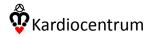
# Long-term outcome of patients with congenital heart disease undergoing cardiac resynchronization therapy

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

- (1) Children's Heart Centre, 2<sup>nd</sup> Faculty of Medicine, Charles University in Prague and Motol University Hospital, Prague, Czech Republic
- (2) Department of Cardiac Surgery, Na Homolce Hospital, Prague, Czech Republic
- (3) Cardiology Department, Institute for Clinical and Experimental Medicine, Prague, Czech Republic



# CRT indications in children/congenital heart disease (CHD)

- Systemic LV
  - LBBB
  - RV pacing
- Systemic RV
  - RBBB
  - LV pacing
- Single ventricle
  - Any bundle branch block
  - "Single site" pacing
- Subpulmonary RV?
  - RBBB

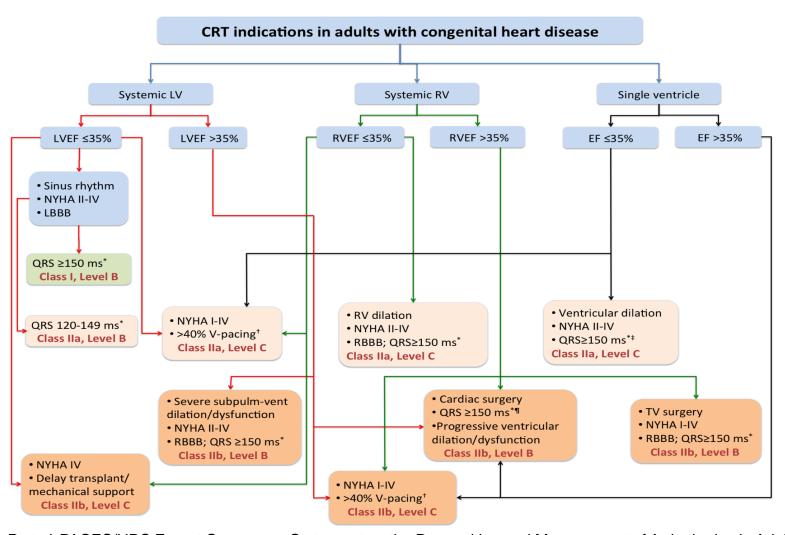


"classic" CRT

Electrical activation delay within failing ventricle required for CRT indication!

Specific for CHD

# CRT indication in adults with CHD



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

#### 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

Single ventricle

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

13 (21.7)

	Janousek et al, <sup>37</sup> 2004	Strieper et al, <sup>38</sup> 2004	Moak et al, <sup>39</sup> 2006	Khairy et al, <sup>40</sup> 2006	Jauvert et al,41 2009	Cecchin et al, <sup>42</sup> 2009	Perera et al, <sup>43</sup> 2013
Total patients, n	8	7	6	13	7	60	67
Age (range), y	Median, 12.5 (6.9–29.2)	Mean, 11 (2.3–28)	Mean, 11.3 (0.5-23.7)	Mean, 7.8 (0.8–15.5)	Mean, 24.6 (15-50)	Median, 15 (0.4–47)	Unknown
Follow-up duration	Median, 17.4 mo	Median, 19 mo	Median, 10 mo	Mean, 16.5 mo	Mean, 19.4 mo	Median, 0.7 y	Mean, 2.75 y
CHD population, n (%)	8 (100)	7 (100)	3 (50)	10 (76.9)	7 (100)	46 (76.7)	50 (74.6)
Systemic RV	8 (100)	1 (14.3)		4 (30.8)	7 (100)	7 (11.7)	
Systemic LV		6 (85.7)	3 (50)	6 (46.2)		26 (43.3)	

### Aim

 To evaluate long-term impact of CRT in pts with CHD and systemic ventricular dysfunction

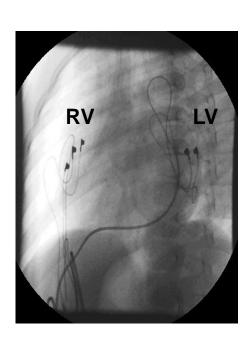
### **Patients**

### Single centre, CRT implantation 2002 – 2014

- N = 30, 15 ♀, 15 ♂
- Underlying substrate
  - Structural CHD (N = 28/30)
  - Systemic ventricle
    - Left = 12
    - Right = 14
    - Single = 4
- Age at CRT implantation: median 12.9 (IQR 6.5 18.2) years
- Follow up: median 9.0 (IQR 4.5 11.4) years on CRT

