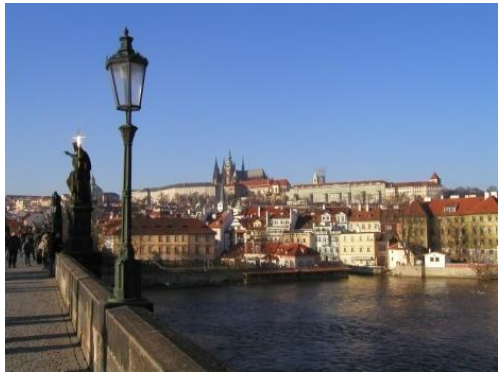
Dyssynchronopathy in Congenital Heart Disease

Diagnosis, Prevention and Therapy

J. Janoušek

Children's Heart Center
University Hospital Motol
Prague, Czech Republic



Indications for CRT in CHD

- Systemic LV failure
  - Left bundle branch block
  - RV paced
- Systemic RV failure
  - Right bundle branch block
  - LV paced
- Single-ventricular failure
  - Any bundle branch block
  - Single site pacing
- Pulmonary RV failure?
  - Right bundle branch block

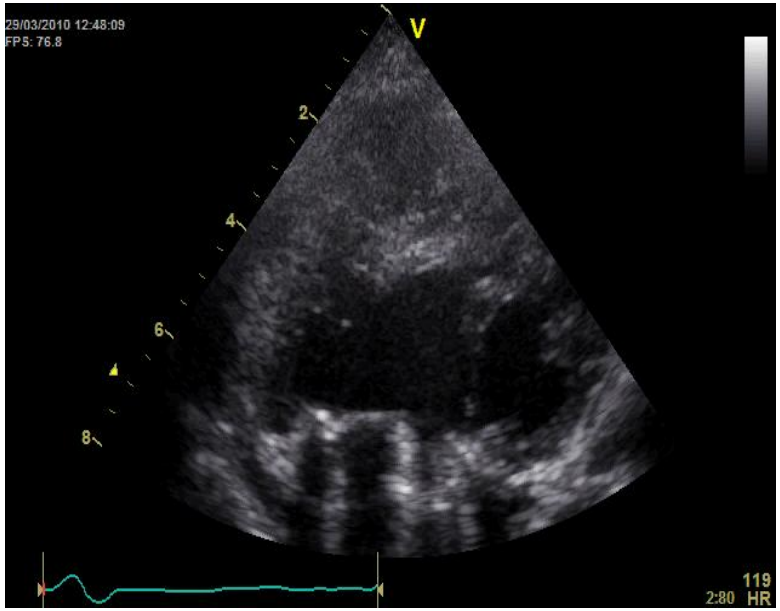
→ The well-studied setting

In any case:

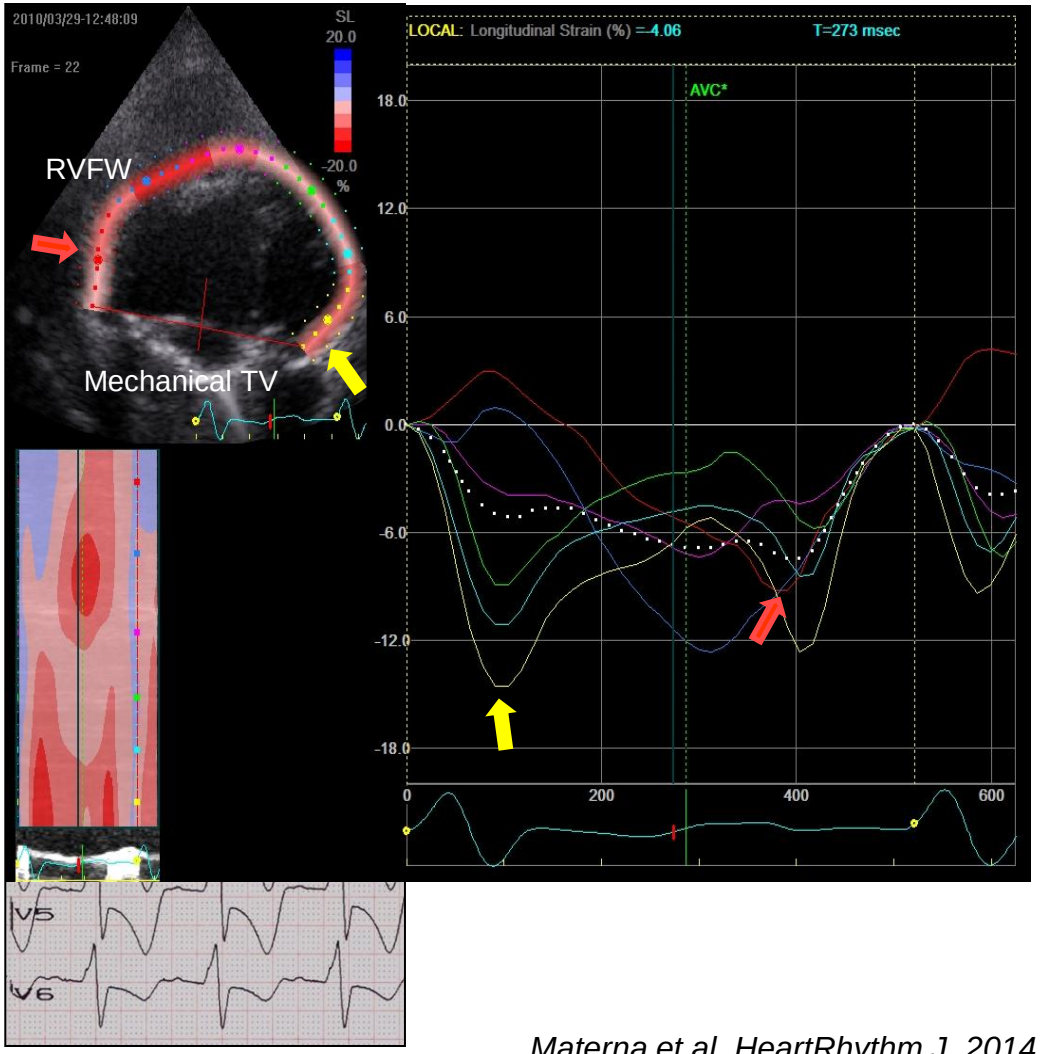
Major electrical activation delay within the failing ventricle needed for indication!

