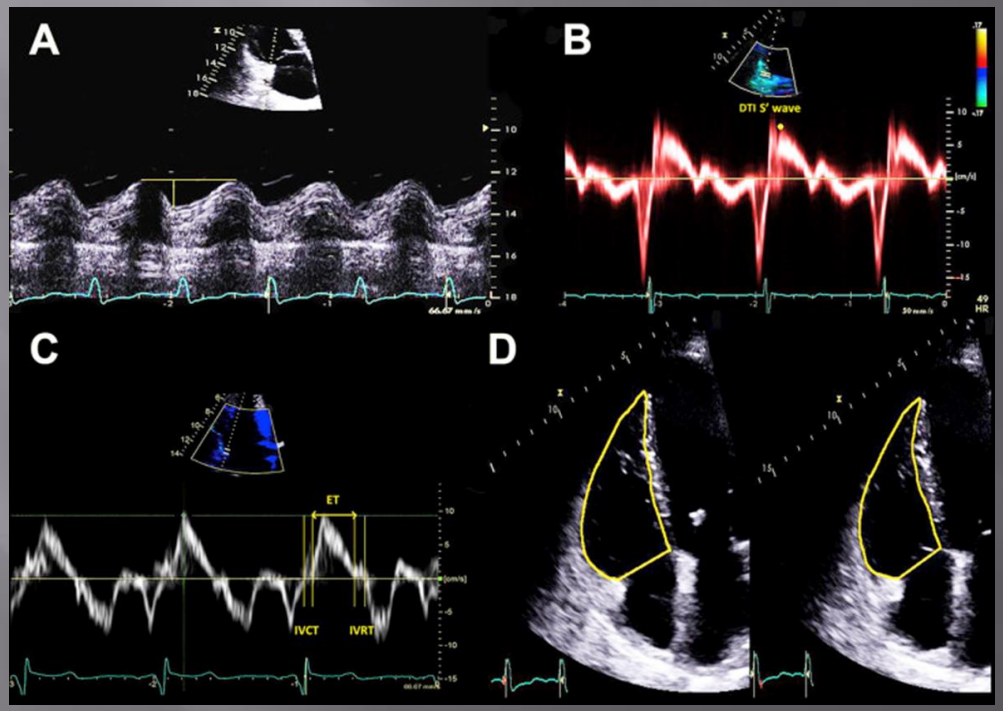
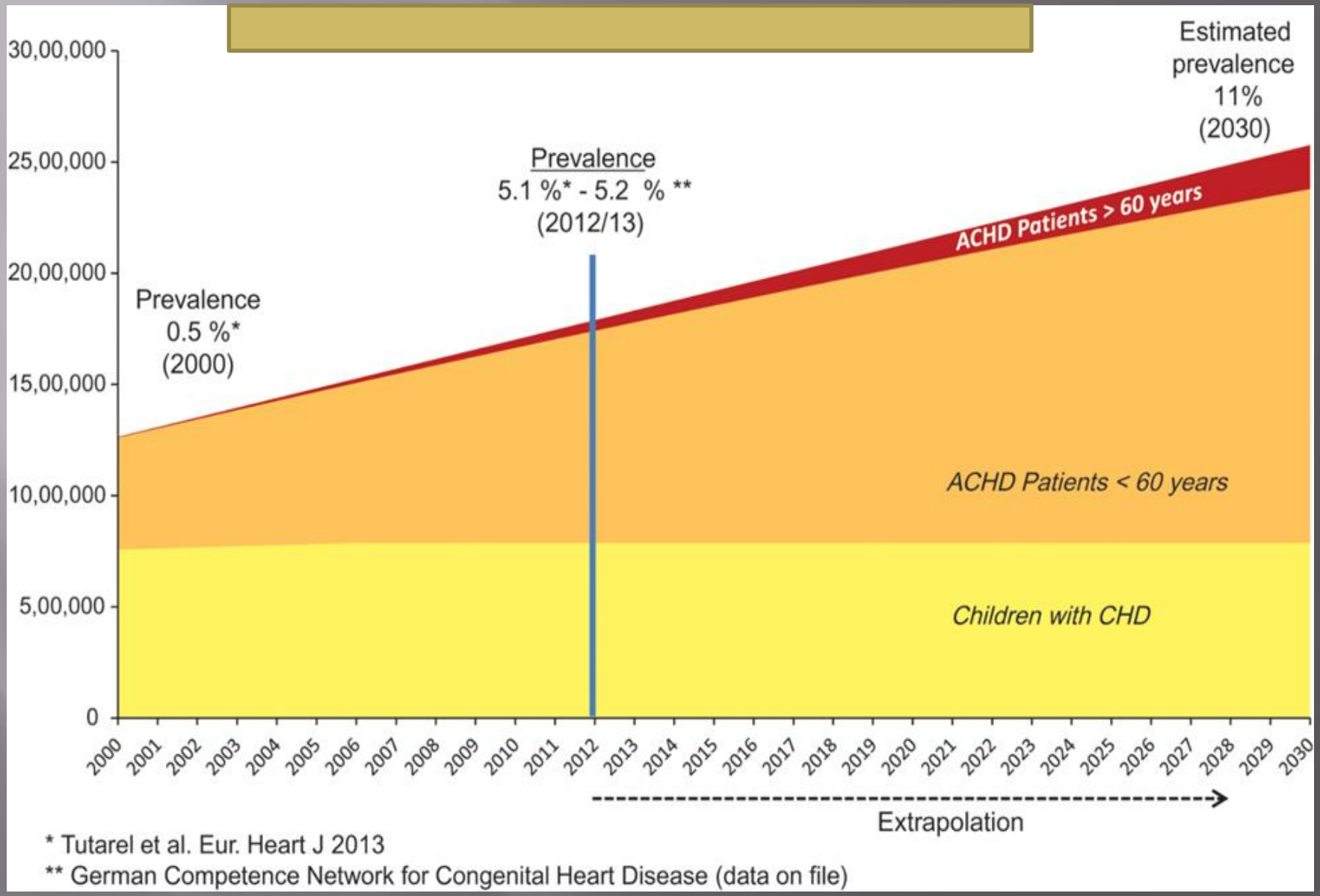
SHADES OF GREY – NOVEL ECHO PARAMETERS OF IMPORTANCE IN THE FAILING CONGENITAL HEART



Ivor Gerber

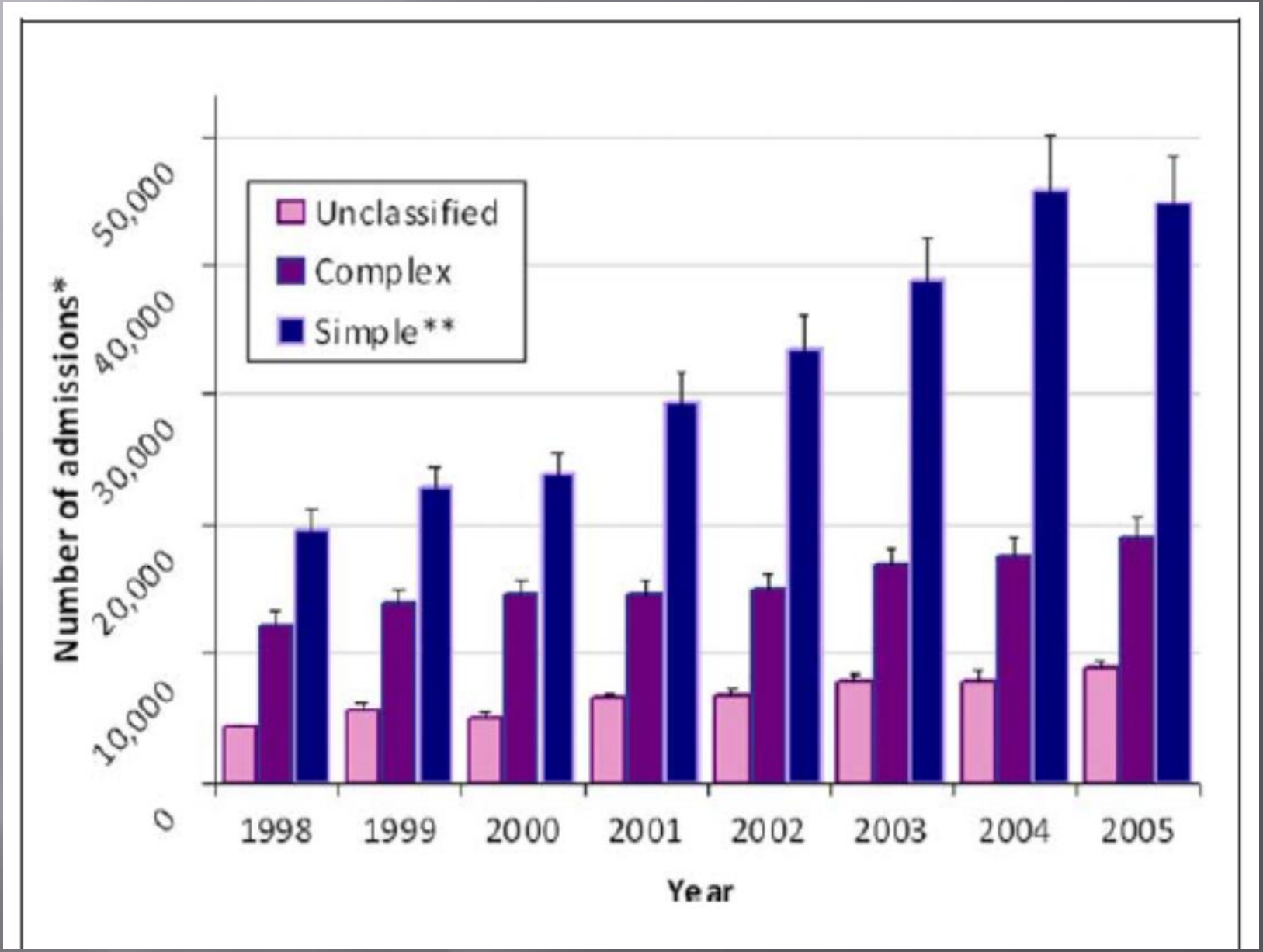
Medical advances 1950-2000

The Adult Population



* Tutarel et al. Eur. Heart J 2013

** German Competence Network for Congenital Heart Disease (data on file)



Opotowsky, et al. JACC. 2009;54:460-7.

