

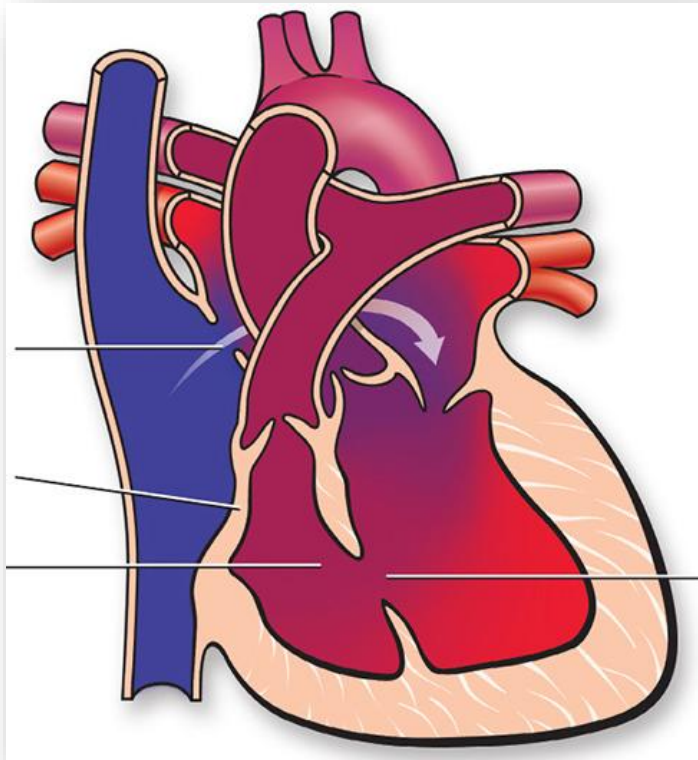
Functional Single Ventricle (FSV)

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Terminology



- Single ventricle
- Univentricular heart
- Functional Single Ventricle (FSV)
- Functionally Univentricular Heart

