Resynchronization therapy in complex CHD, including systemic RV and single ventricle

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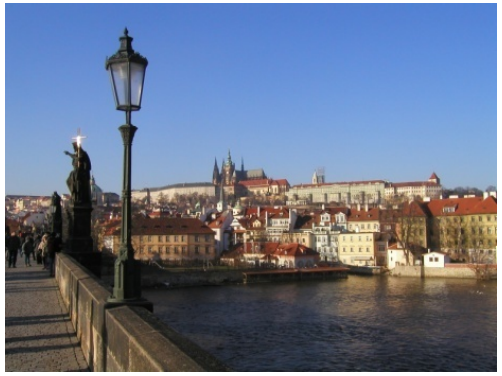
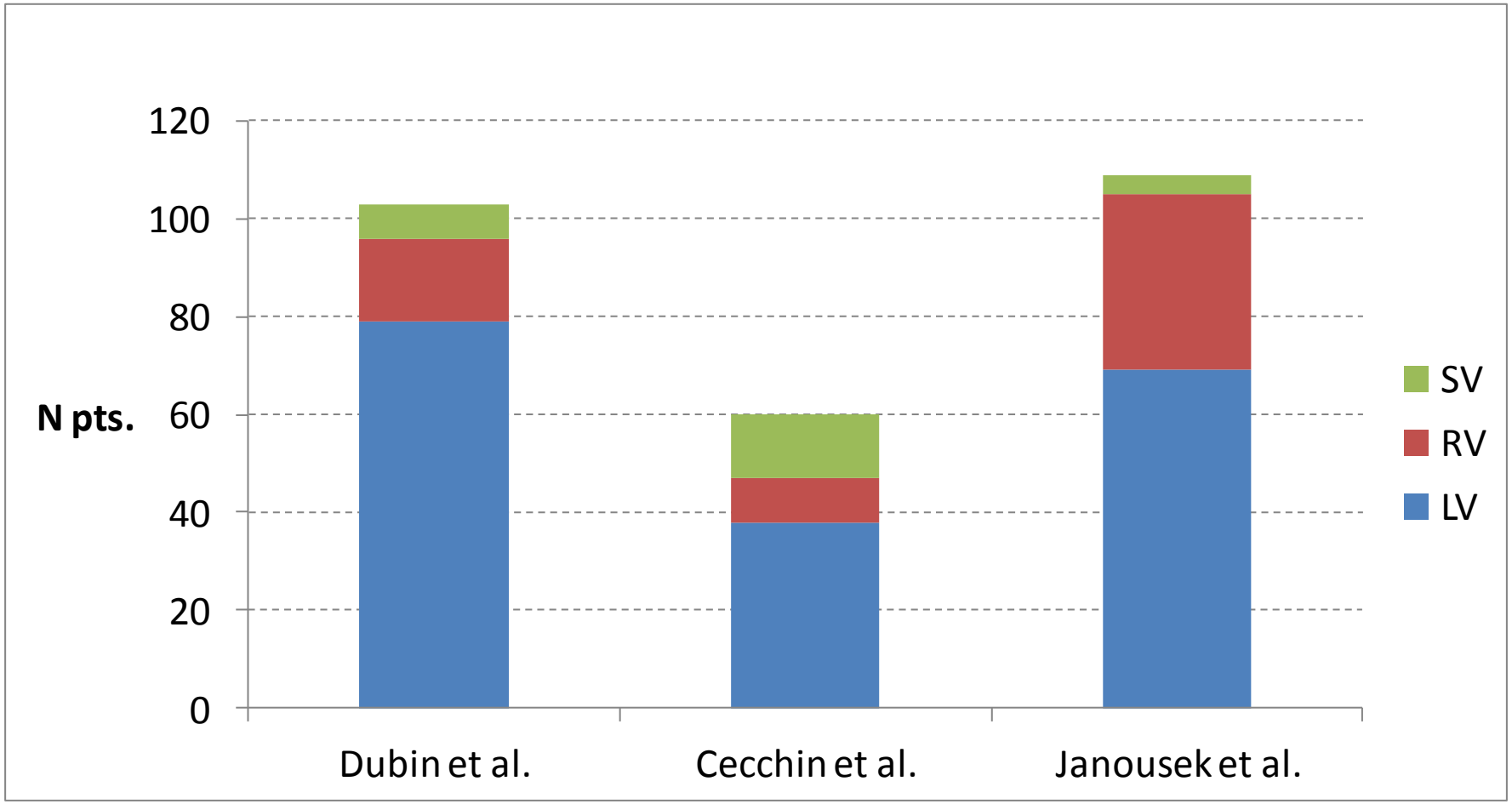


