

Echocardiographic and MR investigation

What are the signs of ventricular dysfunction?

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“Frequent PVCs in structurally normal heart”

- 1. The role of imaging in exclusion of structural heart disease**
- 2. The role of imaging in functional evaluation**

“Frequent PVCs in structurally normal heart”

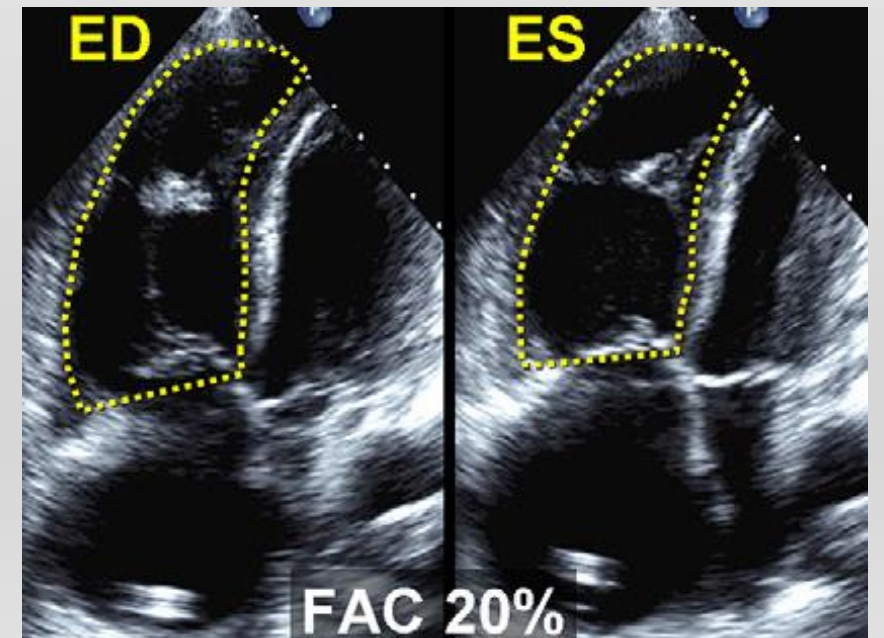
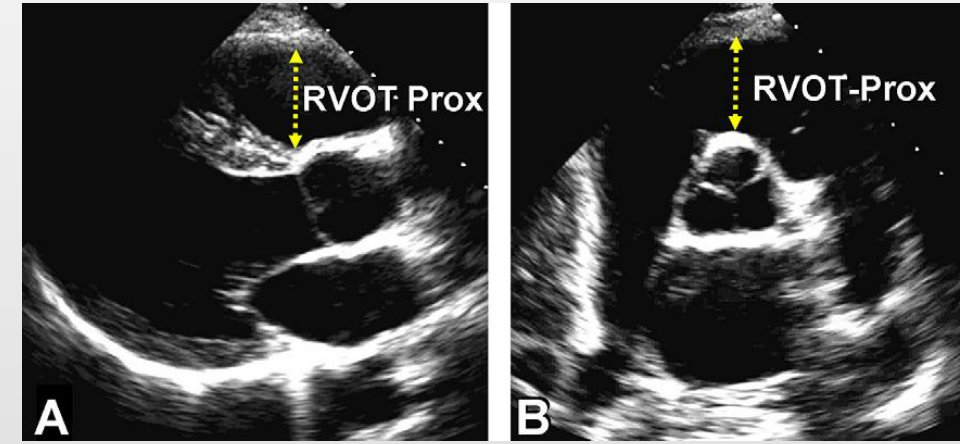
- **Imaging is very important step of evaluation**
- **First imaging tool is echocardiography**
 - **Easy achievable**
 - **Most children have good acoustic windows**
 - **Transthoracic echo generally gives us sufficient information**

Echocardiographic assessment

- **Exclusion of structural heart disease**
 - **Hemodynamically significant CHD**
 - **Cardiomyopathies**
 - **ARVC**
 - **Hypertrophic**
 - **Dilated**
 - **Noncompaction**
 - **Valvular lesions**
 - **Coronary artery abnormalities**
 - **Cardiac tumors**

Echocardiographic evaluation

- Chamber sizes, wall thicknesses
 - LVEDD
 - RVOT (PLAX, PSAX)
- Quantitation of left ventricular systolic function
 - FS %
 - EF %
- Quantitation of right ventricular systolic function
 - TAPSE
 - FAC
- Measurement of diastolic function parameters
- Regional wall motion abnormalities
 - akinesia, dyskinesia or aneurysm



Underlying cardiac pathology

Structural or not

- **Positive family history**
- **Symptoms, particularly exercise induced syncope/presyncope, chest pain**
- **Complex ventricular arrhythmias**
 - **Multiform PVC**
 - **NSVT/SVT**
 - **Polymorphic, bidirectional VT**

Myocarditis

- “Occult” myocarditis
 - there is **no demonstrable structural pathology**
 - there is **no functional abnormality by echocardiography**
- The possibility of myocarditis
 - **acutely symptomatic patients**
 - **Complex ventricular arrhythmias**
 - **Multiform PVCs**
 - **NSVT/VT**

Cardiomyopathy and Myocarditis in Children With Ventricular Ectopic Rhythm

HENRY B. WILES, MD, FACC, PAUL C. GILLETTE, MD, FACC, RUSSELL A. HARLEY, MD,
JANE K. UPSHUR, MD

Conclusions. These results provide evidence that approximately 50% of children with abnormal ventricular ectopic rhythm but a structurally normal heart may have subclinical cardiomyopathy or unsuspected myocarditis.

(J Am Coll Cardiol 1992;20:359-62)

Persistence of Ventricular Arrhythmia After Resolution of Occult Myocarditis in Children and Young Adults

RICHARD A. FRIEDMAN, MD, FACC, DEBRA L. KEARNEY, MD, JEFFREY P. MOAK, MD, FACC,
ARNOLD L. FENRICH, MD, JAMES C. PERRY, MD, FACC

Conclusions. Complex ventricular arrhythmias persist after apparent resolution of occult myocarditis in children. Although these arrhythmias are easier to control after such resolution, the patients may require long-term antiarrhythmic therapy.

(J Am Coll Cardiol 1994;24:780-3)

Cardiac MRI

- **In suspicion of**

- Myocarditis (ongoing or past)
- ARVC
- Doubtful cases



MRI should be a part of evaluation

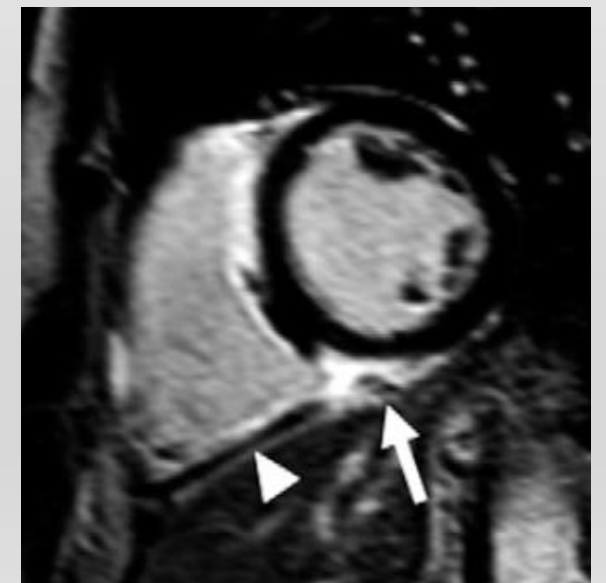
- Cardiac chamber dimensions, wall thicknesses
- Ventricular diastolic and systolic volumes
- EF of both ventricles
- Segmental wall motion abnormalities
- The presence and extent of myocardial edema/inflammation
- Areas of fibrosis (LGE)

