Pediatric Echocardiography

Sarah Clauss MD
Children's National Medical Center
Washington DC

What is your career?

A. Adult Echocardiographic Sonographer
B. Pediatric Echocardiography Sonographer
C. Adult and Pediatric
D. Radiology
E. Other

Children's National ™

Objectives

- Implement current best practice standards in pediatric echocardiography
- Describe the basic pediatric echocardiogram. (views, imaging techniques, etc.)
- Improve the ability of the sonographer to understand and preform high risk pediatric echocardiograms.

Children's National ™

Congenital Heart Defects
7-10/1,000 Live Births

DIAGNOSIS (Balt-Wash)	PERCENT
Ventricular septal defect	26%
Tetralogy of Fallot	9%
Atrioventricular septal defect	9%
Atrial septal defect	8%
Pulmonary valve stenosis	7%
Coarctation of the Aorta	7%
Hypoplastic left heart syndrome	6%
D-Transposition	5%

Children's National ™

CHD in Adults

30,000 babies born with CHD per year

20,000 surgeries for CHD per year

85% survive into adulthood

Over 1.2 million adults with CHD

Increasing at 5% per year

8,500 per year reach adulthood

Less than 10% disabled

Diagnosis	1950's	1960's	1970's	1980's	1990's	2000's
ASD	Rare Repair	Repair older child	Repair age 4	Repair age 2	Repair age 2-3	Device closure
VSD	Rare Repair	Repair >10 kg or palliate	Repair < 1 year or palliate	Repair 6 months or prn	Repair premature infants	
PDA	Repair	Repair	Repair	Repair	Repair	
TOF	Palliate	Late Repair in adults	Repair after palliation		Repair 2-8 months or prn	
TGA	No survivors	Rare Survivors	Atrial Repair	Transitional Decade	Arterial Repair	
Single Ventricle	Comfort care	Palliate	Rare Fontan	Fenestrated Fontan	Lateral Tunnel	Extra-cardiac Fontan
HLHS	Comfort care	Comfort care	Surgery in Boston	Comfort vs. high risk surgery	Surgery & Fetal Diagnosis	

Embryology 101

19 Days: Two endocardial tubes have formed – these tubes will fuse to form a common, single primiative heart tube

22 Days: Heart tube begins to beat

23 Days: Folding commences

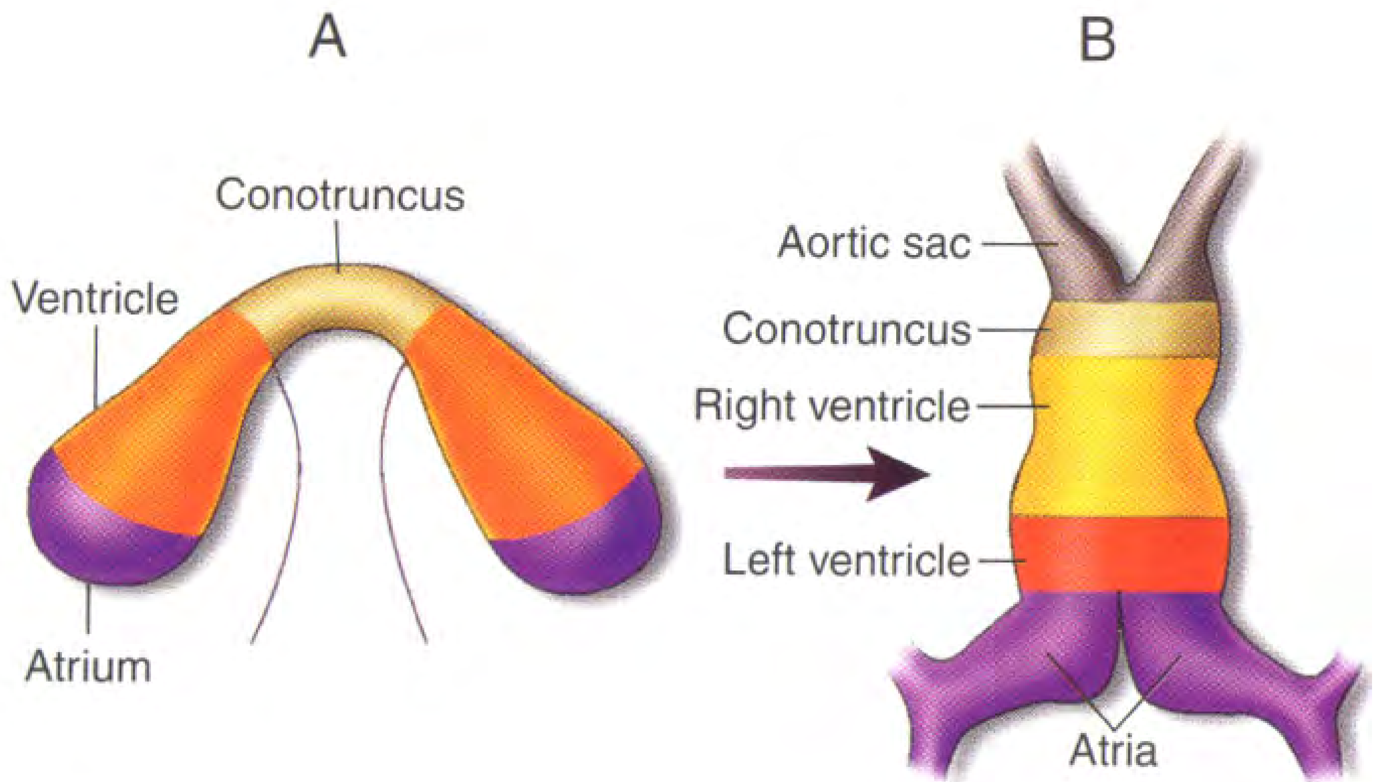
30 Days: Primitive circulation

9 weeks (56 Days): All major structures identified

(In humans, several months of gestation remain for emergence of HLHS, PS, etc)

Children's National ™

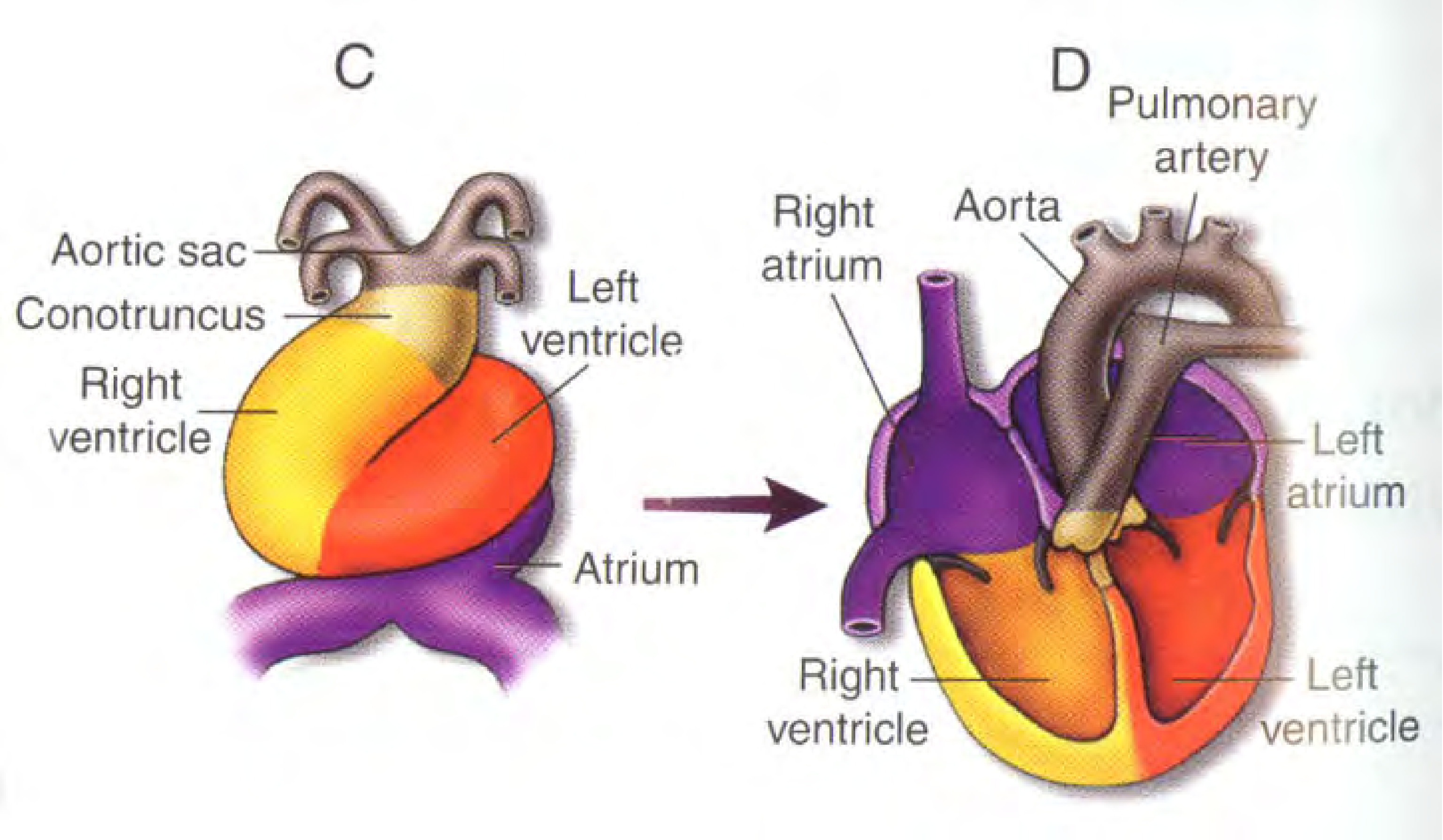
The Cardiac Crescent and the Tube Heart



From *Heart Development*, 1999

Children's National

Looping and Septation



From *Heart Development*, 1999

How do Congenital Heart Defects form?

Complex interaction between environmental and genetic etiology

- Multifactorial
- 5-8% chance of recurrence

Environmental exposures may influence micro-uterine environment and either turn on or off needed protein development

Children's National™

Echocardiography

1793 Italian priest studied bats

1845 Austrian scientist Christian Doppler

WWII Sonar detected submarines

1954 Hertz & Edler

- (A&B mode echocardiogram)

Reflection of US waves by target

Display based on

- Intensity of returned signal
- Time of “flight” or depth

Echo timeline

M-mode ultrasound early 1970's

2D echo late 1970's

Doppler Echo 1980's

- Pulsed wave Doppler
- Continuous wave Doppler
- Color Doppler

Pediatric Echo is Different

Anatomy and physiology over function

Segmental approach for complex patients

Improved resolution

- Heart is closer to chest wall
- Higher frequency transducers
- TEE rarely necessary for diagnosis

Inversion of apical and subcostal images

Diagnostic accuracy depends on image quality

Improve signal/noise ratio

Improve image resolution

Appropriate transducer

Focus depth

OPTIMAL WINDOW SHOULD ALLOW US BEAM TO BE PERPENDICULAR TO AREA OF INTEREST FOR IMAGING AND PARALLAL TO FLOW JETS FOR DOPPLER AND COLOR

5 Standard Views

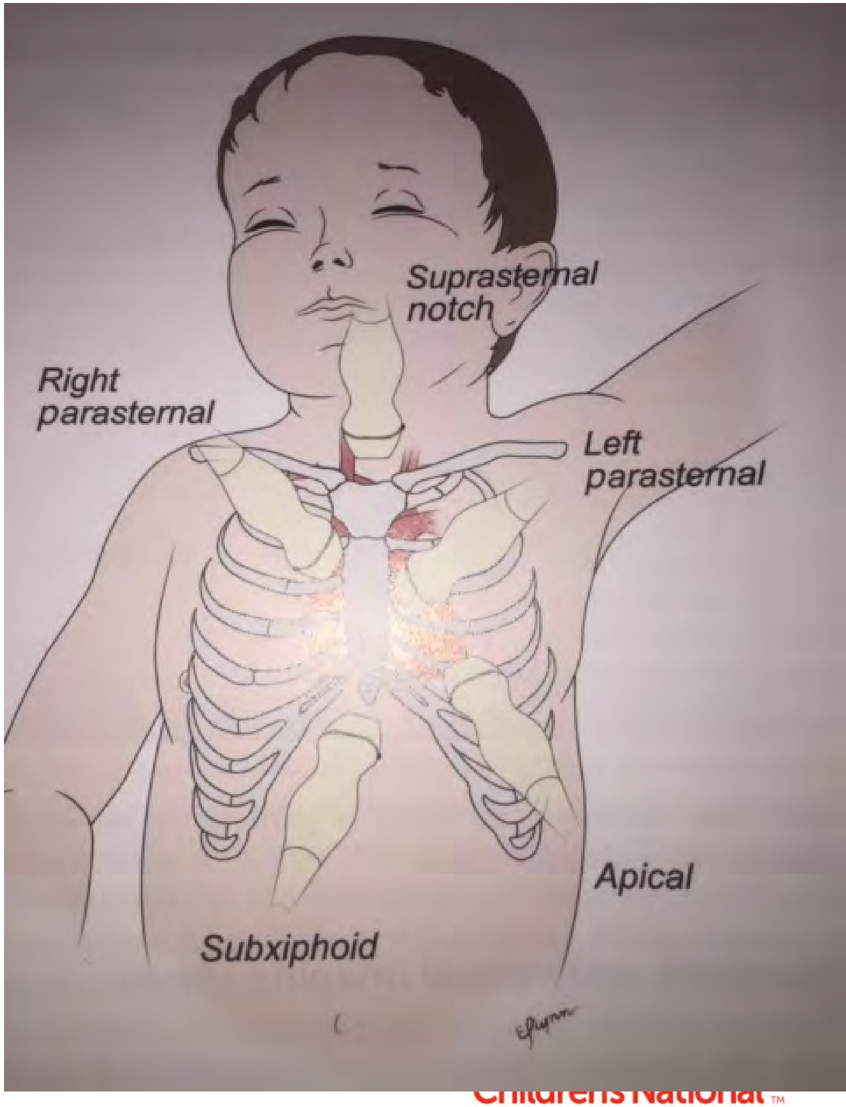
Subcostal

Left Parasternal

Apical

Suprasternal Notch

Right Parasternal

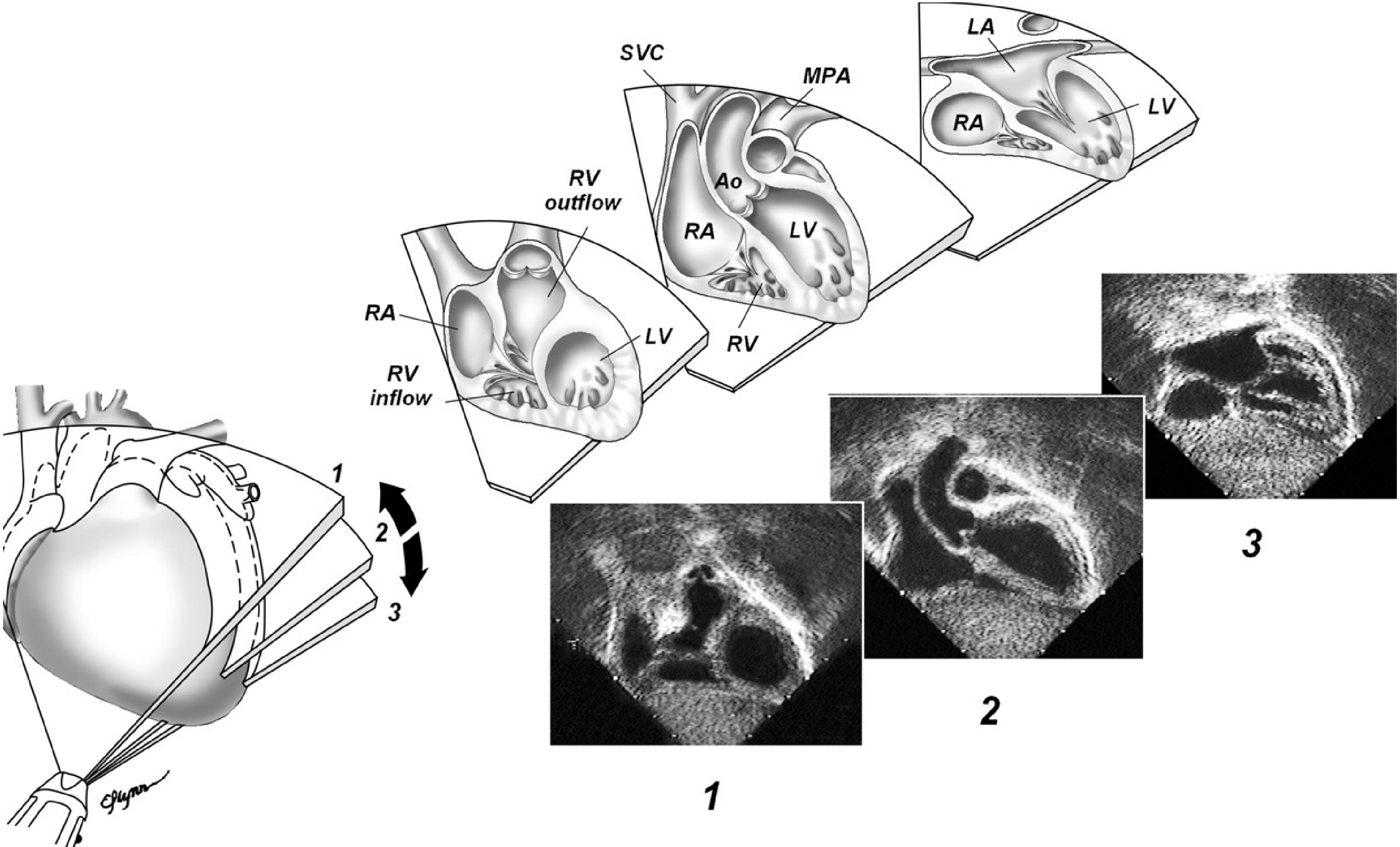


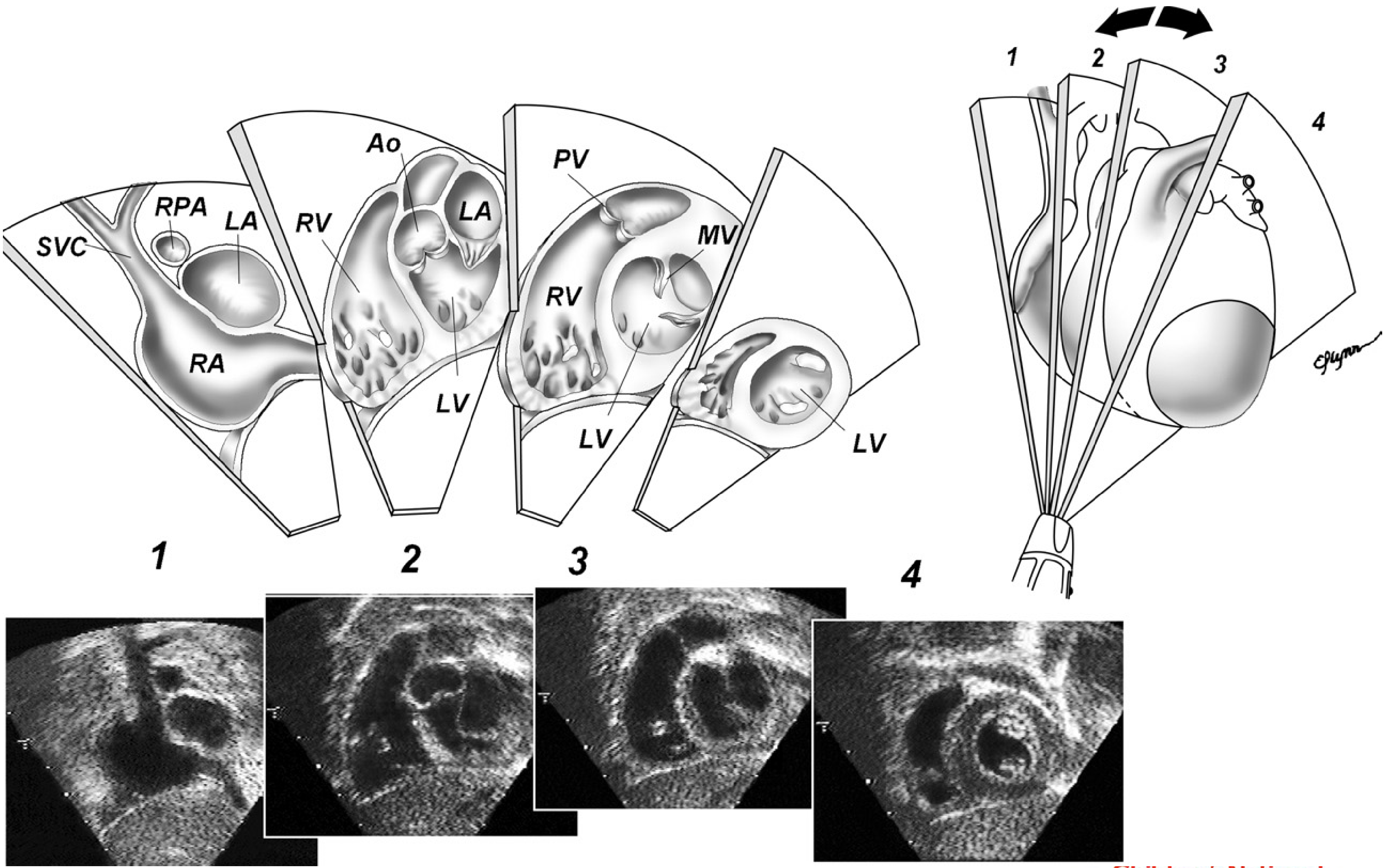
Subcostal structures

- IVC
- Hepatic veins
- Abdominal aorta
- Diaphragm
- SVC
- LA
- RA
- Atrial Septum
- Ascending aorta
- Branch PA

- Coronary sinus
- Pulmonary veins
- Mitral Valve
- Tricuspid Valve
- LV
- RV
- Ventricular Septum
- Aortic Valve
- Pulmonary Valve
- Pericardium

http://www.lai-echo.com/chapter4/video-4-2.asp



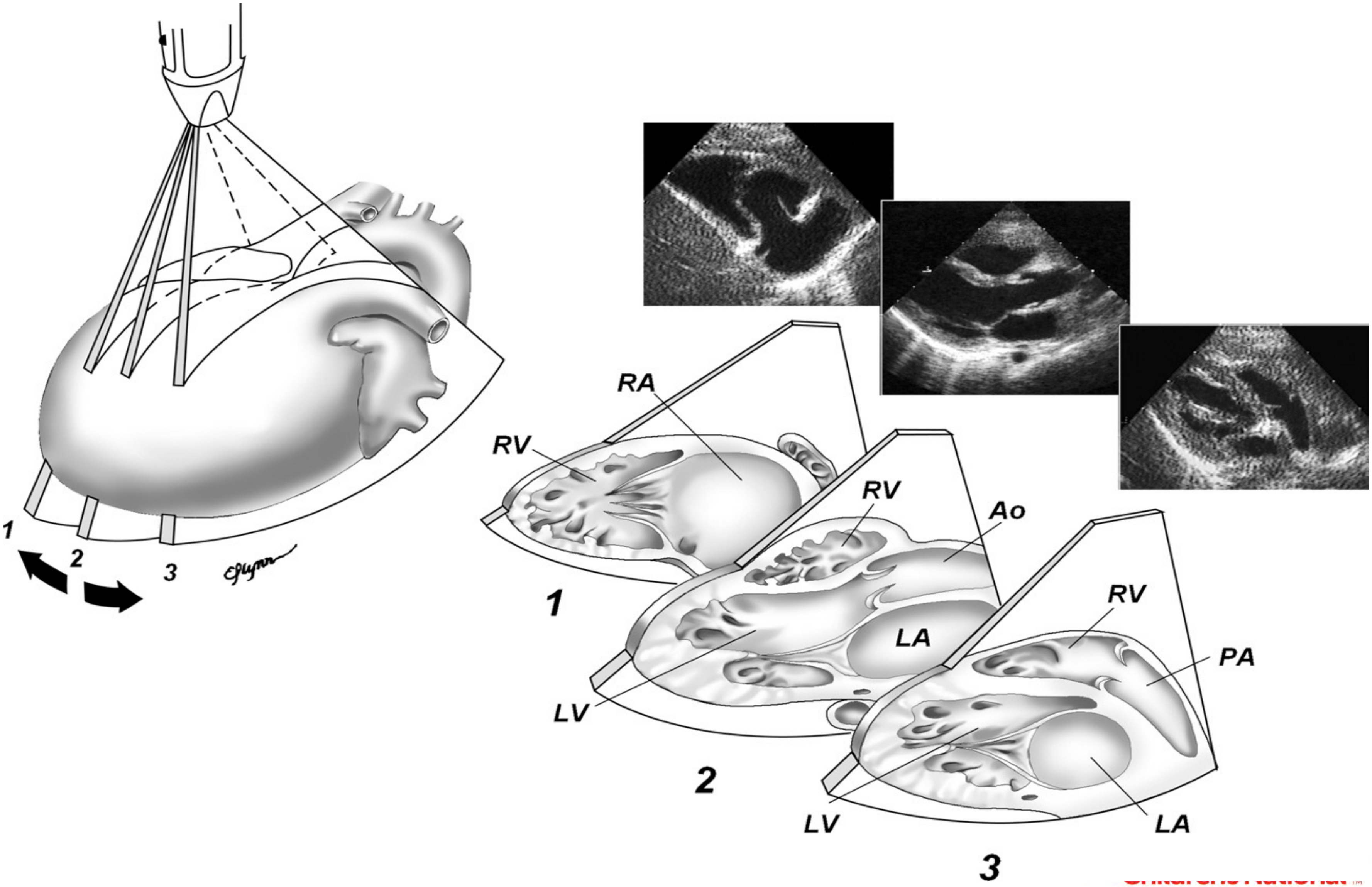

SVC
RPA
LA
RA
RV
Ao
LA
LV
PV
MV
RV
LV
LV
1
2
3
4
1
2
3
4

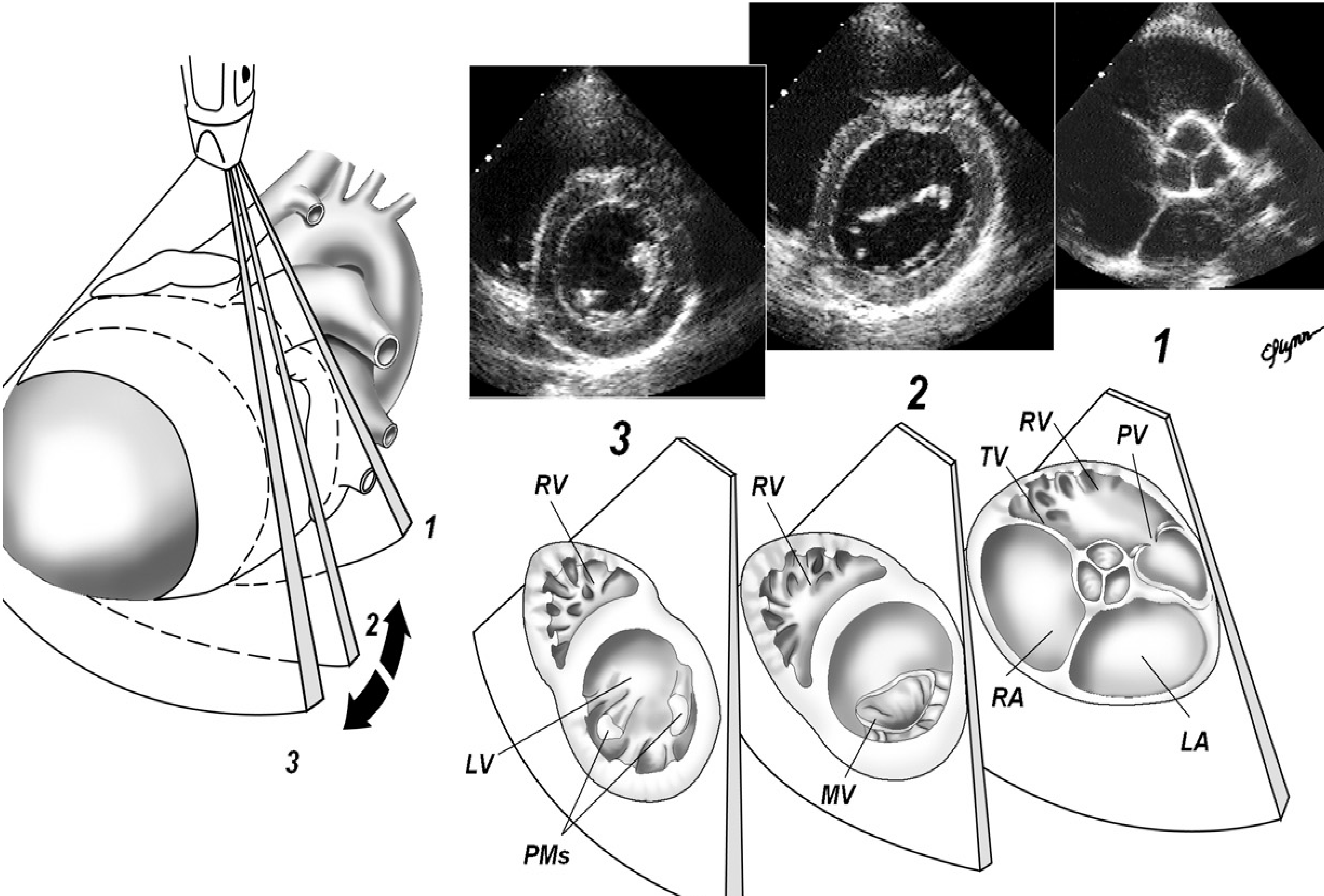
Left Parasternal

- IVC
- SVC
- LA
- RA
- Atrial septum
- Coronary sinus
- Pulmonary veins
- MV
- TV

- LV
- RV
- Ventricular septum
- Aortic Valve
- Pulmonary valve
- Ascending aorta
- Coronary arteries
- MPA/BPA
- Pericardium

http://www.lai-echo.com/chapter4/video-4-6.asp





1
2
3
1
2
3
RV
TV
PV
RA
LA
RV
MV
RV
LV
PMs

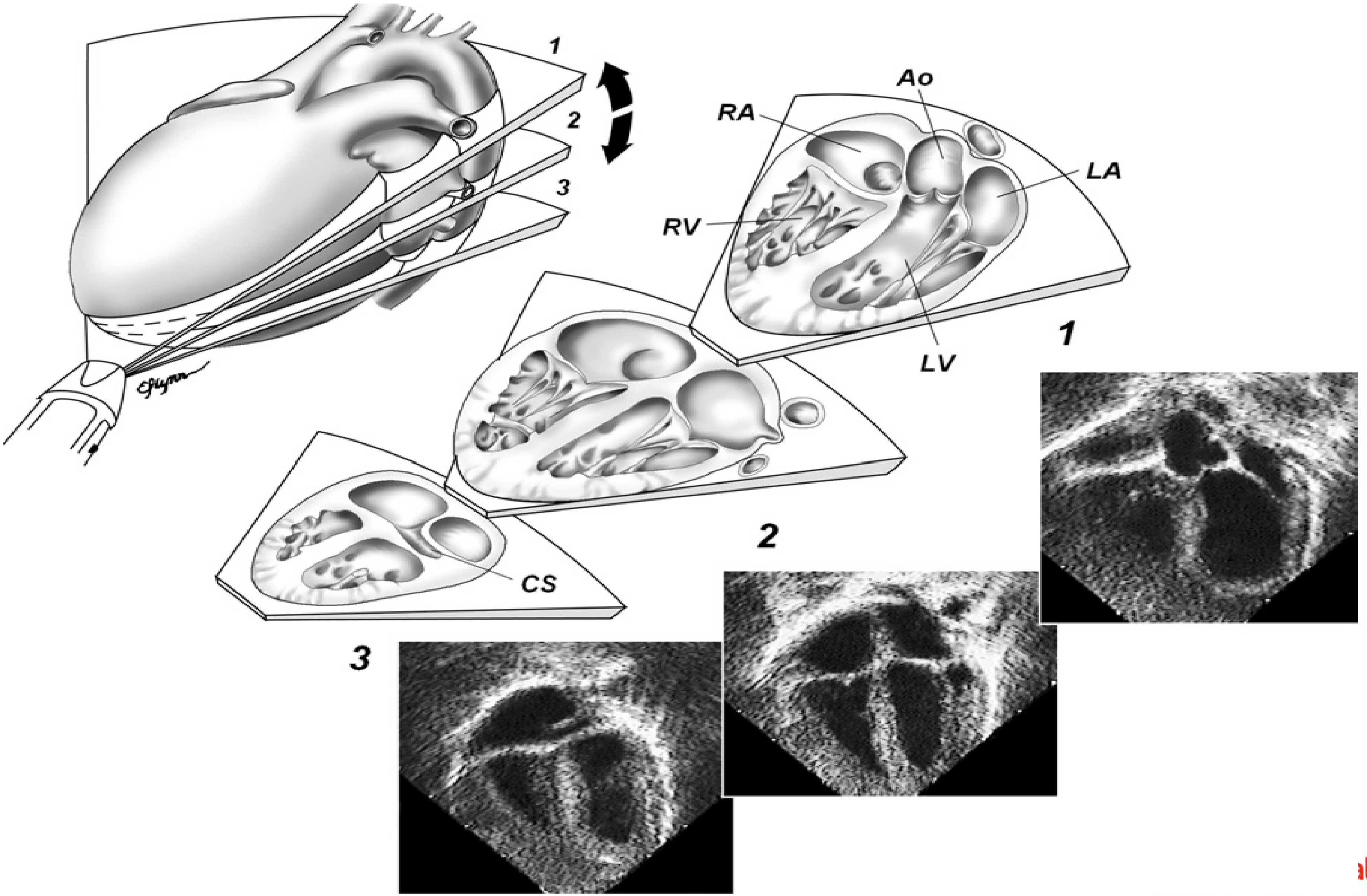
Apical views

- IVC
- LA
- RA
- Atrial Septum
- Coronary sinus
- Aortic valve
- Pulmonary valve
- Ascending Aorta

- Pulmonary veins
- MV
- TV
- LV
- RV
- Ventricular Septum
- MPA/BPA

Children's National ™

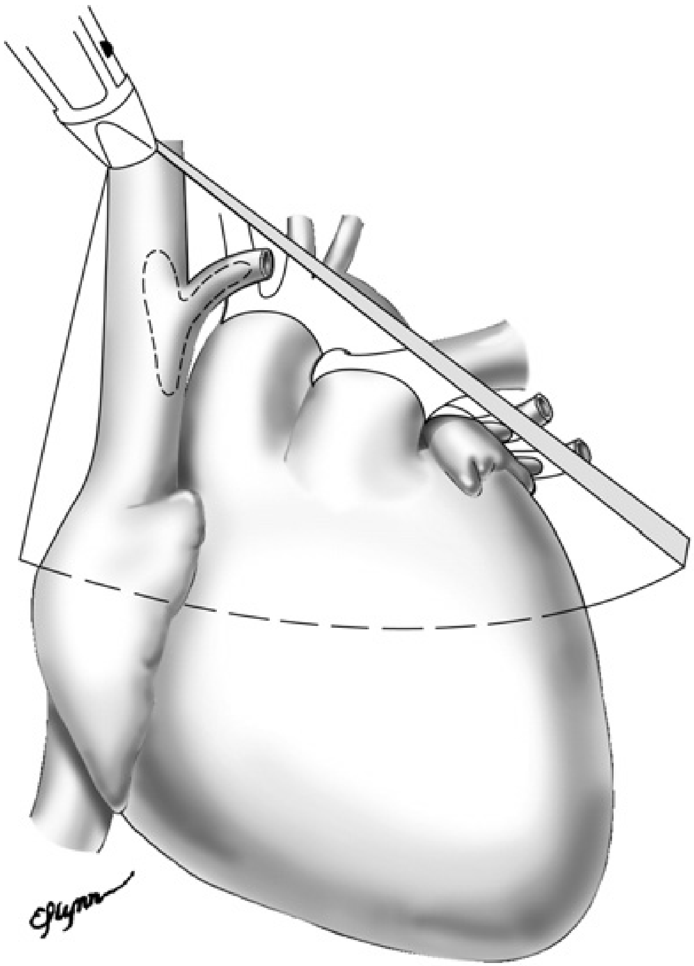
http://www.lai-echo.com/chapter4/video-4-4.asp

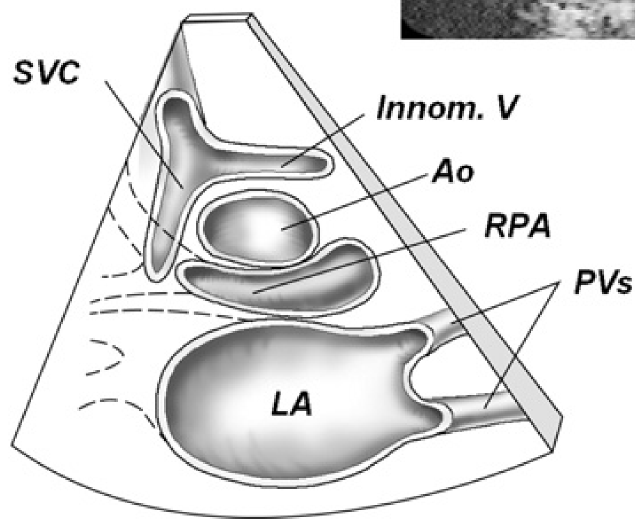


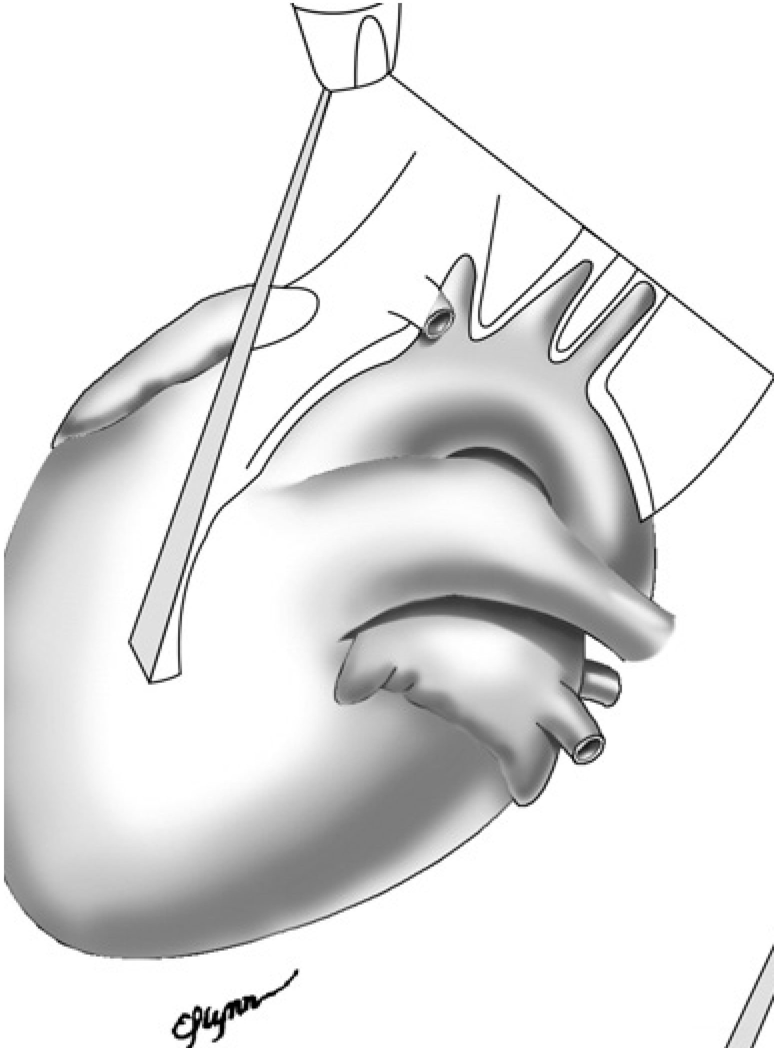
Suprasternal notch

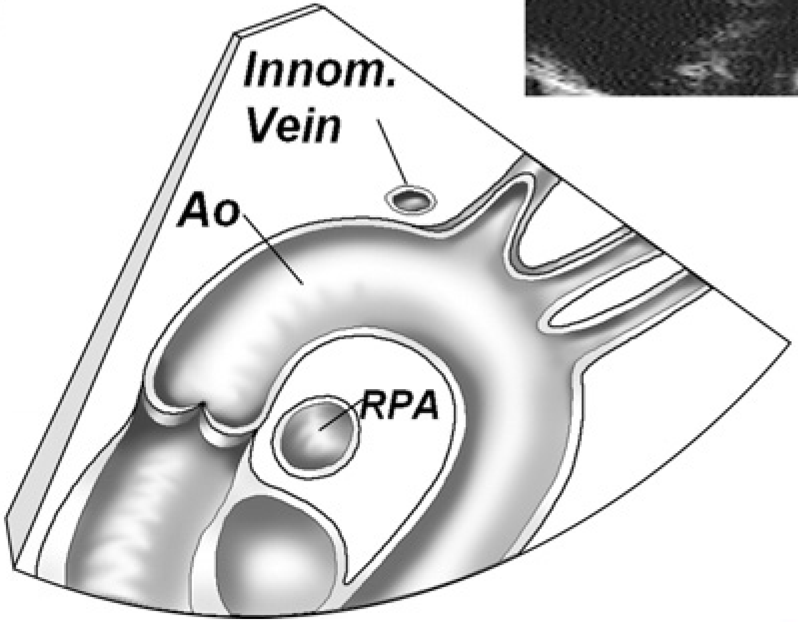
- SVC
- LA
- Pulmonary veins
- Ascending aorta
- Thoracic Aorta
- MPA/BPA
- Aortic Arch
- Left Innominate vein

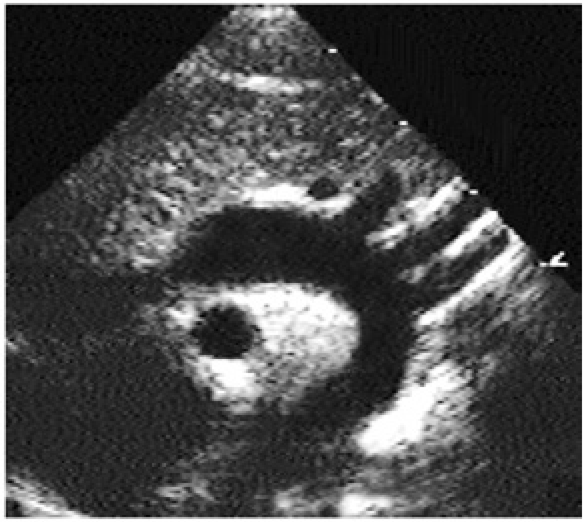
http://www.lai-echo.com/chapter4/video-4-14.asp









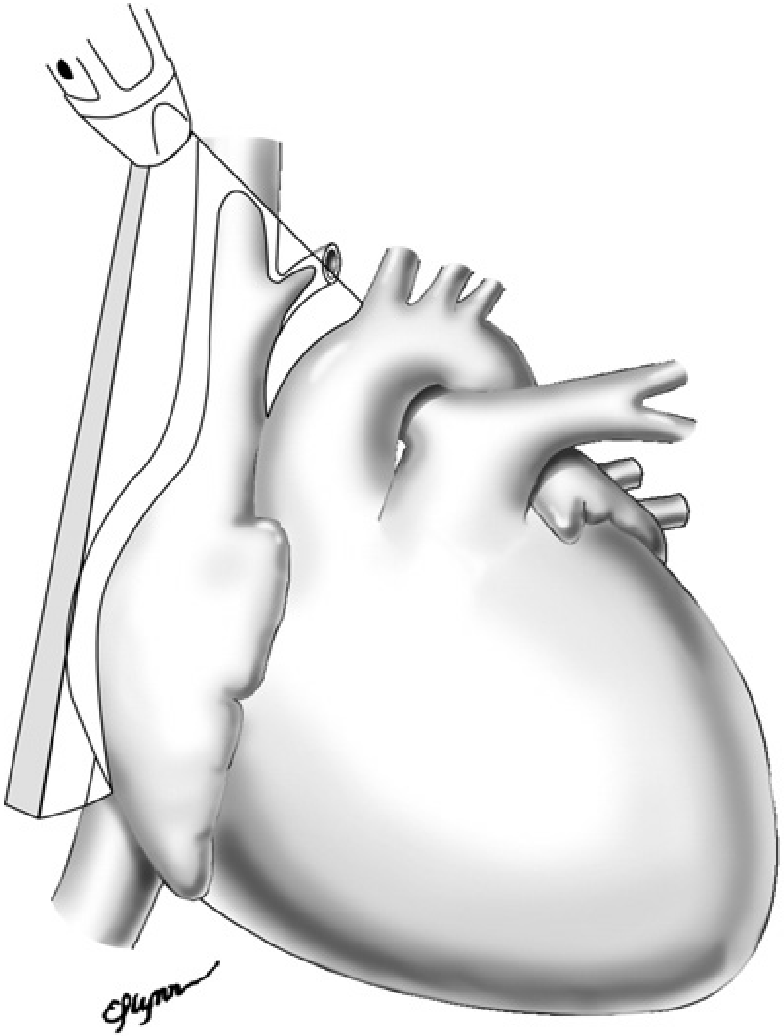


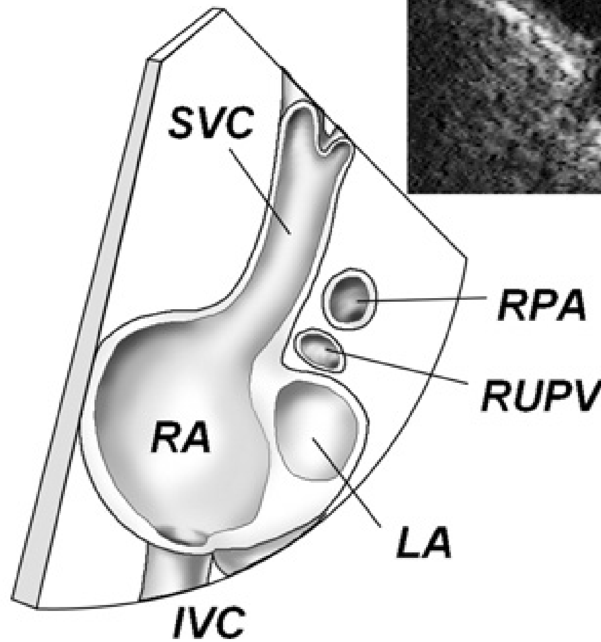
Children's National

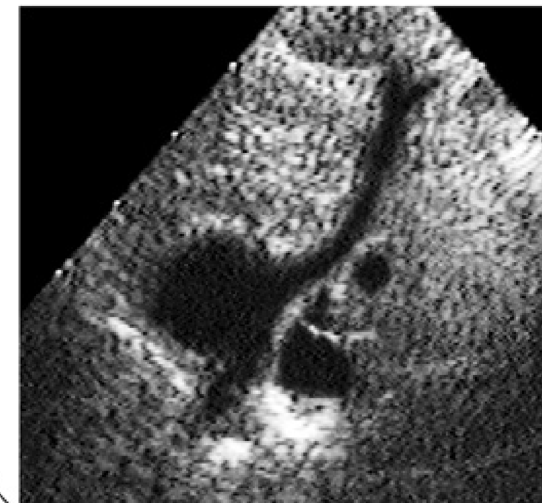
Right Parasternal

- IVC
- SVC
- RA
- Atrial septum
- Right pulmonary veins
- Ascending Aorta
- Right pulmonary artery

Children's National ™




SVC
RPA
RUPV
RA
LA
IVC



Hemodynamic Measurements

Doppler insonation angle

Pressure gradients

Bernoulli equation

- **Modified Bernoulli Equation $\Delta P = 4 \times v_2^2$**

Flow

- Qp=RVOT CSA x RVOT VTI
- Qs= LVOT CSA x LVOT VTI
- Qp/Qs= 1/1 normal , abnormal ≥ 1.5:1

PI velocity for PAEDP

Echo in CHD

Doppler echo

- Pulsed wave Doppler
  - Quantitation of intracardiac hemodynamics
    - Valvar regurgitation
    - Intracardiac shunts
    - LVOT/RVOT obstruction
  - Ventricular function
    - Systolic
    - Diastolic (mitral inflow, pulmonary venous inflow)

Children's National ™

Echo in CHD

Continuous wave Doppler

- Non-invasive measurements of mean and peak transvalvar gradients
  - Valvar stenosis
- Prediction of Ventricular Pressure (modified Bernoulli equation)
  - VSD-→ LV: RV pressure gradient
  - TR/PR→ RV, PA pressure

Children's National ™

Echo in CHD

Color Doppler

- Direction of cardiac flow
  - TAPVR vs. LSVC
- Velocity and Turbulence of cardiac flow
  - Conduit obstruction
  - Identification of intracardiac shunts
    - VSD, PDA, ASD
  - Assessment of Post-op CHD
    - Shunt patency, residual intracardiac shunt

Morphologic/Segmental approach

Define morphologic—not spatial—anatomy

- Which atrium is the Right? Left?
- Which ventricle is the Right? Left?
- Which great artery is which?

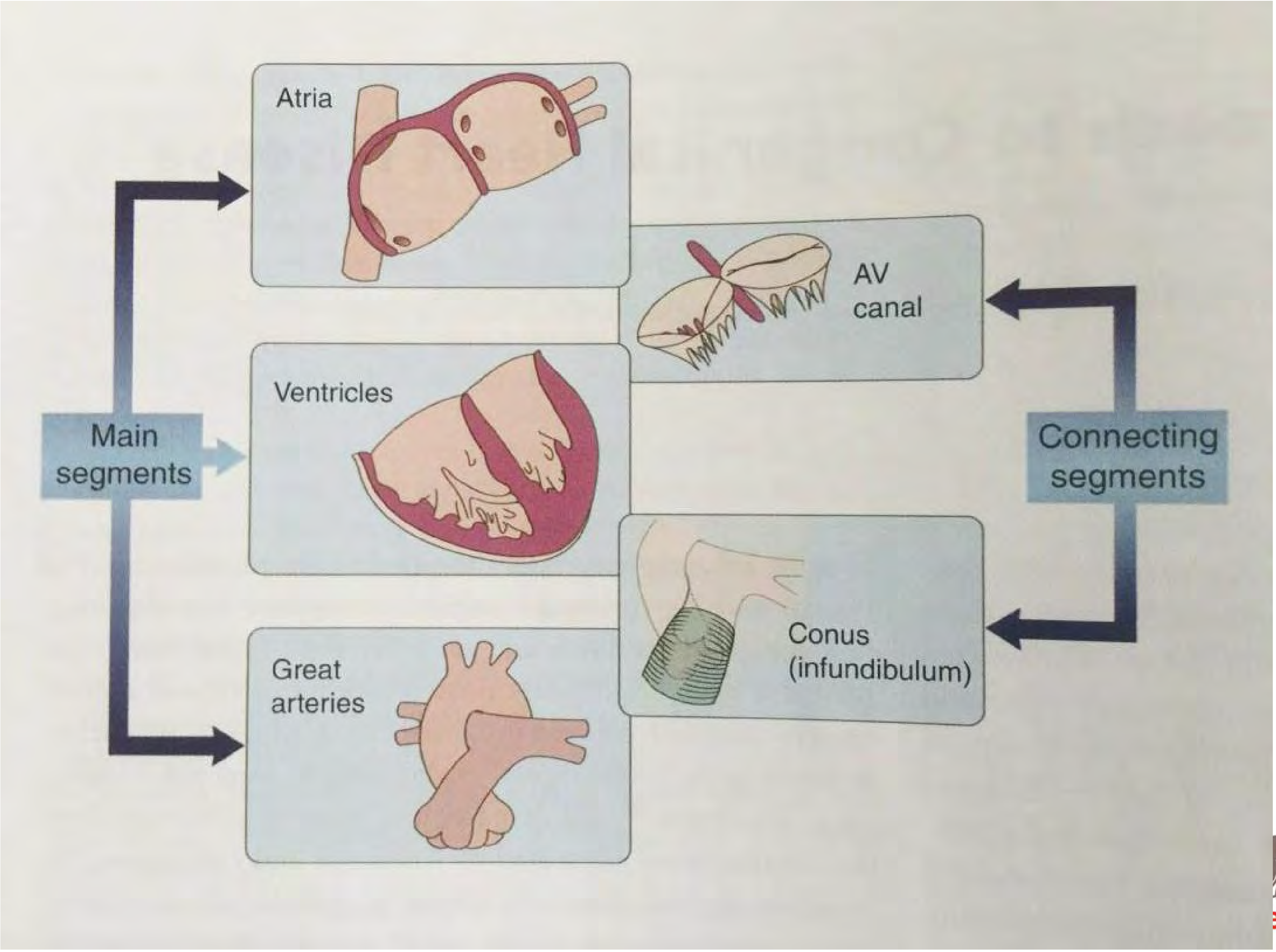
Define segmental anatomy

- Segments: Atrium, Ventricles, Great Arteries
- What is the position of each segment relative to each other?
  - Is the RA on the right? Is it connected to the RV? Is it connected to the PA?
  - Is the LA on the left? Is it connected to the LV? Is it connected to the Aorta?

Predict the physiology

- What is the physiology predicted by the segmental connections?
  - Normal? Transposition? Obstructed flow?
- What is the physiology predicted by flow in the ductus? Across the foramen?

The Cardiac Segments



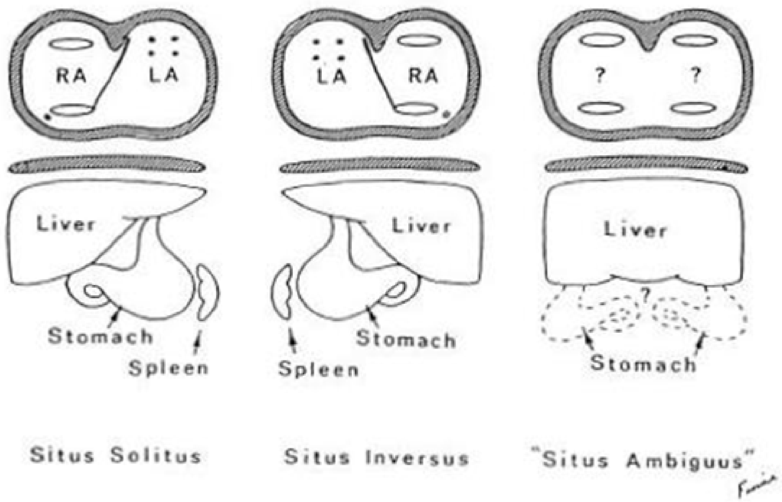
Abdominal and Atrial Situs

Cardiac position

- Levocardia, Dextrocardia, Mesocardia, Dextroposition

Situs abnormalities

- Inversus
  - Not often associated with CHD
- Ambiguous
  - Heterotaxy syndromes
    - Asplenia/polysplenia
    - Abdominal malrotation
    - Cardiac defect
      - » AV canal defect
      - » Conotruncal defects
      - » Systemic and pulmonary venous anomalies



The Endocardial Cushion

Define the connections

- Concordant: RA to RV, LA to LV
- Discordant: RA to LV, LA to RV
- Common inlet: AV canal defect
- Atretic inlet: mitral, tricuspid valve atresia
- Double inlet

Assess AV valve anatomy and function

- Morphology
  - Ebstein’s tricuspid valve, parachute mitral valve
  - Hypoplastic
- Physiology
  - Stenosis
  - Atresia
  - Insufficiency

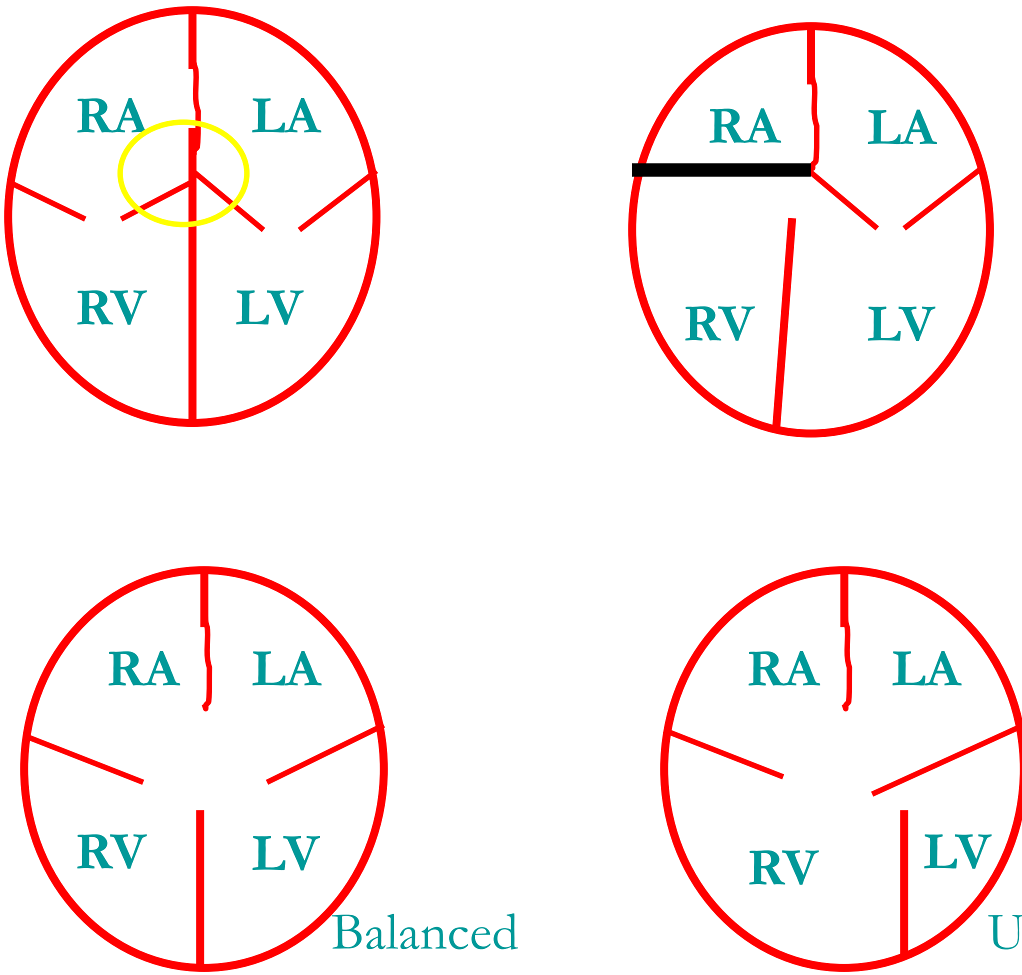
Children’s National™

The Endocardial Cushion

Normal

Atretic AV inlet

Common AV inlet



Balanced

Unbalanced

The Ventricles

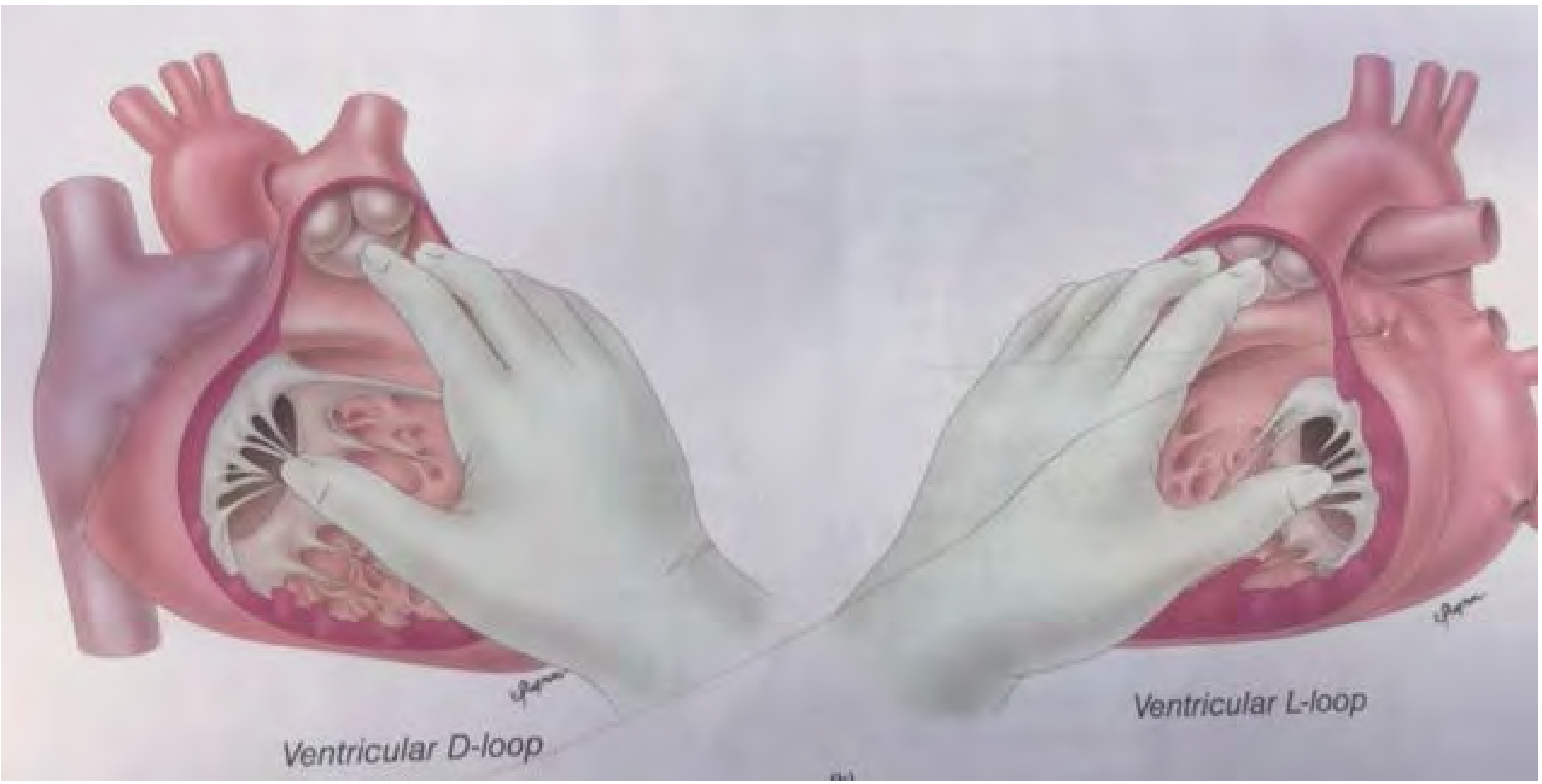
The Right Ventricle

- Coarsely trabeculated
- Moderator band
- "Septophilic" tricuspid valve chordal insertions

The Left Ventricle

- Finely trabeculated
- 2 prominent MV papillary muscles
- No septal attachments of valve

Children's National™


Ventricular D-loop
Ventricular L-loop

The Great Arteries

Identify the great arteries:

- Aorta
  - Coronary artery origins
  - Origin of brachiocephalic vessels from arch
  - “Candy cane?”
- Pulmonary artery
  - Proximal bifurcation into branch PAs
  - No brachiocephalic vessel from the ductal arch
  - “Hockey stick?”

How many outlets?

- One? = truncus arteriosus or semilunar valve atresia
- Two? Are they normal? In position? In size?

Do the great arteries arise from the correct ventricles?

