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# Surgical Options in Fontan-Kreutzer Failure


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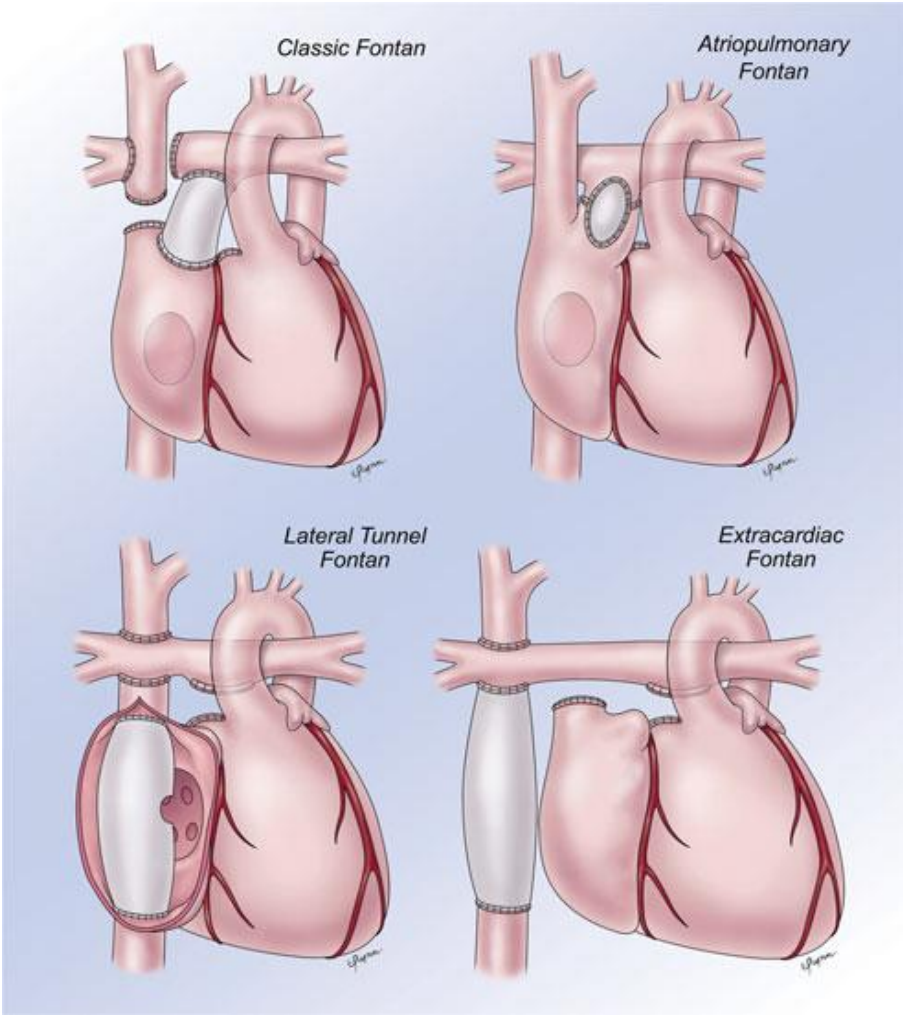
# Fontan-Kreutzer Failure

Early vs. Chronic

Mode of Failure:

Pump vs. Non Pump




Valente AM, Landzberg MJ, Powell AJ: Adult congenital heart disease. Edited by Libby P. In Essential Atlas of Cardiovascular Disease. New York, New York: Springer; 2009:231–246

# Therapy: Fontan-Kreutzer Failure

- Medical
  - Ventricular Dysfunction
  - Pulmonary Vascular Resistance
  - Arrhythmia

- Invasive
  - EP - Arrhythmia
  - Catheter Based – Baffles and Fenestrations
  - Lymphatic

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  - Lymphatic
  - Valve Repair and Replacement
  - Conversion
  - Mechanical Support
    - Destination?
  - Transplant
    - Multiorgan

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# Surgical and Catheter-Based Reinterventions Are Common in Long-Term Survivors of the Fontan Operation

Tacy E. Downing, MD; Kiona Y. Allen, MD; David J. Goldberg, MD; Lindsay S. Rogers, MD; Chitra Ravishankar, MD; Jack Rychik, MD; Stephanie Fuller, MD; Lisa M. Montenegro, MD; James M. Steven, MD; Matthew J. Gillespie, MD; Jonathan J. Rome, MD; Thomas L. Spray, MD; Susan C. Nicolson, MD; J. William Gaynor, MD; Andrew C. Glatz, MD, MSCE

*Circ Cardiovasc Interv*. 2017;10:e004924

**Table 2. Post-Fontan Surgical Reinterventions**

| Surgery | Total | Early* | Surgery | Total | Early* |
|---|---|---|---|---|---|
| Pacemaker | 100 | 11 | Fontan takedown | 10 | 10 |
| Hepatic vein inclusion | 21 | 6 | Aortic valve repair/replace | 7 | 0 |
| Fontan revision | 19 | 4 | Atrial septectomy | 7 | 2 |
| Heart transplant | 19 | 4 | Pulmonary arterioplasty | 5 | 4 |
| Miscellaneous† | 17 | 7 | Maze procedure | 3 | 0 |
| AV valve repair/replace | 14 | 1 | Arch revision | 1 | 0 |
| Create fenestration | 13 | 7 | … | … | … |

AV indicates atrioventricular; and VSD, ventricular septal defect.
*Within 1 year post-Fontan.
†Miscellaneous: aortic root replacement (3), fenestration closure (3); pericardial window (2), aortic aneurysm repair, repair intraoperative injury, resect subaortic stenosis, VSD enlargement, thoracic duct ligation, ligate left superior vena cava, thrombectomy, and sutureless pulmonary vein repair.

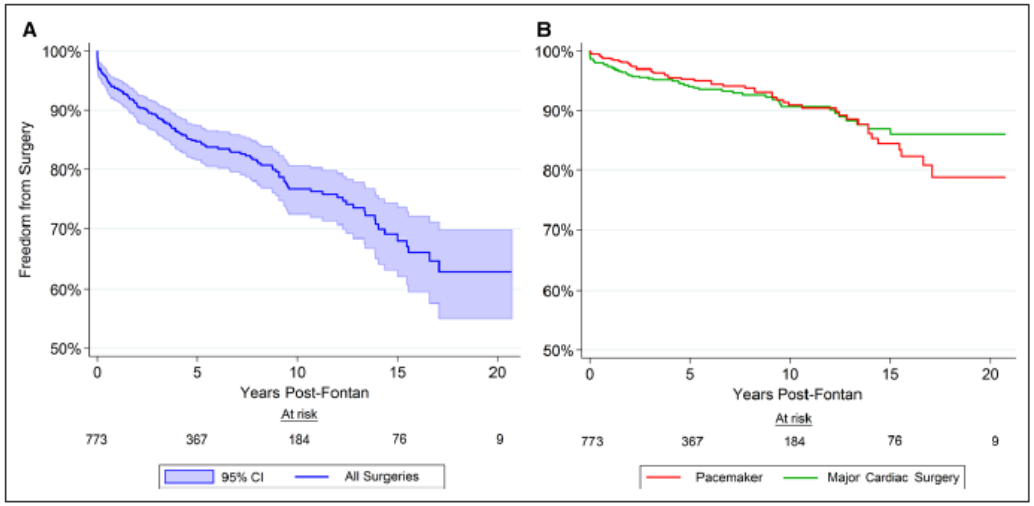



**Figure 2.** Freedom from surgical reintervention. **A**, Kaplan–Meier freedom from first surgical reintervention. Shaded area represents 95% confidence intervals (CIs) of the survival function. **B**, Kaplan–Meier freedom from first pacemaker and first major cardiac surgery in patients with intact Fontan. Y axis in both panels begins at 50%.

