

2005

< Pros & Cons >

***M/3, cc-TGA, PS, BCPC(+)  
Double Switch Operation***



# Congenitally corrected TGA

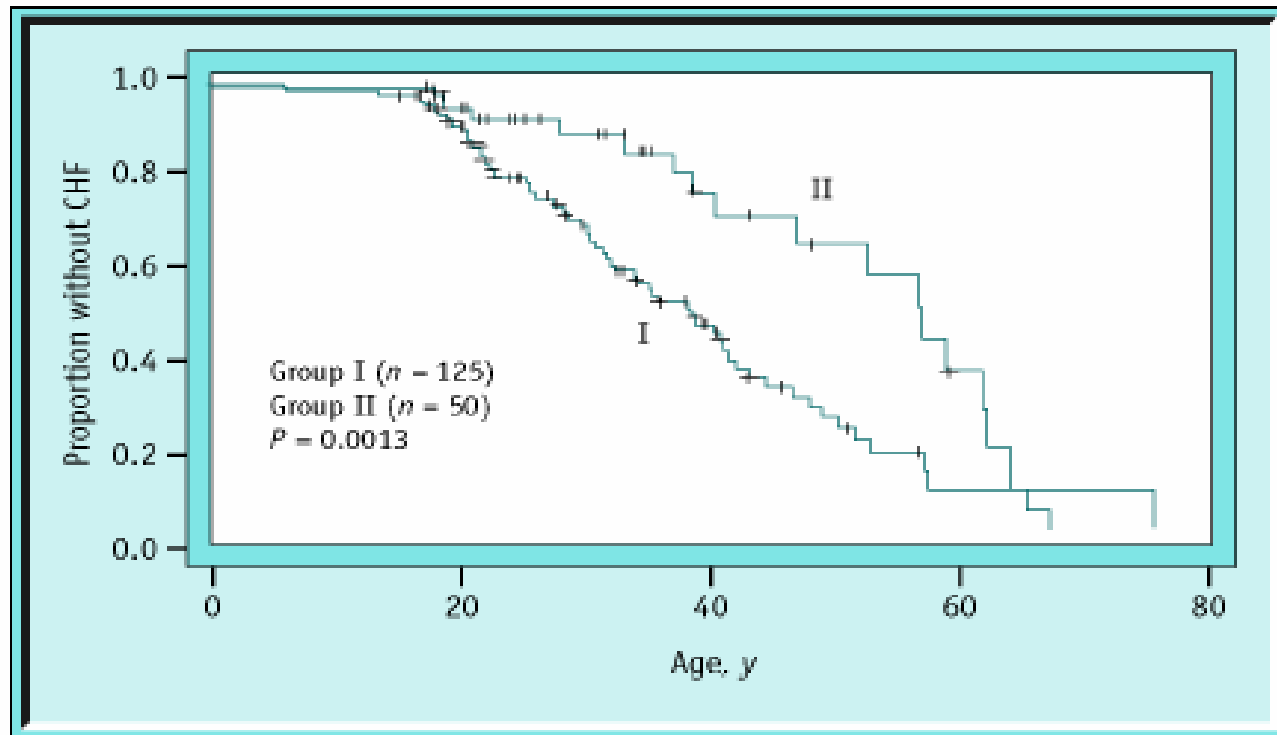
- Atrio-Ventricular & Ventriculo-Arterial discordance
- Physiologically corrected circulation with the morphologic right ventricle (RV) serving as the systemic pump.

Associated lesion	Frequency noted in major series, %
Tricuspid valve abnormalities	90–97
Ventricular septal defect	60–80
Pulmonary stenosis or atresia	55–65
Complete heart block	22–45 (increases with age)
Aortic regurgitation	25–36
Coronary artery abnormalities	Up to 45 (in autopsy series)
Dextrocardia	20
Atrial septal defect	12
Systemic right ventricular hypoplasia	Approximately 10

# Long-term outcome in cc-TGA

Graham et al. (JACC 2000)

- Freedom from Congestive Heart Failure



group I - with associated lesions

group II - no significant associated lesions

# ***Systemic RV fx***

- Hornung et al. (*Am J Cardiol* 1999)
  - Abnormal RV myocardial perfusion with fixed defects at rest and further reversible defects with exercise nearly universal among the 20 patients with cc-TGAs tested.
- Tulevski (*Cardiol Young* 2003)
  - Regional and global right ventricular dysfunction in asymptomatic or minimally symptomatic patients with cc-TGA
  - Right ventricular hypertrophy
    - Progressive ischemia
    - Diastolic dysfx

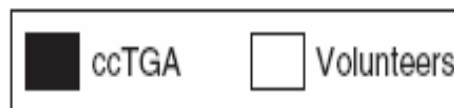
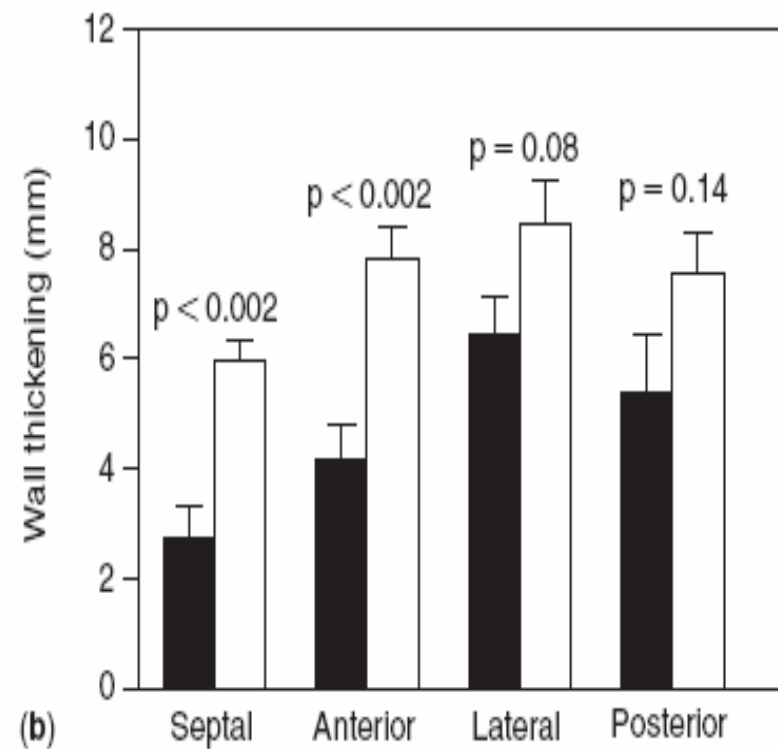
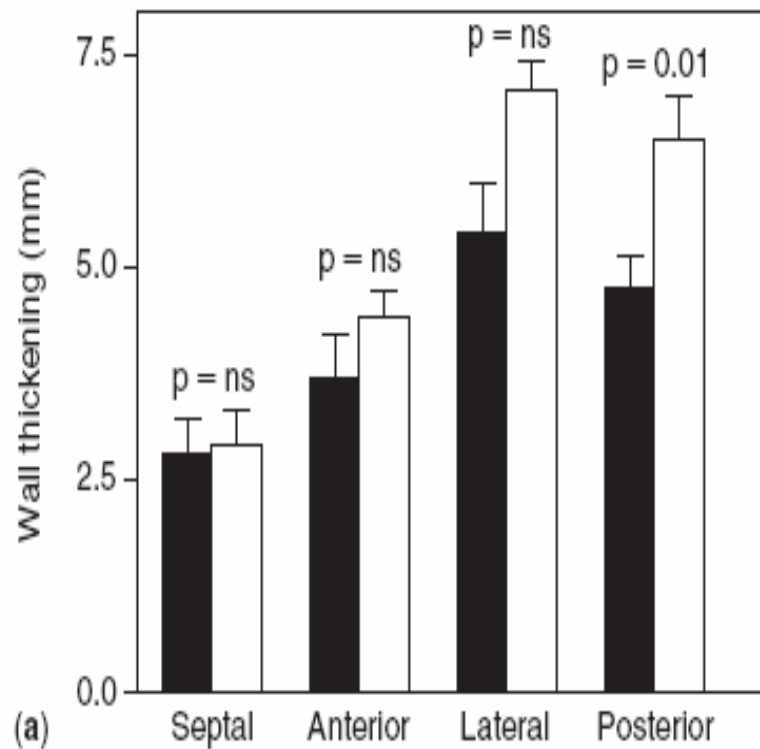
# Systemic RV fx

Table 1. Effects of dobutamine stress.