- Aorta from LV, PA from RV = solitus (normal) GA
- Aorta from RV, PA from LV = transposition of the GA
- Both from RV = DORV

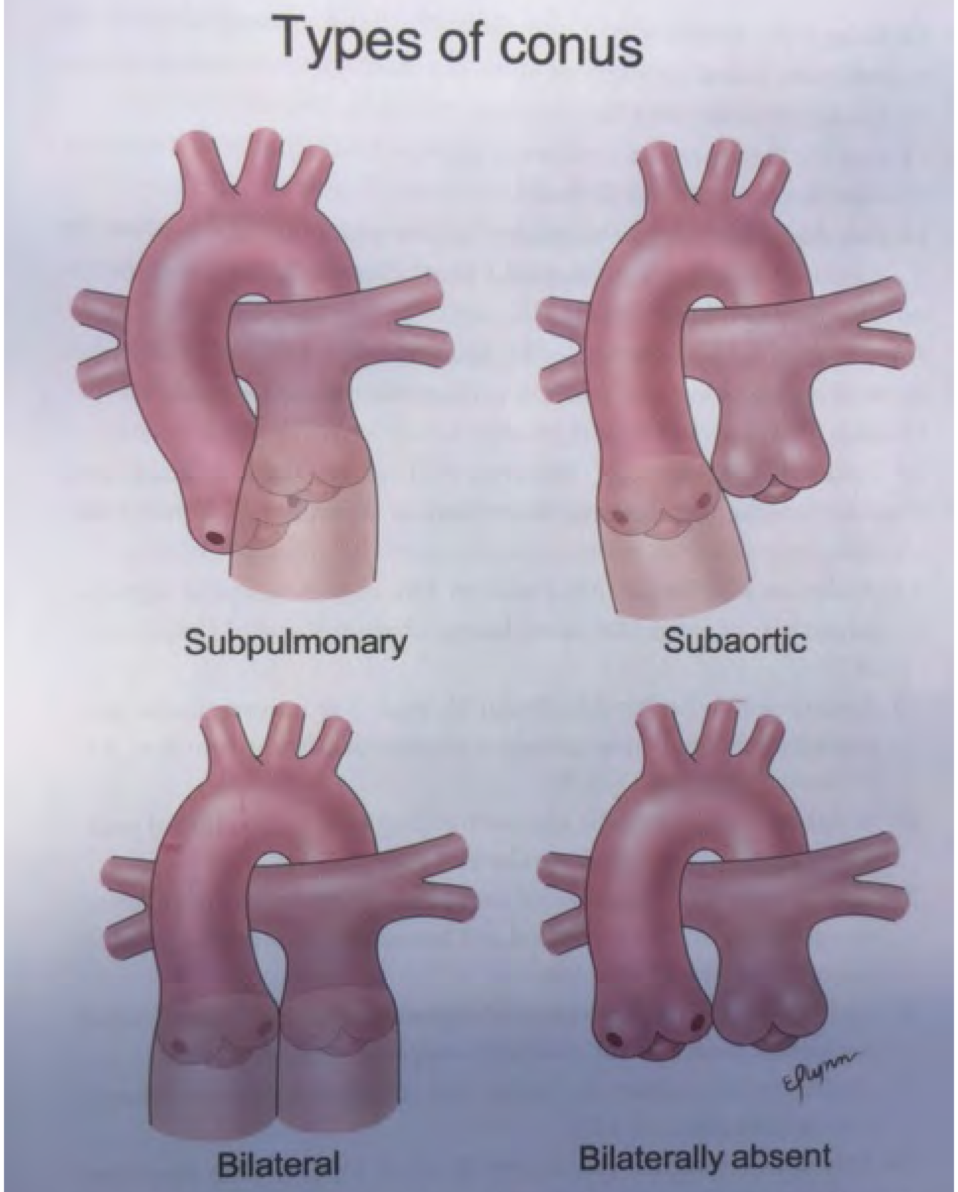
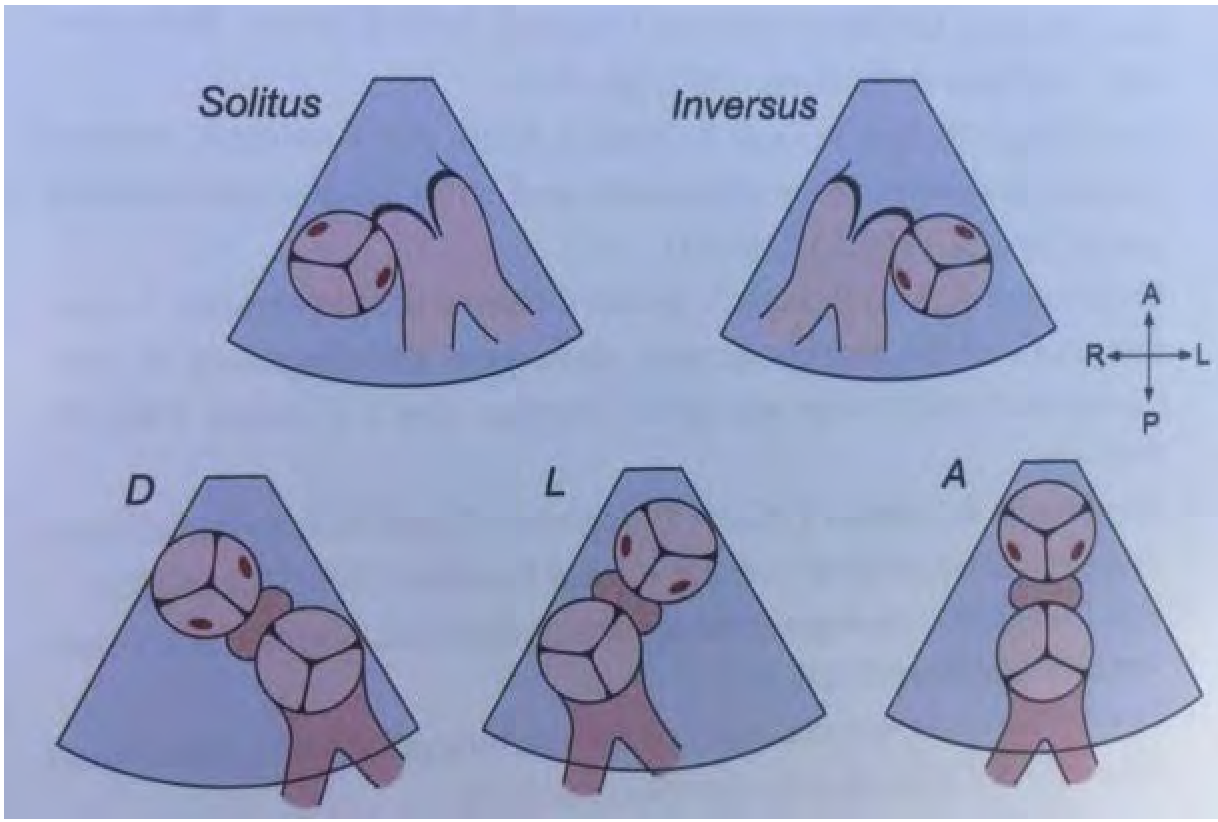
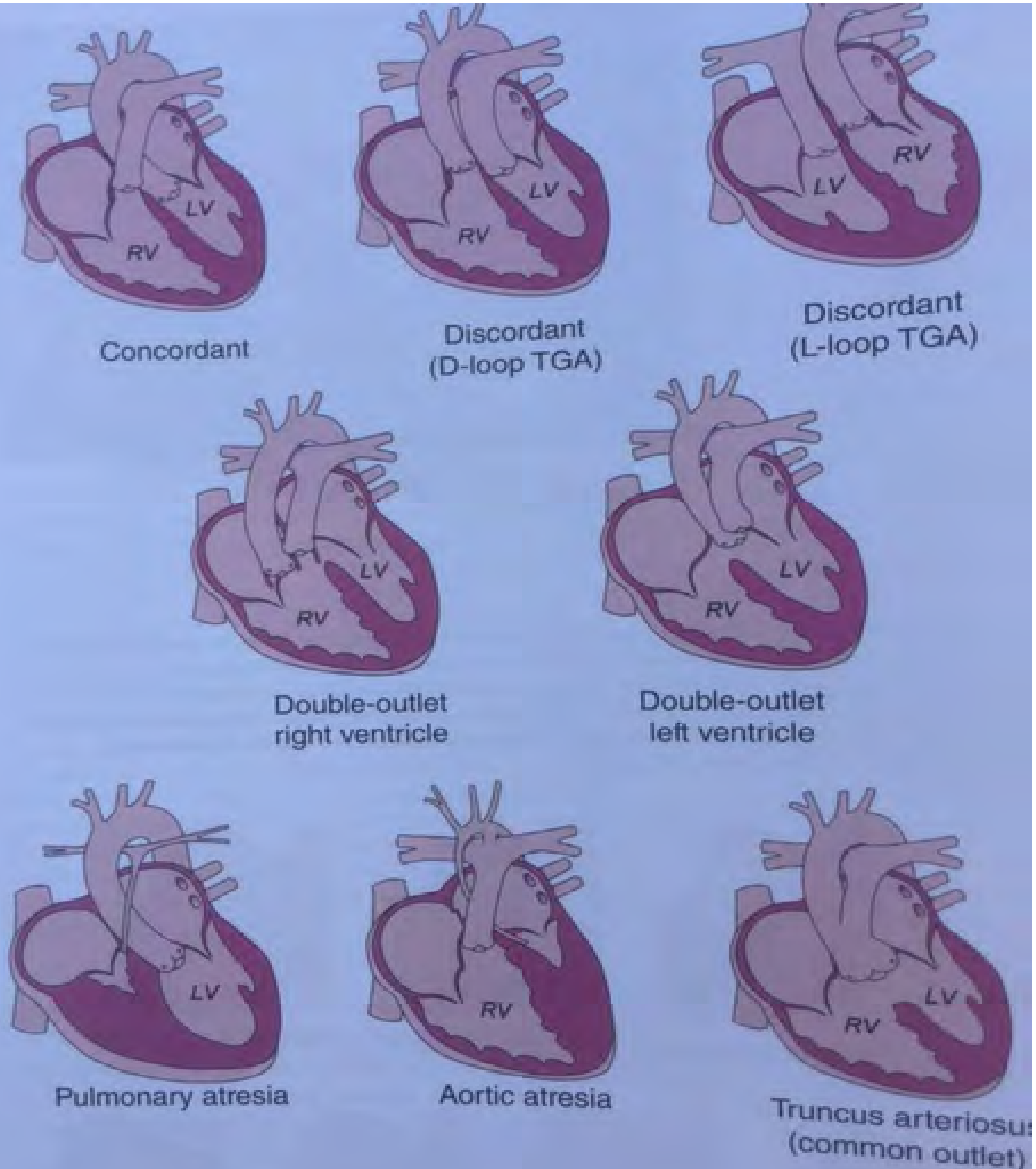


Figure 3.13 Type of infundibulum. In general, there are four types of conus


Solitus
Inversus
A
R
L
P
D
L
A



the aorta and biventricular origin of the main pul-

Common Lesions

Atrial Septal Defects

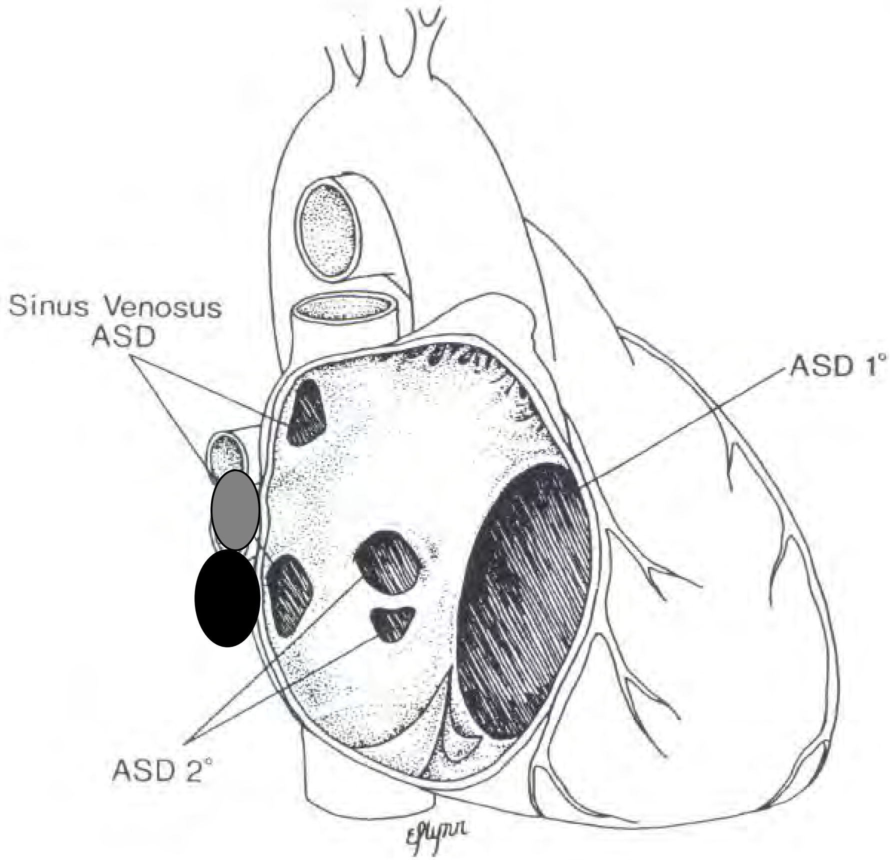
Secundum ASD

Primum ASD

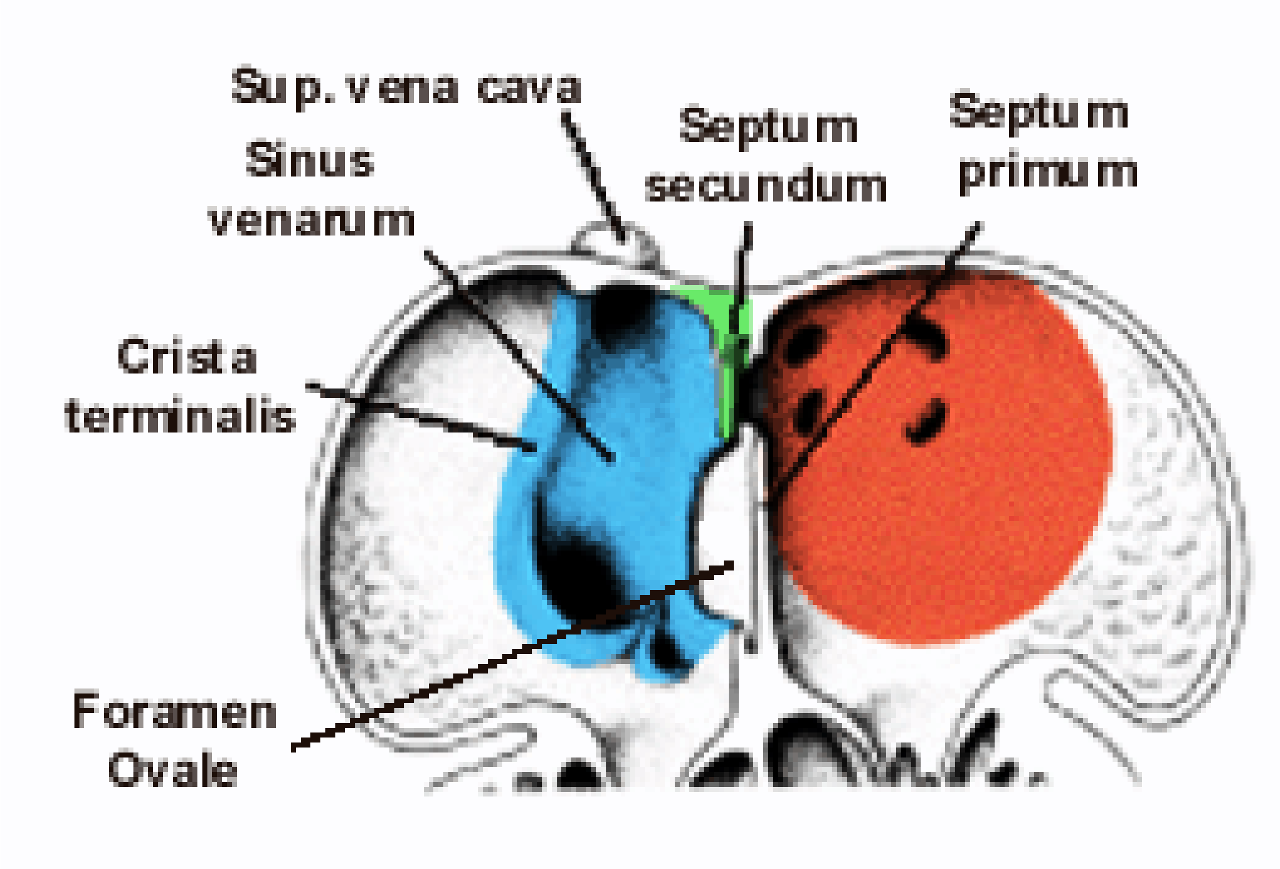
Sinus Venosus defect

- Not truly a deficiency of tl same physiology as an A

Common atrium



Atrial Septal Development



http://www.med.unc.edu/embryo_images/unit-welcome/welcome_htms/contents.htm

ASD: Clinical Correlation

Usually diagnosed in childhood

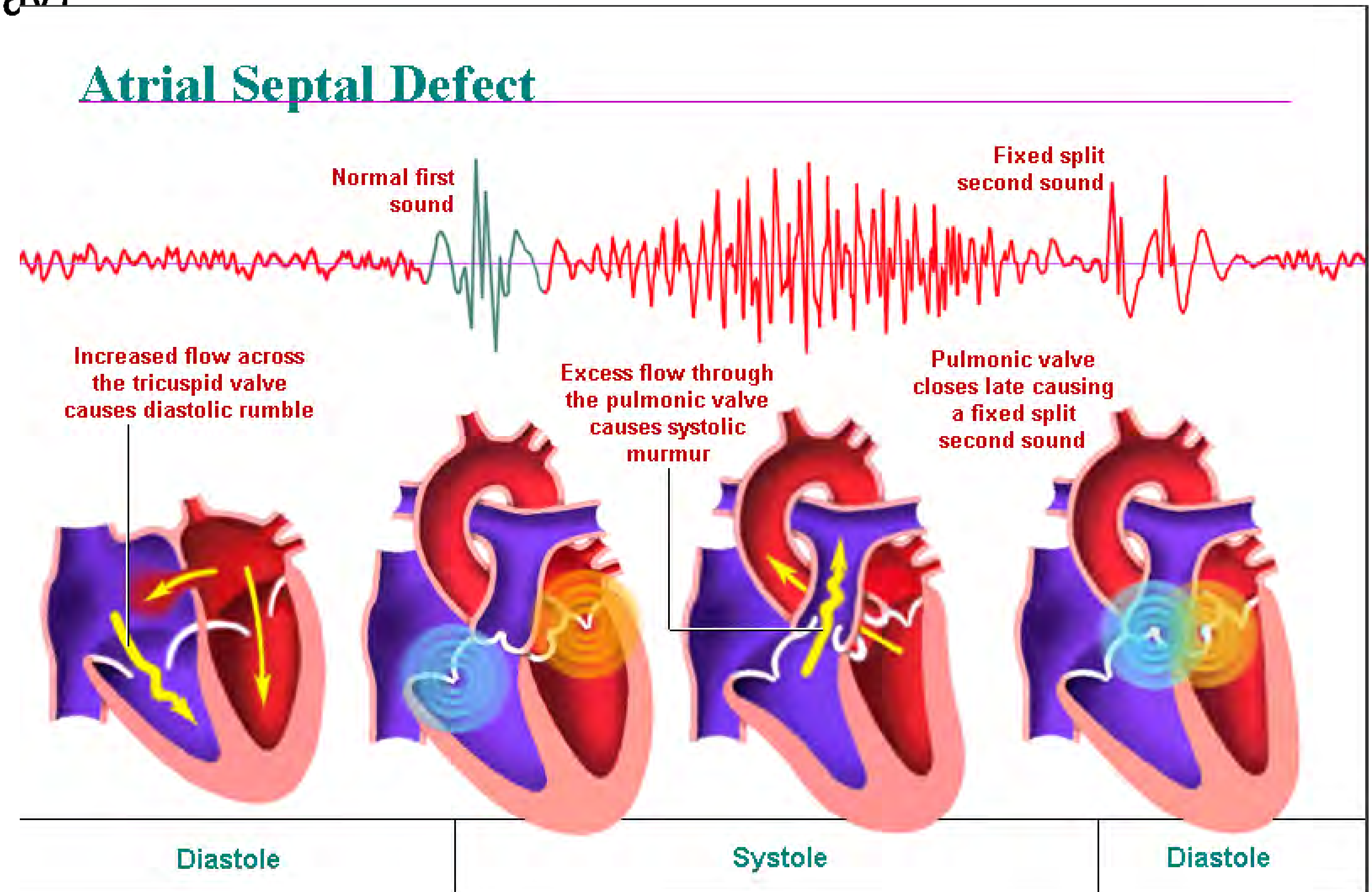
Asymptomatic

F>M

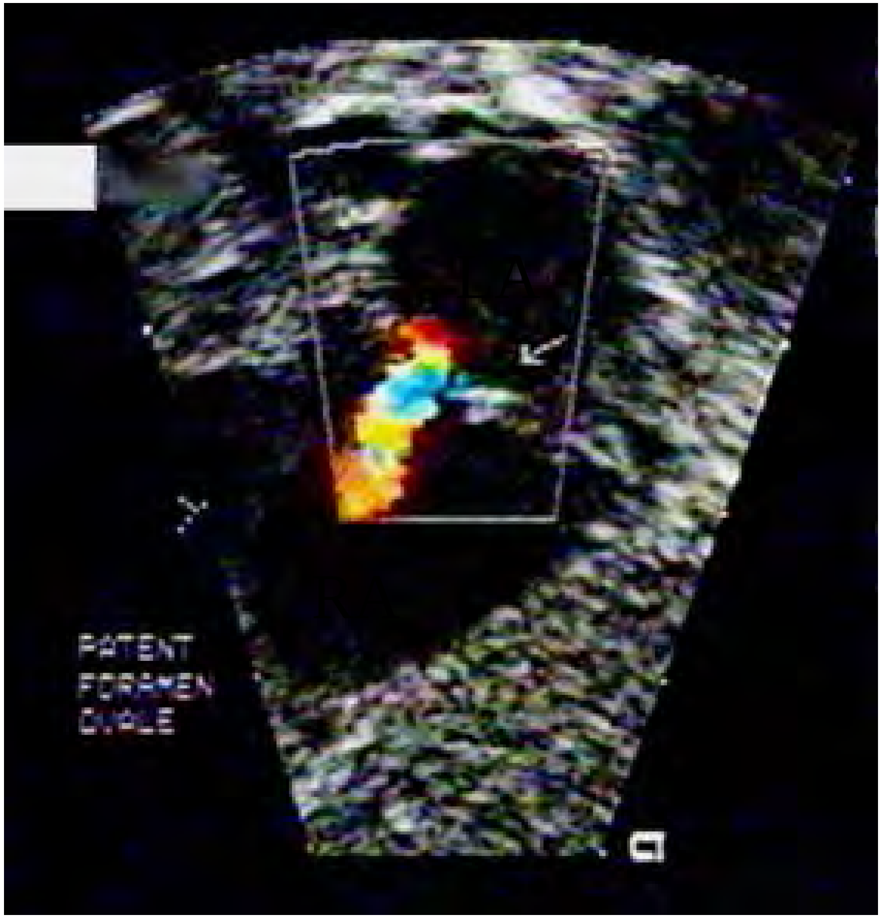
Systolic ejection murmur and widely split fixed S_2

EKG may show RBBB or RVH

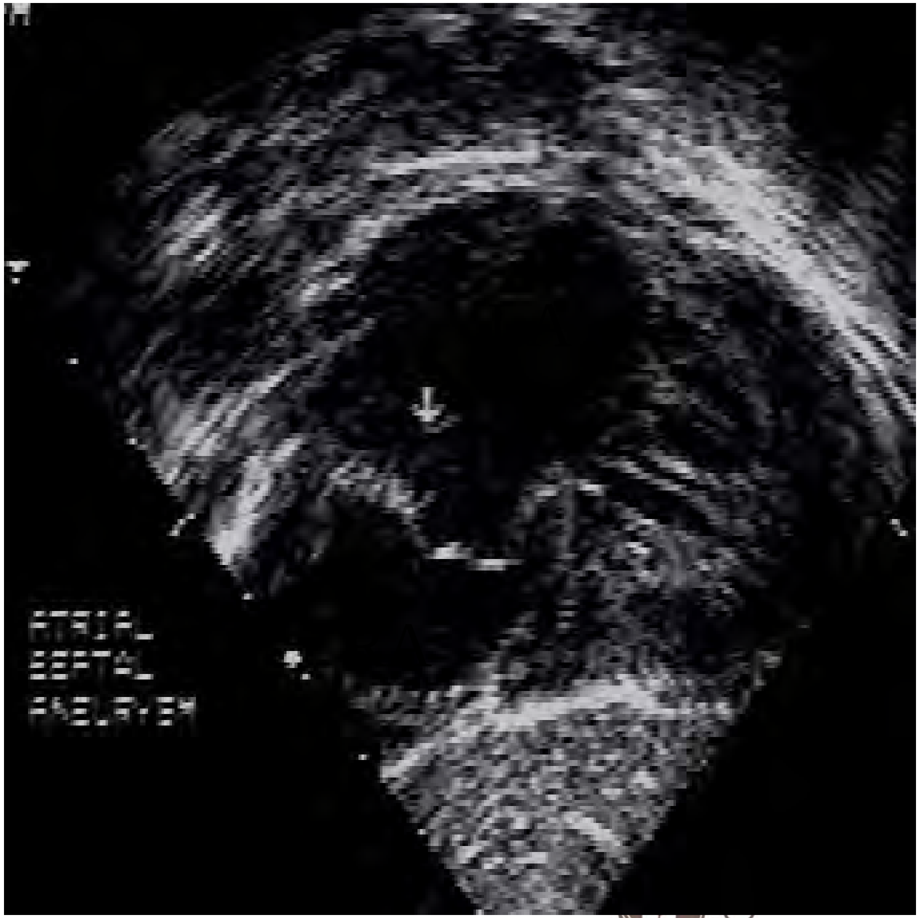
(.wav)


Atrial Septal Defect
Normal first sound
Fixed split second sound
Increased flow across the tricuspid valve causes diastolic rumble
Excess flow through the pulmonic valve causes systolic murmur
Pulmonic valve closes late causing a fixed split second sound
Diastole
Systole
Diastole

Atrial Septum
Normal Variants

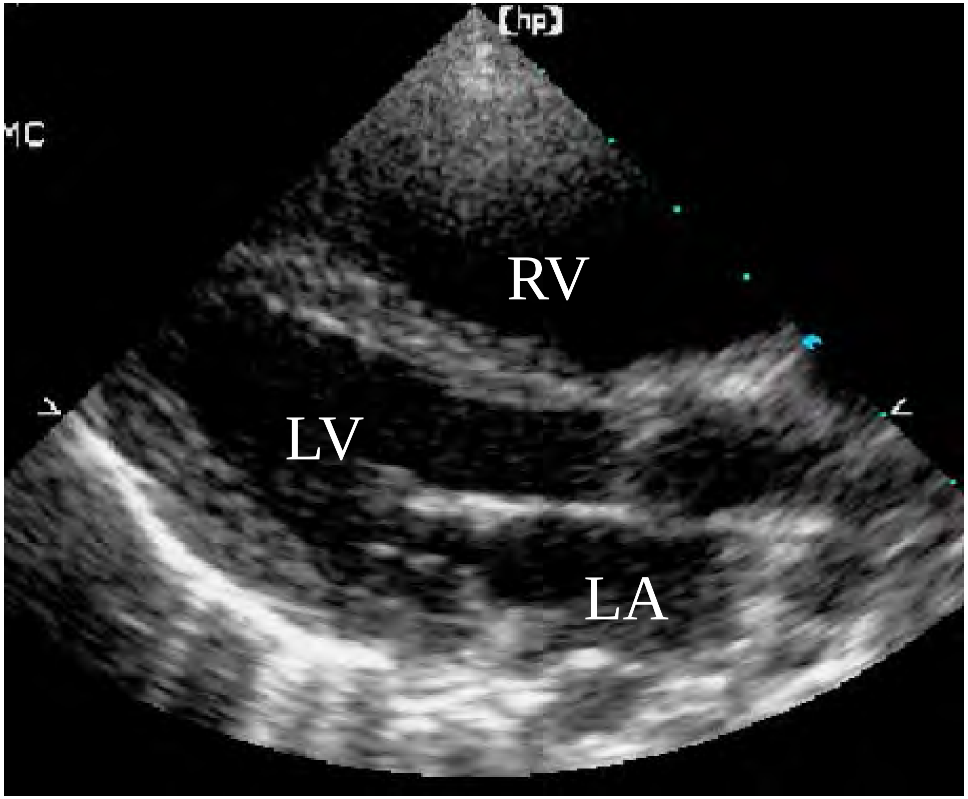


Patent Foramen Ovale

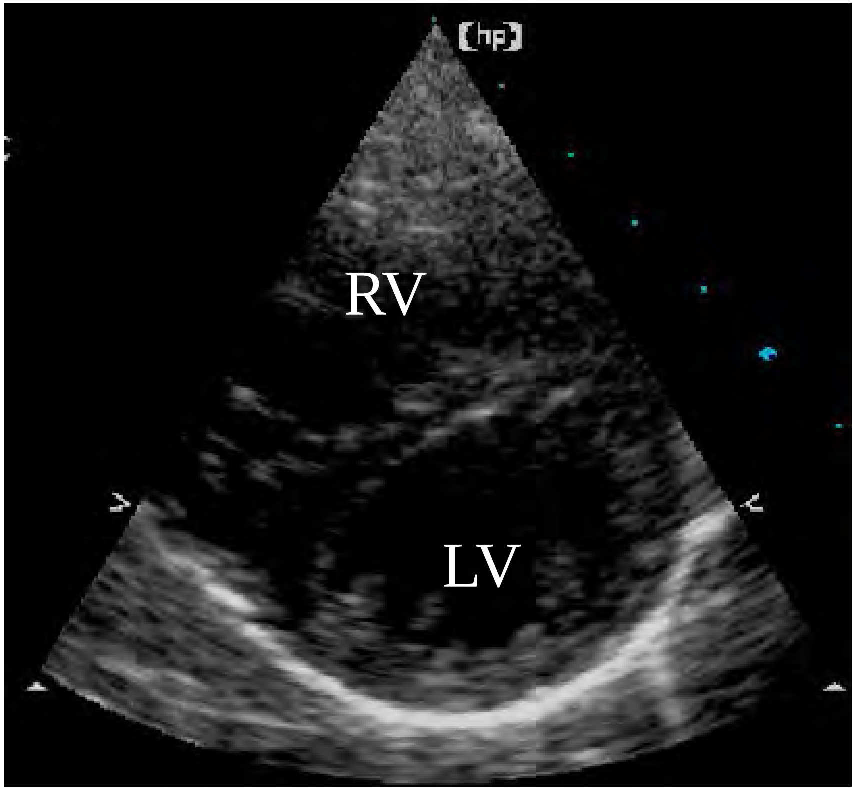


Atrial Septal Aneurysm

Secundum ASD



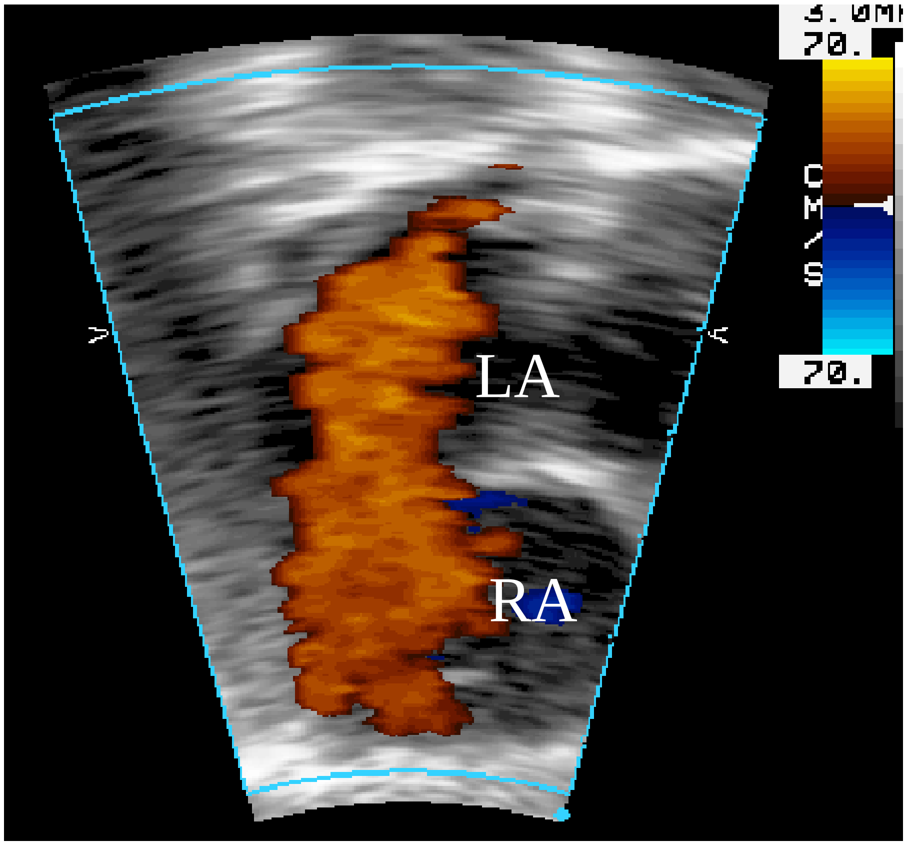
RV Dilation



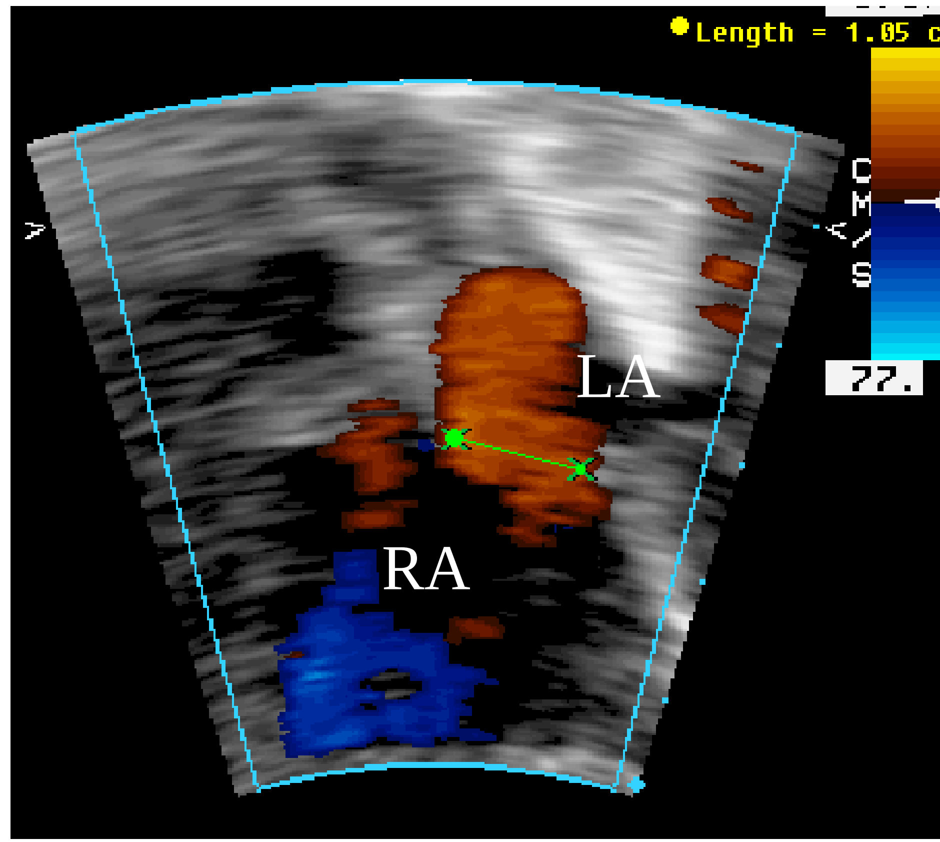
Diastolic Septal Flattening

Children's National

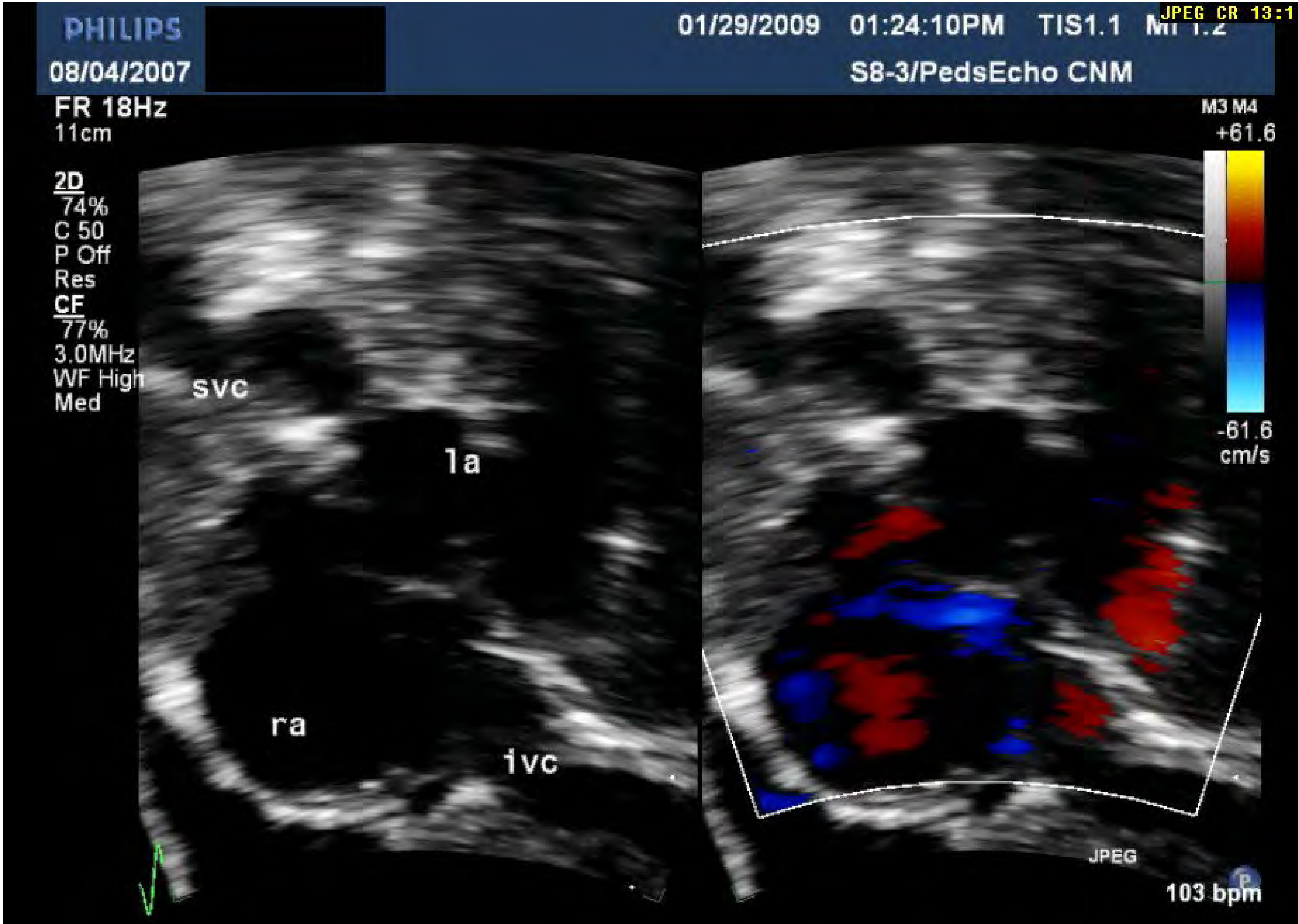
Secundum ASD



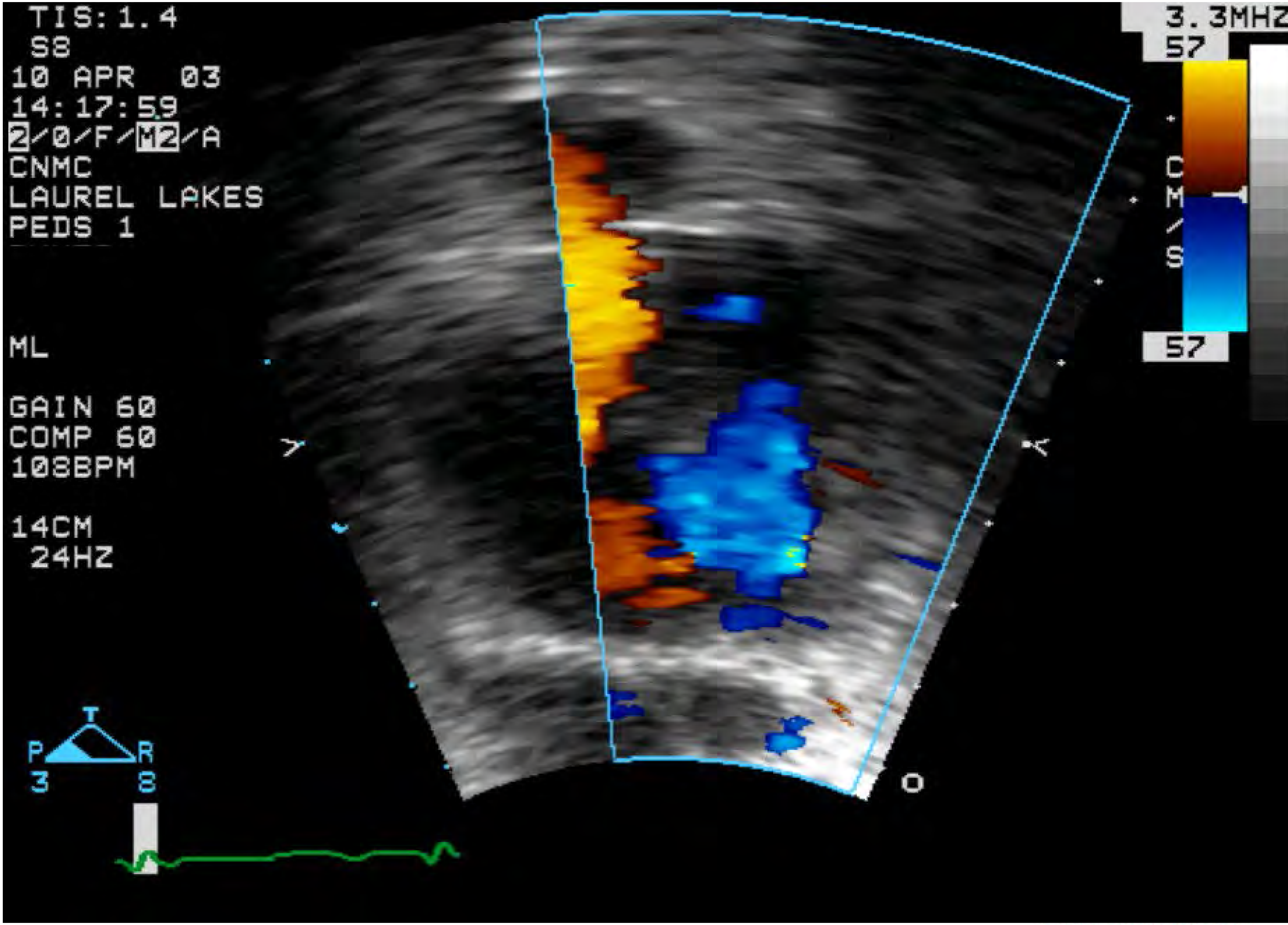
Subcostal Coronal



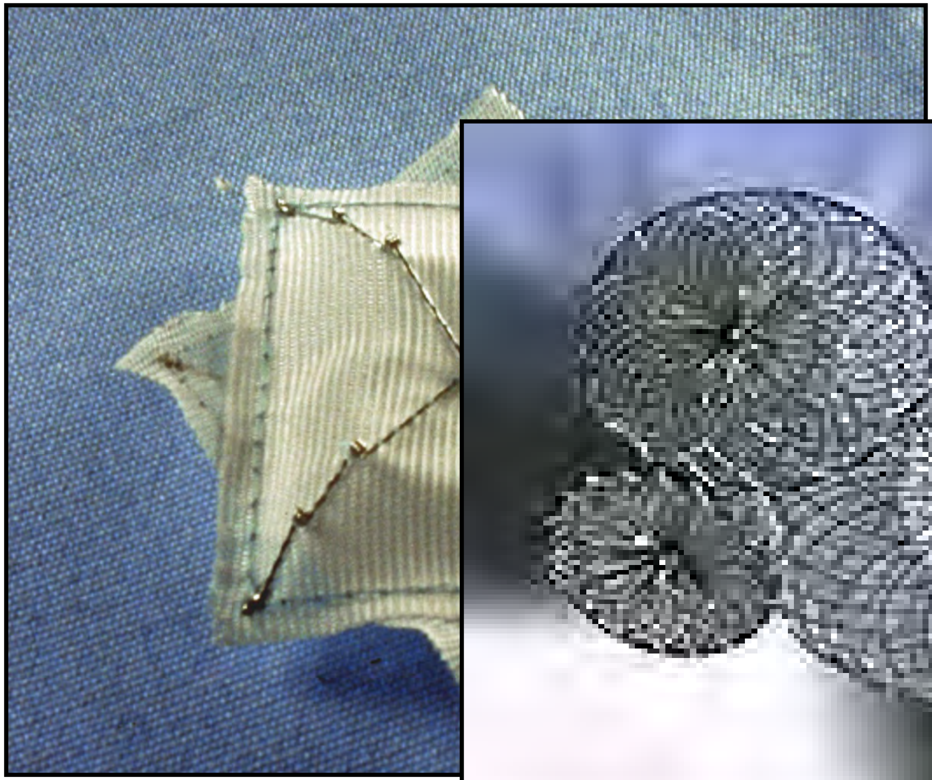
Subcostal Sagittal

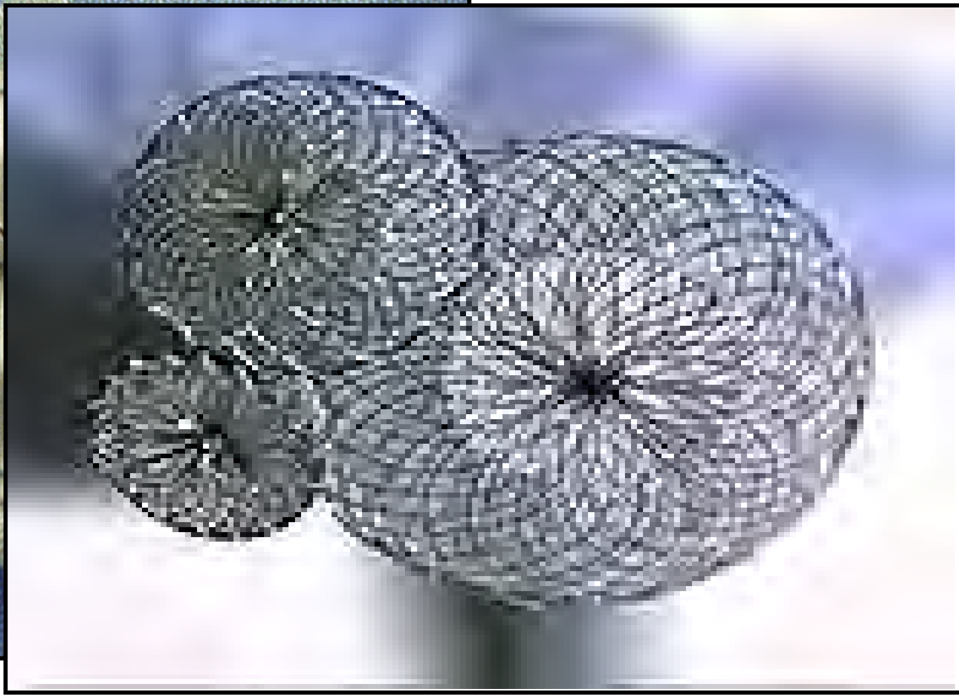


PHILIPS
08/04/2007
[redacted]
01/29/2009 01:24:10PM TIS1.1
JPEG CR 13:1
S8-3/PedsEcho CNM
FR 18Hz
11cm
2D
74%
C 50
P Off
Res
CF
77%
3.0MHz
WF High
Med
svc
la
ra
ivc
M3 M4
+61.6
-61.6
cm/s
JPEG
103 bpm


TIS:1.4
S8
10 APR 03
14:17:59
2/0/F/M2/A
CNMC
LAUREL LAKES
PEDS 1
ML
GAIN 60
COMP 60
108BPM
14CM
24HZ
P
T
R
3
8
3.3MHZ
57
C
M
/
S
57
0

Devices for ASD Closure

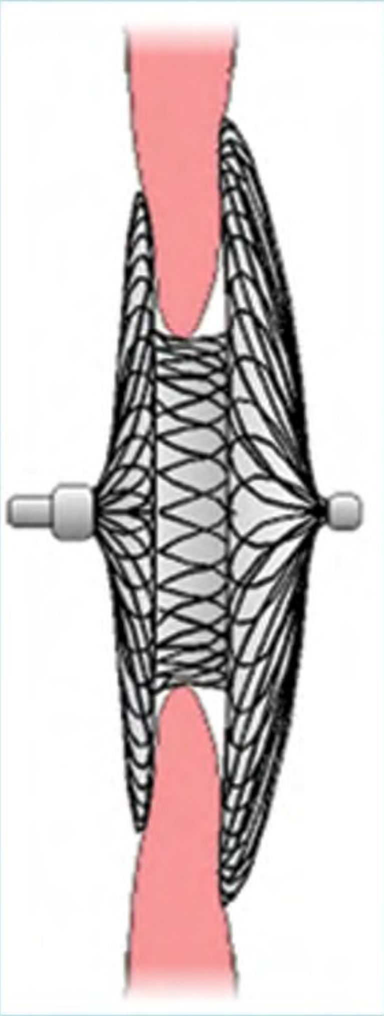


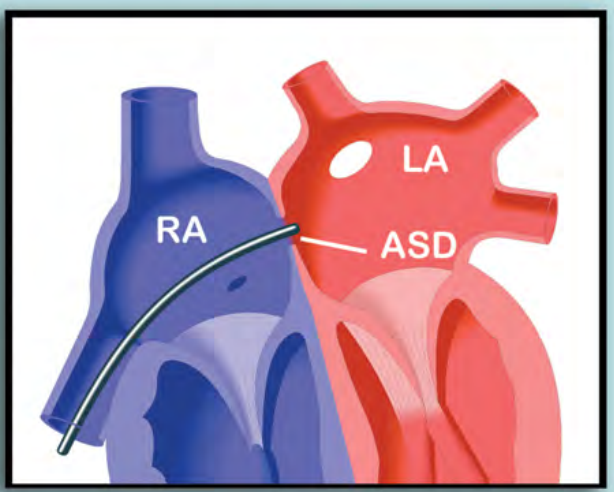


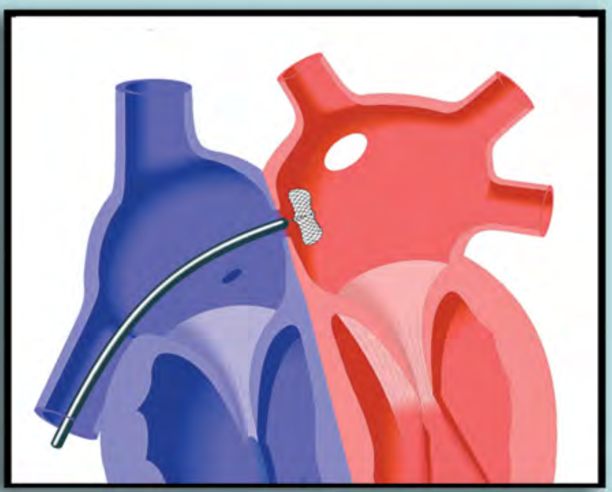
Cardio-SEAL

Amplatzer

Amplatzer Occlusion of Atrial Septal Defect

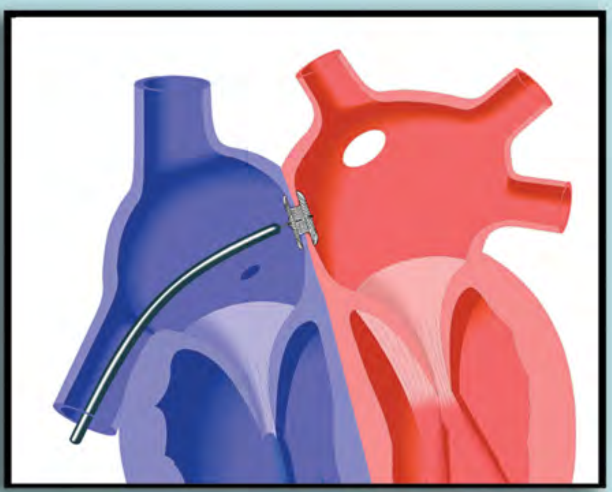




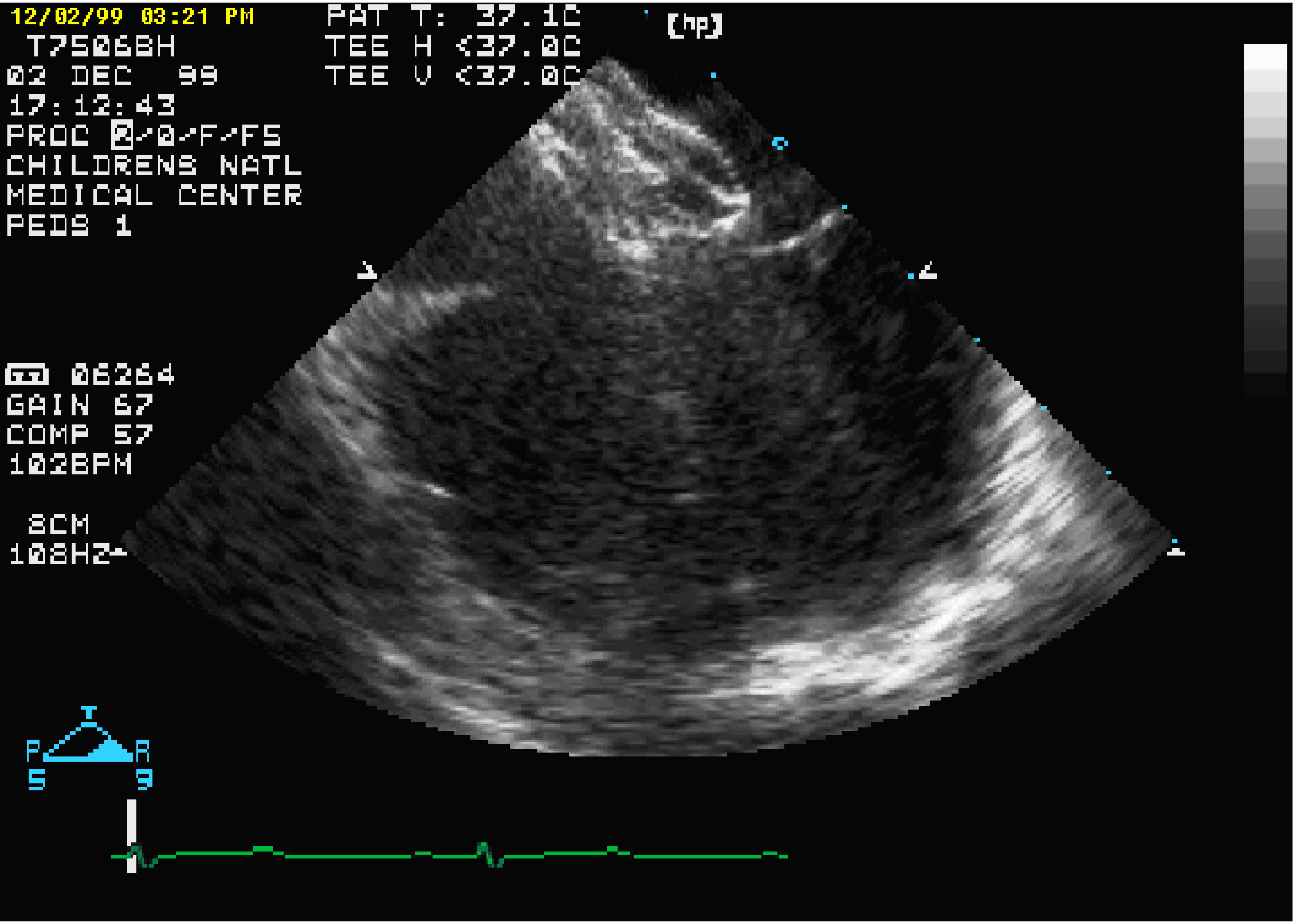


Clockwise from above: Transcatheter delivery of Amplatzer device, which is positioned across the atrial septal defect

Left: Amplatzer device in place



ASD device


12/02/99 03:21 PM
T7506BH
02 DEC 99
17:12:43
PROC 2/0/F/F5
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
PAT T: 37.1C
TEE H <37.0C
TEE V <37.0C
06264
GAIN 67
COMP 57
102BPM
8CM
108HZ
P
R
5
9

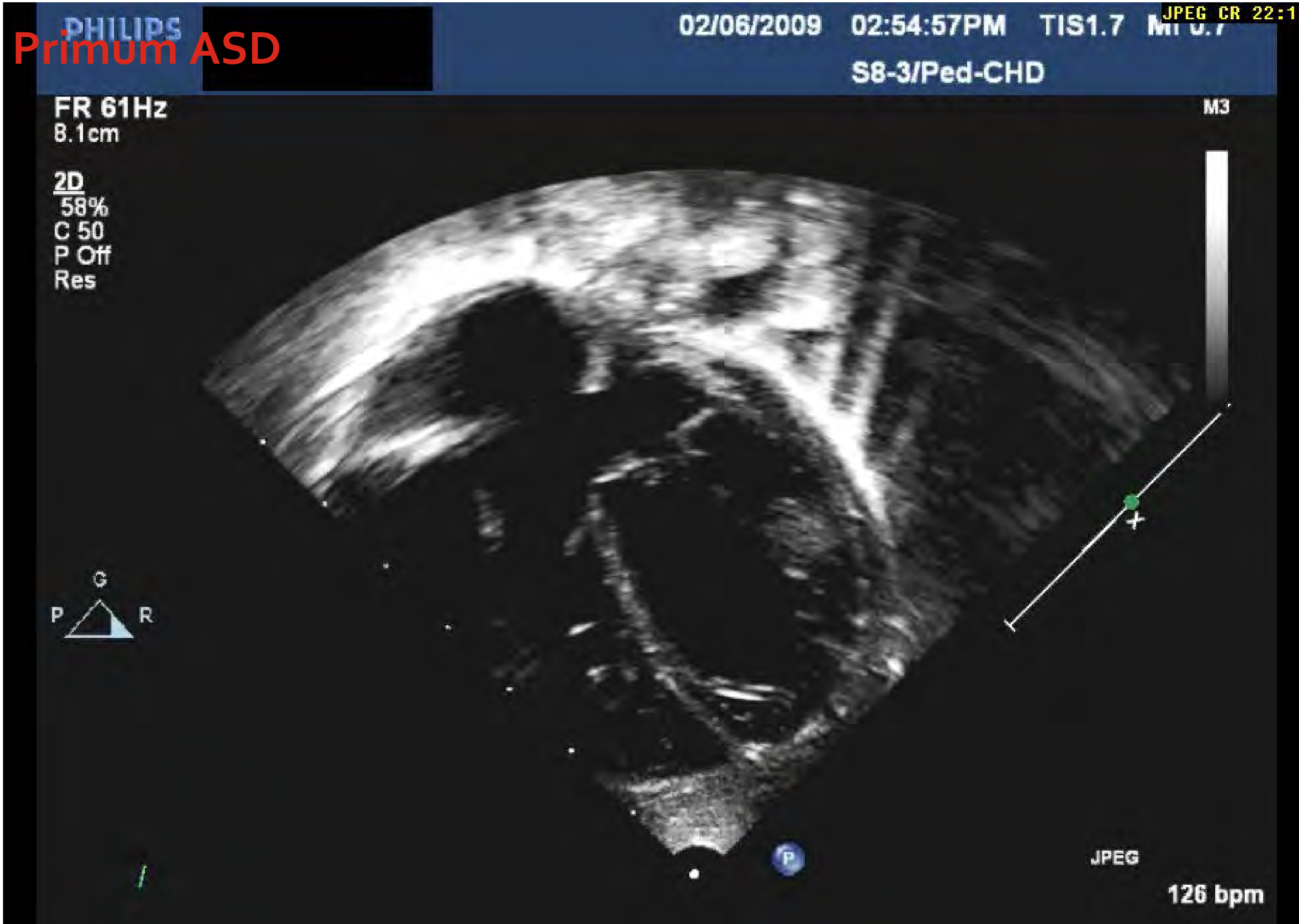
Primum ASD

Part of spectrum of AV canal defects

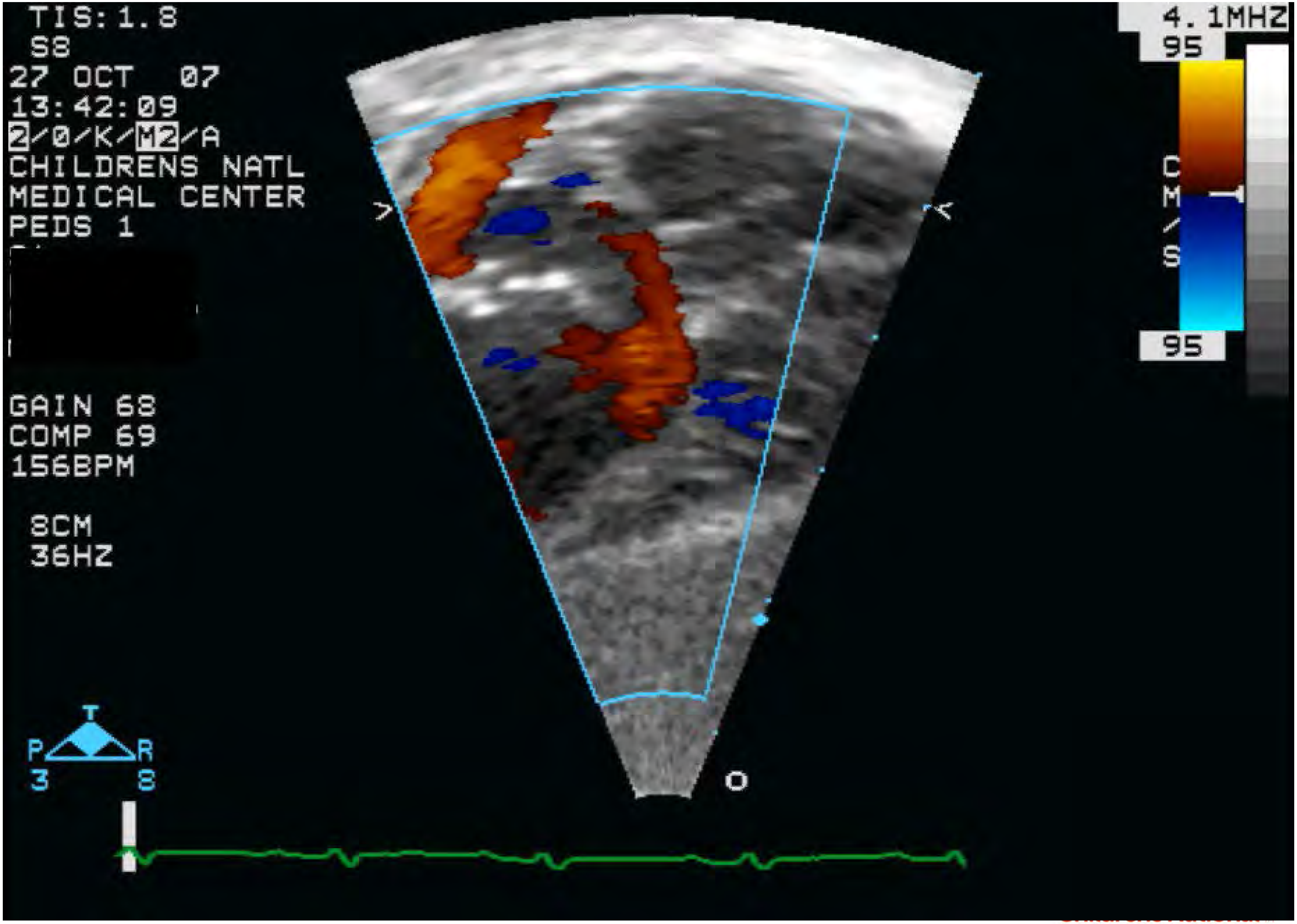
Defect is contiguous with AV valves

Associated with cleft mitral valve

Primum ASD



Children's National

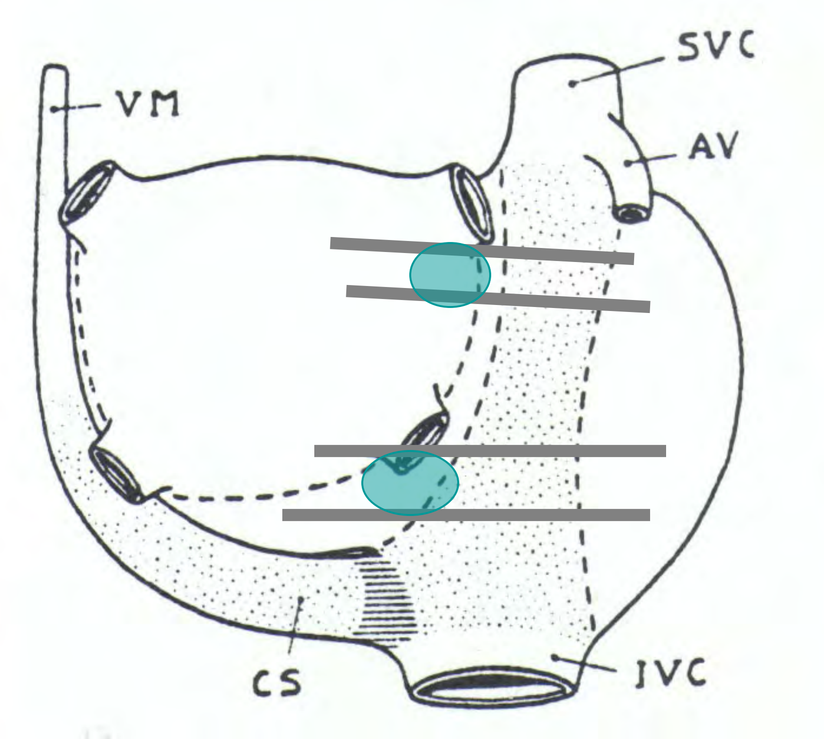

TIS:1.8
S8
27 OCT 07
13:42:09
2/0/K/M2/A
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
[redacted]
GAIN 68
COMP 69
156BPM
8CM
36HZ
T
P
R
3
8
4.1MHZ
95
CM/S
95

