Heart failure in Fallot survivors

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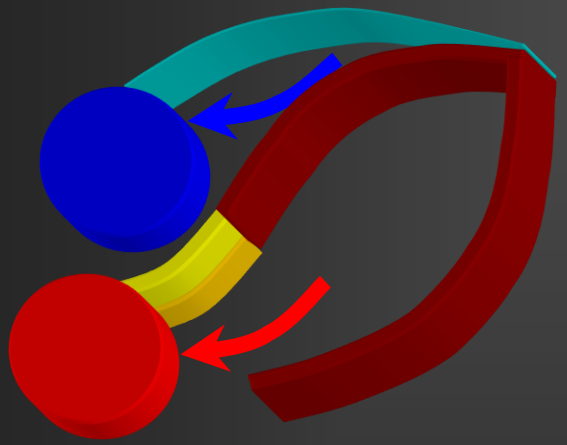
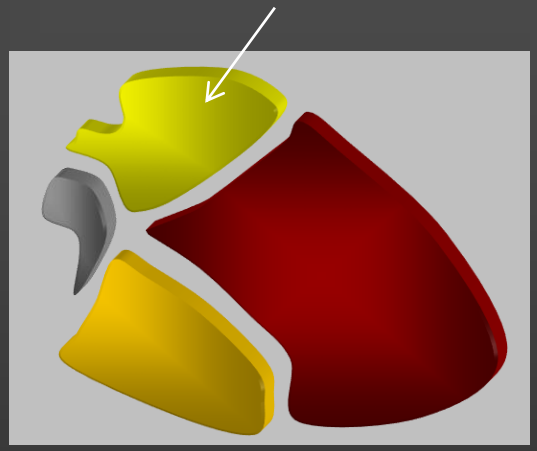
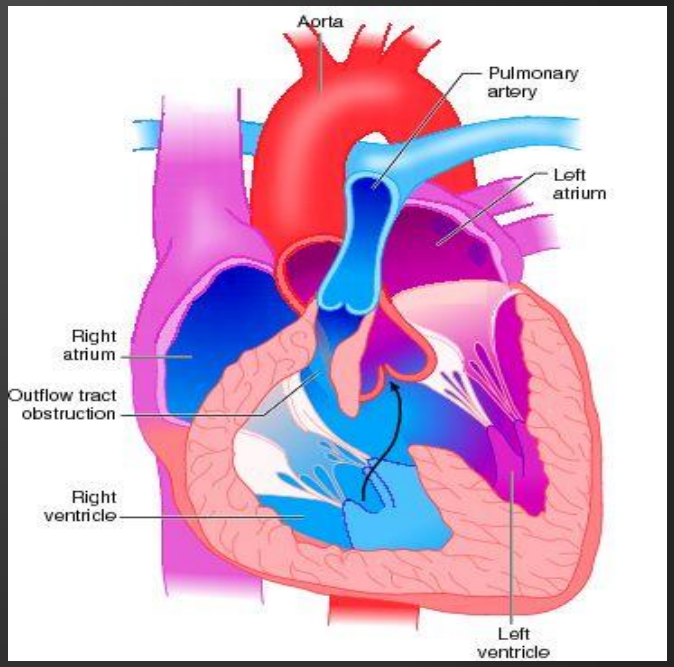
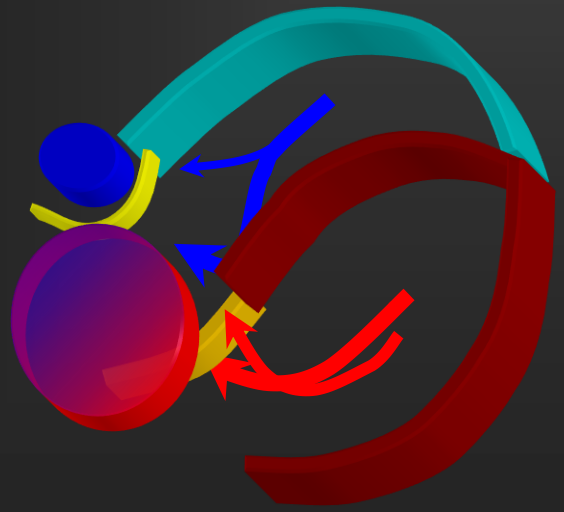
Tetralogy of Fallot

= anterior malalignment of the outlet septum









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

Heart Failure (HF) in TOF

- TOF is the most common cyanotic CHD treated since the 1940s
- There are a large number of adults with TOF
- Surgery in TOF results in pulmonary regurgitation (PI) that leads to RV enlargement and dysfunction and predisposes to:
  - HF
  - Arrhythmias
  - Sudden cardiac death (SCD)
- PI can be ameliorated with pulmonary valve replacement (PVR) that:
  - decreases RV size and PI
  - has not yet been correlated with improved clinical outcomes

Pathophysiology of HF in CHD

Table 1 **Pathophysiology of heart failure with impaired systolic function: triggers (examples)**

1. Systolic dysfunction of the systemic morphological left ventricle
   - Pressure overload (sub-, supravalvular or valvular aortic stenosis, coarctation of the aorta)
   - Volume overload (aortic valve regurgitation, VSD, patent ductus arteriosus, or mitral regurgitation)
   - Myocardial injury (limited myocardial protection during bypass, ventriculotomy)
   - Altered myocardial architecture (non-compaction)
   - Altered geometry of the sub-pulmonary ventricle interfering with diastolic filling of the systemic ventricle (severe pulmonary regurgitation in ToF)
2. Systolic dysfunction of the sub-pulmonary morphological right ventricle
   - Volume overload (severe pulmonary regurgitation in ToF, atrial septal defect with large left-to-right shunt)
   - Pressure overload (severe RV outflow tract obstruction)
3. Systolic dysfunction of the morphological systemic right ventricle
   - Pressure overload [congenitally corrected transposition of the great arteries, dextro-transposition of the great arteries after atrial switch repair (Mustard or Senning)]
   - Myocardial injury by functional ischaemia (single right coronary artery)
4. Systolic dysfunction of the systemic single ventricle
   - Volume under-load after initial volume overload (Fontan repair)
   - Myocardial injury (limited myocardial protection during bypass, ventriculotomy)
   - Myocardial architecture
5. Systolic dysfunction of the cyanotic systemic and/or sub-pulmonary ventricle with or without pulmonary hypertension
   - Myocardial injury by chronic hypoxia (VSD with pulmonary stenosis)
   - Pressure overload (Eisenmenger syndrome)
6. Acquired ischaemic heart disease and ventricular dysfunction
   - Cardiovascular risk factors (hypertension, hyperlipidaemia, diabetes mellitus, smoking)
   - Congenital coronary artery abnormalities (anomalous origin and/or course, extrinsic compression by a dilated pulmonary artery, coronary kinking after re-implantation of coronary arteries)
7. Systolic dysfunction of the systemic ventricle due to tachyarrhythmias

European Heart Journal (2016) 37, 1419–1427

Pathophysiology of HF in repaired TOF

- RV Systolic dysfunction

  - ↑ RV volume and RV mass → ↓ RVEF with preserved CO and wall stress

  - RV fibrosis and altered geometry, abnormal electromechanical coupling, and abnormal perfusion → RV dysfunction

  - RV dilation causes left septal deviation and ↓ RV and LV function

- RV Diastolic dysfunction

  - RV stiffness → ↓ EDV → RV acts just as a conduit between RA and PA

- LV dysfunction due to Ventricular Interdependence

  - mechanical, electric, and neurohormonal coupling of LV and RV

  - RV and LV share myofibers

  - Altered RV size and geometry → pericardial constraint affecting LV

  - Diastolic shift of the ventricular septum → ↓ LV diastolic expansion

  - Poor RV output → low LV preload

- Myocardial fibrosis and scarring from ↑ RV volume and previous surgery as well as neurohormonal activation → malignant VT and SCD

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Symptoms – Signs of HF

Table 2 Signs and symptoms of heart failure in congenital heart disease

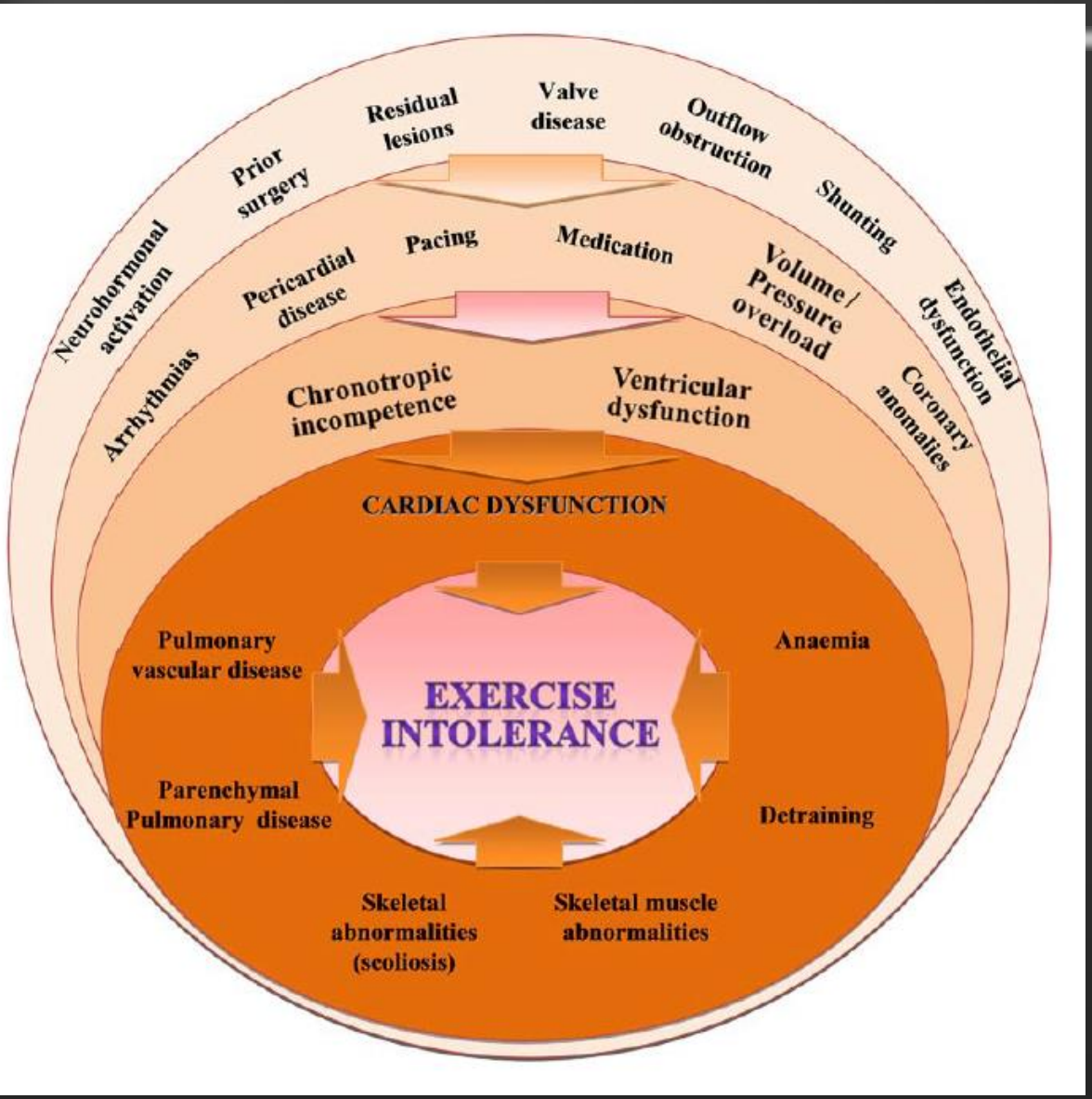
Symptoms of systemic ventricular failure	Signs of systemic ventricular failure
Fatigue	Third or fourth heart sound (gallop)
Breathlessness	Laterally displaced apical impulse
Dry cough especially lying flat	Pulmonary crepitations
Reduced exercise tolerance	Absent BS and dull percussion lung bases due to pleural effusions
Orthopnoea	
Paroxysmal nocturnal dyspnea	
Wheezing	
Symptoms of sub-pulmonary ventricular failure	**Signs of sub-pulmonary ventricular failure**
Fatigue	Elevated JVP
Bloating	Hepatomegaly
Weight gain (> 2kg/week)	Ascites
Loss of appetite	Pitting leg oedema, sacral oedema, scrotal oedema
Reduced exercise tolerance	
Increased abdominal girth	
Symptoms of congestive (biventricular) failure	**Signs of congestive (biventricular) failure**
Combined systemic and sub-pulmonary symptoms	Combined systemic and sub-pulmonary signs

European Heart Journal (2016) 37, 1419–1427

HF Diagnosis – Stress test - BNP

- Cardiopulmonary exercise test (CPET)
  maximal O2 consumption (mVO_2) and F/U
- In complex CHD mVO_2 depends on *cyanosis*, *rhythm disturbances*, *abnormal physiology* etc and is *difficult to assess*
- BNP: - in acute HF the levels ~ with severity
  - BNP > 300 pg/mL → bad prognosis

Exercise tolerance



Decreased exercise tolerance may be due to myocardial dysfunction but also to other factors such as physiology, cyanosis, previous operations, rhythm disturbances, systemic effects of HF

Circulation. 2016;133:770-801

Imaging modalities

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

Repaired TOF: RV - PI - TR

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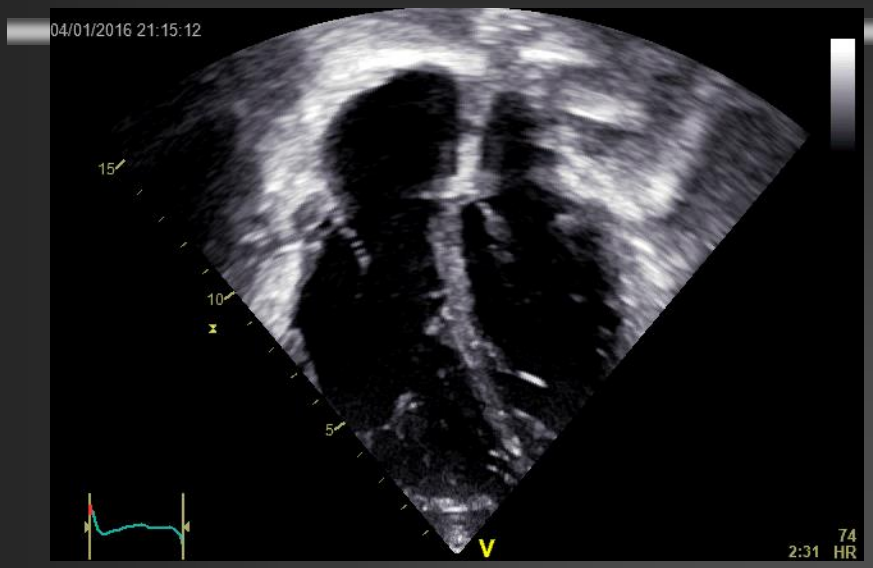

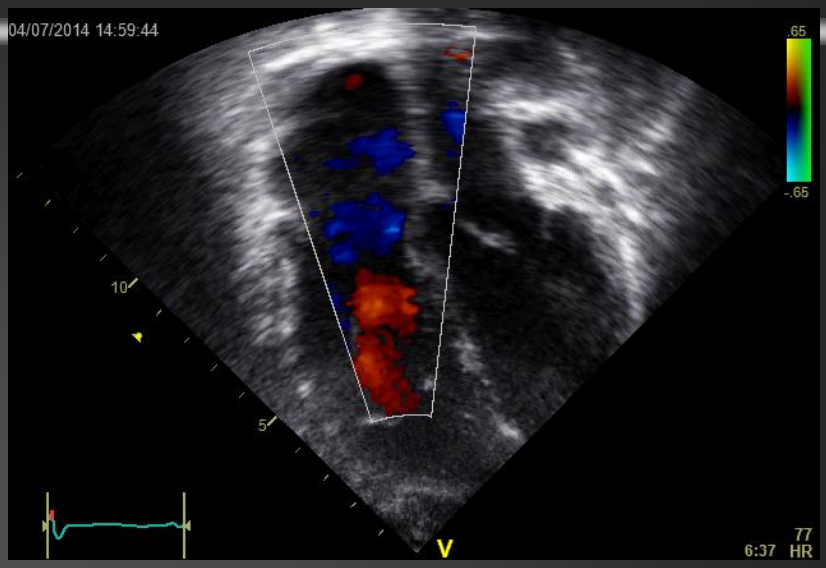

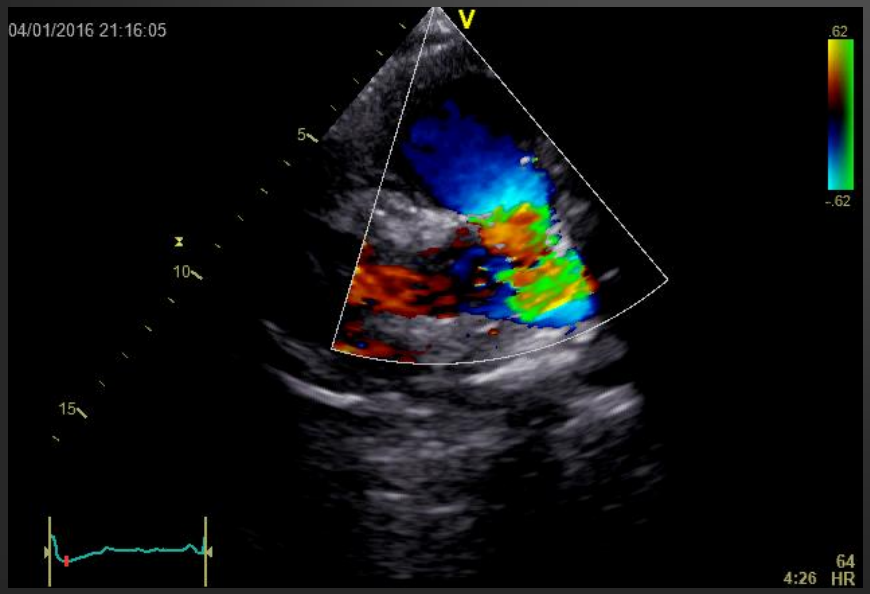

Table 3 Echocardiographic reference values of RA and ventricular size and function in healthy adults (adapted from Rudski *et al.*[23])

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

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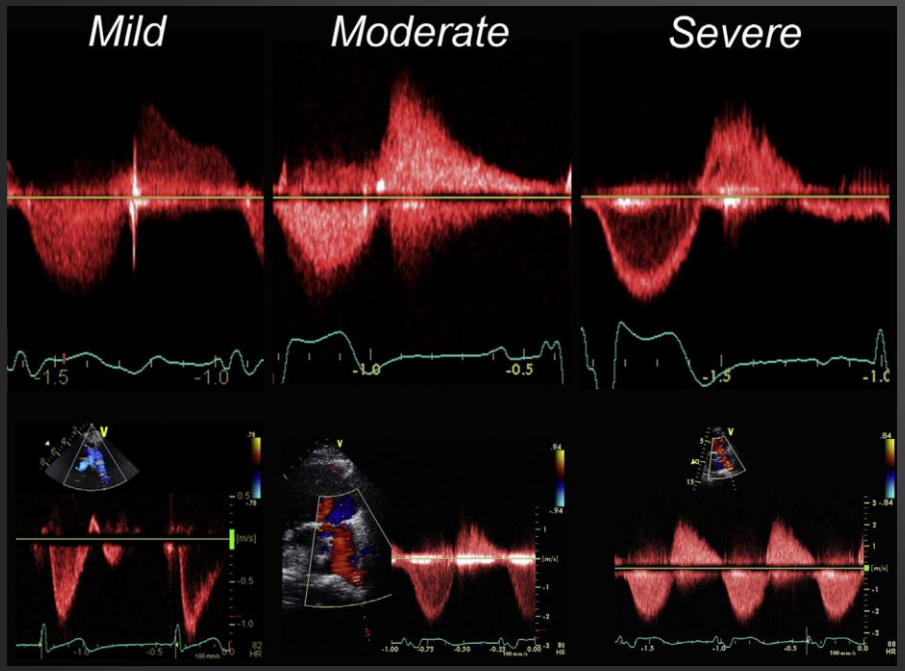
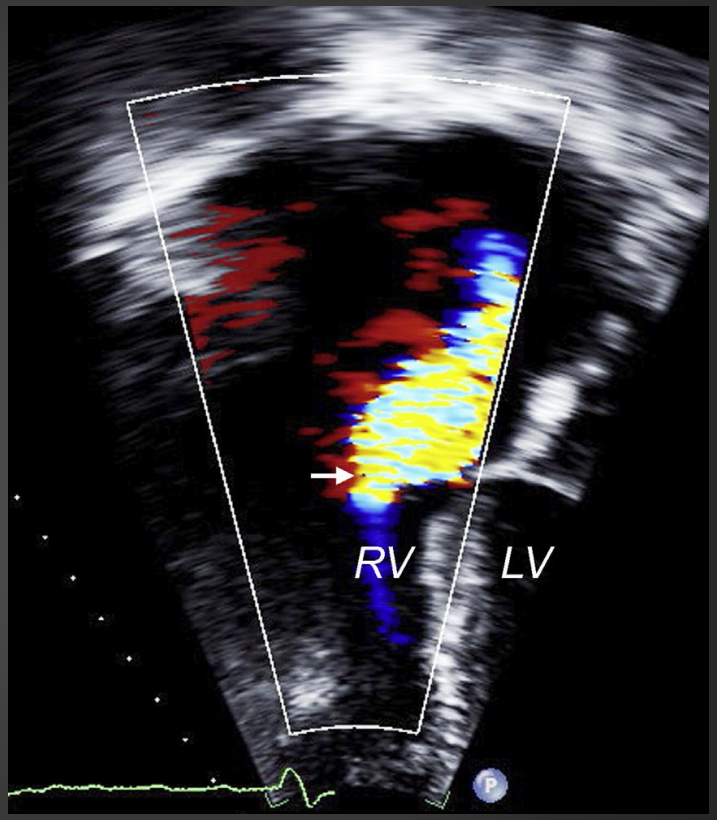
Mild PI: persistent flow gradient at end-diastole