	Segment	ccTGA		p	Controls		p	ccTGA vs Controls	
		Rest	Dobutamine		Rest	Dobutamine		Rest	Dobutamine
Wall motion (mm)	Septal	5.1 (0.5)	5.0 (0.8)	ns	7.1 (0.4)	11.2 (0.6)	<0.001	<0.02	<0.001
	Anterior	7.3 (0.7)	9.5 (1.1)	ns	9.5 (0.3)	13.2 (0.6)	<0.001	<0.008	0.02
	Lateral	9.0 (0.7)	10.7 (1.2)	ns	11.6 (0.5)	13.6 (0.8)	0.04	<0.009	ns
	Posterior	8.5 (0.5)	10.2 (1.6)	ns	11.1 (0.5)	13.2 (0.8)	ns	<0.01	0.01
	Overall	7.2 (0.5)	8.8 (1.0)	ns	9.8 (0.4)	12.8 (0.4)	<0.001	<0.006	<0.006
Wall thickening (mm)	Septal	2.8 (0.4)	2.7 (0.6)	ns	2.9 (0.4)	6.0 (0.4)	<0.004	ns	<0.002
	Anterior	3.6 (0.5)	4.2 (0.6)	ns	4.4 (0.3)	7.8 (0.6)	<0.007	ns	<0.002
	Lateral	5.4 (0.6)	6.5 (0.7)	ns	7.1 (0.3)	8.4 (0.8)	ns	ns	ns
	Posterior	4.8 (0.4)	5.4 (1.1)	ns	6.5 (0.5)	7.6 (0.8)	ns	<0.02	ns
	Overall	4.1 (0.4)	4.7 (0.6)	ns	5.2 (0.4)	7.5 (0.4)	<0.02	ns	<0.005
Ejection fraction (%)		44 (3)	48 (5)	ns	64 (5)	72 (4)	<0.05	<0.001	<0.001

Abbreviations: ccTGA: congenitally corrected transposition of the great arteries; ns: not significant. Values between brackets denote standard error of the mean

# Systemic RV fx



## ***Theoretical Risk Factors for RV failure in cc-TGA***

- Volume overload
- Systemic afterload
- Conduction and rhythm disturbances
- Associated LV pressure and geometry
- Coronary artery anomalies
- Surgical injury
- Hypoxemia
- Advancing patient age (bacterial endocarditis, pregnancy)

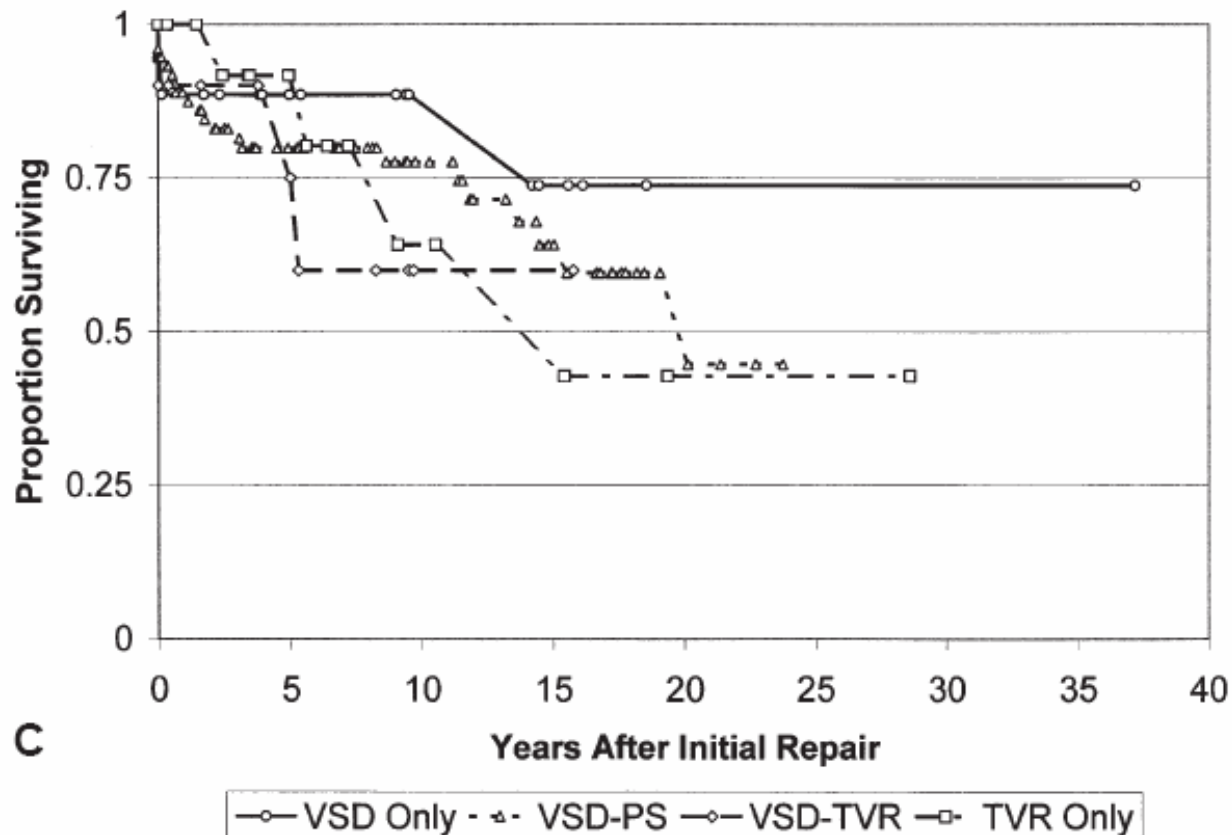
# Conventional repair

				Early mortality	10 yr survival	20 yr survival	Free of reop
Yeh (Toronto)	1999 JTCS	118	1959~1997	6%	75%	48%	65% at 10 yrs
Biliciler-Denktaş (Mayo)	2001 JTCS	111	1971~1996	16%	67%	57% (at 15yr)	59% at 10 yrs
Rutledge (Texas)	2002 Pediatr Cardiol	121 (47)	1952~1999		91%	75%	
Hraska (Boston)	2005 JTCS	123 (96)	1963~1996	15% at 1yr	68%	59%	



# Conventional repair

- Yeh et al. (*J Thorac Cardiovasc Surg* 1999)



## Comparative Anatomy of the Ventricles

	RV	LV
Trabeculation	Coarse	Fine
Wall thickness		Wall thickness
AV valve	Round, 30-40% larger than mitral valve and encompassed by an elastic annulus Leaflets are attached to multiple papillary muscles of differing size including the IVS	Oval shape, reinforced with firm annulus Leaflets are anchored by two distinct papillary muscles
Coronary circulation	During systole and diastole	During diastole
Ventricular architecture	Suited for the changes in preload	Suited for the changes in afterload

