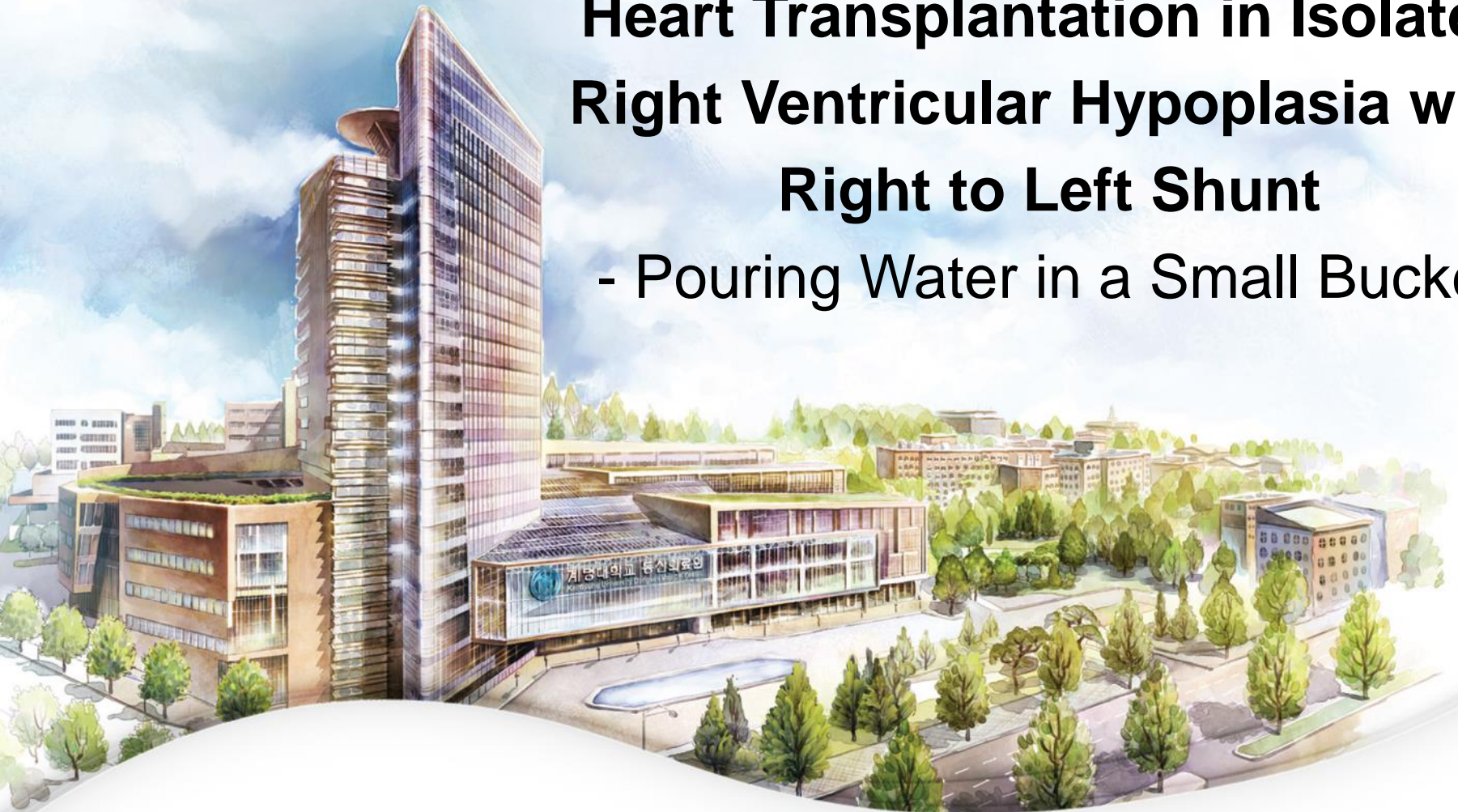


Heart Transplantation in Isolated Right Ventricular Hypoplasia with Right to Left Shunt - Pouring Water in a Small Bucket



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November 2018

35 years old / Male

Smoking (-), Alcohol (-)

C/C : DOE

Isolated RV hypoplasia (s/p BCPS 2003.3)

Varicose vein

Brain abscess s/p op (2017.10)