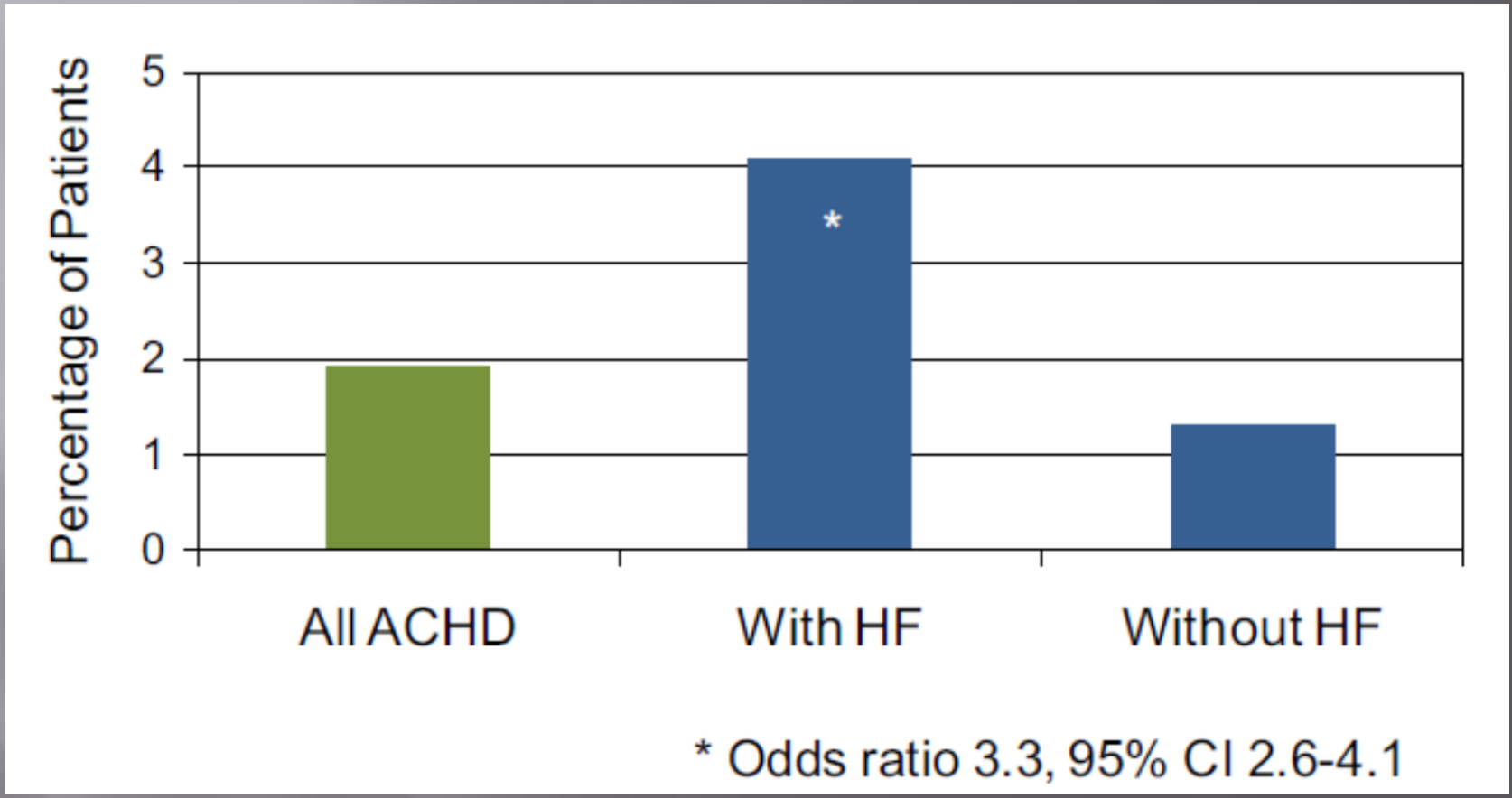
- Many interventions, especially for complex CHD lesions, are palliative rather than curative

Table 3 Frequency of Specific Diagnoses and Procedures Associated With Hospitalizations for ACHD

Year	1998	1999	2000	2001	2002	2003	2004	2005	Total
Diagnoses*									
Arrhythmia	11,742 (1,012)	12,717 (1,047)	13,218 (994)	16,332 (1,261)	17,713 (1,352)	20,174 (1,597)	22,458 (1,896)	24,882 (1,961)	139,237 (5,303)
CAD†	8,574 (745)	9,395 (727)	10,770 (853)	12,843 (1,037)	14,275 (1,105)	16,191 (1,279)	18,440 (1,549)	18,788 (1,455)	109,275 (4,235)
Heart failure	7,453 (624)	8,168 (588)	8,389 (584)	9,628 (649)	10,427 (793)	12,1 (908)	12,939 (1,064)	13,604 (1,008)	83,130 (2,905)
Pulmonary hypertension	3,923 (294)	4,747 (402)	4,774 (372)	4,989 (383)	5,617 (446)	6,284 (506)	6,652 (652)	6,528 (525)	43,516 (1,702)
Pregnancy	1,706 (162)	2,217 (204)	2,149 (202)	2,036 (179)	2,568 (226)	2,669 (235)	2,990 (270)	3,431 (312)	19,765 (822)
Bacterial endocarditis	574 (71)	648 (80)	578 (69)	765 (79)	802 (89)	802 (90)	1,103 (116)	867 (99)	6,138 (296)
Procedures									
Percutaneous ASD/PFO closure	134 (90)	165 (59)	308 (133)	627 (150)	2,205 (662)	3,517 (1,008)	4,415 (1,043)	3,219 (566)	14,589 (2,413)
Pacemaker	974 (150)	1,332 (145)	1,310 (148)	1,427 (52)	1,638 (198)	1,798 (191)	2,021 (208)	2,188 (235)	12,687 (674)
PCI	830 (103)	806 (103)	1,065 (136)	1,130 (130)	1,405 (155)	1,832 (198)	1,967 (247)	2,227 (239)	11,262 (633)
ICD	208 (45)	368 (69)	326 (52)	602 (85)	570 (81)	662 (88)	901 (130)	840 (109)	4,476 (312)

Opotowsky, et al. JACC. 2009;54:460-7.

- Heart failure is the leading cause of death in adults with CHD.
- 1 year mortality rate of 24% after first heart failure admission



Rodriguez et al. Congenital heart disease. 2012.

Independent risk factors for first HF-admission in adulthood.

	HR	95% CI
Patient characteristics		
Multiple defects	2.2	1.7–2.9
Main defect		
Ventricular septal defect	–	–
Atrial septal defect	1.1	0.7–1.7
Aortic coarctation	0.4	0.2–0.9
Tetrology of Fallot	2.1	1.3–3.6
Aortic stenosis	1.9	1.0–3.4
Pulmonary stenosis	0.6	0.3–1.4
Bicuspid aortic valve	0.7	0.4–1.5
AVSD	2.7	1.5–5.1
Marfan syndrome	0.8	0.3–2.2
TGA	5.0	2.5–9.9
Patent arterial duct	0.6	0.2–1.7
Ebstein malformation	0.7	0.2–2.2
ccTGA	5.2	1.8–15.4
PA + VSD	3.0	1.2–7.4
FUH/DILV	11.4	5.9–22.0
Other	3.1	2.2–6.1
Interventions in childhood		
Surgery	2.5	1.8–3.5
Reoperation	1.8	1.2–2.6
Pacemaker implantation	3.1	1.5–6.3

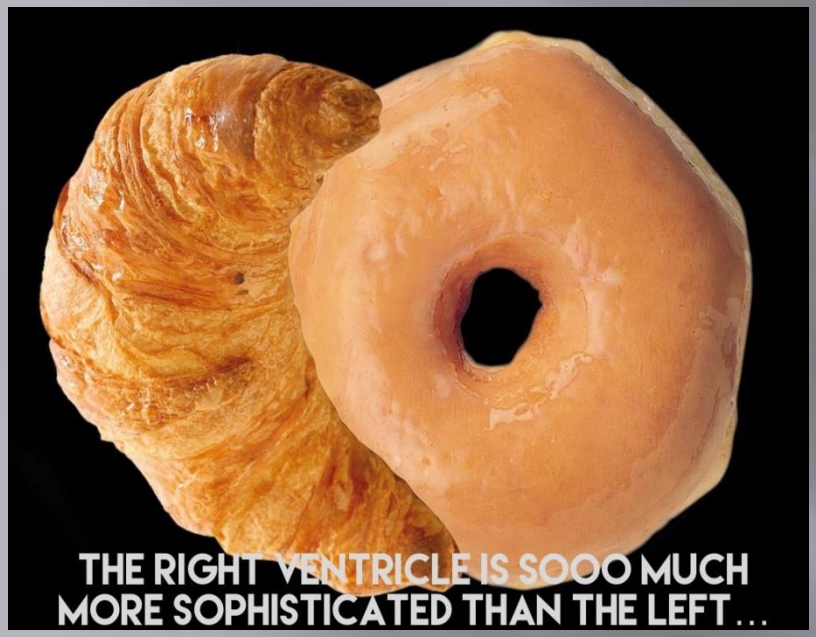
Zomer et al. Heart failure admissions in adults with congenital heart disease

Causes of heart failure in patients with CHD

- Volume overload
  - Left to right shunts
  - Valvular regurgitation
- Pressure overload
  - Valvular disease
  - Other obstructive lesions such as aortic coarctation
- Intrinsic myocardial dysfunction
  - Single ventricle
  - Systemic right ventricle
  - Previous cardiopulmonary bypass
  - Myocardial fibrosis
- Pulmonary hypertension
- Systemic arterial hypertension
- Coronary artery disease
- Cyanosis
- Intractable atrial arrhythmias
- Pericardial constriction

Heart failure in ACHD – which ventricle?

