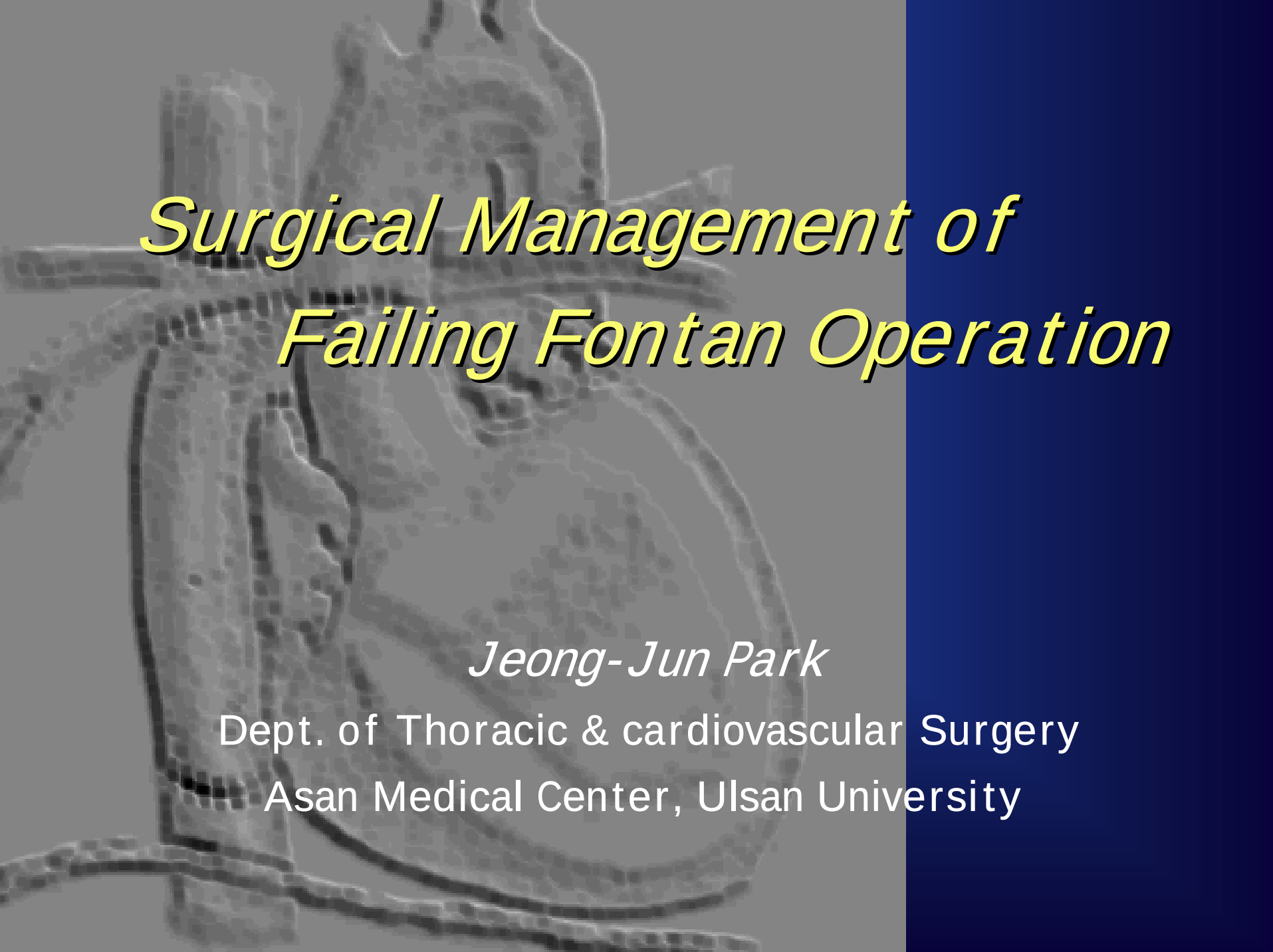

Surgical Management of Failing Fontan Operation

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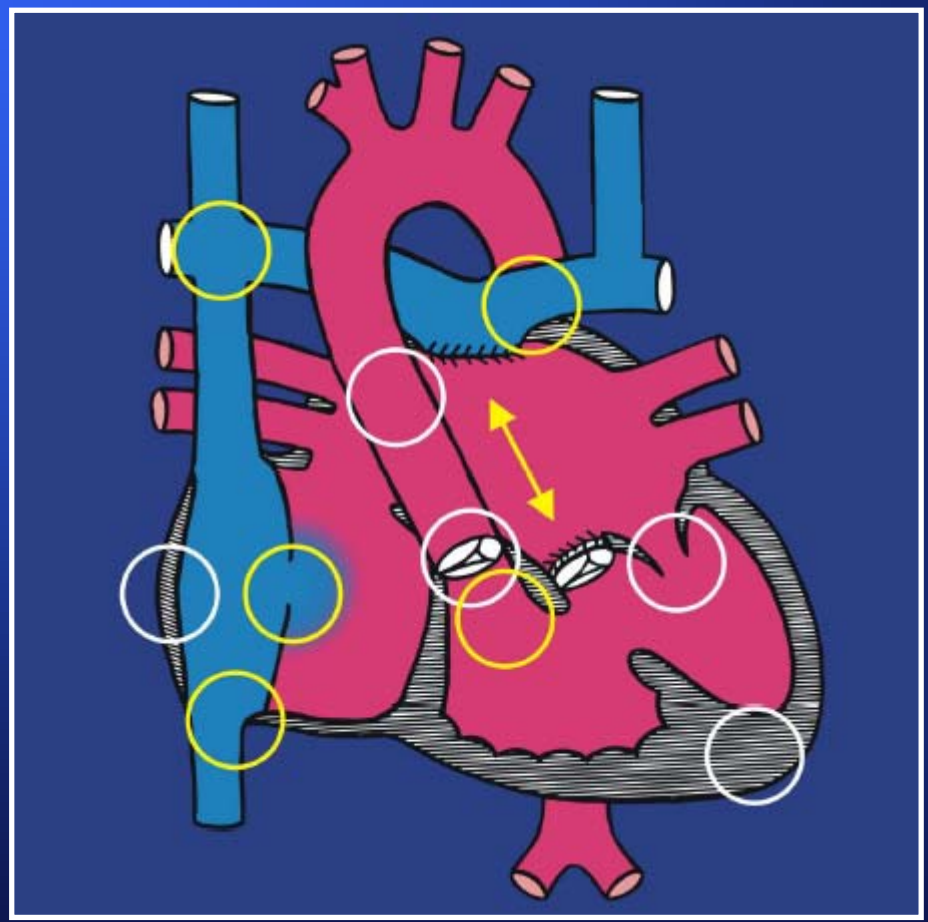
Fontan operation



Surgical reinterventions following the Fontan procedure☆

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Abstract

Objective: The Fontan procedure is utilized as a final reconstructive procedure for patients with functional single ventricle. Short- and long-term outcomes have improved significantly, however, some patients require additional cardiac procedures following the Fontan operation. The outcomes for these reinterventions are not known. **Methods**: Cardiac Surgery and Cardiac Intensive Care Unit databases at The Children's Hospital of Philadelphia were reviewed to identify all patients who underwent cardiac surgery after a previous Fontan operation between January 1, 1995 and December 31, 2001. **Results**: During the study period, 123 procedures were performed in 71 patients. The median time from Fontan to reoperation was 3.6 years (range 0.1–20 years). Indications for reintervention included arrhythmia, cyanosis, 'failing' Fontan circulation or exercise intolerance, protein losing enteropathy, atrioventricular valve (AVV) regurgitation, and other indications. Procedures