Obstacles droits : de la vie foetale à l'âge adulte

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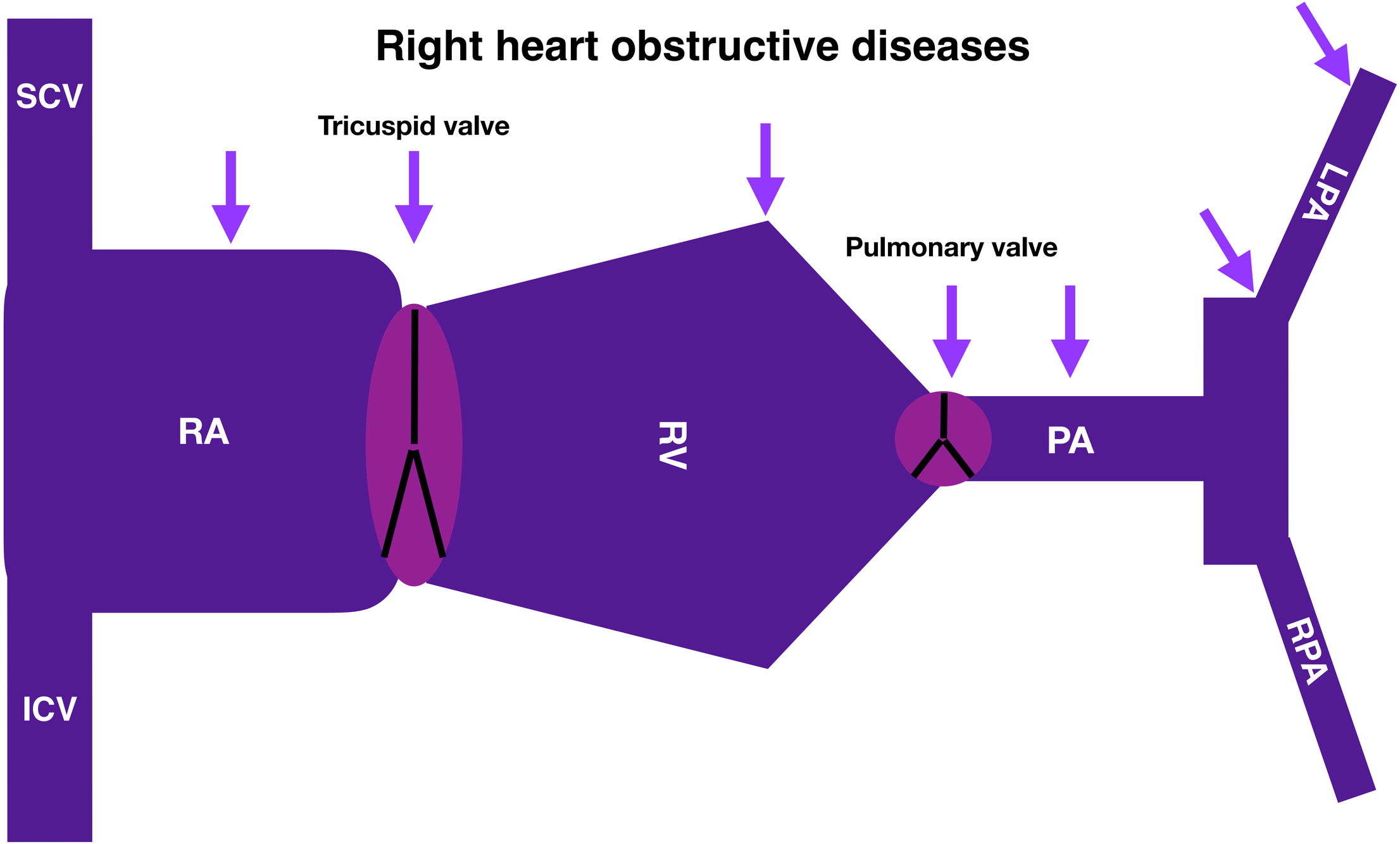
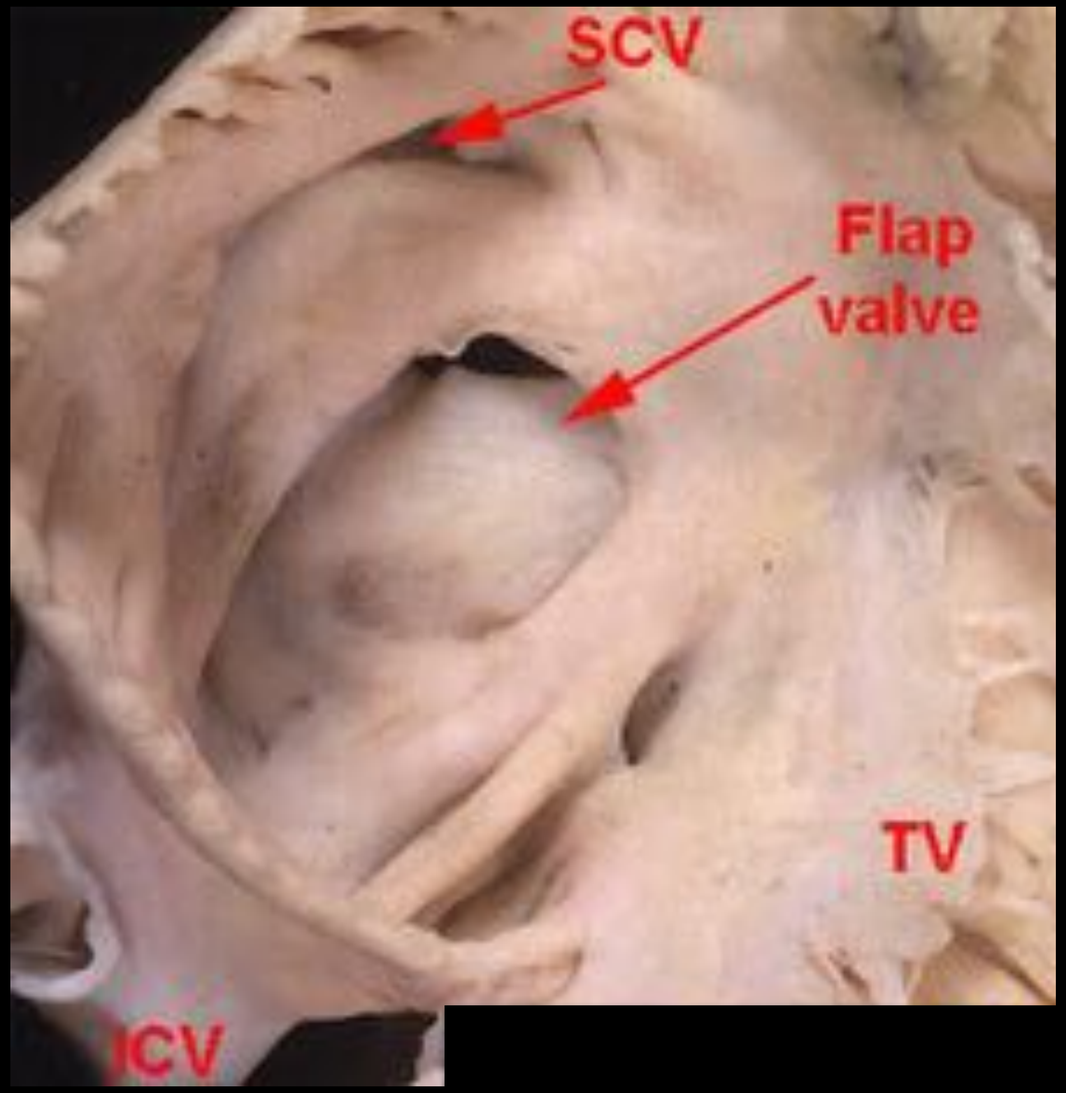
Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

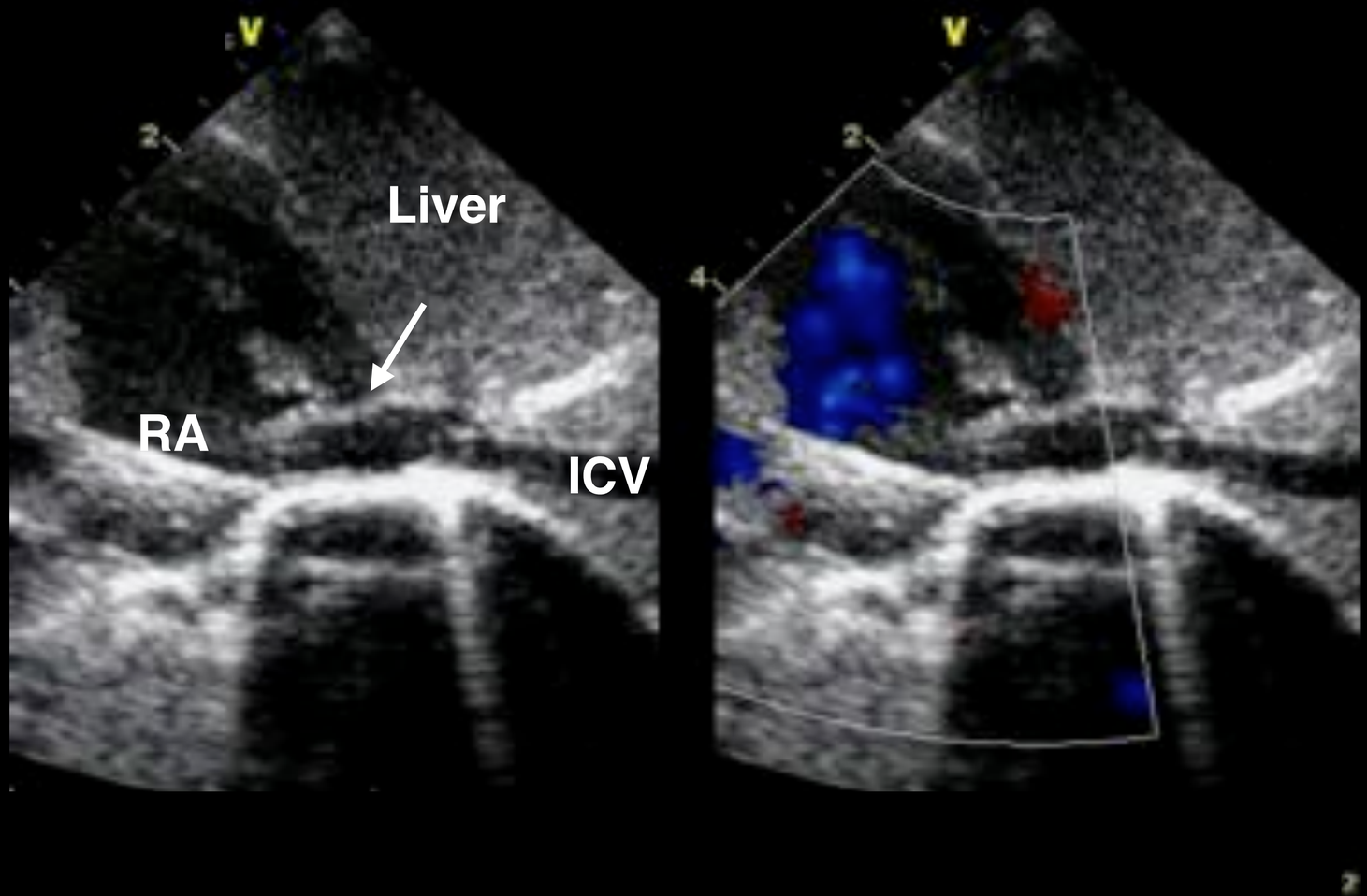
Right heart obstructive diseases



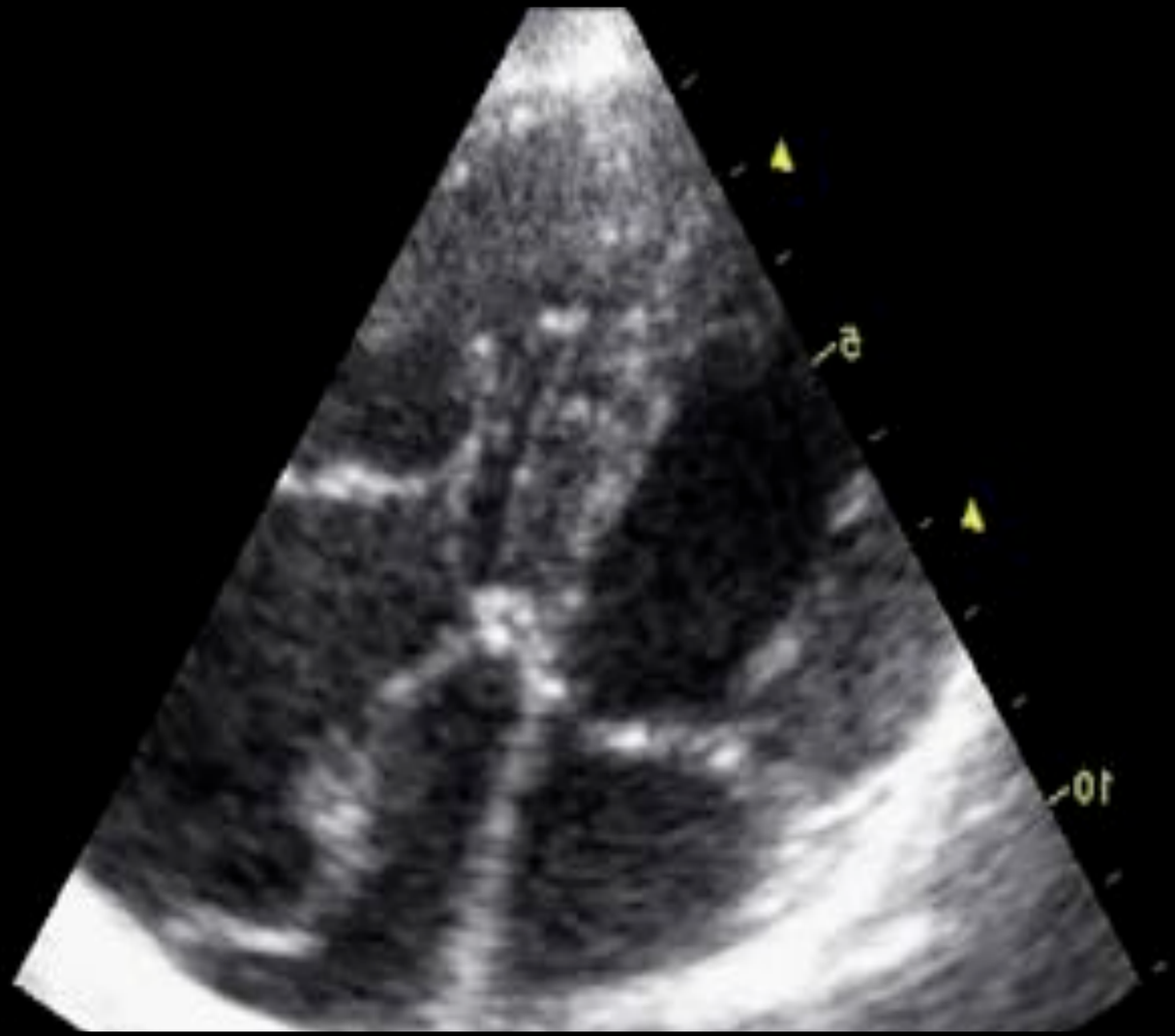

SCV
Flap
valve
TV
ICV


PV

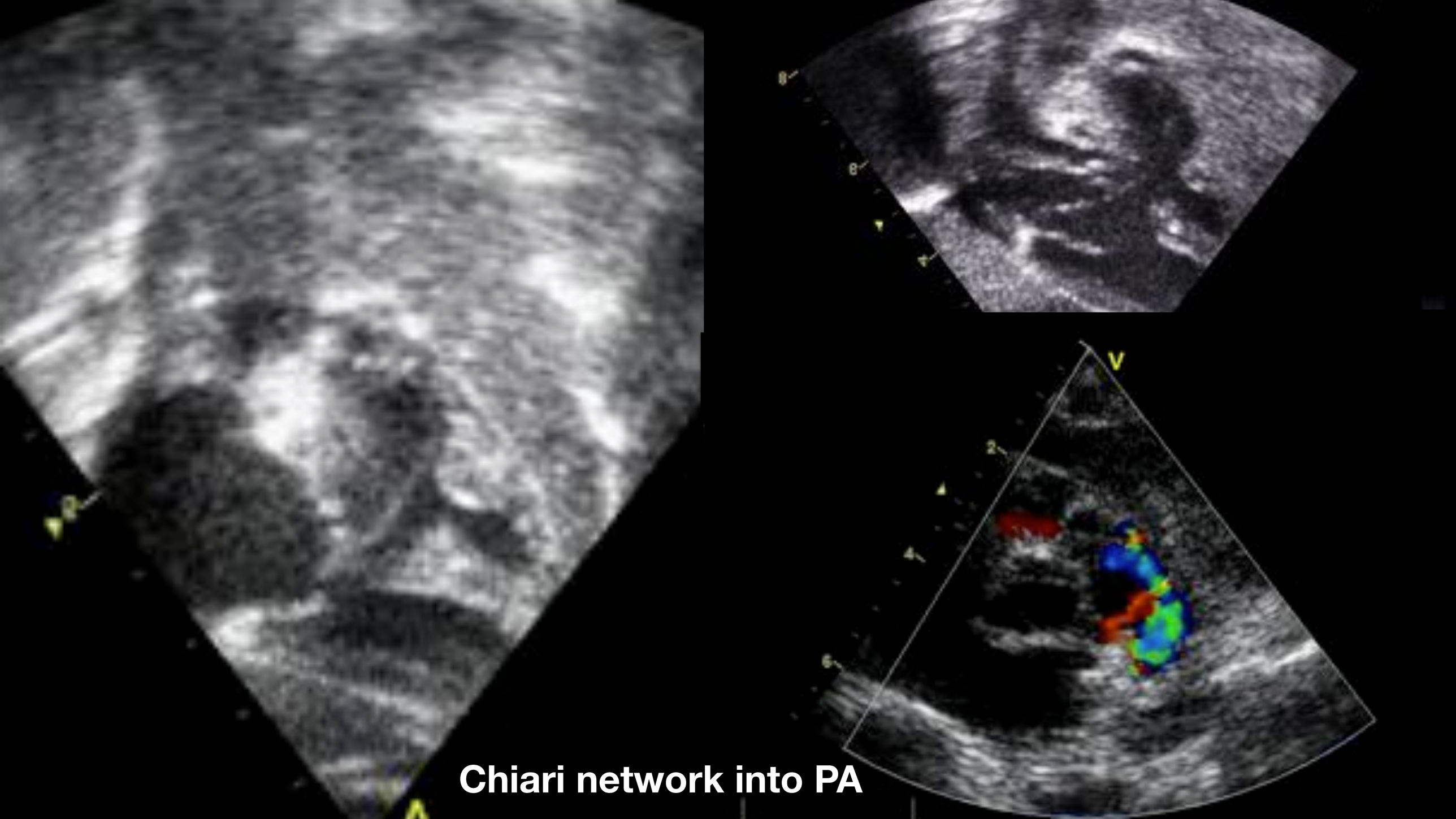
Eustachi Valve



Eustachi Valve
& Chiari network

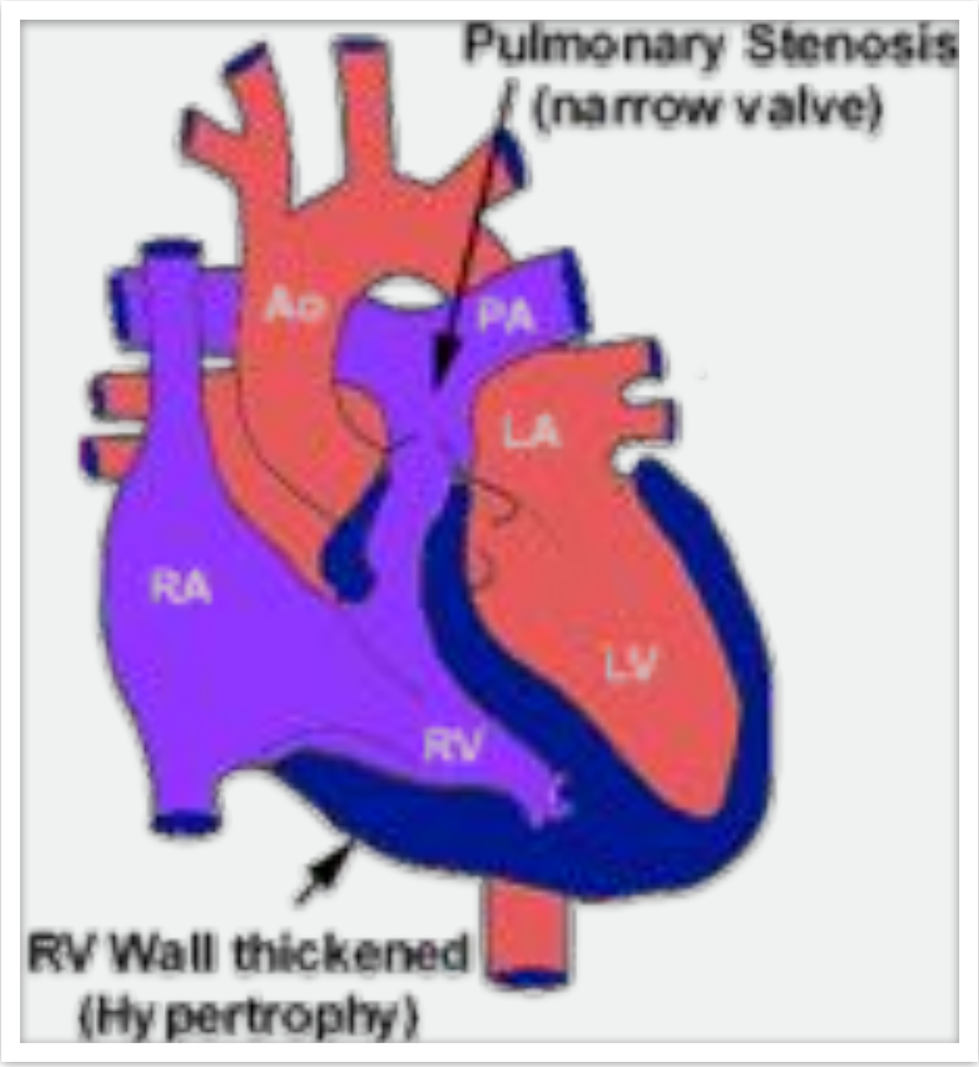


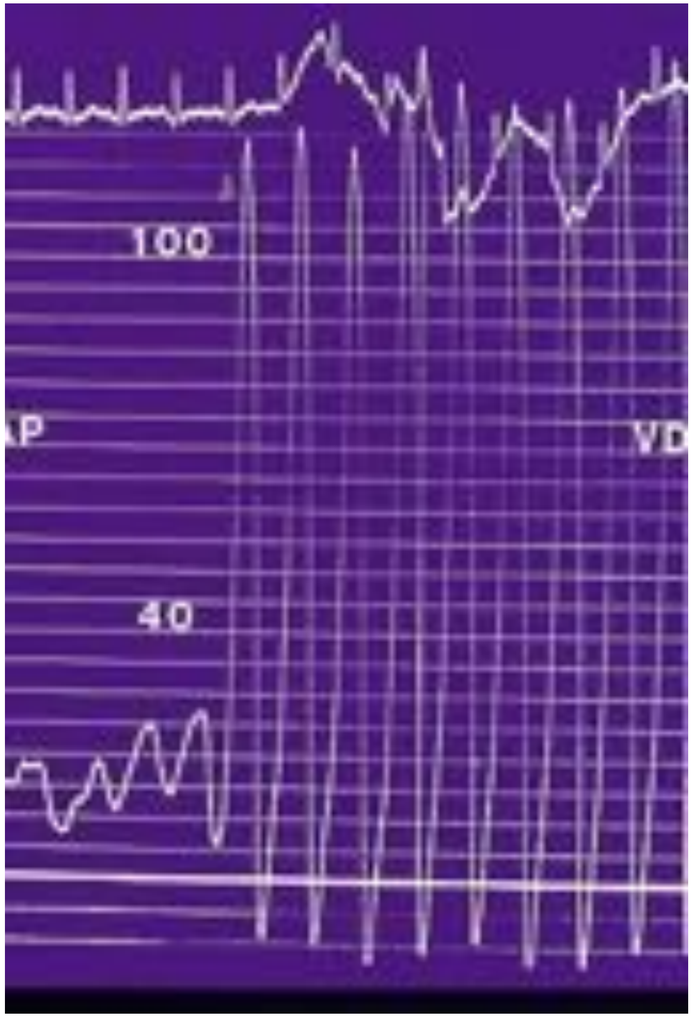
Cor triatriatum dexter


Chiari network into PA

Pulmonary valve

Pulmonary valve stenosis


Pulmonary Stenosis
(narrow valve)
Ao
PA
LA
RA
LV
RV
RV Wall thickened
(Hypertrophy)

