Pravá komora a dyssynchronie
Jak na to?

Jan Janoušek

Dětské kardiocentrum 2. LF UK a FNMH

European Reference Network
for rare or low prevalence complex diseases

Network
Heart Diseases
(ERN GUARD-HEART)

Member
Motol University
Hospital — Czechia

What are the indications for CRT?

2023 HRS/APHRS/LAHRS guideline on cardiac physiologic pacing for the avoidance and mitigation of heart failure

EF <45 %

Recommendations for CHD

COR	LOE	Recommendations	References
2a	C-LD	1. In patients with CHD on GDMT with a systemic LV, LVEF <45%, and ventricular dyssynchrony (as defined by a QRS duration z score of ≥3 or ventricular pacing ≥40%), CRT with BiV pacing is reasonable to reduce the risk of mortality or need for transplant.	400–408
2a	C-LD	2. In patients with CHD and a systemic single ventricle who require pacing, apical pacing is reasonable in preference to nonapical pacing.	409
2b	C-LD	3. In patients with CHD and a systemic single ventricle with symptomatic HF on GDMT, CRT with multisite ventricular pacing may be considered to maintain functional class or ventricular function.	400,402,410,411
2b	C-LD	4. In patients with CHD and a systemic RV with symptomatic HF on GDMT associated with ventricular electrical delay or requiring substantial ventricular pacing, CRT with BiV pacing may be considered to improve or maintain functional class or ventricular function.	400–408,412–415
2b	C-LD	5. In patients with CHD and a subpulmonary RV with RV dysfunction and RBBB, CRT with fusion-based pacing may be considered to improve RV function.	416–418
2b	C-LD	6. In patients with CCTGA and AV block in whom anatomic repair has not been performed, CSP with HBP or LBBAP may be considered to improve functional status.	419,420

Post-repair tetralogy of Fallot

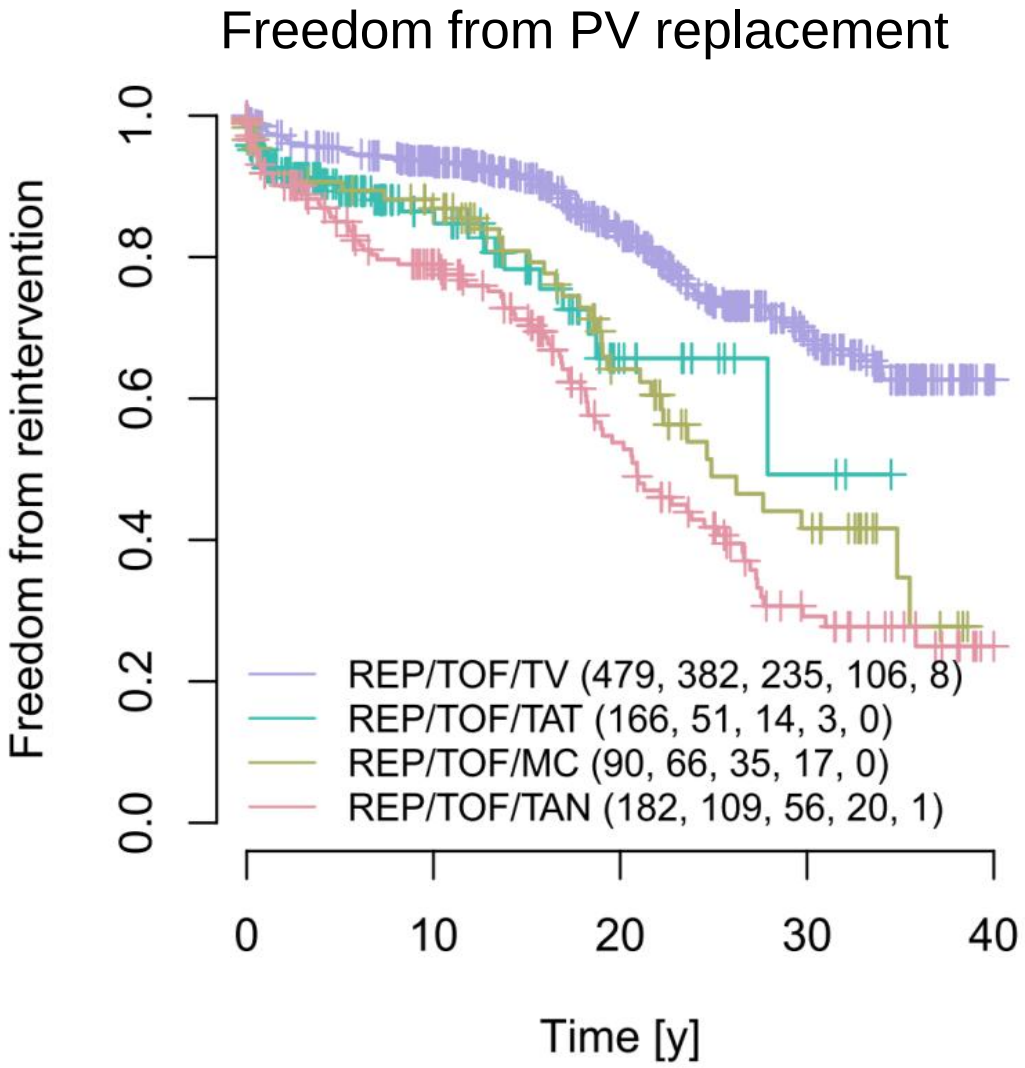
Chronic RV failure model
+
right bundle branch block

Journal of the American Heart Association

ORIGINAL RESEARCH

Survival and Freedom From Reinterventions in Patients With Repaired Tetralogy of Fallot: Up to 42-Year Follow-Up of 917 Patients

Roman Gebauer, MD; Václav Chaloupecký, MD, PhD; Bohumil Hučín, MD, DSc; Tomáš Tláskal, MD, PhD; Arnošt Komárek, PhD; Jan Janoušek, MD, PhD



Post-repair tetralogy of Fallot

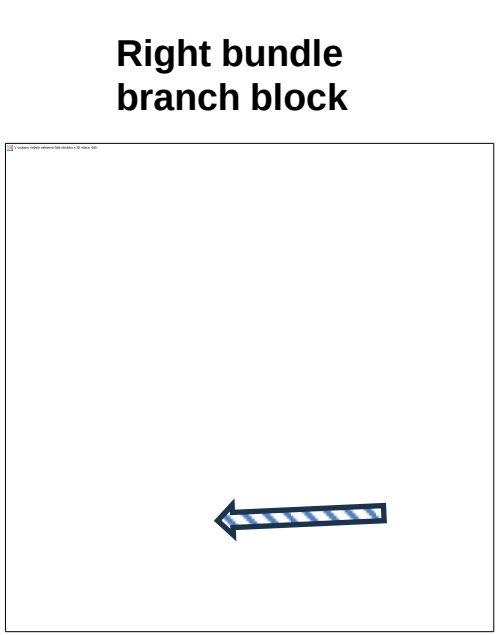
- Decreased probability of RV reverse remodeling after pulmonary valve replacement
  - RVEDV >150 to 170 mL/m2 or RVESV > 82 to 90 mL/m2
  - RV EF ≤45%
  - **QRS ≥160 ms!**
- PVR alone may not lead to RV myocardial performance normalization
  - Myocardial fibrosis/scar?
  - Dyssynchronopathy?
    - RBBB is frequent