Epilepsy

Vital sign & Physical Examination

- Vital sign : 90/60 mmHg – 80/min - 22/min – 36.8 °C



Pitting edema (+/+)
Peripheral cyanosis
Varicose vein

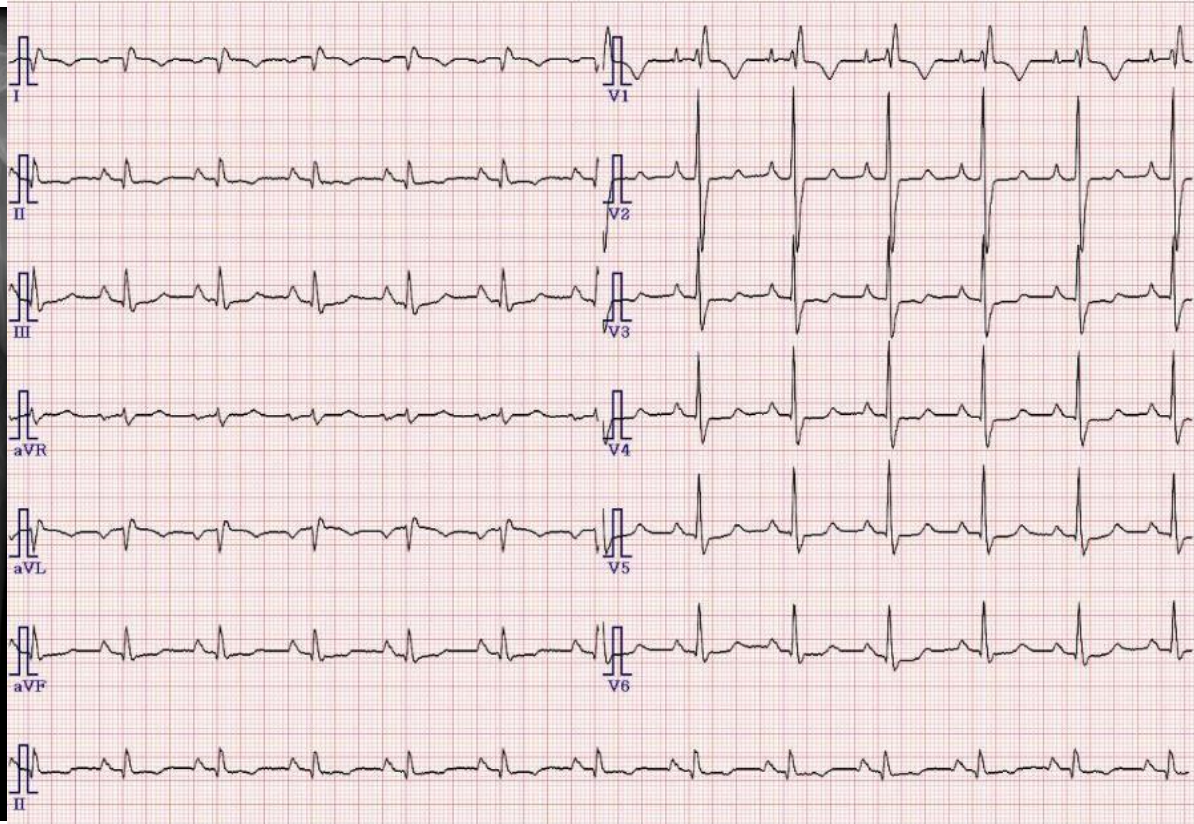


Clubbing (+)



Clubbing (+)
Schamroth sign (+)

Chest PA and ECG