Diagnosis

Tricuspid Atresia

Mitral Atresia

Double Inlet left ventricle

Double inlet right ventricle

PA with intact ventricular septum

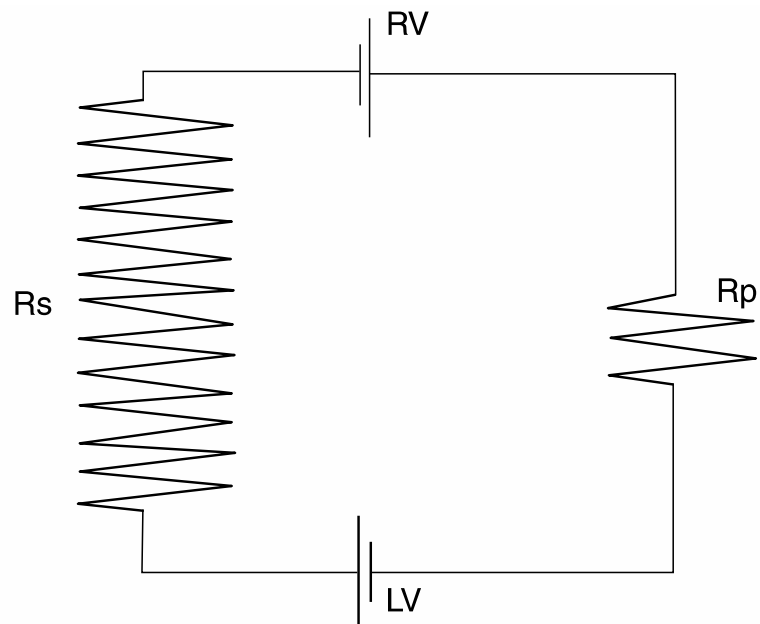
Hypoplastic left heart syndrome

Unbalanced AV canal

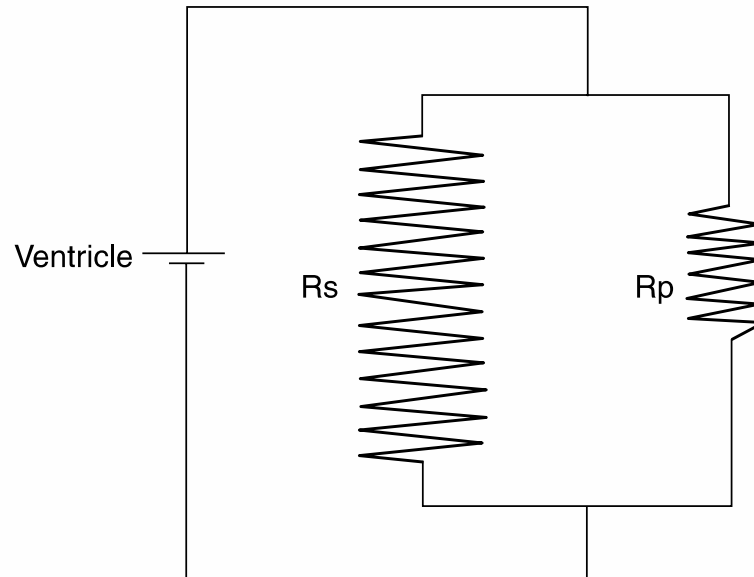
Heterotaxia syndrome

Others

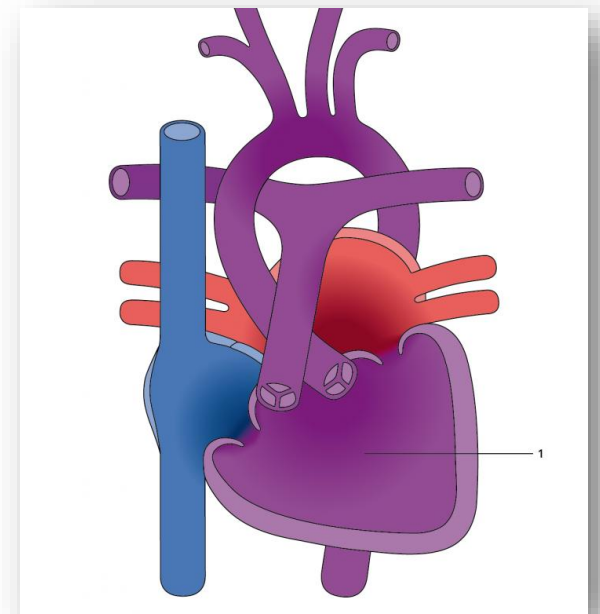
BV vs. FSV



Normal circulation: **serial**

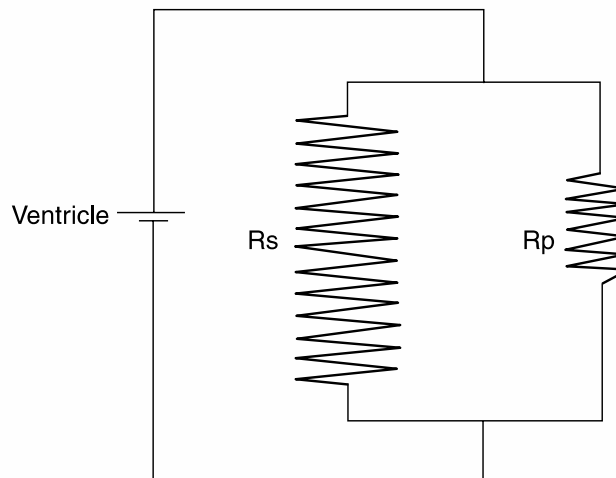


FSV circulation: **parallel**

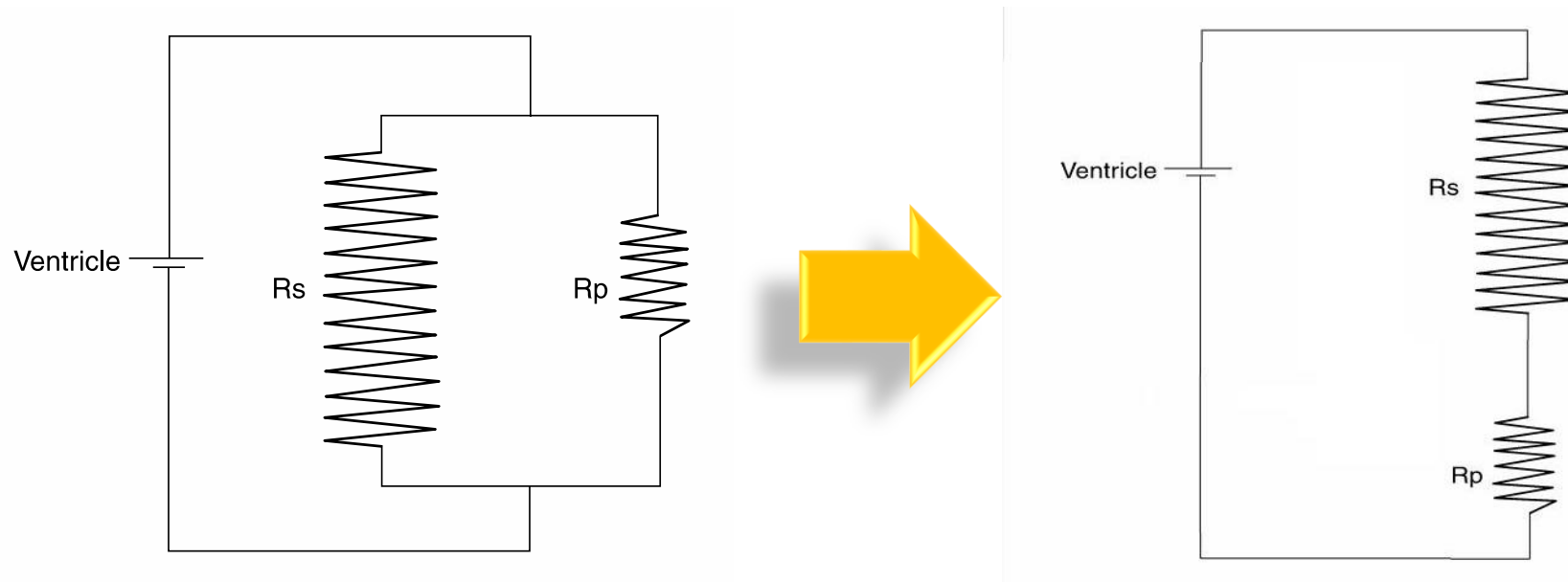


Parallel circulation

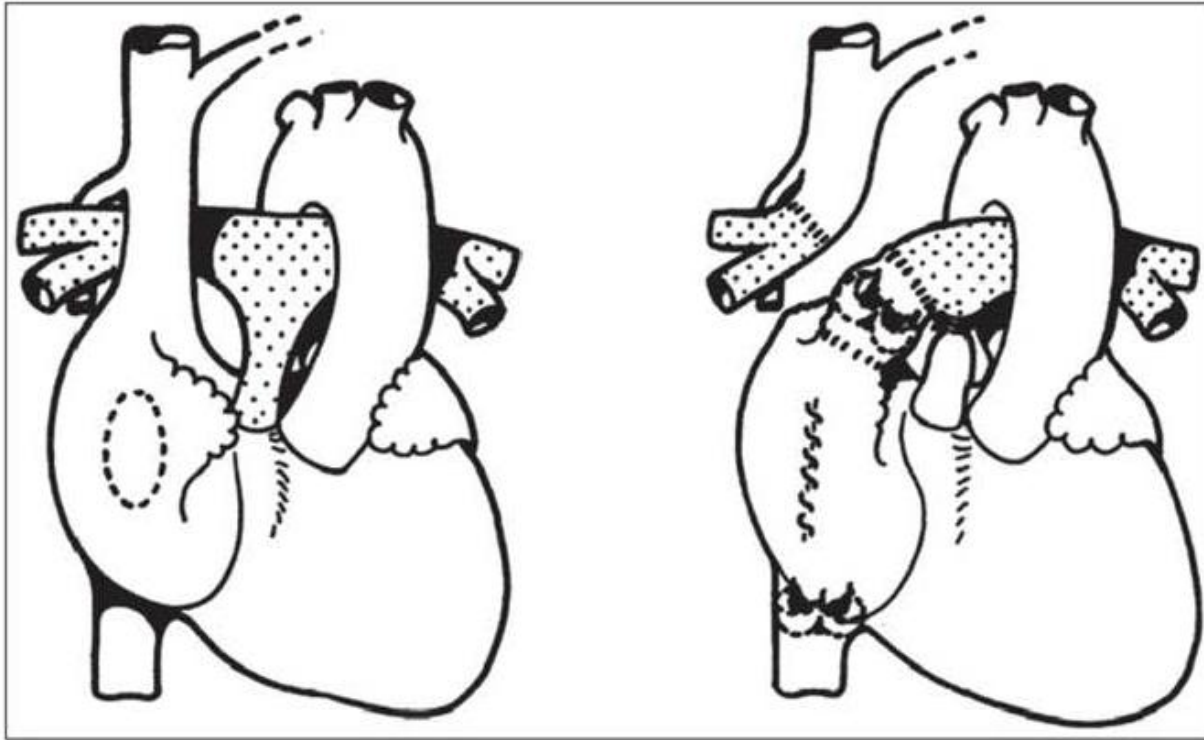
- Disadvantages
 - Arterial desaturation
 - Chronic ventricular volume overload



