ČASR ČESKÁ ASOCIACE PRO SRDEČNÍ RYTMUS
ČESKÁ KARDIOLOGICKÁ SPOLEČNOST

Srdeční resynchronizační léčba u vrozených srdečních vad

J. Janoušek

Dětské kardiocentrum 2. LF UK a FN Motol

Praha, Česká Republika

Kardiocentrum

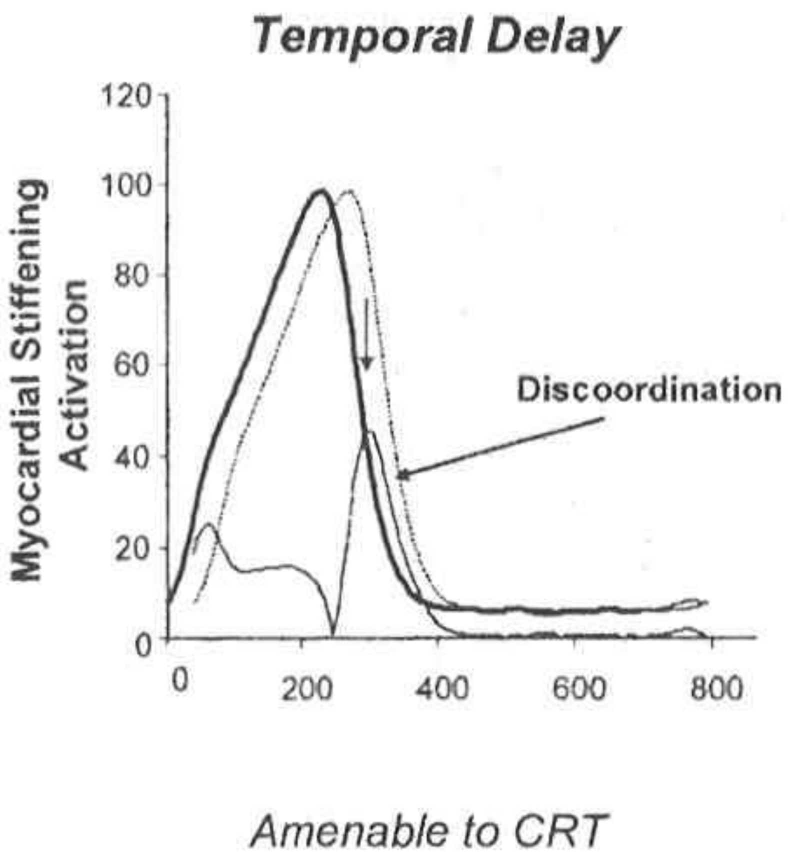
Indications for CRT in CHD

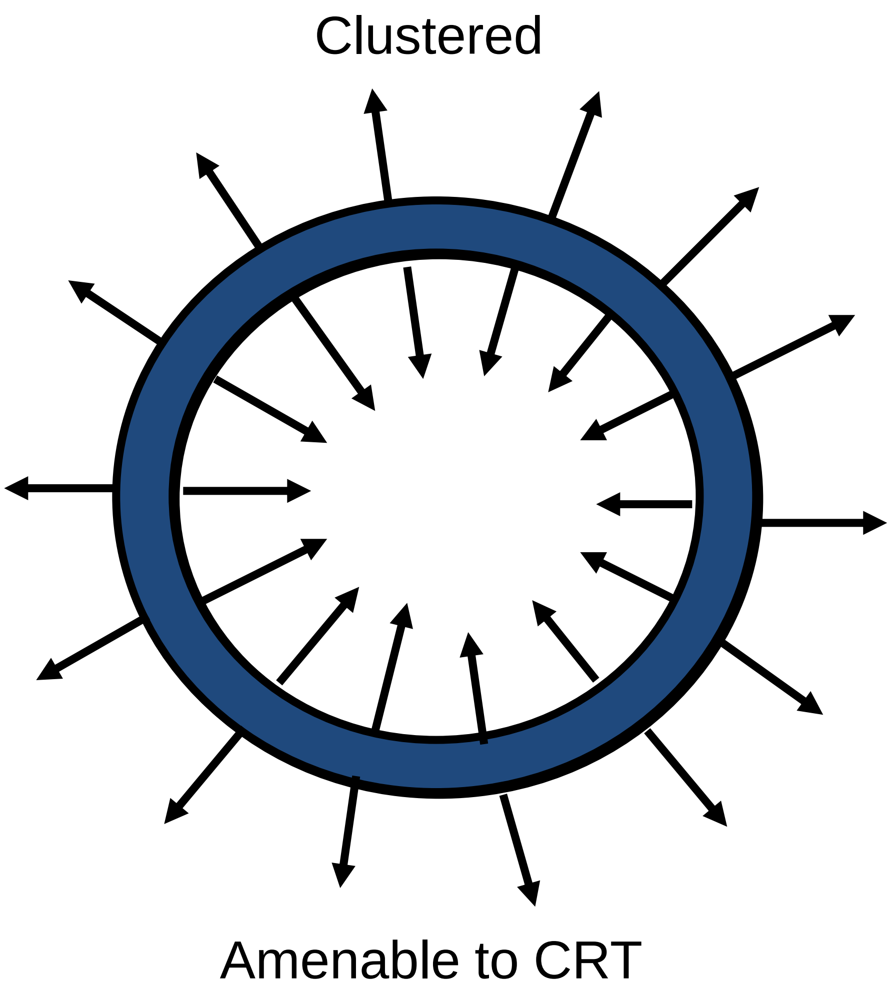
- Systemic LV failure → The well-studied setting
  - Left bundle branch block
  - RV paced
- Systemic RV failure
  - Right bundle branch block
  - LV paced
- Single-ventricular failure
  - Any bundle branch block
  - Single site pacing
- Pulmonary RV failure?
  - Right bundle branch block

} Specific for CHD

Pathophysiological substrate:

Clustered dyssynchrony due to temporal activation delay

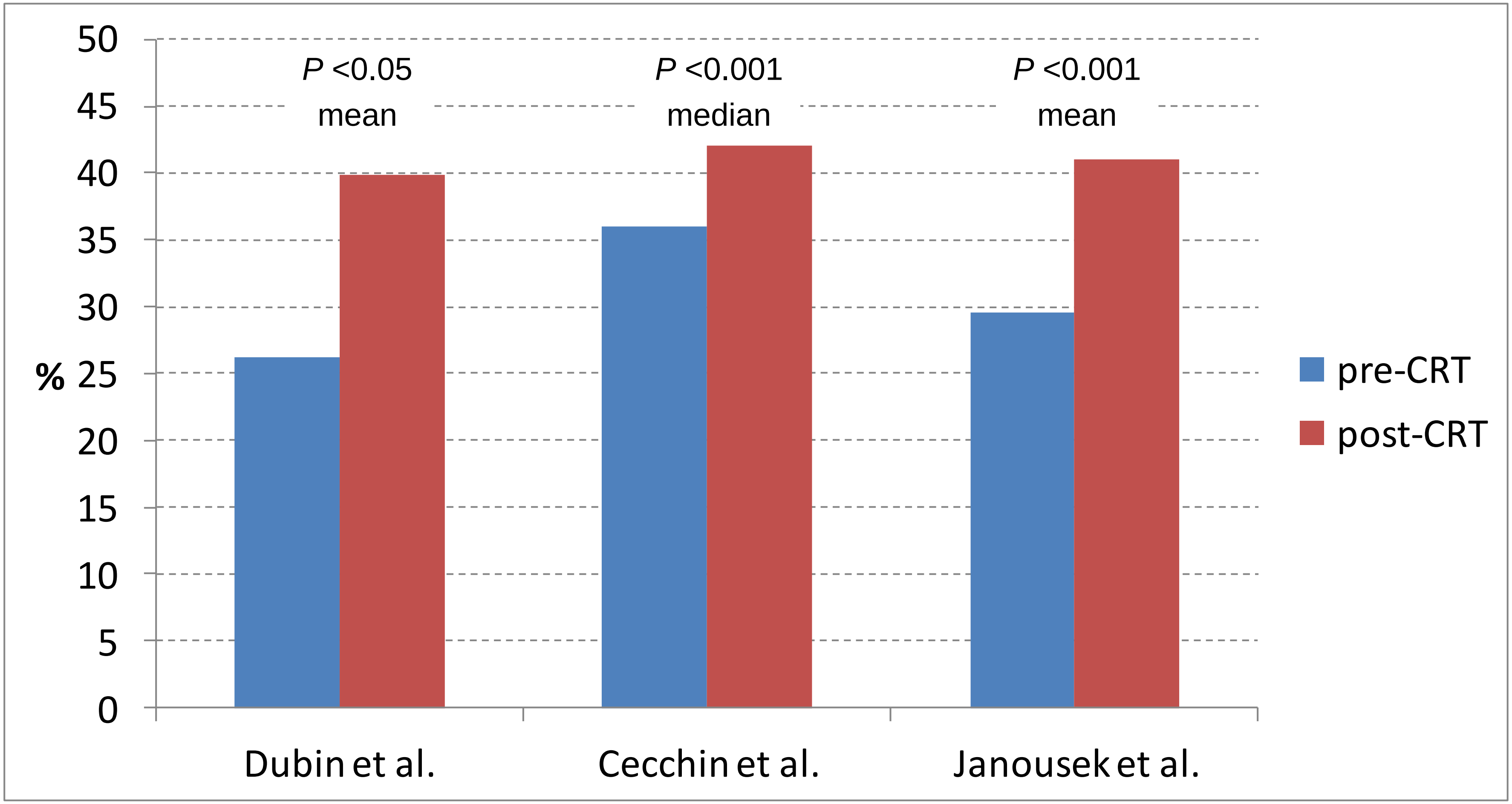




Kass DA, JACC 2008

Helm RH et al.,Circulation 2005

Systemic ventricular ejection fraction in CRT studies in CHD

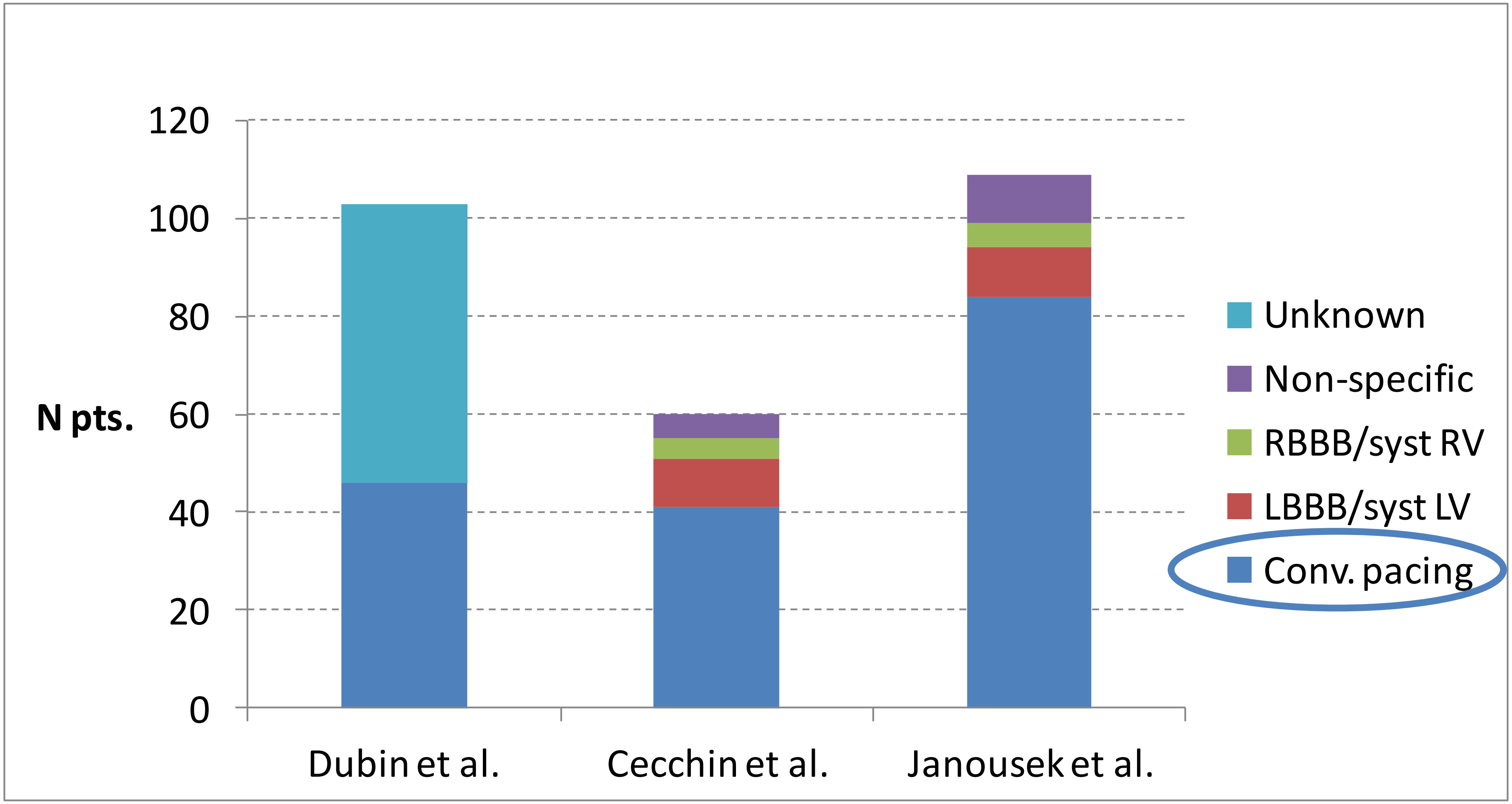


Dubin AM et al. J Am Coll Cardiol 2005;46:2277-83

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

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

Types of electrical dyssynchrony in CRT studies in CHD



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

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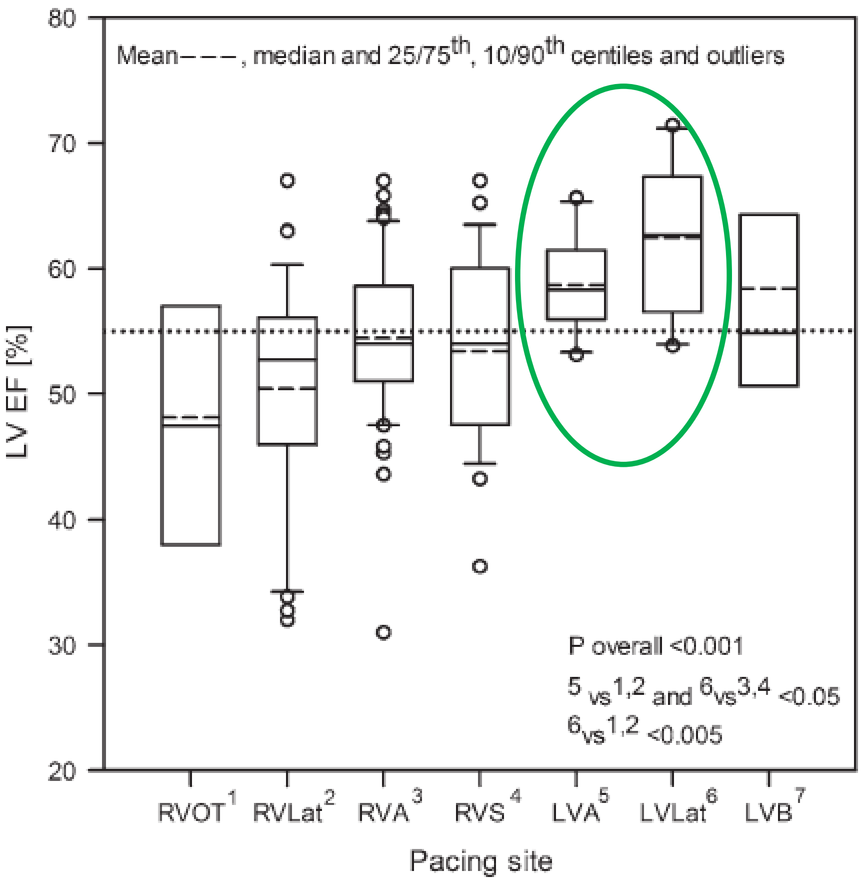
Janousek J et al. Heart 2009, 95:1165-71

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

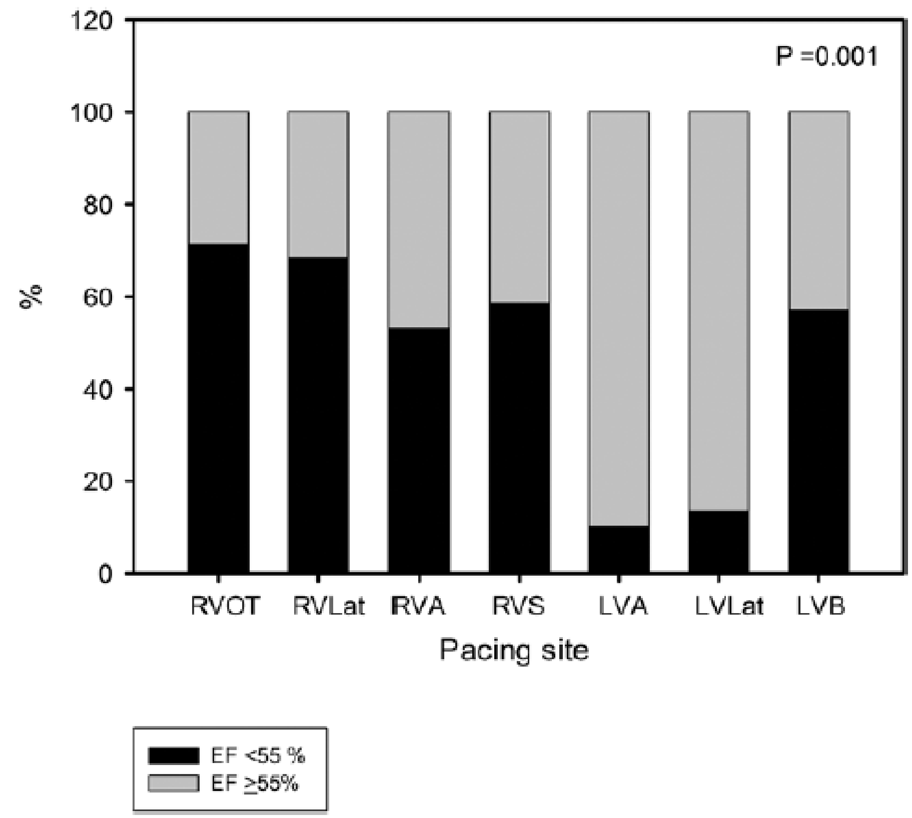
Permanent Cardiac Pacing in Children - Choosing the Optimal Pacing Site: A Multi-Center Study

(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
- May not expect full reverse remodeling

Cardiac Resynchronization Therapy for Pediatric Patients With Heart Failure and Congenital Heart Disease

A Reappraisal of Results

Kara S. Motonaga, MD; Anne M. Dubin, MD

(*Circulation*. 2014;129:1879-1891.)

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.

CRT in systemic RV failure

RBBB or LV paced

N pts.