included pacemaker insertion or revision ($n = 59$, 48%), reinclusion of previously excluded hepatic veins ($n = 16$, 13%), revision to either a lateral tunnel or extra-cardiac conduit Fontan ($n = 13$, 11%), cardiac transplantation ($n = 9$, 7%), enlargement or creation of a baffle fenestration ($n = 6$, 5%), isolated AVV repair or replacement ($n = 2$, 2%), and other procedures ($n = 18$, 14%). There were five early and five late deaths. Hospital mortality was greatest for patients undergoing cardiac transplantation (4/9, 44%), accounting for 80% of the early deaths. **Conclusions**: Surgical reinterventions following the Fontan procedure may be necessary for multiple indications which result in impairment of the Fontan circulation. Most reinterventions can be performed with minimal morbidity and mortality. Survival for patients requiring cardiac transplantation following the Fontan procedure remains poor.

1995-2001, 123 procedures in 71 pts

Final modes of failure

1/ Systemic Venous congestion
2/ „Deep“ Cyanosis
3/ Arrhythmia
4/ Protein Losing Enteropathy

Multifactorial

1.Systemic venous congestion

1/ Sten./Thromb. in cavo-pulm.pathway — intervention / surgery

2/ Pulmonary artery stenosis — intervention / surgery

3/ Pulmonary venous obstruction — surgery / intervention ?