No relationships to disclose

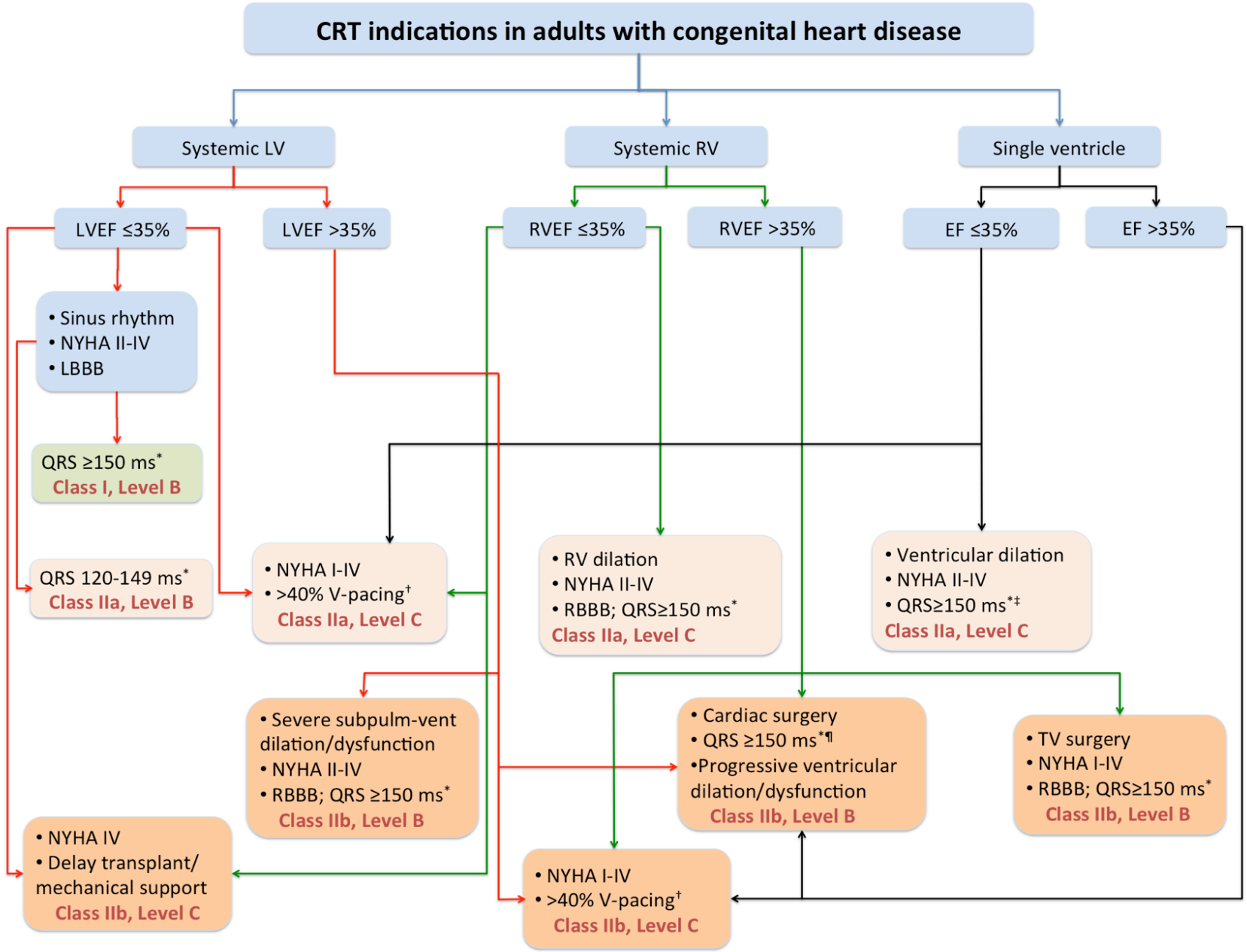
Proportion of complex CRT substrates



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



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

Pathophysiology

- More complex than just dyssynchrony
  - Intrinsic myocardial dysfunction
    - Suboptimal myocardial fiber arrangement
    - Decreased myocardial perfusion reserve
    - Contractile disparity rather than activation delay
  - AV valve regurgitation
  - Fontan physiology
- May prevent reverse remodeling

Is this dyssynchrony good for CRT?

- Look at QRS morphology and duration
  - Conduction delay within the failing ventricle!
- Do I need an ECHO study?
  - Yes for all atypical substrates
    - Degree of pathologic remodelling
    - Exclusion of scars
    - Confirmation of mechanical dyssynchrony
- What ist the golden parameter to look at?
  - None, put all pieces together

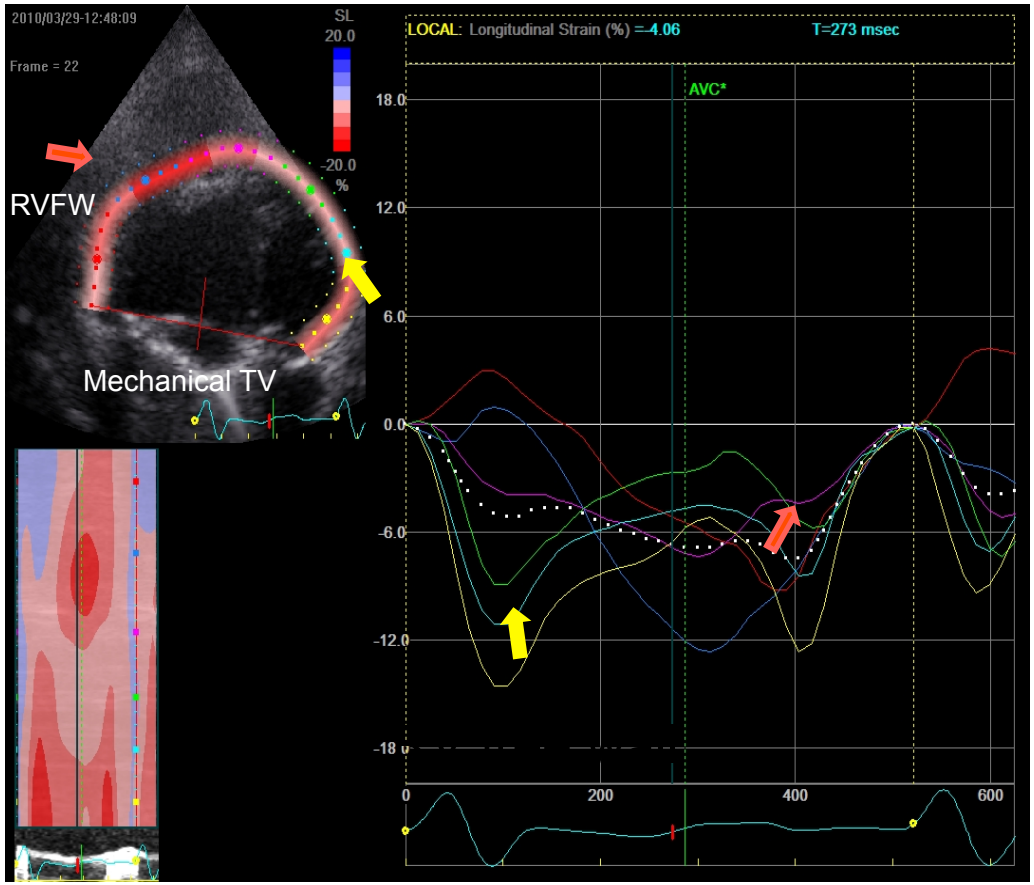
Where is the sweet pacing spot?

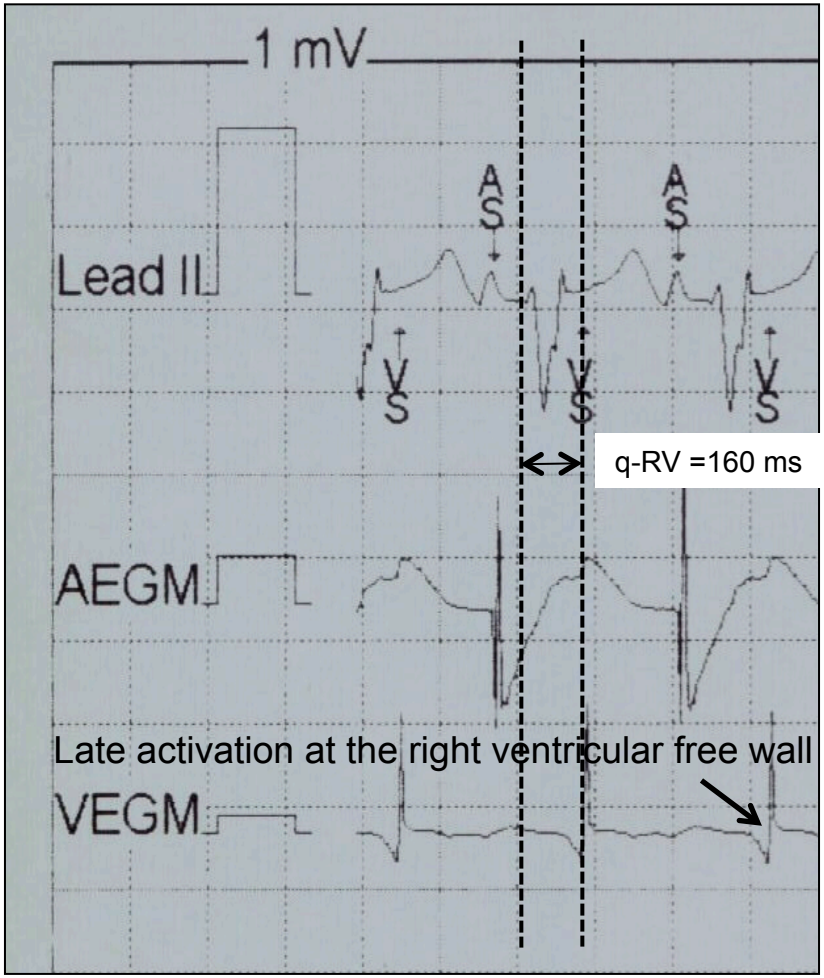
- Site of late activation
  - Electrical
    - Surface ECG (BBB QRS morphology)
    - Intra-operative activation mapping
  - Mechanical
    - Echo – speckle tracking
  - Should be concordant

