Malpositions of the great arteries

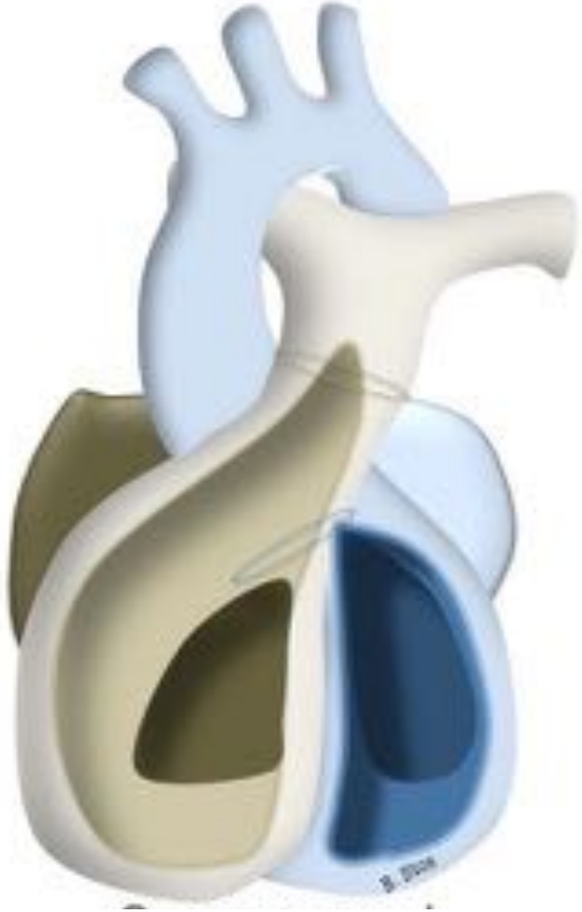
Damien Bonnet

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

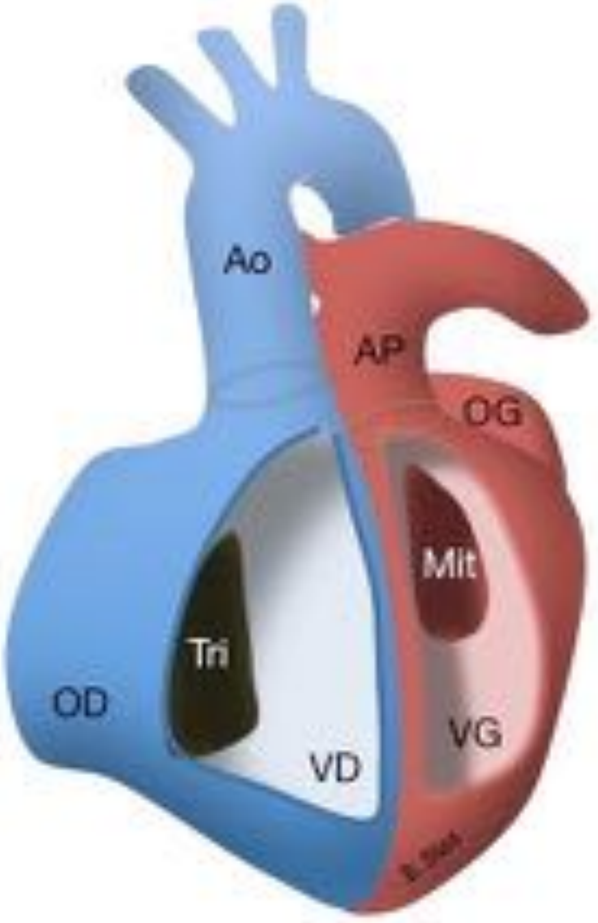
Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

ANATOMY OF THE HEART WITH TGA

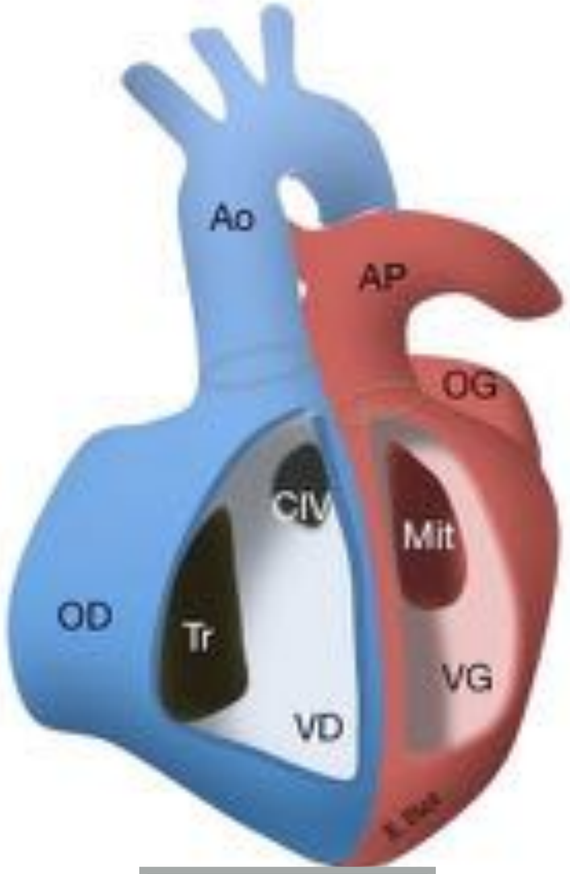


Normal heart

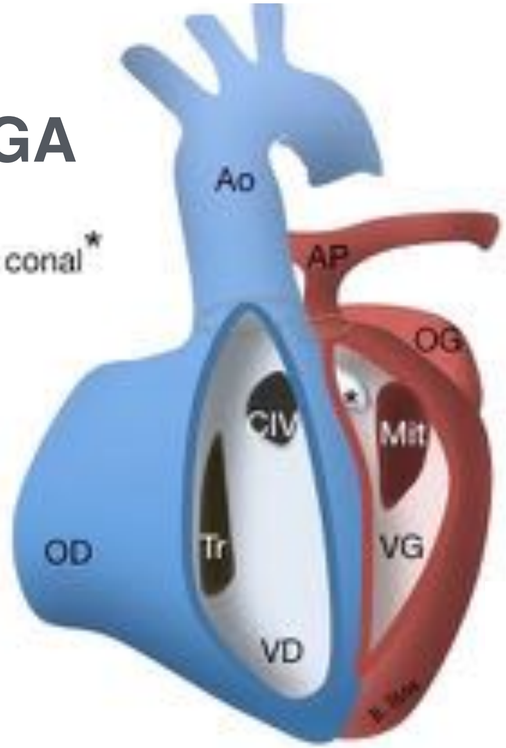


TGA

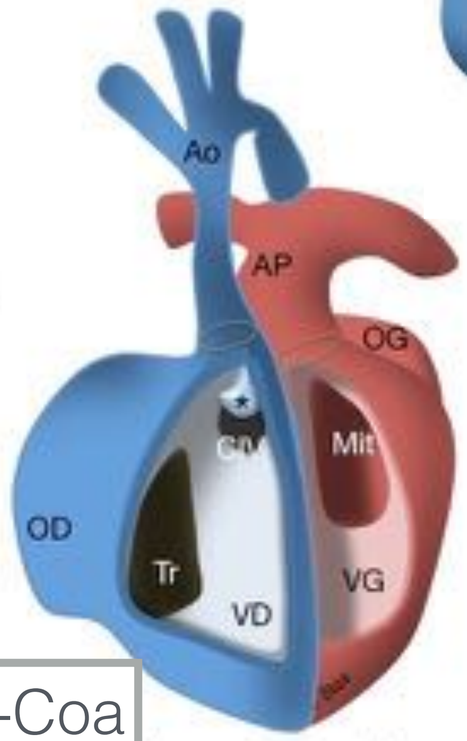
Other complexe malpositions of the GA



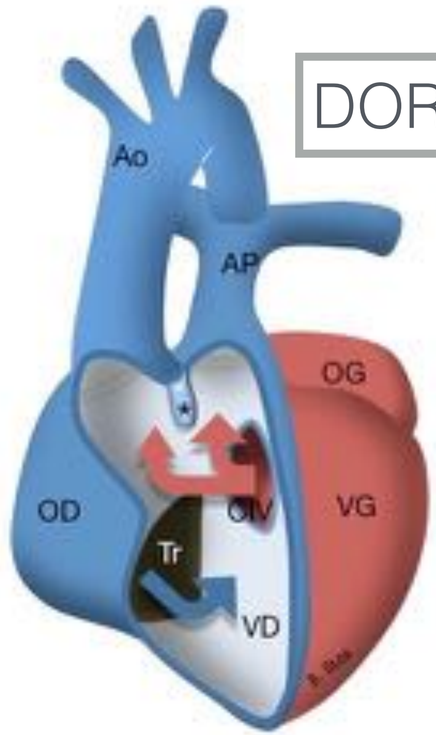
TGA-VSD



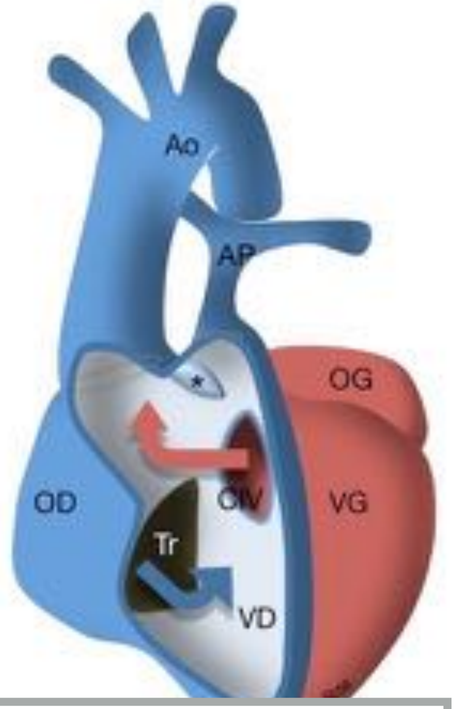
TGA-VSD-PS



TGA-VSD-Coa



DORV-Sub aortic VSD

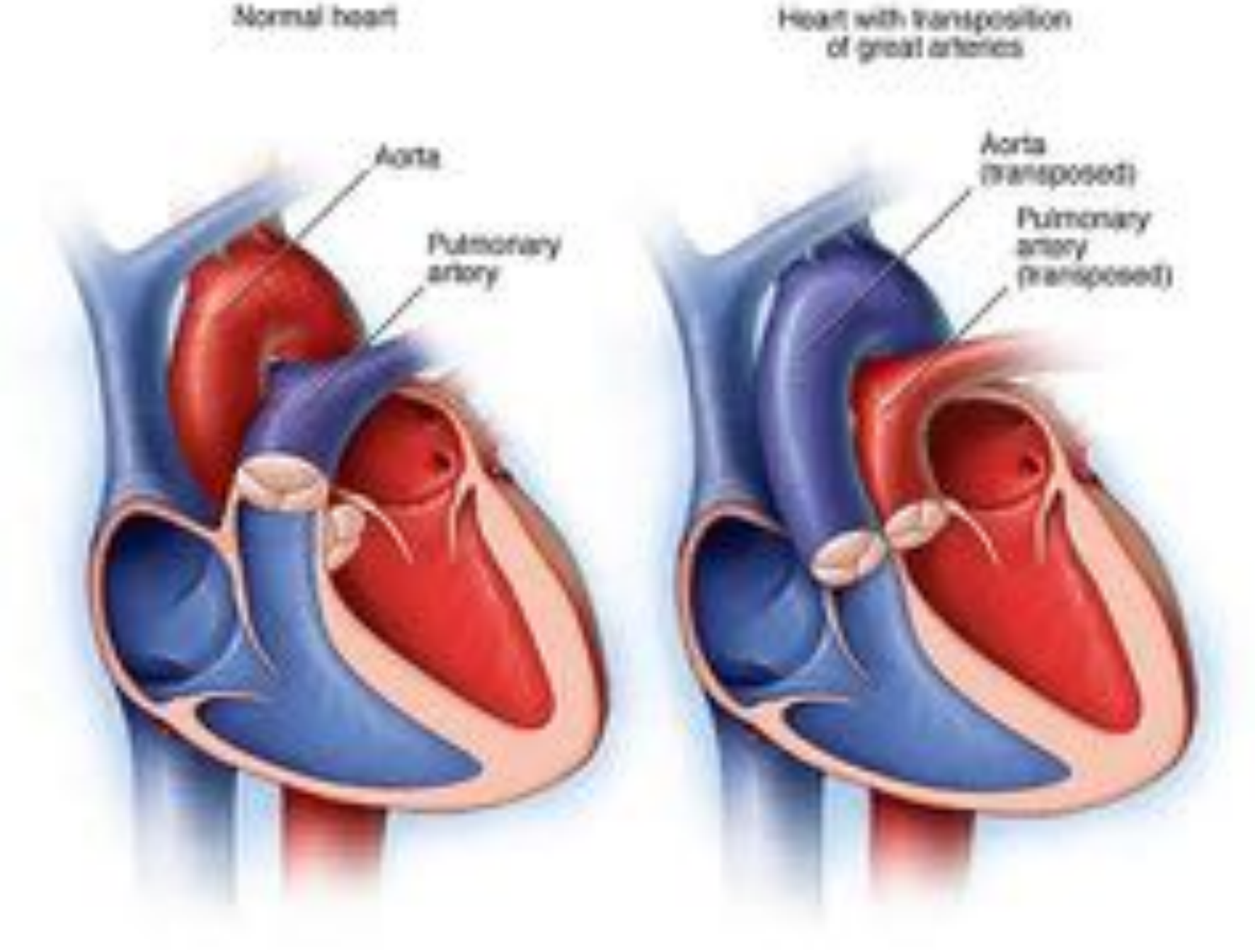


DORV-Sub aortic VSD-SP

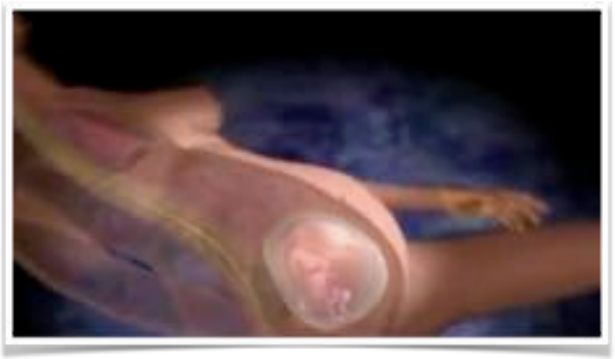
Malpositions of the great arteries

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)

Foetal TGA



The French system during pregnancy

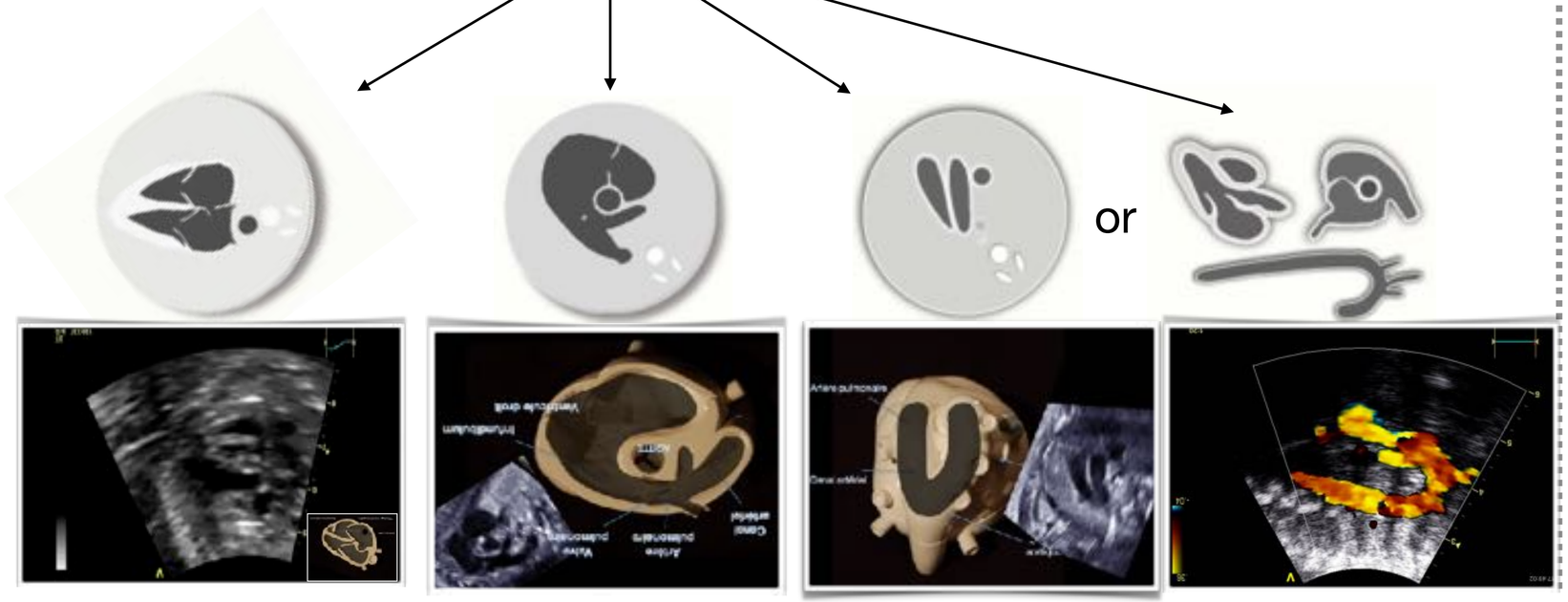
3 systematic foetal echographies - Level 1

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

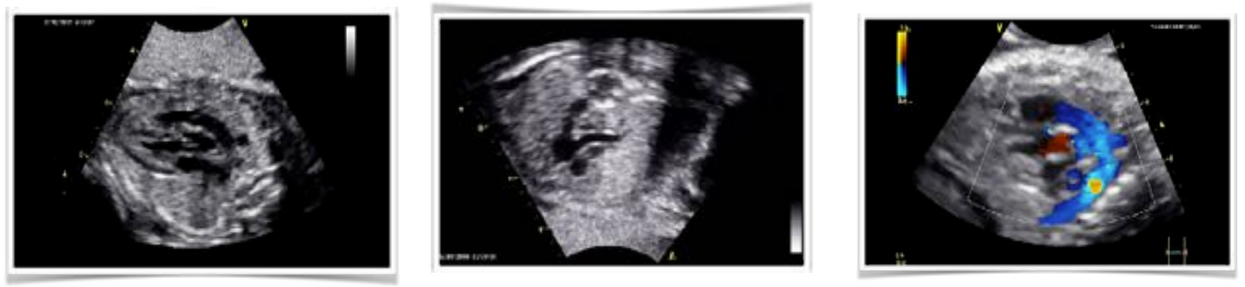
↓

Level 3

Fetal echocardiography
by expert

Prenatal diagnosis of TGA

TGA



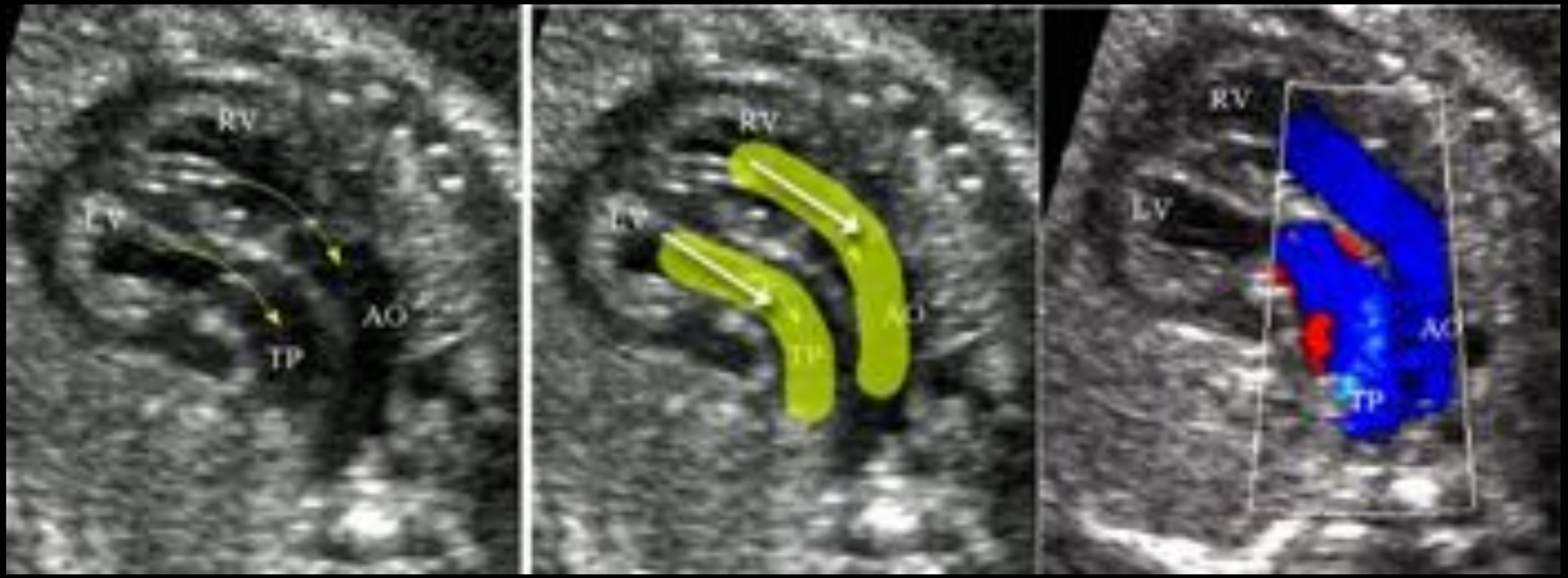
Preoperative mortality in TGA = 4-6%
(*vs./+)* Surgical mortality = 1-2%

Comparison of Characteristics of Patients in the Prenatal and Postnatal Groups

	Postnatal Group	Prenatal Group	*P*
Isolated TGA	304	57	NS
Associated defects	46	11	NS
VSD	31	8	NS
VSD + CoA	14	3	NS
CoA	1	1	NS
Age at admission, h	73 ± 216	2.2 ± 2.8	<0.01
Mechanical ventilation	95 (28)	12 (17.6)	<0.01
Metabolic acidosis + MOF	52	6	<0.05
PGE, infusion	95	22	NS
ISAS	168	54	NS
Preoperative mortality	15	0	<0.05
Coronary artery pattern	222 400	68 800	
Normal	168	47	NS
Abnormal	60	21	NS
Postoperative mortality	20	0	<0.01
Hospital stay, d	30 ± 17	24 ± 11	<0.01

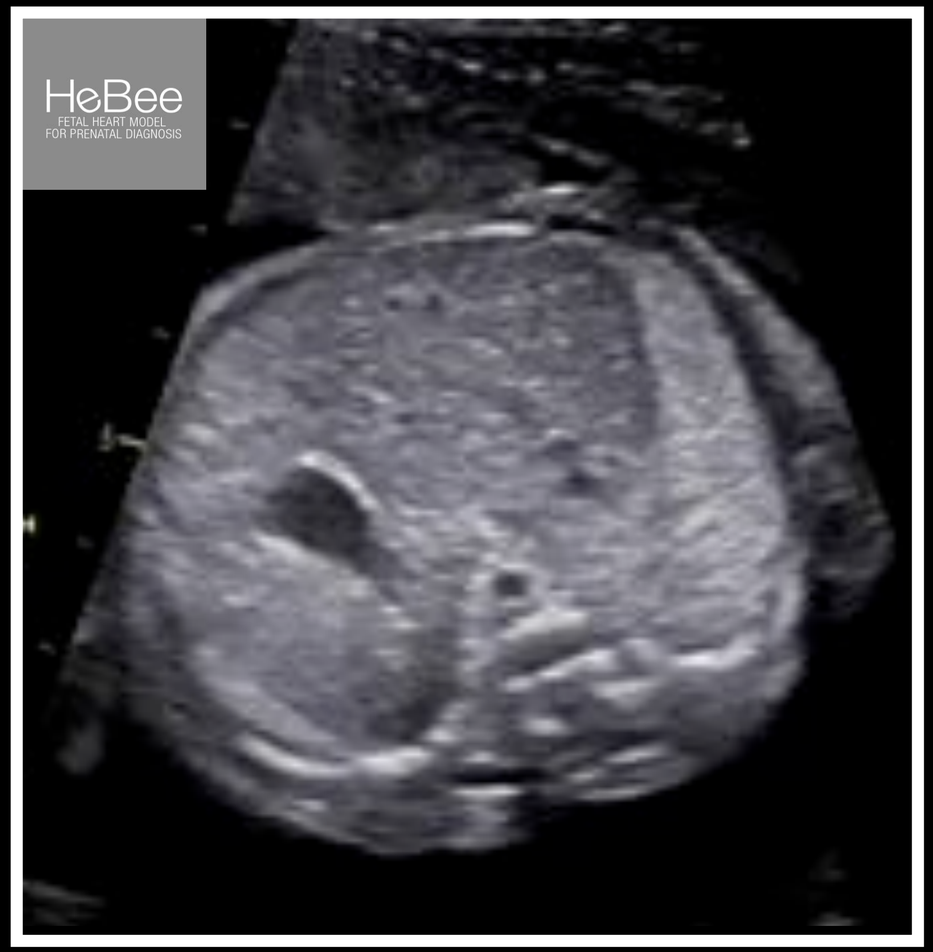
VSD indicates ventricular septal defect; CoA, coarctation; MOF, multiorgan failure; PGE, prostaglandin E; SAS, balloon atrioseptostomy; and ASO, arterial switch operation. Values are n (%).

