Heart Failure in Adult CHD:

Systemic RV or Single Ventricle

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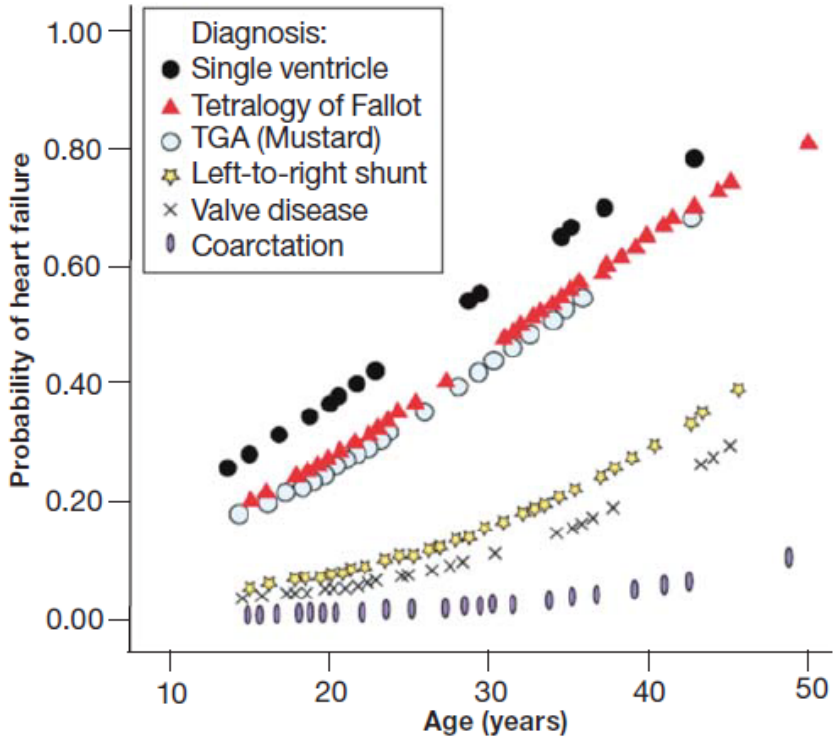
Heart Failure

Single ventricle or systemic RV

- incidence of heart failure 10 to 22%
- the probability of heart failure likely increasing over time.



Norozi K, AJC 2006;97:1238-43

Heart Failure

After a Mustard/Senning palliation

- One-third to one-half of patients have demonstrated reduced systemic RV function at 15 to 18 years follow up.

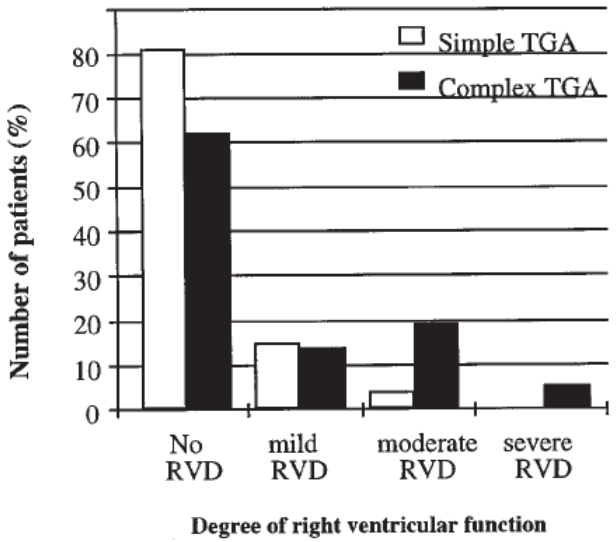


Fig 4. Right ventricular function at last follow-up, with a mean follow-up time of 12.8 years: echocardiographic assessment.

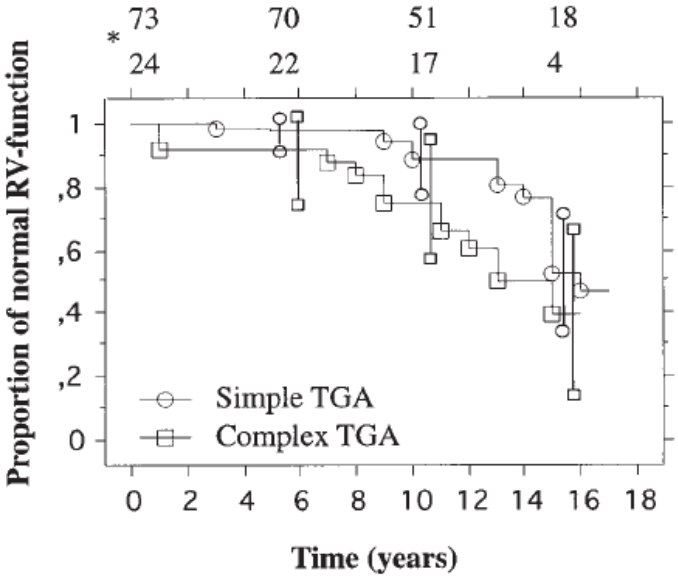


Fig 5. Probability of normal right ventricular function after the Senning operation. *Number of patients at risk. *Vertical lines* represent 95% CI at 5-, 10-, and 15-year follow-up. (Kaplan-Meier cumulative survival plot; $P = .03$, log-rank test.)

Kirjavainen M, JTCS. 1999;117:488-95

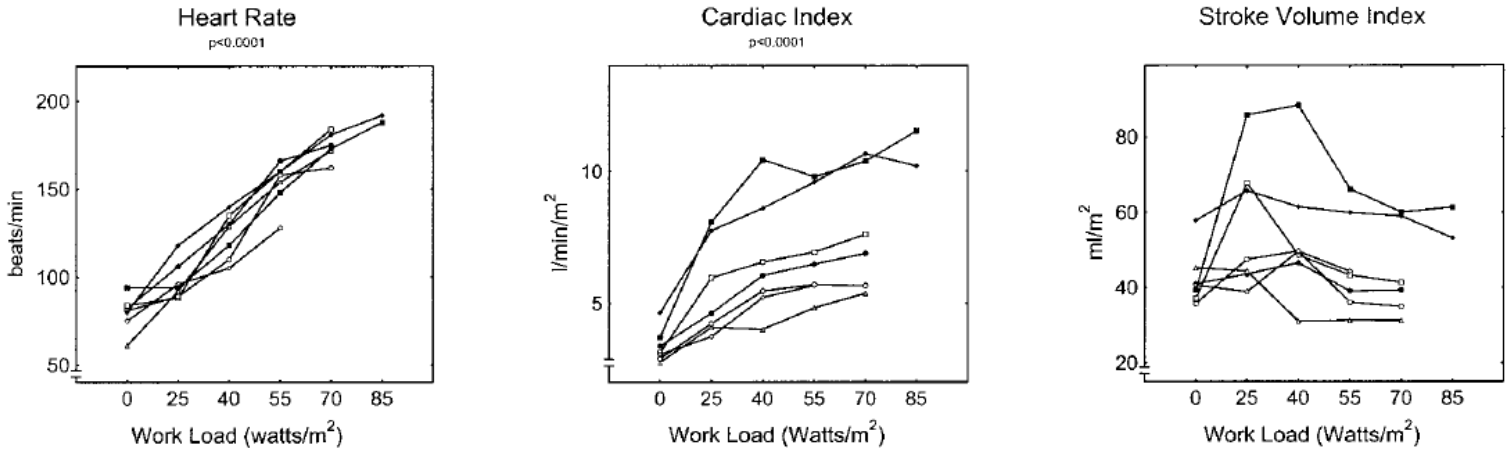
Heart Failure –Mechanism-

After Mustard operation

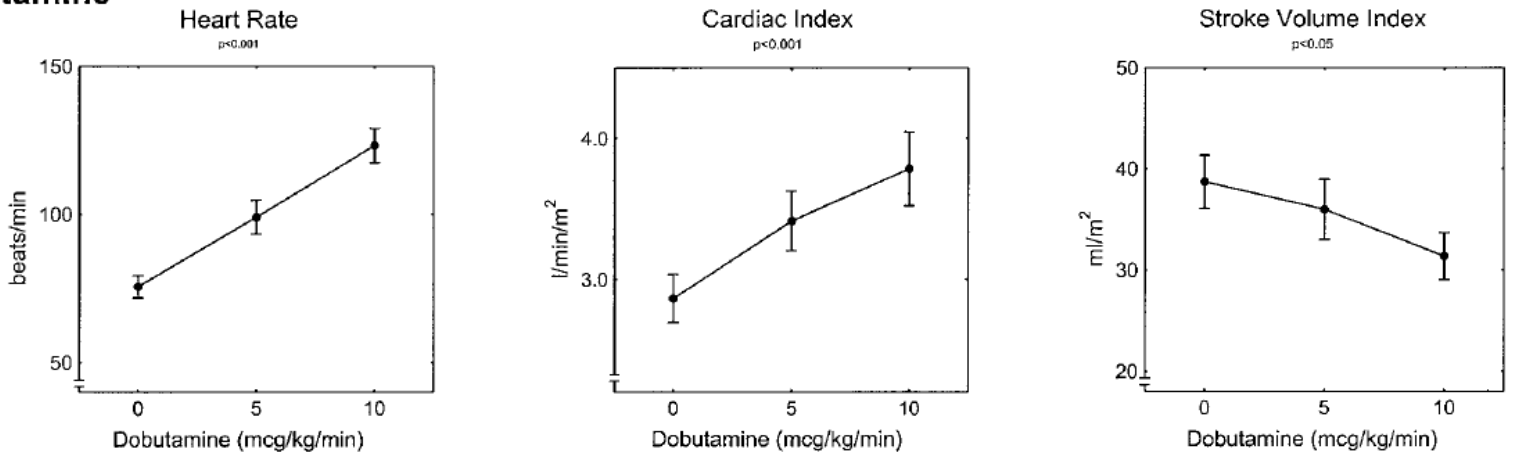
- Impaired increase in cardiac index and stroke volume in response to stress (exercise or dobutamine)
- Inability to augment ventricular filling with tachycardia

Heart Failure –Mechanism-

Exercise

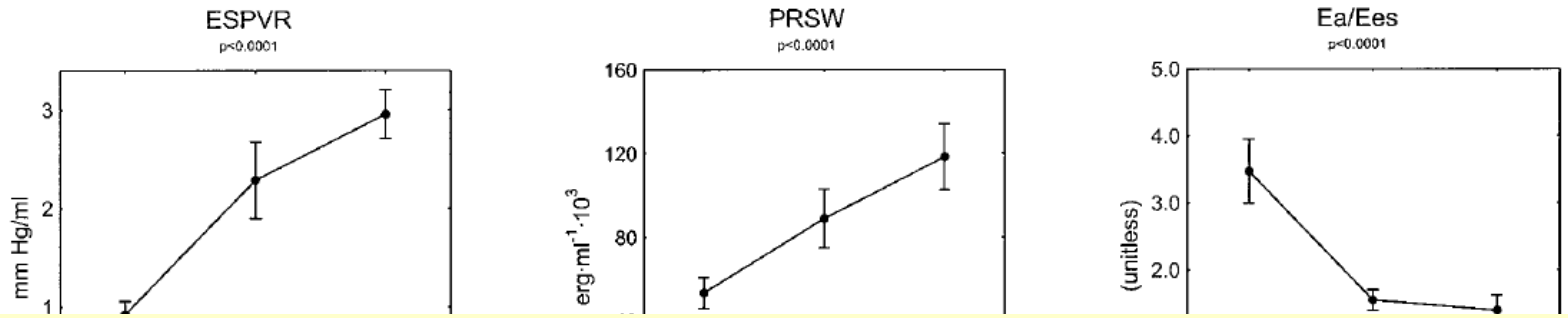


Dobutamine

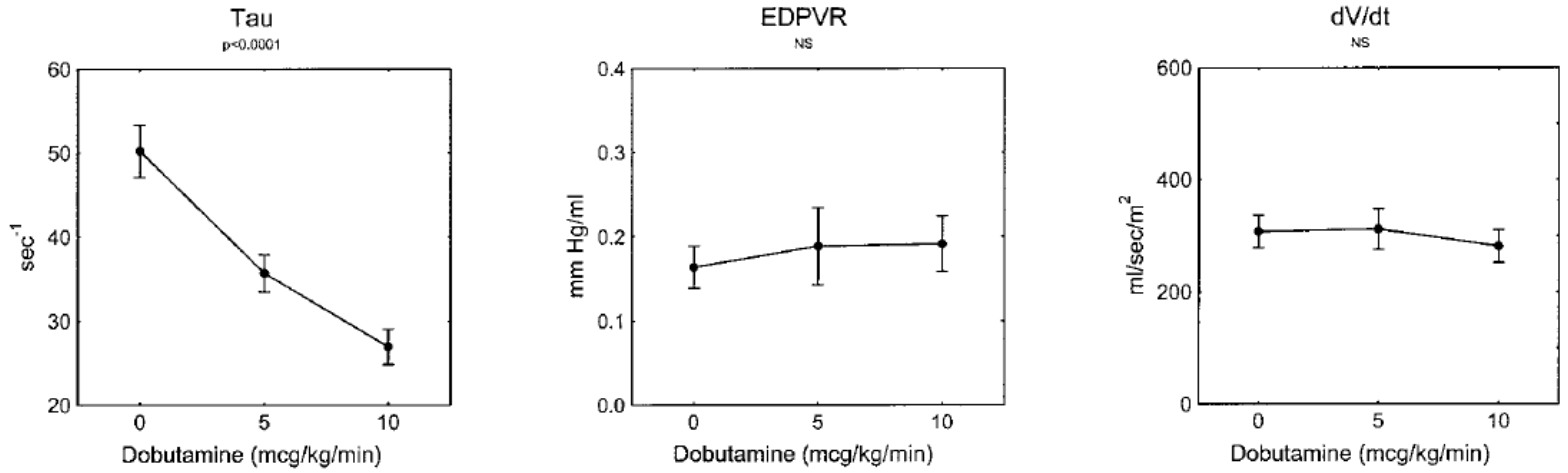


Derrick GP, Circulation 2000;102:III154-9

Heart Failure –Mechanism-



Failure to augment **right ventricular filling rates** during tachycardia, presumably as a result of **impaired AV transport**, consequent to the abnormal intra-atrial pathways