20 Year Freedom from Any Reintervention: 35%
Median time to reintervention: 9.8 years
Median number of interventions: 1 (0-7)

# 40-Year Follow-Up After the Fontan Operation

## Long-Term Outcomes of 1,052 Patients

Kavitha N. Pundi, MD,* Jonathan N. Johnson, MD,*† Joseph A. Dearani, MD,‡ Krishna N. Pundi, BS,§ Zhuo Li, BS,‖ Cynthia A. Hinck, RN, BSN,* Sonja H. Dahl, RN, DNP,* Bryan C. Cannon, MD,*† Patrick W. O'Leary, MD,*† David J. Driscoll, MD,* Frank Cetta, MD*†

(J Am Coll Cardiol 2012;60:1018–25)

**TABLE 6** Risk of Death or Cardiac Transplant

| | HR | 95% CI | p Value |
|---|---|---|---|
| Surgical era 1991–2000* | 0.63 | 0.44-0.90 | 0.01 |
| Surgical era 2001 onward* | 0.12 | 0.02-0.84 | 0.03 |
| Intraoperative sinus rhythm | 0.64 | 0.47-0.88 | 0.005 |
| Pre-operative mean catheter PAP >17 mm Hg | 1.42 | 1.15-1.77 | 0.001 |
| Asplenia | 1.55 | 1.07-2.25 | 0.02 |
| Pre-operative use of diuretics | 1.58 | 1.22-2.04 | <0.001 |
| Post-operative Fontan pressure >20 mm Hg | 2.29 | 1.72-3.05 | <0.001 |
| Post-operative LA pressure >13 mm Hg | 1.85 | 1.39-2.47 | <0.001 |
| Longer bypass time† | 1.12 | 1.05-1.20 | 0.001 |

*Compared with the surgical era from 1973 through 1990. †For every 30-min increase in overall bypass time.

Abbreviations as in Tables 2 and 3.

**TABLE 7** Risk of Death or Reoperation

| | HR | 95% CI | p Value |
|---|---|---|---|
| Bidirectional Glenn prior to Fontan | 0.41 | 0.24-0.70 | 0.001 |
| Intraoperative sinus rhythm | 0.69 | 0.52-0.94 | 0.02 |
| Pre-operative mean catheter PAP >17 mm Hg | 1.43 | 1.17-1.76 | <0.001 |
| Asplenia | 1.81 | 1.23-2.64 | 0.002 |
| Pre-operative use of diuretics | 1.77 | 1.39-2.26 | <0.001 |
| Atriopulmonary Fontan connection | 1.48 | 1.16-1.88 | 0.002 |
| AVV replacement at the time of Fontan | 2.42 | 1.39-4.22 | 0.002 |
| Post-operative Fontan pressure >20 mm Hg | 3.16 | 2.45-4.07 | <0.001 |
| Longer bypass time* | 1.08 | 1.009-1.16 | 0.03 |

*For every 30-min increase in overall bypass time.

Abbreviations as in Tables 2 and 3.

# Longitudinal Outcomes of Patients With Single Ventricle After the Fontan Procedure

Andrew M. Atz, MD,[a] Victor Zak, PhD,[b] Lynn Mahony, MD,[c] Karen Uzark, PhD,[d] Nicholas D'agincourt, MS,[b] David J. Goldberg, MD,[e] Richard V. Williams, MD,[f] Roger E. Breitbart, MD,[g] Steven D. Colan, MD,[g] Kristin M. Burns, MD,[h] Renee Margossian, MD,[g] Heather T. Henderson, MD,[i] Rosalind Korsin, RN,[j] Bradley S. Marino, MD,[k] Kaitlyn Daniels, RN,[f] Brian W. McCrindle, MD, MPH,[l] for the Pediatric Heart Network Investigators

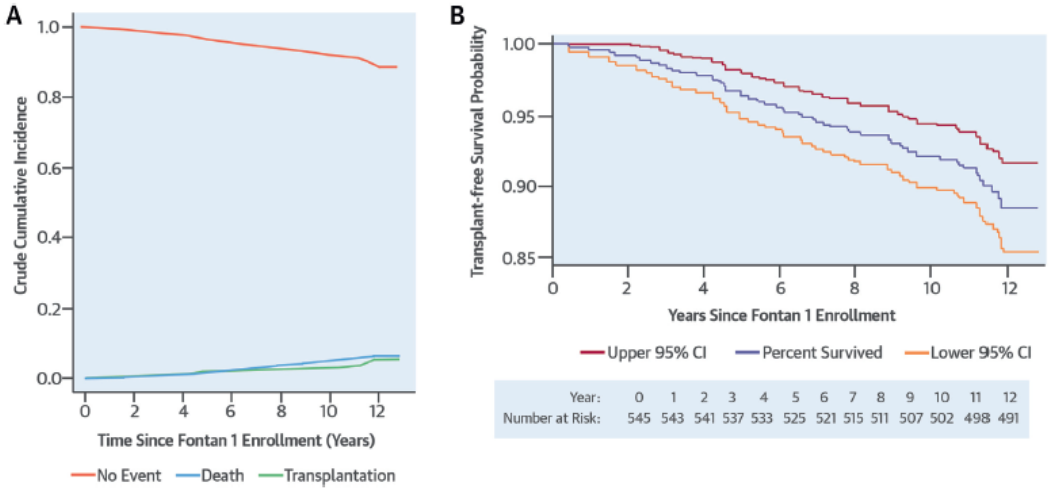
(J Am Coll Cardiol 2017;69:2735–44)

**TABLE 1 Complications and Interventions After Fontan Procedures**

| | Fontan 1 (N = 546) | Fontan 2 (N = 416) | Fontan 3 (N = 373) |
|---|---|---|---|
| Time since Fontan procedure, yrs | 8.7 ± 3.4 | 15.2 ± 3.4 | 17.8 ± 3.4 |
| Cardiac surgery | 23 | 28 | 32 |
| Catheter intervention | 48 | 57 | 62 |
| Electronic device | 13 | 13 | 16 |
| Stroke | 2 | 2 | 4 |
| Seizures | 3 | 5 | 7 |
| Thrombosis | 8 | 9 | 12 |
| Arrhythmia treatment | 20 | 28 | 32 |
| Protein-losing enteropathy | 4 | 7 | 8 |
| Cirrhosis | 0.4 | 4 | 8 |
| Plastic bronchitis | 0.1 | 0.5 | 1 |

Values are mean ± SD or %.

**CENTRAL ILLUSTRATION Survival of Single-Ventricle Patients After the Fontan Procedure**




Atz, A.M. et al. J Am Coll Cardiol. 2017;69(22):2735–44.

**(A)** Cardiac transplant-free survival. Proportion of subjects in each of 3 competing mutually exclusive states: death, cardiac transplantation, and alive without transplantation for all 545 subjects. **(B)** Transplant-free survival since Fontan 1 (with 95% confidence intervals) in all subjects with complete follow-up data. For improved resolution, the scale on the y-axis is limited to 0.85 to 1.0.

# Therapy: Fontan-Kreutzer Failure

Early

- Surgical
  - **Take Down**
  - Valve Repair and Replacement
  - Conversion
  - Transplant
    - Multiorgan

# Early Fontan-Kreutzer Failure

DEFINITION: During same admission as Fontan procedure

Manifestations:

- Low cardiac output
- Hypoxemia
- Pathway obstruction/thrombosis
- End organ dysfunction (liver, kidneys)
- Pleural effusions
- Arrhythmias

