

대한심장학회 제55차 추계학술대회
Mini-Symposium: Long-Term After Fontan Operation

Reoperations After the Fontan Procedure

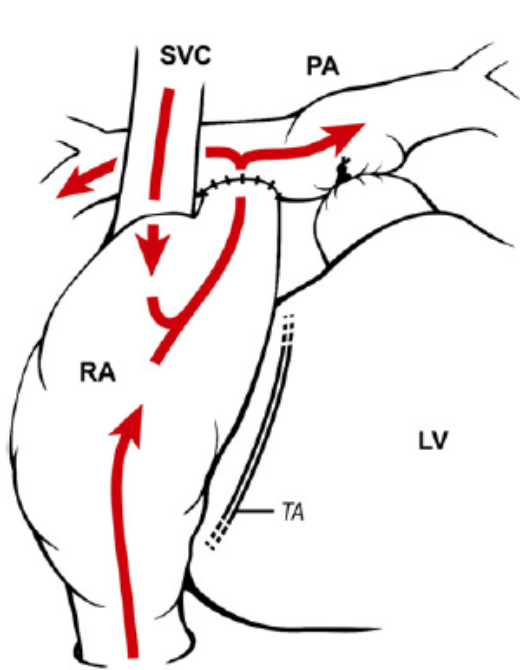
Cheul Lee, MD

**Department of Thoracic and Cardiovascular Surgery
Sejong General Hospital**

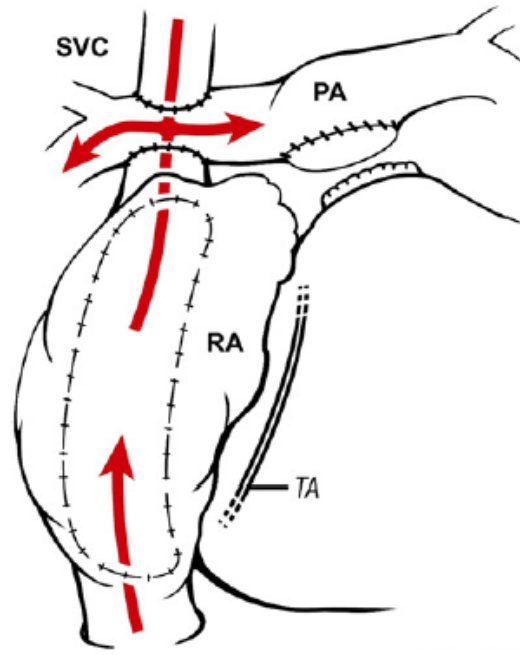
Contents

- 1. Fontan conversion**
- 2. AV valve regurgitation**
- 3. Pulmonary AV fistula**
- 4. Conduit-related problems**

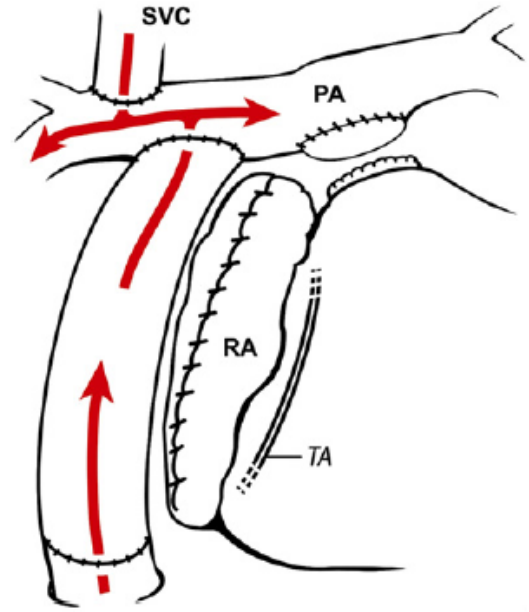
Evolution of the Fontan Procedure



AP Fontan
1971



Lateral Tunnel Fontan
1988



Extracardiac Fontan
1990

Fontan Conversion

Long-term sequelae of the AP Fontan

- * Intra-atrial reentrant tachycardia
- * Pulmonary and systemic venous obstruction
- * Thromboembolism
- * Protein-losing enteropathy
- * Low cardiac output

Thirty-two years after total right heart bypass

Guillermo O. Kreutzer, MD, Buenos Aires, Argentina

F / 53Y

Tricuspid atresia Ib

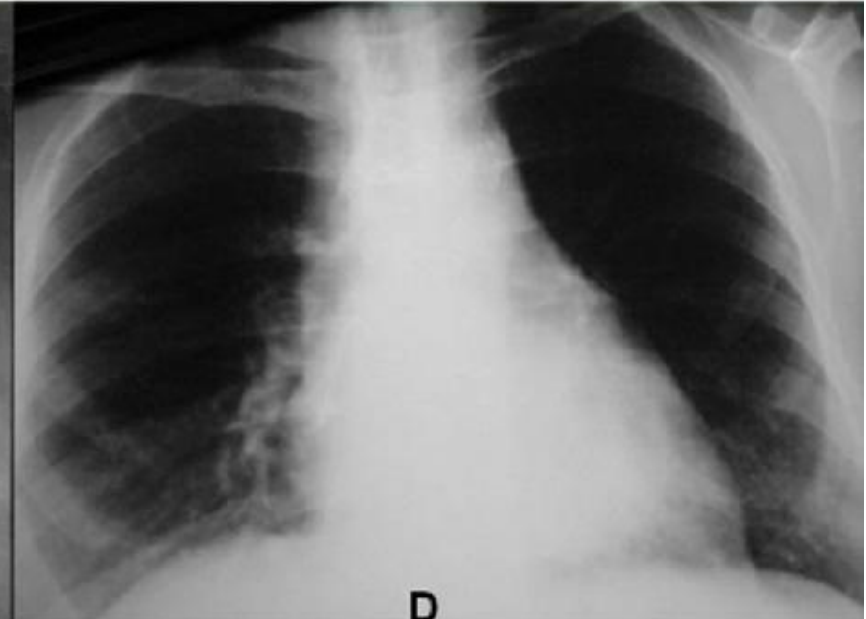
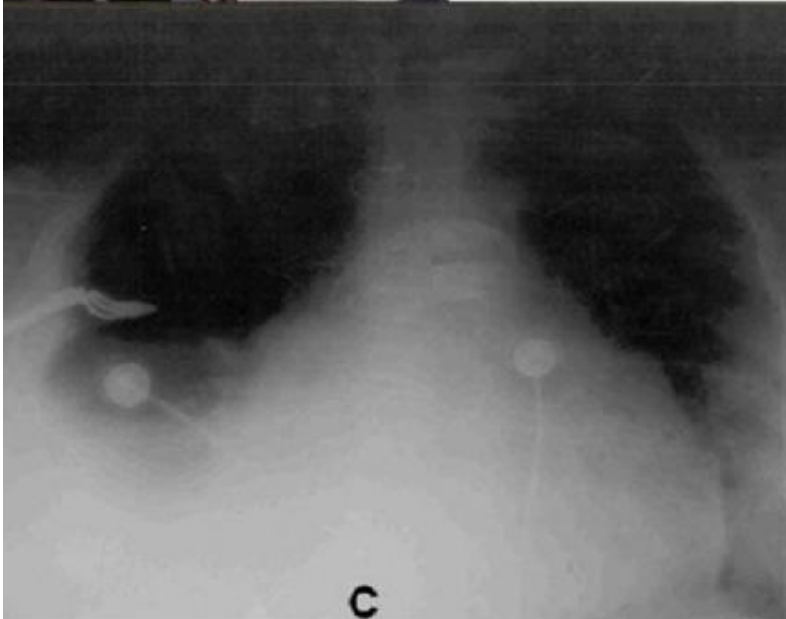
AP Fontan in 1975

Fontan conversion in 2006

The longest survivor in the world



B





111 Fontan Conversions with Arrhythmia Surgery: Surgical Lessons and Outcomes

Constantine Mavroudis, MD, Barbara J. Deal, MD, Carl L. Backer, MD, Robert D. Stewart, MD, MPH, Wayne H. Franklin, MD, Sabrina Tsao, MD, Kendra M. Ward, MD, and R. Andrew DeFreitas, MD

Divisions of Cardiovascular Thoracic Surgery and Cardiology, Children's Memorial Hospital, and Departments of Surgery and Pediatrics, Feinberg School of Medicine, Northwestern University, Chicago, Illinois

Background. The evolving operative strategy and course of 111 consecutive Fontan conversions with arrhythmia surgery and pacemaker therapy were reviewed to identify risk factors for poor outcome.

Methods. Since 1994, 111 patients (mean age 22.5 ± 7.9 years) underwent Fontan conversion with arrhythmia surgery. The series was divided into three time periods: (1) 1994 to 1996 (initial isthmus ablation, $n = 9$, group I); (2) 1996 to 2003 (early modified right atrial maze and Cox-maze III, $n = 51$, group II); and (3) 2003 to 2006 (recent modifications of the modified right atrial maze and left atrial Cox-maze III for both atrial fibrillation and left atrial reentry tachycardia, $n = 51$, group III).

Results. There were one early (0.9%) and six late deaths (5.4%); six patients required cardiac transplantation (5.4%). Two late deaths occurred after transplantation. Renal failure requiring dialysis occurred in four patients (3.6%). Mean hospital stay was 13.7 ± 12.1 days. Mean cross-clamp

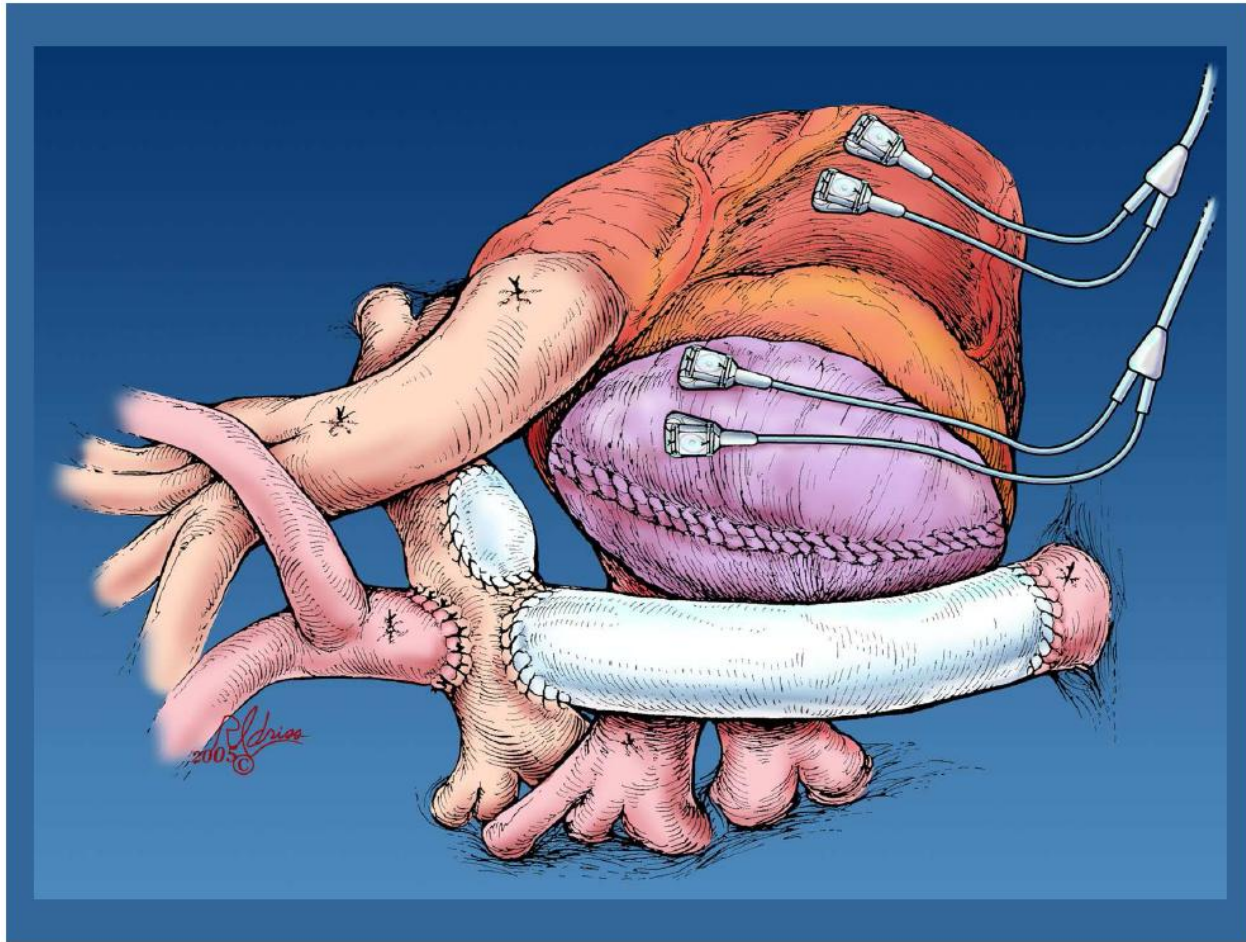
time was 70.8 ± 41.6 minutes. Four risk factors for death or transplantation were identified: presence of a right or ambiguous ventricle, preoperative protein-losing enteropathy, preoperative moderate-to-severe atrioventricular valve regurgitation, and long (>239 minutes) cardiopulmonary bypass time. In intergroup comparisons (groups I and II versus group III), three trends were noted: increased incidence of concomitant surgical repairs ($p = 0.03$), older patients ($p = 0.01$), and increased incidence of left atrial reentry tachycardia and atrial fibrillation ($p = 0.04$). Late recurrence of atrial tachycardia ensued in 15 of 111 (13.5%); 8 of 51 in group II (15.7%) and 4 of 51 in group III (7.8) ($p = 0.3$).