4/ AV-Valve dysfunction — surgery

5/ Aortic valve dysfunction — surgery

6/ Subaortic stenosis — surgery

7/ Coarctatio aortae — intervention / surgery

8/ Myocardial dysfunction — multisite pacing / transplantation / LVAD

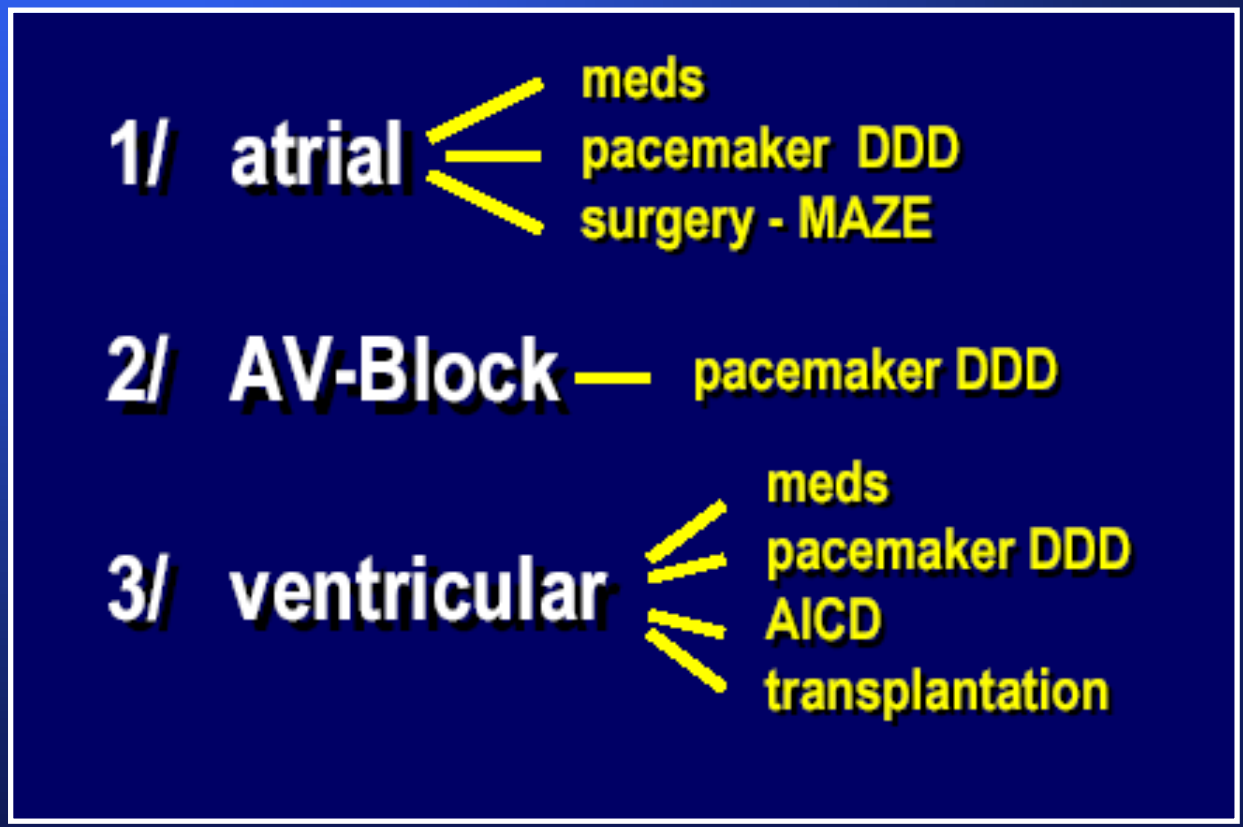
2.Deep cyanosis from:

1/ Large residual interatrial shunt → intervention, surgery

2/ Systemic venous / left atrial shunts → intervention, surgery

3/ Intrapulmonary shunts → intervention, nothing, transplantation

3.Arrhythmia



4.Protein losing enteropathy

1/ Systemic venous congestion
- intervention
- meds
- surgery incl.takedown

2/ No venous systemic congestion
- meds ?
- takedown

3/ Low output
- meds ?
- surgical pacemaker (DDD)
- multisite pacing

Conversion Fontan operation; Multicenter study

REVISION OF PREVIOUS FONTAN CONNECTIONS TO TOTAL EXTRACARDIAC CAVOPULMONARY ANASTOMOSIS: A MULTICENTER EXPERIENCE

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Background: Conversion to total extracardiac cavopulmonary anastomosis is an option for managing patients with dysfunction of a prior Fontan connection. *Methods:* Thirty-one patients (19.9 ± 8.8 years) underwent revision of a previous Fontan connection to total extracardiac cavopulmonary anastomosis at four institutions. Complications of the previous Fontan connection included atrial tachyarrhythmias (n = 20), progressive heart failure (n = 17), Fontan pathway obstruction (n = 10), effusions (n = 10), pulmonary venous obstruction by an enlarged right atrium (n = 6), protein-losing enteropathy (n = 3), right atrial thrombus (n = 2), subaortic stenosis (n = 1), atrioventricular valve regurgitation (n = 3), and Fontan baffle leak (n = 5). Conversion to an extracardiac cavopulmonary connection was performed with a nonvalved conduit from the inferior vena cava to the right pulmonary artery, with additional procedures as necessary. *Results:* There have been 3 deaths. Two patients died in the perioperative period of heart failure and massive effusions. The third patient died suddenly 8 months after the operation. All surviving patients were in New York Heart Association class I (n = 20) or II (n = 7), except for 1 patient who underwent heart transplantation. Early postoperative arrhythmias occurred in 10 patients: 4 required pacemakers, and medical therapy was sufficient in 6. In 15 patients, pre-revision arrhythmias were improved. Effusions resolved in all but 1 of the patients in whom they were present before revision. The condition of 2 patients with protein-losing enteropathy improved within 30 days. *Conclusions:* Conversion of a failing Fontan connection to extracardiac cavopulmonary connection can be achieved with low morbidity and mortality. Optimally, revision should be undertaken early in symptomatic patients before irreversible ventricular failure ensues.
(J Thorac Cardiovasc Surg 2000;119:340-6)

JTCS 2000

- 1992-1999
- n=31
  - AP= 29
  - AV= 1
  - LT=1

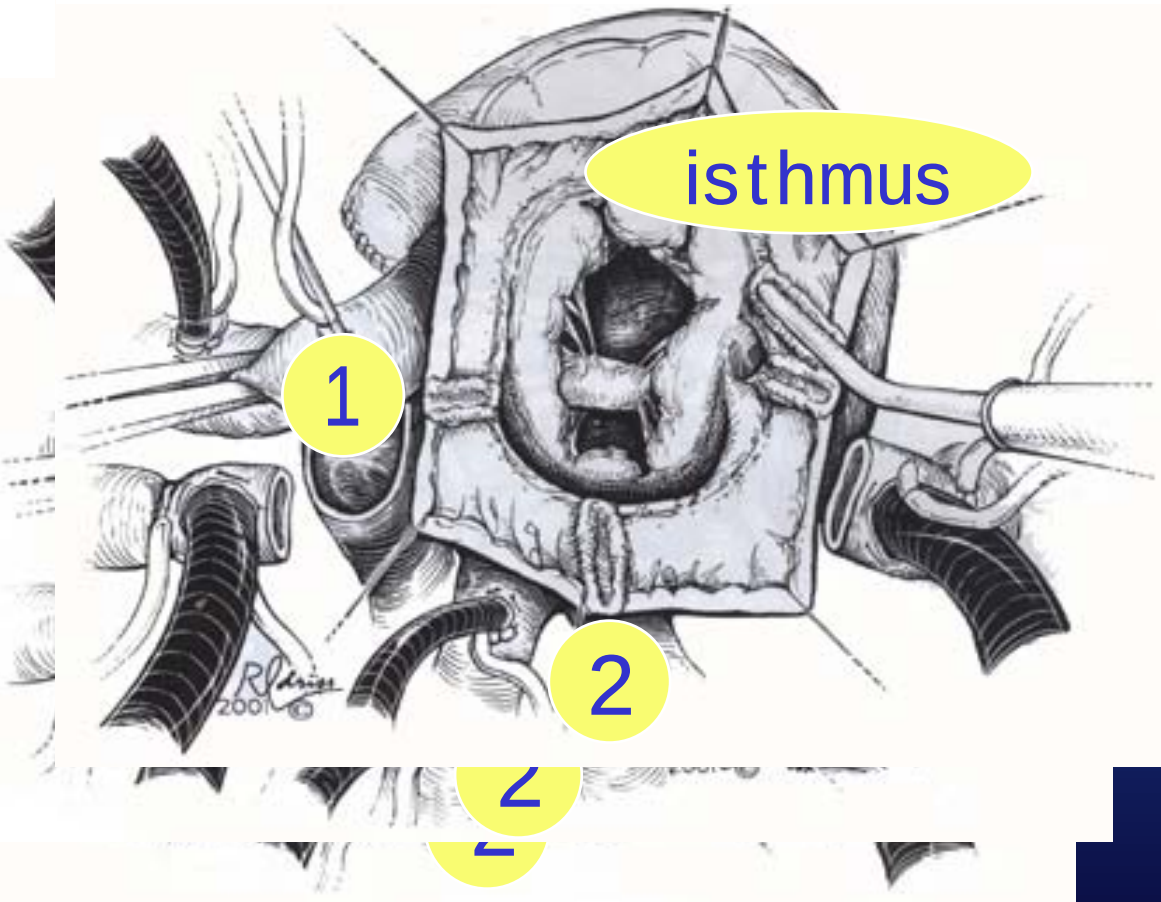
Fontan conversion with arrhythmia surgery

