

# Fontan Deterioration in Pediatric Cardiologist's View

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The background of the slide is a solid blue color. At the bottom, there are several faint, concentric circular patterns that resemble ripples on water, centered around the bottom edge of the text area.


# Outcomes of Fontan operation

- JTCS 2006:131;172-80 Mitchell ME et al
  - Jan. 1992~Dec. 1999, 332 Fontan
  - 85%–LT, 15%–EC, 49%–HLHS
  - Median 8.6yr F/U
  - Freedom from death or Tx; 98.0% at 1yr, 94.9% at 5yr, 93.9% at 8yr
  - Cardiac–related rehospitalization: 54.2%
  - NYHA class I or II: 86.7%
  - School performance: 30.2% above average, 39.9% average, 29.8% below average

# Factors influencing early and late outcome

- *EJCTS 2007*
- Two commandments
  - Preop impaired ventricular function
  - Elevated pulmonary artery pressure

# Late complications associated with Fontan circulation

- Arrhythmia
  - Thromboembolism and hepatic dysfunction
  - Protein-losing enteropathy
  - Worsening cyanosis
- 

# Arrhythmia in failing Fontan

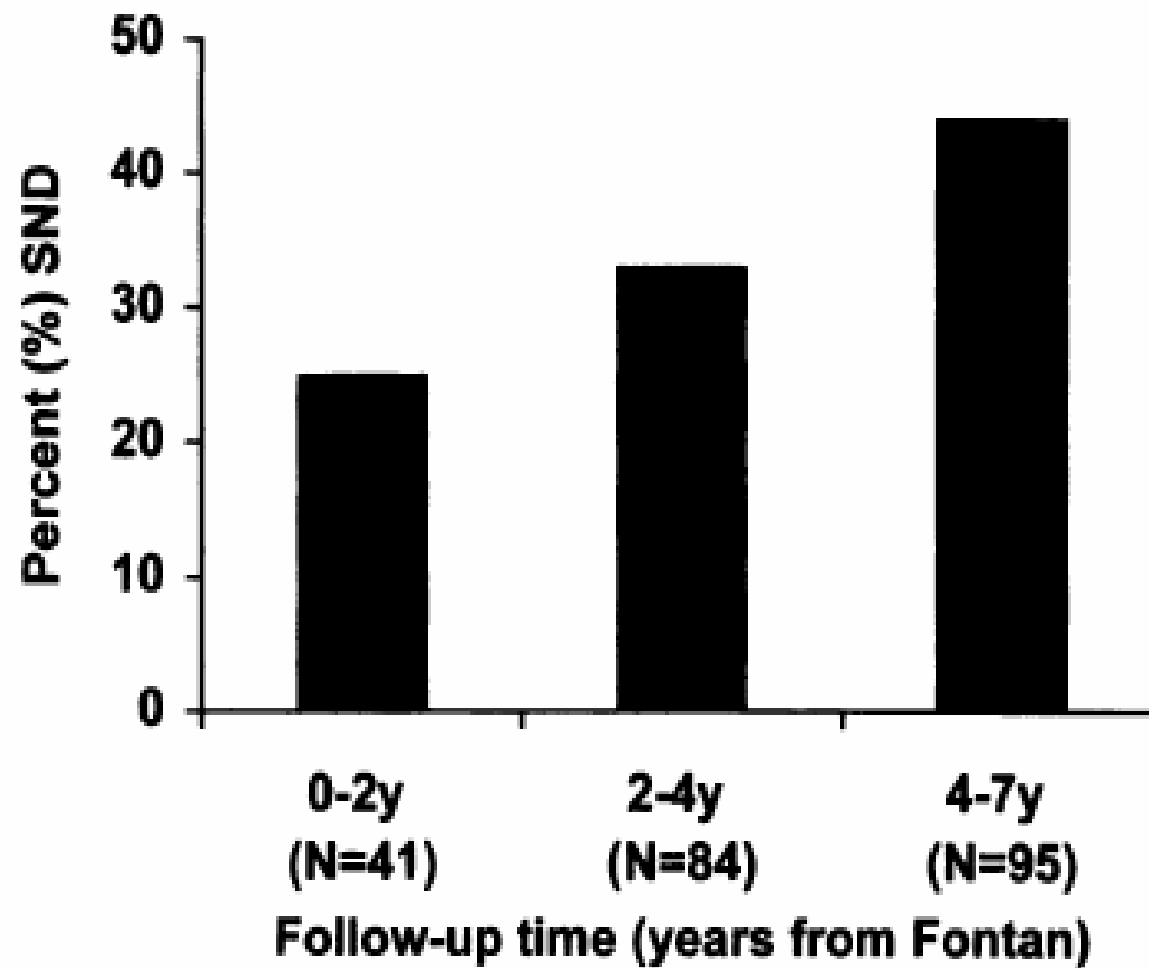
- Mechanism:
  - Injury to sinus node or its blood supply
  - Extensive atrial incision and suture line
  - Chronic exposure of atrial myocardium to elevated pressure with subsequent dilatation
  - Underlying anatomic substrate: discordant AV connection, atrial isomerism
  - Hemodynamic factor: AV valvar regurgitation