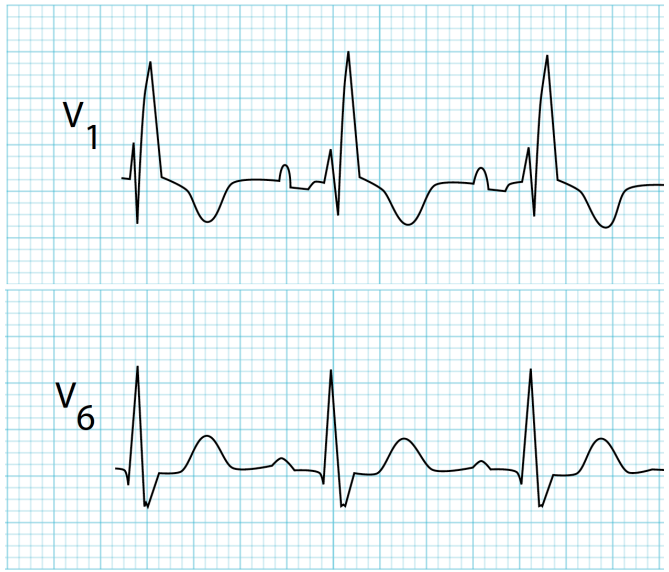
Therrien J, Am J Cardiol 2005
Oosterhof T, Circulation 2007
Henkens IR, Ann Thorac Surg 2007
Baumgartner H et al. EHJ 2010
Kutty S et al. J Am Soc Echocardiogr 2008
Geva T et al. Circulation 2010

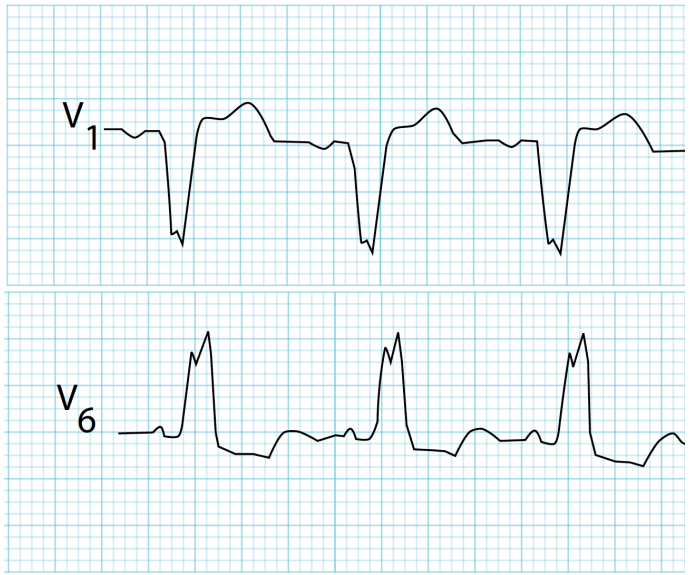
Electrical activation delay as cause of clustered dyssynchrony

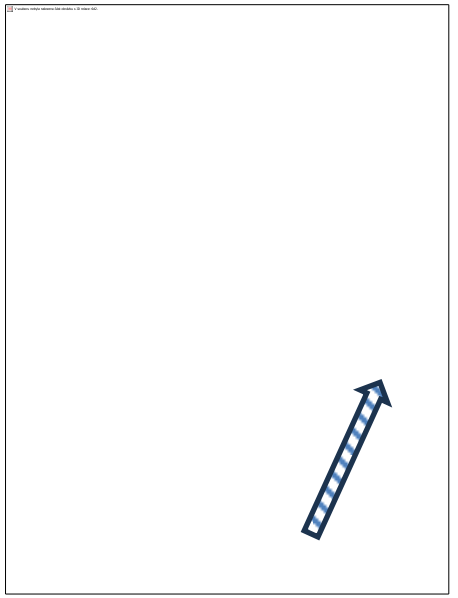
- Is there a conduction delay within the failing ventricle?
- If so, think about CRT!

