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Medical and Interventional Management of Failing Fontan Circulation

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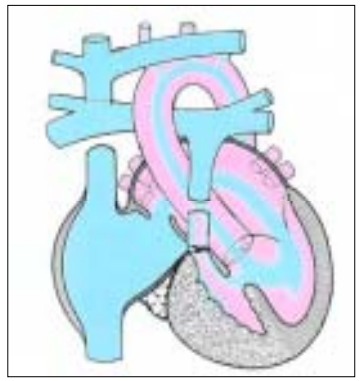
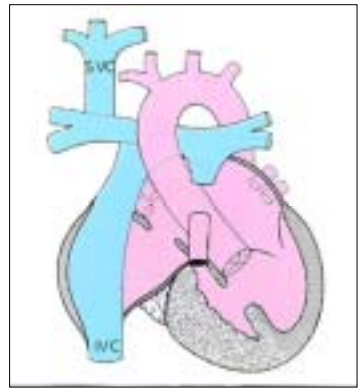
김 영 휘

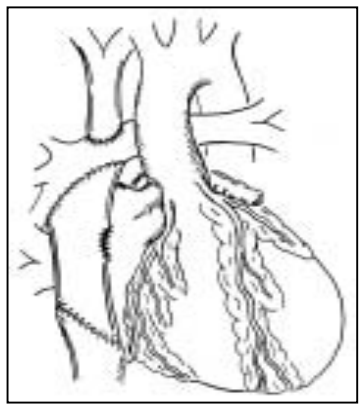
Modification of Classic Fontan

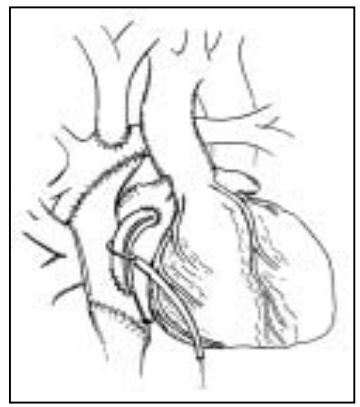
- Staged Fontan: BCS
- Baffle fenestration
- Lateral tunnel operation
- Extracardiac conduit Fontan

⇒ ↓early mortality & morbidity
↑long-term survival









Failing Fontan(?)

- Acute failure

  LCO ± postop Cx

- Late failure

  progressive exercise intolerance

  congestive heart failure

± Late sequale

- Arrhythmias
- Cyanosis
- PLE/ Plastic brochitis
- AVV regurgitation
- Thromboembolism
- Prolonged pleural/pericardial effusion

Acute Failure of Fontan Procedure

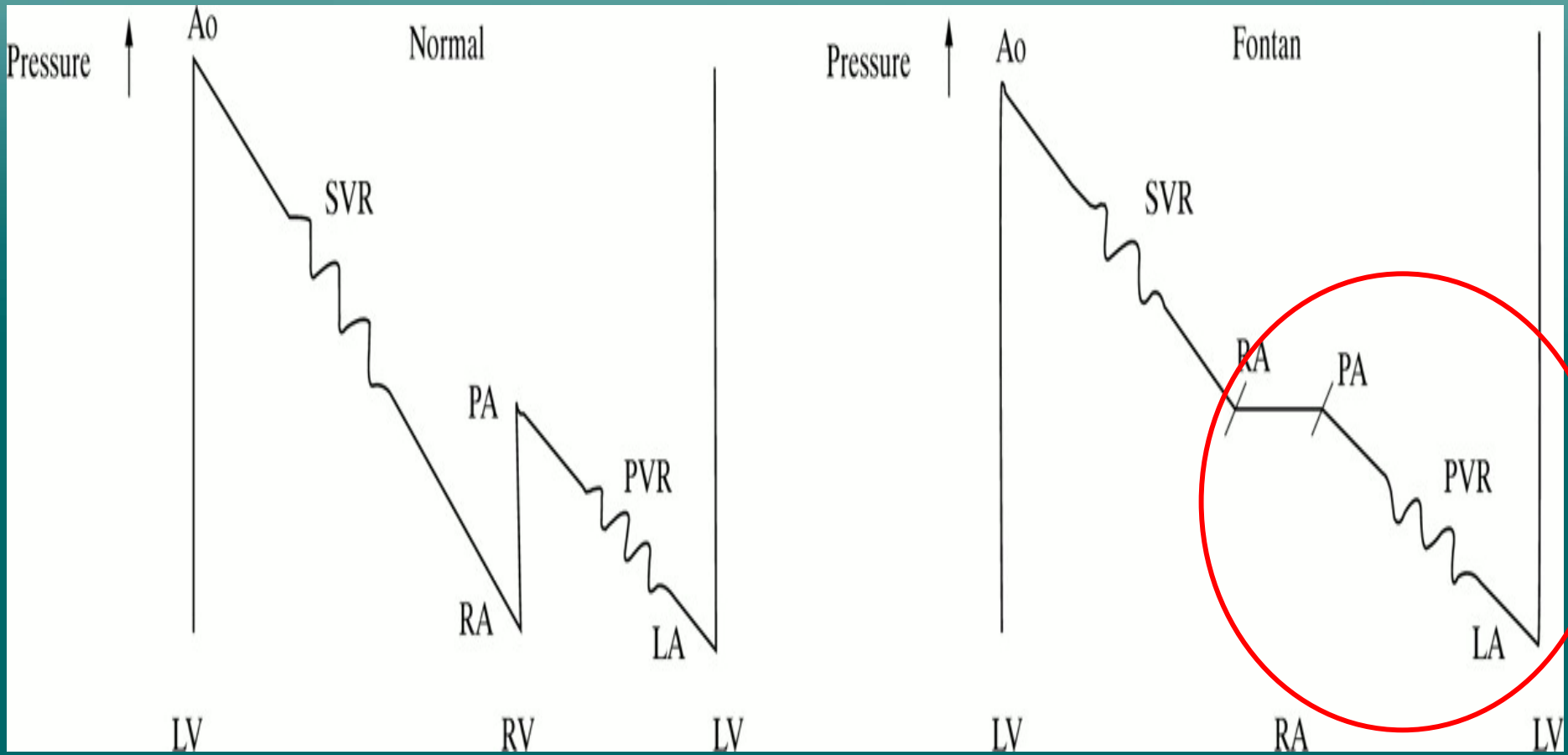
- Low CO
- Cyanosis
- Thrombosis
- Effusion
- Arrhythmias

Factors for progressive deterioration of functional status

- Arrhythmias
- Ventricular dysfunction
- AV valve regurgitation
- Cyanosis
- Fontan pathway obstruction

Prolonged pleural/pericardial effusion
Thromboembolic events
Protein losing enteropathy(PLE)

Pulmonary Circulation



Low Cardiac Output

CVP	LAP	Causes
low	Low	Hypovolemia
high	Low	▪ Fontan pathway obstruction, ▪ ↑PVR - pulmonary vascular disease - PA spasm ▪ Pulmonary artery hypoplasia Branch PA stenosis
high	high	▪ Ventricular dysfunction ▪ AV valve stenosis or insufficiency ▪ Arrhythmia ▪ Tamponade ▪ Outflow obstruction

Prevention of Acute Failure

- Preop. evaluation & correction
  - Aortopulmonary collaterals
  - AV valve regurgitation
  - residual subaortic stenosis
  - residual CoA or PA branch stenosis
- Ultrafiltration
- Fenestration in high risk group
  adjustable or fixed

Management of LCO

- Low CVP → Volume challenge
- High CVP & Low LAP

Fontan obstruction
Branch PA stenosis → Operation

PA spasm → Lowering PVR → NO inhaltion, Prostacyclin, Beraprost, Sidenafil

Pulmonary vascular disease
pulmonary arteial hypoplasia → Fenestration, ECMO, Takedown/HT

Management of LCO

High LAP(>10mmHg) with low TPG

- Arrhythmia
- Tamponade

→ Tx

- AV valve stenosis or insufficiency
- Outflow obstruction

→ Op

- Ventricular dysfunction

→ Inotropic support

VAD / ECMO?