# ***Anatomic repair***

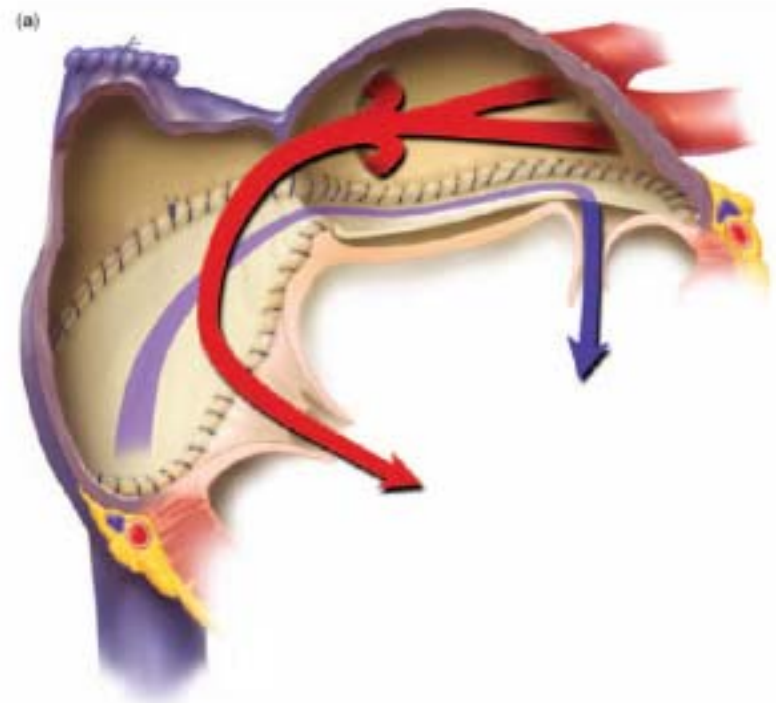
- LV as the systemic ventricle
  - the theoretic advantage of removing the high pressure load from the RV and ameliorating progressive RV and TV dysfunction
- LV 50% of the RV size, LVP > 70% of systemic pressure
- Atrial switch (Senning/Mustard)
  - Position of cardiac apex
- Arterial switch/Rastelli type
  - Normal pulmonary valve, pulmonary outflow tract obstruction (<mild), coronary artery anomalies
  - Good branch PAs with low PVR, Routability of VSD

# ***Anatomic repair***

- Predictable disadvantages
  - Venatrial baffle obstruction
  - Atrial arrhythmia
    - Atrial overload is usually avoided in anatomic correction of cc-TGA (cf. d-TGA)
  - Sinus node dysfunction
  - Long myocardial ischemic time
  - After Rastelli
    - Insufficient space from anatomic RV to PA
      - Conduit compression
    - Reoperation for replacement

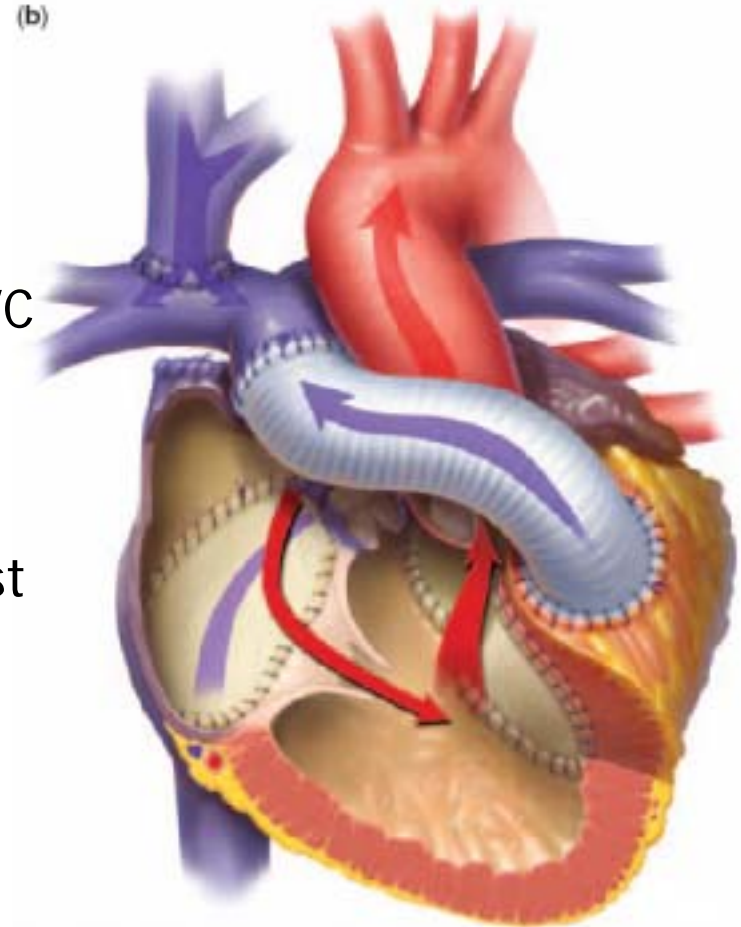
# ***Hemi-Mustard procedure***

- Reduces complexity of the intraatrial procedure
- Reduces myocardial ischemia time
- Recognized complications, such as SVC obstruction, pulmonary venous obstruction, sinus node dysfx are eliminated.
- mRV-pulmonary trunk conduit will last longer
- If the mRV size is reduced, BCPS may provide superior hemodynamics.



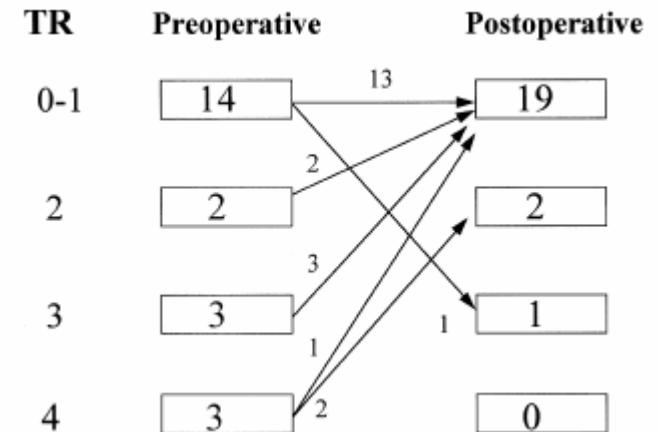
# Hemi-Mustard procedure

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# Outcomes after Double Switch

- Imamura *et al.* (*Ann Thorac Surg* 2000)
  - 27 patients operated at the Cleveland Clinic
  - Age at operation : 3 Mo ~ 55 yrs
  - 22 Double switch operation
    - 10 Senning + ASO
      - mean CPB time 210min, ACC time 107min
    - 12 Senning + Rastelli
      - mean CPB time 194min, ACC time 101min
  - 6 Previous banding
  - no early or late mortality
  - Only 2 patients required pacemakers
  - TV function improved in all patients except one.
  - No systemic or pulmonary venous obstruction or atrial arrhythmia
  - follow-up : mean 27.8 (1 ~ 58) Mo