Treatment of FSV: Parallel to Serial



Fontan operation

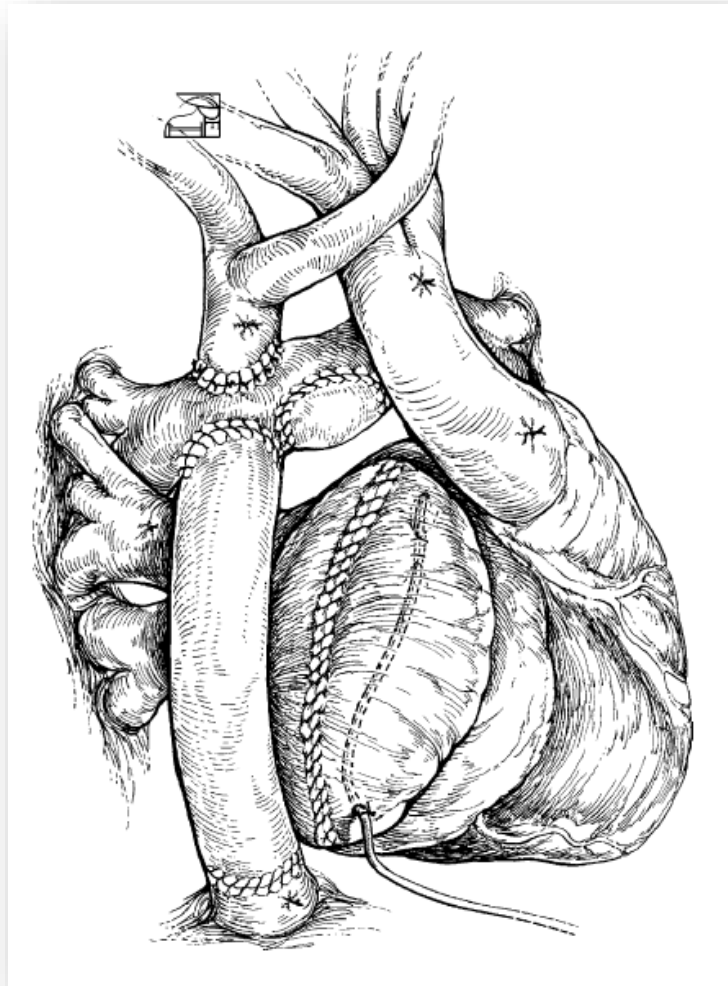


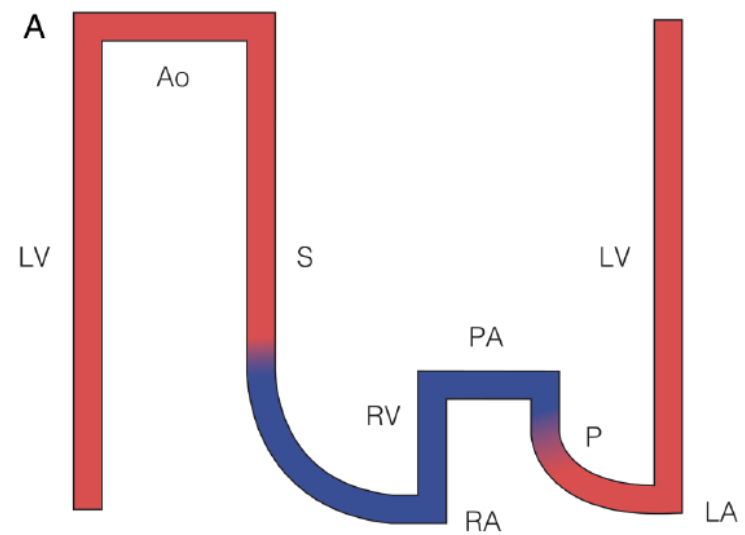
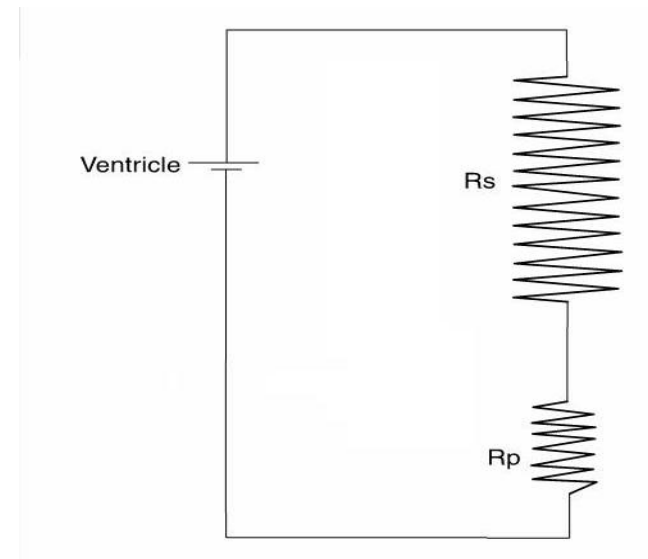
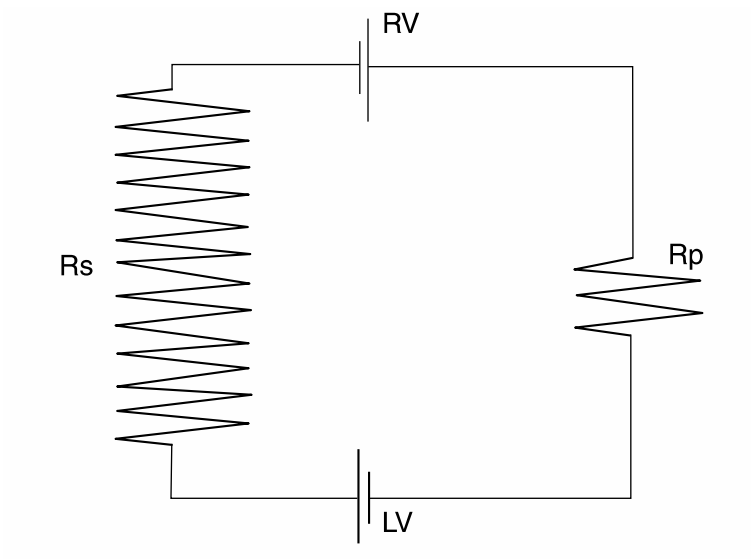
Thorax (1971);26:240-48



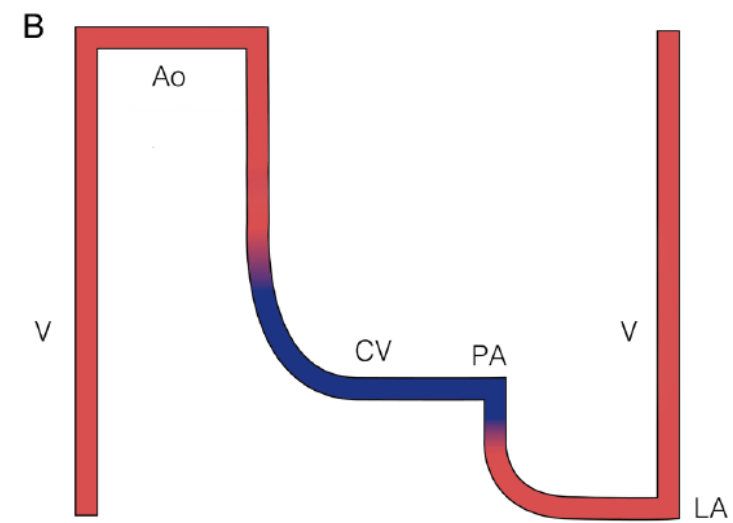
Dr. Francis Fontan

Extracardiac conduit Fontan





Normal circulation



Fontan circulation

Fontan palliation

- Disadvantages
 - **Systemic venous congestion (hypertension)**
 - PLE (protein losing enteropathy)
 - Plastic bronchitis
 - Lymphatic disorder
 - **Decreased cardiac output**



Current practice for patients with FSV

Fontan circulation

- Success depends on
 - Adequate pulmonary artery architecture
 - Low pulmonary vascular resistance
 - Good ventricular function
 - Competent valve

SUCCESS NEEDS
PREPARATION



Multi-stage palliation



- PAB
- Shunt
- Observation

- BCPS

- Fontan

Multi-stage palliation

- First-stage palliation
 1. Controlled source of pulmonary blood flow
 2. Unobstructed systemic outflow
 3. Unobstructed pulmonary venous return

Multi-stage palliation

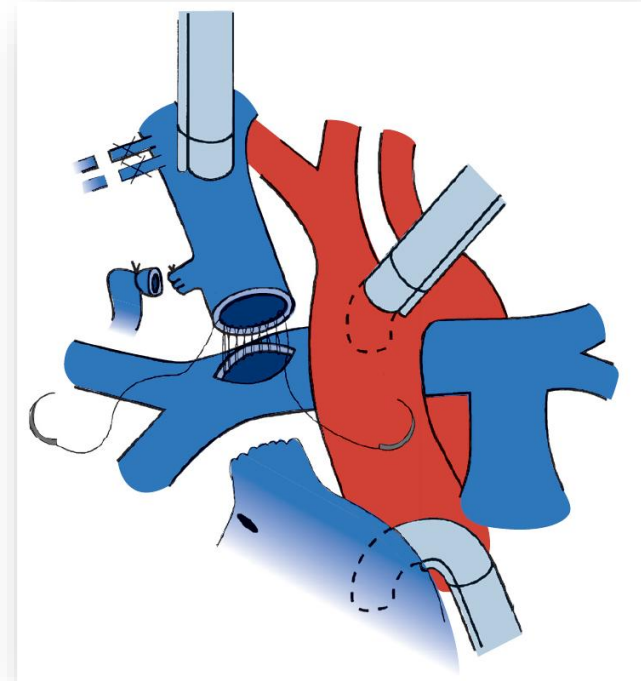
- First-stage palliation
 1. Controlled source of pulmonary blood flow
 - Unrestricted: PAB
 - Severely restricted: mod. B-T shunt
 - Balanced (naturally): observation

Multi-stage palliation

- First-stage palliation
 2. Unobstructed systemic outflow
 - Coarctation of the aorta: **arch repair**
 - LVOT obstruction: **DKS, etc.**
 3. Unobstructed pulmonary venous return
 - Obstructed TAPVR: **TAPVR repair**

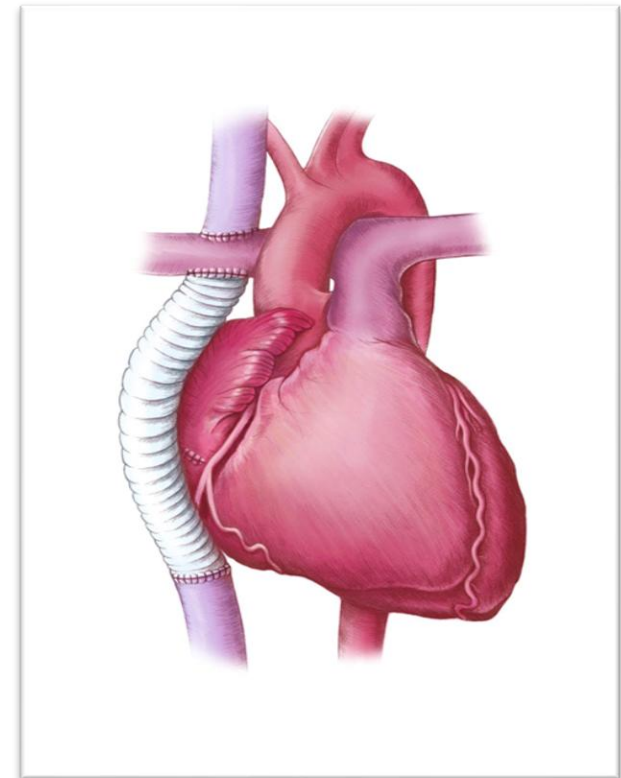
Multi-stage palliation

- Second-stage palliation
 - BCPS (Glenn)
 - Around 6 months of age
 - Improved Fontan outcomes
 - cf) non-pulsatile vs. pulsatile

