Transposition of the great arteries

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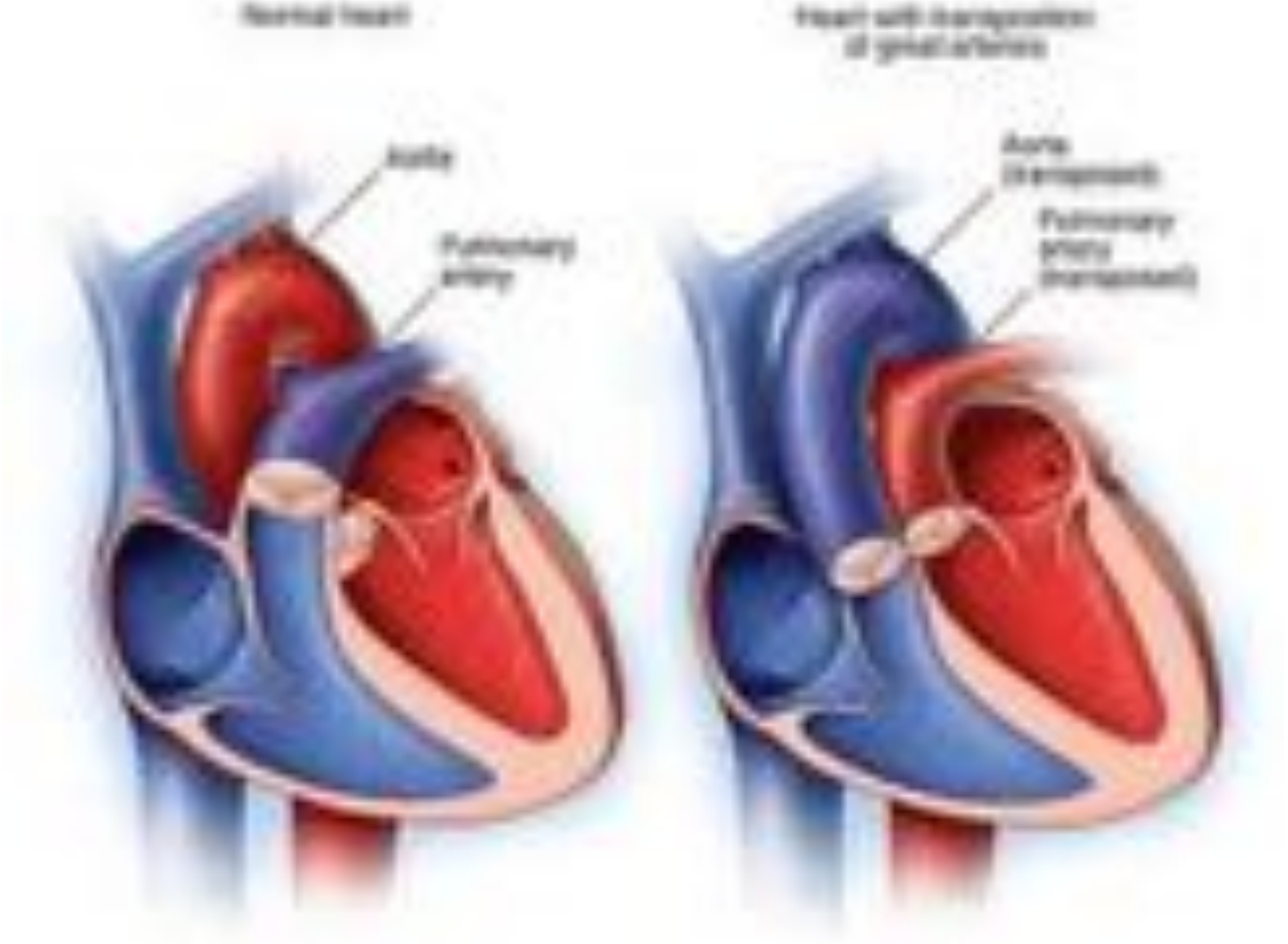
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

Malpositions of the great arteries

4 categories

- Transposition of the great arteries
- Double outlet right ventricle
- Double outlet left ventricle
- Anatomically corrected malposition of the great arteries


Normal heart
Aorta
Pulmonary artery
Heart with transposition of great arteries
Aorta (transposed)
Pulmonary artery (transposed)

ANATOMY OF THE HEART WITH TGA

Normal heart

TGA

Other complexe malpositions of the GA

TGA-VSD

TGA-VSD-PS

TGA-VSD-Coa

DORV-Sub aortic VSD

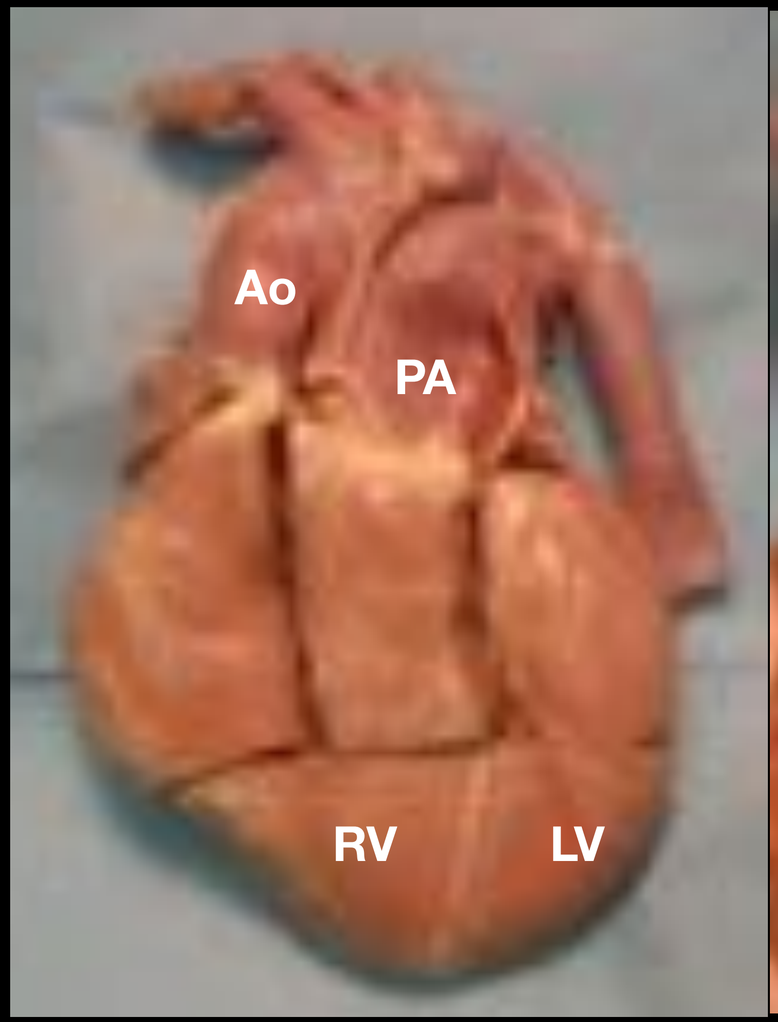
DORV-Sub aortic VSD-SP

Anatomy of transpositions of the great arteries

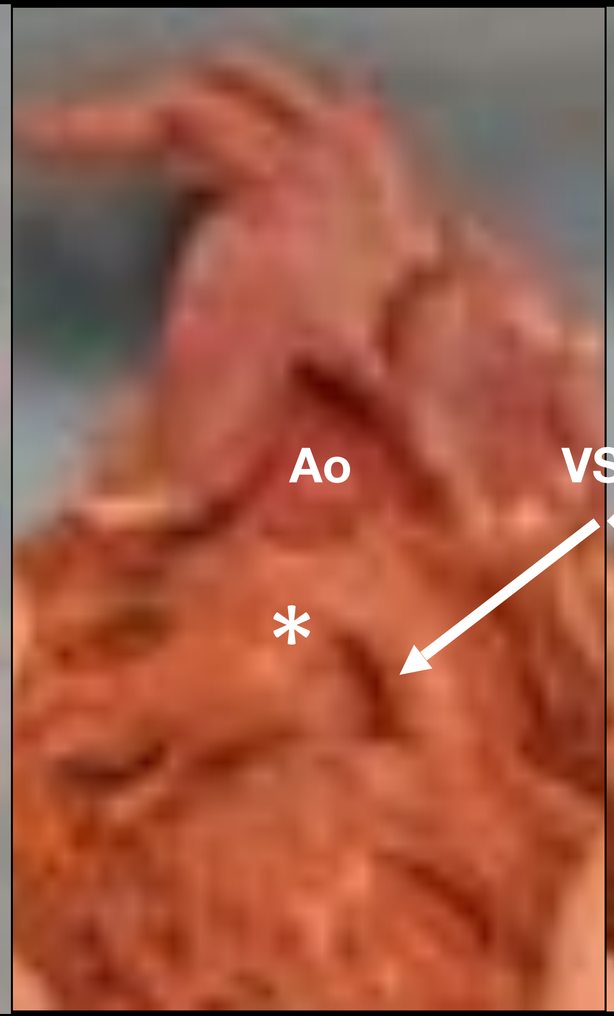
TGA or Ventricle-Arterial Discordance

- Great vessels have parallel course
- Sub-aortic conus
- Fibrous continuity mitral and pulmonary valve
- D-transposition with Aorta anterior and right
- Segment analysis
  - most frequently {S,D,D}
  - sometimes {I,L,L} (mirror image)
  - rarely exception to the looping rule {S,D,L} or {I,L,D}

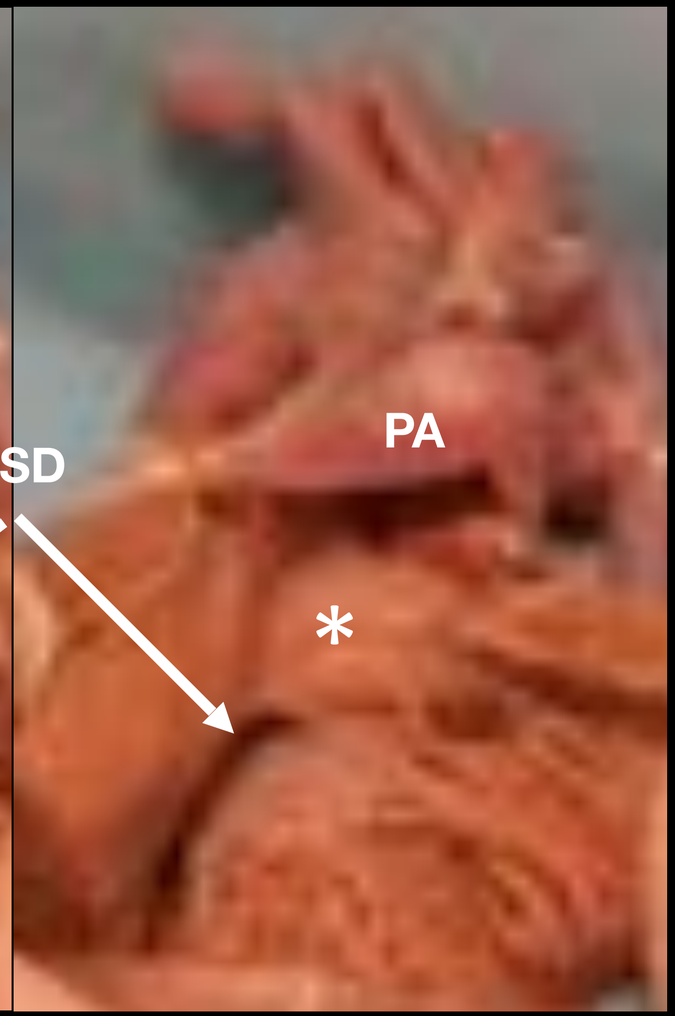
TGA with bilateral conus



TGA {S,D,D}

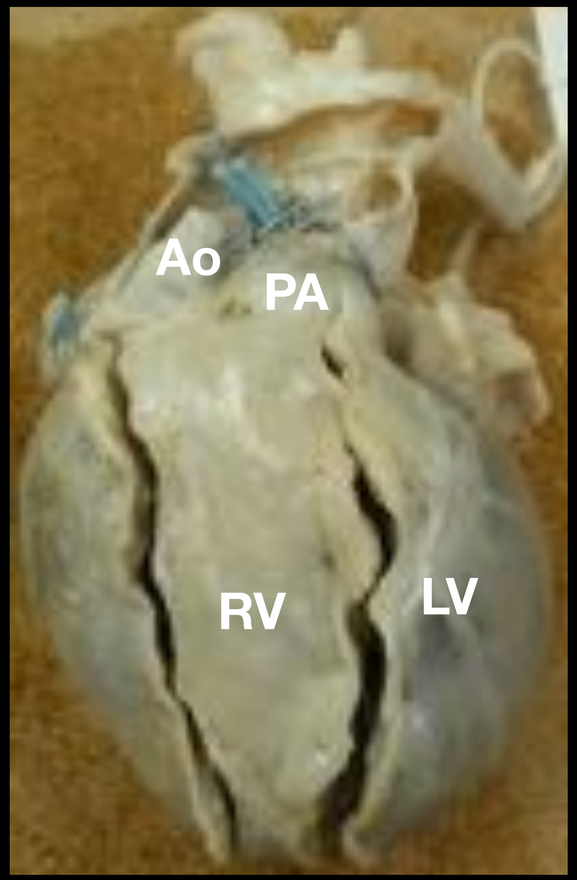


Right ventricle

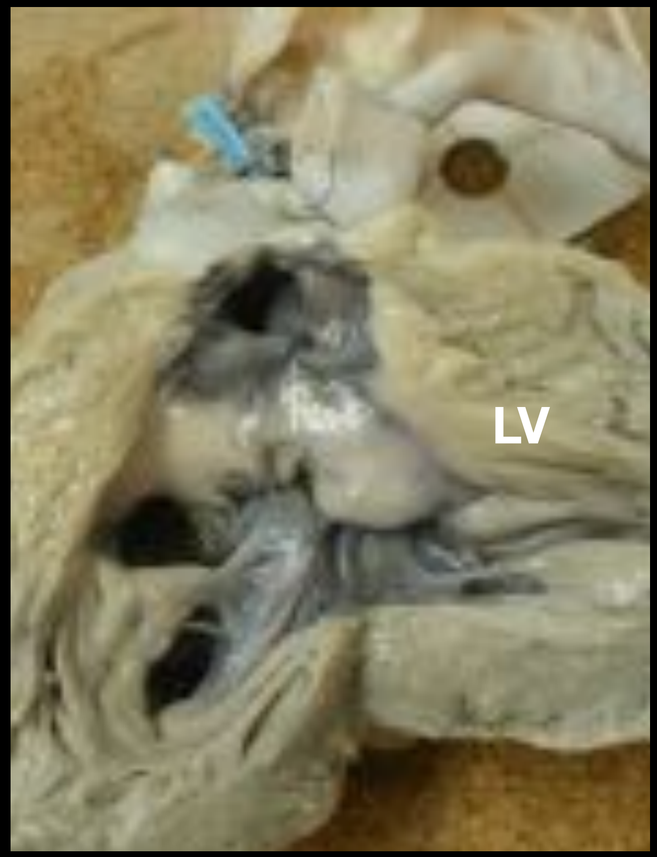


Left ventricle

TGA with bilateral conus

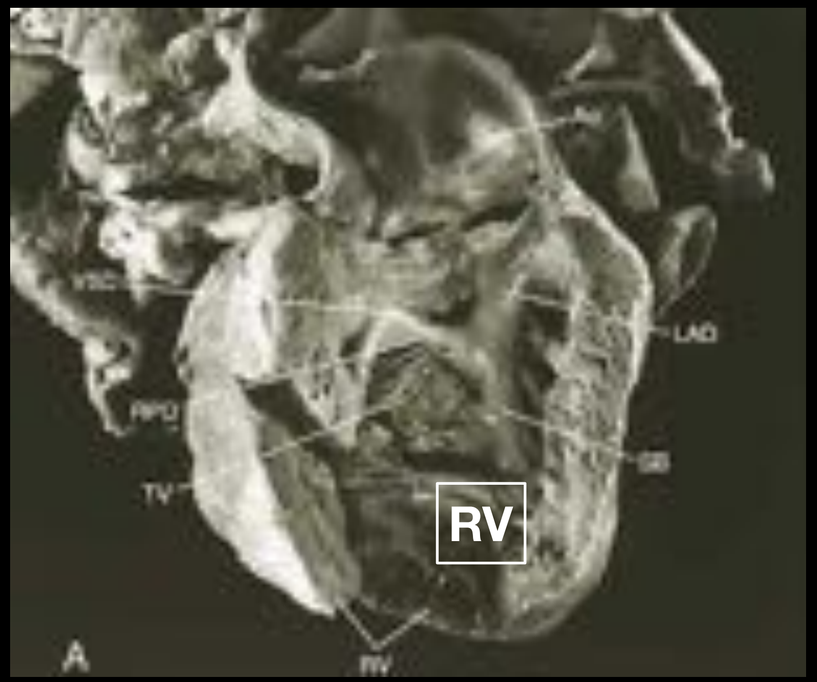


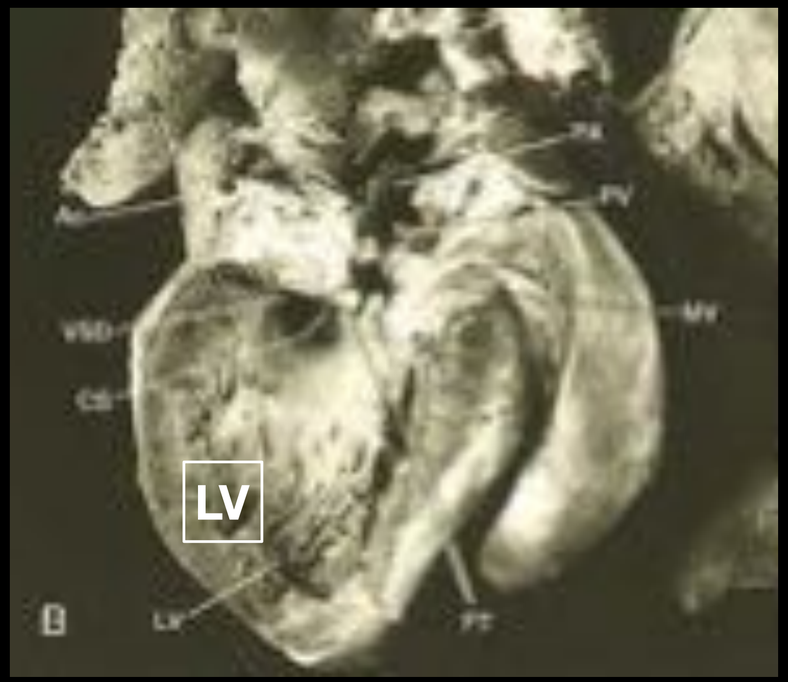
Sub-aortic conus



Sub-pulmonary conus

TGA {S,D,L}

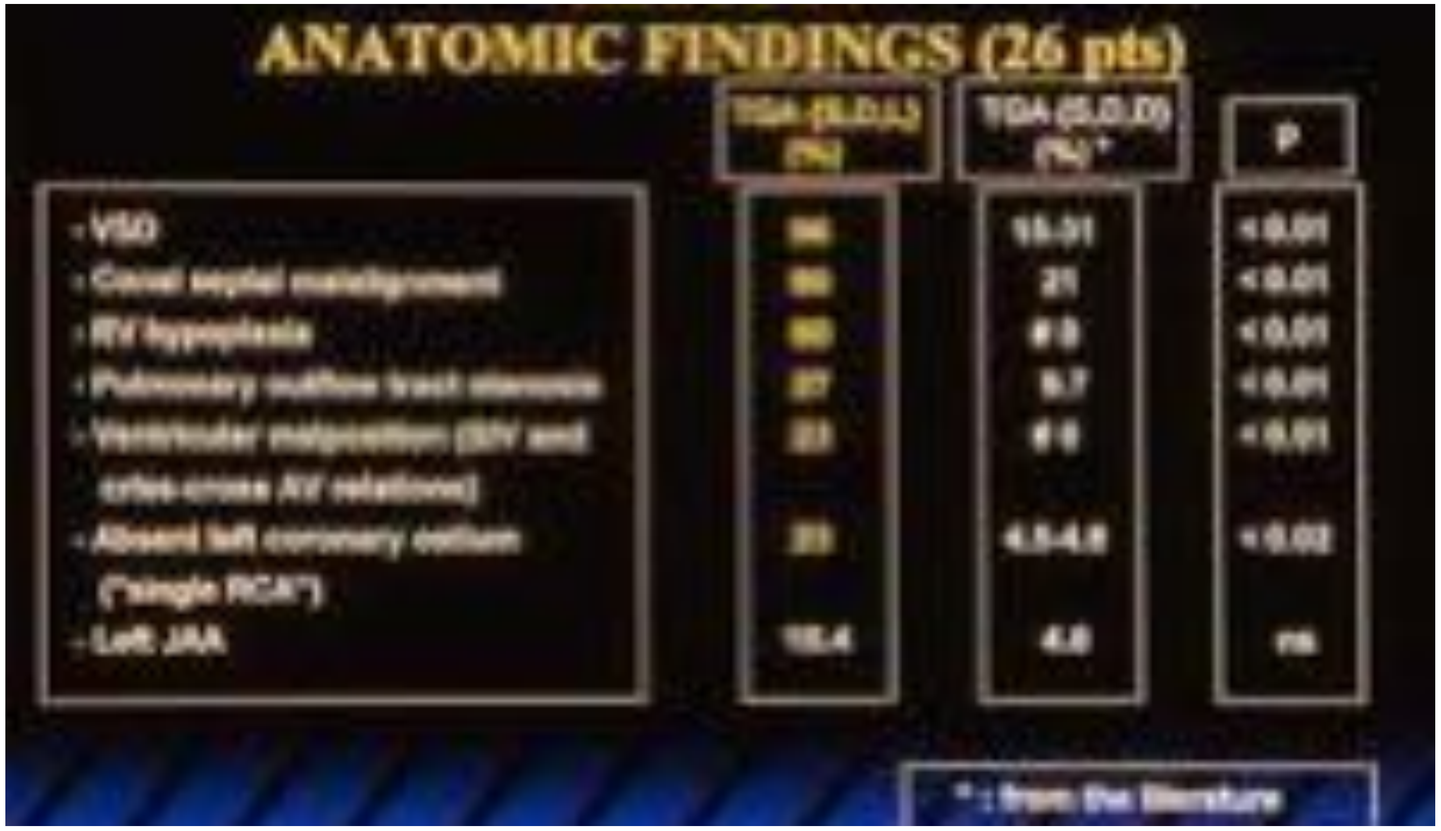




Houyel L, Van Praagh R. et al.
Circulation 1995;110:613-24.

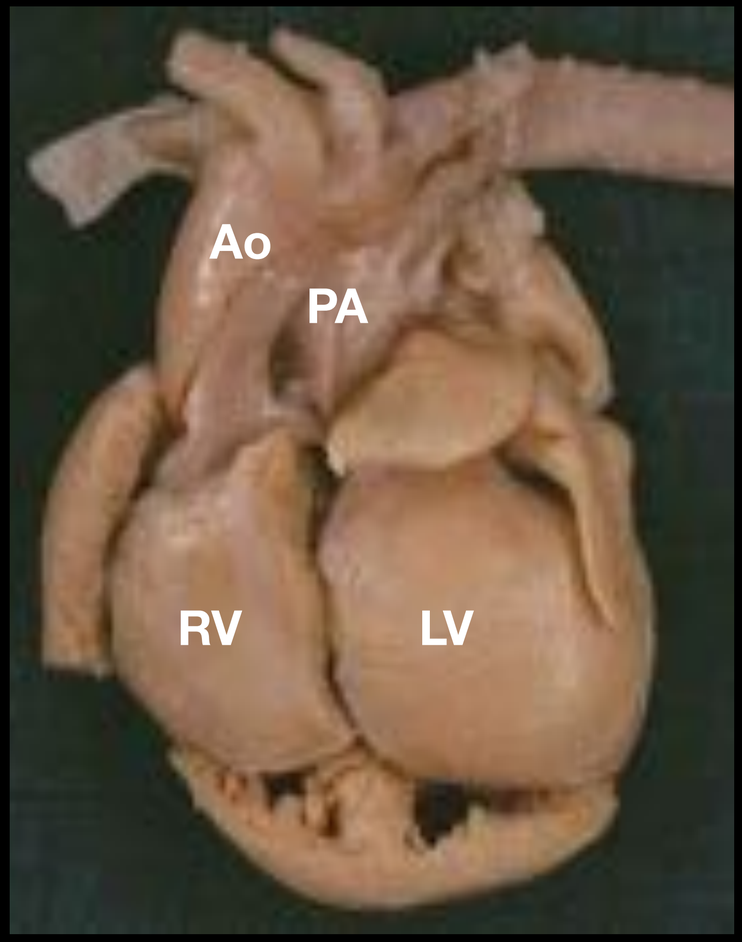
TGV {S,D,L} : pas seulement une variante anatomique de TGV mais une entité anatomique distincte

Implique non seulement le conus mais aussi les ventricules

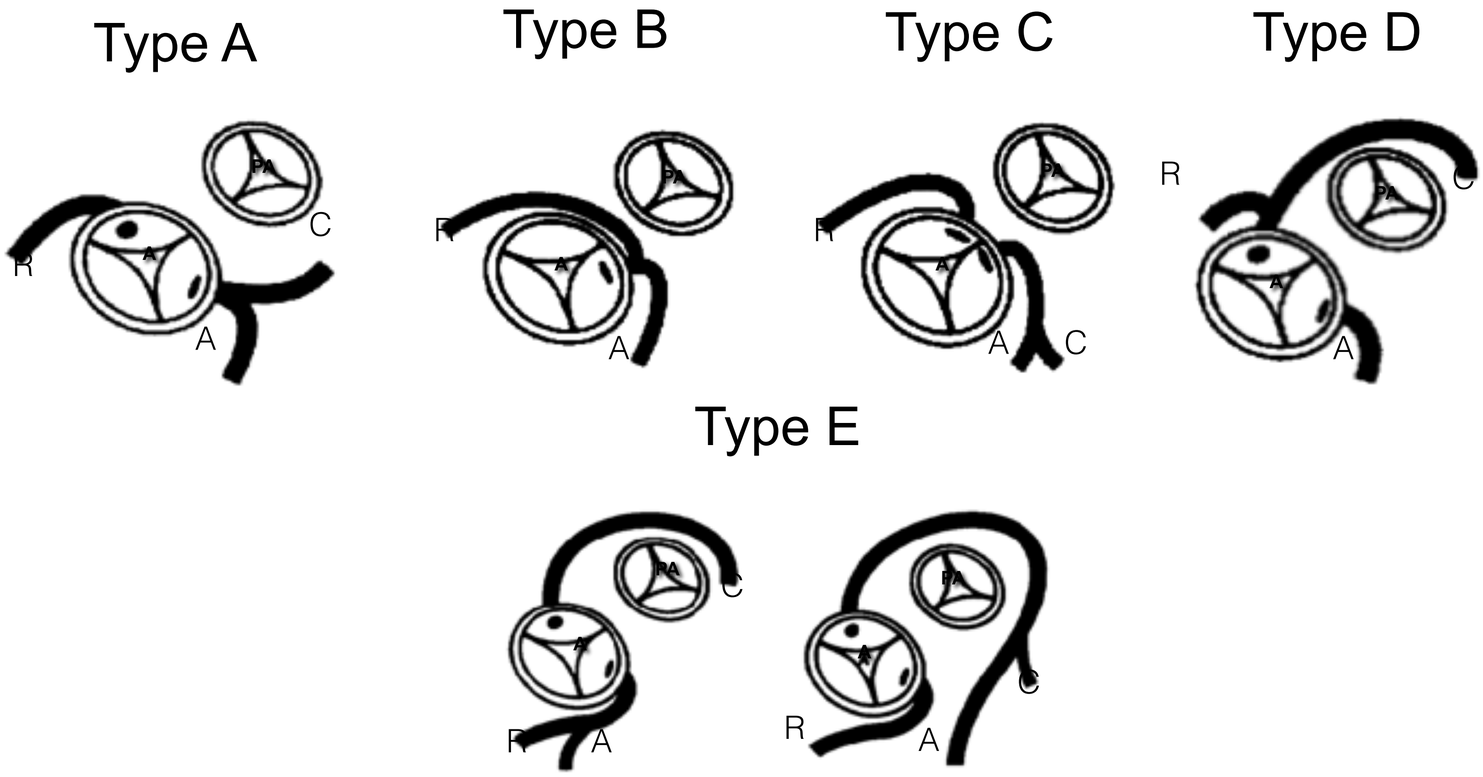


Left juxtaposition of the atrial appendages

- 2 to 5% of cases
- Frequently associated with other anomalies : dextrocardia, hypoplasia of right ventricle, bilateral conus



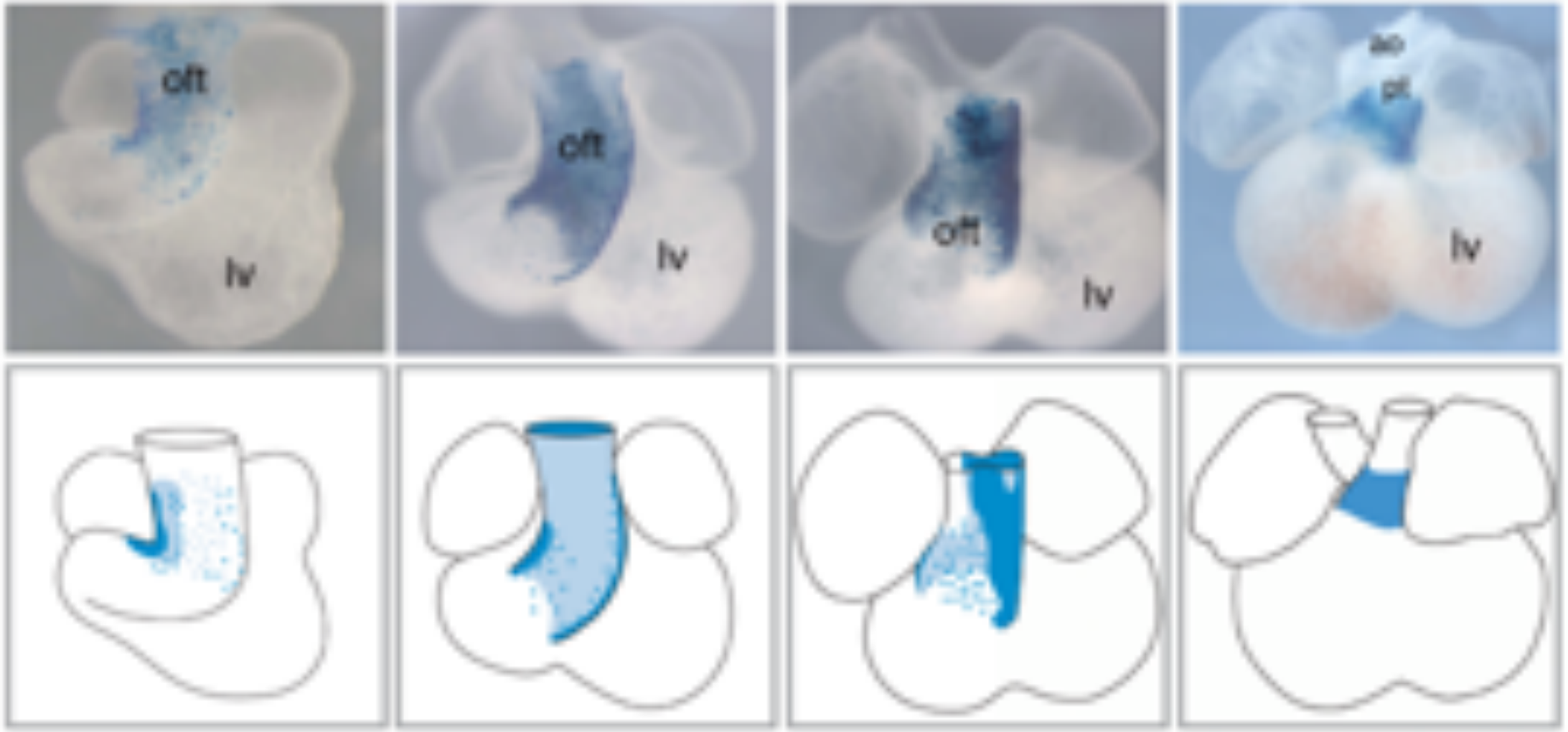
Coronary arteries in TGA



Yacoub and Radley-Smith classification

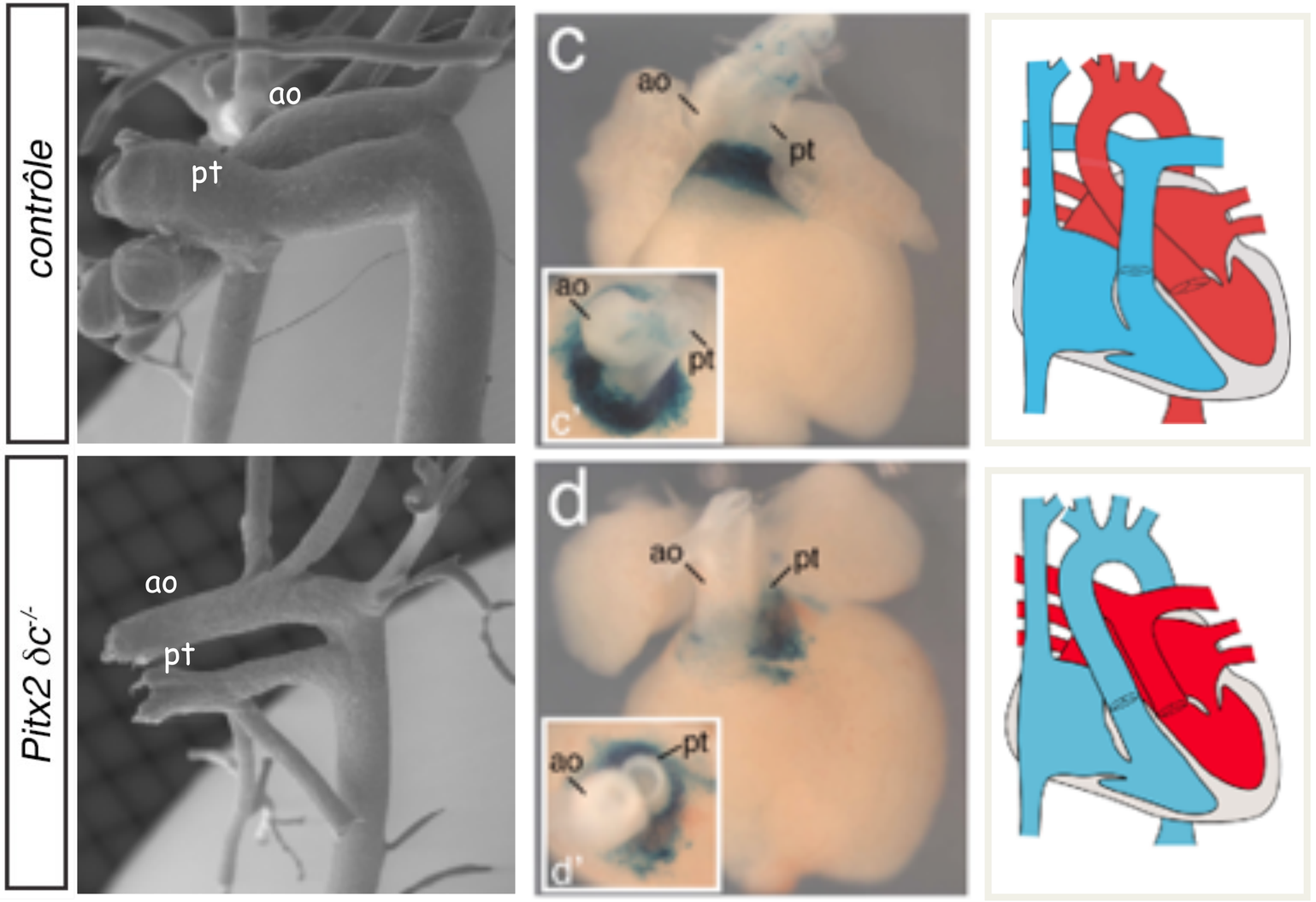
What causes transposition of the great arteries ?

96-16 provides direct evidence for rotation of outflow tract myocardium

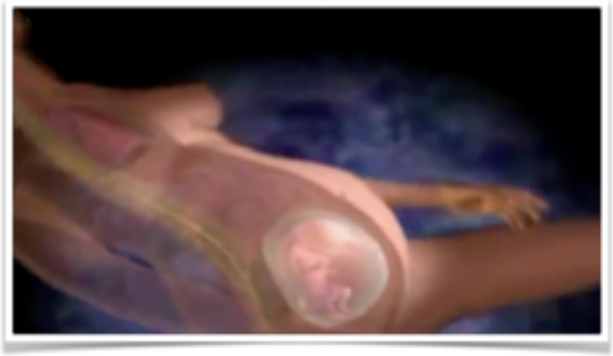


96-16 expression in Pitx2δc heart with TGA

- Transposition of the great arteries with a rotation defect
- Normal septation and normal neural crest cell migration
- Defect of left-right signaling



Foetal TGA



The French system during pregnancy

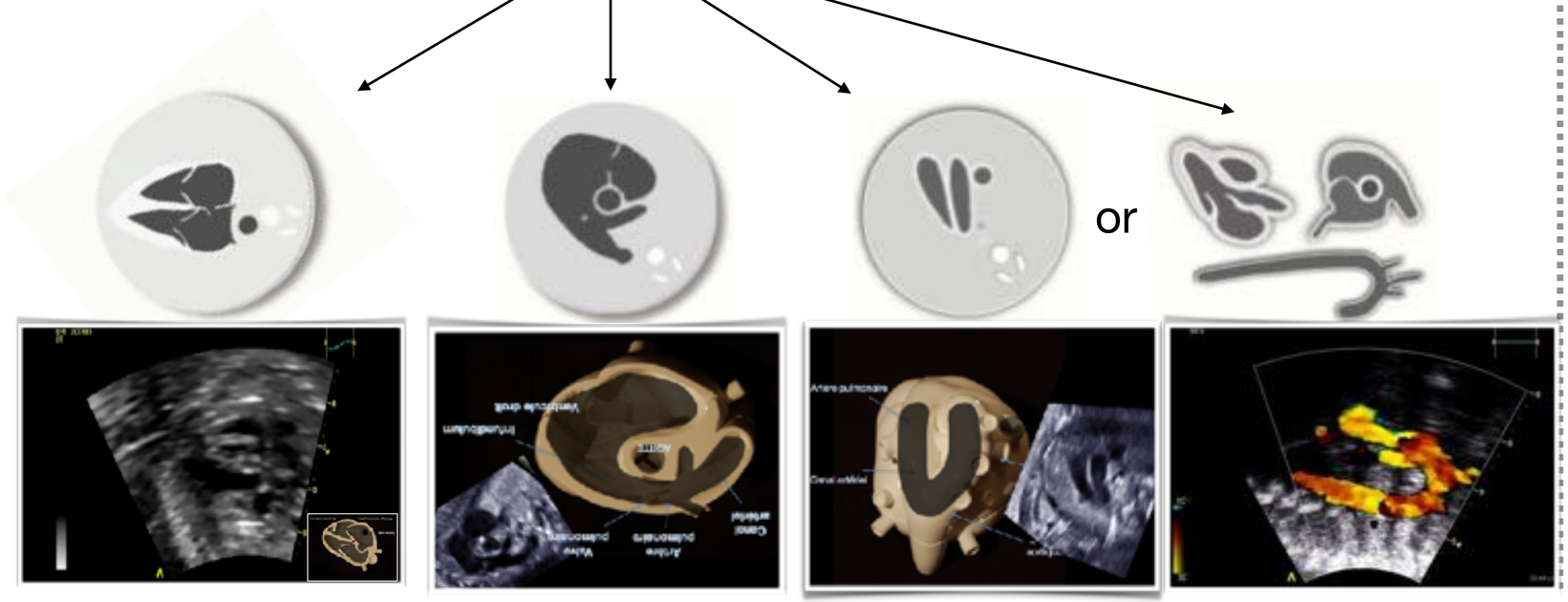
3 systematic foetal echographies - Level 1

11 Weeks — 18-22 Weeks — 32-34 Weeks

Nuchal translucency
Term
Number fetuses

Morphology

Growth
Malformations



In case of anomaly
or difficulty in assessing
normality

Level 2

Expert foetal
echography

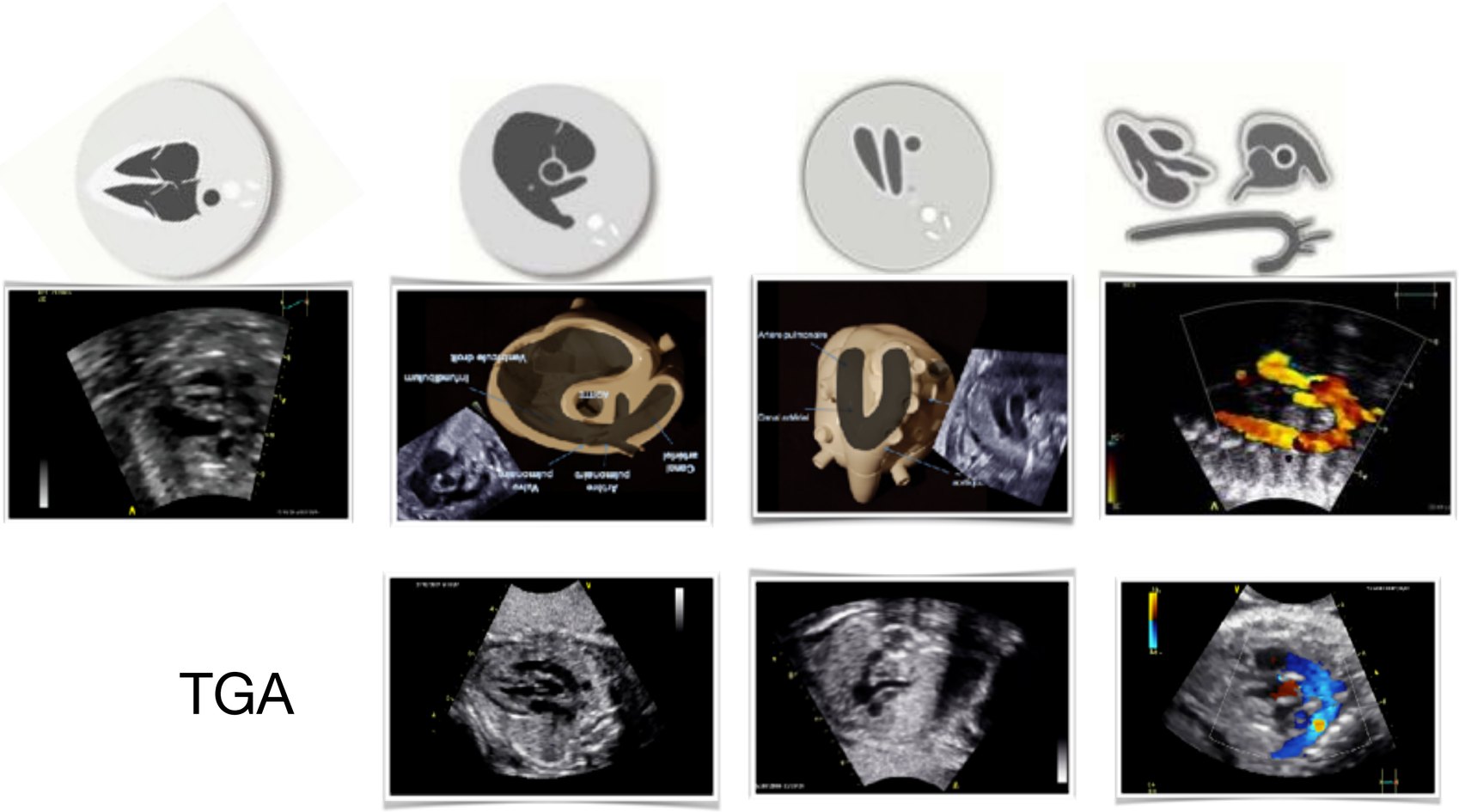
If heart anomaly is confirmed

↓

Level 3

Fetal echocardiography
by expert

Prenatal diagnosis of TGA



TGA

Preoperative mortality in TGA = 4-6%
(*vs./+)* Surgical mortality = 1-2%

Comparison of Characteristics of Patients in the Prenatal and Postnatal Groups

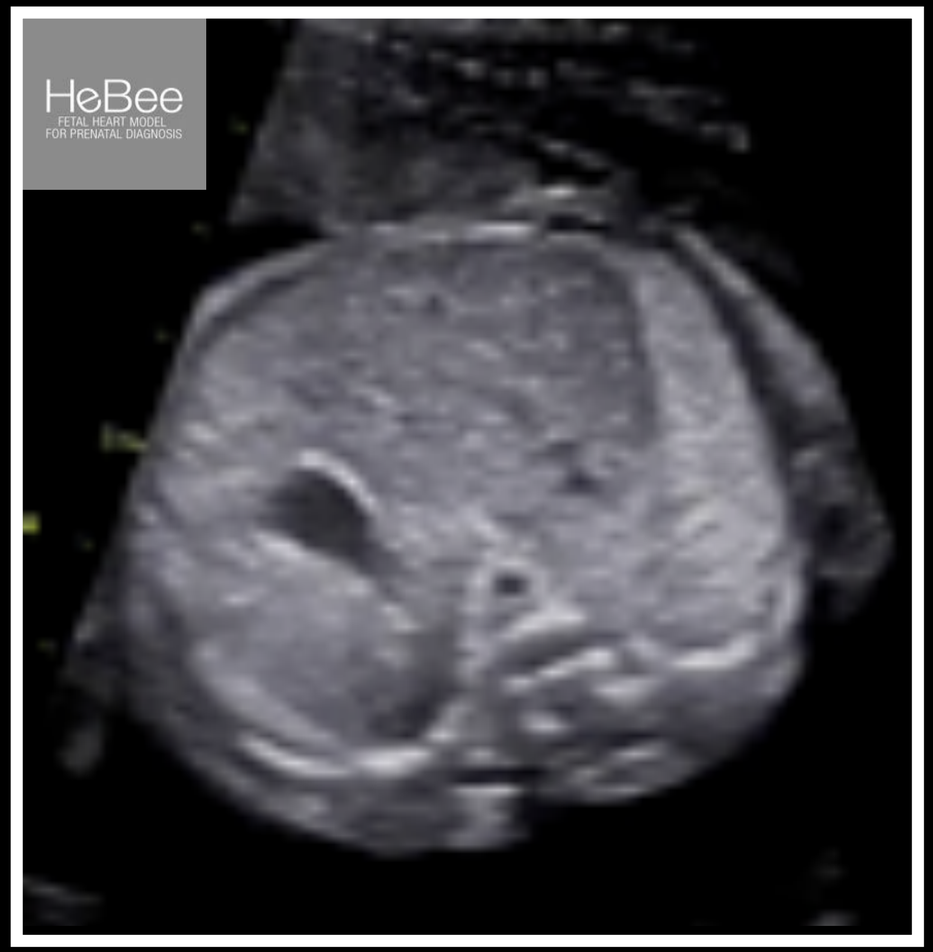
Isolated TGA			
Associated defects			
VSD			
VSD + CoA			
CoA			
Age at admission, y			
Mechanical ventilation			
Inotropic support + IABP			
PGE, infusion			
BAS			
Preoperative mortality			
Coronary artery pattern			
Normal			
Abnormal			
Postoperative mortality			
Hospital stay, d			

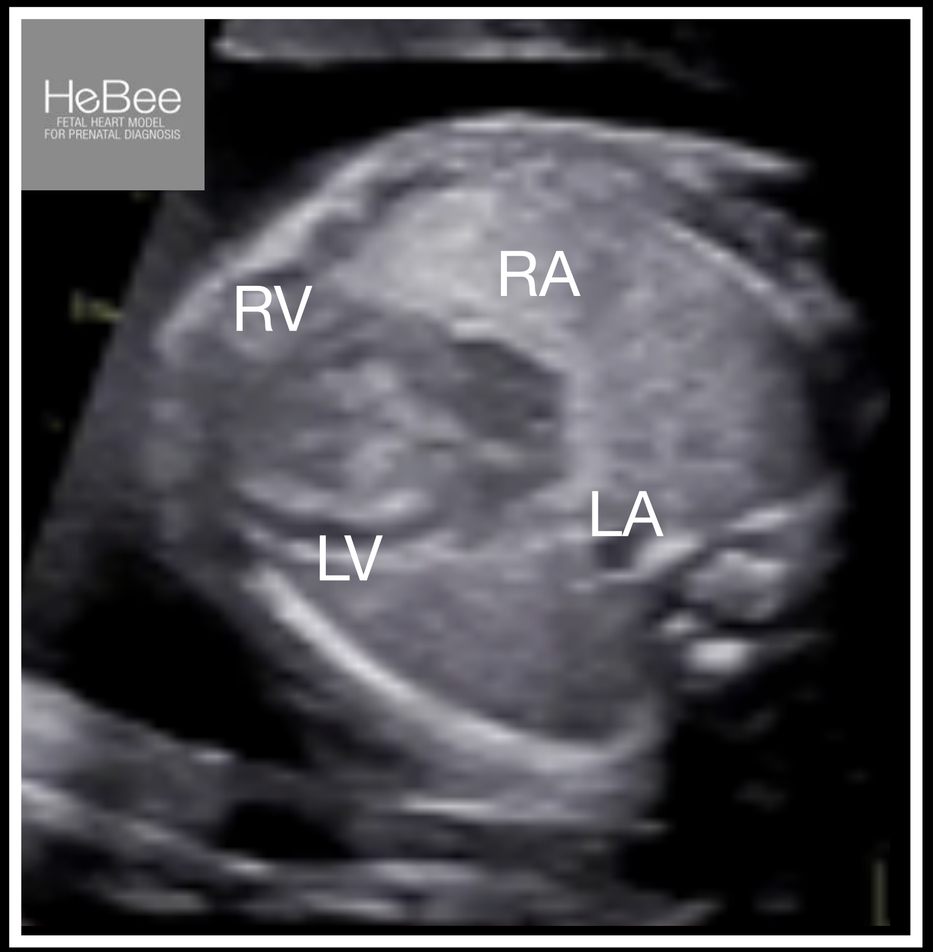
VSD indicates ventricular septal defect; CoA, coarctation; IABP, multiorgan failure; PGE, prostaglandin E; BAS, balloon atrioseptostomy; and ASO, arterial switch operation. Values are n (%).

Bonnet D et al. Circulation 1997

TGA

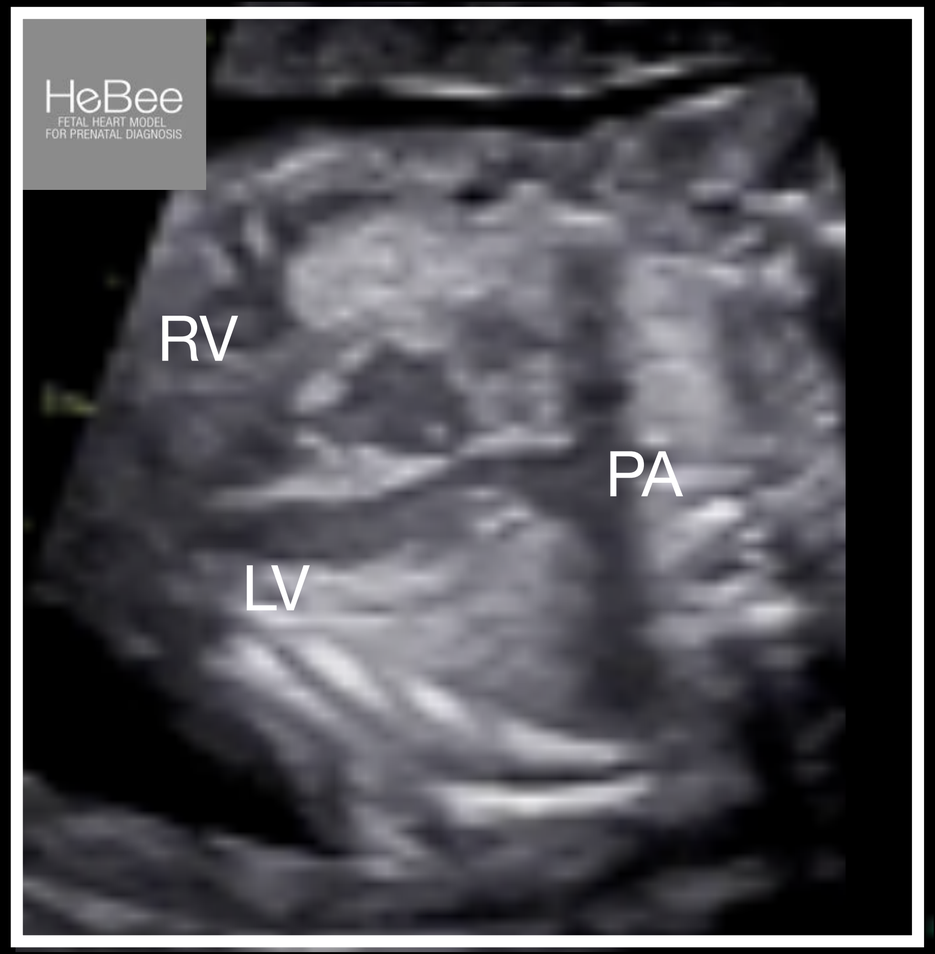
situs & 4 chamber view

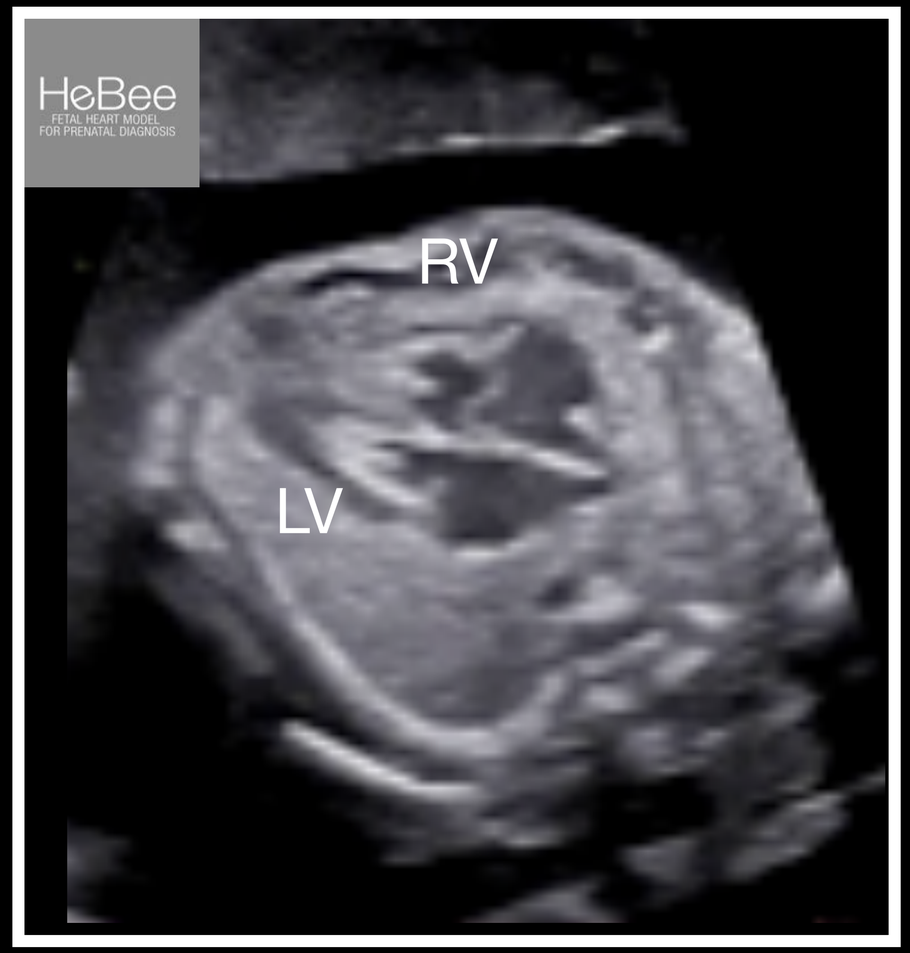




TGA

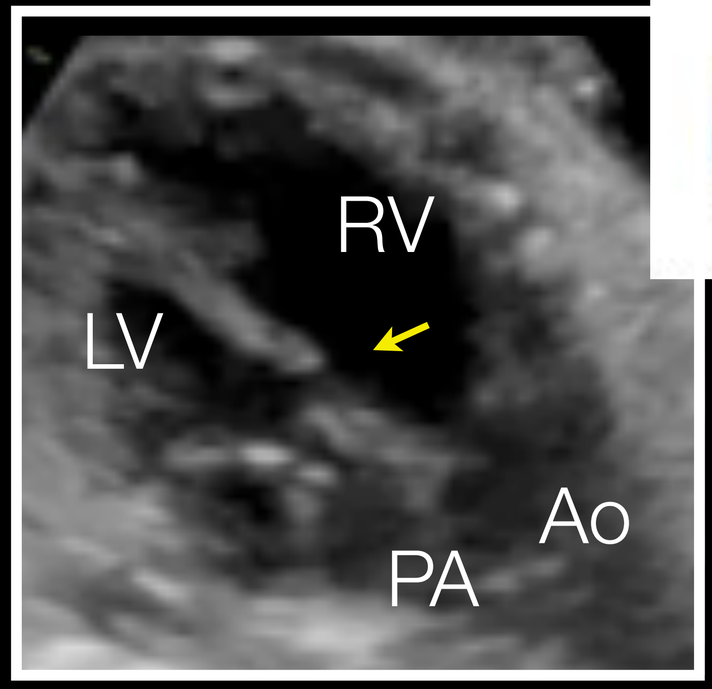
LVOT view

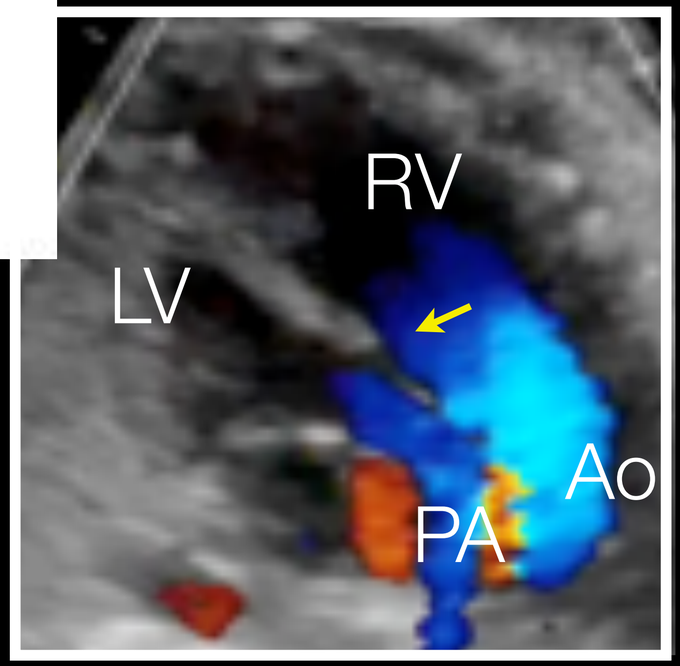




TGA

Complex forms

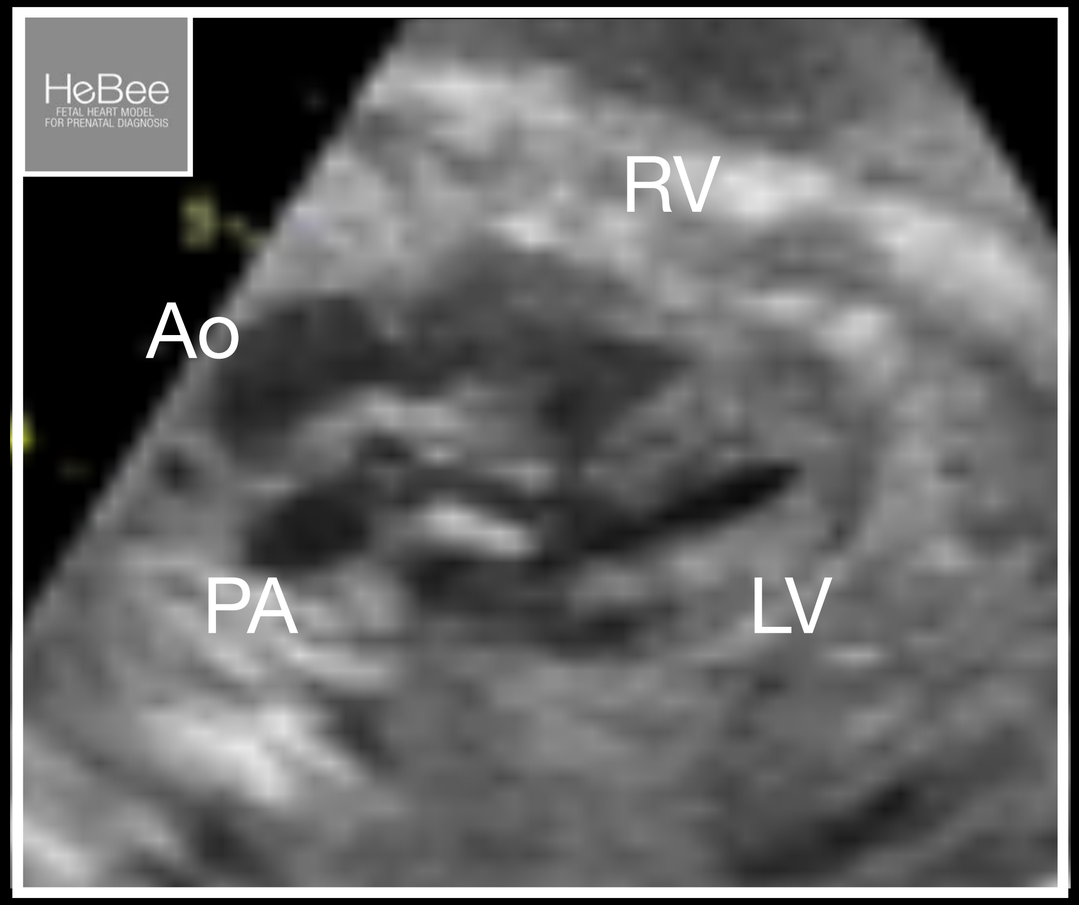




TGA-membranous VSD

TGA

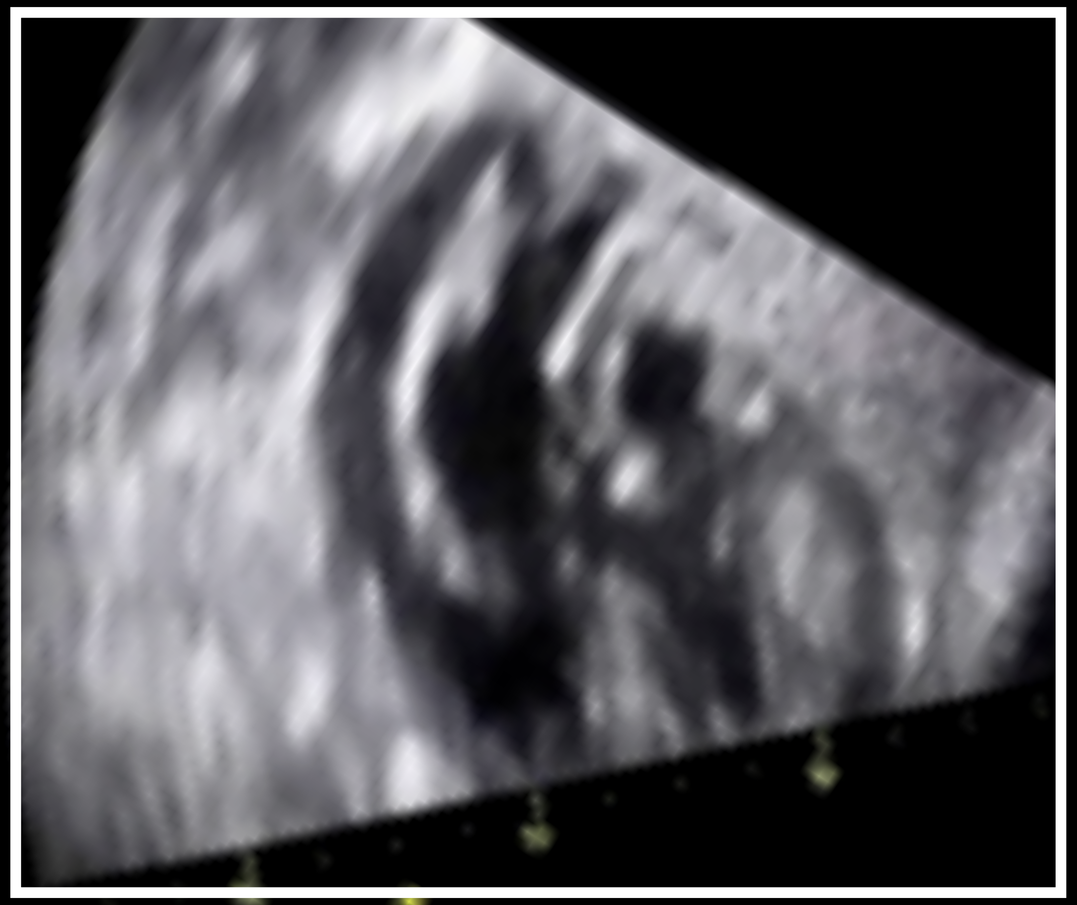
Complex forms



TGA-VSD-PS

TGA

Complex forms



TGA-VSD-Coa

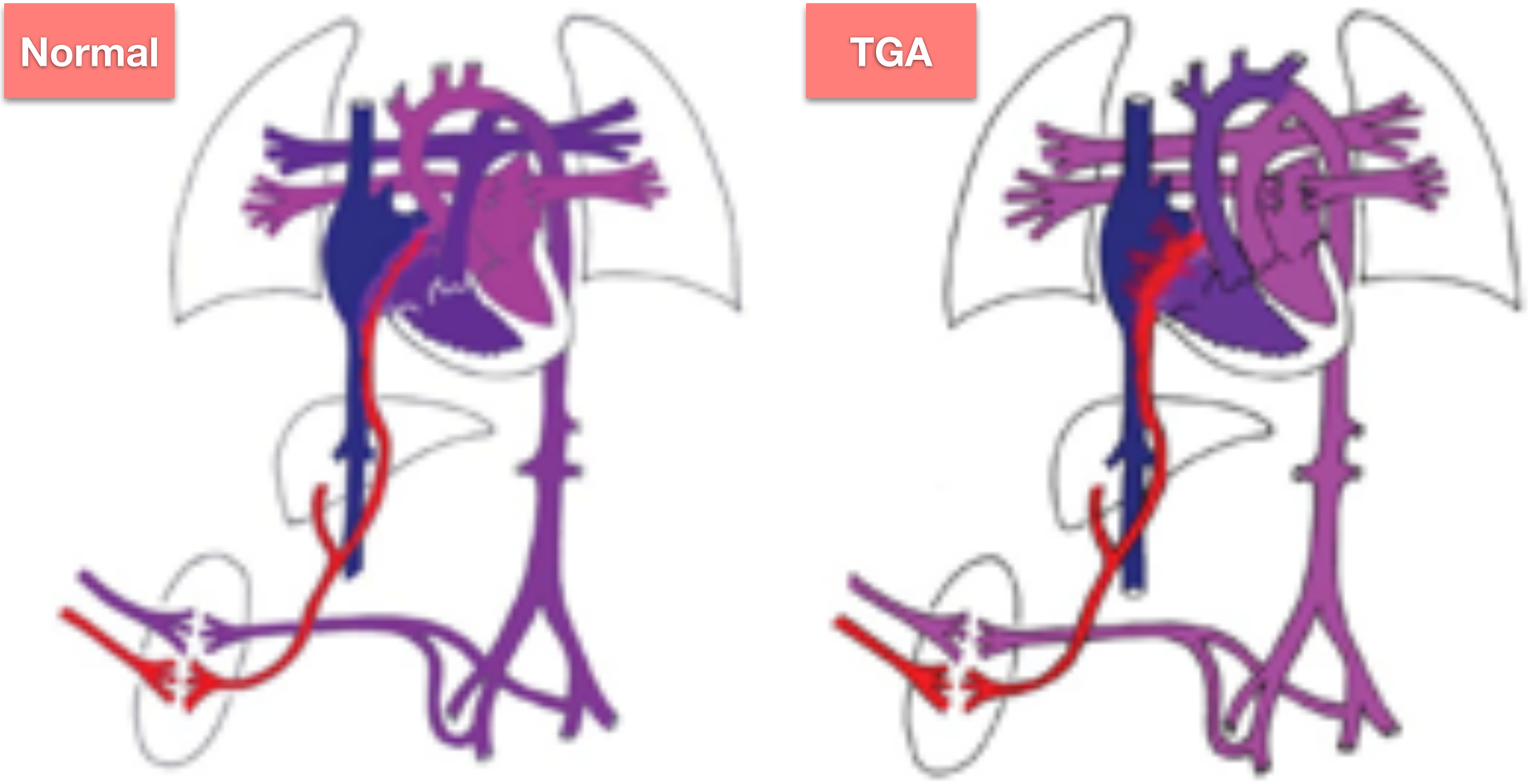
Chromosomal anomalies in fetal CHD

548 CHD-18.5%

PA-IVS and PS	0	0
Left heart obstruction	12/130	9.2%
6 XO; 3 T18; 3 translocations		
Conotruncal defects	23/91	25%
20 del22q11; 1 T21; 2 translocations		
AVSD	32/68	47%
28 T21; 3 T18; 1 XXX		
VSD	12/74	16%
9 trisomies, 2 del22q11, 1 del5		
Transposition of the great vessels	0	0
DORV	7/38	18%
Univentricular heart	2/24	8%
2 T18		

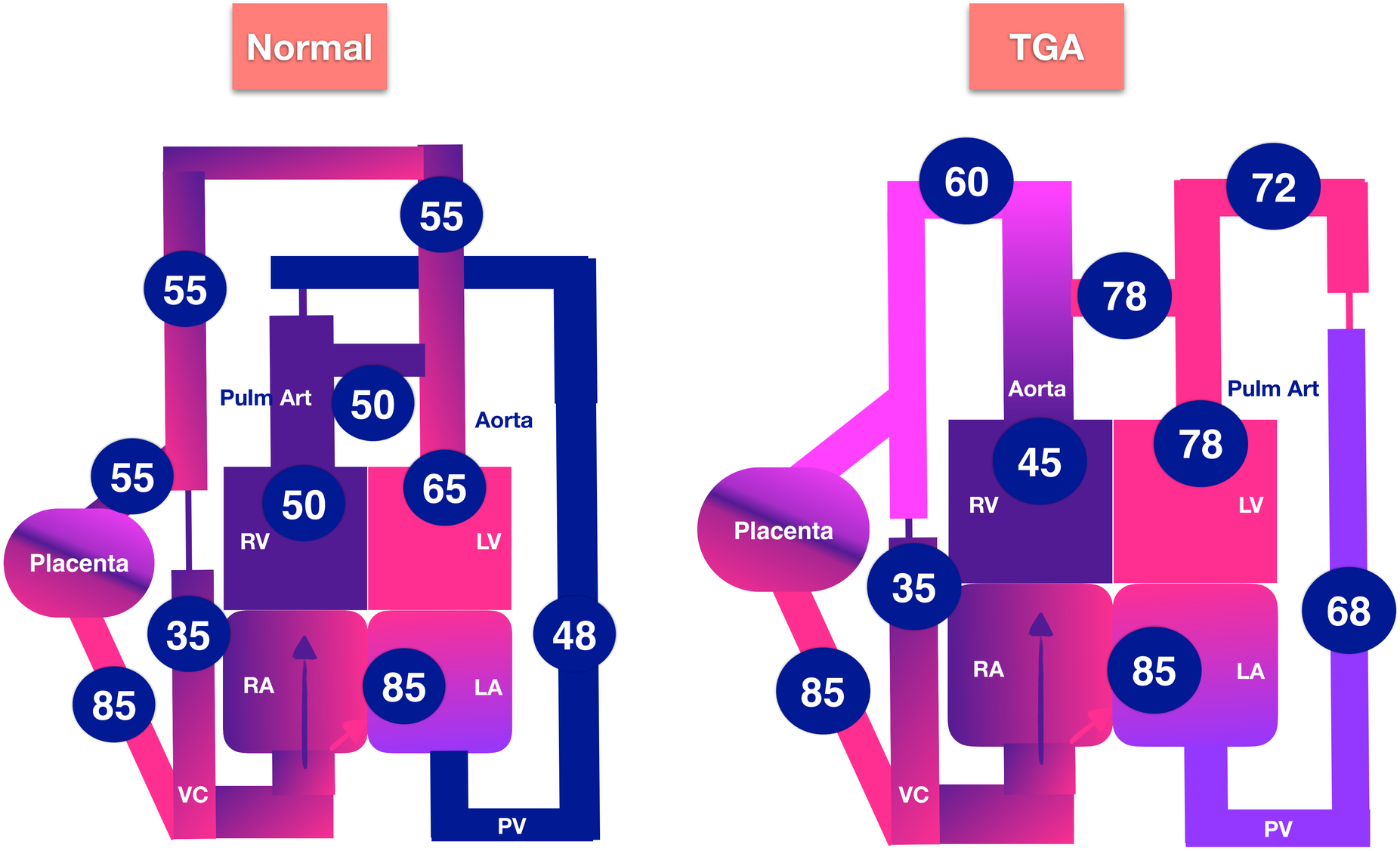
Physiology of TGA

Morphological and physiological consequences of the fetal circulation in TGA



Sun L et al. Circulation. 2015;131:1313-1323.