Specific for CHD

Mechanical activation mapping



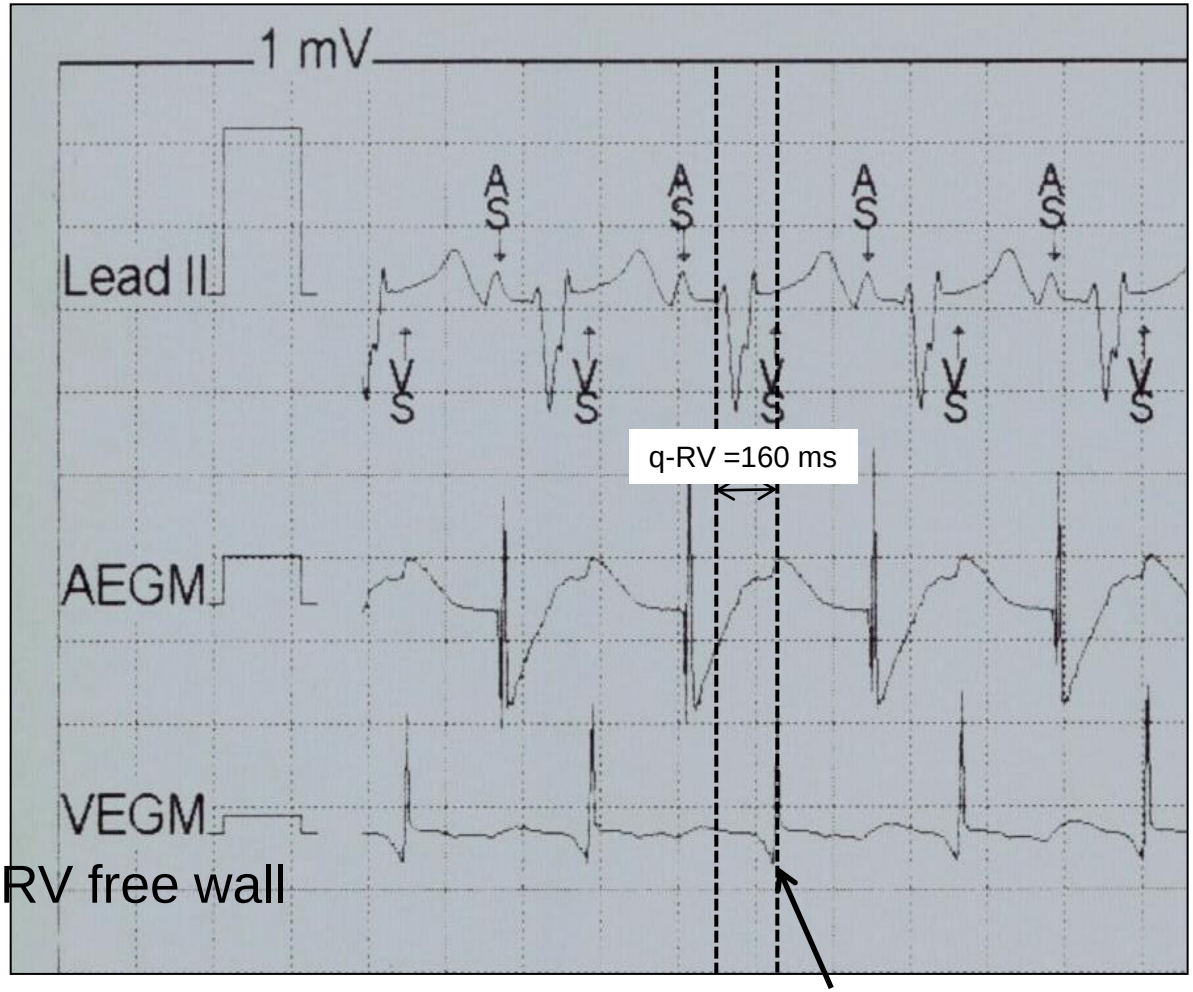
HLHS, st.p. BCPA and TV replacement
Failing dyssynchronous RV due to RBBB



Materna et al. HeartRhythm J. 2014

Peri-procedural electrical activation mapping

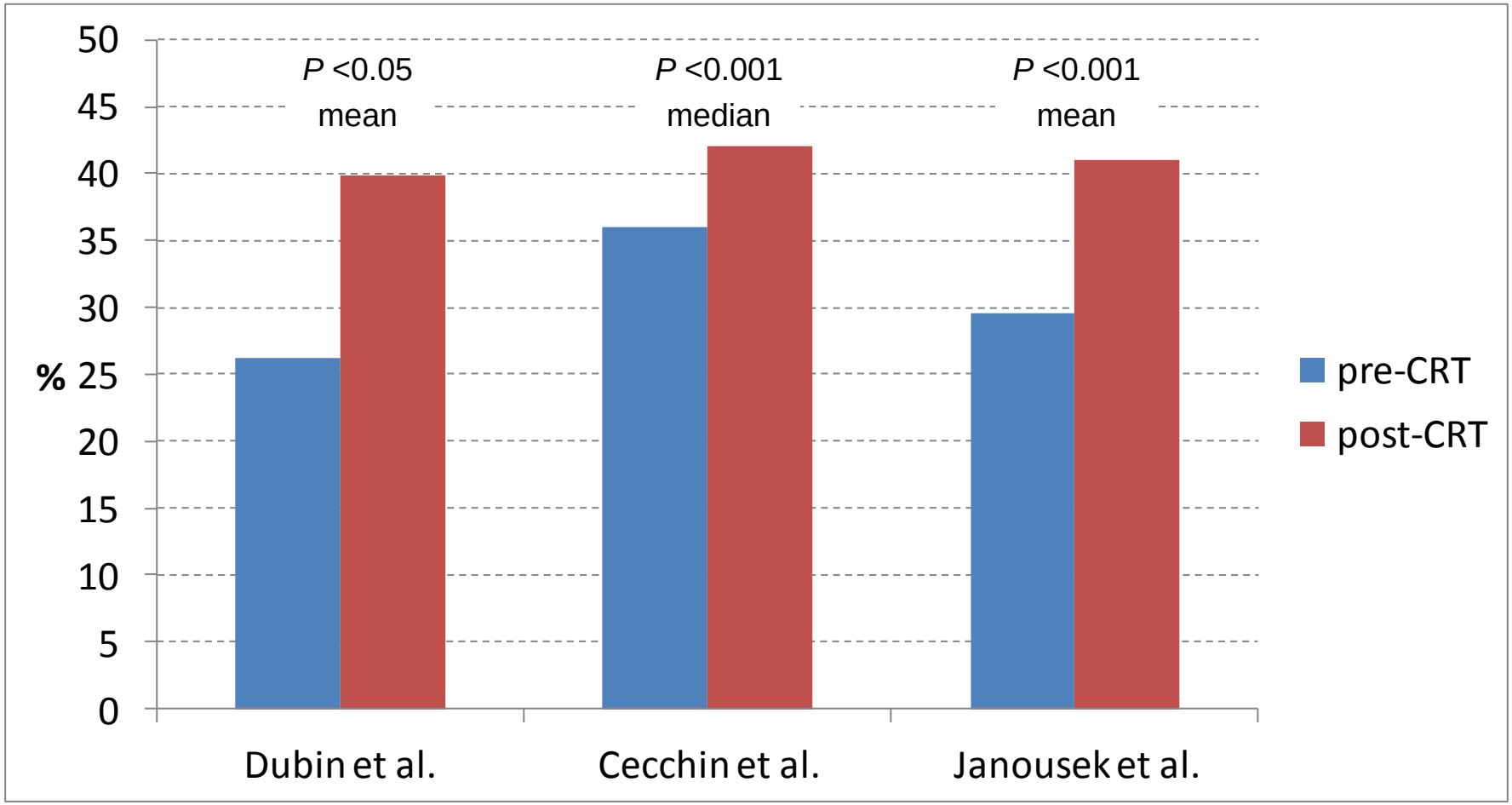
Search for latest local electrical activation during baseline rhythm