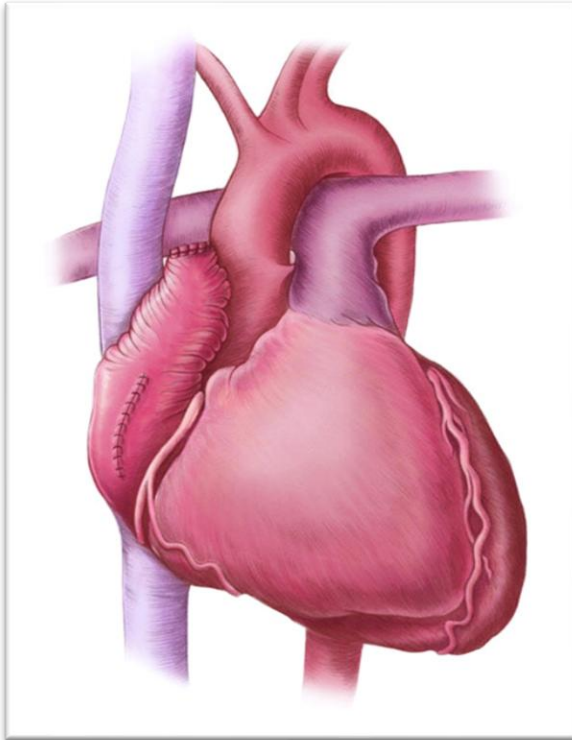


Multi-stage palliation

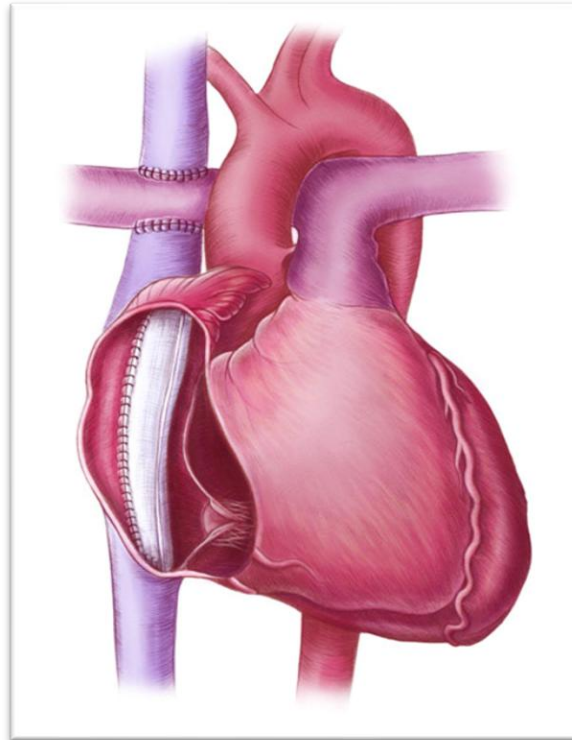
- Last-stage palliation
 - Fontan operation
 - Around 2-4 years of age
 - Improved oxygen saturation



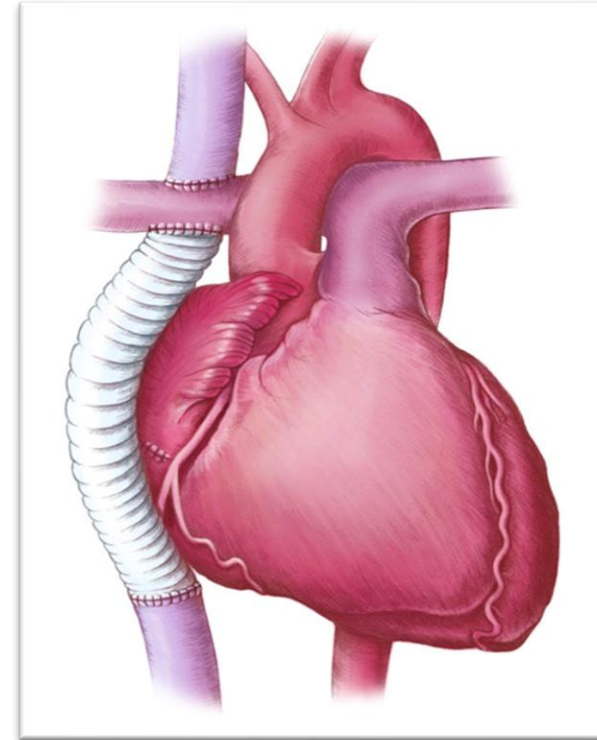
Fontan operation



**Atrio-pulmonary
(AP)**



**Lateral tunnel
(LT)**



**Extracardiac conduit
(ECC)**

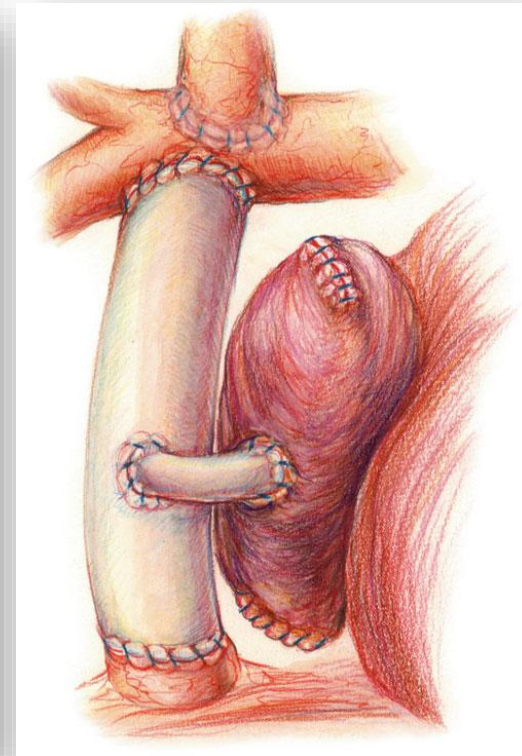
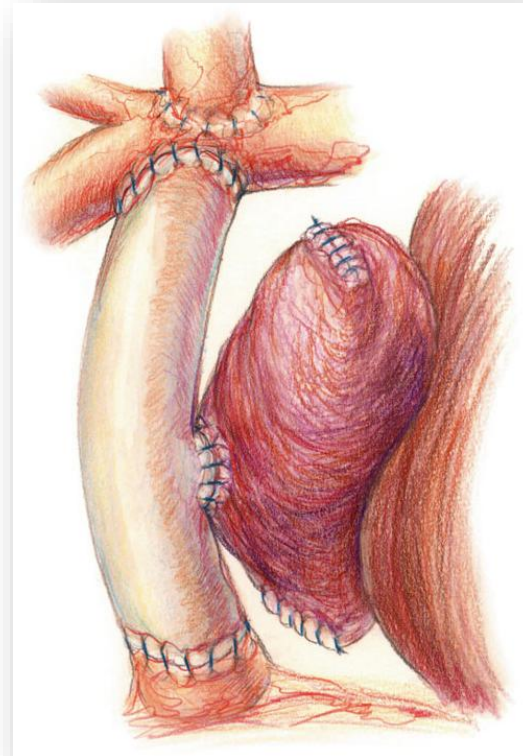
Fontan operation

- Risk stratification
 - ECG, EchoCG, CT (MRI) / **Cardiac Catheterization**

	Low Risk	High Risk
PVR	$< 2 \text{ WU} \cdot \text{m}^2$	-
PAP	-	$> 15 \text{ mmHg}$
PA anatomy	Normal	-
Ventricular function	Normal	-
AV valve regurgitation	-	$\geq$ moderate
Rhythm	Normal	-
AP collaterals	-	Abundant