Right bundle branch block





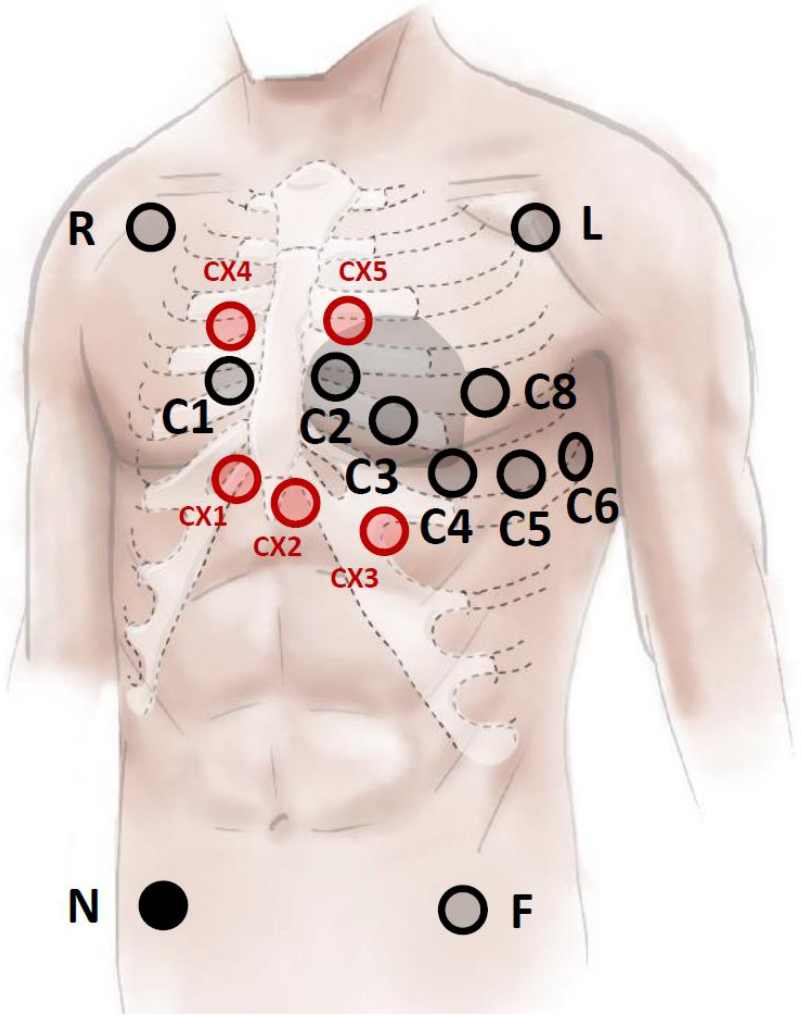


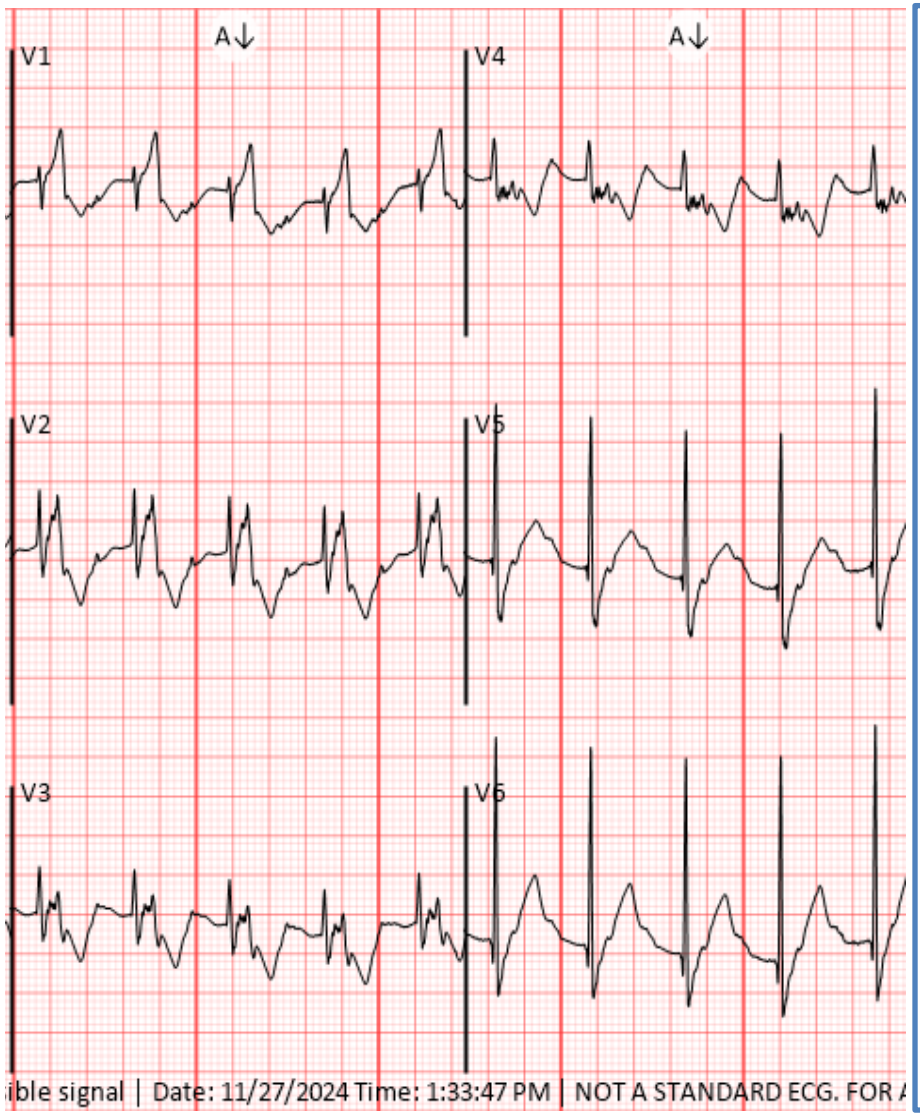


Electrical imaging in dyssynchrony

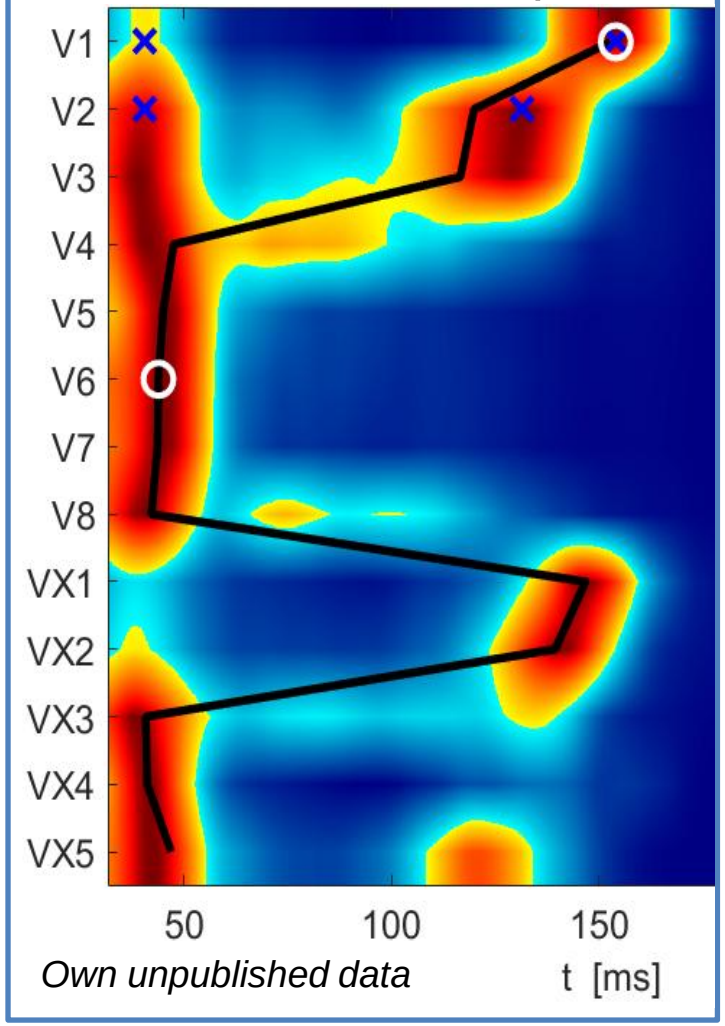
Ultra-high frequency ECG

UHF-ECG lead positions





UHF ECG map

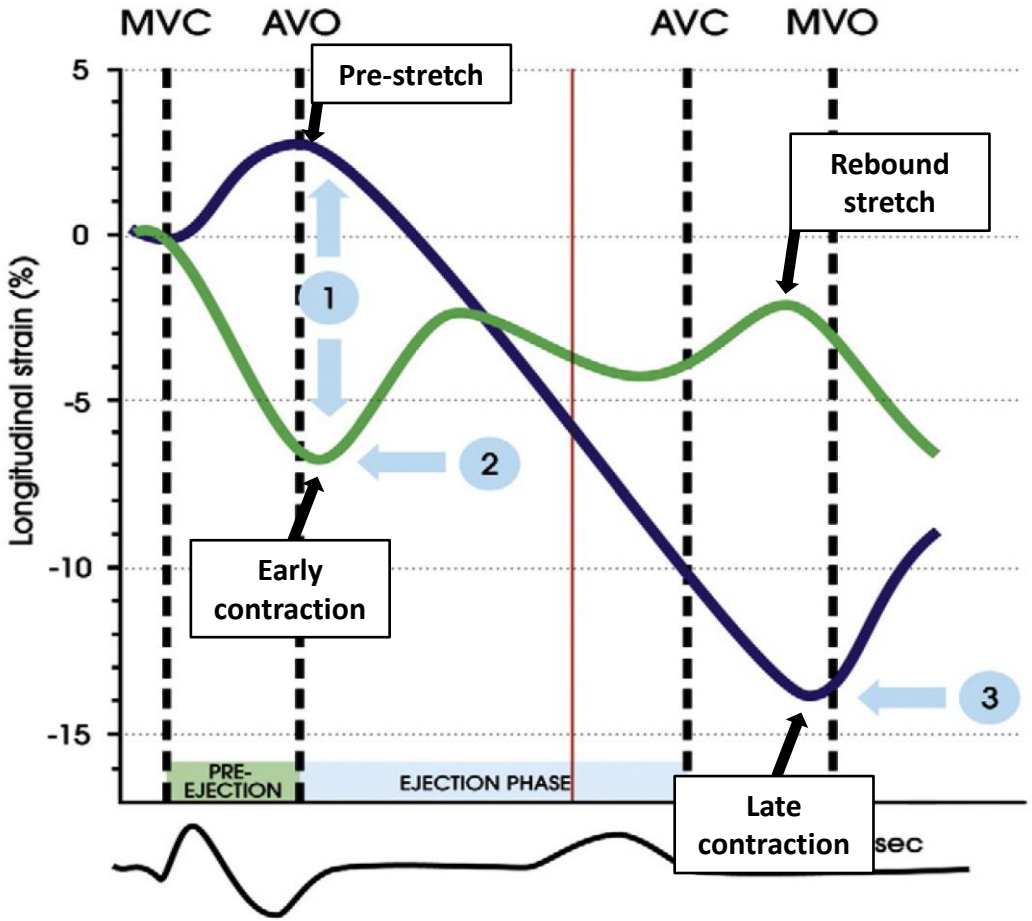


Own unpublished data

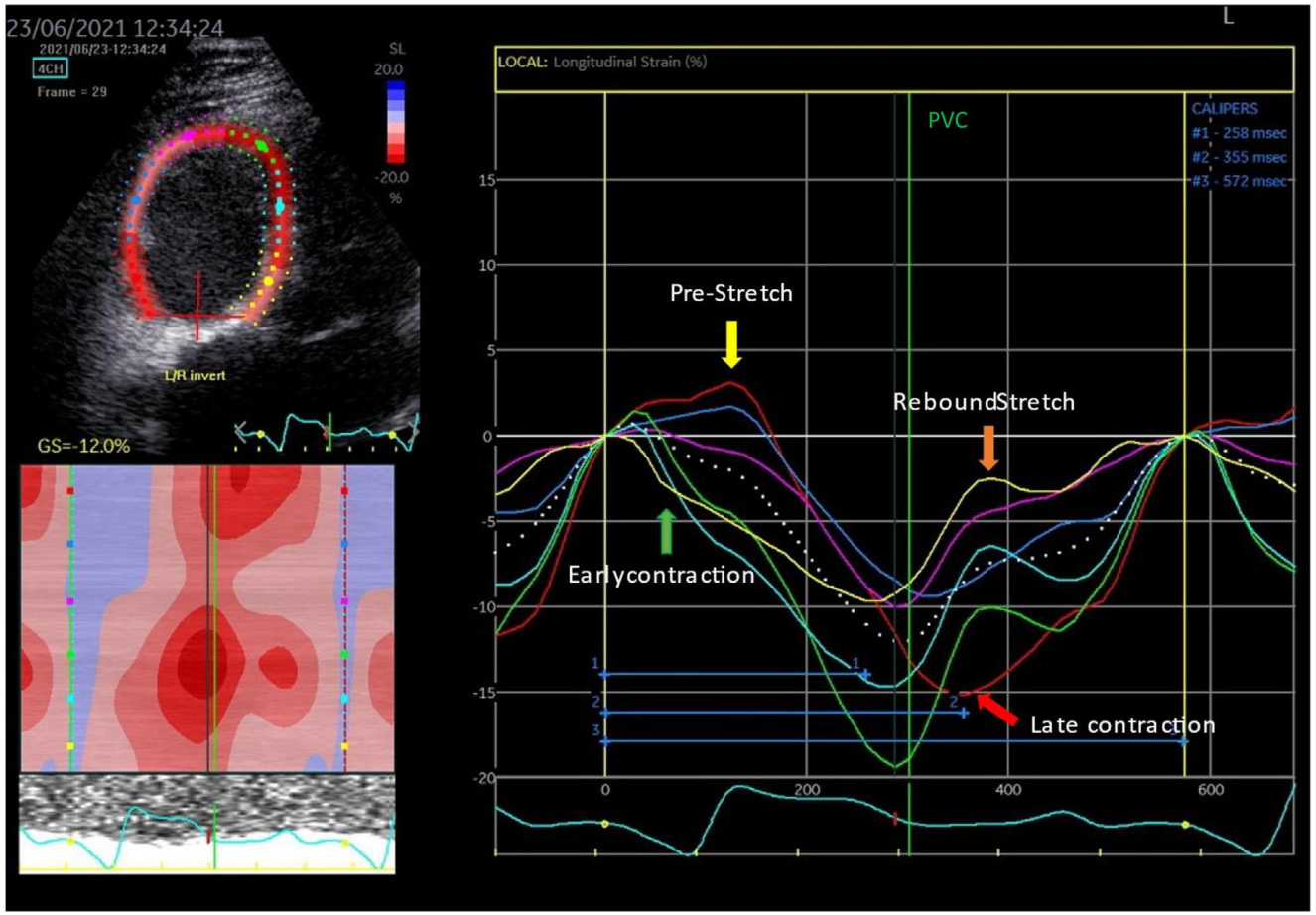
Mechanical imaging in dyssynchrony

Speckle tracking echocardiography

Classic-pattern dyssynchrony



Speckle tracking: RBBB



Risum N, Am Heart J 2012

