Pulmonary right ventricular resynchronization in congenital heart disease (RV-CRT)

J. Janoušek

Children's Heart Center

Univ. Hosp. Motol

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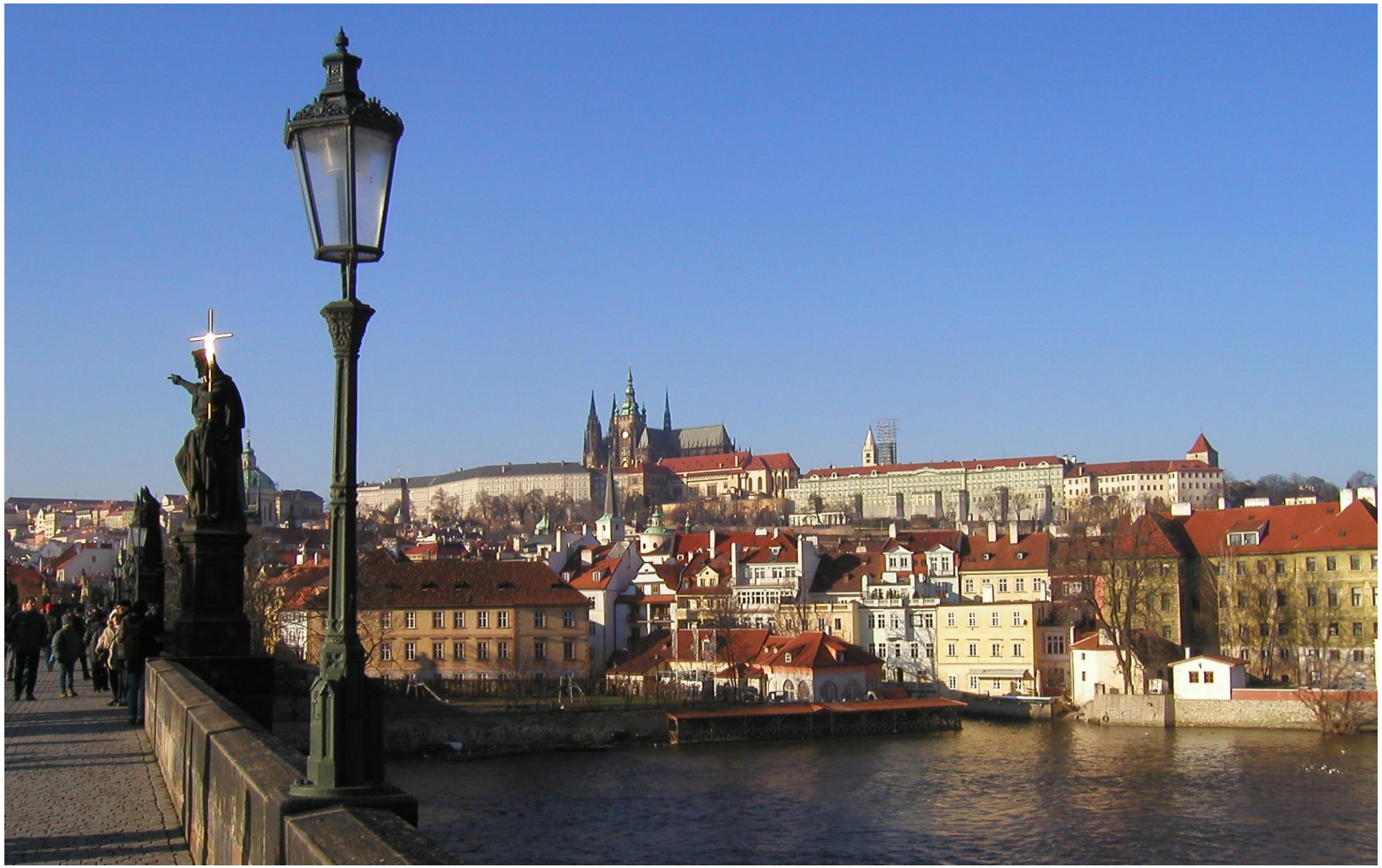
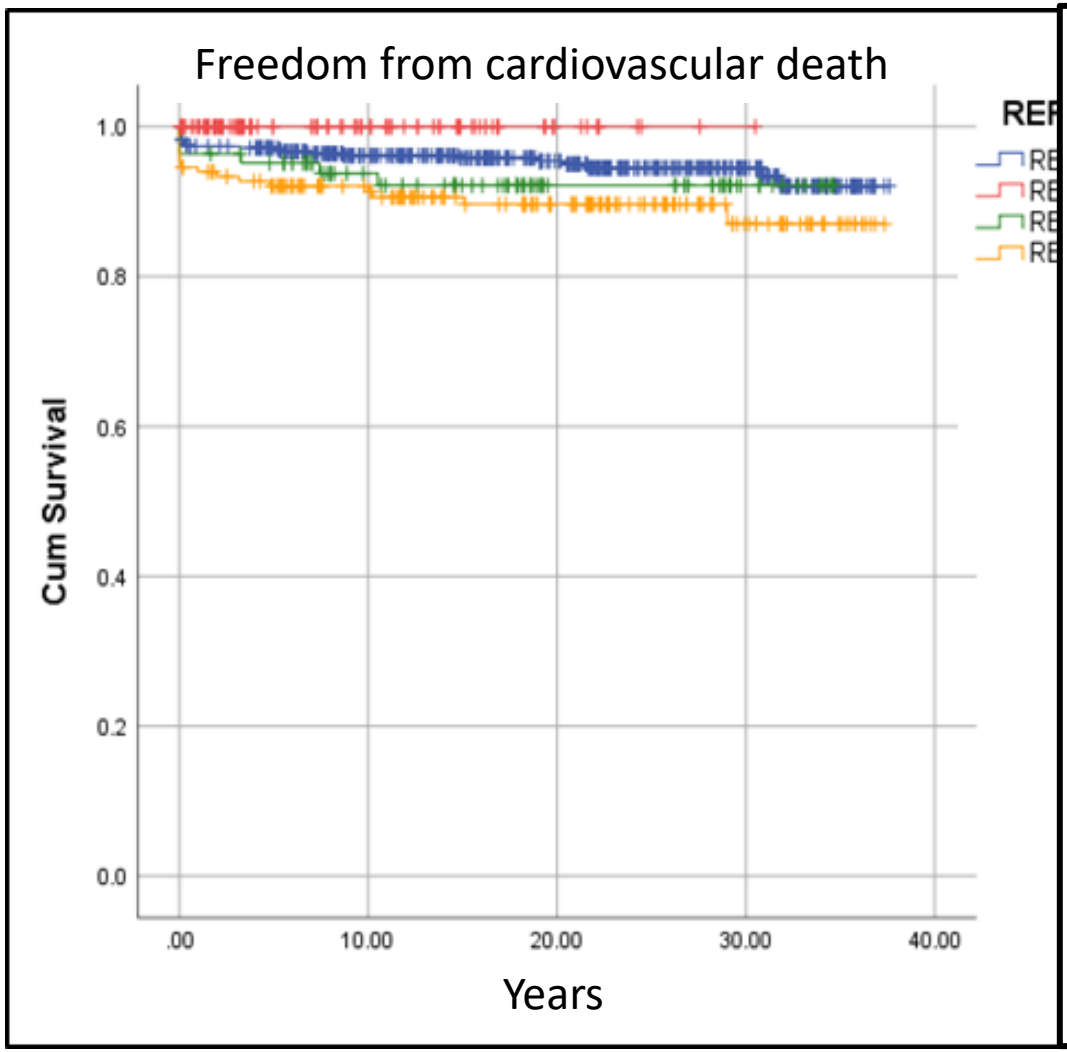
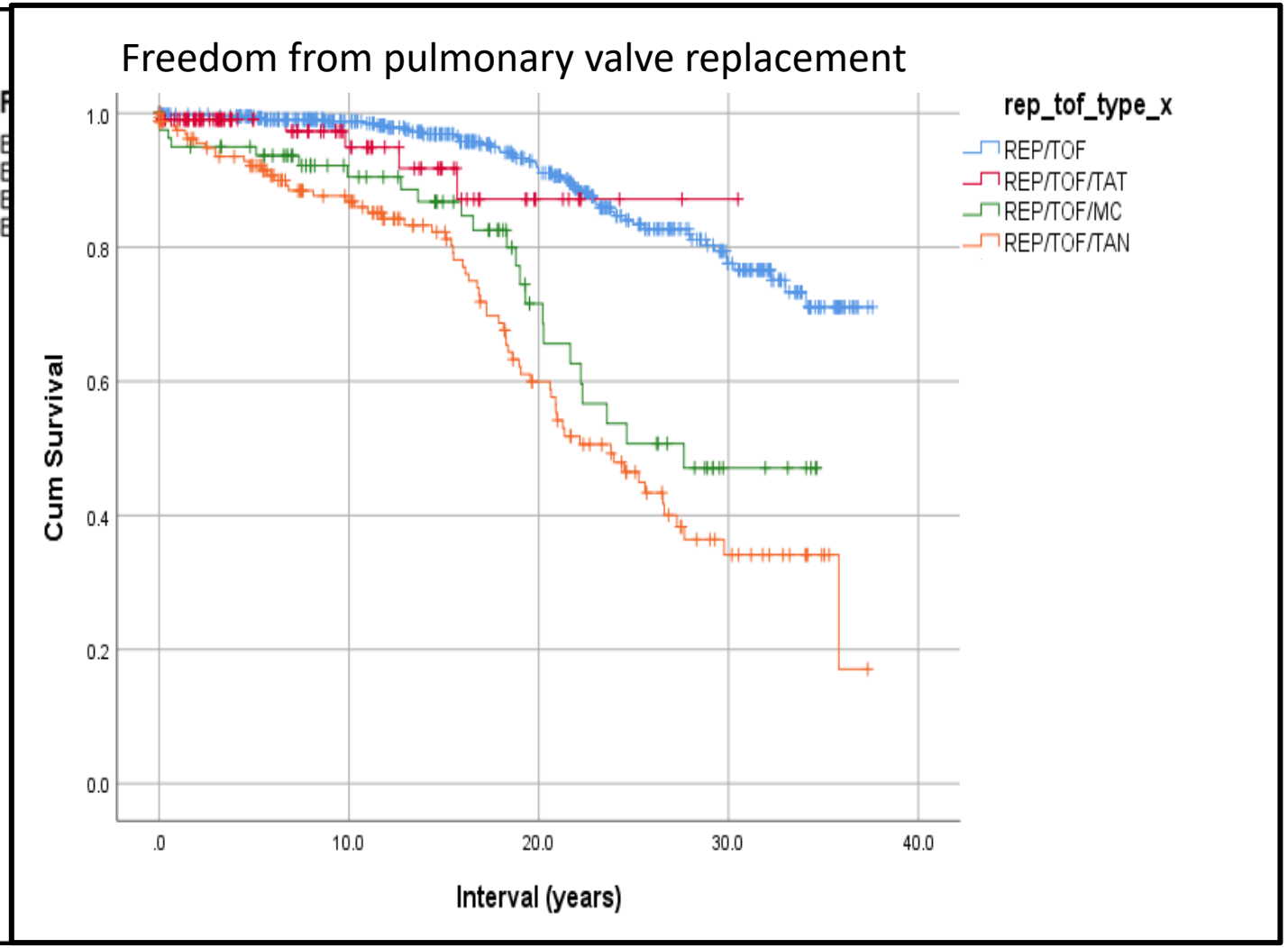