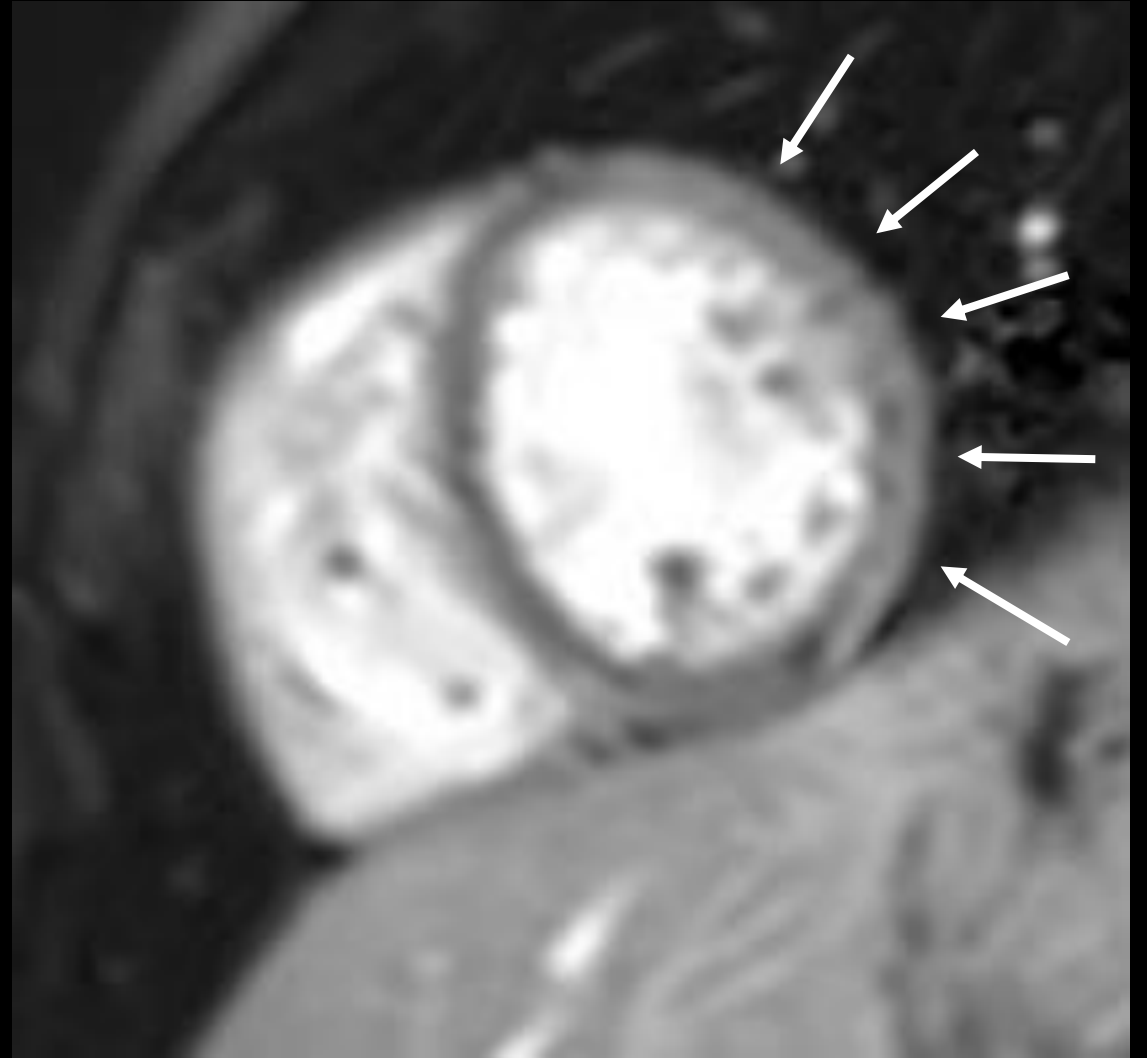
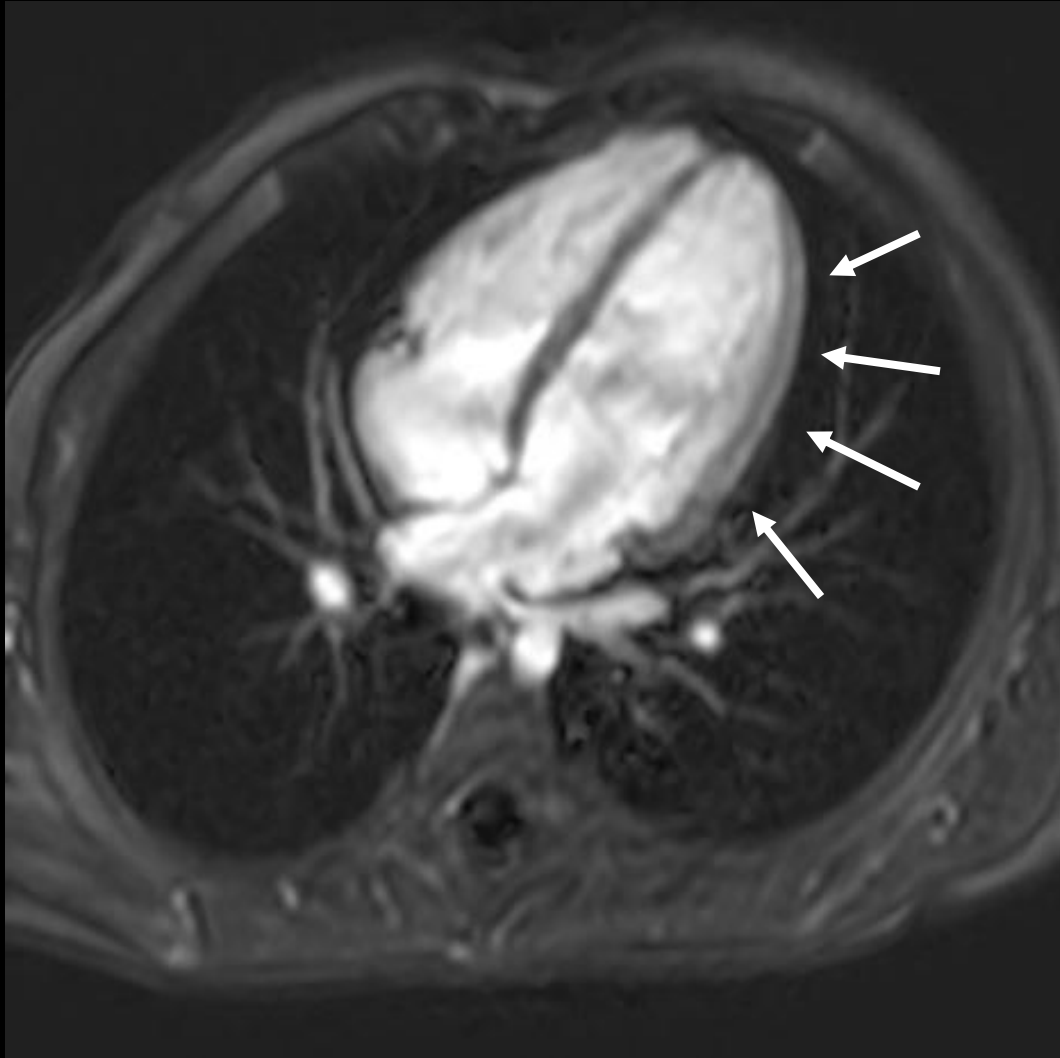
Prevalence and clinical relevance of the morphological substrate of ventricular arrhythmias in patients without known cardiac conditions detected by cardiovascular MR

¹J WEISSER-THOMAS, MD, ^{2,3}VA FERRARI, MD, ⁴A LAKGHOMI, MD, ¹LM LICKFETT, MD, ¹G NICKENIG, MD, ⁴HH SCHILD, MD and ⁴D THOMAS, MD
Br J Radiol 2014;87:

Inclusion (n=76):
- 39 male, 37 female
- premature ventricular beats > lown II (n=30)
- ventricular tachycardia (n=34)
- ventricular flutter (n=1)
- ventricular fibrillation (n=7)

Diagnosis	No. of patients (%)
Dilated cardiomyopathy	3 (3.9%)
Hypertrophic cardiomyopathy	1 (1.3%)
CMR criteria for ARVC	3 (3.9%)
Post-myocarditis scar	5 (6.6%)
Post-myocardial infarction scar	2 (2.6%)
Myocarditis	6 (7.9%)
Total	20 (26.3%)





Arrhythmogenic right ventricular cardiomyopathy ARVC

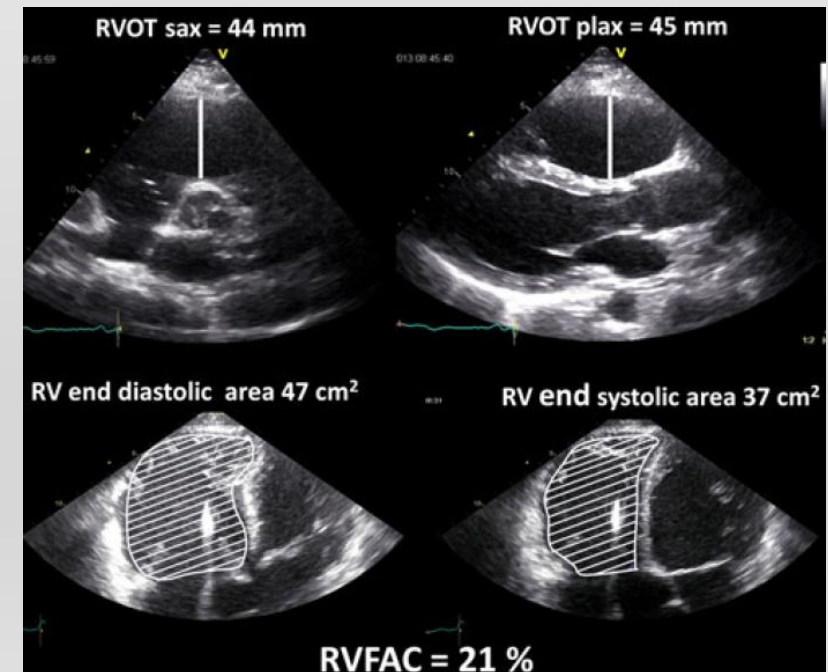
Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

Proposed Modification of the Task Force Criteria

(*Circulation*. 2010;121:1533-1541.)