Mavroudis, Ped Card Surg Ann 2002

- Conversion without arrhythmia surgery: higher recurrence
- Evolution of technique: EP study
  1) Isthmus block
  2) Rt. Sided Maze operation: Atrial reentrant tachy
  3) Maze-Cox III operation: Afib
  + Permanent Pacemaker implantation

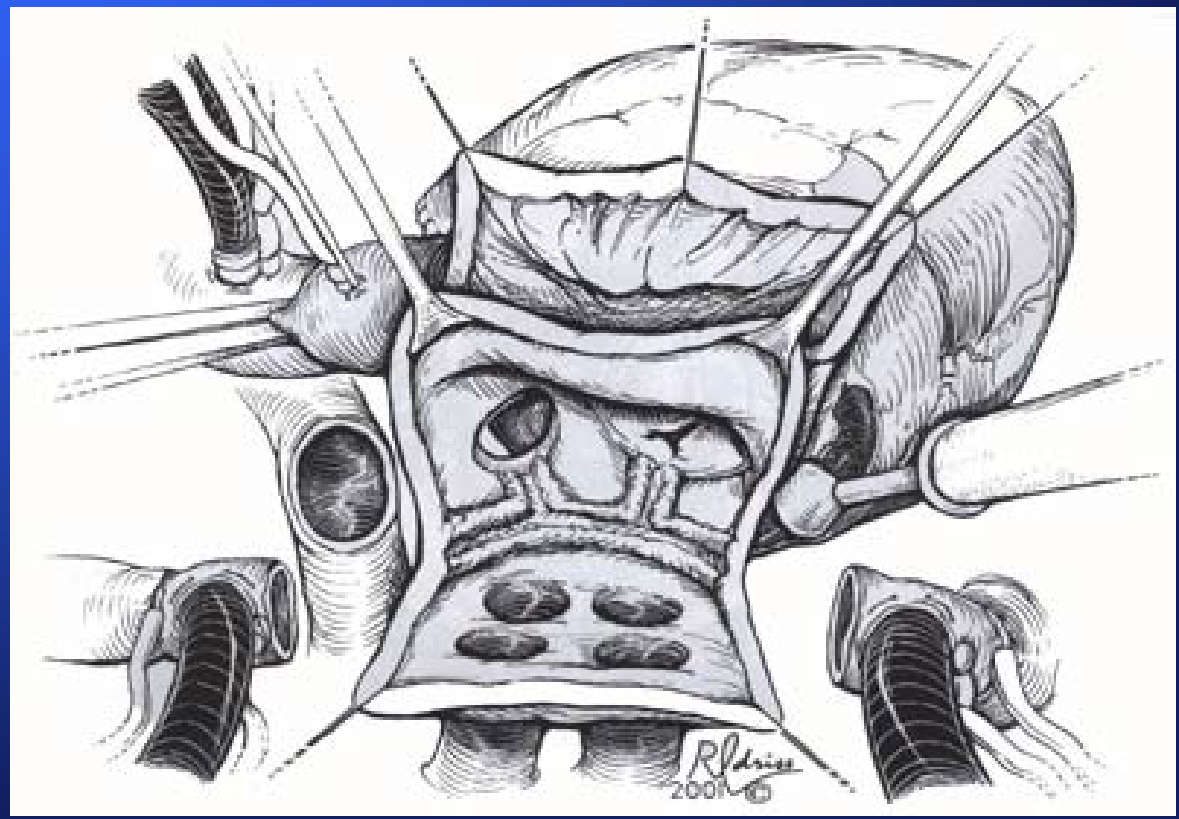
Right-sided Maze operation

Unbalanced AVSD



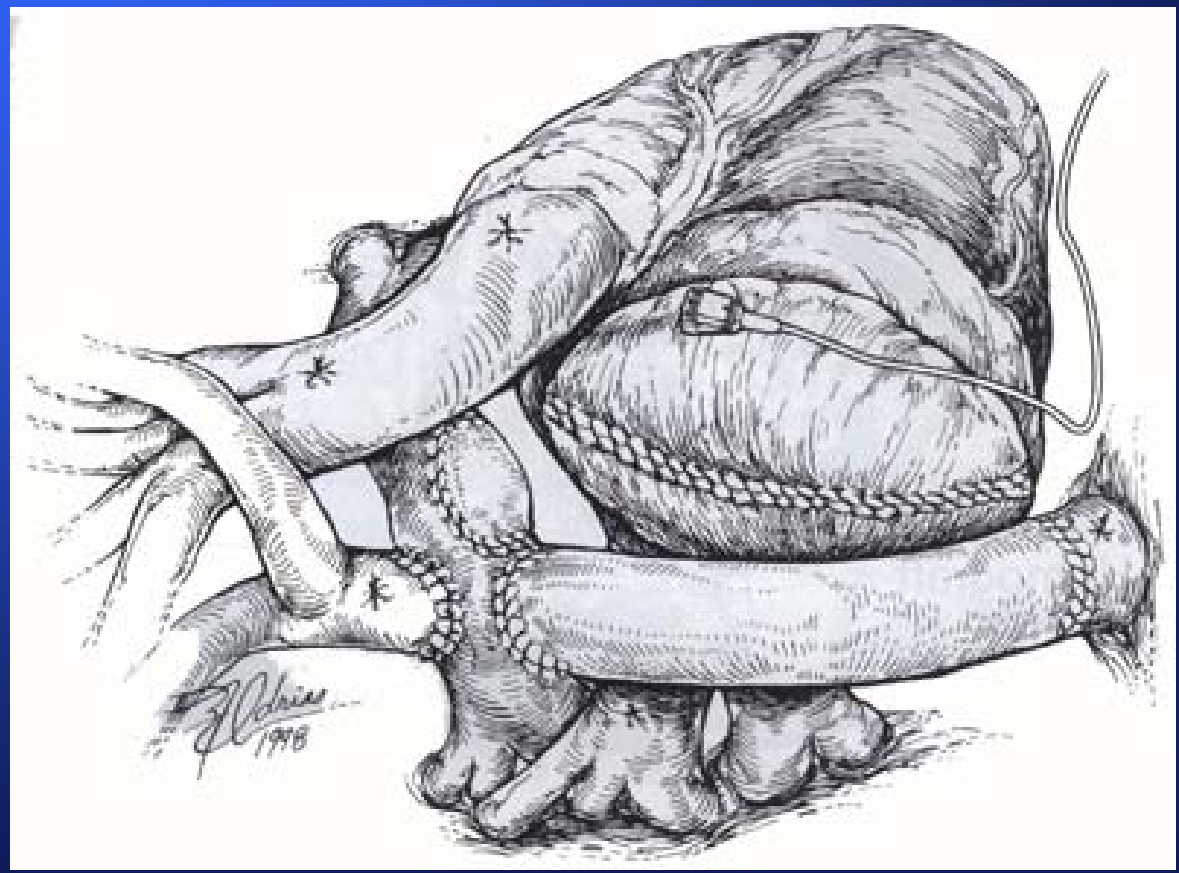
RA reduction
Cryoablation
: -60°c for 90 sec