Morphological and physiological consequences of the fetal circulation in TGA

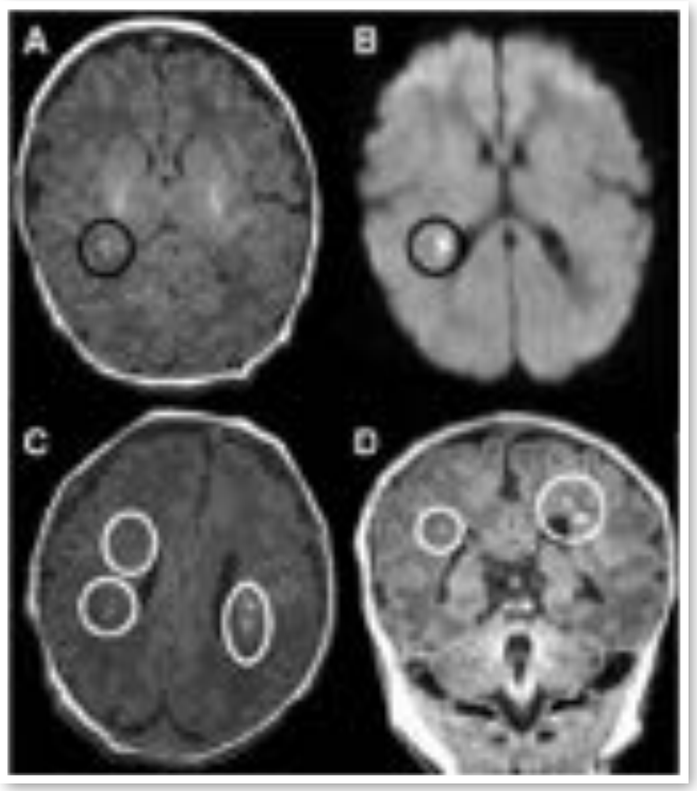


Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

Prenatal white matter MRI anomalies in children with cyanotic congenital heart diseases

- **White matter lesions in** 30 to 40% of newborns with TGA (Miller et al., 2004; Licht et al., 2009)

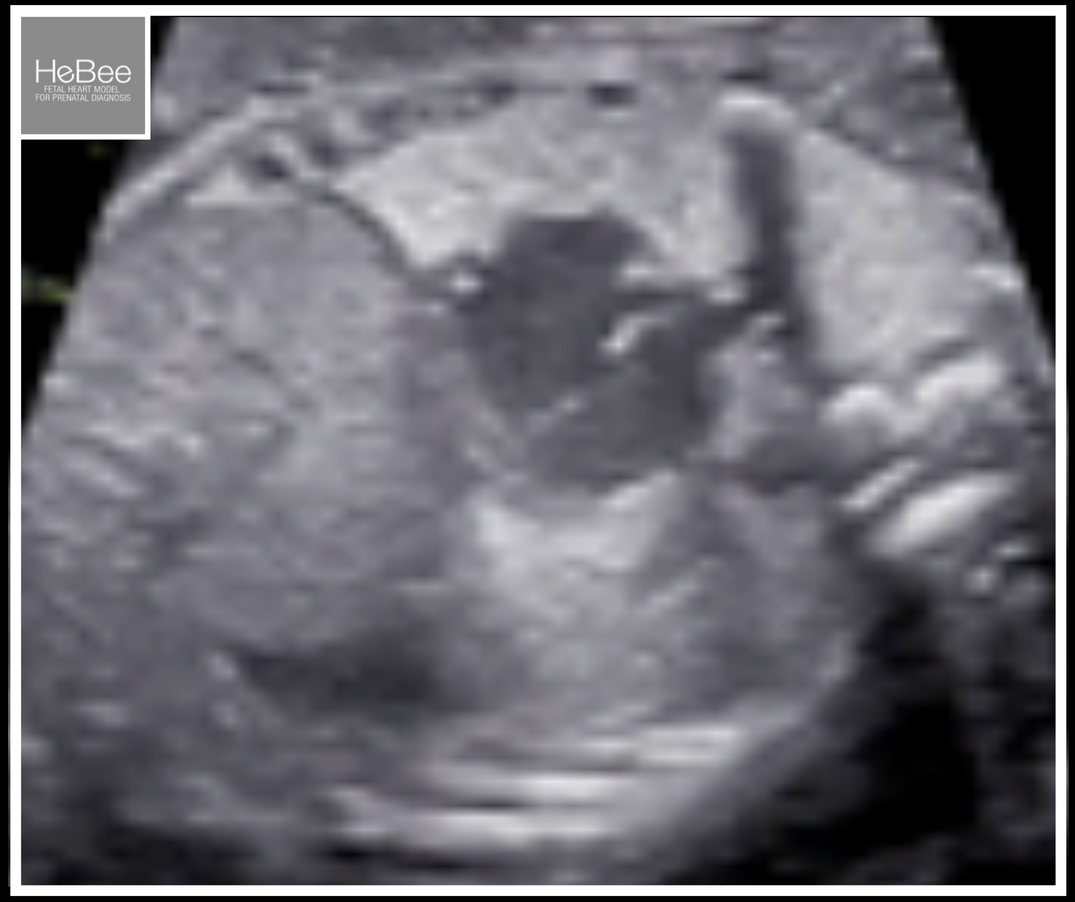
- Same type of anomalies but more severe in complex CHDs such as HLHS (Mahle et al., 2002).



Periventricular white matter lesions in a child with TGA **before** the arterial swicth.
Petit et al., 2009 *in Circulation*

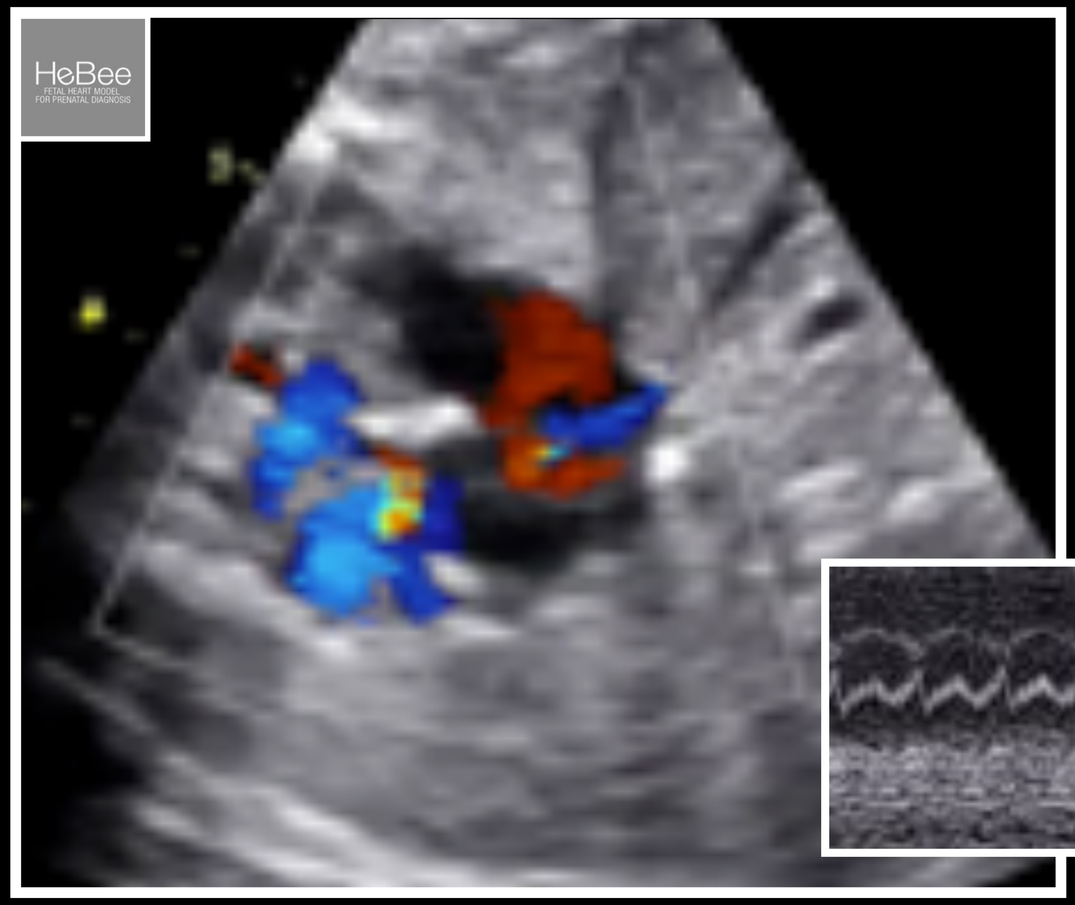
TGA

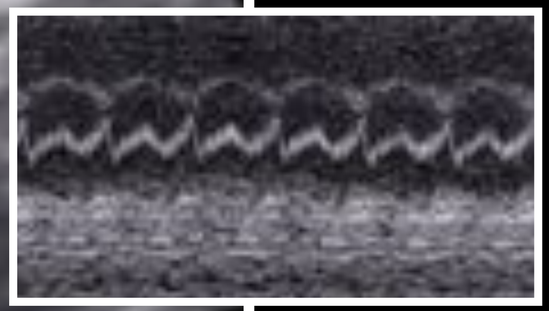
Foramen Ovale



TGA

Foramen Ovale

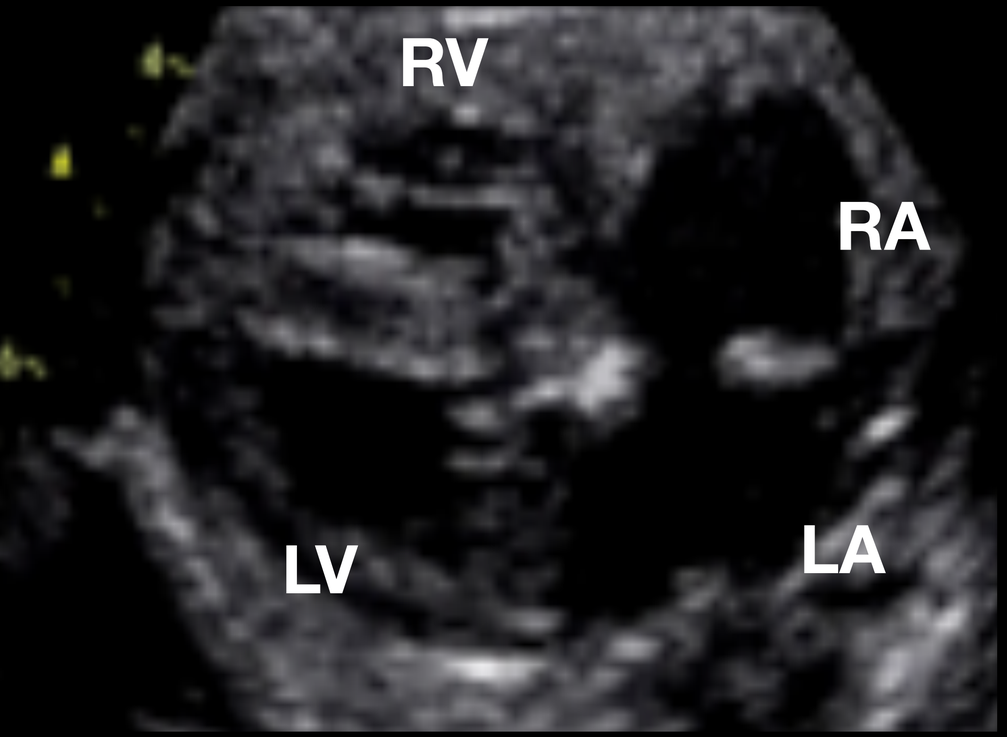


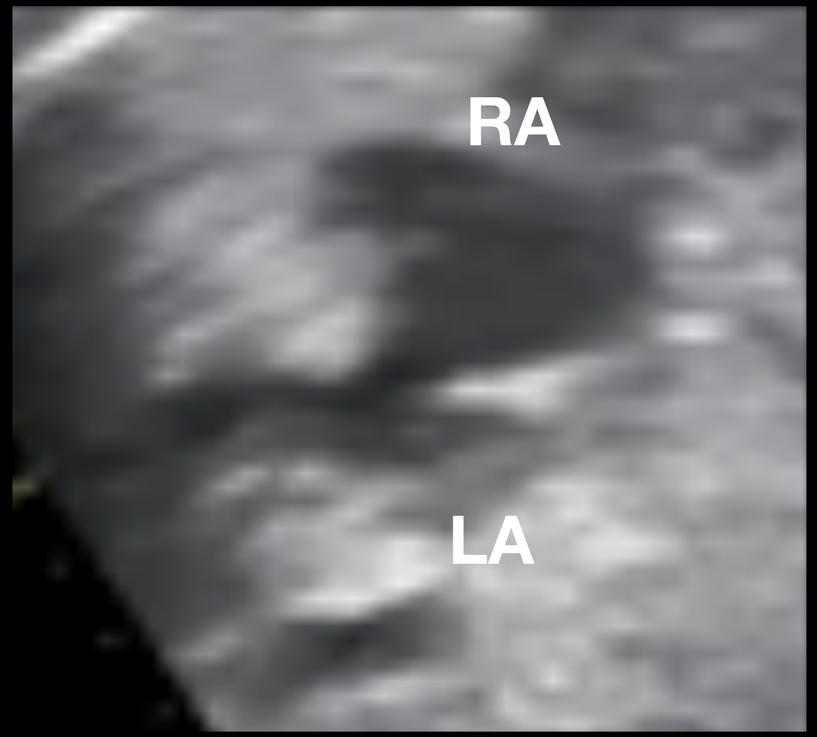


-Floppy Mb
-Small FO
-Small septal length
-pulmonary veins velocity > 0,41 m/s

TGA

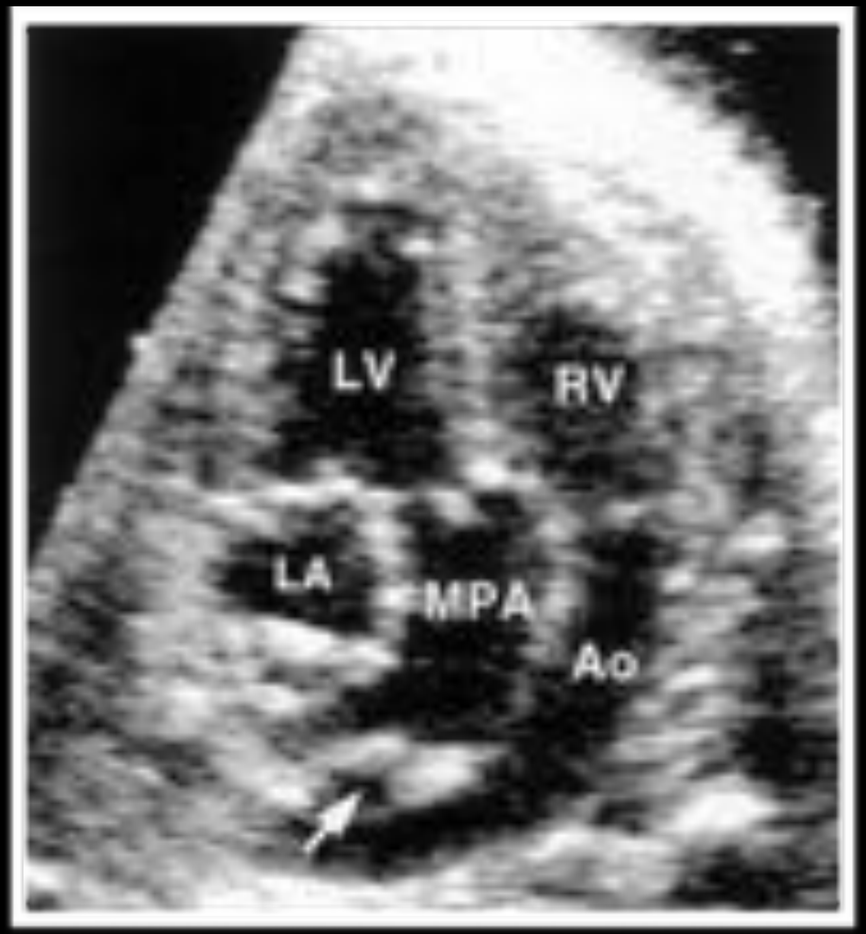
Foramen Ovale





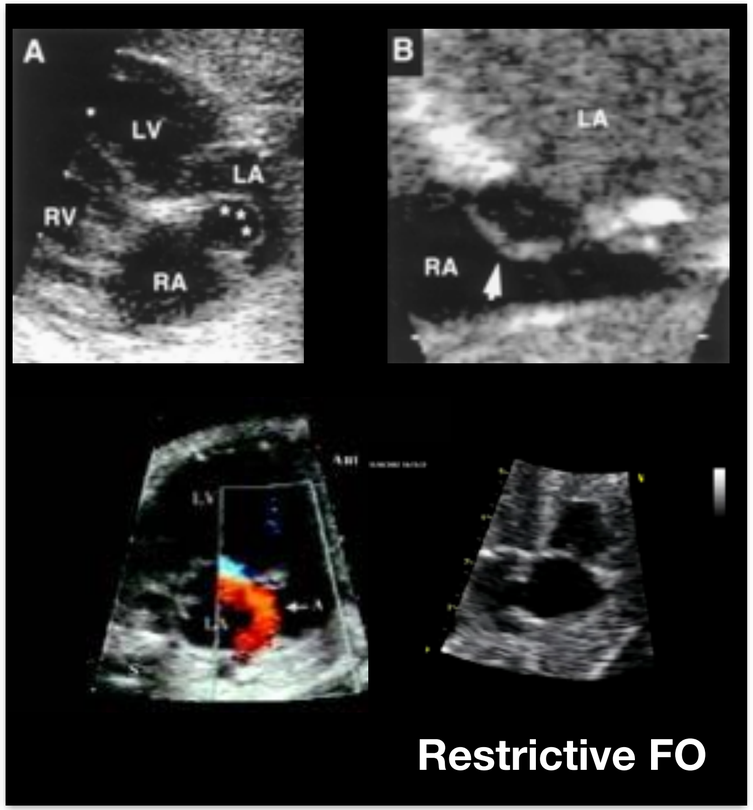
TGA

Arterial duct

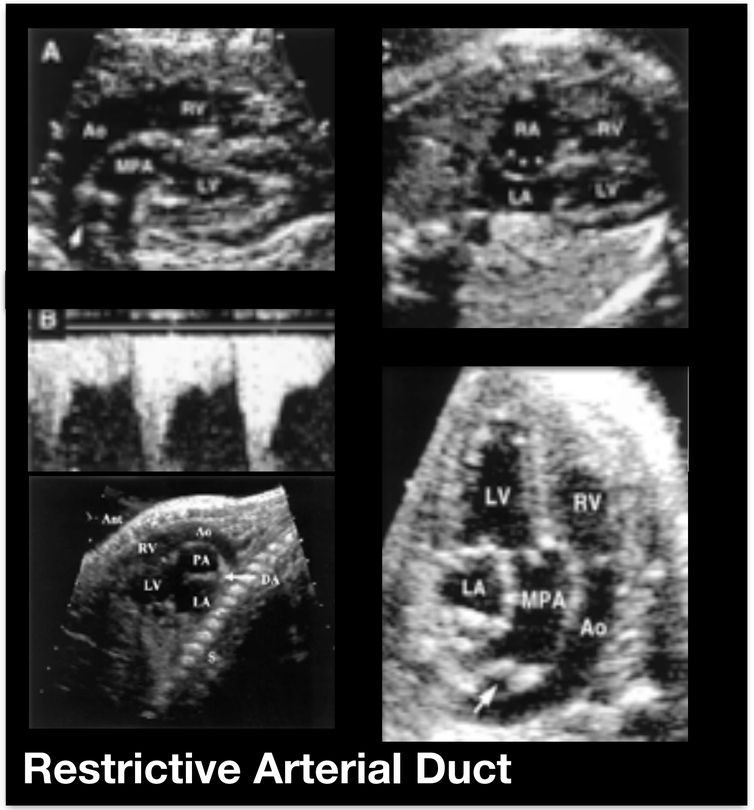


Neonatal management of TGA

Prevention of early neonatal demise



Restrictive FO



Restrictive Arterial Duct

Abnormal Prenatal Shunts and Neonatal Condition

	Abnormal (N=24)		
	FO and DA	FO or DA*	Normal (N=95)
N total	4	20 (19 FO; 1 DA)	95
Critical condition (n=7)	4	3 (2 FO; 1 DA)	6
Stable condition (n=17)	0	17	89

FO indicates foramen ovale; DA, ductus arteriosus.
*This subgroup included 1 fetus in whom the FO was restrictive but the DA could not be analyzed.

Additional criteria [2]
A hypermobile septum and reverse diastolic patent ductus arteriosus

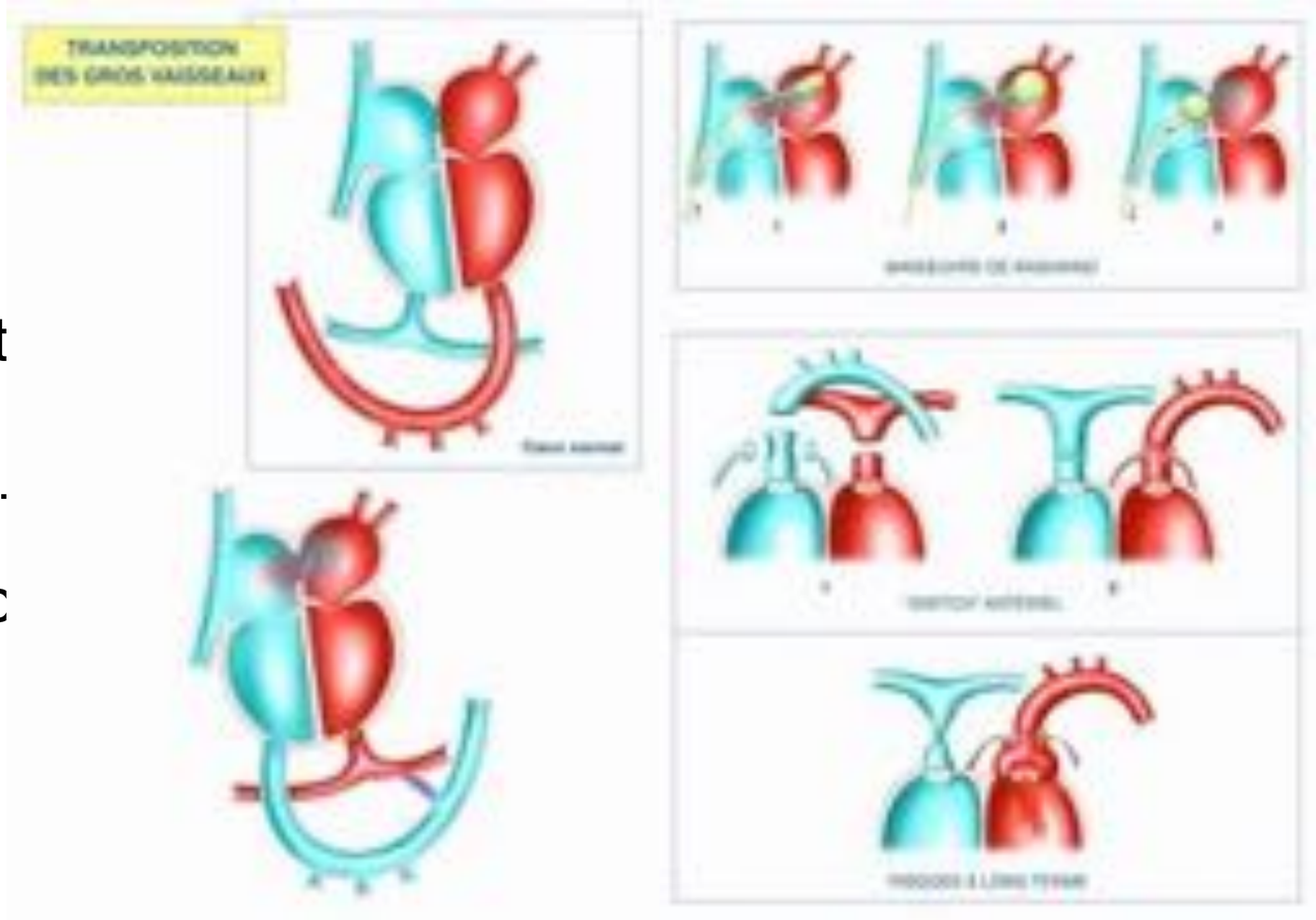
Finally, we do not care
All TGA are delivered on site with the same protocol

Maeno YV et al. Circulation 1999
Jouannic JM et al. Circulation 2004;110:1743-6
2-Punn R, Silvermann N. JASE 2011;24:425-30
Mary T. Donofrio Circulation. 2002;105:e65-e66

Prenatal diagnosis of transposition of the great arteries

Perinatal organization in Paris

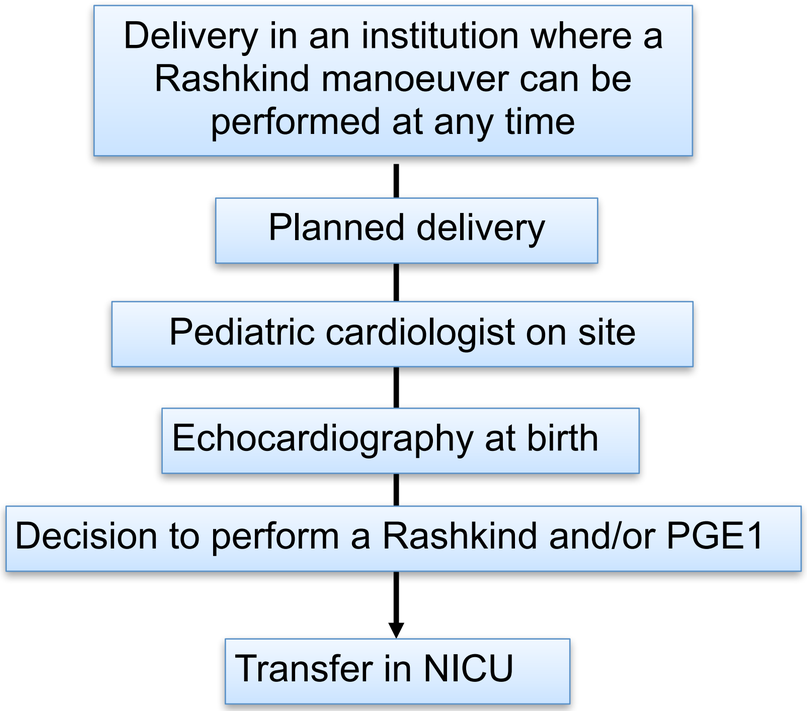
- Organisation of foetal cardiac growth surveillance
  - Foramen ovale and arterial duct
- *In utero* transfer organisation
- Organisation of perinatal management
- Prevention of early neonatal demise
- Prepare the parents to the future even
- Post-natal management and follow-up



Bonnet et al. Circulation 1997; Maeno YV et al. Circulation 1999; Jouannic et al. Circulation 2004; Van Velzen C et al. Ultrasound Obstet Gynecol 2015; 45: 320–325
Escobar-Diaz MC et al. Ultrasound Obstet Gynecol. 2015 June ; 45(6): 678–682.

Prenatal diagnosis of transposition of the great arteries

Perinatal organization



From 1992 to 217

717 prenatally diagnosed TGA (IVS or complex)

6 had congenitally corrected TGA

3 deaths immediately after birth in the delivery room

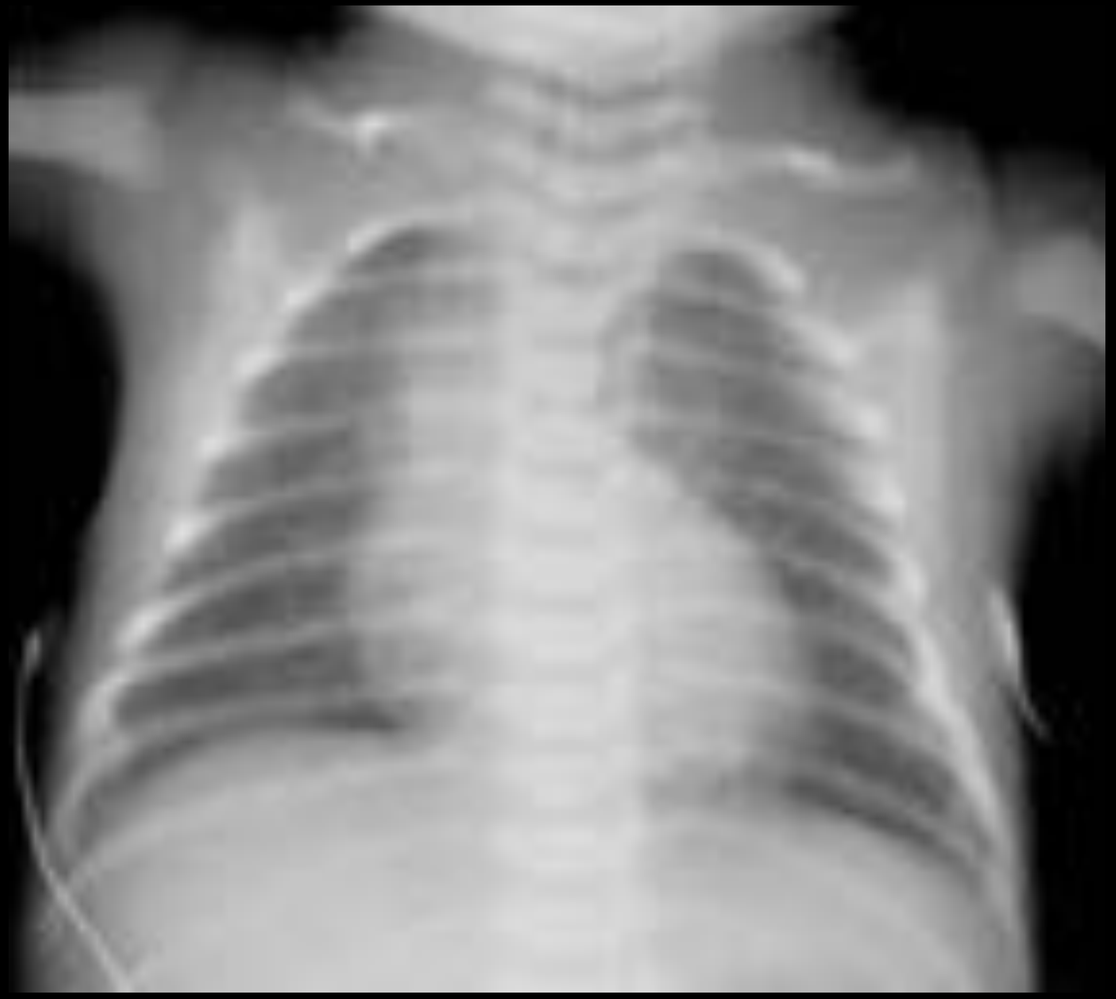
3 additional preoperative deaths

1 extra cardiac malformation in a CHARGE syndrome,
1 necrotizing enterocolitis
1 during the Rashkind procedure (perforation of the left atrium in left juxtaposition of the atrial appendages)

Surgical mortality 1.7 % : 693 survivors at discharge

Neonatal diagnosis of TGA

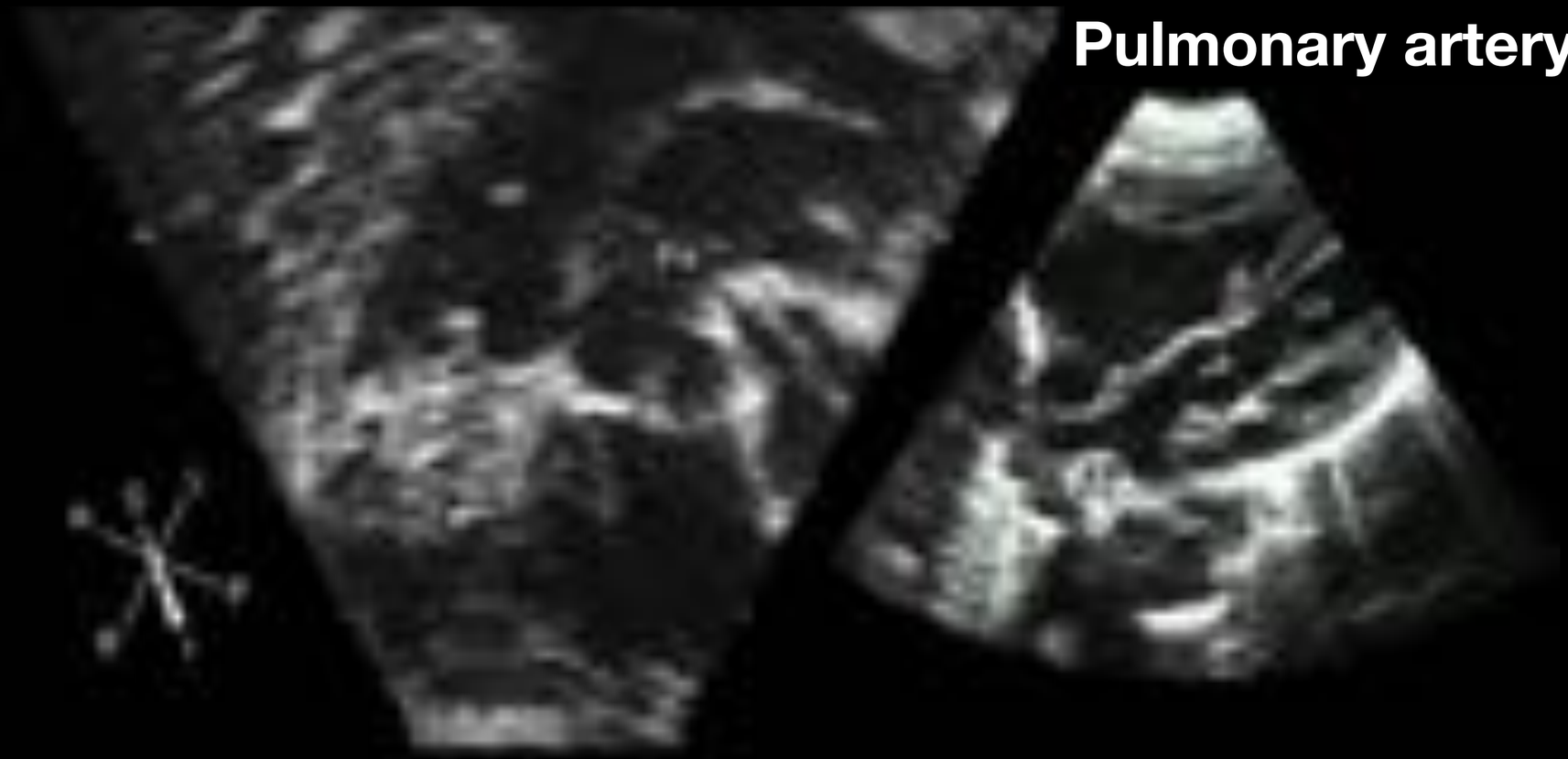
Isolated cyanosis



TGA

Rapid diagnosis

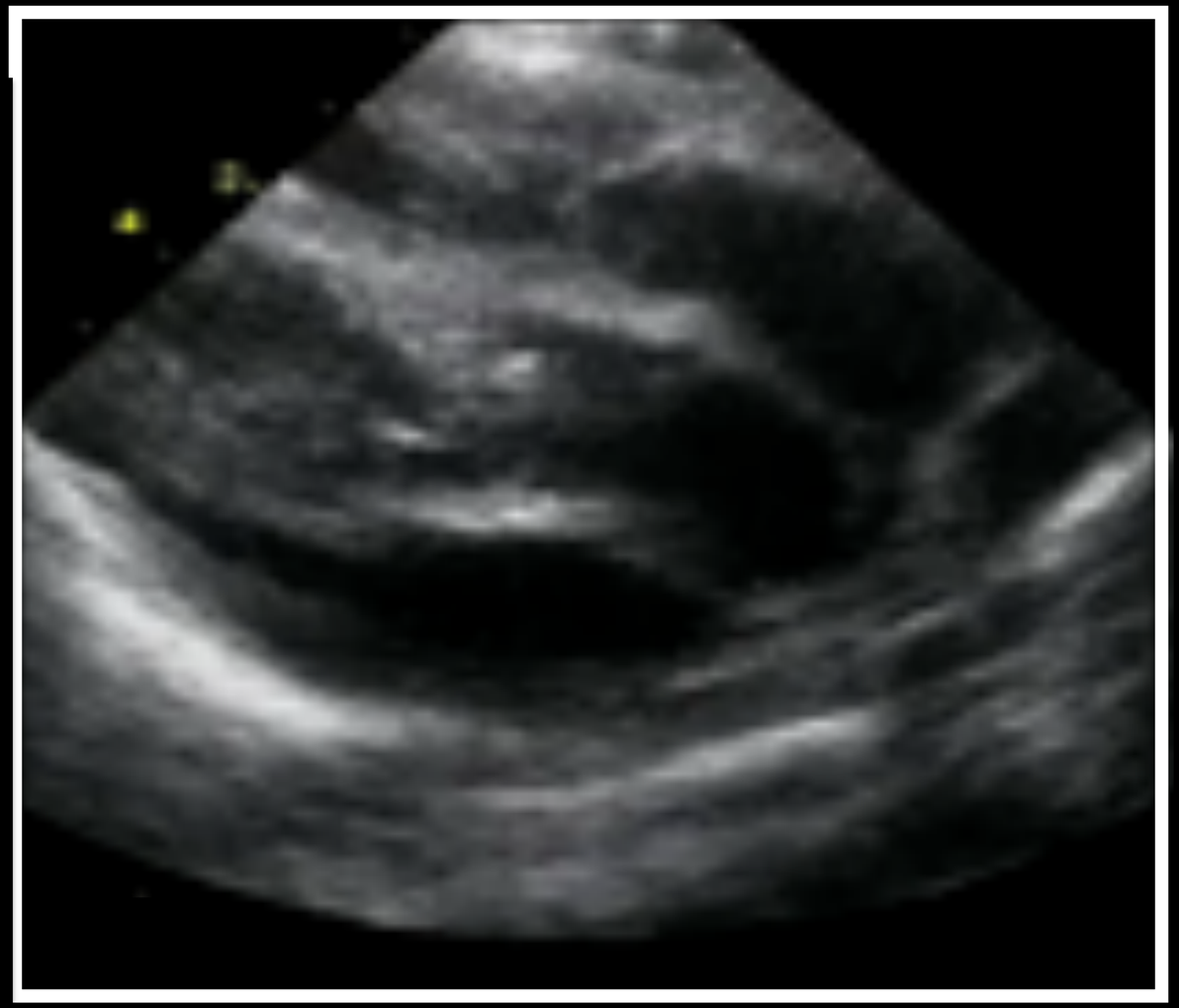
Parallel course of great vessels



Pulmonary artery from LV

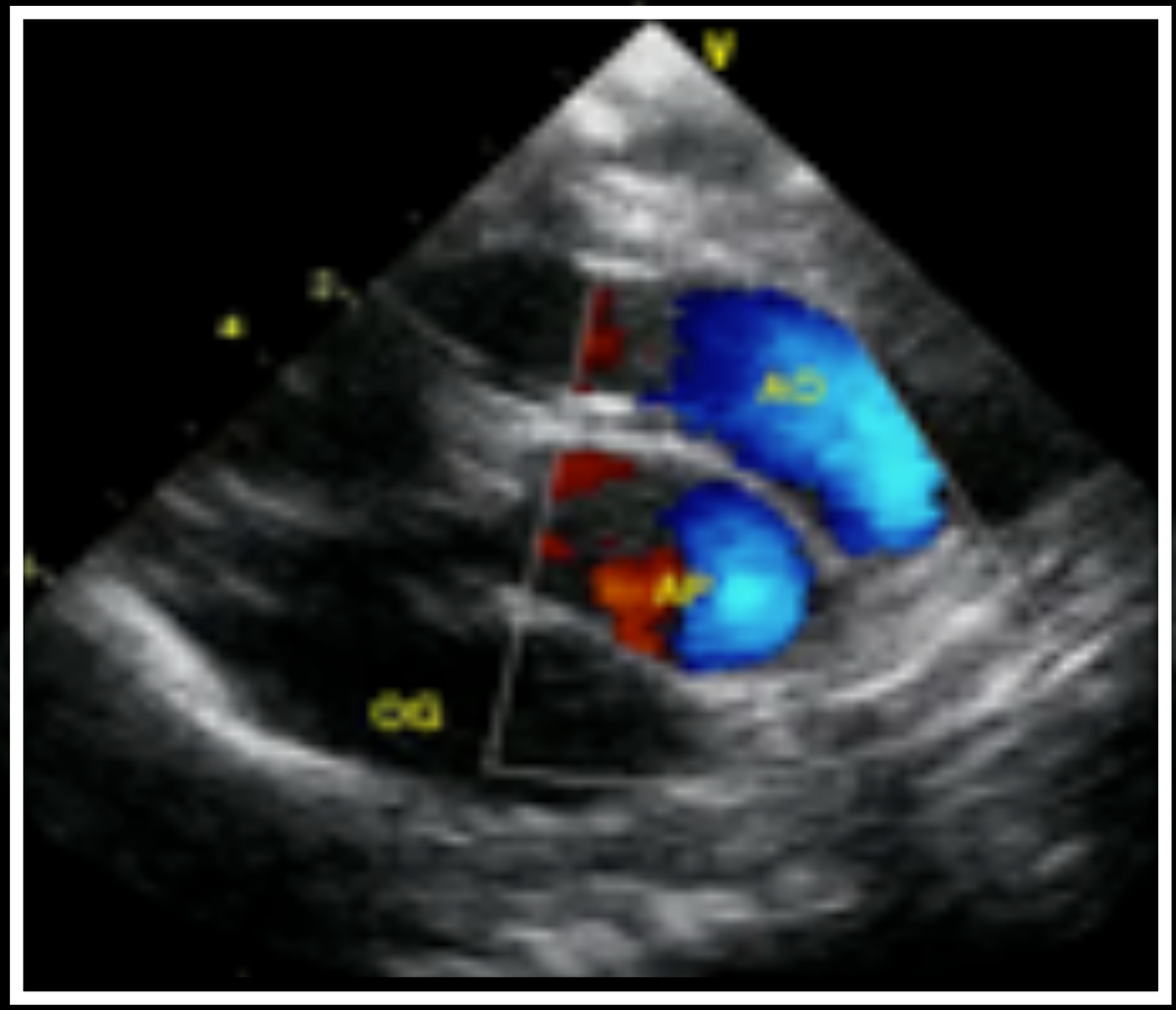
TGA

Rapid diagnosis



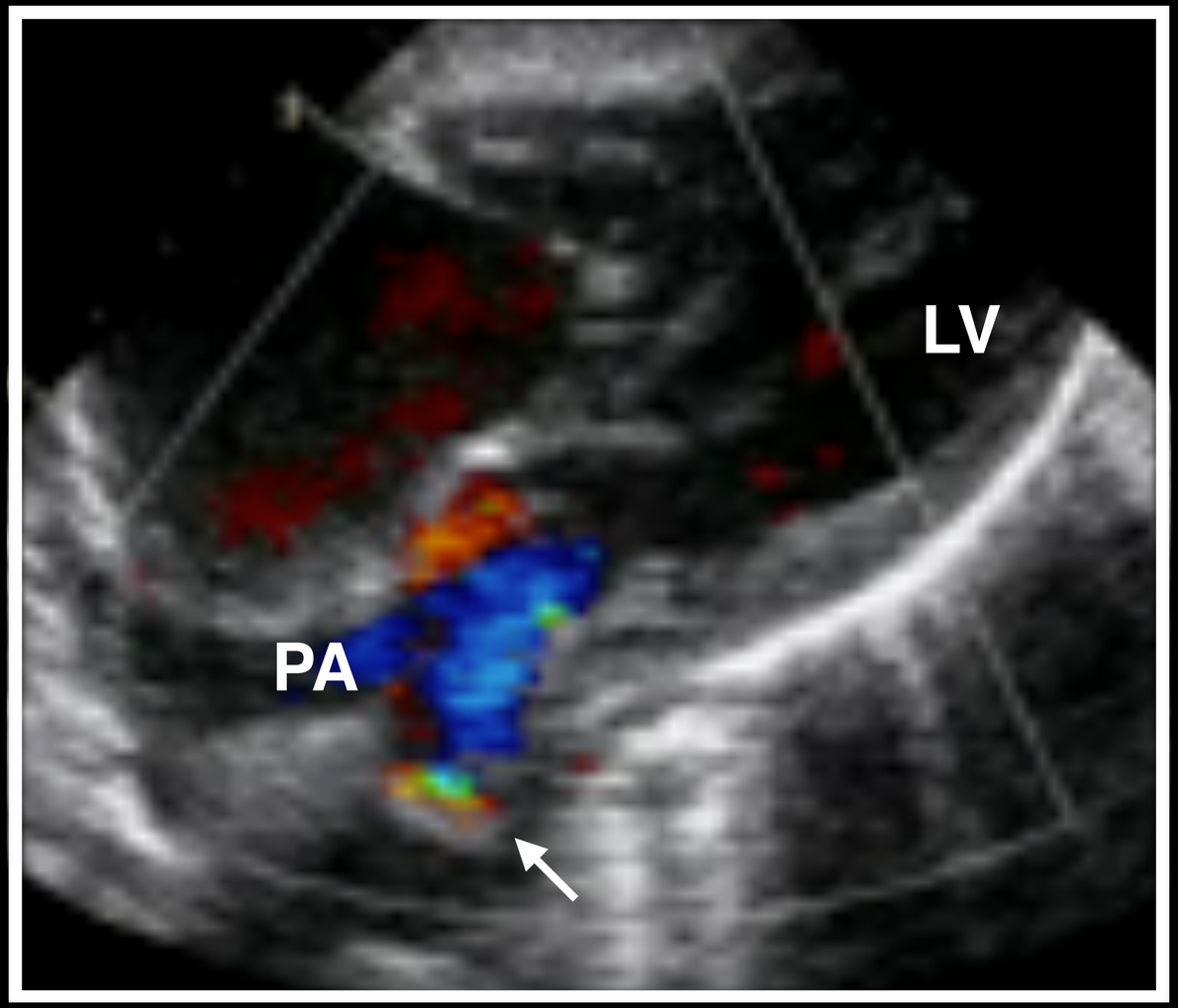
TGA

Rapid diagnosis



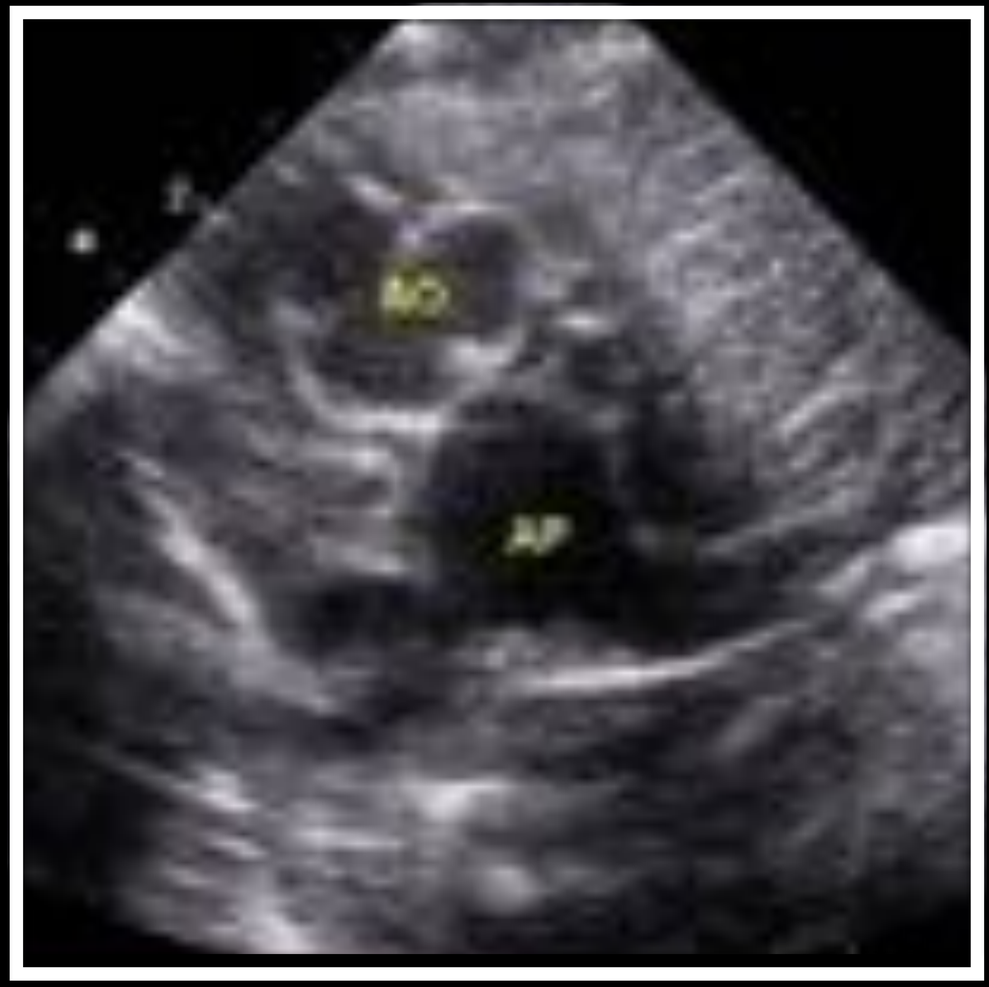
TGA

Rapid diagnosis

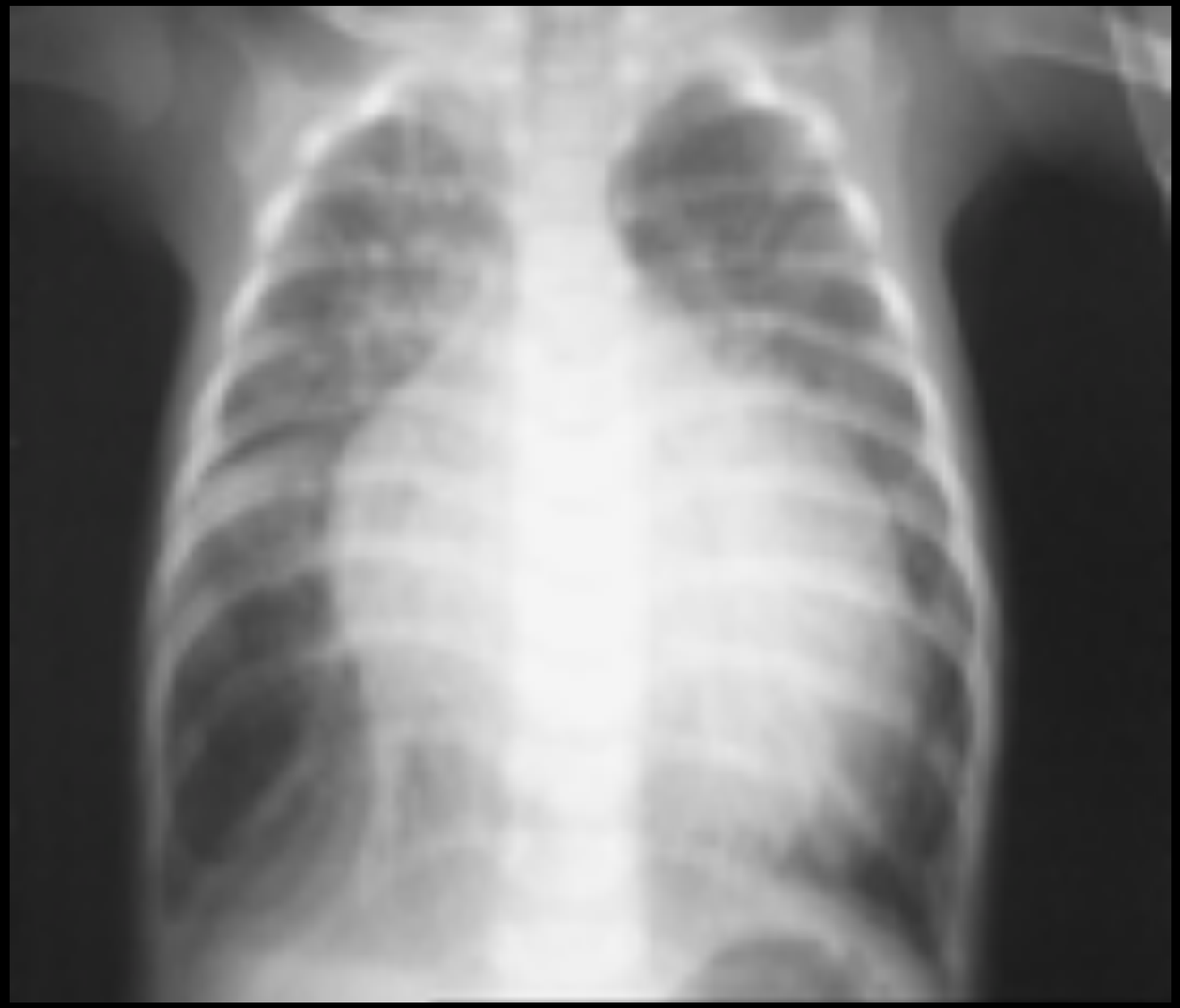


TGA

D-TGA



D-TGA



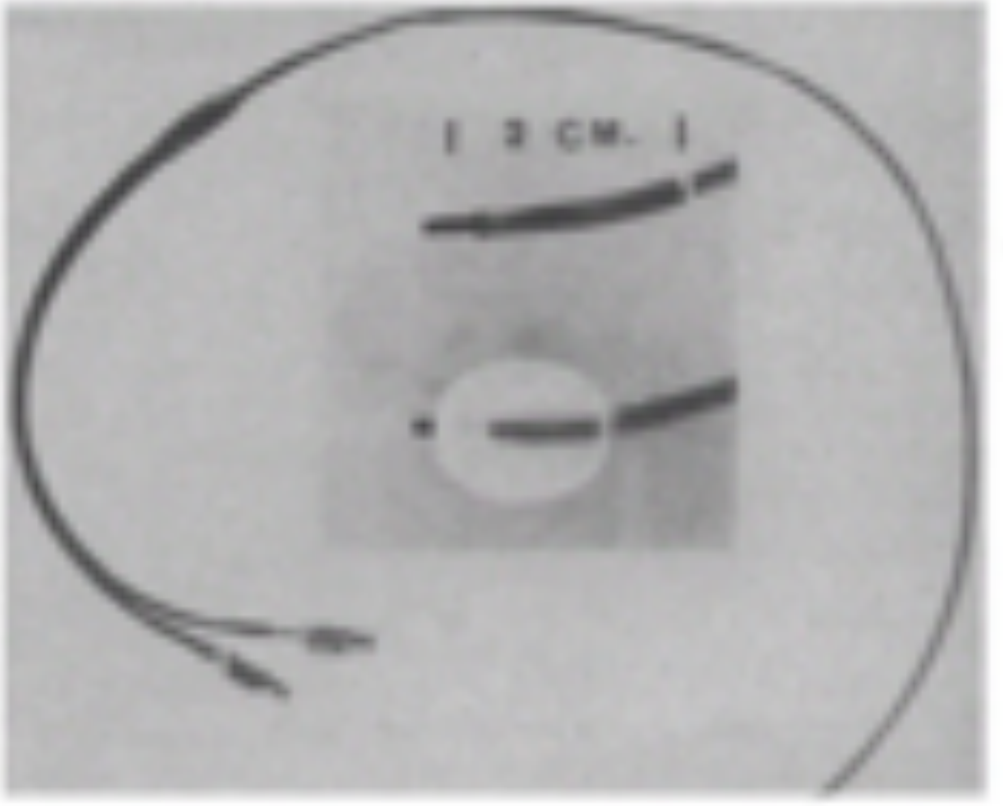
TGA with heart failure and restrictive PFO

Creation of an Atrial Septal Defect Without Thoracotomy

A Palliative Approach to Complete Transposition of the Great Arteries

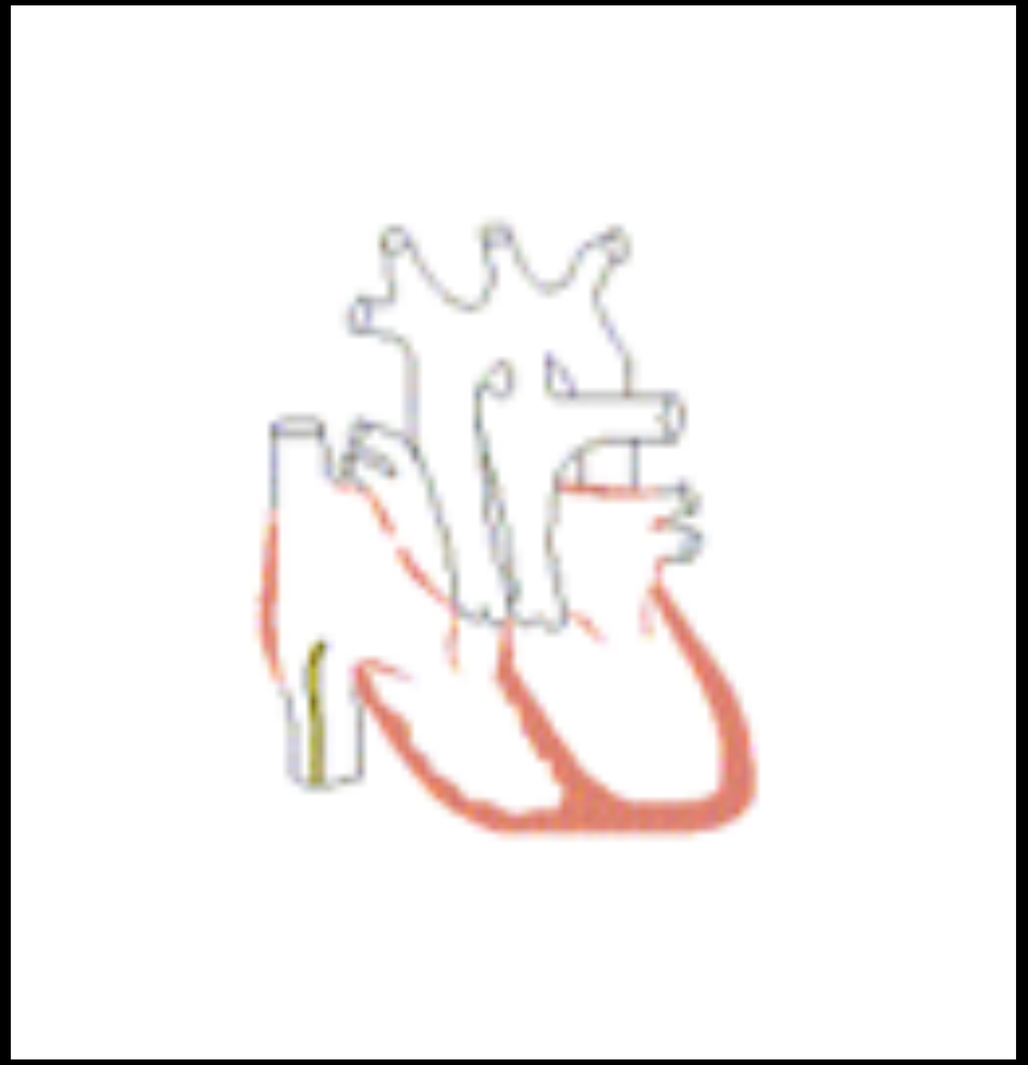
William J. Rashkind, MD, and William W. Miller, MD

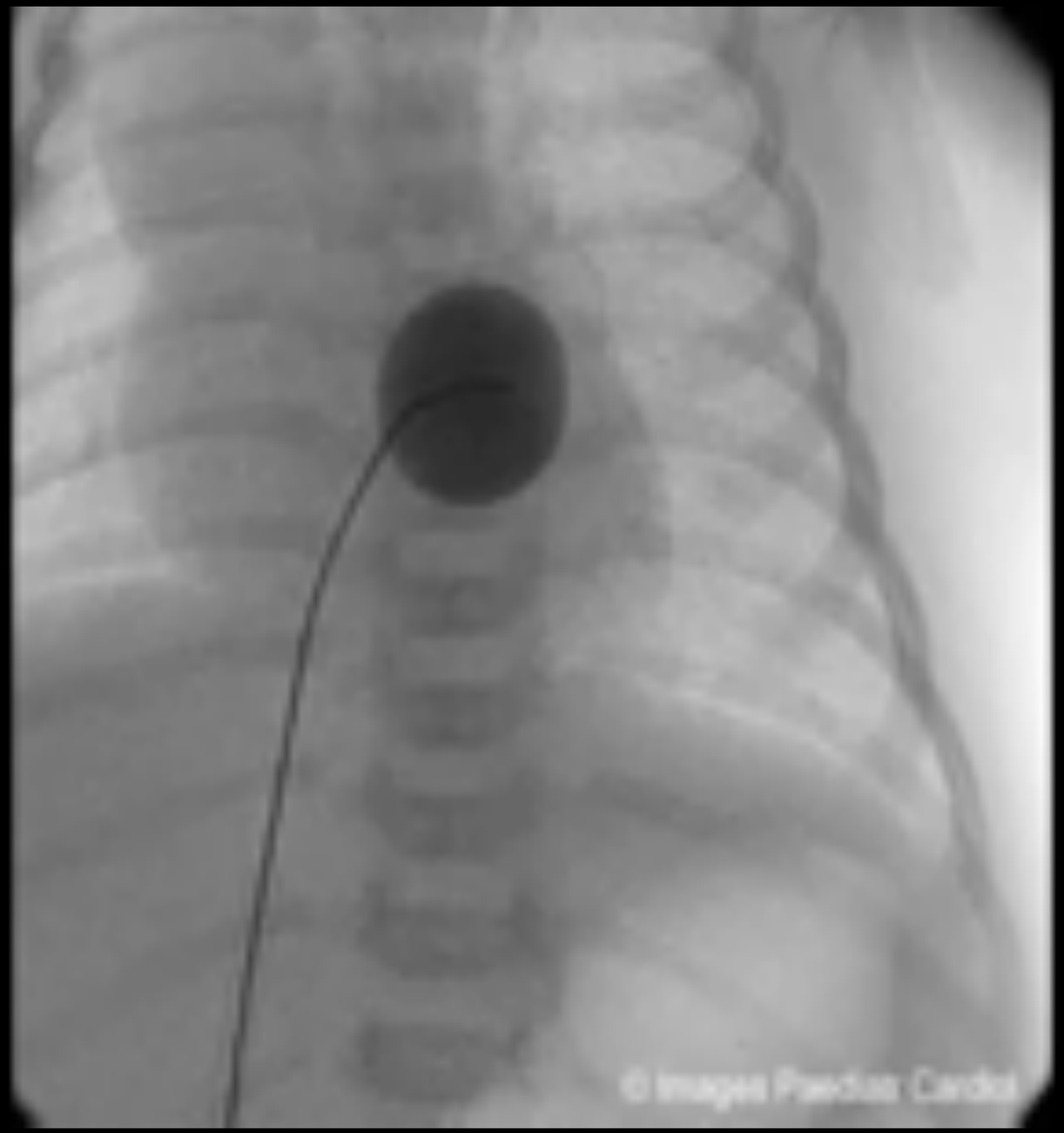
Transposition of the great vessels (TGV) occurs in approximately 50% of children who die with congenital heart disease.[1] With rare exceptions, patients with this lesion die in the first 6 months of life (30% within the first month). Approximately 40% of patients with TGV have an otherwise normal heart. In recent years, various types of complete corrections for this lesion have been attempted. Mustard et al[2] has simplified these procedures and has reduced mortality to reasonable levels. Best results are obtained in children well beyond 6 months of age. Therefore, it is imperative to provide early palliation that is effective until the optimal age for complete correction and that does not interfere significantly with subsequent surgery. Creation of an interatrial communication seems the best available choice to suit these requirements. The Blalock-Hanlon technique,[3] or some



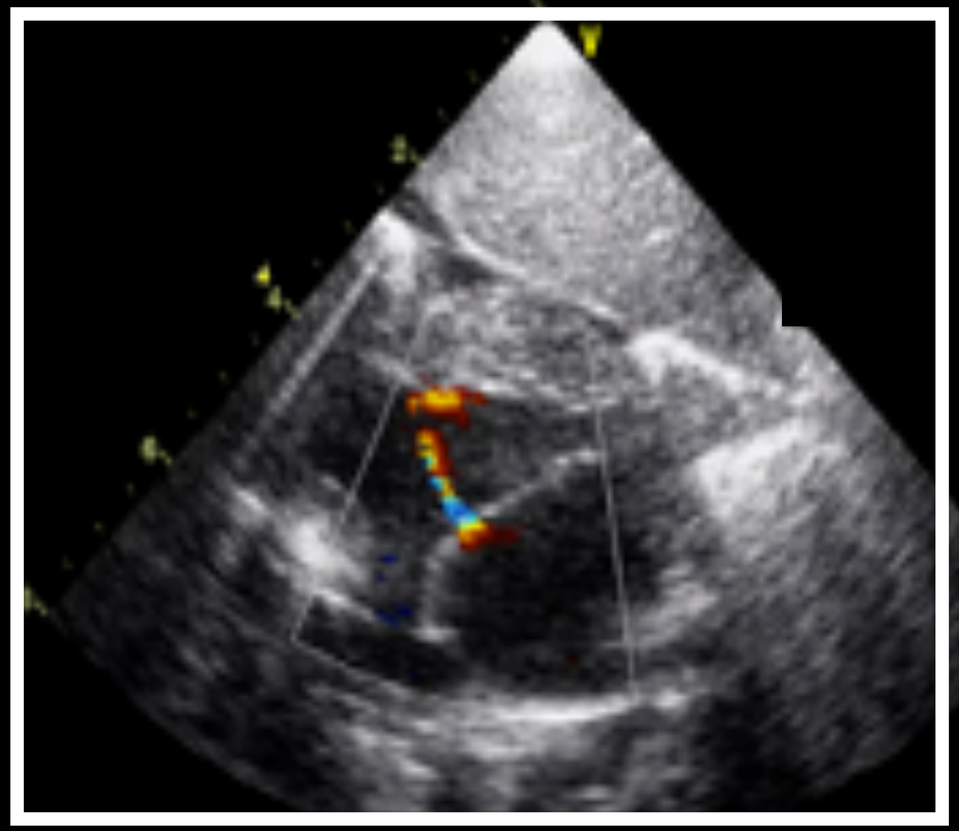
Rashkind WJ and Miller WW. JAMA 1966

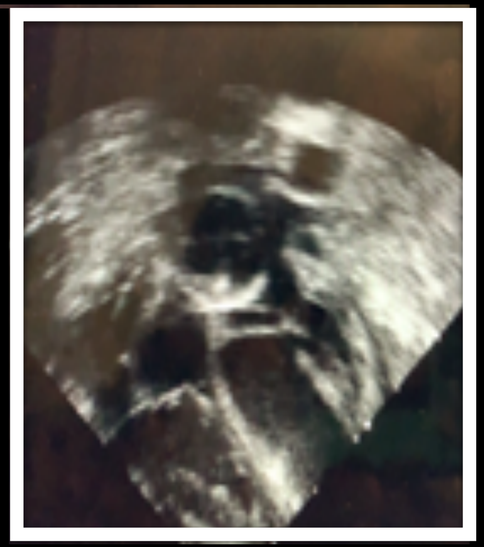
William Rashkind

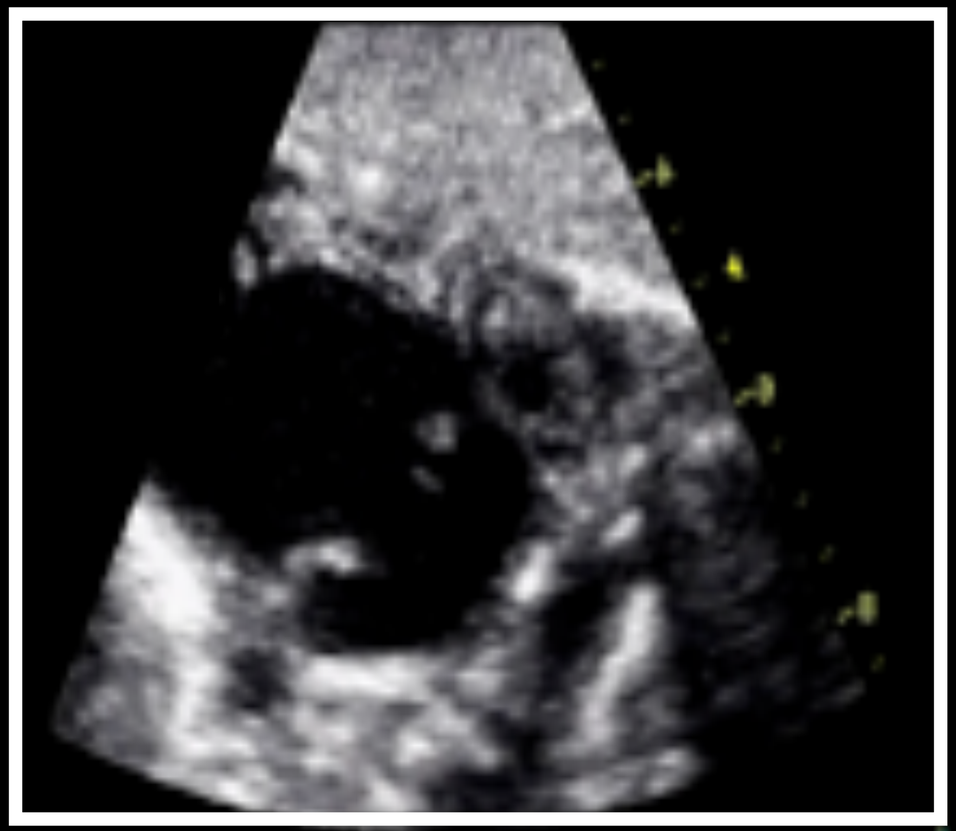




TGA Rashkind







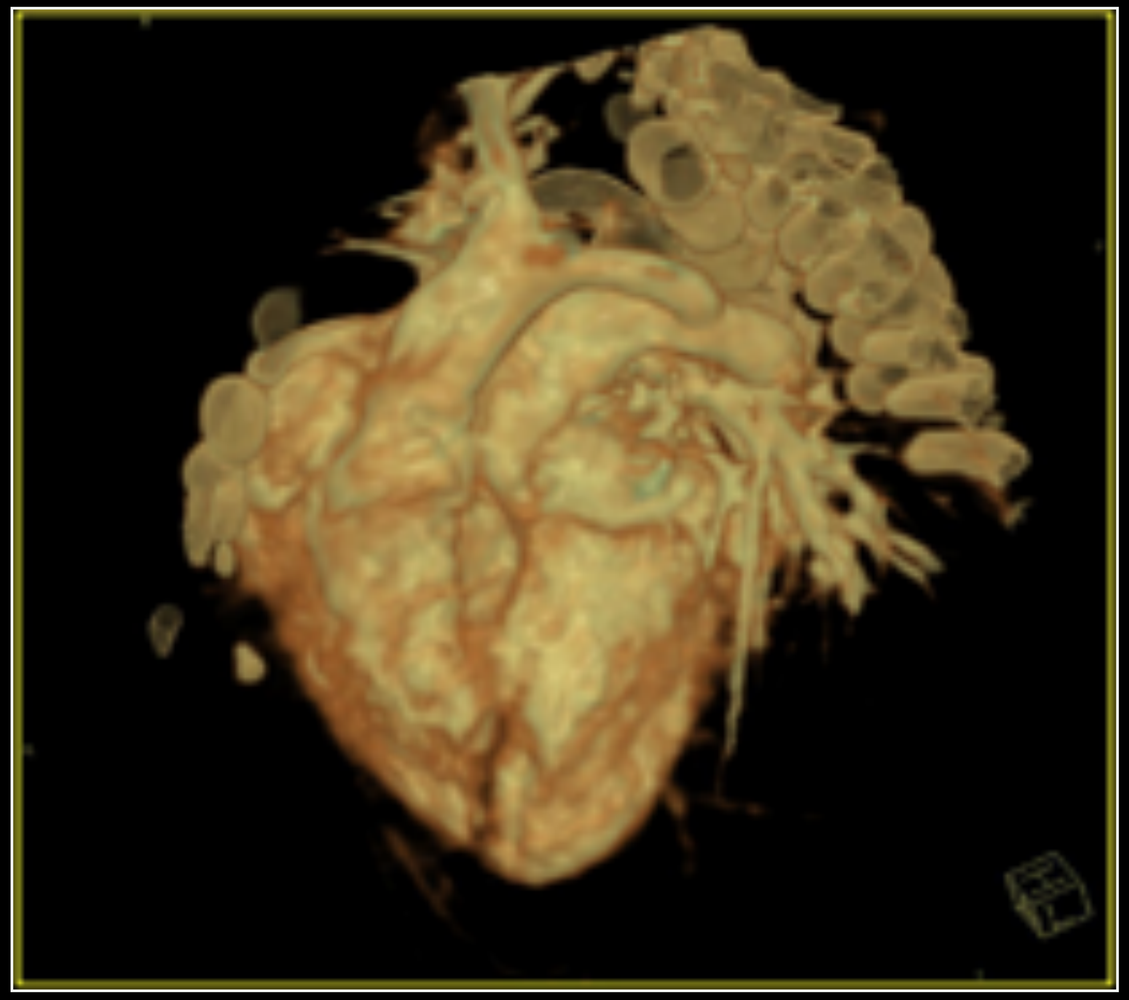
It is a challenging procedure that needs trained interventional/congenital cardiologists and a well prepared catheterization laboratory, with the possibility for surgical or circulatory back-up [1]

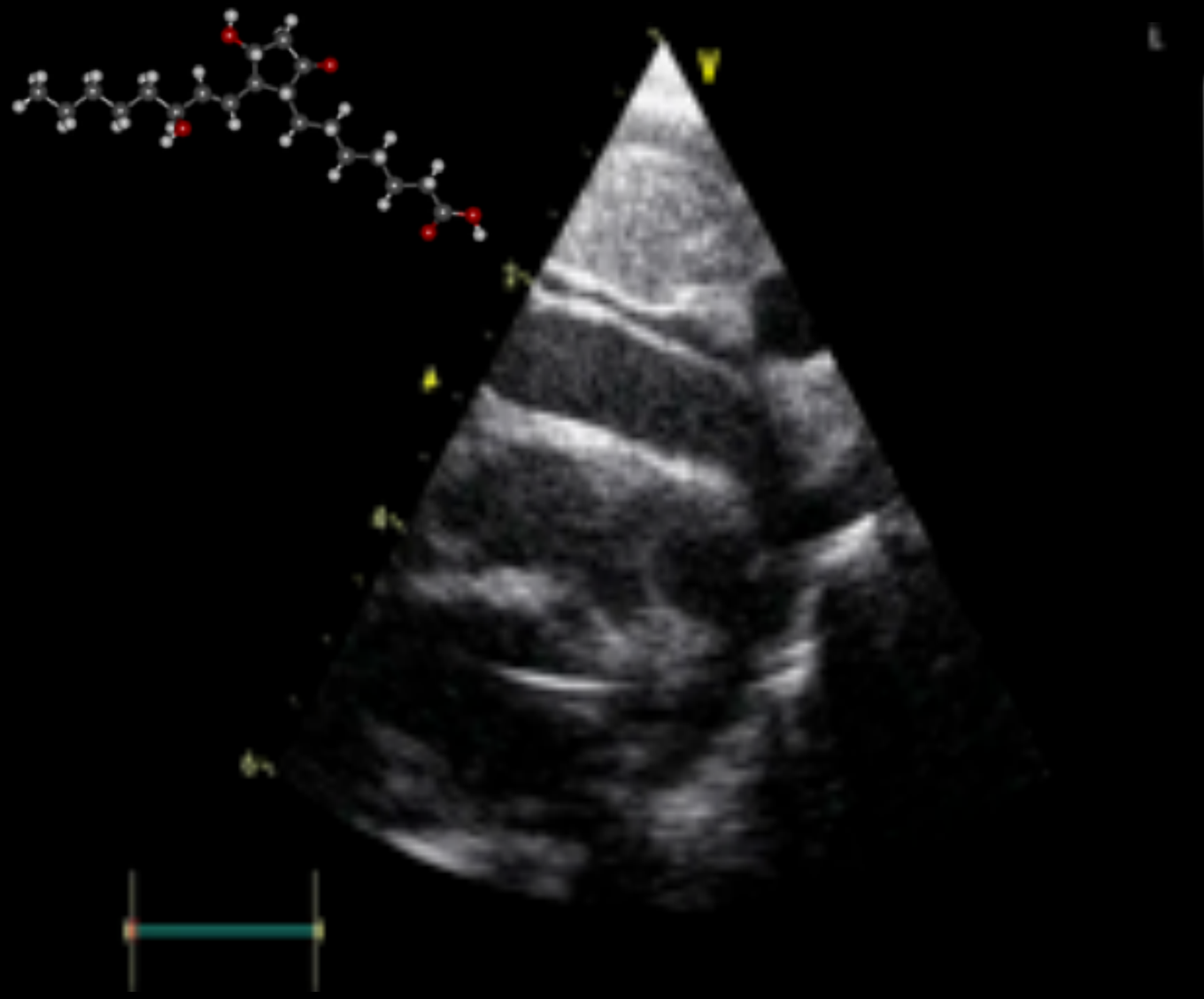
Balloon atrial septostomy performed out-of-hours produced higher complication rates as opposed to balloon atrial septostomy performed during routine hours. Only essential cases should be undertaken at night, and all other cases should be deferred to the daytime to limit unnecessary adverse complication [3]

Rashkind procedure was not associated with increased risk of necrotising enterocolitis, but was associated with nearly twice the risk of clinically recognised stroke (1% versus 0%, p = 0.046) [2]

1-Cinteza, Maedica (Buchar). 2013;8:280-284.
2-Mukherjee D Cardiol Young. 2010;20:373-80.
3-Vimalesmaran. Cardiol Young. 2013;23:61-7.