By 2D echo:

- Regional RV akinesia, dyskinesia, or aneurysm
- *and* 1 of the following (end diastole):
 - PLAX RVOT ≥ 32 mm (corrected for body size [PLAX/BSA] ≥ 19 mm/m²)
 - PSAX RVOT ≥ 36 mm (corrected for body size [PSAX/BSA] ≥ 21 mm/m²)
 - *or* fractional area change $\leq 33\%$



ARVC

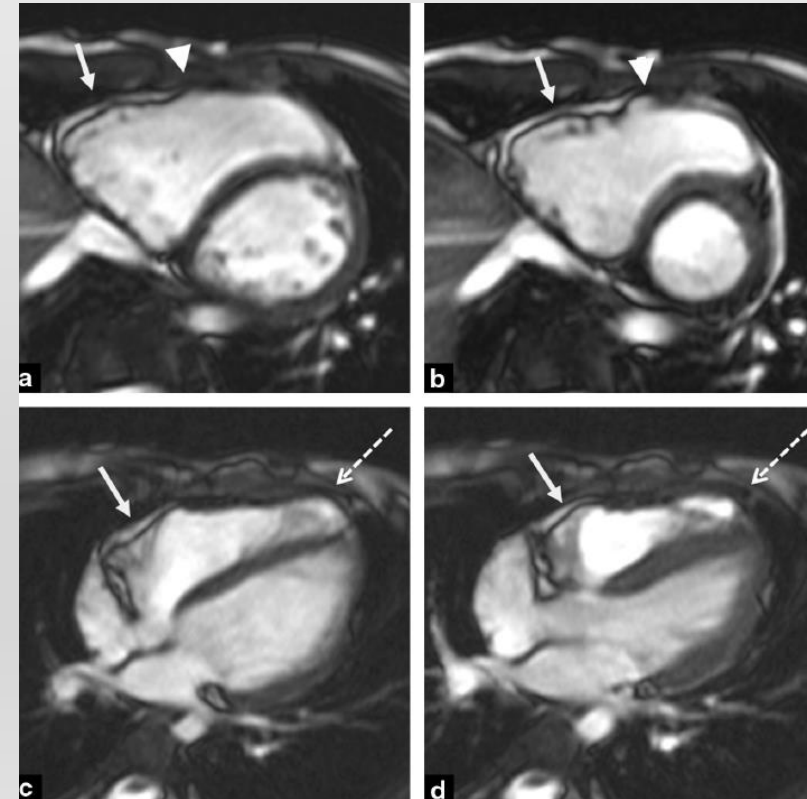
Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

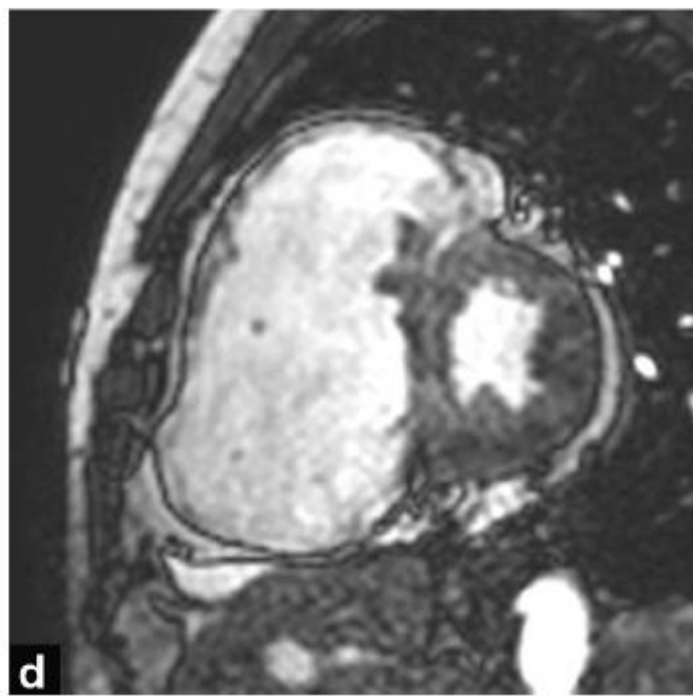
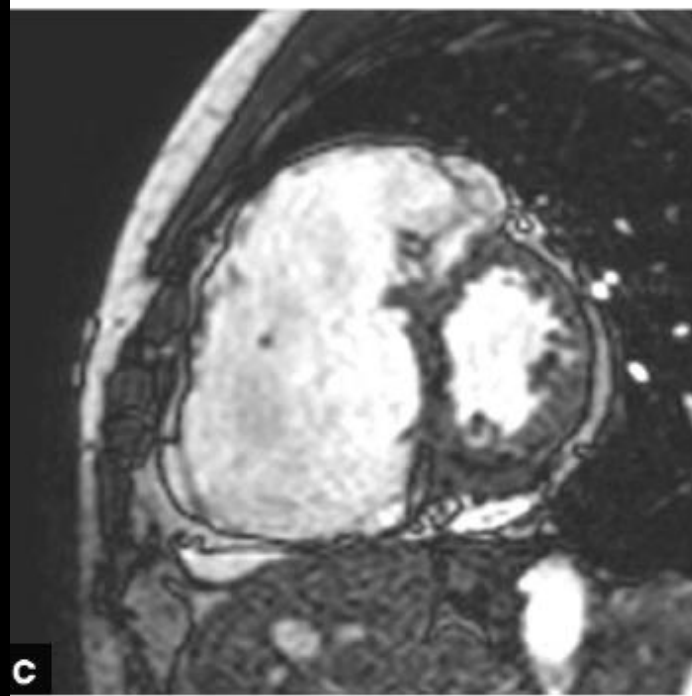
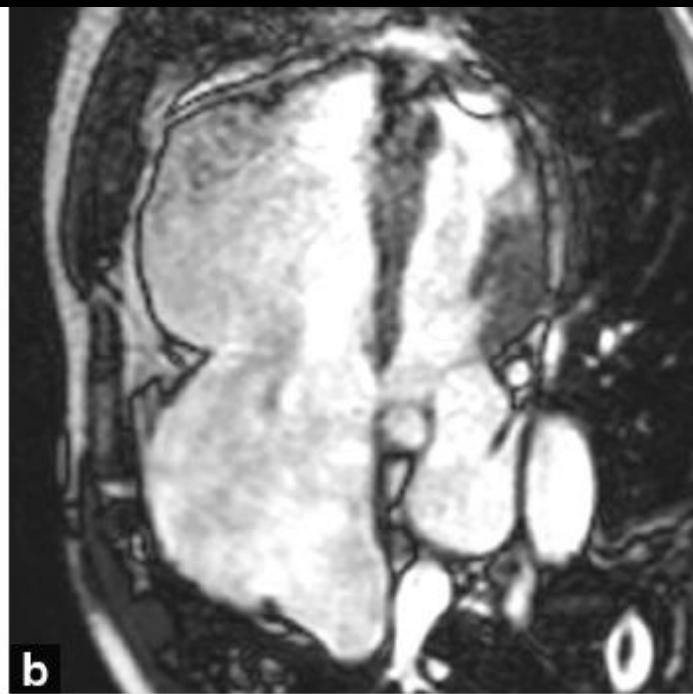
Proposed Modification of the Task Force Criteria

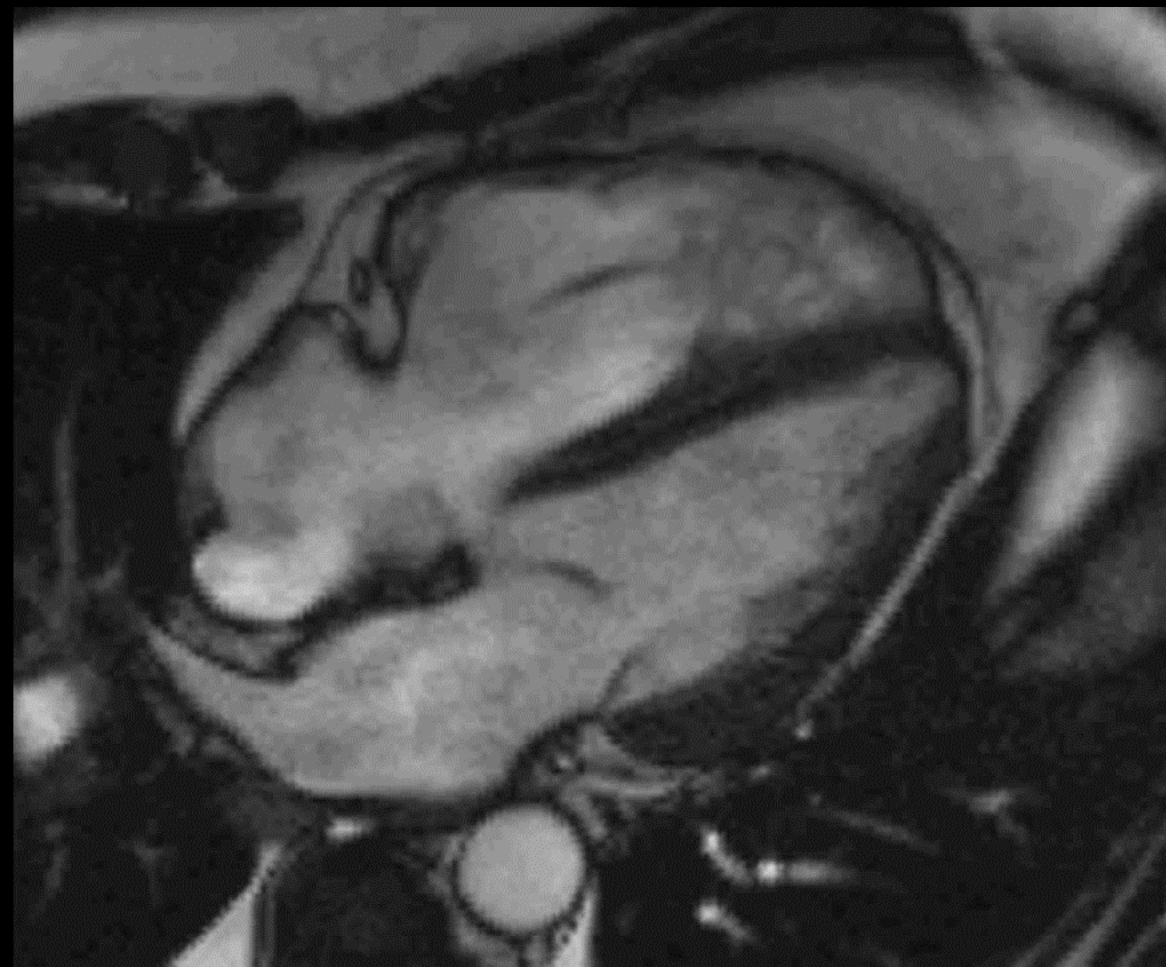
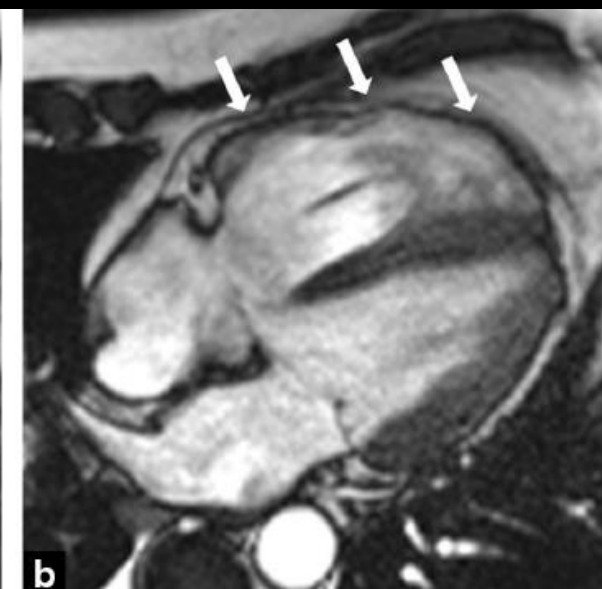
(*Circulation*. 2010;121:1533-1541.)