# Arrhythmia in failing Fontan: sinus node dysfunction

- Reduce CO
  - Limiting HR in patients with relatively fixed stroke volume
  - ↓ ventricular filling ← junctional rhythm and loss of AV synchrony

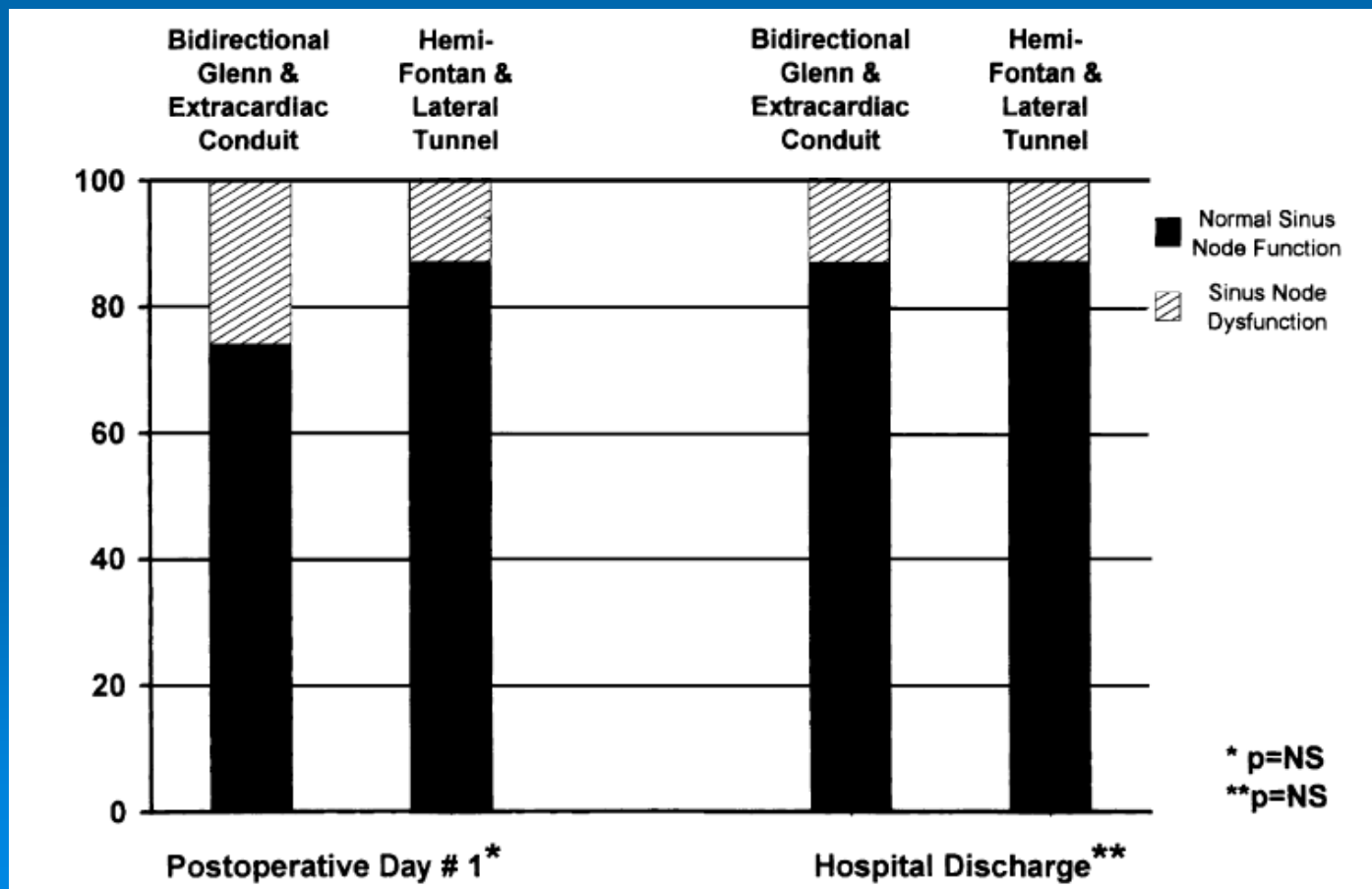
# Arrhythmia in failing Fontan: sinus node dysfunction