- **In the non-ACHD patient, heart failure usually reflects the failing left ventricle**
  - Ischaemic cardiomyopathy
  - Dilated cardiomyopathy
  - Valvular abnormalities
- **Many treatment options that focus on the left ventricle**
  - Medical therapy
  - CABG
  - Valve replacement
- **The right ventricle is considered less often – why?**
  - Less common
  - Less data
  - Less treatment options
  - More difficult to assess

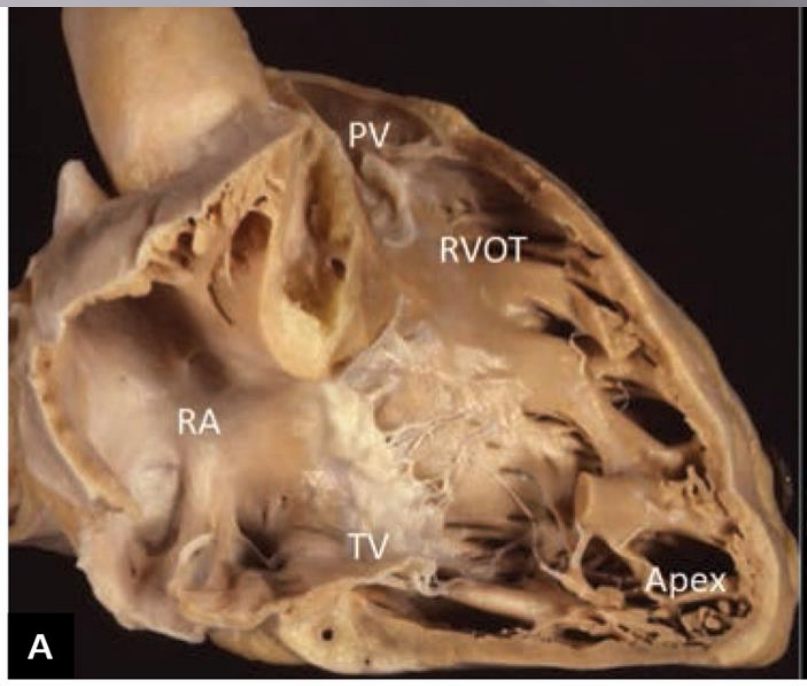


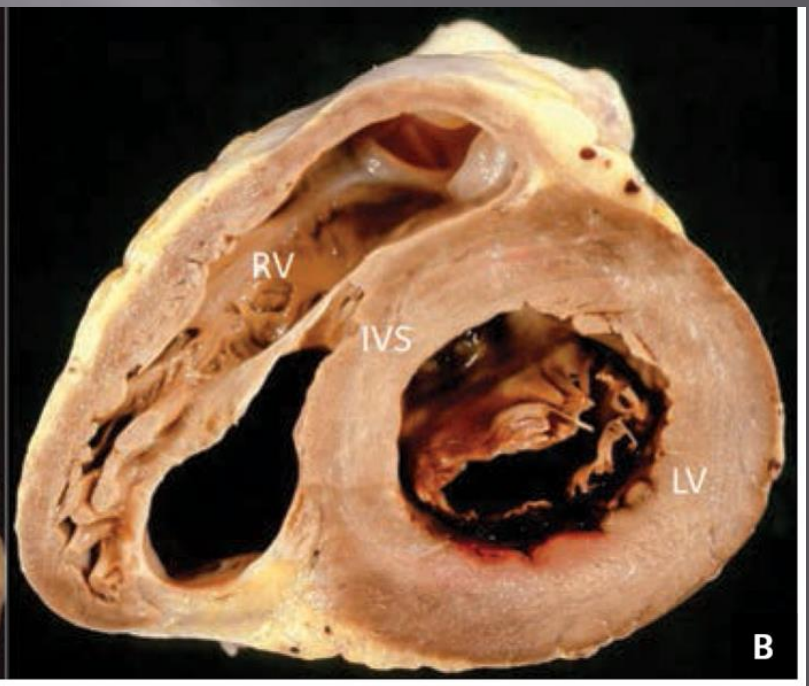
- Very anterior position
- Complex geometric shape
- Prominent trabeculations
- Muscle fibres are arranged mainly longitudinally

- RV contraction
  - Bellow like motion of the free wall towards the septum
  - Longitudinal motion of the base toward the apex
  - Bulging of the septum into the RV cavity

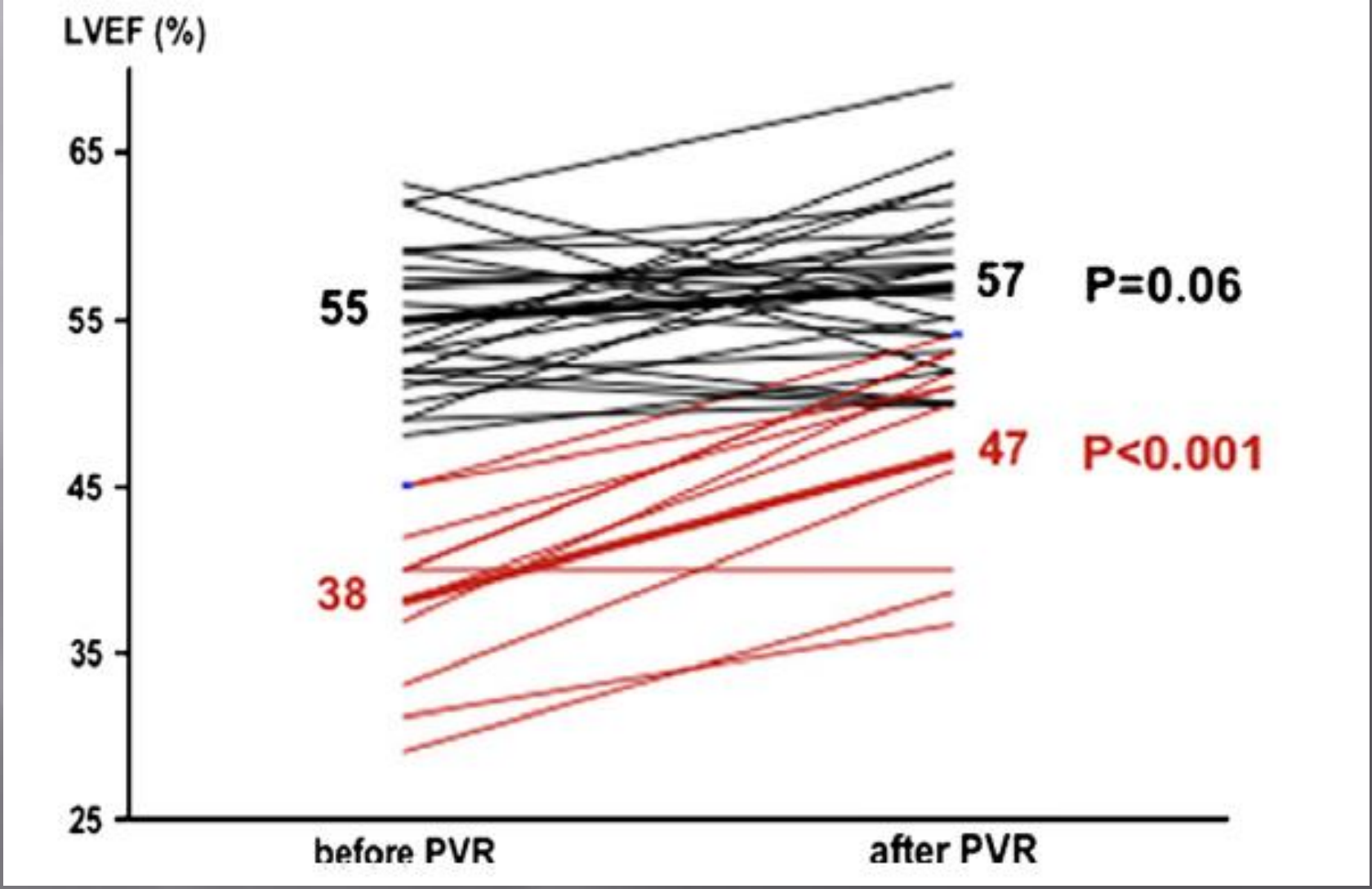
  - In disease states, RV shifts from mainly longitudinal contraction