Fontan fenestration

- Pros
 - lowering Fontan pressure / Increasing preload
 - Decreased pleural effusion
- Cons
 - Desaturation
- Indications
 - High-risk patients
 - Routine





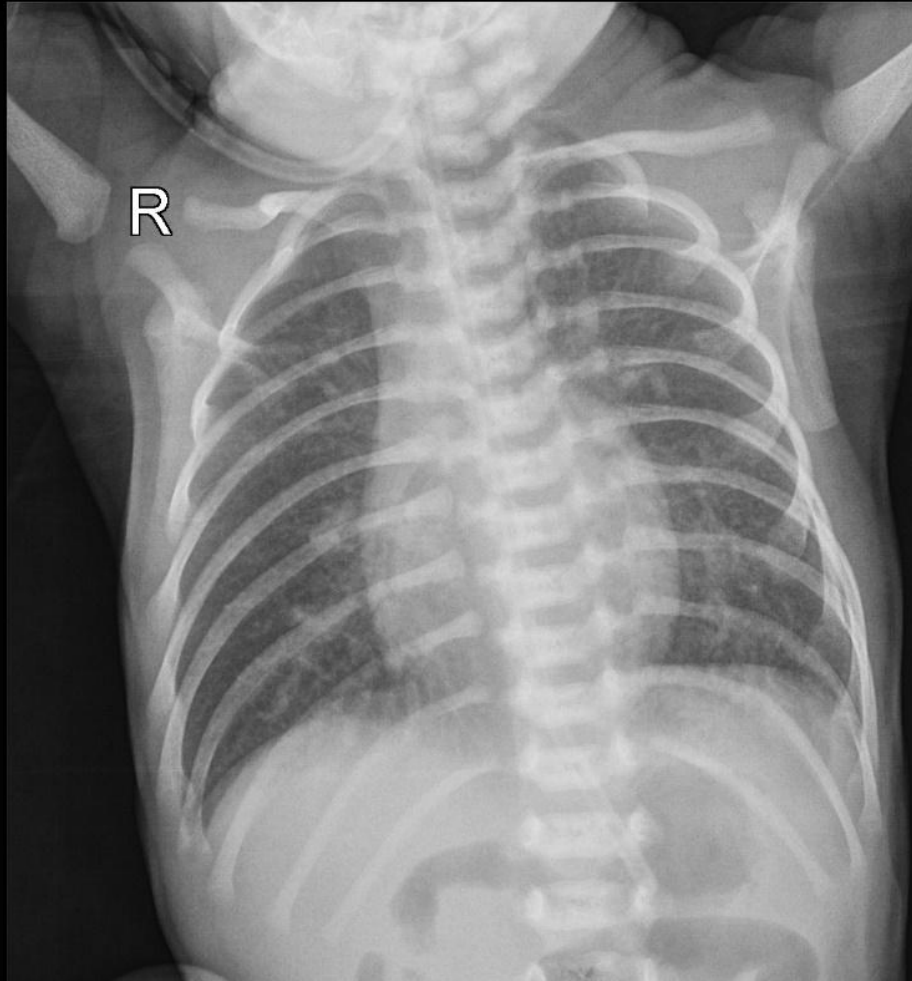
CASE PRESENTATION

Case

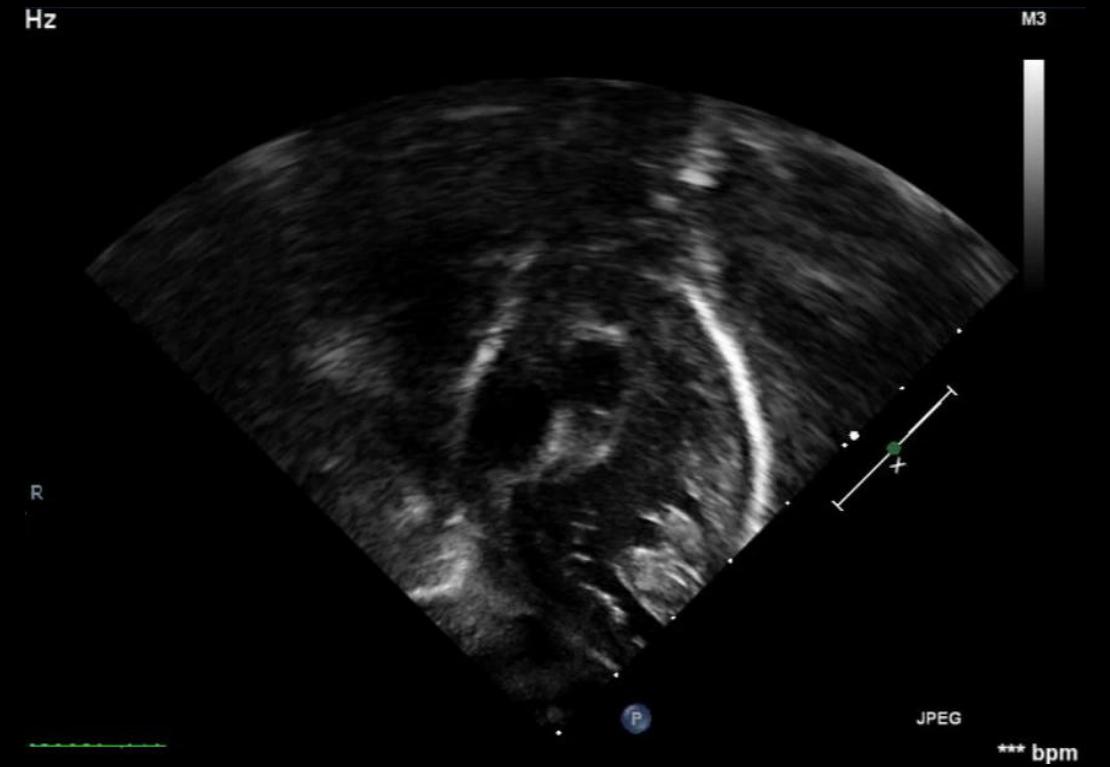
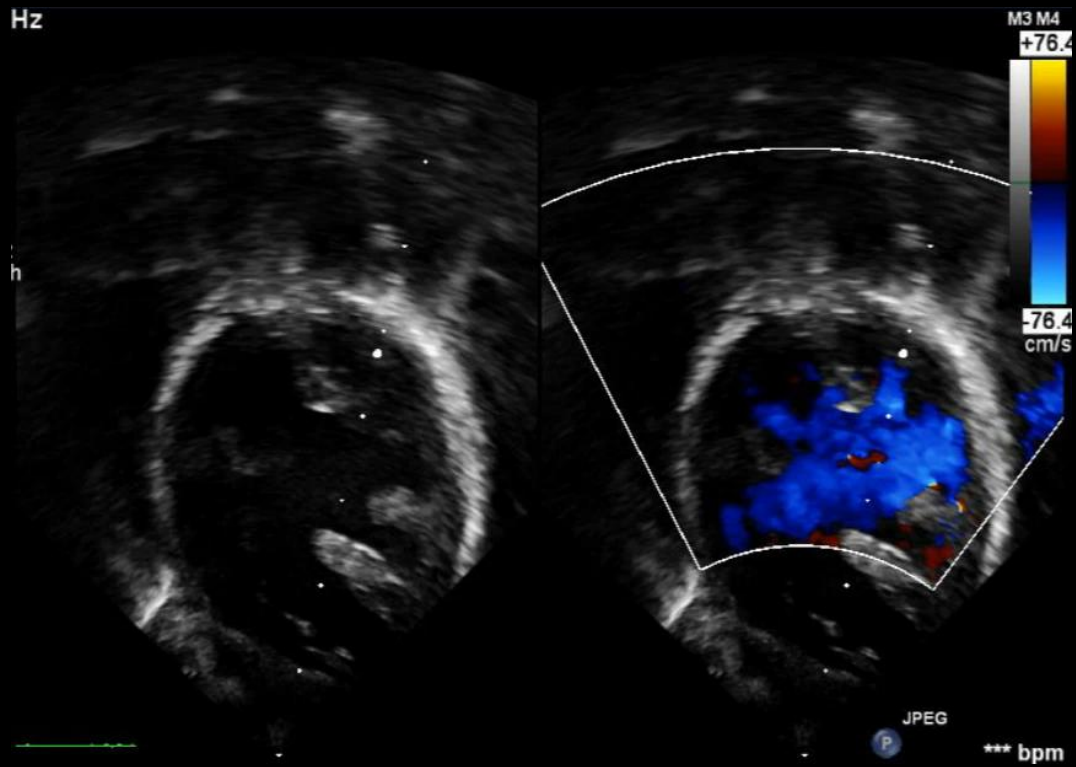
- GA: 37+4 wks, 2.5kg, male
- Duodenal atresia