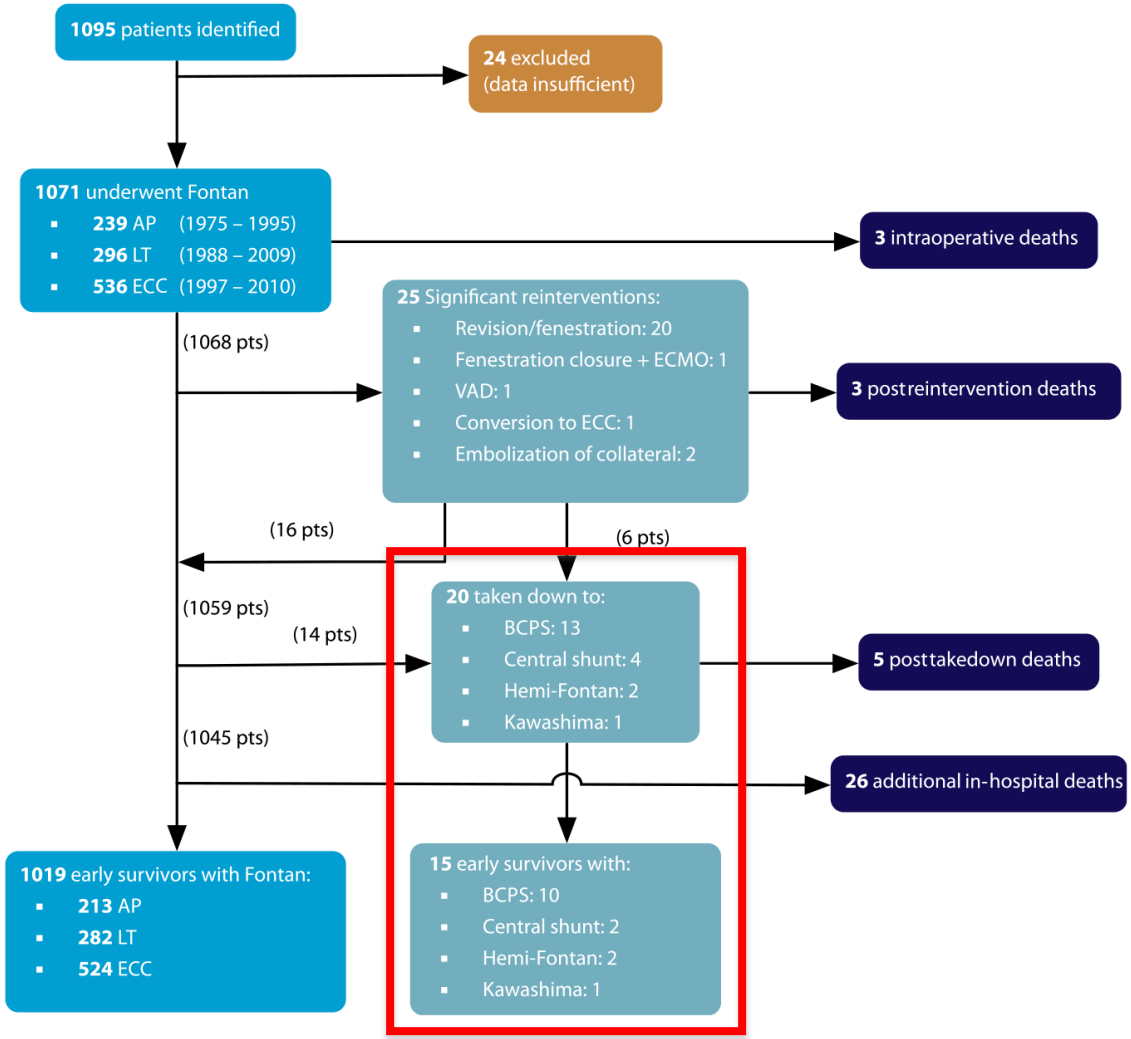
Options:

- Medical management
- Revision (transcatheter or surgical)
- ECMO
- Takedown

# Trends in Fontan surgery and risk factors for early adverse outcomes after Fontan surgery: The Australia and New Zealand Fontan Registry experience

Ajay J. Iyengar, MBBS(Hons), BMedSci,[a,b] David S. Winlaw, MBBS(Hons), MD, FRACS,[c]
John C. Galati, BSc, PhD,[b,j] David S. Celermajer, MBBS, PhD, DSc, MSc, FAHA, FRACP, FAA,[d]
Gavin R. Wheaton, MBBS, FRACP,[e] Thomas L. Gentles, MBChB, FRACP,[f] Leeanne E. Grigg, MBBS, FRACP,[g]
Robert G. Weintraub, MBBS, FRACP,[a,b] Andrew Bullock, MBBS, FRACP,[h]
Robert N. Justo, MBBS, FRACP,[i] and Yves d'Udekem, MD, PhD[a,b]

(J Thorac Cardiovasc Surg 2014;148:566-75)



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(J Thorac Cardiovasc Surg 2014;148:566-75)

**TABLE 3. Continued**

| | Early mortality (n = 37) | | | | Early failure (n = 65) | | | | Prolonged effusions (n = 75) | | | | Composite adverse early outcome (n = 165) | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | Univariate | Multivariate | | | Univariate | Multivariate | | | Univariate | Multivariate | | | Univariate | Multivariate | | |
| Variable | *P* value | OR | 95% CI | *P* value | *P* value | OR | 95% CI | *P* value | *P* value | OR | 95% CI | *P* value | *P* value | OR | 95% CI | *P* value |
| BCPS | <.001 | —§ | | | .003 | —§ | | | .09 | | | | <.001 | —§ | | |
| Previous or concomitant PA reconstruction | .8 | | | | .6 | | | | .2 | | | | .9 | | | |
| Pre-Fontan hemodynamics‖ | | | | | | | | | | | | | | | | |
| Arterial oxygen saturation, (per %; n = 839) | .2 | | | | .1 | | | | .3 | | | | .1 | | | |
| PA pressure (per 5 mm Hg; n = 869) | .01 | 1.5 | 1.0-2.4 | .06 | .003 | 1.4 | 1.0-2.1 | .08 | .4 | | | | .03 | 1.3 | 0.9-1.8 | .1 |
| AV valve regurgitation (n = 871) | —† | | | | .08 | 0.4 | 0.04-3.0 | .3 | .5 | | | | .6 | | | |
| Arteriovenous malformations (n = 809) | .1 | | | | .08 | 2.1 | 0.6-7.2 | .3 | .2 | | | | .03 | 2.6 | 1.2-5.8 | .02 |
| Collaterals (n = 799) | .6 | | | | .08 | | | | .09 | | | | .5 | | | |
| Operative variables | | | | | | | | | | | | | | | | |
| Concomitant procedure | .1 | 2.2 | 0.8-6.0 | 0.1 | .2 | | | | .007 | 2.2 | 1.2-3.8 | .008 | .01 | 1.9 | 1.1-3.3 | .02 |
| Fenestration | .007 | —§ | | | .2 | | | | .1 | | | | .04 | —§ | | |
| Postoperative effusions (mortality and failure end points only) | .3 | | | | <.001 | 14.5 | 6.5-32.5 | <.001 | — | | | | — | | | |
| RSV seasonality (May-Sept prolonged effusions end point only) | — | | | | — | | | | .8 | | | | — | | | |

Regression parameters are presented for covariates entered into multivariate models. *OR*, Odds ratio; *CI*, confidence interval; *Ref*, reference; *BiV*, biventricular; *TA*, tricuspid

## 18 Years of the Fontan Operation at a Single Institution

### Results From 771 Consecutive Patients

Lindsay S. Rogers, MD,*† Andrew C. Glatz, MD,*† Chitra Ravishankar, MD,*†
Thomas L. Spray, MD,*‡ Susan C. Nicolson, MD,*§ Jack Rychik, MD,*†
Christina Hayden Rush, BSN,* J. William Gaynor, MD,*‡ David J. Goldberg, MD*†
*Philadelphia, Pennsylvania*

(J Am Coll Cardiol 2012;60:1018–25)

**Table 9** Summary of Contemporary Reports of Early Postoperative Outcomes

| | Cohort Years | n | Mortality | Takedown | Effusion (days) | Length of Stay (days) | CPB Time (min) |
|---|---|---|---|---|---|---|---|
| O'Brien et al. (7) | 1997–2008 | 145 | 5.5% | 2.8% | 12 | 8 | 82 (63–102) |
| Hirsch et al. (9) | 1992–2007 | 636 | 4.0% | 3.0% | | 10 | 66 (55–93) |
| Tweddell et al. (8) | 1994–2007 | 256 | 2.0% | 0.8% | 5 | 9 | 110 (89–129) |
| Salvin et al. (10) | 2001–2005 | 226 | 0.9% | 0.9% | 6 | | 107.9 ± 46.9 |
| Petrossian et al. (11) | 1992–2005 | 285 | 1.1% | 1.4% | 8 | 11 | |