Quantification of Right Ventricular Electromechanical Dyssynchrony in Relation to Right Ventricular Function and Clinical Outcomes in Children with Repaired Tetralogy of Fallot

Journal of the American Society of Echocardiography
Volume 31 Number 7

Deane Yim, MD, Wei Hui, MD, Guillermo Larios, MD, Andrea Dragulescu, MD, Lars Grosse-Wortmann, MD, Bart Bijnens, PhD, Luc Mertens, MD, PhD, and Mark K. Friedberg, MD, *Toronto, Ontario, Canada; and Barcelona, Spain*

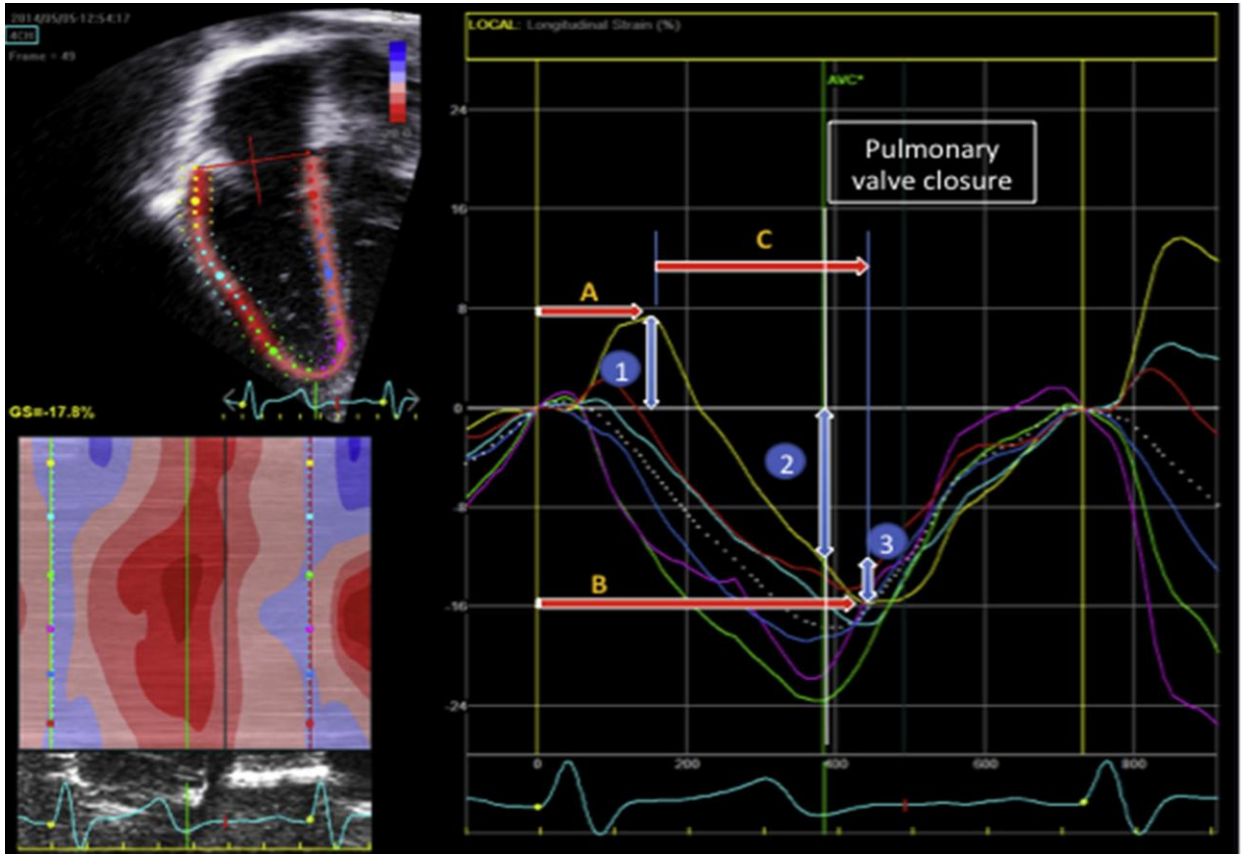
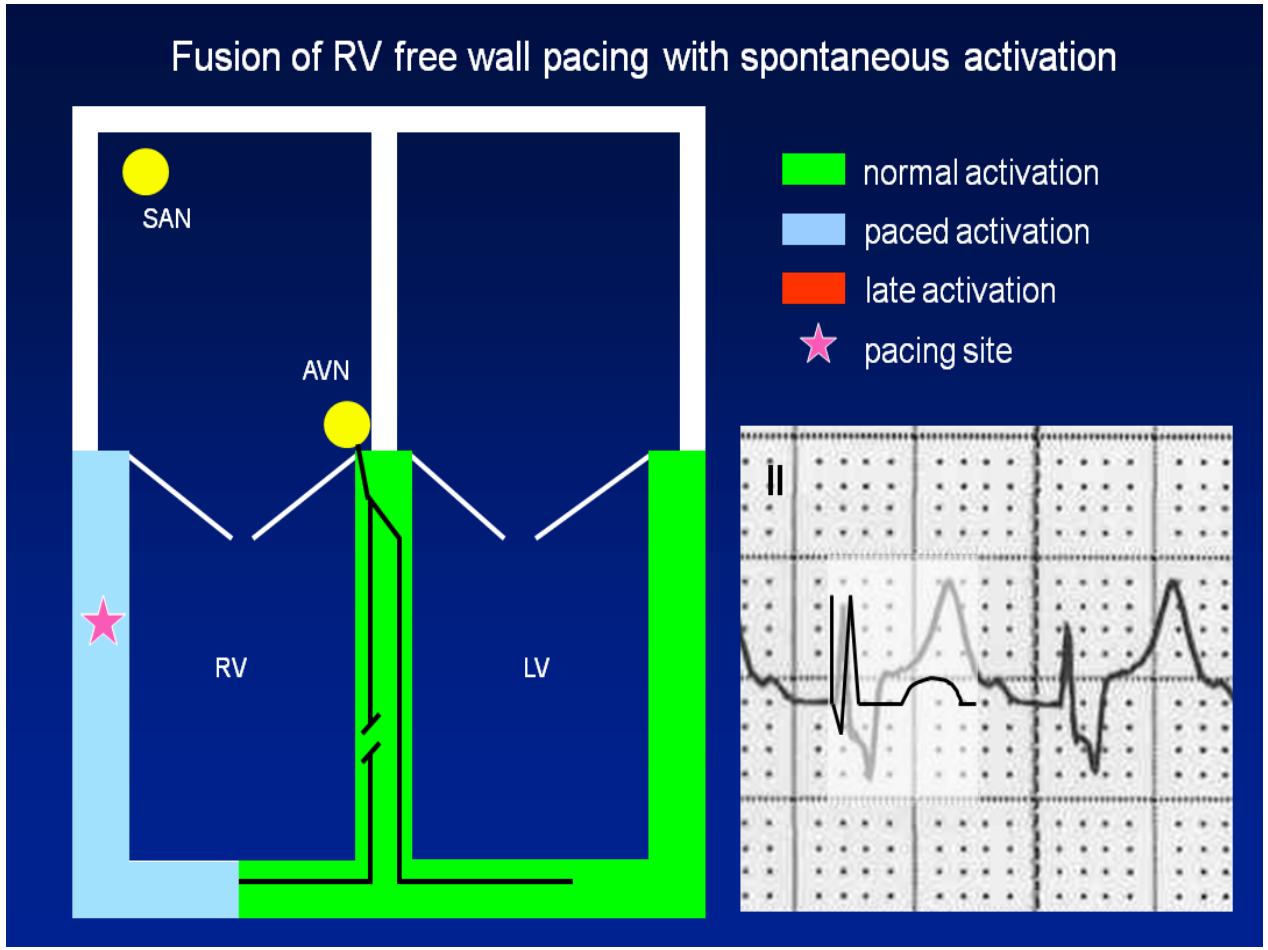