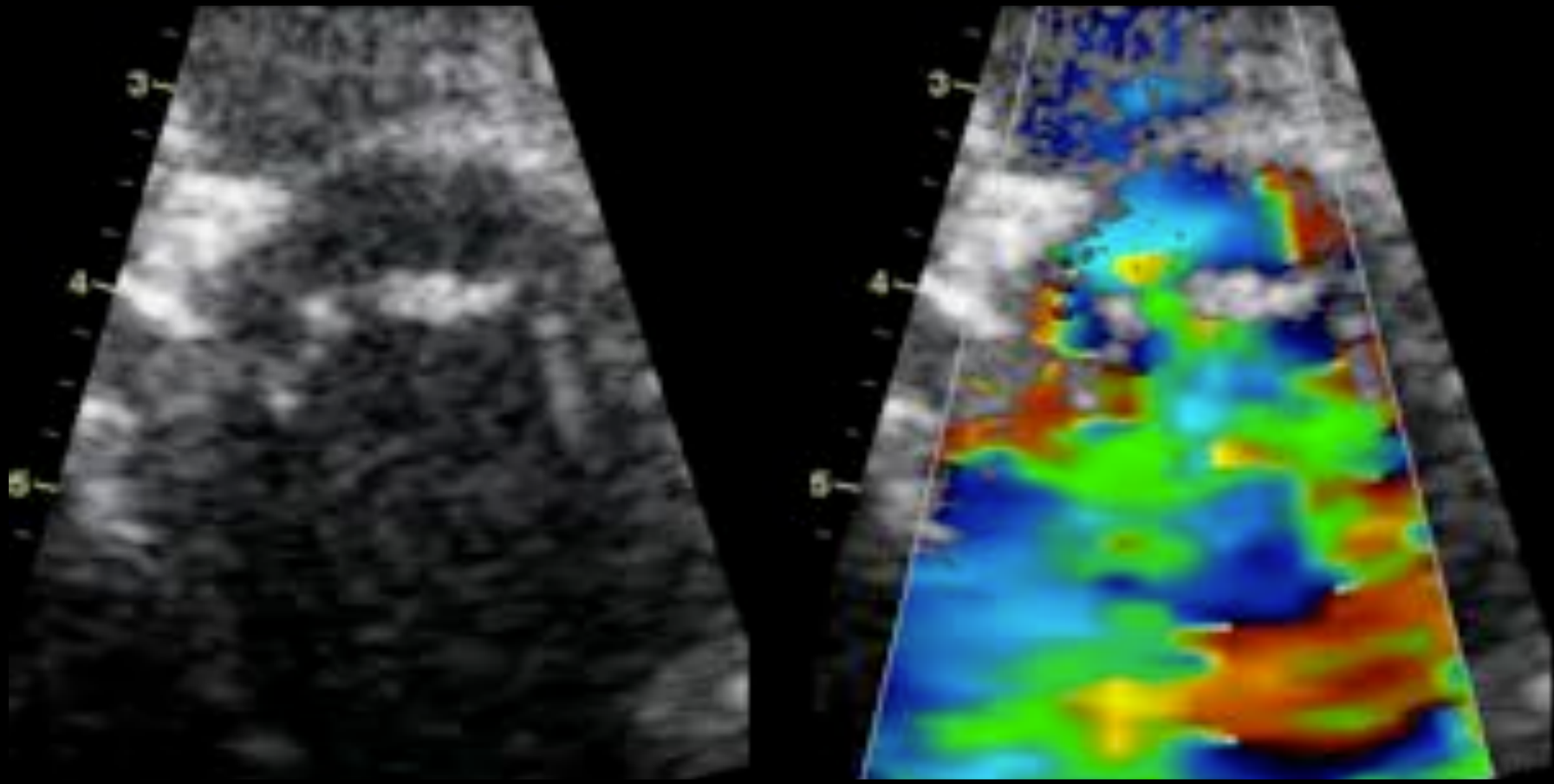

100
40

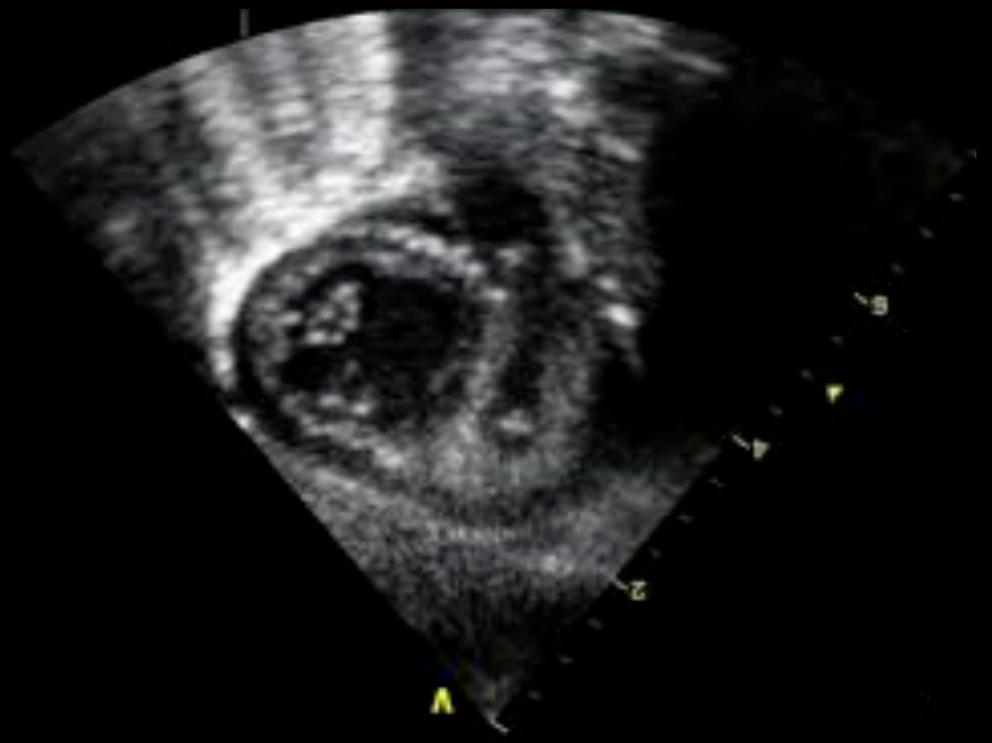
Questions in pulmonary valve stenosis

- Critical or not ?
- Associated cardiac and extra cardiac anomalies ?
- How to predict success of percutaneous dilatation :
  - valve, annulus, pulmonary arteries
- Evolution during infancy
- Restenosis