Lead placement by mechanical/electrical activation mapping

HLHS, st.p. BCPA and TV replacement

Failing dyssynchronous RV due to RBBB





Materna et al. HeartRhythm J. 2014

Studies on CRT in systemic RV

Table 3. Studies That Reported Response to CRT in Patients With Systemic Right Ventricles

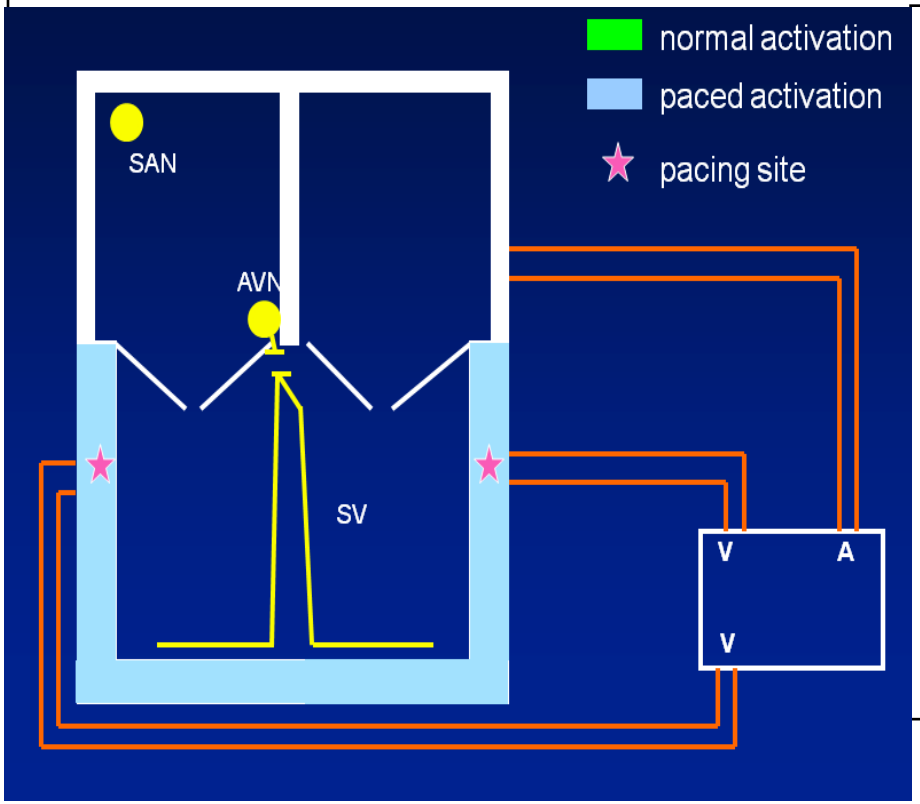
	Janousek et al,[37] 2004	Dubin et al,[44] 2005	Cecchin et al,[42] 2009	Janousek et al,[45] 2009	Jauvert et al,[41] 2009
Total patients with systemic RVs, n	8	17	9	27	7
Age (range), y	Median, 12.5 (6.9–29.2)	Median,12.7 (4.9-50)	Median,27 (0.5-43)	Median,28.8	Mean, 24.6 (15–50)
Follow-up duration (range), mo	Median, 17.4	Median, 4	Median, 8.4	Median, 7.3	Median, 19.4
CRT pacing method, n	7 BiV 1 multisite RV	BiV	BiV	26 BiV 1 single-site RV	BiV
Pre-CRT QRSd, ms	161±21	…	Median, 165	Median, 160	160±31
Pre-CRT sysV EF, %	…	…	Median, 28	28.8±10	…
Pre-CRT NYHA FC	Mean, 2	…	…	Median, 2	Mean, 3
Outcomes after CRT					
Change in QRSd, ms	↓ 45 (mean)	↓ 38.2±29.4 (mean±SD)	↓ 15 (median)	↓ 21 (median)	120±28 (mean±SD)
Change in sysV EF units	↑ 4 (mean)	↑ 13.3±11.3 (mean±SD)	↑ 14 (median)	↑ 7.2±9.9 (mean±SD)	…
NYHA improvement	Mean, ↓ 0.7 FC	…	…	Median, ↓ 1 FC	Mean, ↓ 1.4 FC
Clinical improvement, n (%)	8/8 (100)	13/17 (76.5)	2/8 (25)	19 (86.4)	7 (100)
Nonresponders (%N)	…	4/17 (23.5)	6/8 (75)	3/22 (13.6)	…

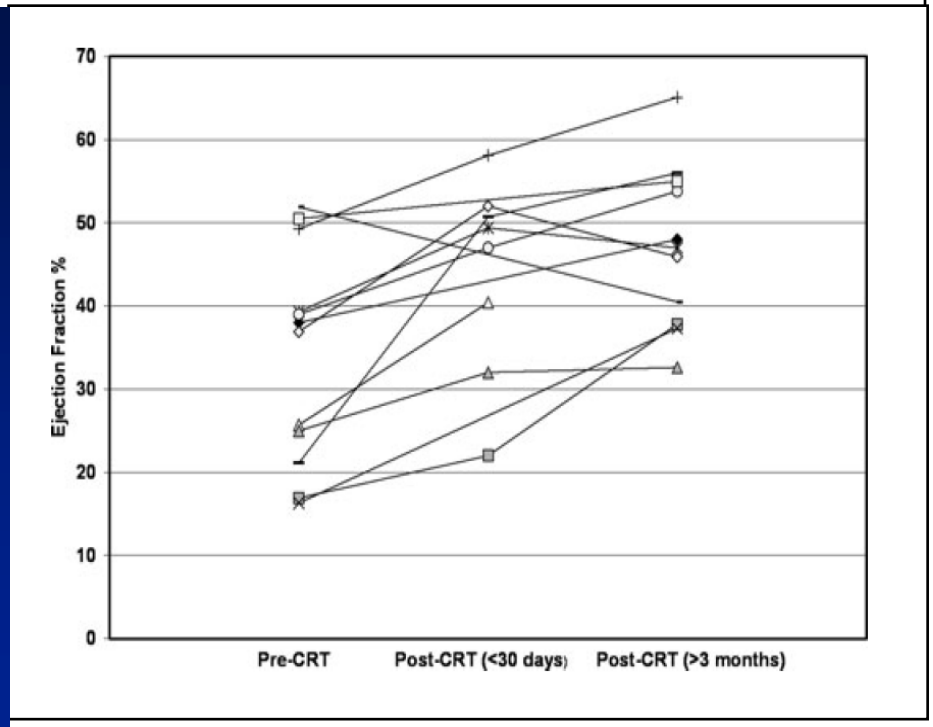
BiV indicates biventricular; CRT, cardiac resynchronization therapy; EF, ejection fraction; FC, functional class; NYHA, New York Heart Association; QRSd, QRS duration; RV, right ventricle; and sysV, systemic ventricle.