Transplantation

Extracorporeal Membrane Oxygenation Support of the Fontan and Bidirection Glenn Circulation

Booth KL, Roth SJ, Thiagarajan RR, Almodovar MC, del Nido PJ, Laussen PC
Department of Pediatrics, Surgery, and Anesthesia, Children's Hospital Boston, Harvard Medical School, Boston, Massachusetts

Patients: Dec. 1984- June, 2004
20 with cavopulm. Connection
(14: Fontan, 6: BDG)
median age: 4.4 yr in Fontan(1-42 yr)
17.1 mo in BDG(4.7-26.9 mo)
median Wt : 14.5 kg(7.9-82 kg) in Fontan
8.4 kg(4.4-10.5 kg) in BDG

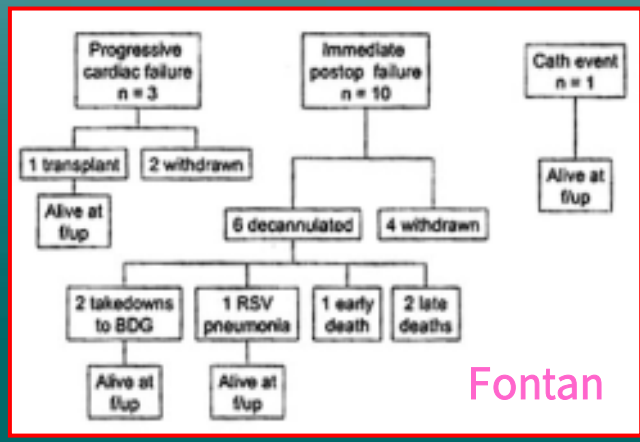
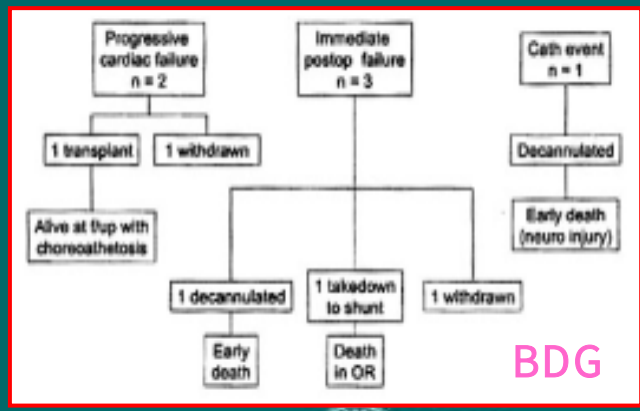


Table 1. Published Literature of Fontan and BDG Patients Supported With ECMO

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.



Ann Thorac Surg 2004;77:1341-8

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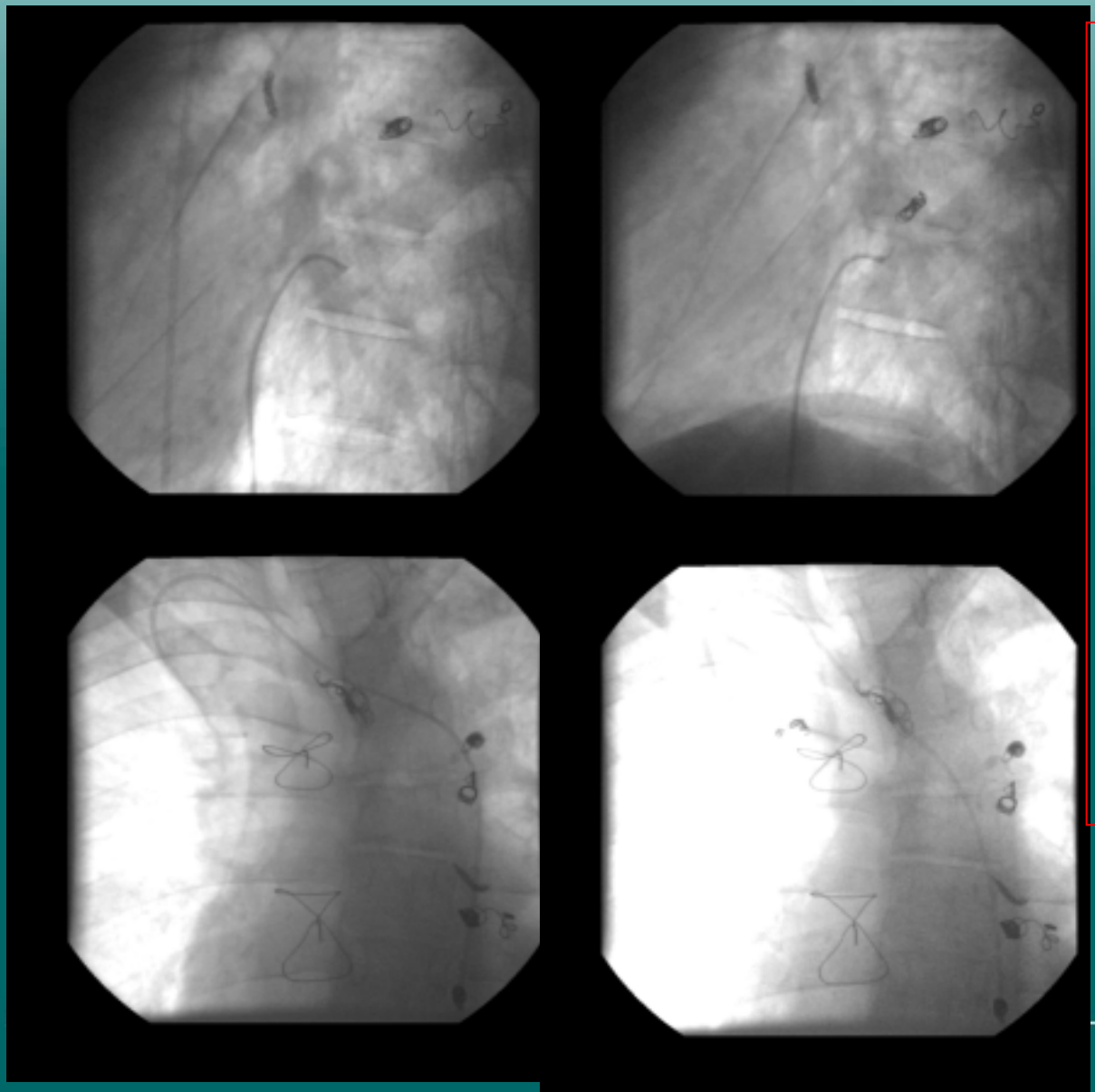
Aortopulmonary collaterals

- Ventricular volume loading
  Increase CVP
  → prolonged effusion
- Catheter intervention coil:
  Giantruco coil
  Tornado coil
  (microinfusion cath)

Coil embolization case



Case: 조 ㅇ ㅇ
Dx : T. atresia I a
1st op : both m-BT, BAS
central sh
2nd op : BCS at 9yr
3rd op : APC with
fenestration d/t small PA
LV function↓
2002.1: massive hemoptysis
cath: SaO2 88%
PAP:m=5, LVEDP 9
multiple collateral
→ coil
2002. 7: hemoptysis recur
→ coil

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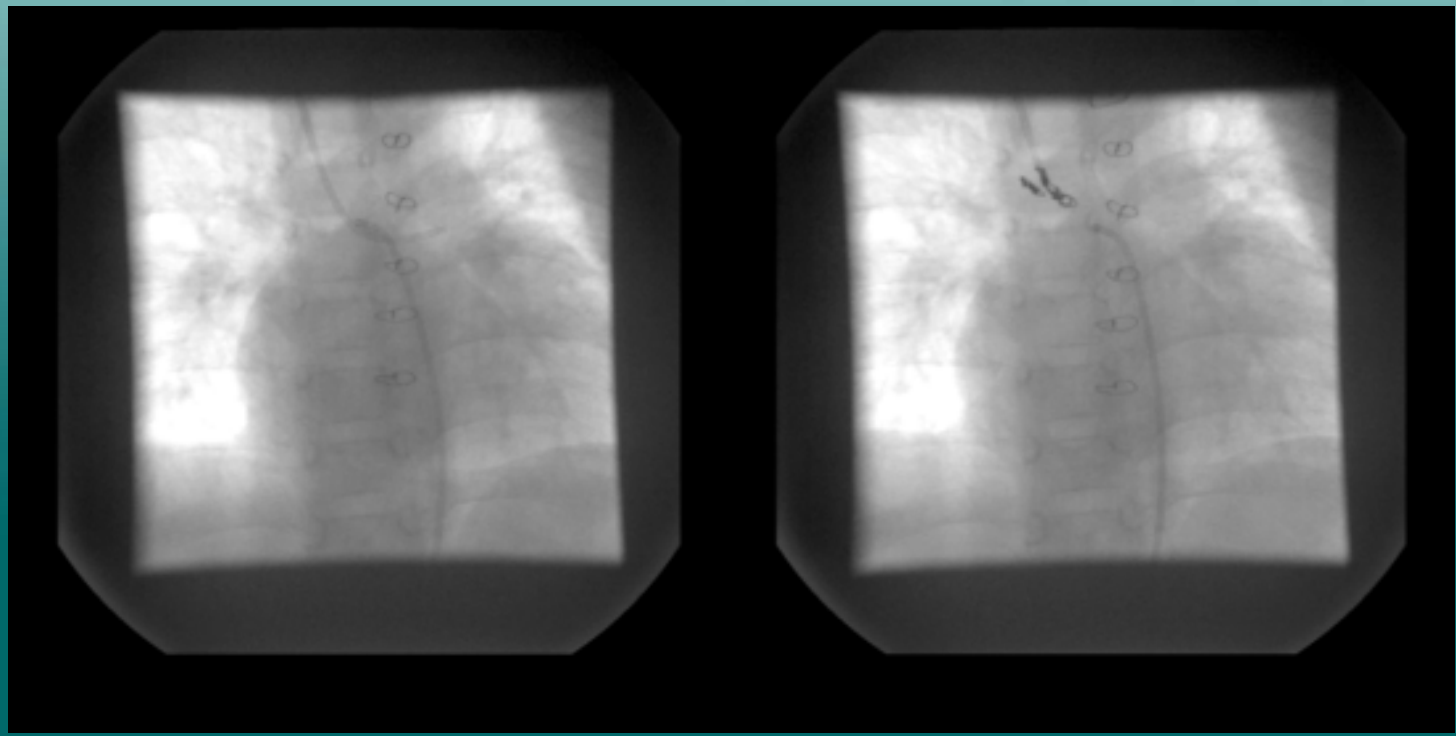
Coil embolization case