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

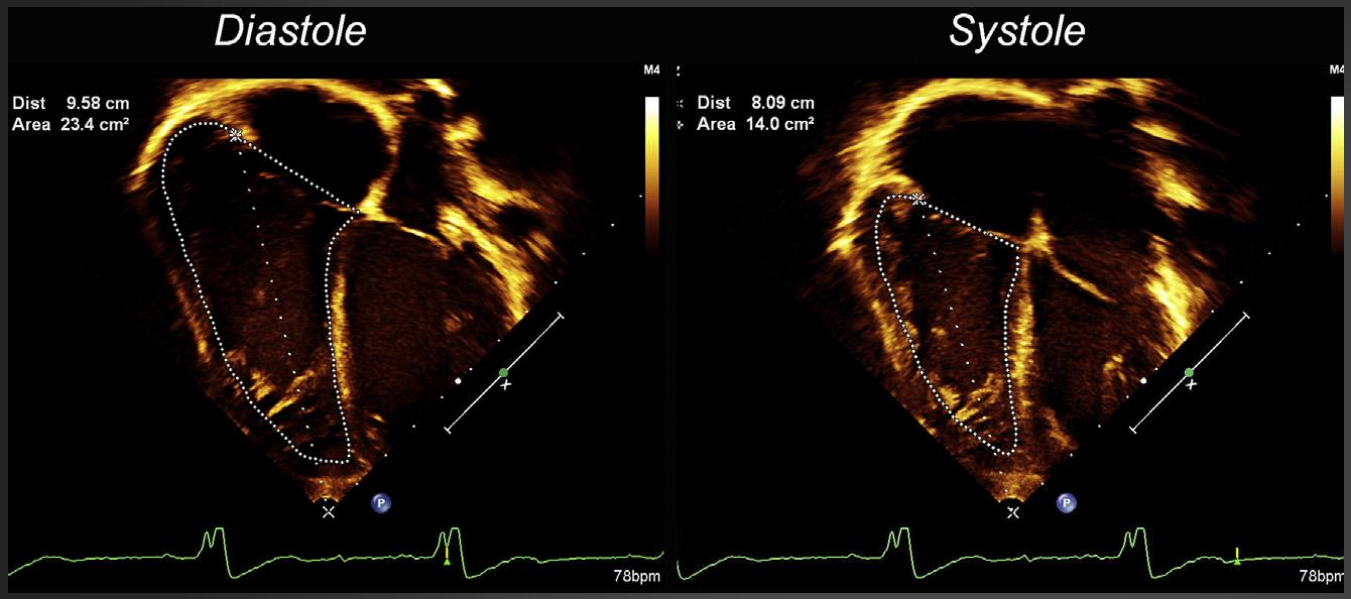


TR morphology

RVp estimate = $4v^2$ + RAP

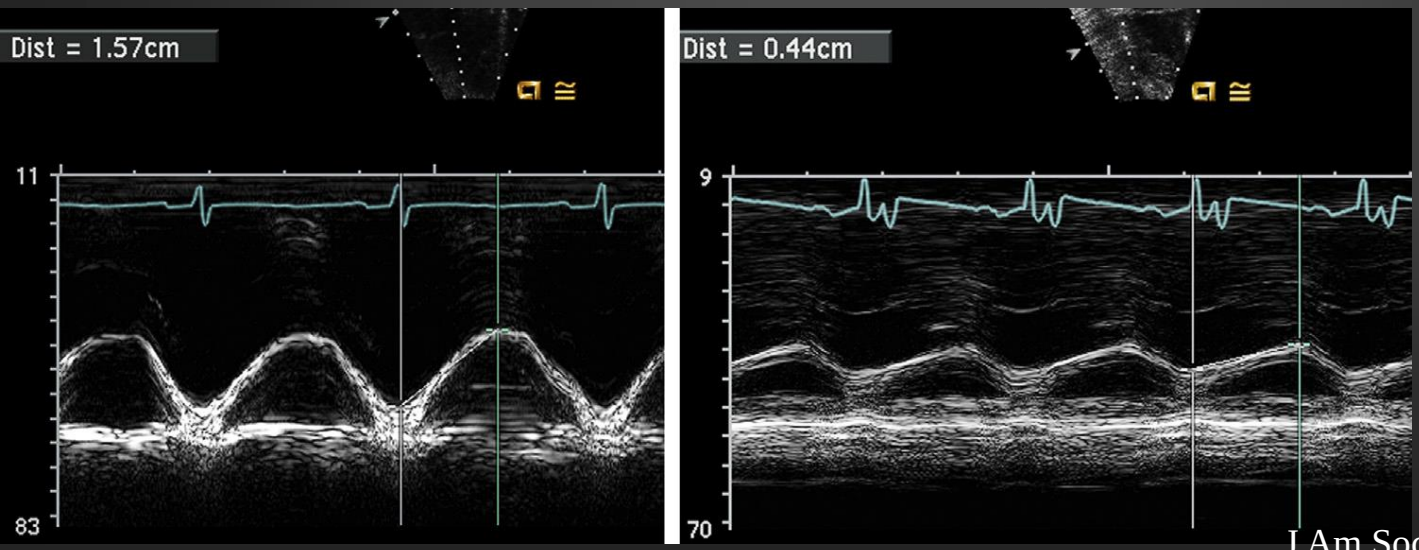
J Am Soc Echocardiogr 2014;27:111-41

Repaired TOF: RV



RV fractional area change (%)

(modest correlation with CMR in TOF)

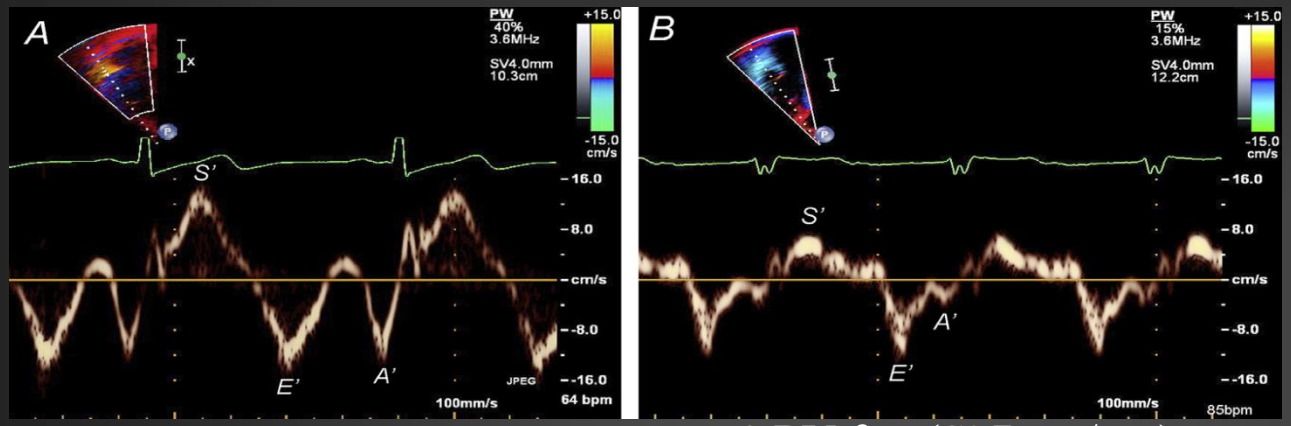


TAPSE

(weak correlation with CMR in TOF)

J Am Soc Echocardiogr 2014;27:111-41

Repaired TOF: Newer ECHO techniques



Nl RV fxn (S′ 14 cm/sec)

↓ RV fxn (S′ 7 cm/sec)

TDI

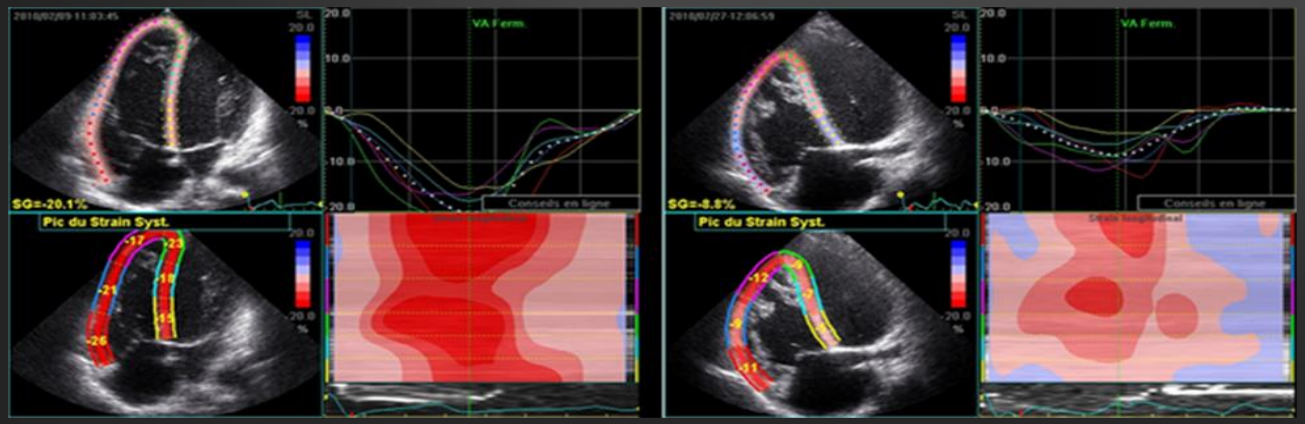
(no available guidelines in TOF)

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 TOF)

Echocardiography. 2014;31(4)

TOF 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:
  - assess RV contractile reserve
  - correlation with severity (NYHA, BNP, peak VO_2, 6MWD)
  - assess medium and long-term prognostic value of SE

TOF CMR

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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 – Angio – RVOT / PAs / Collaterals

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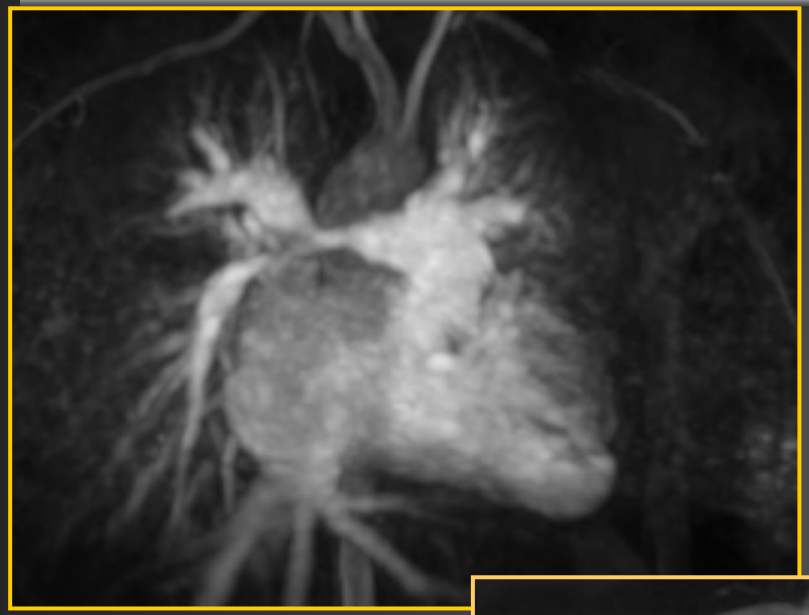

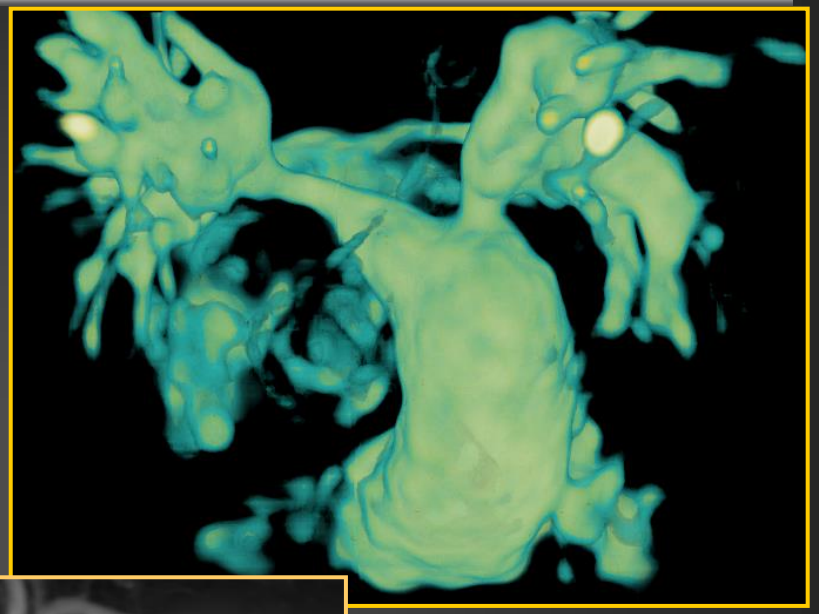

CMR – Anatomy – Cine – PS/PI

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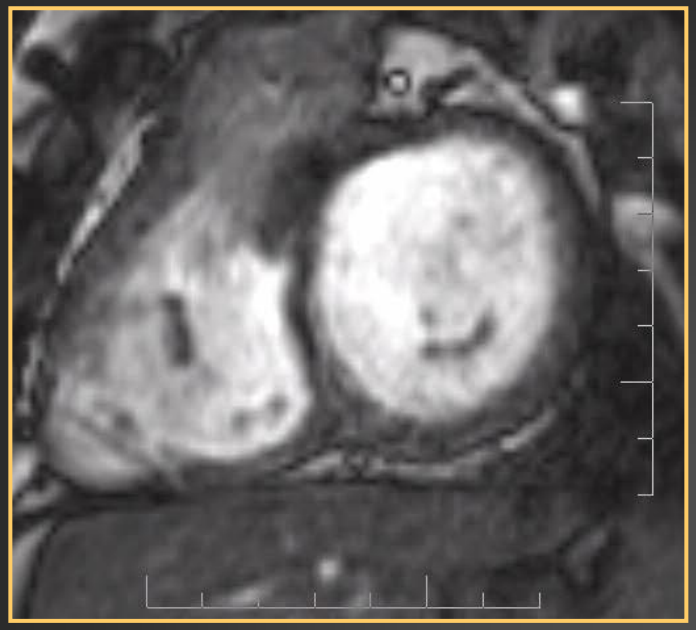

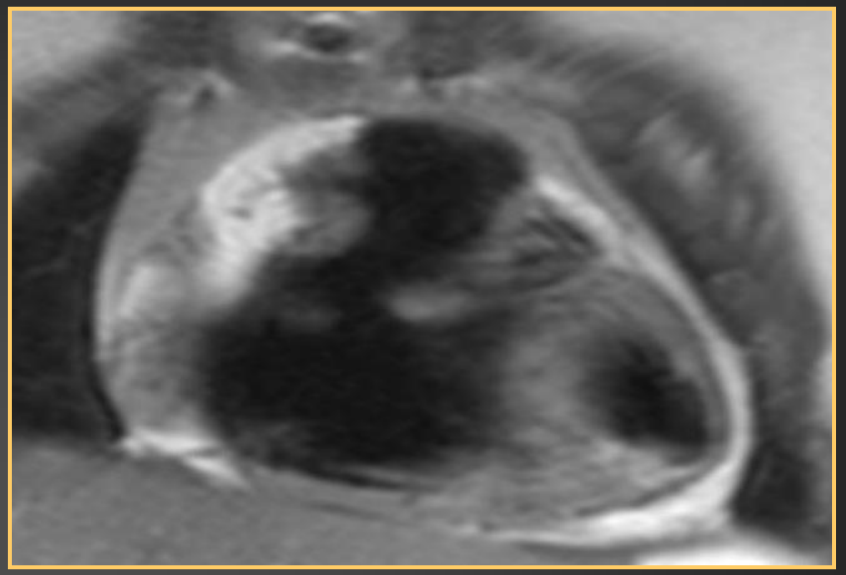

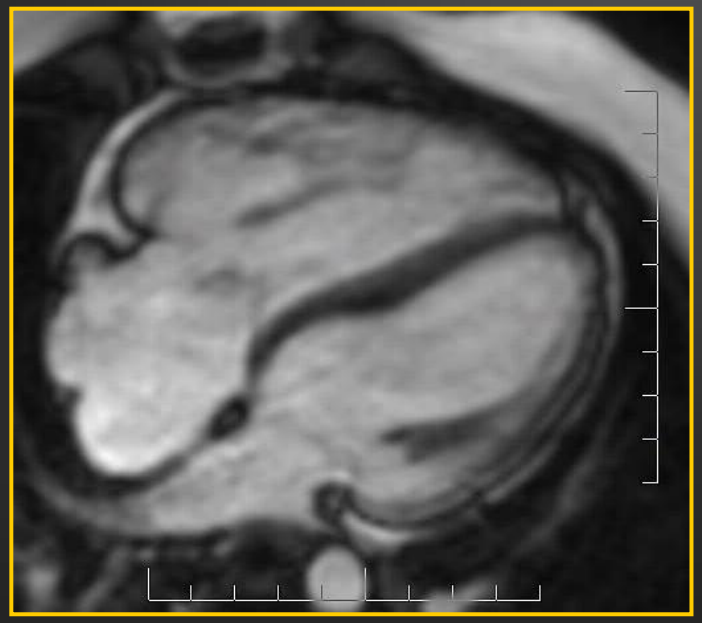

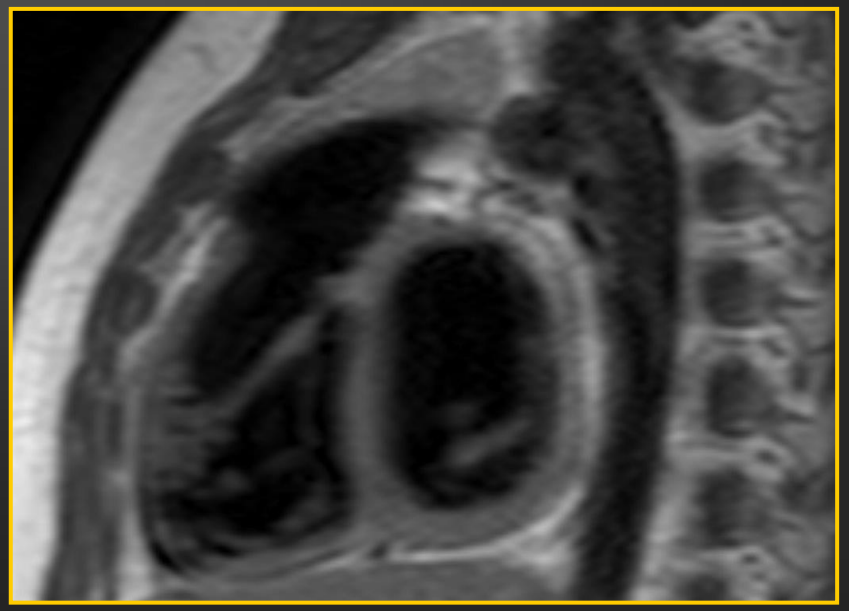

CMR – Cine – RVOT / Flows

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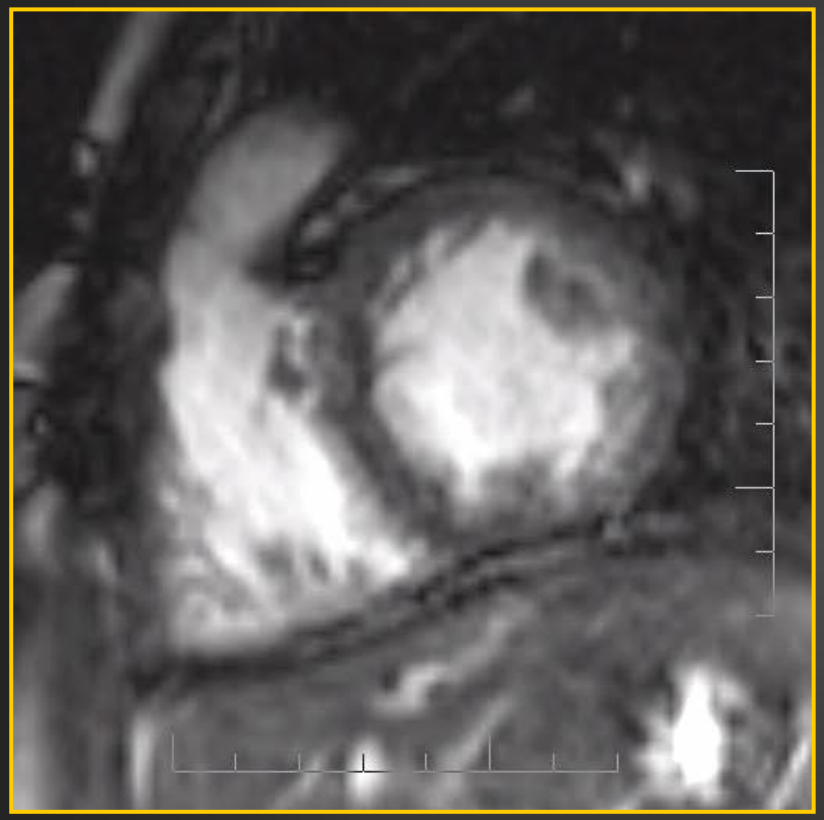

