Echocardiography in Pulmonary Arterial Hypertension in Congenital Heart Disease

Mark K. Friedberg, MD

SickKids®
The Labatt Family Heart Centre

Outline

- Introduction
- Congenital lesions that cause PAH (increased pulmonary pressures)
- Assessment of RV function in PAH
- Evaluation of ventricular-ventricular interactions

PAH in CHD

Definition
PAH: mean pulmonary artery pressure (PAP) >25 mm Hg at rest or >30 mm Hg during exercise, with a left atrial pressure <15 mm Hg and a PVR <3 Wood units.

- Common (15% of the CHD population is thought to develop PVD; including patients whose PVR exceeds systemic vascular resistance with reversed shunting (Eisenmenger syndrome).
- Pre-operative (usually) increased shunt.
- Post-operative (endothelial dysfunction).

Shunts

- VSD
- ASD
- AVSD
- PDA
- Aorta-pulmonary window

Lt obstructive lesions

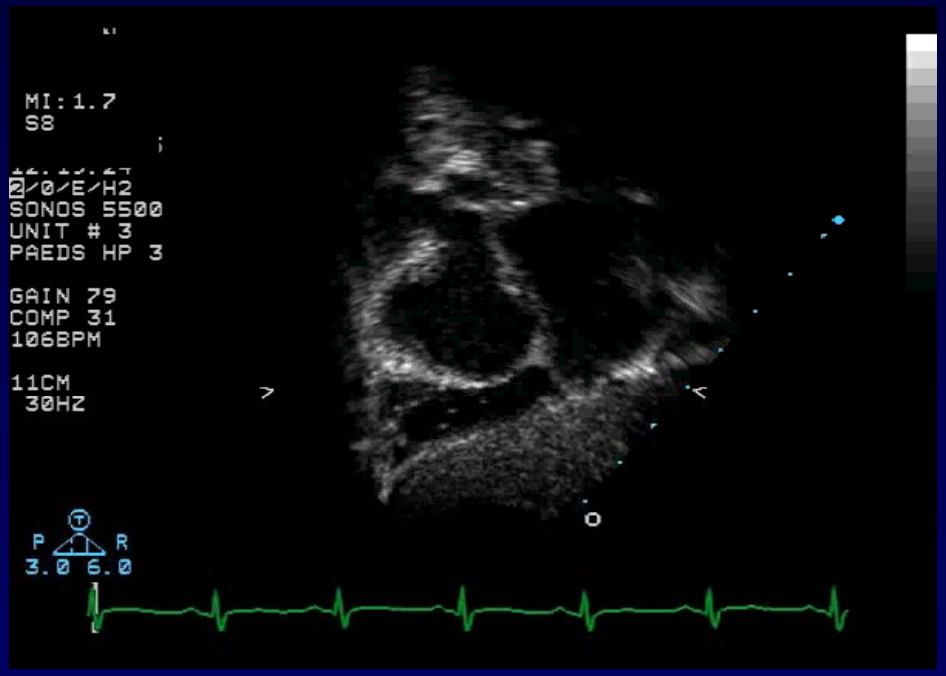
- Pulmonary vein stenosis
- Cor tritriatum
- Supramitral ring
- Mitral stenosis
- Shones
- Aortic coarctation

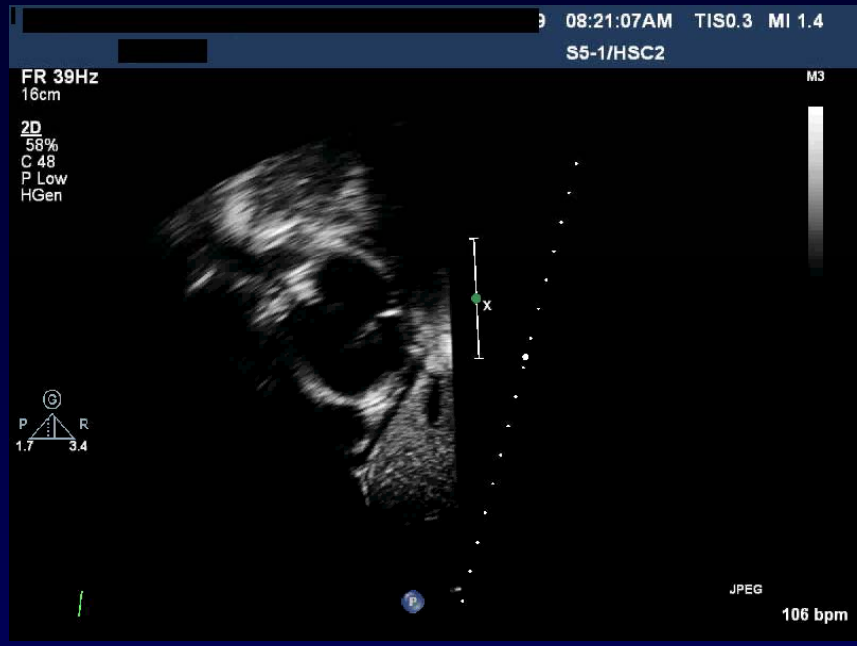
Pulmonary venous hypertension

Clinical questions to be answered by echo:

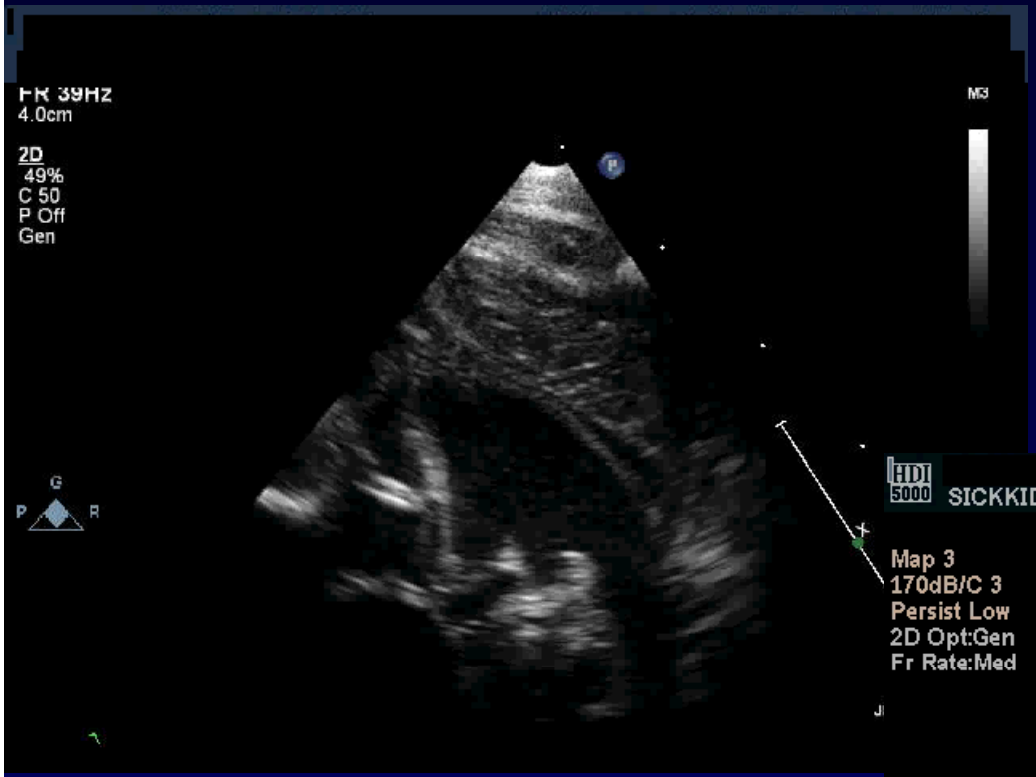
- Severity of pulmonary hypertension
  - Pulmonary artery systolic and diastolic pressures
- Underlying lesions / shunting across intra-cardiac communication
- Associated lesions
- Right and left ventricular function
- Right ventricular hypertrophy
- Tricuspid regurgitation

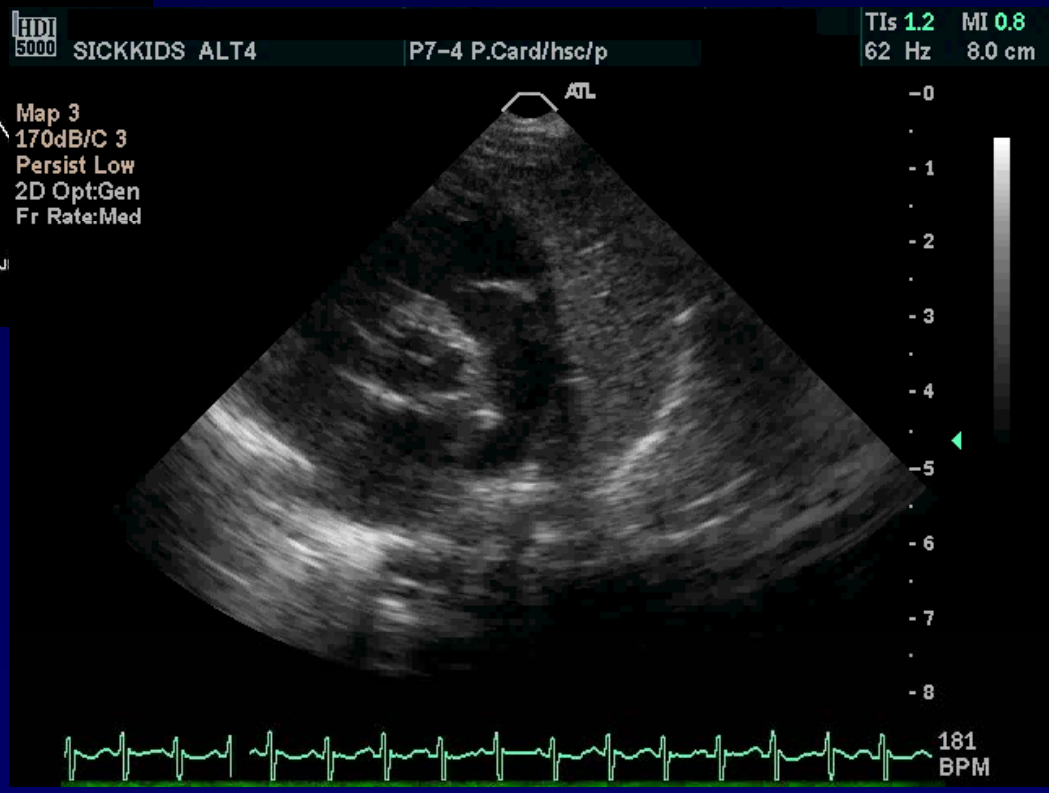
Shunt lesions



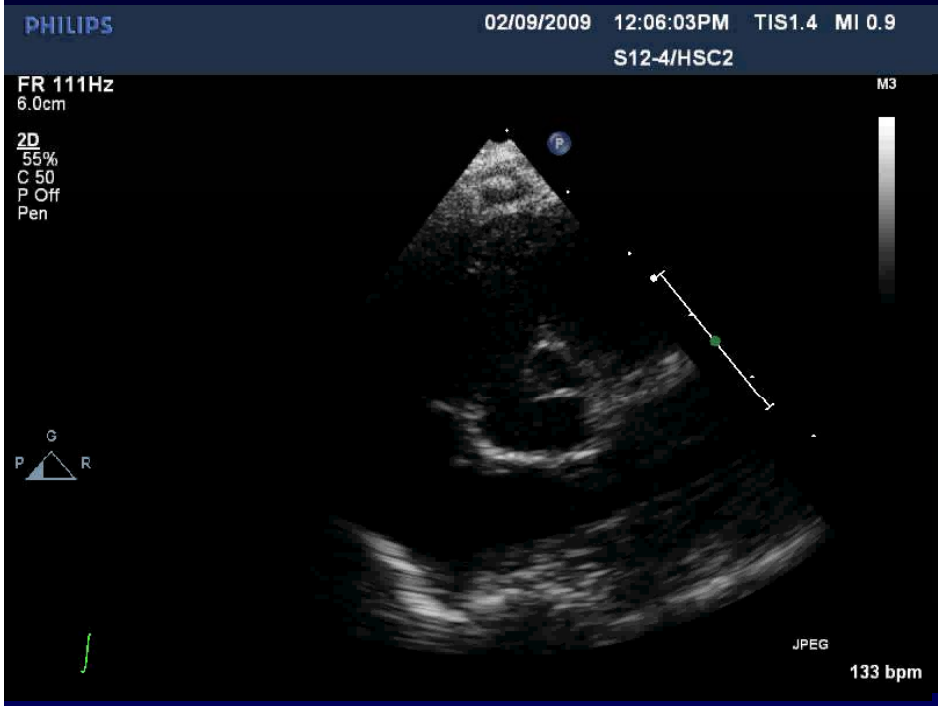


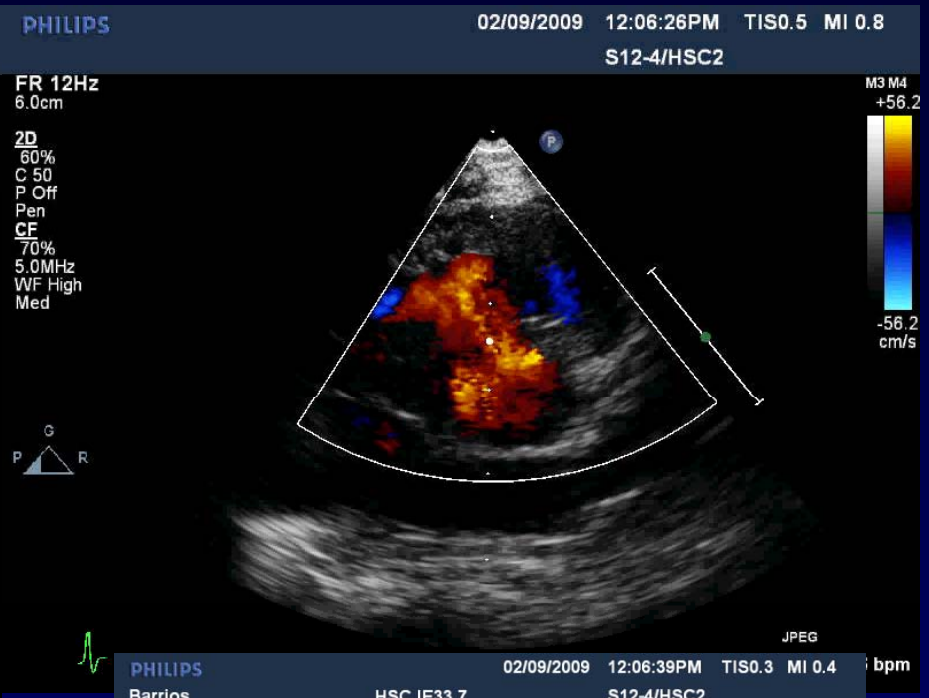
Shunt lesions

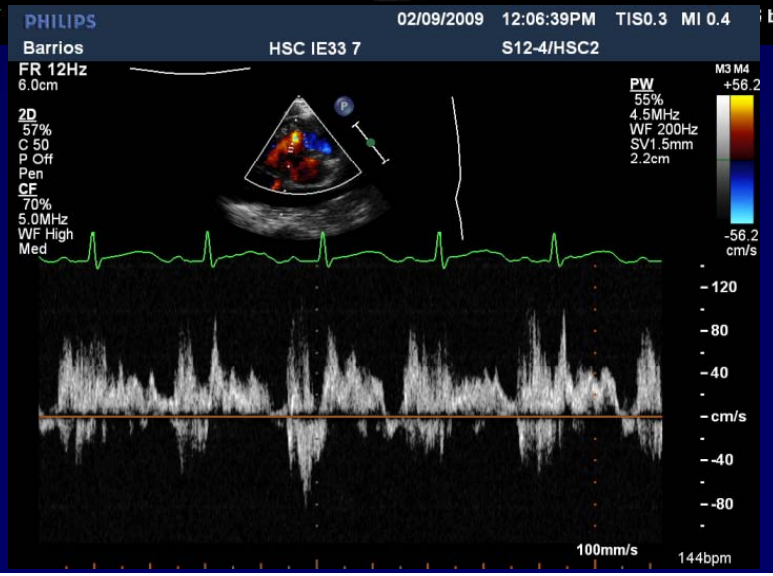




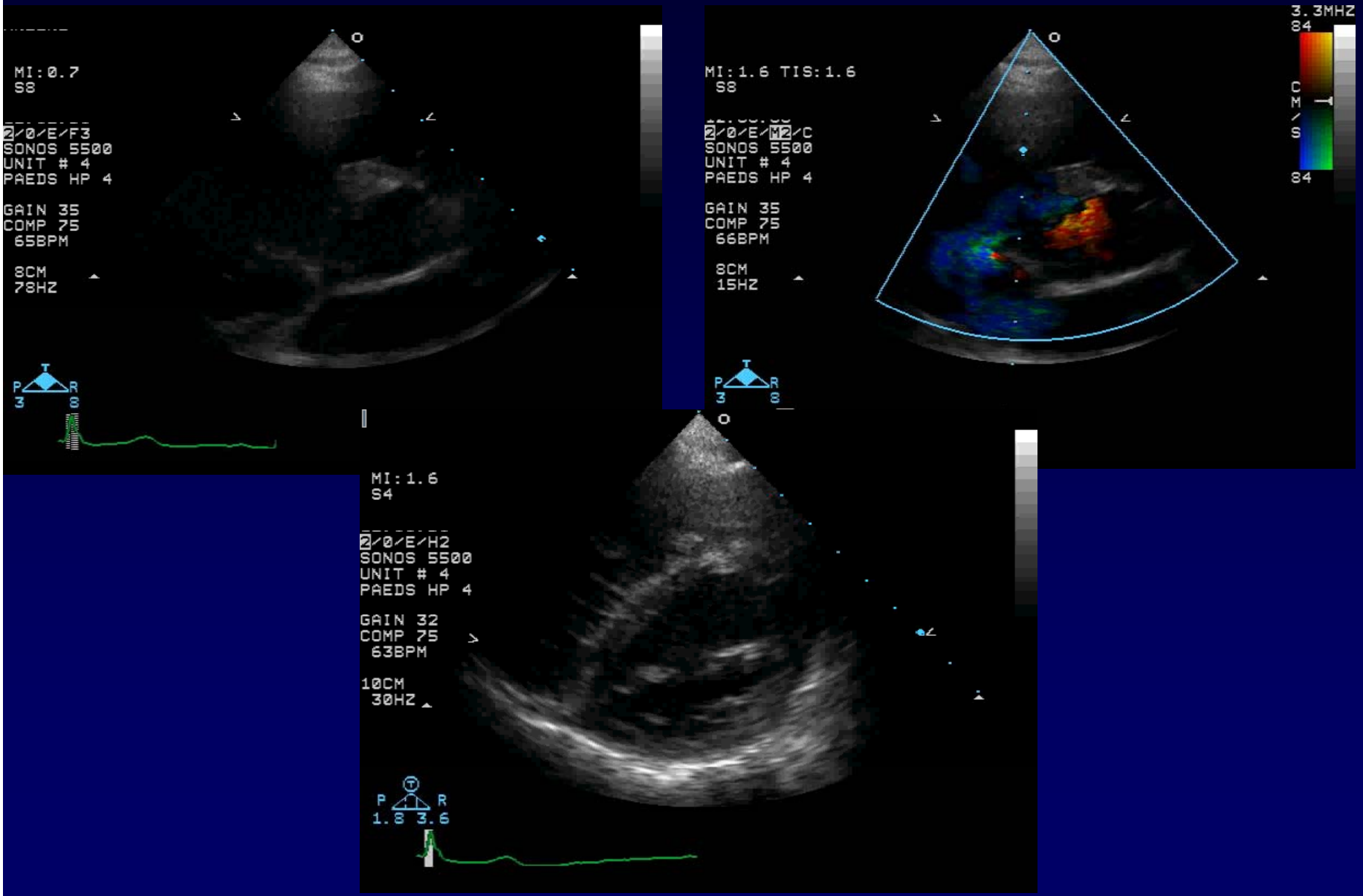
Perimembranous VSD







Perimembranous VSD

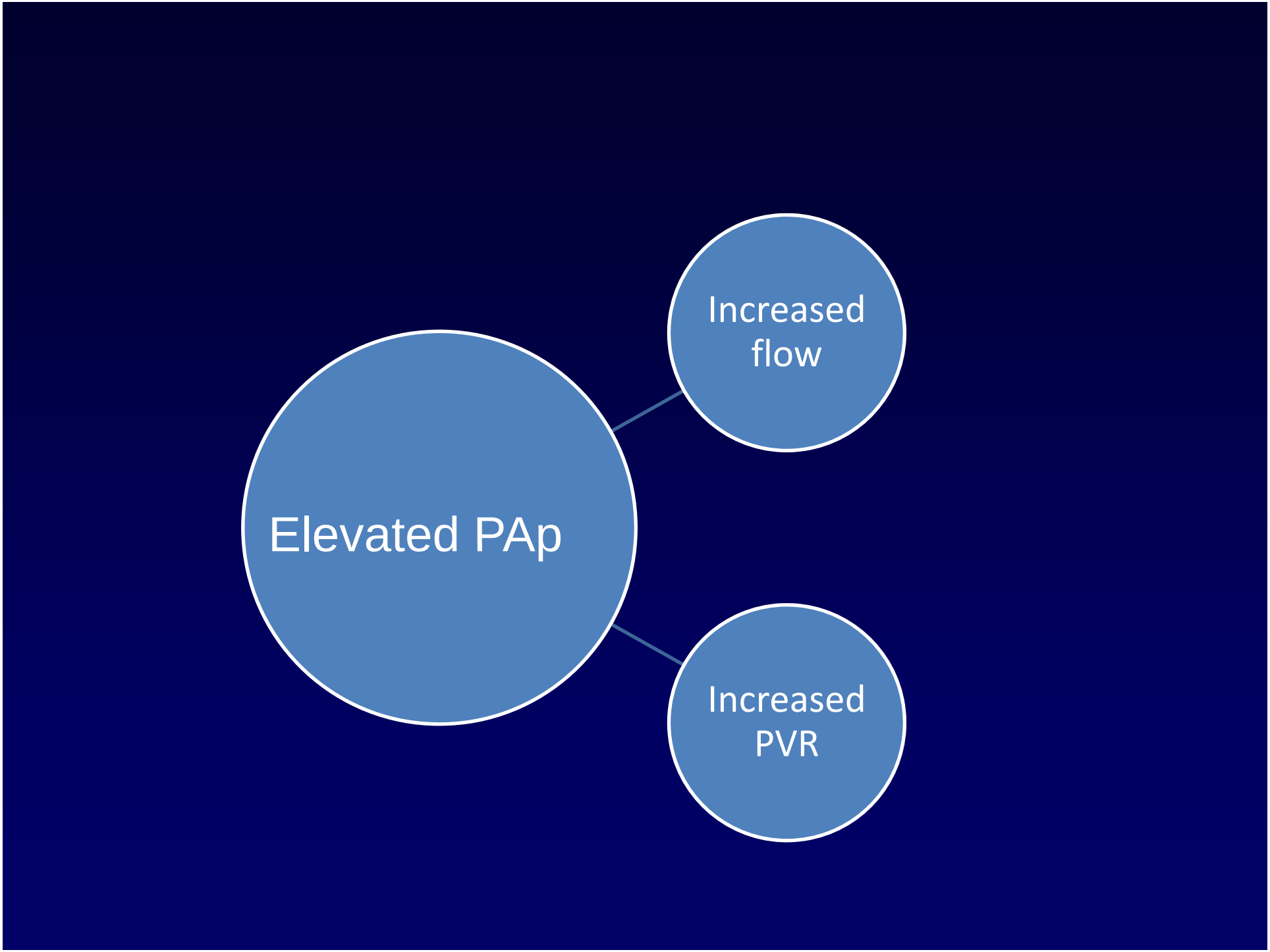


Eisenmenger's syndrome

- Irreversible pulmonary vascular disease from a systemic-to-pulmonary communication (ASD, non-restrictive VSD, non-restrictive PDA, atrioventricular septal defect, aortopulmonary window, surgical systemic-to-pulmonary shunt).
- An initial L > R shunt reverses direction following an increase in PVR.

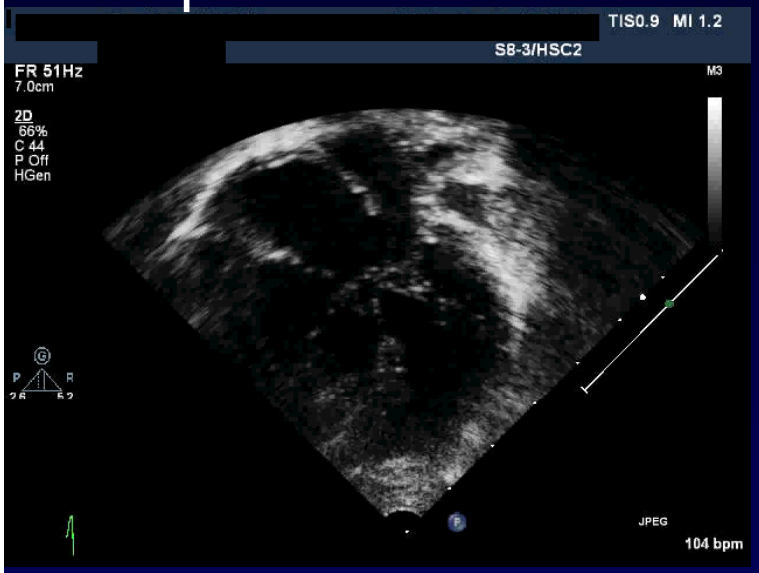
Evaluating worsening in Eisenmenger

- Signs of impending RV failure:
  - RV enlargement and dysfunction.
  - Septal flattening in diastole as well as systole.
  - Worsening tricuspid regurgitation
    - increasing afterload
    - annular dilatation
    - RV dysfunction
  - Right atrial enlargement
    - TR
    - impaired RV compliance.