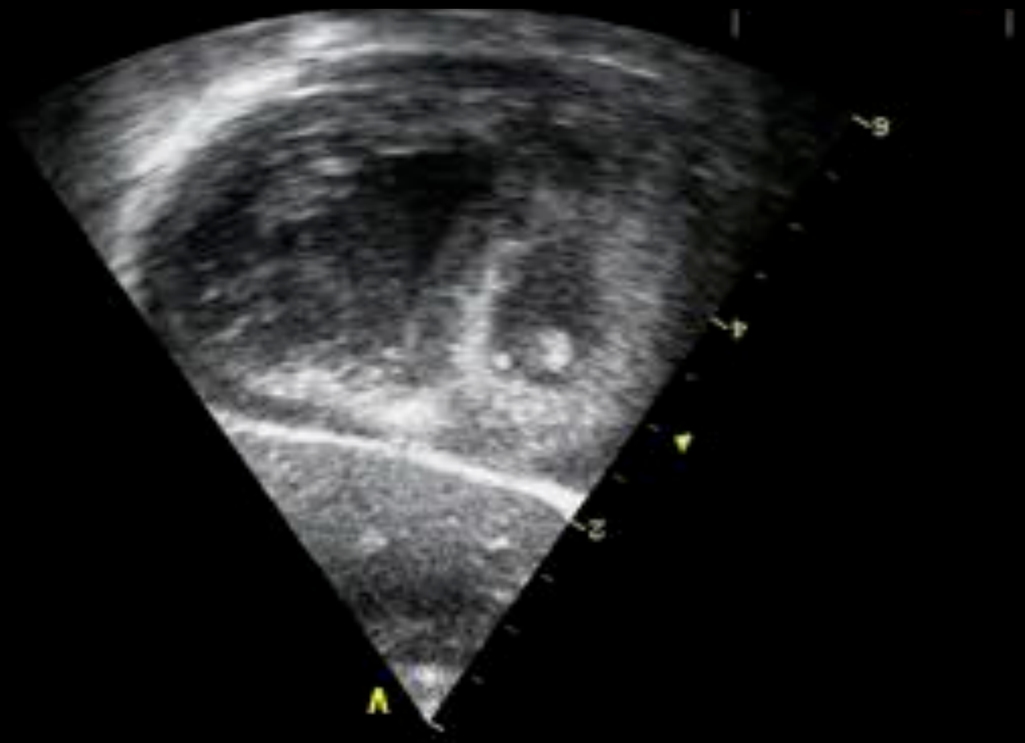
Pulmonary valve stenosis

Pulmonary valve stenosis



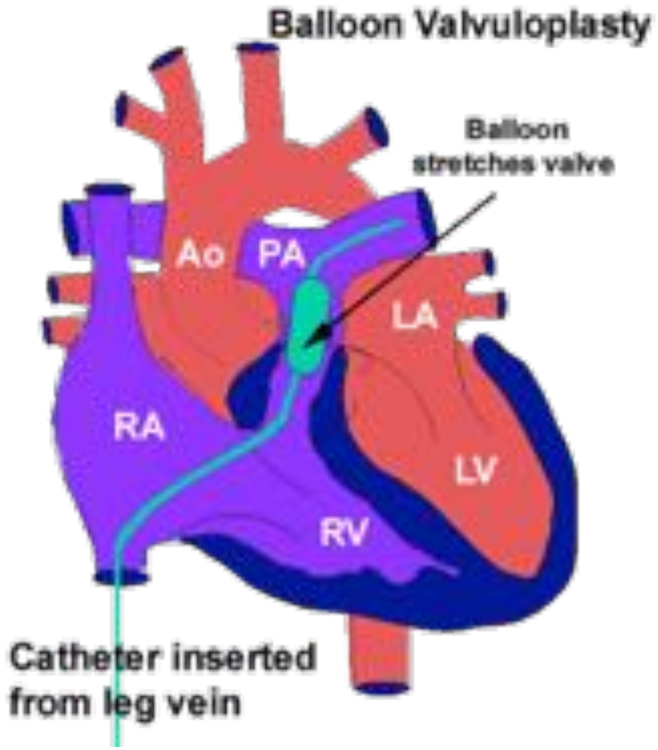


Pulmonary valve stenosis

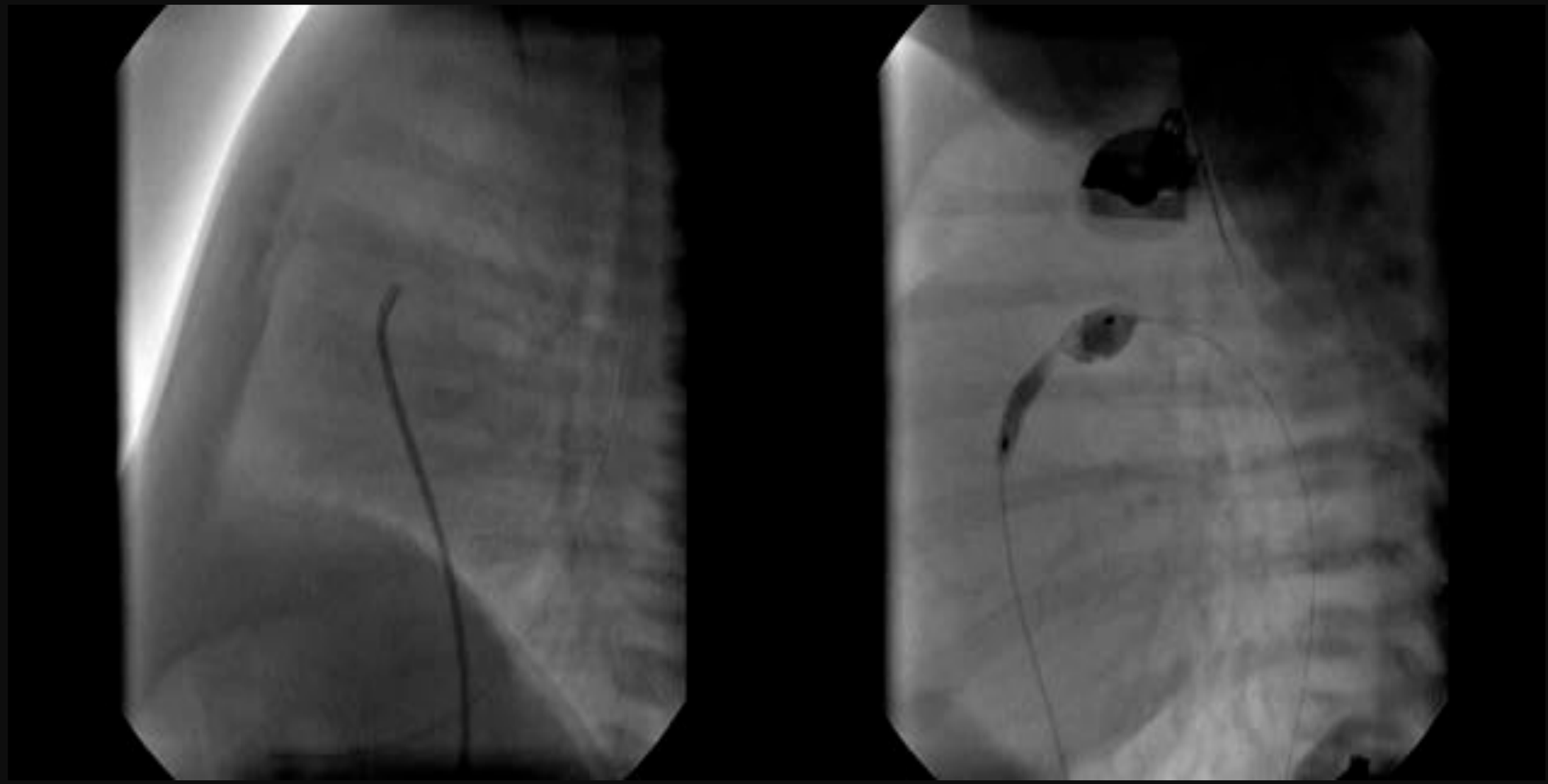


Pulmonary atresia
intact ventricular septum

Percutaneous dilatation



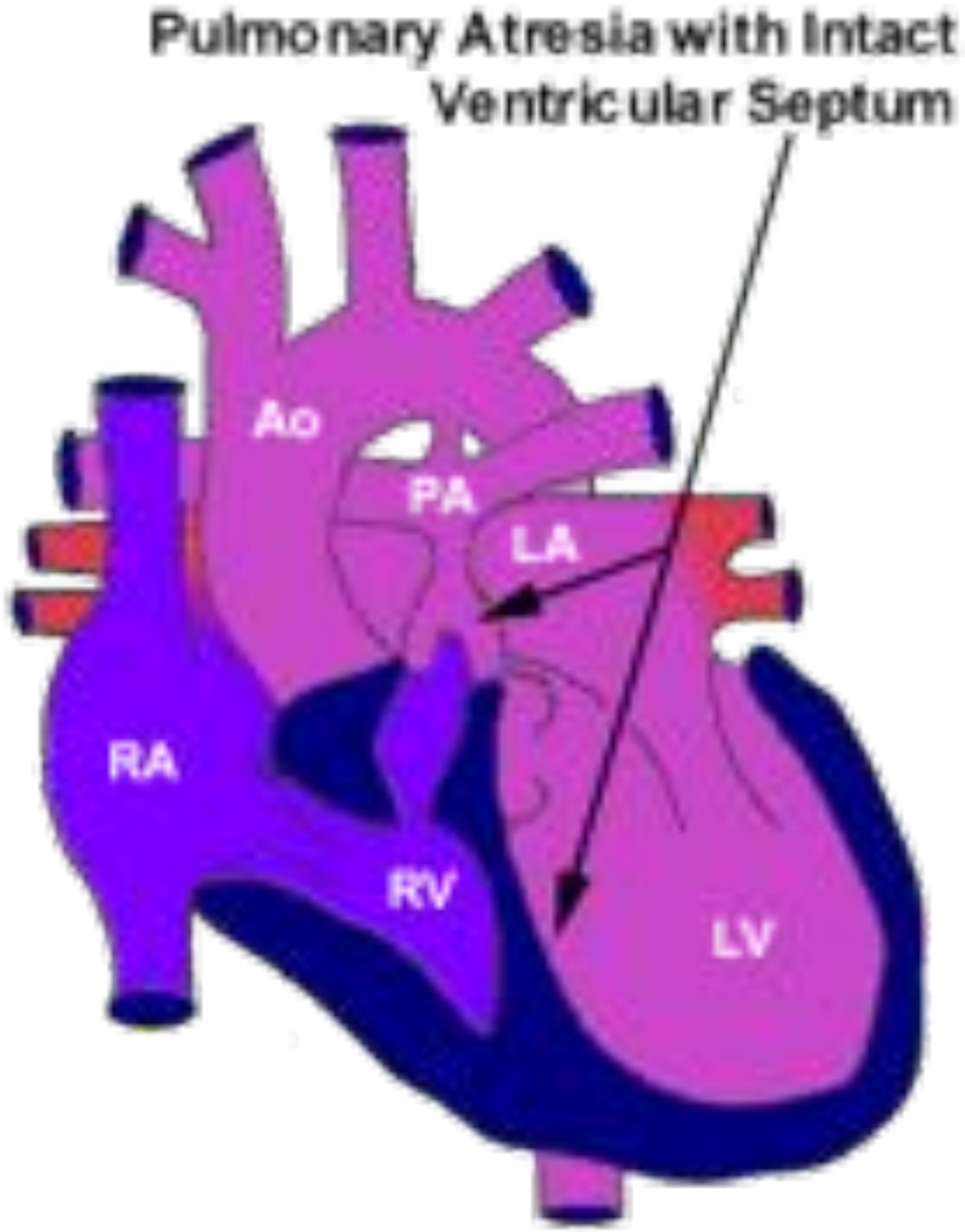
Percutaneous dilatation



Pulmonary valve after dilatation

Pulmonary valve

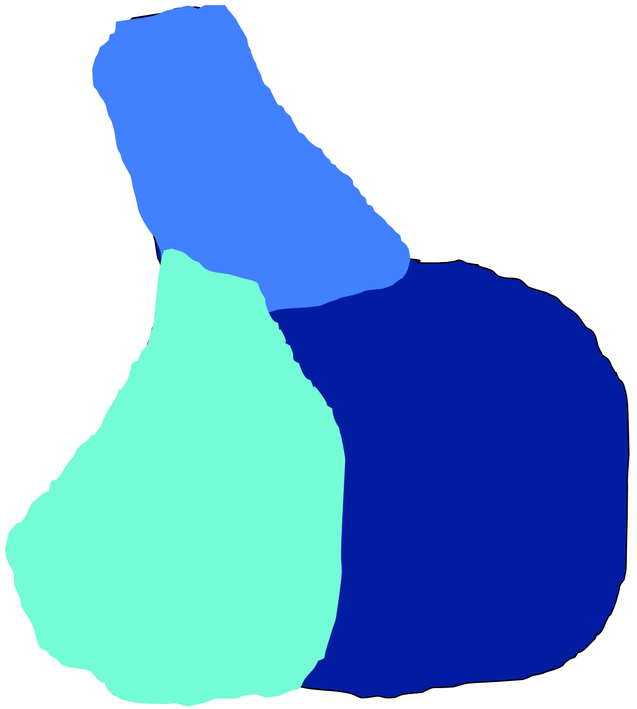
PA-IVS

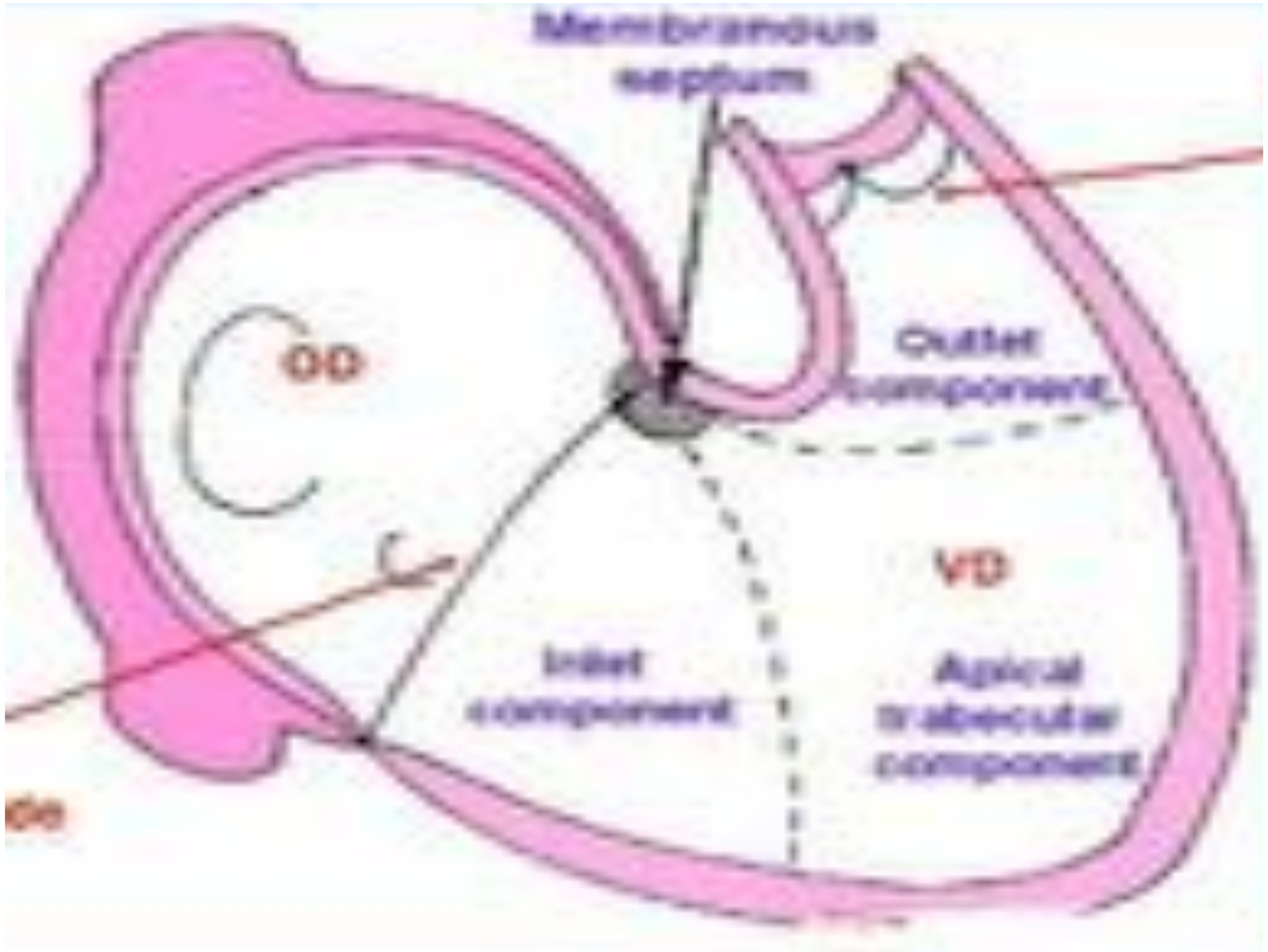

Pulmonary Atresia with Intact Ventricular Septum
Ao
PA
LA
RA
RV
LV

PA-IVS

- Evolution during foetal life
- Right ventricular anatomy
  - 1,2,3 parts
  - Size of tricuspid valve and function
  - Pulmonary annulus
- Coronary anatomy
  - Fistulae
  - RV coronary perfusion


PV



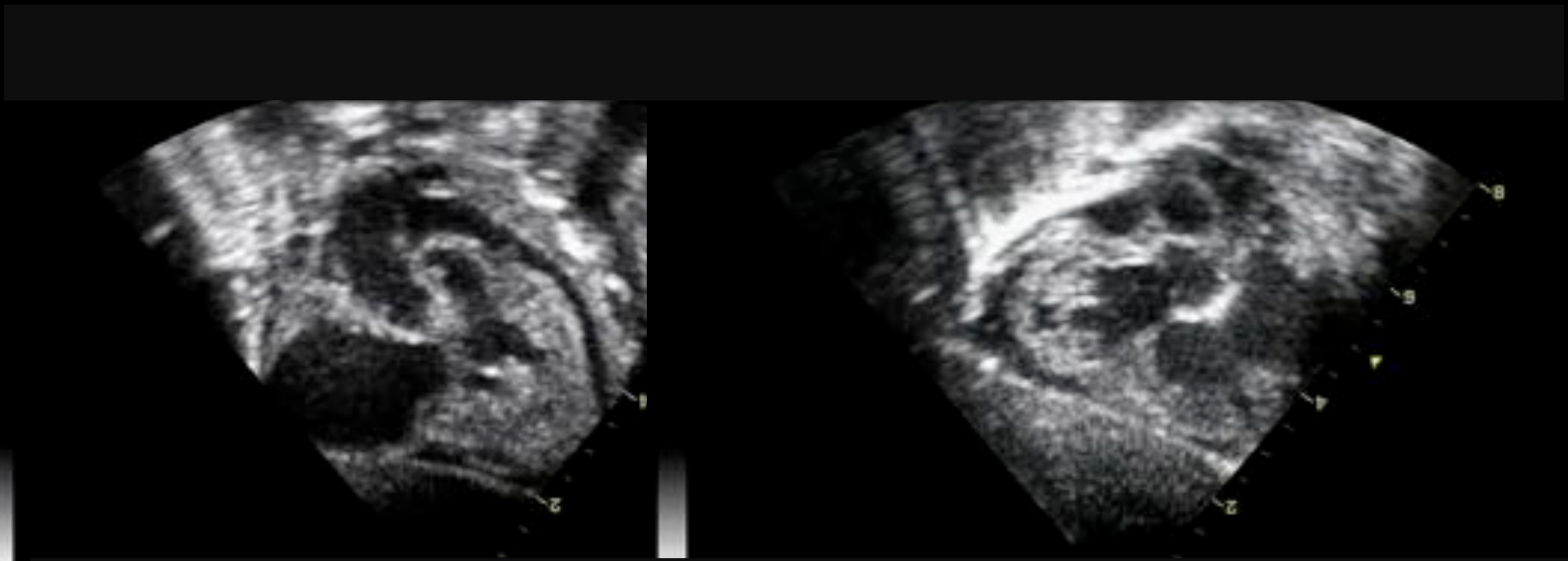

Membranous septum
OD
Outlet component
VD
Inlet component
Apical trabecular component

PA-IVS
Therapeutic strategy

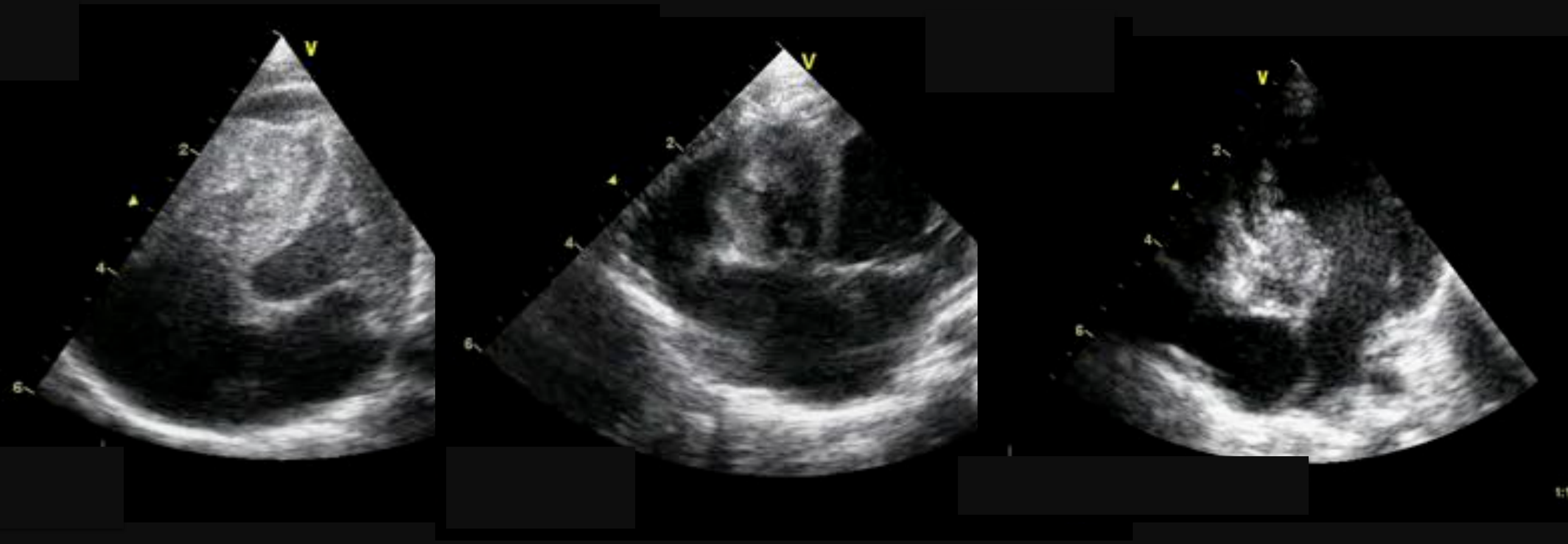
- Tripartite
  - 2 ventricles
- Bipartite : inlet + infundibulum
  - 1,5 ventricle
- Unipartite
  - 1 ventricle

Most important information z-score of the tricuspid valve

Pulmonary atresia intact septum Tripartite Right ventricle

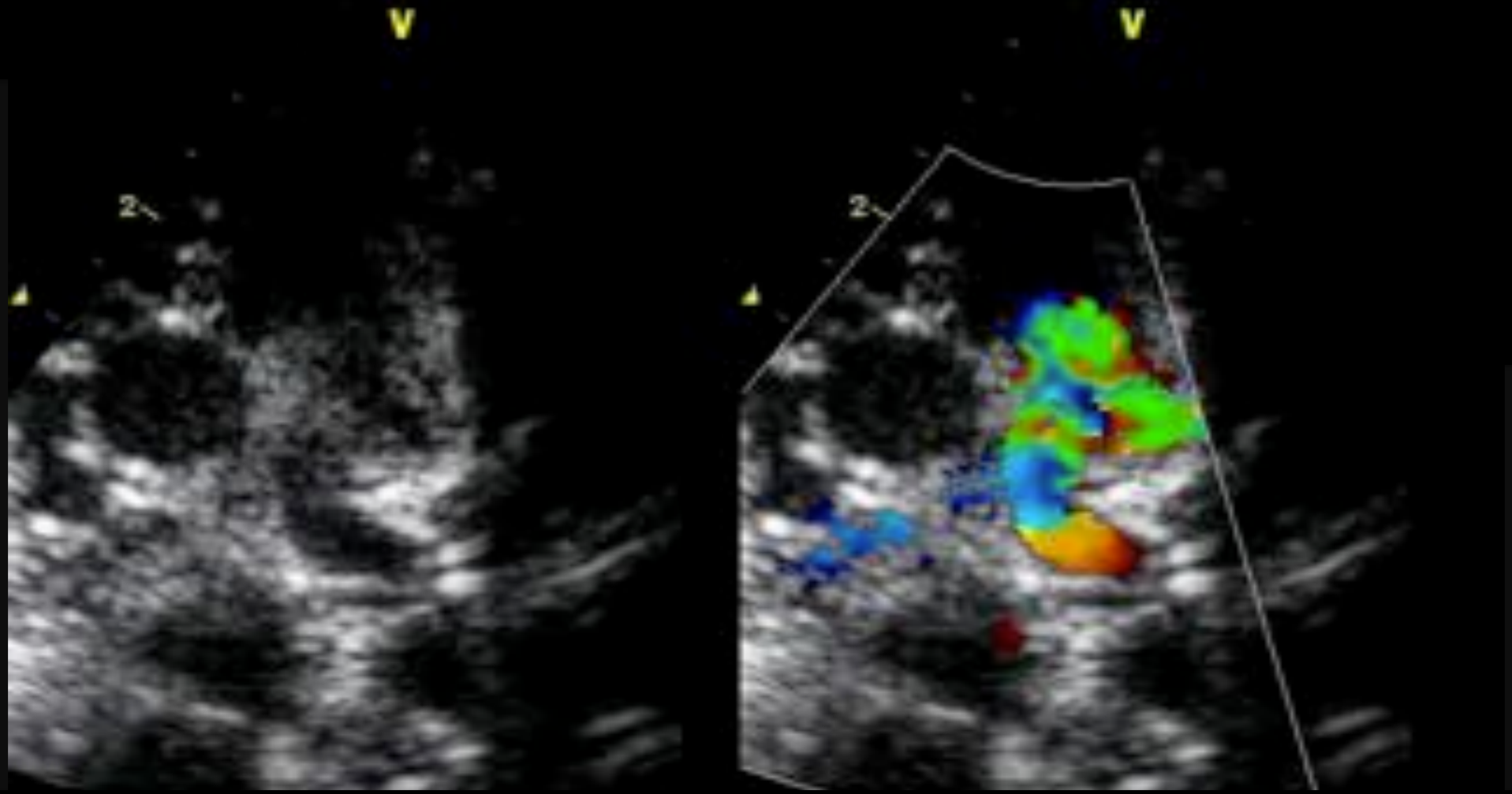


Pulmonary atresia intact septum Right ventricle (3-2-1)

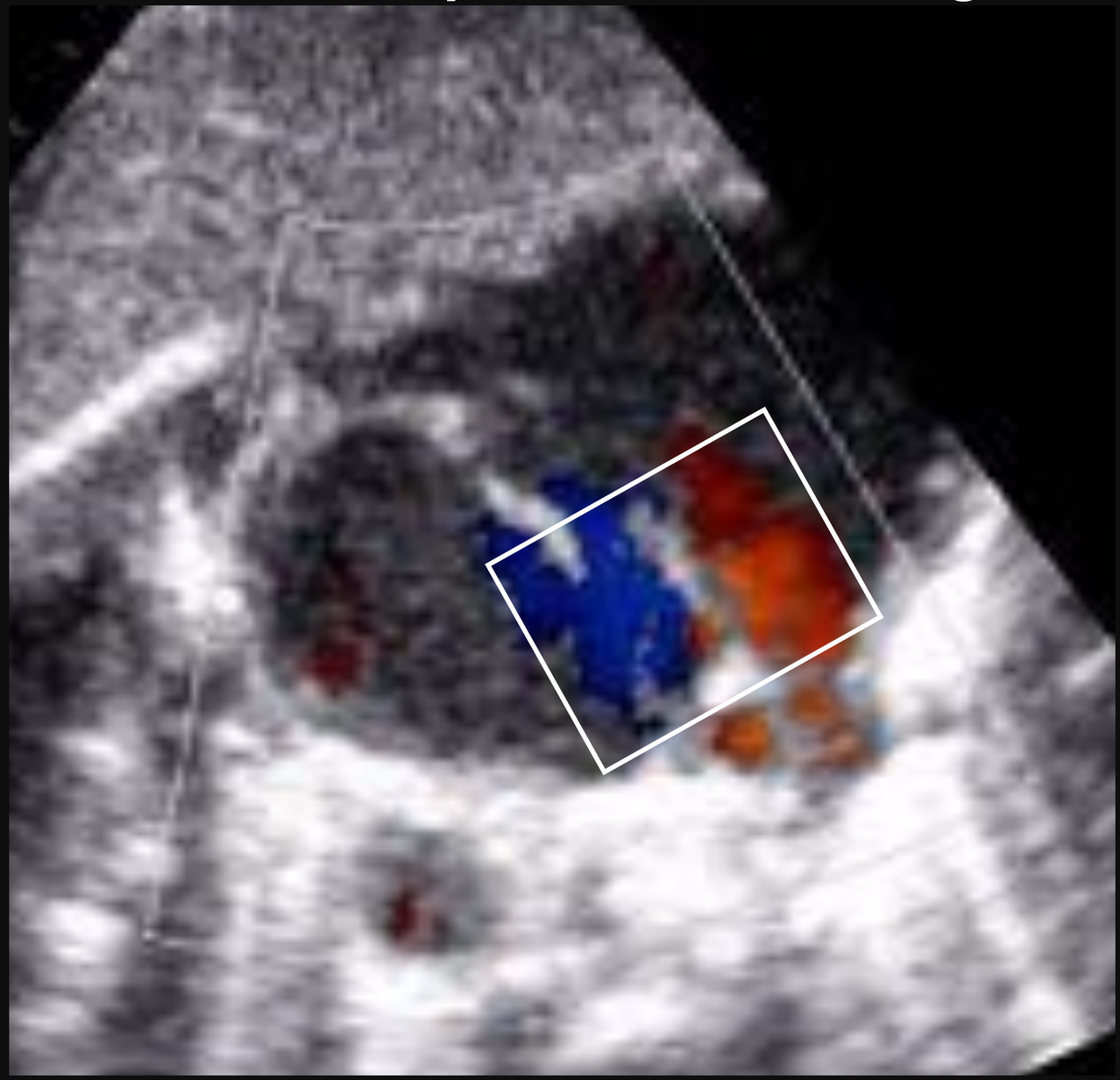


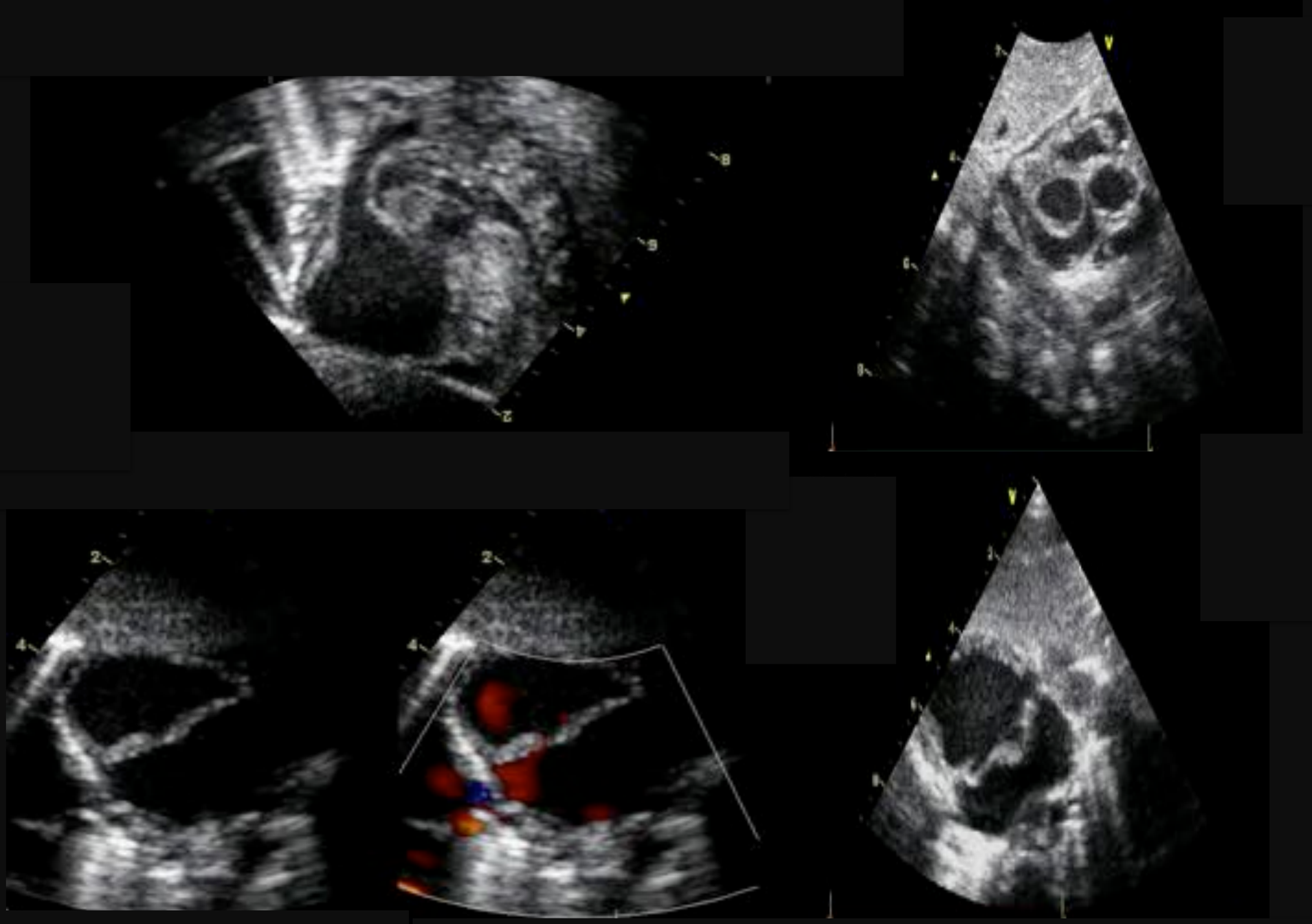
Pulmonary atresia intact septum Pulmonary valve

