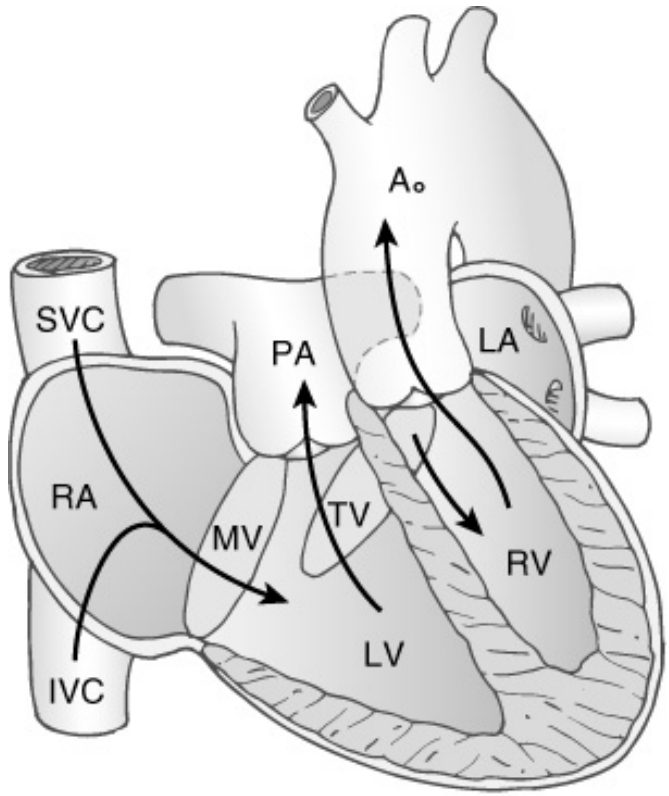
Surgical Options for Congenitally corrected TGA with VSD and PS

Han Ki Park

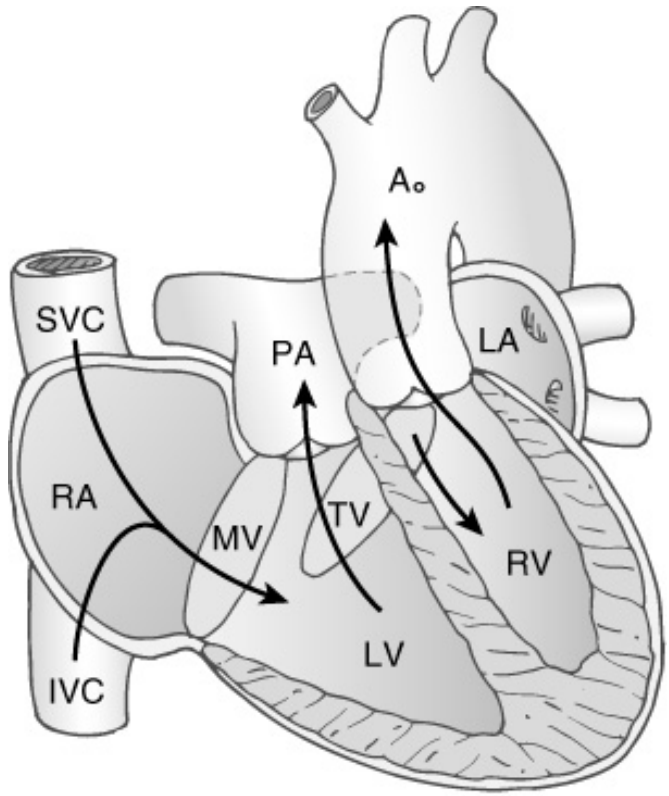
Yonsei University College of Medicine

Congenitally corrected TGA is a complex congenital heart defect characterized by the presence of both atrioventricular & ventricular discordance.



Commonly associated with

- Tricuspid valve regurgitation
- VSD
- Pulmonary stenosis or atresia
- Complete heart block



Case

- 3 years old boy TGA, VSD, small ASD, subpulmonic PS
- BCPC was done at 6 months old to relieve cyanosis
- Currently well with mild cyanosis (SaO_2 of 82%)
- Echo:

  good RV and LV function
  mild/moderate TR;
  mild MR;
  mild Ebstein malformation of TV;
  severe subpulmonic PS without pulmonary regurgitation;
  VSD of not so large in size and with possible dynamic obstruction;
  small ASD;
  mild aortic regurgitation with mild aortic root pathology;
  good MPA and both pulmonary arteries

Surgical Options

I. Classic Repair (Physiologic Repair)

- Classic Repair (Physiologic Repair)
  - VSD closure
  - LV-PA conduit or direct connection

Hraska et al.

Long-term outcome of surgically treated patients with corrected transposition of the great arteries