FN MOTOL

Survival probability after repair of tetralogy of Fallot in childhood according to type of repair

Children's Heart Center, N=814, 1979-2016





Postoperative tretralogy of Fallot

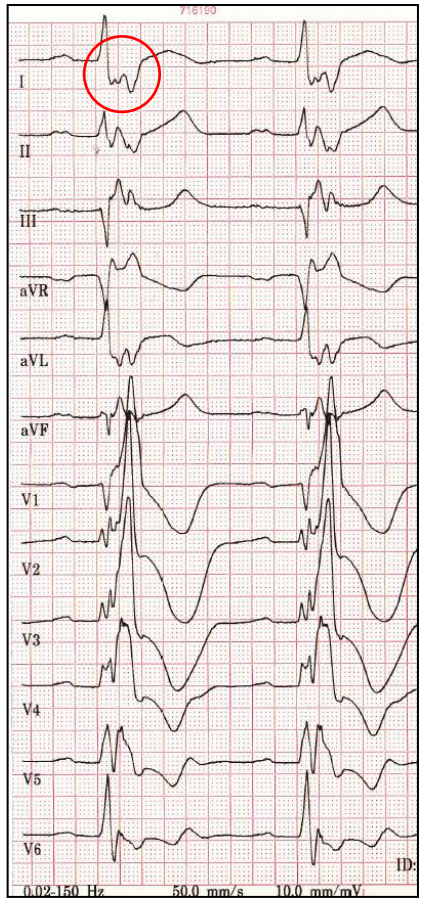
- Chronic RV failure model
- Decreased probability of RV reverse remodeling after pulmonary valve replacement
  - RVEDV >150 to 170 mL/m2 or RVESV > 82 to 90 mL/m2
  - RV EF ≤45%
  - **QRS ≥160 ms!**
- PVR alone may not lead to RV myocardial performance normalization
  - Myocardial fibrosis/scar?
  -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

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

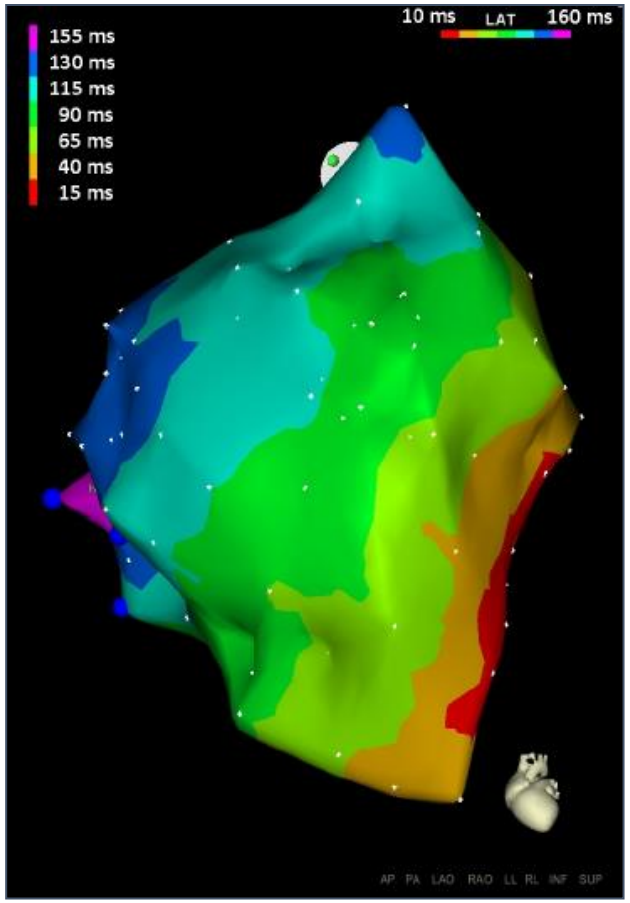
Electromechanical dyssynchrony and RV dysfunction after repair of ToF