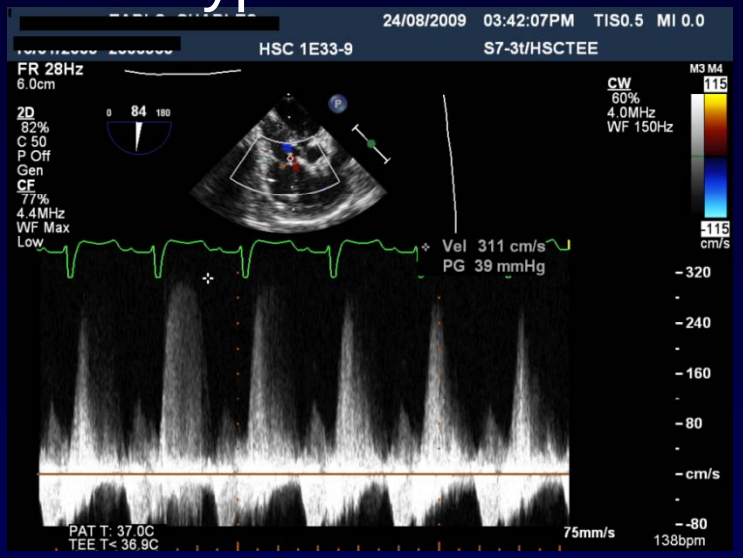

Elevated PAp
Increased flow
Increased PVR

5 months, AVSD

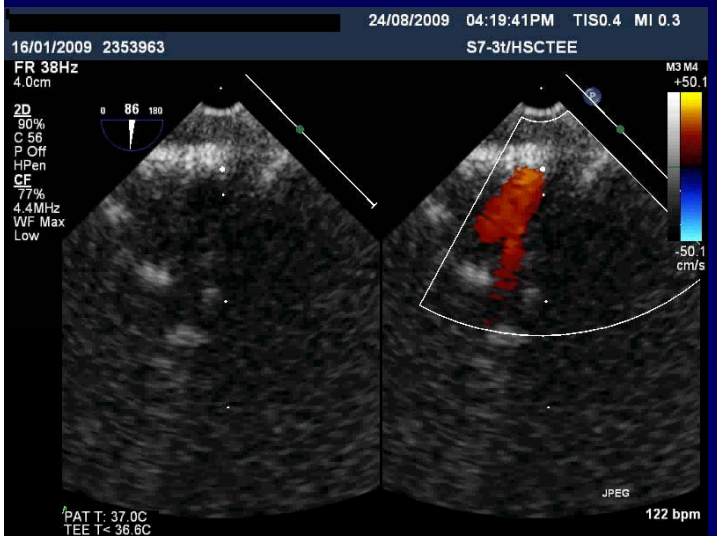
Pre-op



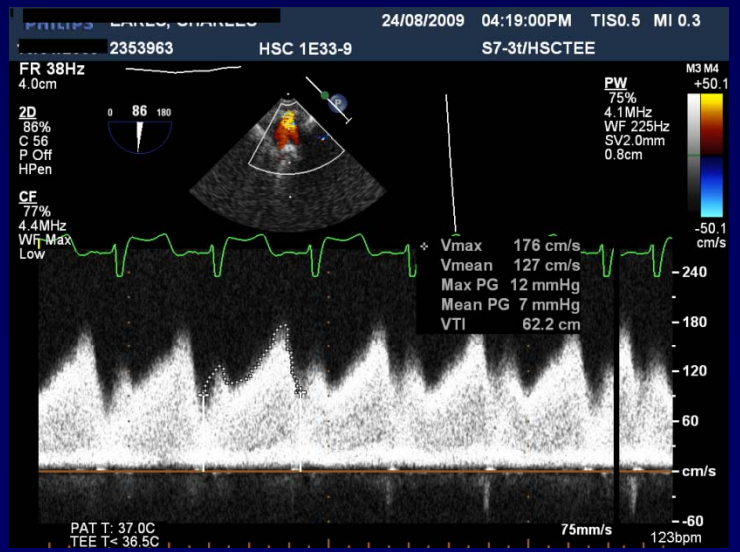
Post bypass TEE



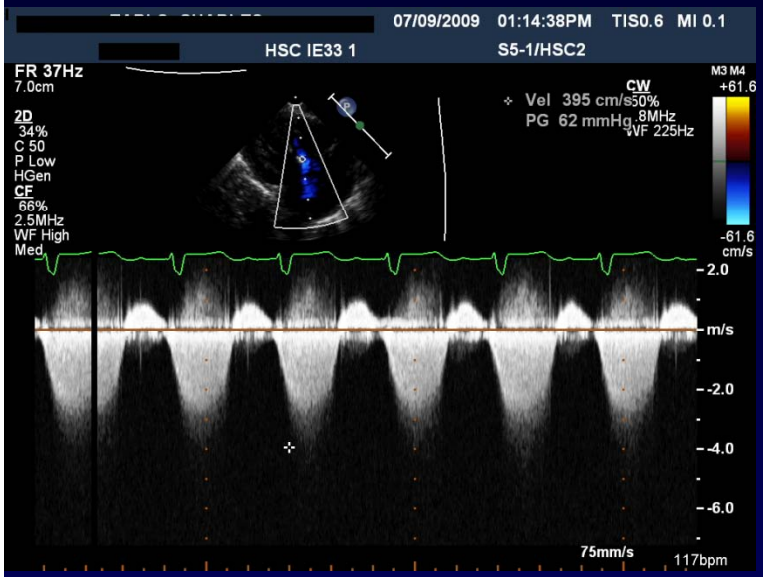
Post bypass TEE

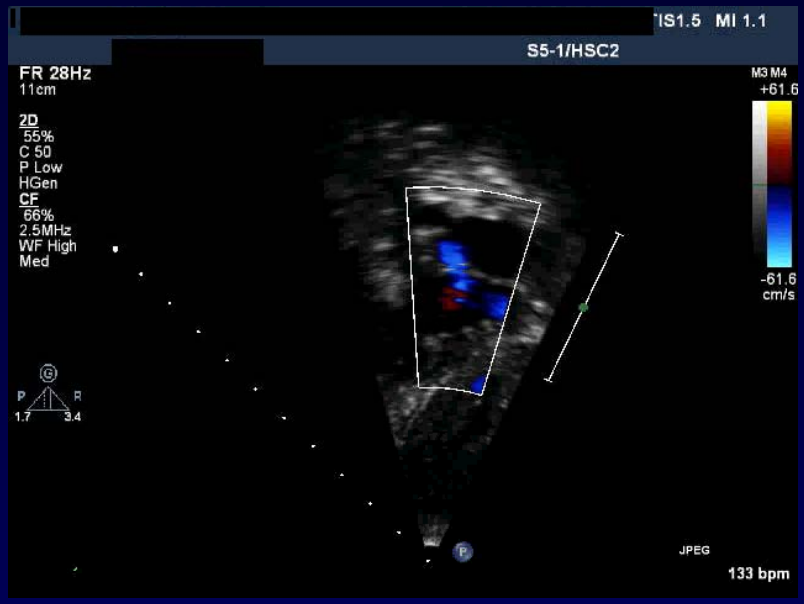


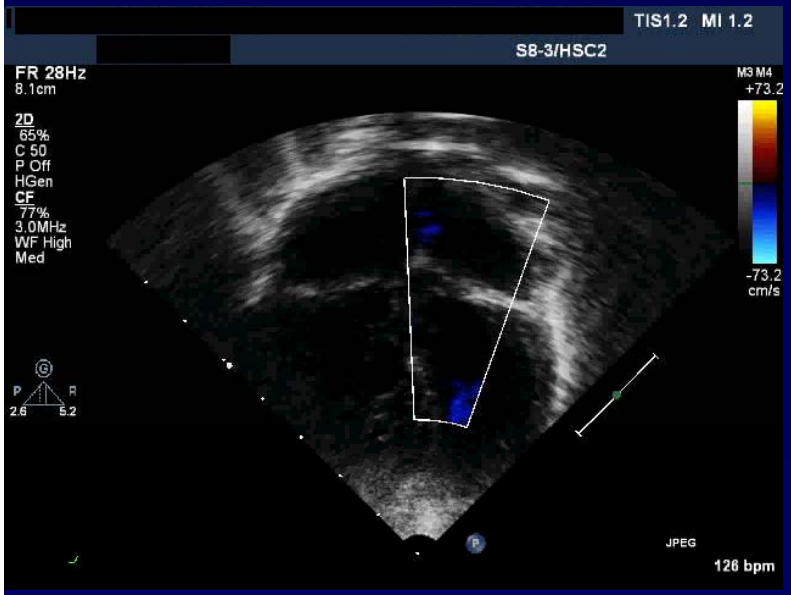
Post bypass TEE

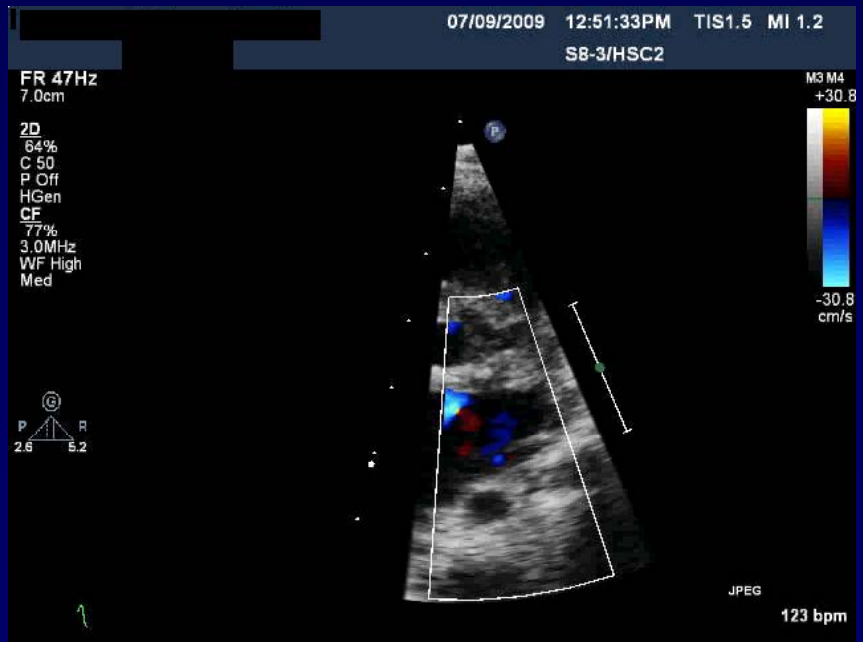


Infant, AVSD, post-op cant wean from ventilator

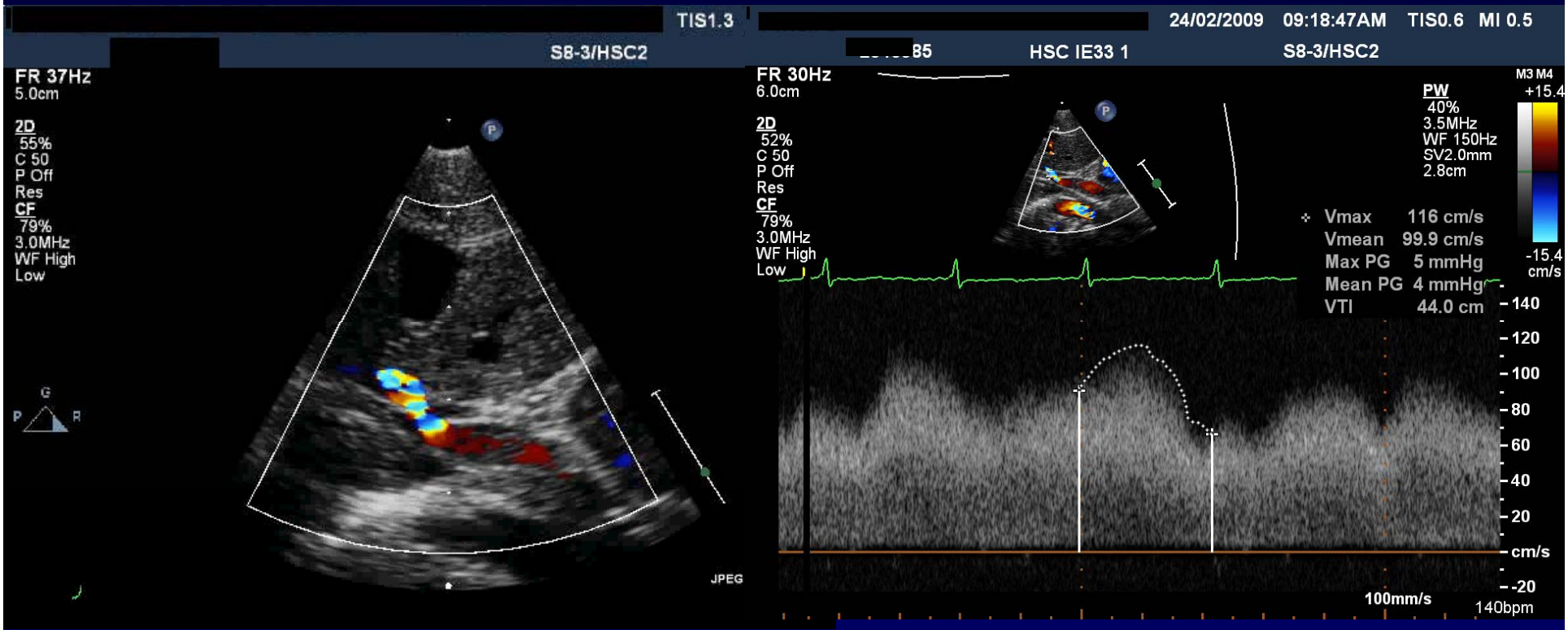




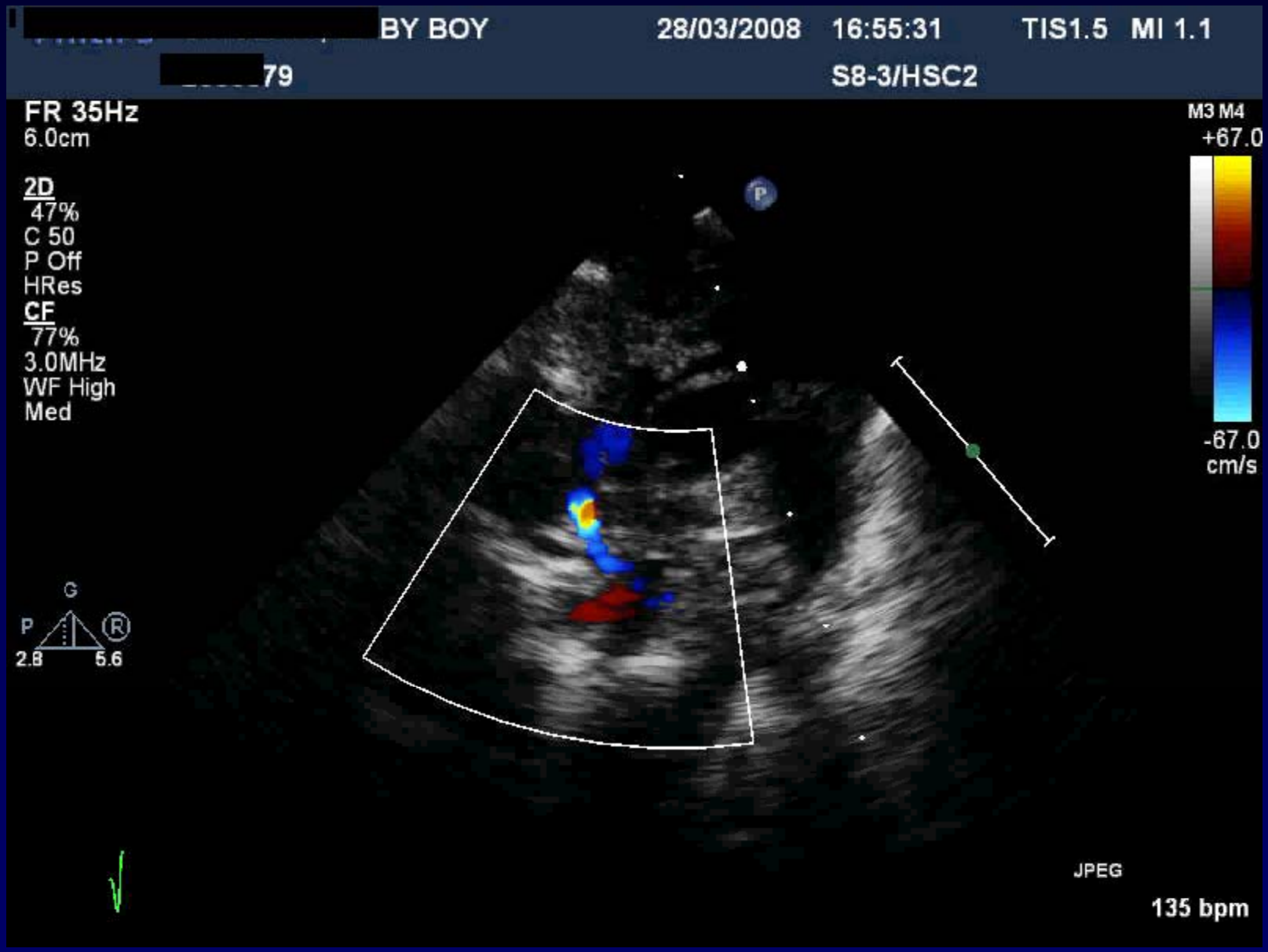




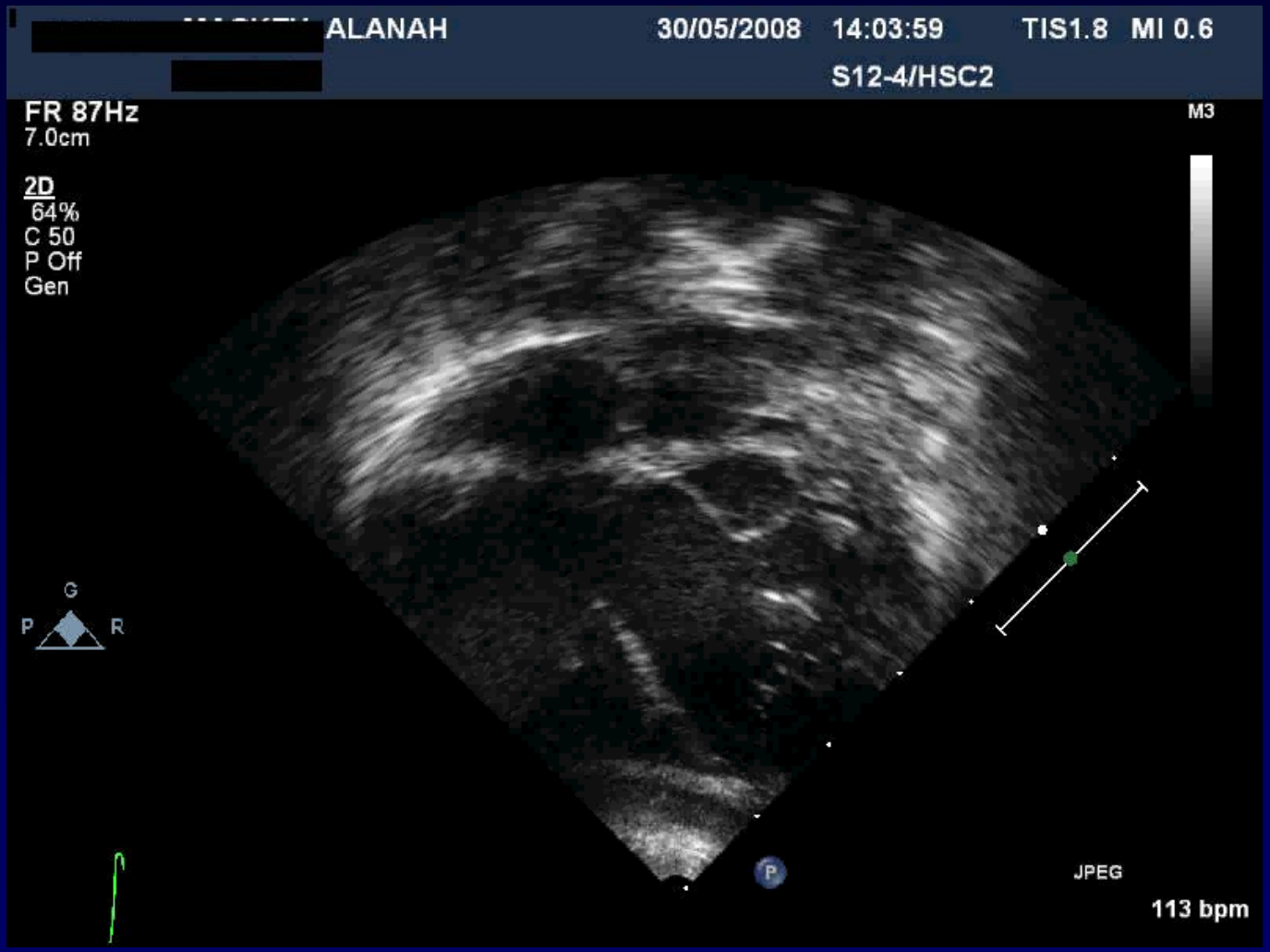
Ill neonate, pre-ECMO echo



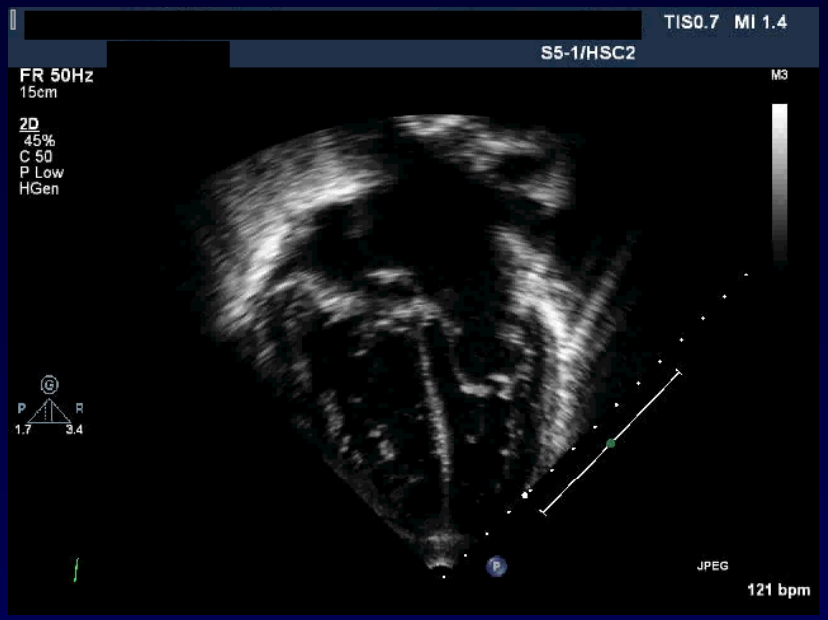
Pulmonary vein stenosis

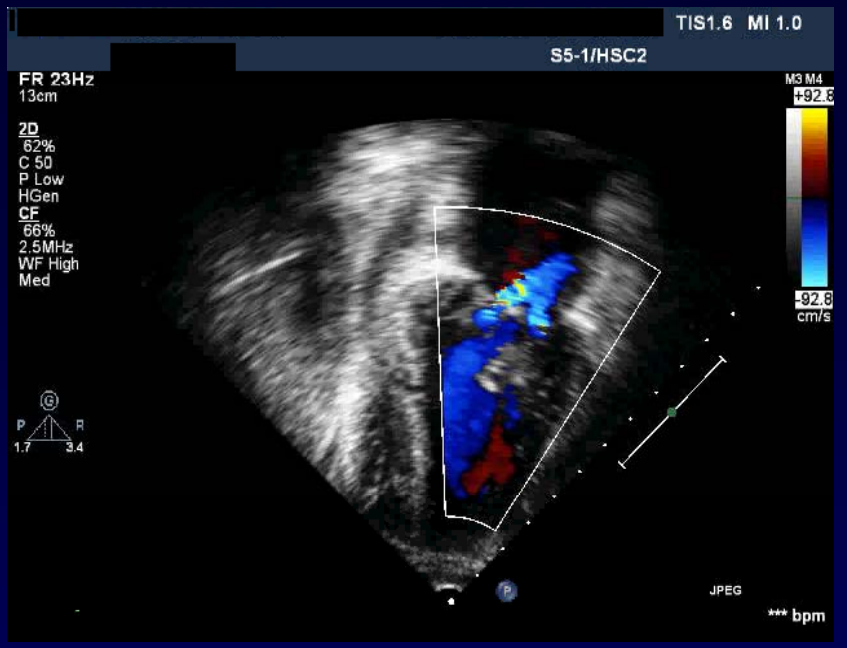


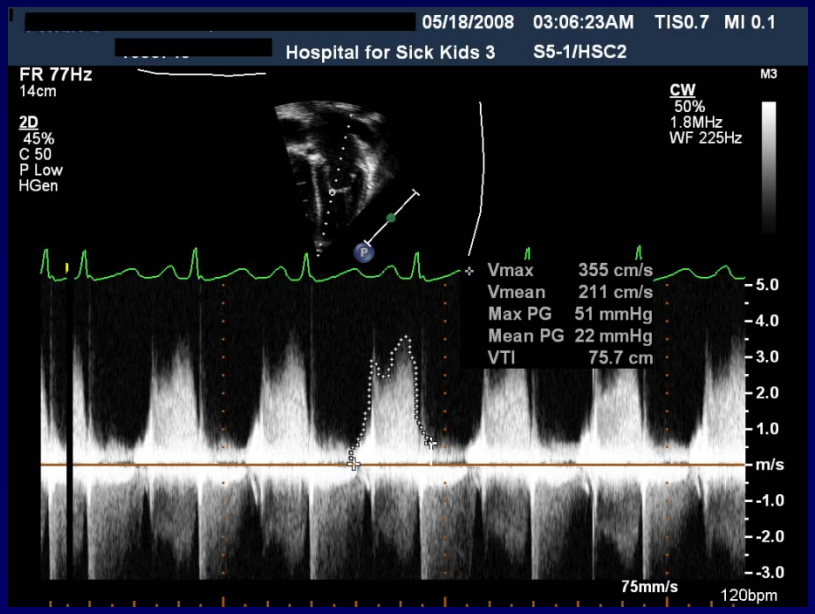
Cor tritriatum

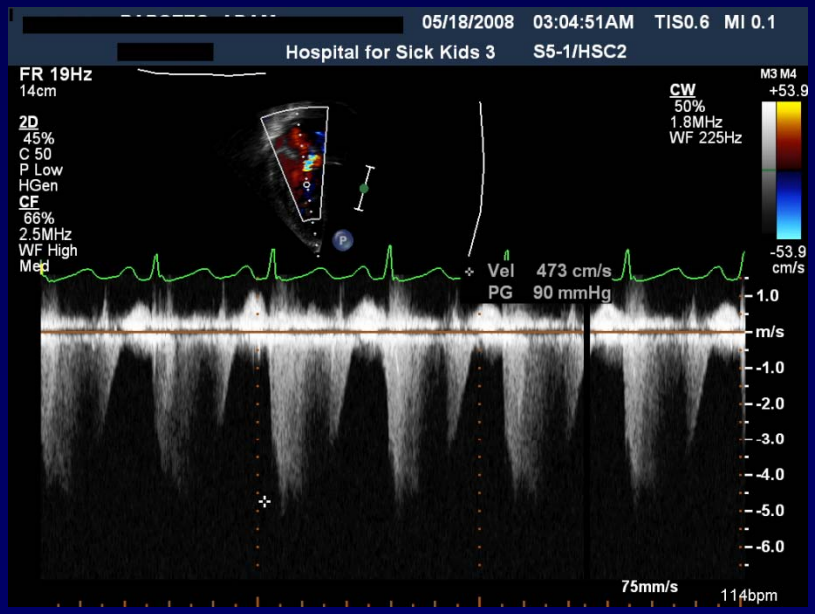


9 months, tachypnea

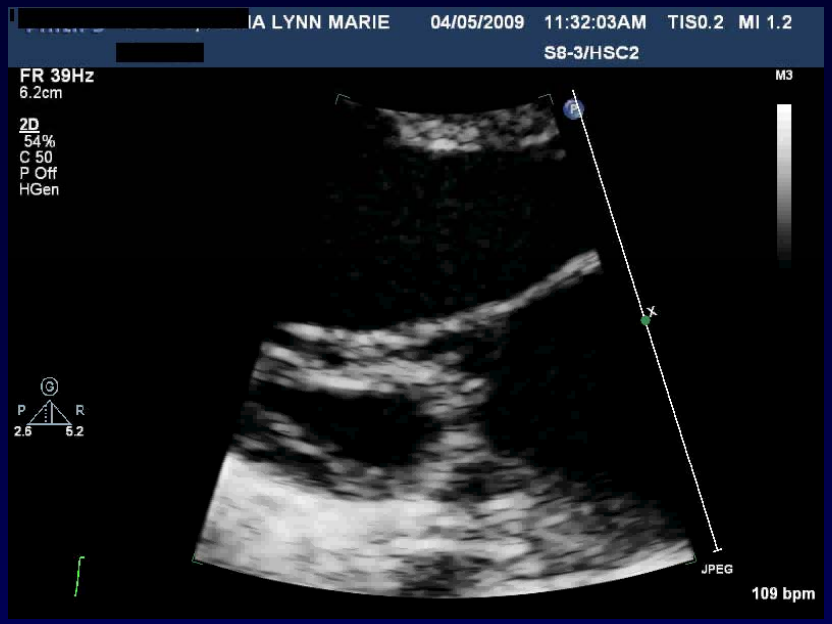


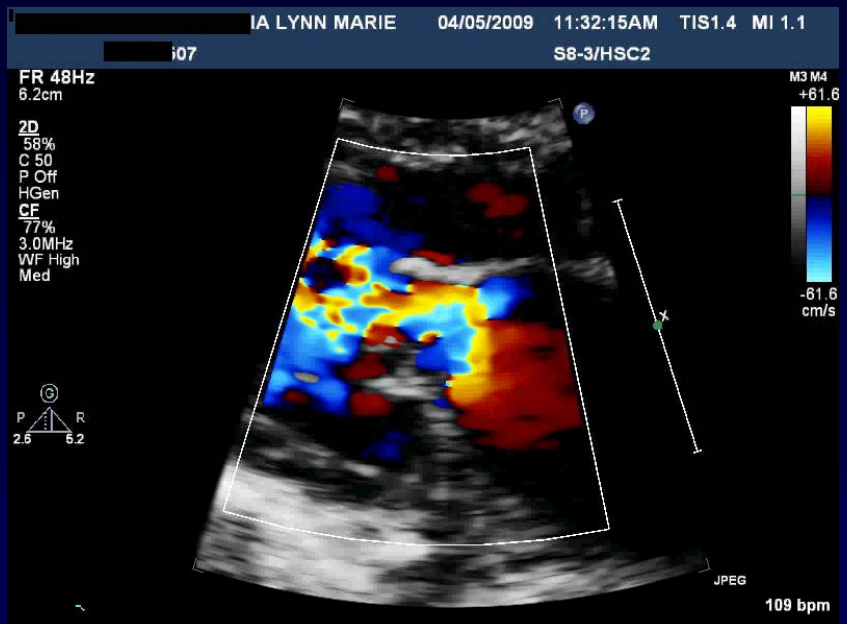