Table 4 Relationship of RV dyssynchrony parameters and function

Variable	RV remodeling	RVEF* and/or strain†
Prestretch amplitude	+	+
Prestretch amplitude %	+	+
Prestretch duration	–	++
Prestretch duration %	–	+
PSS amplitude	–	+
PSS amplitude %	–	+
PSS duration	–	+
PSS duration %	–	+
Mechanical dispersion index	+	+
RV-septal delay	+	++

Pulmonary RV CRT

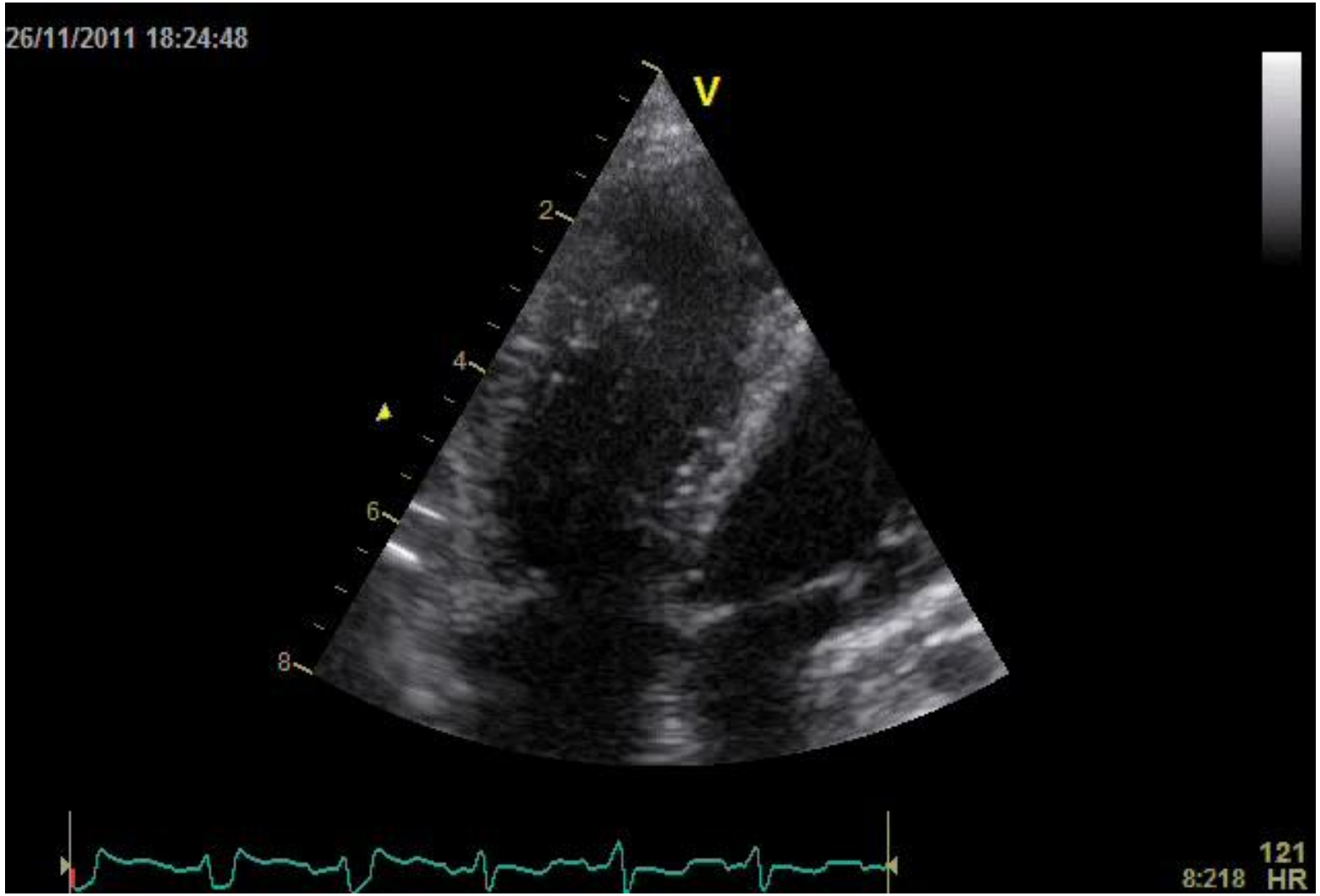
Acute hemodynamic study



- N=7
- RV dysfunction and RBBB
- Results
  - ↓ QRS duration
  - ↑ cardiac index
  - ↑ RV dP/dt