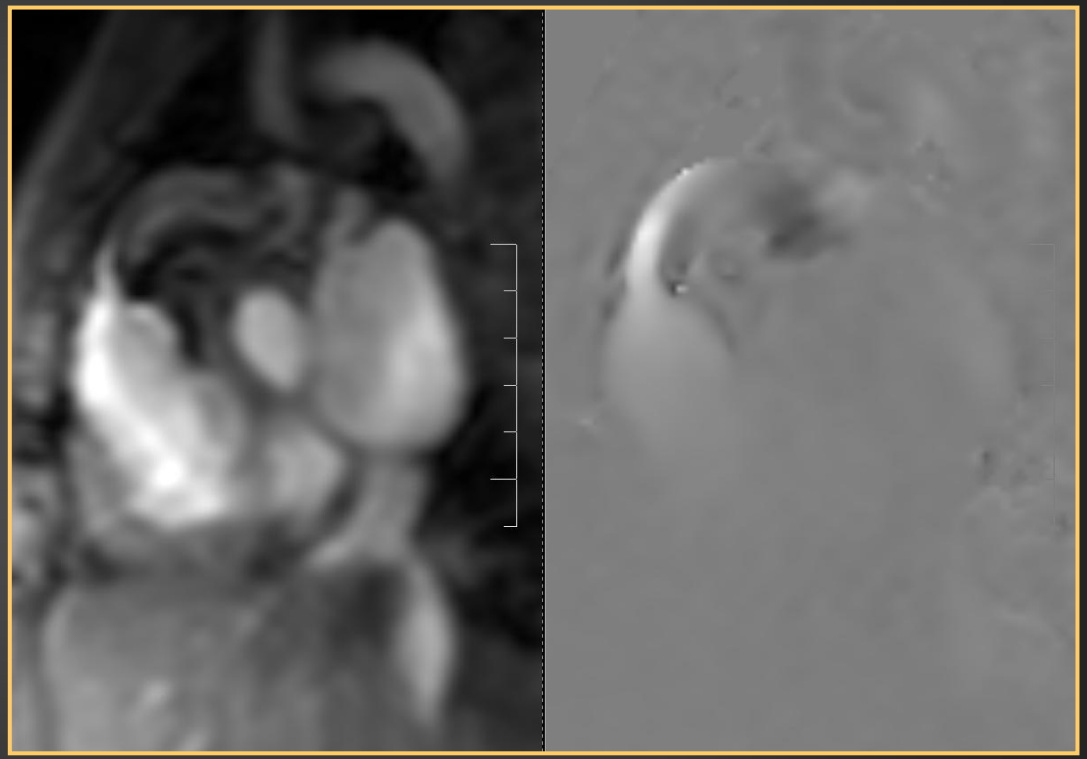

CMR – Anatomy - Cine – Tagging

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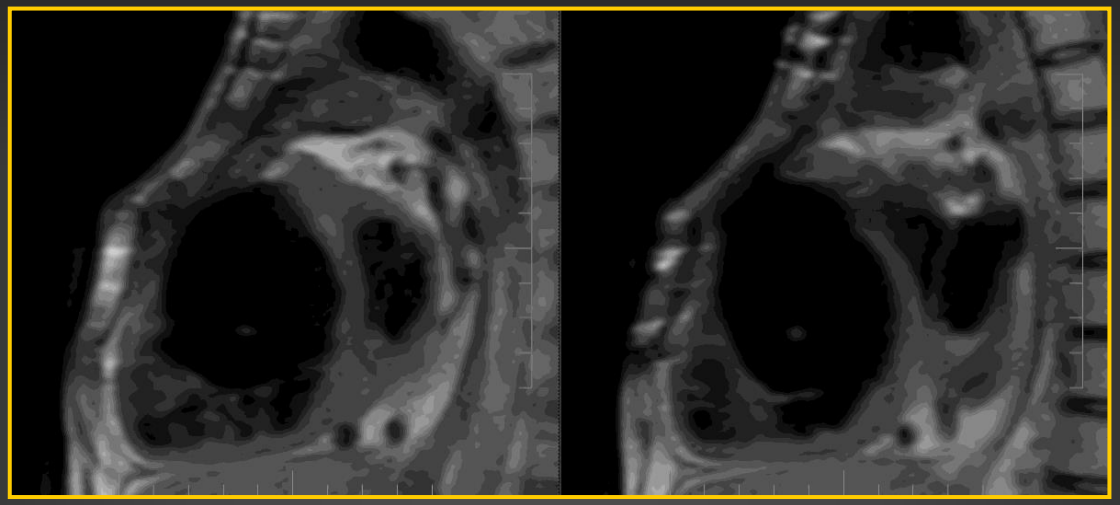

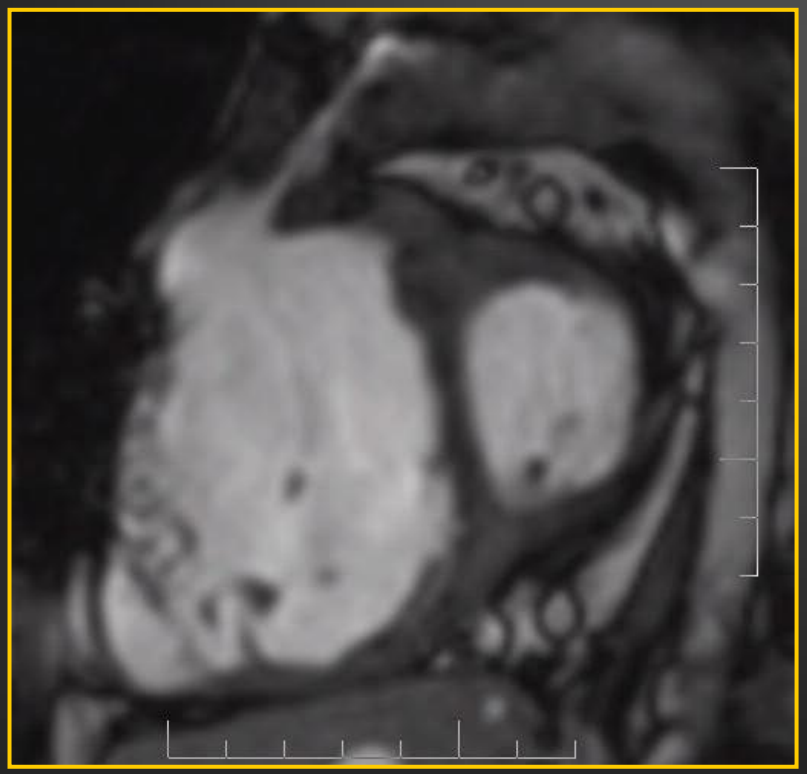

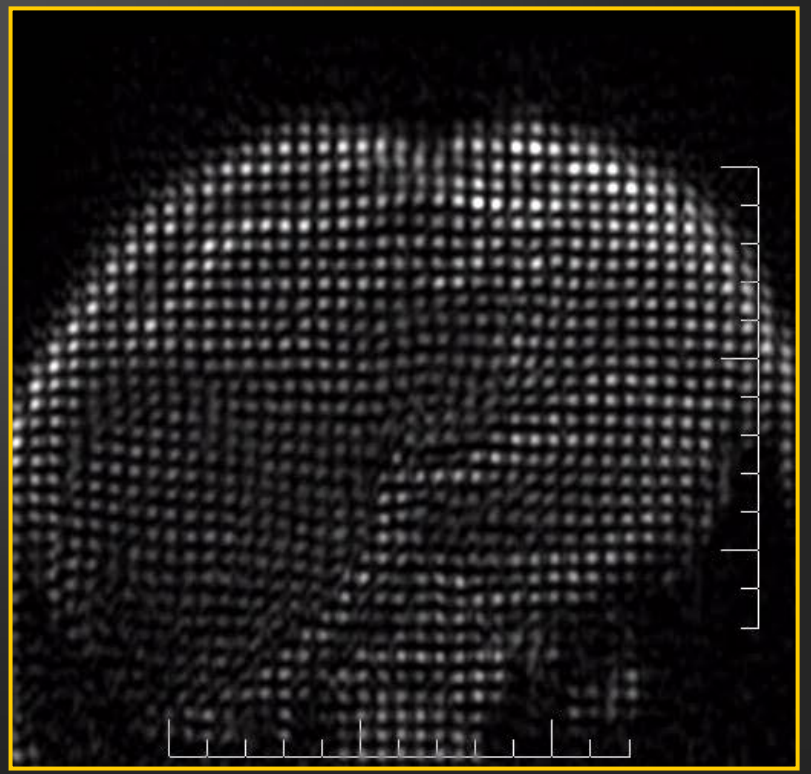

CMR – Measurements – RV/LV/PI

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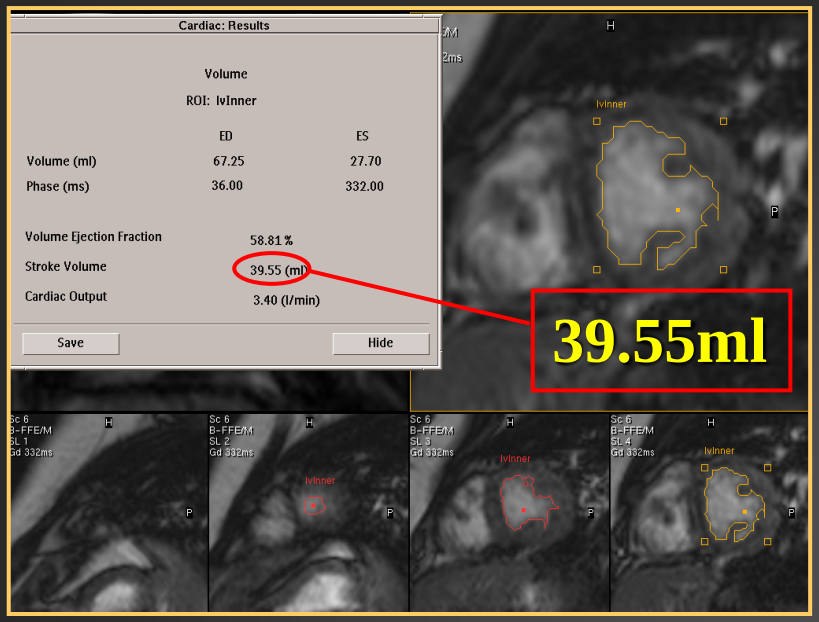

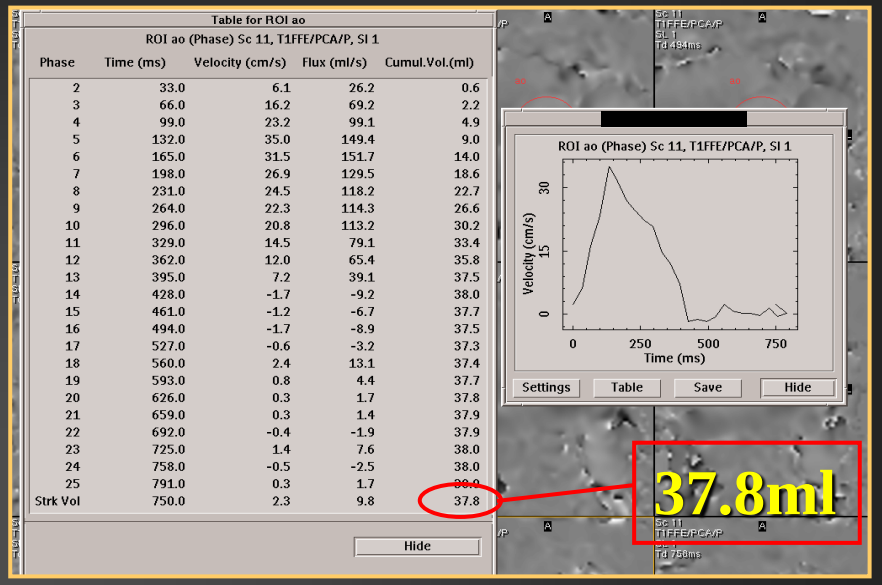

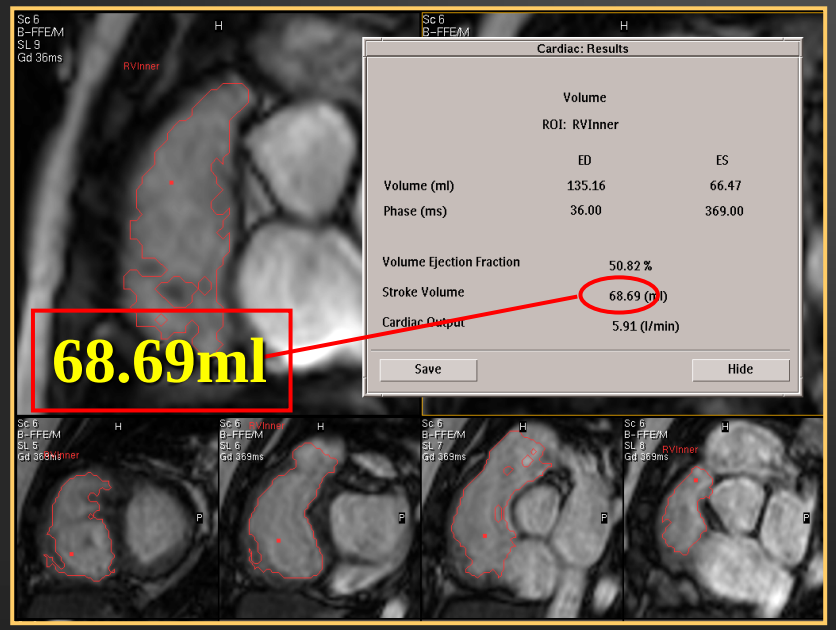

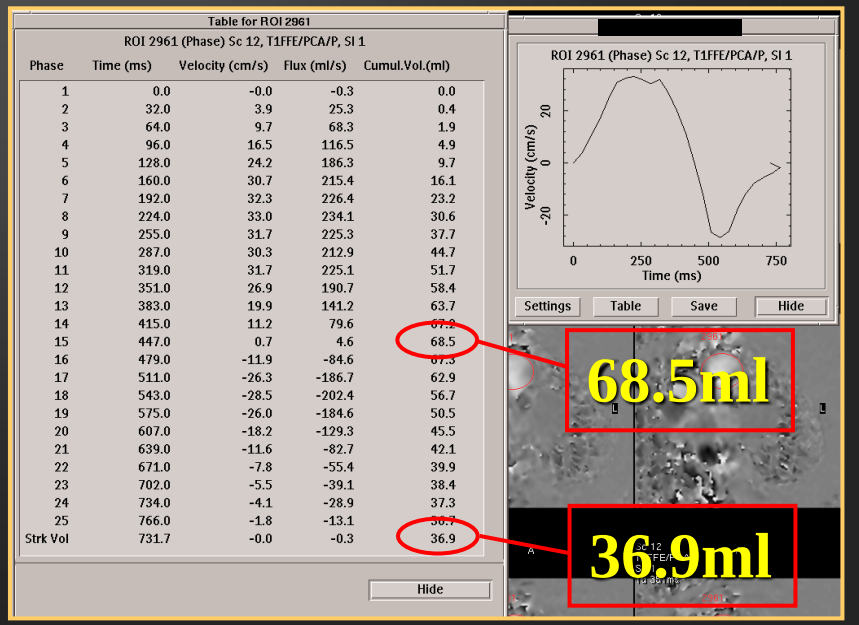

CMR - Myocardial Enhancement

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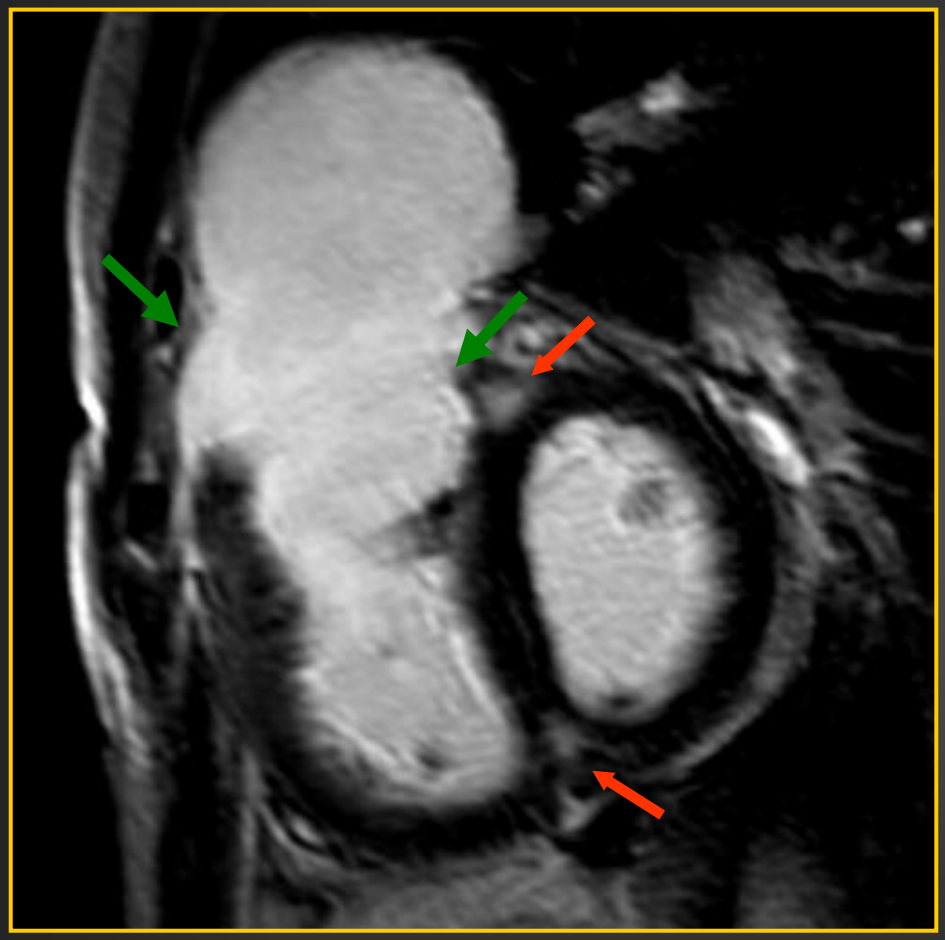

