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- Other special considerations

Selection Criteria for Fontan Op

“The Ten Commandments”

(Fontan and Choussat, 1977)

1. Minimum age of 4 years
2. Sinus rhythm
3. Vena cavae - normal drainage
4. Right atrium of normal volume
5. Mean PAP of < 16 mm Hg
6. Pulmonary resistance of < 4 units/m²
7. PA/Aorta diameter ratio > 0.75
8. Normal function of primitive ventricle (EF $> 60\%$)
9. No mitral incompetence
10. No impairing effects of previous shunts

Contraindications to Fontan Op

(Handy JR Jr and Sade RM,1995)

- **Absolute contraindications**
 1. Early infancy (age < 6-10 months)
 2. Severe hypoplasia of parenchymal pulmonary atresia
 3. Pulmonary vascular resistance > 4 clinical units
- **Relative contraindications**
 1. Age < 1-2 years
 2. Mean pulmonary artery pressure > 15 mm Hg
 3. Ventricular end-diastolic pressure > 15 mm Hg
 4. Pulmonary vascular resistance 2-4 clinical units
 5. Previous pulmonary artery band
 6. **Substantial ventricular hypertrophy**
 7. Mitral or aortic valve insufficiency > mild

Updated Version of the Optimal Criteria for Fontan Op

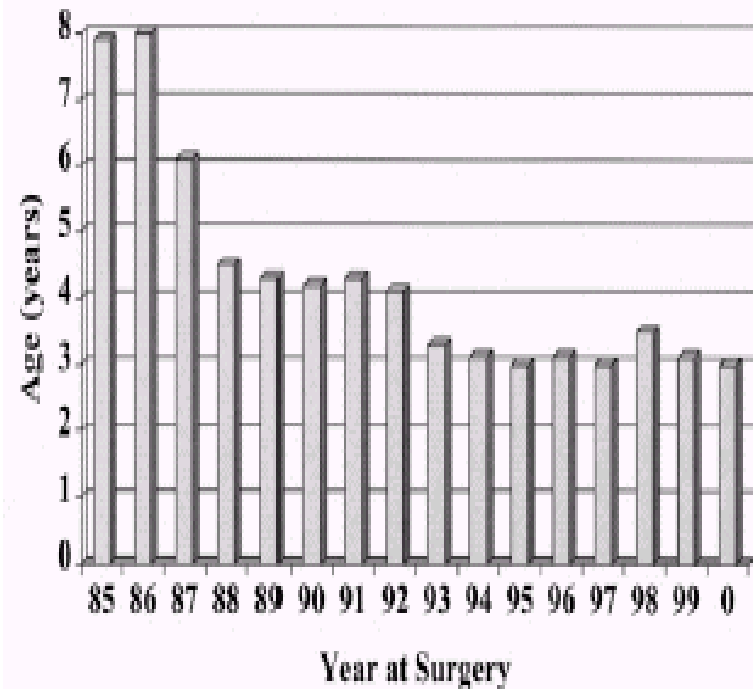
(Tchervenkov and Tsang, 1999)

1. Normal sinus rhythm
2. Normal caval and pulmonary venous connections
3. Normal PVR, with a mean PAP < 15-20 mm Hg
4. No significant PA branch stenosis that would preclude surgical repair
5. PA-Aorta ratio > 0.75
6. Normal ventricular function
7. No systemic AVVR
8. Normal diastolic ventricular function
9. Optimal minimal age: uncertain, probably 2-4 Y
10. Unobstructed systemic circulation (no aortic arch obstruction or SAS)

Age for Fontan Op

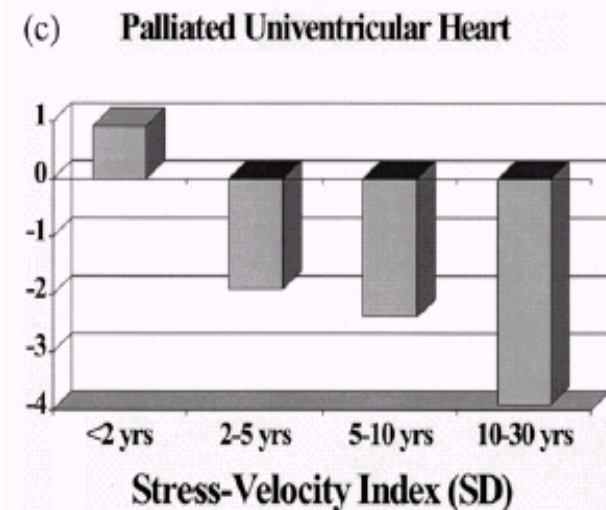
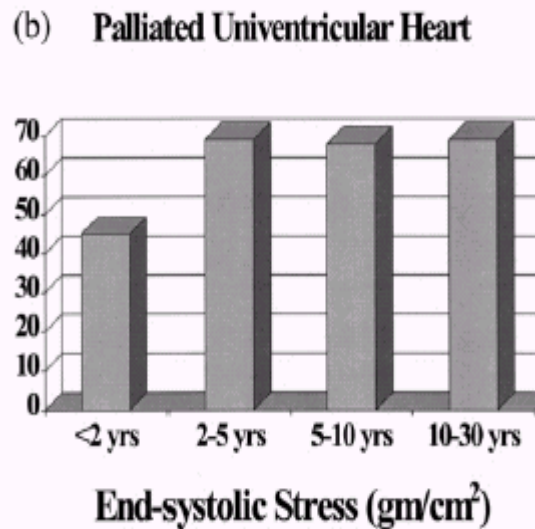
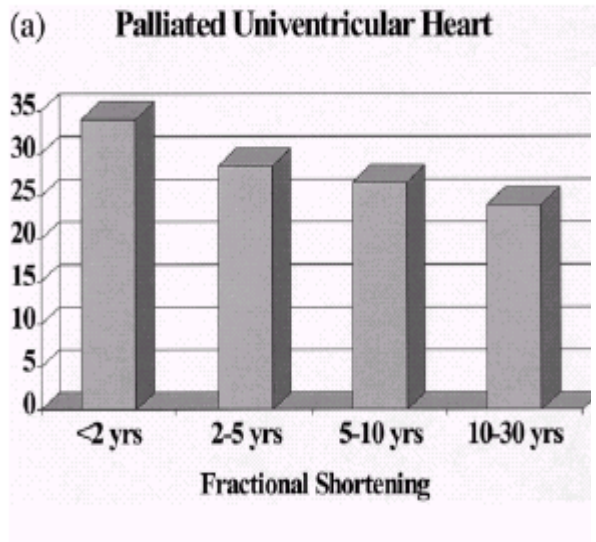
- Impacts of old age on single ventricle
 - Chronic hypoxia
 - Chronic volume loading
 - Ventricle hypertrophy

Median Age at Fontan Op; Boston data
(Colan SD, 2002)



Relationship of Age to Myocardial Mechanics in Patients with Palliated Univentricular Heart

(Sluysmans T, 1992)



Age at Fontan op – How Young?

- Not younger than one who can walk alone
 - No muscle pump effect on venous return
- Suitable < 2 years of age (Weber HS,1992)
- One-stage Fontan < 2 Y (Chowdhury UK,2001)
 - No significant difference in mortality in < 2Y
 - Fenestration ameliorates morbidity/mortality.
 - Noncompliance with > 2 of Choussat's criteria appears to be additive in unsatisfactory outcome.

Age at Fontan op – How Old?

- About 2 mm Hg ↑ in LVEDP every decade after 4th
- More strict selection criteria? (Gates RN,1997,UCLA)

➤ Ventricle dysfunction

early results (Mott AR,2004,Texas)

Mott AR,2004,Texas)

proved functional class after Fontan

tional class after Fontan

tra-operative treatment of arrhythmia

ew onset arrhythmias are sources of peri-

hythmias are sources of peri-operative

Rhythm

- Sinus rhythm is not prerequisite.
- Intra-operative RF- or cryo-ablation
- Pacemaker insertion

Systemic & Pulmonary Venous Drainage

- Especially in isomerism
- IVC interruption
- Separate hepatic veins
 - Pulmonary AVF
- Combined use of baffle & conduit

PVR & PAP

- PVR > 4 units; the only absolute CI x
- Mean PAP < 15 mm Hg
- PAP up to 25 mm Hg if
 - High PBF or
 - Significant AVVR (Mair DD,1985)

PA Size & Fontan Op

- Unfavorable outcome in pts with small PA

- McGoon ratio < 1.8 (Fontan F,1989)

- oon ratio < 1.8 (Fontan F,1989)