# ***Outcomes after Double Switch***

- Ilbawi et al. (Ann Thorac Surg 2002)
  - 12 pts
  - mean age at op. :  $9 \pm 3.6$  Mo
  - Mustard + Rastelli(10) or ASO (2)
  - Concomitant TV valvuloplasty(2) and VSD enlargement (1)
  - 1 hospital death
  - 1 Pacemaker insertion
  - 1 SVC obstruction
  - All survivors had normal LV fx with good hemodynamic parameters and all remained asymptomatic
  - mean follow-up of 7.6 (0.5 to 10) years



# ***Outcomes after Double Switch***

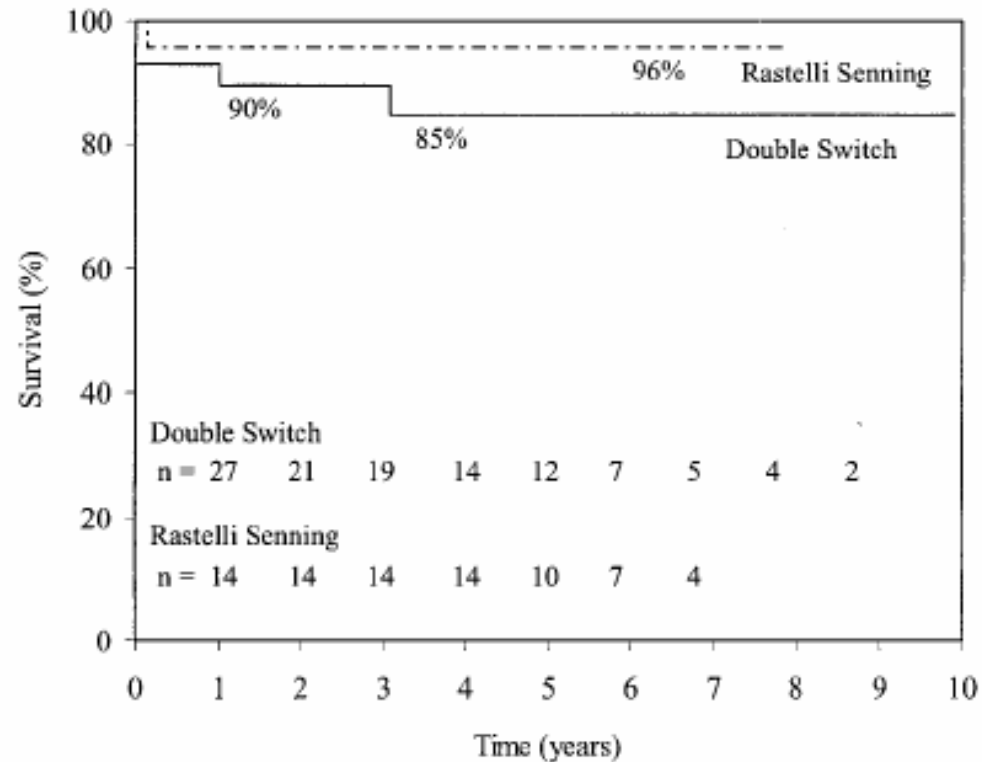
- Imai et al. (*Semin Thorac Cardiovasc Surg* 2001)
  - 76 pts
  - Mean age at op. :  $6.6 \pm 3.6$  yrs (3 Mo ~ 15 yrs)
  - 47 Mustard and 29 Senning
  - 14 ASO, 40 Rastelli, 21 REV
  - 14 previous banding
  - hospital mortality : 7.9%
  - no hospital deaths in the last 22 patients
  - Mean follow-up : 4.9 yrs
  - 4 late deaths
  - 2 reoperation

# ***Outcomes after Double Switch***

- **Langley et al.** (*J Thorac Cardiovasc Surg* 2003)
  - 54 pts
  - Median age at op. 3.2 yrs (7 wks ~ 40 yrs)
  - Senning + ASO (29) or Rastelli (22)
  - 9(31%) Previous banding
  - hospital mortality : 5.6%
  - median follow-up : 4.4 yrs
  - 2 late deaths
  - 94% of survivors in NYHA Fc I

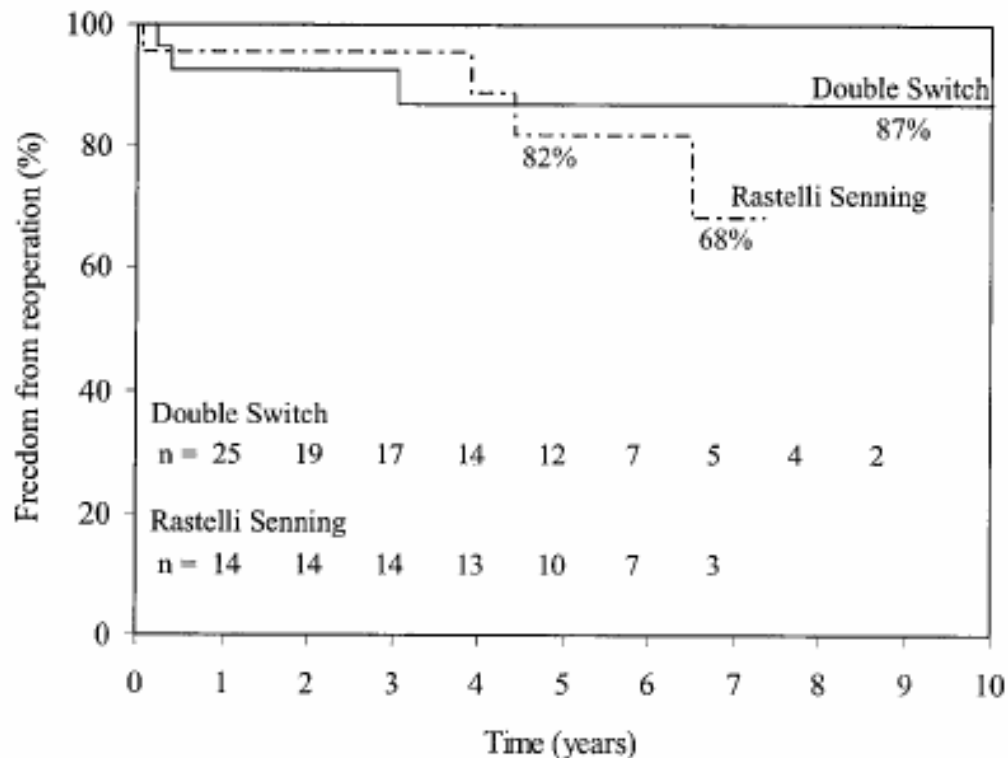
# Outcomes after Double Switch

- Langley et al. (*J Thorac Cardiovasc Surg* 2003)
  - survival



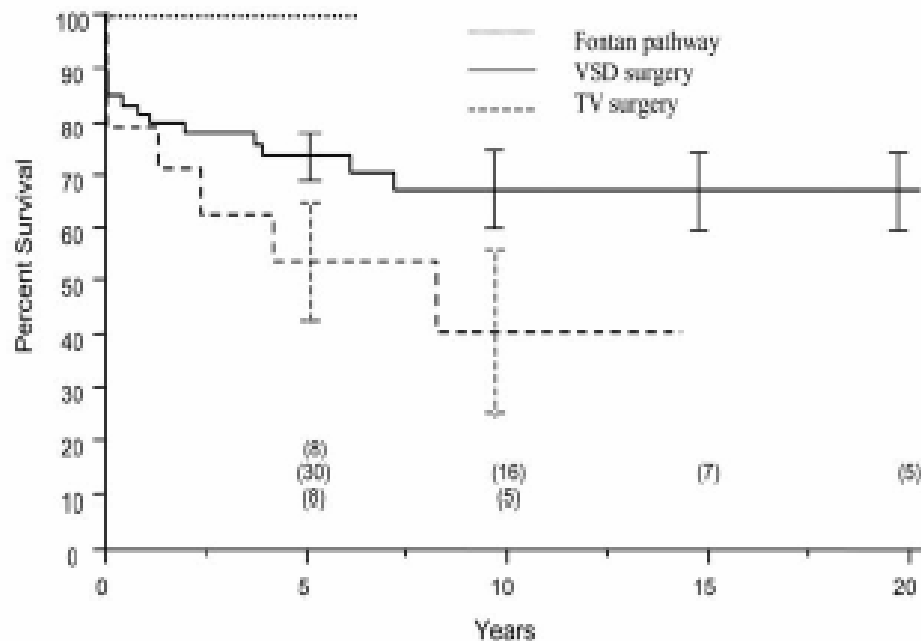
# Outcomes after Double Switch

- Langley et al. (*J Thorac Cardiovasc Surg* 2003)
  - Freedom from reop.