Conclusions. Fontan conversion with arrhythmia surgery is safe and efficacious. Based on improved results and evolving surgical techniques, selection criteria can be more clearly defined.

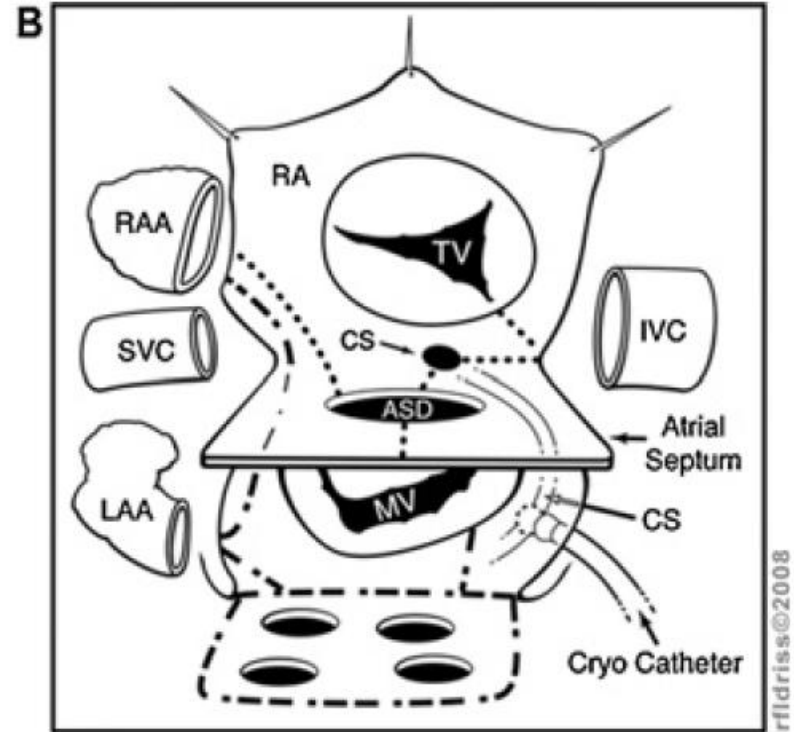
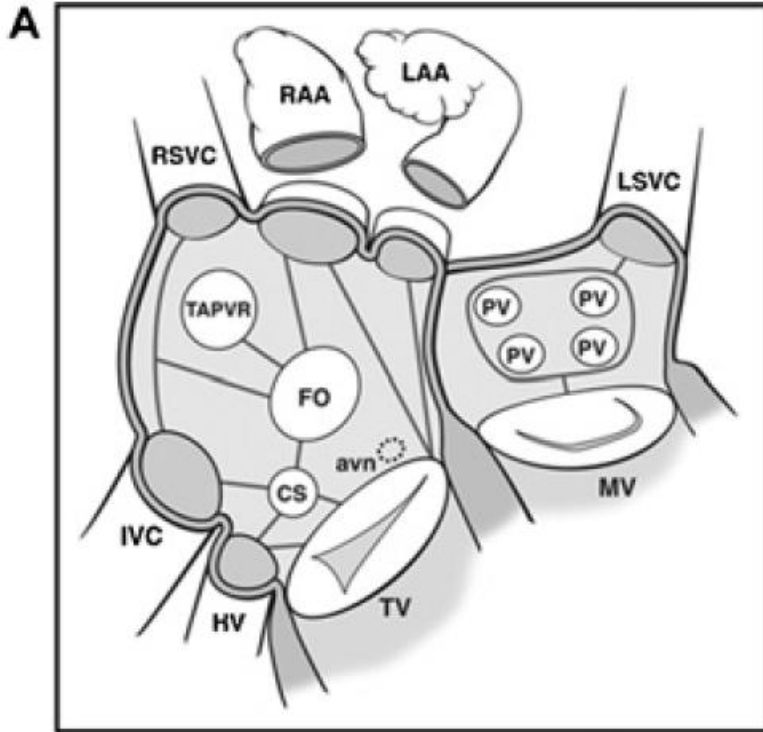
(Ann Thorac Surg 2007;84:1457-66)

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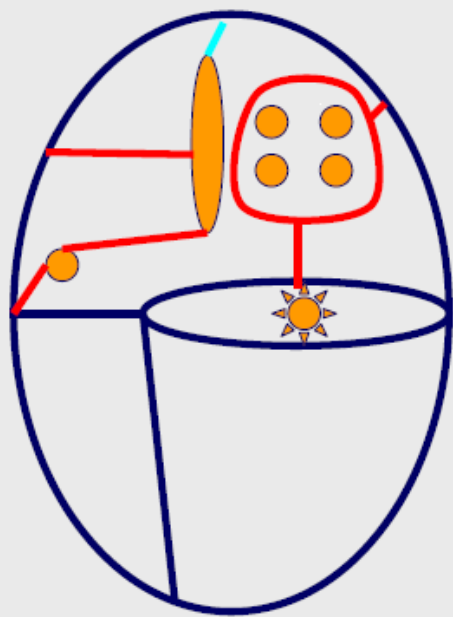
Conversion to Extracardiac Fontan



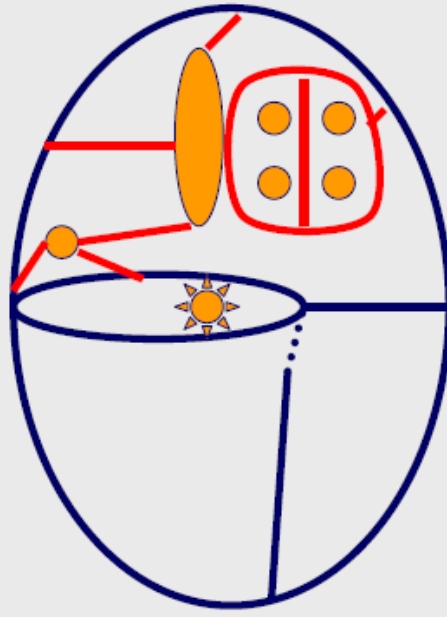
Arrhythmia Surgery



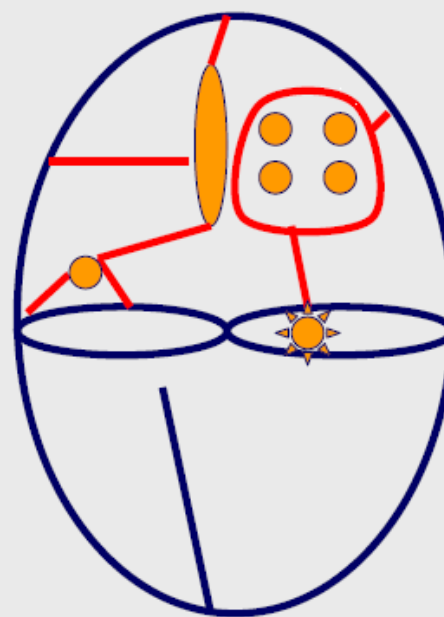
Surgical Modifications of Maze: Complex Anatomy



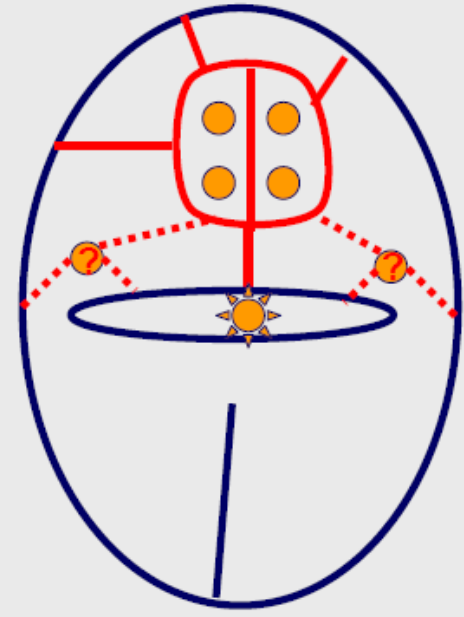
**Tricuspid
atresia**



**HLHS
DORV/MA**



DILV



Heterotaxy

Mavroudis C et al:

Semin Thorac Cardiovasc Surg Pediatr Surg Annu 2007;10:136

Device management of arrhythmias after Fontan conversion

Sabrina Tsao, MD,^{a,c} Barbara J. Deal, MD,^{a,c} Carl L. Backer, MD,^{b,d} Kendra Ward, MD,^{a,c}
Wayne H. Franklin, MD,^{a,c} and Constantine Mavroudis, MD^e

Objectives: We assessed our pacemaker strategy, use of antitachycardia therapies, generator longevity, and need for programming changes in patients having Fontan conversion with arrhythmia surgery.

Methods: Between 1994 and 2008, of 121 consecutive patients having Fontan conversion and arrhythmia surgeries, 120 patients underwent pacemaker implantation at the time of Fontan conversion (mean age 22.9 ± 8.1 years). Prior pacemakers were in place in 32/120 (26.7%) patients. Between 1994 and 1998, single-chamber atrial antitachycardia pacemakers were implanted ($n = 12$); atrial rate-responsive pacemakers ($n = 31$) were implanted between 1998 and 2002. Dual-chamber rate-responsive pacemakers ($n = 16$) were used between 2002 and 2003, and subsequently dual-chamber antitachycardia pacemakers ($n = 61$) have become the pacemaker of choice. Leads have evolved from transatrial endocardial leads to epicardial unipolar and subsequently bipolar leads.

Results: Among 87 patients with adequate follow-up, all are currently atrially paced at a minimum lower rate ≥ 70 beats per minute. Single-chamber atrial pacemakers were implanted in 43 (antitachycardia in 12), and dual-chamber pacemakers in 77 (antitachycardia in 61); multisite ventricular leads were placed in 7 patients. Far-field R-wave sensing in 78.6% of unipolar atrial leads led to use of epicardial bipolar leads. Late atrioventricular block (24%) led to routine implantation of dual-chamber pacemakers. Antitachycardia pacing was utilized in 7%. One patient required acute lead revision and 4 required late upgrade to dual-chamber pacemakers. There was no pacemaker-related infection. Twenty patients required generator change, and the mean device longevity was 7.53 years.

Conclusions: Customized pacemaker therapy can optimize management of patients following Fontan conversion. Device longevity is excellent. Based on our experience with 120 Fontan conversions, we recommend placement of a dual-chamber antitachycardia pacemaker with bipolar steroid-eluting epicardial leads in all patients at the time of the conversion.



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Fontan conversion with arrhythmia surgery[☆]

Woong-Han Kim^a, Hong Gook Lim^a, Jeong Ryul Lee^a, Joon Ryang Rho^a,
Eun Jung Bae^b, Chung Il Noh^b, Yong Soo Yoon^b, Yong Jin Kim^{a,*}

^a*Department of Thoracic and Cardiovascular Surgery, Seoul National University Children's Hospital, Seoul National University College of Medicine, 28 Yongon-Dong, Chongno-Gu Seoul 110-799, Korea*

^b*Department of Pediatrics, Seoul National University Children's Hospital, Seoul National University College of Medicine, 28 Yongon-Dong, Chongno-Gu Seoul 110-799, Korea*

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Abstract

Objective: Hemodynamic abnormalities and refractory atrial arrhythmias in patients late after the Fontan operation result in significant morbidity and mortality. We reviewed our experience with Fontan conversion and concomitant arrhythmia surgery. **Methods:** Between January 1996 and February 2004, 16 patients underwent Fontan conversion and arrhythmia surgery. Mean age at the initial Fontan operation was 5.1 ± 3.5 (range: 2-15) years and mean age at Fontan conversion was 17.0 ± 5.8 (range: 6-30). The initial Fontan operations were atriopulmonary connections in 14 patients, extracardiac lateral tunnel in 1, and intracardiac lateral tunnel in 1. The types of arrhythmia included atrial flutter in 10 patients and atrial fibrillation in 3. Fontan conversion operation was performed with intracardiac lateral tunnel in 5 patients and extracardiac conduit in 11. Arrhythmia surgery included isthmus cryoablation in 10 patients and right-sided maze in 3. **Results:** There has been no mortality. At Fontan conversion operation, 7 patients required permanent pacemaker. All patients have improved to New York Heart Association class I or II. With a mean follow-up of 26.9 ± 30.6 (range: 1-87) months, 16 patients had sinus rhythm, 2 patients had transient atrial flutter which was well controlled, and 2 patients required permanent pacemaker during follow-up. **Conclusions:** Fontan conversion with concomitant arrhythmia surgery and permanent pacemaker placement is safe, improves New York Heart Association functional class, and has a low incidence of recurrent arrhythmias. In most patients, concomitant permanent pacemakers are needed.