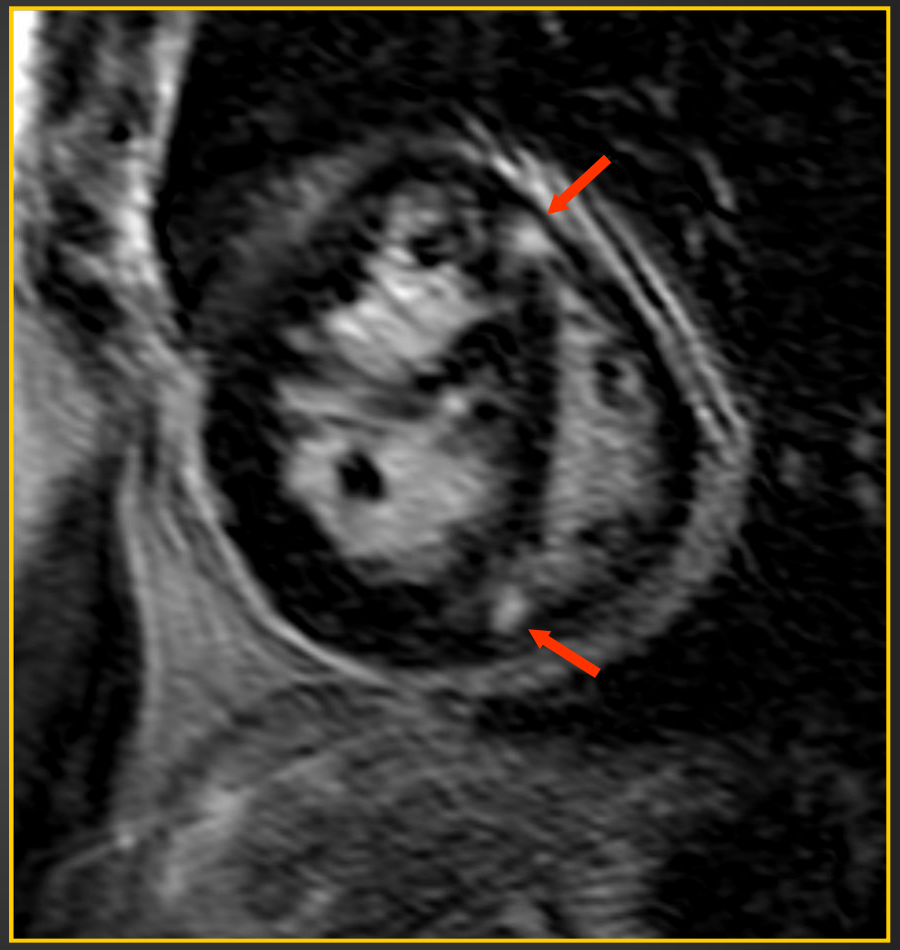

Gadolinium uptake in areas of fibrosis 10 to 20 min after contrast injection

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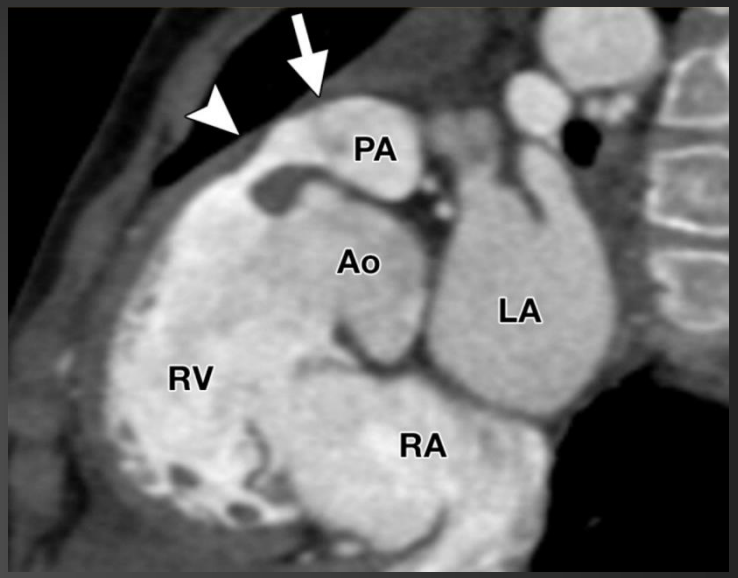
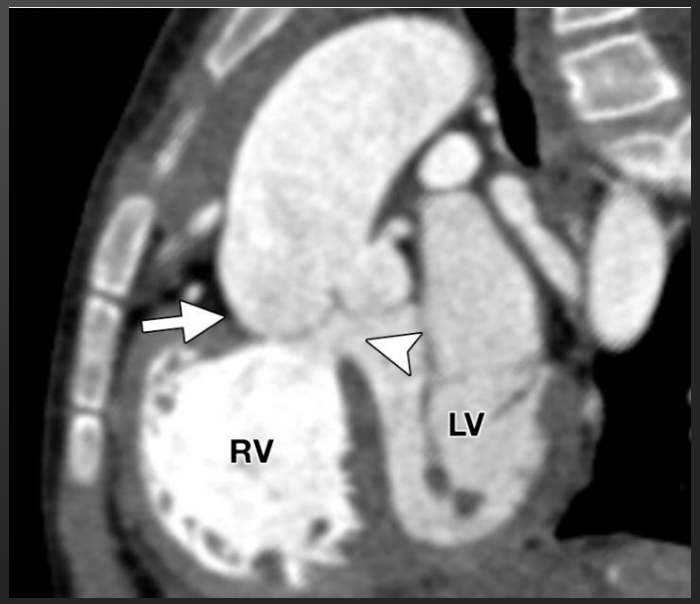
- Anatomical delineation, excellent spatial resolution
- Static and Cine imaging
- 3D reconstruction
- ↑ delineation of small vessels (coronaries, distal PA branches)
- Compatible with pacemakers and defibrillators
- Less artifacts by stainless-steel metallic artifacts
- Radiation, No hemodynamic information





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

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- 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

TOF – Angio – PAs – MAPCAs

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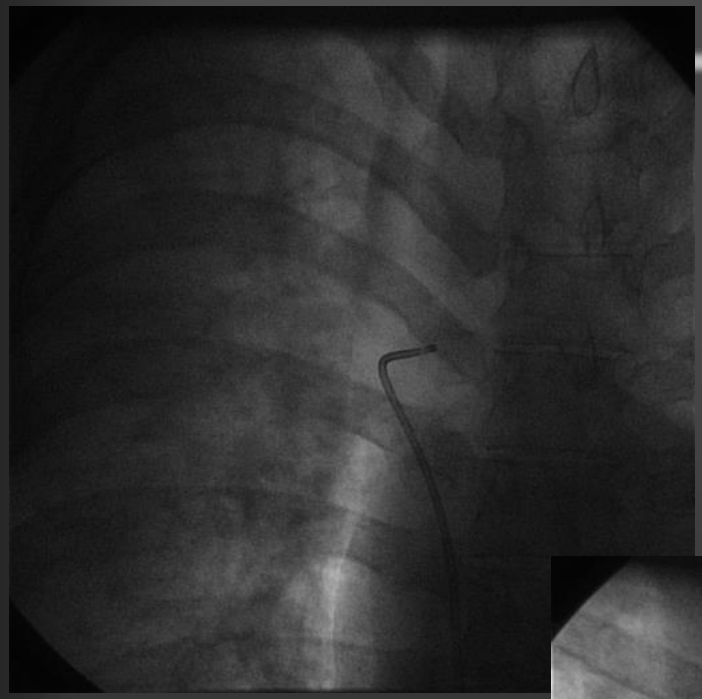

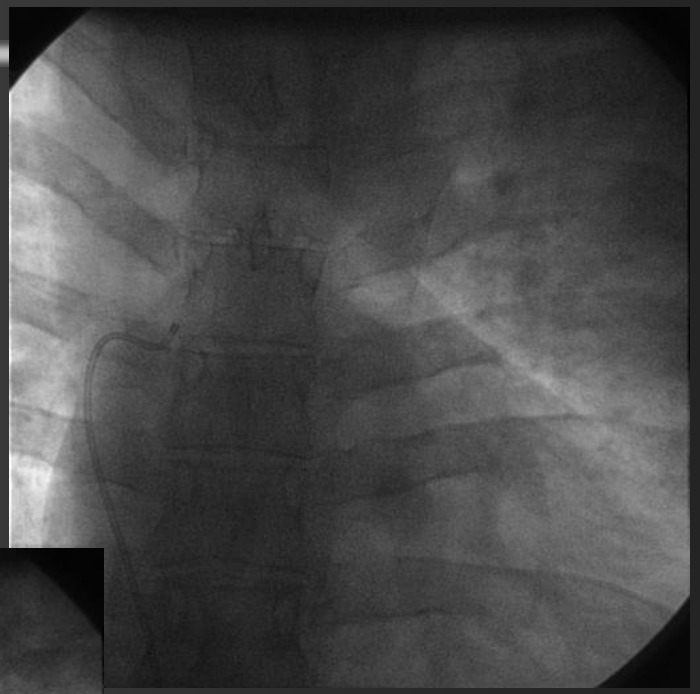

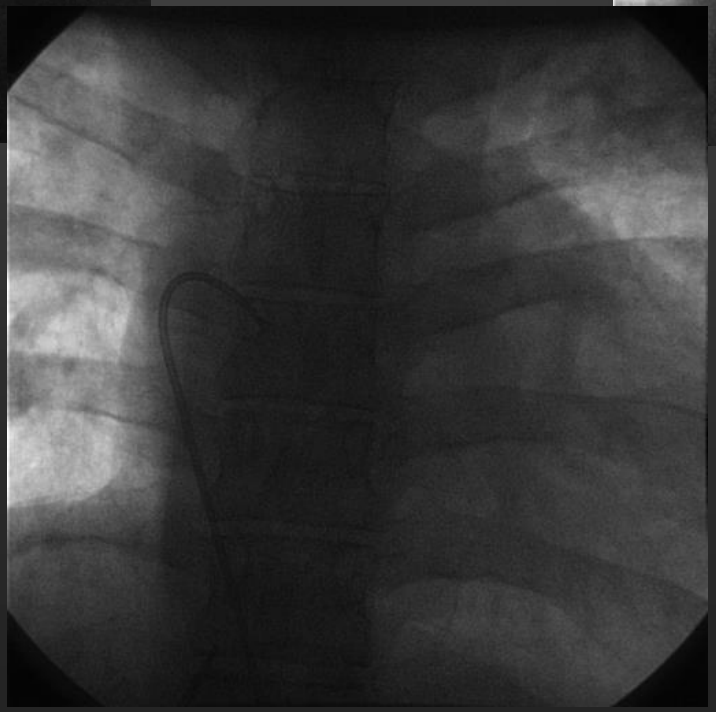

Repaired TOF – Angiography

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Medical management in HF

Table 3 Medical treatment for heart failure related to intrinsic myocardial dysfunction

Systolic HF		
Systemic ventricle		
Morphological left ventricle (EF < 40%)	Asymptomatic or symptomatic	RAAS blockers β-Blockers Mineralocorticoid receptor antagonists Diuretics (loop and thiazide) Digoxin
Morphological right ventricle (EF < 40%)	Asymptomatic Symptomatic	No medical treatment RAAS blockers Beta-blockers Mineralocorticoid receptor antagonists Diuretics (loop and thiazide) Digoxin
Sub-pulmonary ventricle		
Morphological left or right ventricle (EF < 40%)	Asymptomatic	No medical treatment
	Symptomatic	Diuretics (loop and thiazide) Mineralocorticoid receptor antagonists Pulmonary vasodilators (PAH)
Single ventricle		
Fontan circulation (EF < 40%) Morphological left ventricle	Asymptomatic	RAAS blockers β-Blockers Mineralocorticoid receptor antagonists Digoxin
Morphological right ventricle	Asymptomatic	No medical treatment
Morphological left and right ventricle	Symptomatic	RAAS blockers β-Blockers Mineralocorticoid receptor antagonists Diuretics (loop and thiazide) Digoxin
Persistent right-to-left shunt	Asymptomatic Symptomatic	No medical treatment Diuretics (loop and thiazide) Agents reducing afterload

- Different behaviour RV/LV
- Therapy in RV failure extrapolated from LV failure
- Therapy in RV dysfunction without definitive proof

European Heart Journal (2016) 37, 1419–1427

Device Therapy in TOF

- Pacemaker indications as per the general population
- Prevalence of SCD, VT, or appropriate ICD shocks 6-14%
- Standard guidelines for secondary prevention of SCD in TOF
- Unclear primary prevention strategy without optimal risk stratification
- Risk factors

↑ age at repair
QRS ≥180 msec
RV dilatation and dysfunction
frequent or complex ventricular ectopy
SYNCOPE

transannular patch
↑rate of QRS prolongation
LV dysfunction, ↑ LVEDP
VT at EP study

Circulation. 2016;133:770-801

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Cardiac Resynchronization Therapy (CRT)

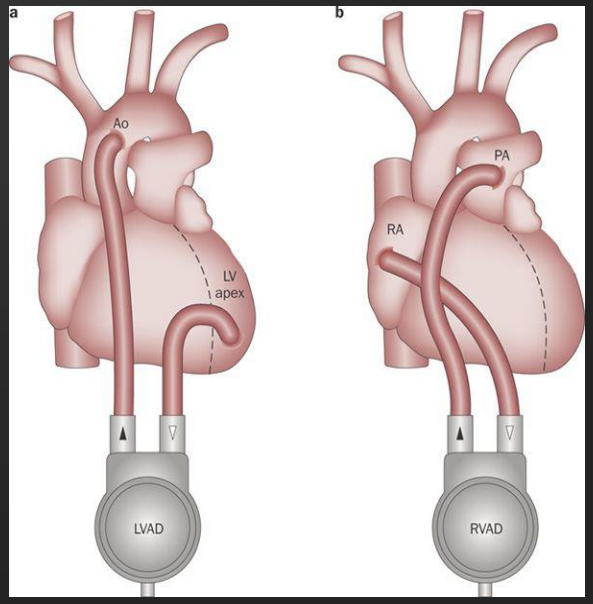
- Optimal technique and measurements for selecting patients for CRT *unclear*
- Both RV and LV systolic dysfunction must be taken into account because of interventricular interaction
- When there is LV systolic dysfunction → guidelines for acquired heart disease
- Will CRT benefit TOF patients with RV dysfunction and RBBB and prevent LV dysfunction?
- If there is no correctable cause of ventricular dysfunction, patients should be managed on a case-by-case basis
- *Outcome of CRT in TOF yet unknown*

Circulation. 2016;133:770-801

Transplantation – Mechanical Support

- Heart transplantation
  - CHD comprises 10% of referrals
  - 30d mortality 20–30%, same long term mortality at 10 years
  - Referral for severe symptoms, ↓ exercise tolerance, frequent hospitalizations, malignant arrhythmias
  - *ATTENTION* to pulmonary hypertension, organ dysfunction, surgical risk

- Ventricular assist devices (VADs)
  - Less frequent use as bridge to transplantation or destination therapy
  - ↑ complications due to comorbidities



Heart transplantation in CHD

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Table 4. CHD-Specific Issues That May Affect Candidacy for and Risk of Transplantation

Issue	Reason	Outcome
Sensitization[271,272]	Use of homografts	Requirement for a prospective crossmatch or presence of PRA >25% associated with wait-list time and increased mortality[271,273]
		Strategies to address sensitization, including need for negative crossmatch, delaying time to transplantation; desensitization strategies may increase risk[263]
	Previous blood transfusions	Presence of donor-specific antibodies increases risk of antibody-mediated rejection and allograft vascular disease[274–278]
Pulmonary hypertension[37,270]	High left atrial filling pressures, cyanosis, volume overload, high shear force, and abnormal development of the vasculature and lungs	Increased risk of right heart failure after transplantation associated with increased perioperative mortality[279]
Surgical challenges	Adhesions, AP collaterals	Increased risk of bleeding, prolonged operative times
	PA reconstruction	Increased mortality[280]
	Previous sternotomy	Increased ischemic times
Liver issues	Passive congestion	Increased morbidity and mortality with increasing MELD scores
	Cirrhosis	
	Portal hypertension	Increased frequency in CHD[281]
	Hepatitis B and C	
Fontan physiology	PLE, liver dysfunction secondary to passive congestion	Increased risk vs other CHD diagnosis, increased risk of bleeding and infection
Eisenmenger syndrome	Severe pulmonary hypertension	Need for heart-lung transplantation associated with poorer outcomes (ISHLT registry 2012 data)[264]
		Consider lung transplantation with primary cardiac repair[282]

Circulation. 2016;133:770-801

Follow up in TOF with HF

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- F/U in Adult CHD Centers
- Nutrition
- Exercise programs
- Psychological Support
- Social Support
- HF Programs (as in acquired heart disease): education, nutrition, exercise, planning of home therapy, close F/U, social and psychological support

Surveillance frequency in repaired TOF

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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

HF in TOF - Conclusions

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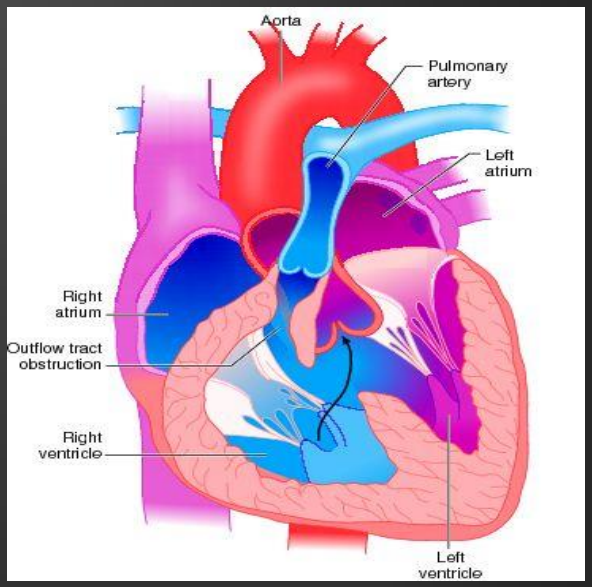
- HF affects the RV but in later stages also the LV
- Progress in understanding the pathophysiology and management
- F/U with clinical, imaging, exercise, biomarker and arrhythmia indices (incidence of SCD)
- Complex evolving management (nutrition, exercise, medical, surgical, interventional, CRT and device therapy, mechanical support and transplantation)
- Comprehensive HF programs could be beneficial as in acquired heart disease


Aorta
Pulmonary artery
Left atrium
Right atrium
Outflow tract obstruction
Right ventricle
Left ventricle

CMR – Anatomy – Angio

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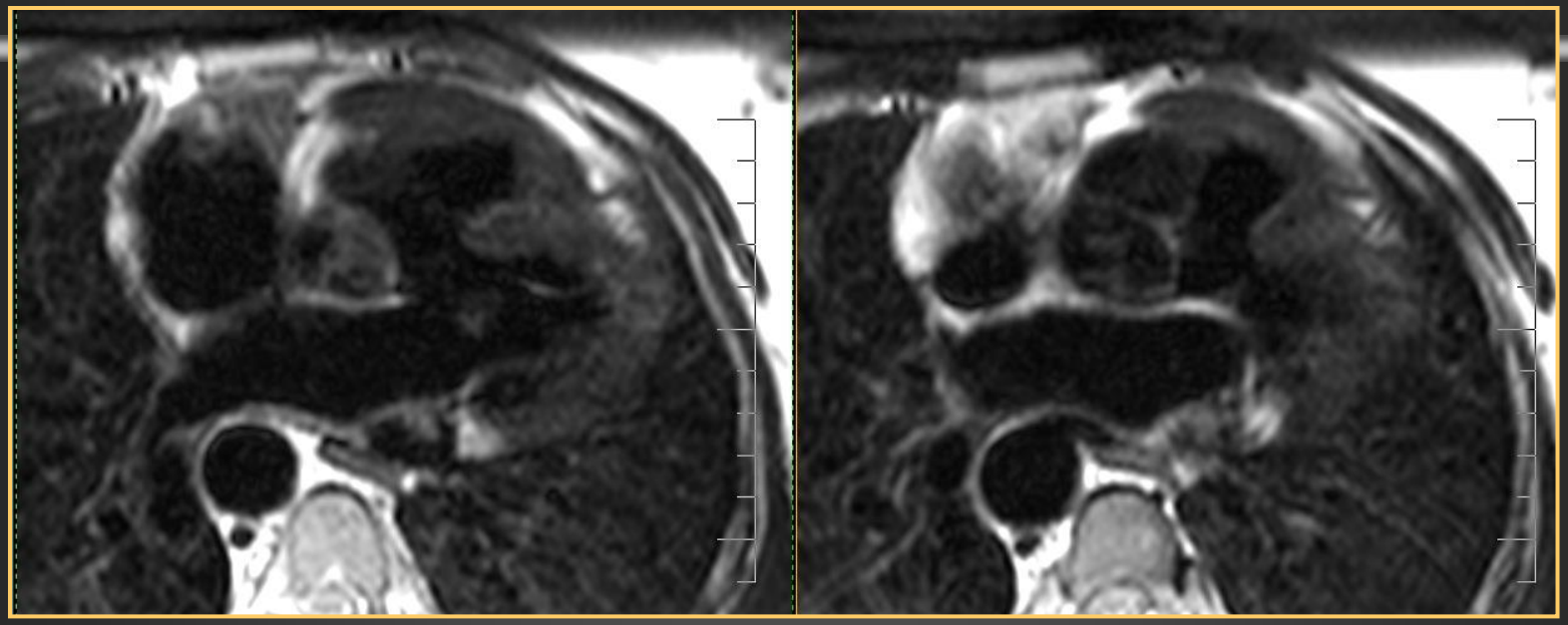