Motonaga, KS et al. Circulation 2014

Resynchronizing the single ventricle

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

Studies on CRT in single V

Table 4. Studies That Reported Response to CRT in Patients With Single Ventricles

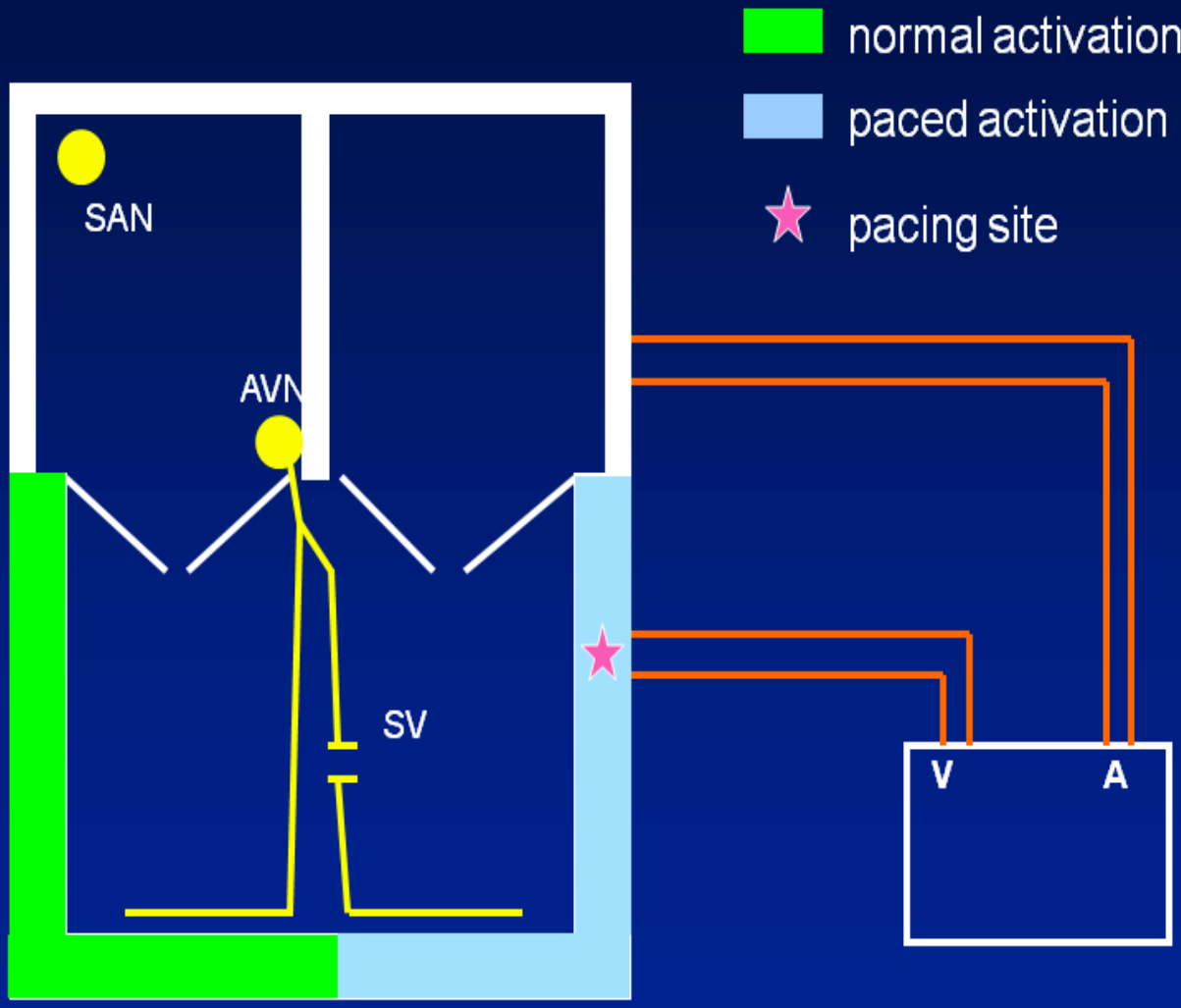
	Dubin et al,[44] 2005	Cecchin et al,[42] 2009	Janousek et al,[45] 2009
Total patients single ventricles, n	7	13 but only 11 with >3 mo of follow-up	4
Median age (range)	3.1 y (5 mo–23.7 y)	17.3 y (0.5–42.5 y)	10.3 y (3.7–30.3 y)
Conventional pacing before CRT, n (%)	…	8 (61.5)	3 (75)
Median pre-CRT QRSd, ms	…	129	…
Median pre-CRT EF, %	…	37	…
Outcomes after CRT			
Change in QRSd, ms	↓ 44.8±26.2 (mean)	↓ 13 (median)	…
Change in EF units	No change	↑ 11 (median)	…
Clinical improvement, n (%)	2 (28.6)	10 (90.9)	3 (75)
Nonresponders, n (%)	5 (71.4)	1 (9.1)	1 (25)

CRT indicates cardiac resynchronization therapy; EF, ejection fraction; and QRSd, QRS duration.