Case: 이 O O, 2yr/M
DX: DILV, cc-TGA, VSD, mild PS
1st OP: Atrial septectomy, PAB at 8 mo of age
2nd OP: BCS, BVF widening at 19 mo of age
Cath(04, 9,16): PAP m=8, VEDP 10, collaterals(+)

Catheter intervention for Fenestration

- Fontan without fenestration
  Transseptal puncture
  - Transseptal needle
  - RF perforation catheter(5J)
- Fontan with ↓ fenestration

Balloon dilatation
Stent implantation

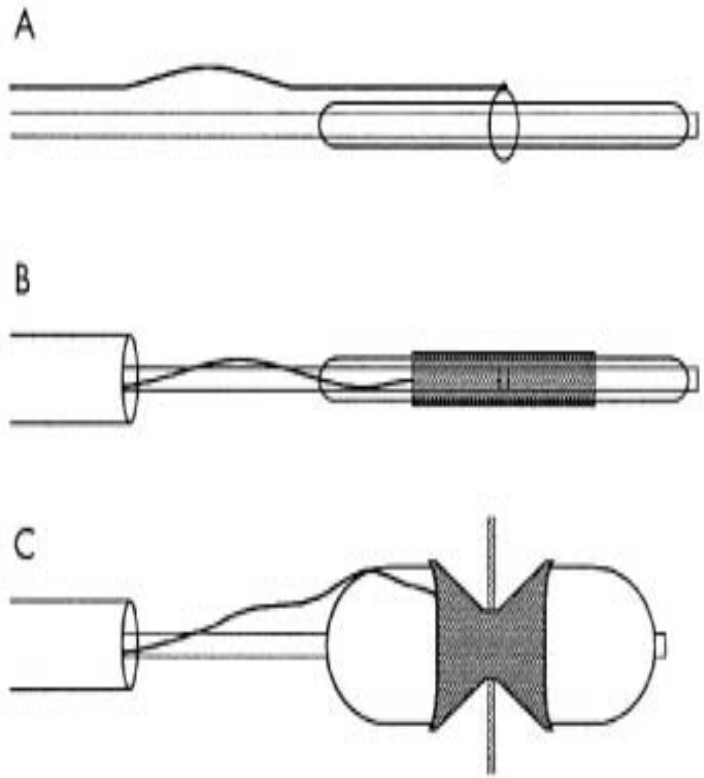
Fenestration size ?

- Balloon : oversized→ cyansis↑
- Stent : fixed-size (3-4 mm ?)
- Fenestrated Amplatzer septal Occludor

Stent Fenestration

Technique

- 3-4 mm loop with epicardial pacing wires
- distal 5 cm of two wire tie→ 90cm length
- resultant loop placed over mid-portion of balloon catheter(15-25 mm): *A*
- sl. dilate stent with the help of 10-14 Fr dilator
- Bare stent mounted on the balloon: *B*
- 11-14 Fr long Mullins sheath across the atrial septum
- stent-mounted balloon with radio-opaque wire loop centered at atrial septum
- Delivery sheath withdrawn to IVC and balloon was fully inflated : *C*



- **Goal for failing Fontan**

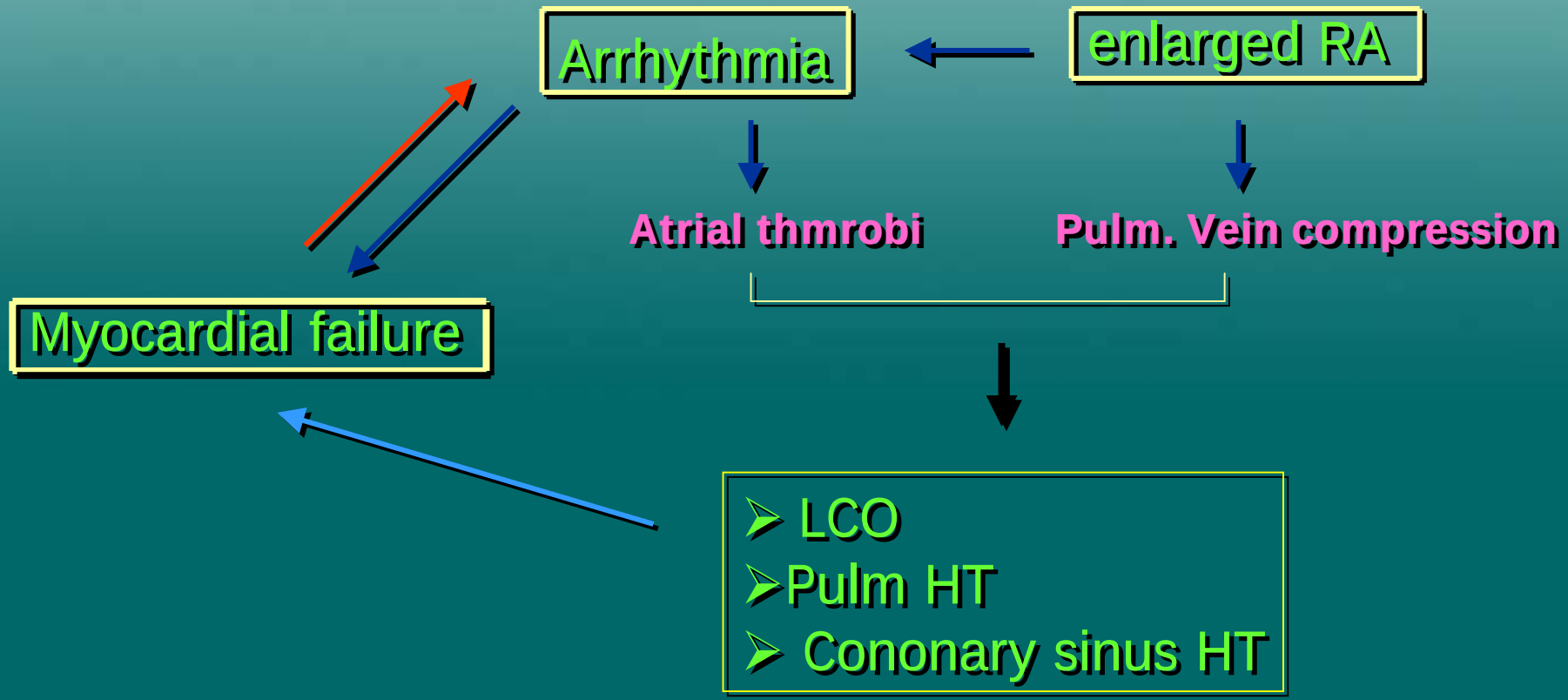
  SaO2 80-85% at room air

Arrhythmia after Fontan Op

- 40% of Fontan pts at 10 yr FU
- Most common : APC
- Atrial tachyarrhythmia(IART)
- SSS
- Arrhythmia substrate

persisting preop. arrhythmia
scar/patch/fibrotic area by atrial dilation
natural barrier : AV valve, IVC, Pulm vein
hemodynamic theory(?)

