

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

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

A Reappraisal of Results

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Table 1. Single-Center Retrospective Studies of Permanent CRT in Pediatric and CHD-Related HF

	Janousek et al, ³⁷ 2004	Strieper et al, ³⁸ 2004	Moak et al, ³⁹ 2006	Khairy et al, ⁴⁰ 2006	Jauvert et al, ⁴¹ 2009	Cecchin et al, ⁴² 2009	Perera et al, ⁴³ 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)	...
Single ventricle	13 (21.7)	...

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) yrs
- Follow up: median 9.0 (IQR 4.5-11.4) years on CRT

Procedures

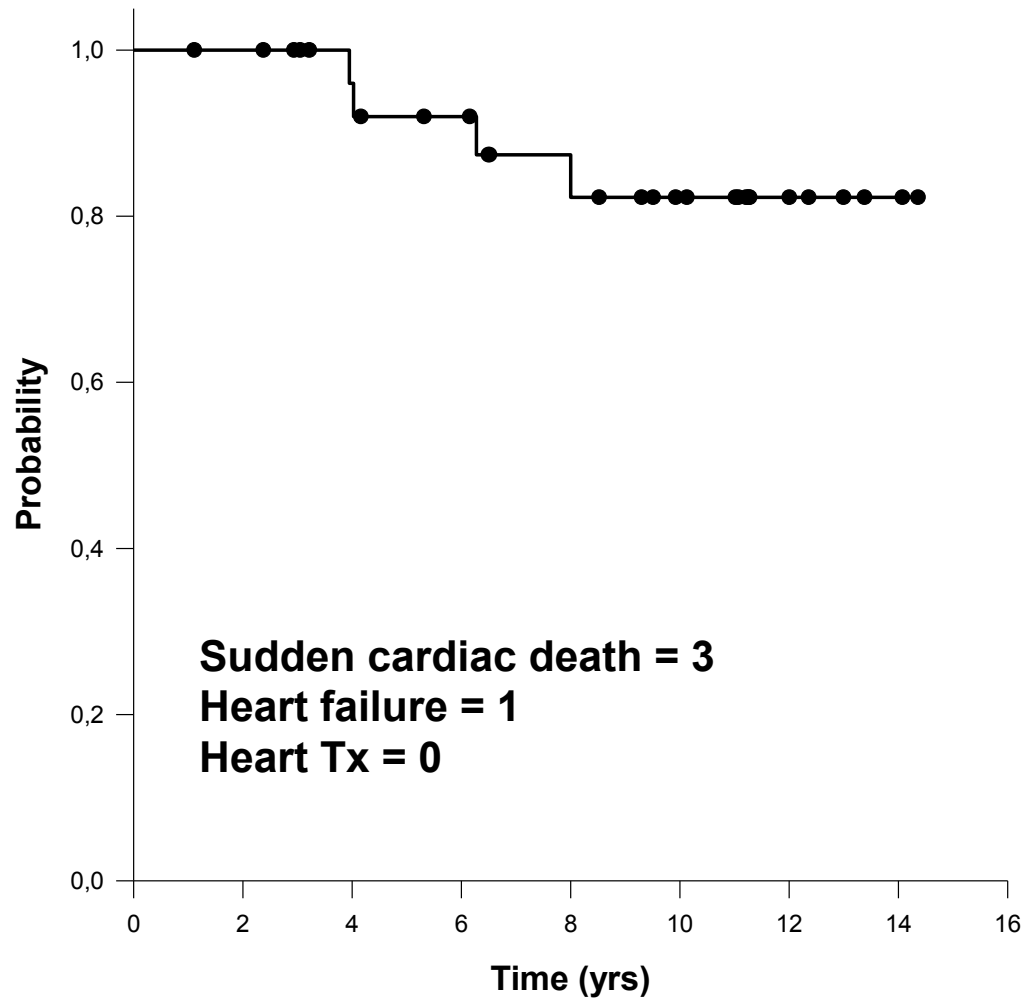
- Type
 - Primary CRT implantation = 11
 - Upgrade from conventional pacing = 19
- CRT-P in all
 - later upgrade to CRT-D in 1/30
- Implantation
 - Transvenous = 3
 - Thoracotomy = 19
 - Mixed = 8
- Associated with other cardiac surgery = 13/30