TOF - 3D printing

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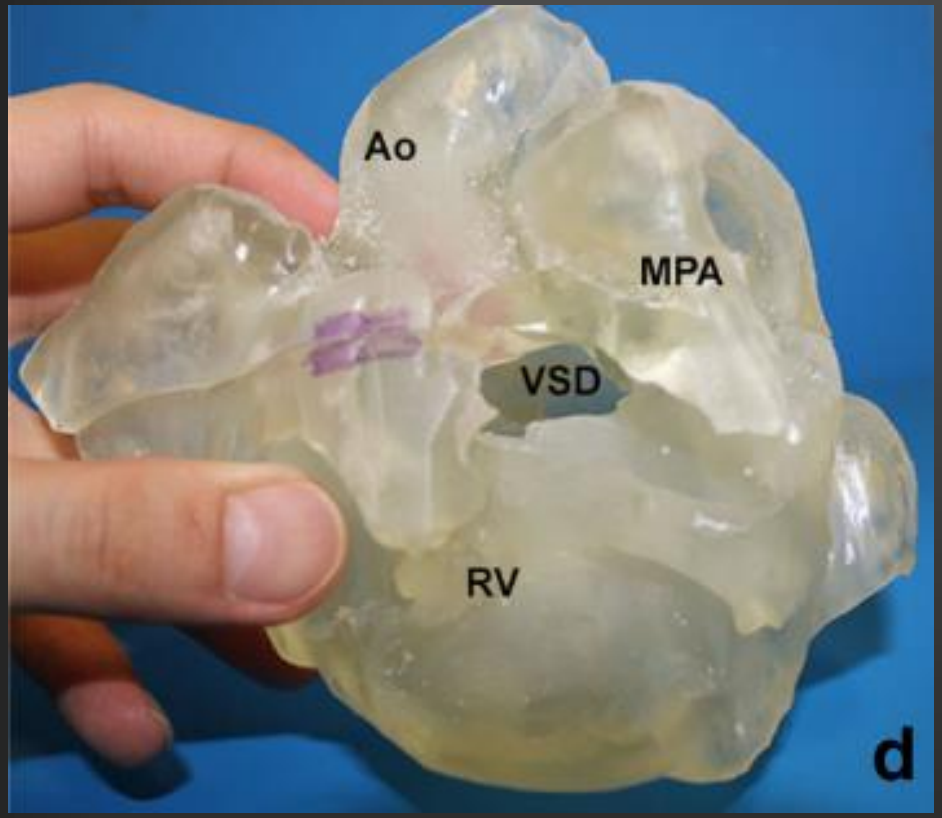

Unrepaired TOF (from ECHO)

Repaired TOF (from CMR)

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

Ορισμός Καρδιακής Ανεπάρκειας (ΚΑ)

- **Αδυναμία της καρδιάς και των νευροορμονικών συστημάτων να παράγουν ικανή καρδιακή παροχή για τις μεταβολικές ανάγκες**
- **Προοδευτικό κλινικό και παθοφυσιολογικό σύνδρομο οφειλόμενο σε καρδιαγγειακές και μη ανωμαλίες που συνοδεύεται από:**

- **χαρακτηριστικά συμπτώματα και σημεία όπως *οίδημα, αναπνευστική ανεπάρκεια, μειωμένη ανάπτυξη* και *ανοχή στην κόπωση***
- **κυκλοφορικές, νευροορμονικές και μοριακές διαταραχές**

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Cardiac causes of HF

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Table 3. Causes of HF in Patients With CHD

Volume overload resulting from left-to-right shunt lesions and valvular regurgitation
Pressure overload resulting from valvular disease and other obstructive lesions
Ventricular failure related to intrinsic myocardial dysfunction
Pulmonary hypertension caused by CHD lesions, ventricular dysfunction, or comorbidities such as obstructive sleep apnea
Systemic arterial hypertension resulting from coarctation, acquired renal disease, essential hypertension, or arteriosclerosis
Coronary artery disease related to CHD, atherosclerosis, or comorbidities such as diabetes mellitus
Cyanosis
Intractable atrial arrhythmias

«Normal heart»

- Primary cardiomyopathy
  - Dilated
  - Hypertrophic
  - Restrictive
- Secondary
  - Arrhythmogenic
  - Ischemic
  - Toxic
  - Infiltrative
  - Infectious

Circulation. 2016;133:770-801

Unrepaired TOF – Angio

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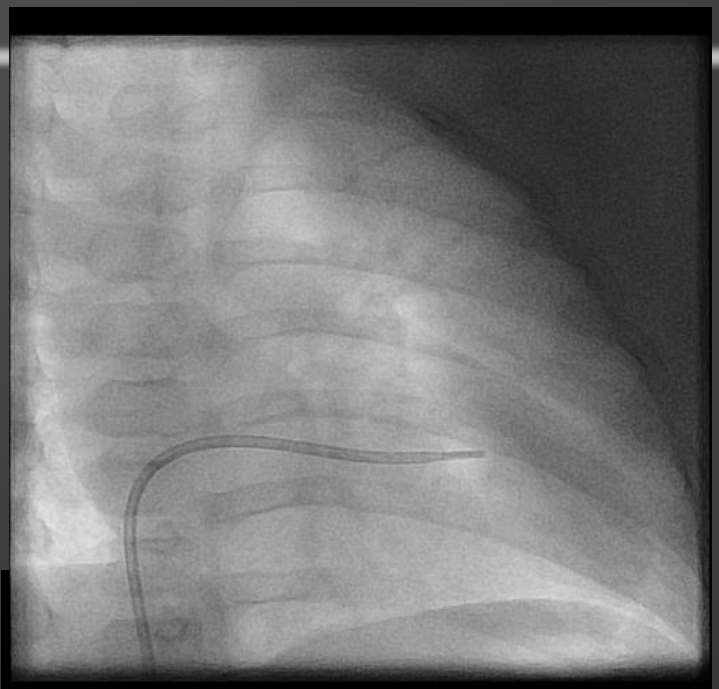

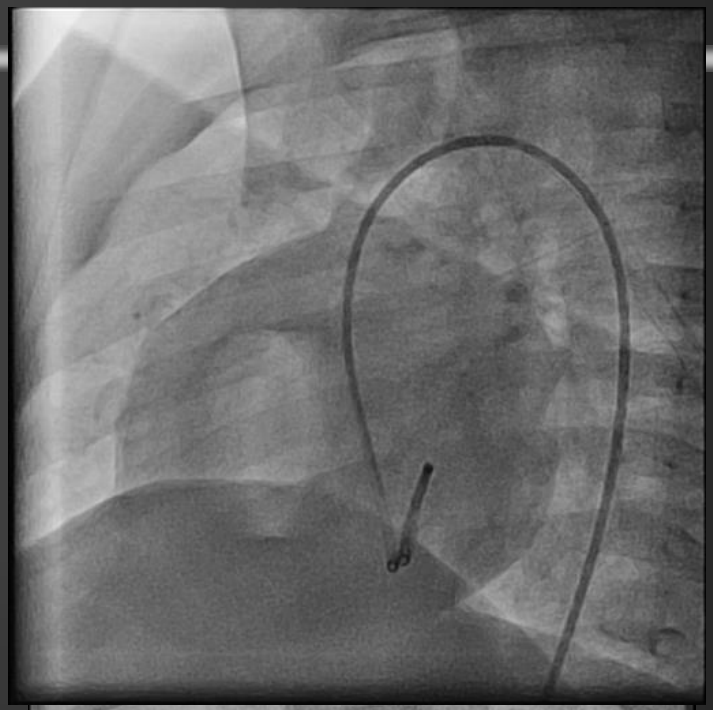

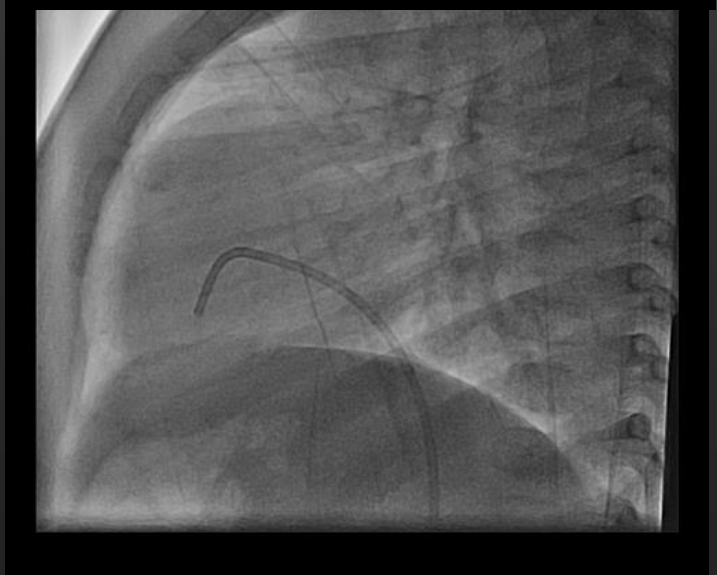

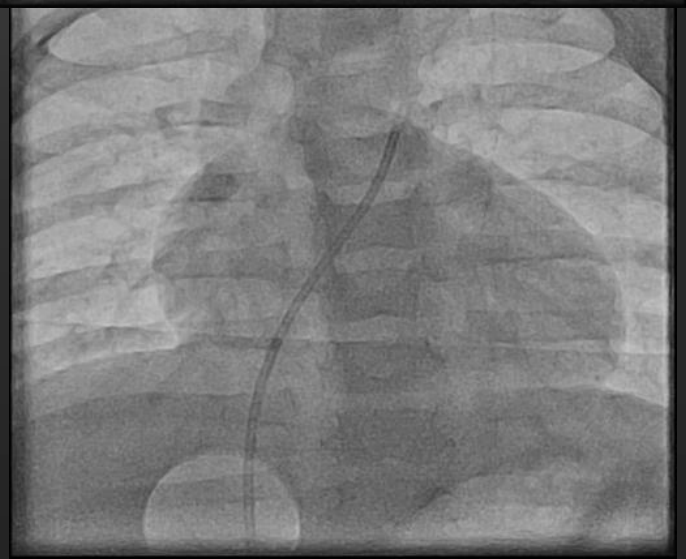

TOF – X-Ray Angiography

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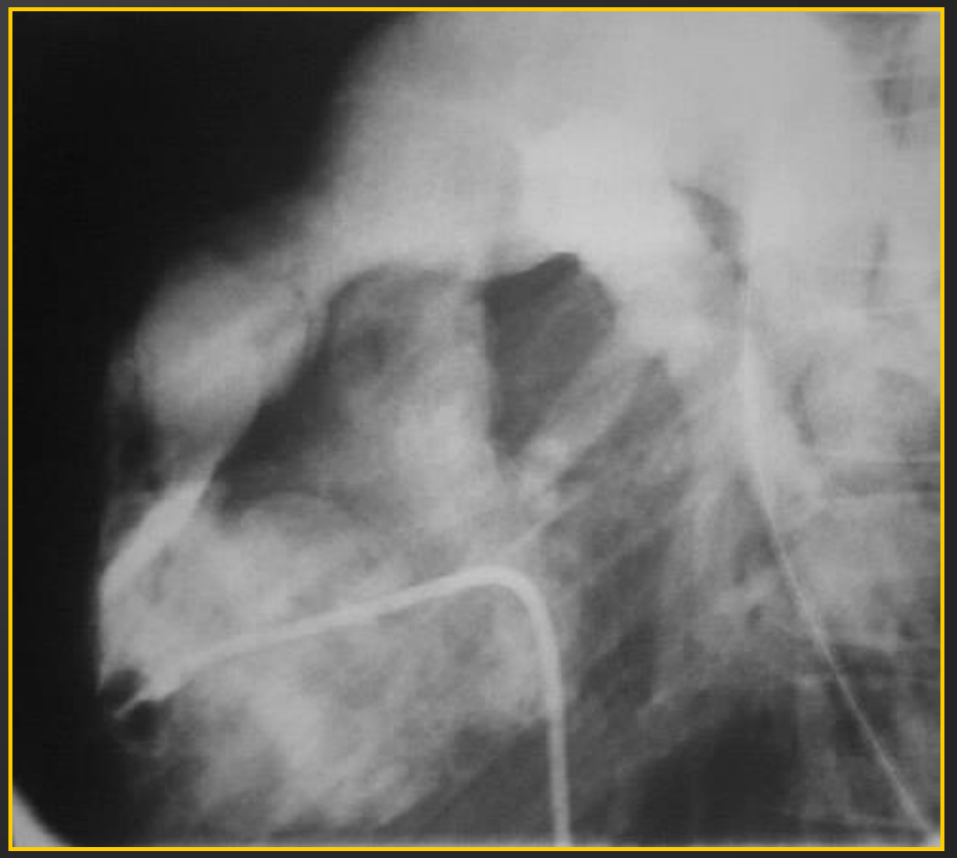

Παρακολούθηση ασθενών με ΚΑ

- **Θρέψη (πολύ σημαντική σε αναπτυσσόμενα βρέφη):**
  - ↑ θερμίδων (Infantrini, Caloreen, MCT oil)
  - ↑ προσλαμβανόμενων (ρινογαστρικός, στερεά)
  - Όχι περιορισμός Na (απαραίτητο στην ανάπτυξη)
- **Άσκηση στην ΚΑ:**
  - Σε ενήλικες βελτιώνει νευροορμονικούς δείκτες
  - Σε παιδιά βελτιώνει ανοχή στην κόπωση, συμπεριφορά, ψυχολογία
- **Ψυχολογική υποστήριξη ασθενών και οικογένειας**
- **Κοινωνική υποστήριξη ασθενών και οικογένειας**
- **Προγράμματα ΚΑ:**
  - εκπαίδευση, διατροφή, άσκηση, σχεδιασμός αγωγής στο σπίτι, στενή παρακολούθηση, κοινωνική και ψυχολογική υποστήριξη

Comparison of imaging modalities

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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 TOF
- 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

Αίτια-Αποτελέσματα ΚΑ

- Αίτια:

- Μυϊκή δυσλειτουργία (συστολική ή διαστολική)
- Μηχανικές ανωμαλίες (π.χ. παθήσεις βαλβίδων)

- Αποτελέσματα: ανωμαλίες παραγόντων καρδιακής παροχής
  - Καρδιακή συχνότητα (ΚΣ)
  - Συσταλτικότητα
  - Προφόρτιο
  - Μεταφόρτιο
  - ↑ νευροορμονικών και φλεγμονωδών παραγόντων (ρενίνη, αλδοστερόνη, νορεπινεφρίνη, BNP, NT-BNP, TNF)

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

TOF – Chest X-ray

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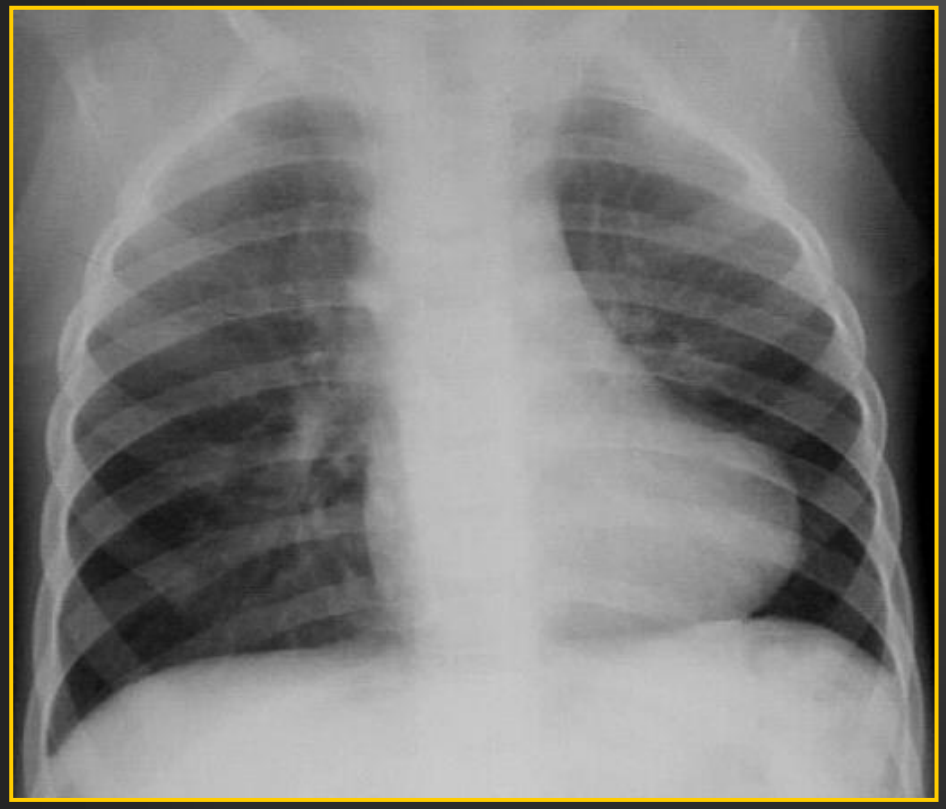

- **Boot-shaped heart (not common)**
- **↓ pulmonary markings in ↓ pulmonary blood flow**

Comprehensive ECHO views

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Subxiphoid coronal

Subxiphoid saggital

4 chamber

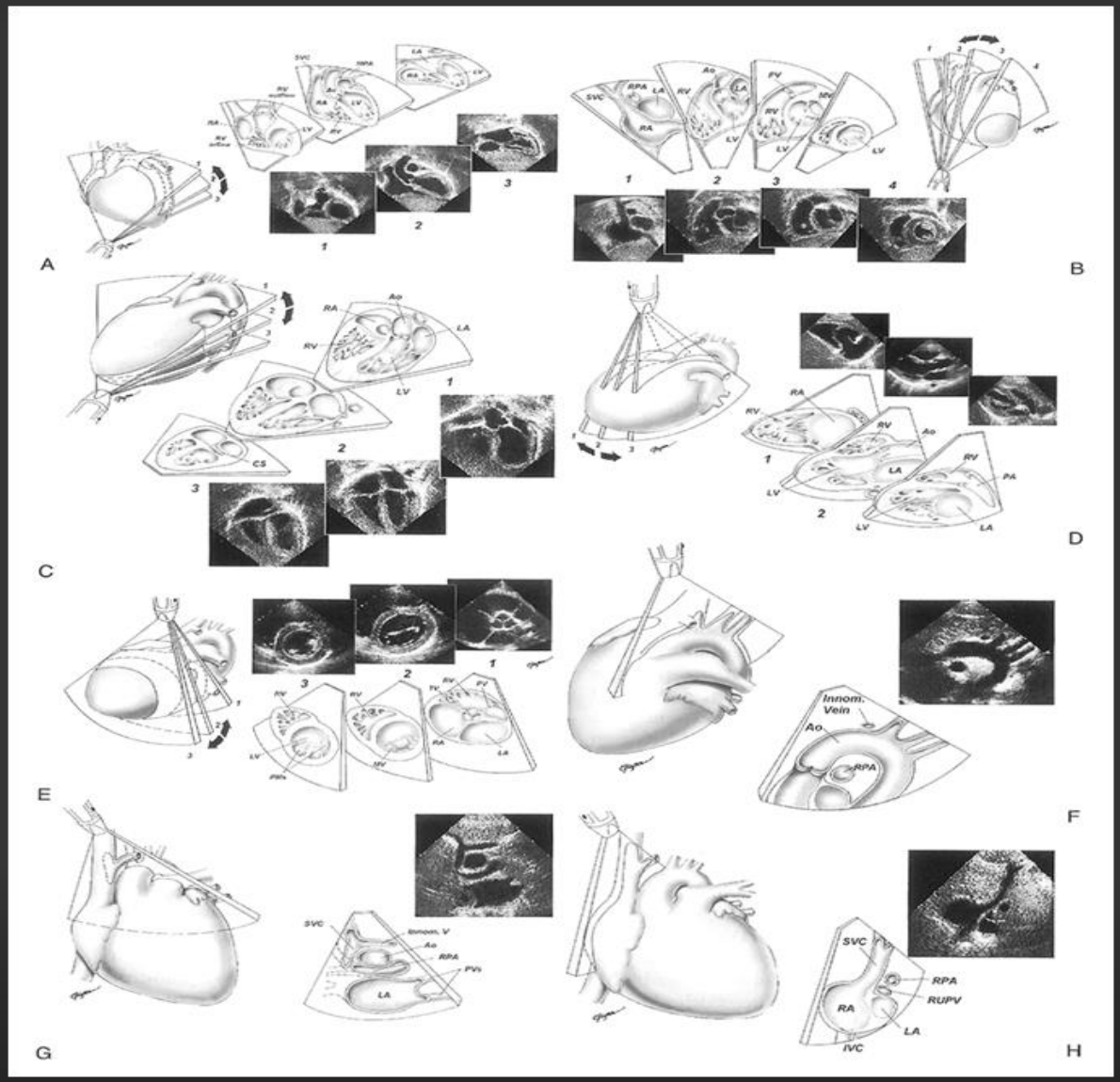
Parasternal long axis

Parasternal short axis

Suprasternal long axis

Suprasternal short axis

Suprasternal right



J Am Soc Echocardiogr 2006;19:1413-30

Unrepaired TOF - ECHO

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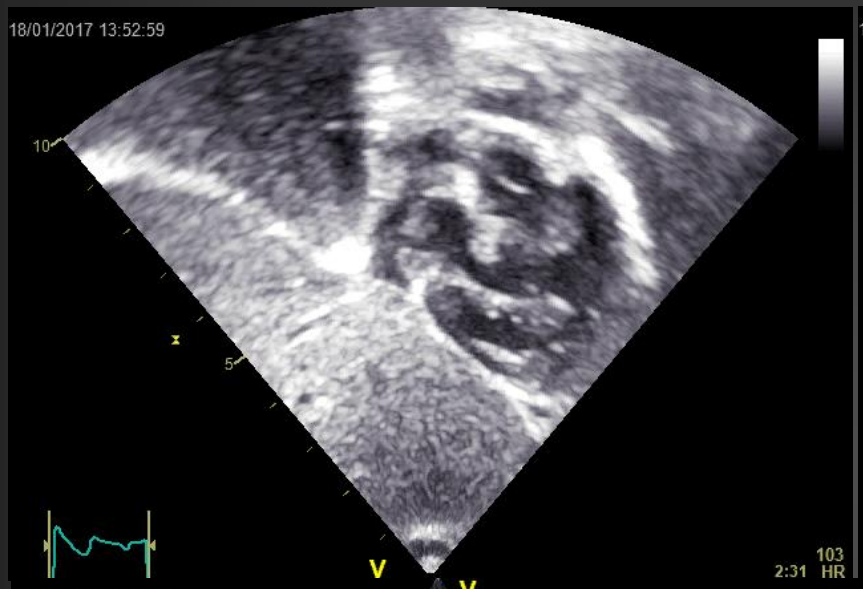

