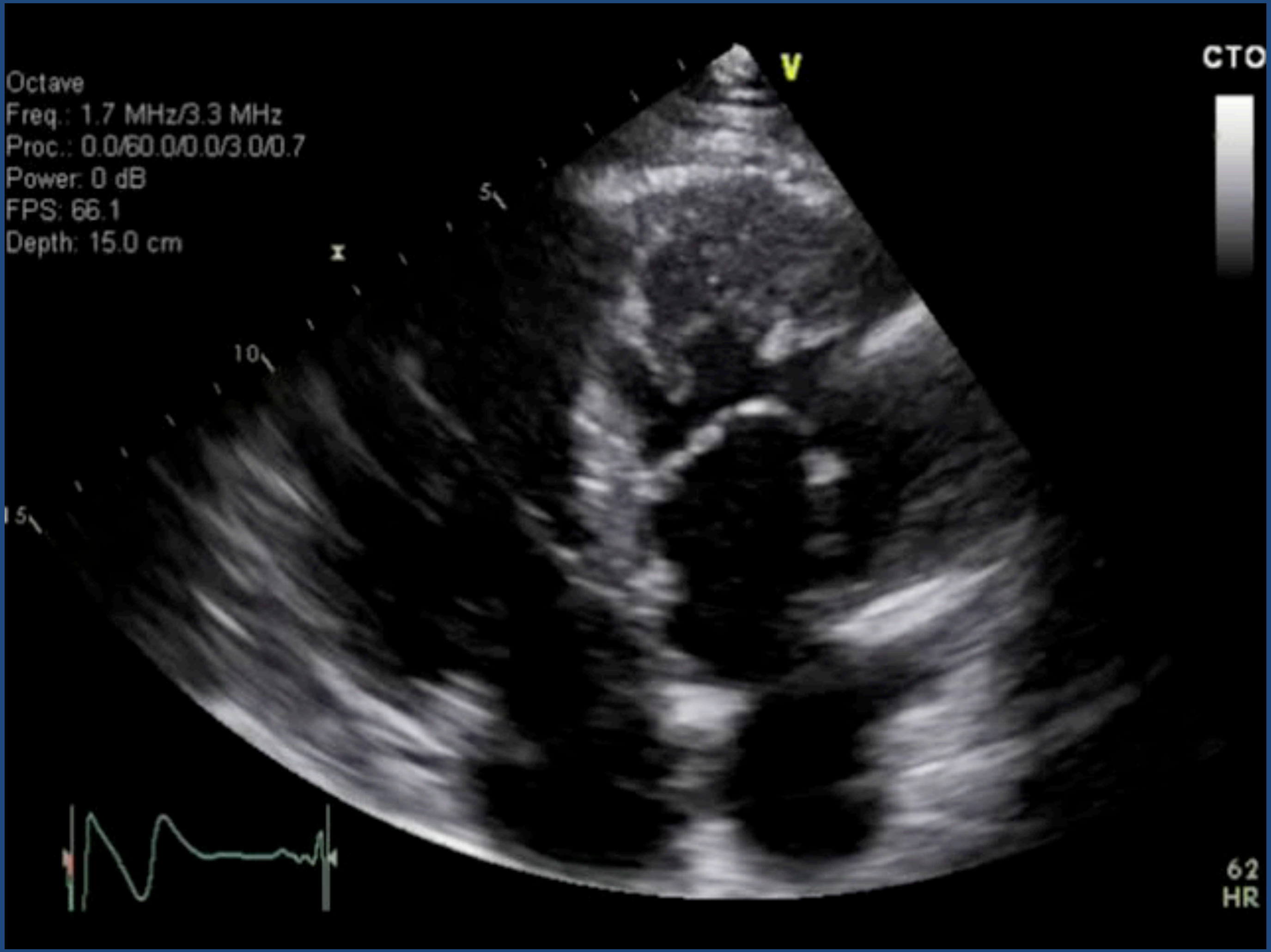
| Predictor | Points |
|---|---|
| Prior cardiac events or arrhythmias | 3 |
| Baseline NYHA 3–4 or cyanosis | 3 |
| Mechanical valve | 3 |
| Systemic ventricular dysfunction LVEF<55 % | 2 |
| High-risk valve disease or left ventricular outflow tract obstruction (aortic valve area <1.5 cm$^2$, subaortic gradient >30, or moderate to severe mitral regurgitation, mitral stenosis < 2.0 cm$^2$) | 2 |
| Pulmonary hypertension, RVSP >49 mmHg | 2 |
| High-risk aortopathy | 2 |
| Coronary artery disease | 2 |
| No prior cardiac intervention | 1 |
| Late pregnancy assessment | 1 |