Motonaga, KS et al. Circulation 2014

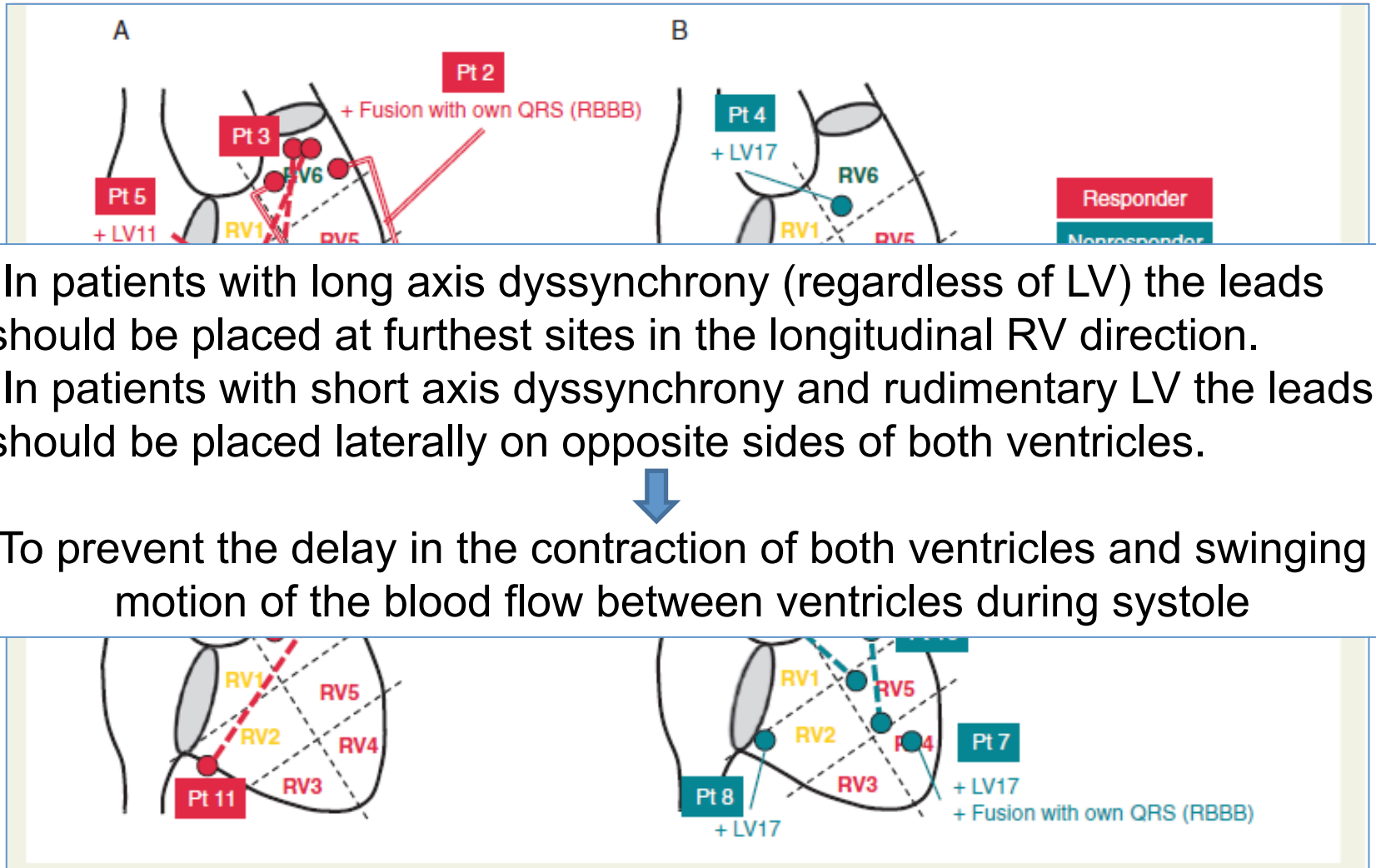
Resynchronizing the single ventricle

The bundle branch block patient



Single-site pacing in fusion with intrinsic activation

Europace (2016) **18**, 100–112
doi:10.1093/europace/euu401



- In patients with long axis dyssynchrony (regardless of LV) the leads should be placed at furthest sites in the longitudinal RV direction.
- In patients with short axis dyssynchrony and rudimentary LV the leads should be placed laterally on opposite sides of both ventricles.

To prevent the delay in the contraction of both ventricles and swinging motion of the blood flow between ventricles during systole

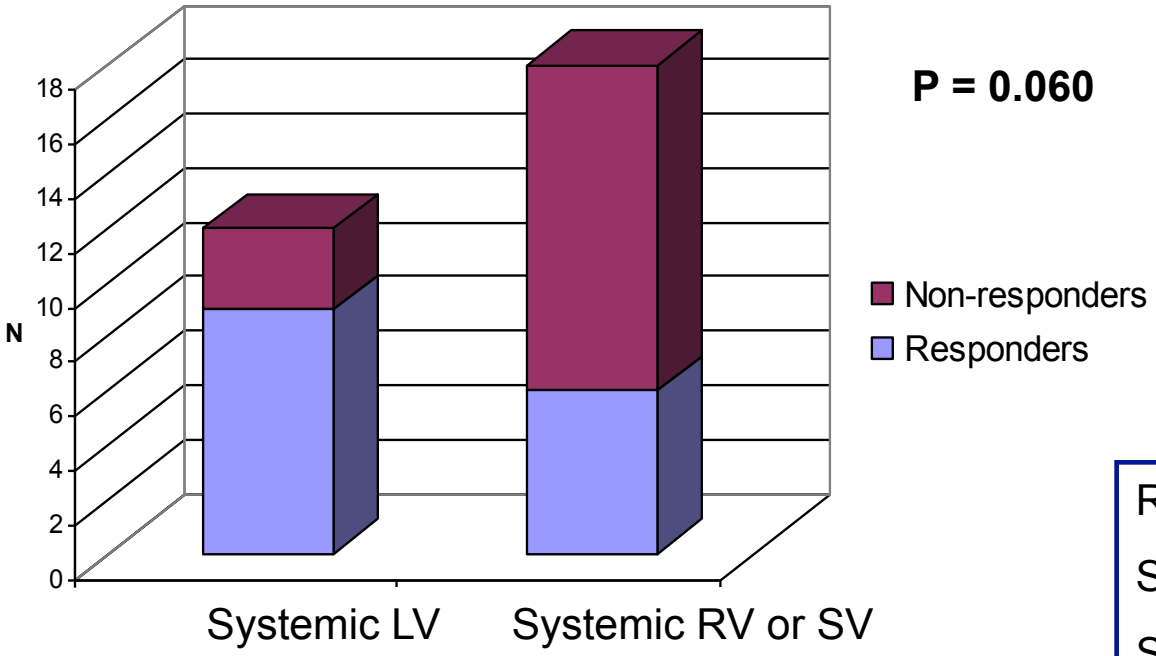
Figure 4 Lead positions. Cardiac resynchronization therapy responders are represented by red squares and nonresponders by turquoise squares. The patient numbers are as in *Table 1*. The dotted lines indicate the procedures that resulted in LD of the RV before the CRT, thick solid lines indicate those that resulted in SD between the rLV and RV, double lines indicate those that resulted in both, and thin solid lines indicate those that resulted in neither. (*A*) Responders with an sRV and rLV. (*B*) Nonresponders with an sRV and rLV. (*C*) Responders with an sRV without an rLV. (*D*) Nonresponders with an sRV without an rLV. The abbreviations are as in *Table 1* and *Figure 1*.

Long-term outcome of patients with congenital heart disease undergoing cardiac resynchronization therapy

Peter Kubuš (1), Jan Rubáčková Popelová (2), Jan Kovanda (1), Kamil Sedláček (3), Jan Janoušek (1)

- N=30
- Age at CRT implantation: median 12.9 (IQR 6.5-18.2) yrs
- Follow up: median 9.0 (IQR 4.5-11.4) years on CRT

Long term CRT response



P = 0.060

Responders (15/30 = 50 %):

Systemic LV = 9/12

Systemic RV or SV = 6/18

Testing of CRT effect prior to implantation

- May play a role in difficult substrate

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

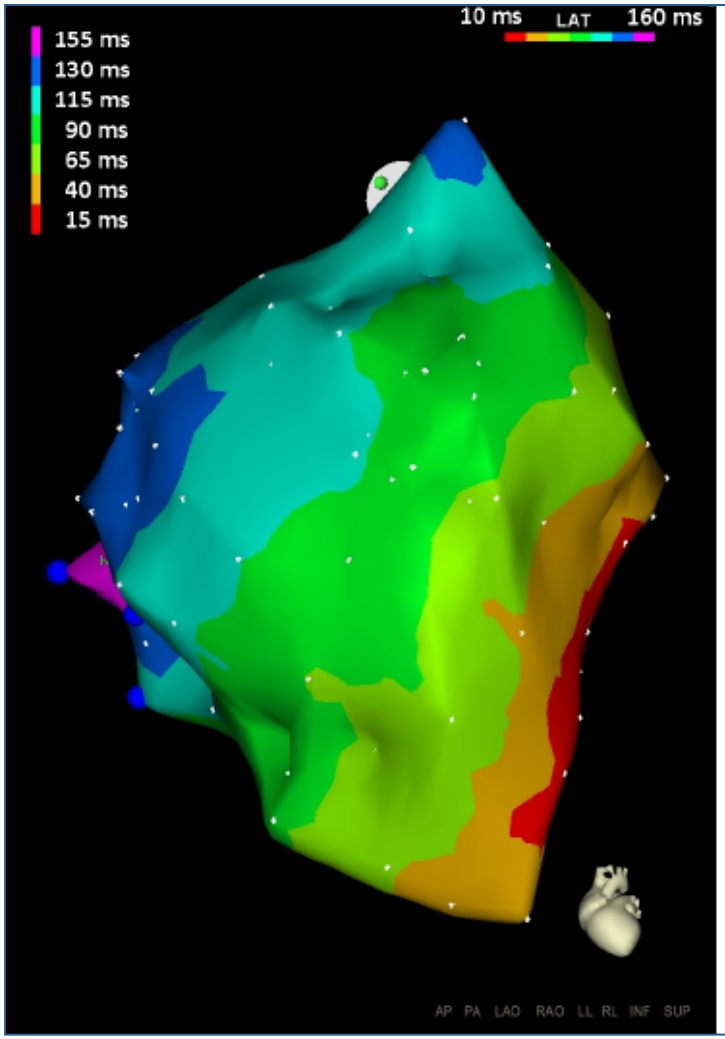
Parameter	CRT Off Mean (SD)	CRT On Mean (SD)	% Change	p Value
QRS interval (ms)	161 (21)	116 (22)	−28.0	0.002†
Interventricular mechanical delay (ms)	median 60	median 50	−16.7	0.047‡
Dyssynchrony index (ms)	138 (59)	64 (21)	−53.6	0.042†
RV filling time (% RR)	45.1 (6.5)	50.0 (6.1)	10.9	0.002†
Tei index	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†