PGE1 and arterial duct





Echographic evaluation in TGA

Echocardiographic evaluation of TGA

1. Foramen ovale and arterial duct
2. Size of the ventricles
   Small RV : check aorta
   Small LV : check pulmonary artery
3. Atrioventricular valves anomalies
4. Coronary arteries

Peroperative analysis

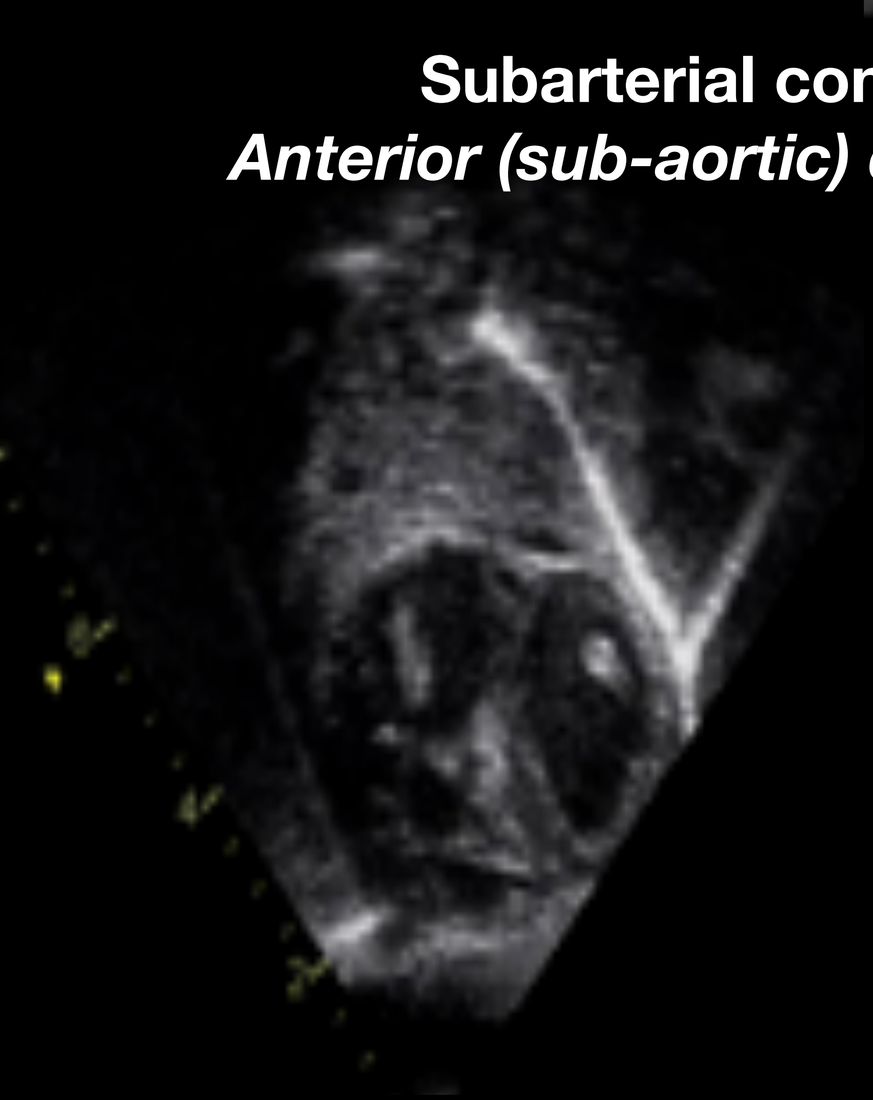
Anything different from the basic form

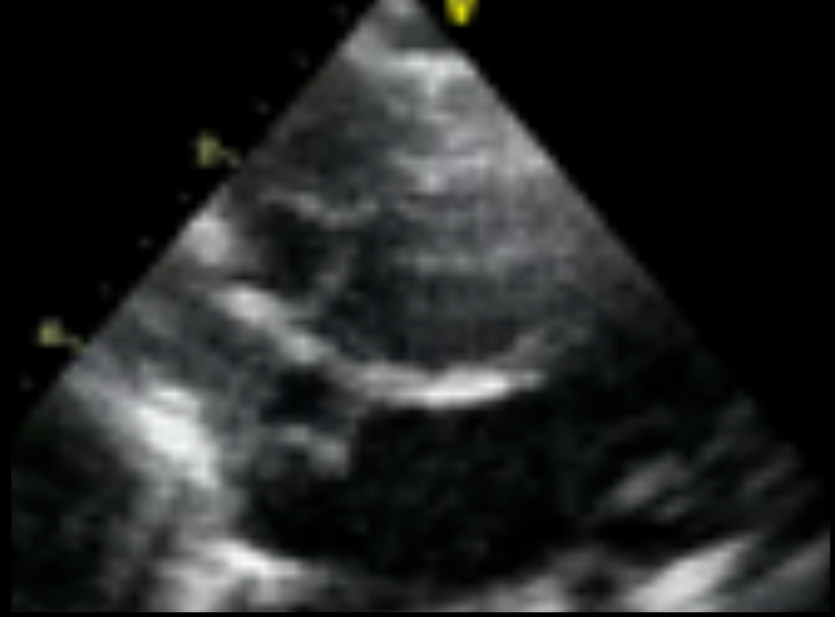
Expected surgical difficulties

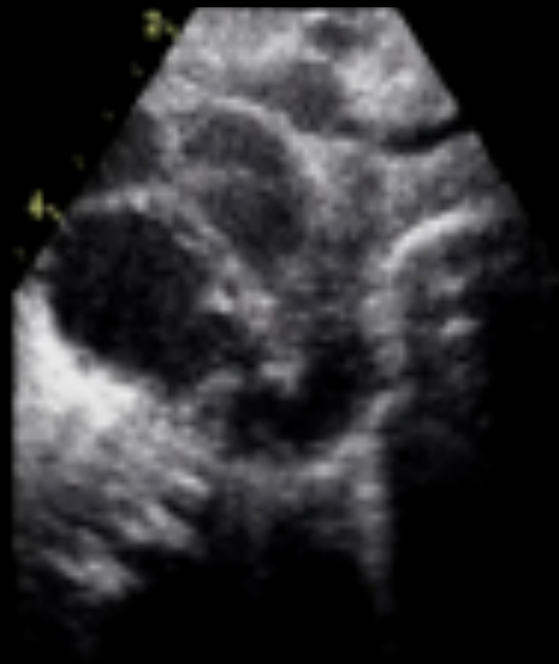
- « Abnormal » coronaries
- Hypoplastic aortic arch/coarctation
- VSD
- Difficult LV to PA routing
- Side by side vessels
- Aorto-pulmonary discrepancy
- Commissural mal-alignement

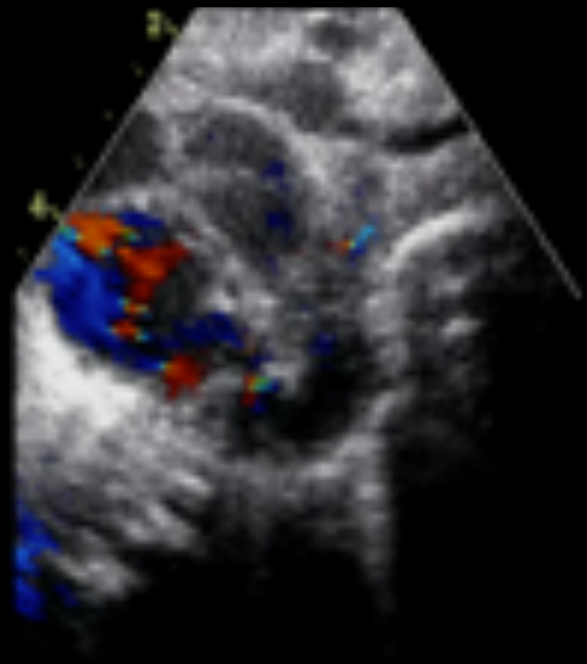
Subarterial conus

Anterior (sub-aortic) deviation



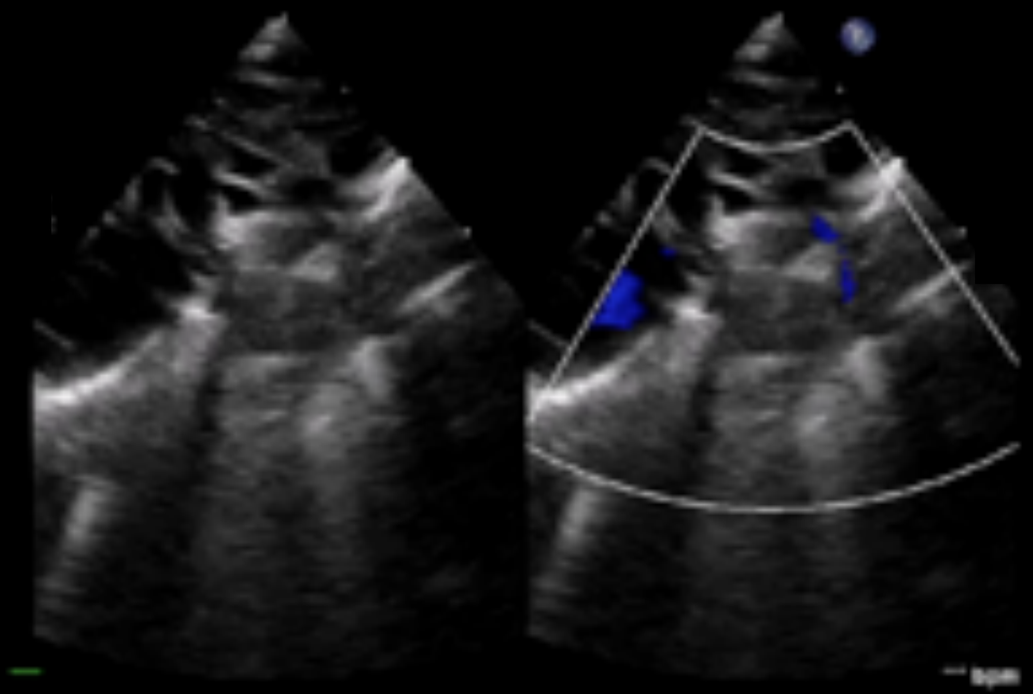






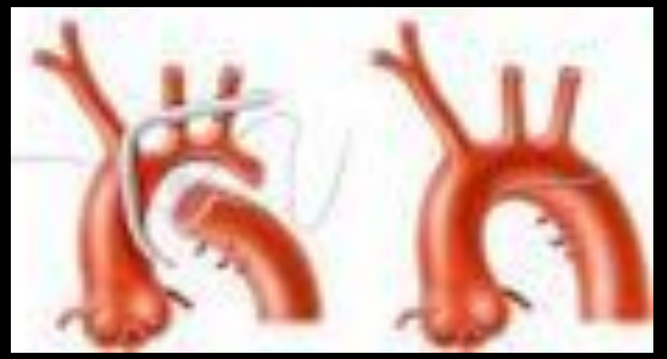
TGA VSD Coarctation

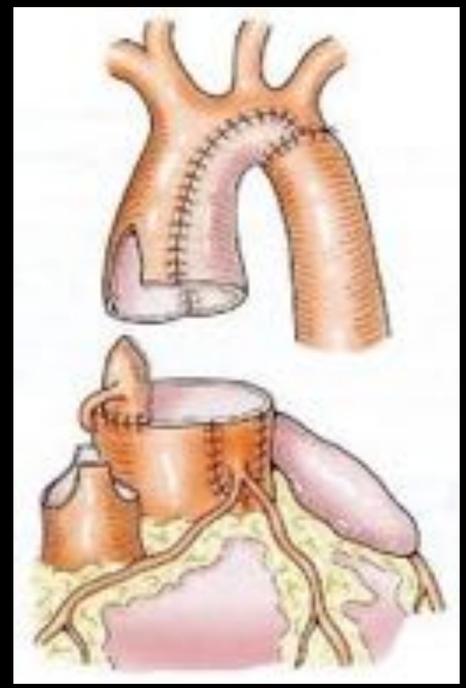
Aortic arch hypoplasia and coarctation



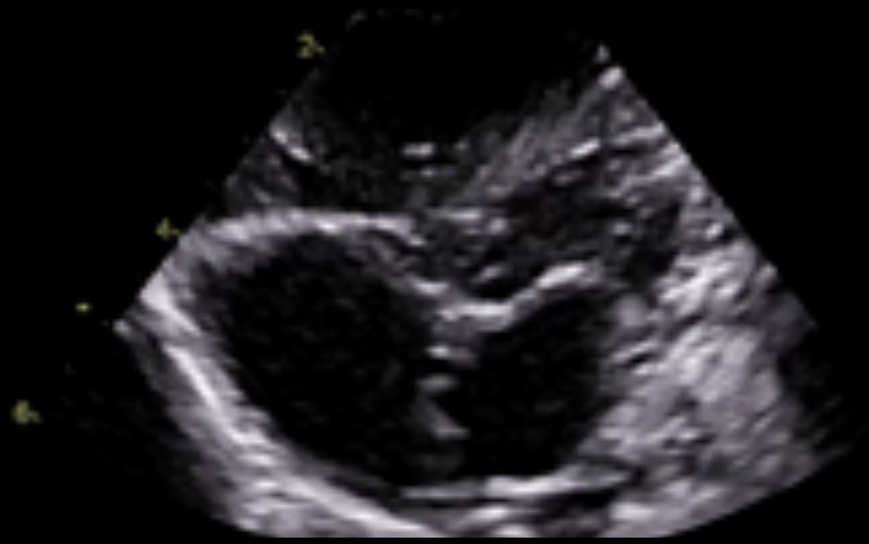
Localization and extension of the narrowed portion

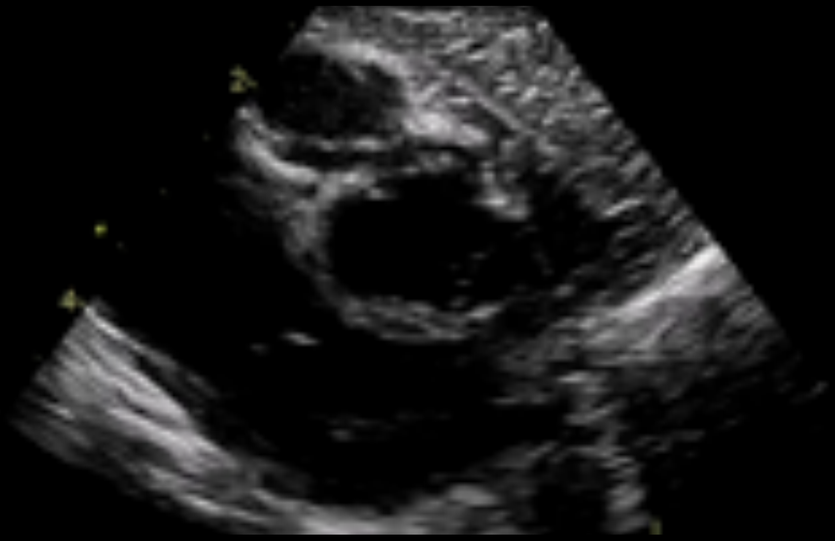
2 techniques: enlargement and extended end to end
=> Discrepancy between aortic and pulmonary roots





