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

Damien Bonnet

Unité médico-chirurgicale de Cardiologie Congénitale et Pédiatrique
Hôpital Universitaire Necker Enfants malades – APHP, Université Paris Descartes, Sorbonne Paris Cité
IcarP Cardiology, Institut Hospitalo-Universitaire IMAGINE

Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

M3C

ARCFA

ASSISTANCE PUBLIQUE HÔPITAUX DE PARIS
NECKER-ENFANTS MALADES

FONDATION imagine
INSTITUT DES MALADIES GÉNÉTIQUES

Necker ENFANTS MALADES HÔPITAL UNIVERSITAIRE

UNIVERSITÉ PARIS DESCARTES

HÔPITAUX UNIVERSITAIRES PARIS OUEST
Corentin-Celton
Hôpital européen Georges-Pompidou
Vaugirard - Gabriel-Pallez

Inserm

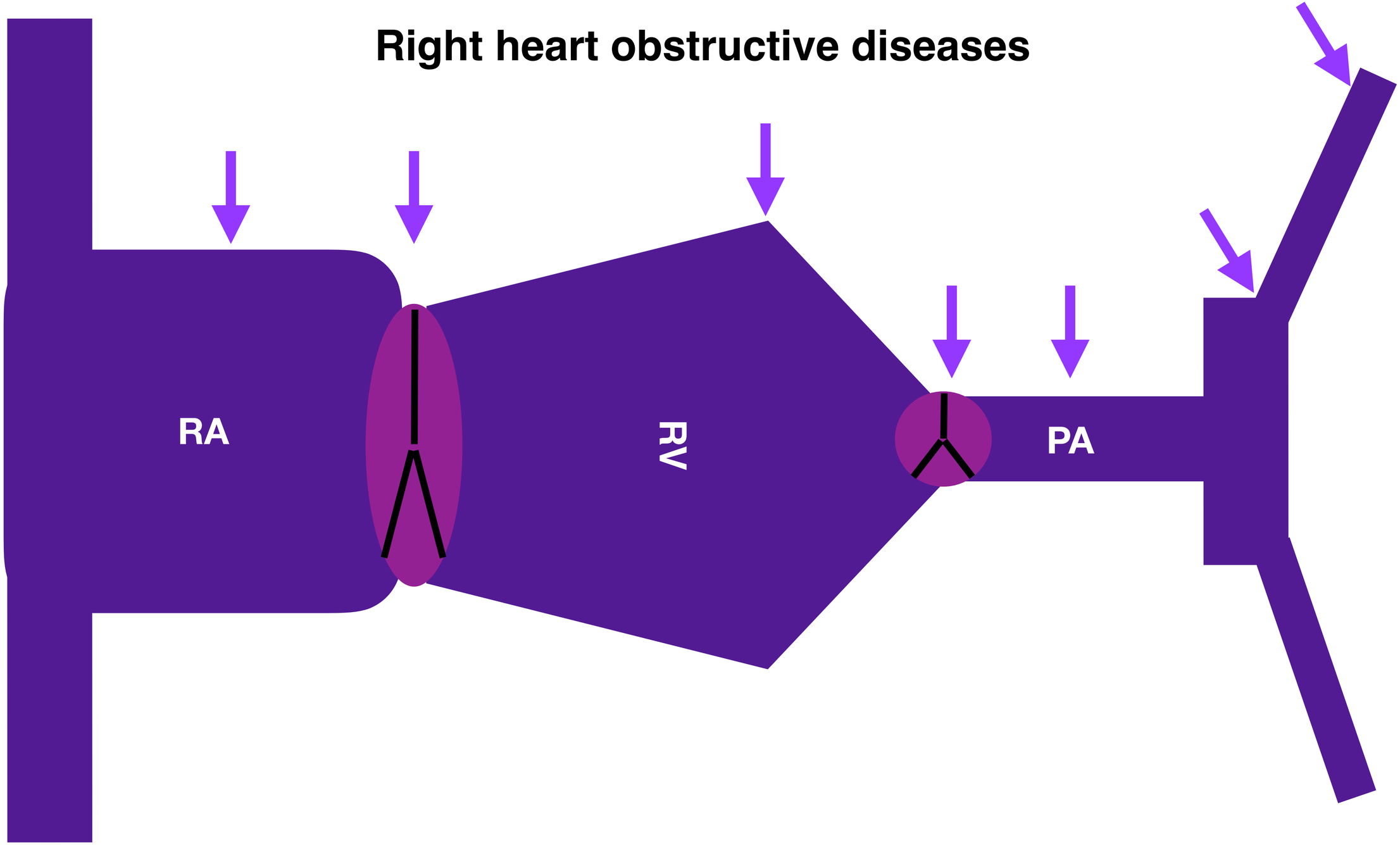
cardiogen
Filière nationale de santé maladies cardiaques héréditaires

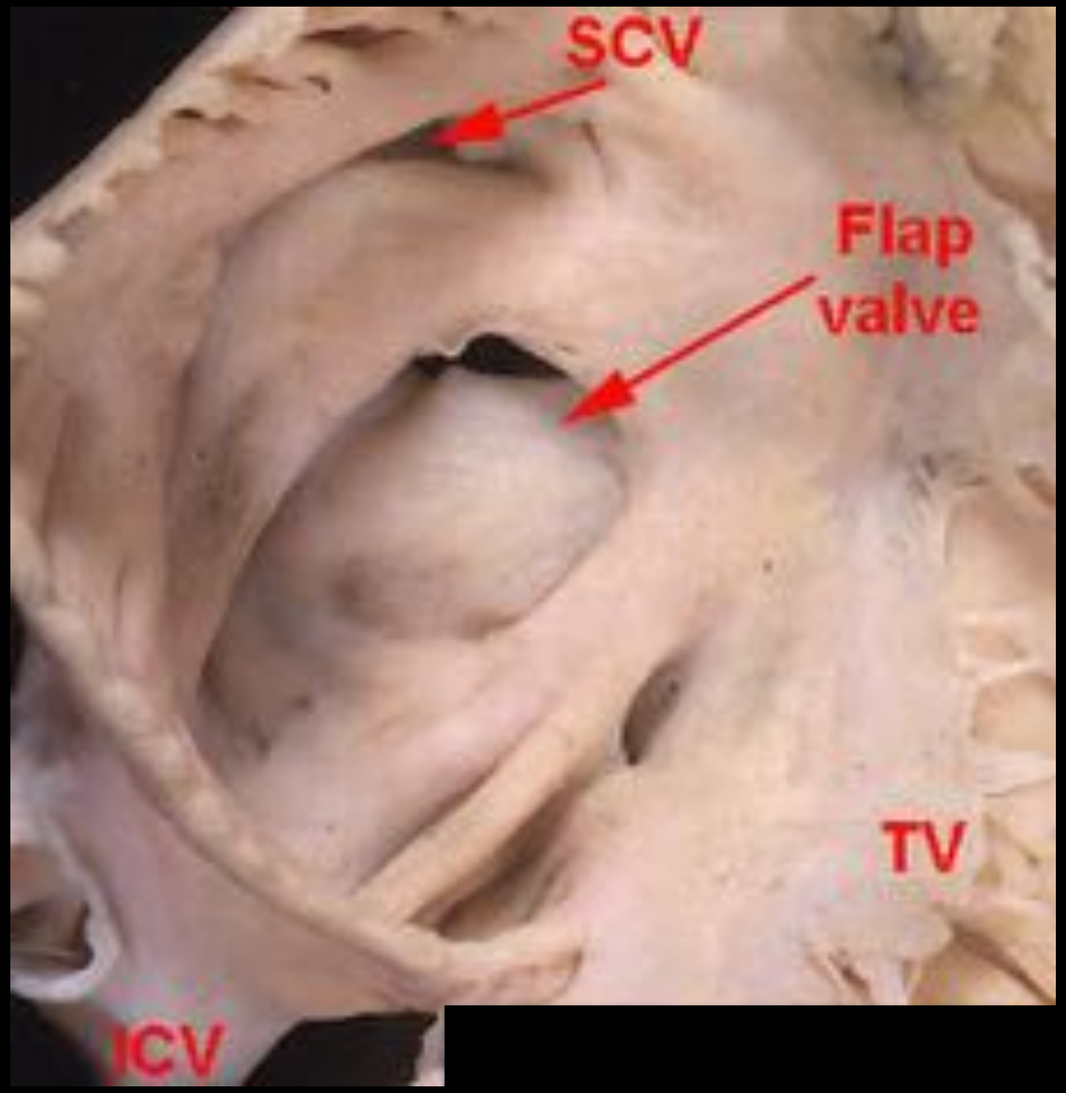
Centre de référence des Maladies Cardiaques Héréditaires

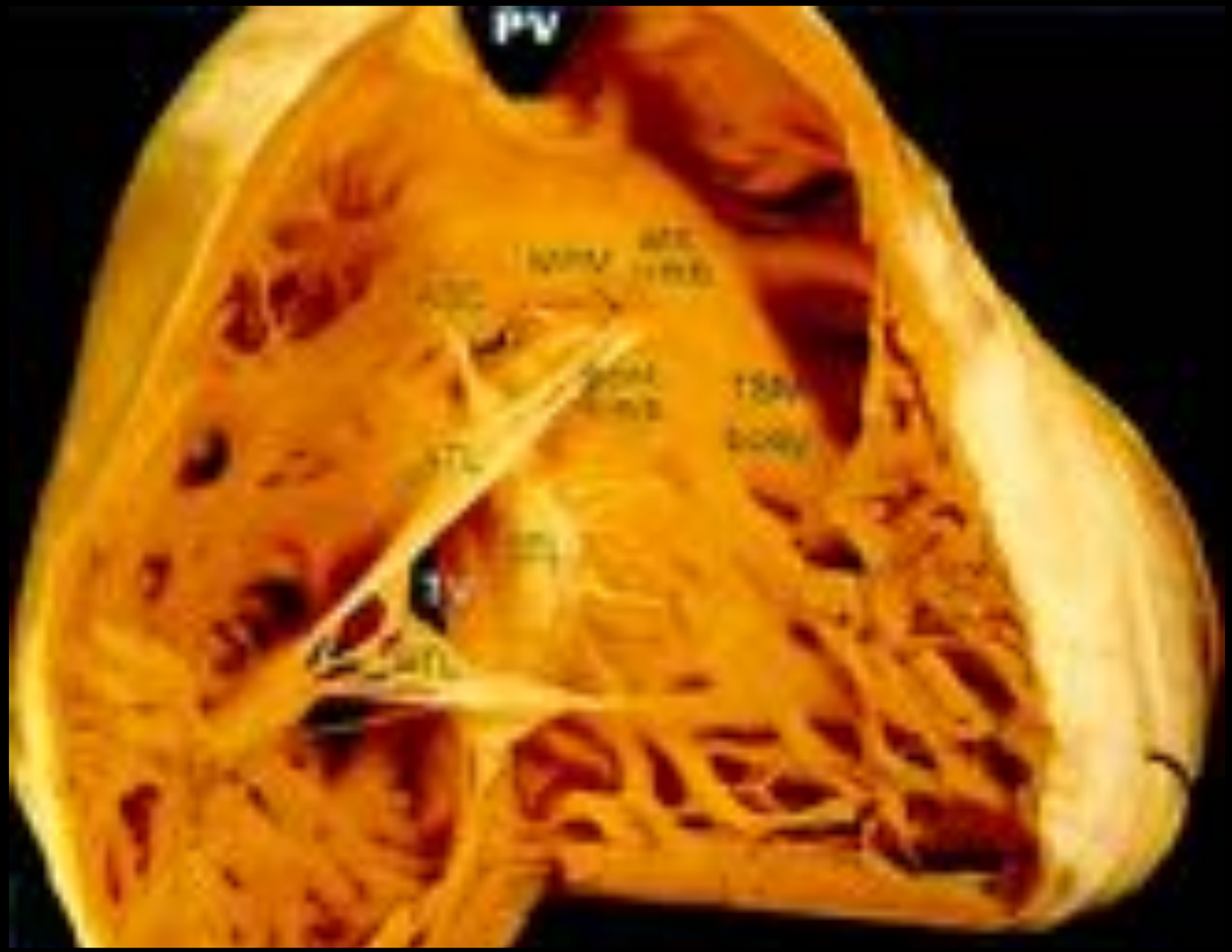
European Reference Network
for rare or low prevalence complex diseases
Network Respiratory Diseases (ERN-LUNG)

European Reference Network
for rare or low prevalence complex diseases
Network Heart Diseases (ERN GUARD-HEART)

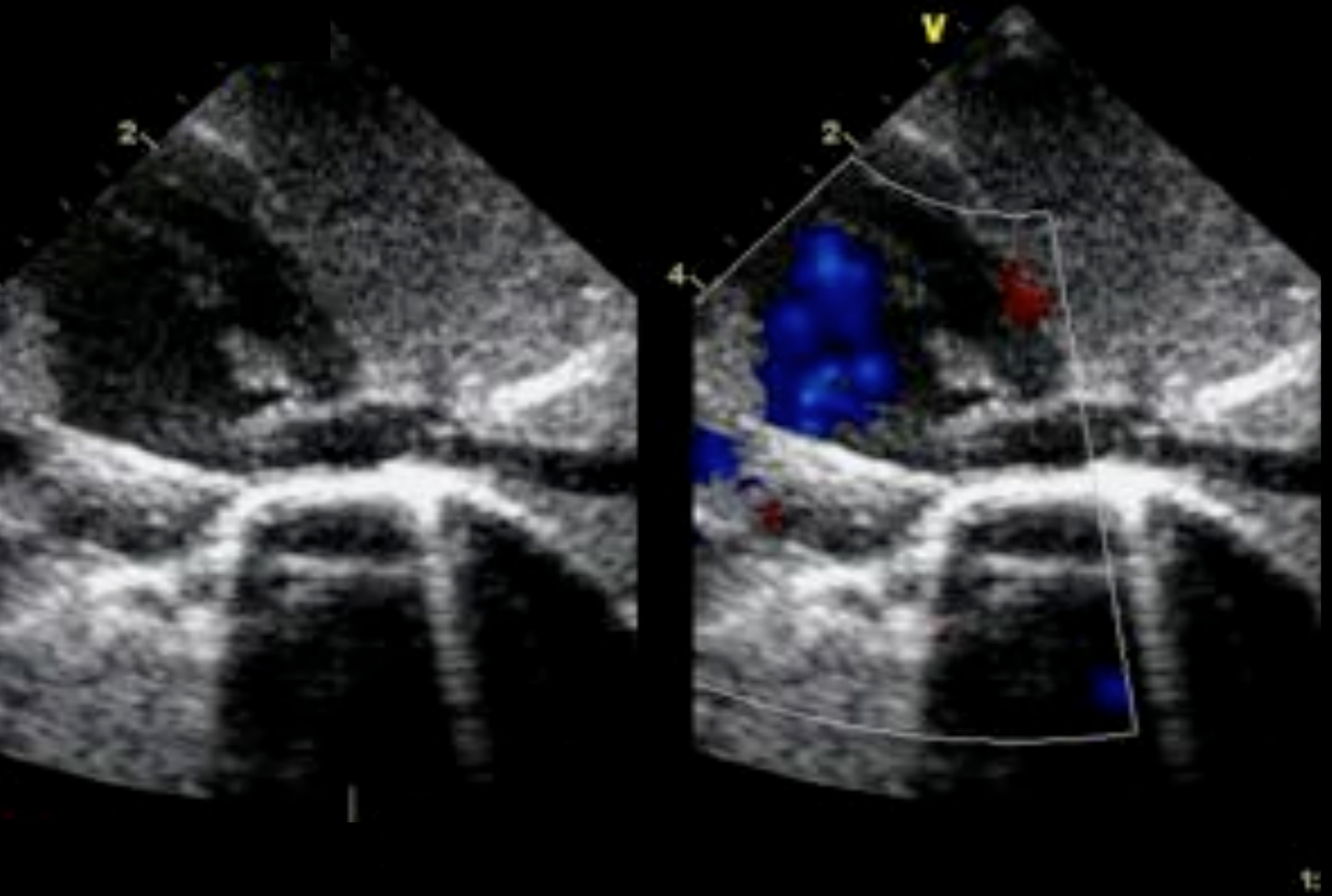
Right heart obstructive diseases



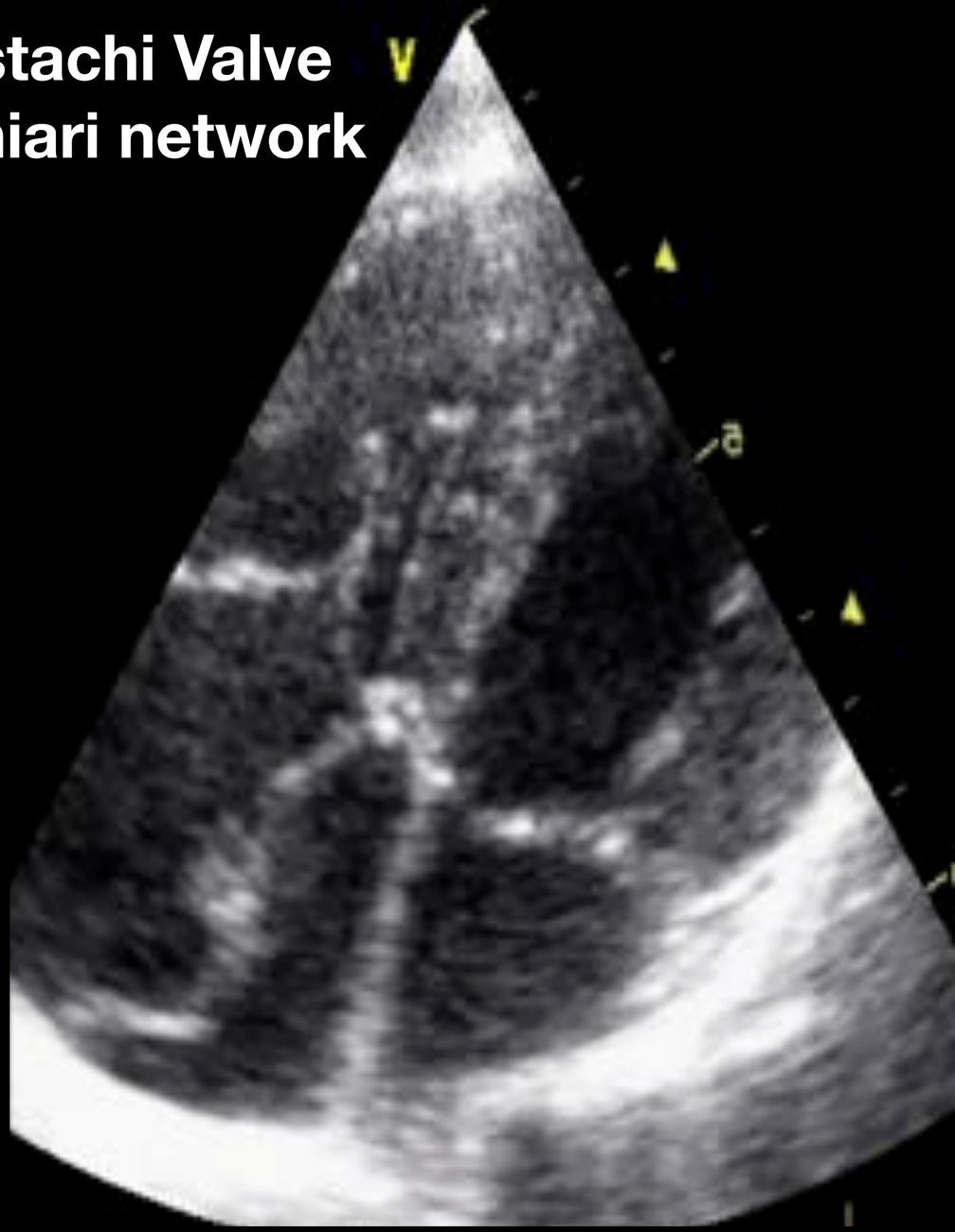

SCV
Flap
valve
TV
ICV


PV

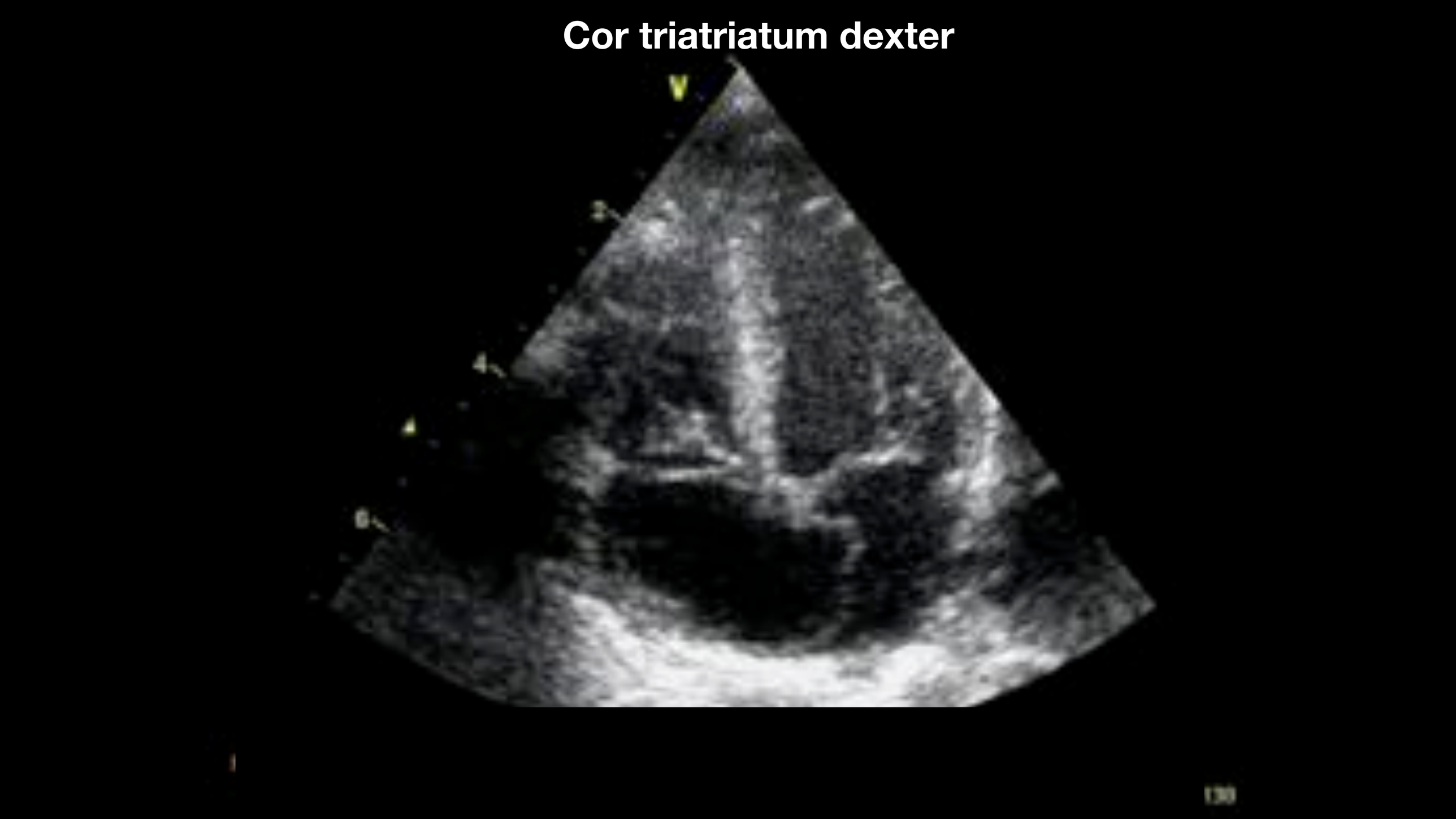
Eustachi Valve

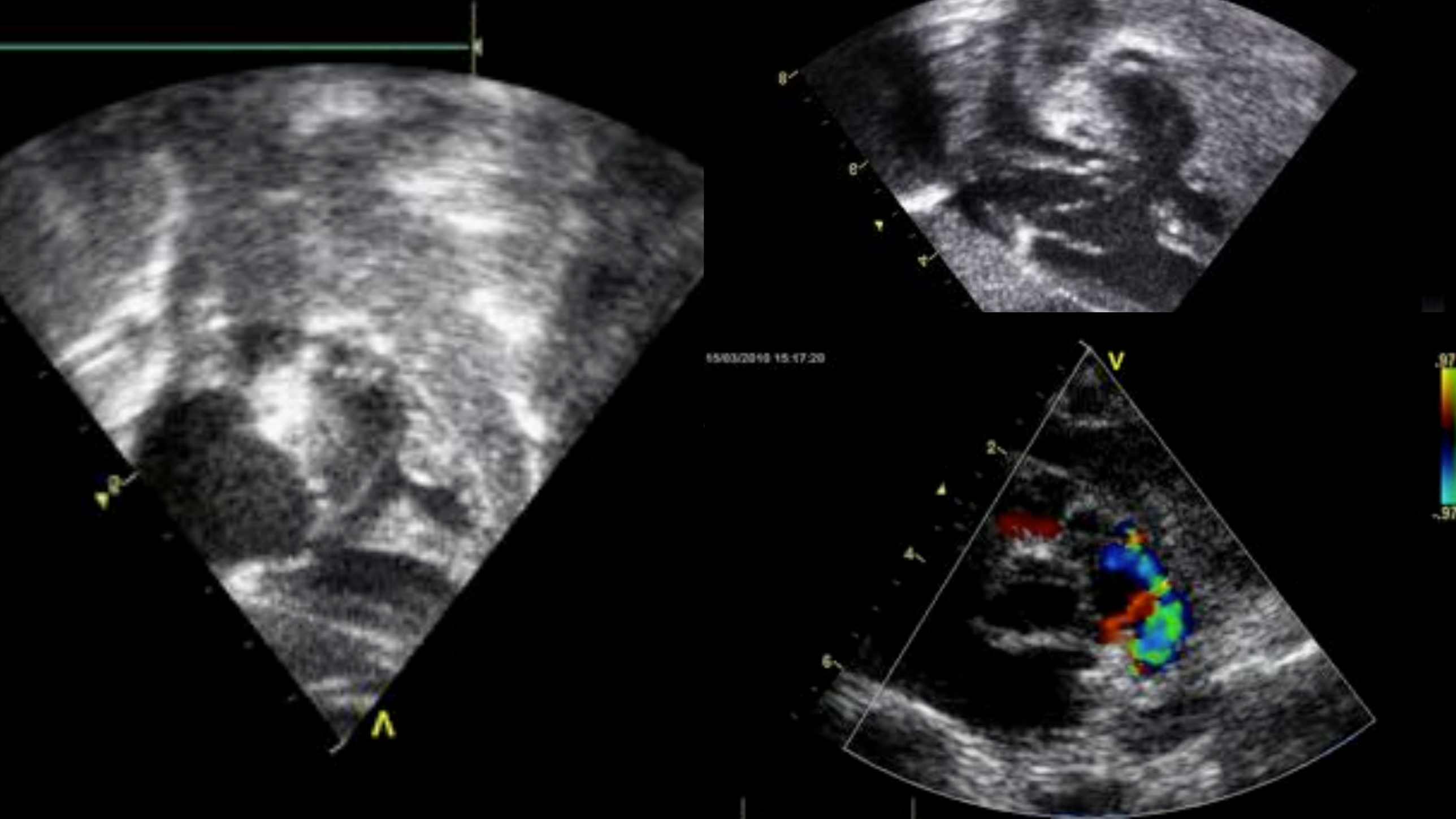


Eustachi Valve & Chiari network



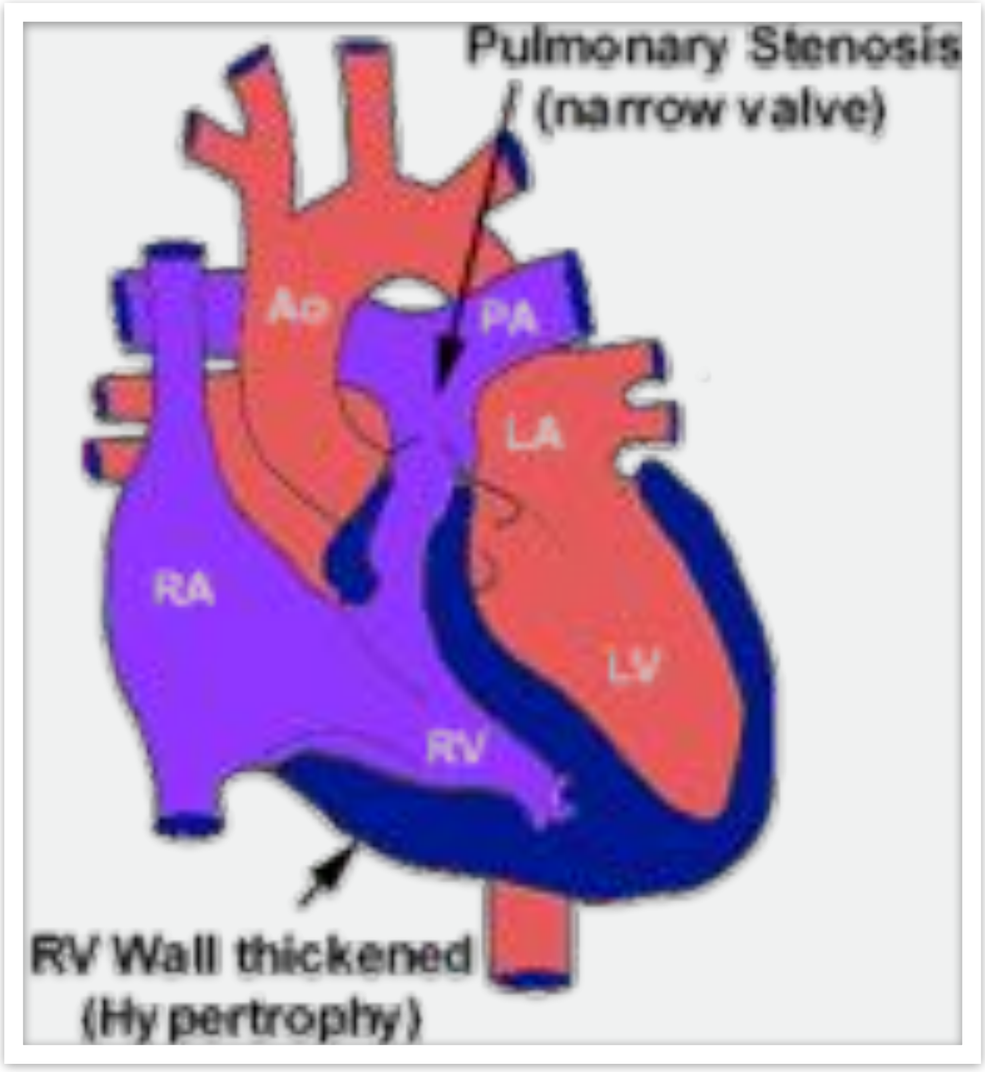
Cor triatriatum dexter

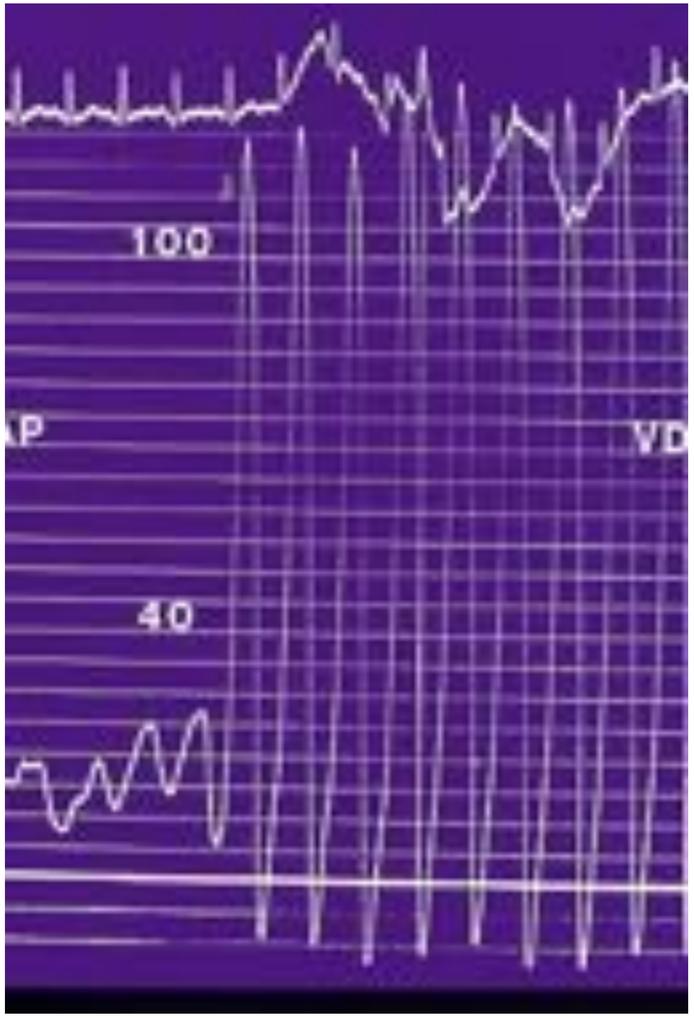



15/03/2018 15:17:20

Sténose valvulaire pulmonaire

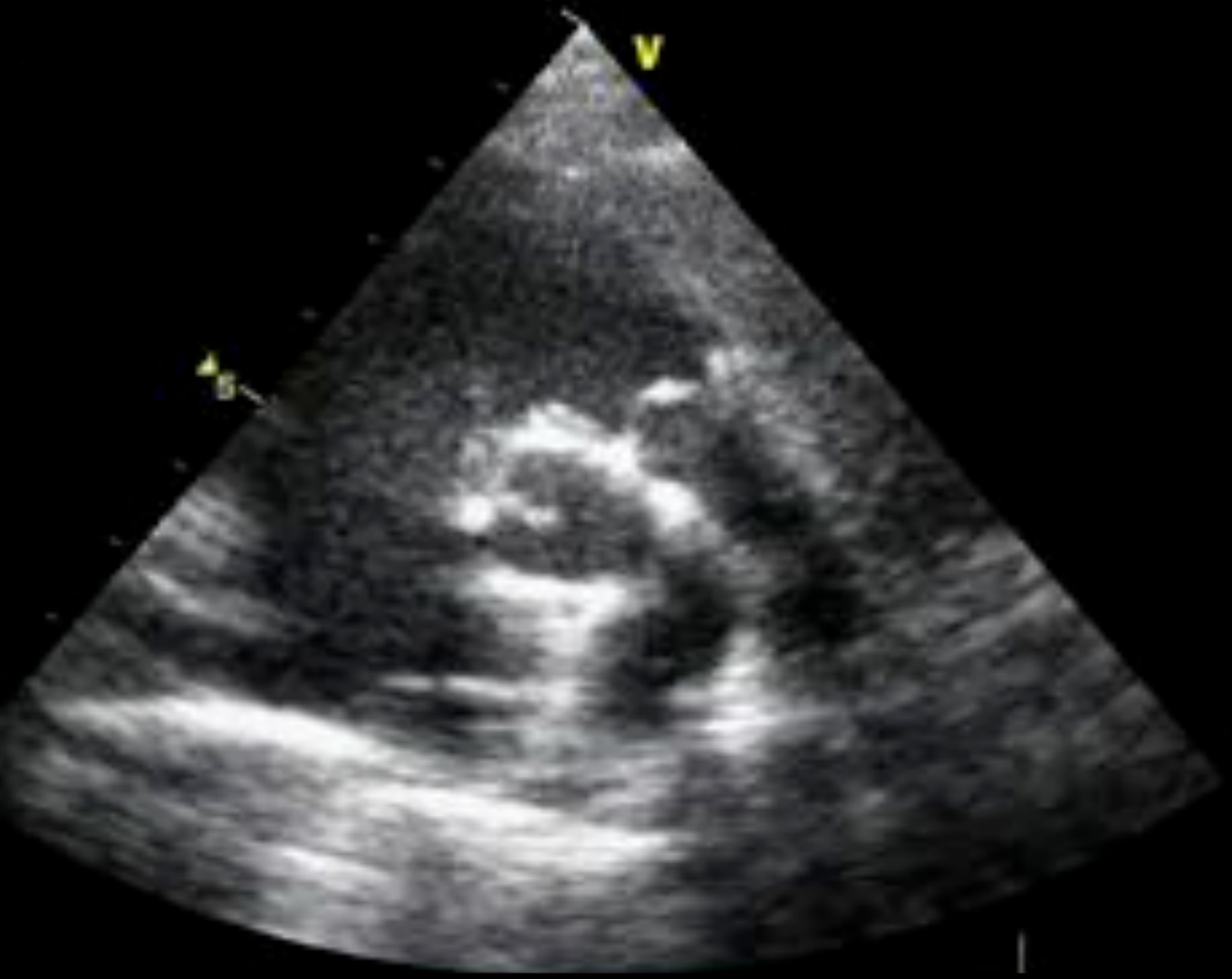
- Critique ?
- Syndromique ?
- Prédire le succès de la dilatation :
  – valve, anneau, dilatation du tronc
- Evolutivité néonatale
- Récidive après dilatation