Materna et al. HeartRhythm J. 2014

Late activation at the right ventricular free wall

Systemic ventricular ejection fraction in pediatric/CHD CRT studies

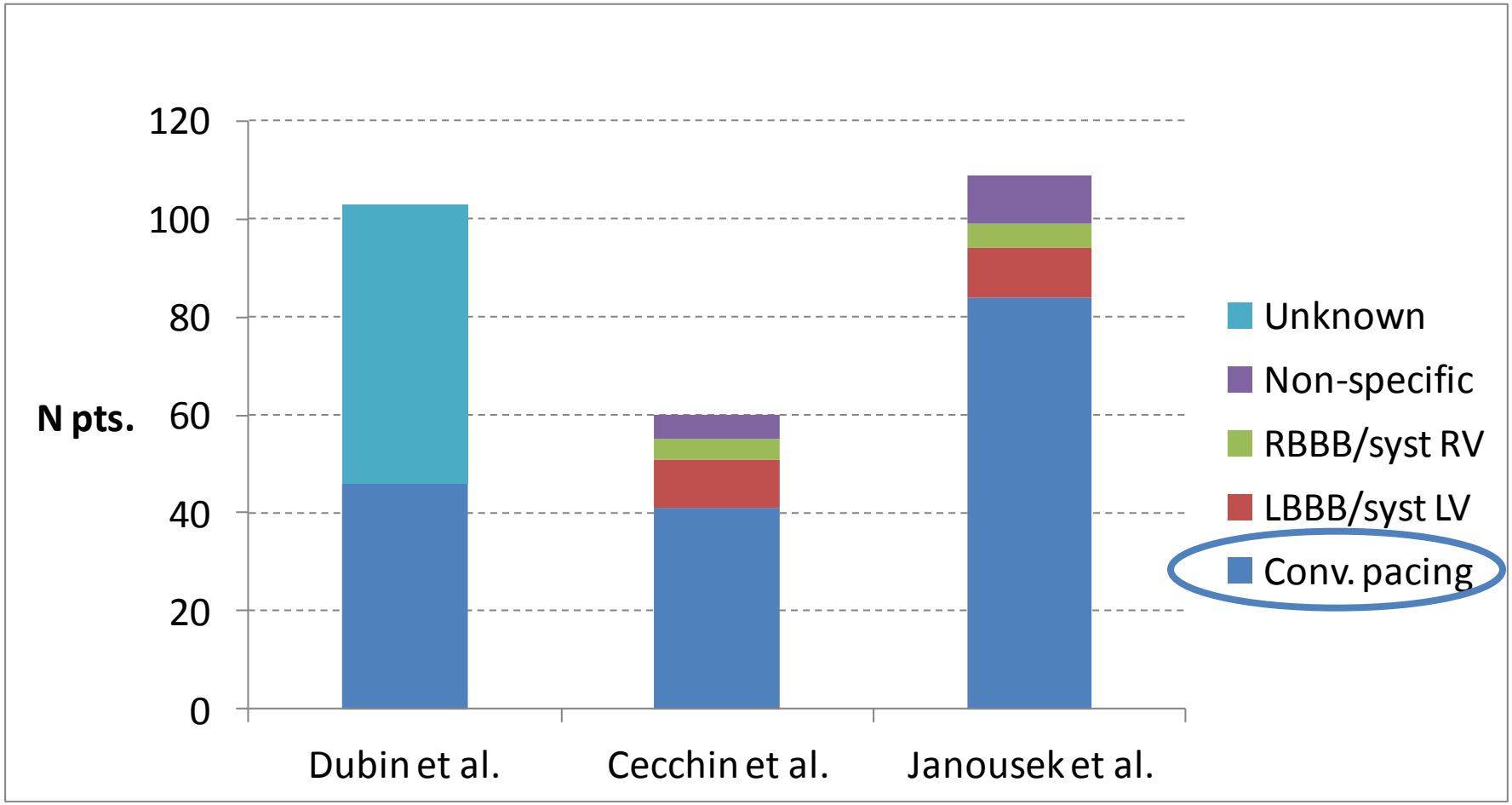


Dubin AM et al. J Am Coll Cardiol 2005;46:2277-83

Cecchin F et al. JCE 2009;20:58-65

Janousek J et al. Heart 2009, 95:1165-71

Types of electrical dyssynchrony in pediatric/CHD CRT studies



Dubin AM et al. J Am Coll Cardiol 2005;46:2277-83

Cecchin F et al. JCE 2009;20:58-65

Janousek J et al. Heart 2009, 95:1165-71

CRT in systemic LV failure

LBBB or RV paced

Change after CRT	EF/FAC units mean (SD)	SVEDD z-score median	NYHA median
Upgrade from RV pacing, systemic LV (N=44)	+14.0 (16.5)	-2.1	Grade 3 → 1
All other pts (N=54)	+7.7 (10.9)	-0.8	Grade 2 → 2
P	=0.101	=0.036	=0.030

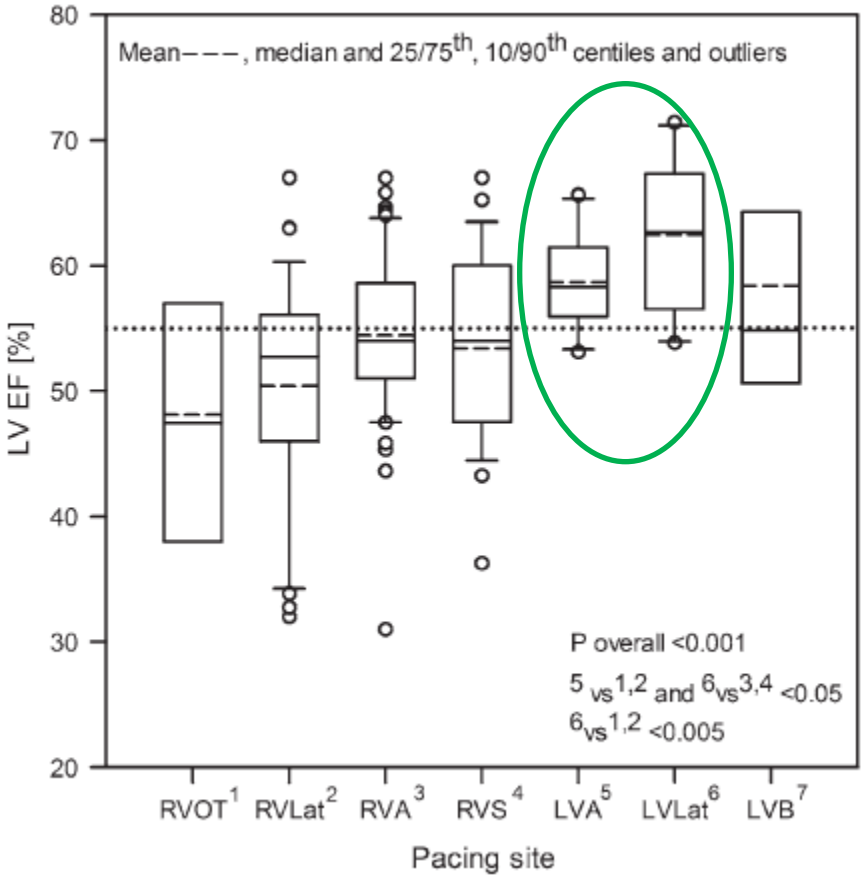
Janousek J et al. Heart 2009;95:1165-71

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION
American Heart Association®
Janousek J et al.
Circulation 2013;127:613-623

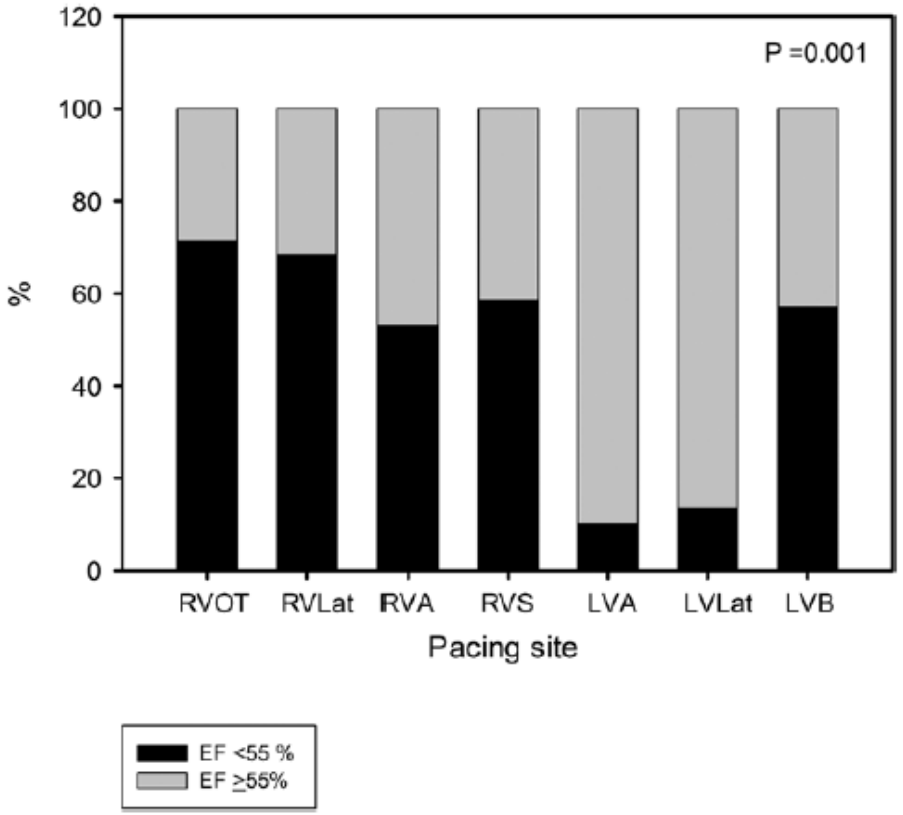
Permanent Cardiac Pacing in Children - Choosing the Optimal Pacing Site: A Multi-Center Study