  - Proper assessment should include various measures



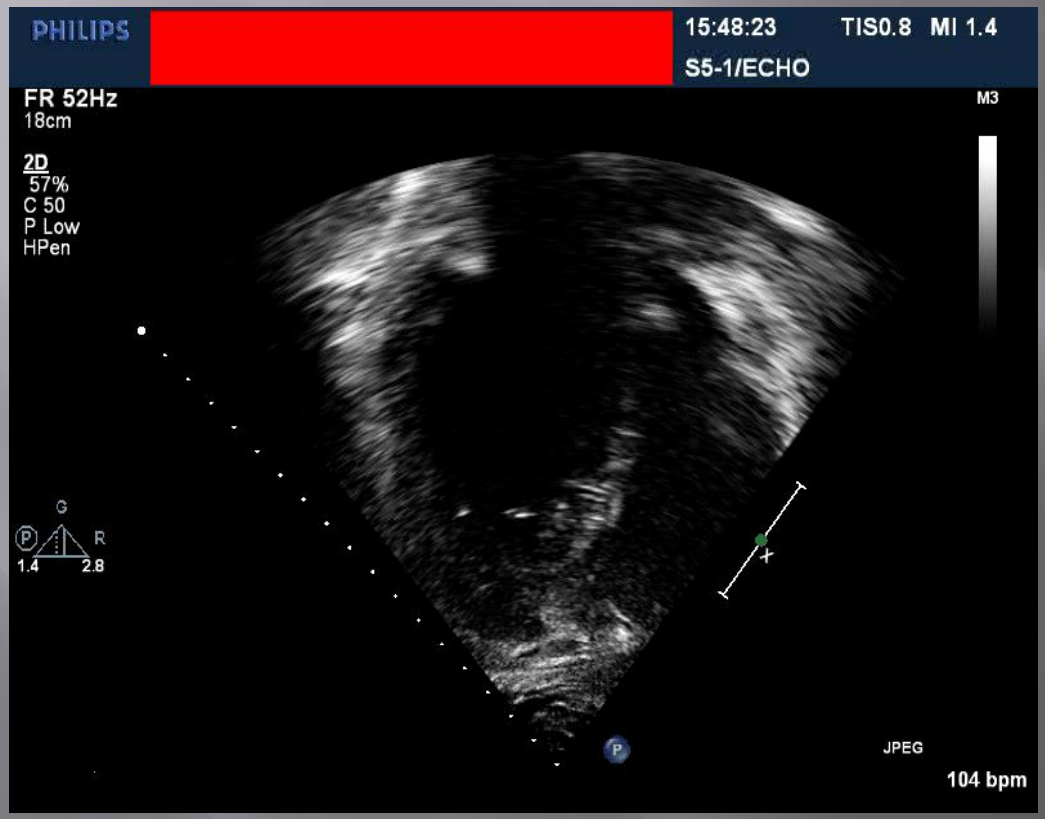


Ventricular interdependence



Echocardiographic assessment of the failing ventricle

Qualitative assessment – "eyeball method"

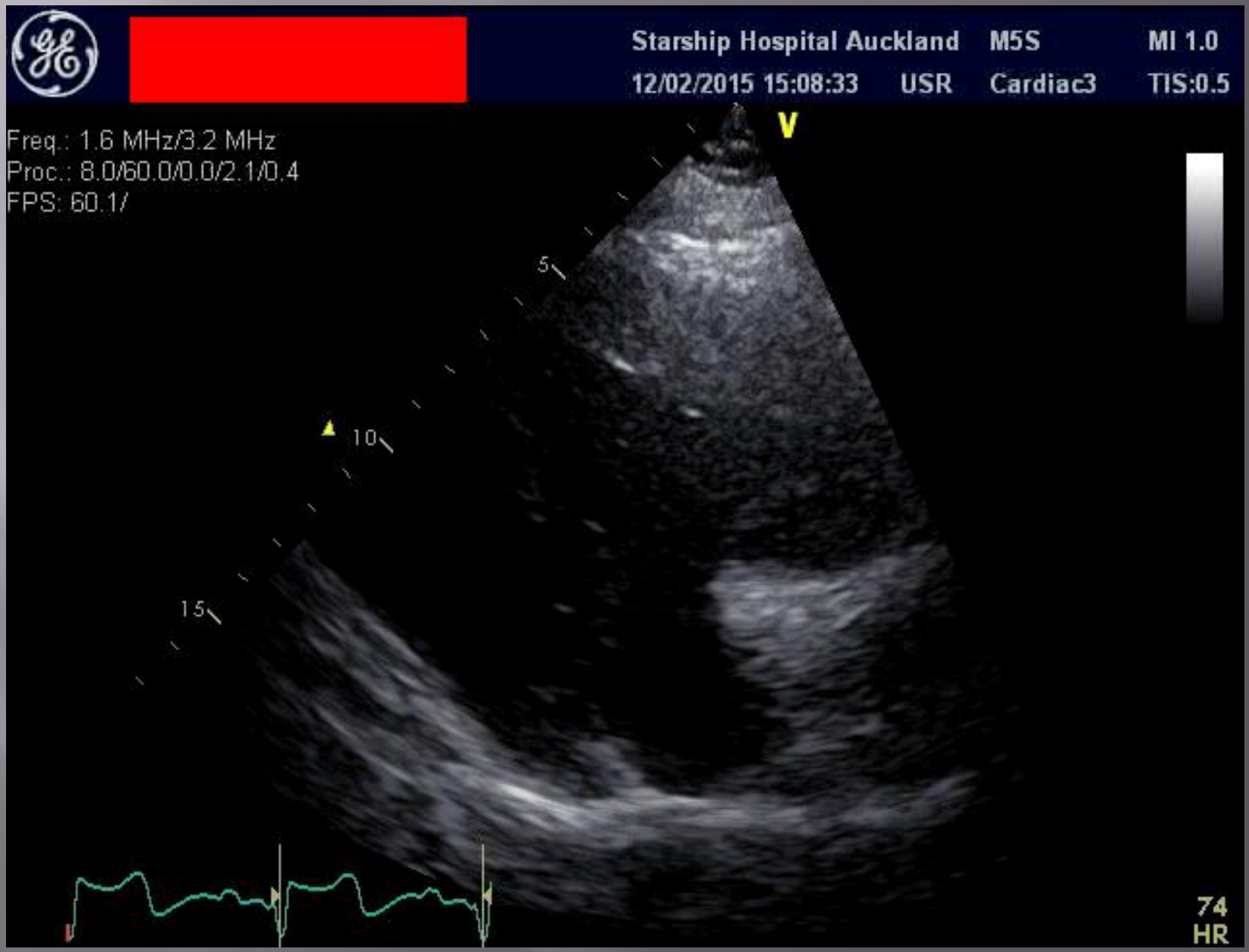


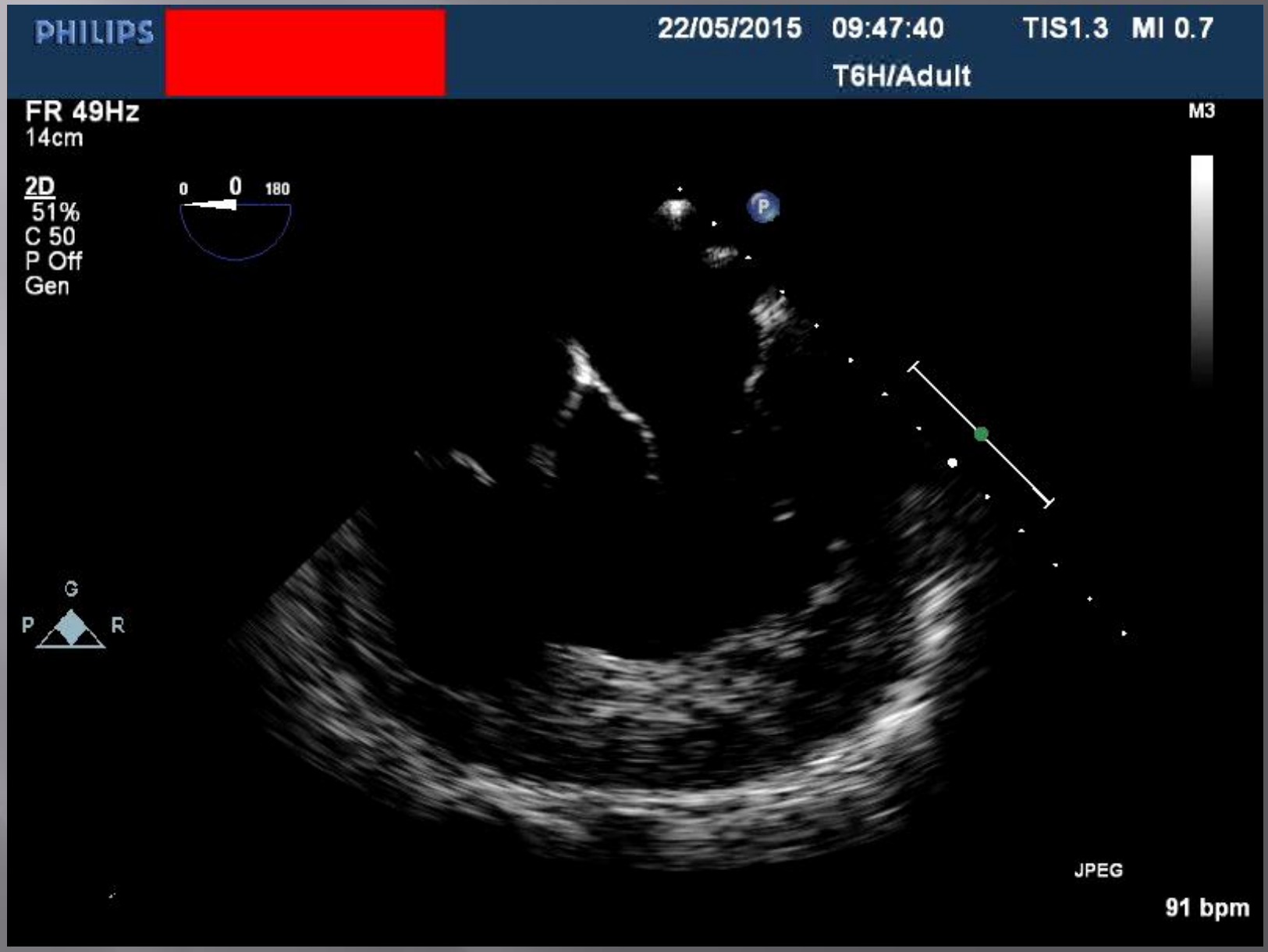
Most frequent method in routine clinical practice