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AV Valve Regurgitation

- **Associated with worse survival**
- **Mechanism**
 - * **Chronic volume overload**
 - * **Structural abnormalities of AV valve**
 - * **Tricuspid valve as a systemic AV valve**
 - * **Ventricular dysfunction**

Timing of AV Valve Surgery

- Rarely before BCPC
- Mostly at the time of BCPC
- Sometimes between BCPC and Fontan
- Sometimes at the time of Fontan
- Rarely after Fontan

Early Surgical Correction of Atrioventricular Valvular Regurgitation in Single-Ventricle Patients

Jae Gun Kwak, MD, Chun Soo Park, MD, Chang-Ha Lee, MD, Cheul Lee, MD, Soo-Jin Kim, MD, Jin Young Song, MD, and Woo-Sup Shim, MD

Department of Thoracic and Cardiovascular Surgery, and Department of Pediatric Cardiology, Sejong General Hospital, Bucheon, South Korea

Background. The purpose of this study is to evaluate the effectiveness of early surgical correction for atrioventricular valve regurgitation (AVVR) in single-ventricle patients.

Methods. The medical records of 39 single-ventricle patients who underwent atrioventricular surgery more than once between 1996 and 2008 were reviewed.

Results. The mean preoperative grade of AVVR was 2.6 ± 0.7 . Four patients underwent valvular operations at first palliative surgery, 3 patients before bidirectional cavopulmonary connection, 13 at bidirectional cavopulmonary connection, 6 in the interstage between bidirectional cavopulmonary connection and Fontan, 10 at Fontan, and 3 after Fontan procedure. Surgical techniques for valve were edge-to-edge sutures for bridging leaflets, leaflet cleft repair, partial or complete annuloplasty with strip, or artificial valve implantation. Although there was no statistical significance, the patients who underwent early operation (AVVR grade less than 2) showed a

tendency toward better atrioventricular valvular function during the postoperative follow-up (57.1 months, range: 2 ~ 129 months). None of the patients whose preoperative AVVR grade less than 2 showed an AVVR more than 2 at the final echocardiography. There were 4 deaths overall due to sepsis, Fontan failure, and sudden cardiac arrest. Final echocardiographic findings showed a mean AVVR of 1.6 ± 0.8 and acceptable cardiac function. All living patients were in a good New York Heart Association functional class (1.07 ± 0.2).

Conclusions. Even though we could not find statistically significant evidence of benefit for early correction of AVVR in single-ventricle patients, the patients undergoing early valvular operation for regurgitation showed a tendency toward better atrioventricular valvular function at midterm.

(Ann Thorac Surg 2010;90:1320–3)

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Table 2. Timing of Valve Operations

| Timing of Valve Operation | Number of Patients |
|------------------------------|--------------------|
| First palliative operation | 4 |
| Before BCPC | 3 |
| Concomitant with BCPC | 13 |
| After BCPC and before Fontan | 6 |
| Concomitant with Fontan | 10 |
| After Fontan | 3 |

BCPC = bidirectional cavopulmonary connection.



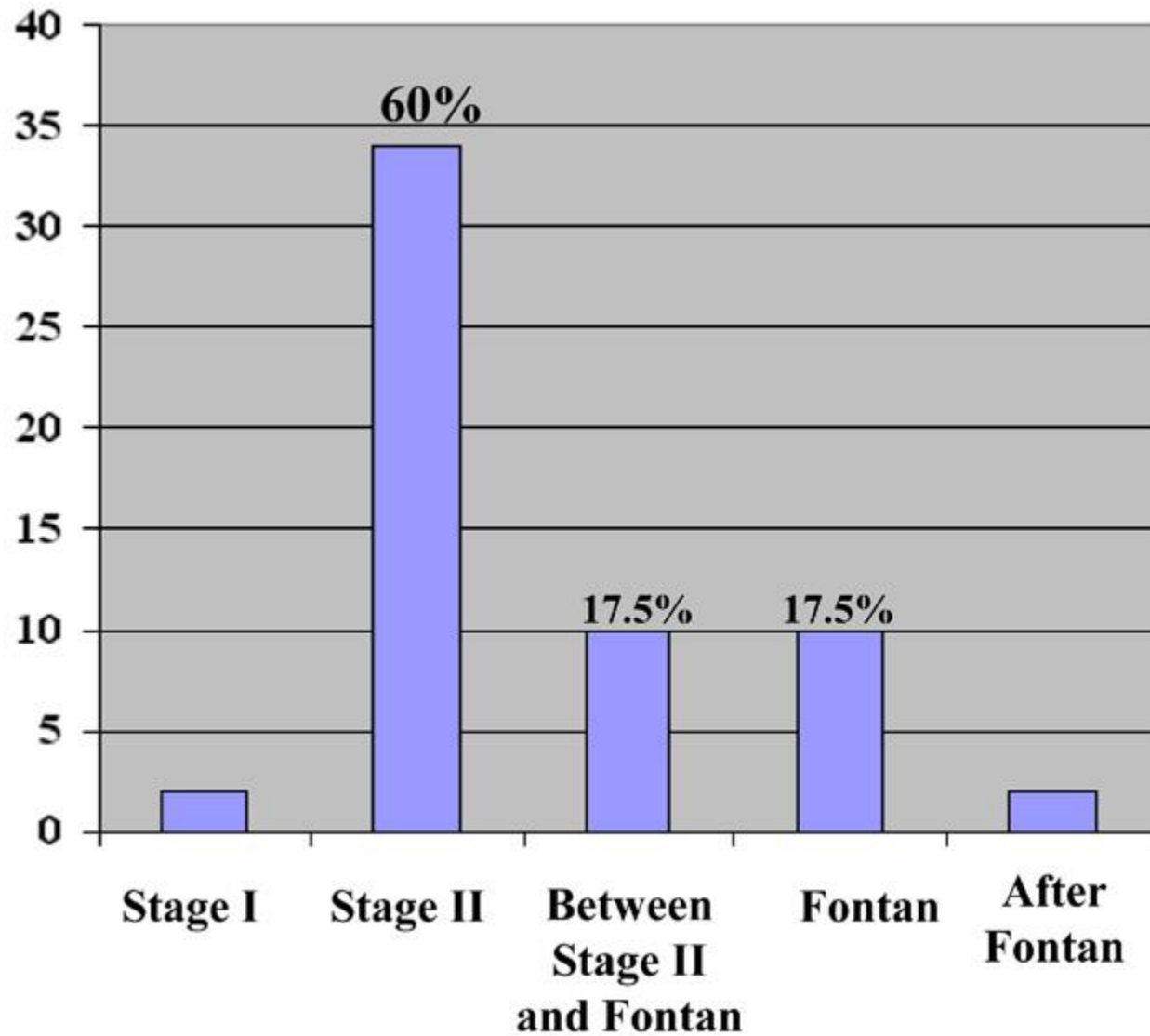
Atrioventricular Valve Repair in Patients With Single-ventricle Physiology: Mechanisms, Techniques of Repair, and Clinical Outcomes

Osami Honjo, MD, PhD, Luc Mertens, MD, PhD, and Glen S. Van Arsdell, MD

Significant atrioventricular (AV) valve insufficiency in patient with single ventricle-physiology is strongly associated with poor survival. Herein we discuss the etiology and mechanism of development of significant AV valve insufficiency in patients with single-ventricle physiology, surgical indication and repair techniques, and clinical outcomes along with our 10-year surgical experience. Our recent clinical series and literature review indicate that it is of prime importance to appreciate the high incidence and clinical effect of the structural abnormalities of AV valve. Valve repair at stage II palliation may minimize the period of volume overload, thereby potentially preserving post-repair ventricular function. Since 85% of the AV valve insufficiency was associated with structural abnormalities, inspection of an AV valve that has more than mild to moderate insufficiency is recommended because they are not likely to be successfully treated with volume unloading surgery alone.

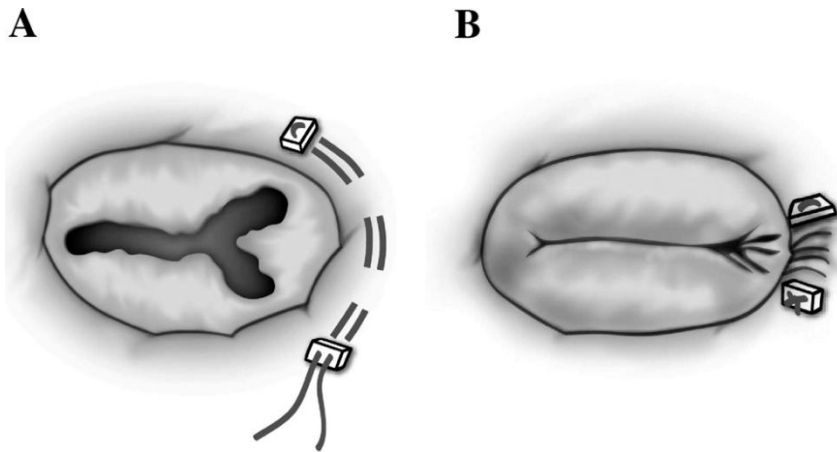
Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 14:75-84 © 2011 Published by Elsevier Inc.

Patients (n)

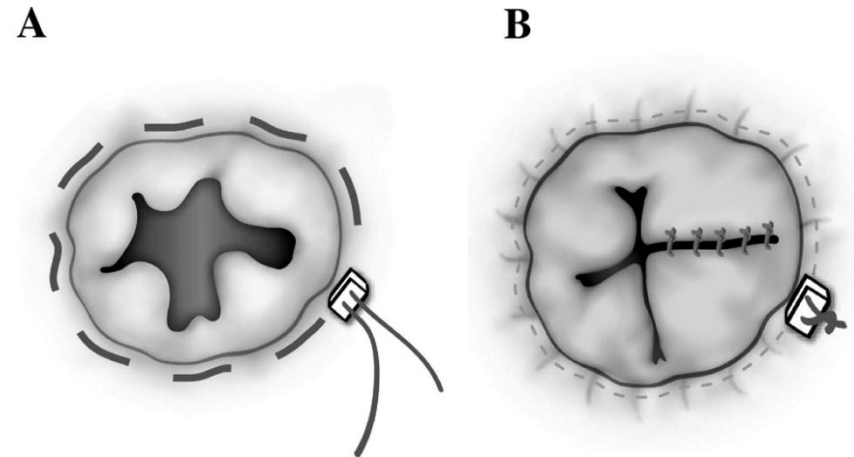


Techniques for AV Valve Repair (1)