Left sided completion of Maze-Cox III procedure



* PV Isolation / LAA resection
* Cryoablation: CS for 180 sec

Fontan conversion with arrhythmia surgery



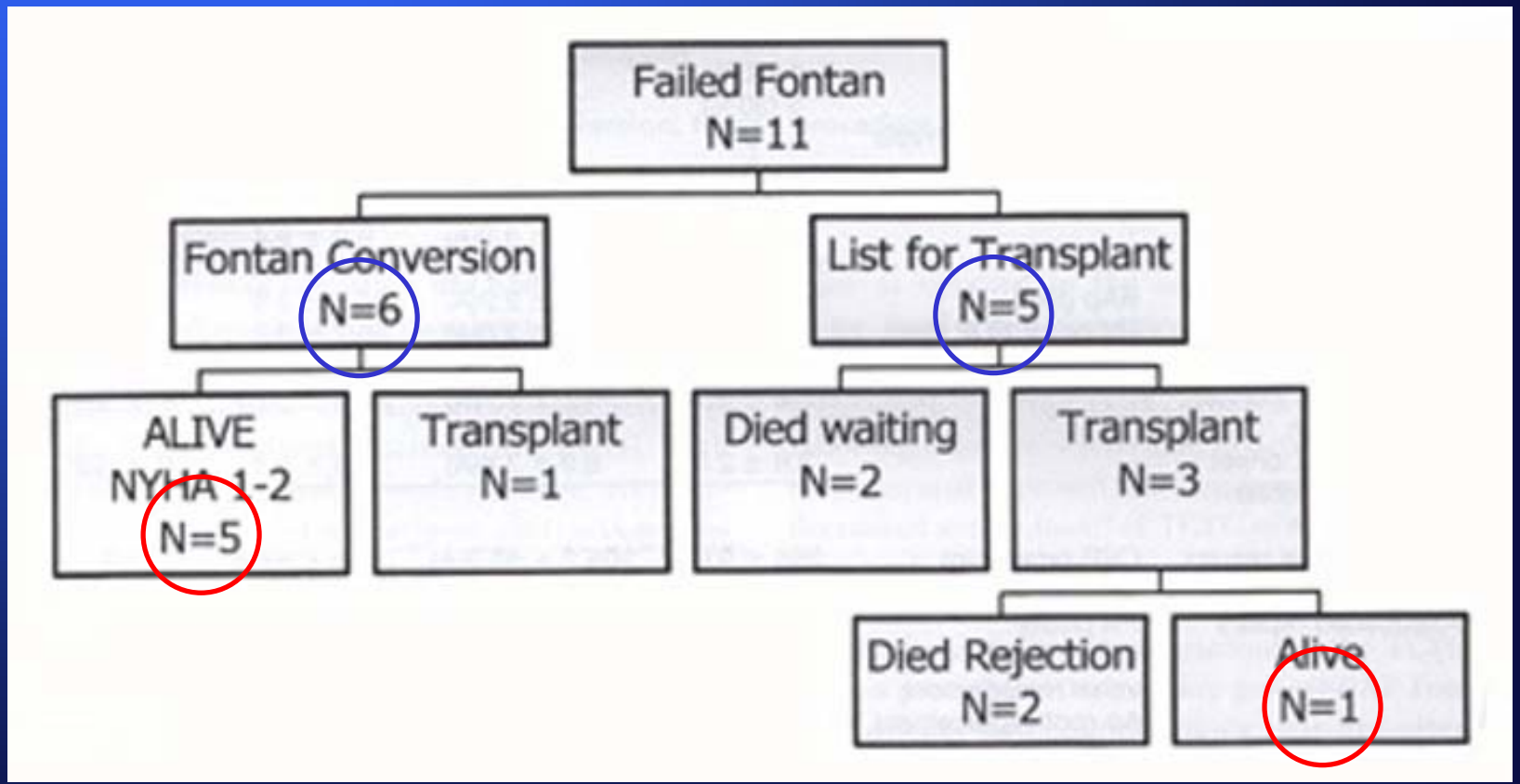
* Steroid-eluting epicardial leads

Fontan conversion with arrhythmia surgery

Mavroudis, Ped Card Surg Ann 2002

- 1994~ 2001, n=41 (1 PLE)
- Arrhythmia surgery
  1) Atrial reentry tachycardia (27)
     : isthmus block(10) / Rt. Sided Maze (17)
  2) Afib (14): Maze-Cox III
  3) PPM: 39 (atrial 34, dual 5)
- Mortality(-) / TPL in 3 pts
- Arrhythmia recur 12.2% / 9% on Mx
- Improvement of functional class & exercise tolerance

Failed Fontan initially considered TPL



Fontan conversion with arrhythmia surgery

Mavroudis, Ped Card Surg Ann 2002

- Early operation before the onset of Afib
  - prevention of progressive myocardial dysfunction
  - cessation of anti-arrhythmic agent
  - improved functional state

Transplantation

* Primary indication: ventricular dysfunction
* Highest risk group for TPL

: Difficulty in

Pre-TPL evaluation, TPL, Postop. management

1) Limited ability to assess hemodynamics
2) Complex anatomy
3) Multiple prior procedure
4) Underlying pathologic state

Pretransplant Evaluation

* Social, psychological, medical compliance
* Prospective cross matching: lymphocytotoxic Ab ↑
* Evaluation for Cardiogenic cirrhosis
* Cardiac & extracardiac vascular anatomy
  - dextrocardia, situs inversus, GA arrangement
  - systemic & pulmonary vein anomaly
  - patency of neck & femoral vss