Mild, moderate, severe

Poor inter-observer agreement

Best used in conjunction with other measures


Starship Hospital Auckland M5S MI 1.0
12/02/2015 15:08:33 USR Cardiac3 TIS:0.5
Freq.: 1.6 MHz/3.2 MHz
Proc.: 8.0/60.0/0.0/2.1/0.4
FPS: 60.1/
5
10
15
74
HR


PHILIPS
[redacted]
22/05/2015 09:47:40
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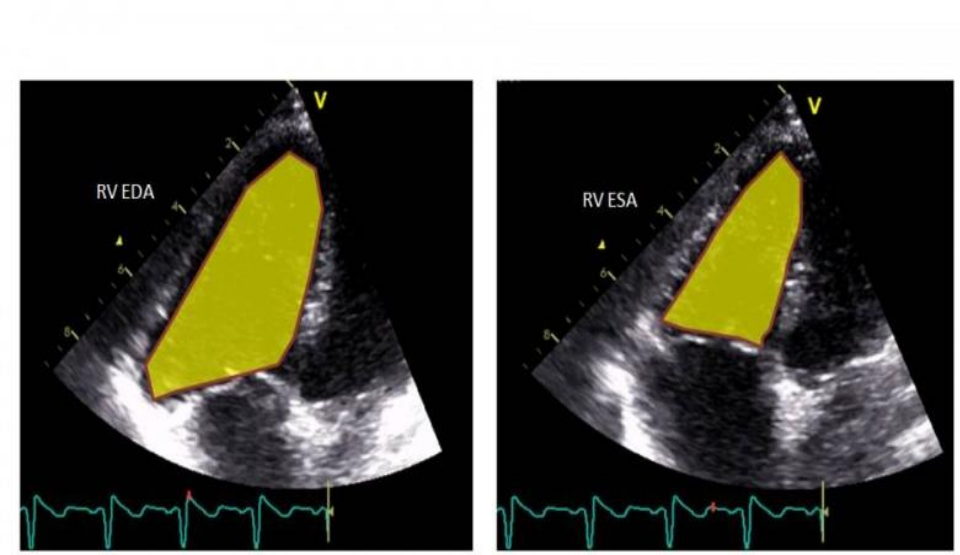
FR 49Hz
14cm
M3
2D
51%
C 50
P Off
Gen
0 0 180
P
G
P R
JPEG
91 bpm

Fractional area change (FAC)

- 2D measure of global RV systolic function
- Encompasses both longitudinal and radial components of RV contraction
- FAC > 35% is considered normal in adults

- Predictor of outcome in PAH
- Can predict RV function after PVR

- Main limitation:
  - Image quality
  - Does not include RVOT, therefore limitation in TOF

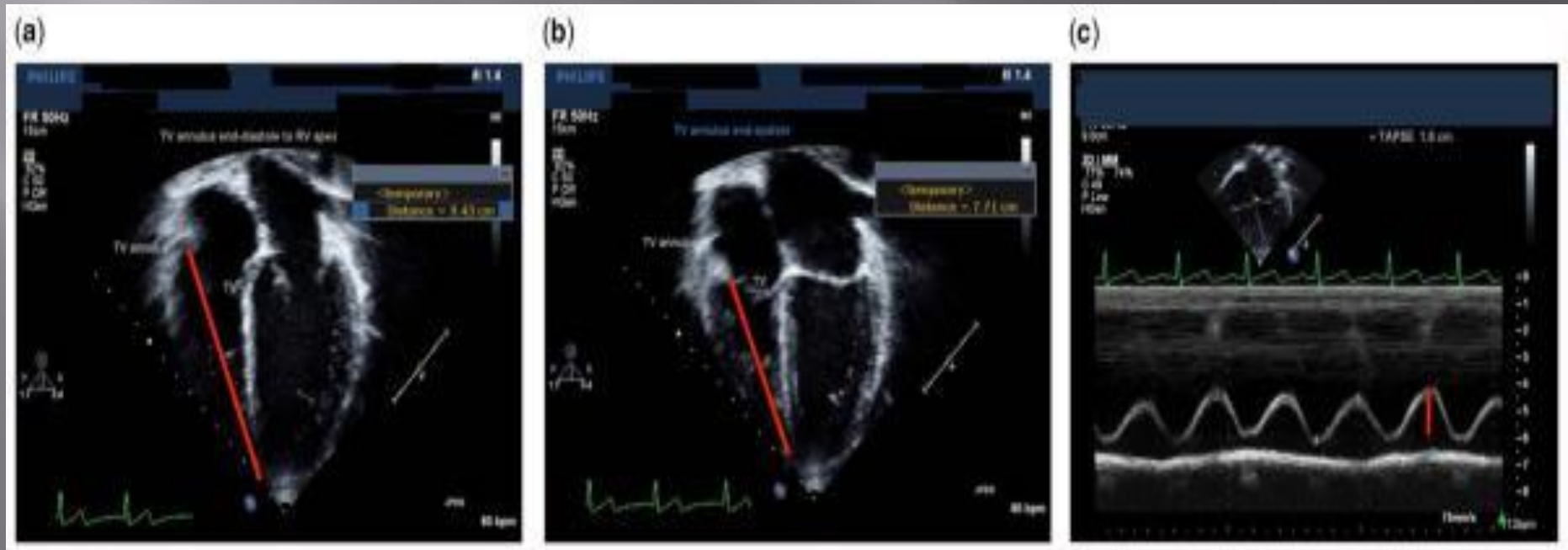


RV FAC % = (RV EDA– RV ESA) / RV EDA x100

Tricuspid Annular Plane Systolic Excursion (TAPSE)

- Measure of RV longitudinal shortening – normal is >16mm.
- Easy to obtain, reproducible

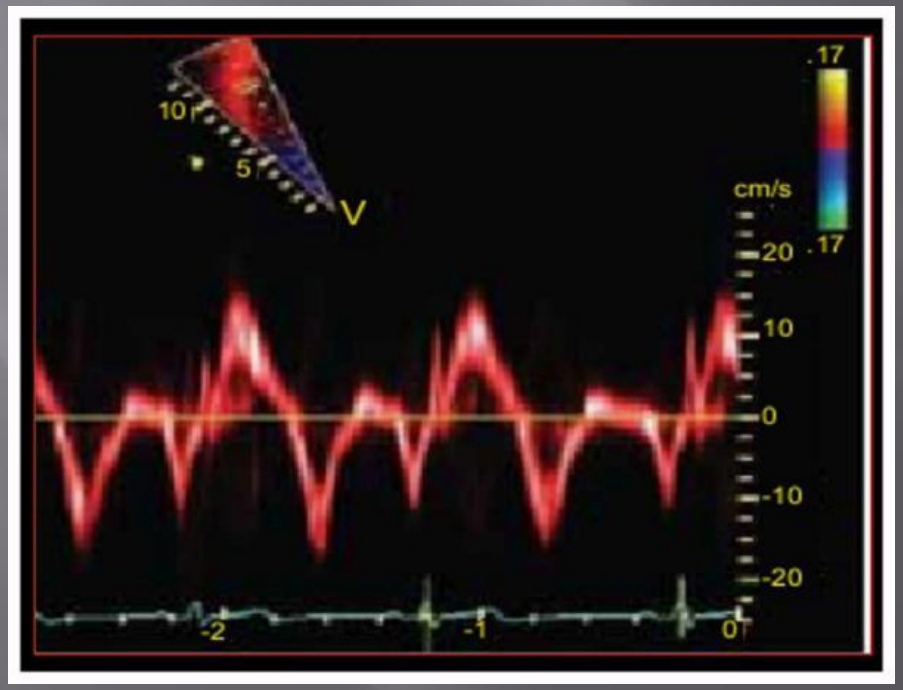
- Angle and load dependent
- Does not take into account the ventricular septum and/or RVOT
- Reflects the function of the basal RV free wall and assumes this represents global RV systolic function



Tissue Doppler-derived RV systolic excursion velocity S'