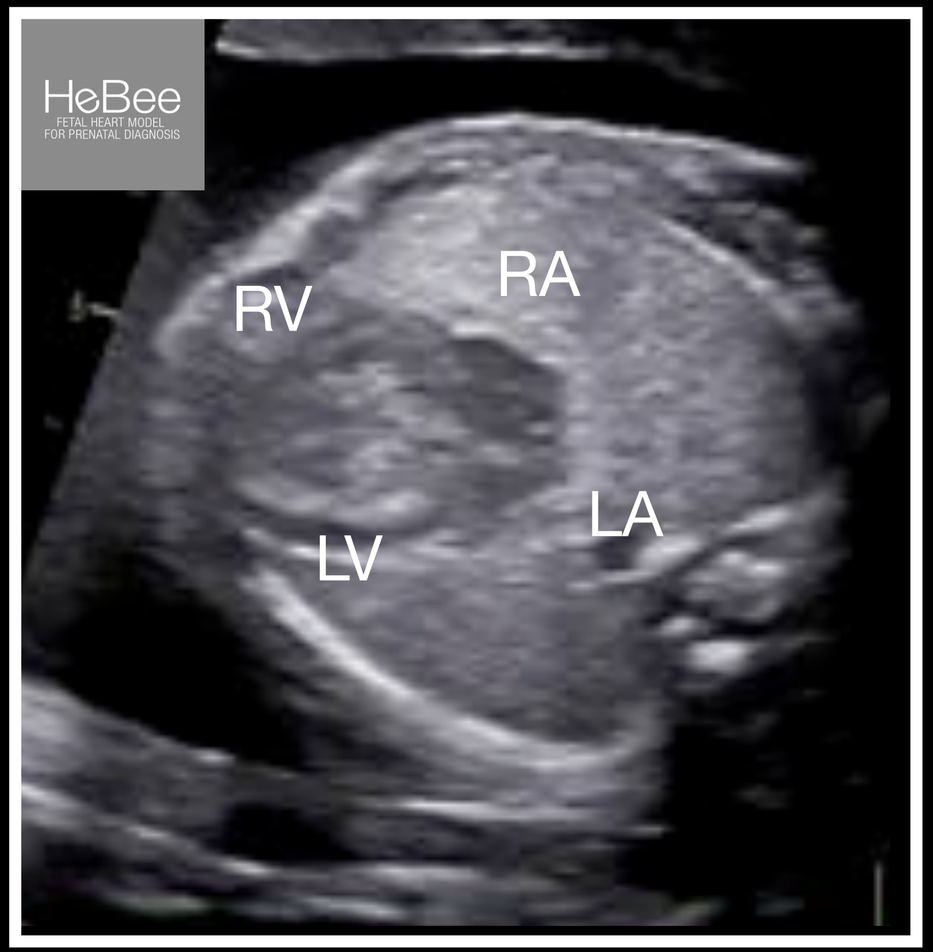
Bonnet D et al. Circulation 1997


RV
LV
AO
TP
RV
LV
AO
TP
RV
LV
AO
TP

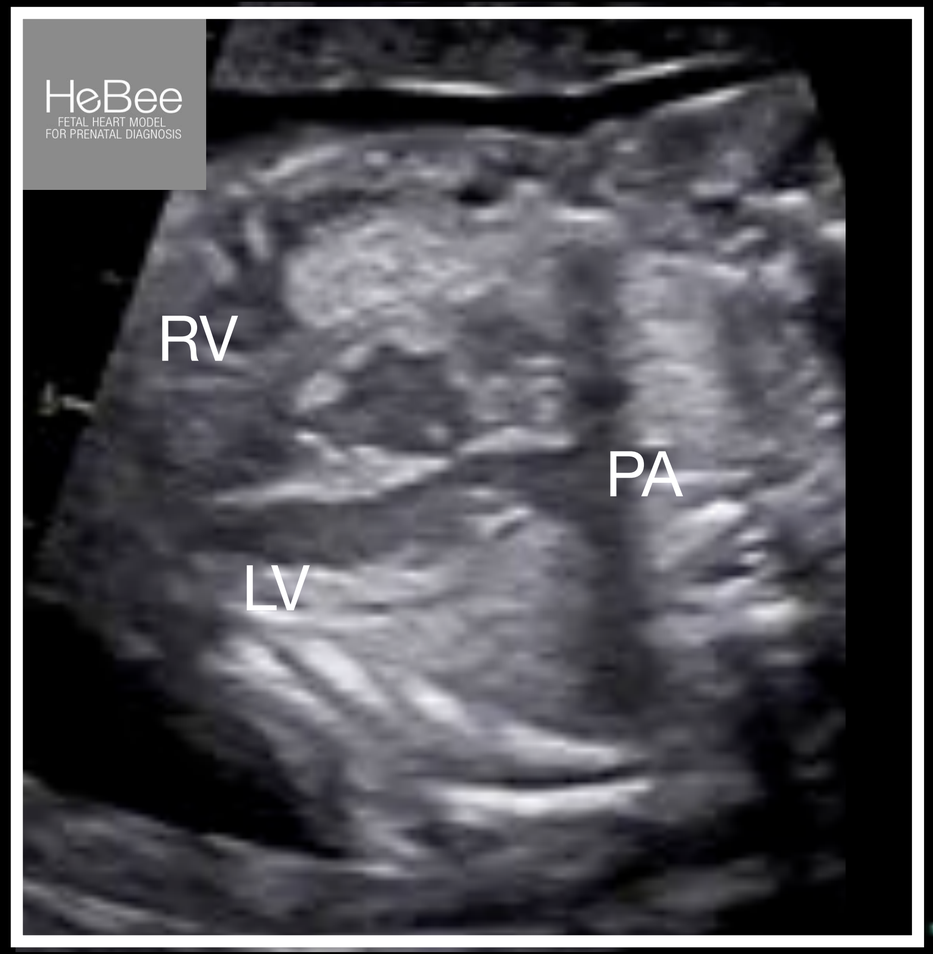
TGA

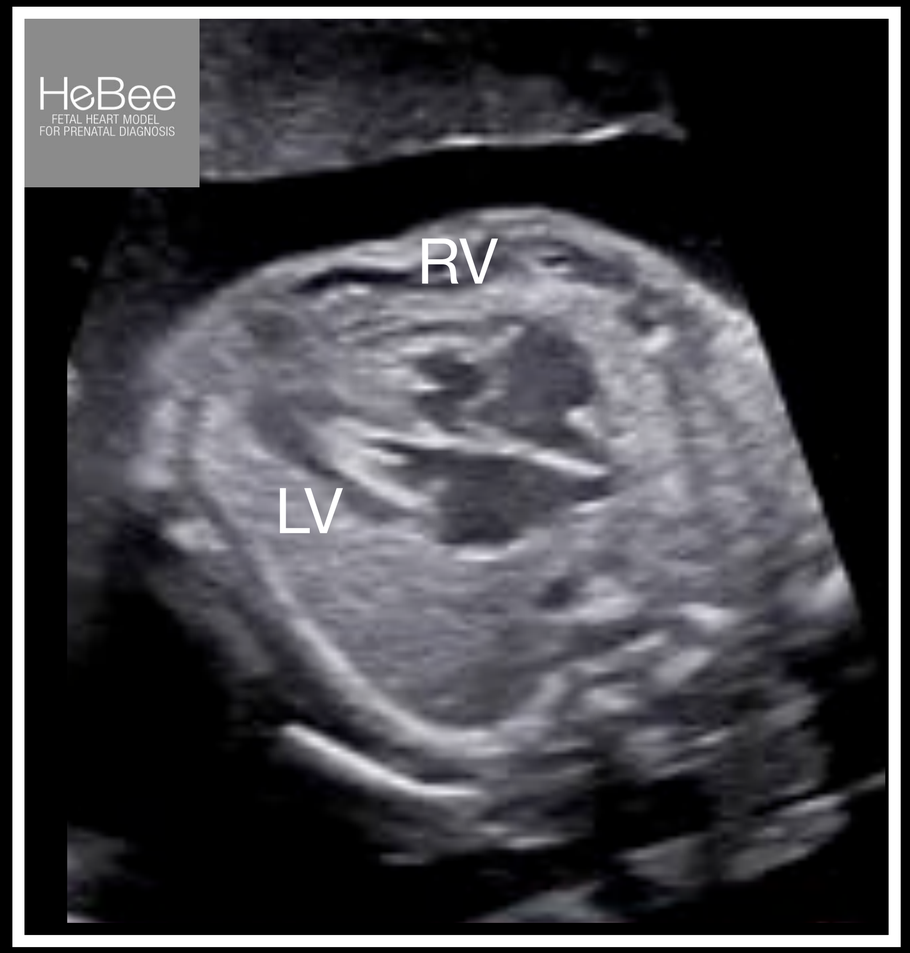
situs & 4 chamber view





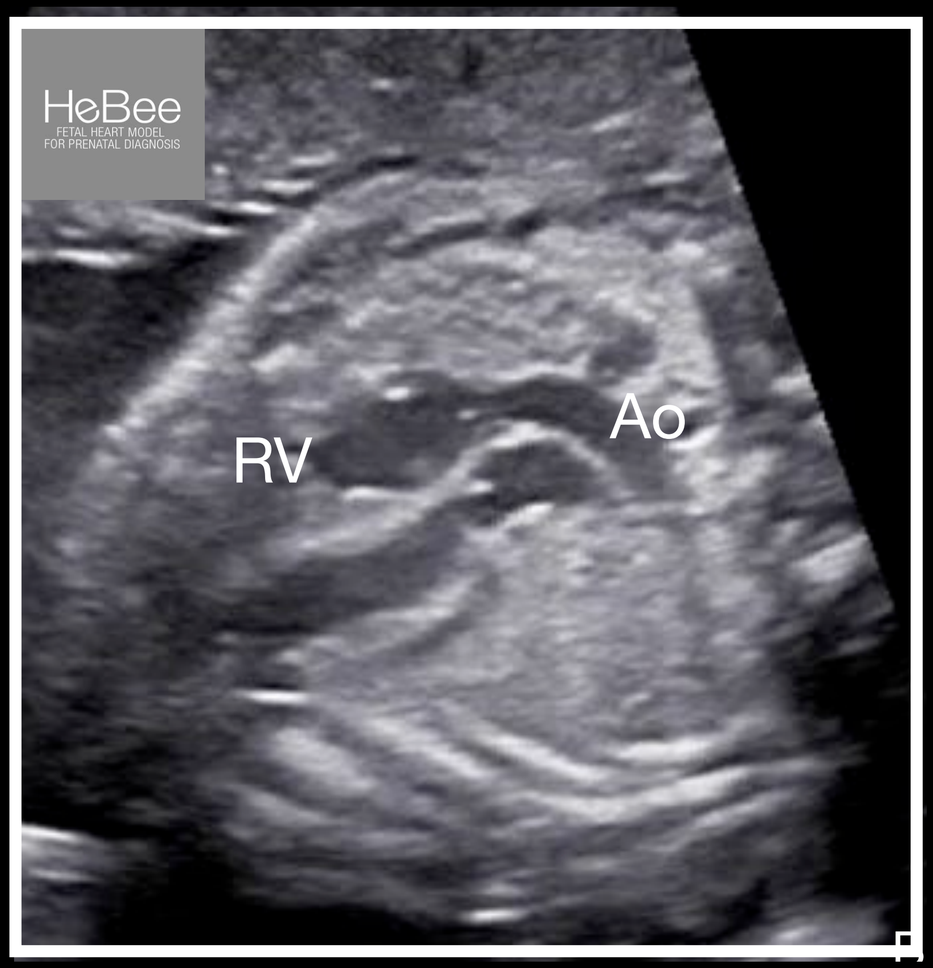
TGA
LVOT view

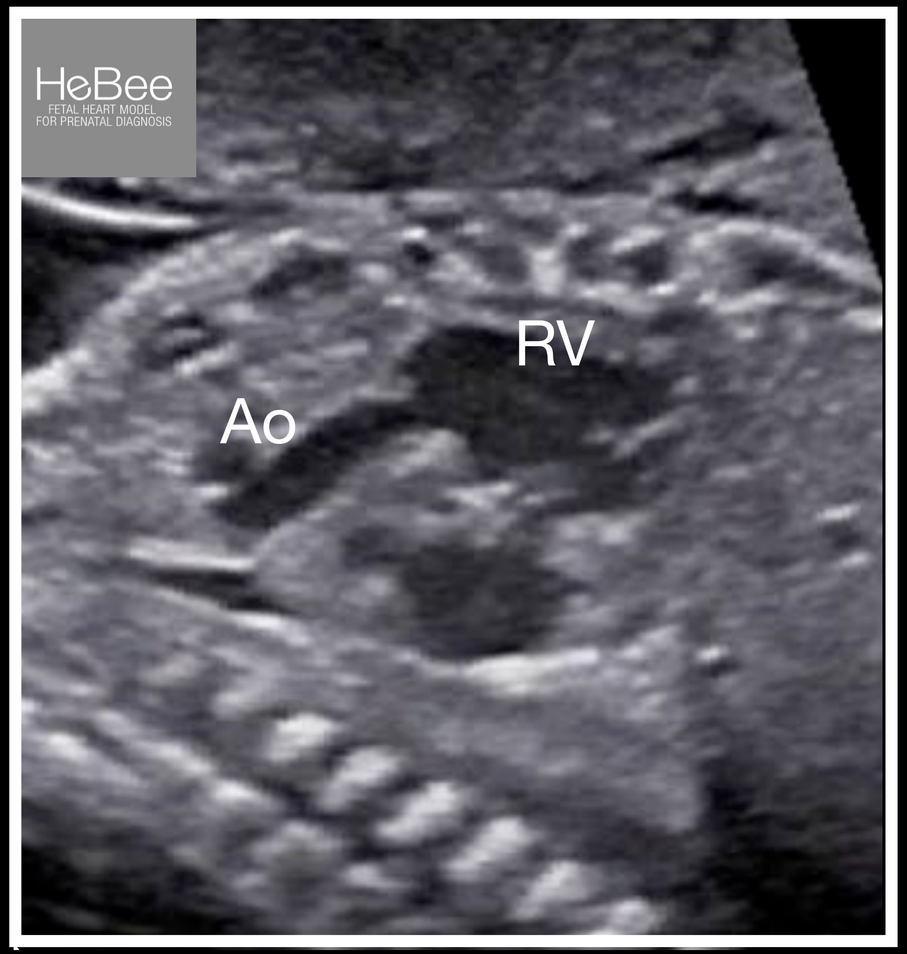




TGA

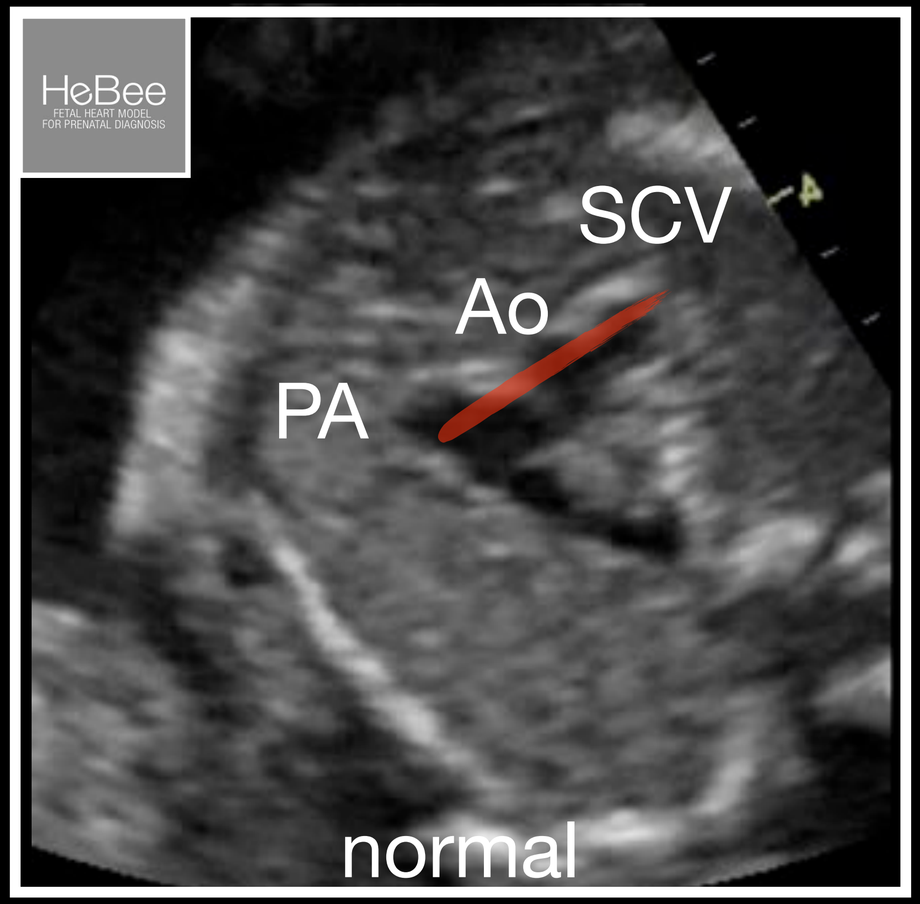
LVOT/RVOT view

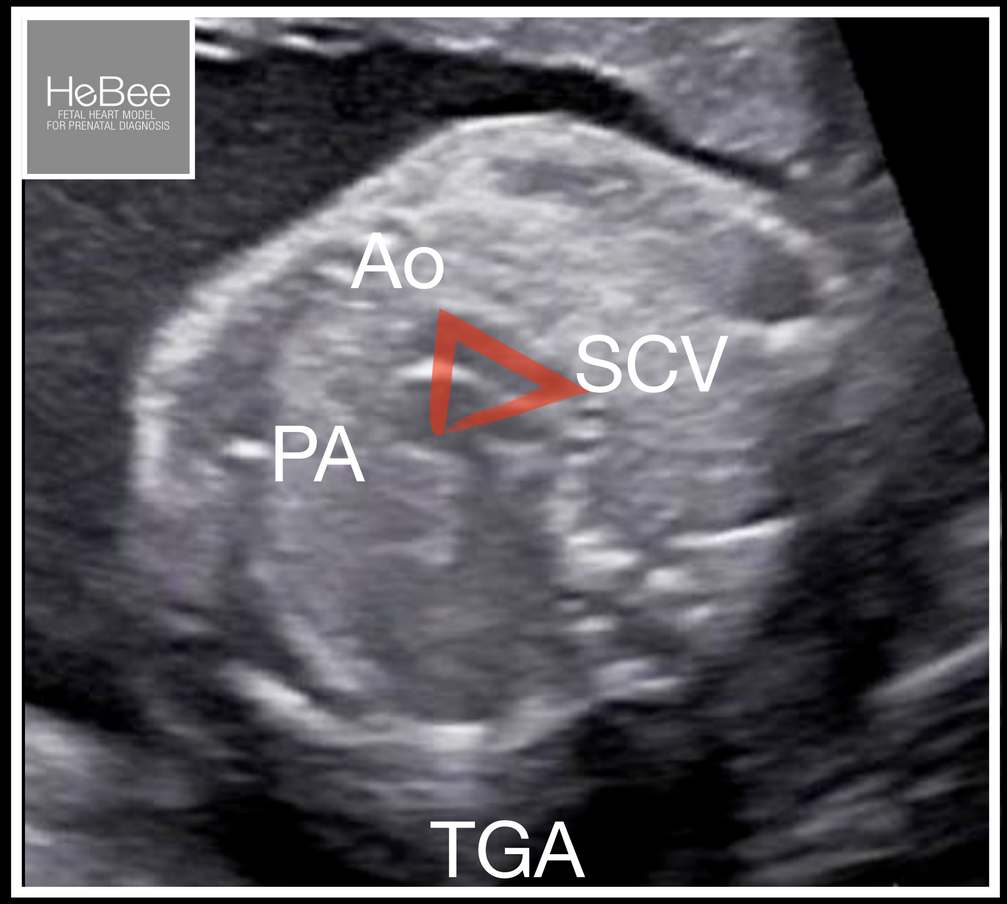




TGA

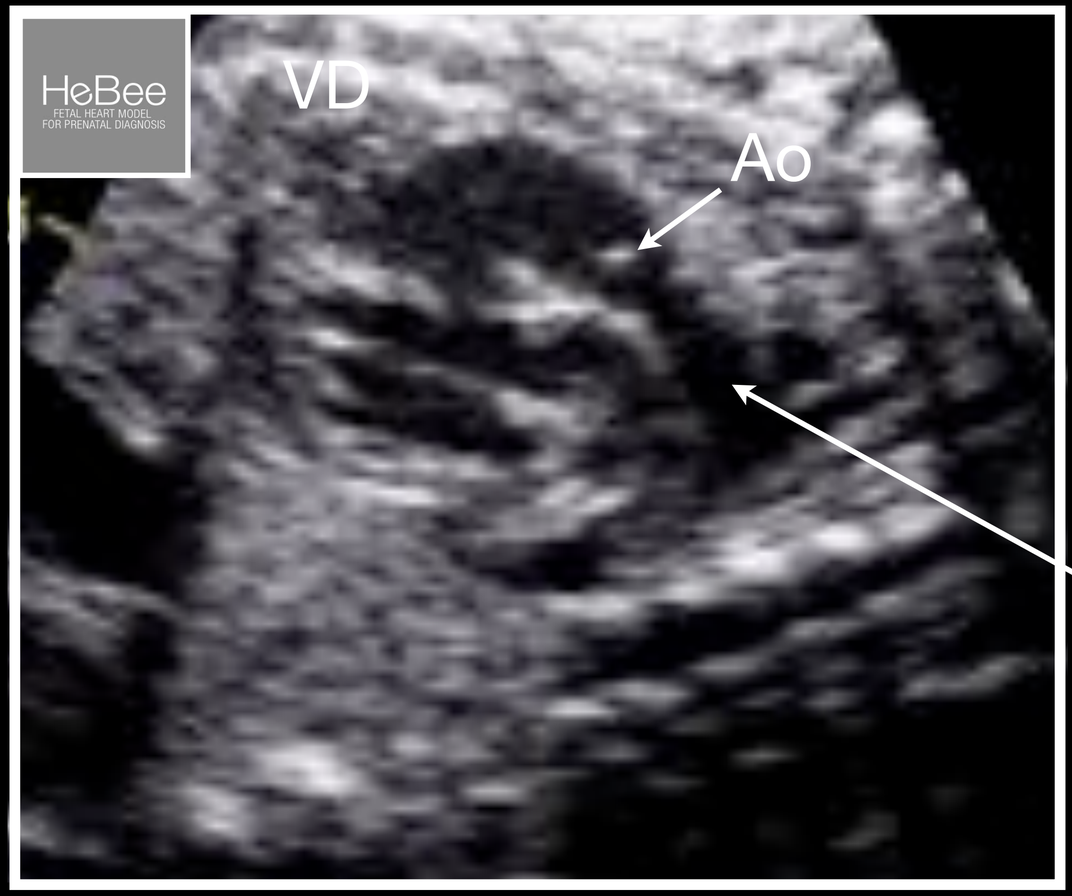
3 vessel view





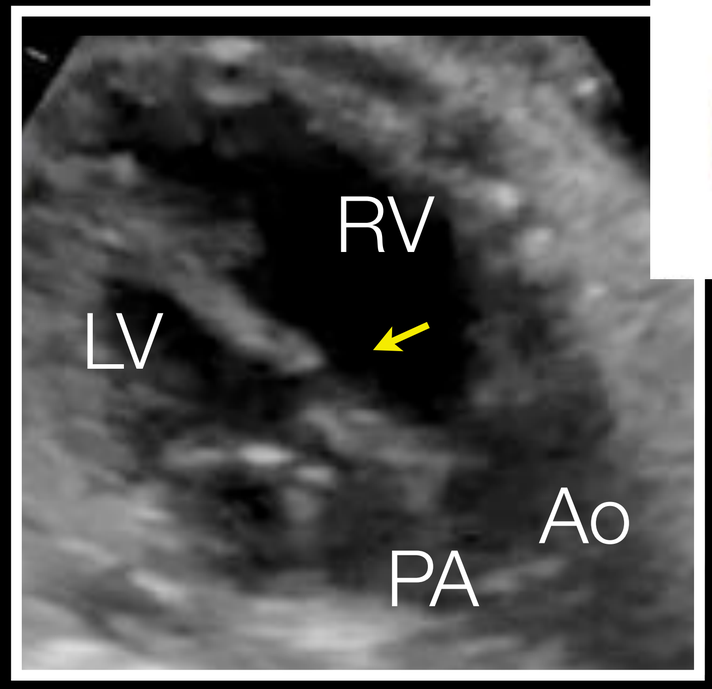
TGA

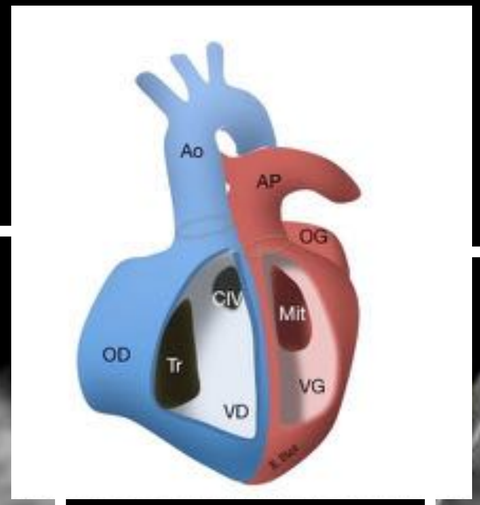
RVOT view

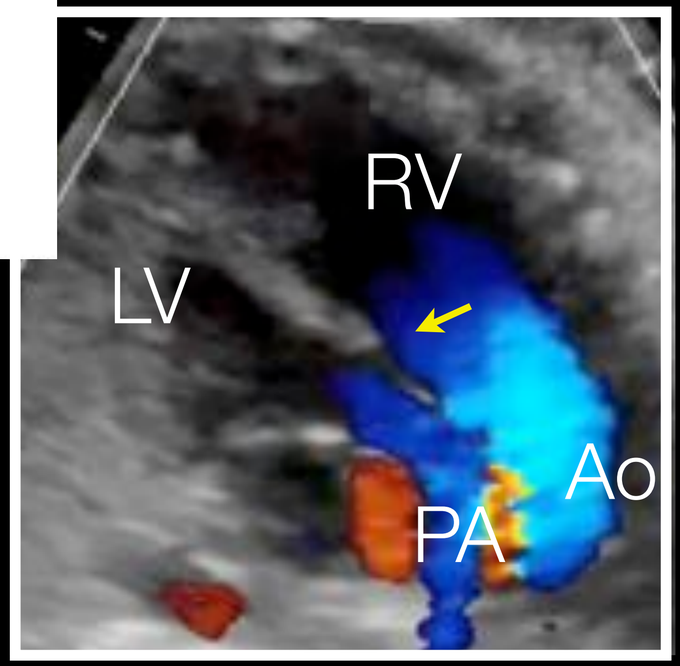


TGA

Complex forms



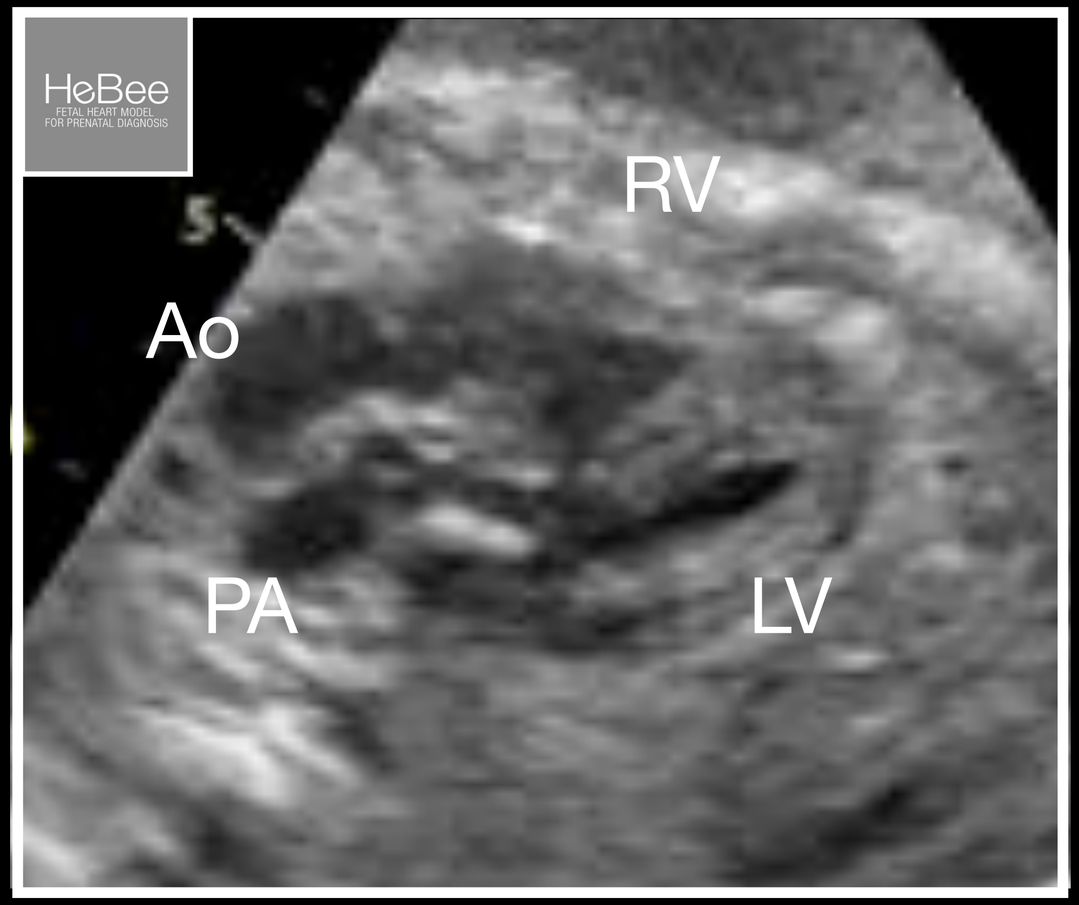




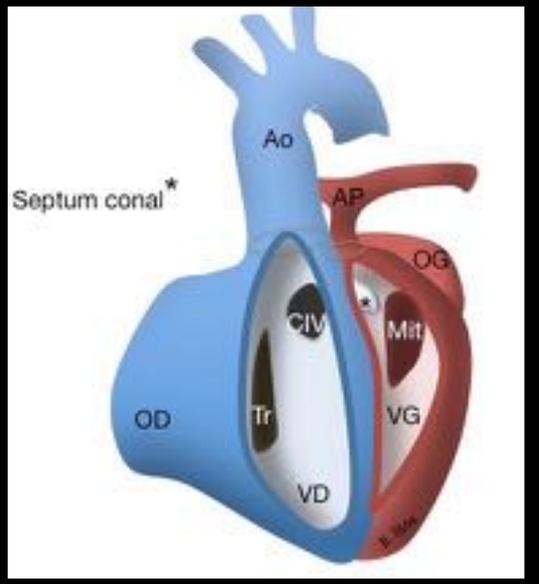
TGA-membranous VSD

TGA

Complex forms

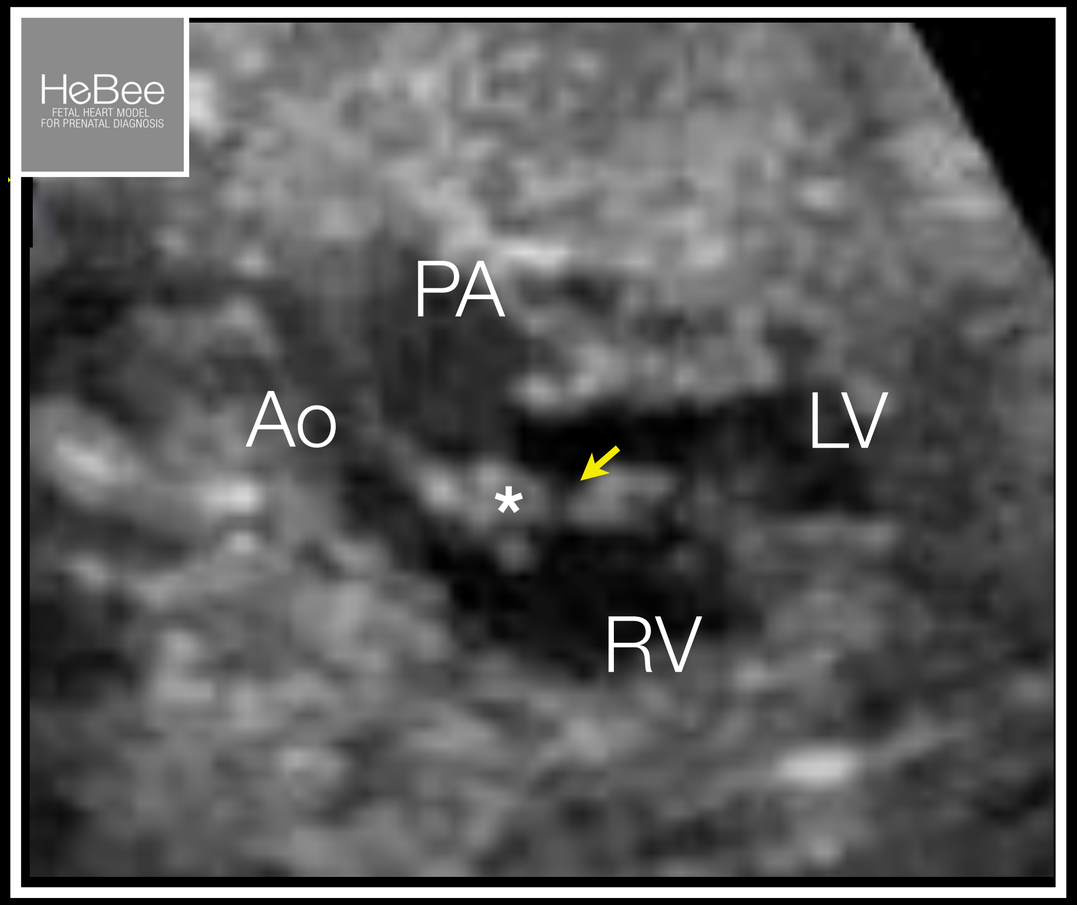


TGA-VSD-PS

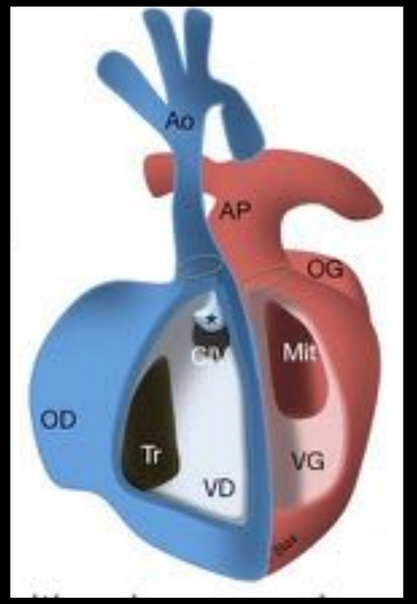


TGA

Complex forms

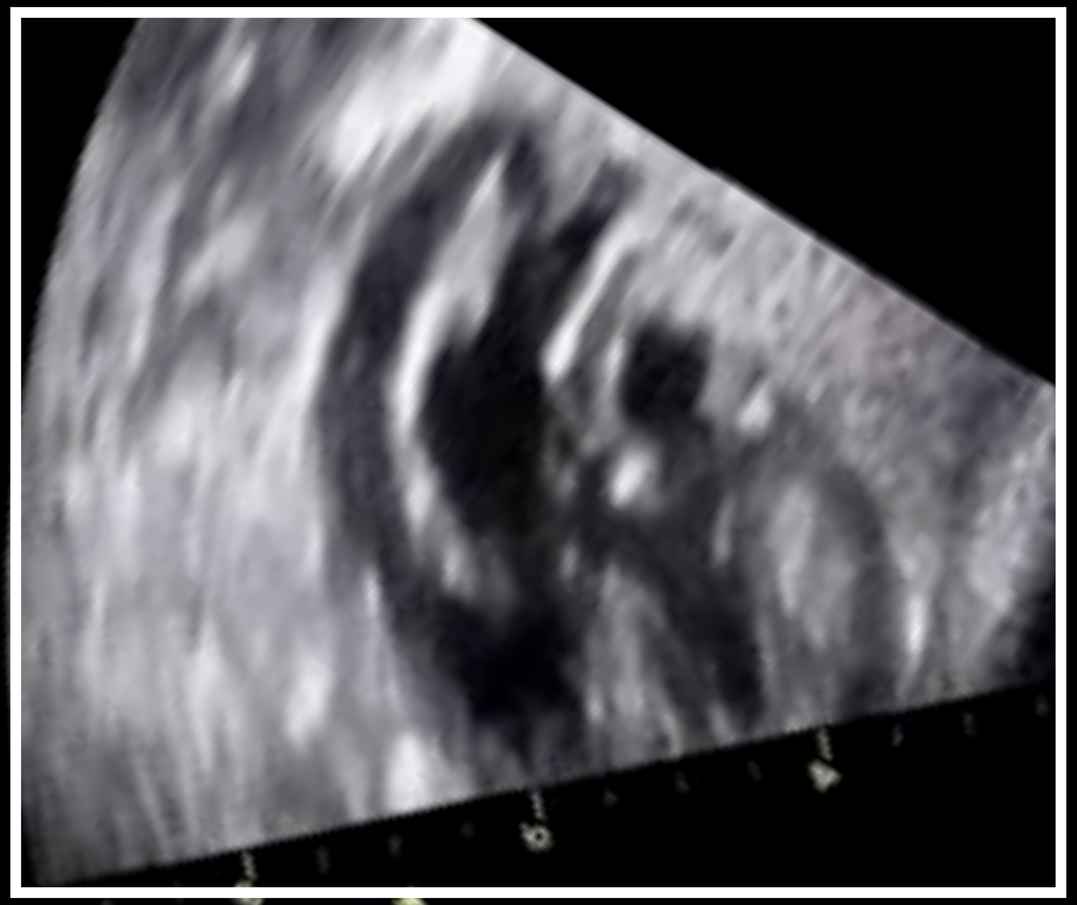


TGA-VSD-Coa

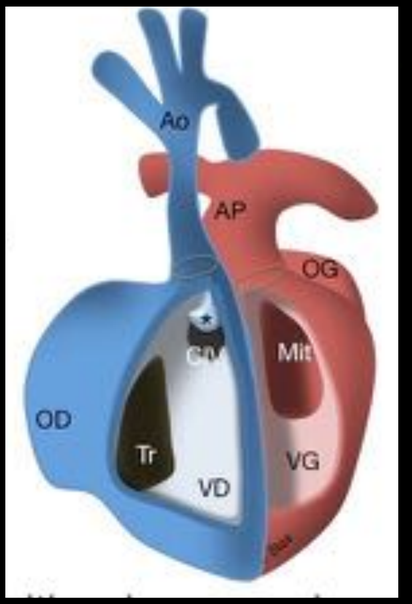


TGA

Complex forms

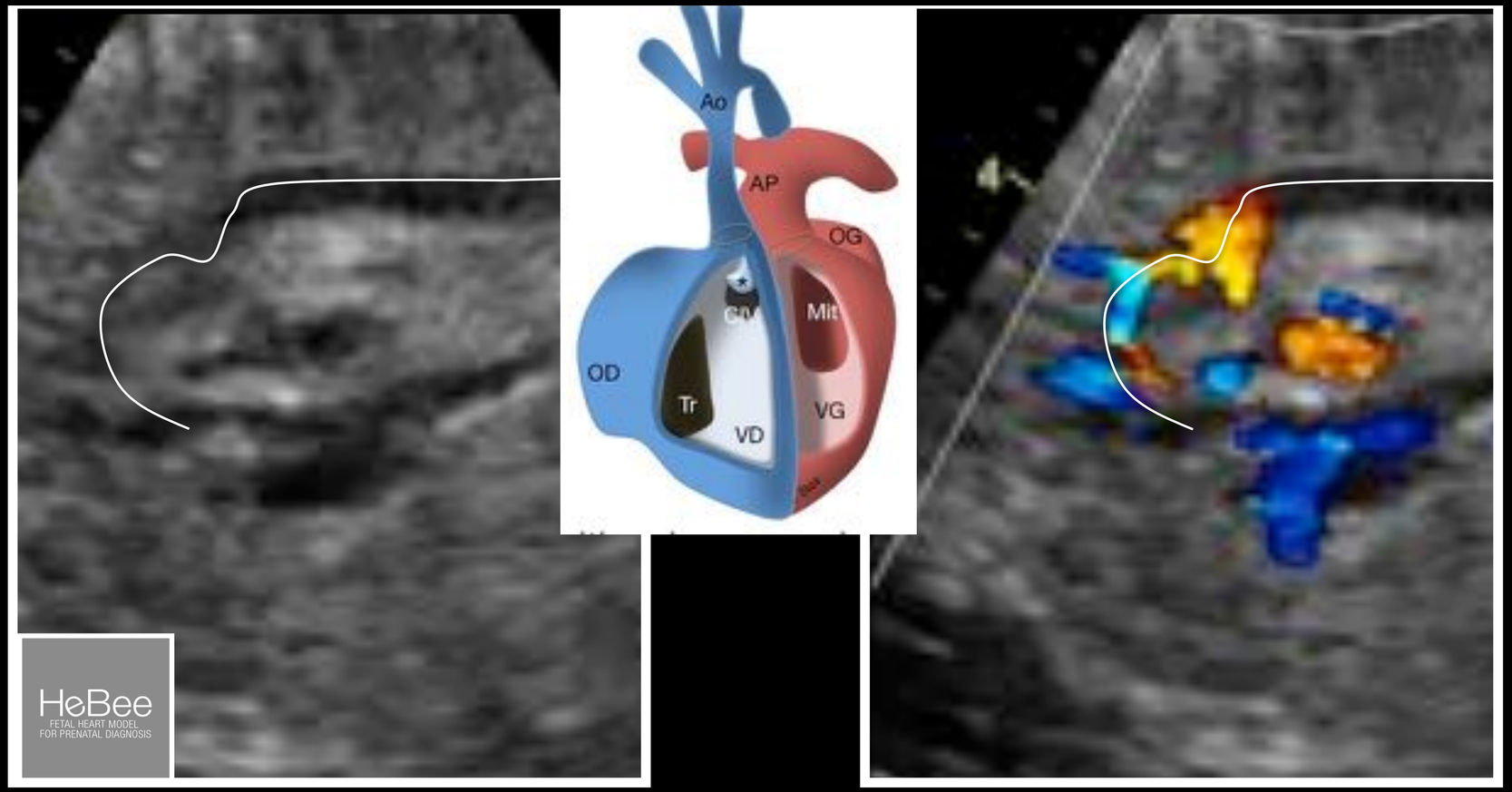


TGA-VSD-Coa



TGA

Complex forms

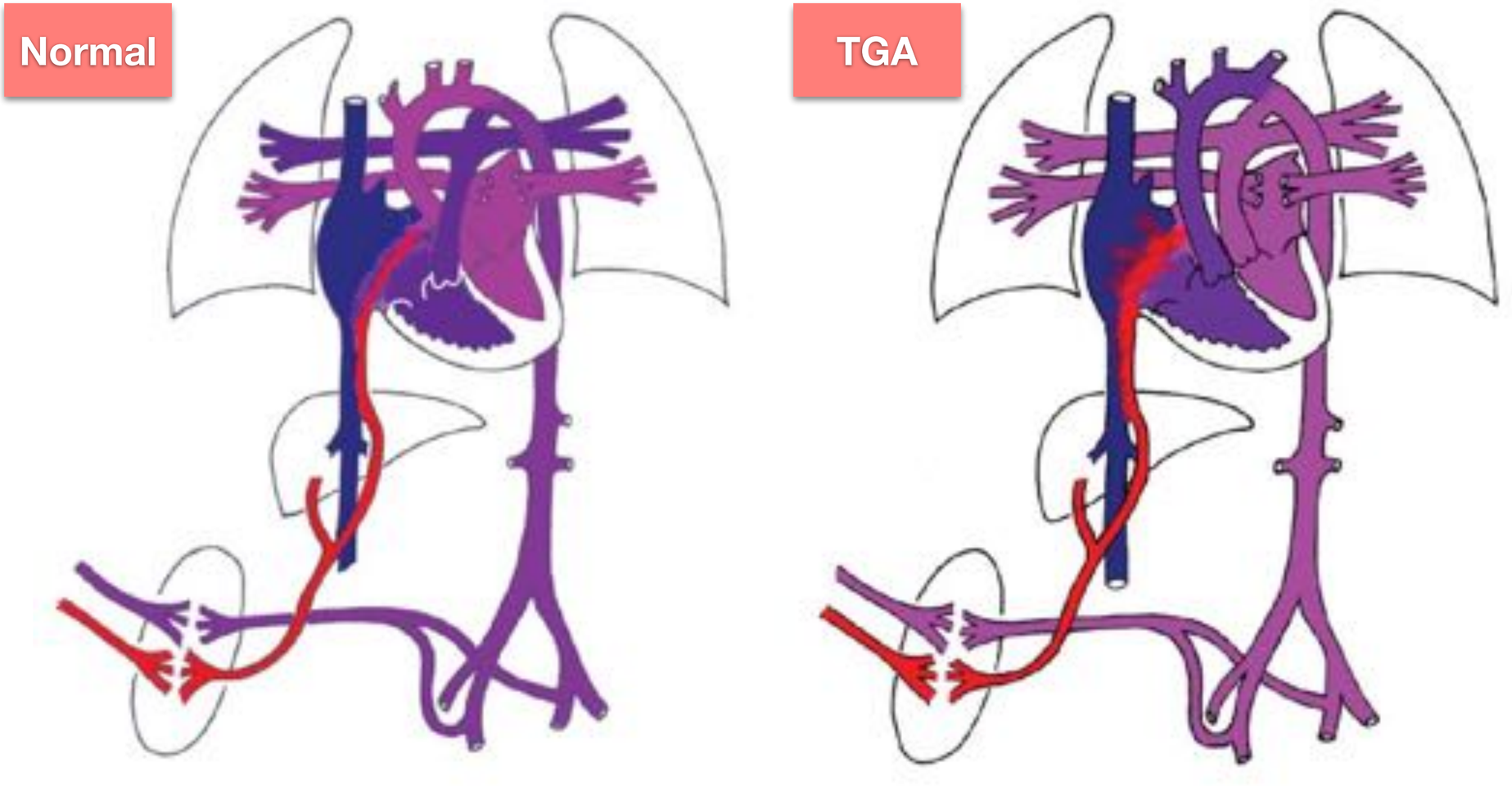


Chromosomal anomalies in fetal CHD

548 CHD-18.5%

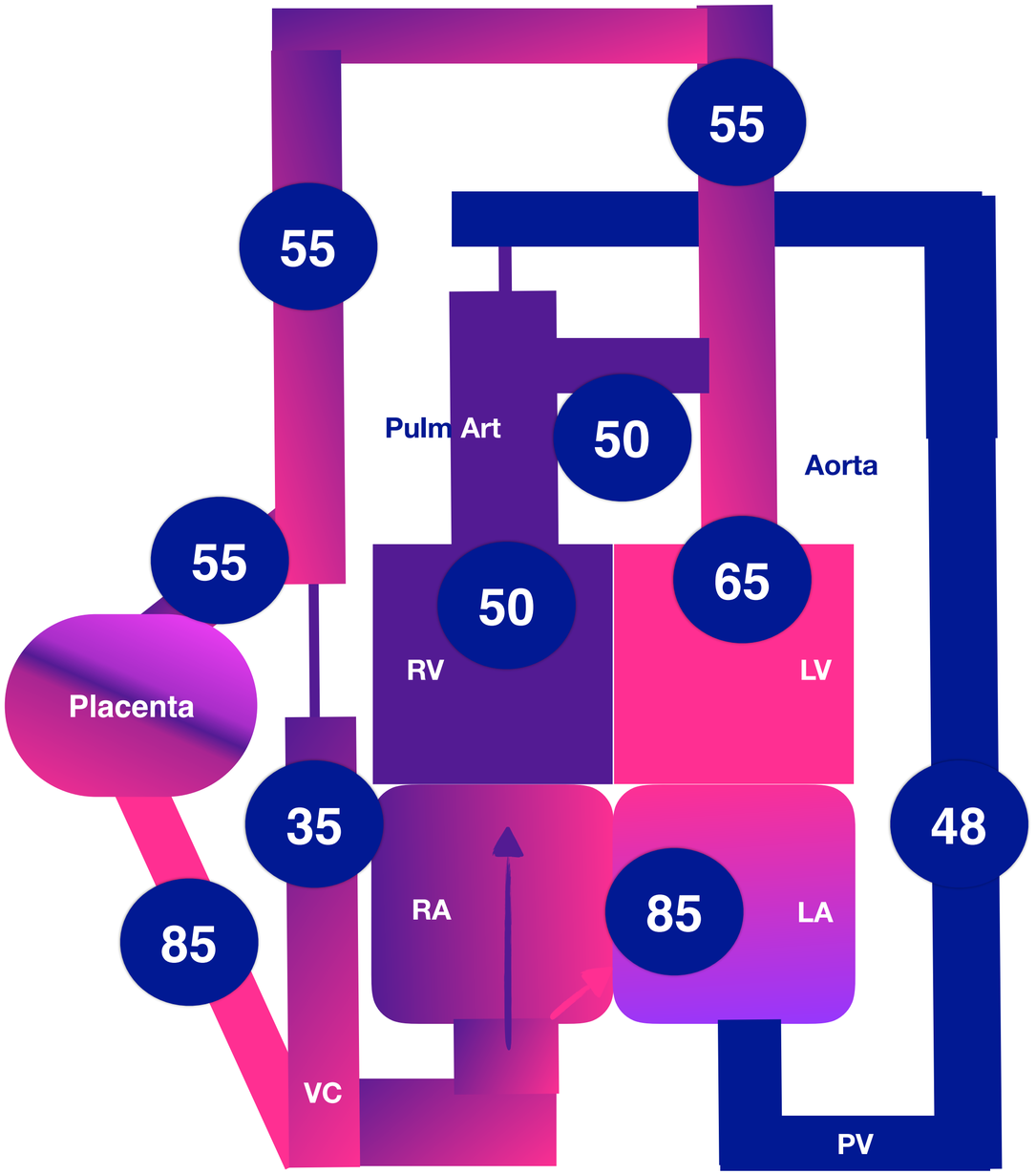
PA-IVS and PS	0	0
Left heart obstruction	12/130	9.2%
6 XO; 3 T18; 3 translocations		
Conotruncal defects	23/91	25%
20 del22q11; 1 T21; 2 translocations		
AVSD	32/68	47%
28 T21; 3 T18; 1 XXX		
VSD	12/74	16%
9 trisomies, 2 del22q11, 1 del5		