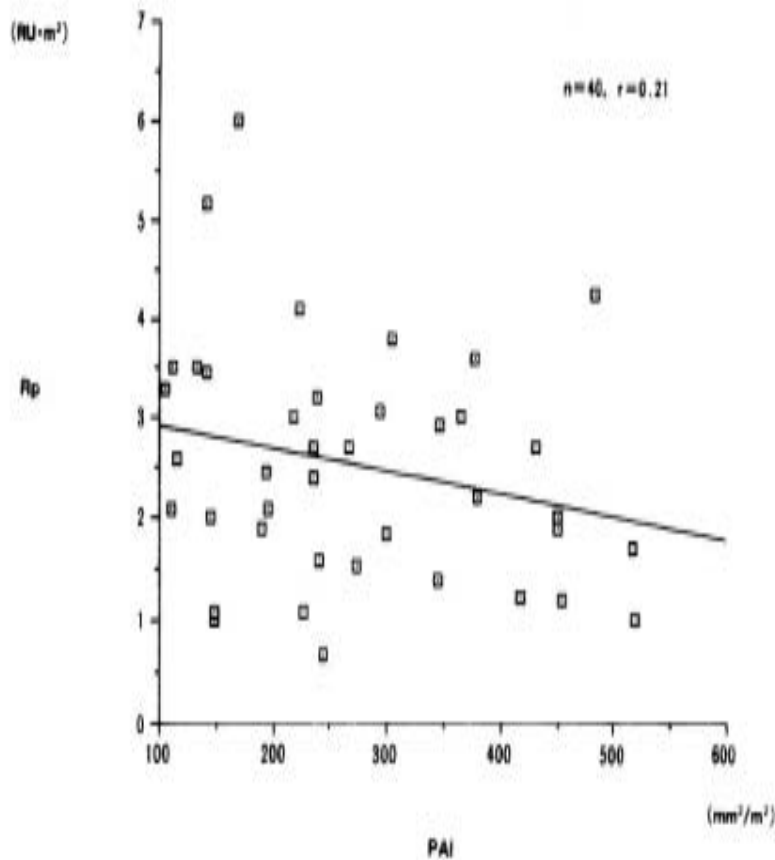
- ada PA index $< 250 \text{ mm}^2/\text{M}^2$ (Nakada S,1984)
984)

rather than PA size

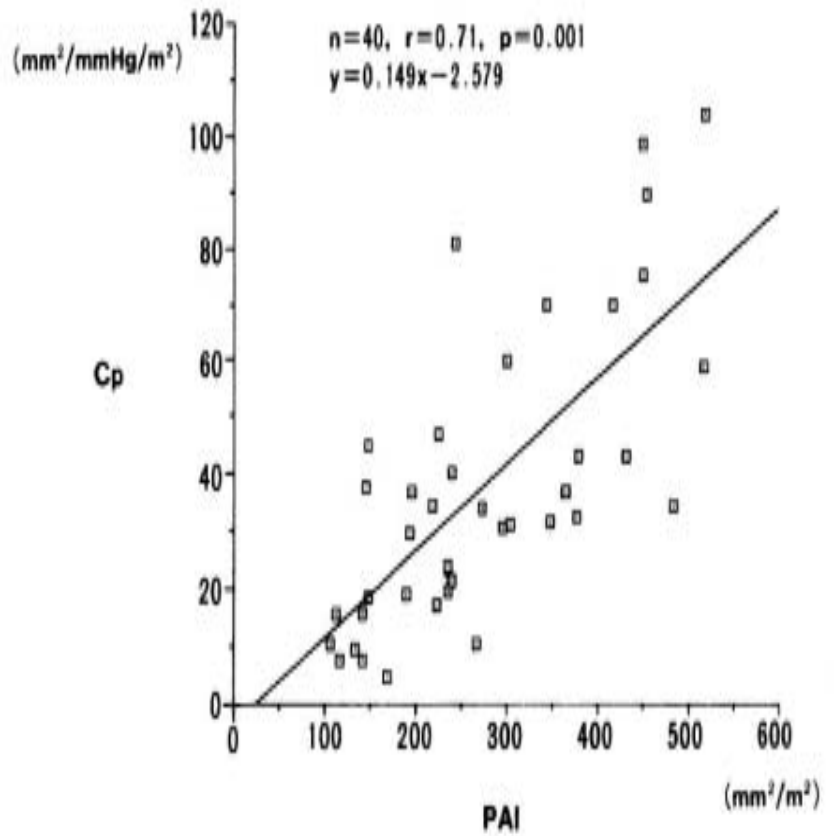
ecially after introduction of BCPS, fenestration
estration

PA Size & Fontan Op

(Senzaki H,1994)



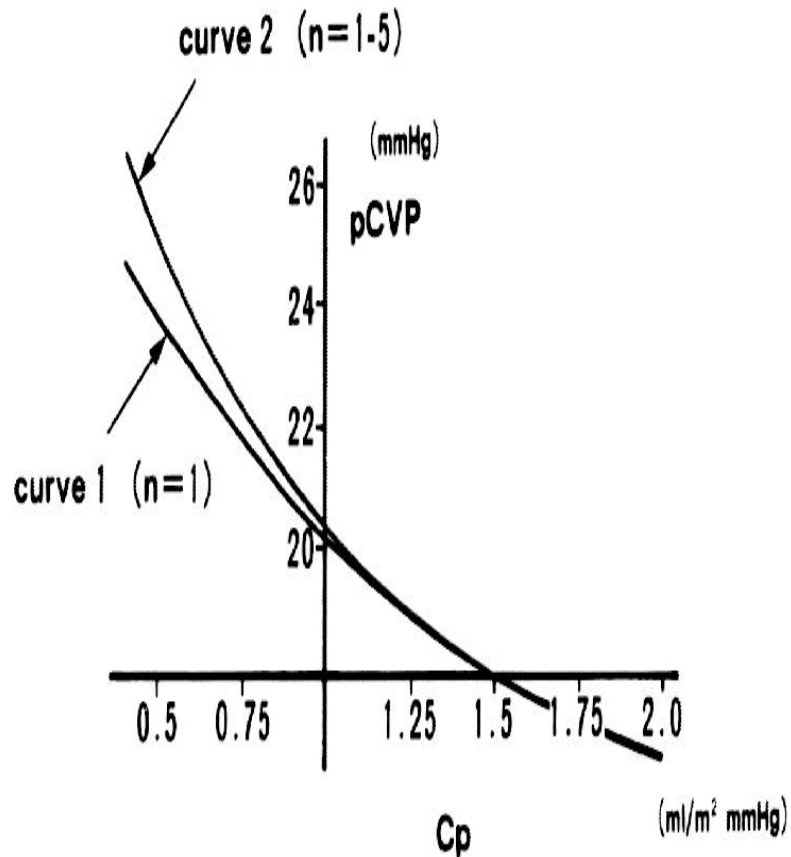
PAI vs PVR



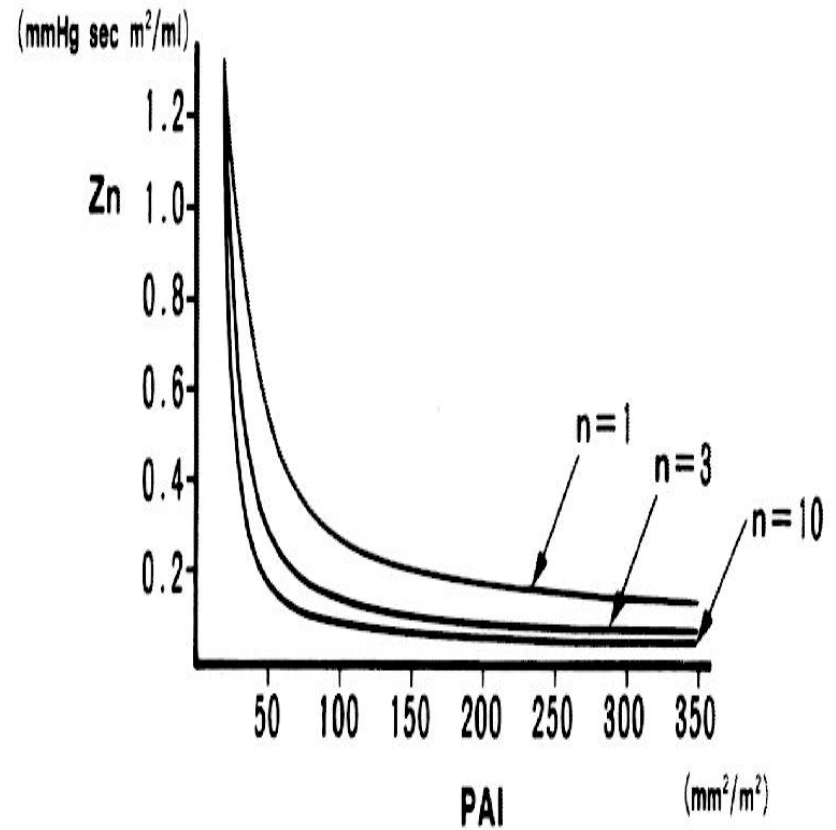
PAI vs Pulmonary vascular compliance

PA Size & Fontan Op

(Senzaki H, 1994)



Vascular compliance vs Peak CVP



PAI vs Total impedance

Hemodynamic Changes after Fontan operation

- Acute increase of SVR

| Changes after Fontan | Impacts on SVR |
|---|---|
| Serial pulmonary circulation from parallel circulation | reduces cross-sectional area & increases total vascular length |
| rise in arterial saturation & fall in cardiac output | peripheral vascular constriction & neurohumoral activation |

- The net impact is **a 75% increase in SVR**.