From ECG to mechanical activation and ventricular function

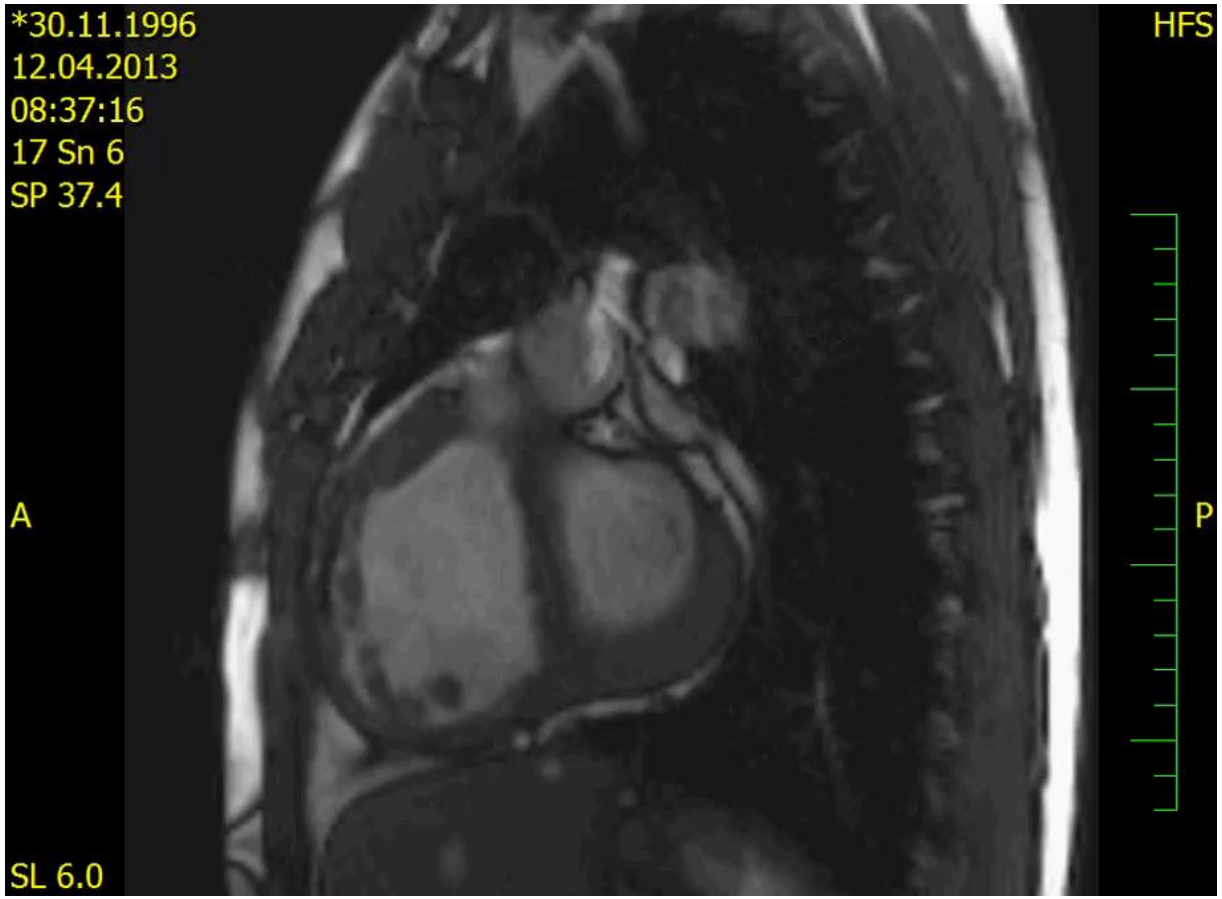
Right bundle branch block



RV activation map



RV function



Kubus P, Circulation 2014

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*

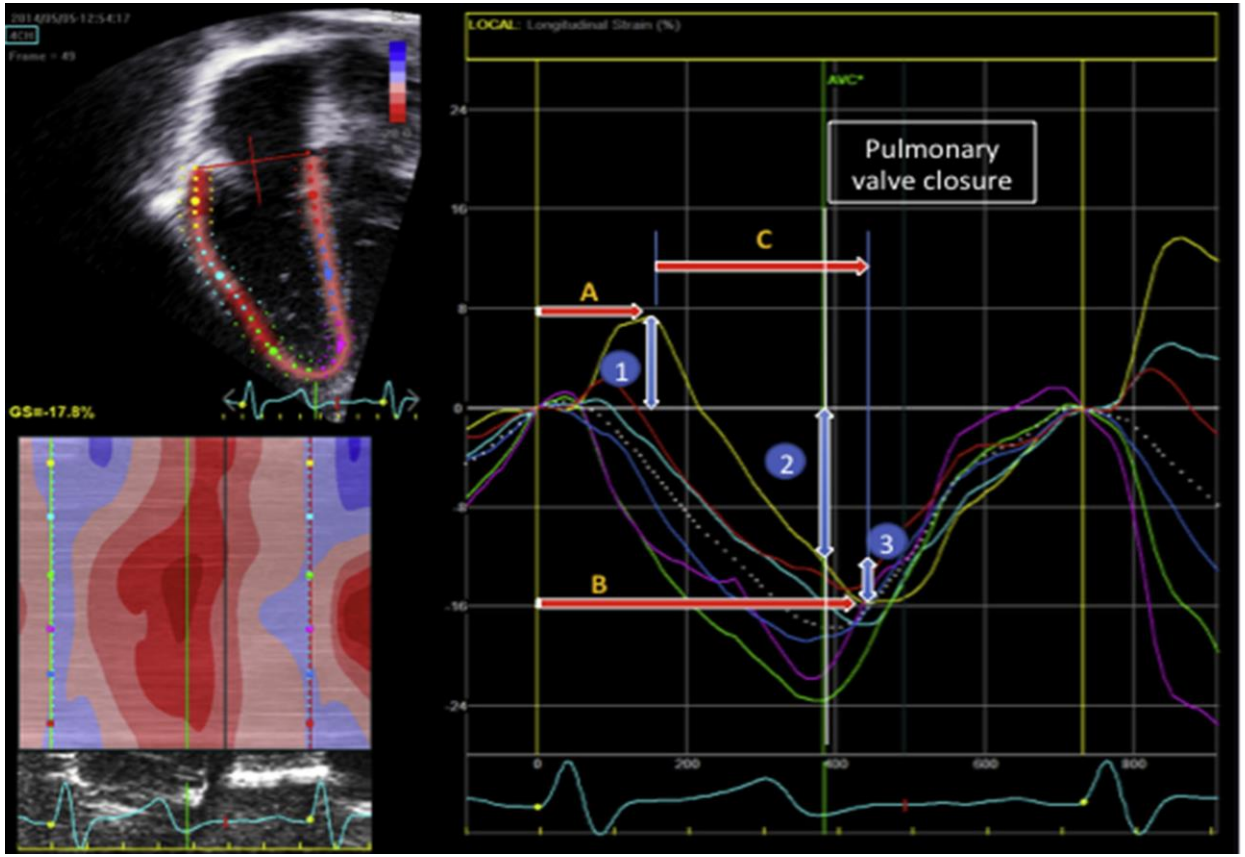


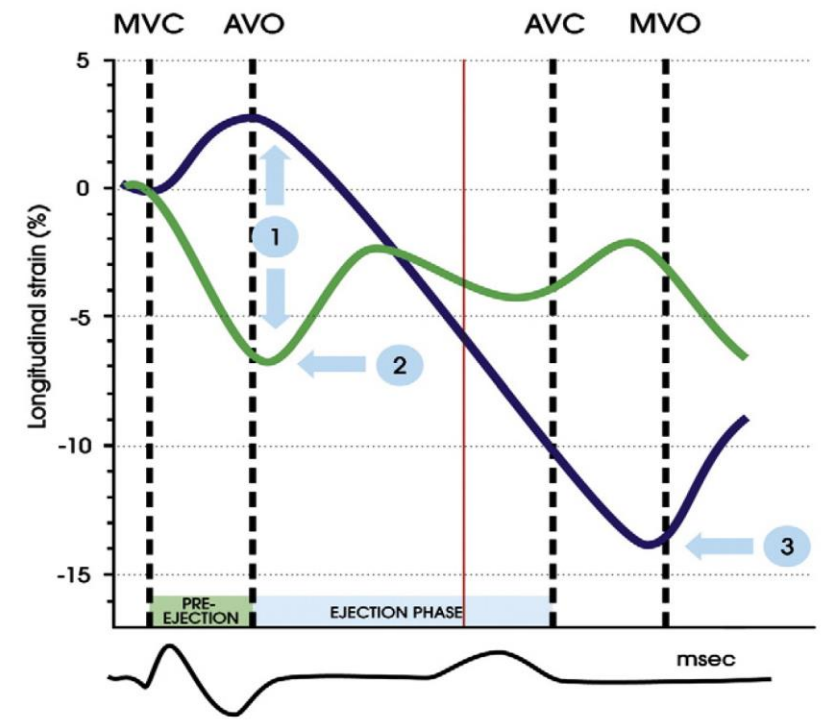
Table 4 Relationship of RV dyssynchrony parameters and function

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	+	++

Simple regional strain pattern analysis to predict response to cardiac resynchronization therapy: Rationale, initial results, and advantages

(Am Heart J 2012;163:697-704.)

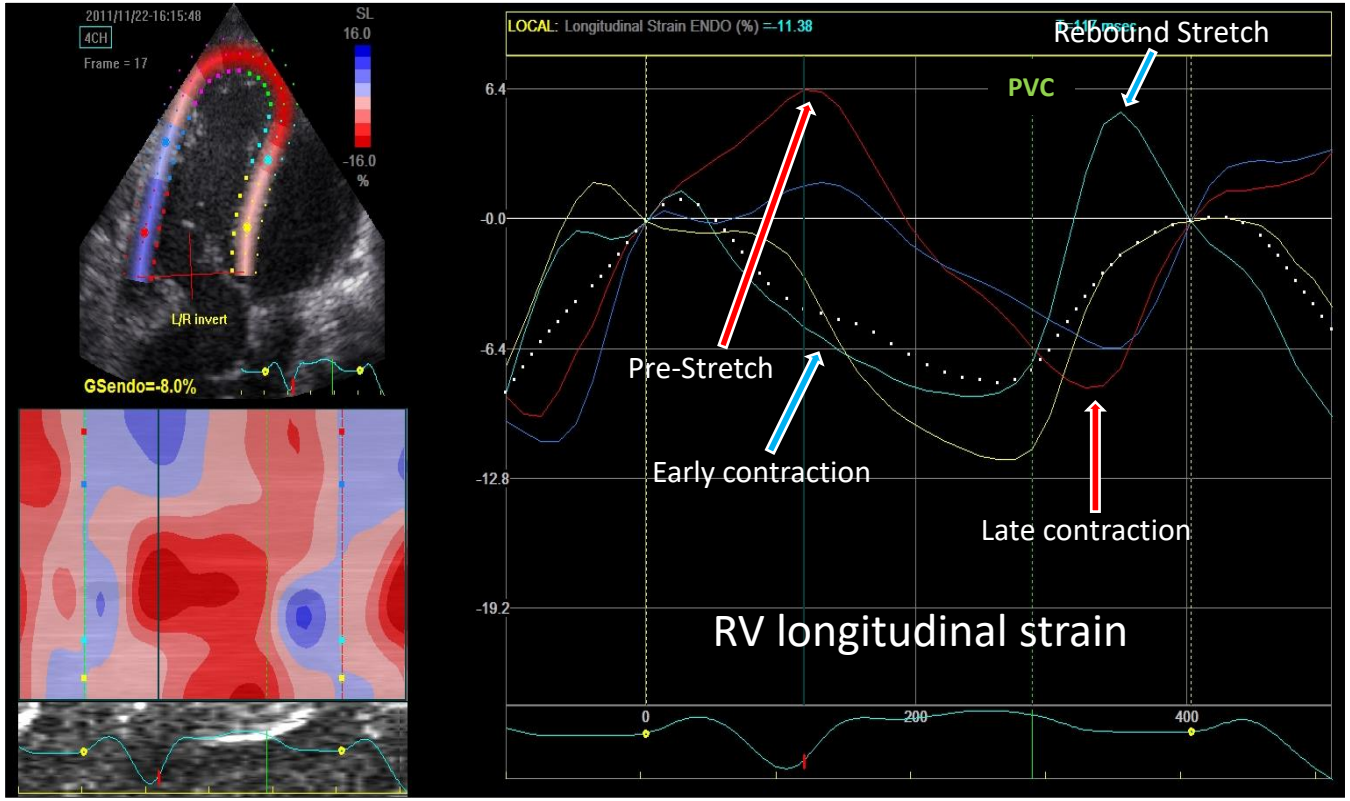
Niels Risum, MD,[a] Christian Jons, MD, PhD,[a] Niels T. Olsen, MD, PhD,[a] Thomas Fritz-Hansen, MD,[a]
Niels E. Bruun, MD, DMSc,[a] Michael V. Hojgaard, MD, PhD,[a] Nana Valeur, MD, PhD,[a]
Mads B. Kronborg, MD, PhD,[b] Joseph Kisslo, MD,[c] and Peter Sogaard, MD, PhD[a] *Skejby, Denmark; and Durham, NC*



Classic-pattern dyssynchrony

1. Early septal contraction and *early lateral wall stretching*
2. Peak septal contraction <70 % of ejection phase followed by *rebound stretch*
3. Peak lateral wall contraction after AVC

Correlates wit CRT efficacy



Indications for CRT in patients in sinus rhythm

For LV only

Recommendations	Class	Level
1) LBBB with QRS duration >150 ms is recommended in chronic HF patients and LVEF ≤35% who remain in NYHA functional class II, and ambulatory IV despite adequate medical treatment. (*)	I	A
2) LBBB with QRS duration 120-150 ms should be considered in chronic HF patients and LVEF ≤35% who remain in NYHA functional class II, and ambulatory IV despite adequate medical treatment. (*)	I	B
3) Non-LBBB with QRS duration >150 ms should be considered in chronic HF patients and LVEF ≤35% who remain in NYHA functional class II, and ambulatory IV despite adequate medical treatment. (*)	IIa	B
4) Non-LBBB with QRS duration 120-150 ms may be considered in chronic HF patients and LVEF ≤35% who remain in NYHA functional class II, and ambulatory IV despite adequate medical treatment. (*)	IIb	B
5) QRS duration <120 ms CRT in patients with chronic HF with QRS duration <120 ms is not recommended.	III	B

* Patients should generally not be implanted during admission for acute decompensated HF. In such patients, guideline-indicated medical treatment should be optimized and the patient reviewed as an out-patient after stabilization. It is recognized that this may not always be possible.