*Primary cardiac event risk: score = 1, 5 % risk, score = 2, 10 % risk, score = 3, 15 % risk, score = 4, 22 % risk and 41 % risk if score greater than 4. NYHA = New York Heart Association Functional Classification; LVEF = left ventricular ejection fraction; RVSP = right ventricular systolic pressure. Source: Silversides et al., 2018, with permission from Elsevier.*[14]

Silversides C et al. Circ 2018

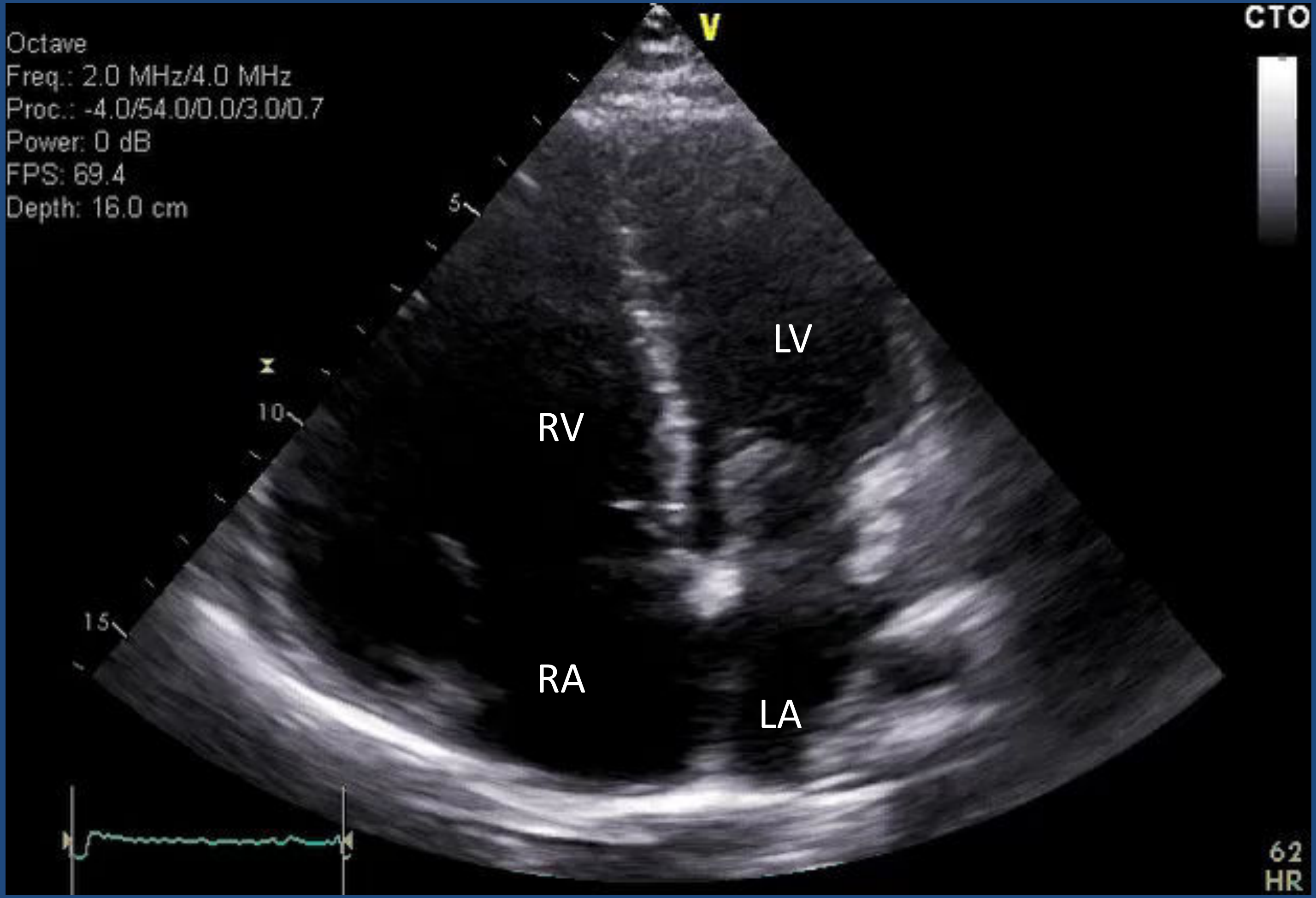
# Rapid Fire

# Diagnosis?



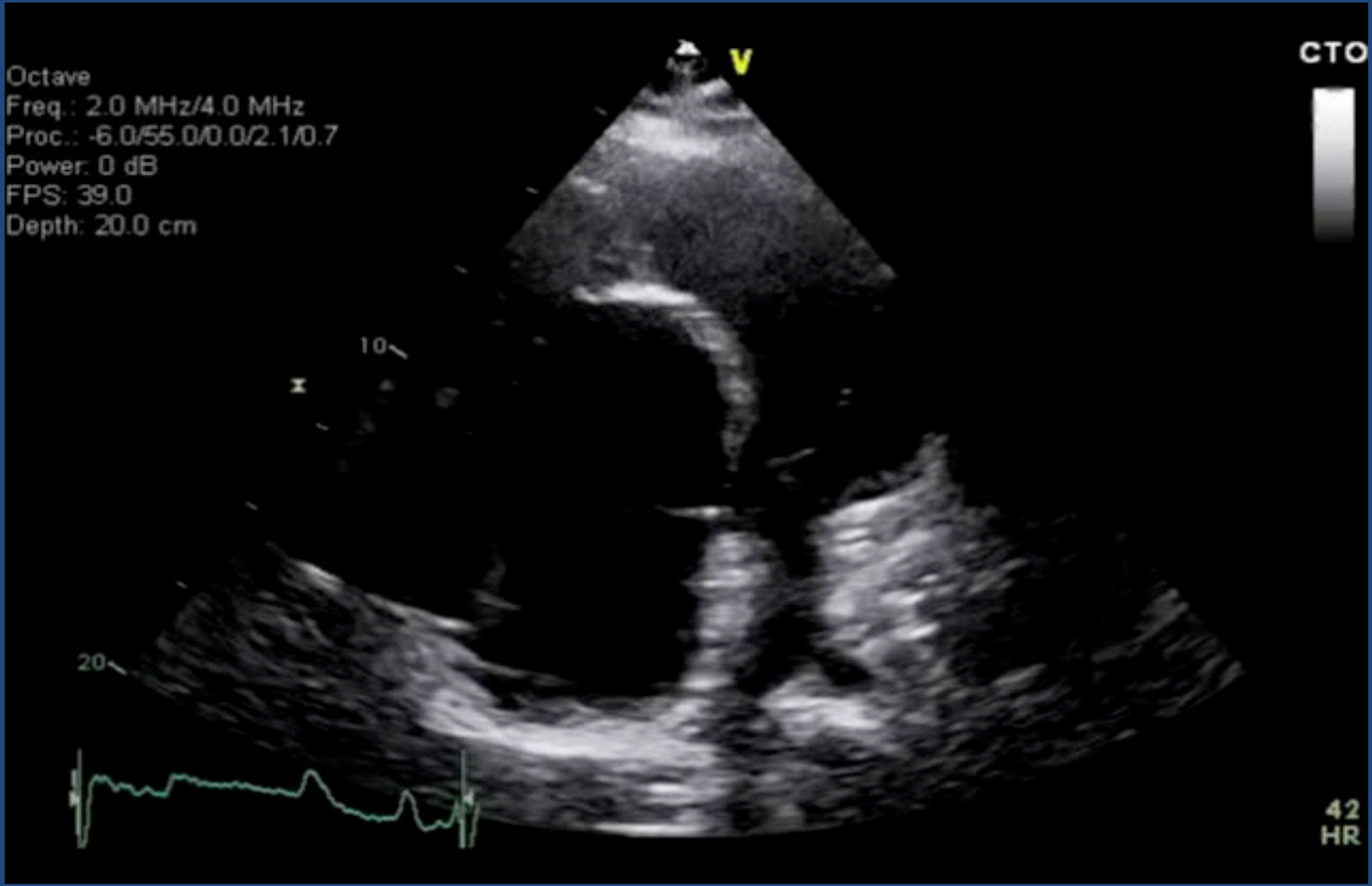

- L-TGA

# Two ACHD defects that could cause this finding?




- ASD, APVR

# Two associated conditions?




Ebstein anomaly can be associated with ASD/PFO, accessory pathway (25%; multiple 50%), RVOTO by TV