- Longitudinal RV systolic function
- Reproducible and easily obtainable
- Normal is >10cm
- <11.5 has a high sensitivity and specificity for RVEF <45%
- S' is reduced and inversely associated with RVEDVi in TOF

- Limitations as per TAPSE

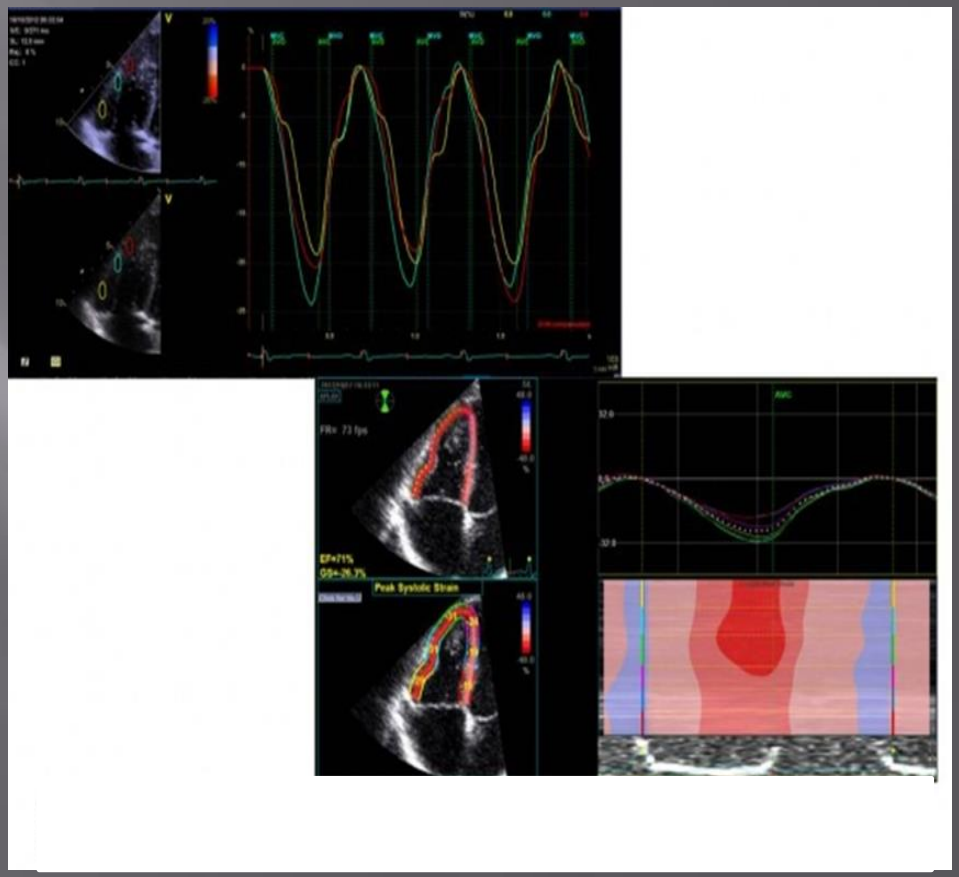


Speckle tracking – strain and strain rate

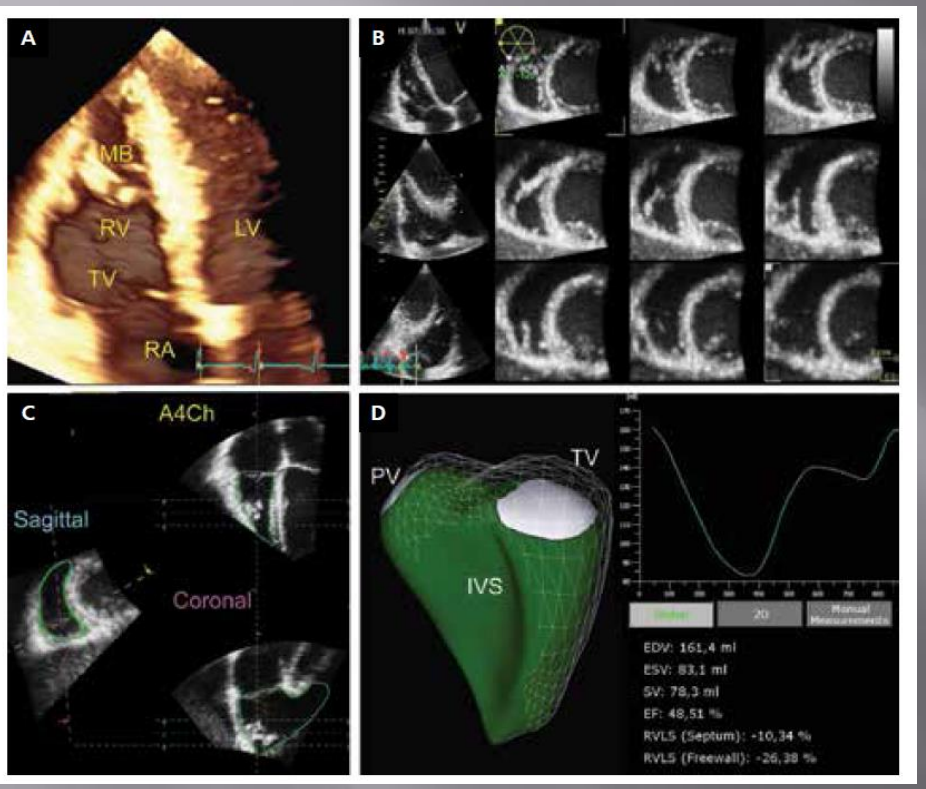
Novel technique to quantitate myocardial contractility

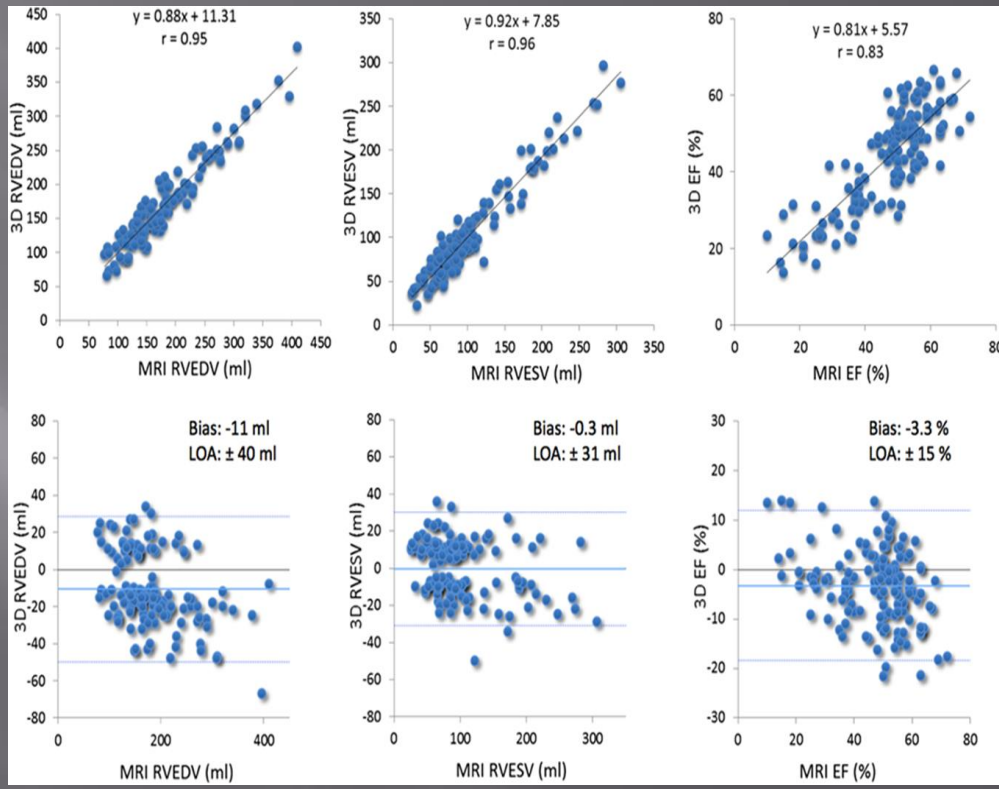
- Reproducible and easy to obtain
- Correlates with RVEF by MRI
- Capable of detecting subclinical RV dysfunction.
- Decline in strain precedes decline in RVEF

- Load dependent
- Developed for the LV and adapted for the RV later – less homogenous with highest values at the apex and RVOT
- Limited to the 4-chamber view
- Normative values are lacking
- May be useful for individual patient follow-up