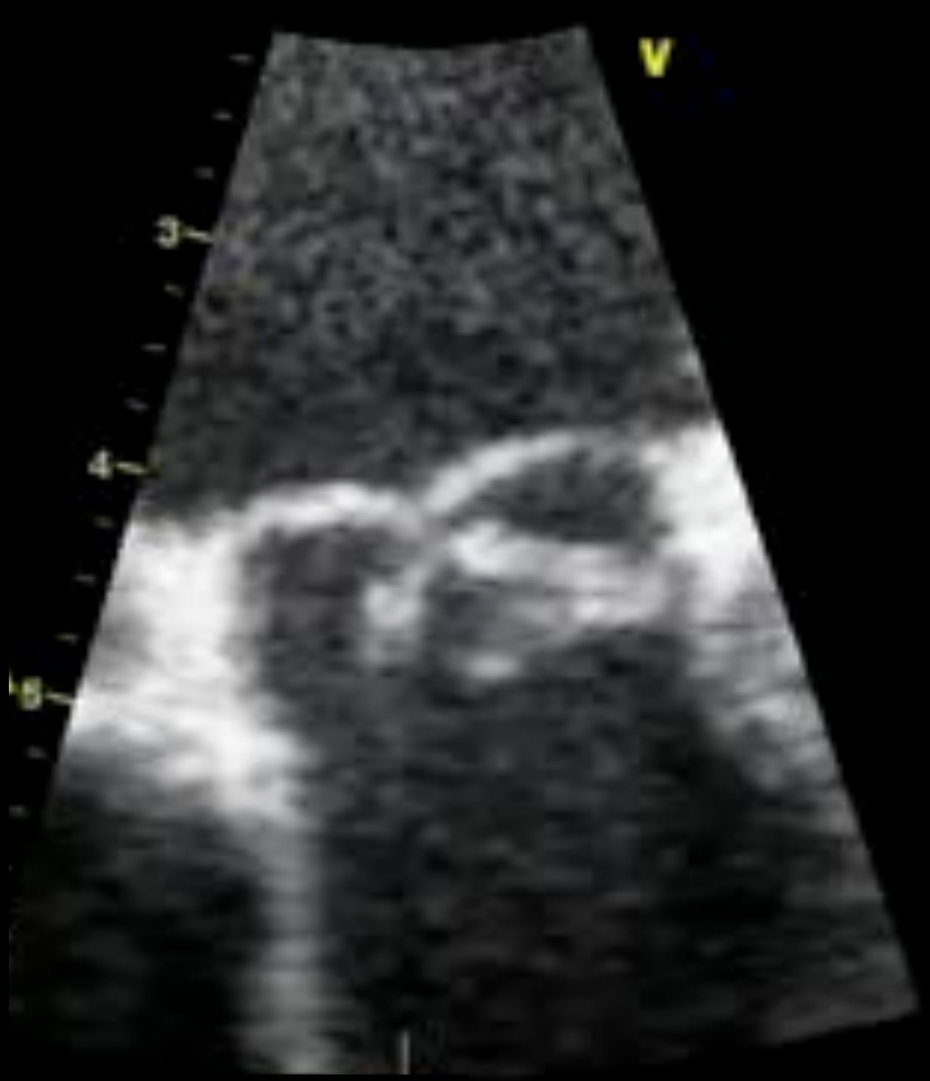

Pulmonary Stenosis
(narrow valve)
Ao
PA
LA
RA
LV
RV
RV Wall thickened
(Hypertrophy)

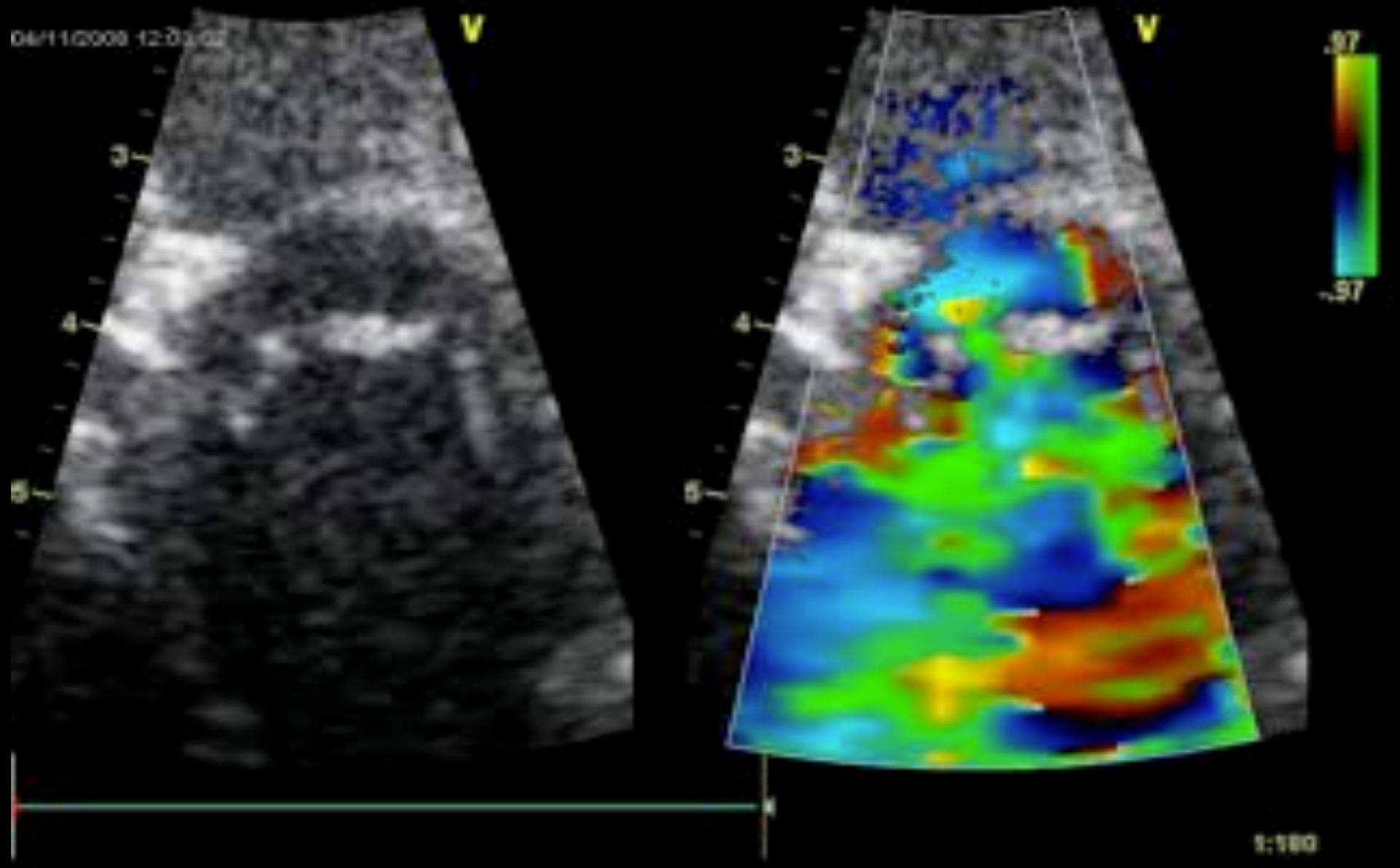

100
40

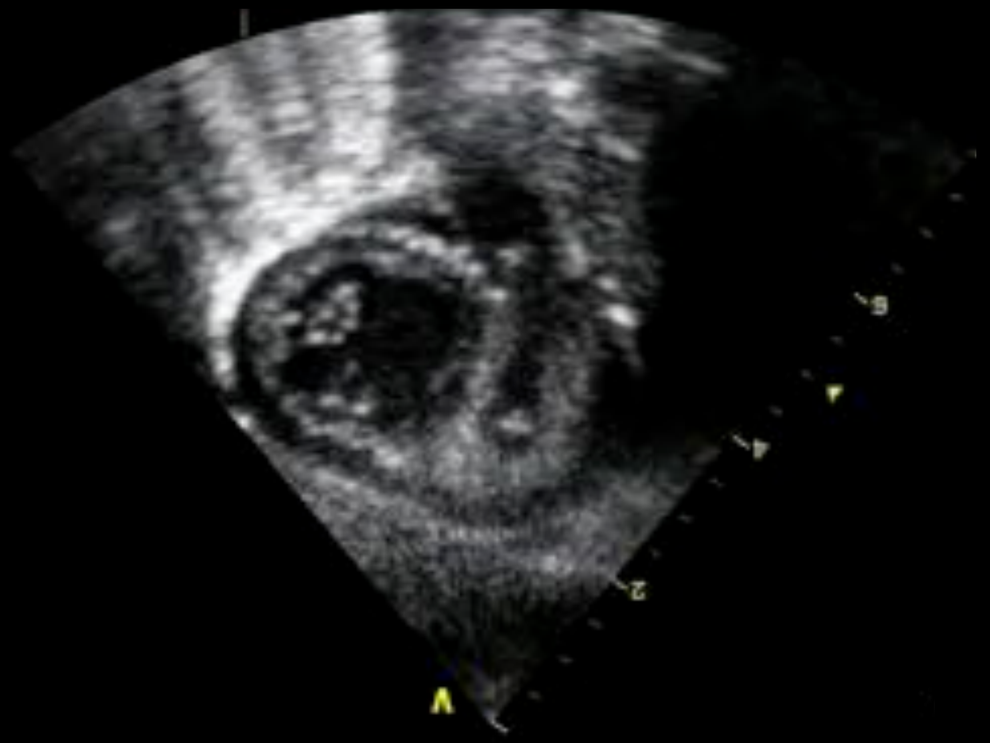
Valve pulmonaire

Sténose pulmonaire valvulaire

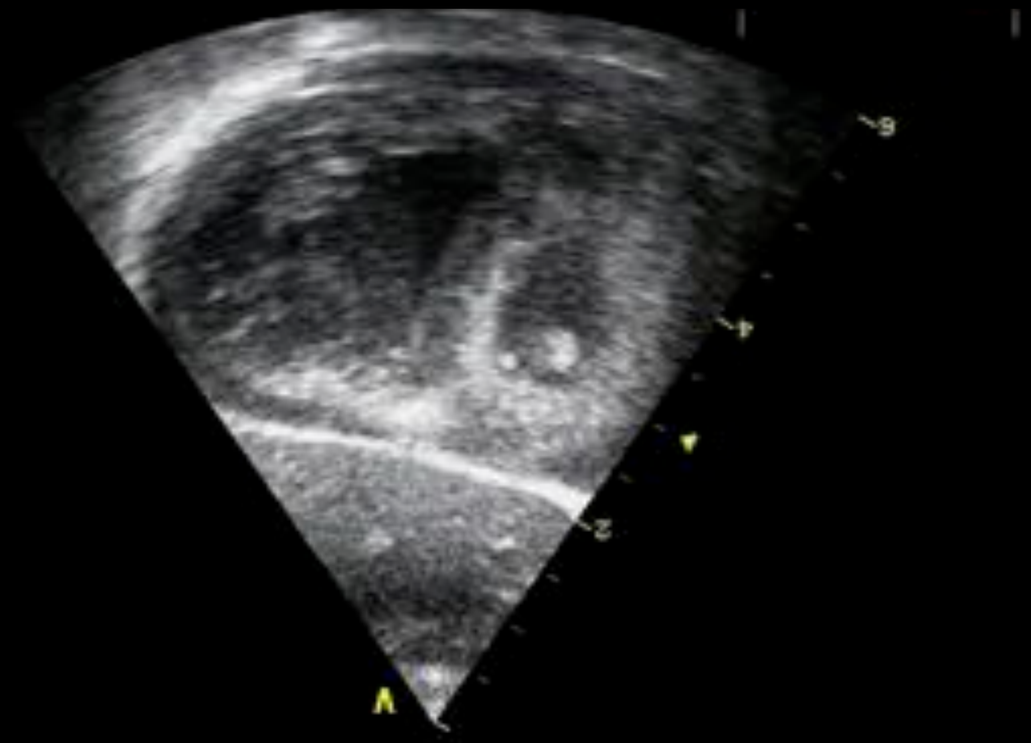

V
5
I


V
3
4
5
I


V
V
.97
-.97
5:180

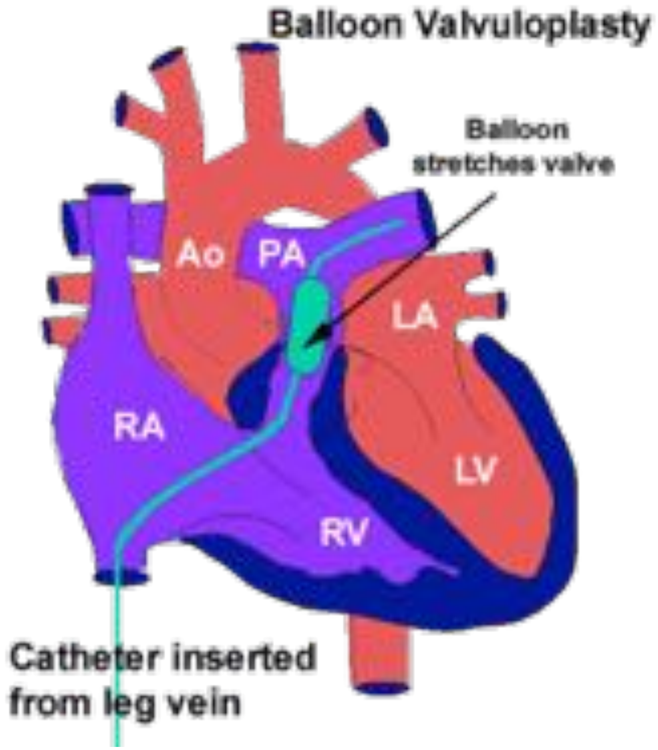


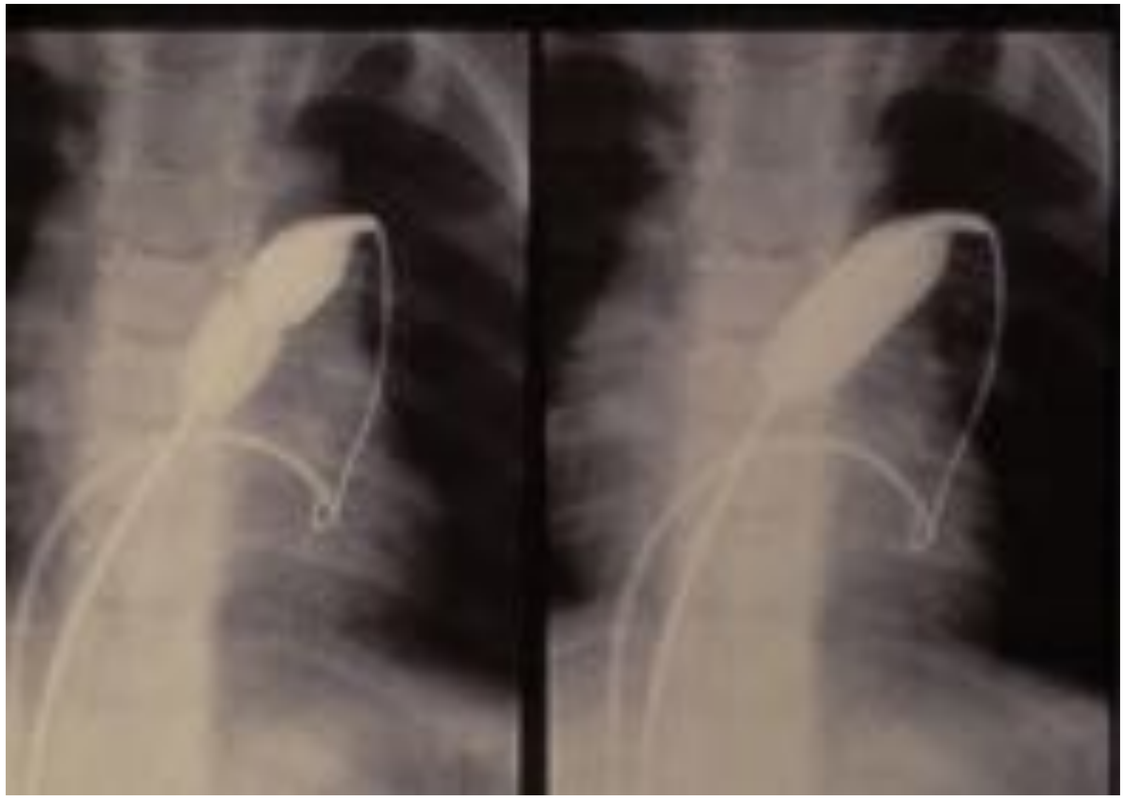
Sténose pulmonaire



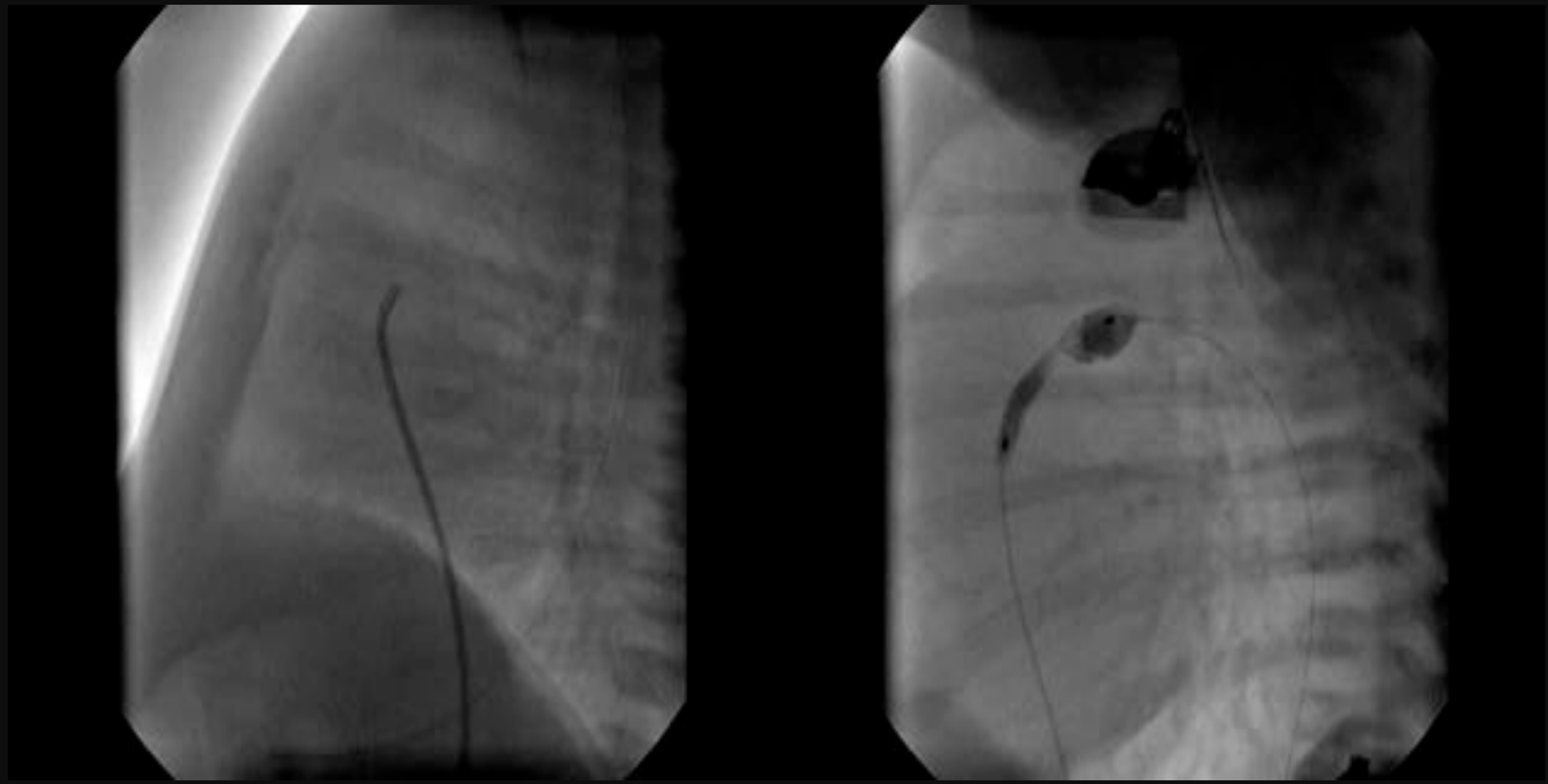
APSI

Dilatation percutanée

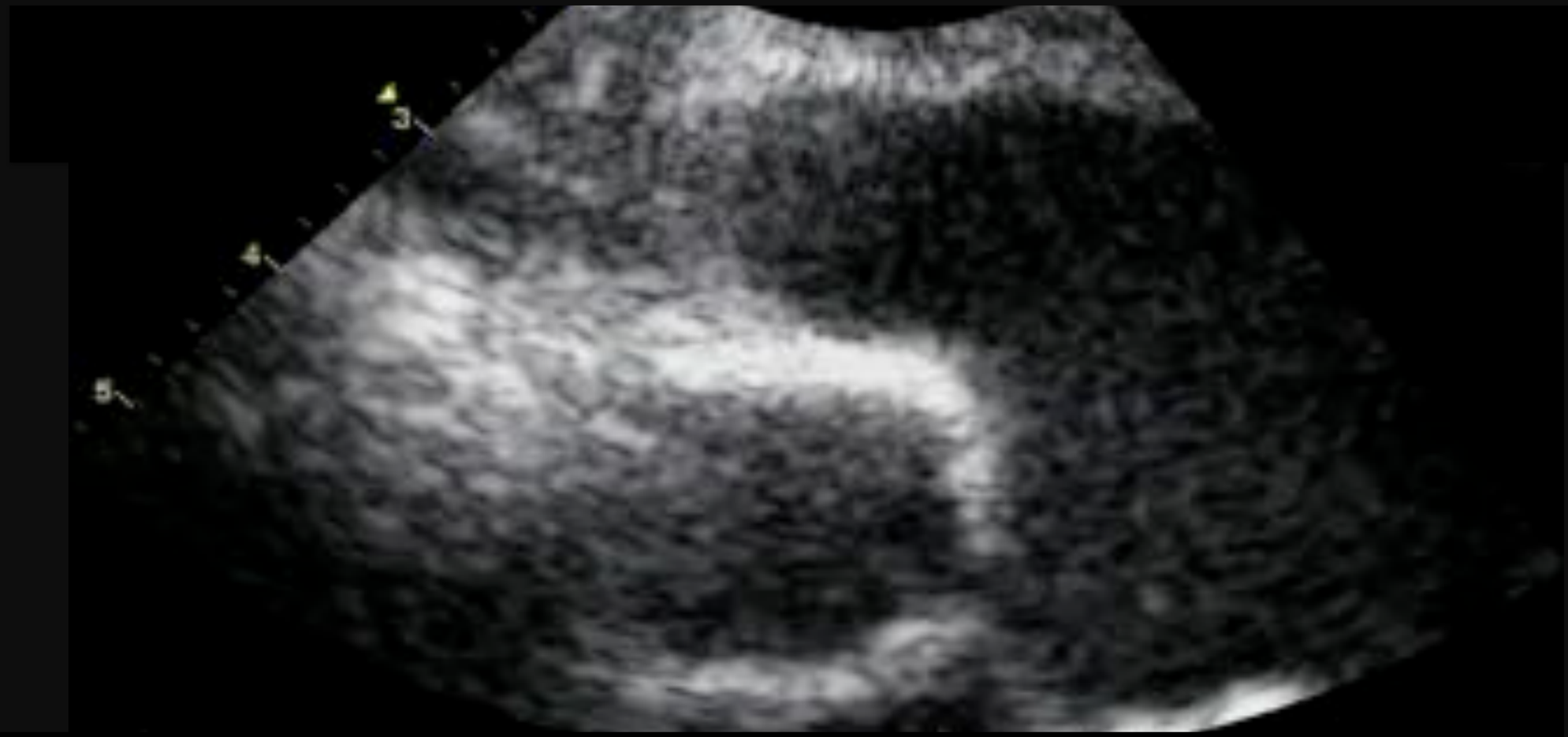




Dilatation percutanée



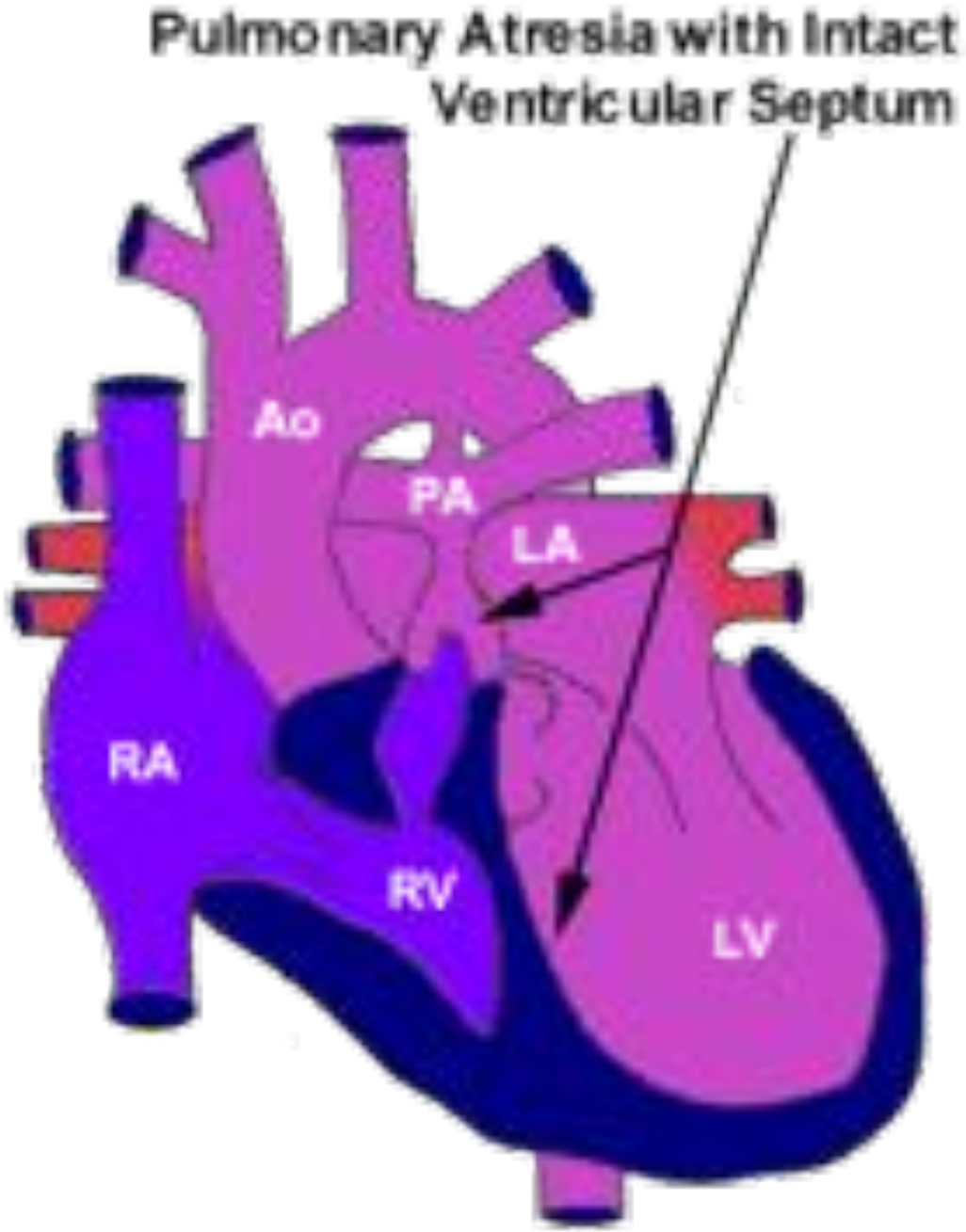
Valve pulmonaire après dilatation percutanée