Sinus Venosus Defects

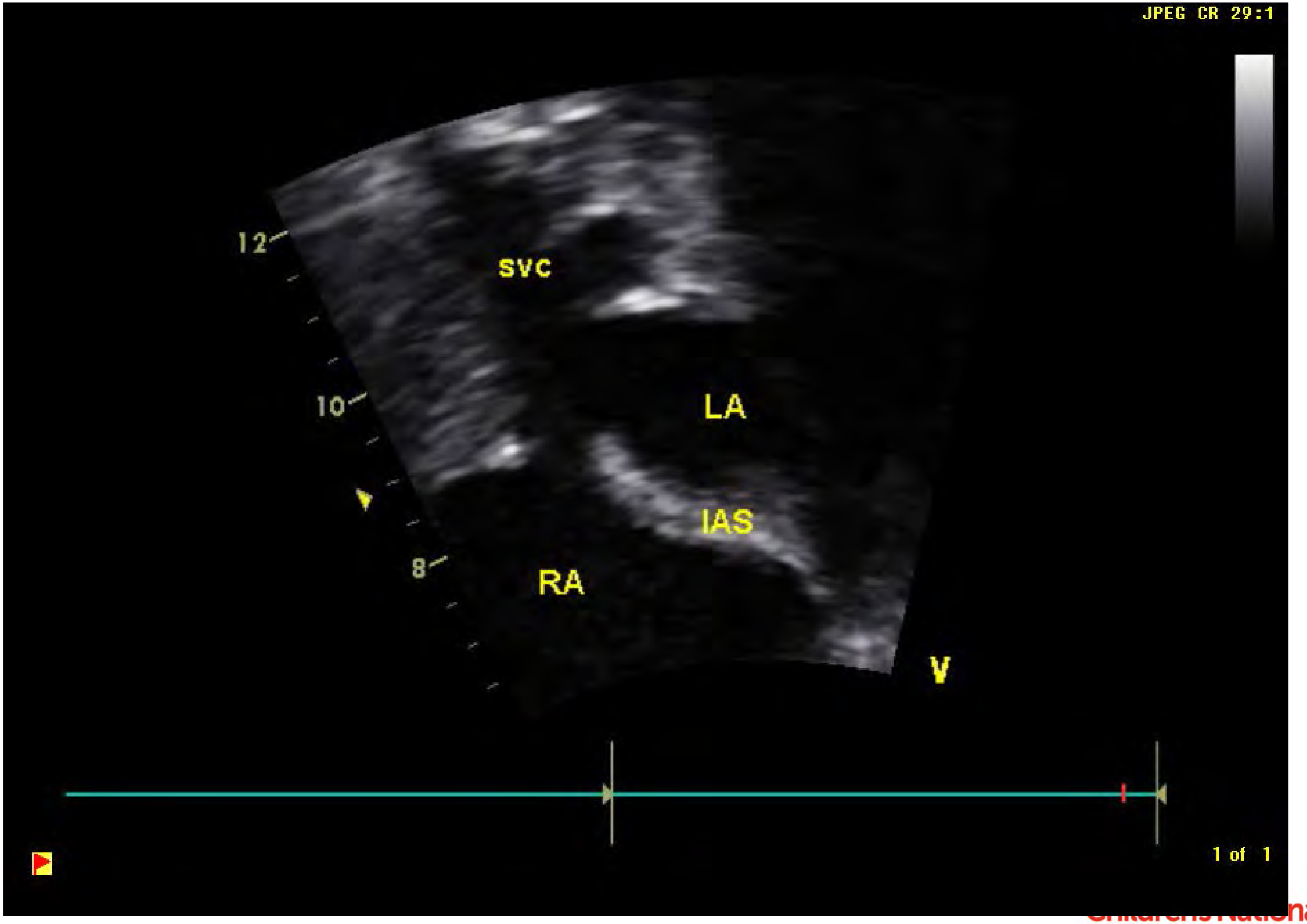
Deficiency in the wall between the right pulmonary veins and the RA

PAPV-DRAINAGE

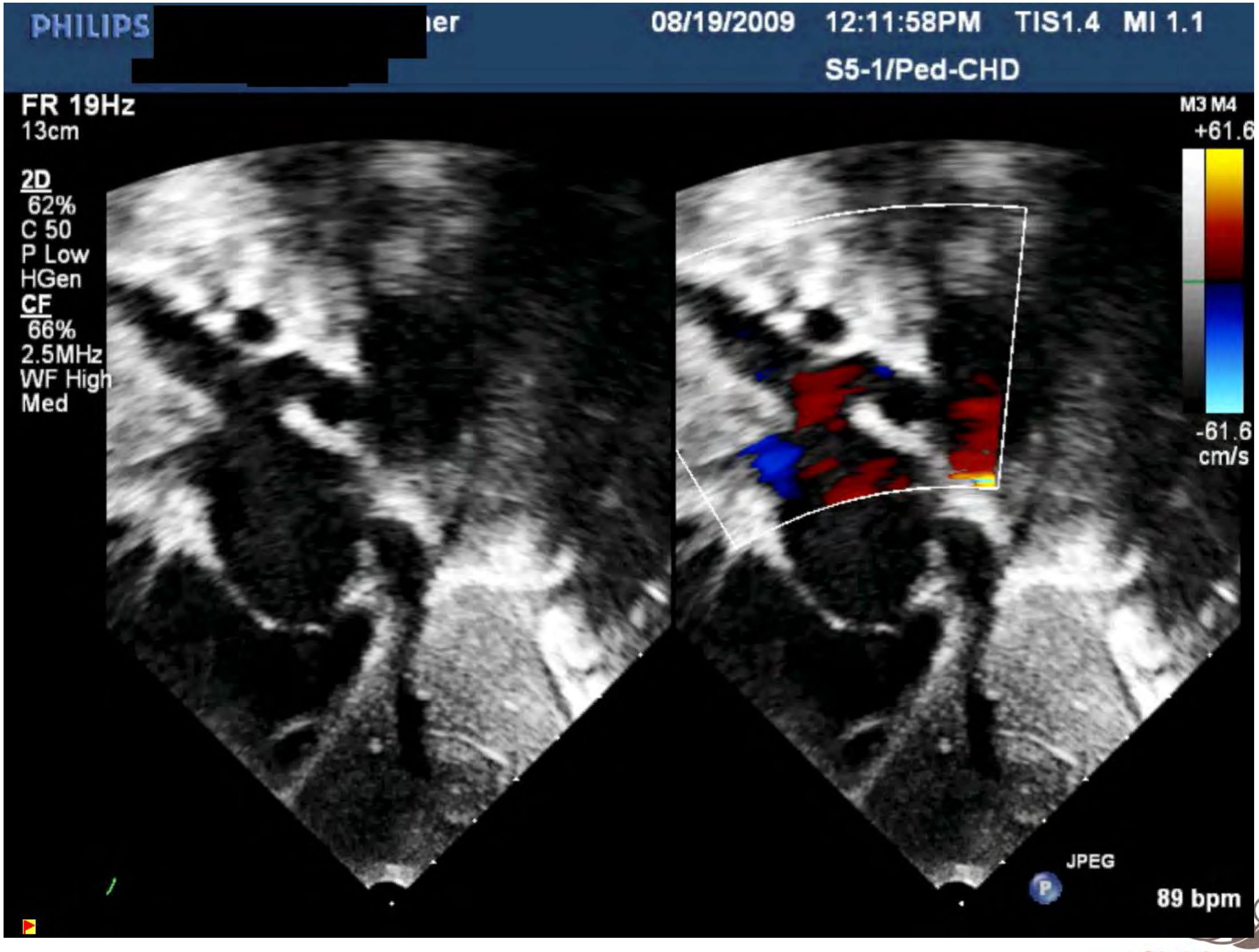
- SVC type = RUPV
- Inferior type = RLPV



Sinus Venosus ASD



Sinus Venosus ASD



Children's National™

Partial Anomalous Pulmonary Venous Return (PAPVR)

Right veins (more common):

RA

SVC (RUPV to the RA or base of the SVC-sinus venosus ASD)

IVC

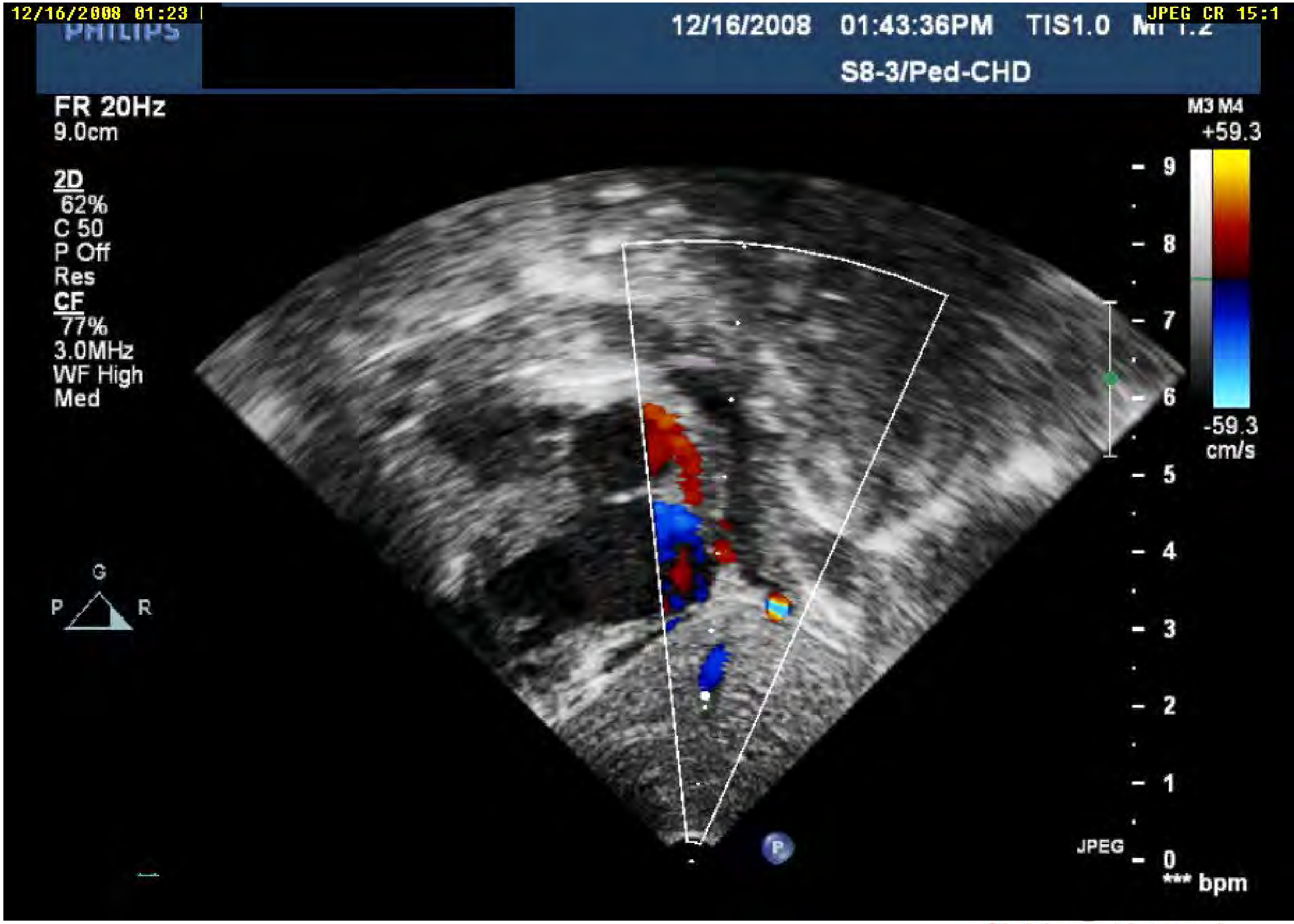
Left veins:

Innominate vein

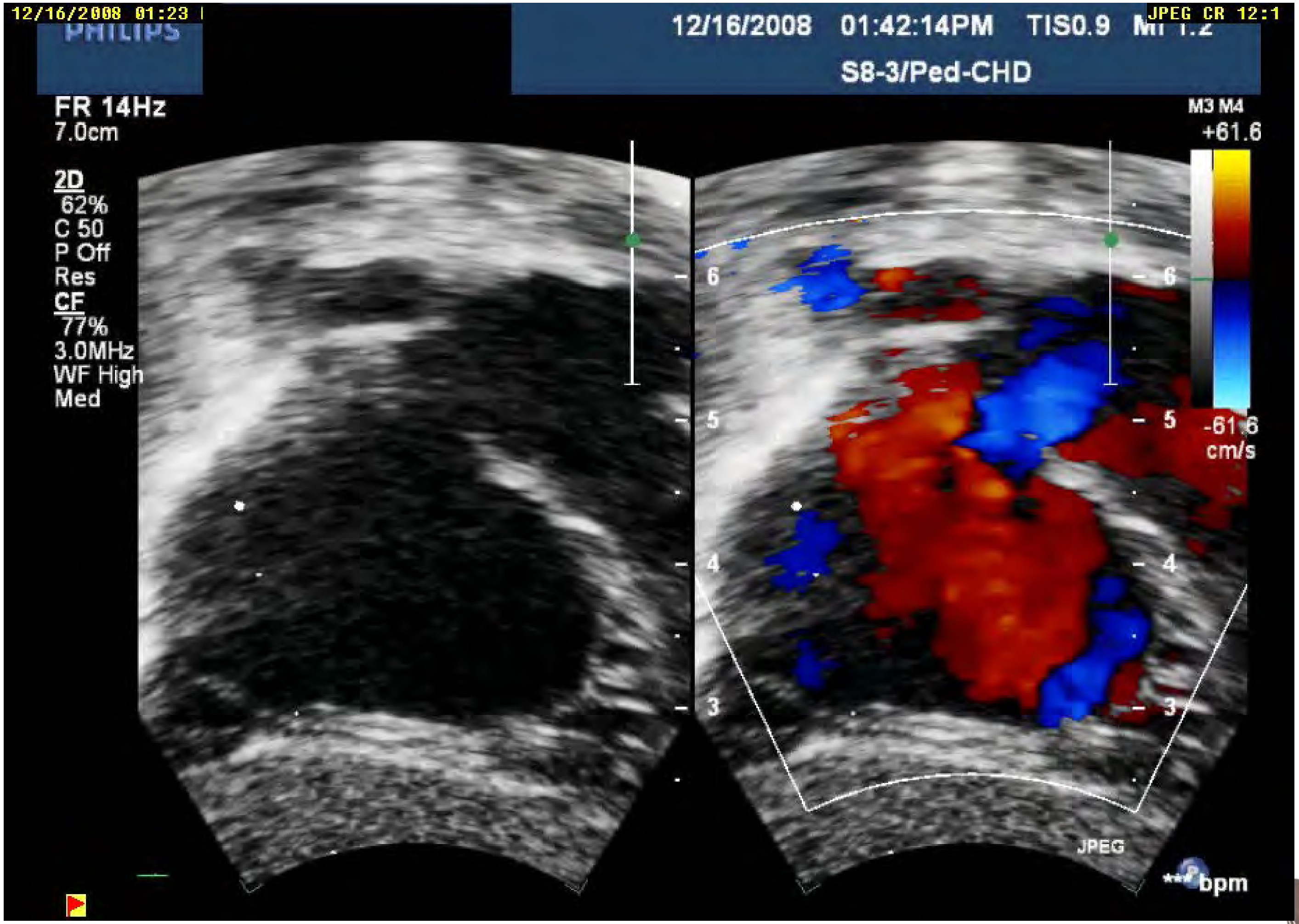
Coronary sinus

Rarely: SVC, IVC, right atrium, or left subclavian vein

PAPVR to IVC


12/16/2008 01:23
[redacted]
12/16/2008 01:43:36PM TIS1.0
JPEG CR 15:1
S8-3/Ped-CHD
FR 20Hz
9.0cm
2D
62%
C 50
P Off
Res
CF
77%
3.0MHz
WF High
Med
G
P
R
M3 M4
+59.3
-59.3
cm/s
9
8
7
6
5
4
3
2
1
0
JPEG
*** bpm

PAPVR to SVC


12/16/2008 01:23
PHILIPS
12/16/2008 01:42:14PM TIS0.9 MI 1.2
JPEG CR 12:1
S8-3/Ped-CHD
FR 14Hz
7.0cm
2D
62%
C 50
P Off
Res
CF
77%
3.0MHz
WF High
Med
M3 M4
+61.6
-61.6
cm/s
6
5
4
3
JPEG
bpm

Total Anomalous Pulmonary Venous Return (TAPVR)

I: Supracardiac: common pulmonary vein drains into the right superior vena cava from the left superior vena cava (vertical vein) and the left innominate vein (50%)

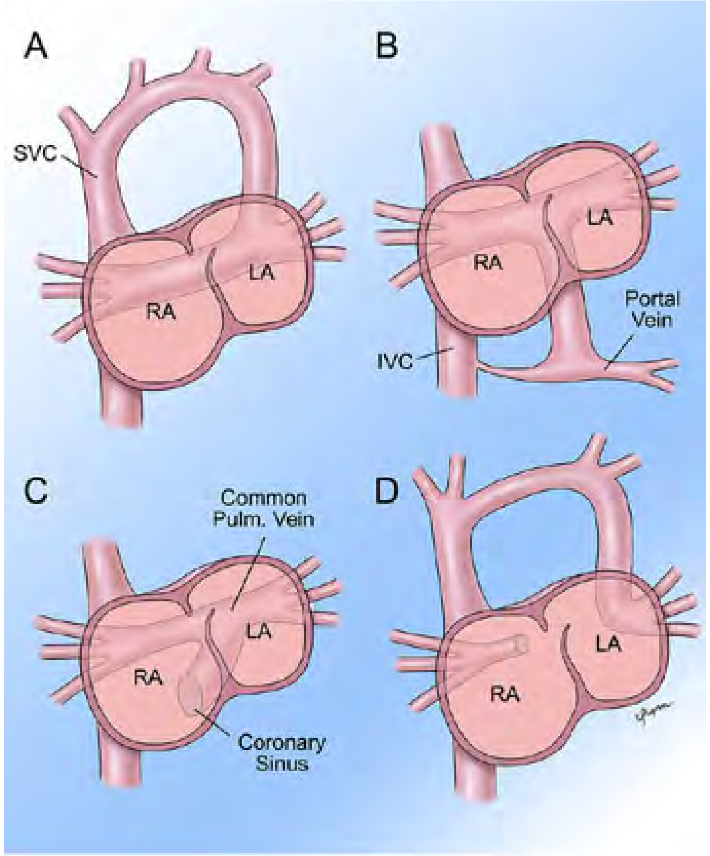
II: Cardiac: coronary sinus, right atrium (20%)

III: Infracardiac: subdiaphragmatic (portal vein, inferior vena cava, ductus venosus) (20%)

IV: Mixed: any combination of types I, II, III, the least common

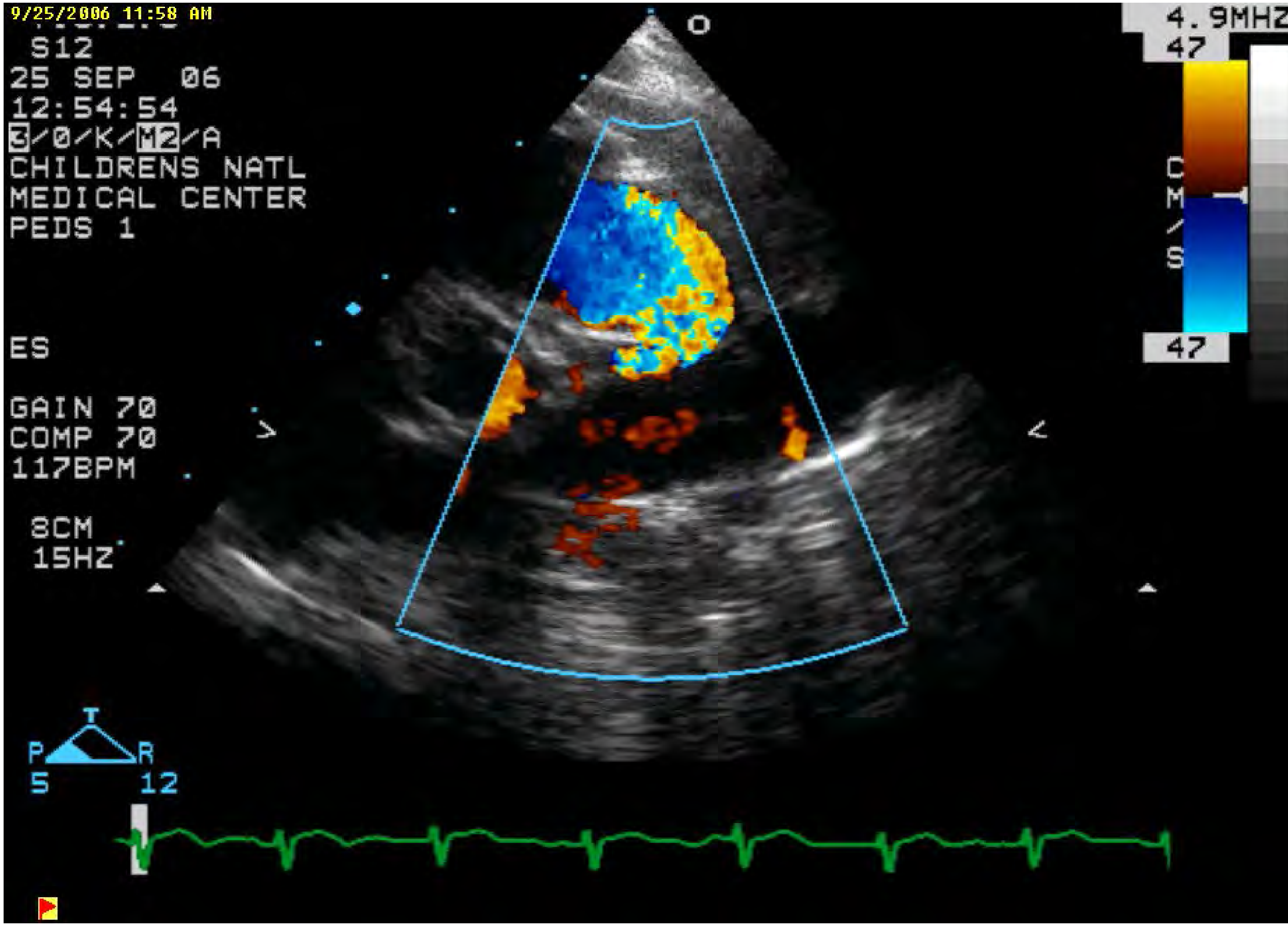
Children's National ™

TAPVR



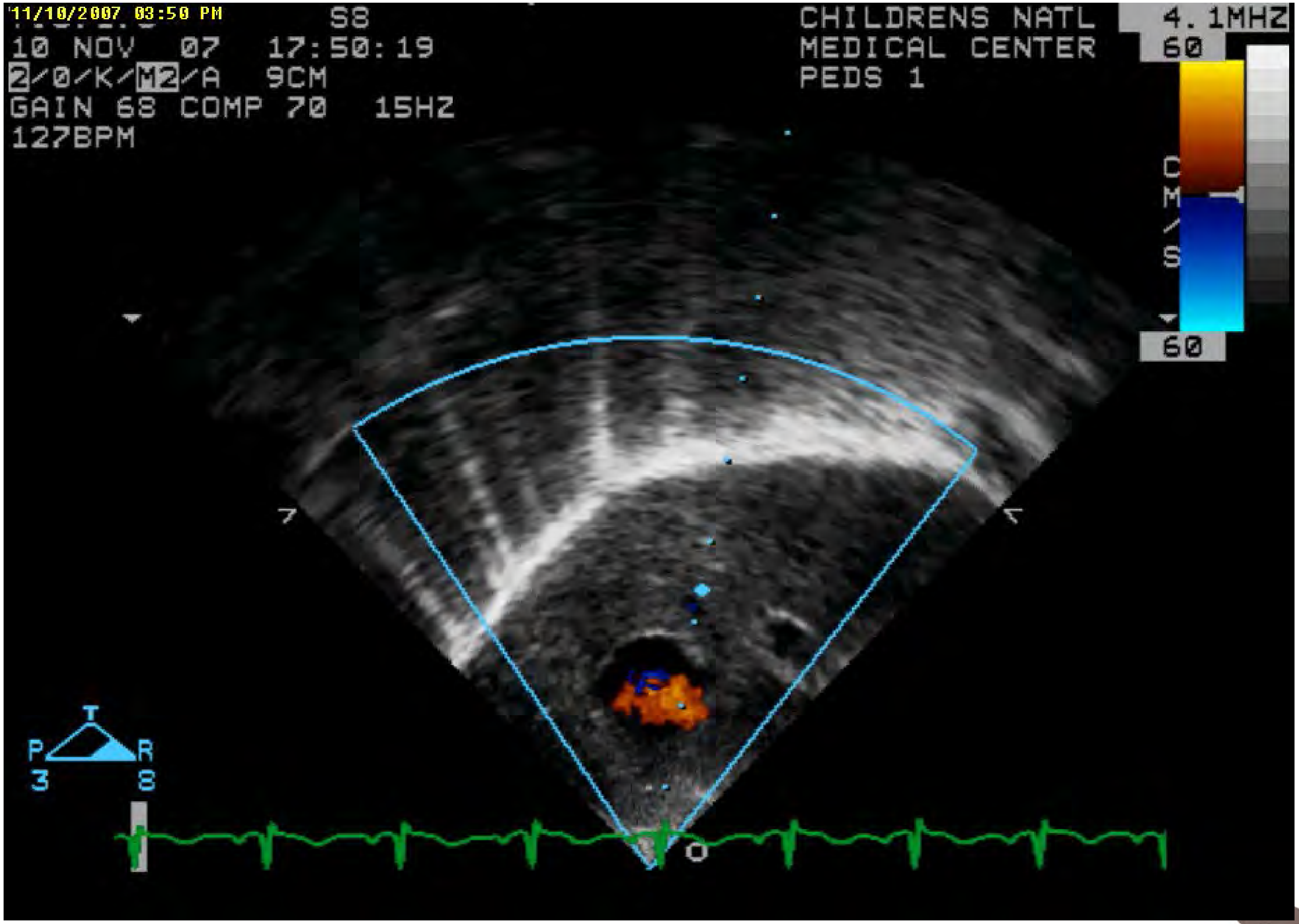
Children's Hospital Boston

TAPVR to Vertical Vein



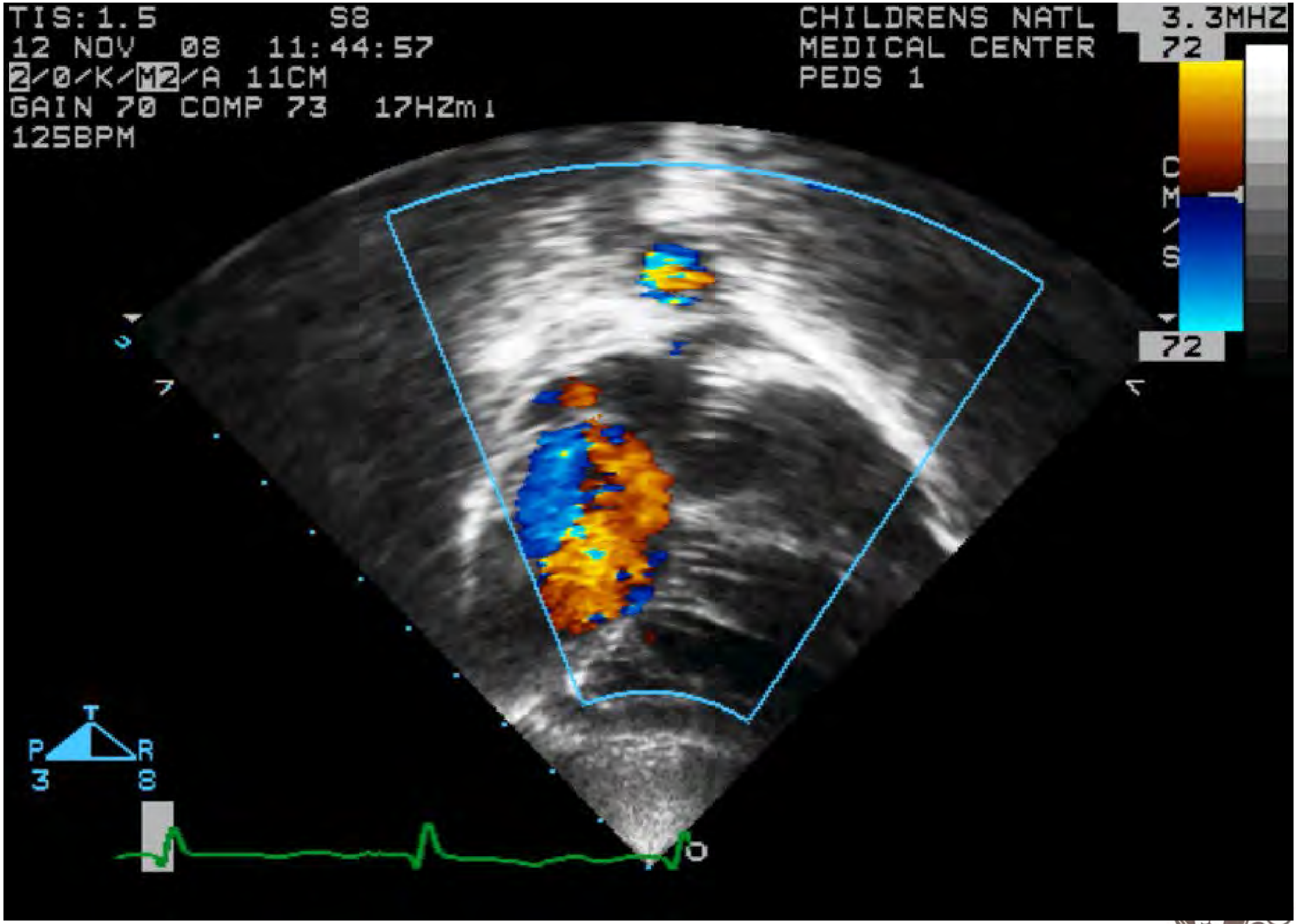
Children's National™

TAPVR to IVC



Children's National™

TAPVR to CS



TIS:1.5
S8
12 NOV 08 11:44:57
2/0/K/M2/A 11CM
GAIN 70 COMP 73 17HZm1
125BPM
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
3.3MHZ
72
CM/S
72
P
T
R
3
8

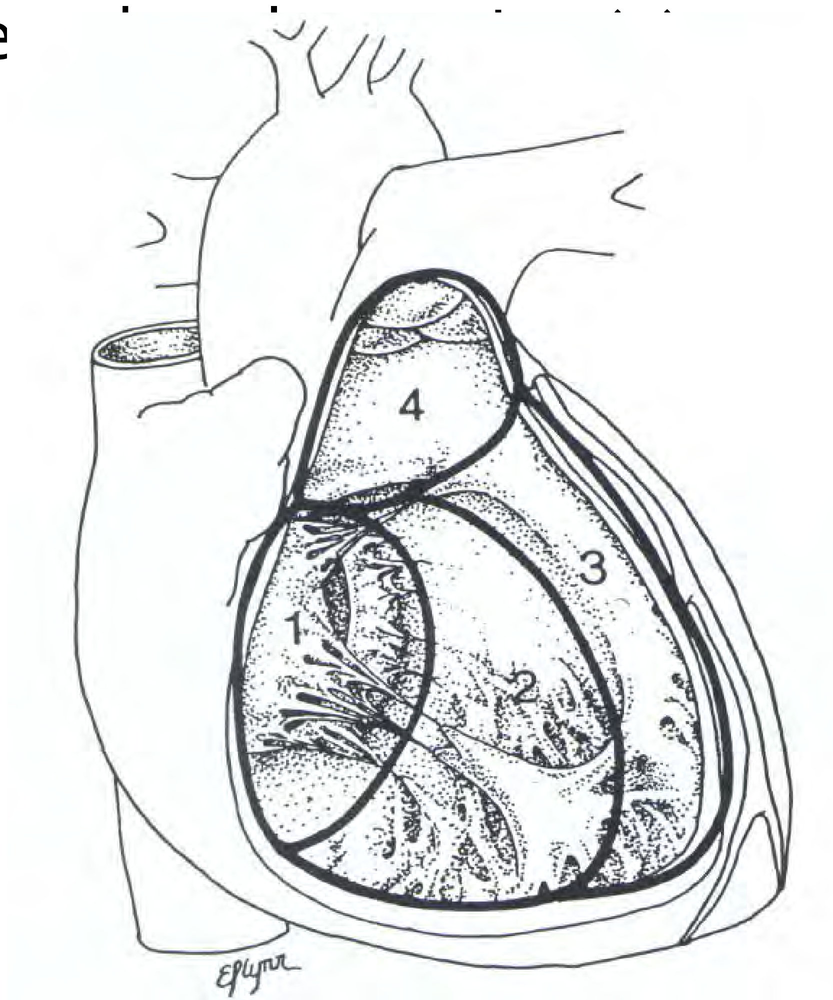
The Ventricular Septum

AV canal septum (1)

Muscular septum including the

and the septal band (3)

Conal septum (4)



The Ventricular Septum

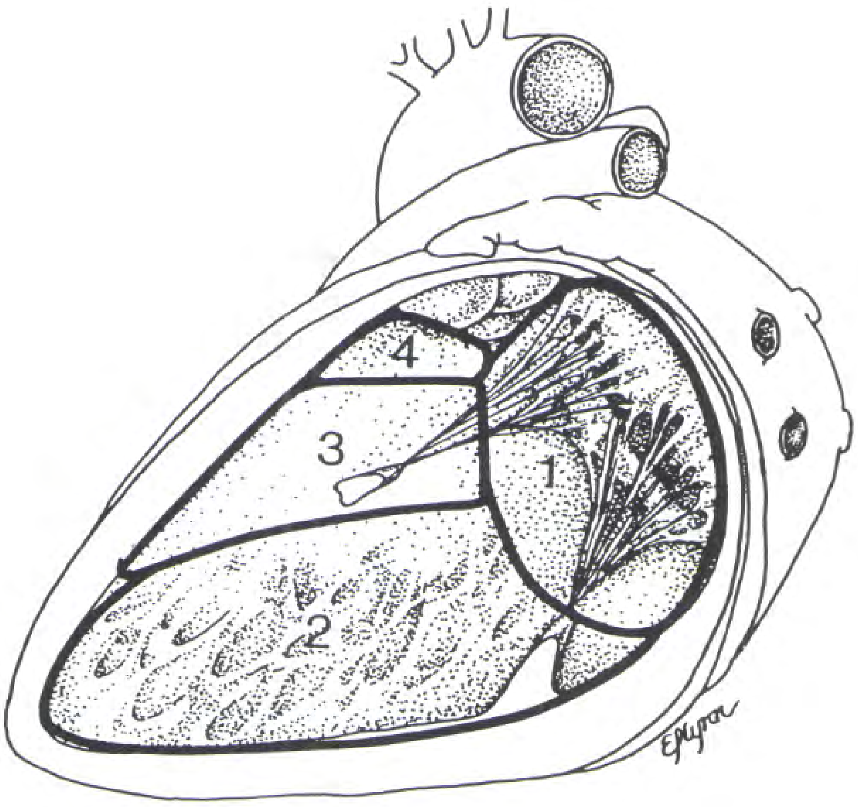
Left ventricular view

AV canal septum (1)

Muscular septum including th

and the septal band (3)

Conal septum (4)



Muscular VSD

Within the muscular ventricular septum

Apical (black)

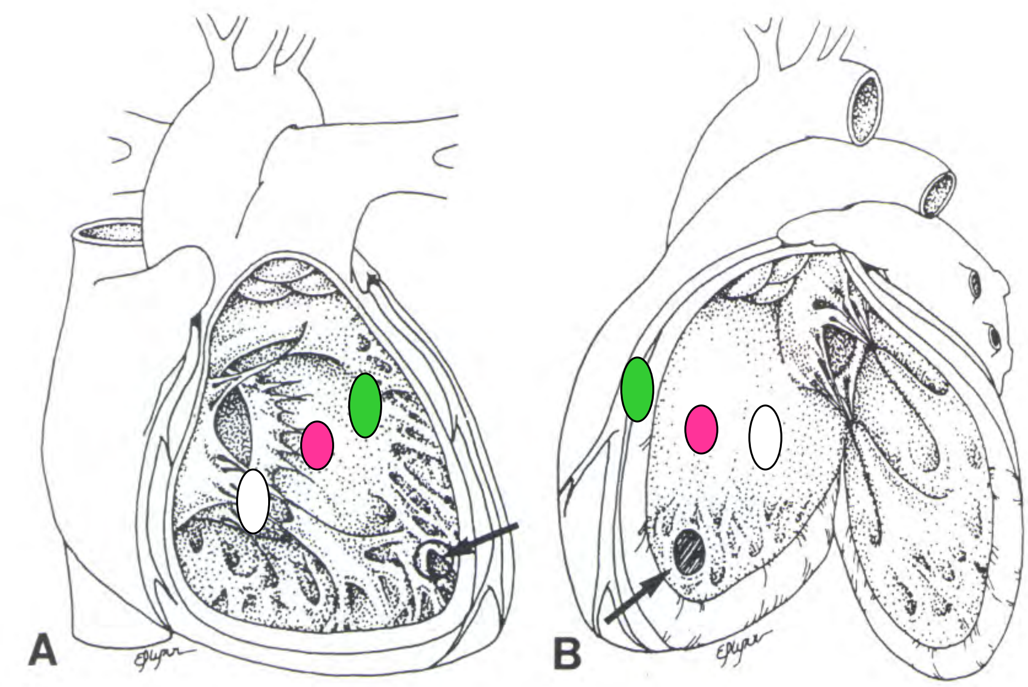
Mid (pink)

Anterior (green)

Posterior/inlet (white)

"Swiss cheese"

Tend to get smaller with tim



Conoventricular VSD

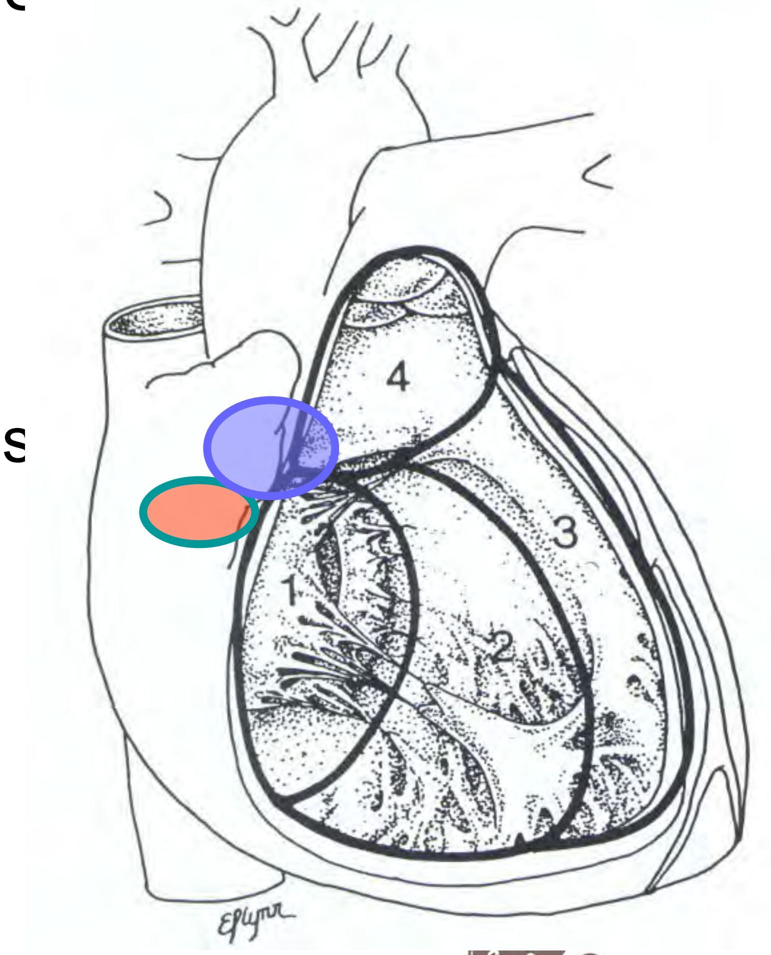
In the area where the AV canal septum, conal septum and muscular septum meet

“Membranous VSD”

- “Para-” or “Peri-” (red)

Malalignment

- “TOF-type”, “VSD in the Y of s



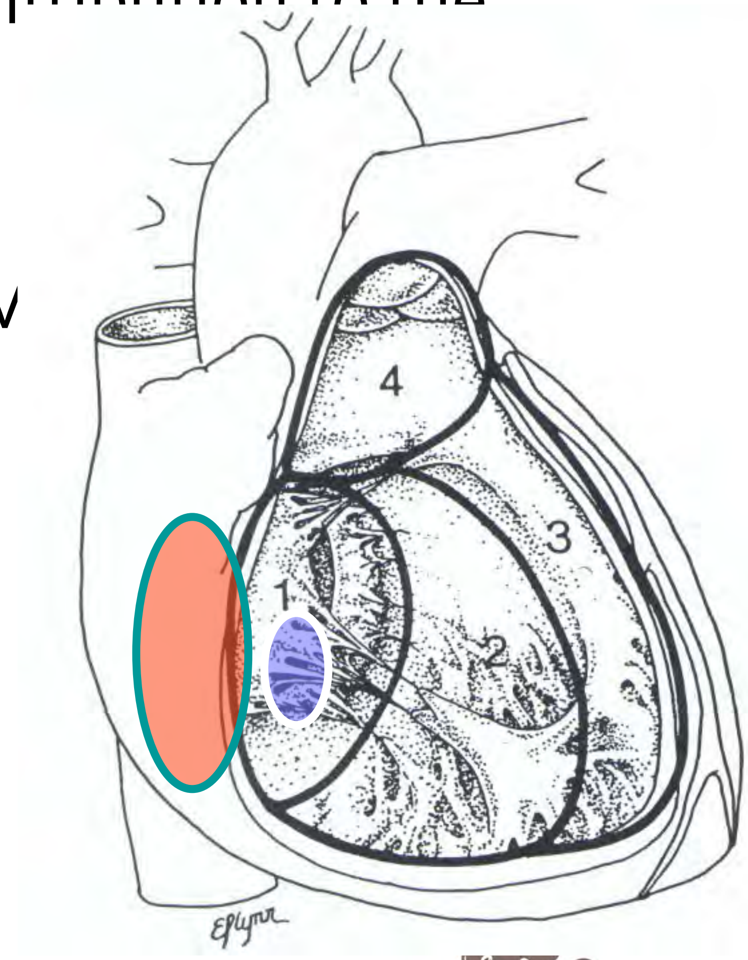
Children's National

AV Canal Type VSD

Deficiency of the AV cushion contribution to the ventricular septum (red)

“Inlet VSD”

Different from “Inlet muscular” V
surrounded by muscle (blue)



Children's National

VSD: Clinical Correlation

Size and pulmonary vascular resistance determines clinical presentation

- Fetal transition

Symptoms are determined by the size of the shunt

- Size of defect
- Presence of other anomalies
- Extracardiac abnormalities

VSD: Clinical Correlation

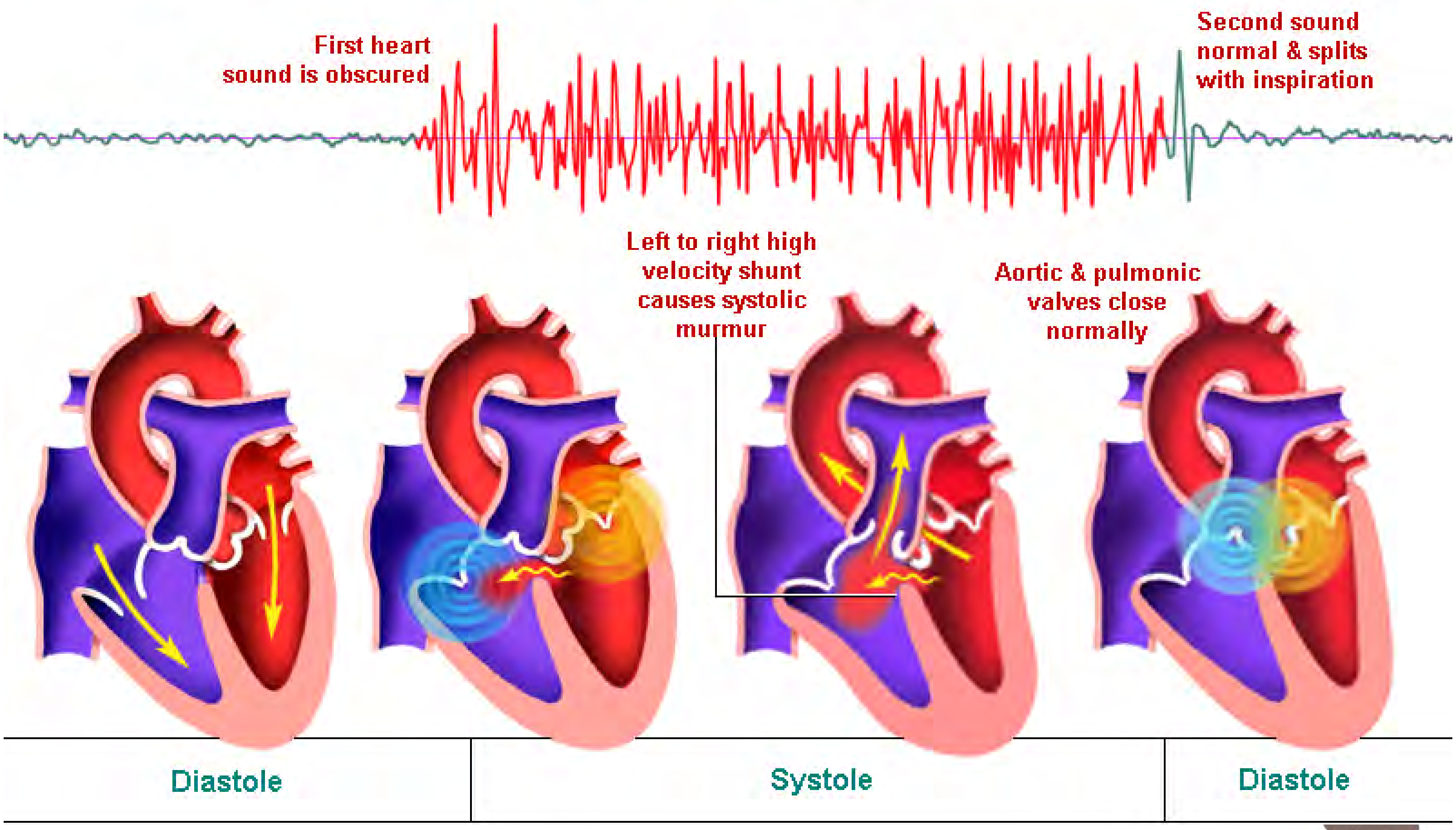
Audible after several days (not immediately after birth), typically picked up at 1st visit

Large defects=congestive heart failure

- Tachypnea (RR>60)
- Poor feeding/poor growth
- Reflux/vomiting

(.wav)

Small Ventricular Septal Defects



Children's National™

VSD: Clinical Correlation

Spontaneous resolution

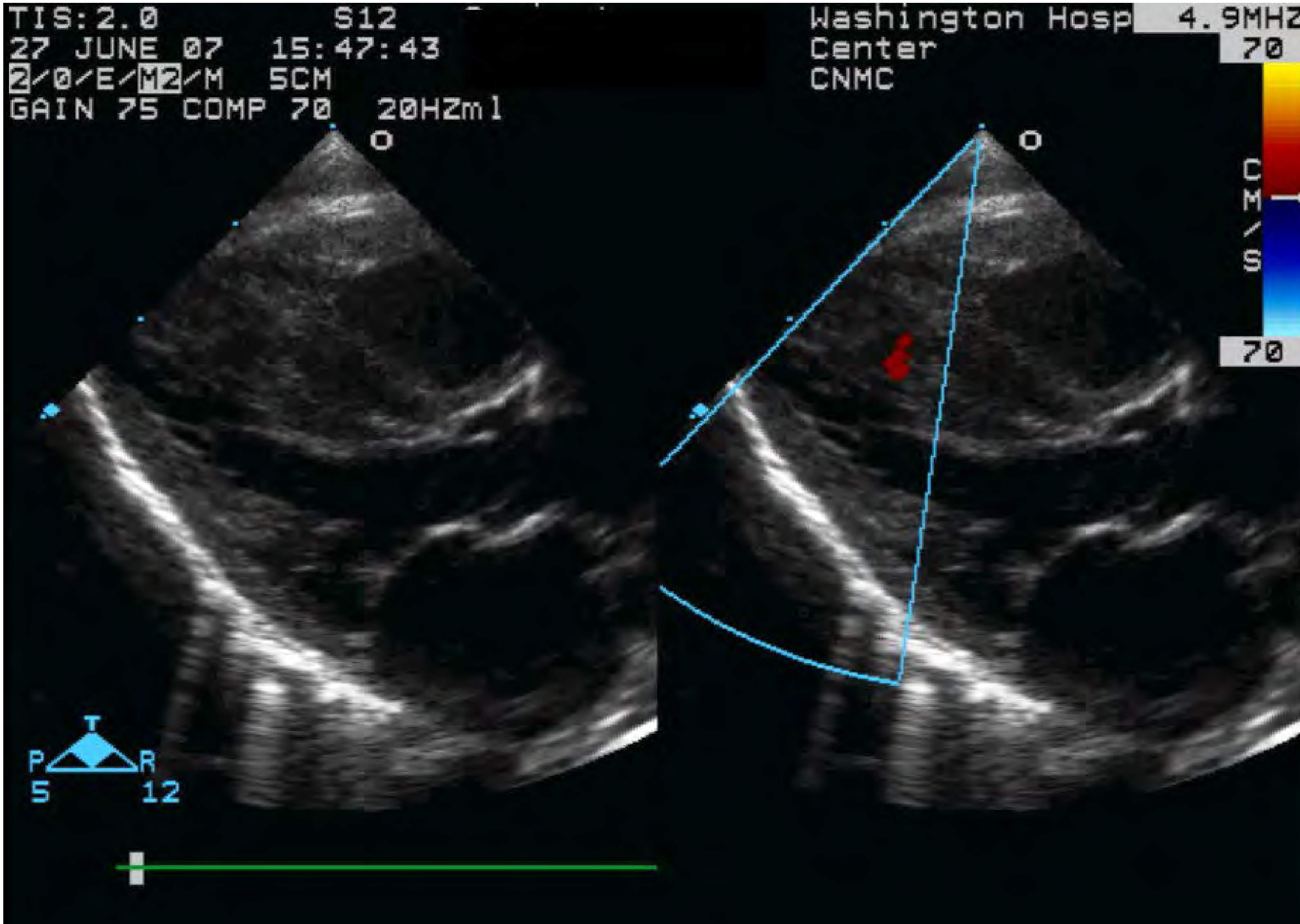
Or not…

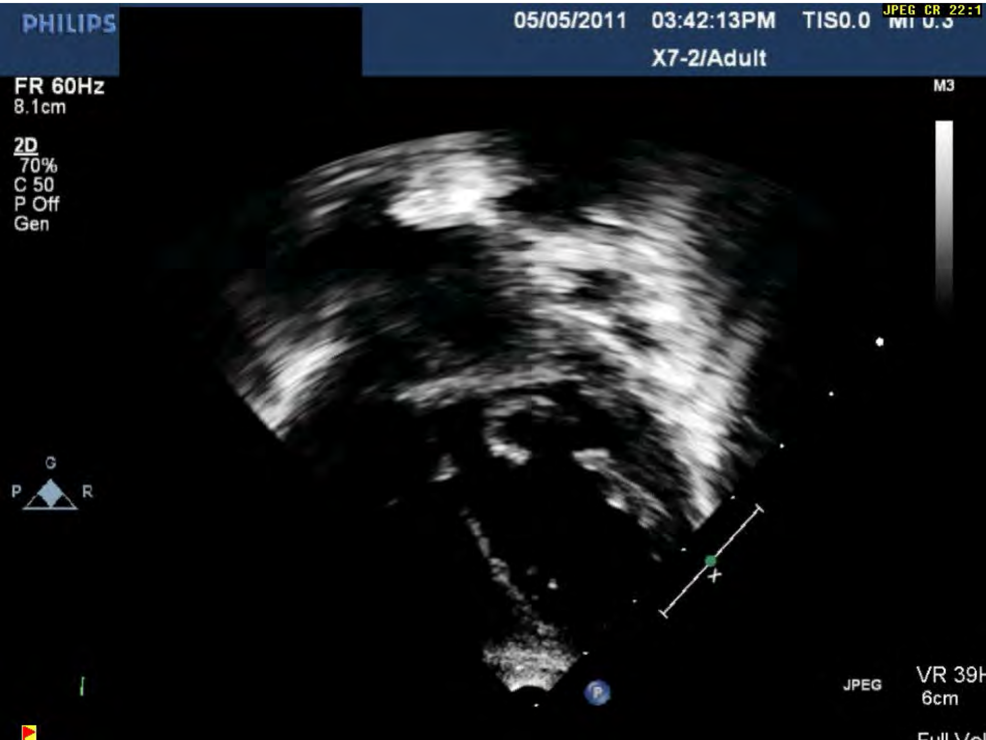
Pulmonary disease

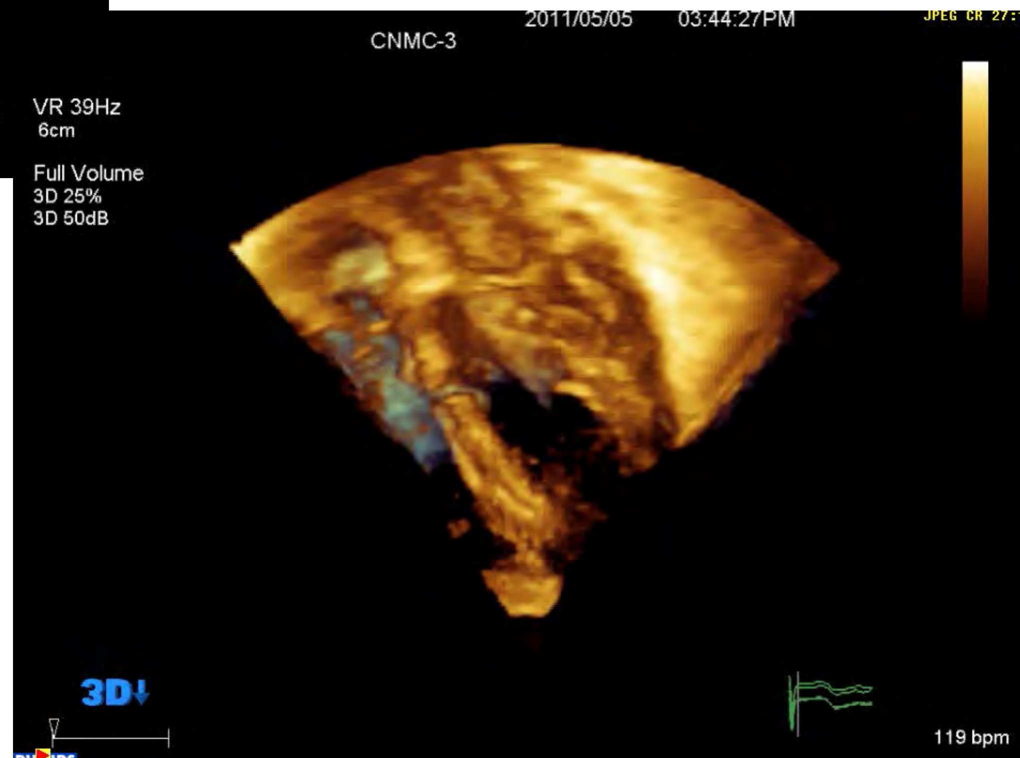
- Eisenmenger’s syndrome

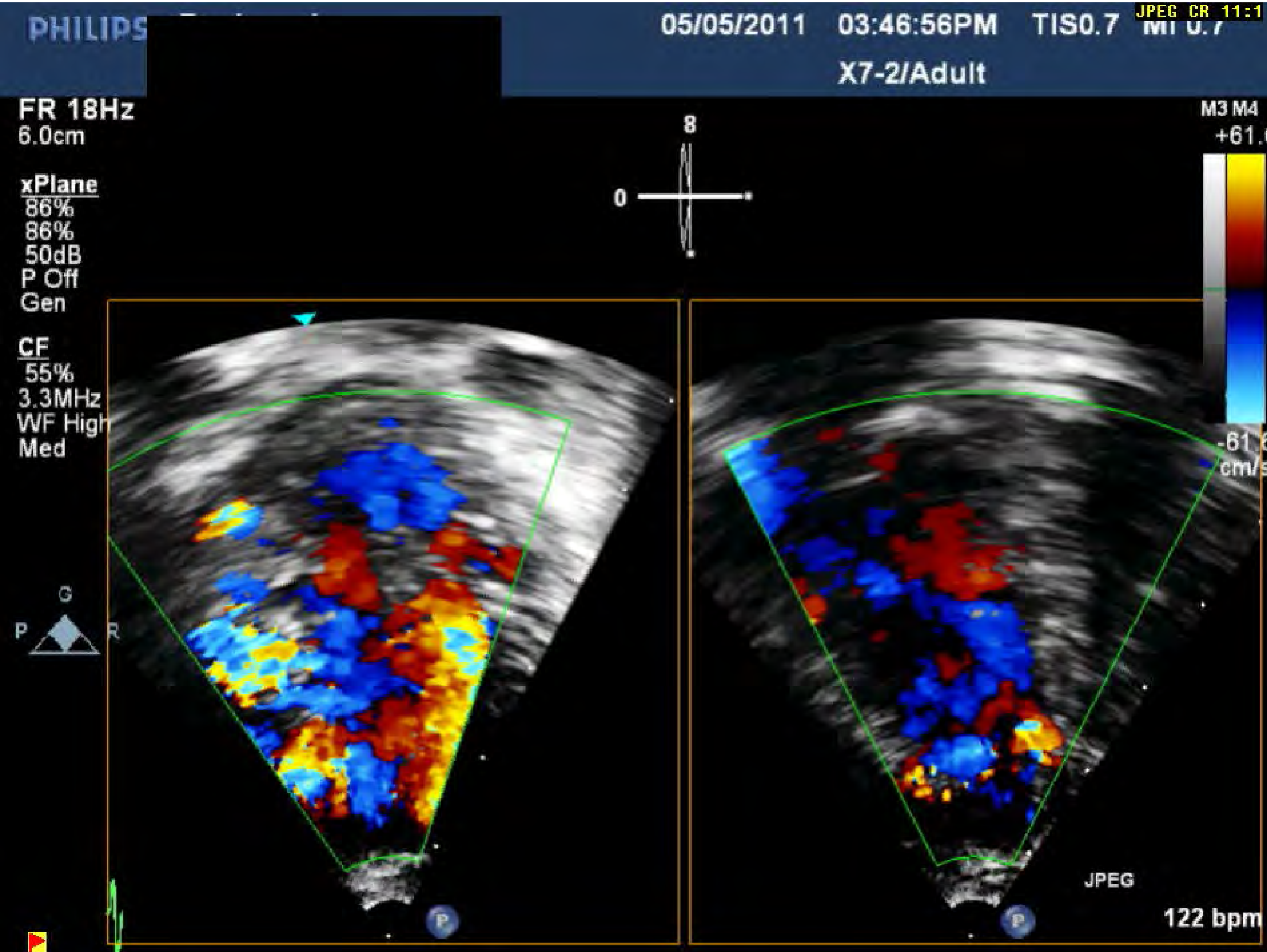
Aortic regurgitation

Muscular VSD

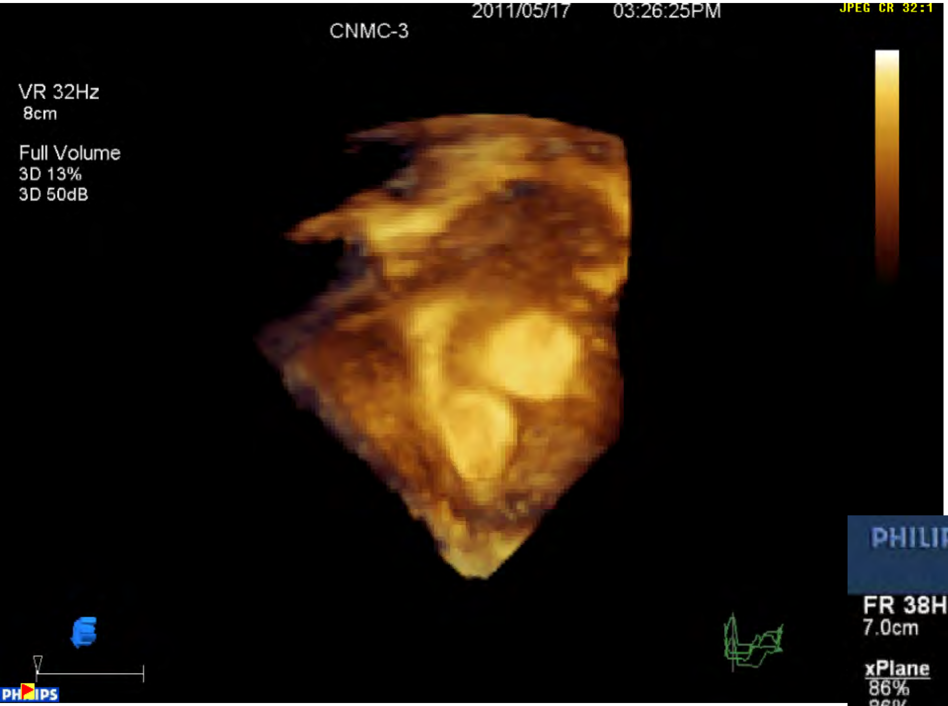


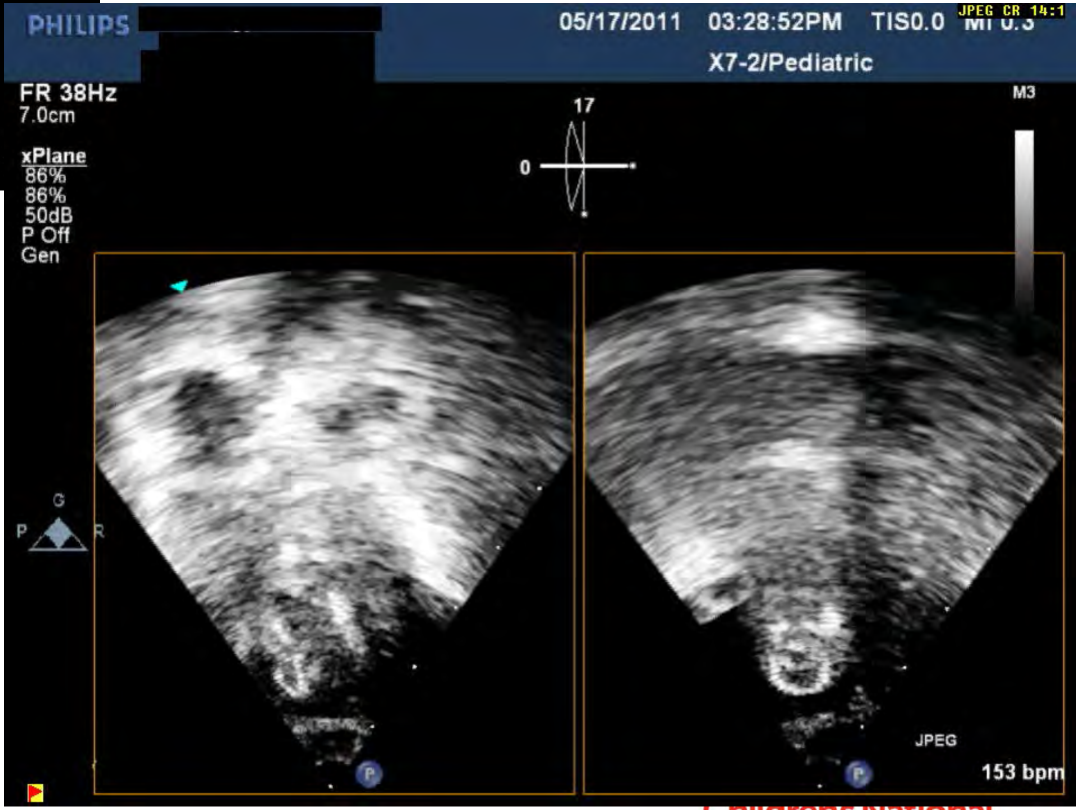

PHILIPS
05/05/2011 03:42:13PM TIS0.0
JPEG CR 22:1
X7-2/Adult
FR 60Hz
8.1cm
2D
70%
C 50
P Off
Gen
M3
G
P
R
JPEG


2011/05/05 03:44:27PM
JPEG CR 27:1
CNMC-3
VR 39Hz
6cm
Full Volume
3D 25%
3D 50dB
3D
119 bpm