Pulmonary atresia intact septum Arterial duct flow



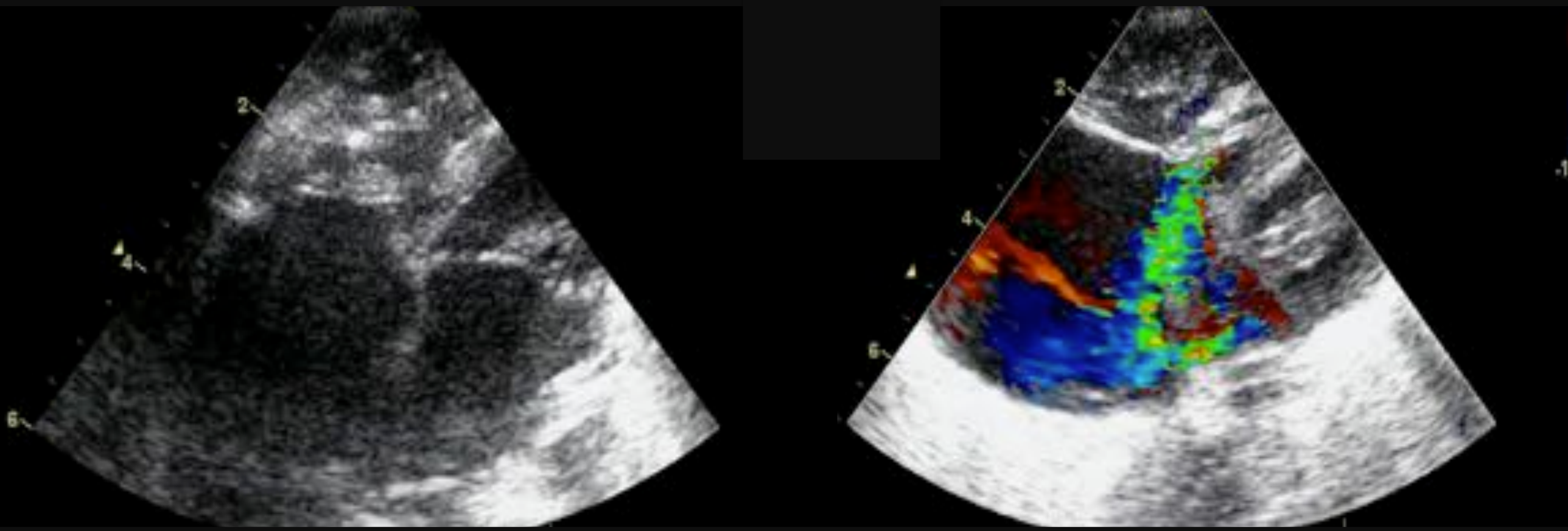
Pulmonary atresia intact septum Interatrial Right to Left shunt



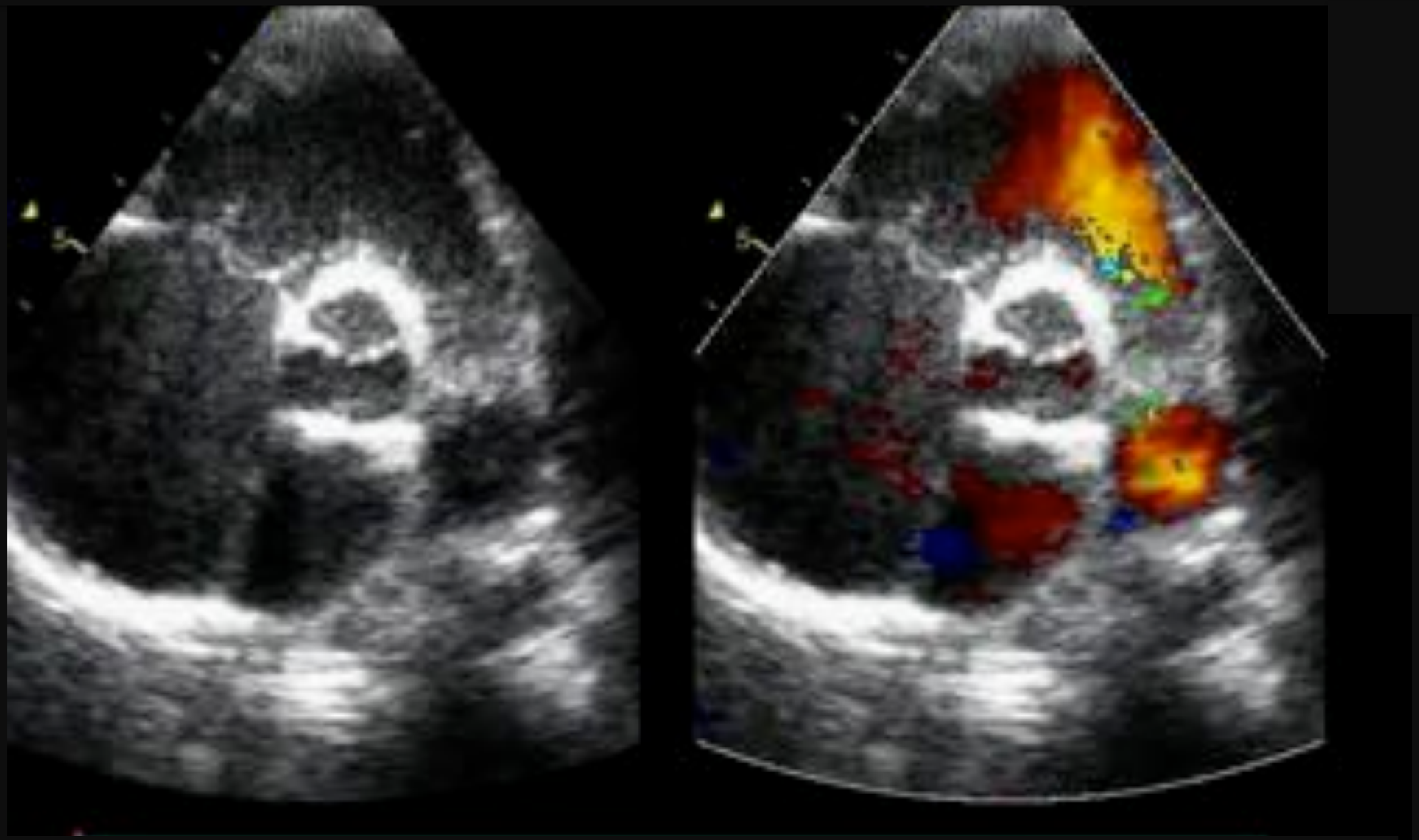


RA and FO in PA-IVS

Tricuspid valve in PA-IVS

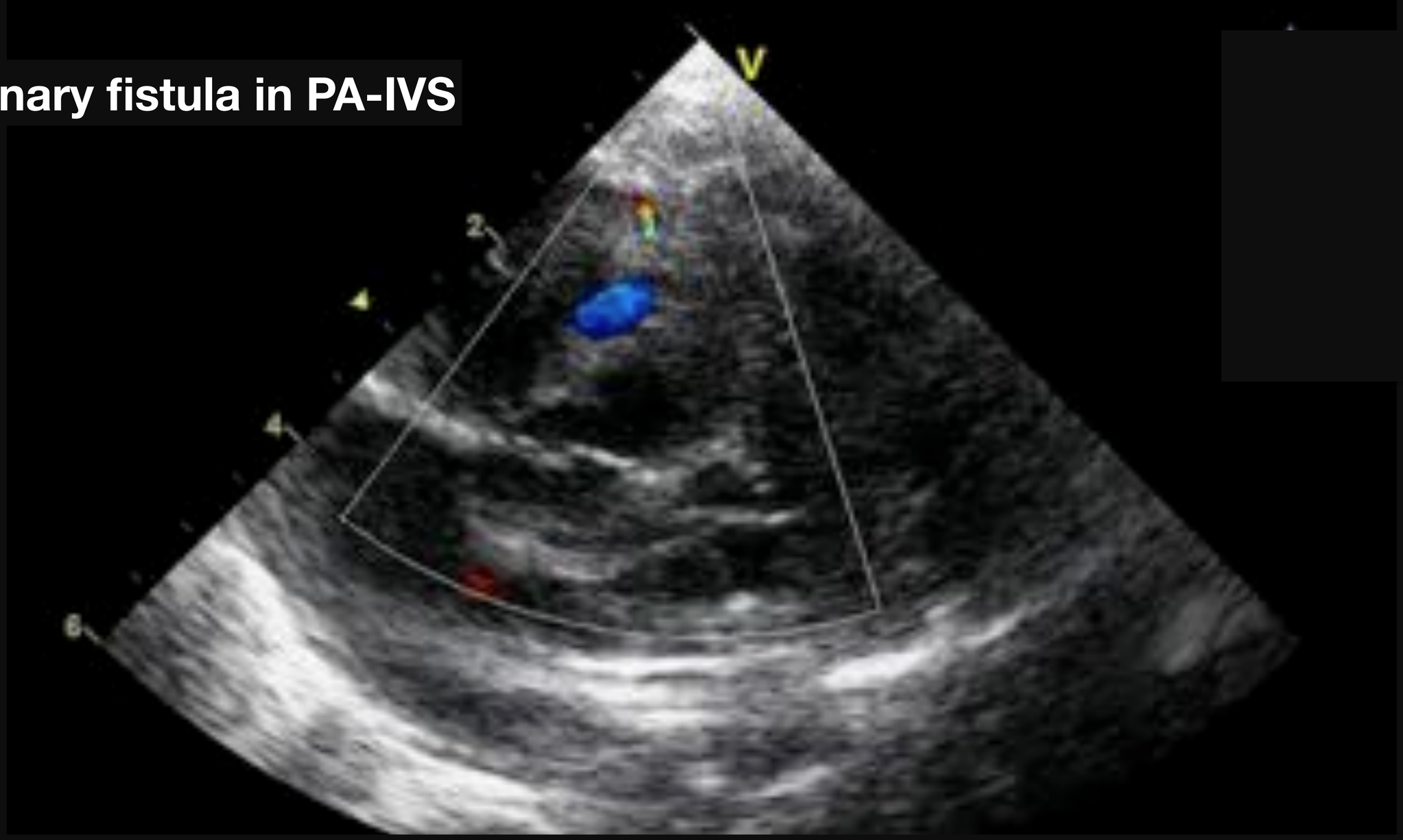


Tricuspid valve dysplasia

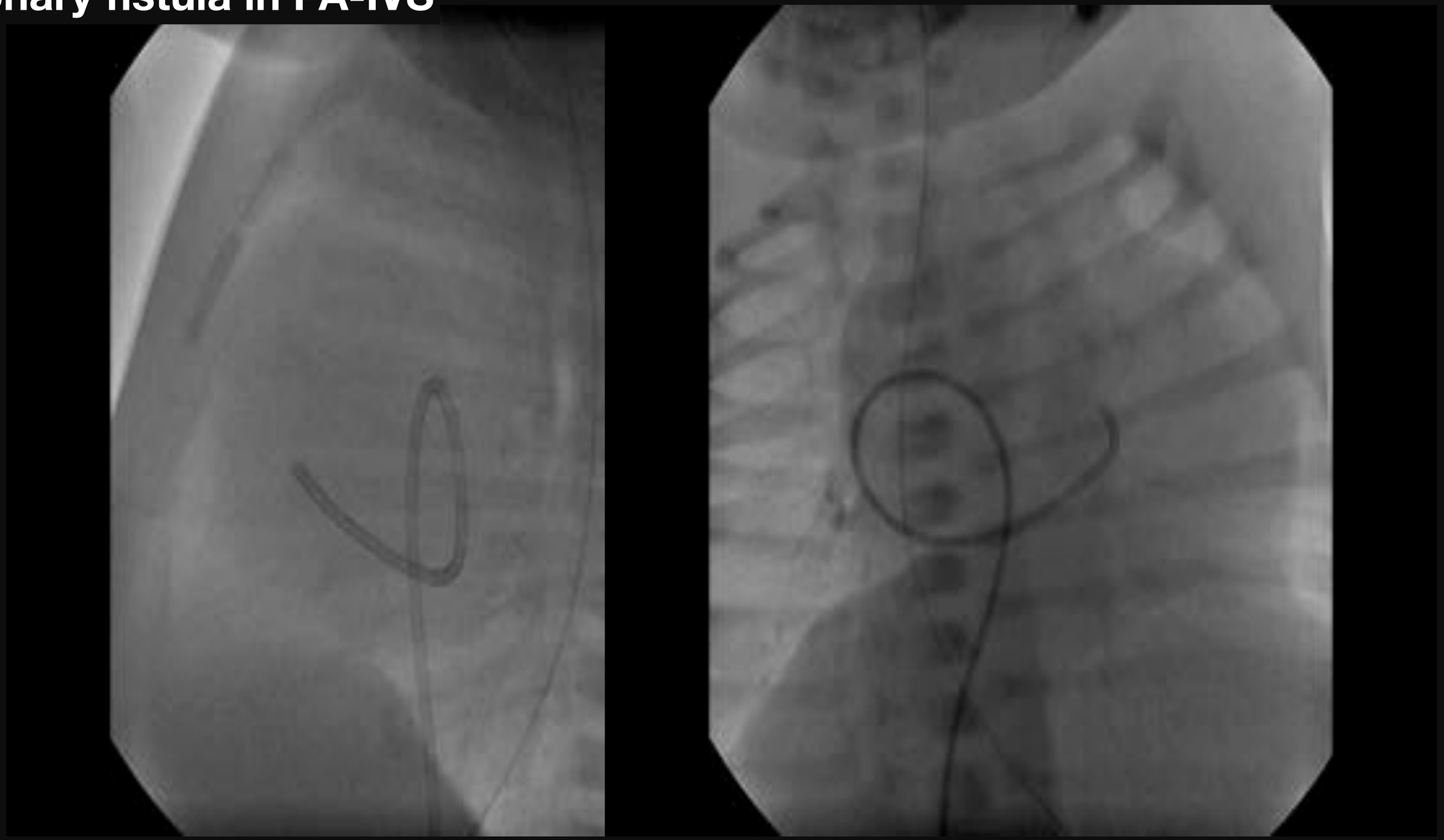


Tricuspid valve dysplasia and PA-IVS/the circle of death

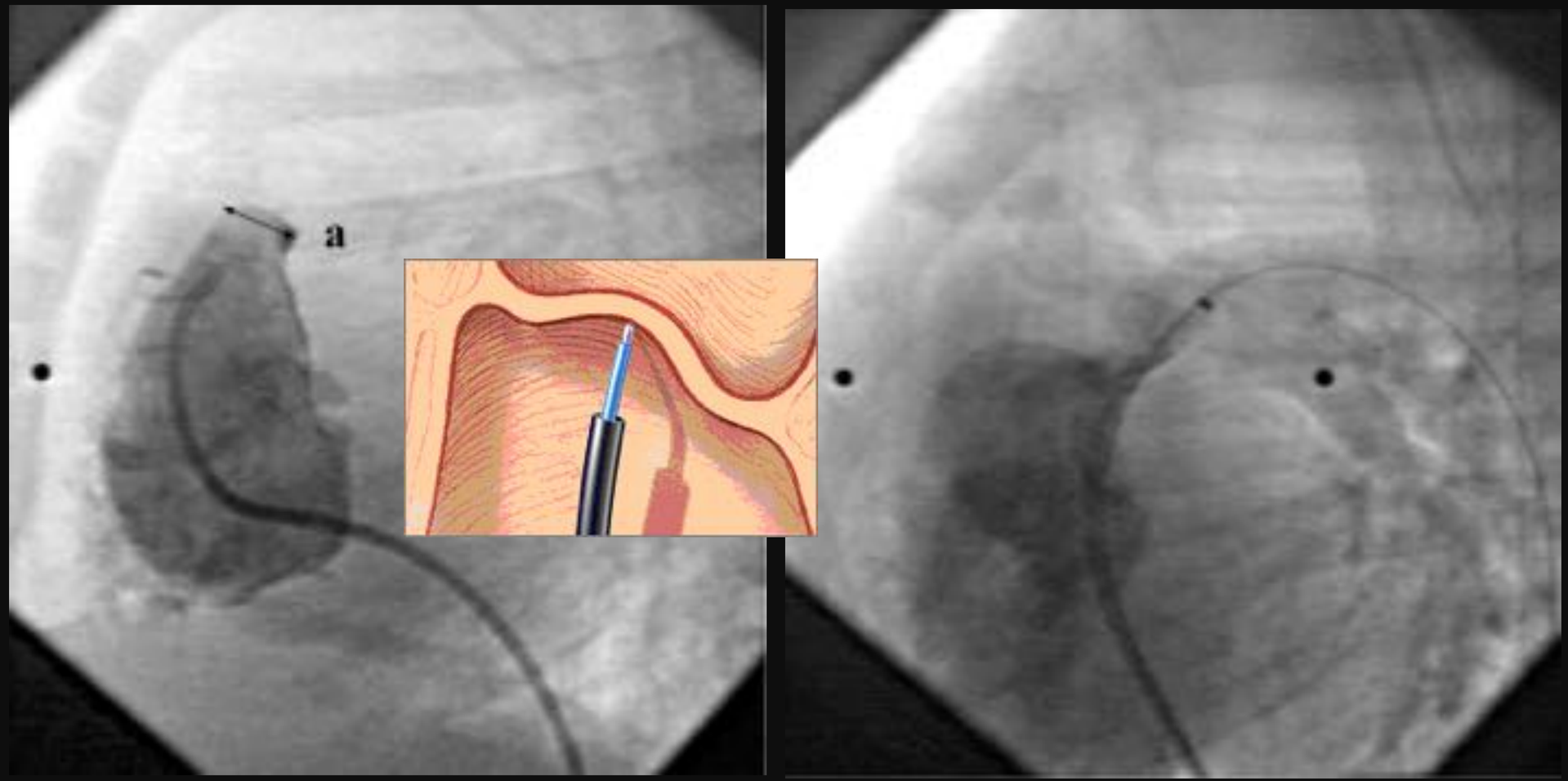
Coronary fistula in PA-IVS



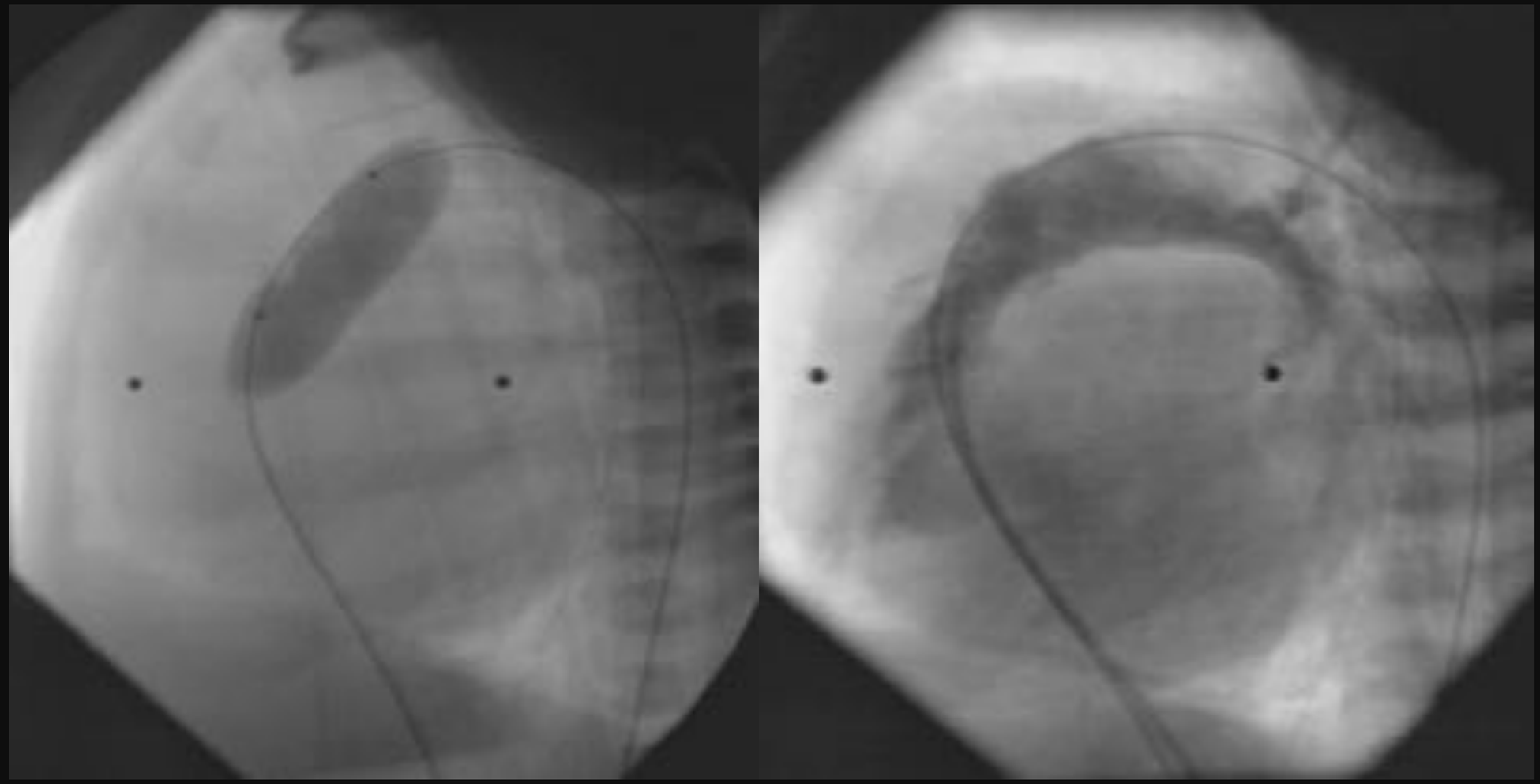
Coronary fistula in PA-IVS



Perforation of pulmonary valve and dilatation in PA-IVS



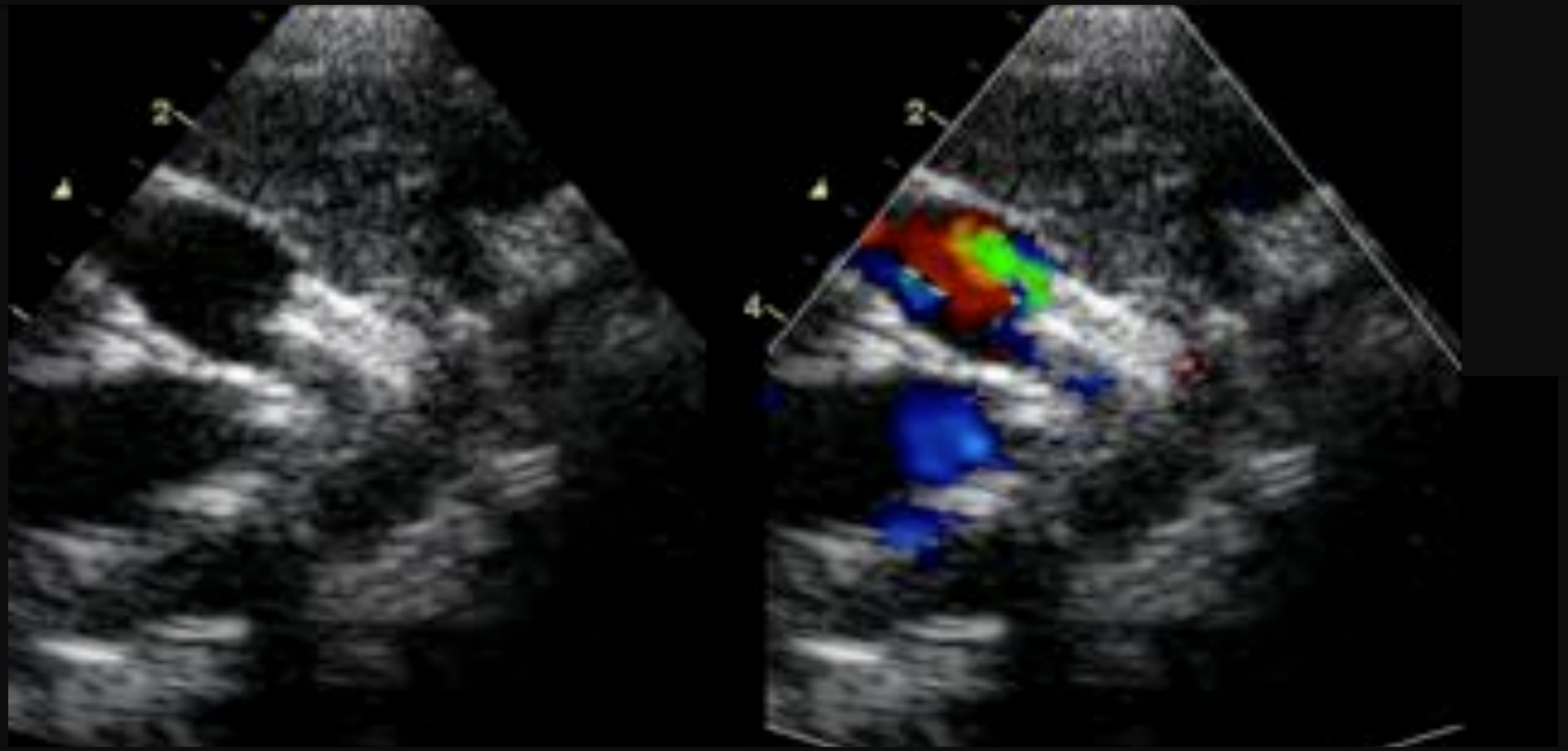
Perforation of pulmonary valve and dilatation in PA-IVS