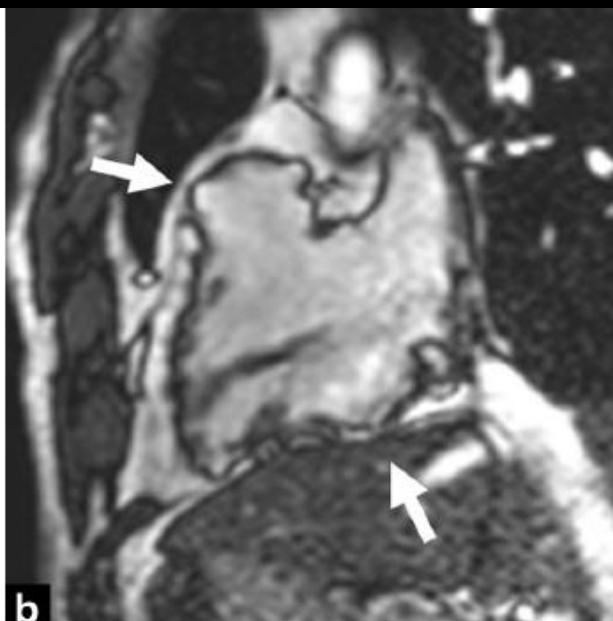
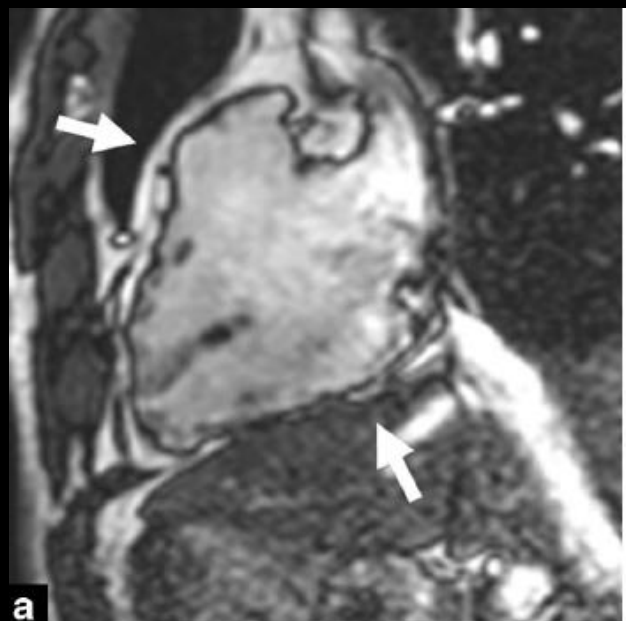
By MRI:

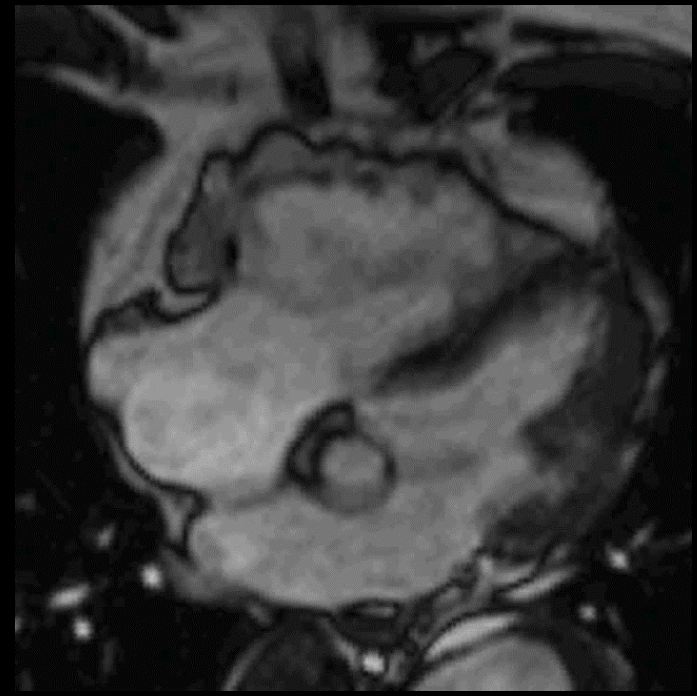
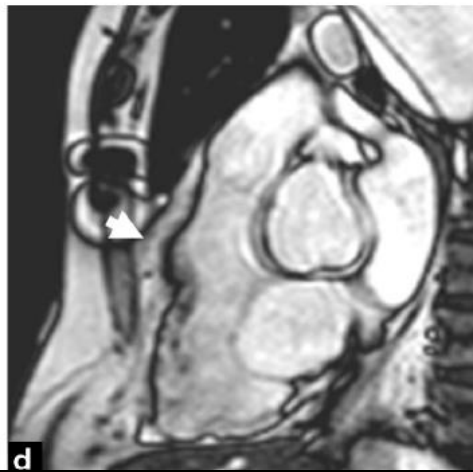
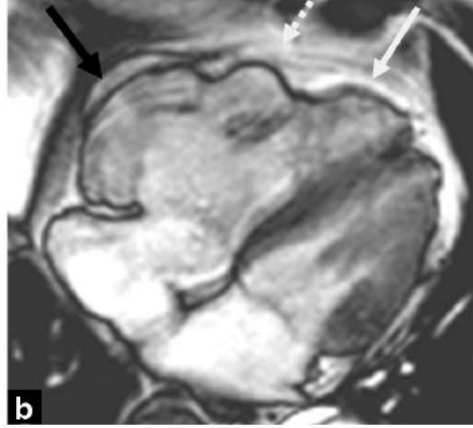
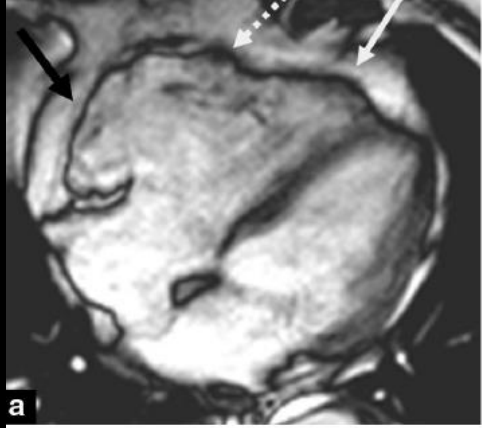
- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
 - Ratio of RV end-diastolic volume to BSA ≥ 110 mL/m² (male) or ≥ 100 mL/m² (female)
 - *or* RV ejection fraction $\leq 40\%$

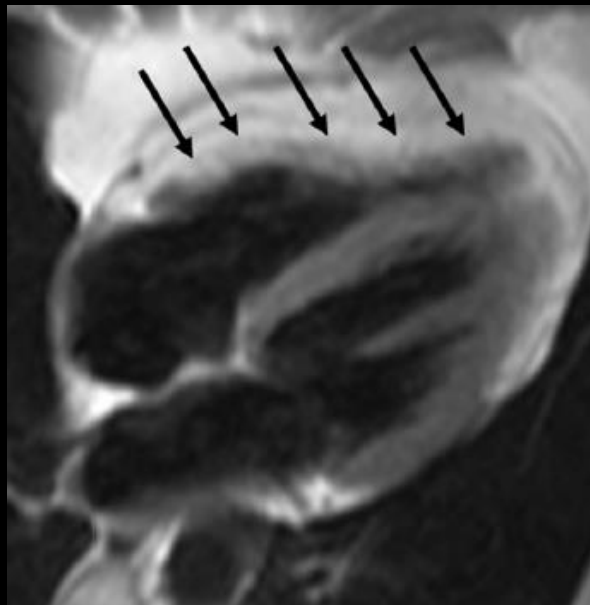
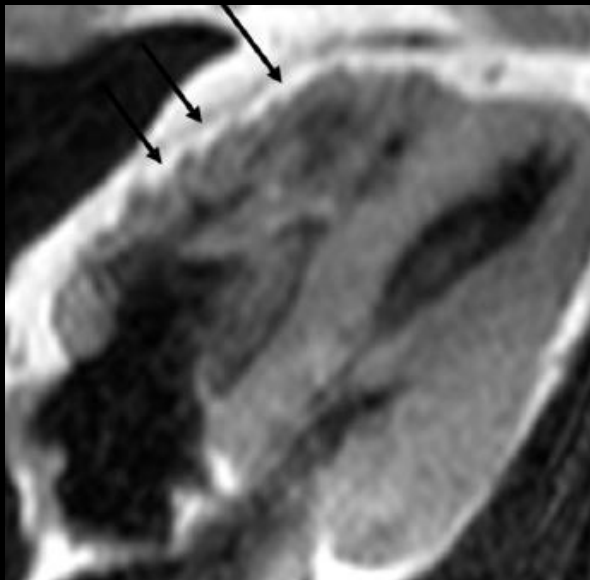