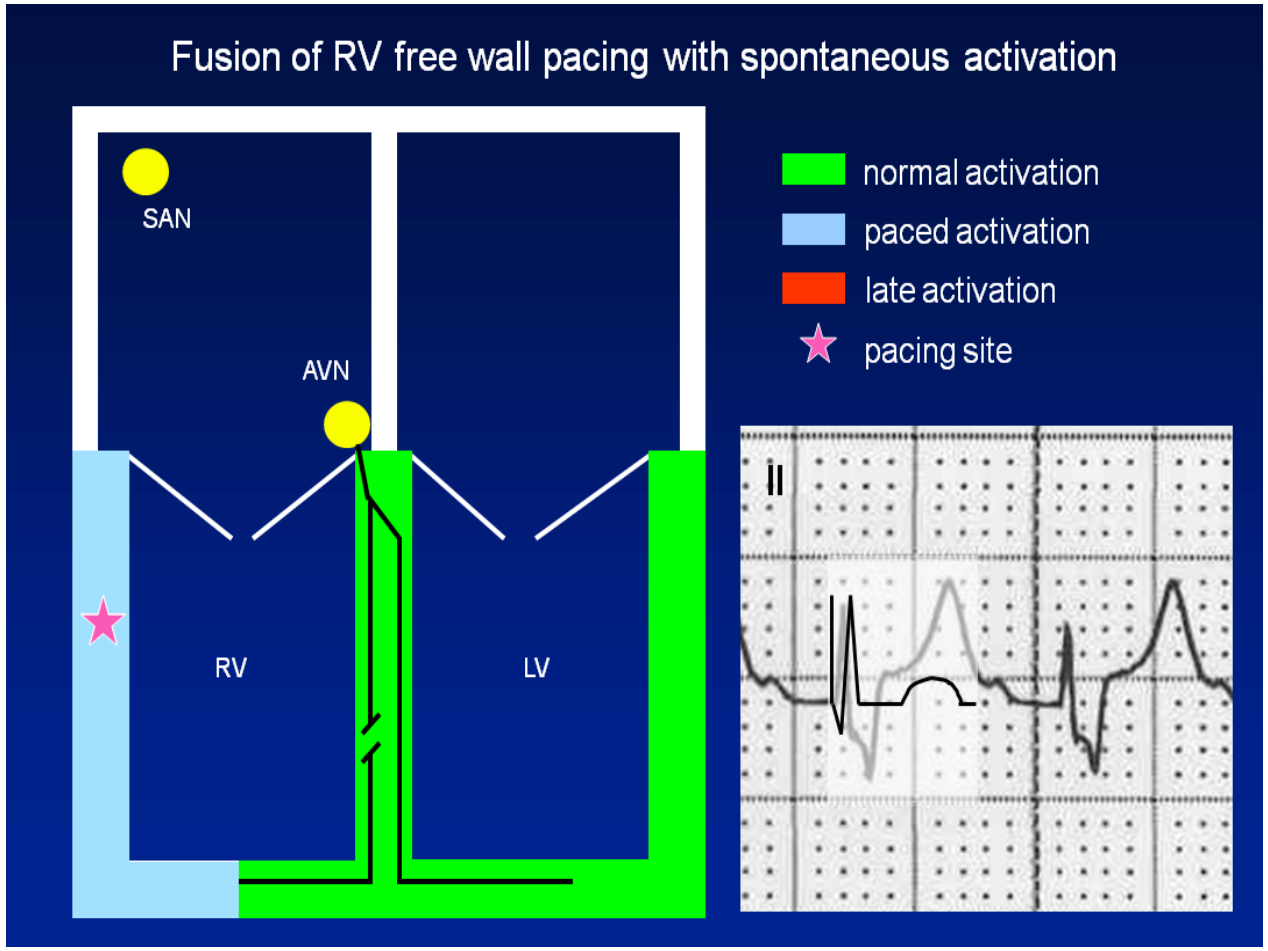
www.escardio.org/guidelines

European Heart Journal 2013;34:2281-2329
Europace 2013;15:1070–1118

EUROPEAN SOCIETY OF CARDIOLOGY®

CRT in pulmonary RV failure

Acute hemodynamic study



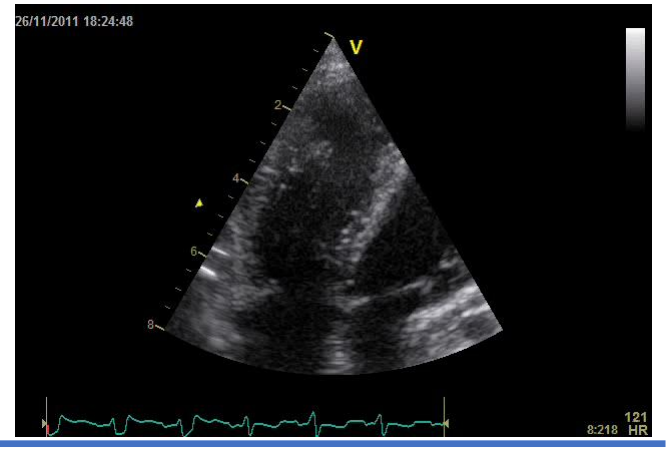
- N=7
- RV dysfunction and RBBB
- Results
  - ↓QRS duration
  - ↑cardiac index
  - ↑RV dP/dt

Dubin AM et al. Circulation 2003

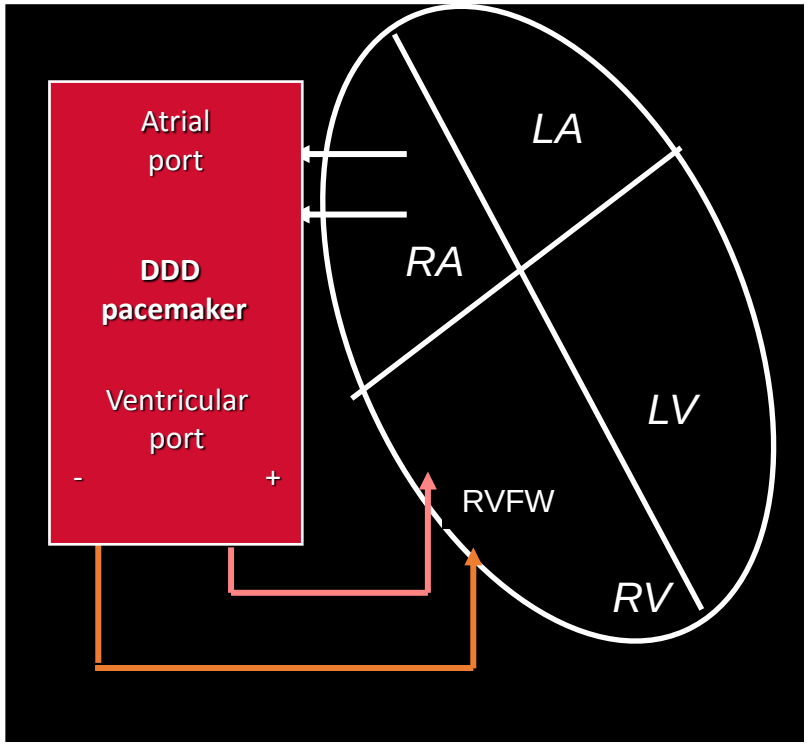
Acute right ventricular resynchronization improves haemodynamics in children after surgical repair of tetralogy of Fallot