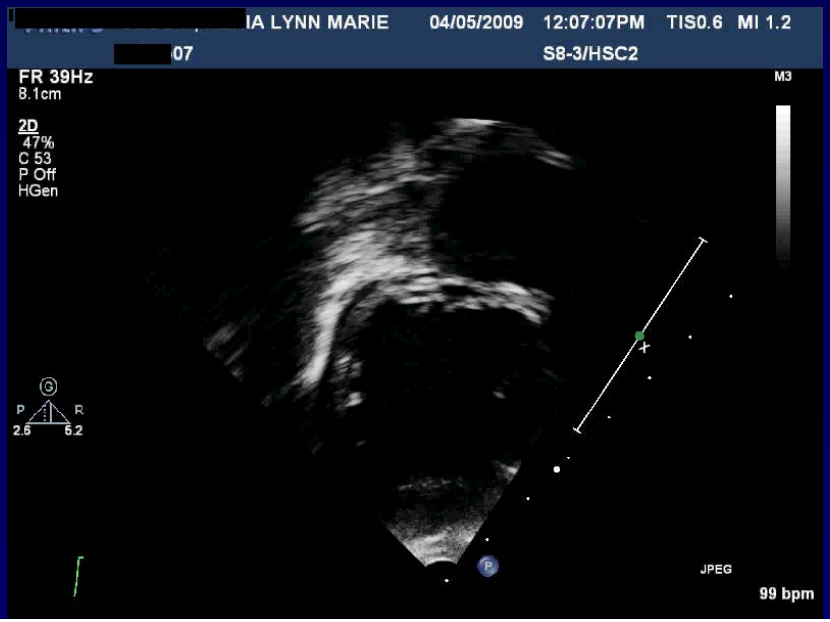


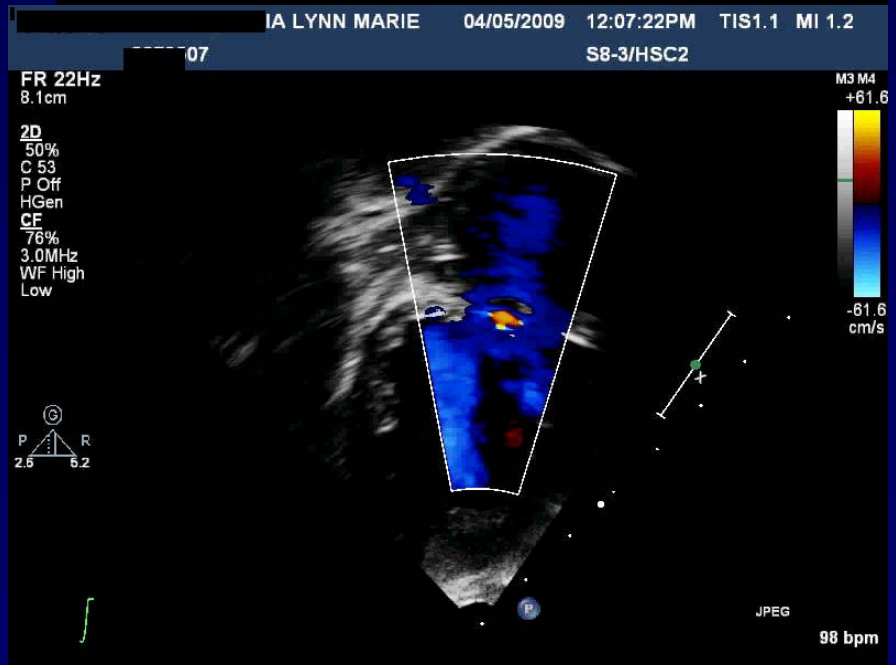


Supra-mitral ring

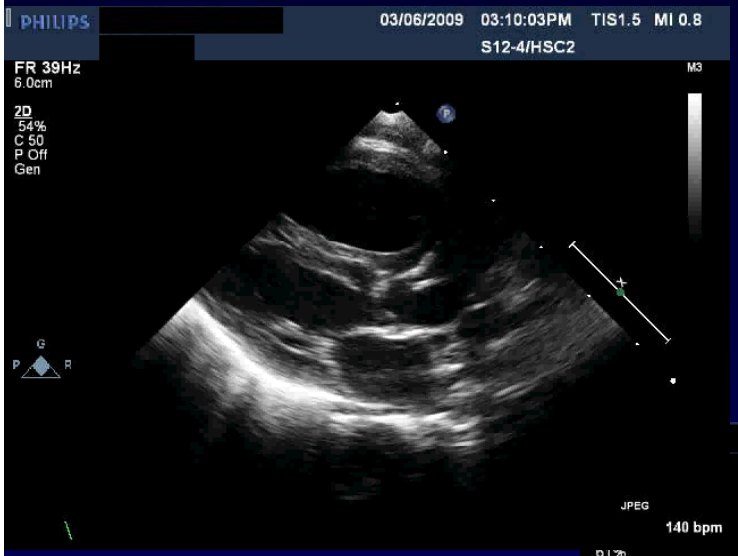


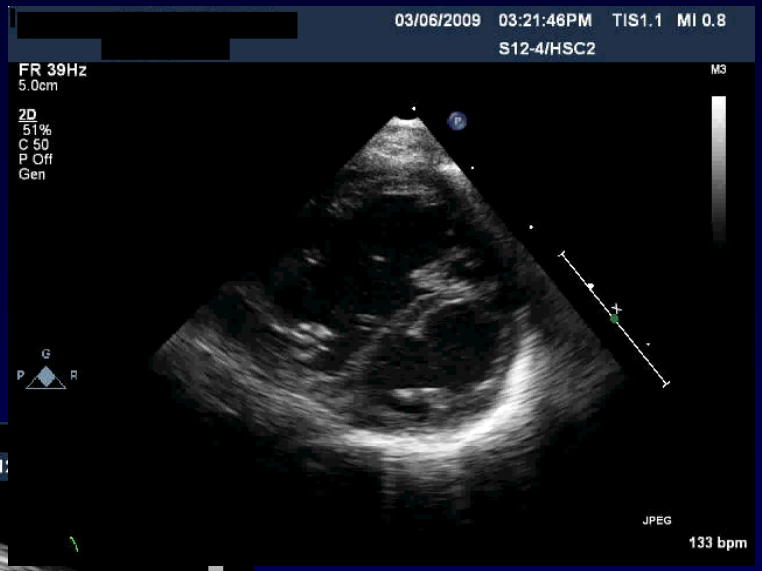


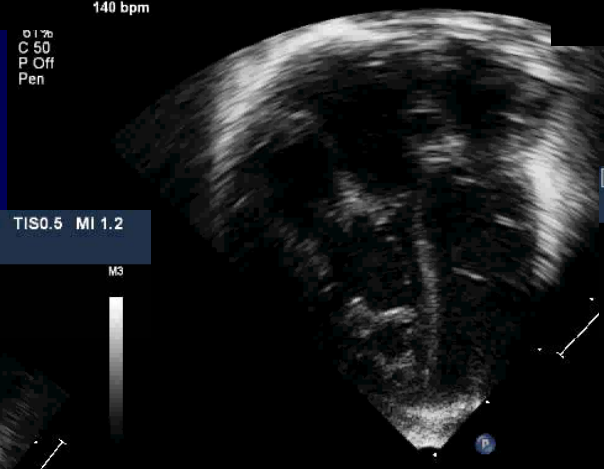


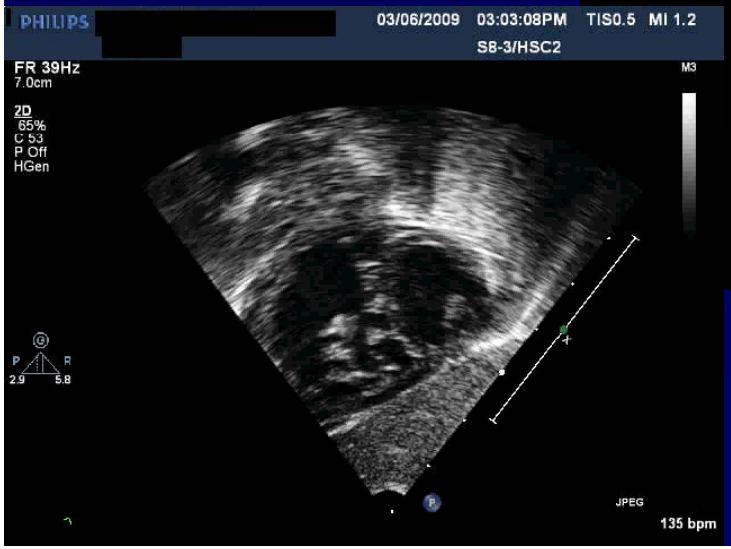


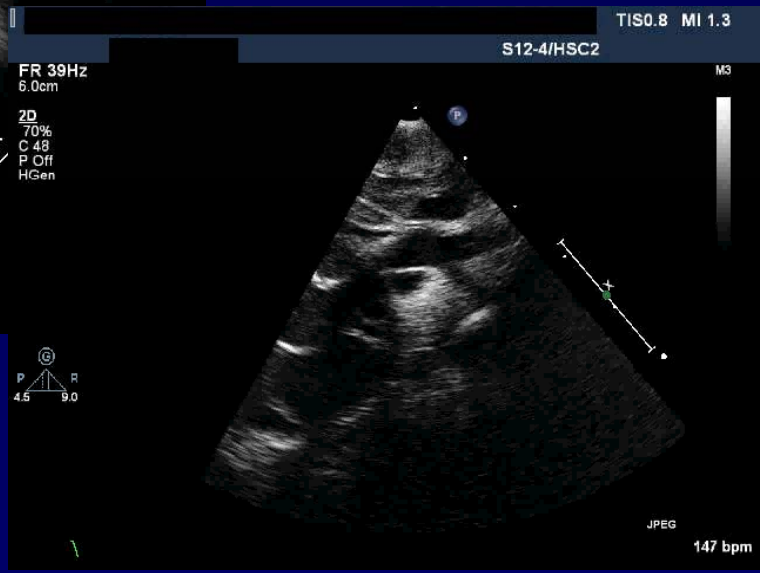
Infant, Failure to thrive



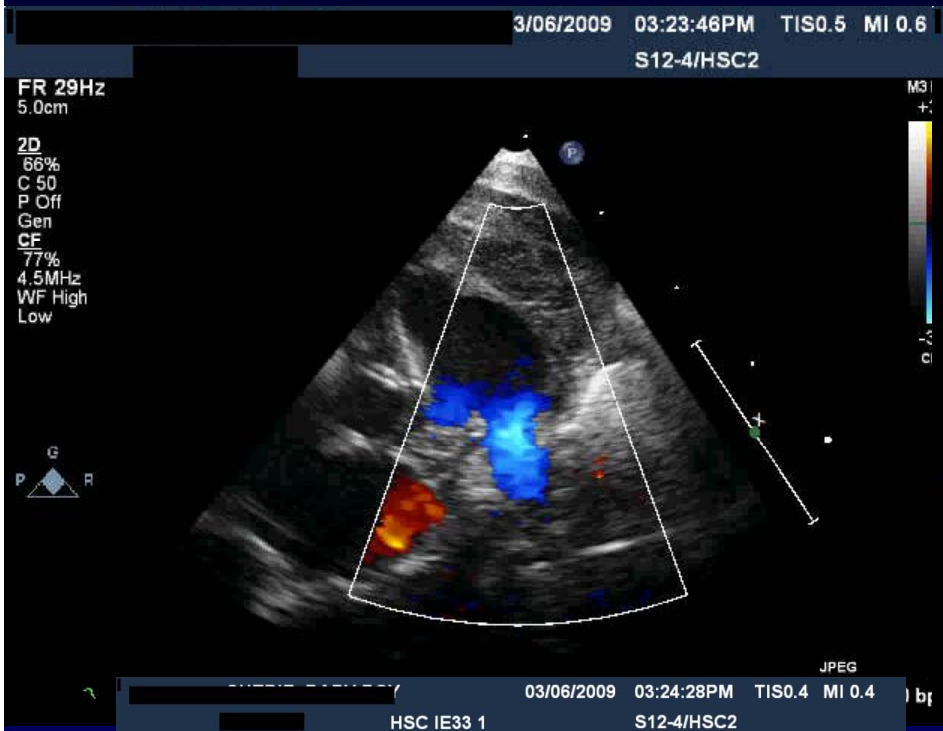


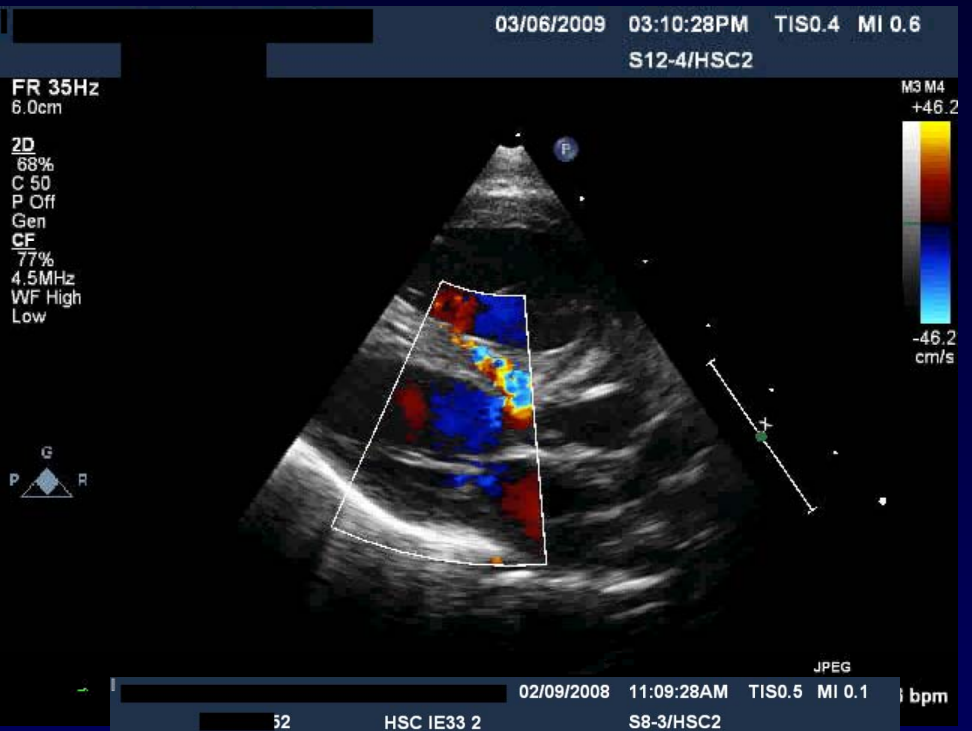


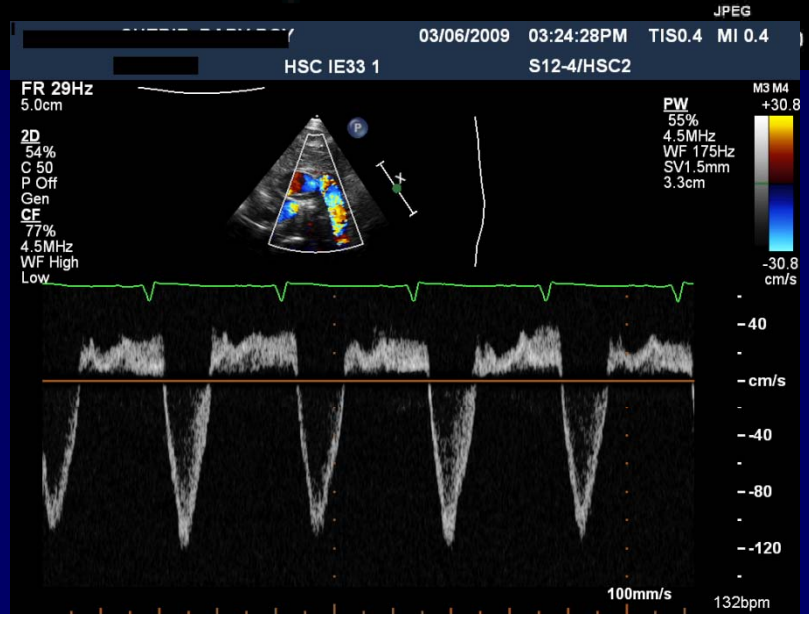


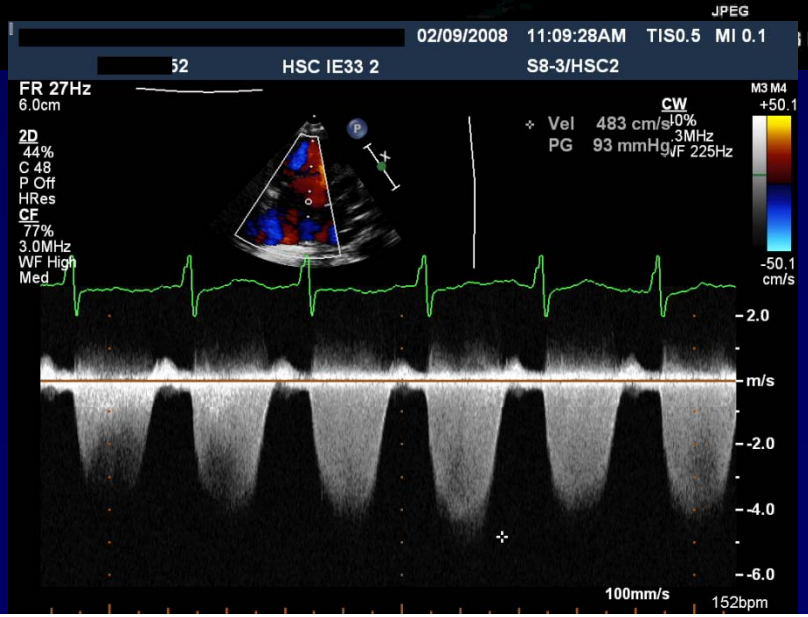


Infant, Failure to thrive

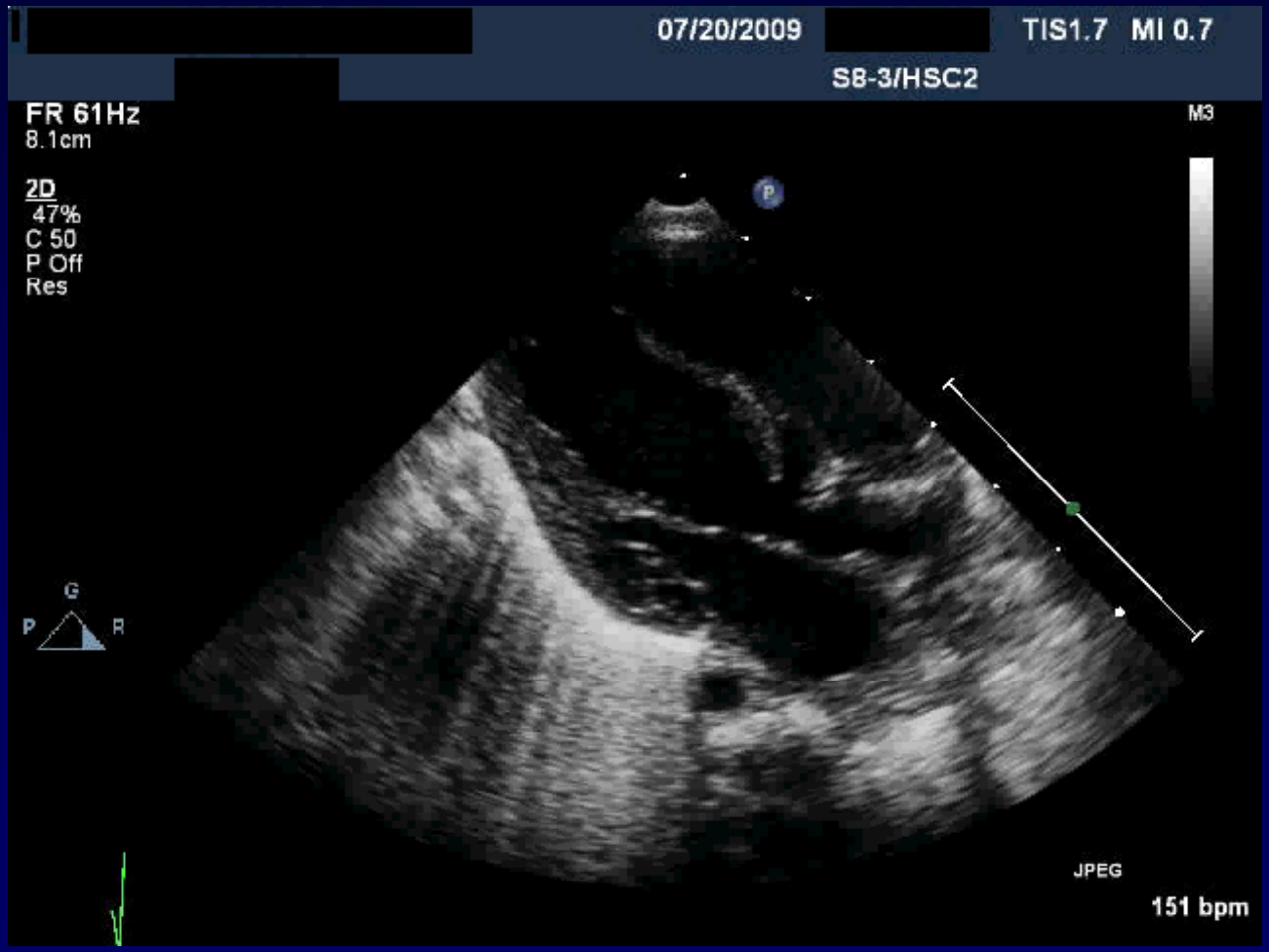




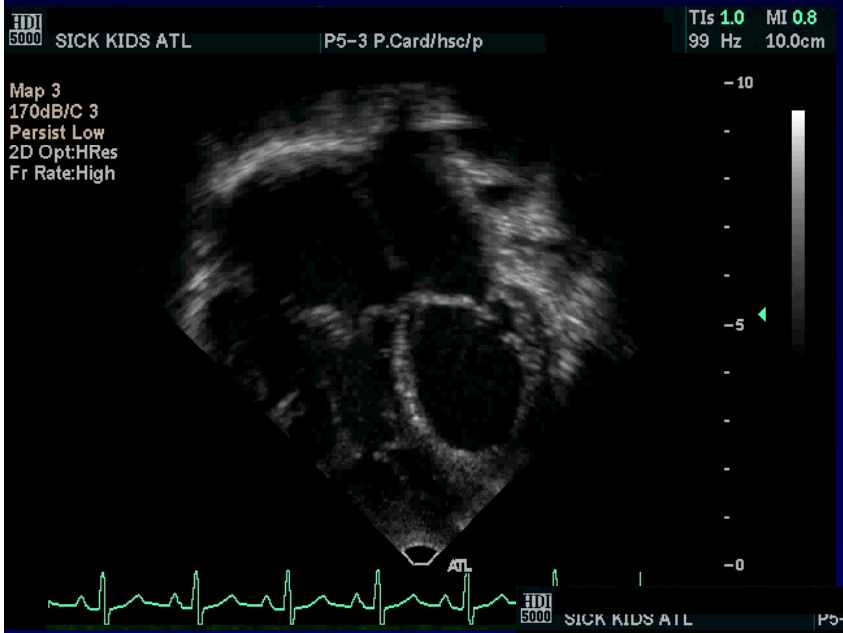


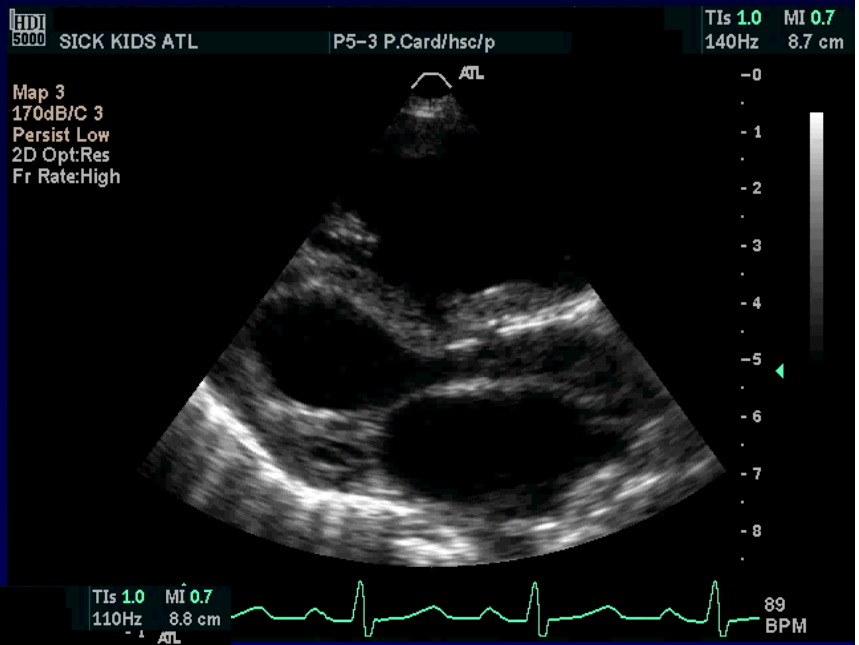


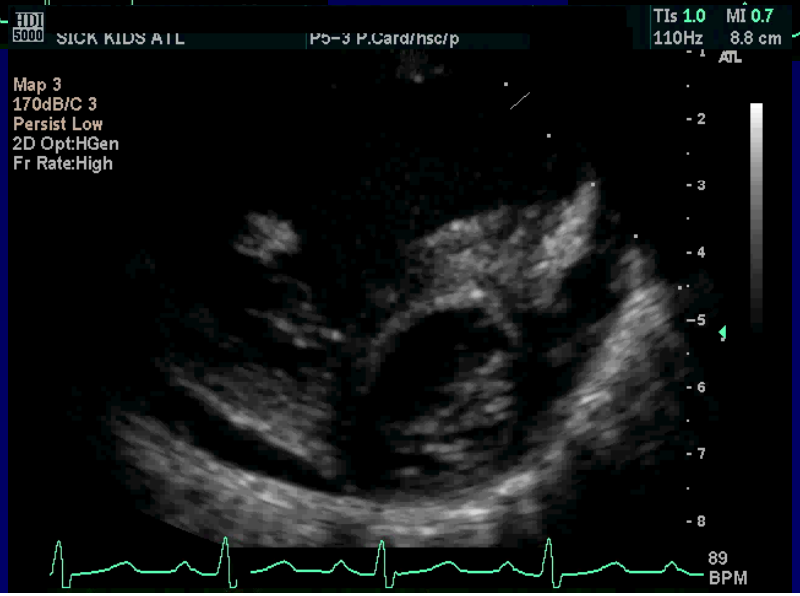
At Follow-up


07/20/2009
TIS1.7 MI 0.7
S8-3/HSC2
FR 61Hz
8.1cm
2D
47%
C 50
P Off
Res
M3
G
P
R
JPEG
151 bpm

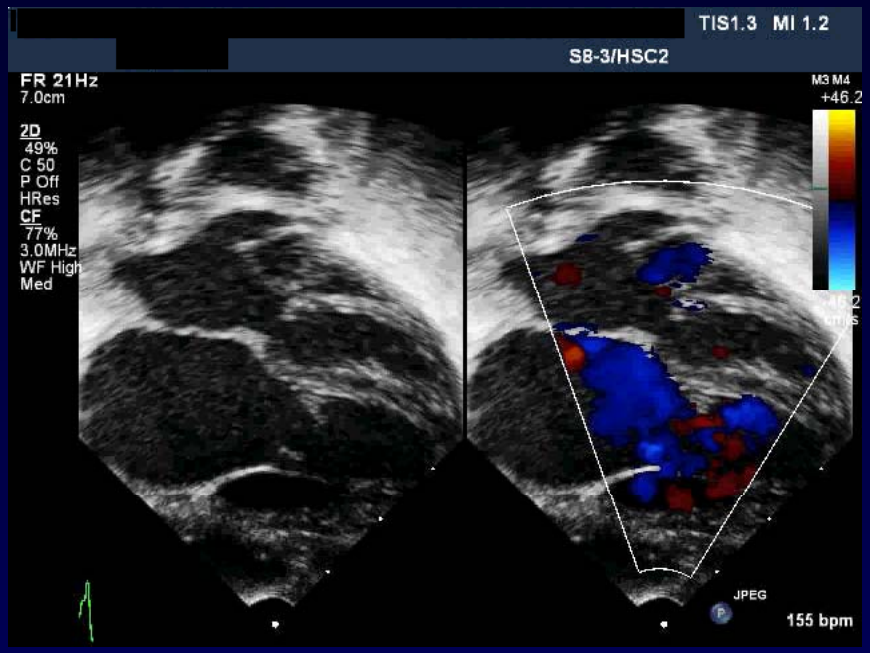
Borderline LV-Irreversible PAH?