Aorto pulmonary discrepancy

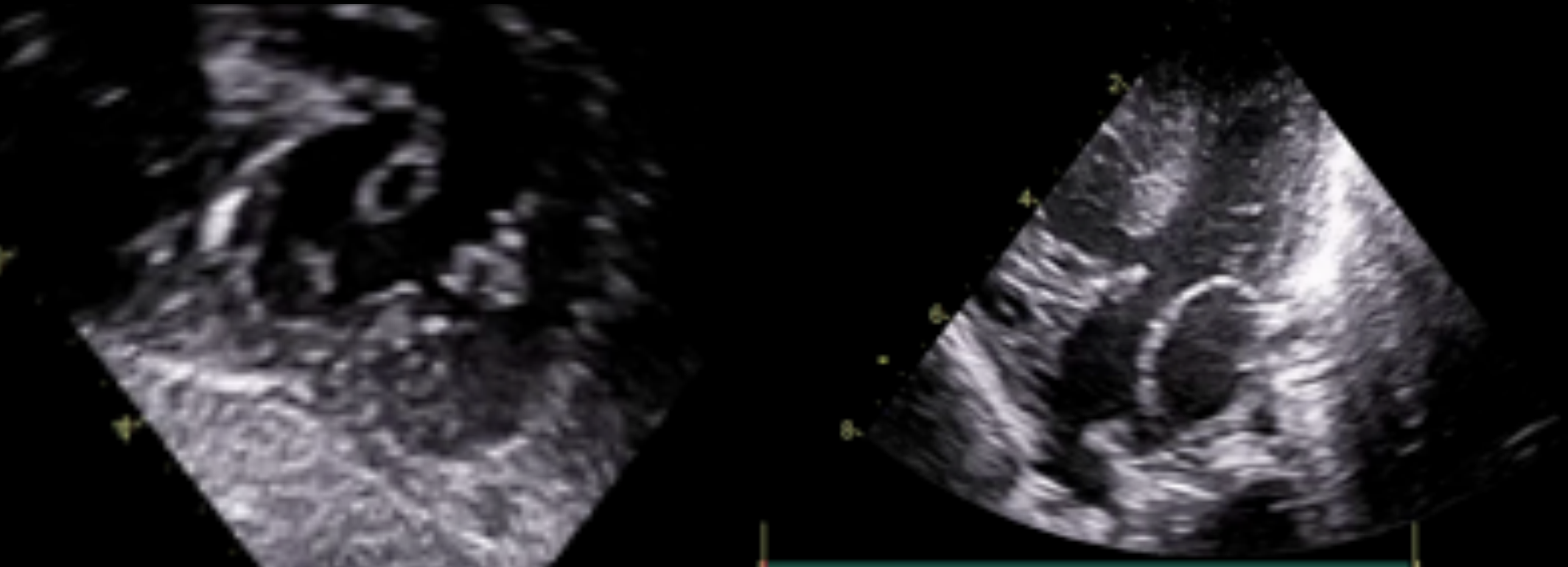




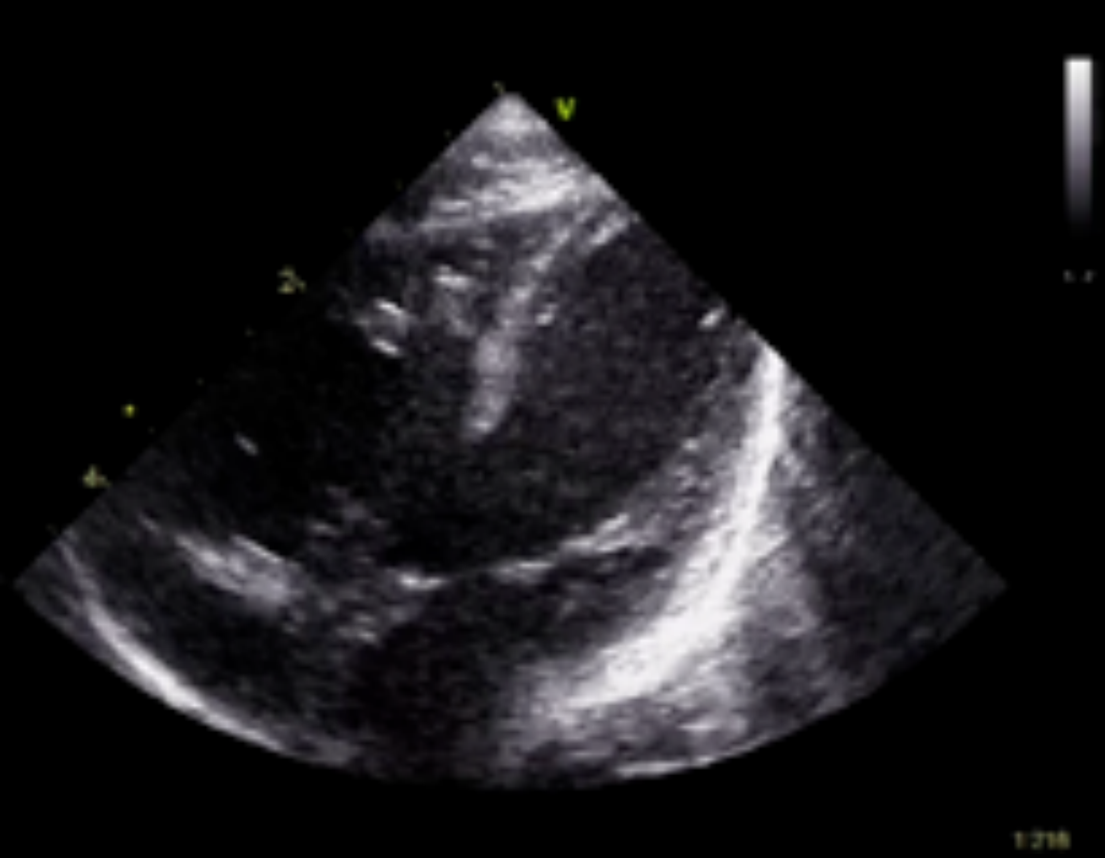
Impact for the type of repair

Subarterial conus

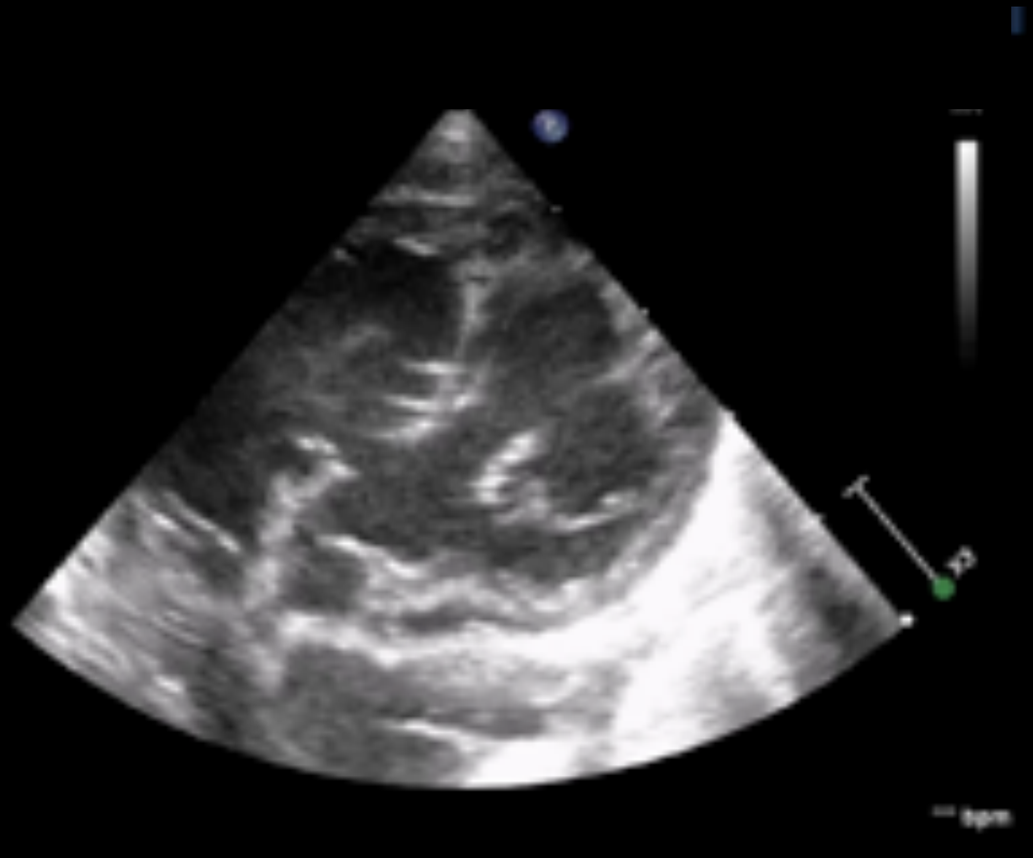
Posterior (sub-pulmonary) deviation



VSD: localization and size

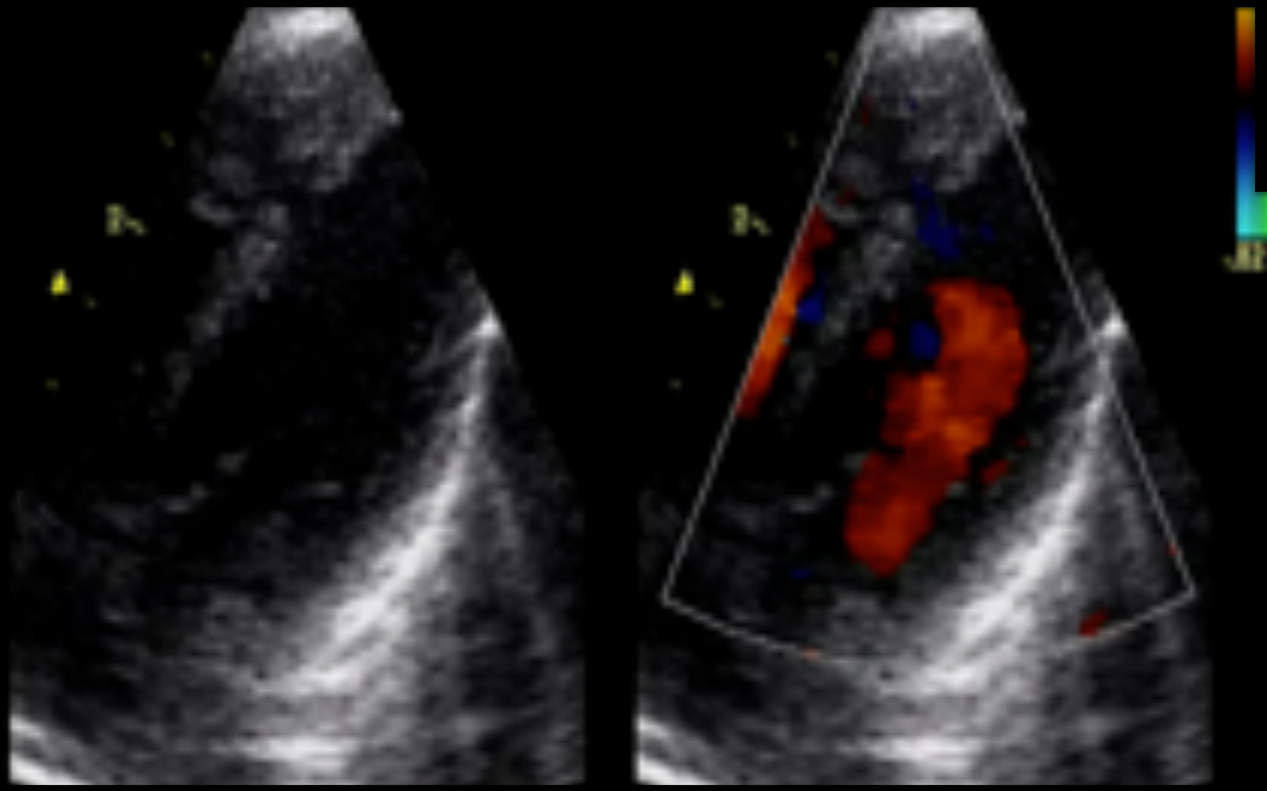


Inlet VSD

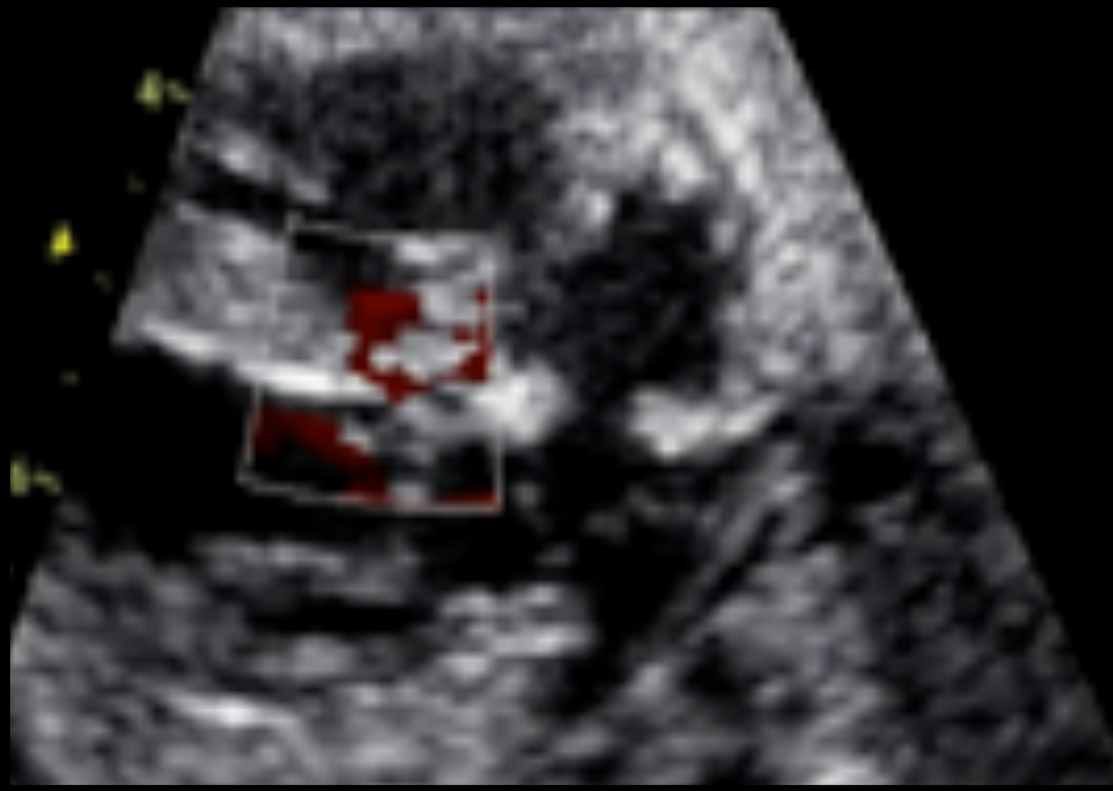


Outlet VSD

VSD: localization and size



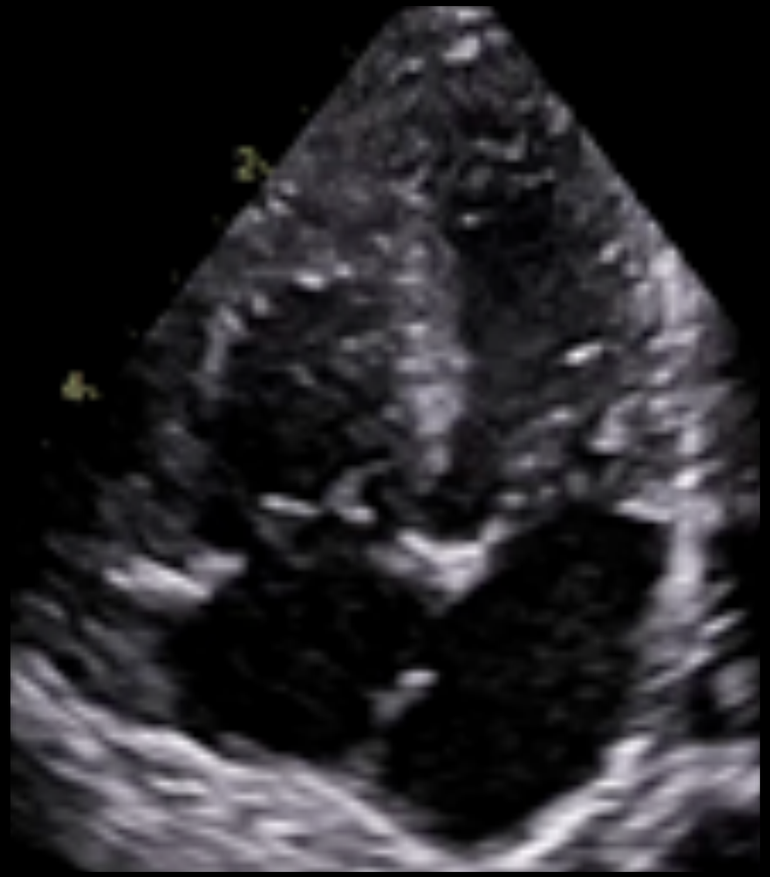
Muscular VSD



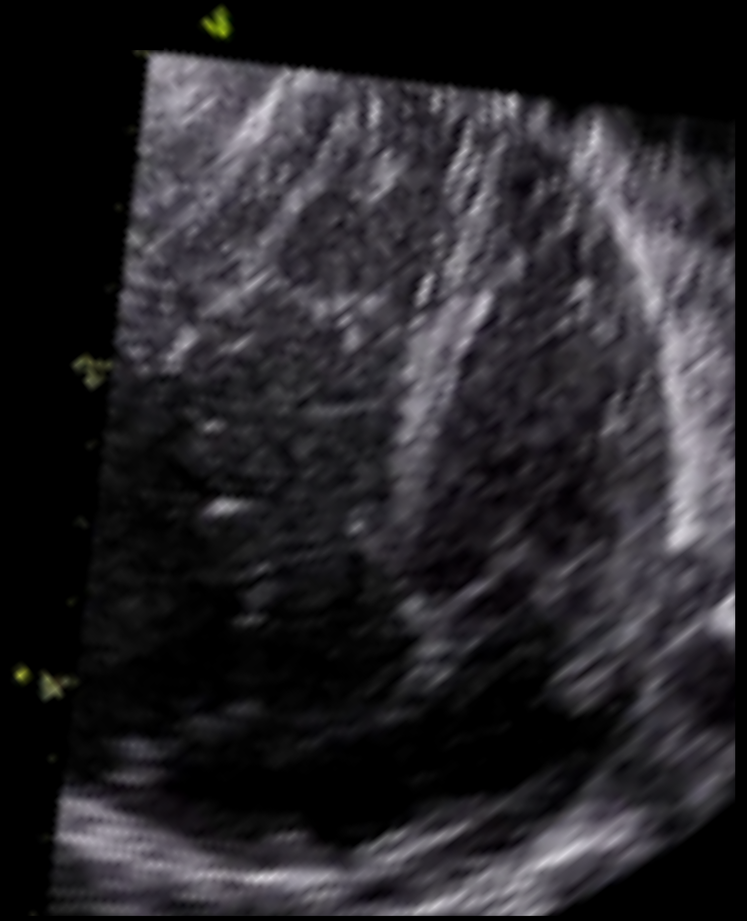
Perimembranous VSD

Unbalanced ventricles

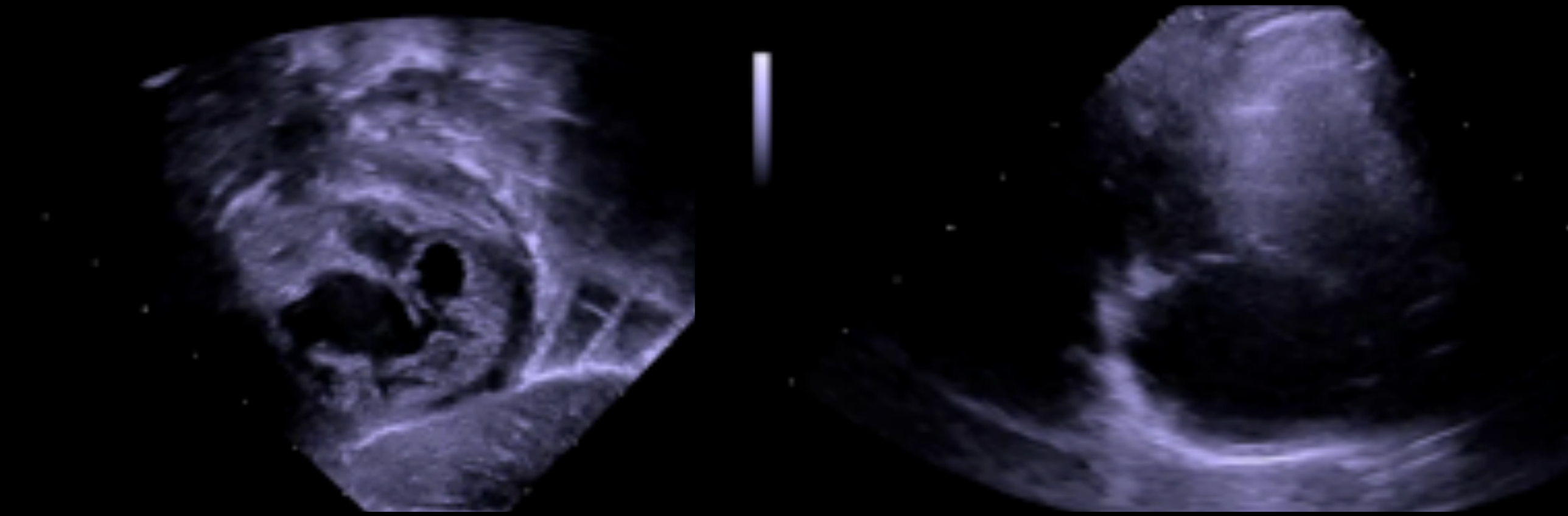
« Small » RV



« Small » LV

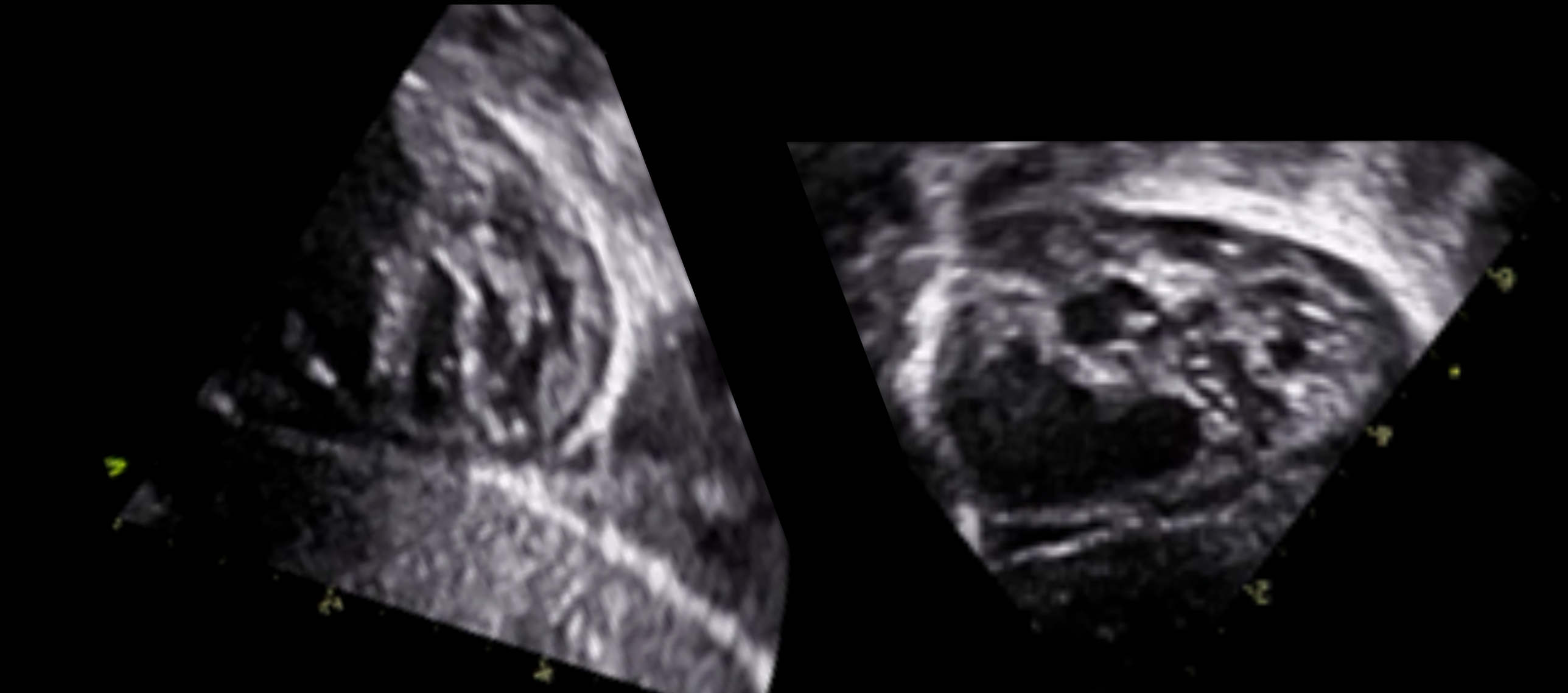


AV valves abnormalities
Straddling and over-riding

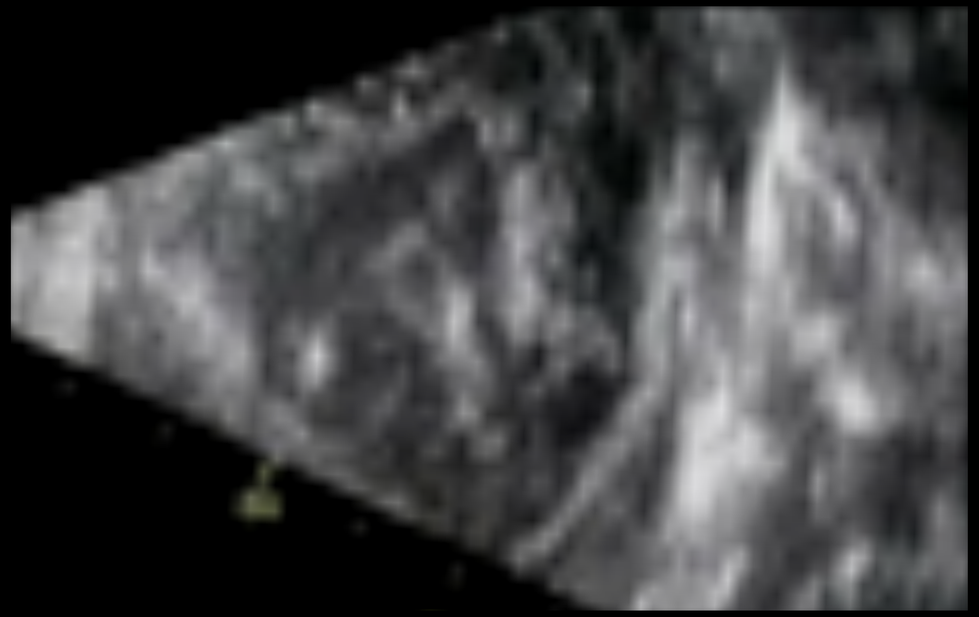


Straddling of tricuspid valve

AV valves abnormalities
Mitral cleft and subpulmonary obstruction

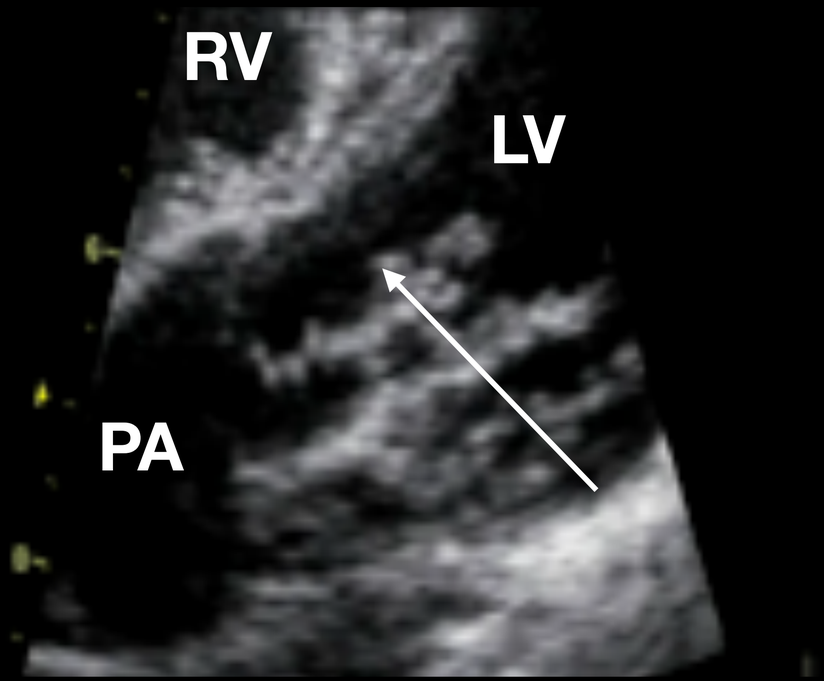


AV valves abnormalities
Mitral cleft

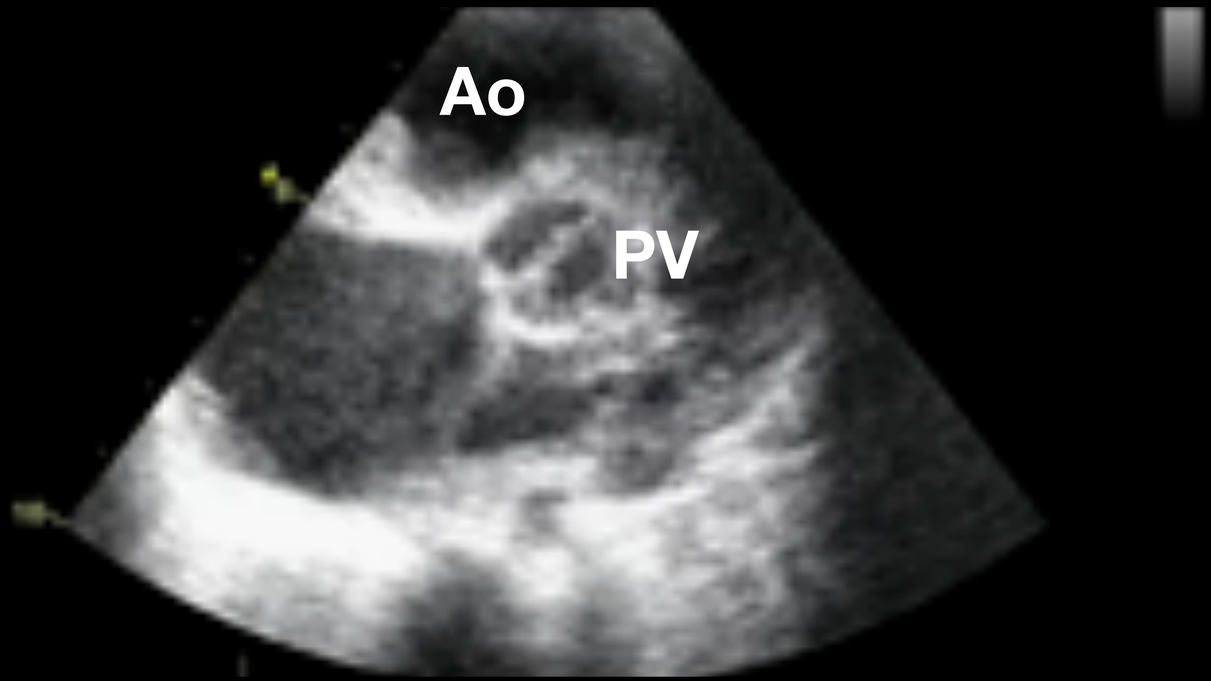


Mitral cleft

TGA left outflow tract obstruction

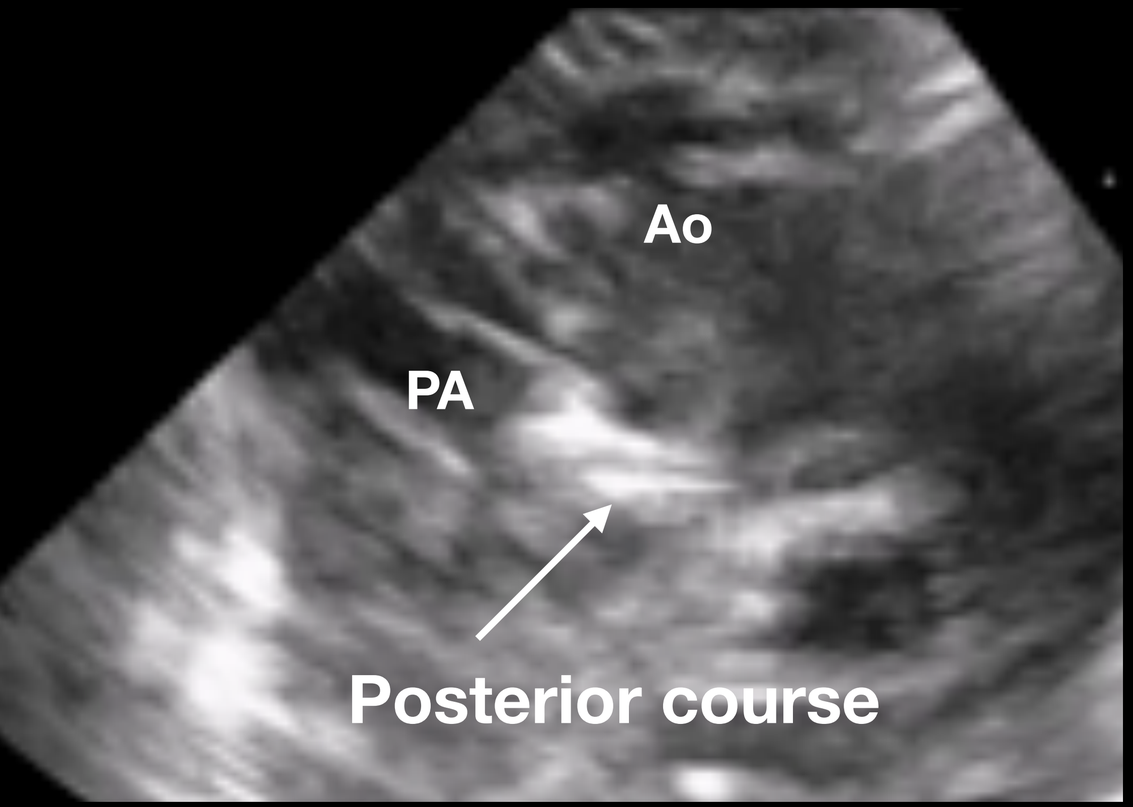


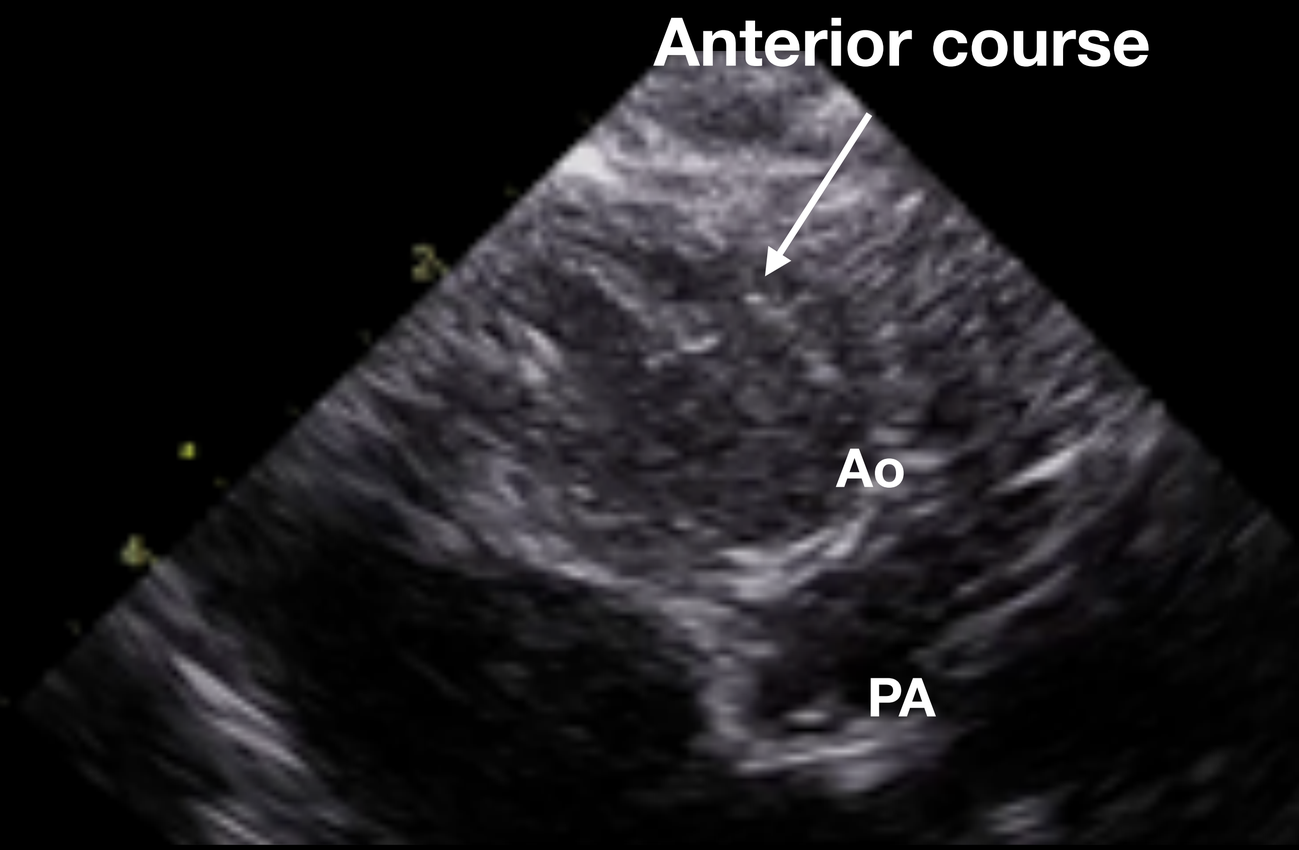
Accessory tissue on mitral valve



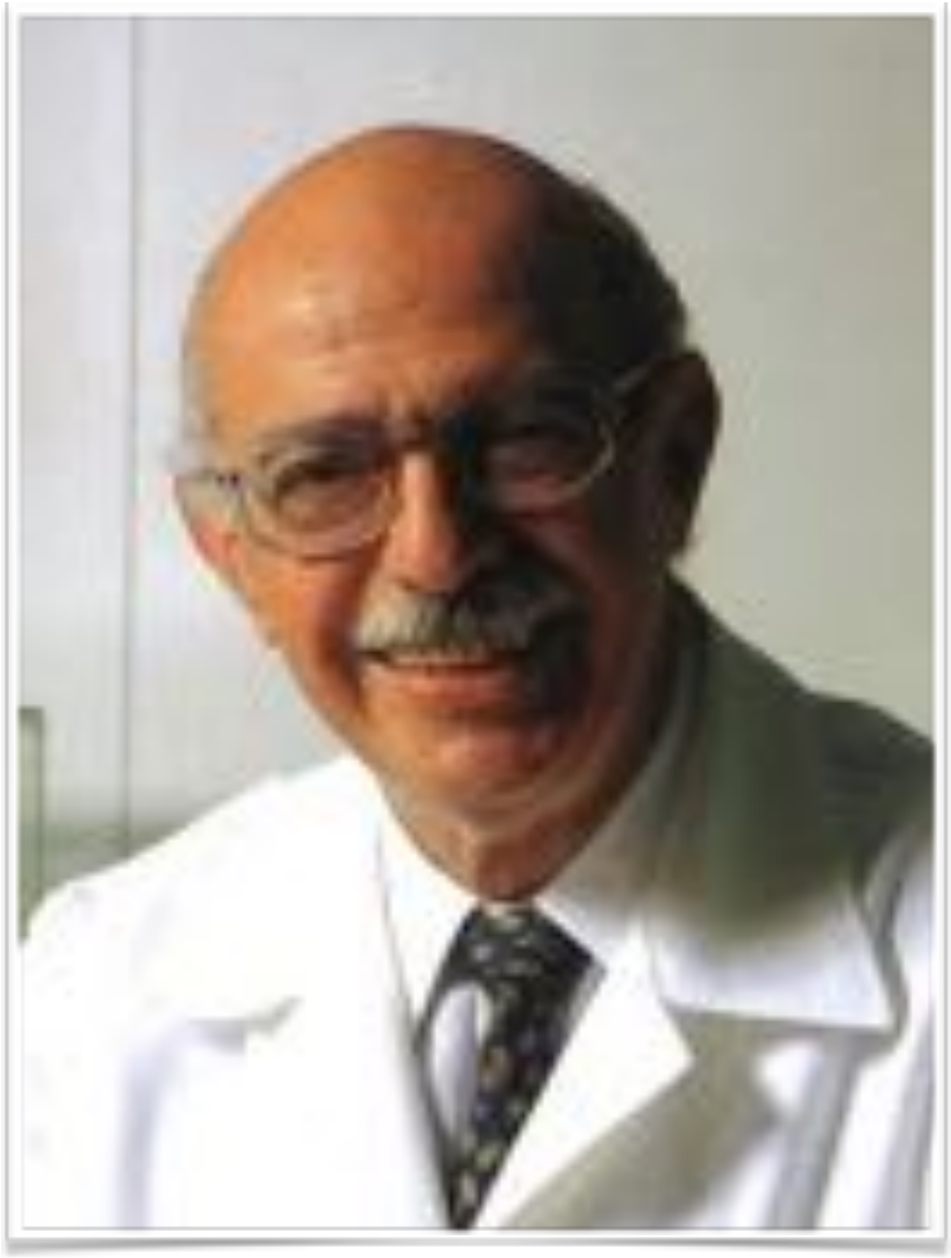
Bicuspid pulmonary valve

Coronary evaluation





The arterial switch operation



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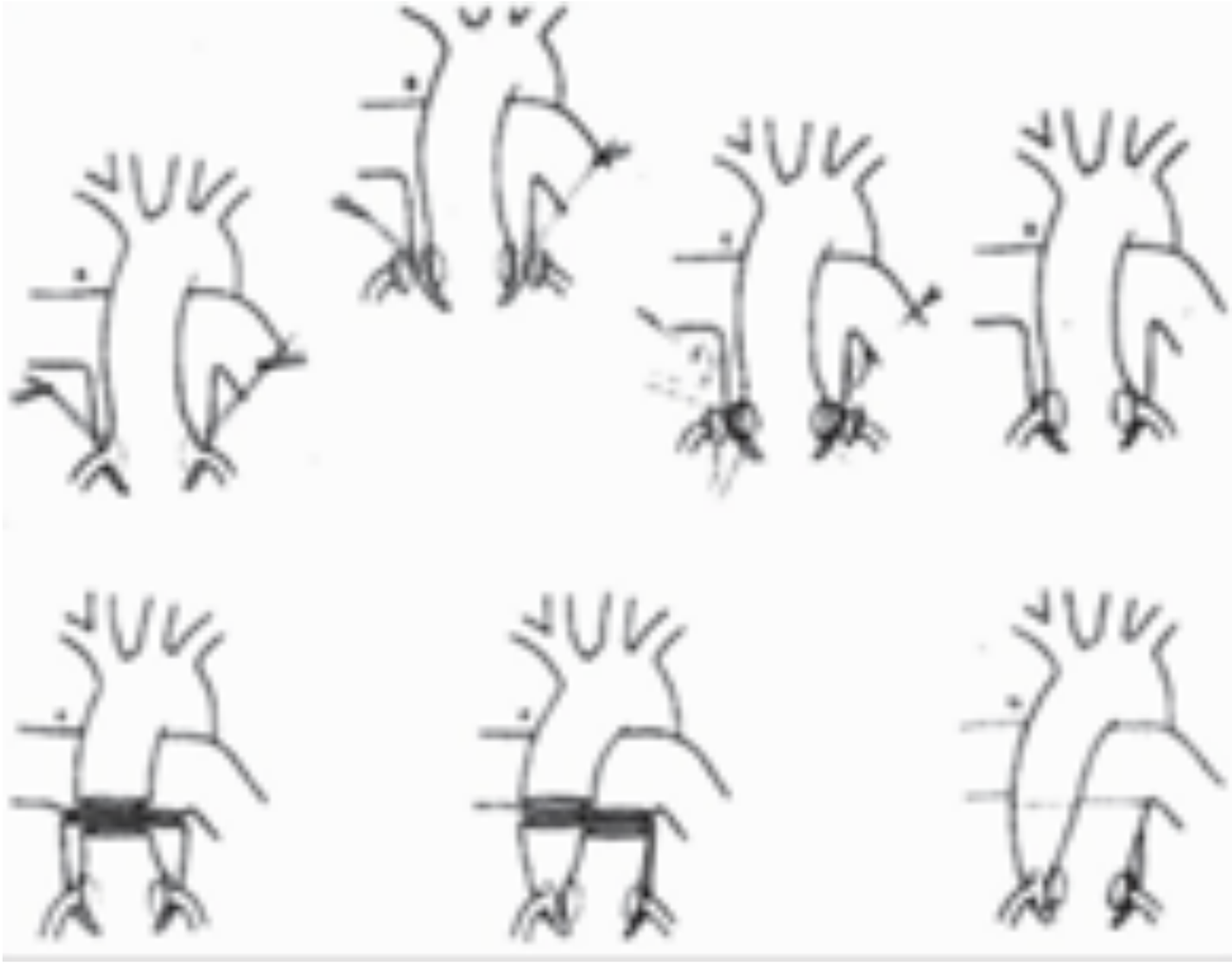
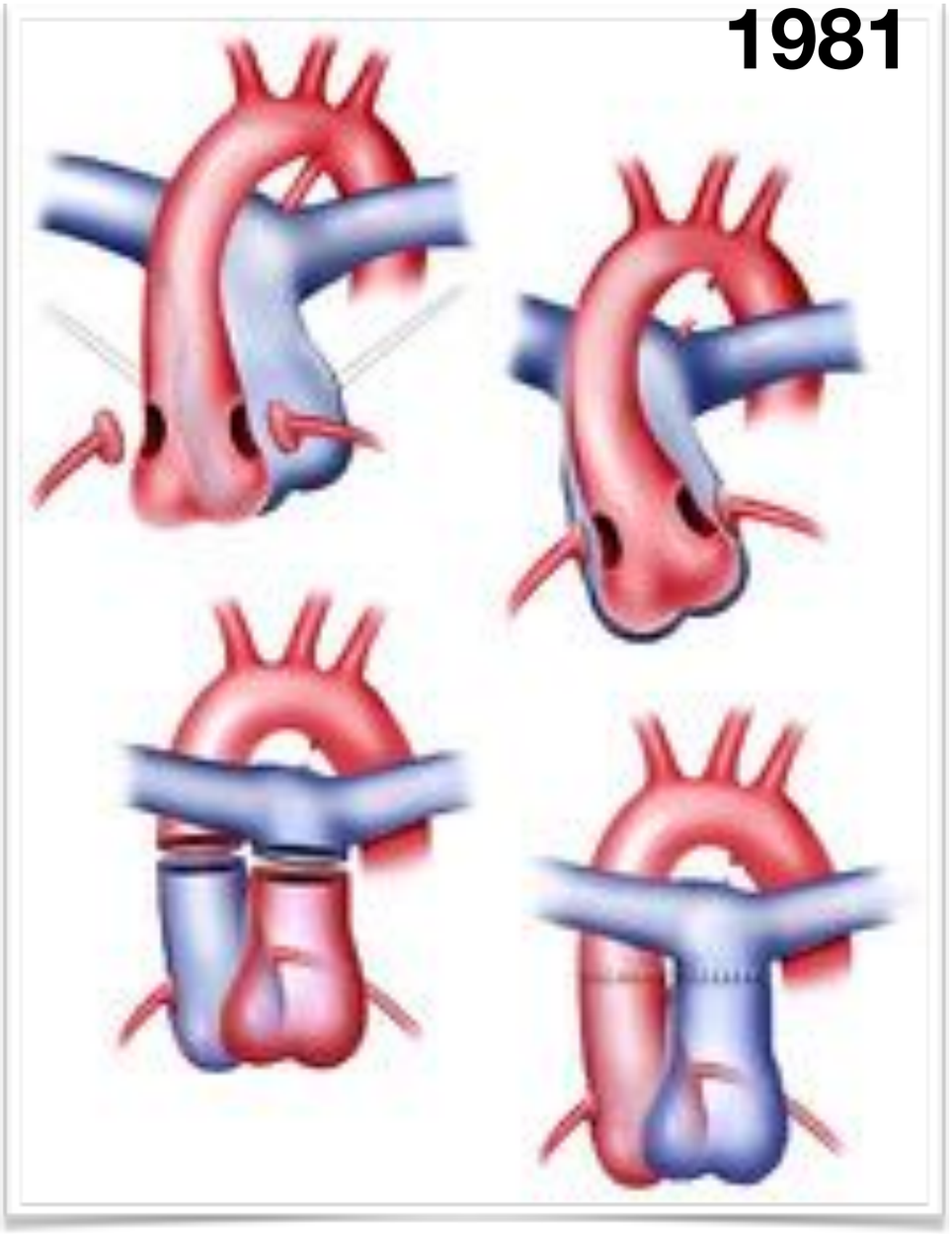
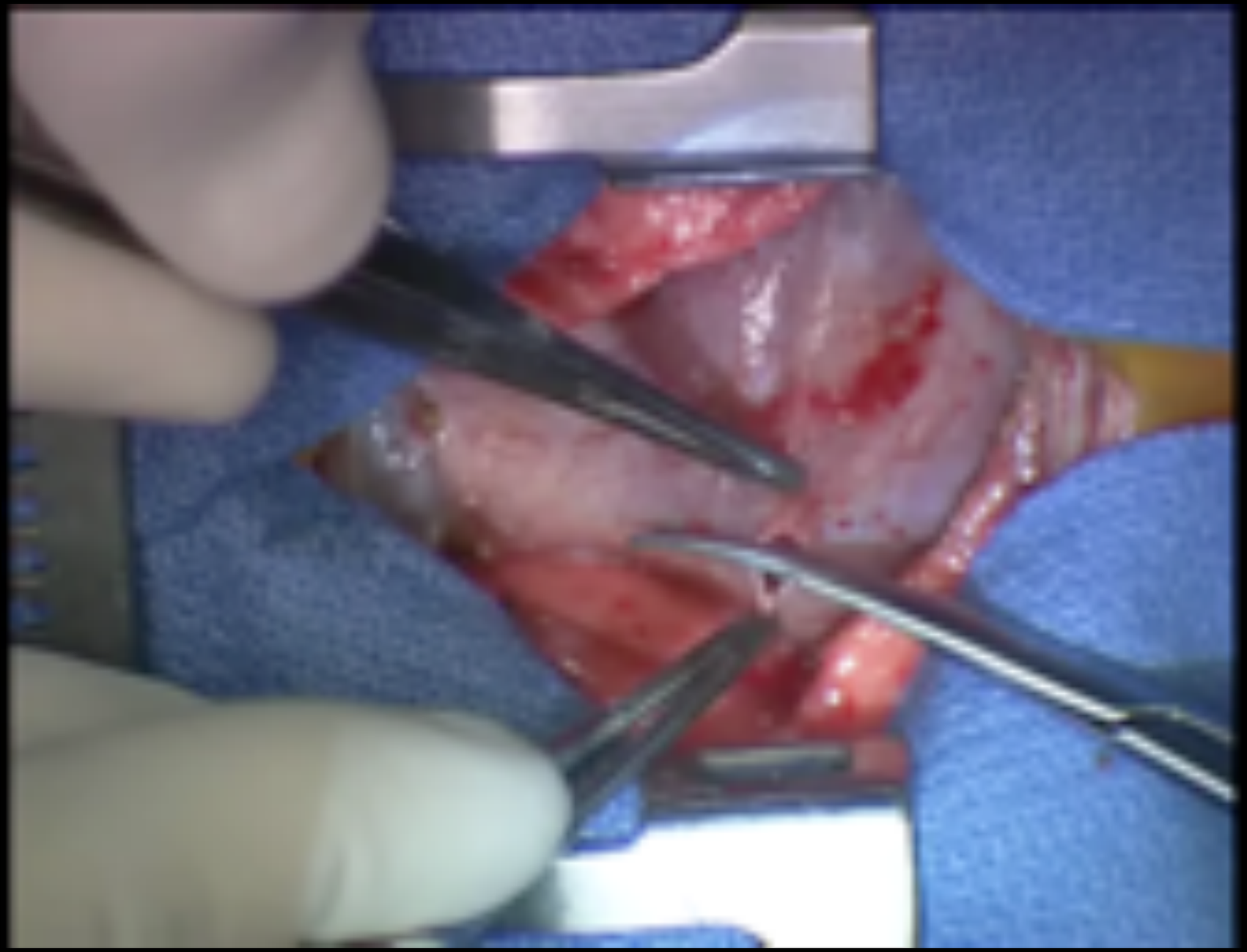
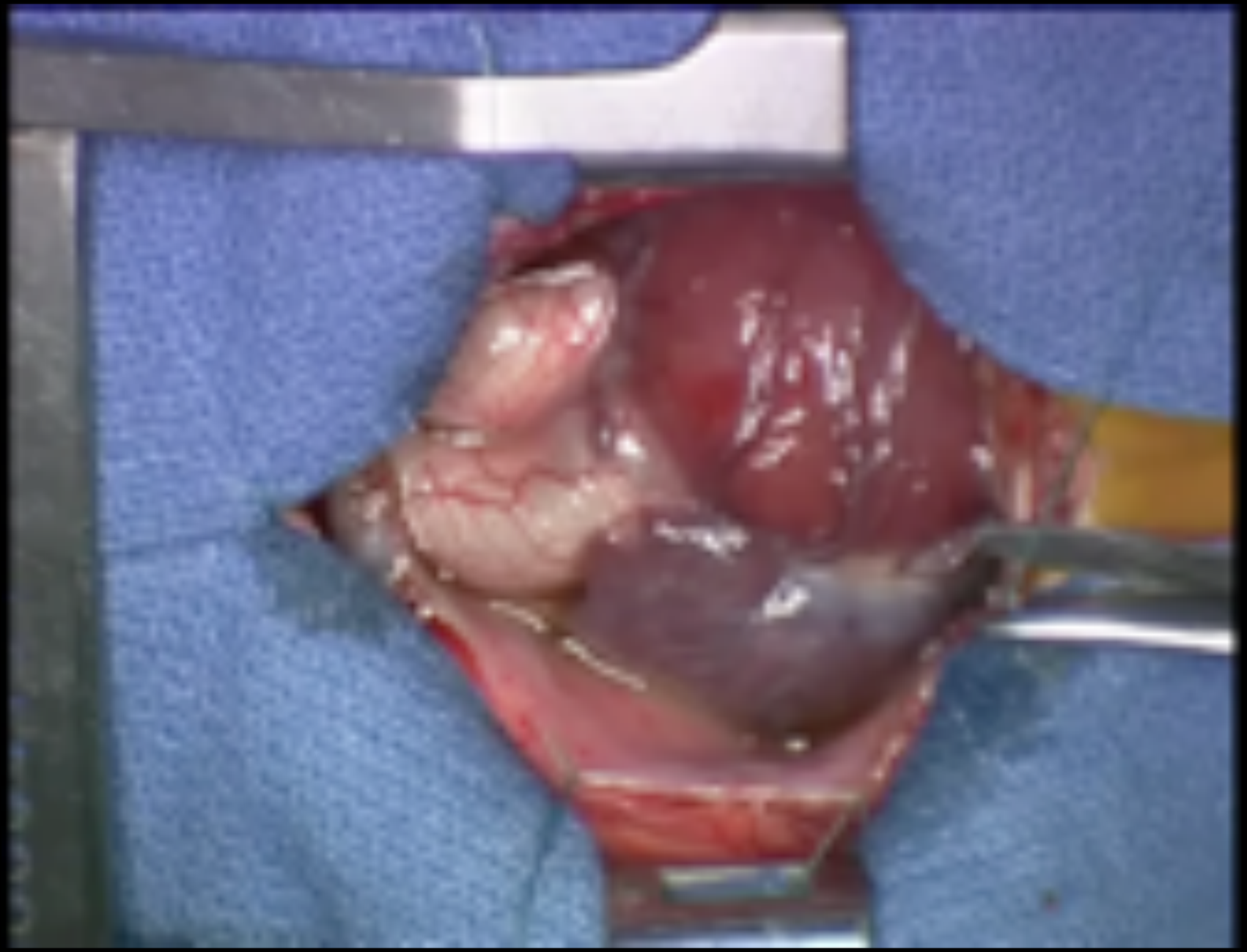
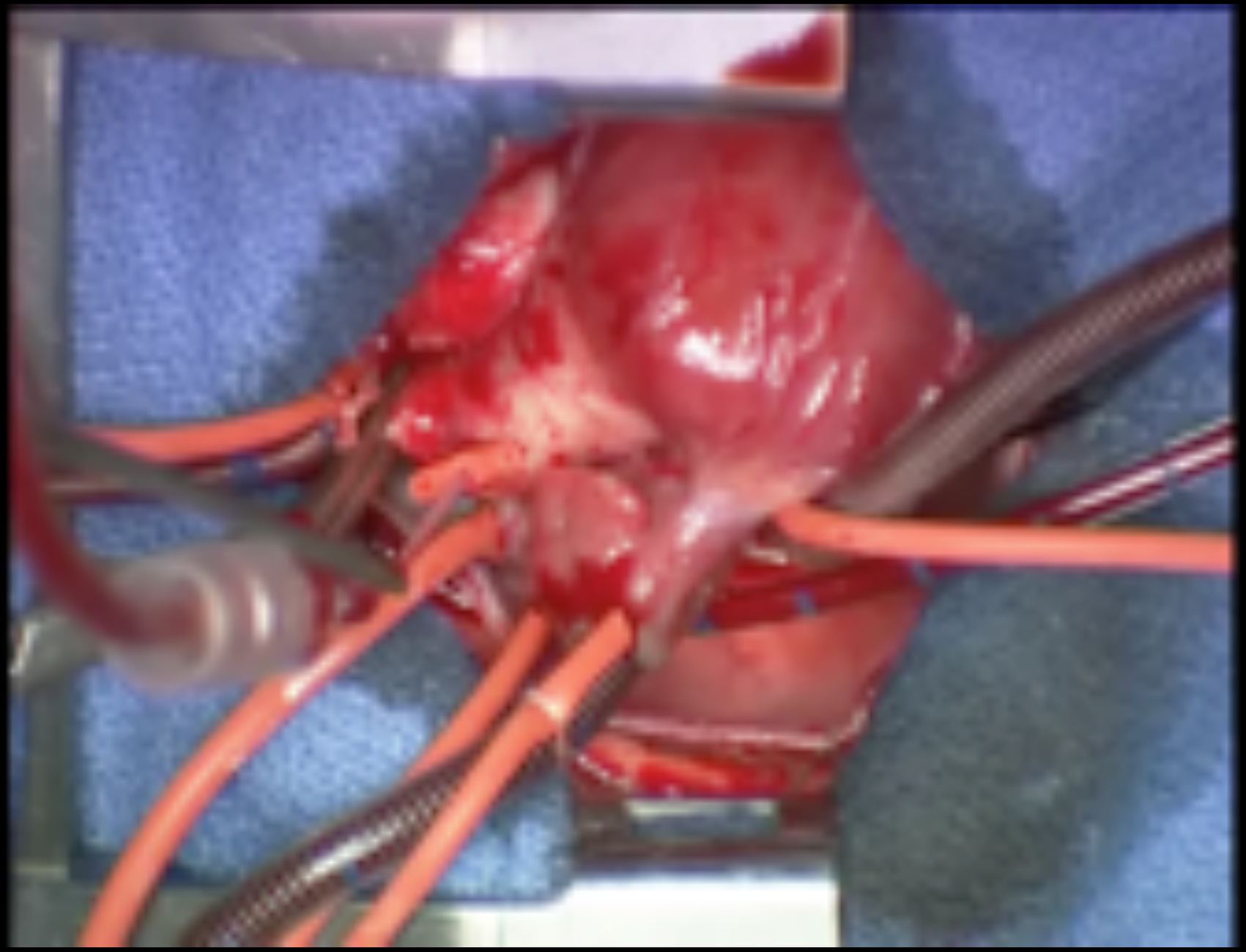


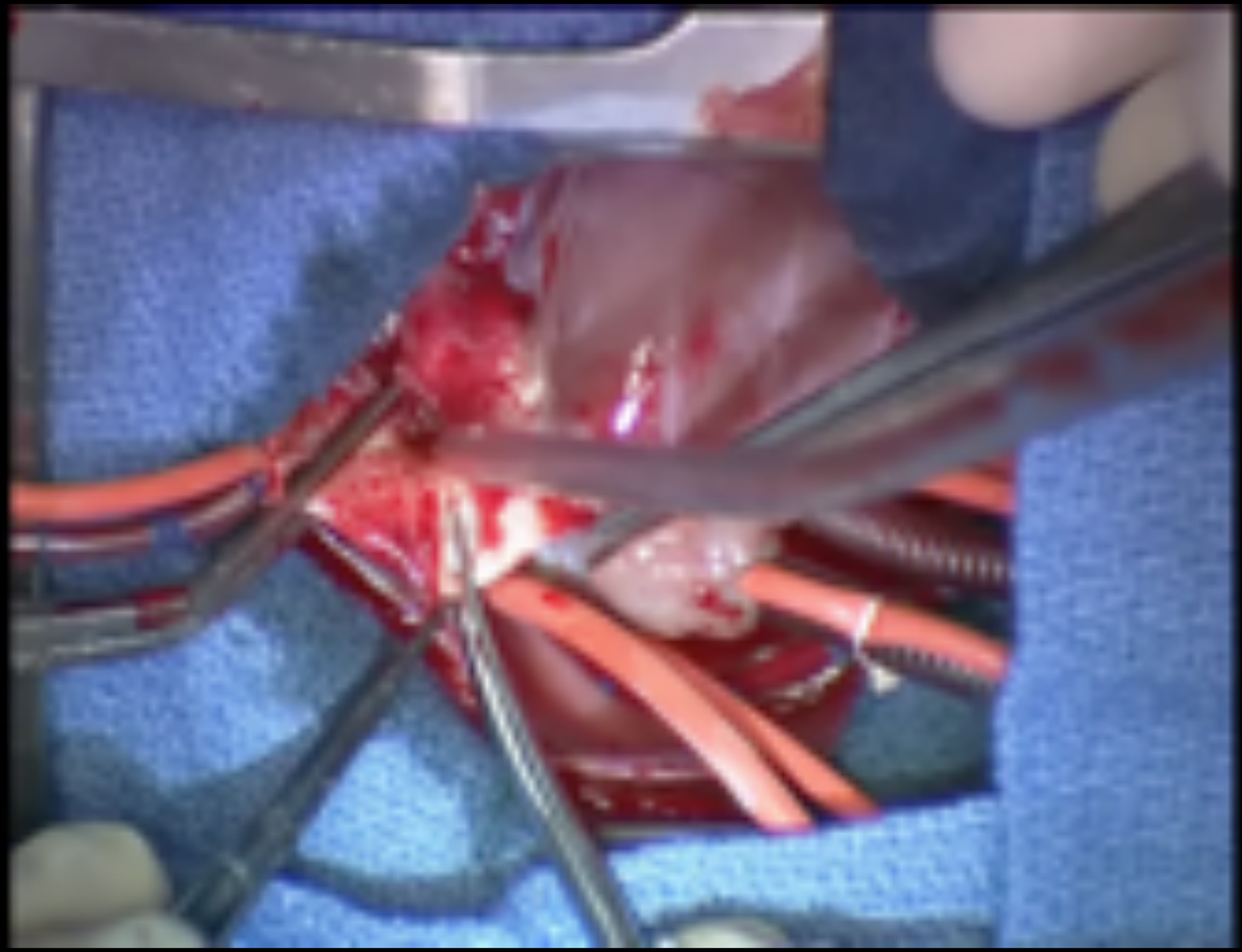
Yves Lecompte

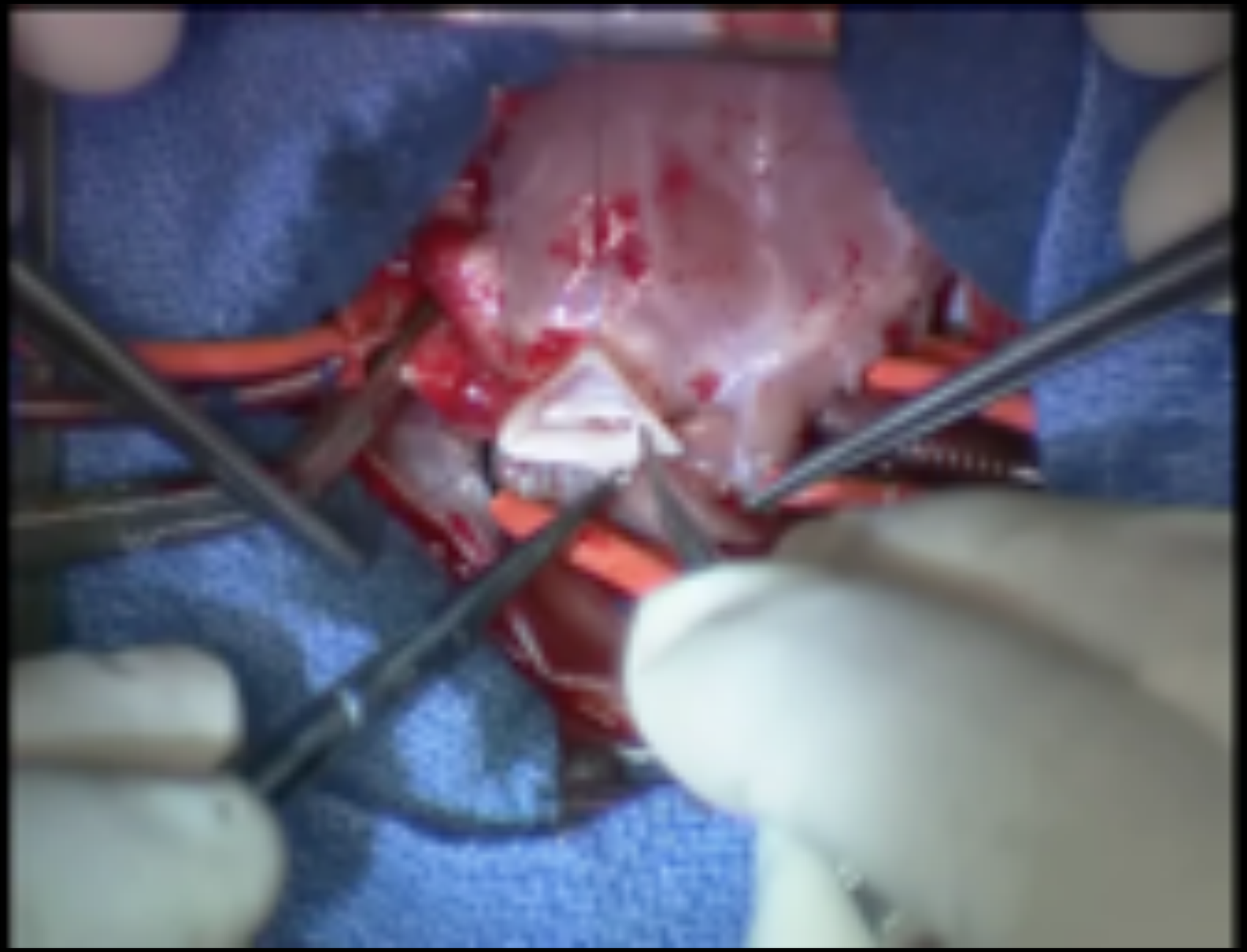


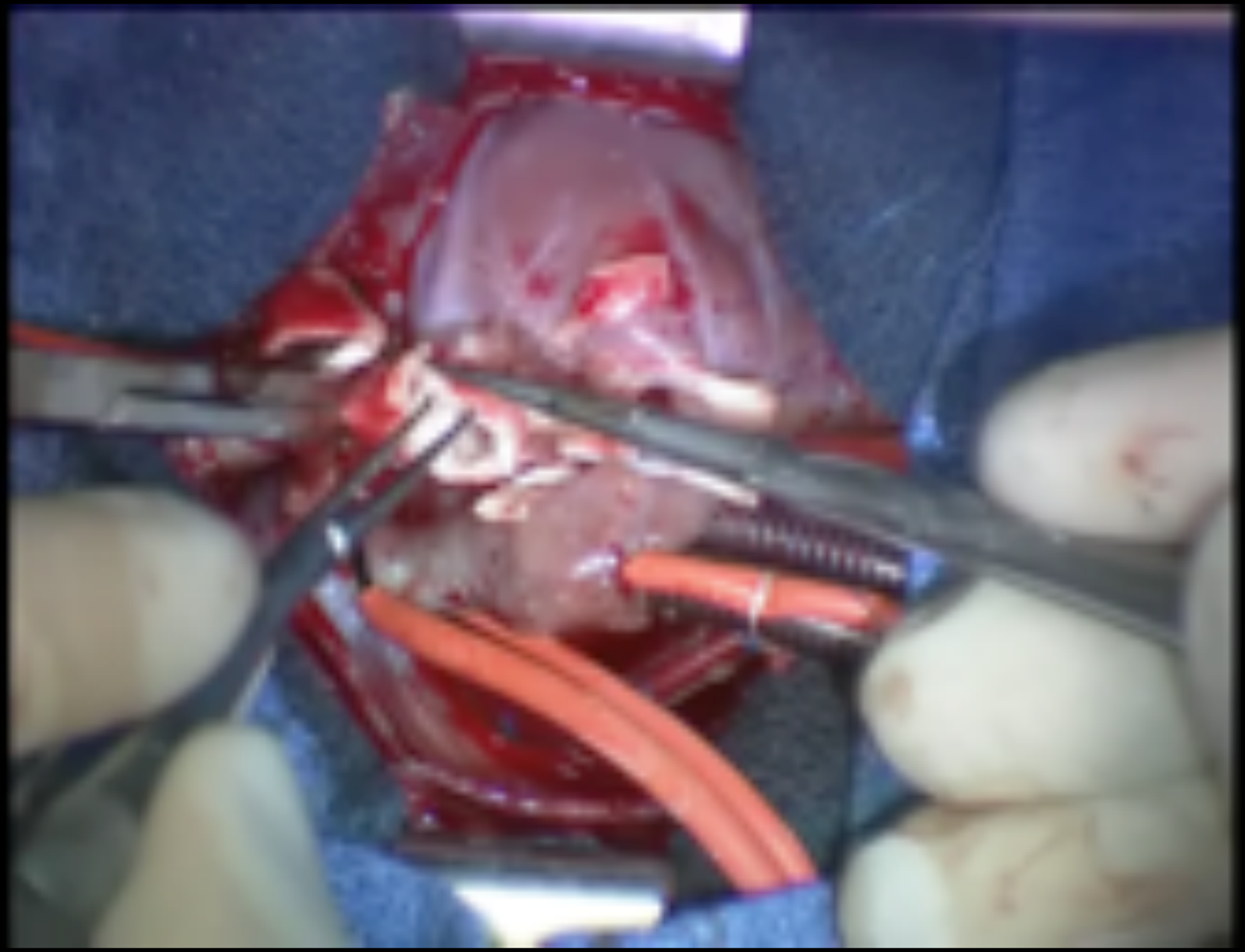


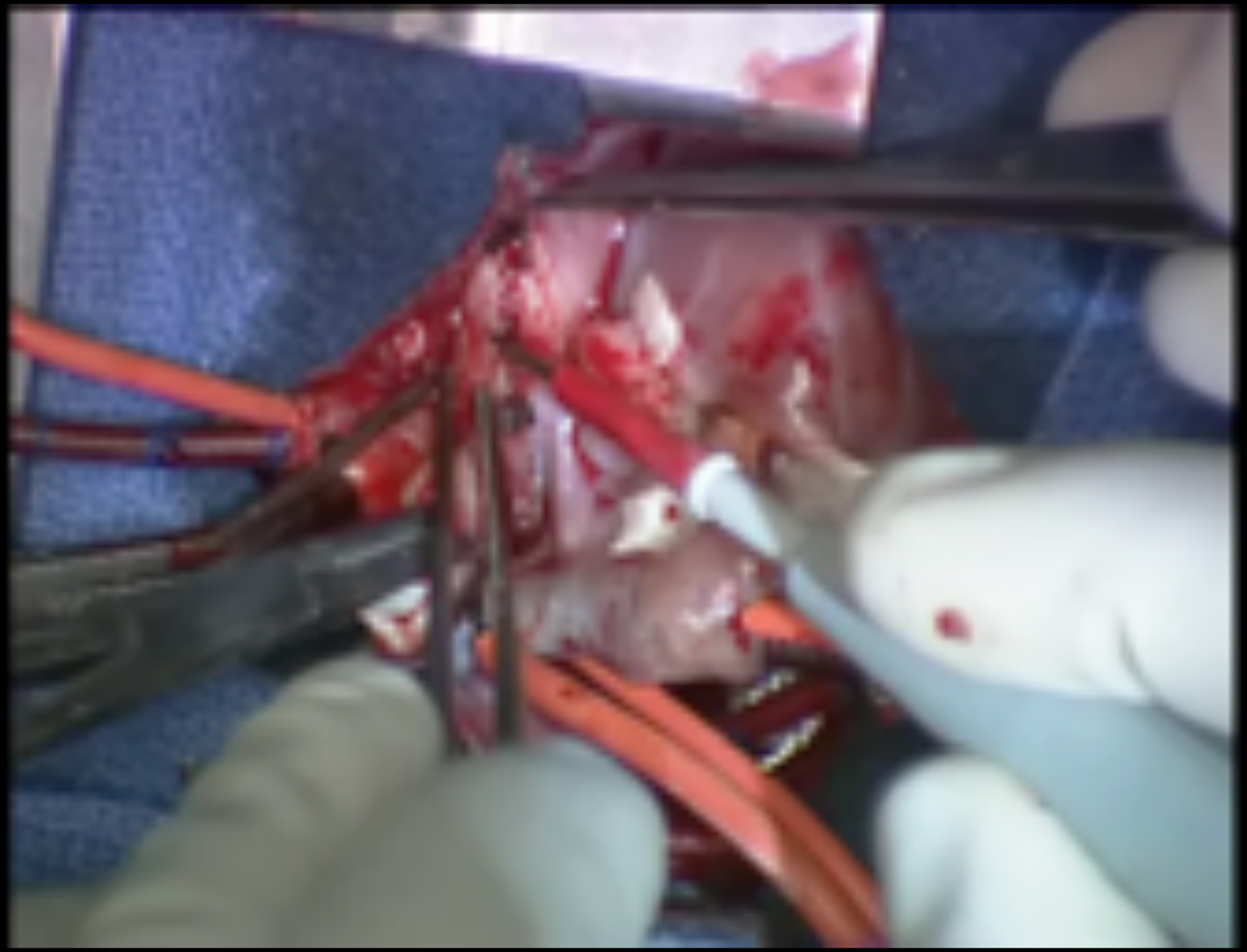


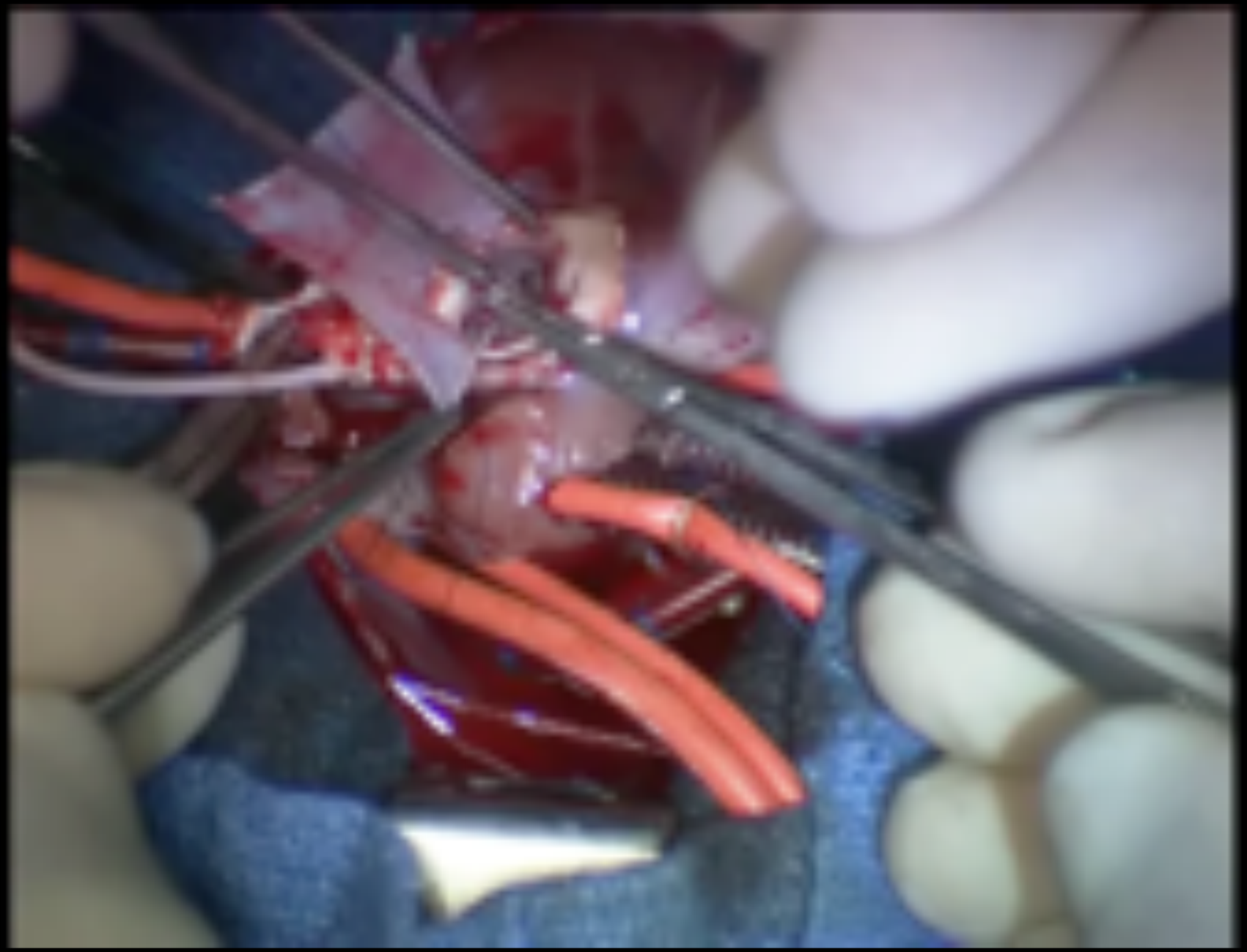


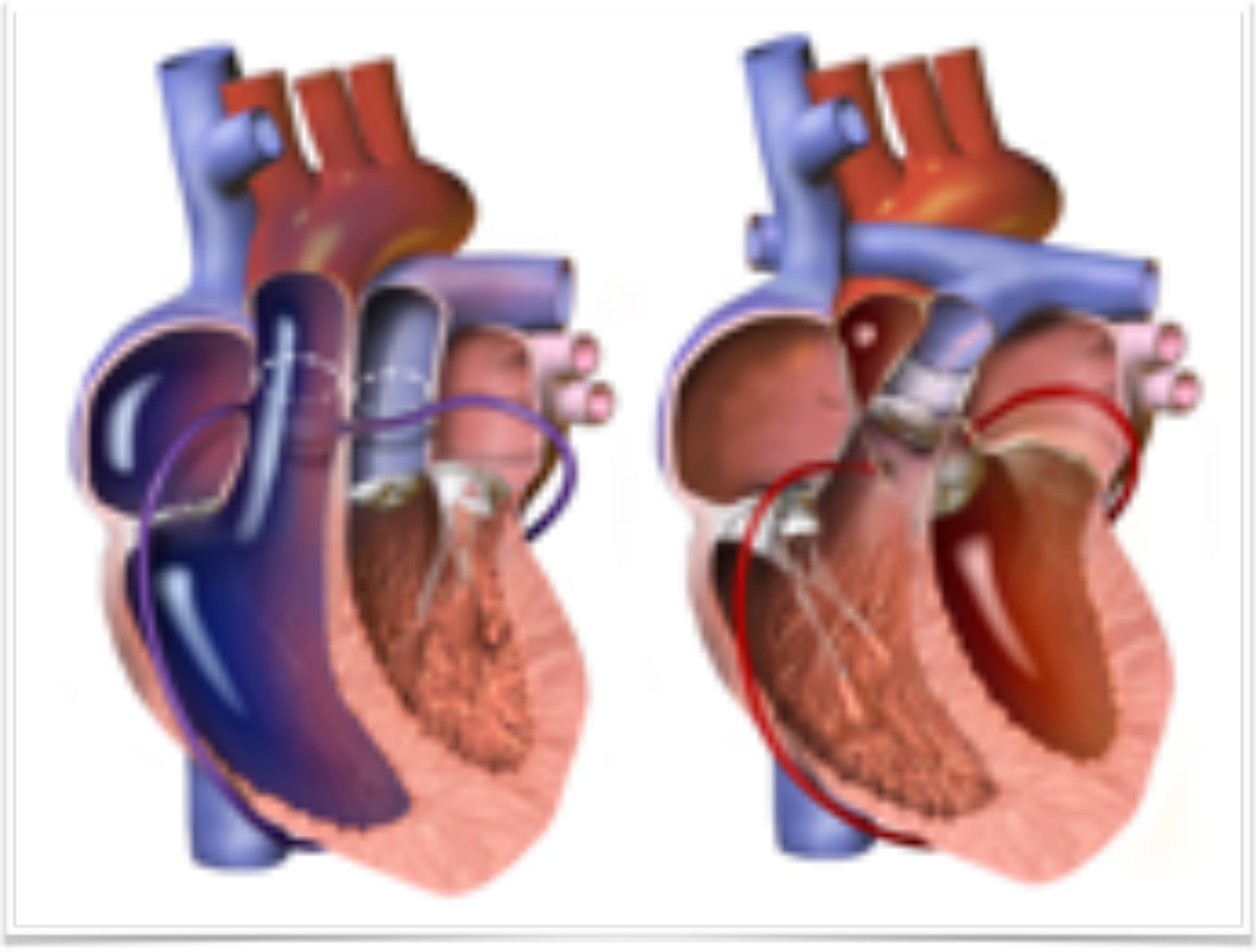




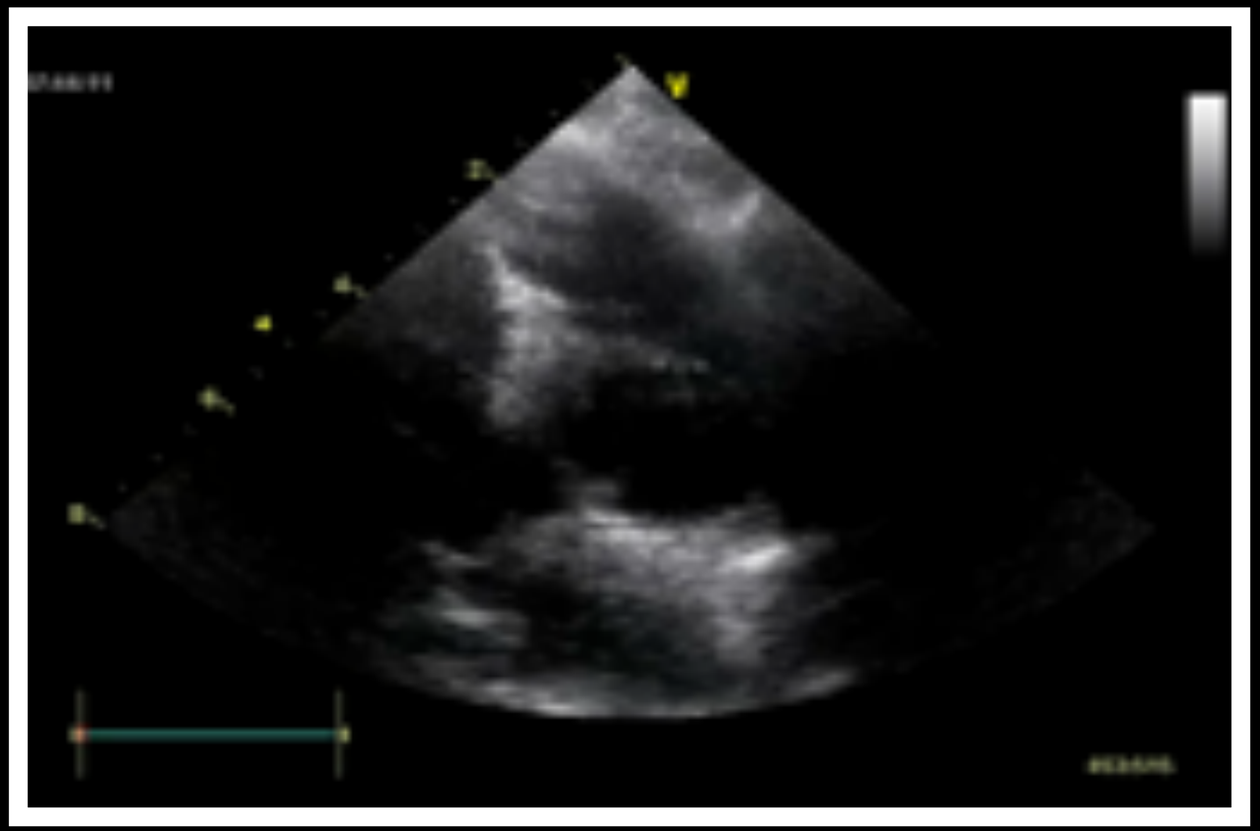


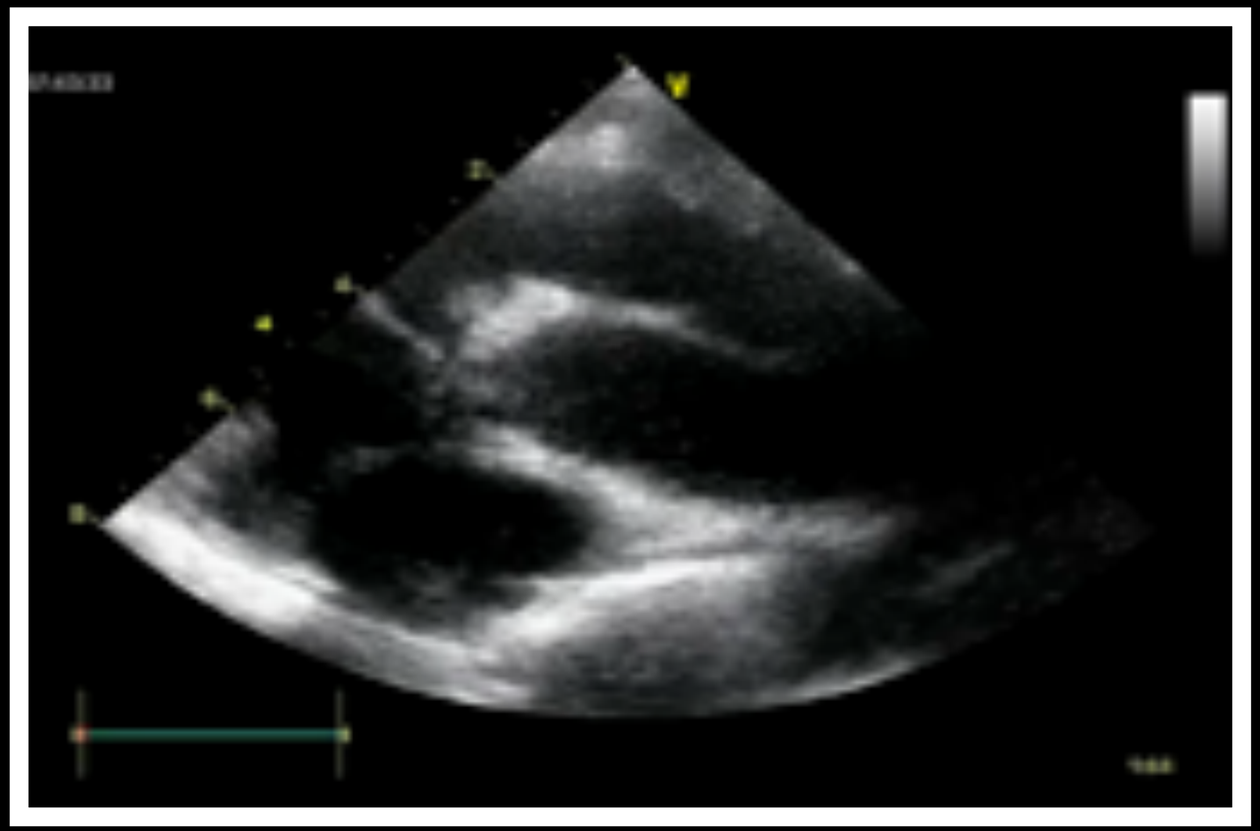






Outflow tracts after the arterial switch for TGA





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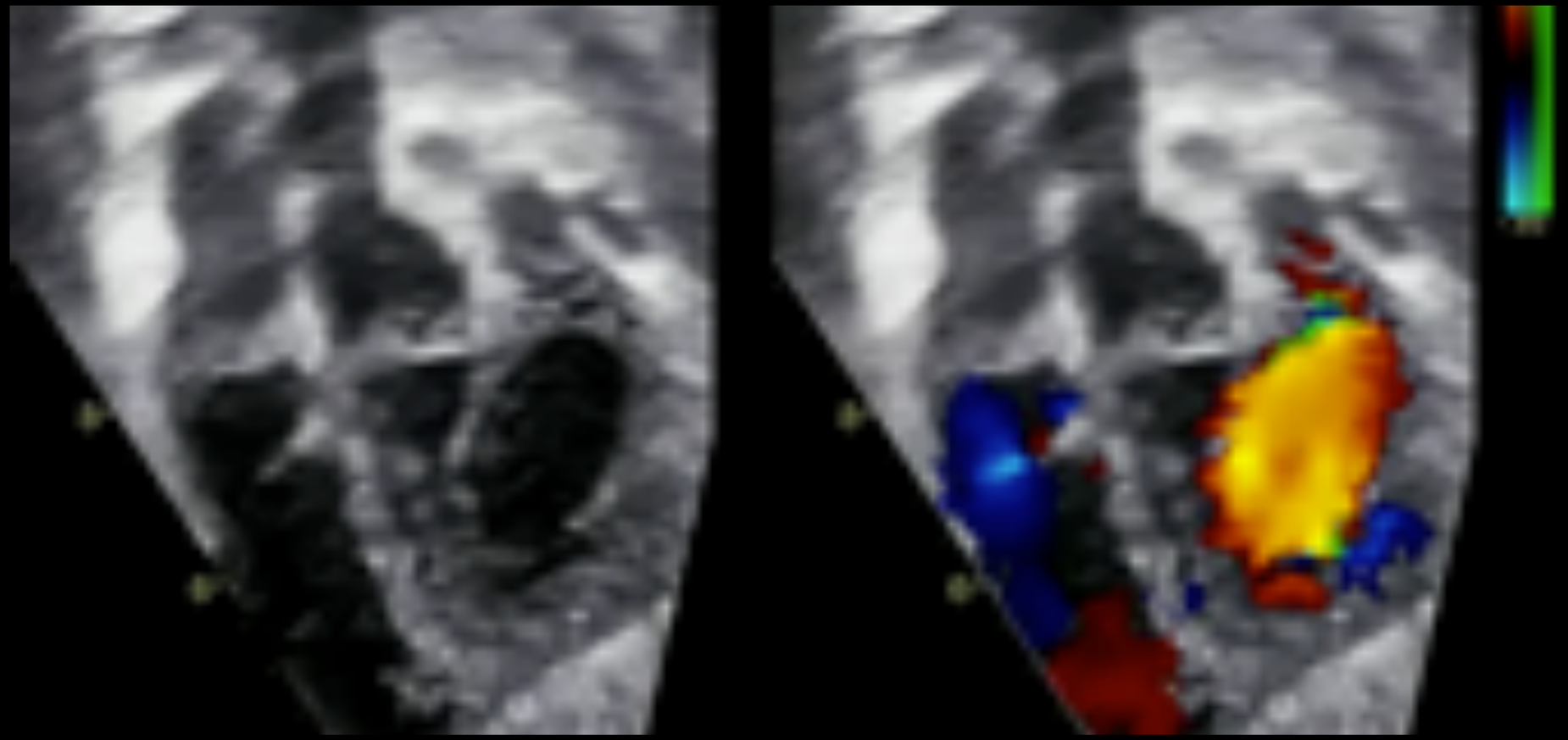
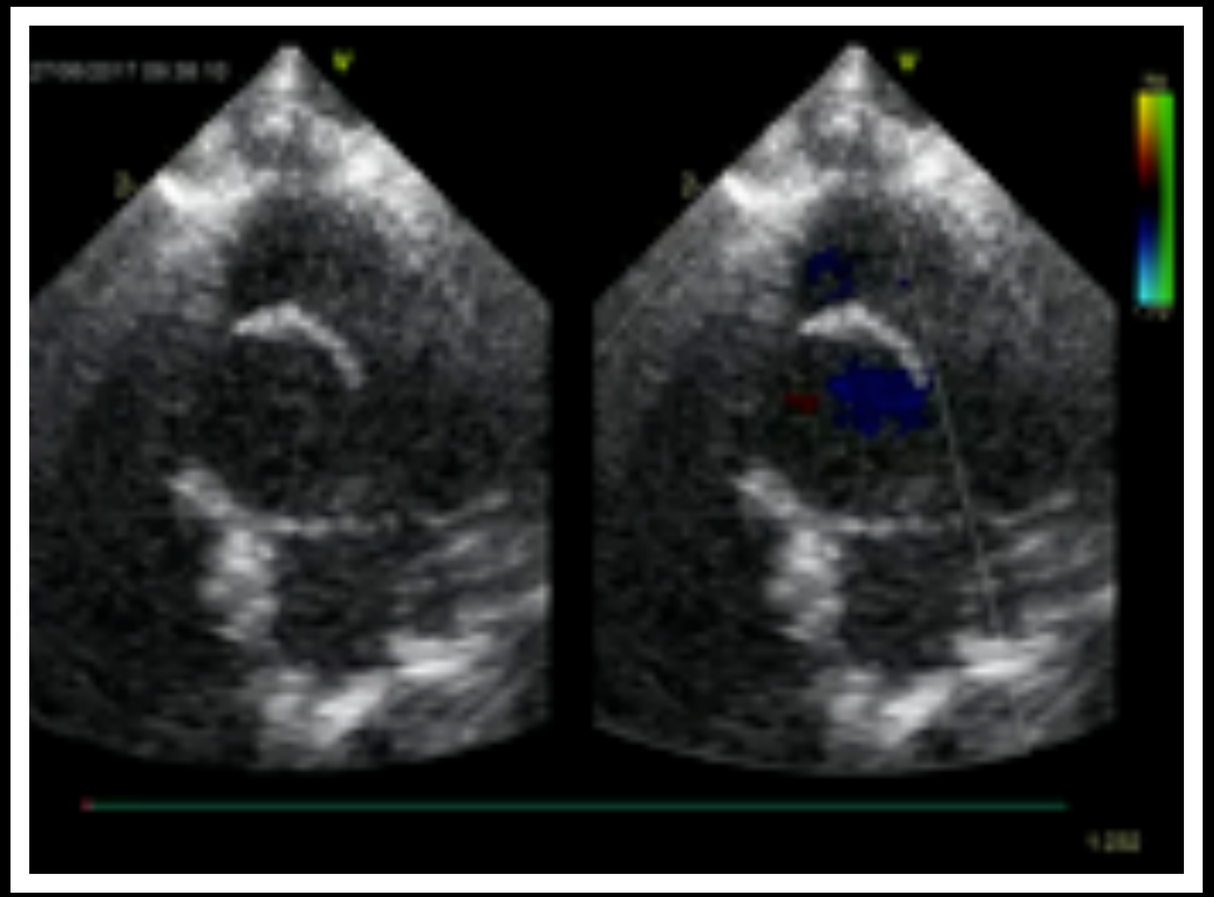
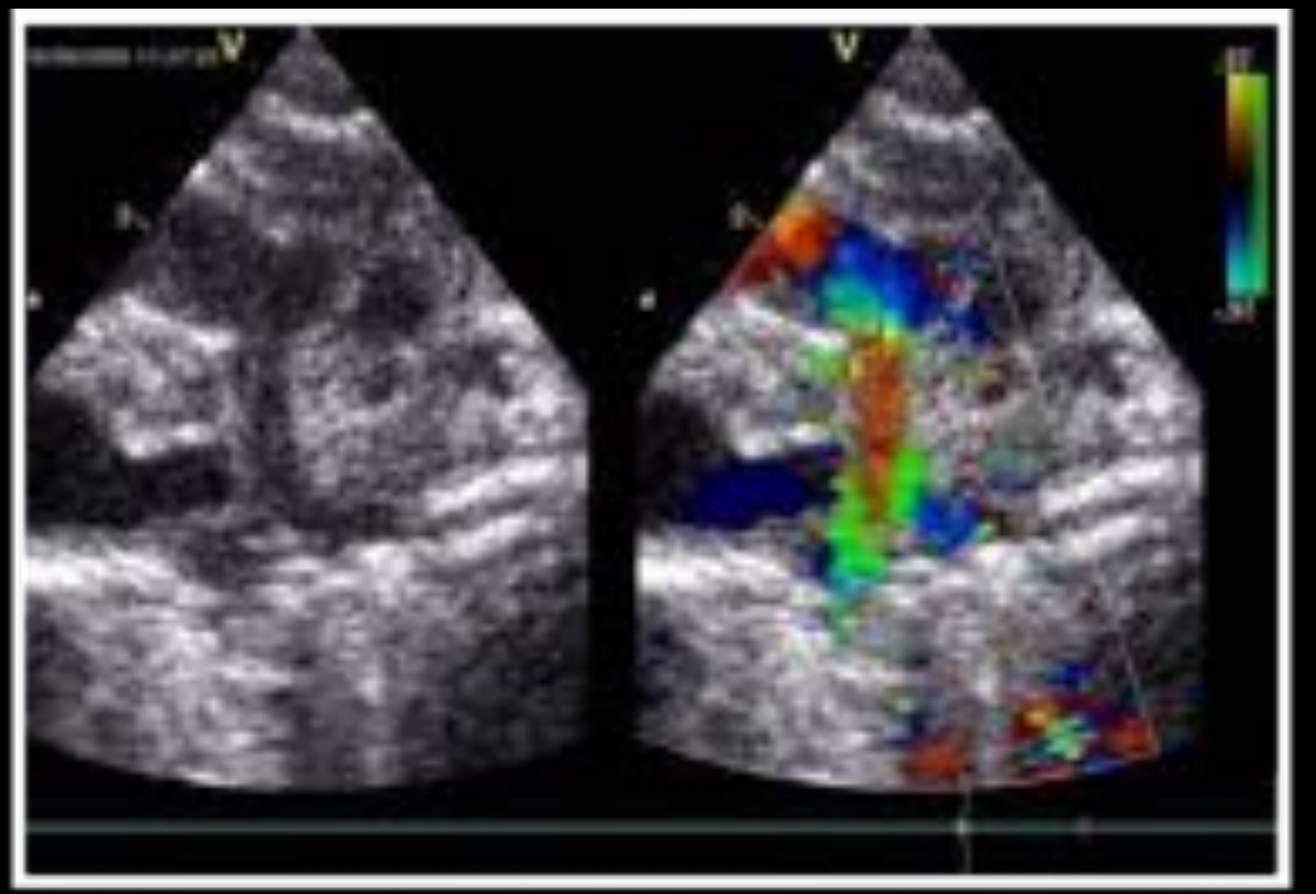


Outflow tracts after the arterial switch for TGA
The Lecompte manoeuver





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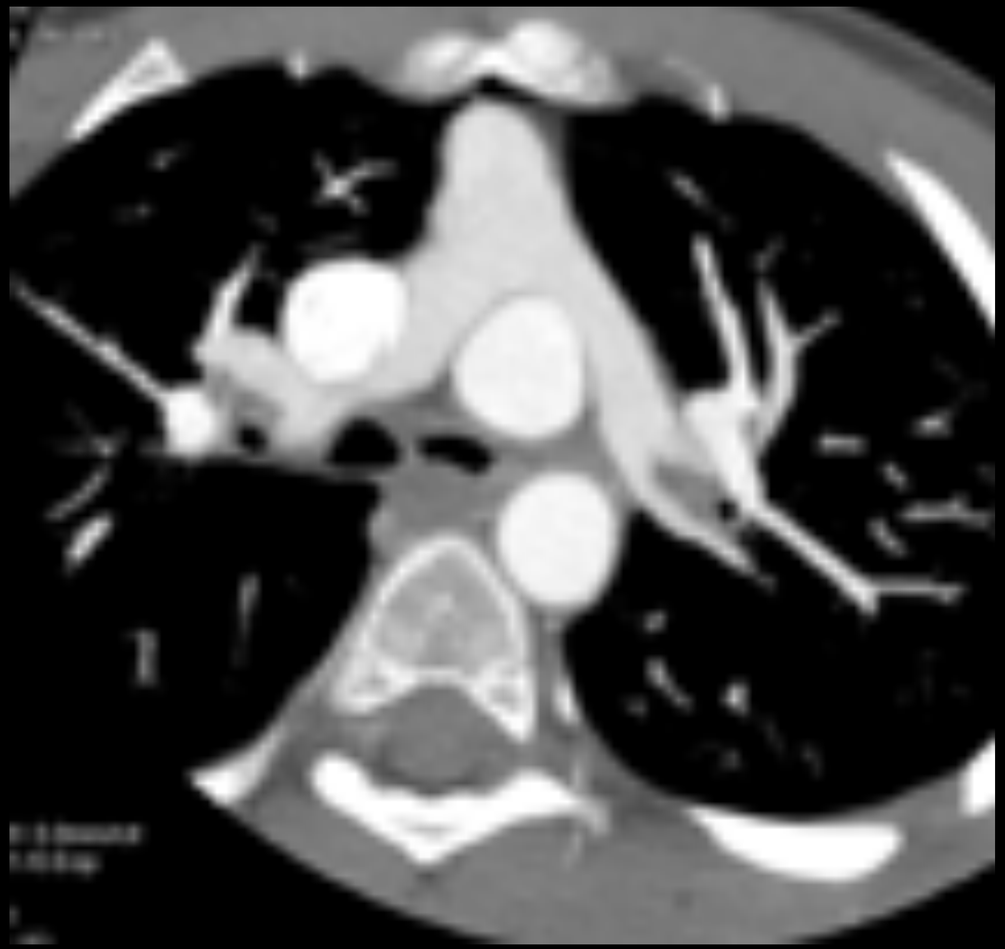
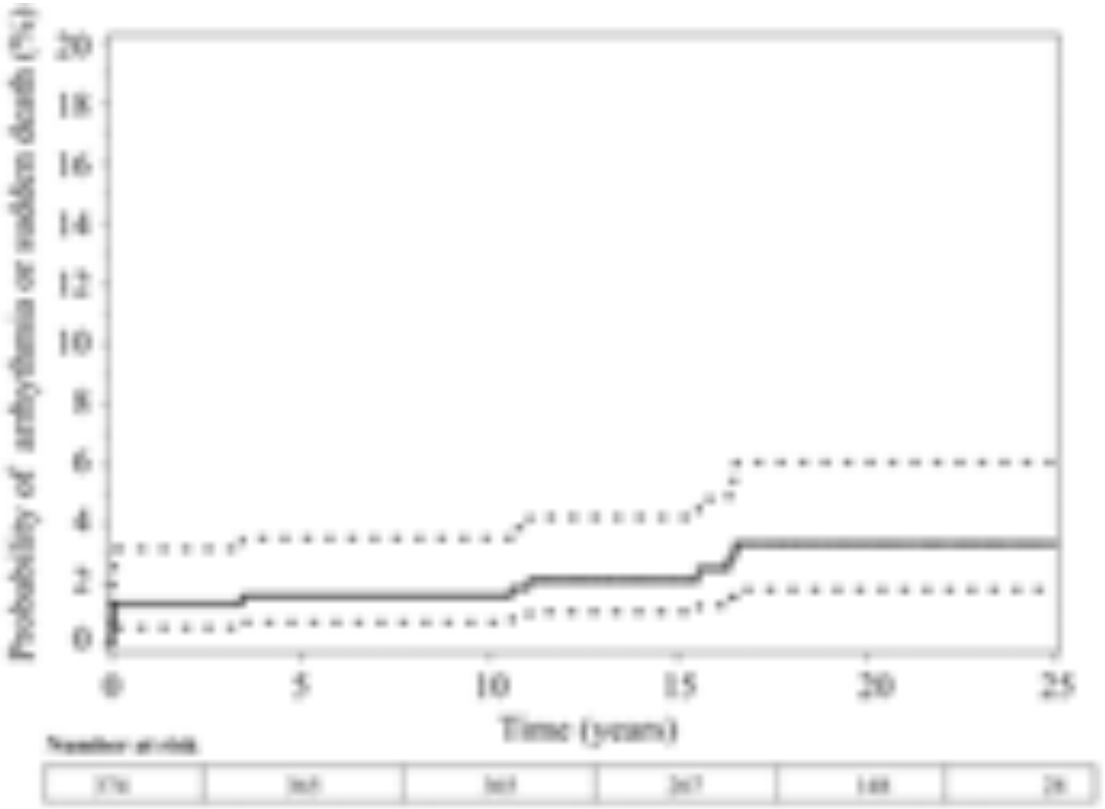
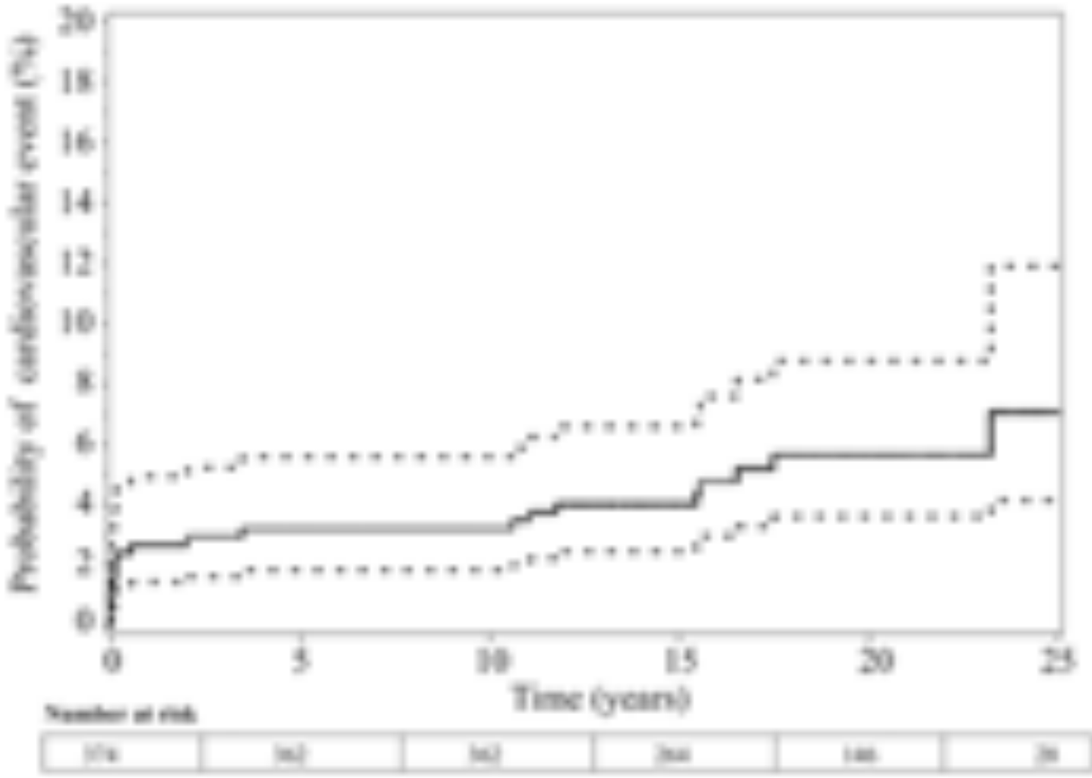


Long term outcomes after the arterial switch operation

Cardiovascular events in the long term



Cumulative probability of arrhythmia or sudden death



Cumulative probability of the combined cardiovascular outcome

Khairy P et al. *Circulation*. 2013;127:331–339.

Life-long management of the ASO population poses several challenges

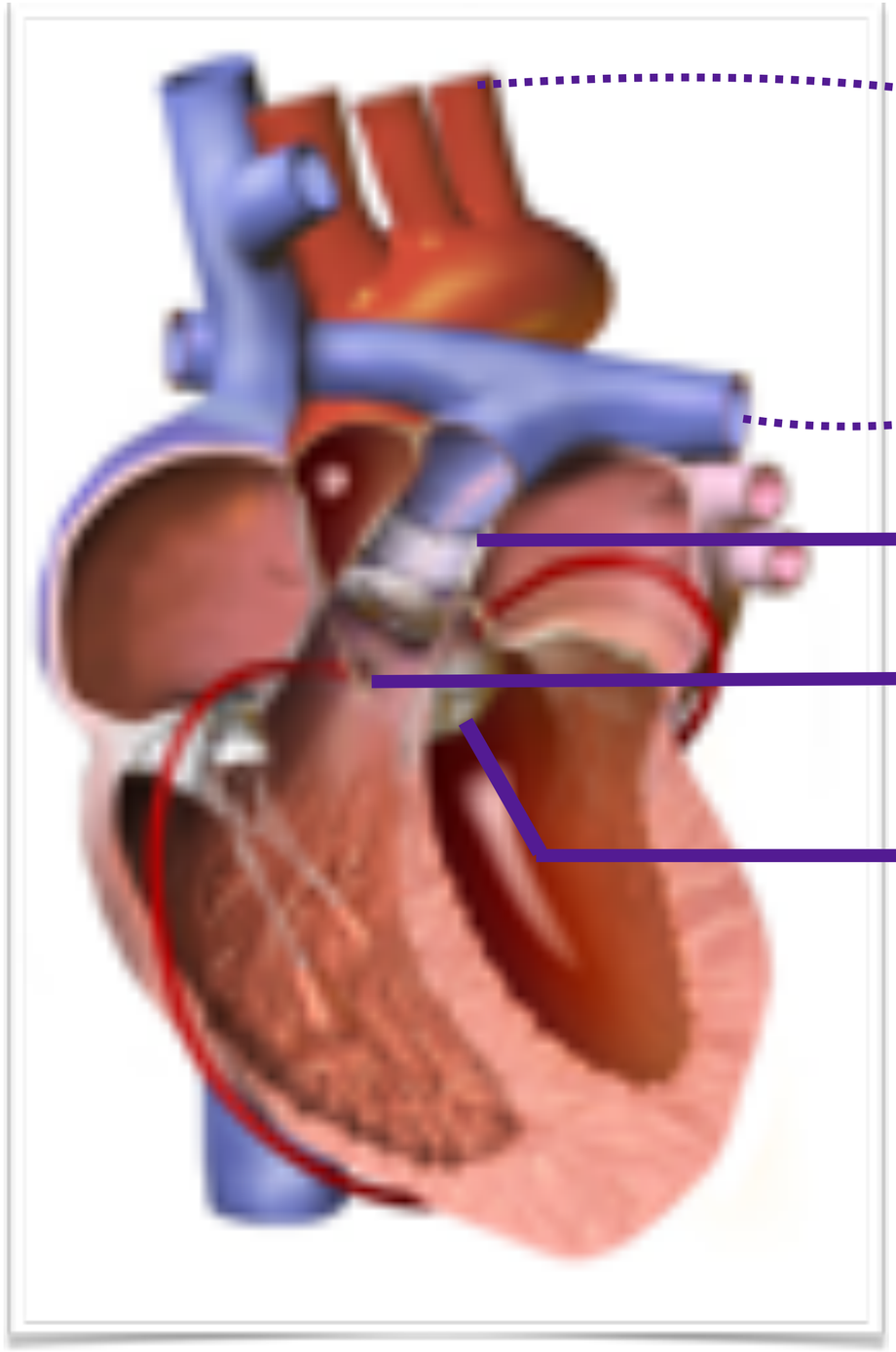
1) absence of current consensus regarding the appropriate interval and modality for surveillance imaging;

2) there is no defined management strategy when subclinical anatomic or physiologic abnormalities are identified;

3) symptoms attributable to potential complications, especially coronary obstruction, are rare and therefore practitioners must be vigilant for classic and atypical presentations;

4) the effects of acquired coronary artery disease superimposed on manipulated coronary arteries remain unknown.

Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511



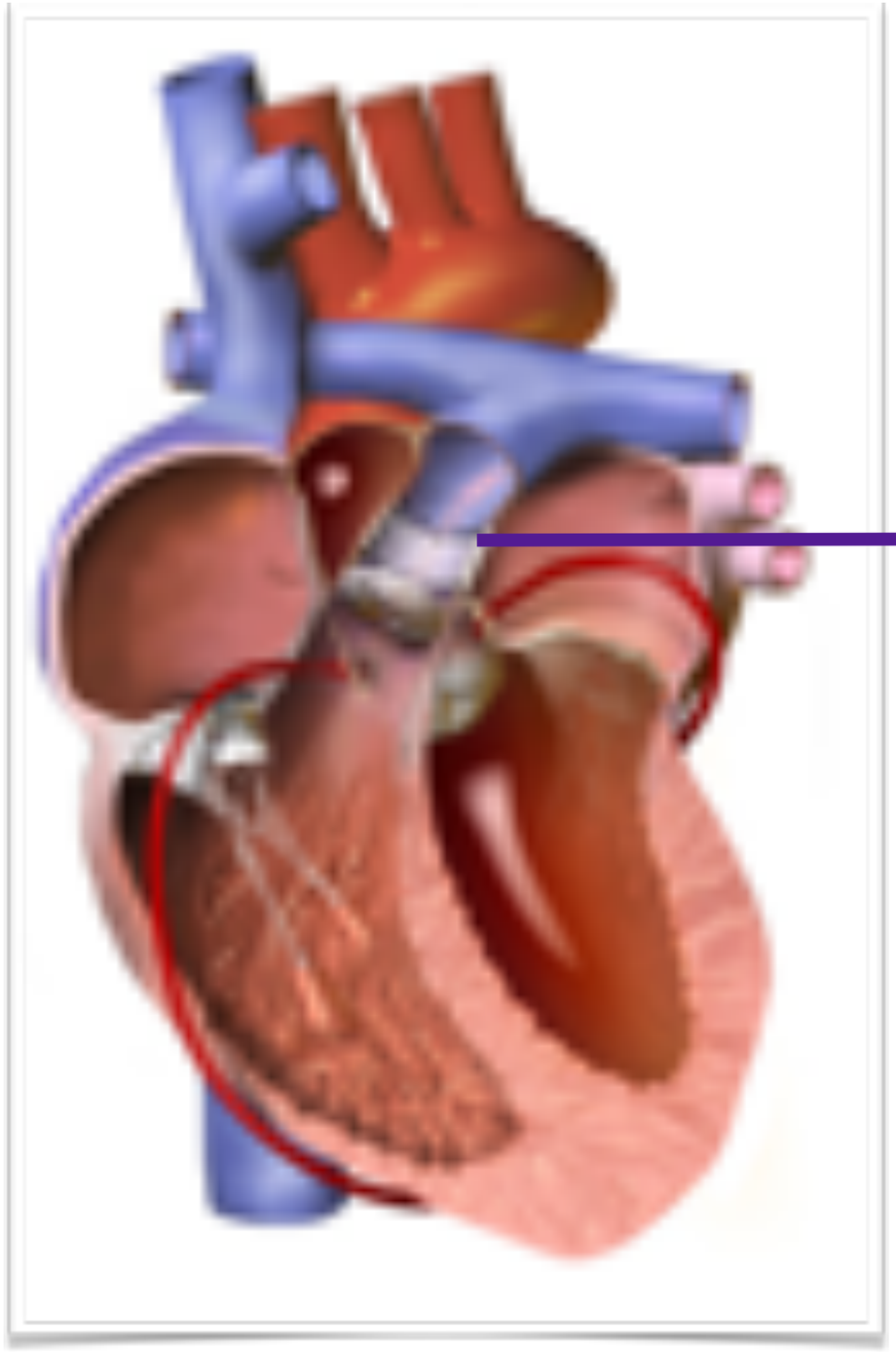
Neurodevelopmental outcomes

Pulmonary arterial hypertension

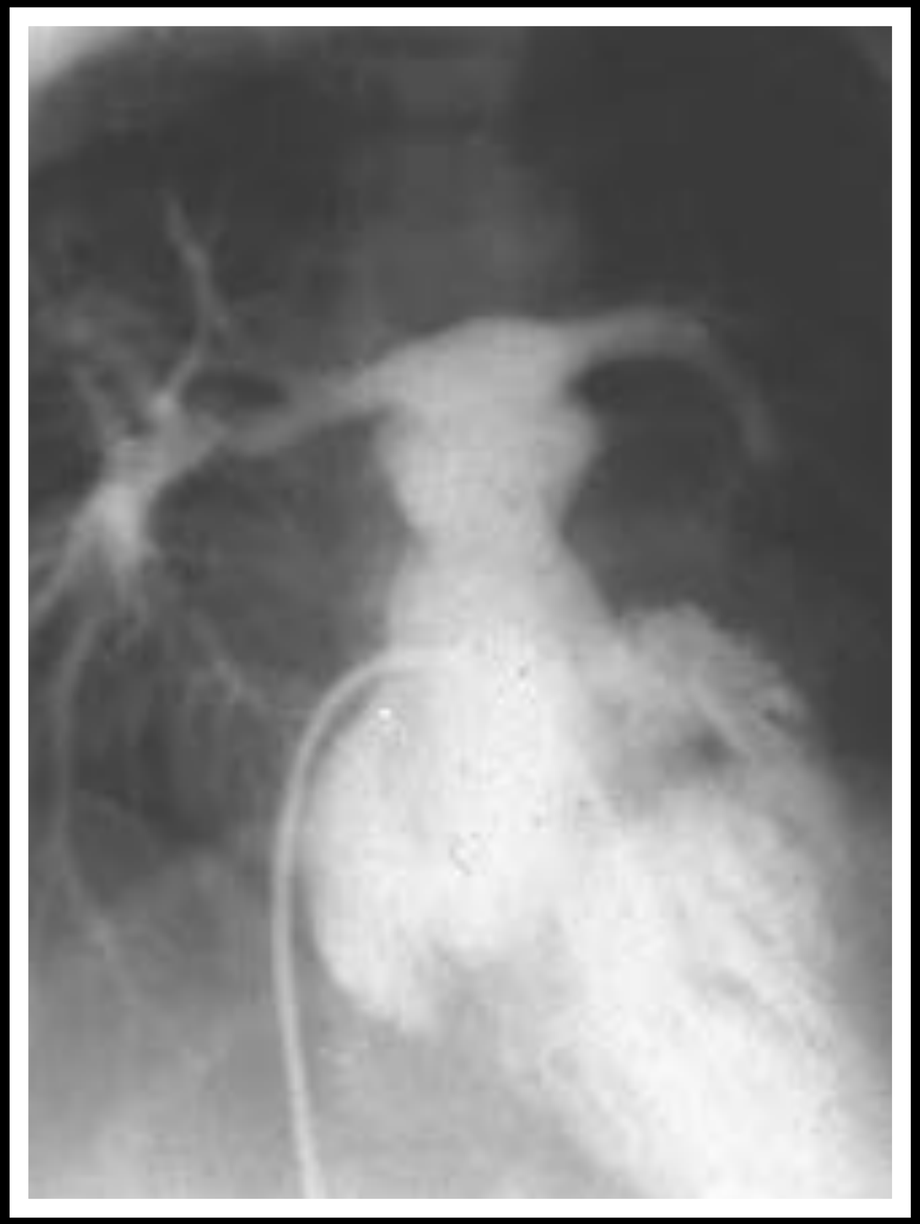
Pulmonary artery stenosis

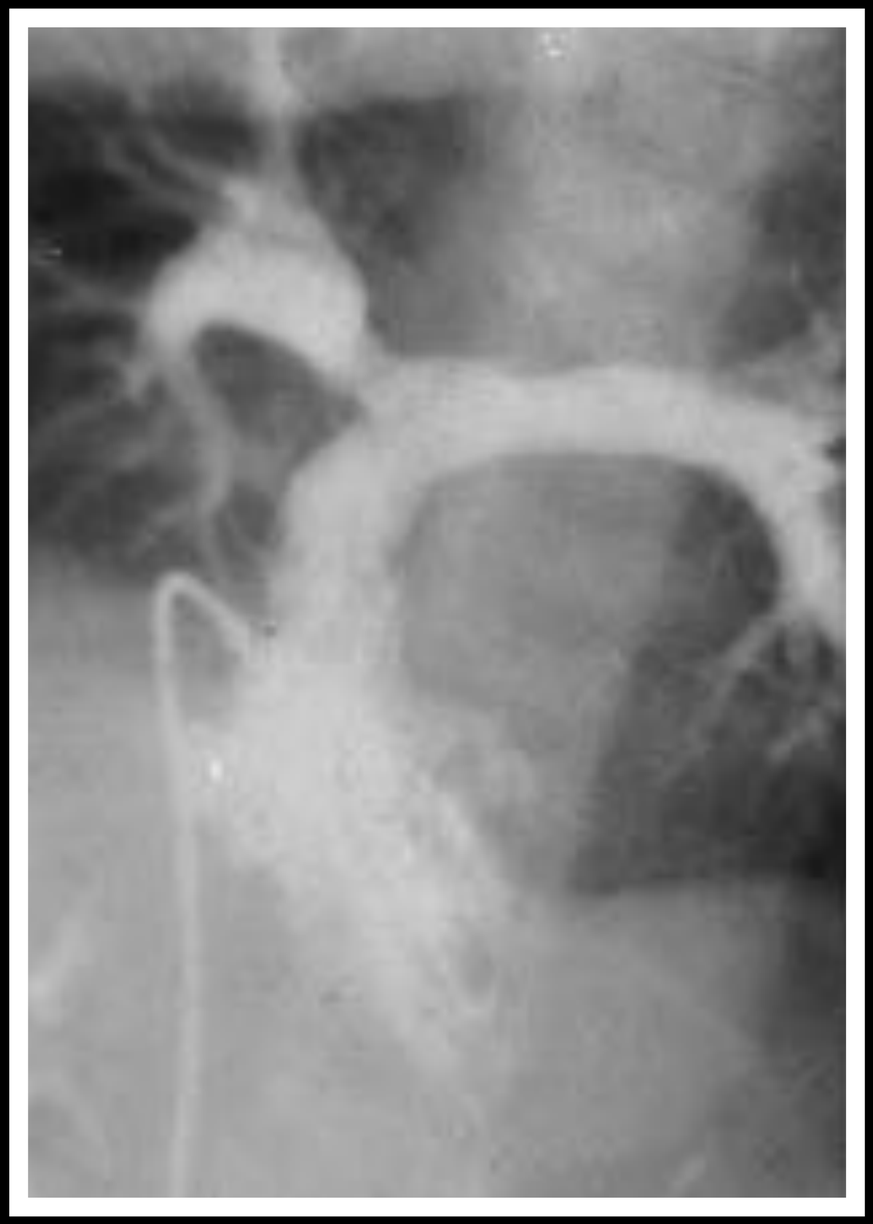
Coronary artery obstruction

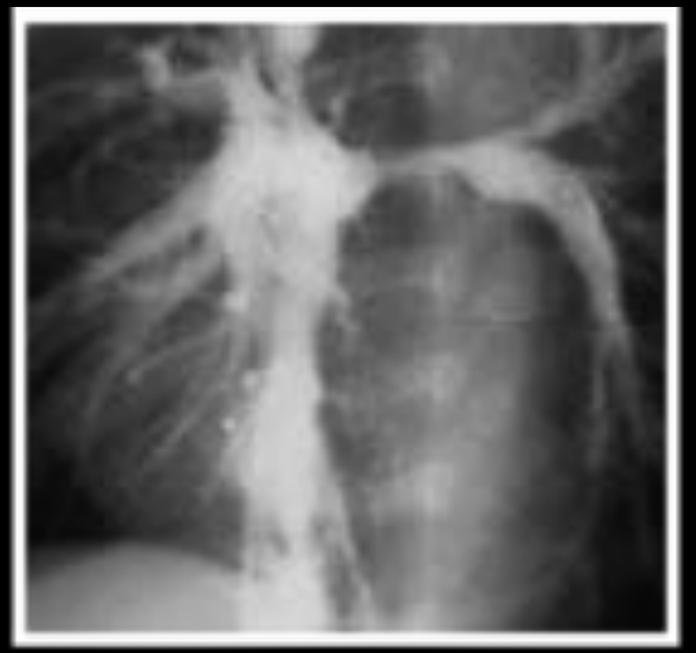
Aortic valve regurgitation

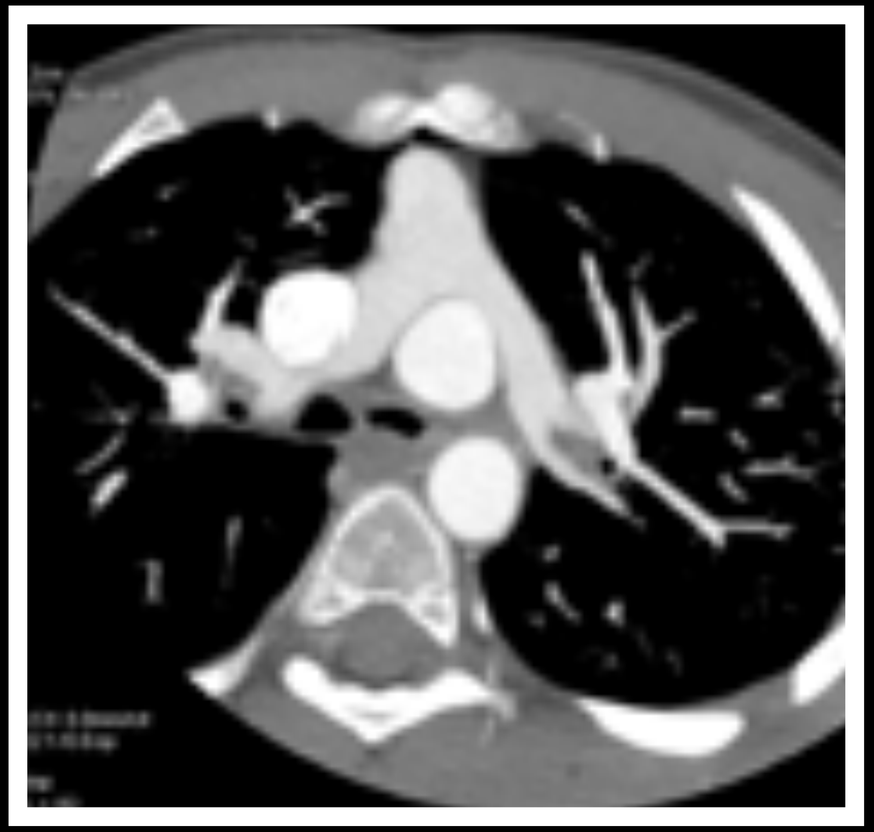


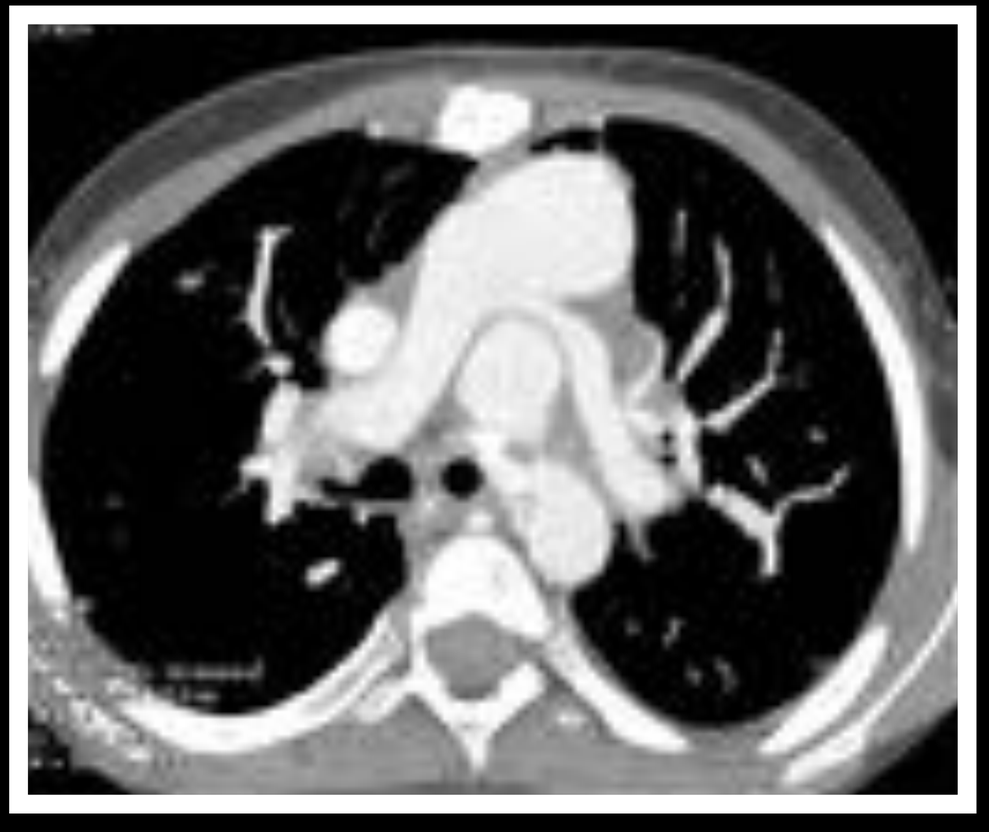
Pulmonary artery stenosis

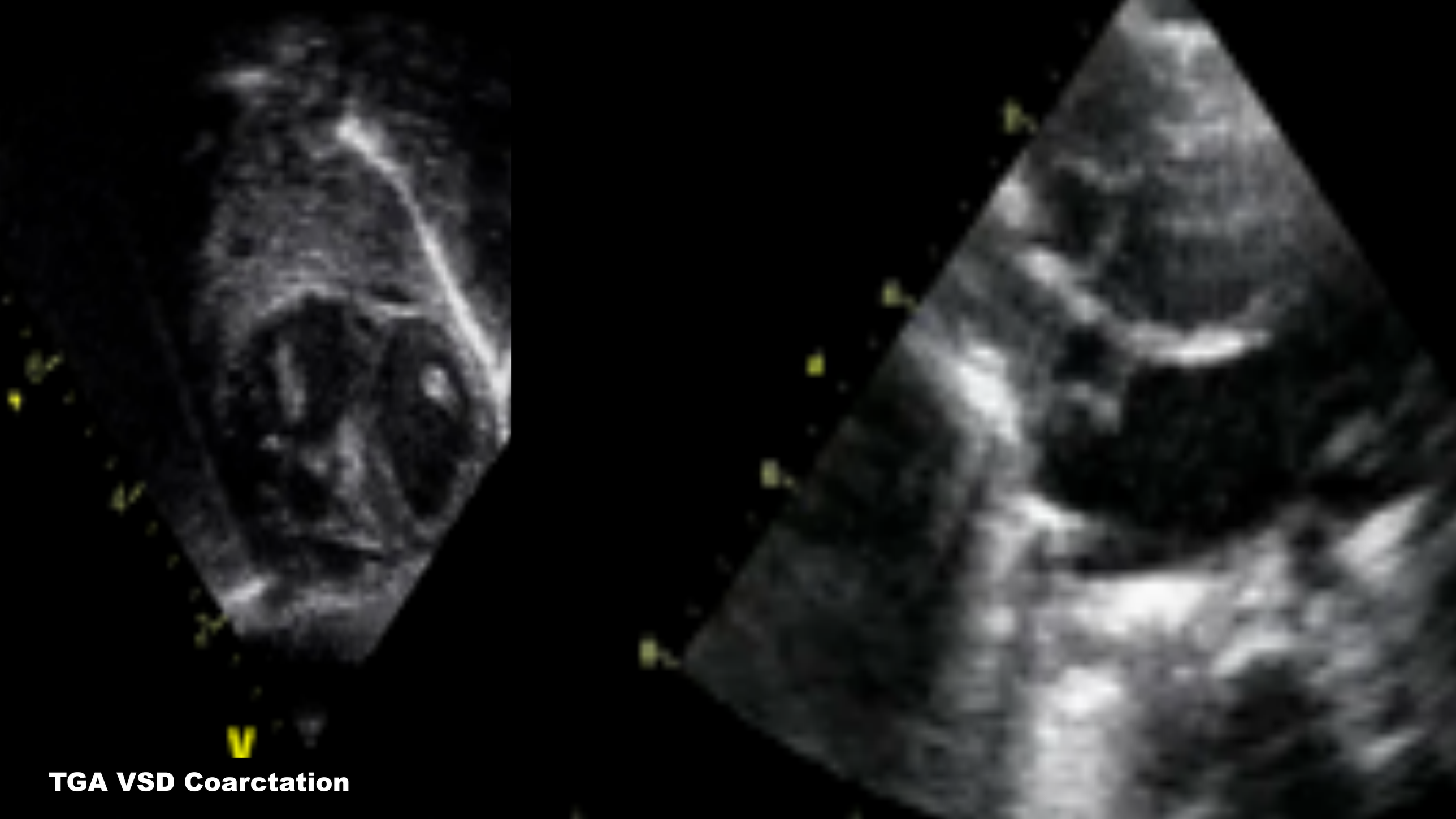


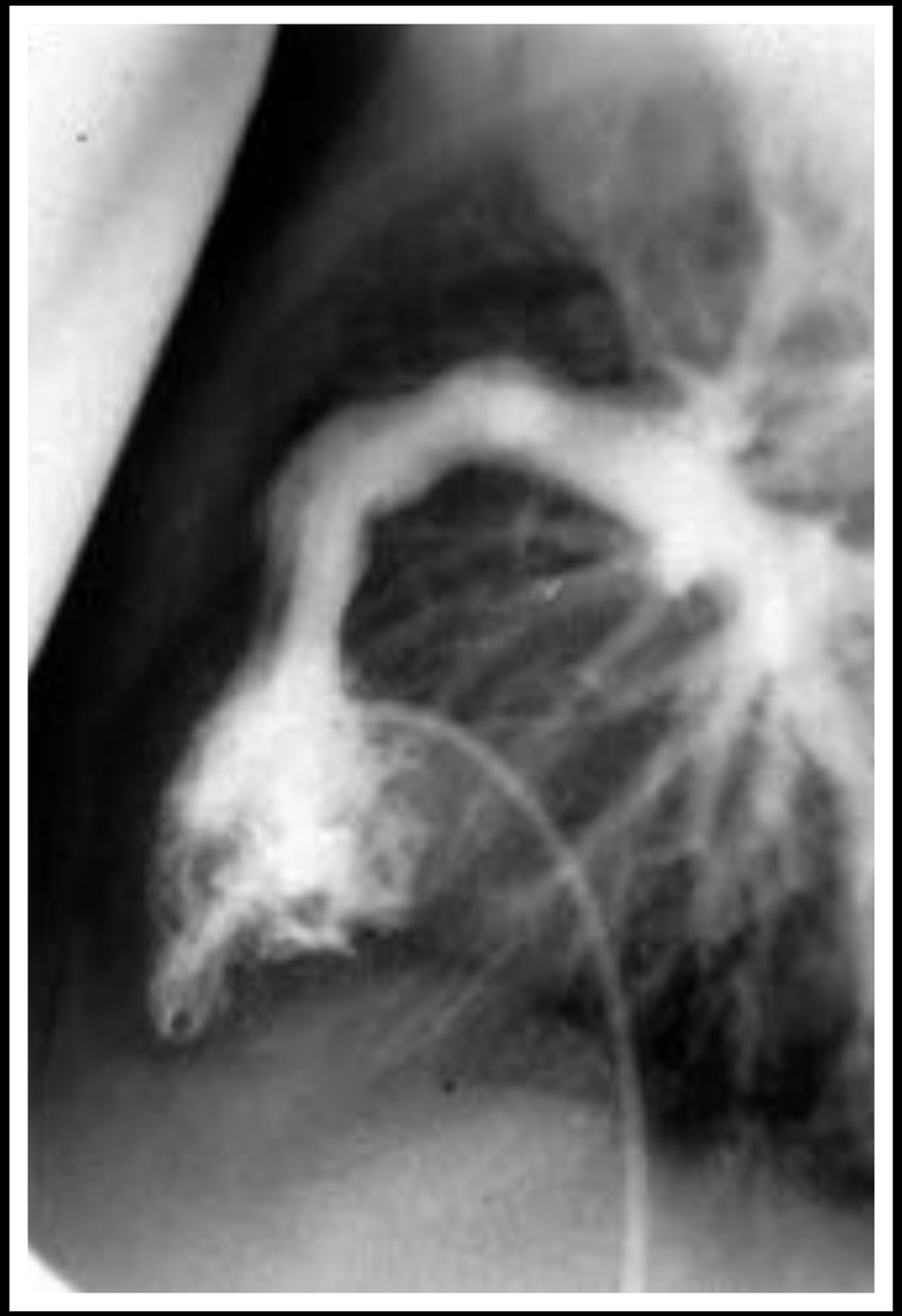


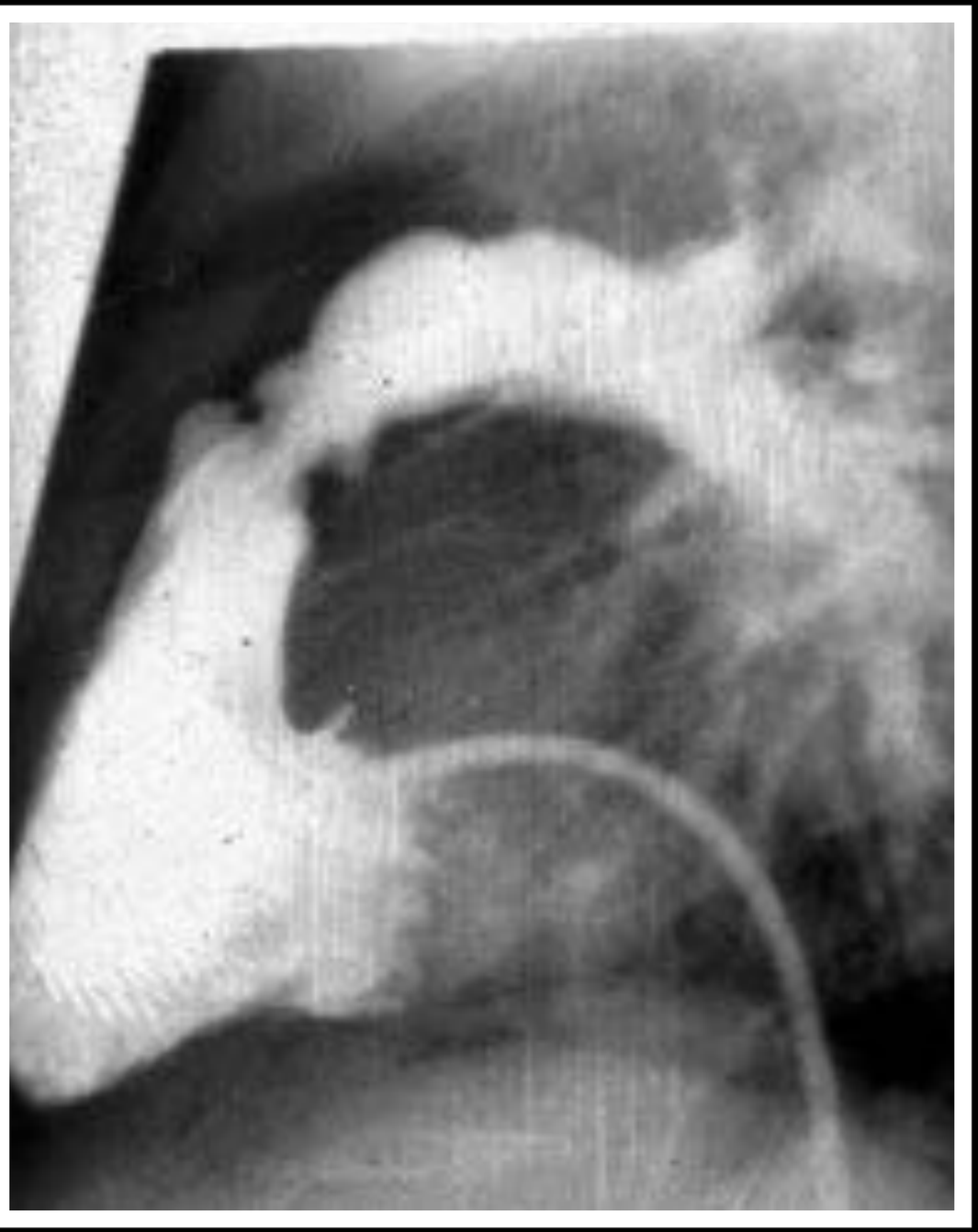


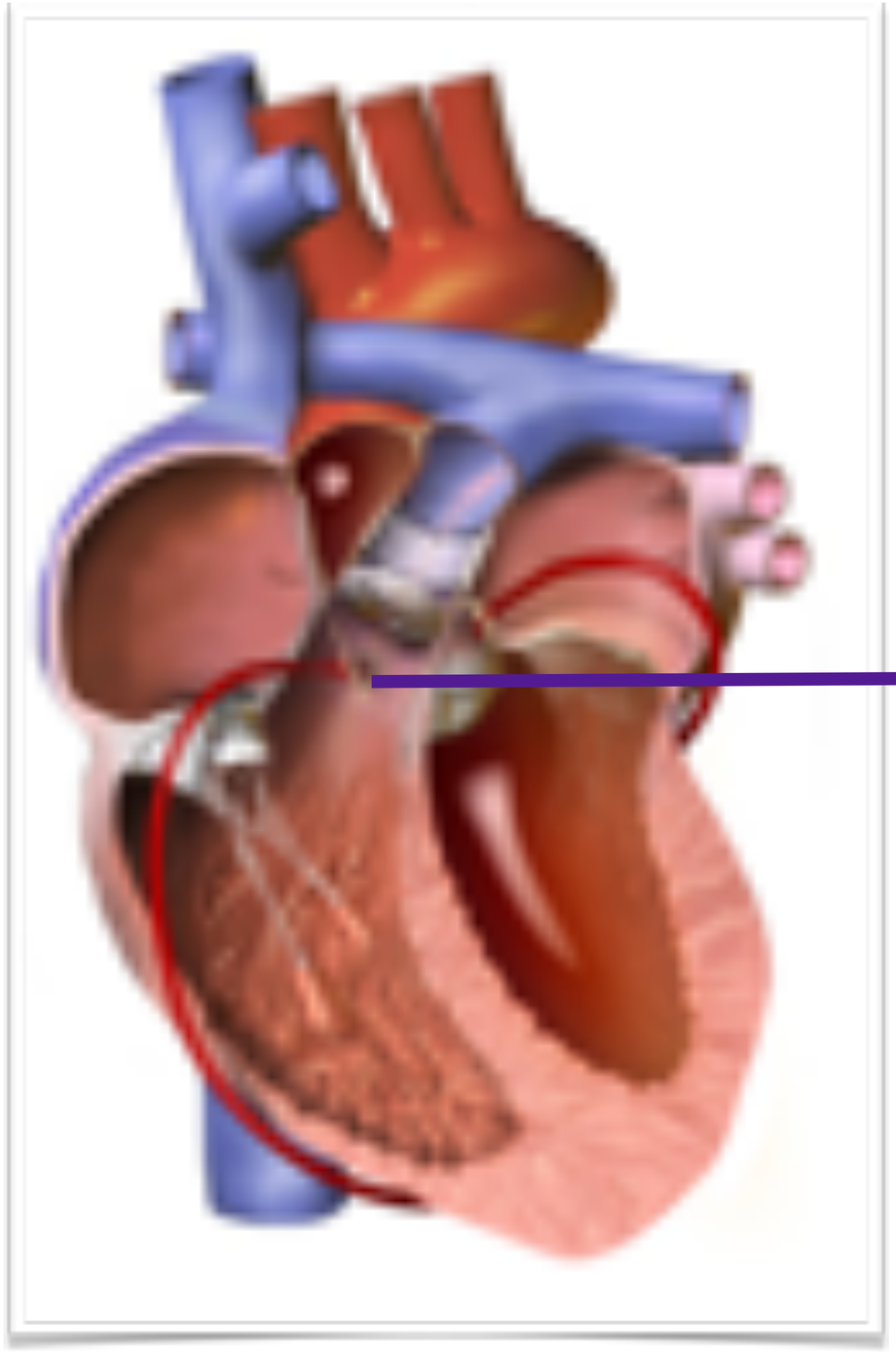





TGA VSD Coarctation



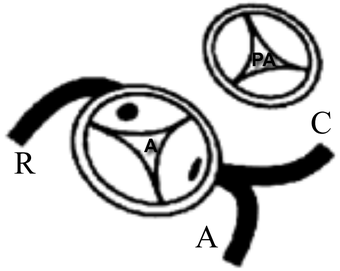




Coronary artery obstruction

Big variations in coronary anatomy (origin, loops, epicardial, intramural)

Type A



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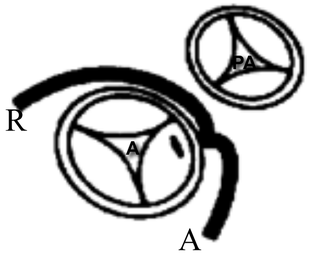
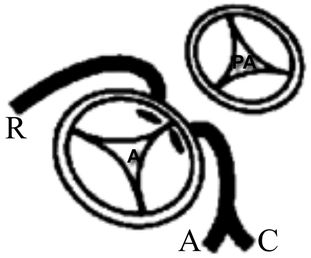
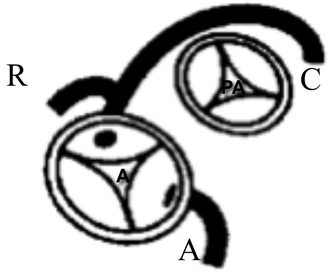


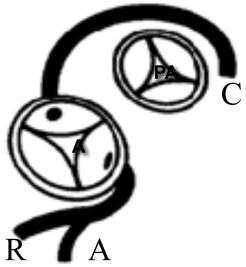
Type C

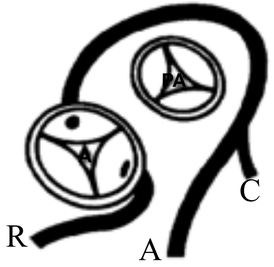


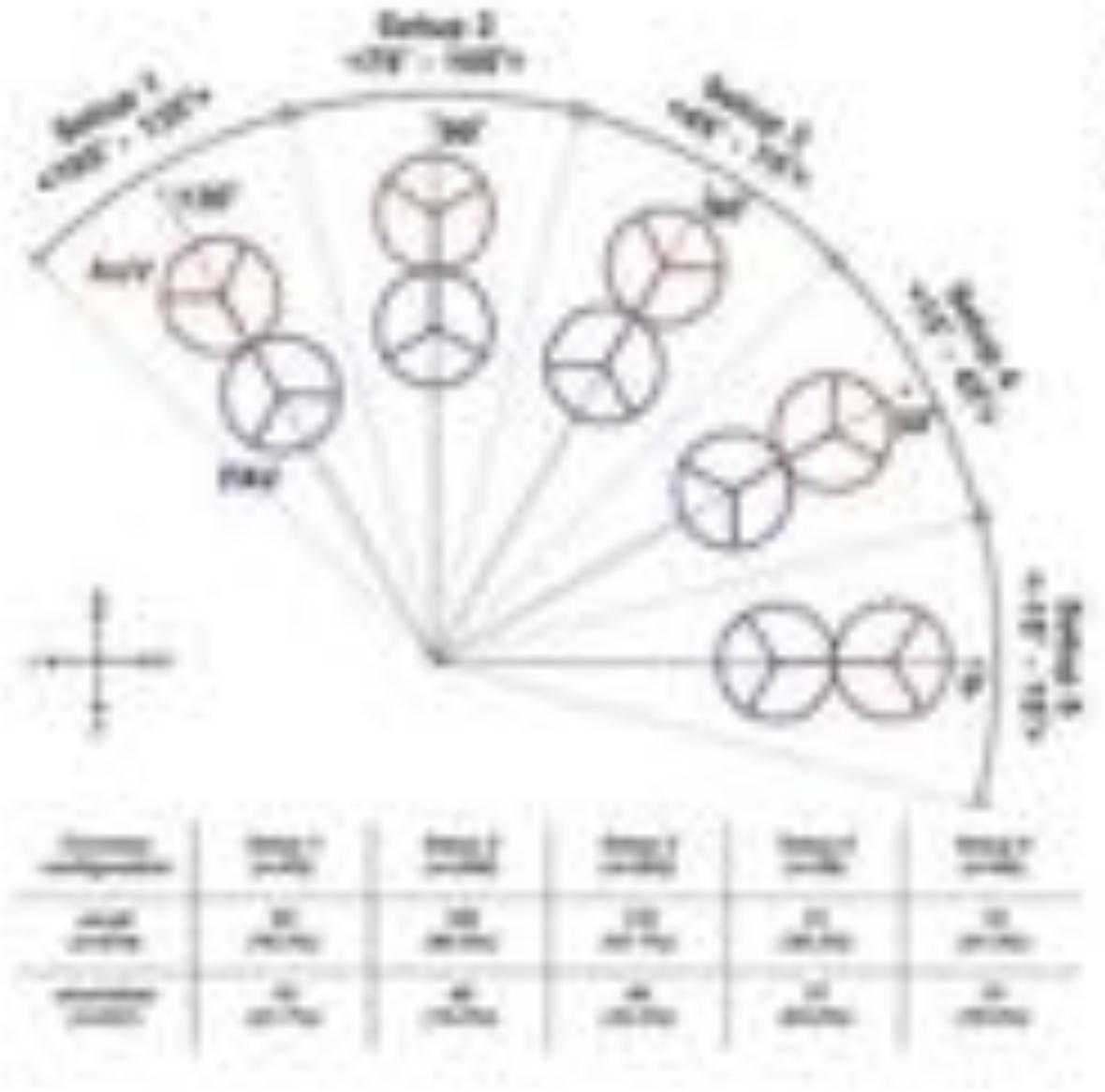
Type D



Type E

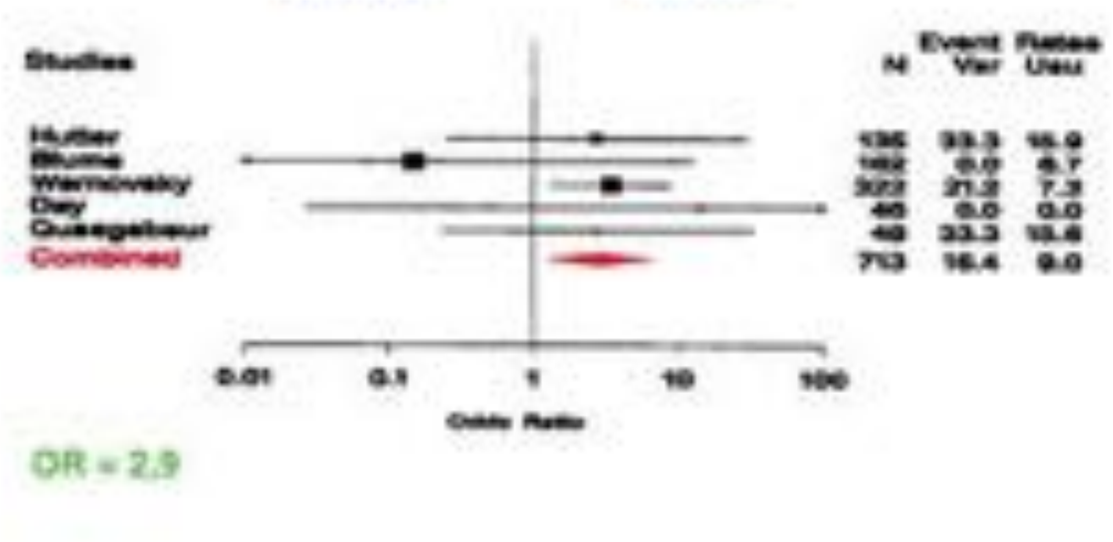




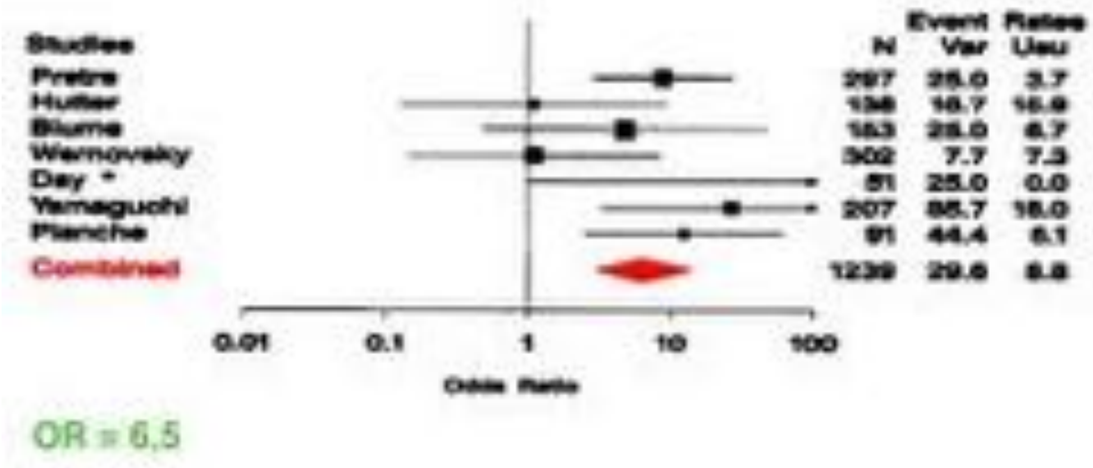


Coronary artery anatomy as a risk factor for early events

Single coronary artery



Intramural course

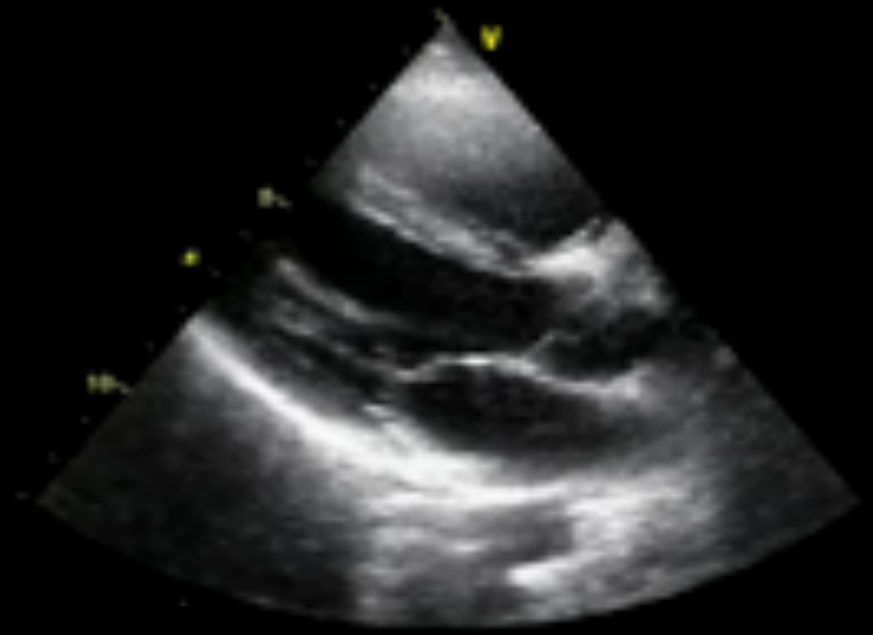


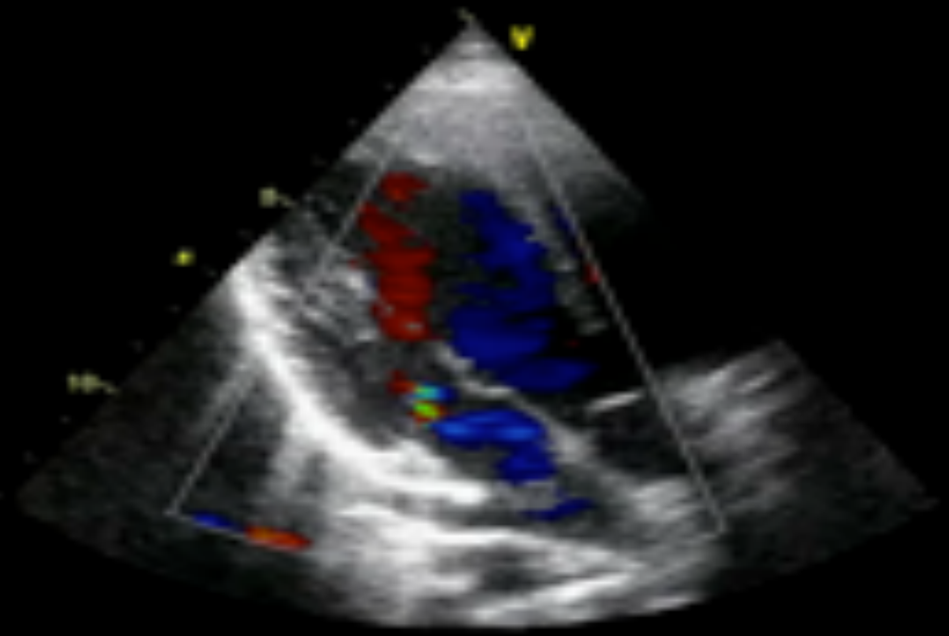
Pasquali, 2002

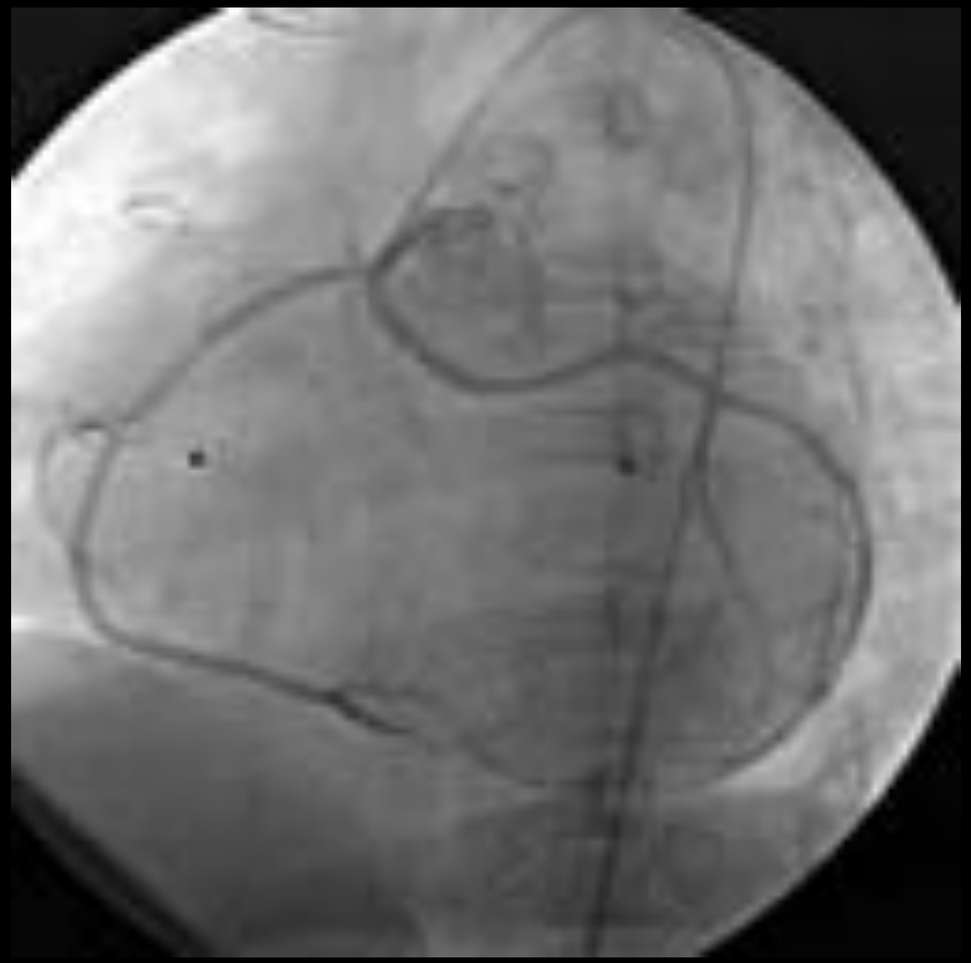
Outcomes and predictors of early mortality of the ASO for TGA with IVS

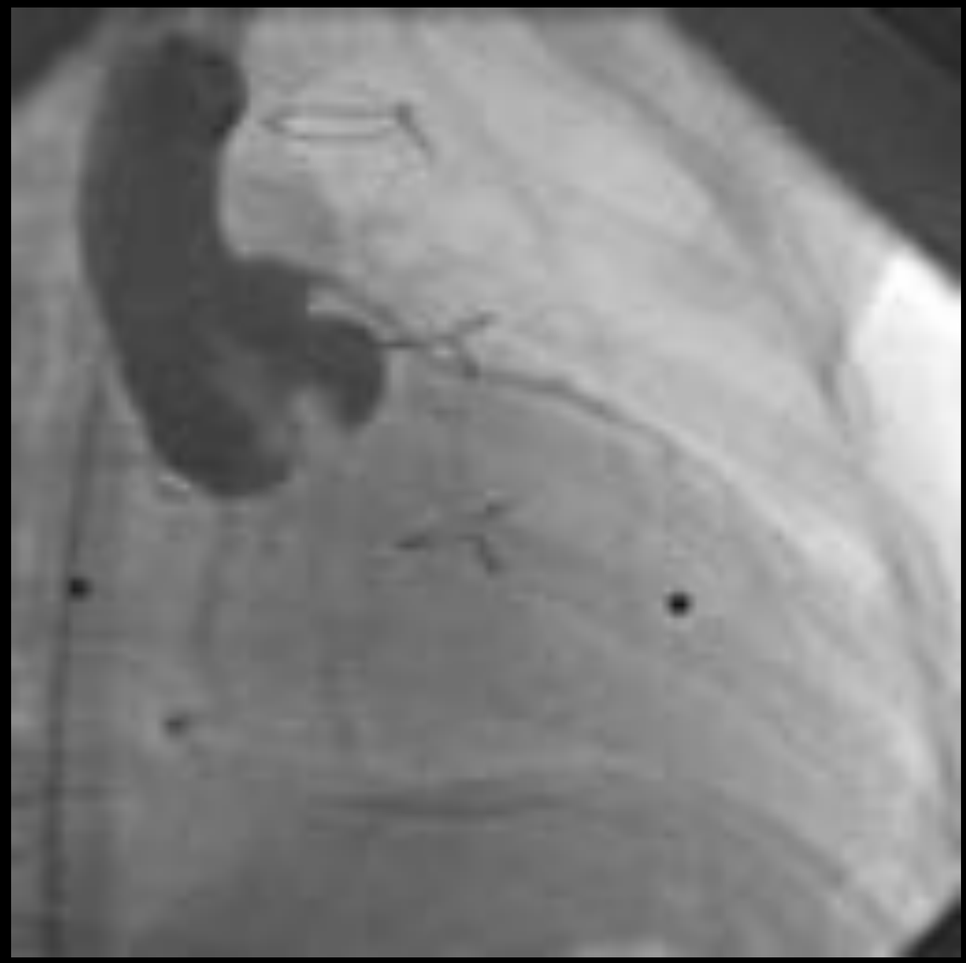
First Author (Ref. #), Year	Inclusive Years	N	% IVS	Early Survival for TGA IVS, %	5-Year Survival, %	10-Year Survival, %	Coronary Anatomic Risk Factors	Other Predictors of Early Mortality
Sarris [43], 2004[a]	1998-2000	613	70	97	NA	NA	Single coronary (univariate analysis only)	Open sternum
Lakovan [5], 2011	1977-2007	332	60.8	88.6	85.8[b]	85.2[b]	Not a risk factor for early mortality	Technical problems with coronary transfer
Fricke [45], 2012	1983-2009	618	64	98.2	98	97	Not a risk factor for early mortality	Weight <2.5 kg ECMO
Khairy [44], 2013	1983-1999	400	59.5	93.5[c]	NA	92.7[c]	Single right coronary artery	Post-operative heart failure
Cain [32], 2014	2000-2011	70	100	98.6	NA	NA	None identified	No predictors of early mortality, but earlier repair <4 days of age was associated with decreased resource utilization
Anderson [34], 2014	2003-2012	140	75	98.6	NA	NA	None identified	No predictors of early mortality, but earlier repair <4 days of age was associated with decreased resource utilization

Myocardial ischemia after the arterial switch for TGA





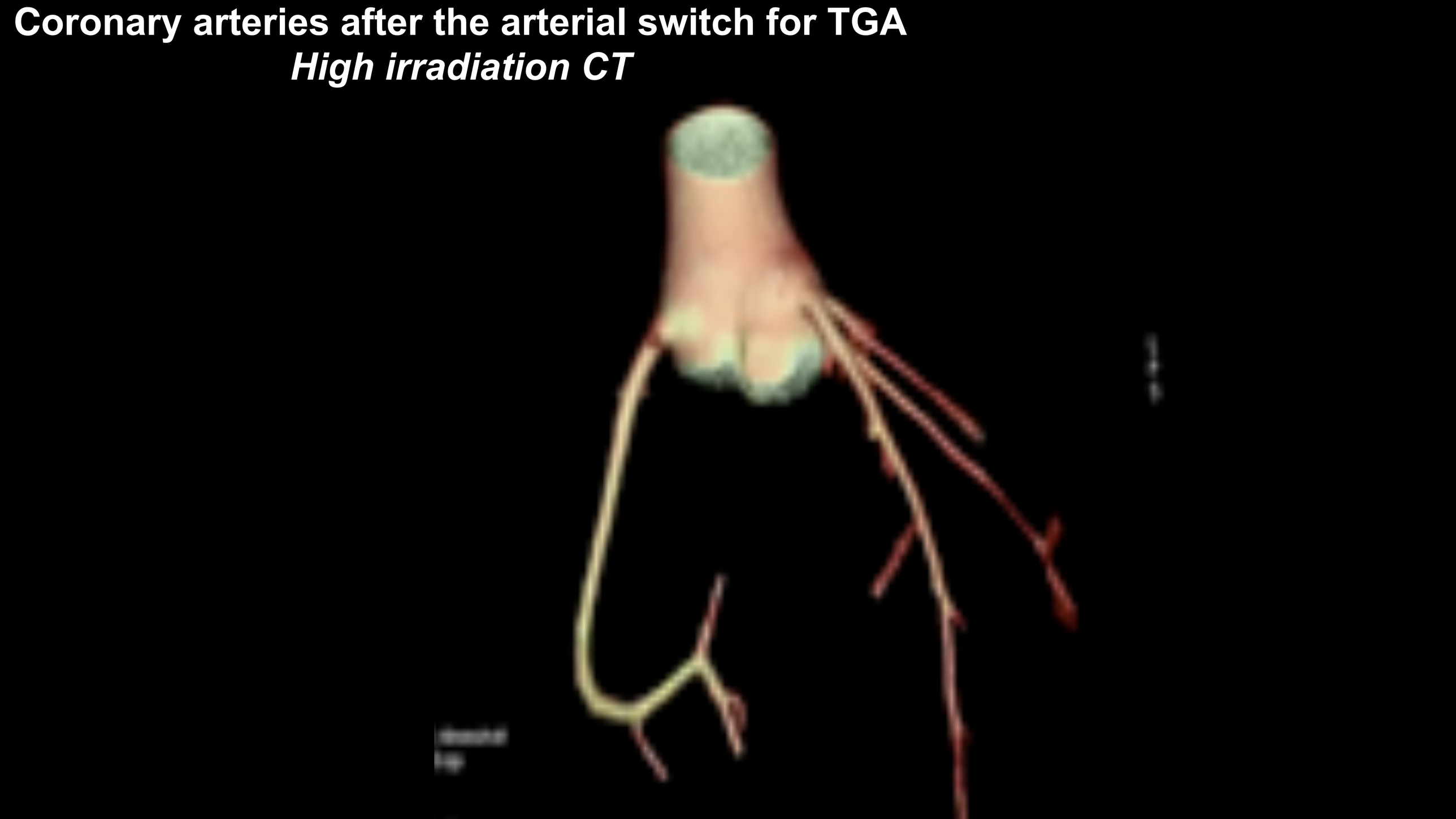




Type D coronary arteries after the arterial switch for TGA

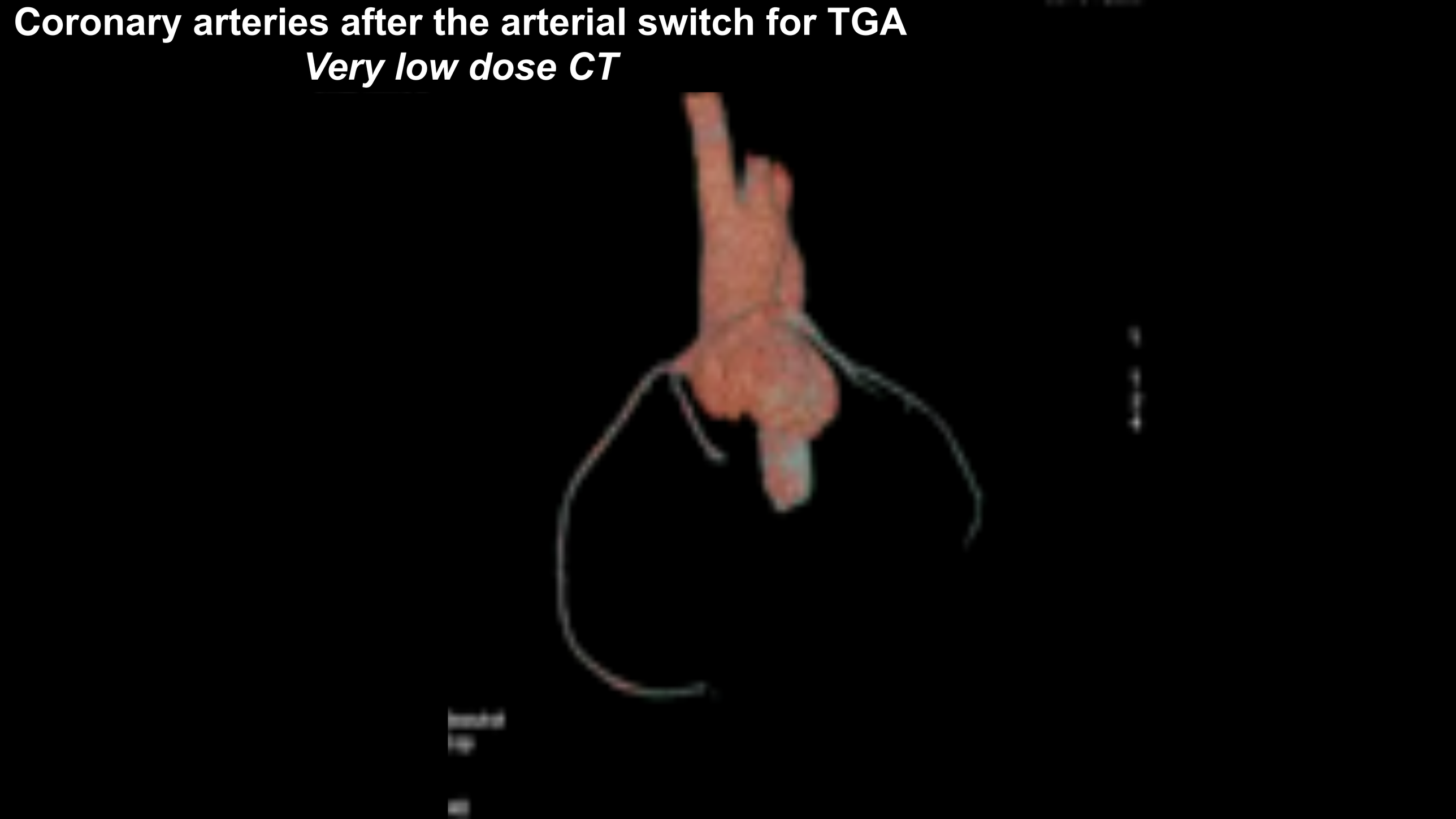
Coronary arteries after the arterial switch for TGA

High irradiation CT



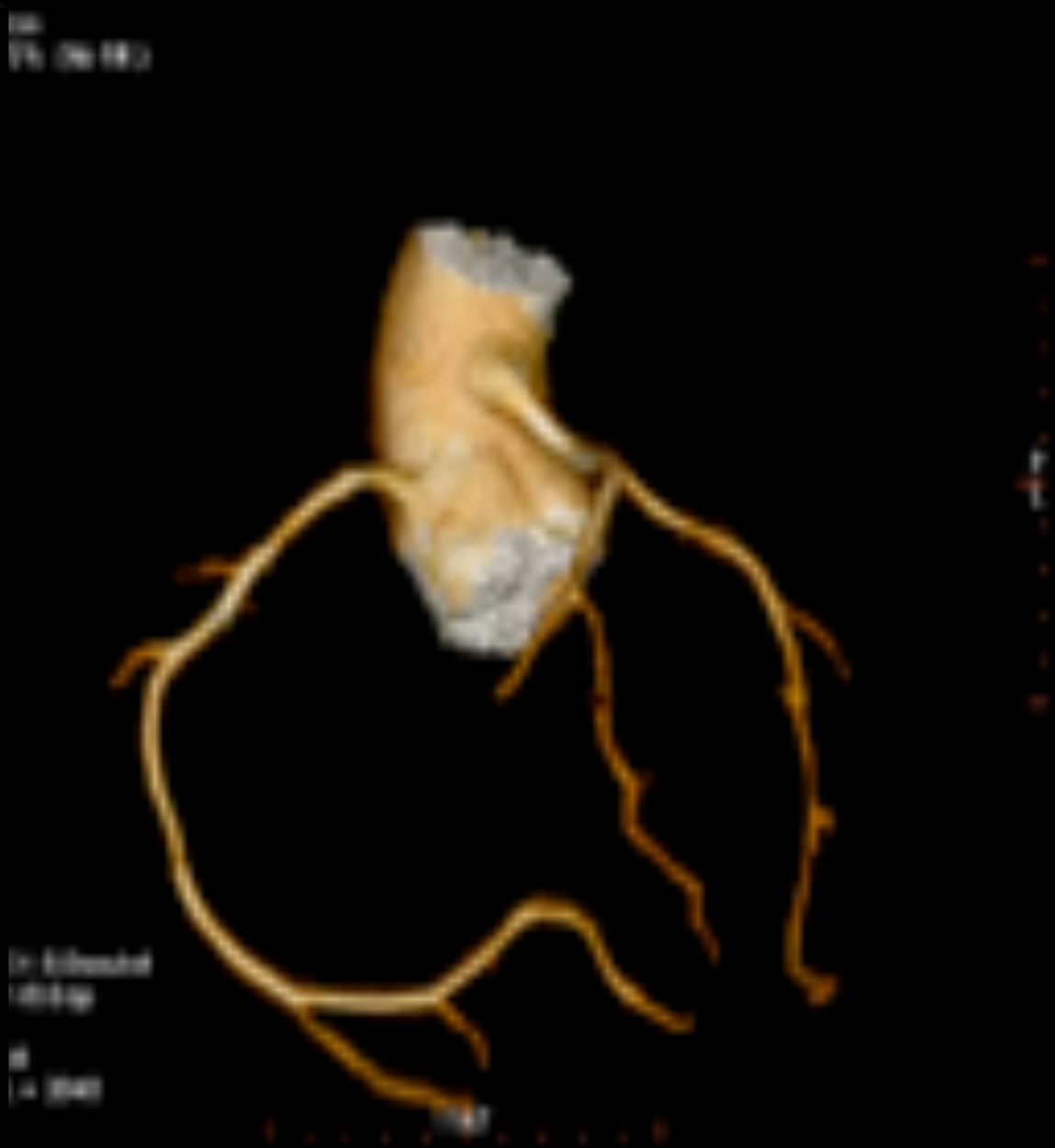
Coronary arteries after the arterial switch for TGA

Very low dose CT



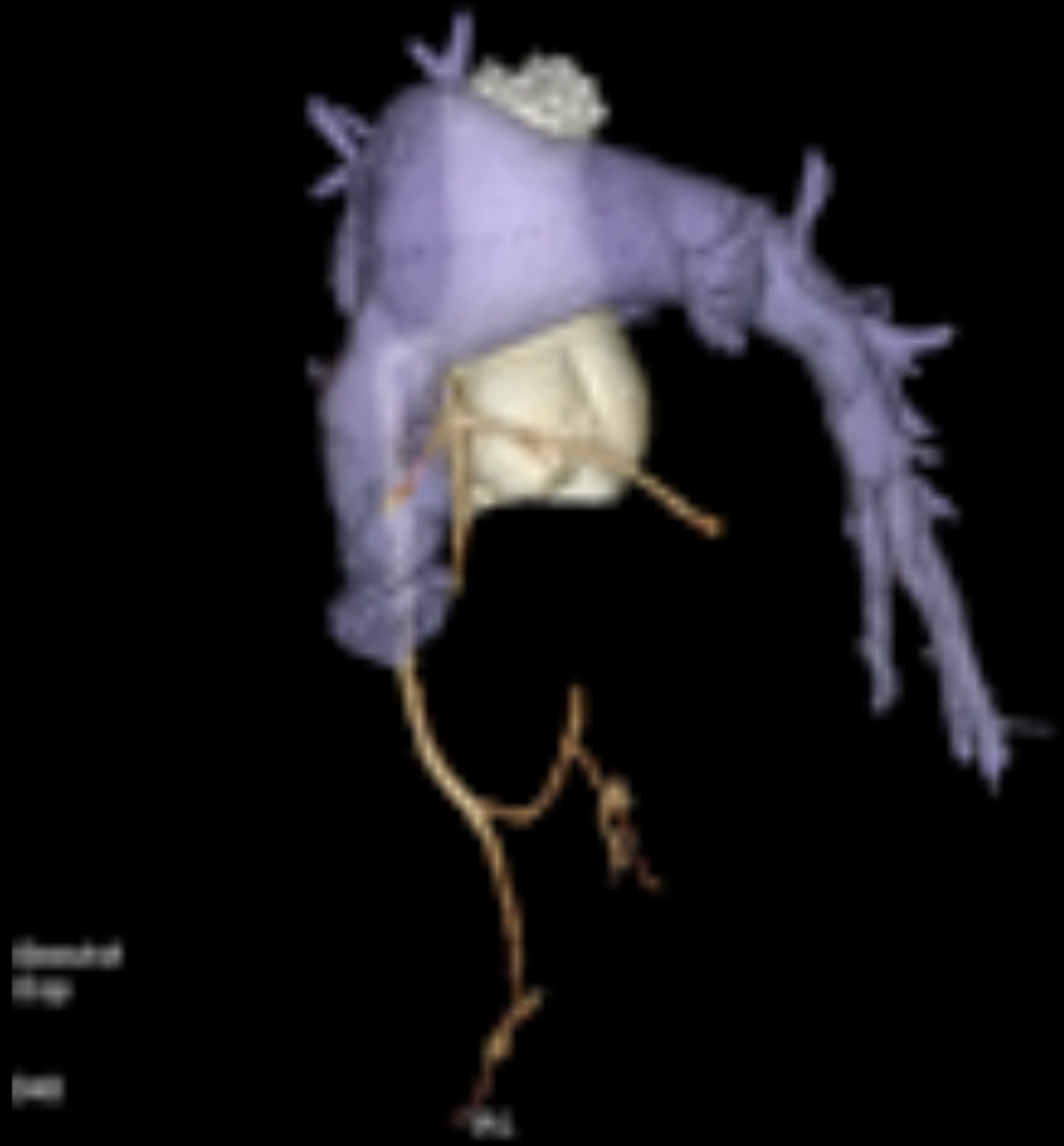
Coronary arteries after the arterial switch for TGA

High reimplantation of left CA



Coronary arteries after the arterial switch for TGA

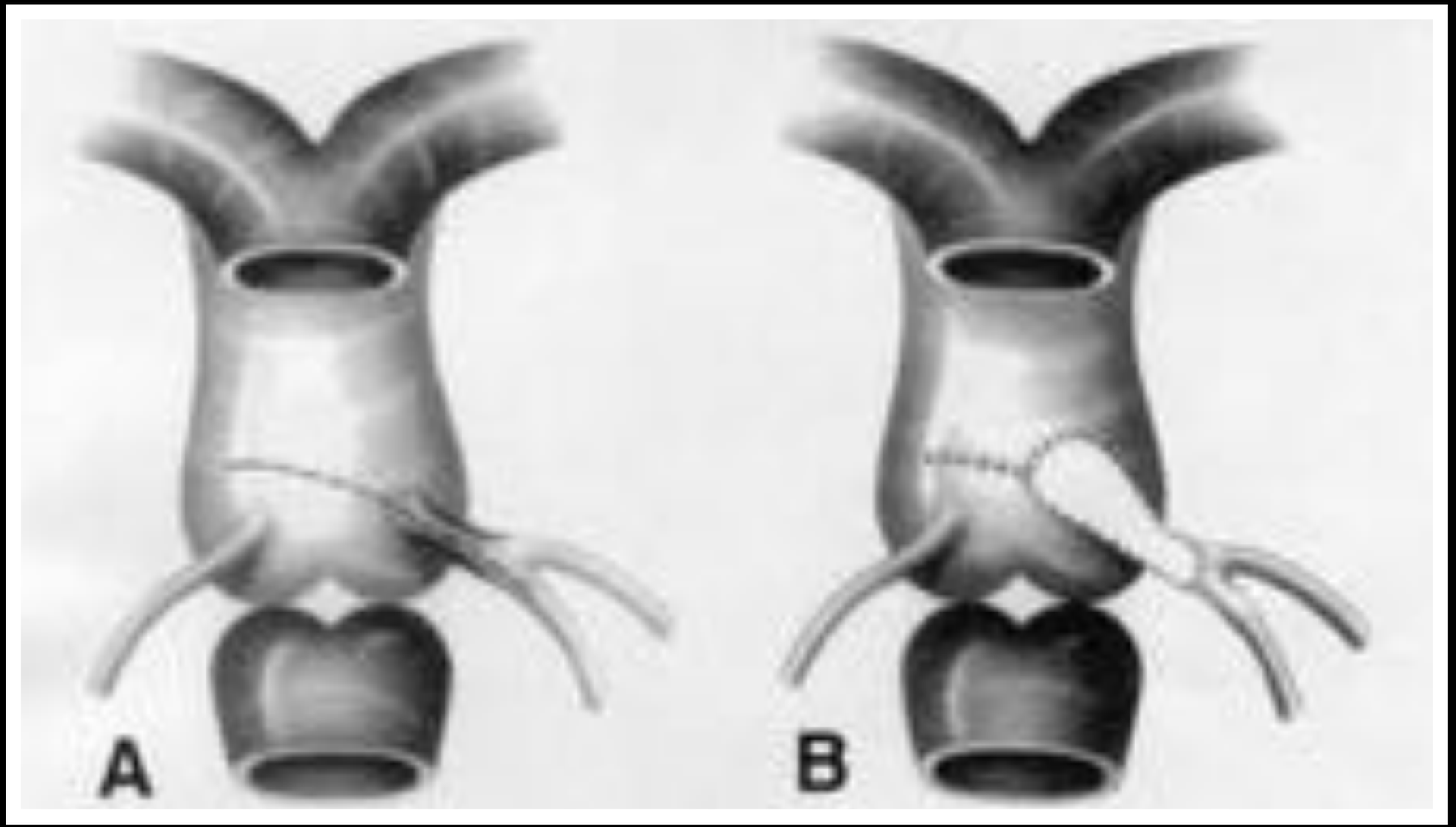
Intervascular course fo left CA

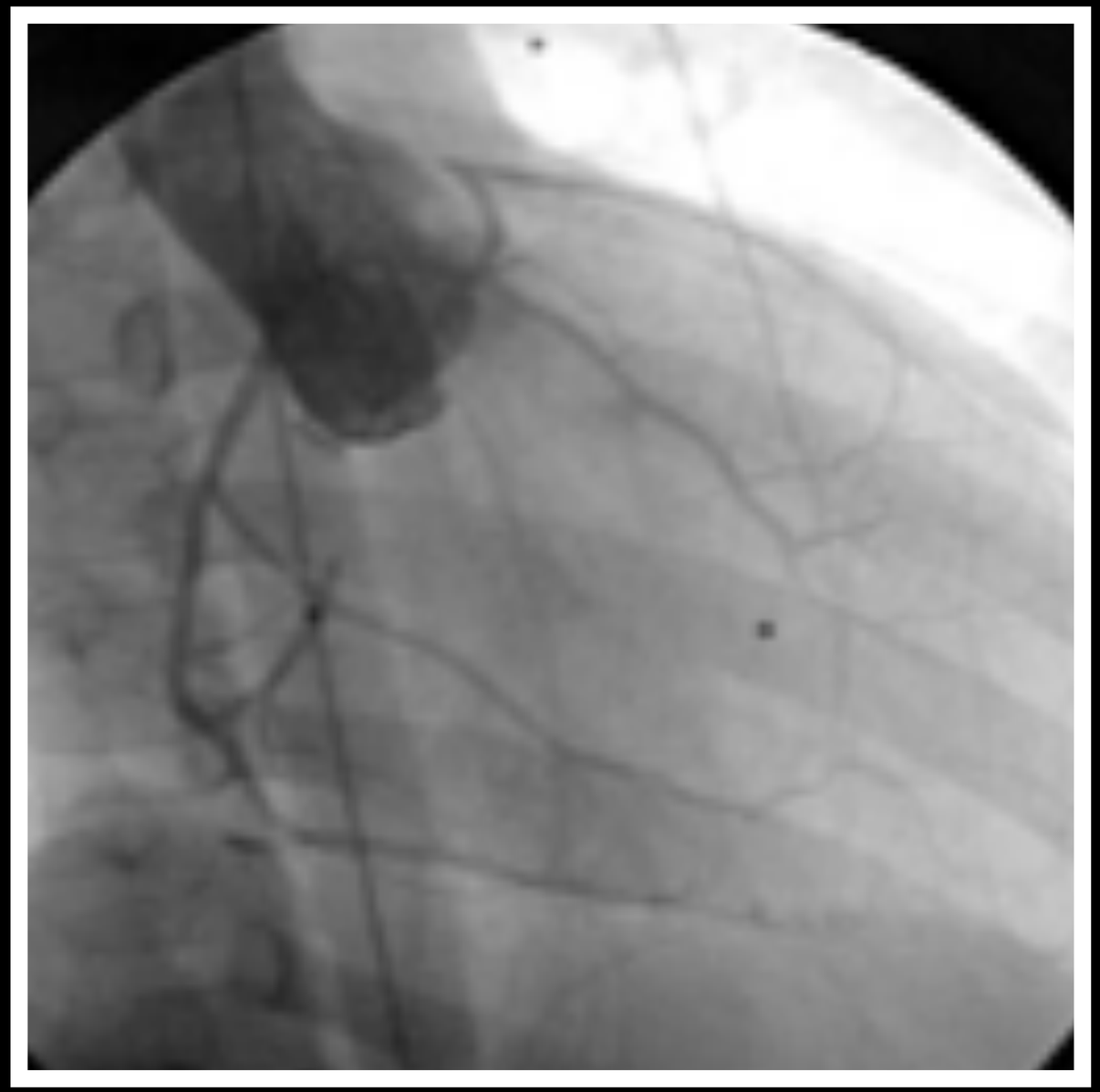


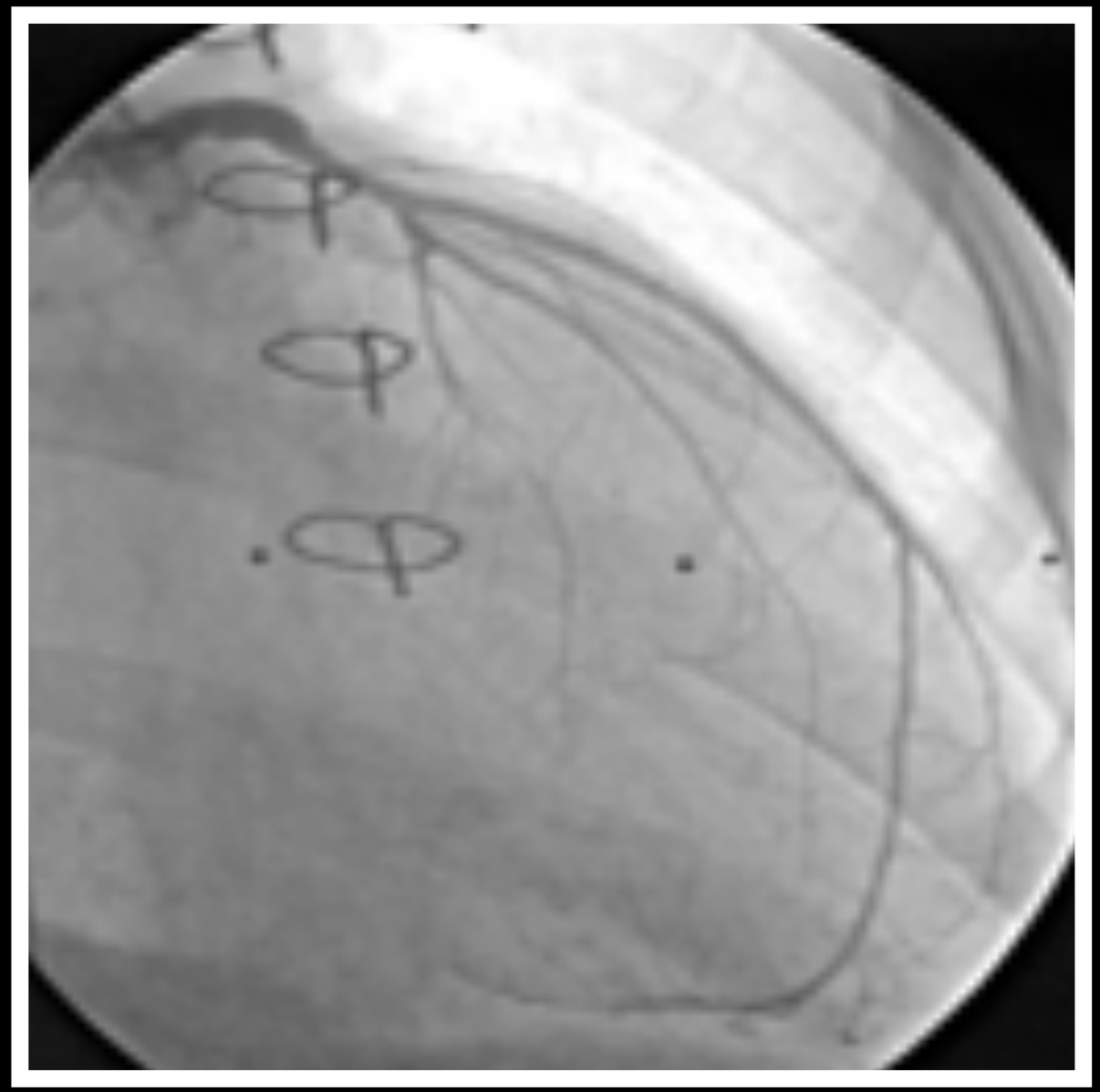
Sudden death due to coronary artery lesions long-term after the arterial switch operation: a systematic review