Partial annuloplasty

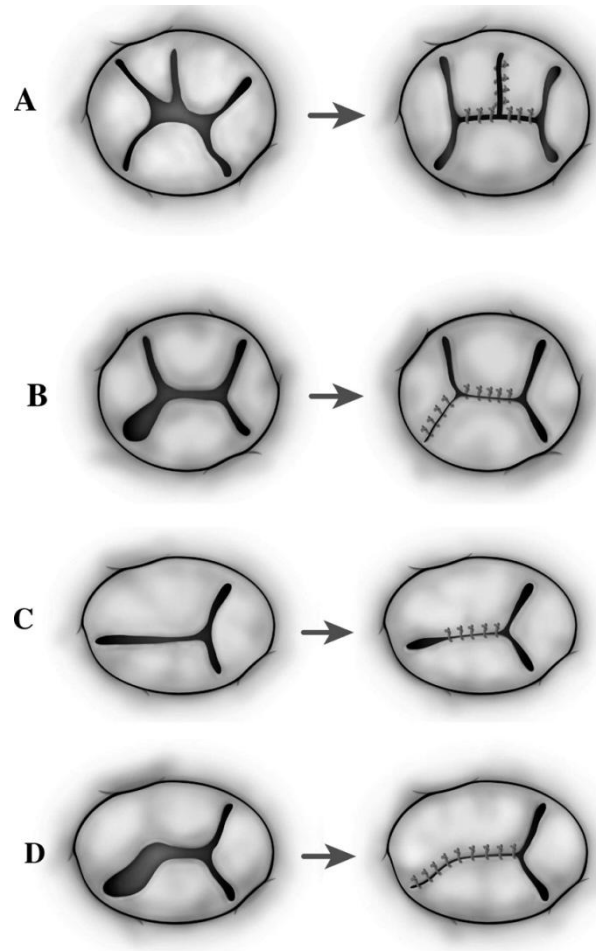


Circular annuloplasty for common AV valve



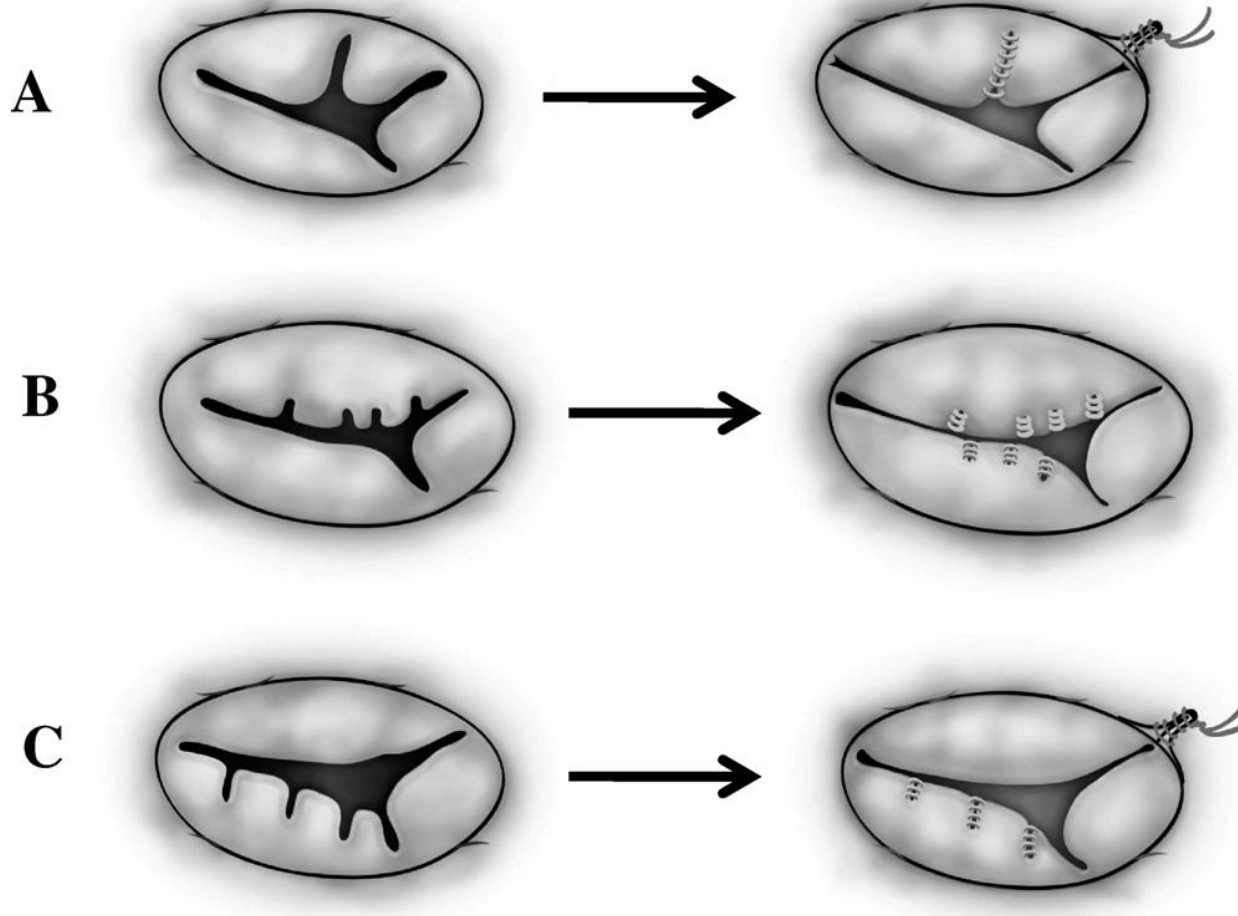
Techniques for AV Valve Repair (2)

Edge-to-edge repair



Techniques for AV Valve Repair (3)

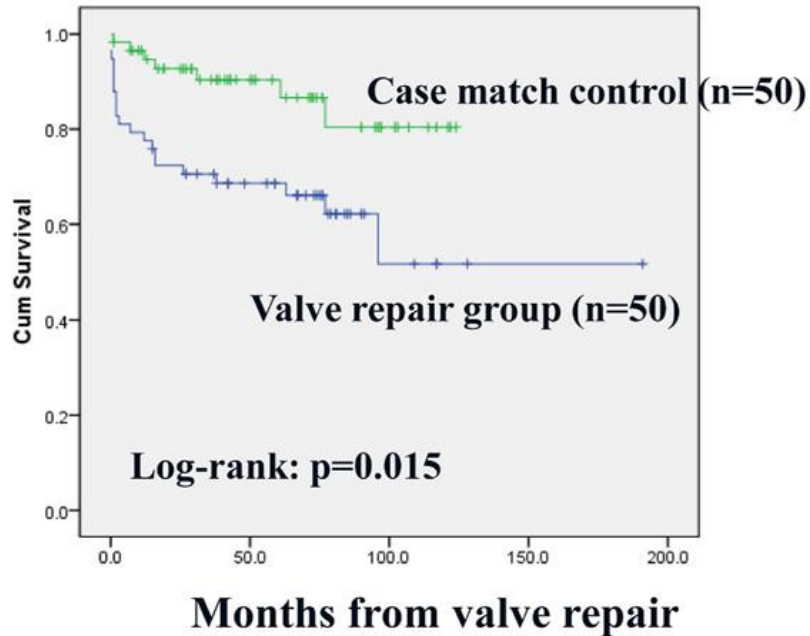
Repair of dysplastic or dysmorphic tricuspid valve in HLHS



Outcome of AV Valve Repair

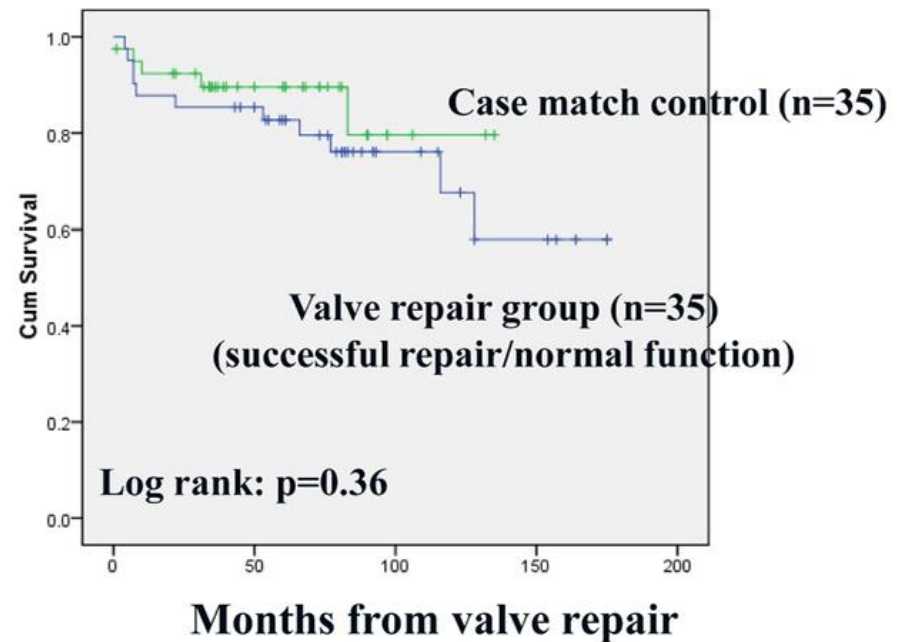
A

**Freedom from death/transplant
Valve repair vs. Matched control**



B

**Freedom from death/transplant
Successful repair/normal function**



Pulmonary AV Fistula

- **“Hepatic factor” in hepatic venous blood**
- **Pulmonary AV fistulae may develop after**
 - * **BCPC**
 - * **Kawashima procedure**
 - * **Fontan completion**

Development of Pulmonary Arteriovenous Fistulas After Bidirectional Cavopulmonary Shunt

Soo Jin Kim, MD, Eun Jung Bae, MD, Do Jun Cho, MD, In Seung Park, MD, Yang Min Kim, MD, Woong-Han Kim, MD, and Seong Ho Kim, MD

Departments of Pediatric Cardiology, Radiology, and Cardiac Surgery, Sejong Heart Institute, Sejong General Hospital, Puchon City, Korea

Background. A high incidence of pulmonary arteriovenous fistulas (PAVF) has been reported after bidirectional cavopulmonary shunt (BCPS) or total cavopulmonary shunt (TCPS; BCPS in patients with interrupted inferior vena cava). However, the definite diagnostic criteria or standard diagnostic modality of PAVF has not yet been defined. The goal of this study was to evaluate the diagnostic modalities and the prevalence of PAVF.

Methods. We selected 10 patients with TCPS and 27 patients with BCPS. Lung perfusion scan, contrast echocardiogram, and pulmonary angiogram were performed. The results were compared among groups of patients and among each diagnostic modality.

Results. All 10 patients with TCPS and 16 and 13

patients with BCPS showed positive results on contrast echocardiograms and lung scans, respectively. Six patients with TCPS and 4 patients with BCPS showed positive results on pulmonary angiograms. All patients with TCPS developed subclinical or clinical PAVF and 19 patients with BCPS developed subclinical PAVF and none of them had clinical PAVF during the short-term follow-up.

Conclusions. Most patients with bidirectional cavopulmonary anastomosis have subclinical evidence of right-to-left intrapulmonary shunting. This problem can be demonstrated with various diagnostic modalities.

(Ann Thorac Surg 2000;70:1918–22)

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Pulmonary Arteriovenous Malformations in Children After the Kawashima Operation

John W. Brown, MD, Mark Ruzmetov, MD, PhD, Palaniswamy Vijay, PhD, Mark D. Rodefeld, MD, and Mark W. Turrentine, MD

Section of Cardiothoracic Surgery, Indiana University School of Medicine, and James Whitcomb Riley Hospital for Children, Indianapolis, Indiana

Background. Pulmonary arteriovenous malformations are a major cause of progressive late cyanosis in patients treated with cavopulmonary anastomoses. Previous experience suggests that exclusion of the hepatic venous effluent from the pulmonary circulation may cause the development of pulmonary arteriovenous malformations after the Kawashima operation in children with interrupted inferior vena cava with azygous continuation.

Methods. From January 1990 to November 2004, 21 children (median age, 2 years) with heterotaxy syndrome and interrupted inferior vena cava with azygous continuation underwent Kawashima operation. The average preoperative arterial oxygen saturation was $76\% \pm 7\%$ (range, 64% to 90%).

Results. Follow-up was complete in all survivors except 1 at a median duration of 4.5 years. Pulmonary arteriovenous malformations were diagnosed at a median of 5 years after Kawashima operation in 11 patients (58%). Completion Fontan operation has been performed in 15 (79%). Five children who underwent a completion Fontan procedure 1 to 1.5 years after Kawashima opera-

tion did not have pulmonary arteriovenous malformations. There were 2 late deaths due to chronic congestive heart failure 4 months and 7 years after Kawashima operation. Overall survival at 10 years is 90%. Univariate and multivariate analysis demonstrated presence of bilateral superior vena cavae ($p = 0.002$) and interval longer than 2 years between Kawashima operation and completion Fontan operation ($p = 0.04$) as predictors of developing pulmonary arteriovenous malformations.

Conclusions. In most patients with heterotaxy and interrupted inferior vena cava with azygous continuation, clinical evidence of pulmonary arteriovenous malformations will develop after Kawashima operation if they are followed up long enough. Early redirection of the hepatic venous effluent to the pulmonary arterial circulation may prevent or lead to regression of pulmonary arteriovenous malformations, with low mortality and morbidity.

(Ann Thorac Surg 2005;80:1592-6)

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PEDIATRIC CARDIAC SURGERY:

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Inclusion of Hepatic Venous Drainage in Patients with Pulmonary Arteriovenous Fistulas

Soo-Jin Kim, MD, Eun-Jung Bae, MD, Jae-Young Lee, MD, Hong-Gook Lim, MD, Cheul Lee, MD, and Chang-Ha Lee, MD

Departments of Pediatric Cardiology and Cardiac Surgery, Sejong General Hospital, Bucheon, Department of Pediatrics, Seoul National University College of Medicine, Seoul National University Children's Hospital, Seoul, Korea

Background. It is well known that hepatic vein (HV) inclusion can ameliorate cyanosis in patients with pulmonary arteriovenous fistulas (PAVFs) during the sequence of Fontan type repair. Previously, we reported that most patients with bidirectional cavopulmonary shunt (BCPS) have clinical or subclinical evidence of a right to left shunt through PAVFs.

Methods. We studied 33 patients who already had clinical and subclinical PAVFs after BCPS. All patients have taken Fontan completion with HV inclusion. The state of PAVFs was reevaluated by pulmonary angiogram, contrast echocardiography, and lung scintigraphy 7.7 ± 2.4 years after HV inclusion.