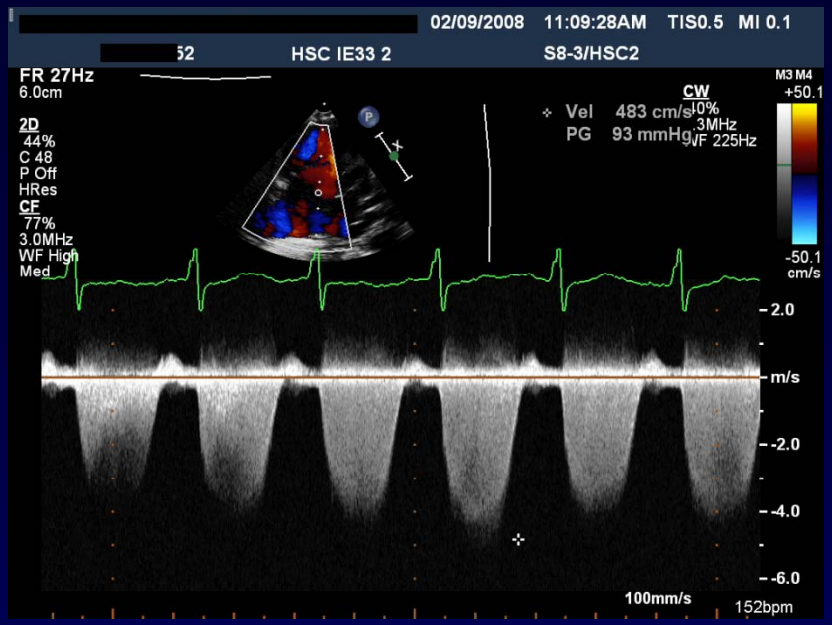


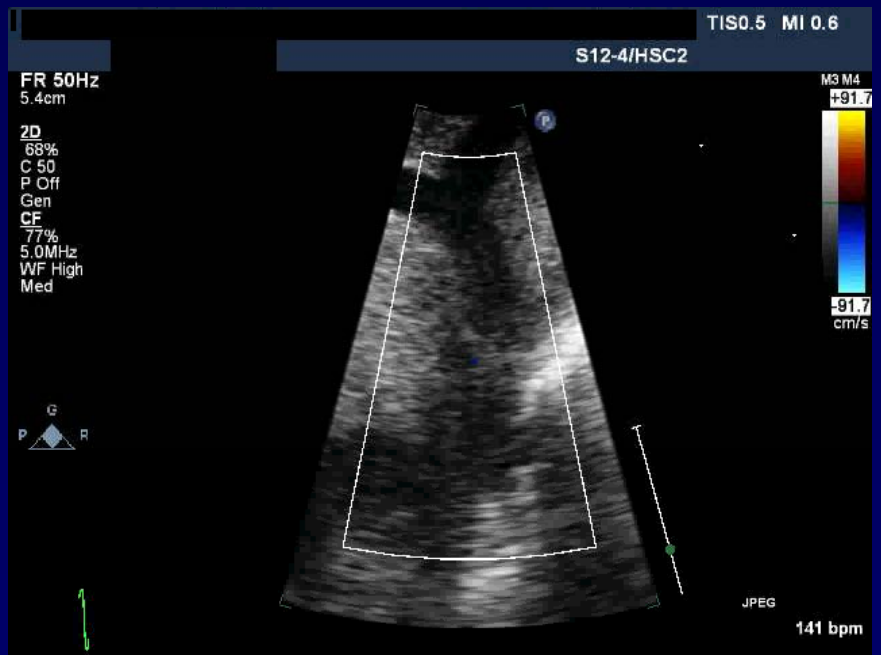




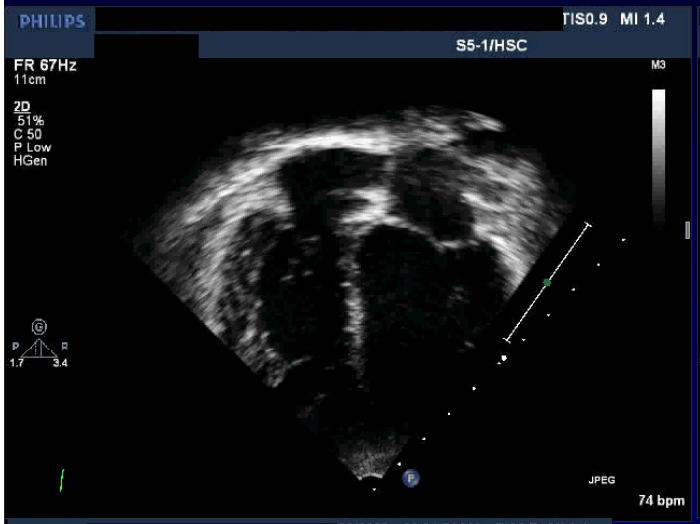
Aortic coarctation

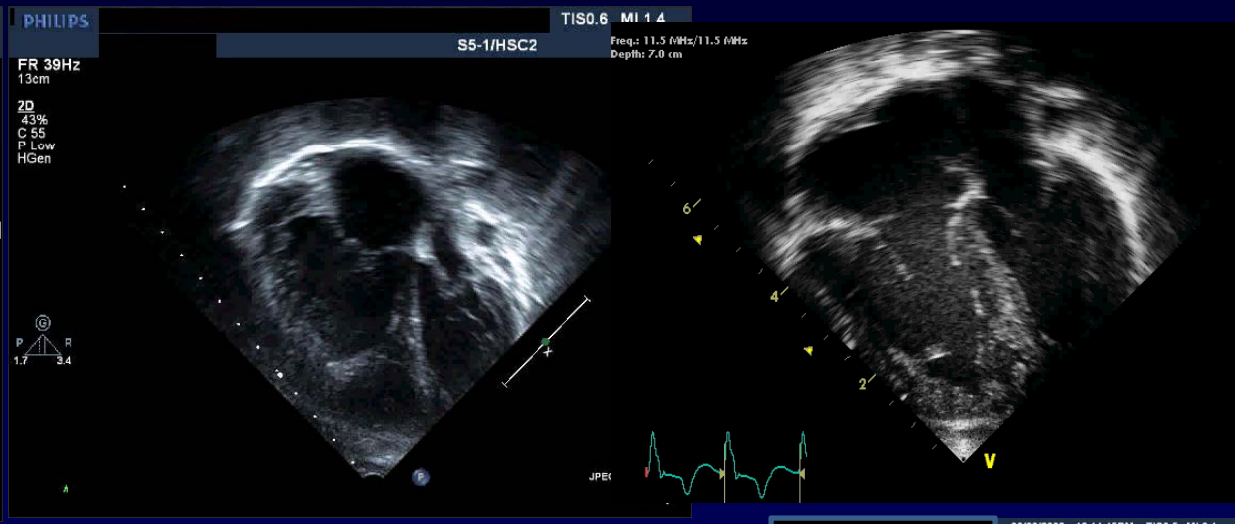


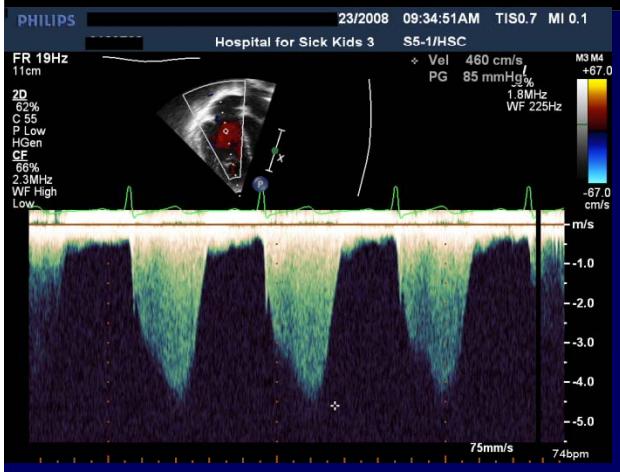


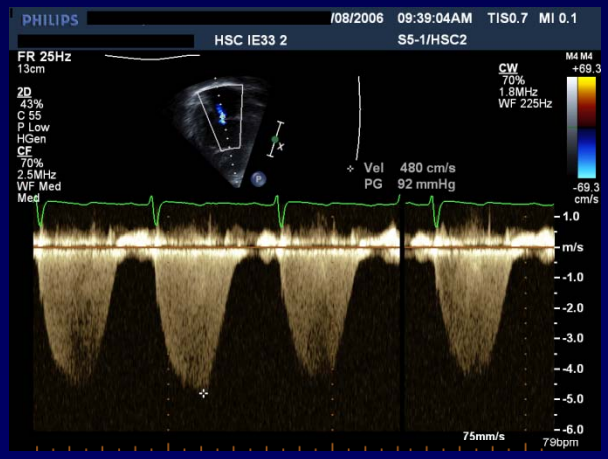


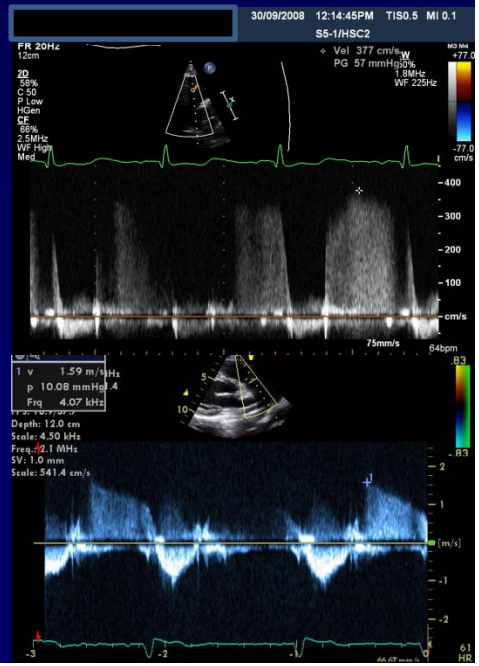
Assessment of RV function



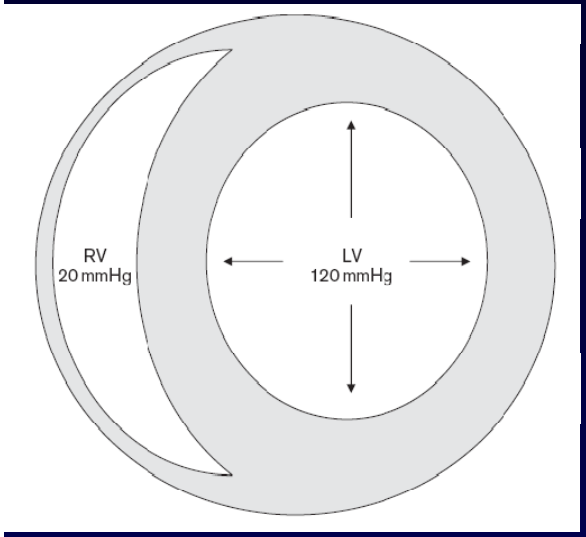


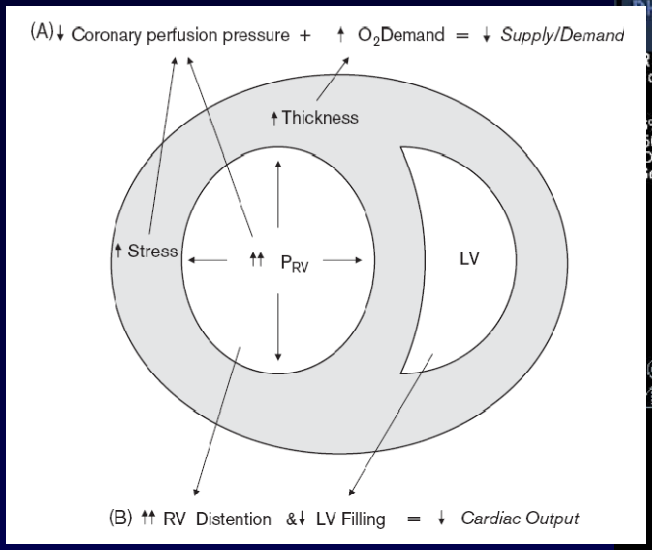


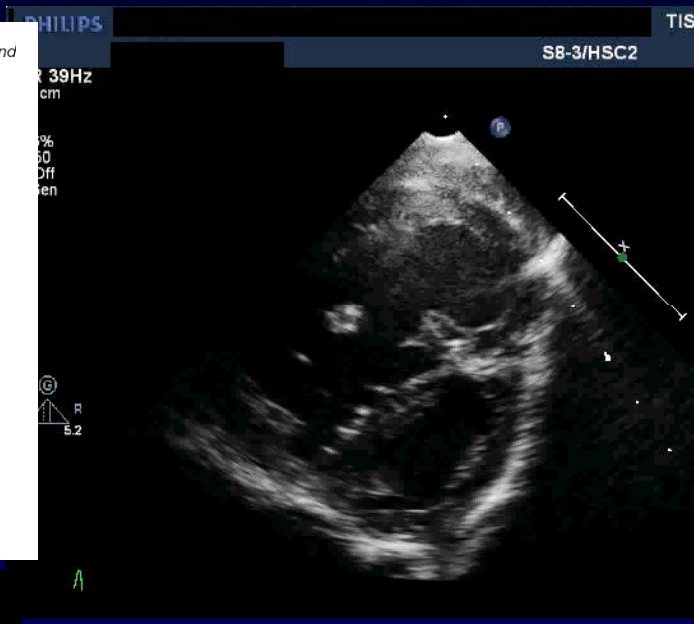


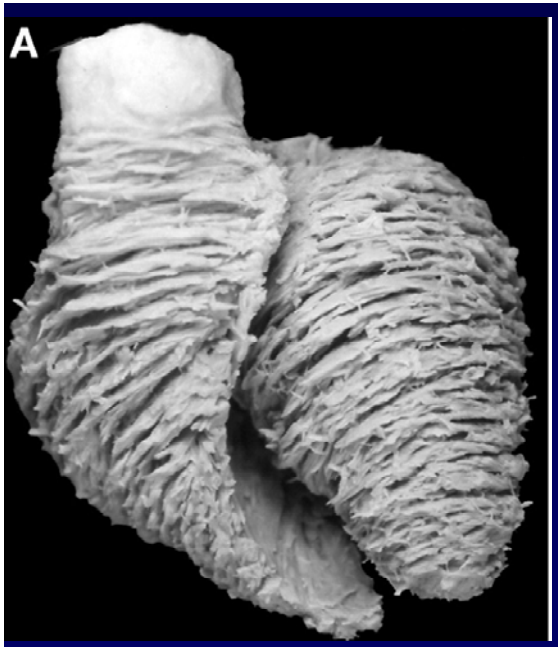


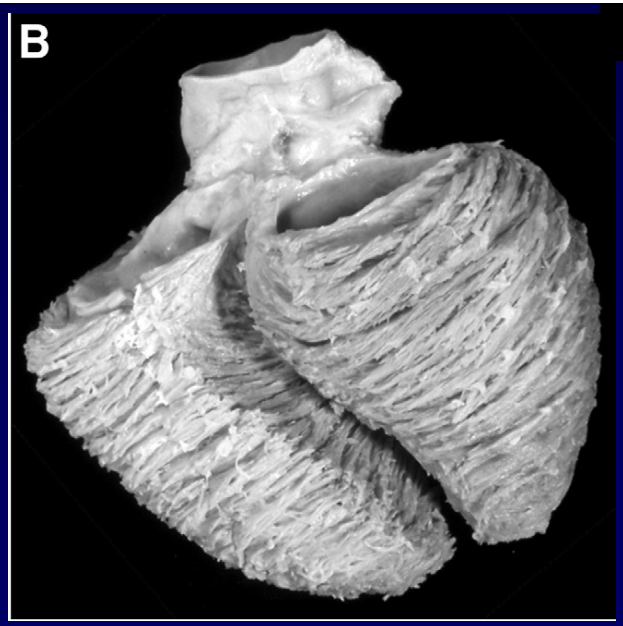
RV structure

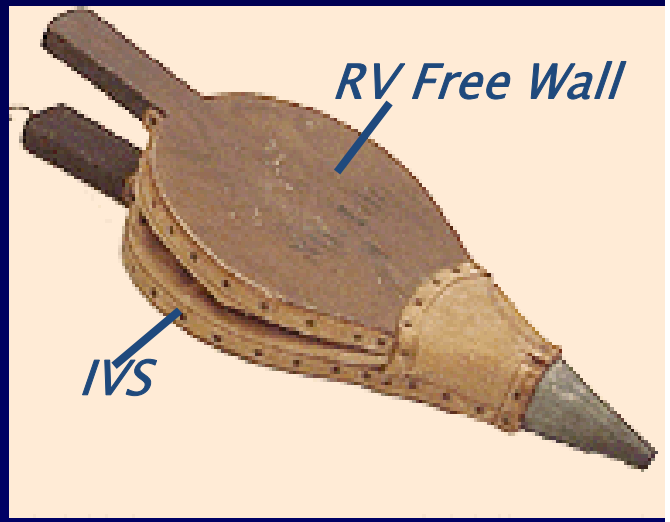








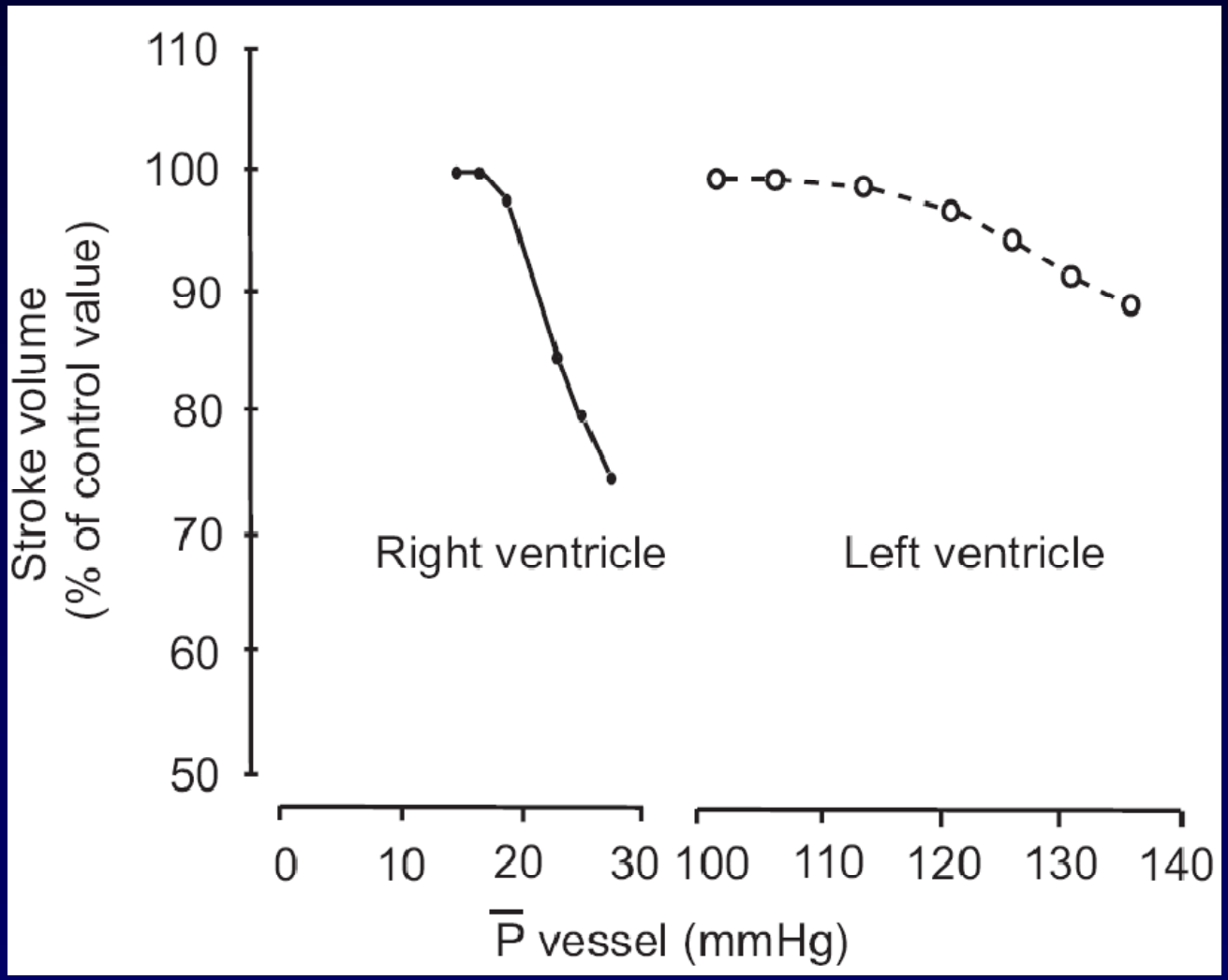




Anderson, Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2007 10:76

Chin, Coronary Artery Disease 2005, 16:13–18

The RV does not handle afterload well



MacNee W. *Am J Respir Crit Care Med*. 1994;150: 833–852.

What should we measure?

Table 1
Echocardiographic measurements used in pulmonary hypertension [2]

Qualitative assessment
Enlarged right atrium and ventricle *
Right ventricle hypertrophy *
D-shaped left ventricular cavity with flattening of the interventricular septum in systole *
Diminished/absent atrial wave of pulmonary valve
Mid systolic closure or notching of pulmonary valve

Hemodynamic assessment
Tricuspid regurgitation velocity *
Pulmonary regurgitation velocity *
Right ventricular outflow tract flow acceleration time
Pulmonary artery systolic flow acceleration time
Right ventricular ejection time
Right ventricular index of myocardial performance
Timing of mid systolic deceleration of right ventricular ejection
Right ventricular long axis function (marker of overall right ventricular systolic function)

* Minimal data set required for echocardiographic evaluation of pulmonary arterial hypertension.

Additional echocardiographic measures described in the literature

RV end-diastolic area index
Right atrial end-systolic area index
RV area change (percentage)
Eccentricity index
Severity of TR/right atrial ratio
Stroke volume
Cardiac index
Pericardial effusion size
Doppler index of global RV dysfunction as suggested by Tei et al.

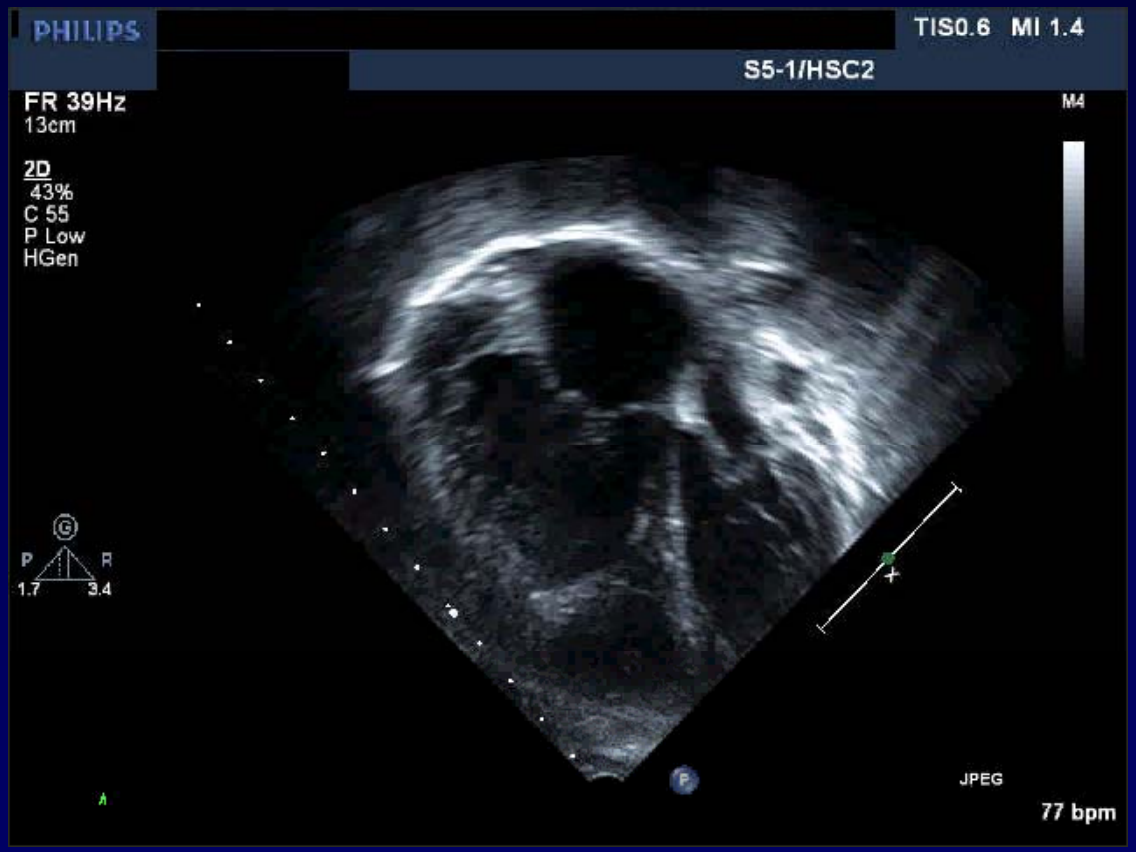
Celermajer and Marwick, Int J Card (2008) 125 :294–303

Assessment of RV function by echo remains difficult

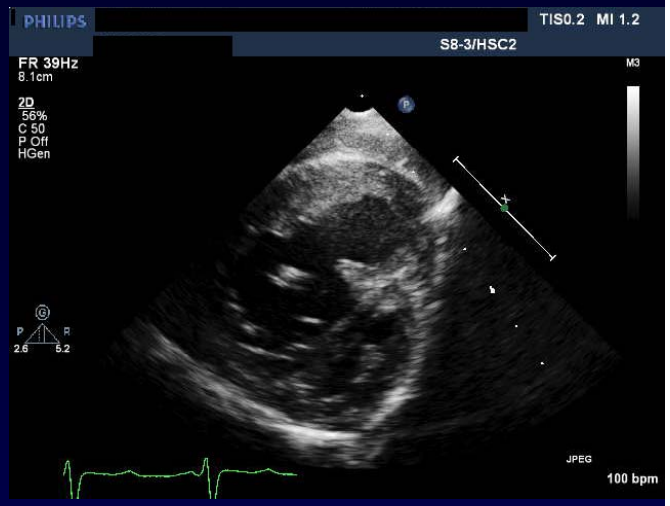
- No axis of symmetry (complicates geometric modeling).
- Endocardial delineation is difficult (prominent endocardial trabeculations)
- Retrosternal location limits acoustic access.

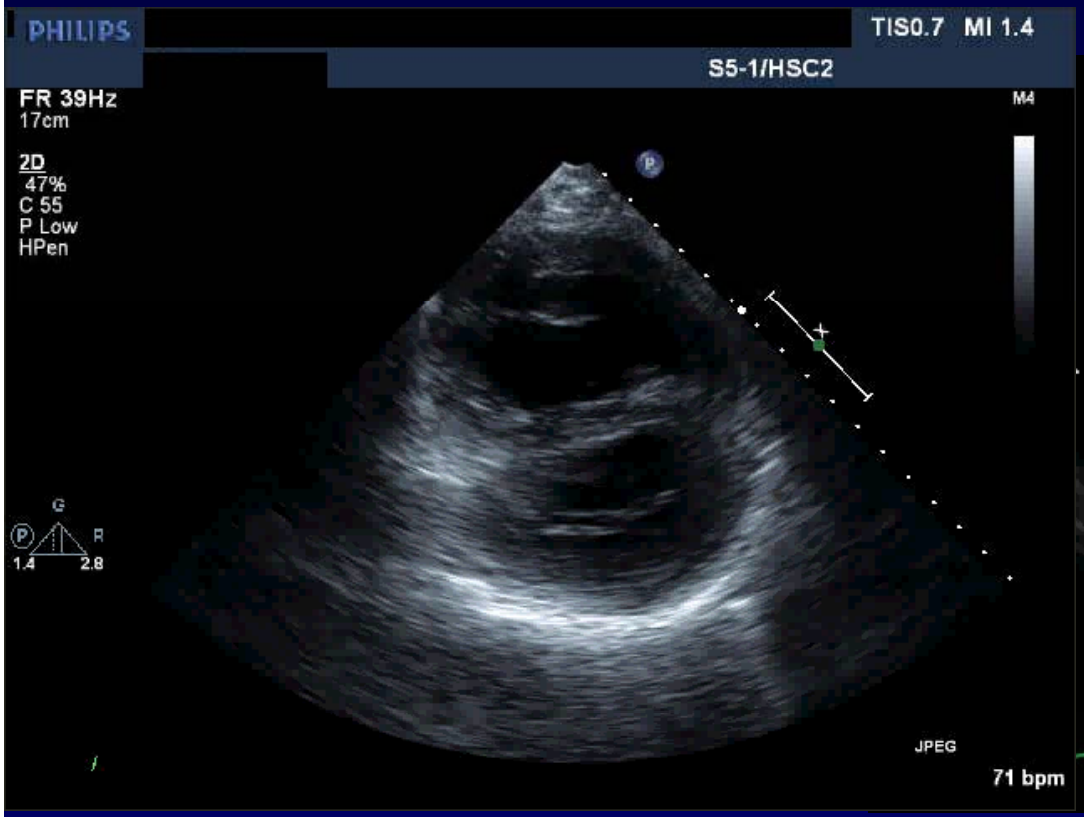
2D Assessment

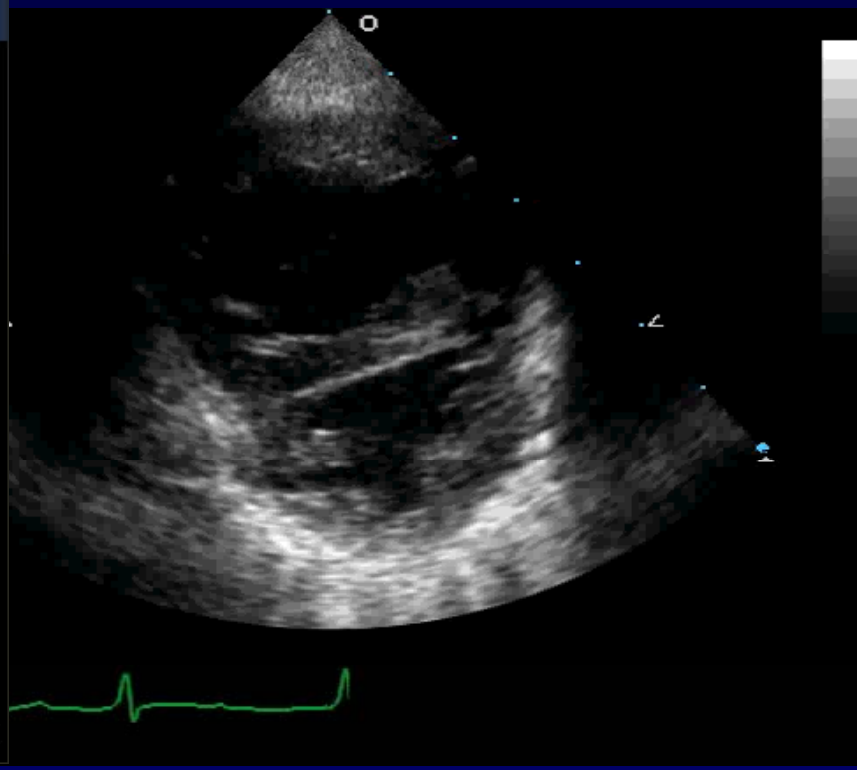
‘Eyeball’ assessment still most prevalent method

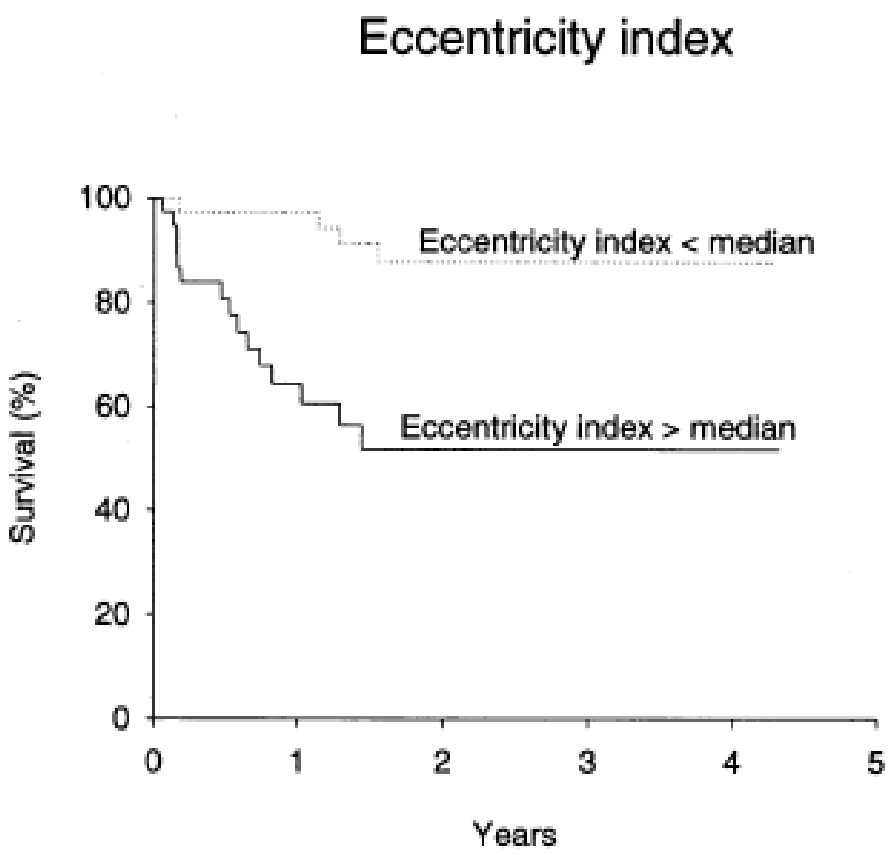


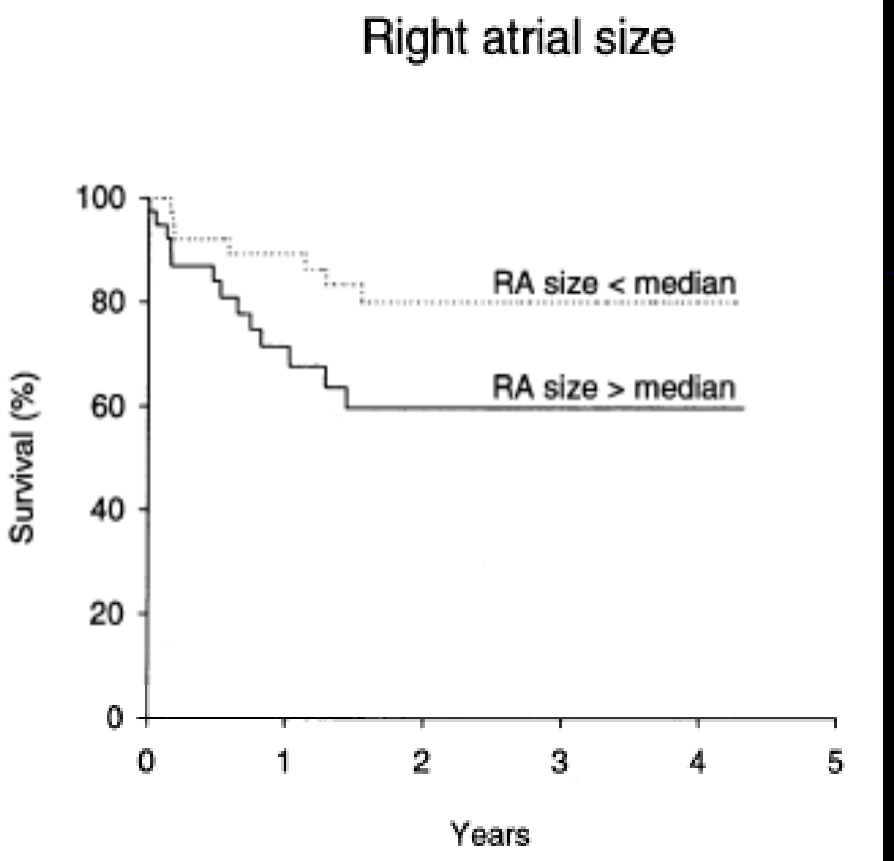
2D assessment





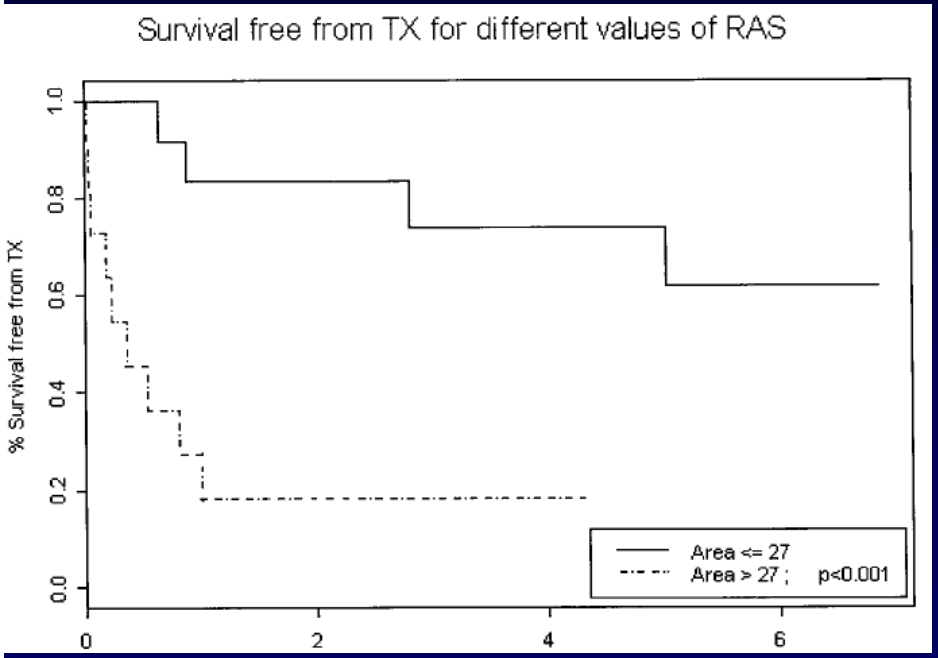






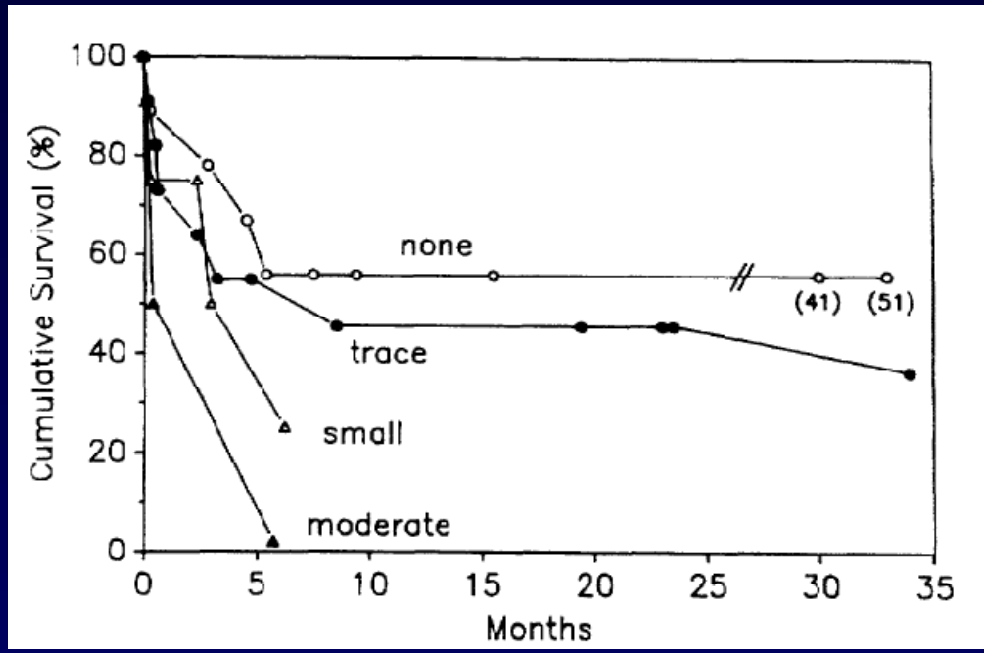
Raymond et al. JACC 2002; 39:1214-9

Right atrial size



Bustamante-Labarta, JASE 2002;15:1160

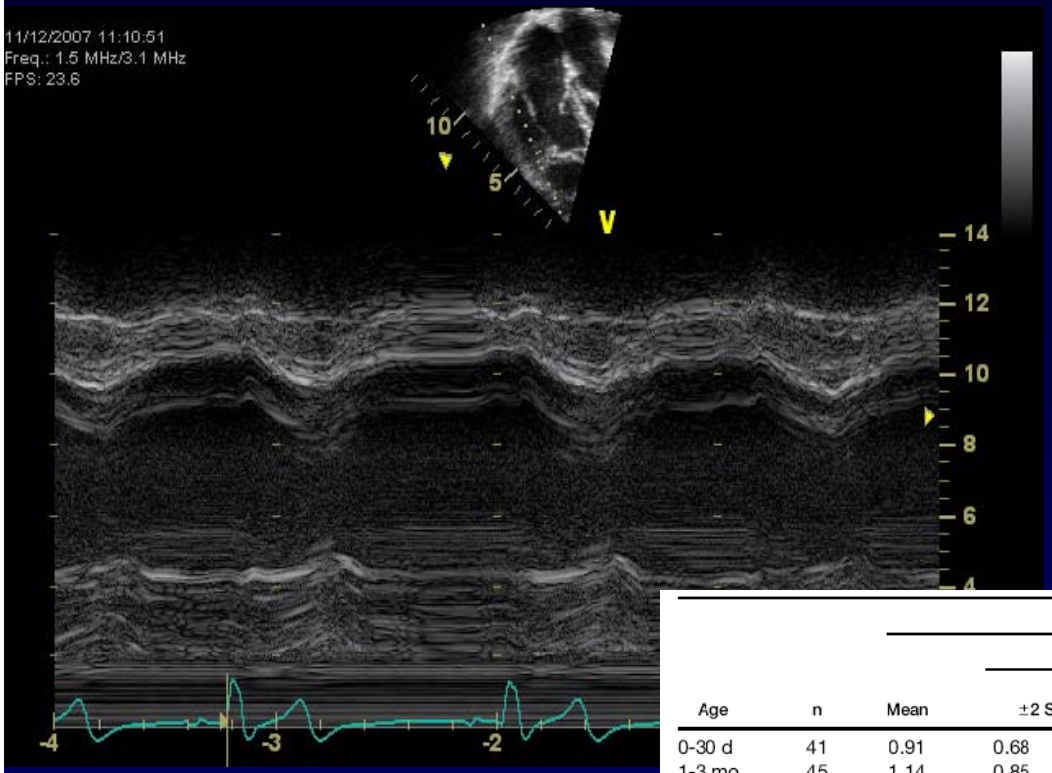
Pericardial effusion



Douglas, Circulation 1989;80:353

Annular Motion

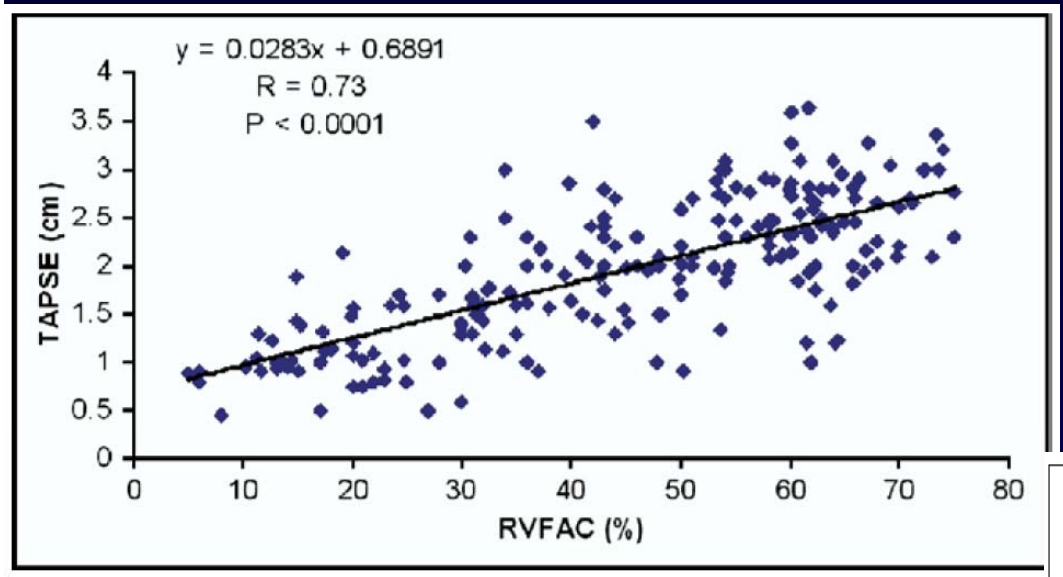
Tricuspid annular plane systolic excursion (TAPSE)