# Management of Early Fontan Failure: A Single Institution Experience

Michael O. Murphy, Andrew C. Glatz, David J. Goldberg, Lindsay S. Rogers, Chitra Ravishankar, Susan C. Nicolson, James M. Steven, Stephanie Fuller, Thomas L. Spray and J. William Gaynor

- Single center cohort study
- July 1st 1995 to December 31st 2009
- Early Fontan Failure was defined as:
  - –early post-operative death
  - –need for Fontan takedown
  - –need for ECMO
  - –emergent transplant

592 patients

Pre-Op Cath data in 83.4%

Prior SCPC 96.8%

Fenestration 91%

MUF 95.9%

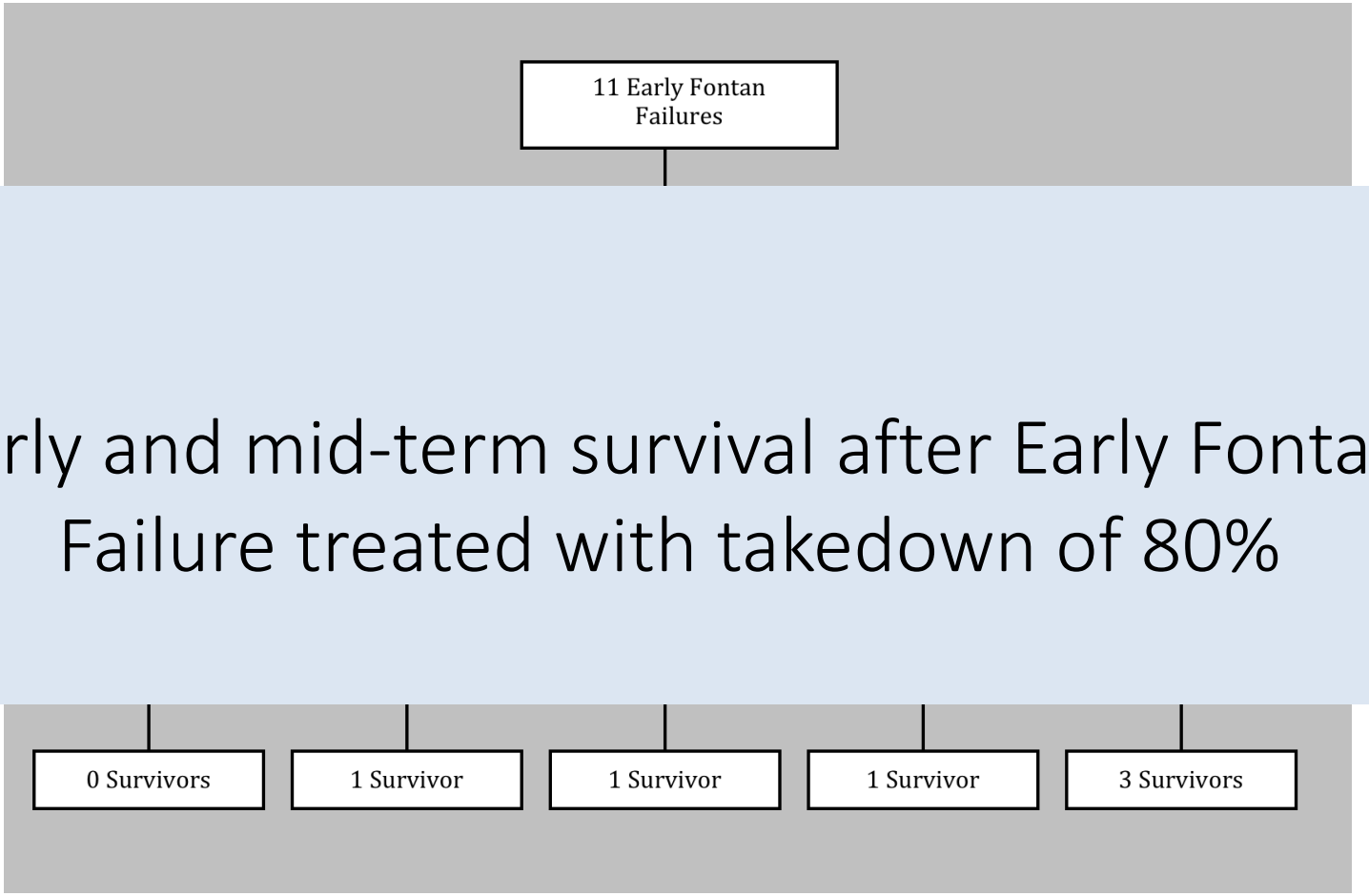
DHCA 97.5%

ECC 60% vs LT 40%

Failure: 11 patients (1.9%)

European Journal of Cardio-Thoracic Surgery 46 (2014) 458–464

# Results




Early and mid-term survival after Early Fontan Failure treated with takedown of 80%

European Journal of Cardio-Thoracic Surgery 46 (2014) 458–464

# Results

| PATIENT CHARACTERISTICS | | | | | |
|---|---|---|---|---|---|
| | Fontan Failure N=11 | | No Fontan Failure N=581 | | P value |
| Heterotaxy | 1 | 9.09% | 58 | 9.98% | 0.922 |
| Common AVV | 2 | 18.18% | 76 | 13.08% | 0.622 |
| AVV regurgitation >mild | 2 | 18.185 | 78 | 14.86% | 0.760 |
| HLHS | 4 | 36.36% | 265 | 46.33% | 0.514 |
| Dominant RV | 7 | 63.64% | 386 | 67.48% | 0.788 |
| No prior SCPC | 3 | 27.27% | 15 | 2.58% | **<0.001*** |
| | | | | | |
| | Mean/median | SD or range | Mean/median | SD or range | P value |
| Age at Stage II (months) | 7.1 | 4.7-14.8 | 6.2 | 1.9-177.6 | 0.934 |
| Age at Fontan (years) | 2.06 | 1.31-16.84 | 2.47 | 1-38.42 | 0.470 |
| Weight at Fontan (kg) | 10.8 | 10-43 | 12.4 | 6.7-59.2 | 0.418 |
| Stage II to Fontan (mths) | 18.9 | 13.3-46.6 | 23.5 | 5.2-155.9 | 0.524 |
| PAP | 12.3 | 3.1 | 11.2 | 3 | 0.258 |
| VEDP | 9.5 | 3.3 | 7.4 | 2.7 | **0.019*** |

European Journal of Cardio-Thoracic Surgery 46 (2014) 458–464

# Results

| SURGICAL CHARACTERISTICS | | | | | |
|---|---|---|---|---|---|
| | Fontan Failure N=11 | | No Fontan Failure N=581 | | P value |
| Fenestration | 9 | 81.82% | 530 | 91.22% | 0.293 |
| Additional procedures | 2 | 18.18% | 103 | 17.73% | 0.969 |
| Arch augmentation | 0 | 0.00% | 2 | 0.34% | 0.413 |
| AVV repair | 1 | 9.09% | 30 | 3.44% | 0.351 |
| Extra-Cardiac Conduit | 11 | 100.00% | 347 | 59.72% | **<0.001*** |
| | | | | | |
| | Mean/median | SD or range | Mean/median | SD or range | P value |
| TS (minutes) | 89 | 70-185 | 65 | 32-274 | **0.001*** |
| CPB (minutes) | 60.5 | 39-126 | 40 | 11-242 | **0.003*** |
| XC (minutes) | 39 | 18-99 | 25 | 0-96 | **<0.001*** |
| DHCA (minutes) | 33 | 0-59 | 23 | 0-80 | 0.255 |

| OUTCOMES | | | | | |
|---|---|---|---|---|---|
| | Mean/median | SD or range | Mean/median | SD or range | P value |
| ICU Duration (days) | 8 | 4-28 | 2 | 0-56 | **<0.001*** |
| Effusion Duration (days) | 15 | 6-50 | 2 | 1-71 | **<0.001*** |
| Hospital Duration (days) | 17 | 4-108 | 7 | 2-71 | **<0.001*** |

European Journal of Cardio-Thoracic Surgery 46 (2014) 458–464

# Therapy: Fontan-Kreutzer Failure

Chronic

- Surgical
  - Take Down
  - Valve Repair and Replacement
  - **Conversion**
  - **Transplant**
    - **Multiorgan**

# Management of the failing Fontan circulation

Barbara J Deal,[1] Marshall L Jacobs[2]

*Heart* 2012;**98**:1098–1104.