Interaction of arrhythmia to failing Fontan



Management of failing Fontan with Atrial tachyarrhythmia

- Extracardac conduit Fontan approach
  - Exclude atrium from systemic vein circuit
  - Avoid additional atrial suture line
  - Extensive atrioplasty with removal arrhythmogenic atrial tissue
- Intraoperative cryoablation
  Intraoperative RF ablation
- Fontan conversion, heart translantatin
- Anti-tachycardial pacemaker insertion

Extracardiac Conduit With a Limited Maze Procedure for the Failing Fontan With Atrial Tachycardias

Shaun P, Setty, MD, Kirsten Finucane, FRACS, Jonathan R, Skinner, MD, FRCPCH, and Alan R, Kerr, FRACS

Table 1. Patient Information and Outcome

Patient No.	Diagnosis	Type of First Fontan	Age at First Fontan (y)	Age at Conversion (y)	Interval (y)	Indication
1 (previously published)	Tricuspid atresia, ASD, VSD	Atriopulmonary anastomosis	7.2	28.2	21.1	Fatigue (NYHA 3) IART Homograft narrowing
2	Tricuspid atresia, d-TGA	Atriopulmonary anastomosis	4	13.7	9.7	IART Fatigue (NYHA 2)
3	Tricuspid atresia, restrictive VSD	Bjork modification	1.6	17	15.4	Atrial fibrillation with giant right atrium Decreased exercise tolerance (NYHA 3)
4	DILV, l-TGA, hypoplastic right ventricle	Atriopulmonary anastomosis	15	27	12	Atrial fibrillation Second-degree heart block caused by medication Fatigue (NYHA 2)
5 (previously published)	TGA, VSD, hypoplastic right ventricle	Atriopulmonary anastomosis	5.5	17.1	11.6	IART Giant right atrium with thrombus Fatigue (NYHA 3)
6	Tricuspid atresia, VSD	Bjork modification	14	34	20	Atrial fibrillation and IART Pre-syncope Fatigue (NYHA 3)

Table 1. Continued

Operative Procedure*	Pacer Mode	Preoperative Antiarrhythmic Medication (Previous Medications)	Current Antiarrhythmic Medication	Outcome
Bilateral Bidirectional Glenn	AAIR	Flecainide Digoxin (Amiodarone—stopped because of pulmonary toxicity)	Digoxin	Postoperative atrial tachyarrhythmia controlled with sotalol. Pacer generator placed 26 days after conversion with epicardial leads in place. Doing well (NYHA 2).
	AAIR	Sotalol Digoxin	Digoxin	Doing well, no arrhythmias (NYHA 1).
	AAI	Digoxin Diltiazem (Flecainide)	None	Doing well, no arrhythmias (NYHA 1).
Resection of subaortic stenosis	DDIR	Digoxin (rate-controlled atrial fibrillation)	Digoxin Sotalol	Surgically induced heart block Postoperative atrial flutter treated with sotalol, doing well (NYHA 1).
	AAIR	Digoxin (Amiodarone)	None	Doing well, no arrhythmias (NYHA 1).
Bilateral bidirectional Glenn	AAIR	Digoxin Diltiazem (Flecainide Sotalol)	Sotalol Digoxin	Atrial flutter day 5 postoperatively, none since sotalol. Improved exercise tolerance (NYHA 2).

* In addition to 22-mm or 24-mm polytetrafluoroethylene extracardiac conduit, right atrial reduction, limited right atrial maze procedure, and pacemaker placement.

AAI = demand atrial pacing; AAIR = rate responsive atrial pacing; ASD = atrial septal defect; DDIR = dual chamber rate responsive pacing; DILV = double-inlet left ventricle; d-TGA = d-transposition of great arteries; IART = intraatrial reentrant tachycardia; l-TGA = l-transposition of great arteries; NYHA = New York Heart Association; TGA = transposition of great arteries; VSD = ventricular septal defect.

Ann Thorac Surg 2002;74:1992-7

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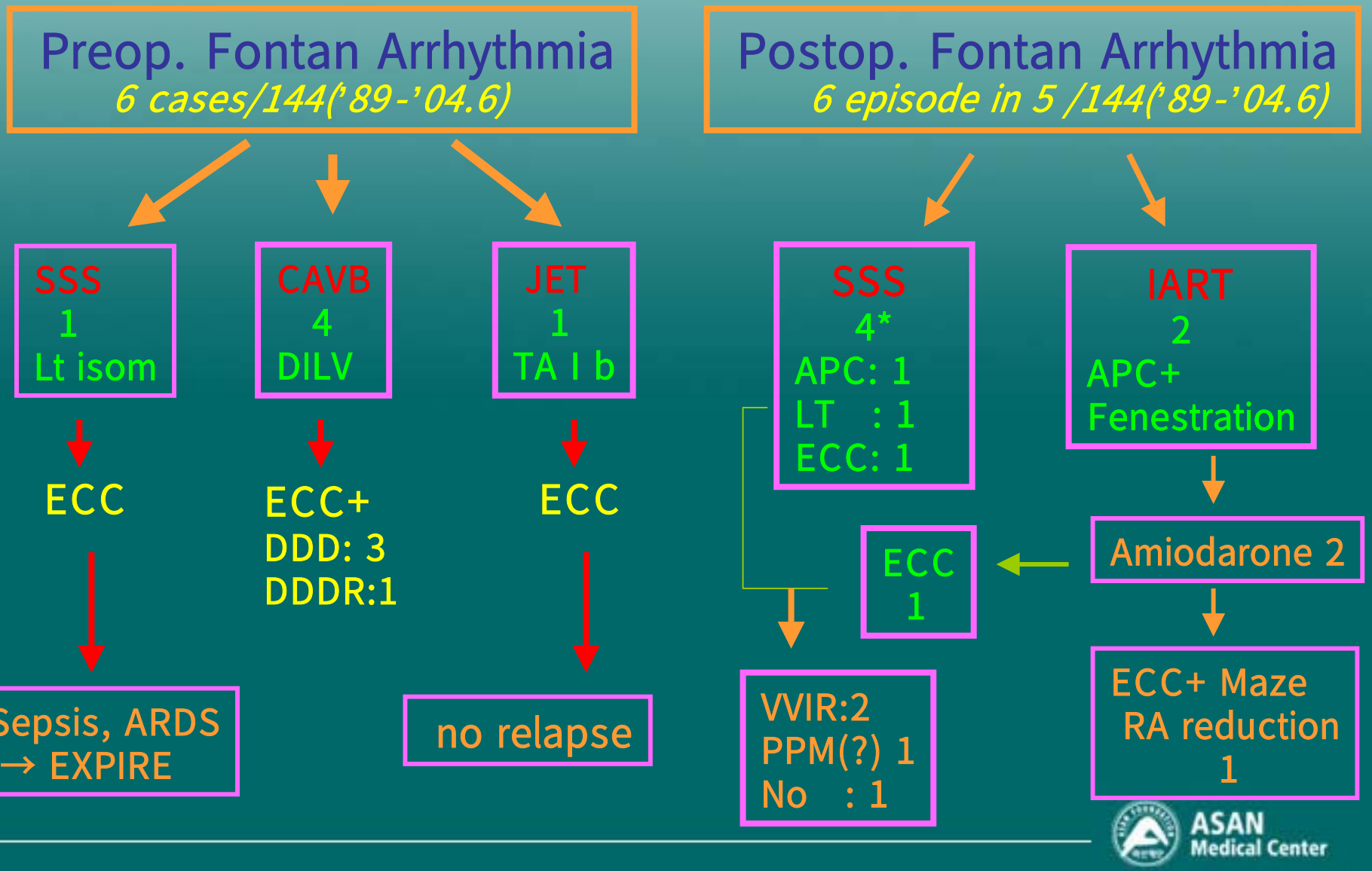
Arrhythmia of Fontan- Patients in AMC