nders

esponders

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

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

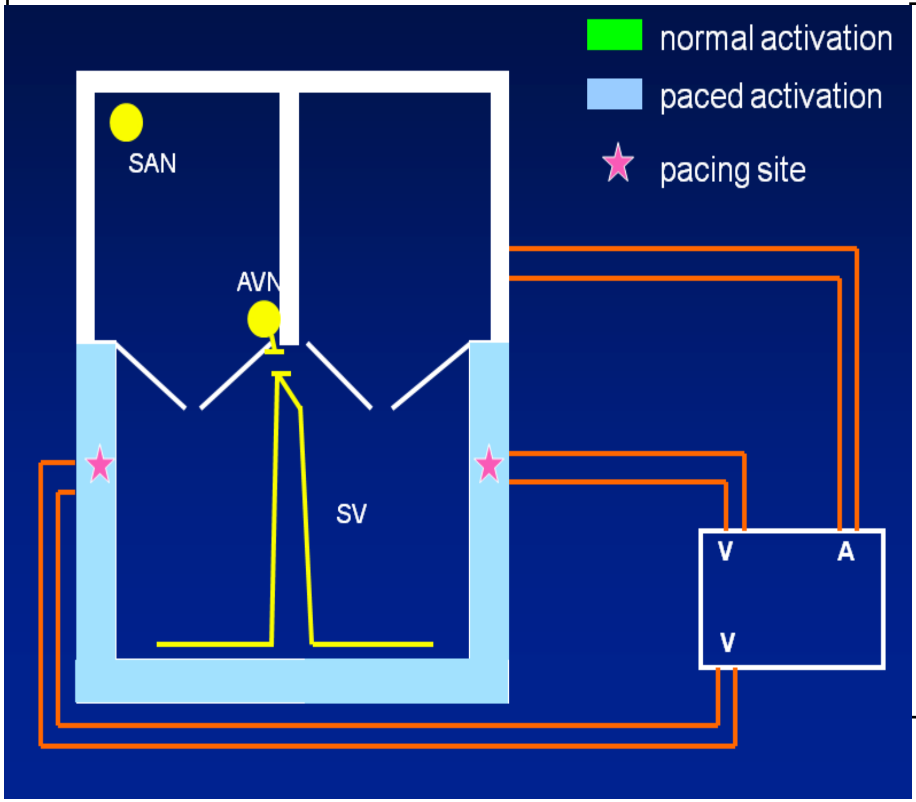
Cecchin F et al. JCE 2009;20:58-65

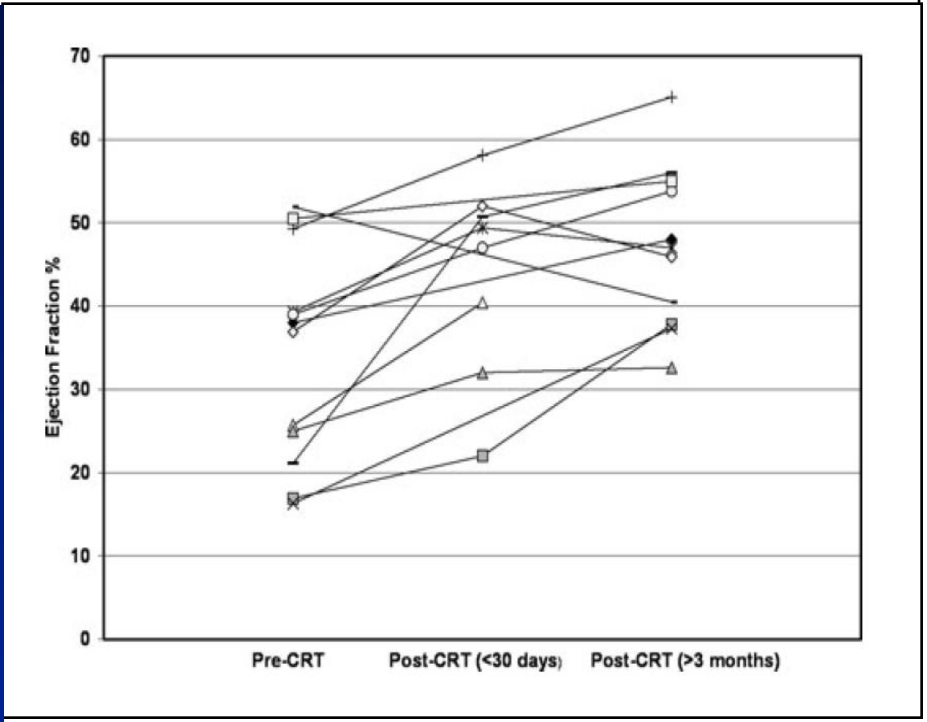
Janousek J et al. Heart 2009, 95:1165-71

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

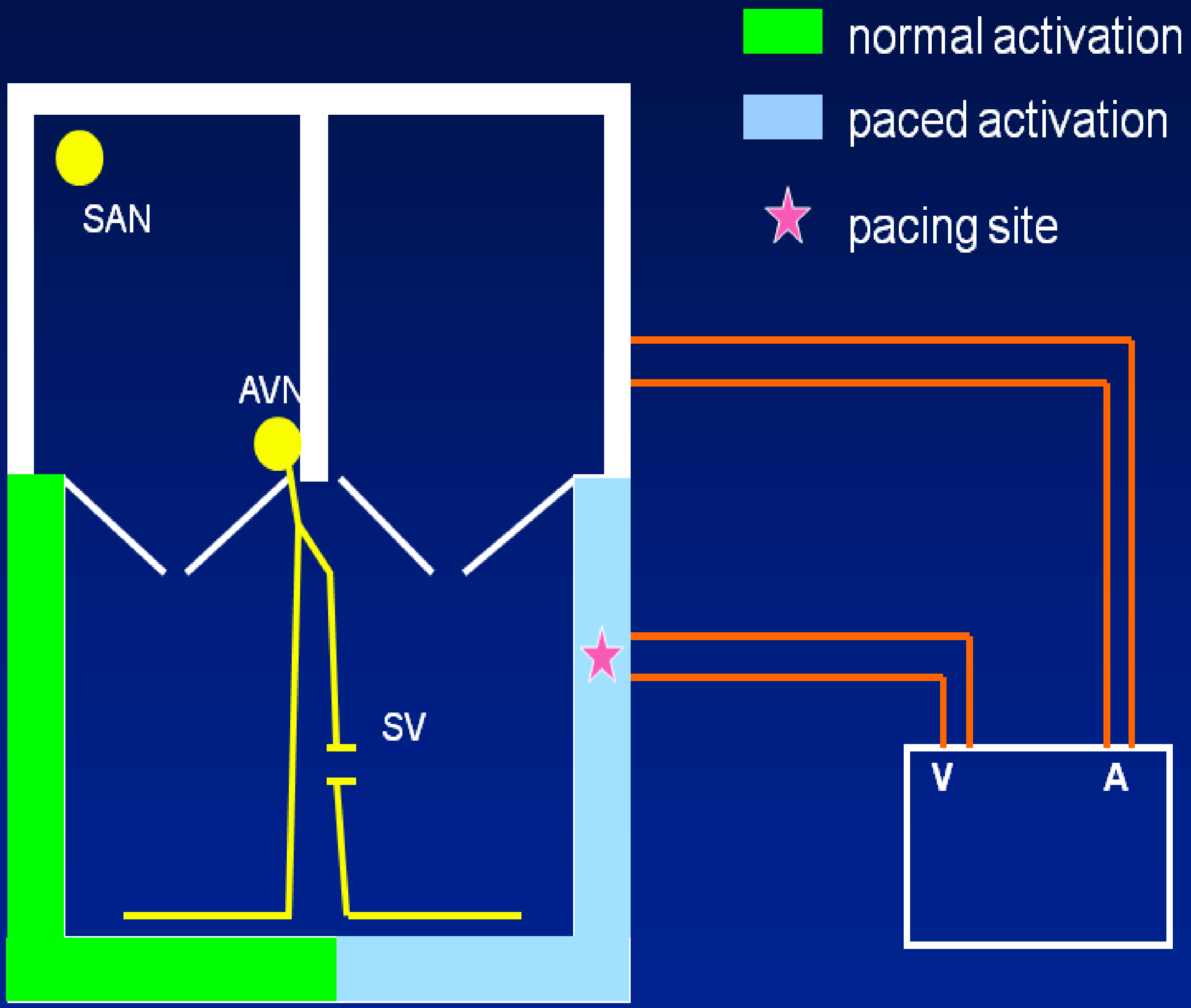




Cecchin F et al. JCE 2009;20:58-65

Resynchronizing the single ventricle

The bundle branch block patient



Single-site pacing in fusion with intrinsic activation

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

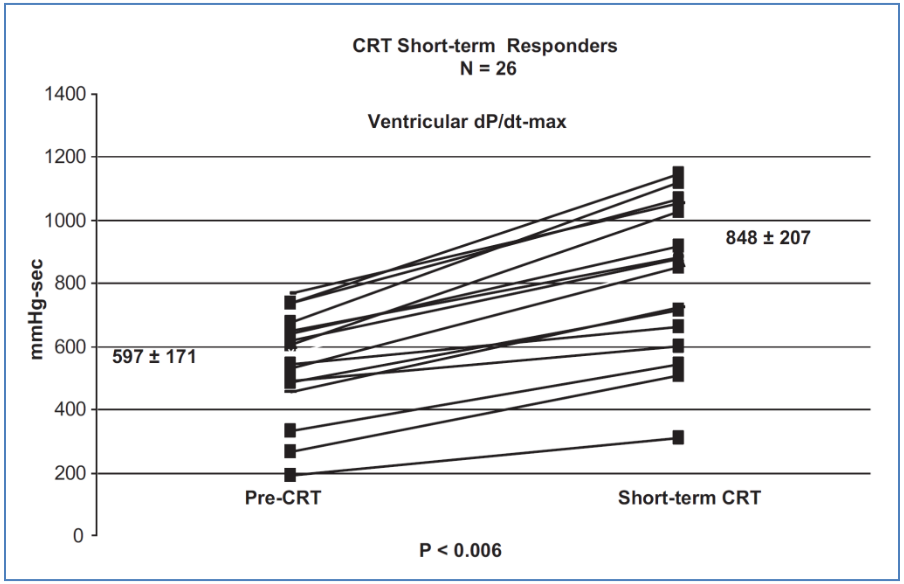
Testing of CRT effect prior to implantation

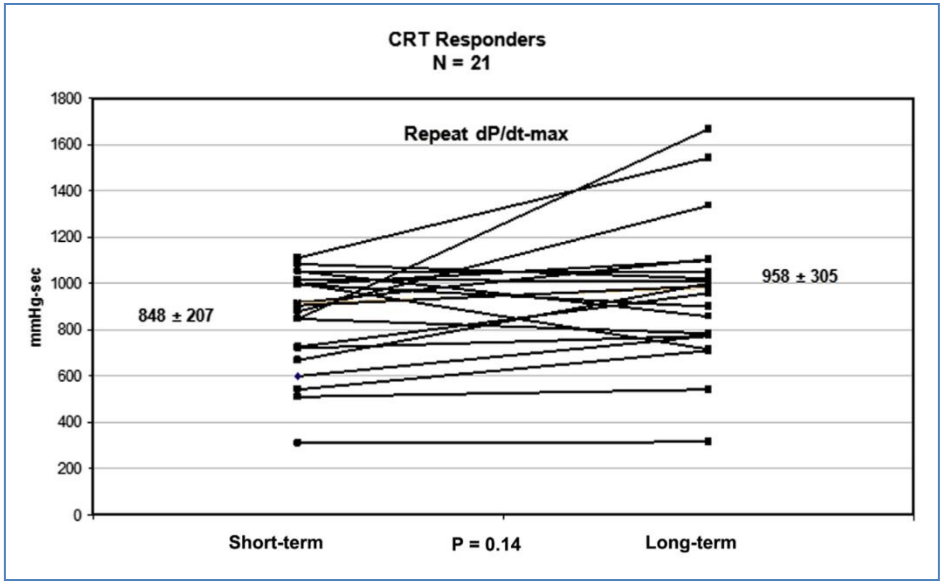
16 Years of Cardiac Resynchronization Pacing Among Congenital Heart Disease Patients

J Am Coll Cardiol EP 2017

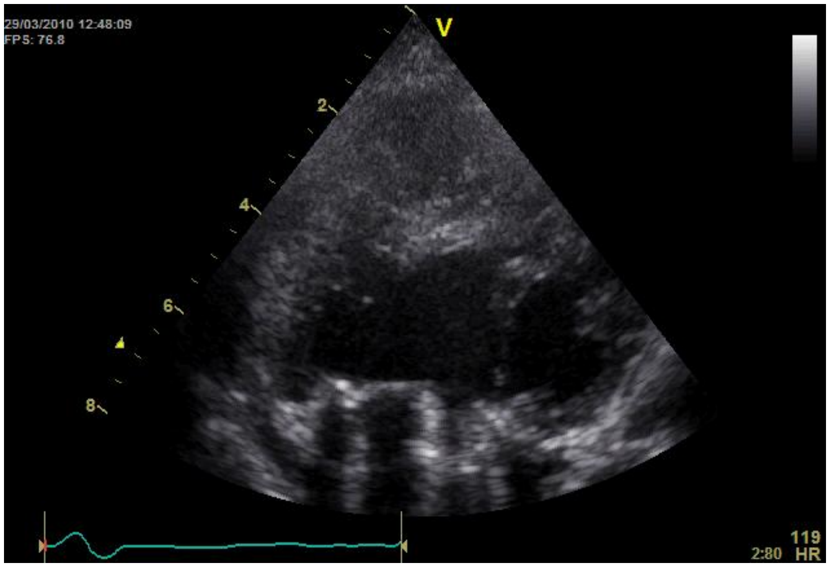
Direct Contractility (dP/dt-max) Screening When the Guidelines Do Not Apply