PA-IVS

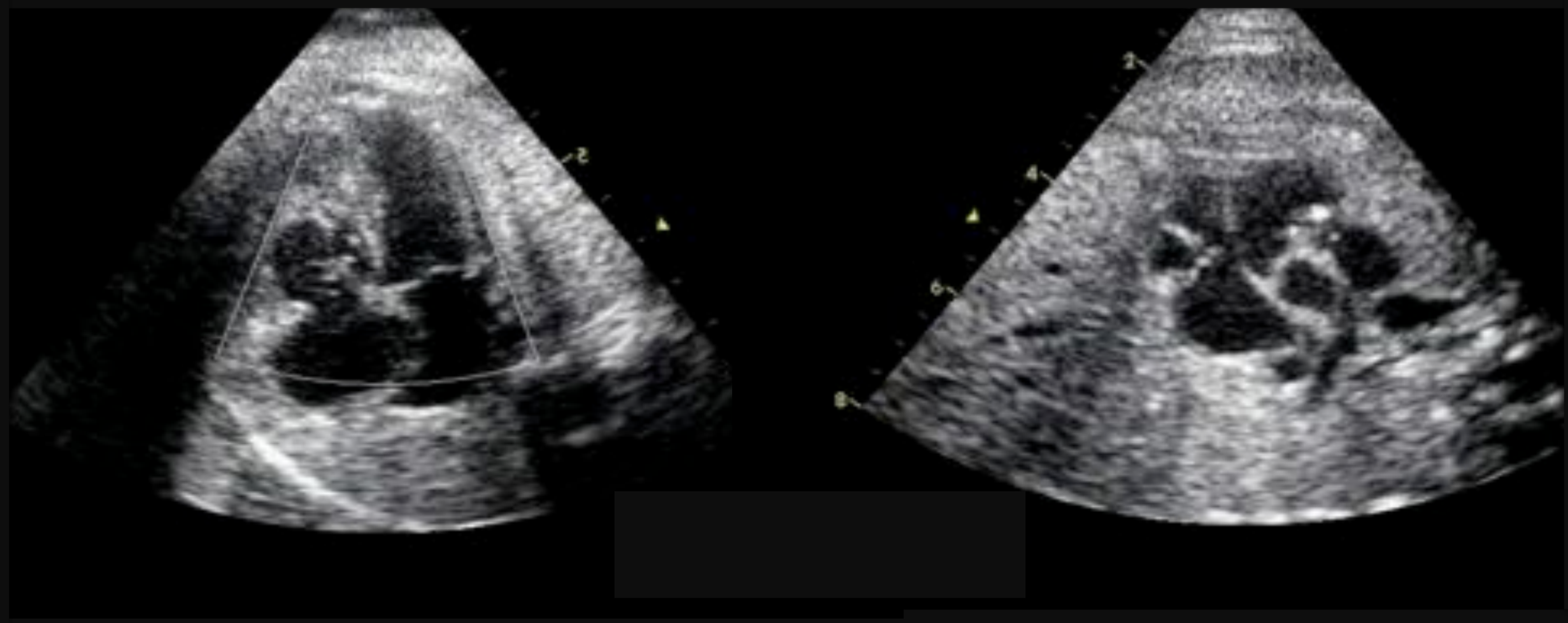
Strategy after pulmonary valve perforation

- Persisting cyanosis
  - RV Compliance
    - Beta-blockers
    - Blalock-Taussig shunt
    - Stenting of arterial duct
- Tricuspid regurgitation
- Growth of right heart structures



Stenting of the arterial duct

Stenting of the arterial duct



Foetal PA-IVS

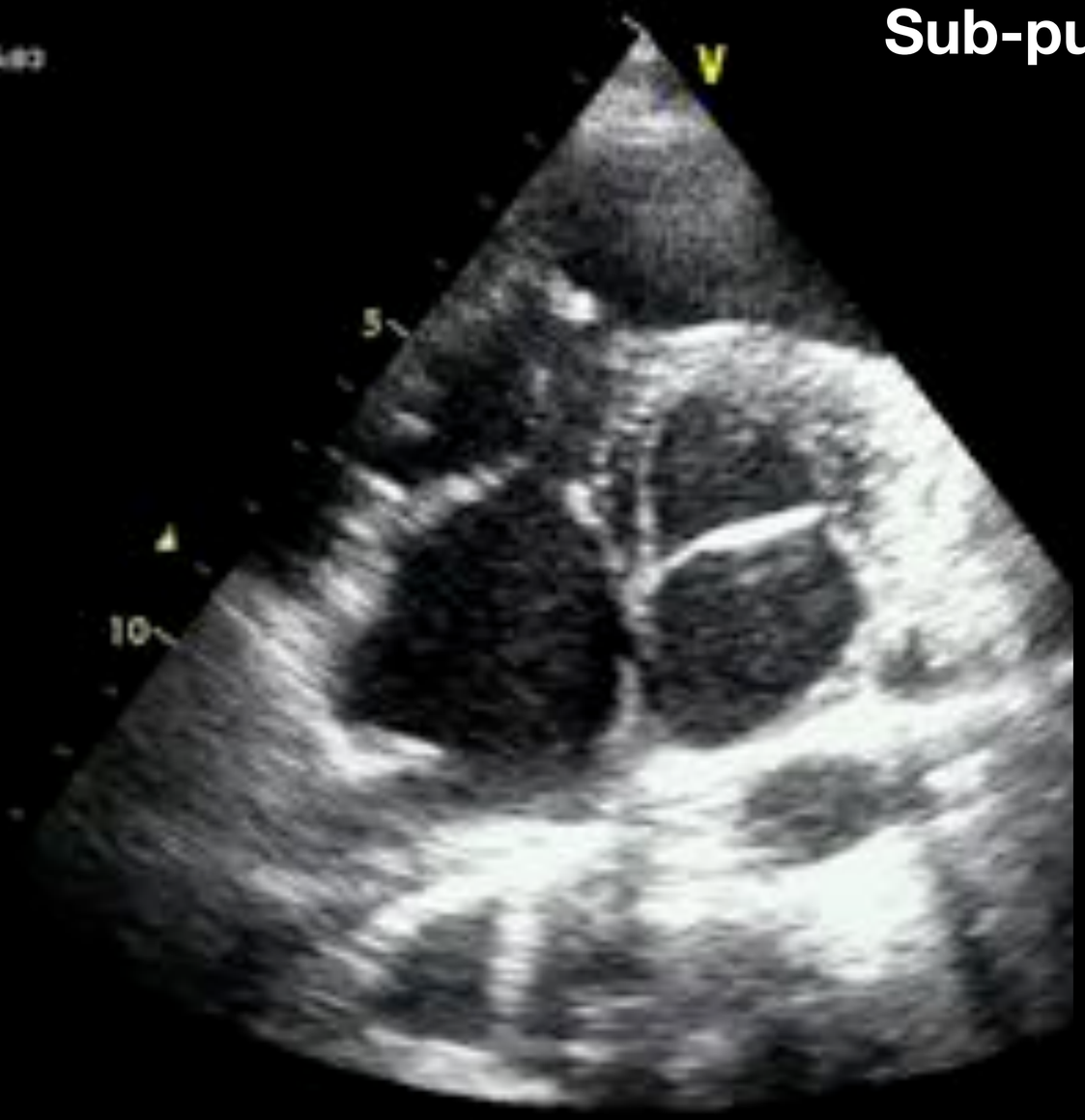
Foetal PA-IVS Interventional cath in utero

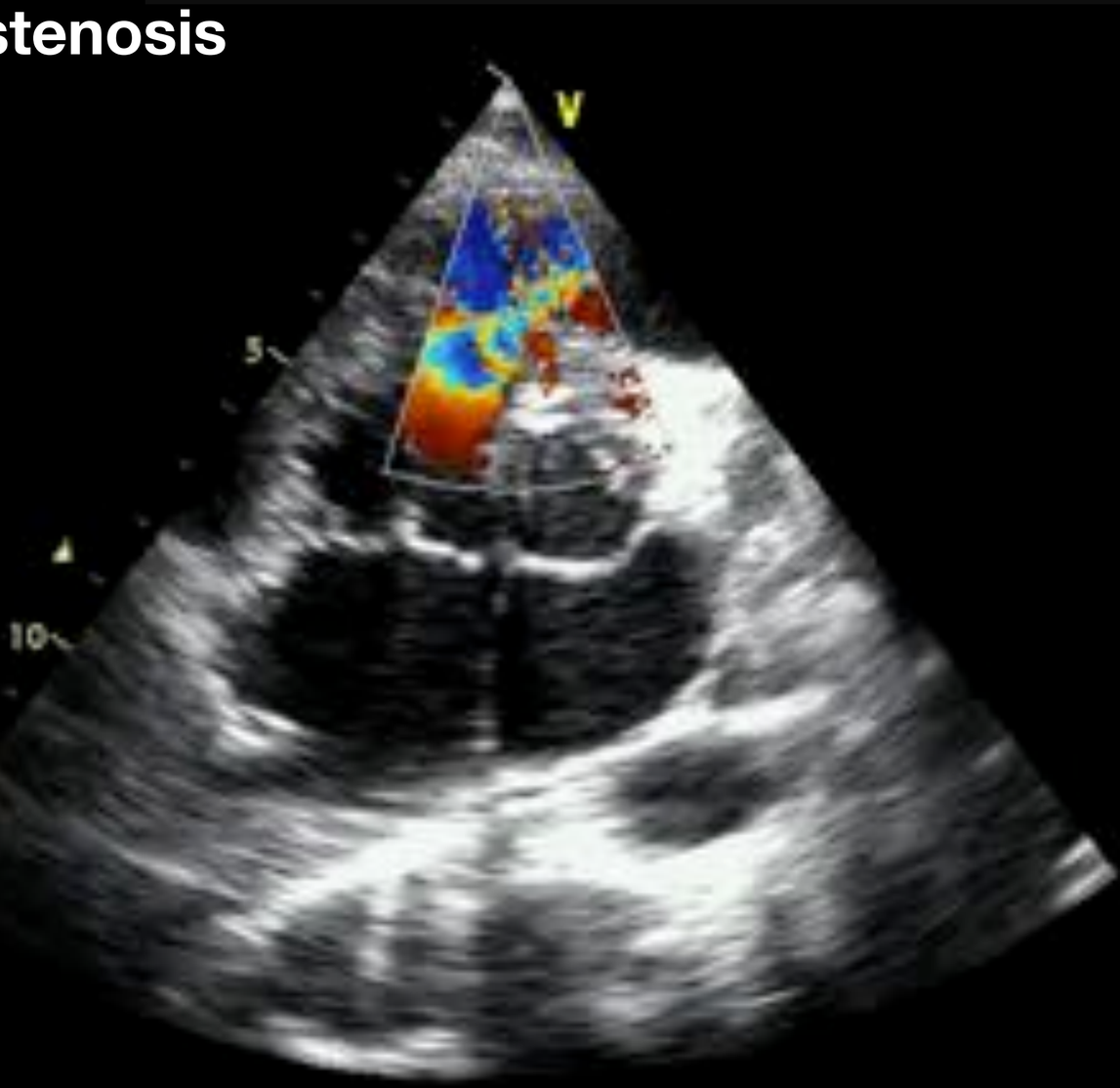
Subpulmonary stenosis

Su-pulmonary stenosis

- Isolated : rare
- Associated with VSD
  - below the obstruction : Fallot-like
  - Above : Left-to-right shunt
    - Echo booby-trap

Sub-pulmonary stenosis



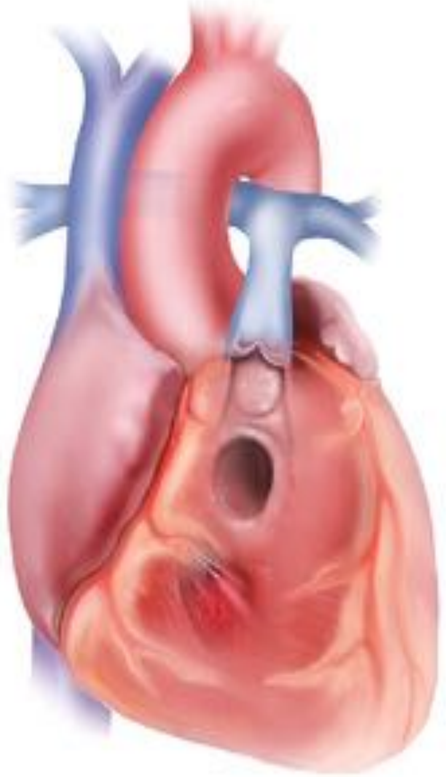


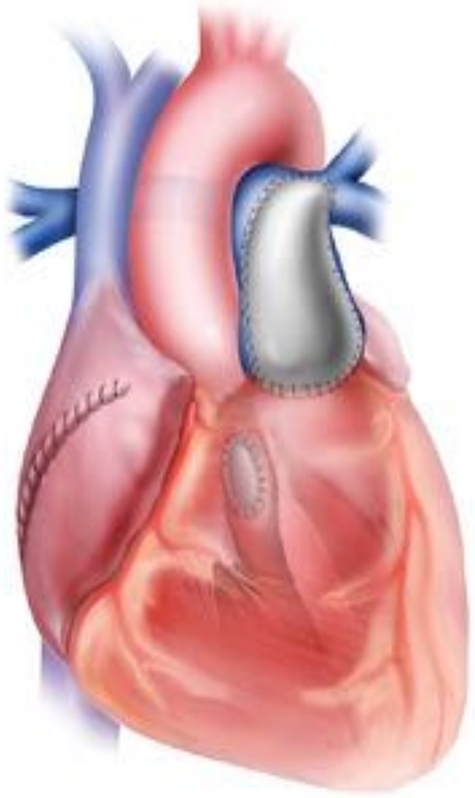

Sub-pulmonary stenosis

RV-PA Conduits

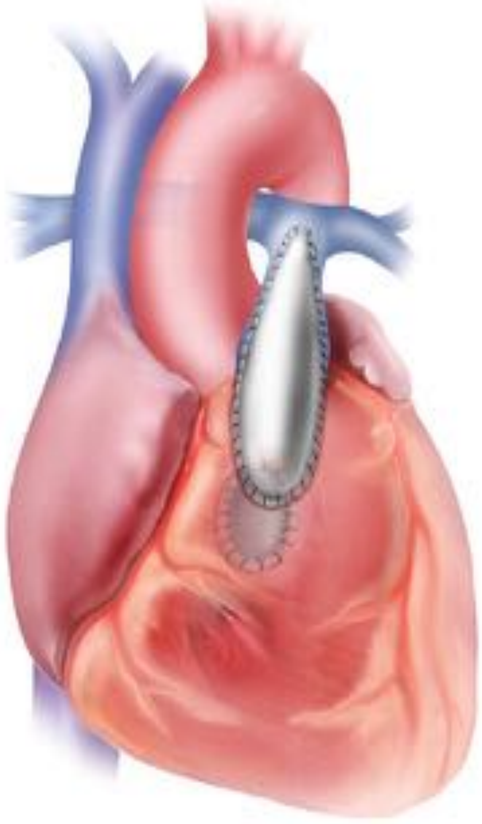
Surgical treatment of tetralogy of Fallot

Repair





Atrial route

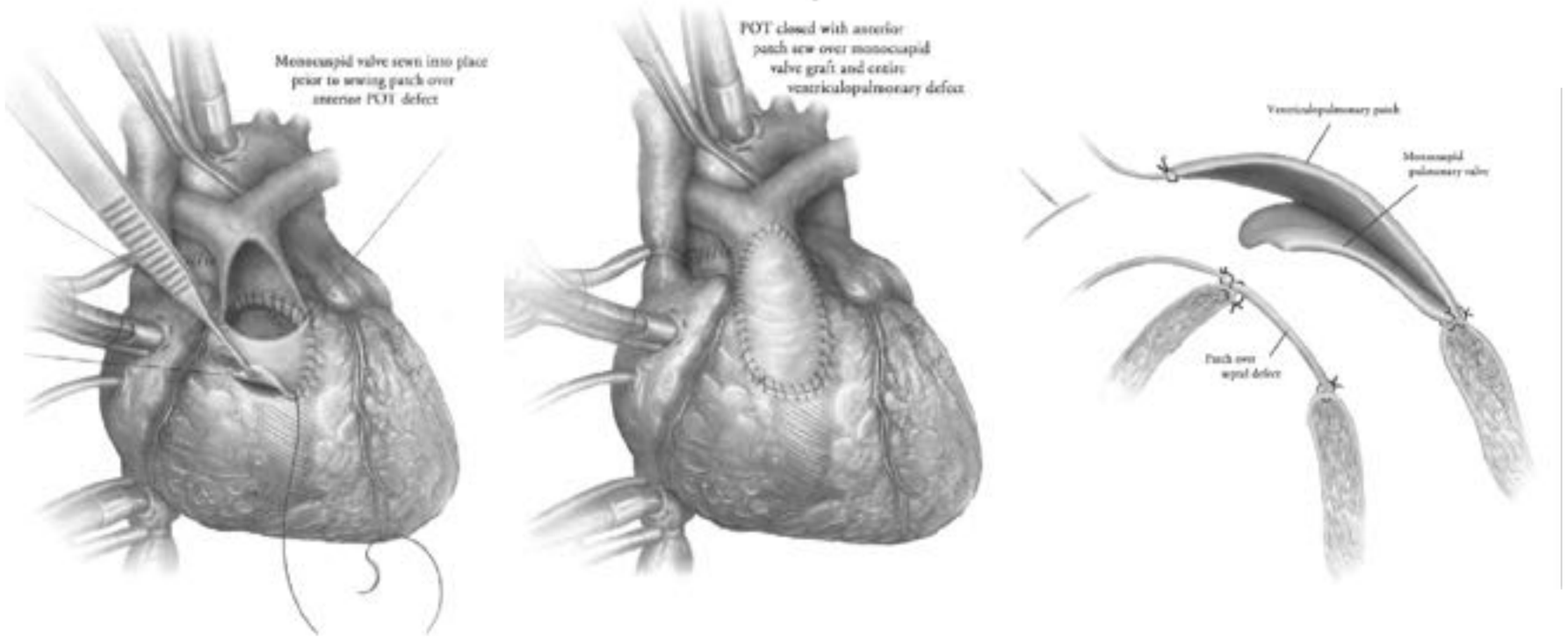


Infundiular route

Right ventricle dilatation after Fallot repair with transannular patch

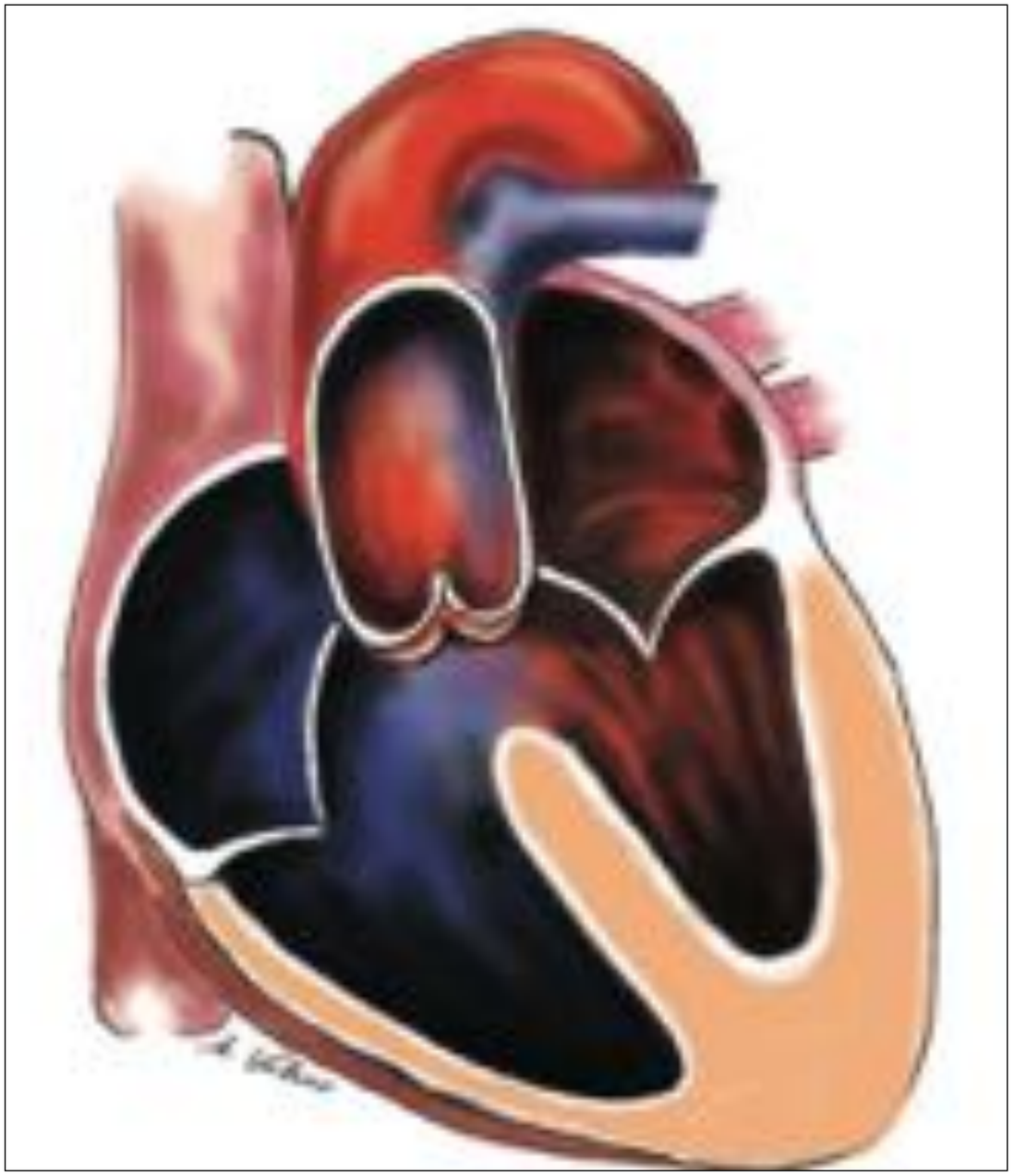
Surgical treatment of tetralogy of Fallot