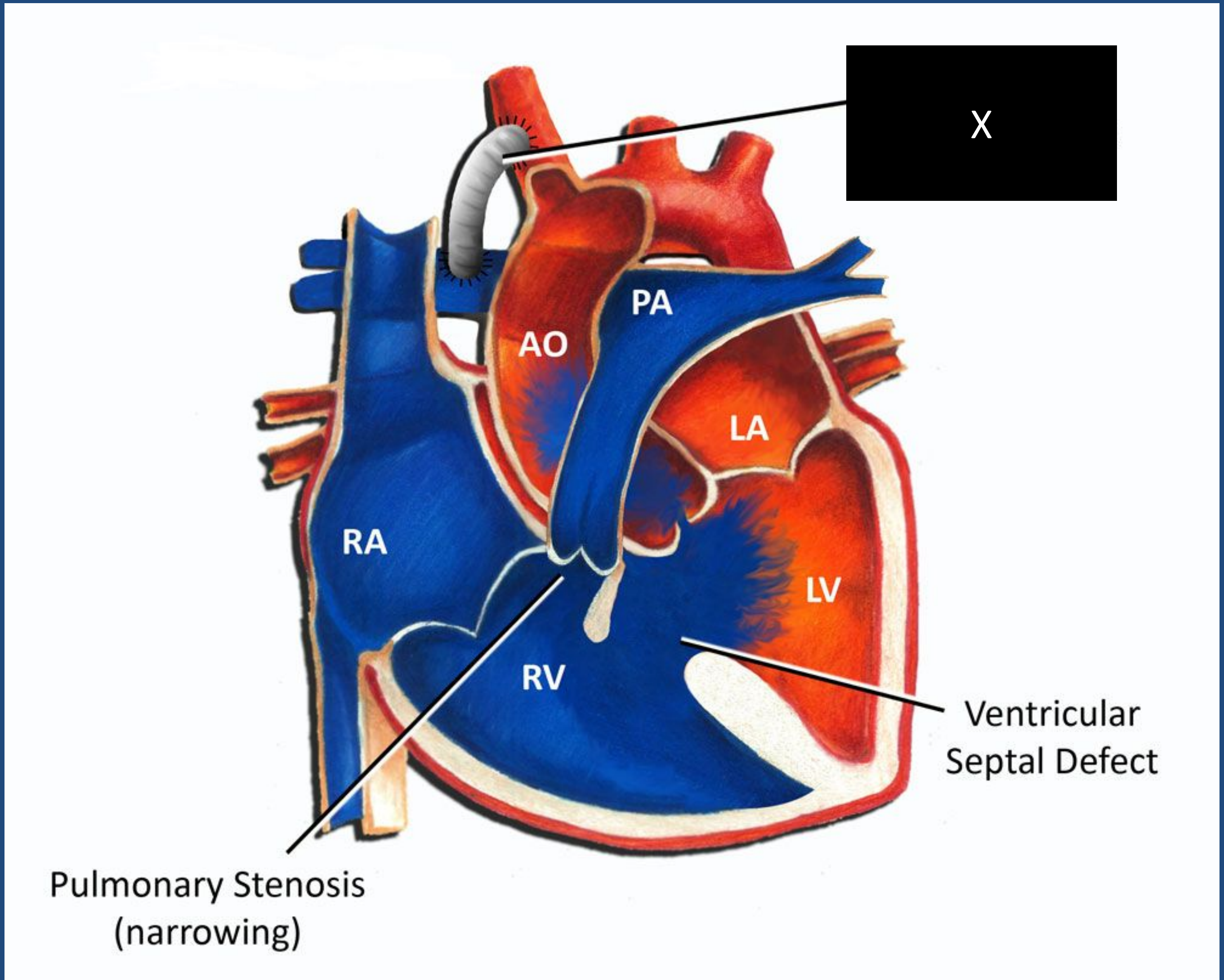
# Diagnosis?