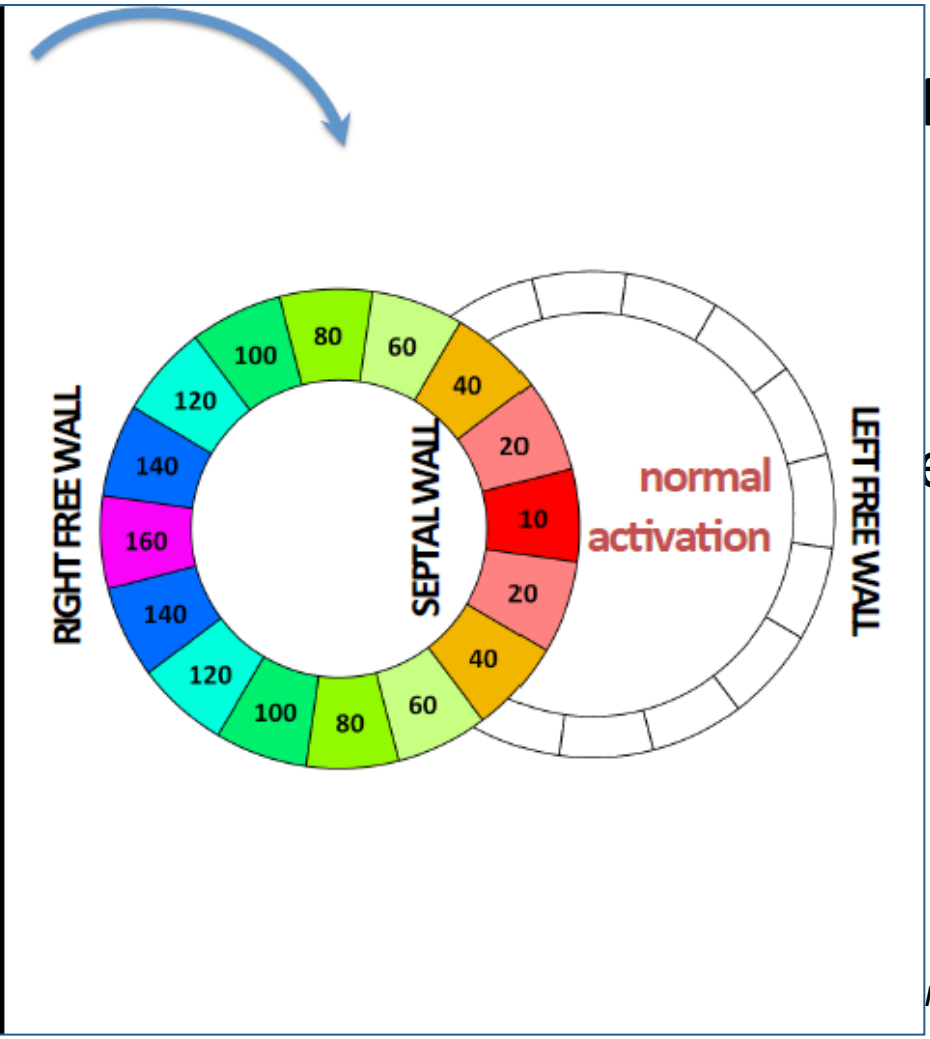
*Measured at a median of 3.8 months after initiation of CRT; †paired *t* test; ‡Wilcoxon signed rank test.
CRT = cardiac resynchronization therapy; EF = ejection fraction; RR = RR interval; RV = right ventricular; SD = standard deviation; VTI = velocity-time integral.

Janousek J et al. JACC 2004

Resynchronizing the pulmonary RV?

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





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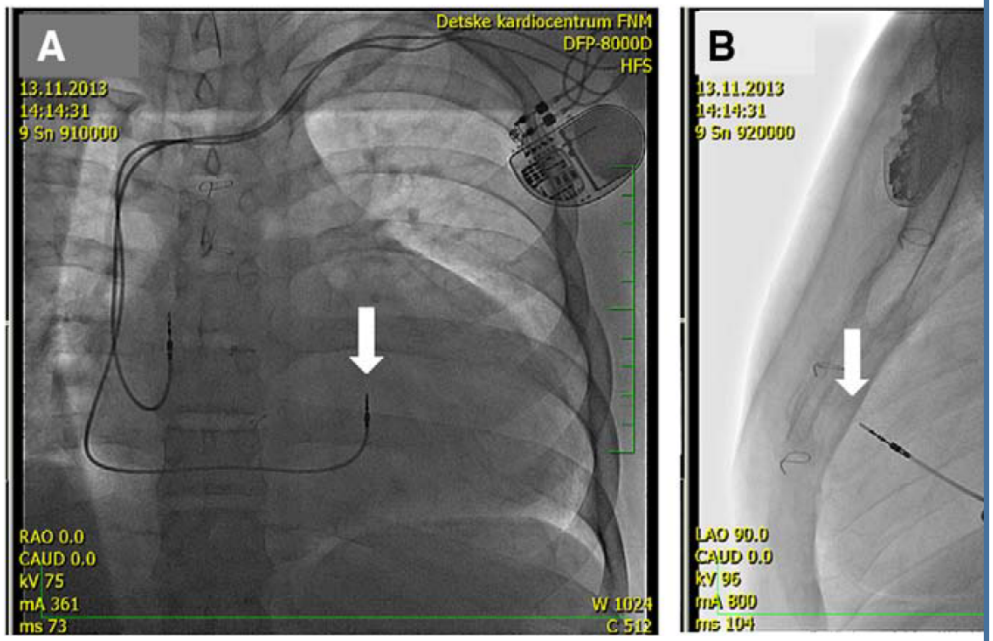
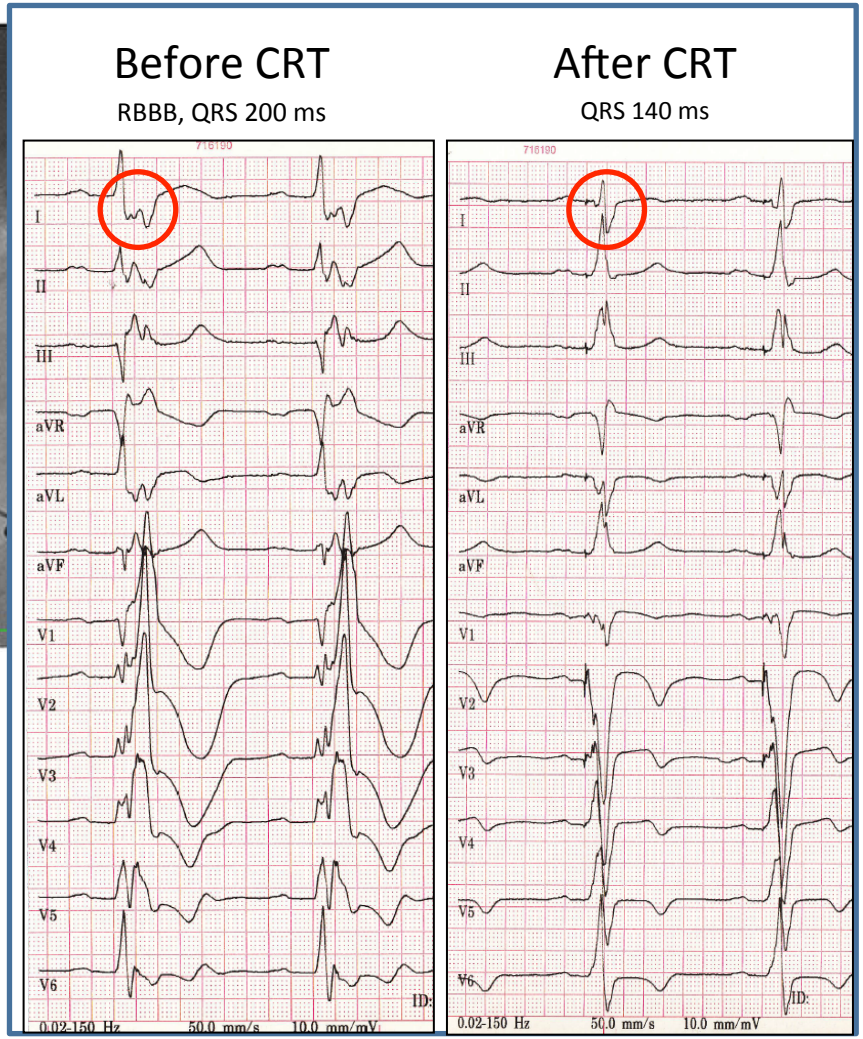
Successful Permanent Resynchronization for Failing Right Ventricle After Repair of Tetralogy of Fallot