J Thorac Cardiovasc Surg 2005;1296:182

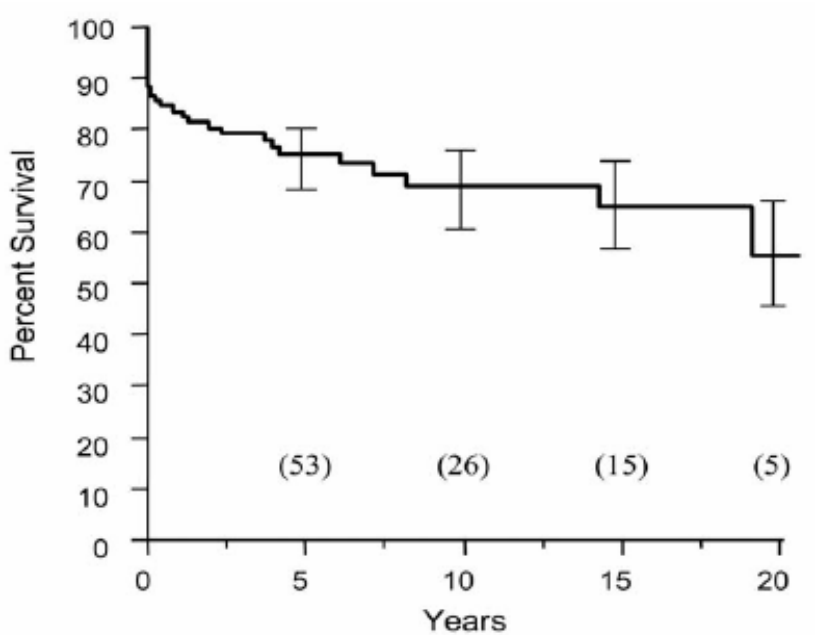
1963 ~ 1996, 123 patient

Palliative surgery and/or Intracardiac Procedure

Surgical Options

I. Classic Repair (Physiologic Repair)

The probability of survival



1 yr	84%
5 yr	75 %
10 yr	68 %
15 yr	61 %

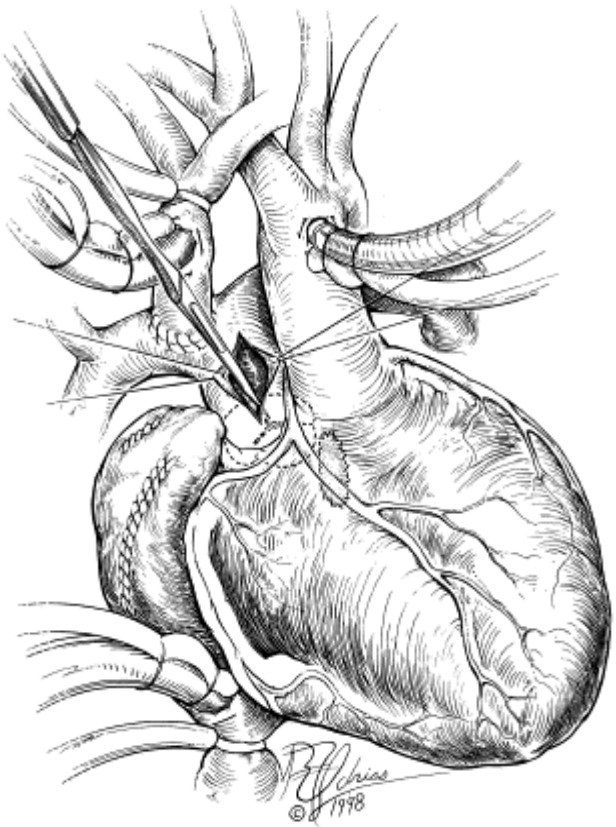
Hraska et al. J Thorac Cardiovasc Surg 2005;1296:182

Surgical Options

I. Classic Repair (Physiologic Repair)

One-and-a-half ventricle repair

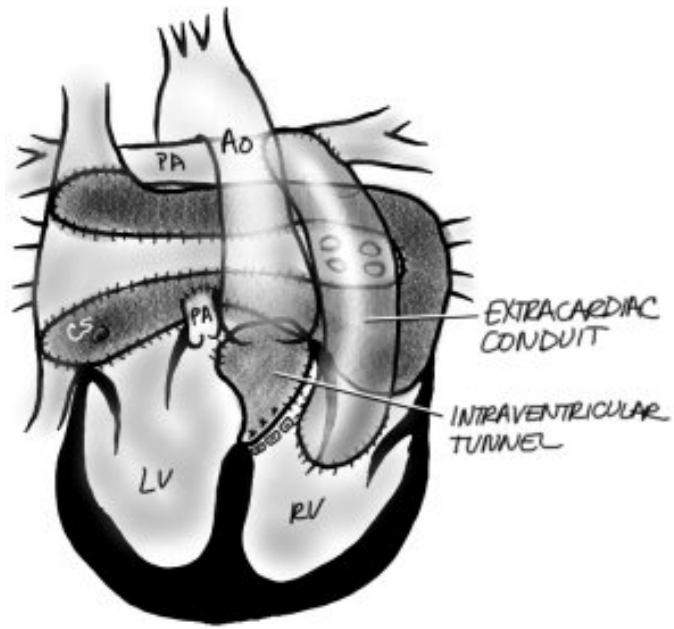
- VSD closure
- Pulmonic valvotomy
- Conal septum resection
- BCPS



Mavroudis et al., Ann Thorac Surg 1999 68:976

Surgical Options

II. Anatomic Repair (Double Switch Operation)

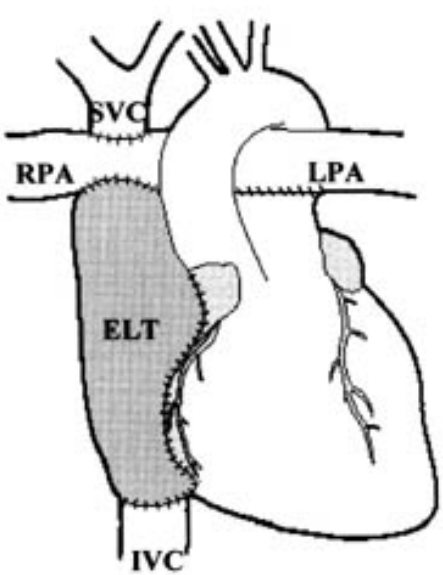


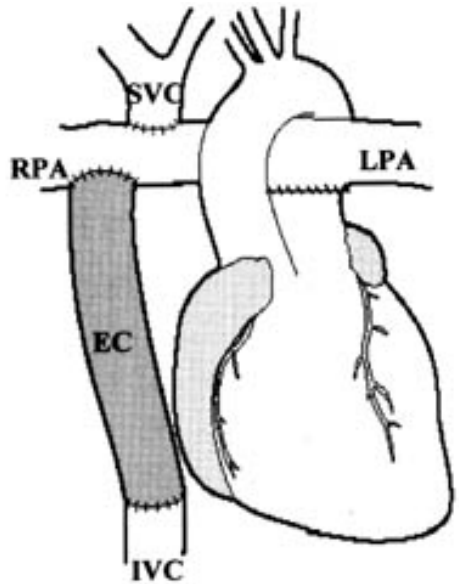
Senning/Mustard procedure

Arterial Switch or Rastelli procedure

Surgical Options

III. Fontan-type Operation





Extension of ASD

Division of MPA

Factors to be considered

- Surgical complexity
- Operative risk
  - Operative mortality
- Late results
  - Late mortality
  - Late complication
  - Re-operation
  - Late functional class

Surgical Complexity

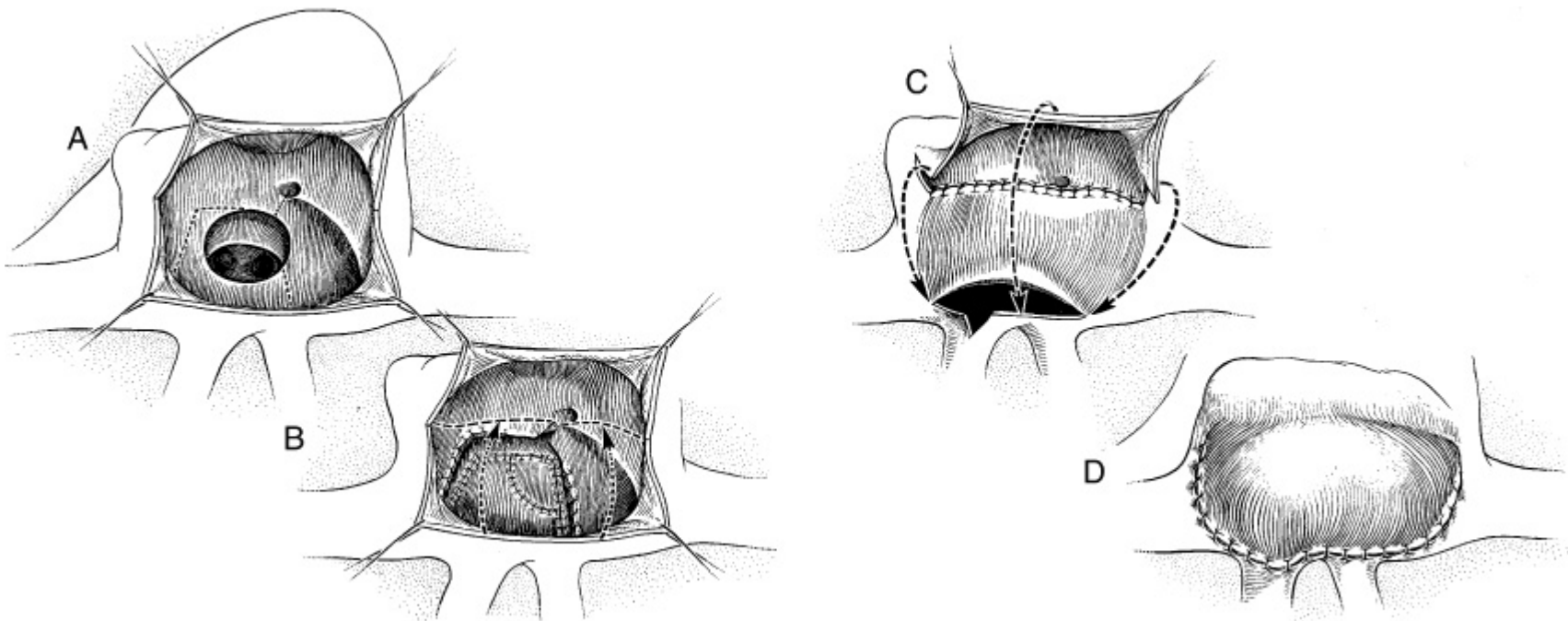
II. Anatomic Repair (Double-Switch Operation)