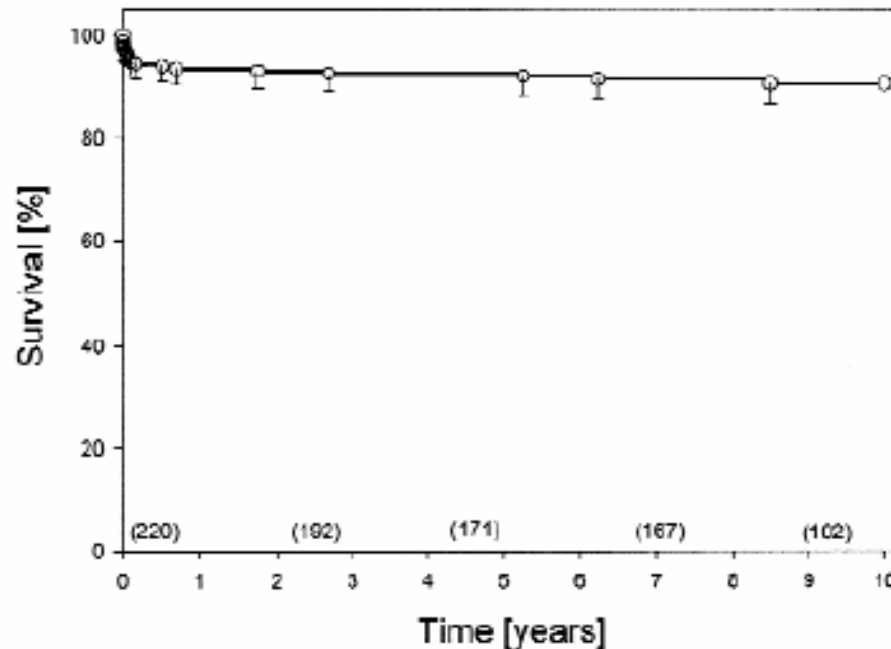
# Outcomes after Fontan op.

- Hraska et al. (*J Thorac Cardiovasc Surg* 2005)
  - Long-term outcome of surgically treated patients with cc-TGA
  - 17 Fontan patients



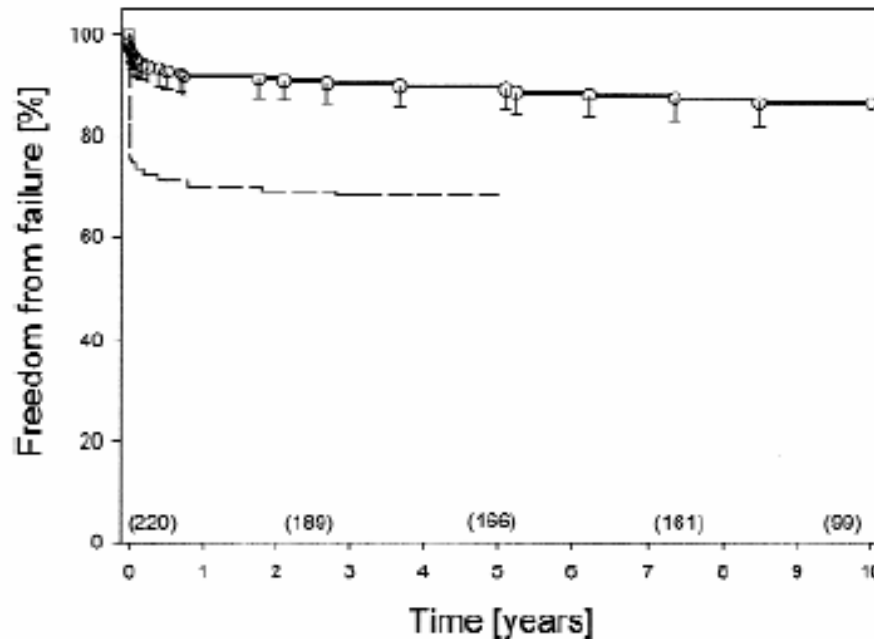
# Long-term Results; Lateral Tunnel Fontan

- Stamm et al. (*J Thorac Cardiovasc Surg* 2001)
  - 220 Fontan patients, Mean FU  $10.2 \pm 0.6$  yrs
  - Survival : 93% at 5yr, 91% at 10 yr



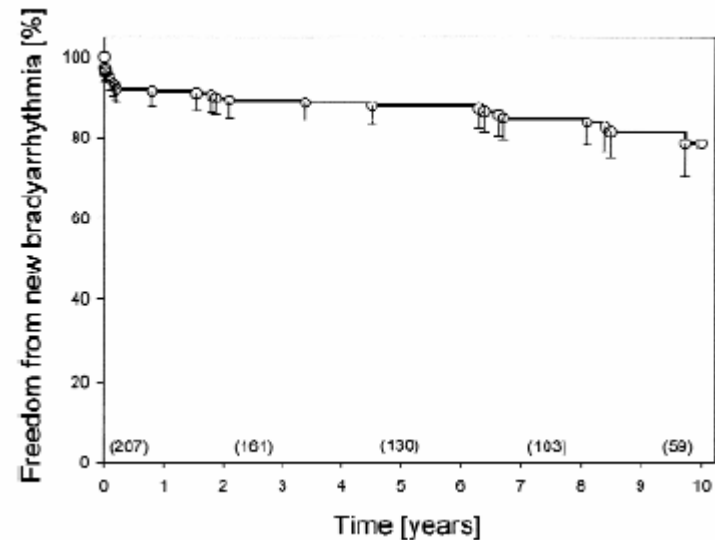
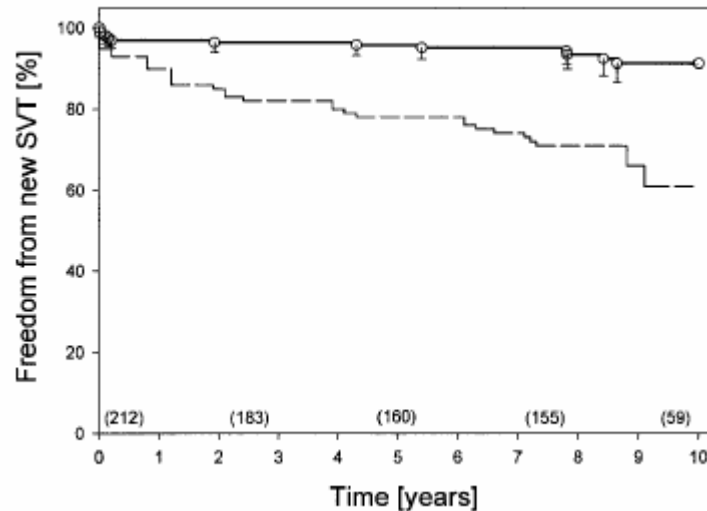
# Long-term Results; Lateral Tunnel Fontan