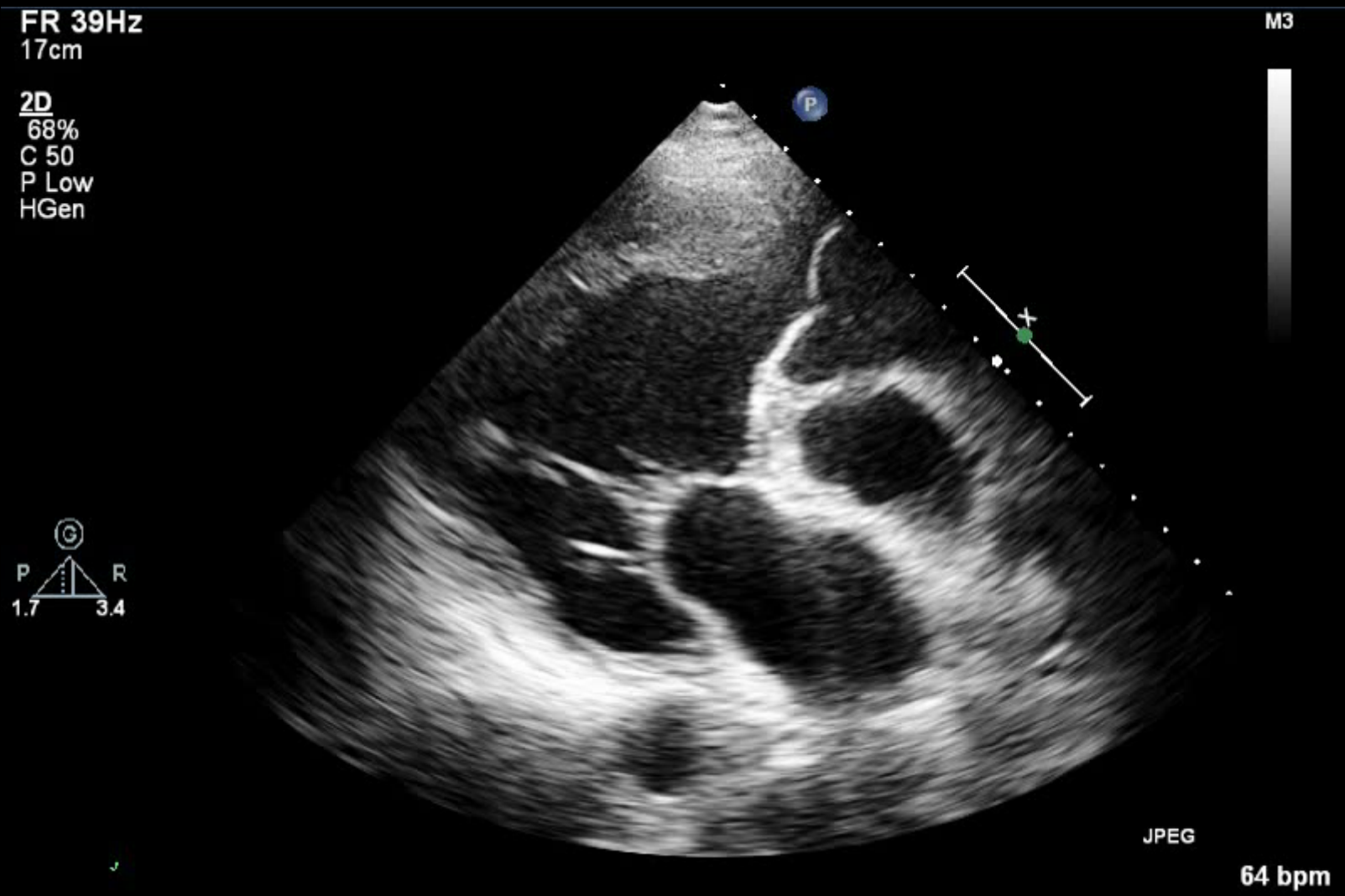


Derrick GP, Circulation 2000;102:III154-9

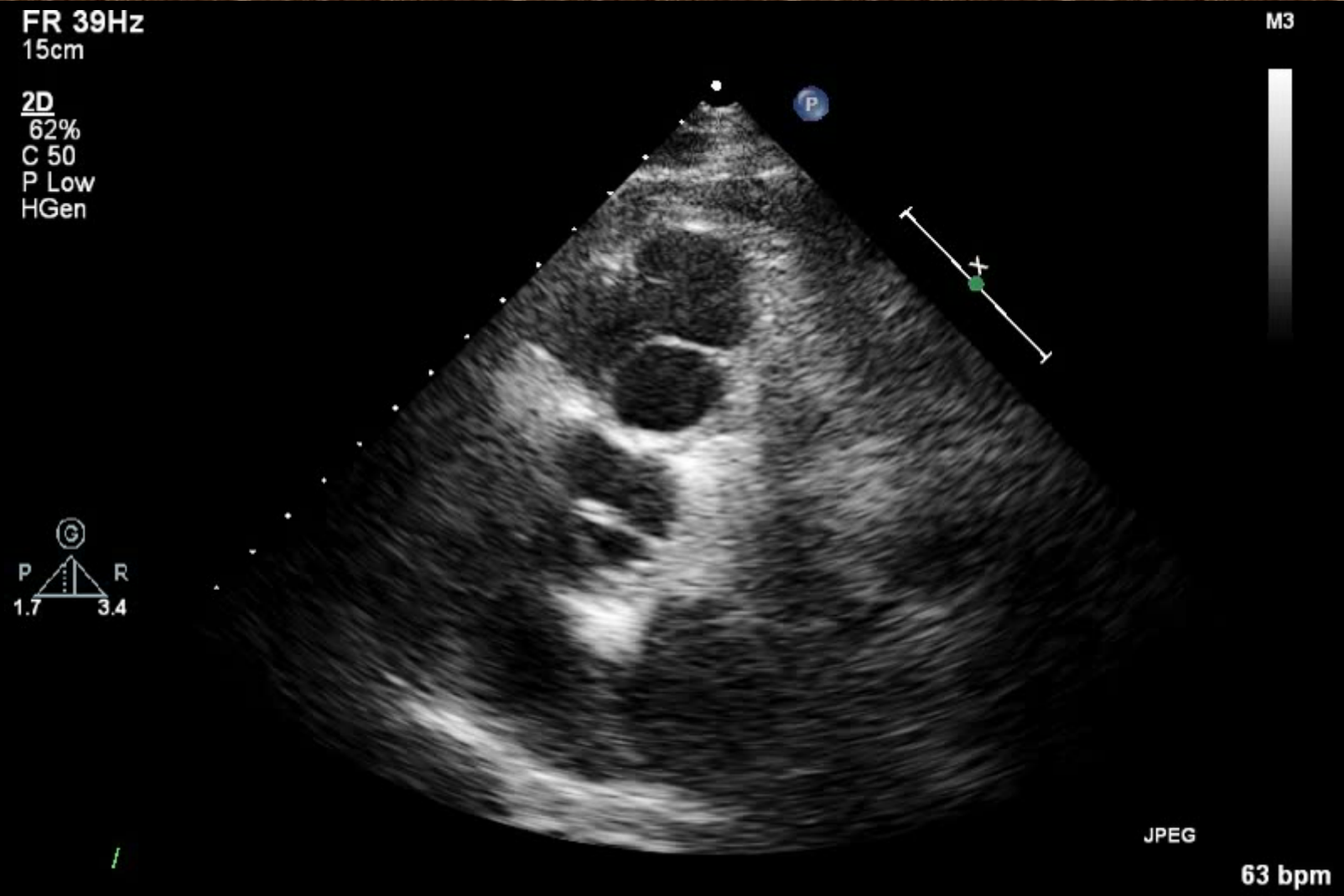
Systemic RV or Single Ventricle

Prognosis

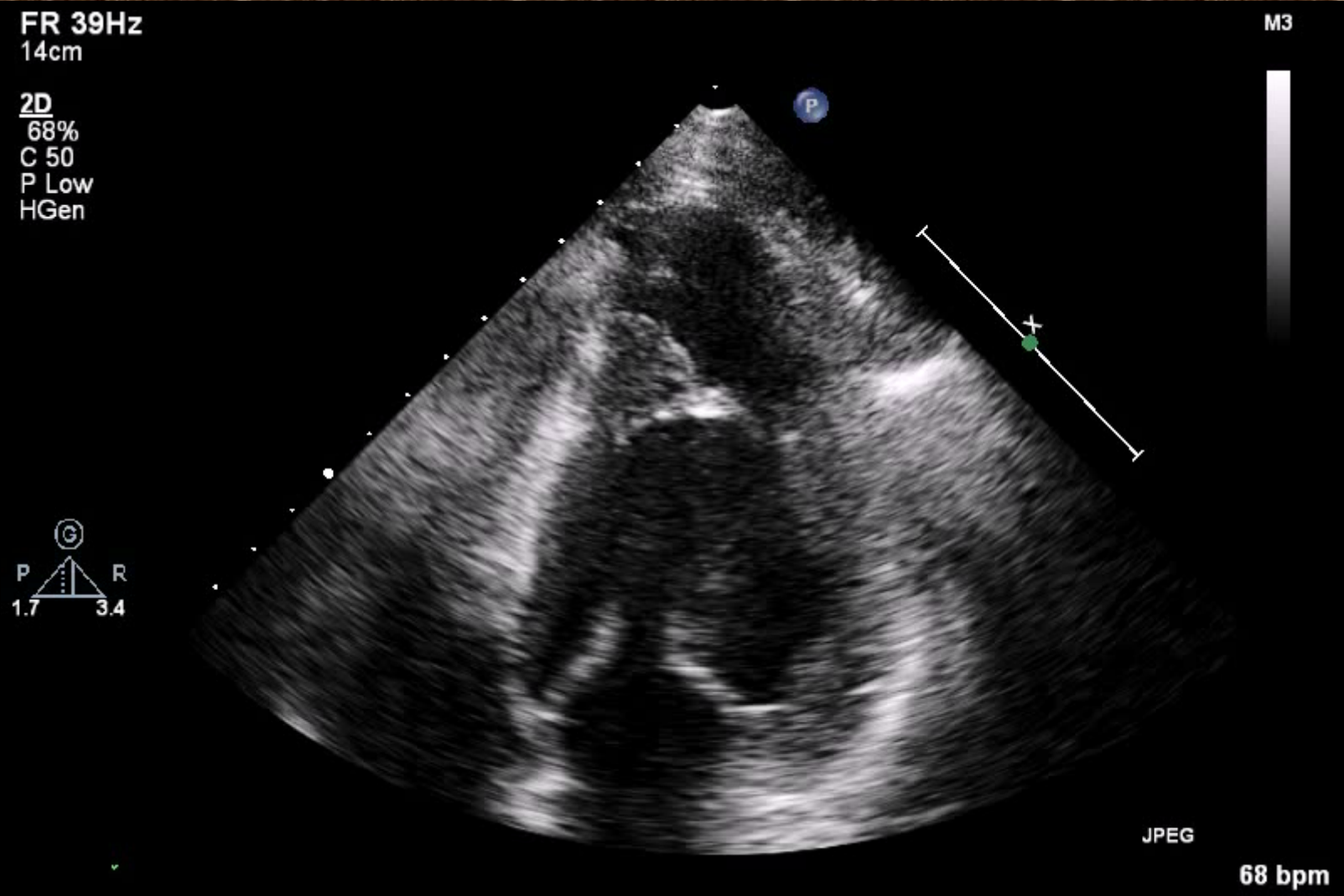
Cc-TGA



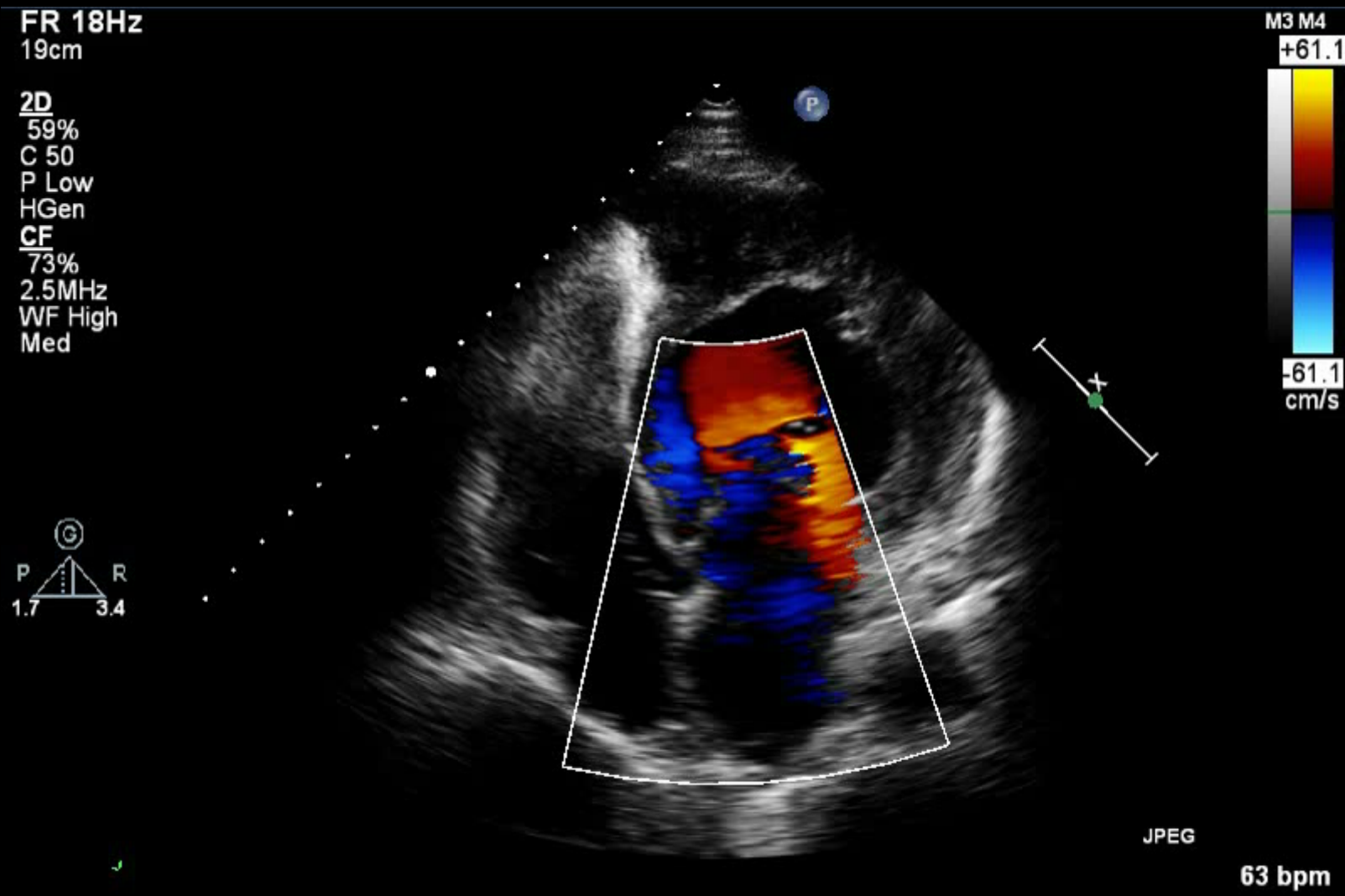
Cc-TGA



Cc-TGA



Cc-TGA


FR 18Hz
19cm
2D
59%
C 50
P Low
HGen
CF
73%
2.5MHz
WF High
Med
M3 M4
+61.1
-61.1
cm/s
P
R
1.7
3.4
JPEG
63 bpm

Systemic RV Dysfunction

Adults with cc-TGA

- Average systemic EF of 41%.