- Children's Hospital of Philadelphia  
*Circulation 1998*
- 287 patients staged with hemi-Fontan followed by lateral tunnel Fontan, 1990-1995
- Sinus node dysfunction in 7% before and in 15% after hemi-Fontan, in 23% at early post-Fontan, in 44% at 4~7yr F/U after Fontan





- *JTCS 2000*
- Modifications to the cavopulmonary anastomosis do not eliminate early sinus node dysfunction – 30 EC Fontan vs 46 LT Fontan

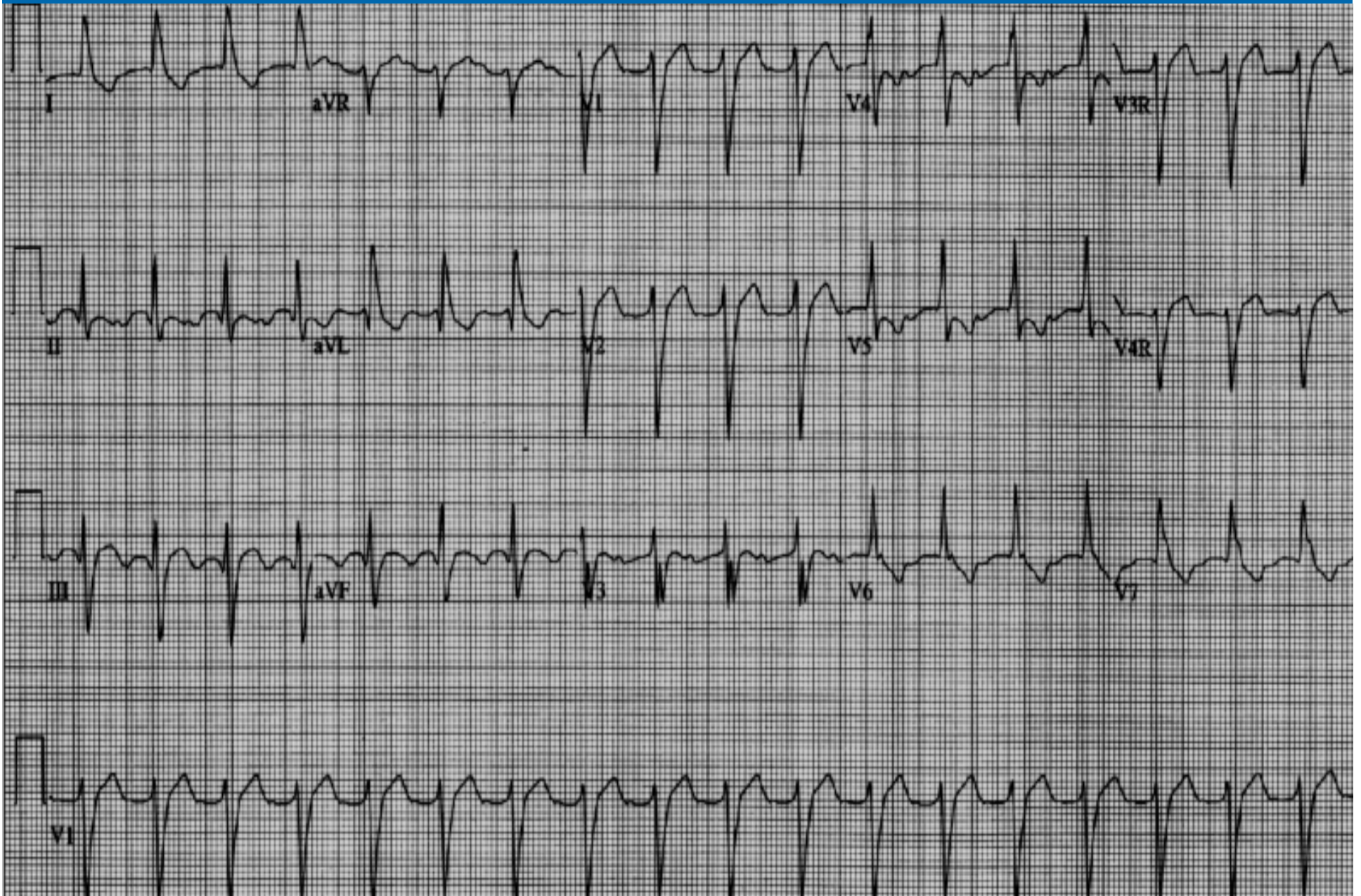


- *ATS 2004;78:1979-88*
- New onset arrhythmia after the EC Fontan op compared with LT procedure
  - 29 LT Fontan (7.9yr), 45 EC Fontan (4.4yr)
  - New onset SVT in early postop  
EC:LT=11%:38%, during F/U 0%:27%
  - Pacemaker due to bradyarrhythmia, in early postop 34%, during F/U +3 patients in LT group, but none in EC group

- *Int J Cardiol 2003;88:285-91*
- SND after Fontan modifications-influence of surgical method
  - SND in situs solitus heart, EC 0/16 vs LT 8/26
  - Staged approach-no risk
- *ATS 2003;76:1389-96*
- LT versus EC Fontan procedure: a concurrent comparison
  - Early postop SND EC:LT=27%:8%, at discharge 10%:0%

# Arrhythmia in failing Fontan: atrial tachyarrhythmia

- Most common form: IART
- IART mimic AF, but longer cycle length ( $> 240\text{ms}$ )
- Often poorly tolerated, limit ventricular filling and cardiac output, associated with atrial thrombus formation
- Risk factor:
  - older age Fontan, prior atrial septectomy, PA reconstruction, worse NYHA class *JTCS 1997*