Cyanosis after Fontan Procedure

Pulmonary AV marfomation → Coil occlusion
Surgical resection

Contrast ECHO
Pulm. Angio

Systemic-pulmo.
Veno-venous coll. → Coil occlusion

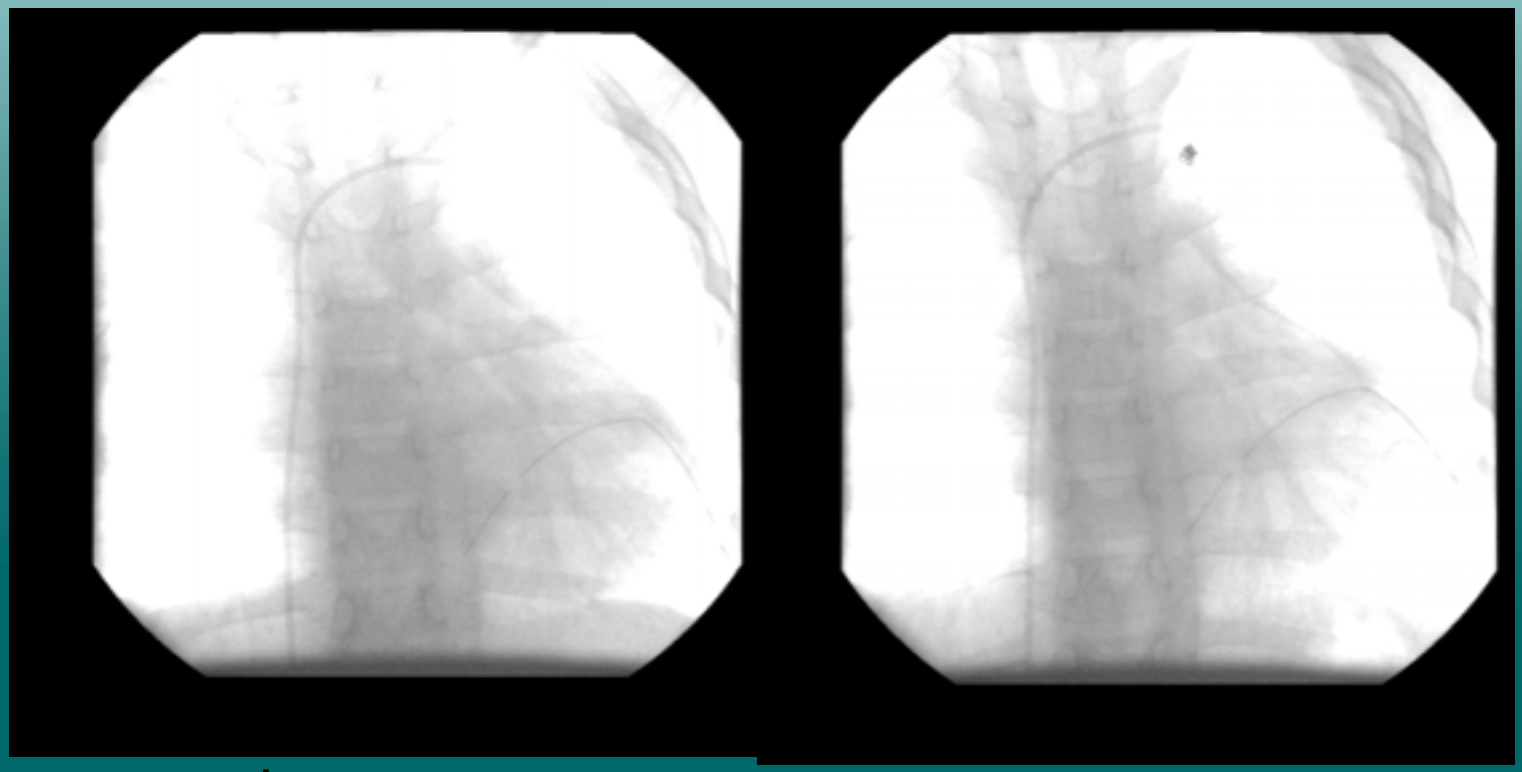
Excessive fenestration

Test occlusion

CVP ≤ 15 mmHG → Device closure

Ventricular dysfunction → Heart Transplantation

systemic-pulmonary venovenous collateral closure



Case: 김 O O, 5yr/M

Dx: cc-TGA, VSD, MV straddling, No PS

1st OP: PAB at 5 mo of age

2nd OP: EC conduit Fontan(18 mm GTVG) at 5 yrs old prolonged pleural effusion for postop 2 mo

→ Cath: SaO2 93%, PAP m=10, VEDP 5

Systemic –pulm. Venovenos collateral(+)

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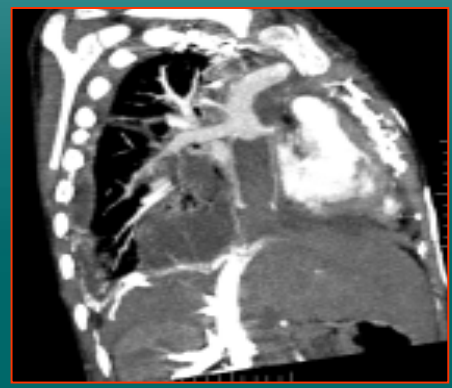
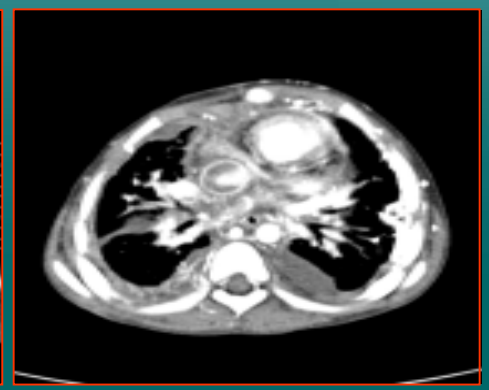
Thromboembolism after Fontan procedure

- True incidence unknown
  → underestimated
- Stroke incidence: 2.6%

Diagnostic tool

- TEE(TTE less sensitive)
- Heart CT/MR
- Cath. Angio





Sx of thrombus formation

- Venous obstruction
- Progressive cyanosis
- paradoxical emboli
- Atrical arrhythmia
- Asymptomatic(40%)

Predisposing Risk Factors

- Slow nonpulsatile flow
- Enlarged thickened atrium
- small TPG, LCO
- Abnormality of clotting system
  (protein C/S deficiency,ATIII↓, VWF↑)

Longterm anticoagulation: controversal

- **good Fontan : no anticoagulation** vs
  all Fontan : antiplatelet/oral warfarin
- **risk of hemorrhagic complication**
- **Wafarin (INR 2-2.5)**

Protein Losing Enteropathy after Fontan OP

- 5-10% of Fontan operation
- Inflammatory cause
  Mechanical cause

Hemodynamic alternation

- ↑SVC pressure
- severe turbulant flow in RPA near Rt BCS

Clinical feature

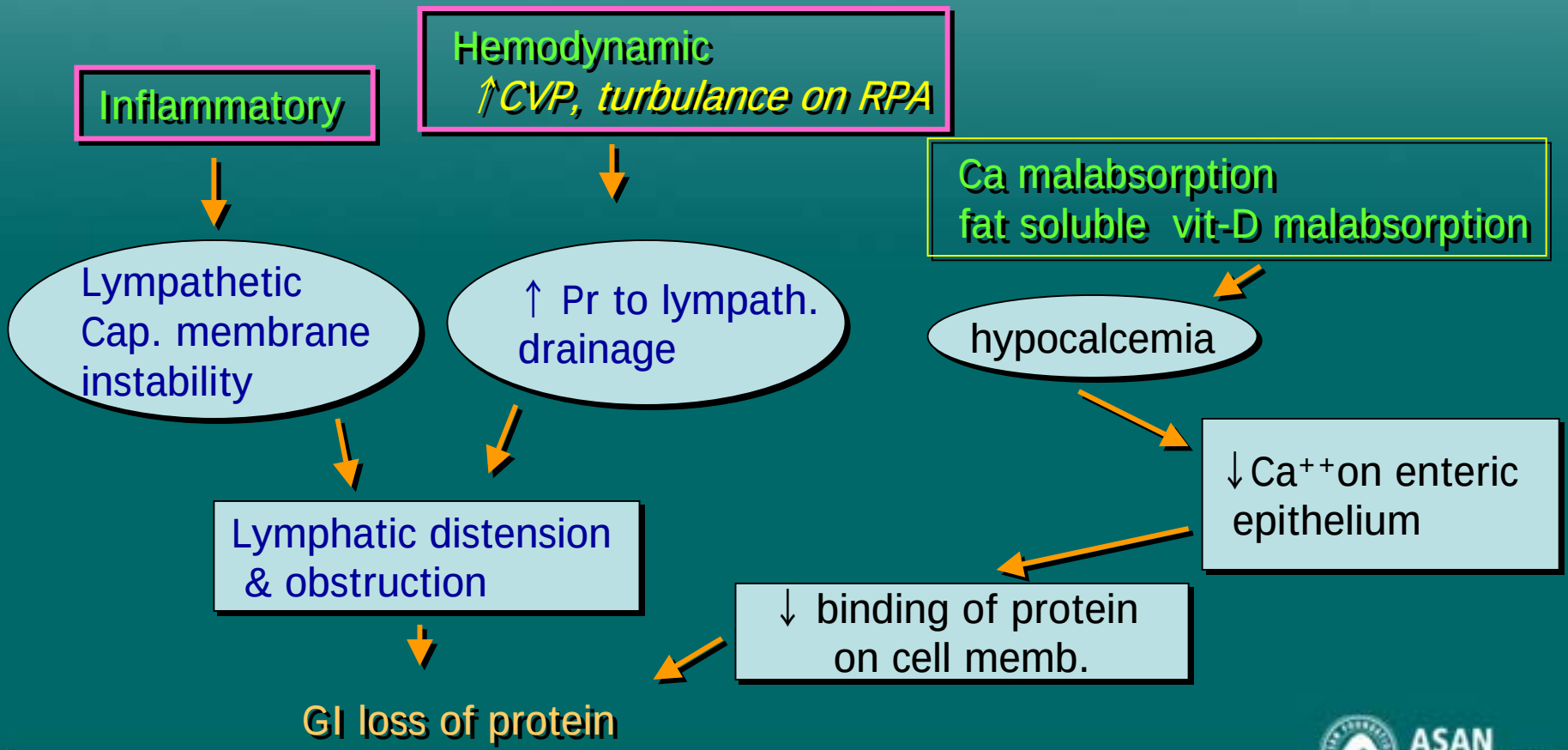
- Hypoproteinemia
- Ascites, edema
- Diarrhea(α-1 antitrypsin↑in stool)
- Immunodeficiency
- Hypocalcemia

