Functional Single Ventricle (FSV)

Eun Seok Choi

Division of Pediatric Cardiac Surgery, Asan Medical Center,

University of Ulsan College of Medicine, Seoul, Korea.

서울아산병원
소아청소년병원 심장외과
Asan Medical Center Children's Hospital Cardiac Surgery

I LOVE HEART
Asan Medical Center
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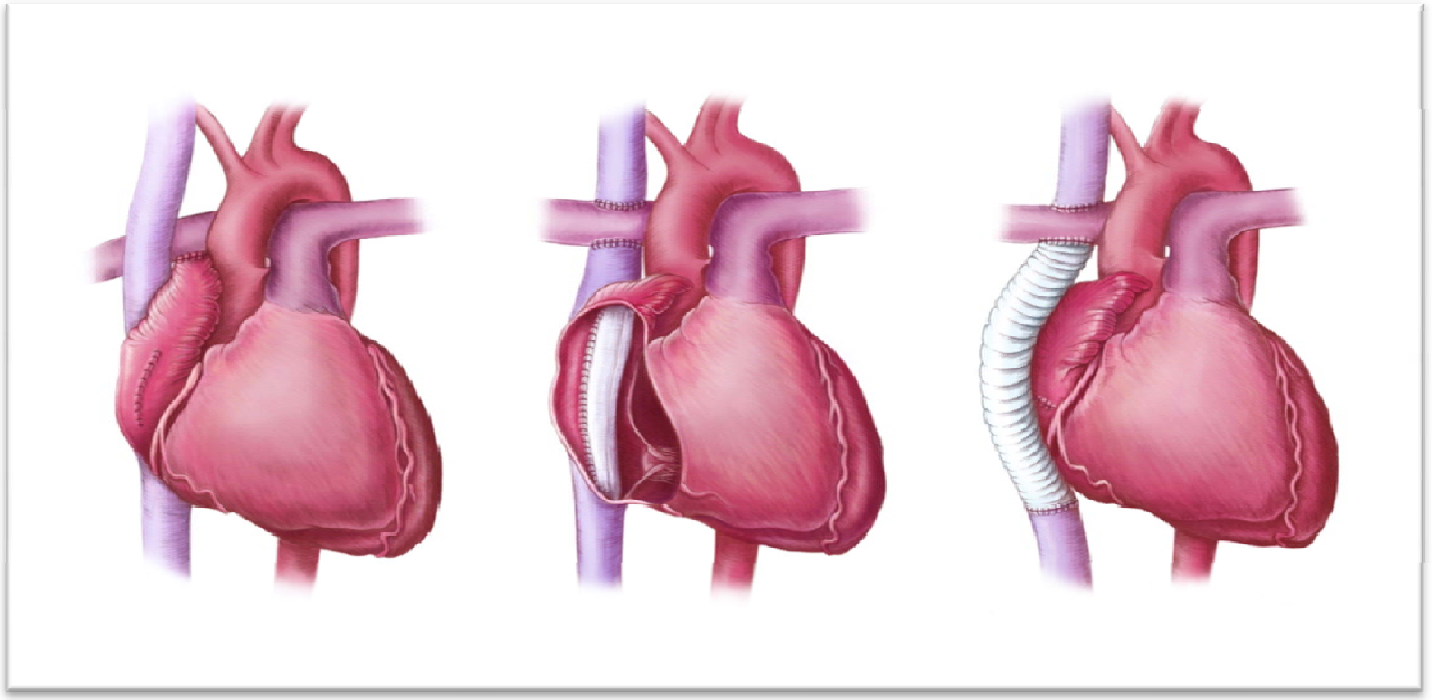
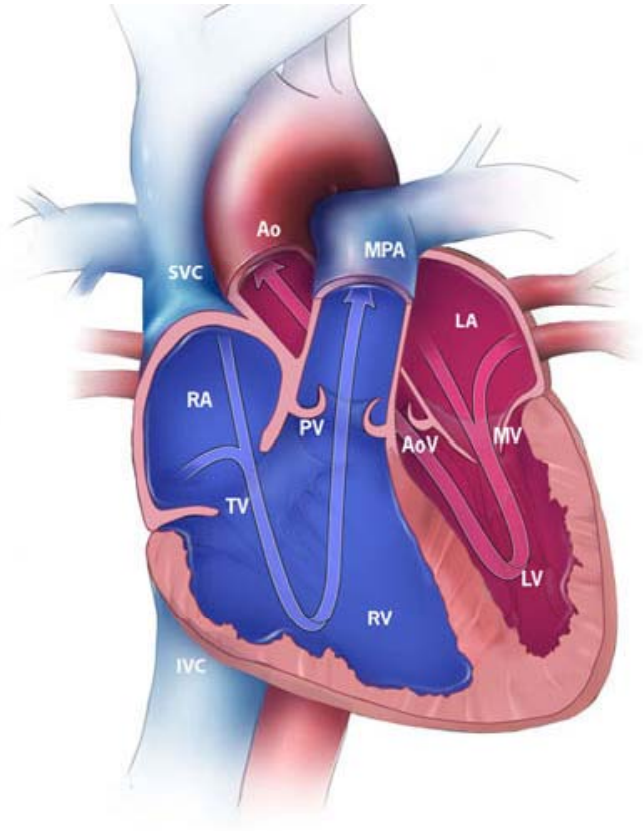
COI Disclosure

The authors have no financial conflicts of interest to disclose concerning the presentation