Results. After Fontan completion, the mean oxygen saturation increased from 80.2 ± 7.4% to 91.5 ± 9.8% in the entire cohort. Moreover, the amount of right-to-left shunting through the PAVFs, measured by lung scintig-

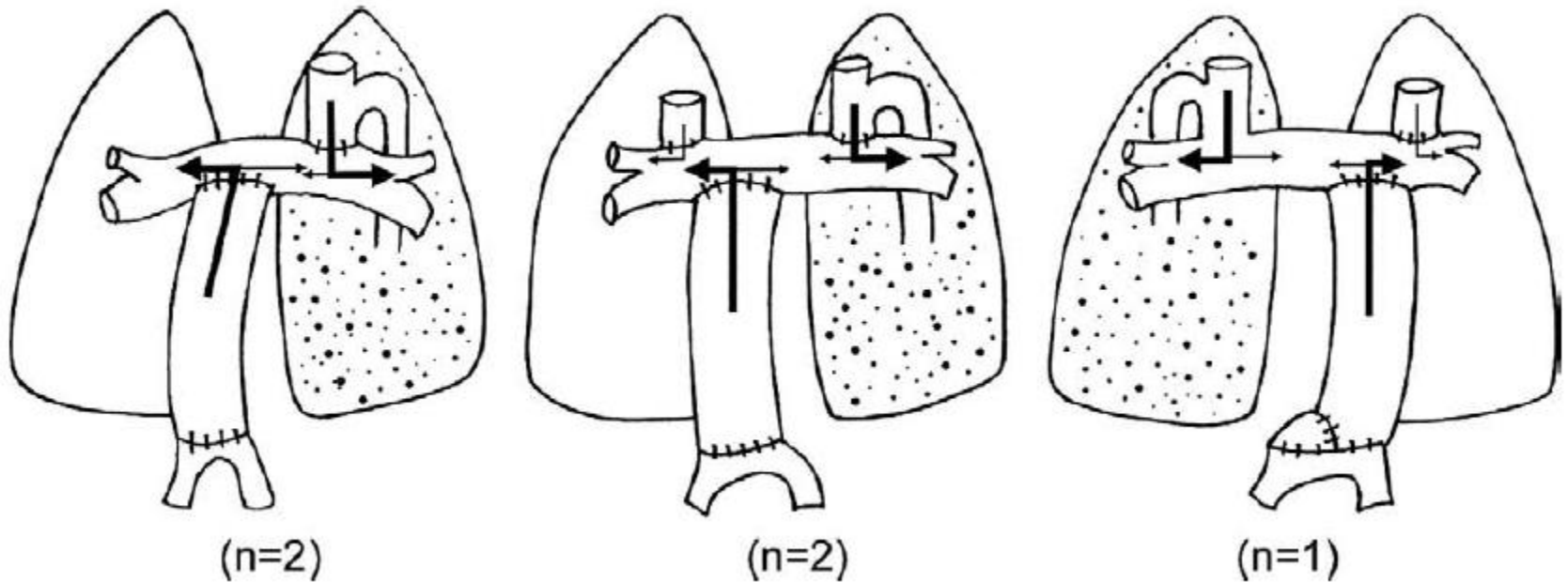
raphy, was decreased from a mean of 23.8 ± 15.1 to 13.0 ± 8.2%. The degree of severity, for most patients, was decreased as demonstrated by contrast echocardiography. However, 5 patients (16.7%) showed persistent PAVFs, even after the HV inclusion. They all had left isomerism with azygous continuation of the IVC and the conduit was positioned on the contralateral side to the SVC with azygous drainage.

Conclusions. Most PAVFs regressed after Fontan completion. Left isomerism with azygous continuation of the IVC had risk for persistent PAVFs when the HV conduit was positioned at the contralateral side to the SVC receiving the azygous drainage. Therefore, appropriate design avoiding unilateral streaming of HV flow should be considered for HV inclusion surgery.

(Ann Thorac Surg 2009;87:548–54)

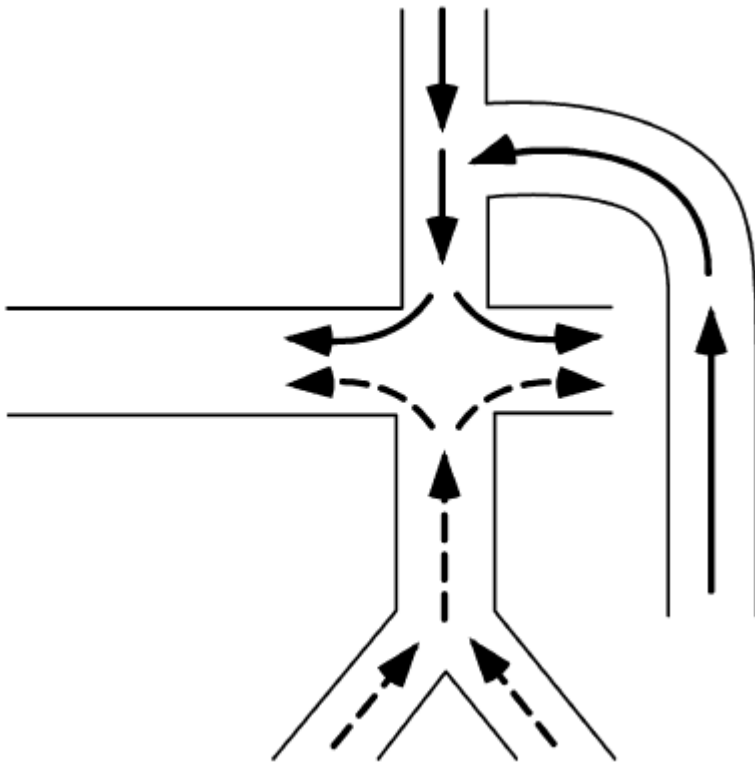
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Uneven Distribution of HV Flow

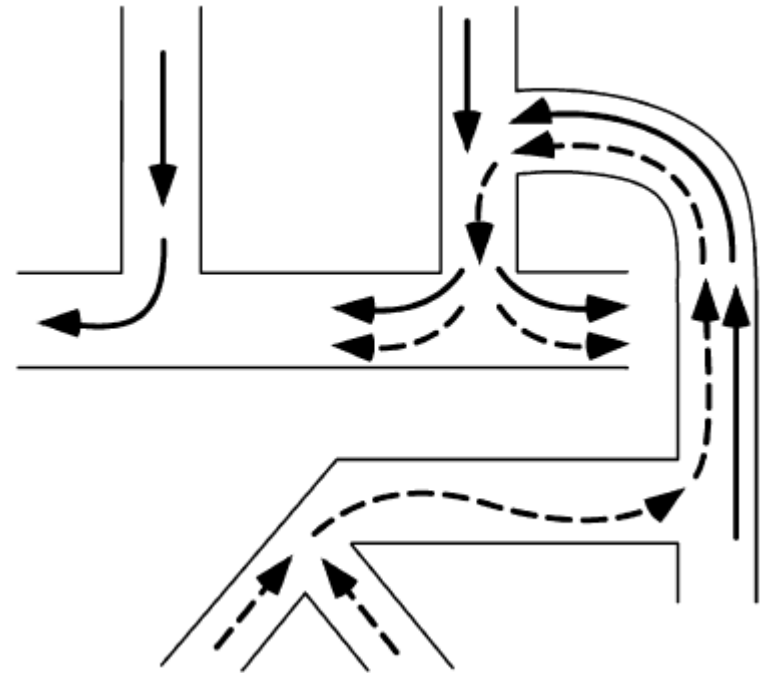


Surgical Solutions

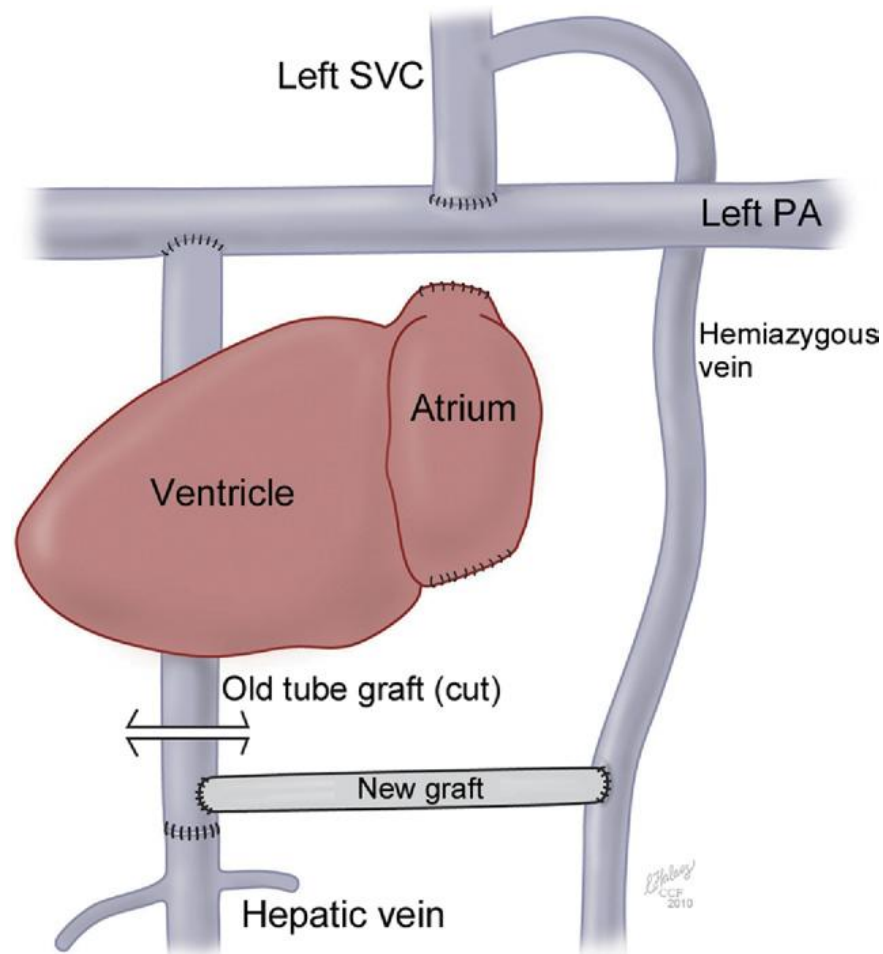
No Offset



HV-to-azygos vein connection



HV-to-Azygos Vein Connection



Cavopulmonary pathway modification in patients with heterotaxy and newly diagnosed or persistent pulmonary arteriovenous malformations after a modified Fontan operation

Doff B. McElhinney, MD,^a Gerald R. Marx, MD,^a Audrey C. Marshall, MD,^a John E. Mayer, MD,^b and Pedro J. del Nido, MD^b

Objective: Pulmonary arteriovenous malformations are an important but uncommon complication of cavopulmonary connection, particularly in patients with heterotaxy. Absence of hepatic venous effluent in pulmonary arterial blood seems to be a predisposing factor. Pulmonary arteriovenous malformations are most common after superior cavopulmonary anastomosis, but may develop, progress, or persist in 1 lung after Fontan completion if hepatic venous blood streams completely or primarily to the contralateral lung.

Methods: Among 53 patients with heterotaxy and inferior vena cava interruption who underwent a modified Fontan procedure from 1985 to 2005, 8 had unilateral streaming of hepatic venous flow and clinically significant pulmonary arteriovenous malformations after hepatic venous inclusion and underwent reconfiguration of the cavopulmonary pathway. In all 8 patients, the hepatic vein-pulmonary artery pathway was contralateral to and offset from the pulmonary artery anastomosis of the single or dominant superior vena cava. Pathway reconfiguration included pulmonary arterial stenting (n = 2), revision of the superior vena cava-pulmonary artery connection (n = 1), construction of a branched hepatic vein-pulmonary artery conduit (n = 2), and surgical or transcatheter construction of a direct hepatic vein-azygous vein pathway (n = 5).