- Stamm et al. (*J Thorac Cardiovasc Surg* 2001)
  - 220 Fontan patients, Mean FU  $10.2 \pm 0.6$  yrs
  - Freedom from Fontan failure : 90% at 5yr, 87% at 10yr



# Long-term Results; Lateral Tunnel Fontan

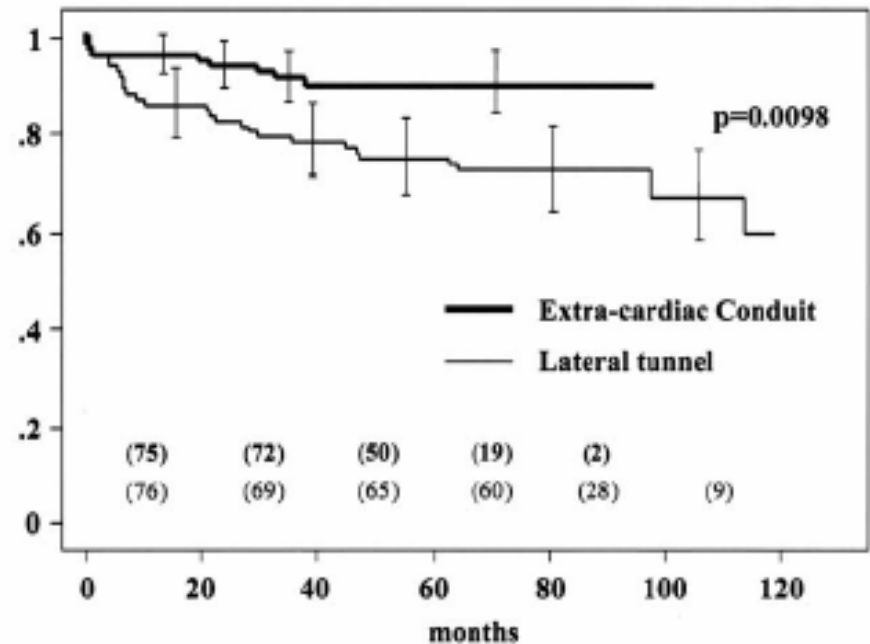
- Stamm et al. (*J Thorac Cardiovasc Surg* 2001)
  - 220 Fontan patients, Mean FU  $10.2 \pm 0.6$  yrs
  - Freedom from new bradyarrhythmia : 88% at 5yr, 79% at 10yr





# Midterm results of Extracardiac conduit Fontan

- Nakano et al. (*J Thorac Cardiovasc Surg* 2004)
  - 88 lateral tunnel vs. 79 extracardiac conduit
  - 8 yr survival 93.2% vs. 94.9%
  - SVT 15.9% vs. 5.1%(5~11%)



Freedom from cardiac related event

# Thromboembolic Cx after Fontan

- Monagle and Karl (*Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2002)
  - Review of 8 studies, total 1585 patients
  - thromboses : 3 ~ 16%
  - stroke or arterial emboli : 3 ~ 19%
- Coon et al. (*Ann Thorac Surg* 2001)
  - echocardiographic findings in 592 Fontan patients
  - Intracardiac thrombus : 52 pts (8.8%)
  - Freedom from thrombus : 92% at 1yr, 82% at 10yr
  - The appearance of thrombus was not influenced by the type of operation or the presence of fenestrations.
  - stroke : 8/52 pts(15%).
  - frequency of thrombus occurrence closely resembled previously published curves for the development of arrhythmia and protein-losing enteropathy

# Thromboembolic Cx after Fontan

- Seipelt et al. (*Ann Thorac Surg.* 2002)
  - 85 pts
  - Thromboembolic event : 13 pts (15.3%)
  - 1<sup>st</sup> peak occurred within the first postoperative year, 2<sup>nd</sup> peak beyond 10 years of follow-up.
  - The type of operation had no influence on the rate of thromboembolism.
  - missed by transthoracic echocardiography alone
- Chun et al. (*Am J Cardiol.* 2004)
  - 139 patients
  - Stroke : 7 pts (3.6%)
  - Events occurred between 2 weeks and 9 years postop.
  - 2 strokes occurred in patients receiving aspirin and warfarin, 2 receiving aspirin alone, and 3 with no anticoagulation

# Aerobic capacity in CHD

(Fredriksen et al. AJCC 2001)

- Exercise test and static lung fx

**TABLE 4** Mean  $\pm$  SD Exercise Test and Static Lung Function Data for the Entire Sample, Subdivided into Diagnostic Groups

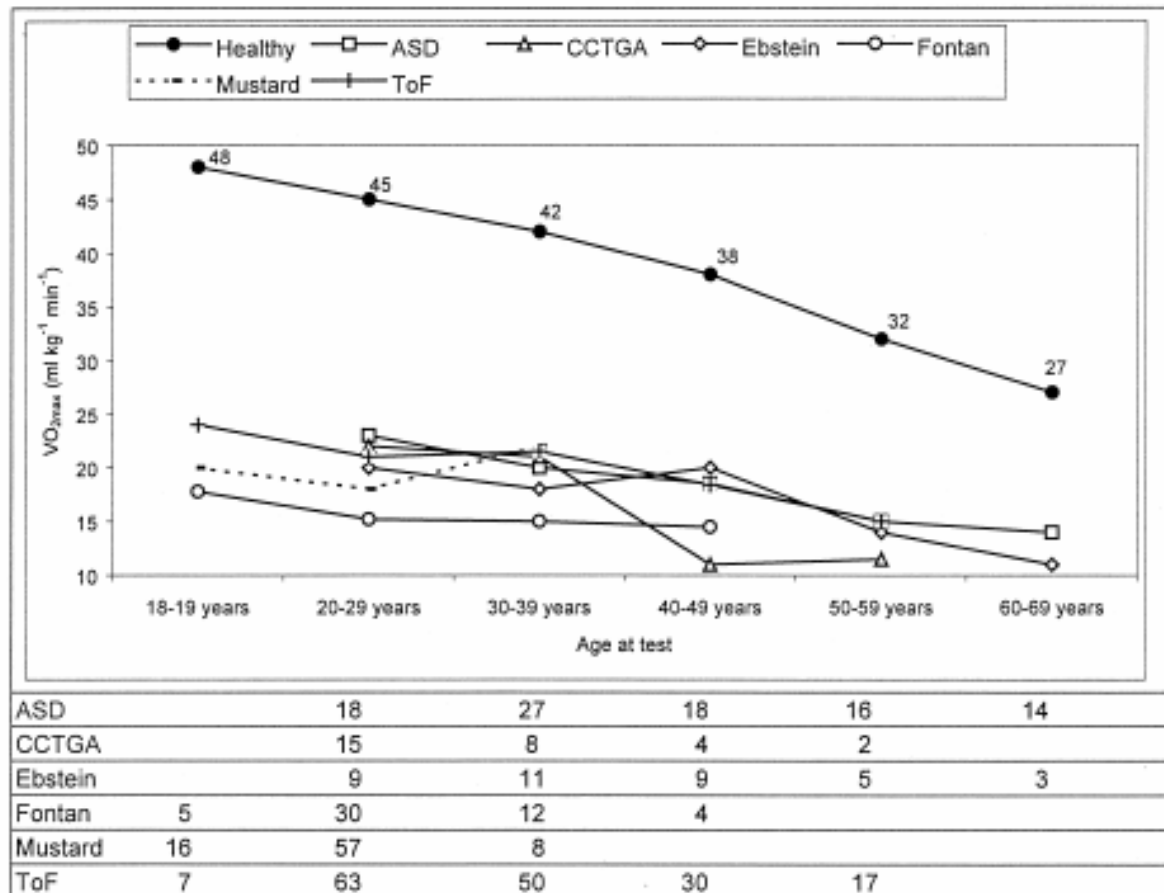
	Max. Oxygen Uptake (ml · kg <sup>-1</sup> min <sup>-1</sup> )	Max. Heart Rate (beats/min)	Max. Systolic Blood Pressure (mm Hg)	Max. Diastolic Blood Pressure (mm Hg)	Oxygen Saturation (%)	% FVC Predicted
All	19.8 $\pm$ 6.9	146 $\pm$ 28	161 $\pm$ 29	80 $\pm$ 20	94 $\pm$ 5	85 $\pm$ 17
ASD	19.8 $\pm$ 7.3	146 $\pm$ 30	—	—	—	97 $\pm$ 20
CCTGA	19.7 $\pm$ 7.4	142 $\pm$ 28	152 $\pm$ 19	81 $\pm$ 13	92 $\pm$ 8	84 $\pm$ 15
Ebstein	18.6 $\pm$ 7.1	144 $\pm$ 30	—	—	92 $\pm$ 4	77 $\pm$ 3
Fontan	15.9 $\pm$ 4.0	139 $\pm$ 25	150 $\pm$ 30	77 $\pm$ 18	87 $\pm$ 5	74 $\pm$ 17
Mustard	20.0 $\pm$ 6.3	147 $\pm$ 25	170 $\pm$ 29	86 $\pm$ 23	89 $\pm$ 5	81 $\pm$ 13
ToF	21.1 $\pm$ 7.2	149 $\pm$ 27	—	—	94 $\pm$ 3	84 $\pm$ 17