Age	n	TAPSE (cm) Mean	Bounds for z-score ranges ±2 SD (95%)		±3 SD (99%)		BSA (m^2) Mean	Minimum	Maximum	Indexed TAPSE mean/BSA mean
0-30 d	41	0.91	0.68	1.15	0.56	1.26	0.22	0.14	0.28	4.13
1-3 mo	45	1.14	0.85	1.42	0.71	1.56	0.29	0.12	0.54	3.93
4-6 mo	20	1.31	1.01	1.65	0.86	1.77	0.34	0.26	0.41	3.85
7-12 mo	22	1.44	1.13	1.77	0.97	1.91	0.40	0.31	0.47	3.6
1 y	25	1.55	1.25	1.88	1.10	2.00	0.47	0.3	0.69	3.29
2 y	39	1.65	1.36	1.94	1.22	2.09	0.53	0.4	0.62	3.11
3 y	27	1.74	1.48	2.02	1.35	2.14	0.63	0.52	0.77	2.76
4 y	47	1.82	1.56	2.07	1.43	2.20	0.70	0.6	0.91	2.6
5 y	29	1.87	1.60	2.13	1.47	2.26	0.77	0.63	0.99	2.42
6 y	41	1.90	1.62	2.18	1.48	2.33	0.82	0.46	1.06	2.31
7 y	32	1.94	1.64	2.25	1.49	2.39	0.94	0.75	1.17	2.06
8 y	23	1.97	1.67	2.28	1.52	2.43	0.97	0.79	1.39	2.03
9 y	20	2.01	1.73	2.30	1.58	2.44	1.00	0.8	1.32	2.01
10 y	27	2.05	1.79	2.31	1.65	2.46	1.15	0.82	1.54	1.78
11 y	25	2.10	1.83	2.36	1.69	2.50	1.28	1.06	1.55	1.64
12 y	18	2.14	1.84	2.43	1.68	2.60	1.39	1.08	1.67	1.53
13 y	20	2.20	1.85	2.54	1.68	2.71	1.48	1.03	1.87	1.48
14 y	35	2.26	1.87	2.65	1.68	2.84	1.55	1.11	1.93	1.45
15 y	25	2.33	1.93	2.75	1.74	2.92	1.59	1.32	1.96	1.46
16 y	34	2.39	1.98	2.78	1.78	3.01	1.66	1.3	2.04	1.43
17 y	27	2.45	2.04	2.88	1.83	3.06	1.77	1.43	2.06	1.38
18 y	21	2.47	2.05	2.91	1.84	3.10	1.79	1.34	2.25	1.37

Koestenberger, JASE 2009;22:715

TAPSE correlates with RV EF



López-Candales, Am J Cardiol 2006;98:973

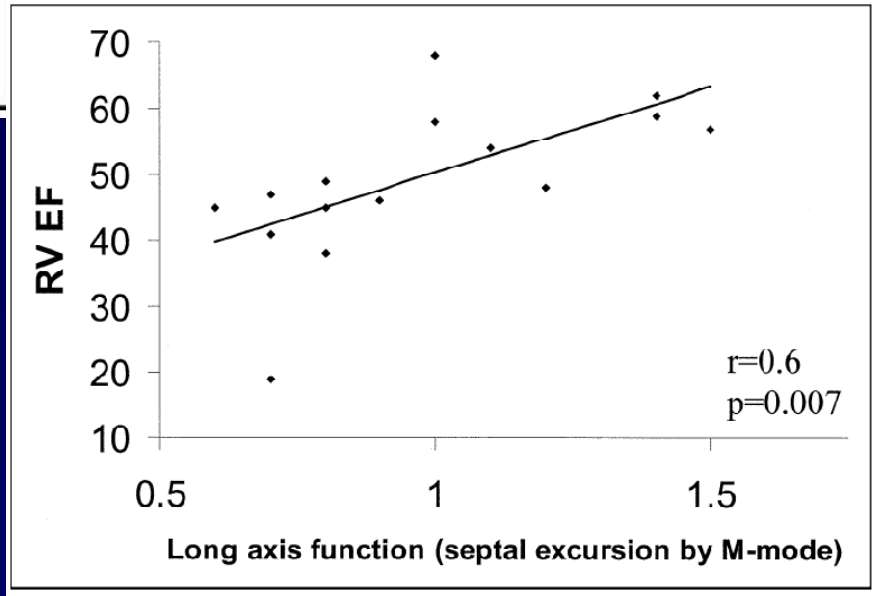
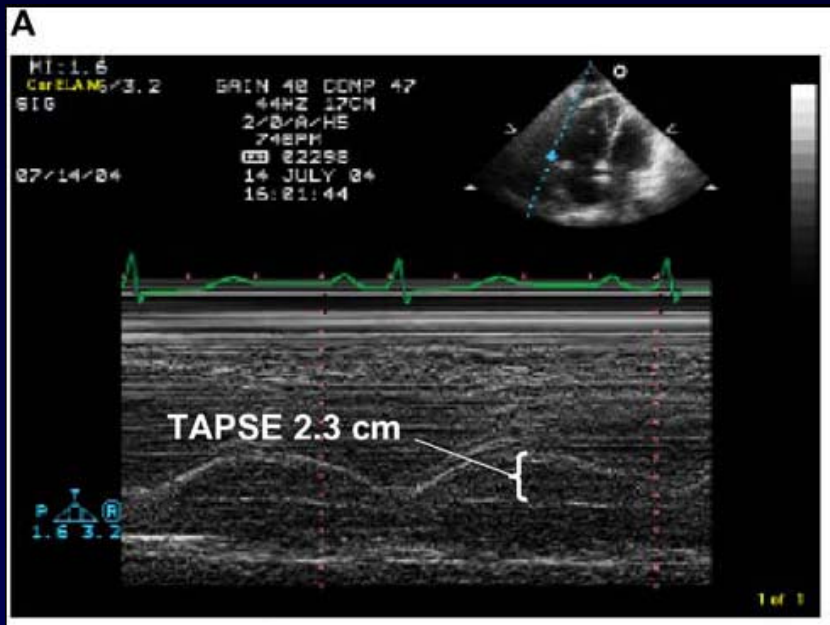


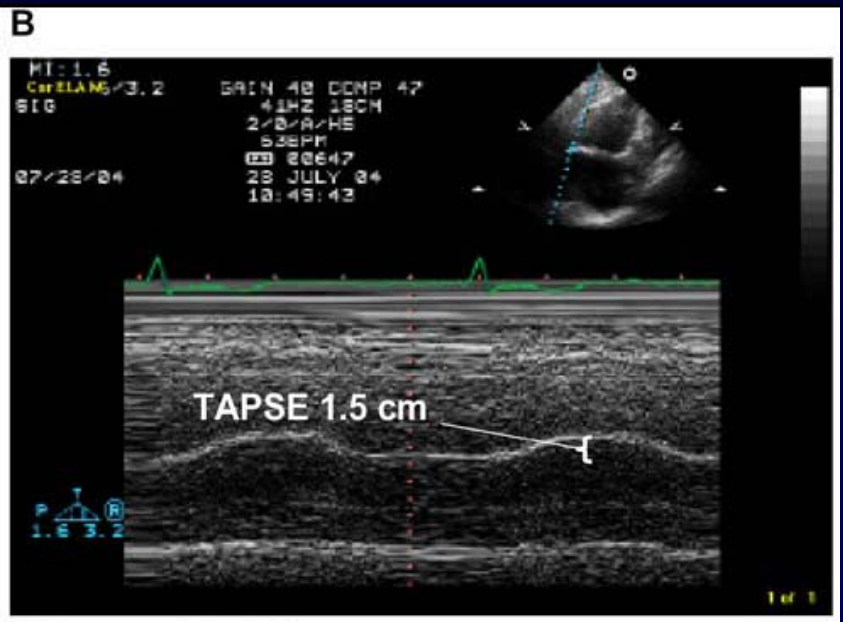
FIGURE 3. Correlation of the RV ejection fraction (EF) by MRI with long-axis ventricular function by M-mode imaging.

Lissin, Am J Cardiol 2004;93:654–657

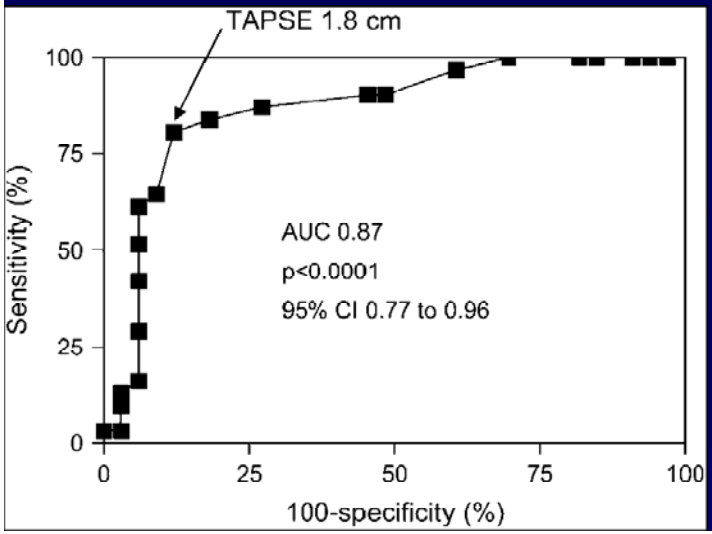
What is the value of TAPSE ?



SVI	37 ml/m^2
CI	2.9 l/min/m^2
RAP	11 mmHg
mPAP	44 mmHg
PVR	6.5 mmHg/l/min



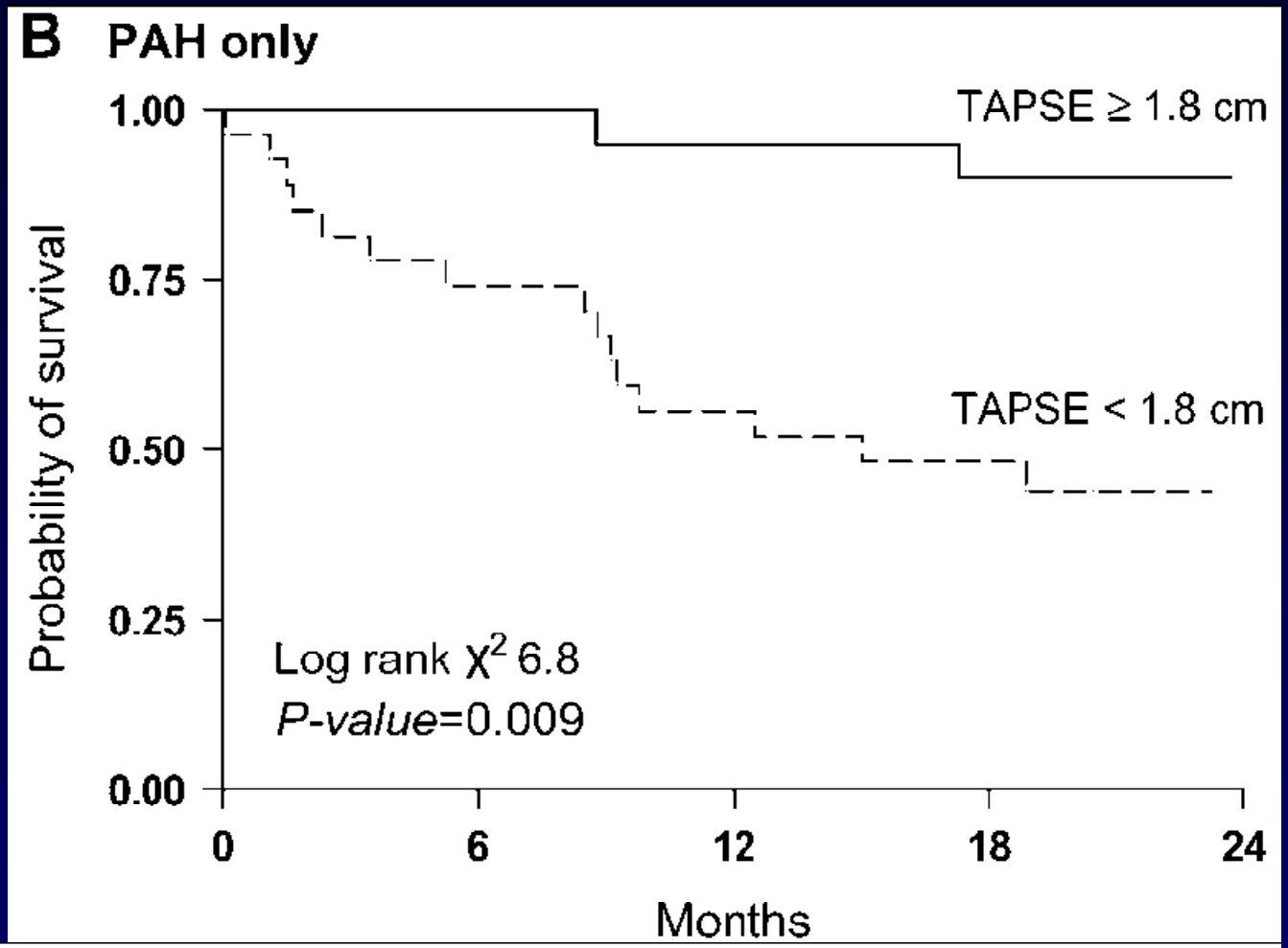
SVI	24 ml/m^2
CI	2.0 l/min/m^2
RAP	10 mmHg
mPAP	50 mmHg
PVR	11.0 mmHg/l/min



Ability of TAPSE to predict RV stroke volume index of 29 ml/m^2.

Forfia, Am J Respir Crit Care Med 2006 174:1034–1041

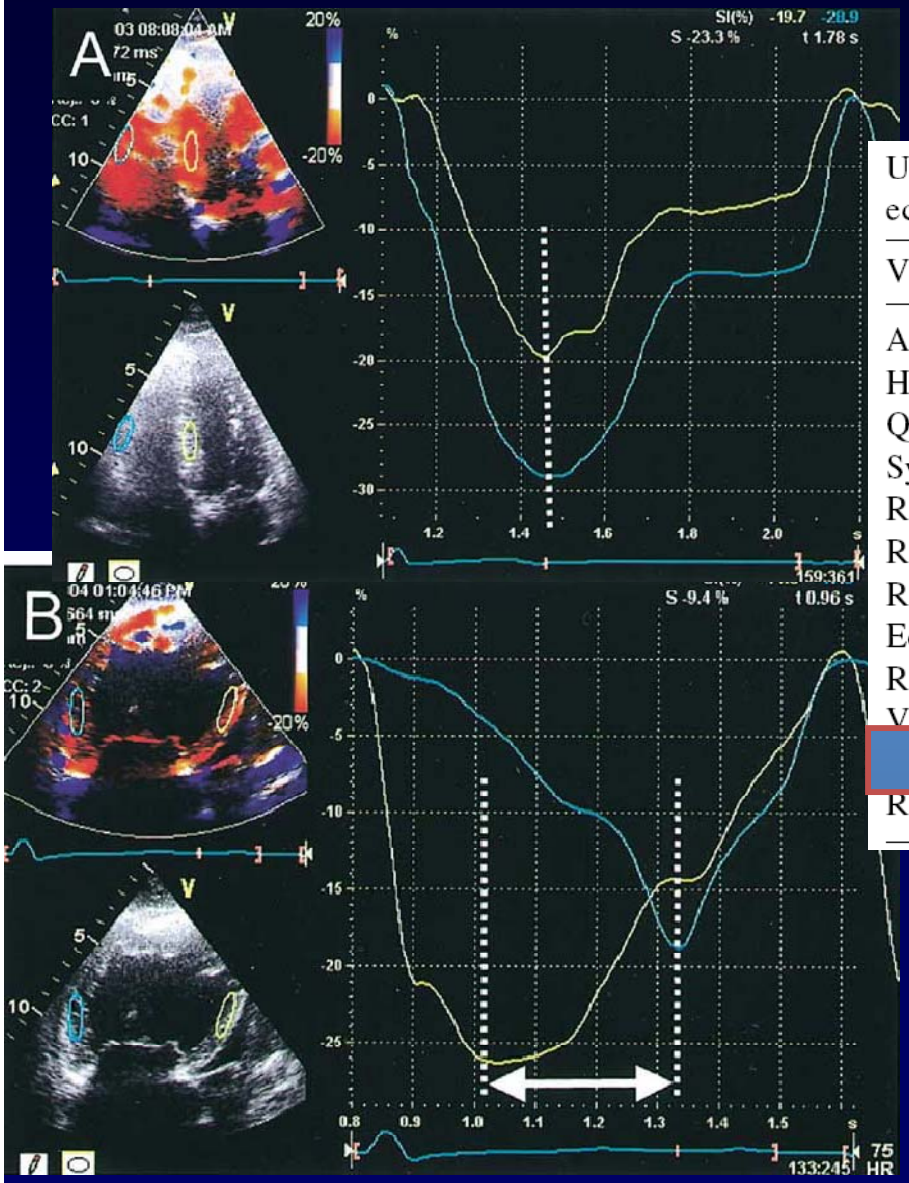
What is the value of TAPSE ?



TAPSE ≥ 1.8 cm (N)	17	17	16	15	15
TAPSE < 1 .8 cm (N)	30	23	18	16	13

Forfia, Am J Respir Crit Care Med, 2006 174:1034

A smaller TAPSE may originate from RV mechanical delay

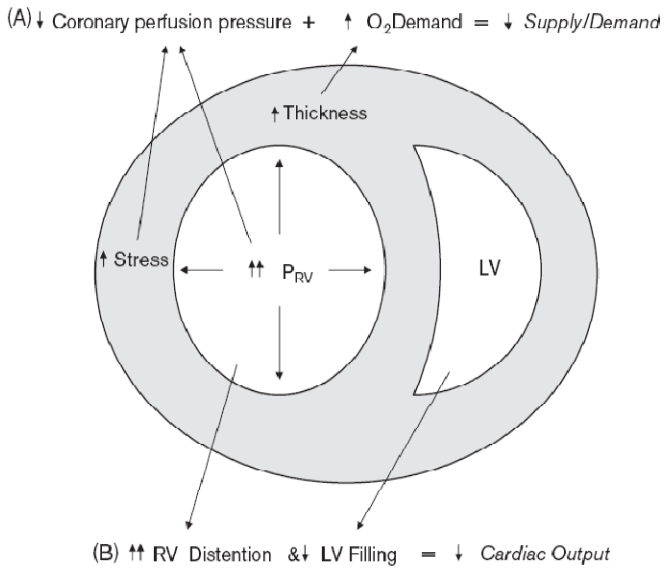


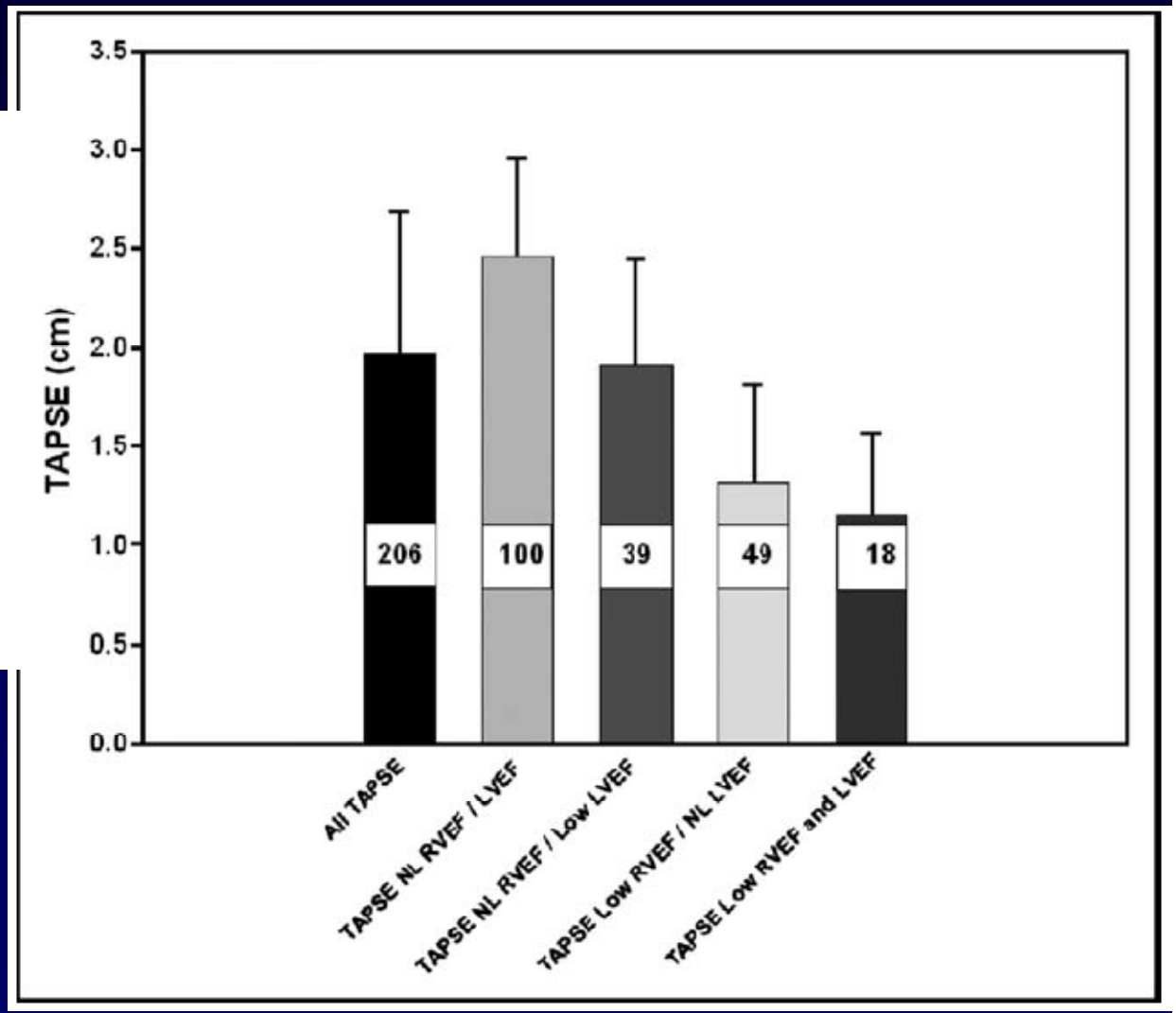
Univariate analysis of RV mechanical delay to other clinical and echocardiographic variables in patients with abnormal RV performance

Variable	r Value	p Value
Age (yrs)	0.06	NS
Heart rate (beats/min)	0.05	NS
QRS duration (ms)	0.67	<0.05
Systolic pulmonary artery pressure (mm Hg)	0.67	<0.05
RV end-diastolic area (cm)	0.73	<0.05
RV end-systolic area (cm)	0.81	<0.05
RV fractional area change (%)	−0.89	<0.05
Eccentricity index	0.75	<0.05
RV myocardial performance index	0.77	<0.05
Vena contracta width	0.63	<0.05
RV free wall peak strain (%)	0.58	<0.05

Lopez-Candales *Am J Cardiol* 2005;96:602

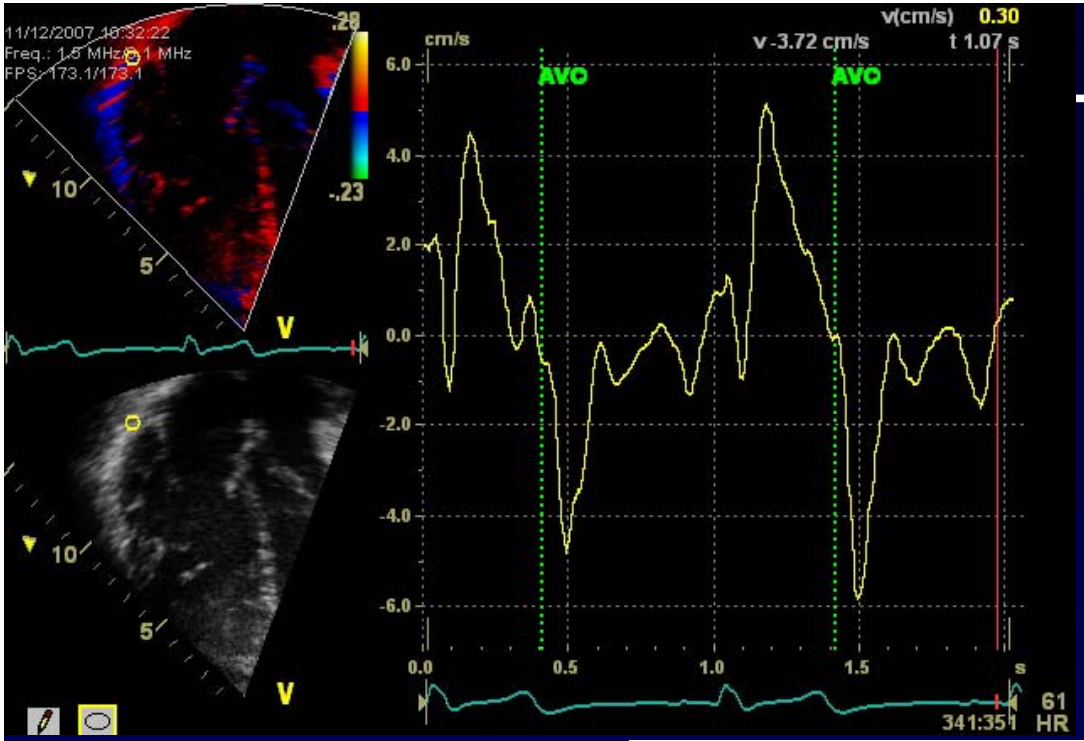
LV function also influences TAPSE

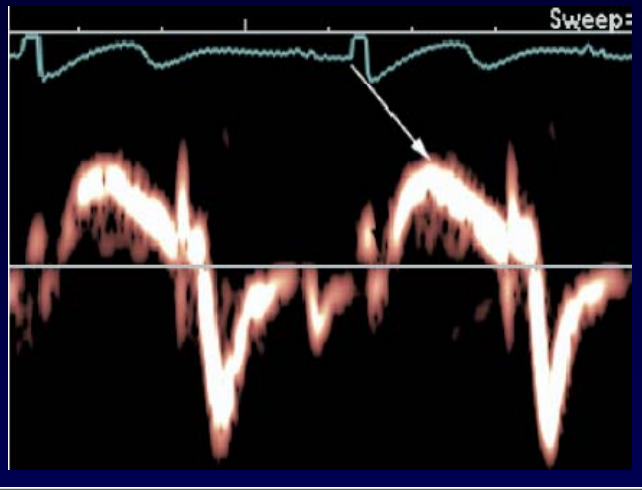




López-Candales, AJC 2006;98:973

Tricuspid velocities

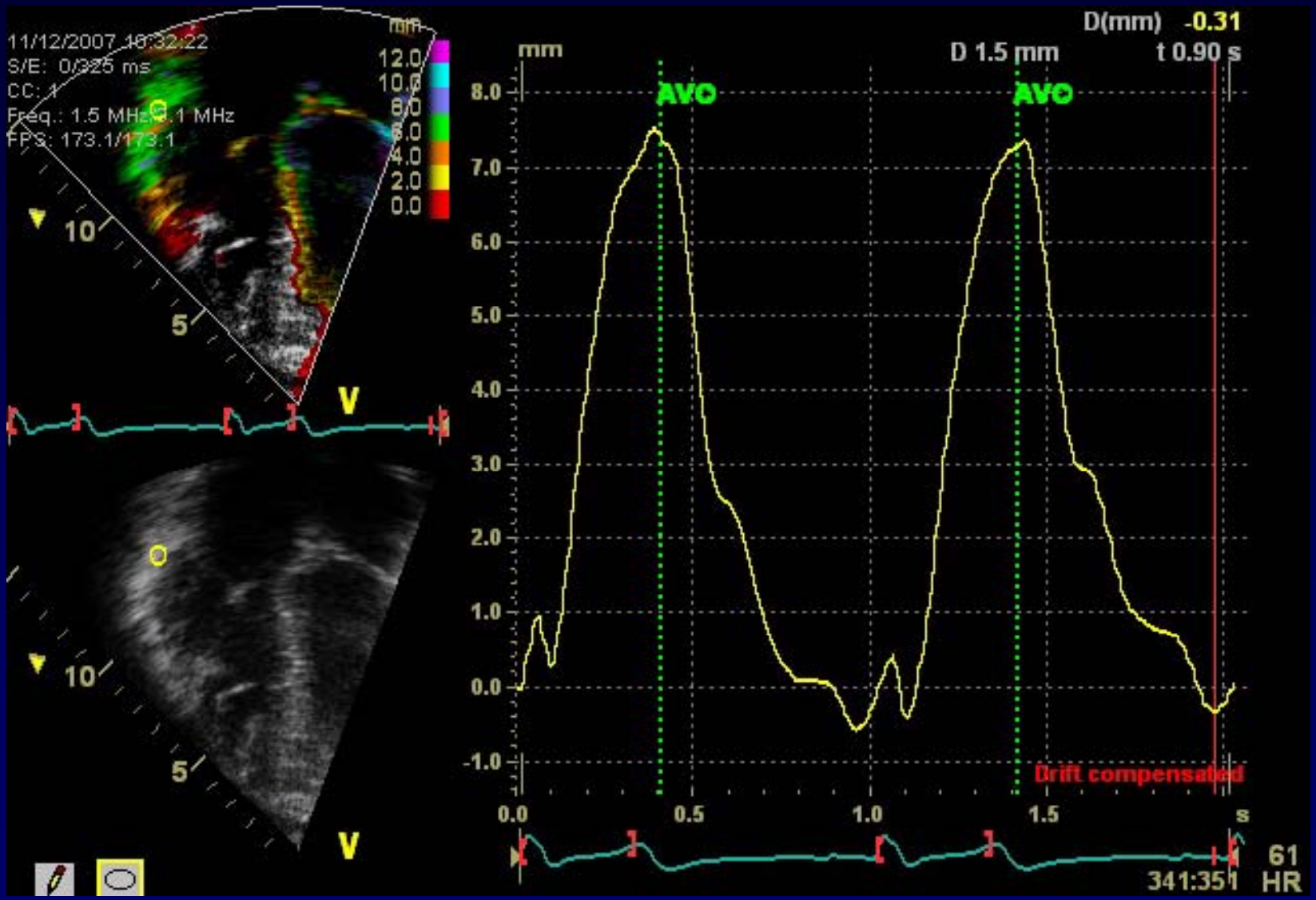




Age group	N	E'-wave velocity	A'-wave velocity	S'-wave velocity
Tricuspid annular				
<1 y	63	13.8 ± 8.2 (11.7–15.9)	9.8 ± 2.4 (9.1–10.5)	10.2 ± 5.5 (8.8–11.7)
1–5 y	68	17.1 ± 4.0† (16.1–18.1)	10.9 ± 2.7 (10.2–11.6)	13.2 ± 2.0† (12.7–13.7)
6–9 y	55	16.5 ± 3.0 (15.7–17.4)	9.8 ± 2.7 (9.0–10.6)	13.4 ± 2.0 (12.8–14.0)
10–13 y	58	16.5 ± 3.1 (15.7–17.4)	10.3 ± 3.4 (9.3–11.2)	13.9 ± 2.4 (13.2–14.5)
14–18 y	81	16.7 ± 2.8 (16.0–17.3)	10.1 ± 2.6 (9.5–10.7)	14.2 ± 2.3 (13.7–14.7)
Total	325	16.1 ± 4.7 (15.6–16.7)	10.2 ± 2.8 (9.9–10.5)	13.0 ± 3.4 (12.6–13.4)