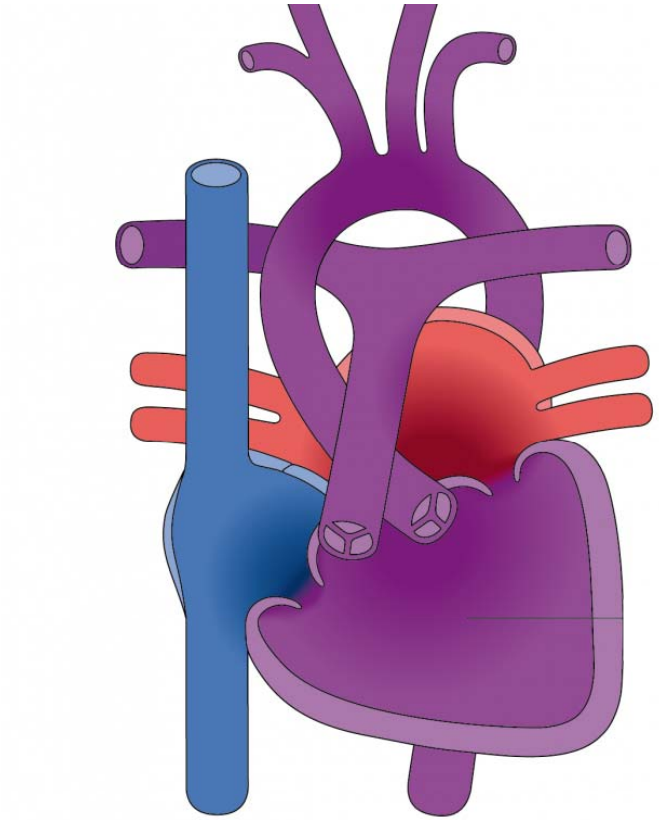


FONTAN CIRCULATION

Terminology



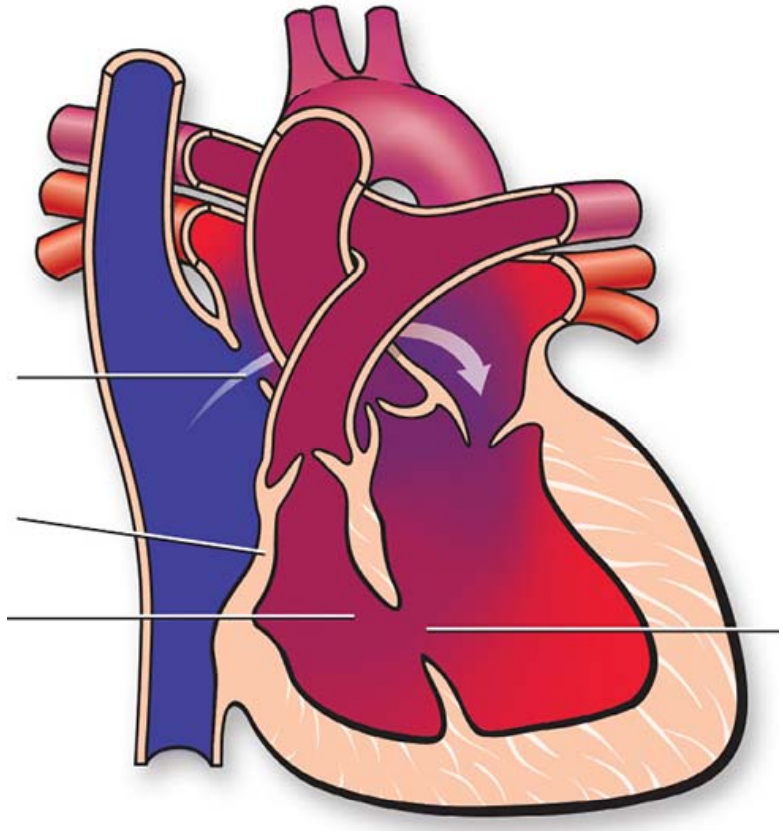
Normal Heart



Single ventricle

Univentricular heart

Terminology



Functional Single Ventricle

Functionally Univentricular Heart

BV vs. FSV

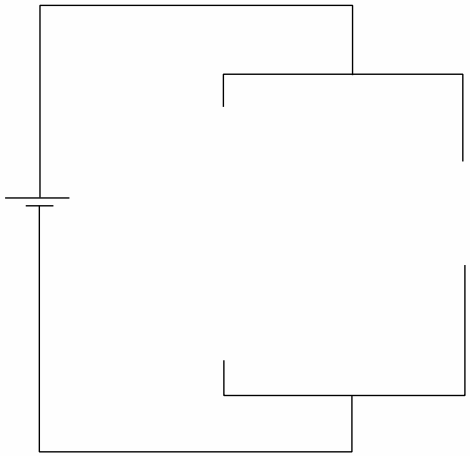
Normal circulation: serial

FSV circulation: parallel

Heart 2016;102:1081-1086

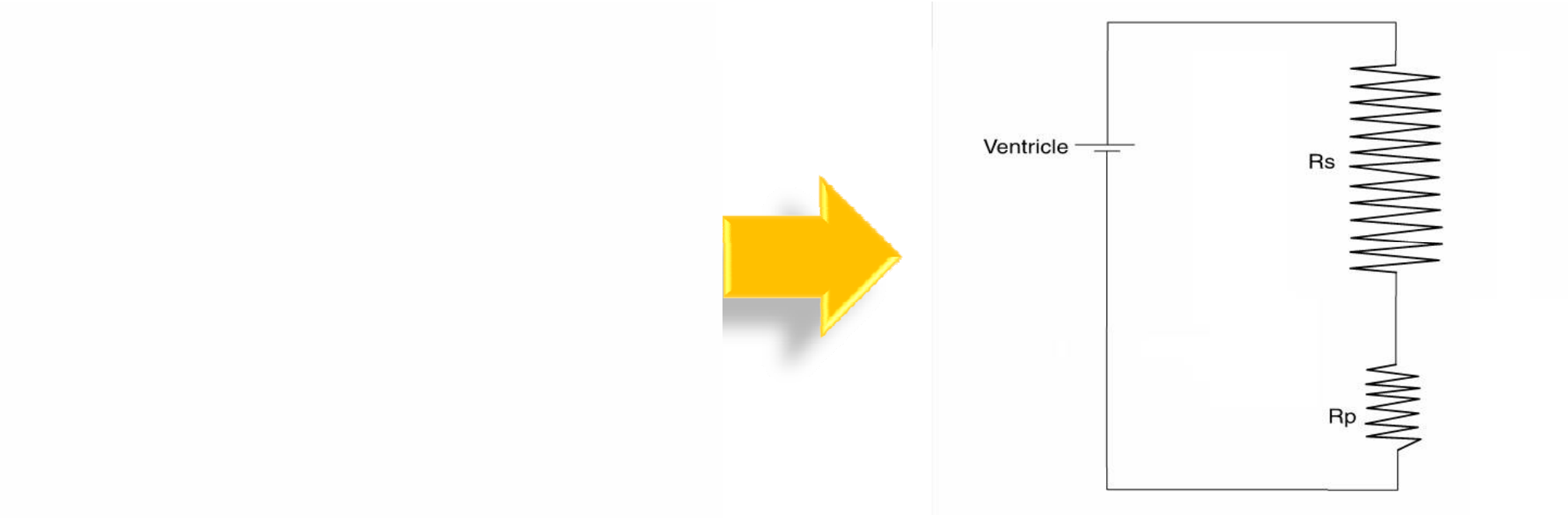
Parallel circulation

- Disadvantages
  - Arterial desaturation
  - Chronic ventricular volume overload