Right hand pink

Left hand and feet blue and clubbed

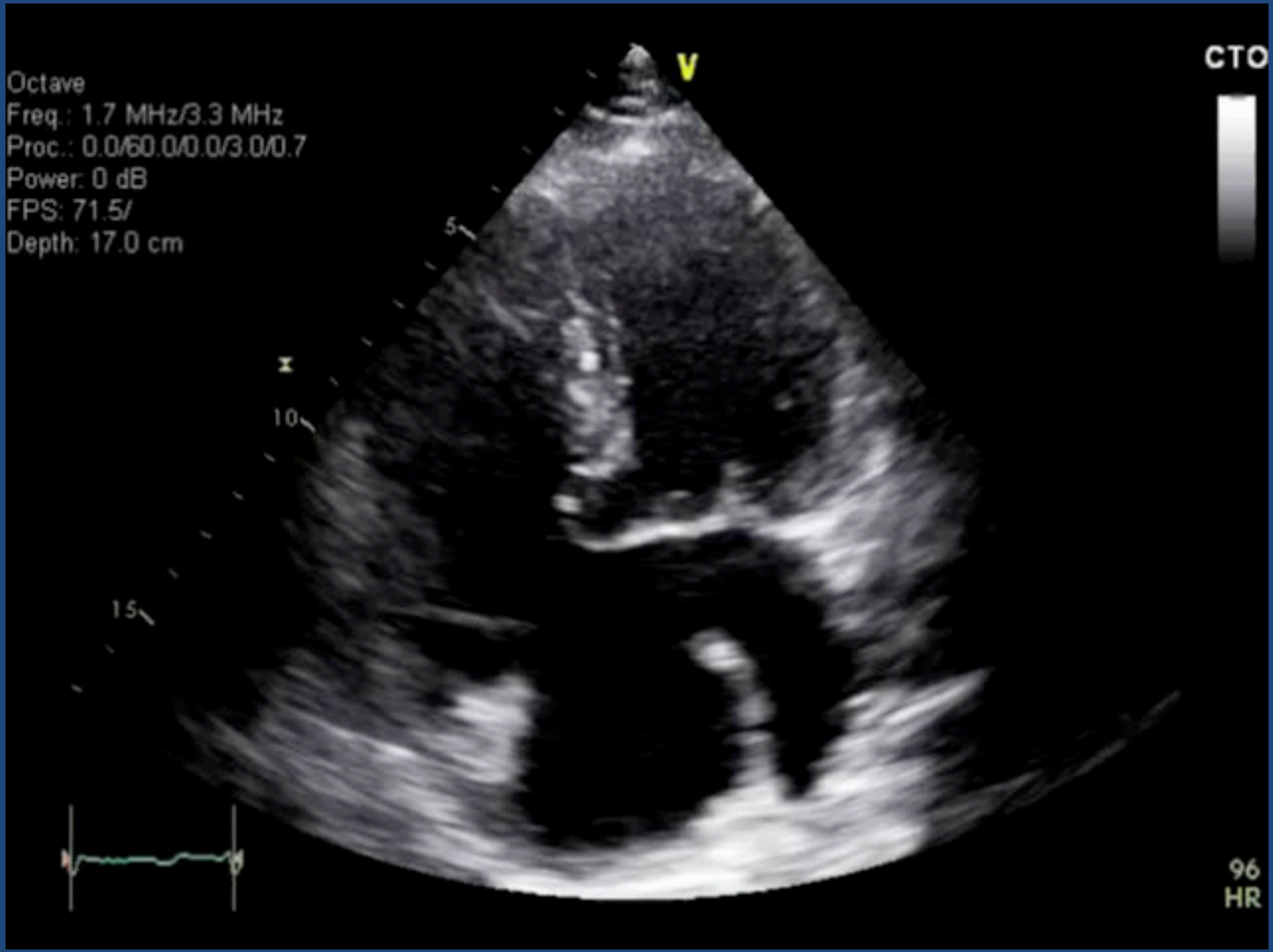
Eisenmenger PDA

# Name the structure “X”?



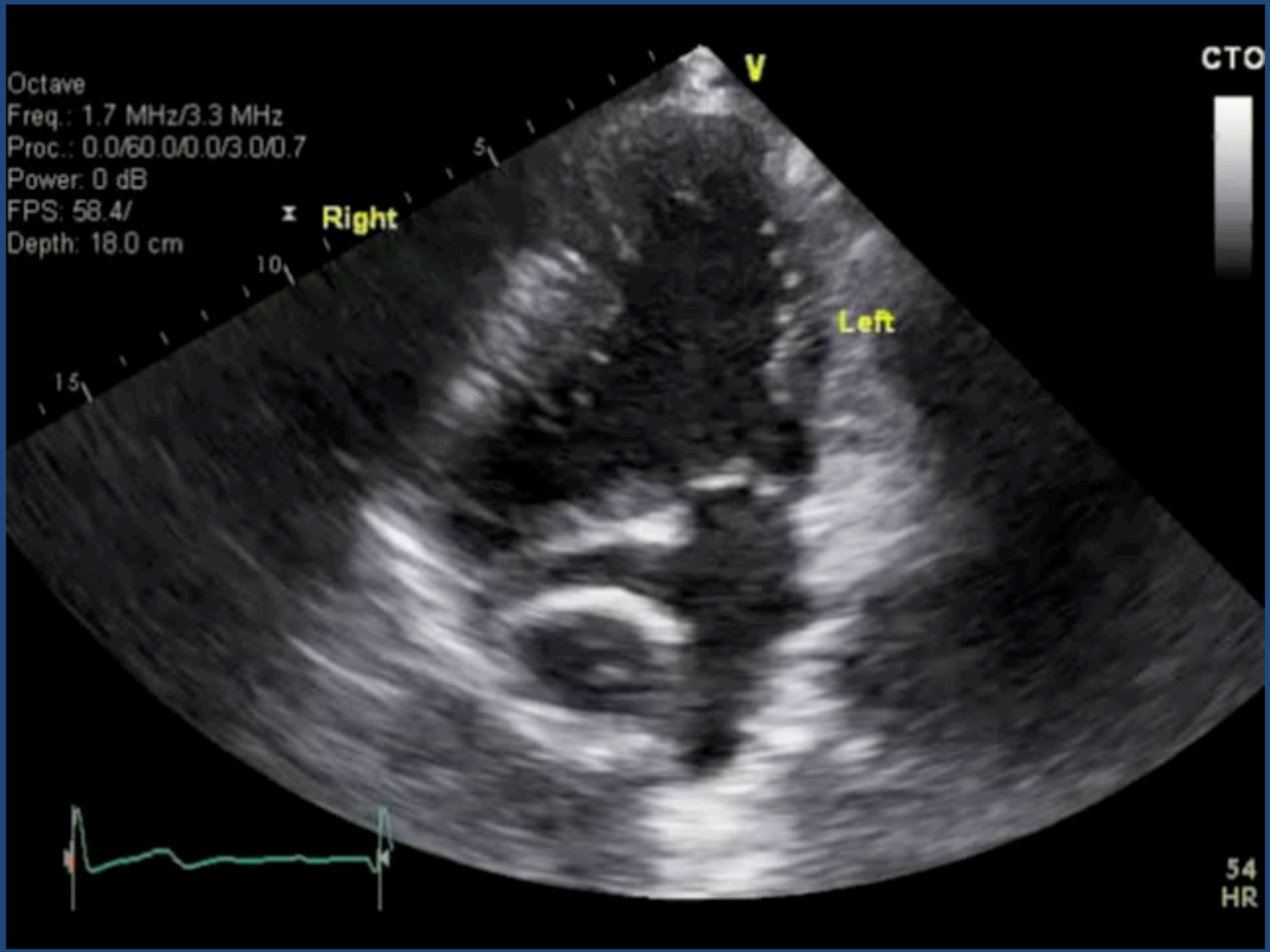

Modified Blalock-Taussig shunt; created to supply  blood to the lungs.  Used to be used as the first step in tetralogy of Fallot repair.

# Diagnosis?




Complete AV canal defect : primum ASD, common AV valve, VSD

# Two complications?




Thrombus, obstruction, leak, arrhythmias (atrial, heart block), ventricular dysfunction, systemic AV valve regurgitation, PLE, liver disease, endocarditis

Questions?