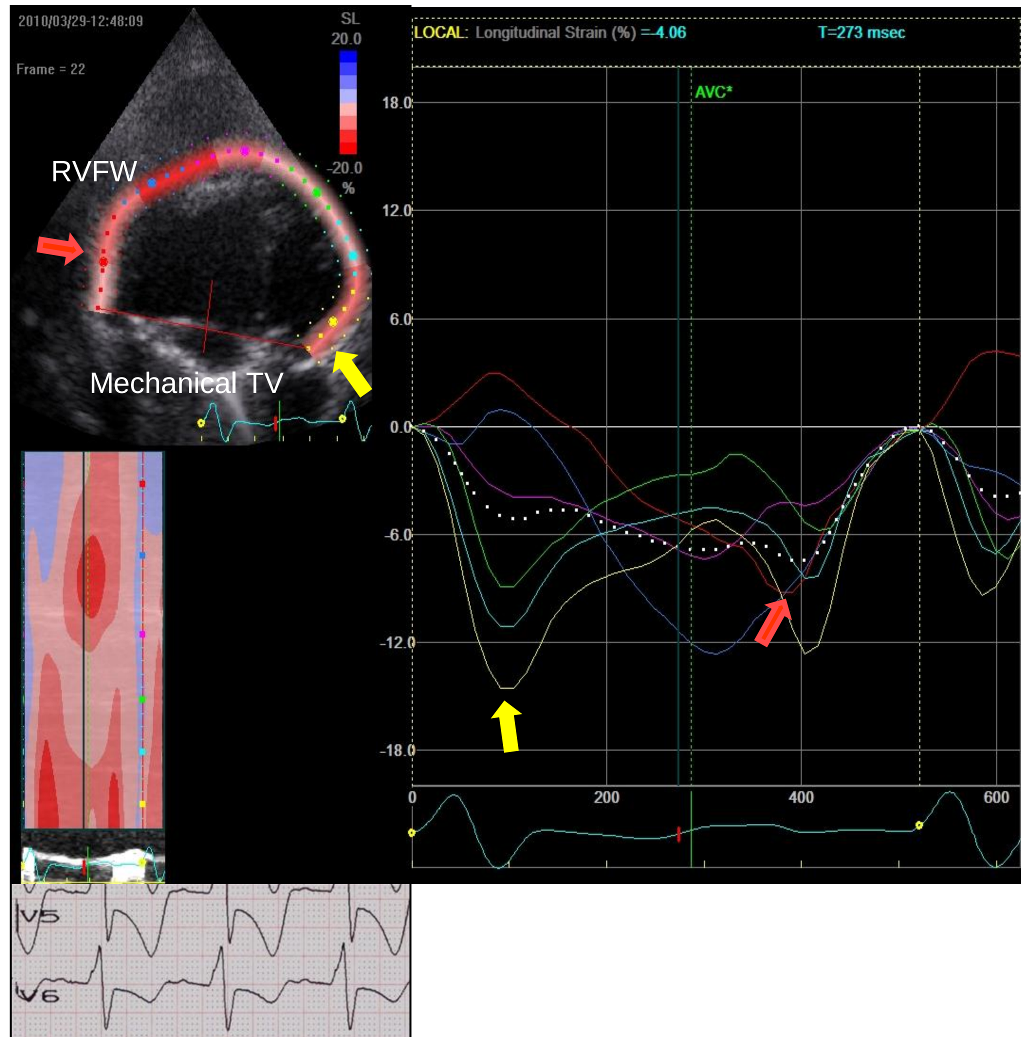
Peter P. Karpawich, MSc, MD, Neha Bansal, MD, Sharmeen Samuel, MD, Yamuna Sanil, MD, Kathleen Zelin, MSN, CPNP





Pre-procedural mechanical activation mapping



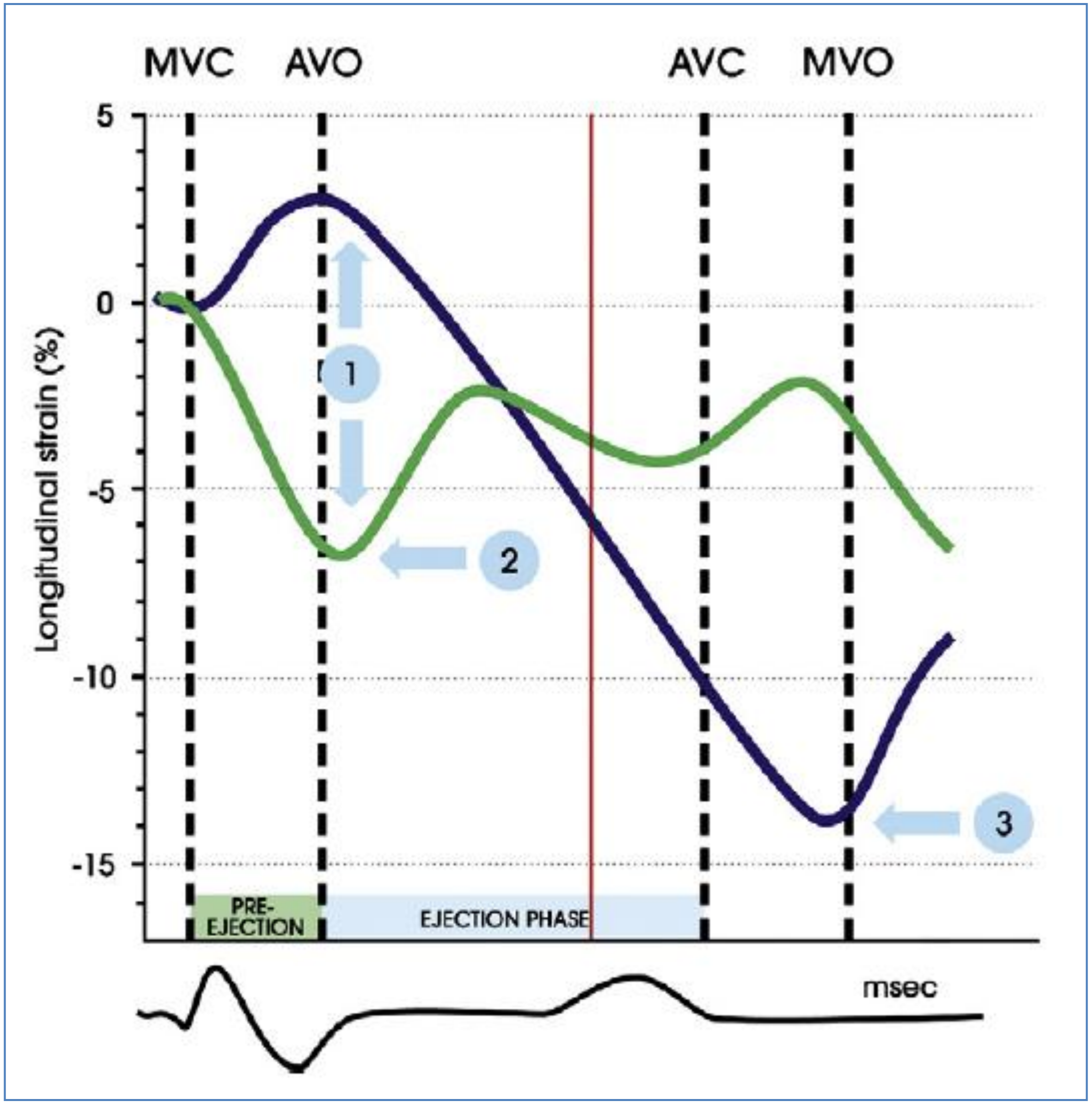


HLHS, st.p. BCPA and TV replacement

Failing dyssynchronous RV due to RBBB

Materna et al. HeartRhythm J. 2014

Detection of classic dyssynchrony pattern

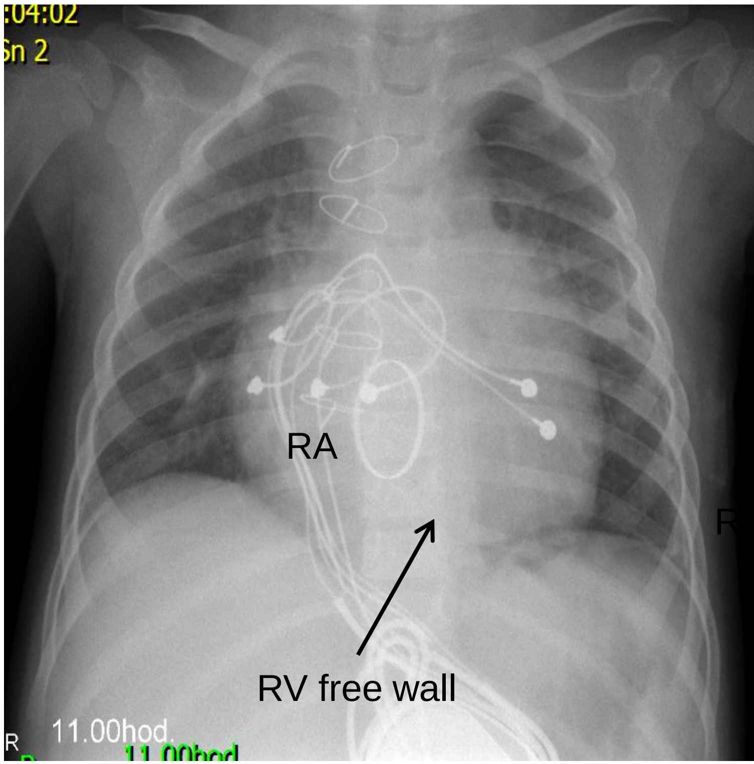


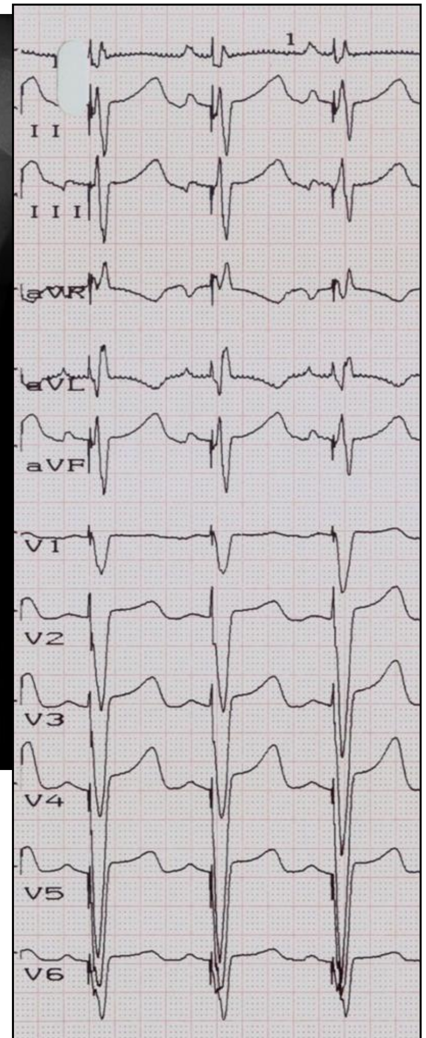
Prediction of positive CRT response in pts. with LBBB