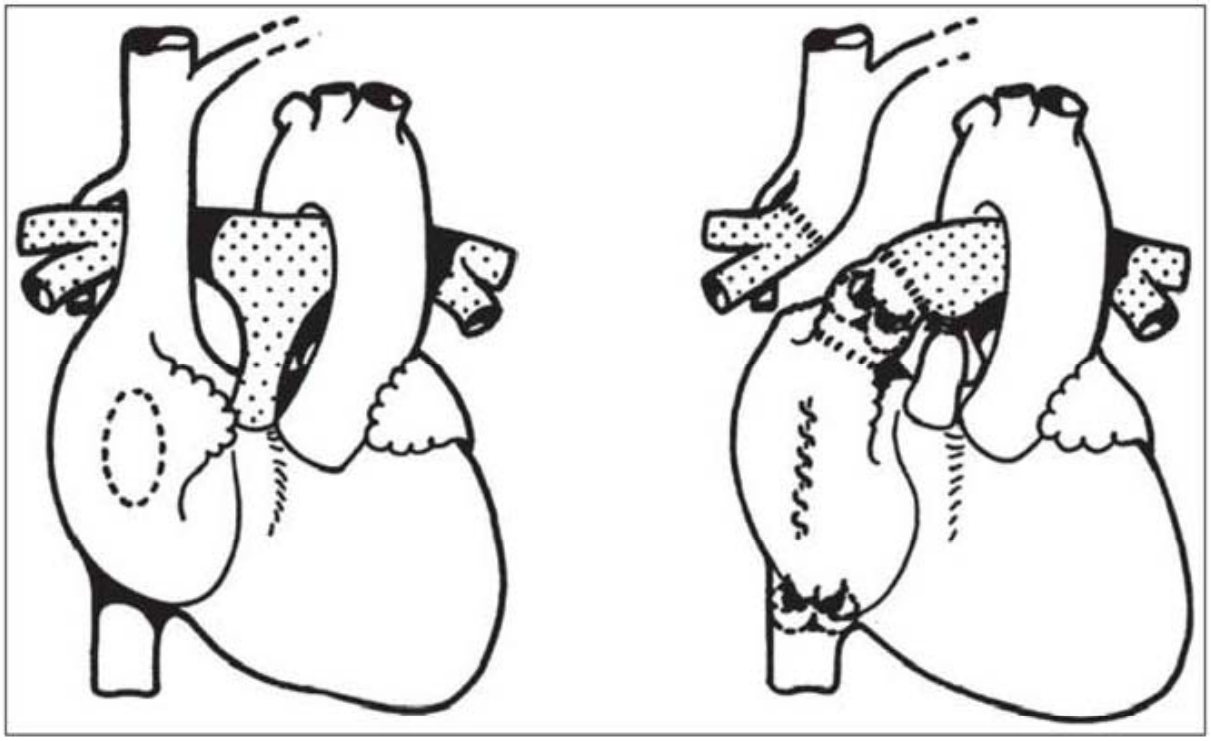


Heart 2016;102:1081-1086

FSV: Parallel to Serial



Fontan operation

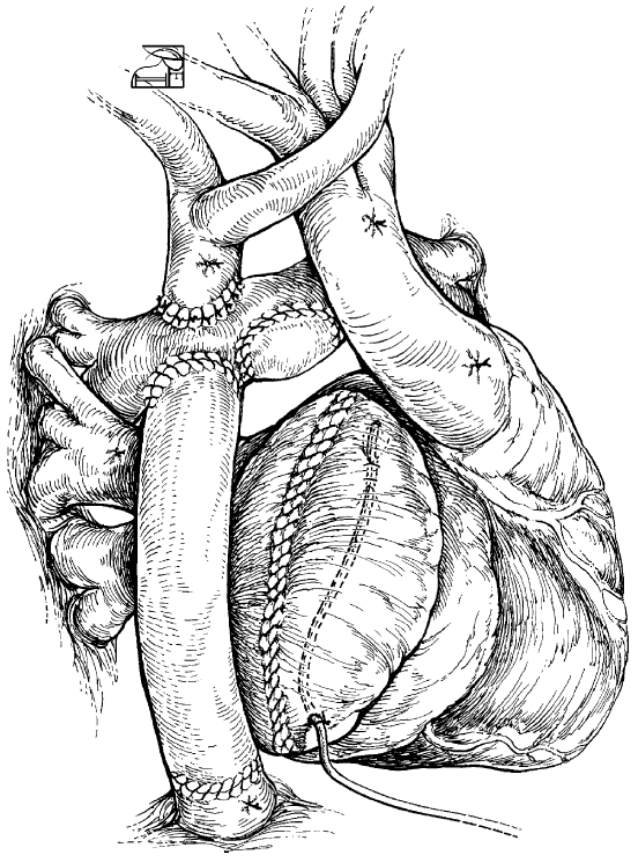


Thorax (1971);26:240-48

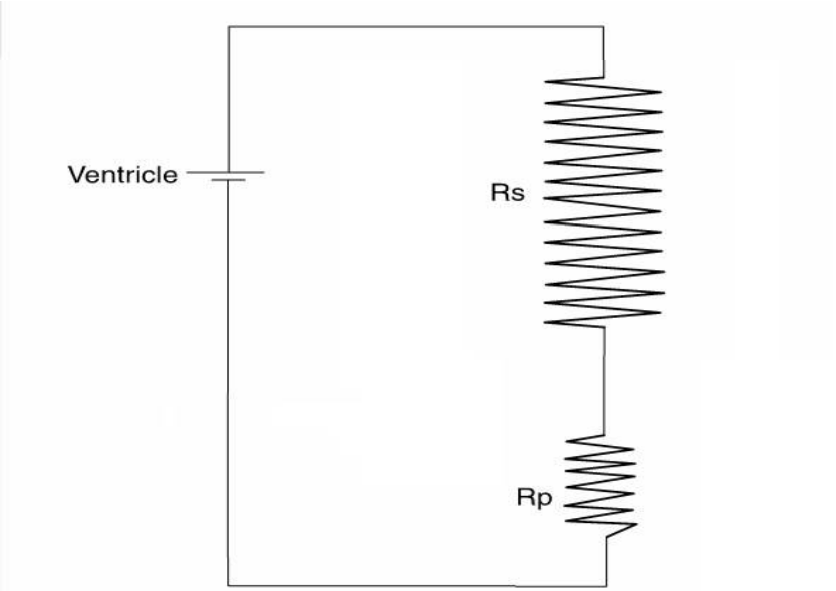
Dr. Francis Fontan

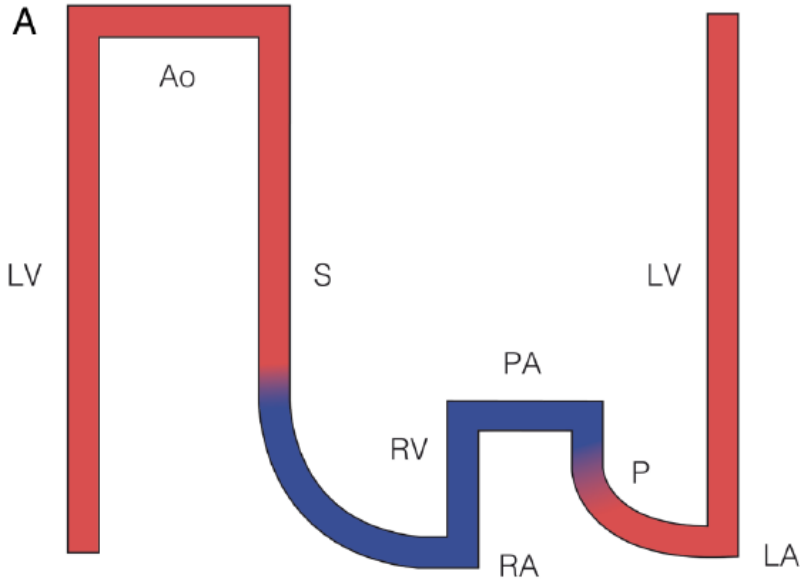
Fontan operation

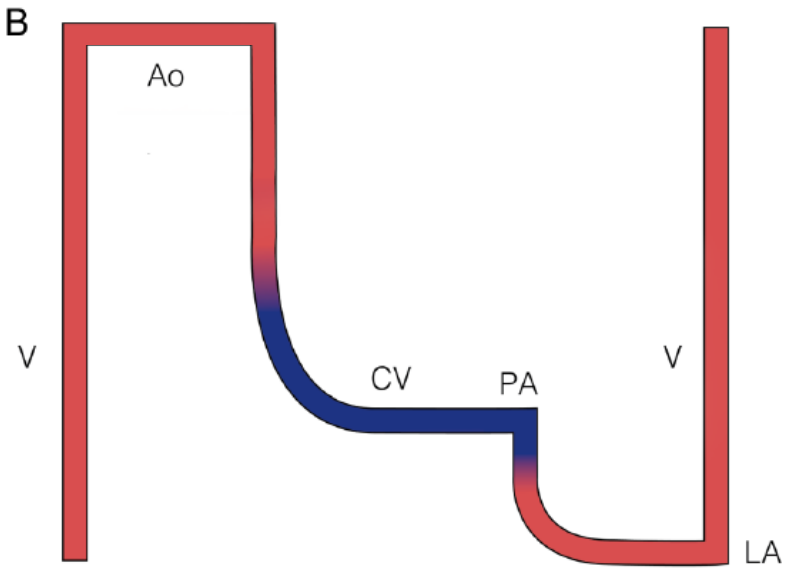
- Total cavopulmonary connection (TCPC)



Mavroudis C. Backer CL. 2013. Pediatric Cardiac Surgery, 4th ed





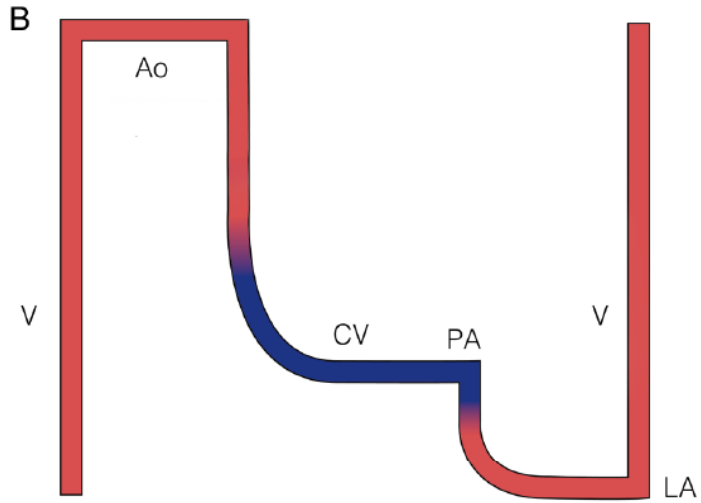


Normal circulation

Fontan circulation

Fontan palliation

- Disadvantages
  - Systemic venous congestion (hypertension)
  - Decreased cardiac output



Heart 2016;102:1081-1086

MULTI-STAGE PALLIATION

Fontan circulation

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

Fontan circulation

- Multi-stage palliation toward Fontan

- 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

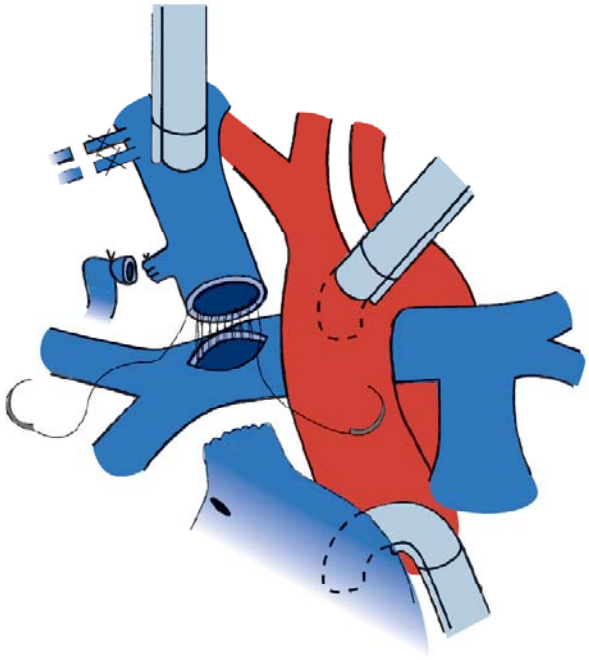
     - Coarctation of the aorta: arch repair
     - LVOT obstruction: DKS, etc.