Transposition of the great vessels	0	0
DORV	7/38	18%
Univentricular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

Morphological and physiological consequences of the fetal circulation in TGA

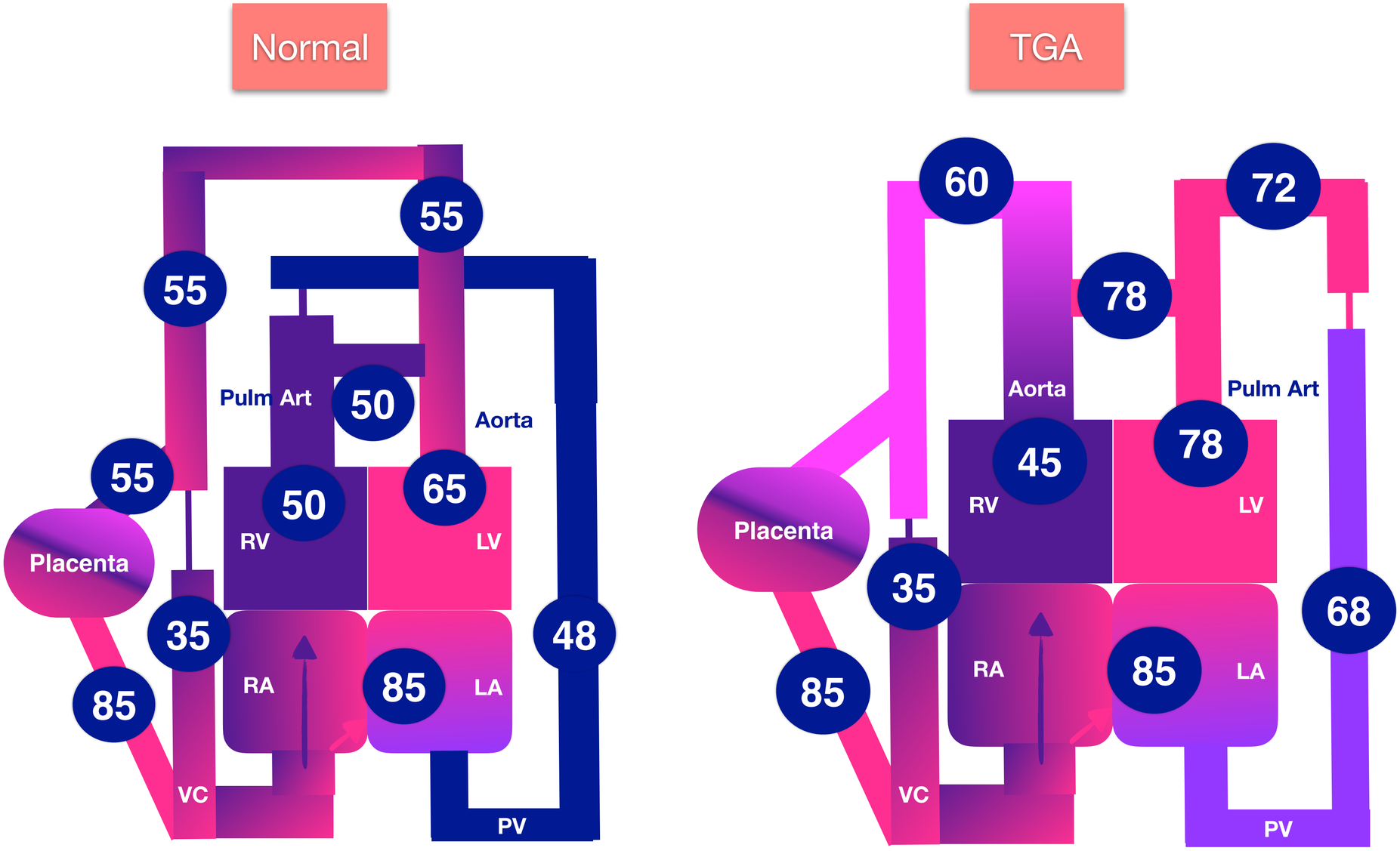


Sun L et al. Circulation. 2015;131:1313-1323.

Morphological and physiological consequences of the fetal circulation in TGA



Morphological and physiological consequences of the fetal circulation in TGA

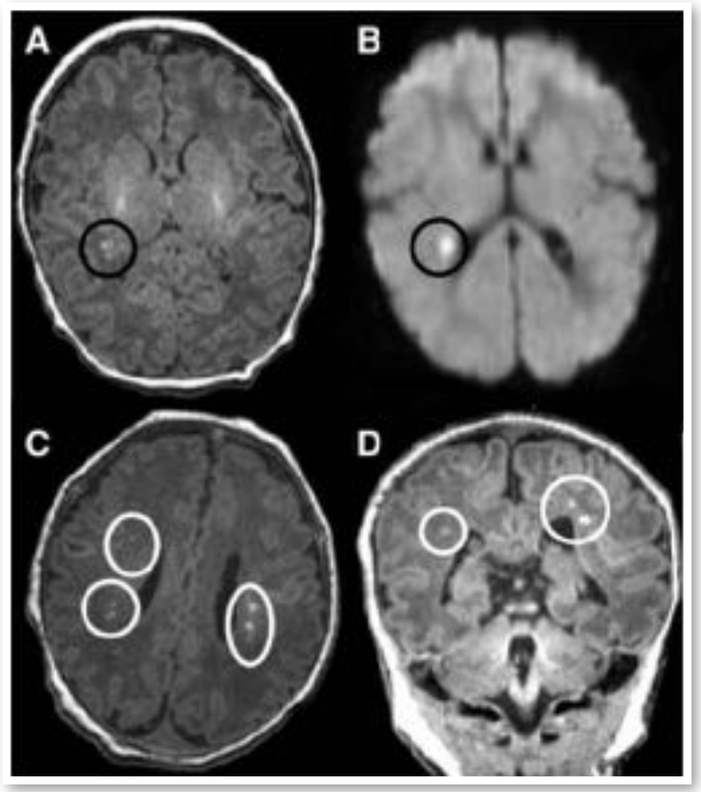


Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

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

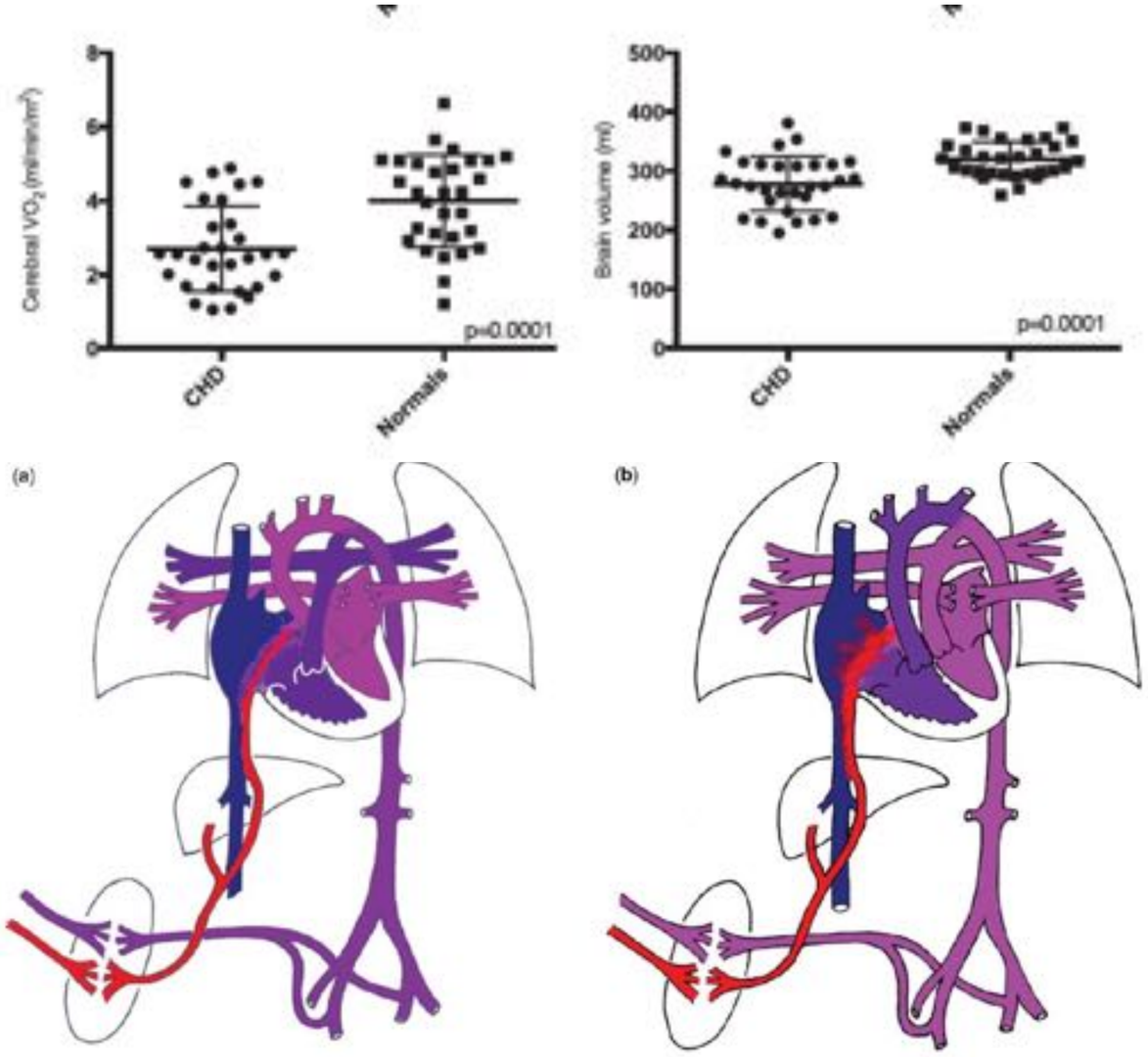
- **White matter lesions in** 30 to 40% of newborns with TGA (Miller et al., 2004; Licht et al., 2009)

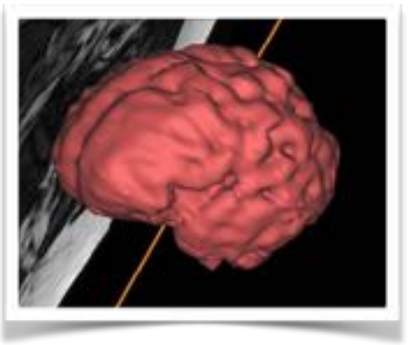
- Same type of anomalies but more severe in complex CHDs such as HLHS (Mahle et al., 2002).



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

Type of CHD and prenatal brain perfusion



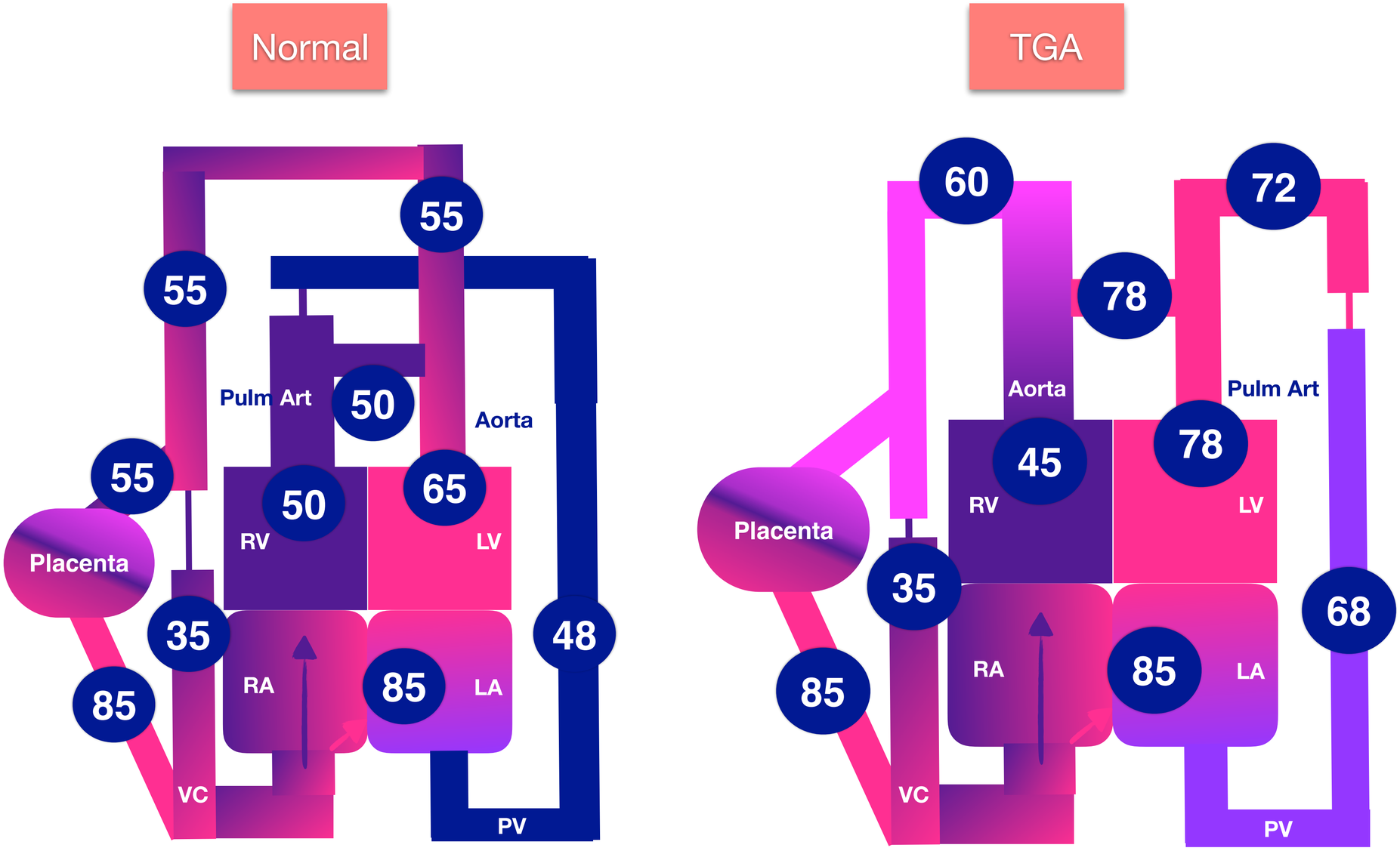


Mechanisms for reduced cerebral oxygenation and impaired brain growth in fetuses with CHD

1-In TGA, streaming results in well oxygenated blood being directed to the pulmonary circulation, whereas the blood supplied to brain is derived largely from more deoxygenated blood returning from the caval veins.
2-Reduction in Umbilical Vein Sao2, which is suggestive of abnormal placental function and results in lower fetal O2 delivery even in the setting of normal CVO and UV flow.