# **Procedures**

- Type
  - Primary CRT implantation = 11
  - Upgrade from conventional pacing = 19
- CRT-P in all
  - Later upgrade to CRT-D in 1/30
- Implantation
  - Transvenous = 3
  - Thoracotomy = 19
  - Mixed = 8
- Associated with other cardiac surgery = 13/30

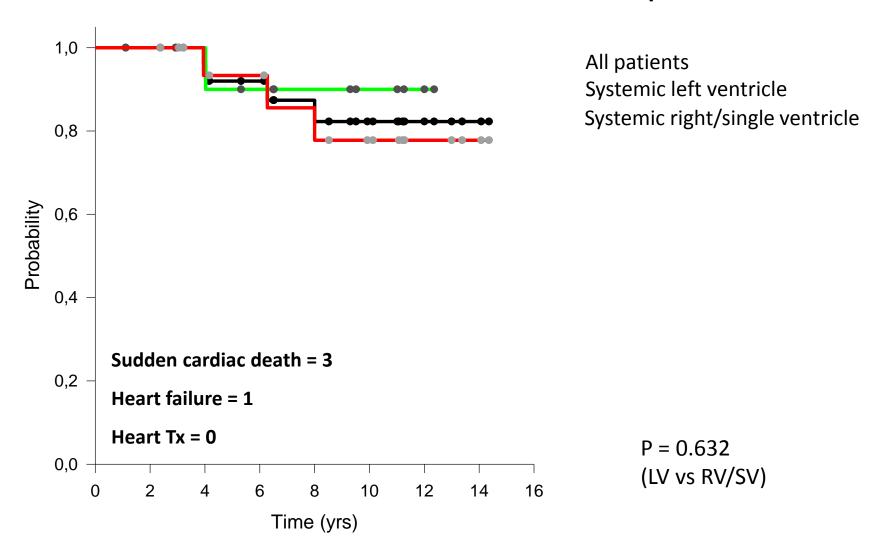


# Follow-up

- Echocardiographic follow-up of systemic ventricular function
- CRT response definition
  - increase in systemic ventricular
    - EF (Simpson biplane, systemic LV) or
    - fractional area of change (FAC, systemic RV/SV) by >10 units and
  - ≤ NYHA class at the end of FUP
- Actuarial survival probability

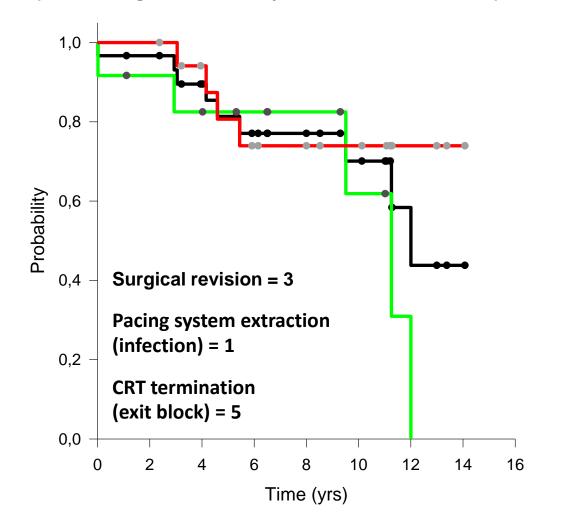
# Cardiovascular death/heart failure hospitalization

#### Freedom from cardiovascular death or heart failure hospitalization



# CRT system complications

Freedom from CRT complications leading to surgical system revision (elective generator replacement excluded) or therapy termination