**Table 1** Failing Fontan circulation

| | | |
|---|---|---|
| Constitutional | Growth failure | Inadequate cardiac output |
| | Exercise intolerance | Chronotropic incompetence, pronounced atrial distension |
| | Depression | Secondary to limitations on functional status |
| Haemodynamic | Obstruction | Atriopulmonary, pulmonary arterial |
| | Systemic venous | Pulmonary venous return, atrioventricular (AV) valve inflow |
| | Systemic outflow | Ventricular outflow, aortic arch |
| | AV valve function | ≥Moderate regurgitation<br>≥Mild stenosis |
| | Ventricular dysfunction | Secondary: atrial dilatation/distortion; AV valve or semilunar valve dysfunction, chronic arrhythmias or antiarrhythmic medications; impaired myocardial perfusion due to coronary sinus hypertension |
| | Thrombosis | Systemic venous, atrial, pulmonary |
| Rhythm | Arrhythmias | Sinus node dysfunction, predominant junctional rhythm, AV block, supraventricular tachycardia/atrial tachycardia, ventricular tachycardia |
| Pulmonary | Cyanosis | Intracardiac right to left shunt, veno-venous collaterals, pulmonary arteriovenous malformations (AVMs) |
| | Pleural effusions | |
| Gastrointestinal | Ascites | Secondary to portal hypertension related to obstruction, versus cirrhosis |
| Metabolic | Metabolic markers | Declining albumin; thrombocytopenia; hyperbilirubinaemia; coagulopathy |

# Indications for Surgical Revision

- Residual Shunt
- Systemic Valve Regurgitation
  - Neoaortic or Atrioventricular
- Conduit Obstruction
- Significant Atrial Flutter or Fibrillation
- Pulmonary Venous Obstruction
- Aortic Root Dilation

Avoid in patients with severe ventricular dysfunction, PLE, plastic bronchitis

# Fontan-Kreutzer Conversion

- Atrial Septectomy
- Biatrial Maze
- Right Atrial Resection
- Extracardiac Fontan
- Dual Chamber Epicardial Pacemaker

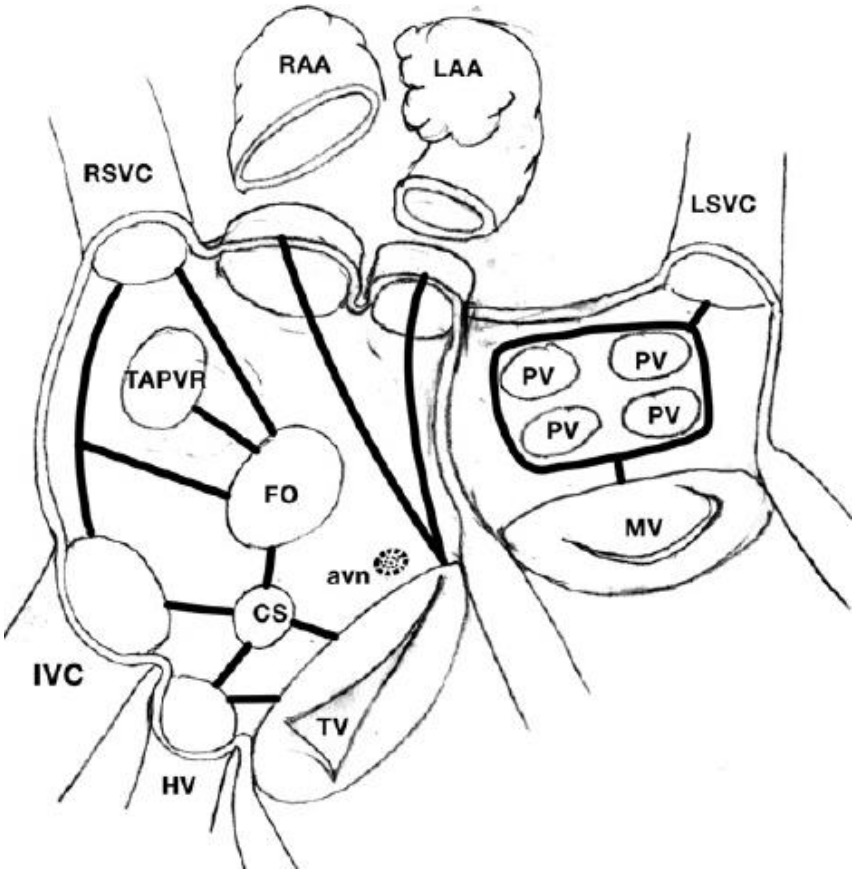



**Figure 3** Surgical modifications of atrial maze procedure in complex anatomy. Solid black lines indicate sites of cryoablation. avn, atrioventricular node; CS, coronary sinus; FO, foramen ovale; HV, hepatic vein; IVC, inferior vena cava; LAA, left atrial appendage; LSVC, left superior vena cava; MV, mitral valve; PV, pulmonic vein; RAA, right atrial appendage; RSVC, right superior vena cava; TAPVR, total anomalous pulmonary venous return; TV, tricuspid valve. Reprinted with permission from Mavroudis C, Deal BJ, Backer CL, Tsao S. Arrhythmia surgery in patients with and without congenital heart disease. *Ann Thorac Surg* 2008;**86**:857–68.

# Rescuing the failing Fontan

Carl L Backer

Backer CL. *Heart* July 2016 Vol 102 No 14

**Table 1** Results of surgical intervention (rescue) for the failing Fontan

| Procedure | n | Early mortality (%) | Late mortality or transplant (%) |
|---|---|---|---|
| Fontan takedown | 38 | 26 | 45 |
| Fontan conversion | 137 | 11 | 26 |
| Heart transplant | 50 | 20 | 34 |

**Table 1** Patients undergoing Fontan conversion at Ann & Robert H. Lurie Children's Hospital from 1994 to 2012 (n = 140)

| | |
|---|---|
| Median age at initial Fontan | 5.6 yrs (range, 1.1 – 34.9) |
| Median age at Fontan conversion | 23.2 yrs (range, 2.6 – 47.3) |
| Interval from initial Fontan to conversion | 16.7 yrs (range, 1.2 – 33.6) |
| Right atrial tachycardia | 48 patients |
| Left atrial tachycardia | 21 patients |
| Atrial fibrillation | 67 patients |

Adapted and reprinted with permission from Deal et al; Ann Thorac Surg 2016;101:717-724.[2]

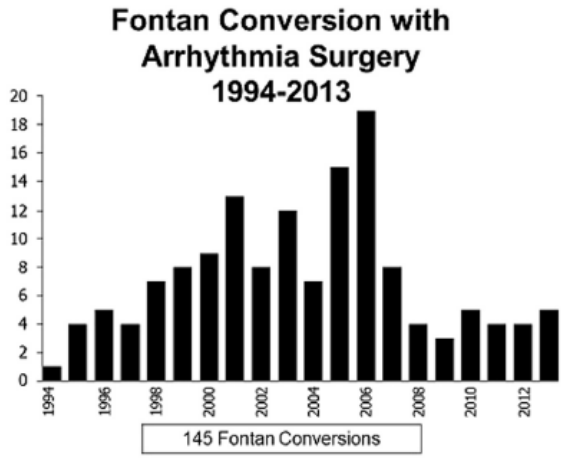



**Figure 8** Fontan conversion with arrhythmia surgery at Ann & Robert H. Lurie Children's Hospital 1994–2013 (145 Fontan conversions).

