Role of imaging in Heart Failure: Heart failure in corrected congenital heart disease

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Imaging modalities

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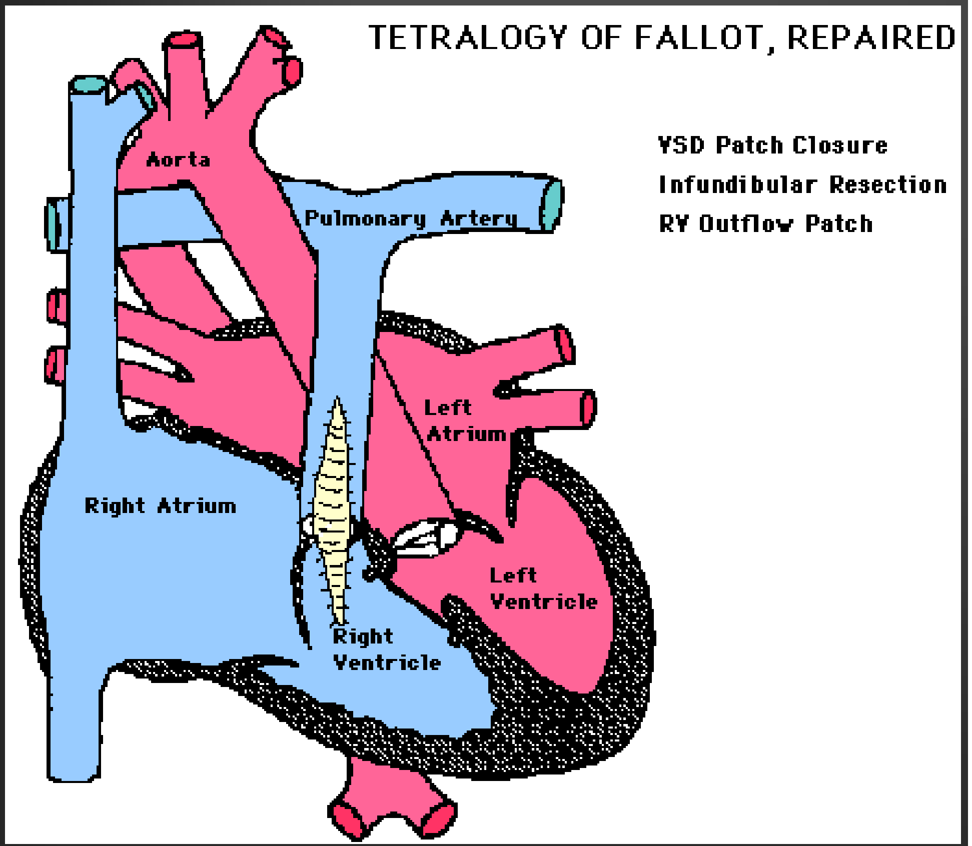
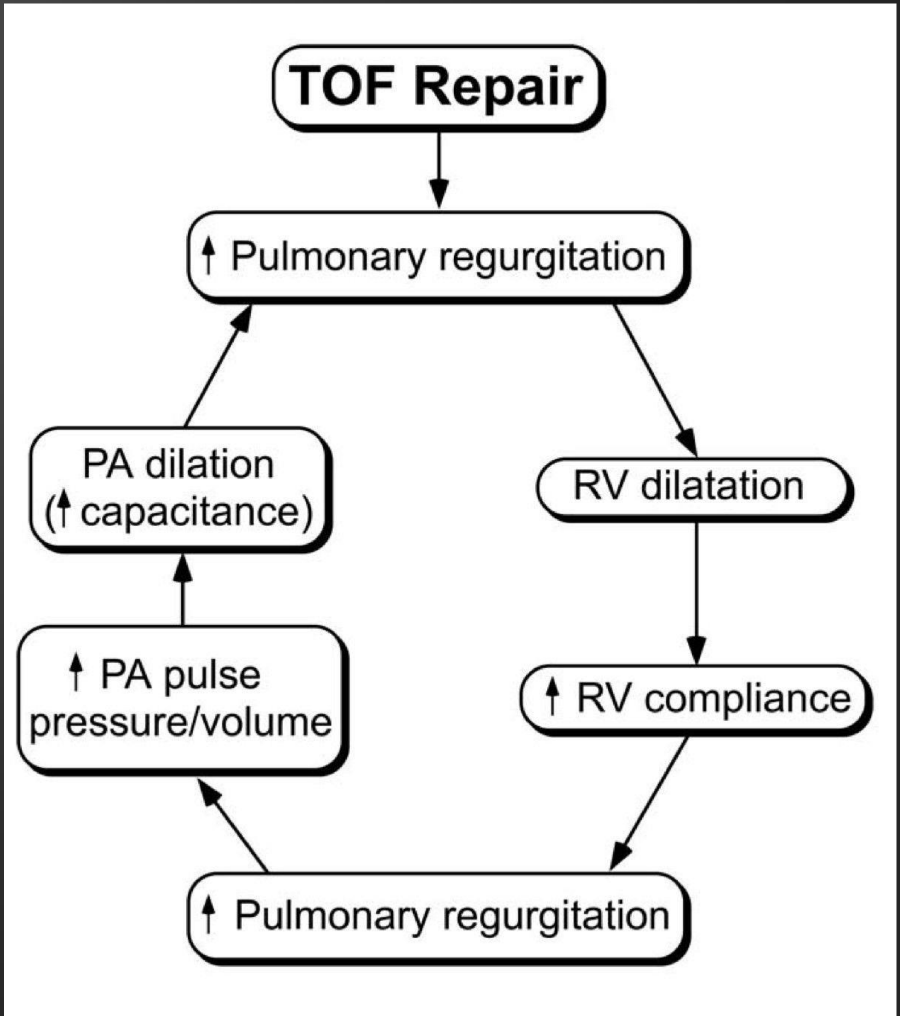
- Chest X-Ray
- Echocardiography
- Cardiovascular Magnetic Resonance
- Cardiovascular Computed Tomography
- Nuclear Scintigraphy
- Barium Swallow
- X-ray Angiography

Tetralogy of Fallot - Repair





Repaired TOF: RV - PI - TR

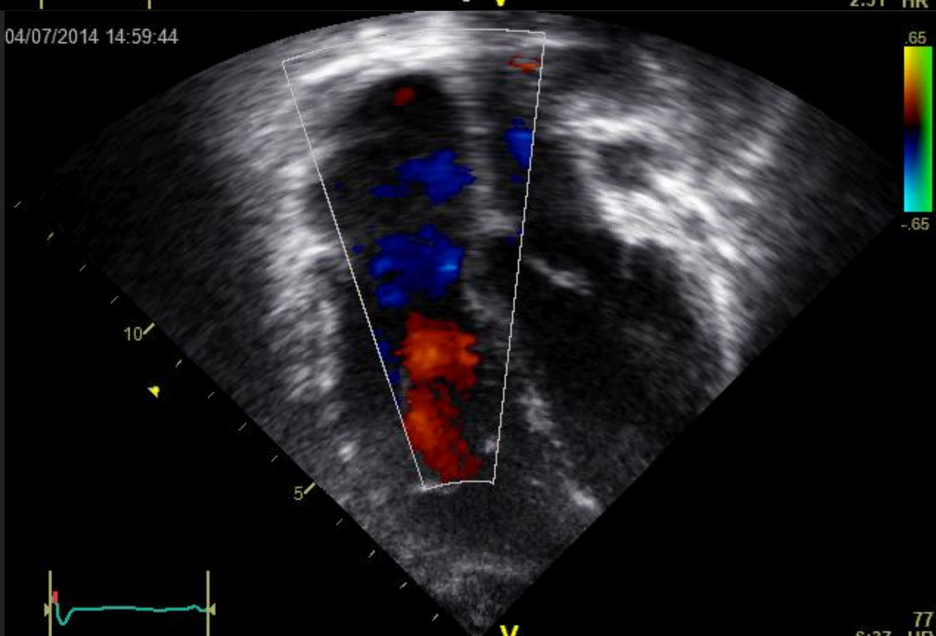


Table 3 Echocardiographic reference values of RA and ventricular size and function in healthy adults (adapted from Rudski *et al.*[23])

Parameter	Abnormal
RV diameter (mm)	
Base	>42
Midventricular level	>35
RV diastolic length (mm)	>86
RV diaphragmatic wall thickness (subcostal view) (mm)	>5
Systolic function	
TAPSE (mm)	<16
Pulsed Doppler peak S′ (cm/sec)	<10
FAC (%)	<35
Diastolic function	
E/E′ ratio	>6
RA end-systolic area (cm^2)	>18
RA length (base to apex) (mm)	>53
RA lateral diameter (mm)	>44

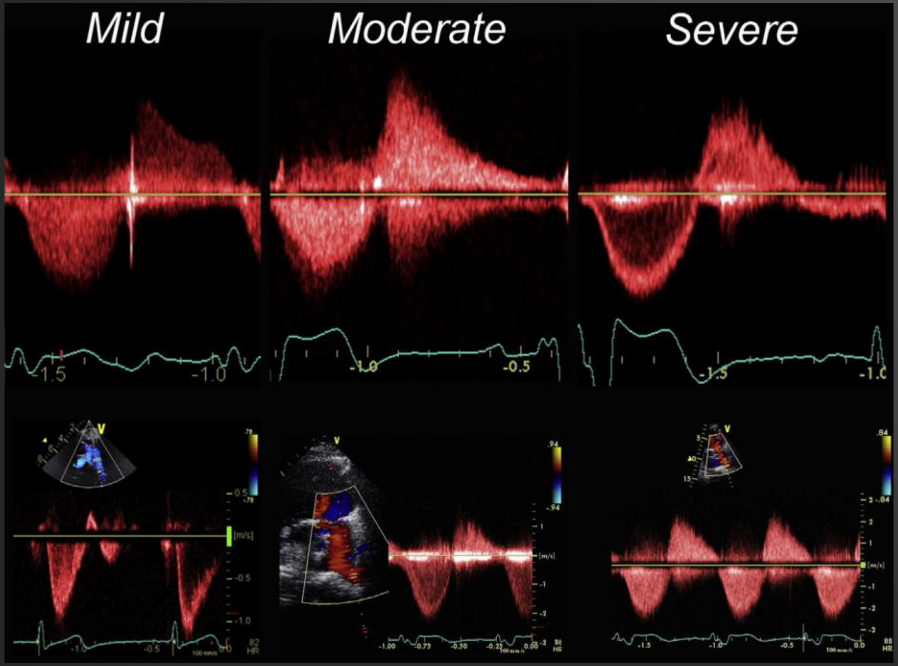
FAC, Fractional area change.

Repaired TOF: PI - TR

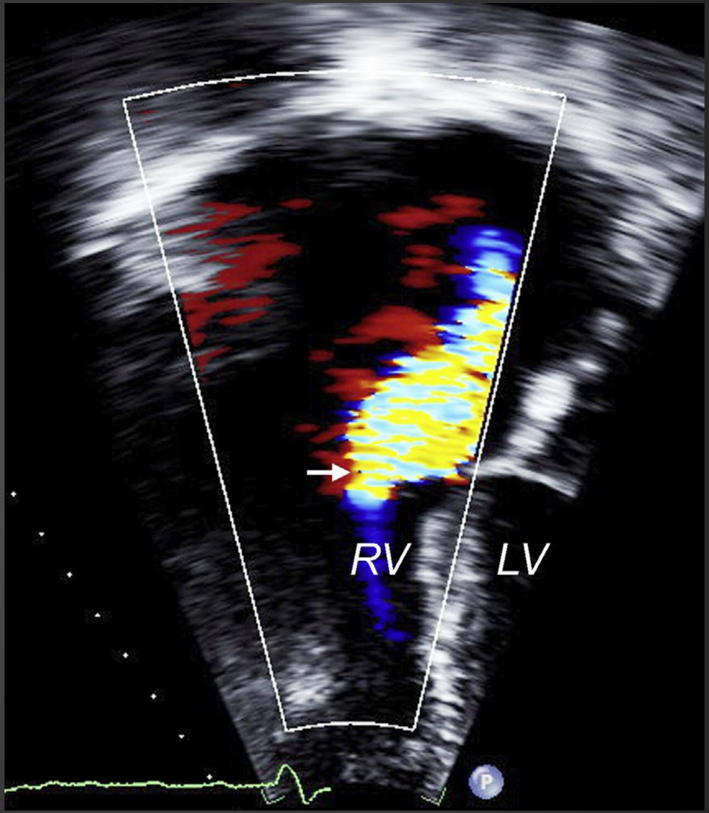
Mild PI: persistent flow gradient at end-diastole

Moderate PI: equilibration of MPA and RV pressures only at end-diastole

Severe PI: early diastolic pressure equilibration



LPA PW showing degrees of diastolic flow reversal

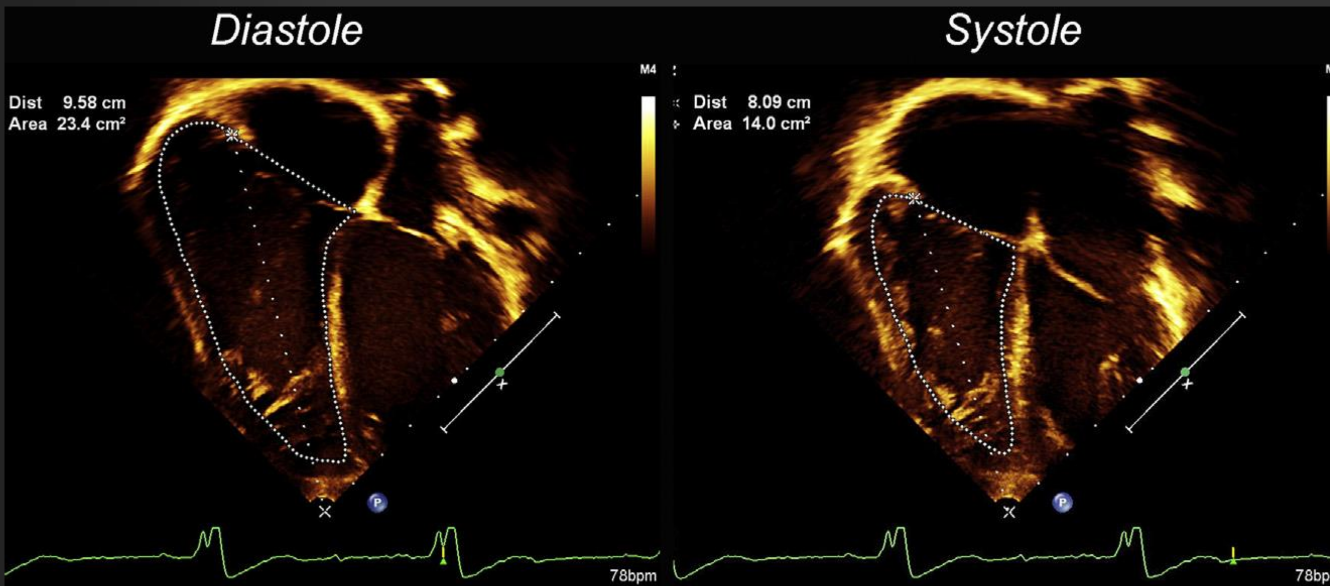


TR morphology

RVp estimate = $4v^2$ + RAP

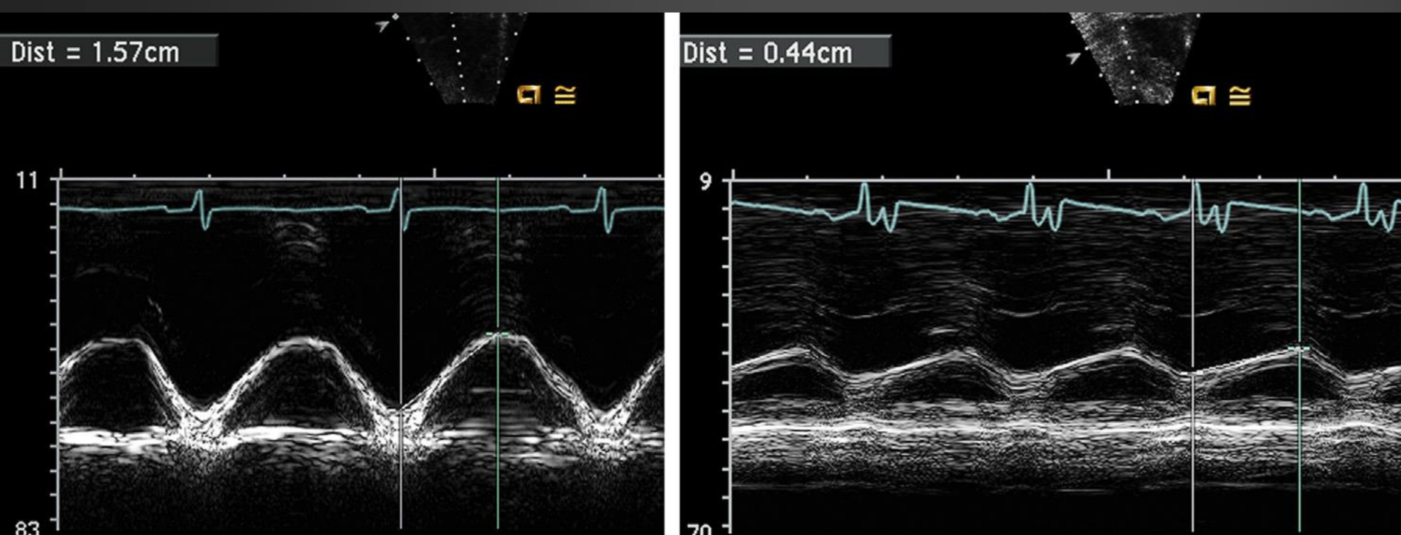
J Am Soc Echocardiogr 2014;27:111-41

Repaired TOF: RV



RV fractional area change (%)

(modest correlation with CMR in TOF)



TAPSE

(weak correlation with CMR in TOF)

J Am Soc Echocardiogr 2014;27:111-41

Repaired TOF: Newer techniques

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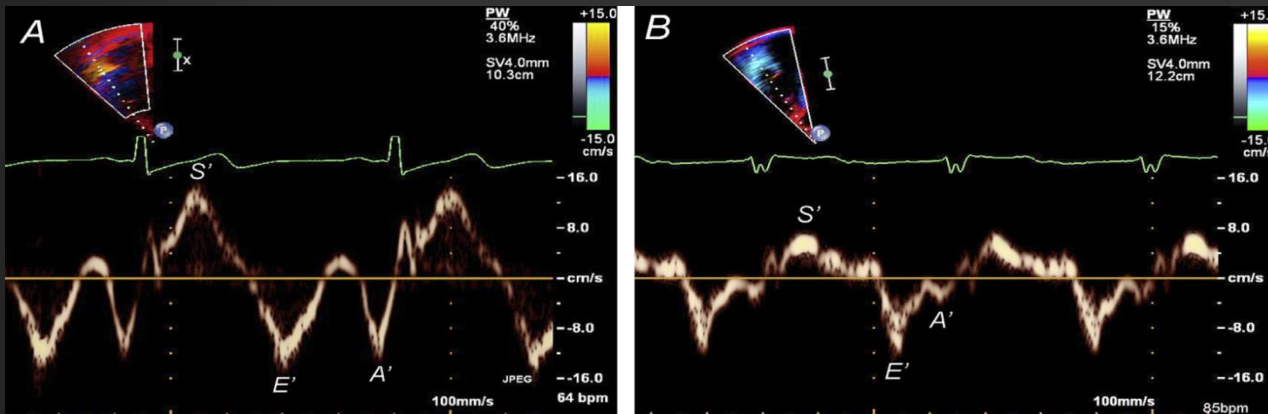


Nl RV fxn (S′ 14 cm/sec)

↓ RV fxn (S′ 7 cm/sec)

TDI

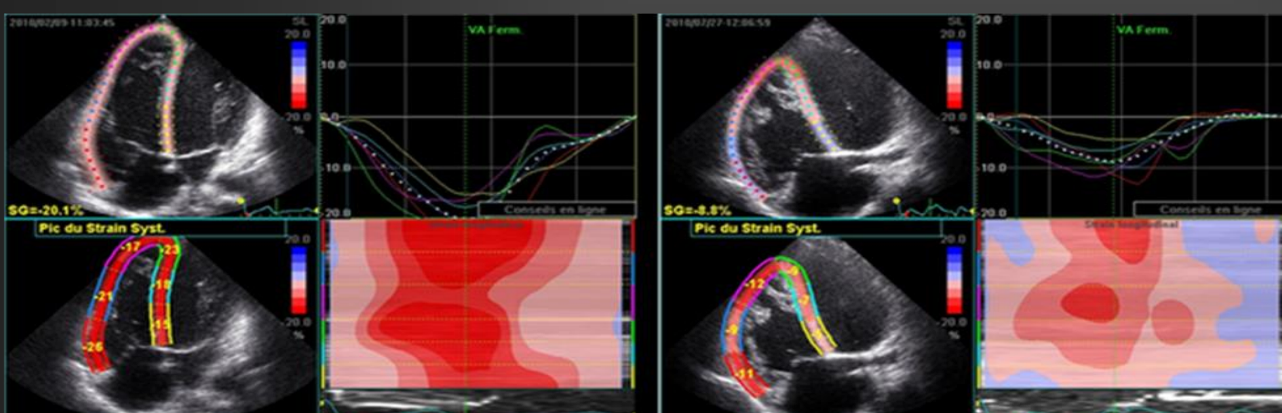
(no available guidelines in CHD)

J Am Soc Echocardiogr 2014;27

RV MPI correlates modestly with CMR RV EF

RV MPI

Circ Cardiovasc Imaging . 2012;5(5)



Nl RV fxn

TOF with ↓ RV fxn

2D Speckle tracking

(no available guidelines in CHD)

Echocardiography. 2014;31(4)

TOF – TDI - Dobutamine

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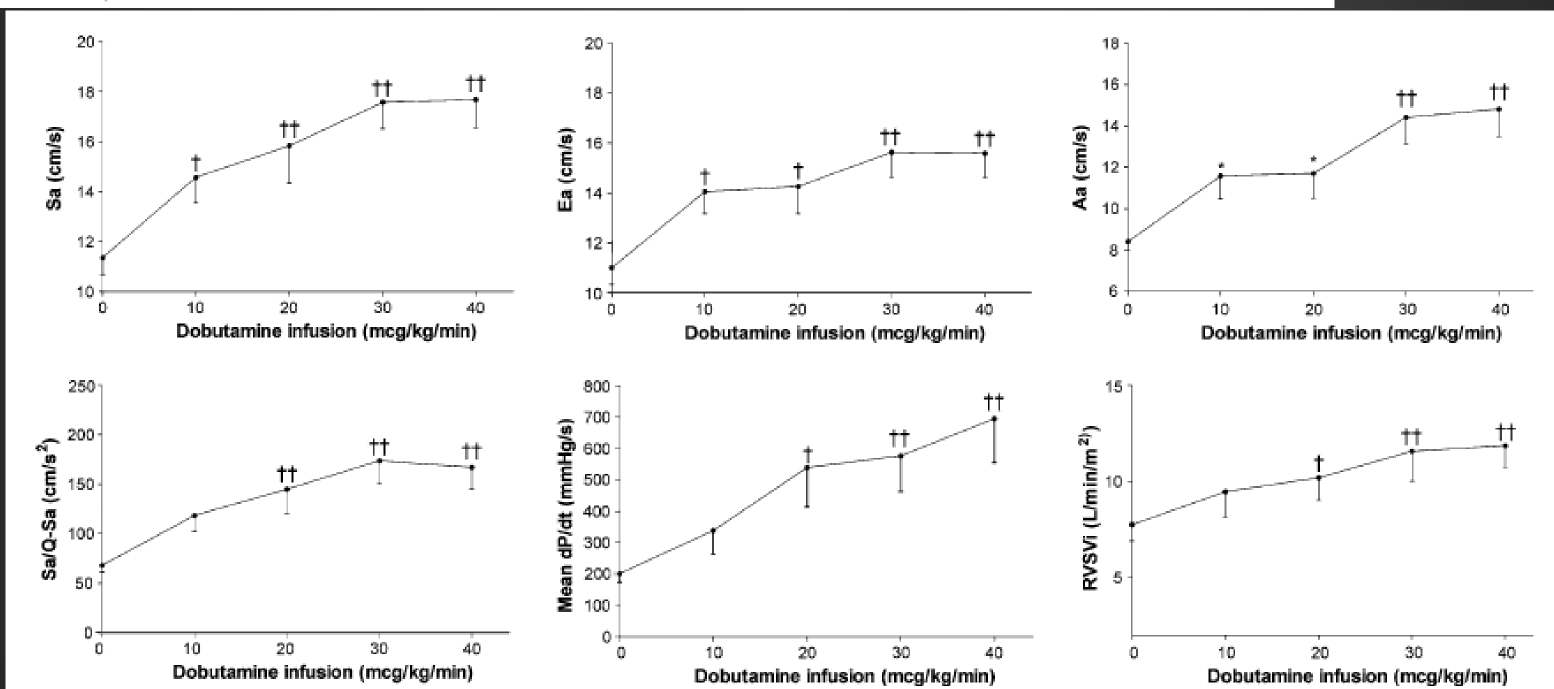
Doppler tissue imaging evaluation of right ventricular function at rest and during dobutamine infusion in patients after repair of tetralogy of Fallot

Table 2 Echocardiographic data

Parameter	A. Controls rest	B. TOF patients rest	C. TOF patients peak dobutamine
Sa (cm/s)	13.7 ± 3.1	11.4 ± 2.8*	17.7 ± 4.7***
Ea (cm/s)	16.3 ± 3.5	11 ± 3.1**	15.6 ± 3.9***
Aa (cm/s)	9.2 ± 3.2	8.4 ± 2	14.8 ± 5***
Sa/Q–Sa (cm/s^2)	92.3 ± 29.4	68.8 ± 26.4**	176.8 ± 84.5***
Mean d*P*/d*t* (mmHg/s)		180 ± 74	537 ± 372***
RVSVi (L/min/m^2)		7.8 ± 3.9	11.9 ± 5.6***

25 patients
11±6 (6–27) yo
Dobutamine



Apostolopoulou et al. Int J Cardiovasc Imaging (2007) 23:25–31

TOF Bike

Exercise Echocardiography Demonstrates Biventricular Systolic Dysfunction and Reveals Decreased Left Ventricular Contractile Reserve in Children After Tetralogy of Fallot Repair

S. Lucy Roche, MB ChB, MRCPCH, MD, Lars Grosse-Wortmann, MD, Mark K. Friedberg, MD, Andrew N. Redington, MD, FRCP, Derek Stephens, MSC, and Paul F. Kantor, MBBCh, DCH, FRCP, *Toronto, Ontario, and Edmonton, Alberta, Canada*