(N=178, 21 centers)

LV ejection fraction at follow-up



Proportion of pts with LVEF<55 %



Systemic RV / Single-V

- More complex than just dyssynchrony
  - Intrinsic myocardial dysfunction
  - AV valve regurgitation
  - Fontan physiology
- Do not expect full reverse remodeling

Presence of a systemic left ventricle was the strongest multivariable predictor of improvement in EF/fractional area of change ($p<0.001$).

Janousek J et al. Heart 2009

CRT in systemic RV failure

RBBB or LV paced

Cardiac resynchronization therapy for adult congenital heart disease patients with a systemic right ventricle: analysis of feasibility and review of early experience

Gerhard-Paul Diller[1,2*], Darlington Okonko[2], Anselm Uebing[1,3], Siew Yen Ho[2], and Michael A. Gatzoulis[1,2]

Europace 2006

- CRT candidates according to current guidelines (QRS ≥120 ms, including NYHA class II pts)
  - TGA: 9.3%
  - ccTGA: 6.1%

CRT in systemic RV failure

RBBB or LV paced

N pts.

Jannousek J et al. Heart 2009, 95:1165-71

Patients' data	Systemic LV (n = 62)	Systemic RV (n = 27)	p Value*
Age at CRT (years), median	13.3	28.8	0.002
Follow-up on CRT (months), median	8.6	7.3	0.965
Initial QRS (ms), median	160	160	0.722
Initial SVEDD (z score), median	4.7	2.1	0.002
Initial EF/FAC (%), mean (SD)	30.6 (15.8)	28.8 (10.0)	0.723
Initial SAVV regurgitation (grade) (median)	1	2	0.025
Initial NYHA class (median)	3.0	2.0	0.215
Change in QRS (ms), median	−40§	−21§	0.877
Change in SVEDD (z score), median	−2.1§	−0.5	0.039
Change in EF/FAC (%), mean (SD)	+13.3 (14.7)§	+7.2 (9.9)§	0.195
Change in SAVV regurgitation (grade), median	−1§	−1§	0.600
Change in NYHA class (median)	−1.0§	−1.0‡	0.380
Non-responders	11/54	3/22	0.745

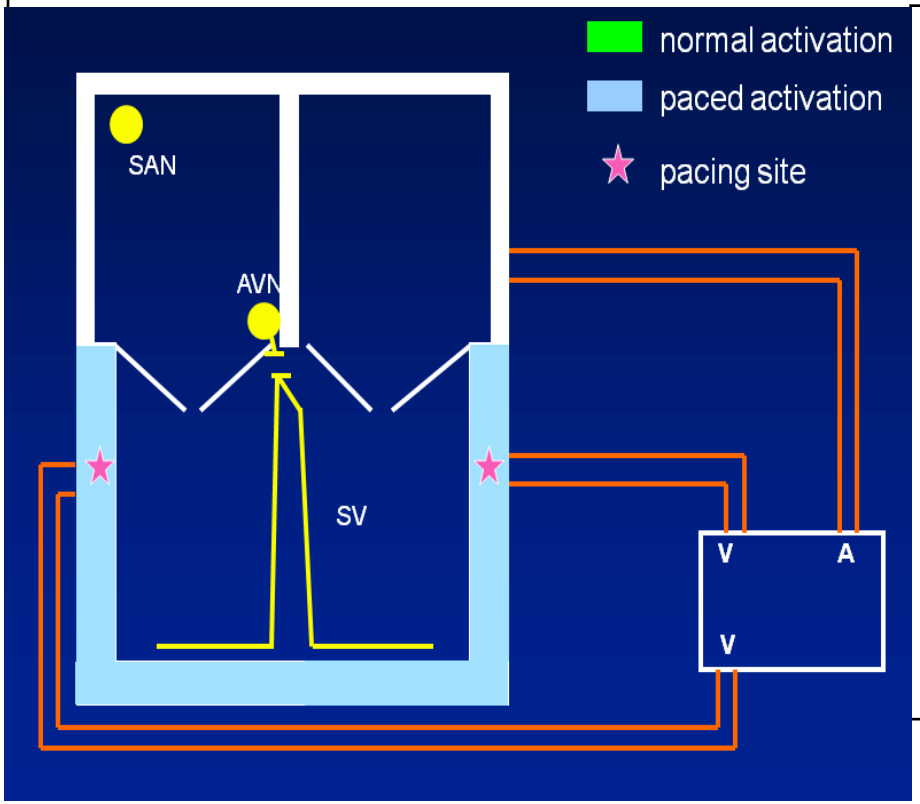
*Significance levels across diagnostic categories; significance levels inside diagnostic categories pre- vs post-CRT: ‡p<0.005; §p<0.001; ¶p<0.05.

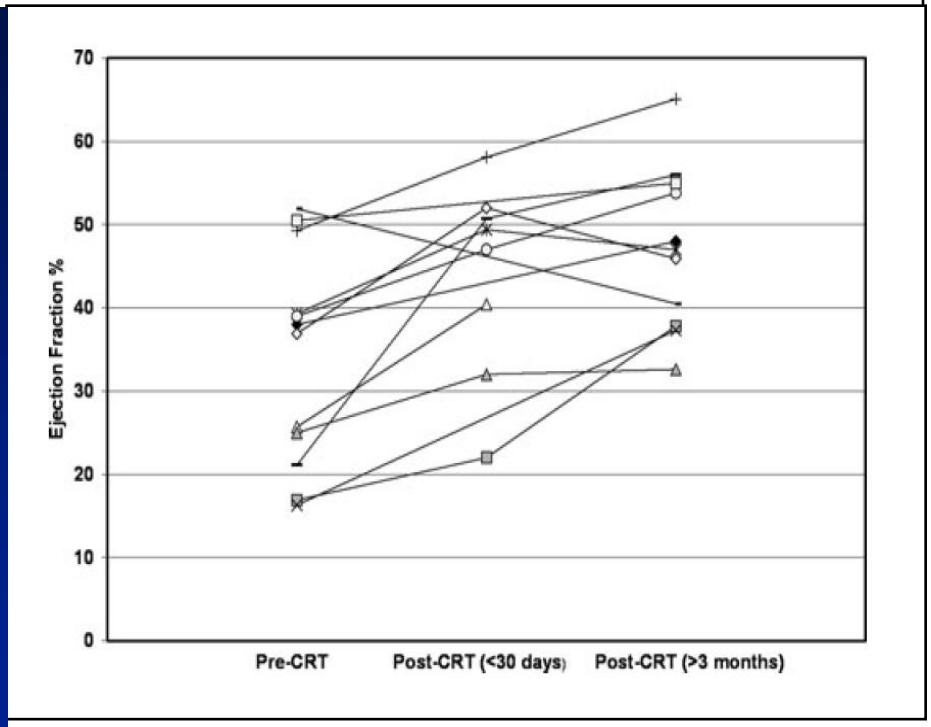
Dubin AM et al. J Am Coll Cardiol 2005;46:2277-83
Cecchin F et al. JCE 2009;20:58-65
Janousek J et al. Heart 2009, 95:1165-71