Valve pulmonaire

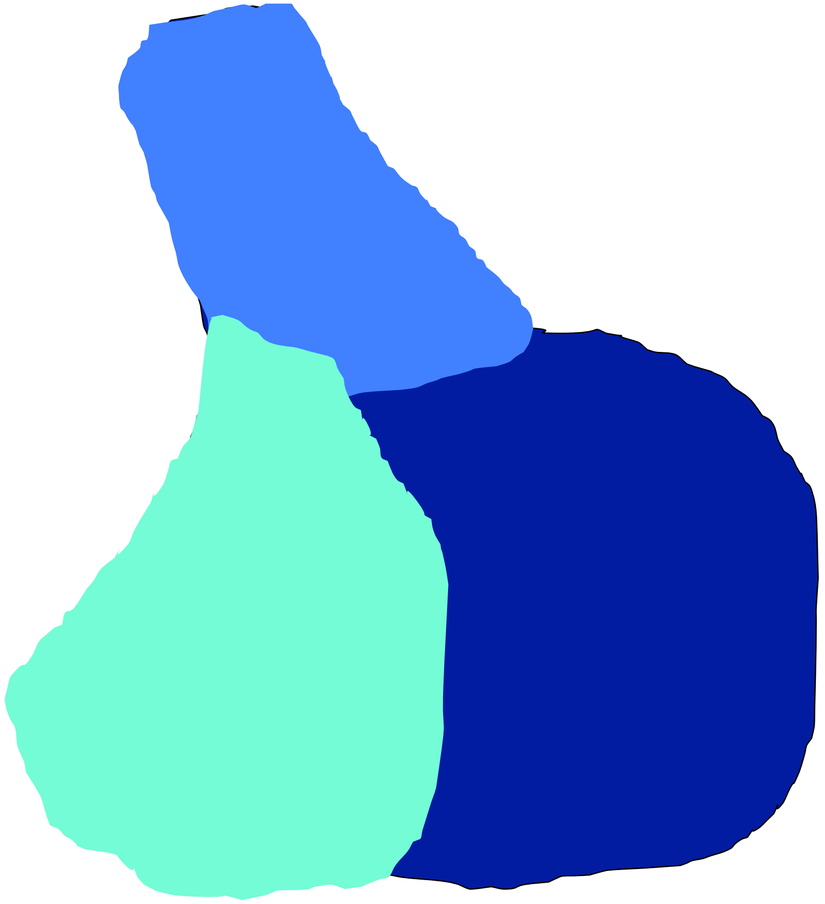
Atrésie pulmonaire à septum intact

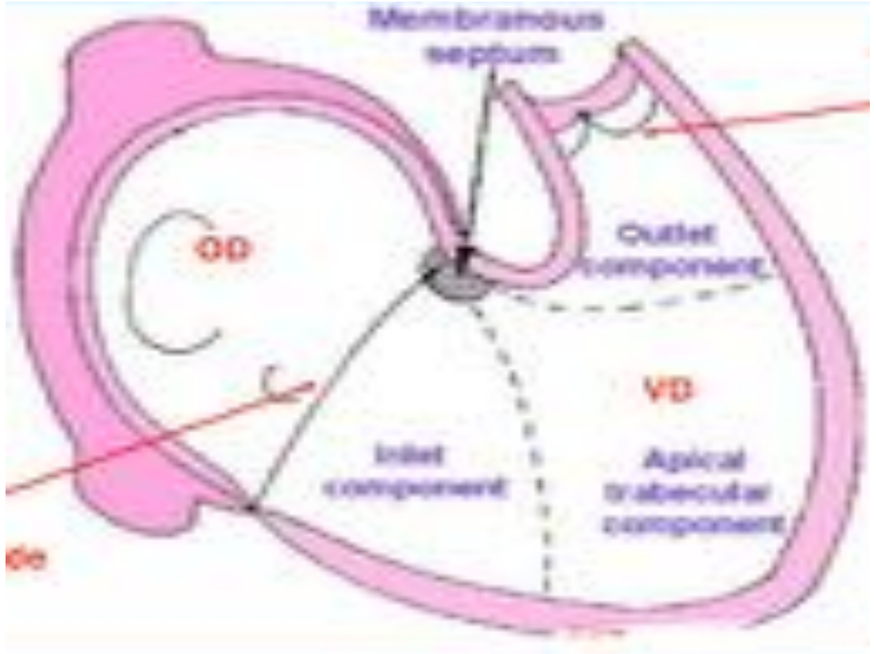
APSI

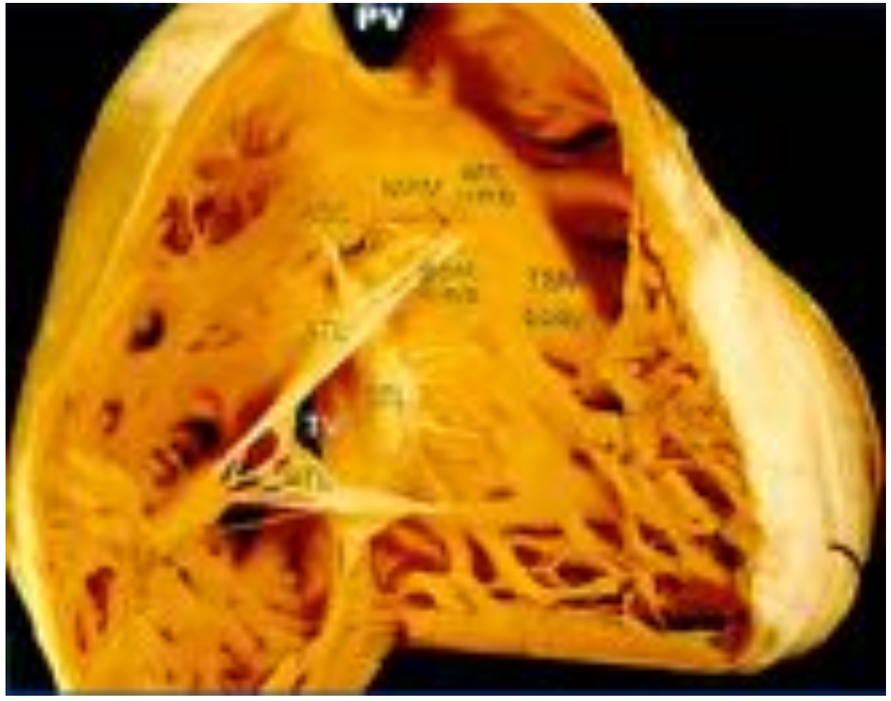

Pulmonary Atresia with Intact Ventricular Septum
Ao
PA
LA
RA
RV
LV

Atrésie pulmonaire à septum intact

- Evolutivité pendant la vie foetale
- Anatomie du VD
  - Nombre de chambres
  - Taille de la tricuspide
  - Anneau pulmonaire
- Perfusion coronaire
  - Fistules/sinusoïdes
  - VD dépendance




Membranous
septum
OD
Outlet
component
VD
Inlet
component
Apical
trabecular
component

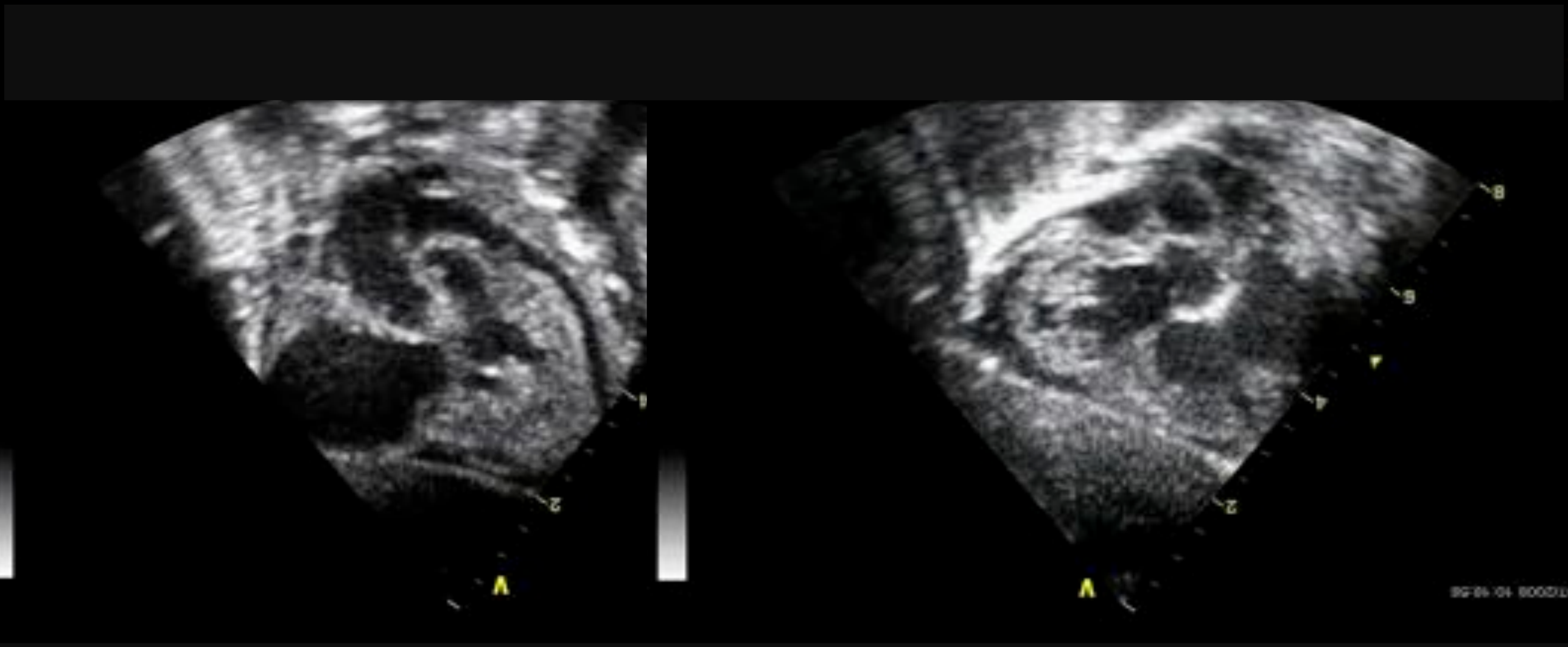

PV

Atrésie pulmonaire à septum intact

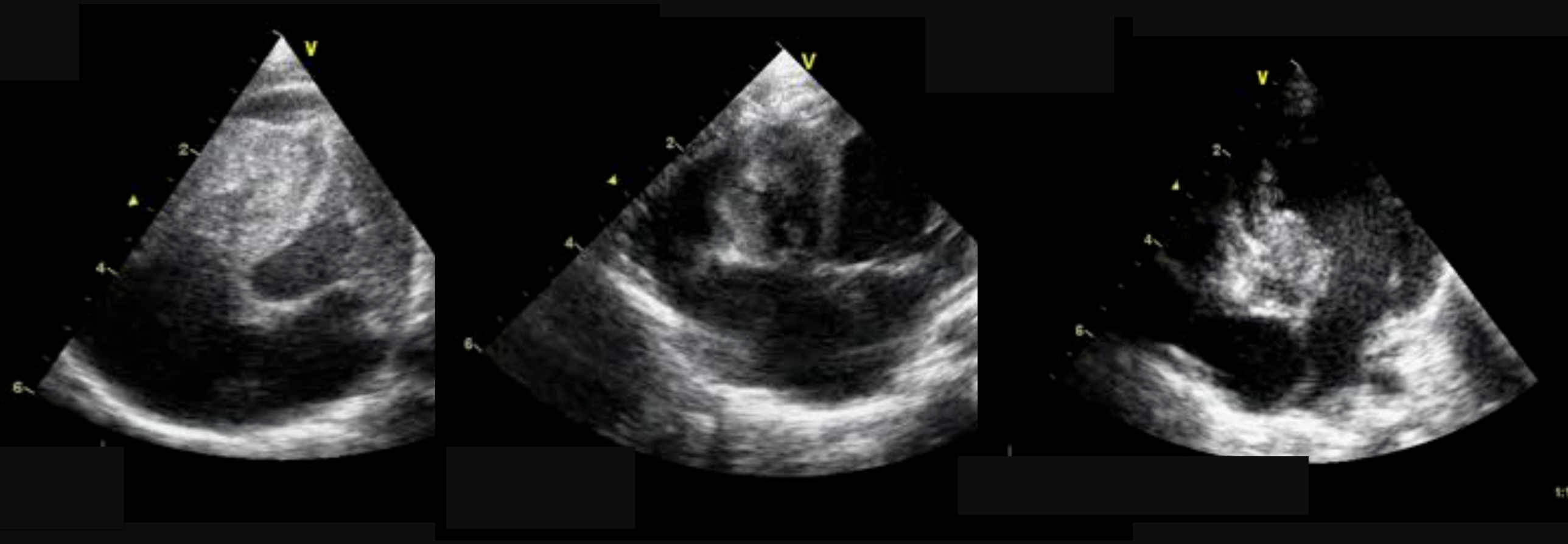
Projet thérapeutique

- Tripartite
  - 2 ventricules
- Bipartite : admission + infundibulum
  - 1,5 ventricule
- Unipartite (croupion)
  - 1 ventricule

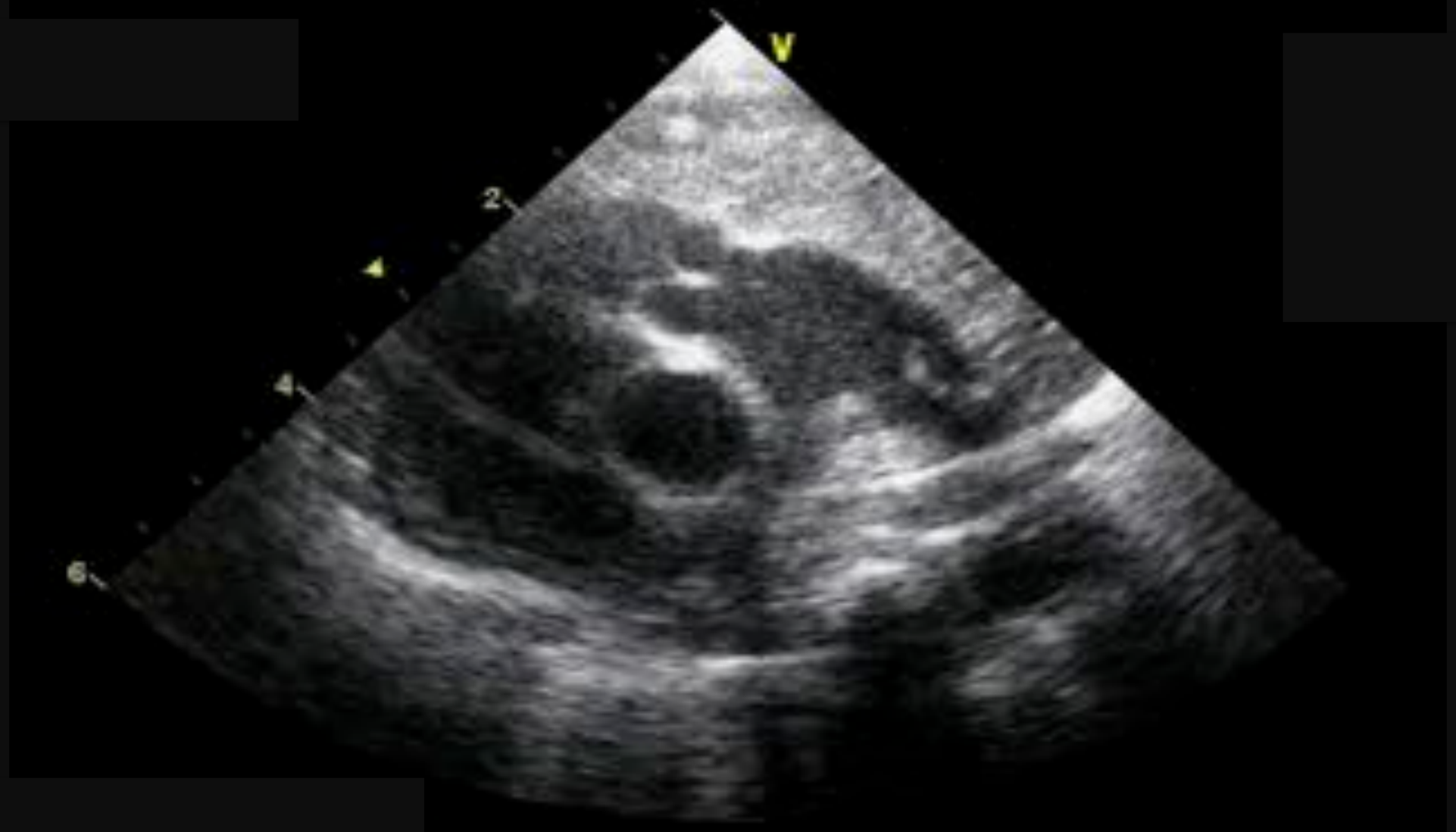
Pulmonary atresia intact septum Tripartite Right ventricle



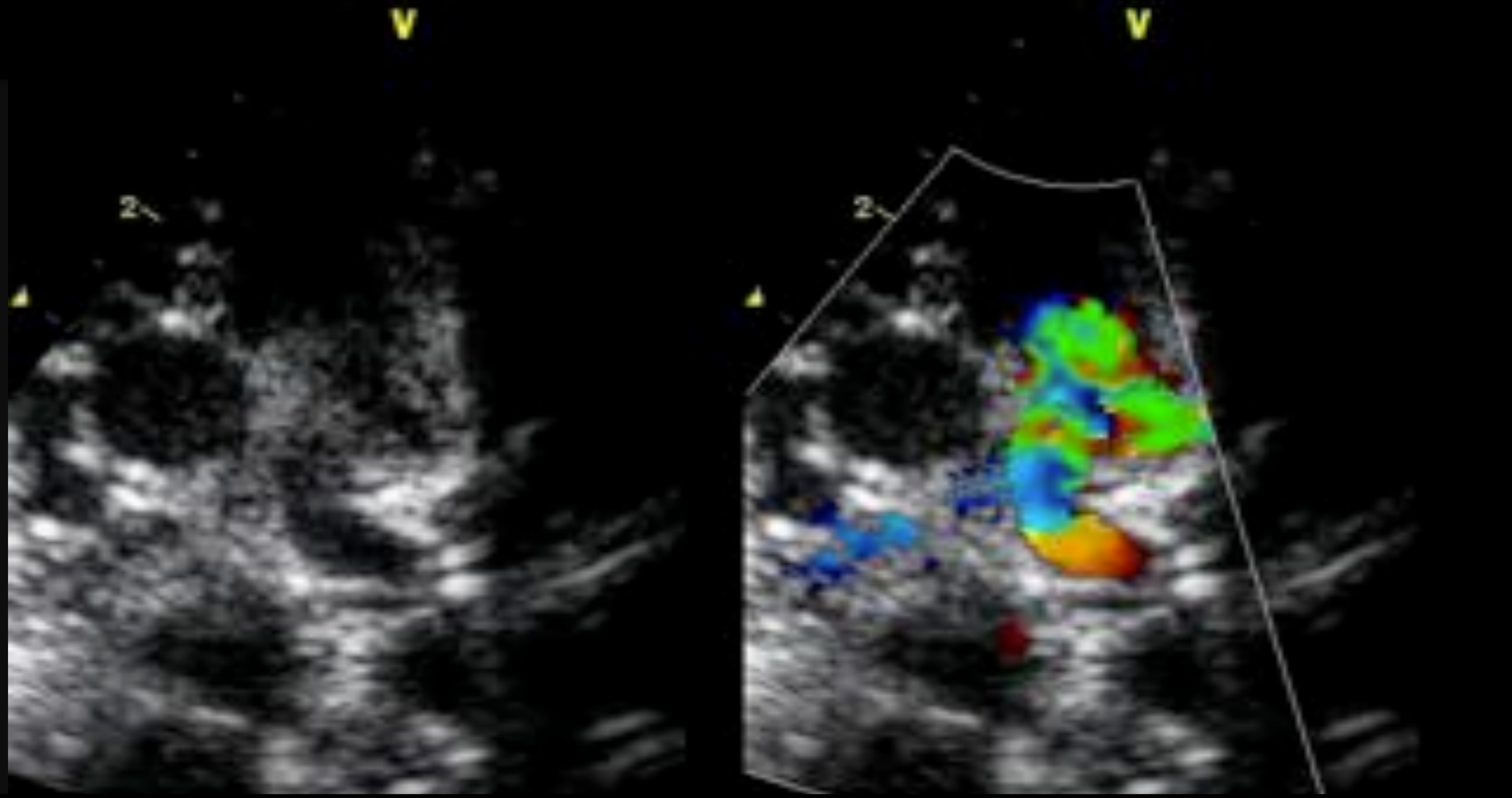
Pulmonary atresia intact septum Right ventricle (3-2-1)



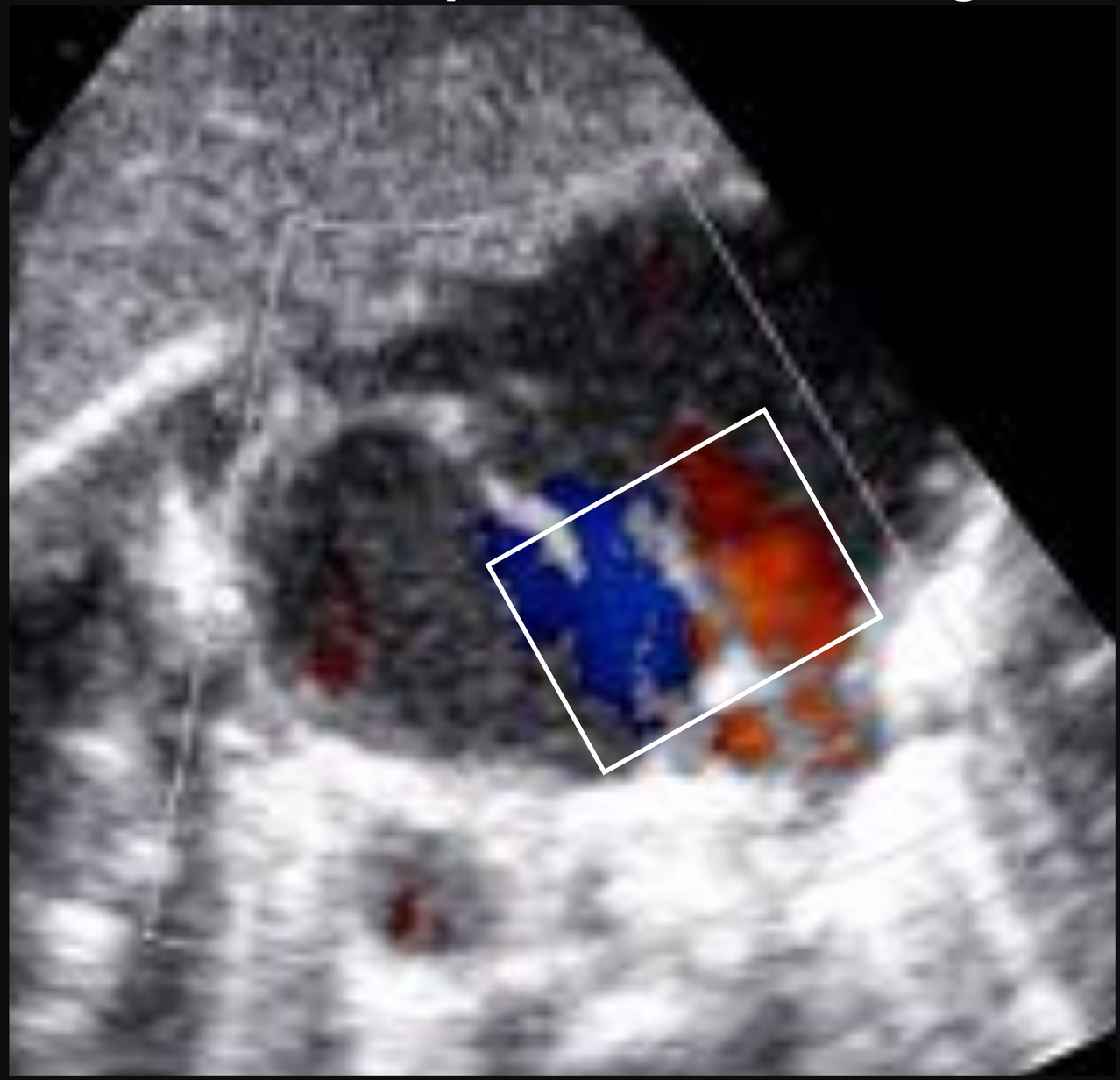
Pulmonary atresia intact septum Pulmonary valve



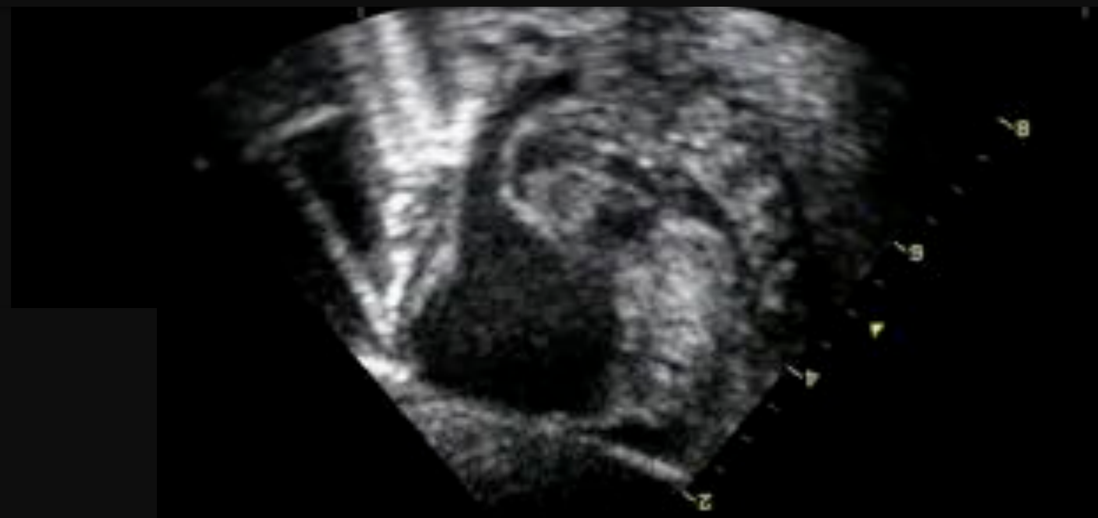
Pulmonary atresia intact septum Arterial duct flow

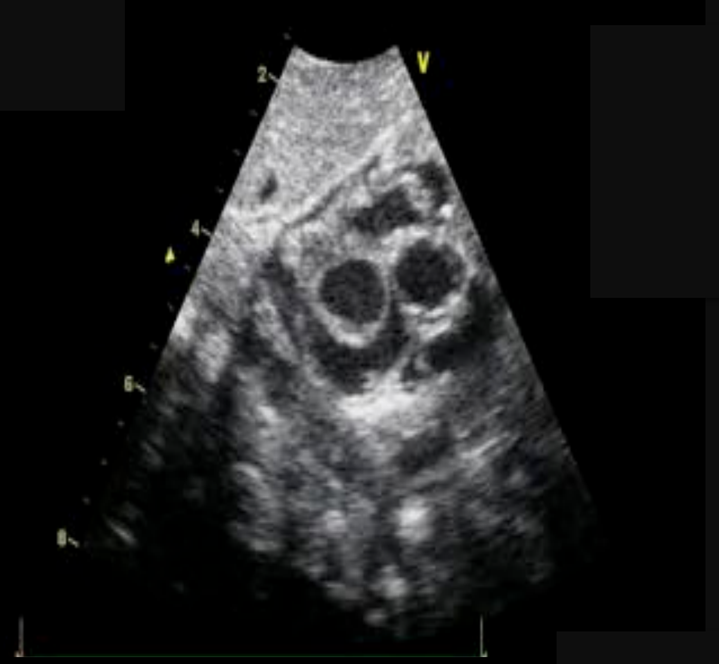


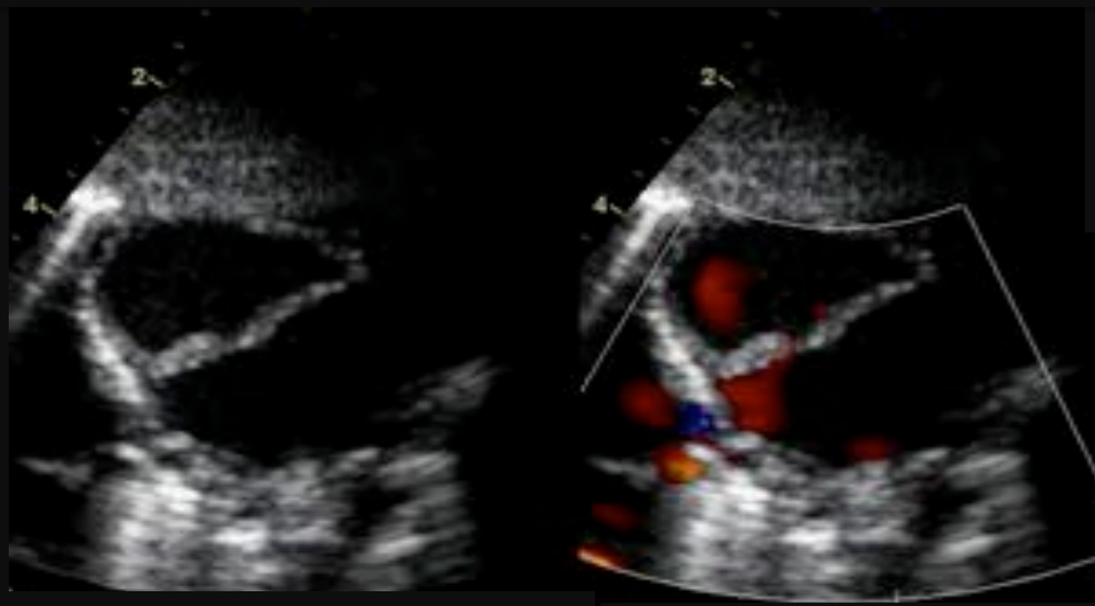
Pulmonary atresia intact septum Interatrial Right to Left shunt