- Atrial Switch
  - Senning / Mustard procedure
- Arterial Switch
- Rastelli Operation

Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

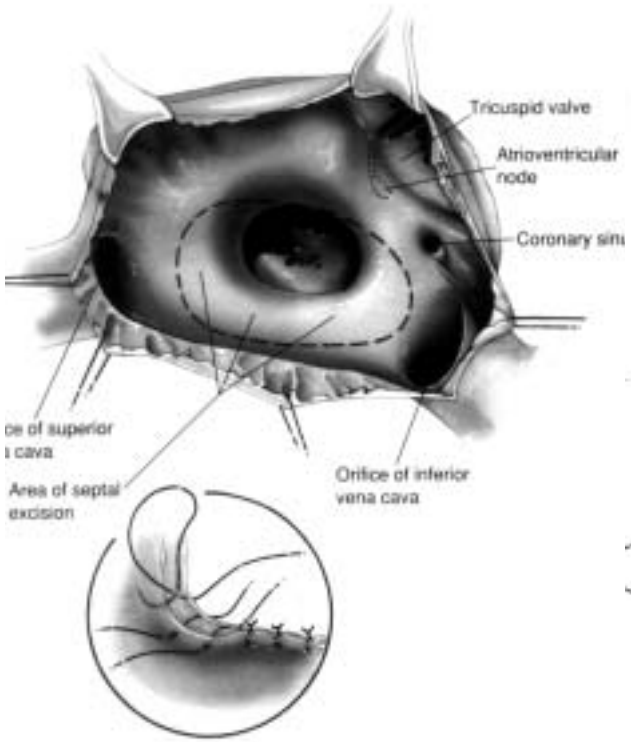
Atrial Switch: Senning procedure

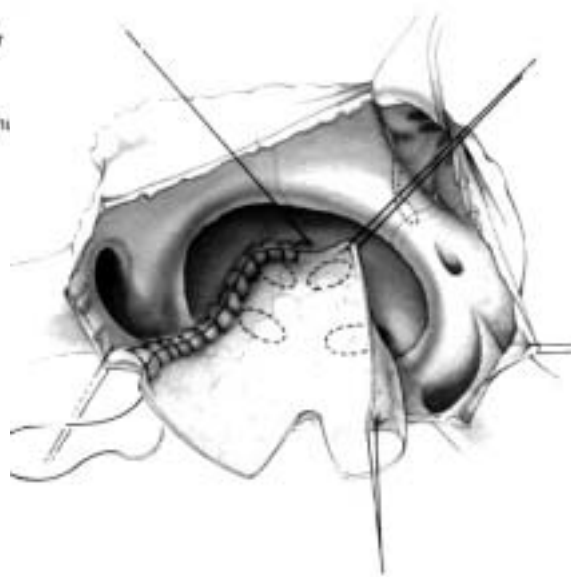


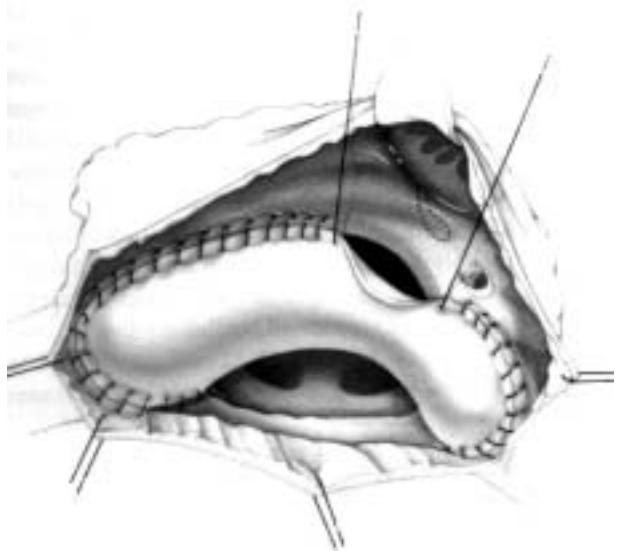
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Atrial Switch: Mustard procedure







Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Arterial Switch

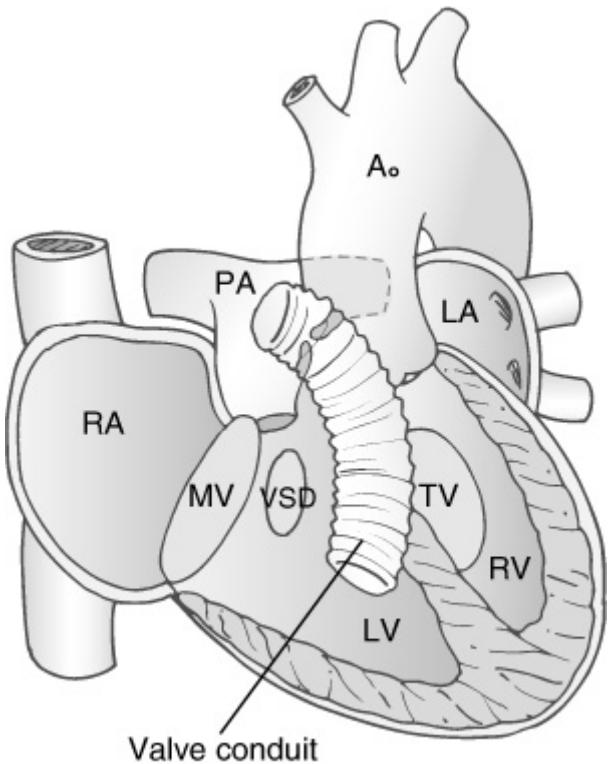
- VSD patch closure
- Resection of Conal septum (subpulmonary obstruction)

Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

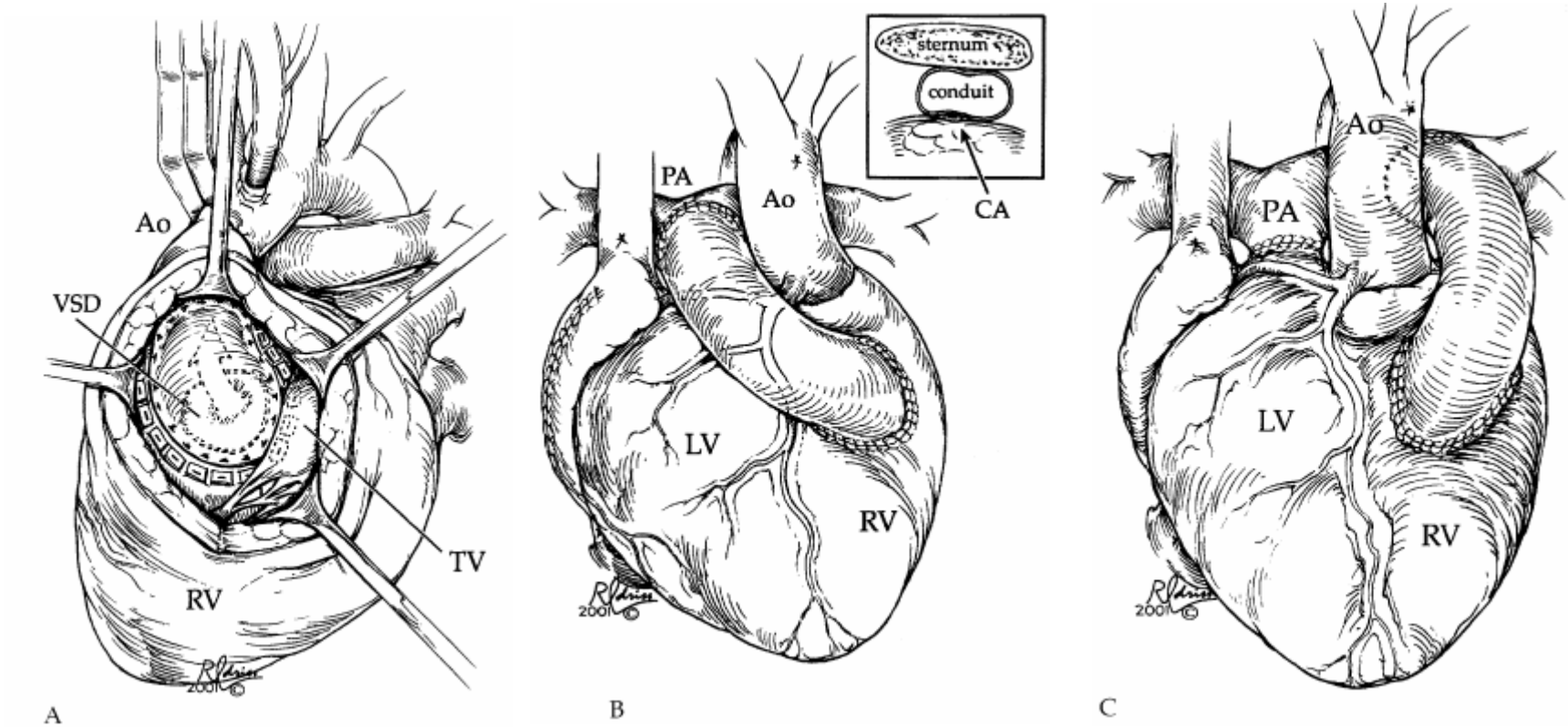
Rastelli Operation

- Baffling of LV outflow thorough VSD to Aorta
- RV-PA conduit



Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)



Ilbawi et al. Ann Thorac Surg 2002;73:594

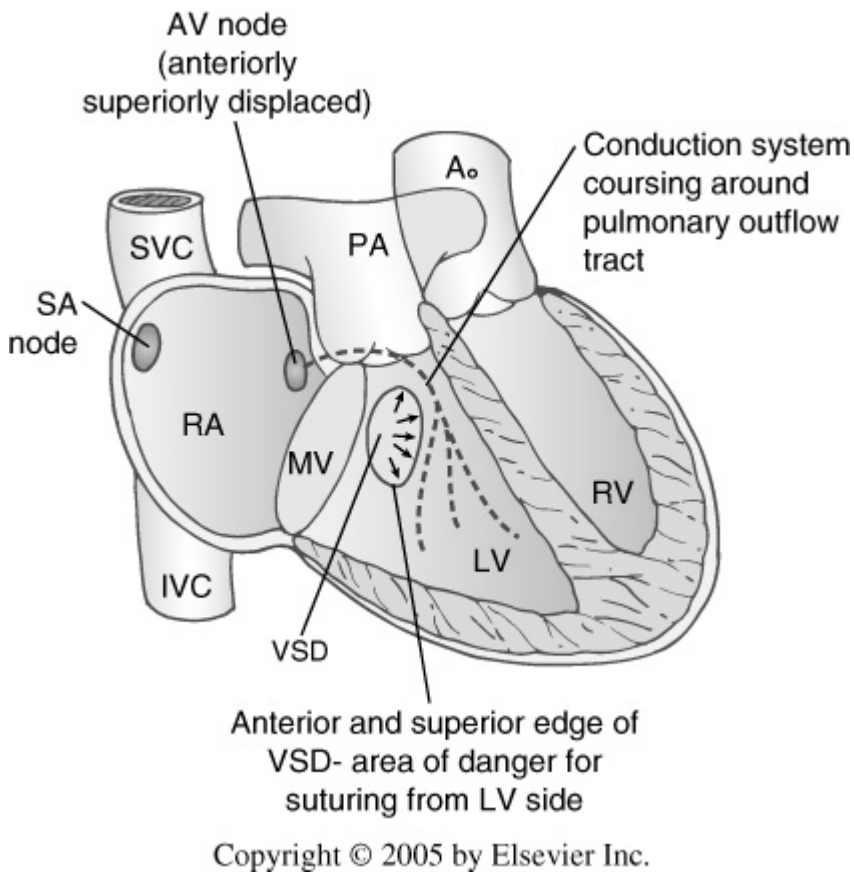
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Conduction system

Bundle of His runs from the anterior node, through the fibrous trigone, to pass anterior and immediately inferior to the pulmonary valve annulus.

When VSD is present bundle of His runs along the superoanterior border fo the VSD



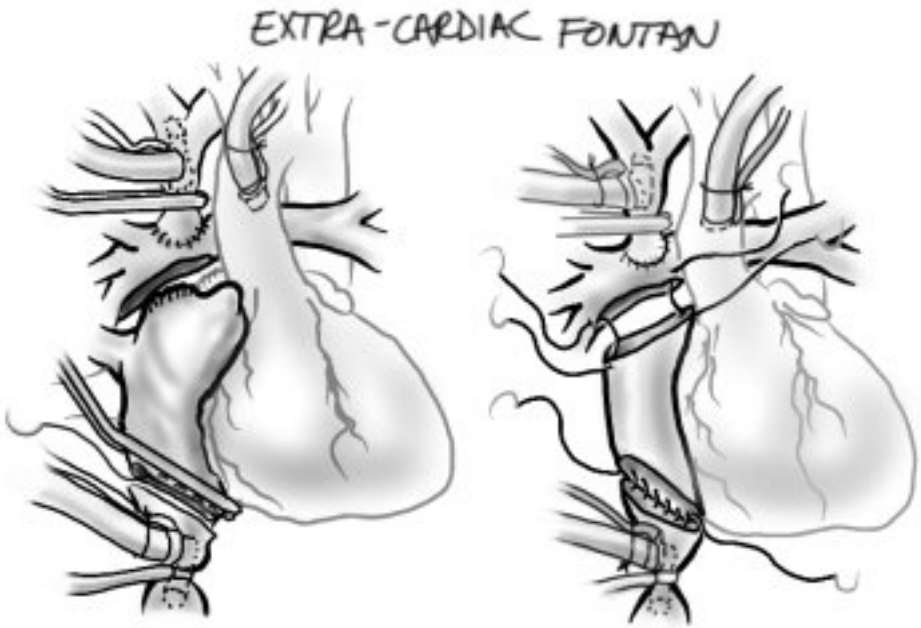
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

- Ilbawi et al.
  - Mustard procedure for venous switch moderate to deep hypothermia (20~24C)
  - CPB time: 180±23 min
  - ACC time: 147±17 min
- Langley et al.
  - Deep hypothermia (18C) in all cases
  - CPB time: 149 min
  - ACC time: 131 min
- Repair of the tricuspid left atrioventricular valve may be technically difficult, because of the posterior location.