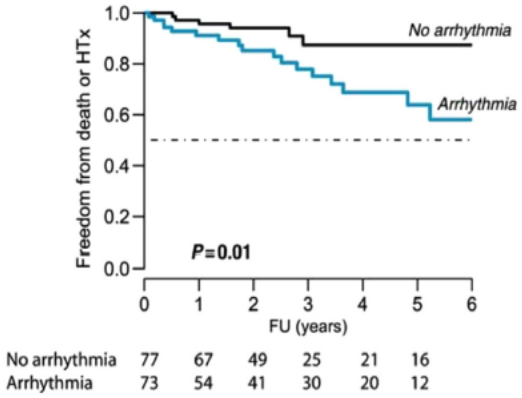



**Table 2** Independent Risk Factors for Cardiac Death or Transplantation after Fontan Conversion

| Variable | Adjusted hazard ratio (95% CI) | *P* value |
|---|---|---|
| Right or indeterminate ventricular morphology | 5.71 (2.37 – 13.75) | <.001 |
| Ascites | 3.69 (1.59 – 8.56) | .002 |
| Protein-losing enteropathy | 4.93 (1.16 – 20.98) | .03 |
| Cardiopulmonary bypass time > 240 min | 2.68 (1.20 – 6.00) | .02 |
| Biatrial arrhythmia operation | 6.17 (1.84 – 20.71) | .003 |

Abbreviation: CI, confidence interval.
Adapted and reprinted with permission from Deal et al; Ann Thorac Surg 2016;101:717-724.[2]

# Surgical options after Fontan failure

Joost P van Melle,[1] Djoeke Wolff,[2] Jürgen Hörer,[3] Emre Belli,[4] Bart Meyns,[5] Massimo Padalino,[6] Harald Lindberg,[7] Jeffrey P Jacobs,[8,9] Ilkka P Mattila,[10] Håkan Berggren,[11] Rolf M F Berger,[2] Rene Prêtre,[12] Mark G Hazekamp,[13,14] Morten Helvind,[15] Matej Nosál,[16] Tomas Tlaskal,[17] Jean Rubay,[18] Stojan Lazarov,[19] Alexander Kadner,[20] Viktor Hraska,[21] José Fragata,[22] Marco Pozzi,[23] George Sarris,[24,25] Guido Michielon,[26] Duccio di Carlo,[27] Tjark Ebels[26]

van Melle JP, *et al*. *Heart* 2016;**102**:1127–1133. doi:10.1136/heartjnl-2015-309235

**Table 3** Indications for failing Fontan surgery

| Indication | Number (%) |
|---|---|
| Arrhythmia | 98 (43.6) |
| Deteriorating functional class | 97 (43.1) |
| Extreme RA dilatation | 87 (38.7) |
| Obstruction | 47 (20.9) |
| Protein losing enteropathy | 31 (13.8) |
| Thrombus | 26 (11.6) |
| AVV regurgitation | 18 (8.0) |
| Pulmonary vein stenosis | 11 (4.9) |
| Cyanosis | 12 (5.3) |
| Systemic obstruction | 4 (1.8) |
| Baffle leak | 4 (1.8) |

AVV, atrioventricular valve; RA, right atrial.

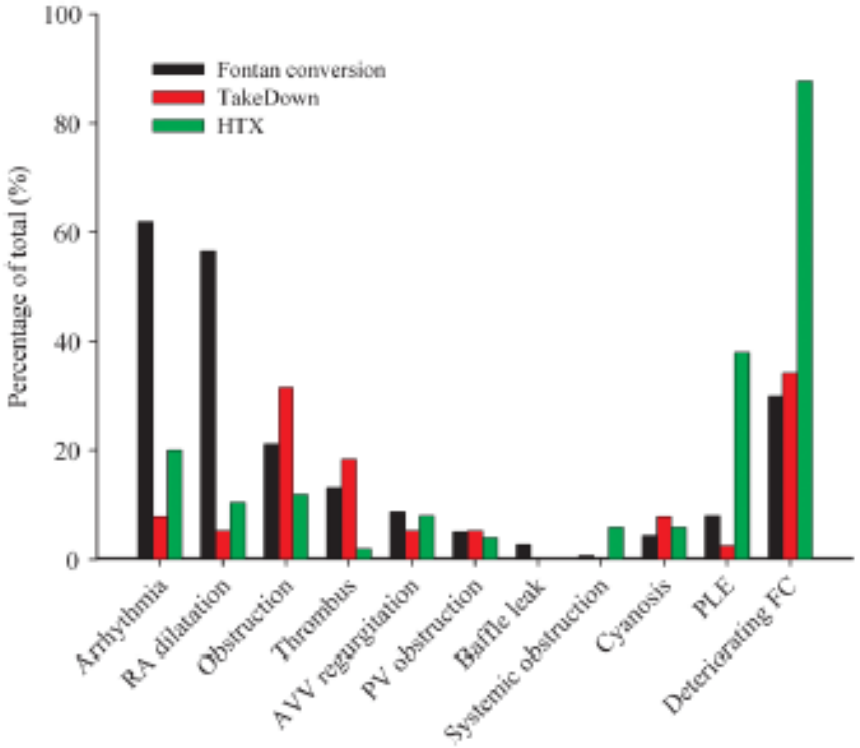



**Figure 2** Indications for failing Fontan surgery. More than one indication is possible within the same patient. RA, right atrial; AVV, atrioventricular valve; PV, pulmonary vein; PLE, protein losing enteropathy; FC, functional class.

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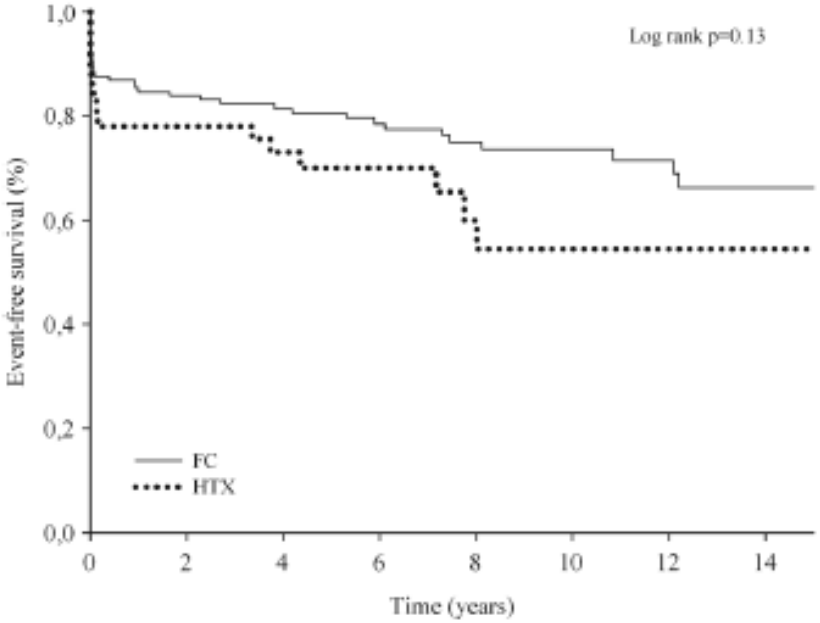



| No at risk | 0 | 5 yrs | 10 yrs | 15 yrs |
|---|---|---|---|---|
| Fontan conversion | 137 | 84 | 41 | 13 |
| HTX | 50 | 23 | 9 | 5 |

**Figure 4** Kaplan–Meier plot showing the event-free survival after heart transplantation (HTX) (n=50) or Fontan conversion (n=137). Time of follow-up is truncated at 15 years. End points are mortality or (re-)HTX. FC, functional class; yrs, years.