Table 2. Baseline Hemodynamics of Subjects Requiring Subsequent Surgical Intervention Versus Medically Treated Subjects (n = 44)

	Subsequent Surgery (30)	No Surgery (14)	p Value
Age, median (range), yr	44 (20–75)	43 (20–79)	0.71
SV EF, mean ± SD (range), %	40 ± 10 (23–65)	43 ± 8 (25–58)	0.16
SAVV regurgitation ≥3/4, no., %	24 (80)	2 (14)	0.0001
CT ratio, mean ± SD, %	0.57 ± 0.10	0.47 ± 0.07	0.005
Ability index ≥2, no., %	25 (83)	8 (57)	0.13
Functional capacity, mean ± SD, %*	74 ± 27	87 ± 24	0.37
Unable to do EST, no., %†	13 (43)	0 (0)	0.01

*Performance on cardiopulmonary testing, expressed as percent of expected when compared with predicted value for gender, age and body size; †Patient too ill to undergo exercise stress testing (EST) at presentation.

CT ratio = cardiothoracic ratio measured on chest radiograph; SAVV = systemic atrioventricular valve; SV EF = ejection fraction of the systemic ventricle.

Beauchesne LM, JACC 2002;40:285-90

Systemic RV Dysfunction

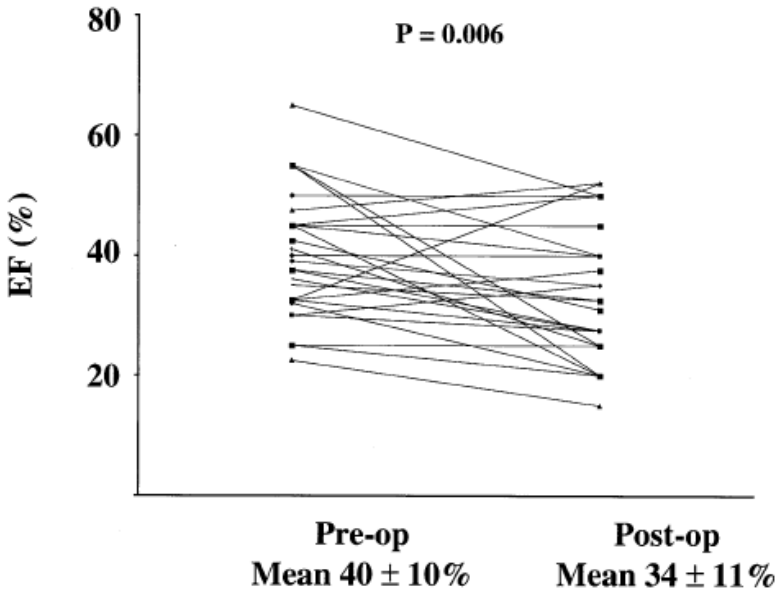


Table 4. Variables Predicting Need for Eventual Orthotopic Heart Transplantation in the Surgical Group*

Variable	p Value
Age at surgery	0.26
Functional capacity†	0.77
Ability index ≥2	0.21
CT ratio	0.31
Preoperative SV EF	0.001

*After initial surgical repair, four patients eventually required orthotopic transplantation—proportional hazards regression; †Performance on exercise stress testing at initial visit, expressed as percent of expected when compared with predicted value for gender, age and body size.

CT ratio = cardiothoracic ratio measured on chest radiograph; SV EF = ejection fraction of the systemic ventricle.

Beauchesne LM, JACC 2002;40:285-90

Systemic RV Dysfunction

cc-TGA associated with PS or VSD

- 70% have systolic dysfunction
- 30 to 50% have symptomatic heart failure

Table 4. Demographic and Clinical Variables by Patient Group

	Group I (Associated Lesions) (n = 132)	Group II (No Associated Lesions) (n = 50)	p Value
Age (yr, mean ± SD)	32 ± 12	34 ± 15	NS
Gender	37% female	52% female	NS
CHF	51%	34%	0.04
RV Dysfunction:			
Any	70%	55%	NS
Moderate or severe	39%	32%	NS
TR:			
Any	82%	85%	NS
Moderate or severe	57%	40%	NS
Pacemaker	45%	27%	0.04
Arrhythmia	47%	29%	0.04
Open heart surgery (excludes transplant)	70%	15%	0.001
LV Dysfunction	25%	7%	0.014
AR	36%	25%	NS

Graham TP Jr, JACC 2000;36:255-61.

Systemic RV Dysfunction

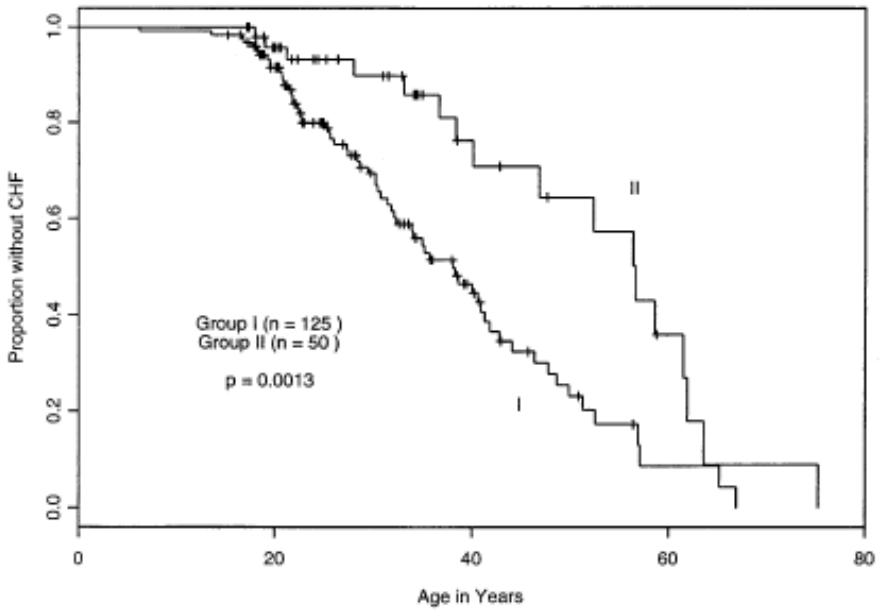


Figure 1. Probability of freedom from CHF for group I (associated lesions) and group II (no significant associated lesions) as a function of increasing age (N = 175 instead of 182 because it was unclear in 7 patients whether they had clinical CHF).

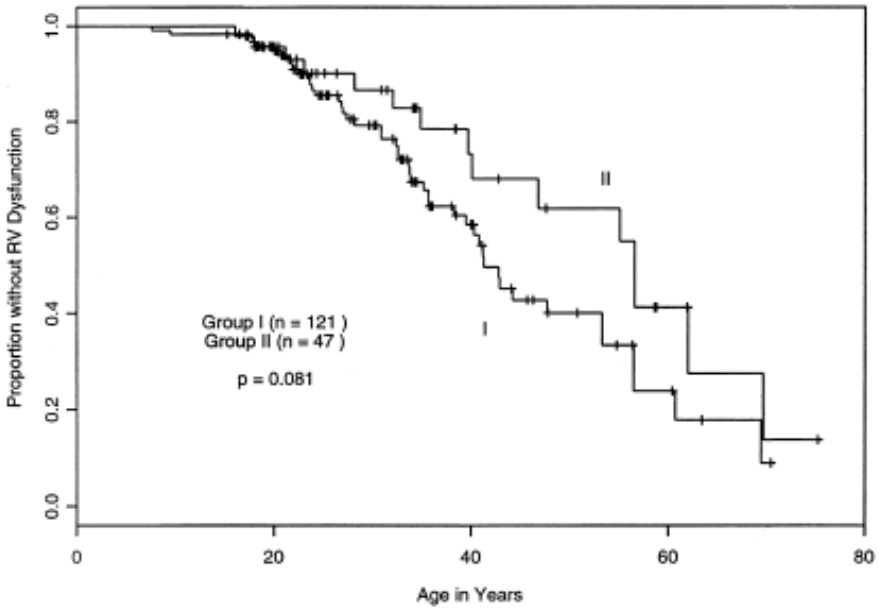
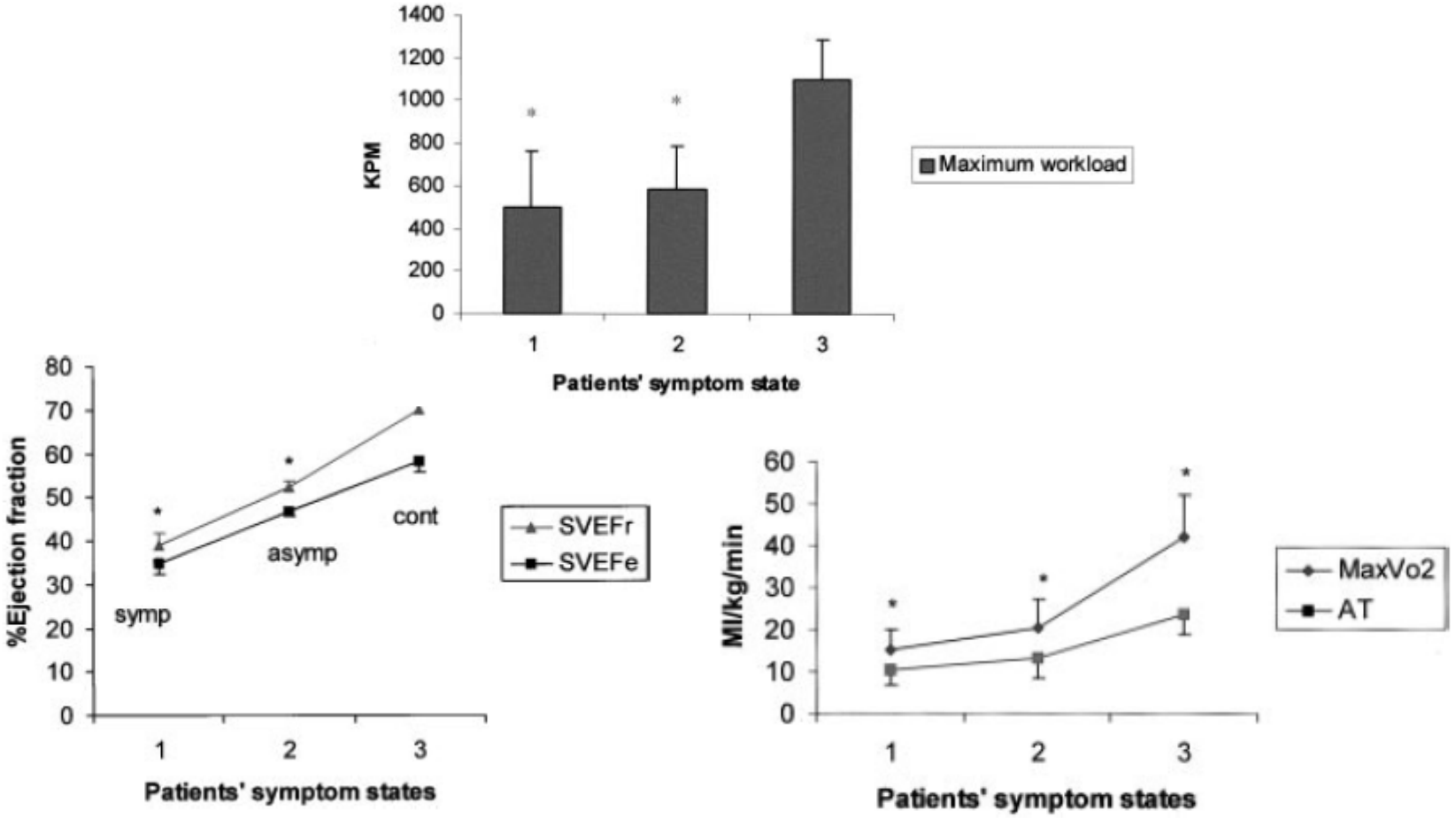


Figure 2. Probability of freedom from moderate or severe RV dysfunction as a function of increasing age. (N = 168 because data were unavailable to make this determination in 14 patients.)

Graham TP Jr, JACC 2000;36:255-61.

Single or Systemic Right Ventricles



Circulation. 2002;105:1189-1194

Single or Systemic Right Ventricles

Mortality

- 47.1% among symptomatic patients
- 5% among asymptomatic patients at 15.7 years of postoperative follow-up.