Surgical Complexity

III. Fontan-type Operation



Extracardiac Lateral Tunnel

Extracardiac conduit TCPC

Operative Risk

II. Anatomic Repair (Double-Switch Operation)

Author		case	Mortality
Langley SM	1991~2001	54	3 (5.6 %)
Devaney EJ	1993~	17	0 (0 %)
Ilbawi MN	1989~2000	12	1 (9 %)
Mee RB	1991~2000	22	3 (33 %)
Imai H	1989~1993	18	2 (11 %)
Yagihara T	1987 ~ 1990	10	3 (30 %)

Operative Risk

II. Anatomic Repair (Double-Switch Operation)

Postoperative complete heart block

Author		case	Incidence
Langley SM	1991~2001	54	7/51 (14 %)
Ilbawi MN	1989~2000	12	1/12 (9 %)
Mee RB	1993~1999	27	0 (0 %)
Karl TR	1989~1996	14	3/10 (30 %)
Yagihara T	1987 ~ 1990	10	1/10 (10 %)

Operative Risk

III. Fontan-type Operation

Author		case	Mortality
Gentles TL	1973~1991	500	16.8% (27.1→7.5)
Stamm C	1987~1991	220	5.4 %
Yoshimura N	1988~2000	76	10.5 %
Azakie A	1994~1998	60	5.6 %
Woods RK	1995~2001	58	5 %

Operative Risk

III. Fontan-type Operation

Yoshimura et al.

Risk factors influencing early and later mortality after total cavopulmonary connection

J Thorac Cardiovasc Surg 1997;114:376

Risk factors for early mortality

- Ventricular morphology (RV)
- Prolonged CPB time (240 min)
- EF < 60%

Operative Risk

III. Fontan-type Operation

Gentles et al.

Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome

J Thorac Cardiovasc Surg 1997;114:376

Risk factors for early failure

- high mean PA pressure
- younger age
- Heterotaxy syndrome
- Tricuspid as only AV valve
- PA distortion
- AP connection
- absence of fenestration
- Longer CPB time

Operative Risk

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

J Thorac Cardiovasc Surg 2001;121:28

Predictors of early failure

- Age at Fontan
- Single RV
- Left AVV stenosis or atresia
- PA distortion
- Preop PVR

Early and Late Results

II. Anatomic Repair (Double-Switch Operation)

Ilbawi et al.

Intermediate Results of the anatomic repair for congenitally corrected transposition

Ann Thorac Surg 2002;73:594

12 patients with ccTGA and VSD, PS (n=10)
Mustard + ASO (n=2), Mustard + Rastelli (10)
Follow up: 0.6 ~ 10 (7.6 ± 3.1) year

Operative mortality	9 %
Surgical complete heart block	9 %
SVC obstruction	9 %
bradyarrhythmia	36 %
conduit replacement in 5.2 year follow up	45%
mild to moderate TR	18%
LV EF = 49 ~ 70%	

Early and Late Results

II. Anatomic Repair (Double-Switch Operation)

Langley et al.

Midterm results after restoration of the morphologically left ventricle to the systemic circulation in patients with congenitally corrected transposition of the great arteries

Ann Thorac Surg 2002;73:594

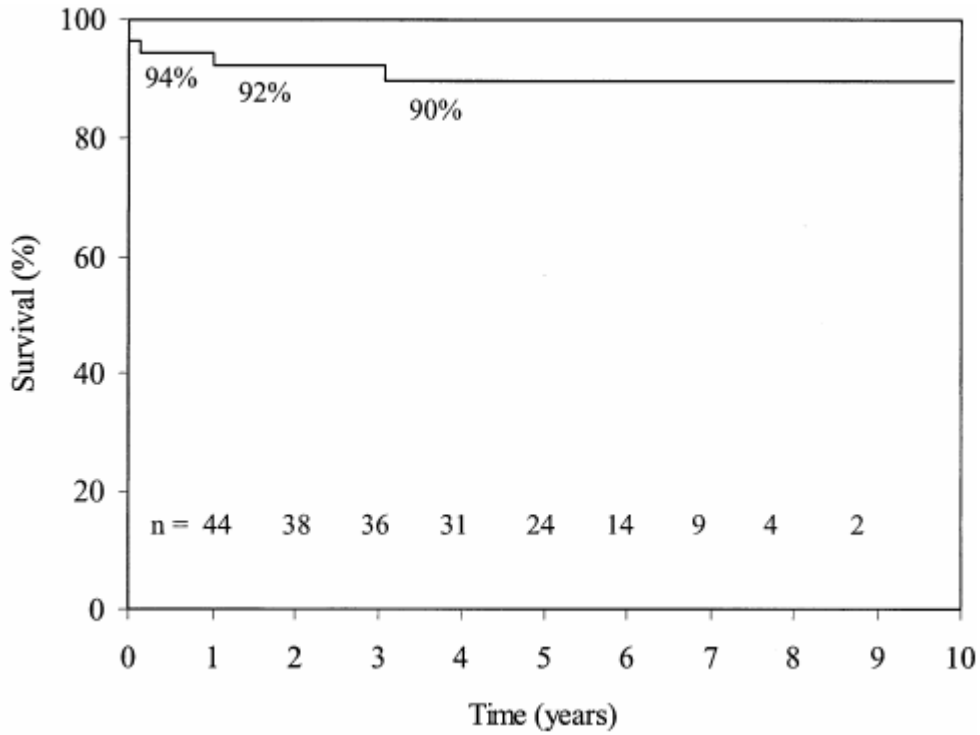
1991 ~ 2001
54 patients
ccTGA
AV discordance with DORV
Senning + ASO
Senning + Rastelli
Senning + intraventricular rerouting
Follow-up 4.4 year

Early mortality	5.6%
Heart block	13.7%
Chest reopen within 24 hour	9.3%
Kaplan-Meier Survival	
1 year	94.4%
9 year	89.7%

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Kaplan-Meier Survival for Anatomic Repair