  3. Unobstructed pulmonary venous return

     - Obstructed TAPVR: TAPVR repair

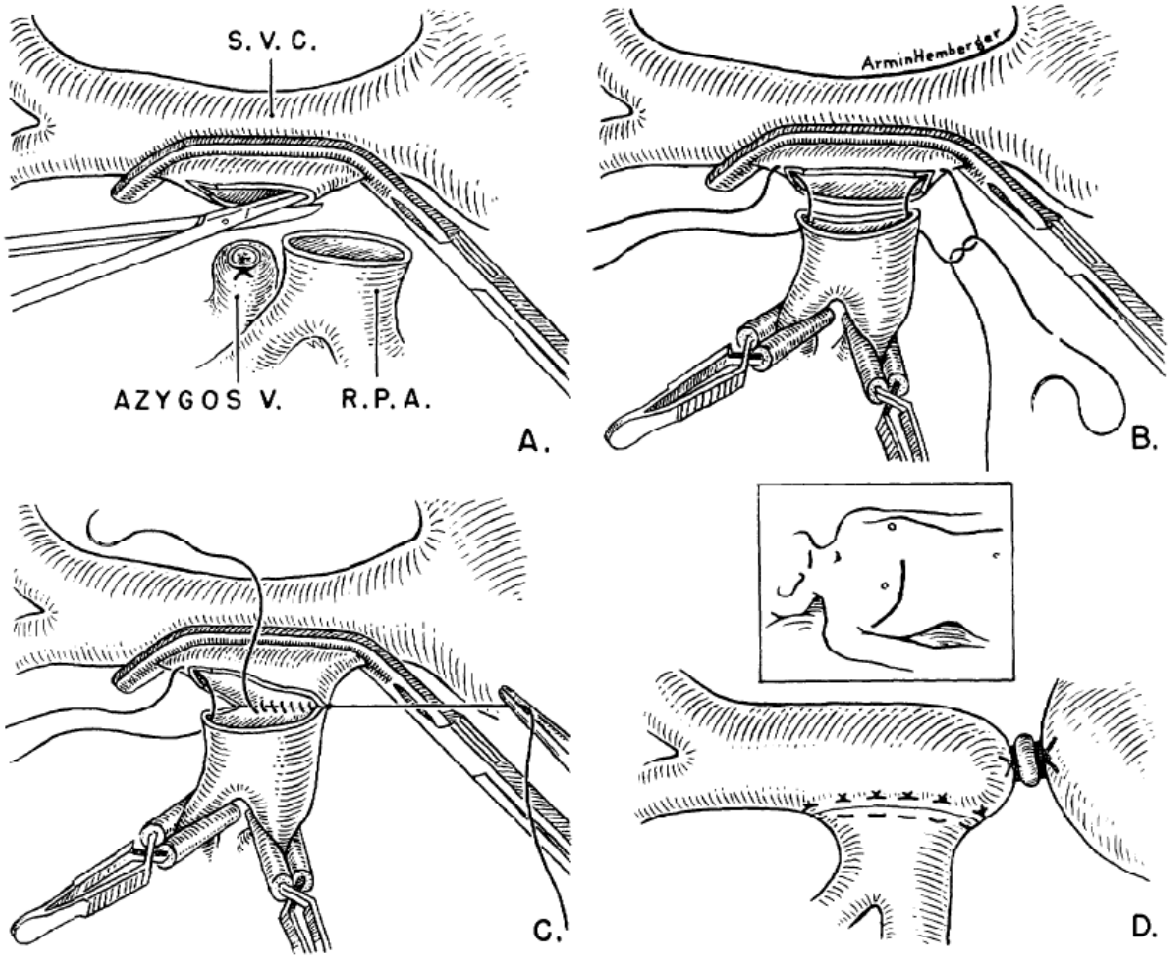
Multi-stage palliation

- Second-stage palliation
  - BCPS (Glenn)
  - 4-6 months
  - Improved Fontan outcomes



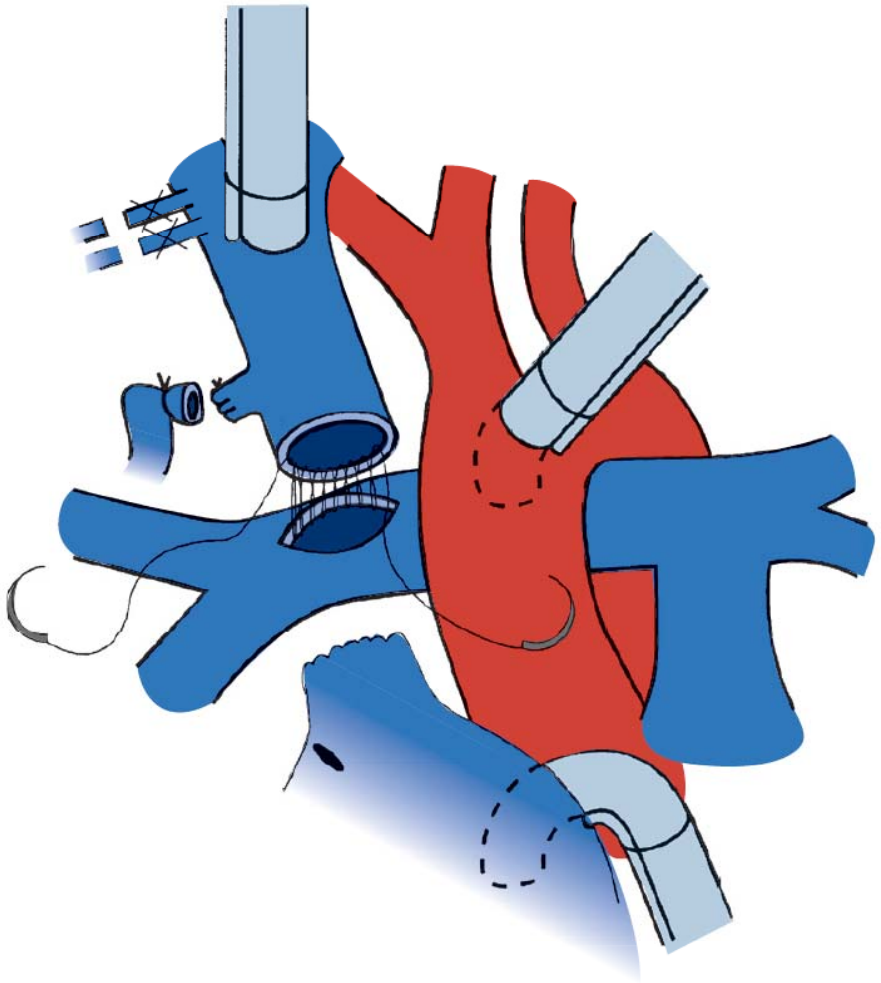
Sabiston and Spencer Surgery of the Chest, 9th ed, 2015

Glenn procedure



Circulation 1965;31:172-189

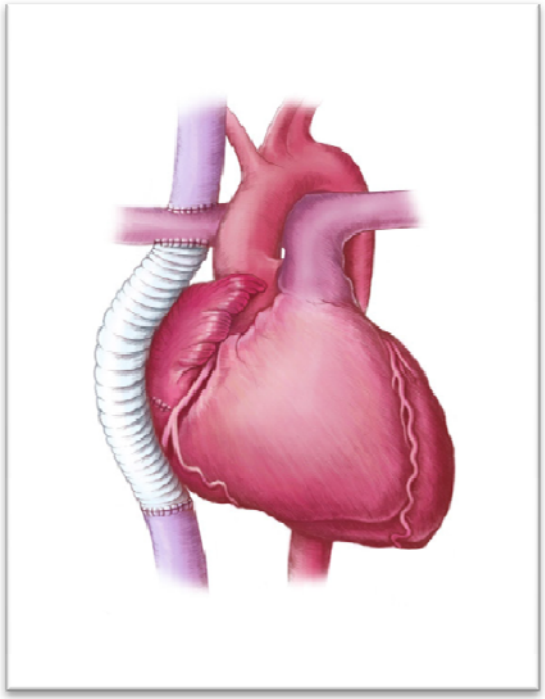
Bidirectional cavopulmonary shunt



Sabiston and Spencer Surgery of the Chest, 9th ed, 2015

Multi-stage palliation

- Last-stage palliation
  - Fontan operation
    - TCPC
  - Around 3 years old
  - Improved oxygen saturation