Eidem, J Am Soc Echocardiogr 2004;17:212-21

Tissue Displacement (tracking)



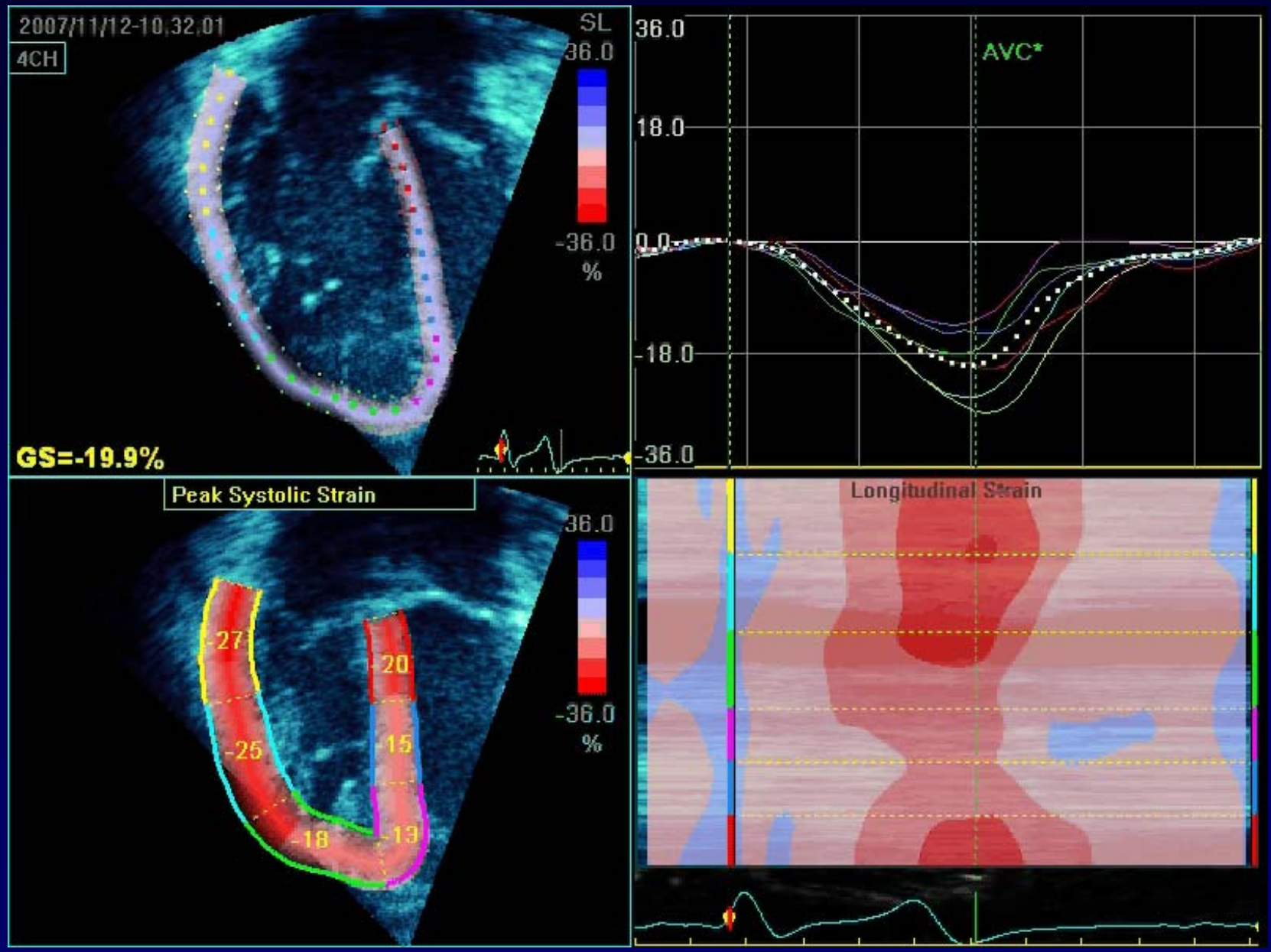
Assessment of myocardial function

Assessing Myocardial Deformation

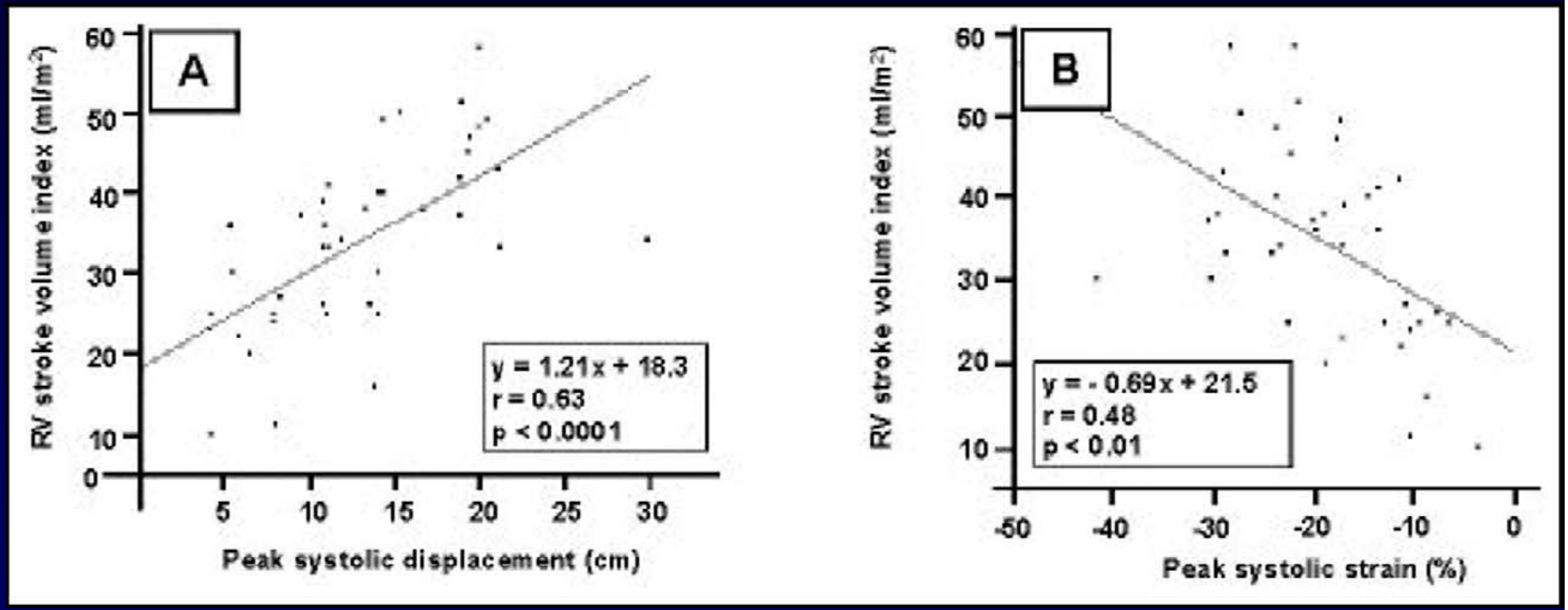
$$S = \frac{l - l_0}{l_0} = \frac{\Delta l}{l_0}$$

Courtesy of Piet Claus

RV strain by speckle tracking



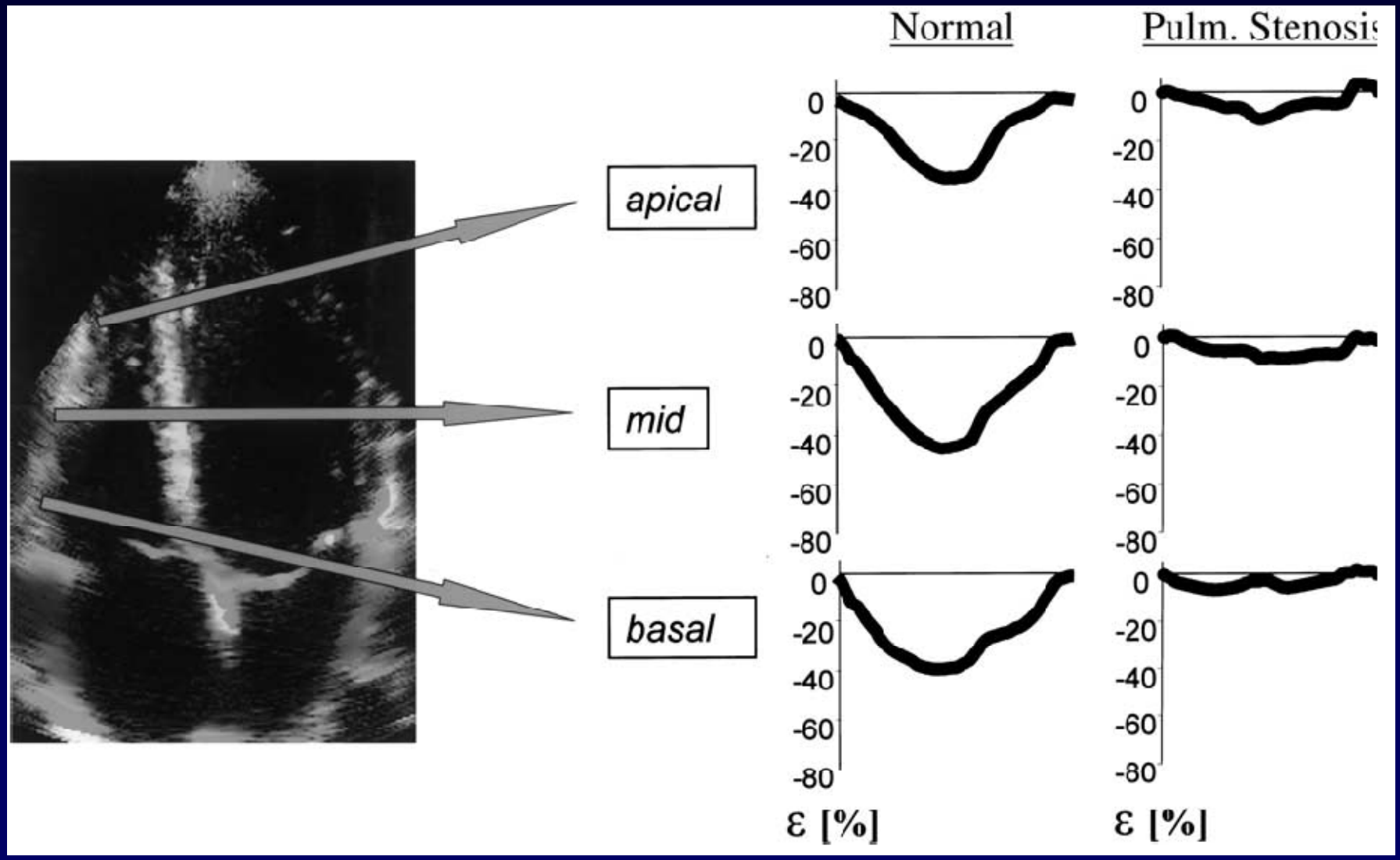
Strain correlates with stroke volume



*SV determined at cath

Urheim, AJC 2005;96:1173

RV strain is influenced by afterload



Weidemann, Ped Cardiol, 2002;23:292

Assessment of regional RV function by strain

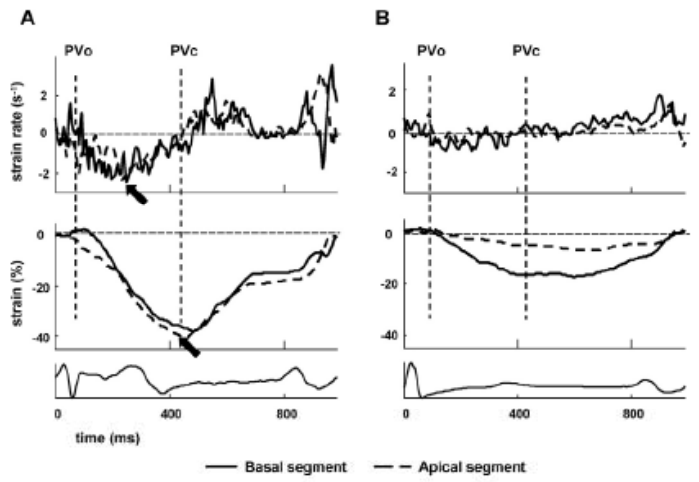
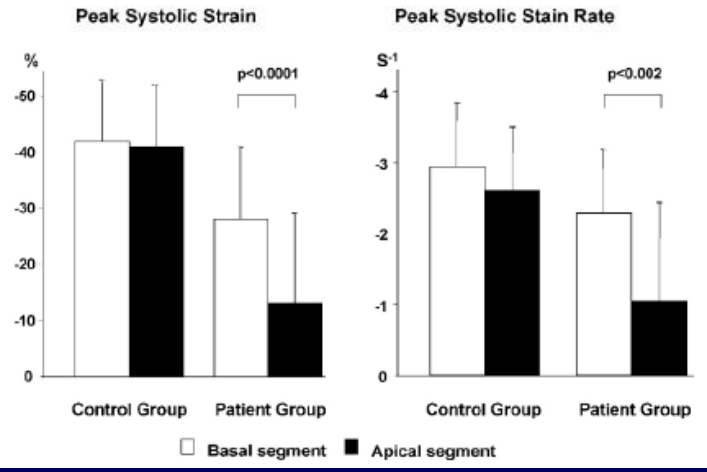
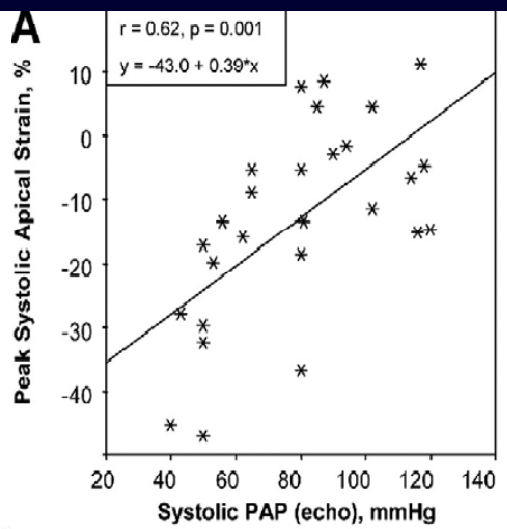
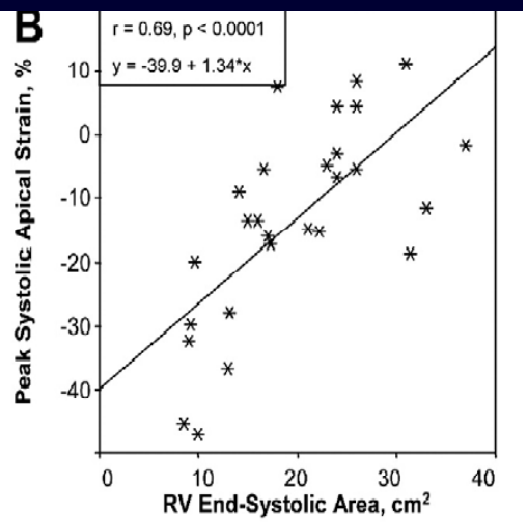
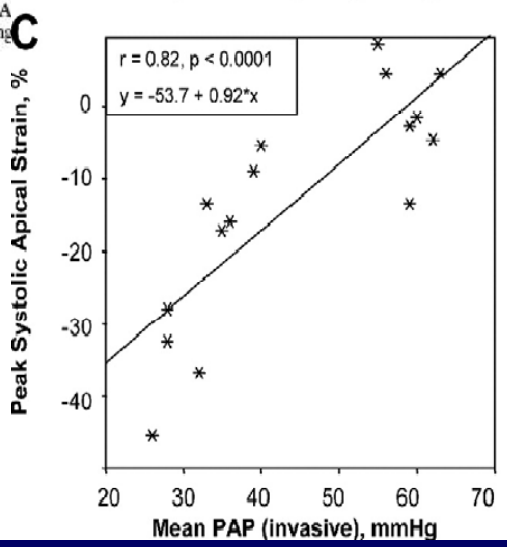


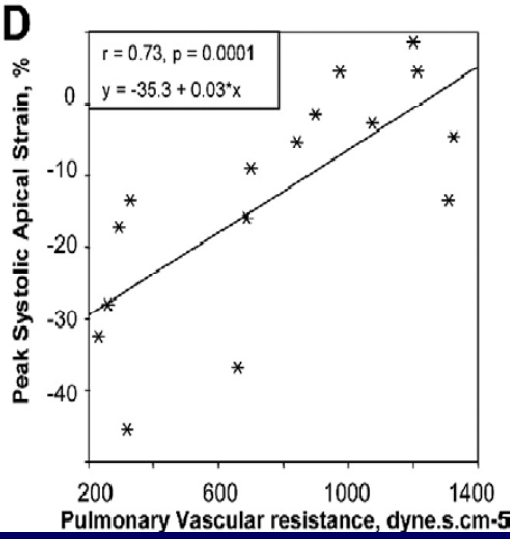
Figure 1 Right ventricular strain rate and strain profiles from basal and apical segments in control subject (A) and patient with pulmonary hypertension (B). *PVc*, Pulmonary valve closure; *PVo*, pulmonary valve opening.







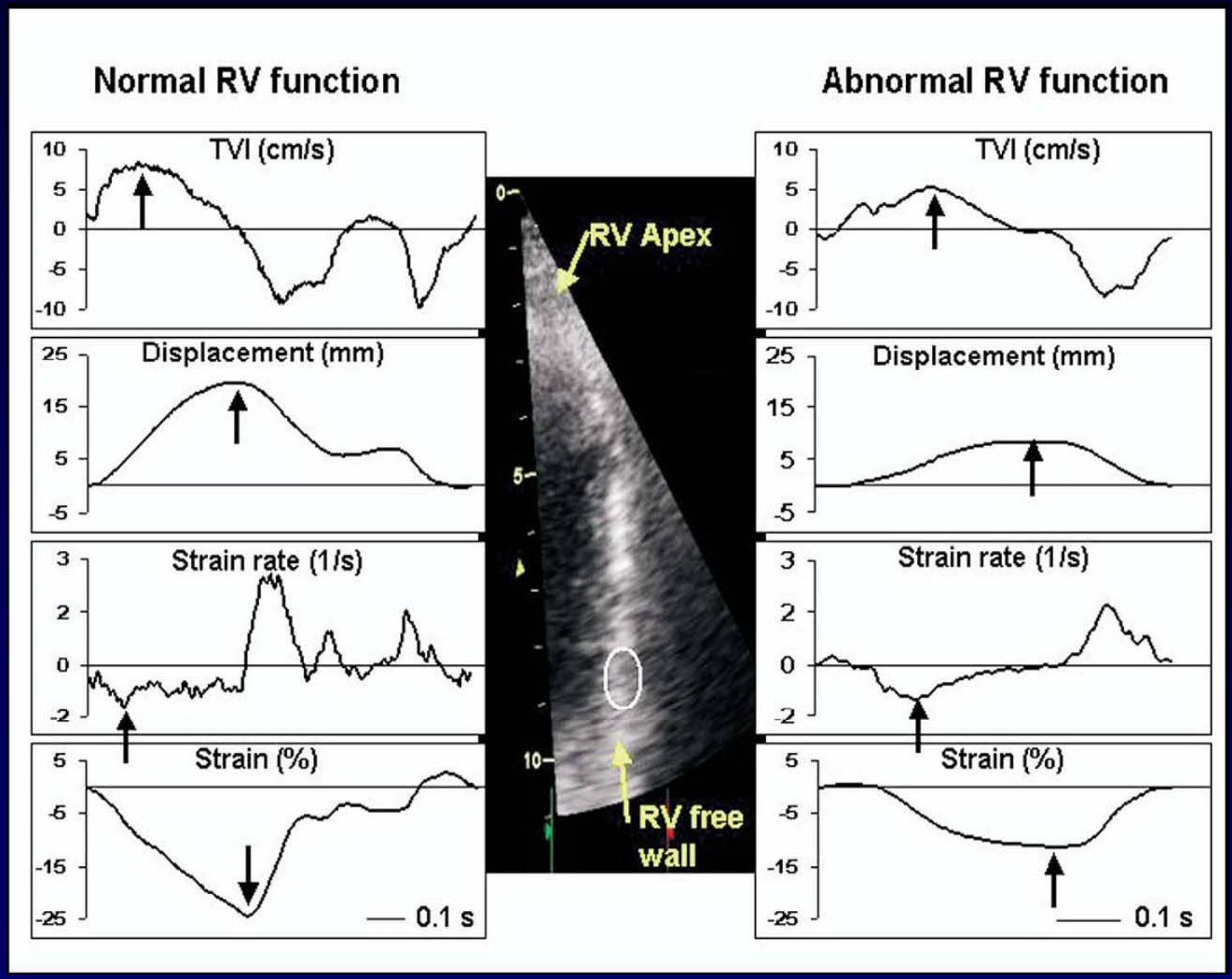




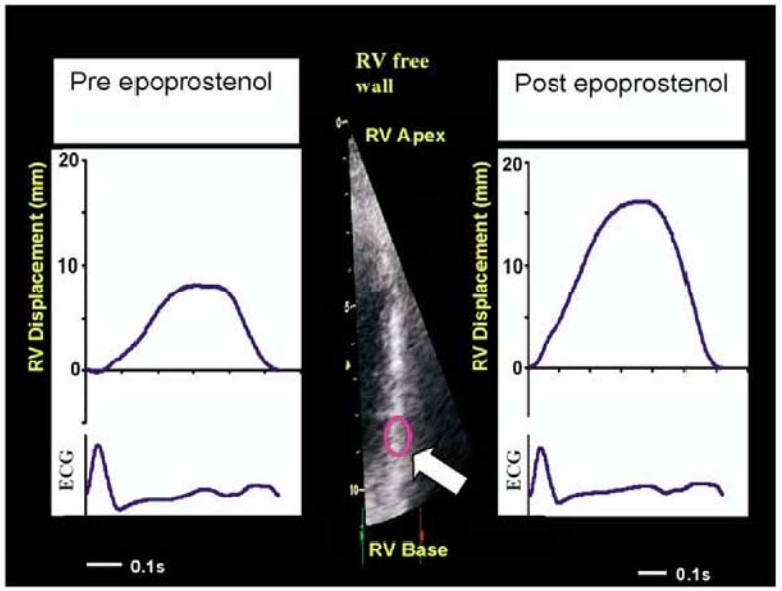
*No significant correlations between deformation and NYHA or 6-min walk test.

Dambrauskaite, J Am Soc Echocardiogr 2007;20:1172

RV myocardial function



Urheim, AJC 2005;96:1173



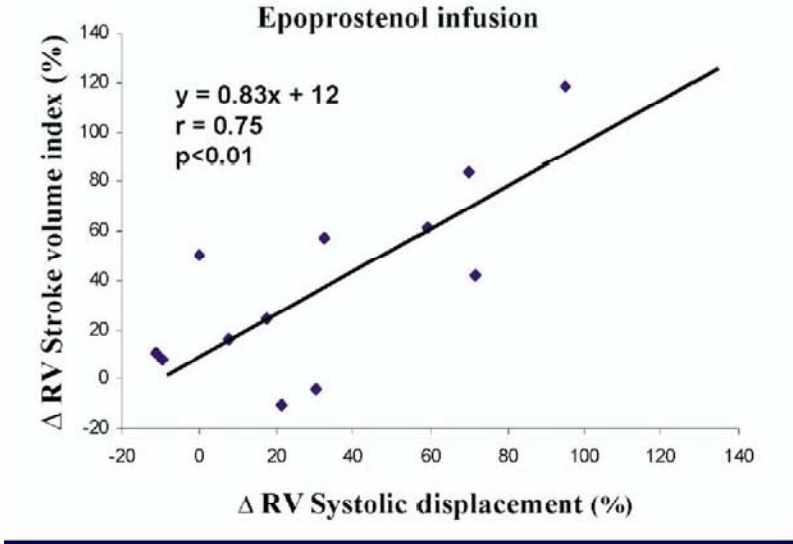


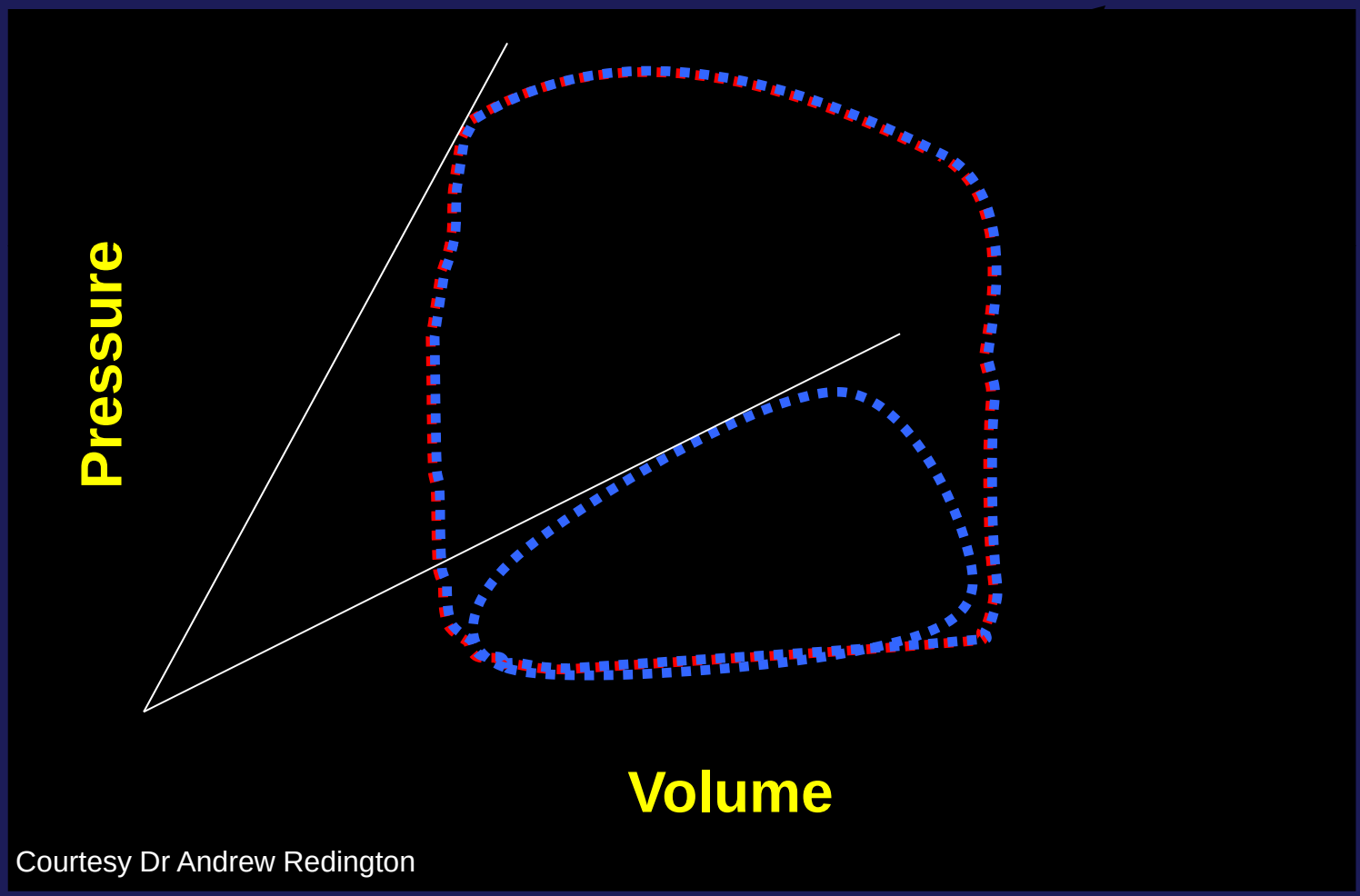
Table 2
Invasive hemodynamic and echocardiographic data before and after epoprostenol infusion

Variable	Before Epoprostenol (n = 12)	Peak Epoprostenol (n = 12)
RV peak systolic pressure (mm Hg)	74.5 ± 20.0	65.7 ± 19.3*
Pulmonary vascular resistance (Wood units)	18.3 ± 12.2	11.4 ± 10.3*
RVSVI (ml/m^2)	27.1 ± 13.5	35.7 ± 15.2*
Isovolumic myocardial acceleration (m/s^2)	1.6 ± 1.0	1.6 ± 1.4
RV peak systolic velocity (cm/s)	5.0 ± 1.9	6.1 ± 2.2*
RV peak systolic displacement (mm)	8.8 ± 4.5	11.1 ± 5.2*
RV peak systolic strain rate (1/s)	−1.2 ± 0.9	−1.5 ± 0.8
RV peak systolic strain (%)	−13.8 ± 8.5	−17.6 ± 9.1*

* p ≤0.05.