Dubin AM et al. Circulation 2003

Mechanical effects of pulmonary RV-CRT

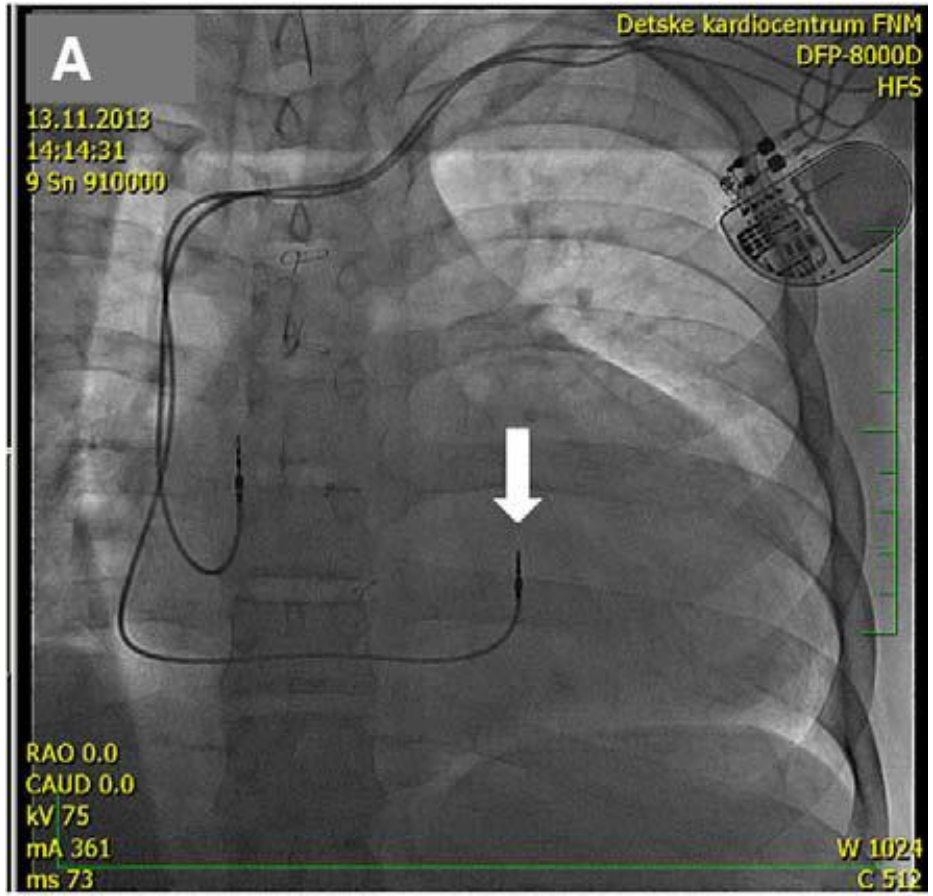


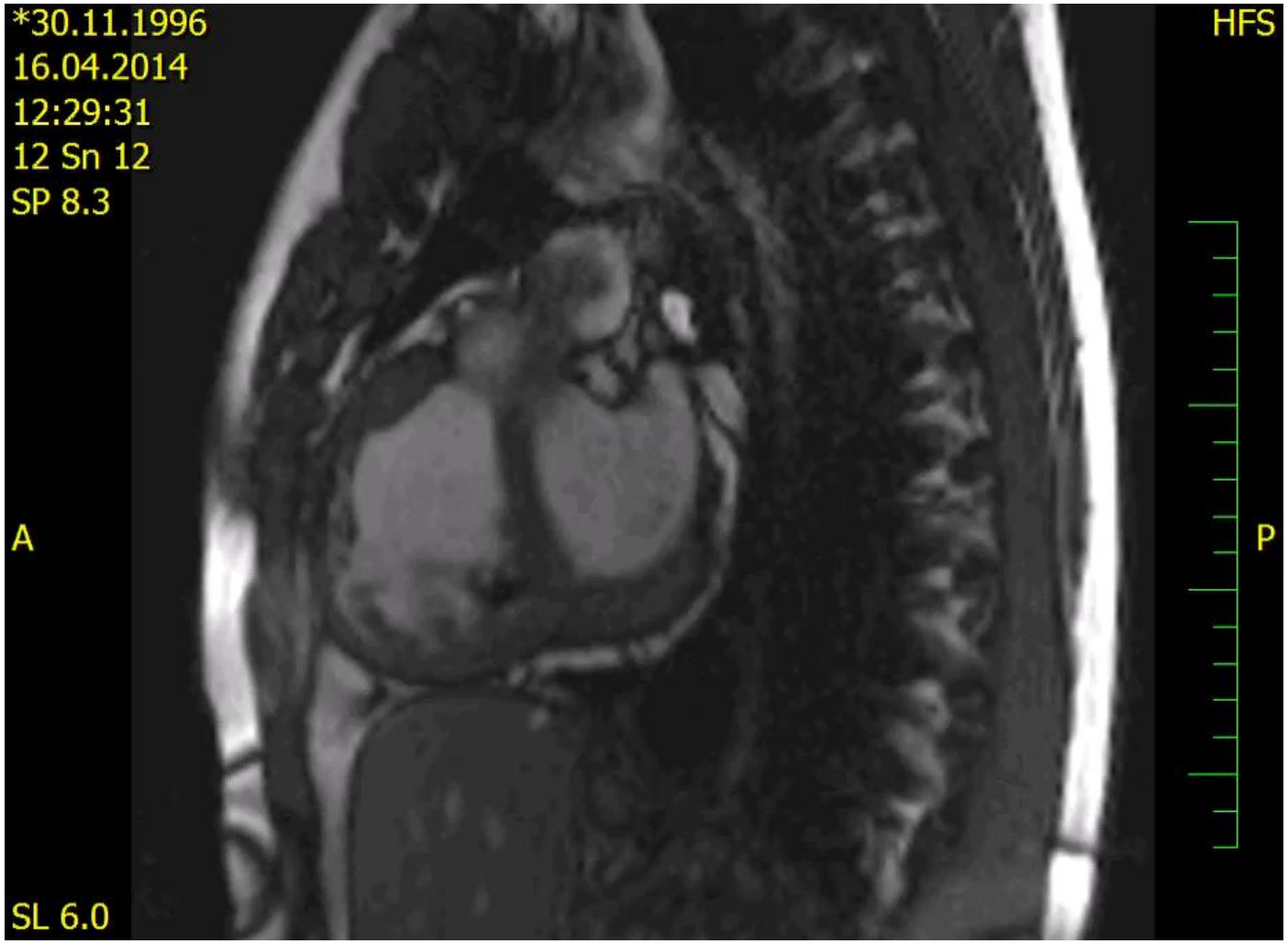
Images in Cardiovascular Medicine

Successful Permanent Resynchronization for Failing Right Ventricle After Repair of Tetralogy of Fallot

Circulation 2014

Peter Kubuš, MD; Ondřej Materna, MD; Petr Tax, MD; Viktor Tomek, MD; Jan Janoušek, MD, PhD





Circulation: Arrhythmia and Electrophysiology

SPECIAL REPORT

Janousek J et al., 2019

Cardiac Resynchronization Therapy for Treatment of Chronic Subpulmonary Right Ventricular Dysfunction in Congenital Heart Disease