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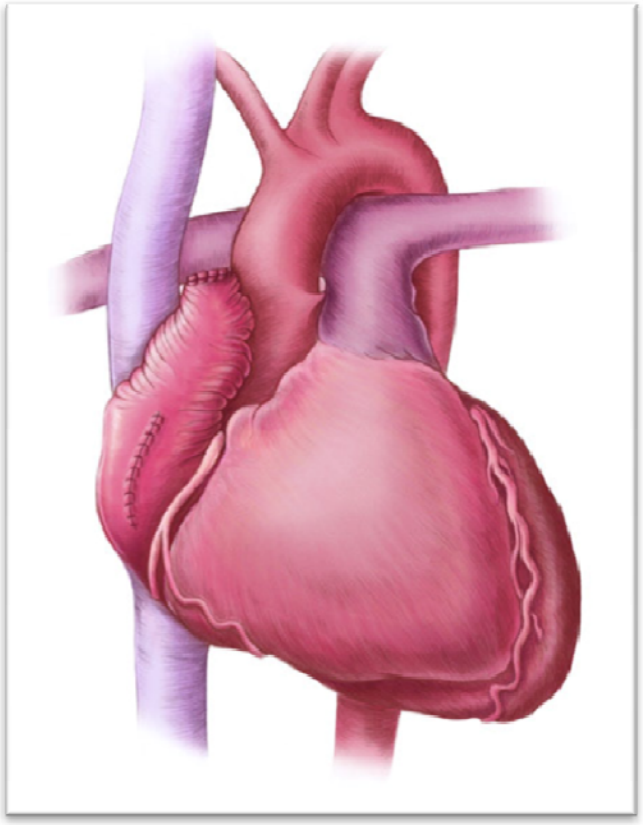
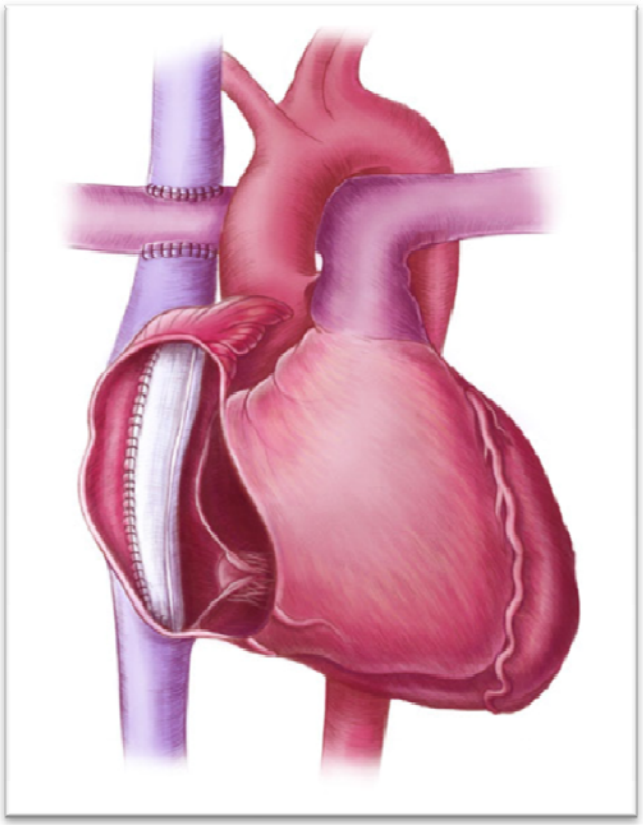
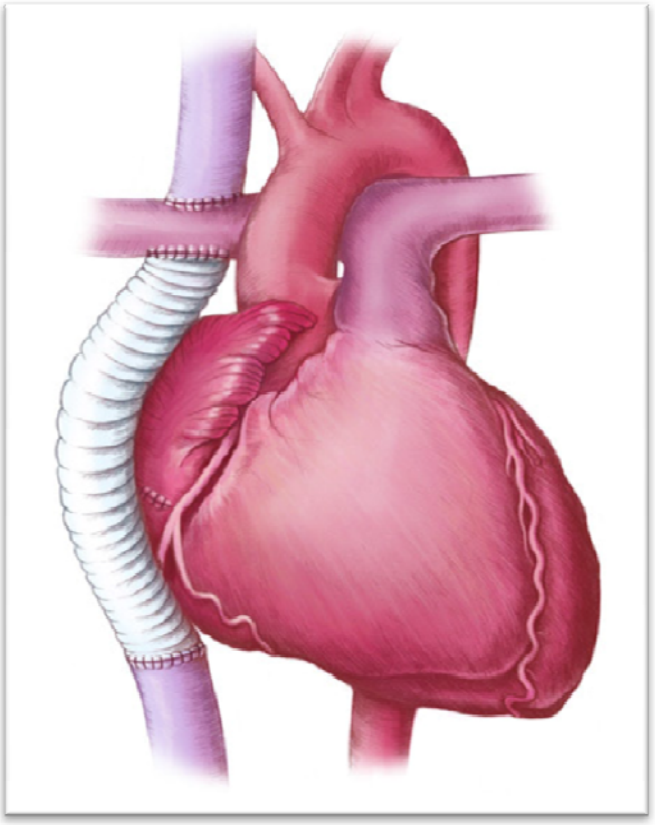
Fontan operation



Atrio-pulmonary (AP)



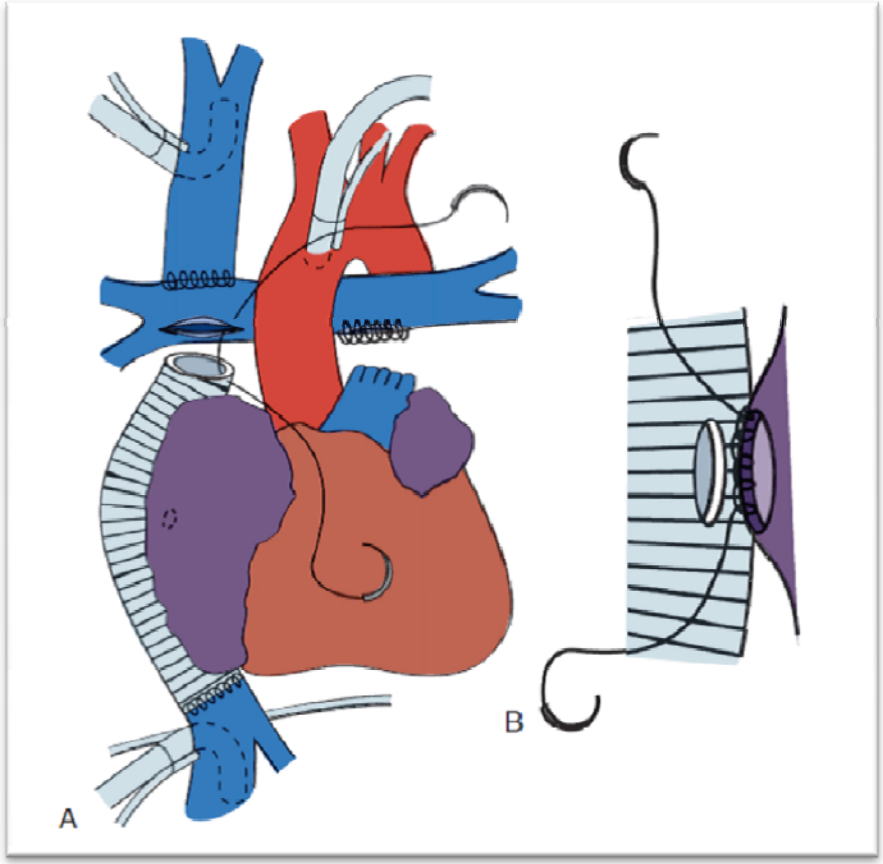
Lateral tunnel (LT)



Extracardiac conduit (ECC)

Fontan operation

- Early outcomes
  - Early mortality: < 5%
  - Early morbidity
    - pleural and pericardial effusion
    - fenestration



Sabiston and Spencer Surgery of the Chest, 9th ed, 2015

Fontan operation

- Late outcomes
  - Survival: 85% at 15 years
  - Late morbidity
    - Atrial arrhythmia
    - Venovenous collaterals
    - Thromboembolism: aspirin vs. warfarin
    - Protein-losing enteropathy (PLE)
    - Plastic bronchitis

Fontan failure

Sabiston and Spencer Surgery of the Chest, 9th ed, 2015

Fontan operation

- Risk stratification
  - Ventricular function
    - Atrioventricular valve regurgitation (AVVR)
  - Pulmonary vasculature
    - Pulmonary vascular resistance (PVR) < 4 U/m^2
    - Mean pulmonary artery pressure (PAP) < 15 mmHg