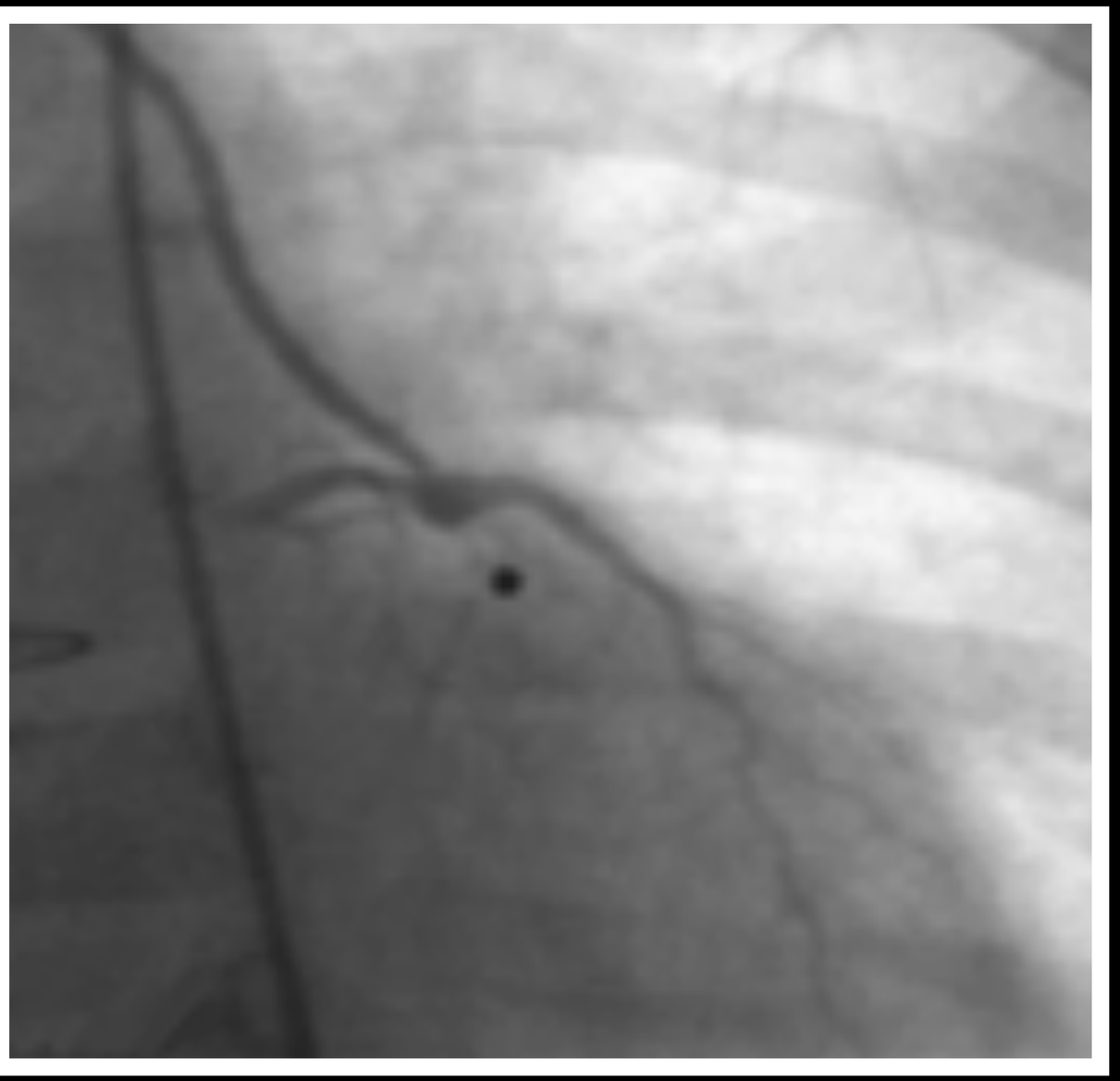
- 52 studies : sudden death because of coronary complications in survivors after 5 years
- 8798 patients: 27 deaths > 5 years post-ASO (0.3%)
  - 10 were known with relevant residual lesions
  - 5 sudden death possibly from cardiac cause, no late death confirmed to be coronary related
- **Routine coronary imaging of asymptomatic single-stage ASO patients is not justified**

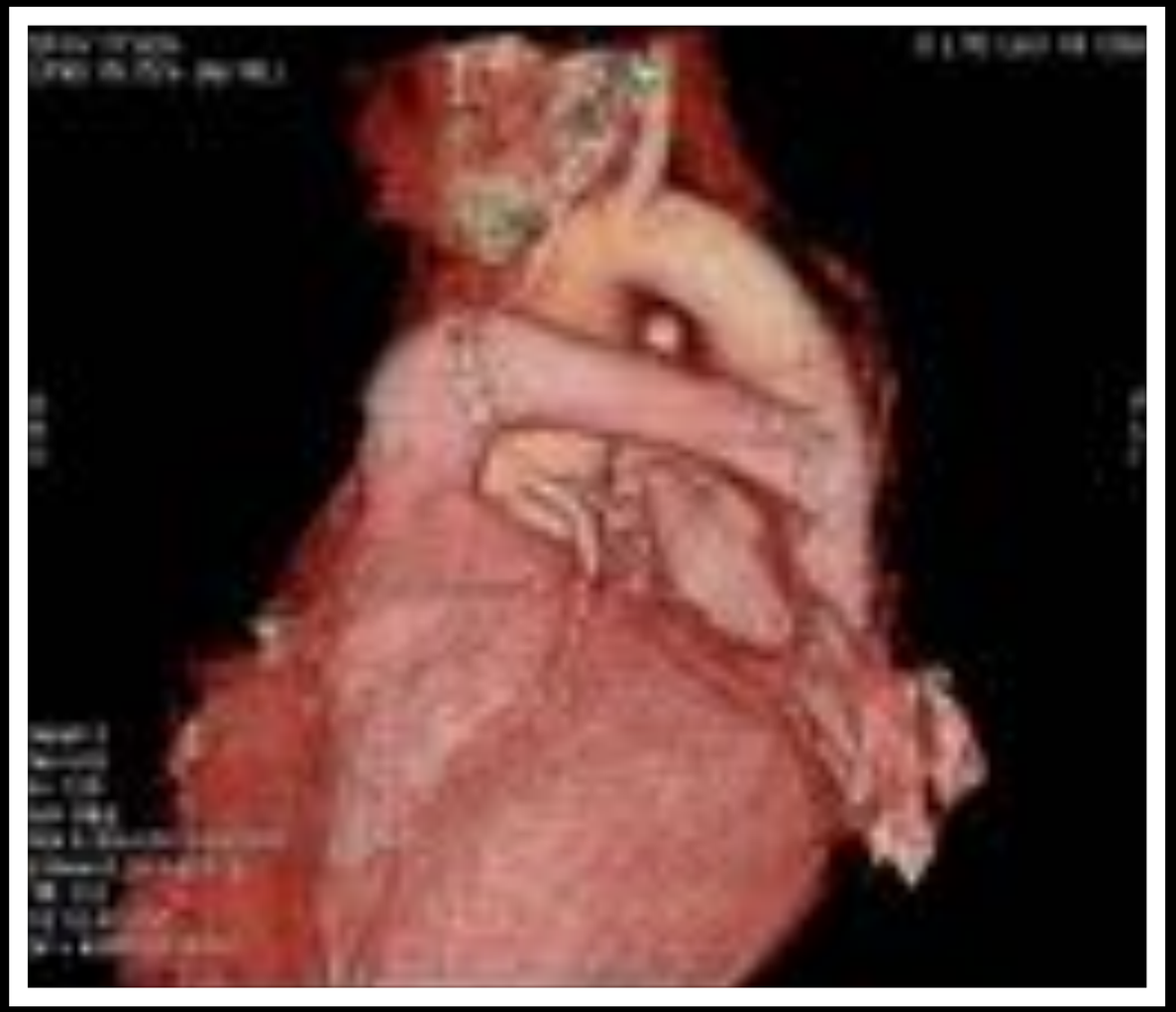
van Wijk SWH et al. Can J Cardiol 2017 33;1180-87


A
B

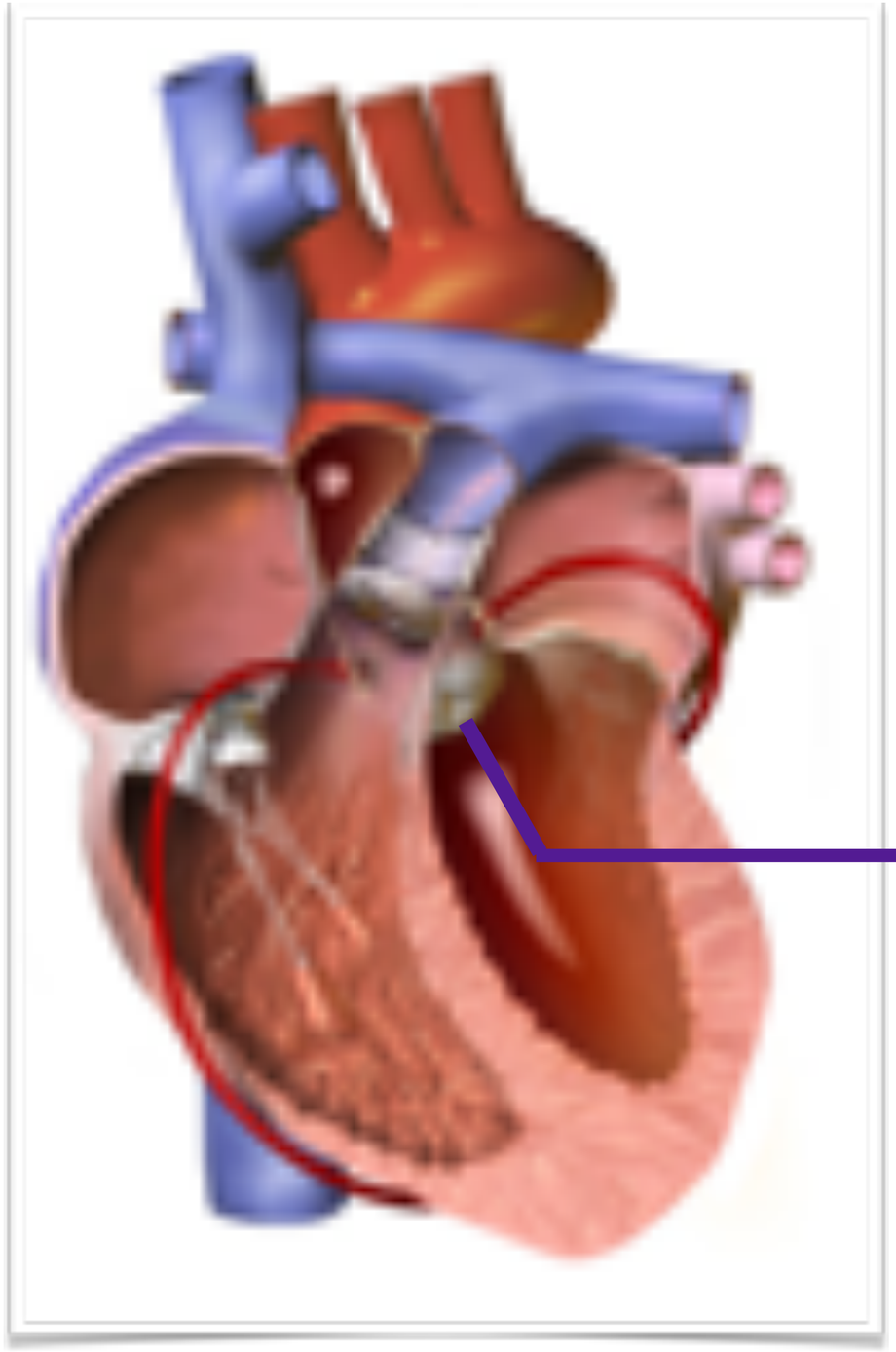






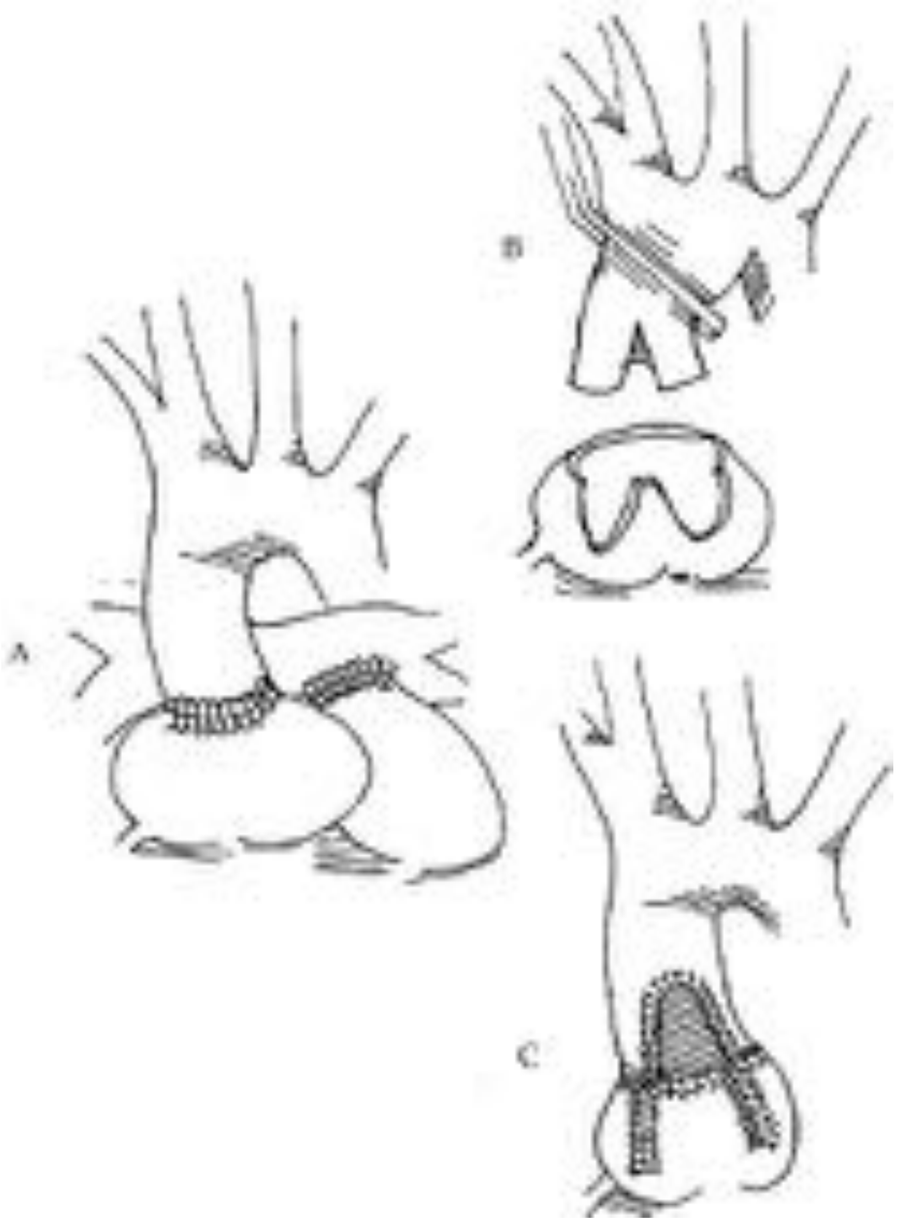


Pontage mammaire interne-IVA post-switch



Aortic valve regurgitation

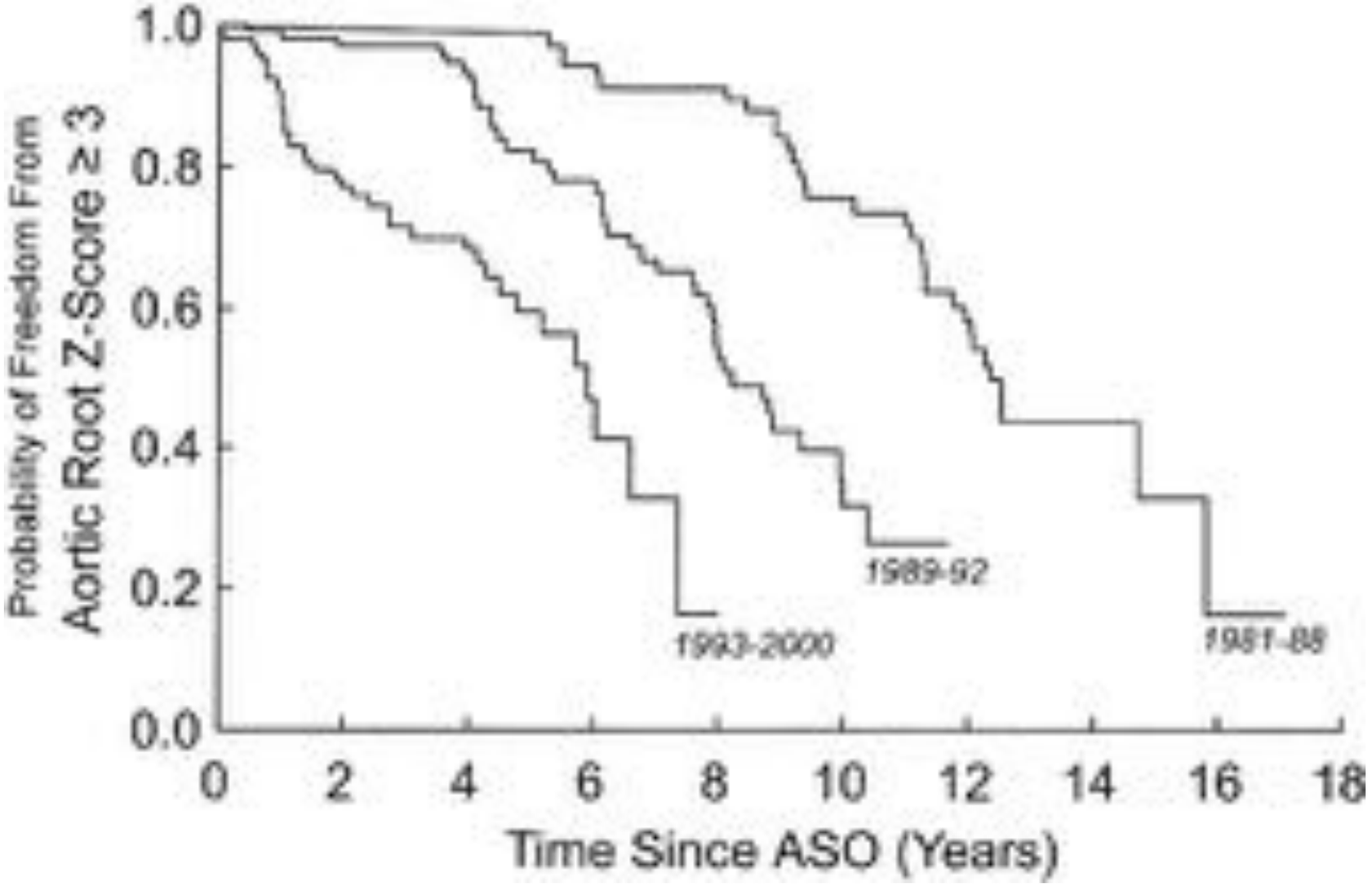
Aortic valve repair for AR after ASO ?



Imamura M. et al., Ann Thorac Surg 2000;69:607-608

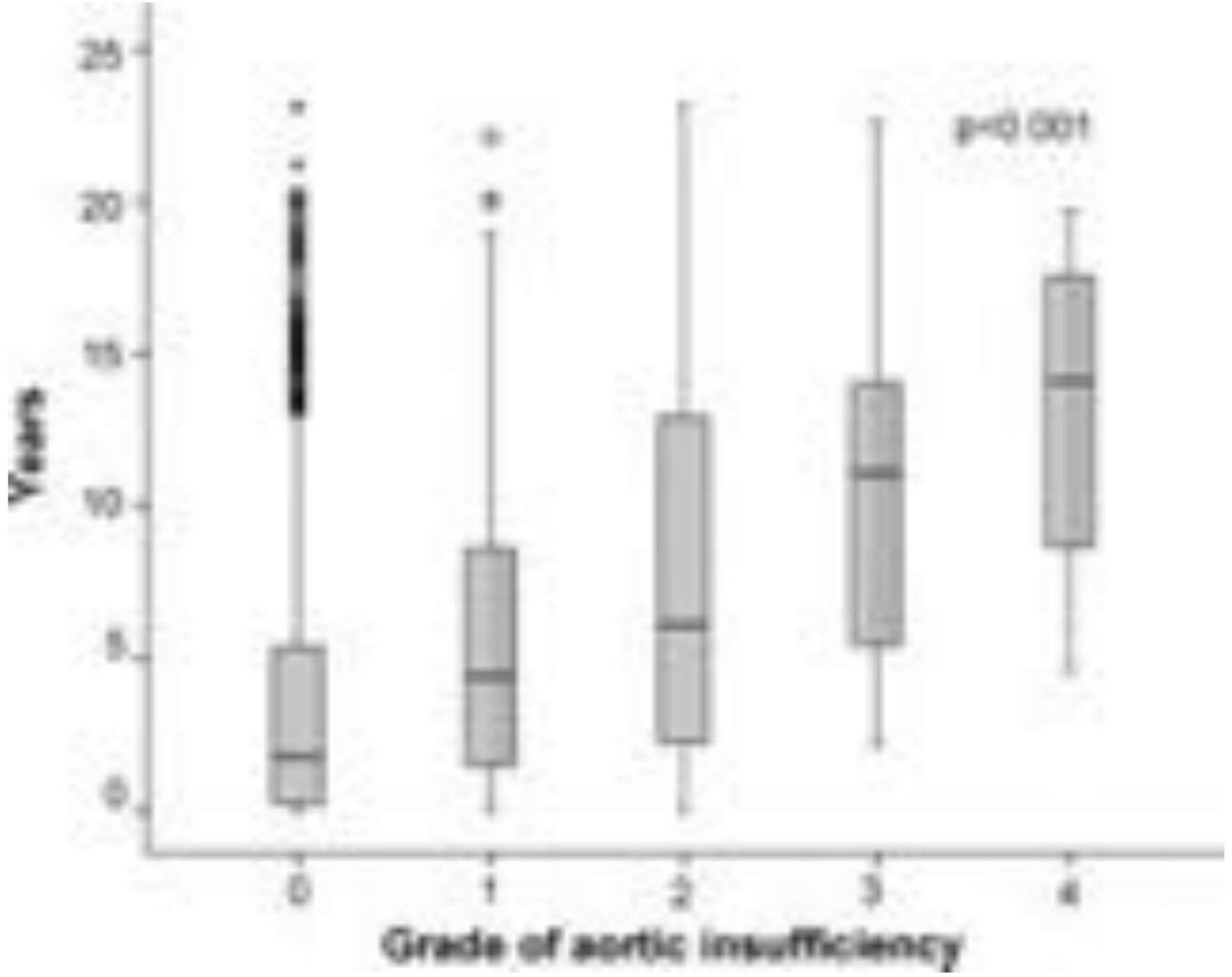
Freedom from aortic root dilation over time since ASO

3 time periods : 1981 to 1988, 1989 to 1992, and 1993 to 2000 ($P<0.001$)



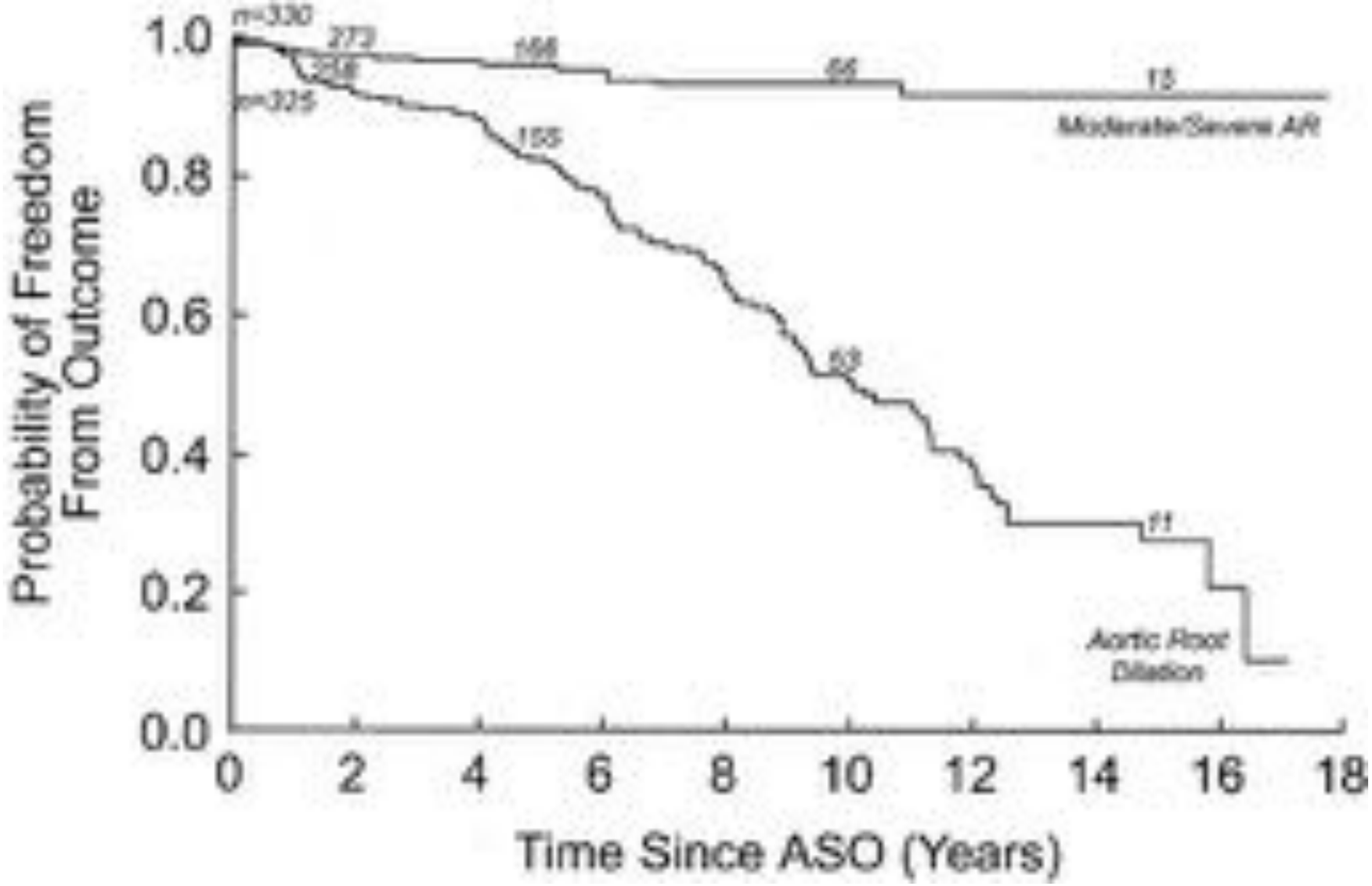
Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132

Development of Aortic Insufficiency over time

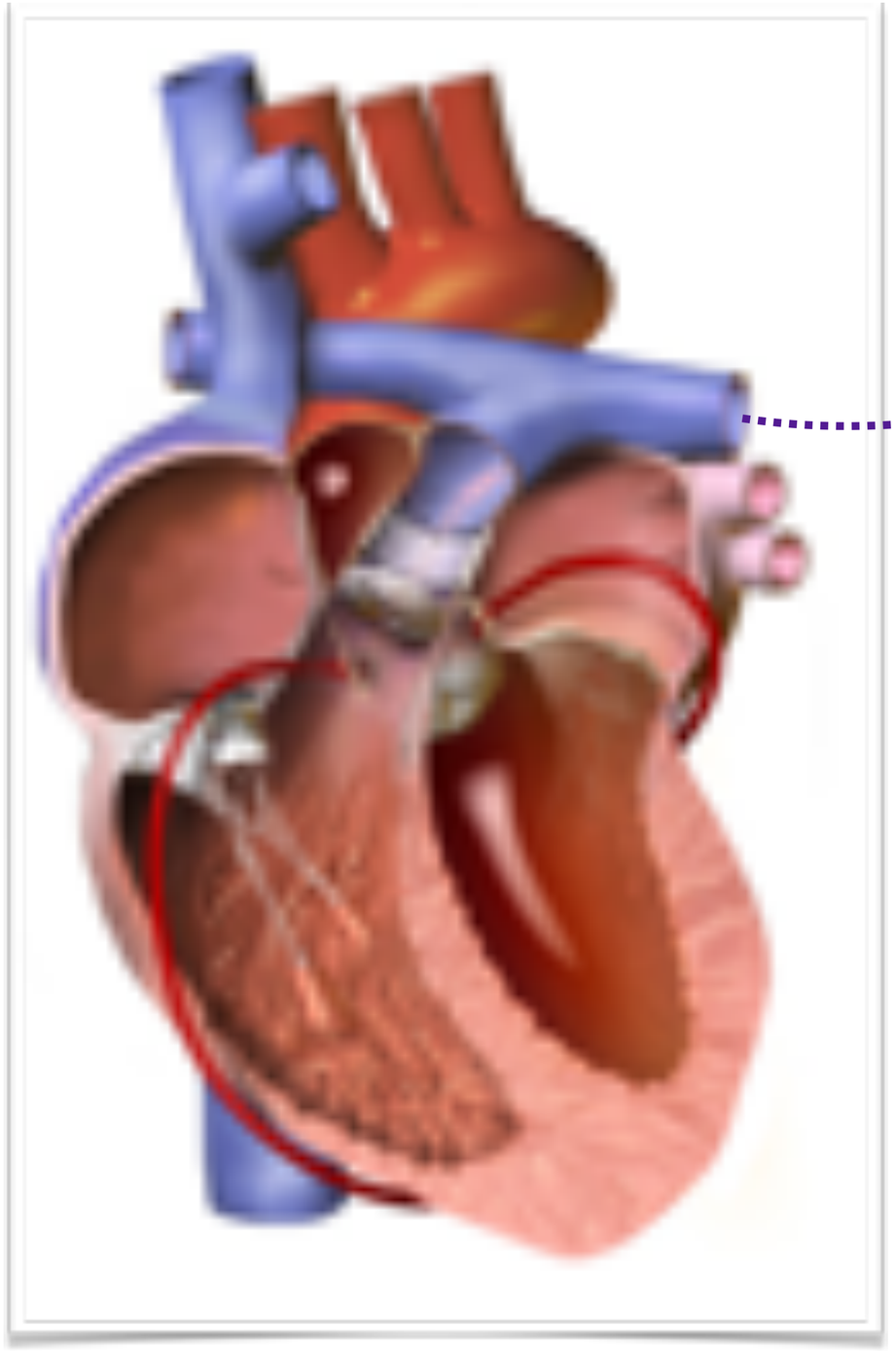


Lange R. et al.; Eur J Cardiothorac Surg 2008;34:711-717

Freedom from neo-aortic root dilation (neo-aortic root z-score >=3.0) and probability from at least moderate neo-aortic regurgitation

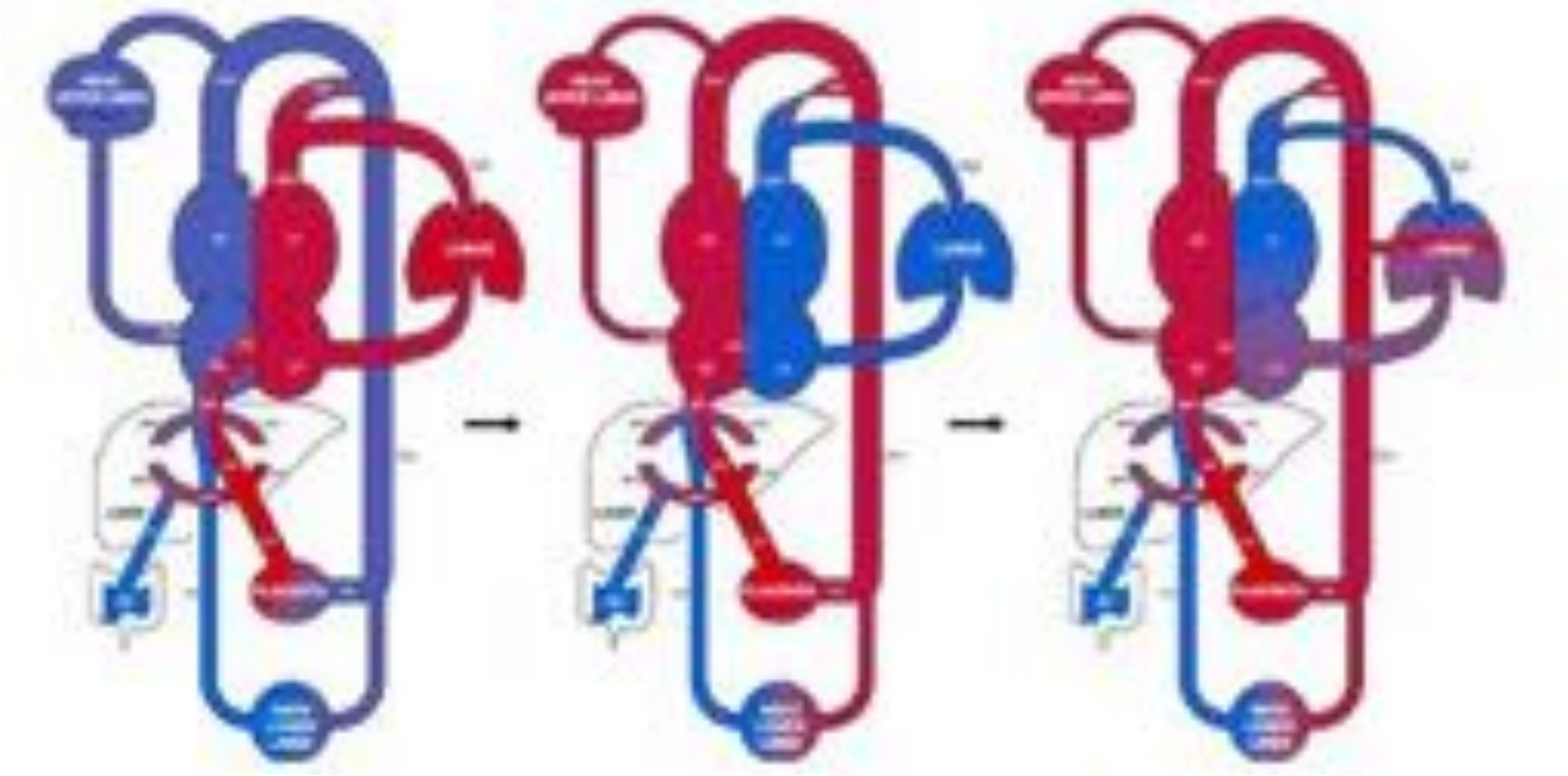


Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132



Pulmonary arterial hypertension

Pulmonary circulation in fetal TGA



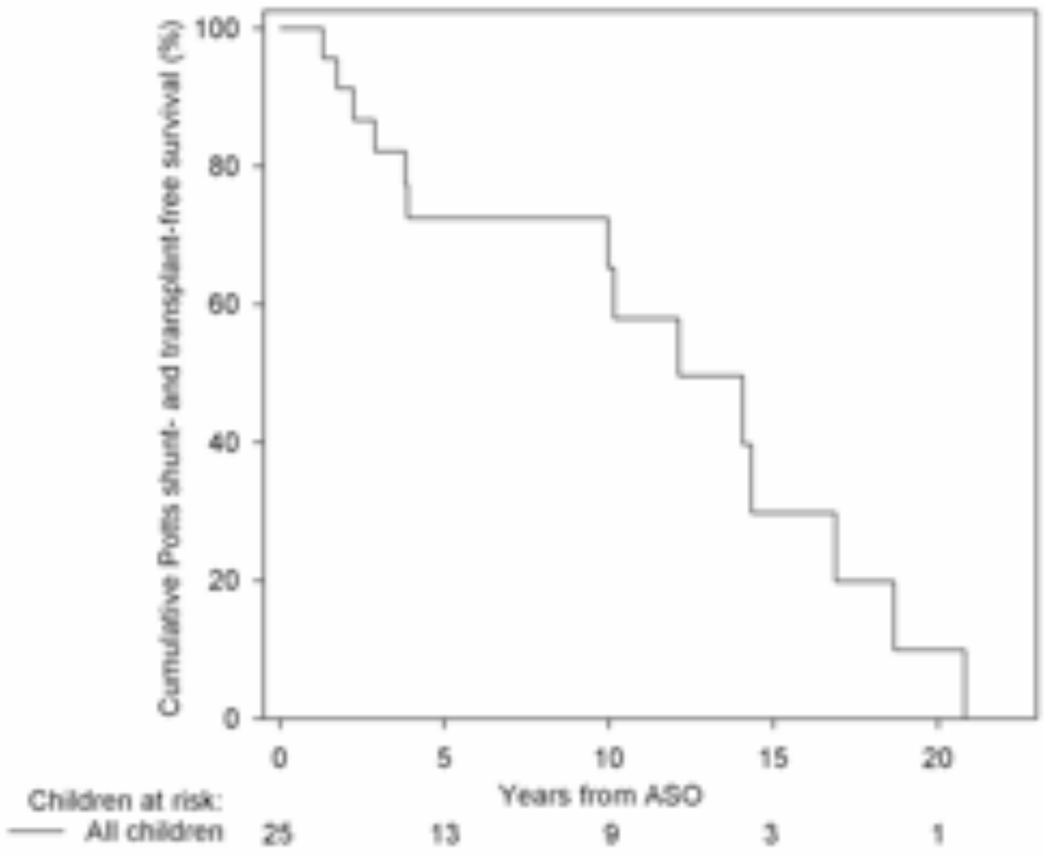
Initial increase in PBF due to vasodilatation with increased oxygen
Increased pulmonary venous return
Reduced size of the FO
Ductal constriction due to oxygen
Isolation of Pulmonary circulation
Increased PVR
Development of aorta-pulmonary collaterals

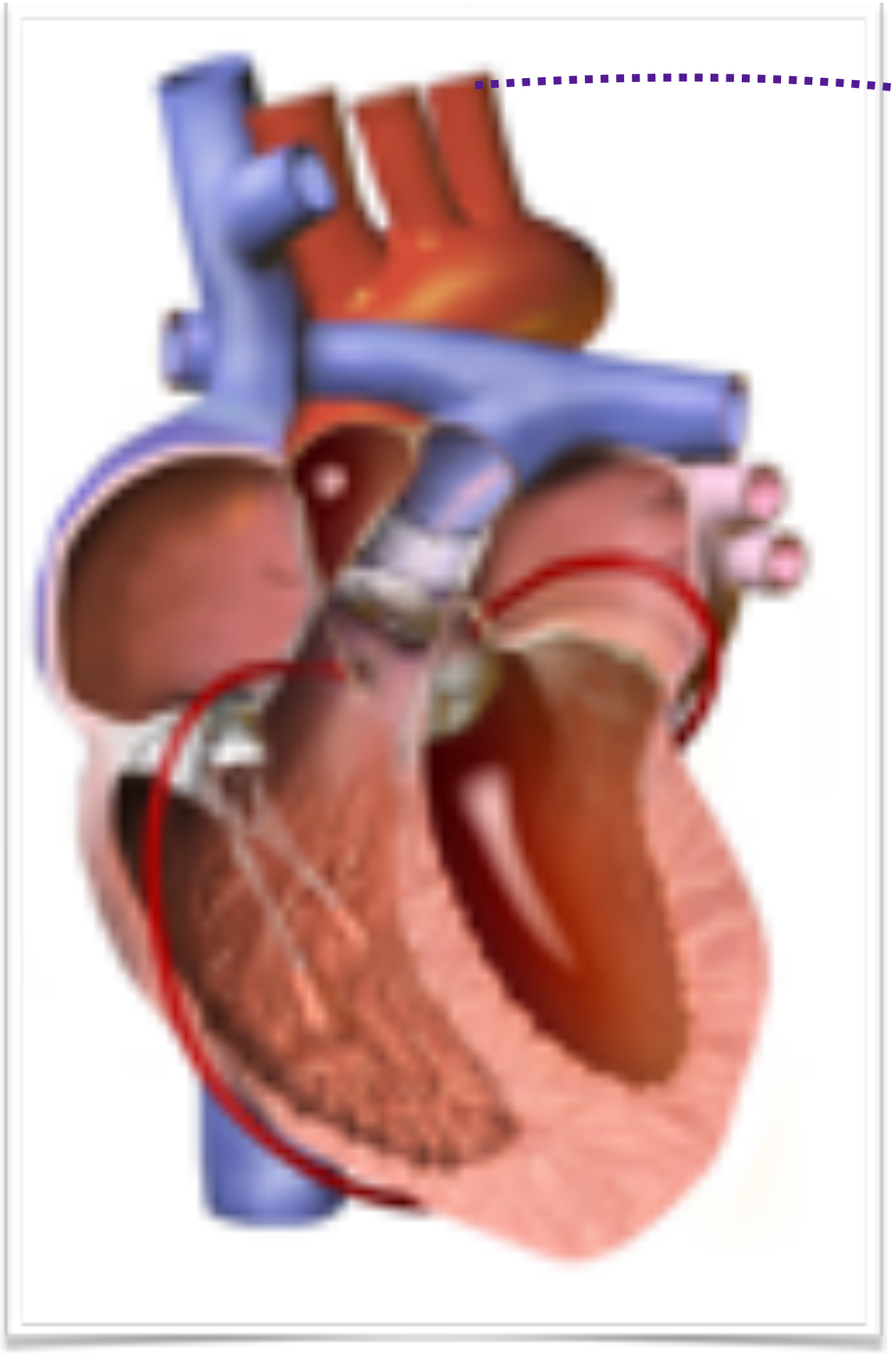
PAH characteristics after ASO for TGA

	Value
Age first PAH detection (months)	4 (1, 25)
PAH detection within one year after ASO	16 (64)
Age first detection (months)	2 (1, 4)
PAH detection more than one year after	9 (36)
Age first detection (months)	40 (25, 93)
Age first RHC (months)	10 (2, 30)
mPAP (mmHg)	48 (37, 55)
mSAP (mmHg)	62 (51, 65)
mPAP/mSAP	0.82 (0.71, 1.07)
mPCWP (mmHg)	10 (8, 12)
PVRi (WU.m²)	11.5 (8.7, 13.0)
PAH therapy at endpoint	
CCB monotherapy	1 (4)
PAH-targeted mono therapy	6 (24)
PAH-targeted dual therapy	8 (32)
PAH-targeted triple therapy	10 (40)

Ziljstra W et al. Heart 2017

Survival in PAH after ASO for TGA





Neurodevelopmental outcomes

ToM tests in TGA vs controls

Cognitive domain	Test	TGA (n=45)	Controls (n=45)	*p*
IQ	CMMS	113 (8.3)	116 (8.85)	*ns*
Receptive Language	NEPSY - Comprehension	12.4 (0.80)	12.5 (0.81)	*ns*
Motor Inhibition	NEPSY – Knock and tap	24.25 (3.81)	25.97 (2.12)	0.01
Cognitive Inhibition	Stroop test (errors)	3.08 (3.02)	1.42 (1.48)	0.001
	Stroop test (Reaction time)	82.42 (31.61)	61.03 (20.53)	0.0002
Verbal working memory	Digit span WISC IV	2.84 (2.49)	3.64 (2.55)	*ns*
Spatial working memory	BEM-144 blocks	3.06 (2.12)	4 (2.03)	0.03
Cognitive flexibility	DCST	7.28 (2.86)	8.66 (2.09)	0.01
Social cognition	Theory of mind tests	0.95 (1.27)	2.15 (1.24)	0.0009

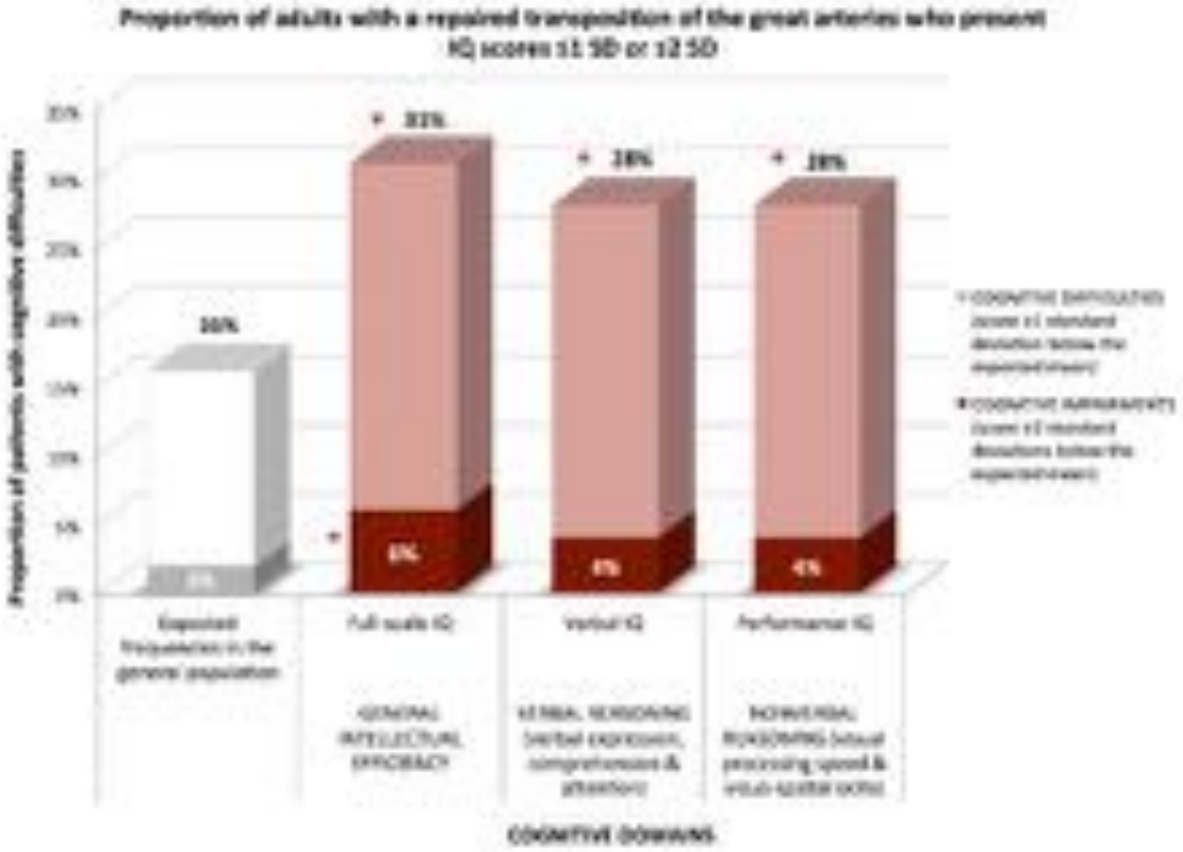
Calderon J et al. J Pediatr 2012

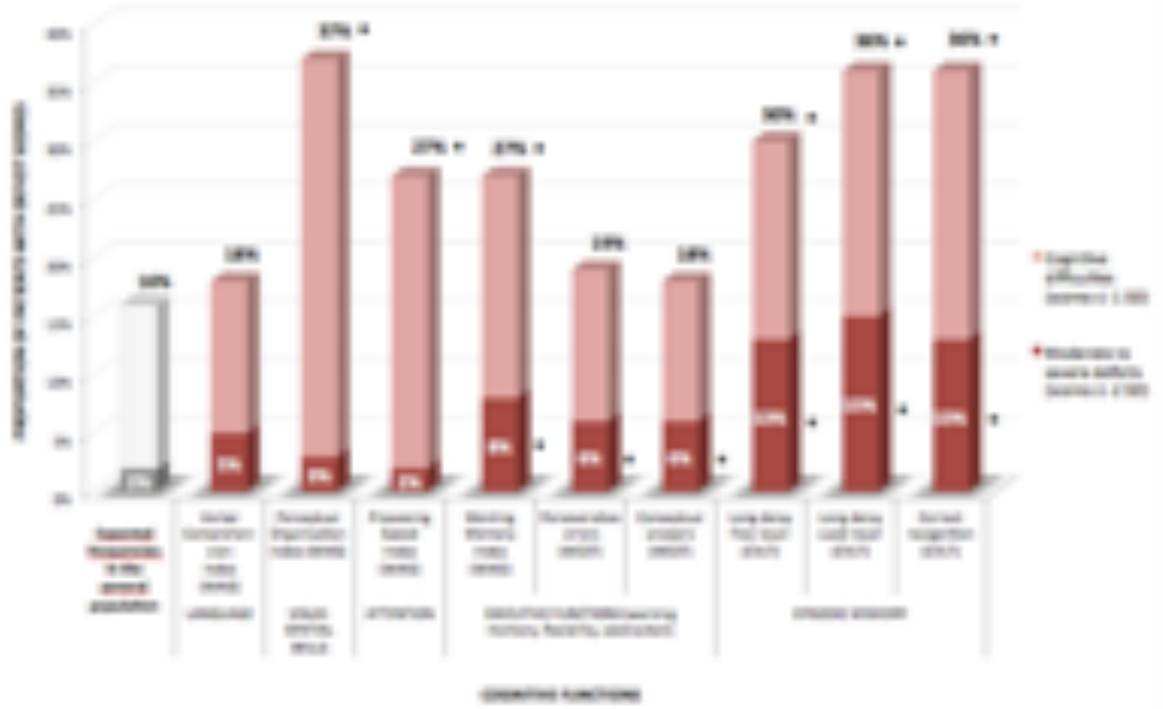
ToM tests in TGA vs controls
role of prenatal diagnosis

Cognitive Domain	Test	Prenatal (n=29)	Postnatal (n=16)	*p*
IQ	CMMS	114.5 (8.50)	112.4 (8.06)	0.4
Receptive Language	NEPSY - Comprehension	12.65 (0.55)	12.25 (1.12)	0.11
Response motor control	NEPSY – Knock and tap	24.31 (2.46)	24.14 (5.82)	0.89
Cognitive control	Stroop test (Number of errors)	2.41 (2.48)	4.31 (3.59)	0.04
	Stroop test (Reaction Time)	77.82 (28.05)	90.74 (36.71)	0.19
Verbal working memory	Digit span WISC IV	2.96 (2.48)	2.62 (2.57)	0.66
Spatial working memory	BEM-144 blocks	3.62 (2.0)	2.06 (2.01)	0.01
Cognitive flexibility	DCST	8.10 (2.65)	5.64 (2.61)	0.006
Social cognition	Theory of mind	1.31 (1.33)	0.31 (0.87)	0.01

Calderon J et al. J Pediatr 2012

Neurodevelopmental outcomes after ASO in adults



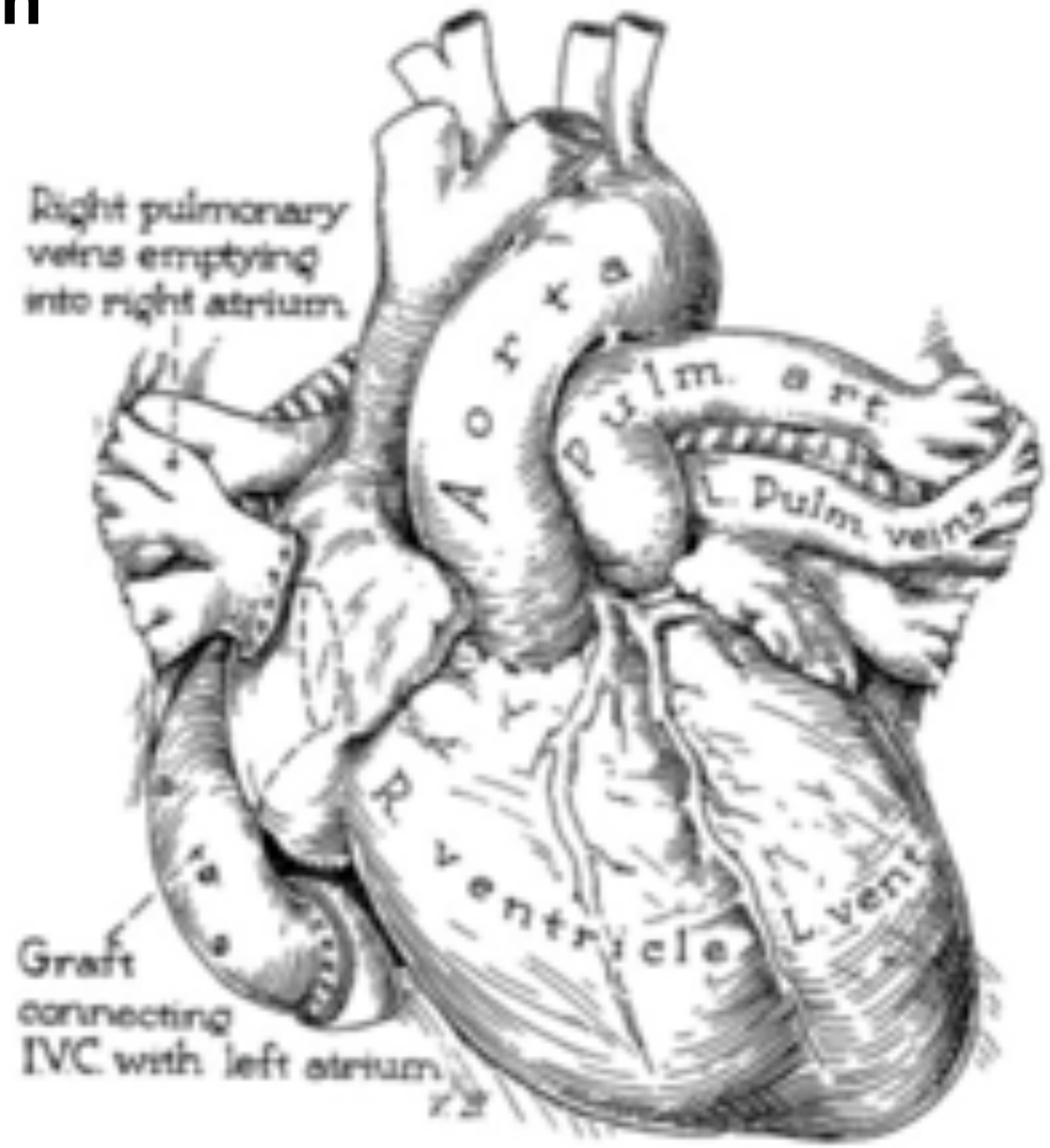


Kasmi L et al. J Thorac Cardiovasc Surg 2017; Ann Thorac Surg 2017

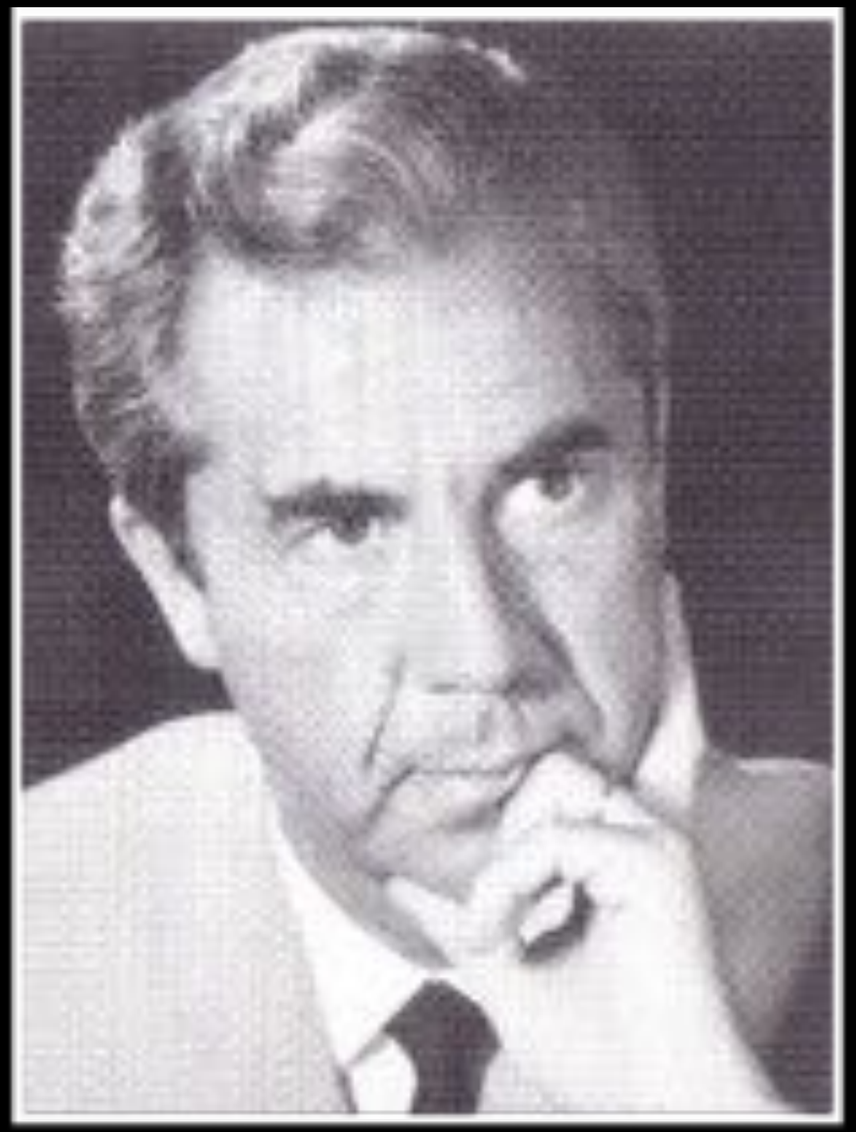
Atrial repair of TGA

The Senning and Mustard operations

Baffes operation



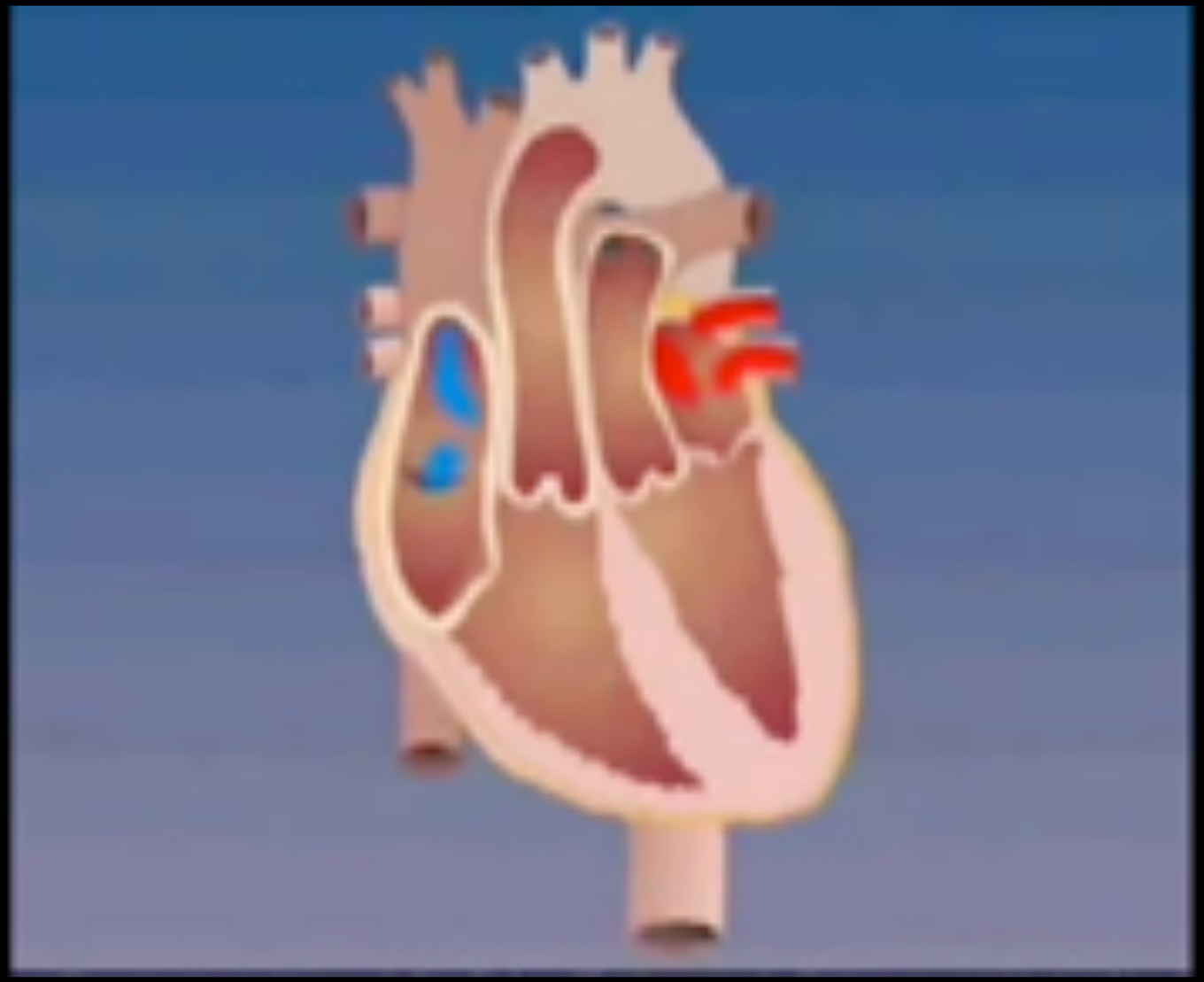
Atrial correction of TGA



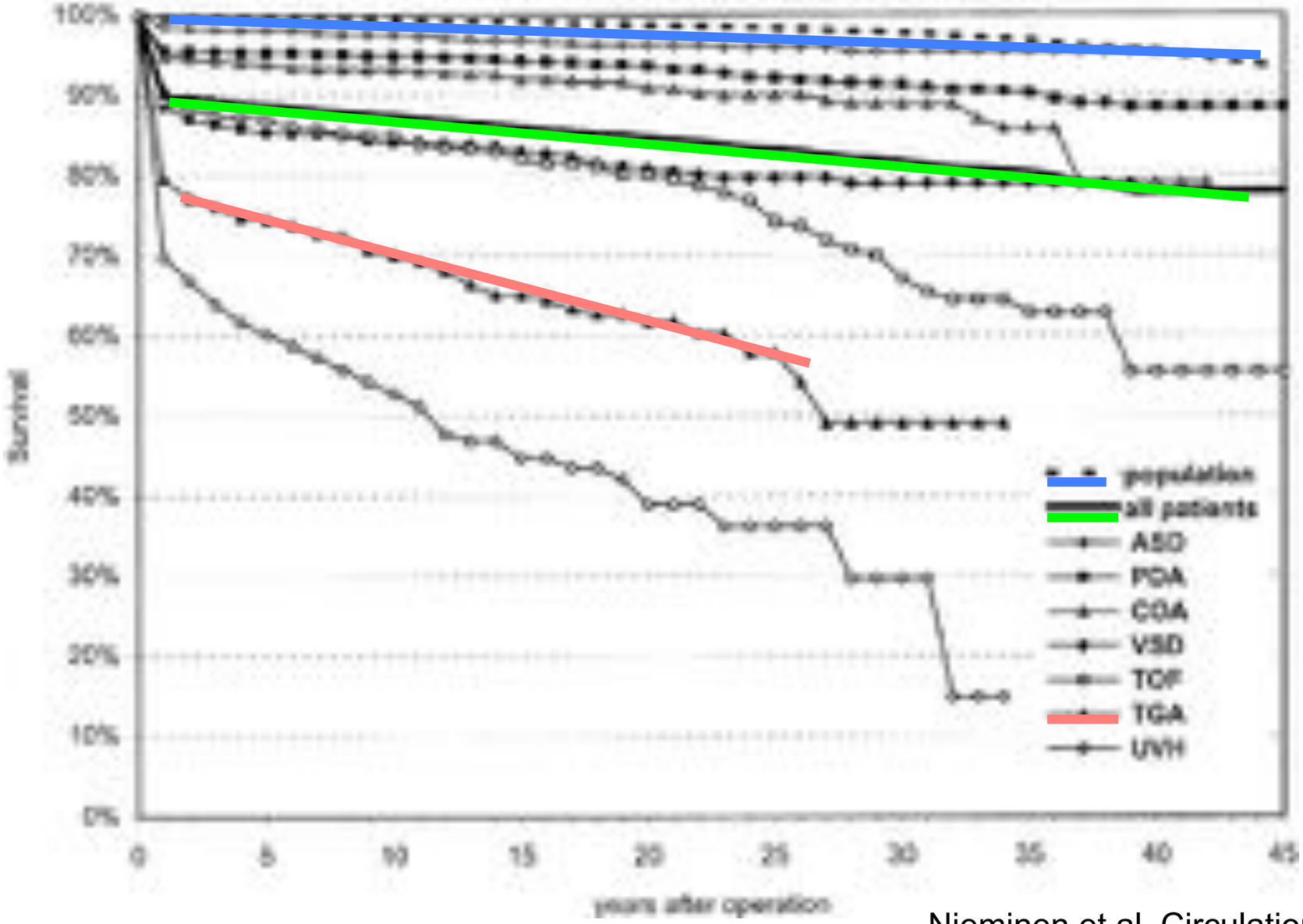
Ake Senning

William Thornton Mustard

Mustard operation

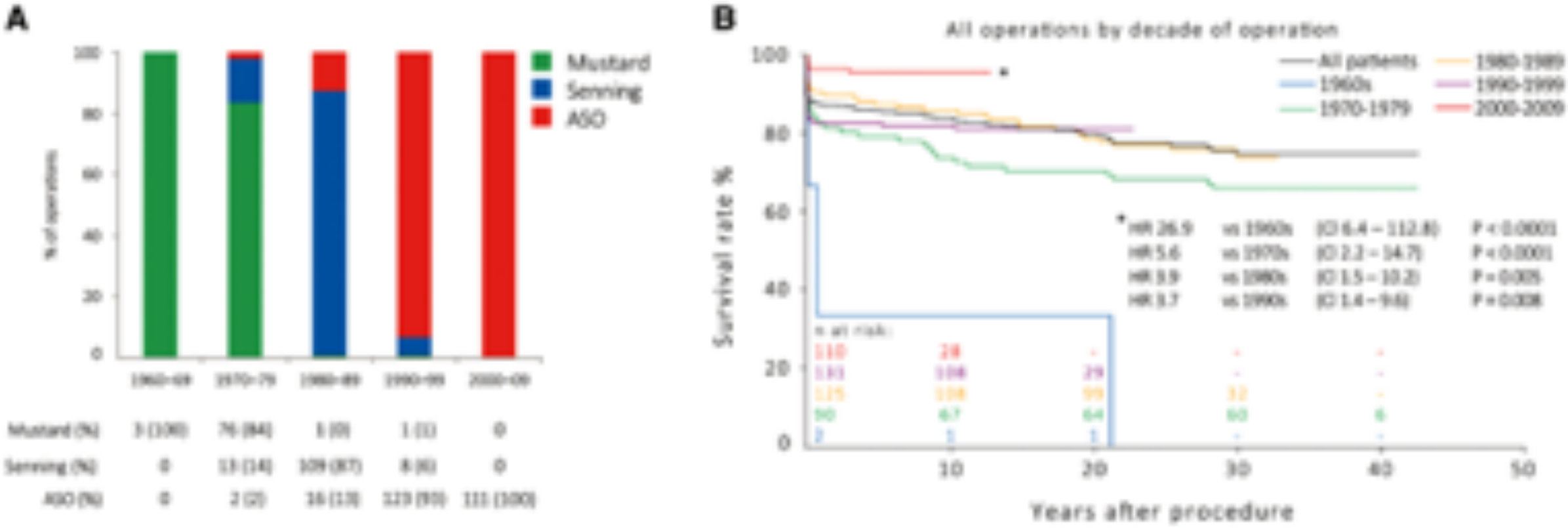


Survival of all patients, separate diagnostic groups, and general population



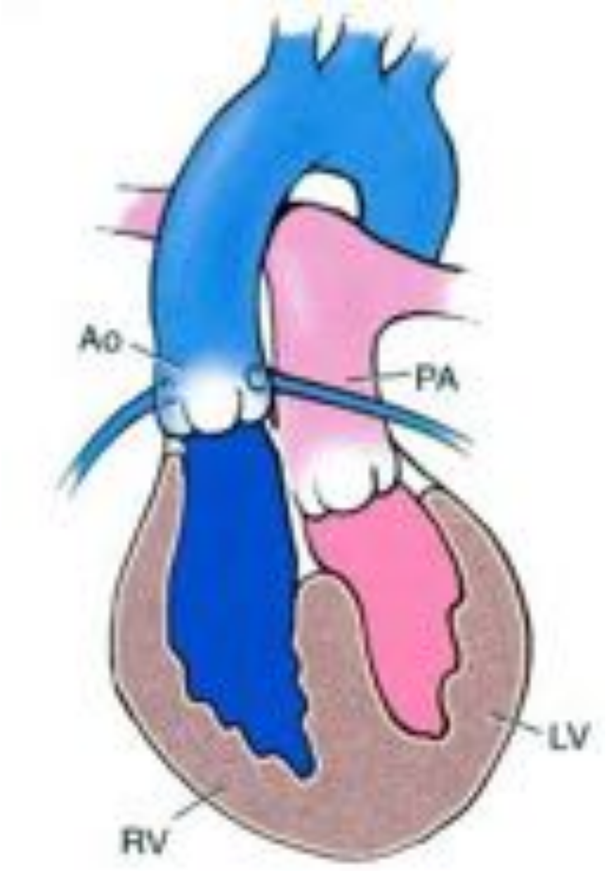
Nieminen et al, Circulation 2001

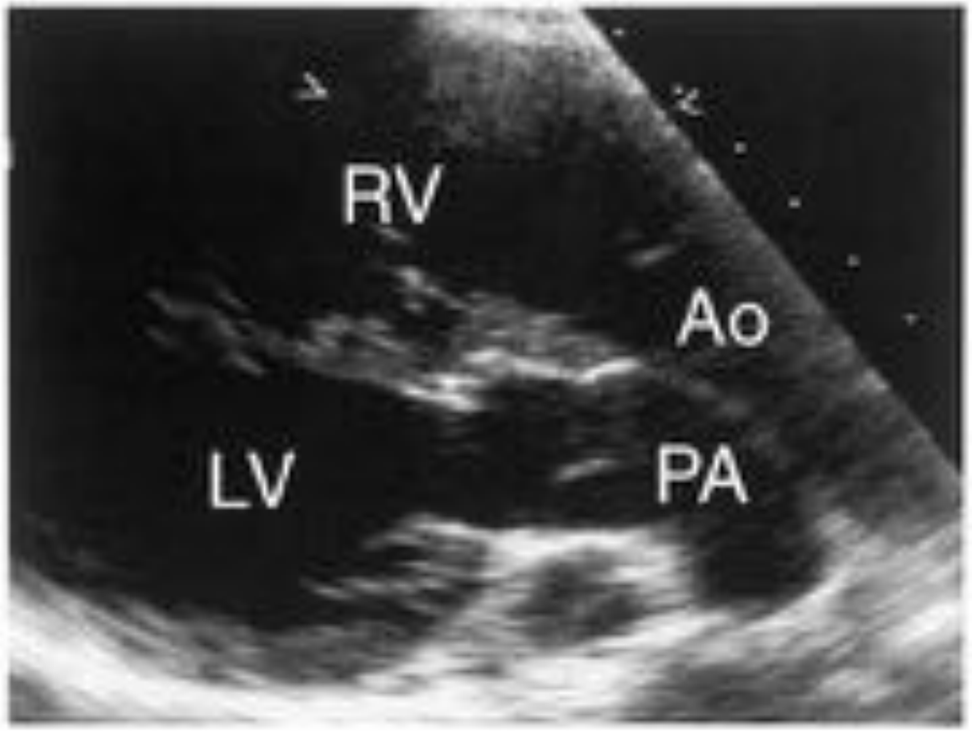
Outcomes after the Mustard, Senning and arterial switch operation for treatment of transposition of the great arteries in Finland: a nationwide 4-decade perspective



Eur J Cardiothorac Surg. 2017;52(3):573-580.