- Decrease of preload

- ↓ of load-dependent systolic function

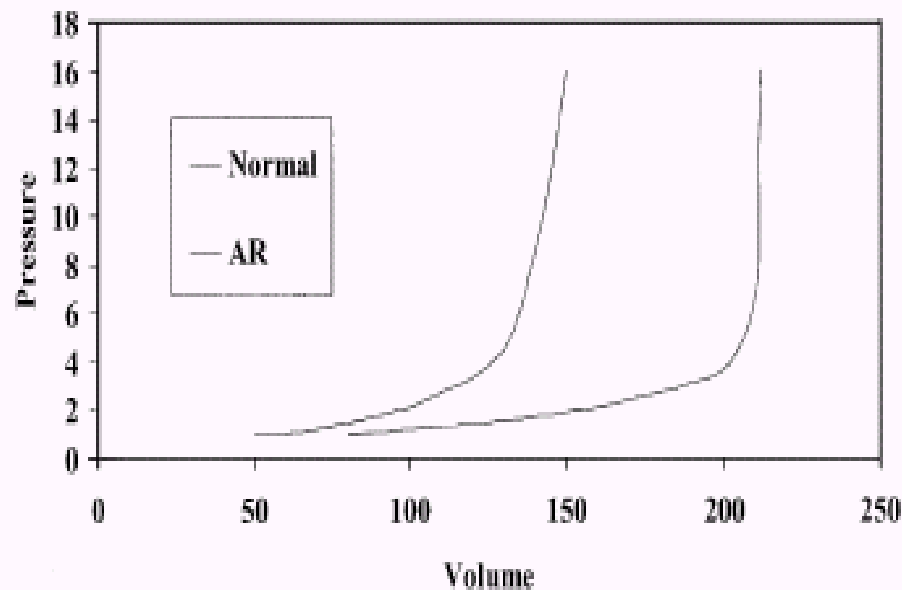
- ➔ ↑ risk of Fontan in ventricle dysfunction

Hemodynamic Changes after Fontan operation

- Constant LV mass & decreased volume
 - Increased mass/volume ratio
- Impaired, incoordinate ventricle relaxation
 - Post-Fontan Doppler; $E\downarrow$, $A\uparrow\uparrow$
 - Abnormal wall motion

Ventricular Compliance

Diastolic Pressure-Volume Curve



Typical response of the left ventricular diastolic pressure-volume curve in patients with a volume overload lesion such as chronic aortic regurgitation. The curve is shifted to the right, such that over the operant range of volume, the pressure is lower than in the normal ventricle, consistent with **enhanced chamber compliance**. (In Colan SD, 2002)

LVH / SAS in Fontan Candidates

- Increased mass/volume ratio after Fontan Op
 - Difficult to predict post-Fontan change
 - Isoprel infusion may unmask latent SAS
- Factors contributing to acquired systemic LVOTO
 - DI LV, rudimentary RV, and a discordant VA connection
 - Size of the VSD
 - Previous PAB & other volume unloading procedures
- Staging with DKS w/wo BCPS

Summaries

- No absolute criteria except PVR
- Multiple factors rather than single factor
- Concept of “threshold” effect, becoming important in those patients who are at higher overall risk by virtue of other factors such as older age, technical approach to the Fontan procedure, or other risk factors

Cardiac Cath before Fontan

Hemodynamic data

- Calculation of PVR
 - PAP (or PV wedge)
 - PV pressure (or PA wedge)
 - O2 consumption
 - Check pressure after closure of APCs and venous collaterals
- Ventricle EDP
- Pressure gradient at subaortic area
 - Can be different after Fontan; Isoprel ?

Anatomy

- PA anatomy
- Pulmonary venous return
- Systemic venous return
 - Separate hepatic vein
 - LSVC
 - Venous collaterals
- LVOTO
- AP collaterals
- Pulmonary AVF
- AVV function - Echo

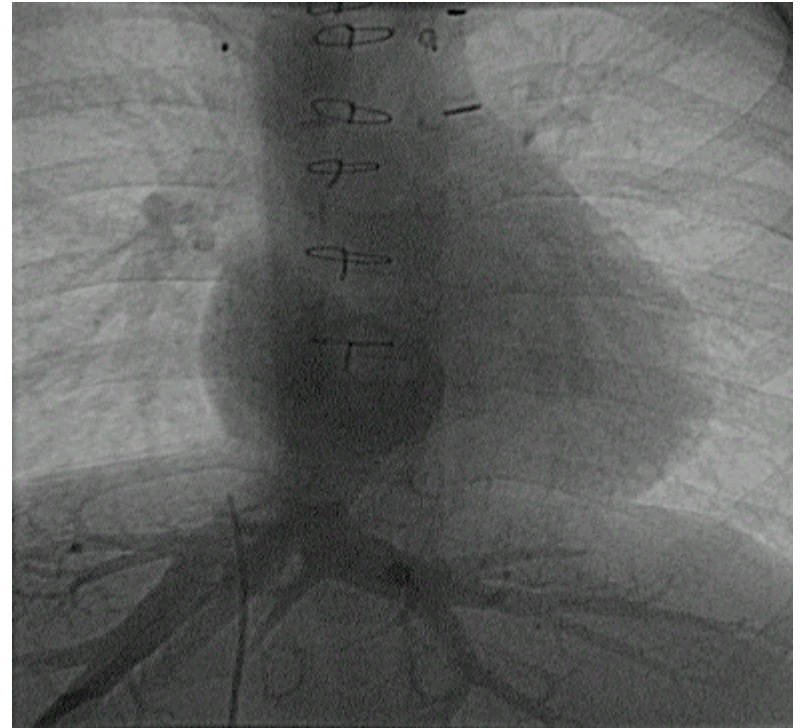
Balloon Occlusion Angiogram at Innominate Vein

- Venous collaterals
 - Closure collaterals except collaterals to IVC
 - Re-open of small LSVC
- Angiogram at both SVCs after bilateral BCPS
- PA anatomy & PV drainage



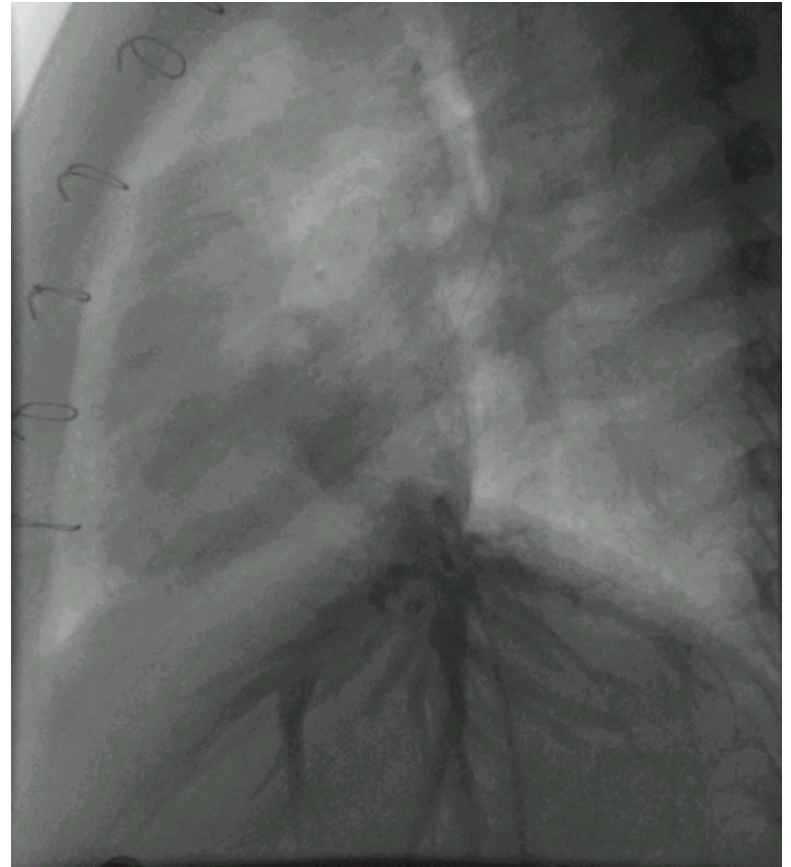
IVC Angiogram

- Draining site of IVC & hepatic veins
- IVC interruption
- Separate hepatic veins