Sabiston and Spencer Surgery of the Chest, 9th ed, 2015

Diagnosis

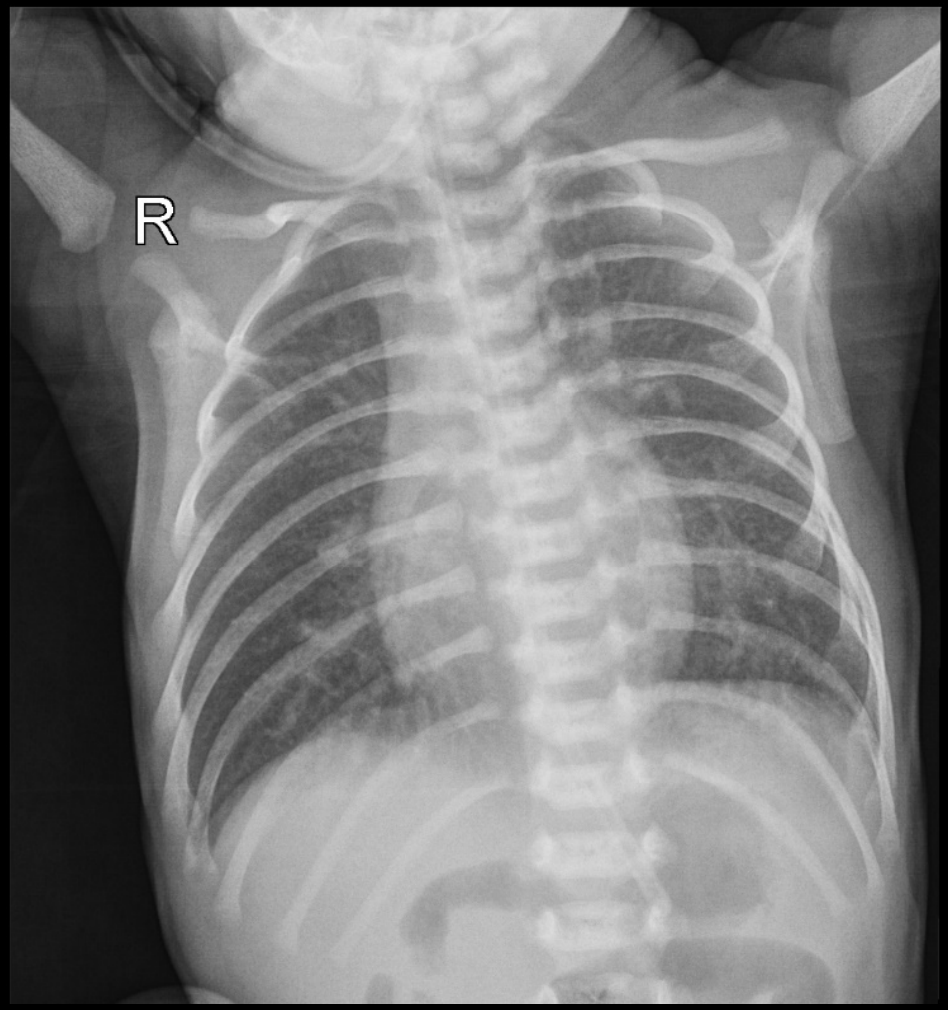
Tricuspid Atresia	Mitral Atresia
Double Inlet left ventricle	Double inlet right ventricle
PA with intact ventricular septum	Hypoplastic left heart syndrome
	Unbalanced AV canal
	Heterotaxia syndrome
	Others