# Meta-Analysis of the Effectiveness of Heart Transplantation in Patients With a Failing Fontan

Nazlee Tabarsi, MD[a], Meijiao Guan, PhD[a], Jacob Simmonds, MA, MB[b], Mustafa Toma, MD[a], Marla Kiess, MD[a], Victor Tsang, MBBS[b], Peter Ruygrok, MD[c], Igor Konstantinov, MD, PhD[d], William Shi, MBBS[d], and Jasmine Grewal, MD[a,*]

Am J Cardiol 2017;119:1269–1274

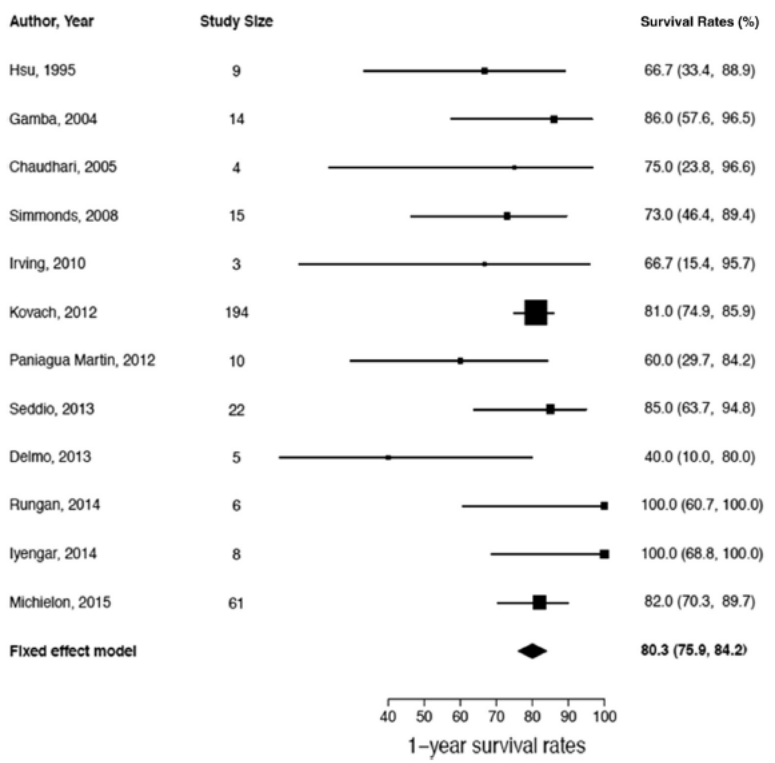



Figure 2. Forest plot of 1-year survival rates using fixed effects model. Studies are arranged by year of publication.

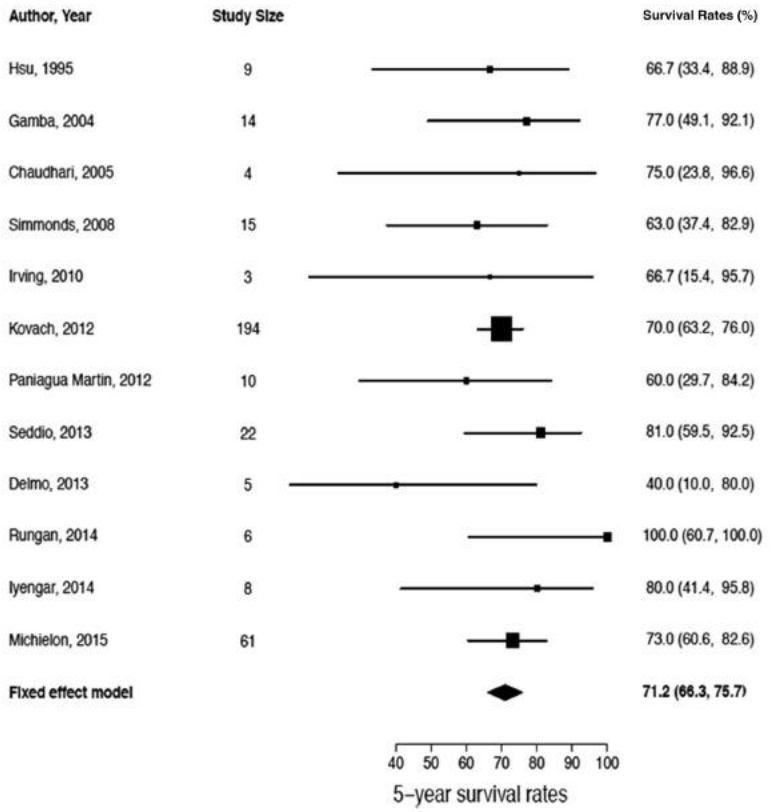



Figure 3. Forest plot of 5-year survival rates using fixed effects model. Studies are arranged by year of publication.

# Mortality Risk Stratification in Fontan Patients Who Underwent Heart Transplantation

Christopher J. Berg, MS[a], Brenton S. Bauer, MD[a,b,c], Abbie Hageman, BS[a,d], Jamil A. Aboulhosn, MD[a,c,d], and Leigh C. Reardon, MD[a,c,d,e,*]

Am J Cardiol 2017;119:1675−1679

Table 3
Mortality risk post-transplant by patient characteristic

| Variable | Hazard Ratio (95% CI) | P value* |
|---|---|---|
| Age <18 at time of transplantation (N=16) | 3.88 (1.36 - 11.05) | **0.011** |
| Female (N=14) | 0.58 (0.21 - 1.66) | 0.31 |
| Transplantation prior to 2004 (N=17) | 4.18 (1.35 - 12.90) | **0.013** |
| Fontan-transplantation interval <10 years (N=15) | 4.38 (1.59 - 12.04) | **0.004** |
| Left morphological ventricle (N=24) | 1.80 (0.58 - 5.56) | 0.307 |
| Number of previous cardiac surgeries | | 0.641 |
| >3 previous surgeries | 0.58 (0.17 − 2.05) | 0.401 |

* Bold values represent significance at p <0.05.

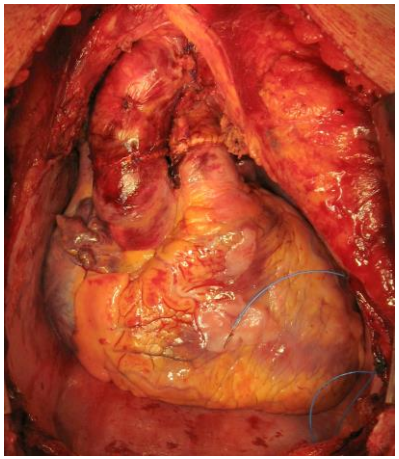
Table 4
Preoperative risk factors for postoperative mortality

| Preoperative risk factor | N | Hazard Ratio (95% CI) | P value* |
|---|---|---|---|
| Systemic ventricle ejection fraction <20% | 12 | 3.40 (1.14 - 10.19) | **0.029** |
| Moderate or severe atrioventricular valve regurgitation | 20 | 8.12 (1.05 - 62.50) | **0.044** |
| Average Fontan circuit pressure (mmHg) | 28 | | 0.330 |
| Fontan pressure >16.5mmHg | 10 | 0.21 (0.05 - 0.84) | **0.027** |
| Transpulmonary gradient (mmHg) | 28 | | 0.160 |
| Comorbidities | | | |
| Protein losing enteropathy | 6 | 1.38 (0.40 - 4.83) | 0.611 |
| Chronic renal insufficiency | 11 | 0.37 (0.10 - 1.29) | 0.118 |
| Cirrhosis | 6 | 0.81 (0.18 - 3.61) | 0.785 |
| Ascites | 7 | 0.21 (0.03 - 1.61) | 0.135 |
| Advanced mechanical support | | | |
| Renal replacement therapy | 2 | 71.29 (5.96 - 852.40) | **<0.001** |
| Extra-corporal membrane oxygenation | 4 | 6.25 (1.94 - 20.20) | **0.002** |
| Serum markers | | | |
| Creatinine (mg/dL) | 32 | | 0.493 |
| Total bilirubin (mg/dL) | 27 | | 0.081 |
| Model of End-Stage Liver Disease, excluding INR score | 27 | 1.17 (1.04 - 1.31) | **0.007** |
| Score ≥19 | 7 | 4.91 (1.40 - 17.27) | **0.013** |
| Cardiopulmonary bypass time (mins) | 28 | | 0.228 |
| Donor ischemic time (mins) | 26 | | 0.367 |

# Operative Strategy

Median Sternotomy

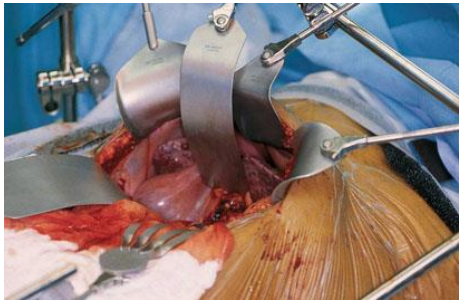
Limited Laparotomy – Liver Exam



Orthotopic Heart Transplant on CPB

*Wean from CPB and turn SVC cannula into right atrium*

Orthotopic Liver Transplant – bilateral subcostal with midline extension



*Reverse heparin*

Close chest and abdomen

# Intermediate-term outcomes after combined heart–liver transplantation in children with a univentricular heart

Seth A. Hollander, MD,[a] Olaf Reinhartz, MD,[b] Katsuhide Maeda, MD,[b] Melissa Hurwitz, MD,[c] David N. Rosenthal, MD,[a] and Daniel Bernstein, MD[a]