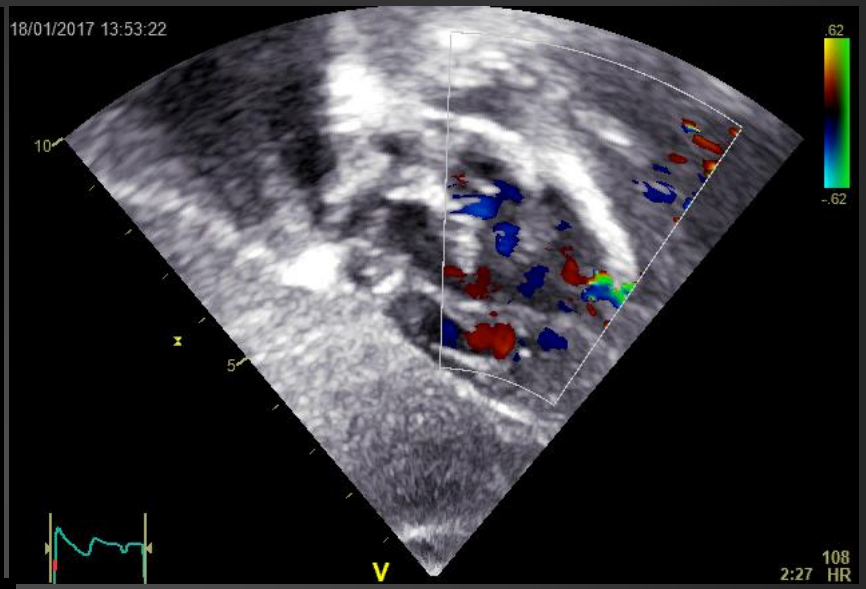

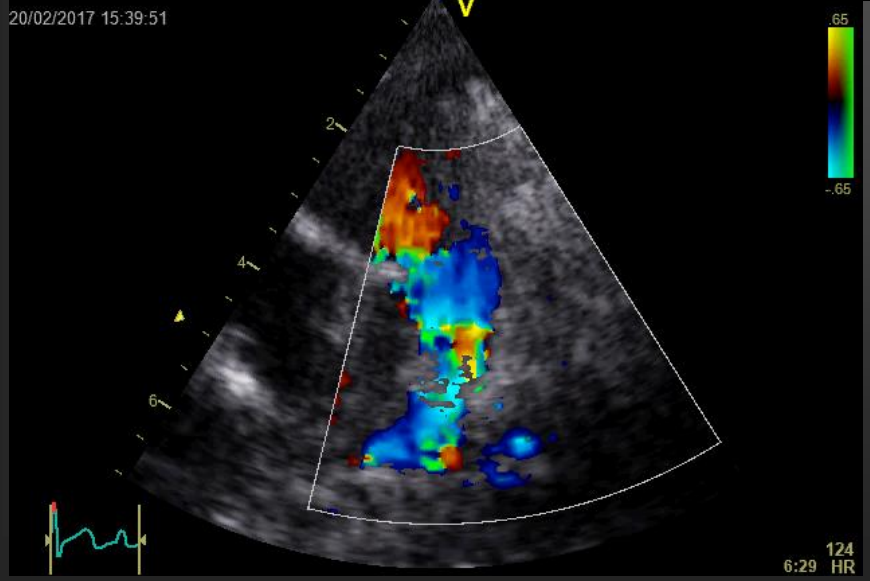

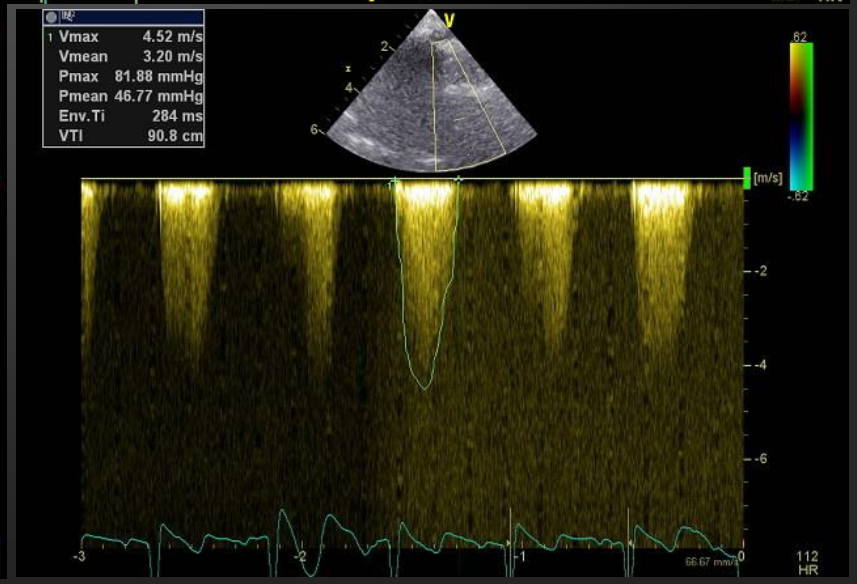

TOF CMR

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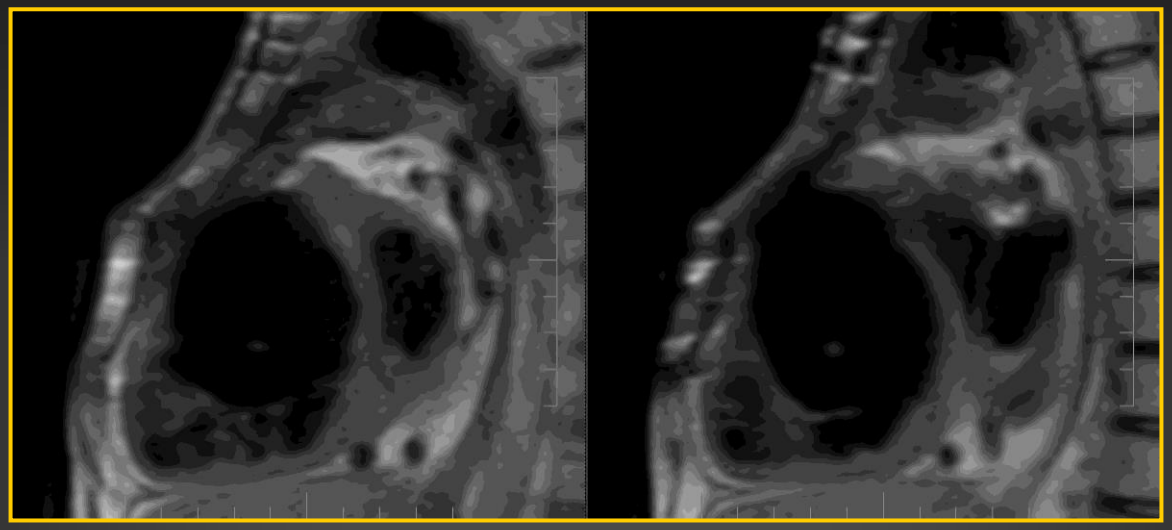
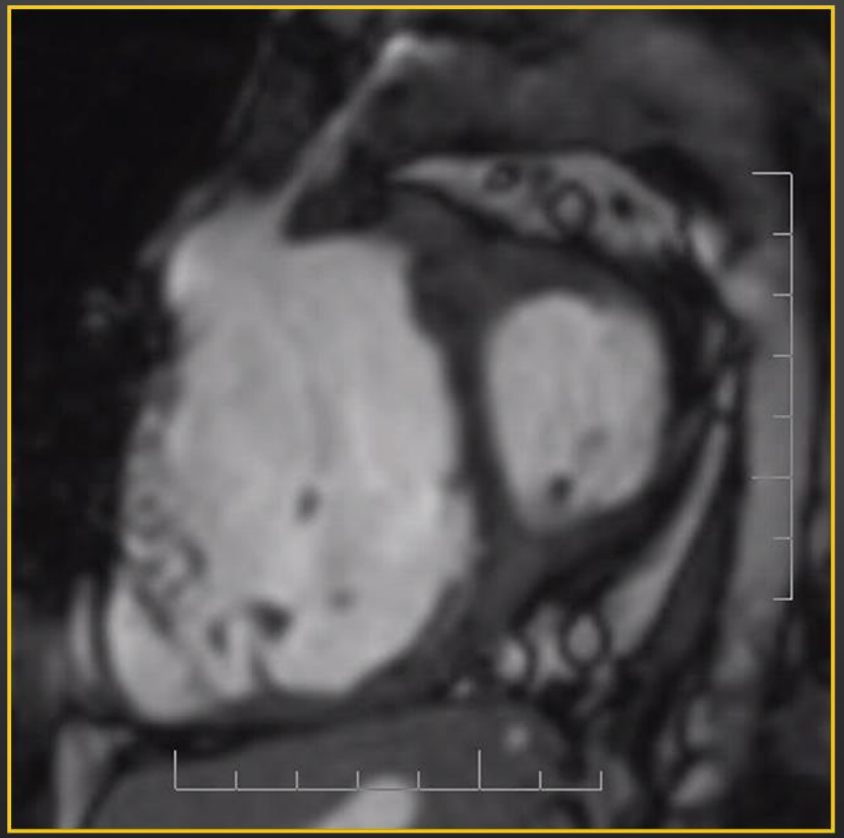
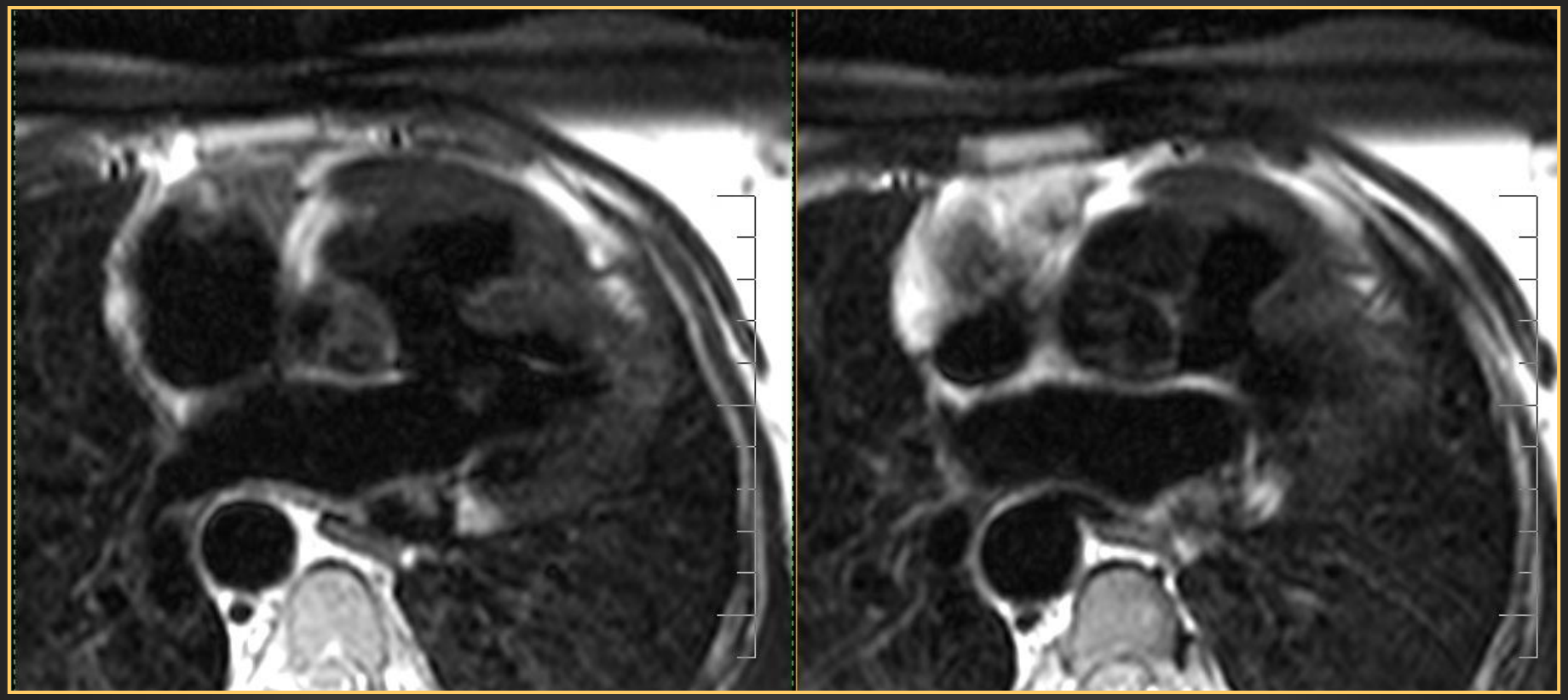
- Steady-state free precession (SSFP) → , which is a type of gradient-echo technique characterized by high signal-to-noise ratio, high T2/T1 contrast ratio, and sharp borders between the blood pool and the myocardium
- Electrocardiographically gated SSFP can be used as a cine magnetic resonance sequence, 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

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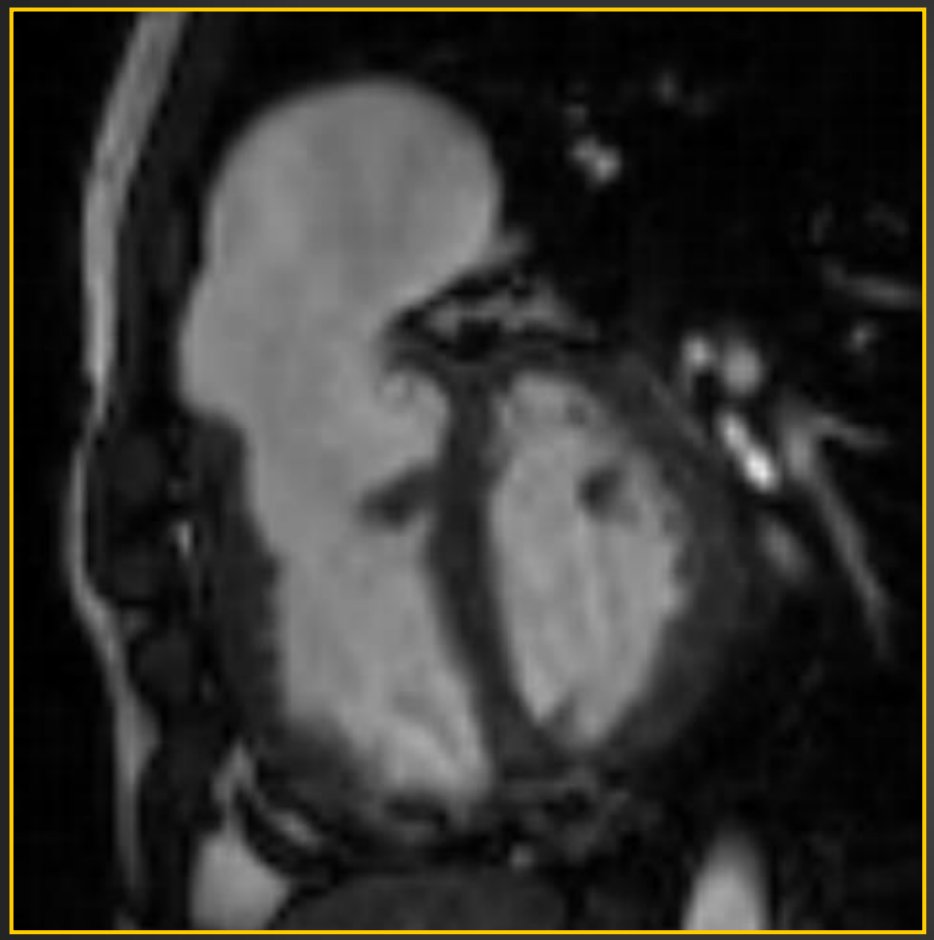