Best predictors for mortality

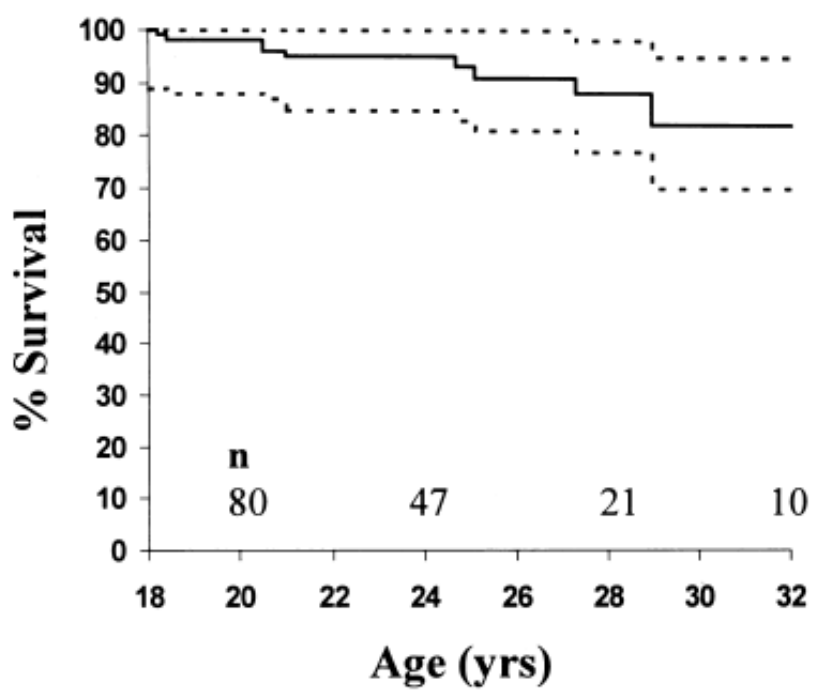
- New York Heart Association class
- Systemic ejection fraction
- Age at operation

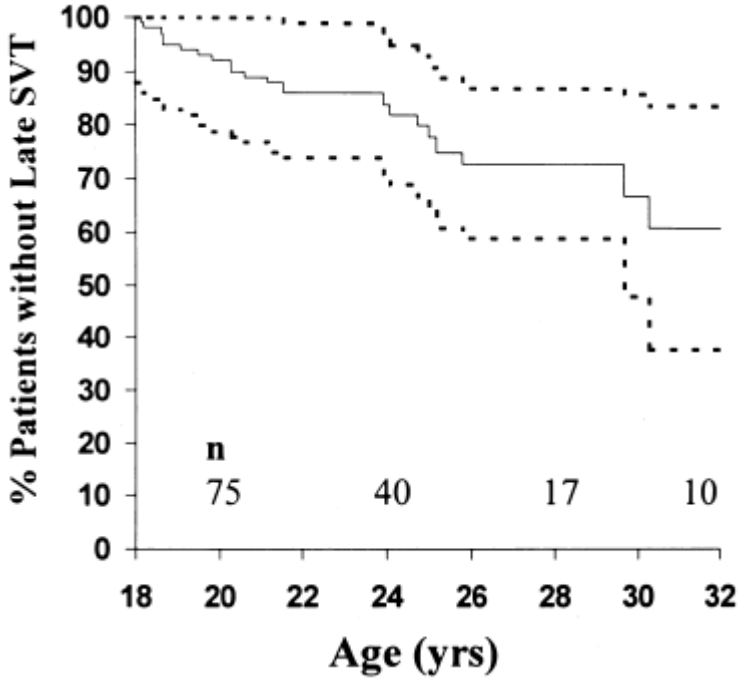
Circulation. 2002;105:1189-1194

Systemic Right Ventricle

Adults after Mustard procedure

- Pulmonary hypertension and systemic ventricular dysfunction were independent risk factors for death or CHF.



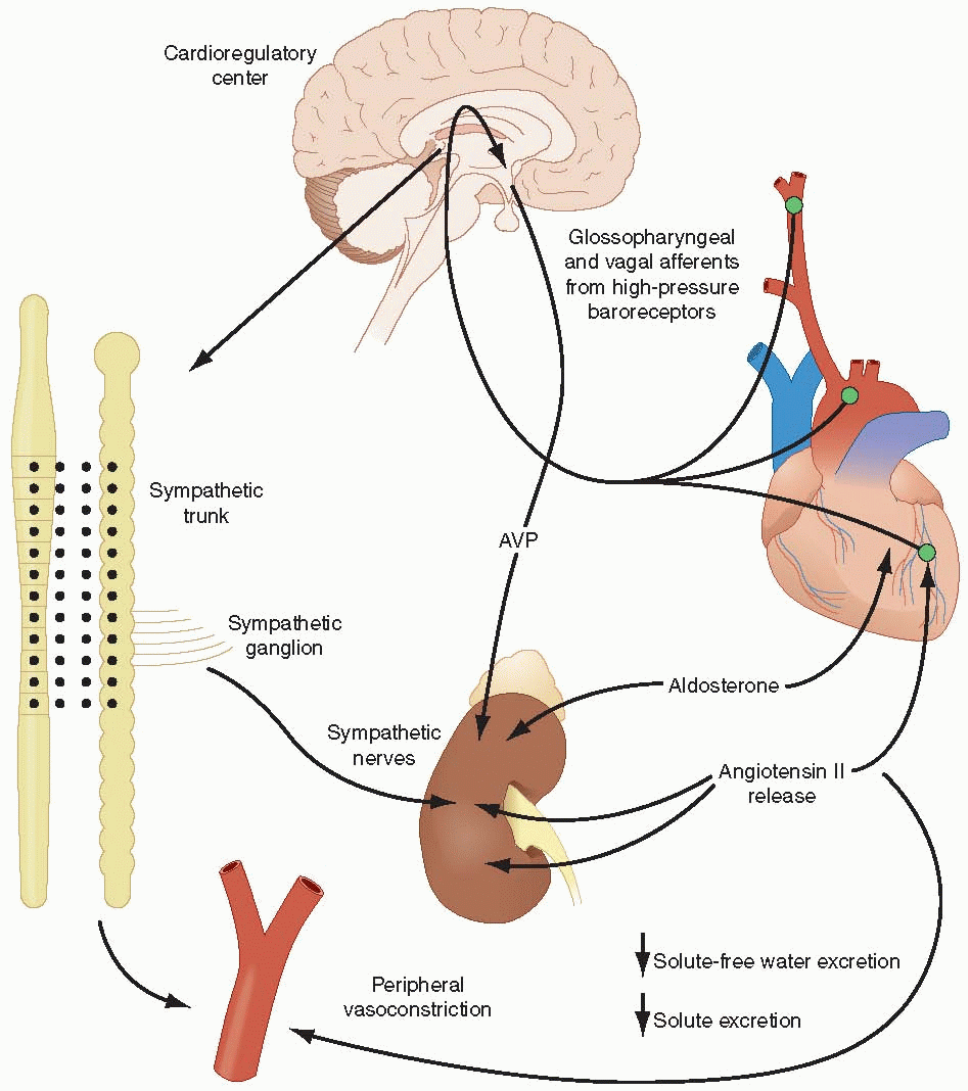


Puley G, AJC 1999;83:1080 –4

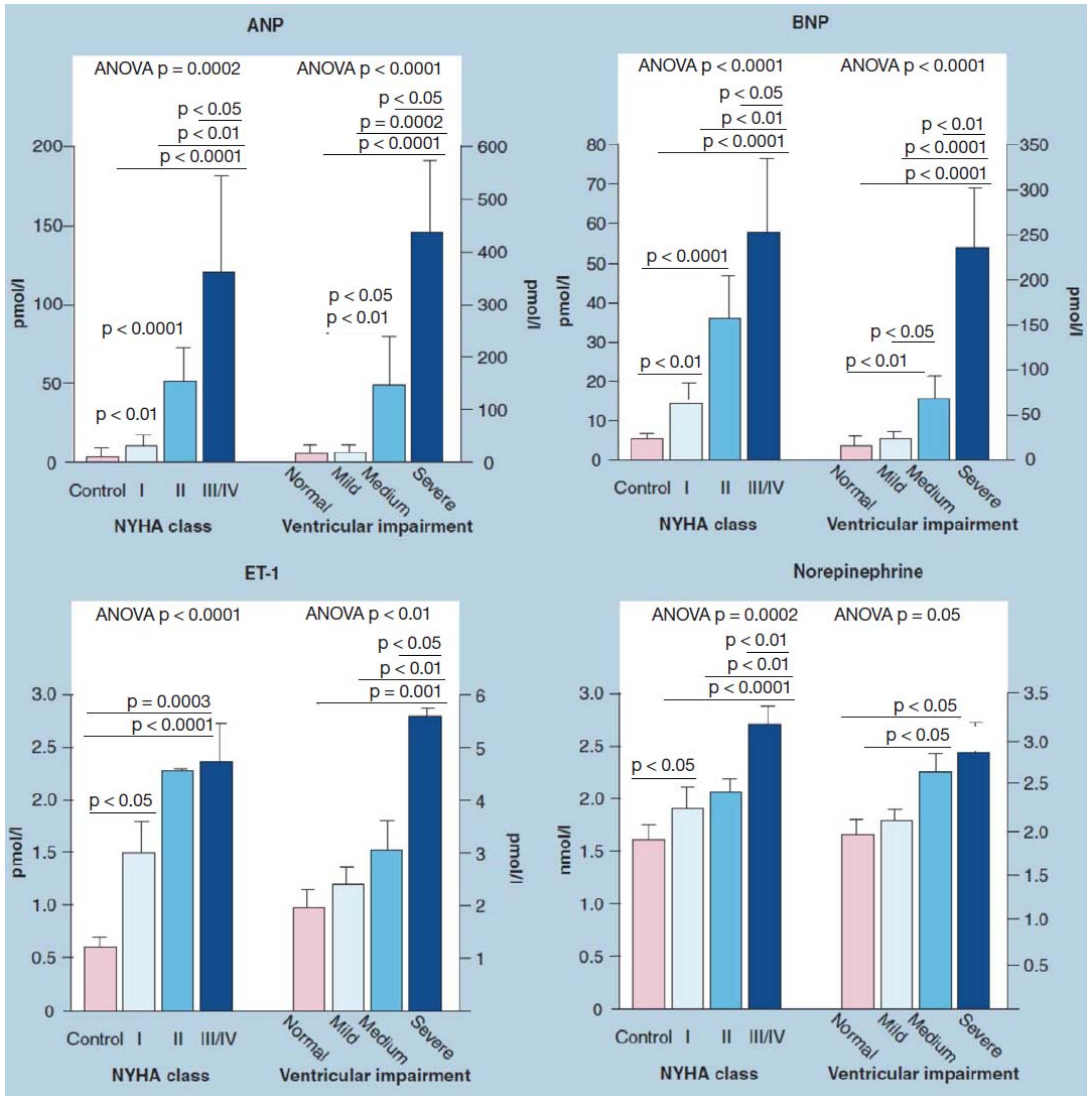
Systemic RV or Single Ventricle

Treatment for HF

Neurohumoral Mechanism



Neurohumoral Activation



Bolger AP, Circulation. 2002;106:92-9

Diuretics

Mechanism

- Control fluid retention in advanced HF

- Furosemide, torsemide, and bumetanide act at the loop of Henle (loop diuretics)
  - Reversibly inhibit the reabsorption of Na+, K+, and Cl- in the thick ascending limb of Henle's loop
- Thiazides and metolazone
  - Reduce the reabsorption of Na+ and Cl- in the first half of the distal convoluted tubule
- Potassium-sparing diuretics (spironolactone)
  - Act at the level of the collecting duct

Diuretics

❍Potency and pharmacologic properties

- **Loop diuretics**
  - ➢ Increase the fractional excretion of sodium by 20-25%
  - ➢ Generally required to restore normal volume status in patients with HF
- Thiazide diuretics
  - ➢ Increase it by only 5-10%
  - ➢ Loose their effectiveness in patients with moderate or severe renal insufficiency (creatinine >2.5 mg/dL)

Diuretics

In CHD

- The balance between adequate volume status and pulmonary perfusion
  - Fontan palliation
    - Passive, nonpulsatile filling for preload of the systemic chamber
  - Shunt-dependent patients
    - Driving pressure and volume

CONSENSUS: Cooperative North Scandinavian Enalapril Survival Study
- TRIAL DESIGN -

Design

Multicenter, multinational, randomized, double-blind, placebo-controlled

Patients

253 patients with severe congestive heart failure (**NYHA class IV**) and heart size >600 (men) or >500 mL/m^2 (women), and receiving a diuretic and digoxin; patients with MI in previous 2 months excluded

Follow up and primary endpoint

Primary endpoint: all-cause mortality. Mean 188 days follow up

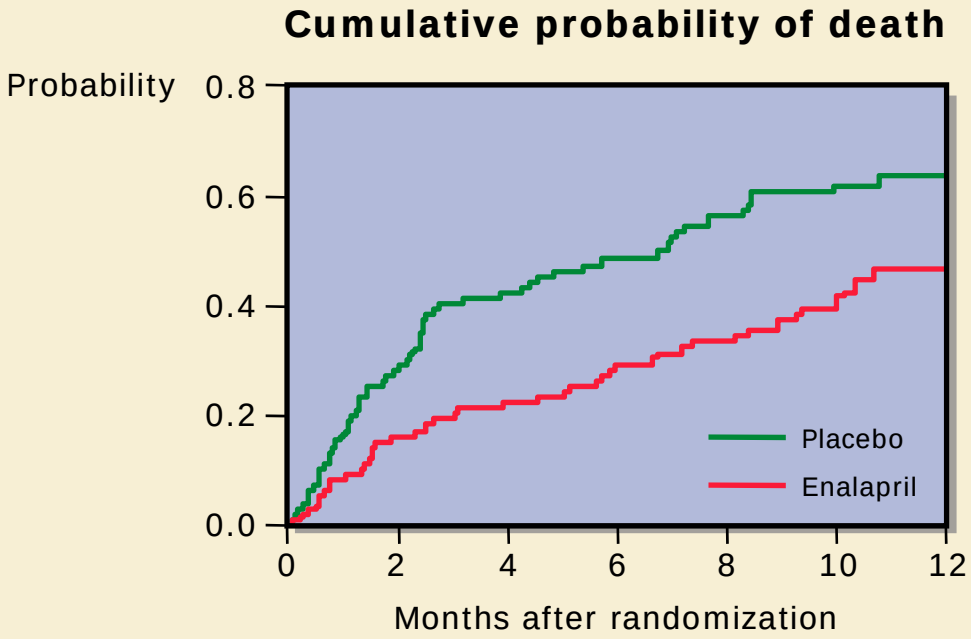
Treatment

Placebo or enalapril initiated at 5 mg twice daily; increased to 10 mg twice daily after 1 week if no side effects, then to maximum 20 mg twice daily according to clinical response

CONSENSUS: Cooperative North Scandinavian Enalapril Survival Study
- RESULTS -

- Trial halted early on recommendation of Ethical Review Committee because of evident benefit of enalapril
- Significant reduction in **all-cause mortality** in enalapril group at 6 months and 1 year, with overall relative risk reduction of **27%** (39 vs. 54%, P=0.003)
- Reduction in mortality entirely attributed to reduction in **death due to progression of heart failure**
- No difference in incidence of sudden cardiac death within the two groups
- NYHA class improved in significantly higher proportion of enalapril group (42 vs. 22%, P<0.001)
- Withdrawal due to hypotension higher in enalapril group, but overall withdrawal rate similar in the two groups

CONSENSUS: Cooperative North Scandinavian Enalapril Survival Study
- RESULTS continued -



Placebo:	126	78	59	47	34	24	17
Enalapril:	127	98	82	73	59	42	26

CONSENSUS Trial Study Group. *N Engl J Med* 1987;**316**:1429–35.