J Heart Lung Transplant 2013;32:368–370

Since 2007 – 47 Heart Liver Transplants, only 3 pediatric

3 patients at 2, 3, 5 years post transplant
- 100% one-year survival

| Age/Gender | Diagnosis | Time from Palliation - Transplant |
|---|---|---|
| 17 Female | DILV, TA, Straddling MV, TGA | 13 years |
| 14 Female | Heterotaxy, Unbalanced AV Canal, Interrupted IVC | 9 years |
| 7 Male | VACTERL, DORV, PS, VSD, PAPVR | 7 years |

# Heart and heart–liver transplantation in adults with failing Fontan physiology

Leigh C. Reardon[1,2] | Eugene C. DePasquale[3] | Jana Tarabay[1] | Daniel Cruz[3] | Hillel Laks[4] | Reshma M. Biniwale[4] | Ronald W. Busuttil[5] | Fady M. Kaldas[5] | Sammy Saab[6] | Robert S. Venick[7] | Jeannette P. Lin[1] | Ali Nsair[3] | Mario C. Deng[3] | Abbas Ardehali[4] | Martin Caderias[3] | Amit Iygengar[8] | Jamil A. Aboulhosn[1]

**Conclusions:** Despite inherent risks and complexities of OHT or CHLT in patients with a failed Fontan, transplant is a reasonable therapy. Peri- and postoperative complications are common and may require surgical reintervention. Continued observation of practices and unifying themes may help improve patient selection, pre- and postoperative treatment and ultimately outcomes.

**Table 2.** Patient clinical characteristics (n = 20 unless otherwise specified)

| Clinical characteristics | |
|---|---|
| Number of cardiac surgeries, Median (range) | 3 (2-6) |
| Age at Fontan, years median (range) | 5.5 (3- 22) |
| Number of CHLT, No. (%) | 5 (25%) |
| Complex venous reconstruction, No. (%) | 8 (40%) |
| EF%, Median (range) (n = 19) | 35 (15-65) |
| Resting saturation, % median (range) | 90 (67-97) |
| Number of cyanotic patients (<90%), No. (%) | 9 (45%) |
| Fontan pressure, mmHg median (range) (n = 17) | 18 (15-23) |
| Single ventricular EDP, mmHg median (range) (n = 12) | 12.5 (9-15) |
| BNP, pg/mL, Median (range) (n = 18) | 394 (27-2710) |
| Creatinine, mg/dL, Median(range) (n = 18) | 1.2 (0.5-2) |
| Albumin, g/dL, Median (range) (n = 18) | 4 (2.5-5) |
| Total bilirubin, mg/dL, Median (range) (n = 18) | 1.65 (0.3-4.7) |
| MELD-XI, median, (range) (n = 17) | 13.5 (9.4-22.9) |
| Cardiopulmonary bypass time, median (range) (n = 14) | 181 (108-465) |
| Days to discharge post-OHT, Median (range) | 23 (8-76) |
| Days to discharge post-CHLT, Median (range) | 51 (26-77) |
| Bleeding | 11 (55%) |
| Hematoma requiring evacuation | 2 (10%) |
| Bleeding requiring chest exploration | 2 (10%) |
| Cardiac tamponade requiring window | 4 (20%) |
| Extensive bleeding | 2 (10%) |
| Pleural effusions requiring thoracentesis | 1 (10) |
| Multi-organ failure | 0 (0%) |
| Stroke | 0 (0%) |
| Infection | 4 (20%) |
| Extracorporeal membrane oxygenation after OHT | 1 (5%) |
| Acute renal failure requiring hemodialysis | 4 (20%) |
| Plasmapheresis/Intravenous Immunoglobulins (IVIG) | 5 (25%) |
| Delayed chest closure | 5 (25%) |
| Acute rejection | |
| Acute Cellular Rejection >1R | 0 (0%) |
| Acute Cellular Rejection ≤Grade 1R, mild | 2 (10%) |
| Antibody Mediated Rejection >1 | 0 (0%) |
| Antibody Mediated Rejection ≤Grade 1, mild | 1 (5%) |
| Arrhythmias | 4 (20%) |
| Ventricular tachycardia | 2 (10%) |
| Atrial fibrillation | 1 (5%) |
| Atrial flutter | 1 (5%) |

# Single-center outcomes of combined heart and liver transplantation in the failing Fontan

Benjamin A. D'Souza[1] | Stephanie Fuller[2] | Lacey P. Gleason[2] | Nicole Hornsby[1] | Joyce Wald[1] | Karen Krok[3] | Abraham Shaked[4] | Lee R. Goldberg[1] | Alberto Pochettino[5] | Kim M. Olthoff[4] | Yuli Y. Kim[1]

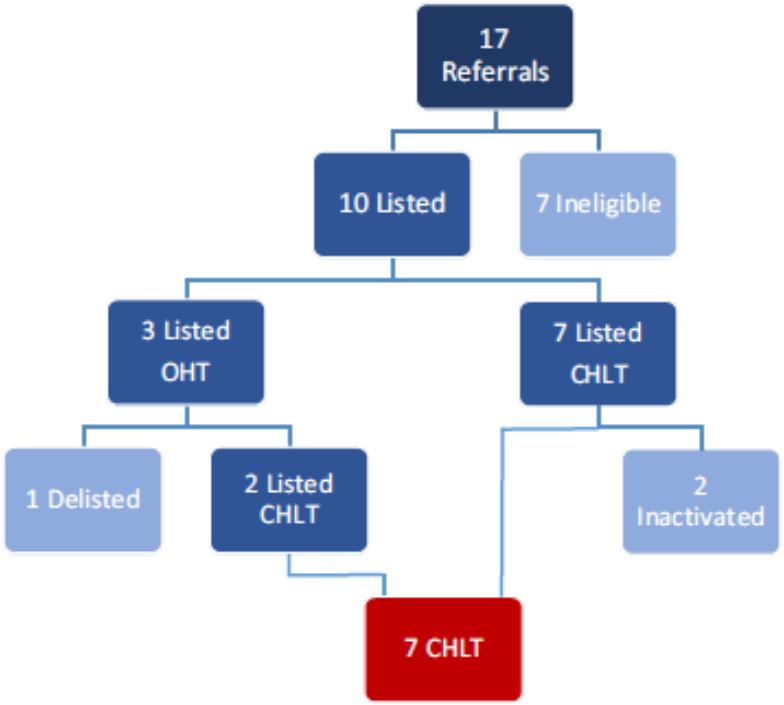



**FIGURE 1** Results of cardiac transplant referrals for Fontan patients between 2000 and 2013. CHLT, combined heart-liver transplantation; OHT, orthotopic heart transplantation

Indication for referral:

7/17 PLE (43%)

57% inotrope dependent

All had cirrhosis by MRI or CT and two had biopsy proven cirrhosis

Median wait 0.7y (0.1-3.5)

Median CPB 218" XC 176"

Median ischemic time 211"

Median LOS 29 days

RESULTS:

AKI 4/7 (57%) with dialysis in 2

No mortality at 4.6 years

1/7: liver rejection treated with steroids

NOW 10 Patients

# Conclusion

- Fontan-Kreutzer operation achieves excellent results
- Survival excellent when early failure detected and addressed with takedown
- Mechanical support exists for the chronic patient with failing Fontan-Kreutzer and results are encouraging
- Transplant remains challenging
  - Organ shortage
  - Comorbidities
- Heart liver transplantation is a viable option for patients with Fontan-Kreutzer association liver disease
- Research efforts need to focus on what constitutes a good adult Fontan-Kreutzer and we need to reproduce this physiology