Follow-up

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

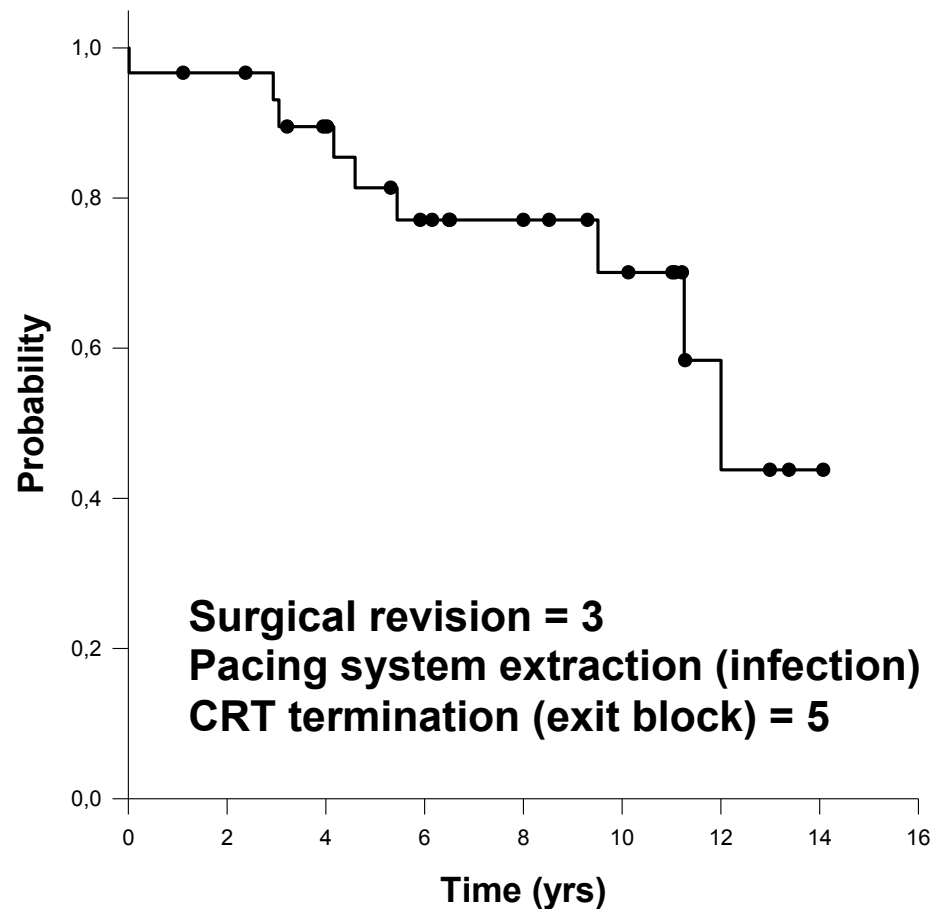
Results (I)