SOLVD: Studies Of Left Ventricular Dysfunction
- TRIAL DESIGN -

Design

Multicenter, multinational, randomized, double-blind, placebo-controlled

Patients

2569 clinically stable patients with chronic CHF and ejection fraction ≤0.35, approximately 90% in **NYHA classes II and III**; patients with MI in previous month excluded

Follow up and primary endpoint

Average 41.4 months follow up. Primary endpoints mortality and hospitalization for worsening heart failure

Treatment

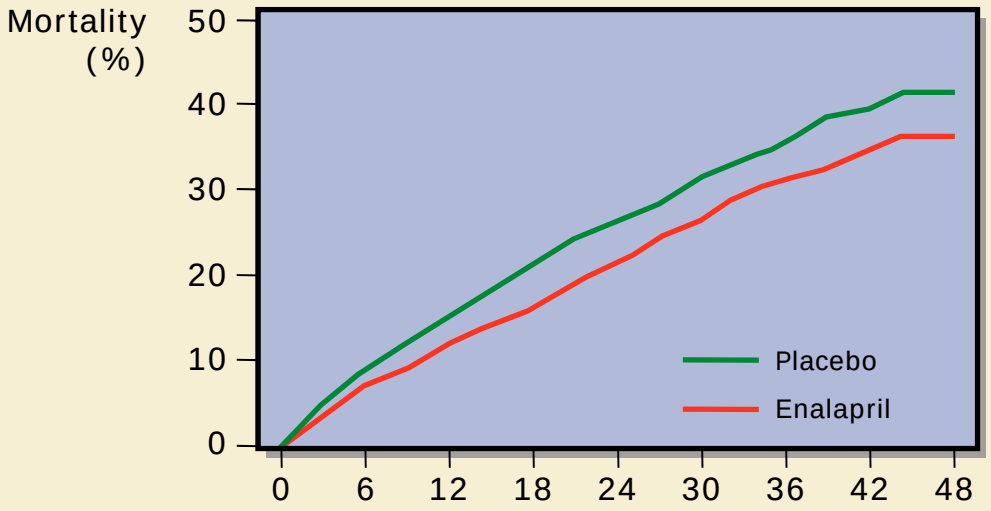
Patients assigned enalapril received 2.5 or 5 mg twice daily initially, then 2.5–20 mg per day

SOLVD: Studies Of Left Ventricular Dysfunction
- RESULTS -

- **All-cause mortality** and **death or hospitalization due to heart failure** significantly reduced in enalapril group compared with placebo
- Significant reduction in several categories of death due to cardiovascular causes, majority attributable to **reduction in progressive heart failure**
- Benefit in terms of death or hospitalization due to heart failure significantly smaller for highest tertile baseline ejection fraction
- No significant difference in MI in placebo and enalapril groups
- Most common side effects hypotension and increased serum creatinine

SOLVD: Studies Of Left Ventricular Dysfunction
- RESULTS continued -

Cumulative all-cause mortality



No. of patients alive									
Placebo	1284	1159	1085	1005	939	819	669	487	299
Enalapril	1285	1195	1127	1069	1010	891	697	526	333

The SOLVD Investigators. *N Engl J Med* 1991;**325**:293–302.

SOLVD: Studies Of Left Ventricular Dysfunction - RESULTS continued -

Death and hospitalization for CHF

	Placebo n=1284 (%)	Enalapril n=1285 (%)	% Risk reduction (95% CI)	One-sided P
Death due to any cause	39.7	35.2	16 (5–26)	<0.0036
Death or hospitalization for CHF	57.3	47.7	26 (18–34)	<0.0001
Cardiovascular death [a]	35.9	31.1	18 (6–28)	<0.002
Cardiac death	34.3	29.3	19 (7–29)	<0.0015
Arrhythmia without worsening CHF	8.8	8.2	10 (-17–31)	–
Heart failure or arrhythmia with CHF	19.5	16.3	22 (6–35)	<0.0045

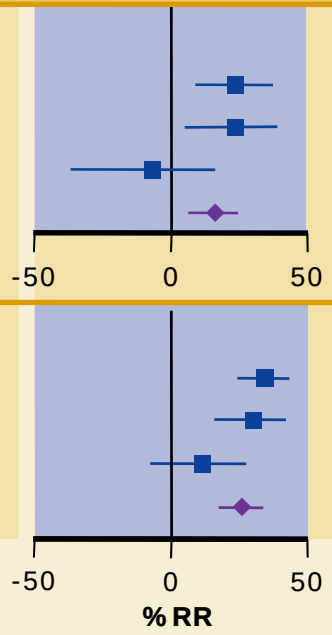
[a] Cardiac causes (including MI), stroke and other vascular causes

The SOLVD Investigators. *N Engl J Med* 1991;**325**:293–302.

SOLVD: Studies Of Left Ventricular Dysfunction
- RESULTS continued -

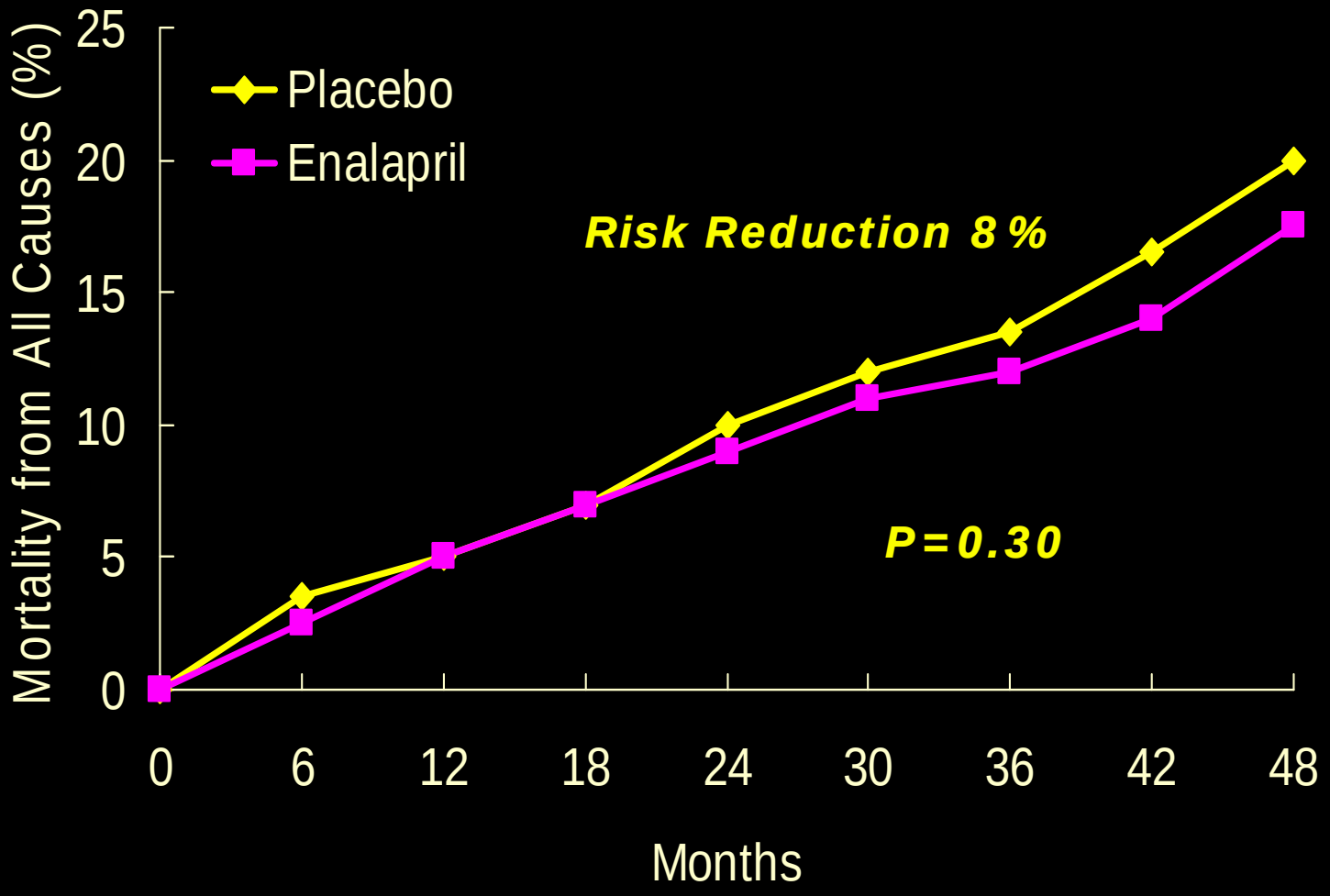
Effect of enalapril on ejection fraction subgroups (% of patients)

		Placebo n=1284	Enalapril n=1285	RR (%)
Death	**Ejection fraction (%)**			
	6–22	50	41	24
	23–29	39	33	24
	30–35	28	31	-7
	Overall	40	35	16
Death or hospitalization	**Ejection fraction (%)**			
	6–22	69	52	35
	23–29	56	47	30
	30–35	45	44	12
	Overall	57	48	26



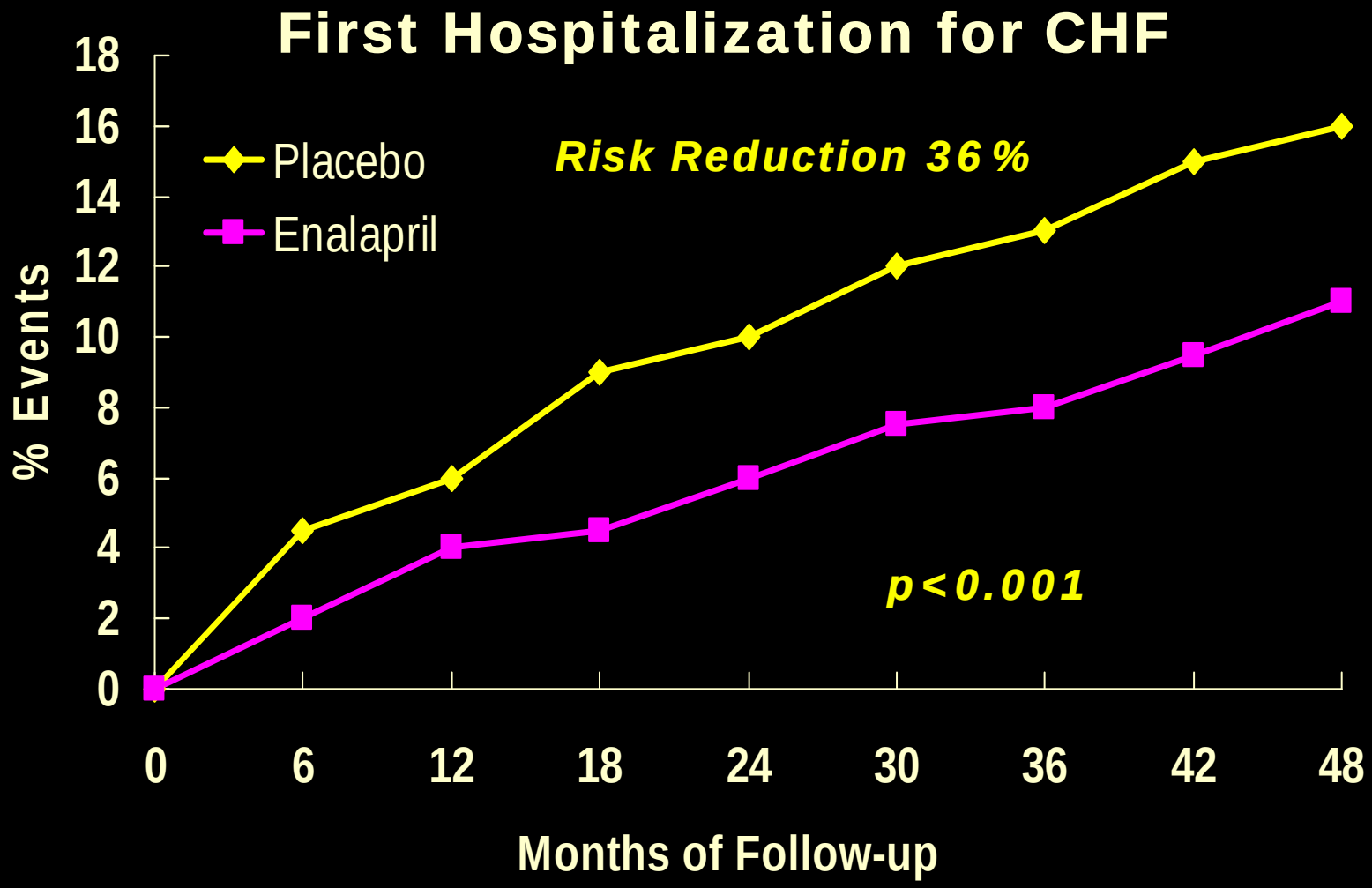
The SOLVD Investigators. *N Engl J Med* 1991;**325**:293–302.