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Variable	Controls (*n* = 27)	Patients with TOF (*n* = 29)	*P*	Percentage of patients with TOF with measurements ≥2 SDs below controls' mean
Peak heart rate	164 ± 15.1	137.2 ± 15.6	.0005	41
RVOT FS (%)	37.7 ± 7.8	25.3 ± 7.1	<.0001	32
RV LAX FS (%)	34.6 ± 7.4	21.2 ± 6.1	<.0001	48
RV IVA (m/sec^2)	10.3 ± 2.0	3.5 ± 1.4	<.0001	100
RV Sm (cm/sec)	13.1 ± 2.5	9.2 ± 2.5	<.0001	34
LV LAX FS (%)	22.3 ± 4.6	23.0 ± 6.3	NS	7
LV IVA (m/sec^2)	9.9 ± 2.6	3.8 ± 1.6	<.0001	75
LV Sm (cm/sec)	9.9 ± 1.7	8.1 ± 2.4	.001	25

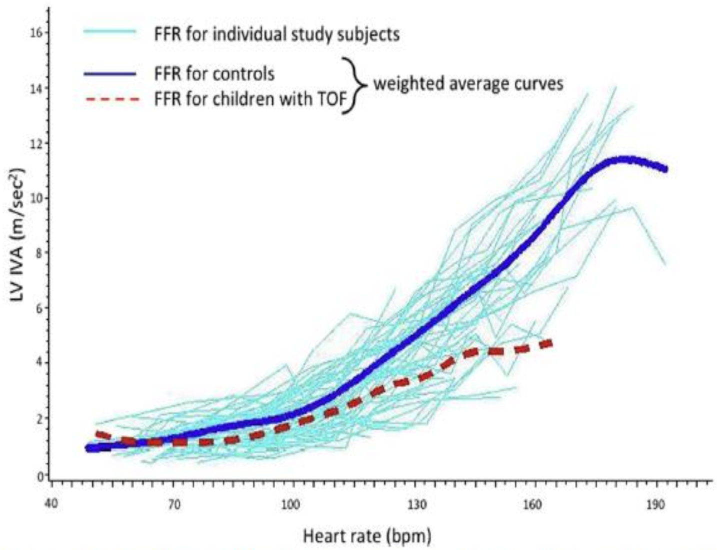

Figure 3 LV force-frequency relationships in asymptomatic children with TOF and healthy controls. Each *light blue line* represents an individual study participant, and the group locally weighted scatterplot smoothing average curves are superimposed as *heavier lines.*

29 TOF patients
Bike

Roche et al. J Am Soc Echocardiogr 2015;28:294-301

CMR in CHD

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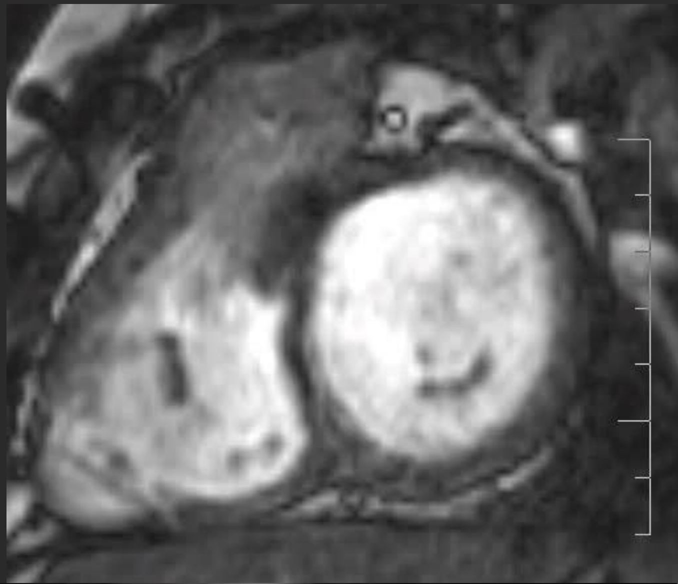
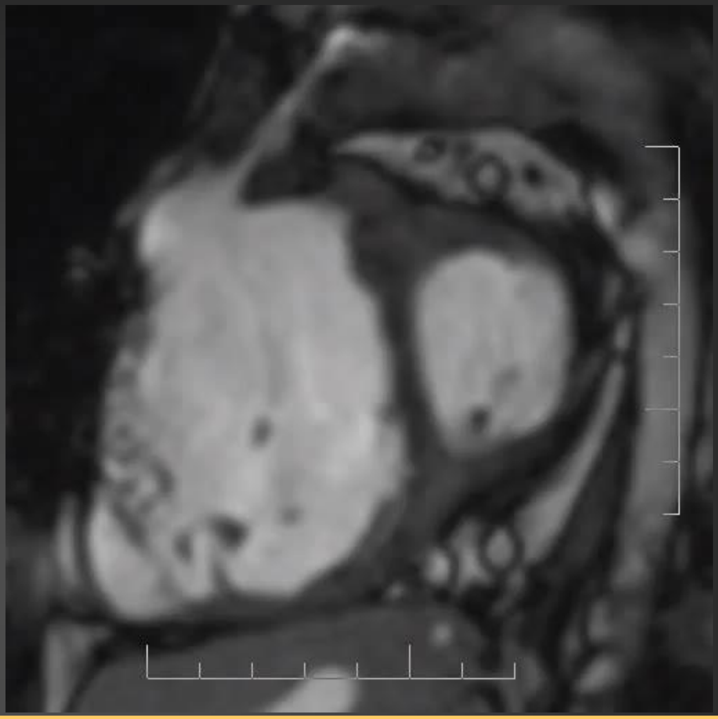
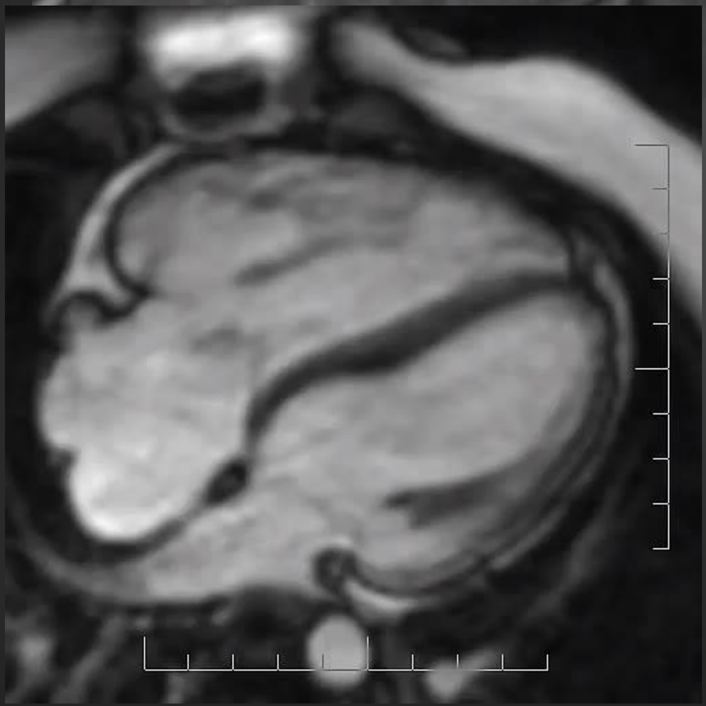
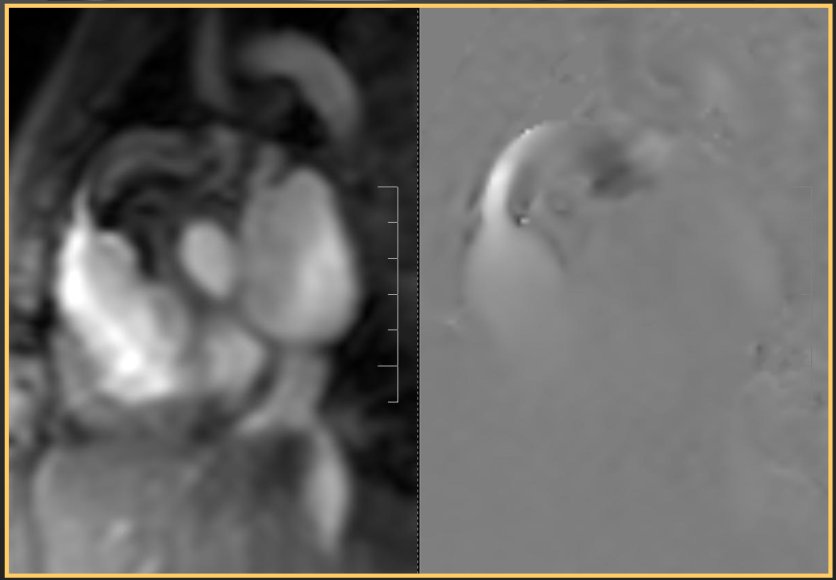
- Anatomical delineation
- Static and Cine imaging
- High–spatial resolution 3D reconstruction
- Phase-contrast (PC) for blood flow measurements
- Late gadolinium enhancement
- Accurate, objective, reproducible and quantitative measurements
  - biventricular size and function
  - pulmonary and systemic blood flow measurements
  - differential PA flow
  - valve regurgitant volumes (e.g., PR, AR)
  - myocardial viability and presence of scarring
  - origin & proximal course of coronaries

CMR – Anatomy – Cine – PS/PI









CMR – Measurements – RV/LV/PI

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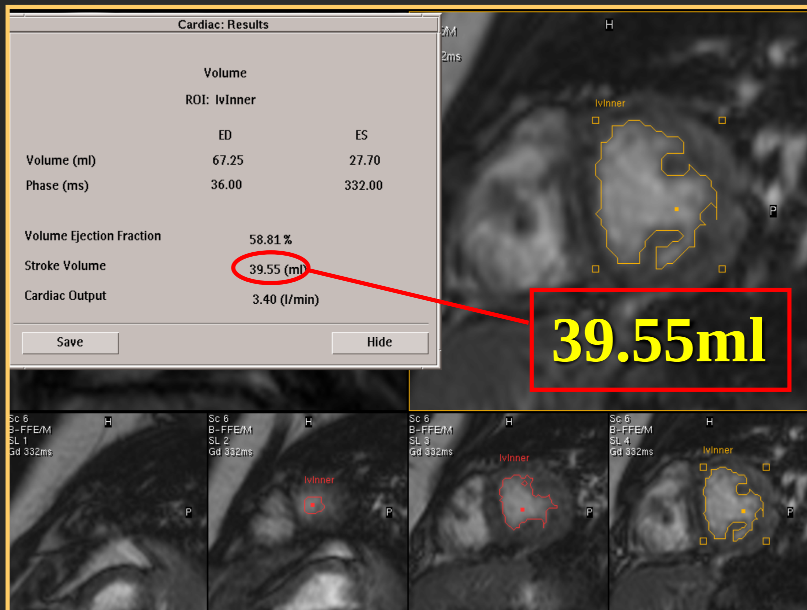
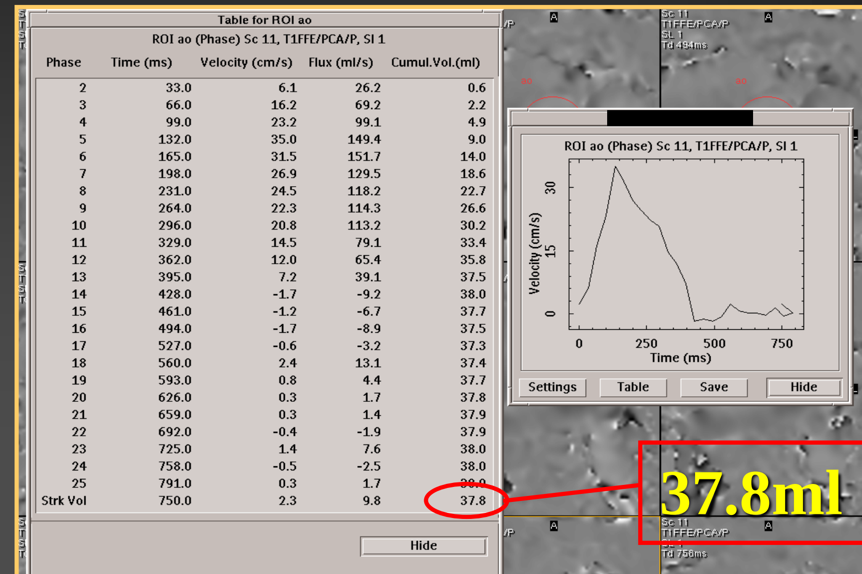
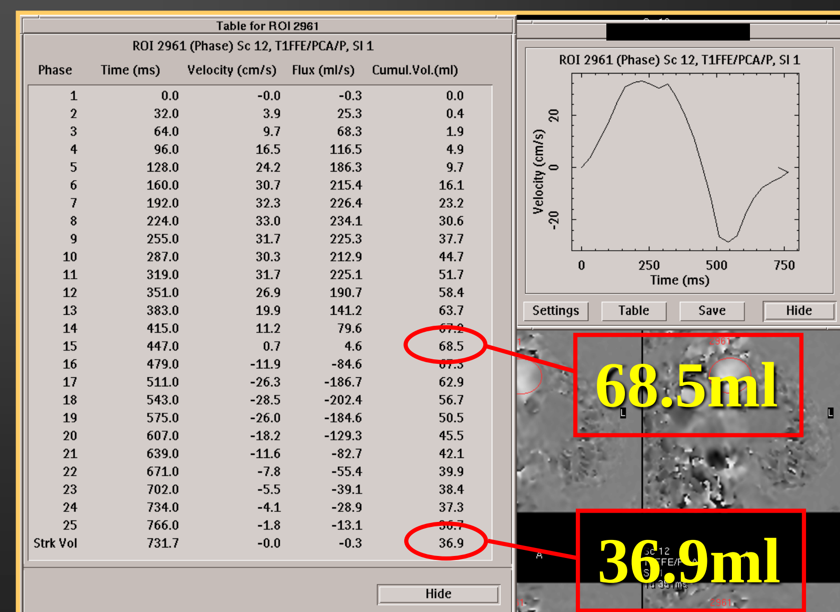


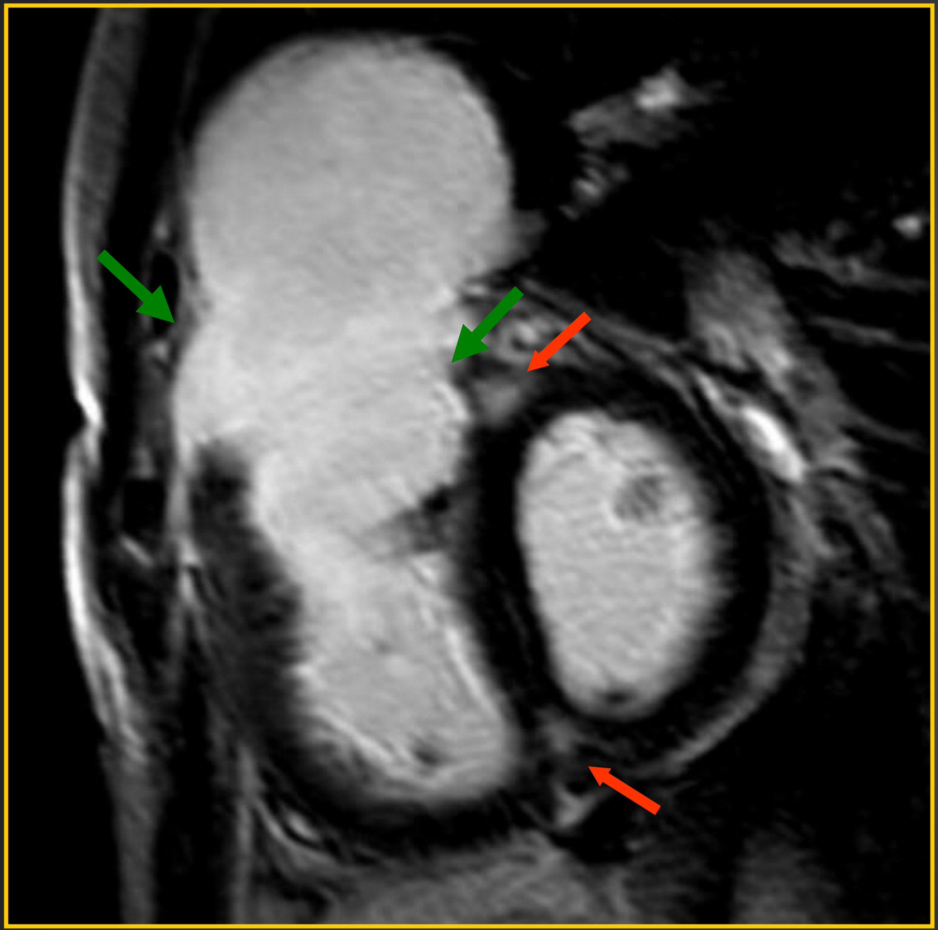


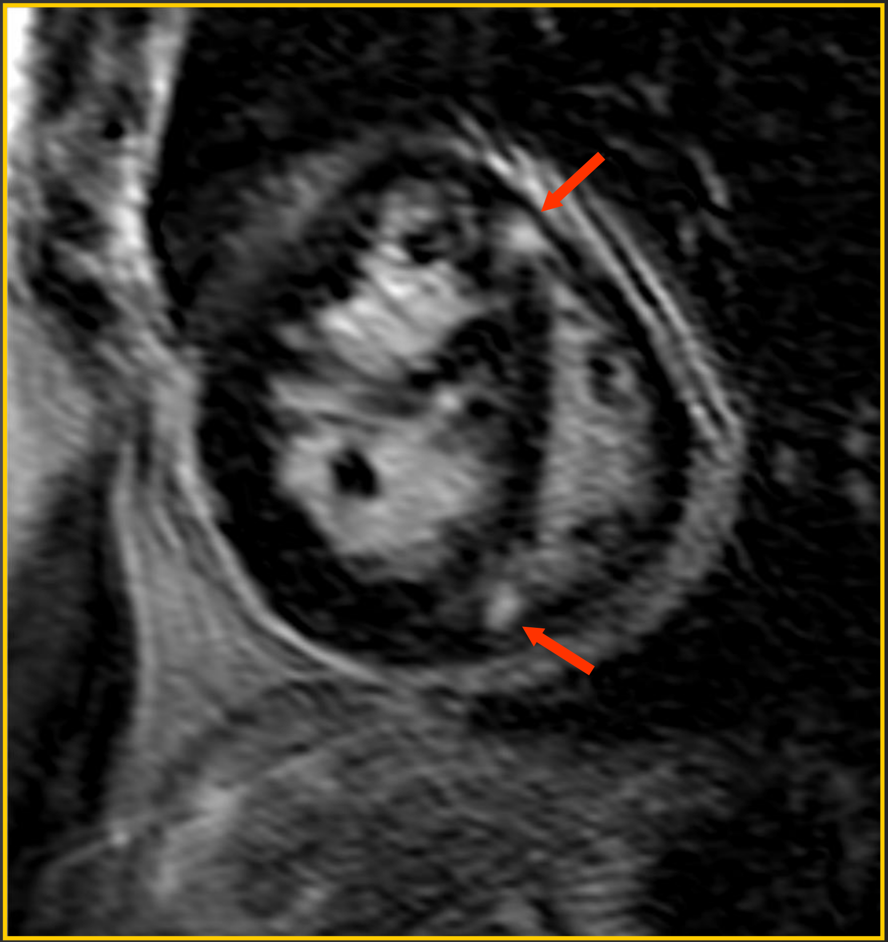
Phase	Time (ms)	Velocity (cm/s)	Flux (ml/s)	Cumul.Vol.(ml)
2	33.0	6.1	26.2	0.6
3	66.0	16.2	69.2	2.2
4	99.0	23.2	99.1	4.9
5	132.0	35.0	149.4	9.0
6	165.0	31.5	151.7	14.0
7	198.0	26.9	129.5	18.6
8	231.0	24.5	118.2	22.7
9	264.0	22.3	114.3	26.6
10	296.0	20.8	113.2	30.2
11	329.0	14.5	79.1	33.4
12	362.0	12.0	65.4	35.8
13	395.0	7.2	39.1	37.5
14	428.0	-1.7	-9.2	38.0
15	461.0	-1.2	-6.7	37.7
16	494.0	-1.7	-8.9	37.5
17	527.0	-0.6	-3.2	37.3
18	560.0	2.4	13.1	37.4
19	593.0	0.8	4.4	37.7
20	626.0	0.3	1.7	37.8
21	659.0	0.3	1.4	37.9
22	692.0	-0.4	-1.9	37.9
23	725.0	1.4	7.6	38.0
24	758.0	-0.5	-2.5	38.0
25	791.0	0.3	1.7	[illegible]
Strk Vol	750.0	2.3	9.8	37.8



Phase	Time (ms)	Velocity (cm/s)	Flux (ml/s)	Cumul.Vol.(ml)
1	0.0	-0.0	-0.3	0.0
2	32.0	3.9	25.3	0.4
3	64.0	9.7	68.3	1.9
4	96.0	16.5	116.5	4.9
5	128.0	24.2	186.3	9.7
6	160.0	30.7	215.4	16.1
7	192.0	32.3	226.4	23.2
8	224.0	33.0	234.1	30.6
9	255.0	31.7	225.3	37.7
10	287.0	30.3	212.9	44.7
11	319.0	31.7	225.1	51.7
12	351.0	26.9	190.7	58.4
13	383.0	19.9	141.2	63.7
14	415.0	11.2	79.6	[illegible]
15	447.0	0.7	4.6	68.5
16	479.0	-11.9	-84.6	[illegible]
17	511.0	-26.3	-186.7	62.9
18	543.0	-28.5	-202.4	56.7
19	575.0	-26.0	-184.6	50.5
20	607.0	-18.2	-129.3	45.5
21	639.0	-11.6	-82.7	42.1
22	671.0	-7.8	-55.4	39.9
23	702.0	-5.5	-39.1	38.4
24	734.0	-4.1	-28.9	37.3
25	766.0	-1.8	-13.1	[illegible]
Strk Vol	731.7	-0.0	-0.3	36.9

CMR - Myocardial Enhancement





Gadolinium uptake in areas of fibrosis 10 to 20 min after contrast injection

Criteria for PVR - Role of CMR

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- Asymptomatic patients with ≥ 2 of:

  1. RVEDV index>150 ml/m^2 or Z-score>4 (large patients RV/LV EDV >2)
  2. RV end-systolic volume index > 80 ml/m^2
  3. RV EF < 47%
  4. LV EF < 55%
  e. Large RVOT aneurysm
  5. QRS duration >140 ms
  6. Sustained tachyarrhythmia related to right heart volume load
  7. Other:
     - RVOT obstruction with RVp ≥ 2/3 systemic
     - Branch PA stenosis (<30% flow to affected lung) not amenable to stenting
     - ≥ Moderate TR
     - Residual ASD or VSD with Qp/Qs ≥ 1.5
     - Severe aortic regurgitation
     - Severe aortic dilatation (diameter ≥ 5 cm)

- Symptomatic patients (exercise intolerance or heart failure) and ≥ 1 of the above

Journal of Cardiovascular Magnetic Resonance 2011

TOF - Cardiac CT

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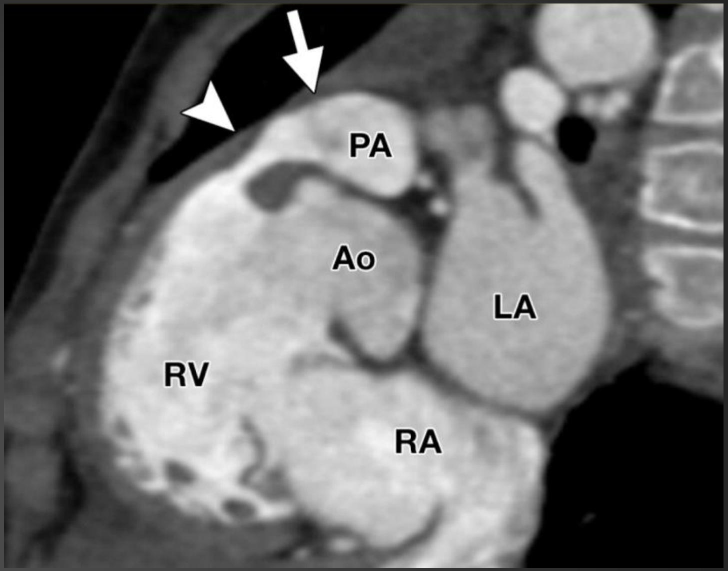
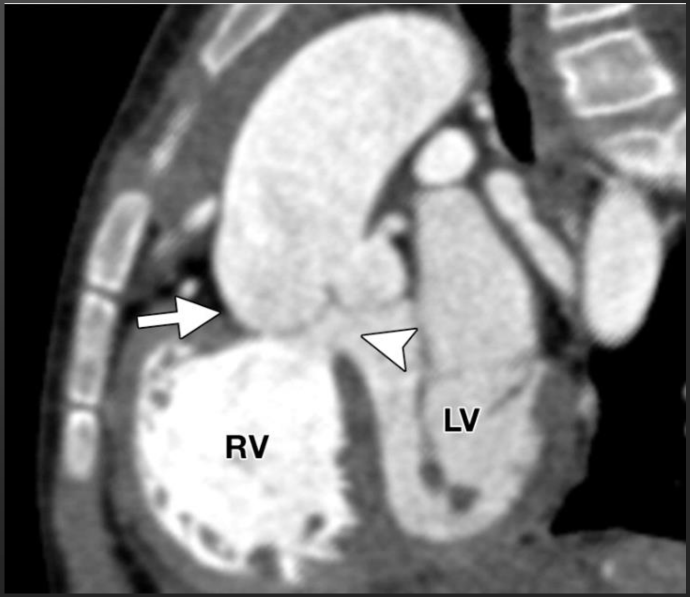
- Anatomical delineation, excellent spatial resolution
- Static and Cine imaging
- 3D reconstruction
- ↑ delineation of small vessels (coronaries, distal PA branches)
- Compatible with pacemakers and defibrillators
- Less artifacts by stainless-steel metallic artifacts
- Radiation, No hemodynamic information





TOF Nuclear Scintigraphy

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- Ventricular size & function
- Pulmonary perfusion
- Quantification of cardiac shunts
- Quantification of differential PA blood flow
- V-scan to assess ventilation-perfusion mismatch
- Myocardial perfusion and viability
- Radiation
- Only used in contraindications to CMR and cardiac CT

TOF X-ray Angiography

- Imaging RVOT/PAs/Aorta
- RV size & function / PI qualitatively
- Hemodynamics
- Coronaries (adults, abnormal course, Melody implantation)
- Catheter interventions:
  - PA balloon dilation and stenting
  - percutaneous PV implantation (Melody/Sapien)
  - occlusion of aortopulmonary collaterals
  - closure of residual septal defects
  - coronary artery interventions
- Critically ill patient
- Invasive, radiation, cost