Freedom from cardiovascular death or heart failure hospitalization



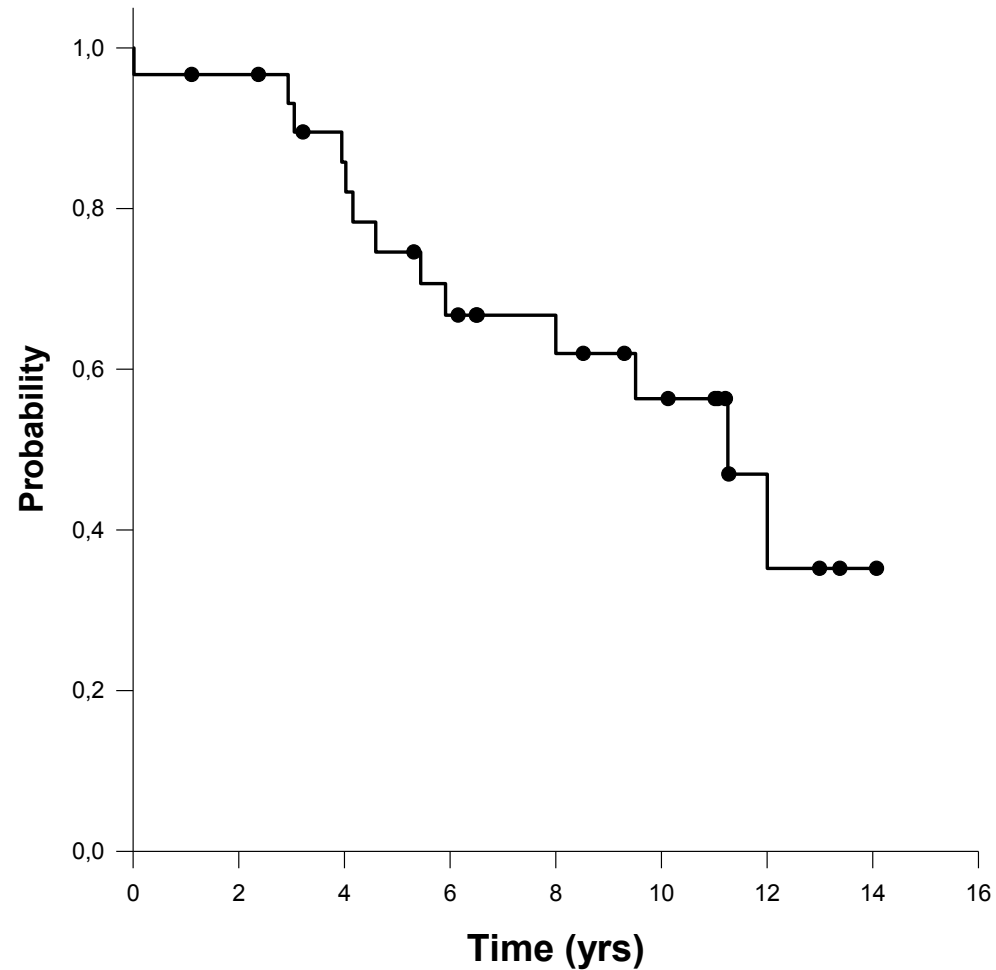
Results (II)

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



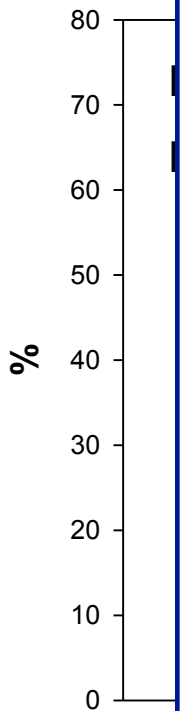
Results (III)

Overall probability of an uneventful therapy continuation

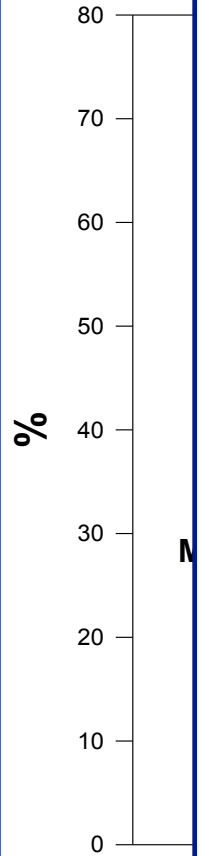


Results (IV)