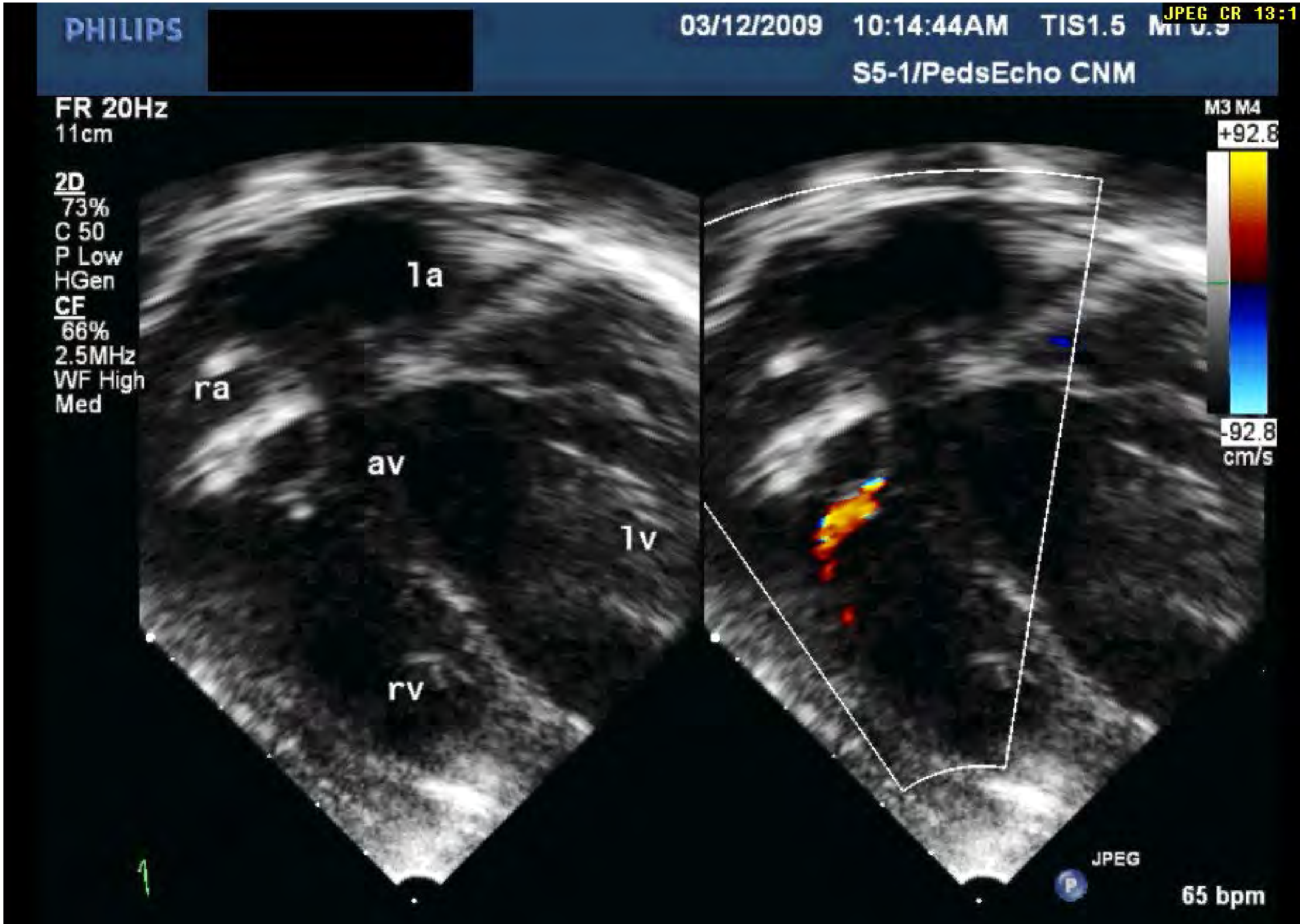


Children's National

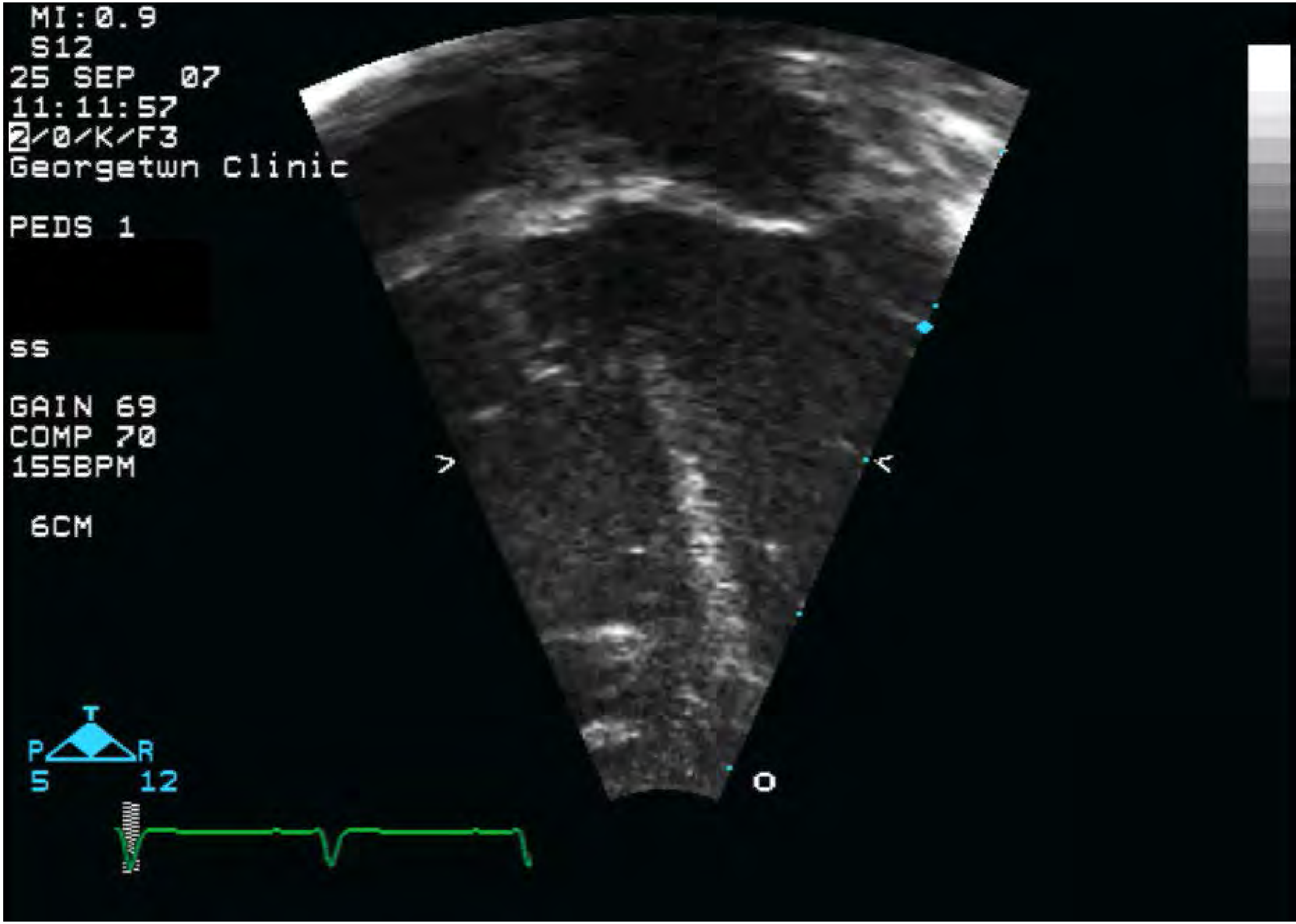

CNMC-3
2011/05/17
03:26:25PM
JPEG CR 32:1
VR 32Hz
8cm
Full Volume
3D 13%
3D 50dB


PHILIPS
[redacted]
05/17/2011
03:28:52PM
TIS0.0
JPEG CR 14:1
X7-2/Pediatric
FR 38Hz
7.0cm
xPlane
86%
86%
50dB
P Off
Gen
17
0
M3
JPEG
153 bpm

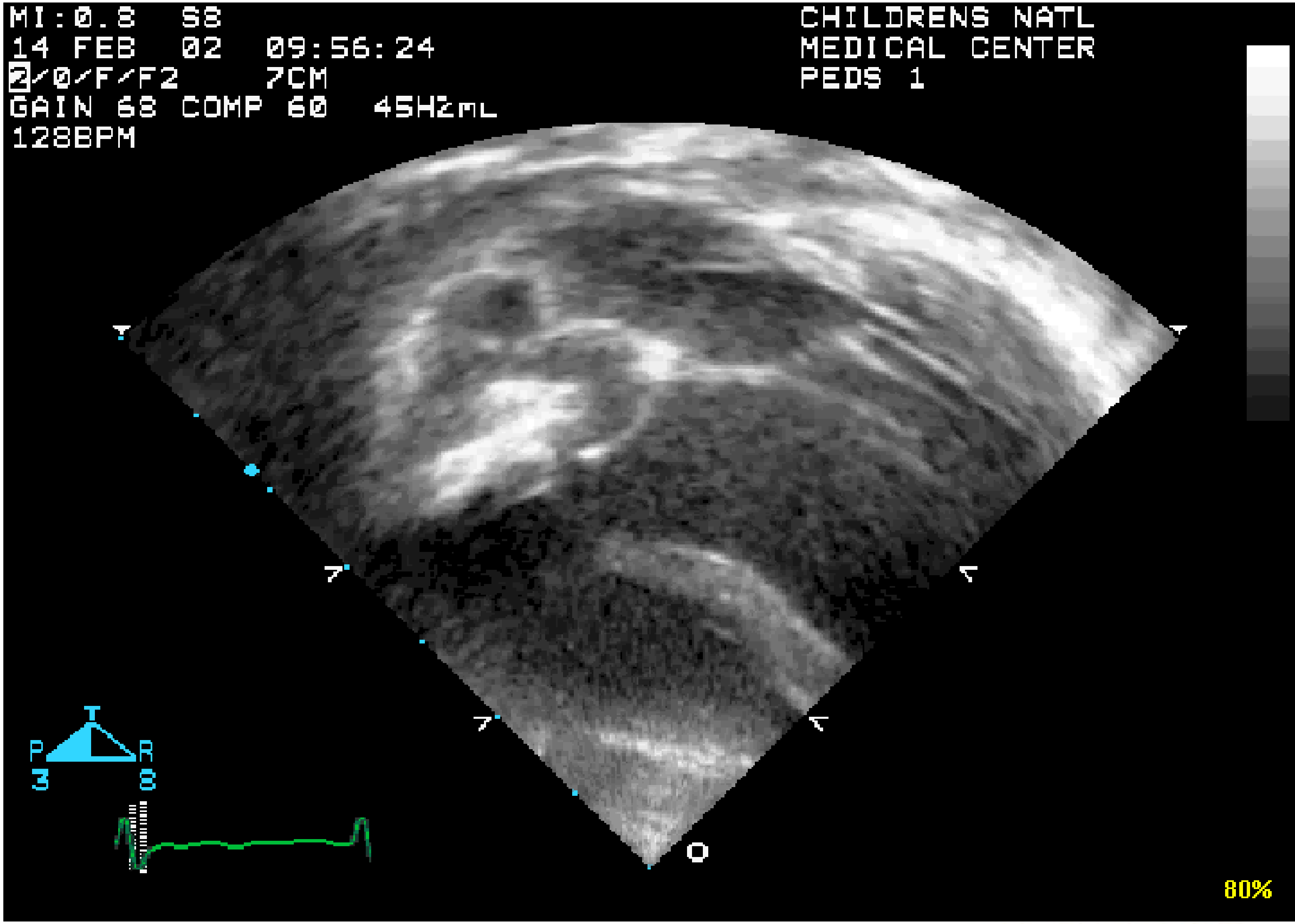
Membranous VSD

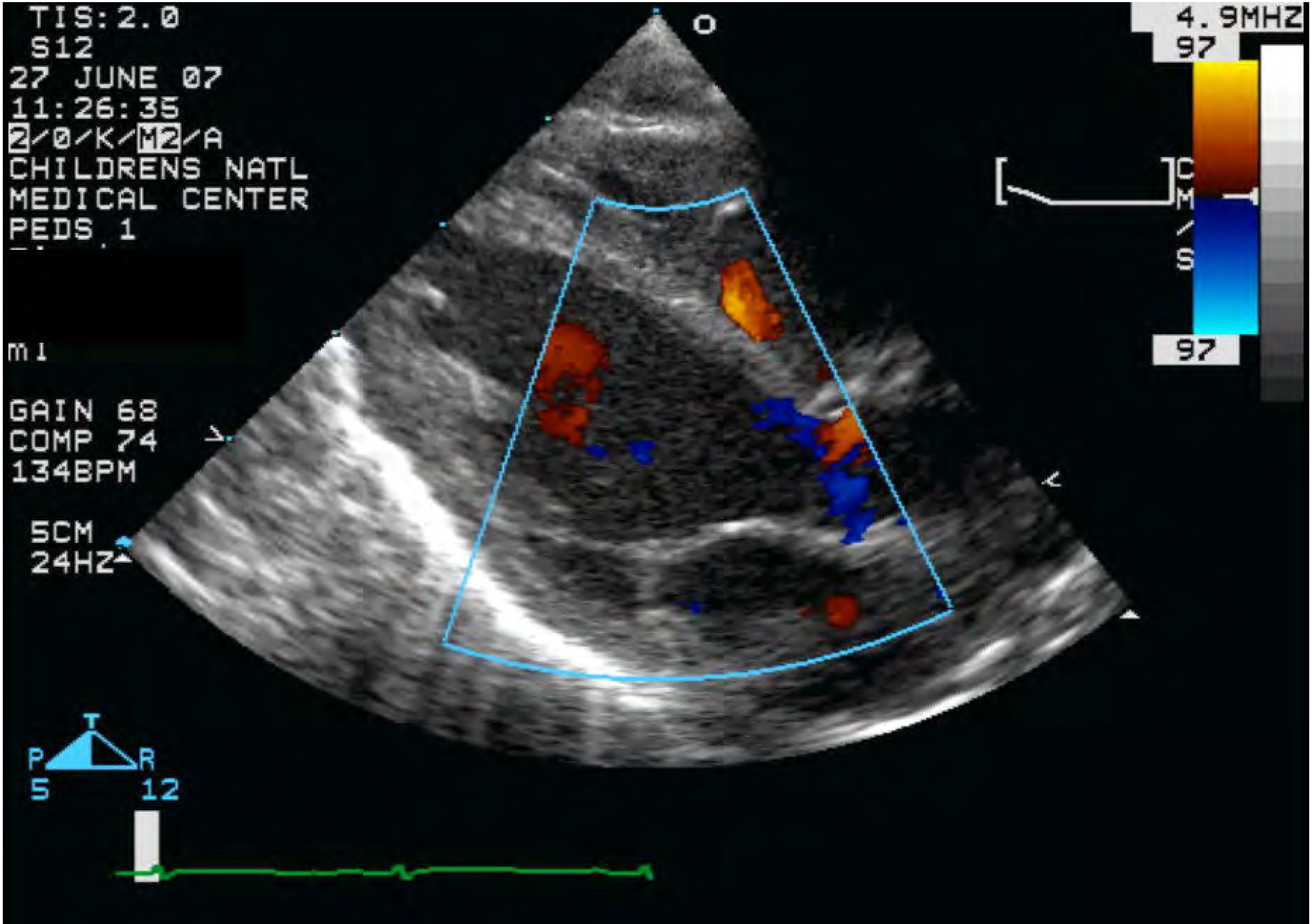


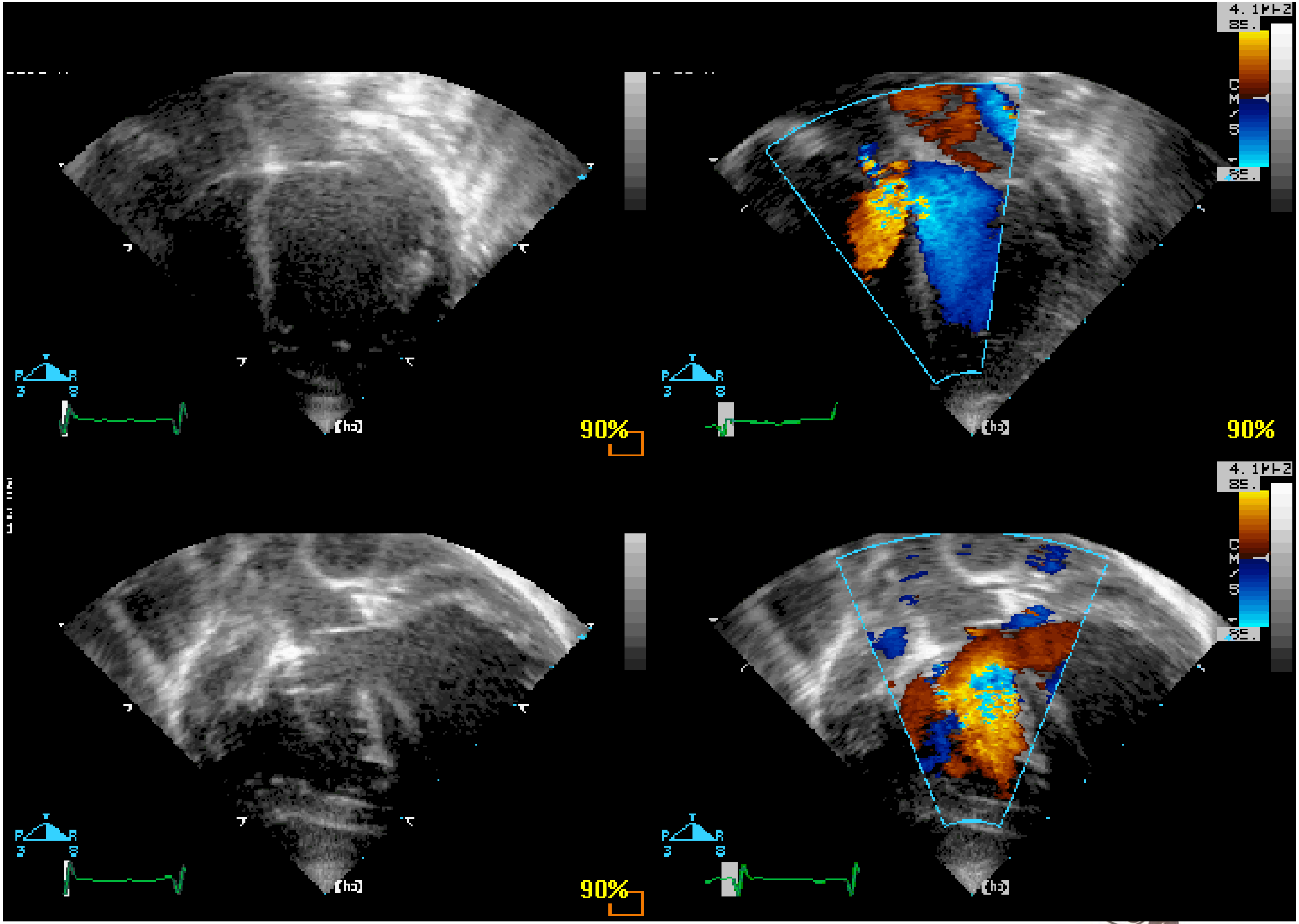
Membranous VSD
w/ TV aneurysm tissue



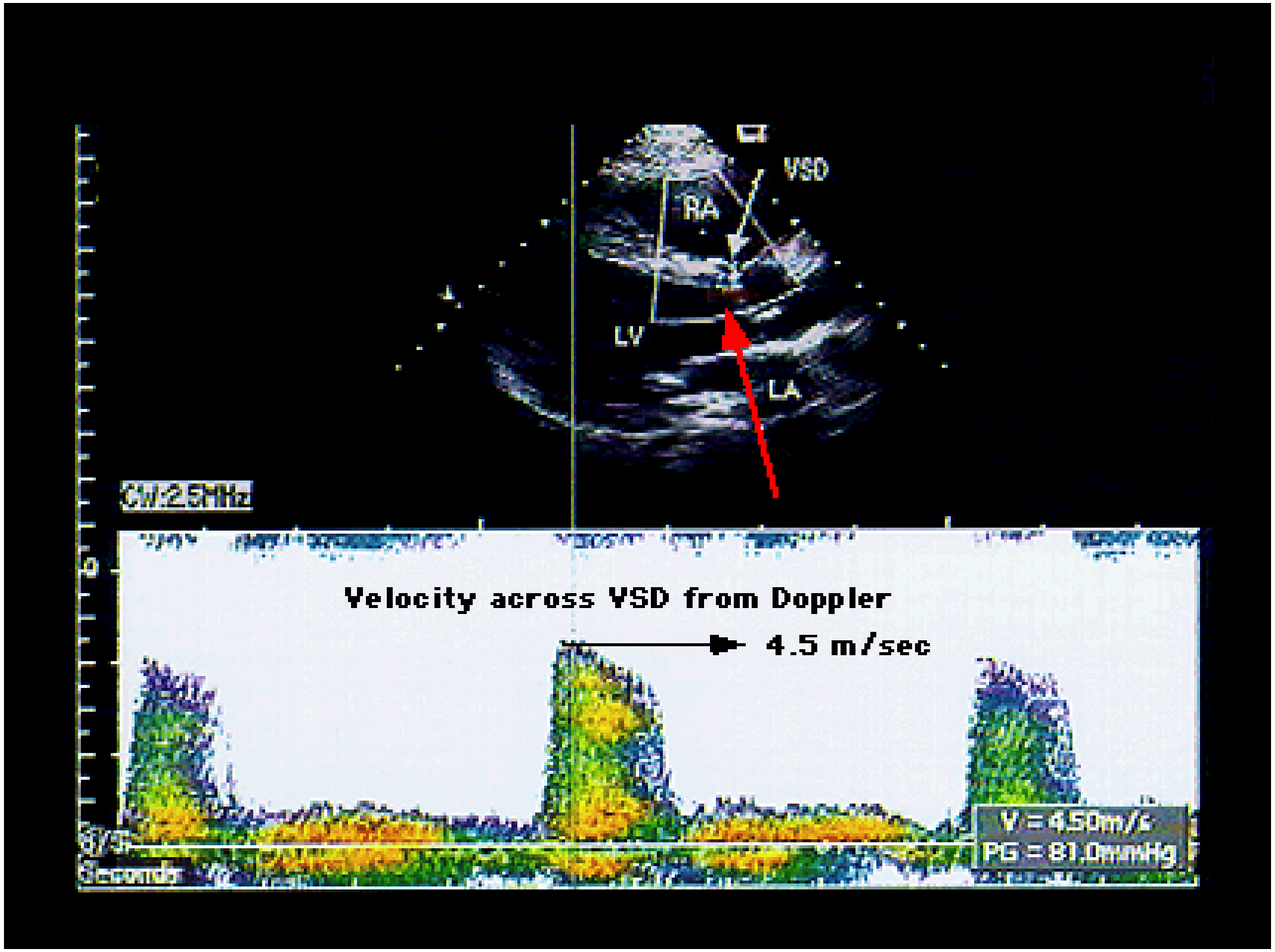
Membranous VSD w/ Aortic Valve Prolapse




TIS:2.0
S12
27 JUNE 07
11:26:35
2/0/K/M2/A
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
[redacted]
m1
GAIN 68
COMP 74
134BPM
5CM
24HZ
4.9MHZ
97
CM/S
97
T
P
R
5
12

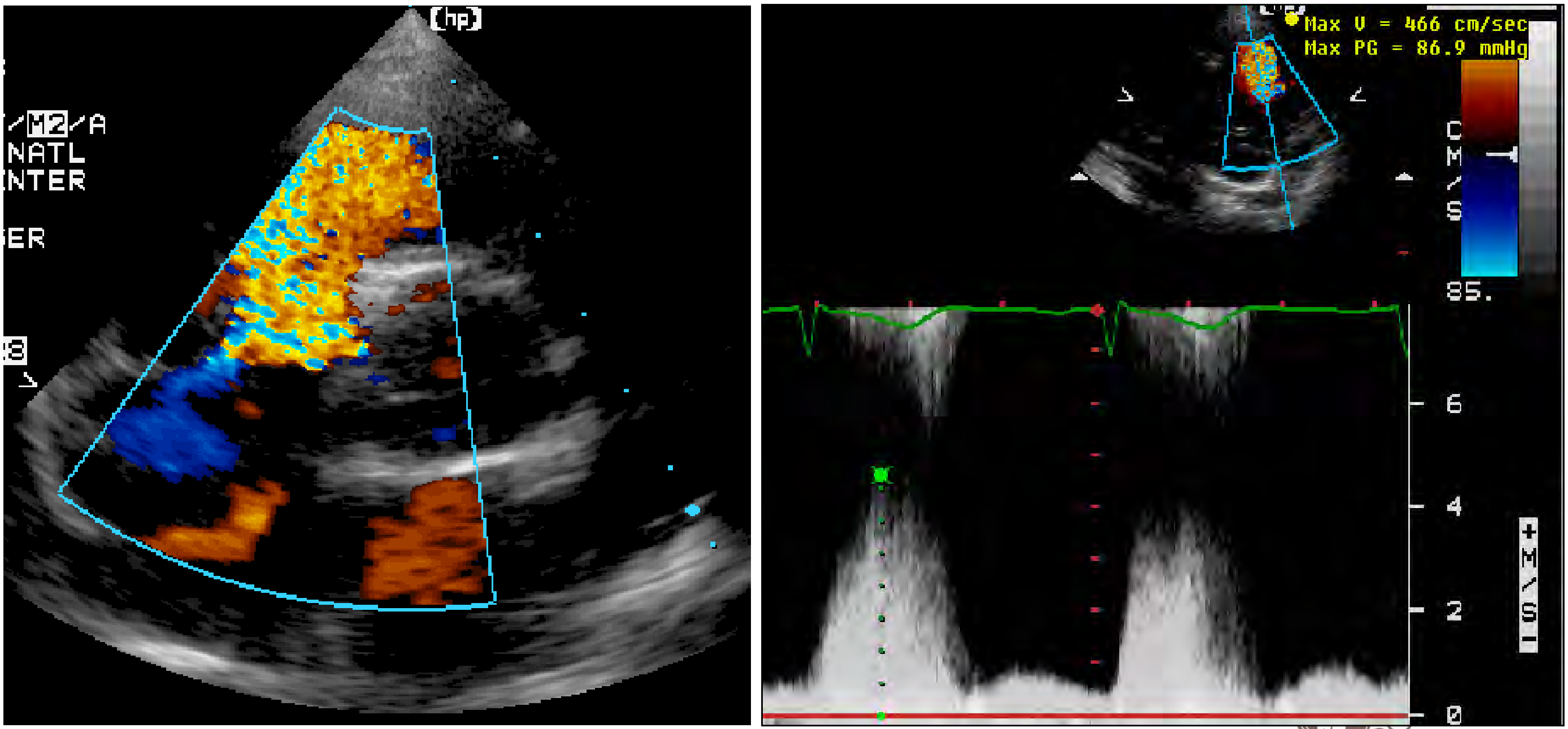


Children's National ™



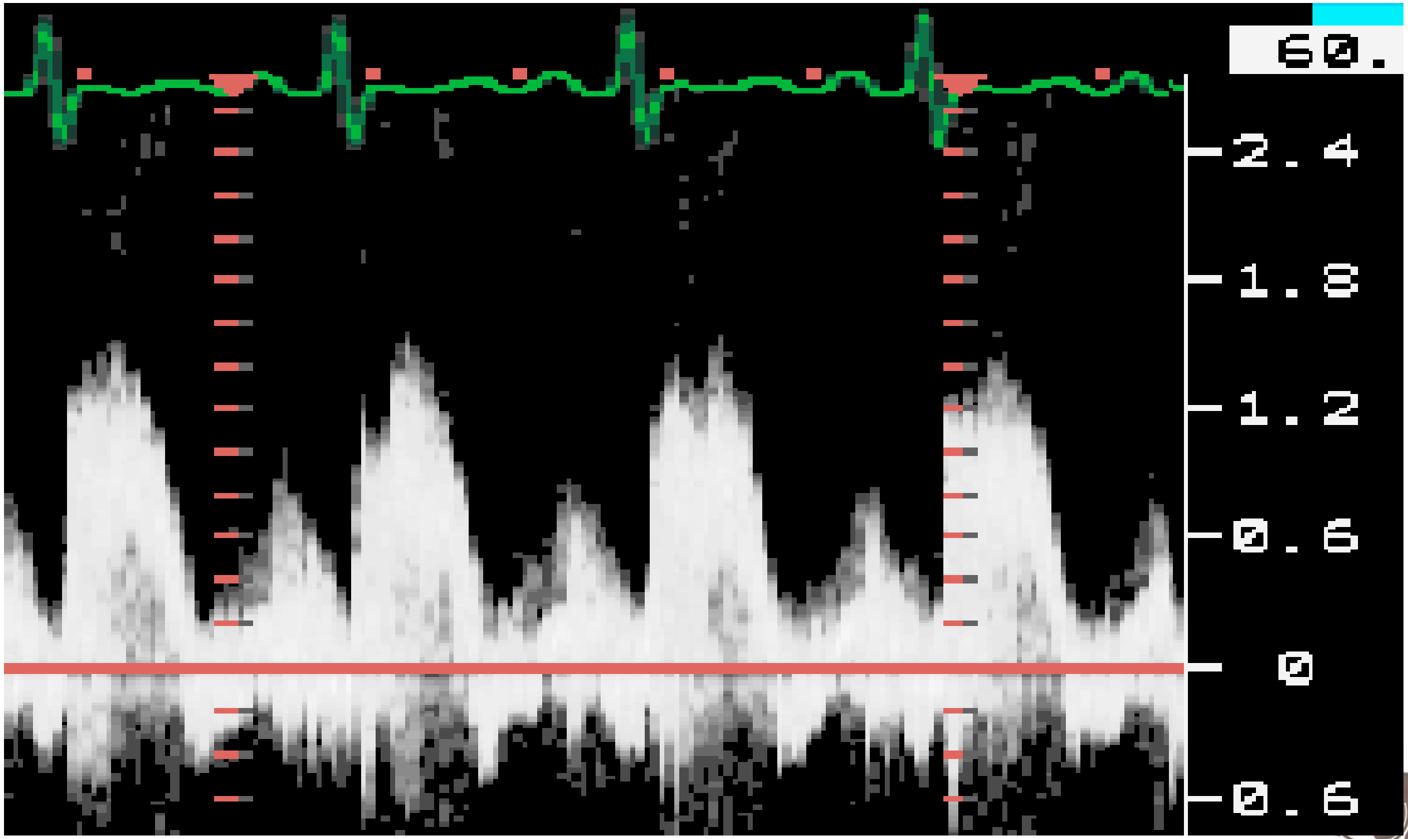
Continuous wave Doppler in ventricular septal defect The echocardiographic frame demonstrated the Doppler determination of pressure gradient across a membranous ventricular septal defect (VSD) (white arrow). The direction of the continuous wave Doppler beam used to obtain the velocity across the ventricular septal defect is illustrated by the red arrow. The velocity (V) is 4.5 m/sec and based upon the modified Bernoulli equation, (pressure = [velocity] 2 x 4) the gradient is 81 mmHg. (Courtesy of Ann Kavanaugh-McHugh, MD.)

Restrictive Membranous VSD



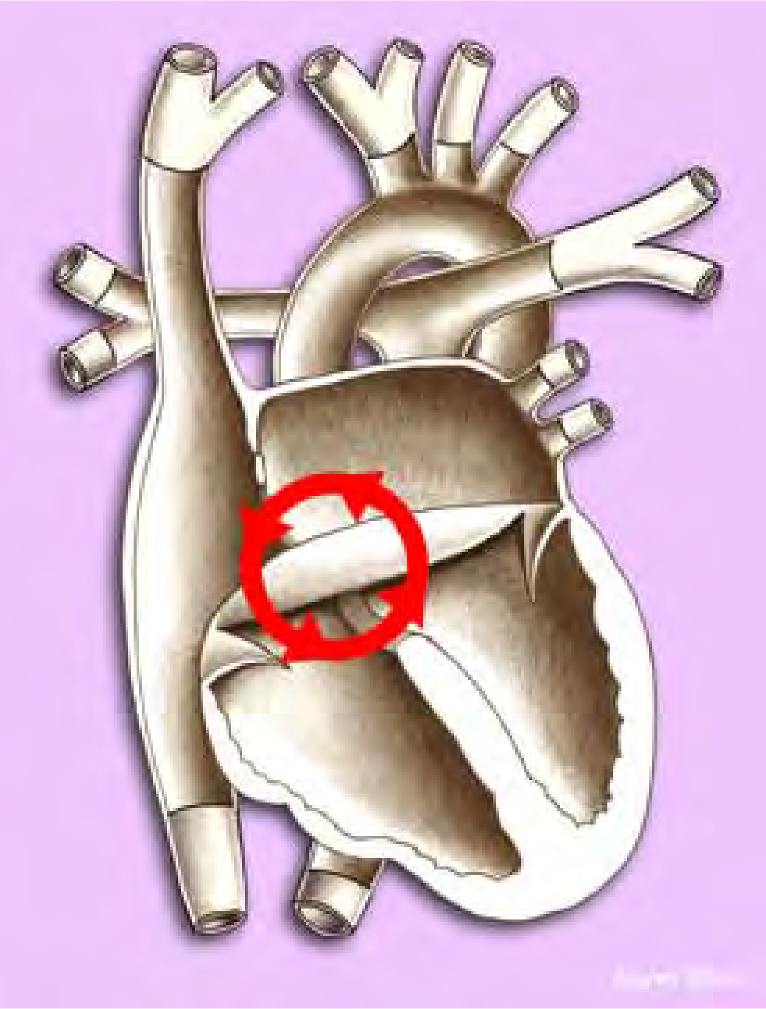
Children's National™

Unrestrictive Membranous VSD



Children's National™

Atrioventricular Canal Defect- Complete



Children's National™

Common AV Canal (CAVC)

Endocardial Cushion Defect (ECD)

Atrioventricular Septal Defect (AVSD)

Failure of the AV canal to develop properly and form tricuspid, mitral valves and portions of atrial and ventricular septae

Spectrum of defects

Children's National ™

Definitions

Incomplete CAVC = lack the VSD component or ASD component

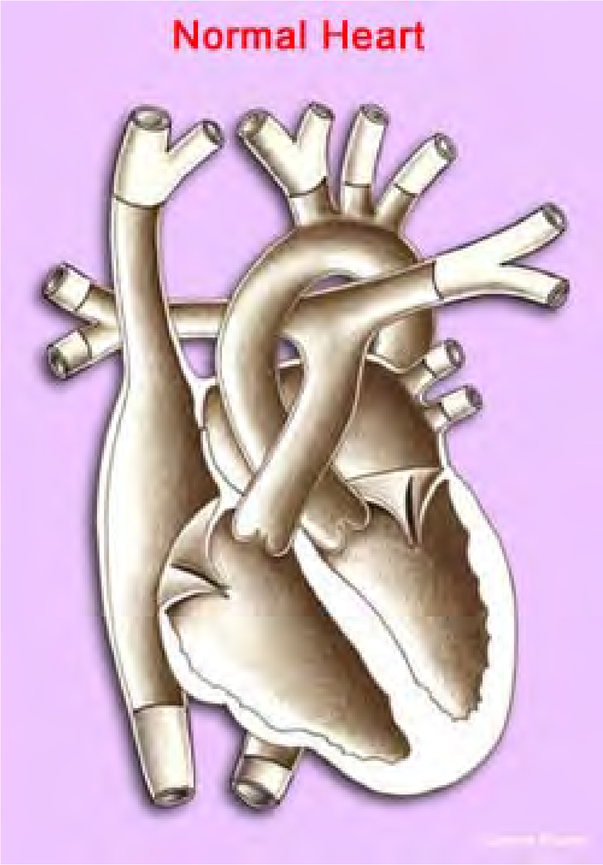
Partial CAVC = synonym for incomplete CAVC *OR* = primum ASD with cleft mitral valve

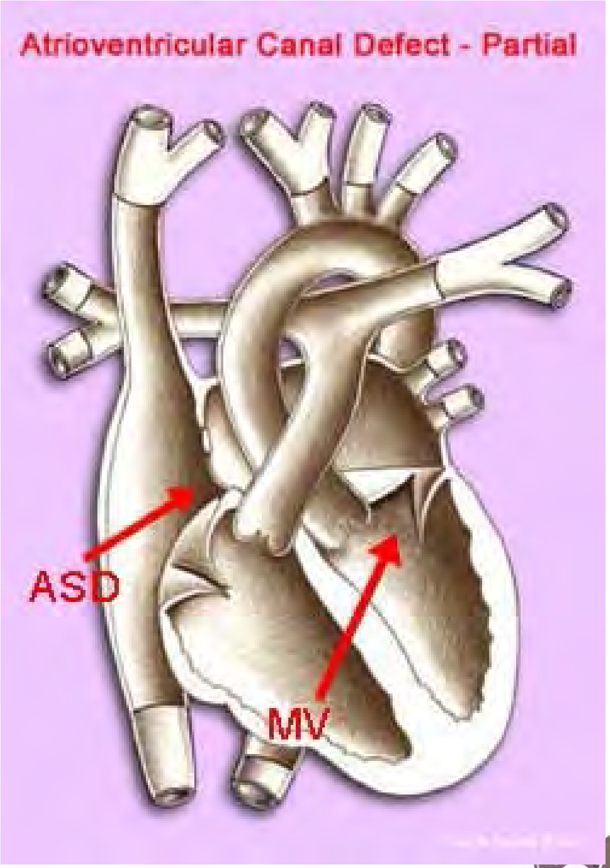
Transitional CAVC = small VSD component

Balanced/Unbalanced

Children's National™

Atrioventricular Canal Defect – Partial

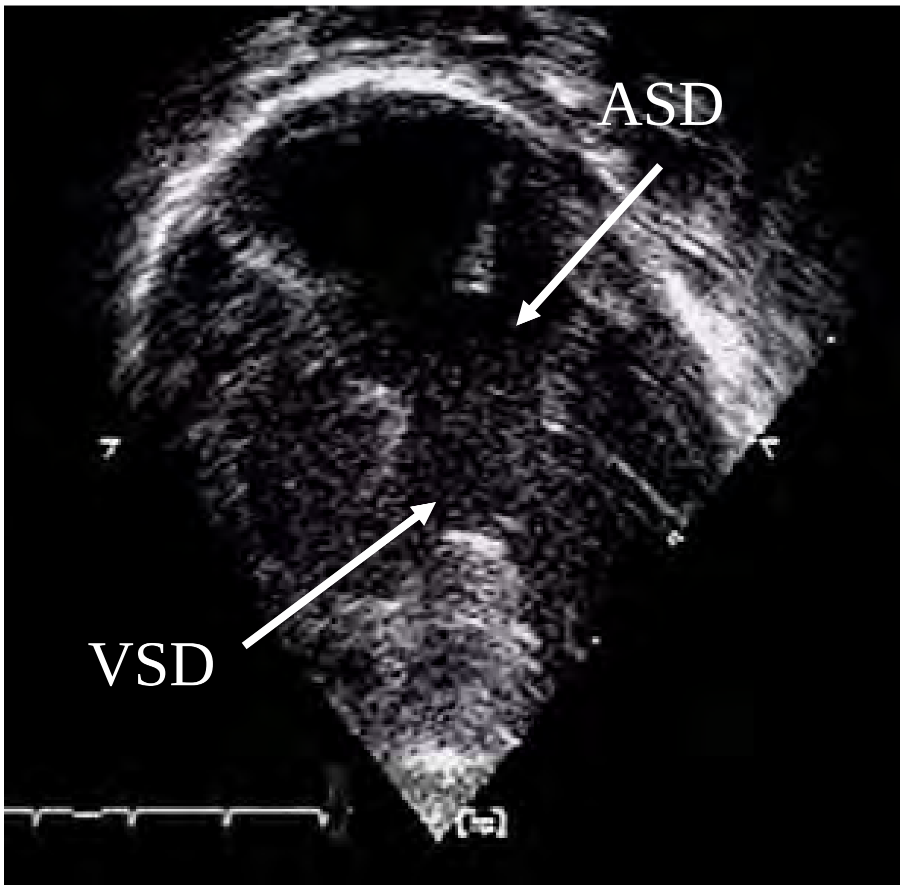


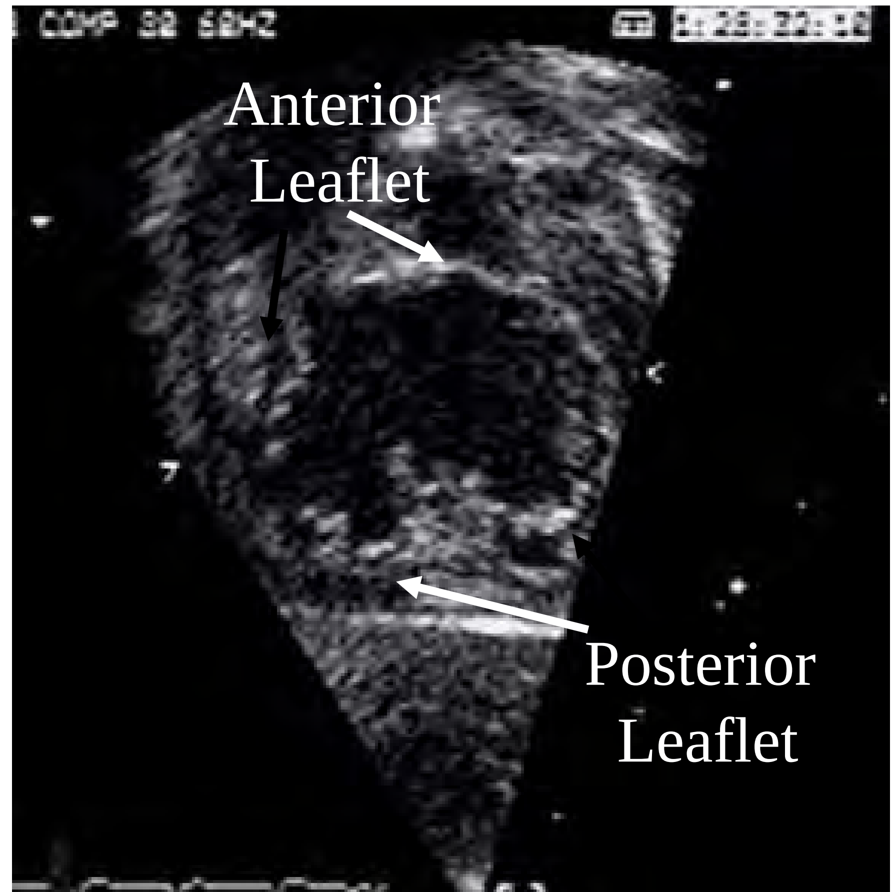


Children's National

AV Septal Defect

Complete





Children's National™

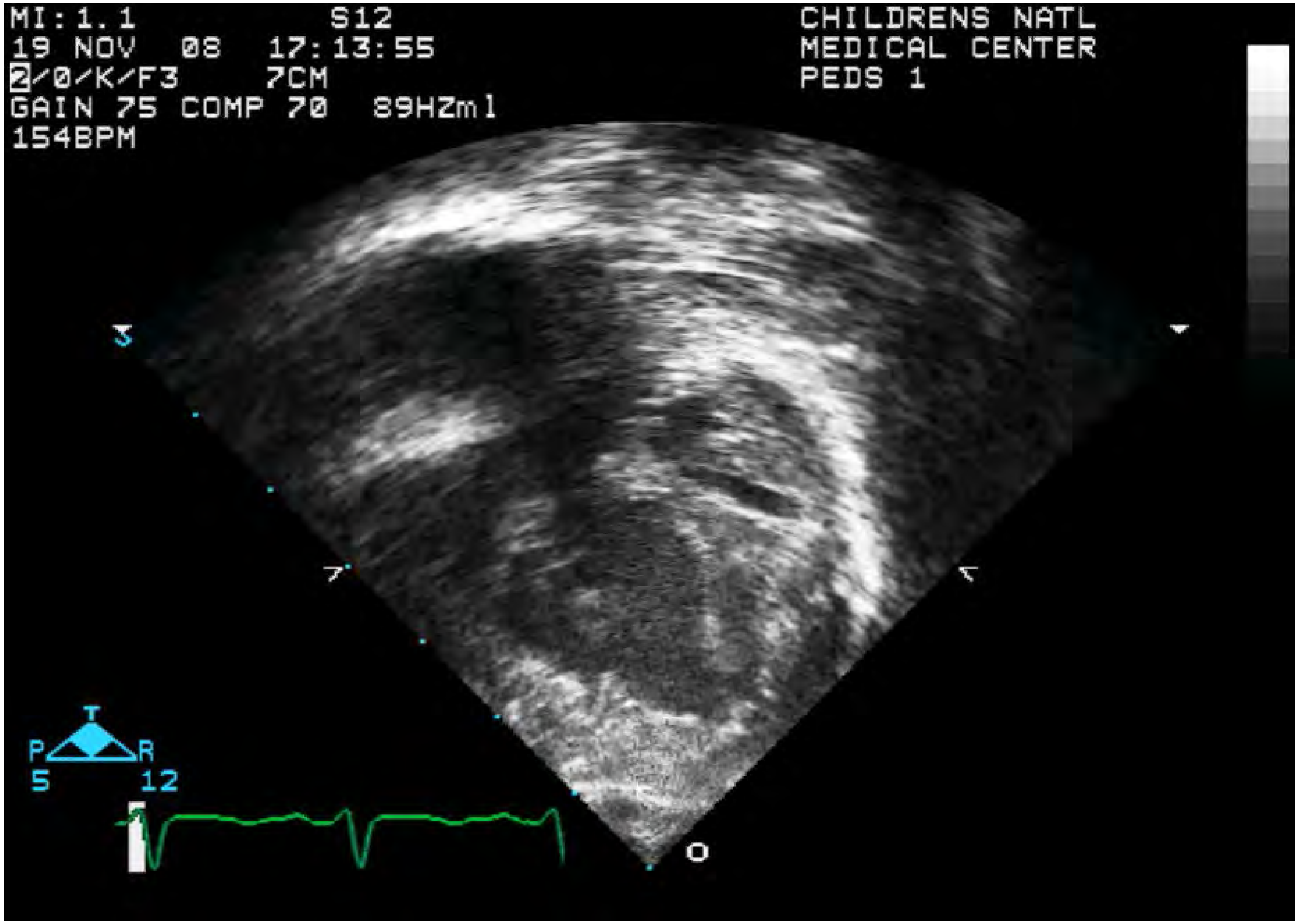

7/26/2008 11:24 AM
S12
26 JULY 08 10:44:33
PROC 2/0/F/F3 5CM
GAIN 75 COMP 70 89HZm1
145BPM
SHADY GROVE AHC
ECHO LAB
CNMC
1:12:00.03
T
P
R
5
12

Best View of CAVC

Children's National™


MI:0.9
S8
15 SEP 08
10:47:00
2/0/K/F3
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
[redacted]
ml
GAIN 78
COMP 70
157BPM
9CM
102HZ
T
P
R
3
8
0

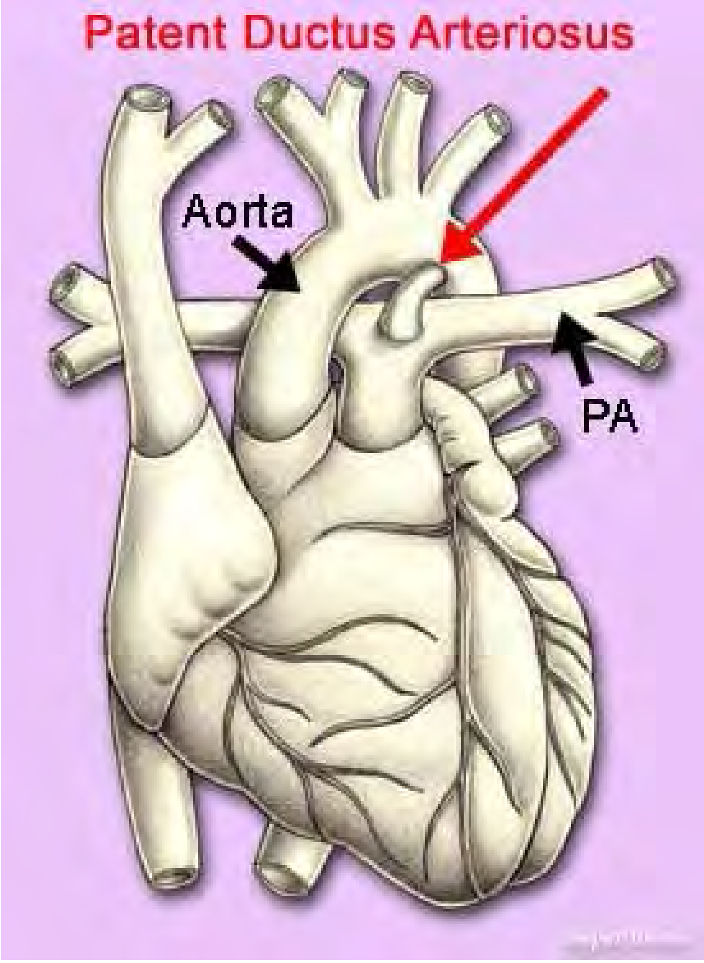
Unbalanced

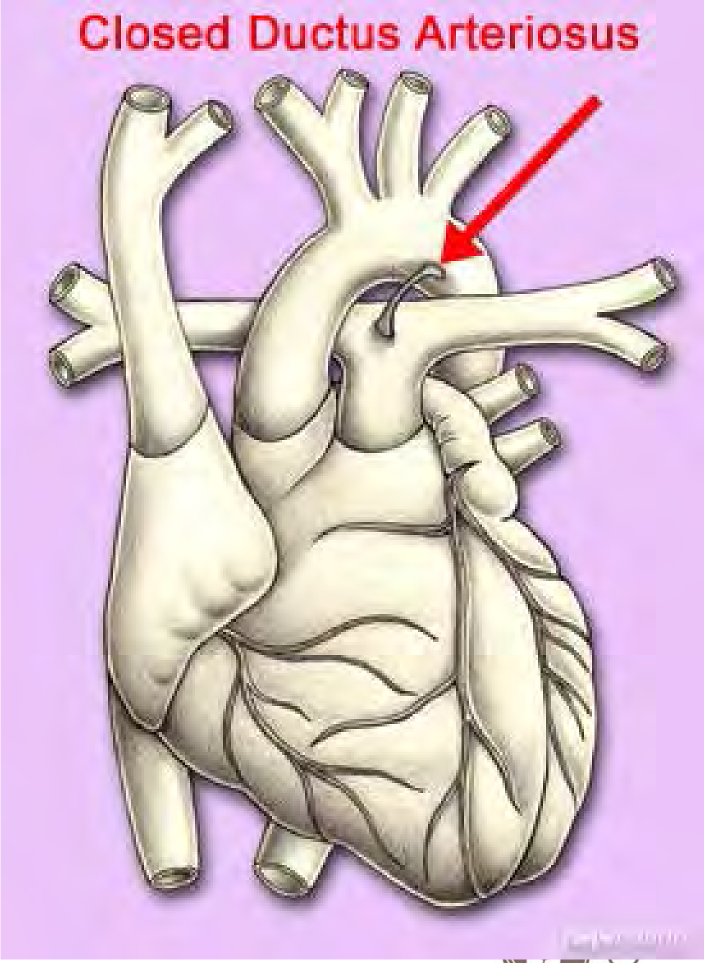


Children's National™

Children's National ™

Patent Ductus Arteriosus





Children's National

PDA: Clinical Correlation

Closed in 90% of infants by 48 hours of life

- Prematuring, altitude

Anatomy

- Derived from the left 6th embryonic arch

Closure

- Muscular constriction→endothelium→thrombosis→fibrous strand

Physiology↔ shunting

- Symptoms proportional to shunting

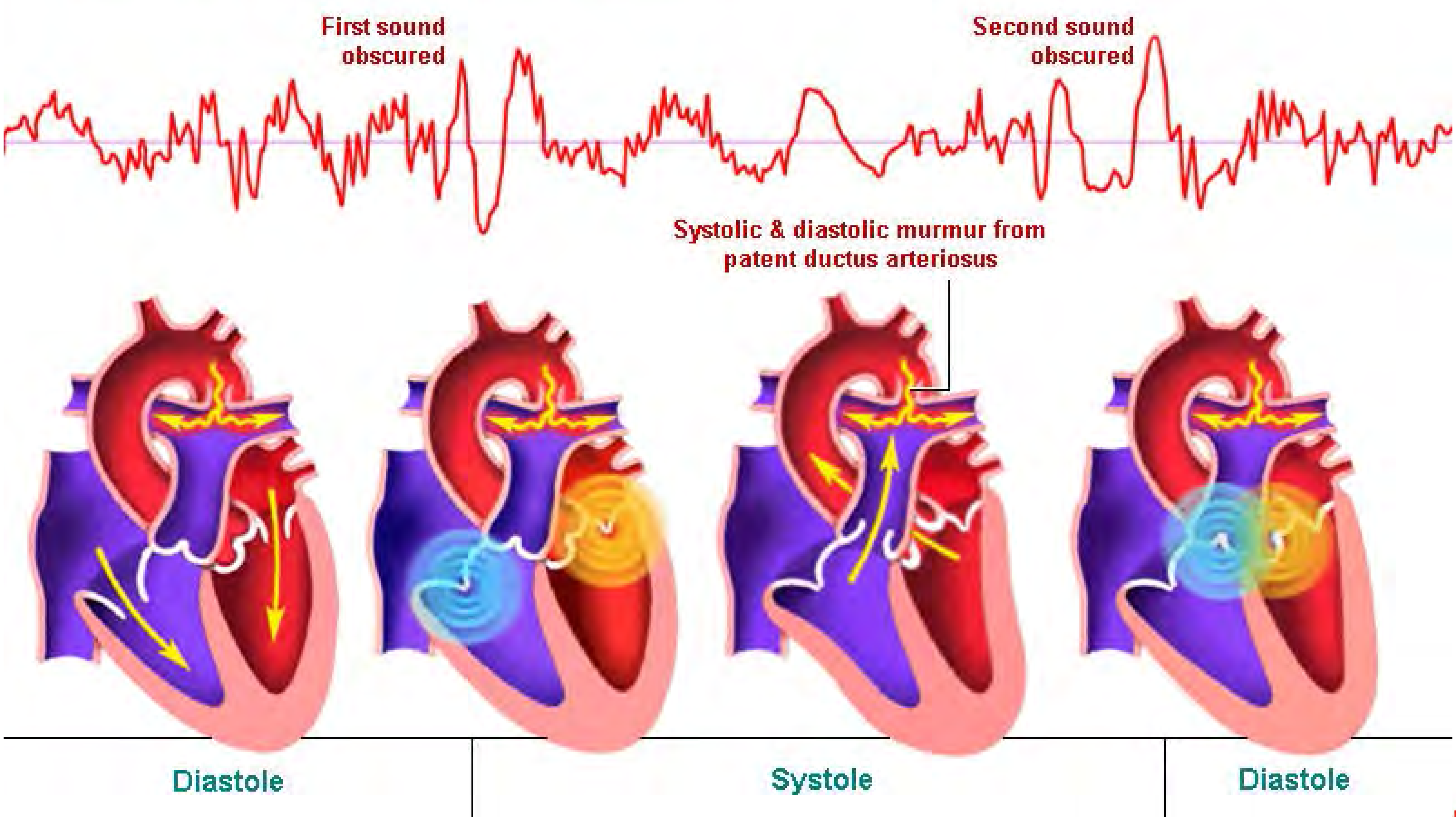
Murmur

EKG

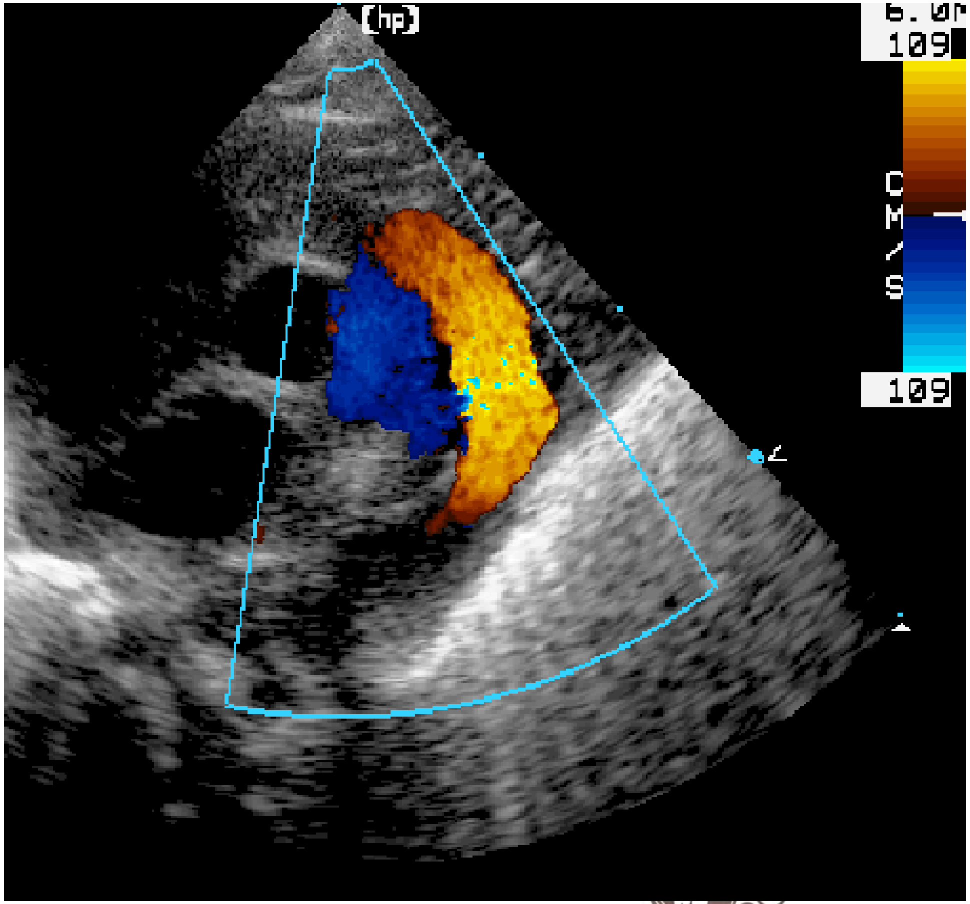
- Ventricular hypertrophy

(.wav)

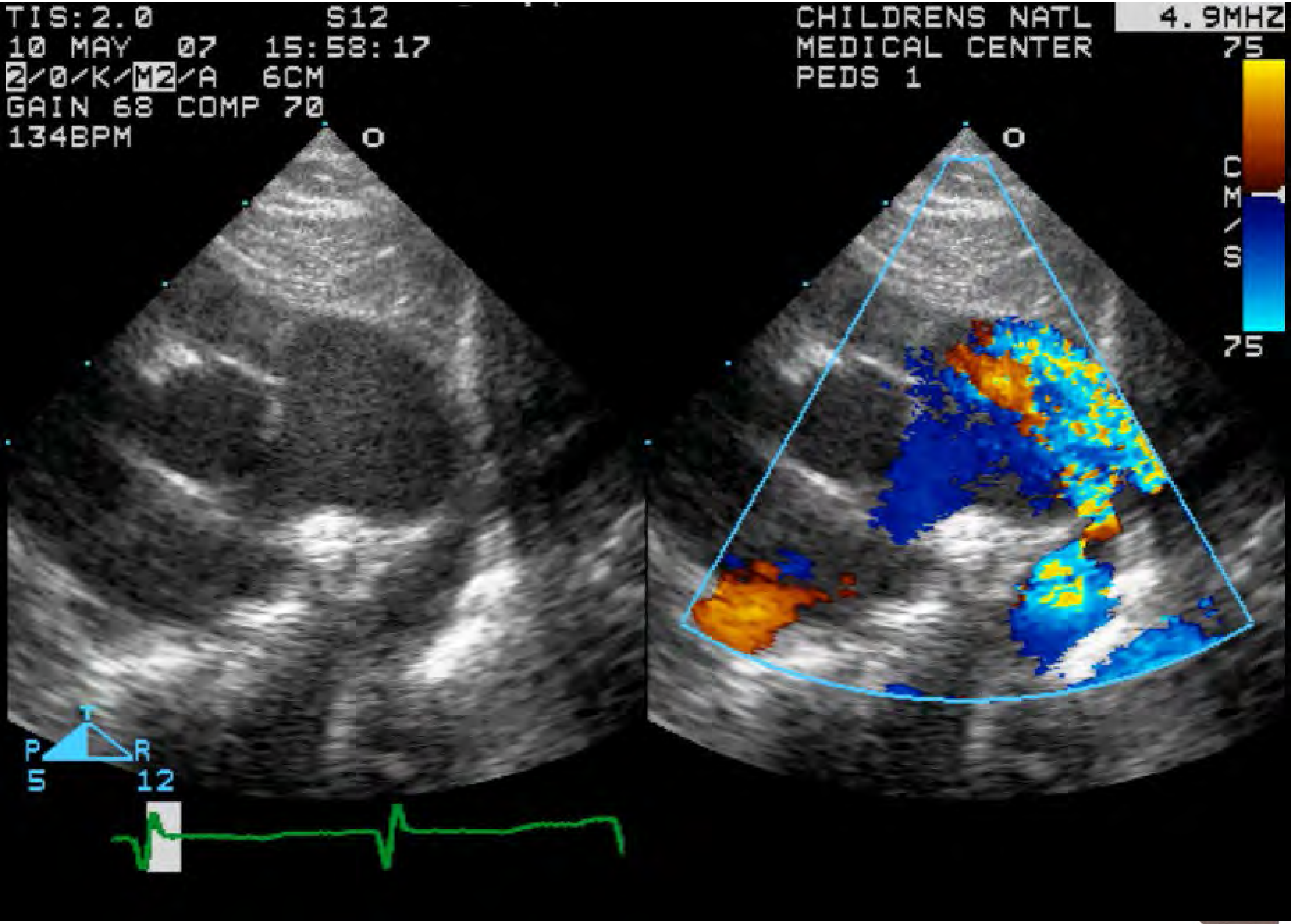
Patent Ductus Arteriosus



Patent Ductus Arteriosus

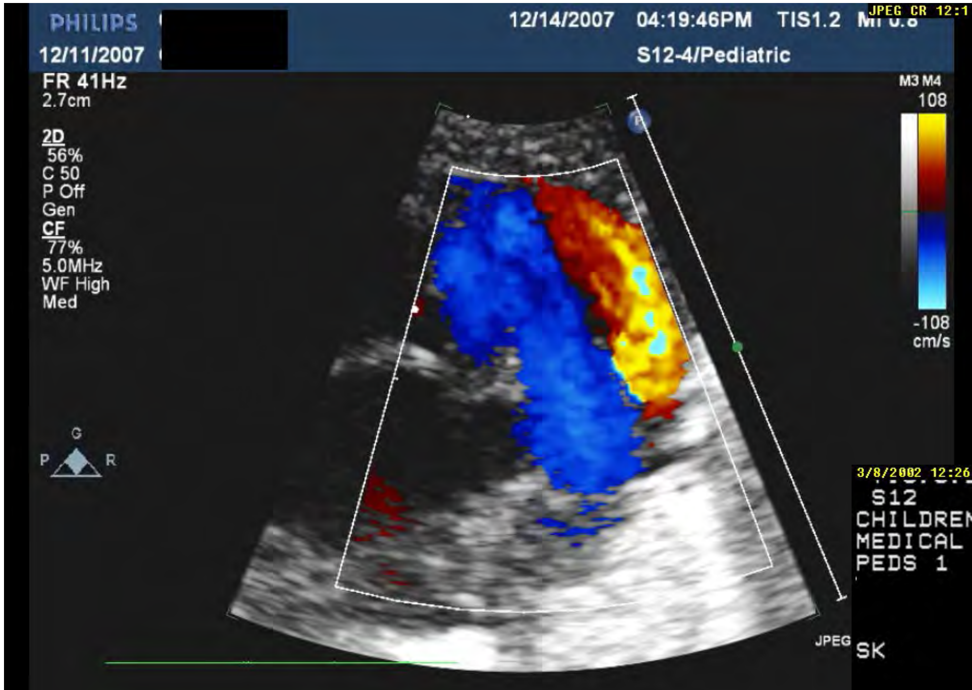


Children's National ™

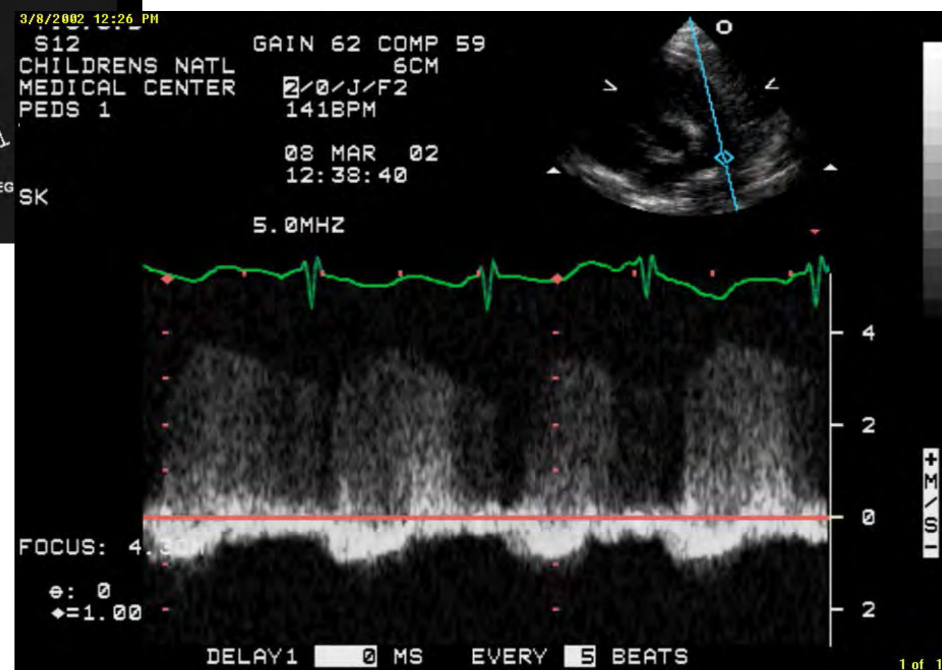


Children's National™

Doppler of the PDA (L-R shunt)