Laboratory findings

pH 7.486 pCO₂ 35.5 **pO₂ 39.5** HCO₃⁻ 26.5 **O₂sat 69.3**

CBC 2,700 - 15.2 / 44.9 - 105K

BUN/Cr 19/0.68

Na/K/Cl 132/4.3/98

TP/Alb 5.2/3.3

AST/ALT 29/23

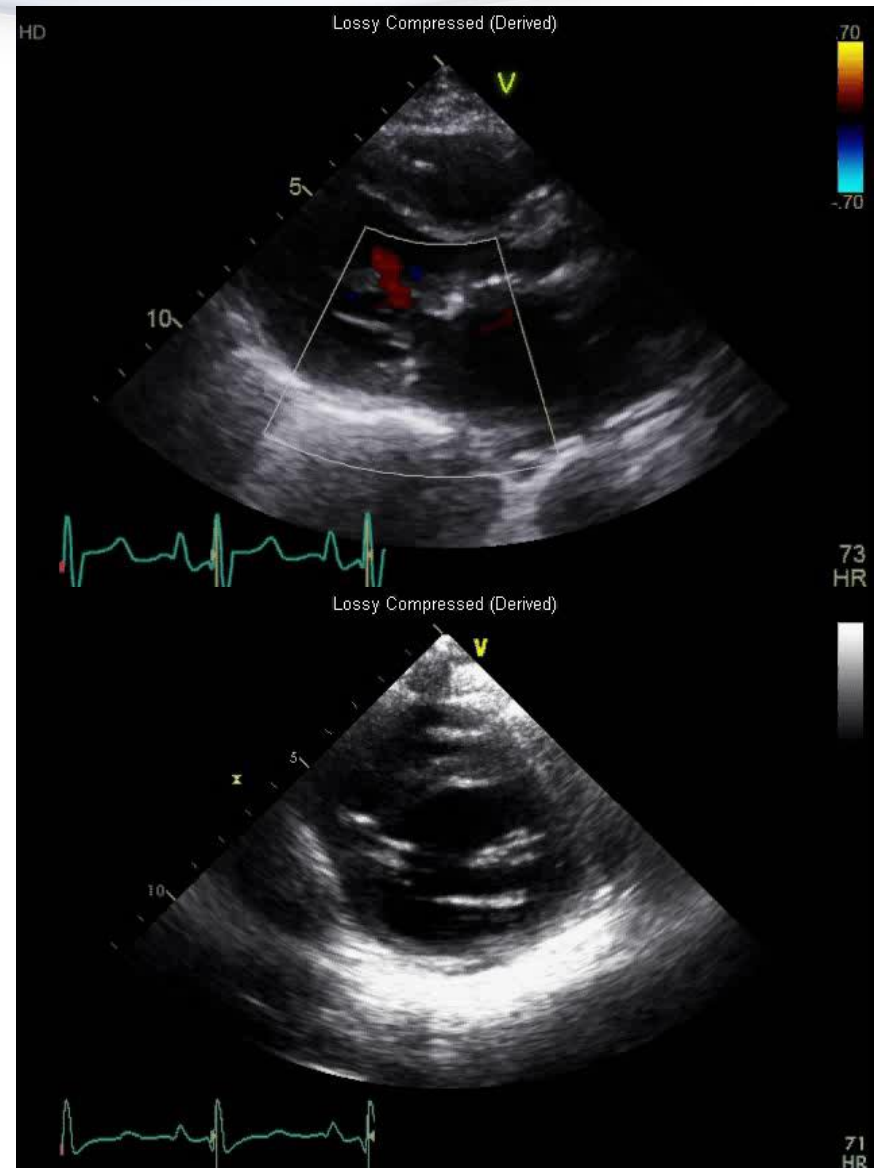
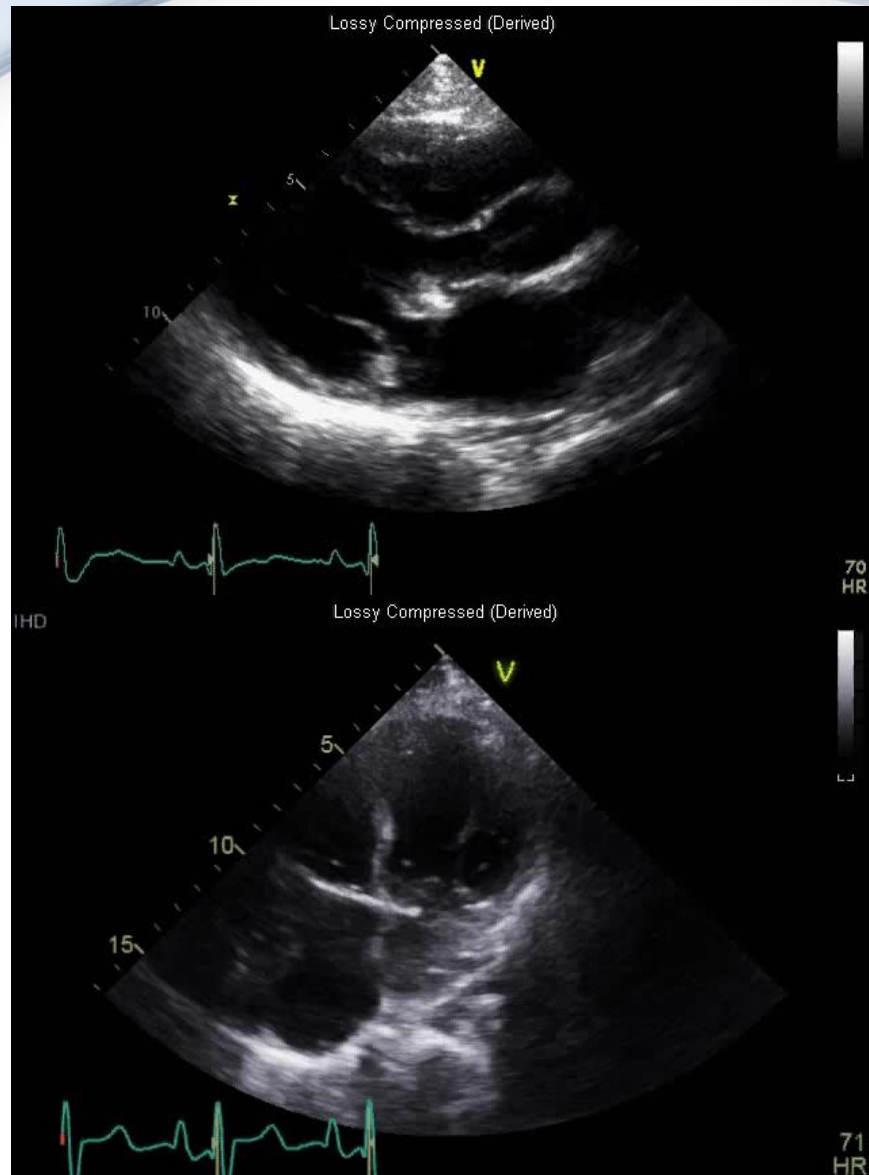
NT-proBNP **2,227**