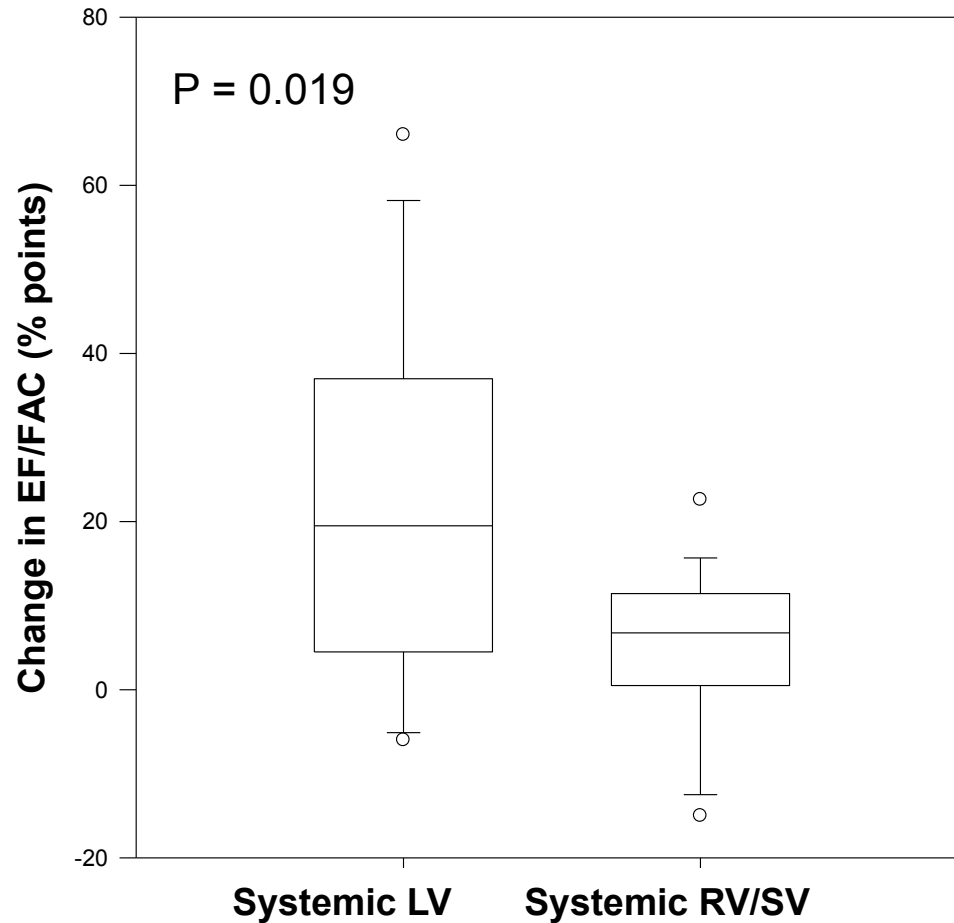
Ejection



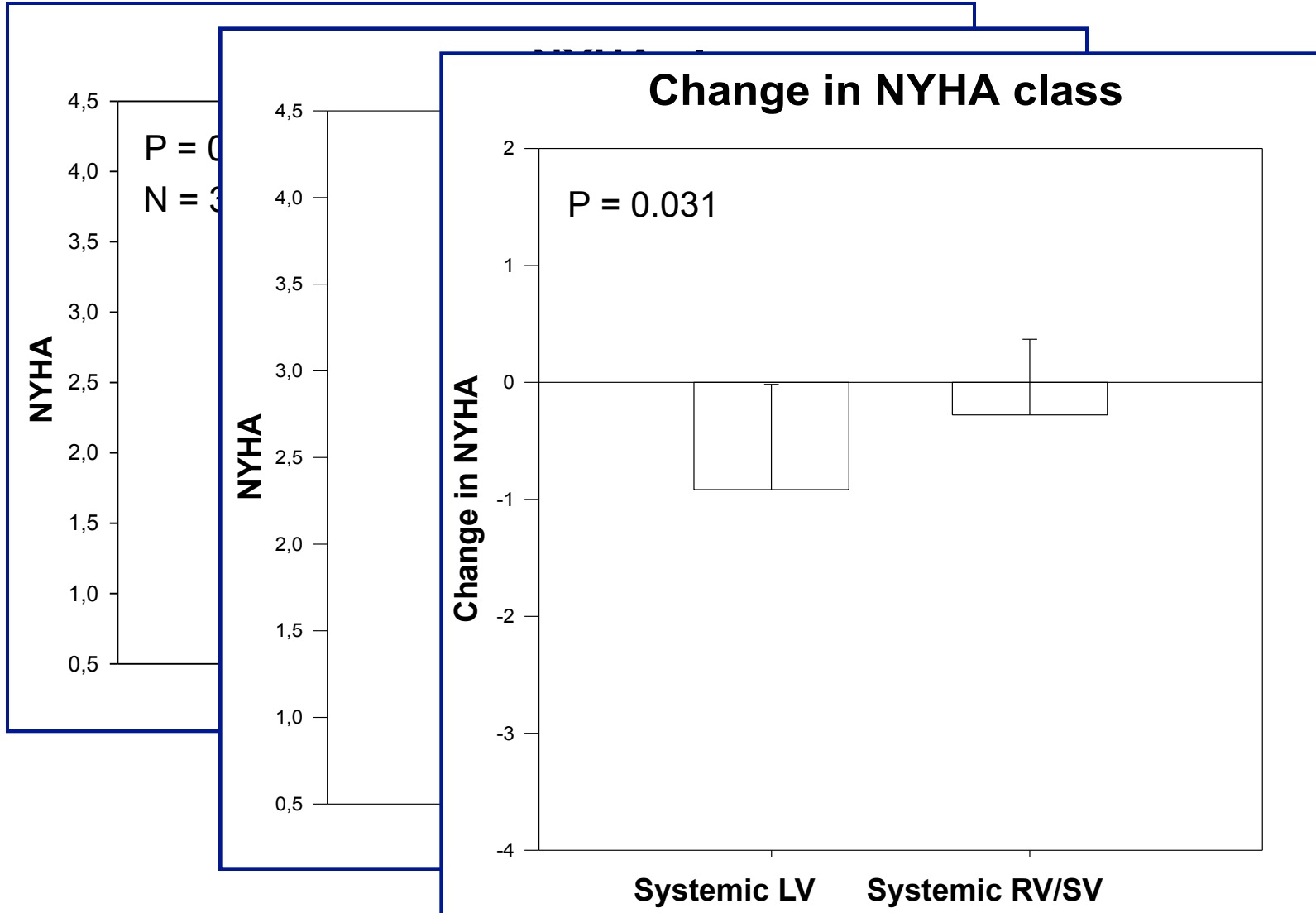
Ejection



Change in ejection fraction/fractional area of change

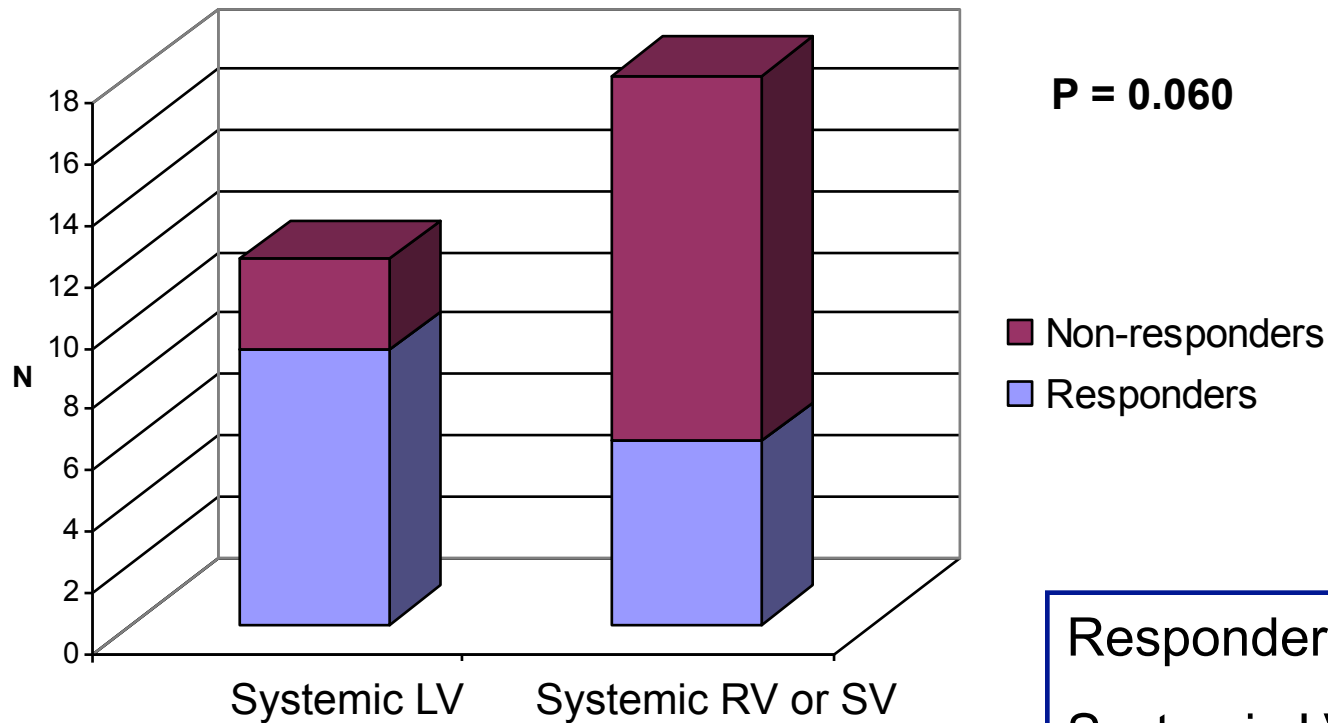


Results (V)



Results (VI)

Long term CRT response



Responders (15/30 = 50 %):

Systemic LV = 9/12

Systemic RV or SV = 6/18

Conclusion

- 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.
- Sudden death rate significant (10% in this cohort)
 - CRT-D should be individually considered in every patient.