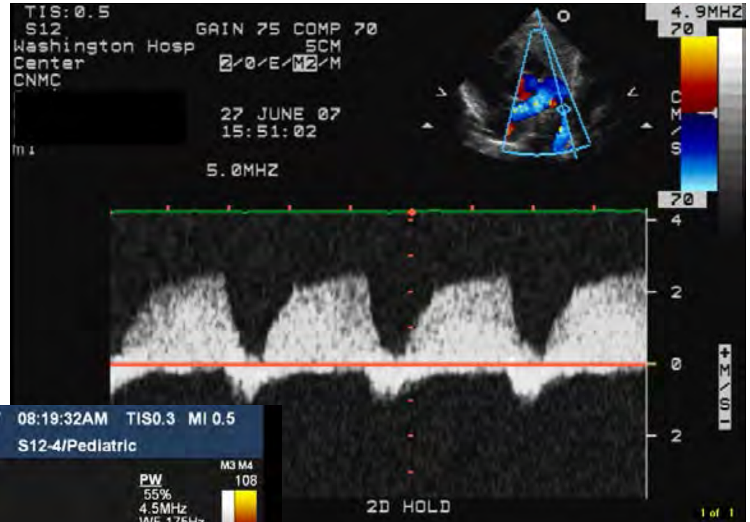


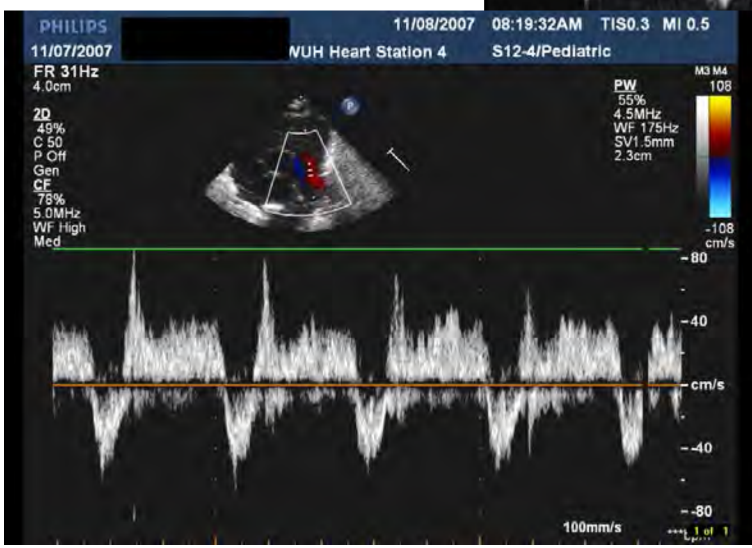
Color flow Doppler (left) showing a L-R shunt from the descending aorta through the PDA to the PA (red: towards the probe)



CW Doppler tracing (right) seen above the baseline indicating flow toward the probe from the descending aorta through the PDA to the PA. The peak velocity is reached in late systole 4 m/s. L-R shunt

Doppler of the PDA (bidirectional shunt)



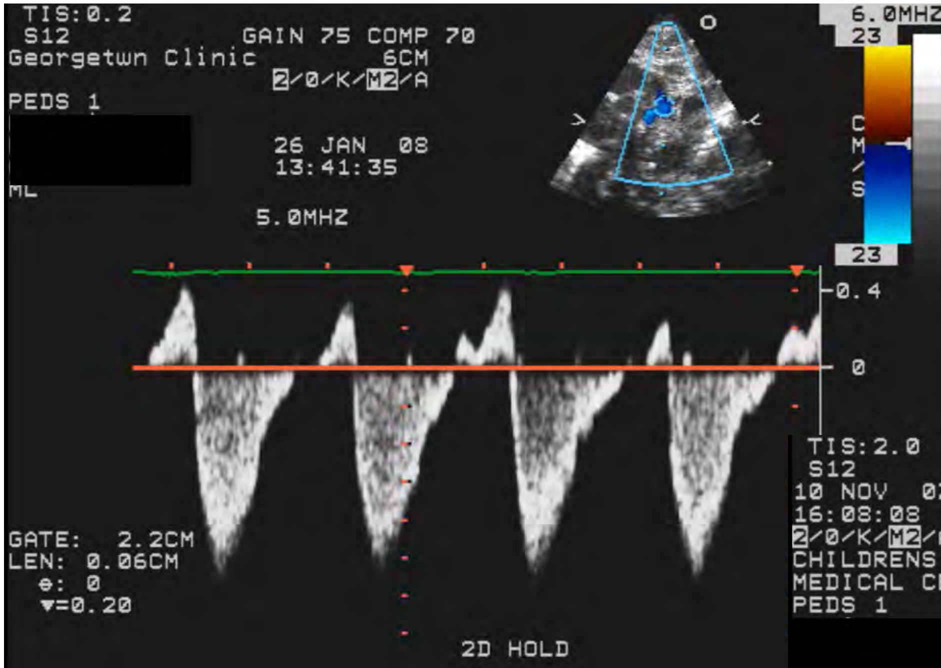


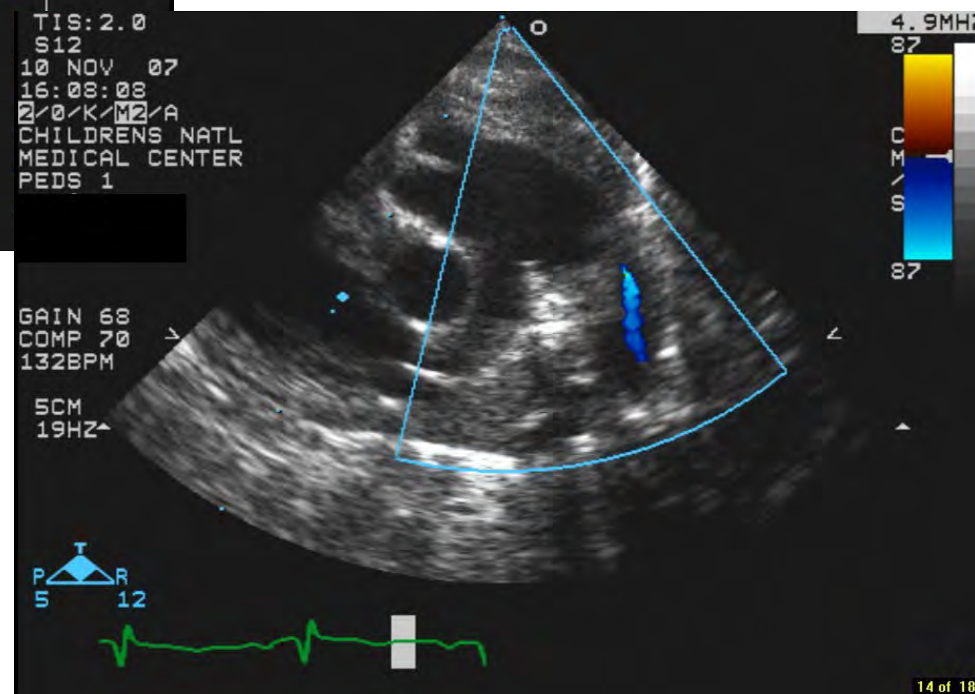
Bidirectional blood flow through the PDA can be a normal finding in newborn infants due to high pulmonary resistance

CW Doppler from an infant with pulmonary artery hypertension and PDA. The negative deflection in systole below the baseline arises from the R-L shunt through the PDA from the PA to the Dao (away from the TDX).
The positive deflection (late systole-into late diastole) arises from L-R shunt through the PDA from the Dao to the PA

Children's National

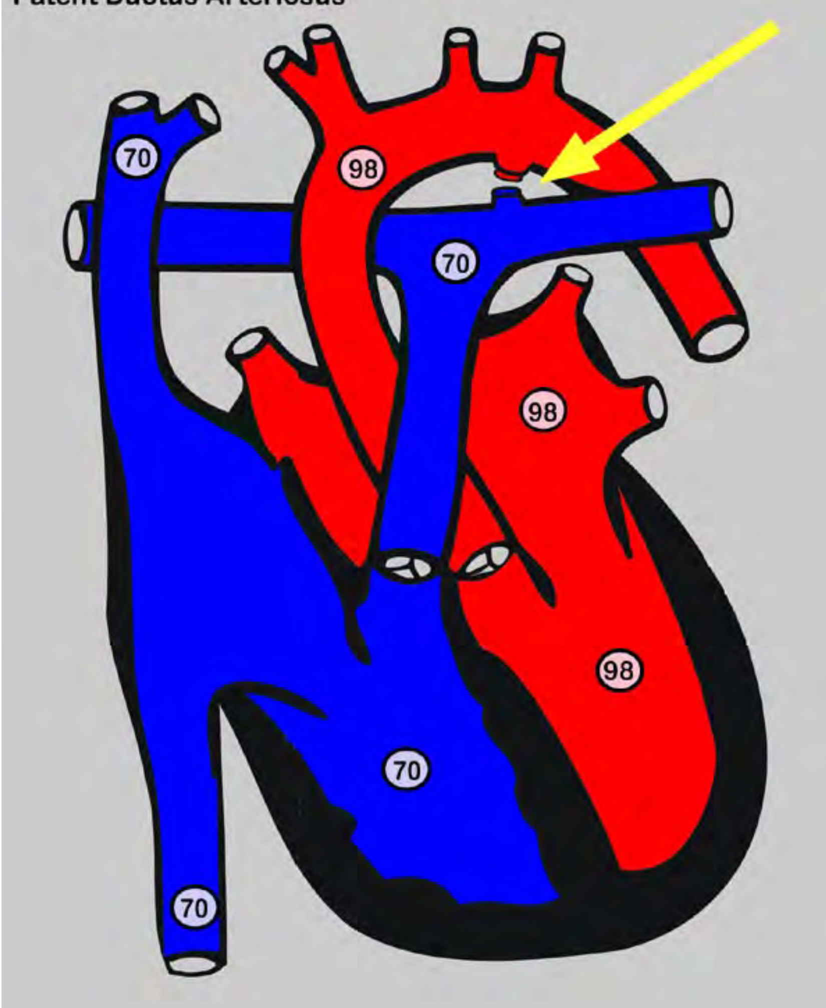
Doppler of the PDA (R-L shunt)

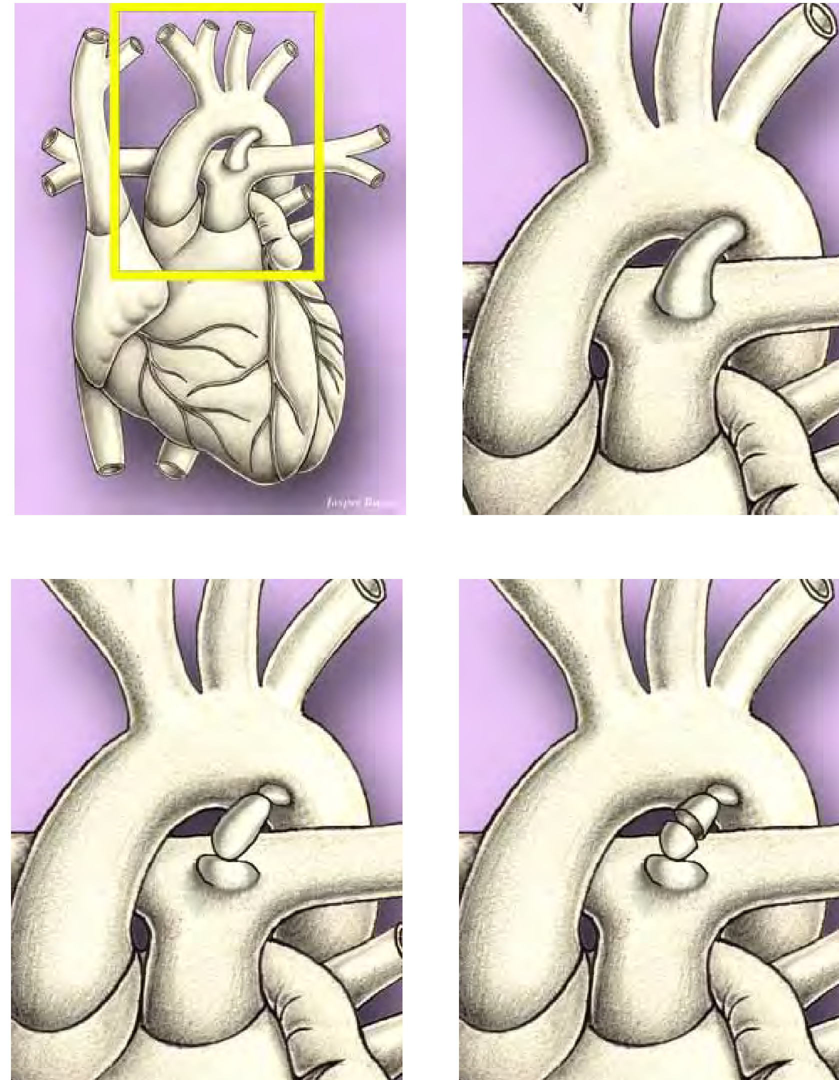




The Doppler spectral tracing shows evidence of severe pulmonary hypertension and no evidence of a L-R shunt through the PDA. The shunt is R-L from the ductus arteriosus to the Dao (blue: away from the TDX)

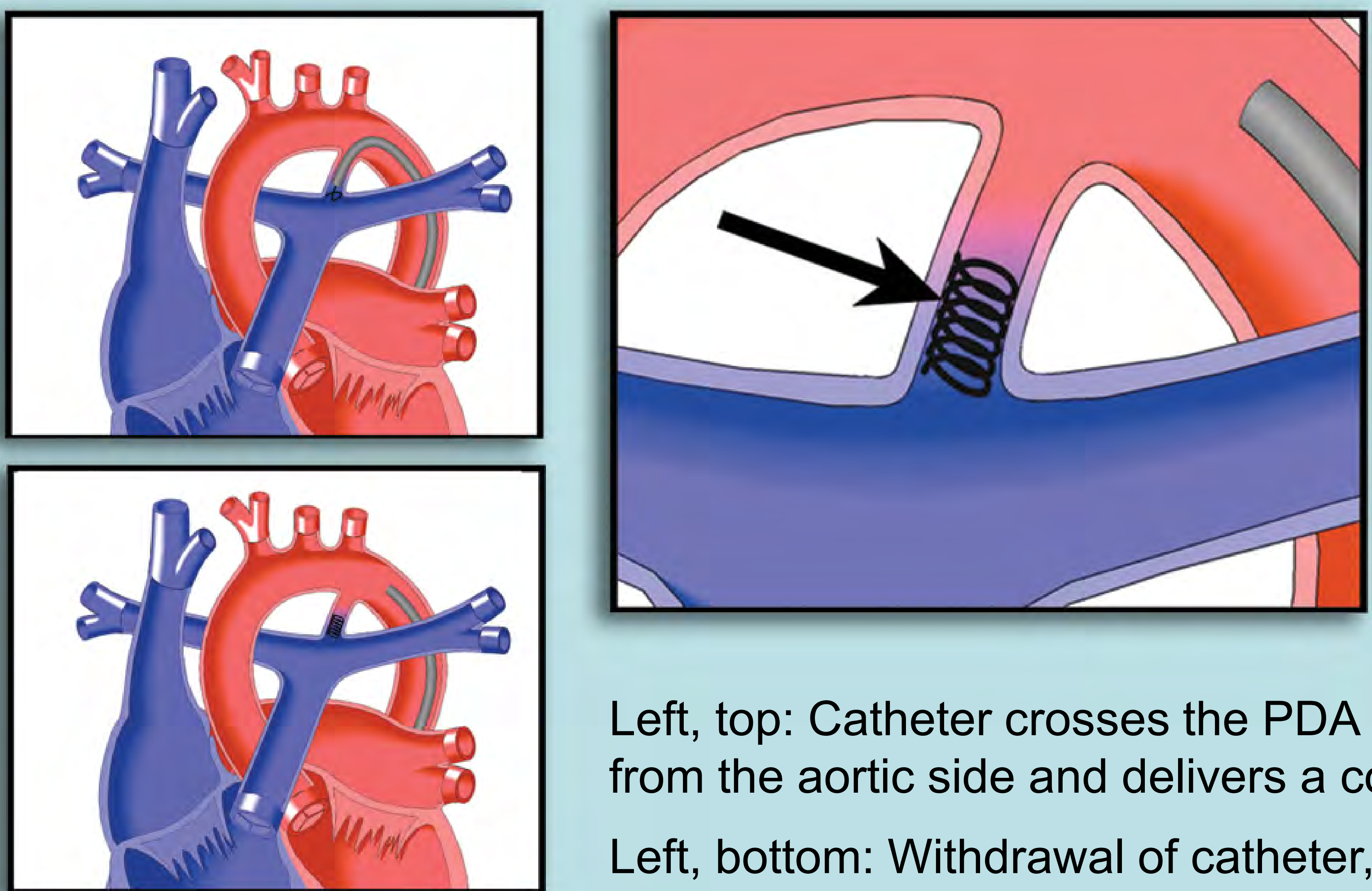
Patent Ductus Arteriosus – Ligation and Division





Occlusion of Intracardiac and Vascular Shunts

Coil embolization of PDA



Left, top: Catheter crosses the PDA from the aortic side and delivers a coil.

Left, bottom: Withdrawal of catheter, leaving coil in PDA

Amplatzer Ductal Occluders

Amplatzer ductal occluder

Illustration courtesy AGA Medical Group

Aorta angiogram with device occlusion of PDA, lateral view

Children's National ™

Right Heart Obstructive Lesions

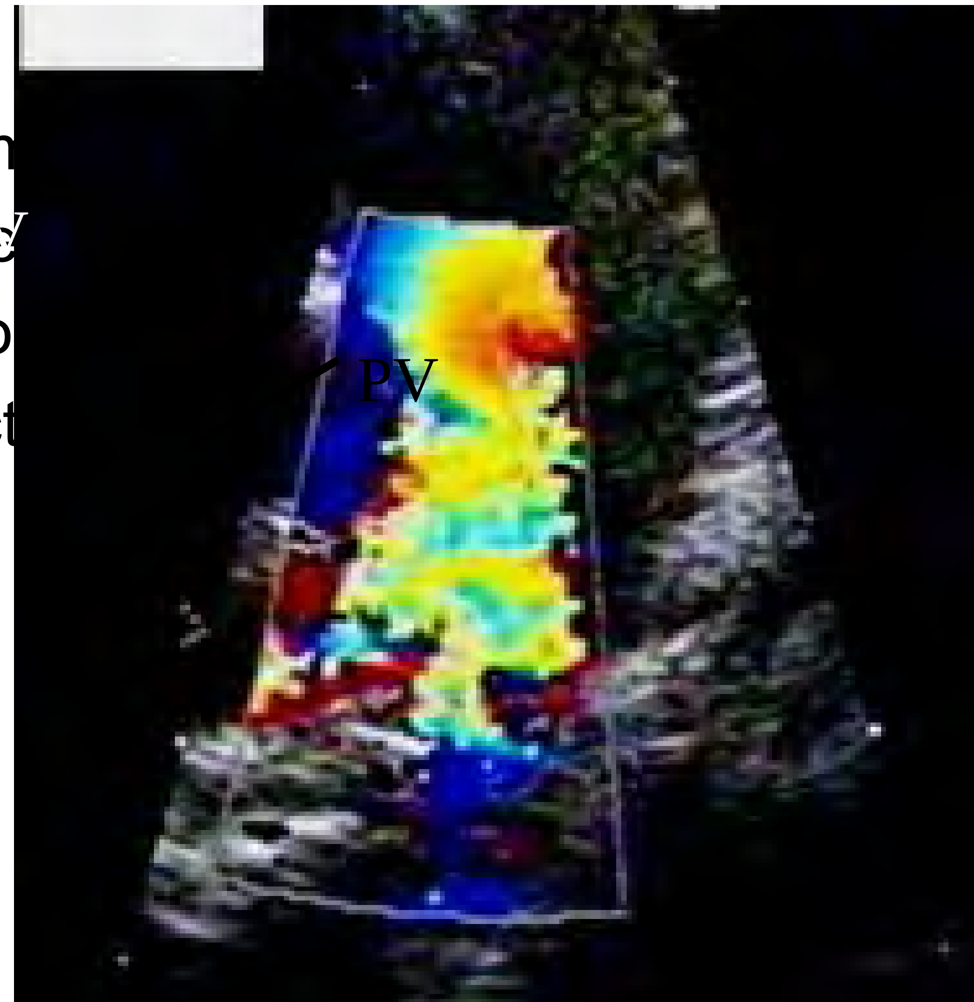
Pulmonary Valve Stenosis

Valve anatomy

- Doming, fused comn
- Thickened, immobile
- Subvalvar obstructio
- Supravalvar obstruct

Post stenotic dilation

RVH



PS: Clinical Correlation

Asymptomatic

Murmur at birth

EKG

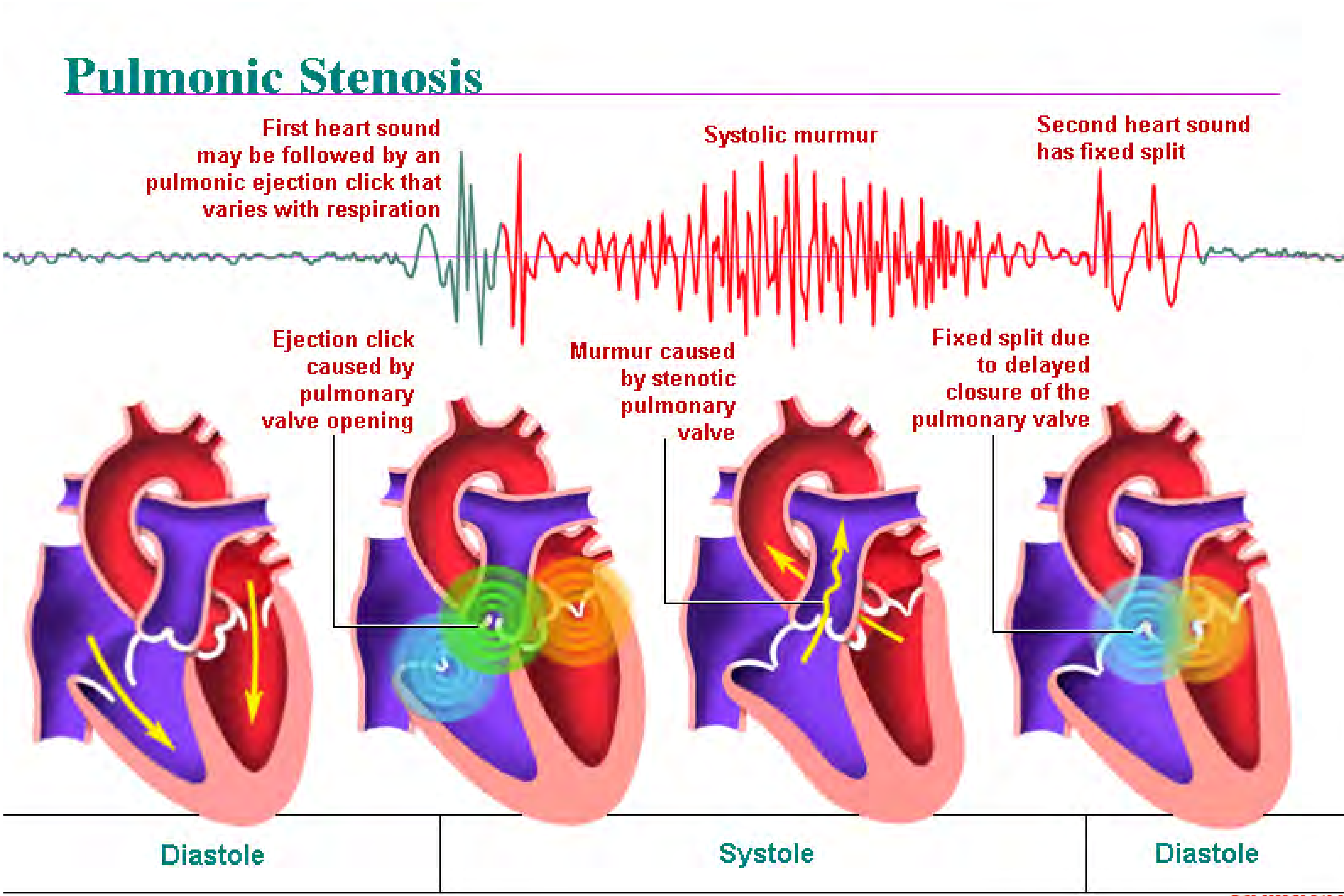
- RAD, RVH proportional to obstruction

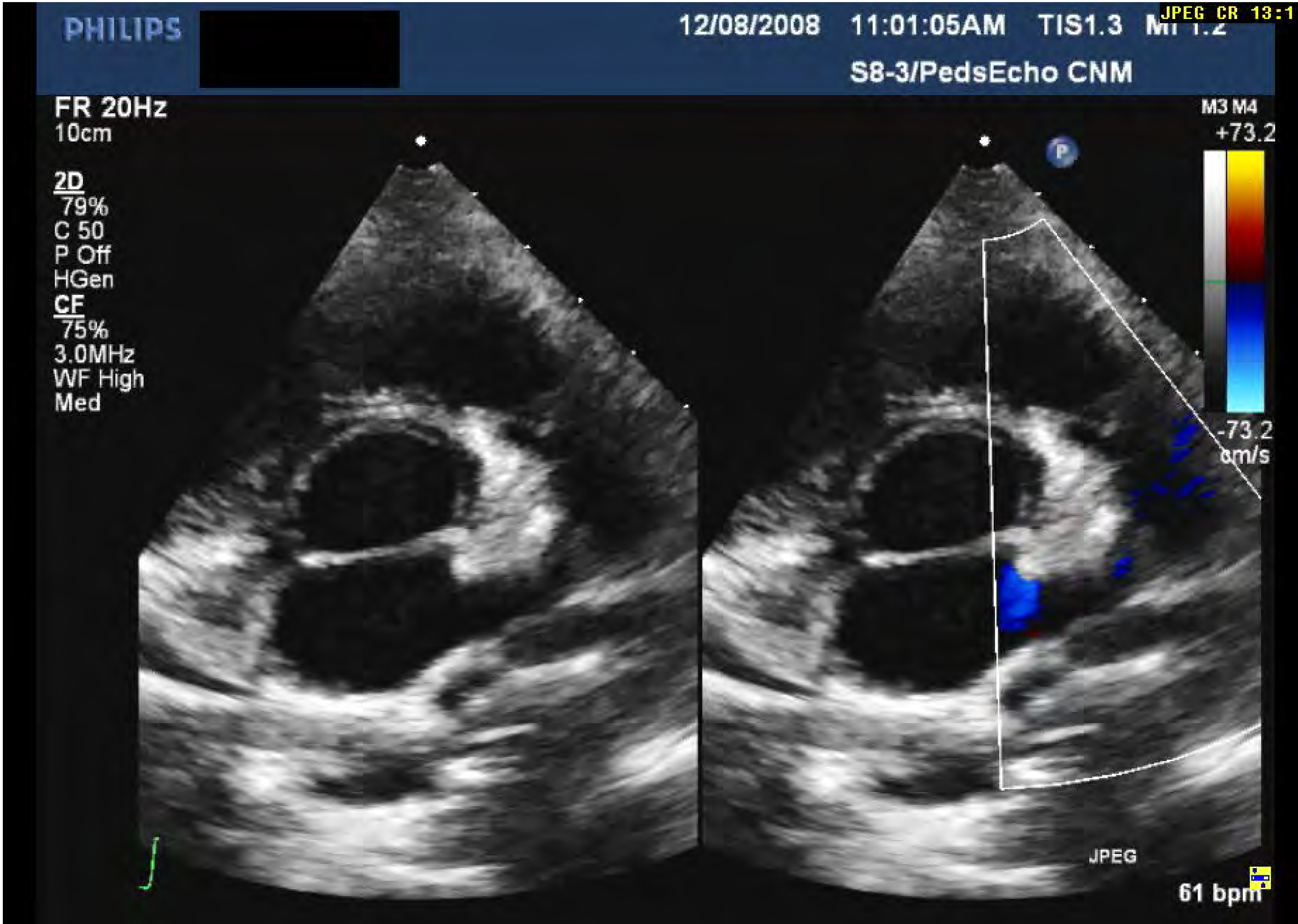
Management

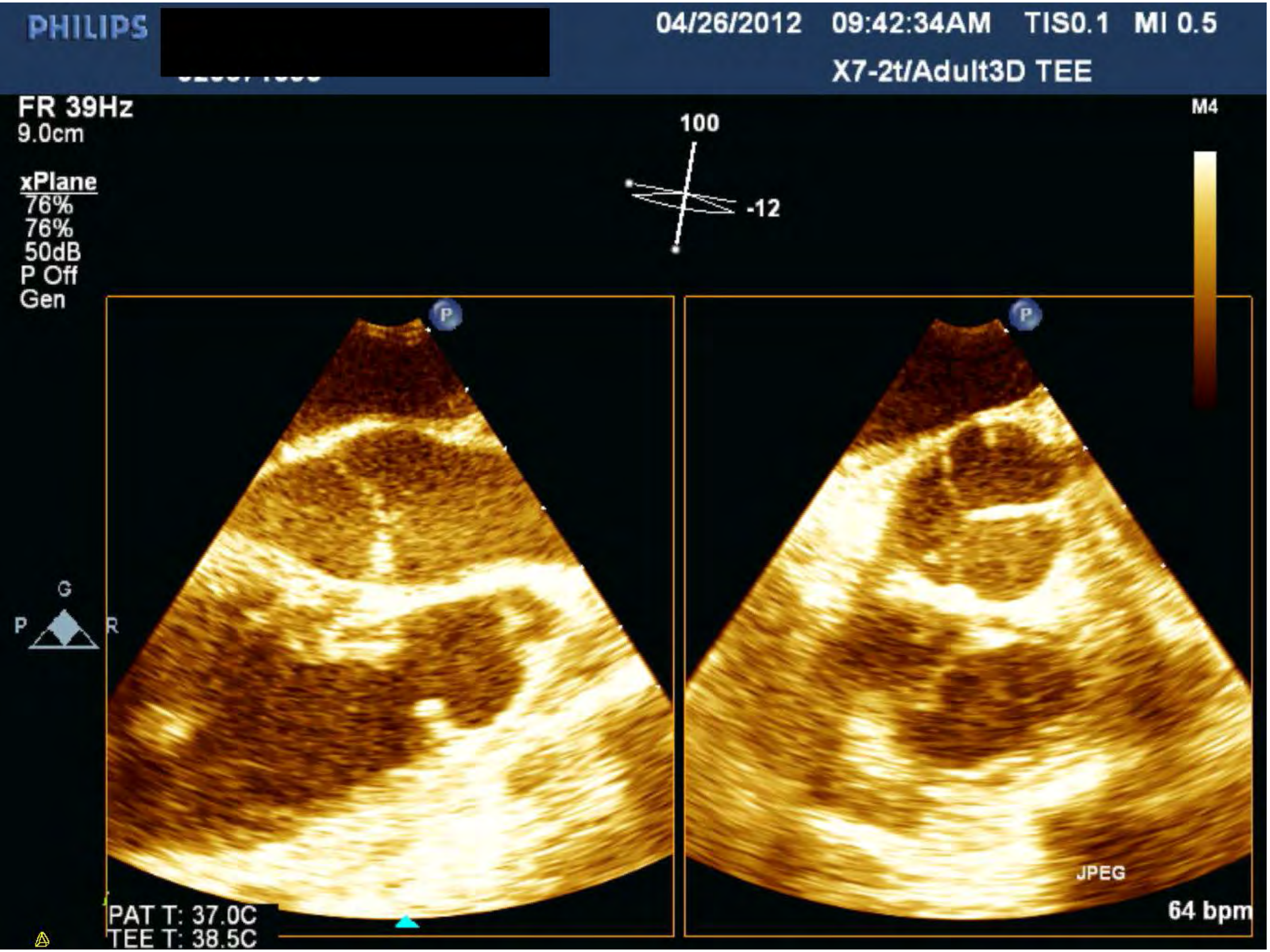
- Balloon dilation

Excellent outcome

(.wav)

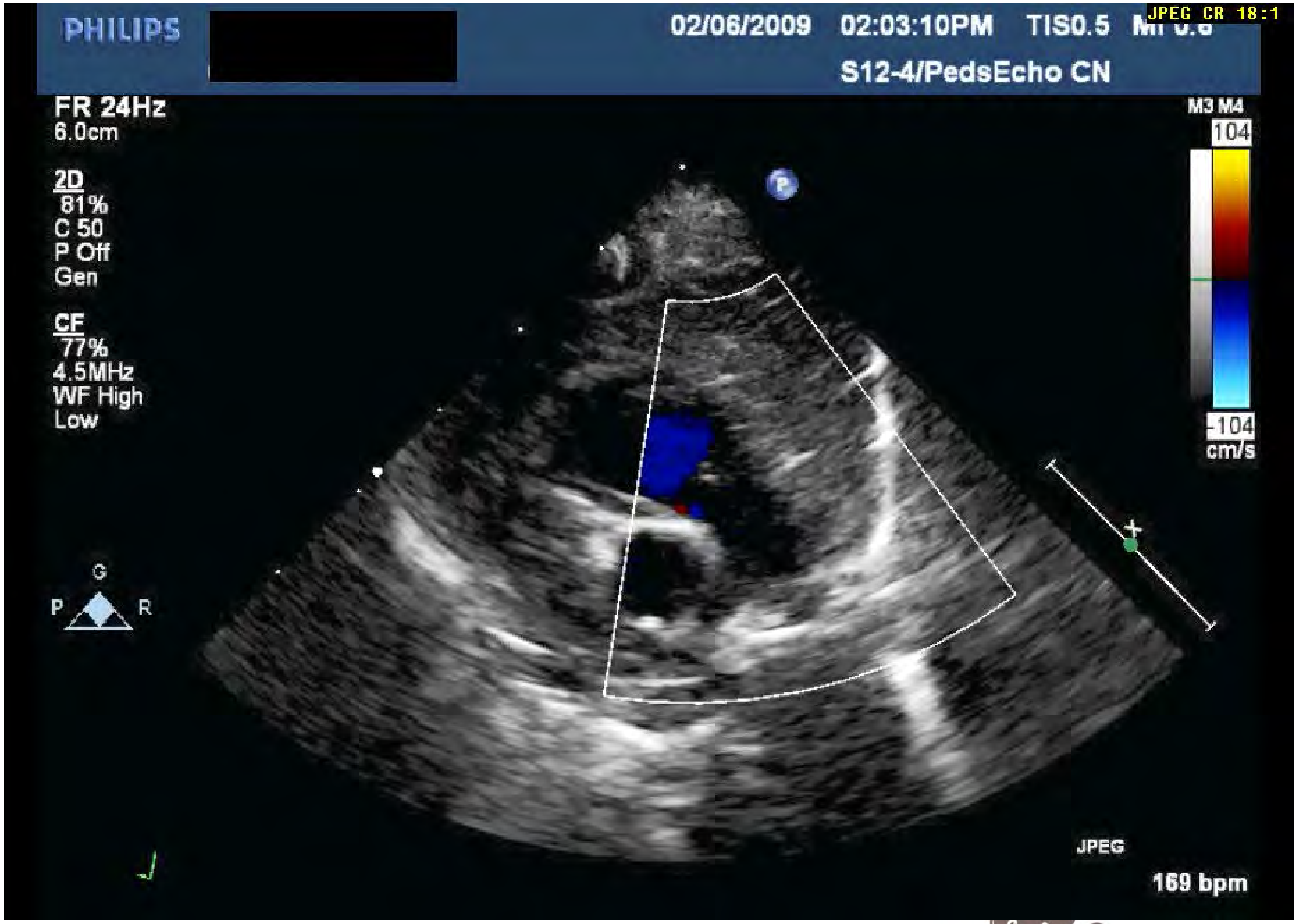

Pulmonic Stenosis
First heart sound may be followed by an pulmonic ejection click that varies with respiration
Systolic murmur
Second heart sound has fixed split
Ejection click caused by pulmonary valve opening
Murmur caused by stenotic pulmonary valve
Fixed split due to delayed closure of the pulmonary valve
Diastole
Systole
Diastole


PHILIPS
[redacted]
12/08/2008 11:01:05AM TIS1.3
JPEG CR 13:1
S8-3/PedsEcho CNM
FR 20Hz
10cm
2D
79%
C 50
P Off
HGen
CF
75%
3.0MHz
WF High
Med
M3 M4
+73.2
-73.2
cm/s
JPEG
61 bpm



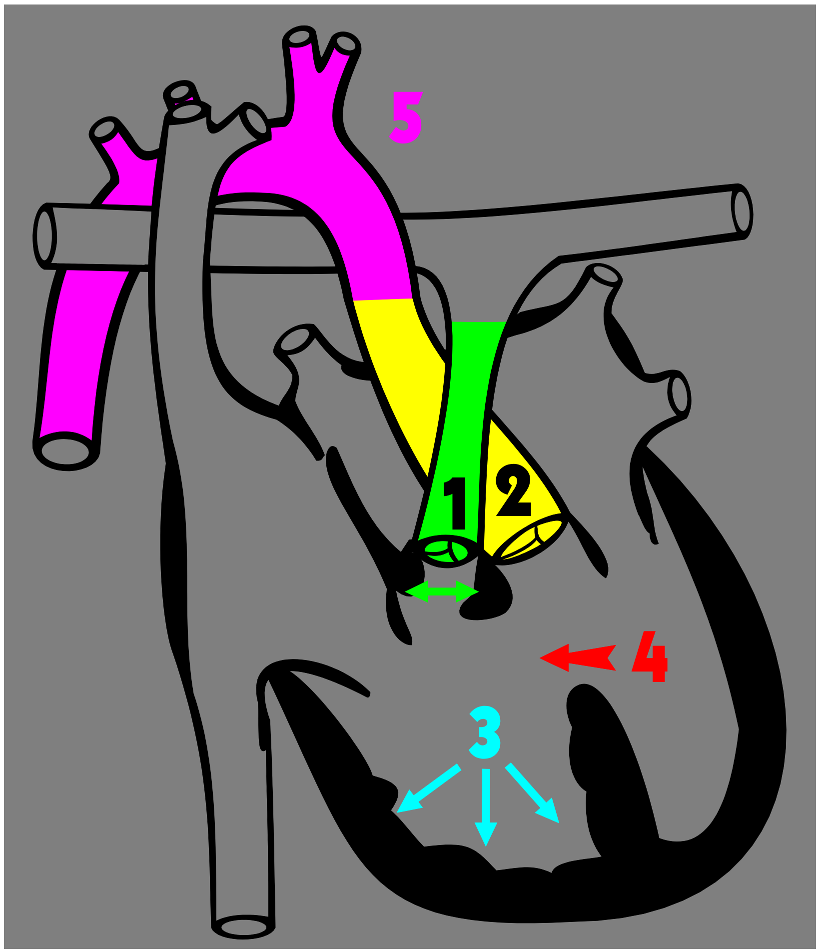
Children's National ™

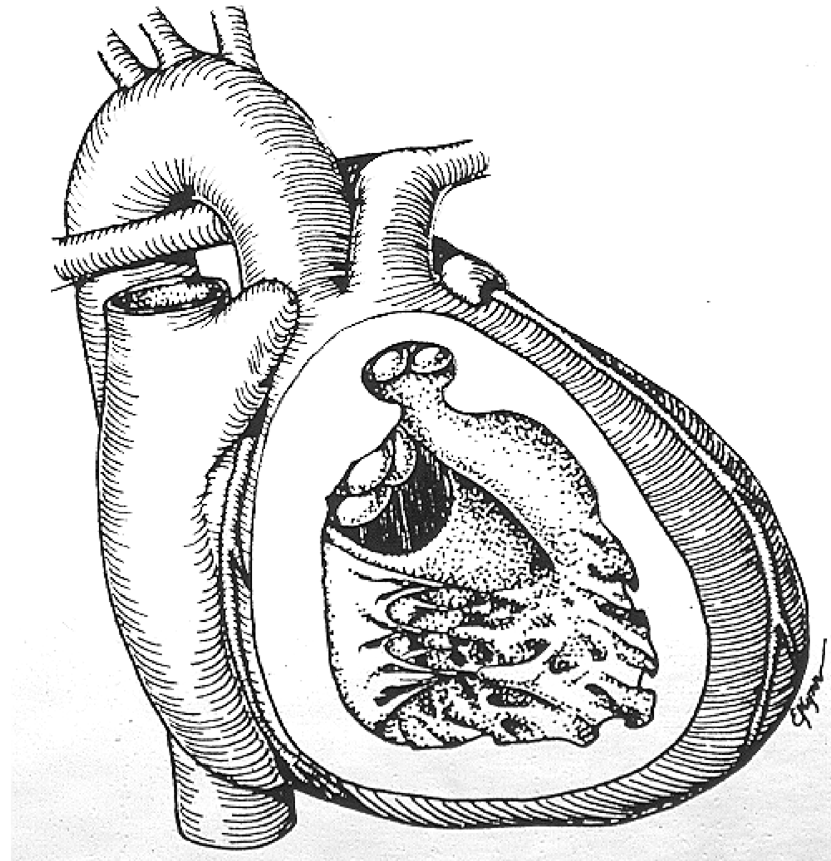
Pulmonary Artery Branch Stenosis



Children's National™

Tetralogy of Fallot





Children's National™

Tetralogy of Fallot

"Maladie Bleu" 1888

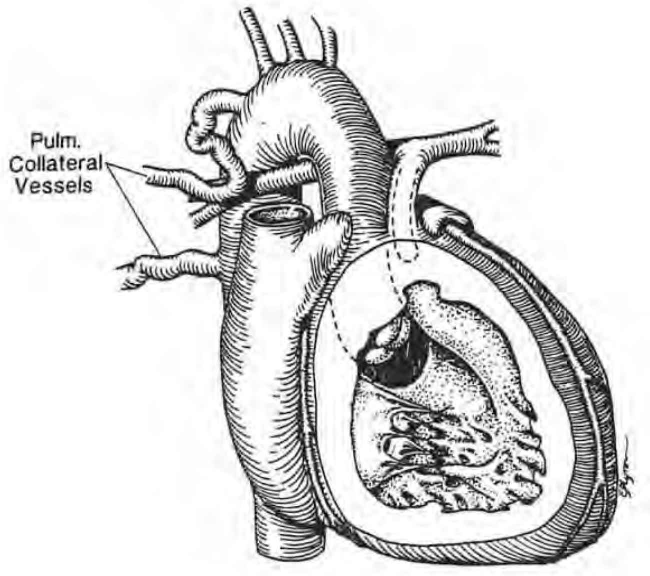
Stensen 1671

Sandifort 1777

Arthus Louis Etienne FALLOT

Children's National ™

Variations of Tetralogy



Tet, pulmonary atresia:

MAPCAS

“Mexican Tet”

- Hypoplastic or absent conal septum

Tetralogy with absent pulmonary valve

- Rudimentary pulmonary valve leaflets result in fetal pulmonary regurgitation, PA dilation
- Airway and lung development is compromised in severe cases

Tetralogy with CAVC

TOF: Clinical Correlation

Most common cyanotic defect

Defective neural crest migration resulting in abnormal conotruncal development

Clinical presentation depends on degree of subpulmonary narrowing

- This may change over time

Presentation

- Fetal dx
- Murmur

TOF: Clinical Correlation

Cyanosis due to right to left shunting at ventricular level

Degree of cyanosis is proportional to amount of RVOTO

Dynamic factors may worsen cyanosis

- Tet Spell→ no murmur→ deeply cyanotic

EKG

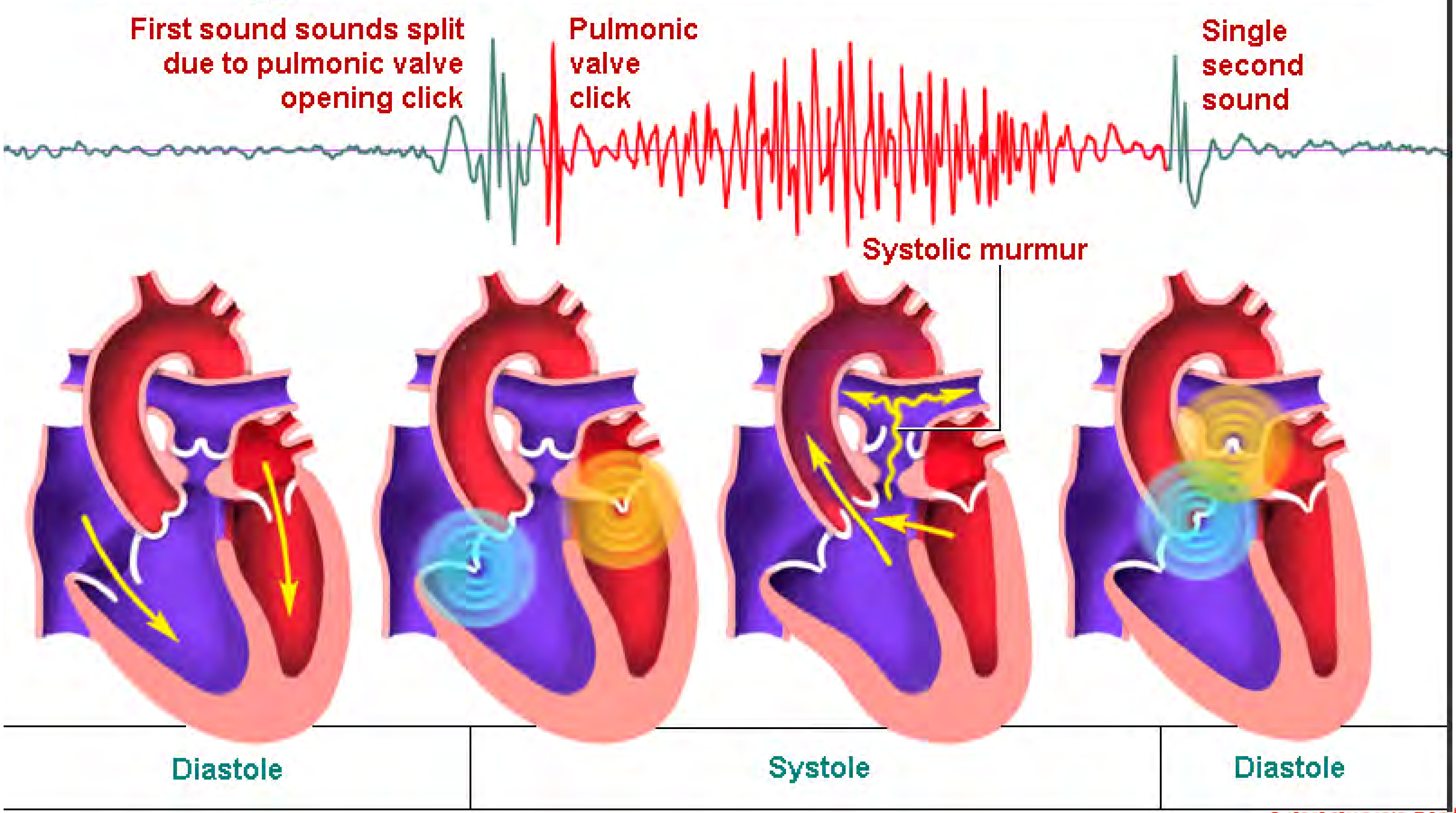
- RVH, RAD, RAE

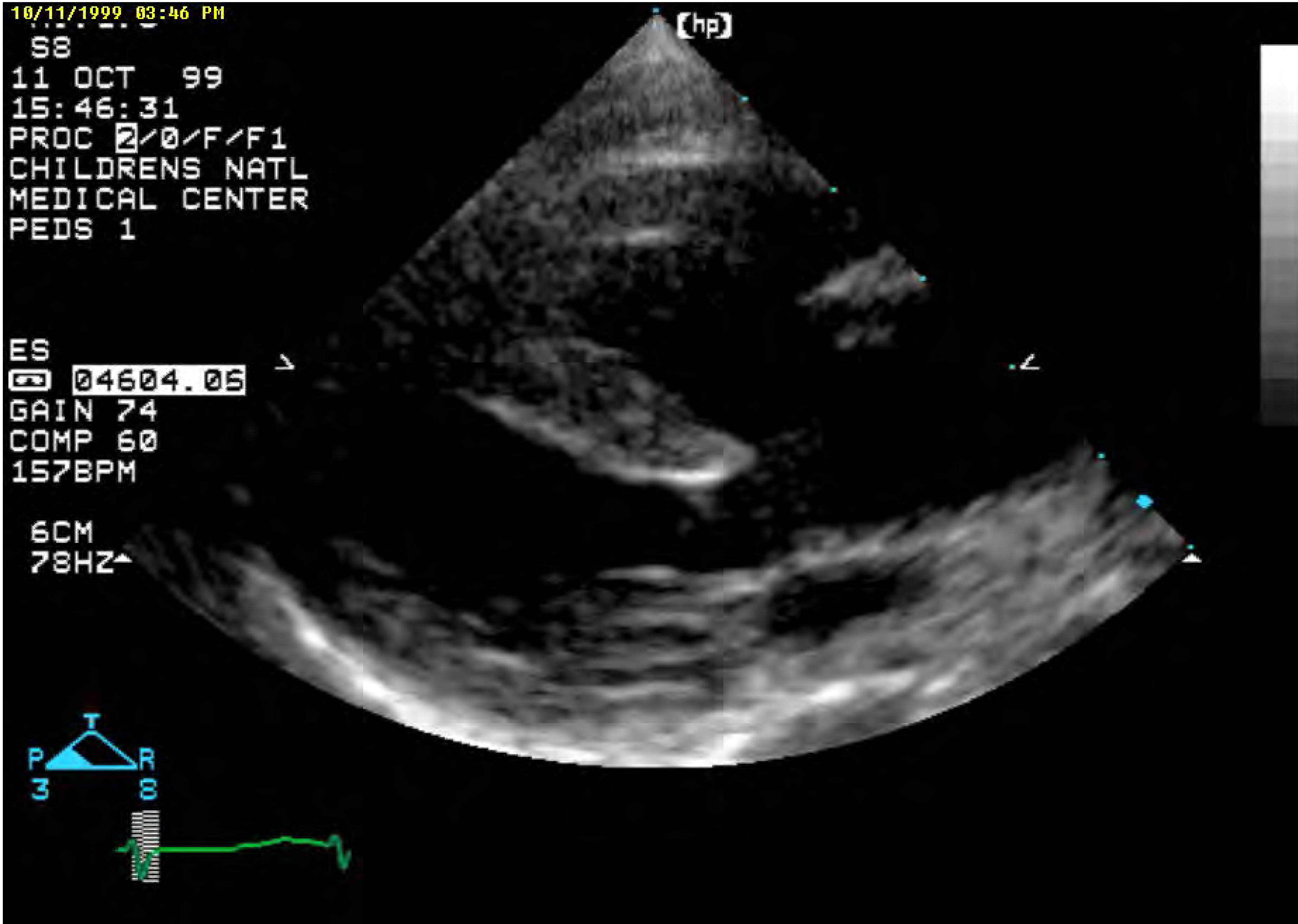
CXR

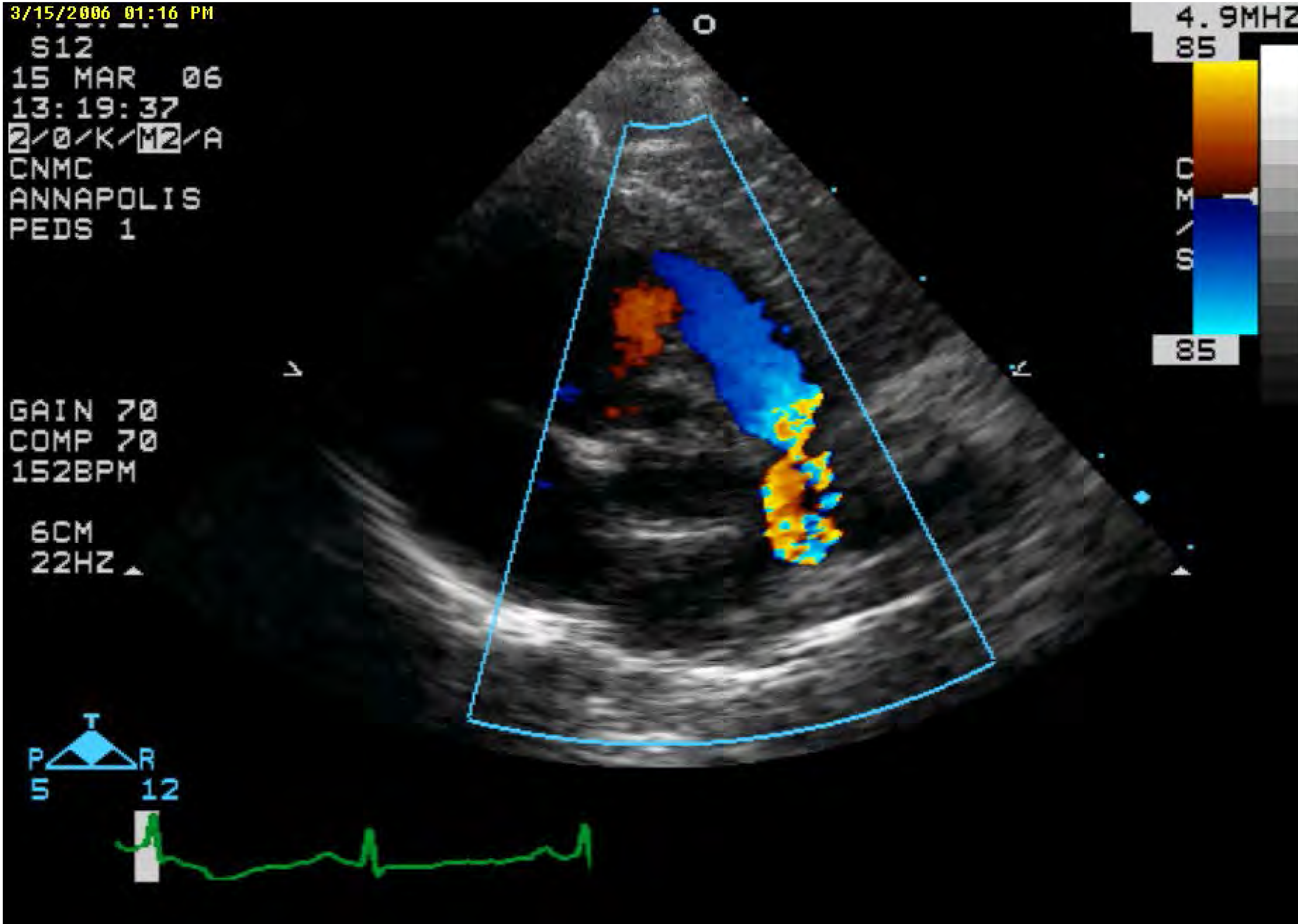
- Boot shaped heart

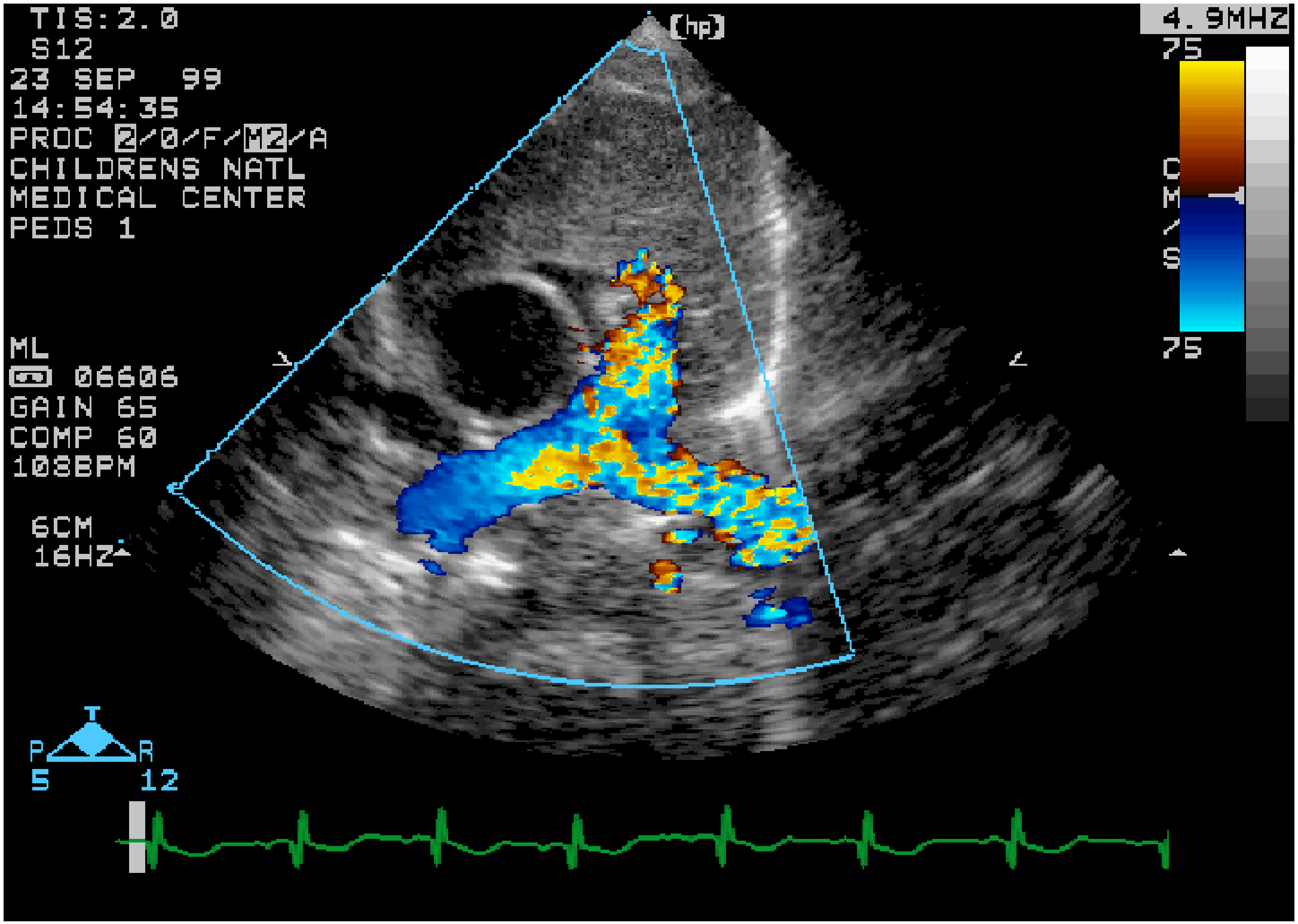
(.wav)

Tetralogy of Fallot



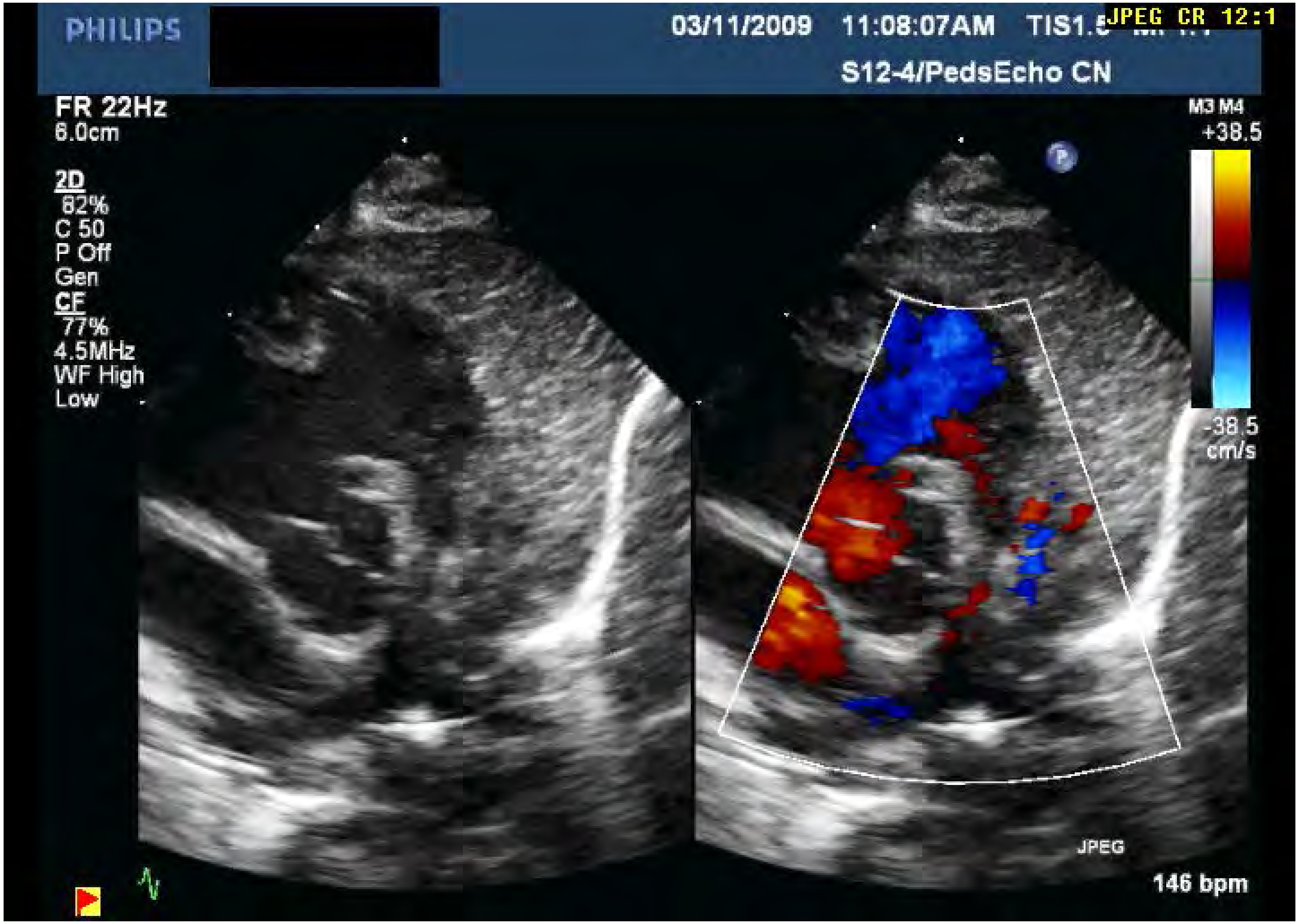

10/11/1999 03:46 PM
S8
11 OCT 99
15:46:31
PROC 2/0/F/F1
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
ES
04604.0S
GAIN 74
COMP 60
157BPM
6CM
78HZ
(hp)
T
P
R
3
8

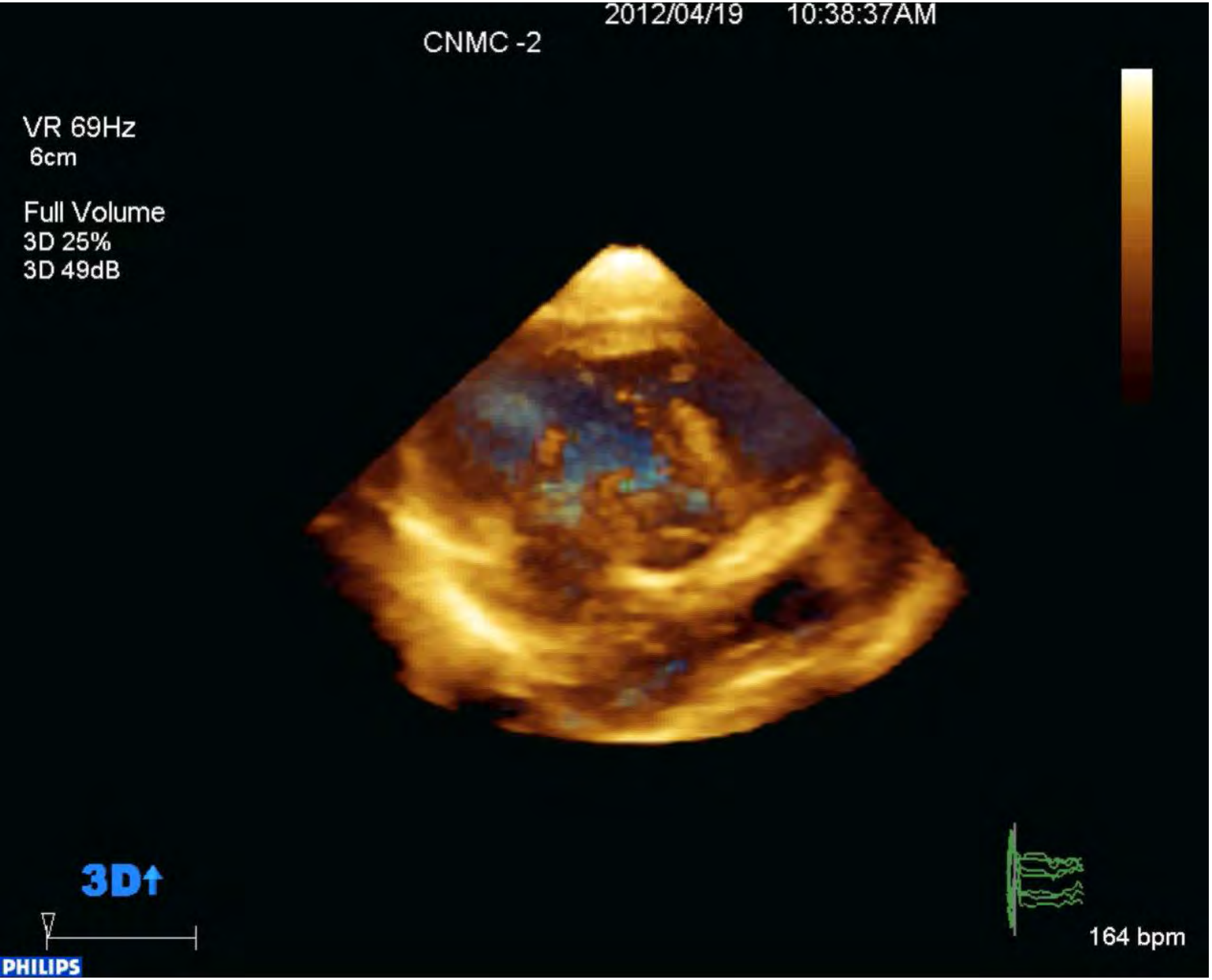

3/15/2006 01:16 PM
S12
15 MAR 06
13:19:37
2/0/K/M2/A
CNMC
ANNAPOLIS
PEDS 1
GAIN 70
COMP 70
152BPM
6CM
22HZ
4.9MHZ
85
CM/S
85
T
P
R
5
12