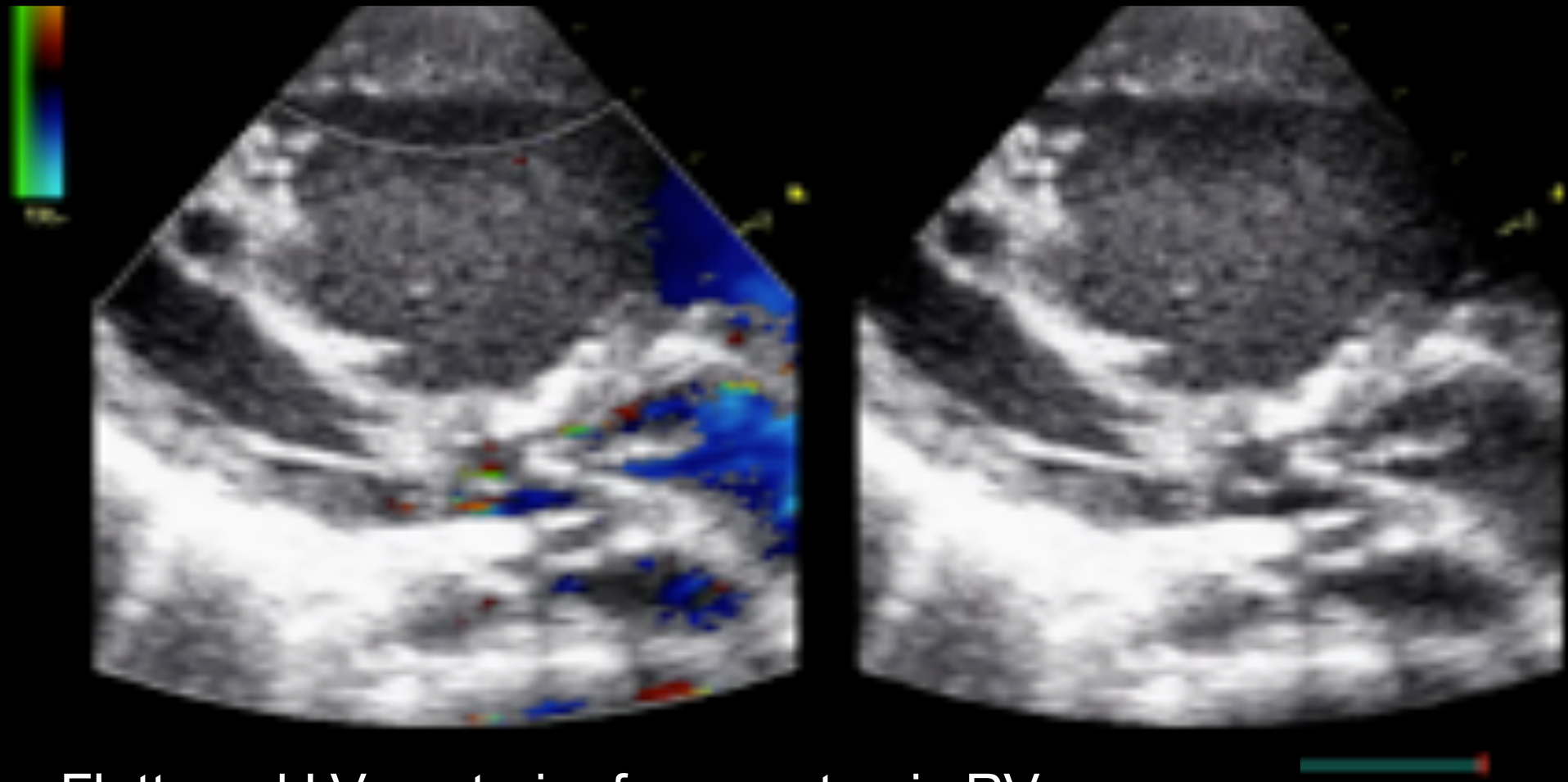
Echocardiogram long axis view





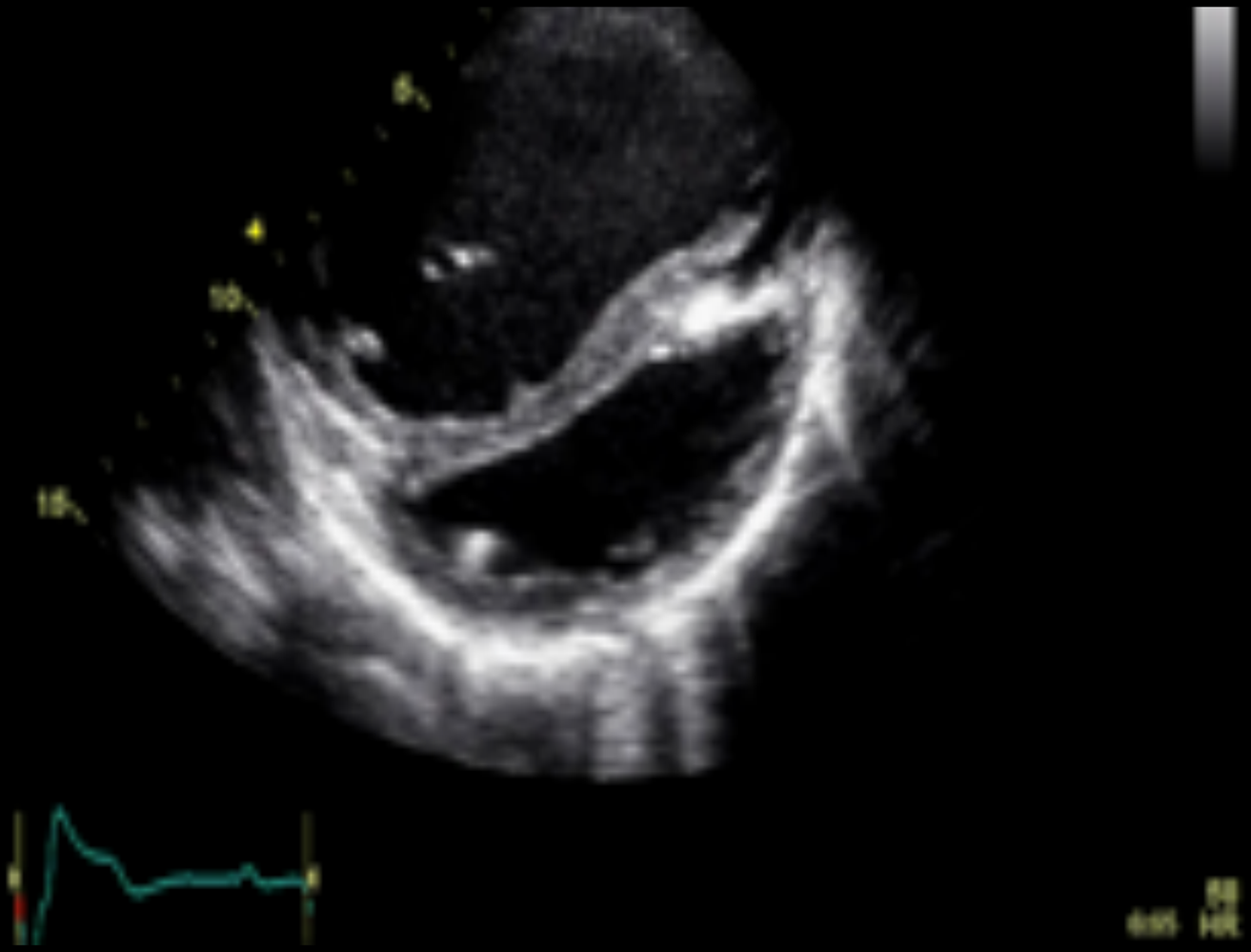
Aorta anterior, PA posterior

Long axis view after atrial correction of TGA



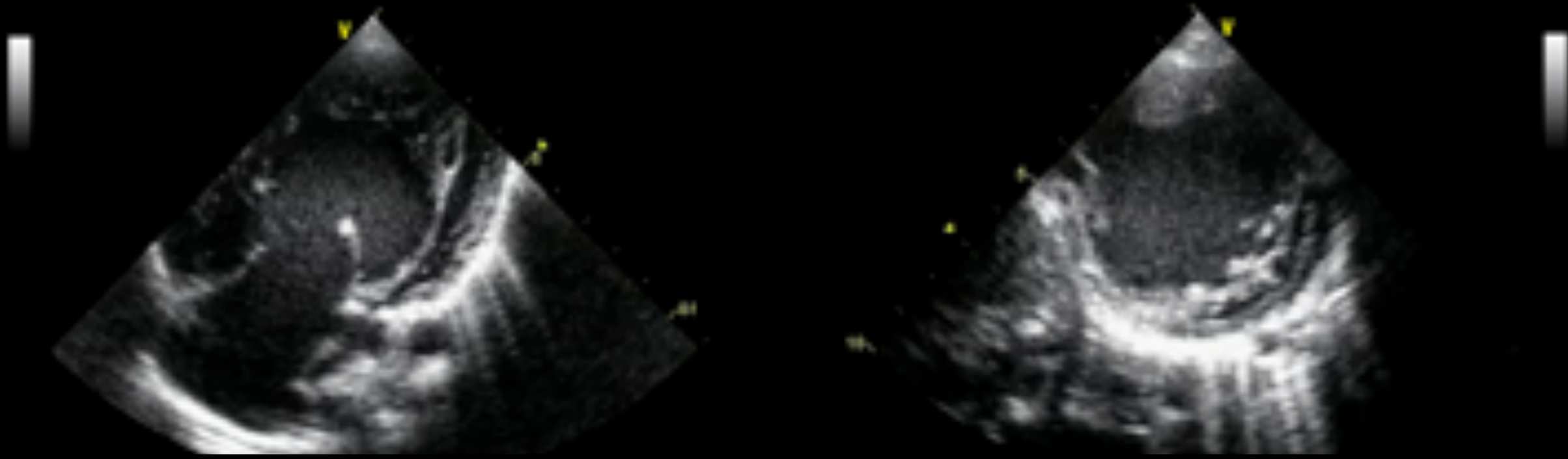
Flattened LV posterior from systemic RV

Short axis view after atrial correction of TGA

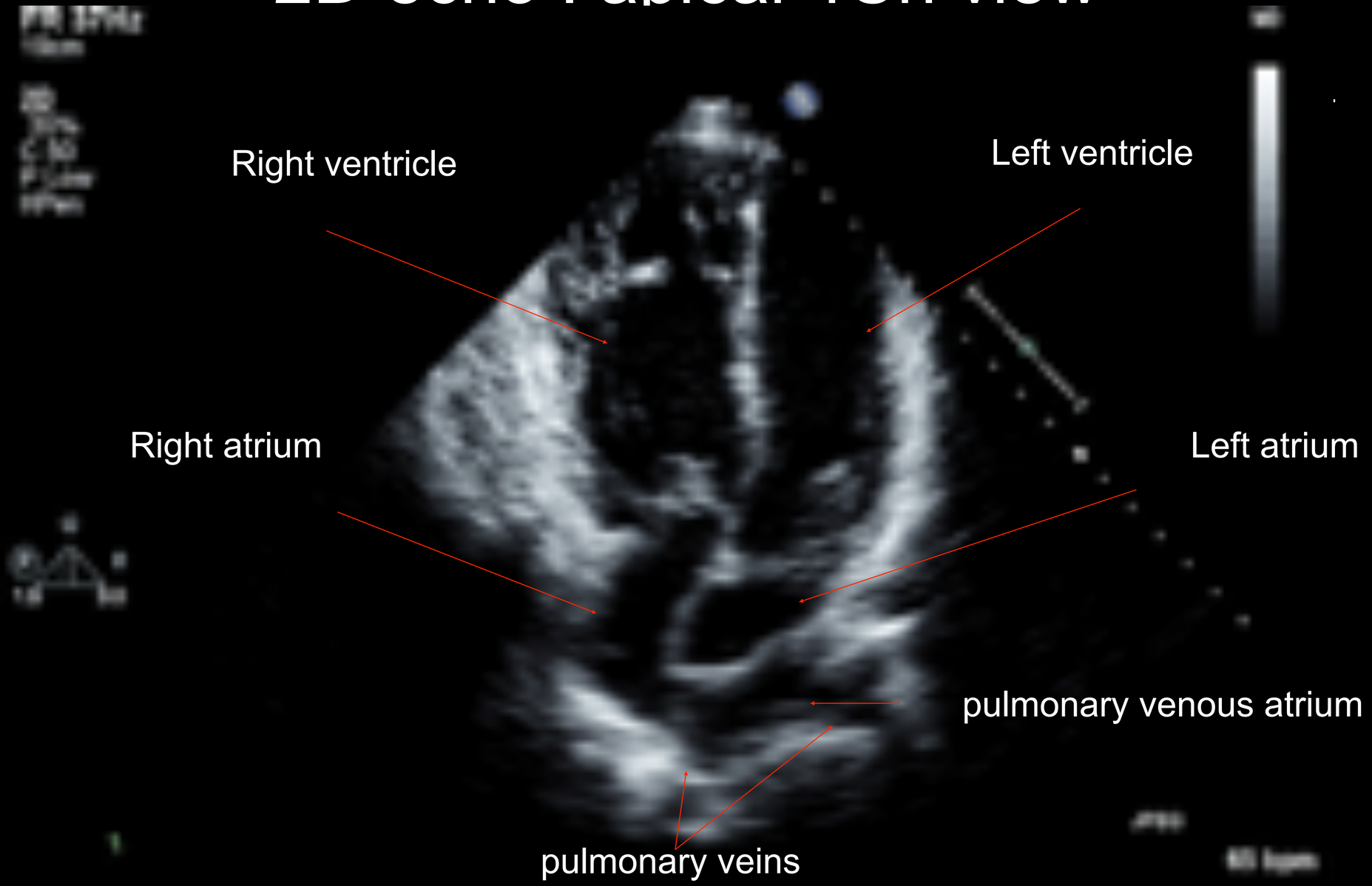


Flattened LV posterior from systemic RV

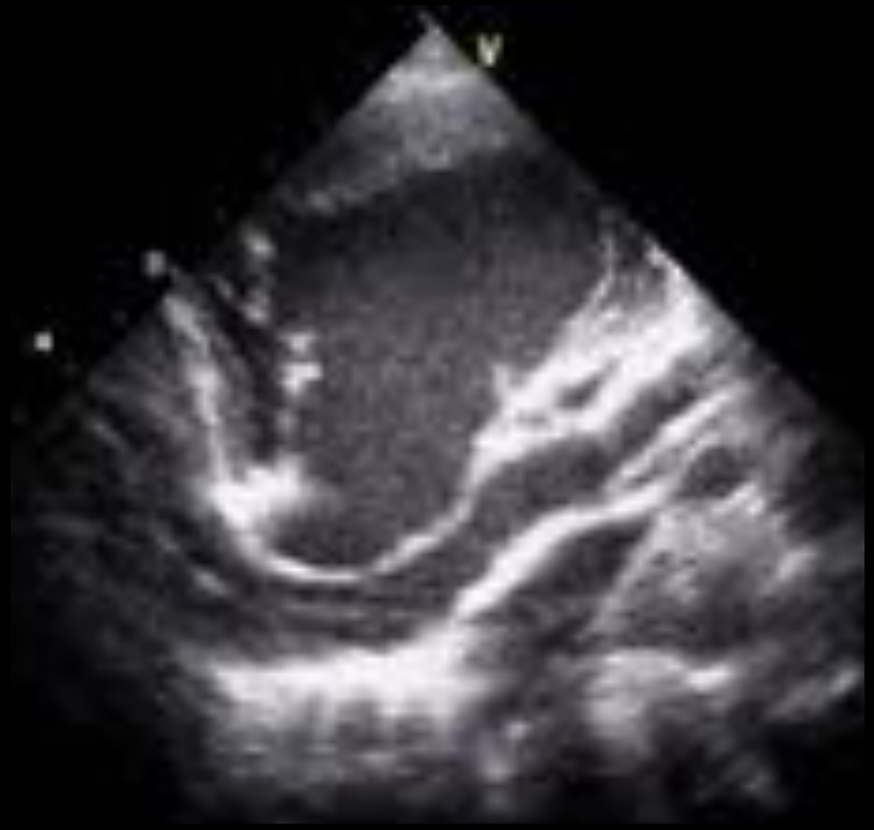
Right ventricle after atrial correction of TGA



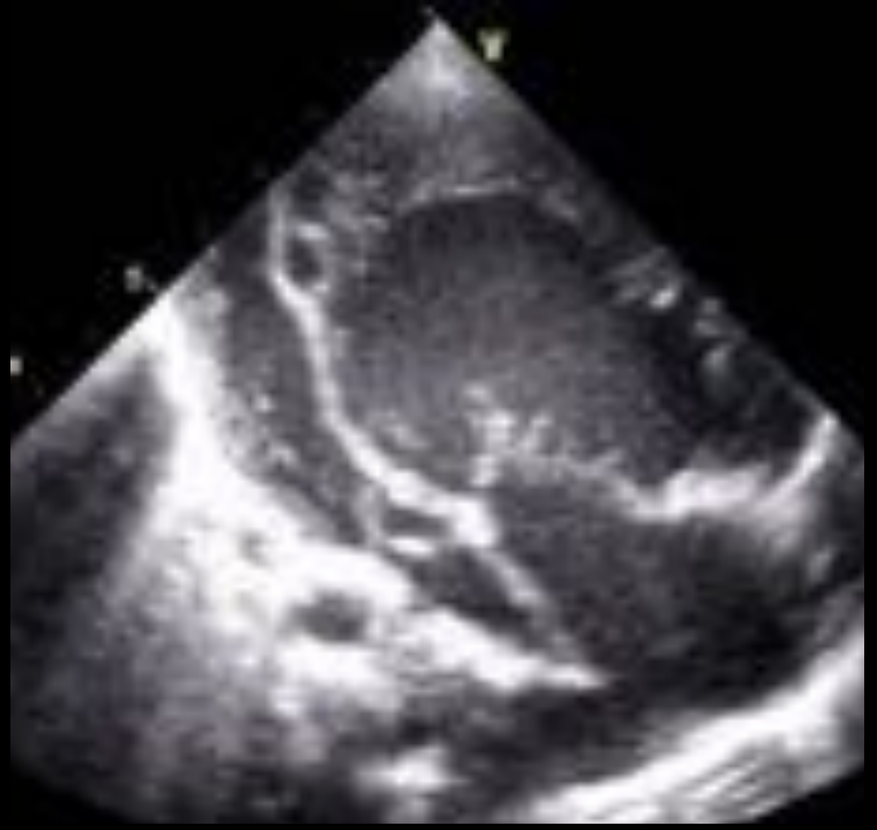
2D echo : apical 4Ch view



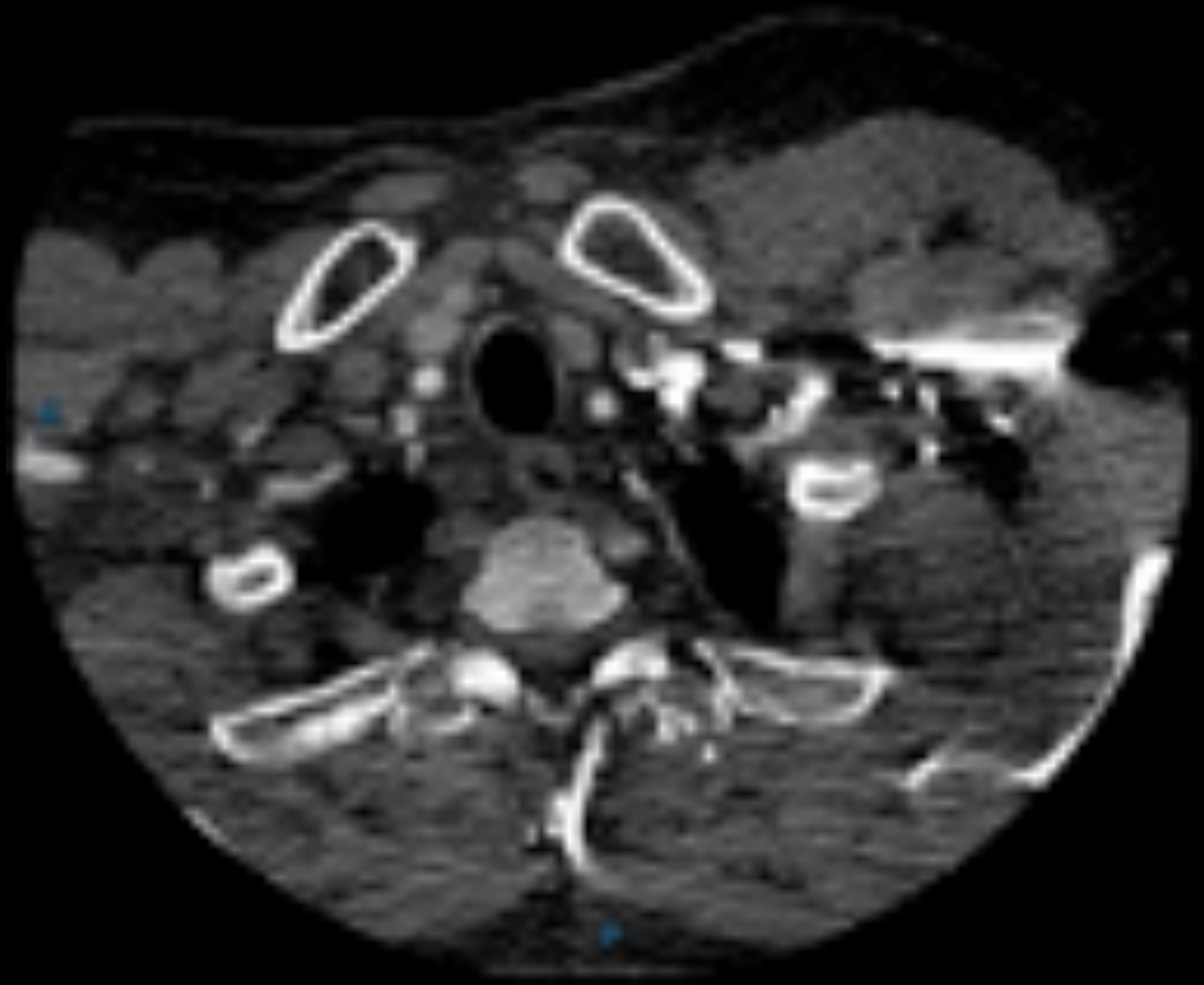
Caval baffle



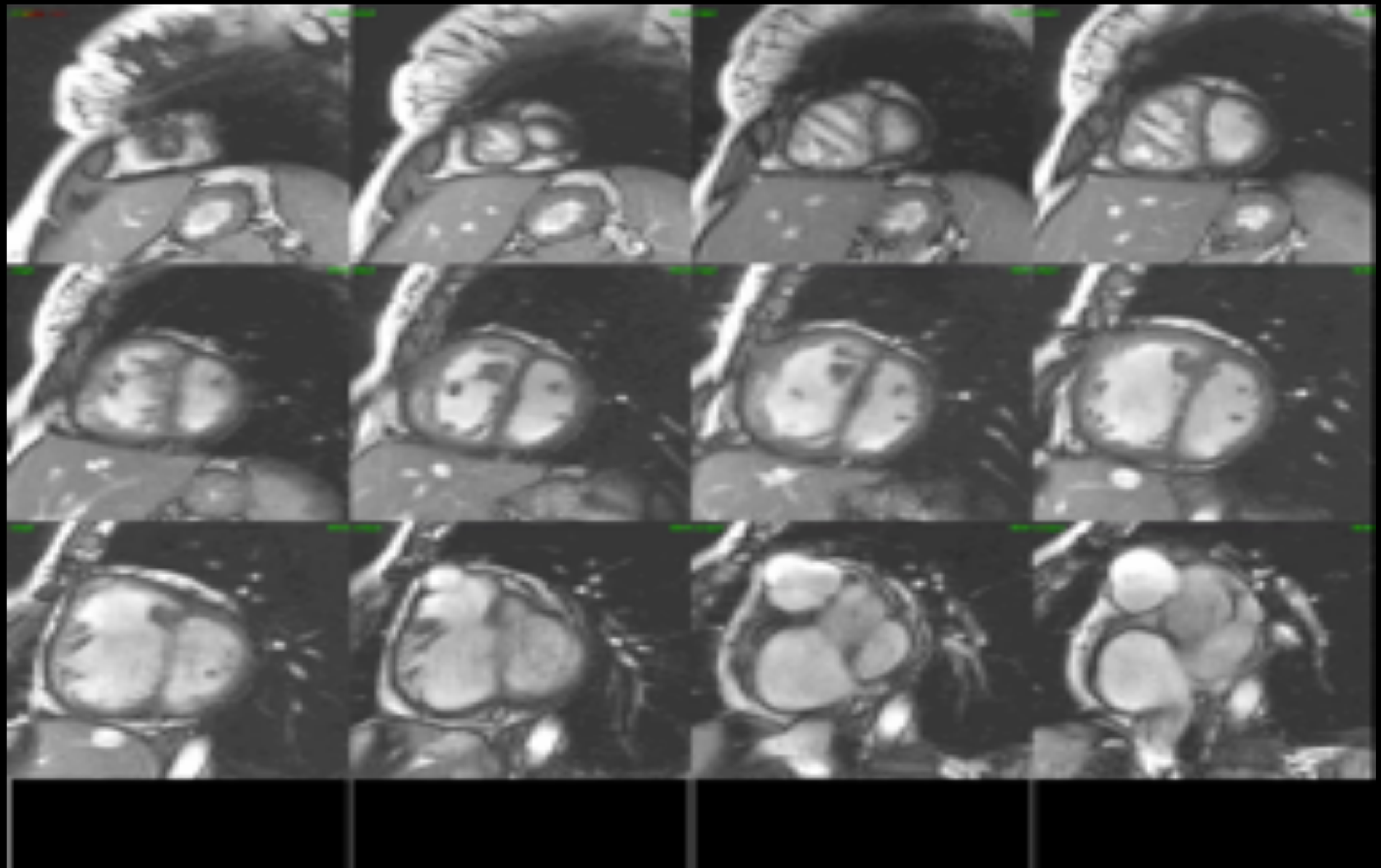
Pulmonary veins baffle



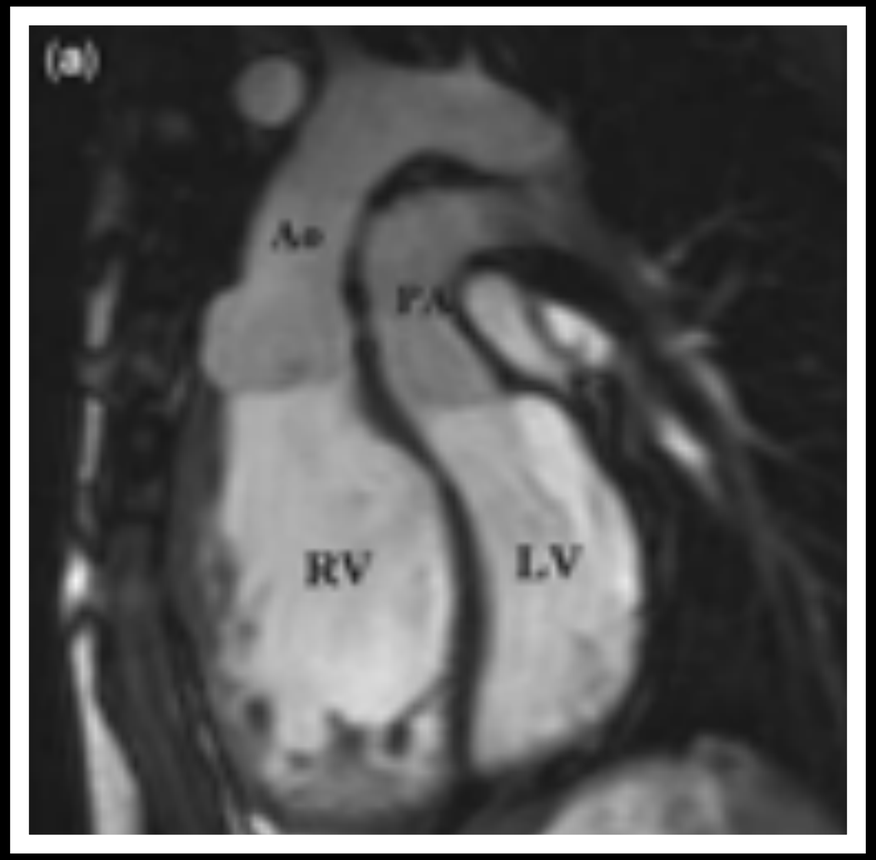
CT Scanner after Senning operation for TGA

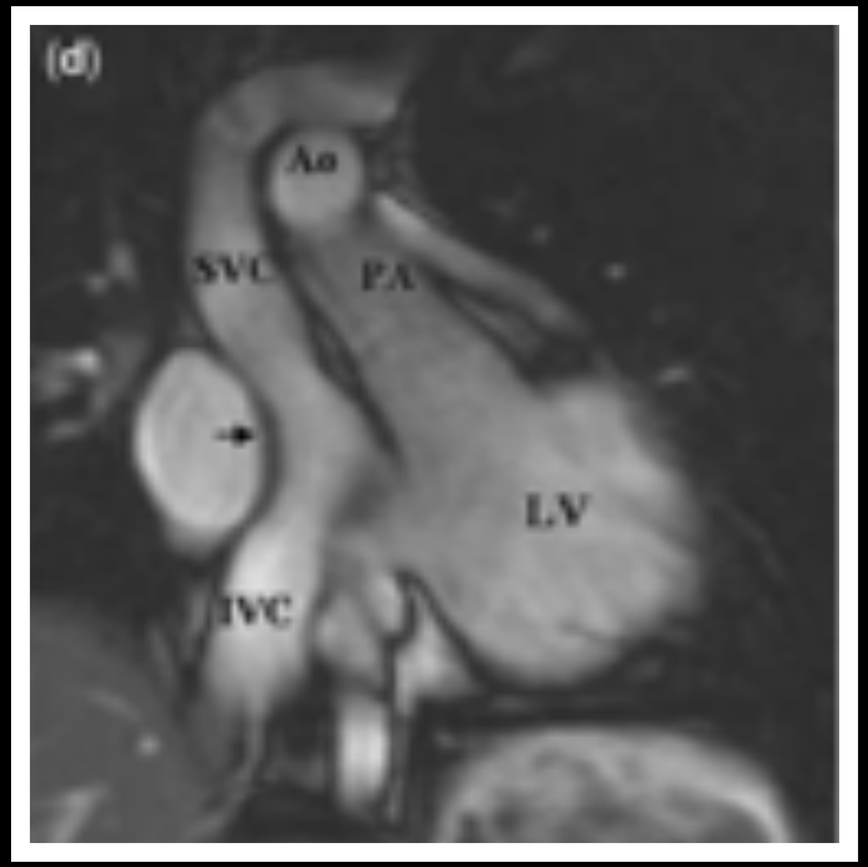


MRI senning



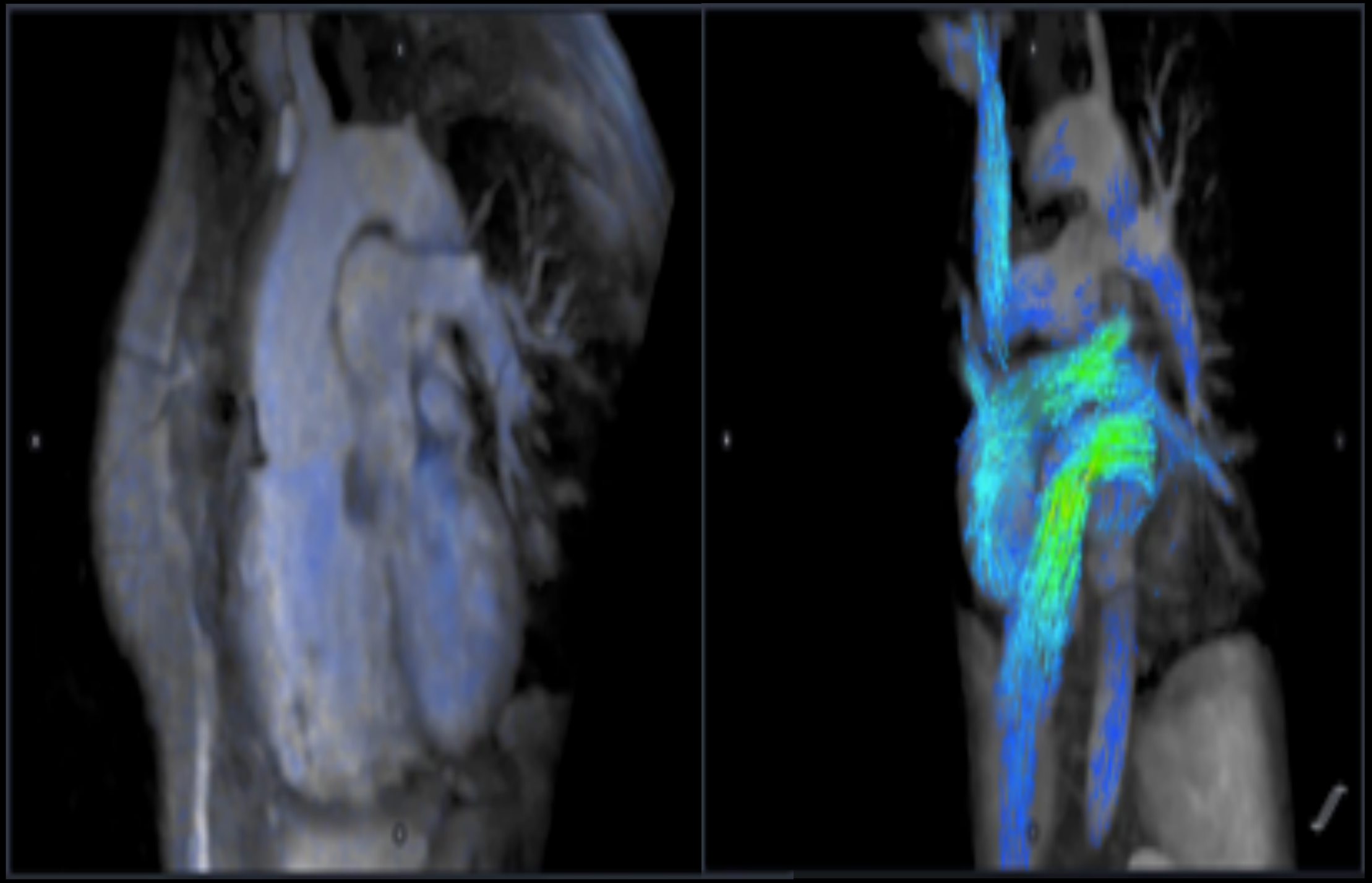
Arterial and atrial pathways in Mustard





Johansson et al, Cardiol Young, 2010

MRI senning

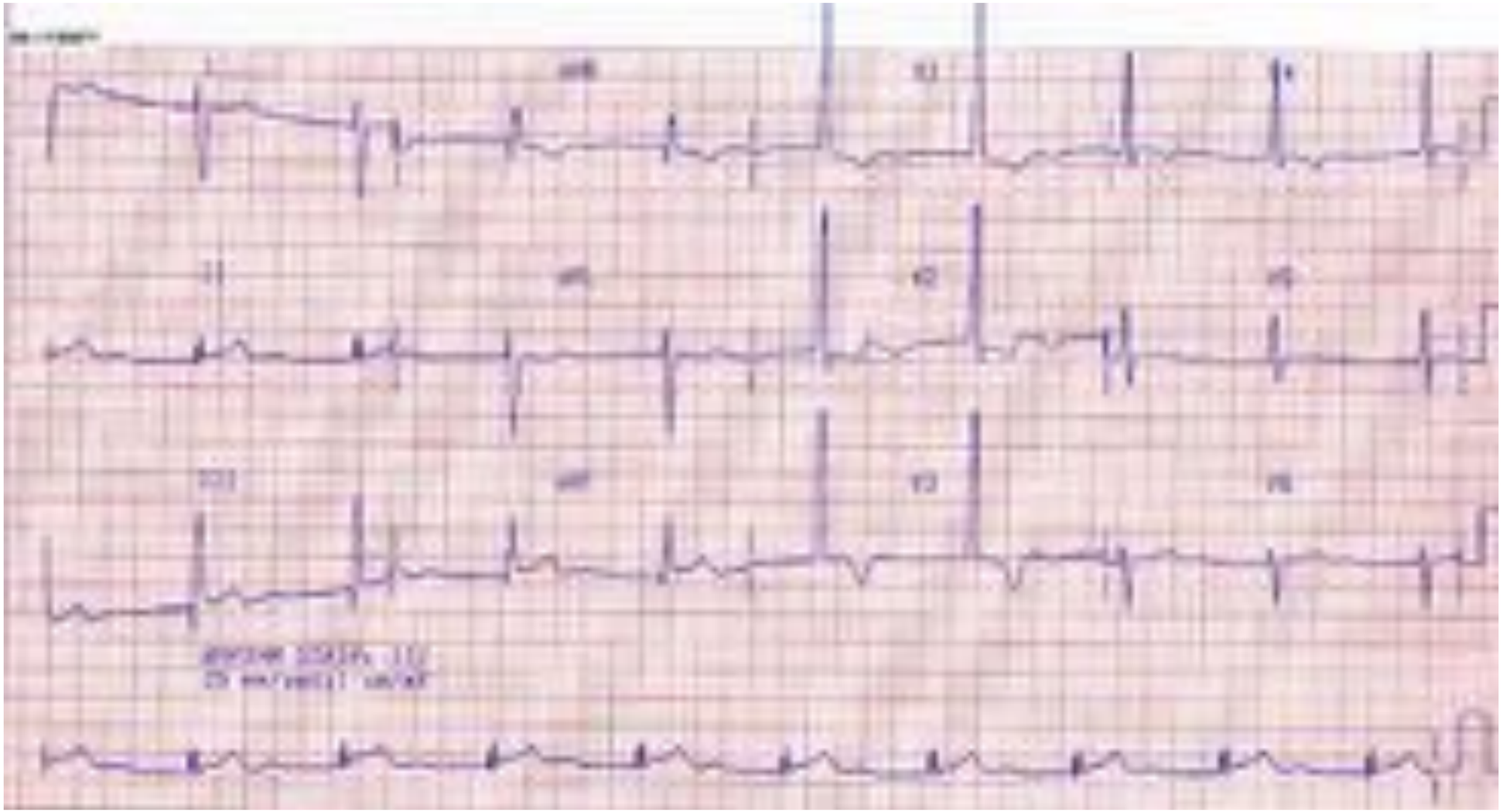


Mortality after atrial switch

- Late yearly mortality 0.5%, due to arrhythmias and heart failure
- Sudden death (42%) most common mode of death
- Independent predictors for mortality:
  - (Atrial)Tachyarrhythmias
  - Advanced functional class

Kammeraad, et al, JACC 2004

ECG after atrial switch



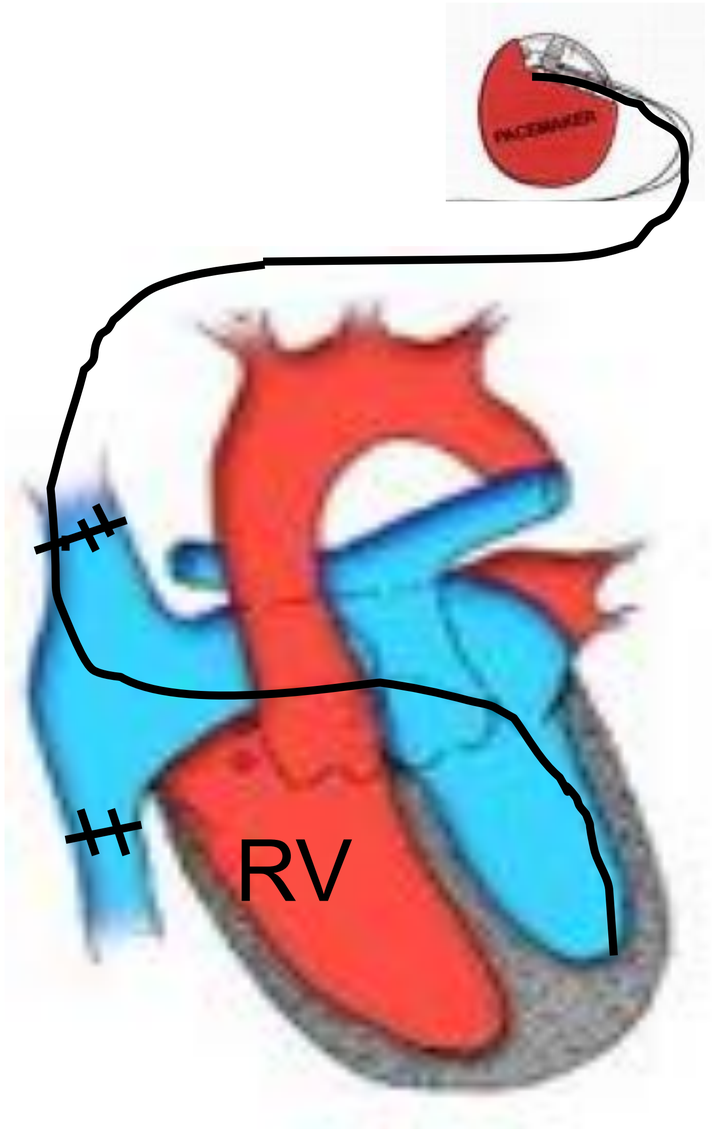
RVH

Right heart axis

Junctional rhythm

Only 40% of patients has SR at age 20

20% of young adults with atrial switch needs PM for sick sinus syndrome



Be aware of altered venous connection:

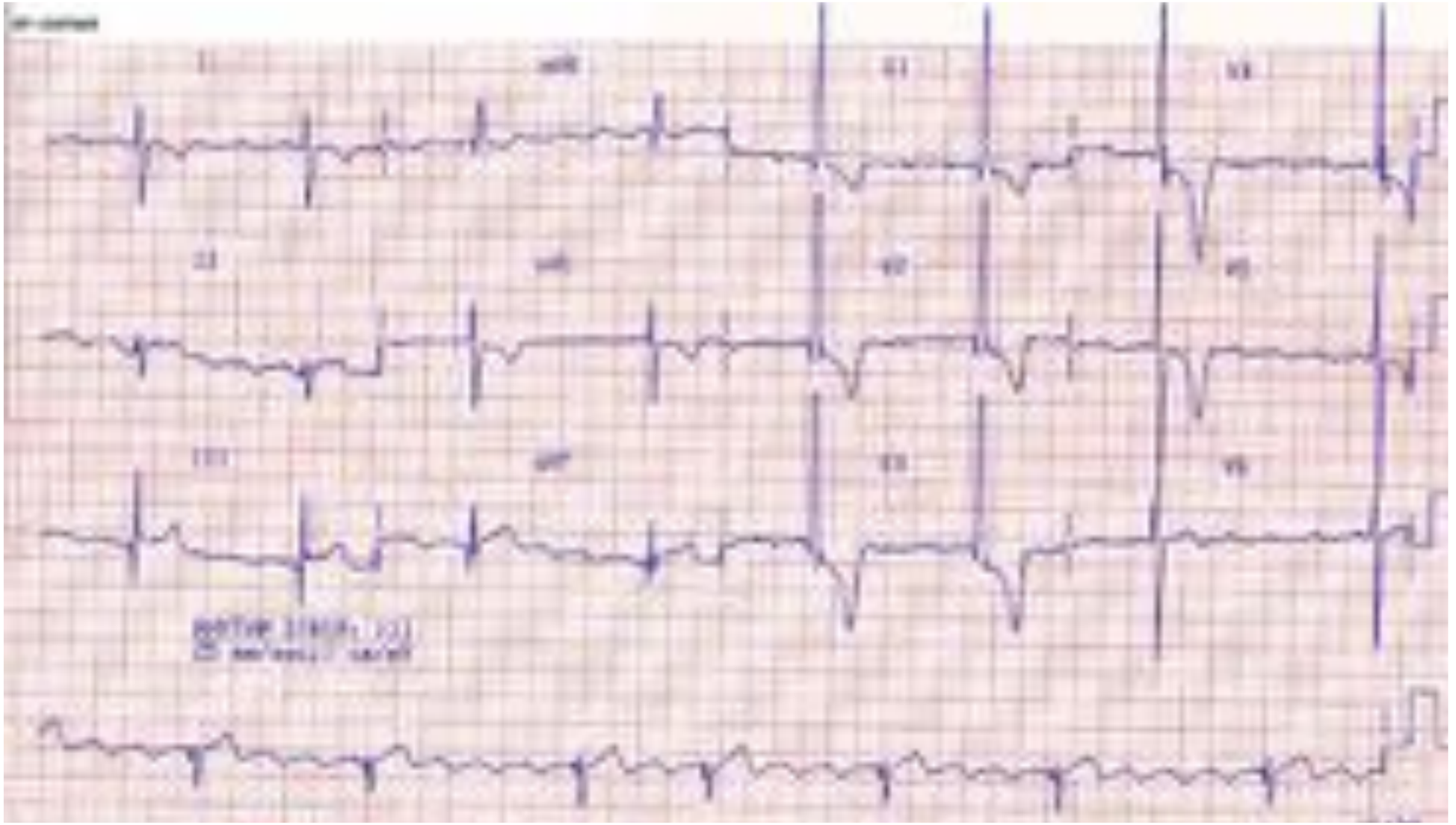
Ventricular lead will end up,

after some unusual loops,

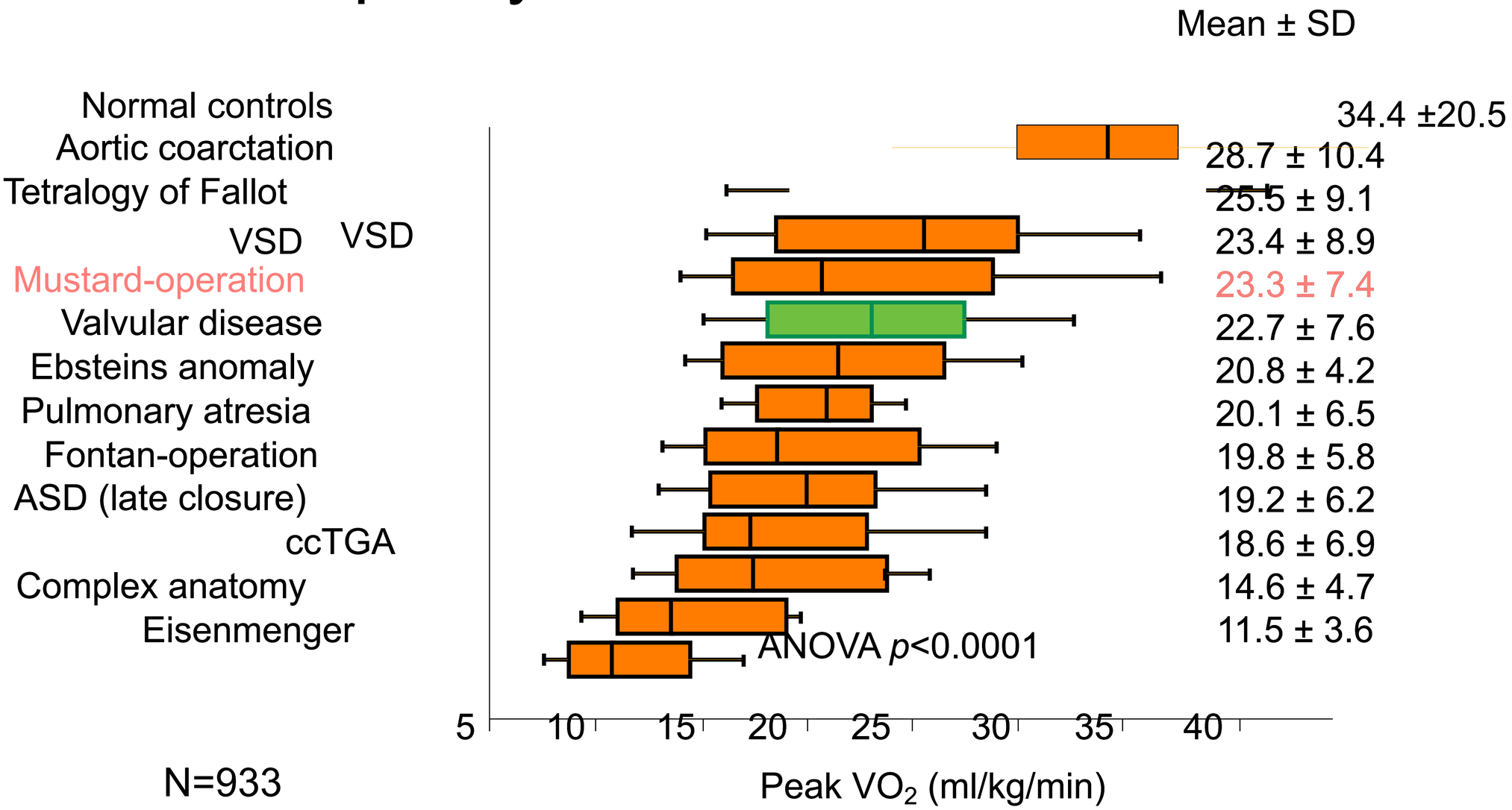
in a smooth-walled LV

Atrial flutter after atrial switch

Atrial arrhythmias may lead to hemodynamic instability and sudden death

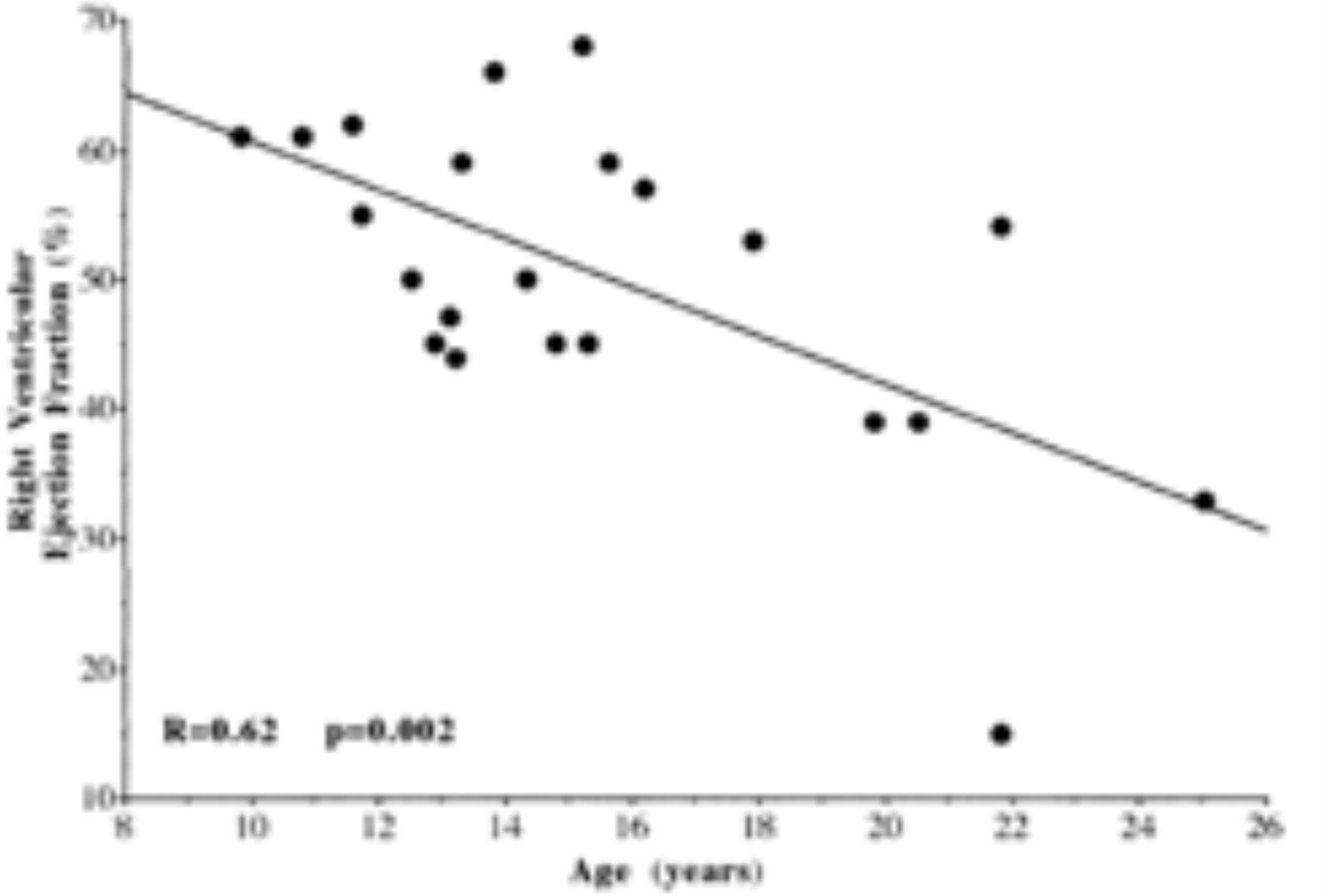


Exercise capacity in CHD



Diller GP, et al. Circulation 2005;

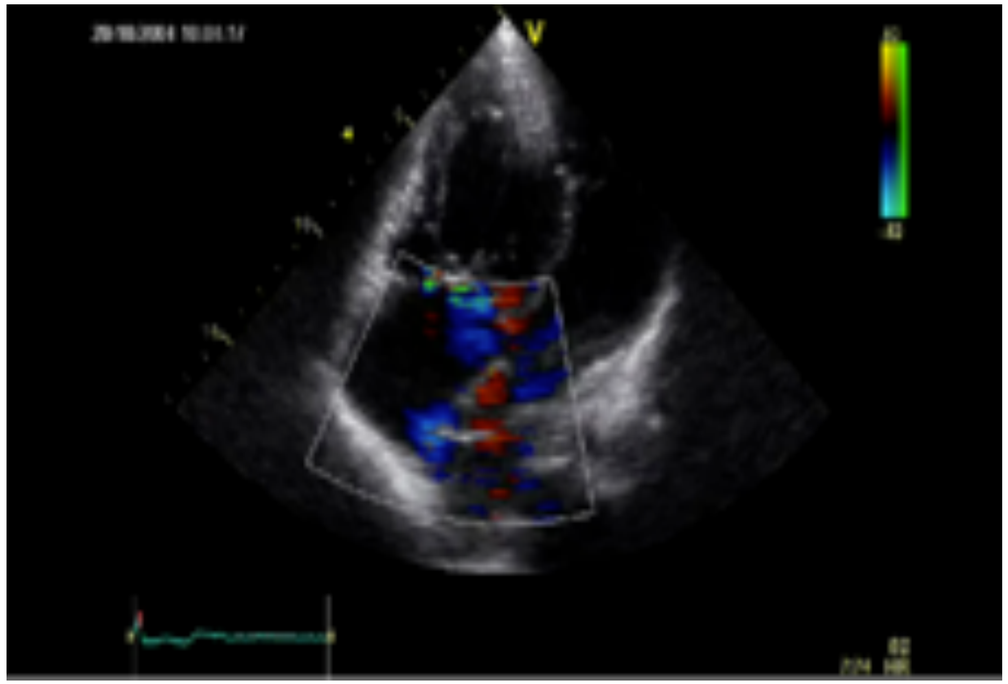
Decline of RV function after atrial switch



Milane et al. , JACC 2000.

Late complications atrial switch

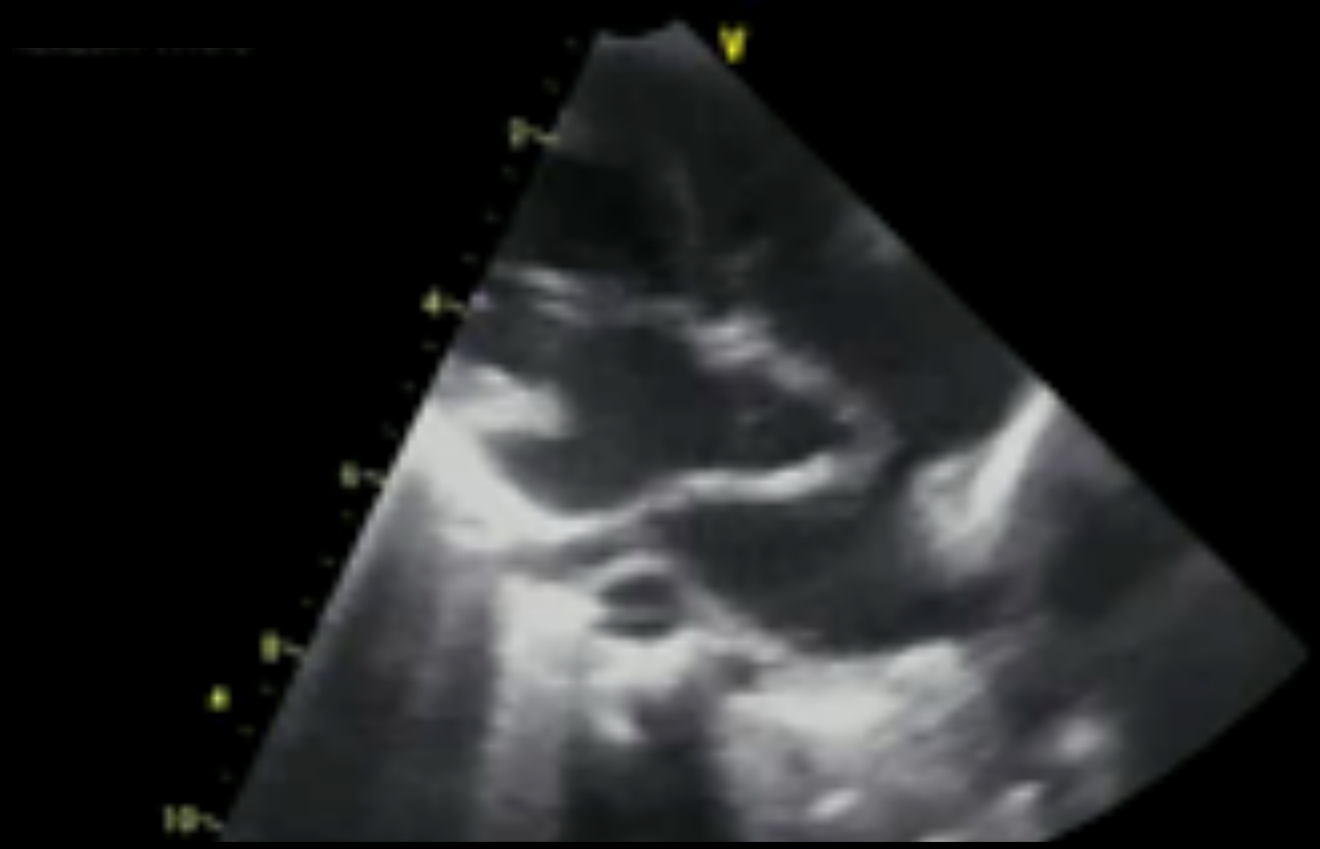
- Early death
- Arrhythmias
- Exercise capacity
- RV dysfunction ⬇
- Tricuspid regurgitation

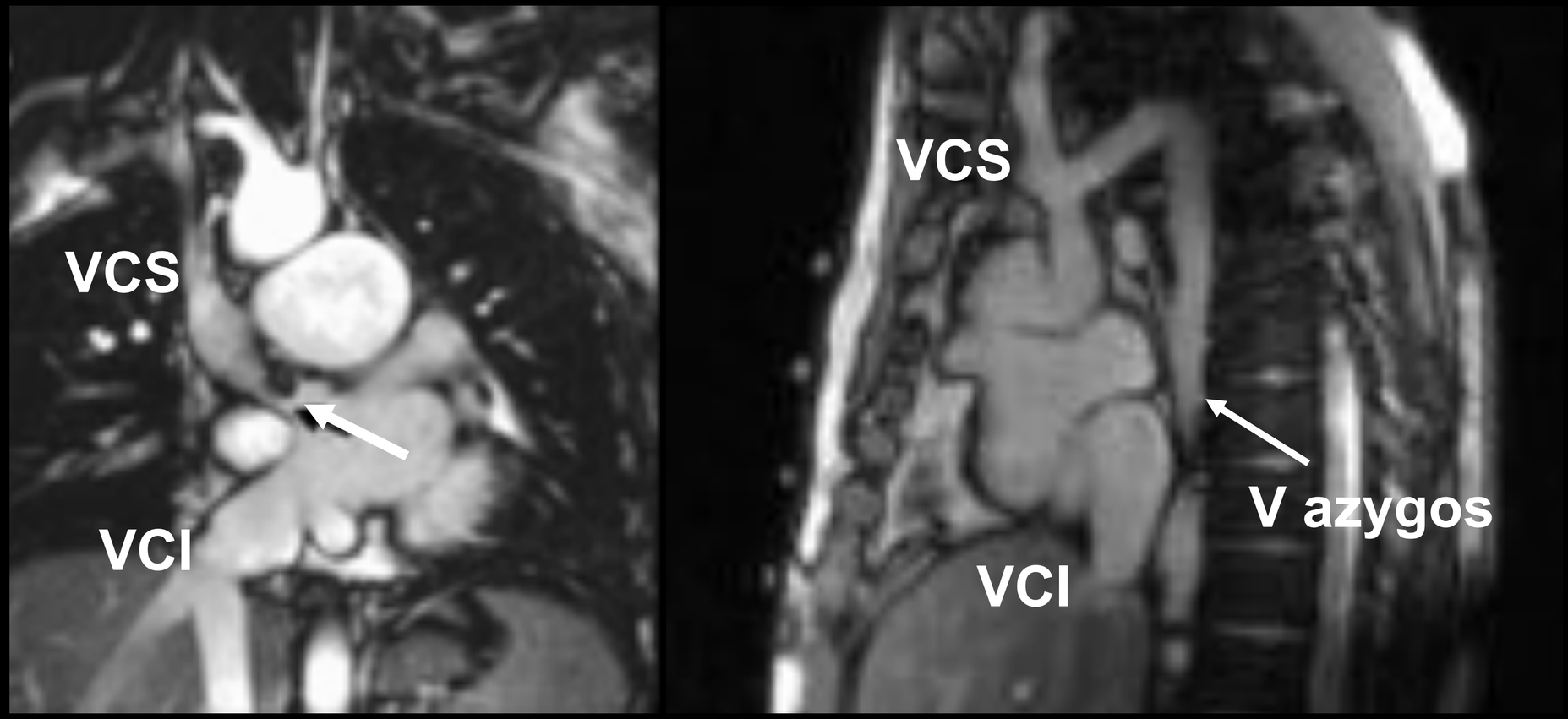


Abnormal septal configuration
RV dilation

Late complications atrial switch

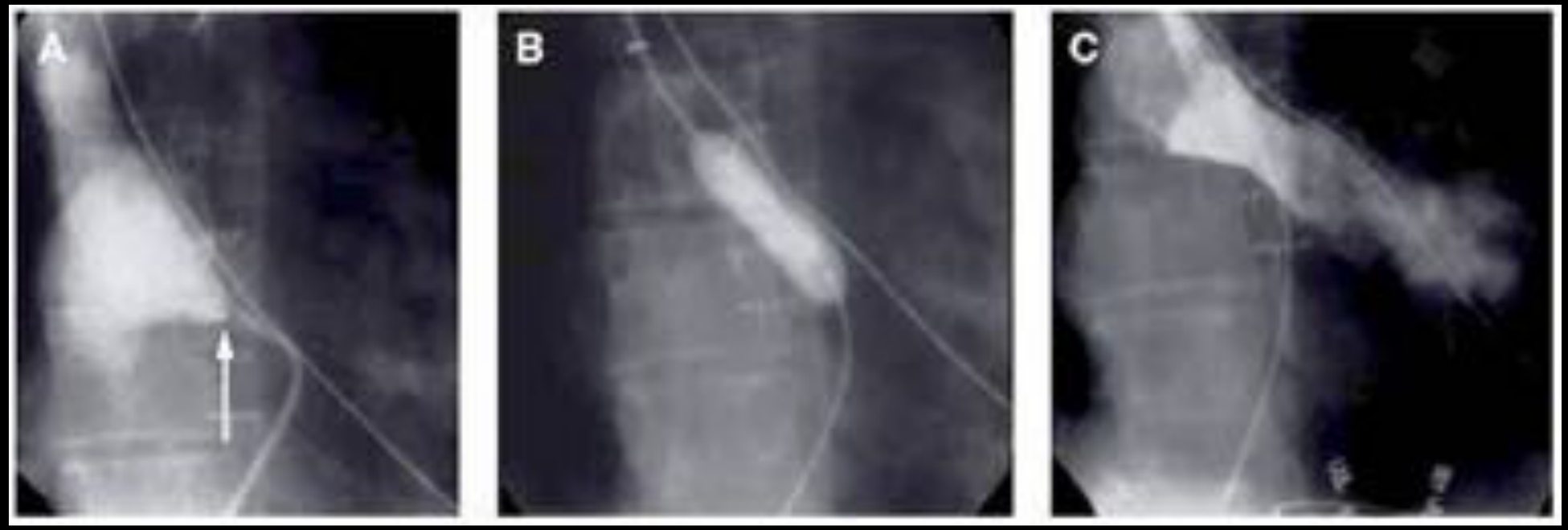
- Early death
- Arrhythmias
- Exercise capacity
- RV dysfunction
- Tricuspid regurgitation
- **Baffle obstructions**





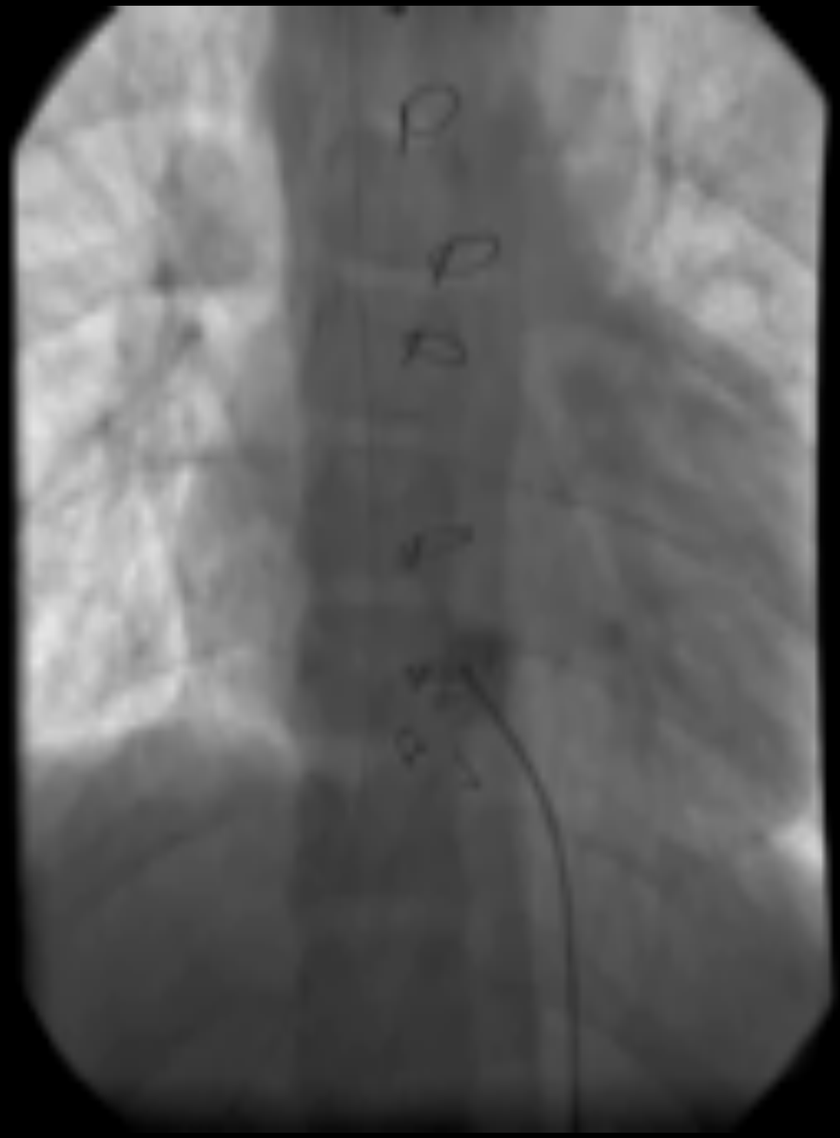
Baffle obstruction with increased azygos flow