CASE PRESENTATION

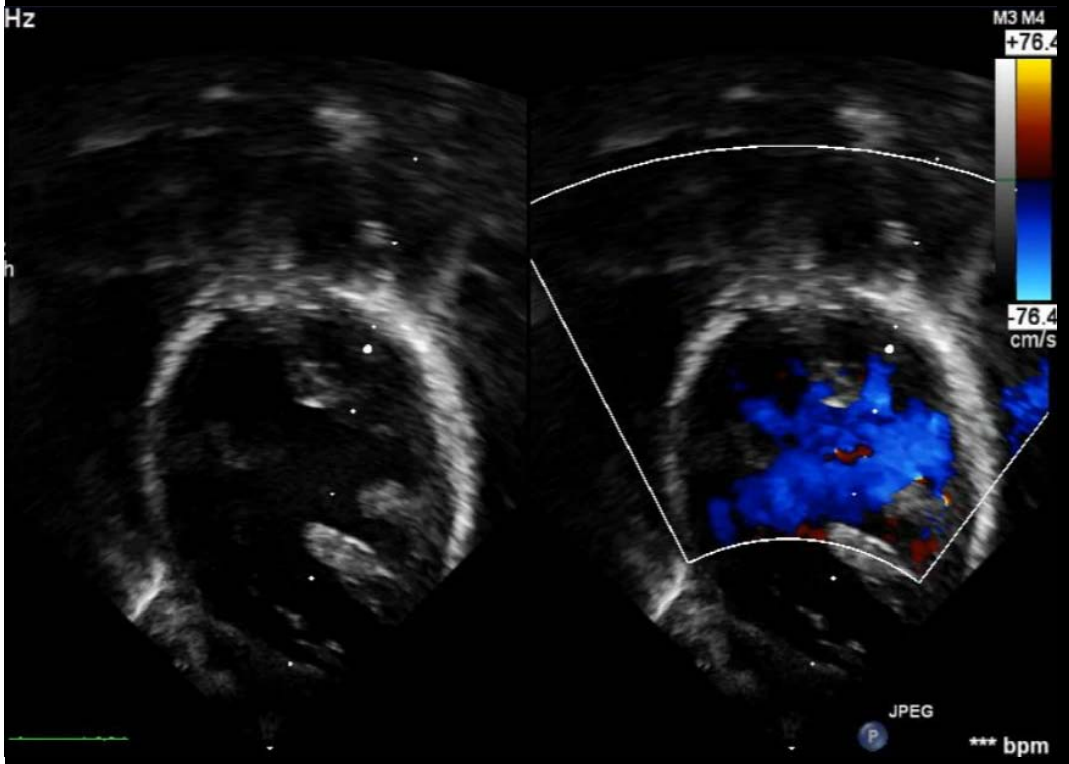
Case

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

After birth



After birth



After birth

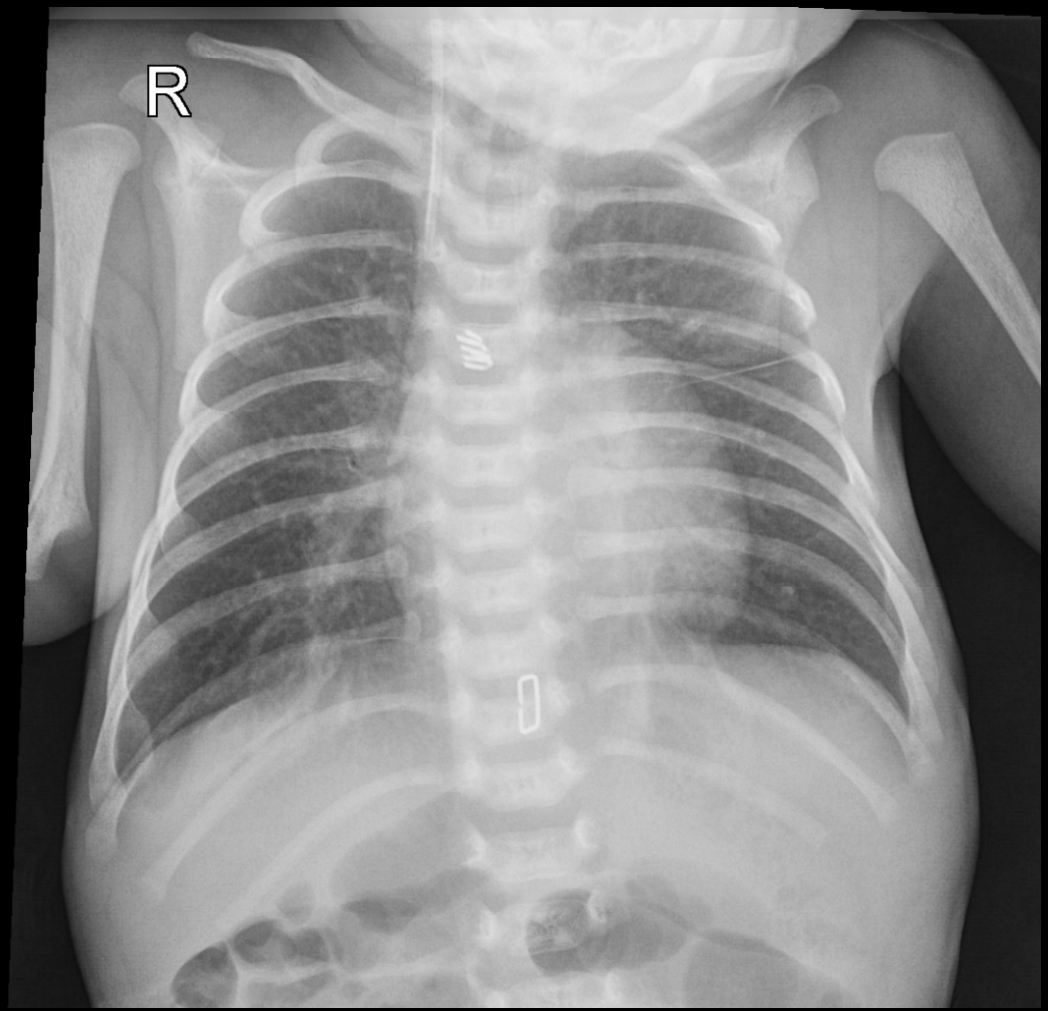
Case

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

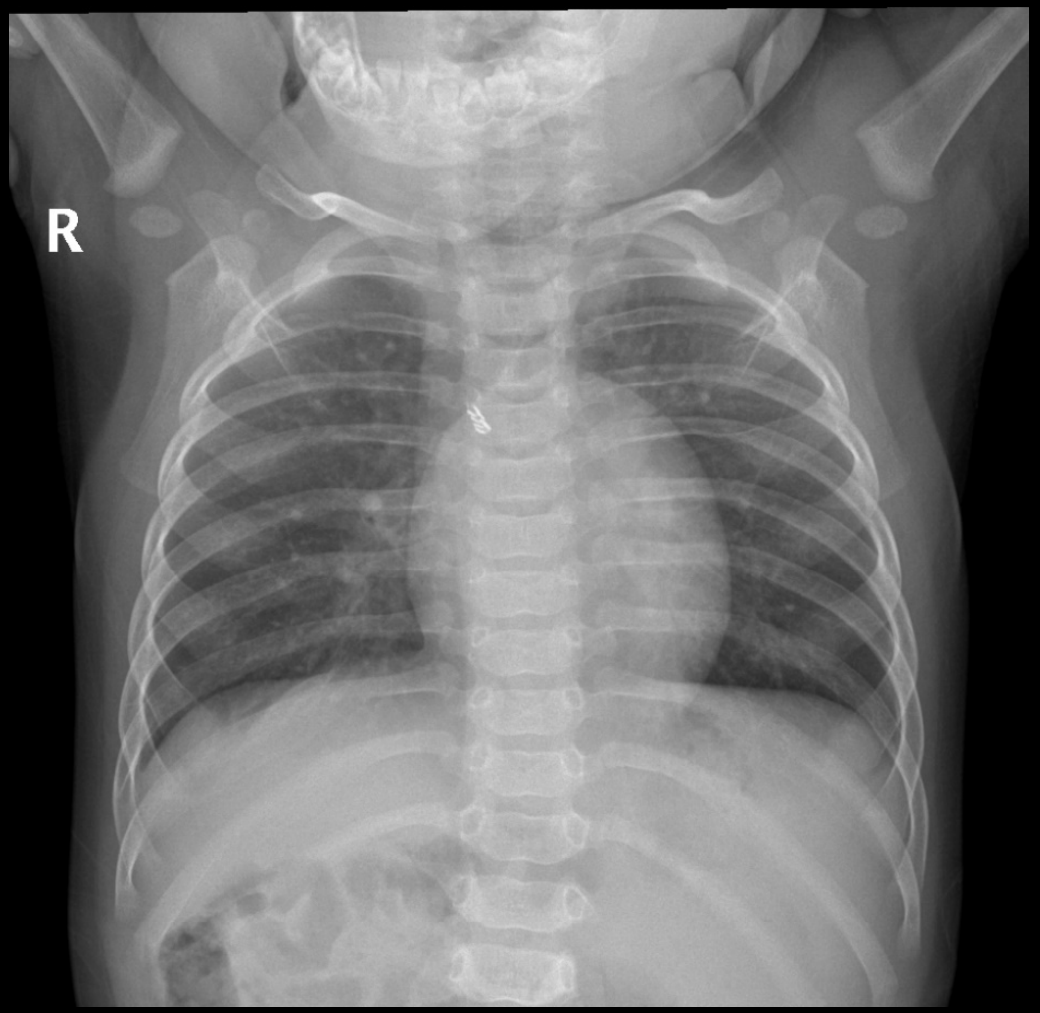
Case

- GA: 37+4 wks, 2.5kg, male
- Diagnosis
  - FSV: Heterotaxy syndrome (RAI), DORV, AVSD
- PAB at 10 days old