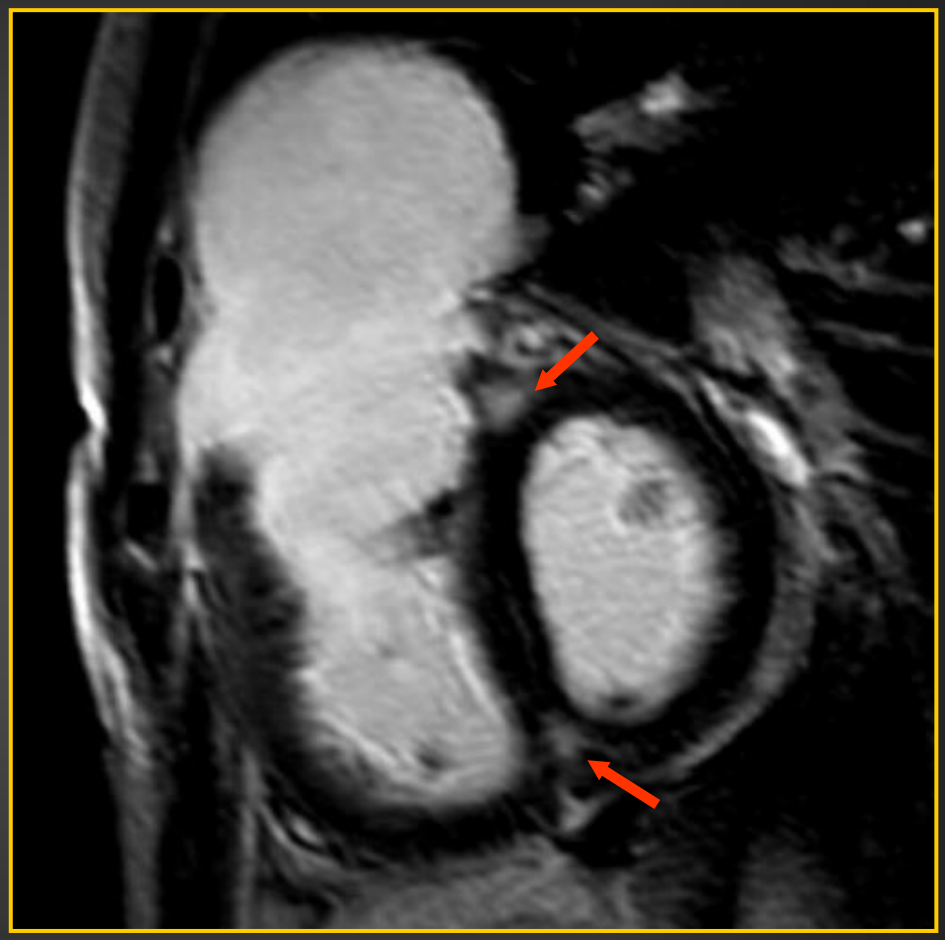

Myocardial disarray and plexiform fibrosis

CMR - Myocardial Enhancement

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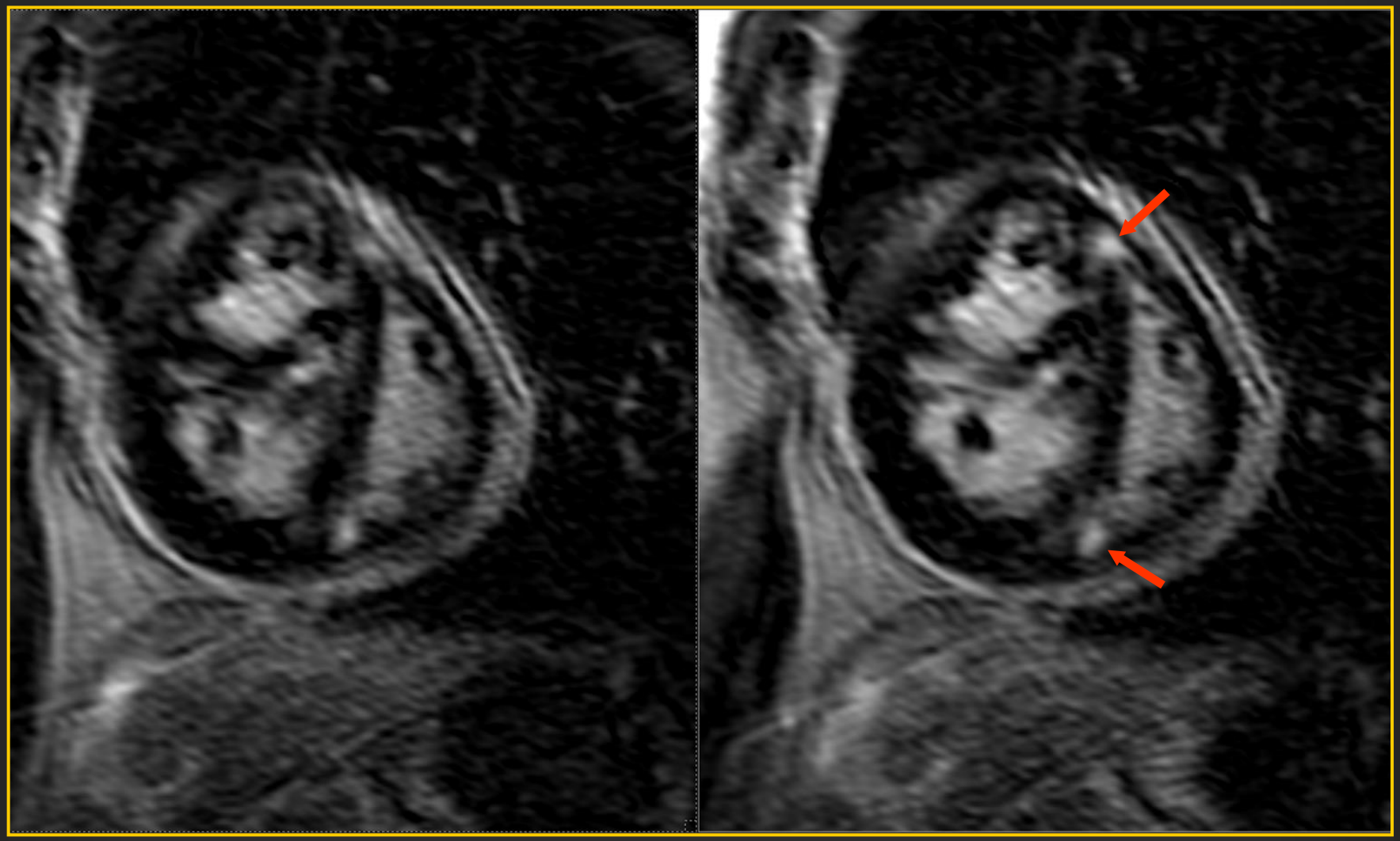
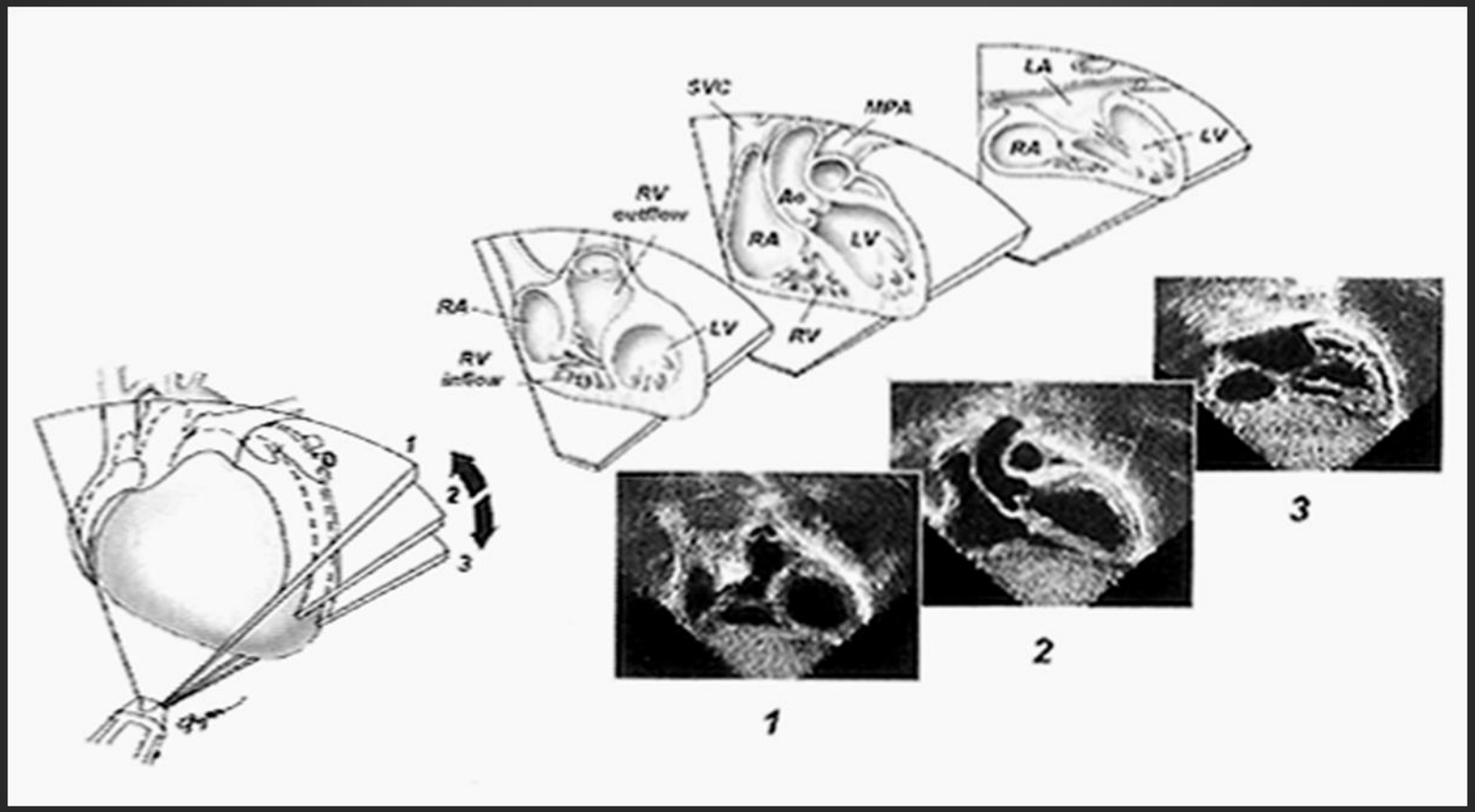


Myocardial disarray and plexiform fibrosis

Υποξιφοειδική Απεικόνιση – Στεφανιαίες τομές



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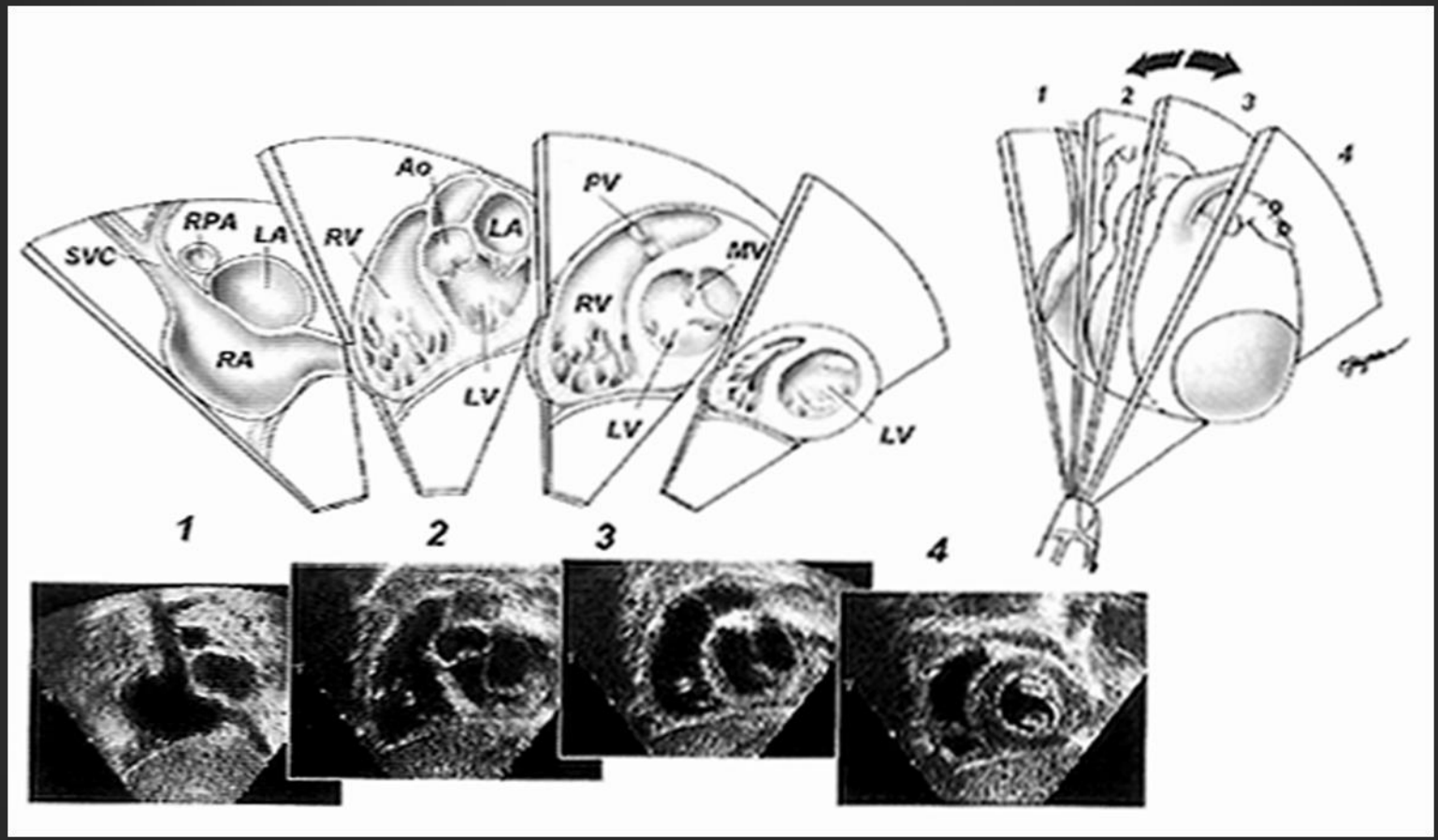
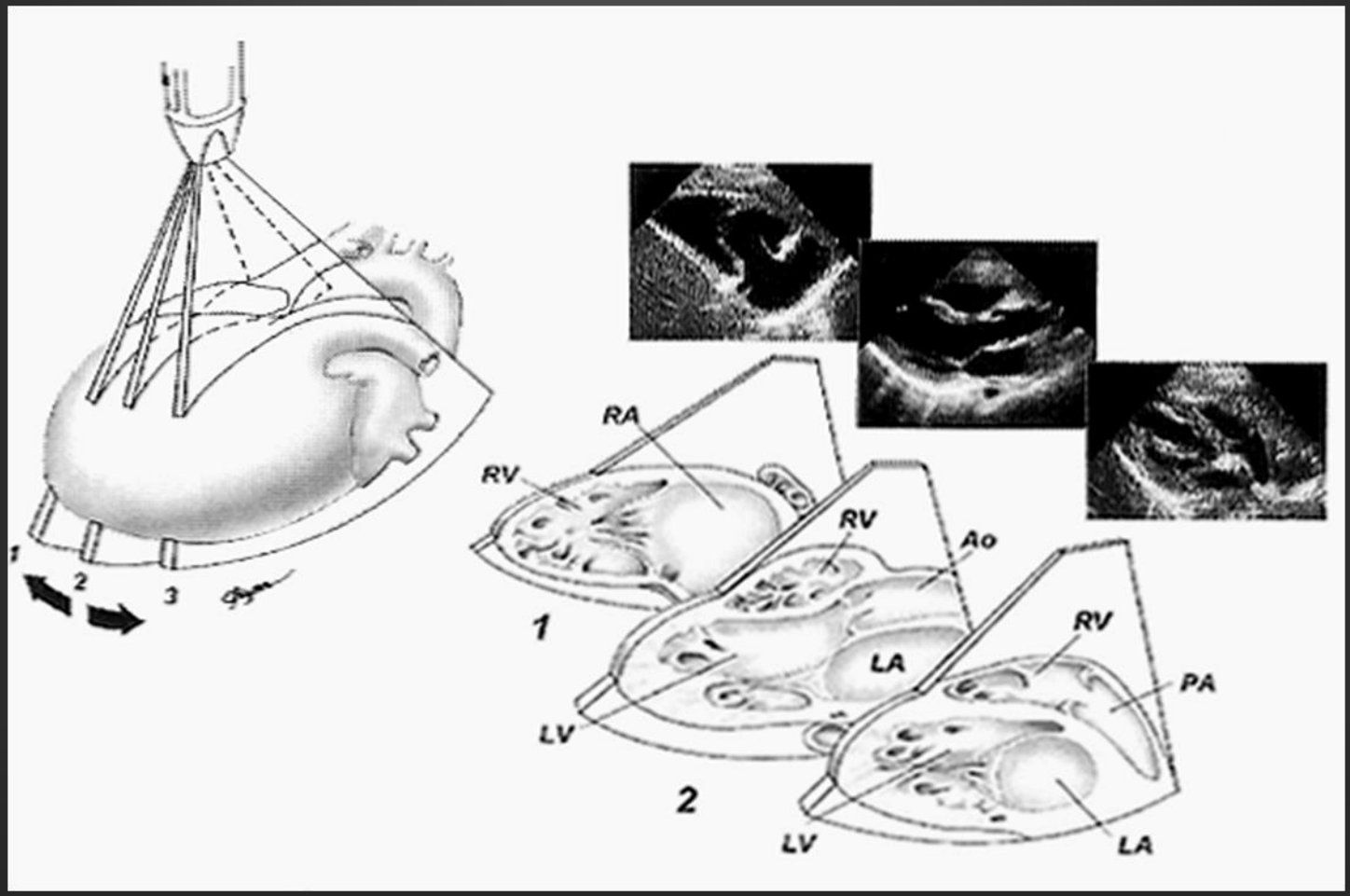


Παραστερνική Τομή κατά το Μακρύ Άξονα



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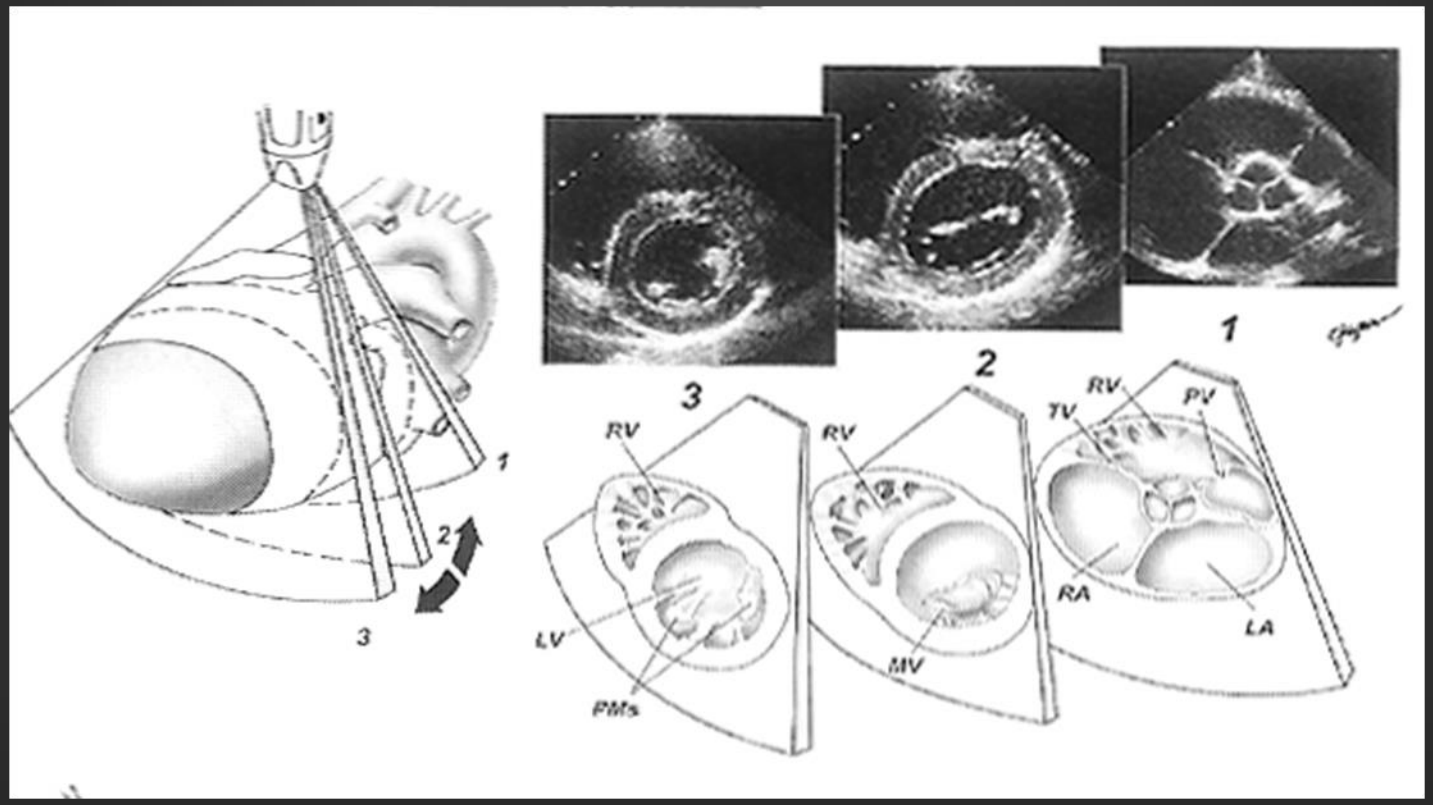
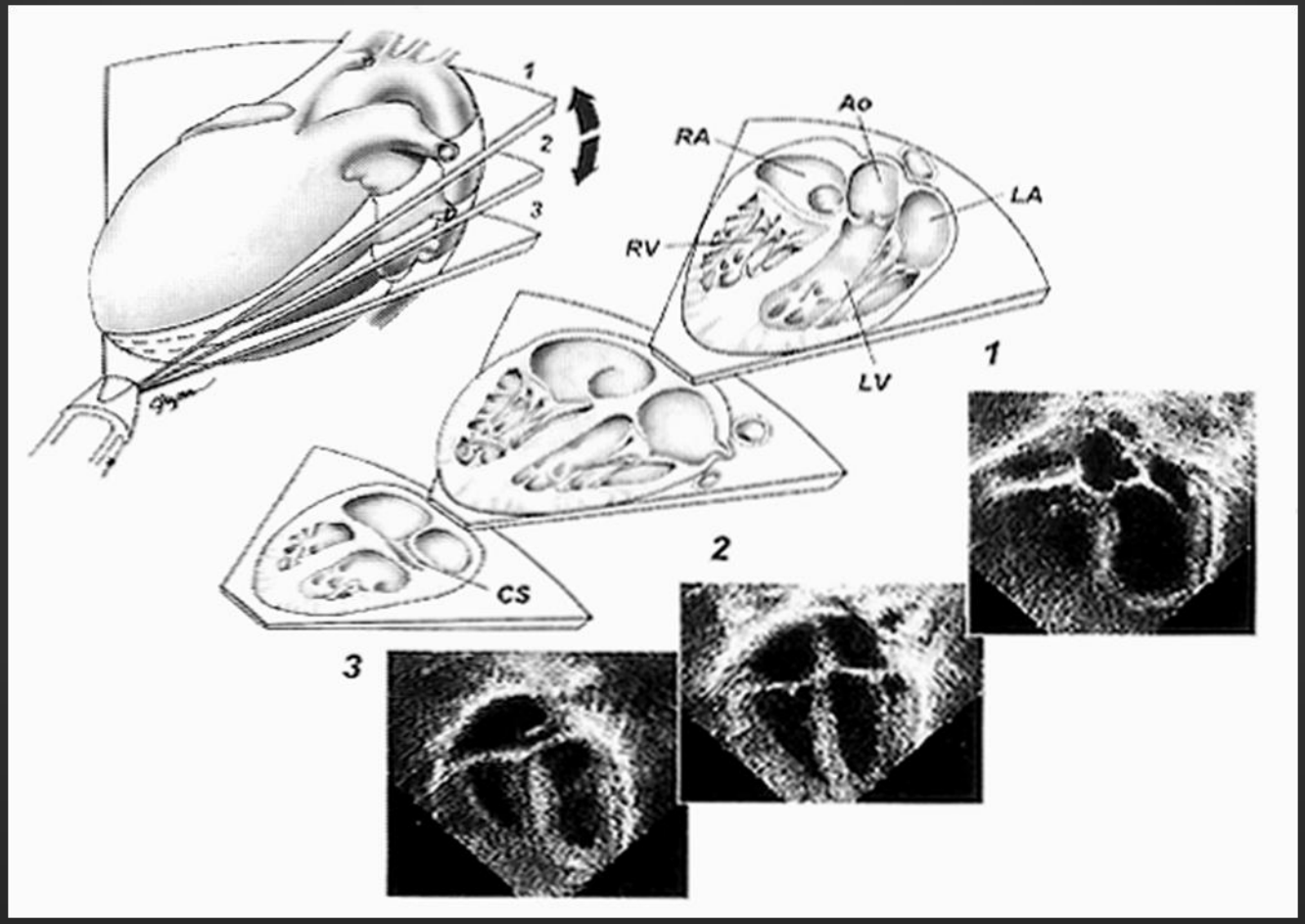
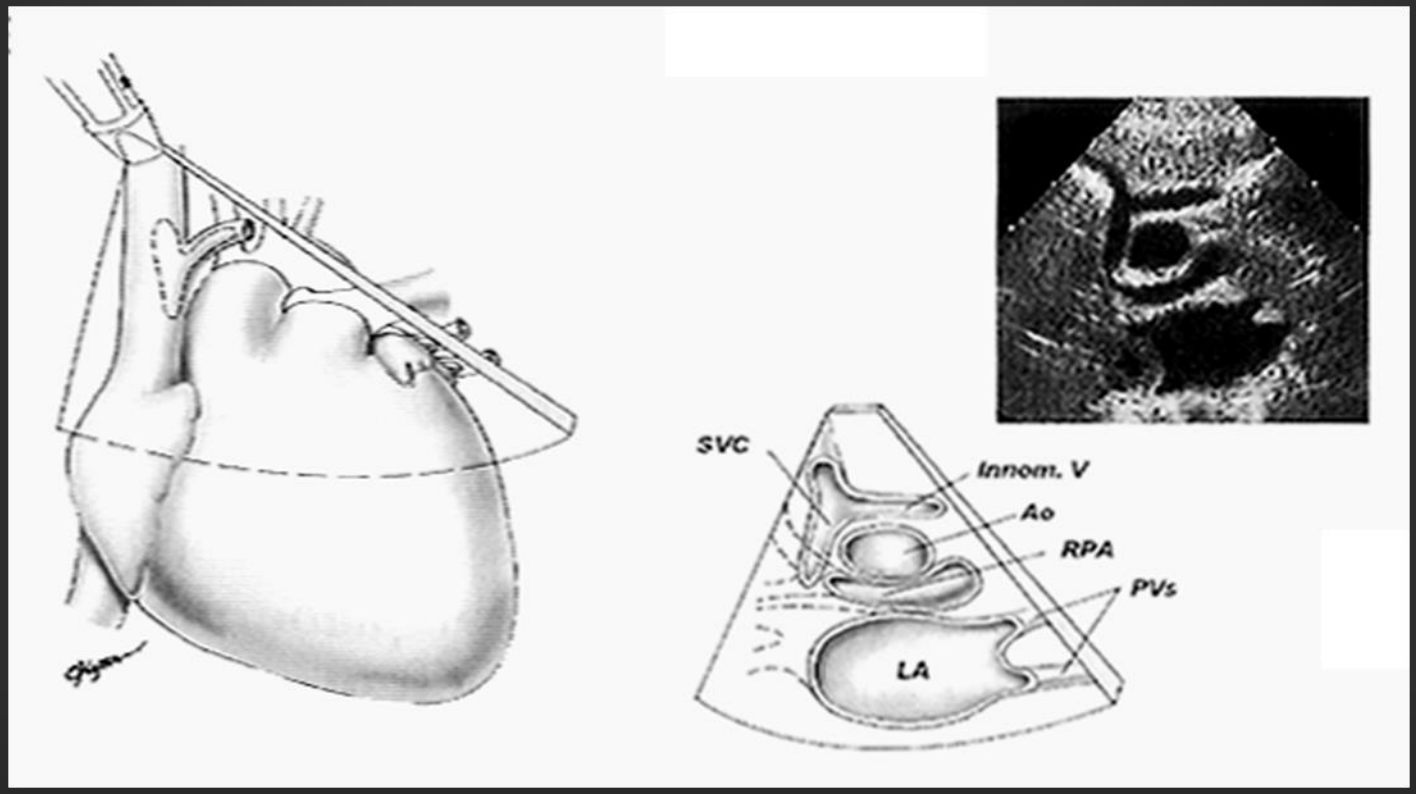