  - Systemic ventricular dysfunction, heterotaxy, anomalous systemic venous drainage, previous BDG *ATS 2005*



# Arrhythmia in failing Fontan: atrial tachyarrhythmia

- Notoriously resistant to antiarrhythmics
- Increased with F/U
- Reported upto 57% *Heart 2000*
- LT Fontan 3-5yr F/U 4.1% *Circulation 1998*
- Efforts minimize risk of arrhythmia
  - Minimize injury to sinus node & its blood supply
  - Minimize atrial incision & suture line particularly in terminal groove
  - Guard against or reduce AV valvar regurgitation

# Arrhythmia in failing Fontan: atrial tachyarrhythmia

- Digoxin, amiodarone, sotalol
- Radiofrequency ablation
  - Initial success rate around 80%, recurrence rate greater than 50% at 2yr F/U
  - 3-D mapping system and irrigated-tip ablation catheter, immediate success in >80%, Recur or new arrhythmia 30-45% in 6-12 mon after ablation  
*Heart Rhythm 2005, AJC 2003*
- Fontan conversion and arrhythmia surgery
  - Fontan conversion without cryoablation-50~67% clinical recurrence of IART
  - No recurrence after right sided Maze procedure at 2.5yr F/U

# Thromboembolism

- Increased risk of thrombosis
  - Atrial arrhythmia
  - Distended and sluggish Fontan pathway
  - Intravascular prosthetic material
  - Hepatic impairment with multiple clotting factor abnormalities: ↓ level of protein C, protein S, antithrombin III
  - Increased platelet reactivity