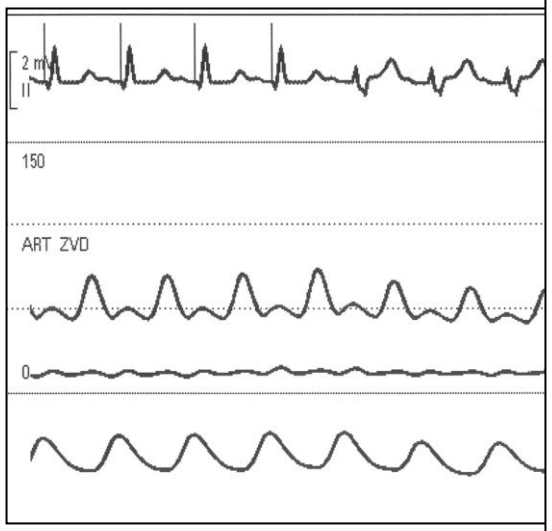
Europace 2018

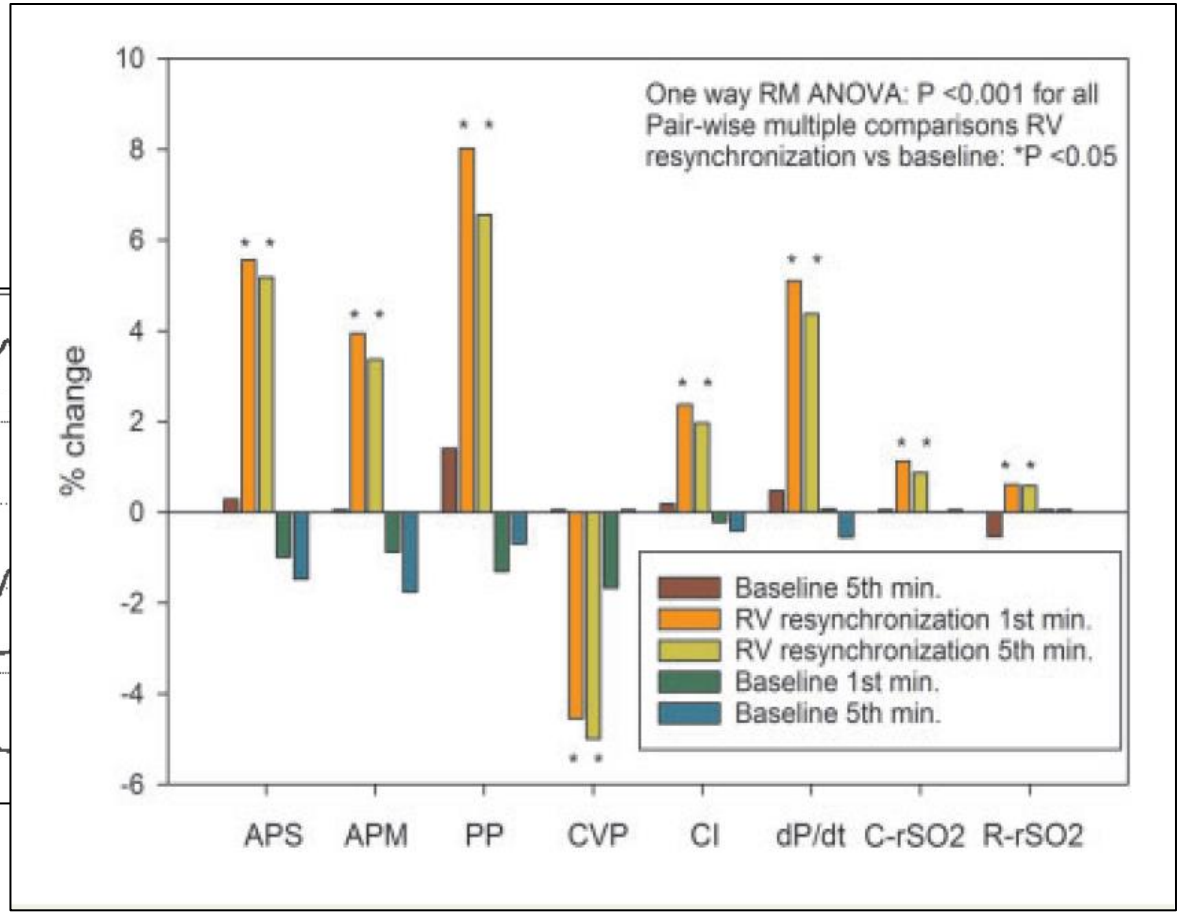
Pavel Vojtovič*, Filip Kučera, Peter Kubuš, Roman Gebauer, Tomáš Matějka, Tomáš Tláskal, Miroslav Ložek, Jan Kovanda, and Jan Janoušek



Atrial-triggered RV free wall pacing
in complete fusion with spontaneous ventricular activation (N=28)