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

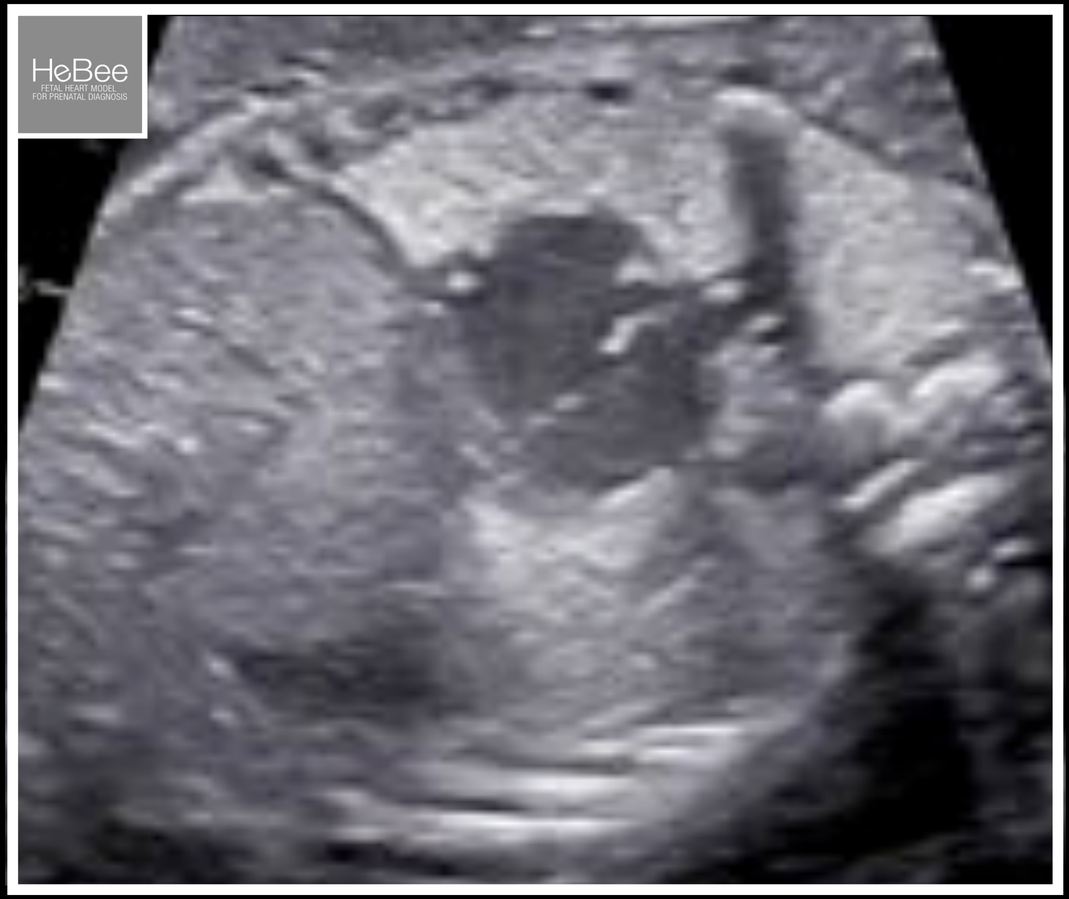
Morphological and physiological consequences of the fetal circulation in TGA



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

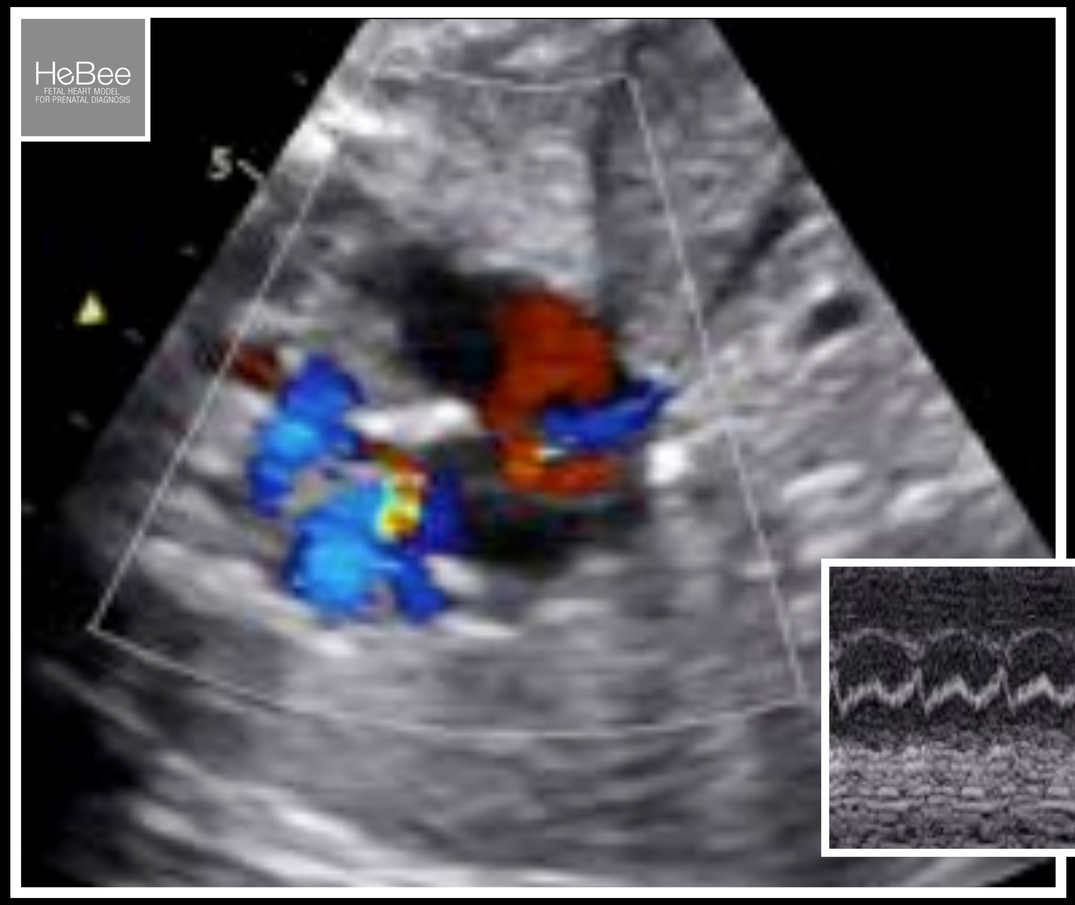
TGA

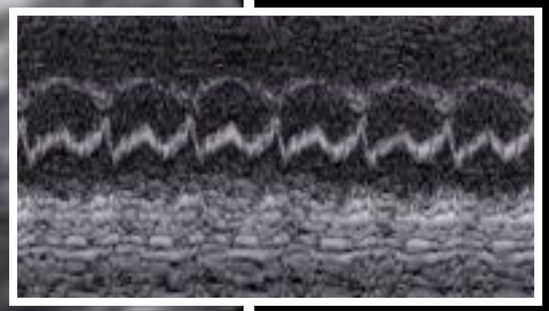
Foramen Ovale



TGA

Foramen Ovale

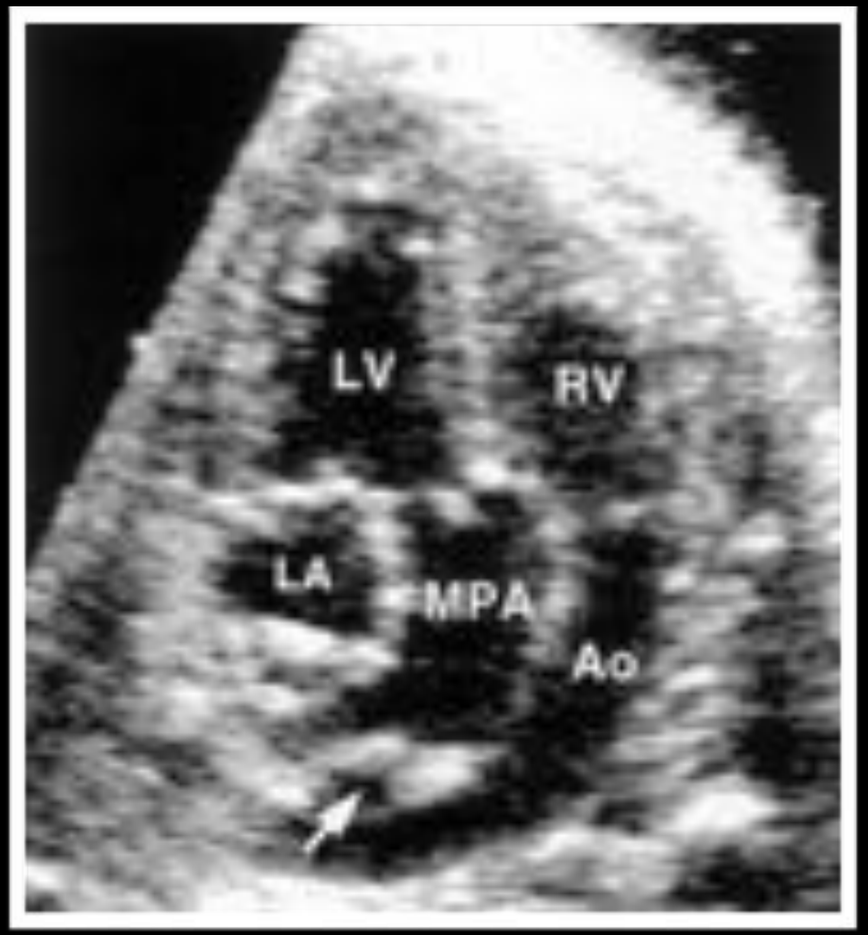


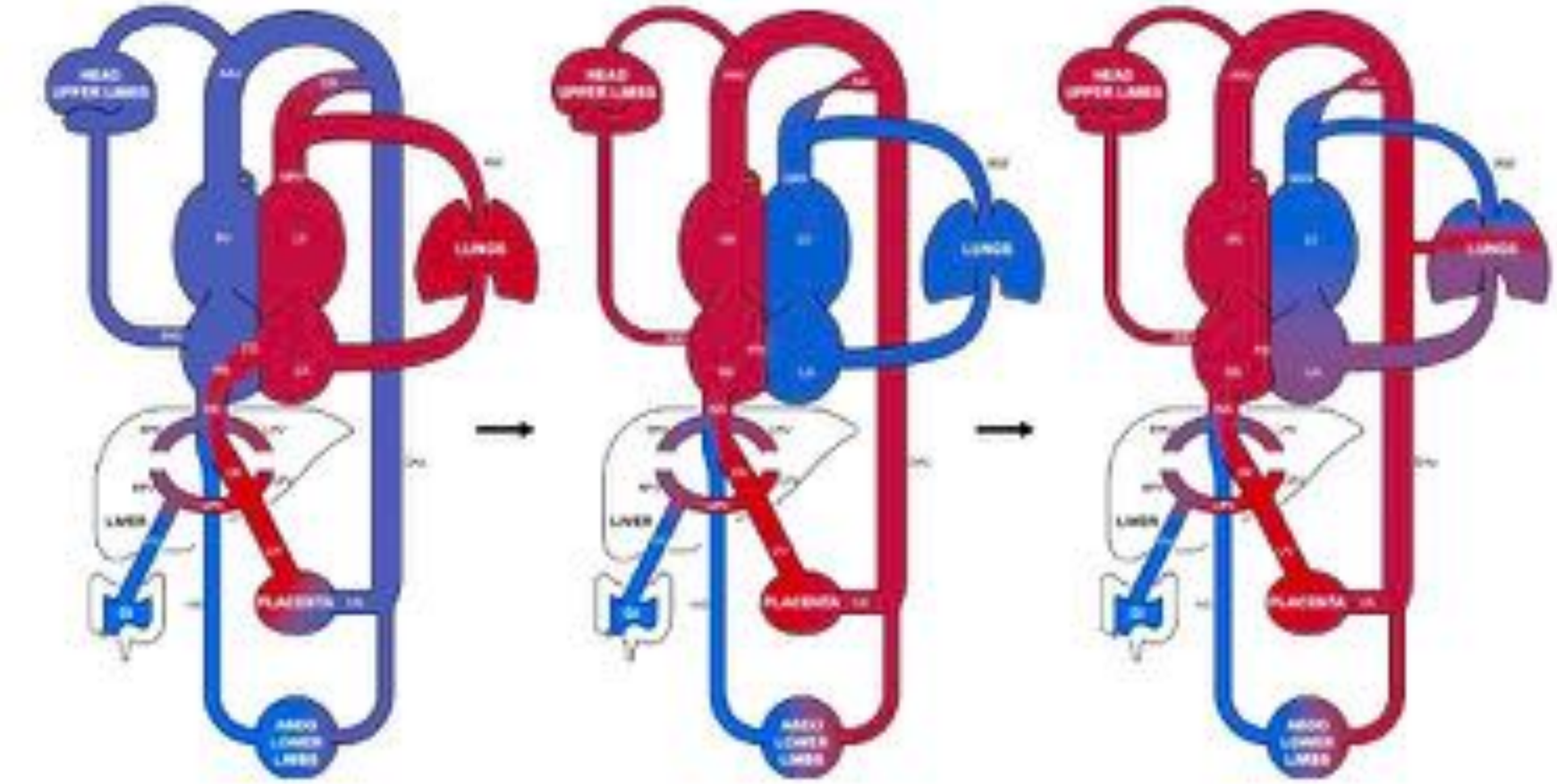


-Floppy Mb
-Small FO
-Small septal length
-pulmonary veins velocity > 0,41 m/s

TGA

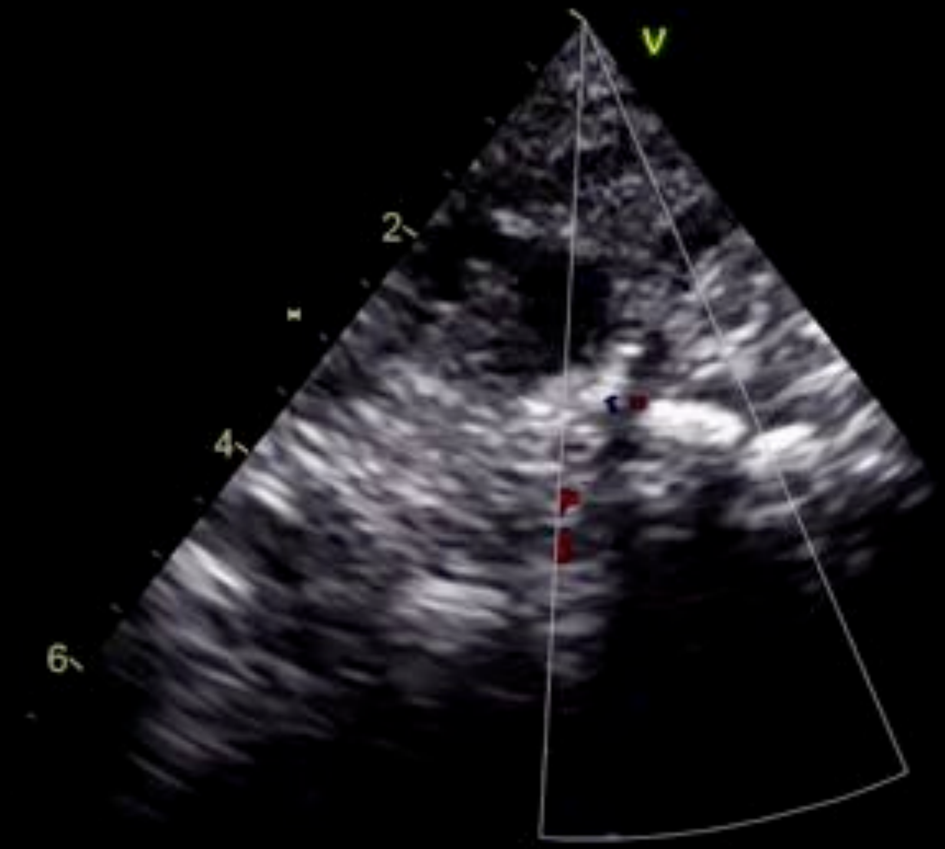
Arterial duct

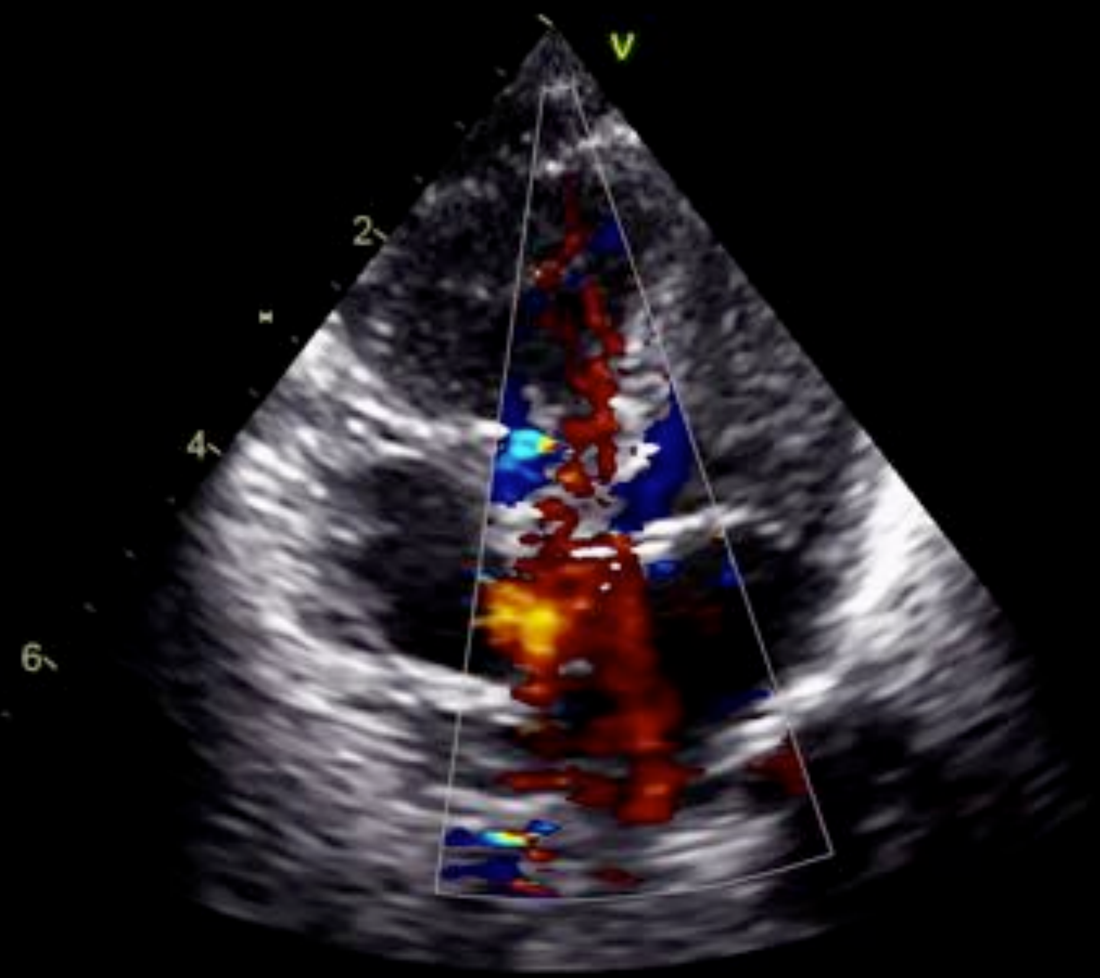




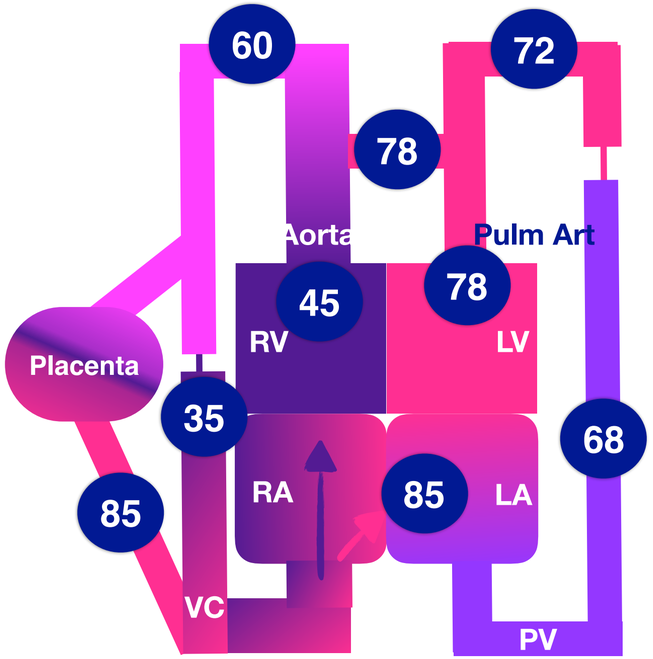
Initial increase in PBF due to vasodilatation with increased oxygen
Increased pulmonary venous return
Reduced size of the FO
Ductal constriction due to oxygen
Isolation of Pulmonary circulation
Increased PVR
Development of aorta-pulmonary collaterals

PDA and Collaterals in TGA

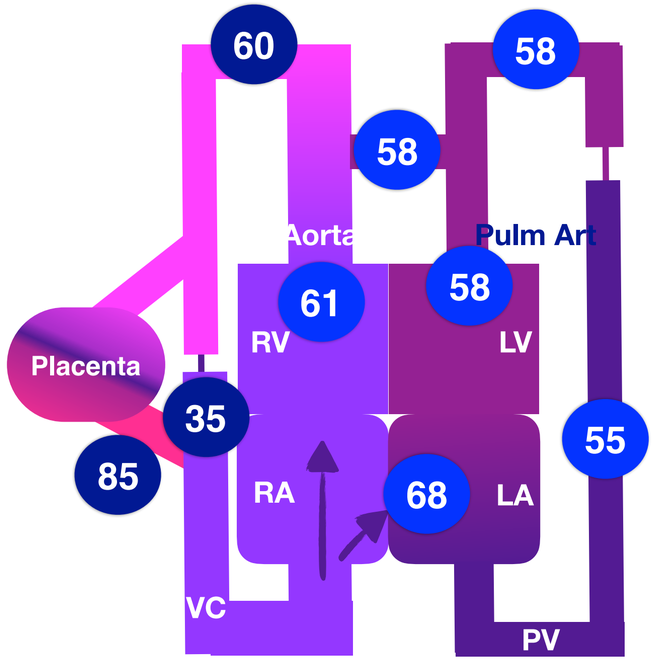




Morphological and physiological consequences of the fetal circulation in TGA



TGA fetus

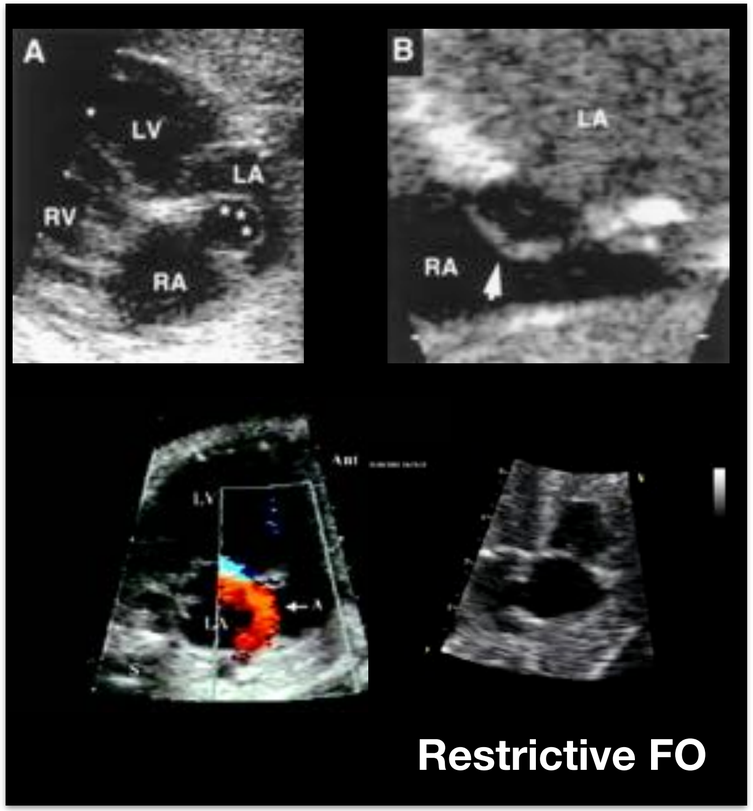


Closure of the ductus venous

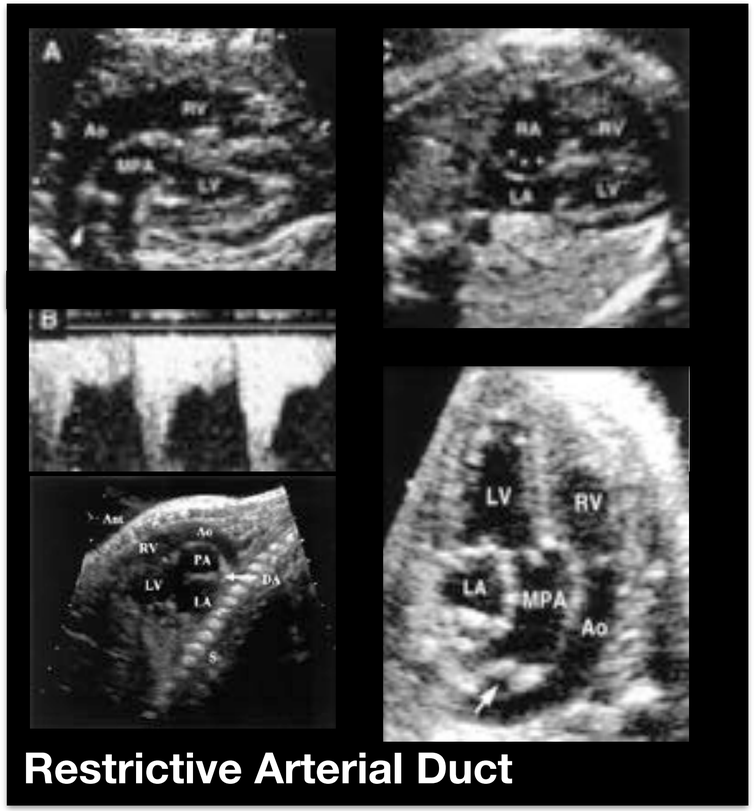
Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

Prenatal diagnosis of transposition of the great arteries

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, pregnancy termination, perinatal and early neonatal mortality for selected (isolated) congenital heart anomalies

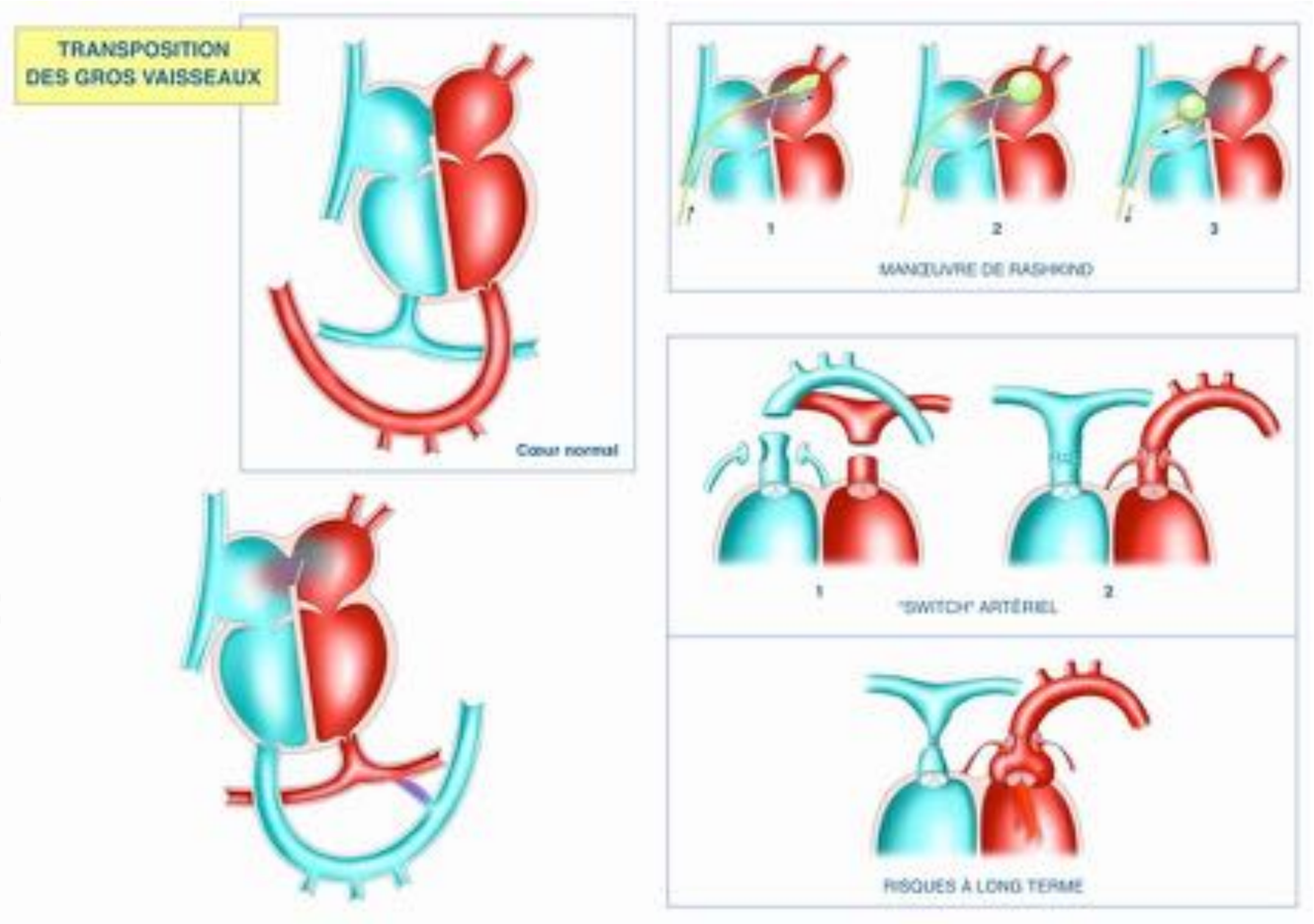
Paris Registry of Congenital Malformations, 1983-2000