# Thromboembolism

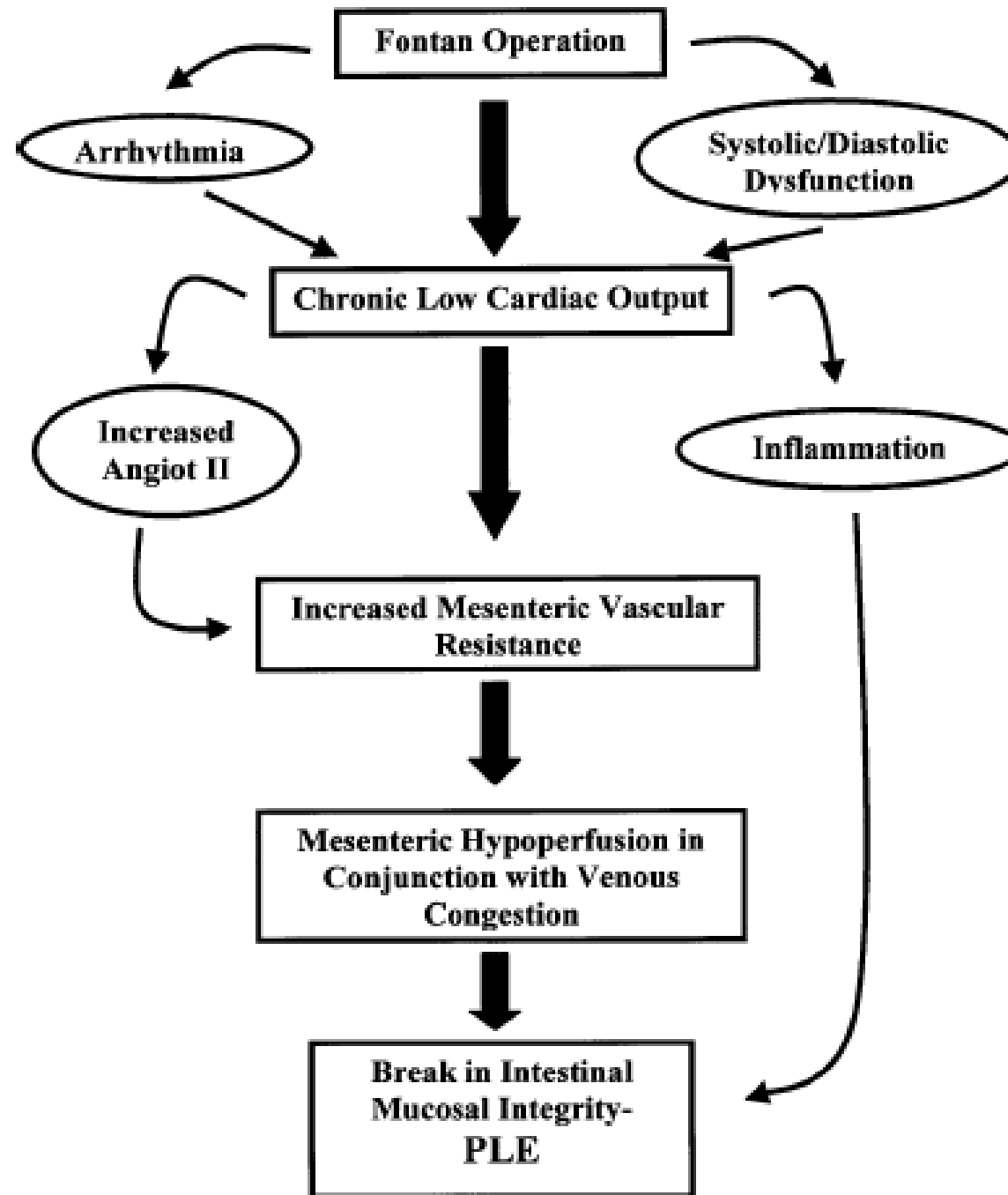
- Exact prevalence: unclear, 3~33%
- Stroke incidence: 2.6% among 645 patients over 15yrs *Pediatr Neurol* 1995
- Freedom from thrombus 92%, 90%, 84%, 82% at 1, 3, 8, 10yr after Fontan *ATS* 2001

# Thromboembolism

- Ligation of pulmonary trunk leaving blind cul-de-sac distal to pulmonary valve: worrisome substrate for occurrence of thromboembolism
- The role of long-term antiplatelet or anticoagulation therapy: poorly defined