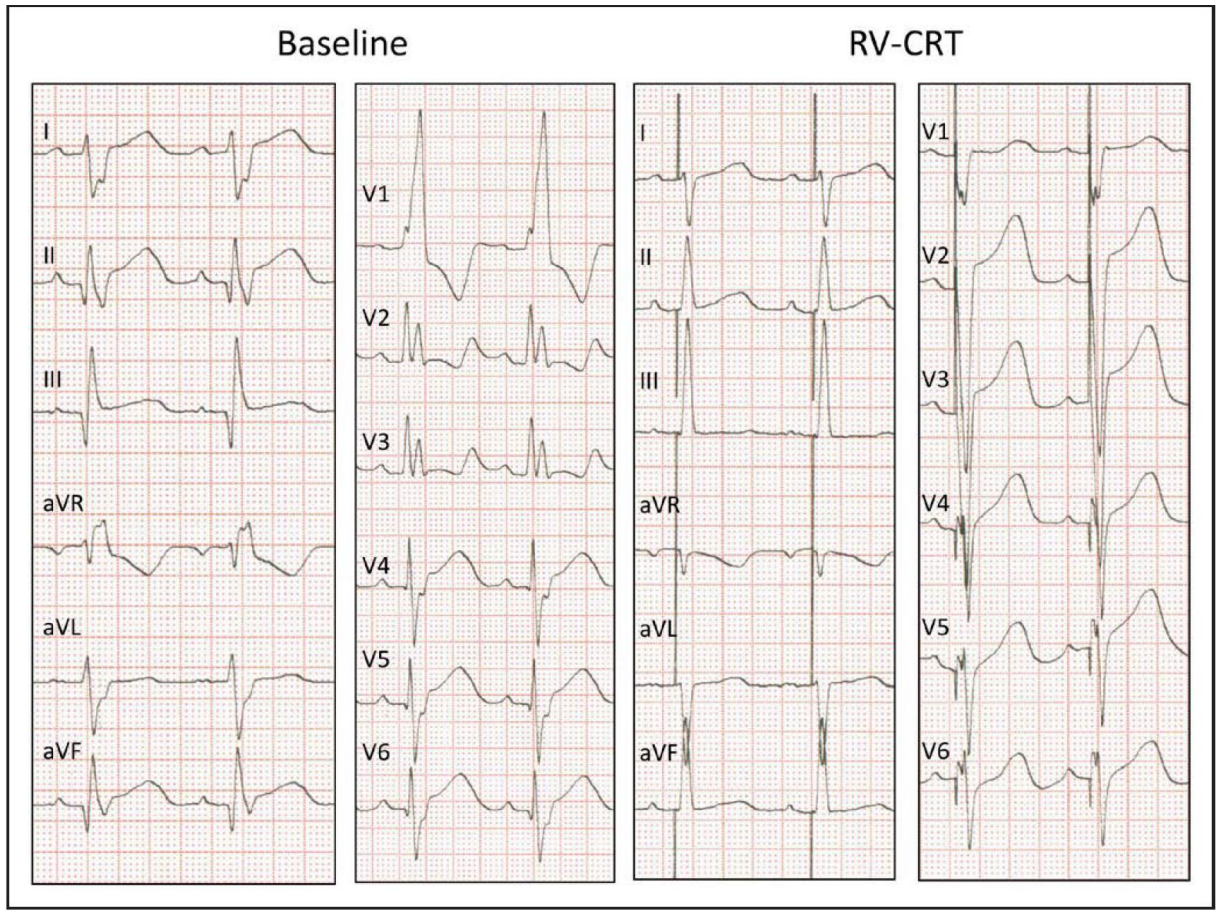


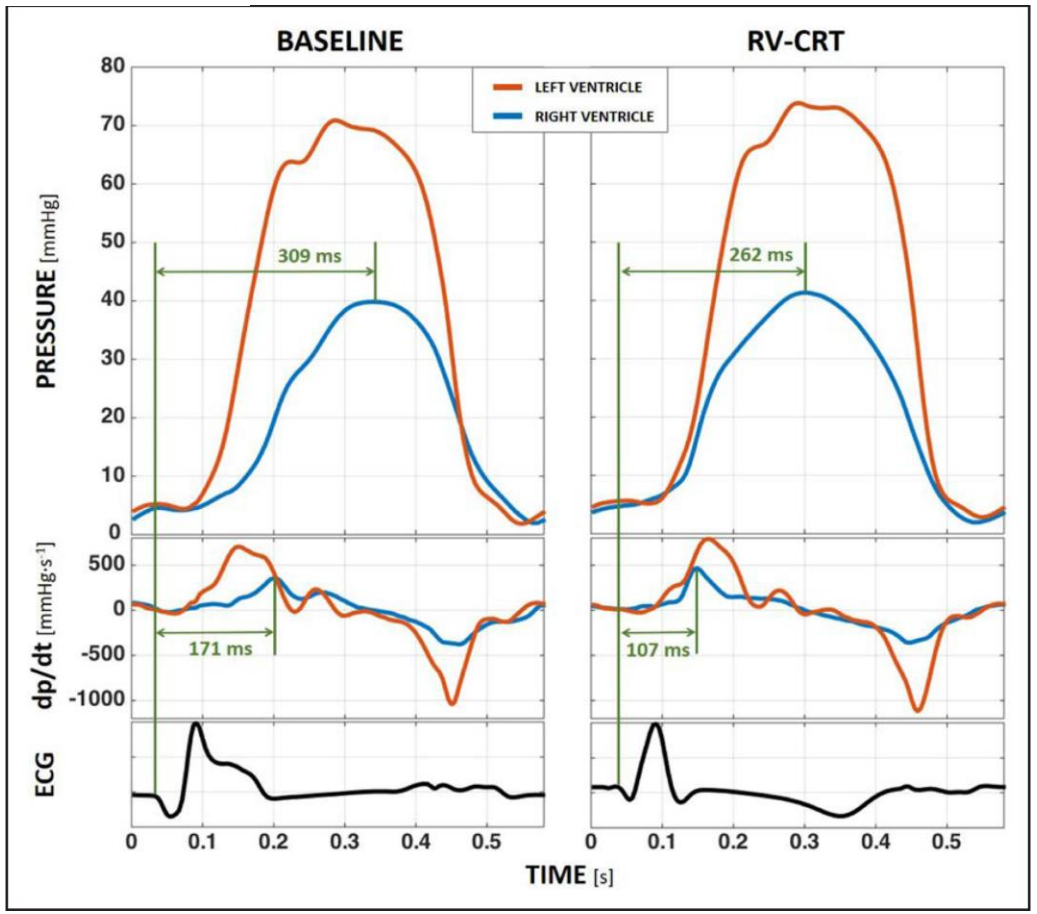
Pulmonary Right Ventricular Resynchronization in Congenital Heart Disease

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





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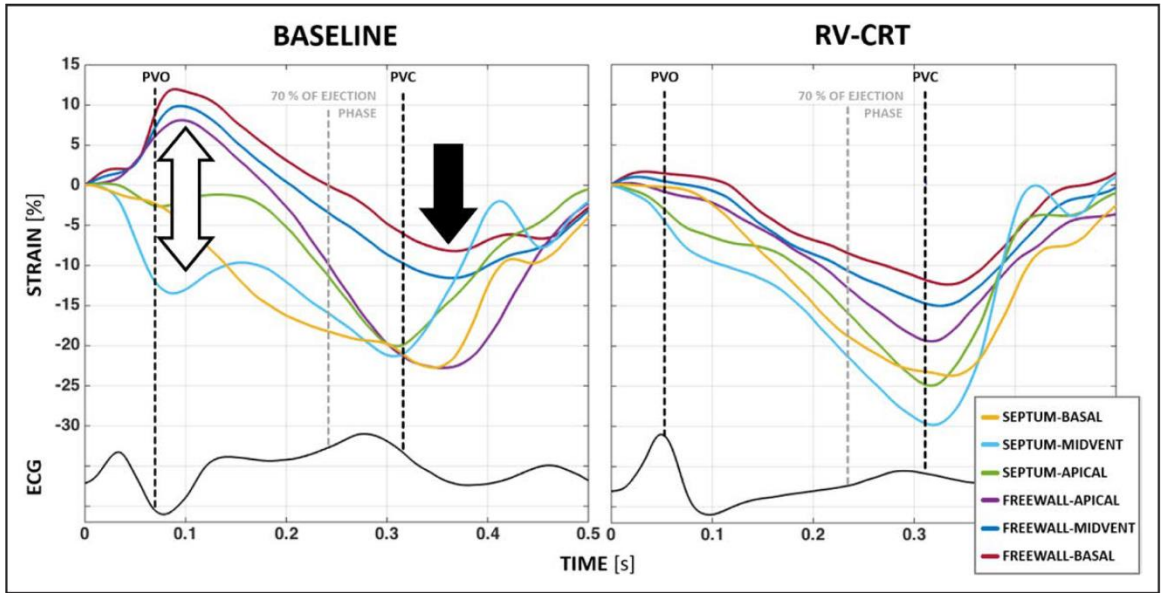
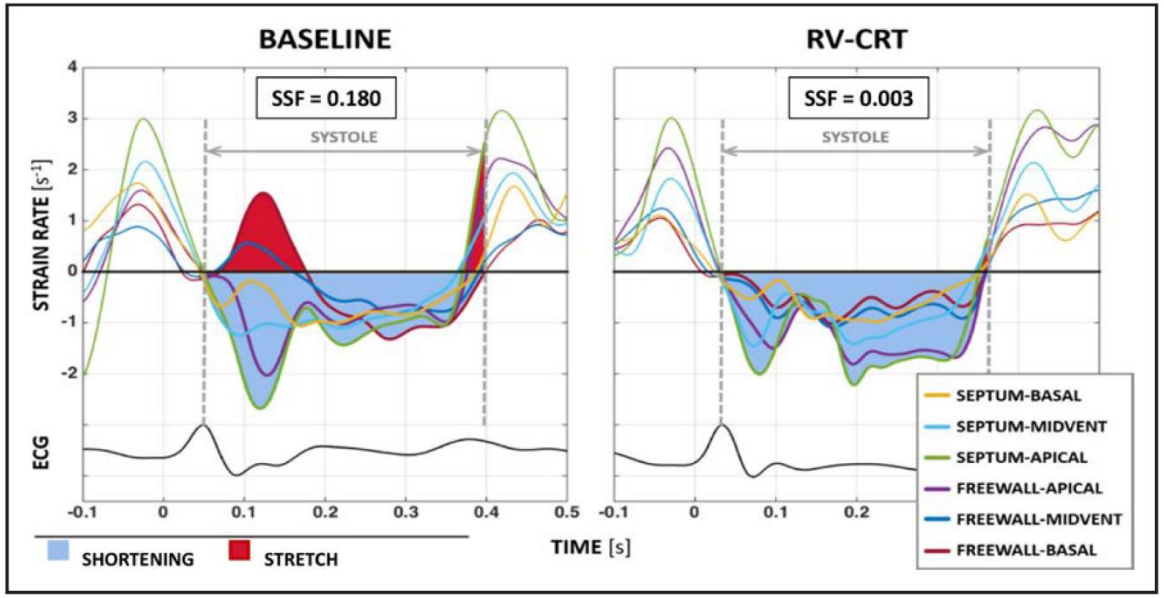
RV mechanical discoordination