Resynchronizing the single ventricle

The complete AV block patient

Our approach to this group has evolved our time, but a high importance was placed on obtaining maximal distance between

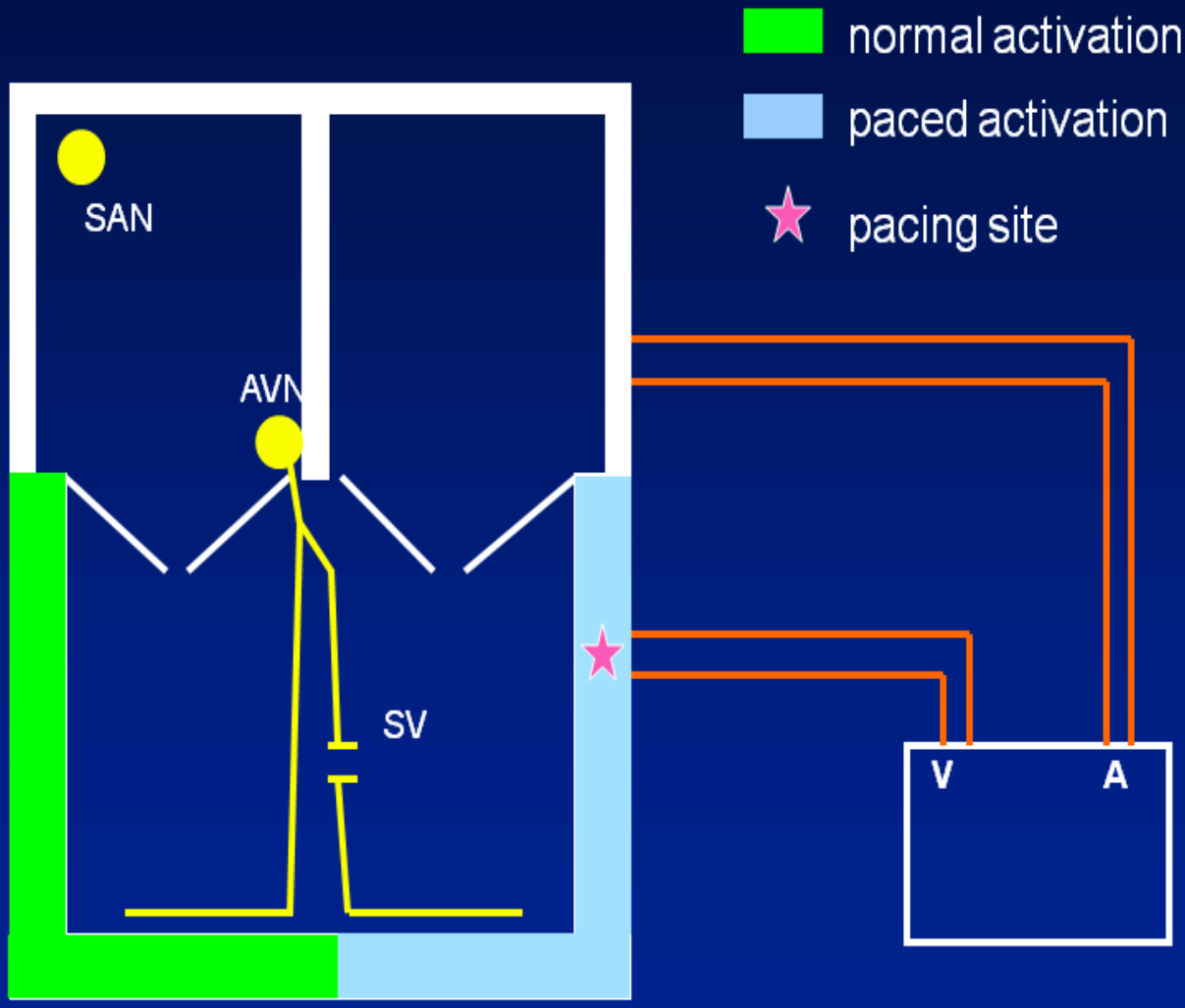




Cecchin F et al. JCE 2009;20:58-65

Resynchronizing the single ventricle

The bundle branch block patient



Single-site pacing in fusion with intrinsic activation

Testing of CRT effect prior to implantation

- May play a role in difficult to reach substrates
  - Systemic RV, functionally single ventricle

Table 2. Acute Hemodynamics Effects of CRT (in systemic RV)

Parameter	CRT Off Mean (SD)	CRT On Mean (SD)	% Change	p Value
QRS interval (ms)	161 (21)	116 (22)	−28.0	0.002†
Interventricular mechanical delay (ms)	median 60	median 50	−16.7	0.047‡
Dyssynchrony index (ms)	138 (59)	64 (21)	−53.6	0.042†
RV filling time (% RR)	45.1 (6.5)	50.0 (6.1)	10.9	0.002†
Tei index	median 0.65	median 0.60	−7.7	0.008‡
RV +dP/dt (mm Hg/s)	630 (142)	919 (211)	45.9	0.007†
Aortic VTI (cm)	17.2 (6.2)	18.4 (6.8)	7.0	0.028†
RV EF (%)*	41.5 (8.1)	45.5 (6.4)	9.6	0.04†

*Measured at a median of 3.8 months after initiation of CRT; †paired *t* test; ‡Wilcoxon signed rank test.
CRT = cardiac resynchronization therapy; EF = ejection fraction; RR = RR interval; RV = right ventricular; SD = standard deviation; VTI = velocity-time integral.

Janousek J et al. JACC 2004

Associated cardiac procedures

Will CRT improve the function so that I can replace the tricuspid valve in a Senning/CCTGA in the same procedure?

Patient data	All (n = 109)	CHD (CRT + concurrent cardiac surgery) (n = 16)
Age at CRT (years), median	16.9	13.9
Follow-up on CRT (months), median	7.5	4.0
Initial QRS (ms), median	160	160
Initial SVEDD (z score), median	3.3	2.6
Initial EF/FAC (%), median	27.0	24.5
Initial SAVV regurgitation (grade), median	1	0
Initial NYHA class (median)	2.5	2.0
Change in QRS (ms), median	−40§	−46§
Change in SVEDD (z score), median	−1.1§	−0.8
Change in EF/FAC (%), mean (SD)	+11.5 (14.3)§	+12.3 (17.1)¶
Change in SAVV regurgitation (grade), median	−1§	0
Change in NYHA class (median)	−1.0§	−1.0†
Non-responders	15/94	0/13