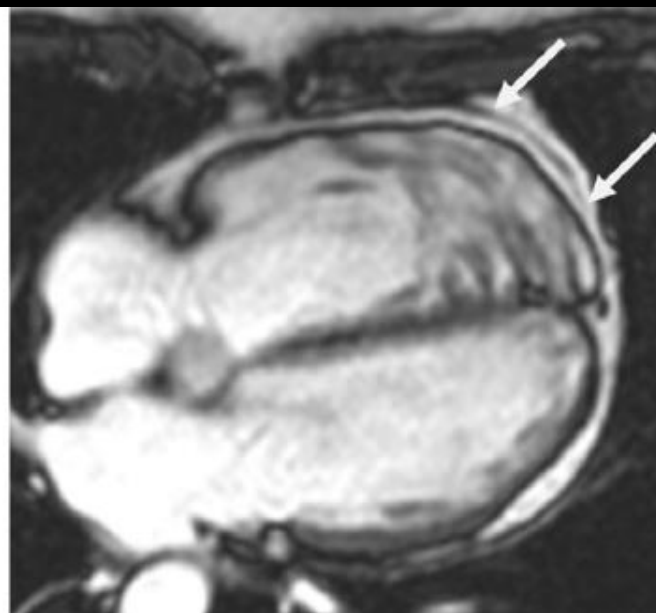
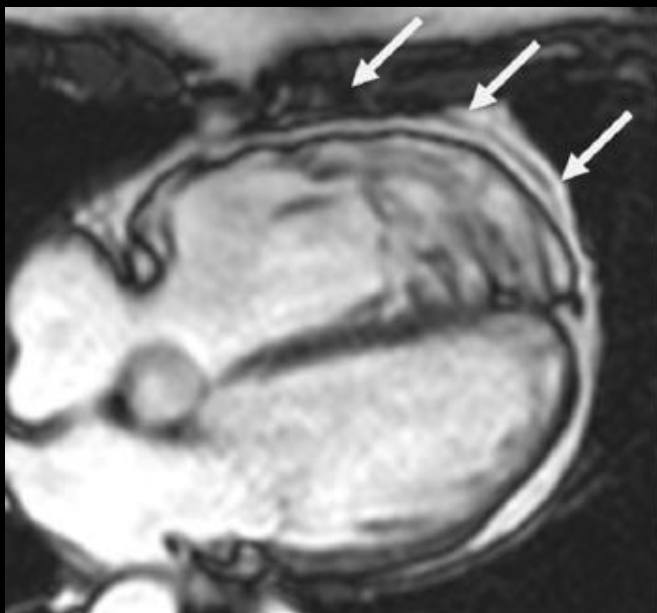




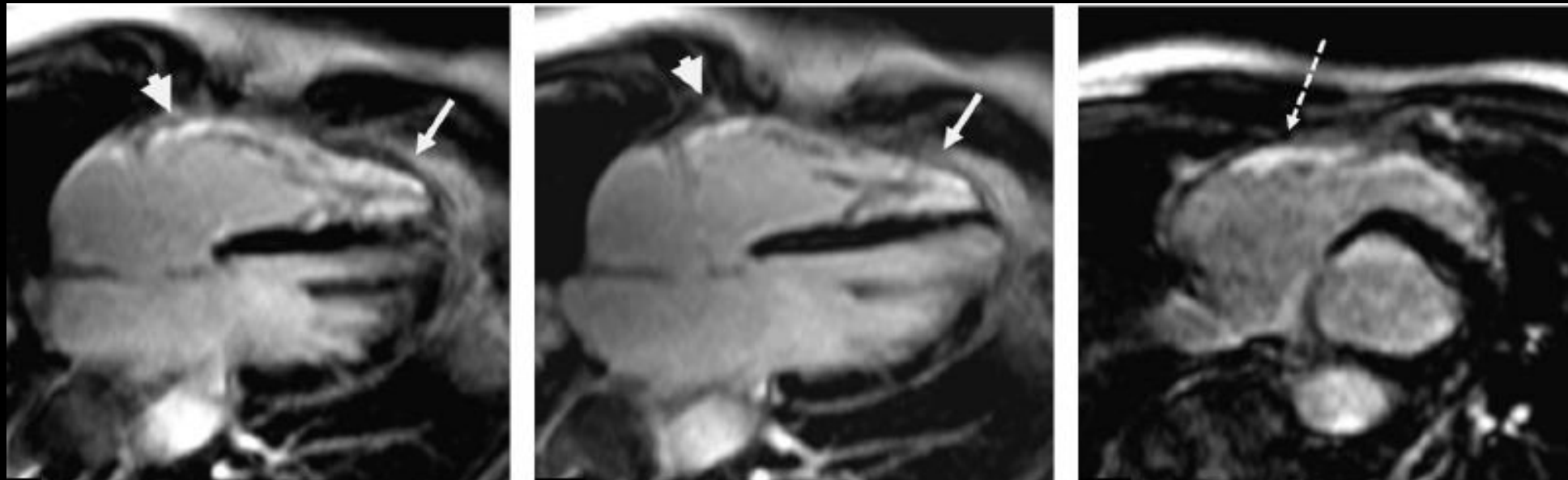




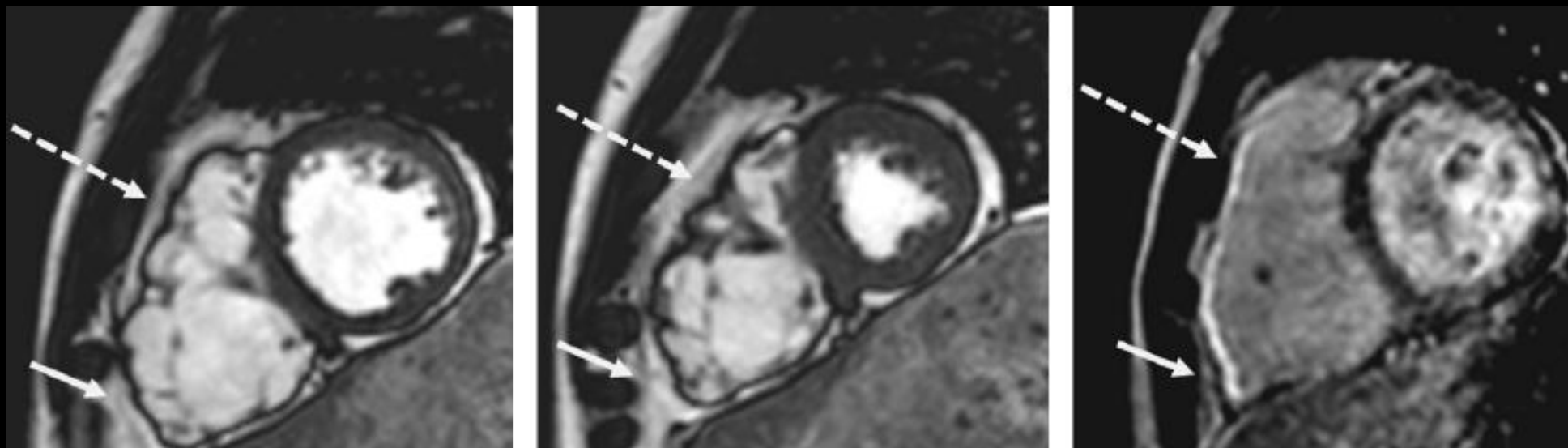
Fat infiltration



Hypertrabeculation



Late gadolinium enhancement



Functional Evaluation

Ventricular dysfunction

- Adults with **high PVC burden** can develop left ventricular dilation, dysfunction consistent with a cardiomyopathy
- It is **reversible with medical therapy or radiofrequency ablation**

Table I PVC burden associated with LV dysfunction

	<i>n</i>	%LVd	%VEs LVd	%VEs normal LV	<i>P</i>
Ban <i>et al.</i> ²¹	127 (28 LVd)	22%	31 ± 11%	22 ± 10%	0.001
Deyell <i>et al.</i> ²⁵	90 (24 LVd)	27%	32 ± 12%	27 ± 12%	0.077
Munoz <i>et al.</i> ²⁶	70 (LVd 17)	24%	29 ± 15%	17 ± 14%	0.004
Olgun <i>et al.</i> ²⁷	51 (21 LVd)	41%	30 ± 11%	14 ± 15%	0.0001
Hasdemir <i>et al.</i> ²⁸	249 (17 LVd)	7%	29 ± 9%	8 ± 7%	0.001
Baman <i>et al.</i> ²⁹	174 (57 LVd)	33%	33 ± 13%	13 ± 12%	0.0001
Kanei <i>et al.</i> ³⁰	108 (21 LVd)	19%	13 ± 11% ^a	7 ± 9% ^a	0.004

Ventricular dysfunction

Usefulness of Ventricular Premature Complexes in Asymptomatic Patients ≤ 21 Years as Predictors of Poor Left Ventricular Function

Karine Guerrier, DO, MPH*, Jeffrey B. Anderson, MD, MPH, Richard J. Czosek, MD, Wayne A. Mays, MS, Christopher Statile, MD, Timothy K. Knilans, MD, and David S. Spar, MD

Am J Cardiol 2015;115:652e655

- **22/123 patients (18%) had VPC burden $>24\%$**
- **none of them had decreased LV FS**

Ventricular dysfunction

Frequent Ventricular Premature Beats in Children With a Structurally Normal Heart: A Cause for Reversible Left Ventricular Dysfunction?