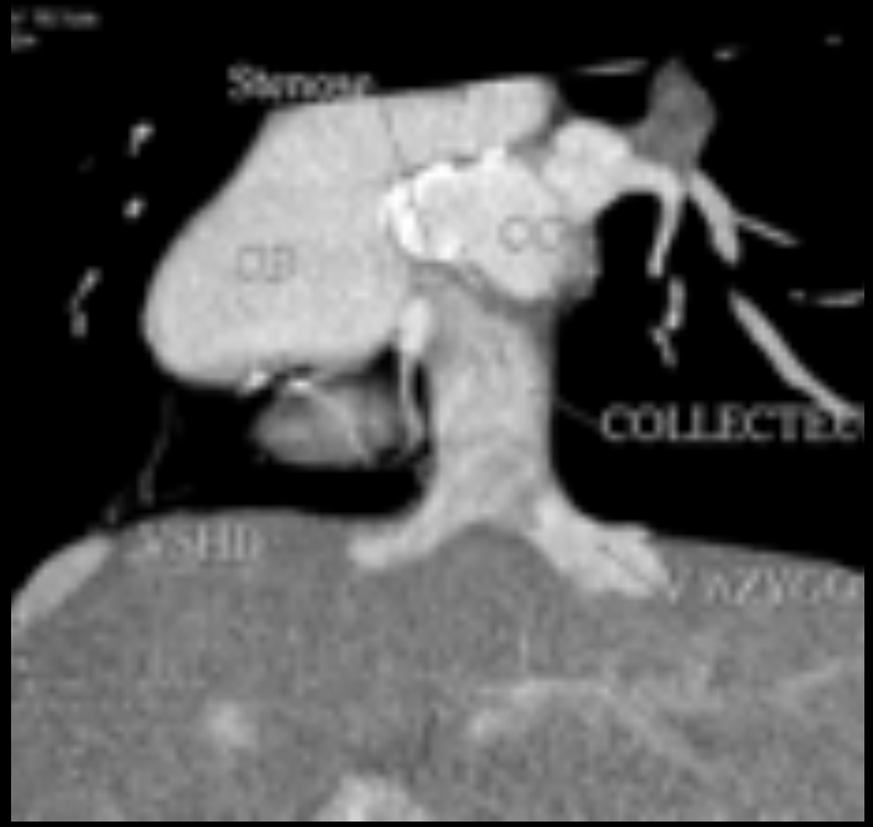
Superior baffle-limb stenosis in TGA after atrial switch and following PM implantation



obstruction Radiofrequency + Stent

Love BA et al., Nat Clin Pract Cardiovasc Med 2008

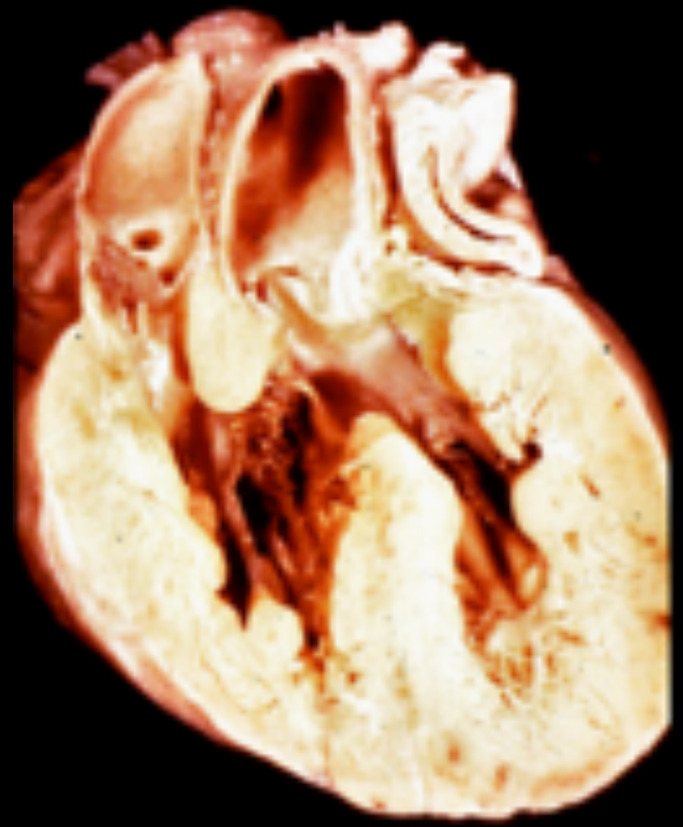



Stenose
COLLECTED

TGA-VSD and Pulmonary stenosis

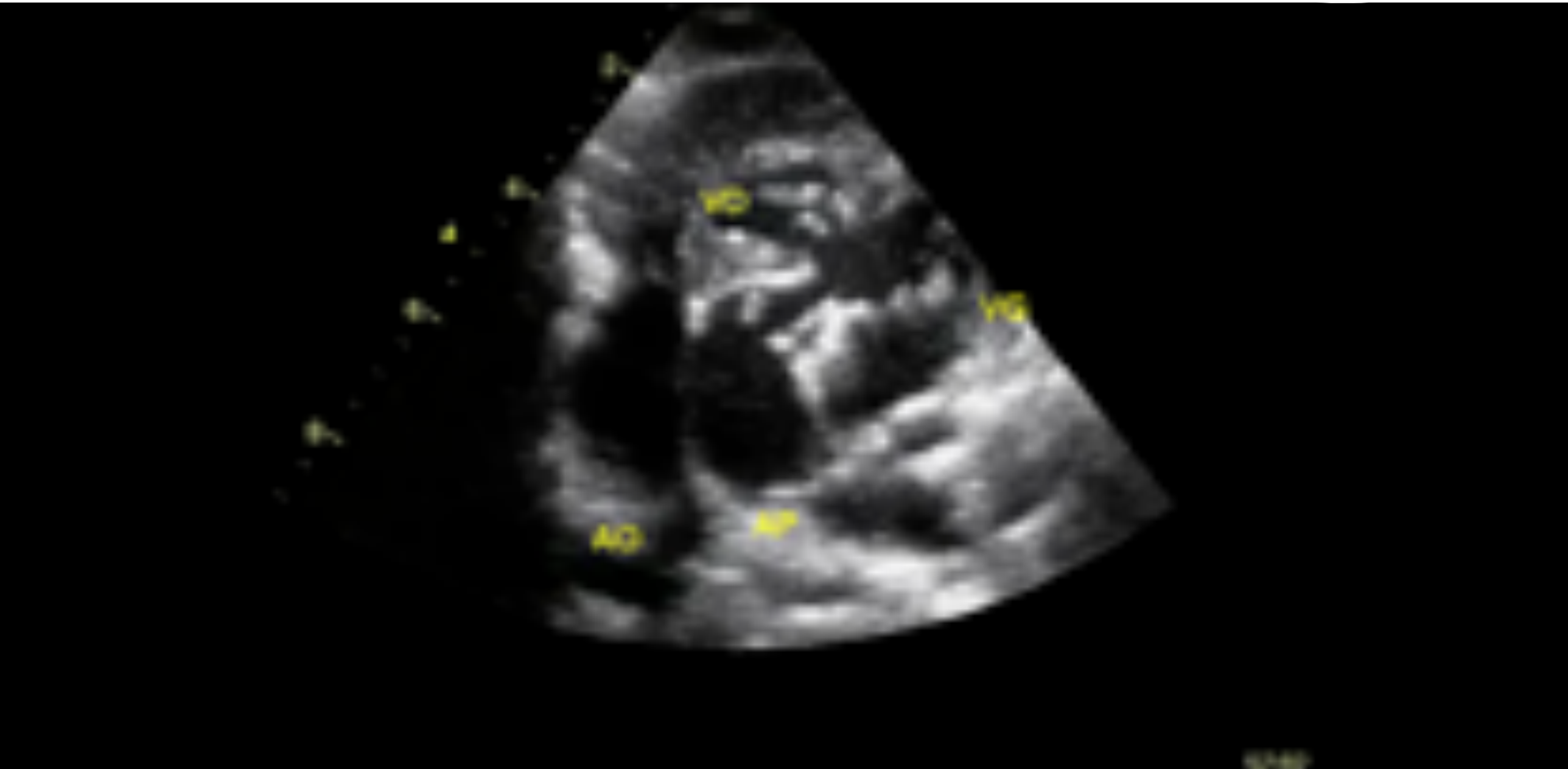
TGA

TGA - VSD - Pulmonary stenosis



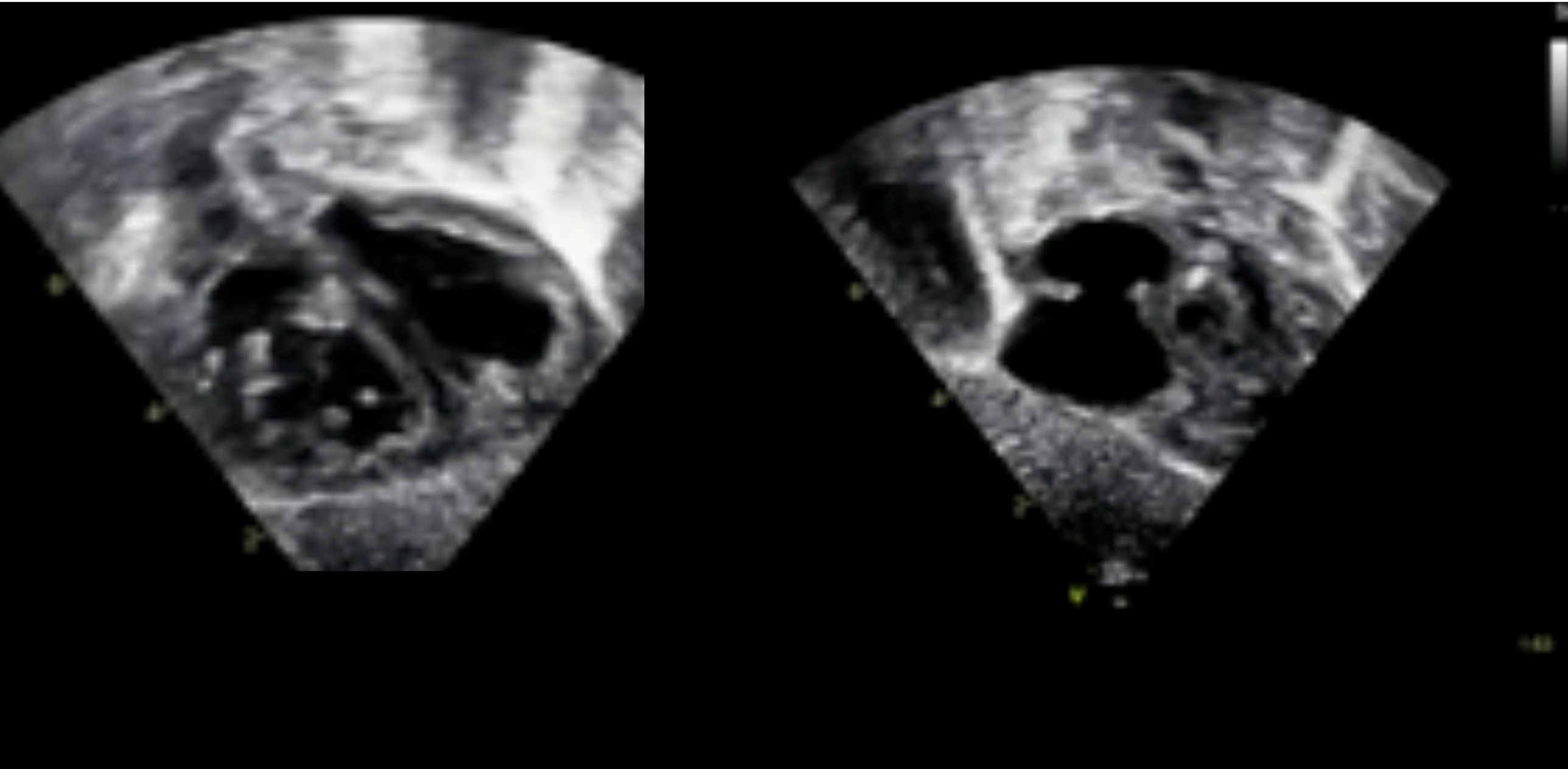
TGA

TGA - VSD - Pulmonary stenosis



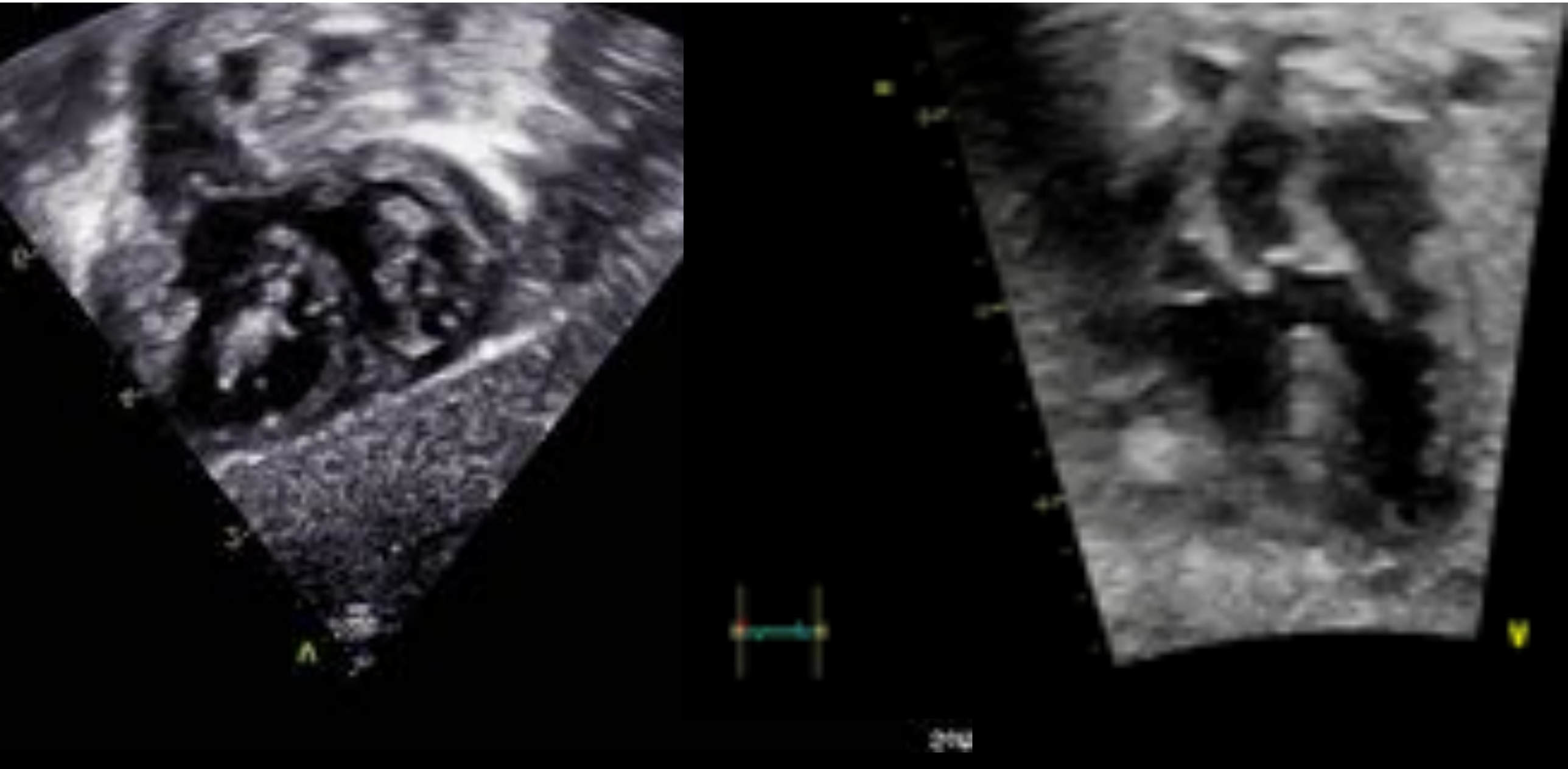
TGA

TGA - VSD - Pulmonary stenosis

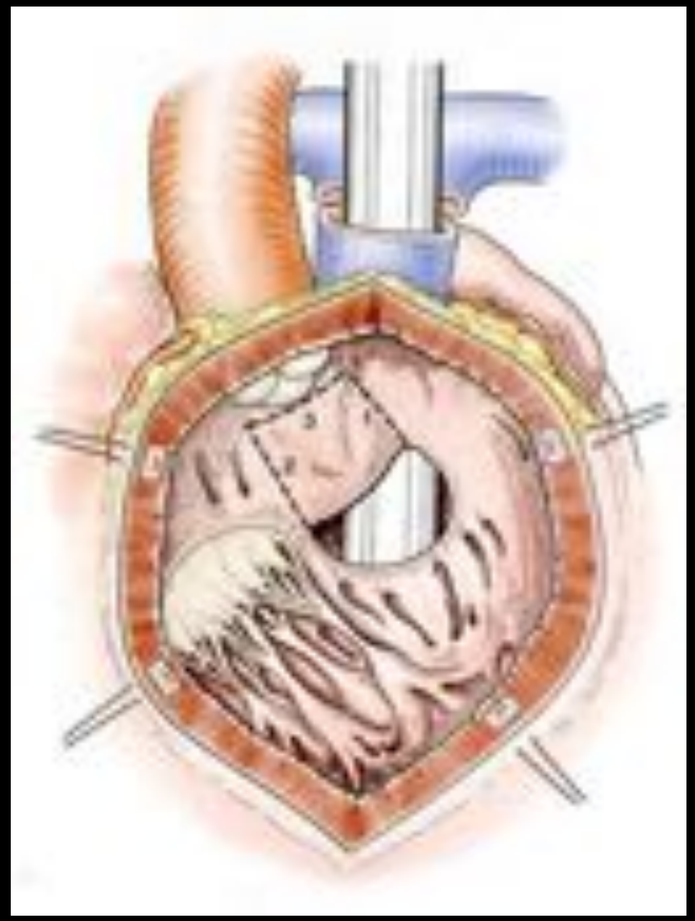


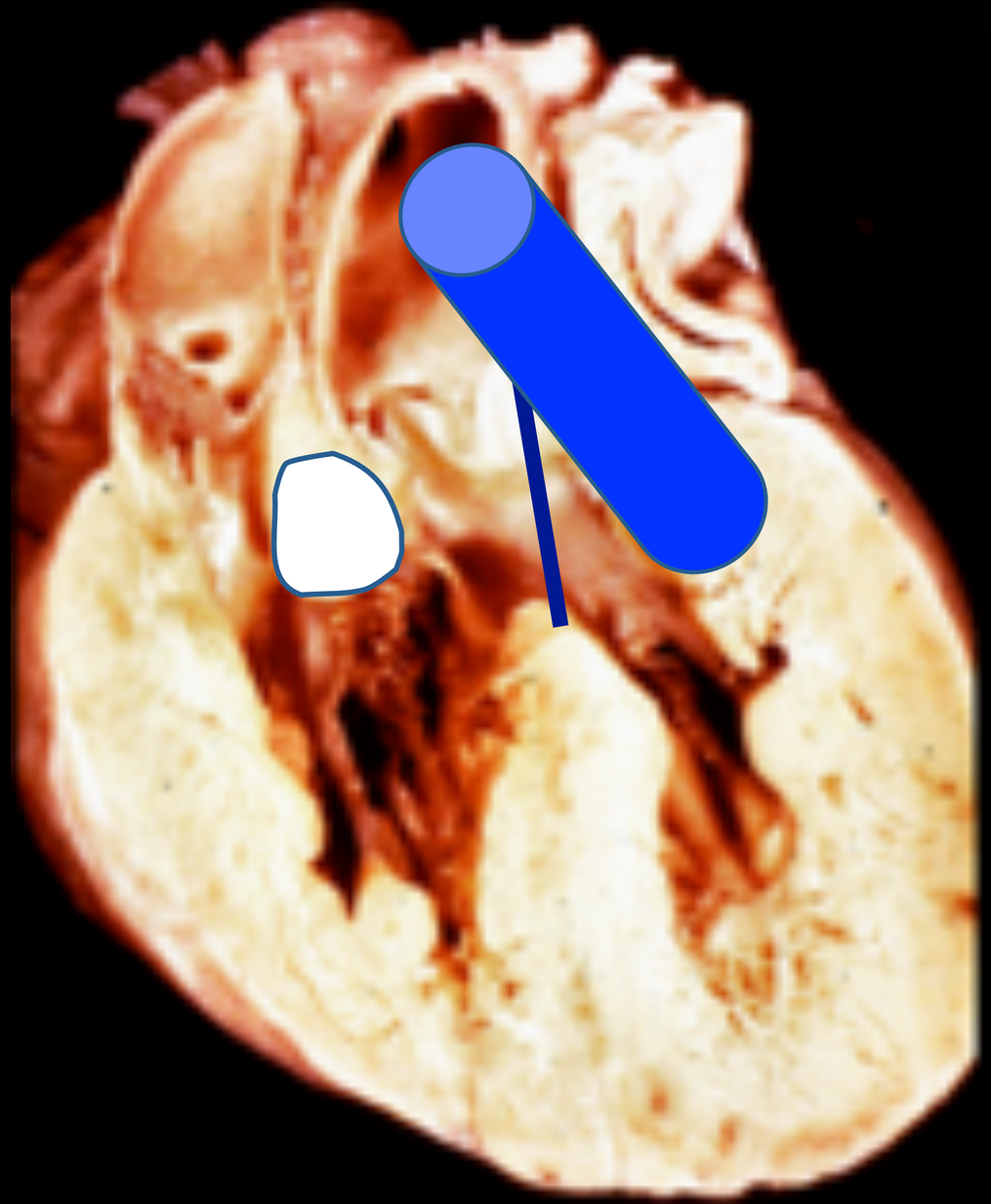
TGA

TGA - VSD - Pulmonary stenosis

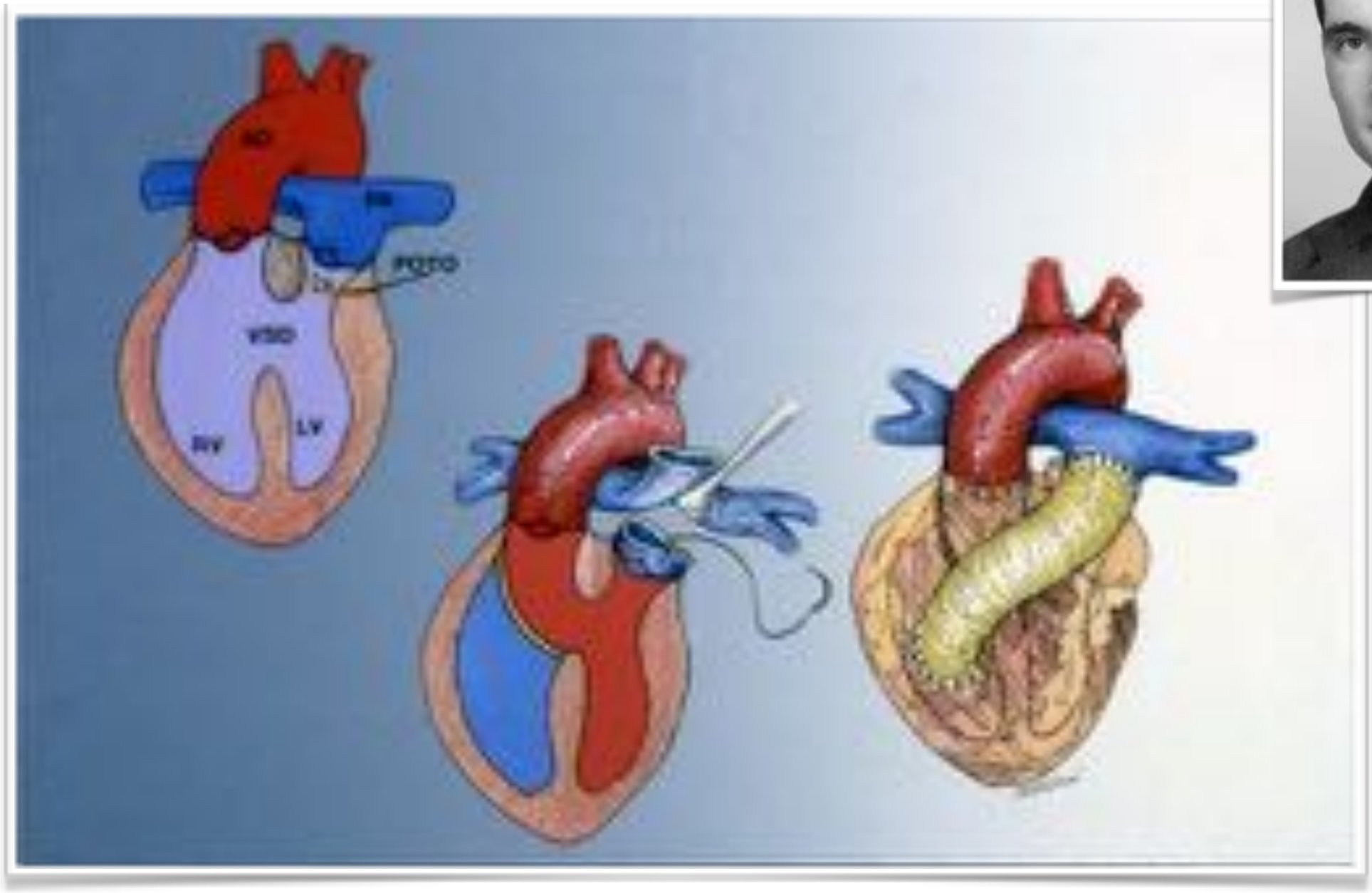


The Rastelli operation

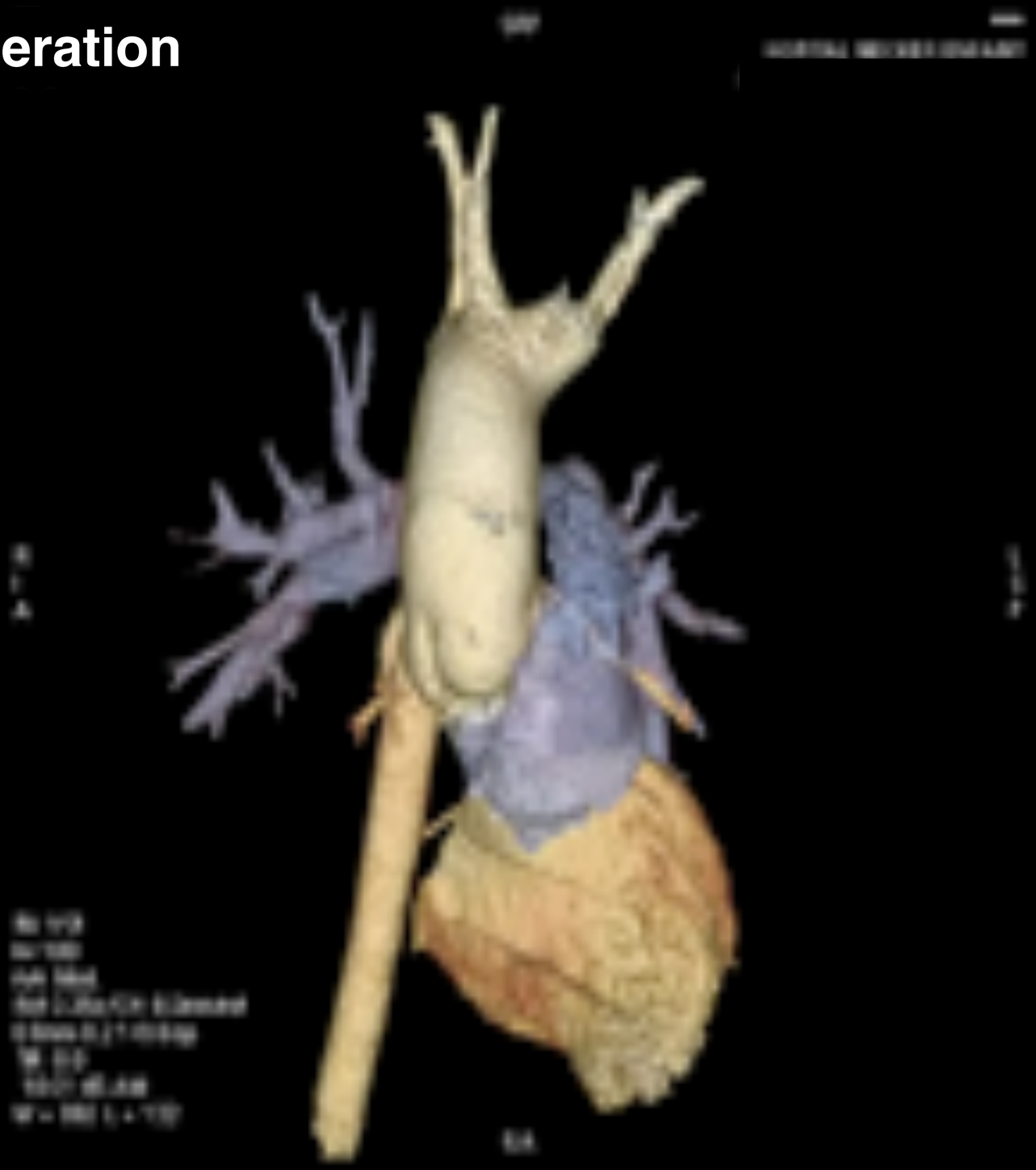




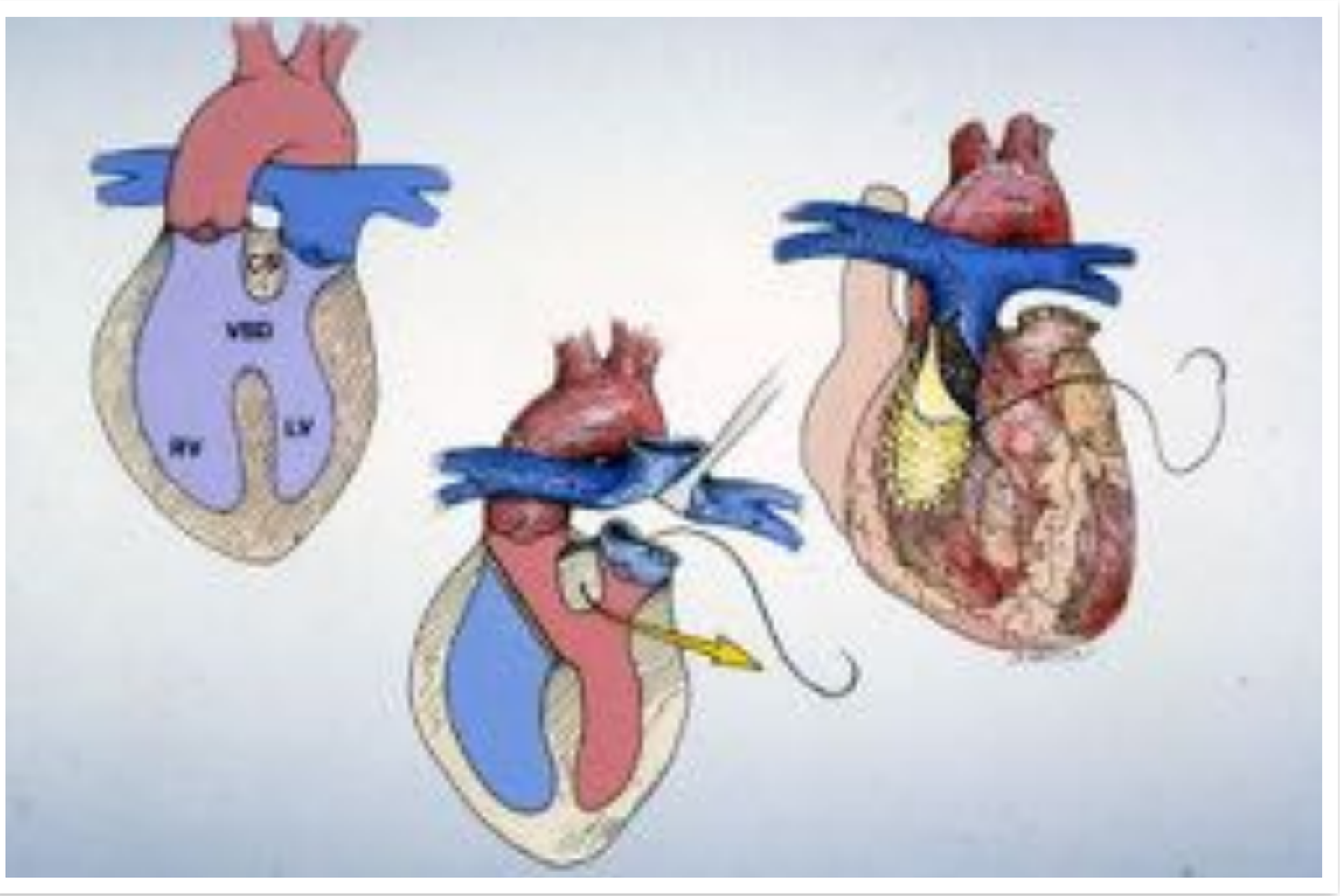
Rastelli procedure

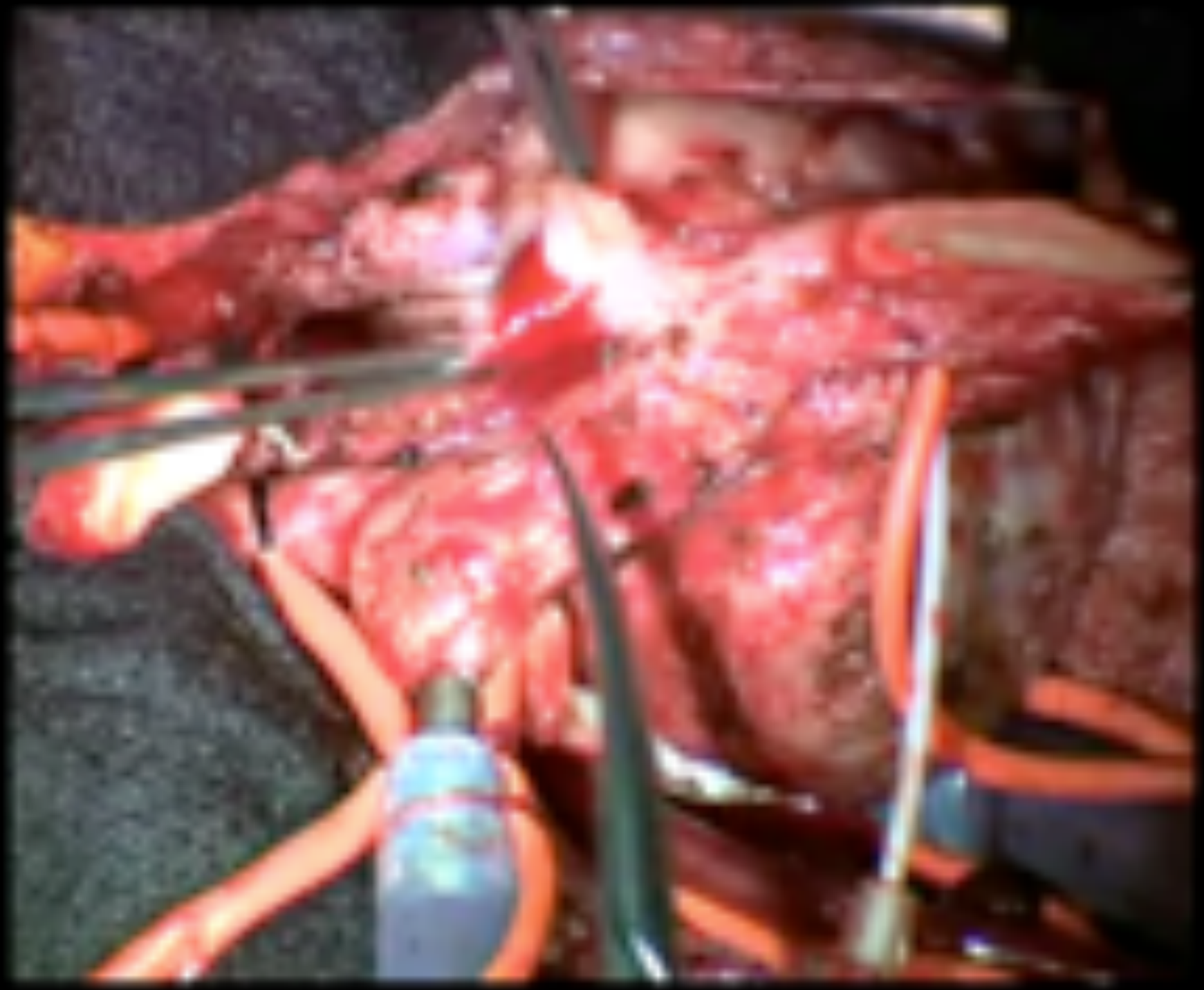


CT after Rastelli operation



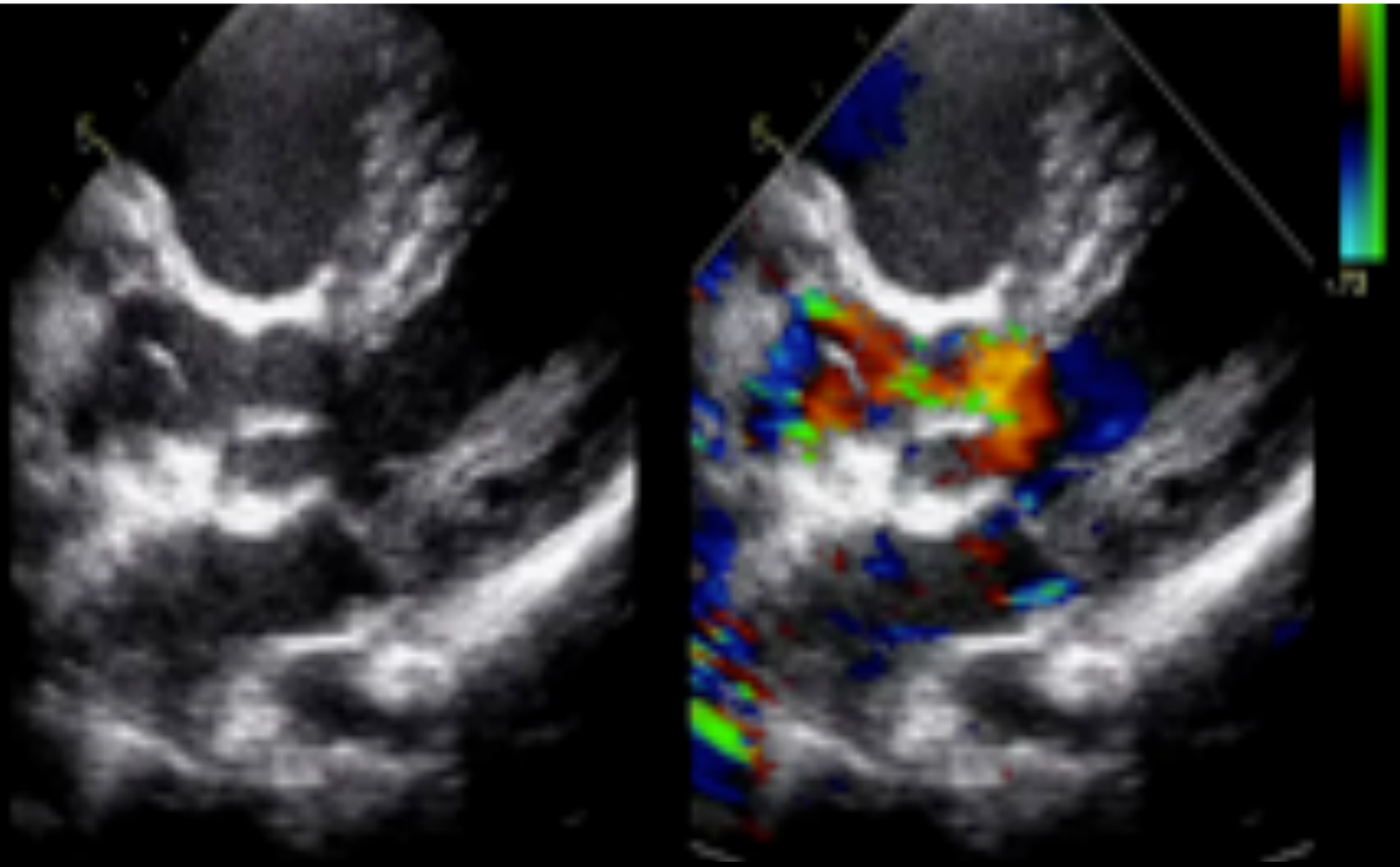
REV operation
(Réparation à l'Etage Ventriculaire)





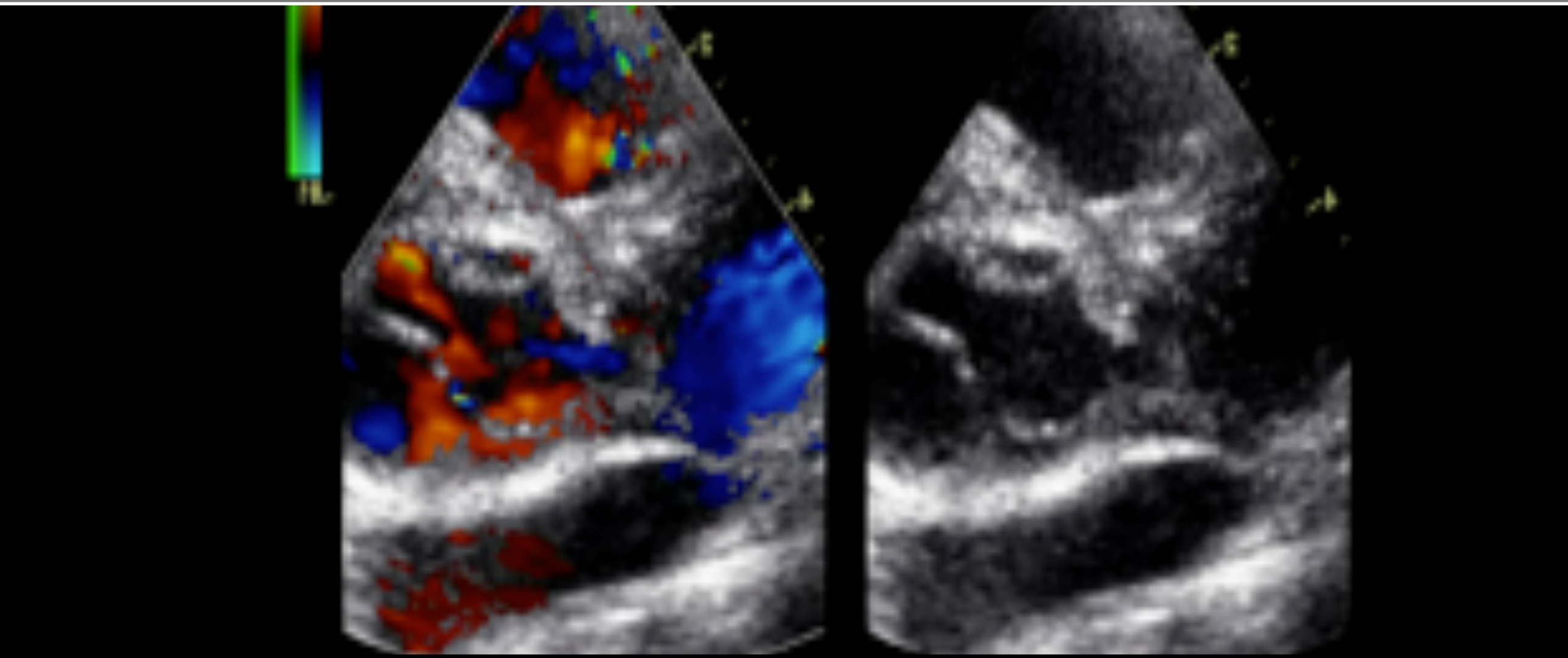
TGA

TGA - VSD - Pulmonary stenosis: tunnel from LV to aorta

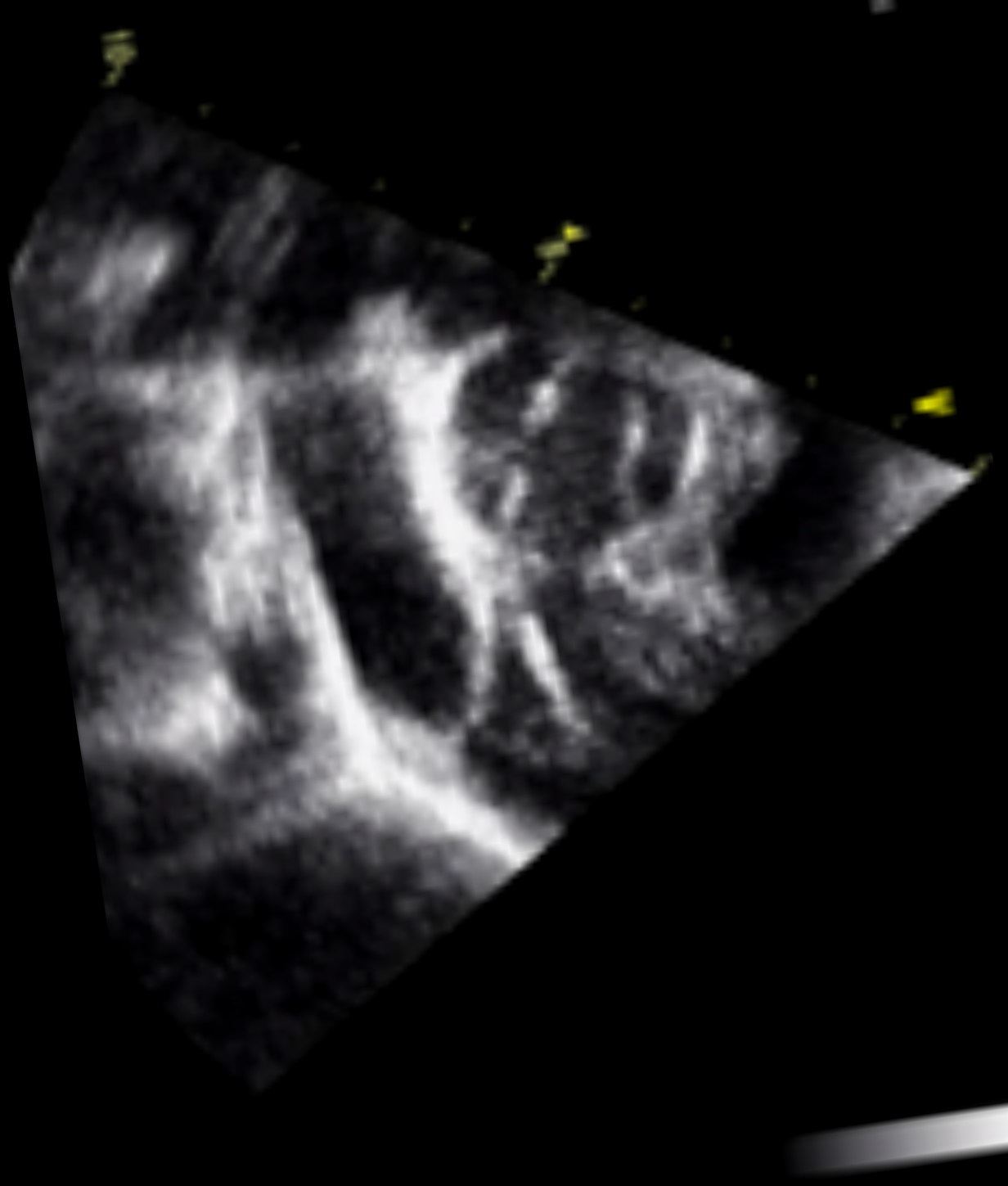


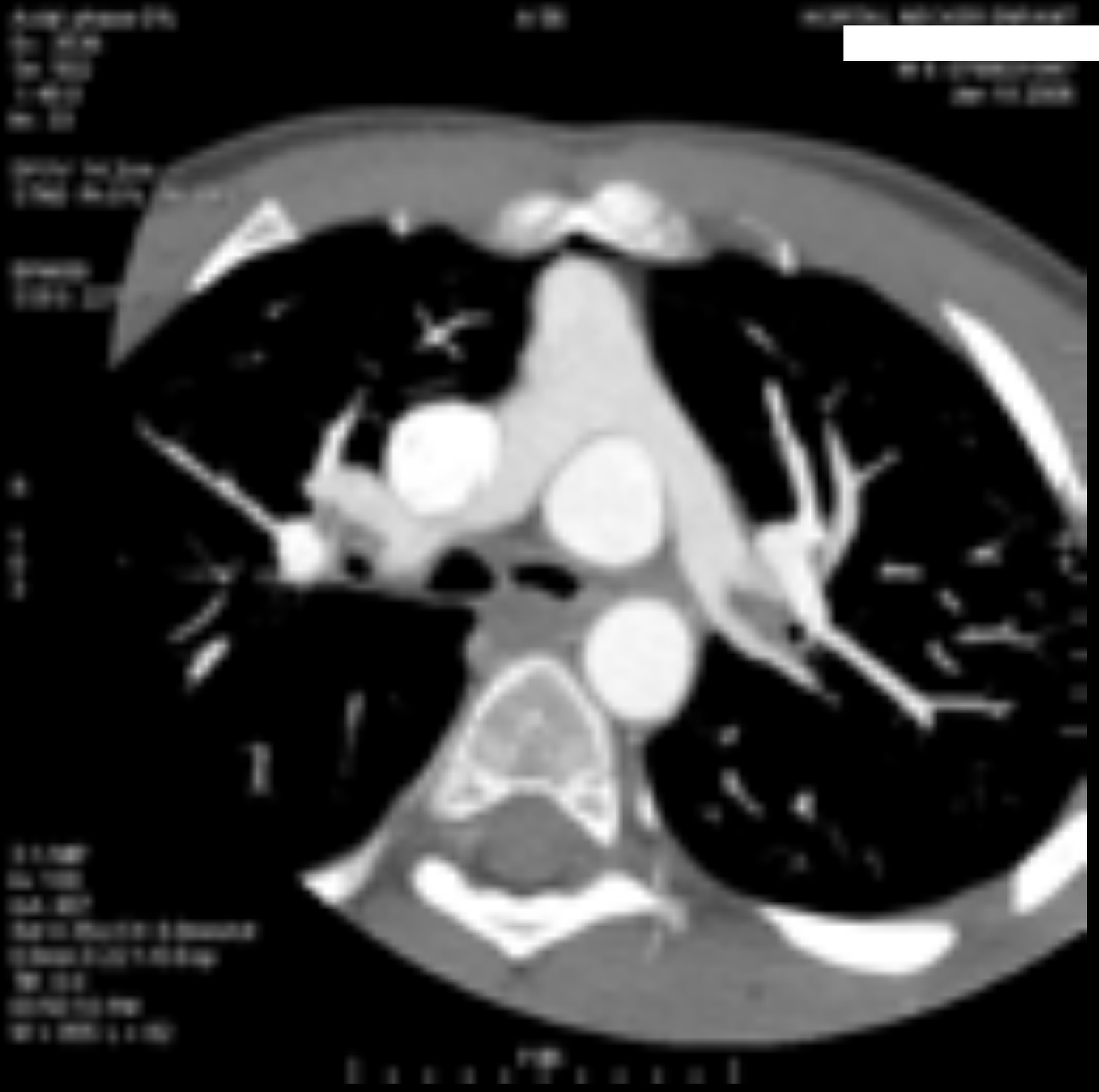
TGA

Sub-aortic obstruction after tunnel from LV to aorta



Rastelli/REV procedure
Sub aortic stenosis





Anatomically corrected TGA

Cardiol Young 2007; 17: 26–34
© Cambridge University Press
ISSN 1047-9511
doi: 10.1017/S1047951106001320

Continuing Medical Education

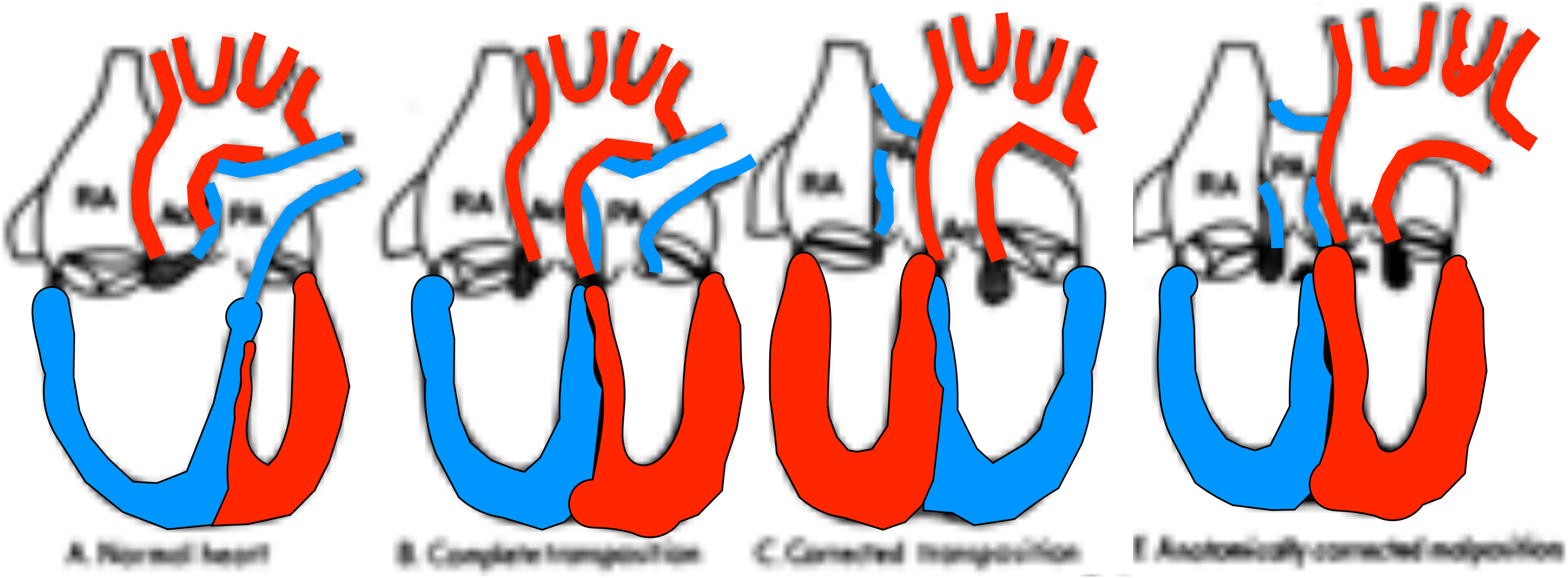
What is anatomically corrected malposition?

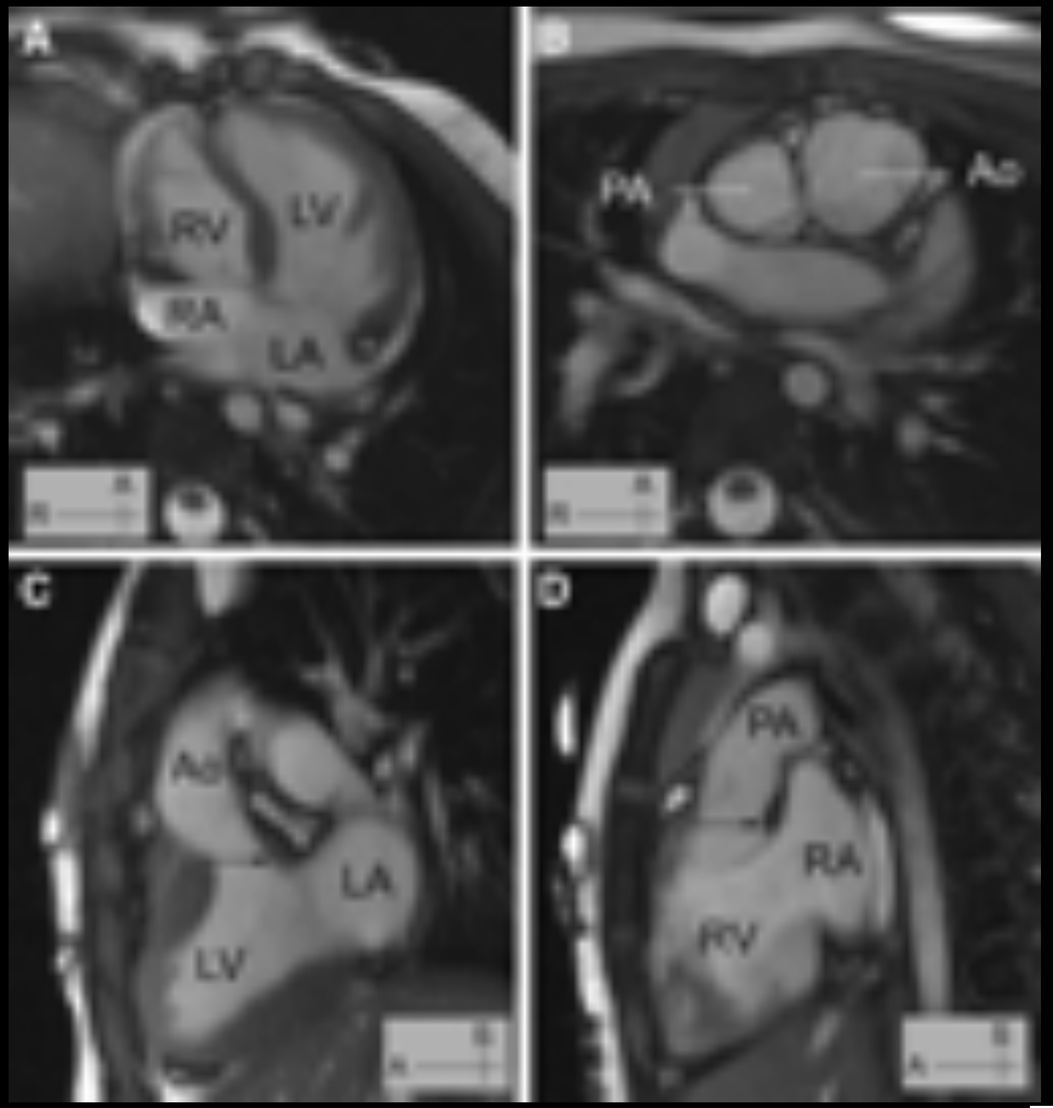
Alessandra Bernasconi,[1] Oscar Cavalle-Garrido,[1] Don G. Perrin,[2] Robert H. Anderson[3]

[1]*Division of Cardiology,* [2]*Division of Pathology, Department of Paediatrics, The Hospital for Sick Children, The University of Toronto, Toronto, Ontario, Canada;* [3]*Cardiac Unit, Institute of Child Health, University College, London, United Kingdom*

«It remains a fact, nonetheless, that many paediatric cardiologists and surgeons remain unaware of the significance of the malformation»

Anatomically corrected malposition of the great arteries





Pediatr Cardiol (2010) 31:562–563
DOI 10.1007/s00246-009-9618-8

IMAGES IN PEDIATRIC CARDIOLOGY

Anatomically Corrected Malposition of the Great Arteries

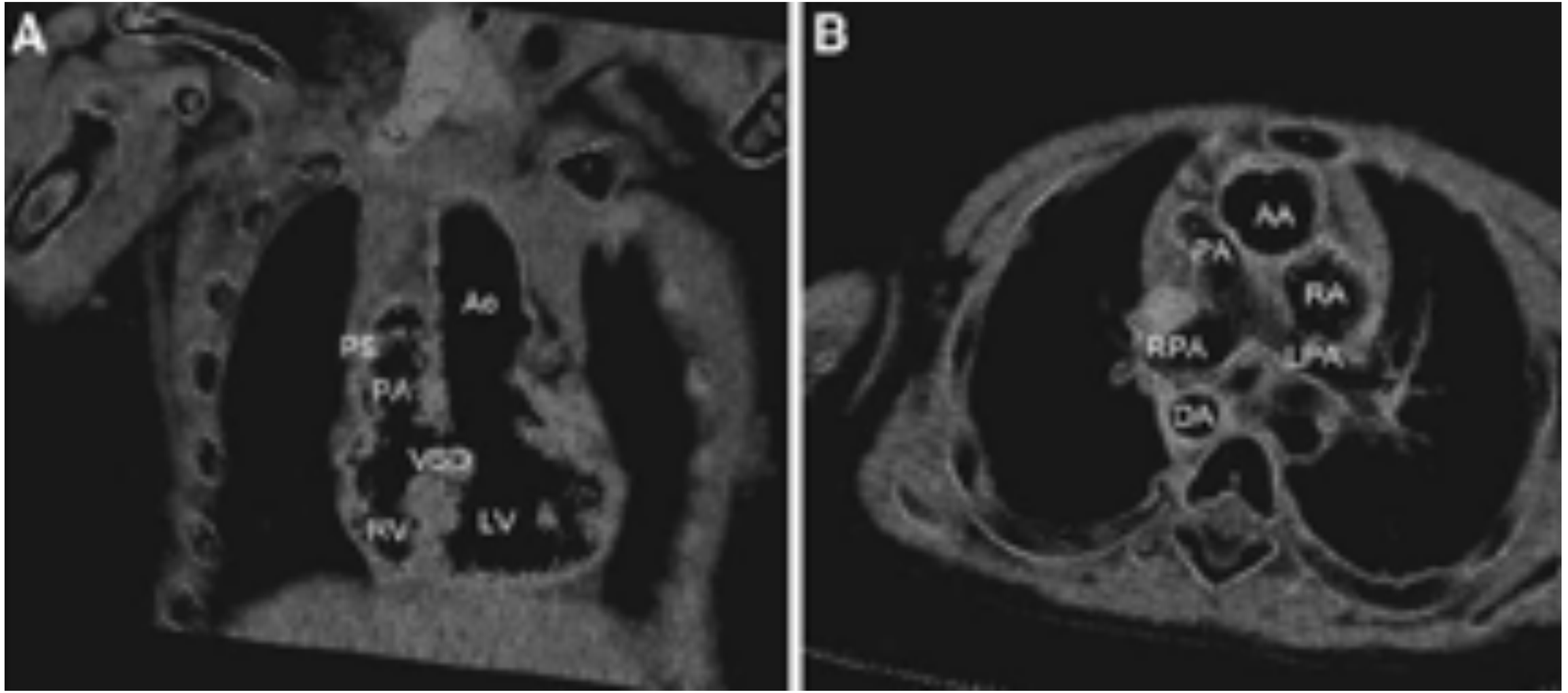
Christopher J. Clarke · K. Anitha Jayakumar · Andrew W. Hoyer

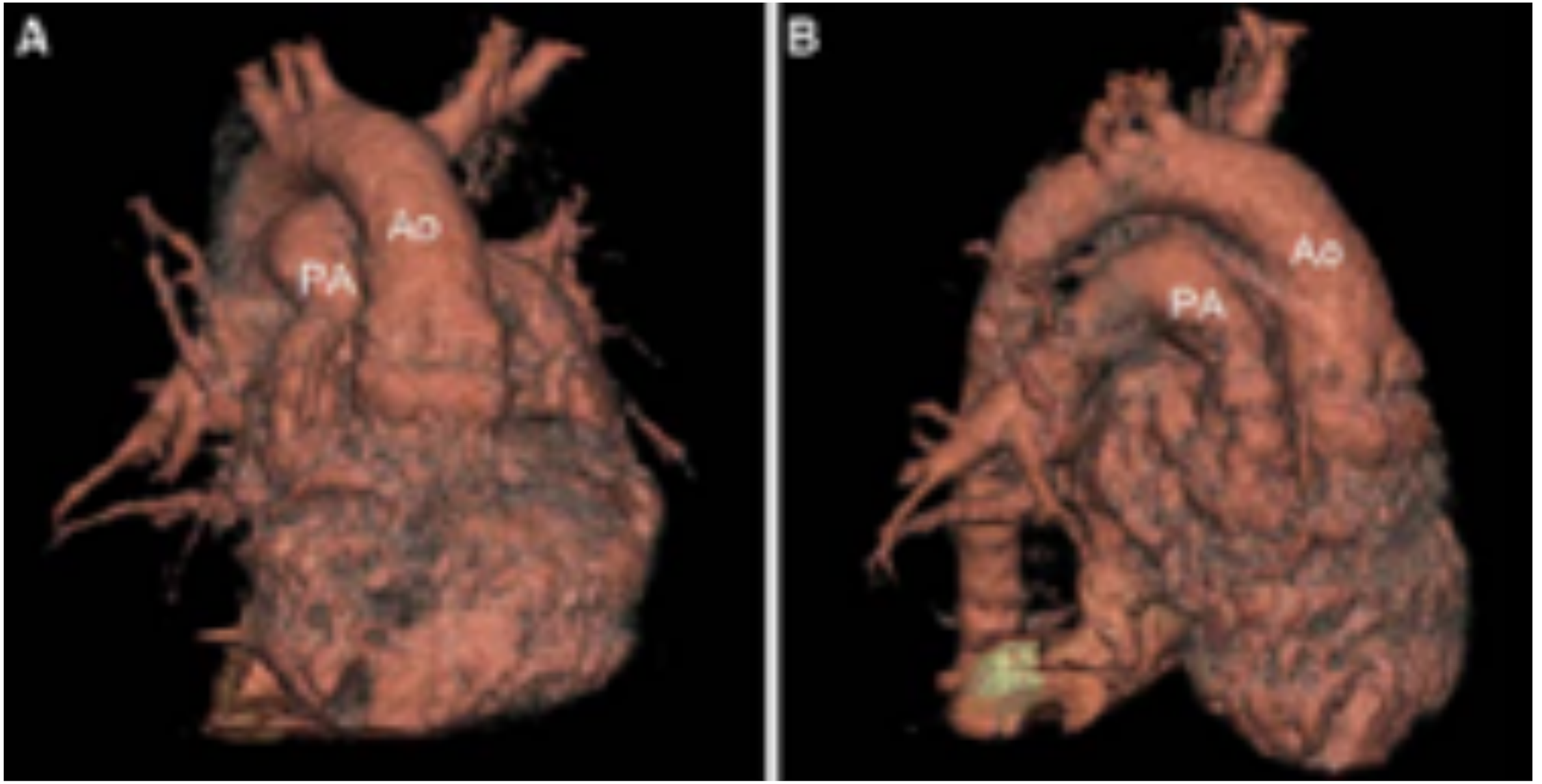
Pediatr Cardiol (2008) 29:467–468
DOI 10.1007/s00246-007-9080-4

IMAGES IN PEDIATRIC CARDIOLOGY

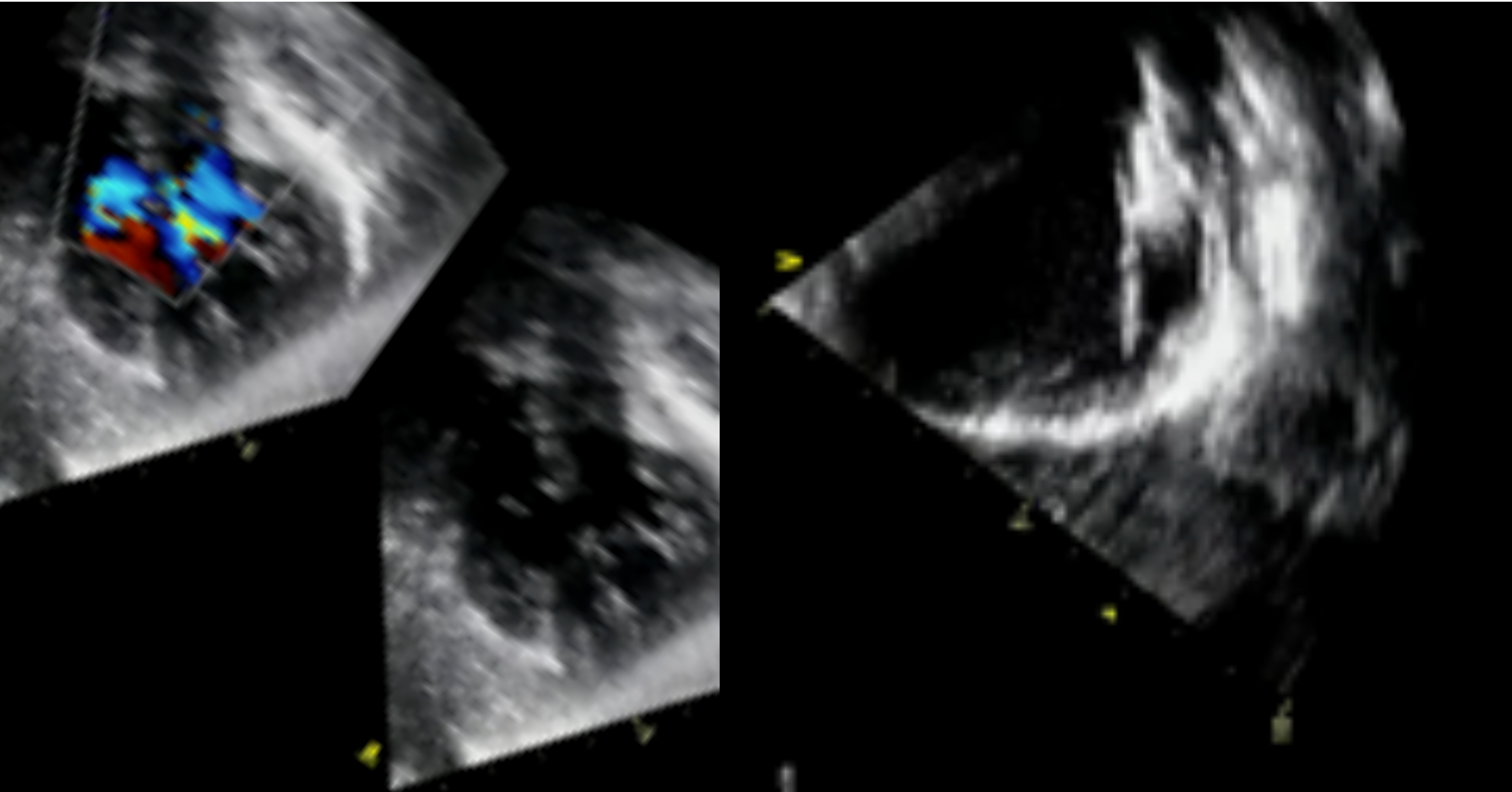
Anatomically Corrected Malposition of the Great Arteries

Ming-Ren Chen




A
PA
Ao
B
PA
Ao

Anatomically corrected malposition of the great arteries



Anatomically corrected malposition of the great arteries