Repaired TOF – Angiography

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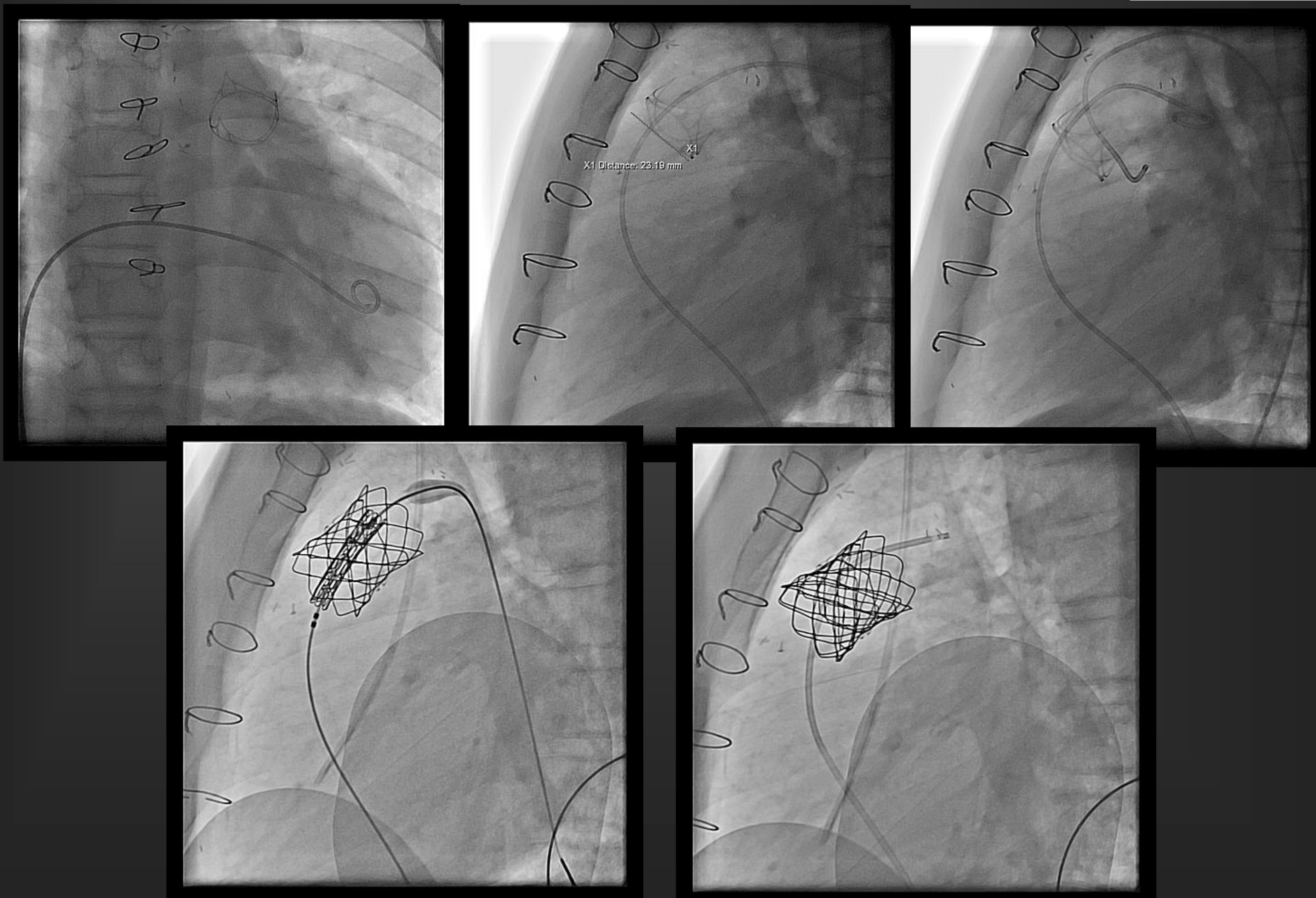
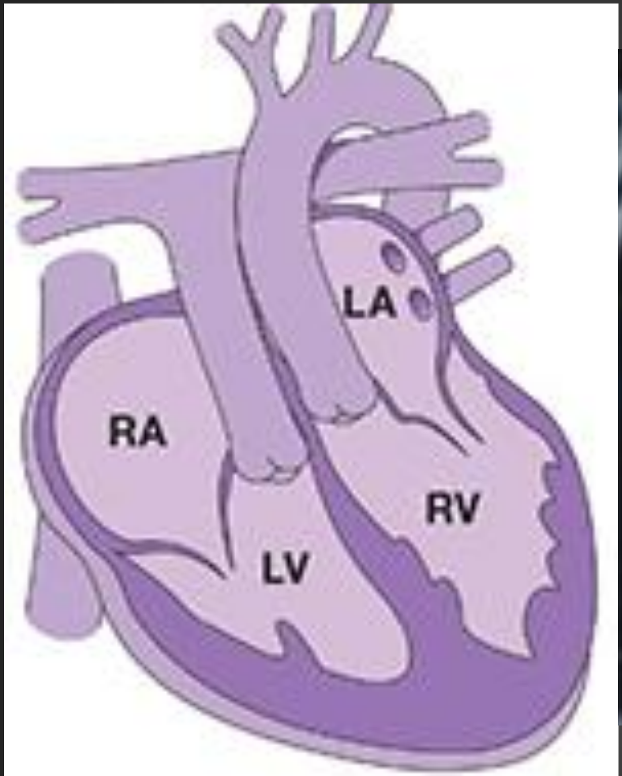
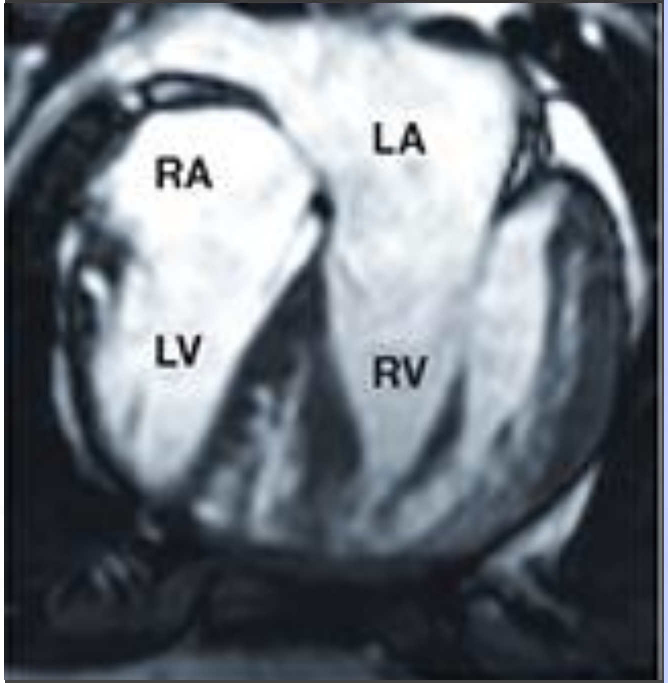
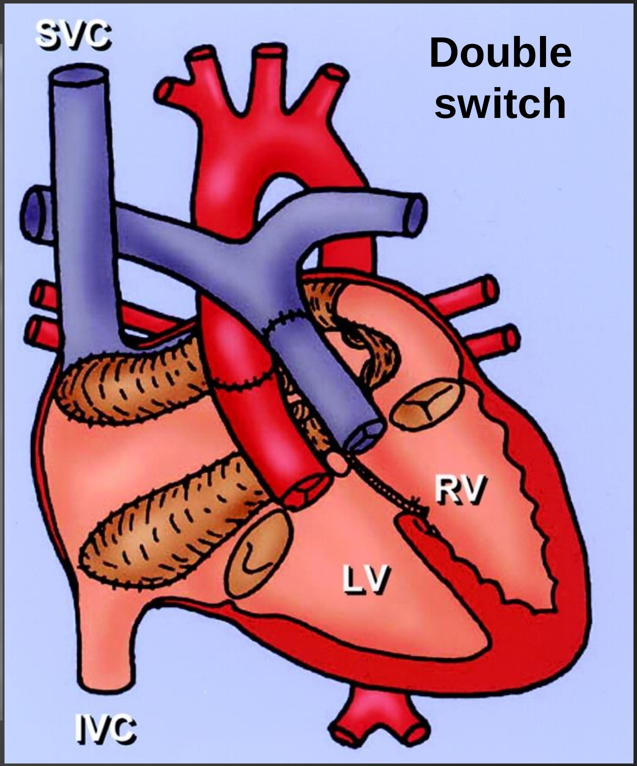


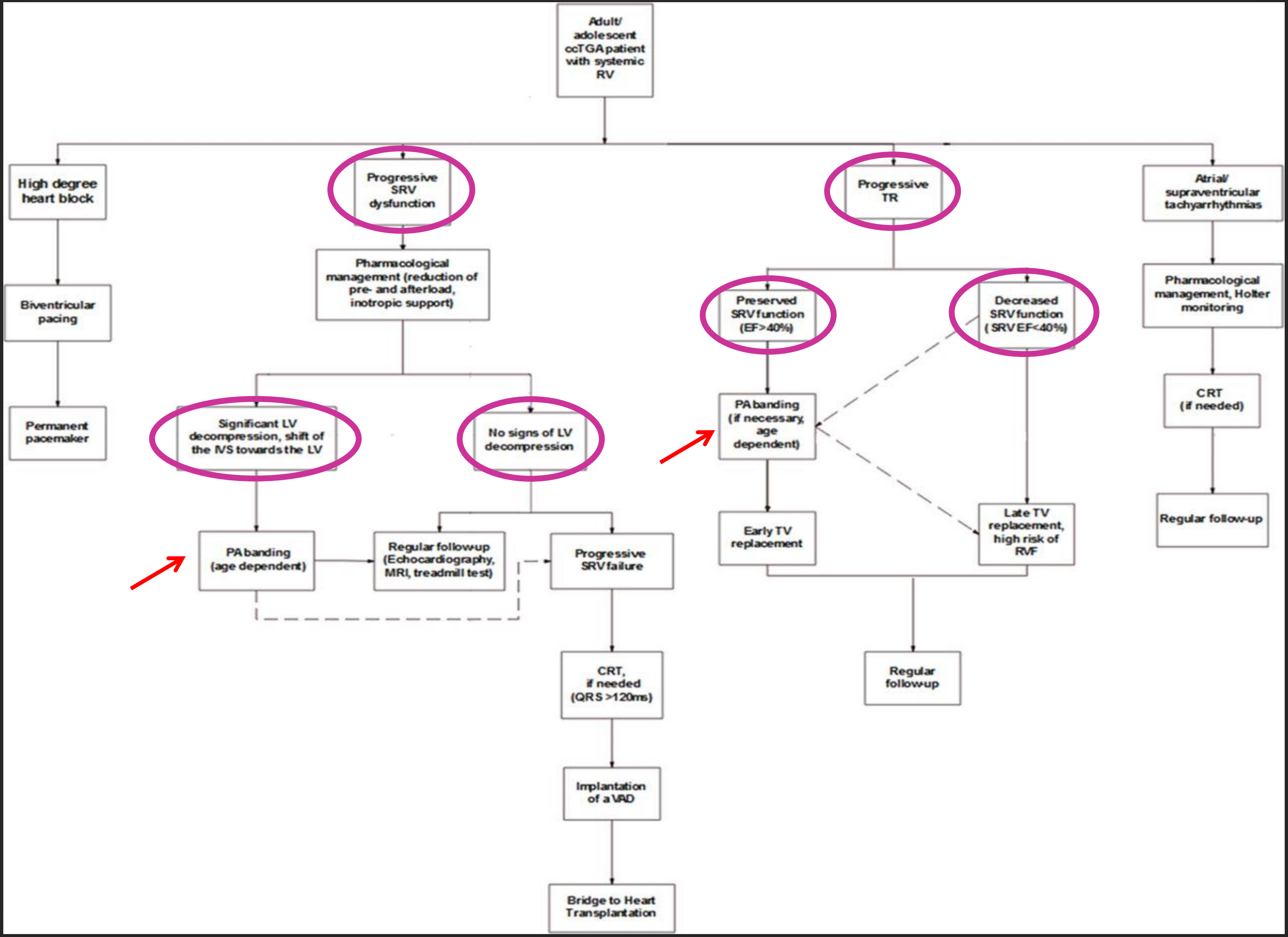
Systemic RV - CCTGA





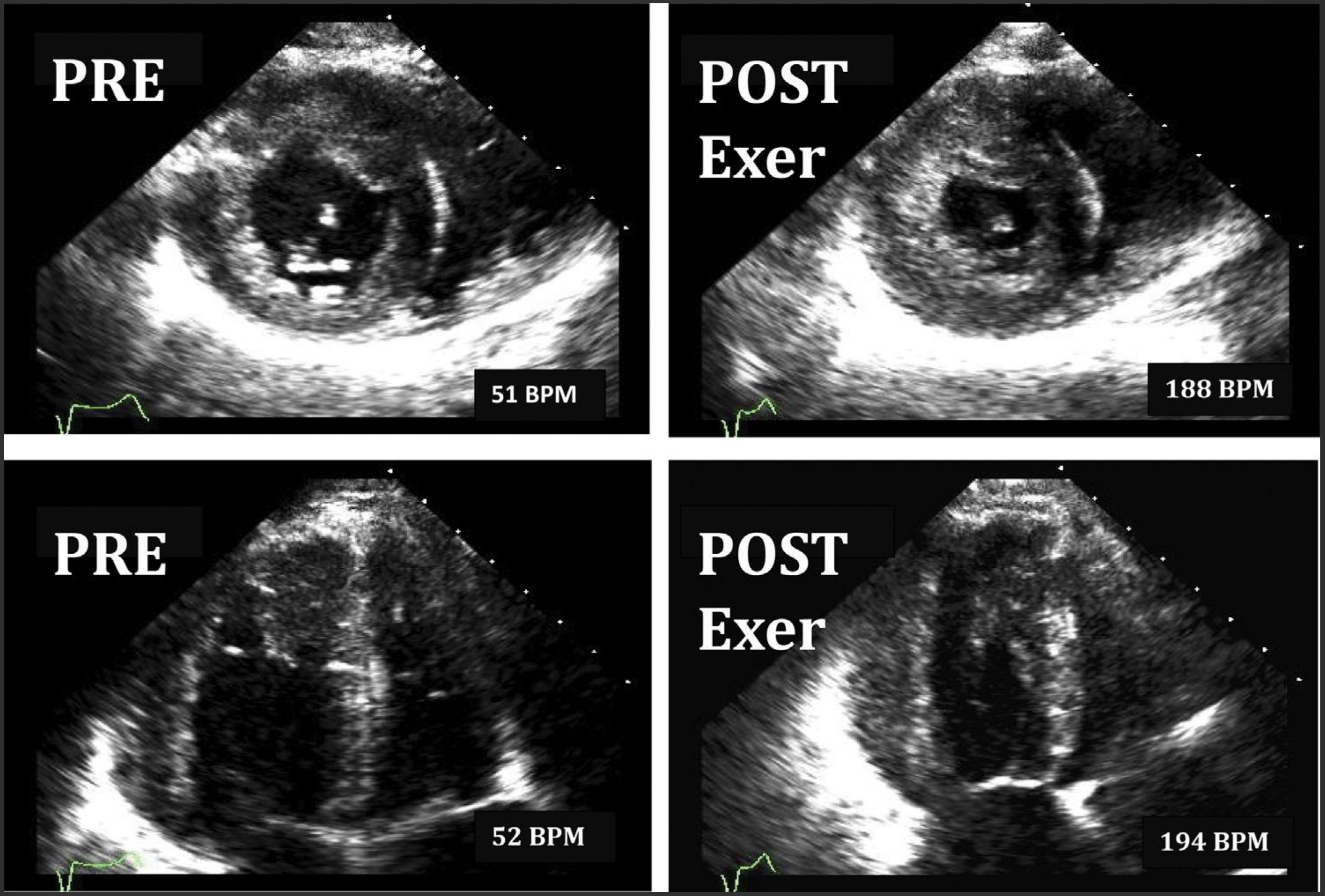


CCTGA – Adult algorithm



Circulation. 2016;134:1293-1302

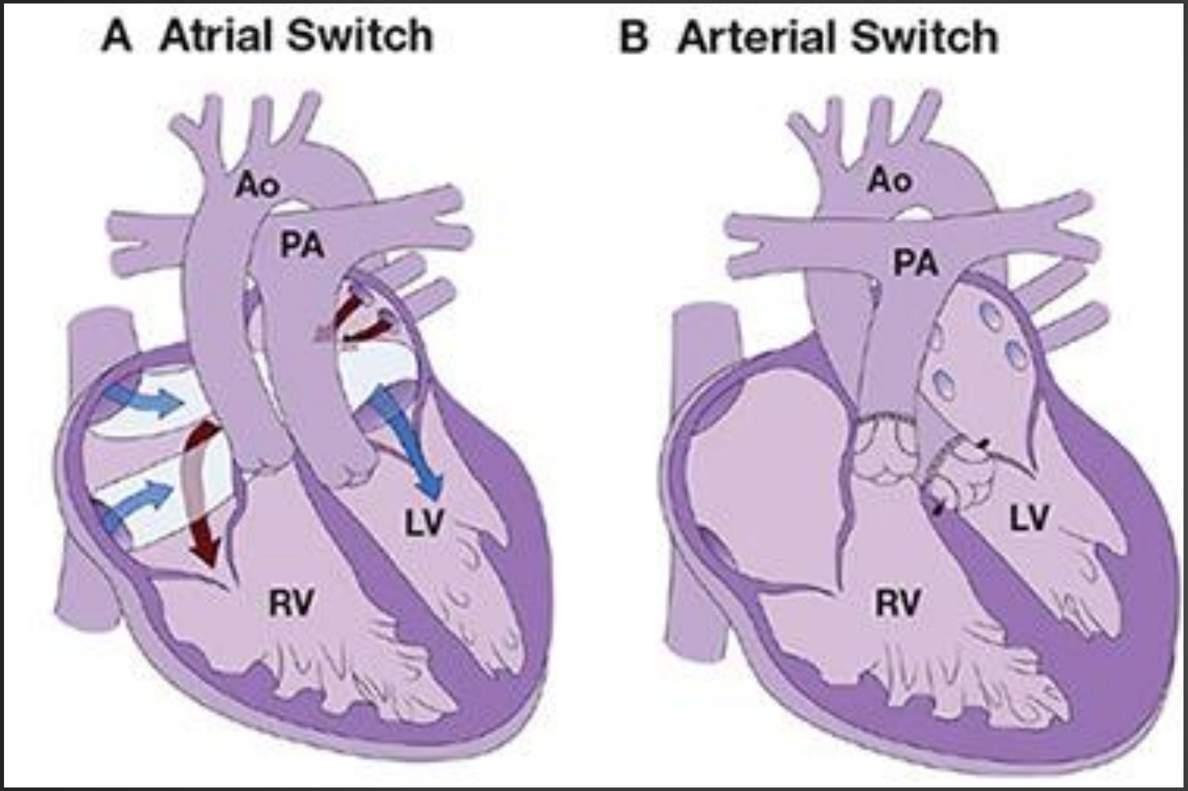
CCTGA - Exercise

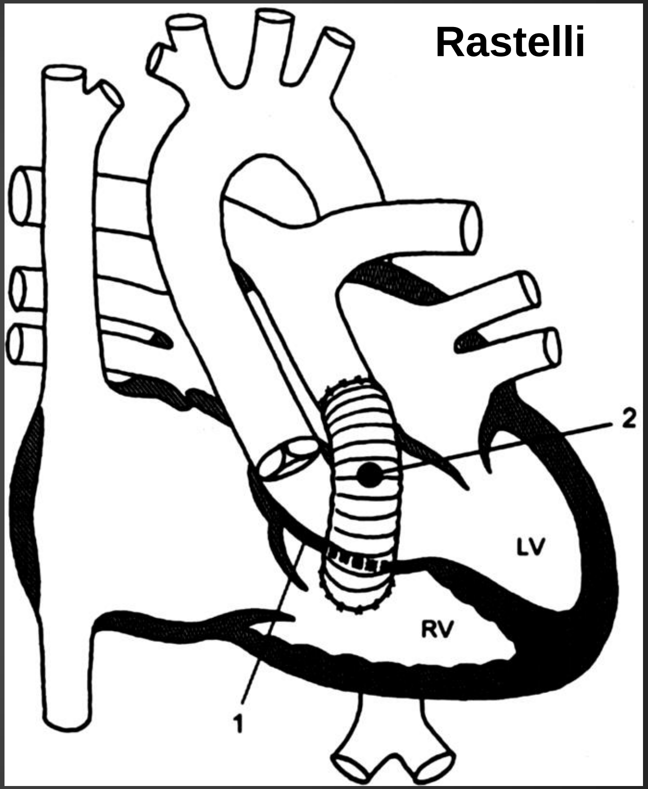


Situs inversus and CCTGA: Increased global myocardial thickening and base-to-apex shortening with significant decrease in RV end-systolic volume

Lancellotti et al. J Am Soc Echocardiogr 2017;30:101-38

Transposition of the Great Arteries





TGA s/p Mustard/Senning - Dobut

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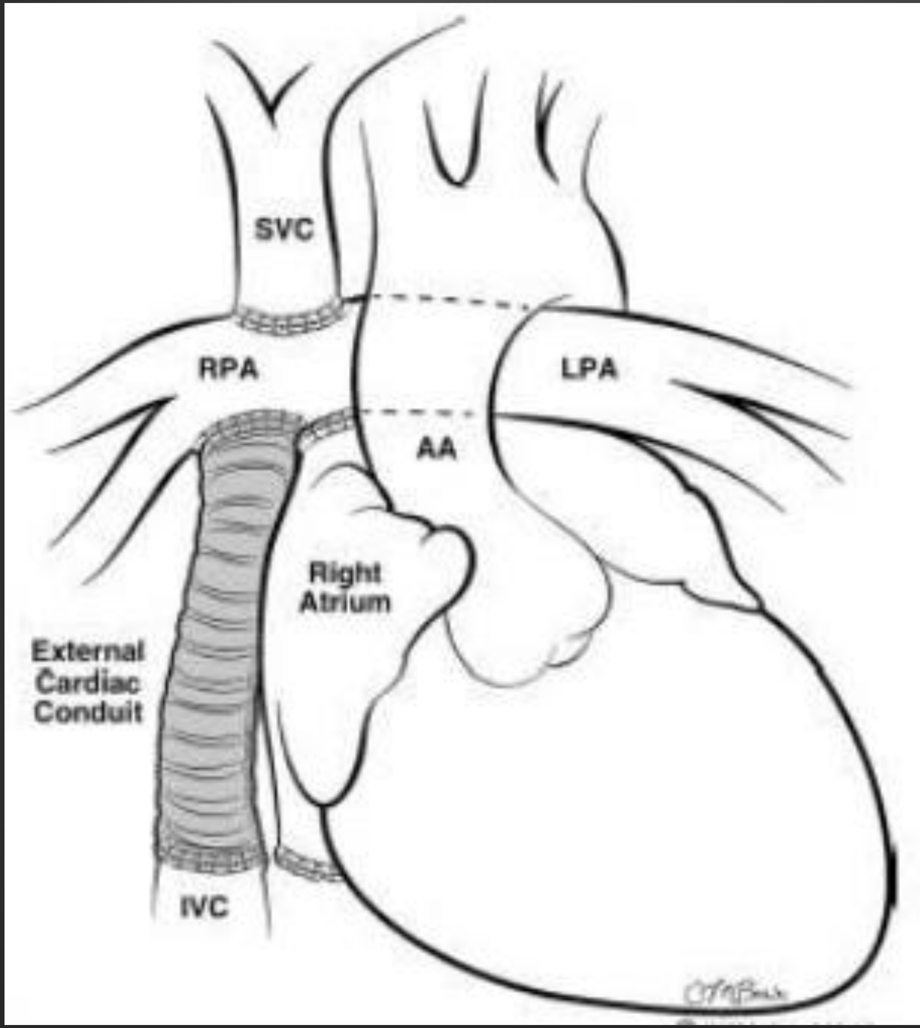
Relation of Biventricular Function Quantified by Stress Echocardiography to Cardiopulmonary Exercise Capacity in Adults With Mustard (Atrial Switch) Procedure for Transposition of the Great Arteries

	Controls		Patients		Patient Versus Control		Stress Versus Rest		Interaction Between Patient/Control and Stress/Rest	
	Rest Mean±SD	Stress Mean±SD	Rest Mean±SD	Stress Mean±SD	*F*	*P*	*F*	*P*	*F*	*P*
Systemic ventricle										
Systolic physiology										
Q to onset of shortening, ms	89.1±17.2	55±12.2	119.6±27.1	95.6±45.4	29.4	<0.001	29.0	<0.0001	0.9	ns
Long-axis excursion, cm	1.55±0.22	1.85±0.2	1.03±0.33	1.01±0.3	92.2	<0.0001	12.5	<0.001	16.1	<0.001
Ventricular systolic velocity, cm/s	8.5±1.8	14.6±3.9	8.4±3.1	10.4±3.7	7.2	0.01	54.7	<0.0001	14.3	<0.001
Peak ventricular outflow velocity, m/s	1.1±0.13	2.1±0.21	0.99±0.15	1.49±0.32	47.2	<0.0001	358.5	<0.0001	46.0	<0.0001
Diastolic physiology										
Time from S2 to onset of lengthening, ms	65.5±13.7	40.9±10.2	90.4±39.7	71.9±45.8	13.3	<0.001	15.2	<0.001	0.3	ns
Tissue Doppler early diastolic velocity, cm/s	13.4±3.6	15.1±2.9	10.8±3.5	12.8±3.5	7.0	<0.05	9.3	<0.01	0.1	ns
Tissue Doppler late diastolic velocity, cm/s	10.9±2.6	14±3.3	7.4±2.8	10.1±4	19.5	0.0001	13.5	<0.001	0.1	ns
Doppler E wave velocity, cm/s	62.7±15.5	82.3±17.2	79.6±29.6	95.4±28.7	6.5	<0.05	21.7	<0.0001	0.2	ns
Pulmonary ventricle										
Systolic physiology										
Q to onset of shortening, ms	82.3±21.9	48.6±11.7	68.1±24	61.9±14.4	0.2	ns	42.0	<0.0001	20.2	<0.0001
Long-axis excursion, cm	2.37±0.16	2.8±0.3	1.73±0.44	2.1±0.47	57.7	<0.0001	37.9	<0.0001	0.1	ns
Ventricular systolic velocity, cm/s	11.2±1.8	20.1±2.6	12.5±5.2	21.6±4.8	2.0	ns	176.3	<0.0001	0.0	ns
Peak ventricular outflow velocity, m/s	0.76±0.11	1.6±0.47	1.46±0.92	2.72±1.29	13.3	<0.001	80.0	<0.0001	2.9	ns
Diastolic physiology										
Time from S2 to onset of lengthening, ms	28.6±22.7	20±18.5	10±34.6	3±17.5	10.3	<0.01	3.2	ns	0.0	ns
Tissue Doppler early diastolic velocity, cm/s	11.6±2.7	14.1±3.5	17±6.4	19.4±3.3	22.9	<0.0001	9.6	<0.01	0.0	ns
Tissue Doppler late diastolic velocity, cm/s	11.5±2.5	19.9±4.9	10±4.4	13.4±3.3	15.0	<0.001	36.5	<0.0001	6.9	<0.05
Doppler E wave velocity, cm/s	43.8±12	60±14.1	81.9±23.8	97.7±26.3	59.9	<0.0001	20.1	<0.0001	0.0	ns