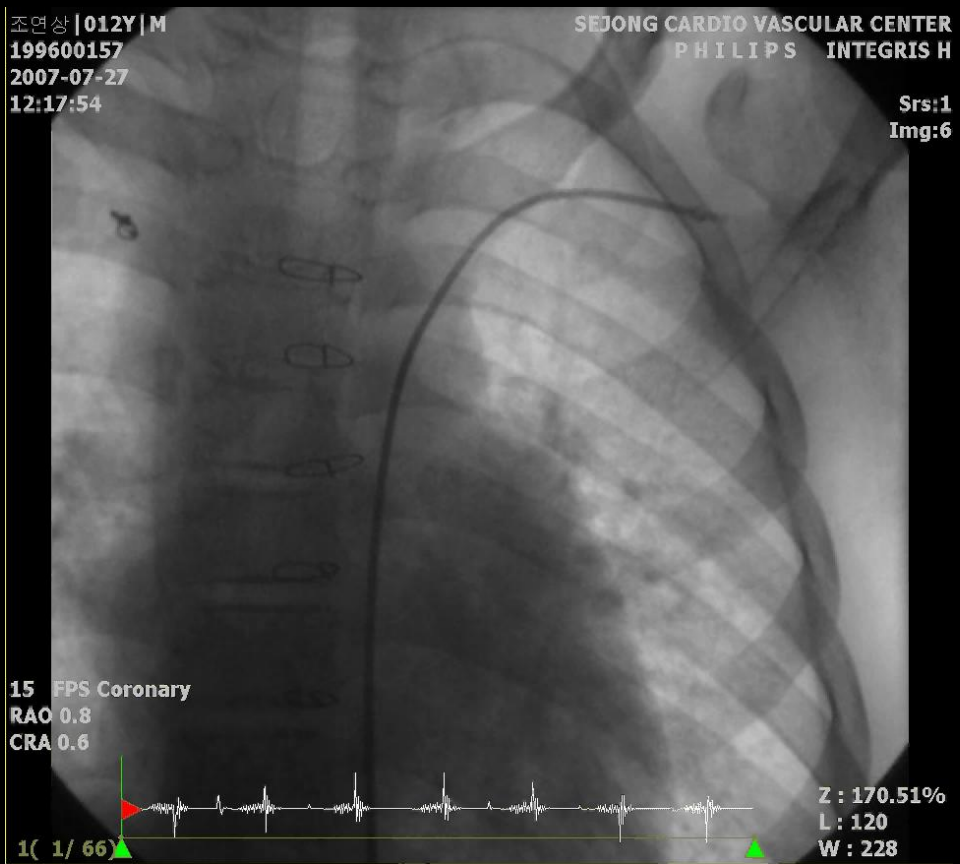
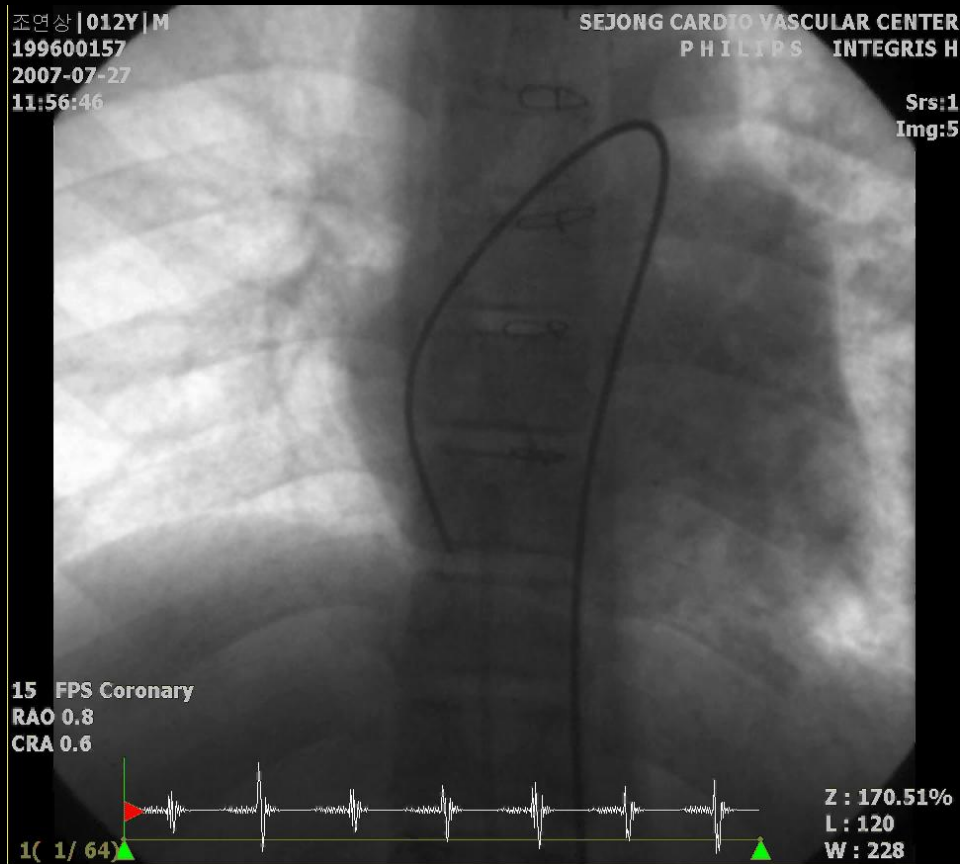
Results: Hepatic vein-azygous vein connection led to improvement in 4 of 5 patients; other approaches typically did not lead to improvement.

Conclusions: Resolution of hypoxemia after cavopulmonary pathway reconfiguration in patients with unilateral pulmonary arteriovenous malformations and hepatic venous flow-streaming after Fontan completion supports the importance of hepatic venous effluent in the pathogenesis of pulmonary arteriovenous malformations and the practice of cavopulmonary pathway revision in such patients. Completion or reconfiguration of the Fontan circulation with direct hepatic vein-azygous vein connection may provide the most reliable mixing and bilateral distribution of hepatic venous blood in this population of patients. (J Thorac Cardiovasc Surg 2011;141:1362-70)

Case

- **M / 12Y**
- **Heterotaxy, bilateral SVC, IVC interruption**
- **s/p Kawashima at 15 m**
- **s/p Lateral tunnel Fontan at 28 m**
- **Progressive hypoxemia, SpO₂ 55%**

Preoperative Angiogram



Lung Perfusion Scan

012Y|M
199600157
2007-07-26
16:07:05

INFINIA
Acc:2007145203
Srs:1
Img:0

ANT

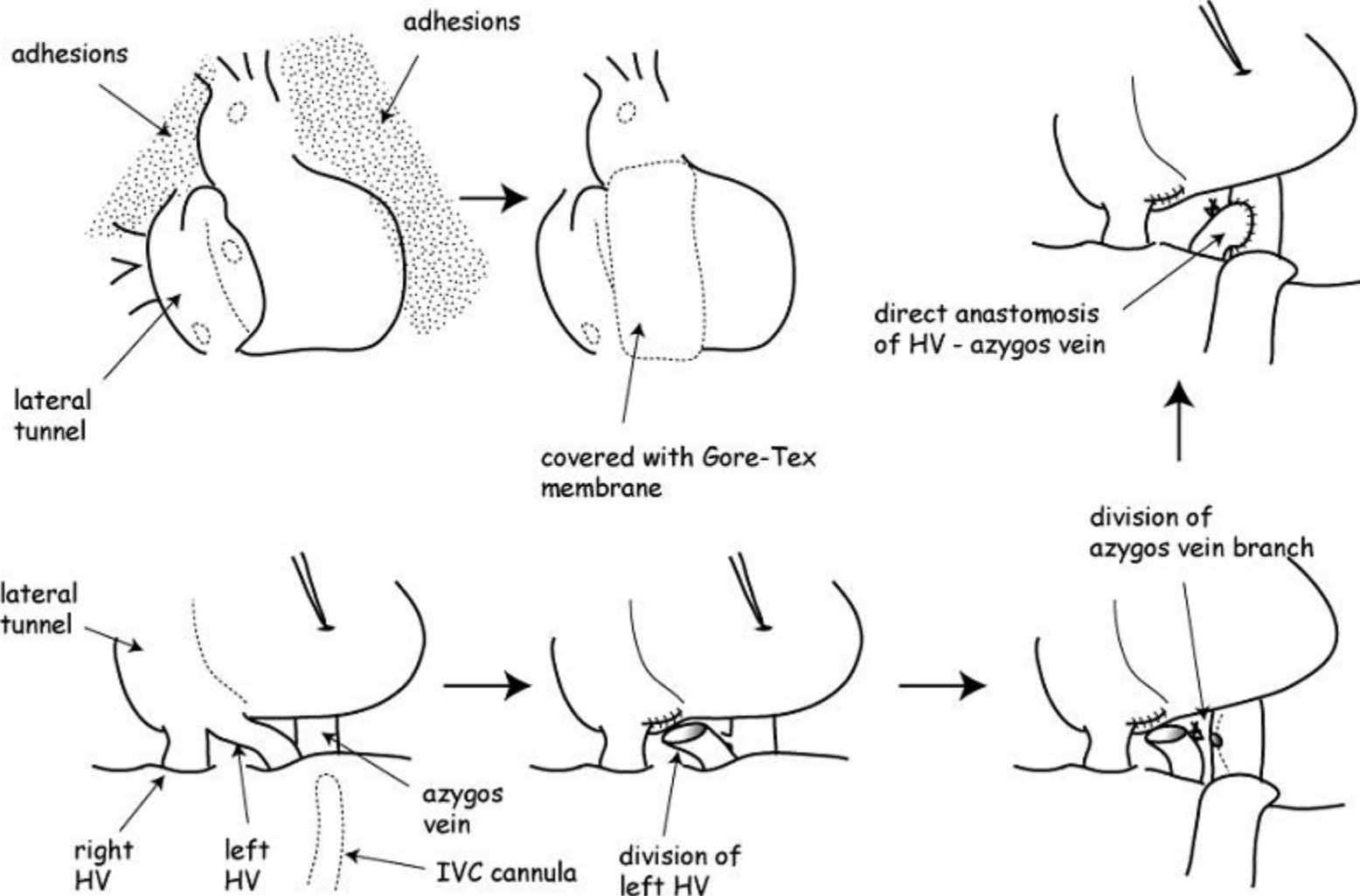
PP:FFS

Shunt fraction = 32%

28cm

Zoom : 344.53%
WL : 298
WW : 595

HV-to-Azygos Vein Connection



012Y|M

2007-08-21

Rapidia
Acc:2007163124
Srs:1002
Img:418

PP:HFS



Azygo

H.V

[L]

11cm

KVP:

[P]

Zoom : 194.27%
WL : 264
WW : 916

Follow-Up Status

- **SpO₂ 90%**
- **Shunt fraction 8% by LPS**

Conduit-Related Problems

- **Conduit thrombosis**
- **Conduit stenosis**

Predisposing Factors for Fontan Conduit Thrombosis

- **Poor hemodynamics**

 - Excessively oversized conduit relative to the IVC size**

- **Coagulation factor abnormalities**

 - Protein C/S deficiency, elevated factor VIII level...**

- **Foreign materials**

 - Artificial graft, venous catheter**

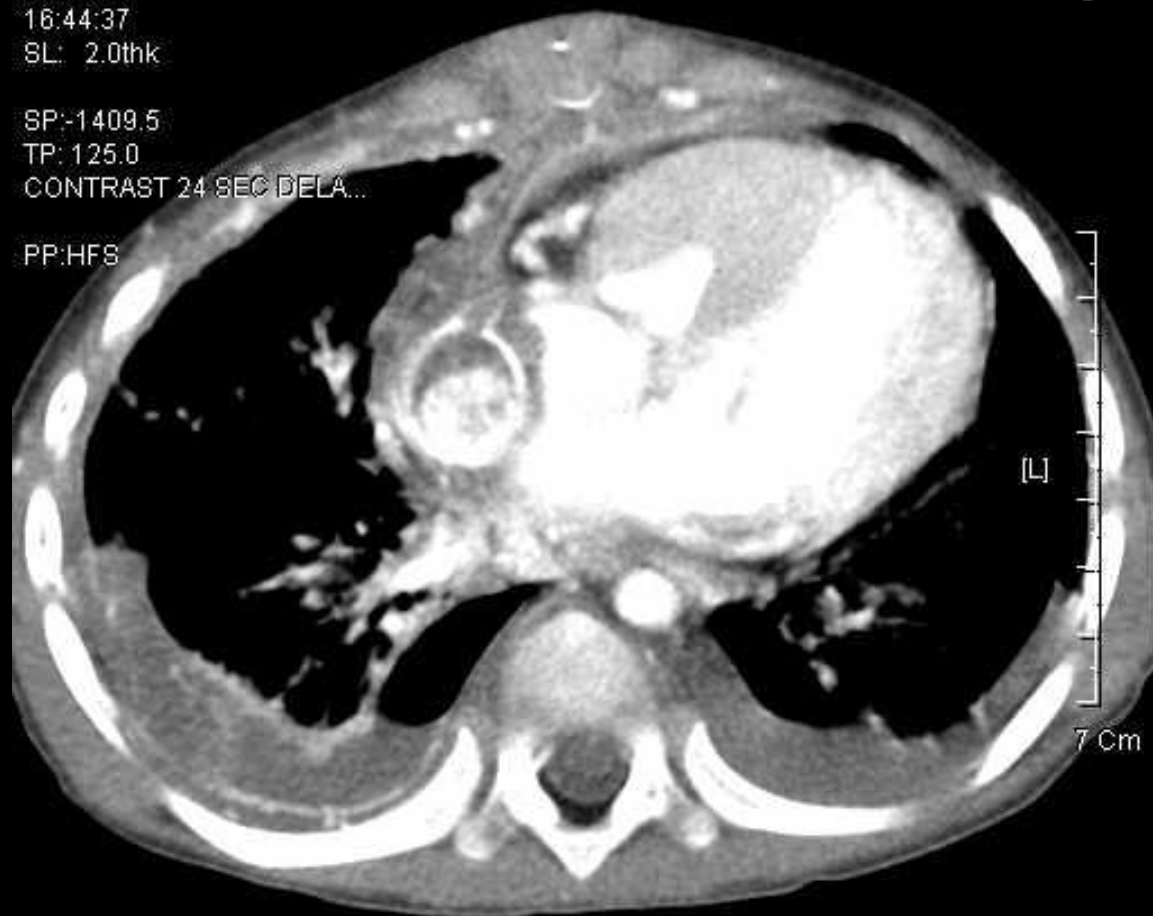
M/38m, PA with IVS, s/p Extracardiac Fontan

003Y/M
200102868
2004-06-14
16:44:37
SL: 2.0thk

Srs:9
Acq:24
Img:104

SP:-1409.5
TP: 125.0
CONTRAST 24 SEC DELA...