– Sensitivity 95 %
– Specificity 91 %

Risum N et.al., Amer.H.J. 2012

Lead placement according to activation mapping





Materna et al. HeartRhythm J. 2014

Europace (2017) 0, 1–8
doi:10.1093/europace/euw386

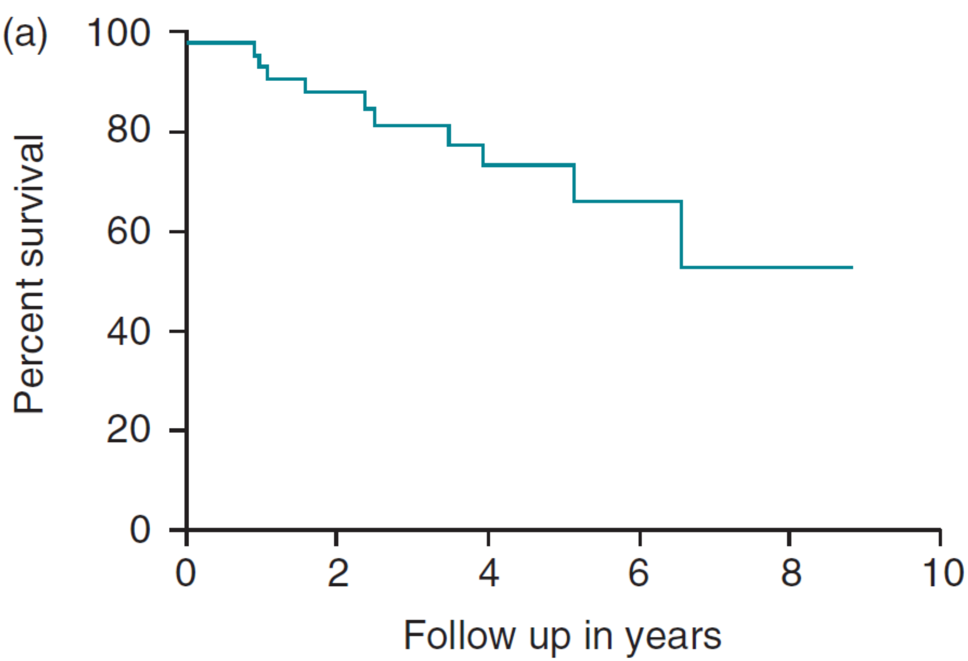
CLINICAL RESEARCH

Cardiac resynchronization therapy in adults with congenital heart disease

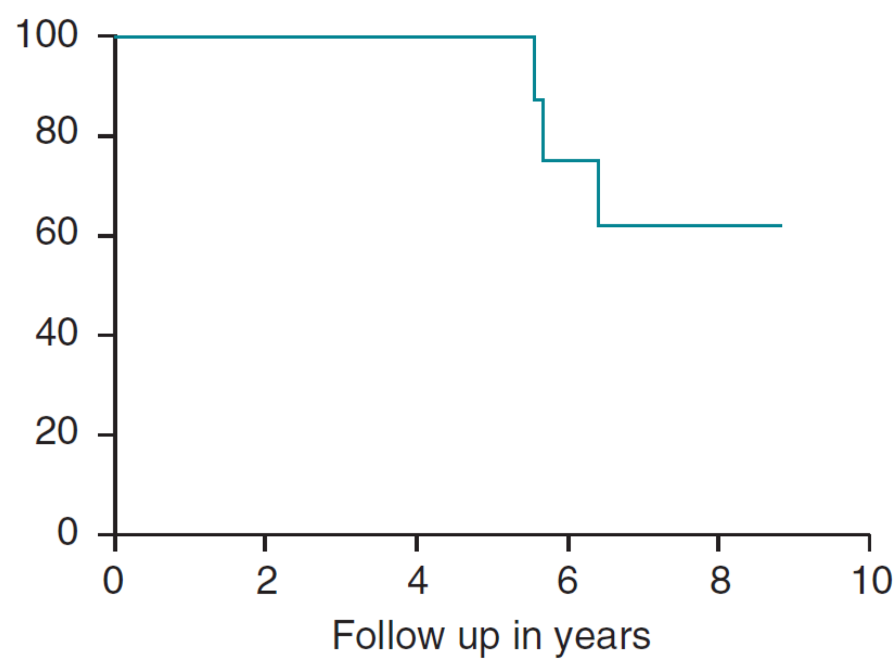
N = 48, median age/FUP: 47.0/2.6 yrs

Zeliha Koyak[1,2], Joris R. de Groot[1], Ahmed Krimly[3], Tara M. Mackay[1], Berto J. Bouma[1], Candice K. Silversides[3], Erwin N. Oechslin[3], Ulas Hoke[4], Lieselot van Erven[4], Werner Budts[5], Isabelle C. Van Gelder[6], Barbara J. M. Mulder[1,2]*, and Louise Harris[3]

Freedom from death/heart transplantation

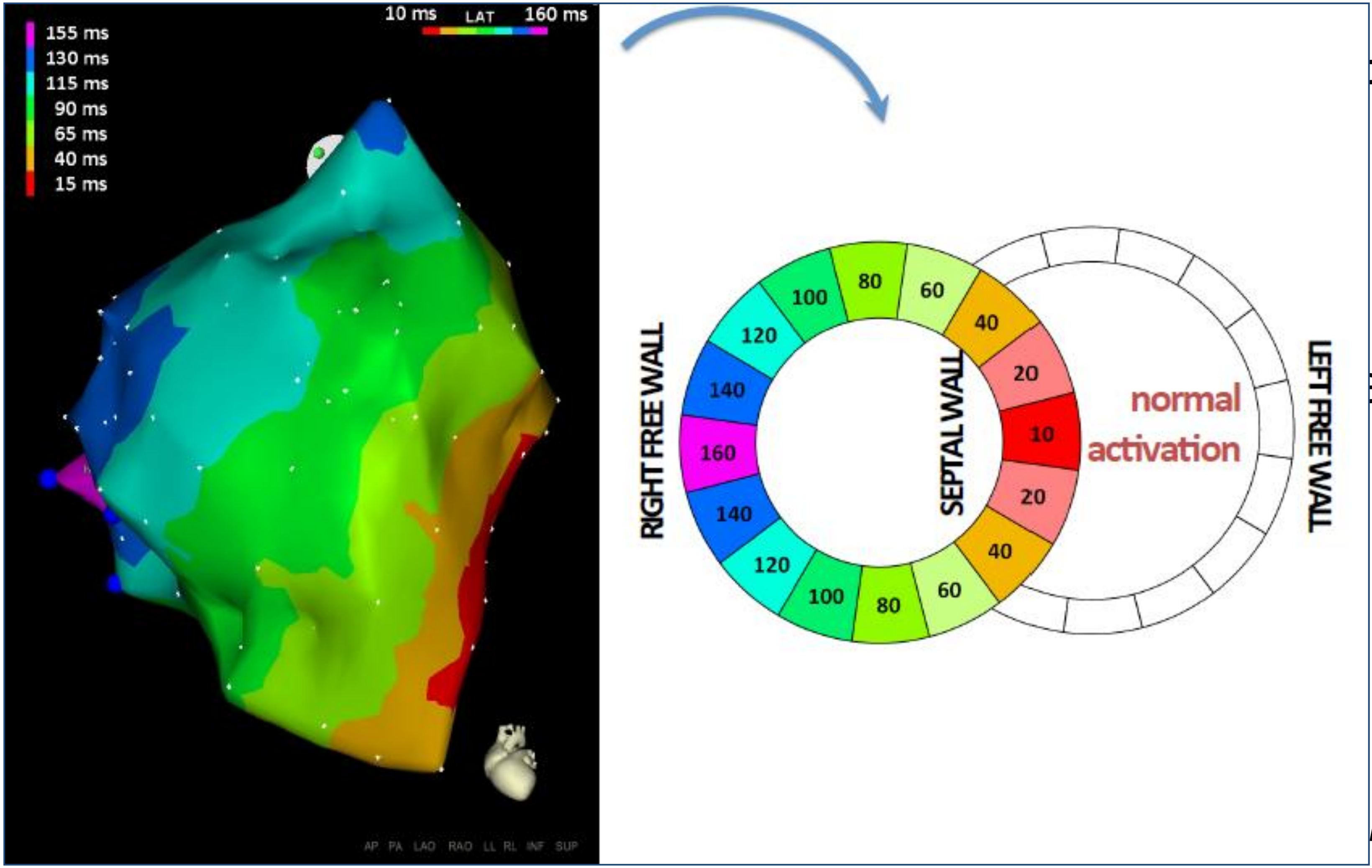


Freedom from CRT system dysfunction



Pulmonary RV-CRT

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



- … PVR
- … e

… r 2008

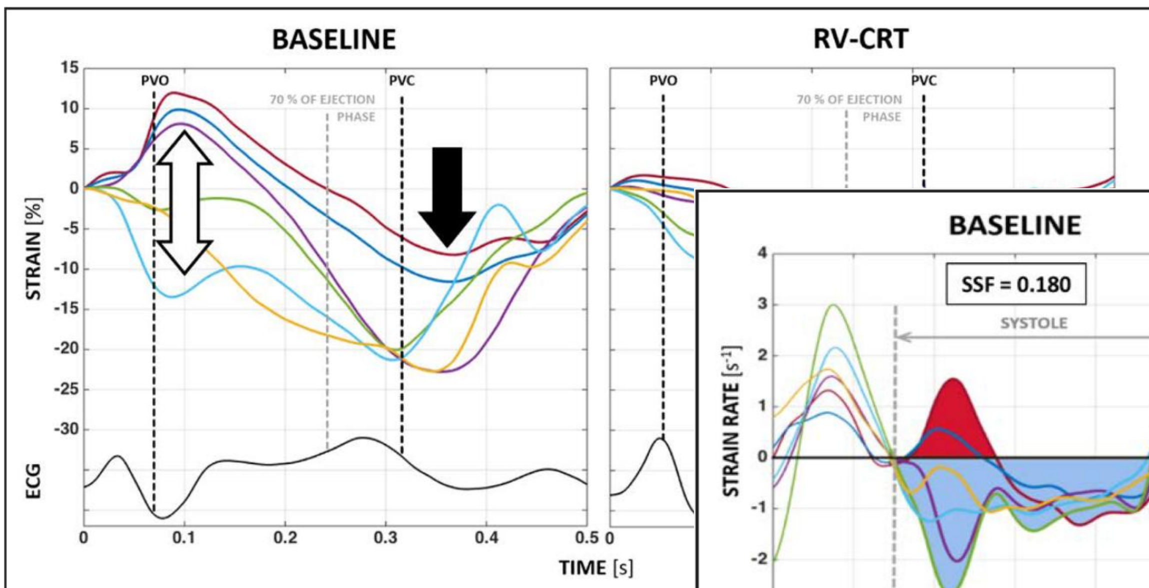
CLINICAL PERSPECTIVE

In patients with congenital heart disease, right ventricular (RV) dyssynchrony caused by right bundle block has been associ-