After birth



After birth



After birth



Case

- GA: 37+4 wks, 2.5kg, male
- Duodenal atresia
- Diagnosis
 - FSV
 - Heterotaxy syndrome (RAI), DORV, AVSD
 - TAPVR (cardiac, nonobstructed)

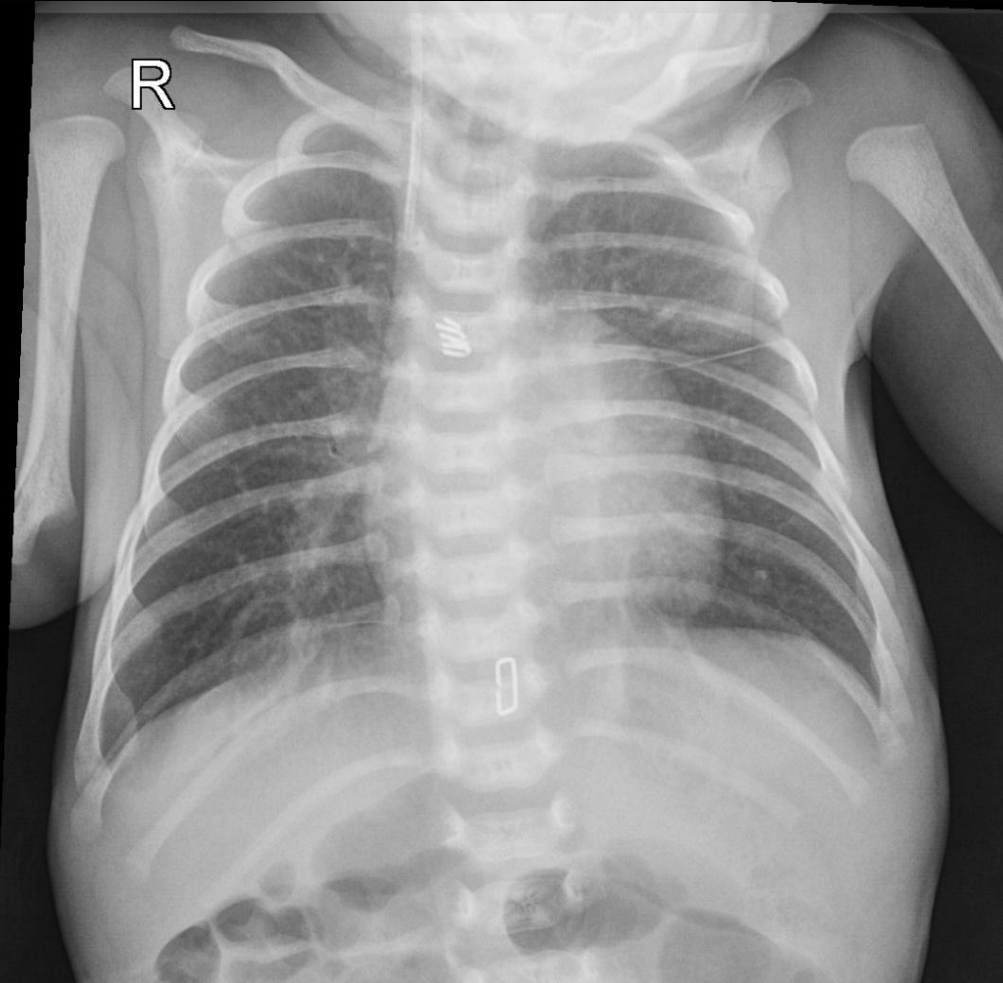


Case

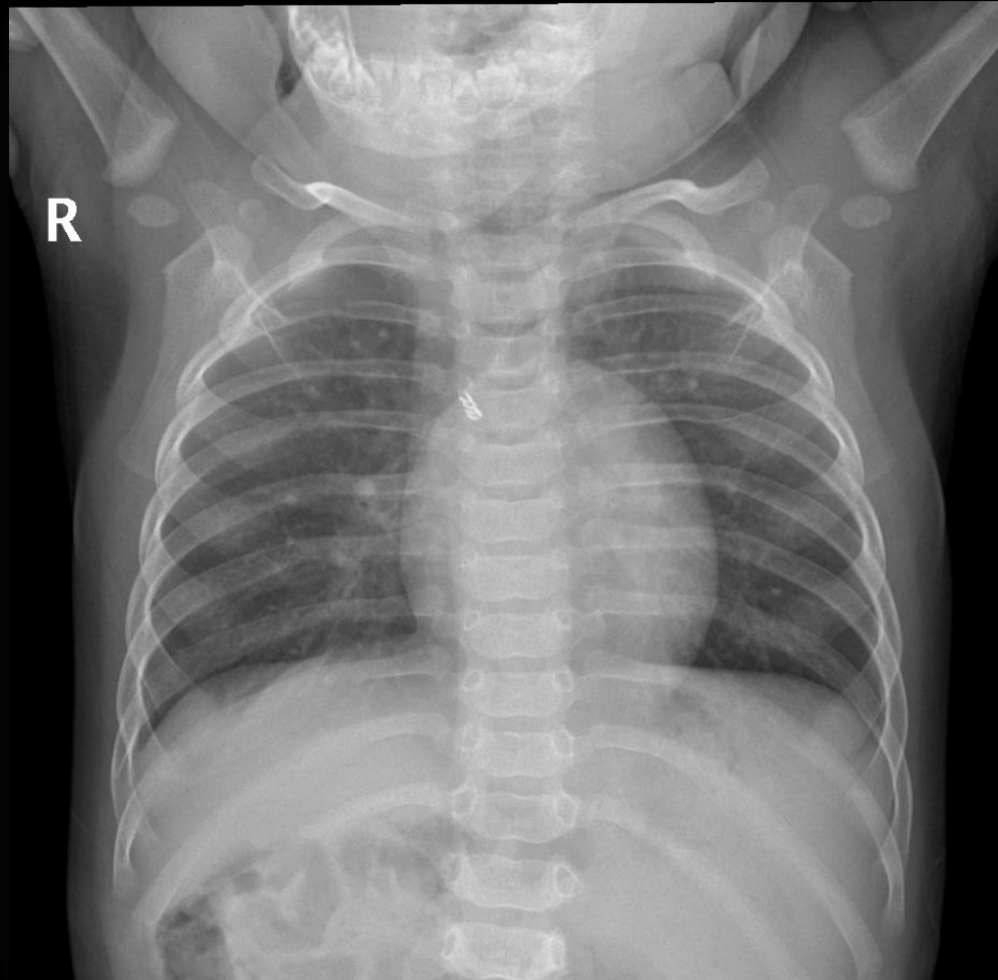
- GA: 37+4 wks, 2.5kg, male
- Diagnosis
 - FSV: Heterotaxy syndrome (RAI), DORV, AVSD, no PS
- PAB at 10 days old