# Protein-losing enteropathy

- 3~15% of Fontan patients
- Sx: fatigue, peripheral edema, pleural and pericardial effusion, ascites, chronic diarrhea
- Dx: low serum albumin, increased fecal  $\alpha_1$ -antitrypsin levels
- Mediated by chronically elevated central venous pressure



# Protein-losing enteropathy

- Other risk factor: longer CPB time, morphologic RV anatomy *AJC 2001*
- In patients with generalized edema, 5yr survival rate  $\approx 50\%$ , 10yr 1/5 *JTCS 1998*

# Protein-losing enteropathy

- Multiple therapeutic approach:
  - Dietary modification with high protein, high MCT
  - Afterload reduction agent
  - Inotropic agent
  - Heparin
  - Albumin infusion
  - Octreotide
  - Prednisone
  - Pacing on significant SND
  - Atrial fenestration
  - Fontan revision
  - Cardiac transplantation

# Worsening cyanosis

- In absence of atrial fenestration, SpO<sub>2</sub> ≥ 94%
- Common cause:
  - Progressive deterioration of ventricular function with or without AV valve regurgitation
  - Shunting through baffle leak or residual interatrial communication
  - Pulmonary vein compression by giant RA or aorta
  - Systemic venous collateralization
  - Pulmonary AV malformation
  - Pulmonary pathology
  - Hepatic venous connection to CS or LA
  - Rt-to-Lt interatrial shunt via small thebesian vein
  - Diaphragmatic paresis

## Effectiveness of Carvedilol for Congestive Heart Failure that Developed Long after Modified Fontan Operation

Naoko Ishibashi · In-Sam Park · Yukiko Takahashi · Mitsunori Nishiyama ·  
Yasue Murakami · Katsuhiko Mori · Shigekazu Mimori · Makoto Ando ·  
Yukihiro Takahashi · Toshio Nakanishi

- 27yr old man
- Intractable CHF due to severe ventricular dysfunction
- Carvedilol 2mg/day → 30mg/day
- ↓ atrial pressure and improved ventricular function



# β-blocker on heart failure

- Mechanism
  - Suppression of oxygen consumption
  - Upregulation of  $\beta$ -adrenergic receptor
  - Antiarrhythmic effect
  - Reduction of cardiac norepinephrine
  - Improvement of ventricular diastolic function

# Carvedilol

- Potent nonselective  $\beta$ -blocker
  - $\alpha_1$ -adrenergic receptor blocker:  
vasodilating effect
  - Antioxidant effect
- a primary drug for treatment of CHF

- Bosentan induces clinical, exercise and hemodynamic improvement in a pre-transplant patient with plastic bronchitis after Fontan operation
  - *J Heart Lung Transplant 2005;24:1174-6*
  - 14yr old boy with high pulmonary artery pressure

# Fontan experience

- Total 45 patients
  - 1994. July 11~2003. Feb., 24 cases at DAUH
  - 2003. Mar~2006. Dec., 21 cases at PNUH
  - Median F/U 4yr 11mon
- Age: 1yr 9mon~ 24yr, median 2yr 11mon
- Body wt: 10.0~63.8kg, median 12.8kg

- **Underlying conditions**

- Tricuspid atresia 13
- Isomeric heart 6, RAI 5/LAI 1
- Common inlet ventricle 5, RV 3/ LV 2
- Mitral atresia or stenosis/LV hypoplasia 6
- PA IVS 4
- DILV 3
- Ebstein 2
- HLHS 1
- DORV/ MV straddling 1
- Others 4

- Op methods
  - Lateral tunnel procedure: 6
  - Extracardiac conduit TCPC: 38
    - 18mm, 2; 19mm, 7; 20mm, 26; 22mm, 2; 24mm, 1
  - Hepatic vein inclusion after Kawashima: 1
  - Staged with BCPC except 3 cases
  - Right Maze in 2 patients
- No early mortality, 1 late mortality
- Intractable chylothorax 3
- Pacemaker in 2, antiarrhythmics in 1

# Follow-up by specialized team

- Regular surveillance
  - Thorough clinical history
  - Physical examination
  - Resting oximetry
  - 12-lead EKG
  - Chest X-ray
  - Echocardiography
  - CBC, LFT, serum protein/albumin
  - Occasional Holter monitoring