7/16 systemic AV valve replacement

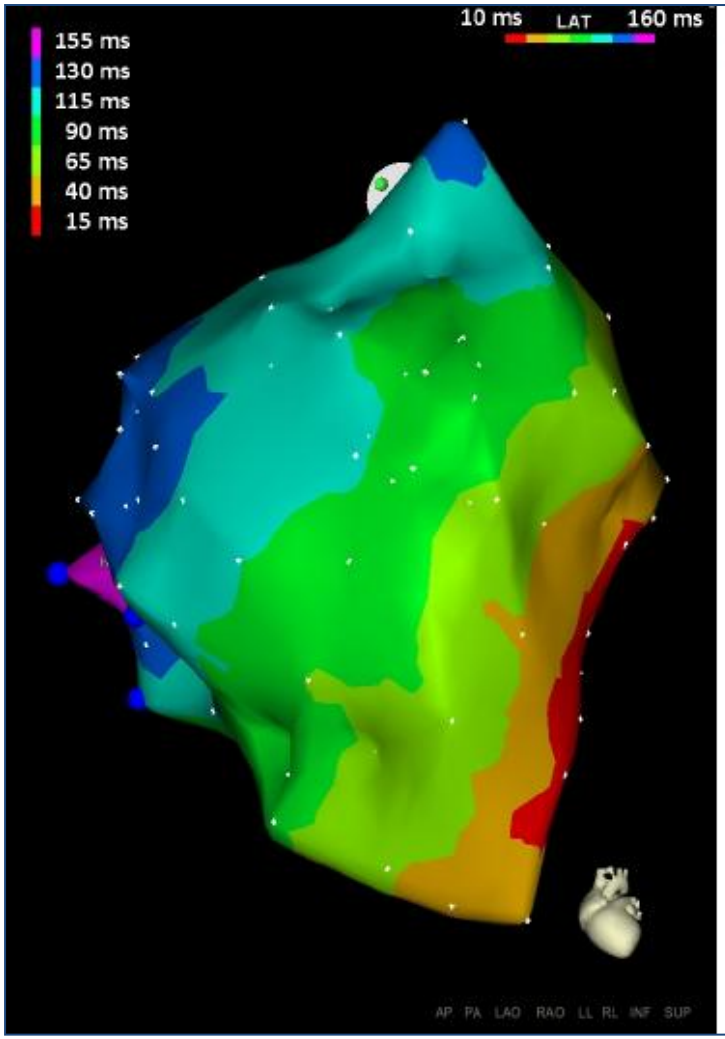
All survived

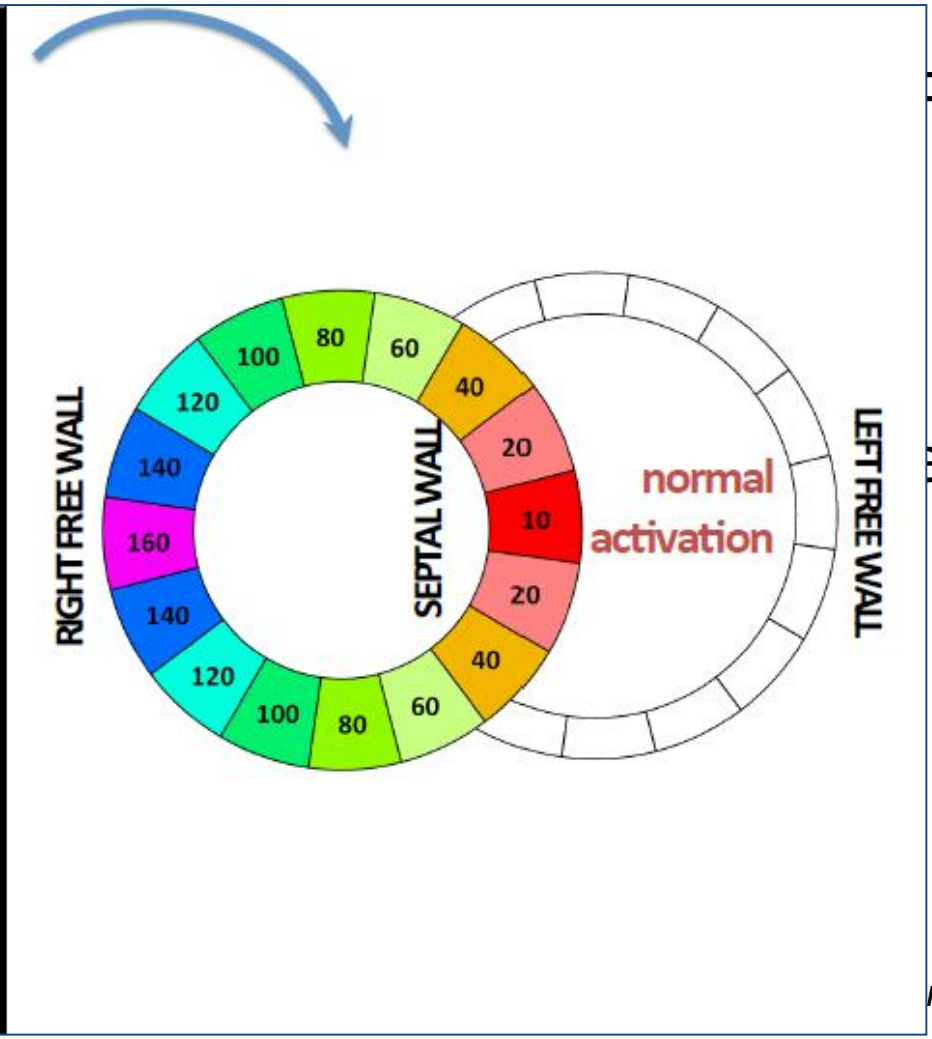
Janousek J et al. Heart 2009

Pulmonary RV-CRT

RBBB is by far the most frequent dyssychrony pattern in CHD!