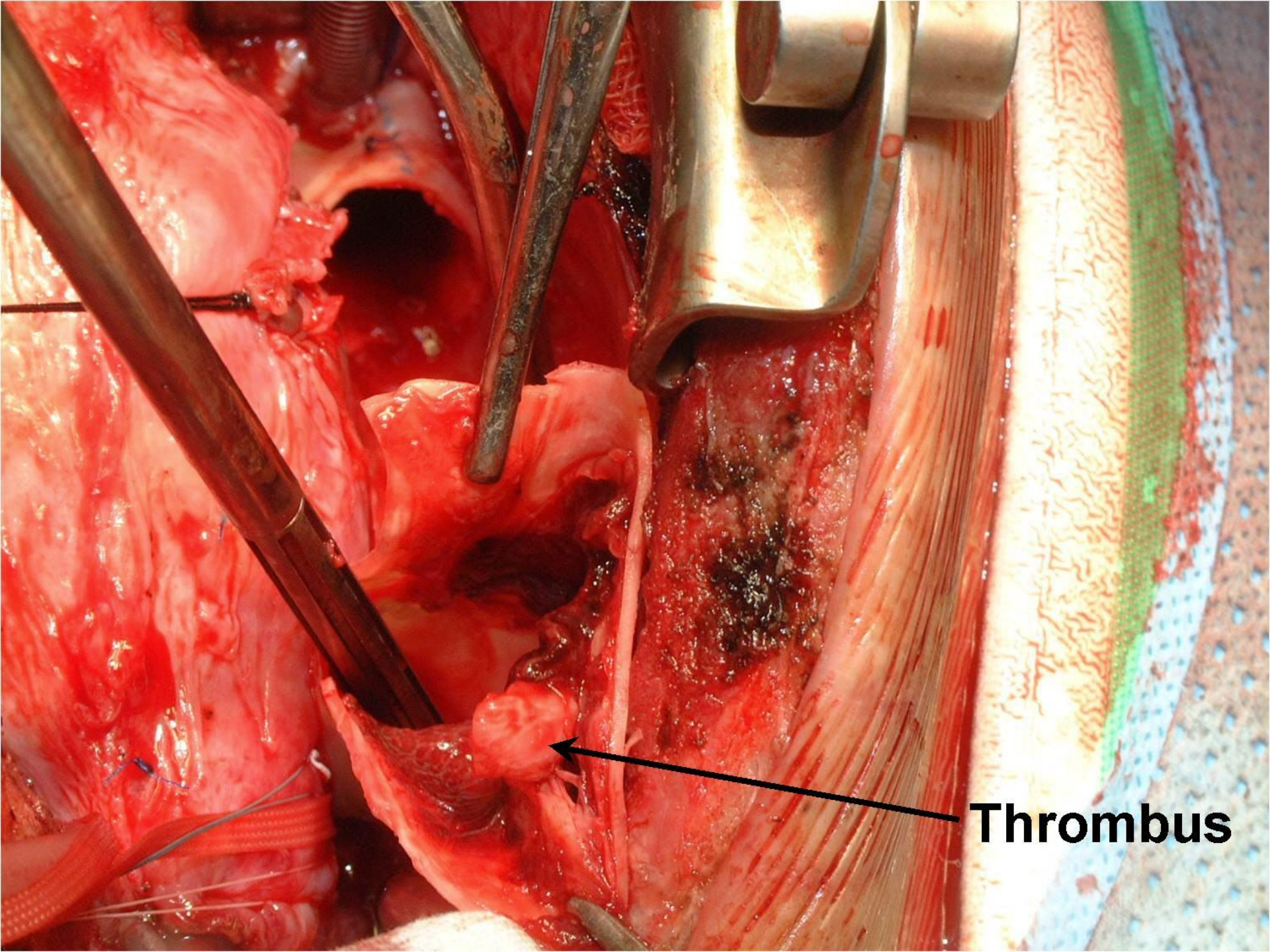
PP:HFS



KVP: 120.0
MAS:110

L:50
W:350

[P]



Thrombus

A Multicenter, Randomized Trial Comparing Heparin/Warfarin and Acetylsalicylic Acid as Primary Thromboprophylaxis for 2 Years After the Fontan Procedure in Children

Paul Monagle, MD, MSc, MBBS,* Andrew Cochrane, MD,* Robin Roberts, MSc,† Cedric Manlhiot, BSc,‡ Robert Weintraub, MBBS,* Barbara Szechtman, BA,† Marina Hughes, DPHIL,§ Maureen Andrew, MD,‡ Brian W. McCrindle, MD, MPH,‡ for the Fontan Anticoagulation Study Group

Melbourne, Victoria, Australia; and Toronto and Hamilton, Ontario, Canada

| | |
|--------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Objectives | The purpose of this study was to compare the safety and efficacy of acetylsalicylic acid (ASA) and warfarin for thromboprophylaxis after the Fontan procedure. |
| Background | Fontan surgery is the definitive palliation for children with single-ventricle physiology. Thrombosis is an important complication; the optimal thromboprophylaxis strategy has not been determined. |
| Methods | We performed a multicenter international randomized trial of primary prophylactic anticoagulation after Fontan surgery. Patients were randomized to receive for 2 years either ASA (5 mg/kg/day, no heparin phase) or warfarin (started within 24 h of heparin lead-in; target international normalized ratio: 2.0 to 3.0). Primary endpoint (Intention to treat) was thrombosis, intracardiac or embolic (all events adjudicated). At 3 months and 2 years after the Fontan procedure, transthoracic and transesophageal echocardiograms were obtained as routine surveillance. Major bleeding and death were primary adverse outcomes. |
| Results | A total of 111 eligible patients were randomized (57 to ASA, 54 to heparin/warfarin). Baseline characteristics for each group were similar. There were 2 deaths unrelated to thrombosis or bleeding. There were 13 thromboses in the heparin/warfarin group (3 clinical, 10 routine echo) and 12 thromboses in the ASA group (4 clinical, 8 routine echo). Overall freedom from thrombosis 2 years after Fontan surgery was 19%, despite thrombosis prophylaxis. Cumulative risk of thrombosis was persistent but varying and similar for both groups ($p = 0.45$). Major bleeding occurred in 1 patient in each group. |
| Conclusions | <u>There was no significant difference between ASA and heparin/warfarin as primary thromboprophylaxis in the first 2 years after Fontan surgery.</u> The thrombosis rate was suboptimal for both regimens, suggesting alternative approaches should be considered. (International Multi Centre Randomized Clinical Trial Of Anticoagulation In Children Following Fontan Procedures; NCT00182104) (J Am Coll Cardiol 2011;58:645–51) © 2011 by the American College of Cardiology Foundation |

Fontan Conduit Stenosis

- **Type of conduit**
- **Size of conduit**

OPERATION RECORD

등록번호:

성명:

Age/Sex: 12 / F (Birthdate:1996-04-27)

체중: 73.2 kg

체표: 1.8 m2

Op. Date 09-03-20 **Op No:** 25800

Surgeon 이철 / 김태식 / 김수미

Physician 김수진



Dx Extracardiac Fontan conduit stenosis
- mean PG 3-4mmHg
- venous collaterals from IVC to PVs

Single ventricle, heterotaxia syndrome, right isomerism, DORV, c-AVSD, TAPVR

s/p Pulsatile BCPS (1996.10.31)
s/p Extracardiac conduit Fontan procedure with
18mm Hemashield graft (1998.2.6)

*** Preop Dx ***
Same as the above

Clinical progressive cyanosis (SpO2 97% -> 88%)

Past Hx s/p Pulsatile BCPS (1996.10.31)
s/p Extracardiac conduit Fontan procedure
with 18mm Hemashield graft (1998.2.6)

Remark

Op. Name

Extracardiac Fontan conduit change with GoreTex graft 22mm

Op. finding

Severe adhesion around the previous conduit
Previous conduit : thin intimal peel(+), concentric narrowing at IVC-to-conduit junction
Could not find the IVC-to-PV collaterals
No intraoperative event

|012Y|F

199611549

2009-01-09

SEJONG HOSPITAL

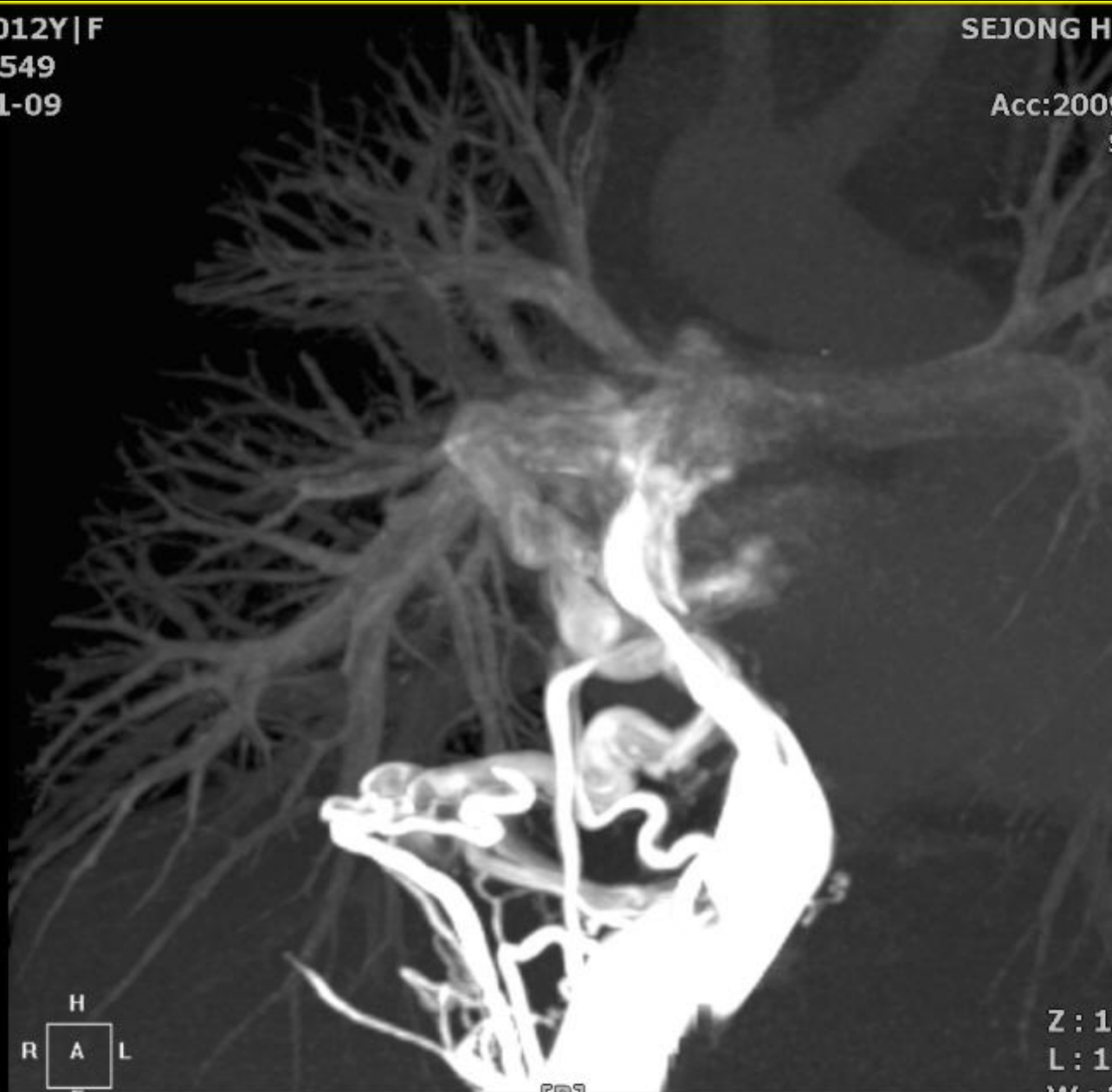
Rapidia

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Srs:1002

Img:307

PP:HFS



[L]

8cm



[P]

Z : 125.36%

L : 125

W : 259

신예찬 | 013Y | M
199622721
2010-02-19

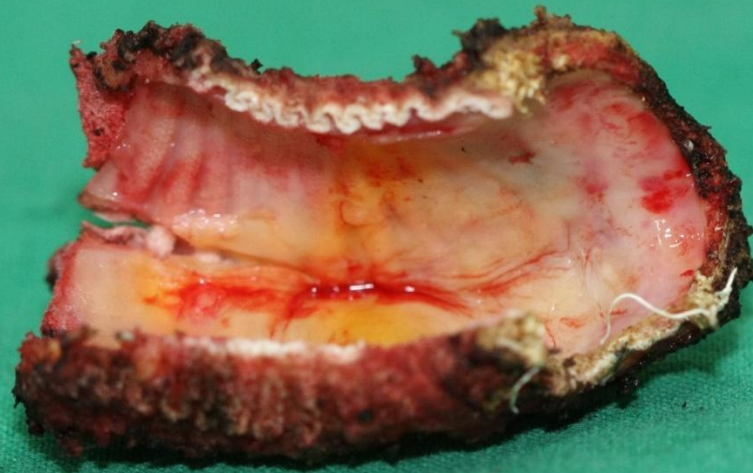
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Img:4

1

(1/96)

15 FPS Coronary
RAO 0.2
CAU 0.4

Z : 164.65%
L : 120
W : 228



Explanted Hemashield conduit

Midterm follow-up of the status of Gore-Tex graft after extracardiac conduit Fontan procedure[☆]

Cheul Lee^a, Chang-Ha Lee^{a,*}, Seong Wook Hwang^a, Hong Gook Lim^a,
Soo-Jin Kim^b, Jae Young Lee^b, Woo-Sup Shim^b, Woong-Han Kim^c

^aDepartment of Thoracic and Cardiovascular Surgery, Sejong Heart Institute, Sejong General Hospital, Bucheon, South Korea

^bPediatric Cardiology, Sejong Heart Institute, Sejong General Hospital, Bucheon, South Korea