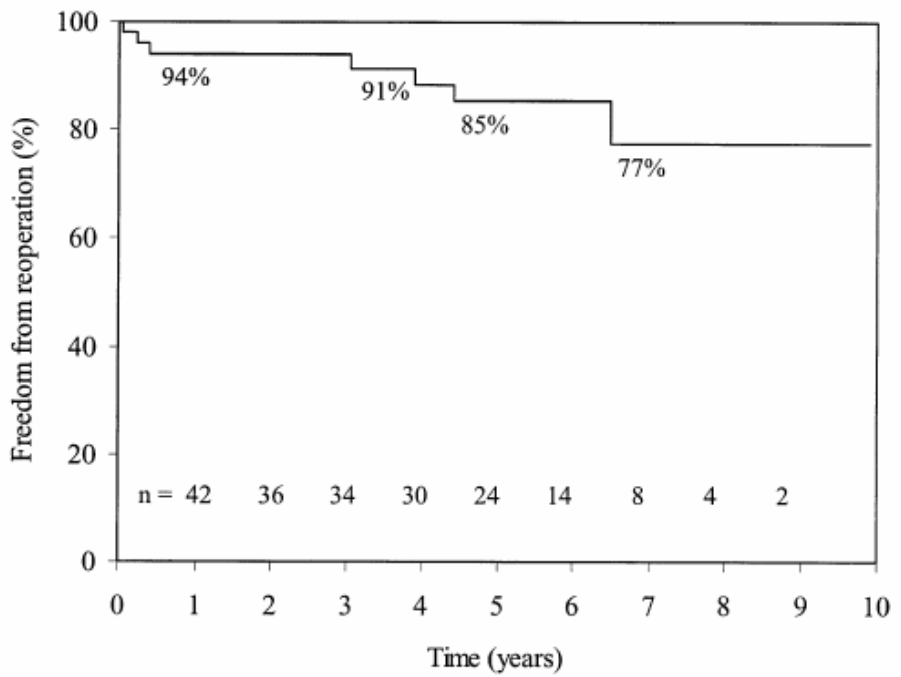


Langley et al. Ann Thorac Surg 2002;73:594

Early and Late Results

II. Anatomic Repair (Double-Switch Operation)

Freedom from reoperation



Langley et al. Ann Thorac Surg 2002;73:594

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

- Still, long-term data are lacking
- Long-term problems associated with Senning procedure for concordant TGA
  - Pathway obstruction
  - RV dysfunction
  - Arrhythmia
- Long-term problems associated with Rastelli procedure for concordant TGA

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Gelatt et al.

Arrhythmia and mortality after the Mustard procedure: A 30-year single-center experience

J Am Coll Cardiol 1997;29:194

534 children who underwent Mustard operation

Follow up: 11.6 ± 7.2 years.

There were 77 late deaths (16.1%)

with sudden death (n=31): the most frequent cause

Sinus rhythm was present in 77% at 5 years and 40% at 20 years.

Reoperation was required in 54 (10%).

Repair of an obstructed or leaking baffle 25 (5%)

Early and Late Results

II. Anatomic Repair (Double-Switch Operation)

Kreutzer et al.

Twenty-five-year experience with Rastelli repair for Transposition of the great arteries

J Thorac Cardiovasc Surg 2000;120:211-23.

1973 ~ 1998, 101 patients with D-transposition, VSD

Pulmonary stenosis (73), Pulmonary atresia (18), LVOTO (10)

Intraventricular Baffling + RV-PA continuity establishment

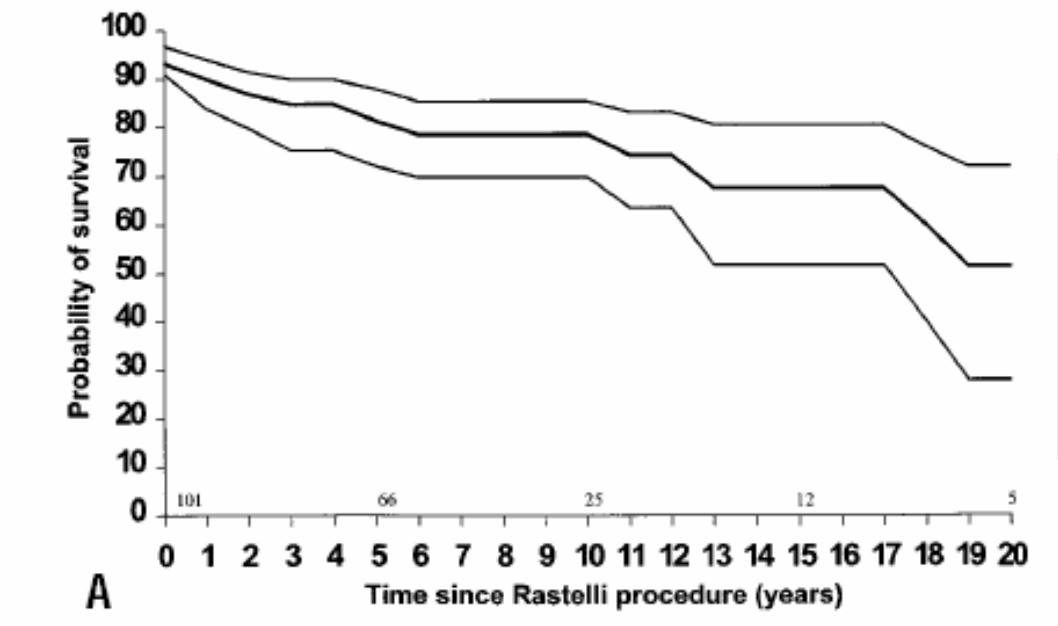
Early mortality: n=7 (7%)

Median follow-up: 8.5 years (0.4 ~ 22 years)

Long-term Resluts

II. Anatomic Repair (Double-Switch Operation)

Overall survival after the Rastelli Repair



5yr 82%

10yr 80%

15yr 68%

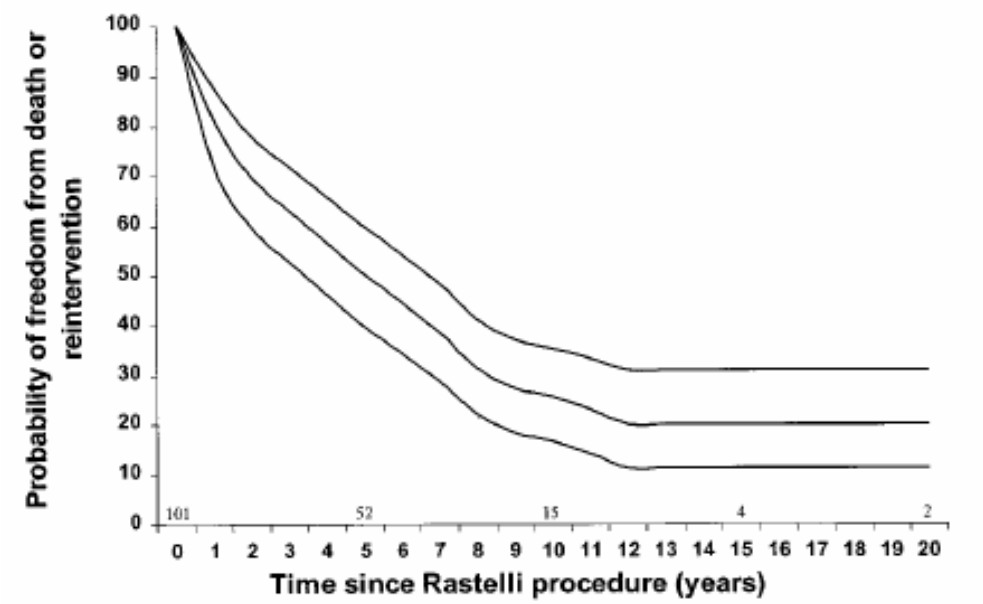
20yr 52%

Kreutzer et al. J Thorac Cardiovasc Surg 2000;120:211-23.