- 27 patients
- Age 29±7 yo
- Dobutamine
- Positive correlation of rest RV free wall long-axis excursion at rest with CPET

Wi et al. Circulation. 2004; 110:1380-1386

Univentricular Heart - Fontan



- Passive flow of blood through connection of SVC and IVC directly to the pulmonary arteries
- Use of the normal size ventricle (right or left) as systemic pump

Fontan - Dobutamine

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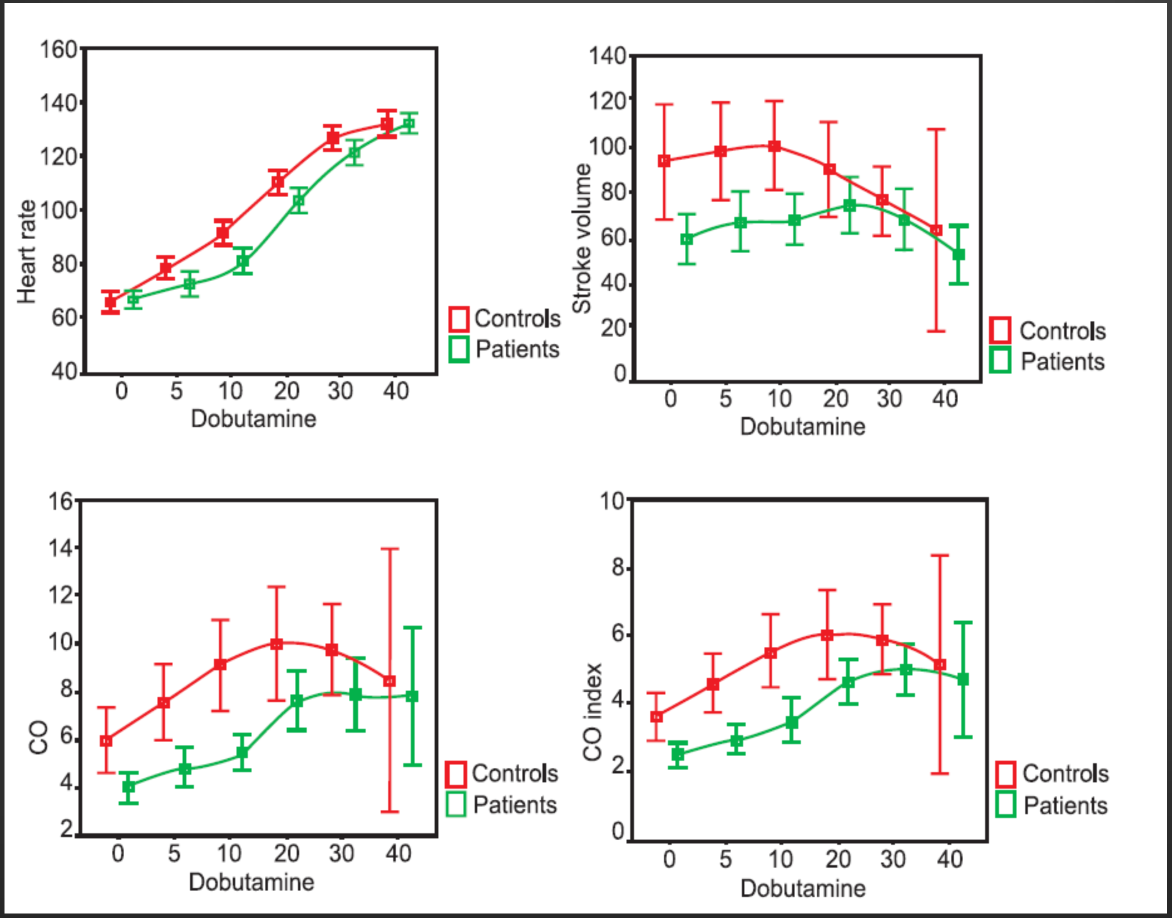
Dobutamine Stress Echocardiography for the Evaluation of Cardiac Reserve Late After Fontan Operation



10 patients

Age: 28 ±5 yo

Brili et al. Hellenic J Cardiol (2007) 48:252-7

Fontan - Dobutamine

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Two-Dimensional Global Longitudinal Strain Rate Is a Preload Independent Index of Systemic Right Ventricular Contractility in Hypoplastic Left Heart Syndrome Patients After Fontan Operation

Table 2. 2D Speckle Tracking

	Baseline				Dobutamine				Baseline vs Dobutamine
	n	Steady State	Preload Maneuver	*P*	n	Steady State	Preload Maneuver	*P*	*P*
Global strain, %	52	−17.70±3.36	−16.89±3.83	0.08	51	−18.37±3.9	−17.34±4.14	0.04	0.24
Global strain rate, s^{-1}	52	−1.30±0.29	−1.34±0.34	0.28	51	−2.26±0.49	−2.31±0.6	0.44	<0.001
Regional strain, %									
Basal septal	50	−12.5±6	−12.7±6.5	0.57	49	−11.1±7.5	−11.9±8.3	0.26	0.46
Mid septal	50	−17.6±4.8	−17.1±5.4	0.80	49	−16.9±6.8	−16.4±6.5	0.64	0.54
Apical septal	39	−21.1±8.4	−19.1±8	0.24	40	−22.0±7.2	−21.4±7.3	0.61	0.98
Apical lateral	45	−18.6±7.7	−17.2±8.4	0.06	44	−19.8±7.7	−20±7.5	0.75	0.35
Mid lateral	50	−19.0±5.2	−17.8±5.8	0.06	52	−20.2±6.2	−17.8±5.9	0.003	0.07
Basal lateral	49	−18.7±5.8	−18.9±5.2	0.95	51	−20.2±5.9	−17.5±6.4	0.01	0.03
Regional strain rate, s^{-1}									
Basal septal	51	−1.12±0.51	−1.23±0.74	0.27	50	−1.7±0.84	−1.97±1.37	0.11	<0.001
Mid septal	50	−1.31±0.39	−1.32±0.39	0.56	49	−2.29±0.88	−2.28±0.88	0.99	<0.001
Apical septal	39	−1.55±0.61	−1.61±0.72	0.46	40	−2.71±1.1	−2.67±0.9	0.98	<0.001
Apical lateral	45	−1.3±0.44	−1.38±0.57	0.68	44	−2.46±1.01	−2.56±0.89	0.88	<0.001
Mid lateral	50	−1.25±0.36	−1.26±0.41	0.84	52	−2.2±0.61	−2.16±0.77	0.79	<0.001
Basal lateral	49	−1.3±0.37	−1.29±0.38	0.78	51	−2.41±0.72	−2.29±0.79	0.44	<0.001

52 patients

Median age: 6.6 (2.9–22) yrs

Schlangen et al. Circ Cardiovasc Imaging. 2014;7:880-886

Surveillance frequency in repaired CHD

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Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography
Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

Modality	Age (y) <2 y*	2–9	10–19	20–49	≥50
Echocardiography	12 mo	12 mo	24 mo	24 mo	24 mo
CMR	Not recommended routinely; ordered to address specific questions not answered by echocardiography		• 36 mo in stable patients • 12 mo if moderate (≥150 mL/m^2) or progressive (increase of >25 mL/m^2) RV dilatation or dysfunction (RV EF ≤ 48% or ≥6% decrease in EF)		
CT	Not recommended routinely; ordered when CMR is indicated but cannot be performed (e.g., metallic artifacts or contraindications to CMR)				
Lung perfusion scan	If predicted RV systolic pressure 60% systemic or smallest branch PA diameter Z score < −2.5; in patients ≥10 y of age, consider CMR flow measurements				
X-ray angiography	Not recommended routinely†; ordered when noninvasive methods either cannot be performed or have failed to provide satisfactory diagnostic data				Coronary angiography when clinically indicated
Chest radiography	Not recommended routinely; may be ordered for evaluation of stent integrity				

J Am Soc Echocardiogr 2014;27:111-41

Comparison of imaging modalities

Characteristic	Echocardiography	CMR	CT	Nuclear scintigraphy
Availability	++++	++	++	+++
Portability	++++	–	–	–
Cost (relative value units)*	9.11[†]	22.51[‡]	14.39[§]	13.59[‖]
Radiation risk	–	–	++++	++++
Artifacts from stainless-steel implants	+	+++	+	–
Sedation requirements in young children	++	++++	+++	++
Spatial resolution (mm)	<1	<1–2	<1	5–10
Temporal resolution (msec)	20	30	75–175	–[¶]
RV size/function	++	++++	+++	+
RV pressure	+++	+	+	–
TR severity	+++	+++	–	–
Mechanism of TR	++++	++	–	–
PR severity	++	++++	–	–
Branch PAs flow quantification	–	+++	–	++++
LV size/function	+++	++++	+++	++
Coronary origins and proximal course	++	+++	++++	–
Aortic dimensions	+++	++++	++++	–
Residual shunts	+++	+++	+	–
Pulmonary-to-systemic flow ratio	+	++++	+[#]	+
Aortopulmonary collateral vessels	–	+++	+++	–
Myocardial viability	+	++++	+	+++

J Am Soc Echocardiogr 2014;27:111-41

Conclusions

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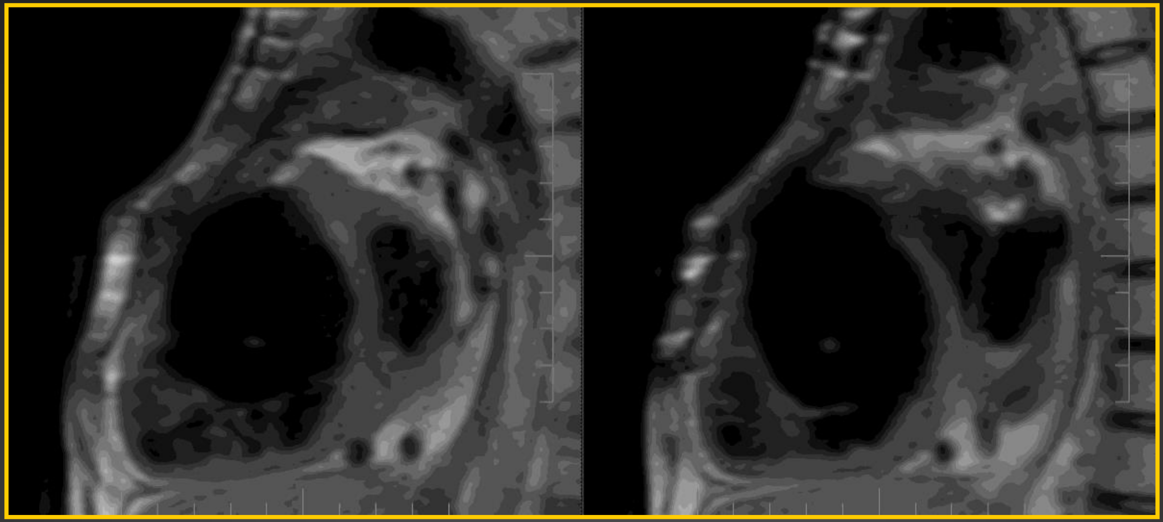
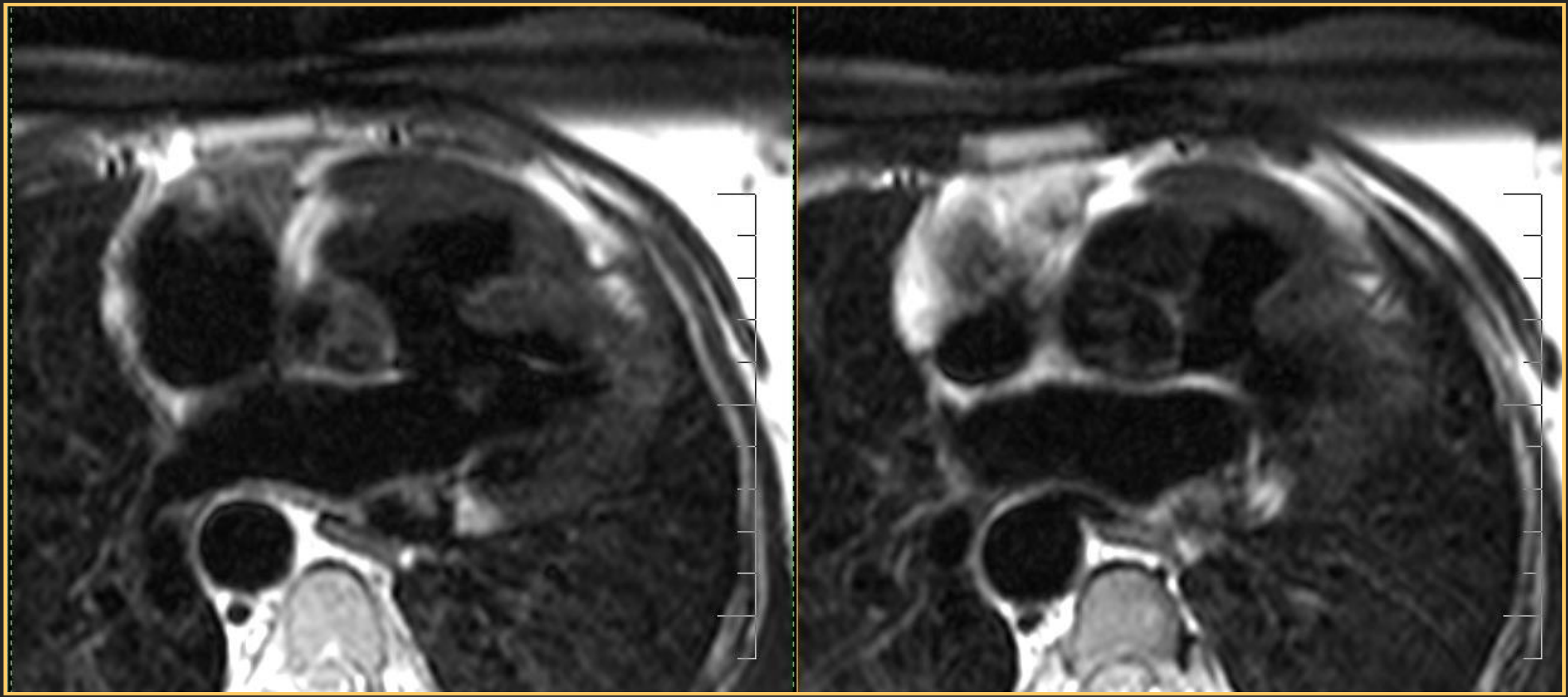
- Multimodality approach in imaging in repaired CHD
- No single test gives all necessary information
- Choice influenced by:
  - age, need for sedation, acoustic windows
  - clinical question
  - local availability and expertise
  - cost
  - radiation exposure
  - pacemakers/defibrillators
- Research to identify predictors of deterioration vs stable course in this growing population

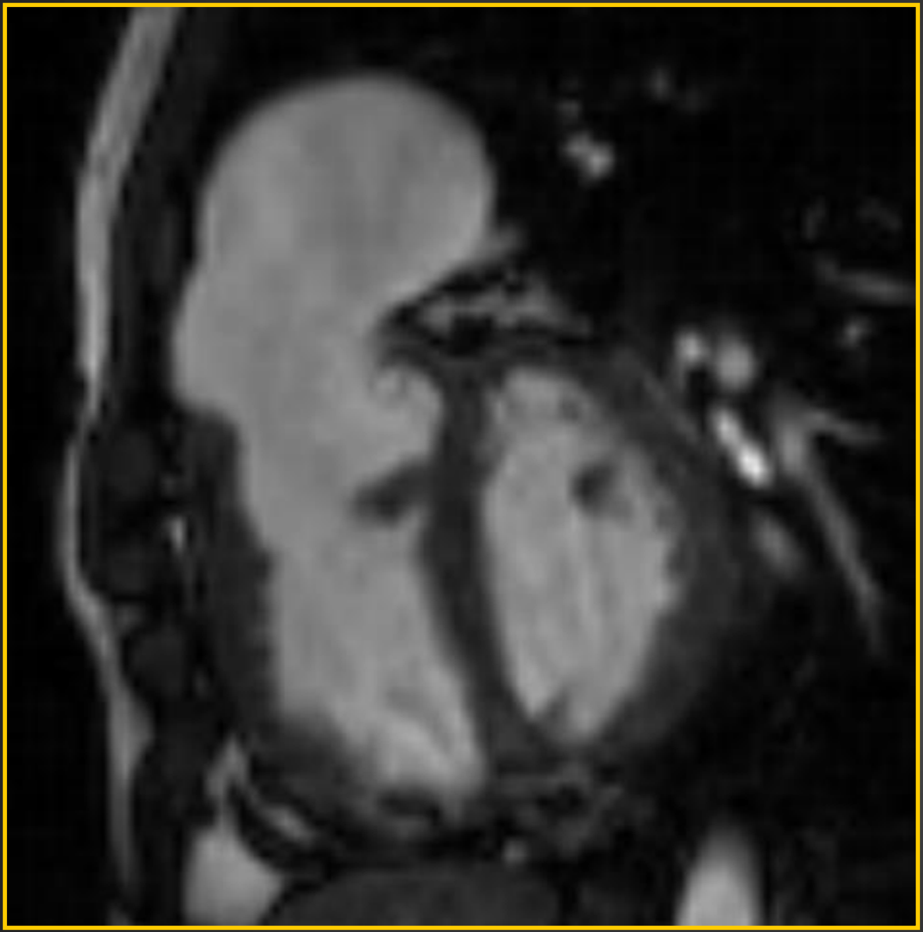
TOF CMR

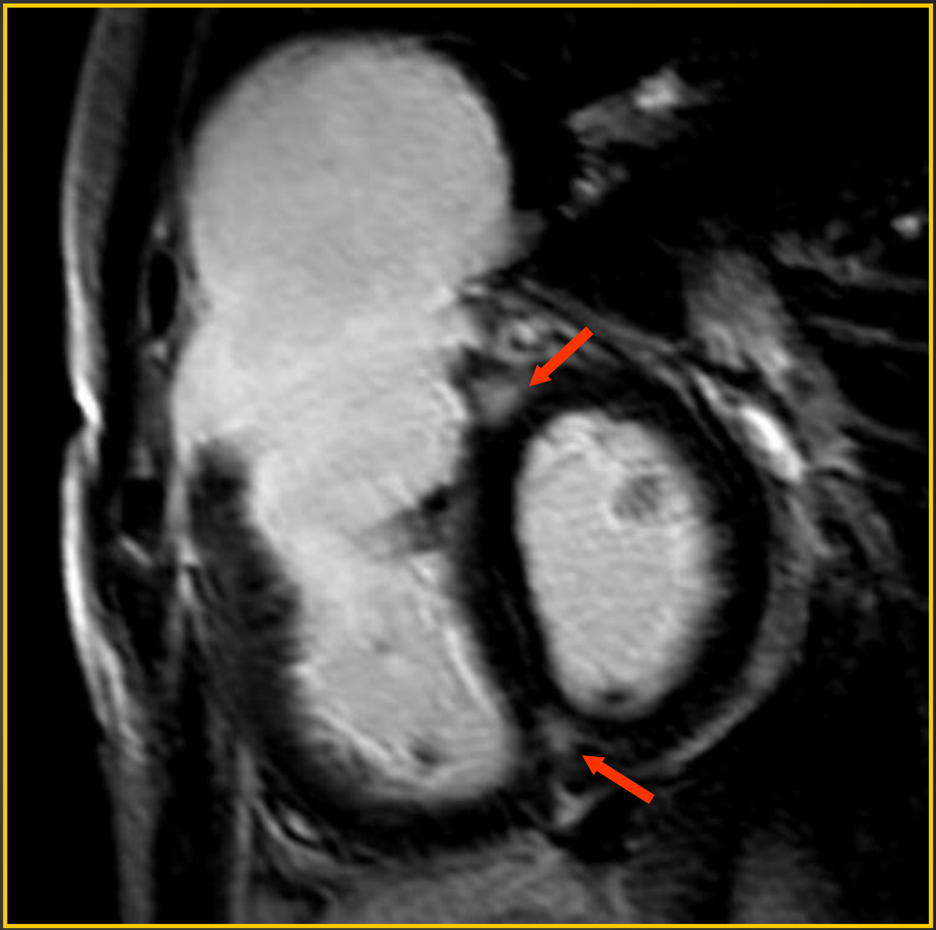
- Steady-state free precession (SSFP) → , which is a type of gradient-echo technique characterized by high signal-to-noise ratio, high T2/T1 contrast ratio, and sharp borders between the blood pool and the myocardium
- Electrocardiographically gated SSFP can be used as a cine magnetic resonance sequence, which is typically used for assessment of ventricular size and function, valve function, and intracardiac and extracardiac anatomy
- electrocardiographically gated, respiratory-navigated SSFP sequence can yield a high–spatial resolution static 3D data set, which is often used for detailed assessment of intracardiac anatomy and/or coronary artery anatomy.
- Electrocardiographically gated turbo (fast) spin-echo (TSE) imaging offers high spatial resolution (submillimeter in-plane), excellent contrast between elements of soft tissue, and decreased sensitivity to metallic artifacts compared with gradient-echo sequences, although it provides only static images.
- Contrast-enhanced magnetic resonance angiography (MRA) represents a robust 3D technique
- Electrocardiographically gated, phase-contrast (PC) flow measurements are used for measurements of blood flow, including flow rates in the great arteries and veins, differential PA flow, and regurgitant volumes (e.g., PR, AR





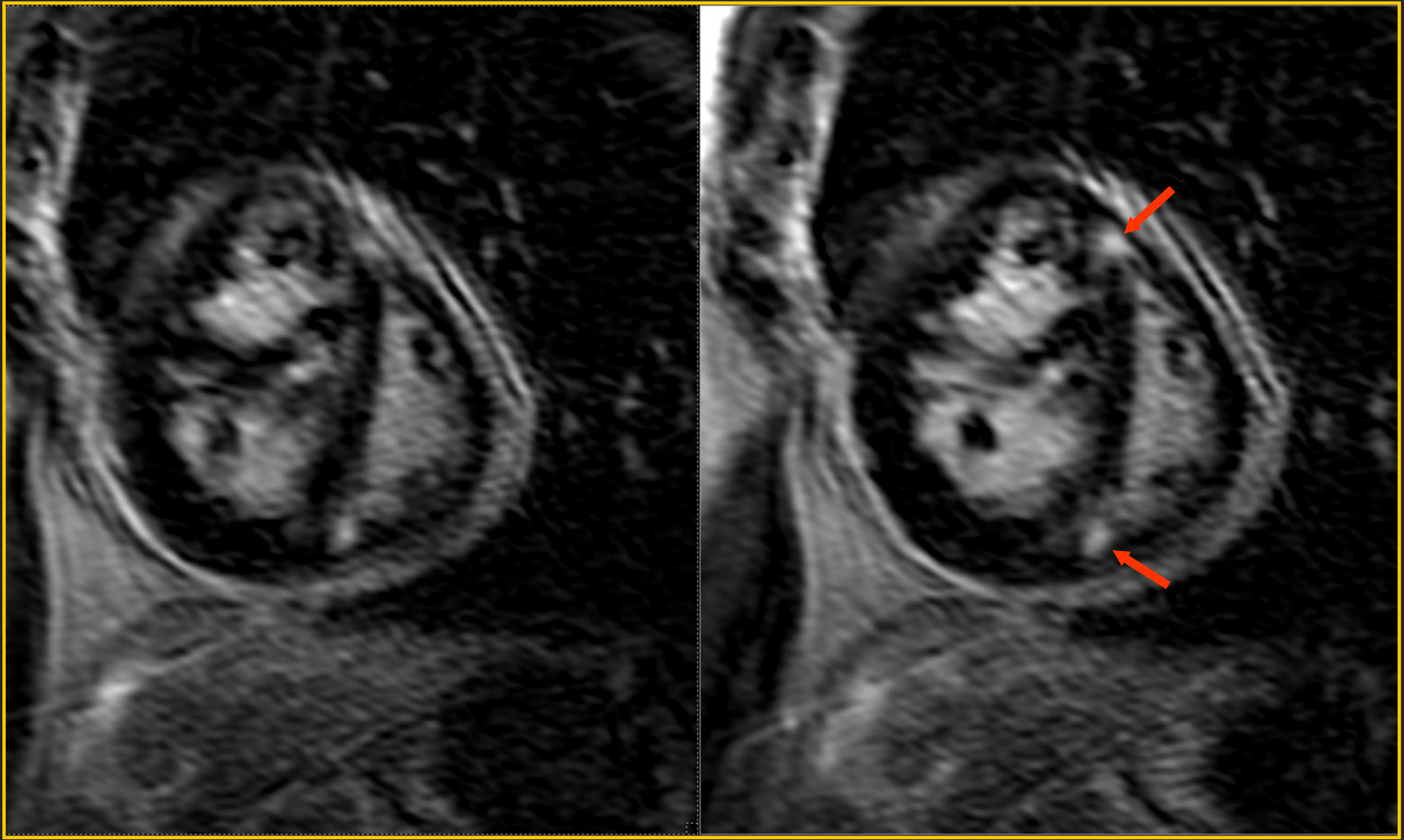
CMR - Myocardial Enhancement





Myocardial disarray and plexiform fibrosis

CMR - Myocardial Enhancement



Myocardial disarray and plexiform fibrosis

Comprehensive ECHO views

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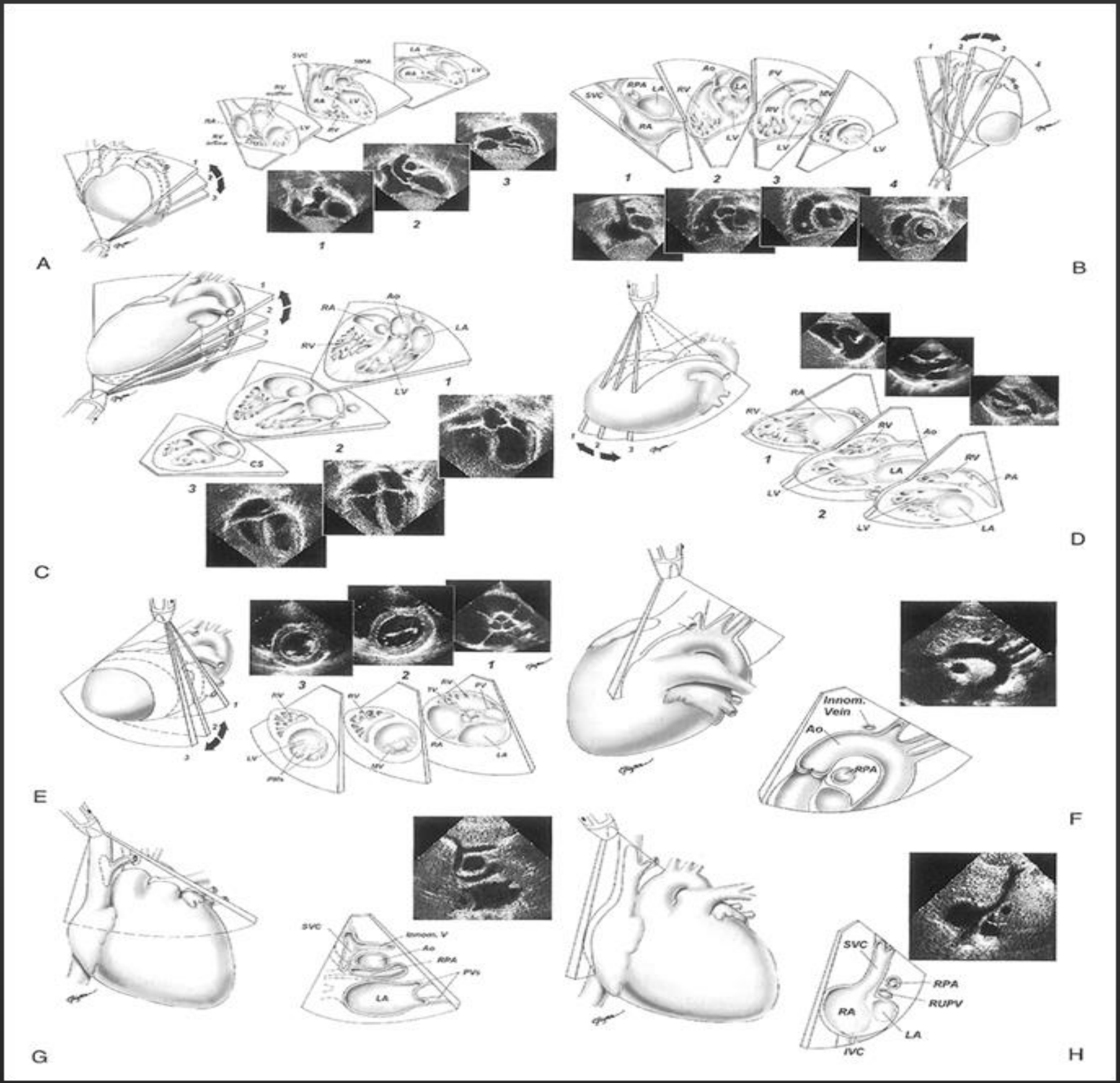
Subxiphoid coronal