SOLVD Prevention Trial



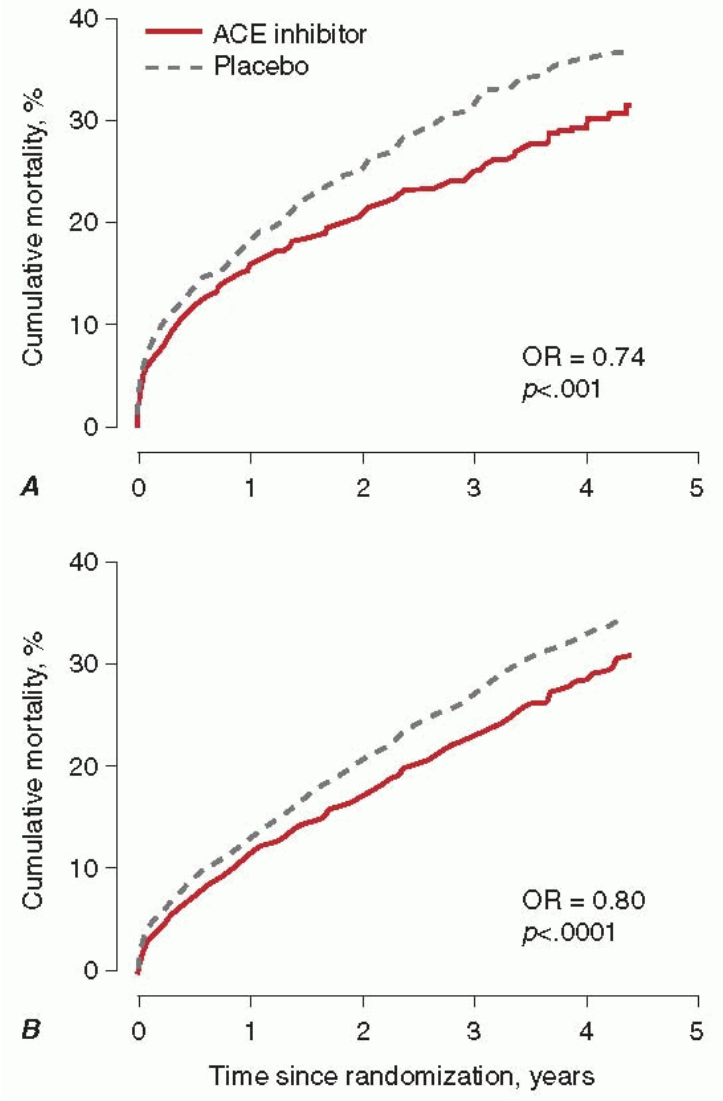
N Engl J Med 1992;327:685-91

SOLVD Prevention Trial



N Engl J Med 1992;327:685-91

ACE Inhibitors in HF



Acute myocardial infarction
(SAVE, AIRE, and TRACE trials)

HF with depressed EF

Flather, Lancet 2000;355:1575

ACE Inhibitors

Adverse Effects

- Decreases in blood pressure and mild azotemia that may occur during the initiation of therapy
  - Generally well tolerated
- Potassium retention
- Nonproductive cough (10-15%), angioedema (1%), skin rash
  - Kinin potentiation
  - Angiotensin receptor blockers (ARBs) are the recommended first line of therapy

ACE Inhibitors

- After Mustard procedure
- Cardiopulmonary exercise test
- Cardiac MRI

TABLE 1 Cardiopulmonary Study Data at Rest Before and After ACE Inhibitor Therapy (minimum of 6 months on ACE inhibitors)

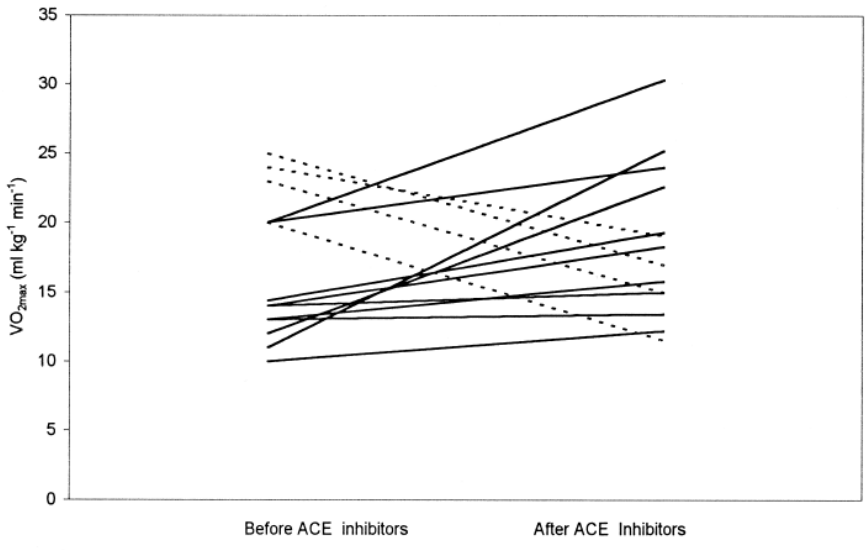
Variable	No.	Before ACE	After ACE	p Value
Forced vital capacity[1]	14	4.2 ± 0.85	4.1 ± 0.91	0.310
Forced vital capacity % predicted	14	80 ± 8	78 ± 9	0.214
Heart Rate (beats/min)	14	75 ± 13	77 ± 15	0.970
Systolic blood pressure (mm Hg)	14	114 ± 11	120 ± 13	0.340
Diastolic blood pressure (mm Hg)	14	68 ± 8	72 ± 12	0.290
Left ventricular ejection fraction (%)	14	58 ± 10	59 ± 12	0.609
Right ventricular ejection fraction (%)	14	47 ± 11	45 ± 11	0.608

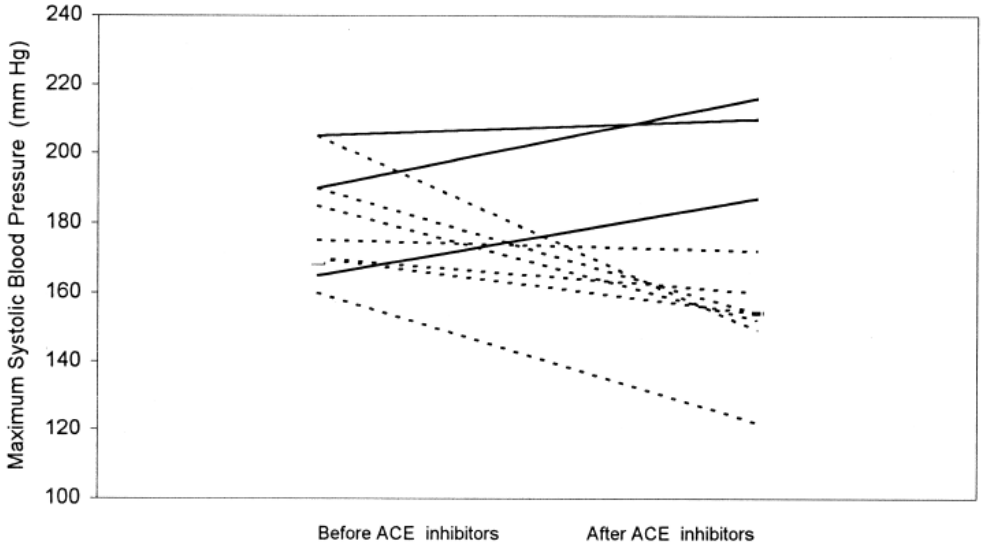
TABLE 2 Cardiopulmonary Study Data at Maximal Exercise Before and After ACE Inhibitor Therapy (minimum of 6 months on ACE inhibitors)

Variable	No.	Before ACE	After ACE	p Value
Exercise time (min)	14	6.6 ± 1.8	7.0 ± 1.9	0.58
Heart rate$_{max}$ (beats/min)	14	148 ± 21	144 ± 27	0.340
Systolic blood pressure$_{max}$ (mm Hg)	14	178 ± 16	166 ± 28	0.148
Diastolic blood pressure$_{max}$ (mm Hg)	14	84 ± 14	85 ± 16	0.740
Maximum oxygen uptake ($ml \cdot kg^{-1} \cdot min^{-1}$)	14	16.7 ± 5.1	18.5 ± 5.4	0.360
Ventilation ($L \cdot min^{-1}$)	14	61.4 ± 18.6	63.9 ± 23.9	0.554
Left ventricular ejection fraction$_{max}$ (%)	14	58 ± 10	59 ± 12	0.609
Right ventricular ejection fraction$_{max}$ (%)	14	47 ± 11	45 ± 11	0.608

Hechter SJ, AJC 2001;87:660–3

ACE Inhibitors





Hechter SJ, AJC 2001;87:660–3

Angiotensin Receptor Blockers

- **Mechanism**
  - Block the effects of angiotensin II on the angiotensin type 1 receptor
- **Alternative therapy to ACE inhibitors**
- **ACE inhibitors + ARBs**
  - Benefit in some trial
- **Beta blockers + ARBs**
  - Reverse the process of LV remodeling
  - Improve patient symptoms
  - Prevent hospitalization
  - Prolong life

Angiotensin Receptor Blockers

❍Losartan

- Seven patients ≥13 years of age
- Surgically palliated TGA
- Who had never received vasodilator therapy

TABLE 1 Summary of Effects of Losartan

Parameter	Immediately Before Losartan Therapy	After 8 Weeks of Losartan Therapy	p Value
Blood pressure (mm Hg)			
Systolic	117 ± 6	107 ± 13	0.04
Diastolic			NS
Ejection fraction (%)	48 ± 10	54 ± 7	0.04
EROA (mm^2)	12.9 ± 6.4	6.3 ± 6.4	0.02
Regurgitant volume (mL)	22.5 ± 11.1	8.2 ± 11.3	0.01
Right ventricular dp/dt (mm Hg/s)			NS
Right ventricular ejection time (ms)			NS
Acceleration time (ms)	147 ± 28	119 ± 19	0.05
Duration of exercise (min)	11.2 ± 2.9	13.2 ± 3.7	0.02

Values are expressed as mean ± SD.

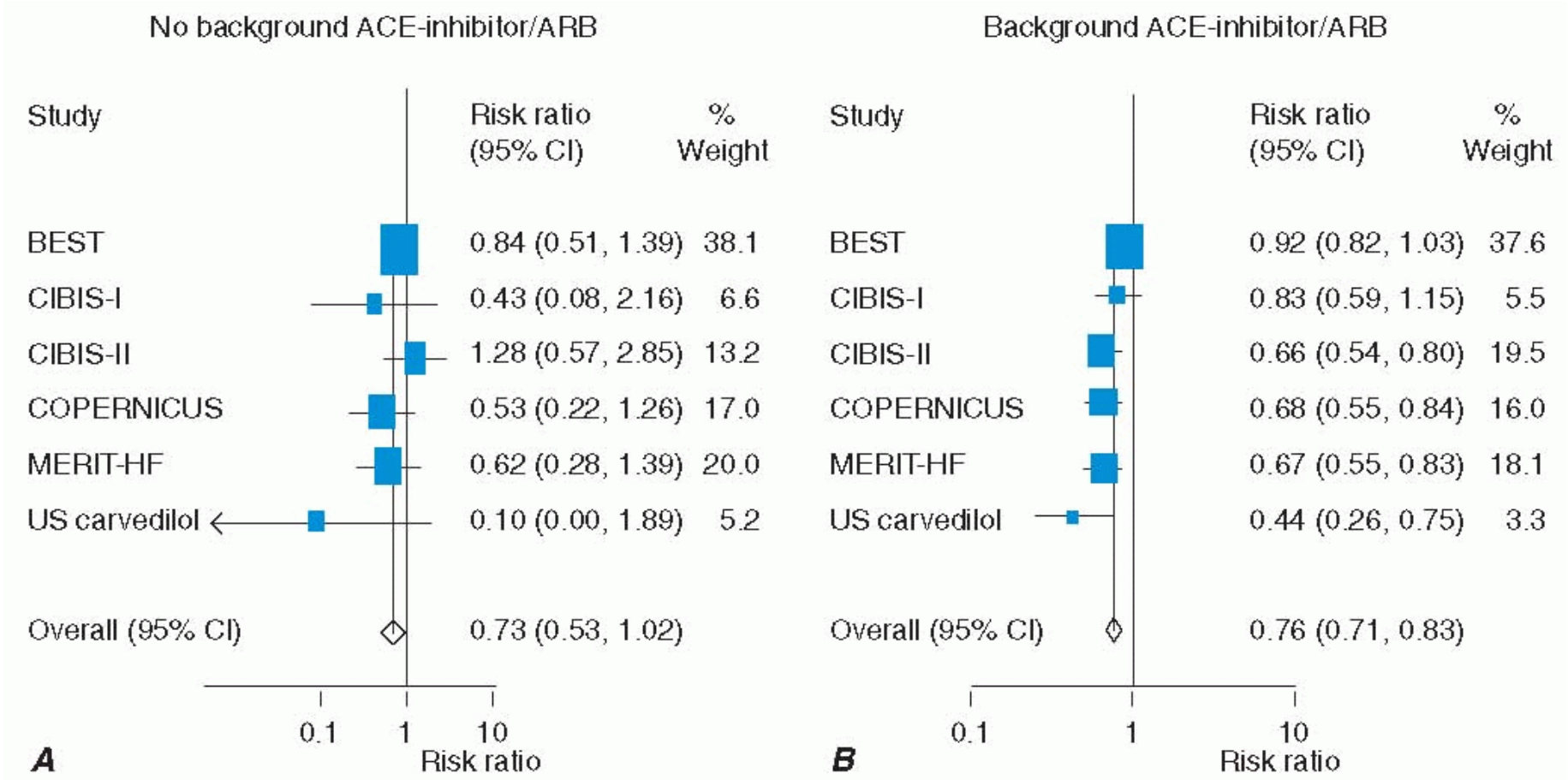
dp/dt = change in pressure over the change in time (rate of rise of ventricular pressure); EROA = effective regurgitant orifice area of systemic atrioventricular valve.