Risk Factors

- ↑ Rp ≥ 4 unit
- Heterotaxic synd(polysplenia)
- Ventricular dysfunction (VEDP ≥ 12 mmHg)

Protein Losing Enteropathy after Fontan OP

Pathophysiology



Protein Losing Enteropathy after Fontan OP

management

- **Medical**
  - dietary manipulation/ albumin infusion
  - correction of hypocalcemia
  - diuretics/ high dose steroid
  - systemic administration of heparin → *osteoporosis*↑
  - High-dose aldosterone receptor antigonist
- **Intervention for ↓ CVP**
  - balloon dilation of narrowed PA
  - occlusion of aortopulmo. collateral arteries
  - creation of fenestration
- **Takedown and /or transplantation**

Effect of High-Dose spironolactone on Proein-Losing Enteropathy in Patients with Fopntan Palliation of Complex Congenital Heart Disease

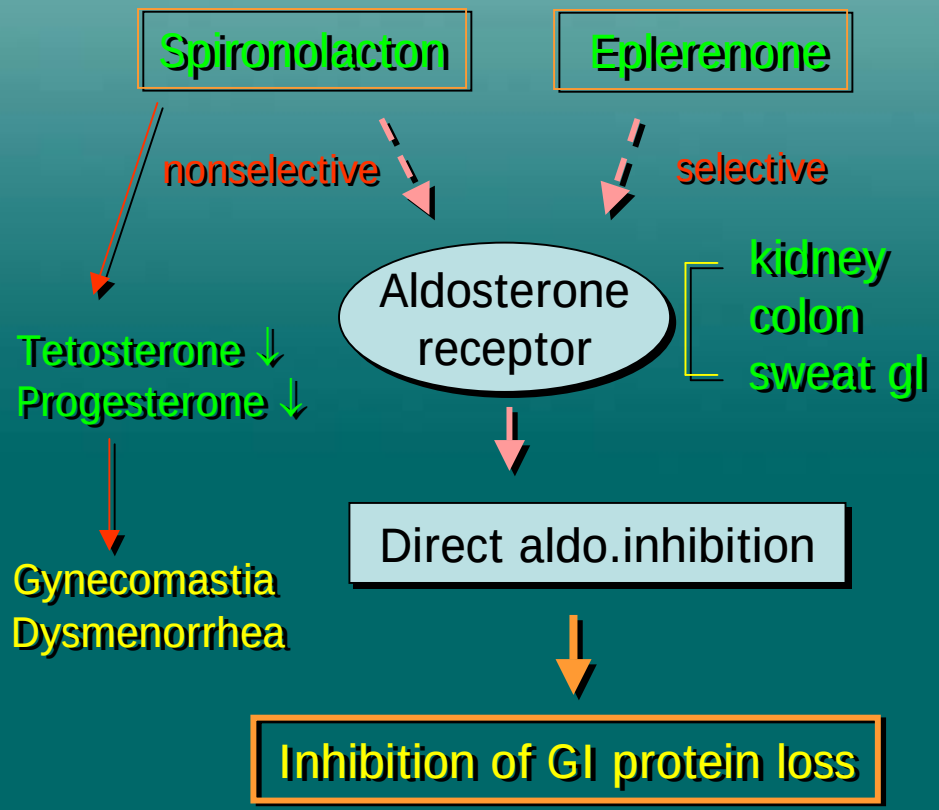


TABLE 1 Patient Summaries and Medical Management of Protein-losing Enteropathy (PLE)

	Patient No.		
	1	2	3
Diagnosis	HLHS	DILV	HLHS
Surgical palliation (age)	Fontan-intracardiac conduit (1 yr)	Fontan-extracardiac conduit (6 yrs)	Fontan-lateral tunnel (2 yrs)
Time interval from Fontan repair to PLE onset	7 yrs	1 yr	10 yrs
Mean pulmonary artery pressure	13 mm Hg	12 mm Hg	13 mm Hg
Presenting total protein and albumin levels	3.8/1.7 g/dl	4.2/1.9 g/dl	4.1/2.4 g/dl
Initial medical therapy	Spironolactone (4 mg/kg/d), ACE inhibitor	Furosemide	ACE inhibitor
Subsequent total protein and albumin levels	5.7/3.5 g/dl	4.2/2.7 g/dl	4.1/2.8 g/dl
Interval medical therapy I	Spironolactone (4 mg/kg/d), ACE inhibitor	ACE inhibitor, furosemide	Spironolactone (1.4 mg/kg/d), ACE inhibitor
Follow-up total protein and albumin levels	3.1/1.8 g/dl	3.9/2.2 g/dl	4.1/2.8 g/dl
Interval medical therapy II	Spironolactone (5 mg/kg/d), ACE inhibitor	Spironolactone (2.3 mg/kg/d), ACE inhibitor, furosemide	Spironolactone (2.8 mg/kg/d), ACE inhibitor
Remission total protein and albumin levels	5.6/3.6 g/dl	5.8/3.5 g/dl	6.5/4.0 g/dl
Duration of remission	2 yrs	3 yrs	2 yrs

ACE = angiotensin-converting enzyme; DILV = double-inlet left ventricle; HLHS = hypoplastic left heart syndrome.