Pretransplant Evaluation

Cardiac catheterization

* Ventricular dysfunction
  - unreliable VEDP
    : underloaded ventricle d/t LCO
    : other correctable anatomic cause
  - Revision vs TPL
    : No published data to guide this prediction
* PVR: PAP & transpulmonary gradient
  - practical value: restrictive
  - low output, multiple level of systemic v. obstruction
    unequal PBF, unilat. PV obstruction, PAVM

Timing of TPL listing

* No established criteria
  - not amenable to medical or surgical Tx
  - hepatic dysfunction
  - early Fontan failure
    (s technically correctable problem)

* Relative stable NYHA II~III pts: subjective

Surgical consideration

* Donor selection: exclude
  - marginal cardiac function
  - donor to recipient bwt ratio <1.0
* Procurement

  through understanding of recipient anatomy
* Transplant procedure
  - moderate hypothemia: 22~25^{o}C
  - SVC, IVC length
  - PA reconstruction

Outcome of TPL

* Small No. with limited F/U
* Primary cause of early mortality
  : hemorrhage & graft failure
* Improvement of PLE

Table 1. Reported Outcomes of Transplantation for Fontan Failure

Study	n	Hospital Mortality	Follow-Up (mos)	Survival Estimate
Hsu et al[1]	9	33%	23	67%
Carey et al[8]	9	33%	17	67%
Lamour et al[9]	8	38%	35	50%
Michielon et al[10]	6	67%	ND	ND
Mitchell et al[17]	15	7%	60	82%

Mitchell, Ped Card Surg Ann 2004

Mechanical Assist

Challenging groups to support with ECMO

Table 1. Published Literature of Fontan and BDG Patients Supported With ECMO

Author	Year	Series size	Fontan/BDG patients
Kanter et. al [13]	1987	13 patients	1 Fontan, died
Klein et. al. [14]	1990	39 patients	4 Fontans, 4 died
Ziomek et. al. [15]	1992	24 patients	3 Fontans, 2 survived including 1 Fontan takedown on ECMO
Saito et. al. [12]	1993	1 patient	Fontan, survived
Dalton et. al. [16]	1993	29 patients	3 Fontans, 2 survived including 1 transplant from ECMO
Kulik et. al. [17]	1996	64 patients	18 cavopulmonary connections, 3 survived
Jaggers et. al. [18]	2000	35 patients (all infants)	1 BDG, died
Aharon et. al. [9]	2001	50 patients	2 Fontans, 2 survived; 1 BDG, died

BDG = bidirectional Glenn; ECMO = extracorporeal membrane oxygenation.

ELSO: 1986~2002, 24.8% survival in Fontan & BDG pts

Extracorporeal Membrane Oxygenation Support of the Fontan and Bidirectional Glenn Circulations

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* n=20 (16Fontan, 4 BDG)
* Main Ix: myocardial failure
* 50% weaned, 30% survival
* Better outcome
  : Good myocardial fxn c an acute reversible event
* Poor outcome
  : Adult-sized pts, progressive myocardial failure

Maximizing venous drainage & systemic perfusion

(multiple venous cannula)

Early before the development of end-organ dysfunction

Conclusions

* Surgical intervention for failing Fontan operation
  - *amenable to anatomic relief*
  - *previous AP Fontan c/s arrhythmia*
    : Conversion c/s arrhythmia surgery
  - *TPL or ECMO for myocardial dysfuntion*
    : challenging

Surgical Approach in Patie
Failing Cavo-Pulmonary Ci

While patients with failing atrio-pulmonary
may benefit from conversion to cavo-pulmonary circulation, a surgical approach in failing cavo-pulmonary circulation is only justified in patients with clearcut pathology amenable to anatomic relief.
Additional atrial arrhythmia surgery and DDD-pacemaker (ventricular multiside ?) may add to benefit.

Table 2
Indications for reintervention

Arrhythmia/pacemaker
Bradyarrhythmia
Tachyarrhythmia
Generator malfunction
Lead malfunction
Cyanosis
Hepatic vein exclusion
Baffle leak
'Failing' Fontan circulation
Progressive exercise intolerance
Congestive heart failure
Protein losing enteropathy
Atrioventricular valve regurgitation
Other
Fontan pathway obstruction
Pleural/pericardial effusion
Osteomyelitis (pacing system related)
Thromboembolic events