3-D echocardiography



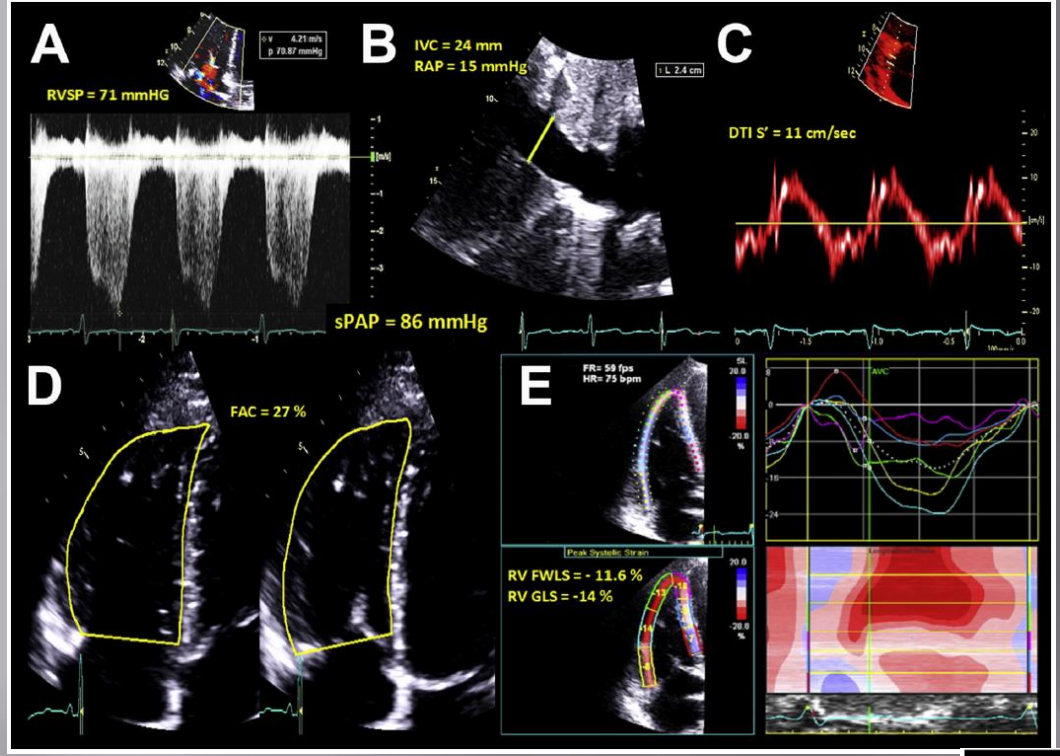


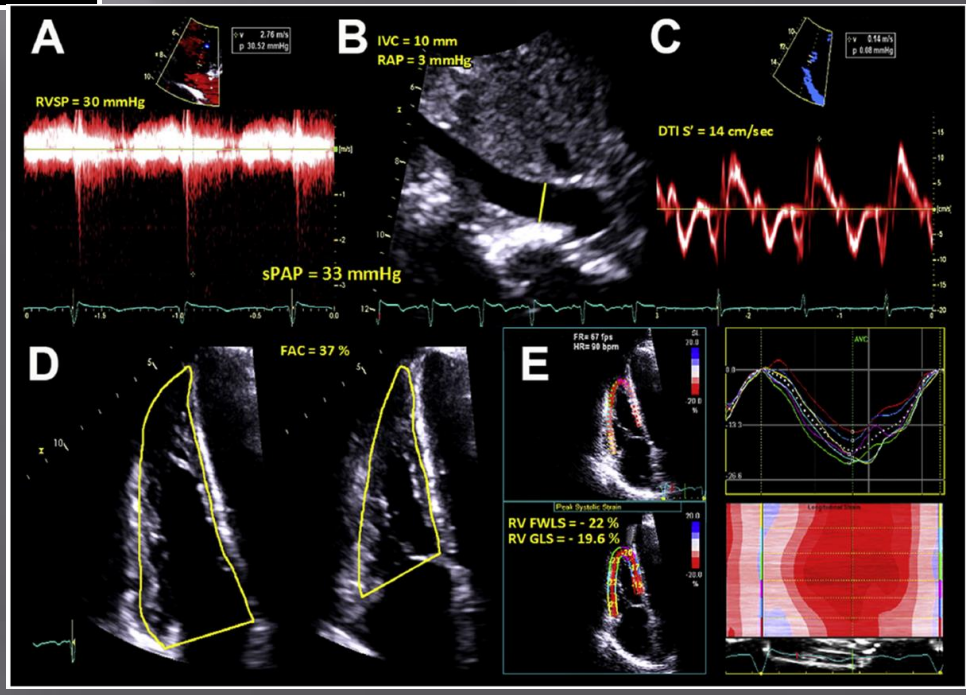
Medvedofsky et al. J Am Soc Echocardiogr 2015

Table 1 Correlation (*r*) between echocardiographic parameters and RVEF by CMR

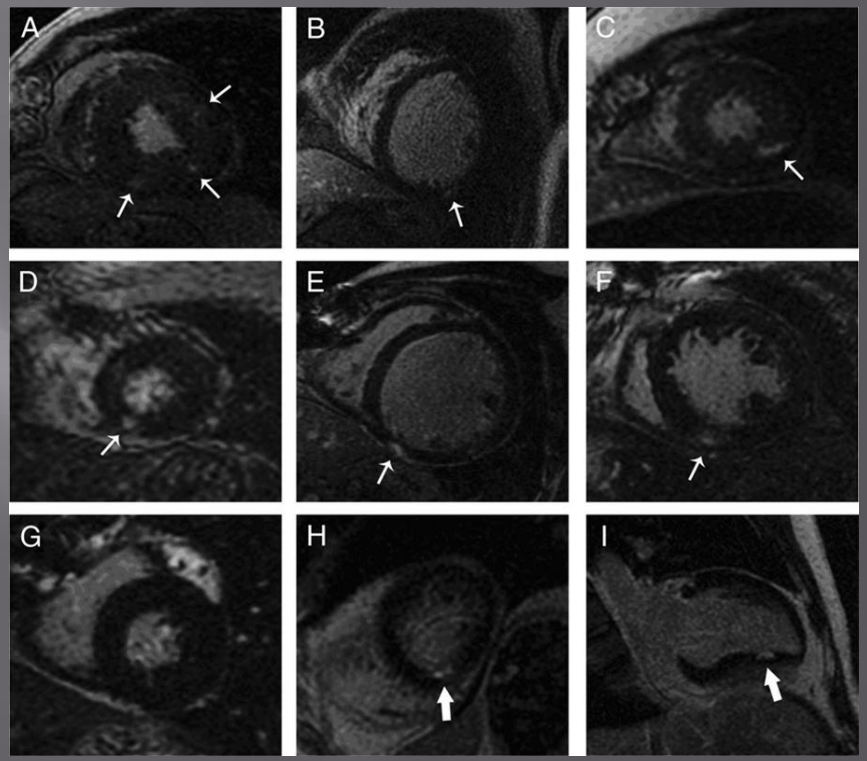
	Population	TAPSE	DTI S′	RIMP	FAC	2D FWLS
Vizzardi *et al.*[6]	Chronic HF (31 patients)	0.54 ($P < .01$)	0.81 ($P < .01$)	NR	0.07 (NS)	−0.76 ($P < .01$)
Focardi *et al.*[7]	Mixed population (63 patients)	0.45 ($P = .01$)	0.52 ($P = .01$)	NR	0.77 ($P < .001$)	−0.86 ($P < .001$)
Li *et al.*[8]	Chronic thromboembolic PH (32 patients)	0.451 ($P = .22$)	0.689 ($P < .001$)	−0.387 ($P = .04$)	0.423 ($P = .02$)	NR
Lu *et al.*[9]	Mixed population (60 patients)	0.27 ($P = .05$)	0.2 ($P > .05$)	−0.36 ($P > .05$)	0.33 ($P = .02$)	−0.54 ($P = .001$)
Leong *et al.*[11]	Systolic HF (83 patients)	0.65 ($P < .001$)	0.51 ($P < .001$)	0.28 ($P = .03$)	0.71 ($P < .001$)	−0.77 ($P < .001$)

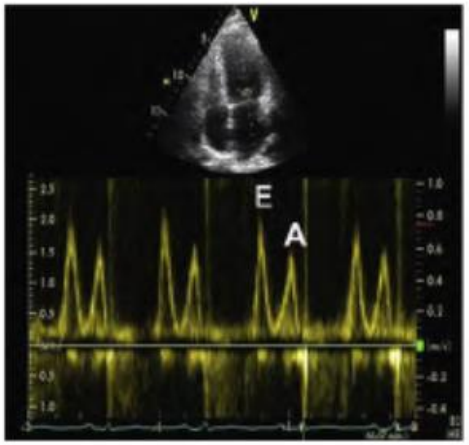
Longobardo et al. J Am Soc Echocardiogr 2017;30:937-46

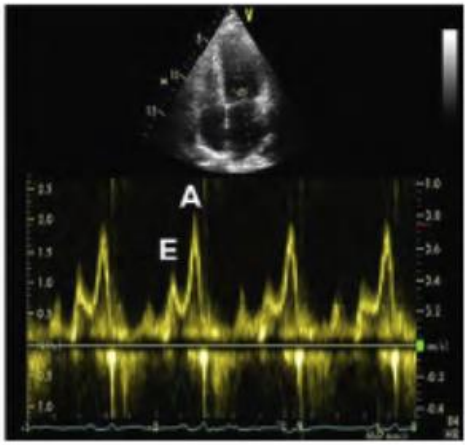

A
RVSP = 71 mmHG
B
IVC = 24 mm
RAP = 15 mmHg
C
DTI S' = 11 cm/sec
sPAP = 86 mmHg
D
FAC = 27 %
E
FR= 59 fps
HR= 75 bpm
Peak Systolic Strain
RV FWLS = - 11.6 %
RV GLS = -14 %