RV contraction efficiency



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

Circulation: Arrhythmia and Electrophysiology

Janousek J et al., 2019

SPECIAL REPORT

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

2019 May;12(5):e007157. doi: 10.1161/CIRCEP.119.007157. IF 4,97, Q1, Cardiovascular journal ranking: 26

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

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association®

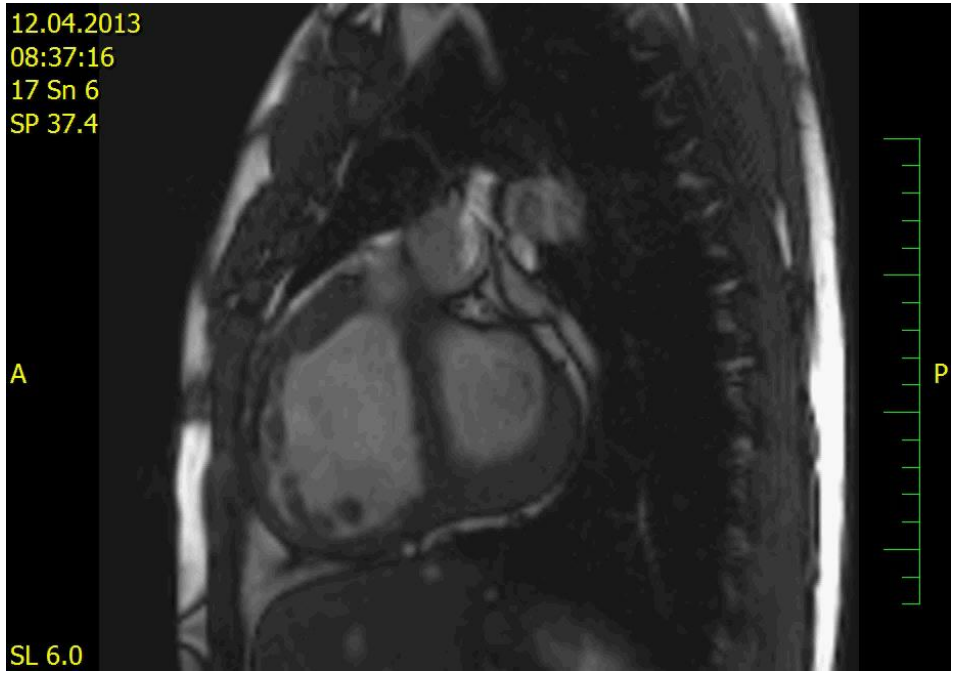
Circulation. 2014;130:e186-e190

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

Peter Kubus, Ondrej Materna, Petr Tax, Viktor Tomek and Jan Janousek

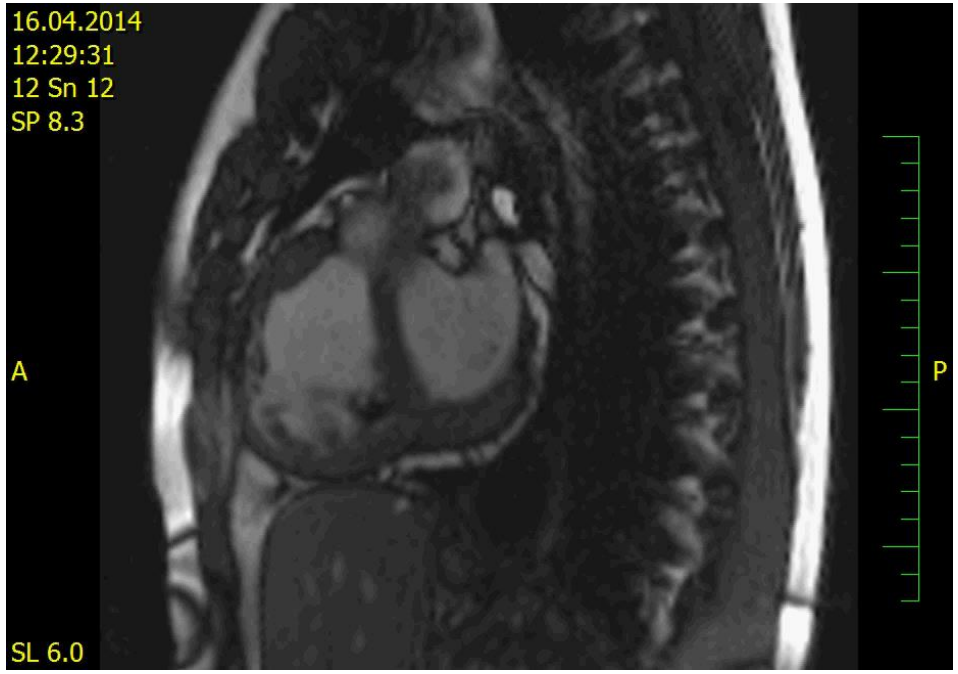
Before

- RV: EDV/ESV 212/172 ml/m2, EF 19 %
- LV: EDV/ESV 80/46 ml/m2, EF 41 %



6 months after

- RV: EDV/ESV 141/87 ml/m2, EF 38 %
- LV: EDV/ESV 63/28 ml/m2, EF 56 %



VO_2 max: 21.0 ➞ 30.4 ml/kg/min., NYHA II ➞ I.

Conclusions

- Pulmonary RV electromechanical discoordination is associated with:
  - Decreased RV contraction efficiency
  - Decreased RV function and pathologic remodeling
- RV-CRT may be used for:
  - Acute management of low cardiac output in the postoperative setting
  - Treatment of chronic dyssynchronous RV failure
- RV-CRT may be used as adjunct to correction of RV volume overload
- Exact indication criteria remain to be established
  - Modeling may be hepful in predicting RV-CRT effect