Lester SJ, AJC 2001;88:1314-6

Beta-Blockers

- **Mechanism**
  - Interfere with the harmful effects of sustained activation of the adrenergic nervous system
- **ACE inhibitors + beta blockers**
  - Reverse the process of LV remodeling
  - Improve patient symptoms
  - Prevent hospitalization, and prolong life.
- **Dose**
  - Should be initiated in low doses followed by gradual increments in the dose (more than 2-week intervals)

Beta Blockers in HF



Krum, Eur Heart J 2005;26:2154

Beta-Blockers

Adverse effects

- Beta-blocker therapy is well tolerated by the great majority (≥85%) of HF patients

- Bradycardia and/or exacerbate heart block
- Worsening fluid retention or symptomatic hypotension
  - Generally occur within several days of initiating therapy
  - Generally responsive to adjusting concomitant medications

Beta-Blockers

Table 1
Baseline characteristics

Variable	β-Blocker Therapy		p Value
	Yes (n = 31)	No (n = 29)	
Men	20 (65%)	18 (62%)	NS
Women	11 (35%)	11 (38%)	NS
Age (yrs)	29 ± 6	27 ± 6	NS
Age at surgery (yrs)	1.5 ± 1.4	1.3 ± 1.2	NS
Other drugs			
Angiotensin-converting enzyme inhibitors	14 (45%)	12 (41%)	NS
Angiotensin receptor blockers	2 (6%)	1 (3%)	NS
Aldactone	11 (35%)	2 (7%)	<0.01
Digoxin	13 (42%)	15 (52%)	NS
Diuretic	11 (35%)	4 (14%)	NS
Pacemaker	24 (77%)	12 (41%)	<0.01
Pacing indications			
Sick sinus syndrome	13 (42%)	10 (35%)	
Atrioventricular block	2 (6%)	2 (7%)	
Paroxysmal atrial fibrillation	9 (29%)	0 (0%)	
Pacing modes			
DDD	12 (39%)	10 (35%)	
VVI	3 (10%)	0 (0%)	
AAI	9 (29%)	2 (7%)	

Data are presented as number (%) and mean ± SD.

- Retrospective analysis
- d-TGA after atrial switch operation
- Systemic RV dysfunction

Doughan AR, AJC 2007;99:704-6

Beta-Blockers

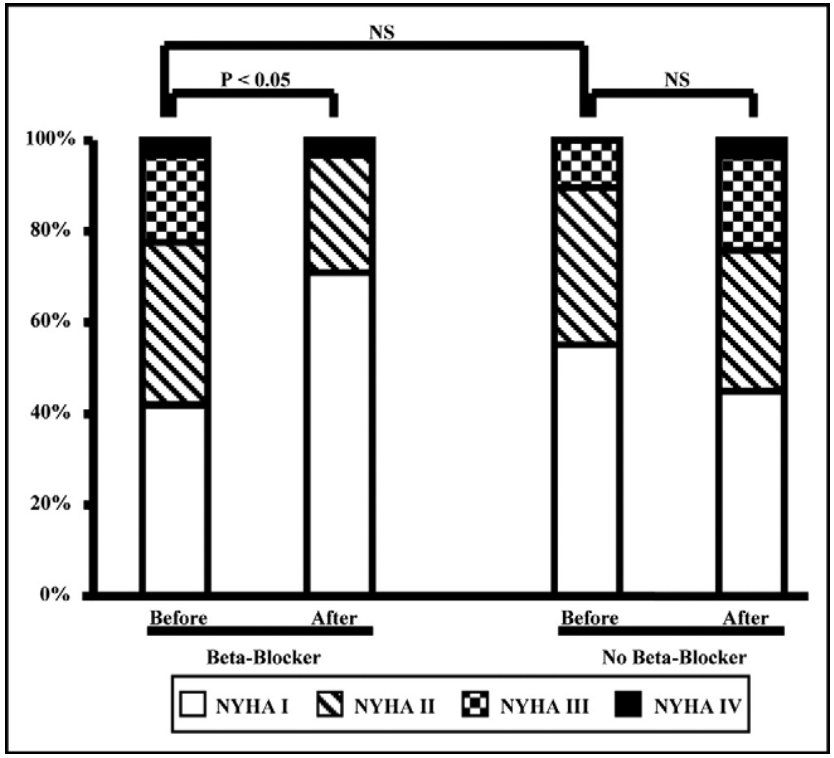


Figure 1. Comparison of NYHA class distribution in patients treated with β blockers and untreated patients at baseline and after a mean follow-up of 4 months.

Table 2
Right ventricular echocardiographic parameters

Variable	β-Blocker Therapy			
	Yes		No	
	Before	After	Before	After
RV ejection fraction (%)†	34 ± 16	36 ± 13	36 ± 8	33 ± 9
RV end-diastolic area (cm^2)†	37 ± 12	39 ± 10	40 ± 6	44 ± 5*
Degree of tricuspid regurgitation‡	2 (1–3)	1 (1–3)	1 (0–2)	1 (0–2)

* $p < 0.05$ versus before.
† Data are presented as mean ± SD.
‡ Data are presented as median (range).

Doughan AR, AJC 2007;99:704-6

Beta-Blockers

- Prospective

- Cardiovascular magnetic resonance (CMR)
- Cardiopulmonary exercise testing

Table 1
Baseline characteristics of the study cohort

	Gender	Age, years	Diagnosis	Associated lesions	Surgical procedures	Age at surgery	Tricuspid regurgitation	Baseline NYHA class	Baseline RVEF	Medications	Carvedilol final dosage
Patient 1	Male	18	D-TGA	–	Senning	8 months	Mild	I	46	ACE-inhibitor	50 mg/day
Patient 2	Male	28	CTGA	–	–	–	Mild	II	31	ACE-inhibitor	50 mg/day
Patient 3	Male	24	D-TGA	VSD	Senning, VSD closure	7 months	Moderate	II	31	ACE-inhibitor, loop diuretic	25 mg/day
Patient 4	Male	19	D-TGA	–	Senning	9 months	Mild	II	40	ACE-inhibitor	50 mg/day
Patient 5	Male	25	D-TGA	–	Senning	10 months	Mild	II	34	ACE–inhibitor	50 mg/day
Patient 6	Female	30	D-TGA	–	Senning	11 months	Moderate	II	32	ACE-inhibitor	50 mg/day
Patient 7	Female	31	CTGA	Ebstein	–	–	Severe	III	26	ACE-inhibitor, loop diuretic	12.5 mg/day
Patient 8	Female	29	D-TGA	–	Senning	14 months	Severe	III	34	ACE–inhibitor, loop diuretic	25 mg/day

ACE indicates angiotensin-converting enzyme; CTGA, congenitally corrected transposition of the great arteries; D-TGA, transposition of the great arteries; NYHA, New York Heart Association; RVEF, right ventricular ejection fraction; VSD, ventricular septal defect.

Giardini A, IJC 2007;114:241-6

Beta-Blockers

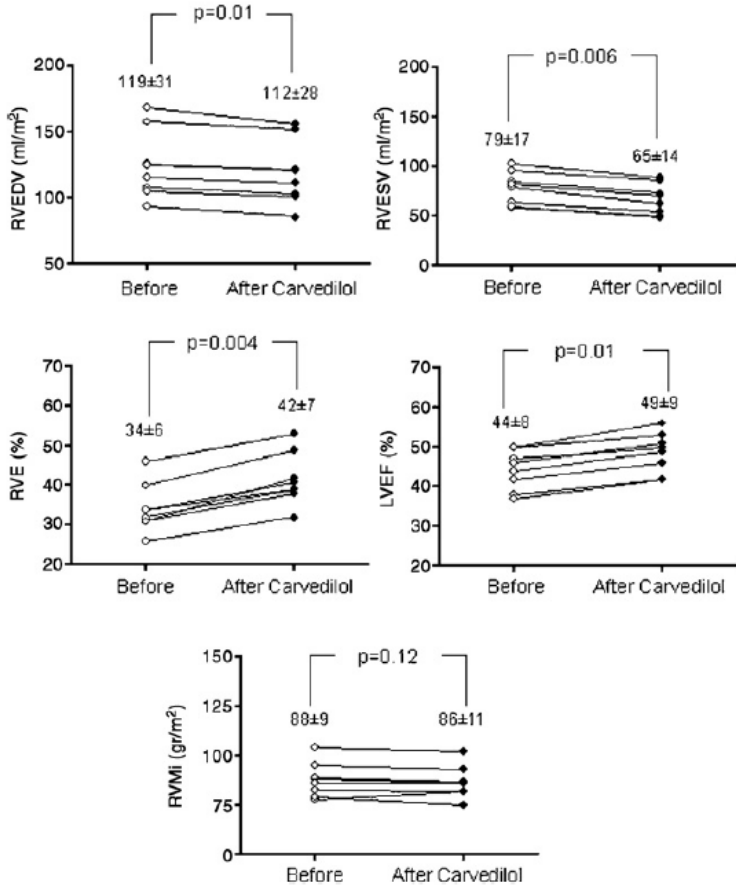


Table 2
Carvedilol-induced changes in exercise capacity

	Baseline	Carvedilol	*p*
Peak VO_2, ml O_2/Kg/min	26.8±5.3	27.3±5.7	0.58
Peak heart rate, beats/min	165±17	161±18	0.01
Peak SBP, mm Hg	163±21	158±19	0.53
Peak SO_2, %	95±3	93±4	0.58
Peak respiratory exchange ratio	1.18±0.10	1.17±0.09	0.86
Exercise duration, min	13.4±2.6	17.3±3.1	0.008
Peak workload, W	131±25	168±29	0.009

VO_2 indicates oxygen uptake.

Giardini A, IJC 2007;114:241-6

Aldosterone Antagonists

Mechanism

- Block the effects of aldosterone (spironolactone or eplerenone)
- Beneficial effects independent of the effects on sodium balance
- Recommended for patients with NYHA class IV or class III HF who have a depressed EF

Aldosterone Antagonists

Adverse Effects

- Life-threatening hyperkalemia
  - Receiving potassium supplements
  - Renal insufficiency
- Aldosterone antagonists are not recommended when the serum creatinine is >2.5 mg/dL
- Painful gynecomastia (10-15%) of patients who use spironolactone

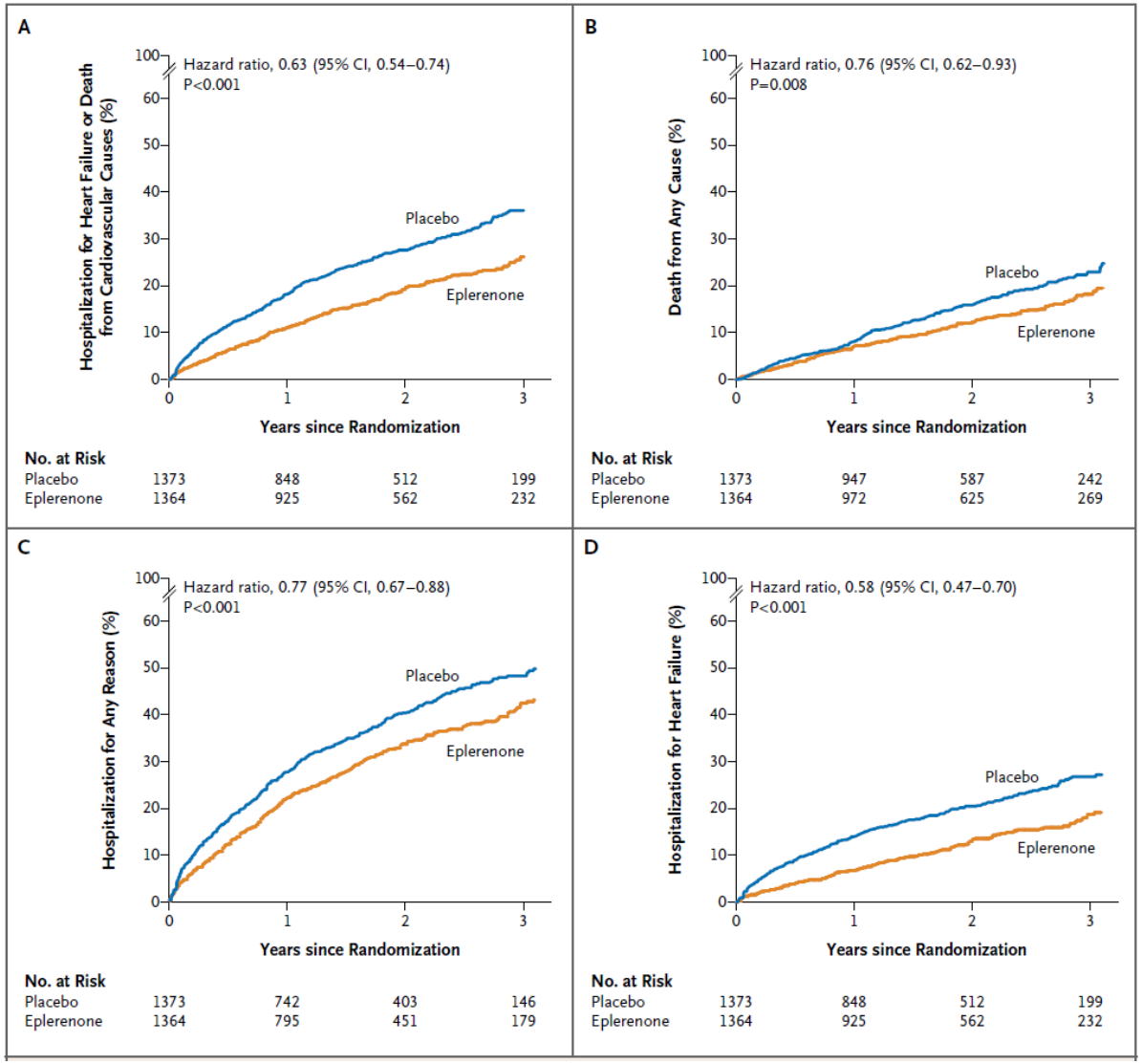
EMPHASIS-HF trial

❍Eligibility criteria

- Age ≥ 55 years
- NYHA functional **class II symptoms**
- Ejection fraction ≤ 30%
  - ➢ if >30 to 35%, a QRS duration >130 msec
- Treatment with an ACEI or ARB
- Treatment with a beta-blocker (unless contraindicated) at the recommended dose or maximal tolerated dose

N Engl J Med. 2011;364:11-21

EMPHASIS-HF trial



N Engl J Med. 2011;364:11-21

Cardiac Resynchronization Therapy (CRT)

•103 patients <21 years of age or with CHD

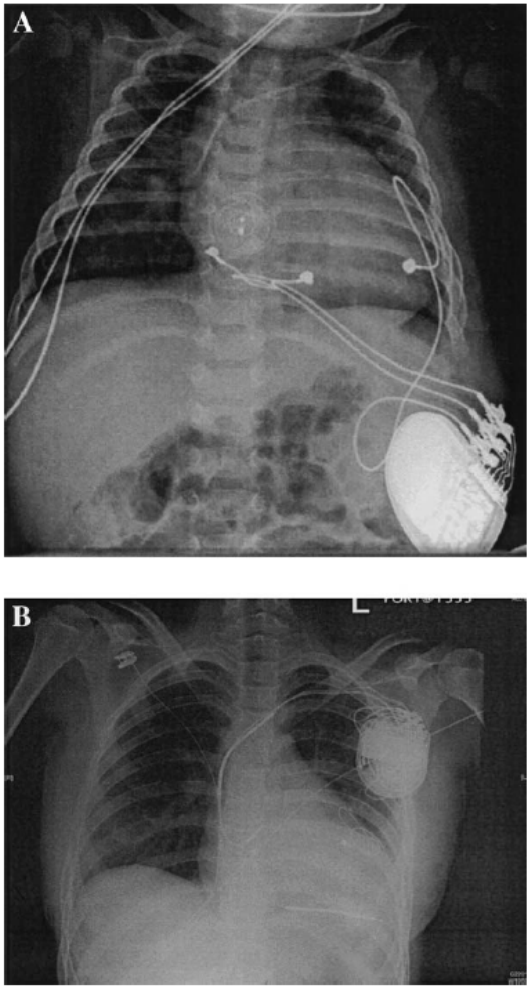


Table 3. Effect of CRT Pacing by Type of Heart Disease

Type of Disease	n	Age (yrs)	EF Improvement (EF units)	QRS Shortening (ms)
Congenital heart disease	73	12.2 (0.5–55.4)	11.9 ± 12.9%	39.1 ± 31.9
Cardiomyopathy	16	15.8 (0.3–19.6)	12.3 ± 13.6%	31.9 ± 37.9
Heart block	14	12.5 (0.3–24.3)	16.1 ± 12.9%	36.8 ± 13.0
p Value		NS	NS	NS

CRT = cardiac resynchronization therapy; EF = ejection fraction.