Post PAB



Pre BCPS

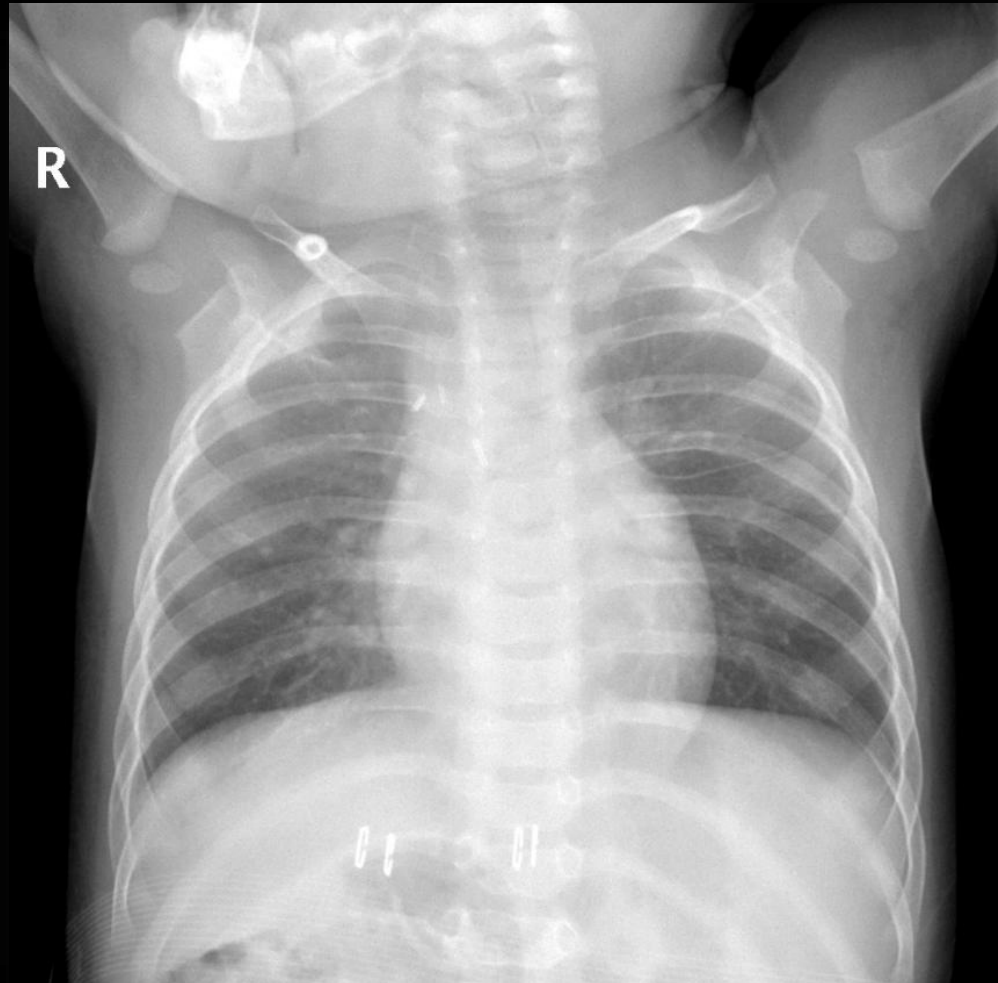


Case

- Diagnosis
 - FSV (Heterotaxy syndrome, DORV, AVSD)
- PAB at 10 days old
- BCS, LPA angioplasty at 8 months of age



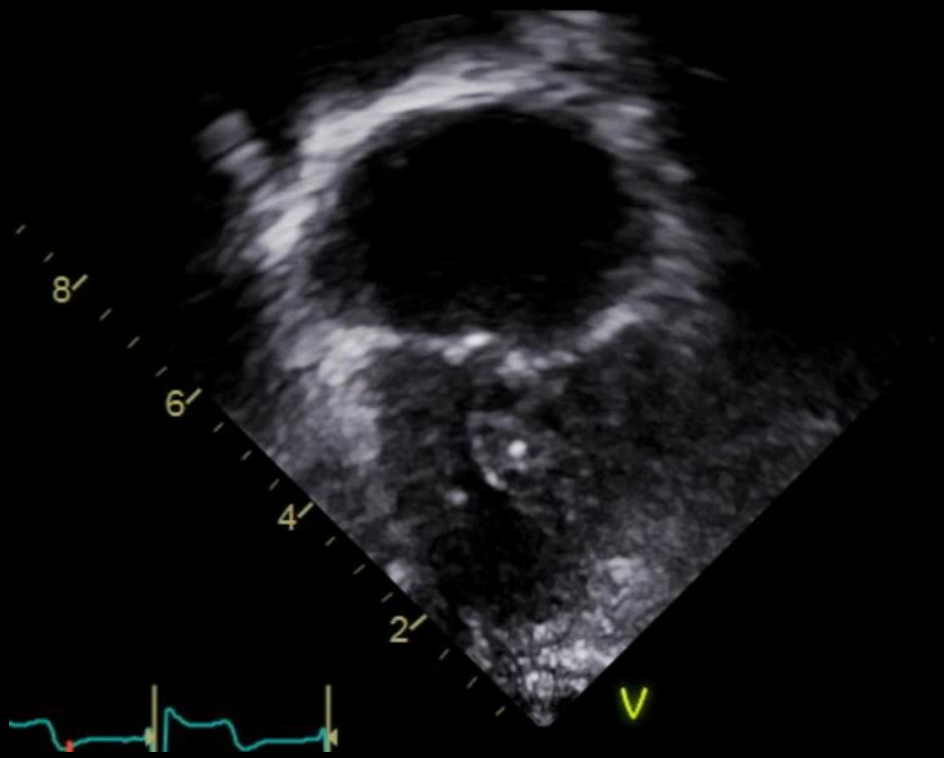
Post BCPS



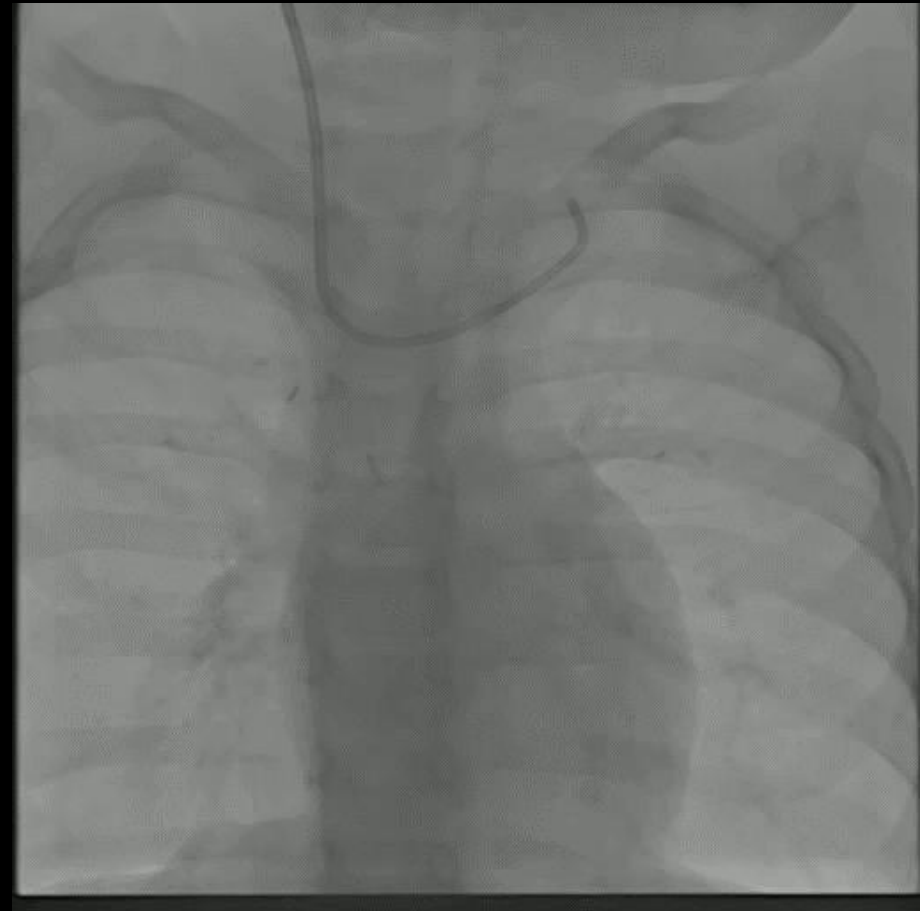
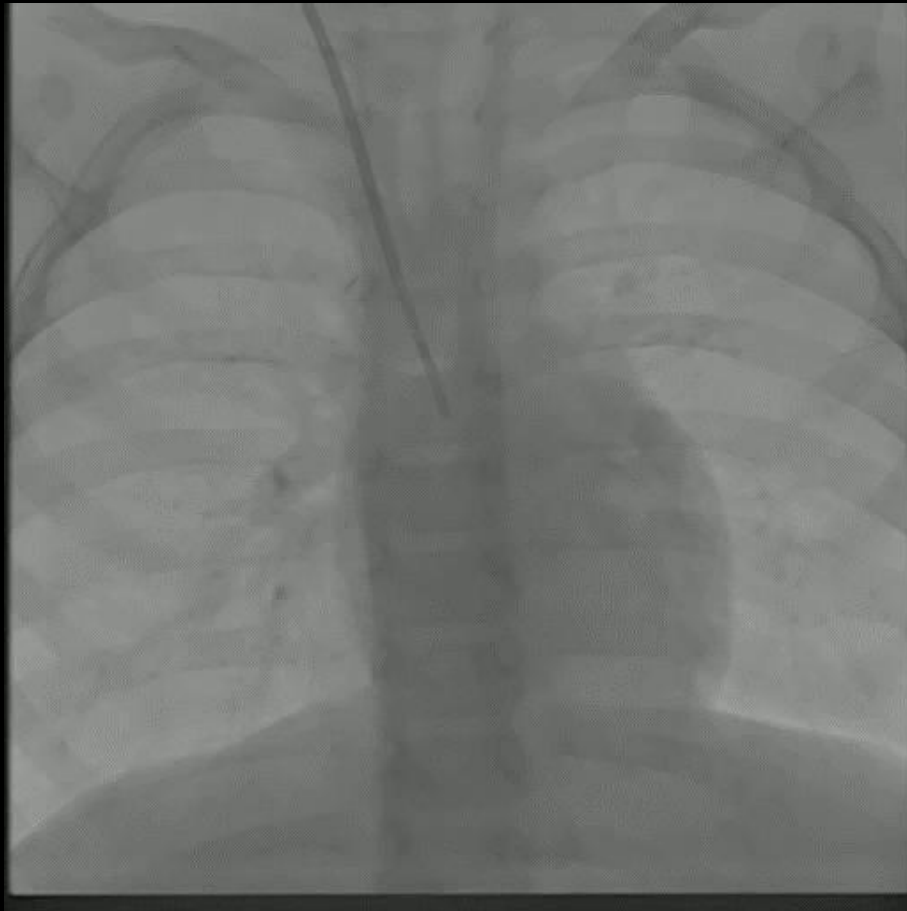
Pre Fontan