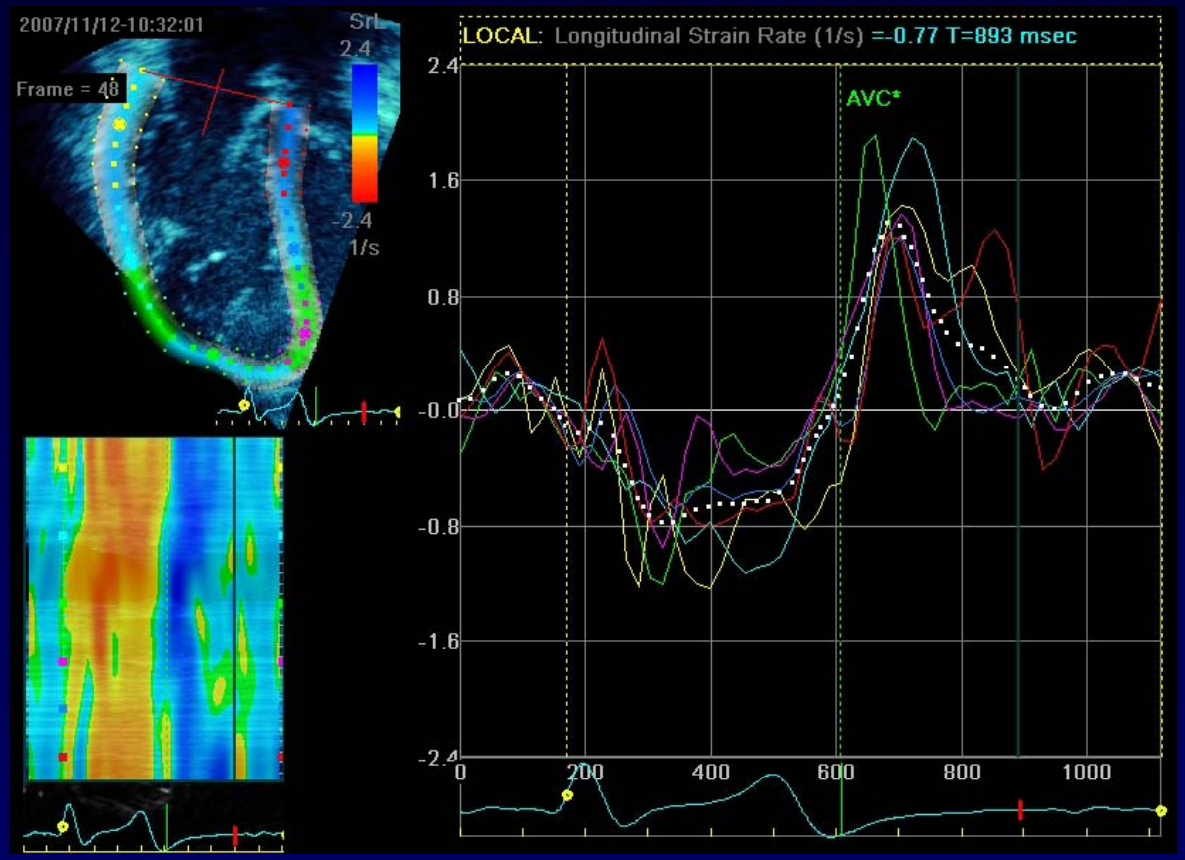
Urheim, Relation of Tissue Displacement and Strain to Invasively Determined Right Ventricular Stroke Volume, **AJC 2005;96:1173**

Assessing RV End-systolic Elastance

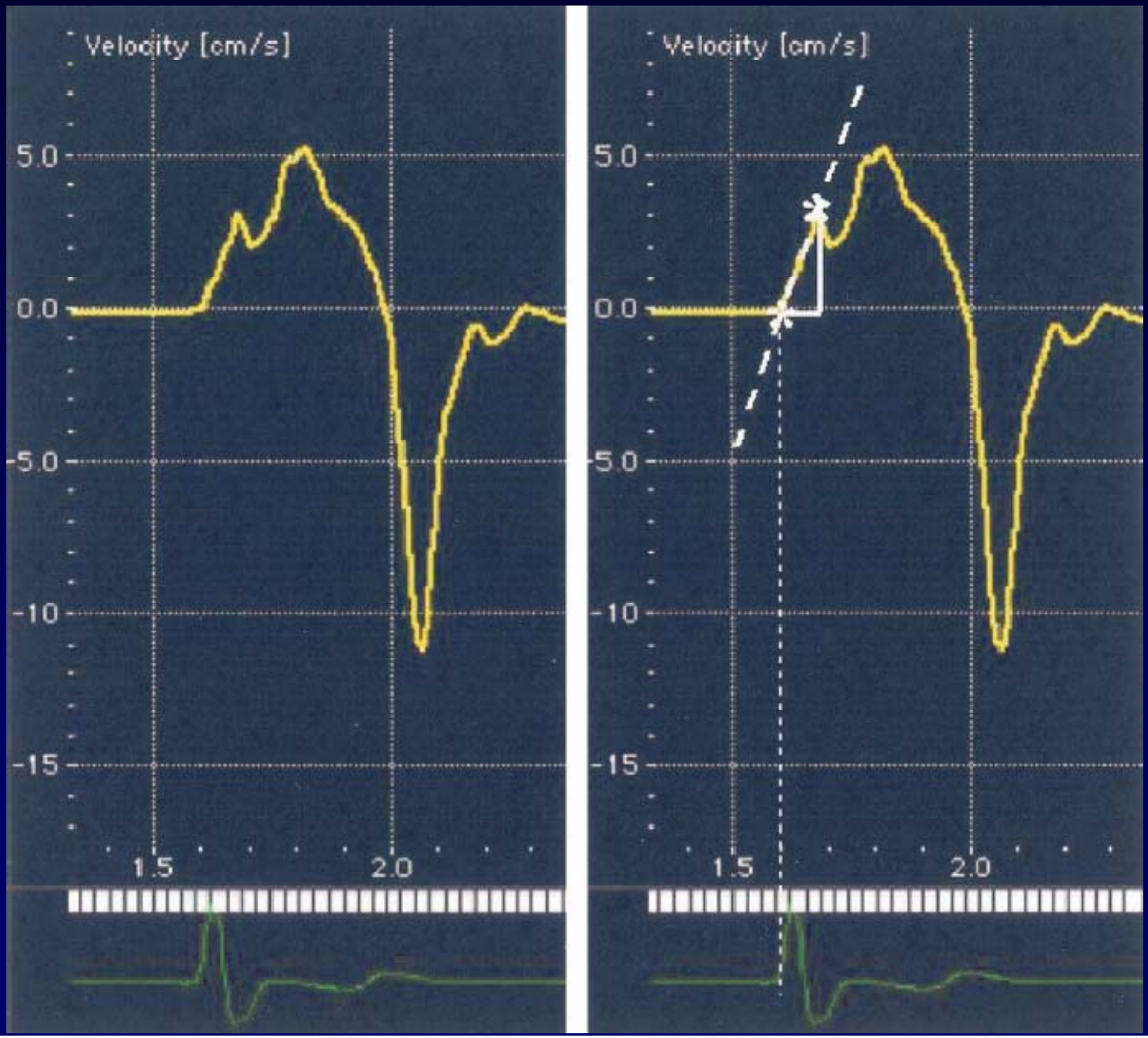


Redington et al. Br Heart J 1988:58:23-8

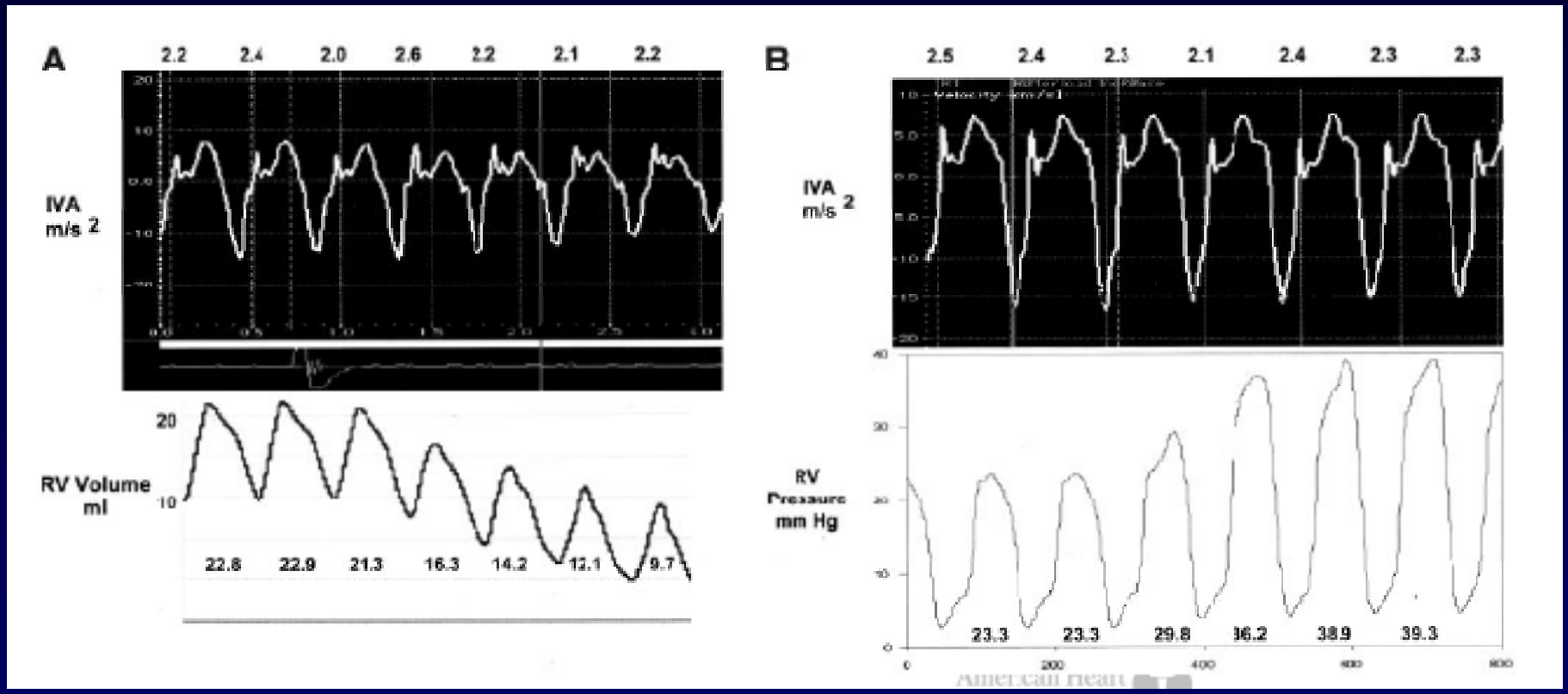
Strain rate


2007/11/12-10:32:01
Srl
2.4
Frame = 48
-2.4
1/s
LOCAL: Longitudinal Strain Rate (1/s) =-0.77 T=893 msec
AVC*
2.4
1.6
0.8
-0.0
-0.8
-1.6
-2.4
0
200
400
600
800
1000

Isovolumic acceleration



Isovolumic acceleration is relatively load independent



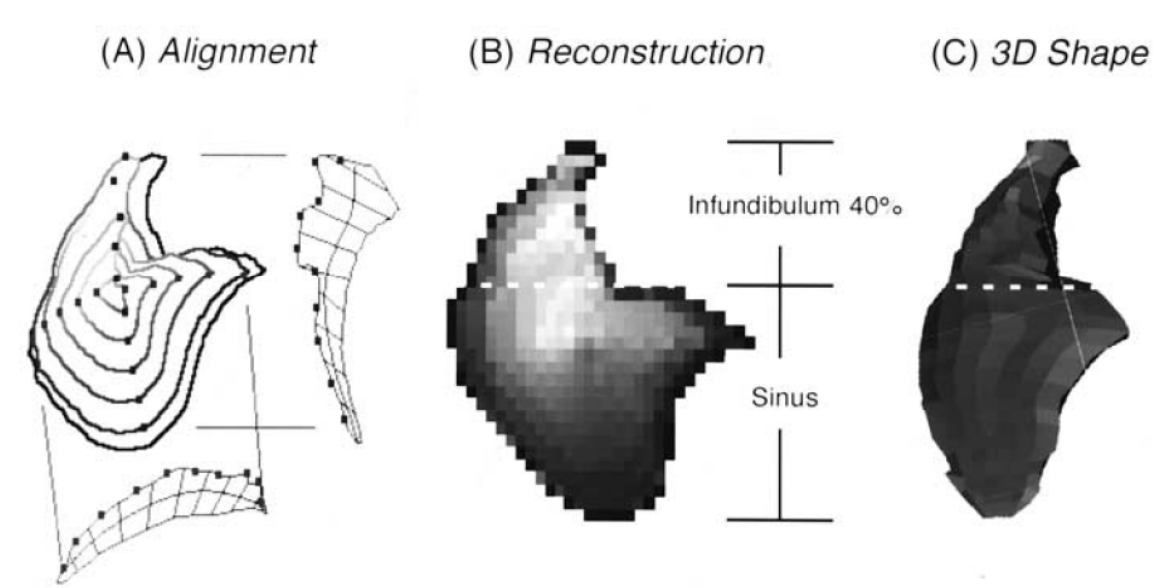
Vogel, Circulation. 2002;105:1693-1699

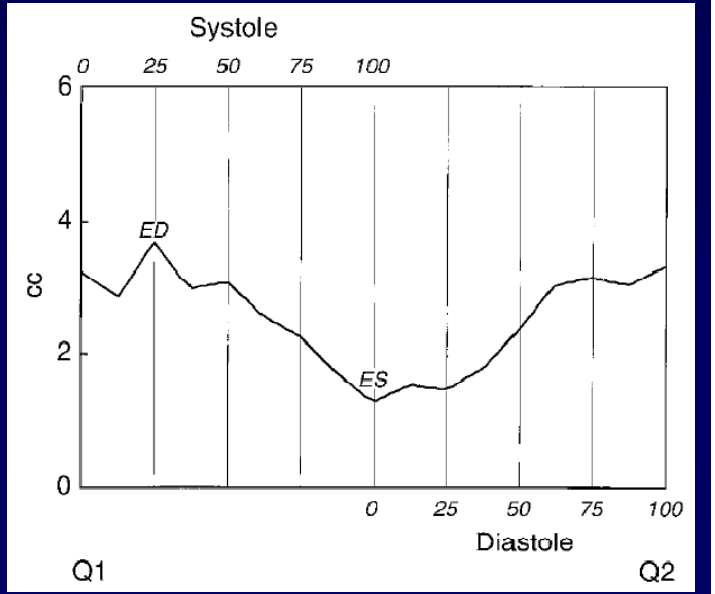
3D Assessment

3D assessment of RV function

Echocardiographic Assessment of the Right Ventricular Response to Hypertension in Neonates on the Basis of Average-shaped Contraction Models

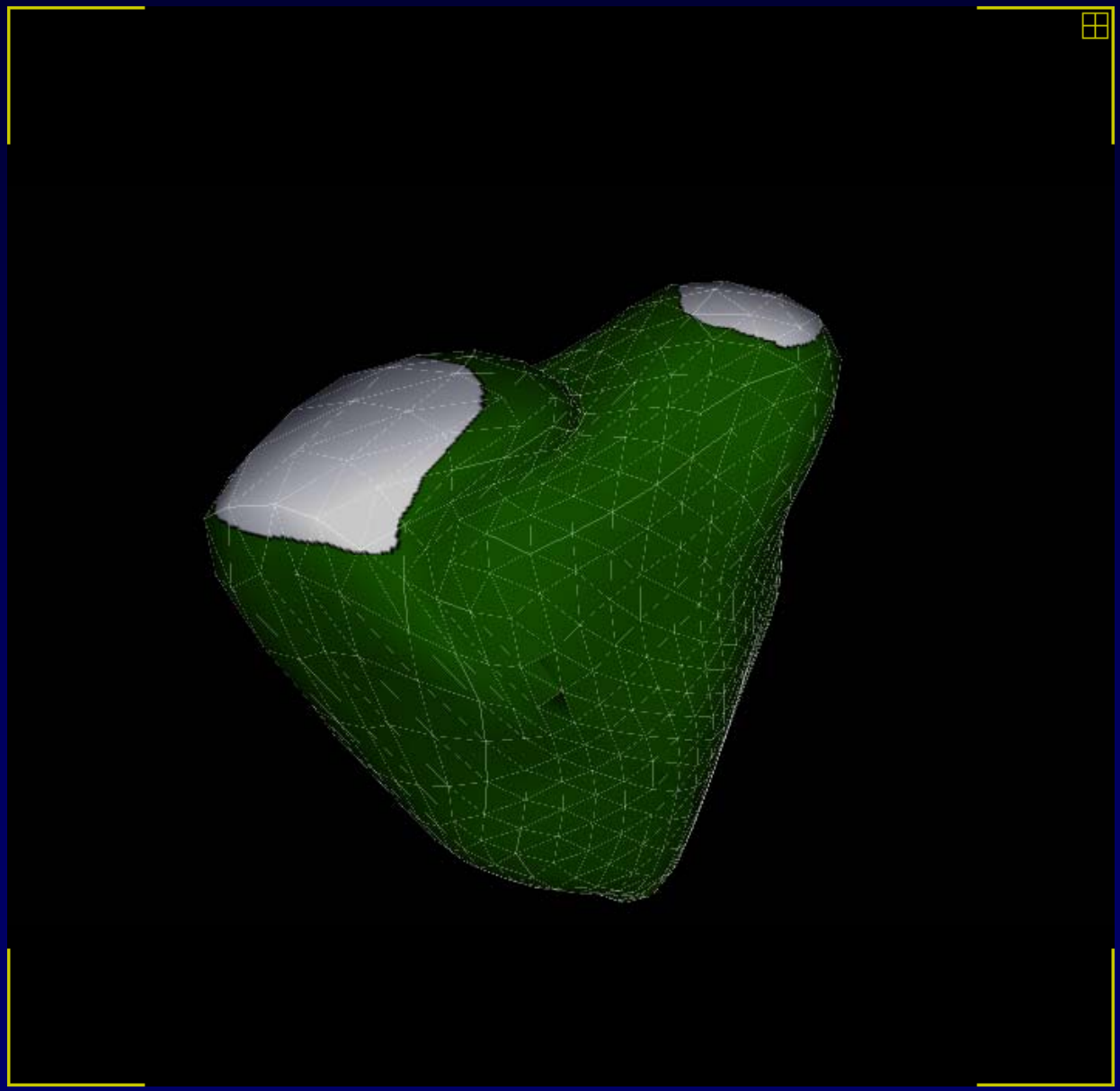
Edward N. Marcus, MSc, Ricardo A. Munoz, MD, Renee Margossian, MD, Steven D. Colan, MD, and David L. Wessel, MD, *Boston, Massachusetts; and Miami, Florida*



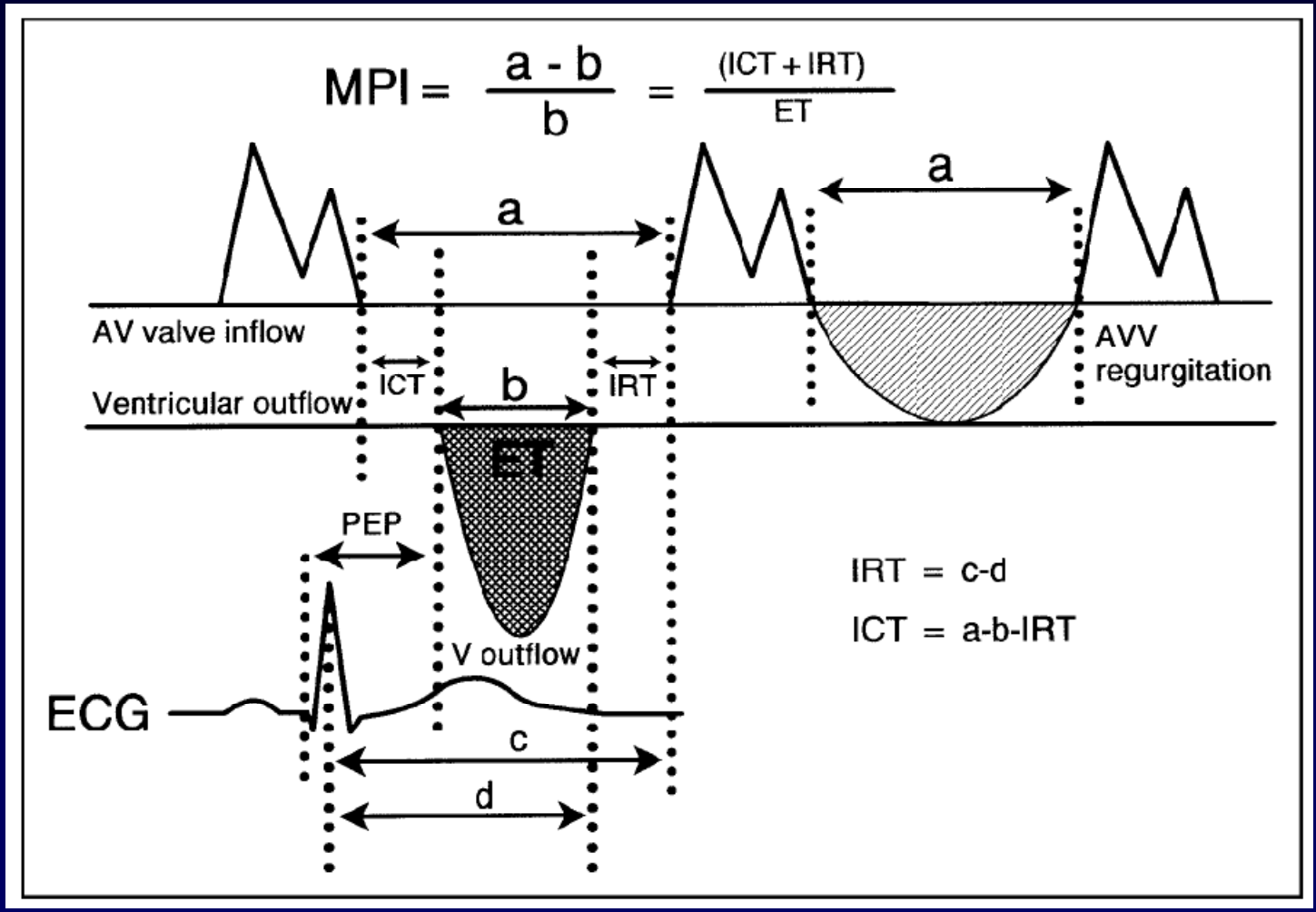


Marcus, J Am Soc Echocardiogr 2002;15: 1145

3D RV function



MPI for RV function in CHD



“a” is duration of tricuspid valve regurgitation

“b” is RV ejection time

Eidem , Am J Cardiol 2000;86:654

Loading and MPI

TABLE 3 Comparison of Doppler Time Intervals: Pre- Versus Postoperative Atrial Septal Defect

	ASD Group I		ASD Group II	
	Pre (n = 16)	Post (n = 16)	Pre (n = 16)	Post (n = 16)
Heart rate (beats/min)	97 (17)	107 (28)†	74 (16)	85 (17)*
RV MPI	0.34 (0.05)	0.33 (0.03)†	0.36 (0.03)	0.35 (.03)†
RV ICT (ms)	35 (13)	24 (9)*	45 (14)	30 (9)*
RV IRT (ms)	62 (14)	55 (13)†	76 (12)	64 (10)*
RV ejection time (ms)	282 (22)	236 (29)*	310 (38)	297 (67)†

*p <0.05 compared with preceding column; †p = NS compared with preceding column.

TABLE 4 Comparison of Doppler Time Intervals: Pre- Versus Postintervention in Pulmonary Stenosis

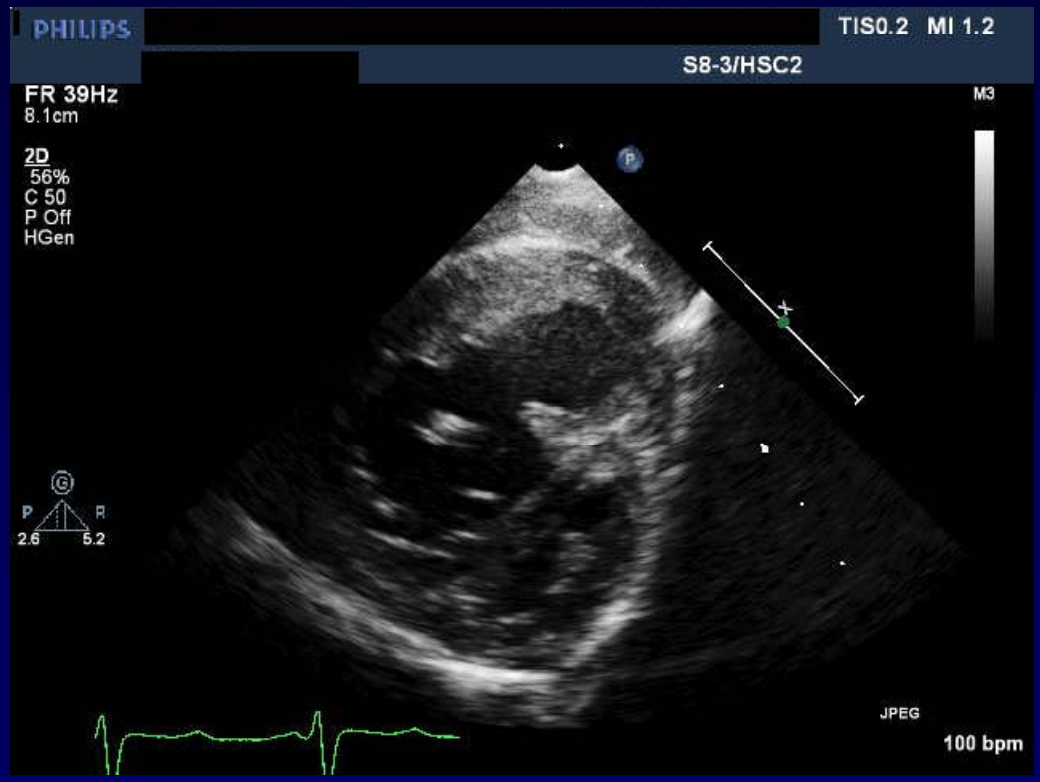
	Preintervention (n = 21)	Postintervention (n = 21)
Heart rate (beats/min)	89 (19)	99 (32)†
Peak gradient (mm Hg)	63 (29)	24 (14)*
RV MPI	0.32 (.06)	0.33 (0.06)†
RV ICT (ms)	63 (33)	57 (22)†
RV IRT (ms)	70 (19)	63 (21)†
RV ejection time (ms)	280 (57)	264 (61)†

*p <0.05 compared with preceding column; †p = NS compared with preceding column.

Eidem , AJC 2000;86:654

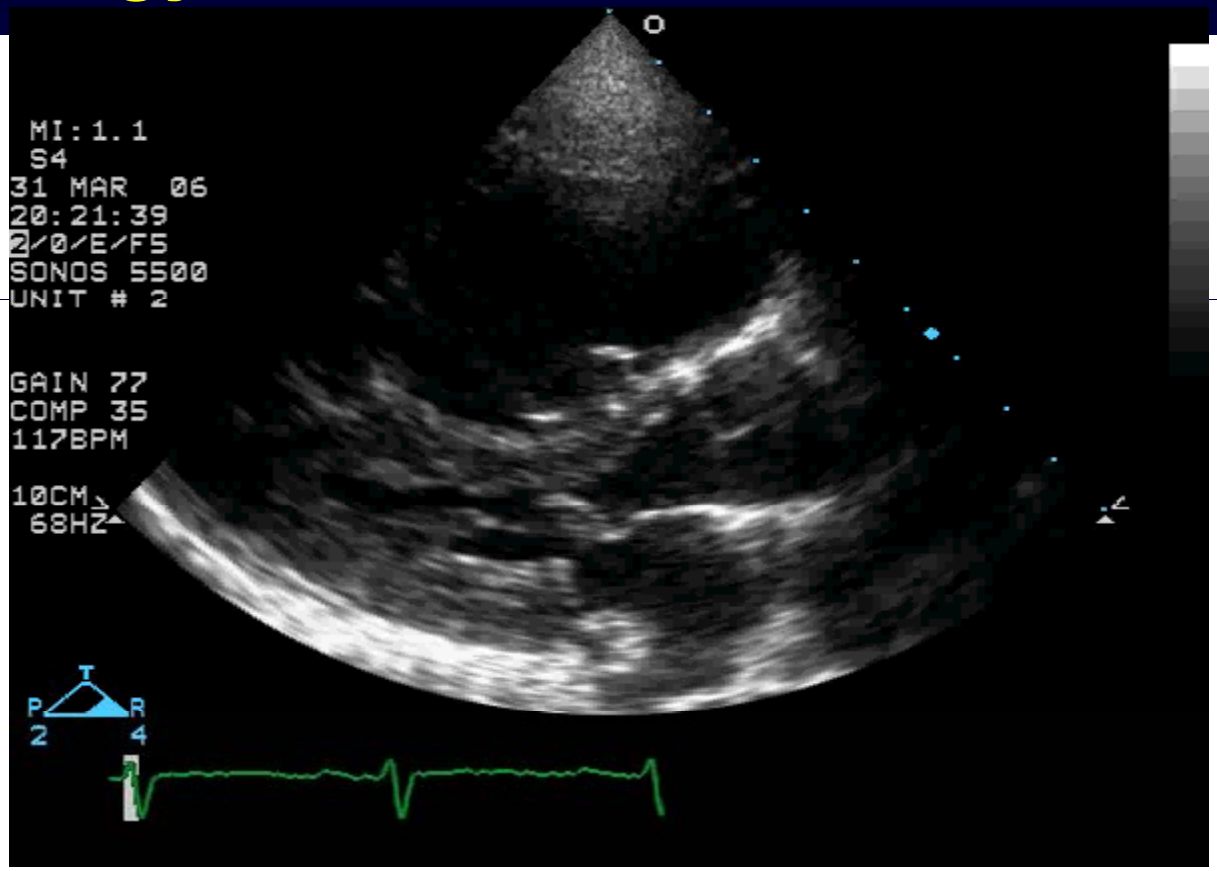
Problems with MPI

- Can’t assess inflow and outflow simultaneously in RV
- Therefore HR has to be similar in both measurements
- ICT is short in the normal RV -> IRT becomes more important (itself an independent index)
- Doesn’t tell you what is wrong


PHILIPS
TIS0.2 MI 1.2
S8-3/HSC2
FR 39Hz
8.1cm
2D
56%
C 50
P Off
HGen
M3
P
P
R
2.6
5.2
JPEG
100 bpm