Children's National

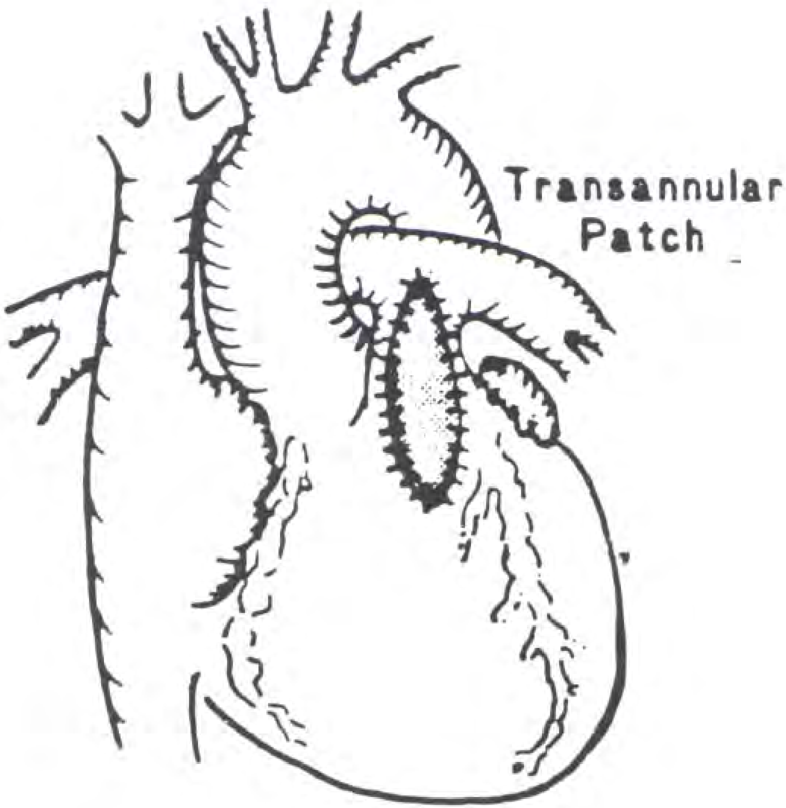

MI:1.6 S8
22 NOV 07 00:13:22
2/0/K/F3 11CM
GAIN 68 COMP 70 78HZcpd
155BPM
[redacted]
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
T
P
R
3
8

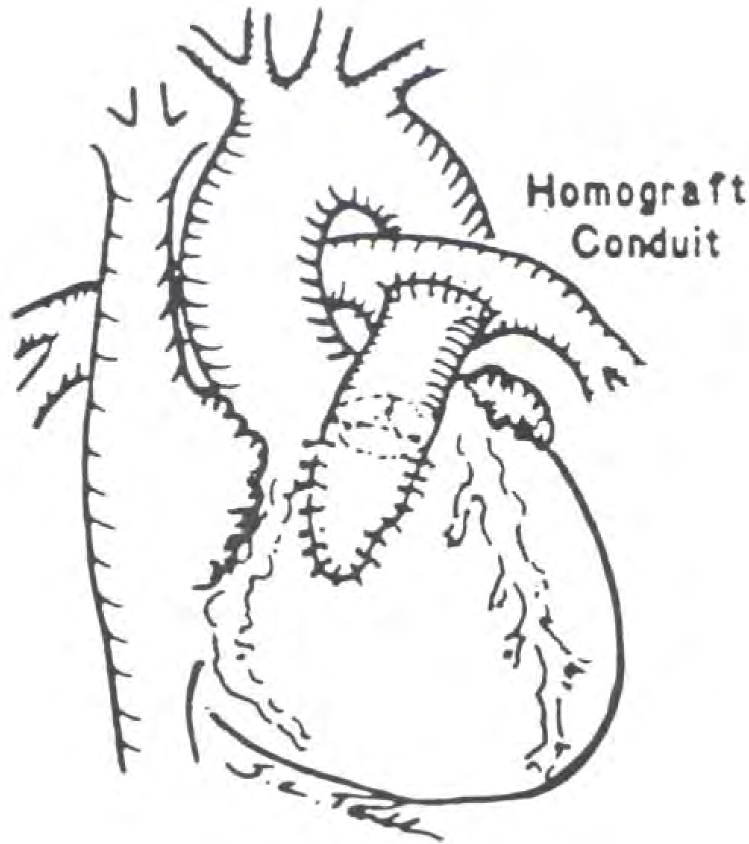

PHILIPS
[redacted]
03/11/2009
11:08:07AM
JPEG CR 12:1
S12-4/PedsEcho CN
FR 22Hz
6.0cm
2D
62%
C 50
P Off
Gen
CF
77%
4.5MHz
WF High
Low
M3 M4
+38.5
-38.5
cm/s
JPEG
146 bpm



Children's National™

Tetralogy of Fallot

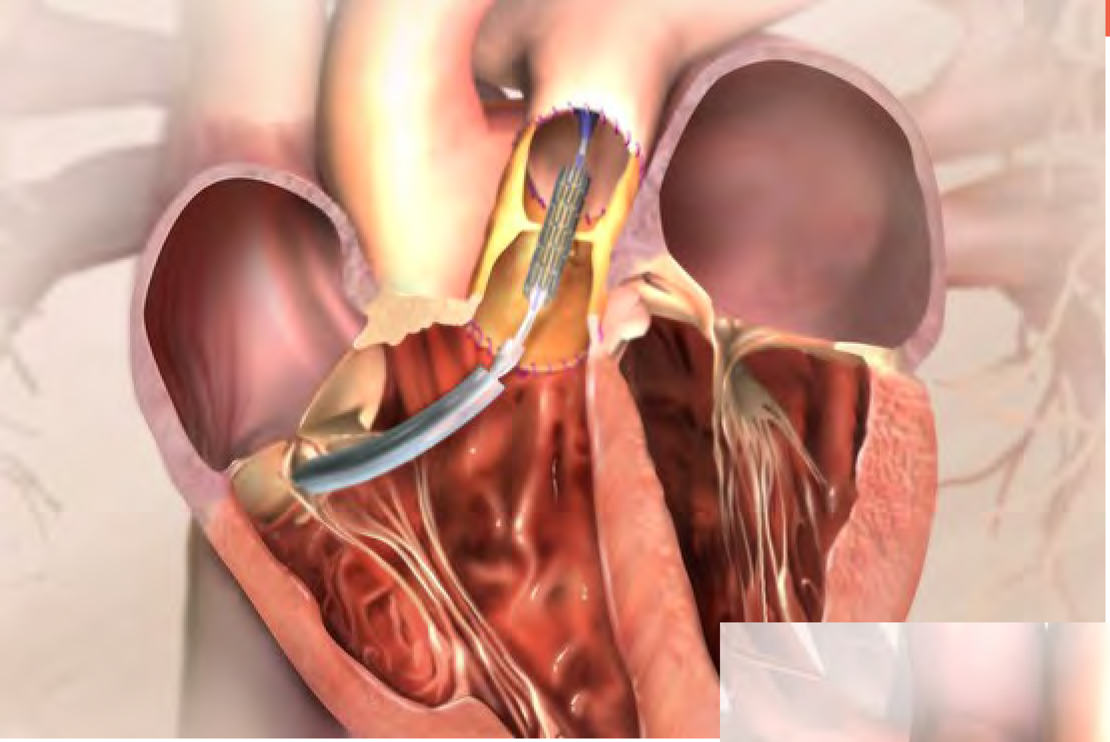
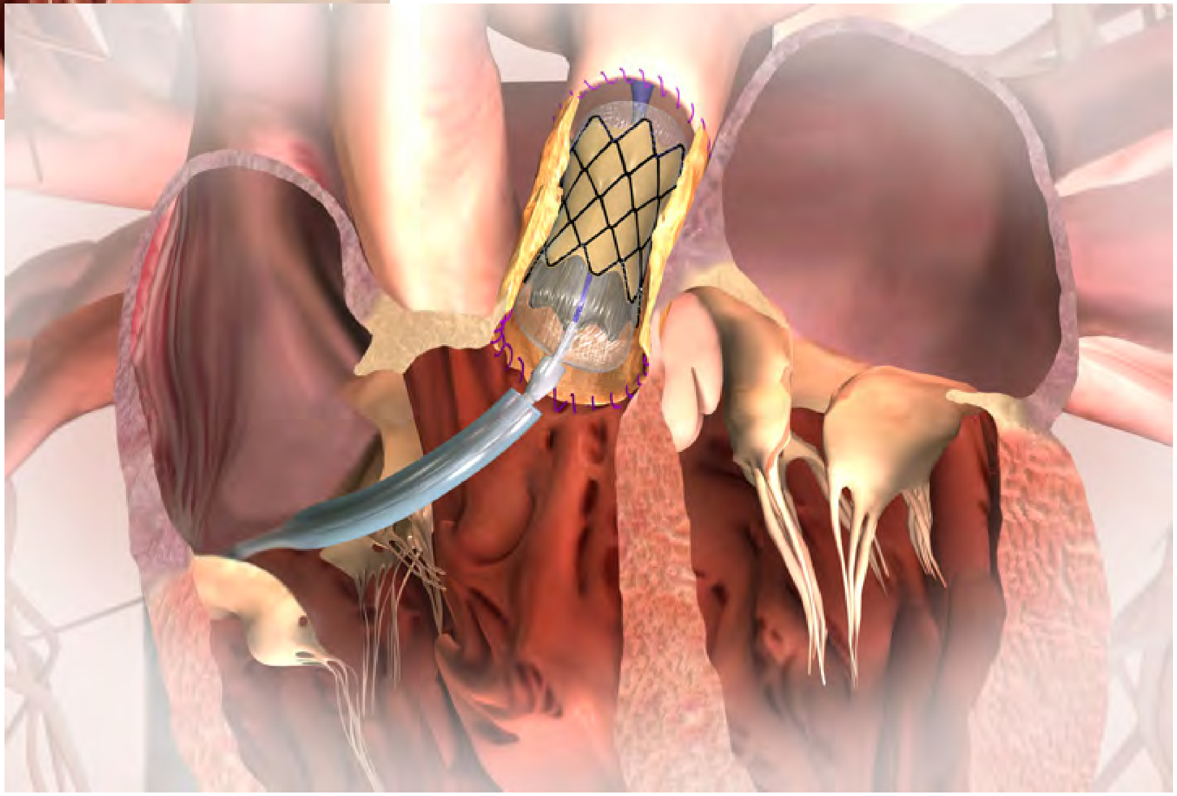




Transcatheter Pulmonary Valve

- Catheter delivered prosthetic pulmonary valve
- Made from bovine jugular vein
- Sewn within a platinum-iridium ballon expandable stent
- For use in patients with a surgically placed conduit from the RV to the PA
- Used to treat significant conduit valve insufficiency and/or stenosis that would otherwise require surgical conduit replacement
- FDA approved 2010

Children's National™

DORV

- Describes a relationship where the PA and Aorta both arise from the anatomic RV
- "DORV" is normal during heart development
- Incidence 1 – 1.5% of patients with CHD
- 1 per 10,000 live births
- Possible association with trisomy 13 and trisomy 18
- Van Praagh – both great arteries arise from the morphologically RV
- NO mitral - aortic fibrous continuity
- Two functional ventricles in which a VSD provides the only outlet for one ventricle
- Anderson - 50% override rule – "if >50% of the aorta is over the RV, its DORV"

Left Heart Obstruction

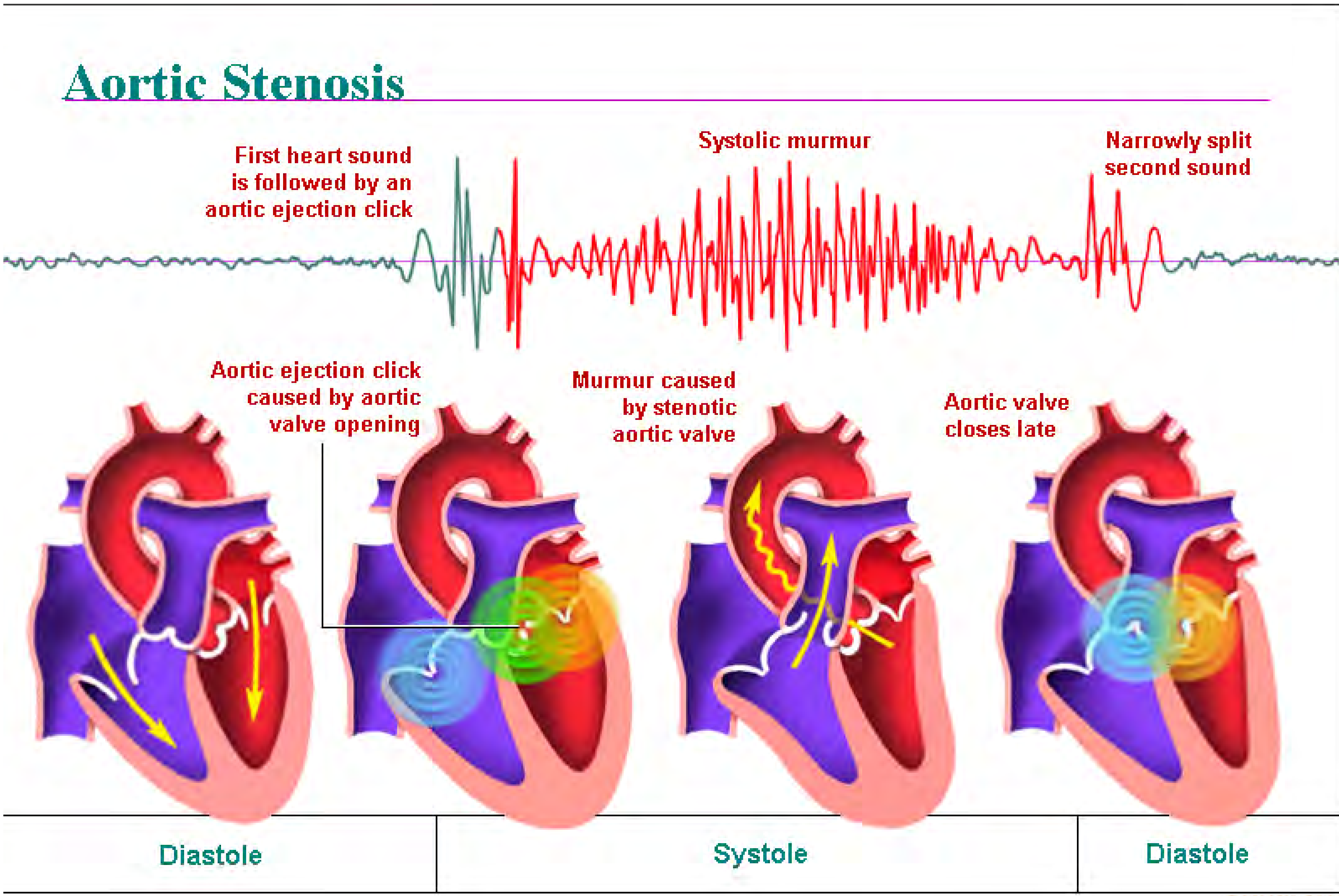
Aortic Stenosis

Valve, sub-valvar or supravalvar

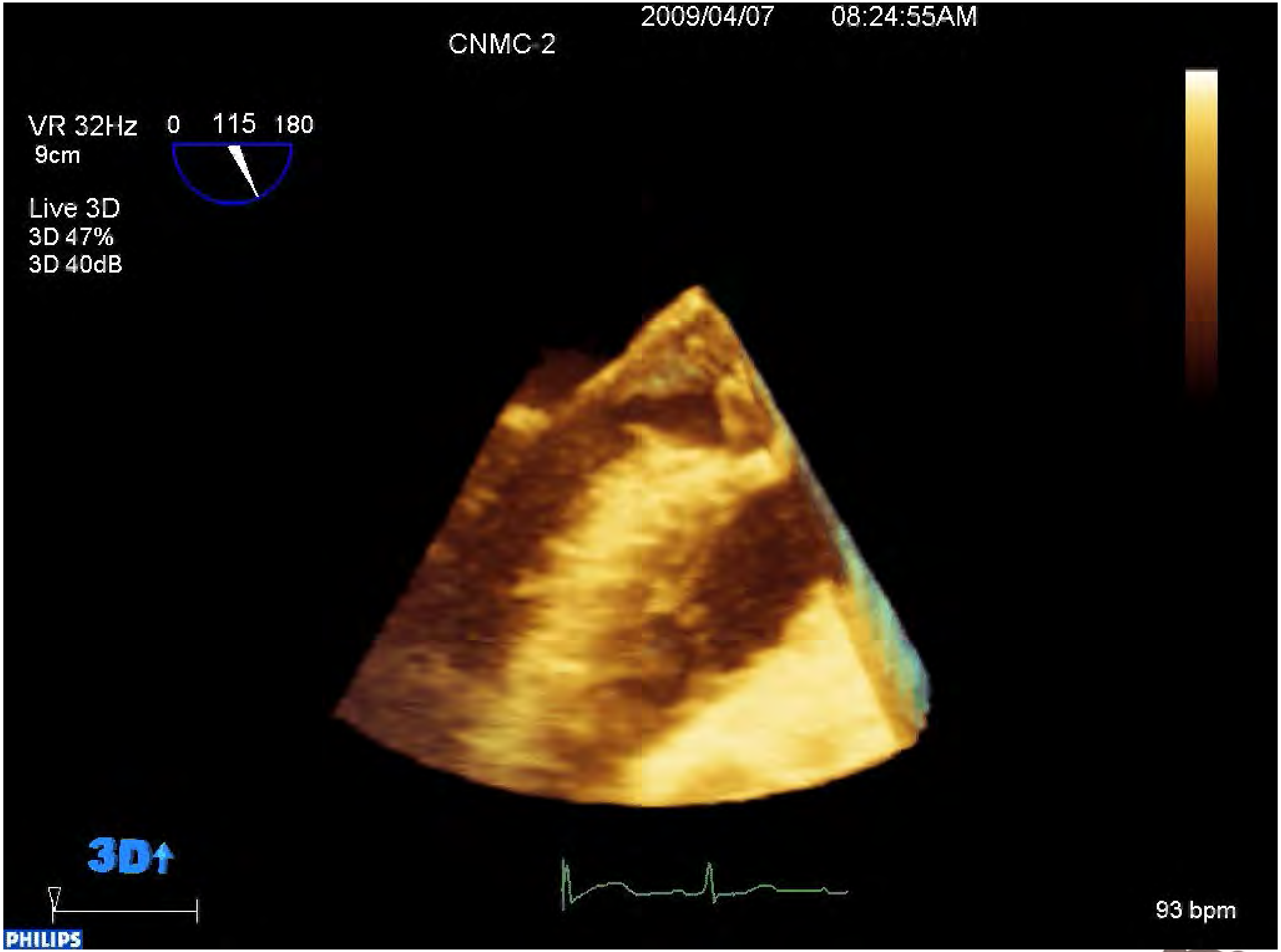
Clinical manifestations

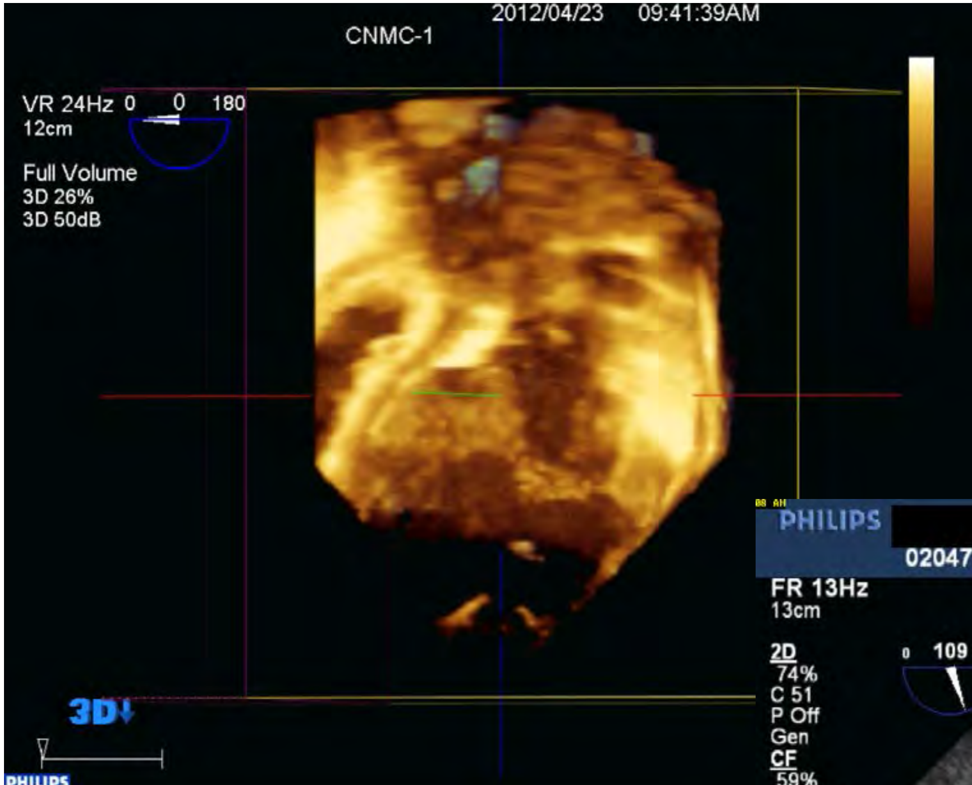
- Mild-moderate assymptomatic
- Severe
  - Depends on age of patient
- Management
  - Cath vs. surgery

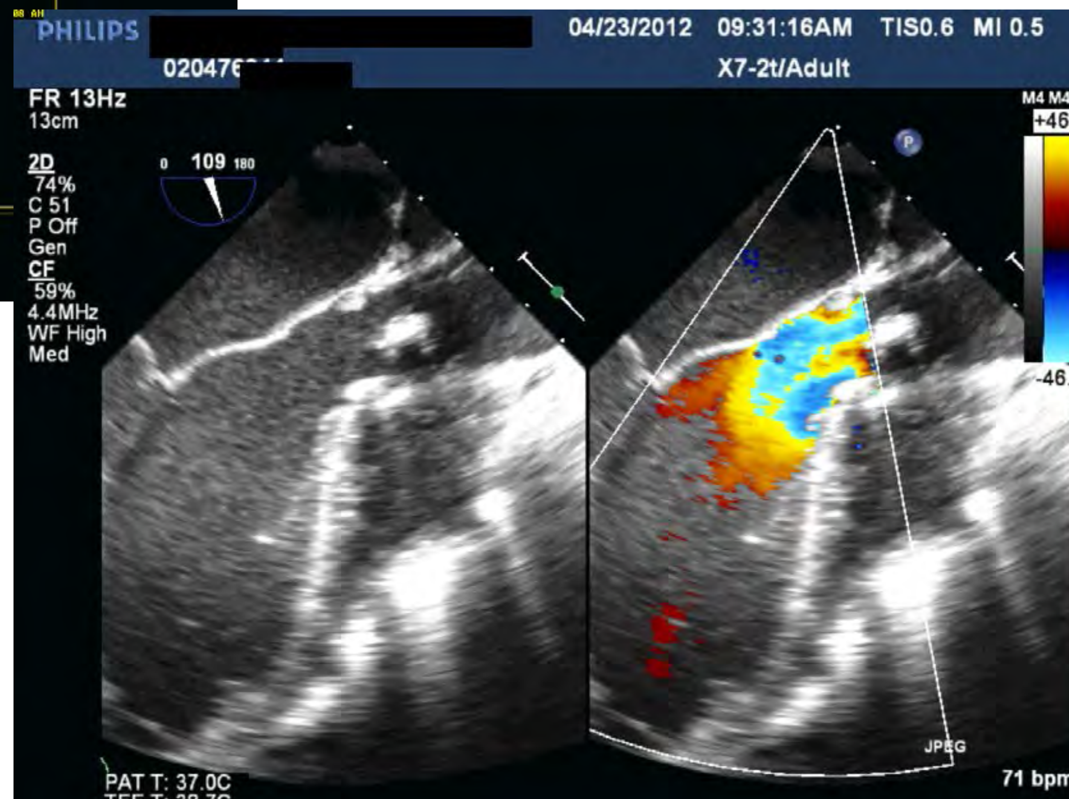
(.wav)

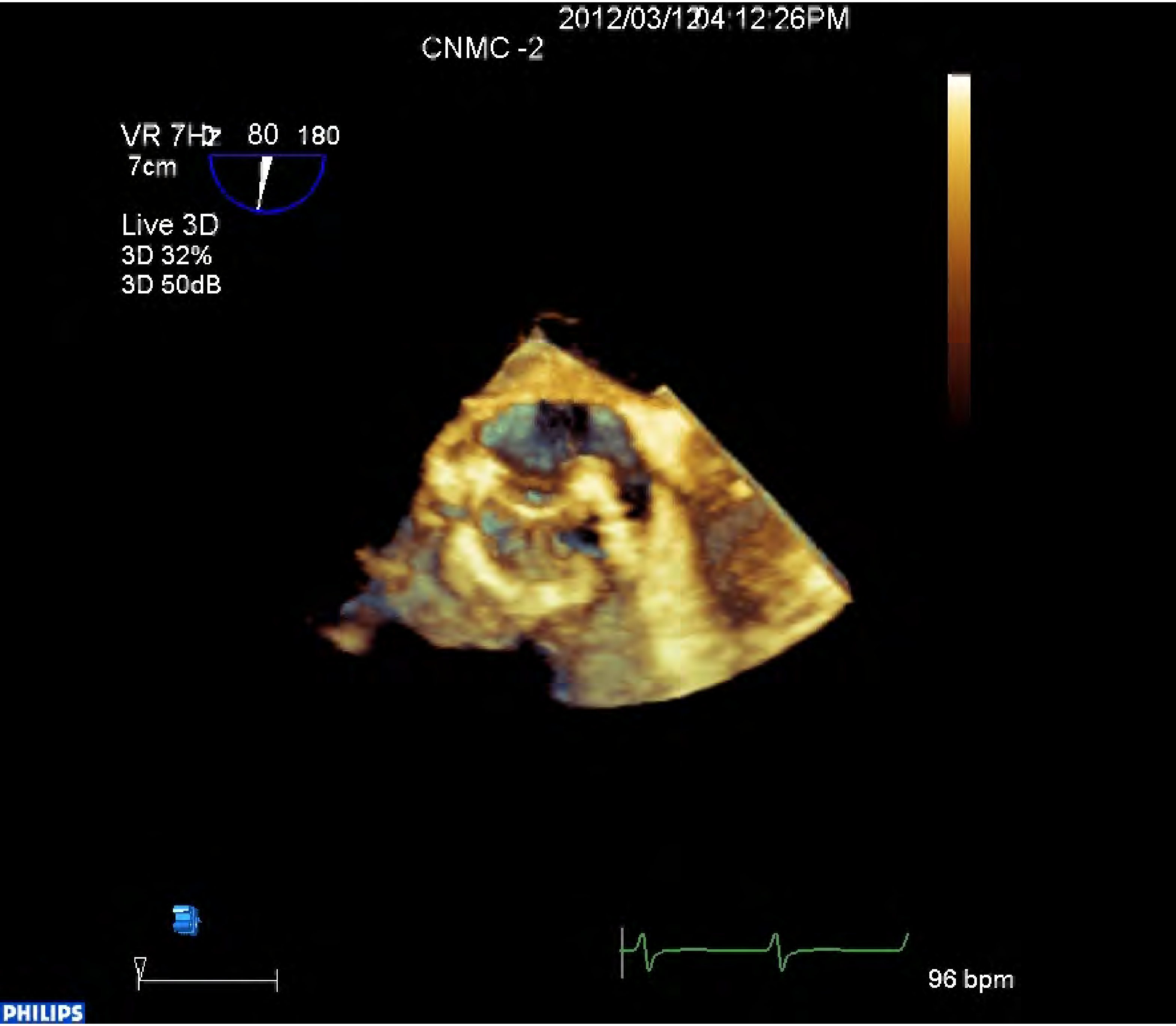


Aortic Stenosis
First heart sound is followed by an aortic ejection click
Systolic murmur
Narrowly split second sound
Aortic ejection click caused by aortic valve opening
Murmur caused by stenotic aortic valve
Aortic valve closes late
Diastole
Systole
Diastole


2009/04/07 08:24:55AM
CNMC 2
VR 32Hz
9cm
0 115 180
Live 3D
3D 47%
3D 40dB
3D
PHILIPS
93 bpm


2012/04/23 09:41:39AM
CNMC-1
VR 24Hz 0 0 180
12cm
Full Volume
3D 26%
3D 50dB
3D
PHILIPS


PHILIPS [redacted]
04/23/2012 09:31:16AM TIS0.6 MI 0.5
02047[redacted]
X7-2t/Adult
FR 13Hz
13cm
2D
74%
C 51
P Off
Gen
CF
59%
4.4MHz
WF High
Med
0 109 180
M4 M4
+46.1
-46.1
JPEG
PAT T: 37.0C
TEE T: 38.7C
71 bpm



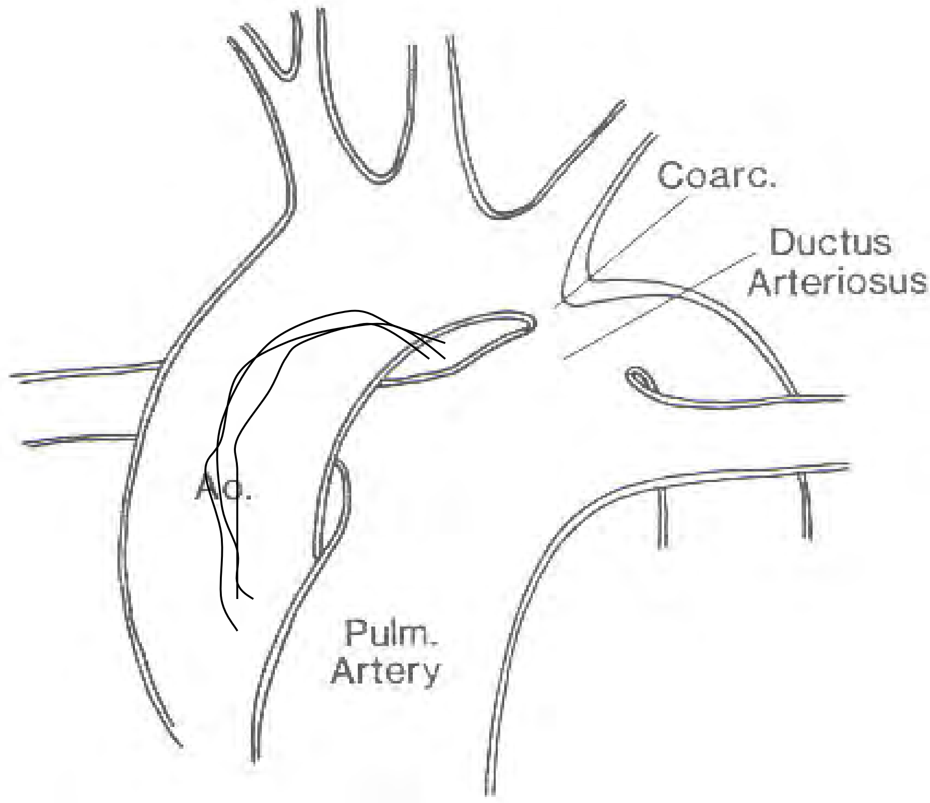
ildren's National

Coarctation of the Aorta

Aberrant ductal tissue within the wall of the aorta

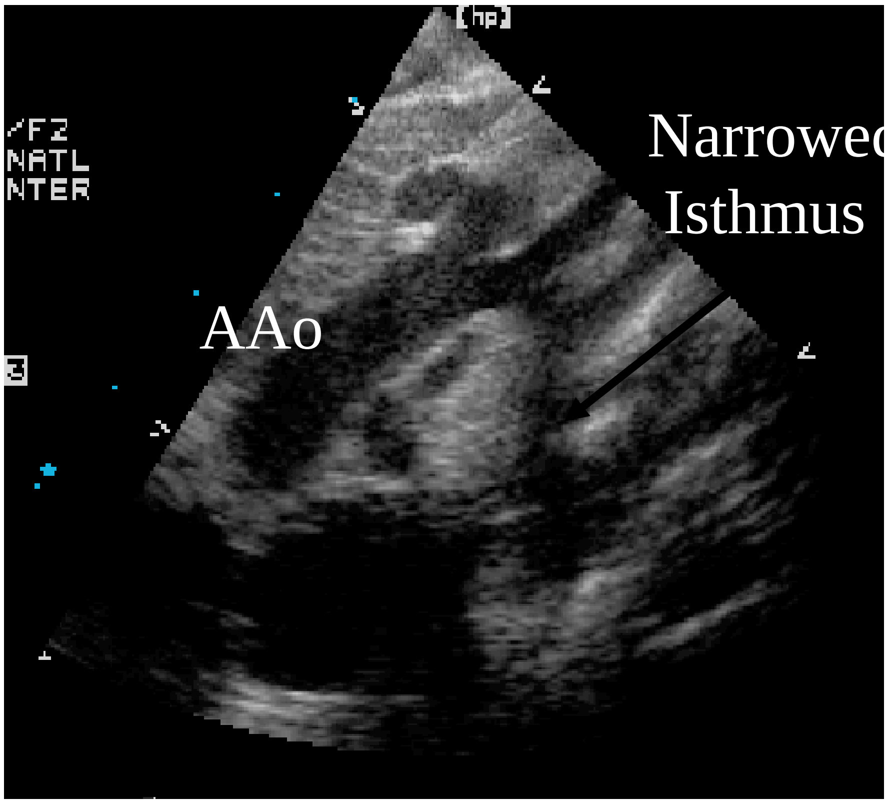
All coarcts are "juxtaductal"

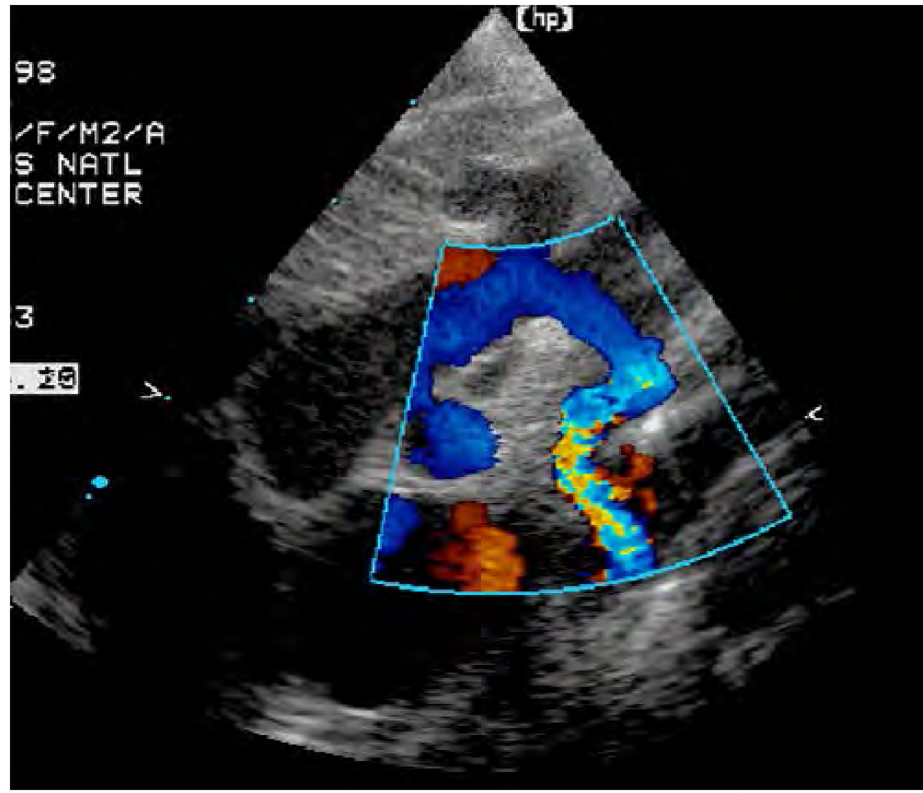
Pseudocoarctation = kinking at t
WITHOUT obstruction



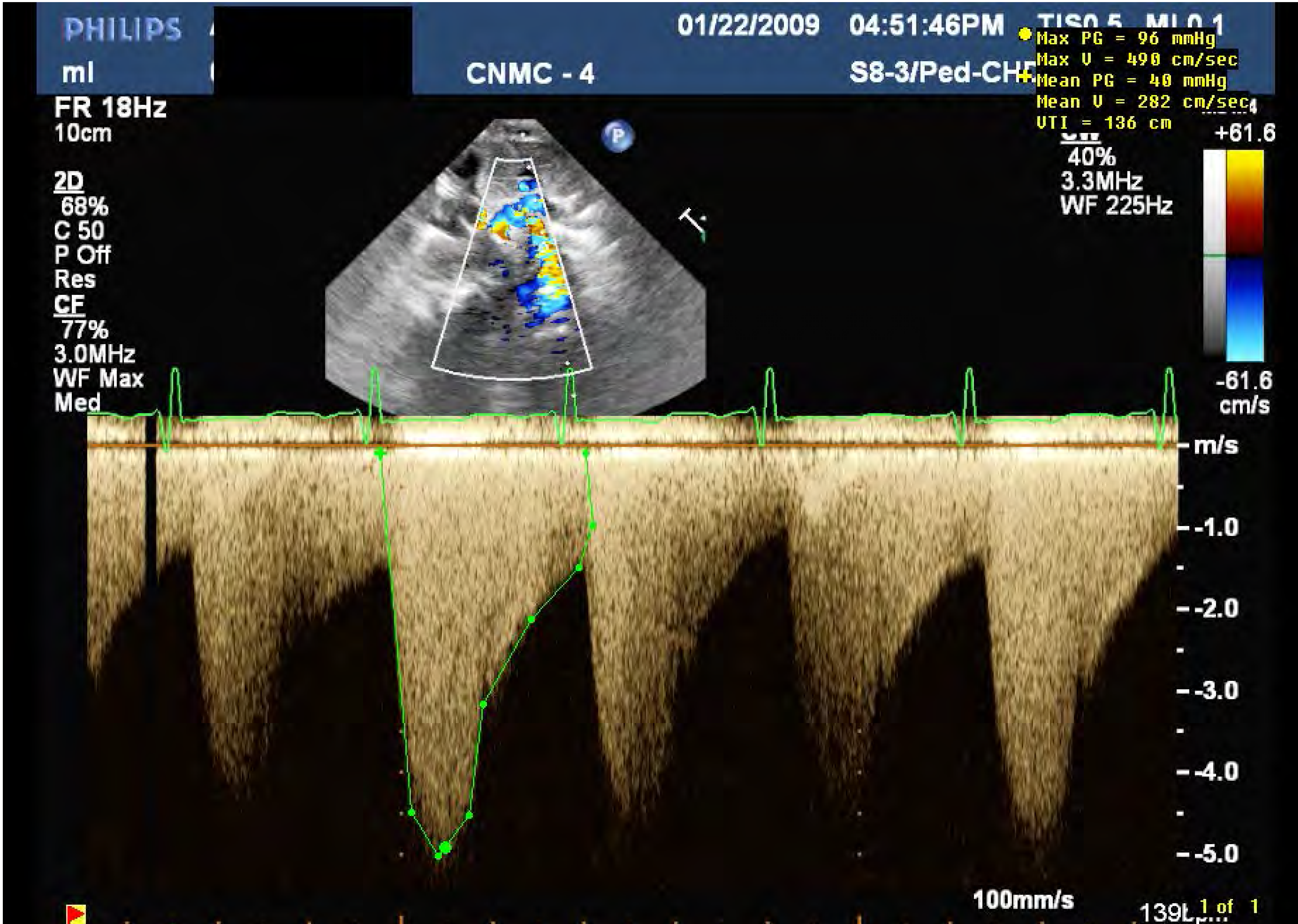
Children's National

Coarctation of the Aorta

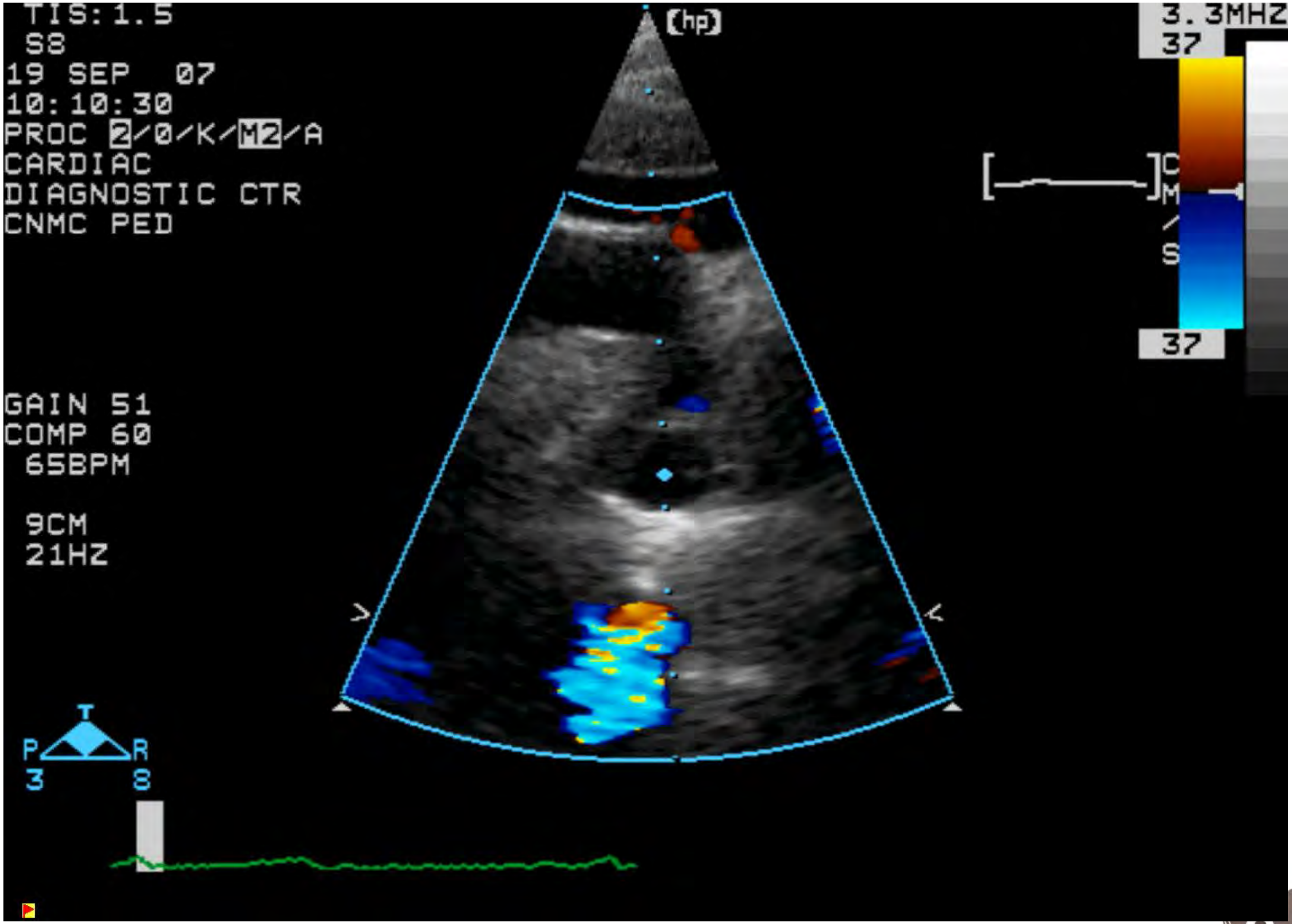




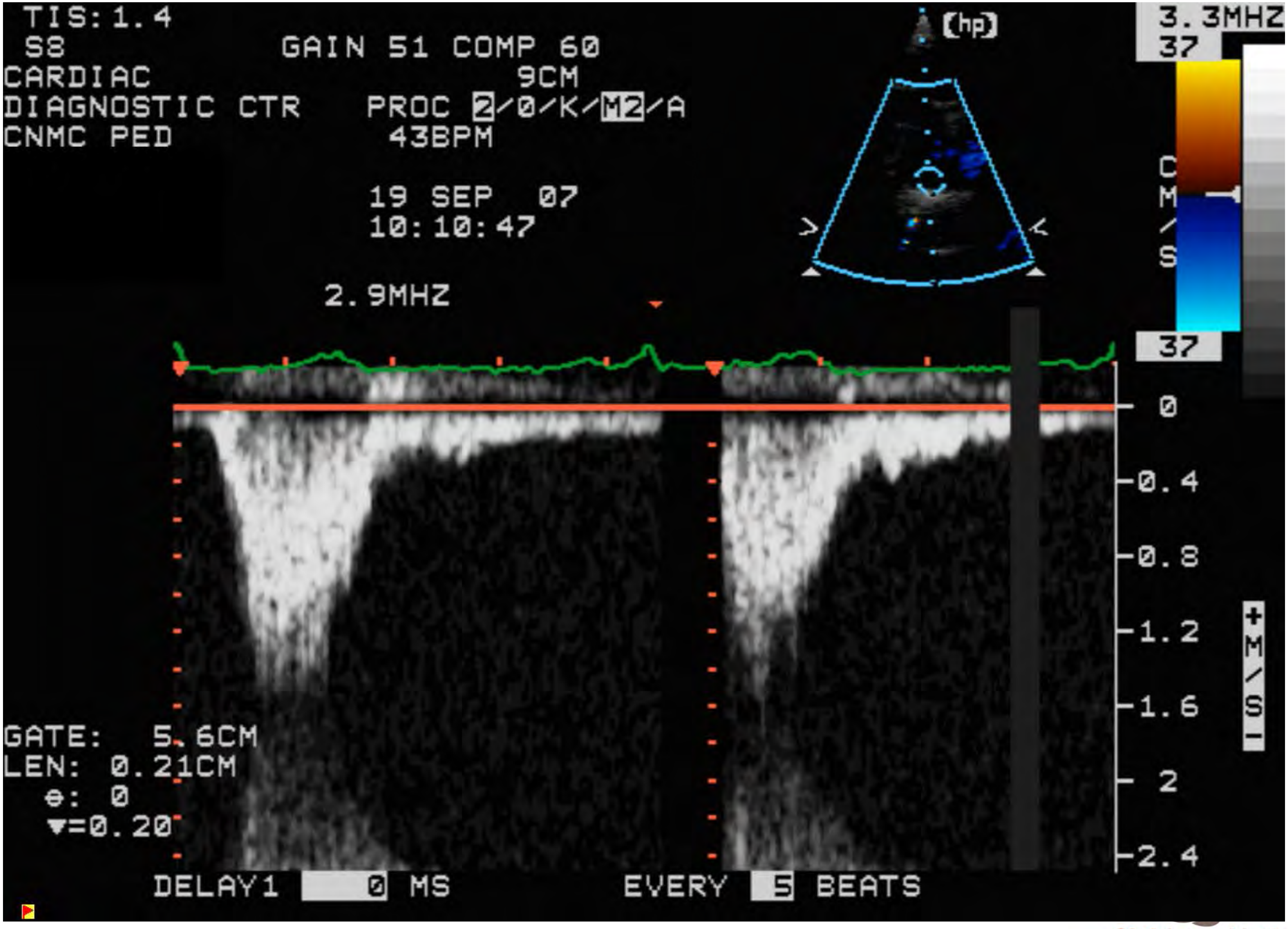
PHILIPS
01/22/2009 11:34:00AM TIS0.4
JPEG CR 18:1
S12-4/Pediatric
FR 17Hz
8.1cm
2D
88%
C 50
P Off
Gen
CF
77%
4.5MHz
WF High
Low
M3 M4
+67.0
-67.0
cm/s
JPEG
157 bpm


PHILIPS
[redacted]
ml
01/22/2009
04:51:46PM
CNMC - 4
S8-3/Ped-CHD
Max PG = 96 mmHg
Max V = 490 cm/sec
Mean PG = 40 mmHg
Mean V = 282 cm/sec
VTI = 136 cm
FR 18Hz
10cm
2D
68%
C 50
P Off
Res
CF
77%
3.0MHz
WF Max
Med
40%
3.3MHz
WF 225Hz
+61.6
-61.6
cm/s
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
100mm/s
1 of 1

Normal or CoA?

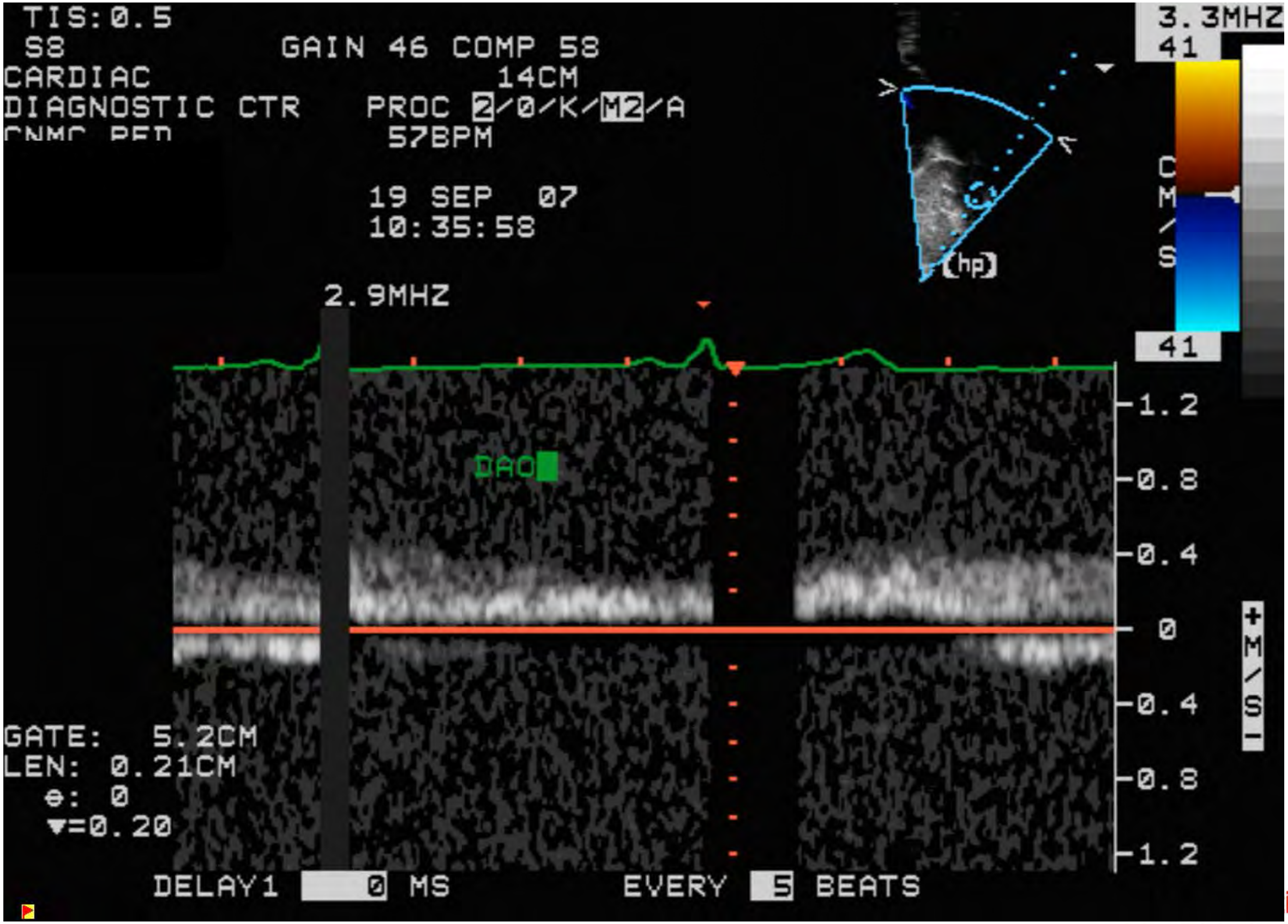


Children's National™



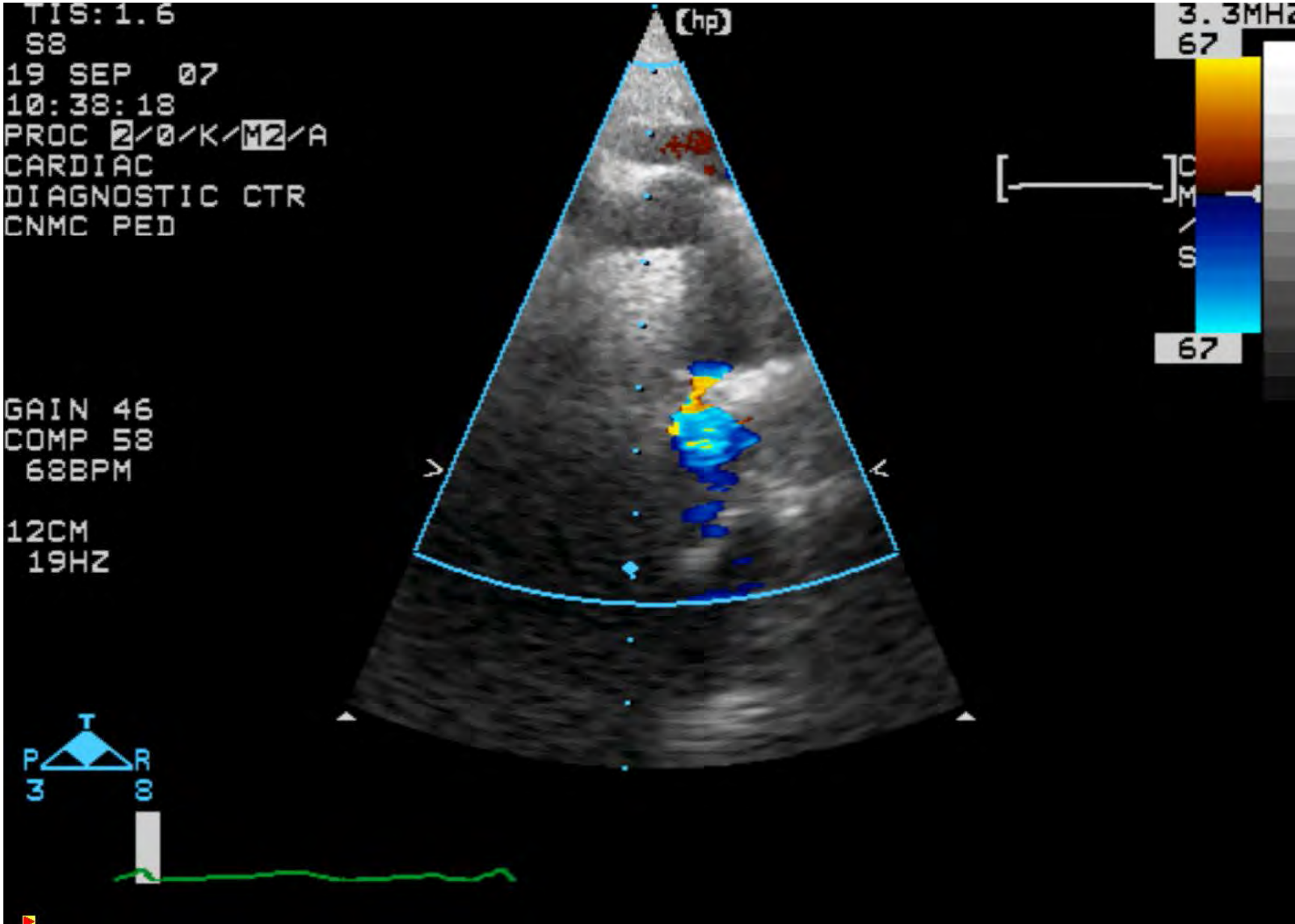
Children's National ™

Descending AO Doppler

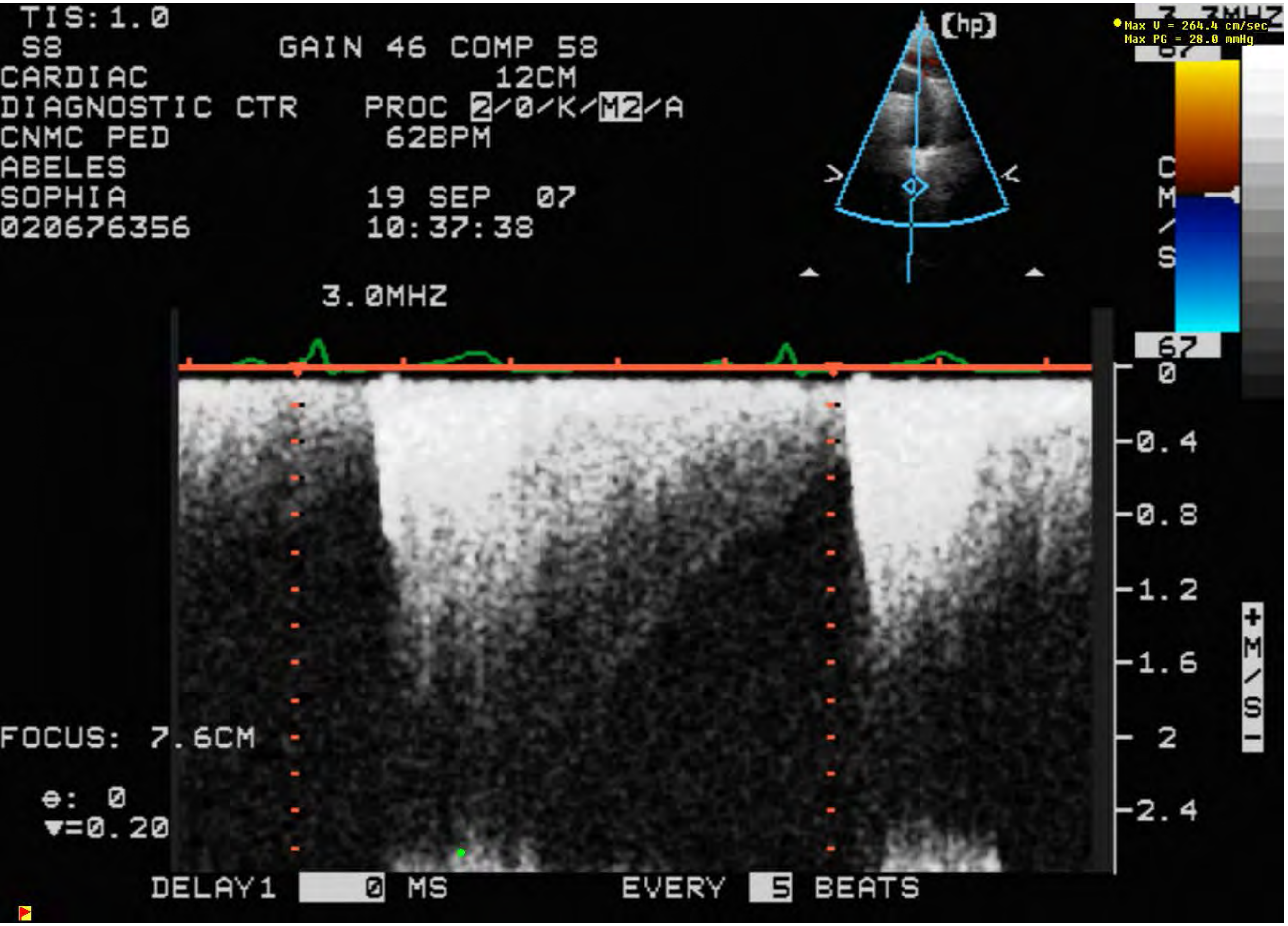


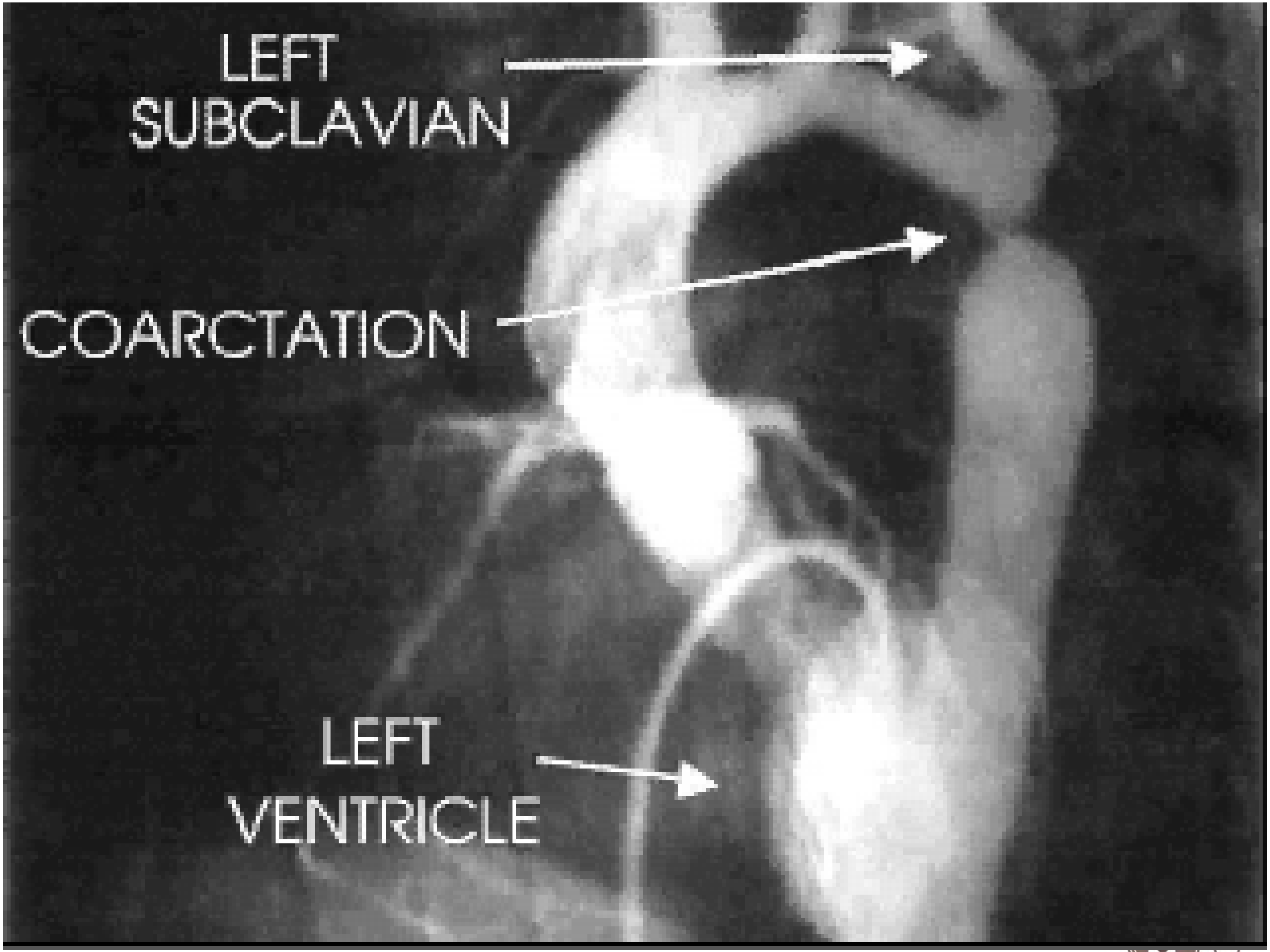
ional ™

Coarctation



Doppler "drag"



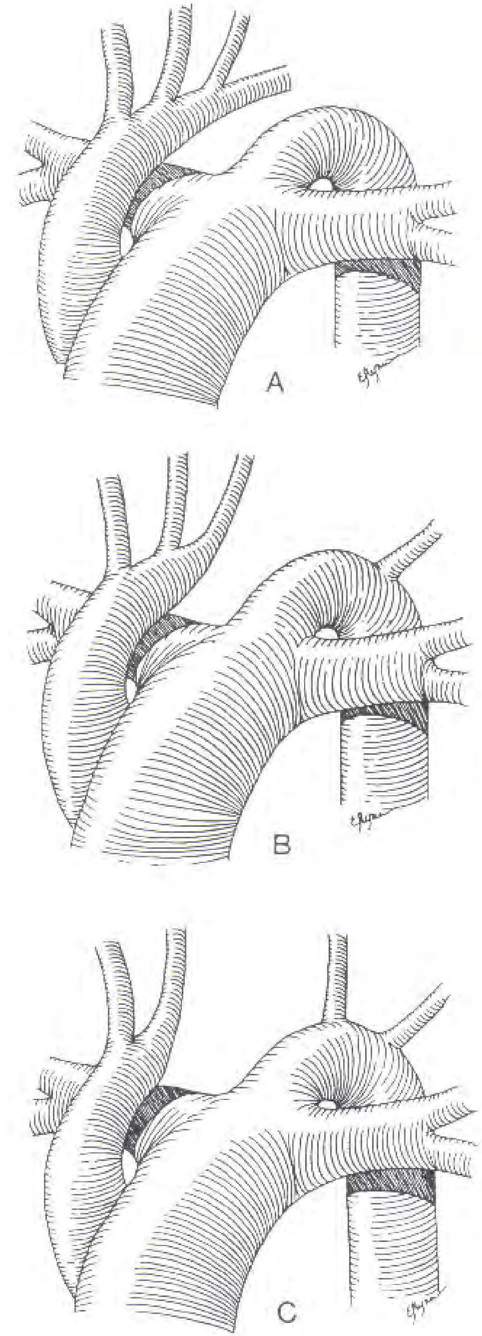

LEFT
SUBCLAVIAN
COARCTATION
LEFT
VENTRICLE

Interrupted Aortic Arch

Type A = After the subclavian artery, prob extreme form of coarctation with obliterat lumen