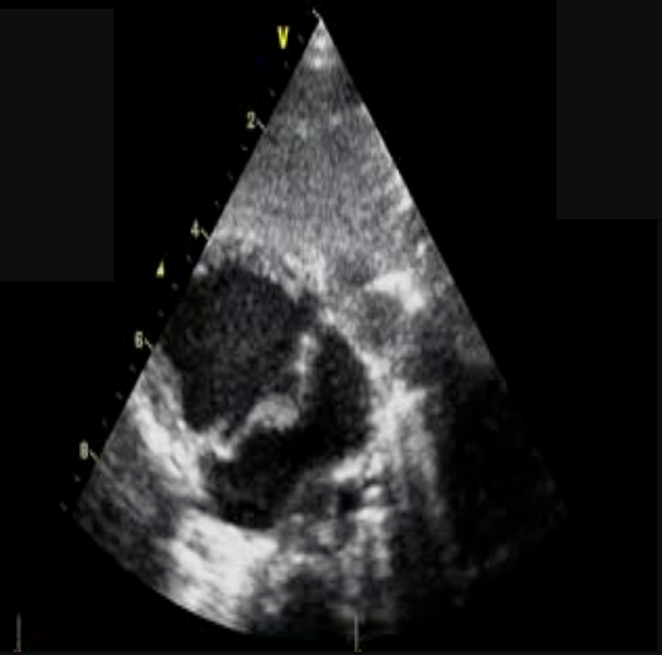


Champions du monde**

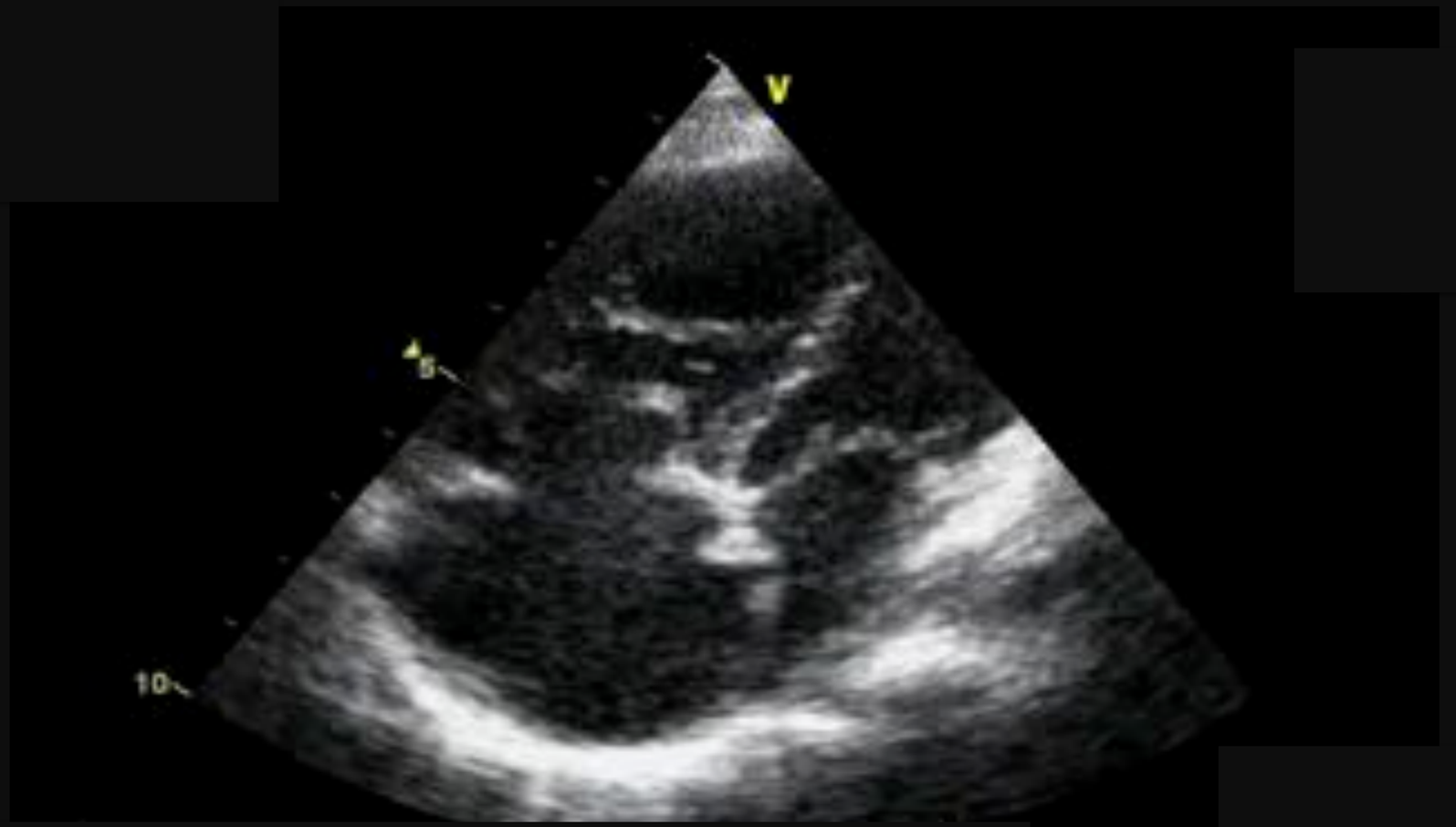




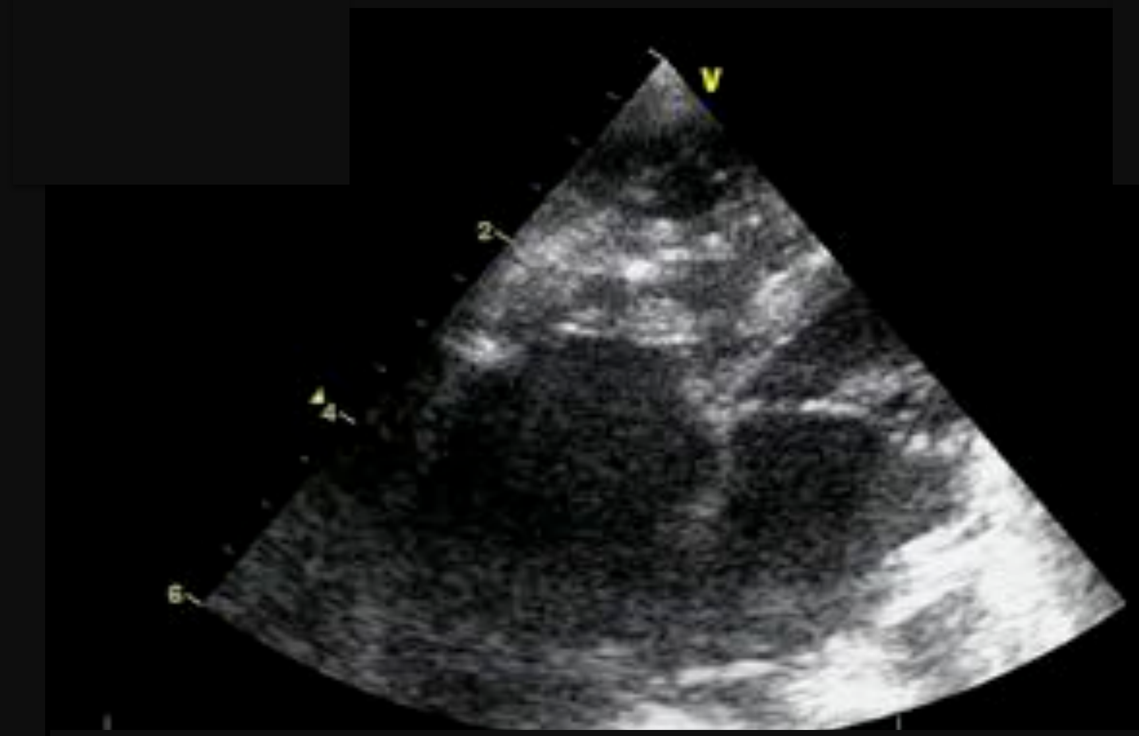




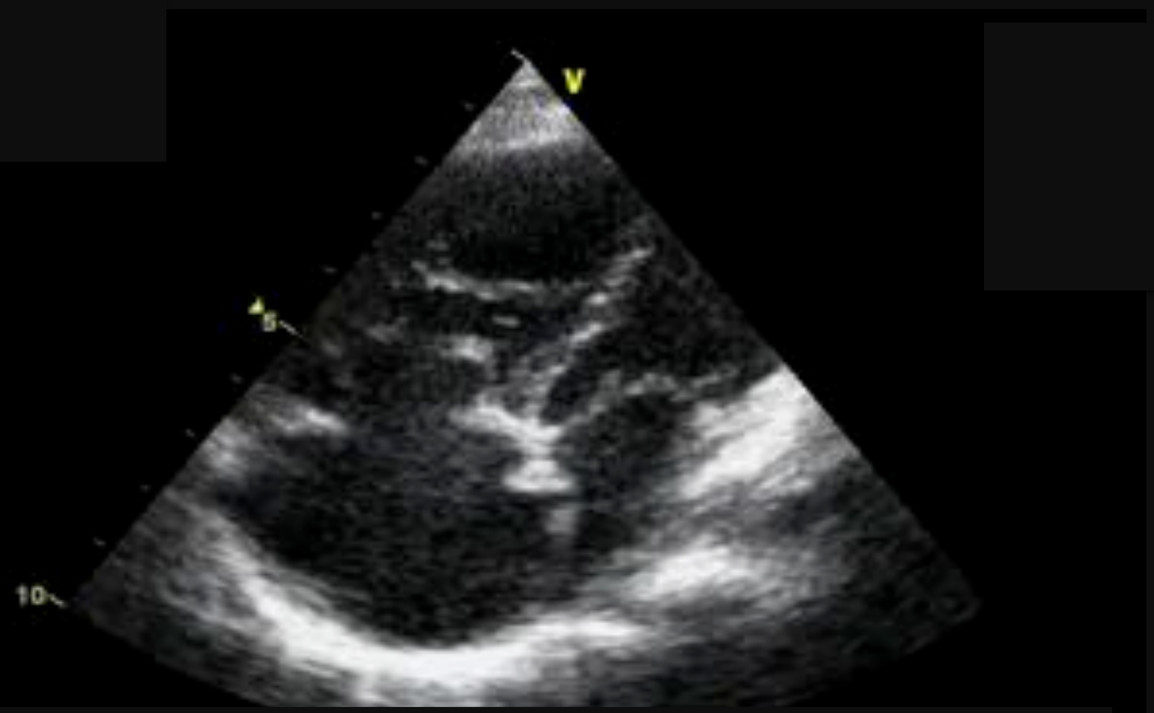
RA and FO in PA-IVS



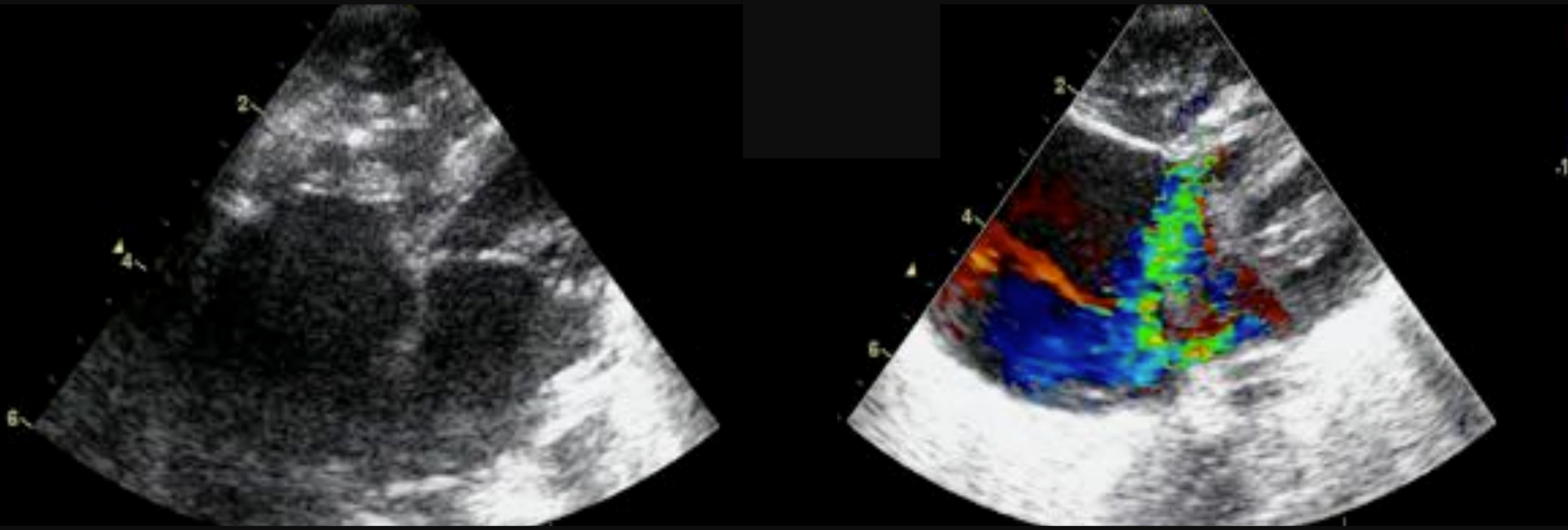
Tricuspid valve in PA-IVS



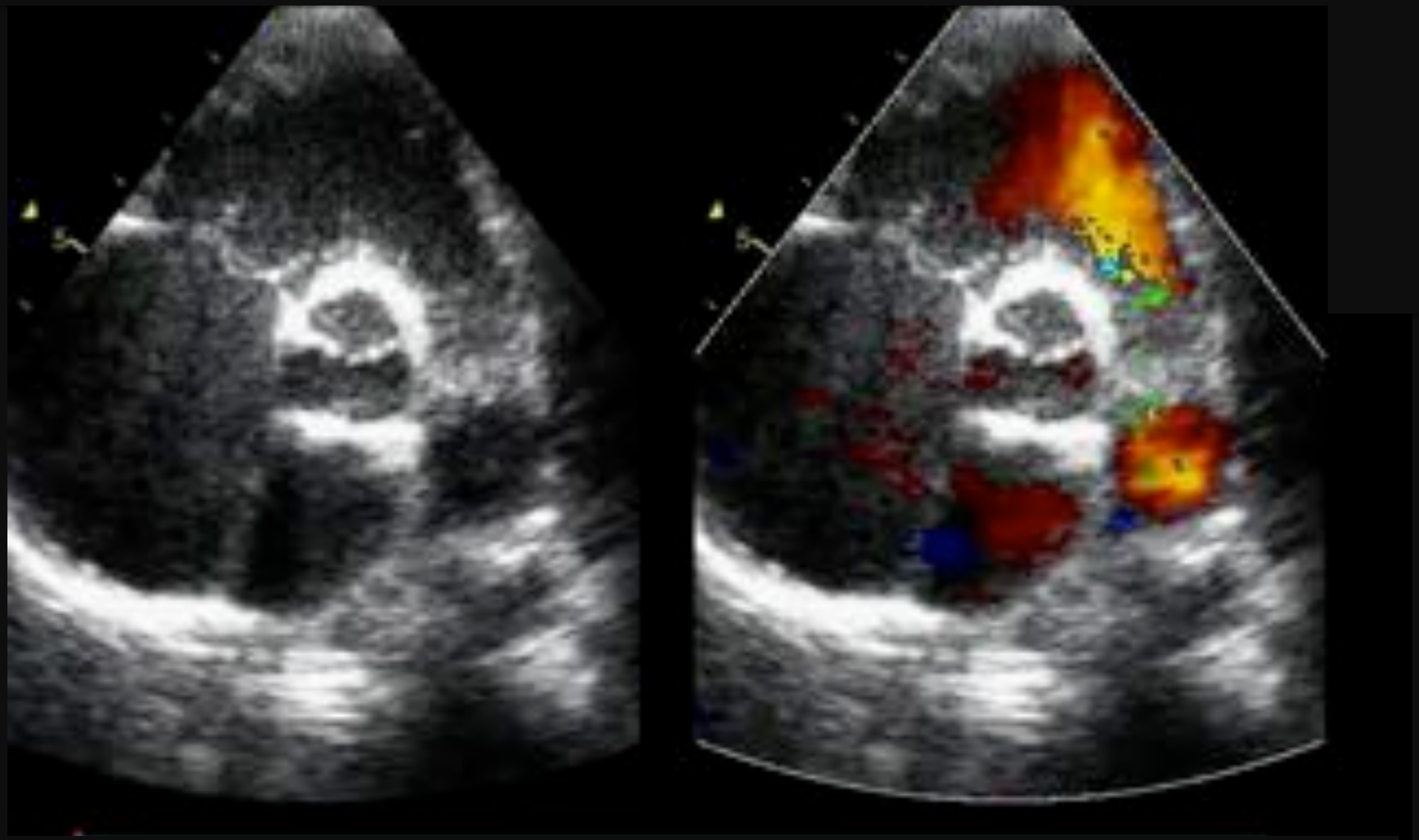
Tricuspid valve dysplasia



Tricuspid valve in PA-IVS

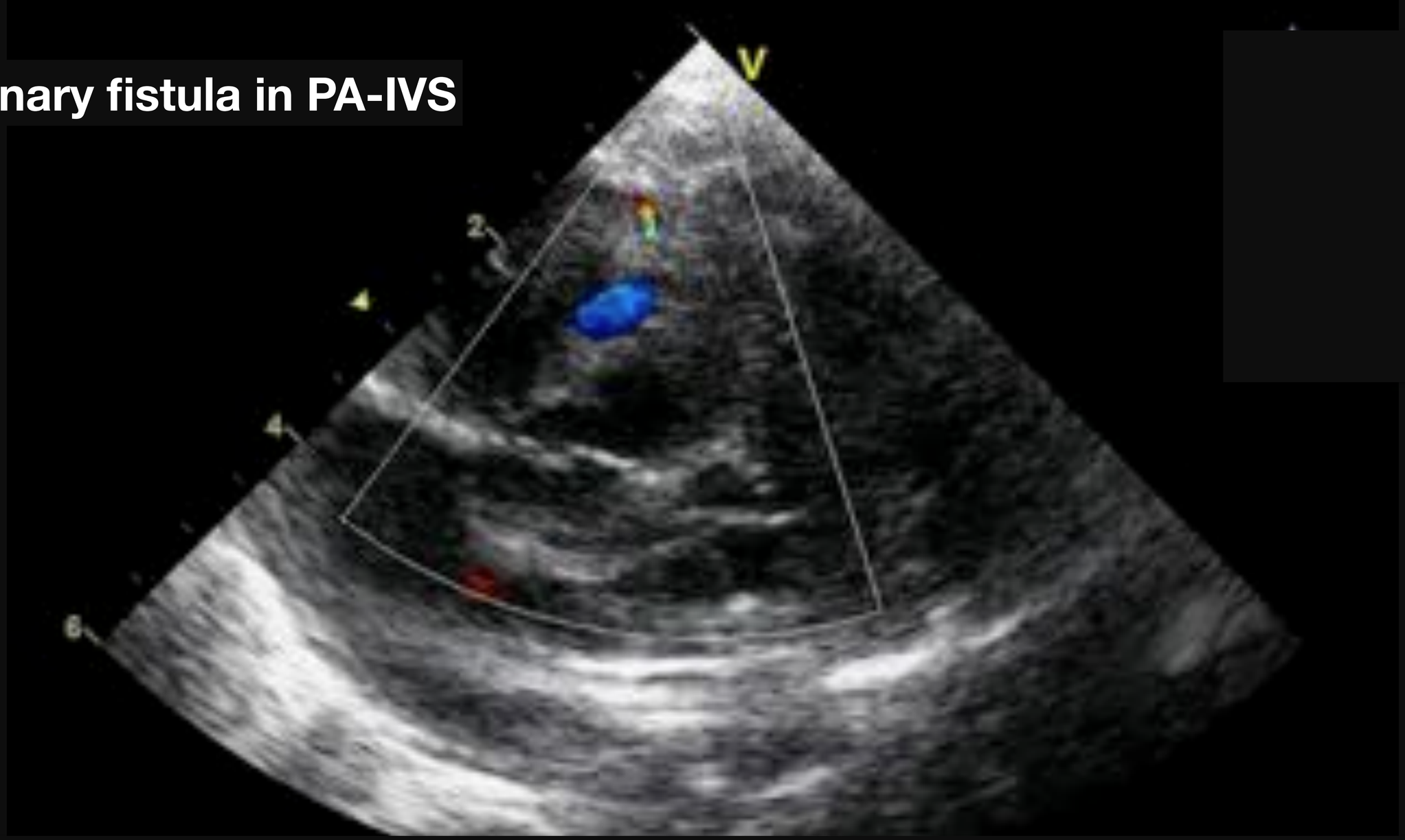


Tricuspid valve dysplasia

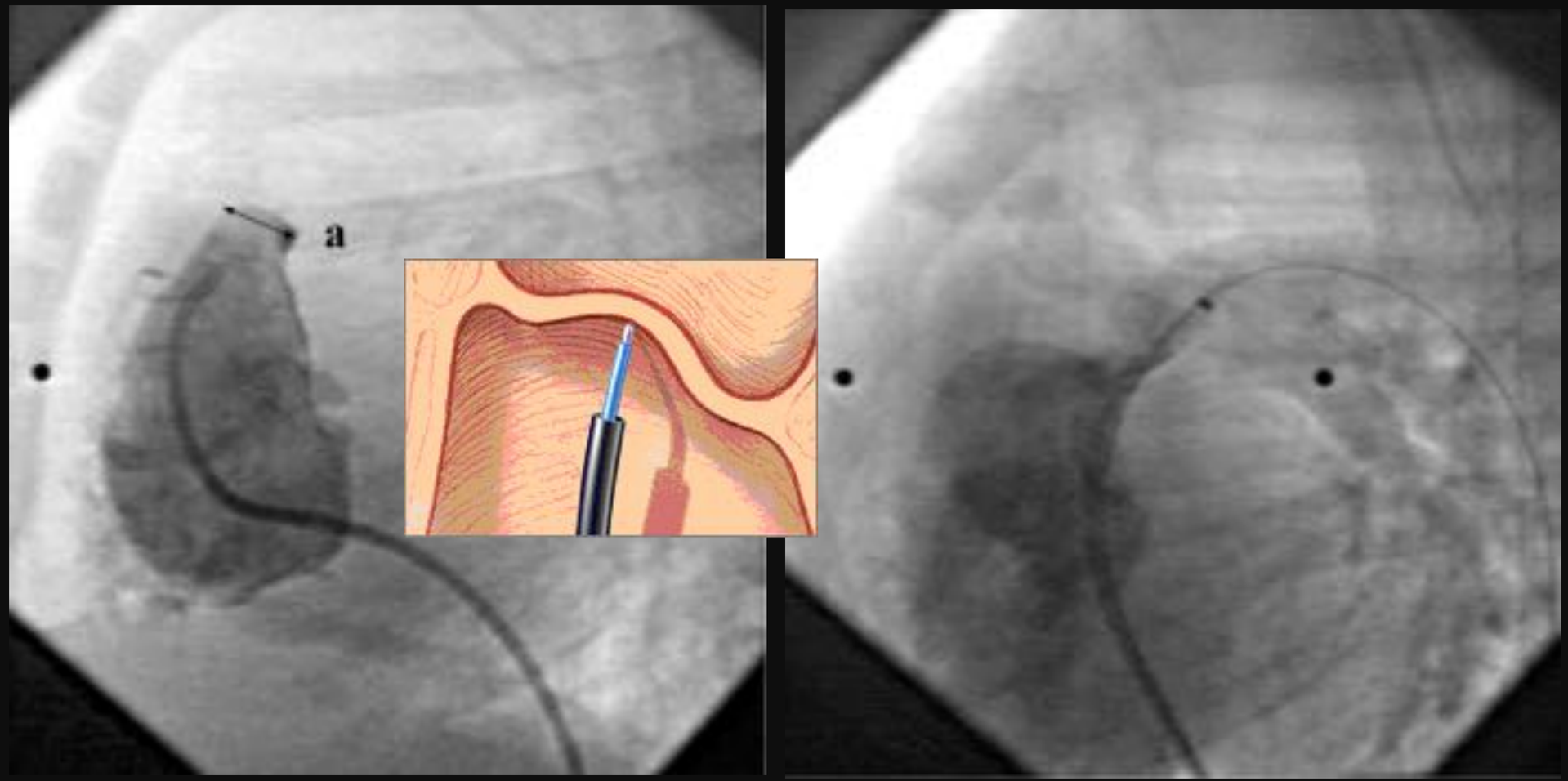


Tricuspid valve dysplasia and PA-IVS/the circle of death

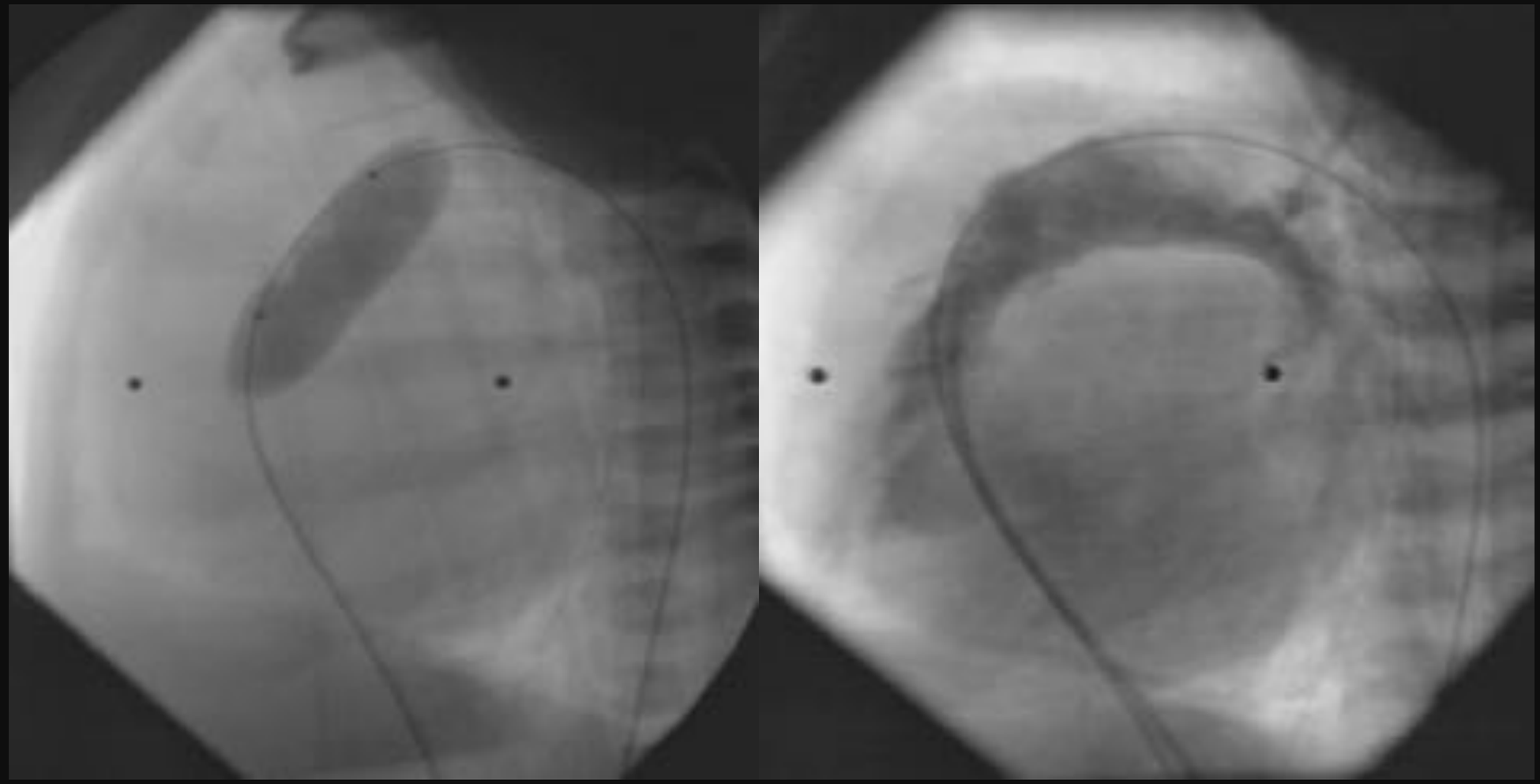
Coronary fistula in PA-IVS



Perforation of pulmonary valve and dilatation in PA-IVS



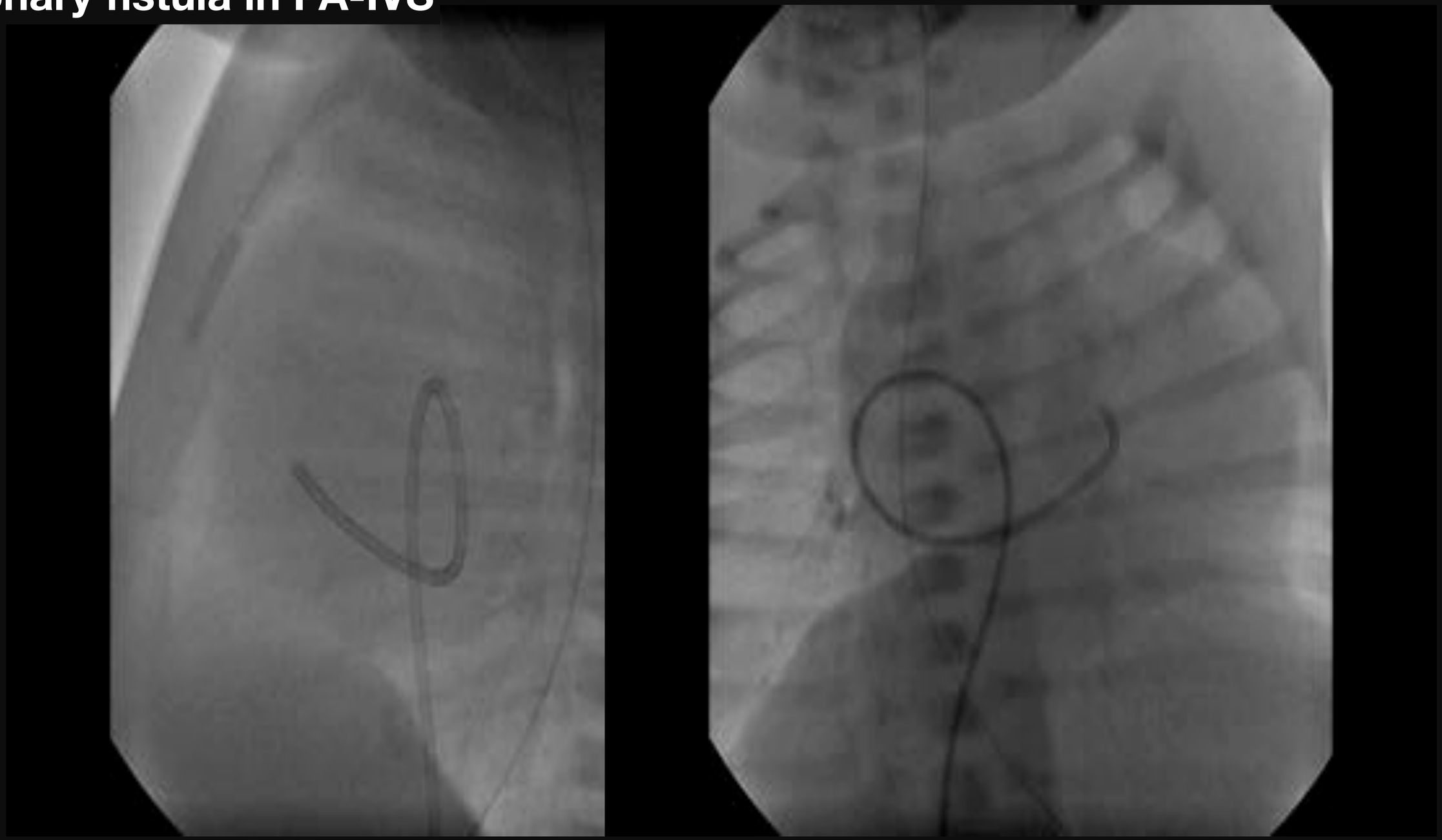
Perforation of pulmonary valve and dilatation in PA-IVS

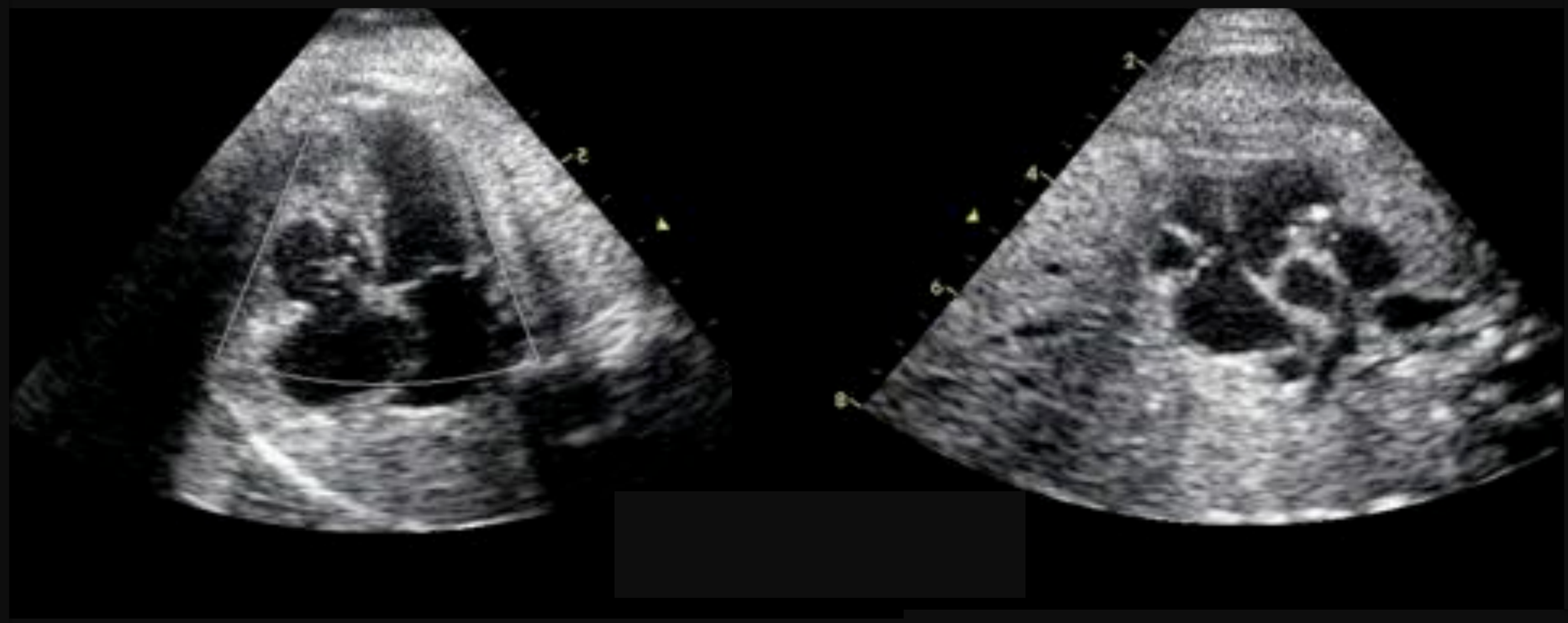


APSI post perforation

- Cyanose persistante
  - Compliance du VD
    - Béta-bloquants
    - Anastomose de Blalock
    - Stent du canal artériel
- Insuffisance tricuspide
- Croissance du VD et de la tricuspide