Bahram Kakavand · Hubert O. Ballard ·
Thomas G. Disessa

Pediatr Cardiol (2010) 31:986–990

- **4/28 children** developed LV systolic dysfunction and dilation
- All 4 CMP resolved, with medicine in 2, spontaneously in 2
- All had > 20% VPC burden

Ventricular dysfunction

Premature ventricular contraction-induced cardiomyopathy in children

Zebulon Z. Spector, Stephen P. Seslar

Cardiology in the Young (2016), 26, 711–717

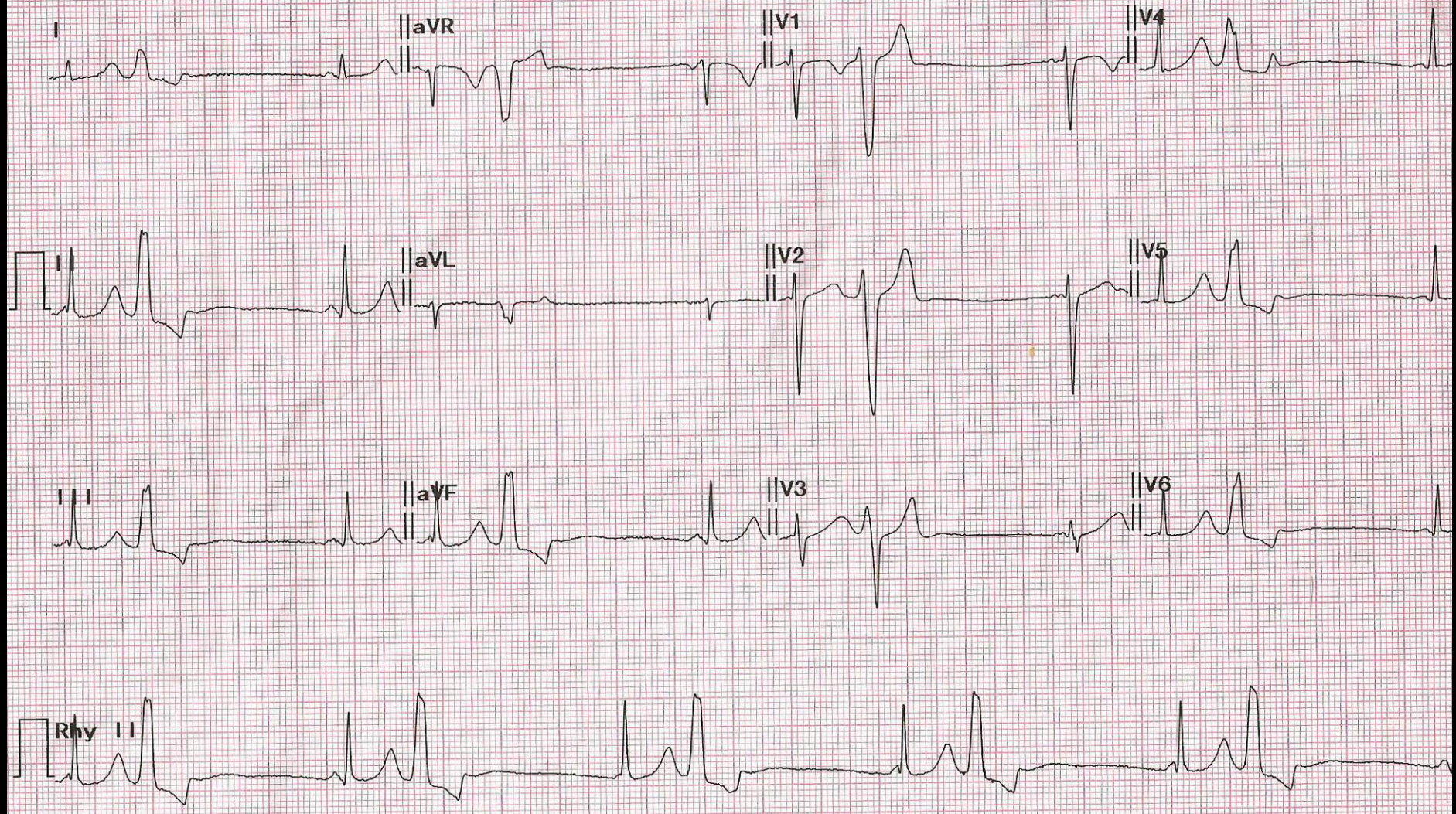
- 36 children with $\geq 20\%$ PVC burden
- 7/36 (19 %) patients with LV dysfunction ($FS \leq 28$)
- PVC burden $34,7 \pm 6,3\%$ vs $27.2 \pm 5,1\%$
- None of them symptomatic

Ventricular dysfunction

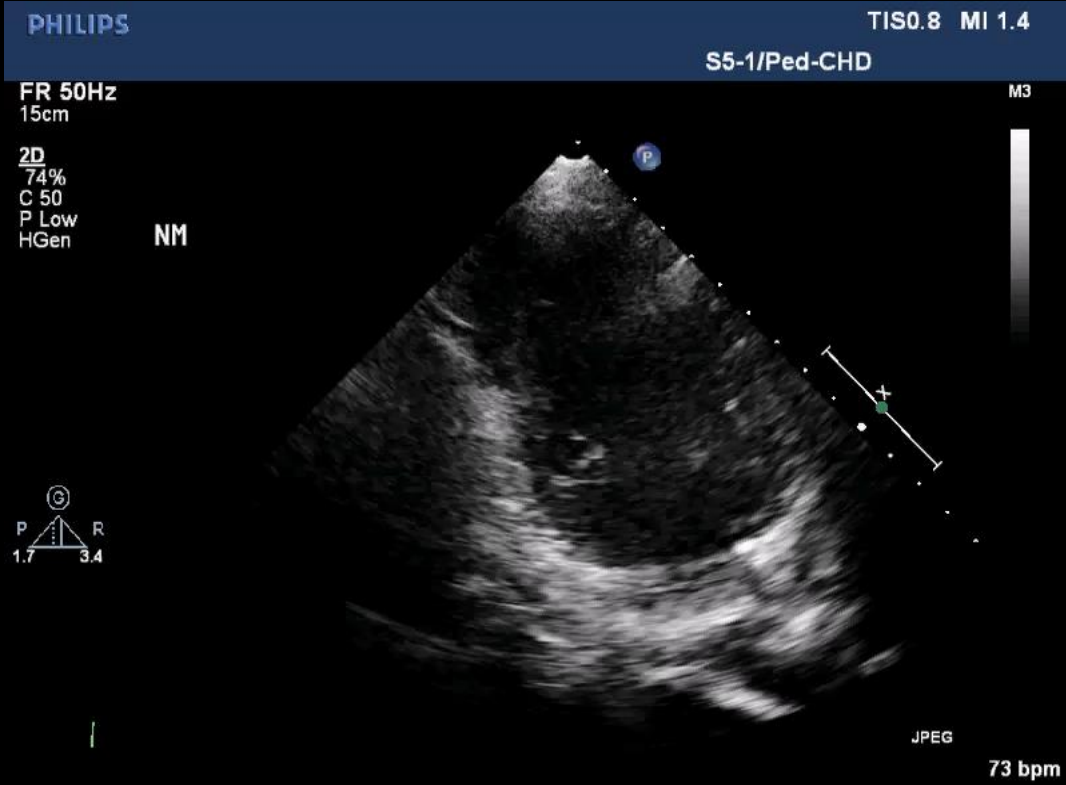
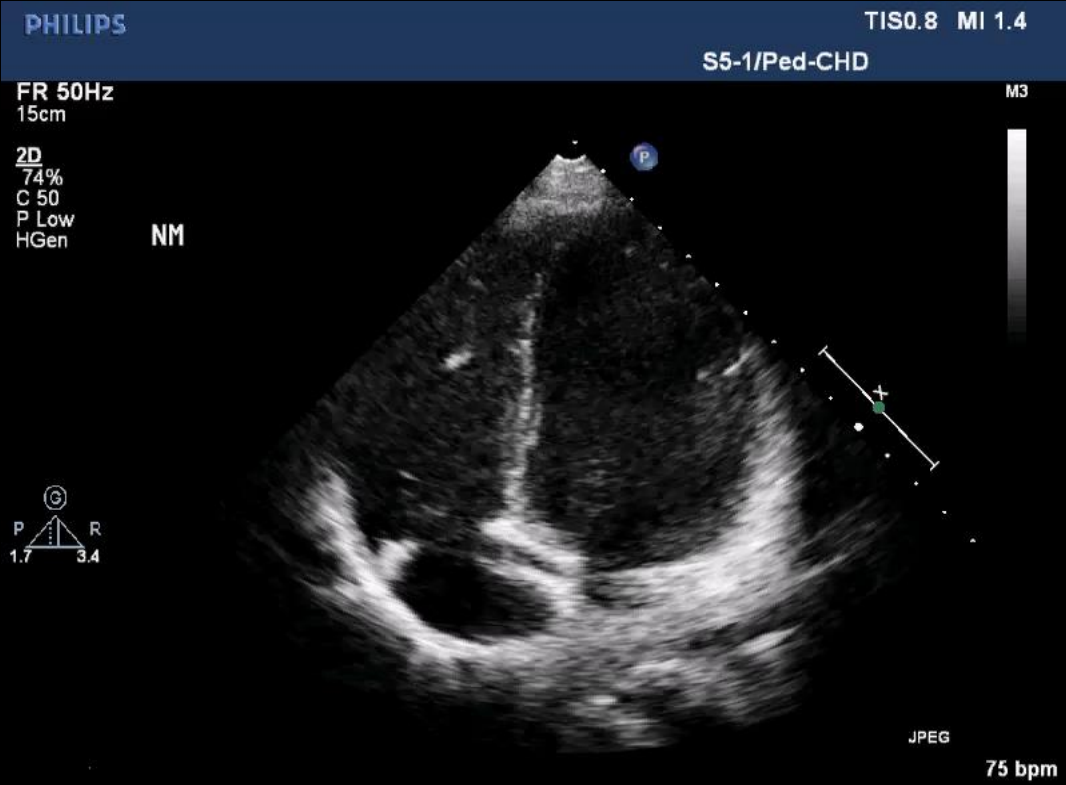
Left ventricular dysfunction is associated with frequent premature ventricular complexes and asymptomatic ventricular tachycardia in children