Ringel et al. Am J Cardiol 2003;91:1031-2

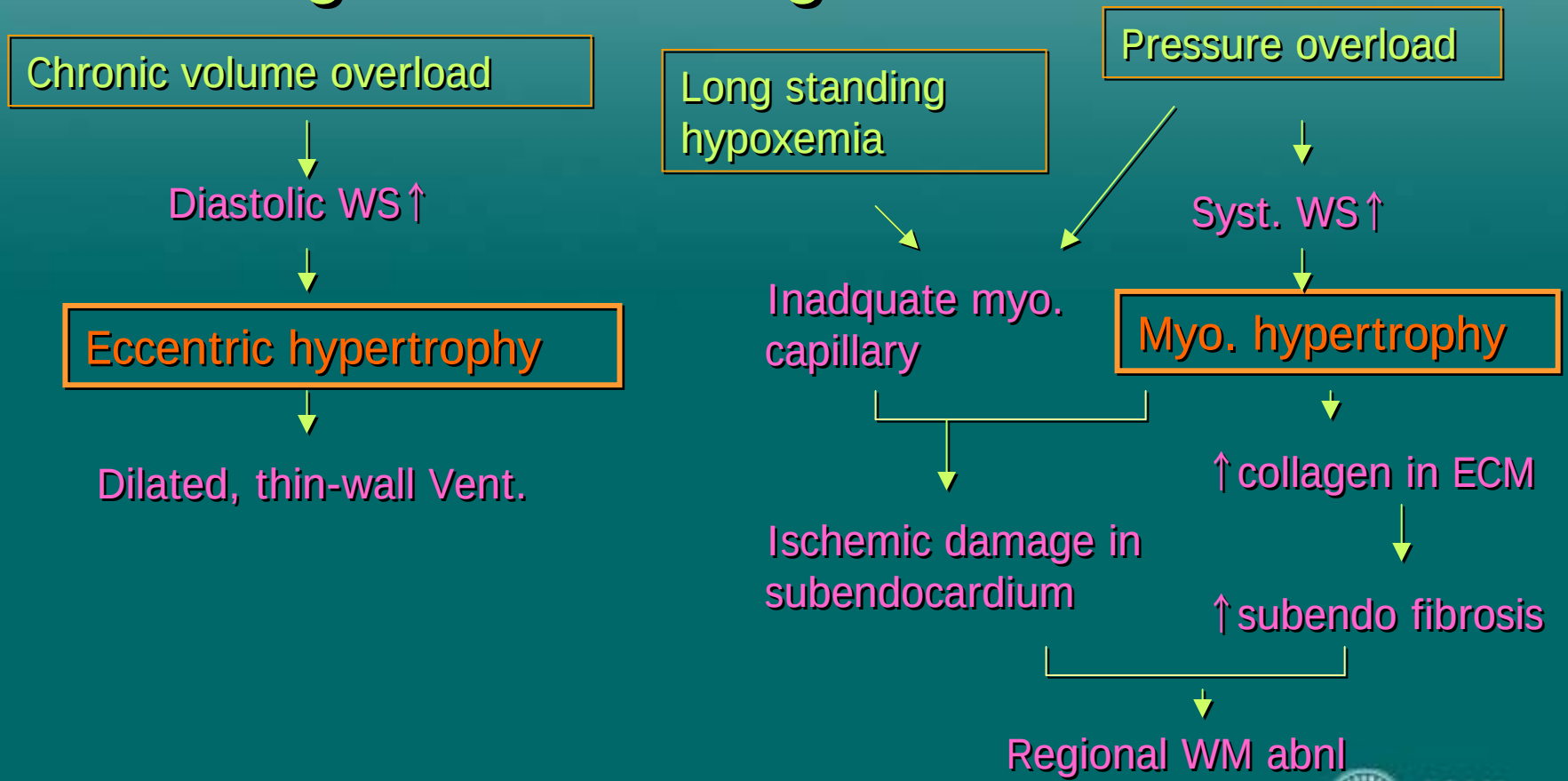
Ventricular dysfunction after Fontan OP

contributing factors

- Increased ventricular mass
- Morphological type of ventricle
- Impaired coronary artery flow

Ventricular remodeling after Fontan OP

Pathologic remodeling



Left ventricular remodeling in hearts with tricuspid atresia : Morphologic observations and possible basis for ventricular dysfunction after surgery

Maria Angelica Binotto, MD, Maria de Lourdes Higuchi, MD, Vera Demarchi Aiello, MD

Method

32 heart with T. atresia at autopsy
27 control heart
die of non-cardiovascular related Ds

Morphometric analysis

- Myocyte transverse ϕ
- capillary volume fraction
- nuclear volume fraction
- quantification of fibrous tissue
- endocardial thickness

TABLE 2. Gross measurements from the inlet and outlet of the LV, comparing hearts with tricuspid atresia and control hearts

	Tricuspid atresia		Control		
	Mean ± SD	Median	Mean ± SD	Median	*P* value
Inlet wall thickness (cm)	0.68 ± 0.17	0.70	0.69 ± 0.22	0.70	.82
Outlet wall thickness (cm)	0.70 ± 0.21	0.70	0.70 ± 0.22	0.60	.94
Inlet length (cm)	3.19 ± 0.76	3.10	2.97 ± 0.63	2.90	.24
Outlet length (cm)	3.69 ± 0.84	3.55	3.05 ± 0.63	2.95	.005

TABLE 3. Myocyte diameter, interstitial fibrosis and endocardial thickness in the inlet, apex, and outlet of the LV in control hearts and those with tricuspid atresia

	Myocyte diameter (μm)		Fibrosis (IFVF %)		Endocardial thickness (μm)	
	Mean ± SD	Median	Mean ± SD	Median	Mean ± SD	Median
Inlet						
Tricuspid atresia	9.6 ± 2.4	9.6 (25)	3.8 ± 2.8	3.1 (29)†	53.9 ± 40.1	41.8 (29)*
Control	10.0 ± 2.6	9.8 (15)	1.0 ± 0.7	1.0 (23)	24.0 ± 19.7	14.3 (18)
Apex						
Tricuspid atresia	9.6 ± 2.1	10.6 (15)	2.6 ± 2.0	2.0 (25)†	20.8 ± 14.7	15.1 (28)*
Control	9.0 ± 2.1	8.7 (14)	0.5 ± 0.5	0.4 (19)	10.7 ± 3.7	9.9 (19)
Outlet						
Tricuspid atresia	8.9 ± 1.9	9.0 (14)	2.1 ± 1.8	1.6 (22)†	34.1 ± 29.0	23.9 (22)
Control	9.0 ± 1.8	9.4 (14)	0.6 ± 0.5	0.4 (18)	19.0 ± 13.2	14.8 (16)

In parentheses are shown the numbers of cases analyzed in each comparison (tricuspid atresia versus control).
*$P < .01$ versus control.
†$P < .001$ versus control.

J Thorc Cardiovasc Surg 2003126(4):1026-1032

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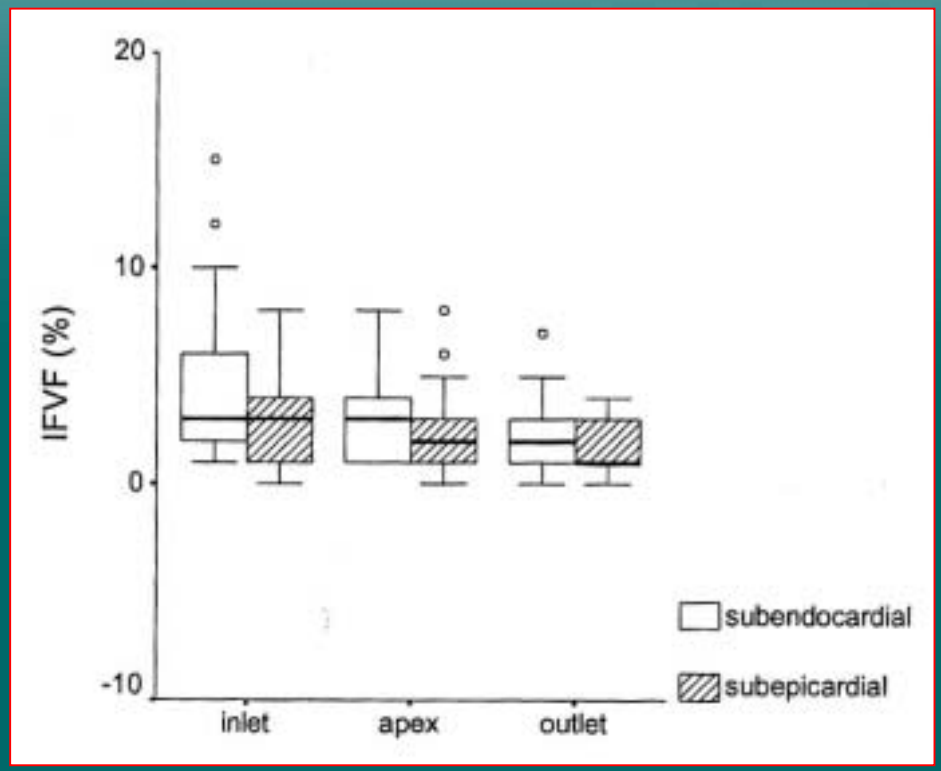