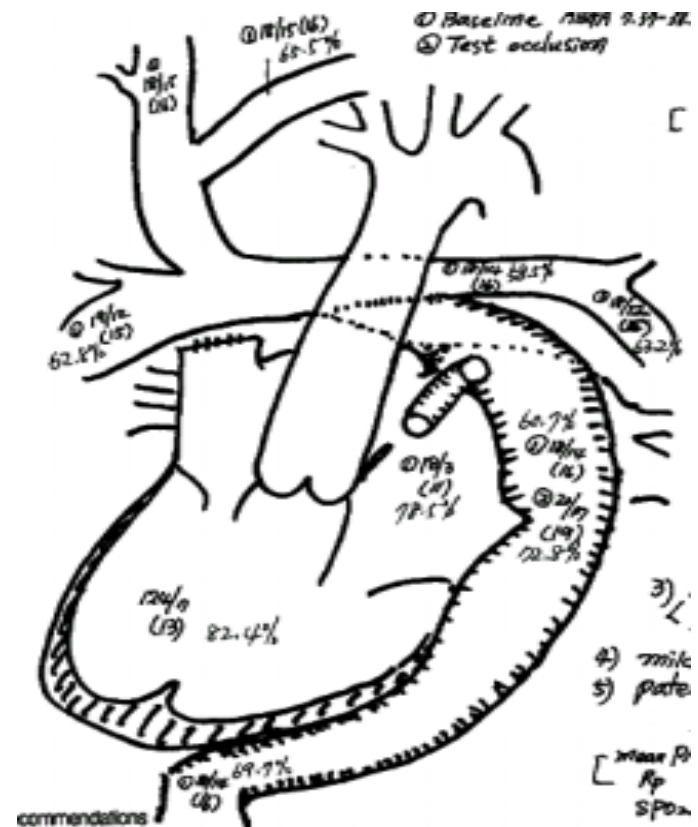


IVC Angiogram

Separate Hepatic Veins?



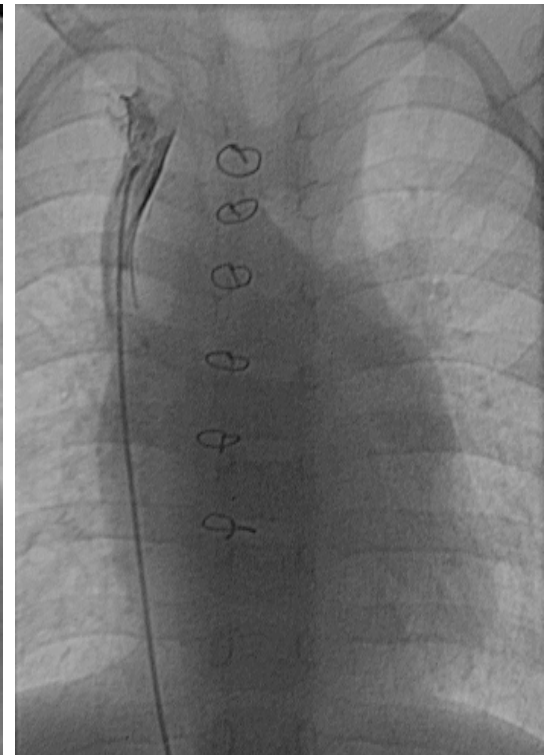
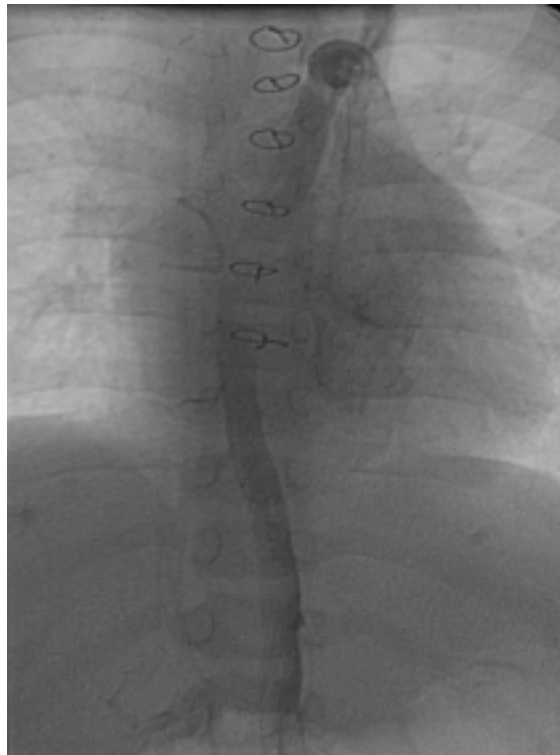
- 2.8Y, S-solitus, dextrocardia, DI LV, large VSD & ASD, PA
- s/p RMBT & PA angioplasty (6W of age) s/p BCPS (14M of age)
- Dextrocardia, right side adhesion, right side IVC



IVC Angiogram

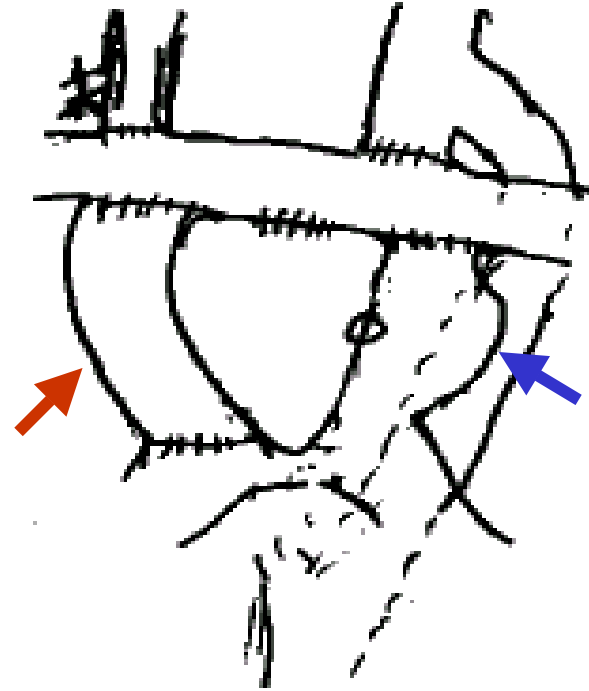
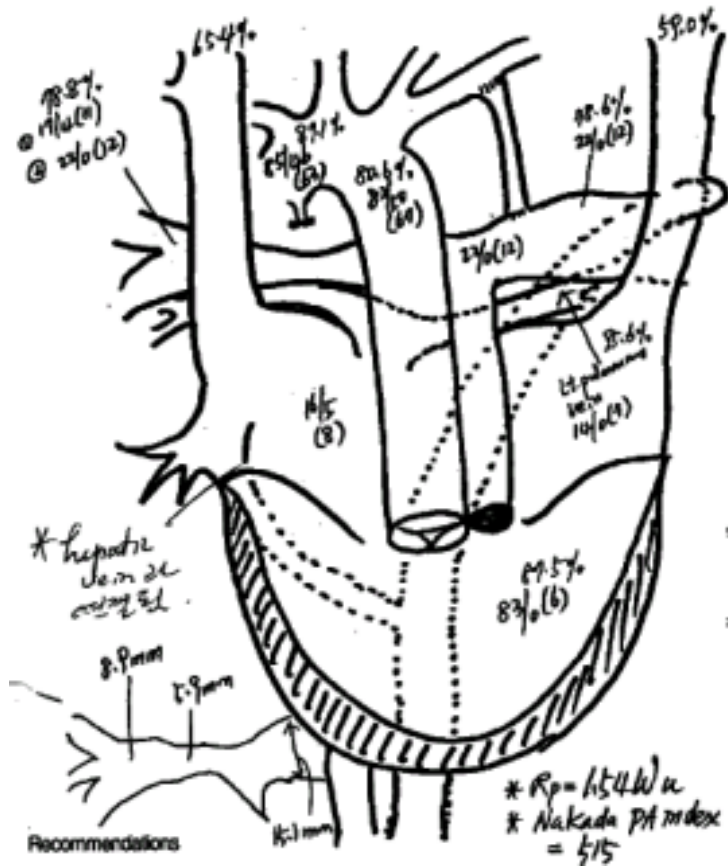
Abnormal Systemic Venous Return

- 23M/10 kg; Lt isomerism
- SA, C-AVSD, DORV, Rt arch, Rt PDA, Bilateral SVCs, IVC interruption with azygos continuation to LSVC, Communication between hepatic veins & IVC
- s/p LMBT 4 mm (2W of age) s/p RPA angioplasty (8M of age)