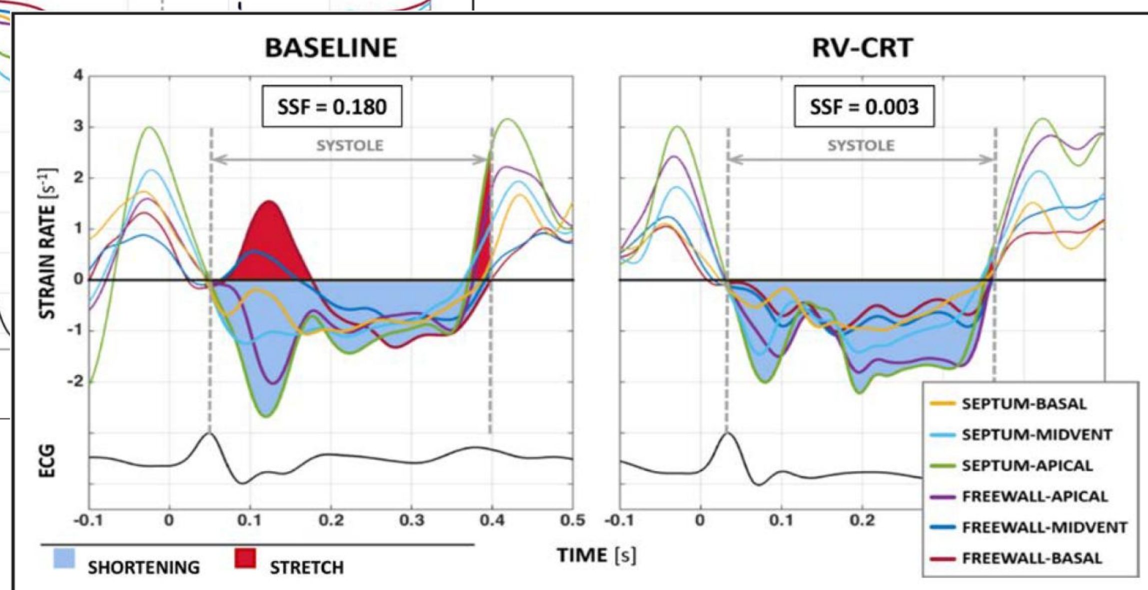
RV-CRT may be beneficial for patients with ToF and similar lesions who do not show reverse RV remodeling after PVR

cardiac resynchronization therapy may be beneficial for patients with tetralogy of Fallot and similar lesions who do not show reverse RV remodeling after pulmonary revalvulation. These data may help to design an ethically acceptable prospective, randomized trial using RV cardiac resynchronization therapy with magnetic resonance imaging compatible pacing systems in patients late after repair of tetralogy of Fallot undergoing pulmonary valve replacement to observe chronic RV functional parameters, exercise capacity, and clinical outcome.

RV mechanics

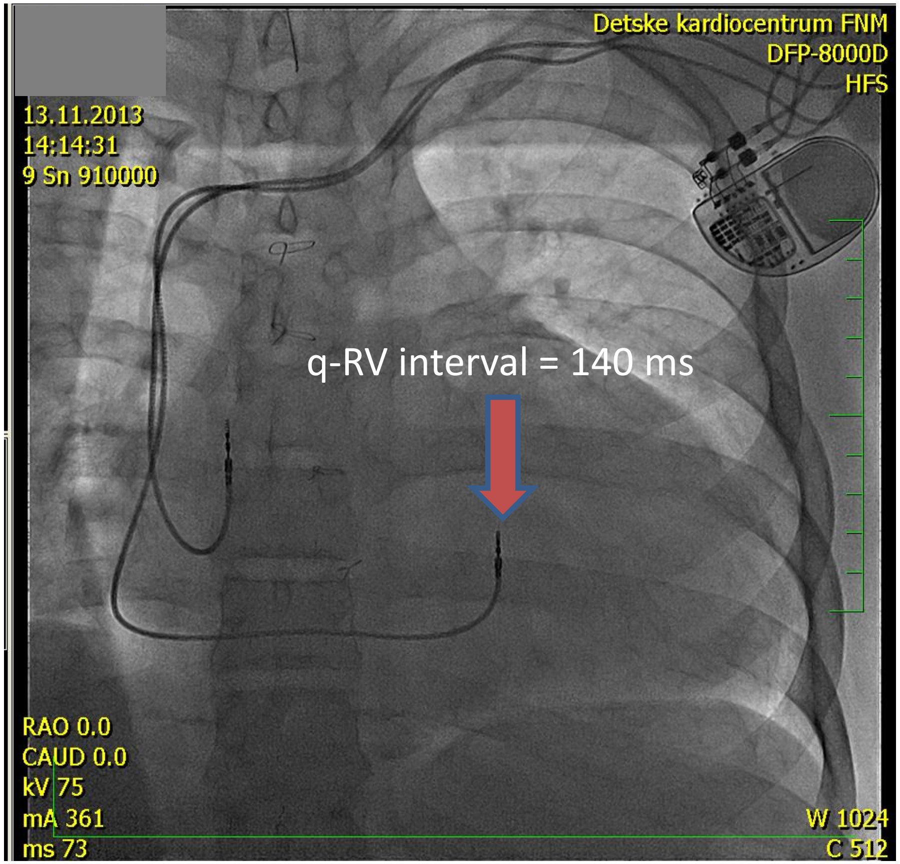


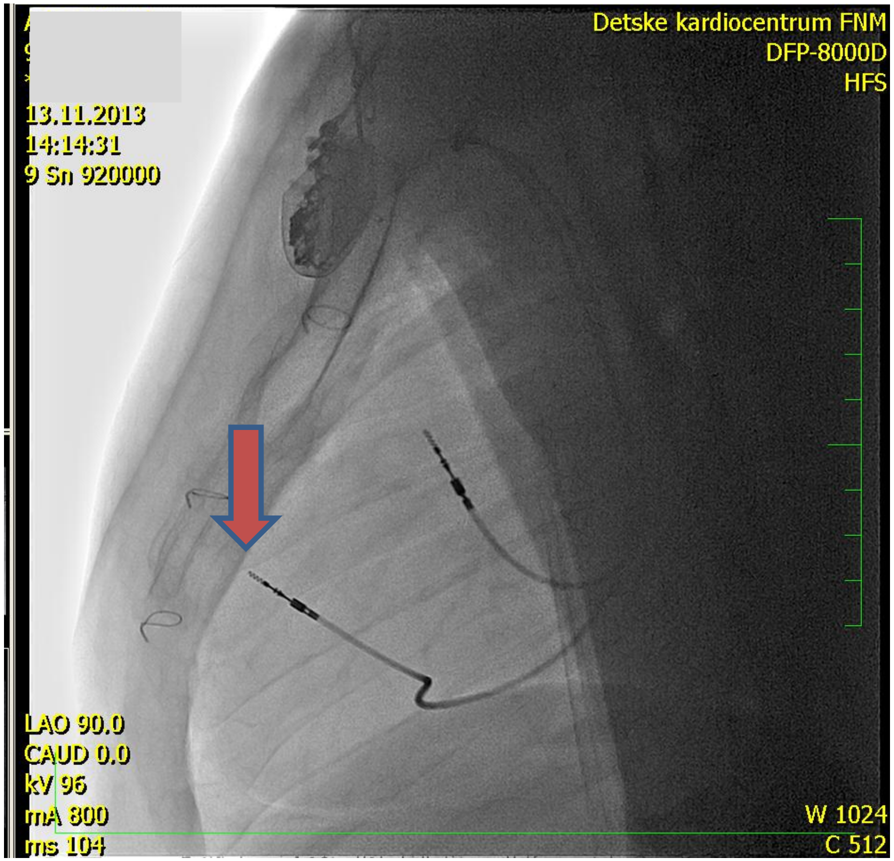
RV contraction efficiency



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

RV-CRT in a failing repaired ToF patient





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

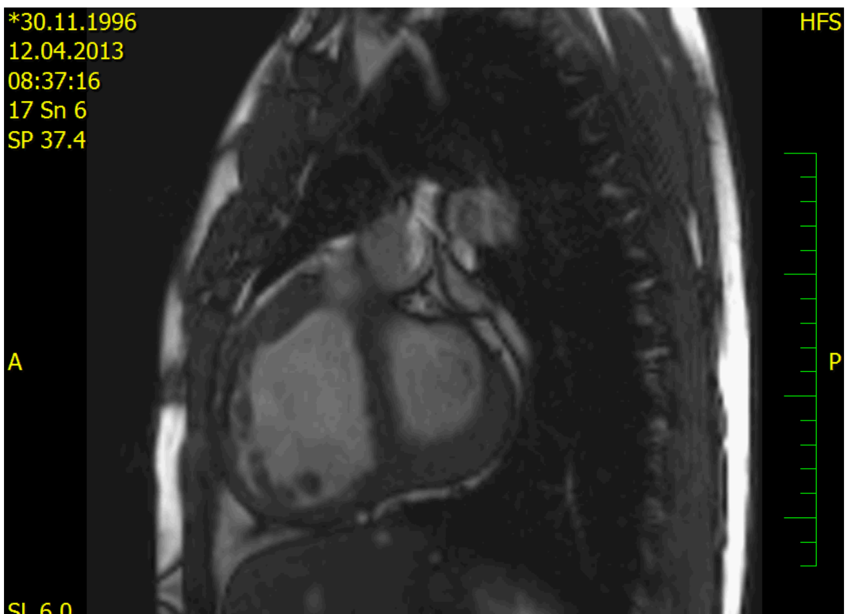
Peter Kubus, Ondrej Materna, Petr Tax, Viktor Tomek and Jan Janousek