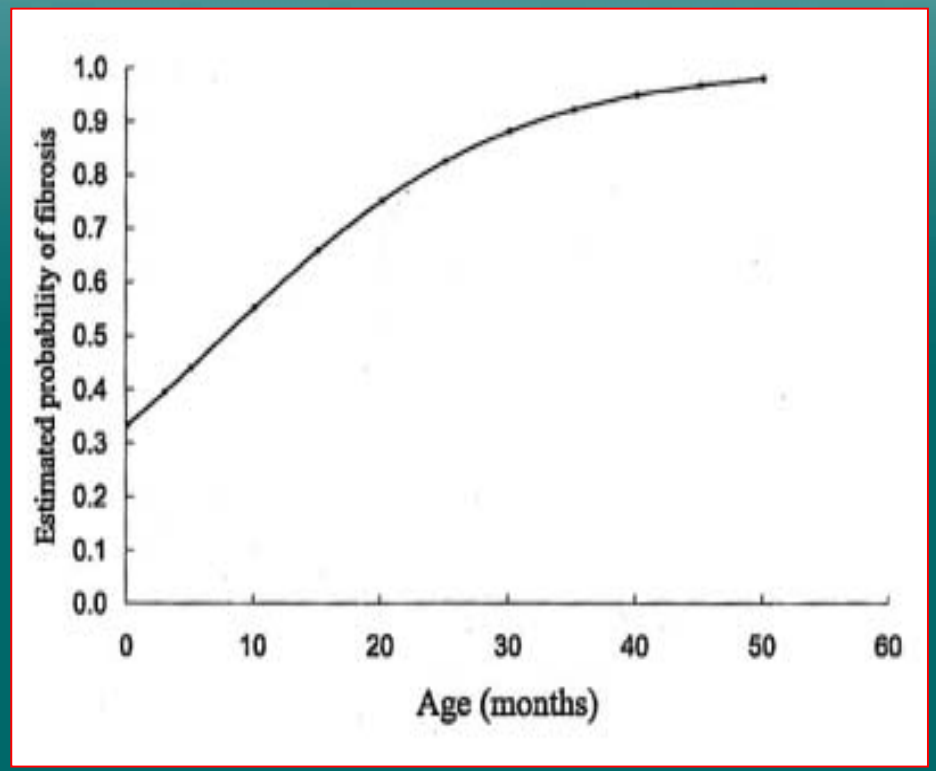

J Thorc Cardiovasc Surg 2003126(4):1026-1032

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Myocardial Perfusion and Coronary Flow Reserve assessed by Positron EmissionTomography in Patients after Fontan-like Operation

Patients: 10 (Fontan-like OP)
M:F=6:4(m age:15.8±5.01yr)
OP age : 5.9±3.7yr
FU after surgery : 9.4±4.2 yr
Control : 10 healthy adults

Method

Clinical exam. ECG, TTE
Cath. Angio/TEE
PETwith N-13 ammonia
at rest & adenosin stress

Result

Vent.dysfunction(VD): 6/10
ESS_m, ESD_m↑ in FLO
MBF higher in FLO than normal
MBF and CFR ↓ in FLO(esp. V.D)
CVR↑ in FLO

Table 3. Hemodynamic and quantitative myocardial flow parameters

Parameter	FLO	Normals
Cardiac index (l/min/m²)	2.34 ± 0.57	
RA pressure (mmHg)	10.3 ± 1.9	
EDP (mmHg)	6.3 ± 2.2	
RA saturation (%)	69.4 ± 5.2	
PA mean pressure (mmHg)	8.6 ± 2.1	
Aortic mean pressure (mmHg)	66.7 ± 10.6	
Aortic saturation (%)	96.1 ± 2.2	
LA pressure (mmHg)	5.6 ± 2.8	
MBF (ml/g/min)	0.91 ± 0.32	0.73 ± 0.15*
MBFN (ml/g/min)	0.99 ± 0.25	0.77 ± 0.17*
$MBF_{adenosine}$ (ml/g/min)	2.12 ± 0.78	3.10 ± 0.8*
CFR	2.5 ± 0.88	4.1 ± 1.01
CVR_{rest} (mmHg/ml/g/min)	98.3 ± 41.3	11 2.4 ± 24.3
$CVR_{adenosine}$ (mmHg/ml/g/min)	38.2 ± 17.4	24.5 ± 8.3

FLO, Fontan-line operation; *RA*, right atrium; *EDP*, end diastolic pressure; *PA*, pulmonary artery; *LA*, left atrium; *MBF*, myocardial blood flow; *MBFN*, myocardial blood flow normalized to the rate pressure product; *CFR*, coronary flow reserve; *CVR*, coronary vascular resistance
* Significant difference (Mann–Whitney test)

Hauser M et al. Pediatr Cardiol 2003;24:36-92

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Medical Center

Plastic Bronchitis after Fontan OP

Formation of large branching bronchial cast*

- Inflammatory cast : fibrin > mucin
  eosinophil Charcot-Leydon crystal
- Acellular cast : mucin > fibrin, eosinophil(-)

↓

symptomatic airway obstruction

Mechanism : unknown

Predisposing factors
- Ventricular dysfunction
- AV valve insufficiency
- Arrhythmias
- PLE

↓

CVP↑

↓

third-space fluid loss → cast (?)

신 ○○, s/p Fontan

Plastic Bronchitis after Fontan OP

Management

- Mucolytics & bronchodilator
- Dnase & steroid← inflammatory
- removal of cast

  repeated bronchoscopy

  aerosolized urokinase

  tissue plasminogen activators
- pacer- induced synchronous

  AV conduction(atrial rate:120/min)

Reports of Plastic Bronchitis

TABLE 1—Reports of Plastic Bronchitis[1]

Authors	n	Associated disease(s)
Mulligan, 1924[5]	1	Pneumonia
Leggat, 1954[17]	1	Bronchiectasis
Sanerkin and Leopold, 1966[1]	1	Asthma
Fruchter et al., 1982[18]	1	Asthma, allergy
Bowen et al., 1985[2]	5[2]	Pericarditis, asthma, bronchiectasis
Liston et al., 1986[19]	3[2]	Asthma, allergy, pneumonia
Muller et al., 1987[20]	3	URI, pericarditis, asthma
Werkhaven and Hollinger, 1987[16]	4	Pneumonia, URI
Colloridi et al., 1990[9]	1	DORV with PVA
Cairns-Bazarin et al., 1992[21]	1	RAD
Duncan et al., 1993[22]	1	TA
Park et al.[23]	1	TOF
Raghuram et al., 1997[8]	3	Sickle-cell/acute chest syndrome
Sesar et al., 1997[11]	9	Asthma/URI/AML with mucositis
Languepin et al., 1999[10]	3	TA, PAPVR, TOF, absent PAV
Quasney et al., 2000[12]	1	TA, PVA
Setzer et al., 2001[24]	1	TA
This report	3	Eosinophilic pneumonia, TOF, TA

[1]URI, upper respiratory tract infection; TOF, tetralogy of Fallot; PAV, pulmonary artery valve; PVA, pulmonary value atresia; PAPVR, partial anomalous pulmonary venous return; TA, triscuspid atresia; AML, acute myelogenous leukemia; DORV, double-outlet right ventricle; RAD, reactive airways disease.
[2]These two reports share one case.

Brogan et al. Pediatr pulmonol 2002;34:482-7

Plastic Bronchitis in Children:

A case series and review of medical literature

TABLE 2—Patient Data by Underlying Illness

	Group I (n = 14)	Group II (n = 17)	Group III (n = 11)
Age (years)	5.7 (4.7)	7.0 (4.1)	4.9 (4.8)
Sex (m/f)	7/6	7/9	7/5
Fever (%)	62	12	67
Mechanical ventilation (%)	31	31	50
Recurrences (%)	31	47	37
Pleural effusions (%)	0	31	25
Air leak	15	6	17
Survival (%)	100	71	83
Type I infiltrates (%)	77	12*	58
Type II infiltrates (%)	8	65	25
No pathology	15	24	17

*$P < 0.05$ compared to group I.

TABLE 3—Cardiac Procedures[1]

Prebronchial cast procedures	
Fontan	10
Bidirectional Glenn	1
Blalock Taussig shunt	2
TOF repair	3
Postbronchial cast procedures	
Pericardiectomy	2
PAPVR repair	1
Thoracic duct ligation	2
Pacemaker placement	1
PA stent/pacemaker placement	1

[1]TOF, tetralogy of Fallot; PAPVR, partial anomalous venous return; PA, pulmonary artery.

Group I : asthma/allergic, Group II : cardiac, Group III : none

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Summary

- To prevent progressive ventricular dysfunction
  - avoid prolonged PAB state till Fontan operation
  - correct residual shunt through catheter intervention to avoid chronic volume overloaded state
  - correct residual anatomical defect surgically to avoid pressure overload
  - High-risk group for Fontan OP→ fenestration
  - avoid long standing hypoxemia through catheter intervention (pulm.AVM, system.to pulm. Venoveno collaterals, fenestration)
  - correct late sequales medically or through catheter intervention
- Life threatening late sequale(esp. PLE or refractory arrhythmia) combined with ventricular dysfunction ➔ Fontan conversion or heart transplantation
- Long-term anticoaguration : controversal
  Fenetrated Fontan , Extracardiac conduit Fontan : warfarin (PT:2-2.5 INR)