IVC Angiogram

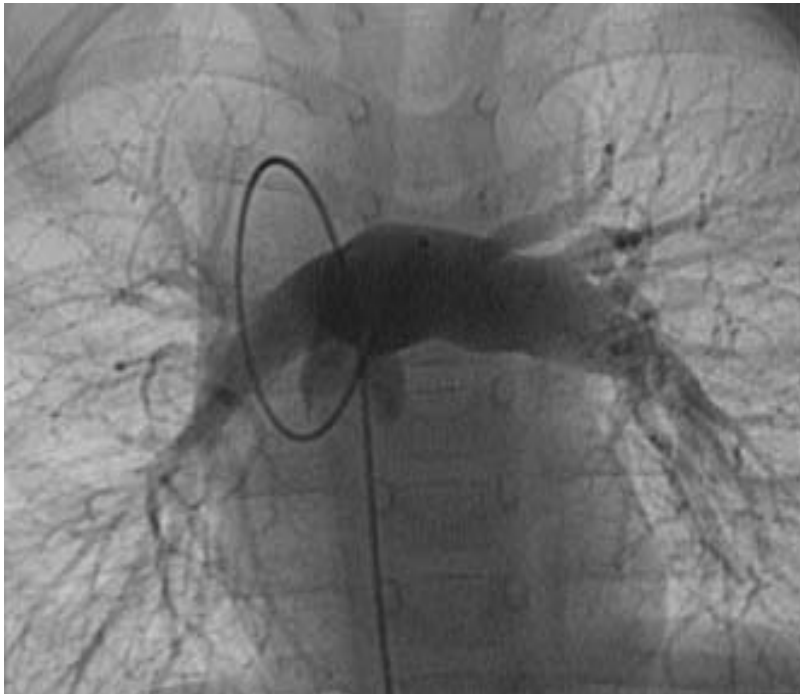
Abnormal Systemic Venous Return



- ↑ Rt side; Extra-cardiac Fontan (16 mm)
- ↑ Lt side; Intra-cardiac lateral tunnel with 4 mm fenestration

Abnormal PV Return

- 3Y 8M/M; Rt isomerism, SA, SV, C-AVSD, DORV, combined PS, TAPVR to IV via VV
- One stage extra-cardiac Fontan with fenestration



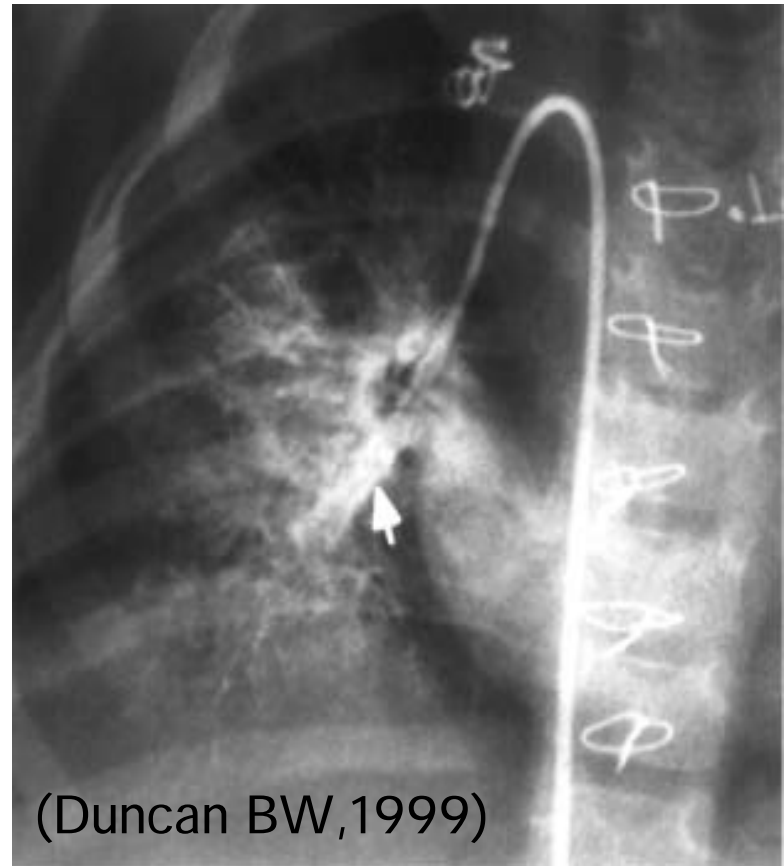
Selective PA Angiogram

- PA anatomy
- Pulmonary venous drainage
- Balloon occlusion angiogram at innominate vein in post-BCPS patients



Pulmonary AVF

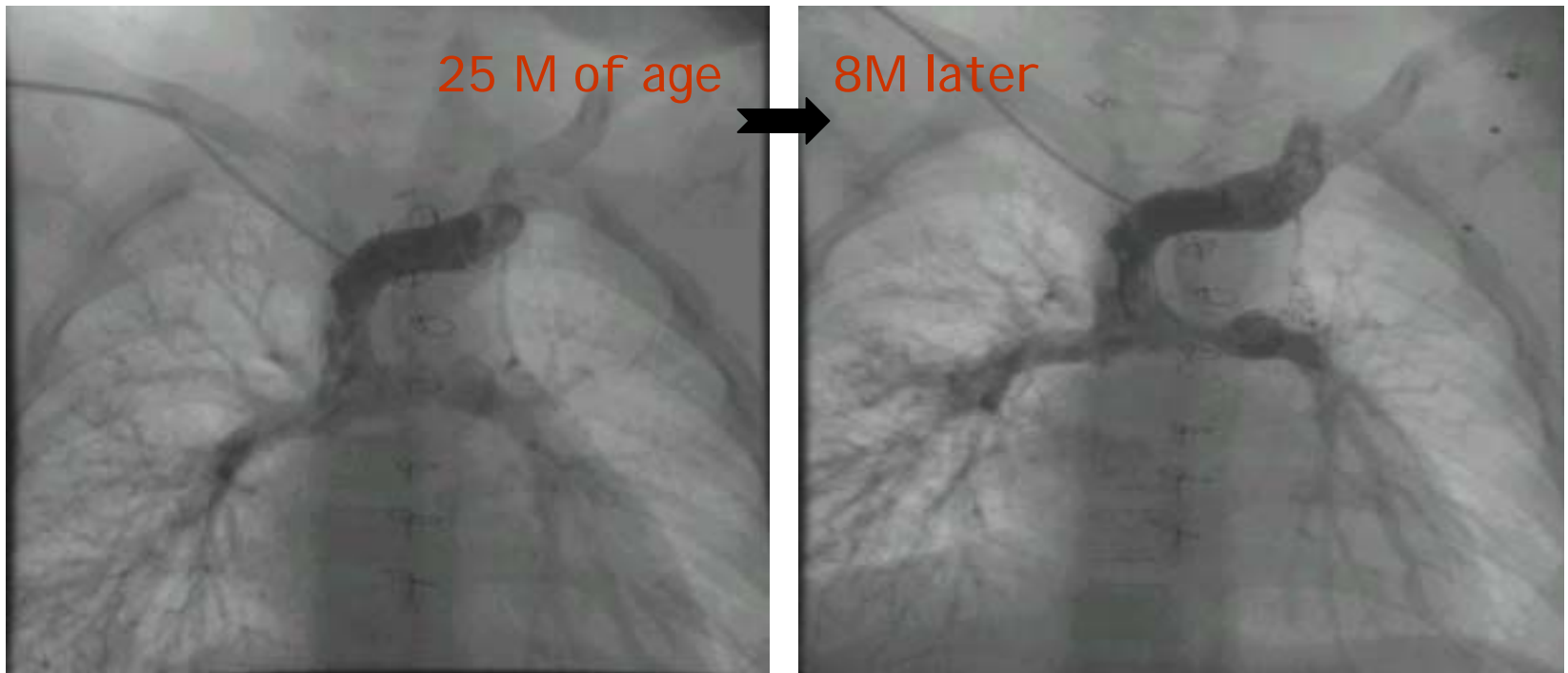
- After BCPS or Kawashima operation
- Unusually low SpO₂
- Pulmonary venous desaturation in the absence of venous collaterals to pulmonary veins or lung problems
- Selective PA angiogram
 - Rapid filling of pulmonary vein
 - "Lake" or "chain"
- Selective contrast echo at suspected PA



Pulmonary AVF

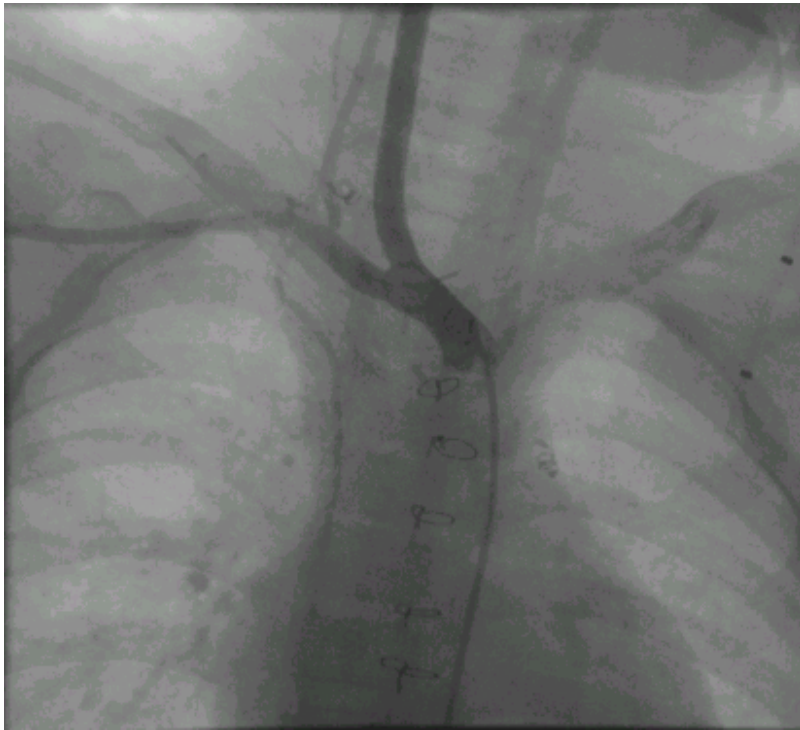
Role of APCs

- PA/IVS, severe TS
- s/p BVP at other hospital (1W) s/p BCPS & RV exclusion (5M)
- TCO of arterial collaterals to the Rt lung (25M)