\*Men and women are analyzed together. The number of patients is listed in Table 3.  
Max. = maximal.

# Aerobic capacity in CHD

(Fredriksen et al. AJCC 2001)

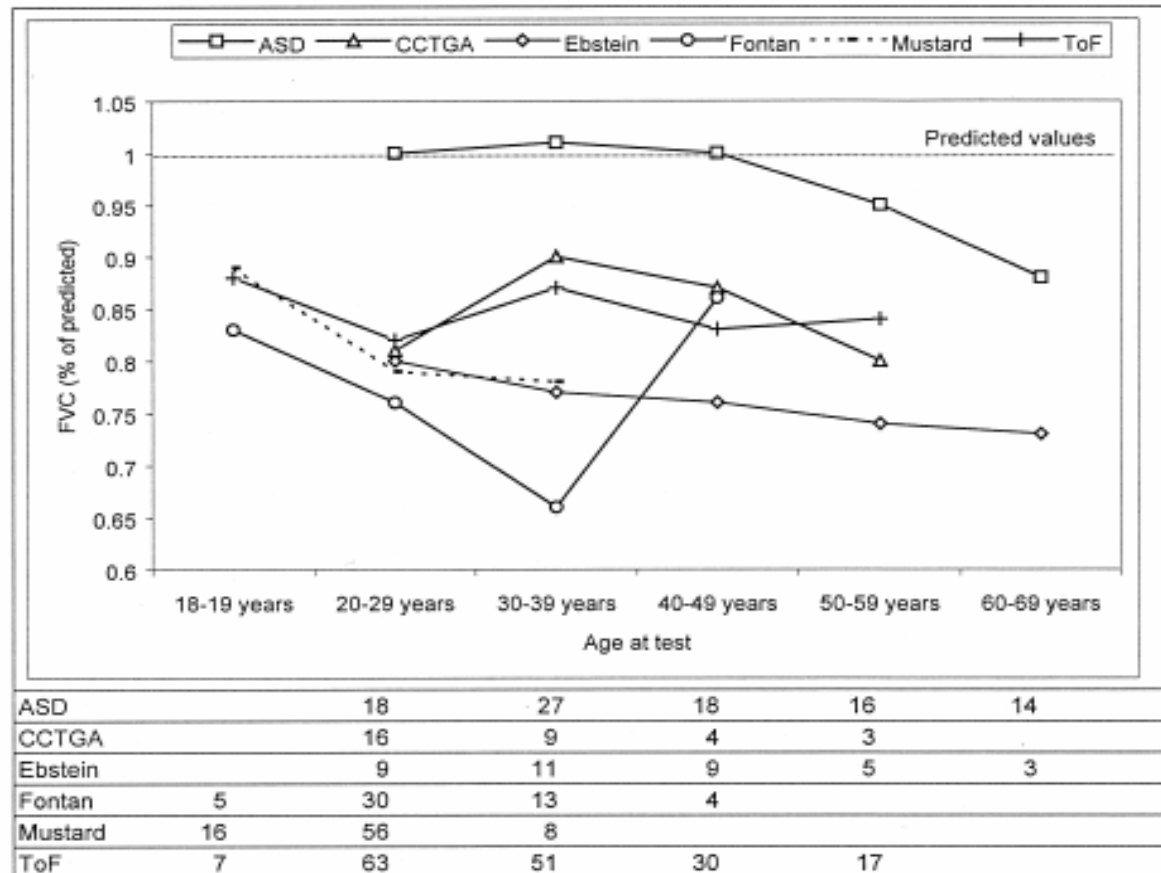
- Maximal O<sub>2</sub> uptake



# Aerobic capacity in CHD

(Fredriksen et al. AJCC 2001)

- Static lung function - FVC



# Pathologic anatomy of cc-TGA

(Van Praagh et al. Am Heart J 1998)

- 33 autopsy case with two ventricles
- With IVS(22%), Ebstein's anomaly was always present, Pulmonary outflow tract obstruction never occurred, aortic outflow tract obstruction was frequent(43% 3/7)
- With VSD(78%), Ebstein's anomaly less common(44%), straddling or overriding of TV(24%), pulmonary outflow tract obstruction (56%), aortic outflow tract obstruction(4%)
- Malformation of the RV sinus and of the TV : 30/32(94%) cases of cc-TGA with situs solitus
  - Ebstein;s anomaly, with dysplasia of the TV and of the RV sinus 10/32(59%)
  - Hypoplasia of the RV sinus 11/32(34%)
  - Straddling 5cases, TS 5 cases, TV overriding 1 1case
- Sparing of the LV was always observed in 100% of the classical cc-TGA {S,L,L/D}
- Systemic Rt coronary artery insufficiency with angiographically normal coronary arteries

# ***Pathologic anatomy of cc-TGA***

*(Van Praagh et al. Am Heart J 1998)*

- Patients with classical corrected TGA {S,L,L} and two ventricles almost never have two normal ventricles
- The morphologically right ventricle is a one coronary ventricle;
- The normal tricuspid valve is not designed to close a circular systemic atrioventricular orifice
- The papillary muscles of the morphological left ventricle are a well-balanced pair;
- The radiations of the conduction system favor the morphologically left ventricle.