- PVR
- e





r 2008

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association®

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

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

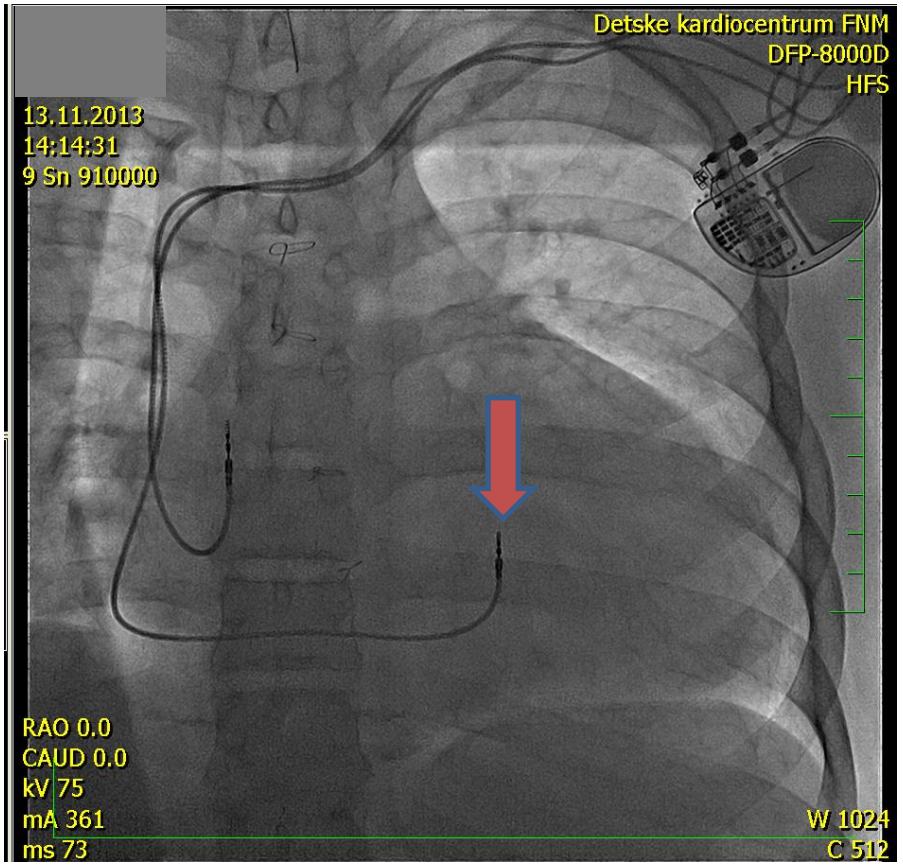
Circulation. 2014;130:e186-e190

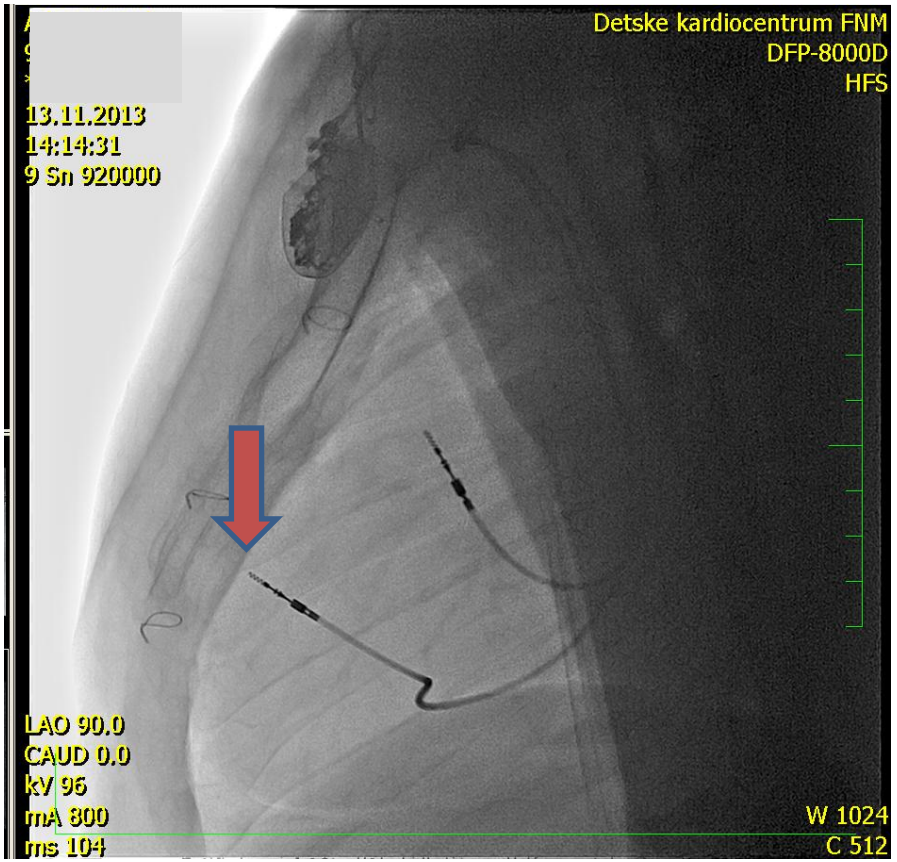
Case report, boy, age 17

- 12/1996 modified BT shunt
- 11/1997 trans-ventricular ToF repair
- 1/2000 RVOT aneurysm resection, PA plasty
- 10/2013 valvuloplasty of moderate PS
- 11/2013
  - Continuing RV dysfunction
  - No significant PR or TR
  - Low exercise tolerance and VO_2max
  - RBBB, QRS = 200 ms

MRI compatible pacemaker implantation

RV lead position: basal to mid-ventricular free wall at the border between inflow and outflow
q-RV interval = 140 ms



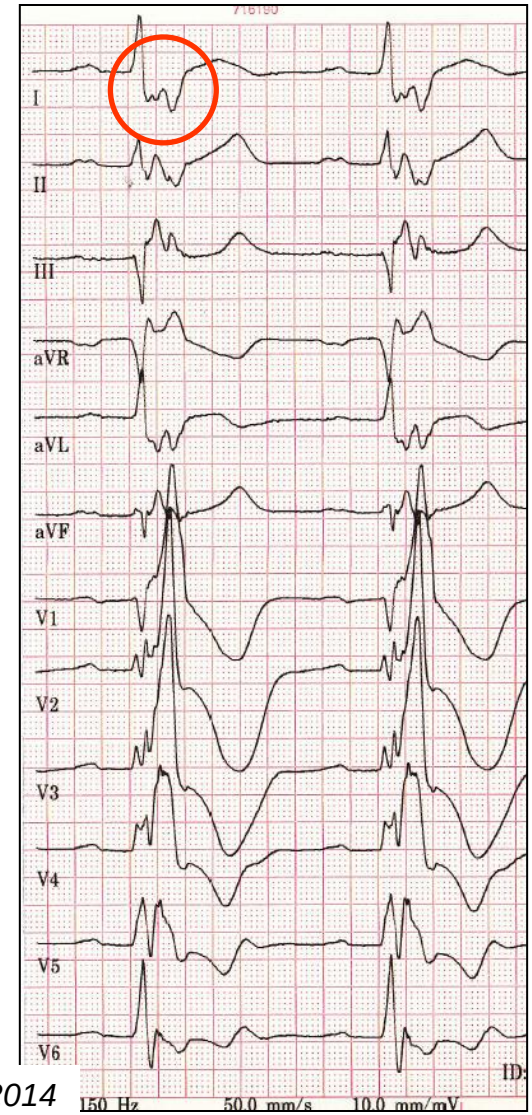


Kubuš P et al. Circulation 2014

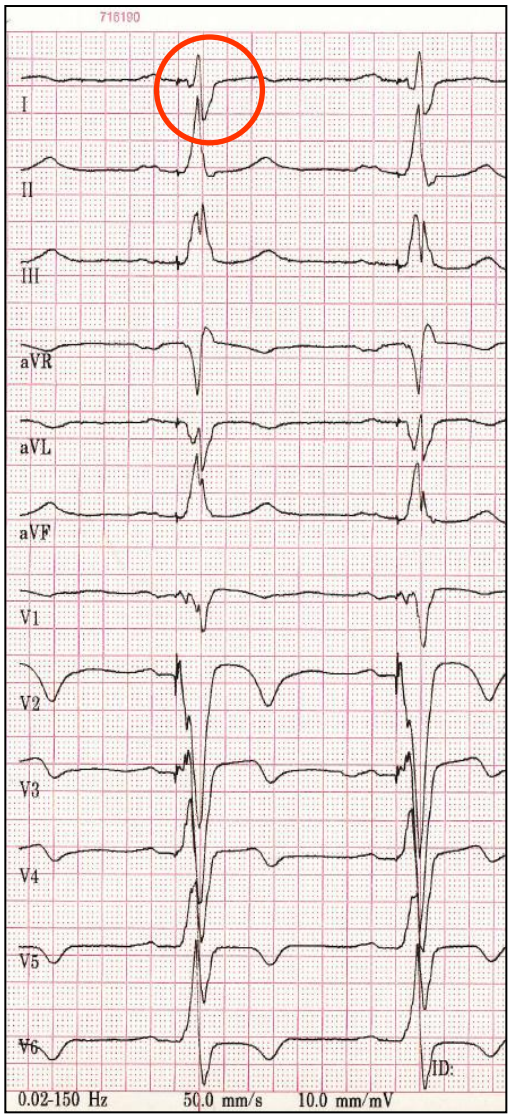
ECG

- **Before CRT**
  - **Sinus r.**
  - **CRBBB**

  (QRS 200 ms)



- **After CRT**
  - **Sinus r.**
  - **Complete fusion of paced and intrinsic activation**
  - **QRS 140 ms**



Kubuš P et al. Circulation 2014

Circulation
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Heart Association®