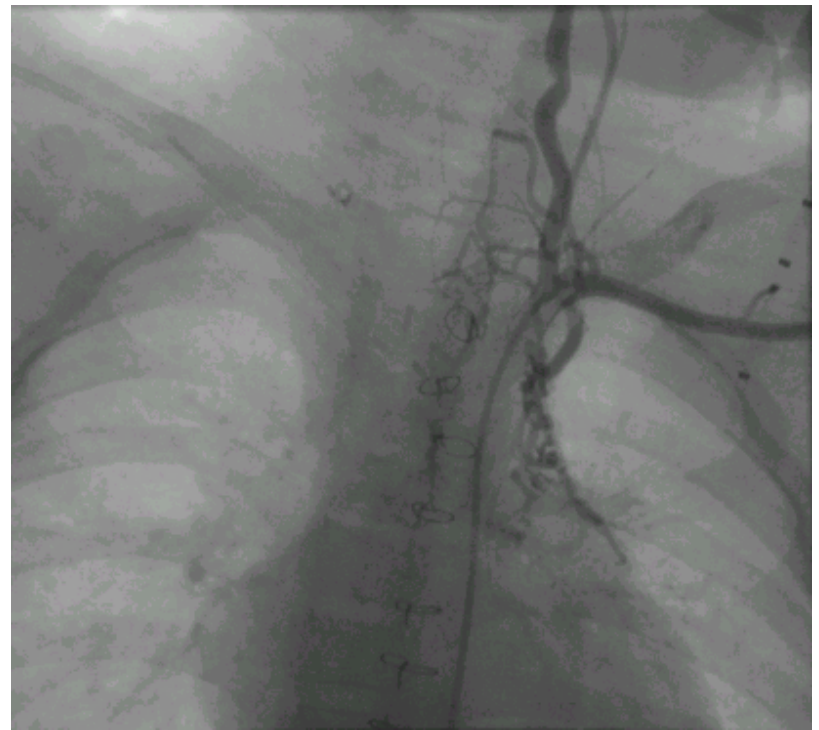


Pulmonary AVF

Role of APCs



No significant APCs in Rt lung



Several APCs in Lt lung

Aorto-Pulmonary Collaterals (APCs) in Fontan Candidates

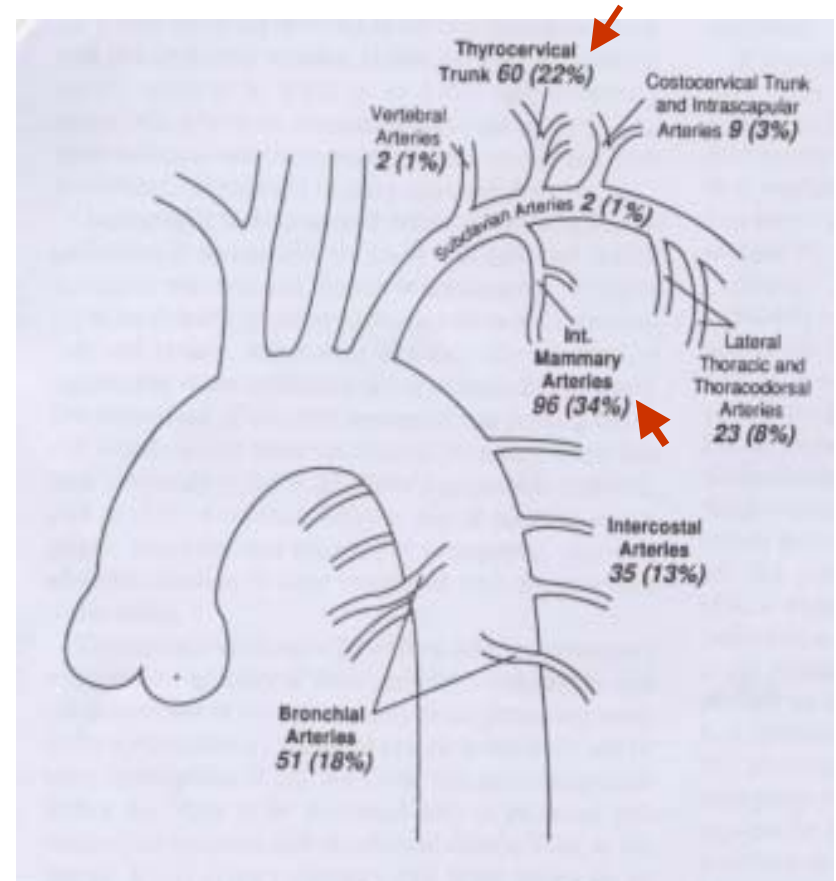
- Increase PBF & ventricle volume loading
- Increase PAP & LAP
- Pleural / pericardial effusion
- Increased mortality? (Ichikawa H, 1995)
 - Direct measurement of the amount APCs at OR
 - If > 33% of cardiac return
Post-op CVP > 17 mm Hg
Mortality in all

Aorto-Pulmonary Collaterals

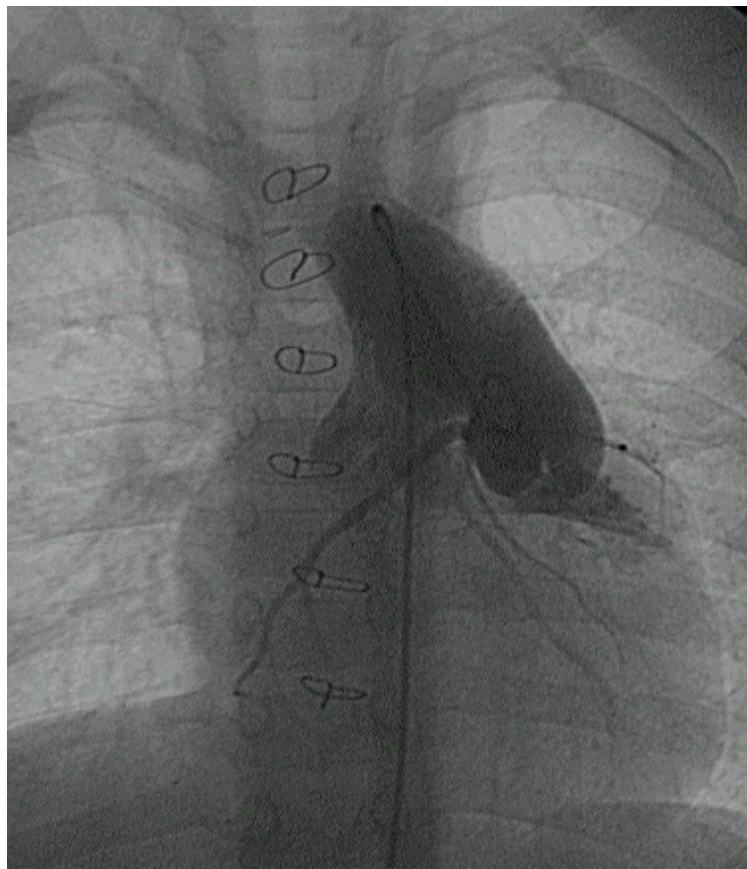
- O₂ jump-up > 5 %
- Negative wash-out on PA angiogram
- Selective angiogram at collaterals
 - Direct visualization of PA
 - Visualization of pulmonary vein on levo-phase
- ➔ Need for closure before Fontan

Source of APCs (Triedman JK, 1993, JACC)