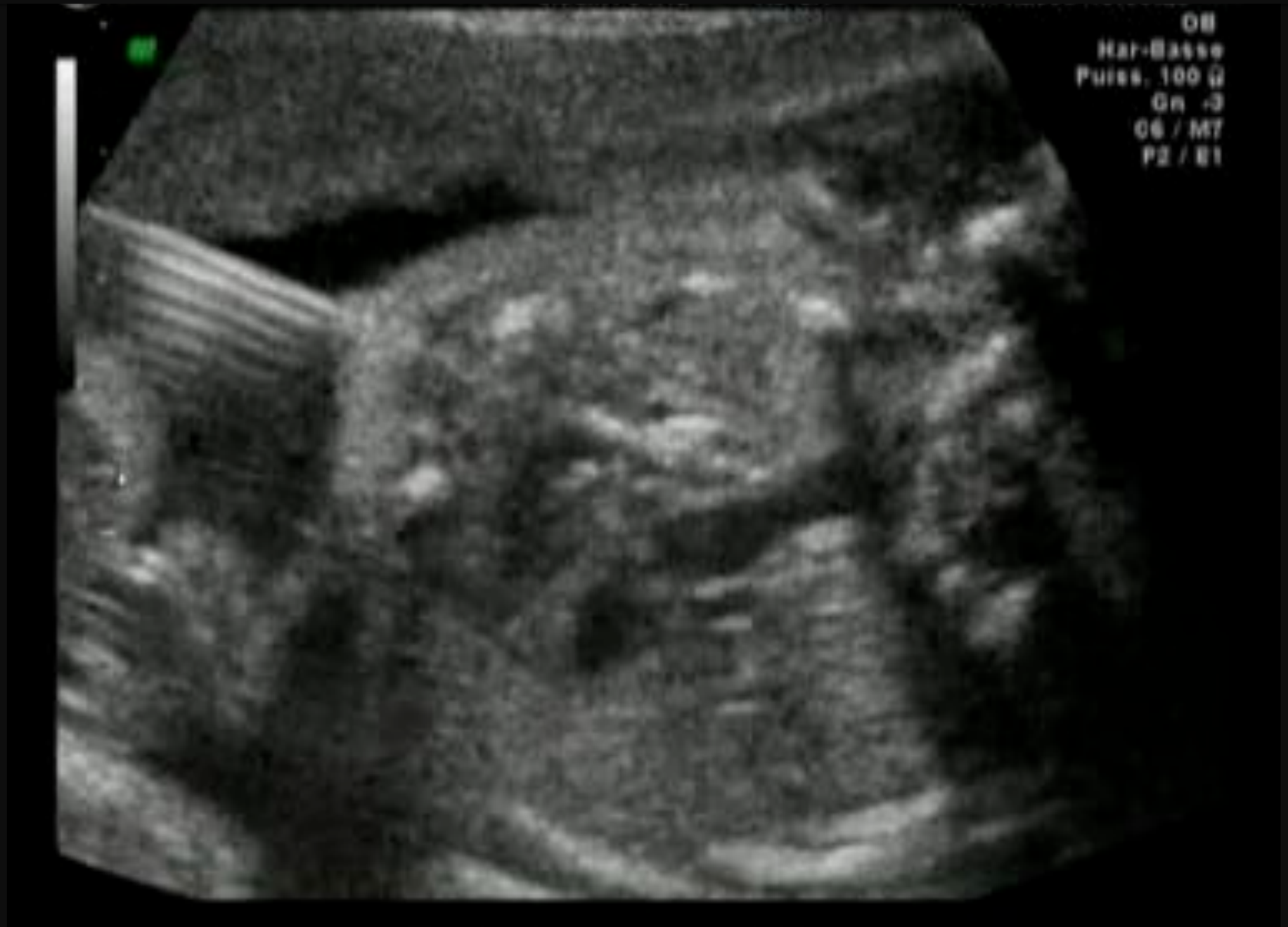
Coronary fistula in PA-IVS

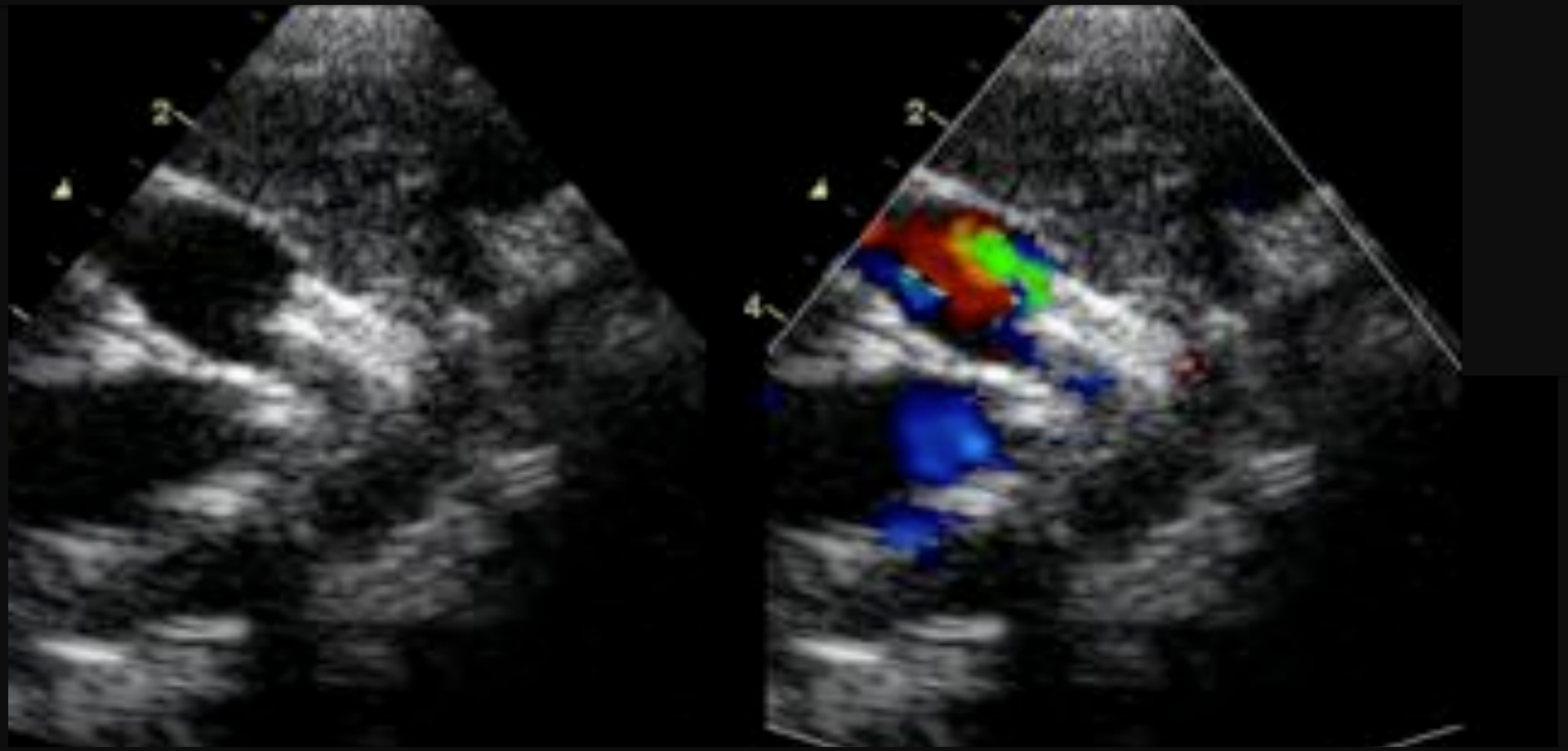




Foetal PA-IVS

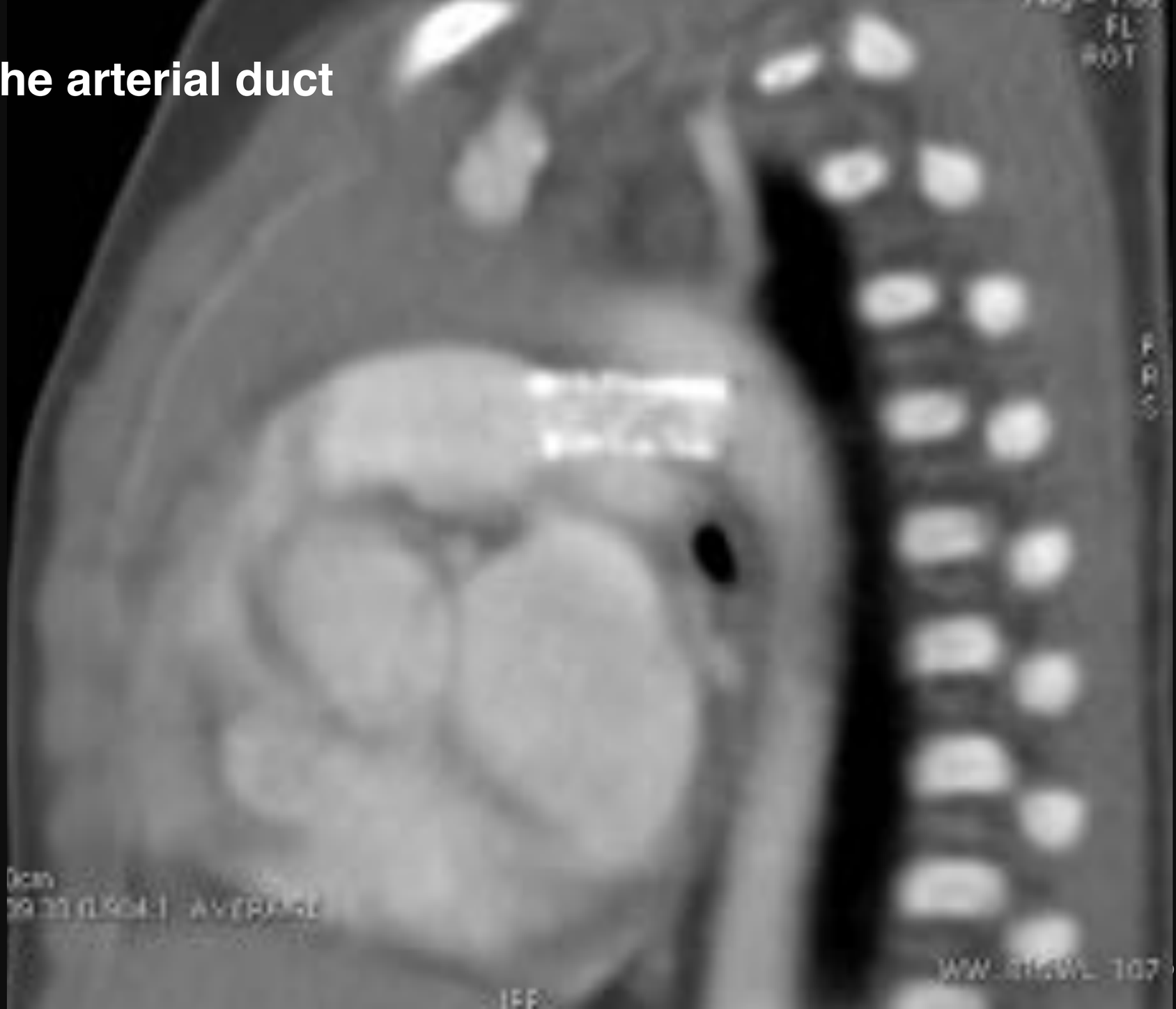
Foetal PA-IVS Interventional cath in utero





Stenting of the arterial duct

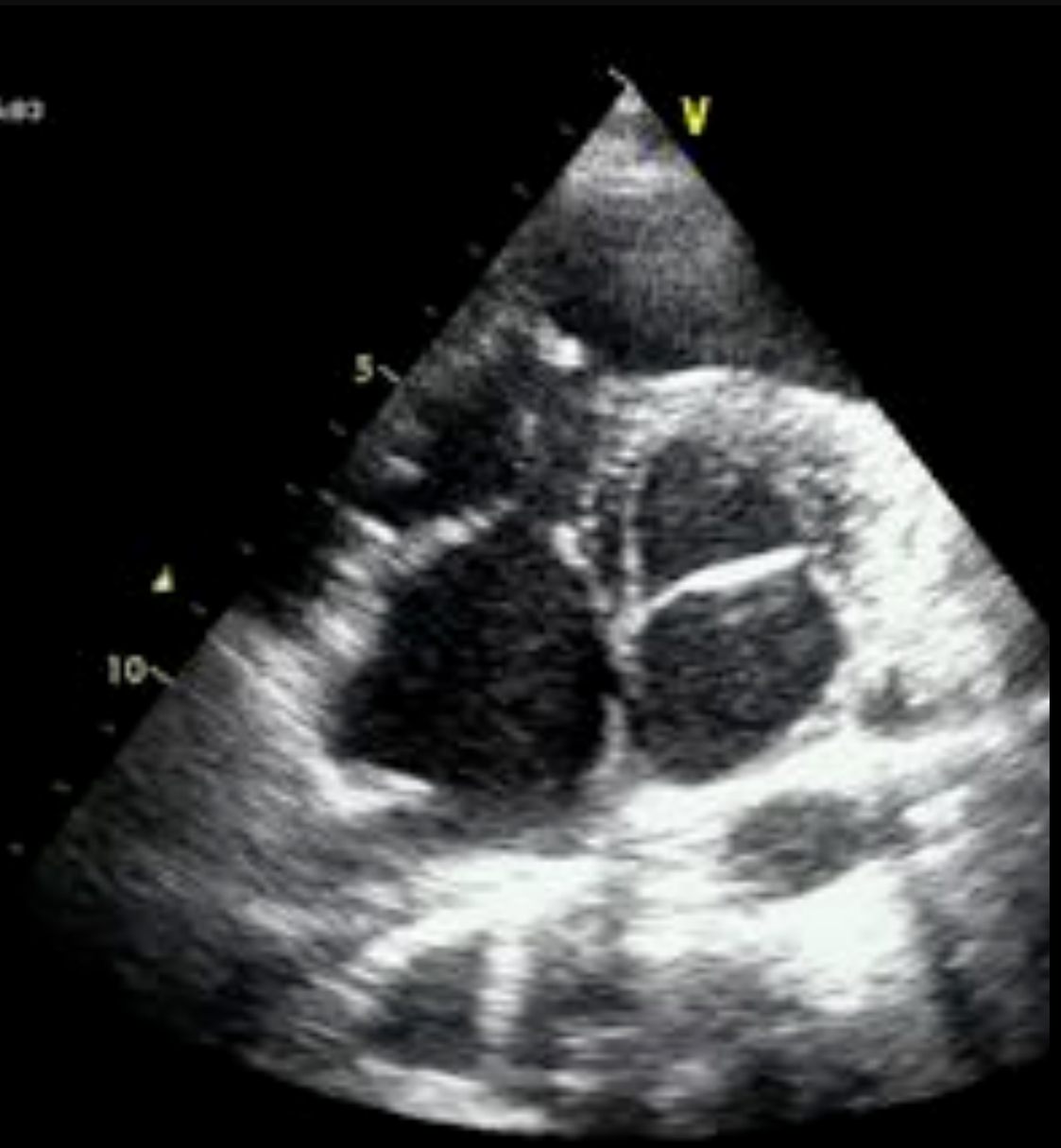
Stenting of the arterial duct

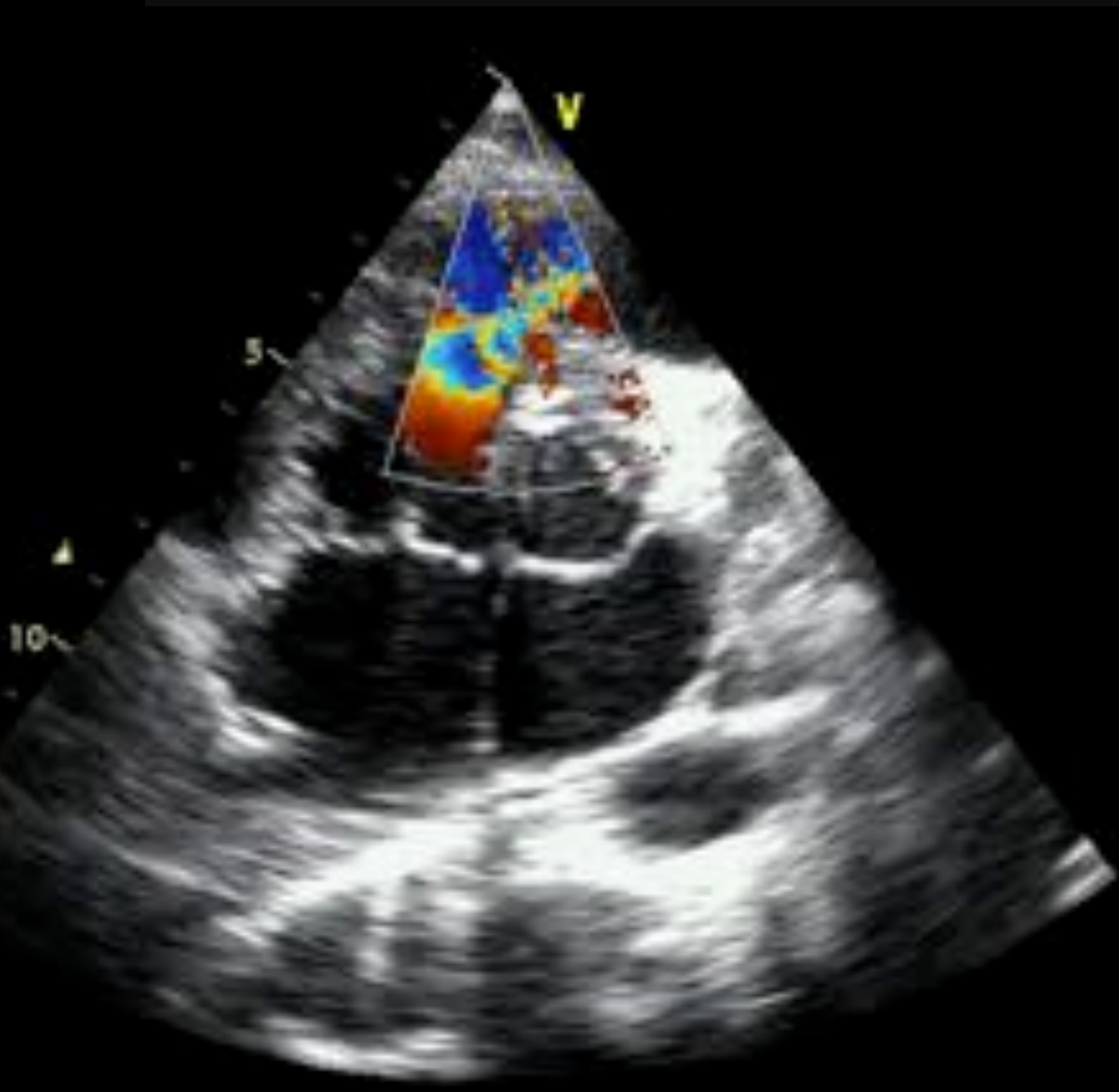


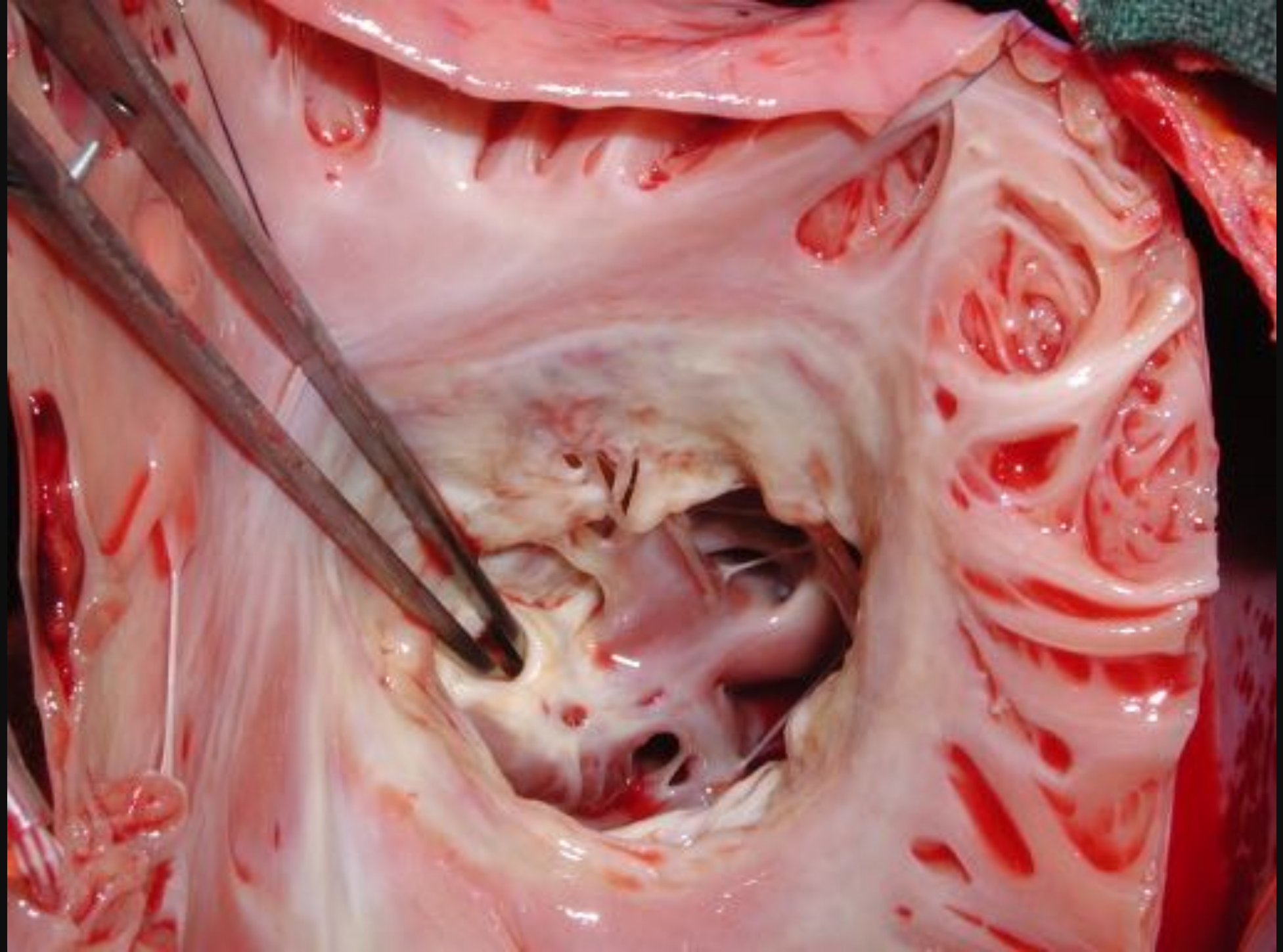
Sténose médio-ventriculaire

Sténose médio-ventriculaire

- Isolée : rare
- Associée à une CIV
  - Sous l'obstacle : Fallot-like
  - Au dessus de l'obstacle : shunt Gauche-Droite
    - Piège échocardiographique


V
5
10

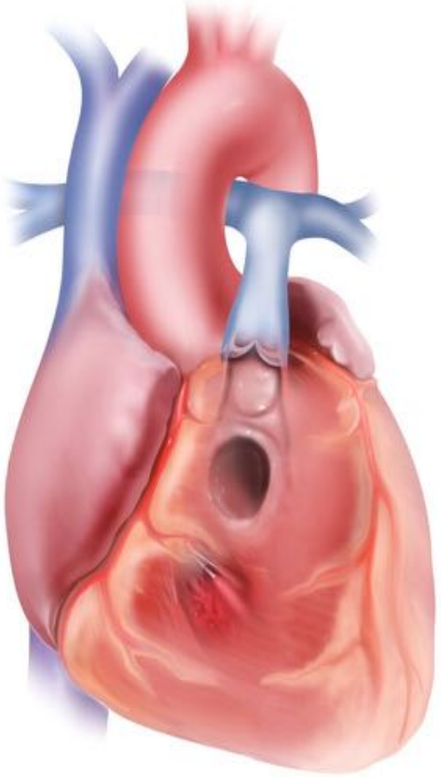

V
5
10

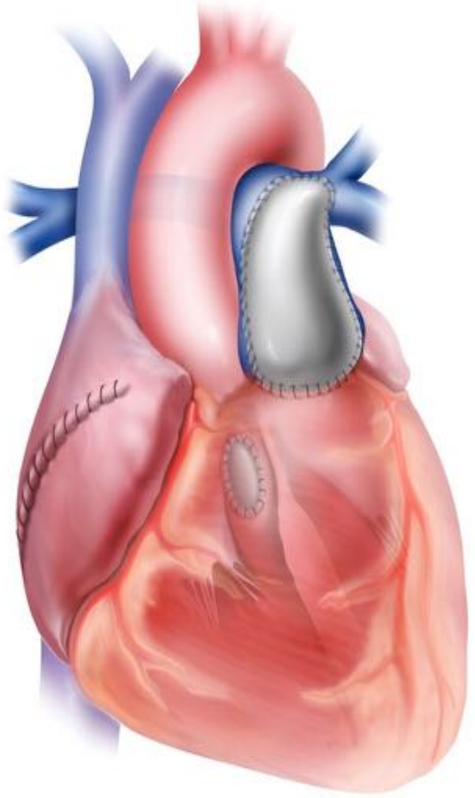


Conduits VD-AP

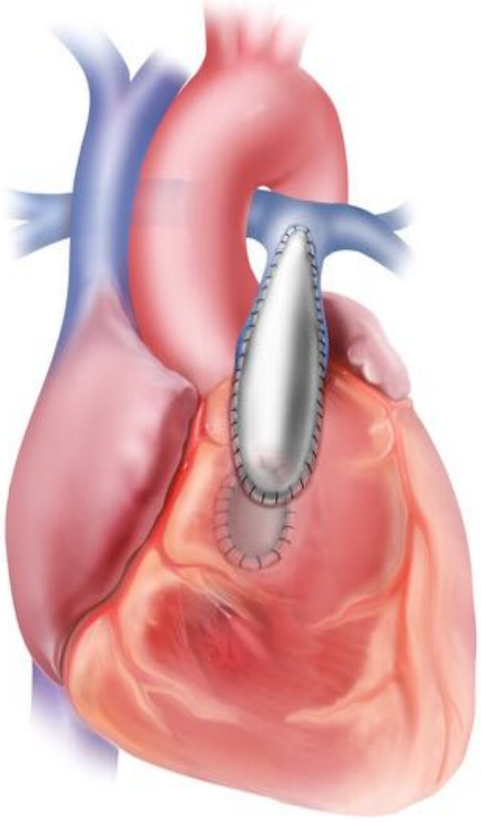
Traitement chirurgical de la tétralogie de Fallot

Réparation



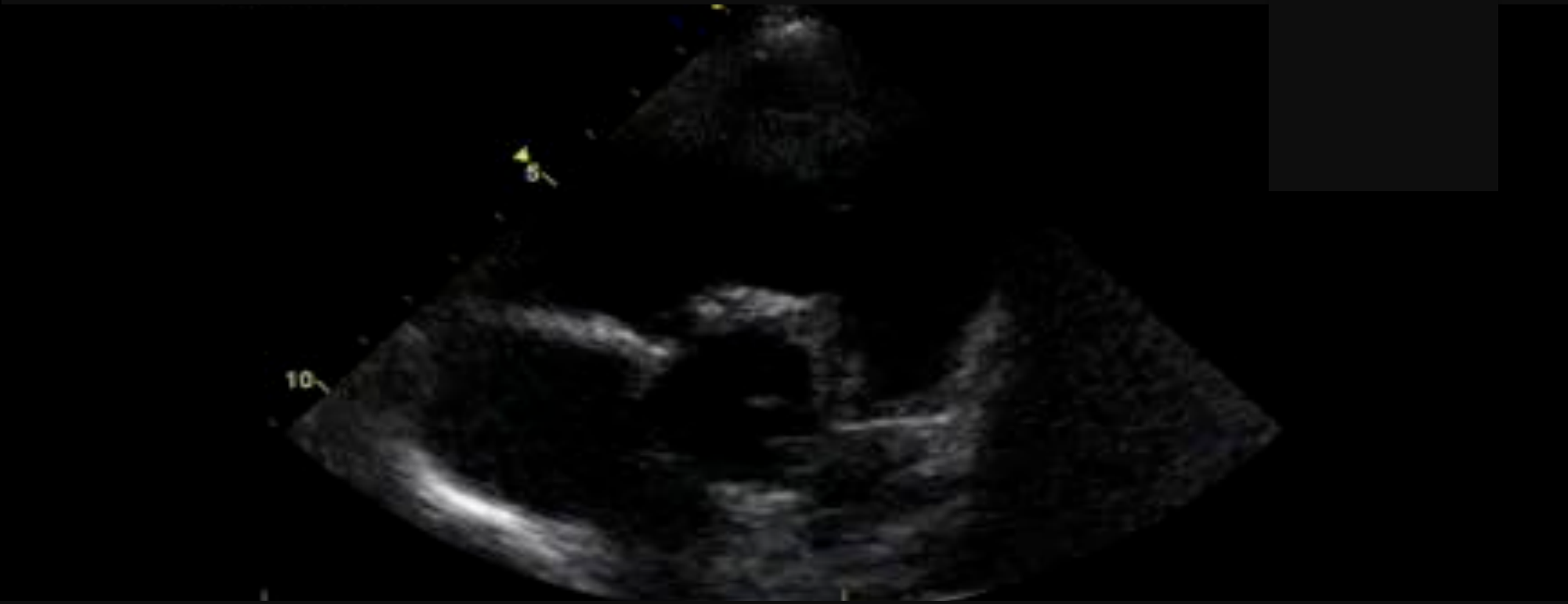


Voix atriale
Patch pulmonaire



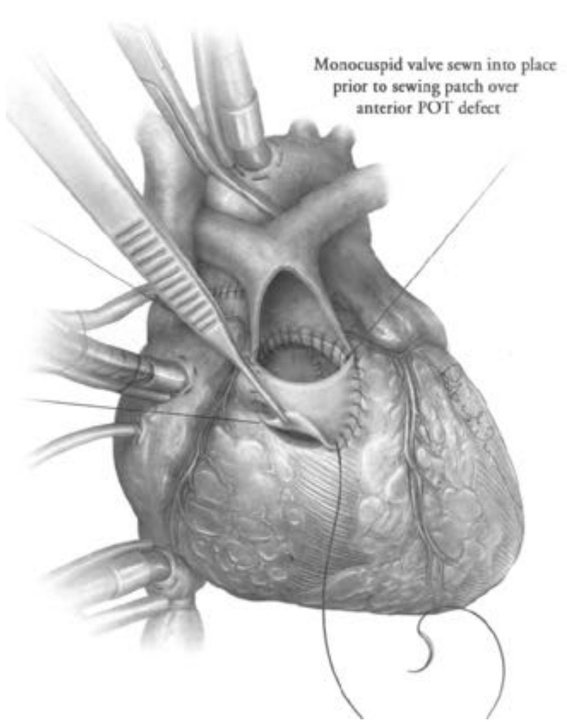
Voix infundibulaire
Patch infundibulo-pulmonaire

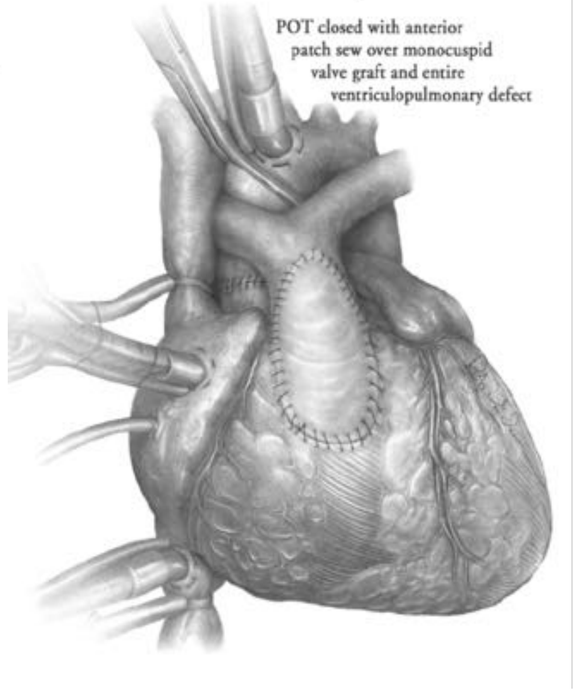
Right ventricle dilatation after Fallot repair with transannular patch

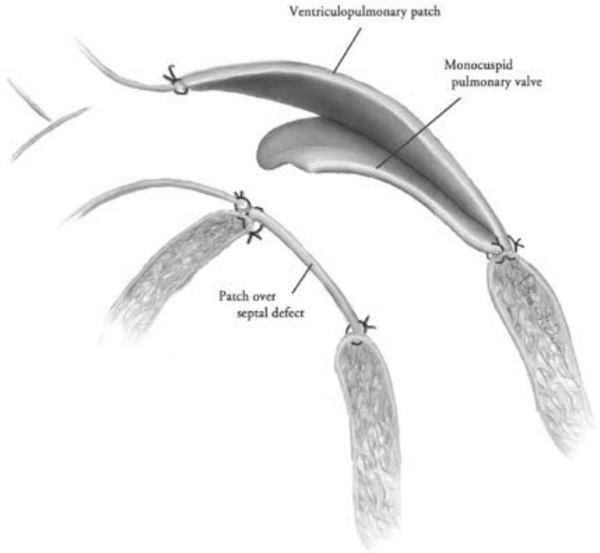


Traitement chirurgical des tétralogies de Fallot

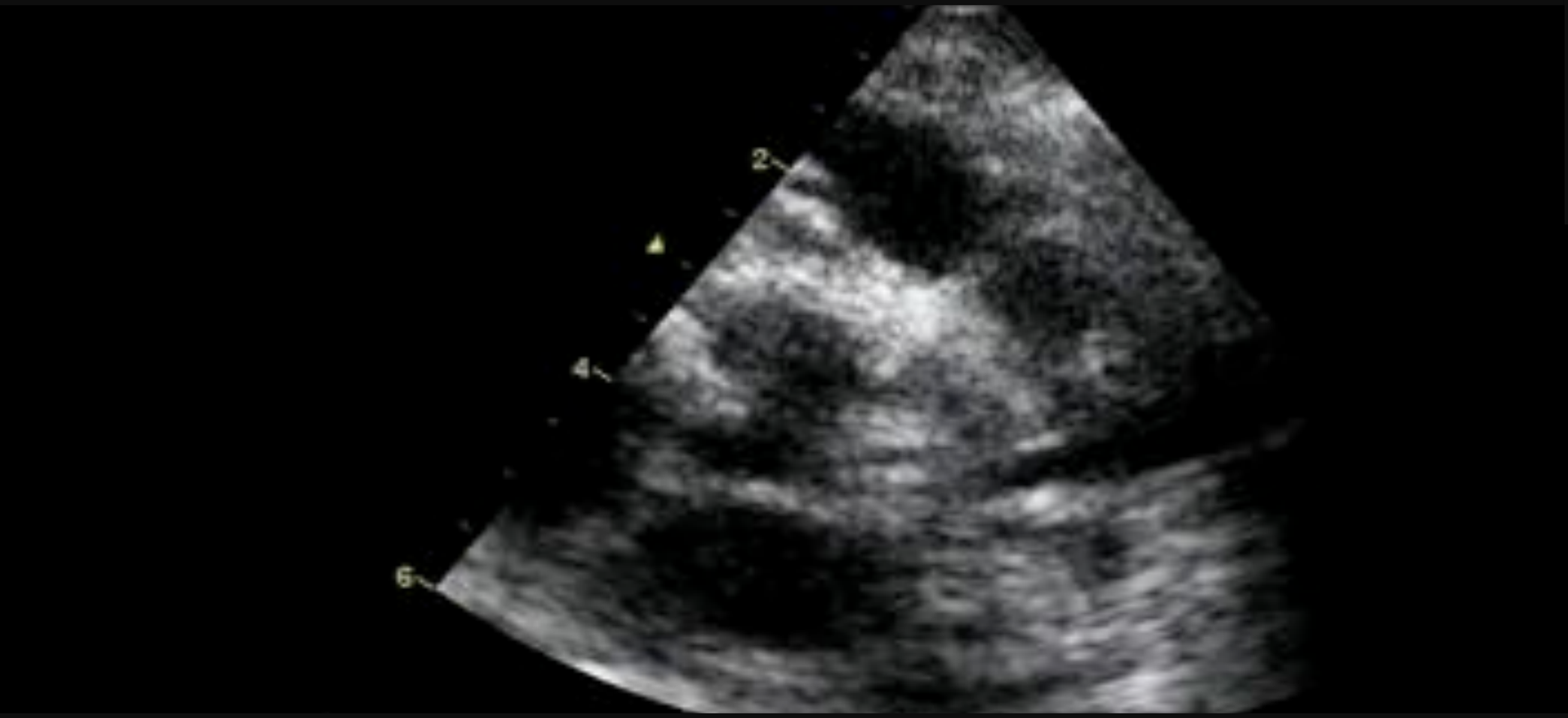
Fallot + Agénésie des valves ou petites branches : « Monocusp »