	1983 - 1988	1989 - 1994	1995 - 2000	
	%	%	%	p
TGA				
Prenatal Diagnosis	12.5	48.1	**72.5**	0.001
Pregnancy Termination	0	7.4	0	0.62
First Week Mortality	18.8	8.3	2.6	0.04
Perinatal Mortality	23.5	12.0	5.0	0.02
HLHS				
Prenatal Diagnosis	31.8	82.8	**88.9**	< 0.001
Pregnancy Termination	13.6	72.4	63.0	< 0.001
First Week Mortality	83.3	75.0	50.0	0.12
Perinatal Mortality	84.2	75.0	50.0	0.10

Prenatal diagnosis of transposition of the great arteries

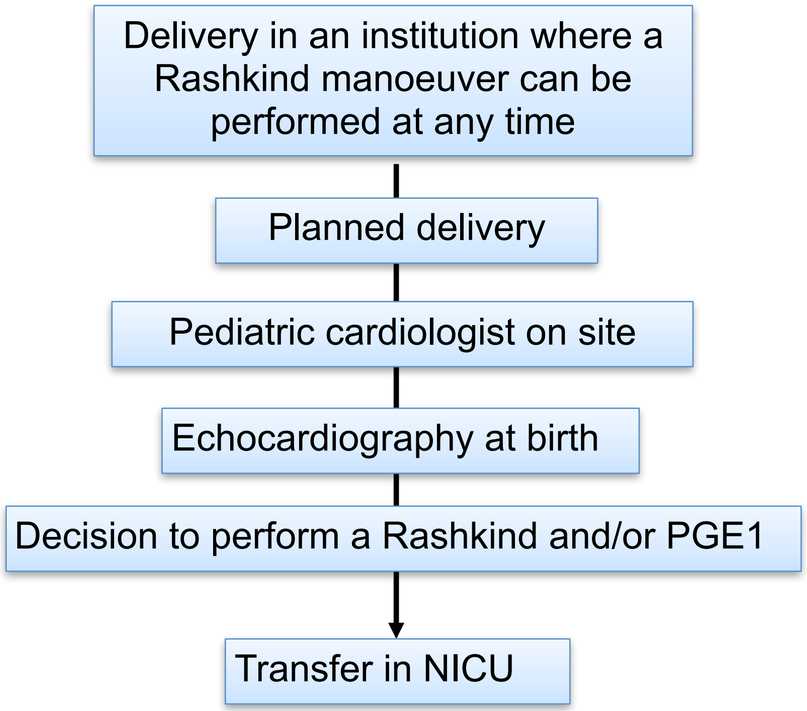
Perinatal organization in Paris

- 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



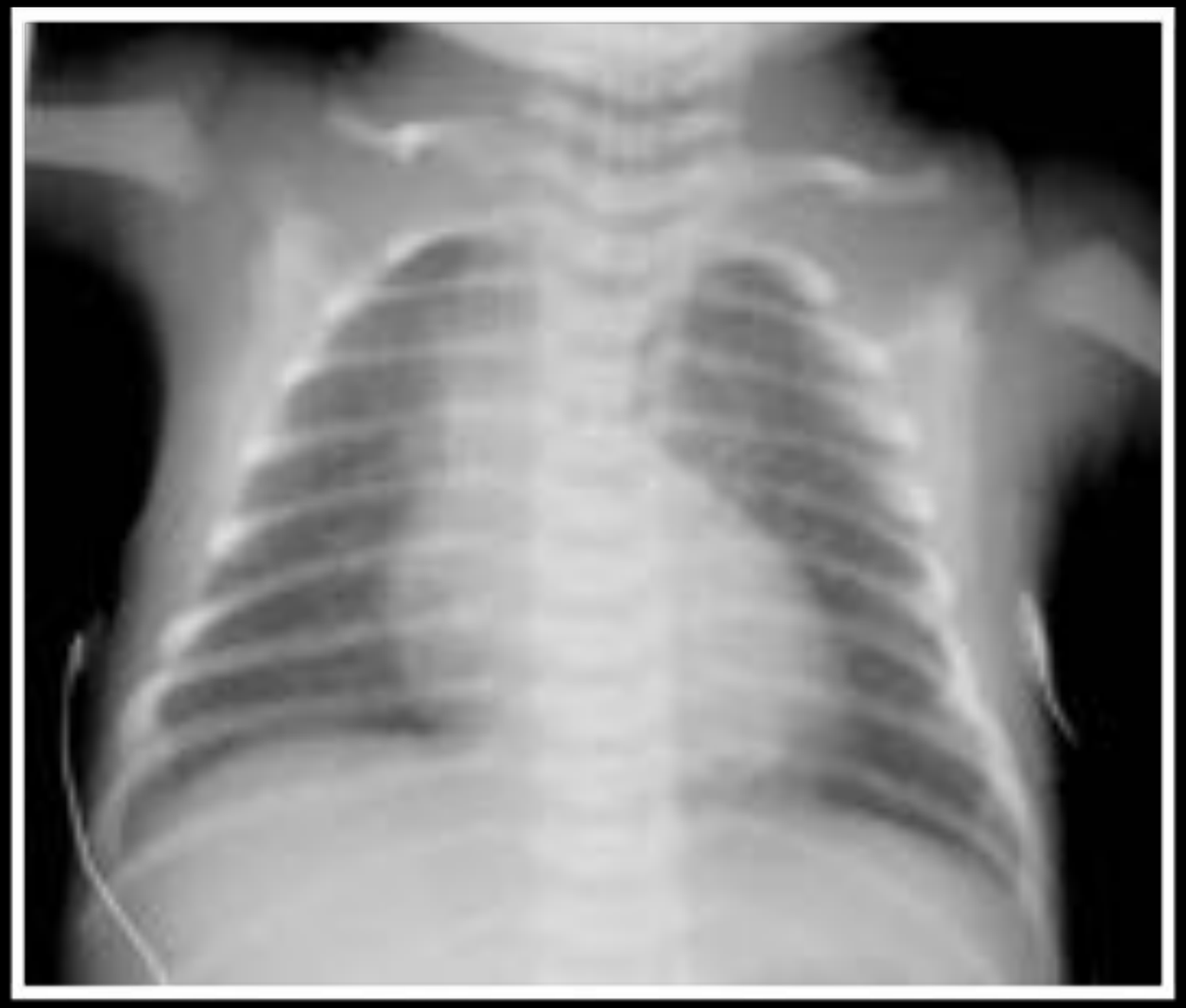
Bonnet et al. Circulation 1997; Maeno YV et al. Circulation 1999; Jouannic et al. Circulation 2004; Van Velzen C et al. Ultrasound Obstet Gynecol 2015; 45: 320–325
Escobar-Diaz MC et al. Ultrasound Obstet Gynecol. 2015 June ; 45(6): 678–682.

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

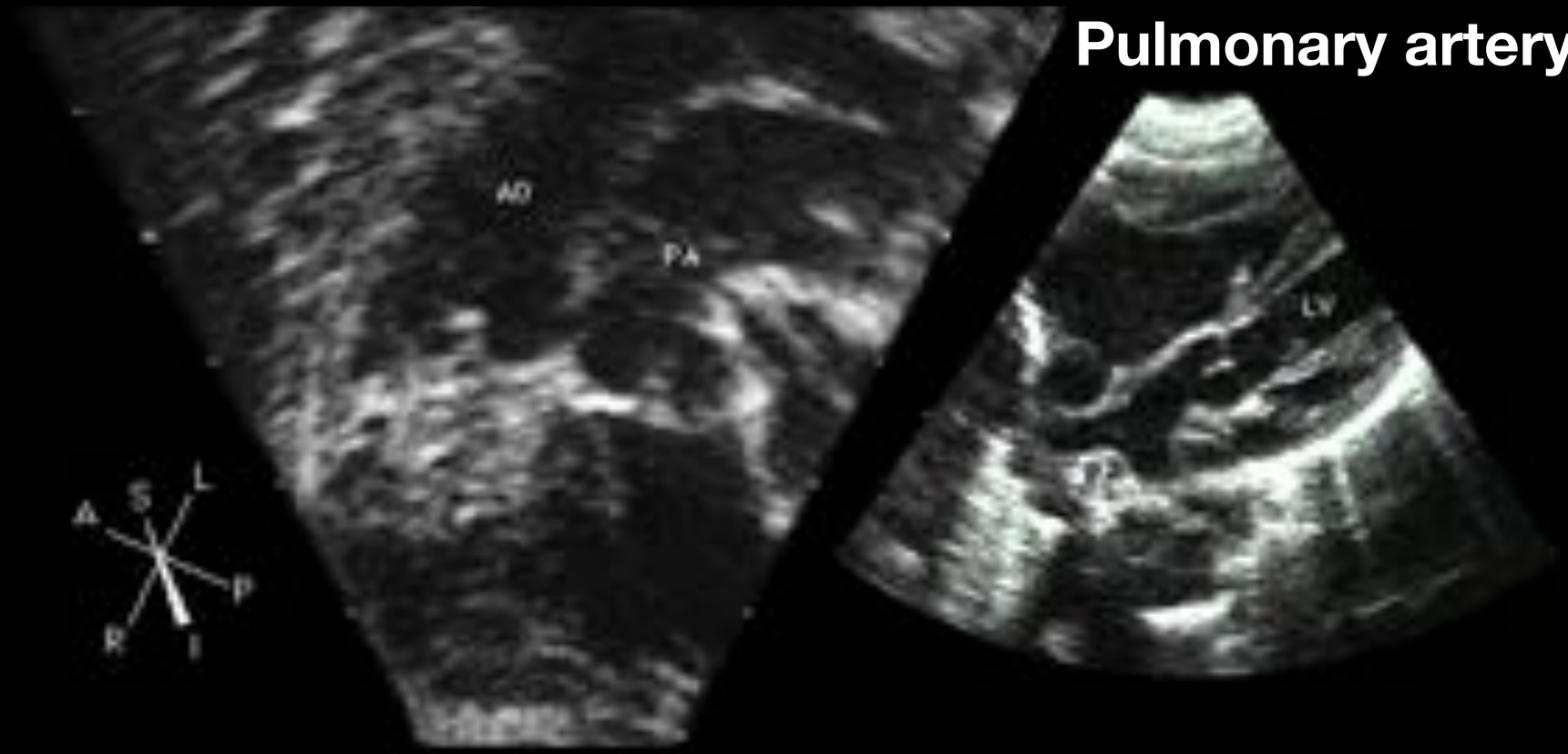
Neonatal diagnosis of TGA
Isolated cyanosis