4 chamber

Parasternal short axis

Suprasternal short axis



Subxiphoid saggital

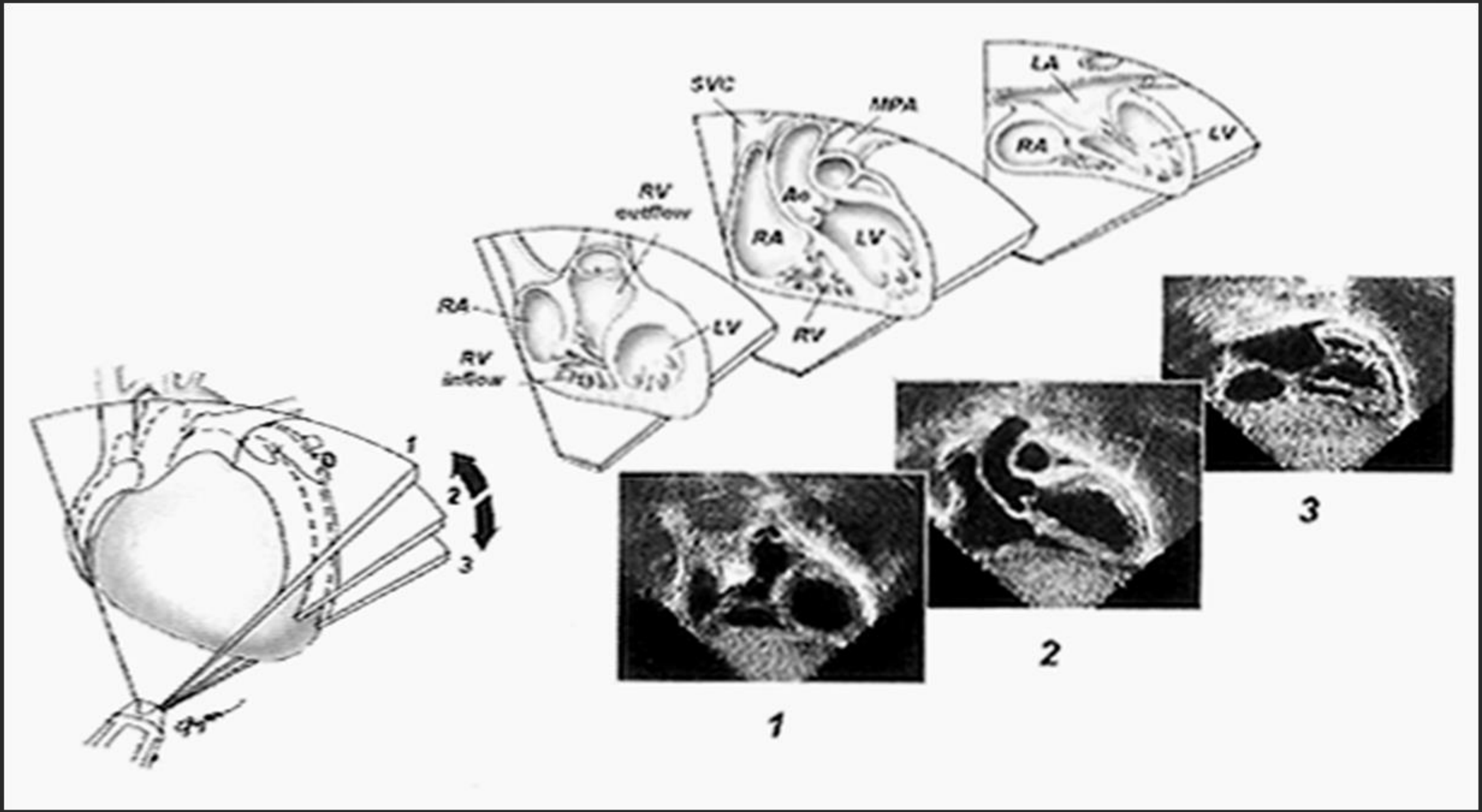
Parasternal long axis

Suprasternal long axis

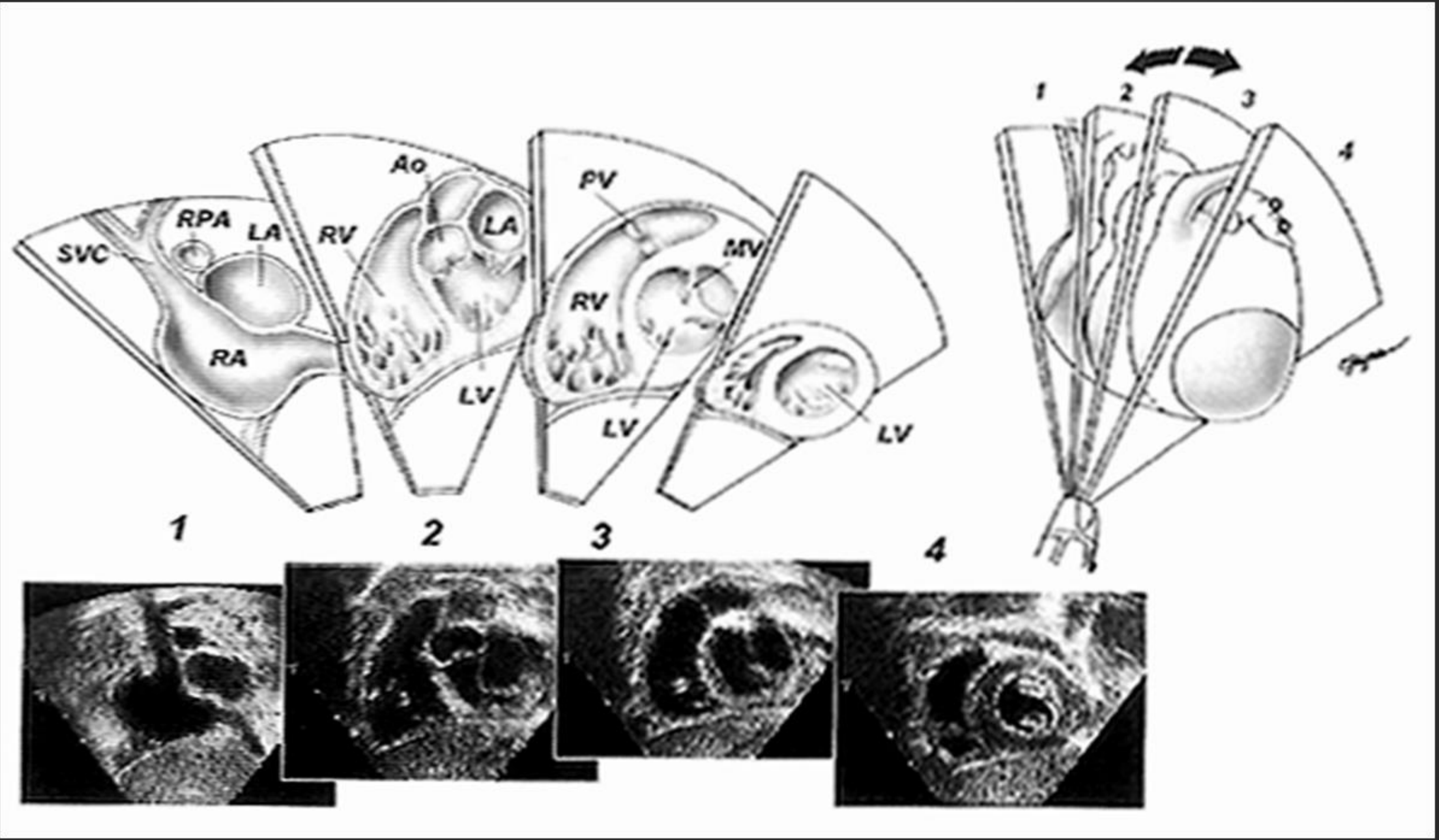
Suprasternal right

J Am Soc Echocardiogr 2006;19:1413-30

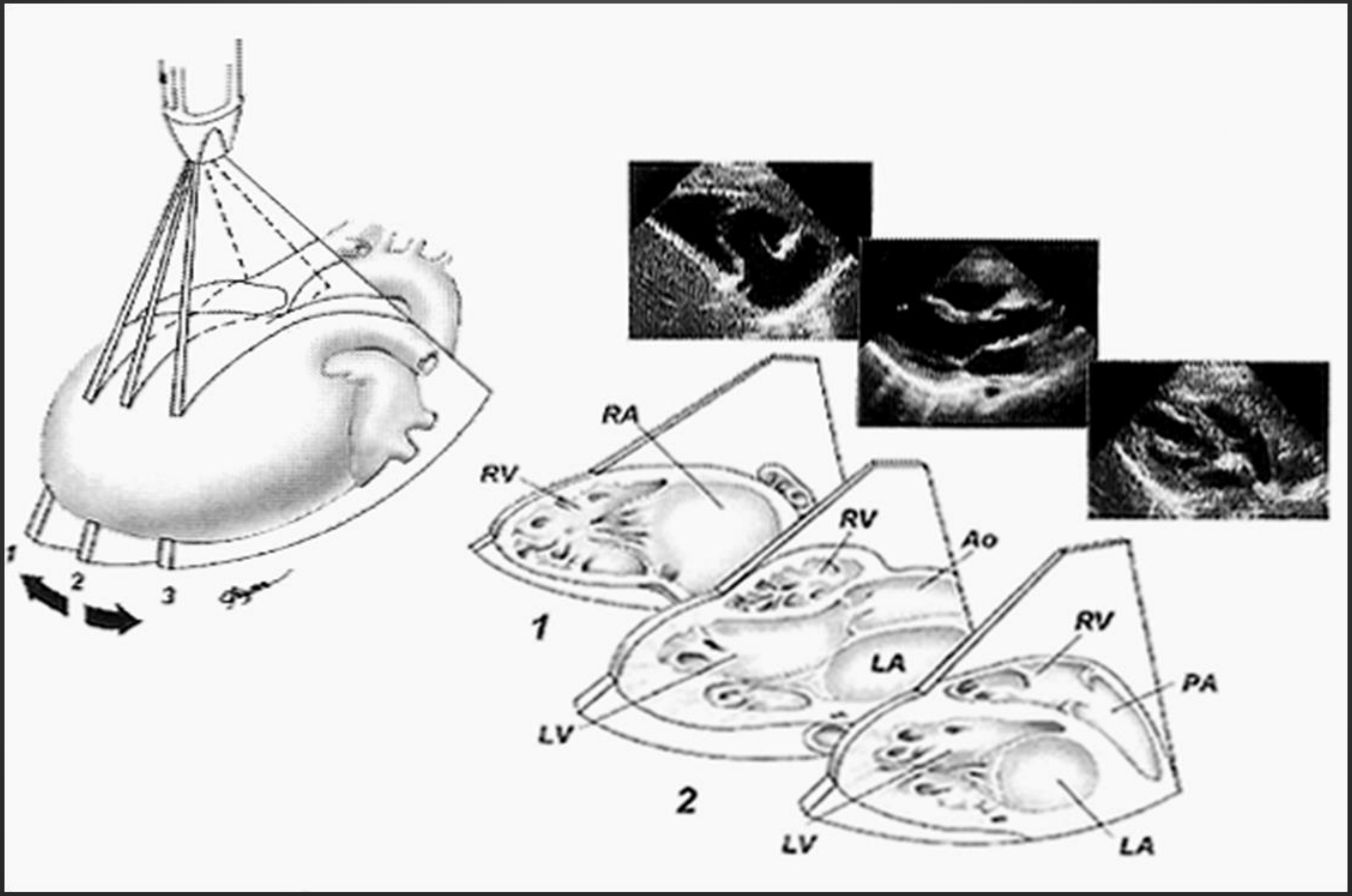
Υποξιφοειδική Απεικόνιση – Στεφανιαίες τομές



Υποξιφοειδική λήψη- Οβελιαίες τομές



Παραστερνική Τομή κατά το Μακρύ Άξονα



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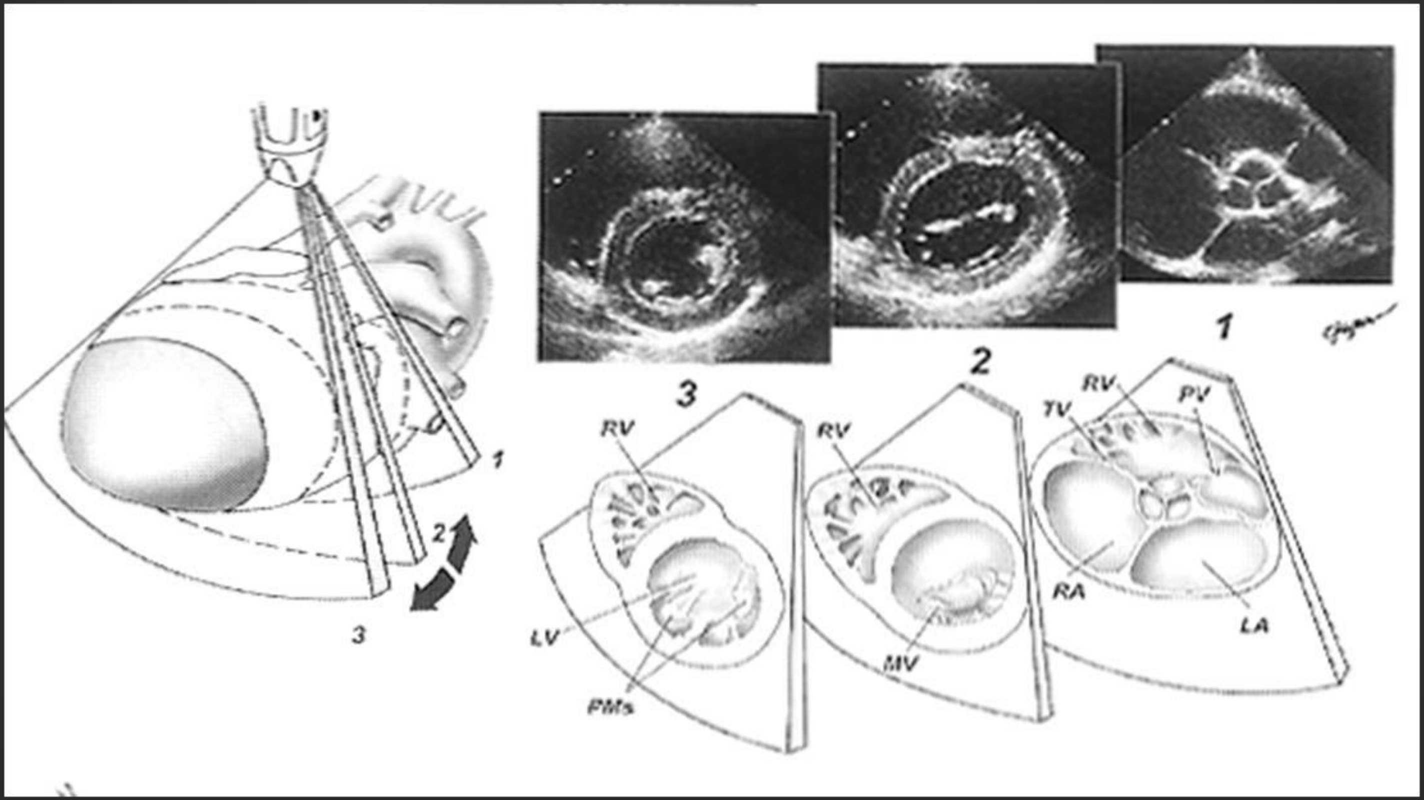
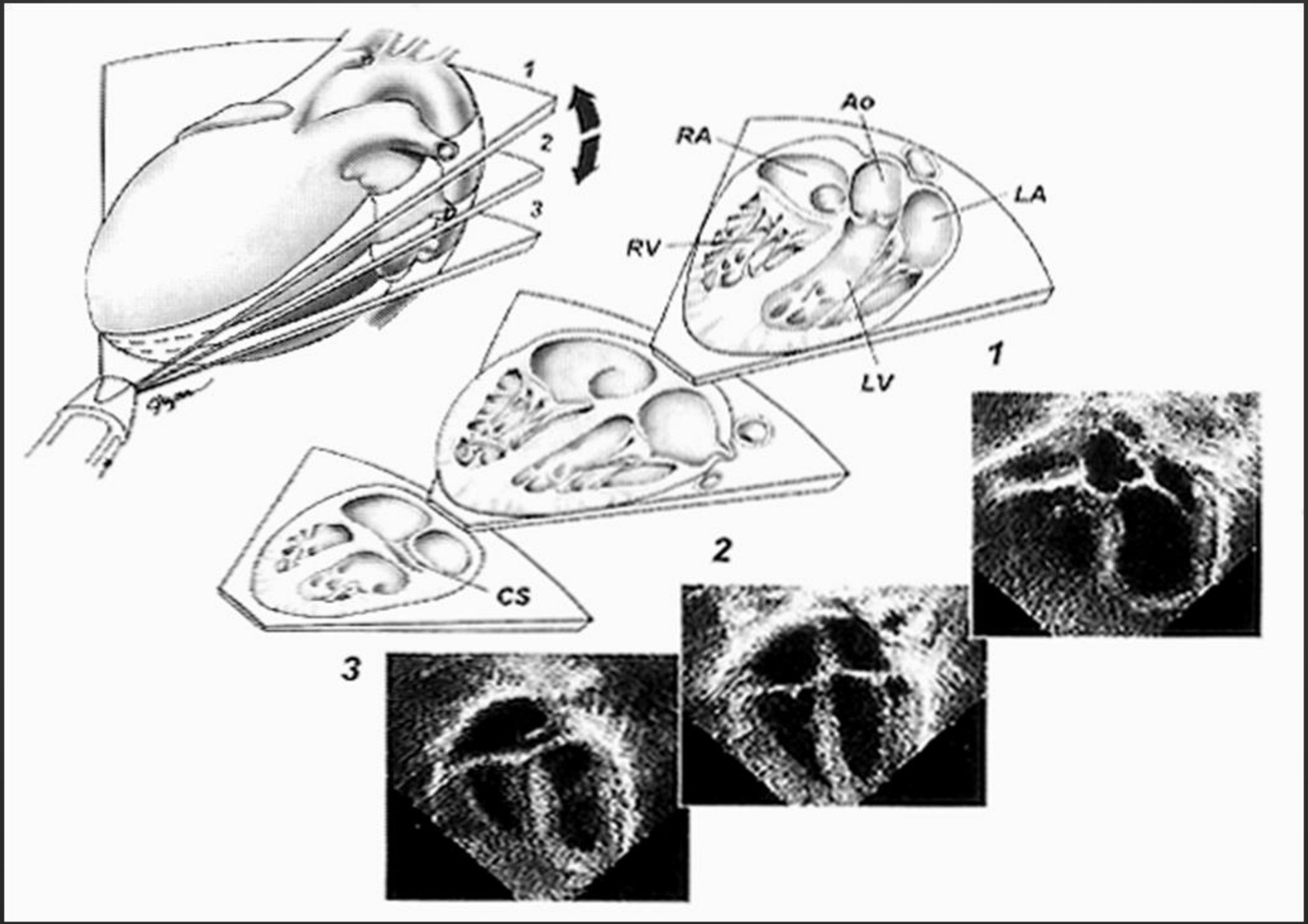