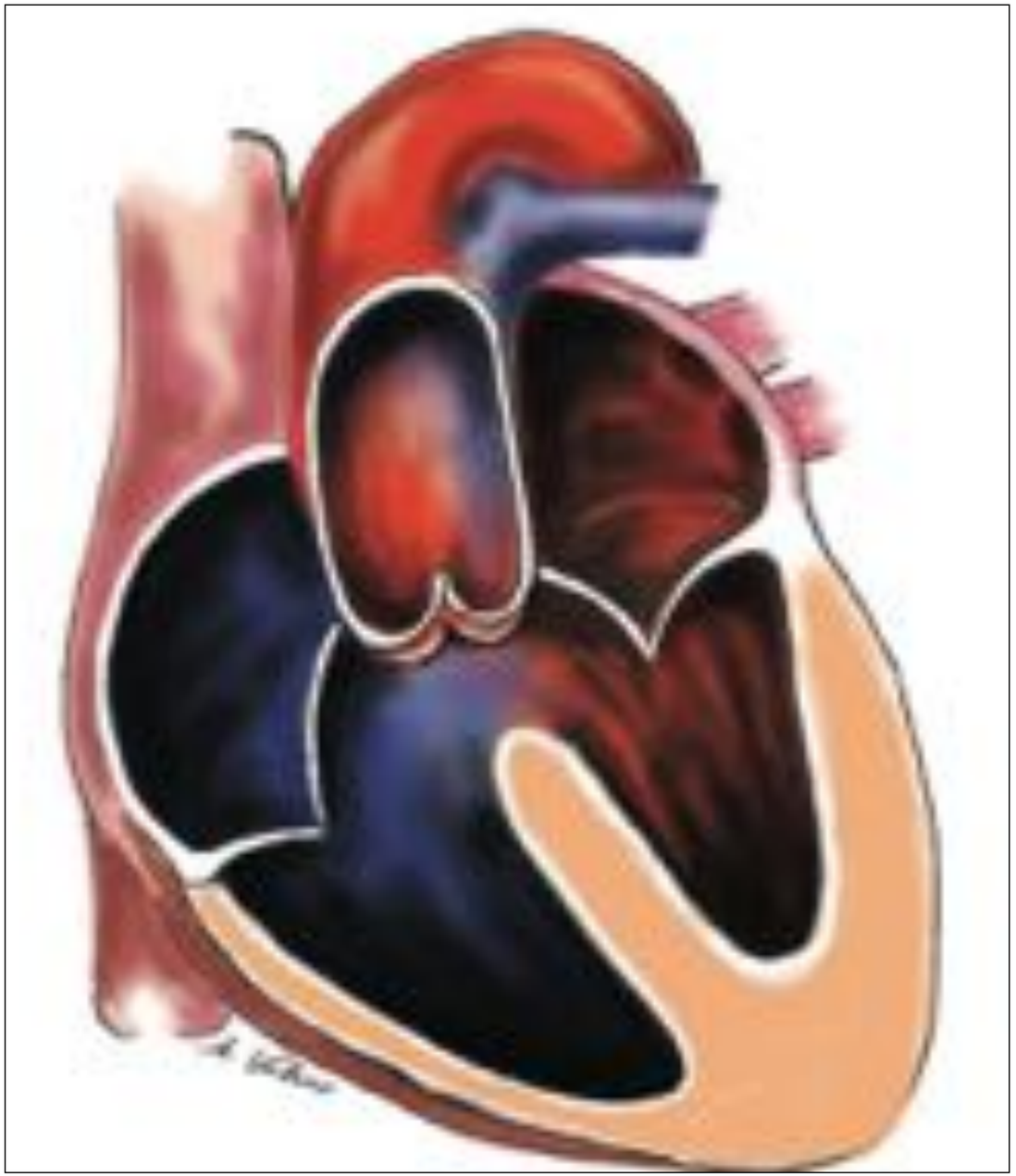


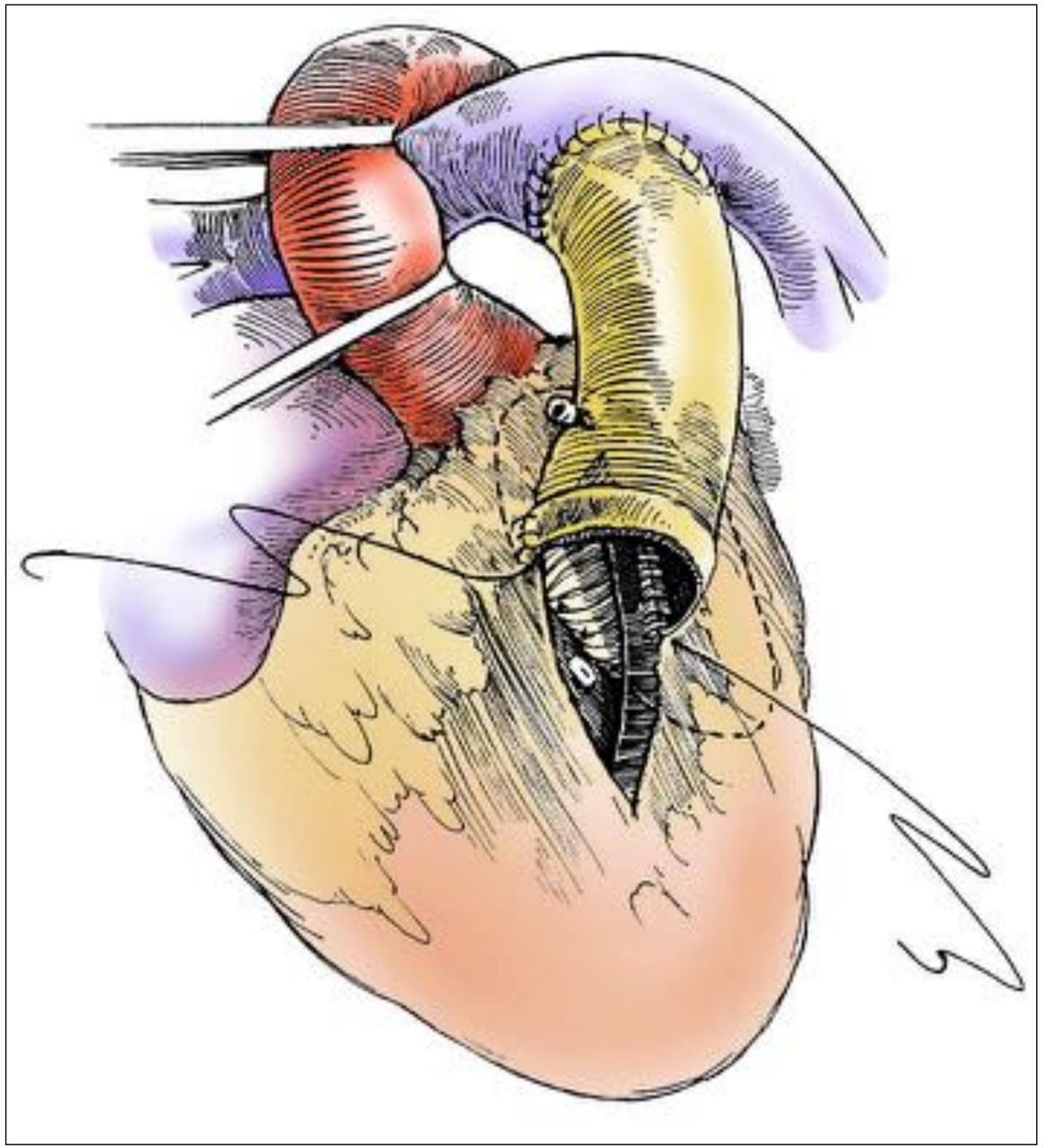


Pulmonary mono-cusp

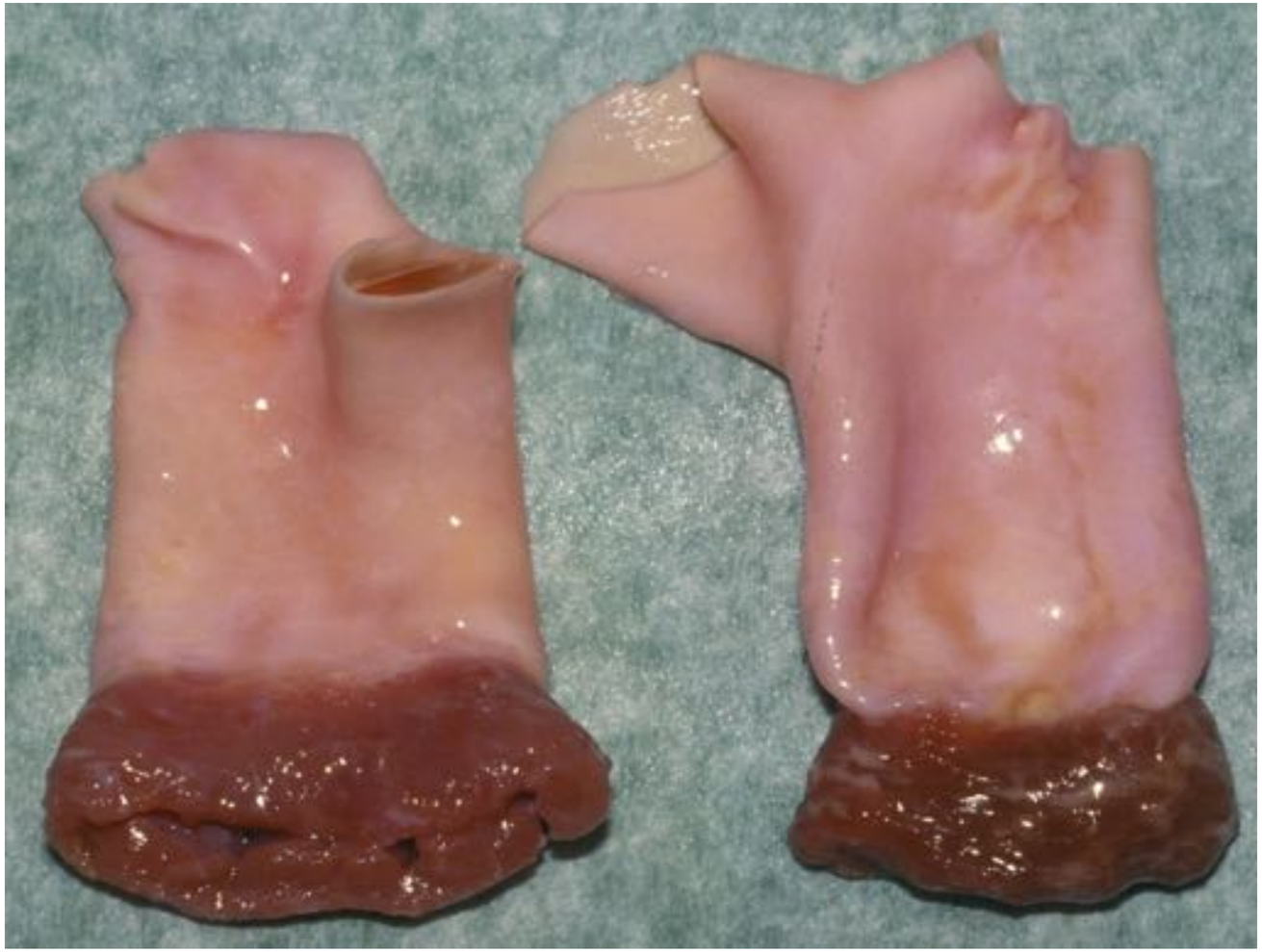


Les réparations avec des conduits valvés ou non valvés

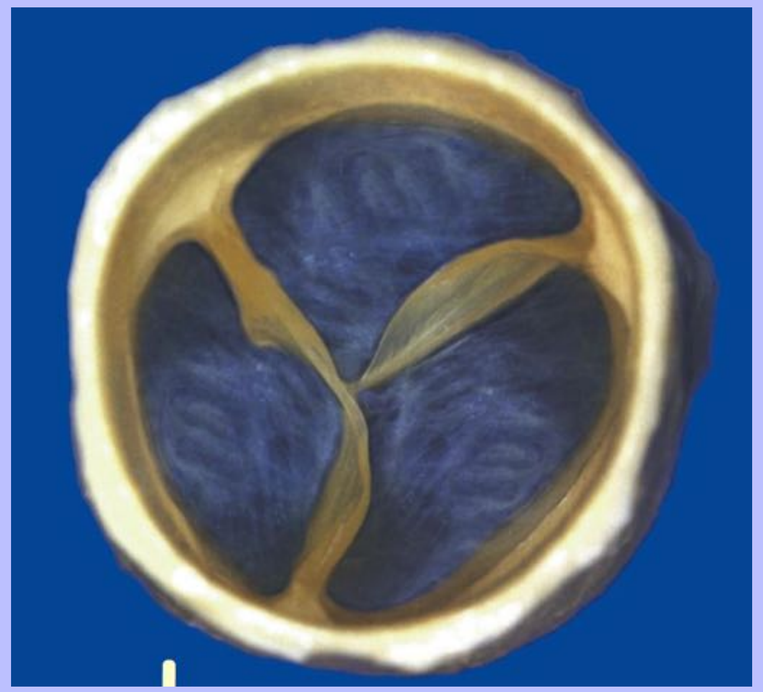


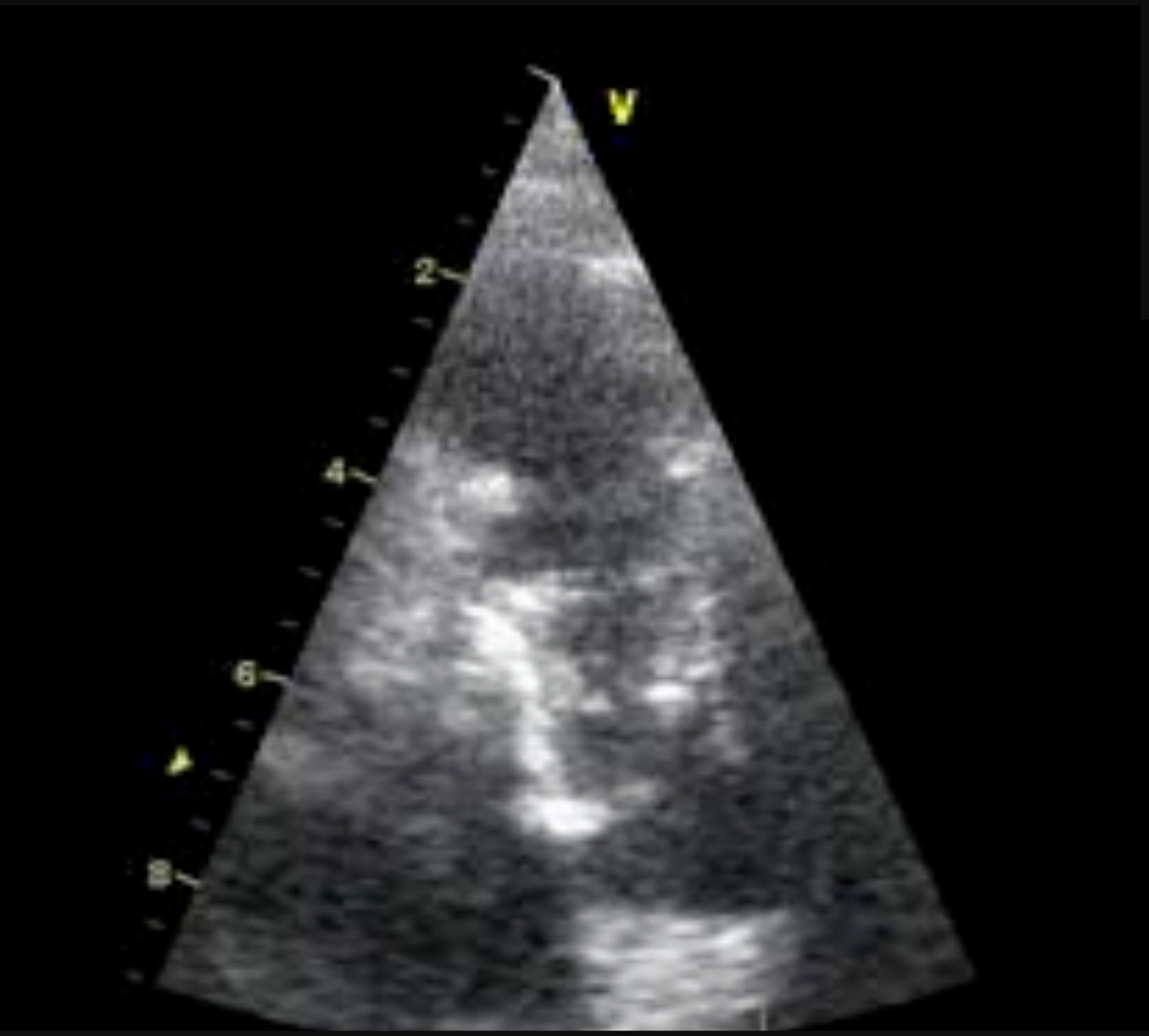


Homogreffe pulmonaire

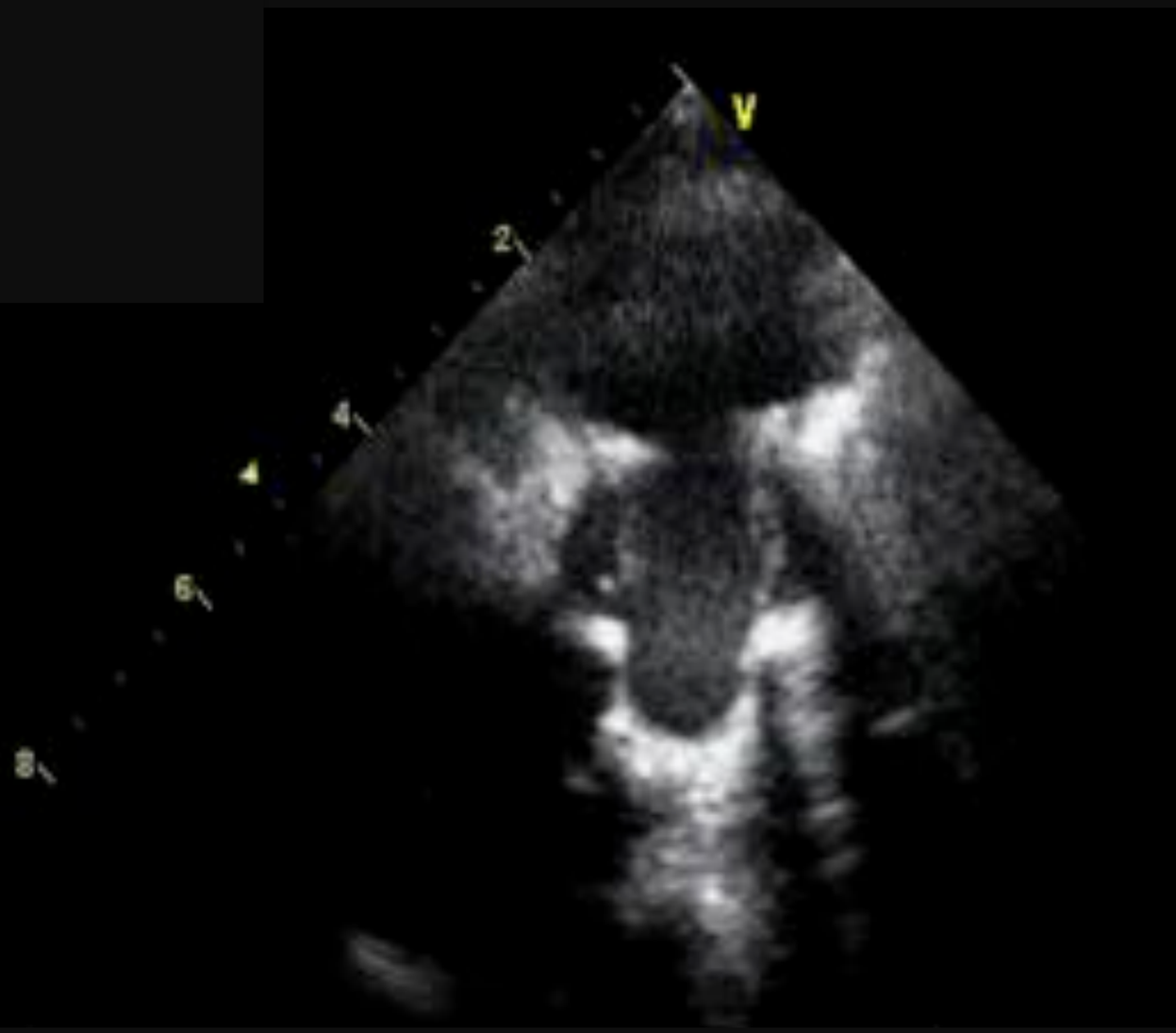


Tube Contegra - Venpro



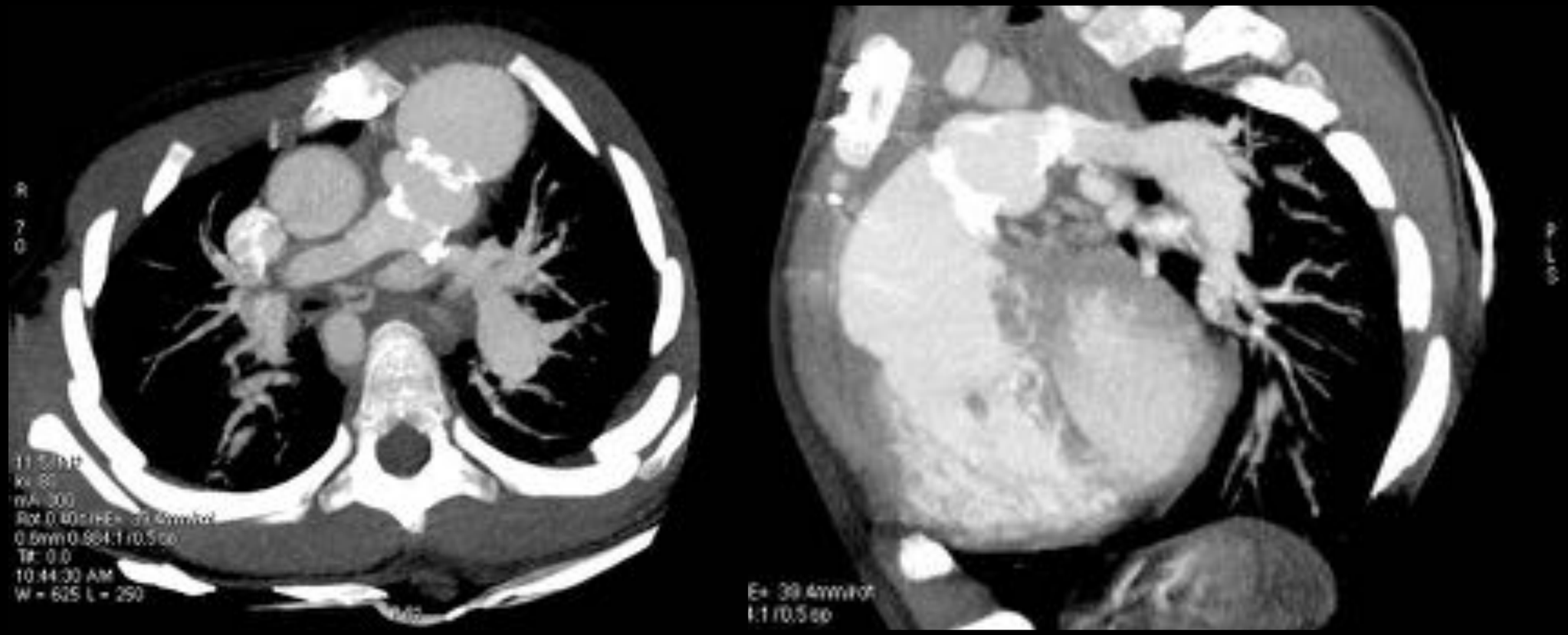


Labcor



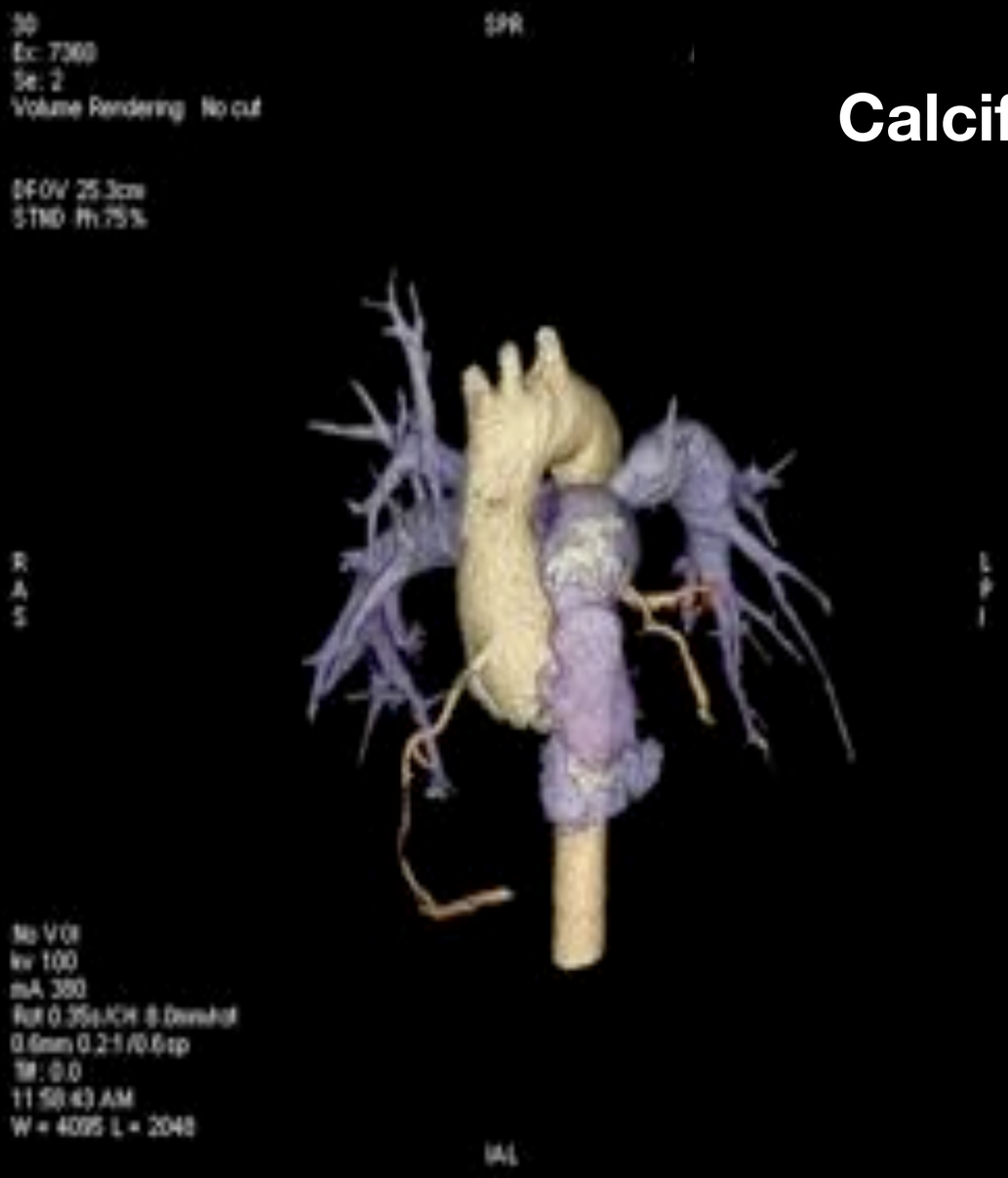
VenPro

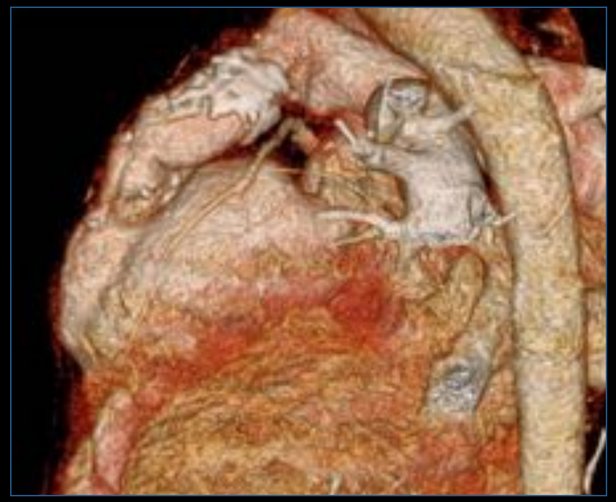
Calcified pulmonary conduit

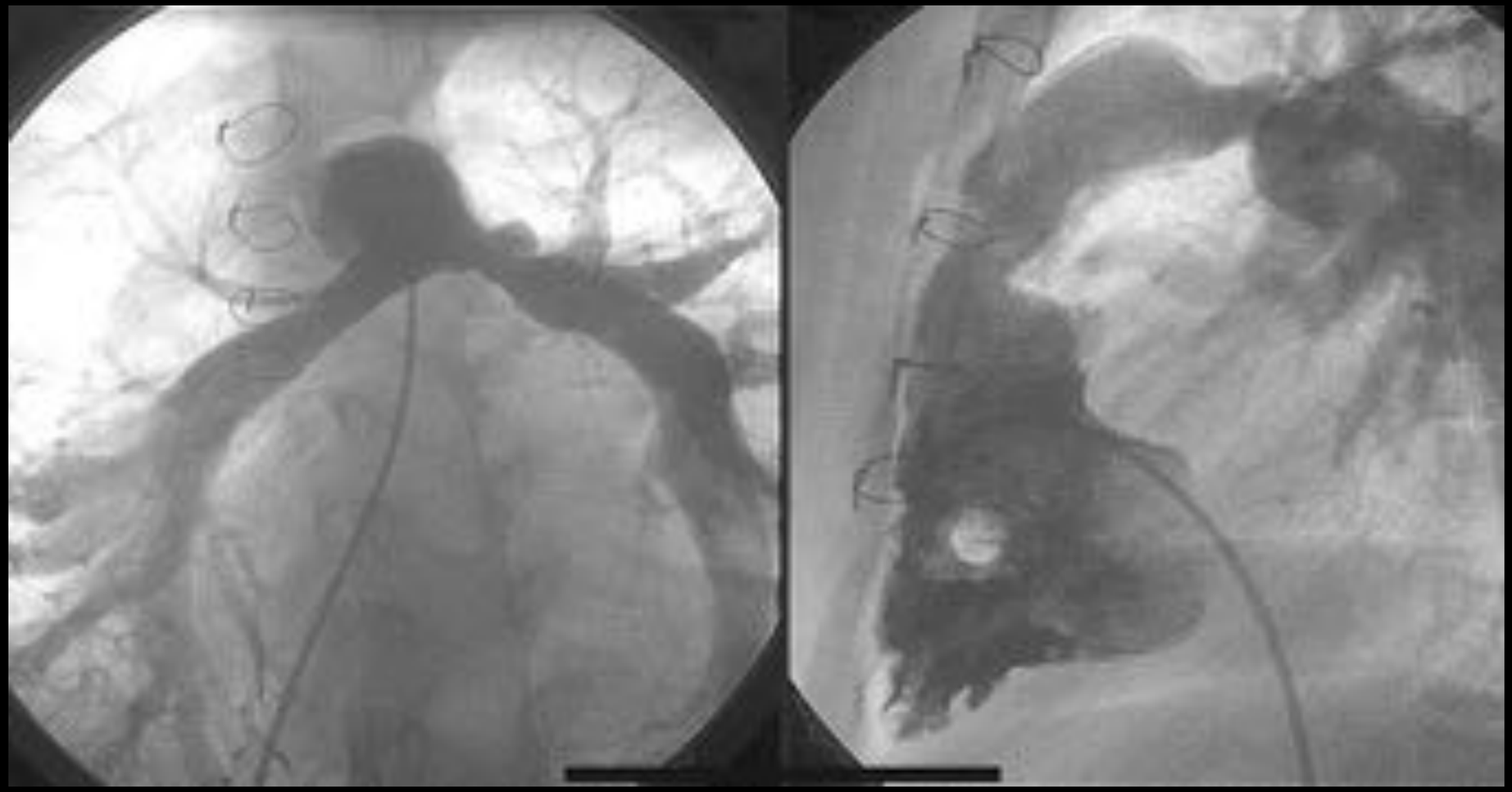


Calcified pulmonary conduit

Calcified pulmonary conduit



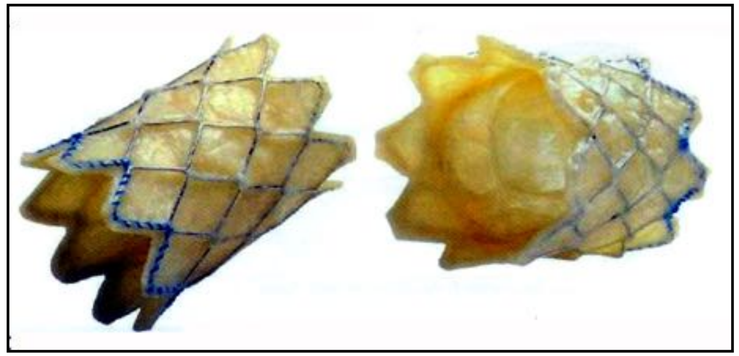




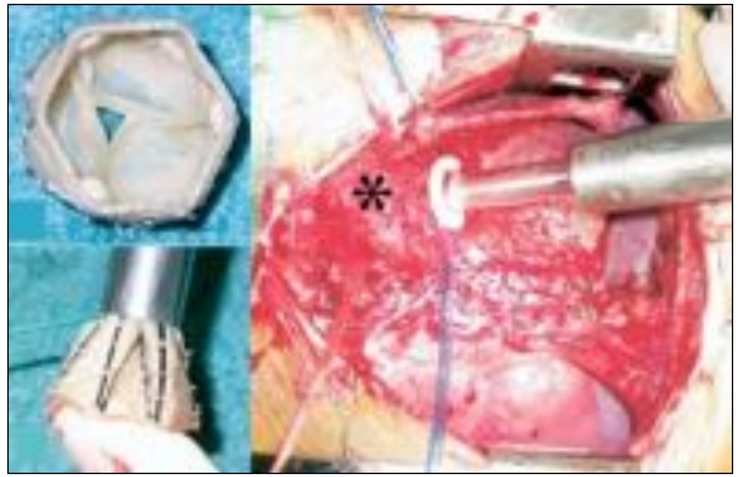
Calcified pulmonary conduit

Valves pulmonaires

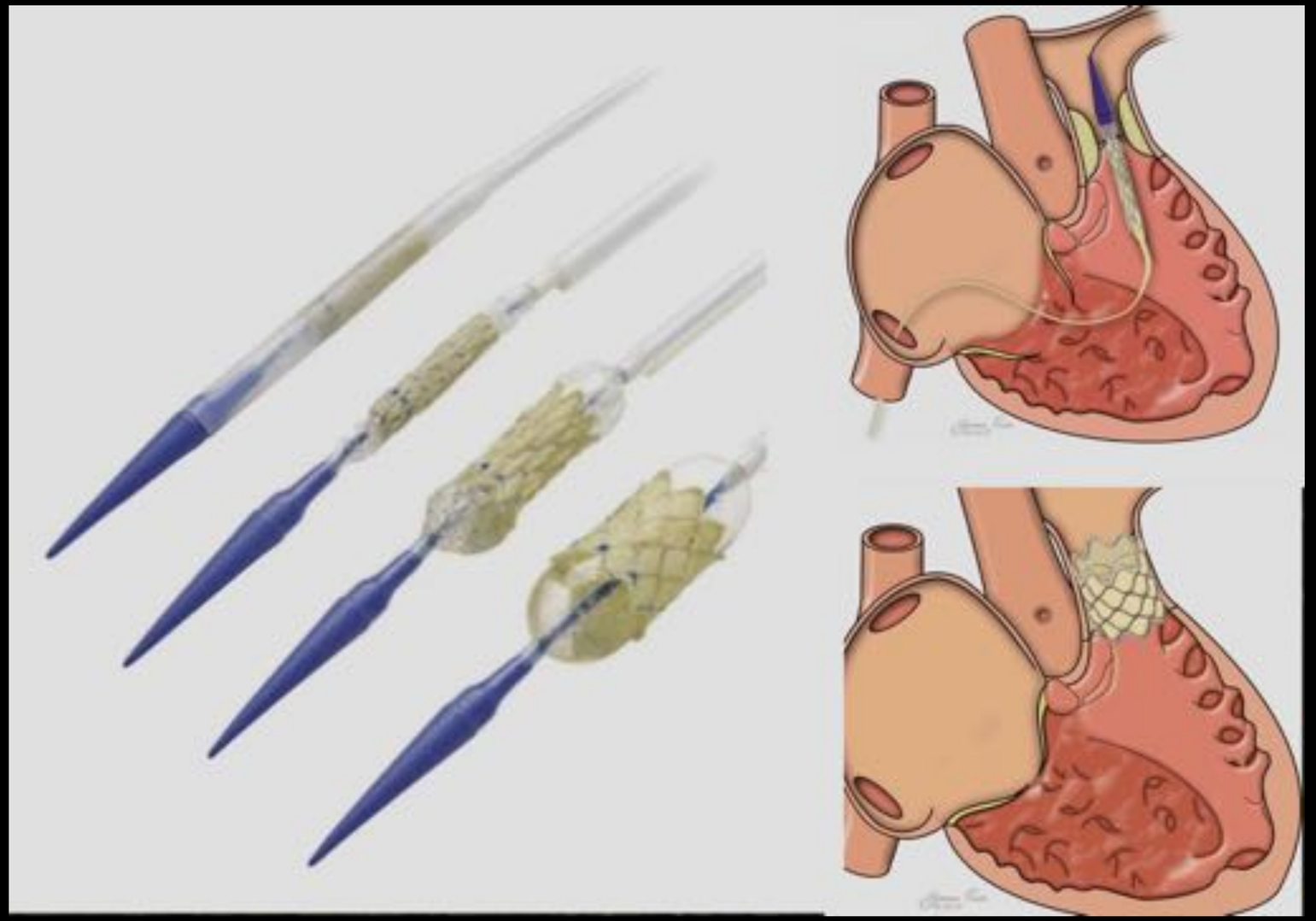
Valve pulmonaire
«Melody»