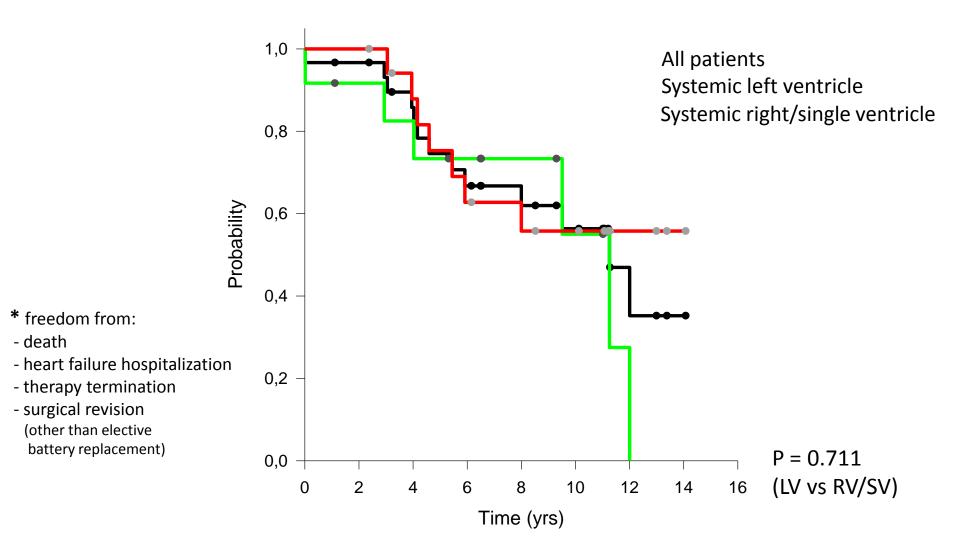


All patients
Systemic left ventricle
Systemic right/single ventricle

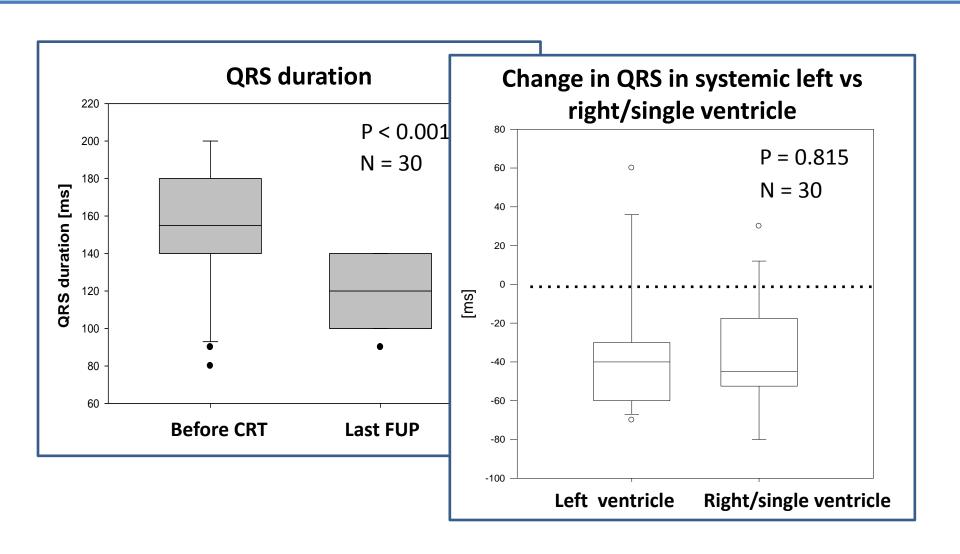
P = 0.418 (LV vs RV/SV)

# Uneventful therapy continuation

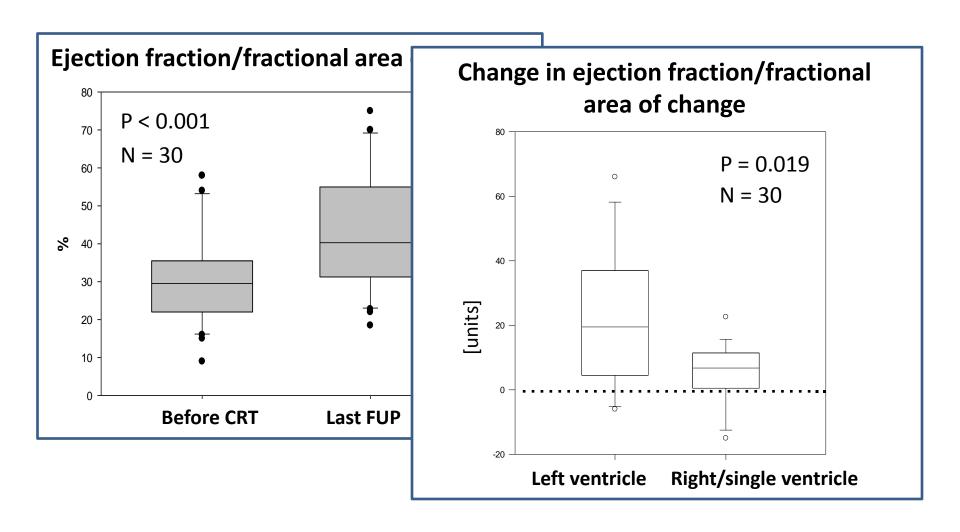
#### Overal probability of uneventful therapy continuation\*