N = 6

Chronic Response	Baseline	Last Follow-Up	*P* Value
QRS duration, ms	158 (29) [200, 180, 150]	113 (20) [140, 120, 90]	0.002
NYHA class ≥2 [n]	6/6 [3/3]	1/6 [0/3]	0.015
NT-proBNP, ng/L	842 (756) [N/A, 361, 556]	233 (175) [81, 123, 460]	0.156
RV fractional area change, %	17.5 (9.2) [18, 32, 24]	35.0 (3.3) [36, 34, 36]	0.006
RV end-diastolic area index, cm^2/m^2 BSA	28.1 (11.4) [32.0, 18.8, 18.2]	20.1 (3.6) [24.3, 17.5, 20.6]	0.198
RV end-systolic area index, cm^2/m^2 BSA	23.7 (11.2) [26.4, 12.8, 13.8]	13.1 (2.1) [15.5, 11.5, 13.2]	0.086
RV dP/dt_{max}, mm Hg/s	316 (153) [113, 301, 374]	444 (161) [305, 386, 409]	0.051
Late systolic right to left septal flash [n]	6/6 [3/3]	1/6 [0/3]	0.015
RV septal to lateral mechanical delay, ms	150 (80) [131, 88, 83]	1 (22) [–62, 81, 49]	0.044
RV systolic stretch fraction, %	28.4 (22.3) [22.5, 15.7, 7.5]	11.7 (4.8) [13.0, 11.0, 4.0]	0.092
LV ejection fraction, %	62 (19) [59, 29, 66]	62 (13) [72, 43, 71]	0.910

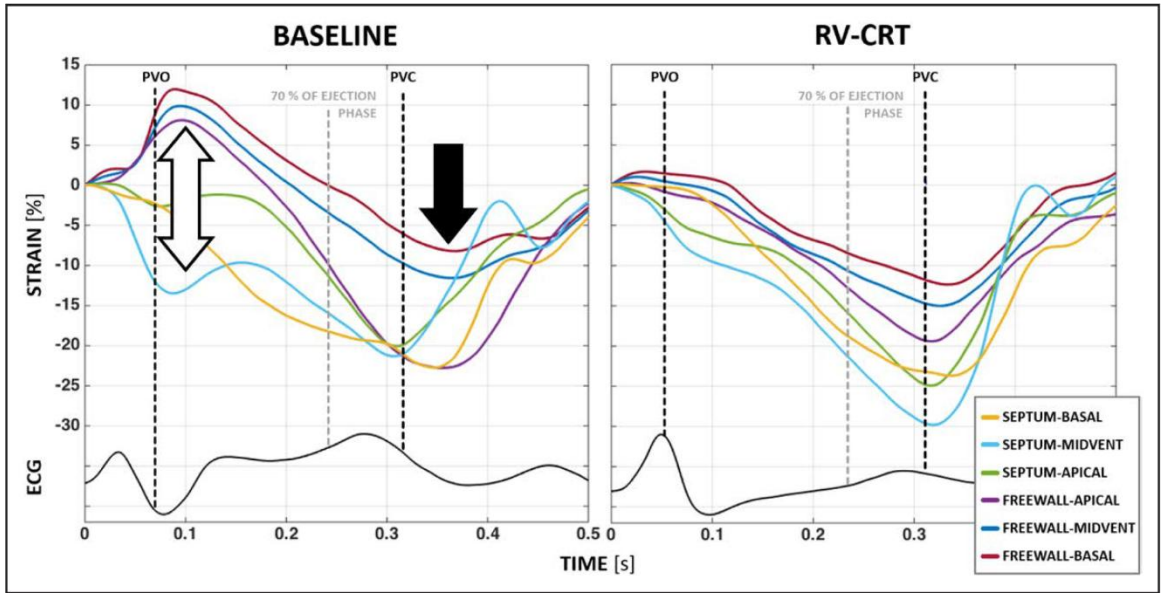
Pulmonary Right Ventricular Resynchronization in Congenital Heart Disease

Acute Improvement in Right Ventricular Mechanics and Contraction Efficiency

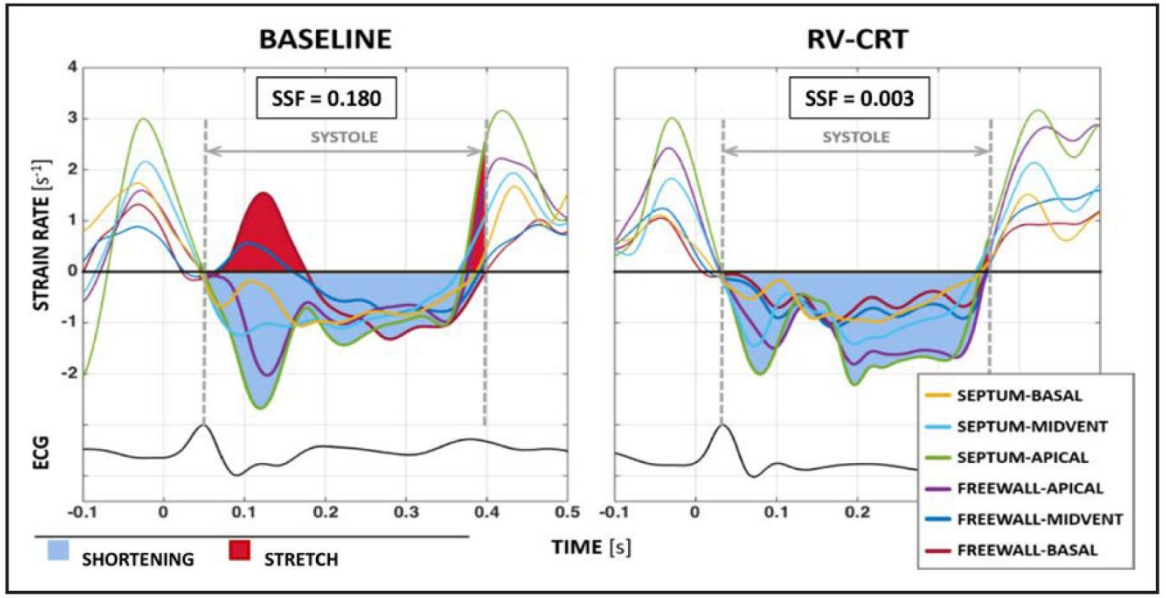
Circ Cardiovasc Imaging 2017

Jan Janoušek, MD, PhD; Jan Kovanda, MD; Miroslav Ložek, MSc; Viktor Tomek, MD, PhD; Pavel Vojtovič, MD; Roman Gebauer, MD; Peter Kubuš, MD, PhD; Miroslav Krejčíř, MSc; Joost Lumens, PhD; Tammo Delhaas, PhD; Frits Prinzen, PhD

RV mechanical discoordination



RV contraction efficiency



Adapted according to: Kirn et al., AJP Heart and Circ. Phys. 2008; 10.1152/ajpheart.00106.2008

Computational modelling of RV dyssynchrony

MECHANICAL WORK

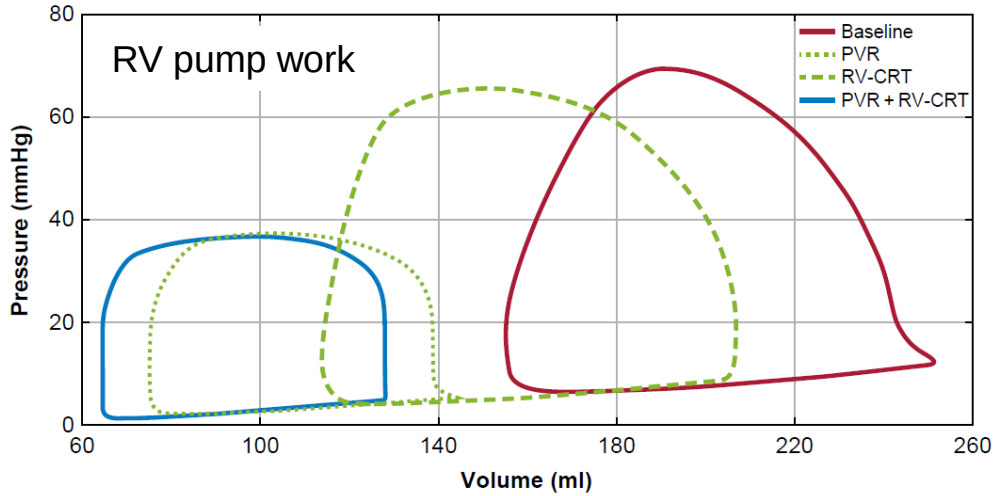
PUMP WORK: **mechanical work**, done by the ventricle