TB/DB 0.71/0.26

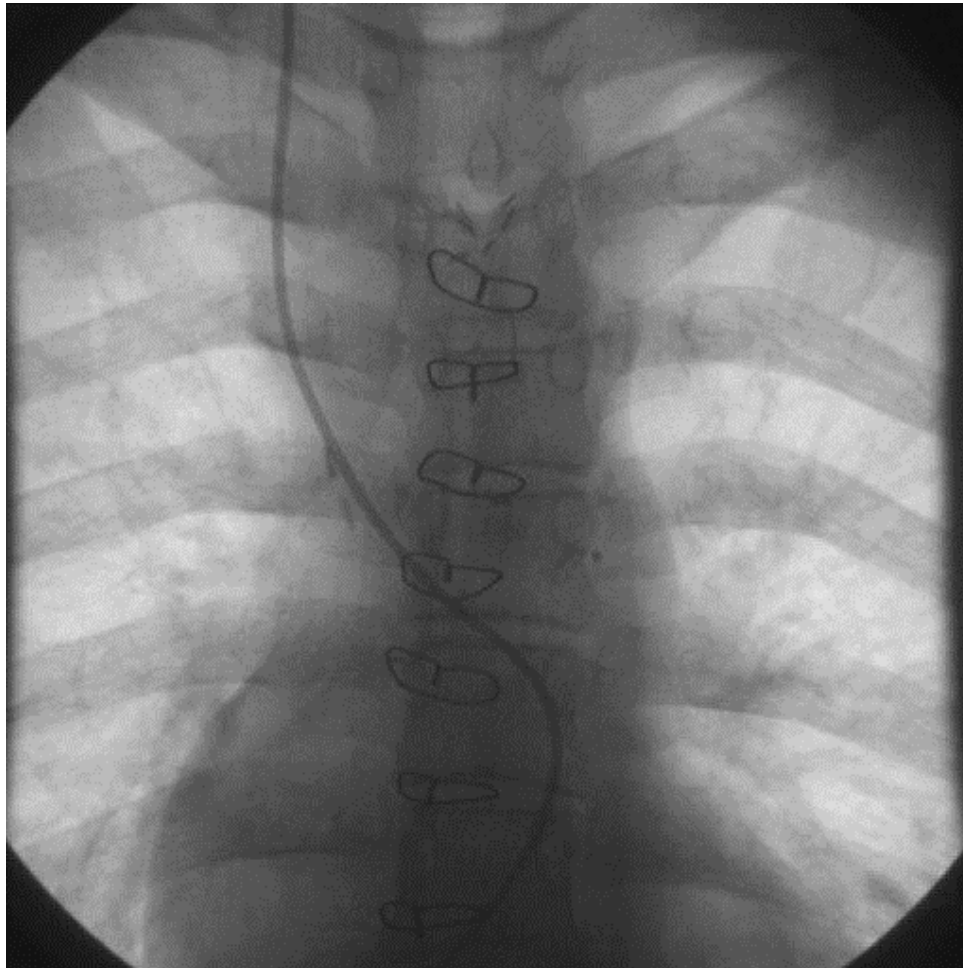
CK-MB/TnI 1.7/0.168

PT(sec/INR) 22.2/1.1

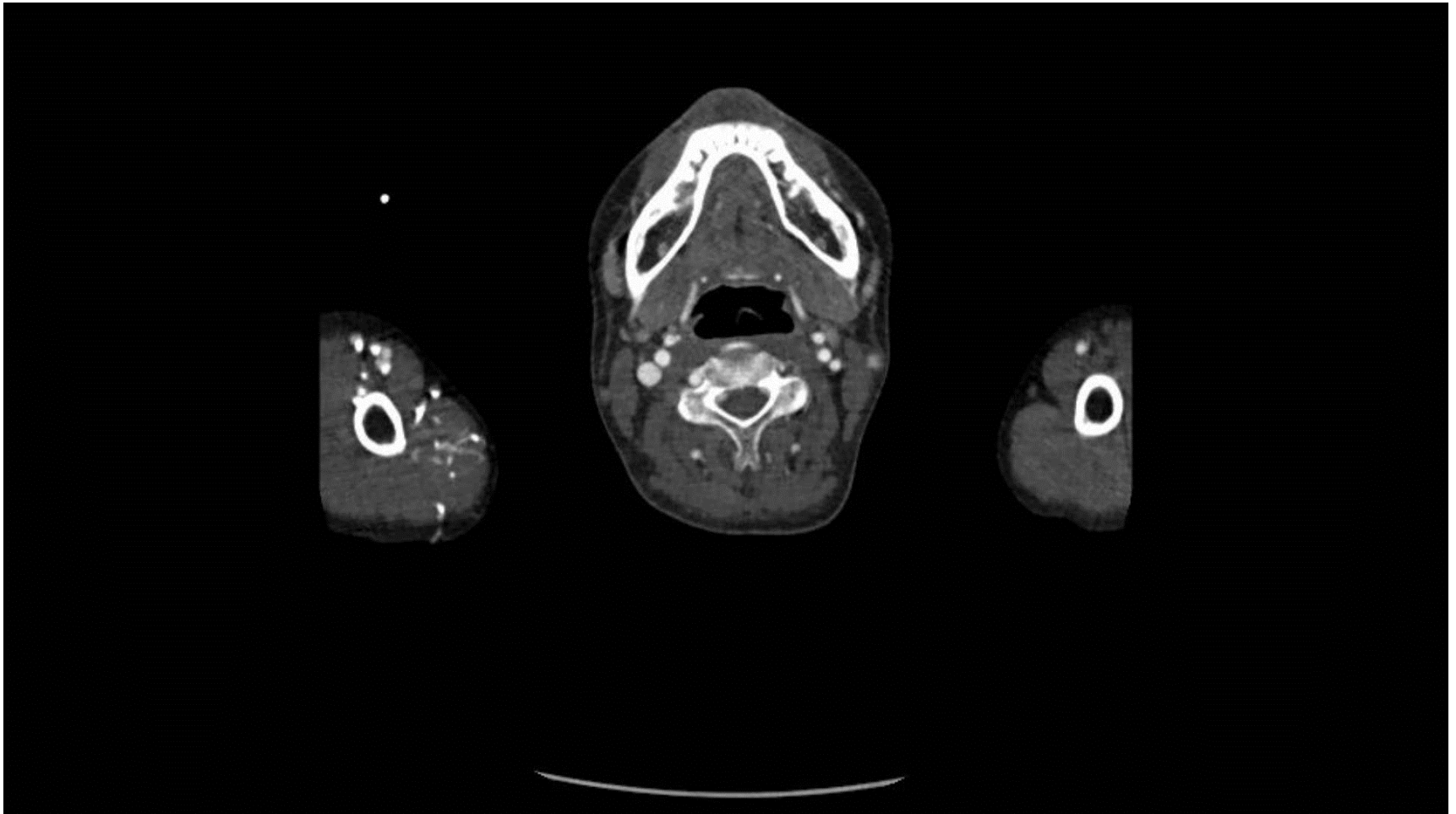
TTE



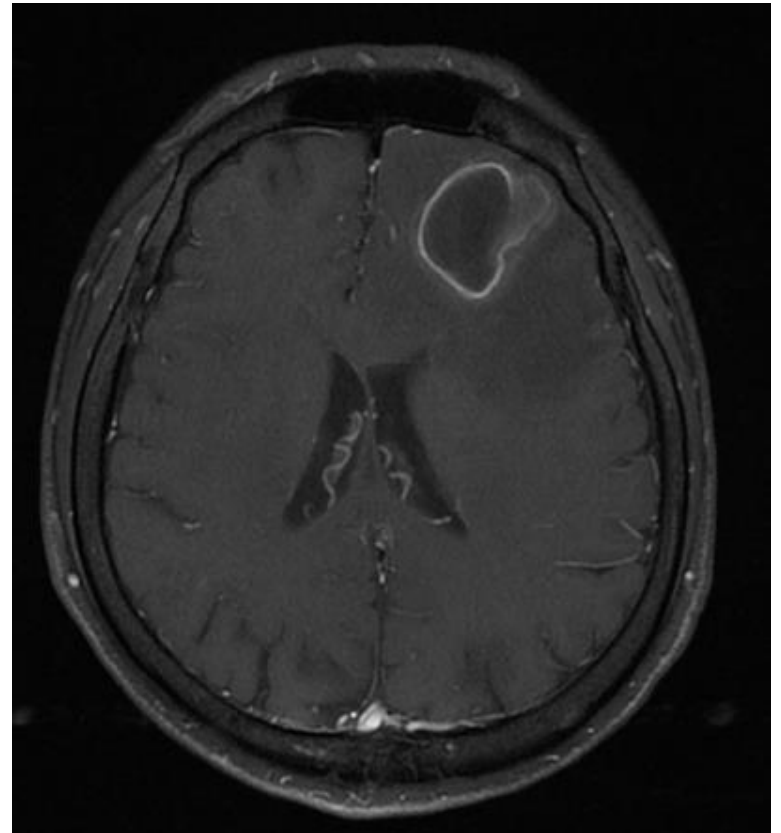
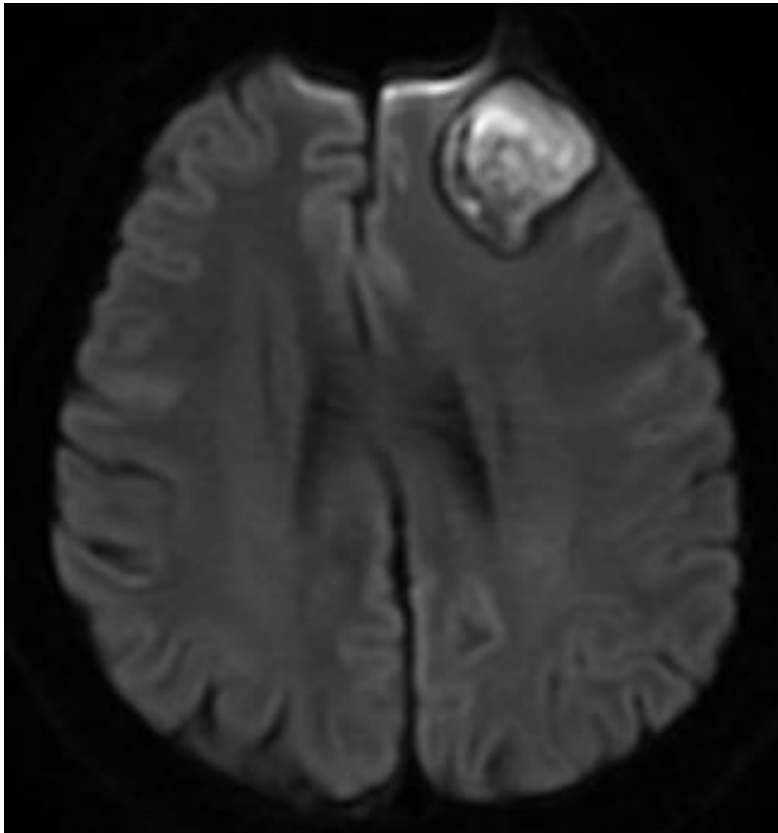
Post-OP Angiography (2003.8)



CT f/u (several years later)



Brain Abscess s/p OP (2017.10)

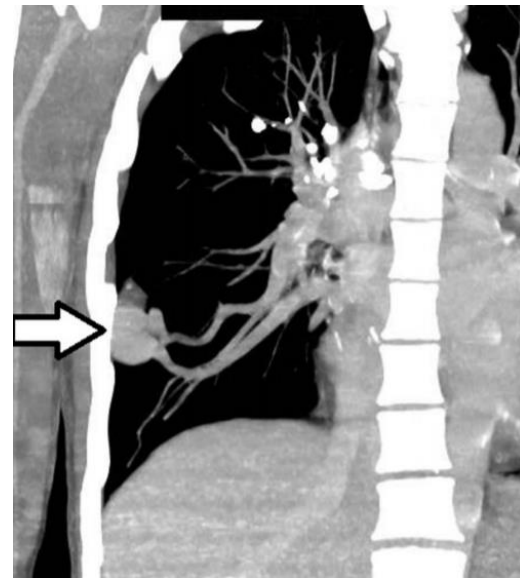


Brain Abscesses Associated with R→L Shunts in Adults

- Avoidance of pulmonary filtering which effectively filters most microorganisms and septic emboli may increase the risk of brain abscess