PHT: A BIVENTRICULAR DISEASE

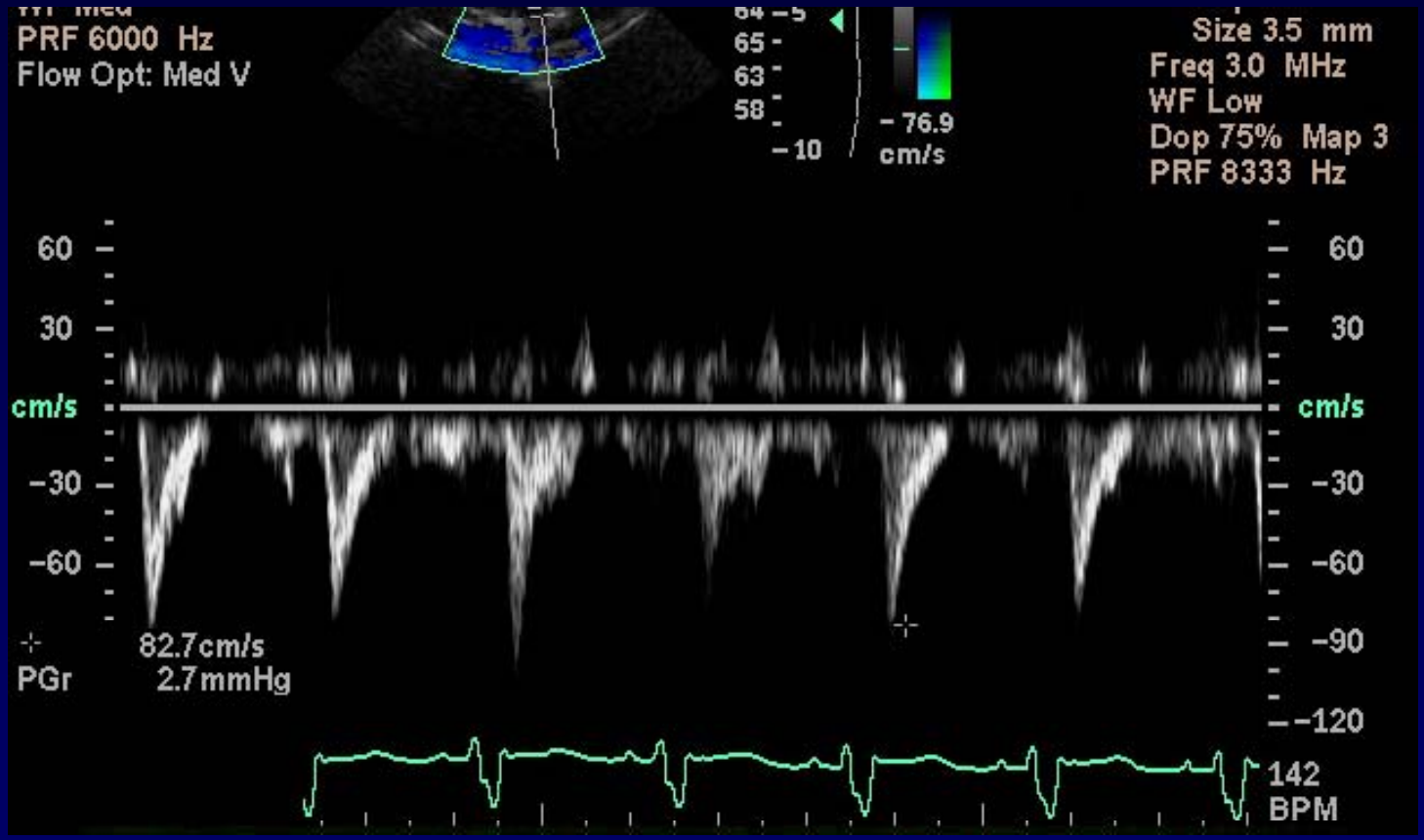
Physiology



Symptomatic deterioration when PVR > SVR

Courtesy Dr Andrew Redington

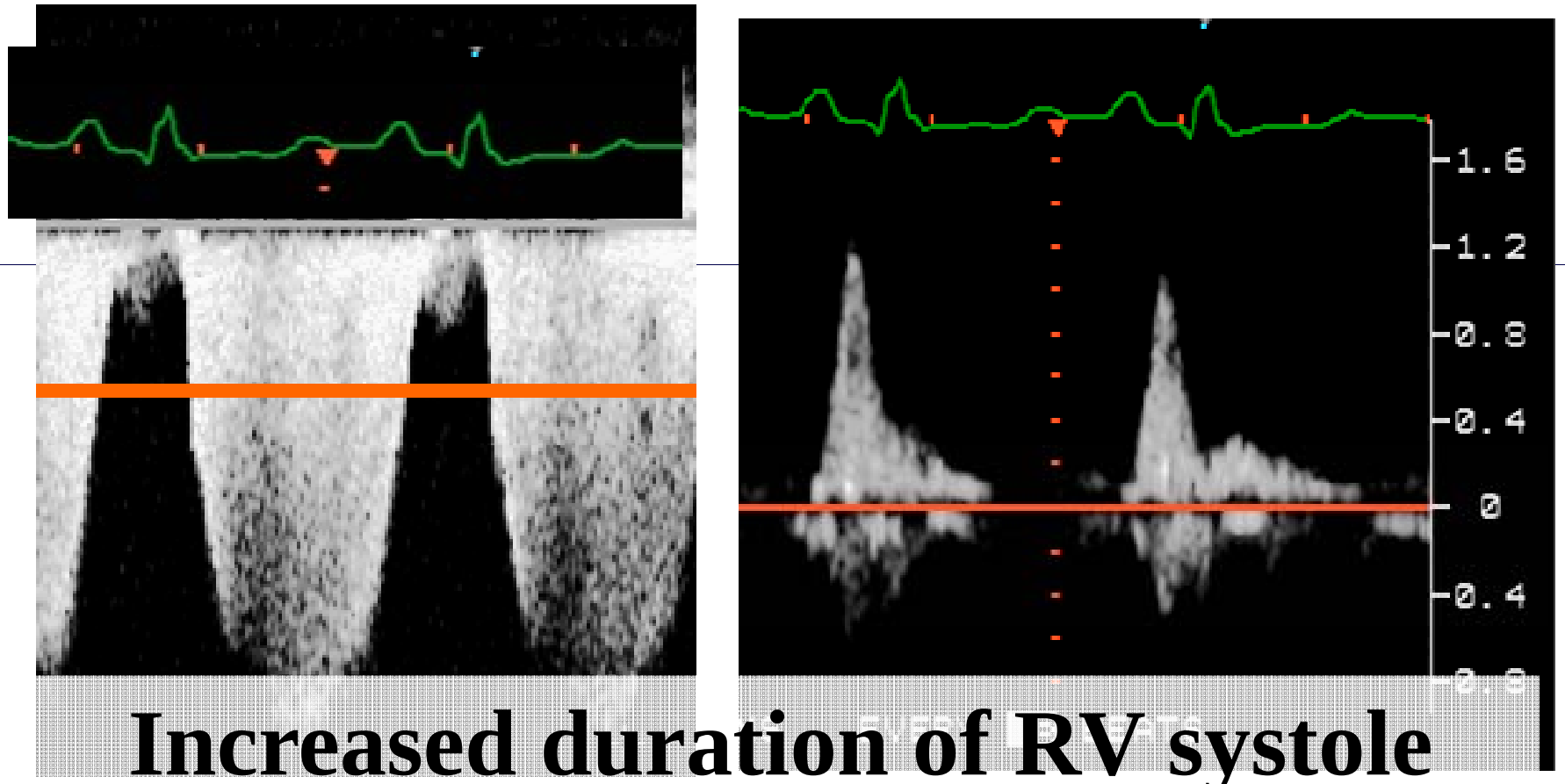
RV ejection


PRF 6000 Hz
Flow Opt: Med V
65
63
58
−10
− 76.9
cm/s
Size 3.5 mm
Freq 3.0 MHz
WF Low
Dop 75% Map 3
PRF 8333 Hz
60
30
cm/s
−30
−60
−90
−120
82.7cm/s
2.7mmHg
PGr
142
BPM

Ventricular interactions

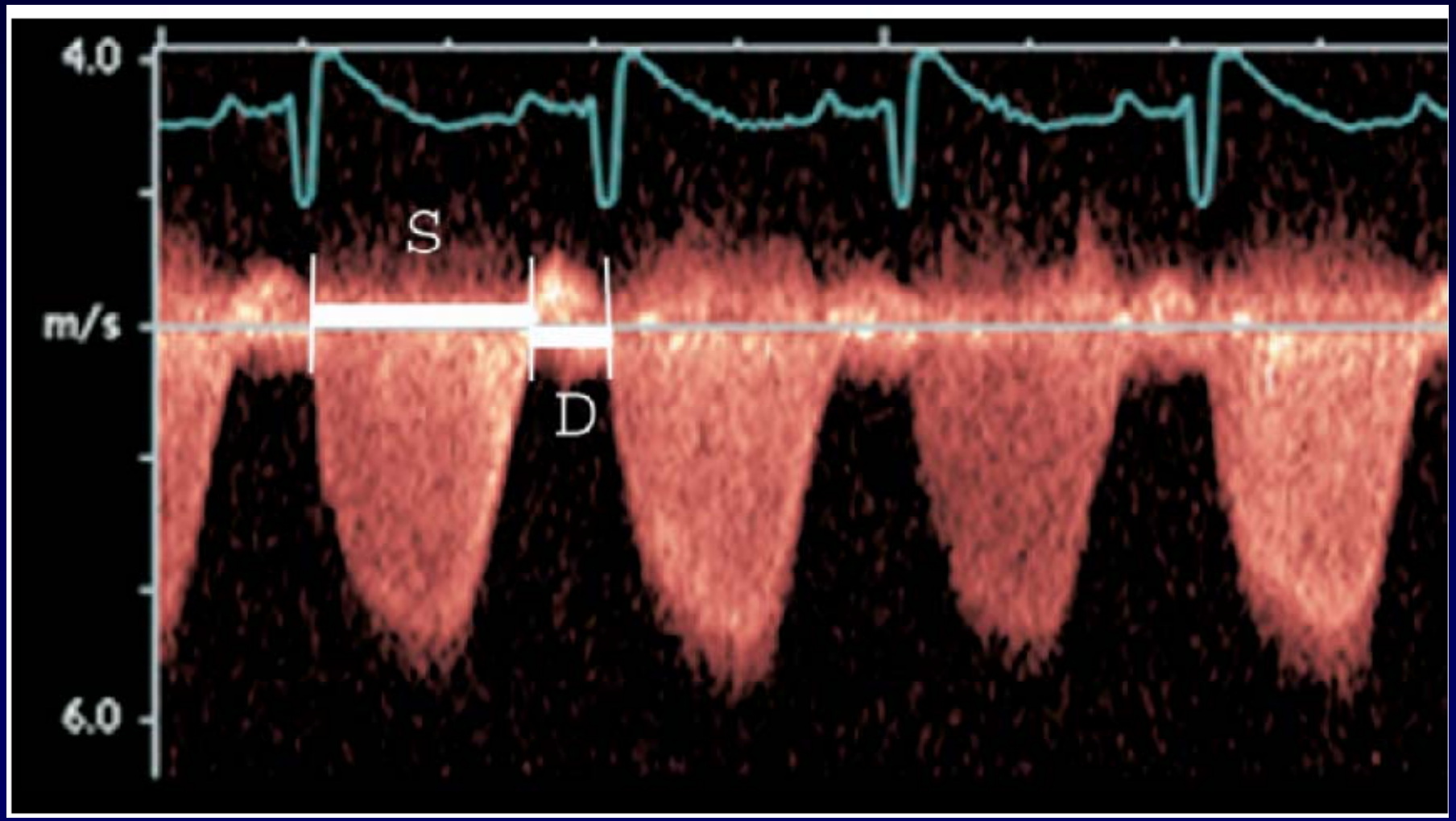
Tricuspid valve

Mitral inflow



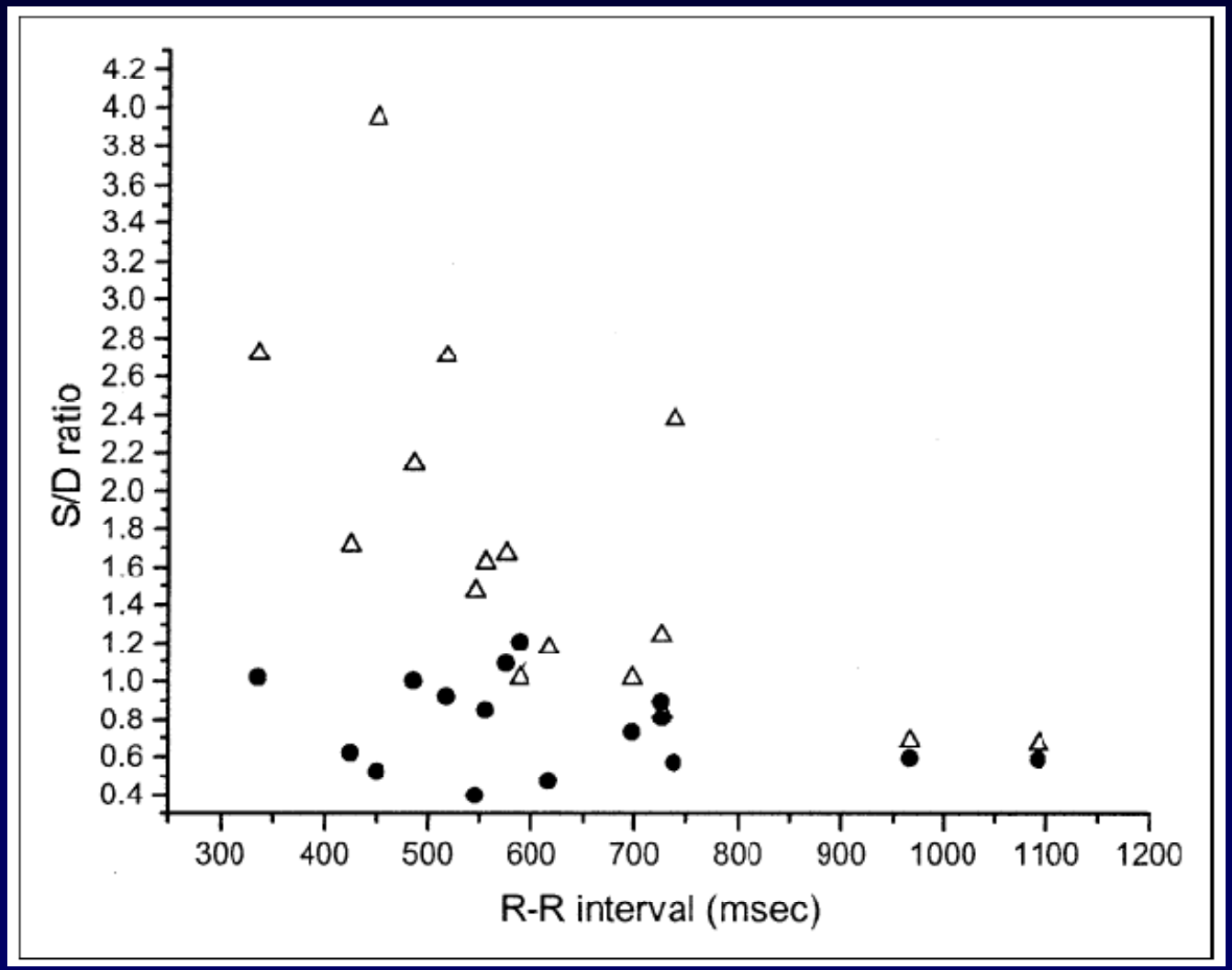
Increased duration of RV systole leads to reduced LV preload

The S/D Duration ratio



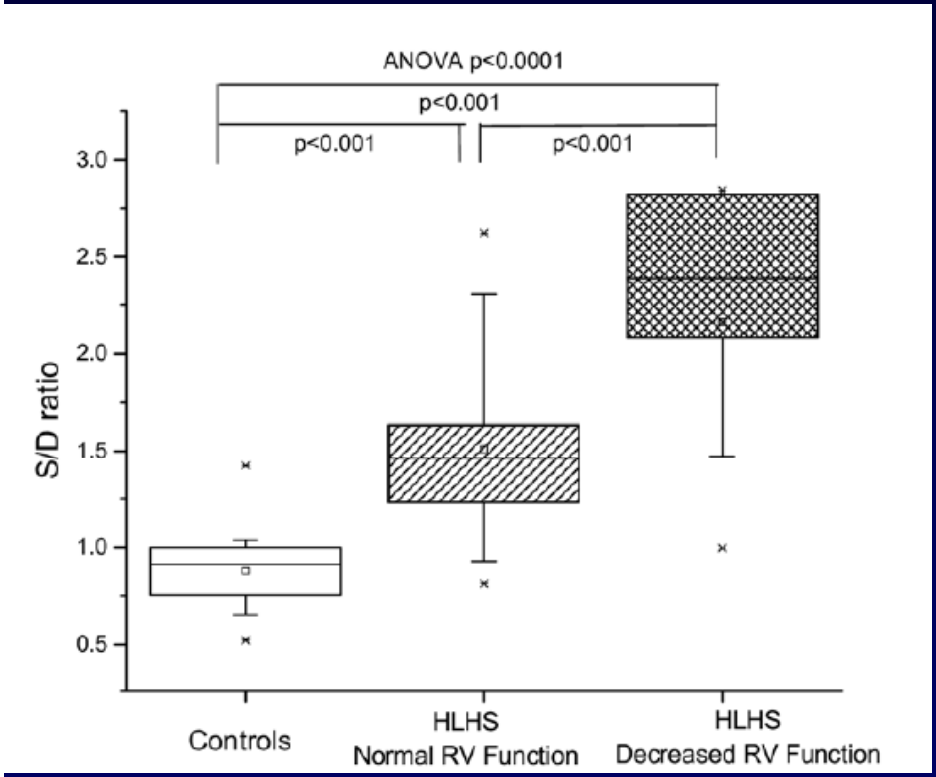
Friedberg, JASE, 2006;19:1326

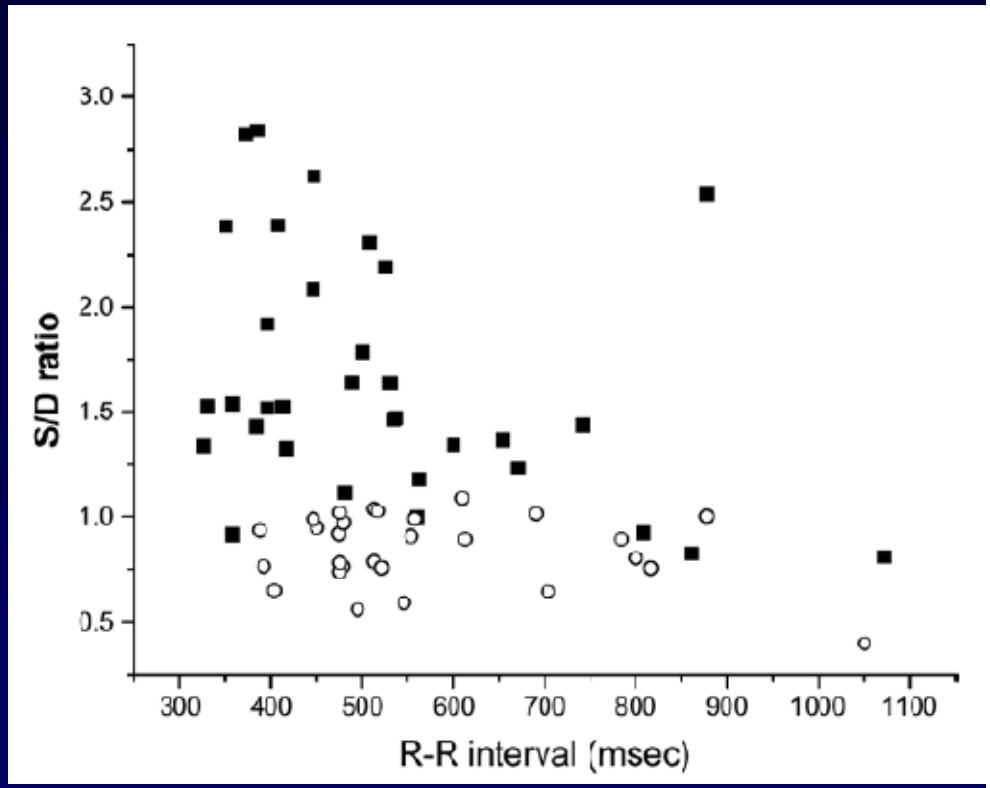
The S/D ratio: Dilated Cardiomyopathy



Friedberg, Am J Cardiol 2006;97:101–105

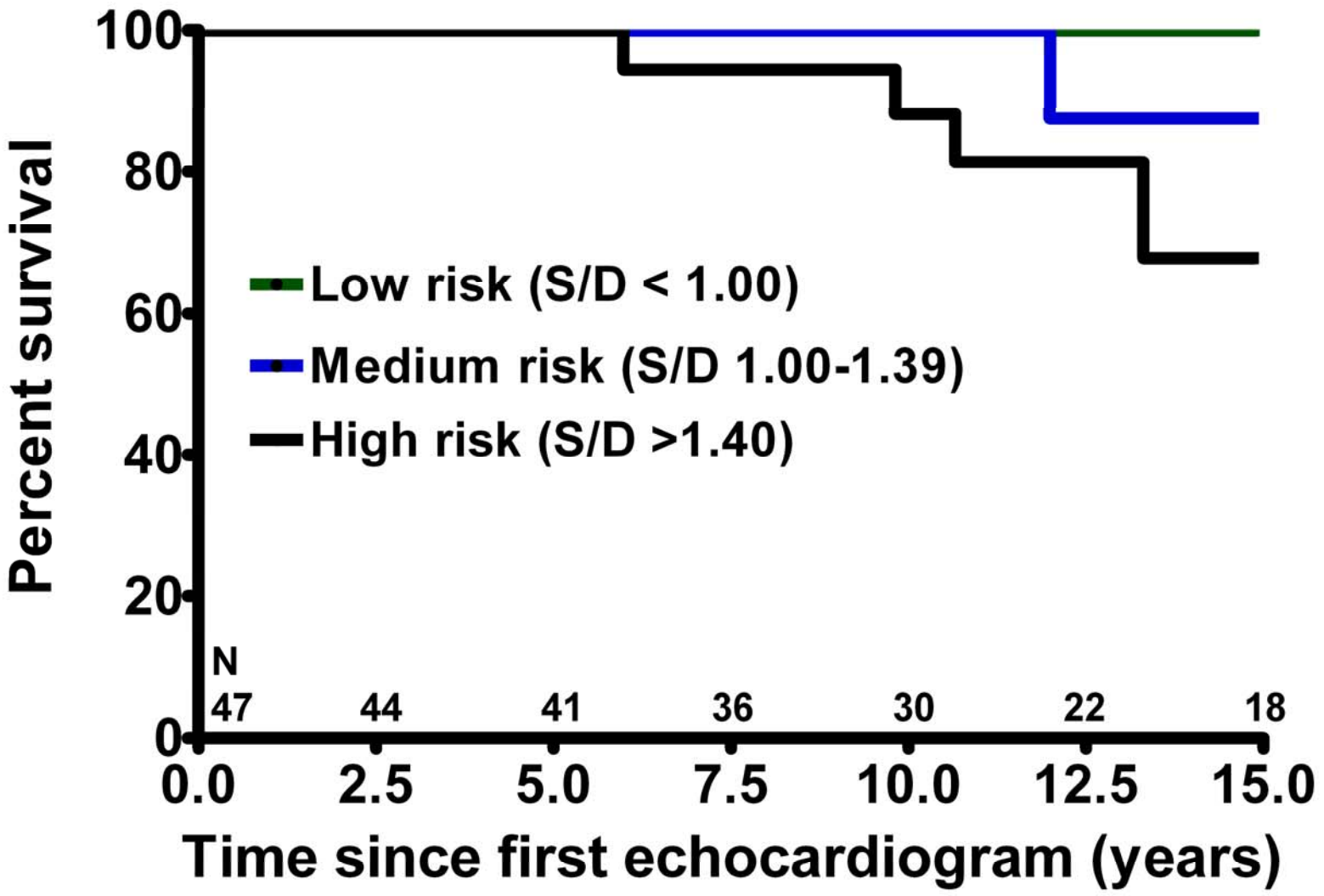
SD ratio in the RV



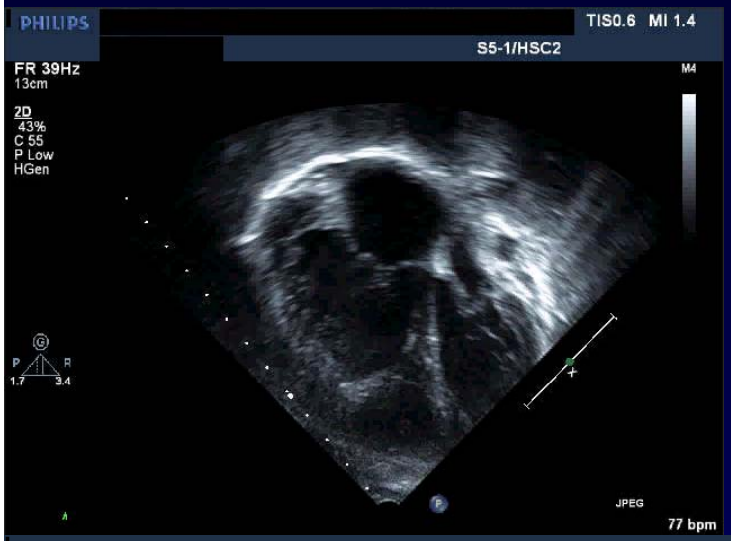


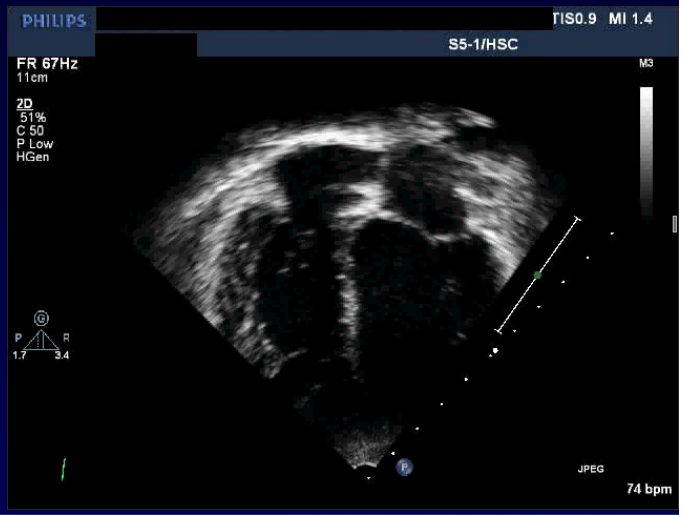
Friedberg, J Am Soc Echocardiogr 2007;20:749

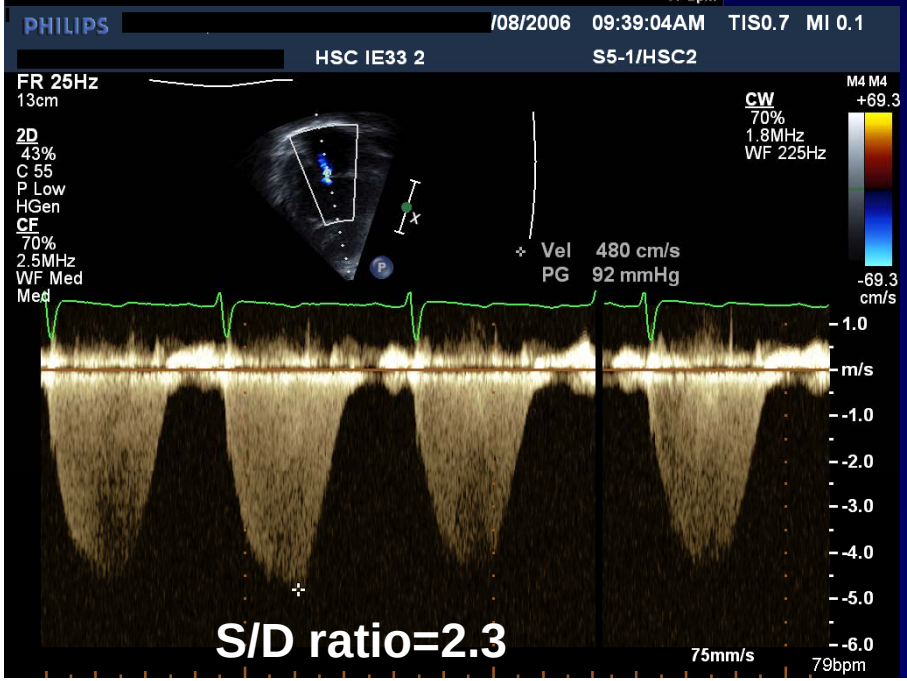
SD ratio in PHTN

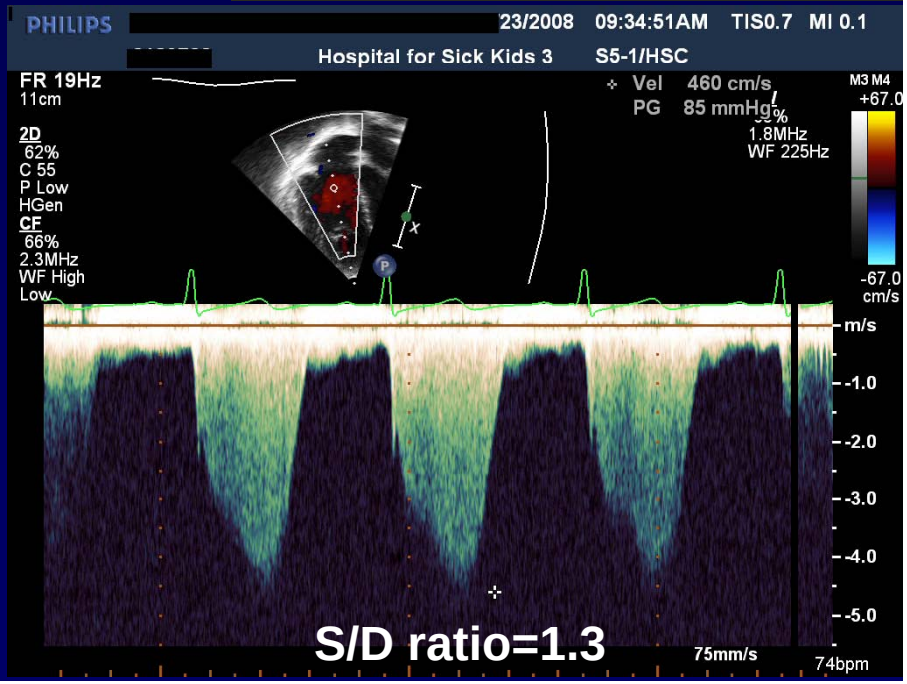


Alkon, AJC 2010;106:430


PHILIPS
TIS0.6 MI 1.4
S5-1/HSC2
FR 39Hz
13cm
2D
43%
C 55
P Low
HGen
M4
JPEG
77 bpm


PHILIPS
TIS0.9 MI 1.4
S5-1/HSC
FR 67Hz
11cm
2D
51%
C 50
P Low
HGen
M3
JPEG
74 bpm


PHILIPS
/08/2006 09:39:04AM TIS0.7 MI 0.1
HSC IE33 2
S5-1/HSC2
FR 25Hz
13cm
2D
43%
C 55
P Low
HGen
CF
70%
2.5MHz
WF Med
Med
CW
70%
1.8MHz
WF 225Hz
M4 M4
+69.3
-69.3
cm/s
Vel 480 cm/s
PG 92 mmHg
1.0
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
-6.0
S/D ratio=2.3
75mm/s
79bpm


PHILIPS
23/2008 09:34:51AM TIS0.7 MI 0.1
Hospital for Sick Kids 3
S5-1/HSC
FR 19Hz
11cm
2D
62%
C 55
P Low
HGen
CF
66%
2.3MHz
WF High
Low
Vel 460 cm/s
PG 85 mmHg
1.8MHz
WF 225Hz
M3 M4
+67.0
-67.0
cm/s
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
S/D ratio=1.3
75mm/s
74bpm

Summary

- Assessment of RV function is a central component in the evaluation, management and follow-up of children with CHD and pulmonary vascular disease.
- Assessment of RV function provides prognostic information.
- Echo remains the mainstay of clinical imaging in this condition, but has important limitations.
- Newer methods (based on older and newer technology) can be combined with conventional assessment to provide a more comprehensive evaluation.
- Further validation of the newer methods in clinical practice is needed.

Thank you