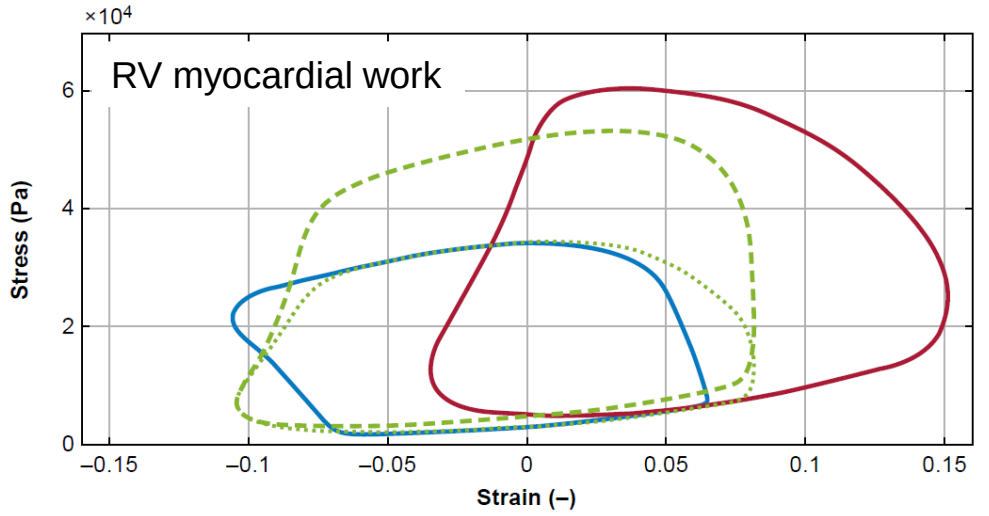
$$PW = \int p \cdot dV$$

MYOCARDIAL WORK: **mechanical work** done by the myocardium

$$MW = \int Stress \cdot dStrain$$

Parameter	Modelled patient #2			
	Baseline	PVR[b]	RV-CRT	PVR + RV-CRT
RV pump work (J/cycle)	0.55	0.26	0.62	0.27
RV myocardial work (J/cycle)	0.97	0.46	0.71	0.37
Standard deviation of segmental RV myocardial work density (J/m^3/cycle)	5171	2640	1813	176
RV myocardial work/cardiac output (J/L)	14.86	7.01	10.86	5.70
RV myocardial work/RV pump work	1.75	1.75	1.14	1.37
Modelled exercise: cardiac output at central venous pressure = 25 mmHg (L/min)	8.3	10.9	10.3	15.2





Ložek M et al. Europace 2024

Computational modelling of RV dyssynchrony

Cardiac output at maximum exercise

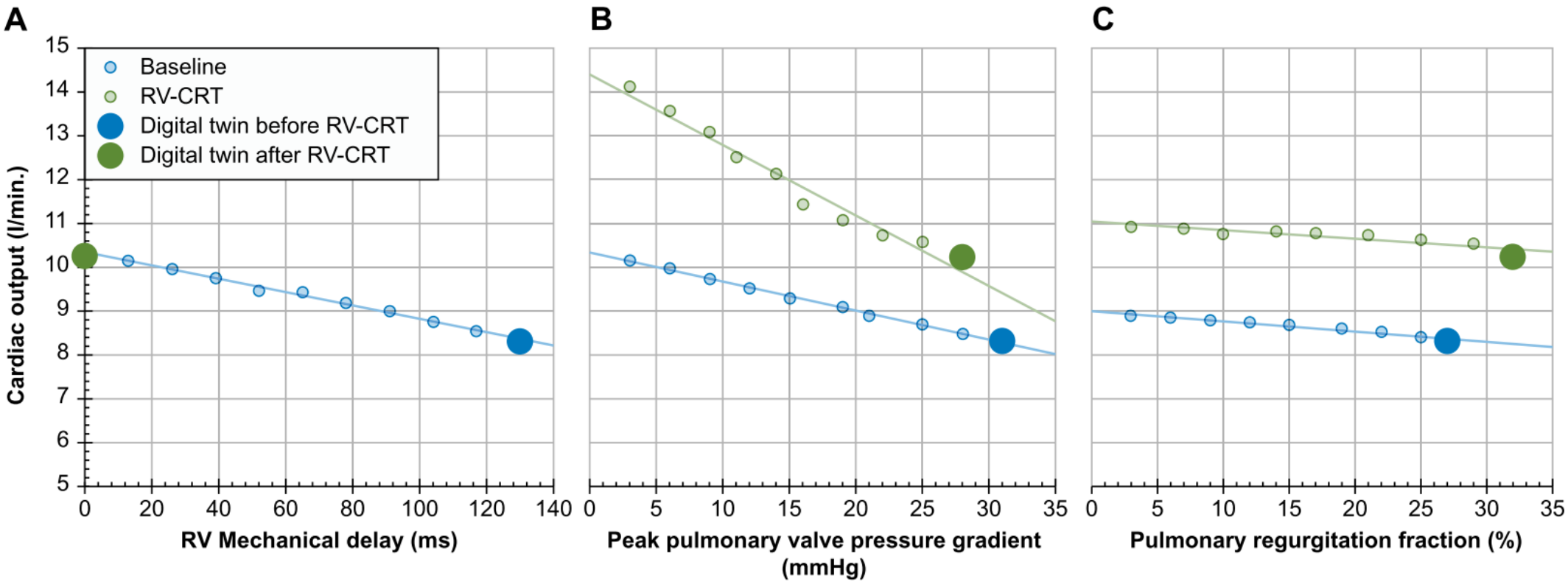


Figure 5 Modelled data from Patient #2. Cardiac output at maximum exercise is displayed at baseline and after RV-CRT as a function of (*A*). RV mechanical delay. (*B*) Pulmonary stenosis. (*C*) Pulmonary regurgitation. RV-CRT, RV cardiac resynchronization therapy.

Ložek M et al. Europace 2024

Summary

- RBBB most common dyssynchronopathy in CHD population
- ECG and ECHO is able to identify RV CRT correctable substrate
  - Classic pattern dyssynchrony
- Basic principles of RV CRT are the same as for LV CRT
- RV CRT improves RV metrics and contraction efficiency
  - Evolves to an additional strategy for treatment of chronic RV dysfunction (ToF, Ebstein and similar)
  - Survival benefit to be seen

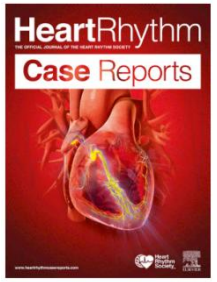
Thank you for your attention

Stay synchronized☺

Pulmonary RV CRT for Ebstein's anomaly?

Electrophysiology and surgery intertwined in complex treatment of Ebstein's anomaly in childhood 2022

Václav Chaloupecký Jr., MD,* Roman Gebauer, MD,* Jan Kovanda, MD,* Karel Koubský, MD, PhD,* Ioana Sus, MD,† Jan Janoušek, MD, PhD, FHRS*



4-years old girl, Ebstein anomaly, recurrent SVT
AP ablation, Cone repair , RV CRT

Table 1 Improvement of hemodynamic parameters during acute right ventricular cardiac resynchronization therapy testing

	RV-CRT off	RV-CRT on
RV filling time [ms]	272	305
RV maximum +dP/dt [mm Hg/s]	233	449
PA velocity time integral [cm] rowhead	15.1	16.7
RV septal-to–free wall mechanical delay [ms]	97	13
Pulm valve closure to peak RV free wall contraction [ms]	57	20
RV systolic stretch fraction	0.37	0.07

PA = pulmonary artery; Pulm = pulmonary; RV = right ventricular; RV-CRT = right ventricular cardiac resynchronization therapy.