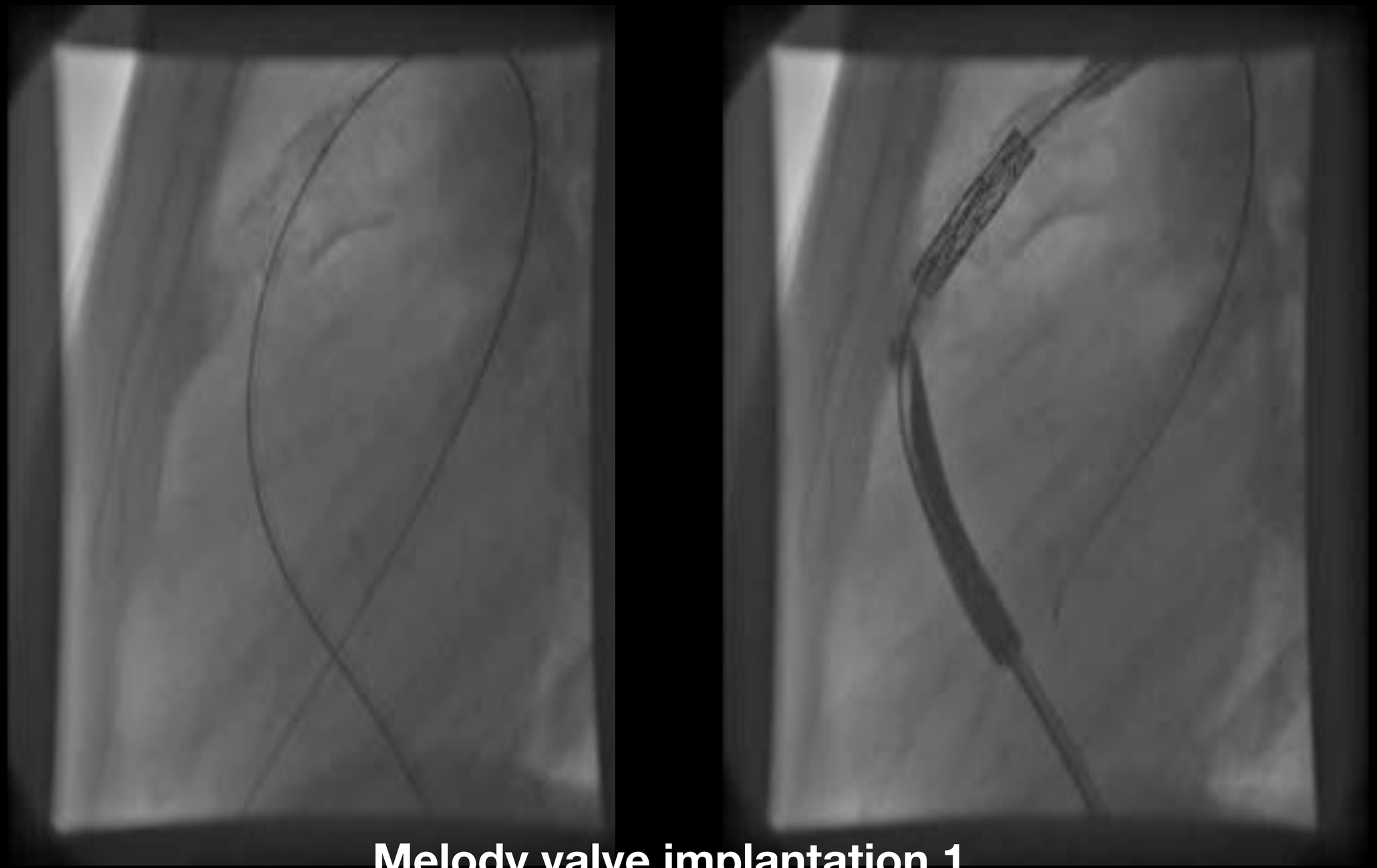
Valve injectable
«Shelhigh»



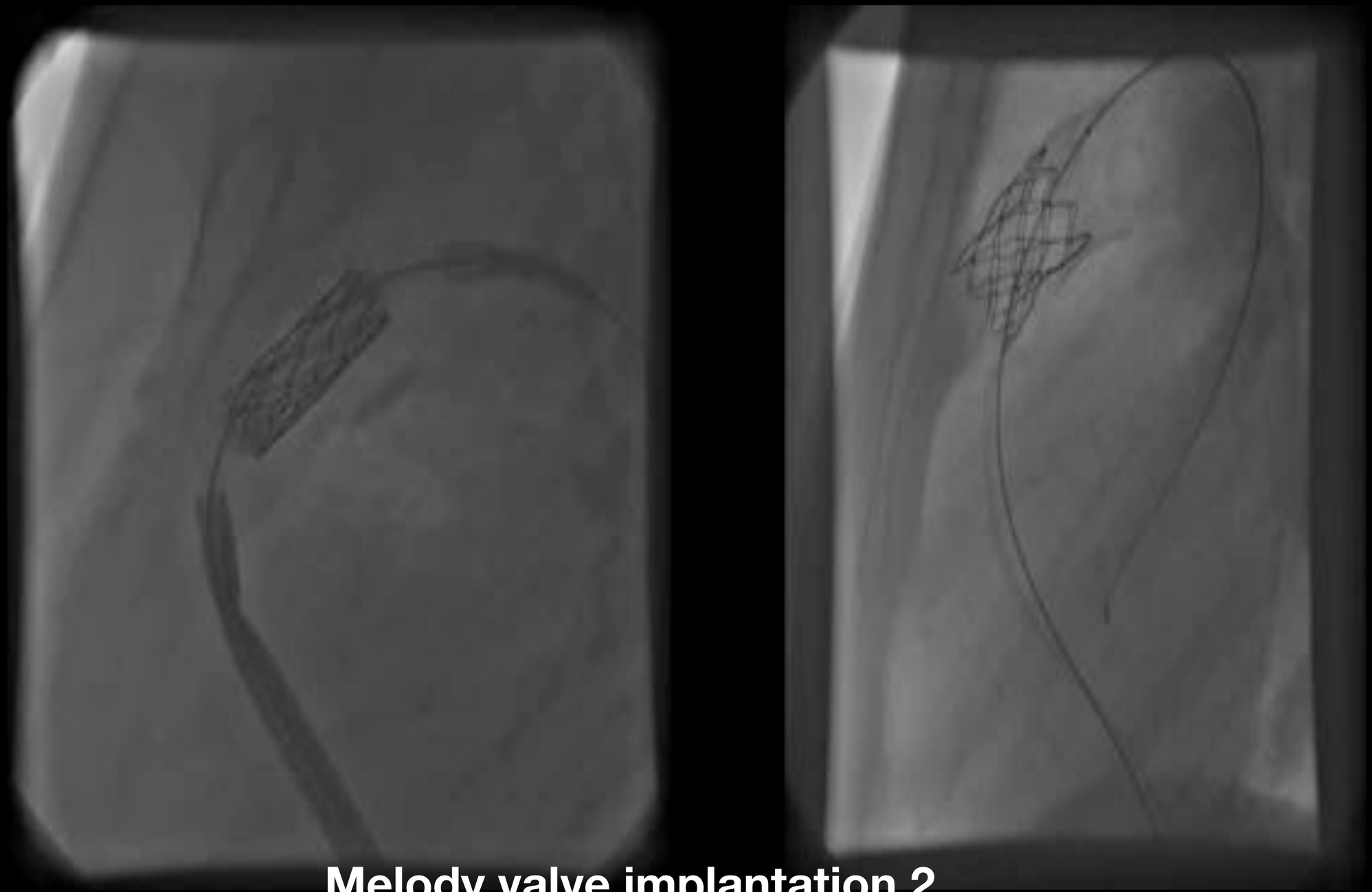
Schreiber. J Thorac Cardiovasc Surg 2006