R.A. Bertels^{1,2*}, L.M. Harteveld^{1,2}, L.H. Filippini^{1,3}, S.A. Clur^{1,4}, and N.A. Blom^{1,2,4}

- **6/72 patients showed LV dysfunction at presentation (2 symptomatic)**
- **Patients with LV dysfunction had**
 - **higher PVC burden (47+16 vs. 16+11%)**
 - **higher prevalence of VT 5 (83%) vs. 27 (41%) and sustained ventricular tachycardia (sVT) 3 (50%) vs. 4 (6%)**
 - **higher number of couplets [6 (100%) vs. 34 (52%)]**



Difficulties in assessing functions

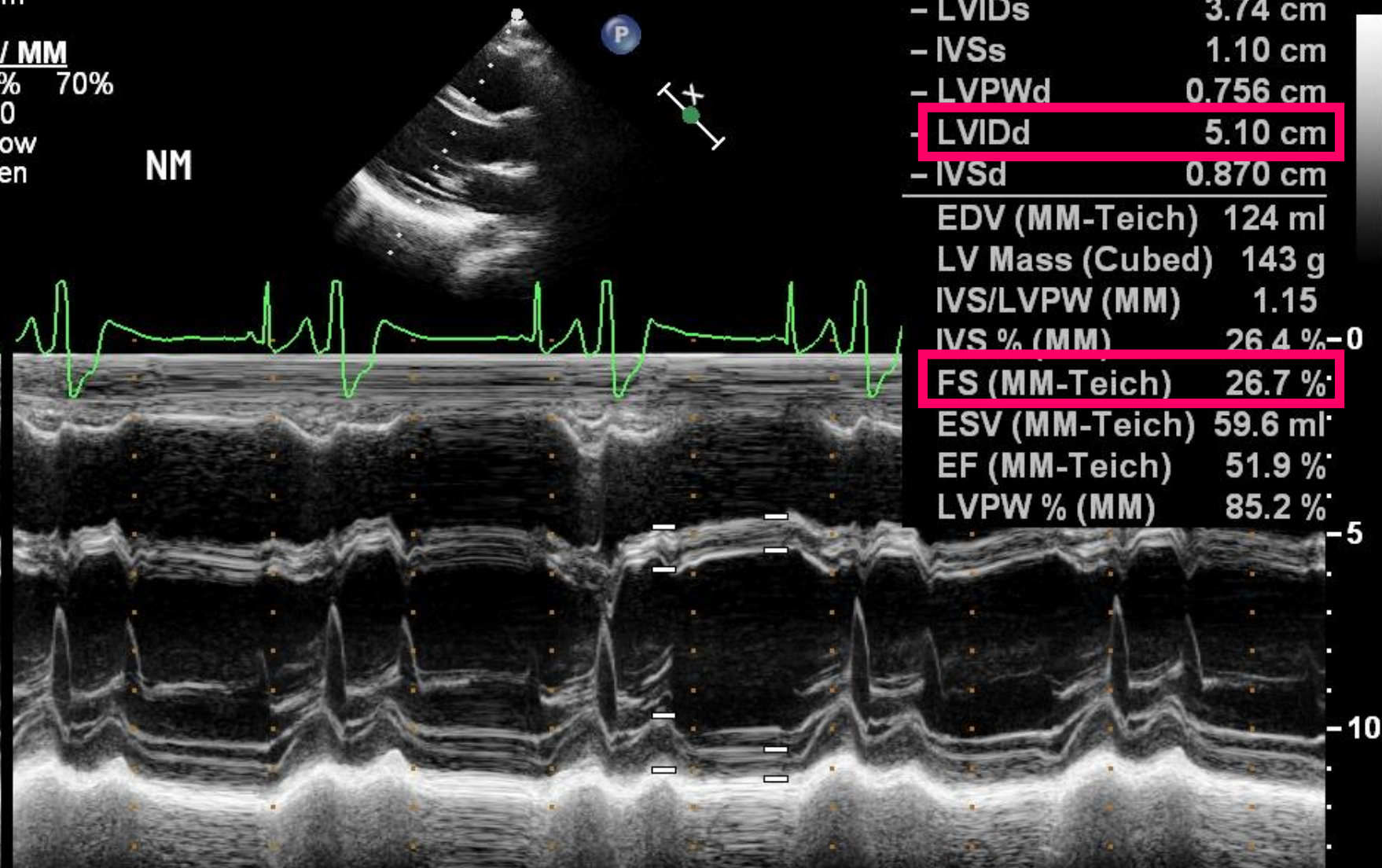


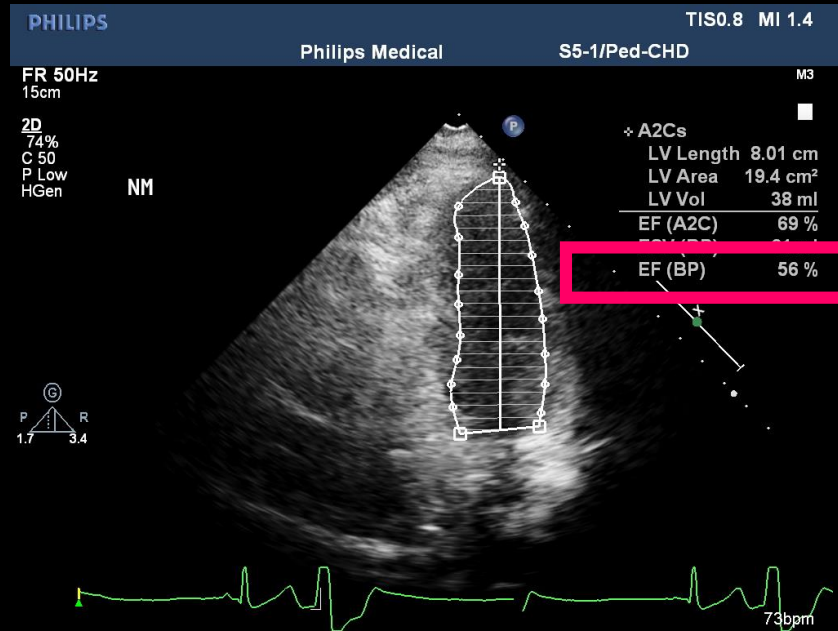
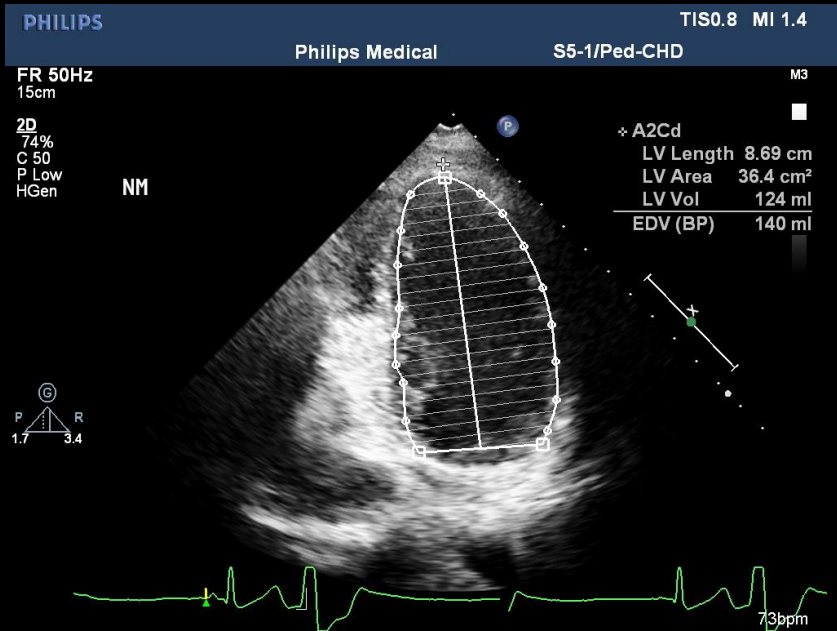
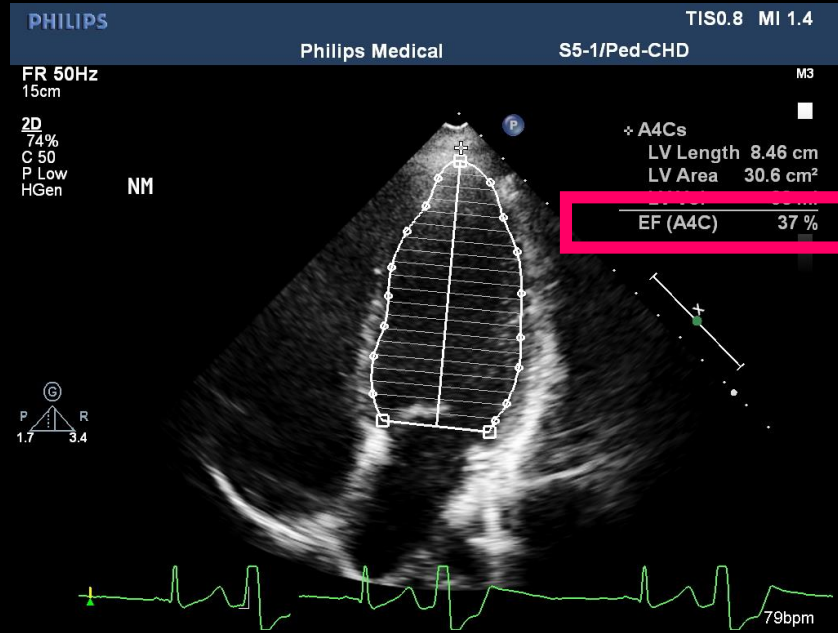
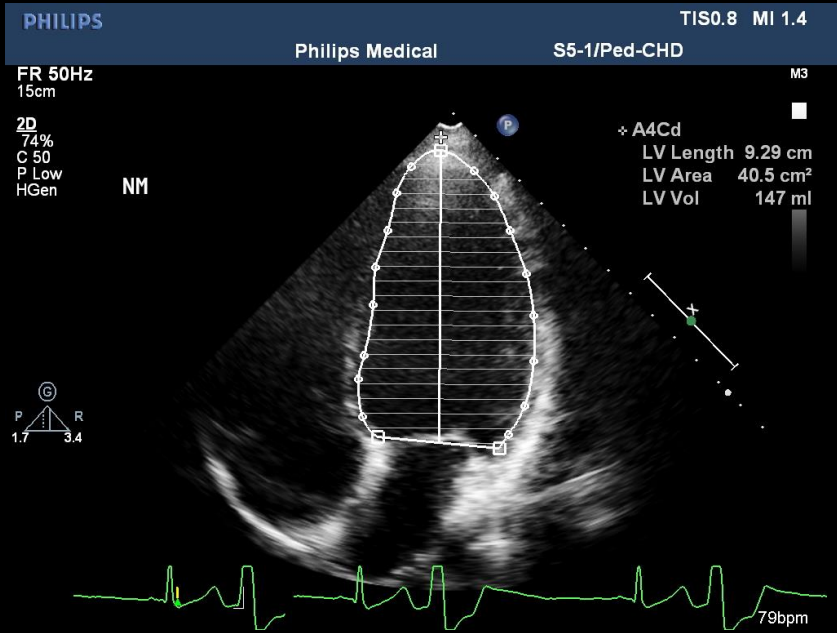
Difficulties in assessing functions