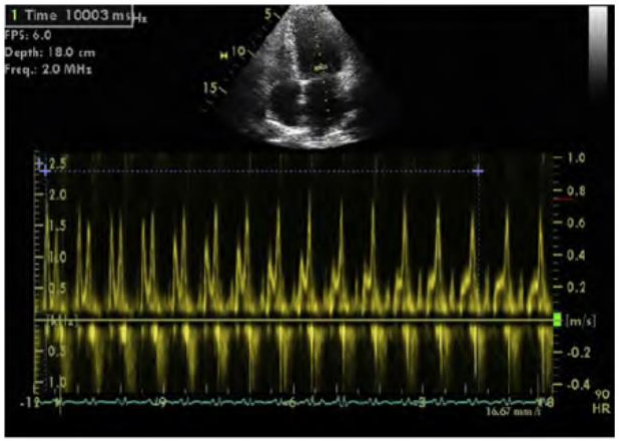

A
RVSP = 30 mmHg
B
IVC = 10 mm
RAP = 3 mmHg
C
DTI S' = 14 cm/sec
sPAP = 33 mmHg
D
FAC = 37 %
E
FR= 67 fps
HR= 98 bpm
Peak Systolic Strain
RV FWLS = - 22 %
RV GLS = - 19.6 %

- **Myocardial fibrosis is frequent as assessed by LGE**
  - TOF (53%)
  - Systemic RV (61%)
  - Eisenmenger's syndrome (73%)
  - Fontan (26%)

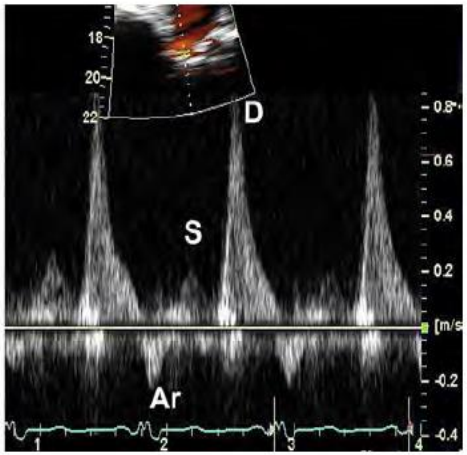


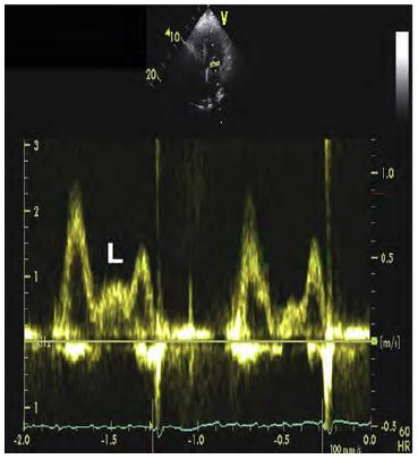

E
A

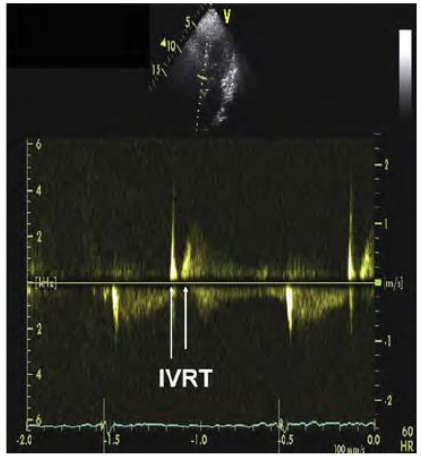

A
E


Time 10003 ms
FPS: 6.0
Depth: 18.0 cm
Freq.: 2.0 MHz
[m/s]
90
HR

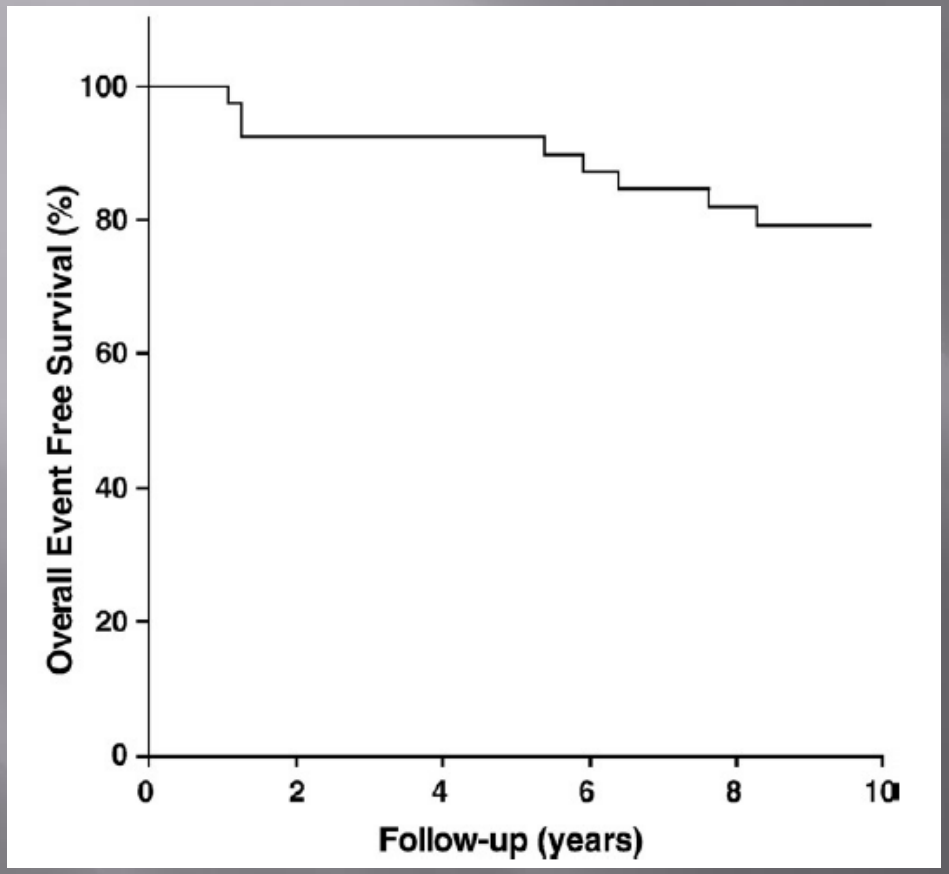

E
A
1.5
1.0
0.5
[m/s]
V
-2
-1
0
78
BPM

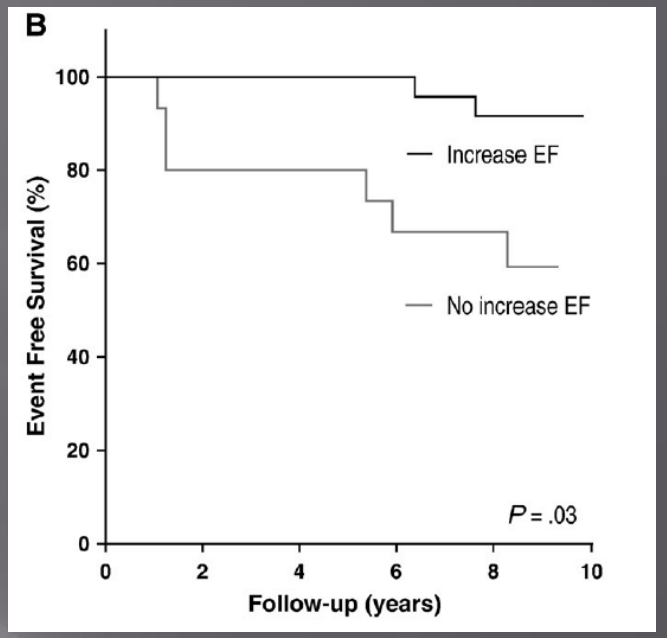

D
S
Ar
0.8
0.6
0.4
0.2
[m/s]
-0.2
-0.4
1
2
3
4

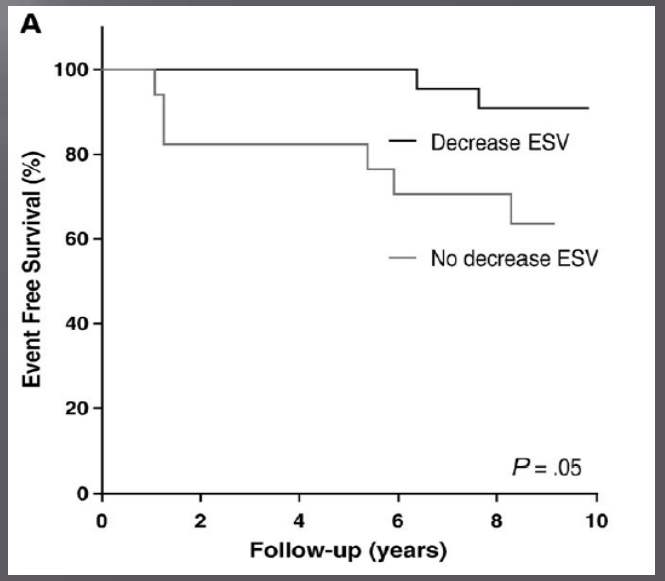

V
L
[m/s]
-2.0
-1.5
-1.0
-0.5
0.0
HR


V
IVRT
[m/s]
-2.0
-1.5
-1.0
-0.5
0.0
HR

39 adults with systemic RV
Stress MRI
8 year follow up







Winter et al. American Heart Journal 2010

Conclusion

- **Increasing non-invasive measures of ventricular dysfunction**
  - Novel, interesting
  - Take special care with technical limitations
  - Use multiple measures together
  - Particularly useful for serial measurement
- **Is there data to advise what to do with the more novel information?**
  - Medical therapy
  - Earlier intervention
  - Increased risk of arrhythmia and SCD