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





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Circulation. 2014;130:e186-e190

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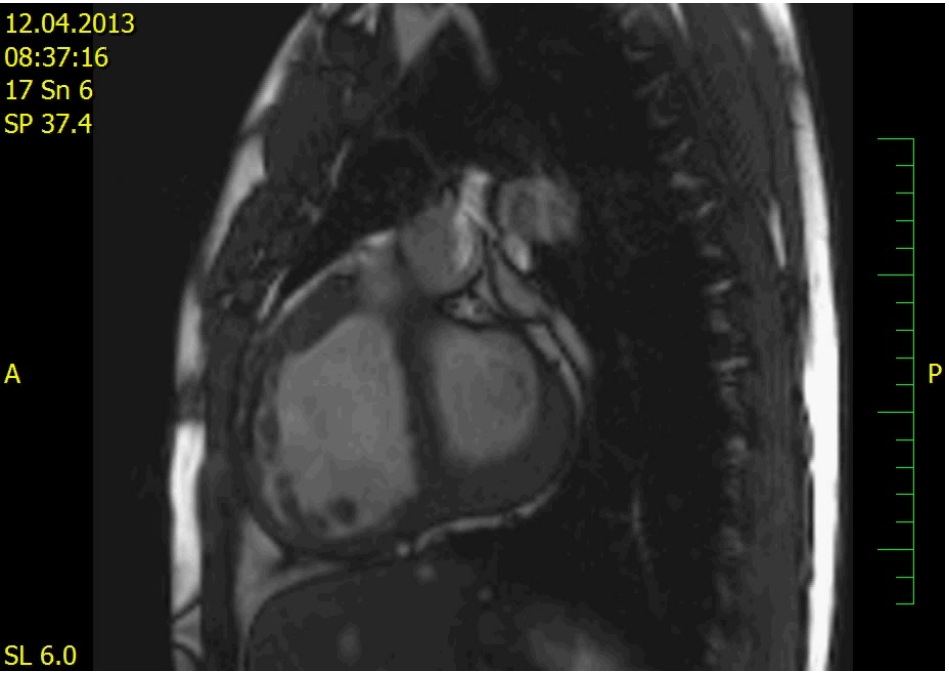
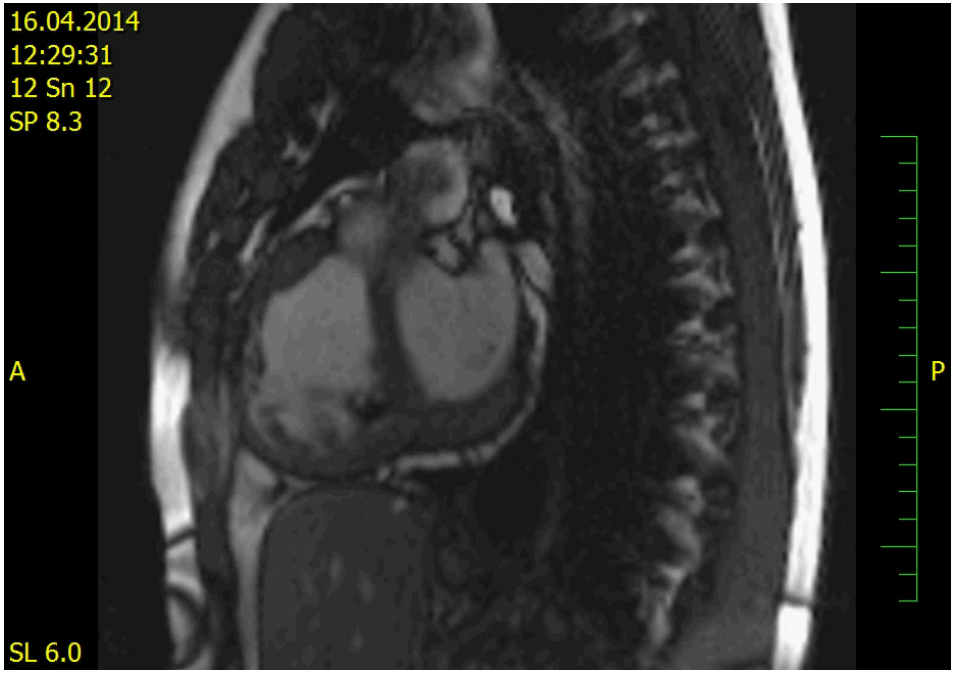
Before