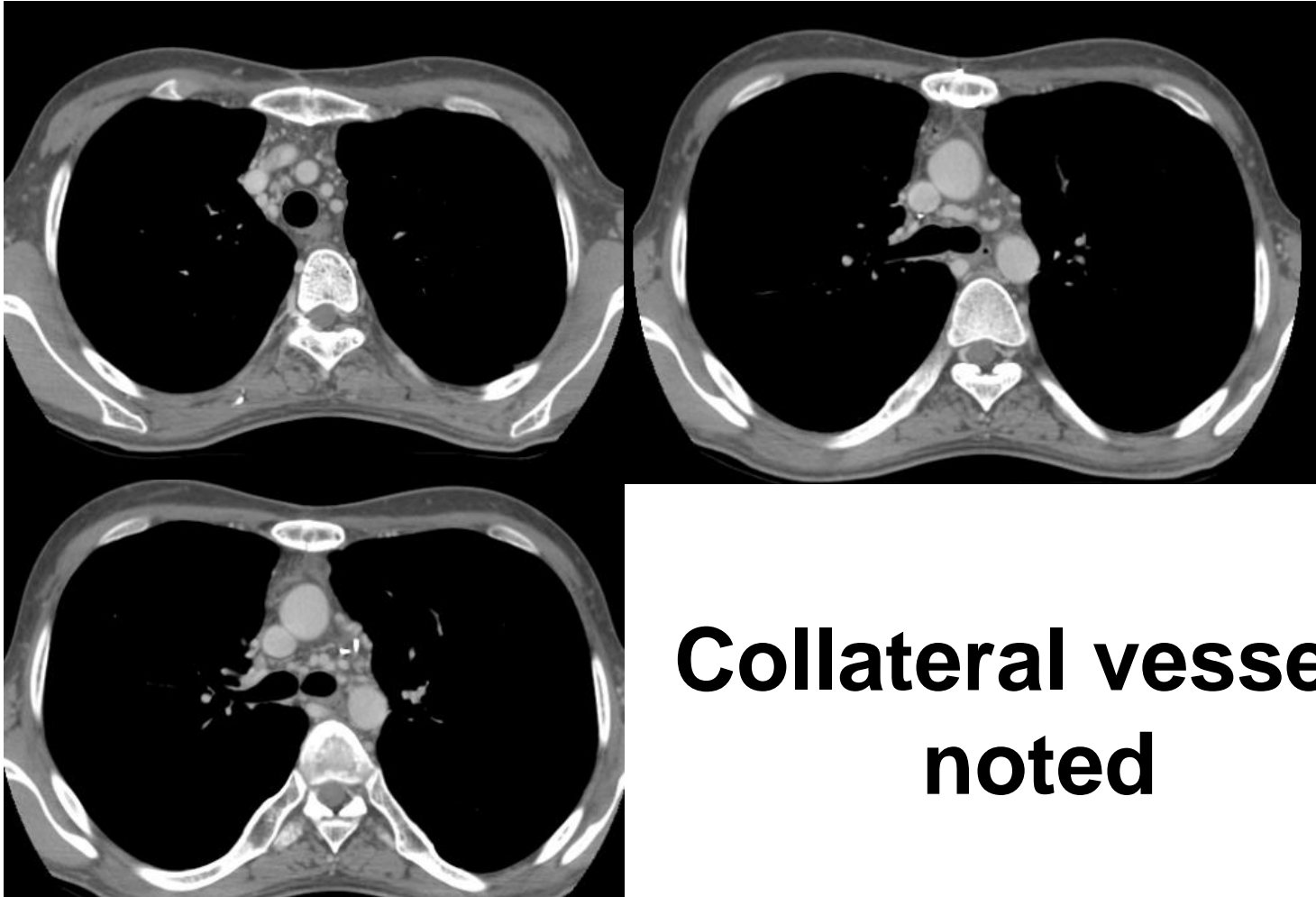


Persistent left SVC



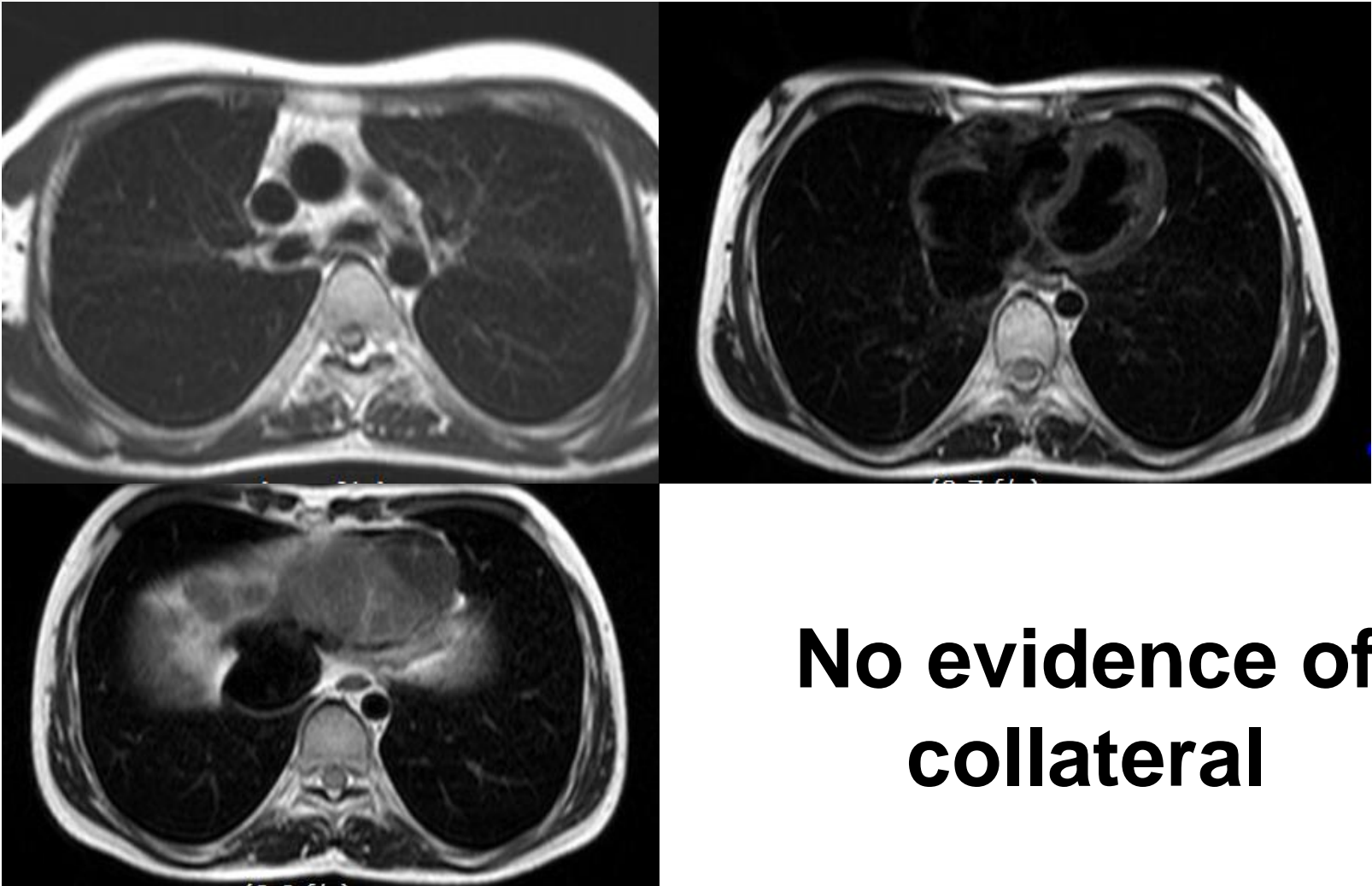
Pulmonary AVM

Chest CT after BCPS (2006)