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 %



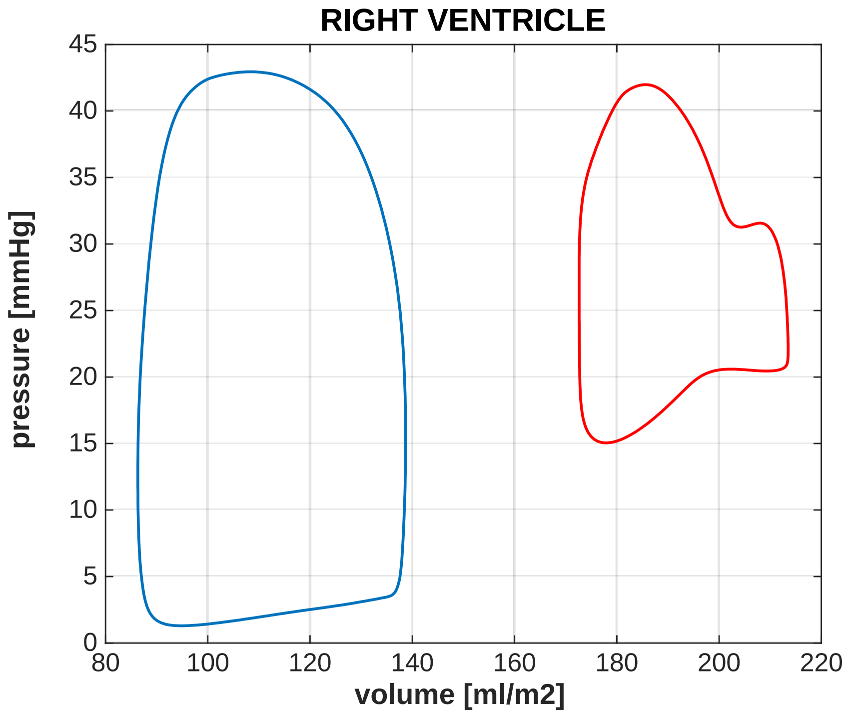
Exercise stress testing - $V0_2$ max: 21,0 (before) ➡ 30,4 ml/kg/min. (6 mos of CRT)