TGA

Rapid diagnosis

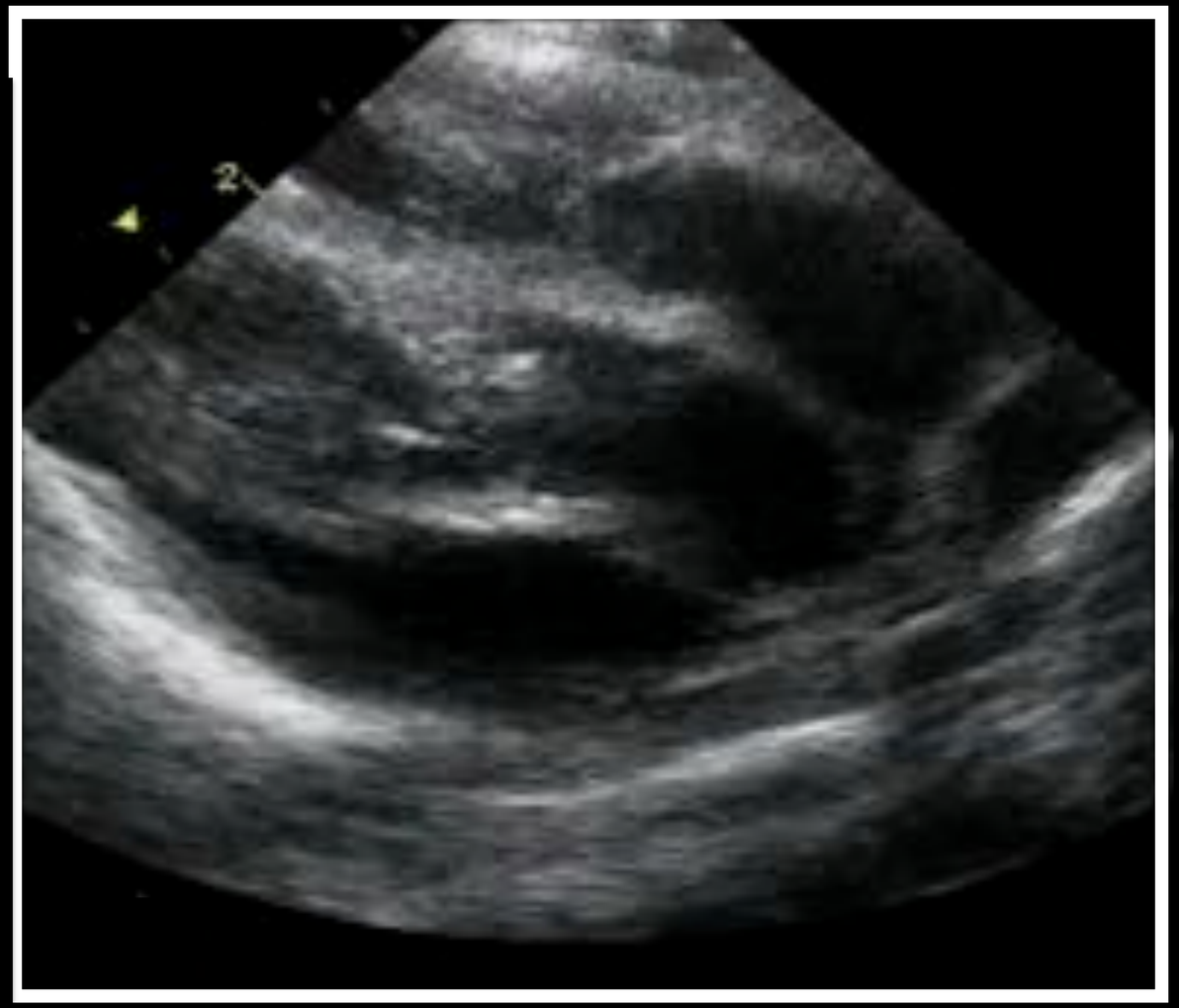
Parallel course of great vessels



Pulmonary artery from LV

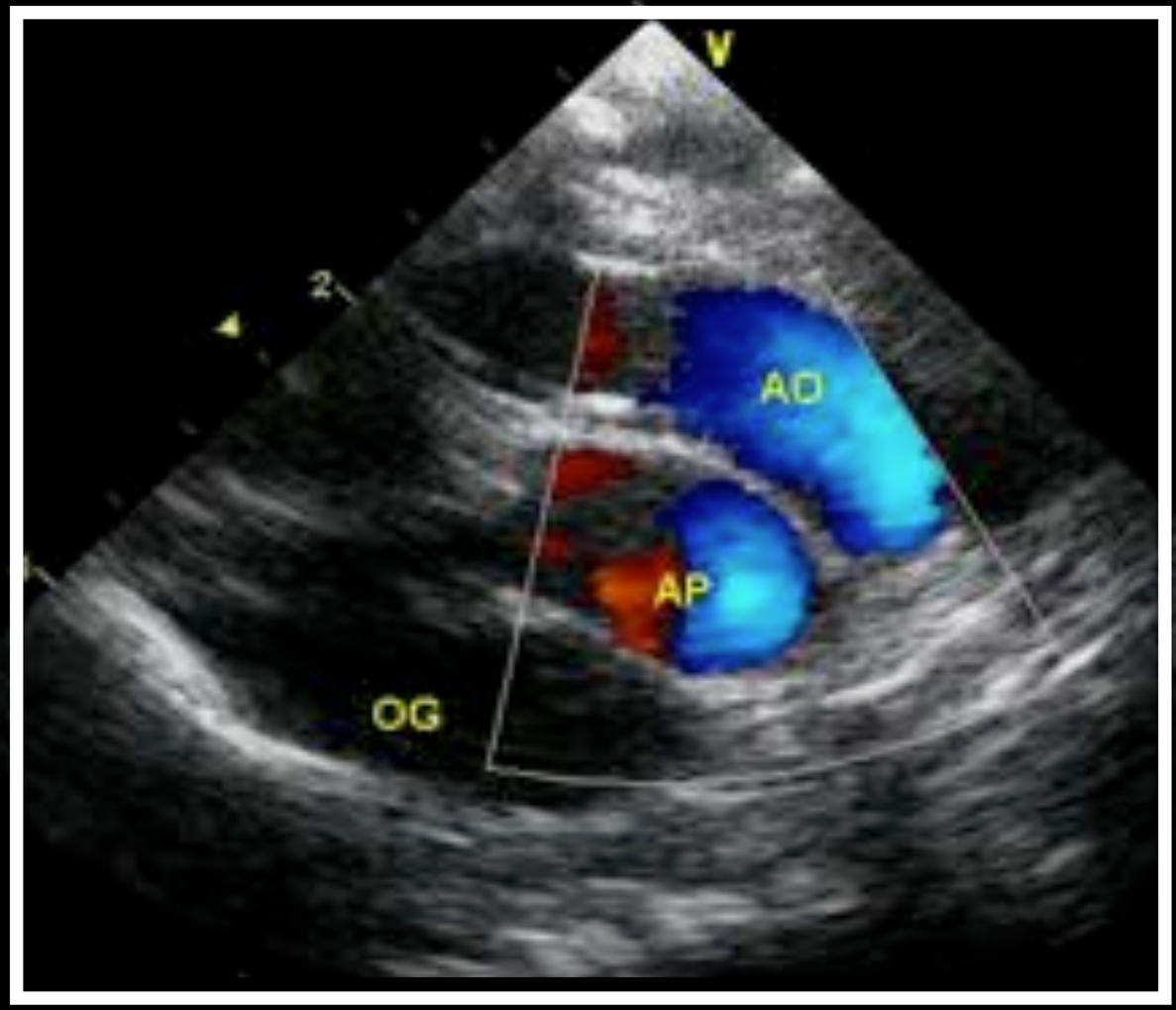
TGA

Rapid diagnosis



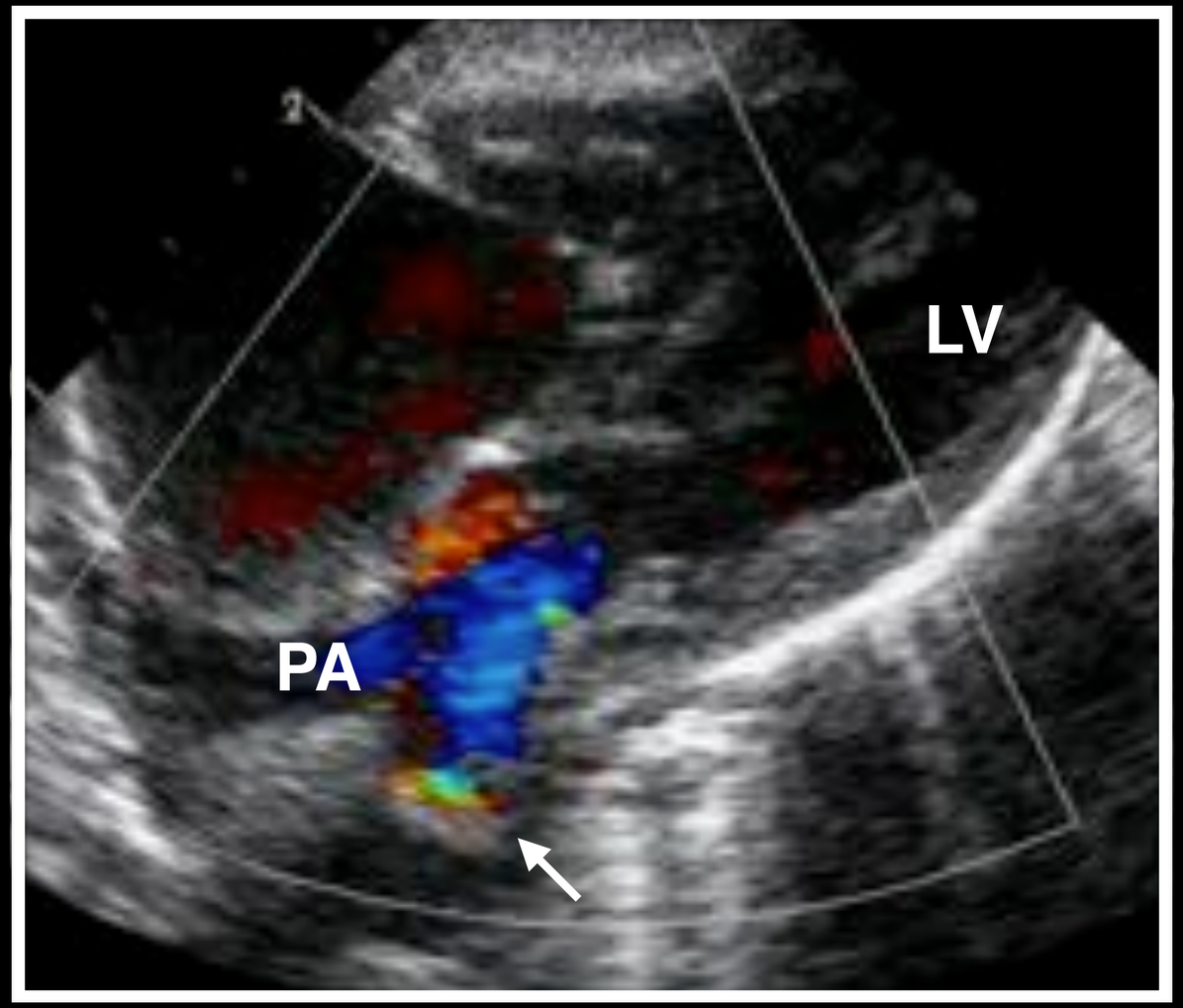
TGA

Rapid diagnosis



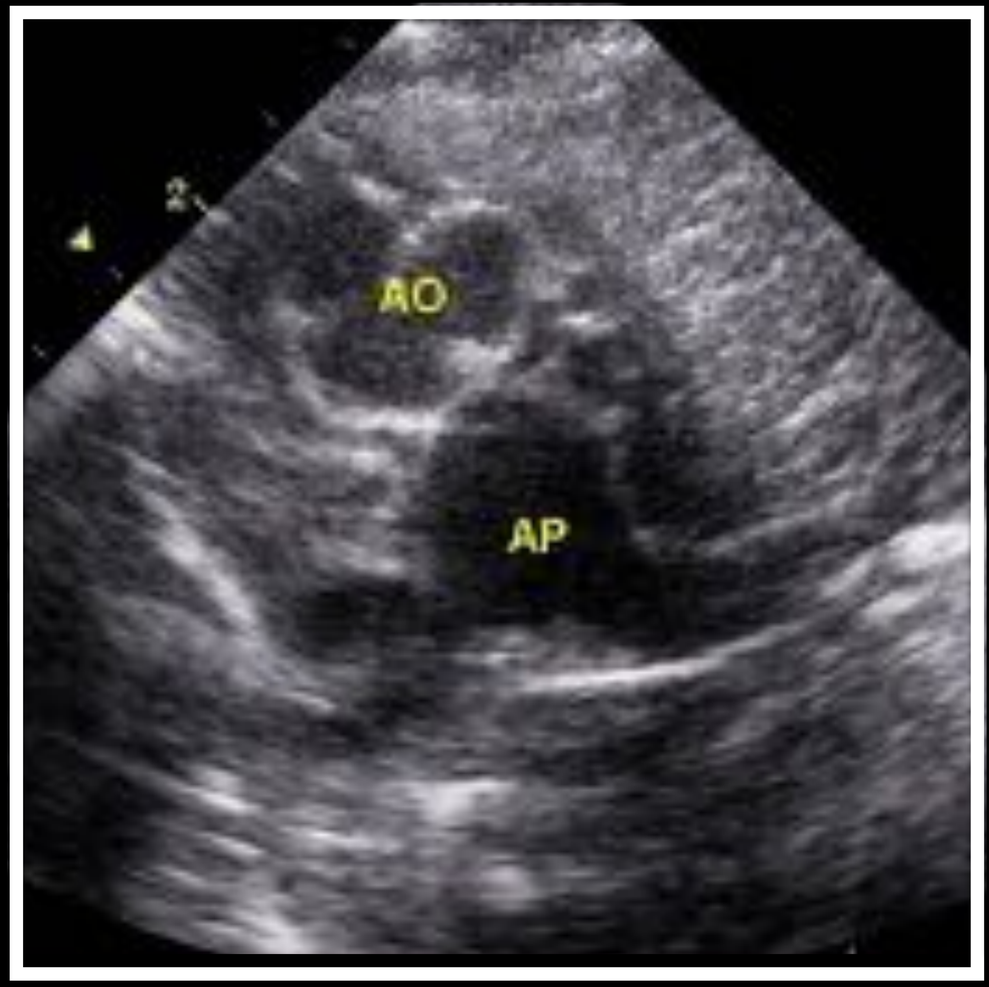
TGA

Rapid diagnosis



TGA

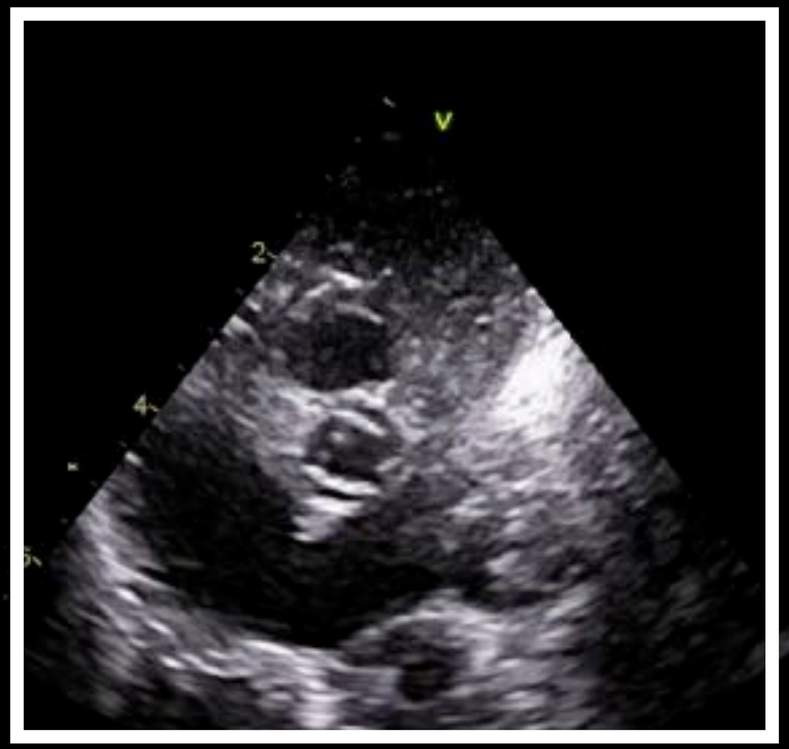
D-TGA

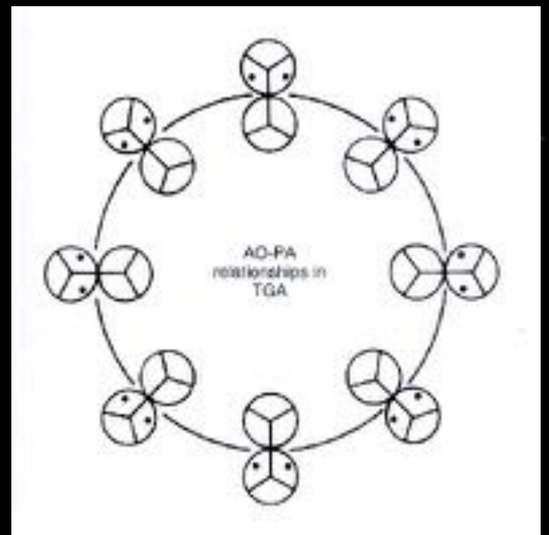


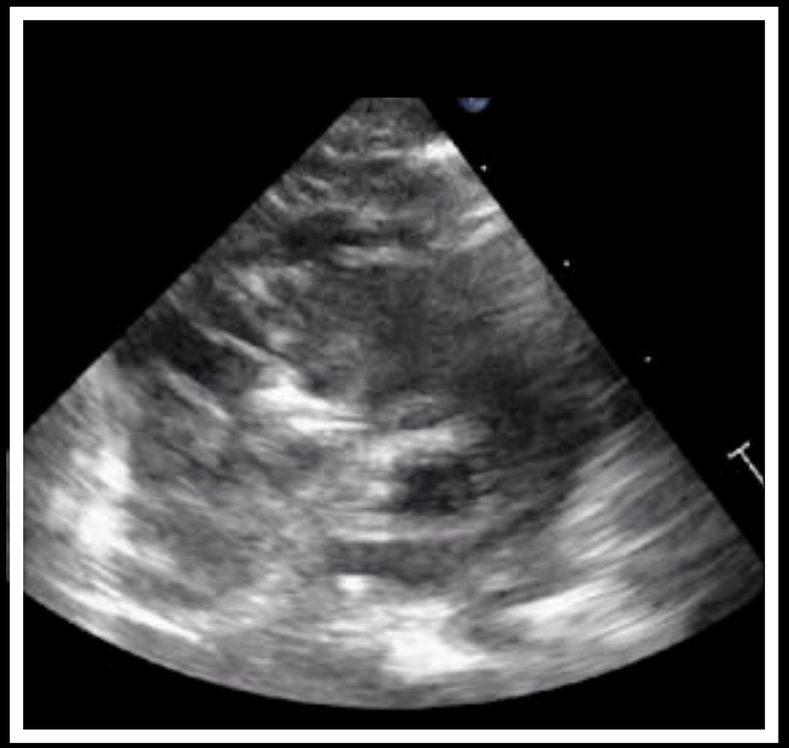
D-TGA

TGA

Aorto pulmonary vessels relationship and discrepancy

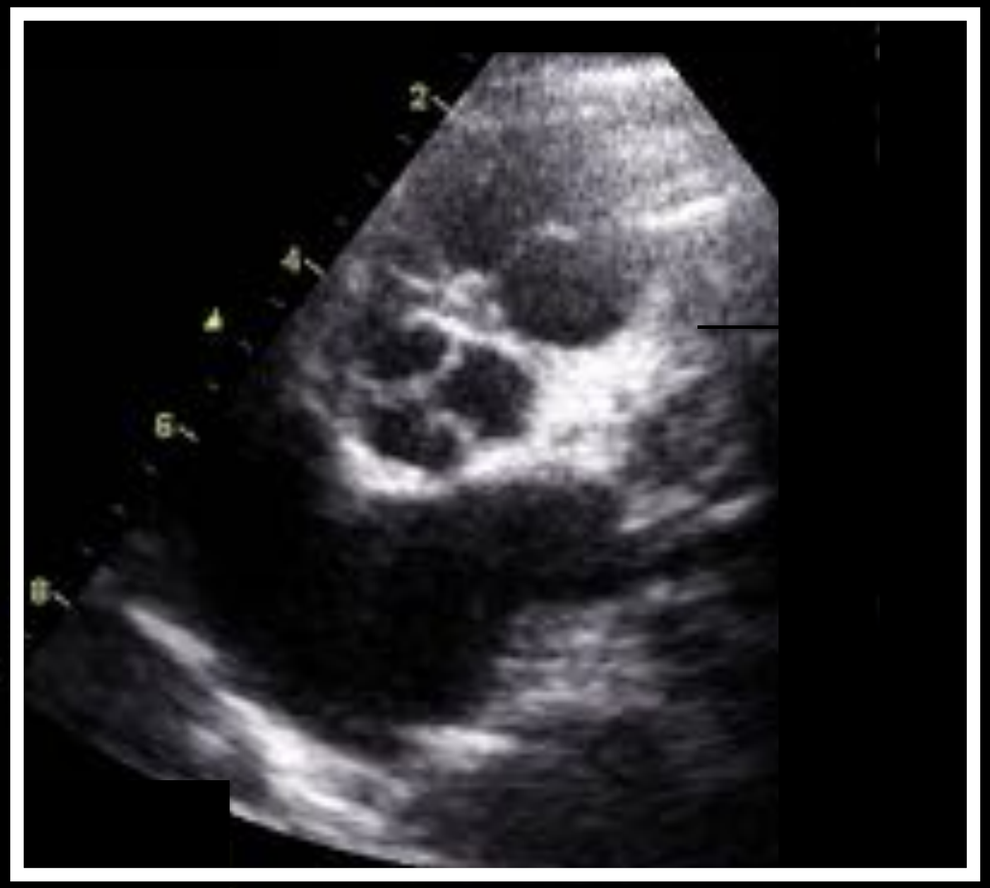




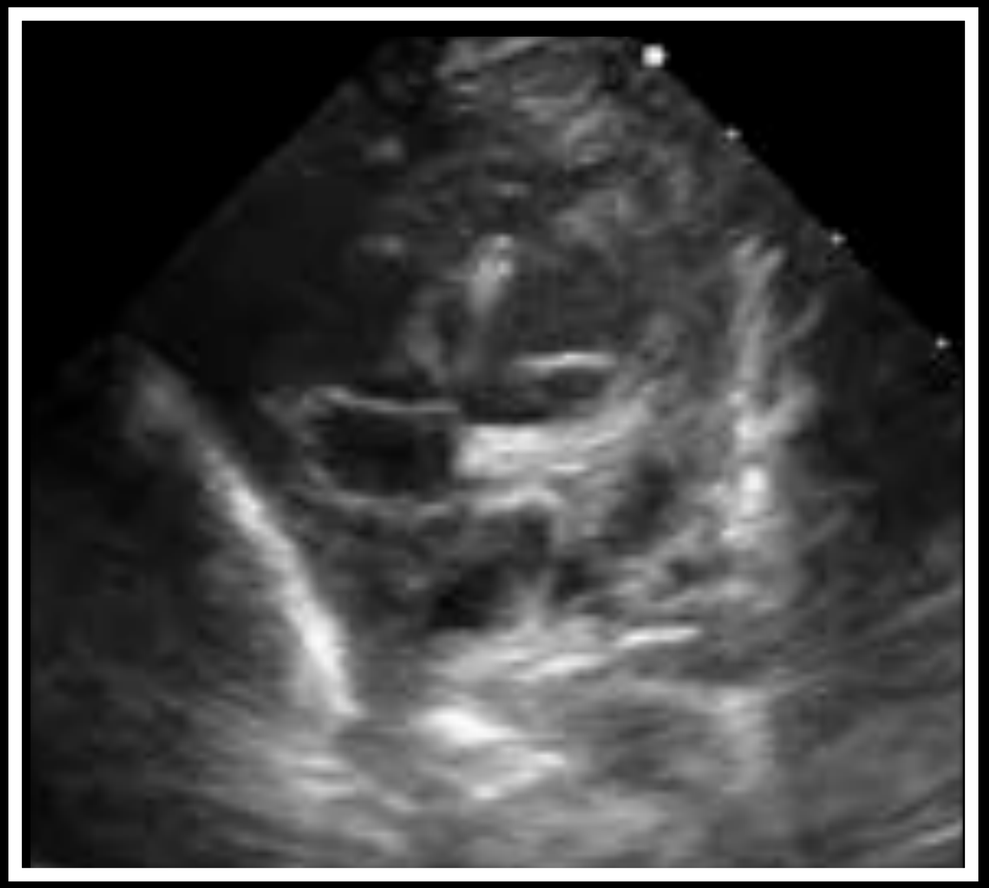


TGA

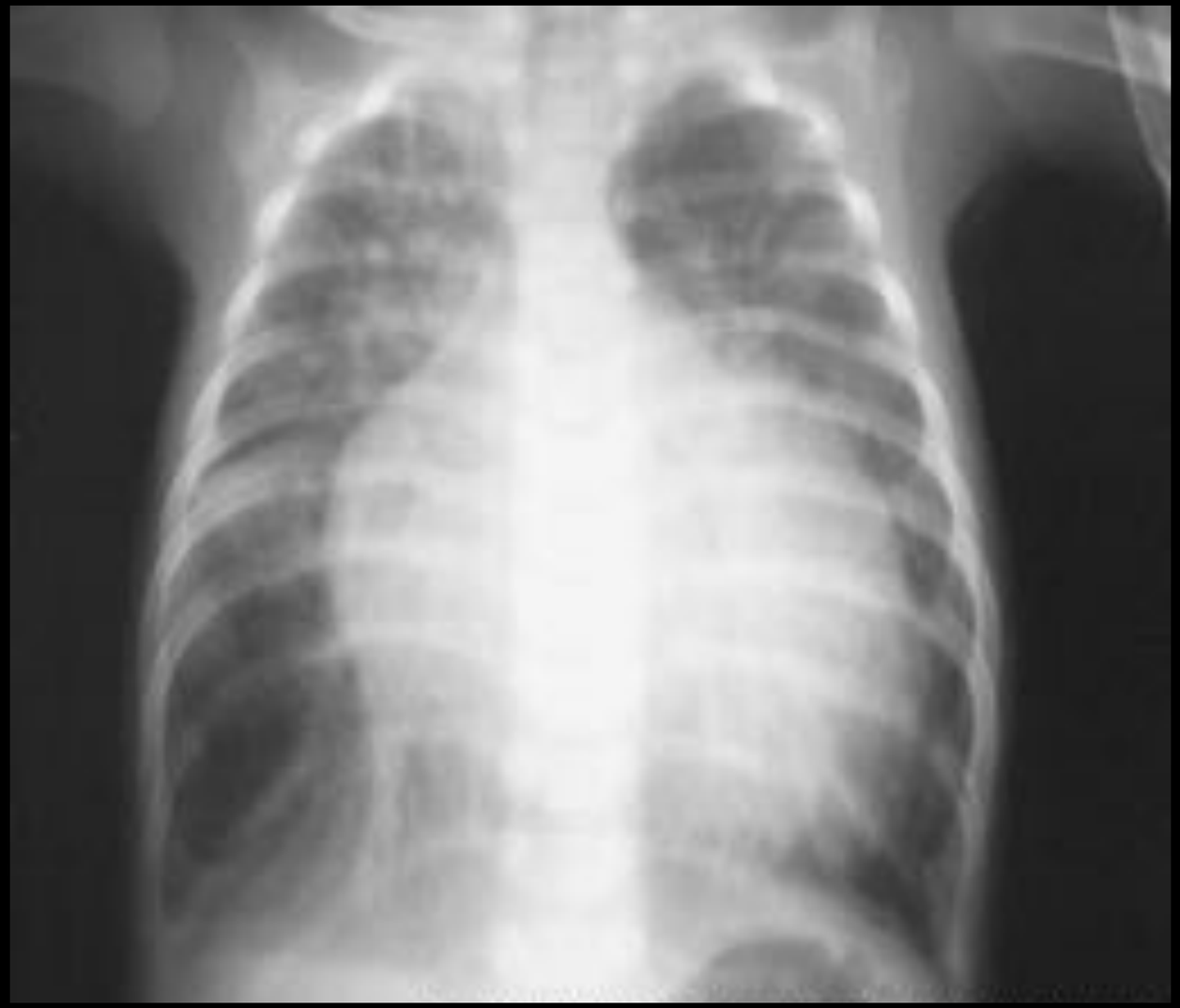
Relative position of the great vessels



L-TGA



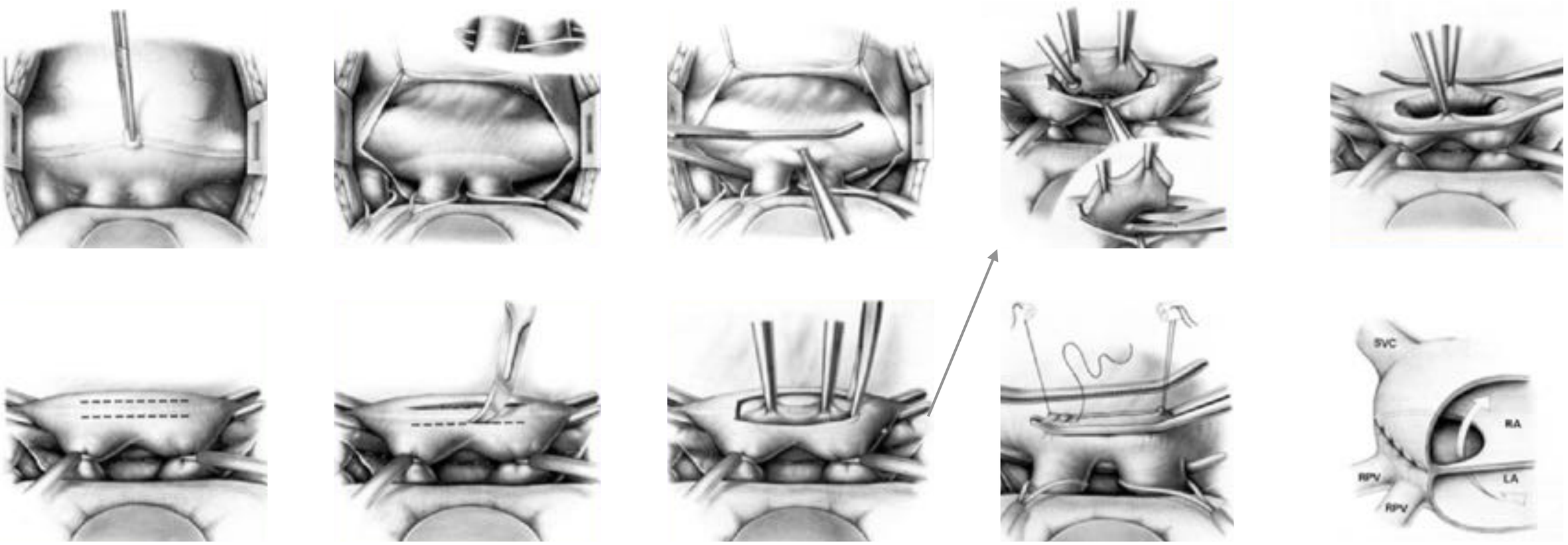
D-TGA with posterior aorta



TGA with heart failure and restrictive PFO

Alfred Blalock

Blalock-Hanlon procedure



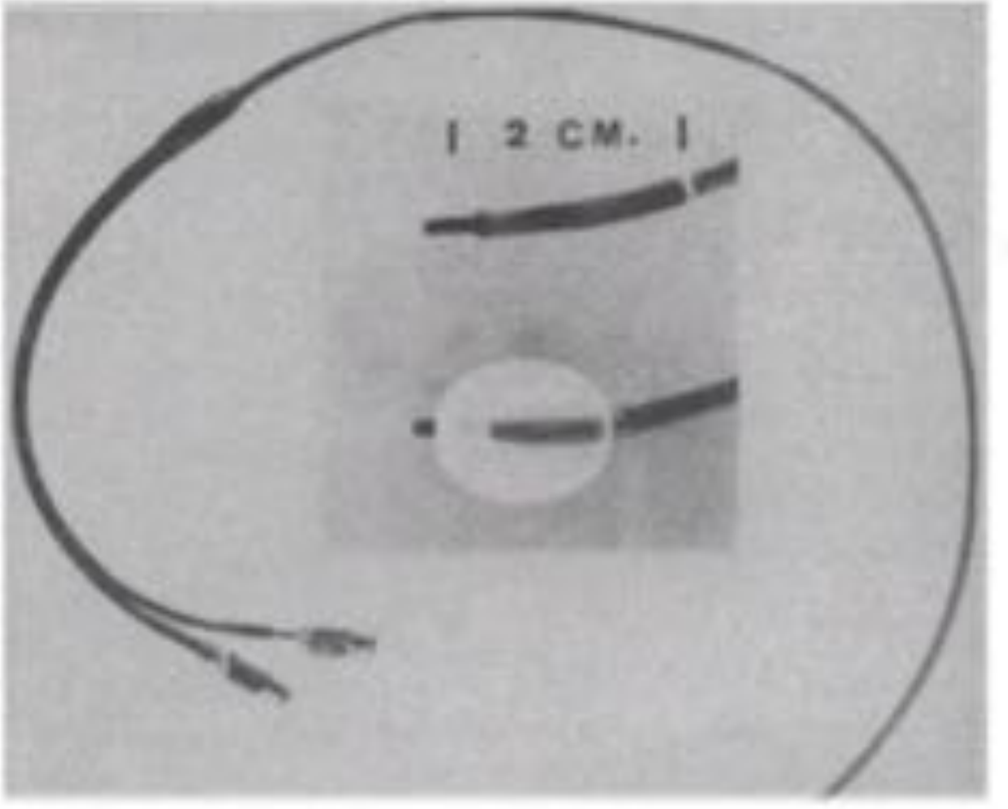
1950

Creation of an Atrial Septal Defect Without Thoracotomy

A Palliative Approach to Complete Transposition of the Great Arteries

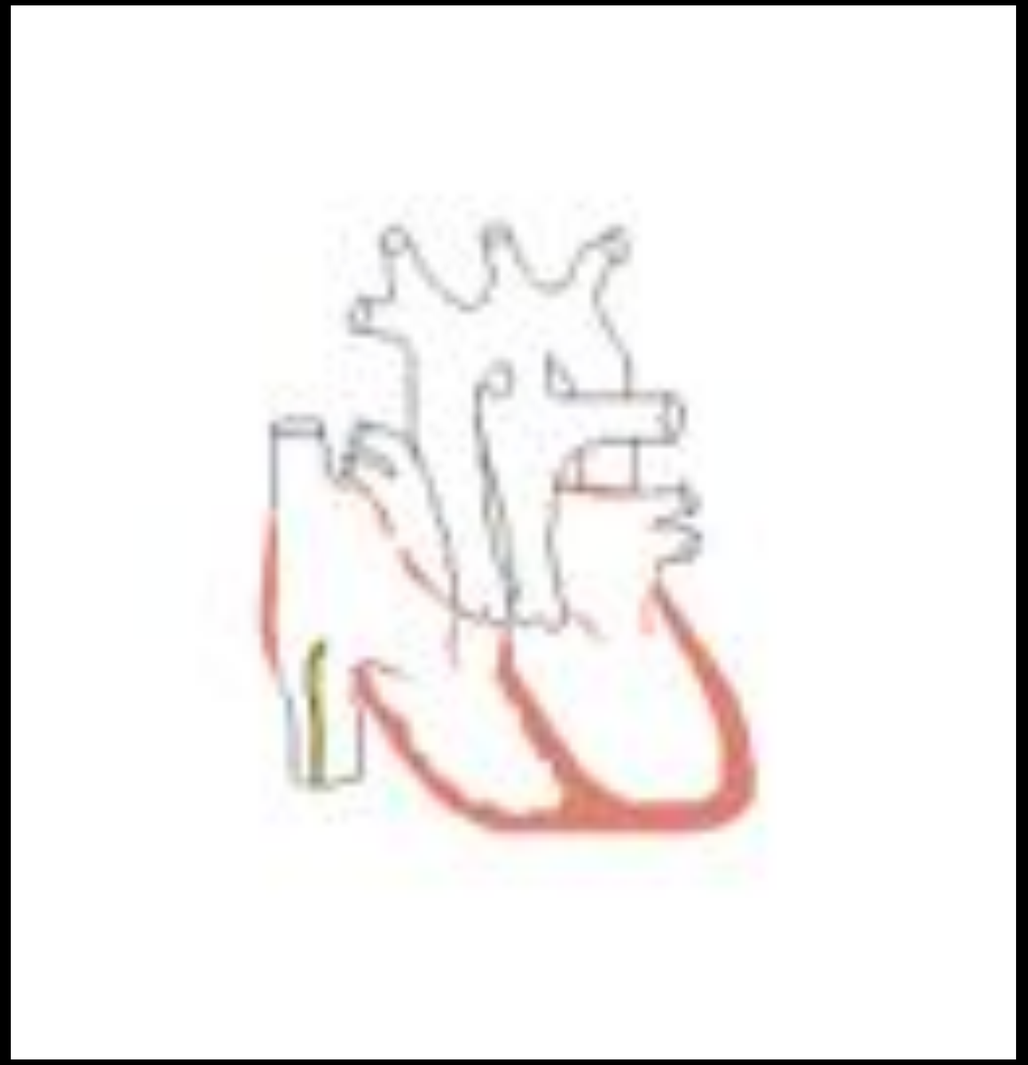
William J. Rashkind, MD, and William W. Miller, MD

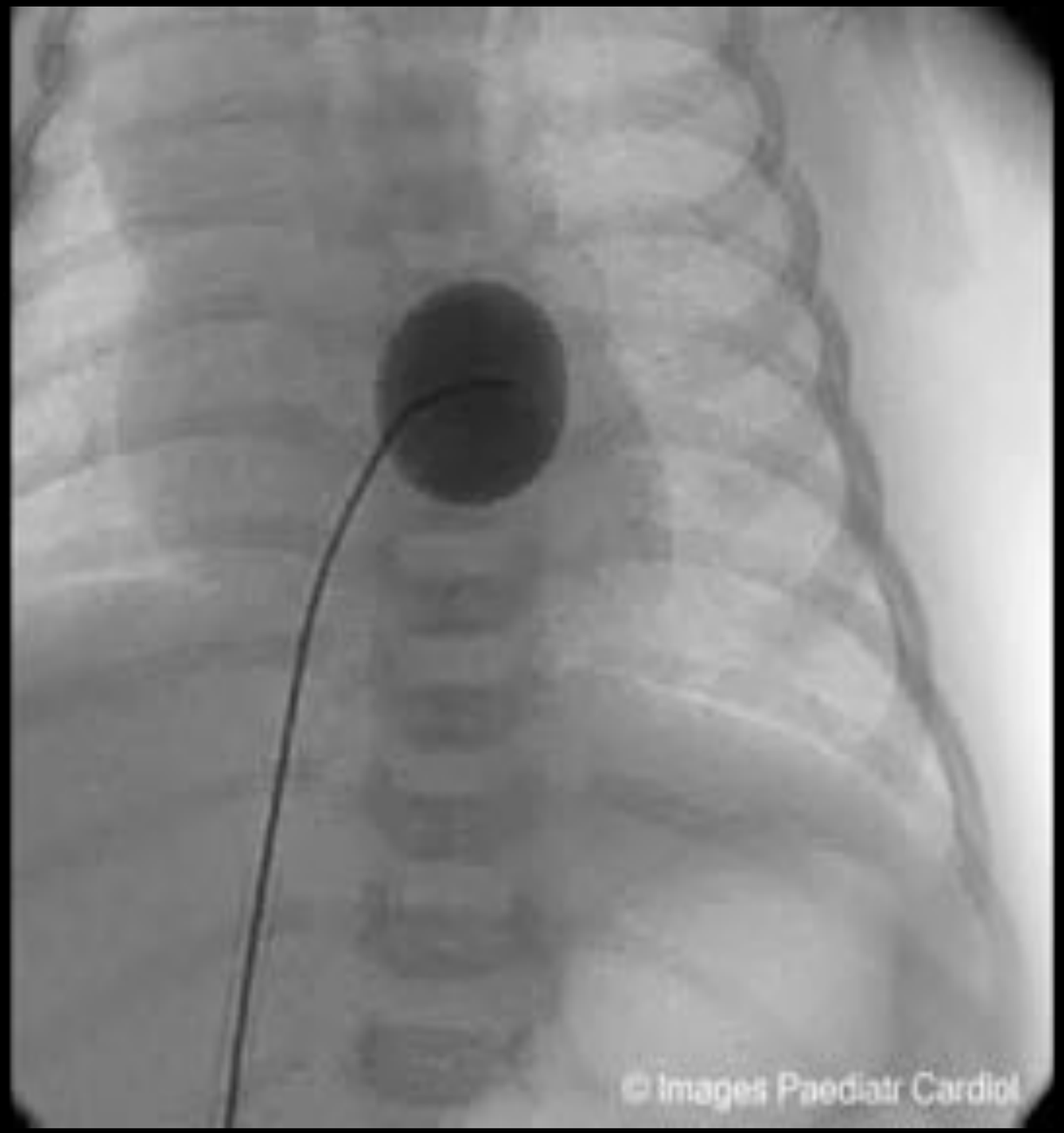
Transposition of the great vessels (TGV) occurs in approximately 20% of children who die with congenital heart disease.[1] With rare exceptions, patients with this lesion die in the first 6 months of life (50% within the first month). Approximately 40% of patients with TGV have an otherwise normal heart. 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



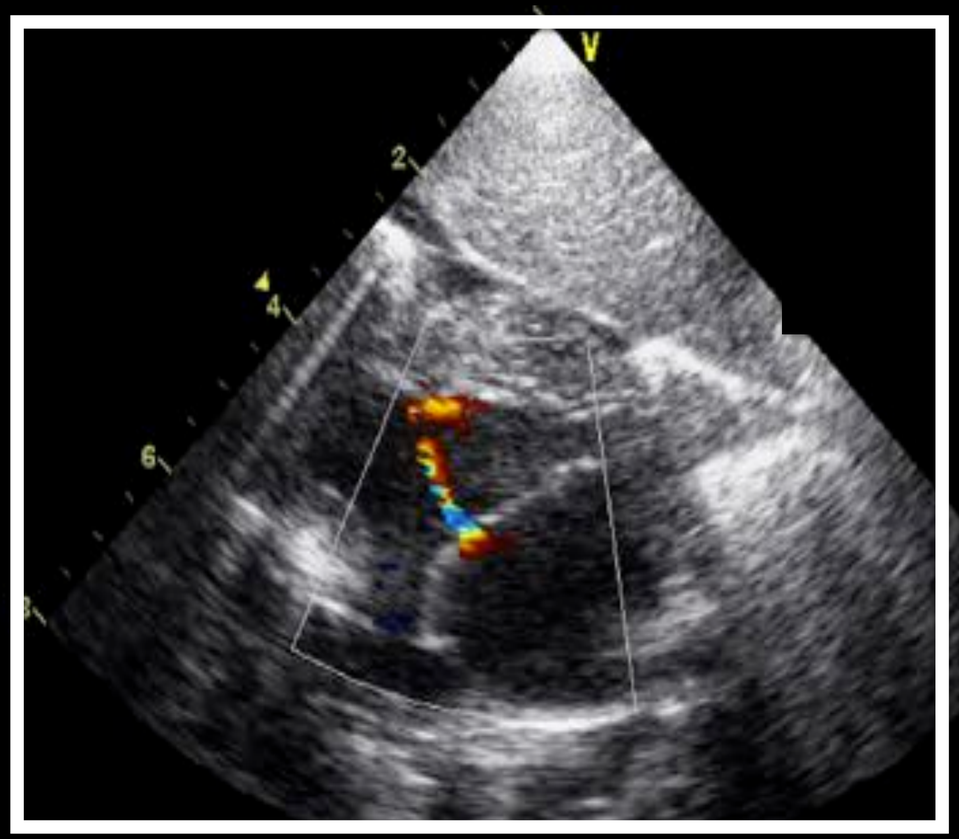
Rashkind WJ and Miller WW. JAMA 1966

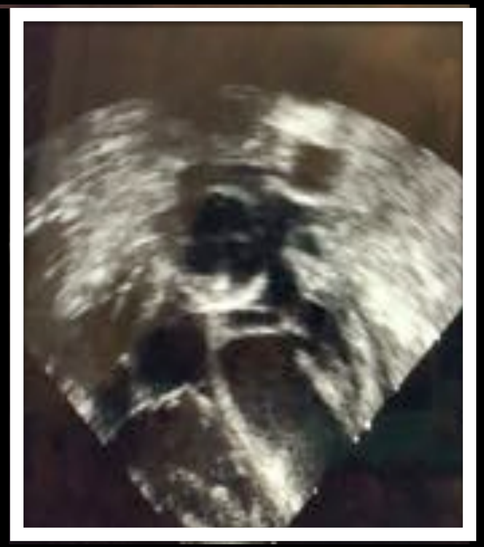
William Rashkind

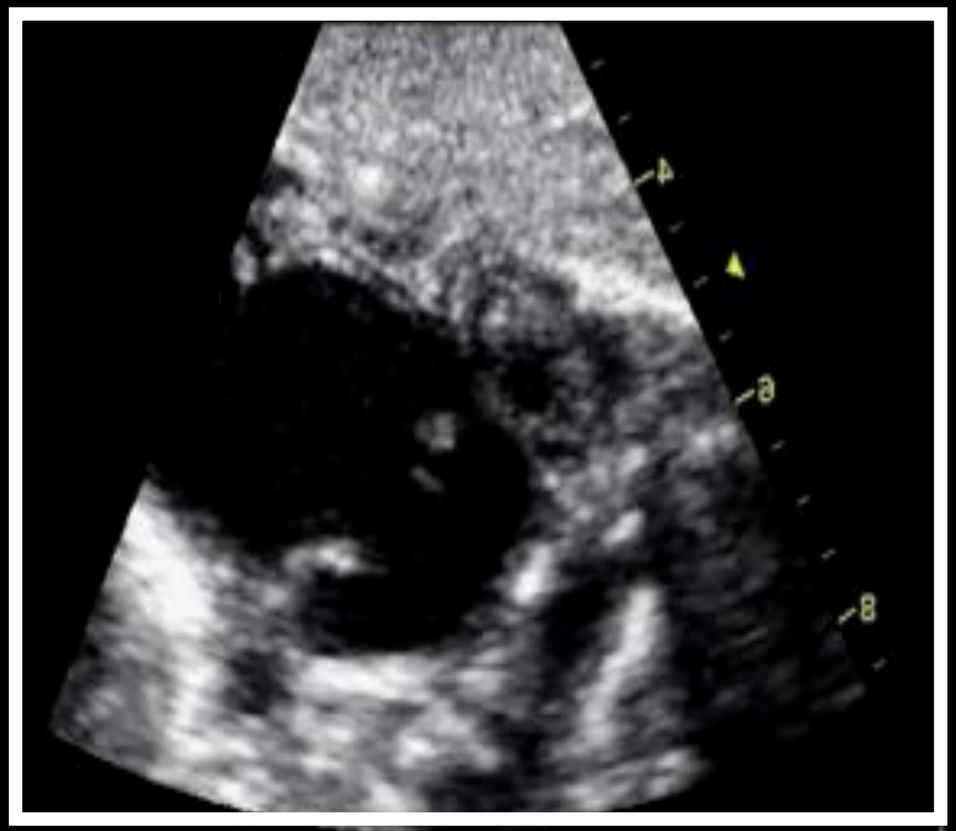



© Images Paediatr Cardiol

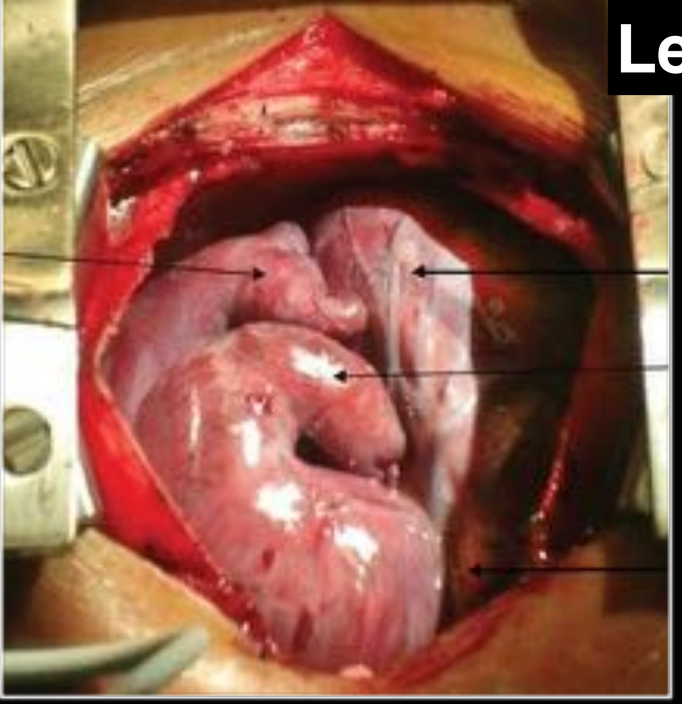
TGA Rashkind







Left juxtaposition of atrial appendages



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.