^cDepartment of Thoracic and Cardiovascular Surgery, Clinical Research Institute, Seoul National University, College of Medicine, Seoul National University Children's Hospital, Seoul, South Korea

Received 13 November 2006; received in revised form 6 March 2007; accepted 9 March 2007; Available online 6 April 2007

Abstract

Objective: Extracardiac conduit Fontan procedure has some theoretical advantages over other types of Fontan procedures, such as optimized flow dynamics, a lower frequency of arrhythmias, and technical ease of procedure. However, lack of growth potential and thrombogenicity of the artificial conduit is the main concern and can possibly lead to reoperation for the conduit stenosis. In this study, we investigated the change and the status of the Gore-Tex graft used in extracardiac conduit Fontan procedure. **Methods:** Between 1996 and 2005, 154 patients underwent extracardiac conduit Fontan procedure using Gore-Tex graft. Among these, 46 patients underwent cardiac catheterization during follow-up period. We measured the internal diameter of the conduit and inferior vena cava angiographically. **Results:** Mean follow-up duration was 36.1 ± 19.7 months. The conduit diameter used was 16 mm in 10 patients, 18 mm in 16, 20 mm in 14, 22 mm in 4, and 24 mm in 2 patients. The mean conduit-to-inferior vena cava cross-sectional area ratio was 1.25 ± 0.33 . According to the conduit size used, this ratio was 1.03 ± 0.17 for 16 mm conduits, 1.33 ± 0.37 for 18 mm, 1.33 ± 0.36 for 20 mm, 1.28 ± 0.26 for 22 mm, and 1.05 ± 0.06 for 24 mm conduits ($p < 0.05$, 16 mm vs 18 mm and 20 mm). The mean percent decrease of the conduit cross-sectional area was $14.3 \pm 8.5\%$, and this did not differ significantly according to the conduit size ($p = 0.82$). Follow-up duration and the percent decrease of the conduit cross-sectional area did not show significant correlation ($r = 0.22$, $p = 0.14$). There was no reoperation due to conduit stenosis. **Conclusions:** During midterm follow-up of about 3 years, the conduit cross-sectional area decreased by 14%, and this did not differ according to the conduit size used. The extent of decrease of the conduit cross-sectional area remained stable irrespective of the follow-up duration. Sixteen millimeters conduit showed no evidence of clinically significant stenosis, but careful follow-up is warranted because of the possible conduit stenosis relative to the patients' somatic growth.

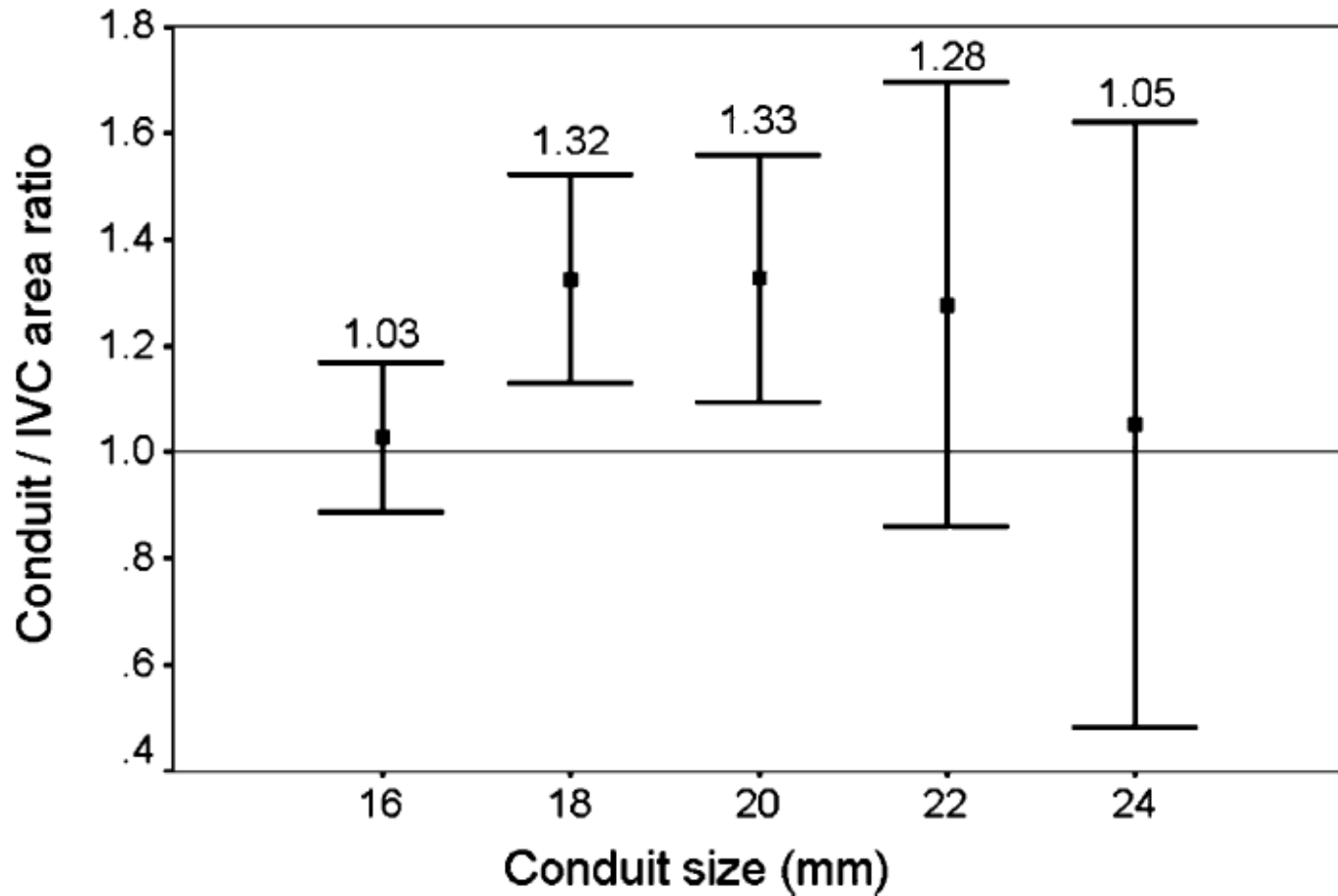


Fig. 5. Conduit-to-IVC cross-sectional area ratio according to conduit size. Filled squares denote mean values (numbers above error bars) and error bars represent 95% confidence interval for means.



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Mid-term follow-up of the status of Gore-Tex graft after extracardiac conduit Fontan procedure[☆]

Yoshie Ochiai^{a,*}, Yutaka Imoto^a, Masato Sakamoto^a, Takashi Kajiwara^a,
Akira Sese^a, Mamie Watanabe^b, Takuro Ohno^b, Kunitaka Joo^b

^a Department of Cardiovascular Surgery, Kyushu Koseinenkin Hospital, Kitakyushu, Japan

^b Department of Pediatric Cardiology, Kyushu Koseinenkin Hospital, Kitakyushu, Japan

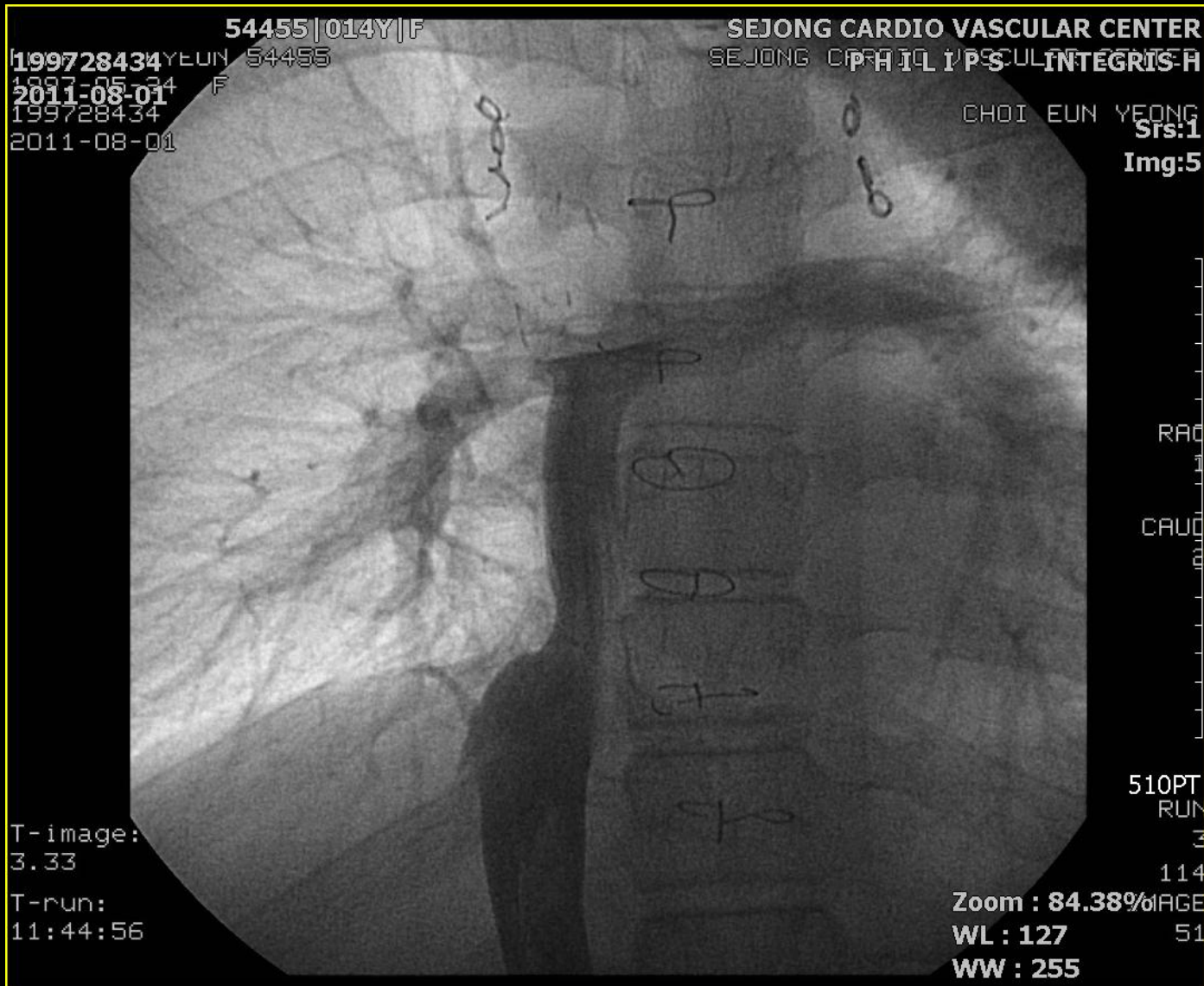
Received 3 September 2008; received in revised form 6 February 2009; accepted 9 February 2009; Available online 28 March 2009

Abstract

Objective: Extracardiac conduit Fontan procedure (ECFP) using Gore-Tex graft has been performed with increasing frequency for the patients with functional single ventricle. However, lack of growth potential and longevity of the conduit are consistent concerns and main points of criticism of the ECFP. In this study, we investigated the mid-term status of the Gore-Tex graft used in the ECFP by comparing the internal diameter of the graft with the inferior vena cava (IVC) diameter at 1 month and 5.2 years after the ECFP. **Methods:** Of 79 patients who underwent ECFP using Gore-Tex graft between November 1997 and December 2007, 33 patients who had completed cardiac catheterization at 1 month (21–73 days) and 5.2 years (3.3–9.6 years) after the ECFP were included in this study. We measured the internal diameter of the Gore-Tex graft and IVC at both catheterizations retrospectively. **Results:** The size of the Gore-Tex graft used in the ECFP was 16 mm in 17 patients, 18 mm in 9 patients, and 20 mm in 7 patients. Laminar flow through the conduits was maintained without any stenosis or kinking of the graft in these 33 patients. No intervention or reoperation related to the extracardiac conduit has been required. There were no significant differences in mean cross-sectional area (CSA) of the conduits at 1 month versus 5.2 years after the ECFP for each conduit size, and no significant changes in the conduit-to-IVC CSA ratio (0.98 ± 0.40 vs 0.82 ± 0.21 for 16 mm, 1.09 ± 0.30 vs 0.92 ± 0.33 for 18 mm, and 1.16 ± 0.55 vs 0.94 ± 0.44 for 20 mm conduit). **Conclusions:** The conduit CSA and conduit-to-IVC CSA ratio remained unchanged in small caliber grafts down to 16 mm at 5.2 years after the ECFP. However, further investigation is necessary to evaluate the fate of the Gore-Tex graft and late hemodynamics in the patients with small conduits after they achieve full somatic growth.

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s/p Extracardiac Fontan with Gore-Tex graft 16mm



s/p Extracardiac Fontan with Gore-Tex graft 16mm

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2011-08-01

SEJONG CARDIO VASCULAR CENTER
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Srs:1
Img:3



Conclusions

- **Reoperations following the Fontan procedure may be necessary for various indications which result in impairment of the Fontan circulation.**
- **Every efforts should be made to prevent the late reoperations during the course of single ventricle palliation toward Fontan completion.**

Francis M. Fontan

3rd Scientific Meeting, World Society for Pediatric and Congenital Heart Surgery
June 2011, Istanbul