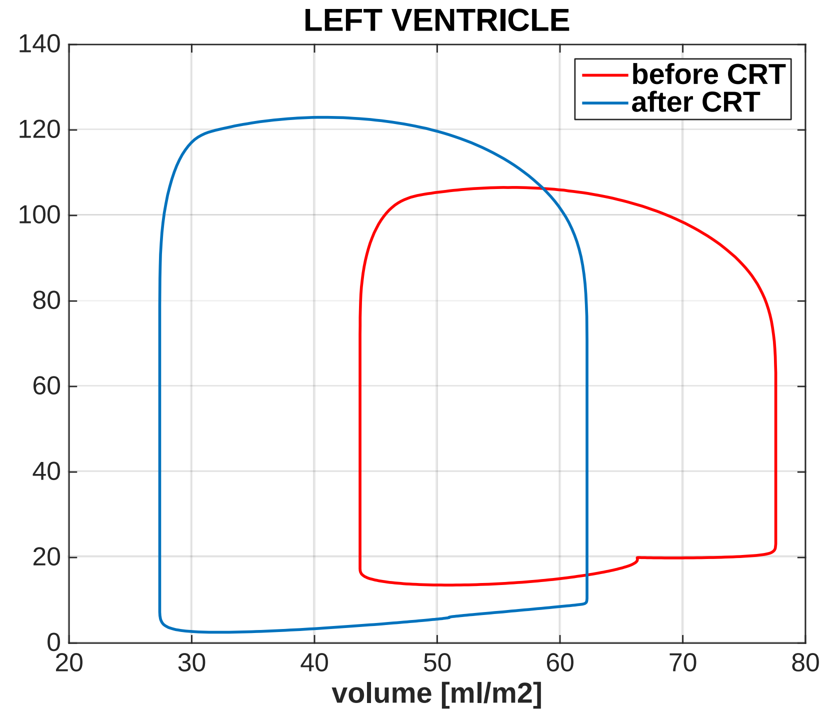
NYHA II ➡ I

Computer modeling of RV-CRT

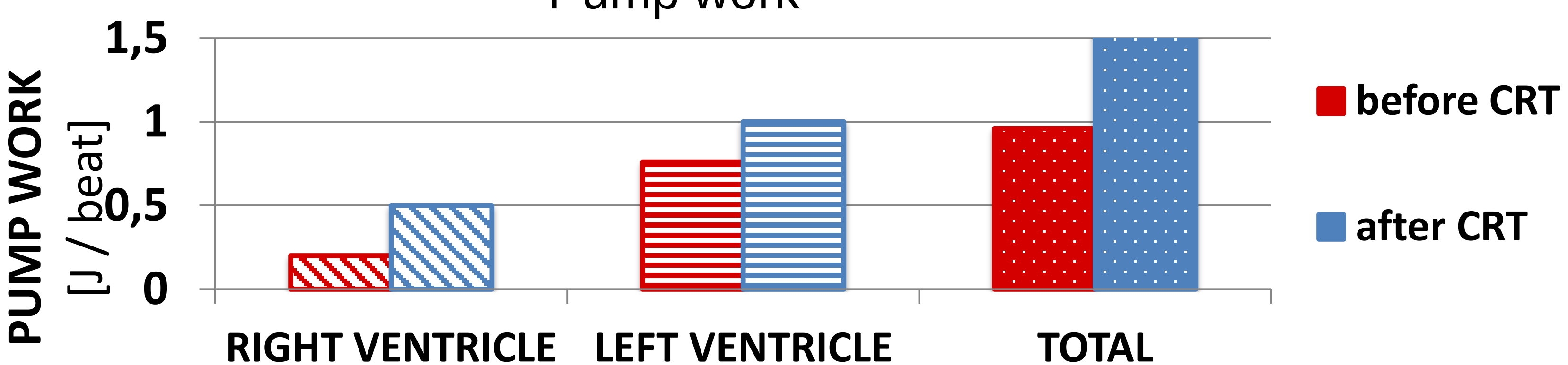
CircAdapt Simulator
Univ. of Maastricht

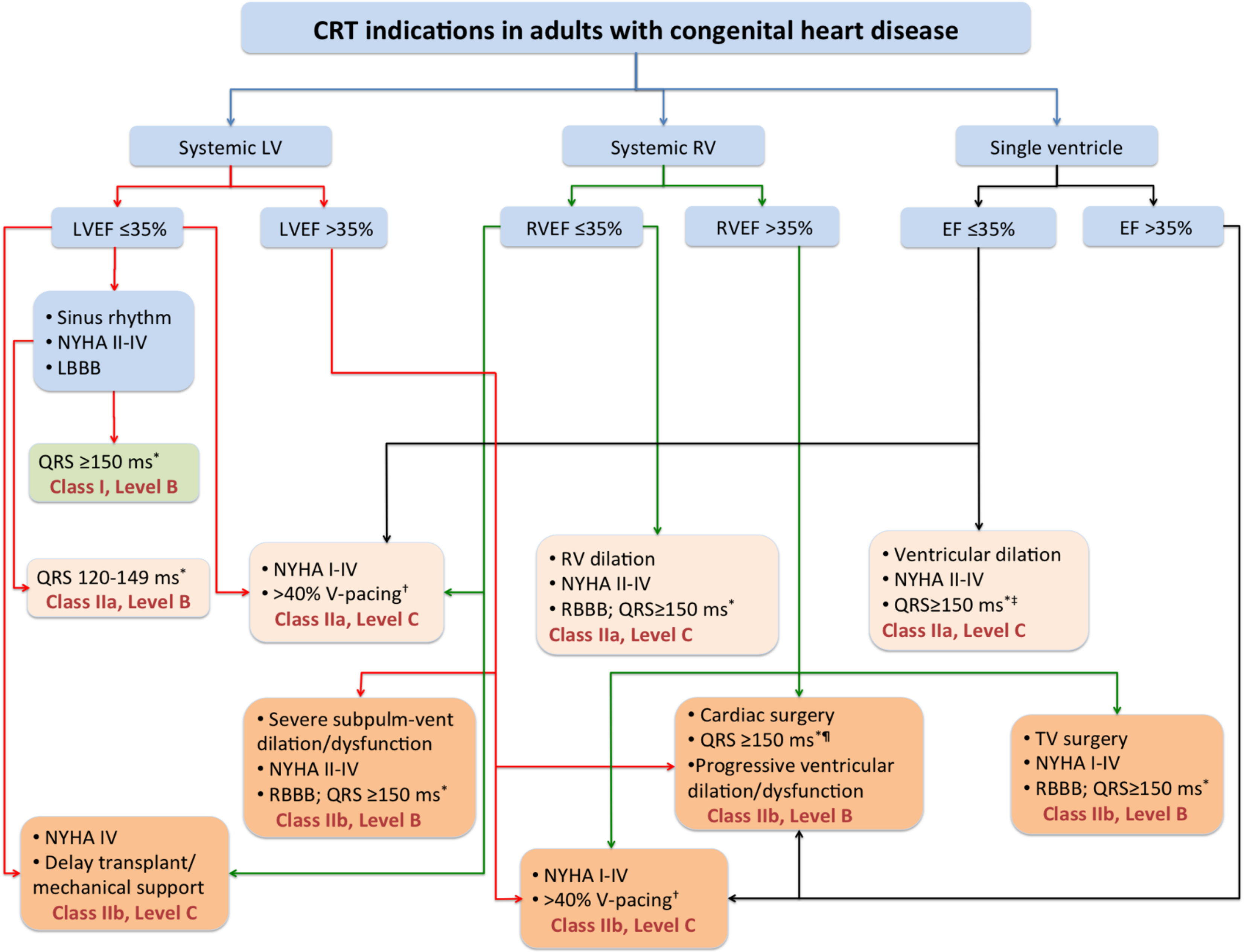
Pressure-volume curves





Pump work



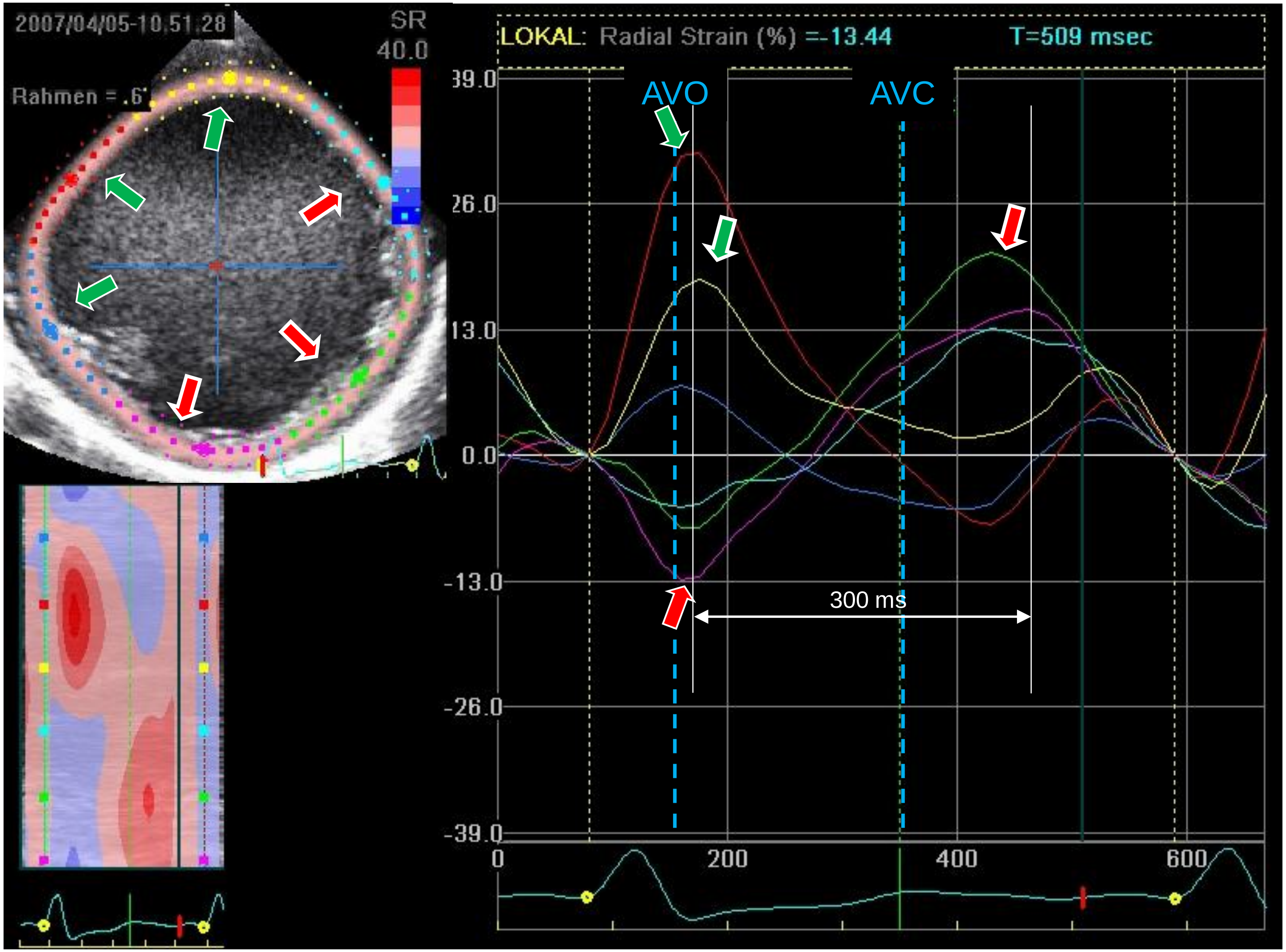


Khairy P et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease. *HeartRhythm Journal 2014*

What to expect in the future?

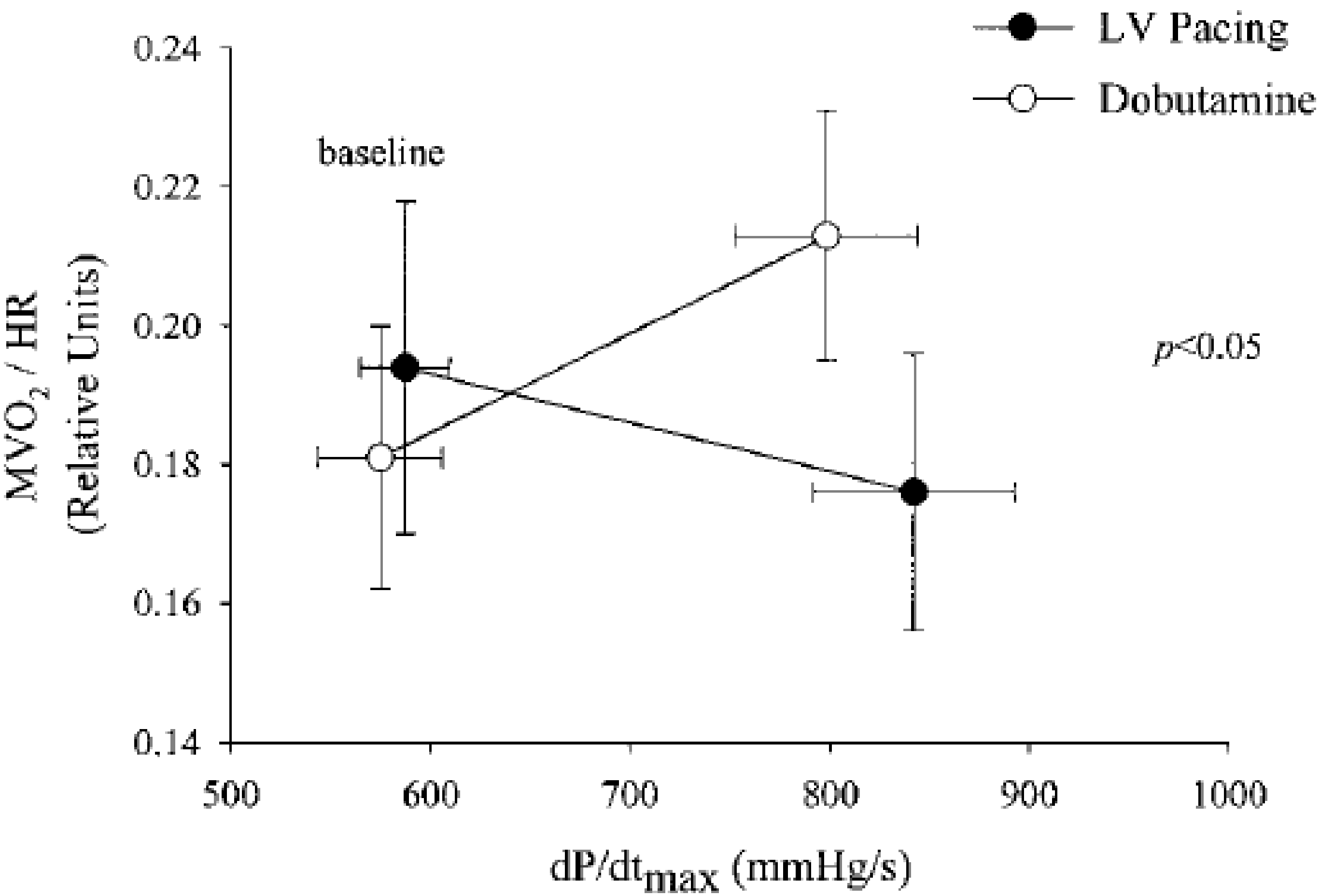
- Imaging studies on predictors of CRT efficacy
  - Classical pattern of dyssynchrony
- Studies on pre-implantation efficacy testing
  - Global and robust contractility parameters
  - Computer modelling
- Increased use of CRT for pulmonary RV failure
- New technologies
  - Leadless CRT pacemakers
- Evaluation of long-term impact of CRT on survival and morbidity
  - CRT in competition or conjunction with other strategies - VAD

Classical pattern of dyssynchrony



Risum N et al. Am Heart J 2012

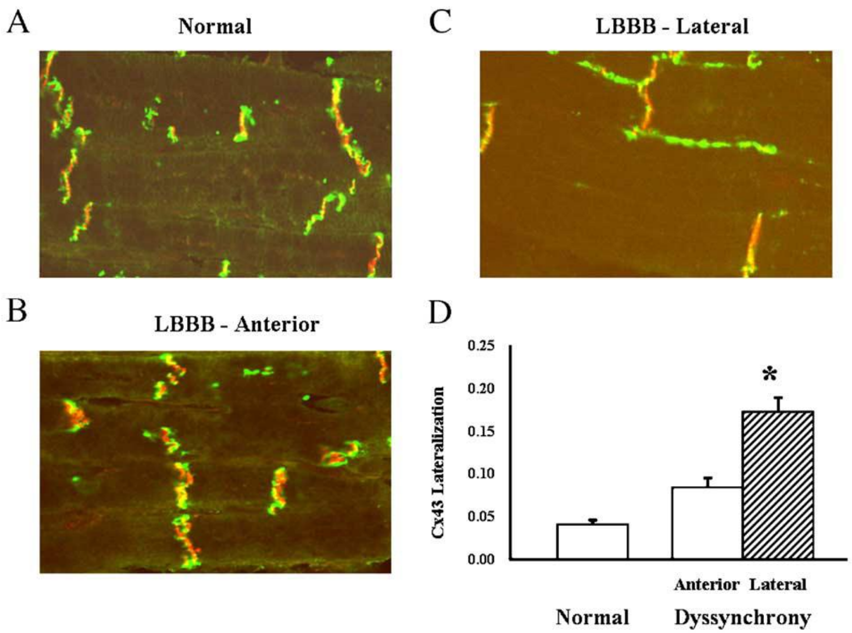
CRT - improved cardiac function at diminished energy cost



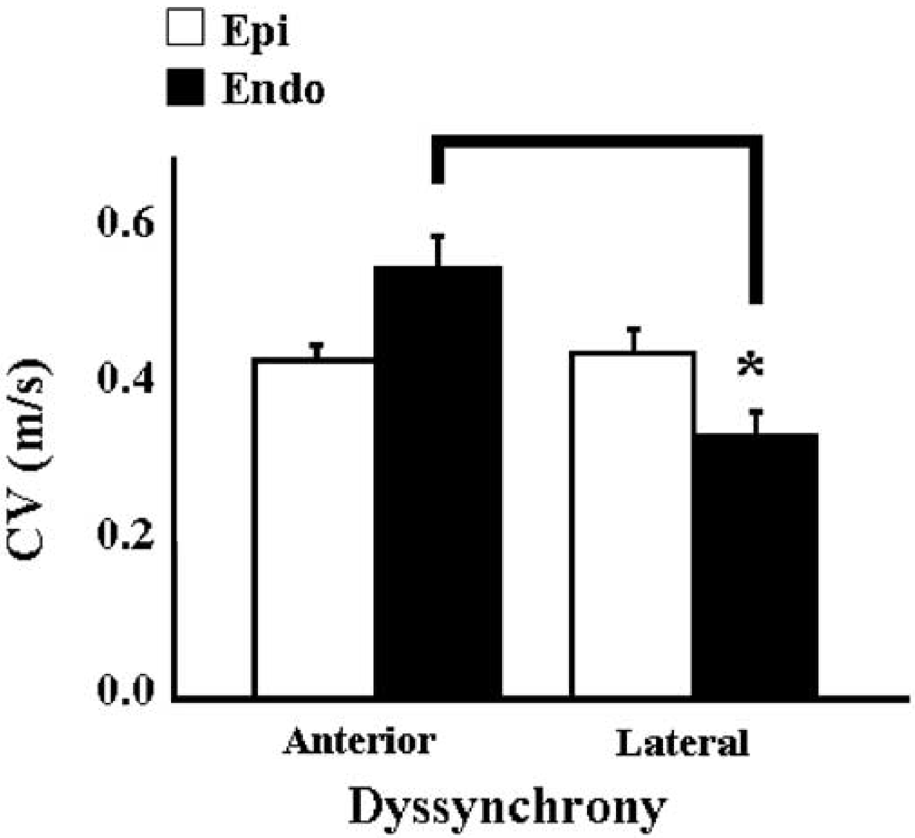
Nelson GS et al., Circulation 2000

CRT – reverse cellular remodeling

Connexine 43 distribution



Conduction velocity



CRT restores normal connexine distribution and conduction velocity in late contracting segments

Spragg DD, Kass DA. Prog Cardiovasc Dis. 2006

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