Κορυφαία Λήψη Τεσσάρων Κοιλοτήτων



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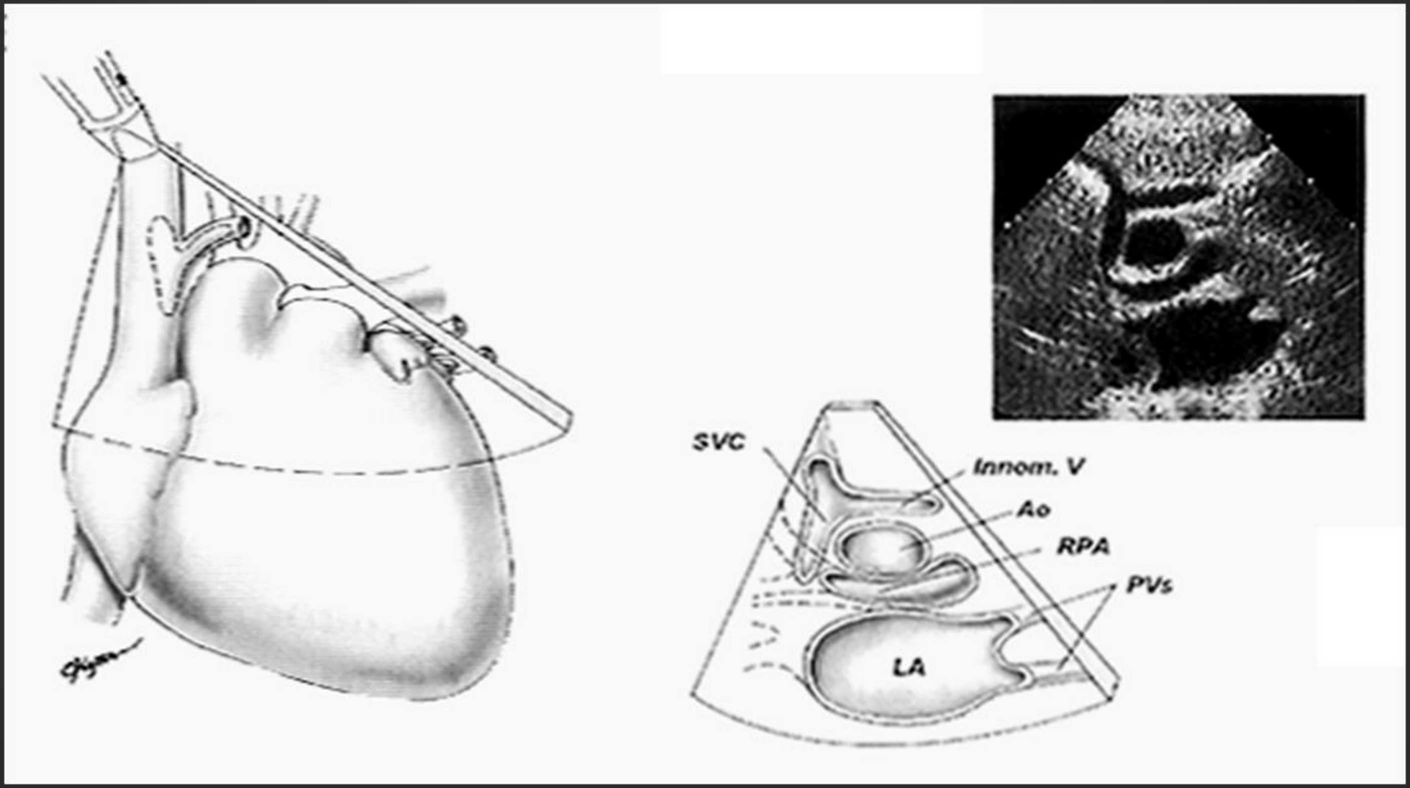
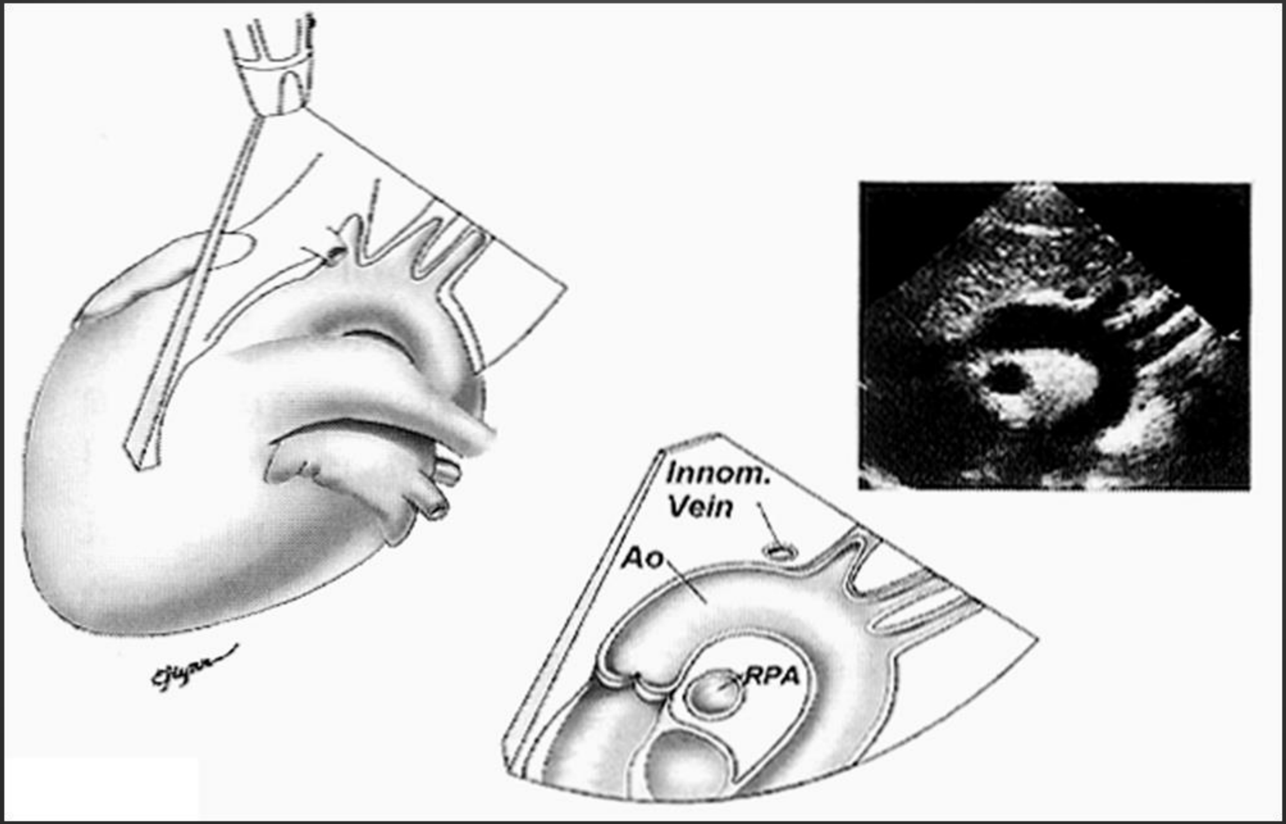
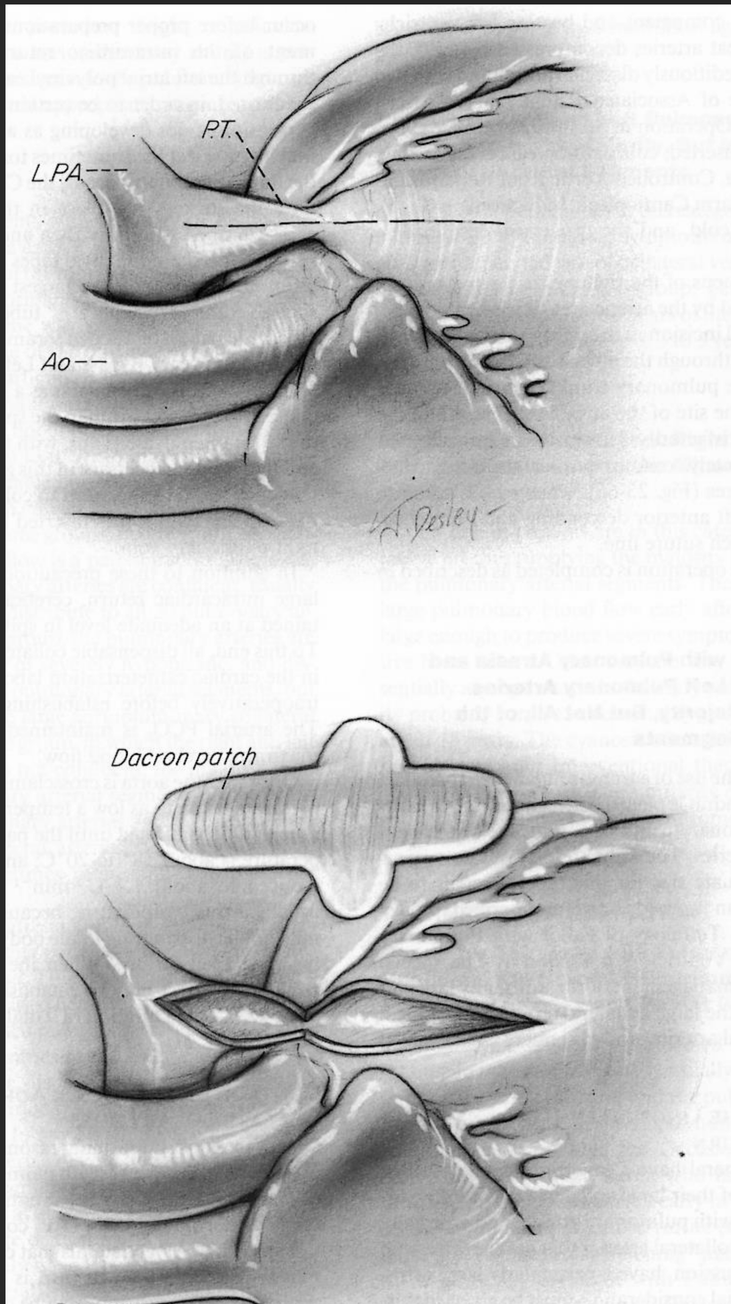
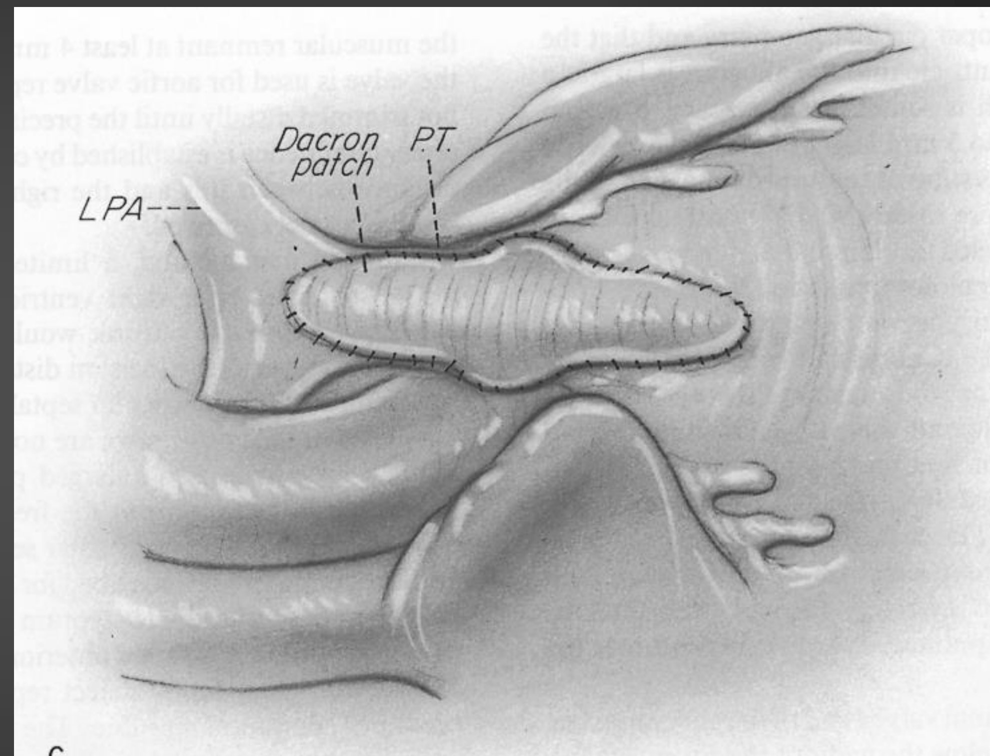


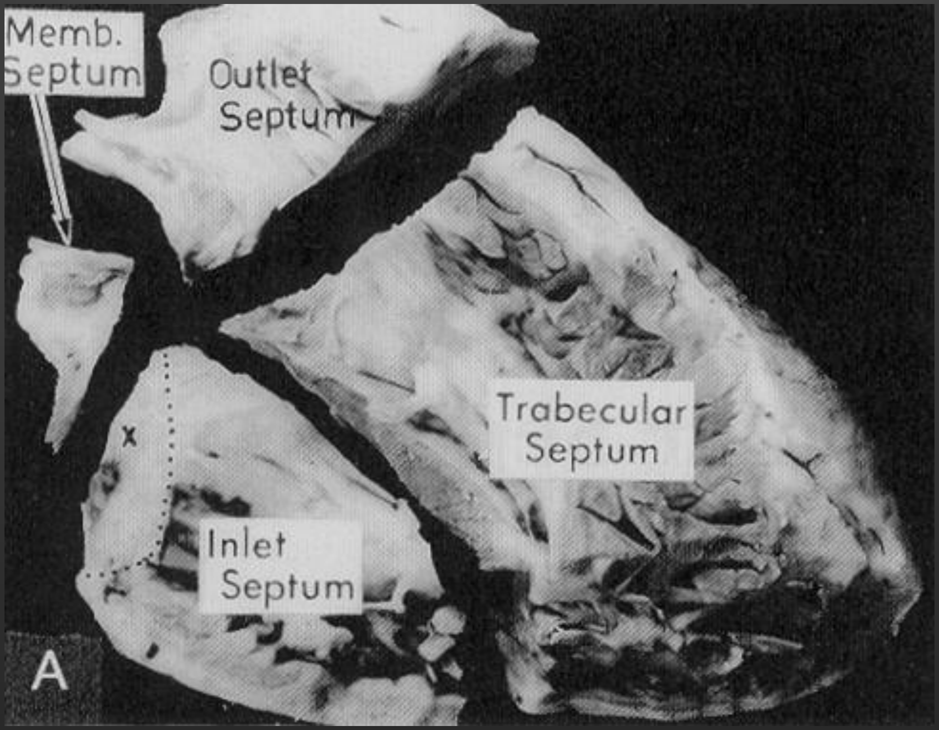
Οβελιαία Υπερστερνική Λήψη



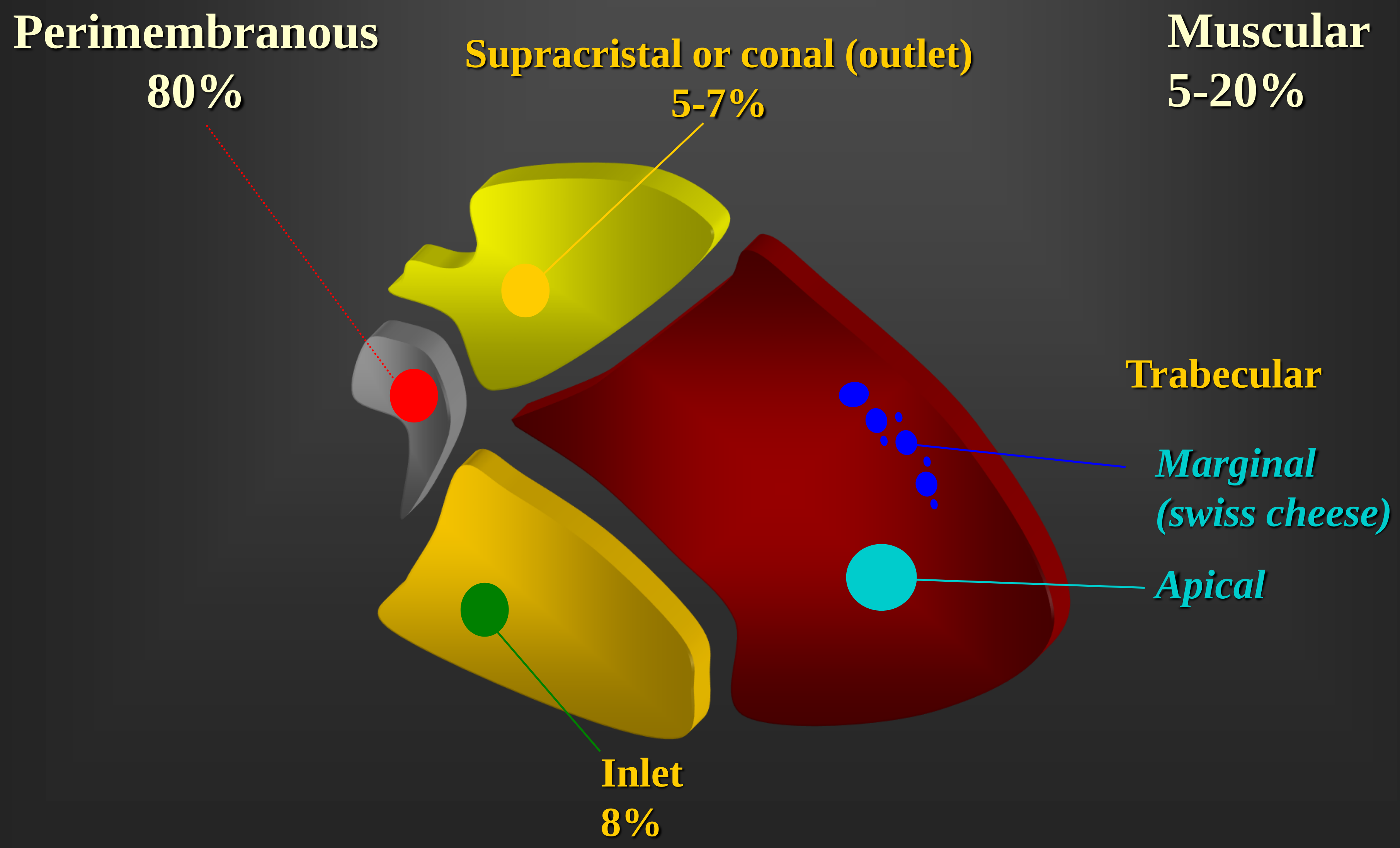

P.T.
L.P.A.
Ao
Dacron patch

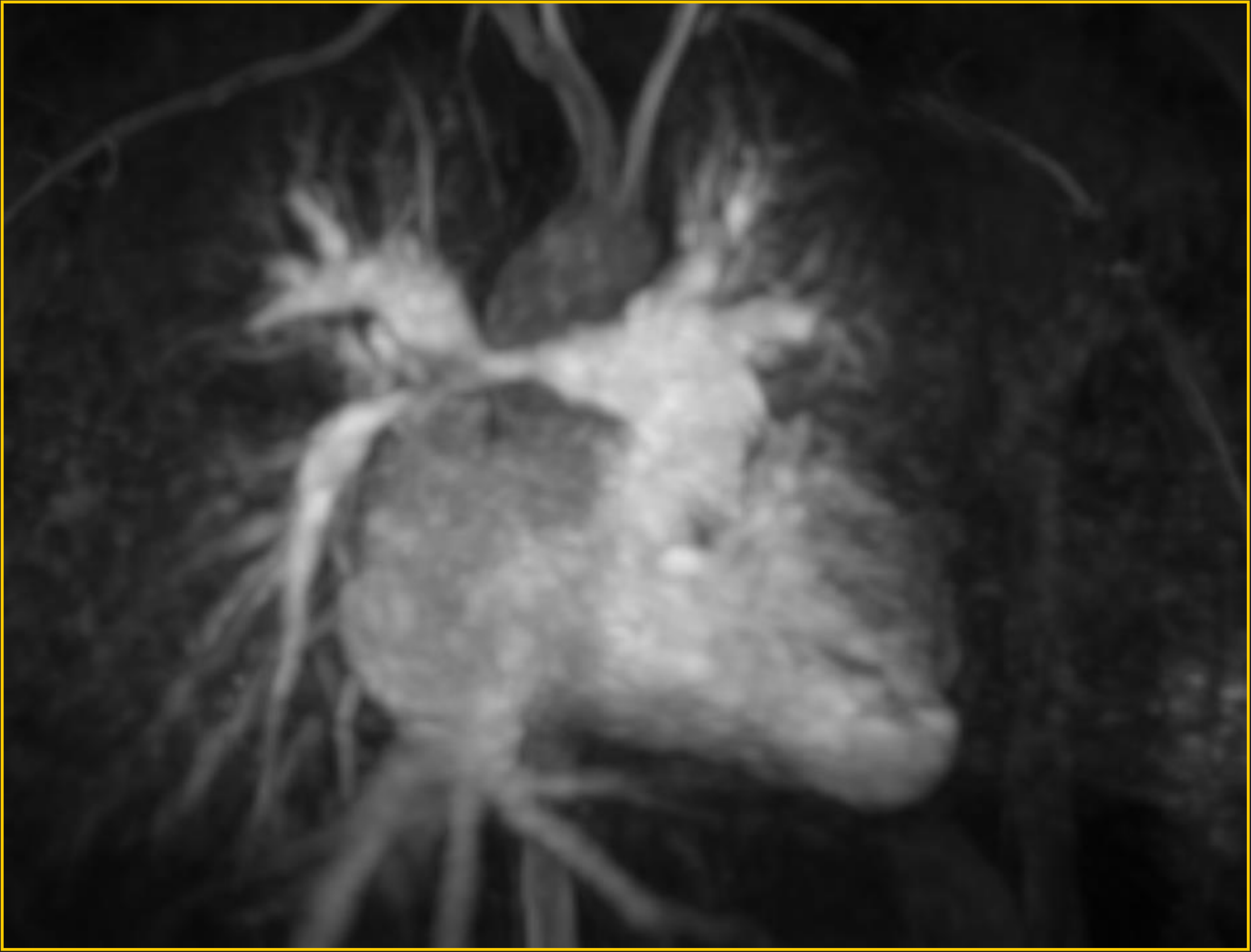

Dacron patch
P.T.
L.PA
c

Ventricular Septal Defects (VSD)



Ventricular Septal Defects (VSD)





Guidelines Repaired TOF

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GUIDELINES AND STANDARDS

Multimodality Imaging Guidelines for Patients with Repaired Tetralogy of Fallot: A Report from the American Society of Echocardiography

Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology

Anne Marie Valente, MD, FASE, Co-Chair, Stephen Cook, MD, Pierluigi Festa, MD, H. Helen Ko, BS, RDMS, RDCS, FASE, Rajesh Krishnamurthy, MD, Andrew M. Taylor, MD, Carole A. Warnes, MD, Jacqueline Kreutzer, MD, and Tal Geva, MD, FASE, Co-Chair, *Boston, Massachusetts; Pittsburgh, Pennsylvania; Massa, Italy; New York, New York; Houston, Texas, London, United Kingdom; Rochester, Minnesota*

(J Am Soc Echocardiogr 2014;27:111-41.)

Imaging surveillance frequency in repaired TOF

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Echocardiography	12 mo	12 mo	24 mo	24 mo	24 mo
CMR	Not recommended routinely; ordered to address specific questions not answered by echocardiography		• 36 mo in stable patients • 12 mo if moderate (≥150 mL/m^2) or progressive (increase of >25 mL/m^2) RV dilatation or dysfunction (RV EF ≤ 48% or ≥6% decrease in EF)		
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J Am Soc Echocardiogr 2014;27:111-41

Goals of Imaging

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- Intracardiac and extracardiac shunts
- TR (degree and mechanism), estimated RVp
- RV evaluation
  - - size and function
  - - regional RV wall motion abnormalities
  - - RVOT (obstruction and/or aneurysm)
- Degree of PR
- Assessment of the main and branch PAs
- LV size and function
- Aorta
  - - size of the aortic root and ascending aorta
  - - degree of AR
  - - aortic arch sidedness
- Origin and proximal course of LCA/RCA
- Systemic-to-pulmonary collateral vessels
- Assessment of myocardial viability

Tetralogy of Fallot

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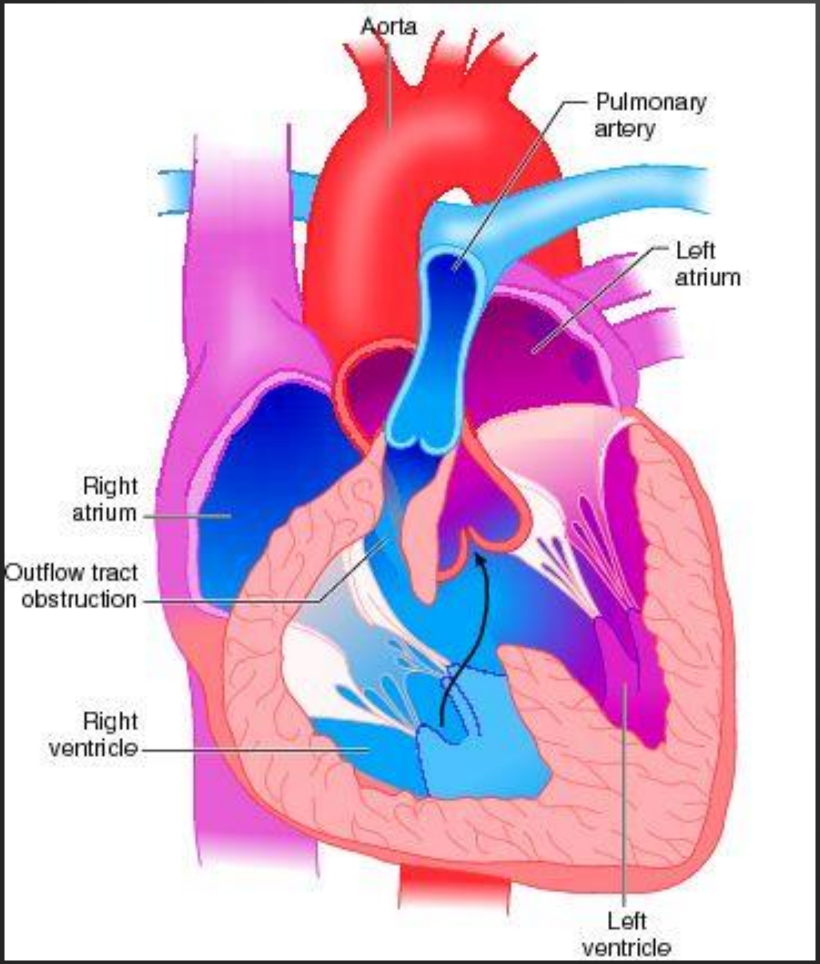
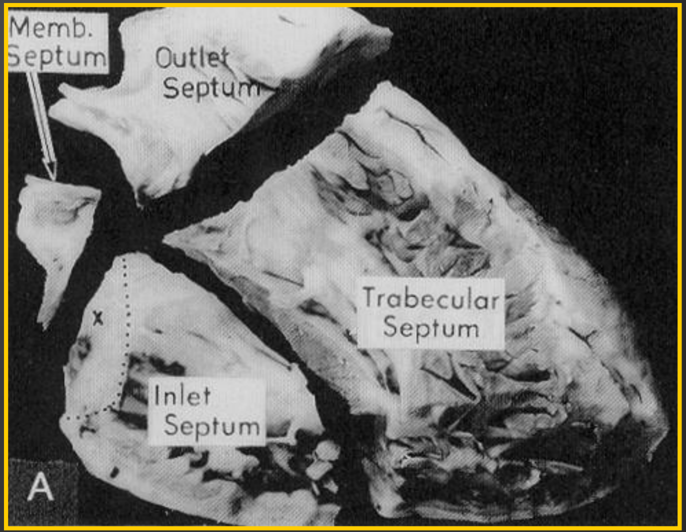
= anterior malalignment of the outlet septum

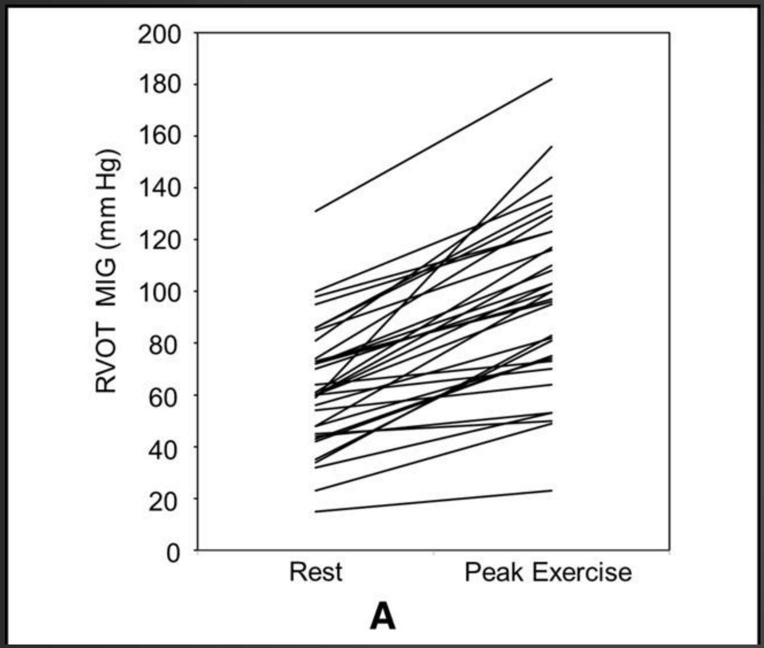


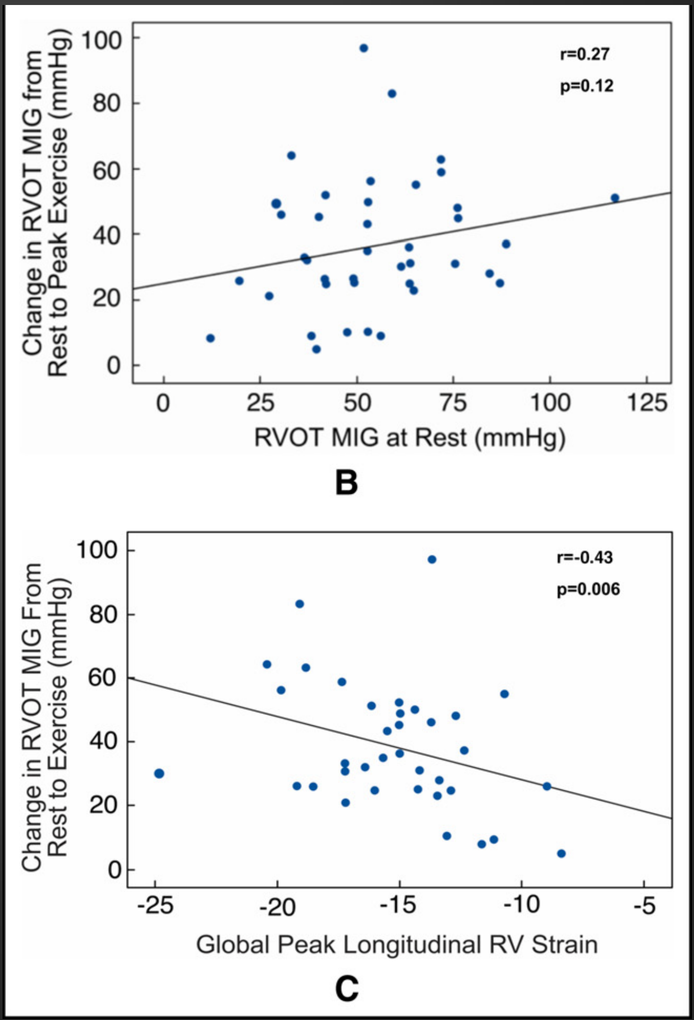


1. Ventricular septal defect
2. Narrow RV outflow tract
3. Aorta overriding interventricular septum
4. Right ventricular hypertrophy