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Freedom from reintervention, reoperation, or death



Kreutzer et al. J Thorac Cardiovasc Surg 2000;120:211-23.

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Causes of late death or failure

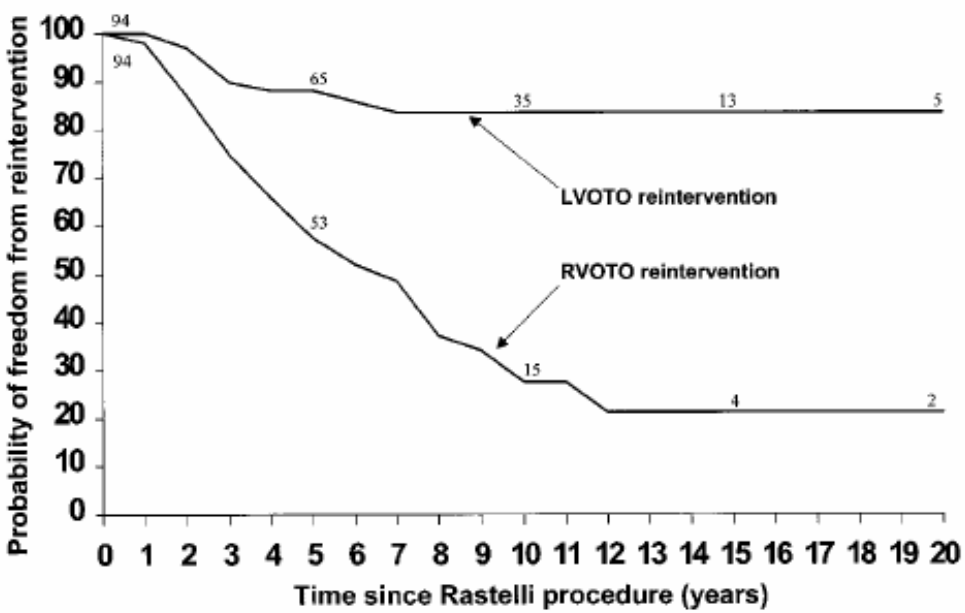
- Sudden death 5
- LV dysfunction 7
- Conduit pseudointima rupture 1
- Myocarditis 1
- Unknown 2
- At re-operation 2

Kreutzer et al. J Thorac Cardiovasc Surg 2000;120:211-23.

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Freedom from RVOTO and LVOTO reintervention



Kreutzer et al. J Thorac Cardiovasc Surg 2000;120:211-23.

Long-term Results

III. Fontan-type Operation

Gentles et al.

Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome

J Thorac Cardiovasc Surg 1997;114:376

1973 ~ 1991

Various modification of Fontan procedures

500 patients

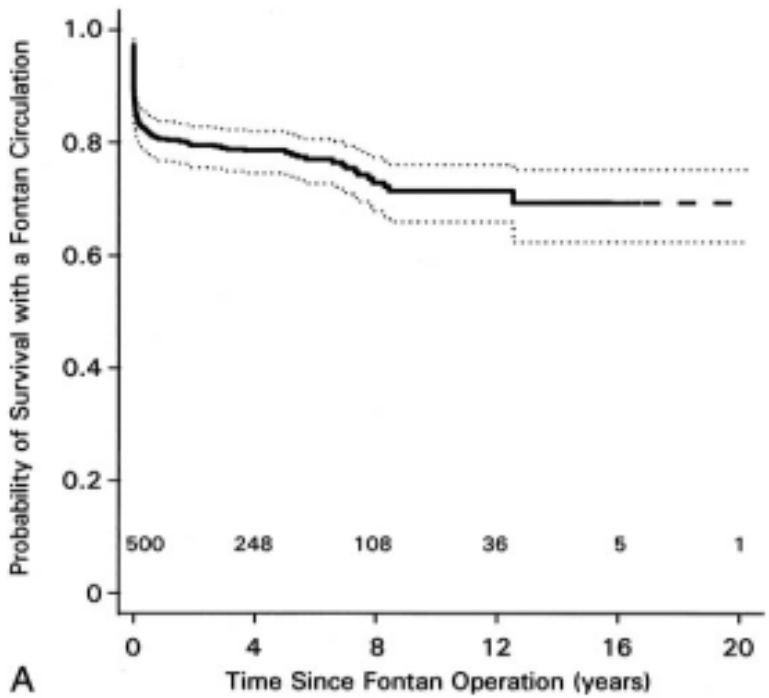
Early failure 16.8% (27.1 → 7.5%)