Table 4. Characteristics of CRT Responders Versus Non-Responders

	Responders (n = 78)	Non-Responders (n = 11)	p Value
Age (yrs)	11.9 (0.4–55.4)	14.8 (3.1–18.4)	NS
Baseline EF (%)	24.3 ± 11.0	32.0 ± 14.2	0.04
Baseline QRS (ms)	166.5 ± 33.2	172.9 ± 21.3	NS
Change in QRS (ms)	36.8 ± 24.7	33.4 ± 18.3	NS
% with CHD	71%	73%	NS
Baseline NYHA functional class 3/4	38%	31%	NS

CHD = congenital heart disease; CRT = cardiac resynchronization therapy; EF = ejection fraction; NYHA = New York Heart Association.

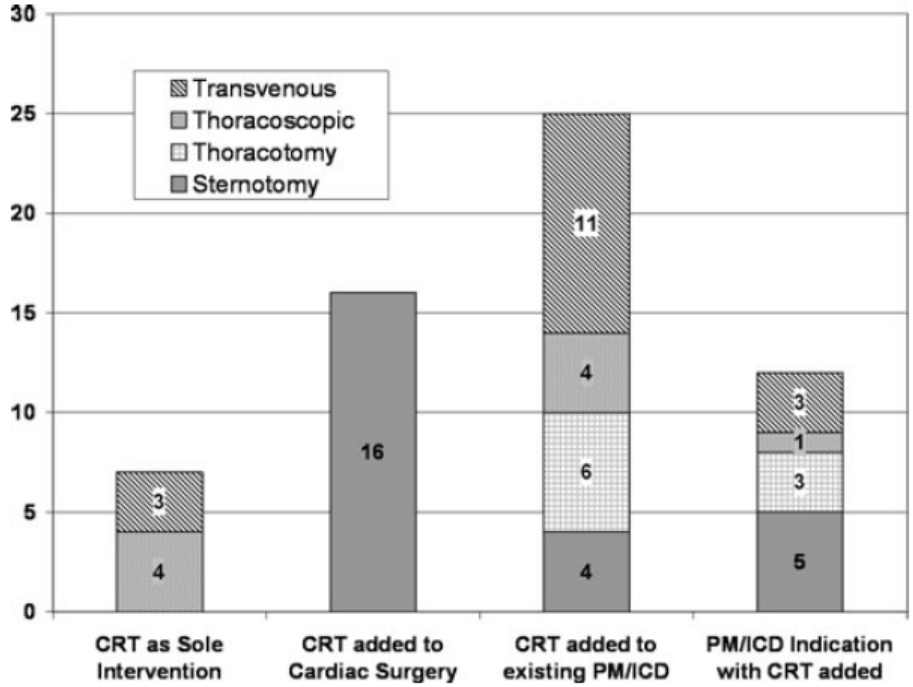
Dubin AM, JACC 2005;46:277-83

Cardiac Resynchronization Therapy (CRT)

TABLE 1

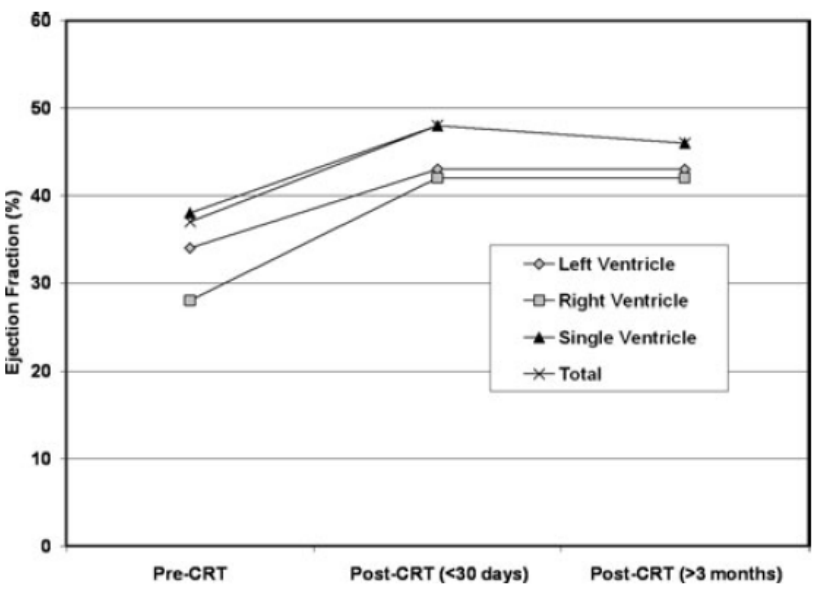
Diagnoses of Patients who Underwent Cardiac Resynchronization Therapy

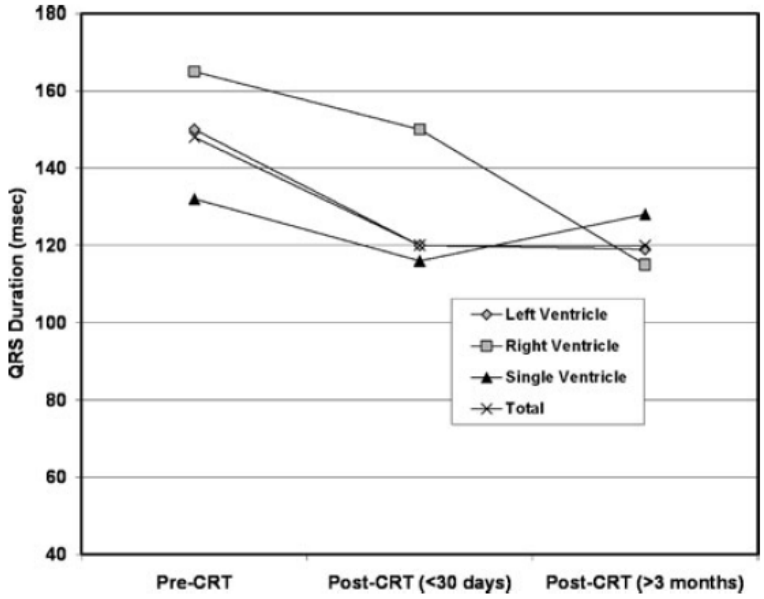
Dilated cardiomyopathy (no CHD)	**N = 14**
Idiopathic	10
Congenital heart block	4
Congenital heart defect	**N = 46**
Single ventricle (s/p bidirectional Glenn or Fontan)	13
Left ventricular outflow tract obstruction	8
L-transposition of the great arteries (systemic LV)	7
L-transposition of the great arteries (systemic RV)	4
Tetralogy of Fallot	6
D-transposition of great arteries (systemic LV)	4
D-transposition of great arteries (systemic RV)	3
Common atrioventricular canal	1



Cecchin F, J Cardiovasc Electrophysiol. 2009;20:58-65

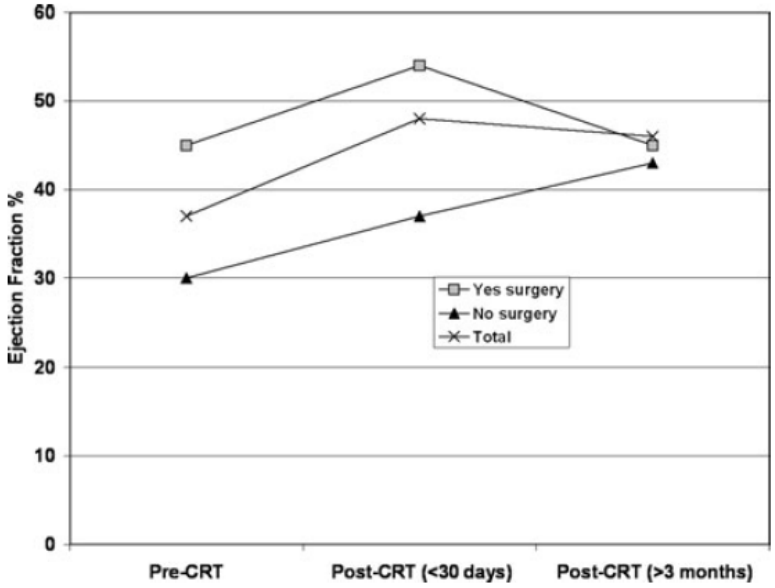
Cardiac Resynchronization Therapy (CRT)

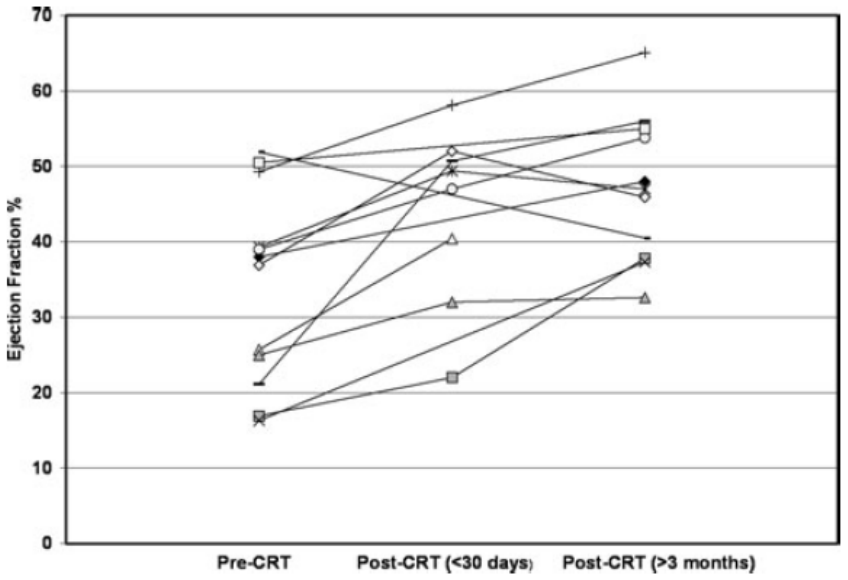




Cecchin F, J Cardiovasc Electrophysiol. 2009;20:58-65

Cardiac Resynchronization Therapy (CRT)





Cecchin F, J Cardiovasc Electrophysiol. 2009;20:58-65

Heart Transplantation

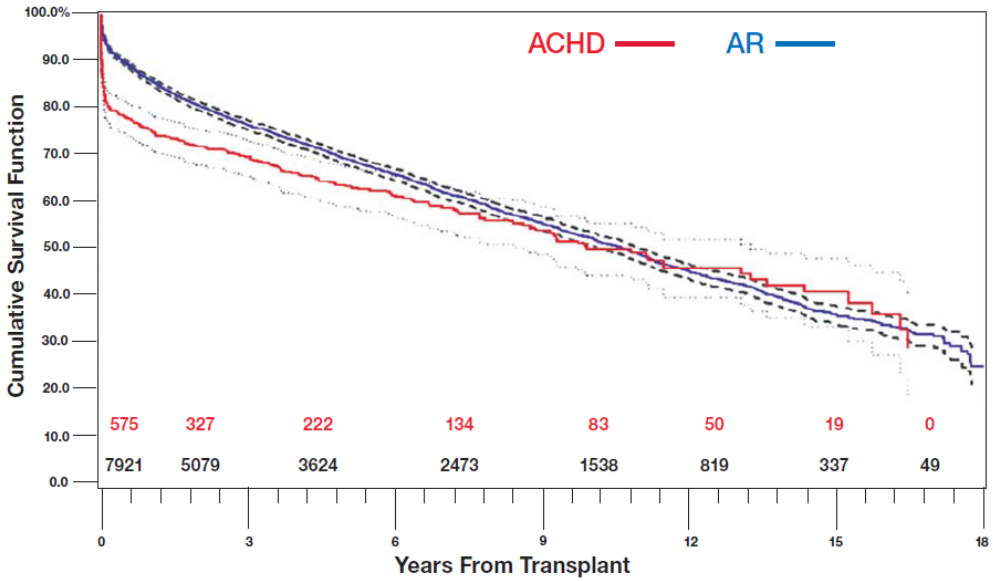


TABLE 2. Multivariable factors associated with posttransplantation mortality

Variable	Parameter estimate (± SE)	Hazard ratio	*P* value
ACHD	0.67 ± 0.15	1.96	<.001
Younger age	0.01 ± 0.002	1.01	.003
Female sex	0.08 ± 0.04	1.10	.03
Longer ischemic time	0.07 ± 0.02	1.07	<.001
No steroid maintenance	0.78 ± 0.09	2.18	<.001
No induction agent	0.19 ± 0.07	1.22	<.001
Status 1	0.09 ± 0.04	1.09	.03
Interaction term between ACHD and steroid maintenance	–	0.51	<.001

SE, Standard error; *ACHD*, adult congenital heart disease.

Karamlou T, JTCVS 2010:140:161-8

Conclusions

- The cohort of adults with congenital heart disease continues to grow, but the clinical and academic infancy of the field results in limited evidence-based applications in clinical practice.
- Extrapolation from adult studies is necessary for those caring for ACHD patients with heart failure.

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