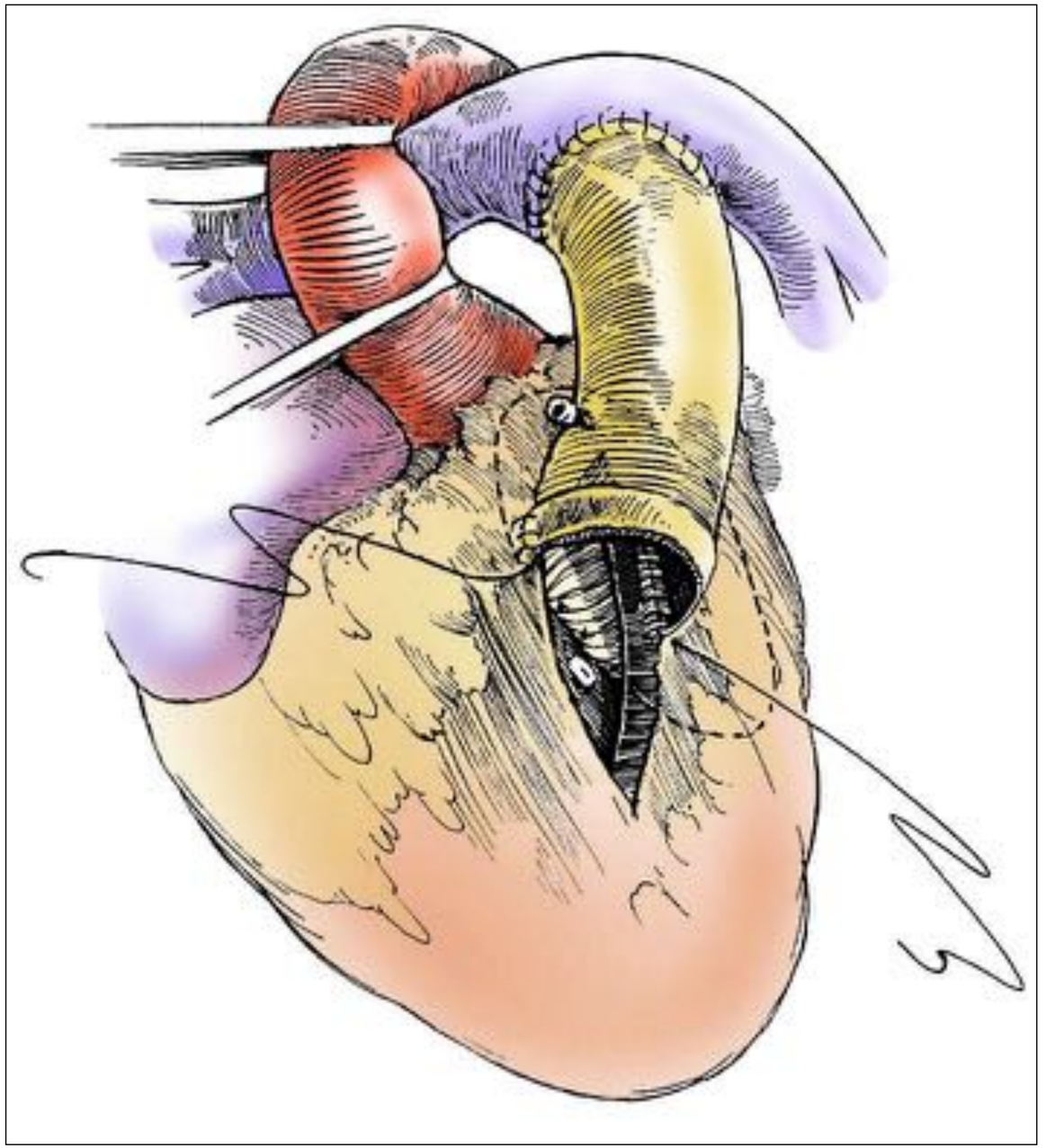
« Monocusp »