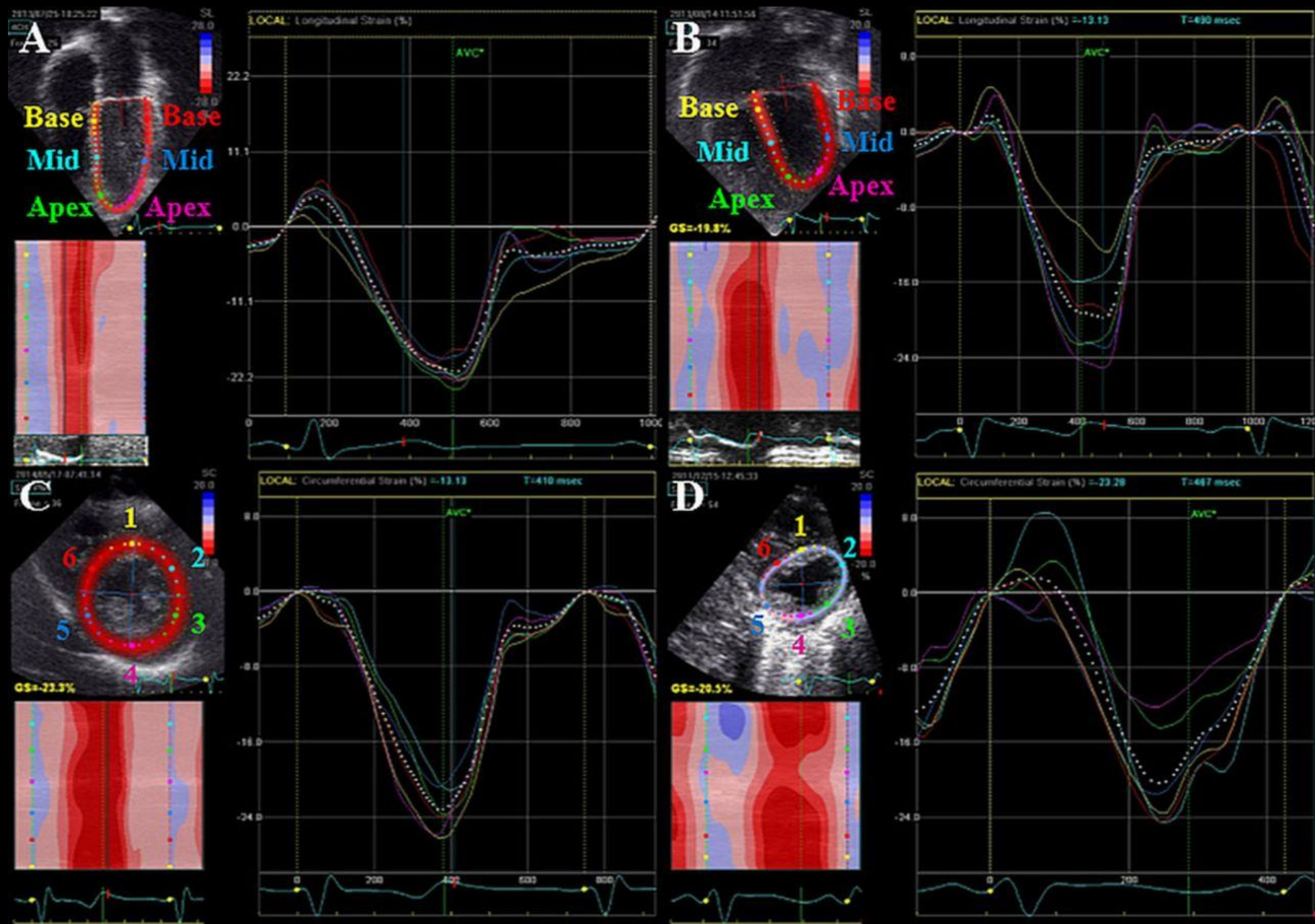
FR 25Hz
15cm

2D / MM
74% 70%
C 50
P Low
HGen

NM

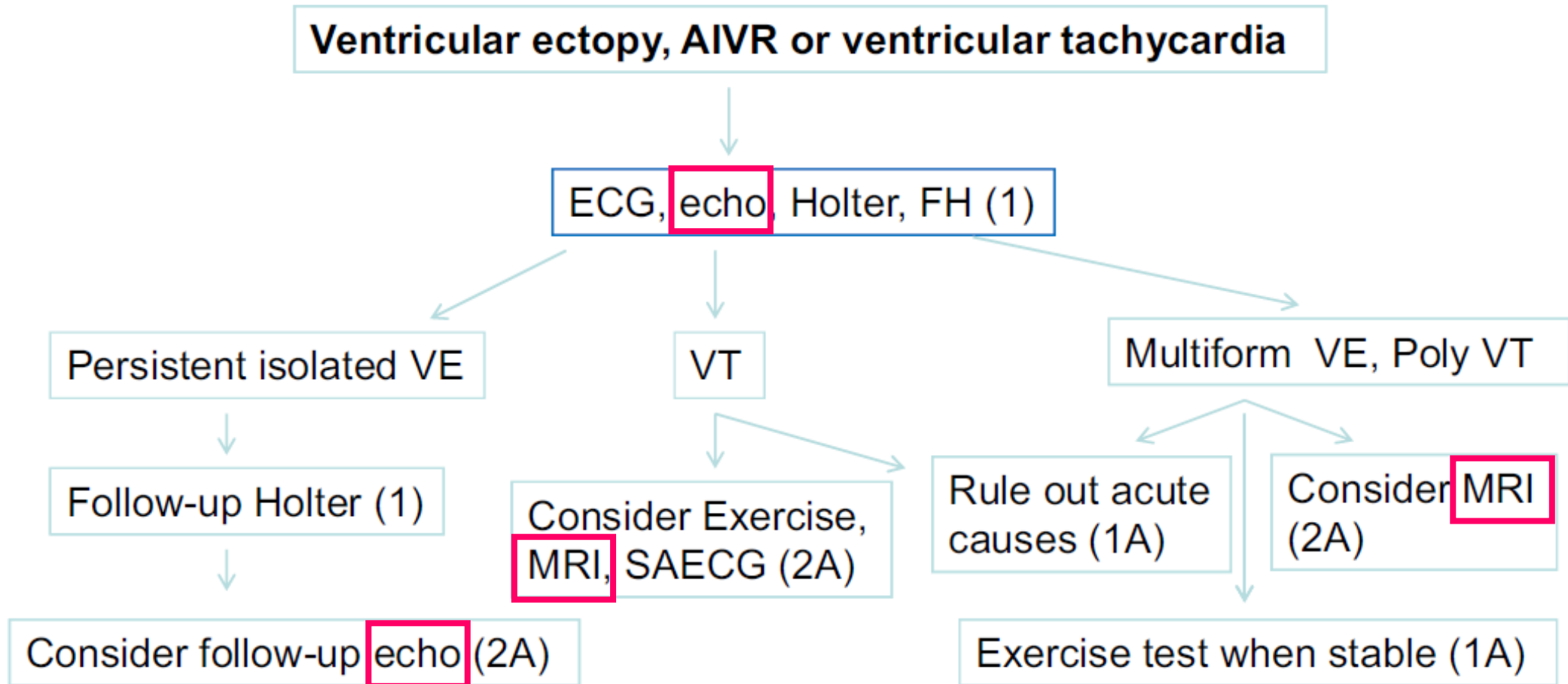






PACES/HRS Expert Consensus Statement on the Evaluation and Management of Ventricular Arrhythmias in the Child With a Structurally Normal Heart

Heart Rhythm, Vol 11, No 9,



Conclusion

- **Echocardiography is an important imaging tool**
 - to exclude structural heart disease
 - functional assessment
- **Myocarditis and ARVC** may have subtle/no signs in echo
- In doubtful cases **MRI with gadolinium** gives invaluable information
 - structure, functions and **tissue characteristics**