Pre Fontan



Pre Fontan



Pre Fontan

Precath Diagnosis

RAI, cAVSD, DORV, TAPVR

s/p PAB, PDA ligation(2017/11/13)

s/p BCPS, LPA angioplasty(2018/07/09)

<cath note> Pre-Fontan study

*pressure tracing

BCS>LPA>RPA, Dao>LV

*Angiography

BCS, Aortic arch (AP/Lat)

* Cath finding >

1. Finding

- ventricular function good.

RVEDP : 12mmHg

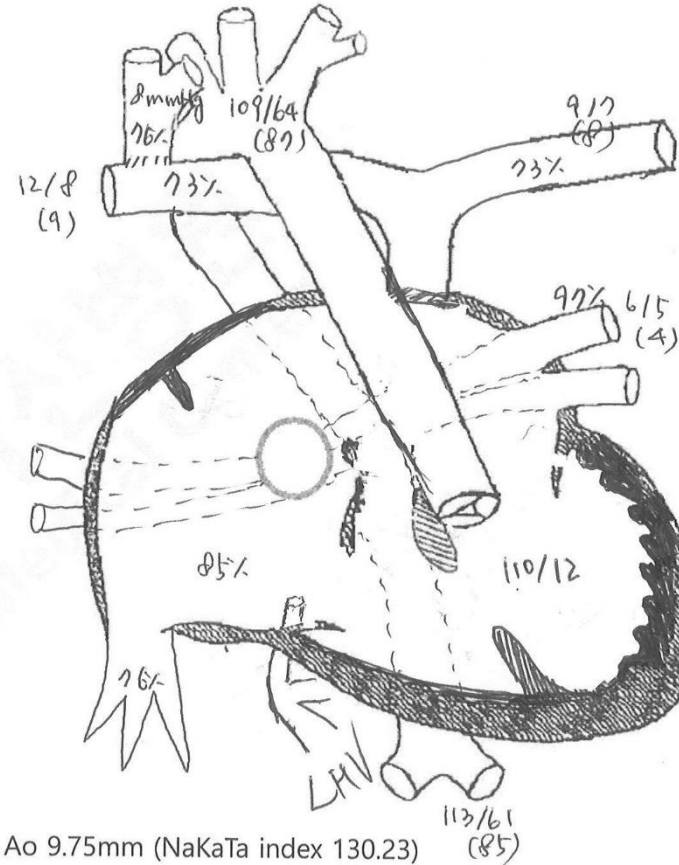
- Wide VSD

- branch PAs sizable : RPA 10.39mm, LPA 8.9mm, thoracic Ao 9.75mm (NaKaTa index 130.23)

- right BCS flow patent

- low PA pressure : mPAP = 9mmHg, Rp 1.36

* TPG = 4mmHg

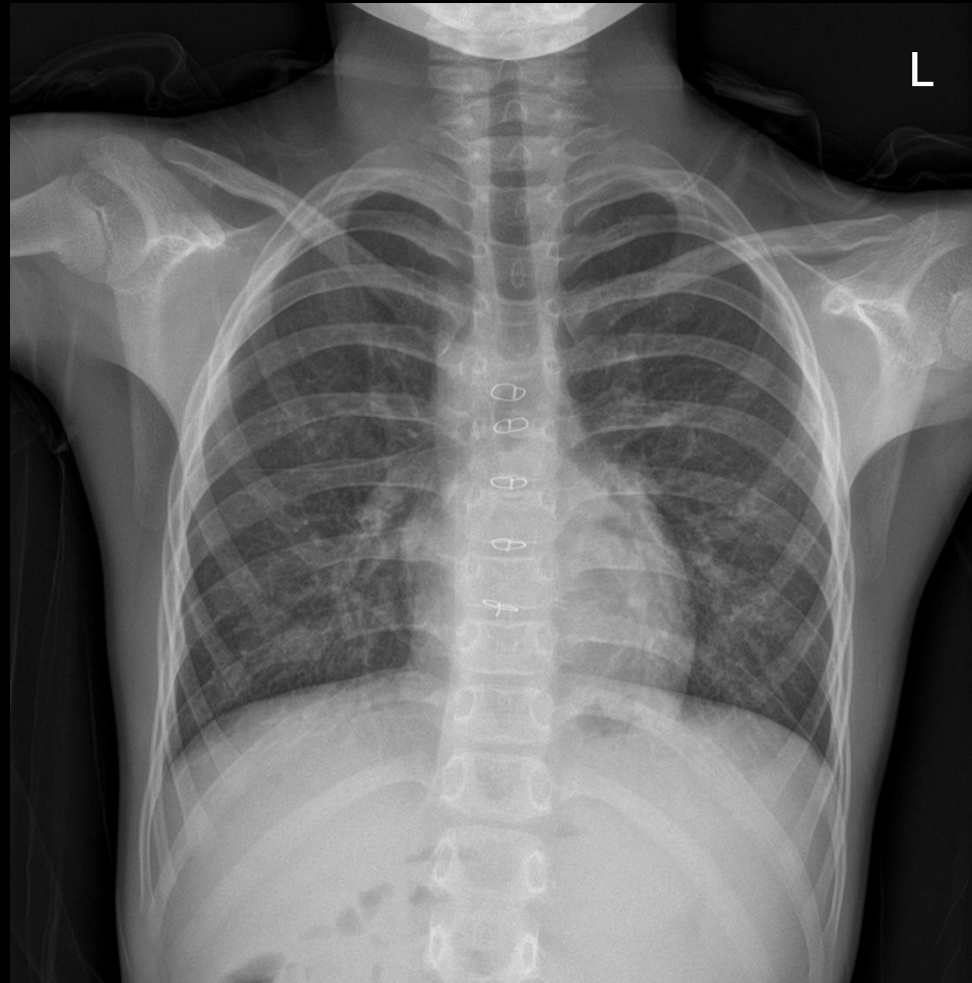


Case

- Diagnosis: FSV (Heterotaxy syndrome, DORV, AVSD)
- PAB at 10 days old
- BCS, LPA angioplasty at 8 months old
- ECC Fontan operation at 3 years old
 - 18mm PTFE vascular graft
 - Fenestration (4mm)



CXR at 8 years of age



Thank you for your attention