Conduit stenosis and RV fxn -

Exercise Stress Echocardiographic Assessment of Outflow Tract and Ventricular Function in Patients With an Obstructed Right Ventricular-to-Pulmonary Artery Conduit After Repair of Conotruncal Heart Defects

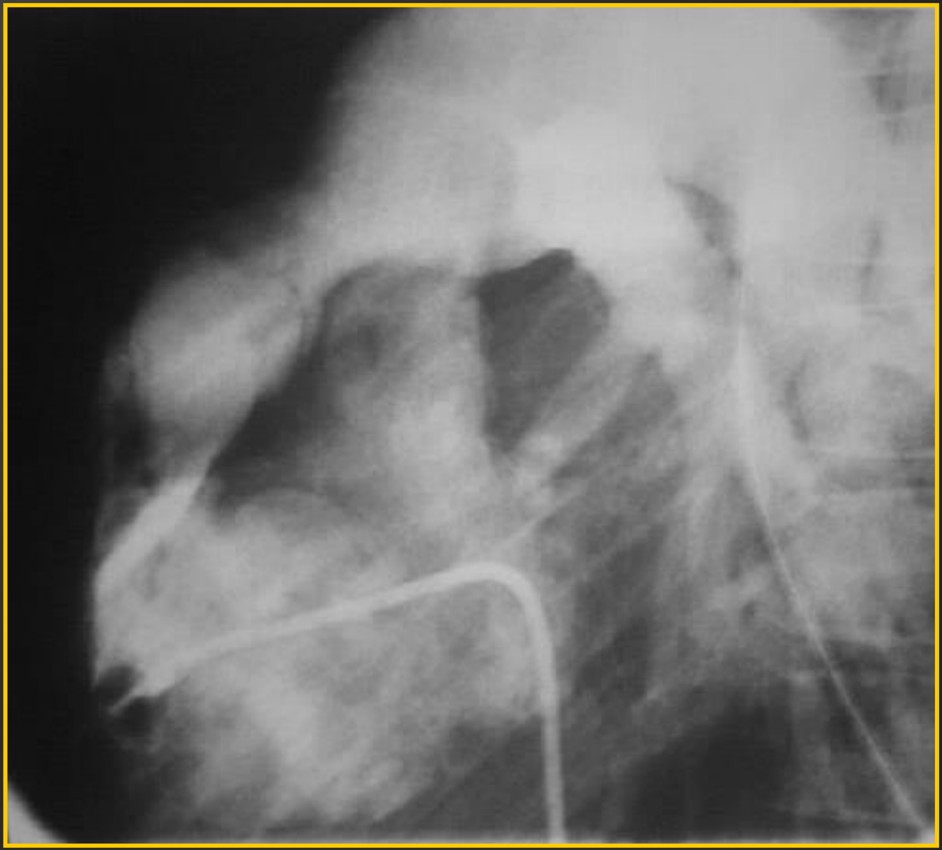




- 35 patients with conduits
- Median age 17 (6-56) yo
- - 25 Fallot
  - 9 truncus arteriosus
  - 6 TGA

Hasan et al. Am J Cardiol 2012;110:1527–1533

TOF – X-Ray Angiography



TOF – Angio – PAs – MAPCAs

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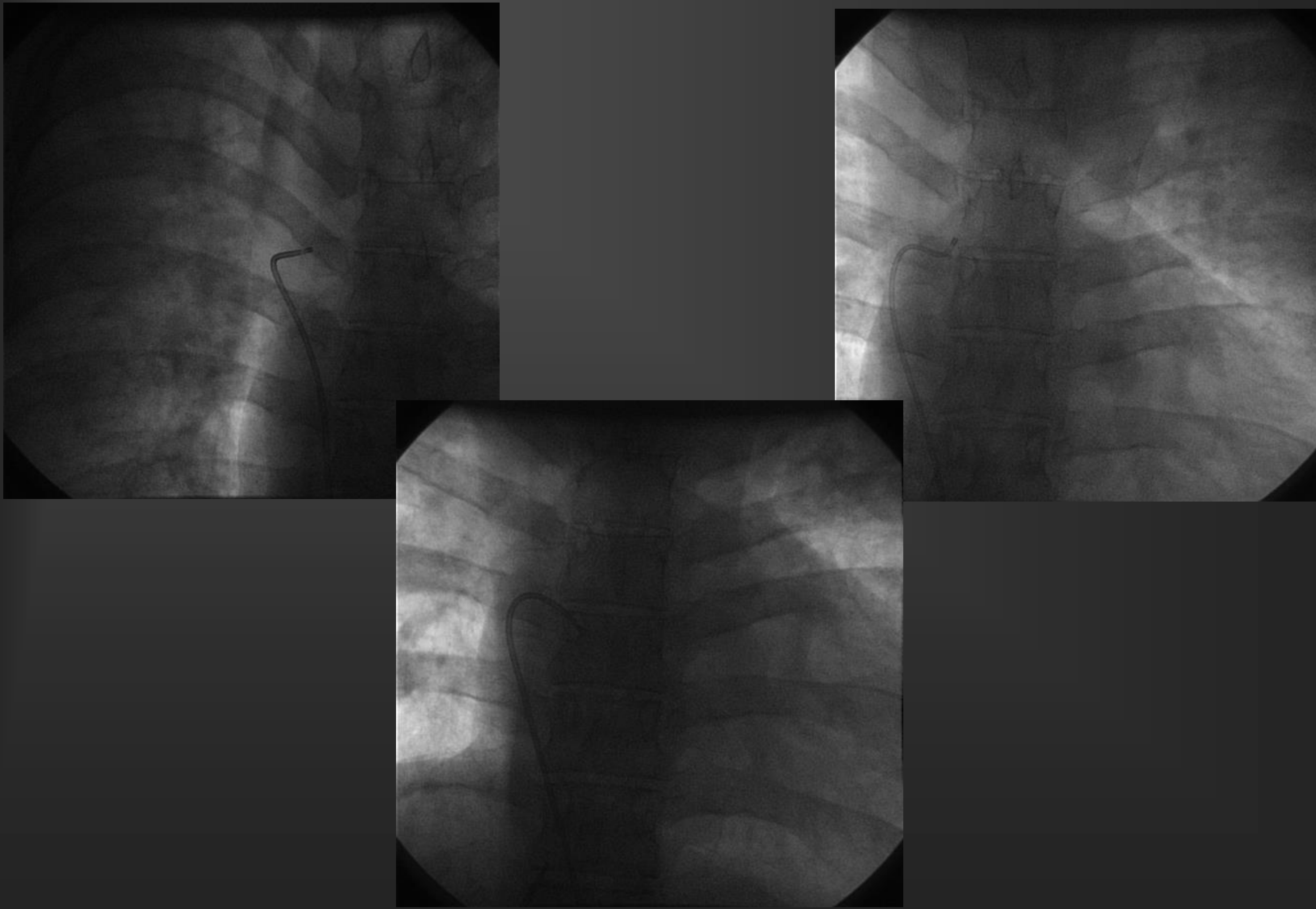

TOF - 3D printing

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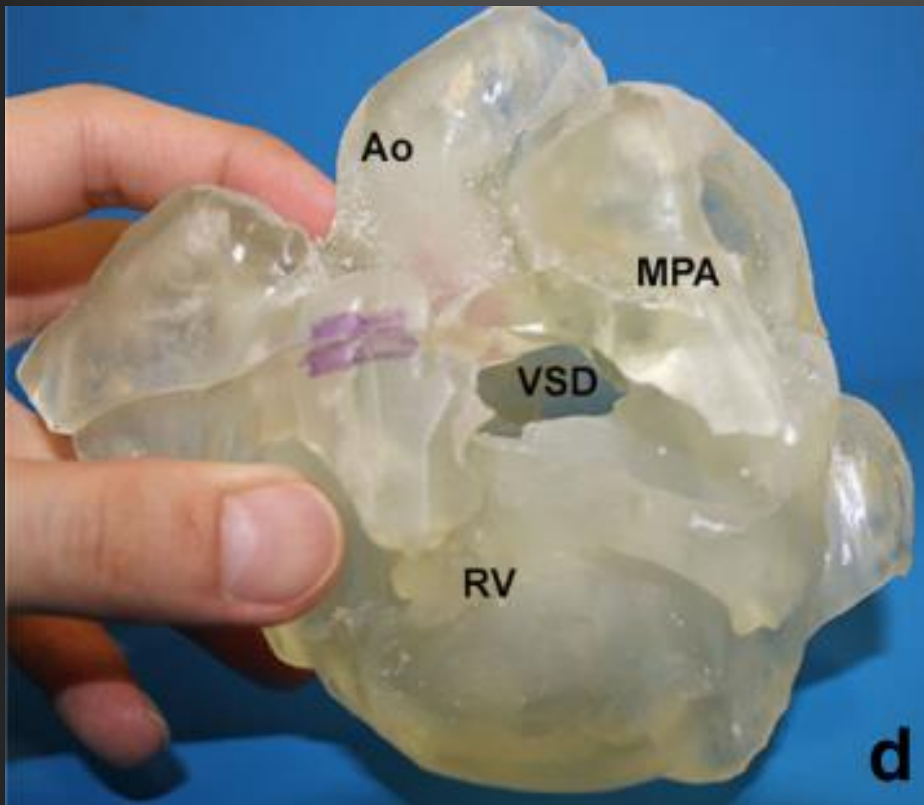
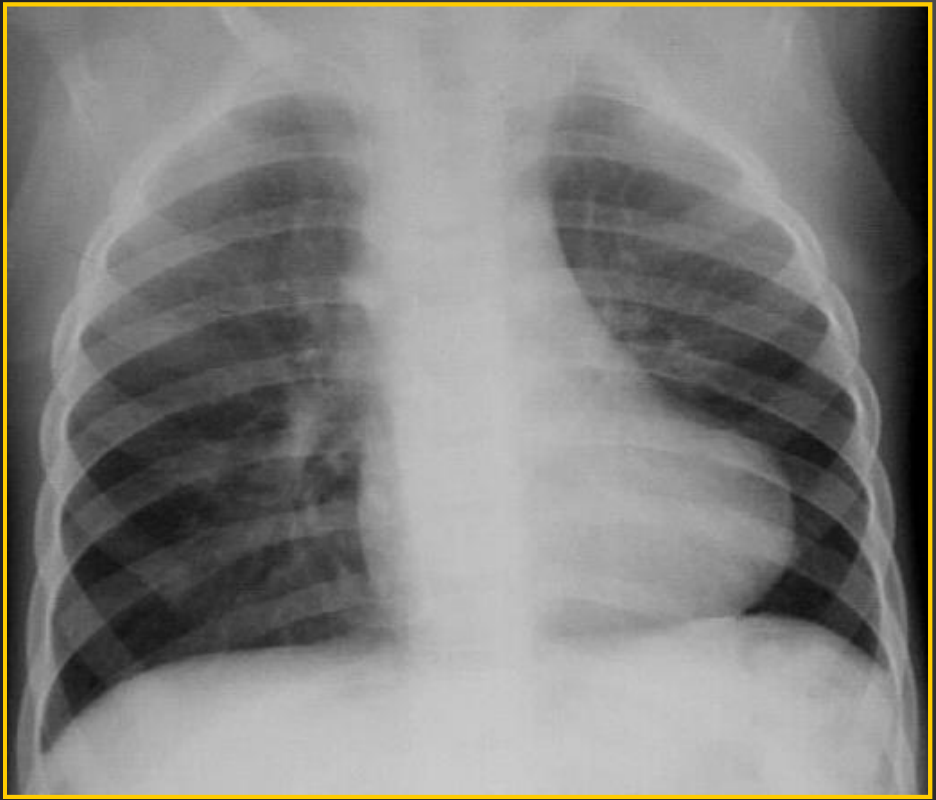


Unrepaired TOF (from ECHO)

Repaired TOF (from CMR)

May be useful in organizing interventions, PV implantation, construction of impantable materials that fit in the patient's RVOT

TOF – Chest X-ray



- **Boot-shaped heart** (not common)
- **↓ pulmonary markings in ↓ pulmonary blood flow**

CMR – Anatomy – Angio

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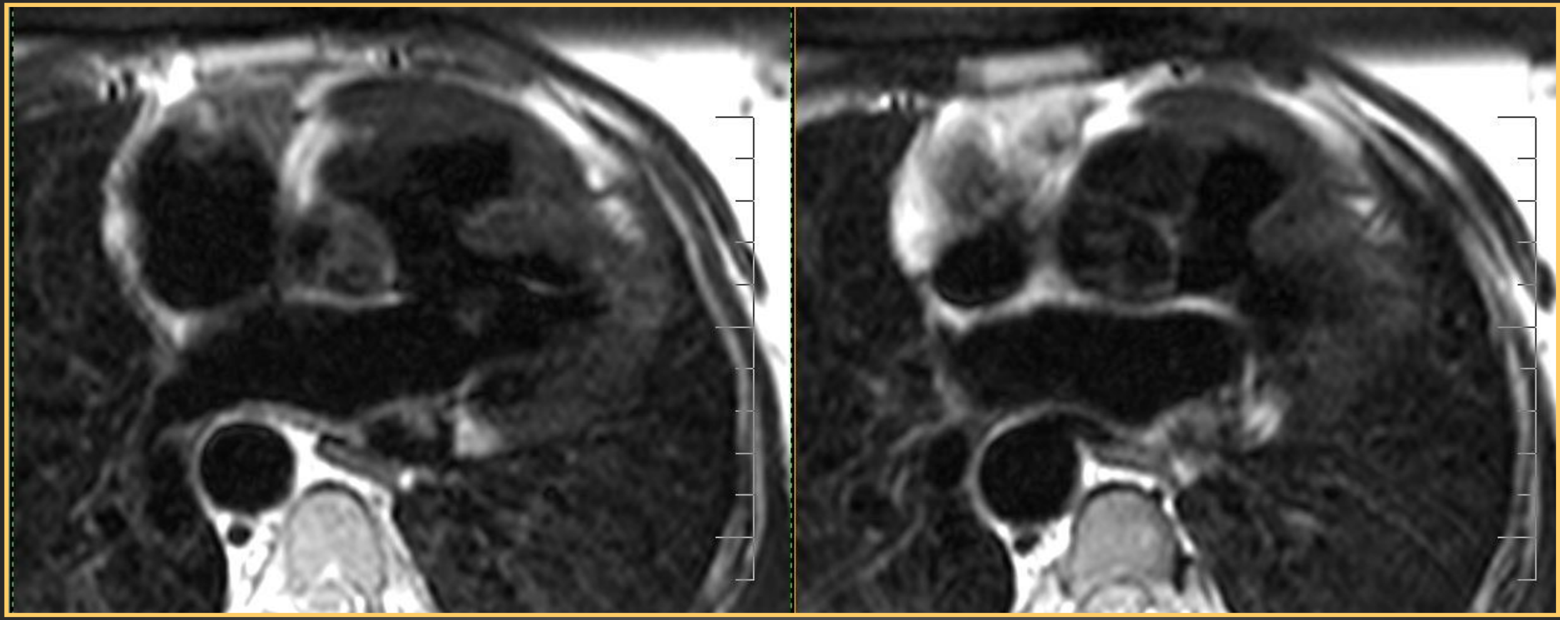

CMR – Angio – RVOT / PAs

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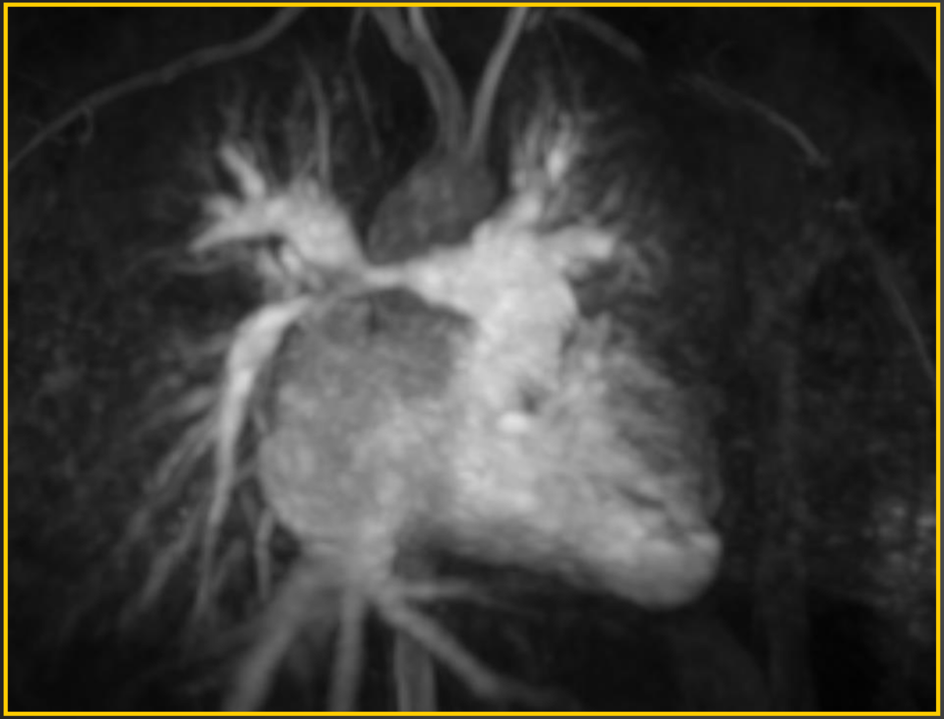
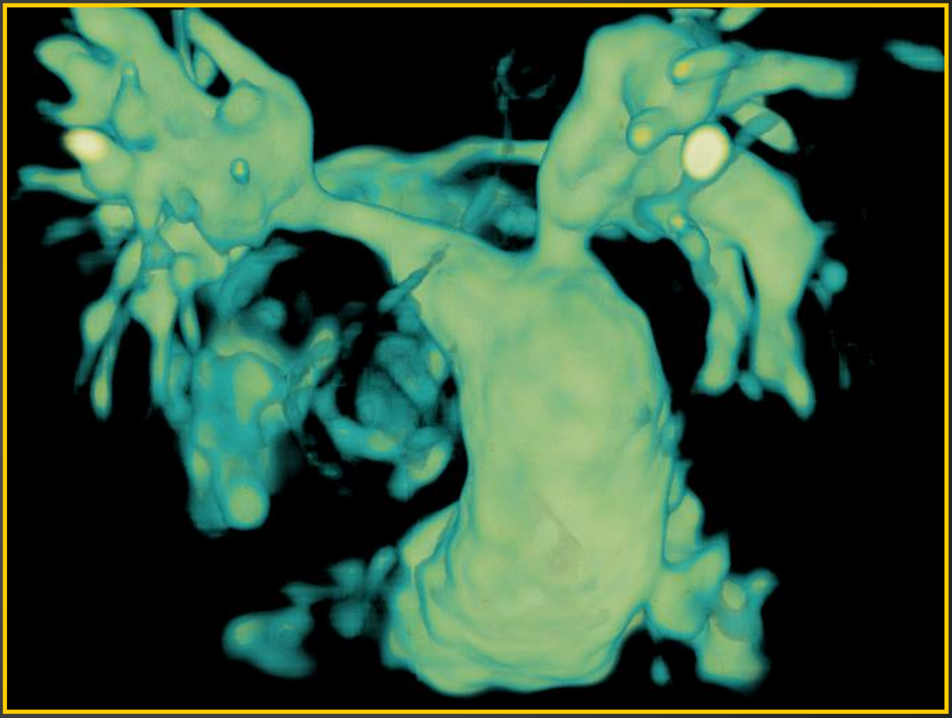
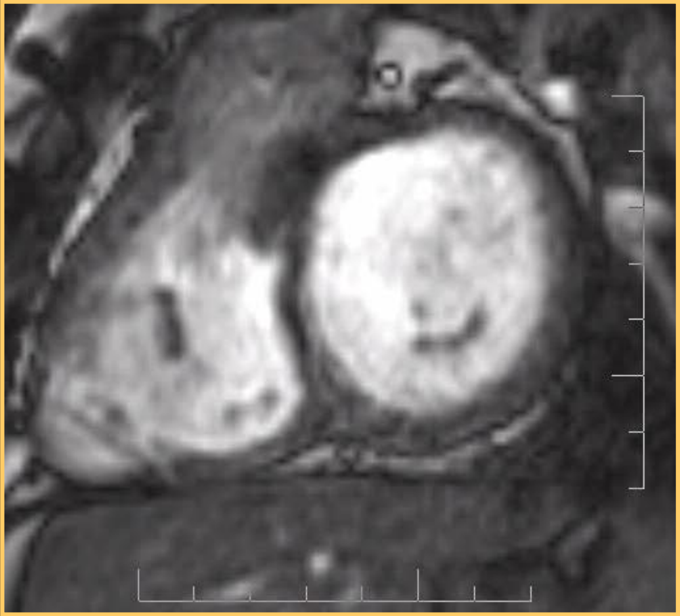
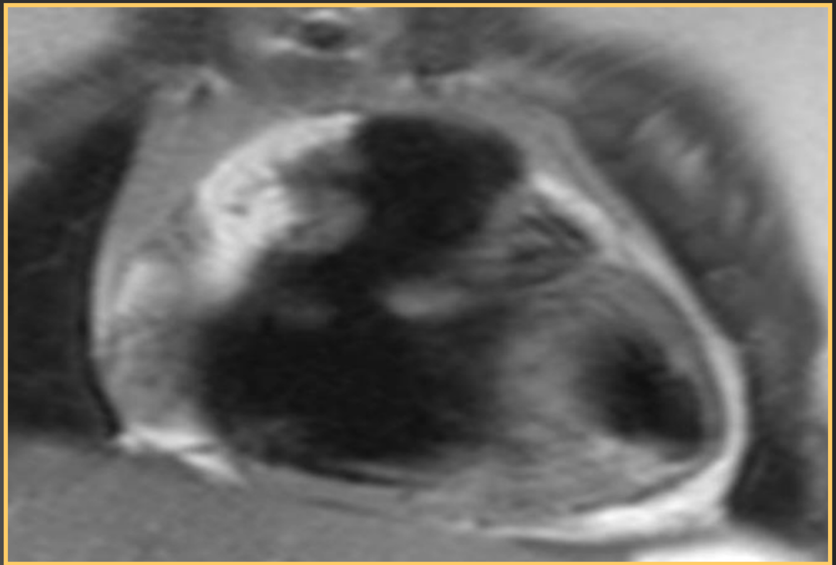
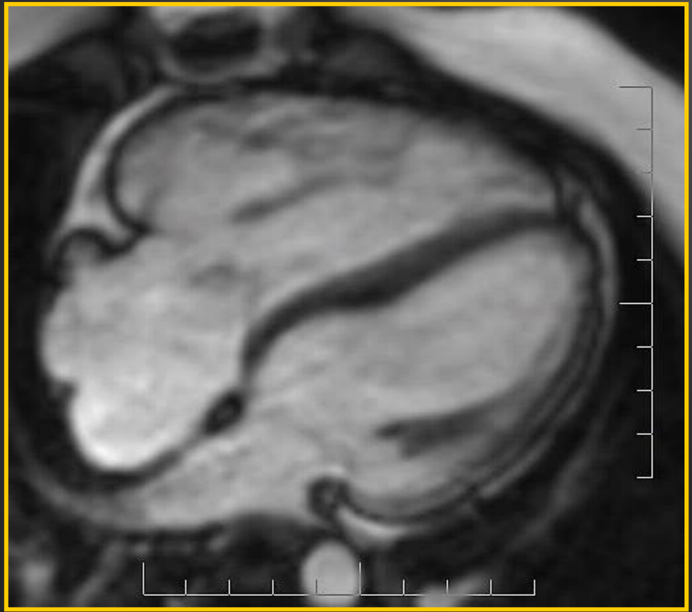