Post PAB



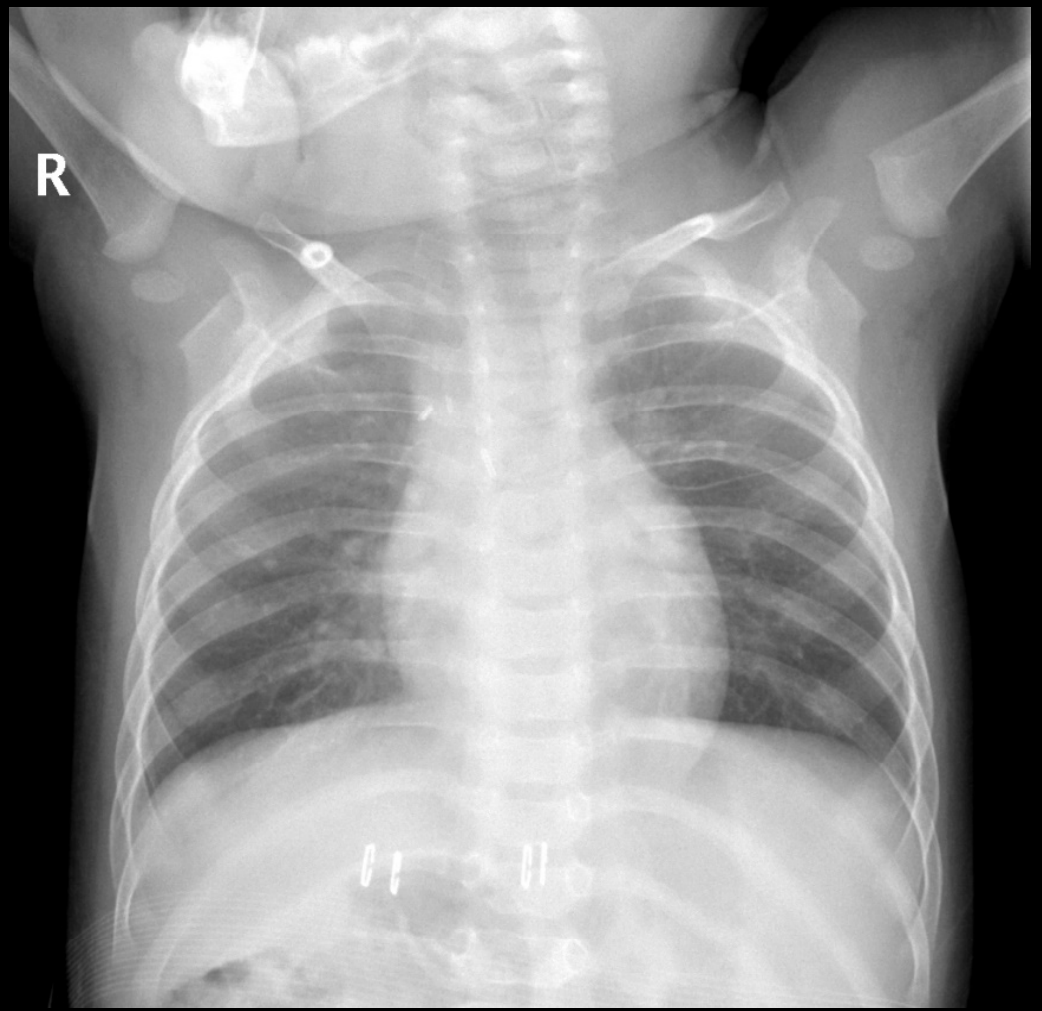
Pre BCPS



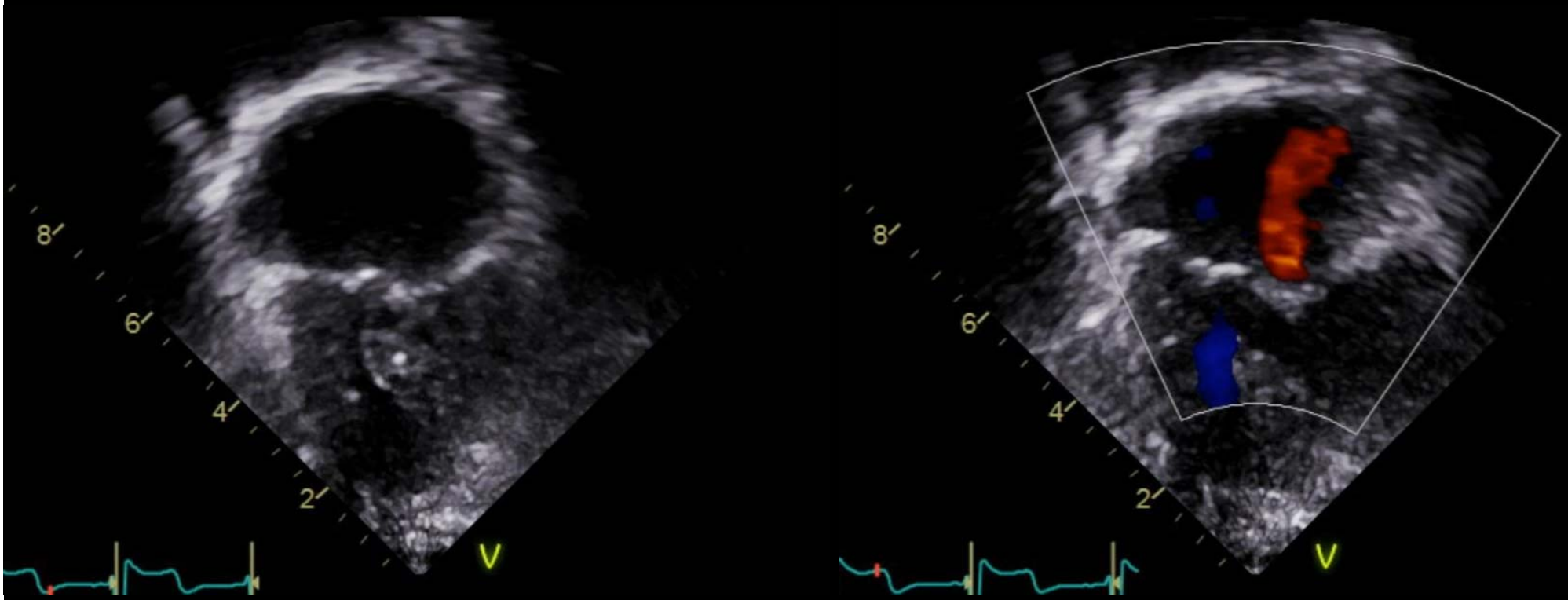
Case

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

Post BCPS



Pre Fontan



Pre Fontan

Pre Fontan

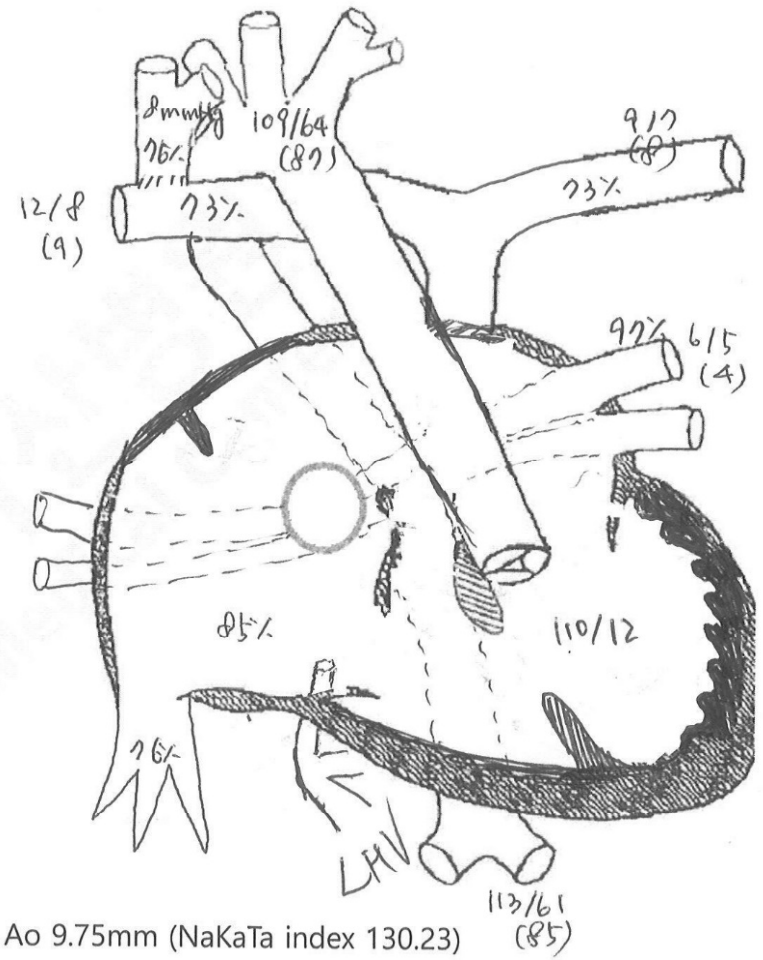
Precath Diagnosis

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)

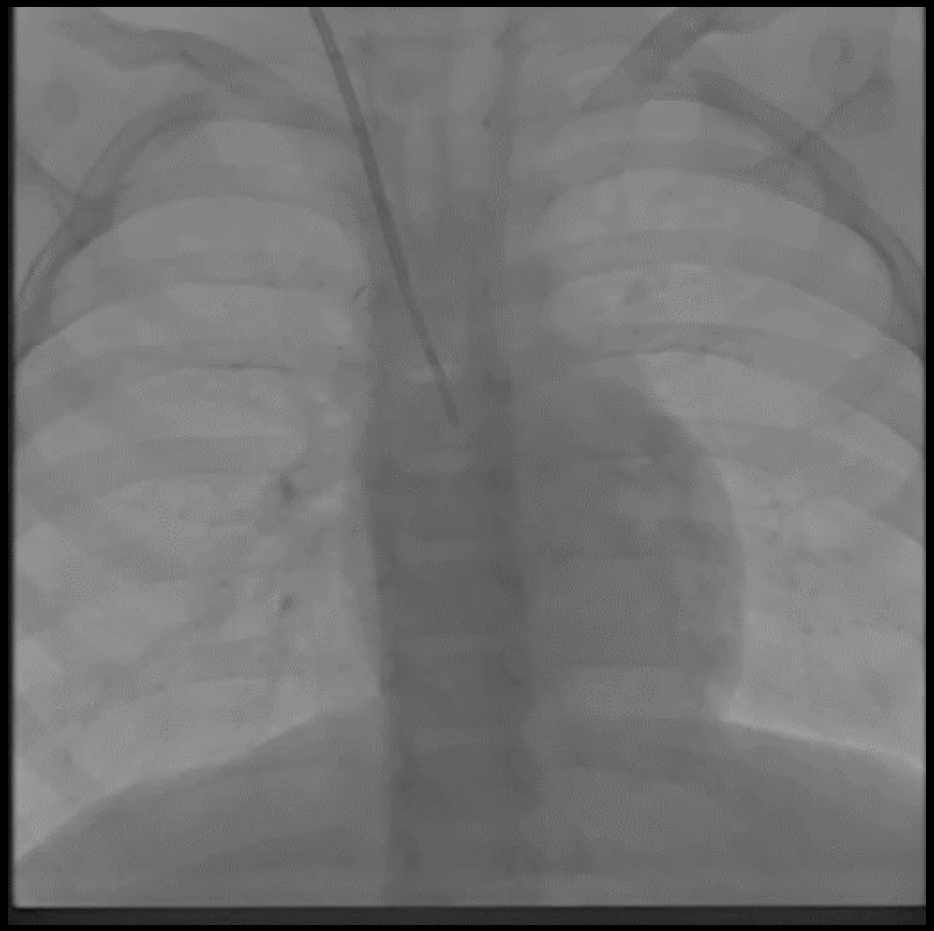
* Cath finding >

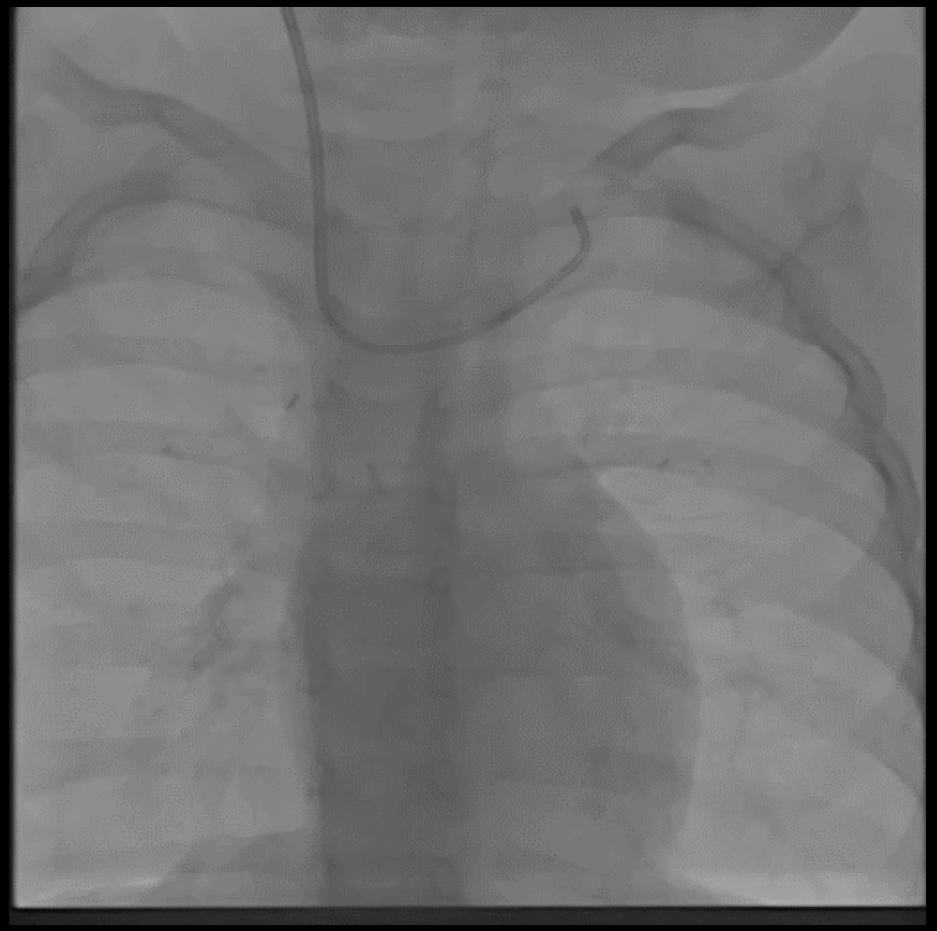
1. Finding

- 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

Pre Fontan





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)

Thank you for your attention