Follow-up: 2464 patient-year

Long-term Results

III. Fontan-type Operation

Probability of survival with a Fontan circulation

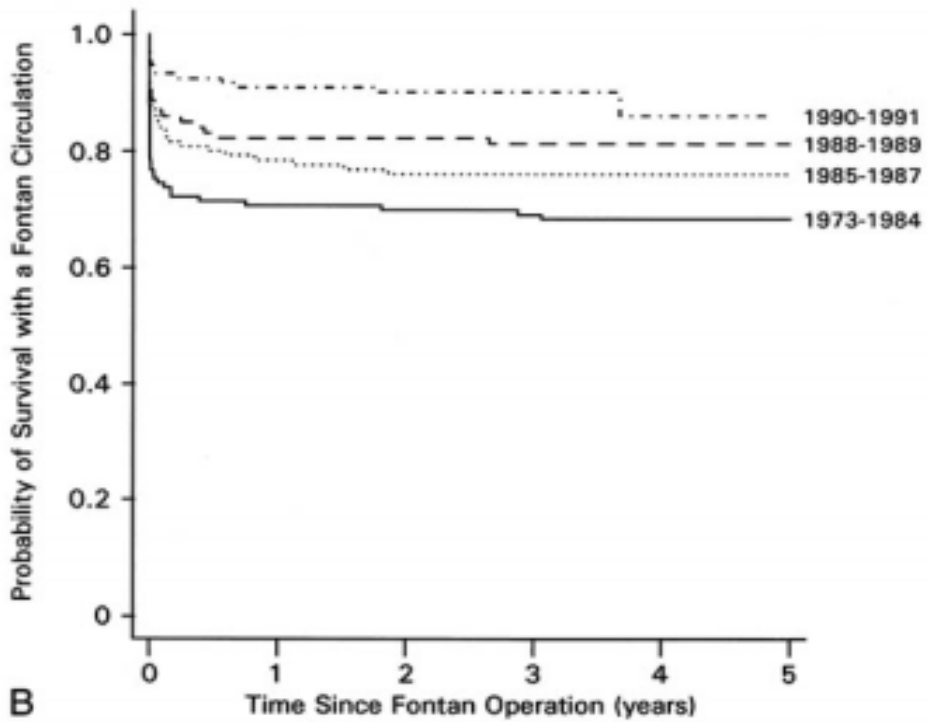


Gentles et al.J. Thorac. Cardiovasc. Surg 1997;114:376-391

Long-term Results

III. Fontan-type Operation

Probability of survival with a Fontan circulation

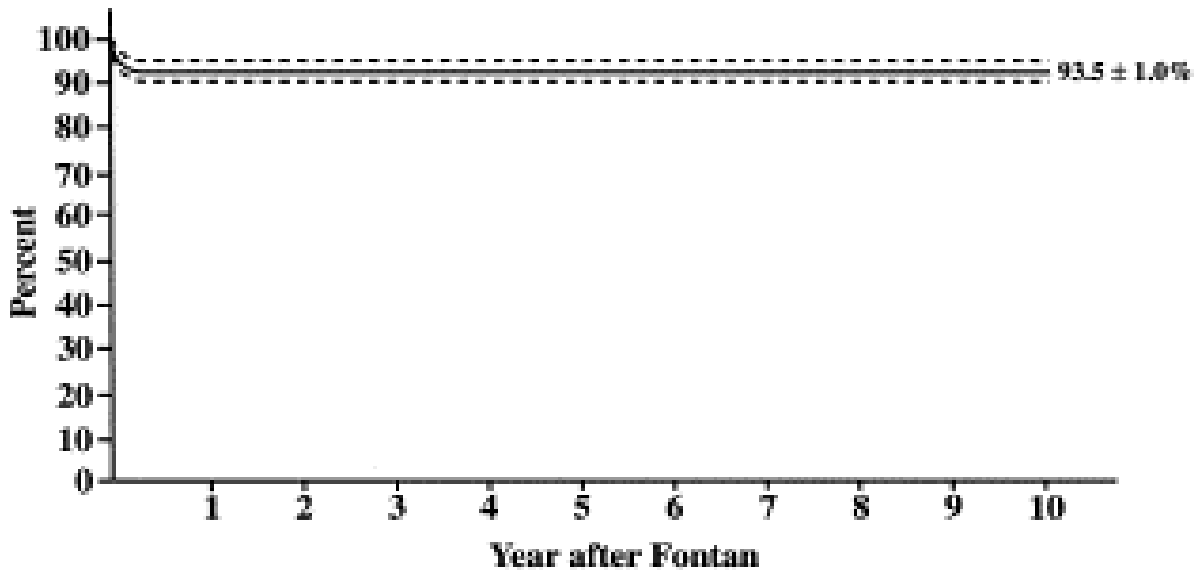


Gentles et al.J. Thorac. Cardiovasc. Surg 1997;114:376-391

Late Results

III. Fontan-type Operation

Actuarial survival after the modified Fontan procedure



Bando et al. Ann Thorac Surg 2000;69:1873.

Long-term Results

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

J Thorac Cardiovasc Surg 2001;121:28

1987 ~ 1991

Lateral tunnel Fontan procesures

220 patients

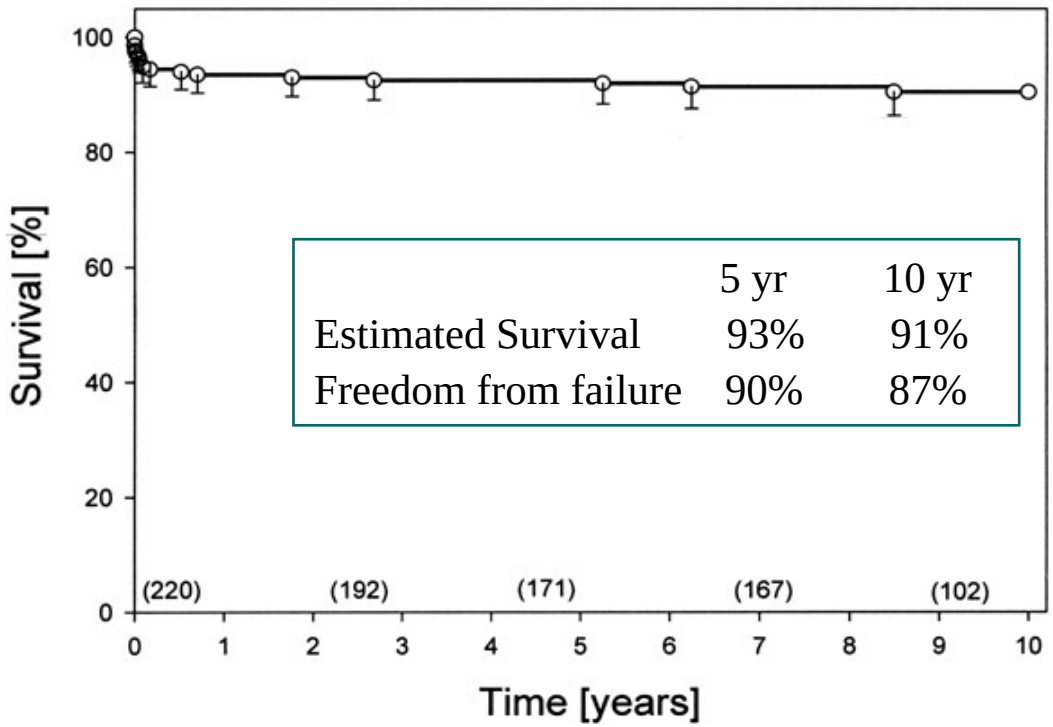
Early failure: 5.4%

Follow-up: 10.2 ± 0.6 (3.1~12.3) year

Long-term Results

III. Fontan-type Operation

Kaplan-Meier estimated overall survival after the lateral tunnel Fontan operation



Stamm et al. JTCS 2001;121:28

Long-term Results

III. Fontan-type Operation

Gentles et al.

Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome

J Thorac Cardiovasc Surg 1997;114:376

Risk factors for early failure

- high mean PA pressure
- younger age
- Heterotaxy syndrome
- Tricuspid as only AV valve
- PA distortion
- AP connection
- absence of fenestration
- Longer CPB time

Risk factors for late failure

- pacemaker before Fontan operation

Late Results

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

J Thorac Cardiovasc Surg 2001;121:28

Predictors of early failure

- Age at Fontan
- Single RV
- Left AVV stenosis or atresia
- PA distortion
- Preop PVR

Predictors of late failure

- Prior systemic-pulmonary shunt
- Prior CoA repair

Long-term Results

III. Fontan-type Operation

Yoshimura et al.

Risk factors influencing early and later mortality after total cavopulmonary connection

J Thorac Cardiovasc Surg 1997;114:376

<u>Risk factors for early mortality</u>

- Ventricular morphology (RV)
- Prolonged CPB time (240 min)
- EF < 60%

<u>Risk factors for late mortality</u>

- Heterotaxy syndrome
- AV valve regurgitation
- Prolonged CPB time (240 min)
- Prolonged ACC time (70 min)

Long-term Results

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

J Thorac Cardiovasc Surg 2001;121:28

Arrhythmia		5 yr	10 yr
Freedom from new supraventricular tachyarrhythmia		96%	91%
Freedom from new bradyarrhythmia		88%	79%
Protein Losing Enteropathy	3		
Thromboembolic event	7		

Long-term Results

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

J Thorac Cardiovasc Surg 2001;121:28

Functional State	
NYHA I	41 %
NYHA II	53 %
NYHA III	6 %
NYHA IV	0 %

Conclusion

III. Fontan-type Operation

- Less complex surgical procedure
- Comparable operative risk
- Fewer development of complete AV block
- Arrhythmogenic suture lines on atrium can be avoided.
- Favorable candidate for Fontan operation
  - Biventricular morphology
  - no PA distortion, normal PVR
- The patient might not need further operations such as conduit changes and atrial baffle revision
- Proven long-term outcome