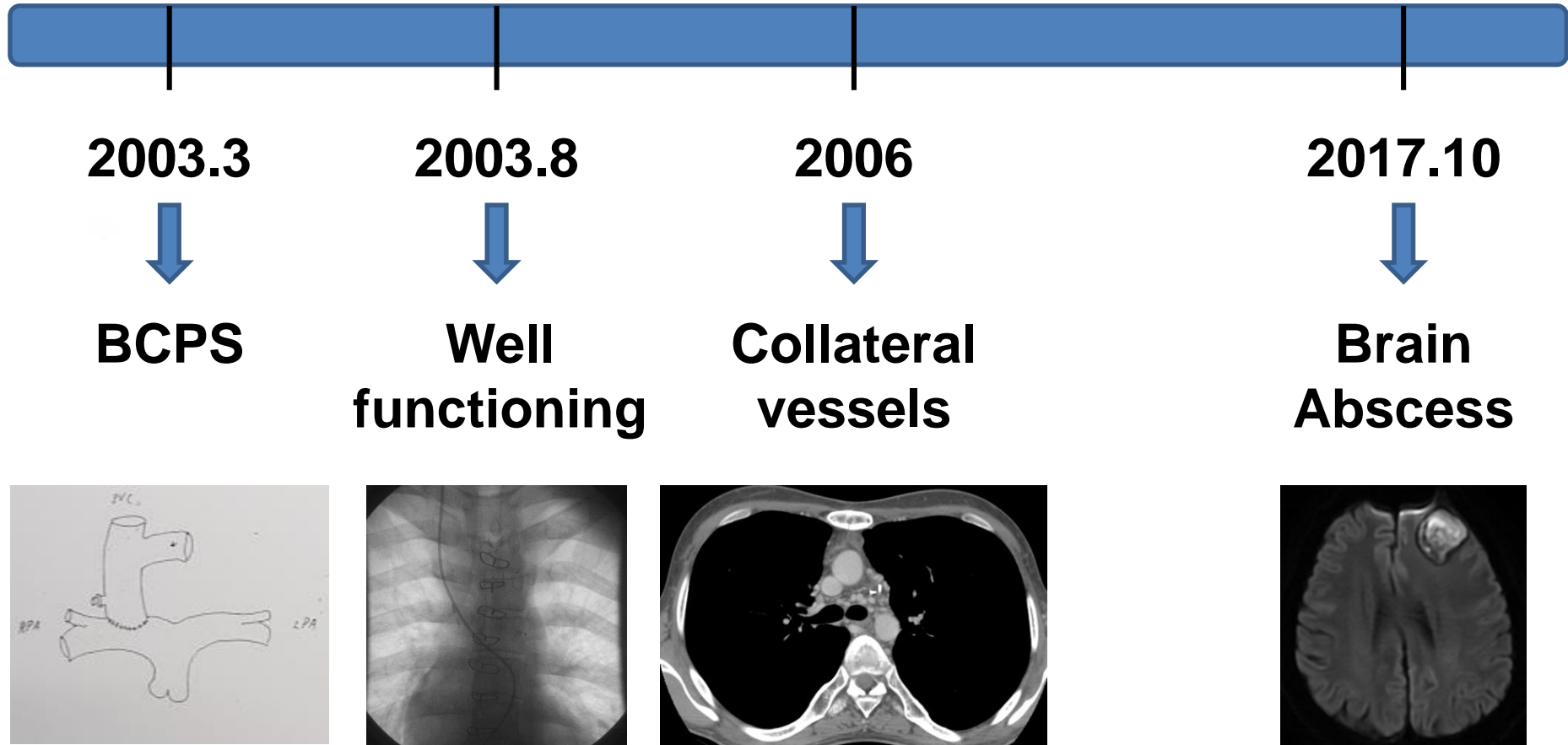
**Collateral vessels
noted**

Before BCPS (2003.3)



**No evidence of
collateral**

Timeline for the Consequence of BCPS



Cardiac MRI (Flow dynamics)



Subclavian vein



Collateral vessel



LA



LV



Systemic artery



IVC



PA & SVC

Hemodynamic information from the MRI

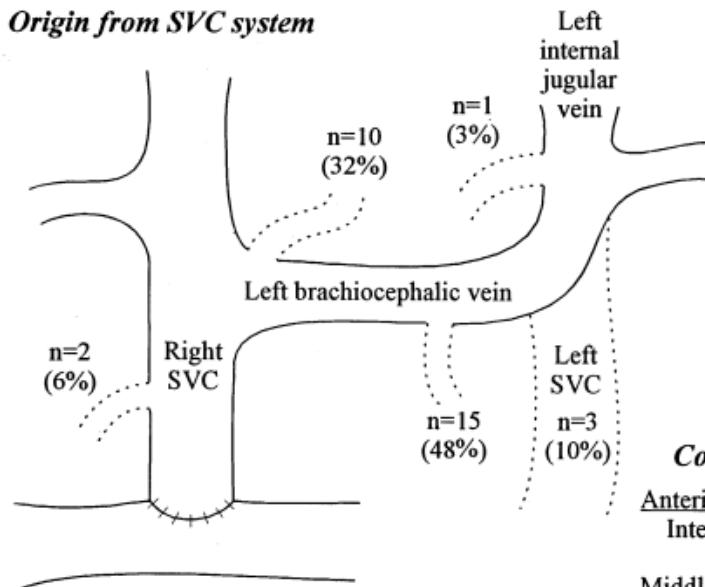
RV Evaluation		LV Evaluation	
RVEDVI (ml/m ²)	26.92	LVEDVI (ml/m ²)	72.64
RVESVI (ml/m ²)	11.74	LVESVI (ml/m ²)	42.5
RVEF (%)	56.4	LVEF (%)	41.4
MPA flow (ml)	27.37	PR (%)	1.93
LPA (ml)	9.8	BCPS forward (ml)	4.09
RPA (ml)	13	backward (ml)	2.95

Systemic venous collateral after BCPS

- 1/3 patients after BCPS procedures have been diagnosed with collateral channels, more than half of them draining below the diaphragm
- Collateral vessels may develop in the absence of mechanical obstruction
- Post op. gradient between the SVC and the RA was independently associated with the presence of collaterals
 - Relative **reduction in caval or PA cross-sectional area after ligation** of one SVC (in the setting of bilateral cava)
 - **PA hypoplasia** or distortion, or obstruction of the anastomosis