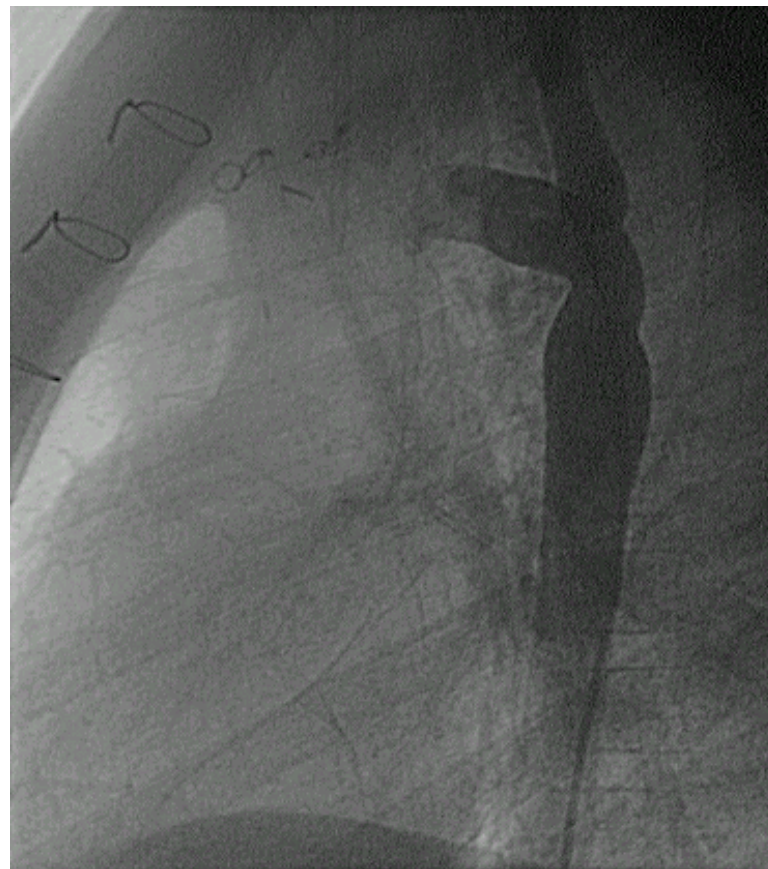
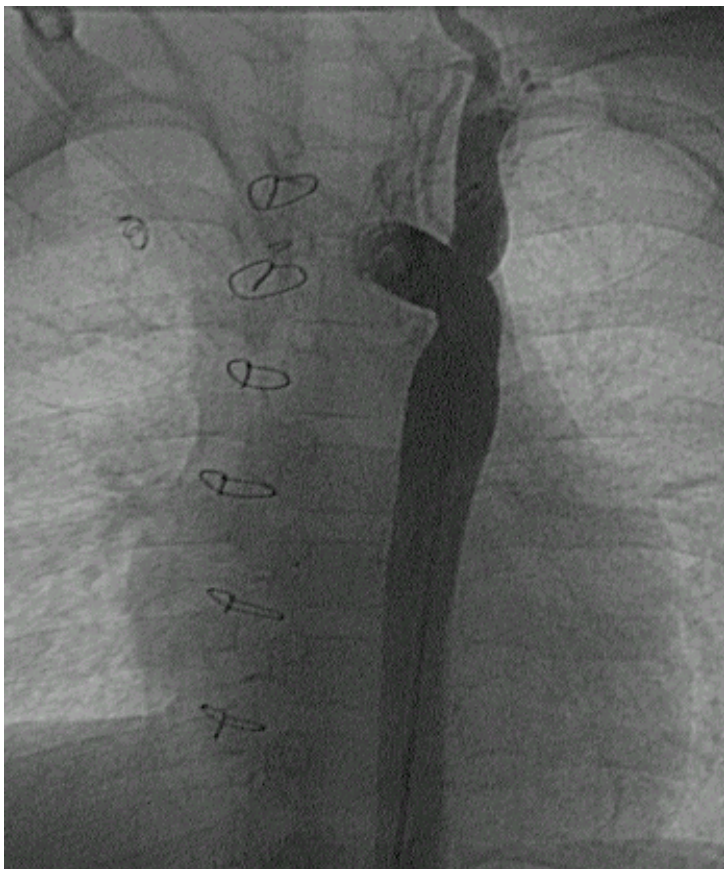
- Aortogram often fails to detect APCs
- Selective angiogram at innominate a. / SCA
- Sometimes diffuse network with multiple small feeding vessels



SAS (NDH; s/p DKS)



SAS (NDH; s/p DKS)



Fontan without Cath

(Ro PS,2004, JACC)

- Cath if positive any one of criteria

Table 1. Characteristics of Low-Risk Patients

Clinical data

Room air pulse oximetry $\geq 76\%$

Hemoglobin ≤ 18 g/dl

Echocardiographic data

Left pulmonary artery visualized without stenosis

No significant atrioventricular valve insufficiency ($\leq 1+$)

No significant ventricular dysfunction (qualitative)

No aortic coarctation

An unrestrictive atrial communication

No evidence of a decompressing vessel

Fontan in Single Lung

(Zachary CH, 1998 & Jacobs ML, 2004)

- M/C LPA loss following a Norwood op
- Usual criteria for hemodynamic acceptability
- Low SaO₂ but no difference in early mortality, length of hospitalization, of effusions
- High incidence of PLE; later fenestration
- All possible means of resuscitating the lost elements of the pulmonary vascular bed and re-establishing PA continuity
 - upper & lower pulmonary vein wedge angiogram
- Fenestrated Fontan if hemodynamically acceptable?

Fontan in Single Lung

(Zachary CH, 1998)

- 1st report in 1980 (Sade RM,1980)
- 7 pts with a single Rt lung undergoing Fontan op
- LPA loss following a Norwood op in 6
- no difference in preoperative hemodynamic data between single lung group and double lung group
- no op mortality, 2 late death (one recurrent PE, the other fulminant RSV infection)
- PLE in 2 of 5 survivors – one death
- 5 additional cases of one lung Fontan (Jacobs ML,2004, Seminars in thoracic & cardiovascular surgery); PLE in 3 patients
 - Totally 3 deaths (2 PLE), PLE in 4 of 9 survivors
 - Usual criteria for hemodynamic acceptability
 - High incidence of PLE

Fontan in Single Lung

- Recruitment methods
 - 1) Surgical shunt with cryopreserved saphenous vein graft as shunt material
 - 2) Catheter intervention (Moore JW,2002)
- The presence of only one PA does not in and of itself preclude satisfactory outcome.
- All possible means of resuscitating the lost elements of the pulmonary vascular bed and re-establishing PA continuity; upper & lower pulmonary vein wedge angiogram
- Suitability for one lung Fontan is based on hemodynamic measurements, just as in other cases.
- Fenestrated Fontan if hemodynamically acceptable?

Fontan in Single Lung

Recruitment by Catheter Intervention I

(Moore JW,2002)

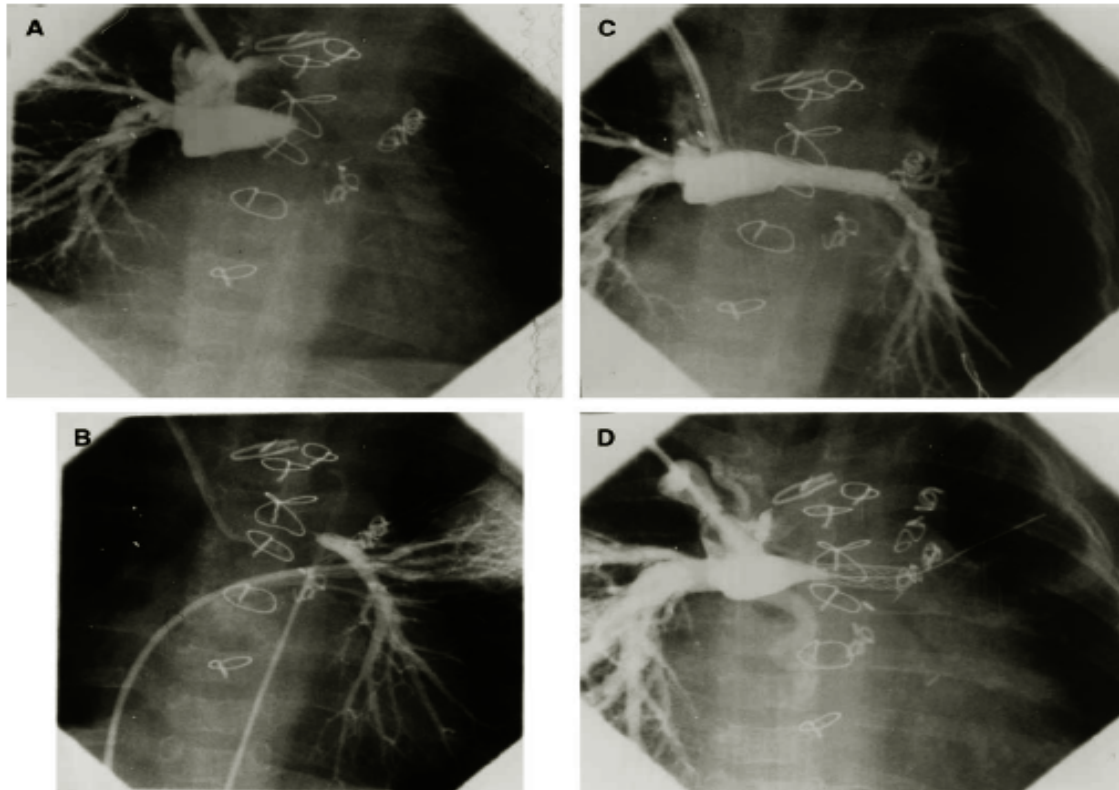


FIGURE 1. Angiograms showing results in patient 1. **A**, right pulmonary artery injection via superior vena cava; **B**, left pulmonary artery opacification via a left pulmonary vein wedge injection with catheter tip in hemi-Fontan reconstruction associated with the right pulmonary artery, demonstrating the long-segment (13 mm) discontinuity of the central pulmonary artery confluence; **C**, angiogram demonstrating patency of the central pulmonary artery confluence after placement of a P204 stent dilated to 5 mm; **D**, right pulmonary artery injection via superior vena cava performed at 3 months follow-up showing in-stent occlusion of previously reconstructed pulmonary arteries (with wire through region of occlusion).