Task Force 3: Training Guidelines for Pediatric Cardiac Catheterization and Interventional Cardiology

Endorsed by the Society for Cardiovascular Angiography and Interventions

Robert H. Beekman, III, MD, FACC, FAAP, Chair;
William E. Hellenbrand, MD, FACC; Thomas R. Lloyd, MD, FACC;
James E. Lock, MD, FACC, FAAP; Charles E. Mullins, MD, FACC, FAHA,
FAAP; Jonathan J. Rome, MD, FACC; and David F. Teitel, MD

Pediatrics 2005;116;1574-1575;

TABLE 1. Recommended Body of Knowledge Covered During Core Training

Indications for and risks of cardiac catheterization and angiography

Indications for and risks of therapeutic catheter procedures

Interpretation of pressure waveforms

Interpretation of O2 saturation data

Fick principle and shunt calculations

Vascular resistance calculations

Cardiac angiography: basic techniques/angles/interpretation

Radiation safety

TABLE 2. Core Training—Recommended Minimum Case

Total cardiac catheterizations	100
Interventional procedures Type of intervention	20
Balloon septostomy	5

TABLE 3. Advanced Training—Recommended Minimum Case Numbers

Total cardiac catheterizations	200
Interventional procedures	100
Type of intervention	
Balloon septostomy	5
Transseptal puncture	10
Pulmonary valve dilation	10
Aortic valve dilation	10
Pulmonary artery dilation	10
Pulmonary artery stent	10
Coarctation dilation	10
Coarctation stent	5
Collateral occlusion	10
Ductus arteriosus occlusion	10
Atrial septal defect occlusion	10

Incidence de la manoeuvre de Rashkind

Environ 780.000 naissances en France

Incidence de la TGV = 0.15 pour mille naissances vivantes

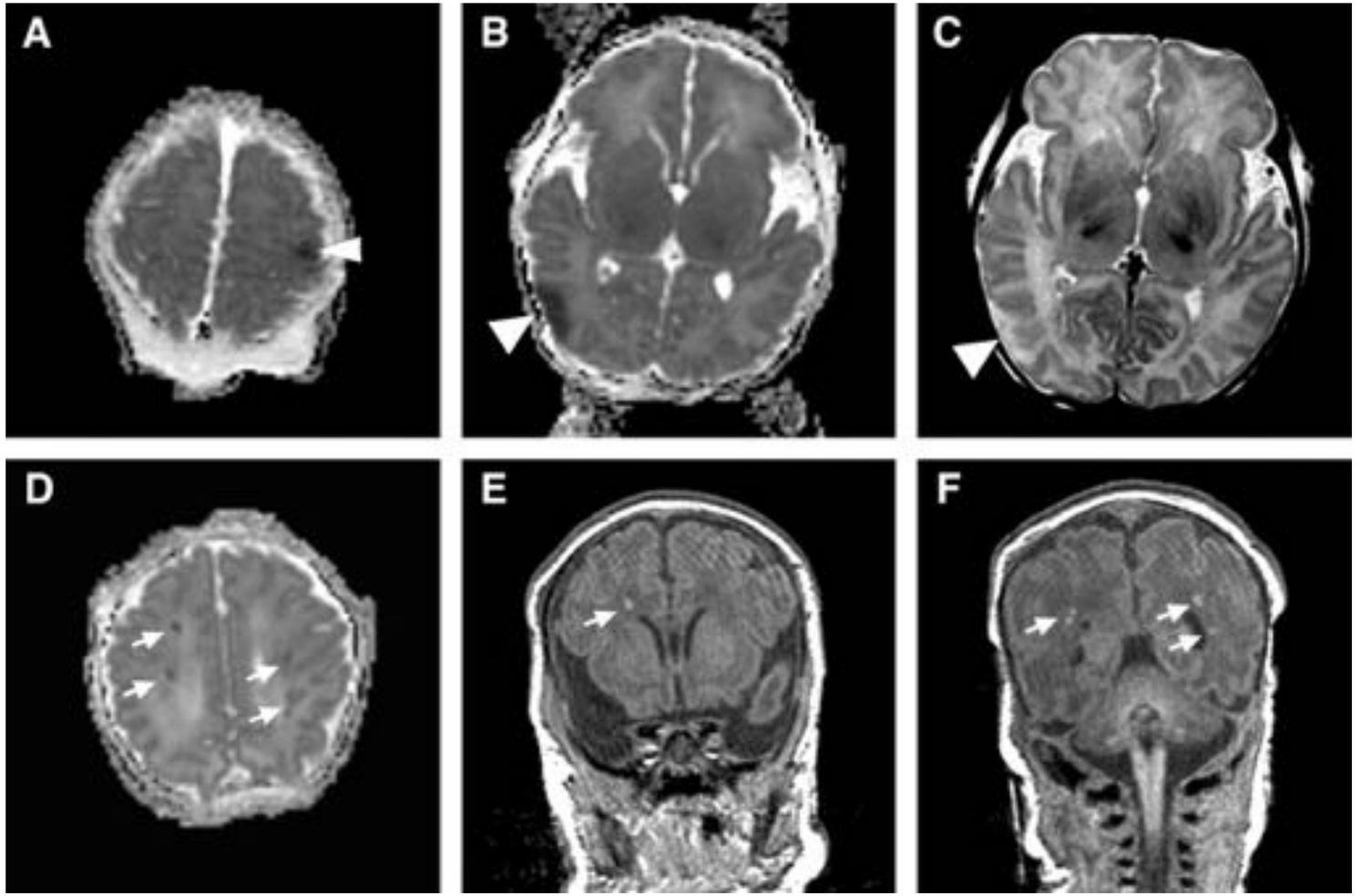
Soit environ 120 transpositions des gros vaisseaux / an

Indication de Rashkind dans 50% des cas environ = 60 Rashkind/an

24 centres de Cardiologie Congénitale identifiés (déséquilibrés en recrutement de TGV du fait du transfert *in utero*)

- -pour de nombreux centres moins de 1 Rashkind /an
- -pour de nombreux cardiopédiatres moins de 1 Rashkind /3 ans

Balloon atrial septostomy is associated with preoperative stroke in neonates with transposition of the great arteries.



A–C, Example of multiple focal strokes.

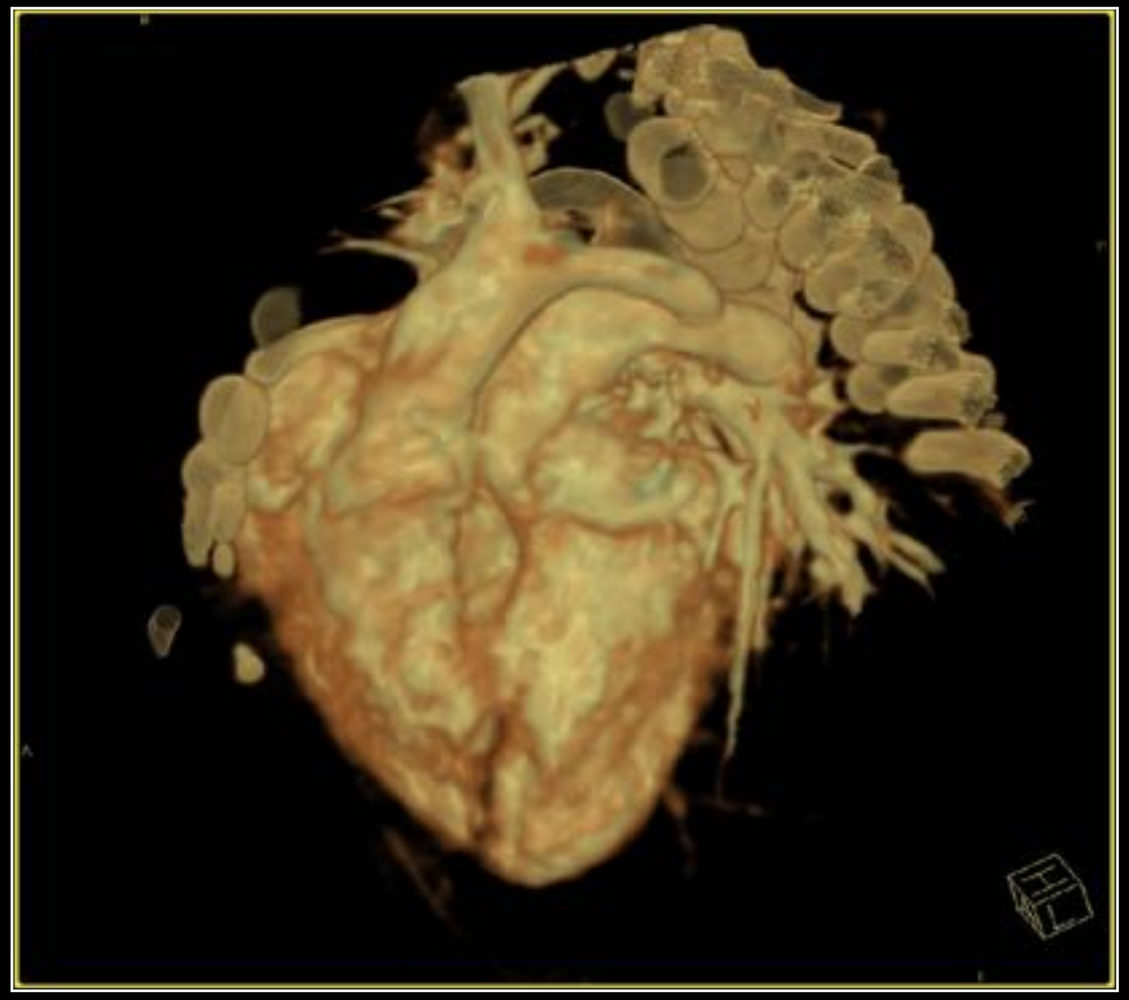
McQuillen P S et al. Circulation 2006;113:280-285

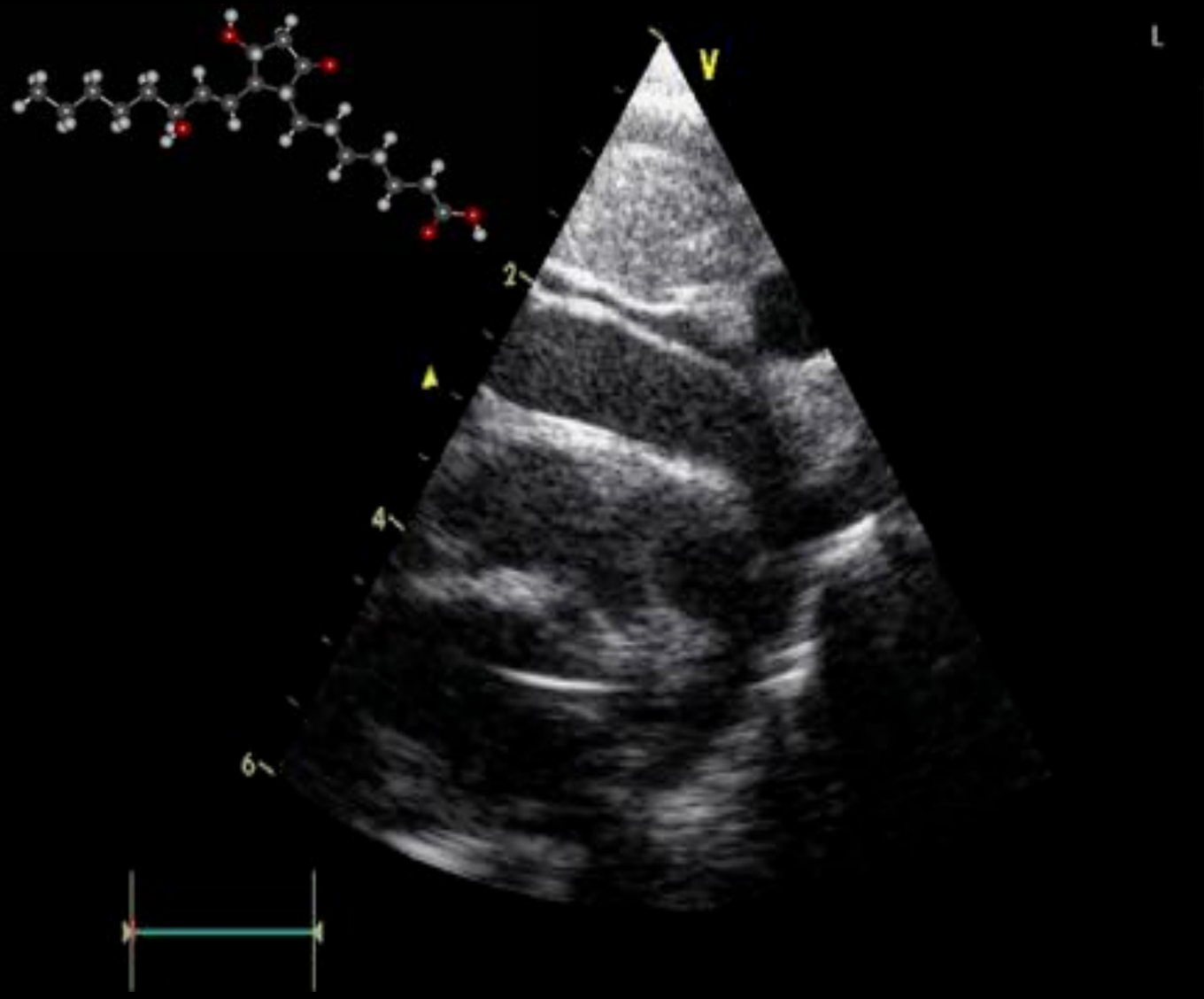
Preoperative brain injury in transposition of the great arteries is associated with oxygenation and time to surgery, not balloon atrial septostomy.

Circulation. 2009;119:709-16.

	PVL (n=10)	No PVL (n=16)	P
BAS, n (%)	6 (60)	8 (50)	NS
Gestational age, wk	39.0±1.2	38.9±1.1	NS
Birth weight, kg	3.41±0.59	3.50±0.64	NS
Head circumference, cm	33.1±1.6	33.8±1.6	NS
TGA with VSD, n (%)	3 (30)	3 (19)	NS
Female, n (%)	5 (50)	6 (37)	NS
Preoperative measures			
pH	7.44±0.06	7.43±0.06	NS
Pco_2, mm Hg	38.8±2.9	39.6±3.8	NS
Po_2, mm Hg	36.9±1.5	41.9±5.0	0.026
Hemoglobin, g/dL	13.8±0.9	14.8±2.1	NS
Base excess, mEq/L	2.43±3.5	1.63±2.9	NS
Lactate	2.9±0.8	3.9±1.2	NS
ABGs per day, n	6.8±2.3	7.1±2.1	NS
Lowest O_2 saturation, %	76.1±9.0	75.3±16.4	NS
Time to surgery, d	5.6±2.9	3.9±2.2	0.028

PGE1 and arterial duct





To Intubate or Not to Intubate? Transporting Infants on Prostaglandin E 1

Garth D. Meckler and Calvin Lowe

Pediatrics 2009;123;e25-e30; originally published online Dec 8, 2008;

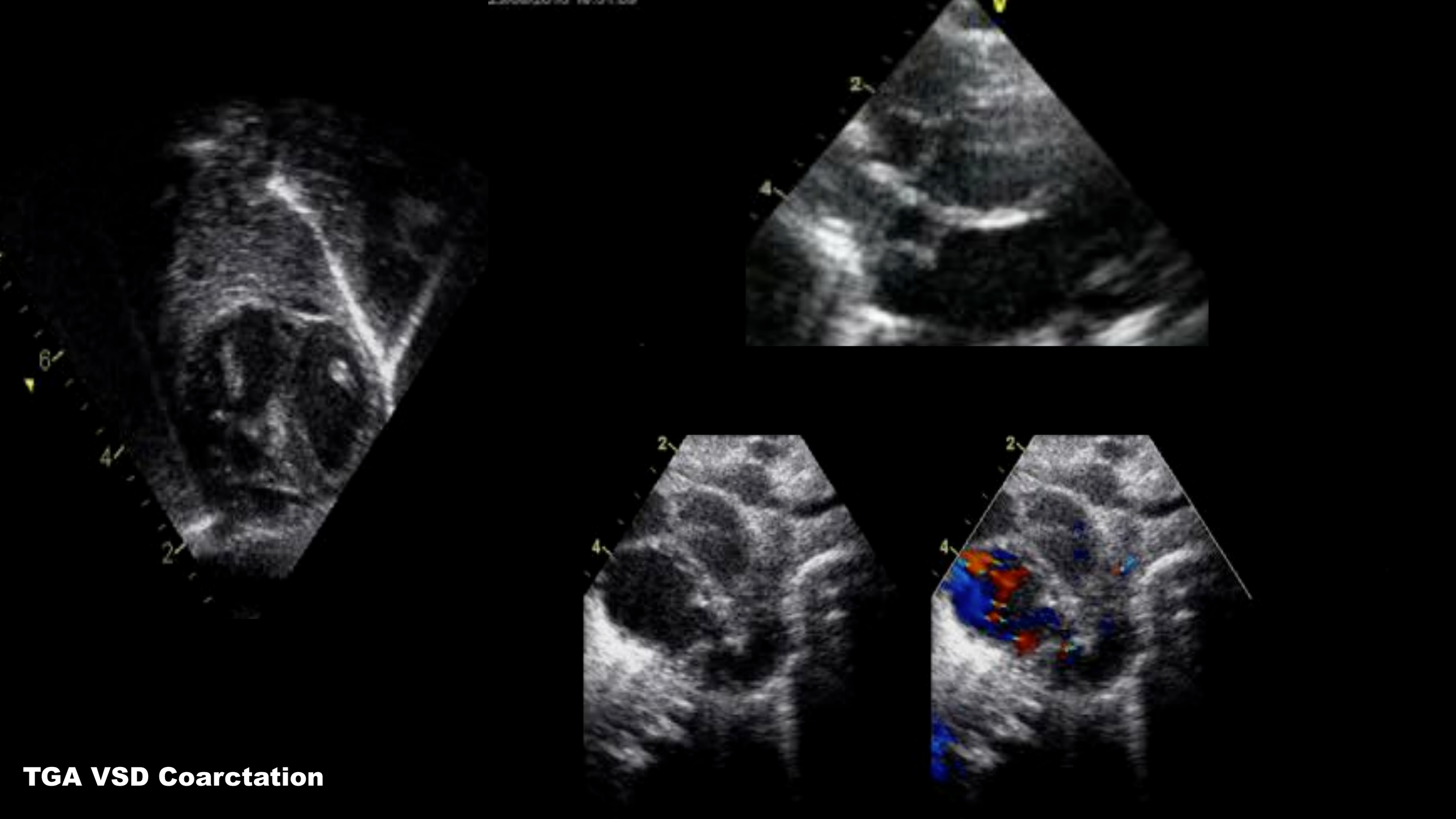
TABLE 5 Multivariate Analysis of Major Transport Complications

Variable	OR	95% CI
Medical comorbidity	2.22	1.02–4.08
PGE, dose		
<0.05 μg/kg per min	(1)	
0.05 μg/kg per min	4.80	1.60–14.40
>0.05 μg/kg per min	3.72	1.10–12.63
Intubation type		
Unintubated	(1)	
Emergent	15.68	3.85–63.83
Elective	7.44	2.82–19.68
CHD physiology		
Single ventricle	1.42	0.66–3.07
Transport mode		
Ground	(1)	
Helicopter	1.17	0.49–2.78
Fixed wing	0.20	0.02–2.59
Transport time		
<30 min	(1)	
30–60 min	0.89	0.36–2.19
60–90 min	0.58	0.18–1.89
>90 min	3.73	0.44–31.39
Gender, EGA	NS	

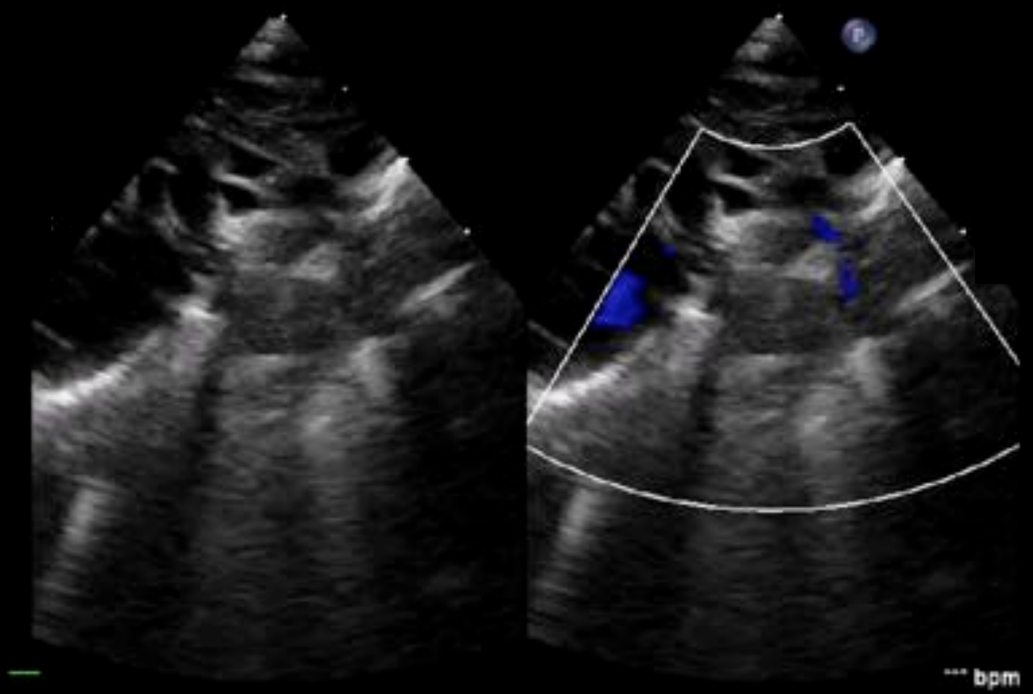
EGA indicates estimated gestational age; NS, not significant.

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

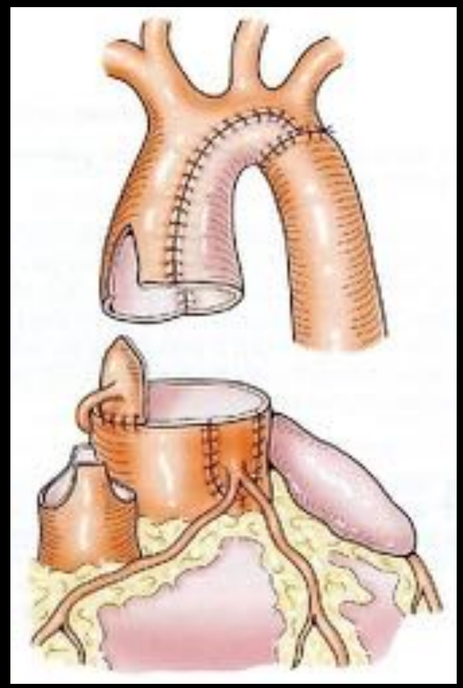

TGA VSD Coarctation

Aortic arch hypoplasia and coarctation

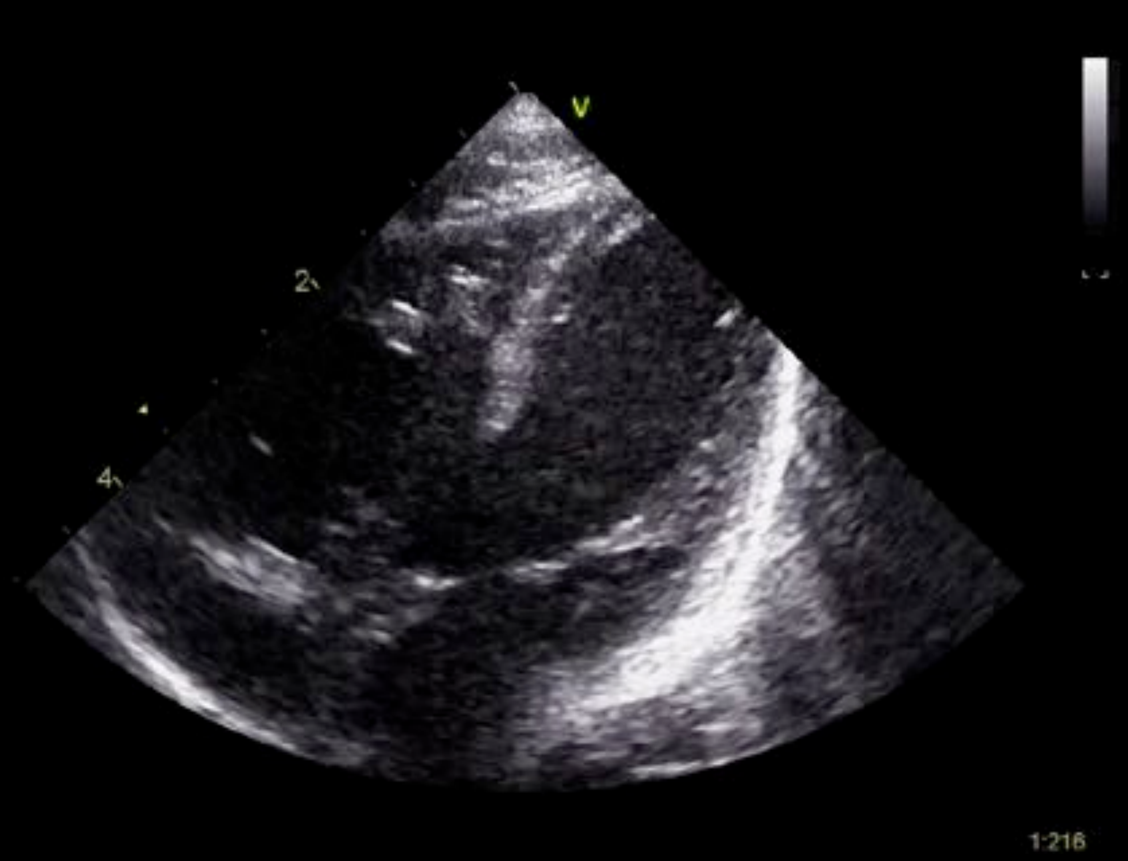


Localization and extension of the narrowed portion

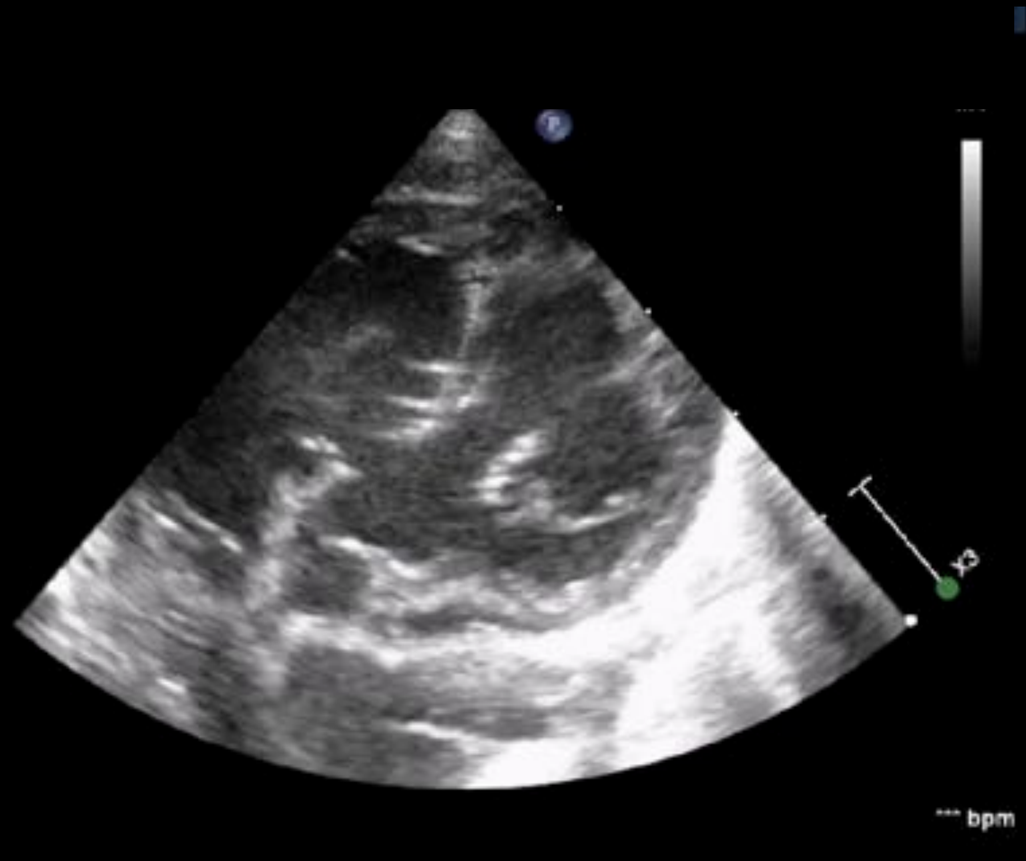
2 techniques: enlargement and extended end to end
=> Discrepancy between aortic and pulmonary roots