Collaterals after BCPS

Origin from SVC system



Course from SVC to IVC

Anterior

Internal thoracic: n=8 (26%)

Middle

Intercostal/pericardial: n=9 (29%)

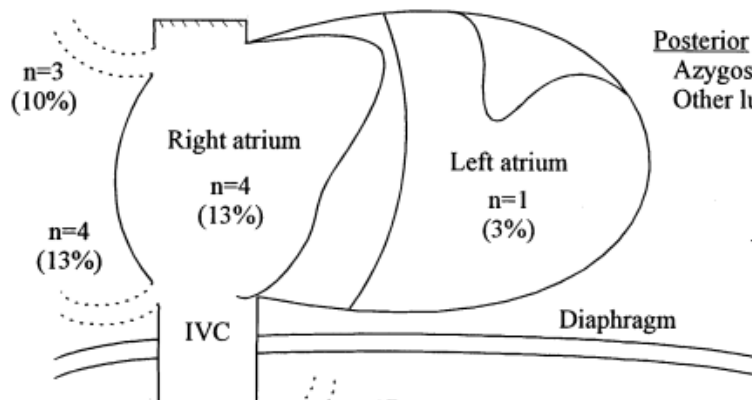
Left SVC: n=3 (10%)

Posterior

Azygos/hemiazygos: n=8 (26%)

Other lumbar: n=3 (10%)

Entry into IVC system

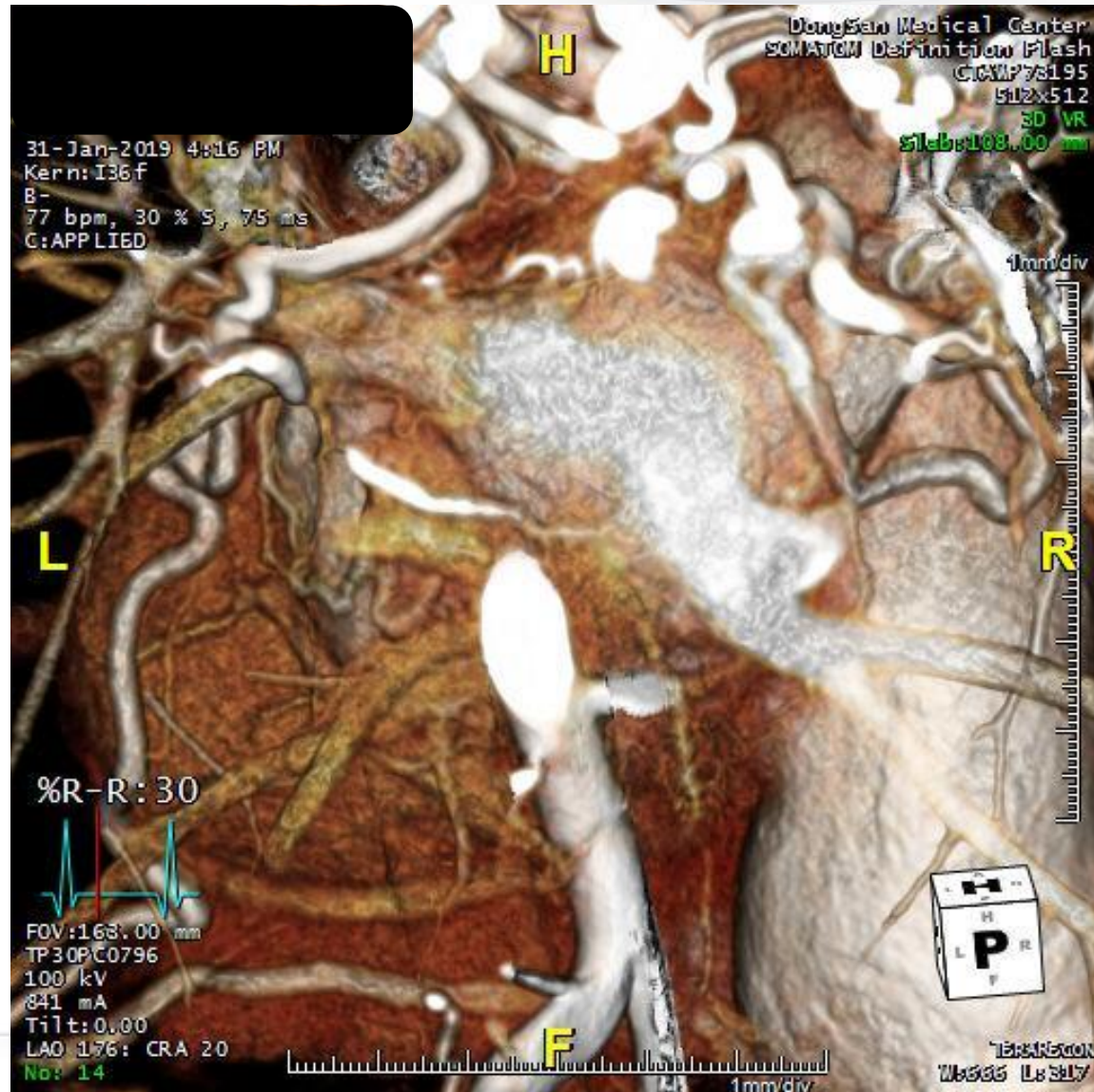


What is the Cause of Collateral Flow?

- **BCPS (SVC to RPA)**
 - **Increased Flow to PA with limited capacity**
(d/t PA hypogenesis)
 - **Increased pressure of PA**
 - **Failure of BCPS**
(Flow congestion to induce co



Cardiac CT with vascular reconstruction



Decided to Proceed Heart Transplantation with Collateral Ligation

Heart Transplantation with Collateral ligation

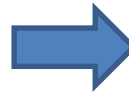


Total pump time : 264 min

Aorta cross clamp time : 100 min

Improved peripheral cyanosis

Pre-OP



POD #14



**Thank You
For Your Attention !**