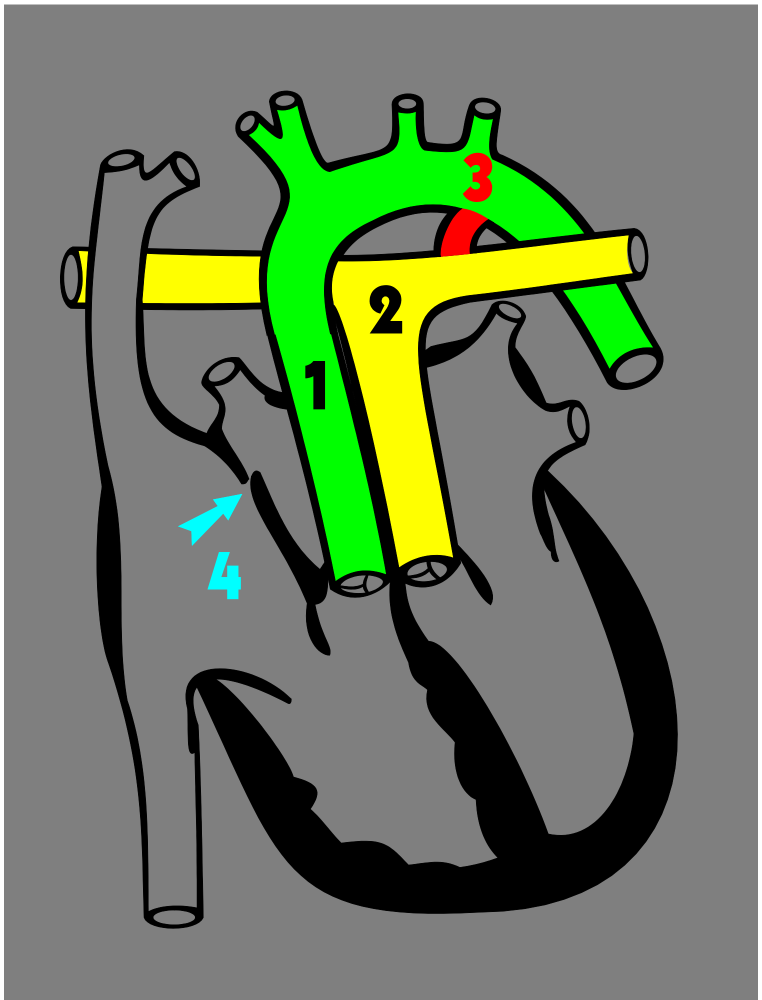
Type B = Between the LCC and LSCA, mos defect of arch remodeling/neural crest

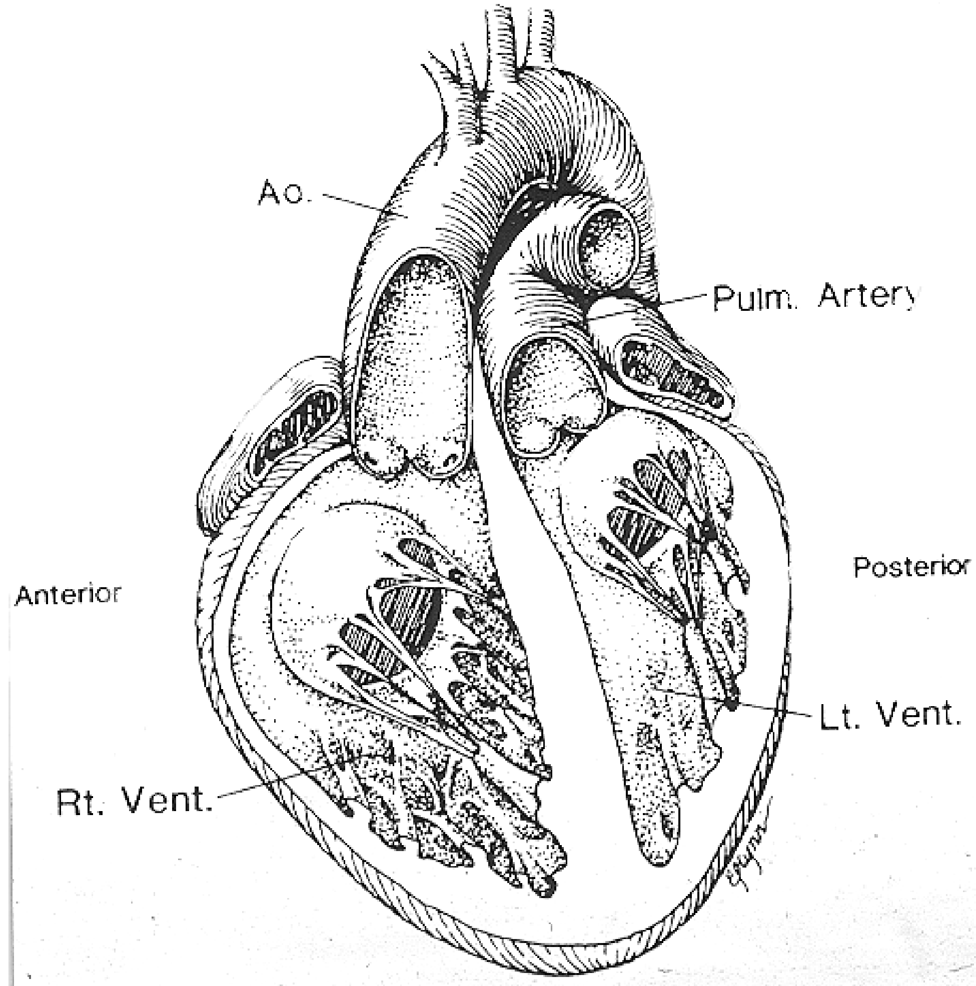
Type C = Between the Carotid arteries, mc



Complex Lesions

D-Transposition of the Great Arteries





D-TGA

First described by Baillie 1797
Natural history: >90% mortality in infancy
Incidence: ~5% of congenital heart disease
Rare association with syndromes or other anomalies
Male:Female = 2:1
Possible association with infant of diabetic mother

D-TGA

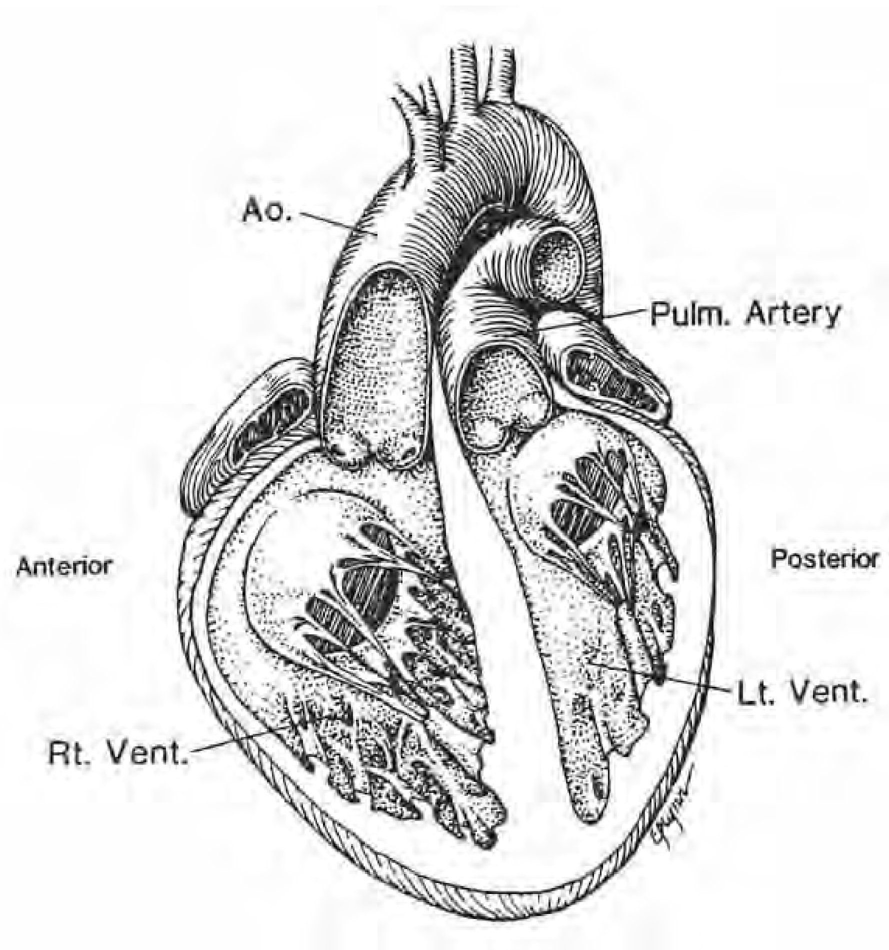
Ventriculo-arterial discordan

Circulation in parallel

RA=>RV=>Ao

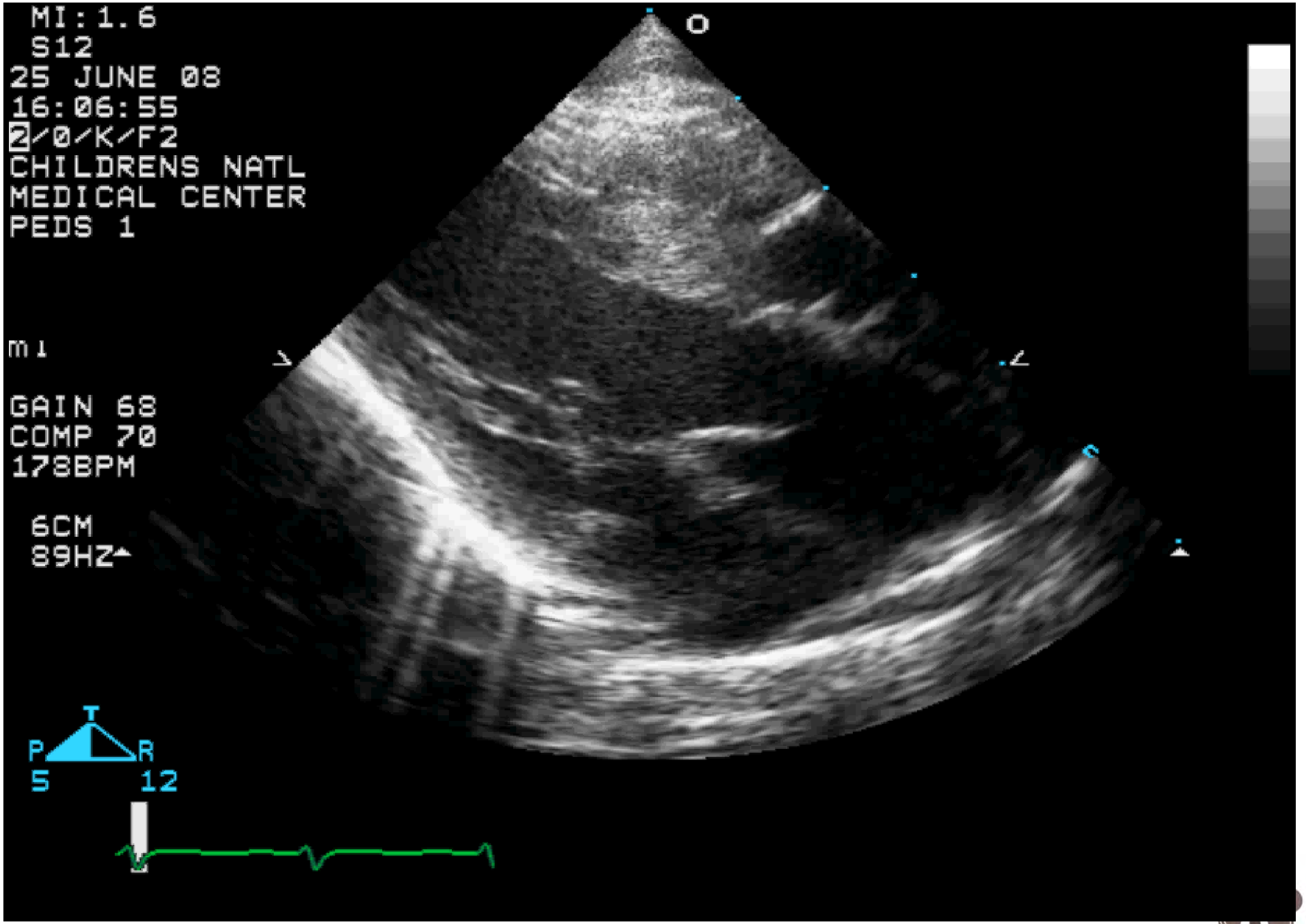
LA=>LV=>PA

Must have mixing at atrial or survive



Children's National™

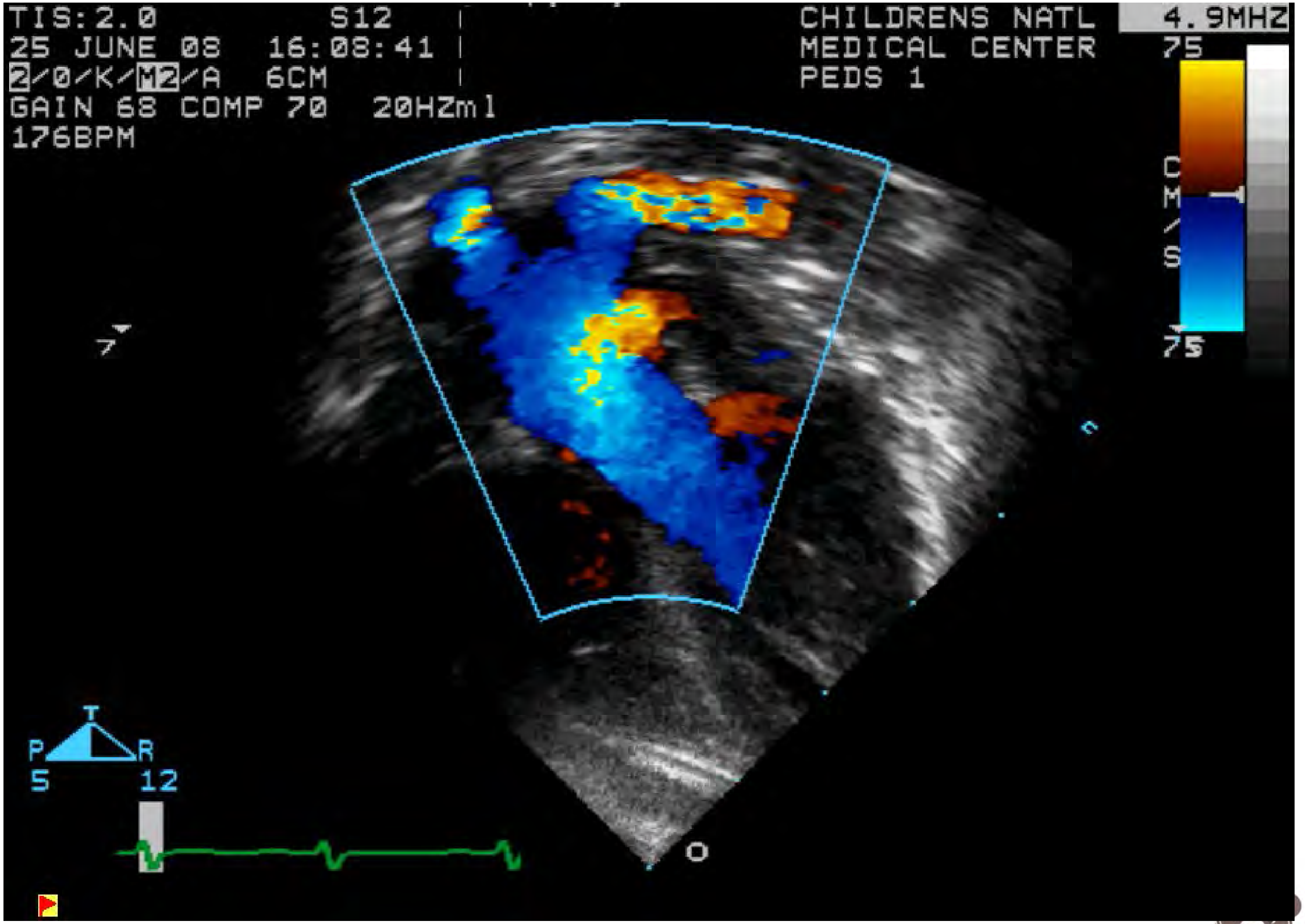
D-TGA



Children's National

11/8/2008 09:55
JPEG CR 18:1
PHILIPS
11/08/2008 10:11:53AM TIS1.5
S12-4/PedsEcho CN
FR 95Hz
5.0cm
2D
60%
C 50
P Off
Gen
M3
G
P
R
JPEG
176 bpm

D-Transposition

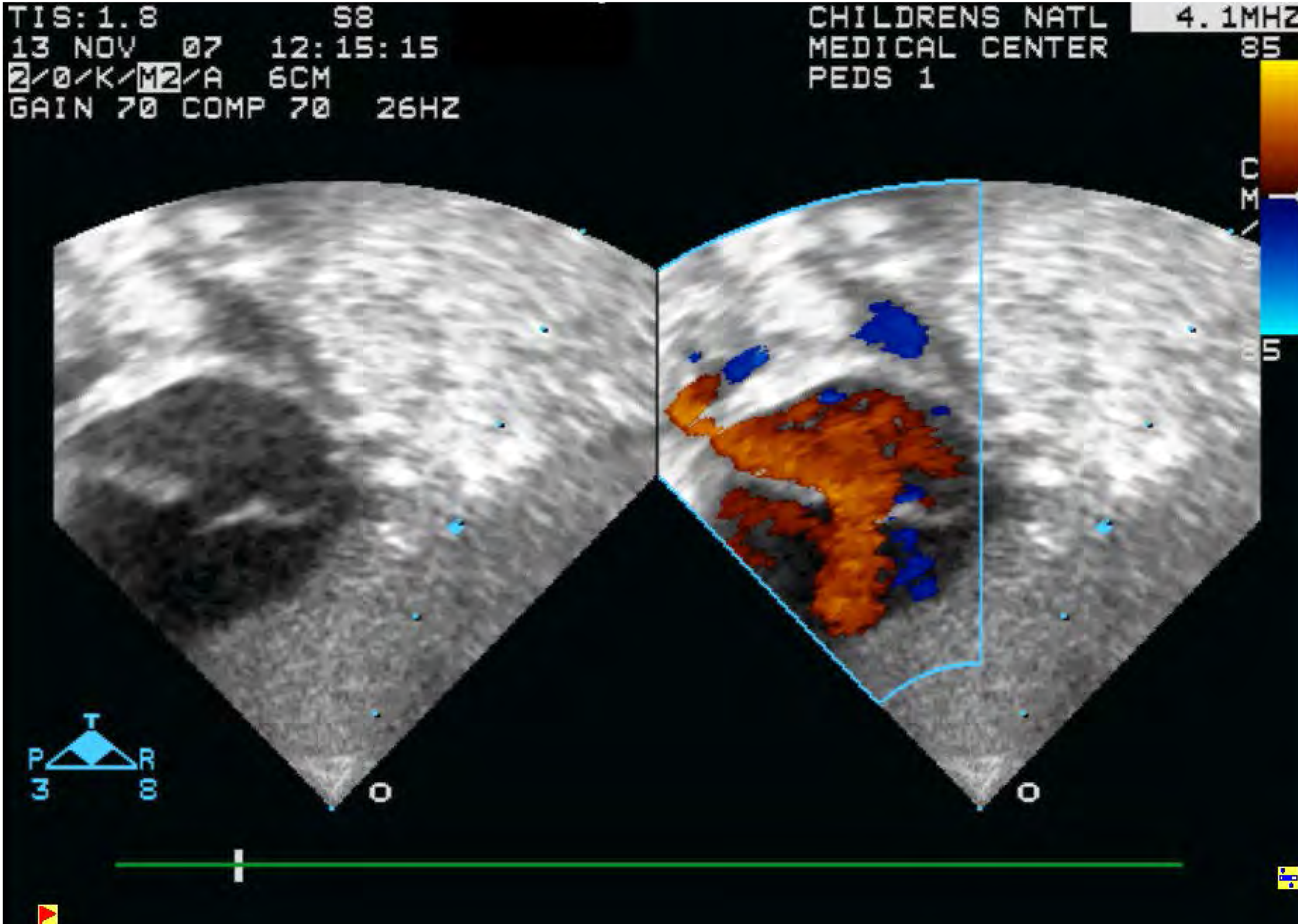


Children's National

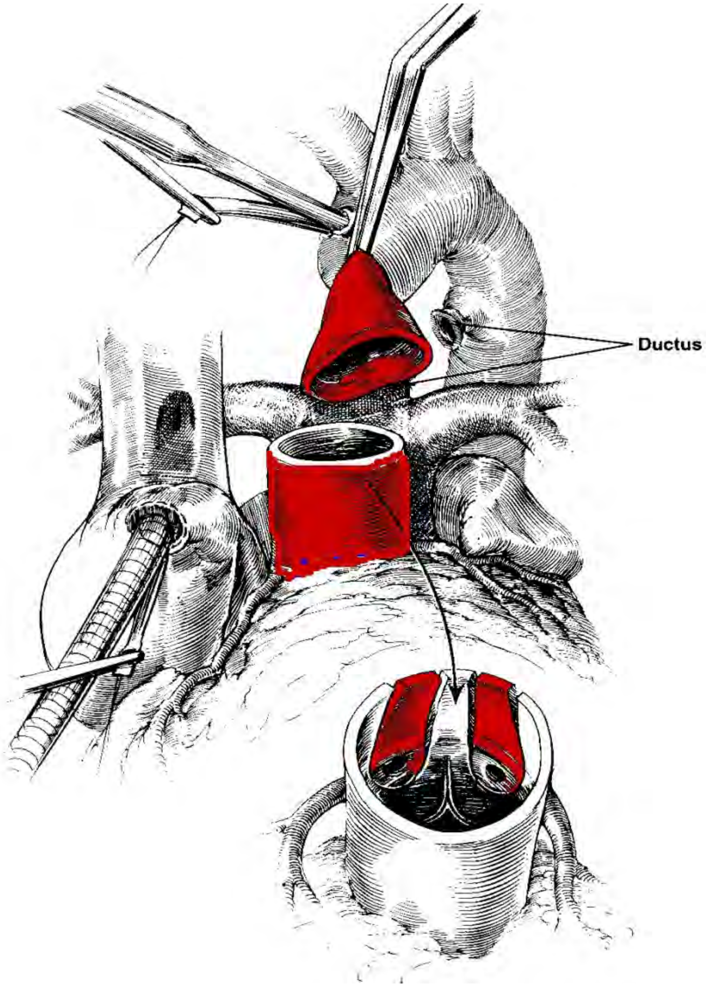
D-Transposition
Balloon Septostomy

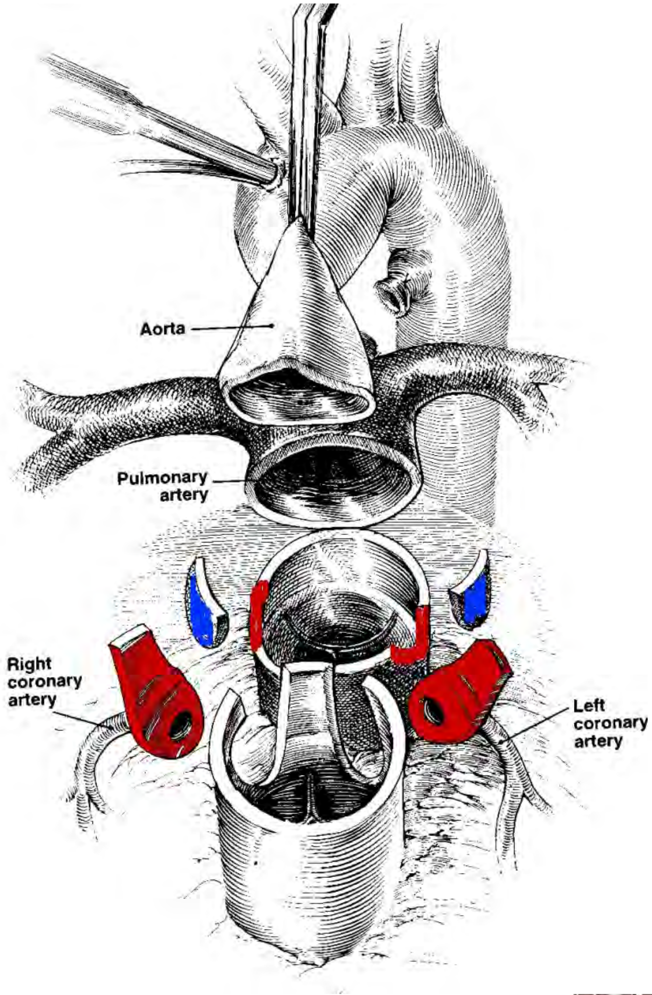
Again...

Children's National™


TIS:1.8 S8
13 NOV 07 12:15:15
2/0/K/M2/A 6CM
GAIN 70 COMP 70 26HZ
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
4.1MHZ
85
P
R
3
8
T
0
0

Arterial Switch Procedure





Children's National™

Long Term Postoperative Concerns
Arterial Switch Operation

Neo-pulmonary stenosis

Coronary abnormalities

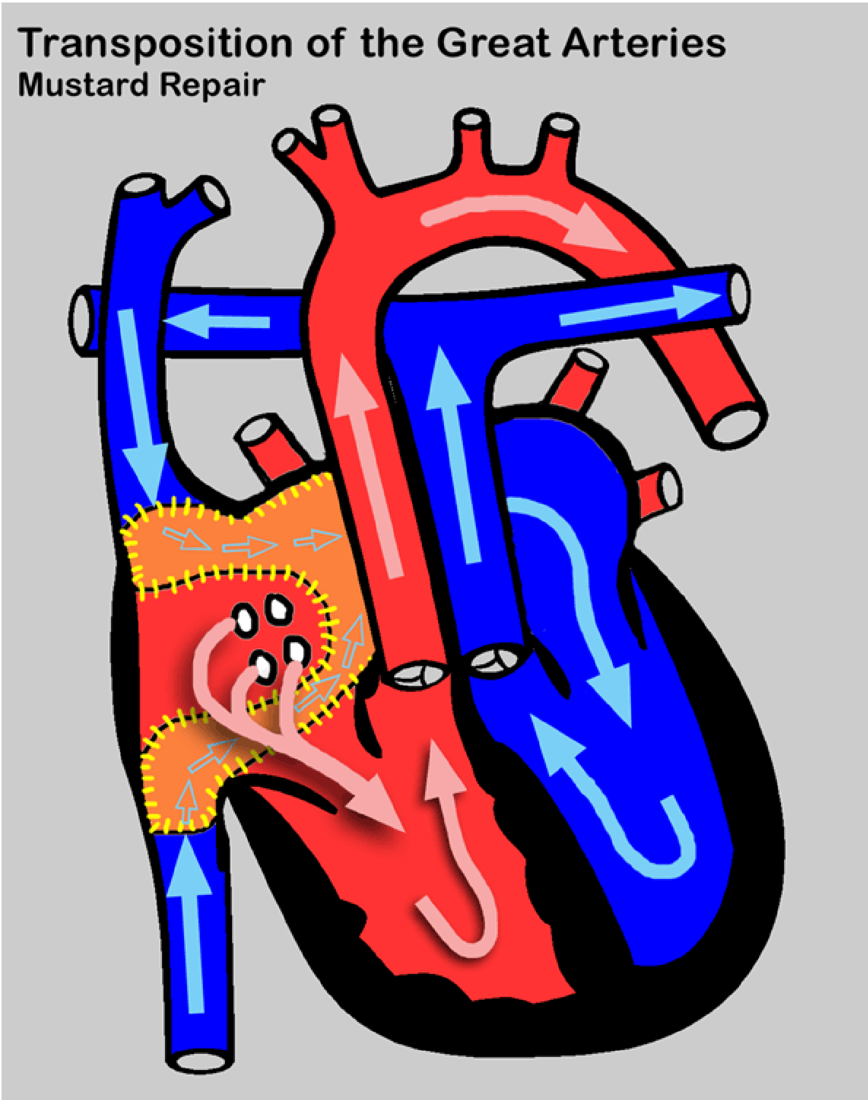
- Obstruction and stenosis
- Decreased flow reserve

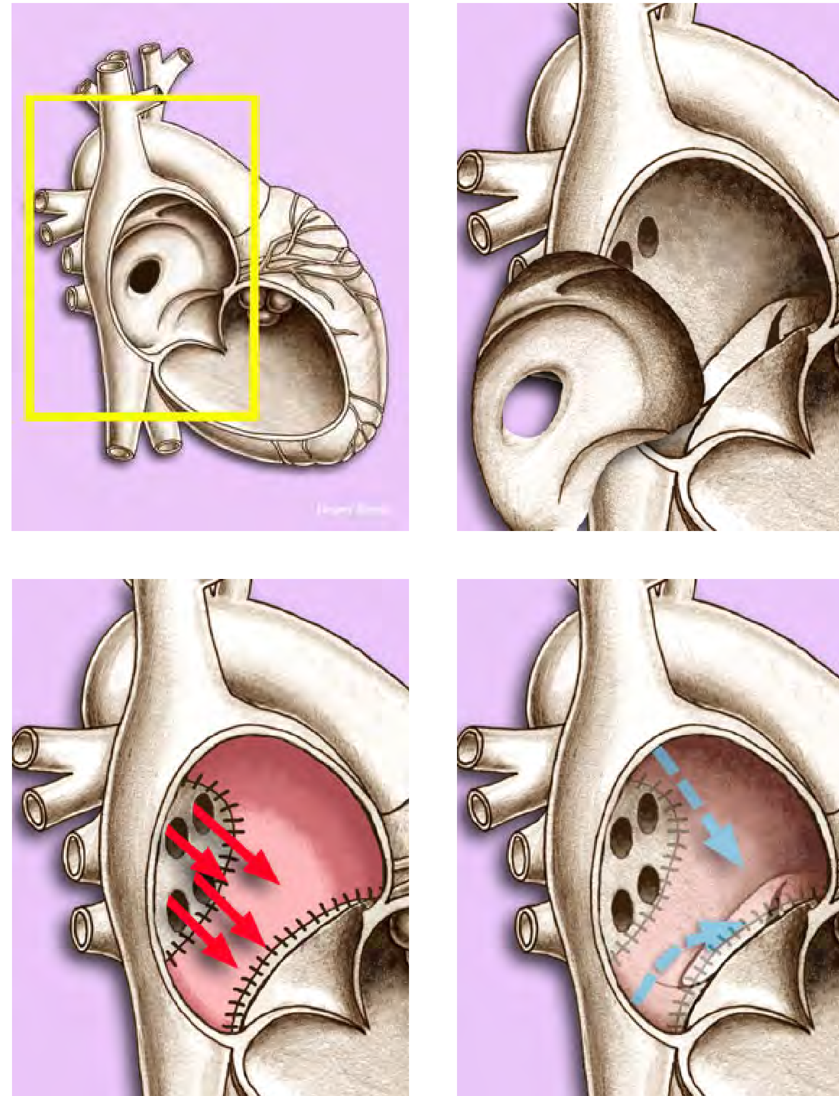
Neo-aortic insufficiency

- Almost always trivial/mild

LV function

Mustard Repair





Children's National

Atrial Baffle Repair
Long Term Sequelae

On going late mortality risk

- 20% mortality at 20 years

Arrhythmia

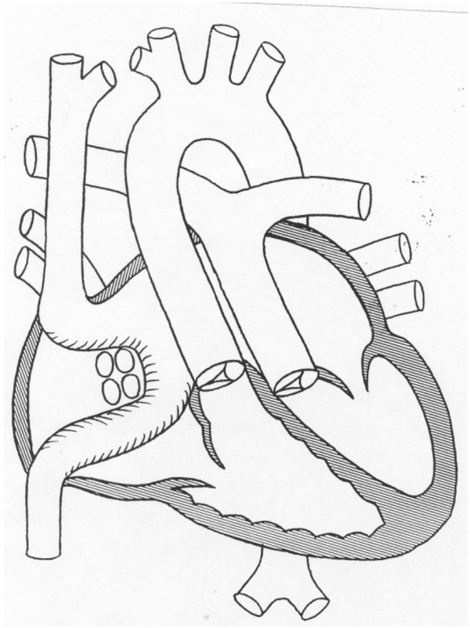
SVC obstruction -- 14-17%

IVC obstruction -- 1%

Baffle Leak -- Significant 1-2%

Systemic AV valve regurgitation -- 30%

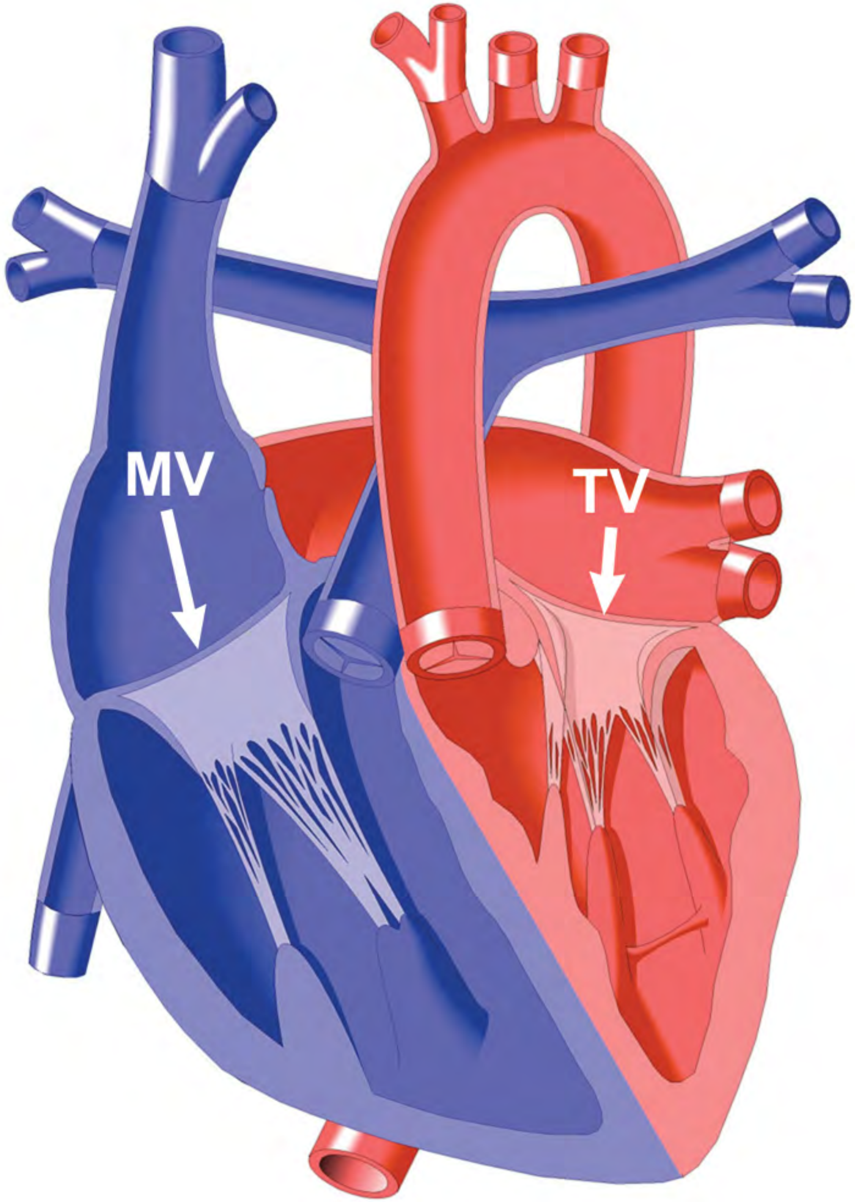
Systemic Ventricular Failure -- 15-20%



Children's National™

Transposition of the Great Arteries L Type

MV – mitral valve
TV – tricuspid valve



L-TGA

"Congenitally Corrected Transposition"

Atrio-ventricular and ventriculo-arterial discordance ("double discordance")

RA $\Rightarrow$ LV $\Rightarrow$ PA

LA $\Rightarrow$ RV $\Rightarrow$ Ao

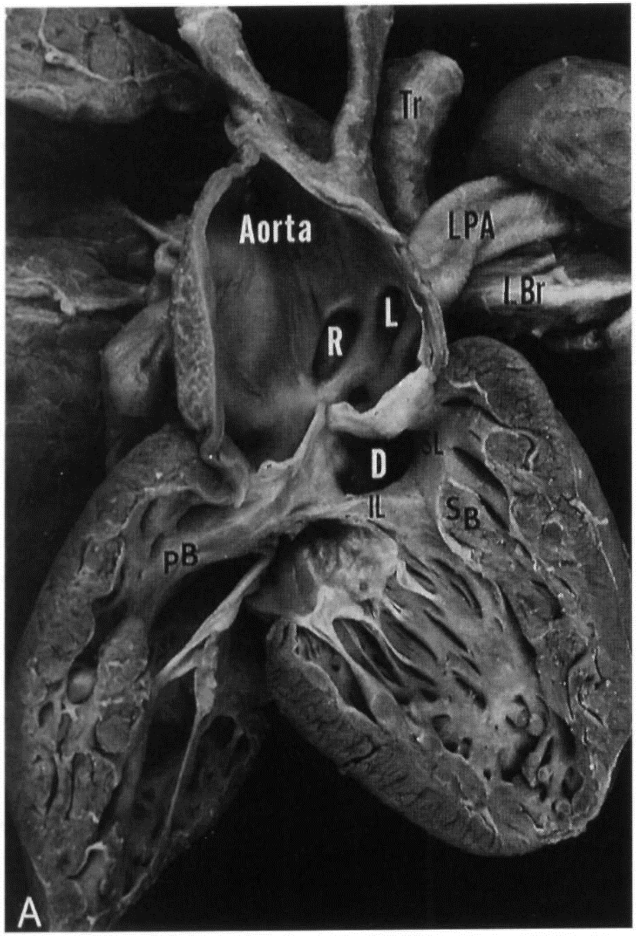
May be an isolated, asymptomatic finding or may be associated with other heart malformations

Children's National™

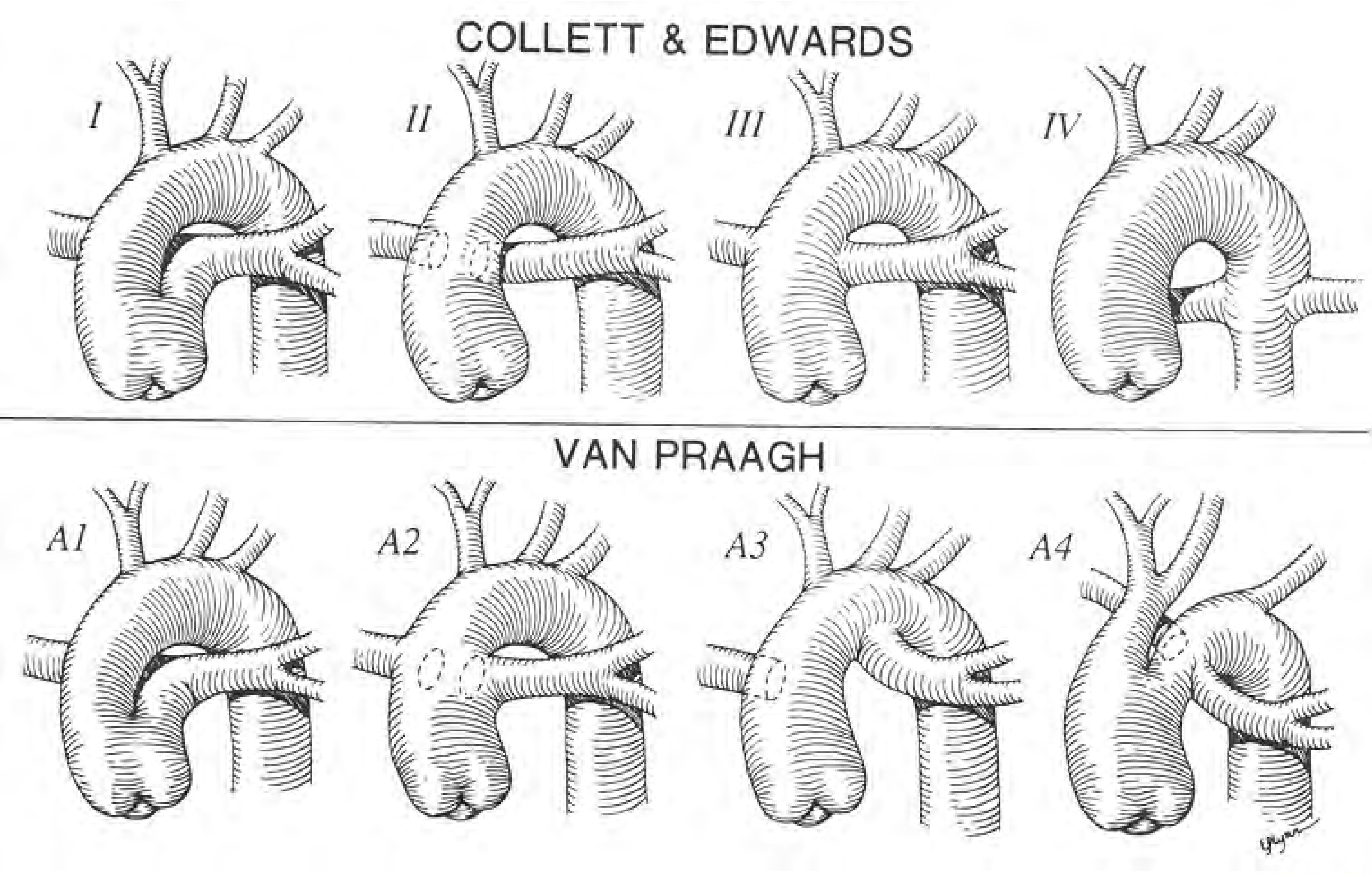
Truncus Arteriosus

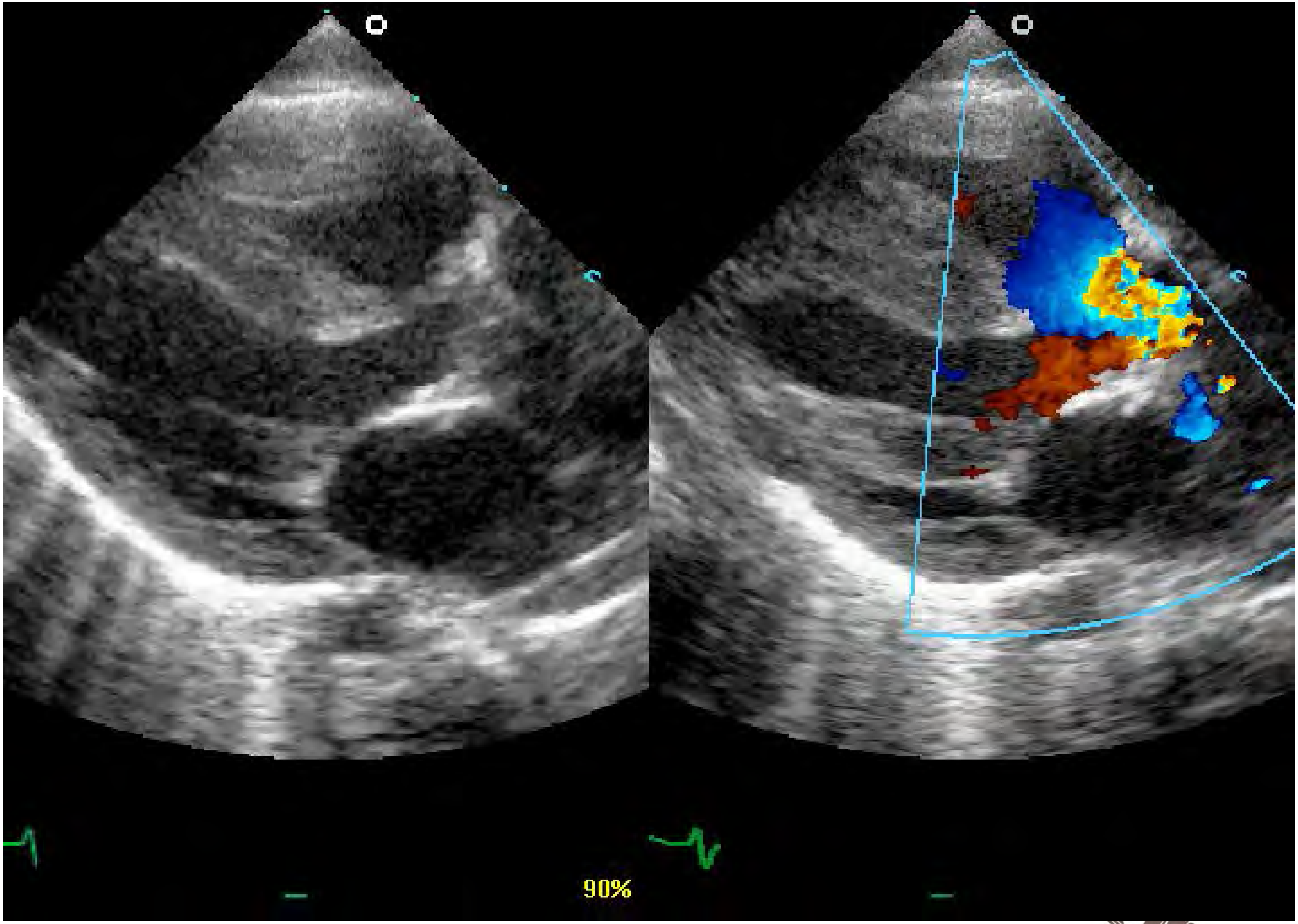
A single vessel arising from the heart and giving rise to the coronary, pulmonary and systemic circulations

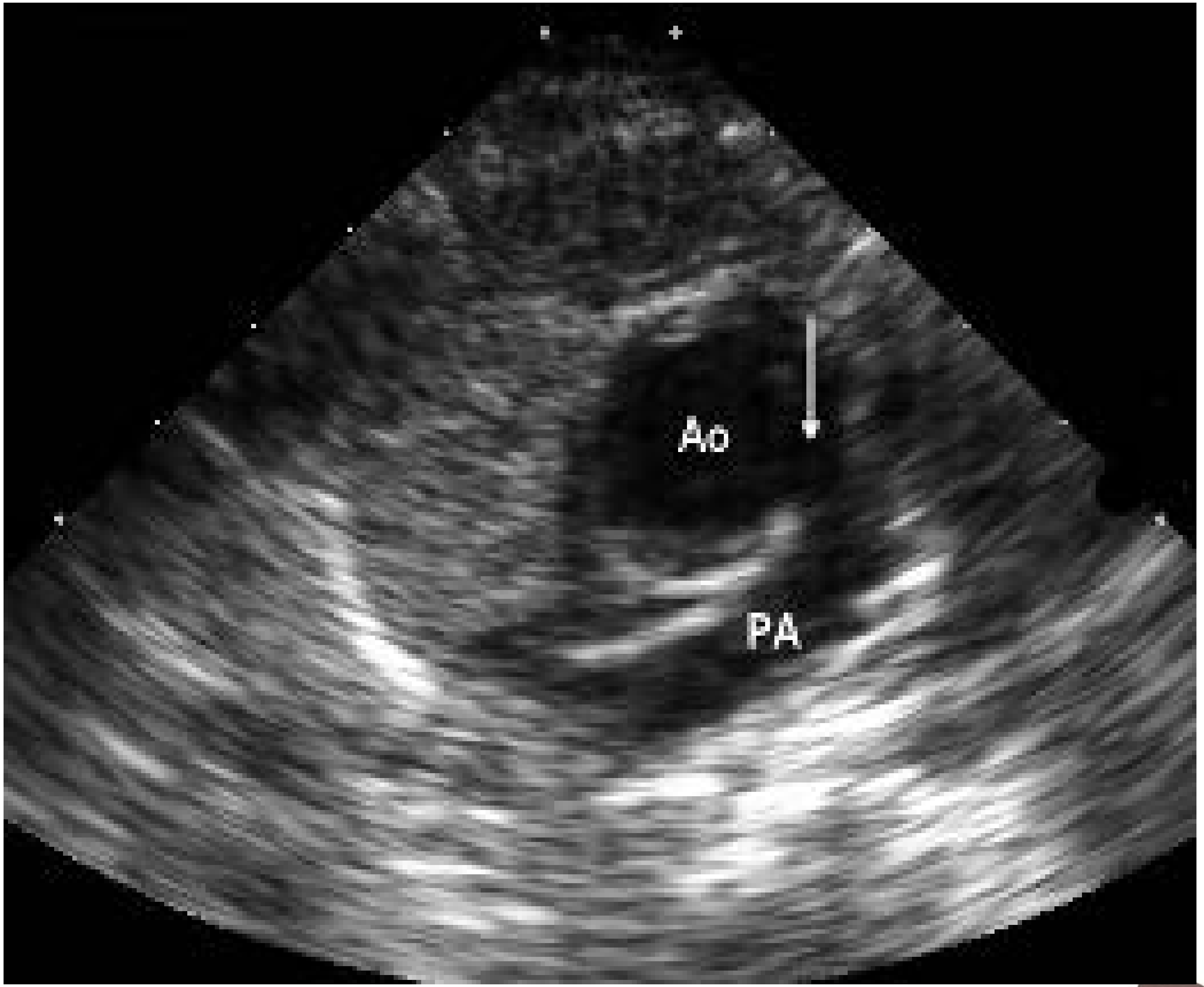
The VSD is the same as TOF



Truncus Arteriosus



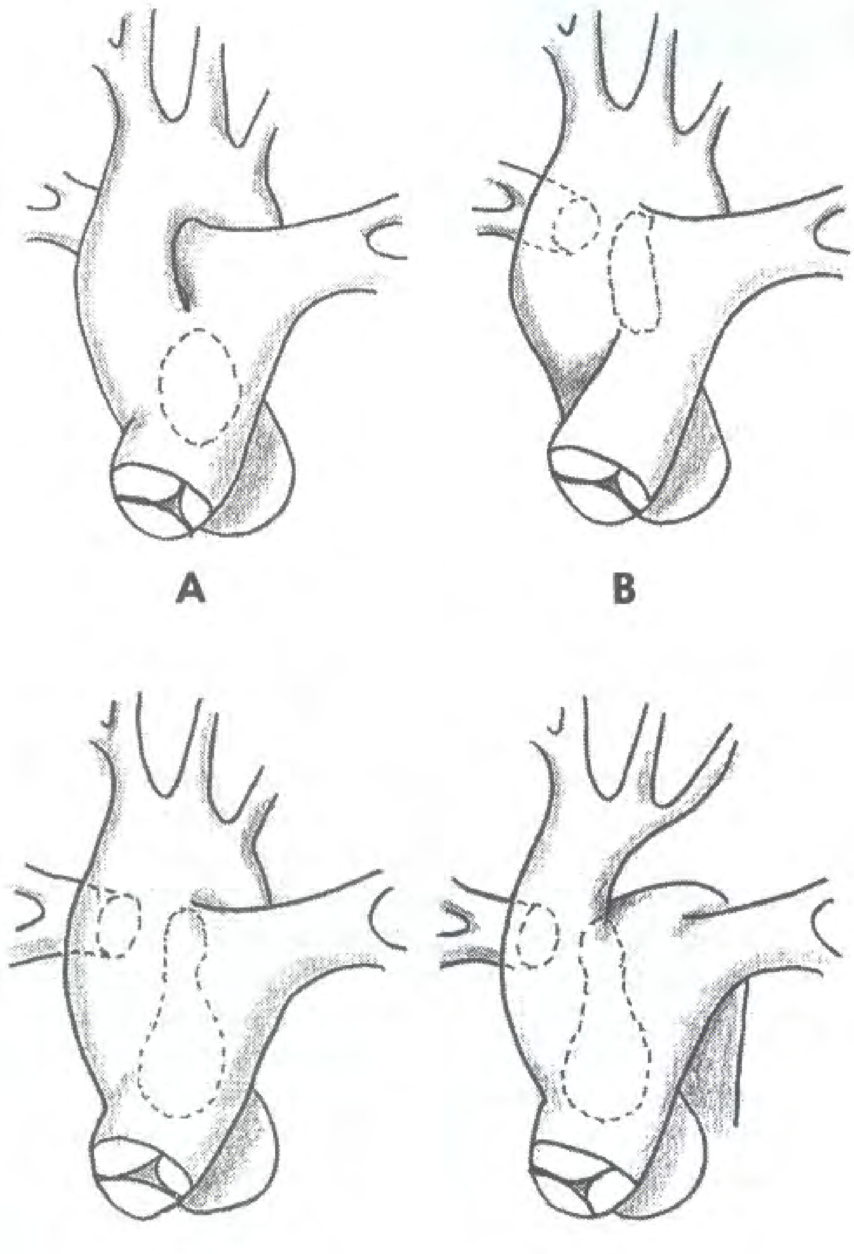

90%



Children's National™

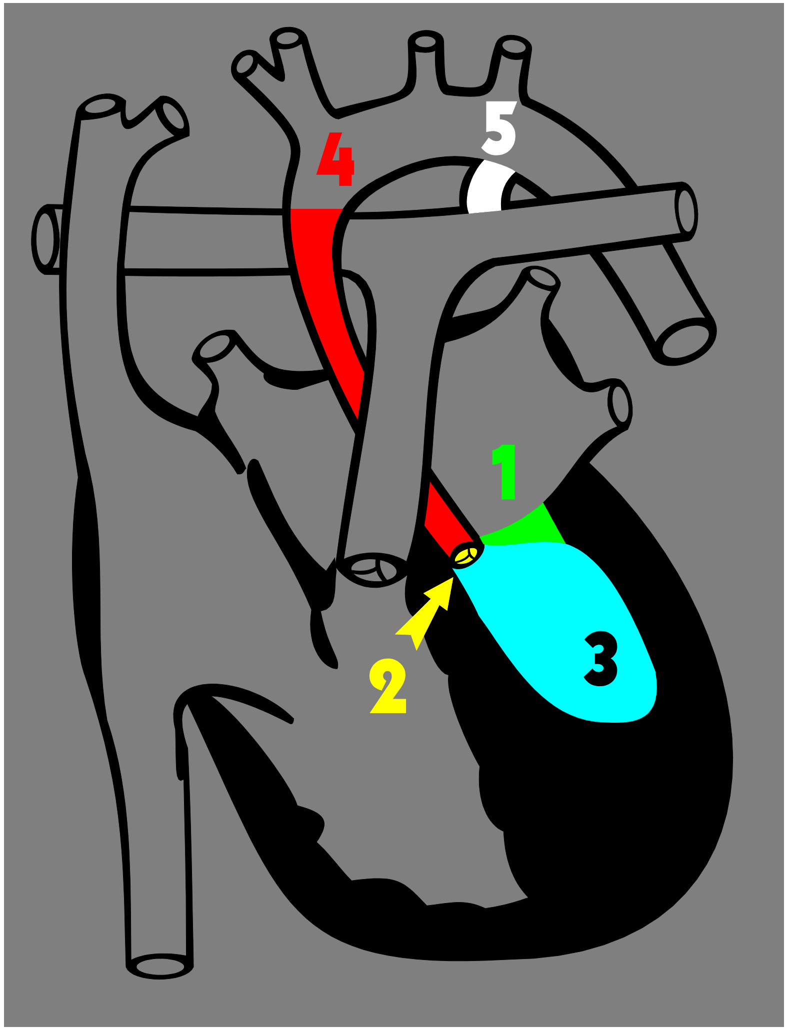
AP Window

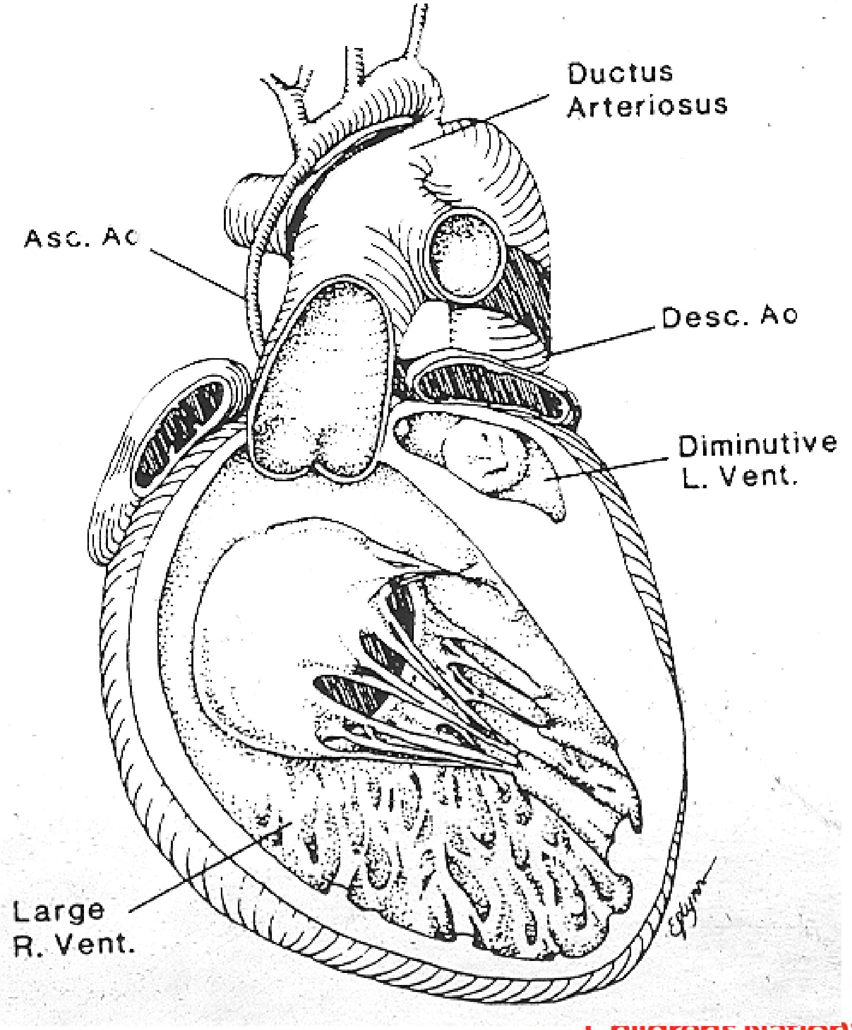
Communication between aort



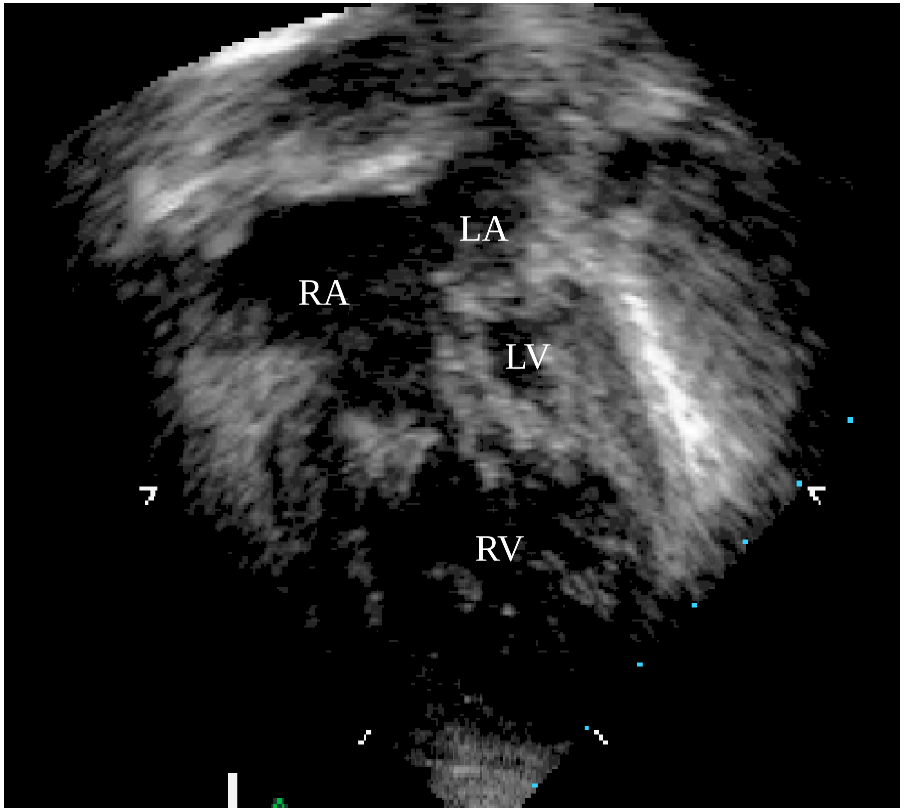
A B C D

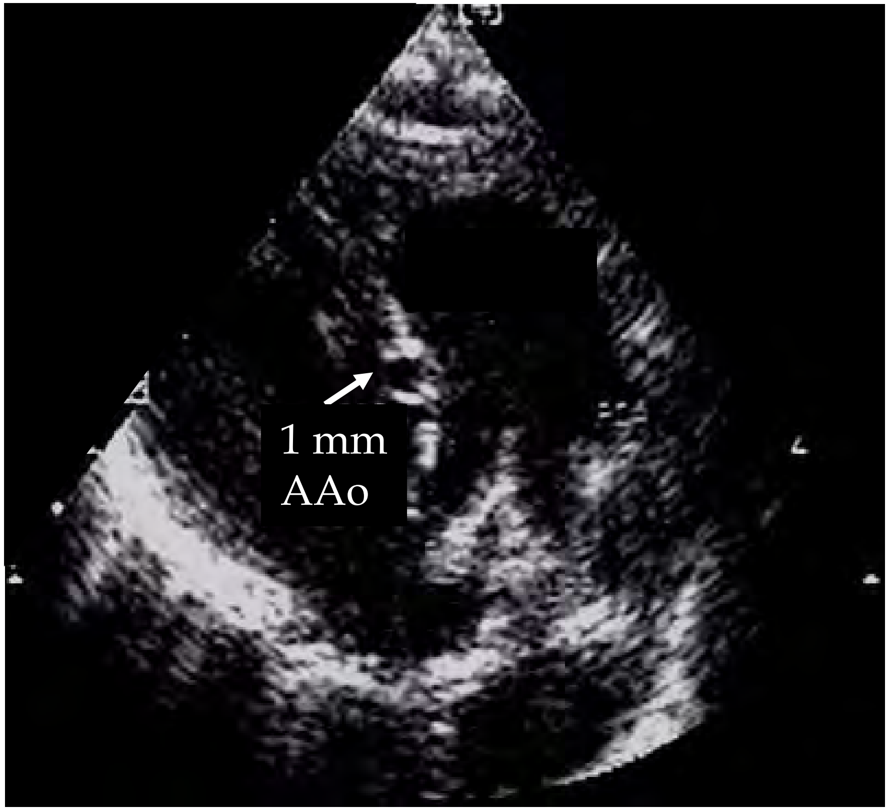
Hypoplastic Left Heart Syndrome



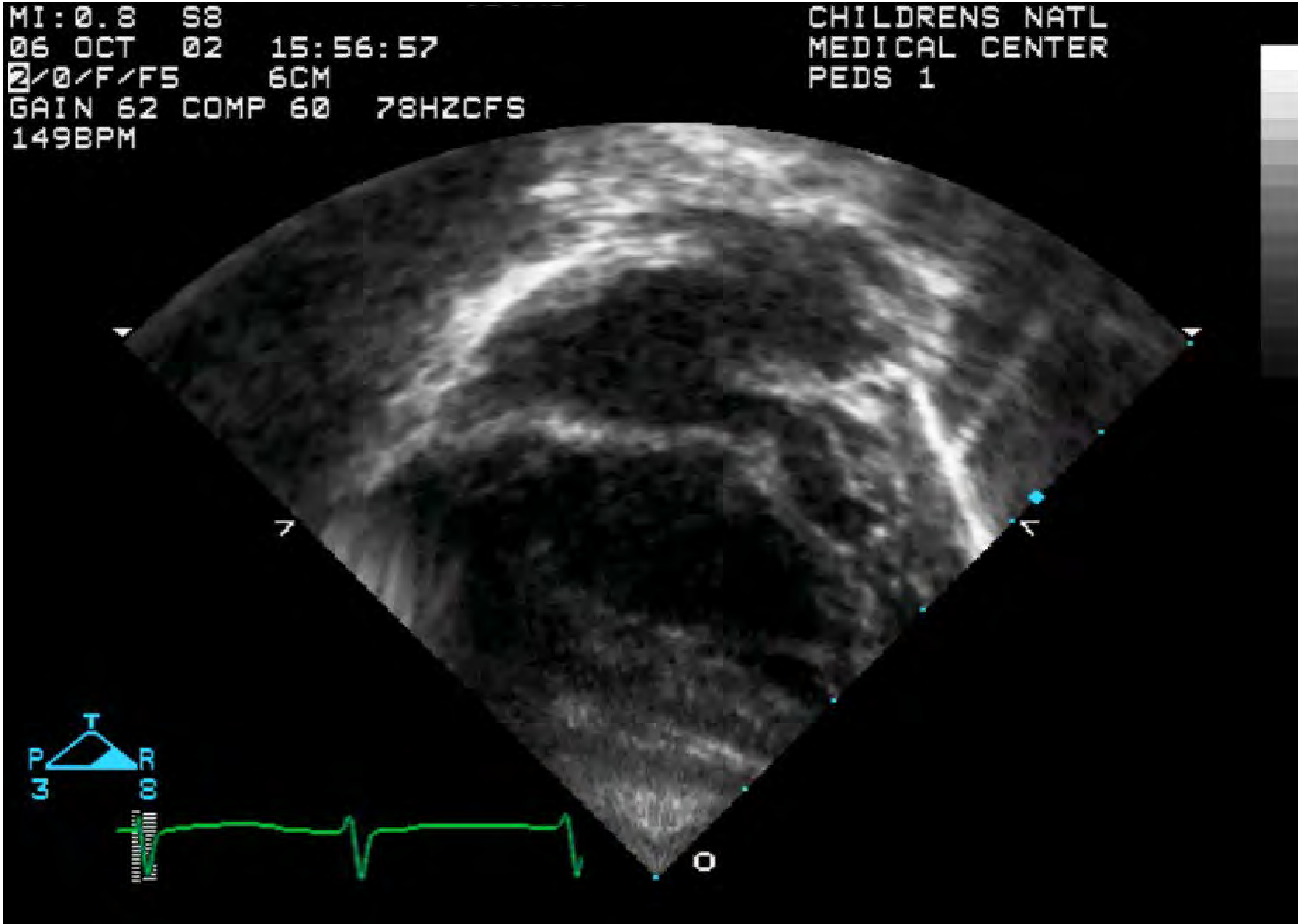


Hypoplastic Left Heart Syndrome





Children's National ™



MI:0.8 S8
06 OCT 02 15:56:57
2/0/F/F5 6CM
GAIN 62 COMP 60 78HZCFS
149BPM
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
T
P
R
3
8
O