- 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 %

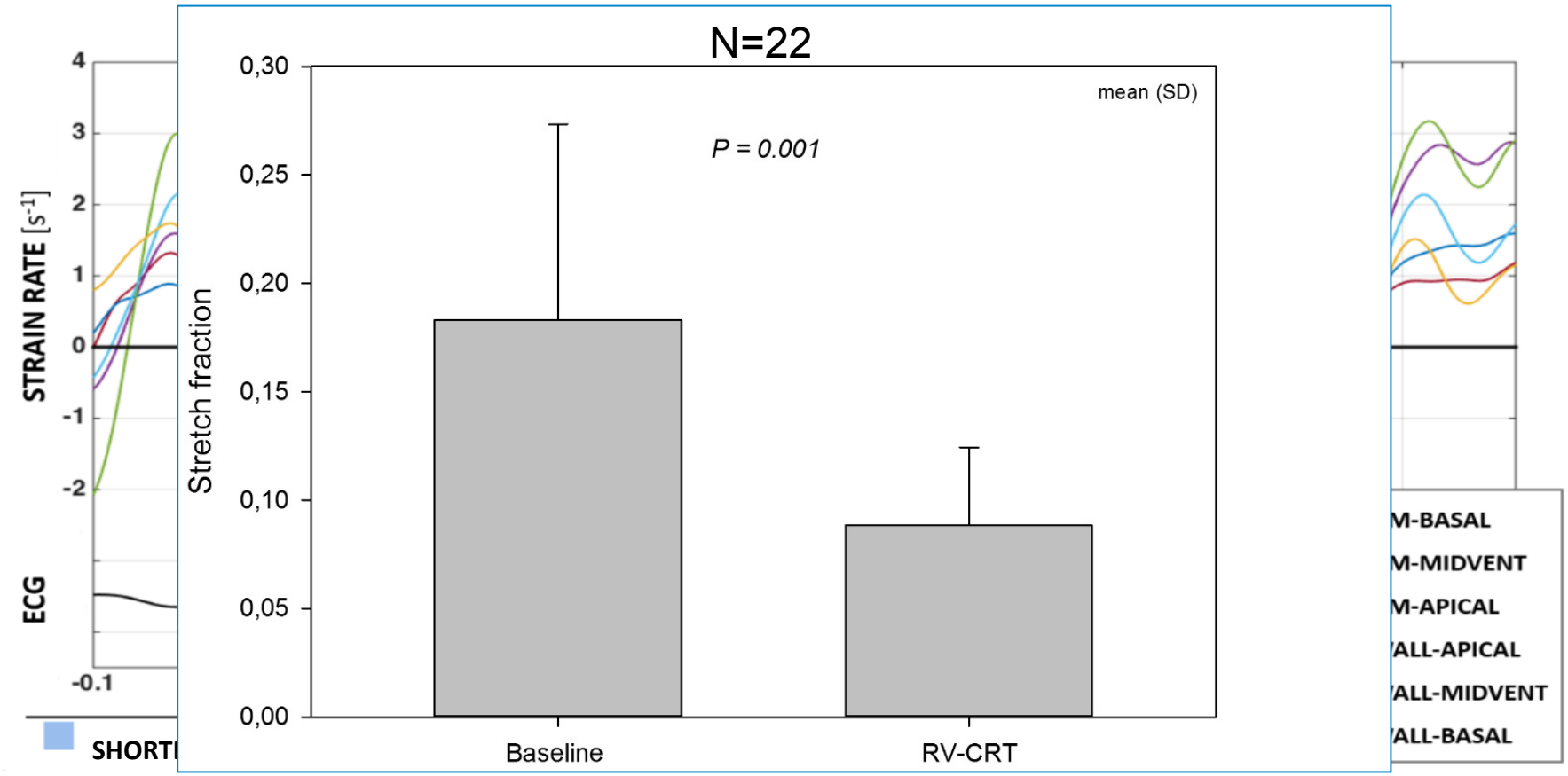


Exercise stress testing - $V0_2$ max: 21,0 (before) ➡ 30,4 ml/kg/min. (6 mos of CRT)

NYHA II ➡ I

Contraction efficiency increased by RV-CRT

Systolic stretch fraction calculation



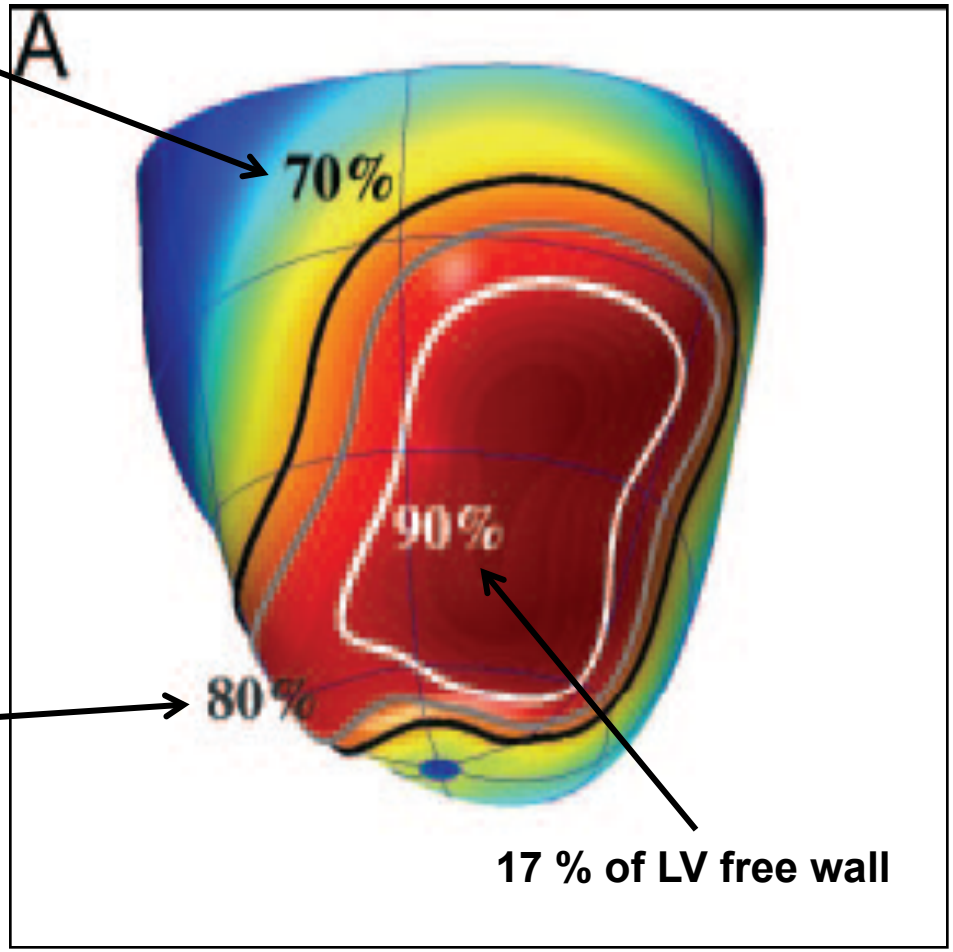
Kovanda J et al., HRS meeting 2016

Can we Really Resynchronize the Single Ventricle or (Systemic) Right Ventricle?

- Yes, at least some
  - If in doubt, check acute CRT response (dP/dt)
  - Other myocardial pathologies may decrease CRT effect
- Impact on mid/long-term survival unknown
  - Long-term follow-up data/registries needed
- Preliminary results seem to open a space for pulmonary RV-CRT
- Terminal heart failure associated with dyssynchrony
  - CRT vs Mechanical support vs HTx listing vs Decreased likelihood being transplanted (adult single-V patients)

How precise do we have to place the leads?

Percentage of max. CRT response related to area of lead placement



Helm RH, Circulation 2007