# ***Pathologic anatomy of cc-TGA***

*(Van Praagh et al. Am Heart J 1998)*

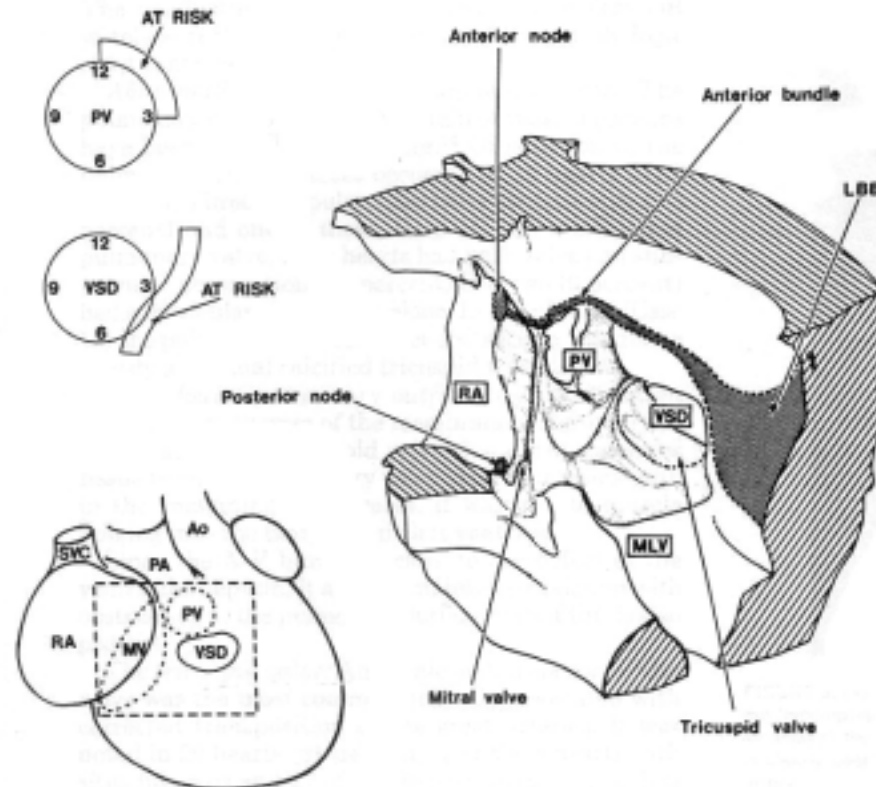
- Many anatomic features suggest that the LV-MV system will be a better long-term pumping system than the RV-TV. Thus the RV-TV system in corrected
- cc-TGA “needs” an arterial switch operation even more than does the RV-TV system in physiologically uncorrected D-TGA50 because the RV sinus and the TV in corrected TGA almost always are malformed

## ***Conclusion***

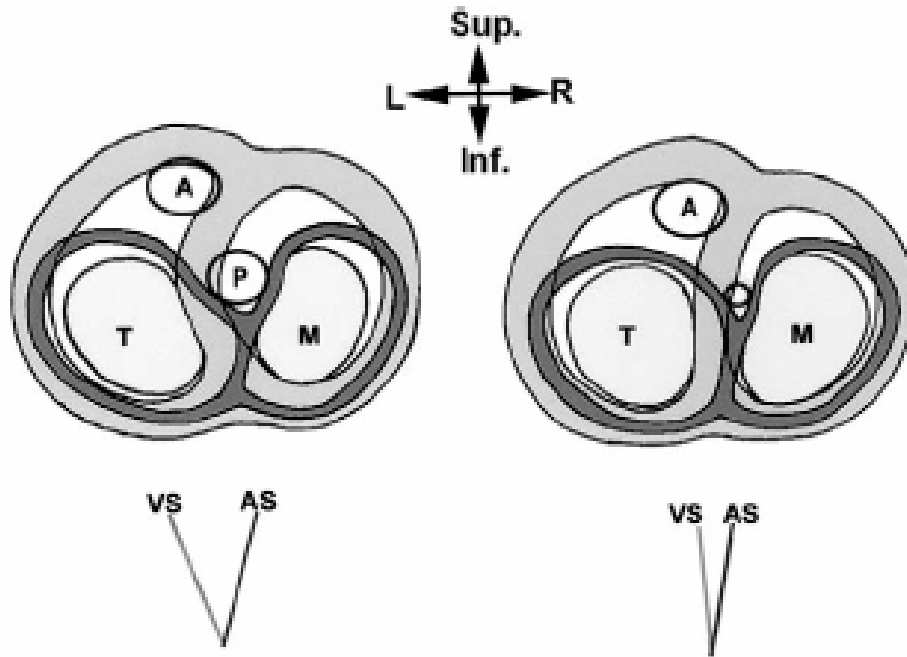
- Conventional repair, which leaves the morphologic RV as the systemic ventricle, has resulted in high incidence of tricuspid regurgitation (TR) and progressive dysfunction of the RV.
- The advent of the Double Switch operation, which restores the morphologic left ventricle (LV) as the systemic ventricle, has yielded favorable outcomes at early follow-up.
- Although long-term follow-up is needed to evaluate the potential superiority of this procedure, several theoretical advantages are expected.



# Conduction system in cc-TGA

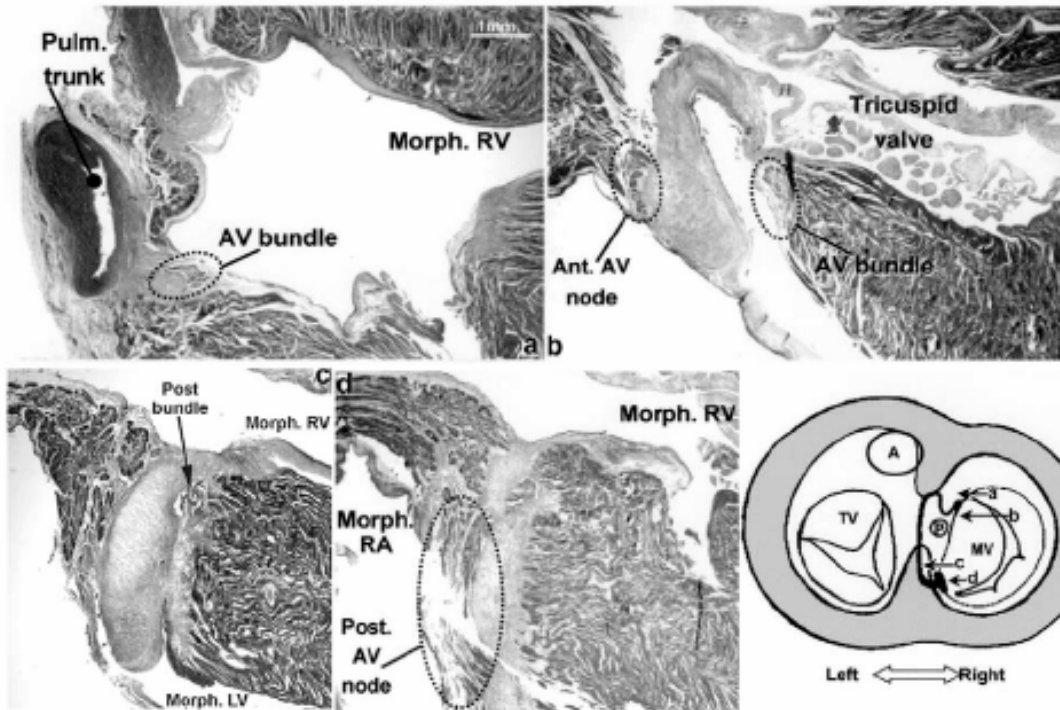


# Conduction system in cc-TGA



- Hosseinpour et al. Ann Thorac Surg 2004
- Pulmonary trunk/Ao ratio < 0.6
- Minimal septal malalignment 0-5 degrees

# Conduction system in cc-TGA



- Hosseinpour et al. Ann Thorac Surg 2004
- posterior node in addition to anterior bundle -> slinglike arrangement of the AV conduction system