BT Shunt: History

1924: Failing to obtain a surgical residency at Hopkins, Alfred Blalock goes to Vanderbilt and begins research on traumatic shock

1930: Vivien Thomas hired as Alfred Blalock's lab assistant

1938: Rabbit models with subclavian to PA anastomosis fail to produce pulmonary HTN

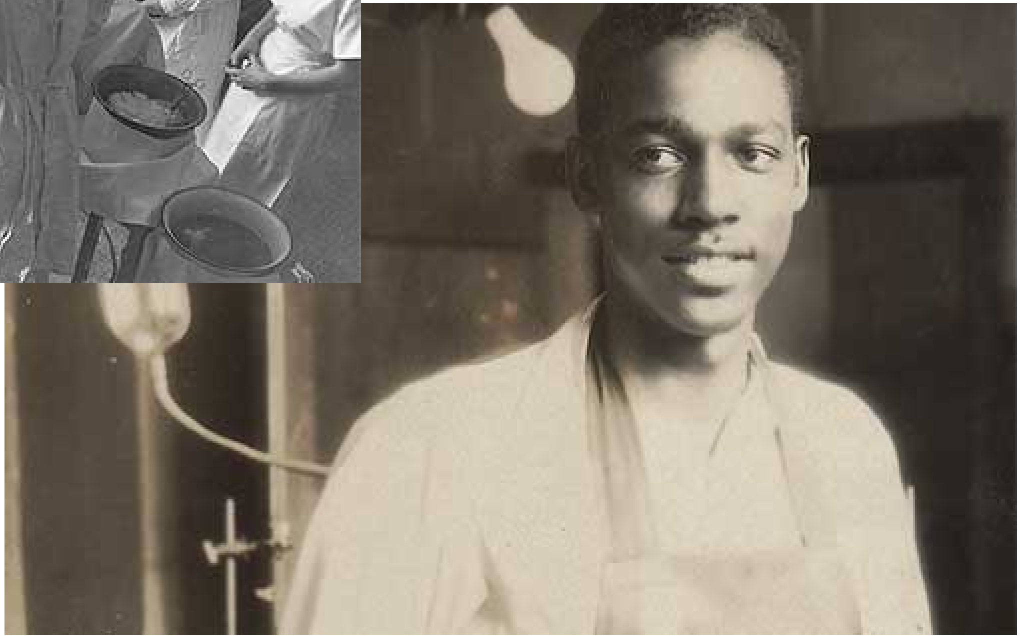
1941: Blalock and Thomas move to Hopkins

1941: Coarctation relief with subclavian to descending aorta shunt

1943: Helen Taussig, a Hopkins pediatrics residency graduate, approaches Blalock about help for "blue babies"

1944: "Anna," a dog with a surgically created mixing lesion, successfully undergoes end-to-side subclavian-to-PA anastomosis, lives 15 years

November 29, 1944: Eileen Saxon, a 15-month-old 4.5 kg undergoes successful systemic-to-pulmonary shunt by Blalock with Thomas directly over his shoulder

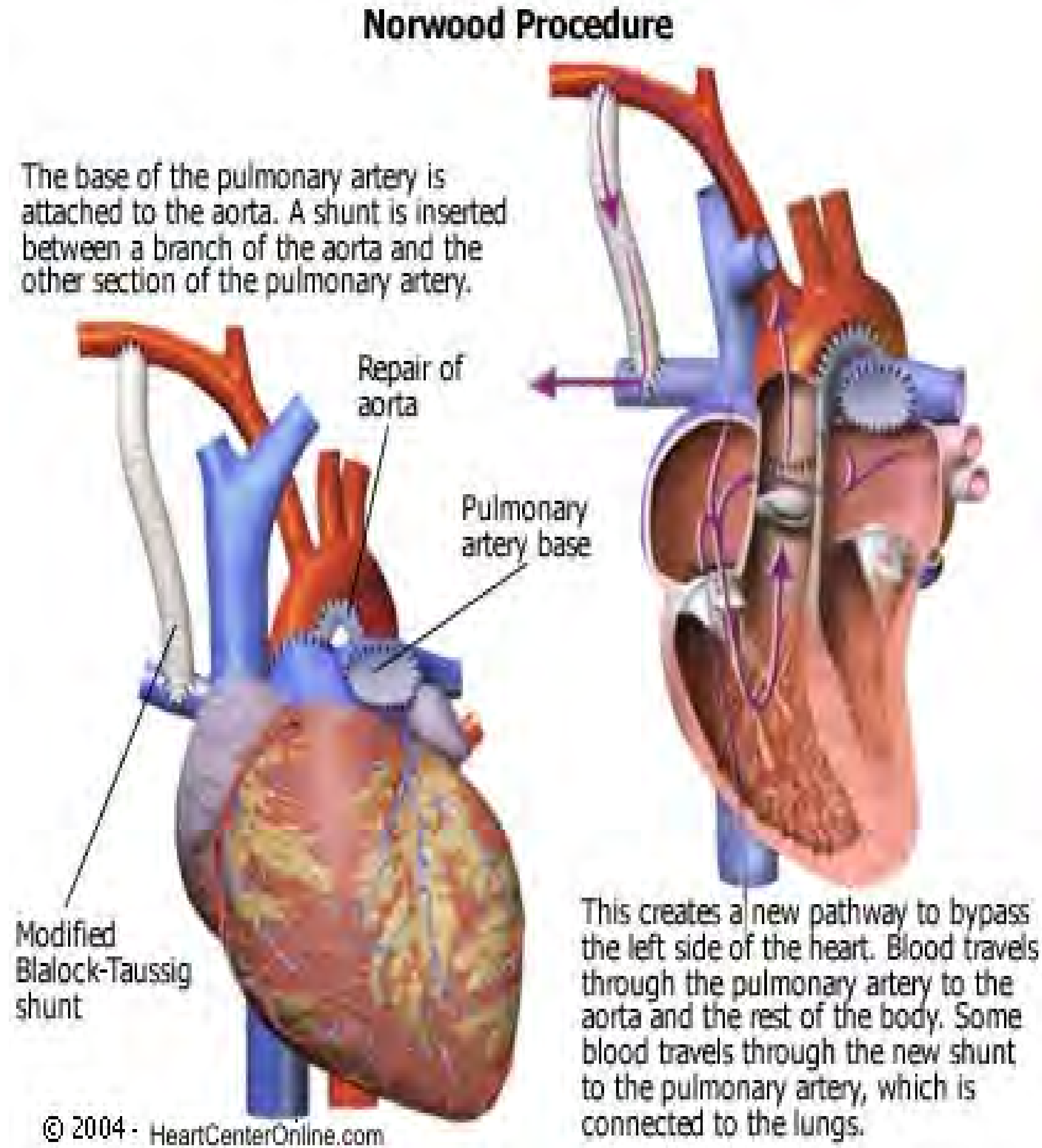


Children's National™

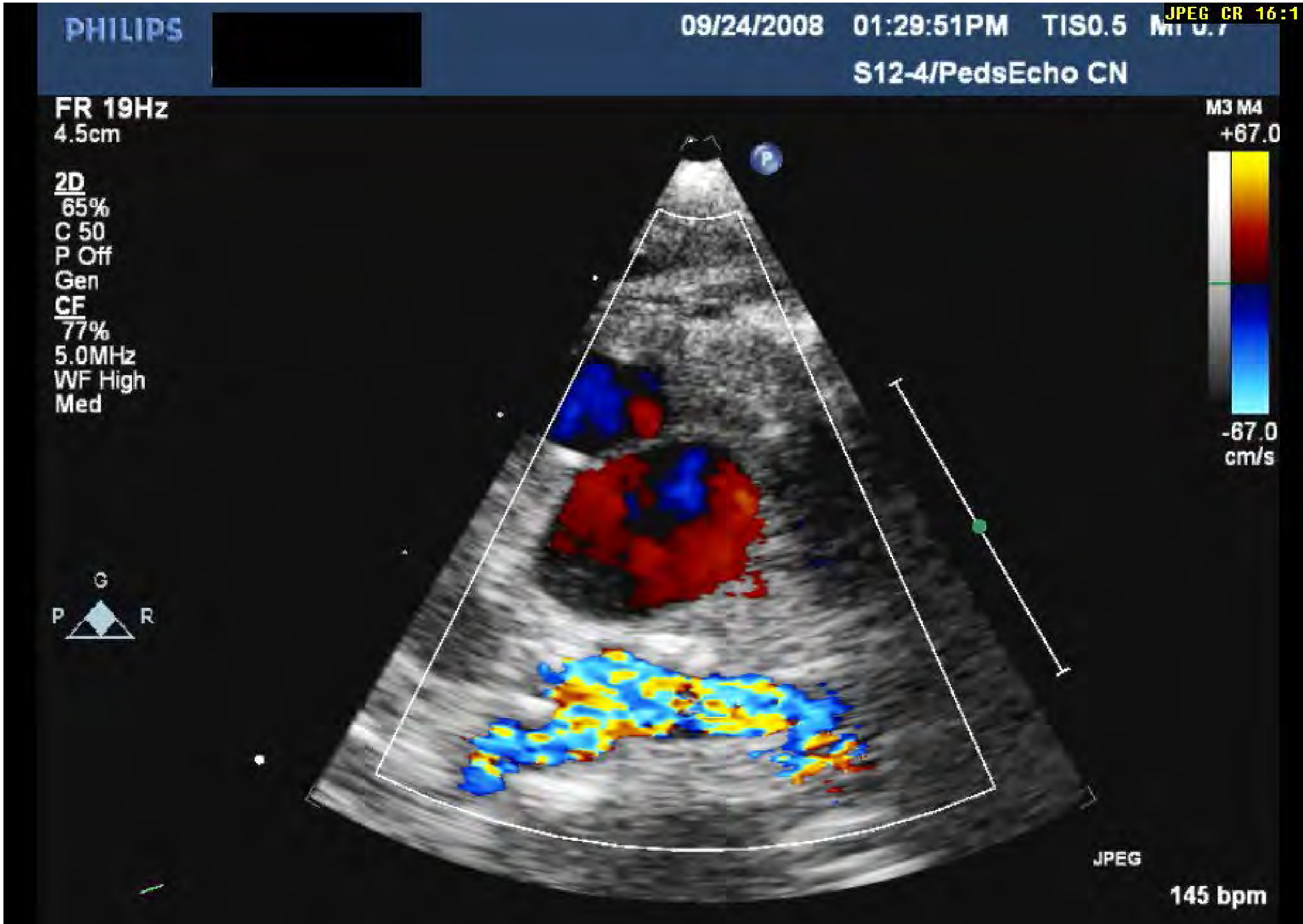
Norwood I: Anatomy

1. Atrial septectomy
2. Ligation of main pulmonary artery and construction of neo-aorta
3. Sano Modification/

Modified BT Shunt



BT Shunt

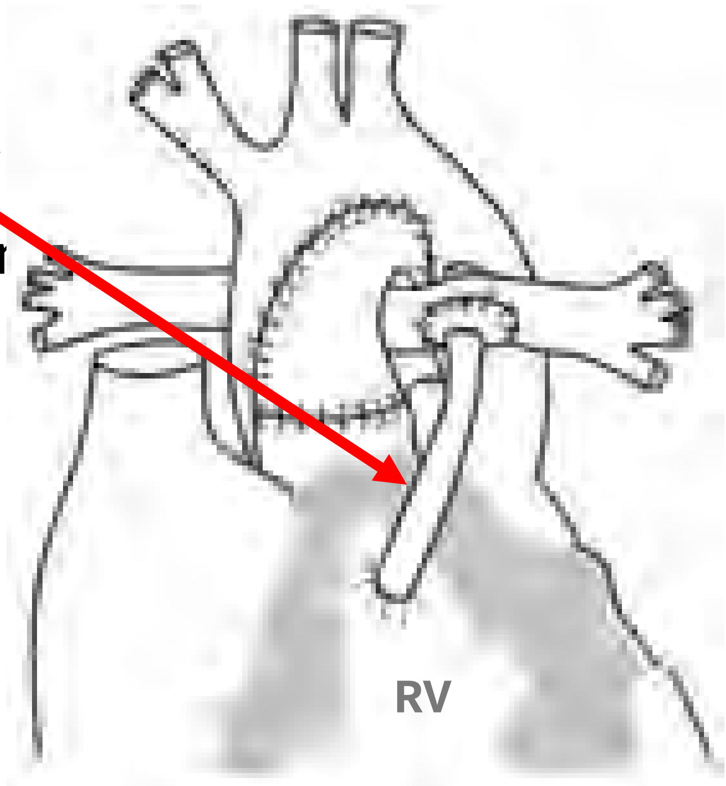


Children's National

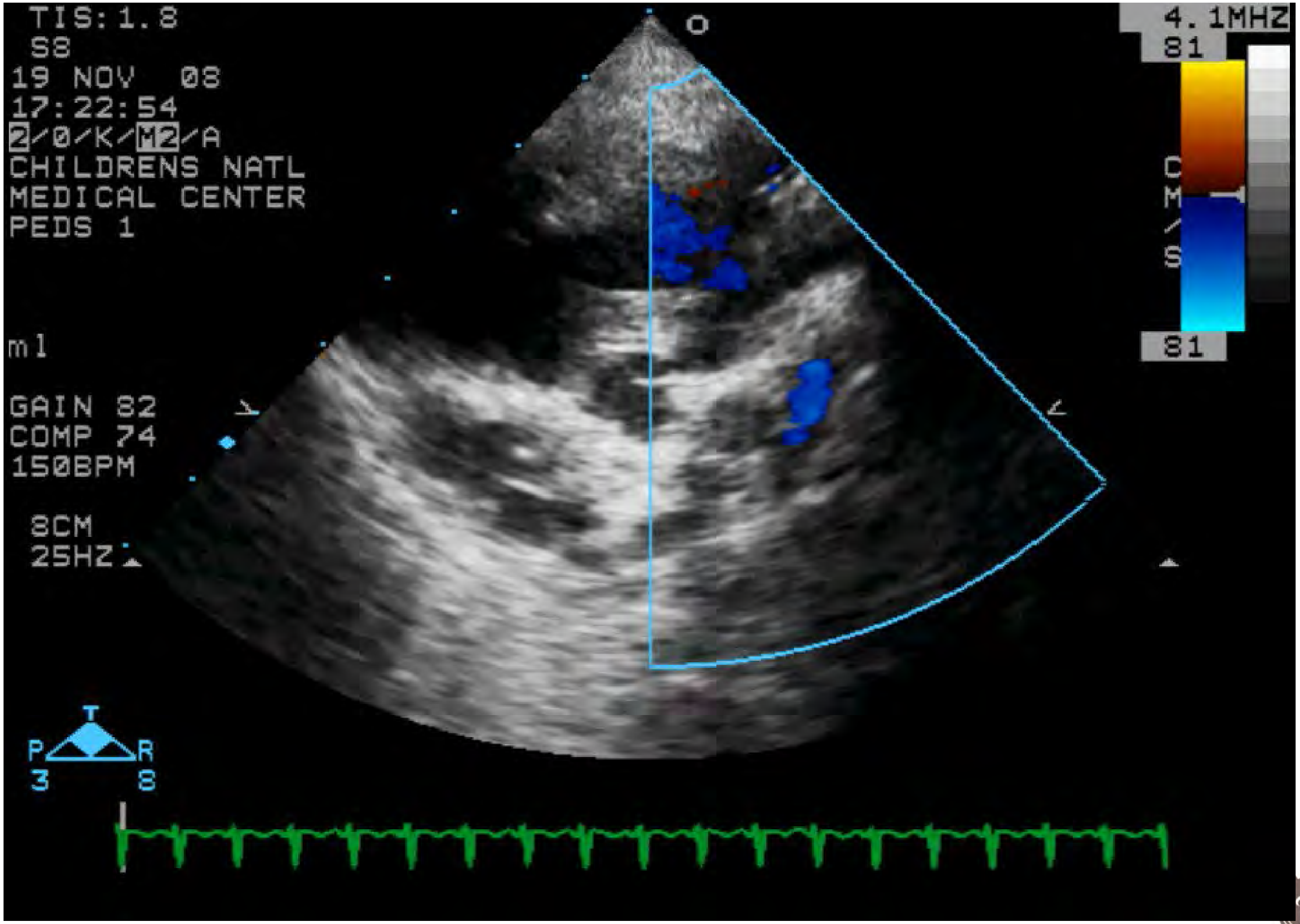
Norwood I: Sano

Sano modification

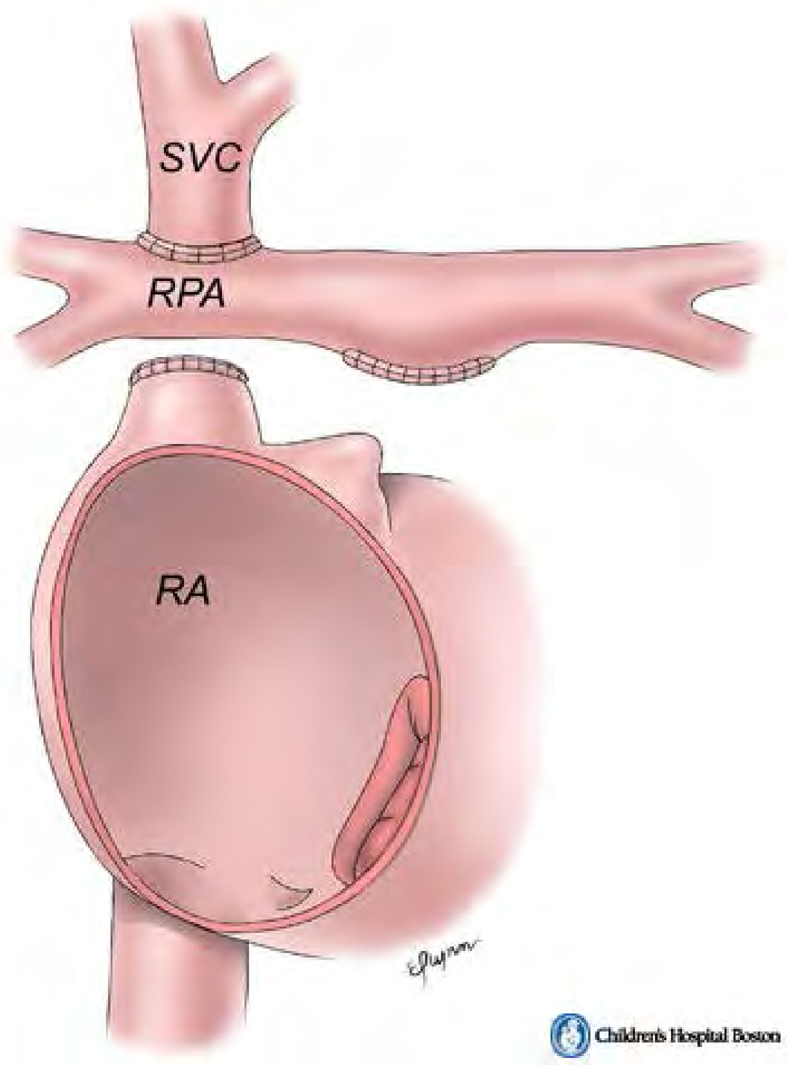
- RV-to-PA conduit
- Eliminates competitive flow
- Enhances coronary perfusion



Sano Shunt


TIS:1.8
S8
19 NOV 08
17:22:54
2/0/K/M2/A
CHILDRENS NATL
MEDICAL CENTER
PEDS 1
ml
GAIN 82
COMP 74
150BPM
8CM
25HZ
4.1MHZ
81
C
M
/
S
81
T
P
R
3
8

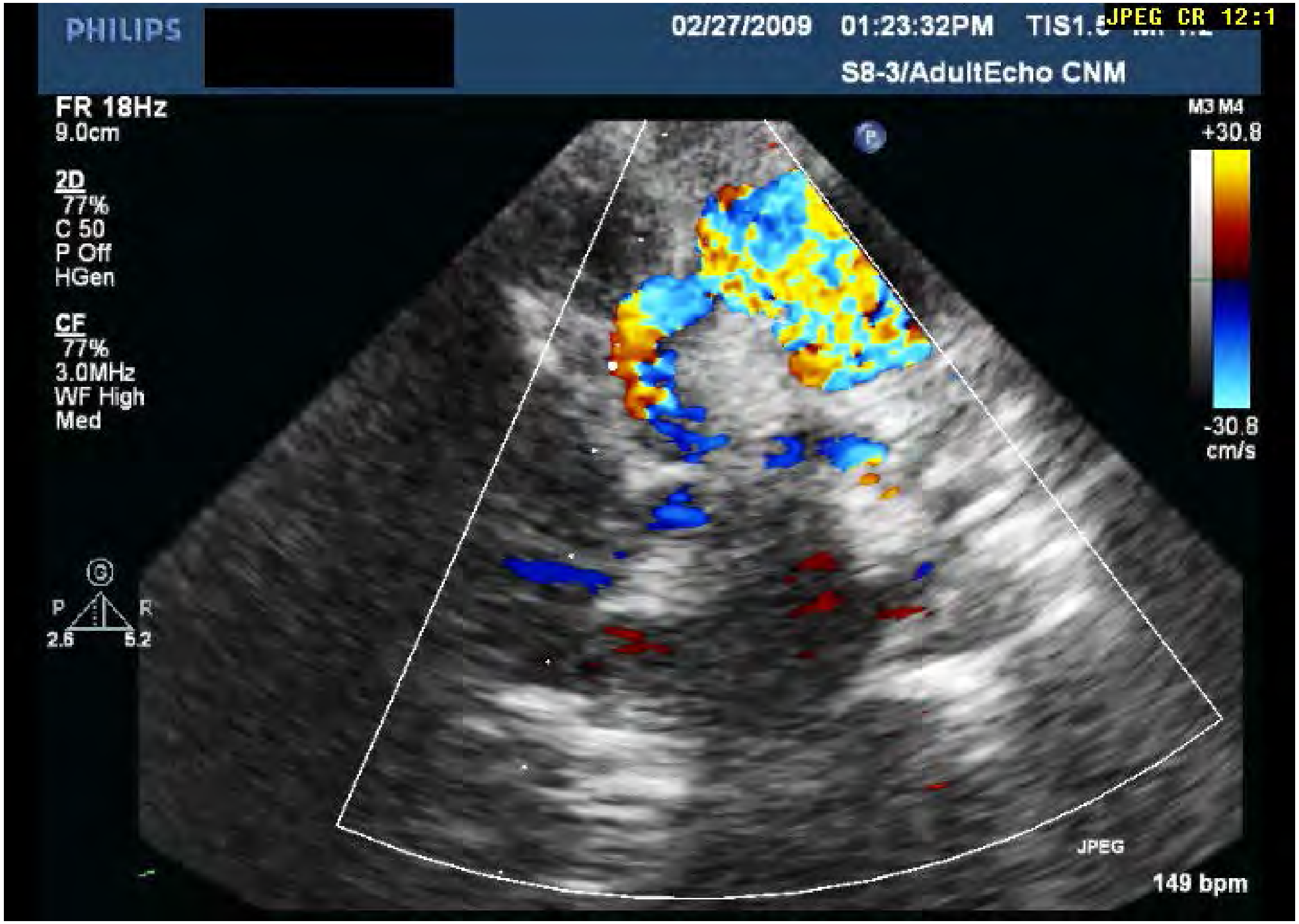
Bidirectional Glenn: Anatomy



- End-to-side anastomosis of SVC to undivided right pulmonary artery
- Includes takedown of BT shunt
- Allows flow to both lungs from SVC via passive flow

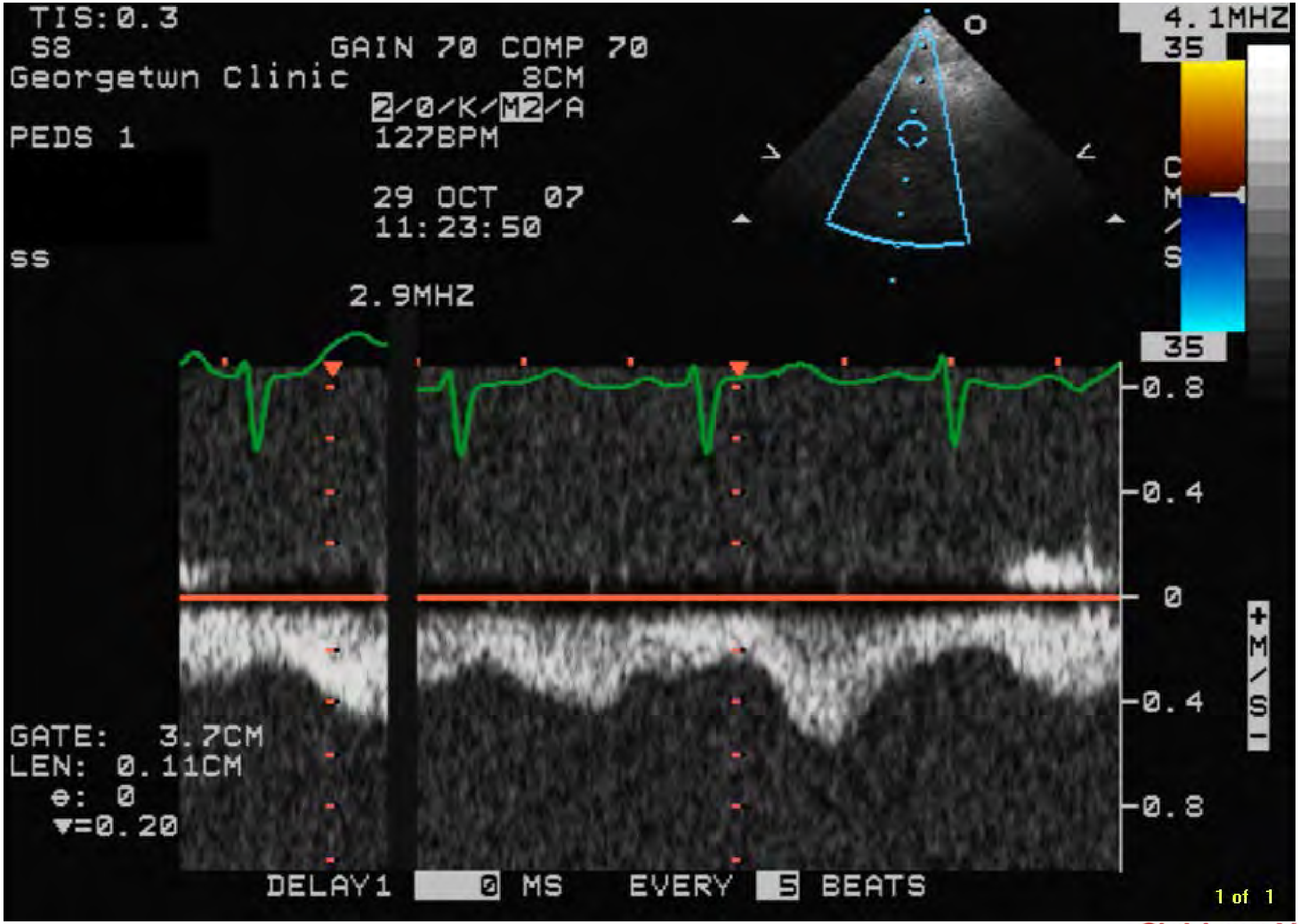
Children's National™

Glenn Shunt

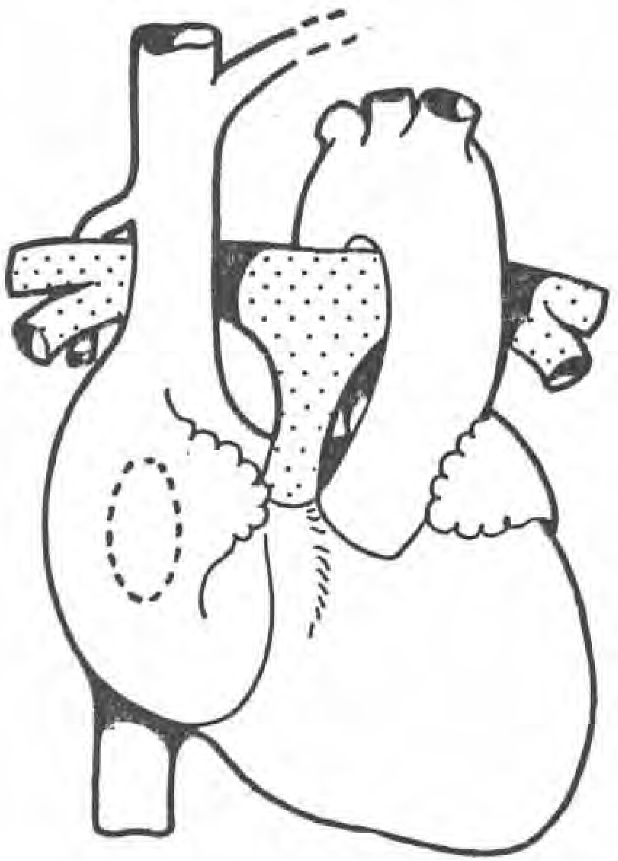


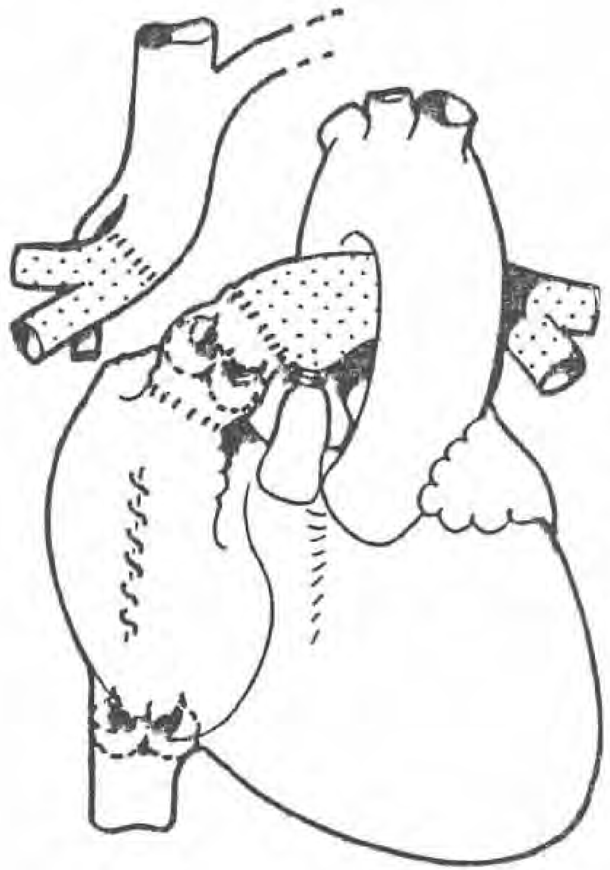
Children's National™

Glenn Doppler



Original Fontan





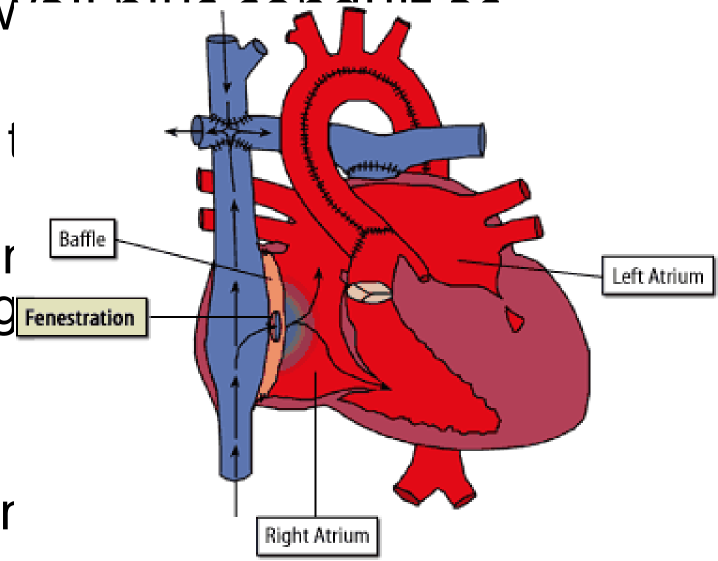
Fontan: Variations

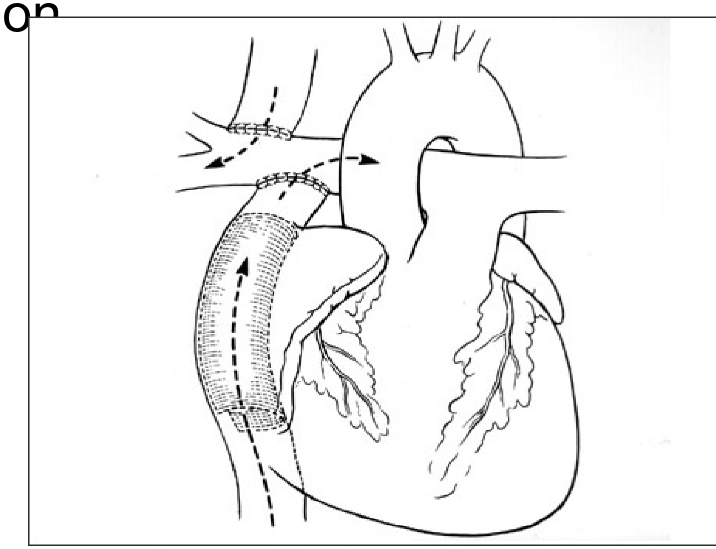
Lateral tunnel runs within RA, using free v
baffle for IVC blood

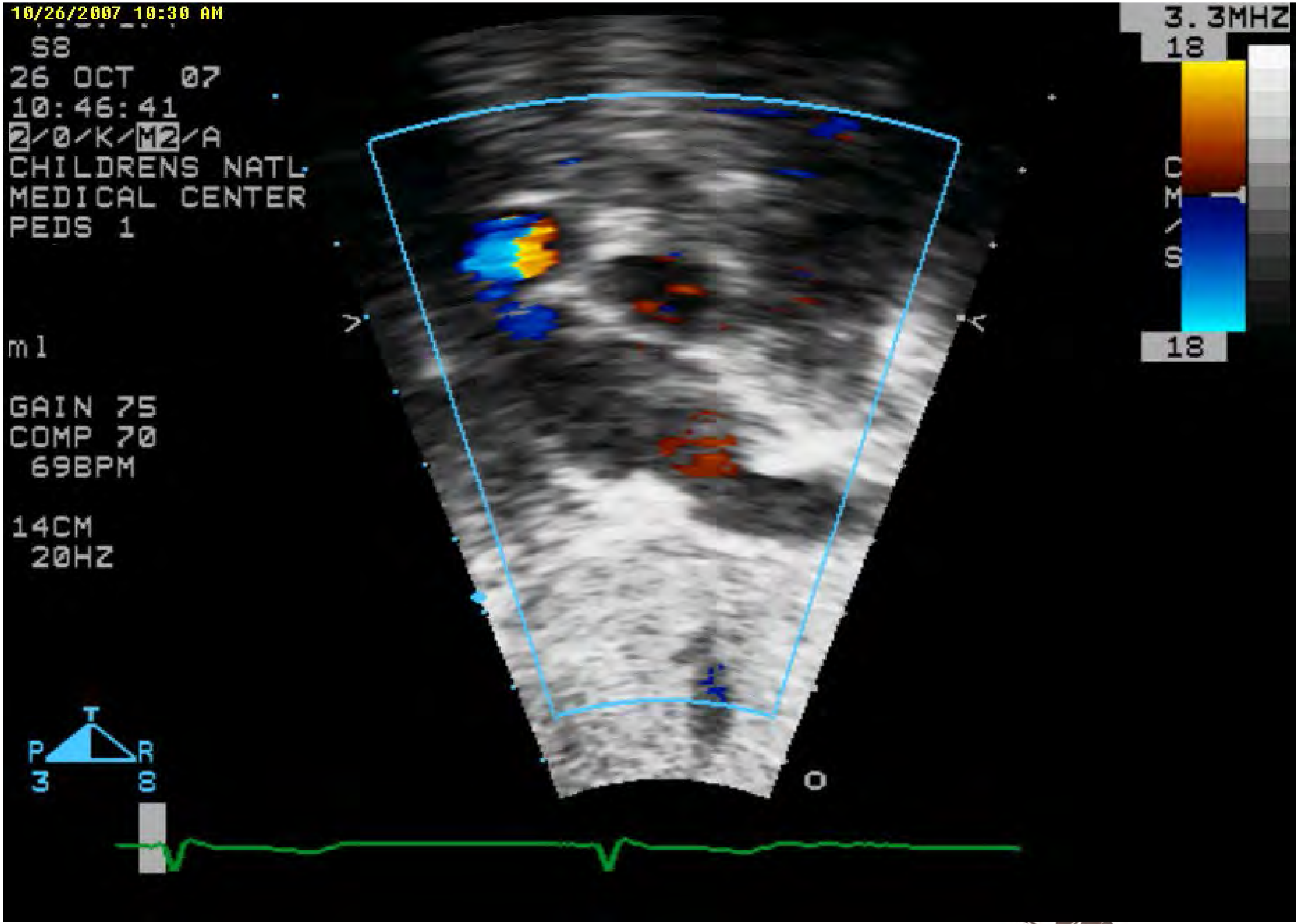
- Fenestrations: R-to-L shunting through t
hypoxemia
- Improve cardiac output, minimize syster
decrease post-op thoracostomy drainag
- Can later be closed by cath

Extracardiac is IVC to MPA

- Generally has lower rate of complicatior
- Foreign material requires anticoagulatio

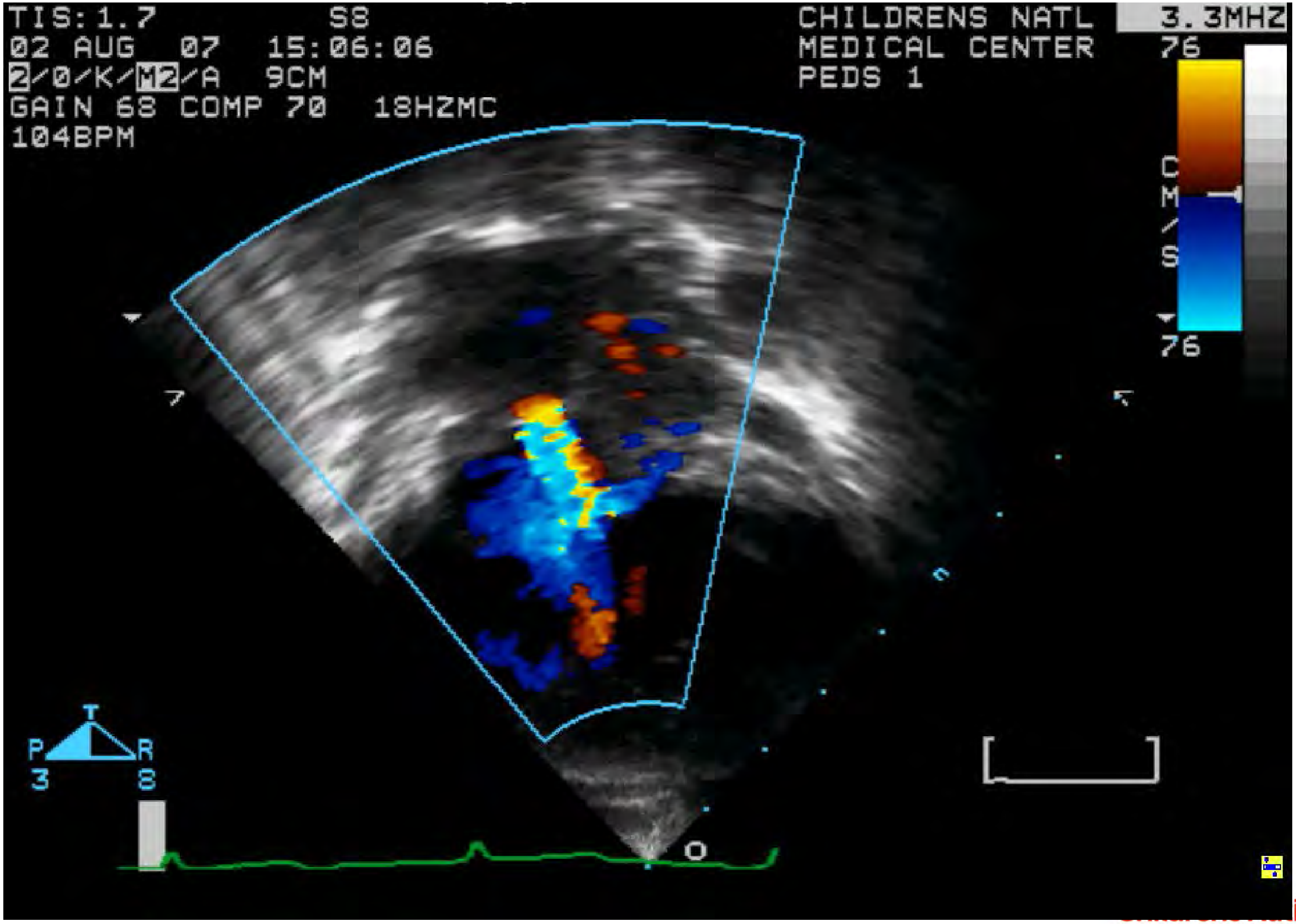






Children's National

Fenestrated Fontan



Hypoplastic Left Heart Syndrome Palliative Reconstruction

Stage I -- Norwood Procedure

- Birth

Stage II -- Bi-directional Cavopulmonary Shunt

- 4-6 months

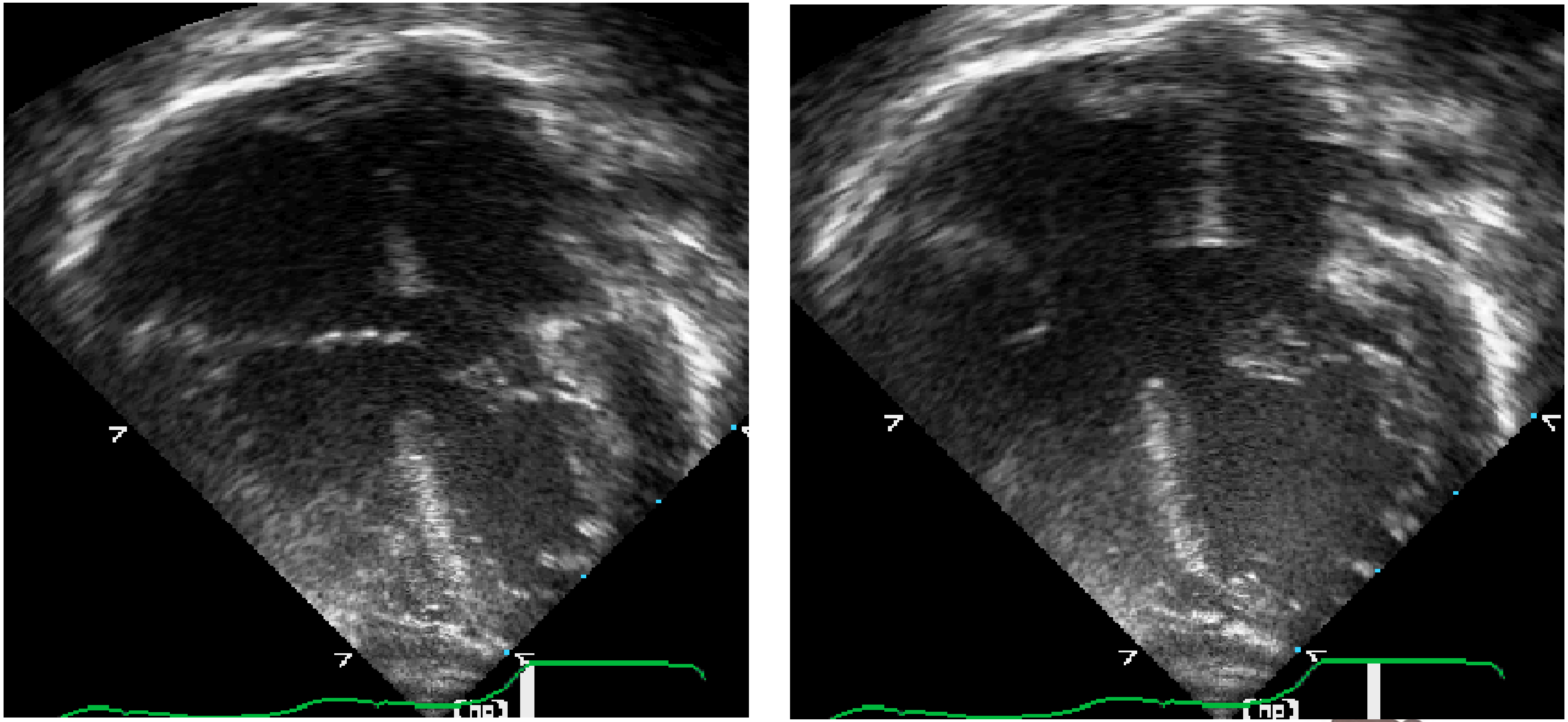
Stage III-- Fontan Procedure

- 18-24 months for lateral tunnel procedure
- > 15 kg for extracardiac procedure

Children's National™

QUESTION 1

A tachypneic 2 month old is not growing well and has a murmur. An echocardiogram is obtained:



SYSTOLE DIASTOLE

Children's National™

E

QUESTION 1 (CONT)

All of the following statements are likely to be true except:

A. The patient is at increased risk to have Down Syndrome

B. The patient's pulmonary artery pressure is normal

C. The patient has an endocardial cushion defect

D. The patient has a normal oxygen saturation

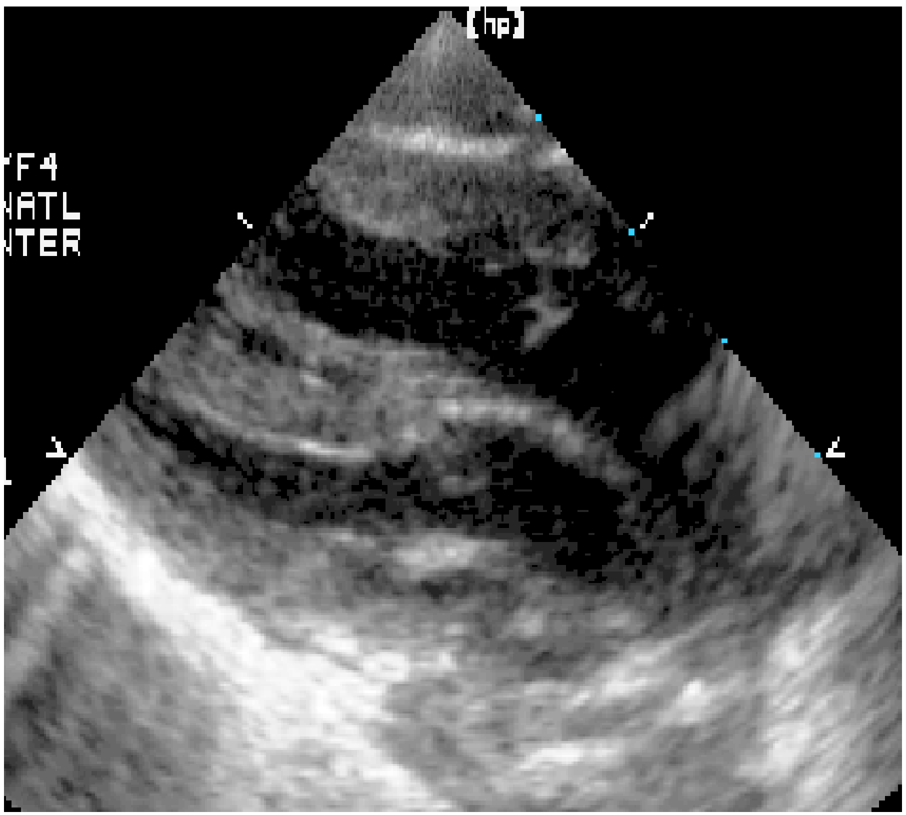
E. The patient may have a small mitral valve cleft after surgical repair

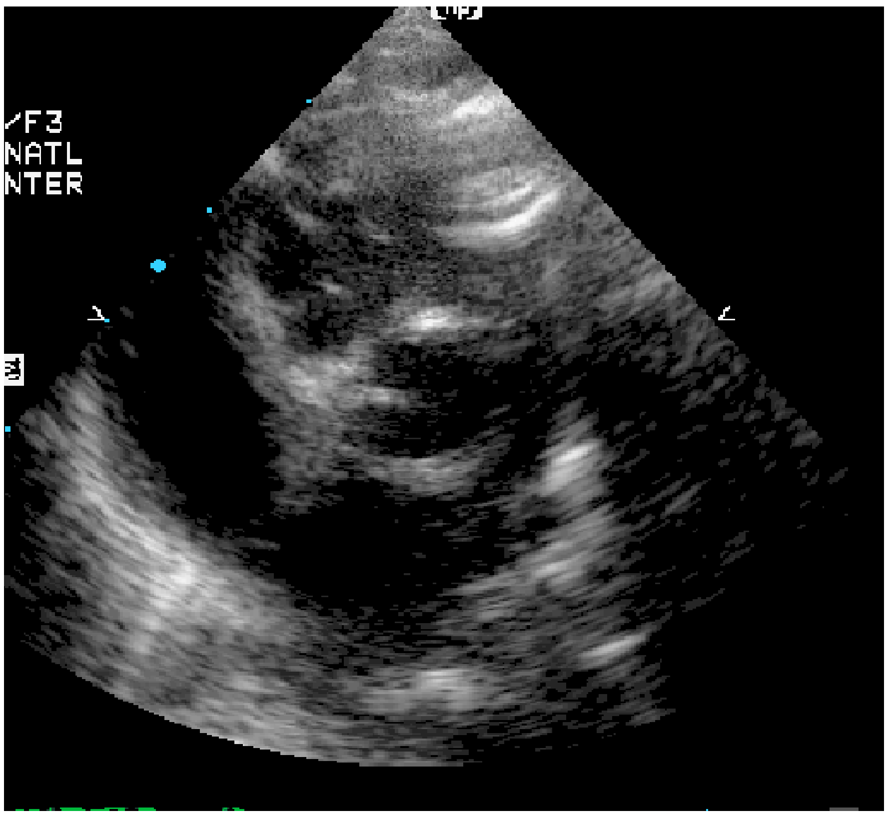
Children's National ™

E

QUESTION 2

A cyanotic newborn has the following echocardiogram:





Children's National

E

QUESTION 2 (CONT)

All of the following statements are likely to be true except:

A. The aorta is malposed anterior and rightward

B. The right ventricle pumps blood to the body

C. Oxygenated blood is pumped to the lungs

D. The left ventricle pumps blood to the body

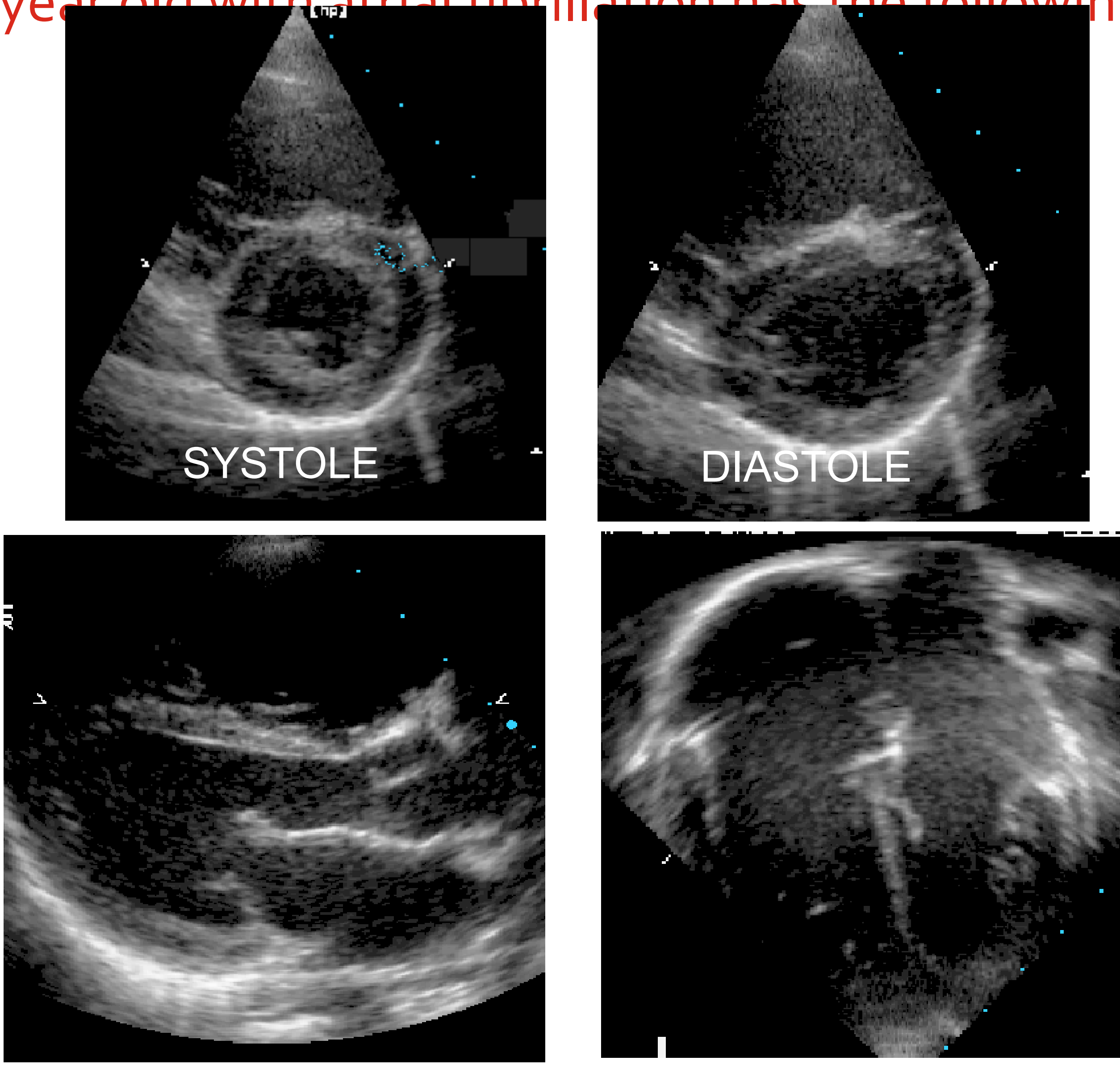
E. The right ventricular pressure is greater than or equal to the left ventricular pressure

Children's National™

E

QUESTION 3

A 40 year old with atrial fibrillation has the following echo:



E

Children's National™

QUESTION 3 (CONT)
Subsequent imaging is most likely to reveal the following

A. Tetralogy of Fallot

B. Large membranous ventricular septal defect

C. Large patent ductus arteriosus

D. Large secundum atrial septal defect

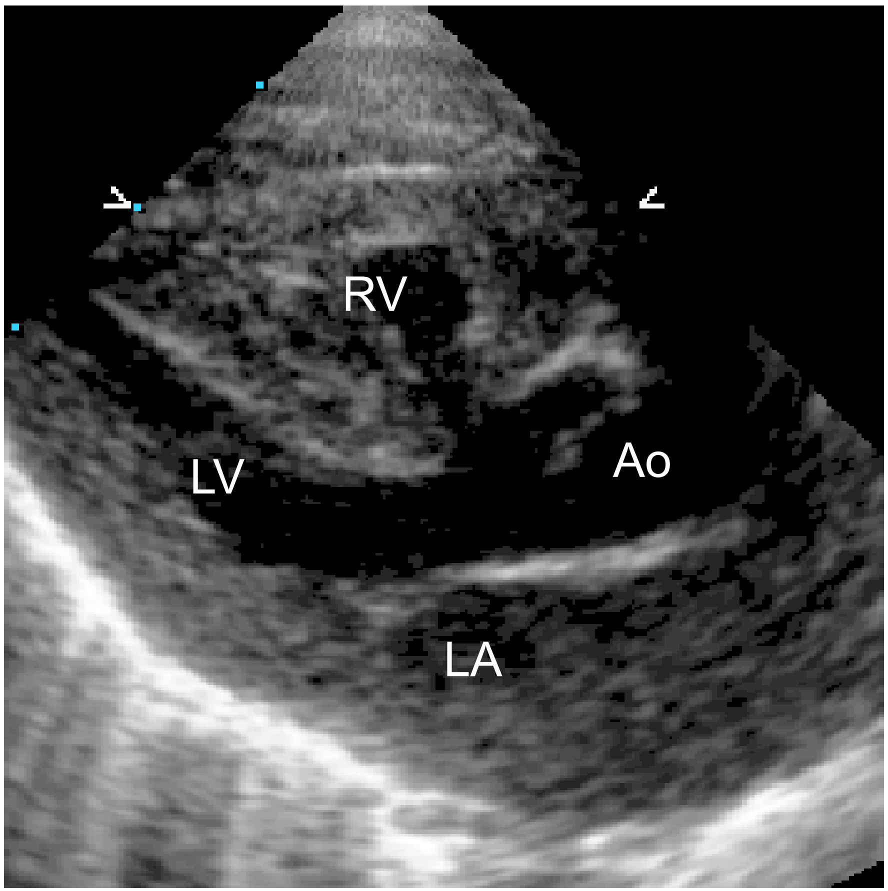
E. No structural cardiac defect

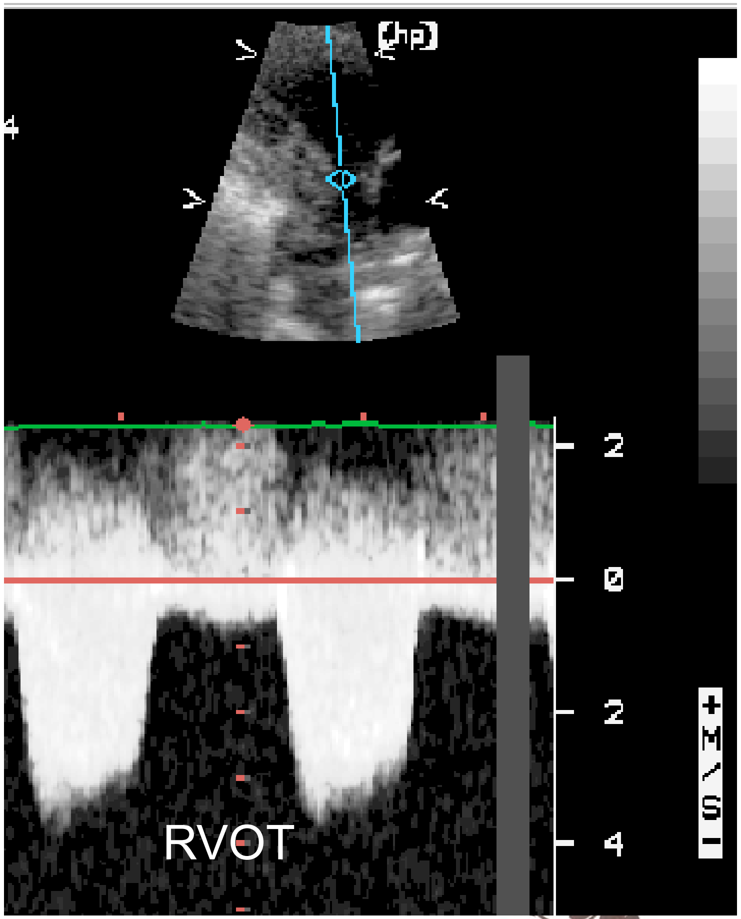
Children's National ™

E

QUESTION 4

A 3 month old with a loud murmur and intermittent perioral cyanosis has the following echo:





E

QUESTION 4 (CONT)
All of the following statements are likely to be true except:

A. The aorta is overriding the left and right ventricle

B. There is a large ventricular septal defect

C. There is pulmonary stenosis

D. The right ventricular pressure is increased

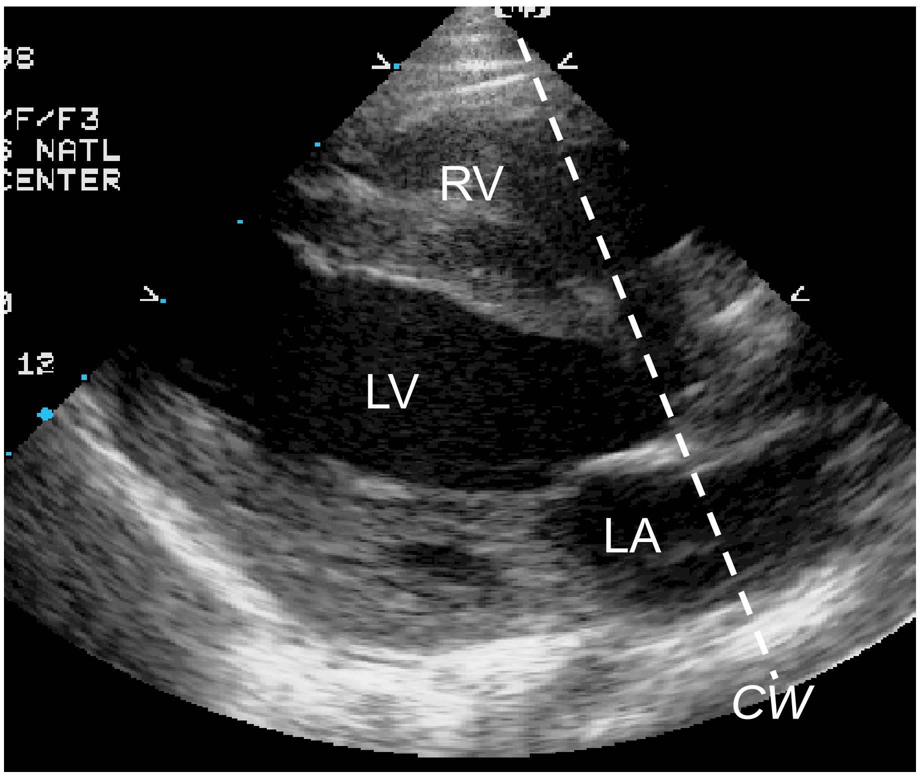
E. The pulmonary artery pressure is increased

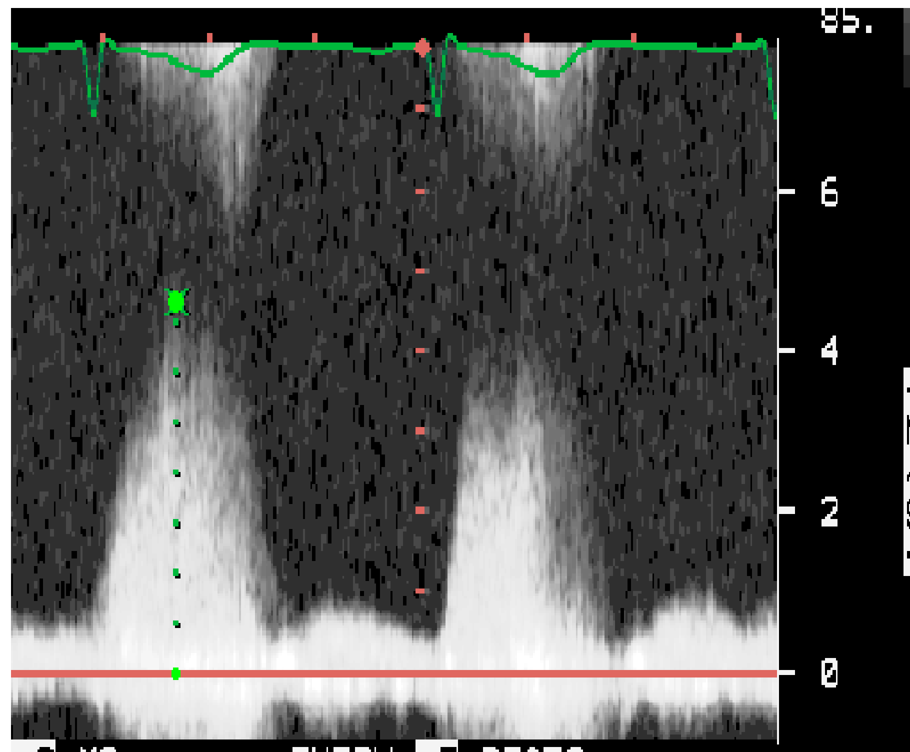
Children's National™

E

QUESTION 5

An asymptomatic 9 month old with a loud murmur and a BP of 79/48 and has the following parasternal long axis 2D and CW Doppler findings:





Children's National

E

QUESTION 5 (CONT)
The most likely diagnosis is:

A. Membranous VSD, normal RV pressure

B. Membranous VSD, elevated RV pressure

C. Muscular VSD, normal RV pressure

D. Muscular VSD, elevated RV pressure

E. Tricuspid regurgitation, elevated RV pressure

E

Acknowledgements

Unattributed illustrations are from Nadas' Pediatric Cardiology

Amy L. Juraszek

Margaret Lasota

Children's National Medical Center

- **202-476-4880 Physician Line**
- **202-476-5579 Echo Lab**

Children's National ™