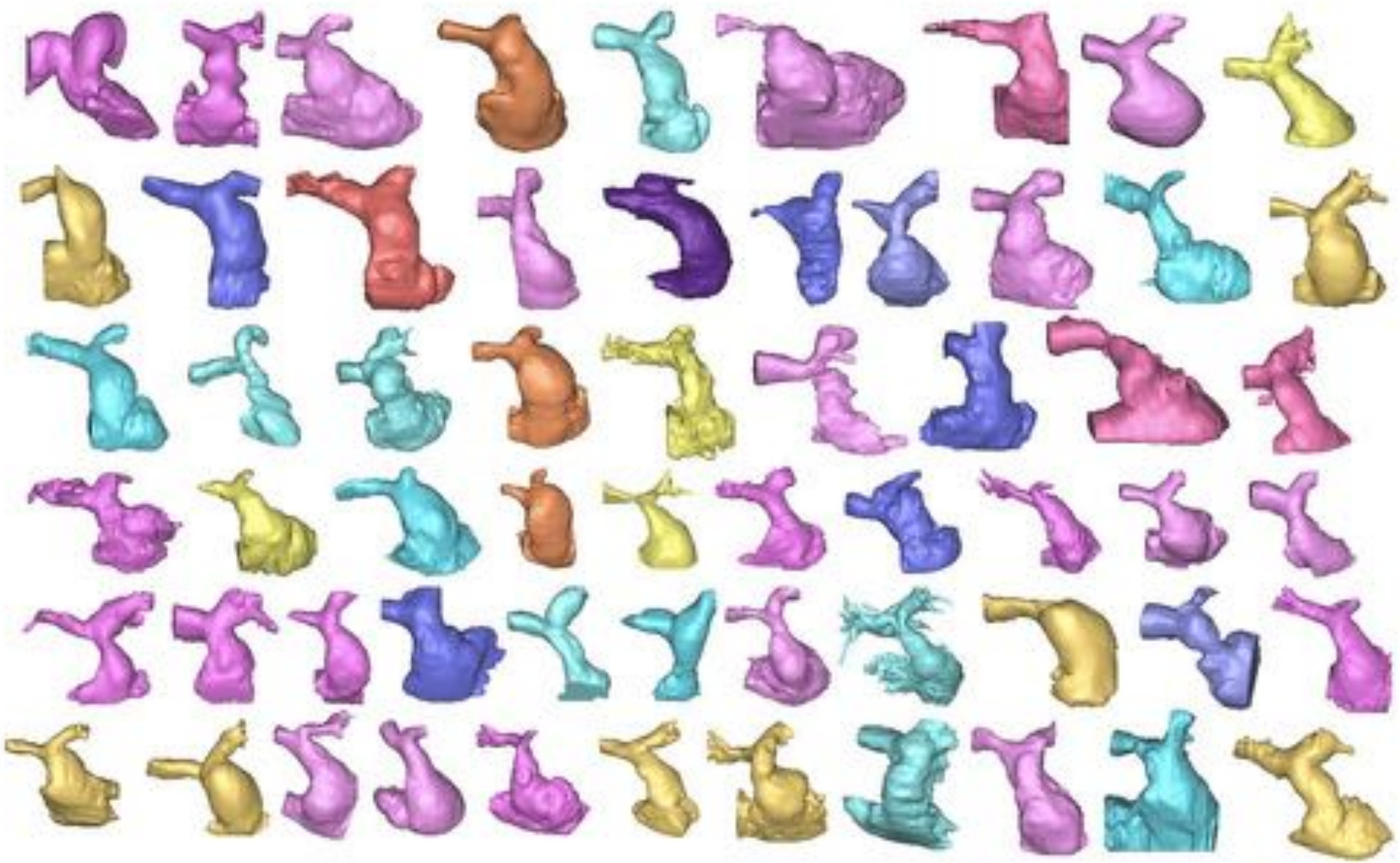
Melody valve implantation - movie



Melody valve implantation 1



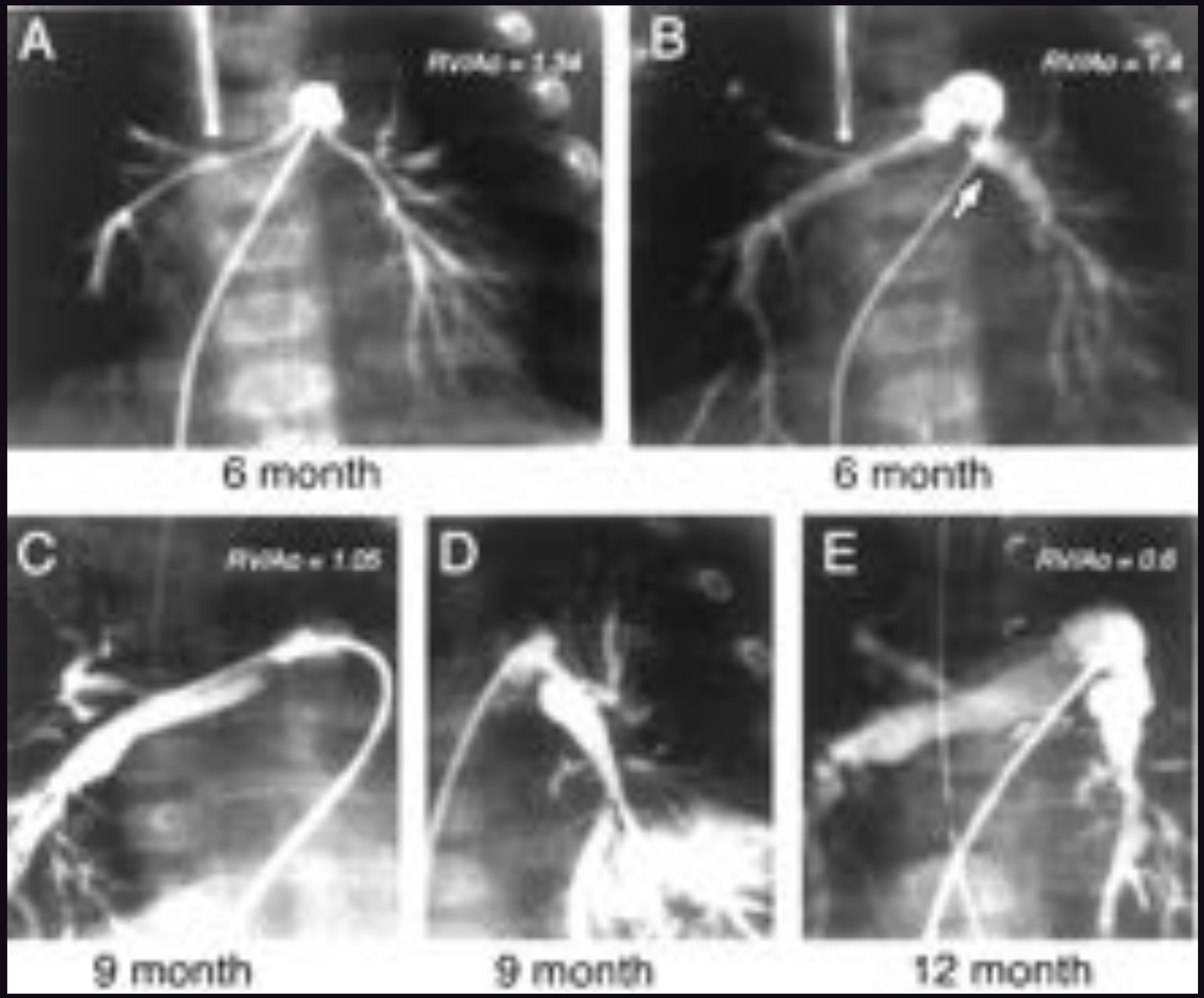
Melody valve implantation 2



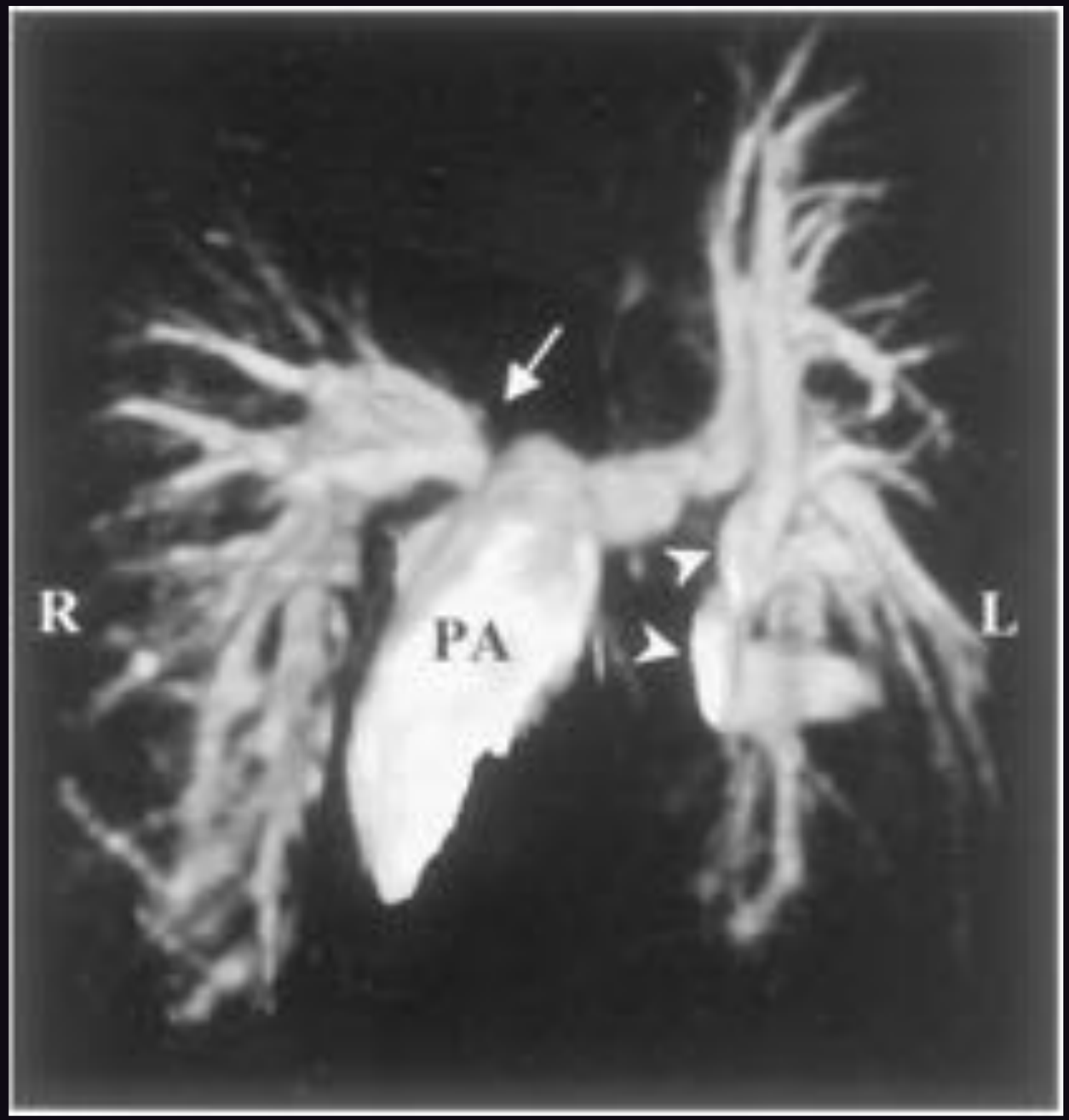
Capelli et al. 2011

Sténoses
des branches pulmonaires

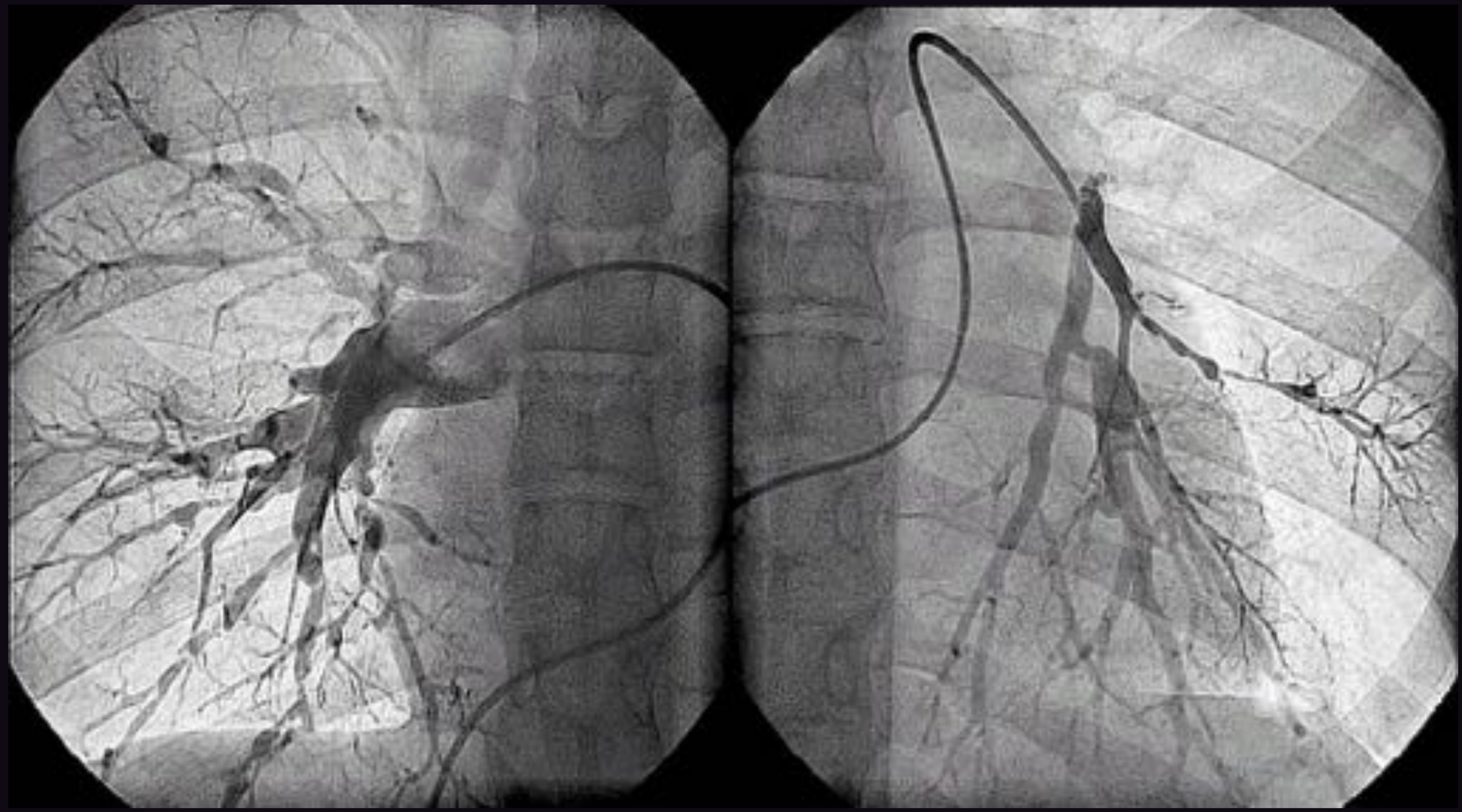
Congénitale



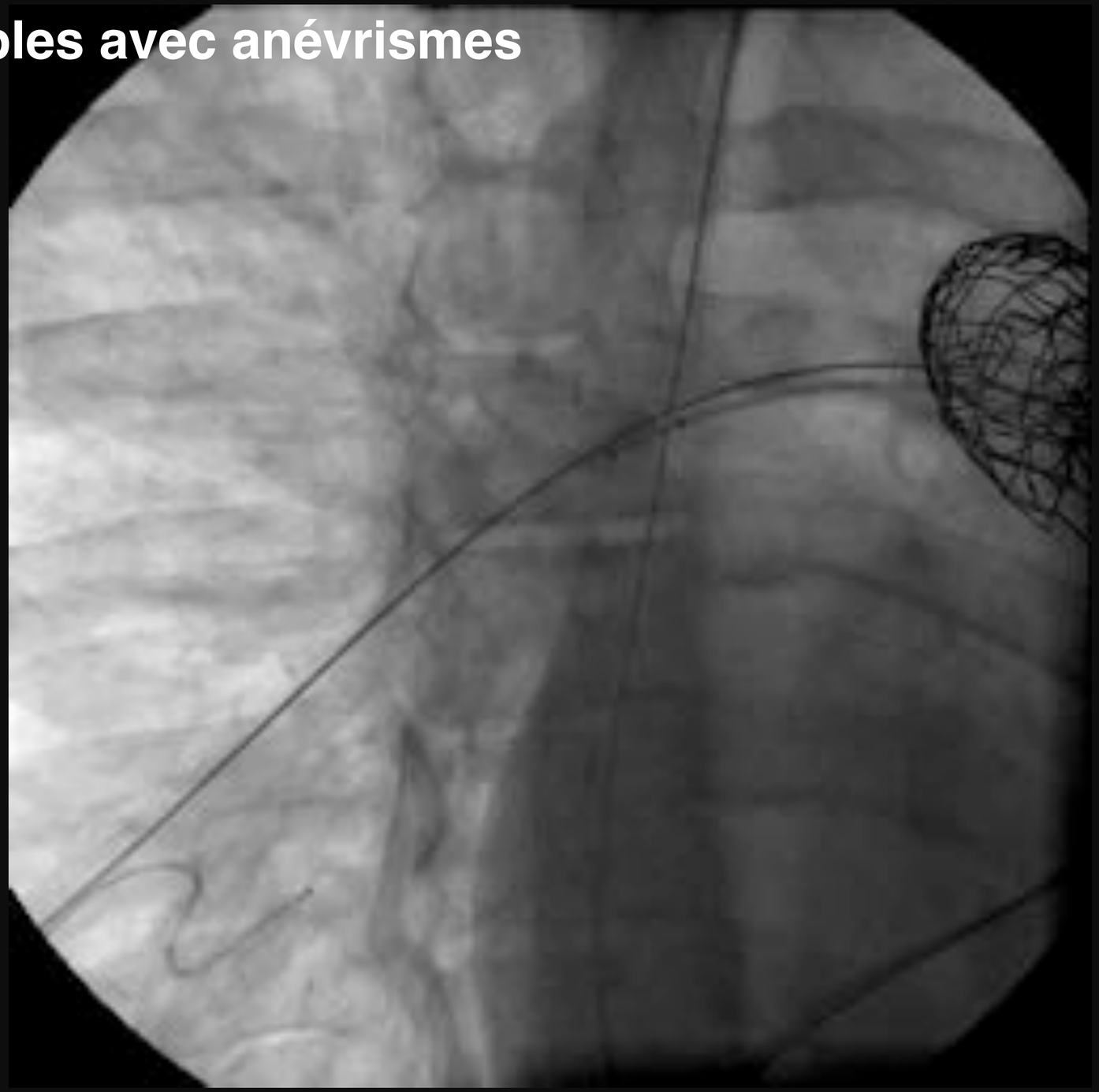
Postopératoire



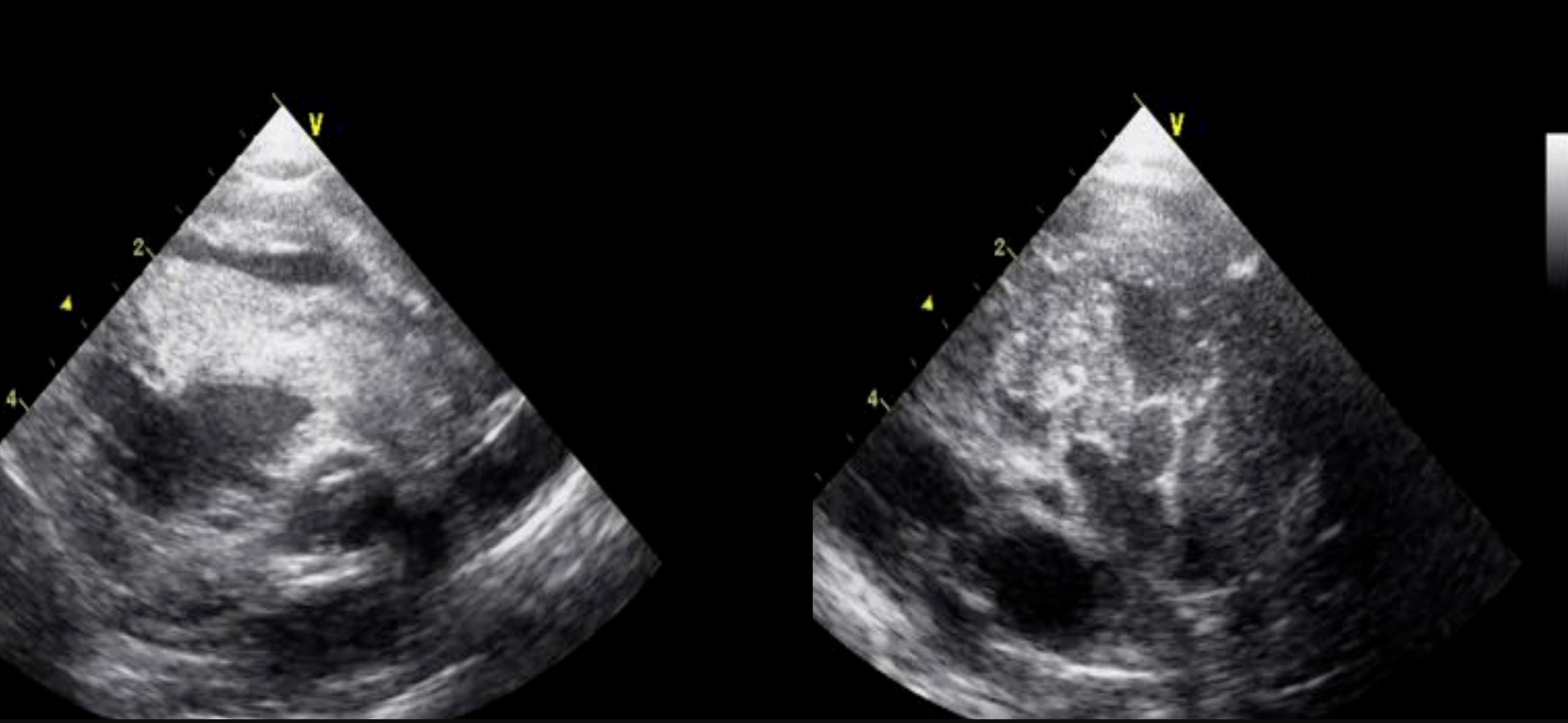
Diffuses multiples et distales



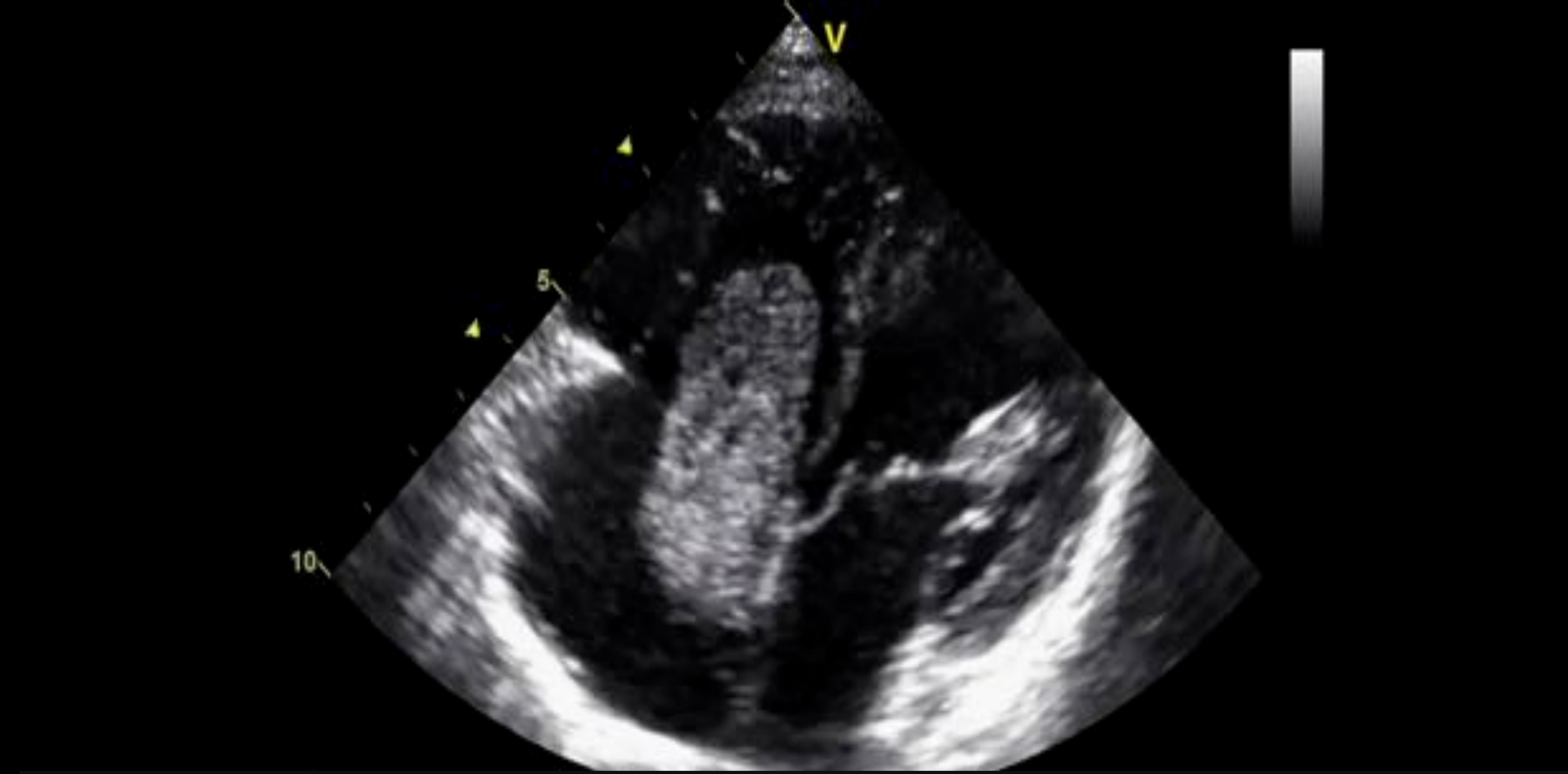
Diffuses multiples avec anévrismes



Obstacles droits inhabituels

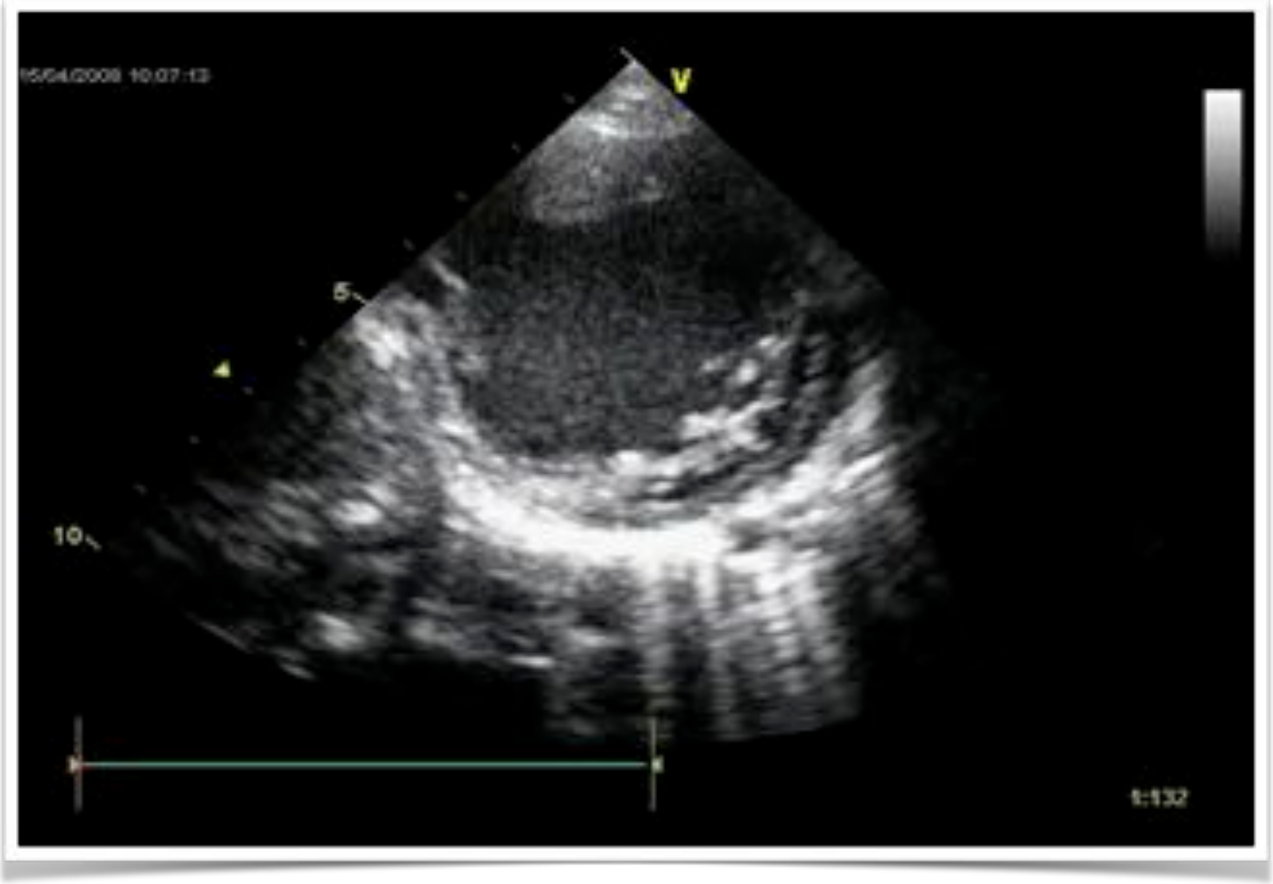


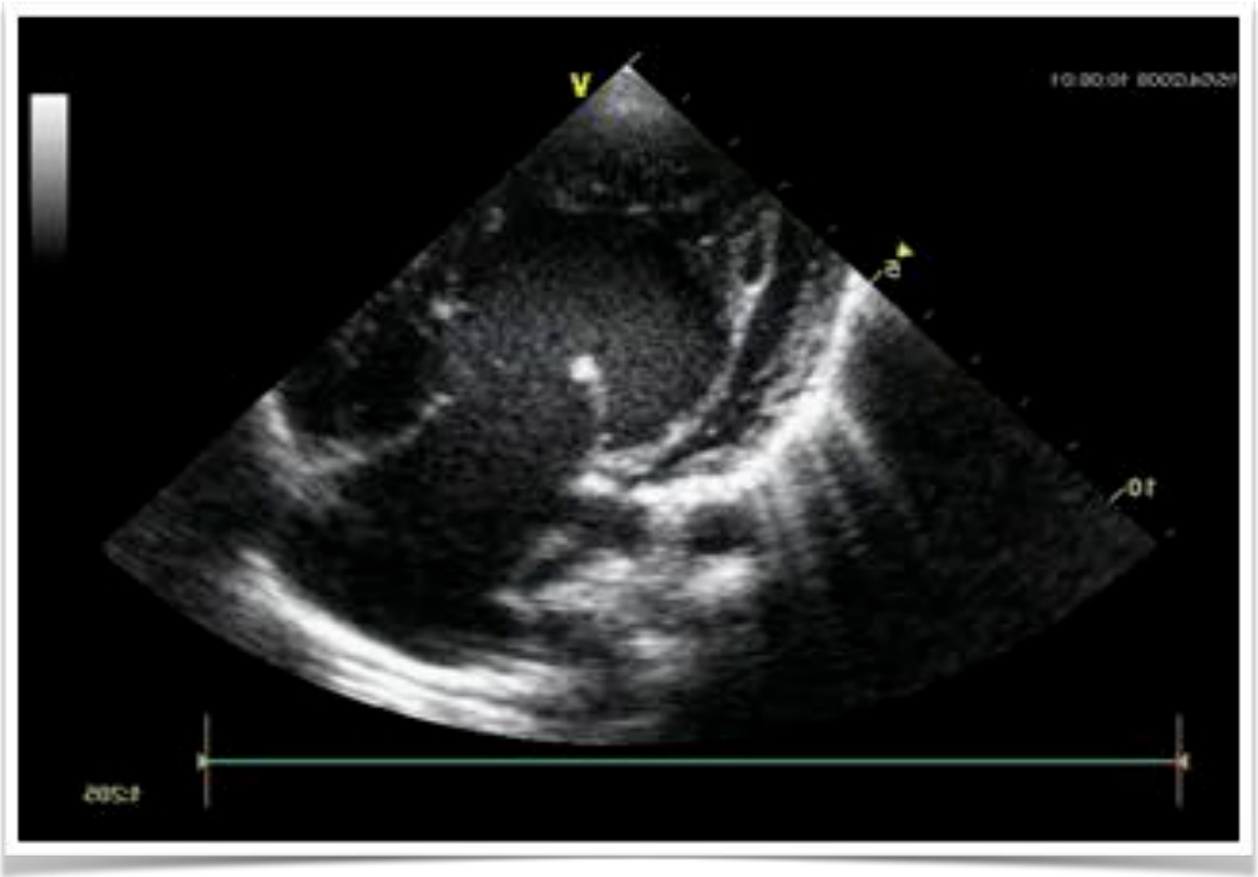
Right ventricular teratoma



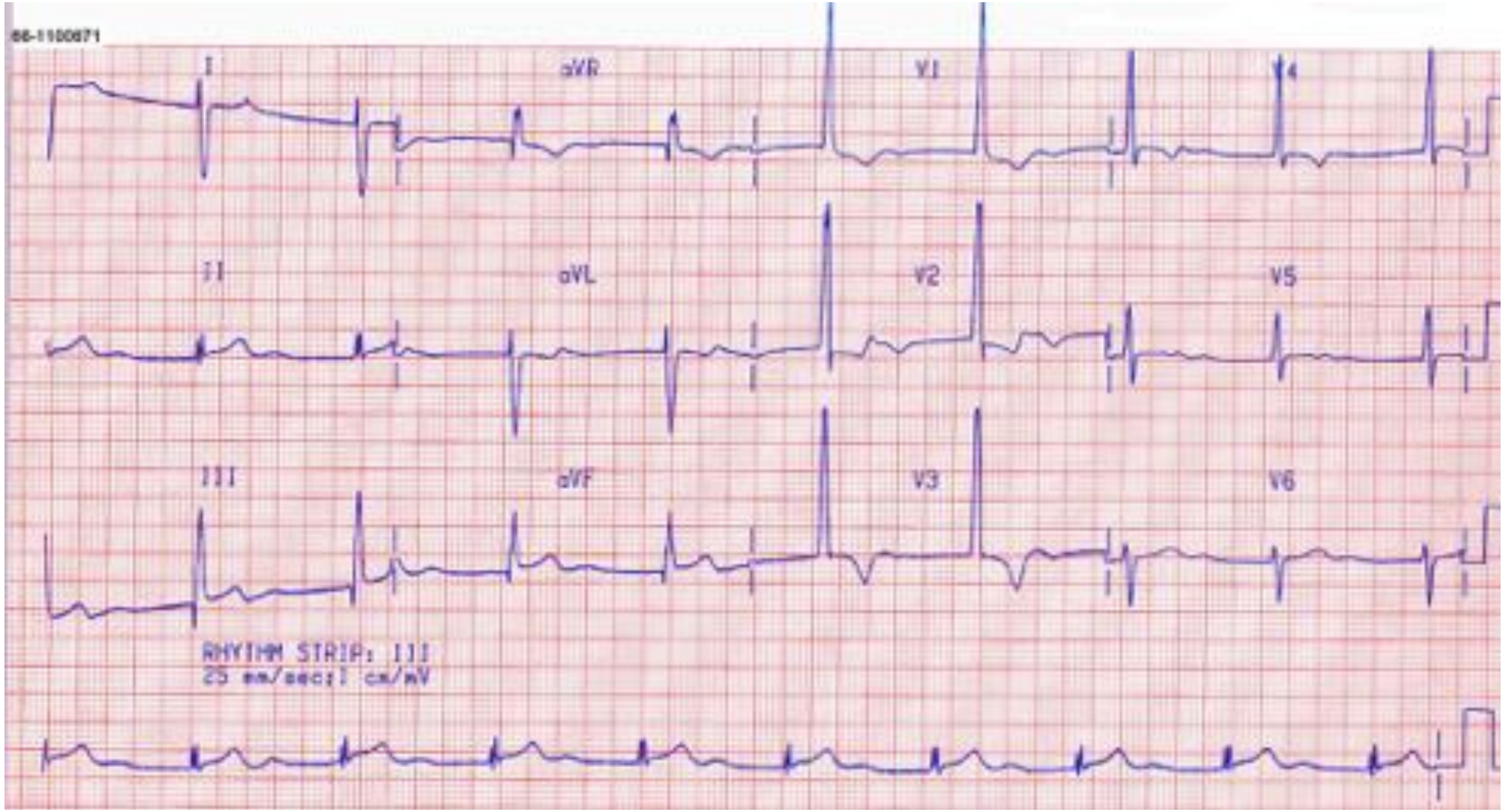
Caval and right atrial extension of a nephroblastoma

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.

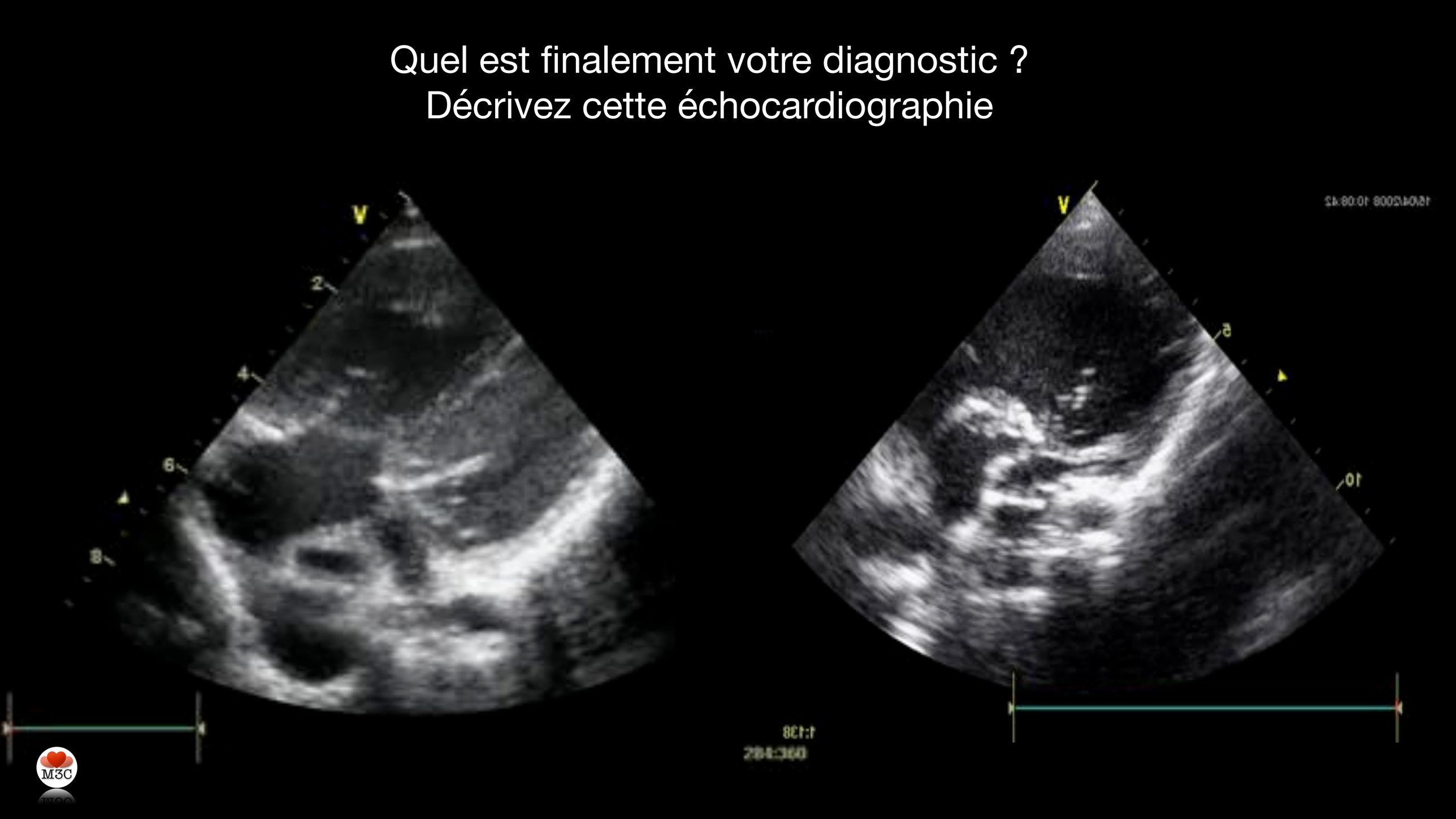




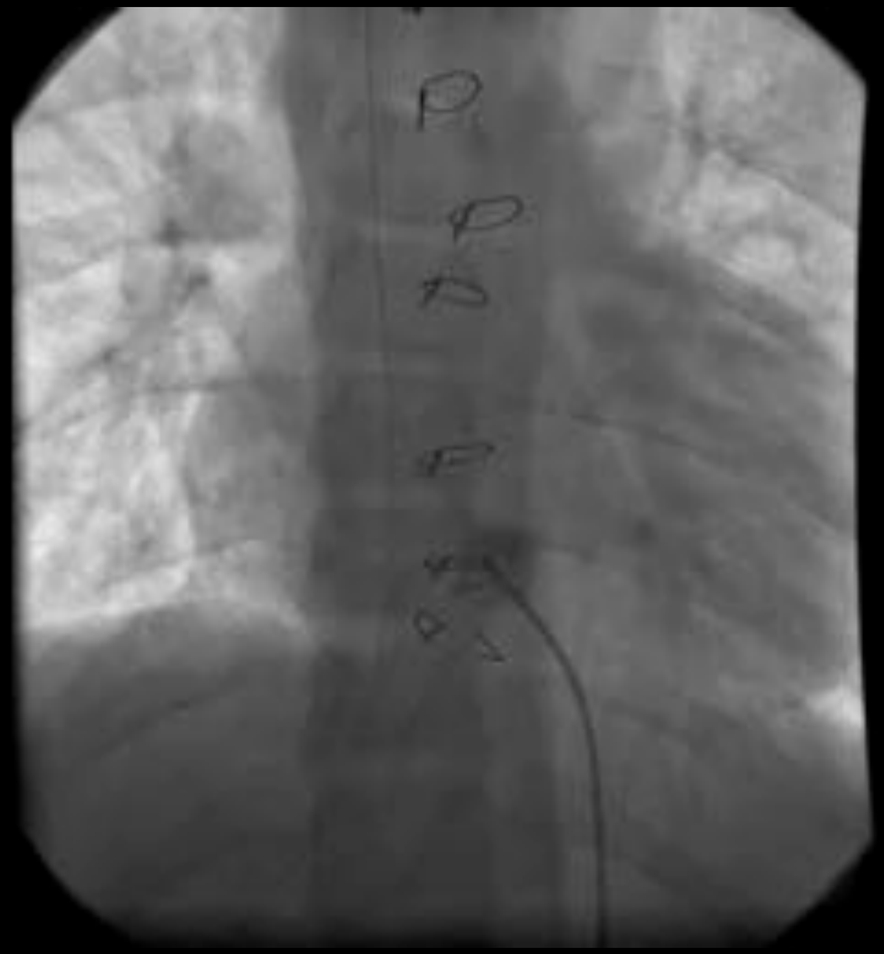
Faites vous ou confirmez vous votre diagnostic sur cet ECG ?

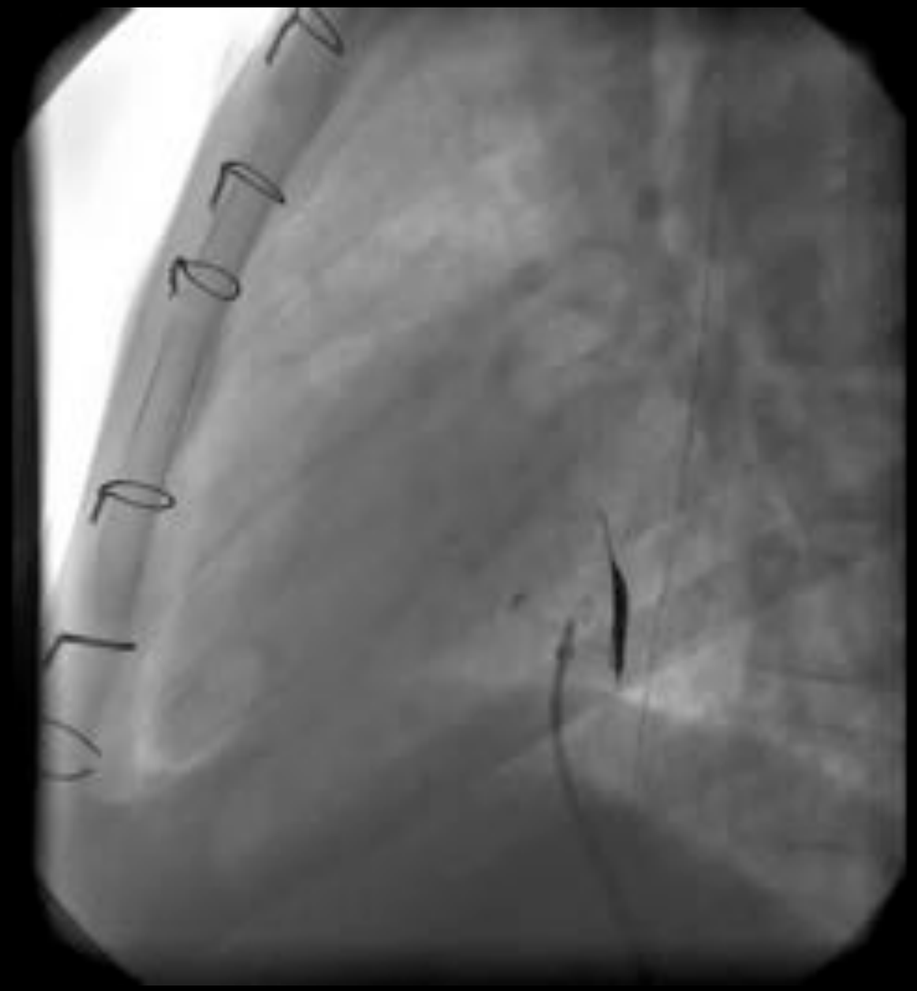


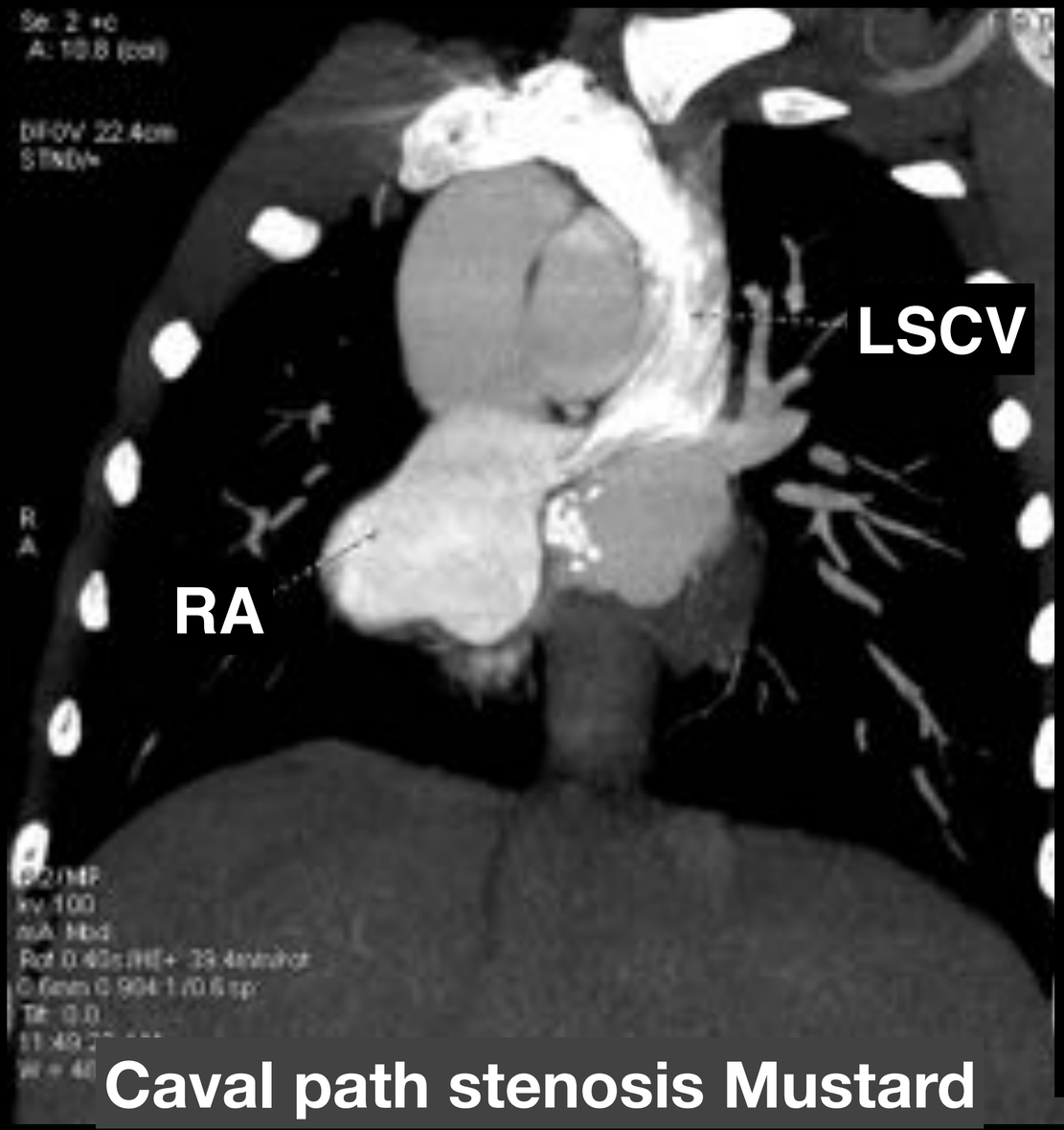
Quel est finalement votre diagnostic ?
Décrivez cette échocardiographie



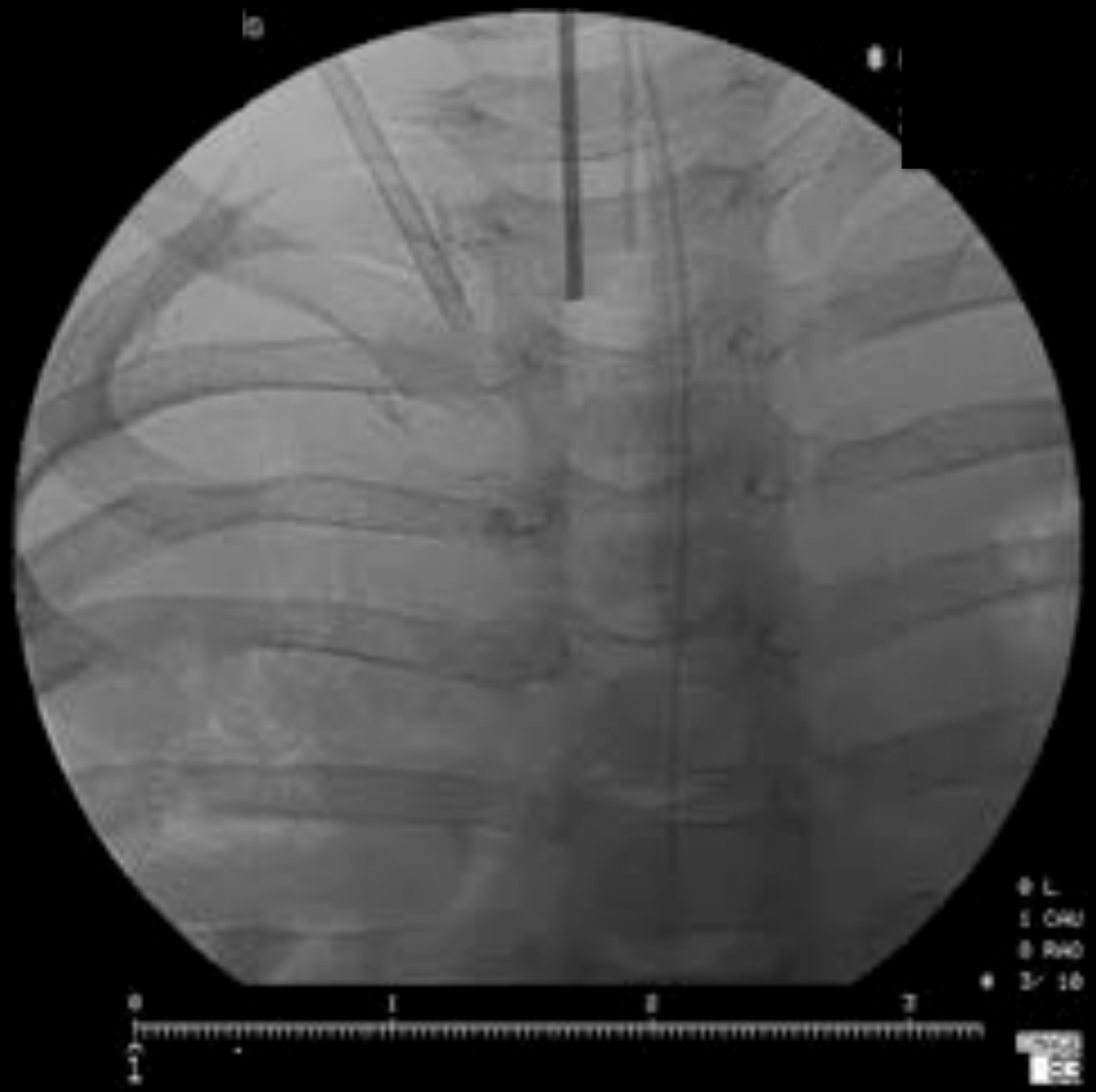
Vous faites un cathétérisme cardiaque car vous ne comprenez pas les symptômes de la patiente chez qui vous avez trouvé une lame d'ascite et une hépatomégalie d'allure cardiaque. Interprétez ces angiographies







Caval path stenosis Mustard



Right Superior Caval Vein stenosis