Pulmonary mono-cusp

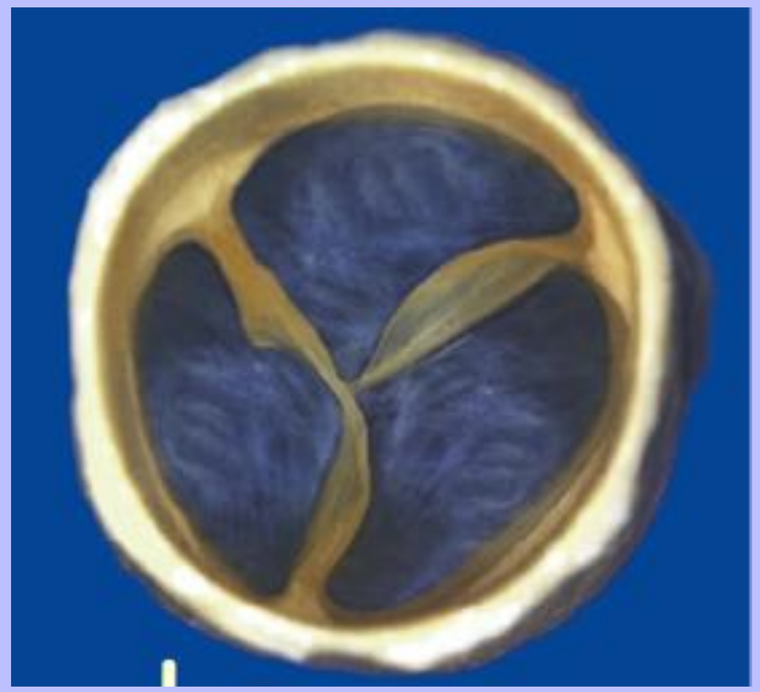
RV-PA conduits : Rastelli type

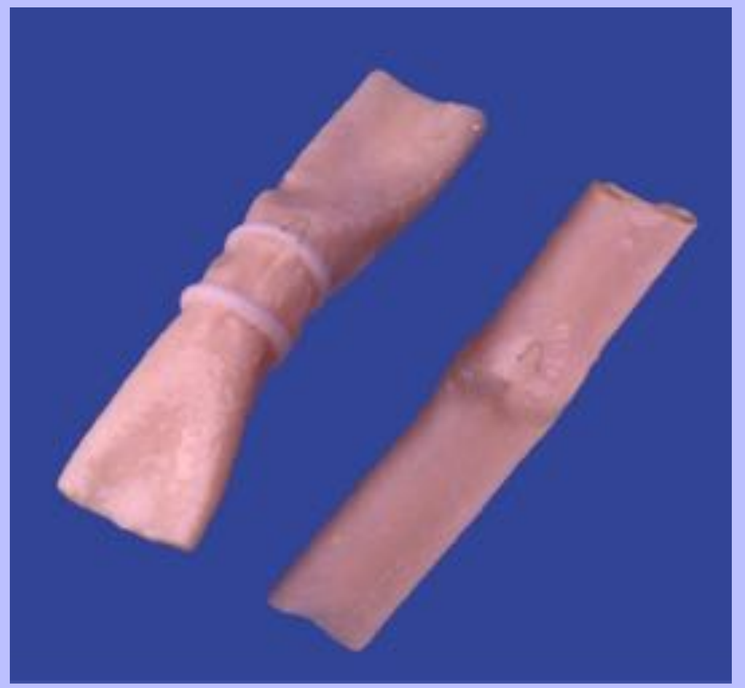




Pulmonary homograft

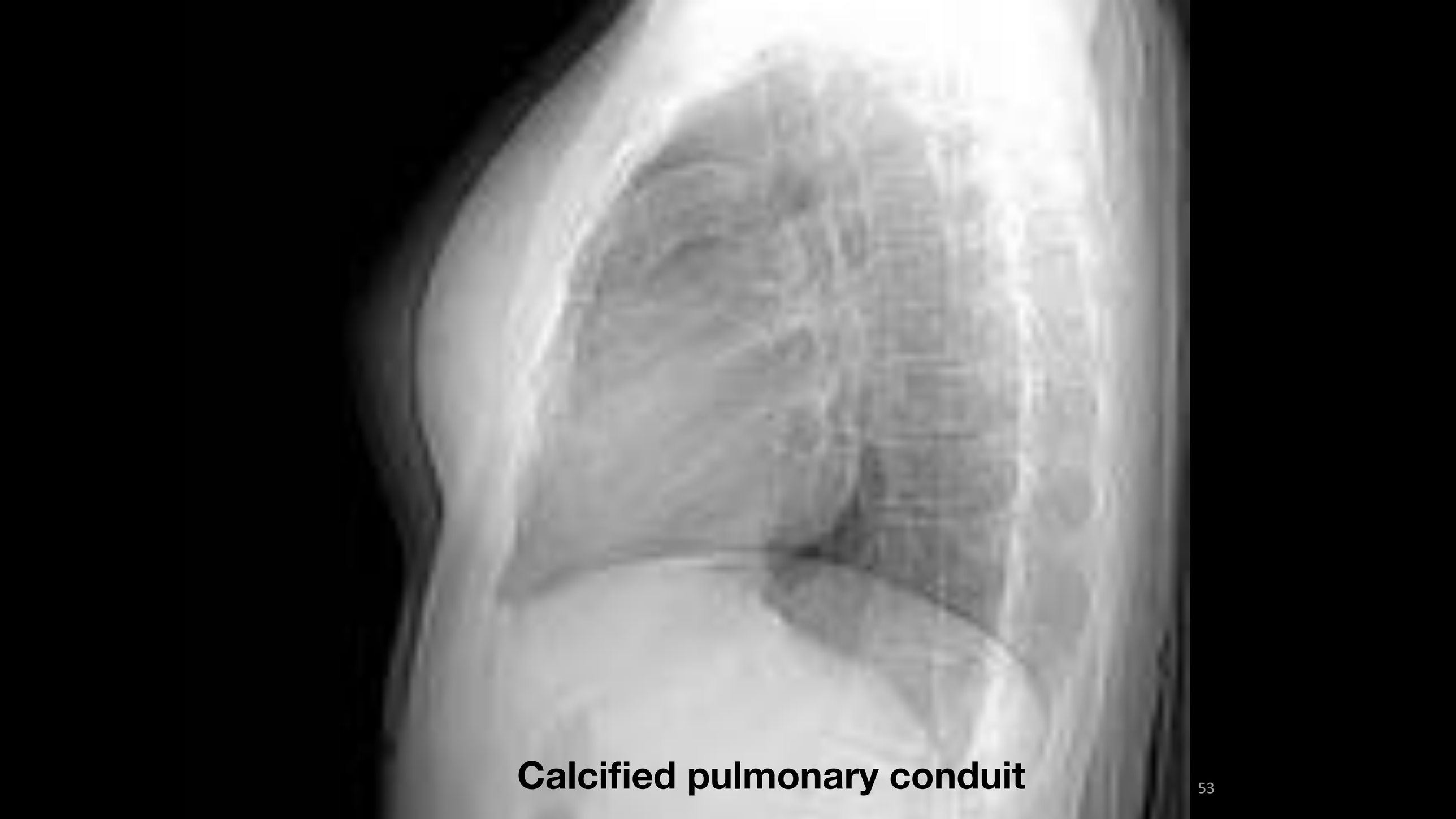
Contegra - Venpro



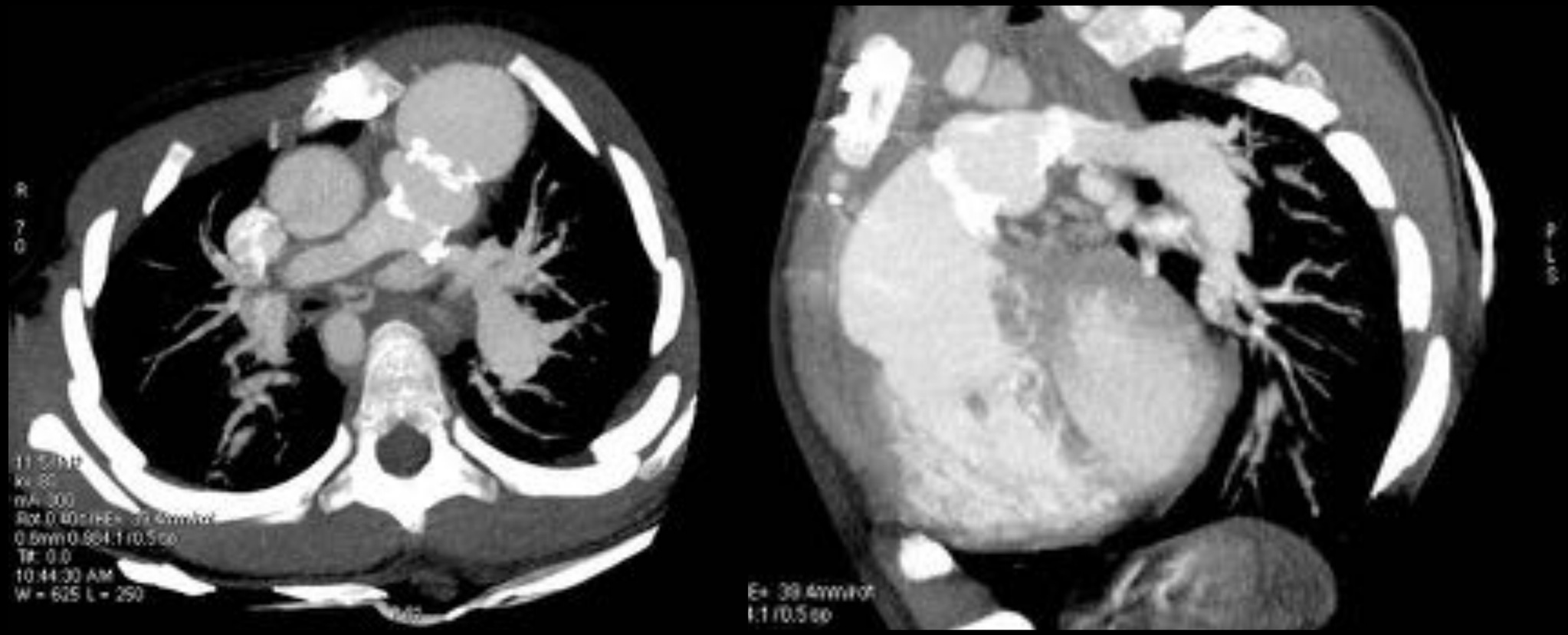


Labcor

VenPro

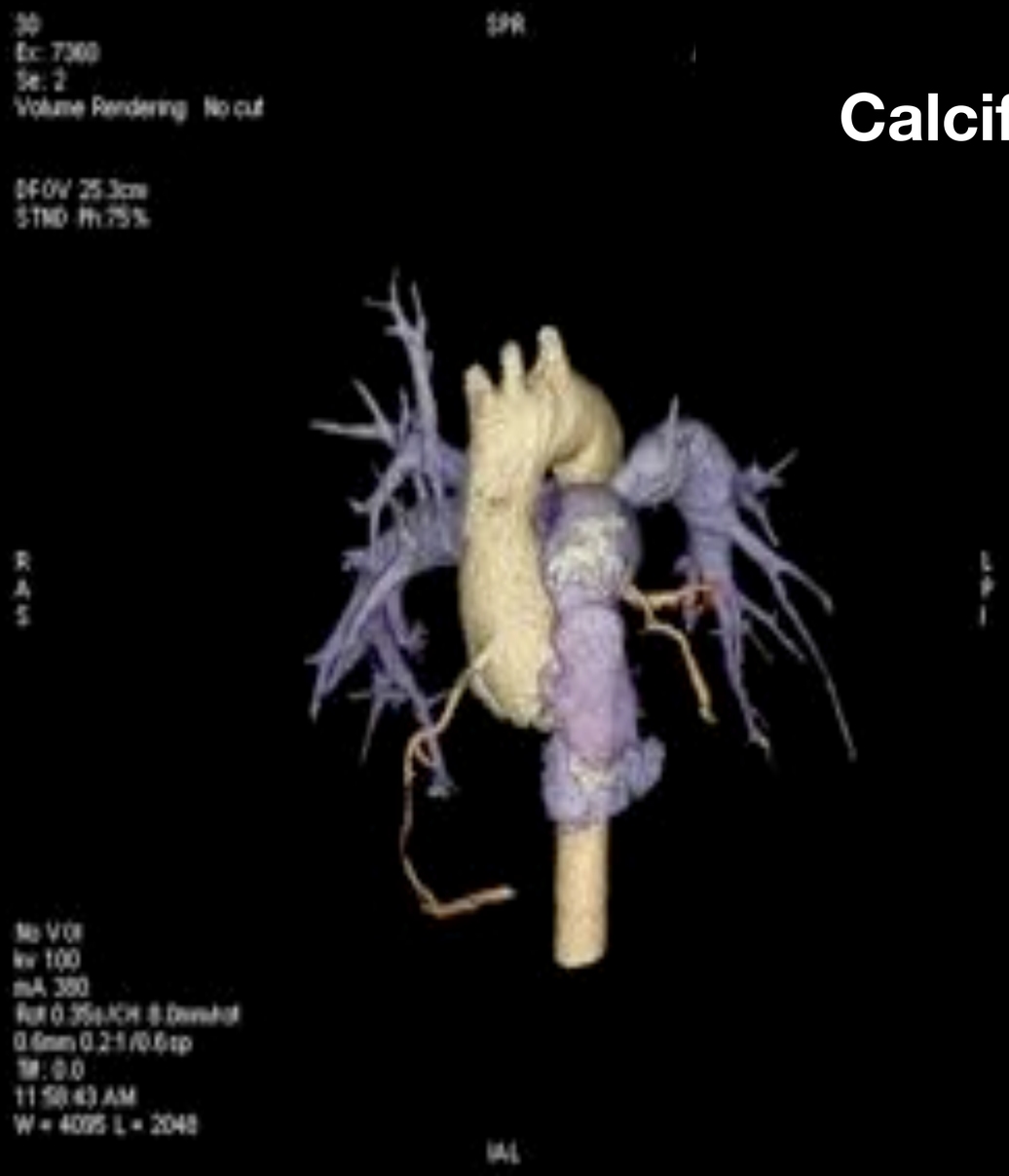


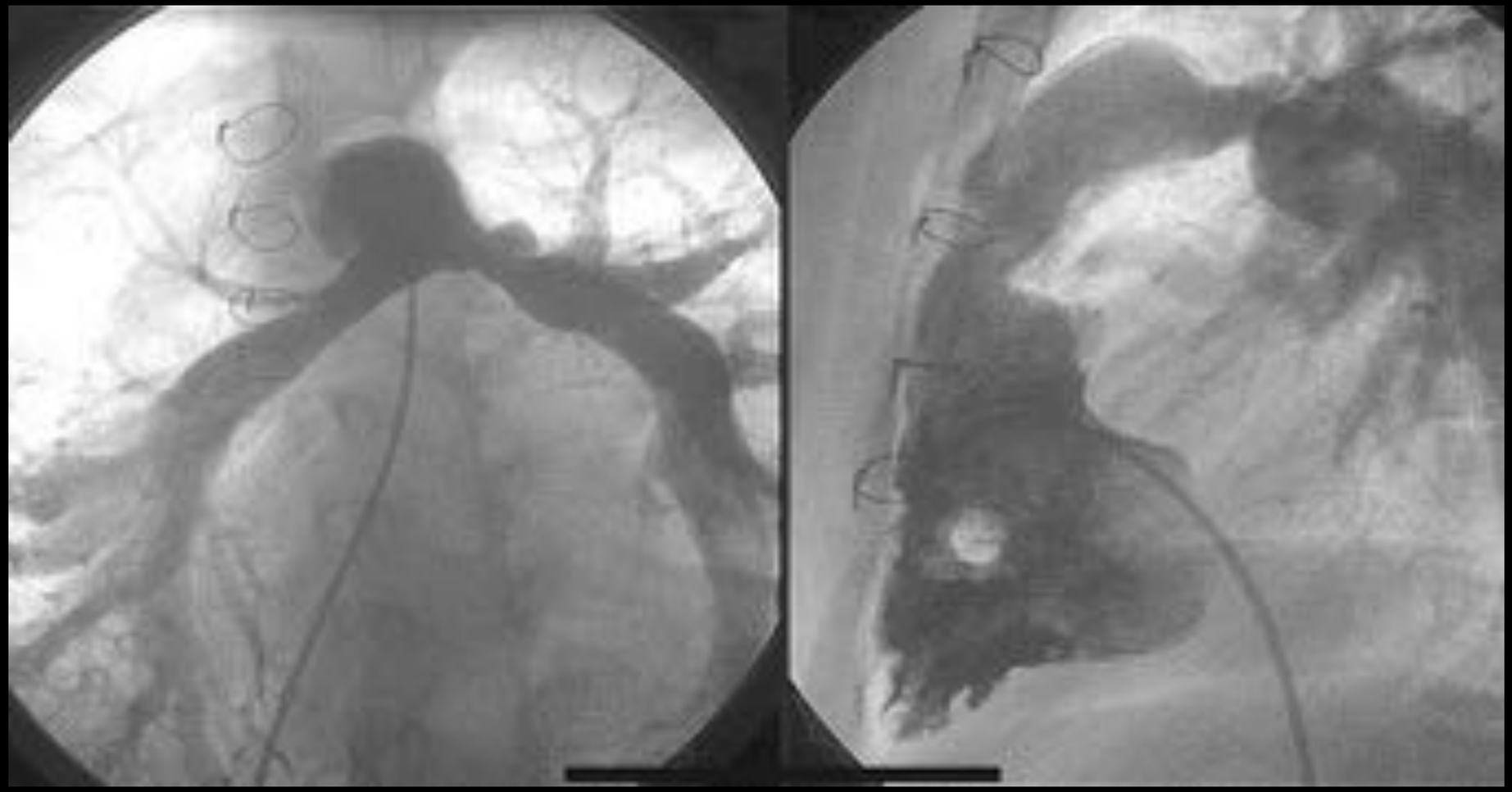
Calcified pulmonary conduit



Calcified pulmonary conduit

Calcified pulmonary conduit

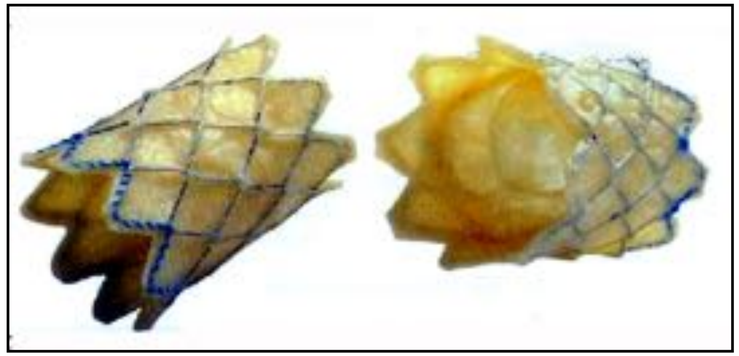




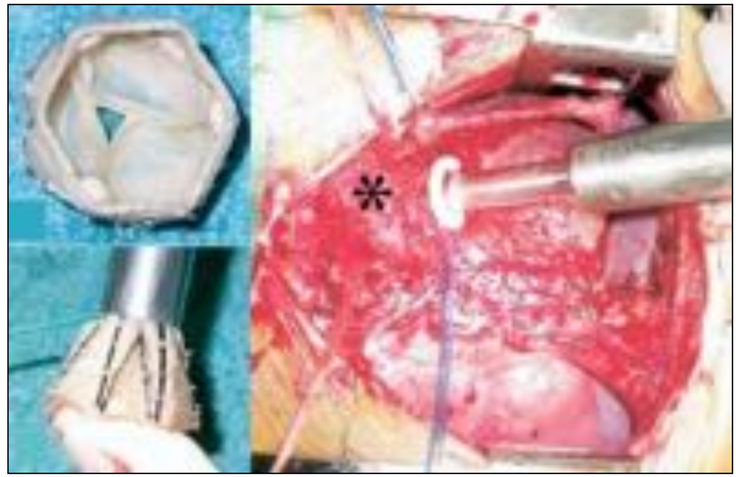
Calcified pulmonary conduit

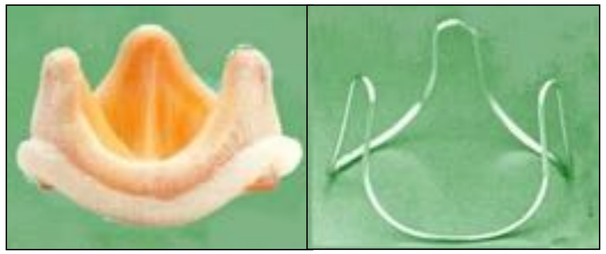
Percutaneous pulmonary valves

«Melody»

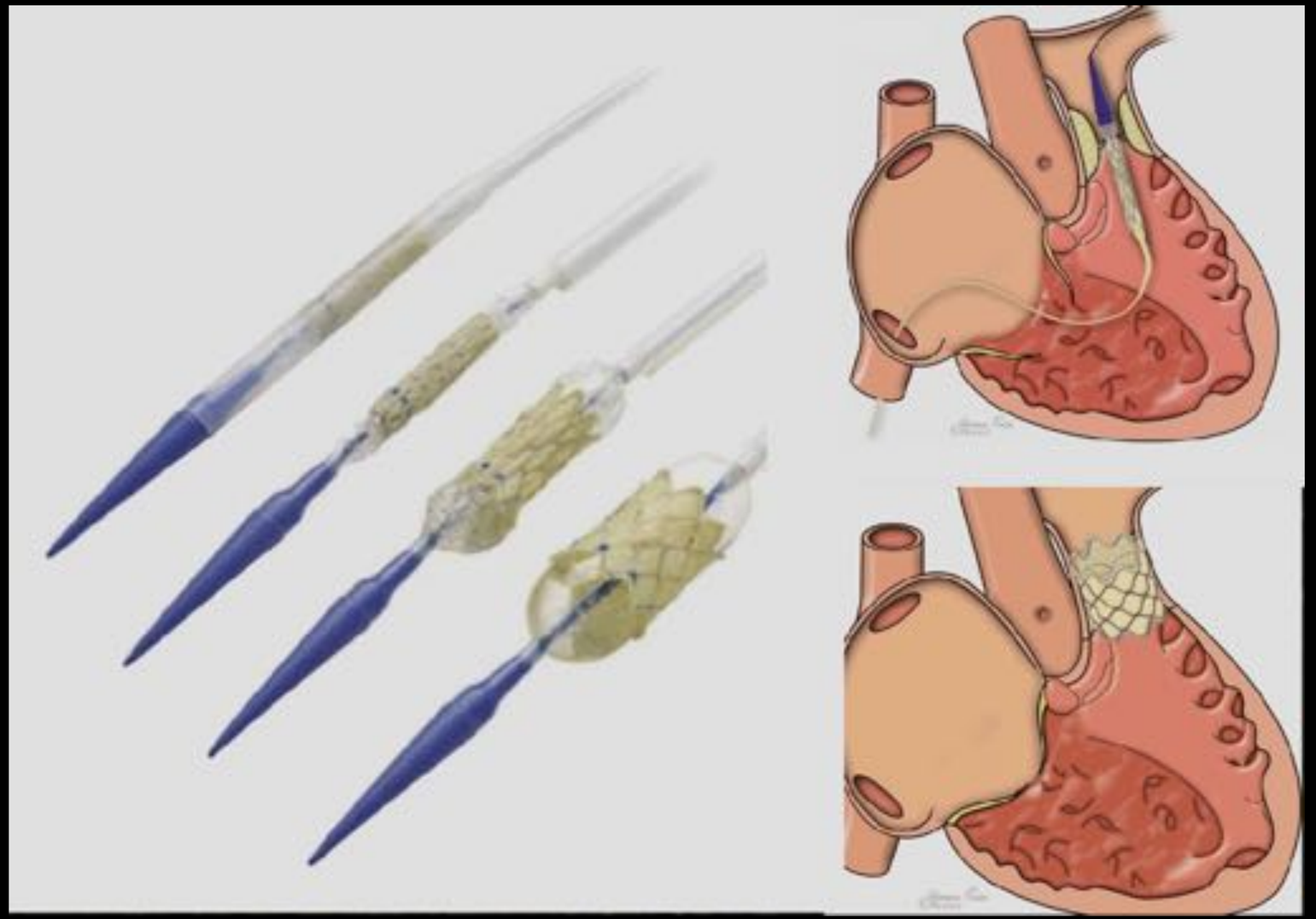


«Shelhigh»





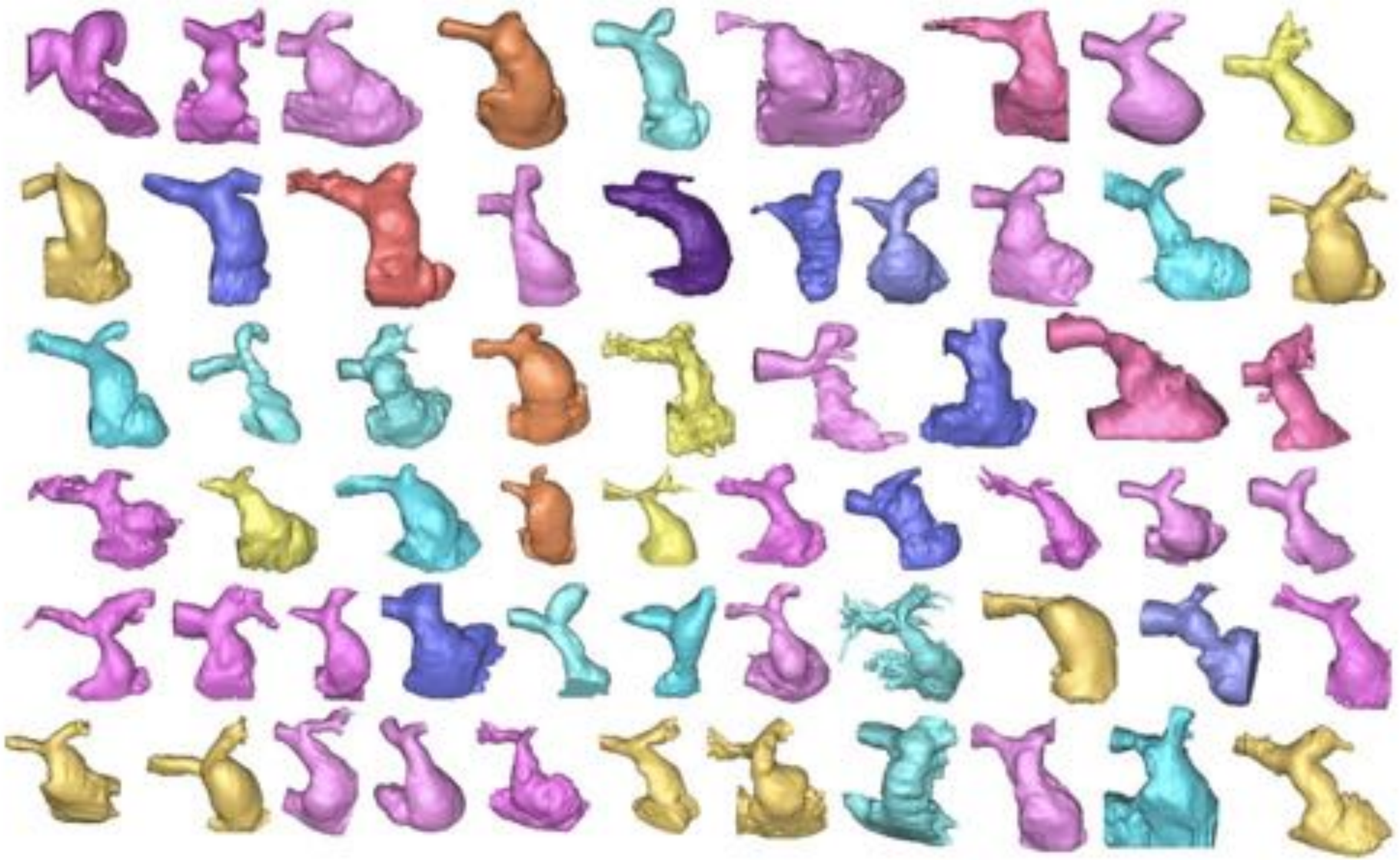
Schreiber. J Thorac Cardiovasc Surg 2006



Melody valve implantation - movie

Melody valve implantation 1

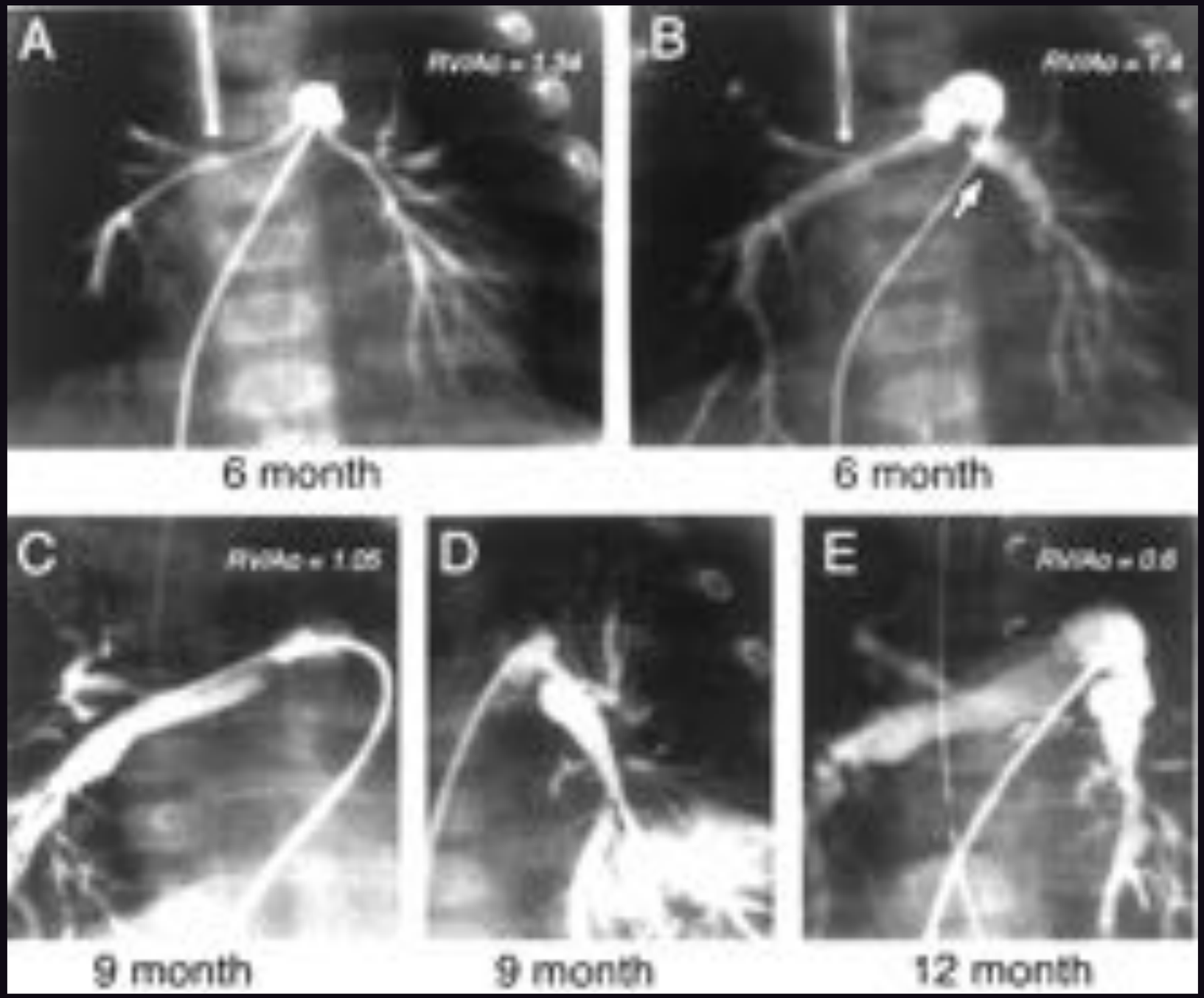
Melody valve implantation 2



Capelli et al. 2011

Pulmonary branch stenosis

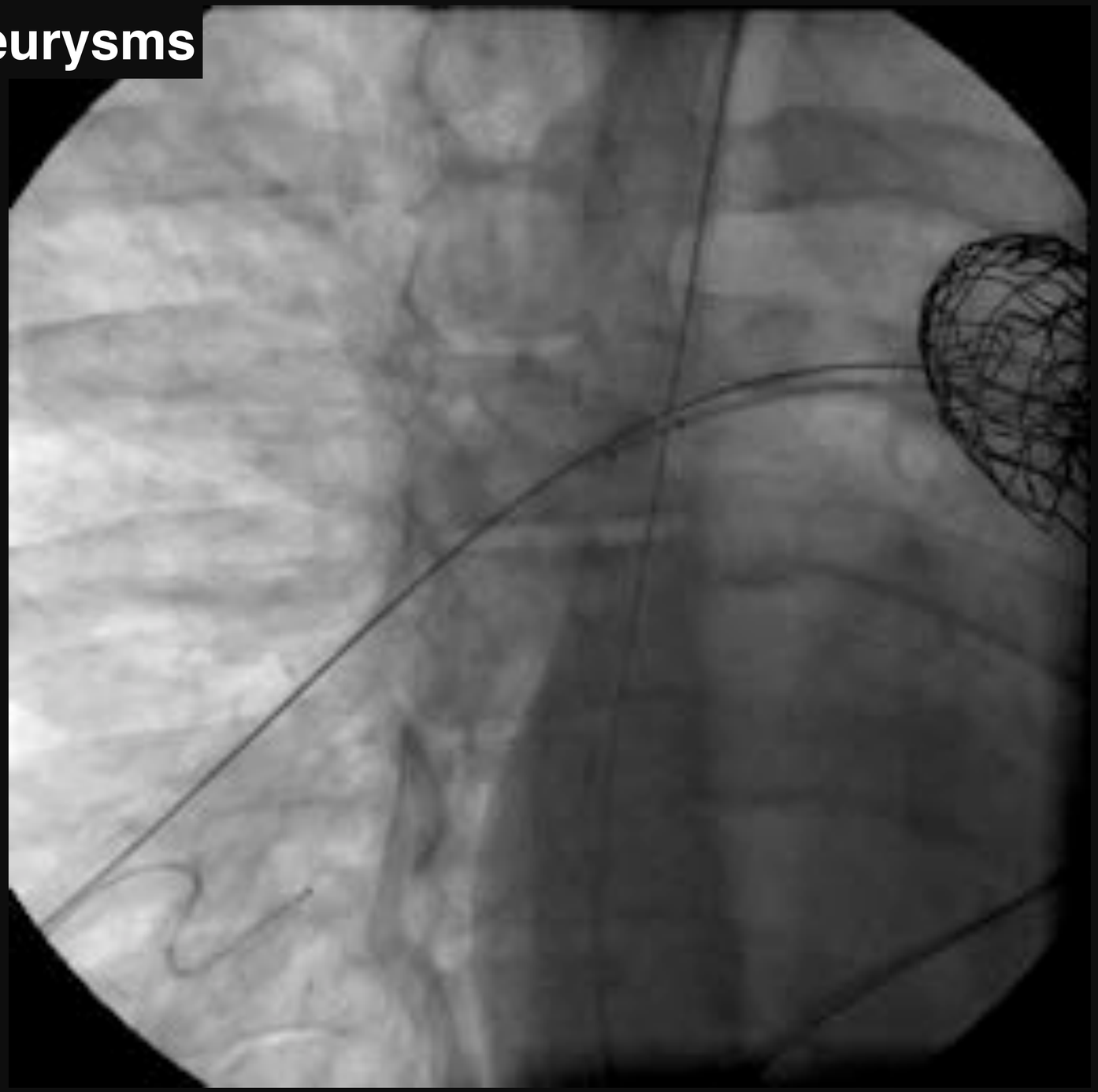
Congenital


A
RVOAo = 1.34
B
RVOAo = 1.4
6 month
6 month
C
RVOAo = 1.05
D
E
RVOAo = 0.6
9 month
9 month
12 month