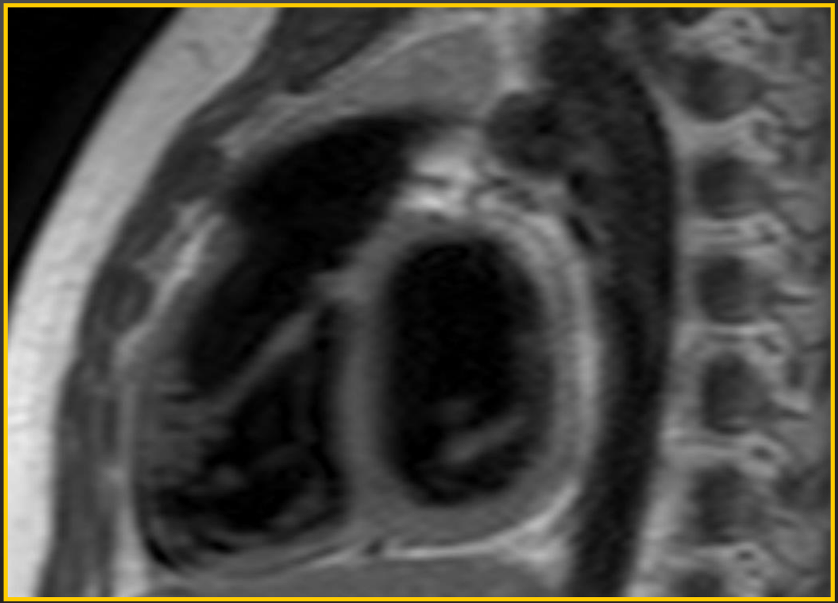


CMR – Anatomy – Cine – PS/PI

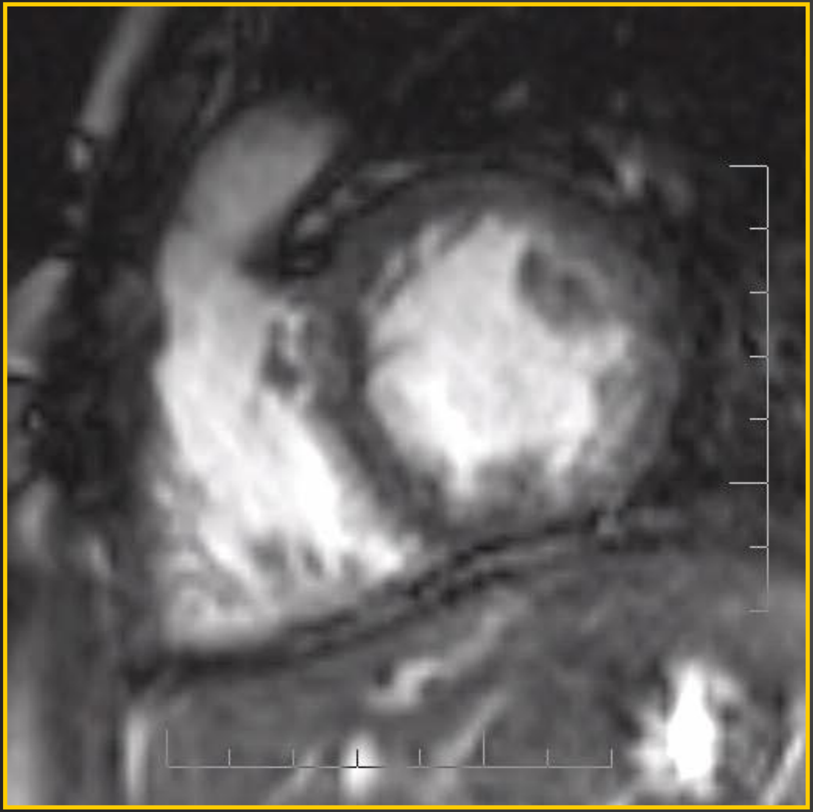


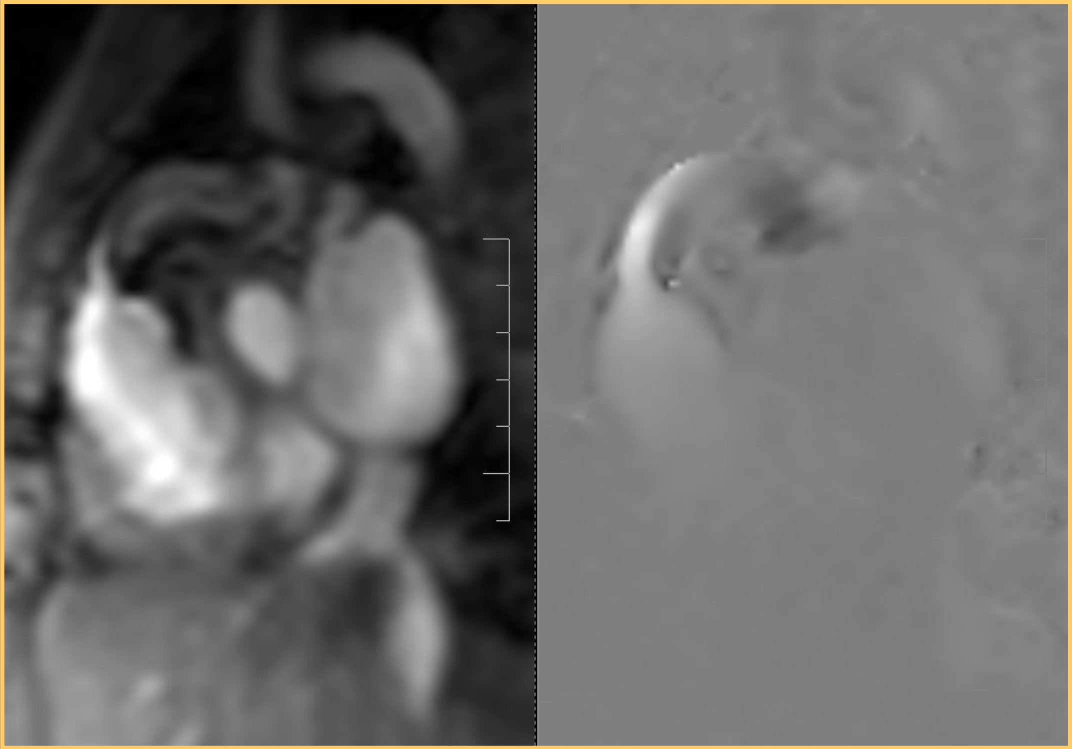






CMR – Cine – RVOT / Flows





CMR – Anatomy - Cine – Tagging

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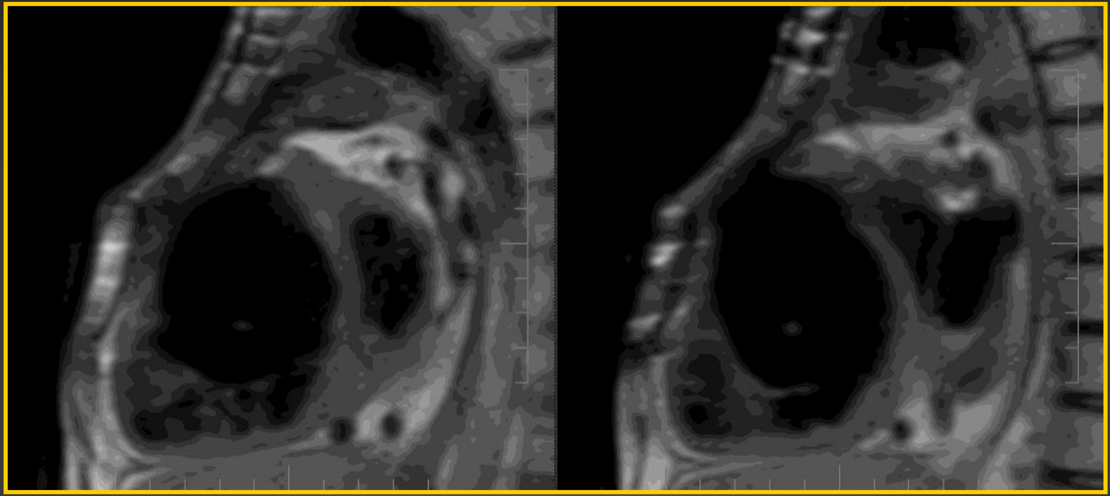

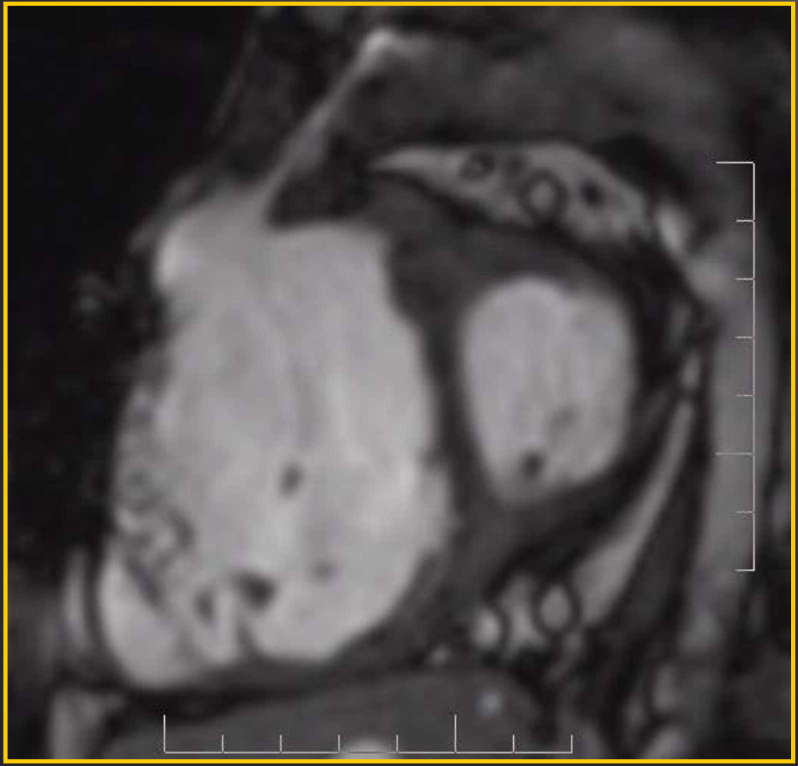

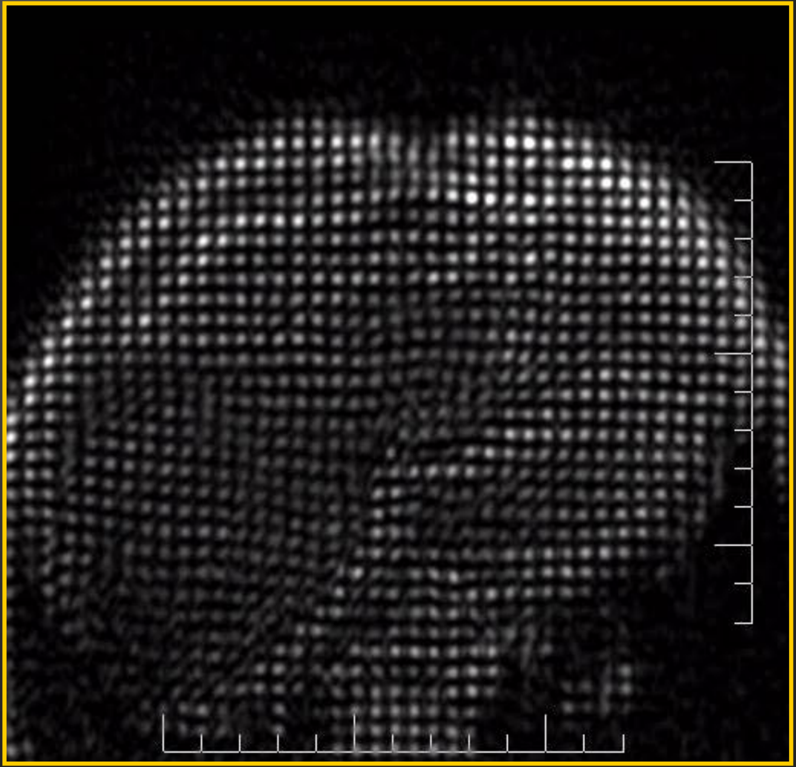

Unrepaired TOF – Angio

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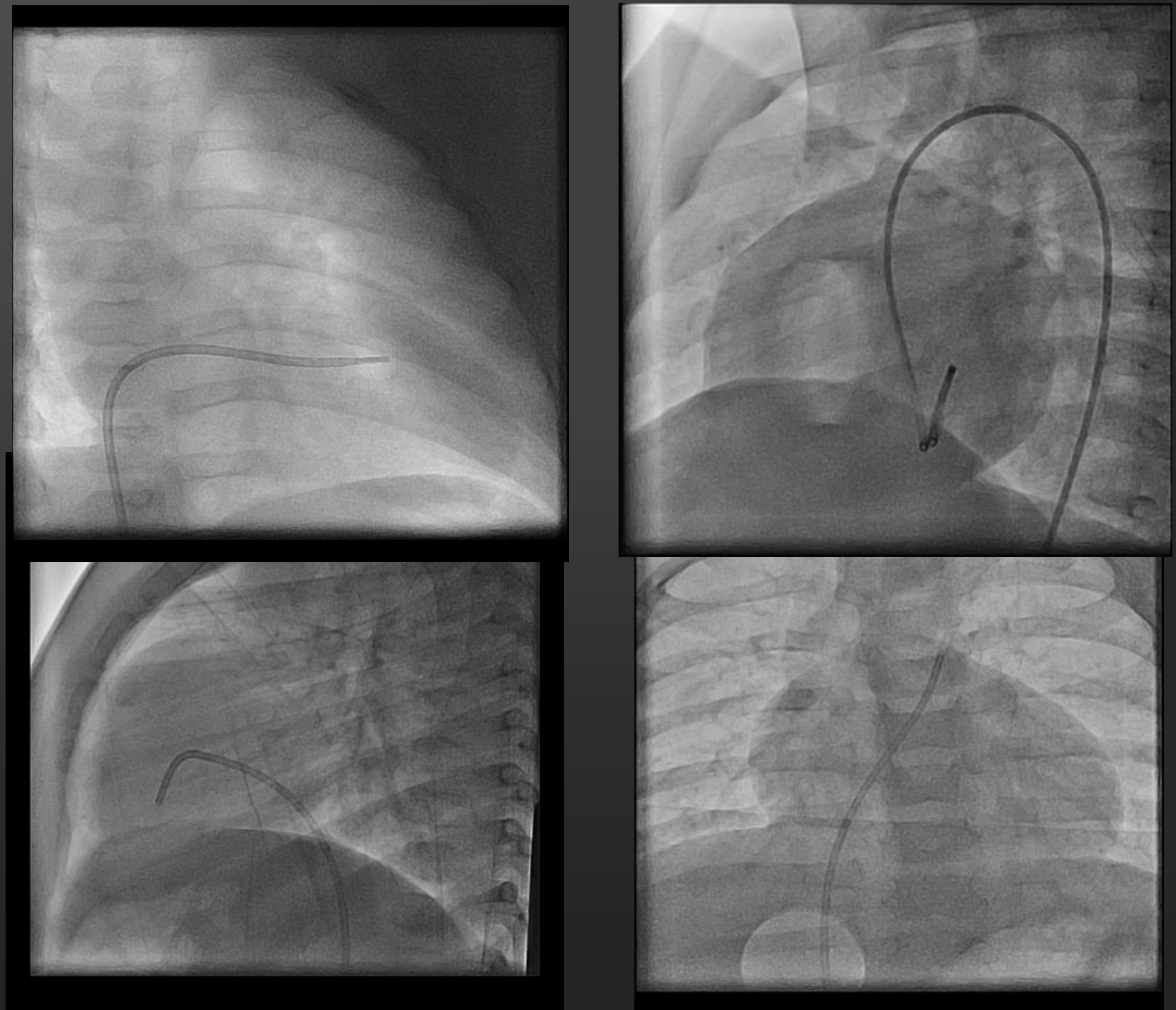

ECHO: 3D / TEE / Stress

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- 3D ECHO:
  - anatomy
  - RV & LV size/fxn (underestimate vs CMR)
- TEE:
  - PFO/ASD
  - TTE challenging
  - infective endocarditis
  - guide to interventional procedures
- Stress ECHO:

  * Stress Echo 2020 study with TOF branch (recruits since 2016)
  - assess RV contractile reserve
  - correlation with severity (NYHA, BNP, peak VO_2, 6MWD)
  - assess medium and long-term prognostic value of SE

Tetralogy of Fallot

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Usefulness of Dobutamine Stress Echocardiography with Tissue Doppler Imaging for the Evaluation and Follow-Up of Patients with Repaired Tetralogy of Fallot

21 patients (median age 27.7 (19-48) years, Dobutamine

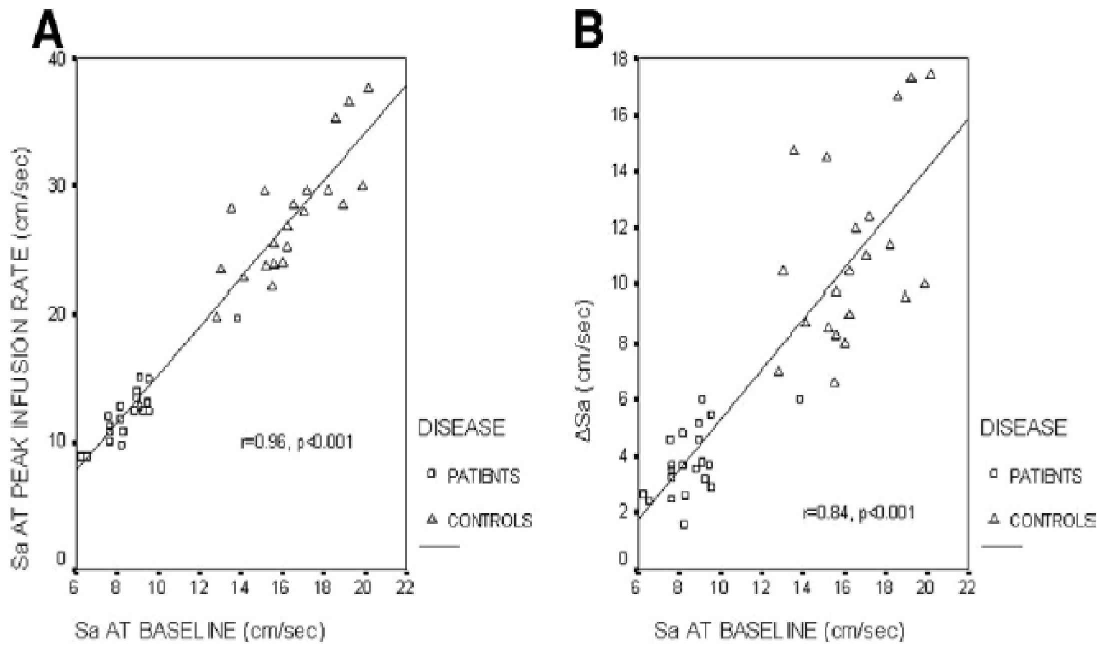

Figure 2 (**A**), Correlation between the baseline values of TDI velocity of the tricuspid annulus during systole (Sa) and the values of Sa at peak infusion rate. (**B**), Correlation between the baseline values of TDI velocity of the tricuspid annulus during systole (Sa) and the absolute dobutamine-induced increase of Sa (ΔSa).

Brili et al. J Am Soc Echocardiogr 2008;21:1093-1098

TGA s/p Mustard/Senning - Dobut

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Reduced contractile reserve of the systemic right ventricle under Dobutamine stress is associated with increased brain natriuretic peptide levels in patients with complete transposition after atrial repair

	Rest	Dobutamine stress	*P*-value
IVA (m/s^2)	0.7 ± 0.25 (0.4–1.1)	1.5 ± 0.9 (0.6–1.2)	< 0.001
s-Velocity (cm/s)	1.8 ± 0.9	3.3 ± 1.6	< 0.002
e-Velocity(cm/s)	4.3 ± 2.2	3.3. ± 1.6	NS
Strain	21.5 ± 4.3	27.5 ± 6.7	< 0.008

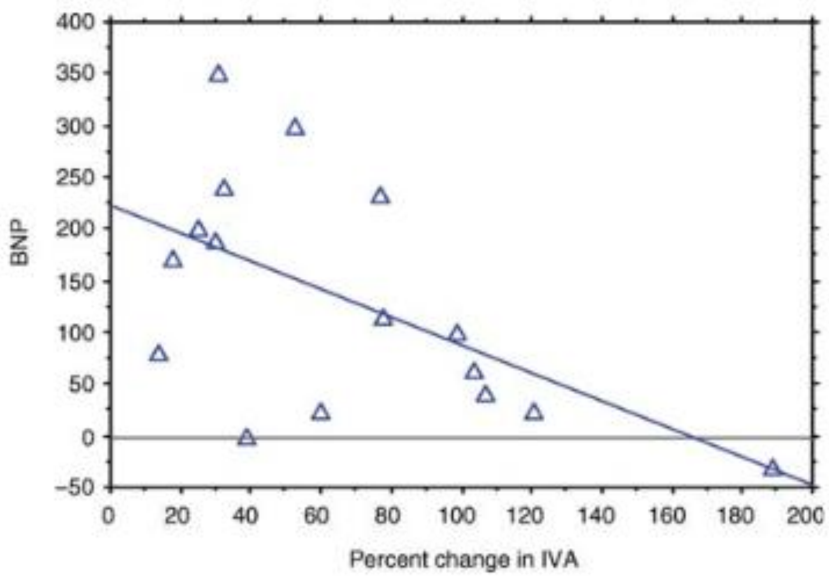

Figure 1 Correlation between change in IVA (*x*-axis) in percent and serum concentration of BNP (brain natriuretic peptide) in pg/mL (*y*-axis) in 16 patients with complete transposition following atrial repair ($r = -0.57$, $P > 0.02$).

- 16 patients (25.6±3.7yo)
- Dobutamine
- At rest, all patients had reduced:
  - IVA
  - s-velocities
  - e-velocities

compared with normal subjects with systemic LV

Vogt et al. European Journal of Echocardiography (2009) 10, 691–694

Pulmonary Hypertension – Master 2 step

ONASSIS CARDIAC SURGERY CENTER

Simple stress echocardiography unmasks early pulmonary vascular disease in adult congenital heart disease

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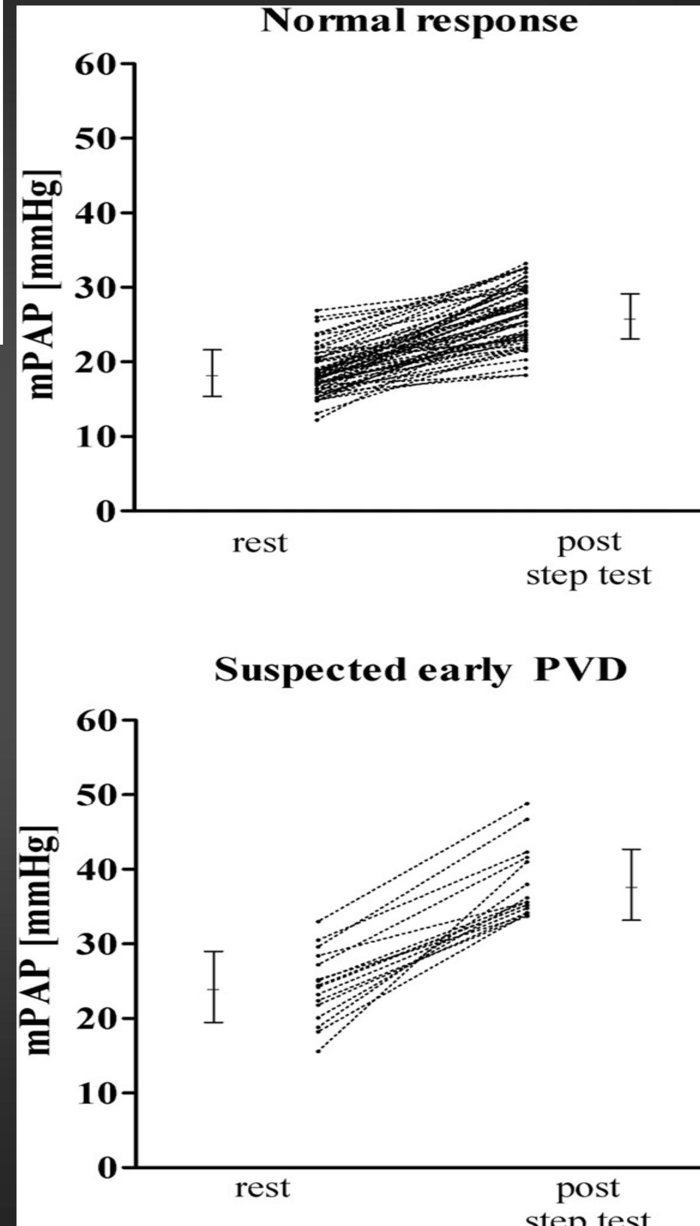
Type of defect				
ASD		35 (58)	10 (63)	
VSD	76	20 (33)	2 (12)	0.10
PDA		1 (2)	2 (12)	
Other		4 (7)	2 (12)	

- 78 patients (43.2 ± 14.5 yo)
- Symptom-limited Master two-step test
- Systolic PAP: TR CW + RAP (from IVC size and collapsibility)
- Mean PAP = 0.6× systolic PAP + 2
- Suspected early PVD: - mean PAP>34 mmHg
  16 patients (21%) - CO>10 L/min
  - mean PAP/CO > 3



Van Riel et al. IJC 197 (2015) 312–314

Pulmonary Hypertension - Bike

Right ventricular load and function during exercise in patients with open and closed atrial septal defect type secundum

- 20 open ASDs
  39.3±17.5 yo
- 30 closed ASDs
  42.4±16.8 yo

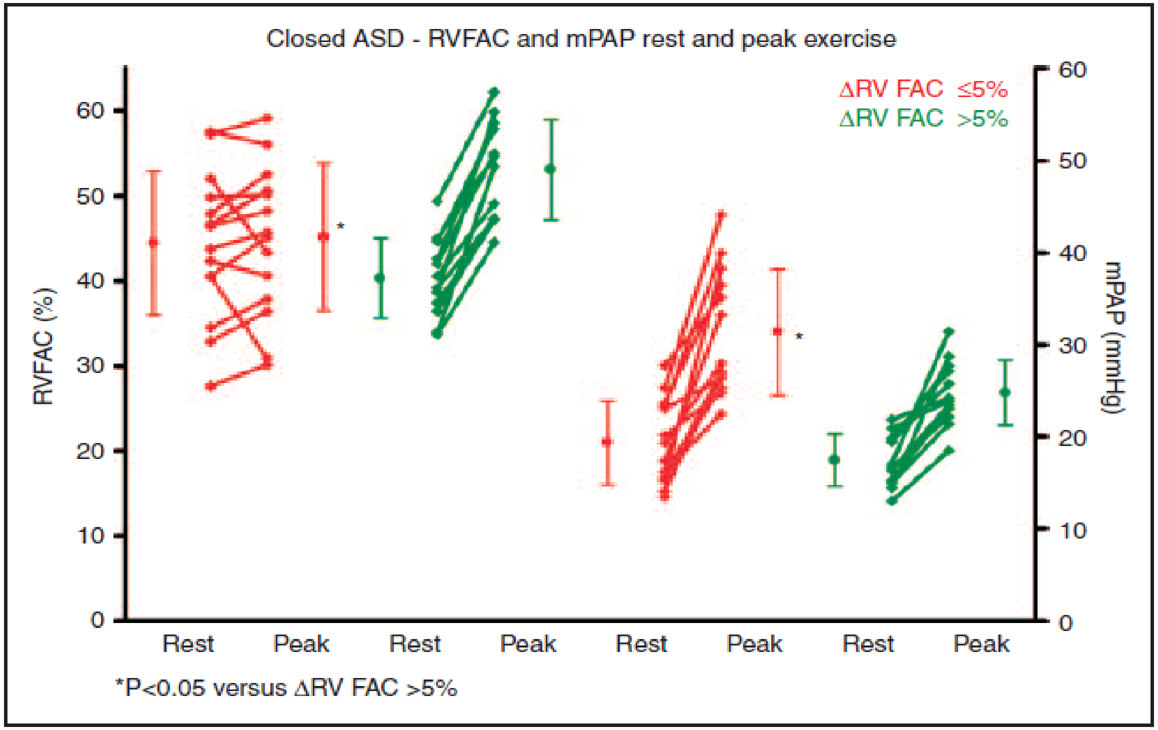


Figure 4. If right ventricular RVFAC increases ≤5% from rest to peak exercise, open and closed atrial septal defect (ASD) patients present with a significantly higher mean pulmonary artery pressure (mPAP) at peak exercise.

Van De Bruaene et al. Eur J Prev Cardiol. 2013 Aug;20(4):597-604

Conclusions

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- Indications for Stress ECHO in pediatric and CHD are evolving
- Serial testing may provide help in
  - diagnosis
  - risk stratification
  - follow-up
  - evaluation of treatment
- The impact of SE on patient outcomes is under research
- Efficiency may be increased with comprehensive TTE
- Deformation and multidimensional imaging may ↑ utility of SE
- SE has great versatility and its use is likely to expand