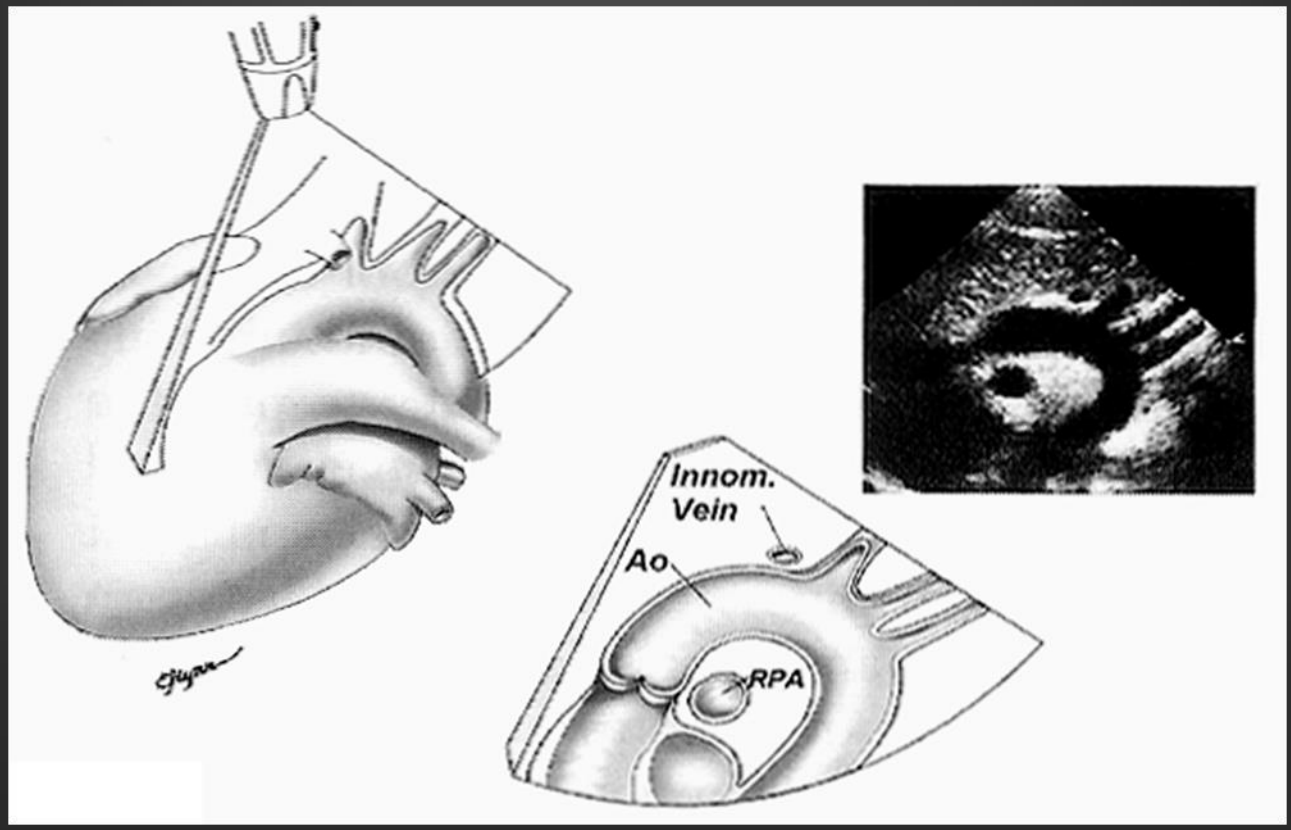
Κορυφαία Λήψη Τεσσάρων Κοιλοτήτων

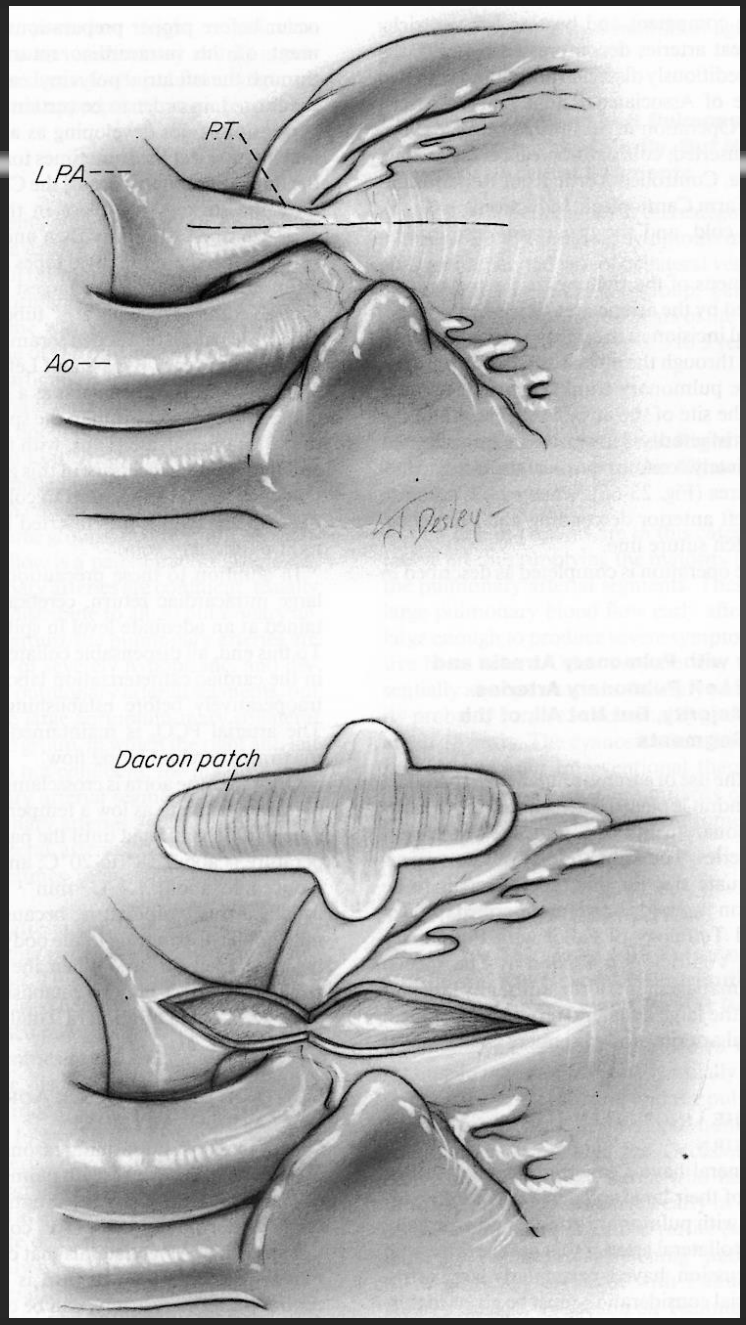


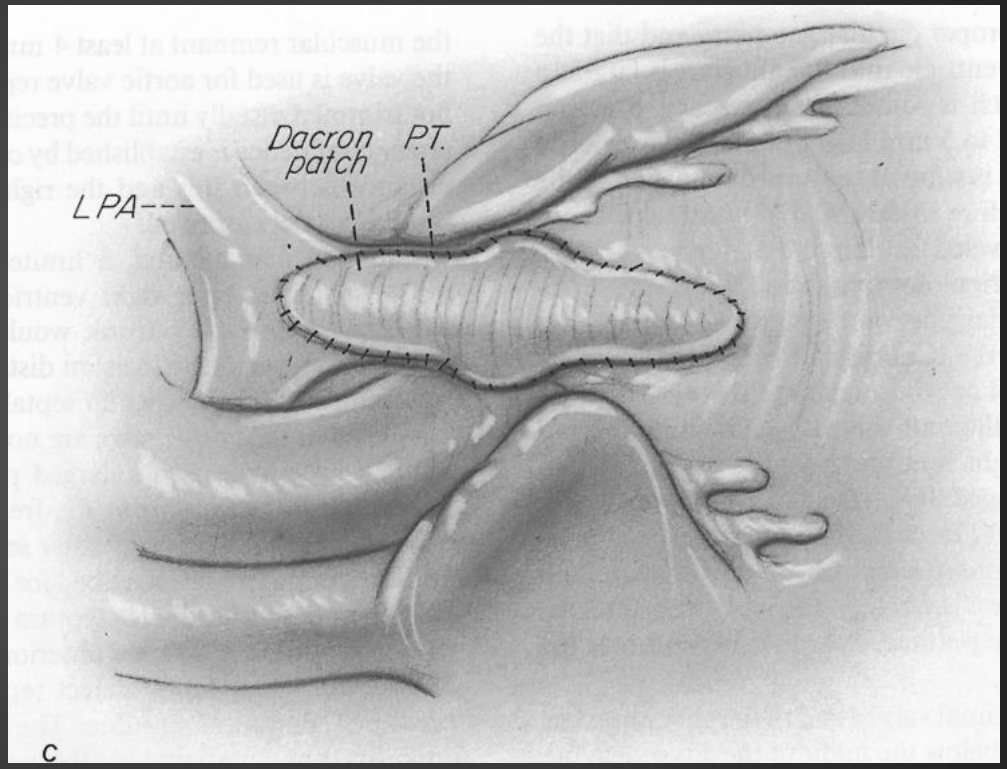
Στεφανιαία Υπερστερνική Λήψη



Οβελιαία Υπερστερνική Λήψη




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


Dacron patch
P.T.
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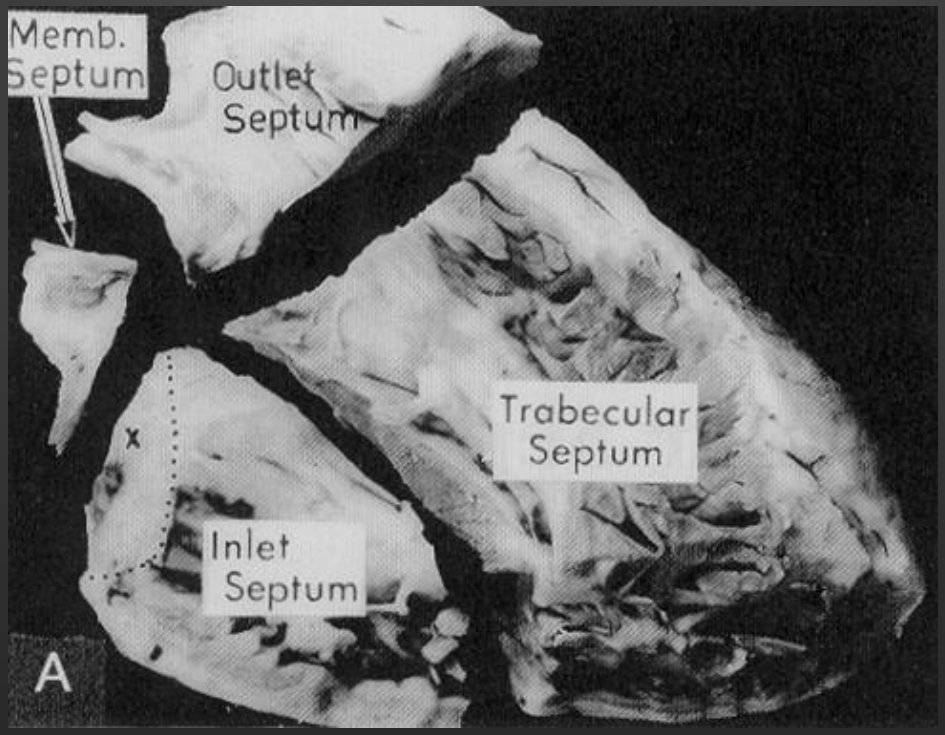
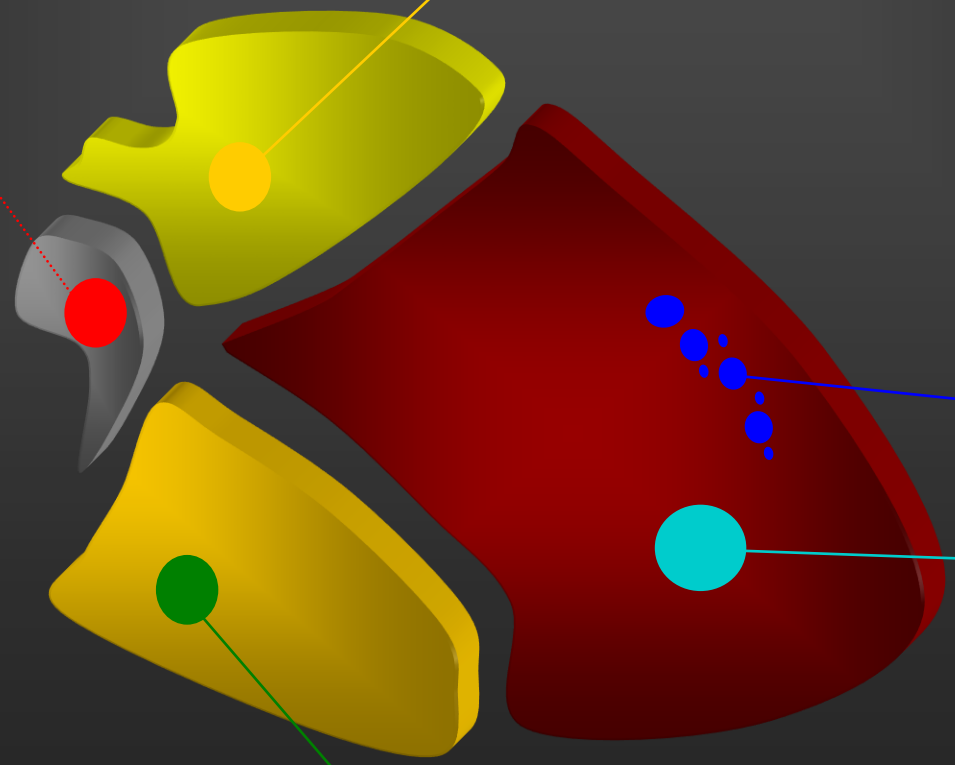
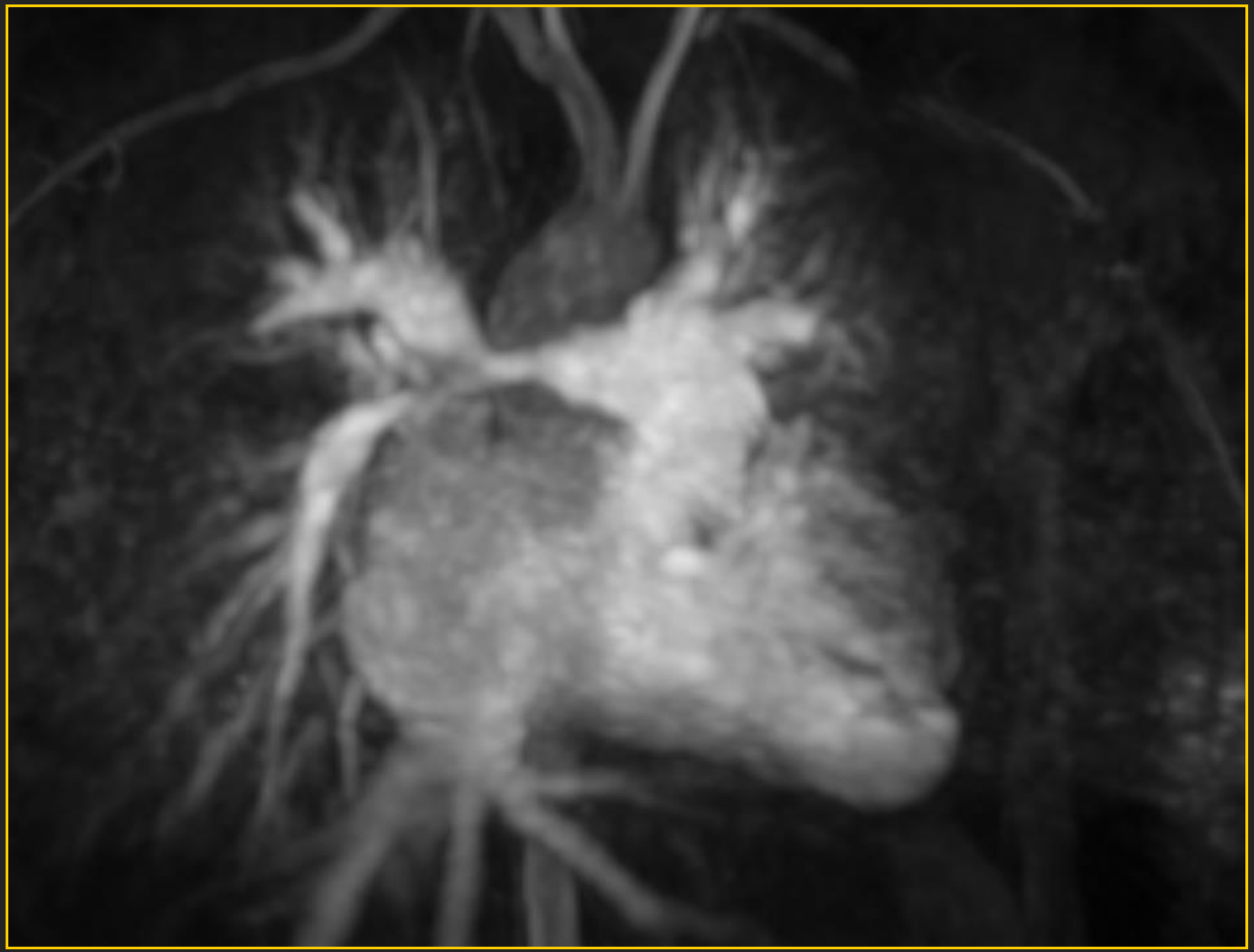
C

Ventricular Septal Defects (VSD)



Ventricular Septal Defects (VSD)





Guidelines Repaired TOF

ONASSIS CARDIAC SURGERY CENTER

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

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

Imaging surveillance frequency in repaired TOF

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