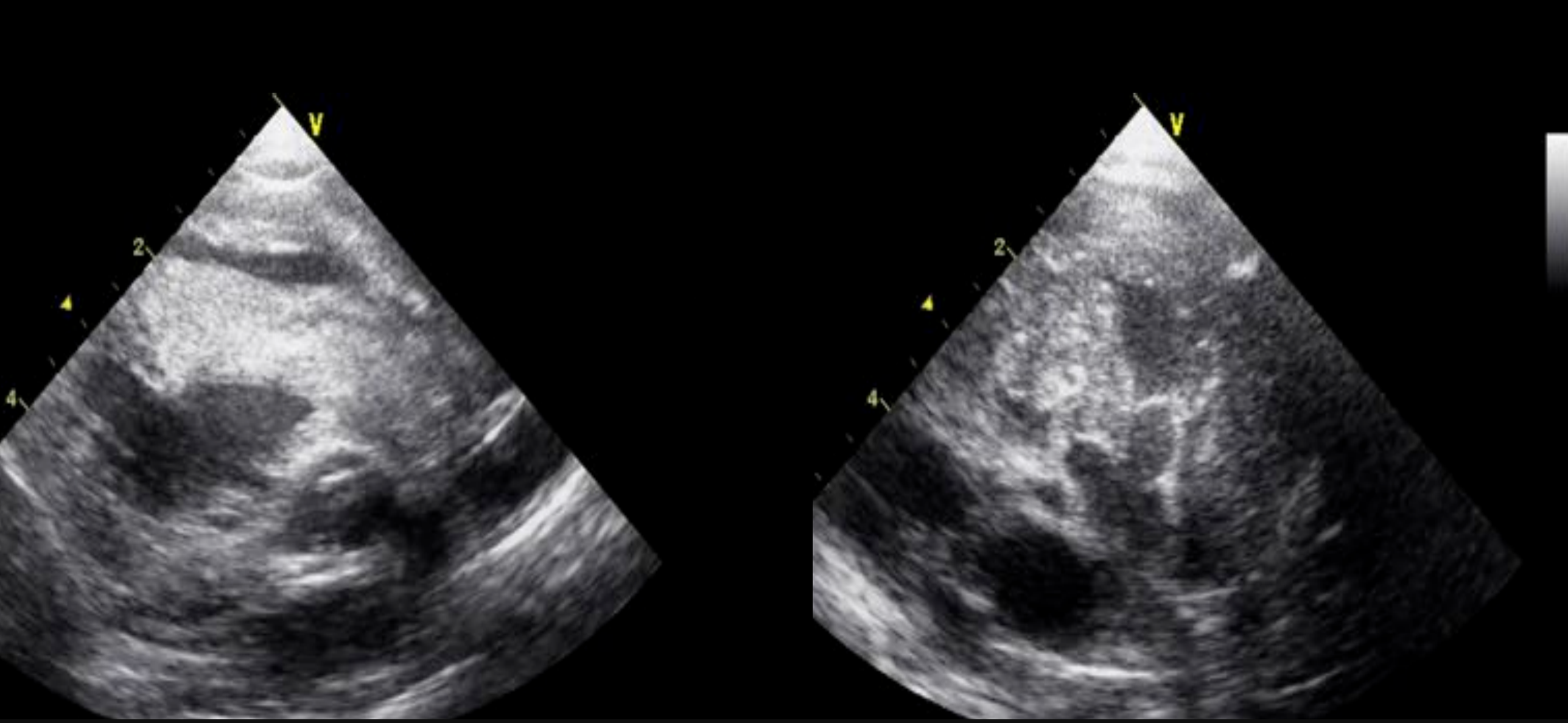
Acquired

Multiple and diffuse

Diffuse with aneurysms



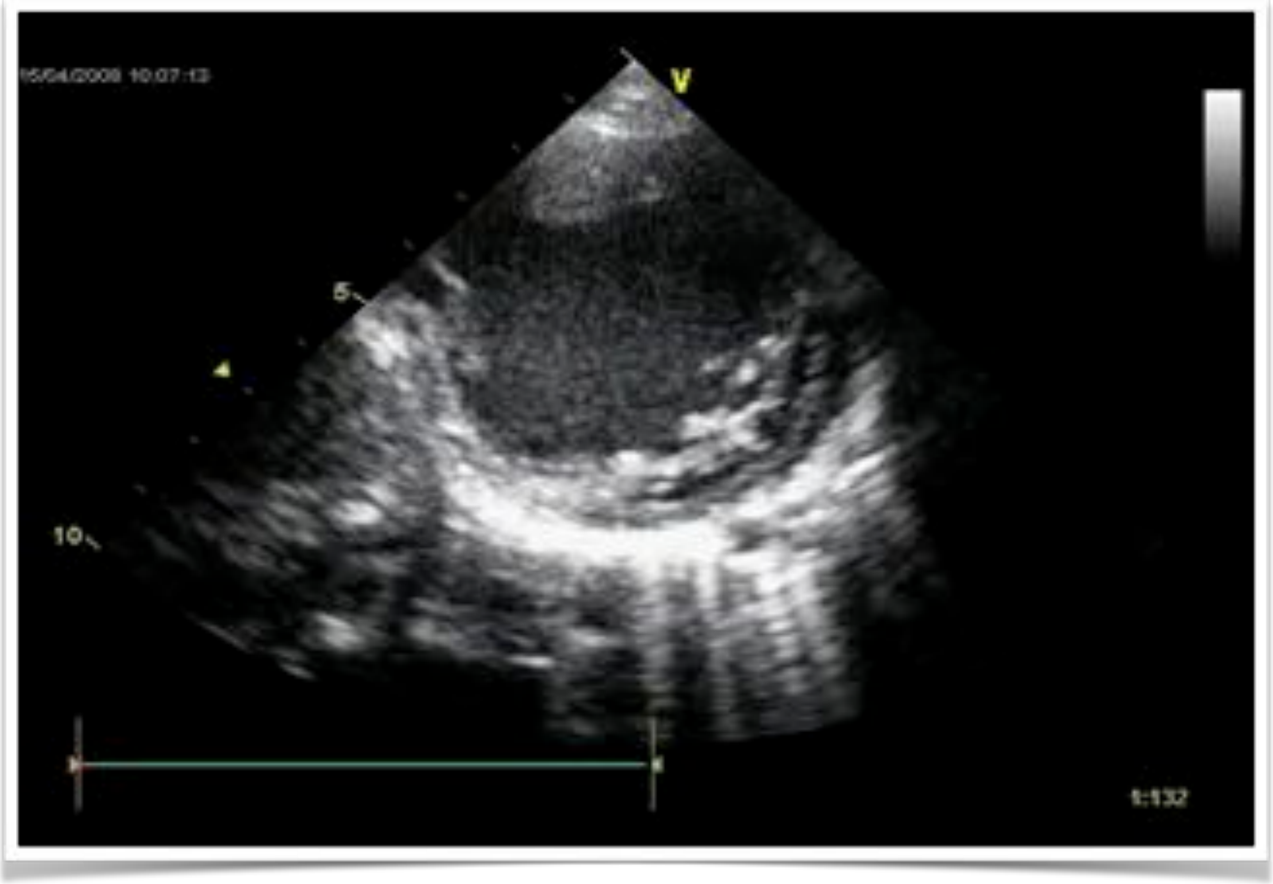
Unusual right obstructions

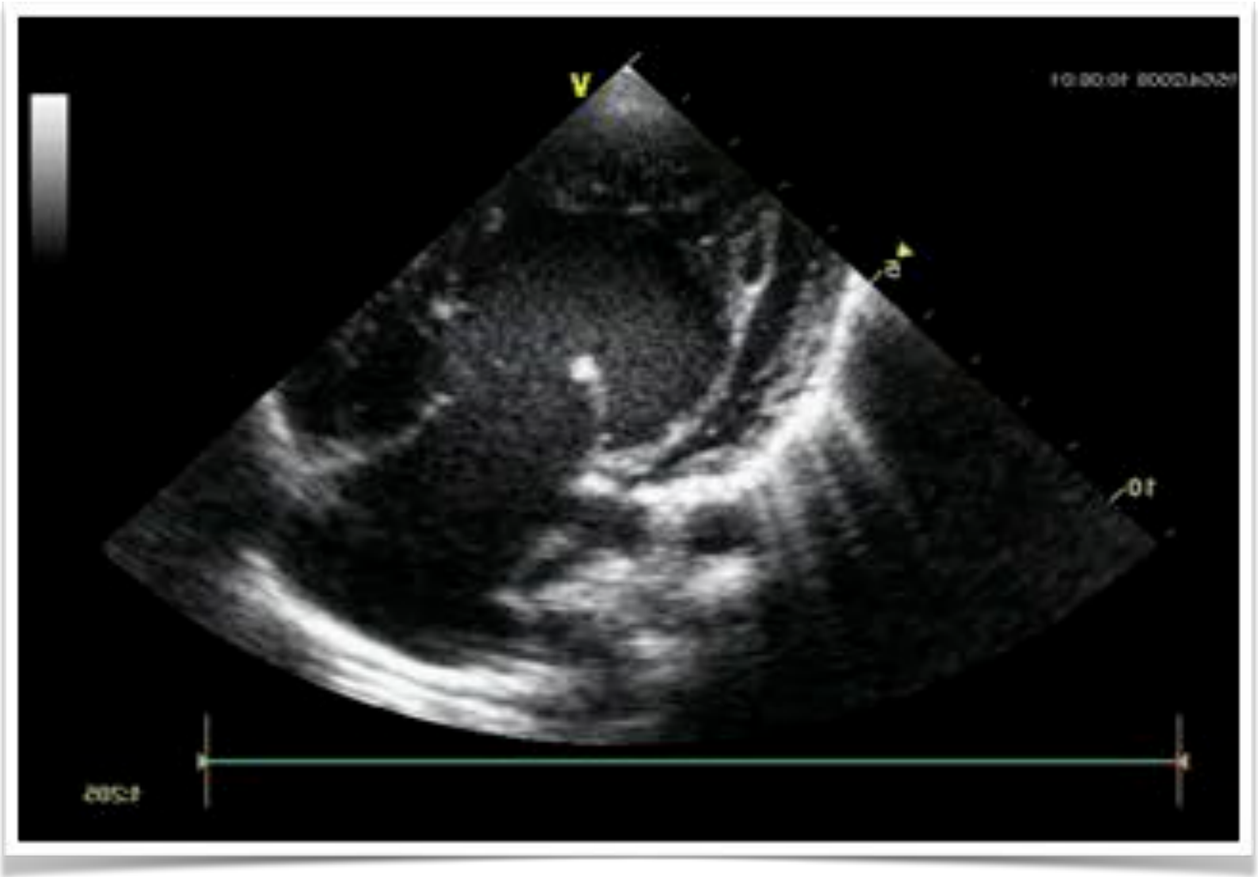


Right ventricular teratoma

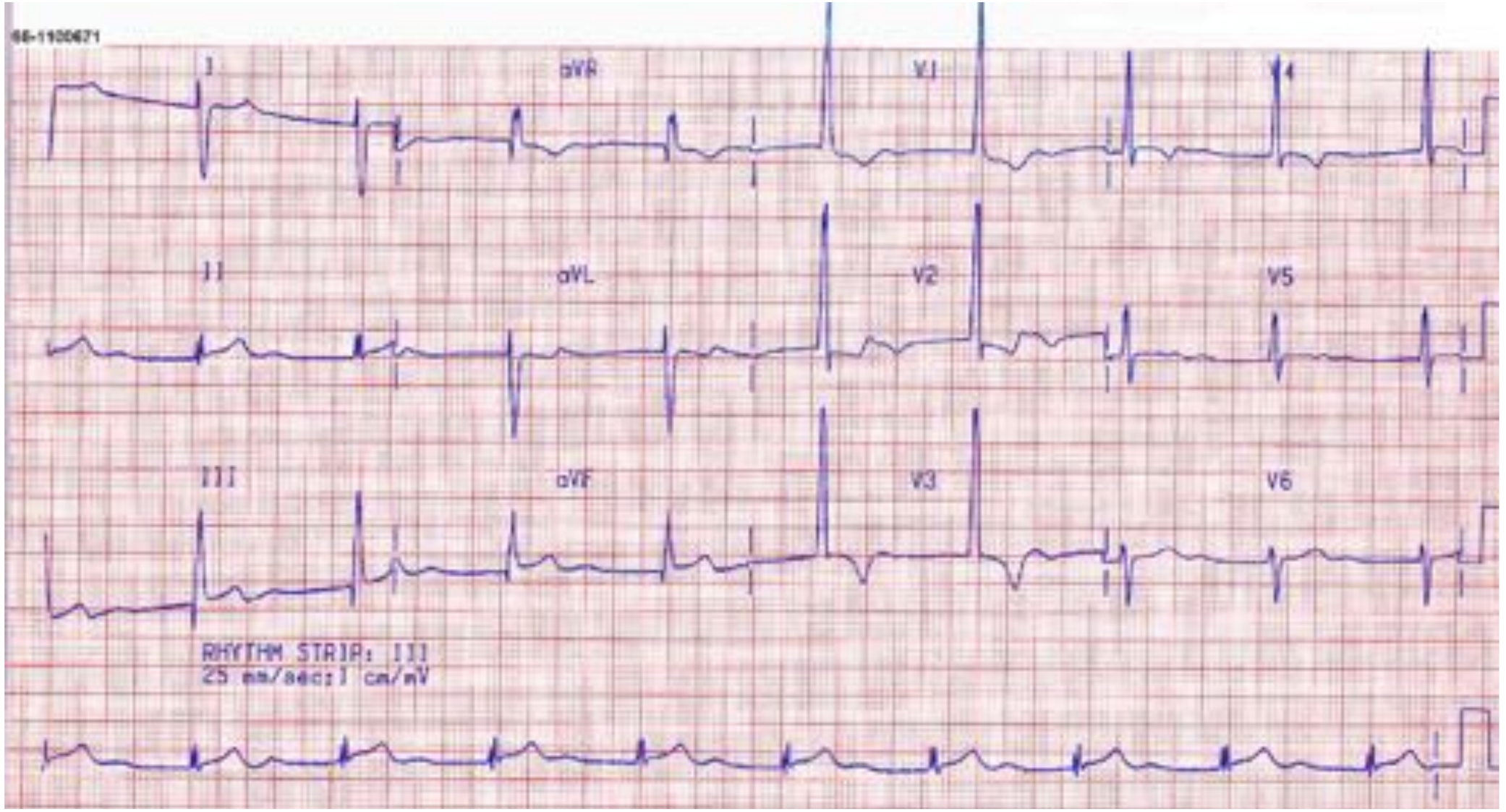
Caval and right atrial extension of a nephroblastoma

Une femme de 33 ans consulte pour des douleurs abdominales à type de pesanteur à l'effort. Elle a été opérée dans l'enfance d'une cardiopathie par sternotomie mais ne se souvient plus laquelle. Elle n'est plus suivie depuis l'âge de 16 ans.

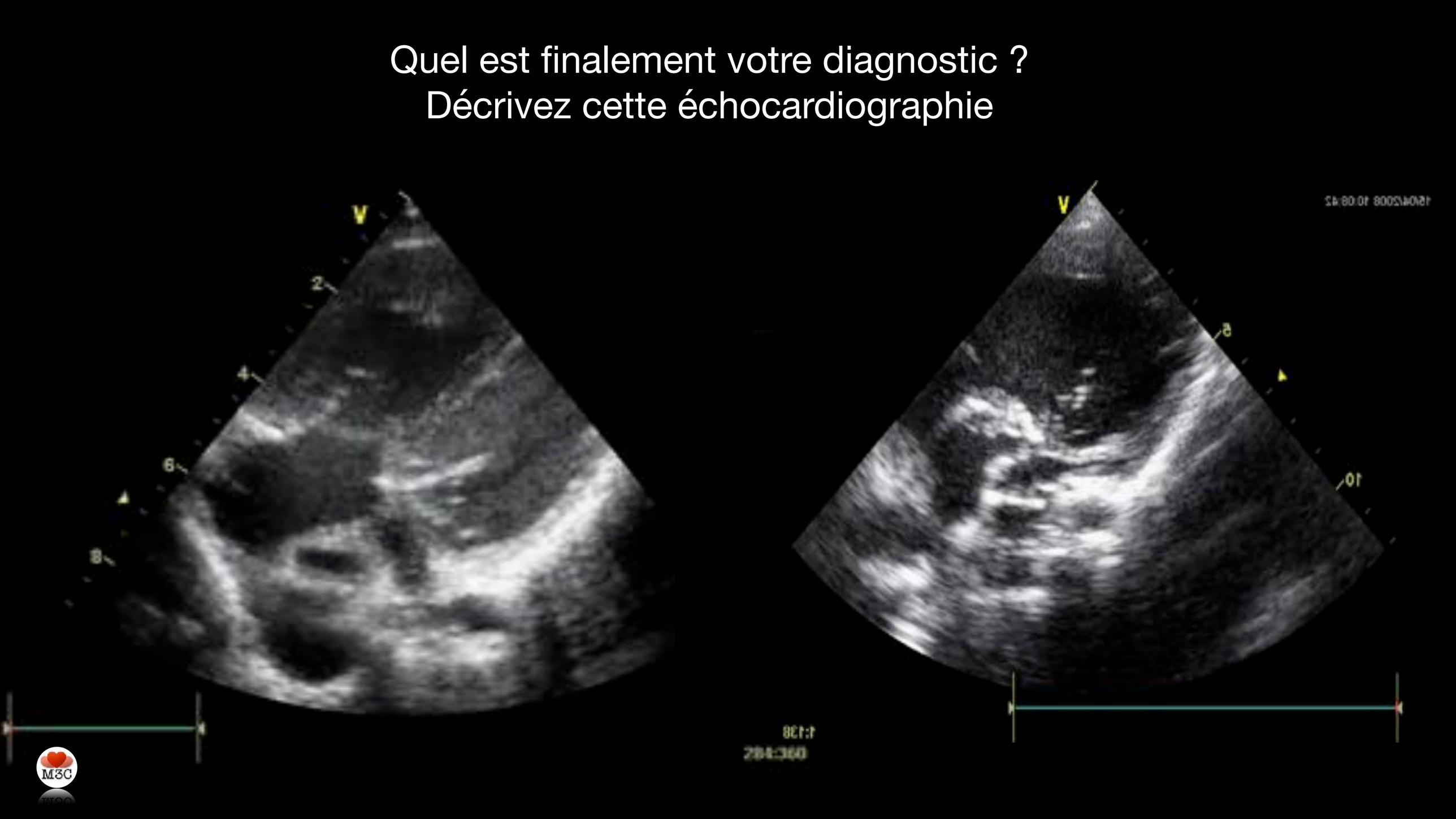




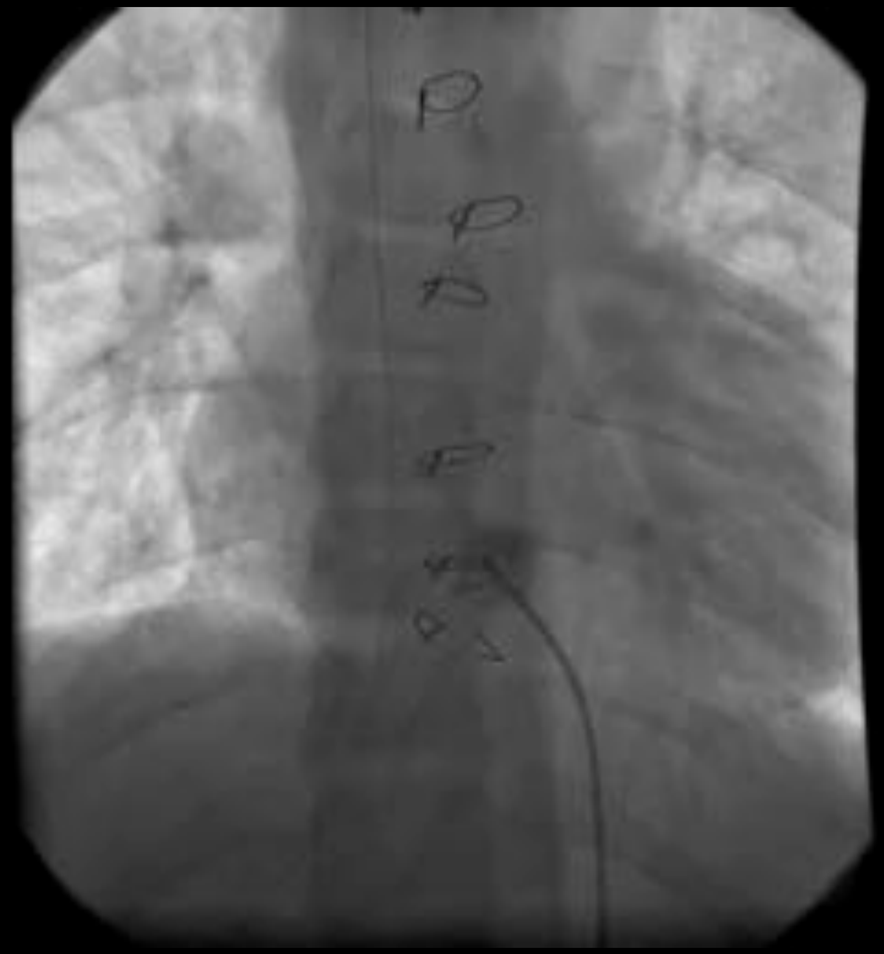
Faites vous ou confirmez vous votre diagnostic sur cet ECG ?



Quel est finalement votre diagnostic ?
Décrivez cette échocardiographie

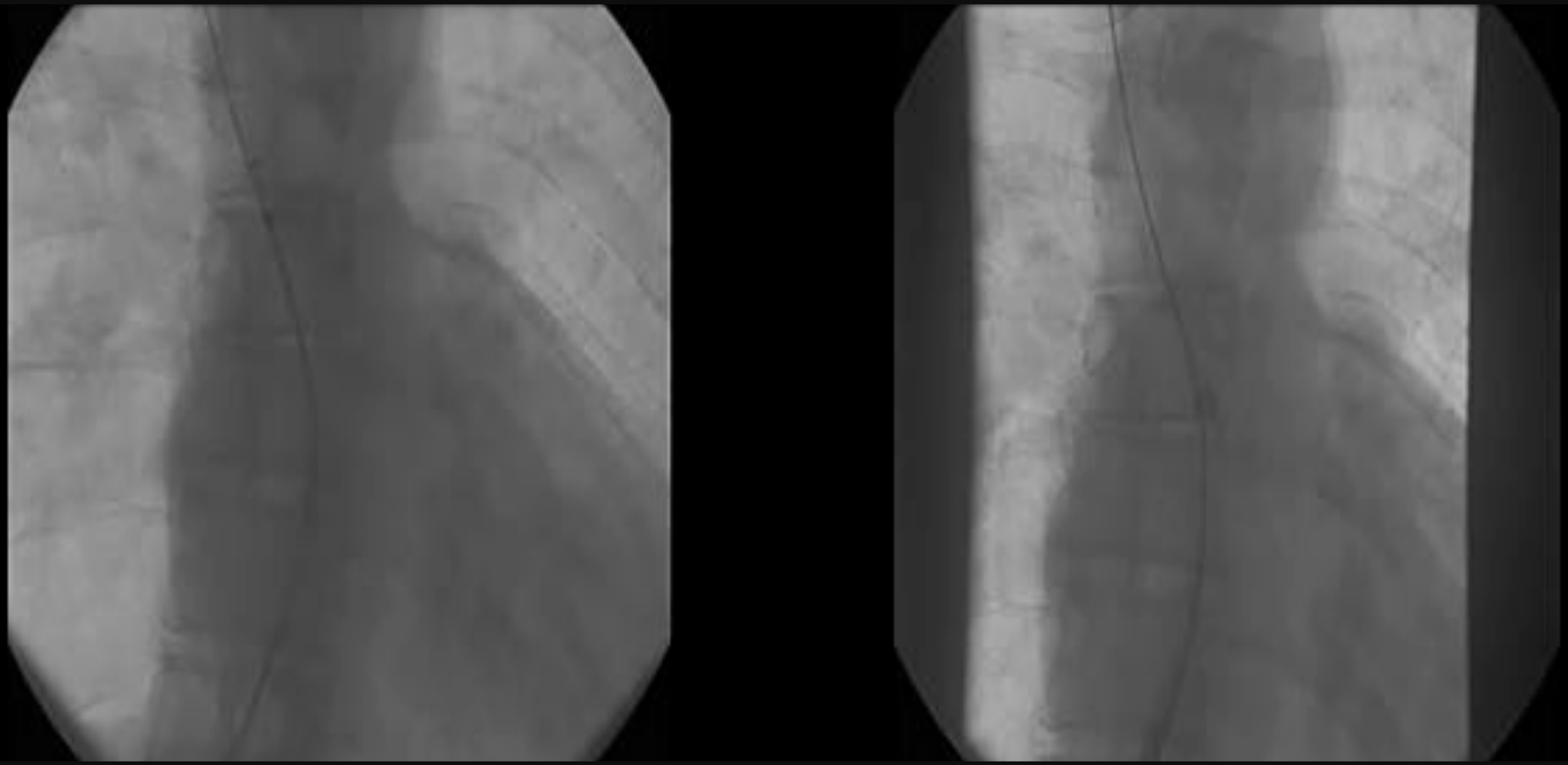


Vous faites un cathétérisme cardiaque car vous ne comprenez pas les symptômes de la patiente chez qui vous avez trouvé une lame d'ascite et une hépatomégalie d'allure cardiaque. Interprétez ces angiographies



Caval path stenosis Mustard

Right Superior Caval Vein stenosis



Superior caval vein stenosis after Senning operation


M3C
Merci