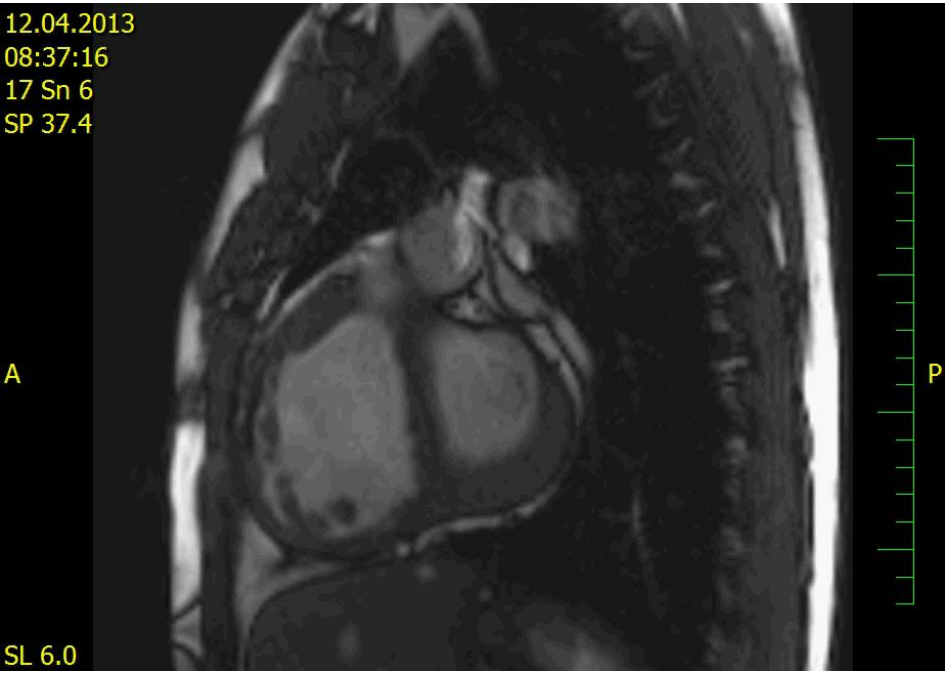
Circulation. 2014;130:e186-e190

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

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

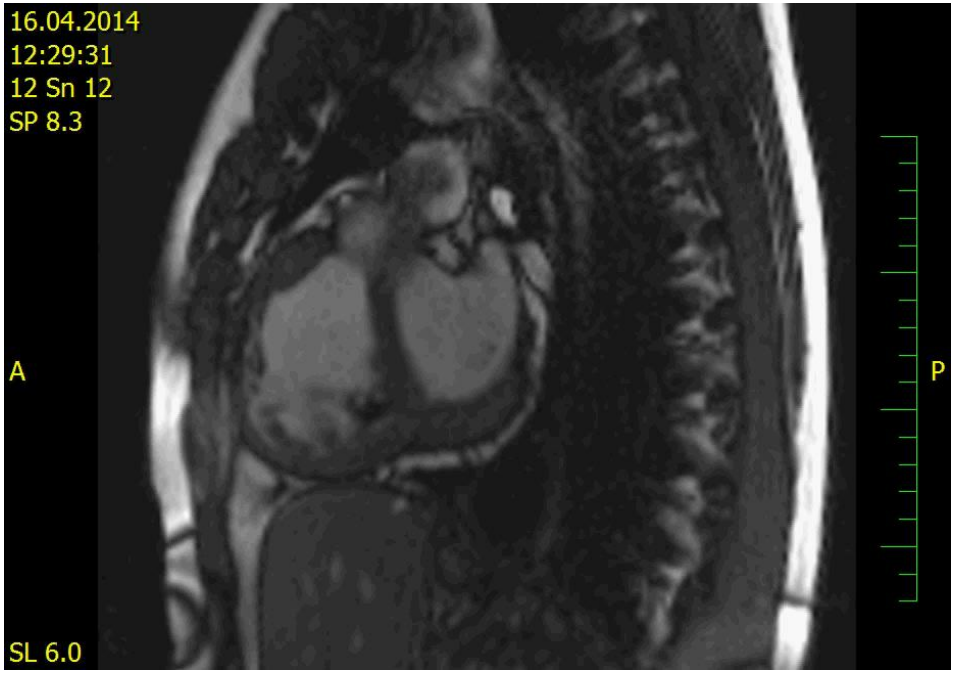
Before

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



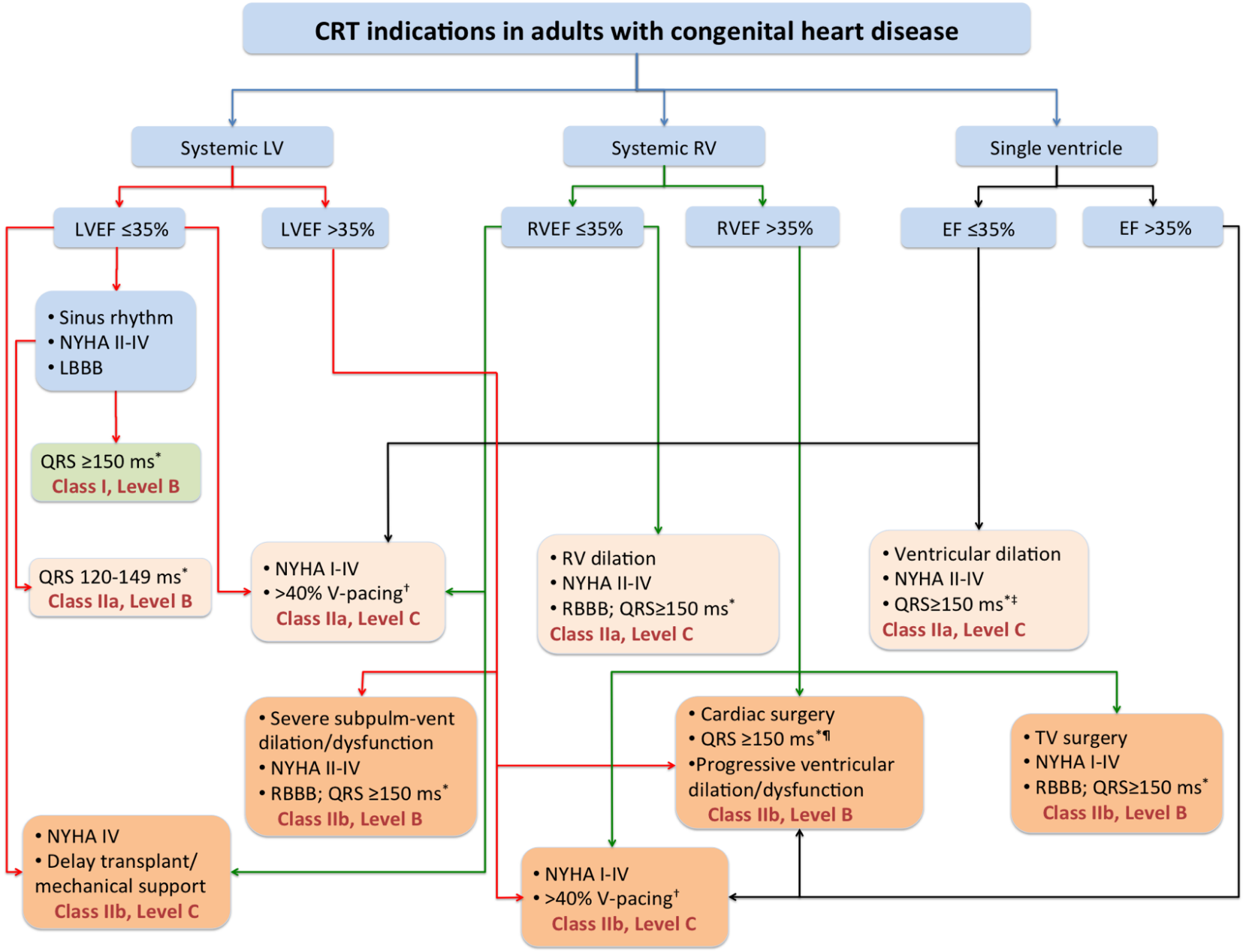
6 months after

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



Exercise stress testing - $V0_2$ max: 21,0 (before) ➡ 30,4 ml/kg/min. (6 mos of CRT)

NYHA II ➡ I



Khairy P et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. *HeartRhythm Journal 2014*

Summary

- CRT is back to it's origins
  - Major electrical activation delay is prerequisite for efficacy
- Most of paediatric CRT preventable
  - Proper conventional pacing
- CRT in systemic LV failure
  - Knowledge/results from adult idiopathic DCMP CRT studies transferrable
- CRT in other substrates
  - Specific approaches, evidence much smaller
- CRT in pulmonary RV failure
  - RV dyssynchrony may play a role in RV failure development
  - Indications?
- Long-term impact of CRT on survival and morbidity unknown
  - *Kubuš et al.: Long-term outcome of patients with congenital heart disease undergoing cardiac resynchronization therapy*