Fontan in Single Lung

Recruitment by Catheter Intervention II

(Moore JW,2002)

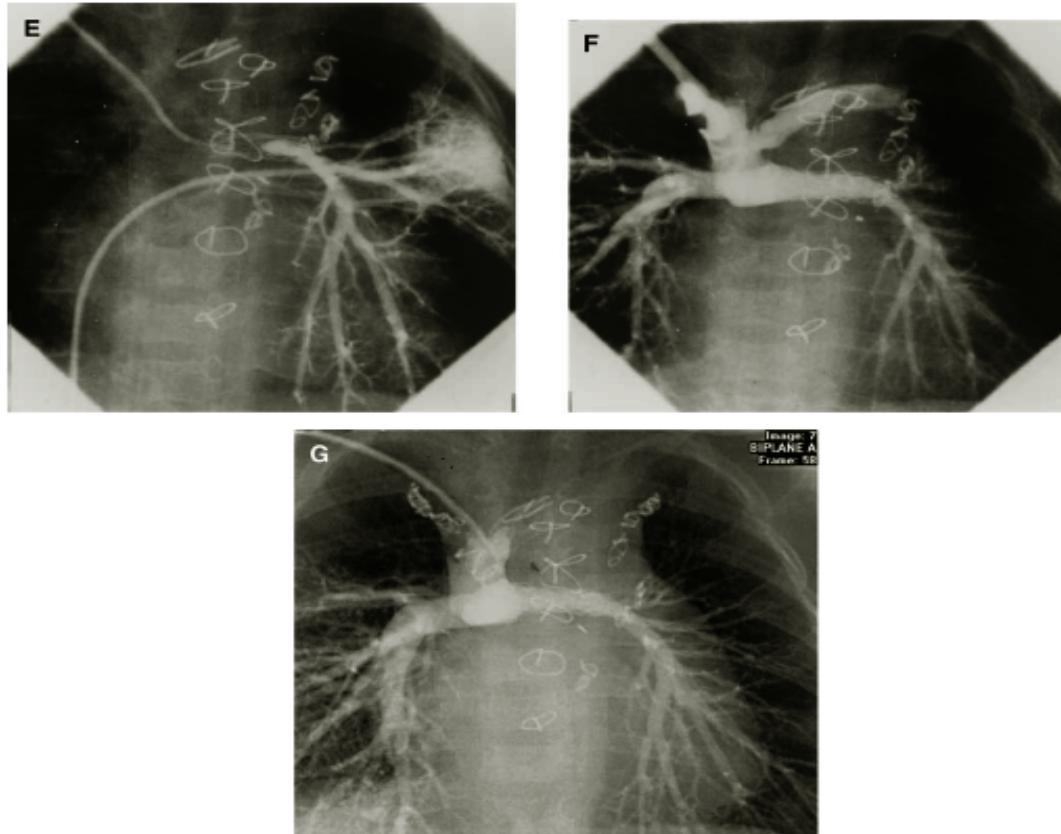
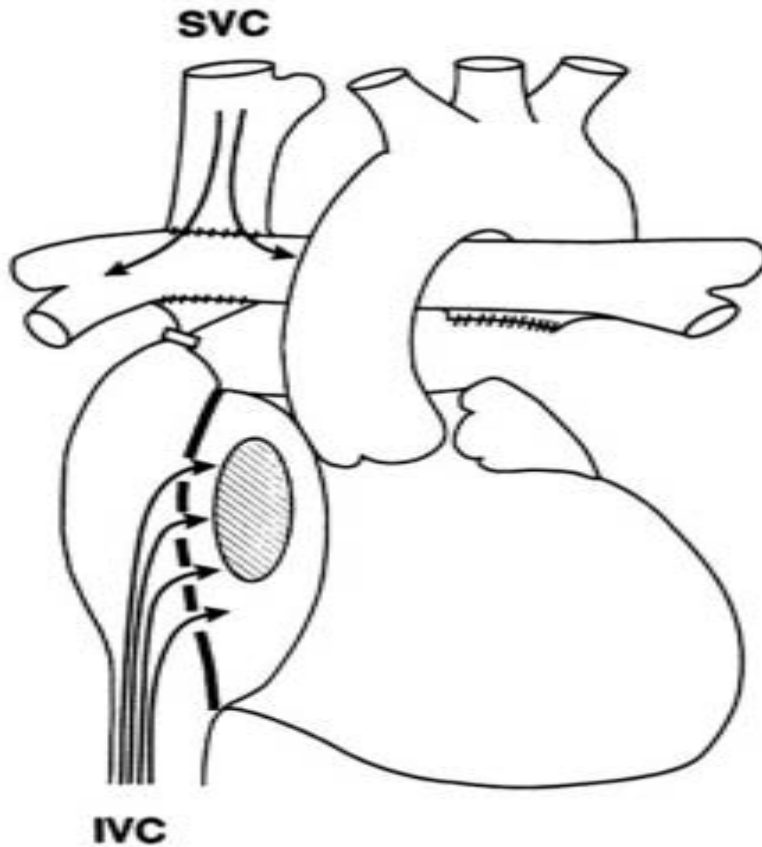


FIGURE 1 CONTINUED. E, left pulmonary artery opacification via a left pulmonary vein wedge injection performed at 3 months follow-up confirming in-stent occlusion; F, angiogram demonstrating patency of the central pulmonary artery confluence after angioplasty and placement of an additional P204 stent dilated to 7 mm; G, follow-up angiogram recorded 11 months after placement of second stent showing long-term patency of reconstructed segment.

Nonsurgical Completion of Fontan

(Klima U, 2000)



(Konertz W, 1995)

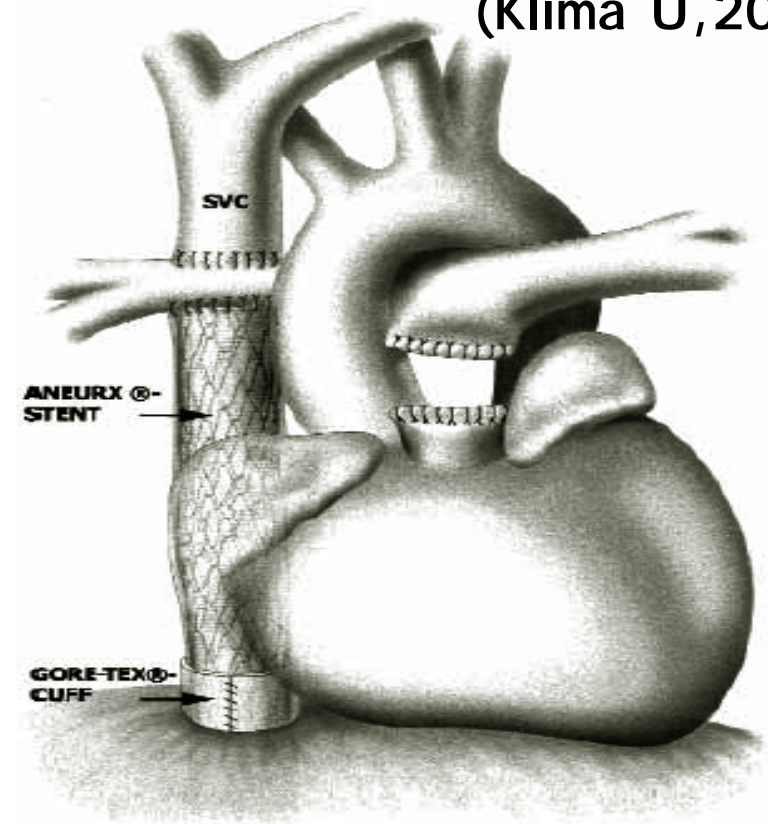


Fig 1. Sketch of the combined operative and interventional procedure. A bidirectional Glenn anastomosis is established, and the Aneurx stent is deployed. A small PTFE (*Gore-Tex*) cuff around the IVC is used as resistance to safely anchor the stent.