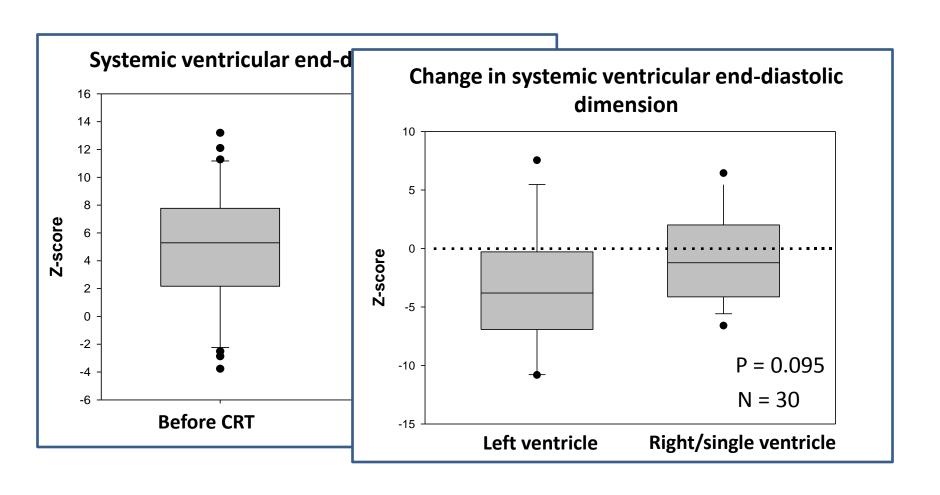
# **QRS** duration



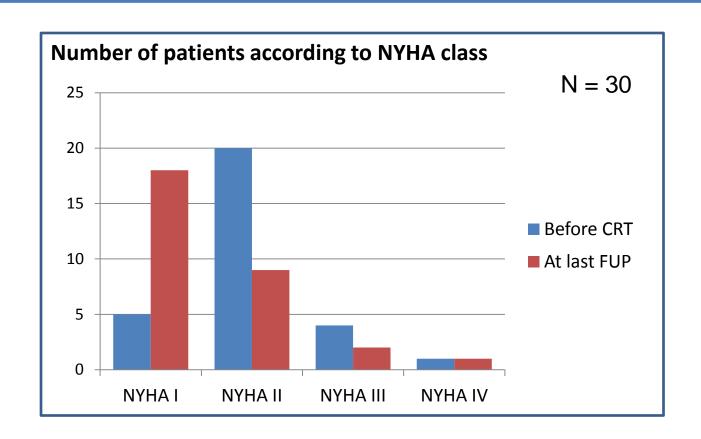
# Systolic function of the systemic ventricle



# Systemic ventricular end-diastolic dimension



# NYHA classification

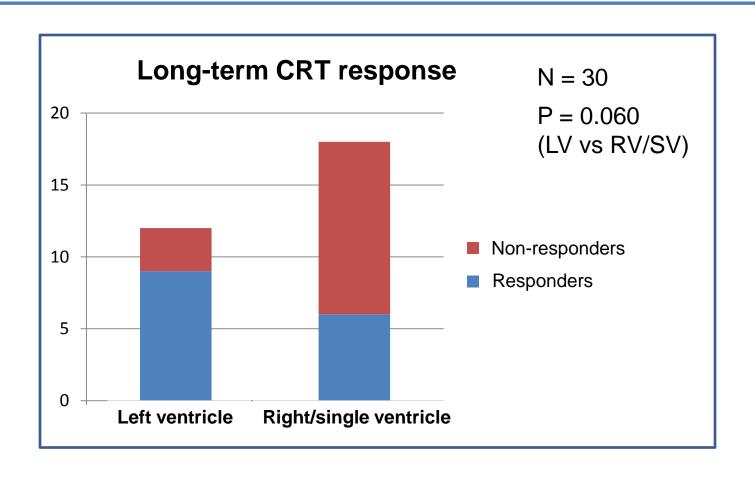


Improved:

Systemic LV: 6/12 pts.

Systemic RV/SV: 6/18 pts.

# Long-term CRT response\*



\*Increase in EF (systemic LV) or FAC (systemic RV/SV) by >10 points and ≤ NYHA class at the end of FUP

# Conclusions

- Long-term CRT in patients with CHD was associated with significant improvement of systemic ventricular function
- CRT was more effective in patients with systemic left ventricle.
- Probability of device complications necessitating surgical revision or therapy termination was high.
- Rate of sudden death was significant (10%)
  - CRT-D should be individually considered in every patient.