VSD: localization and size

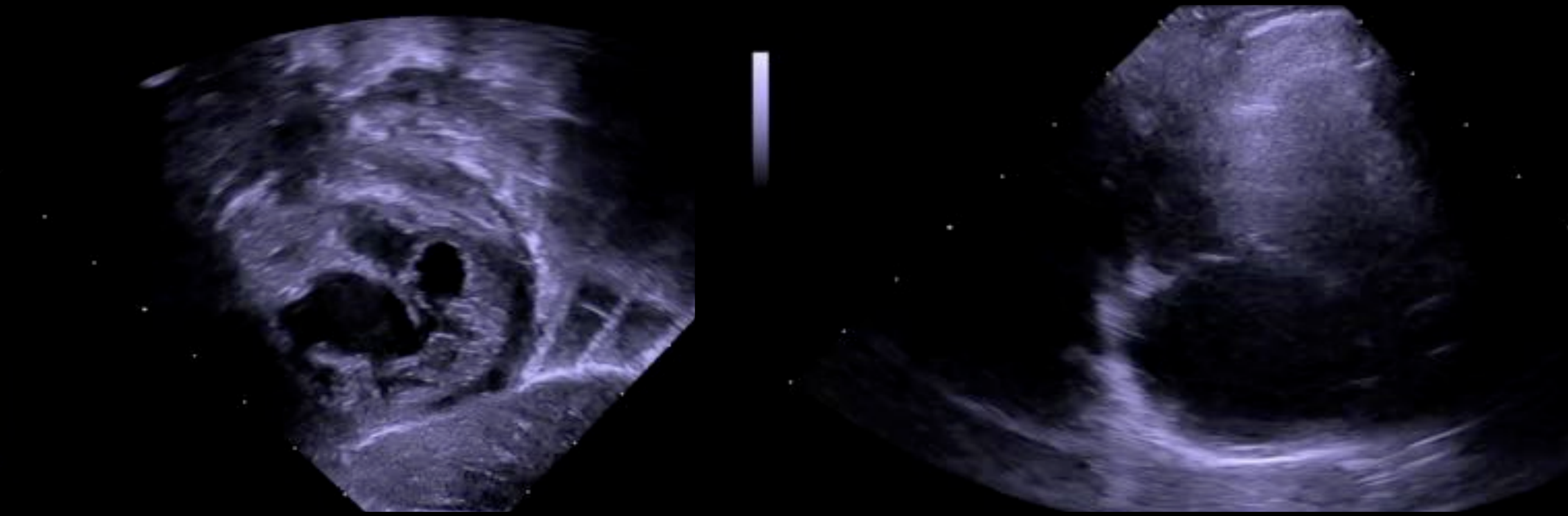


Inlet VSD



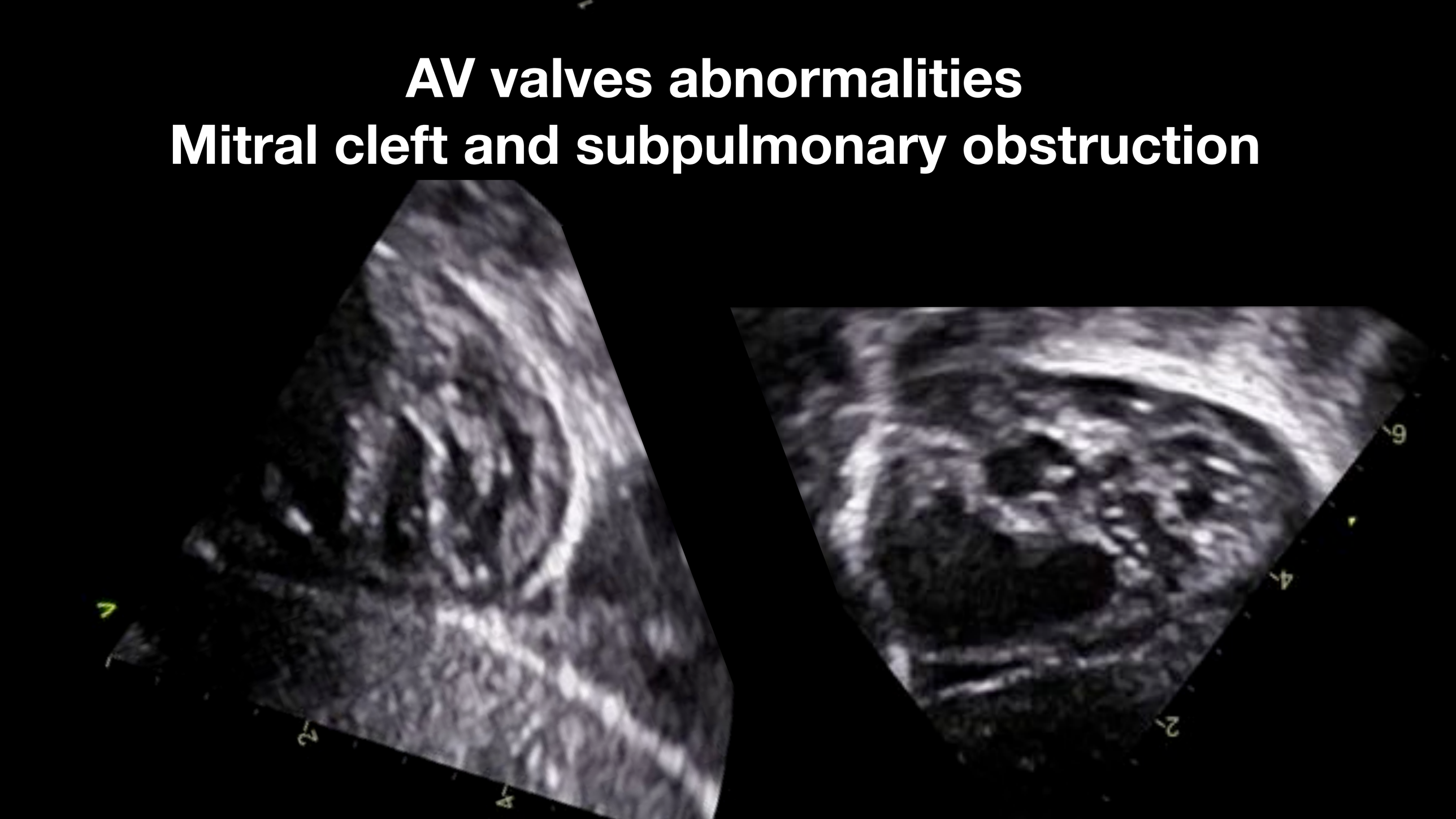
Outlet VSD

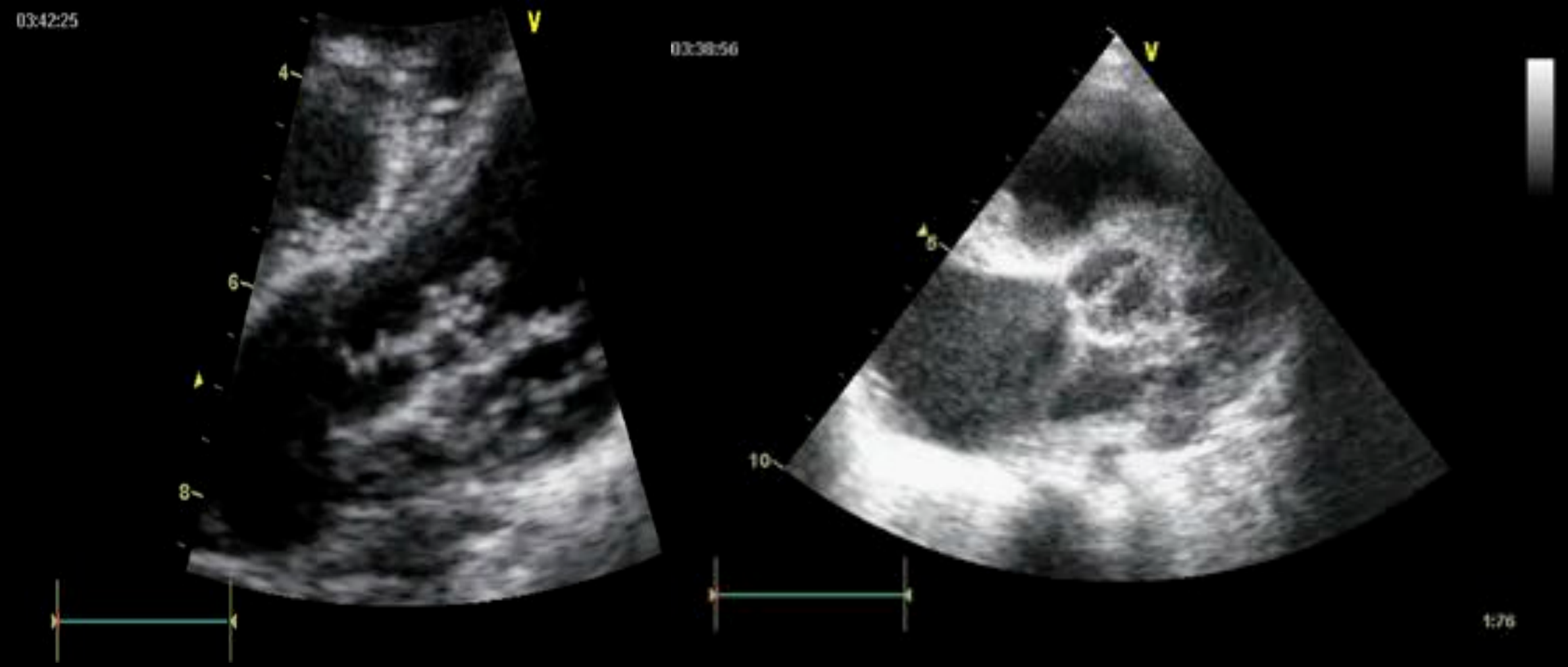
AV valves abnormalities
Straddling and over-riding



Straddling of tricuspid valve

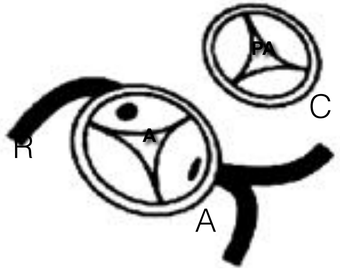
AV valves abnormalities
Mitral cleft and subpulmonary obstruction



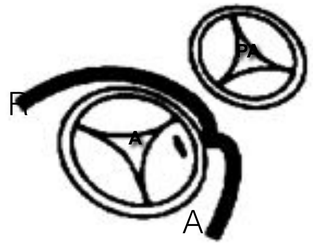


TGA left outflow tract obstruction

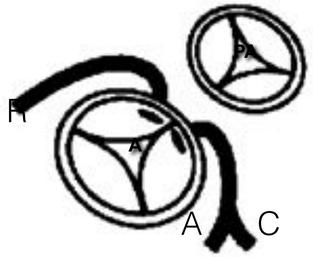
Type A



Type B

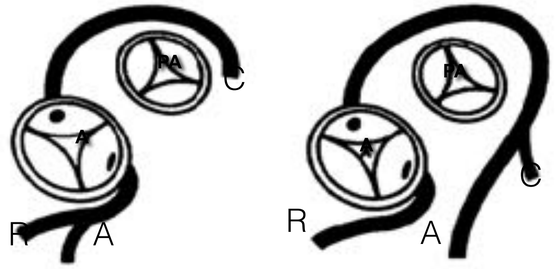


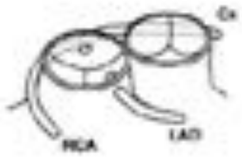
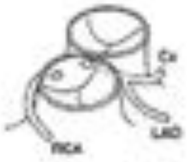
Type C

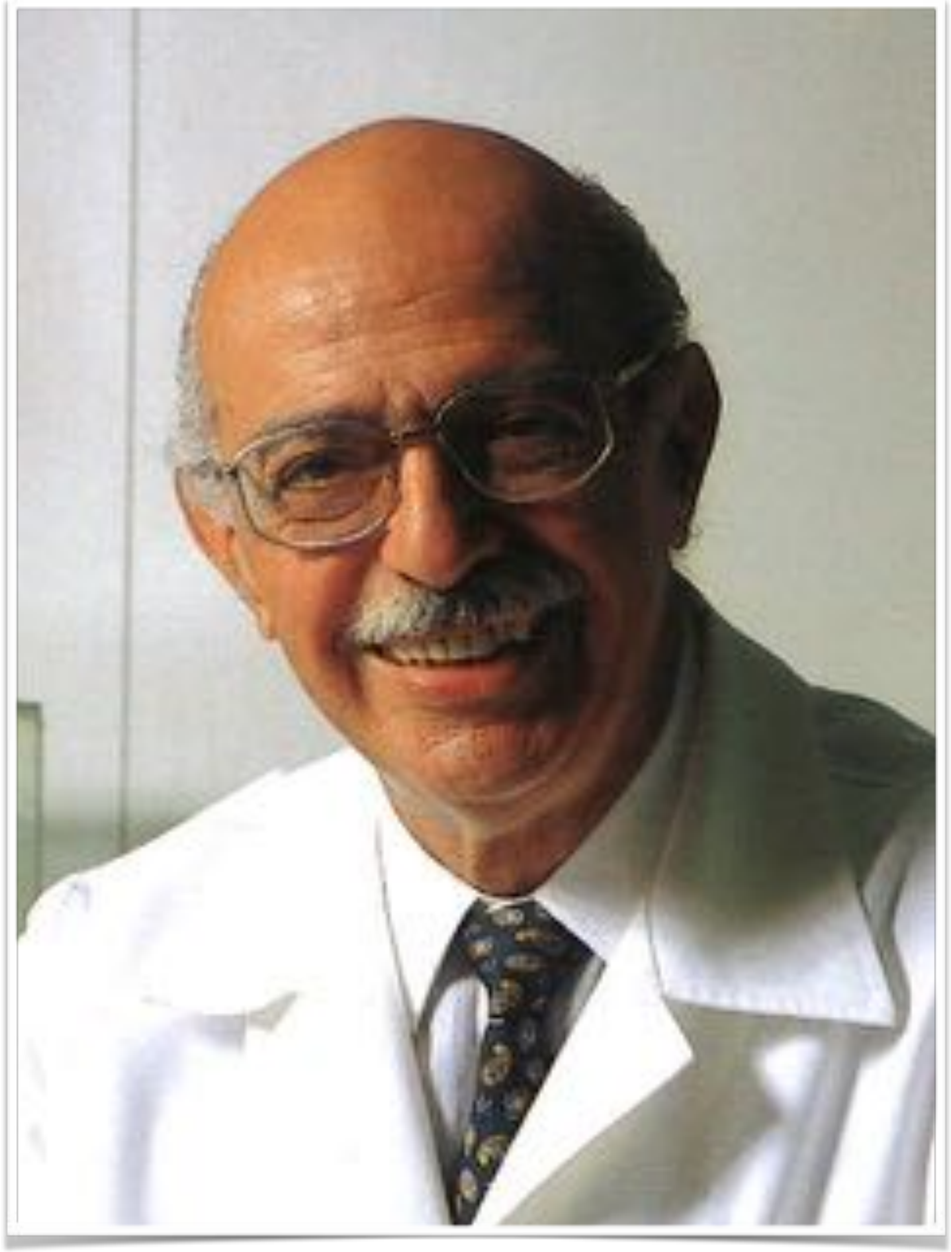


Type D

Type E

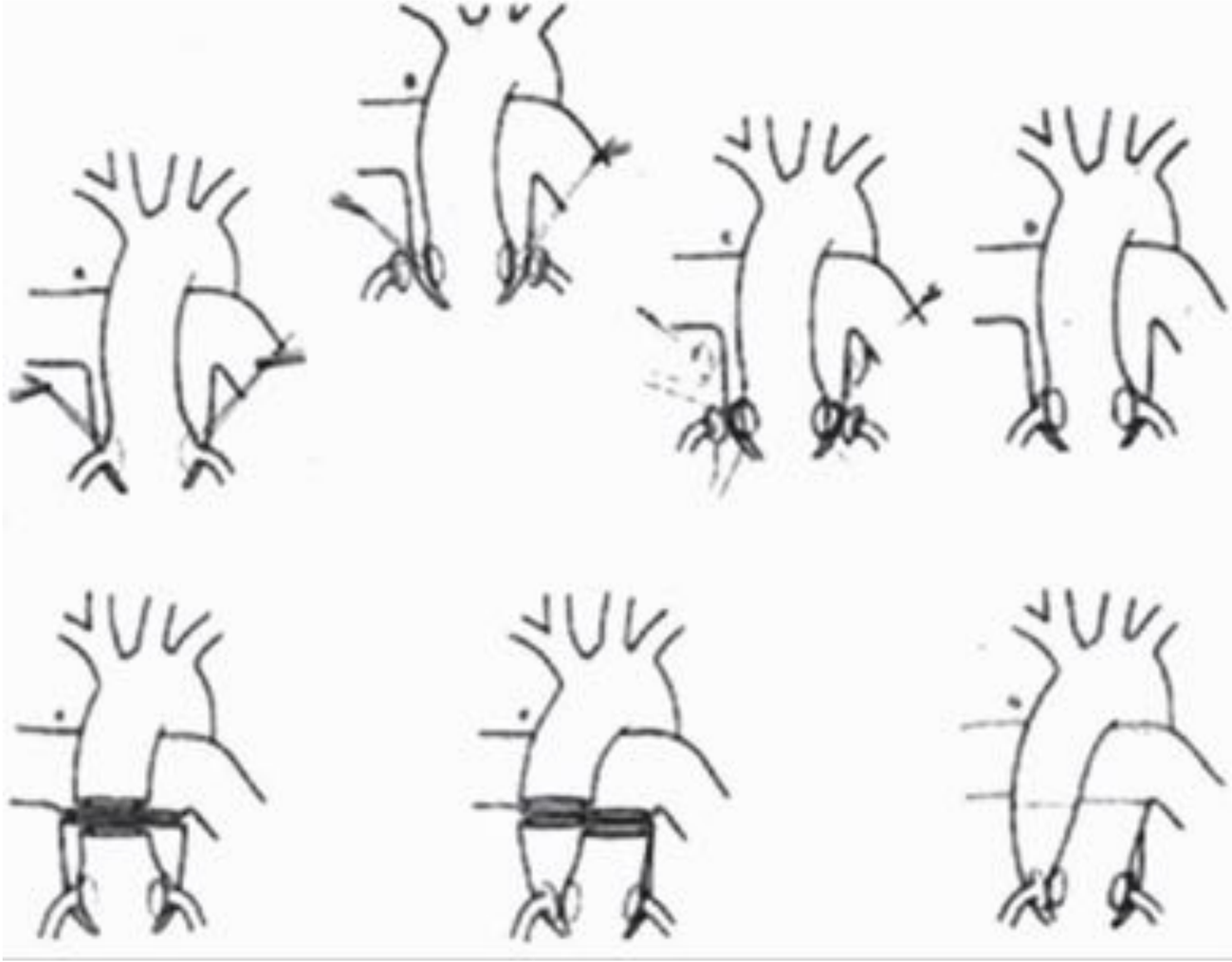


Habituel	Circonflexe de l'ACD	ACG unique	ACD unique	Inversée	ACD inversée et circonflexe	Coronaires Intramurales
						
66,9%	16,1%	1,7%	3,9%	2,4%	4,2%	3,2%

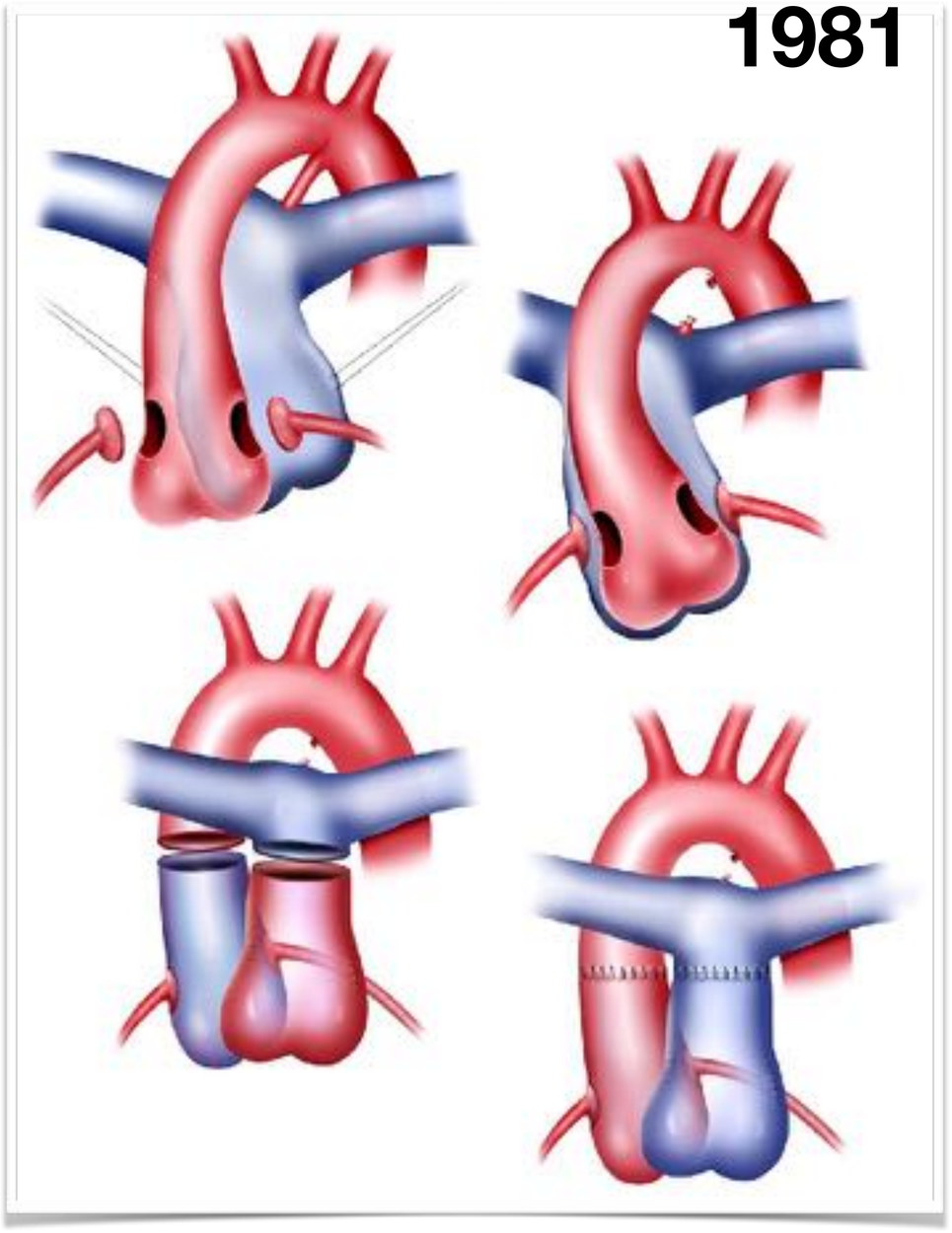


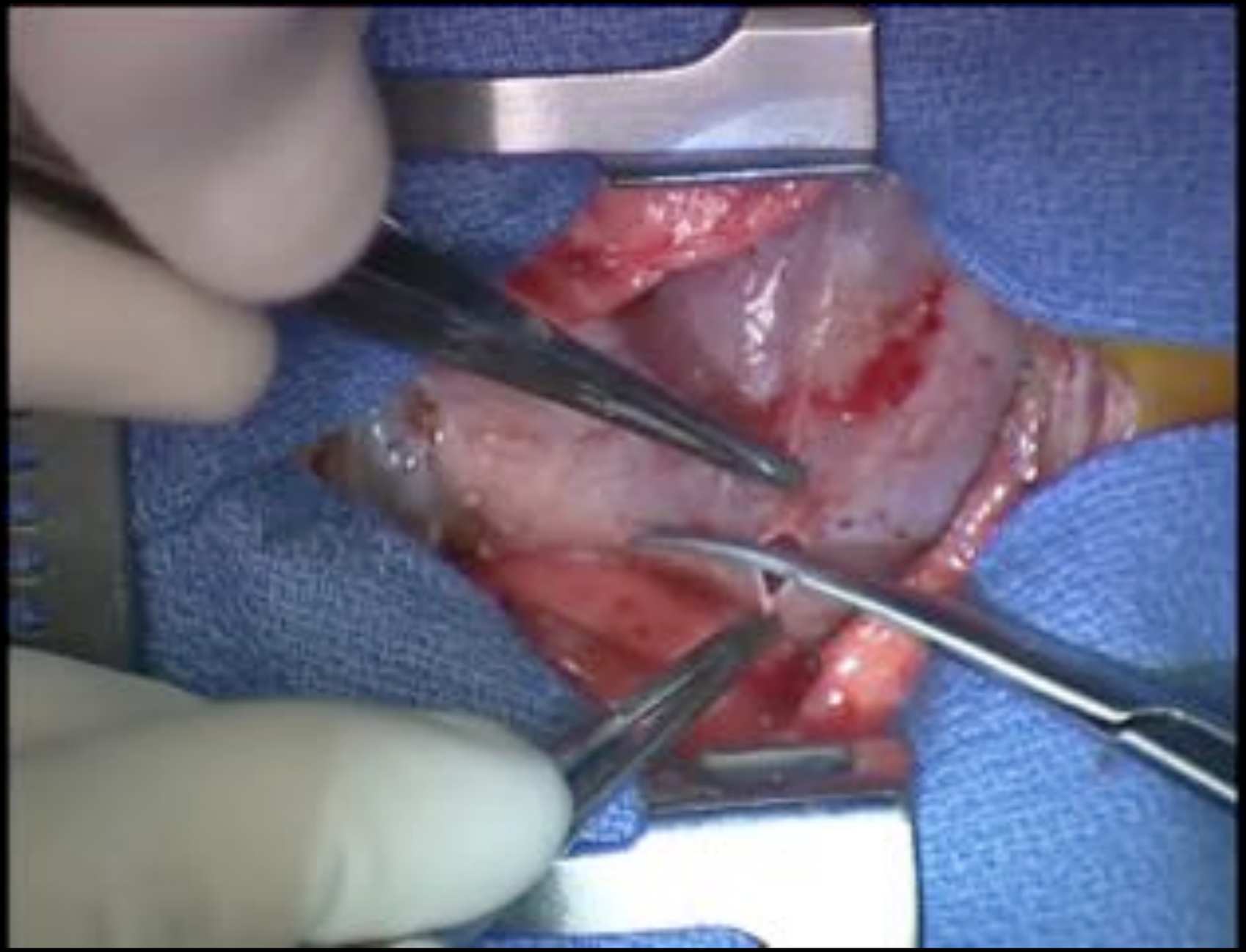
Adib Domingo Jatene

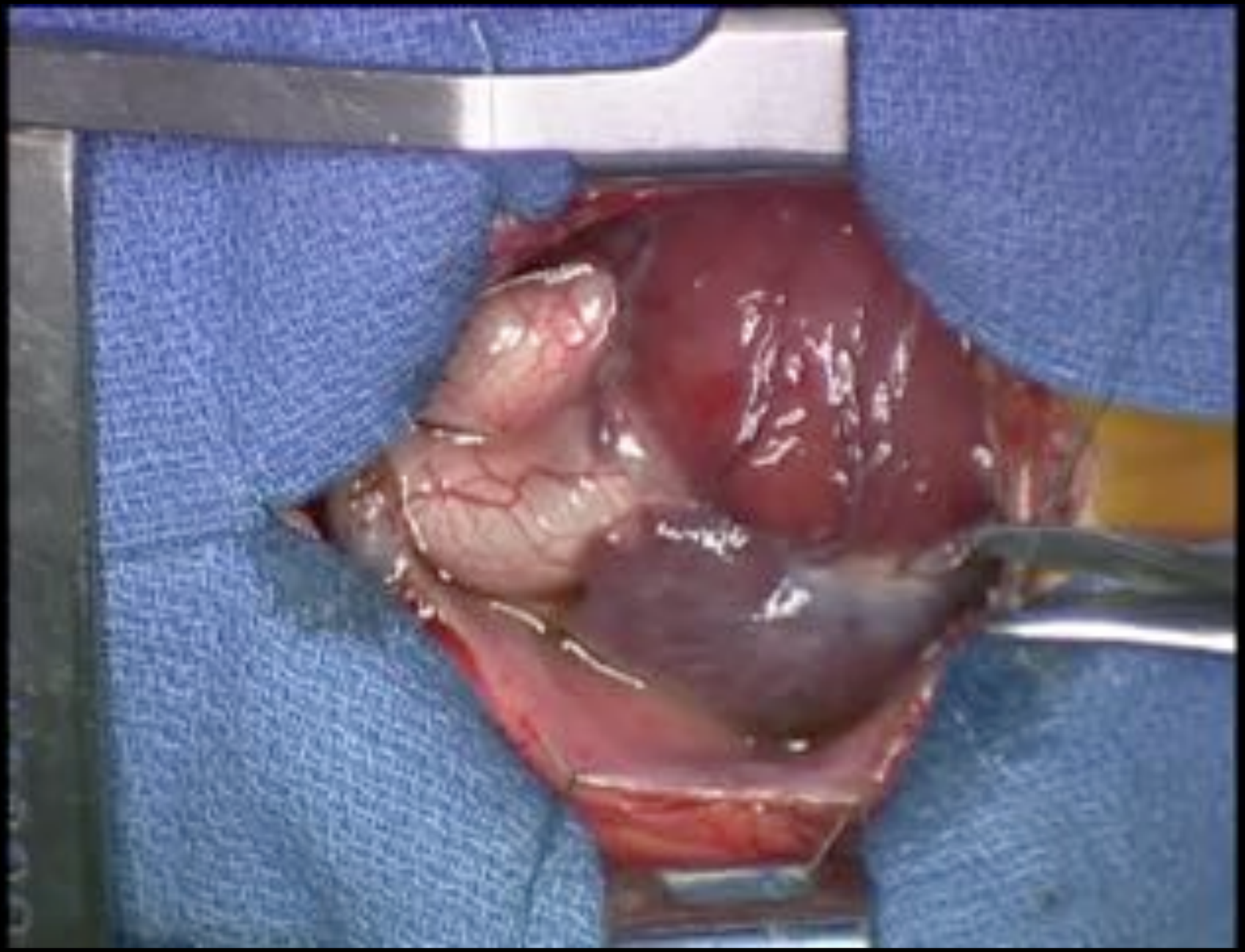
1975

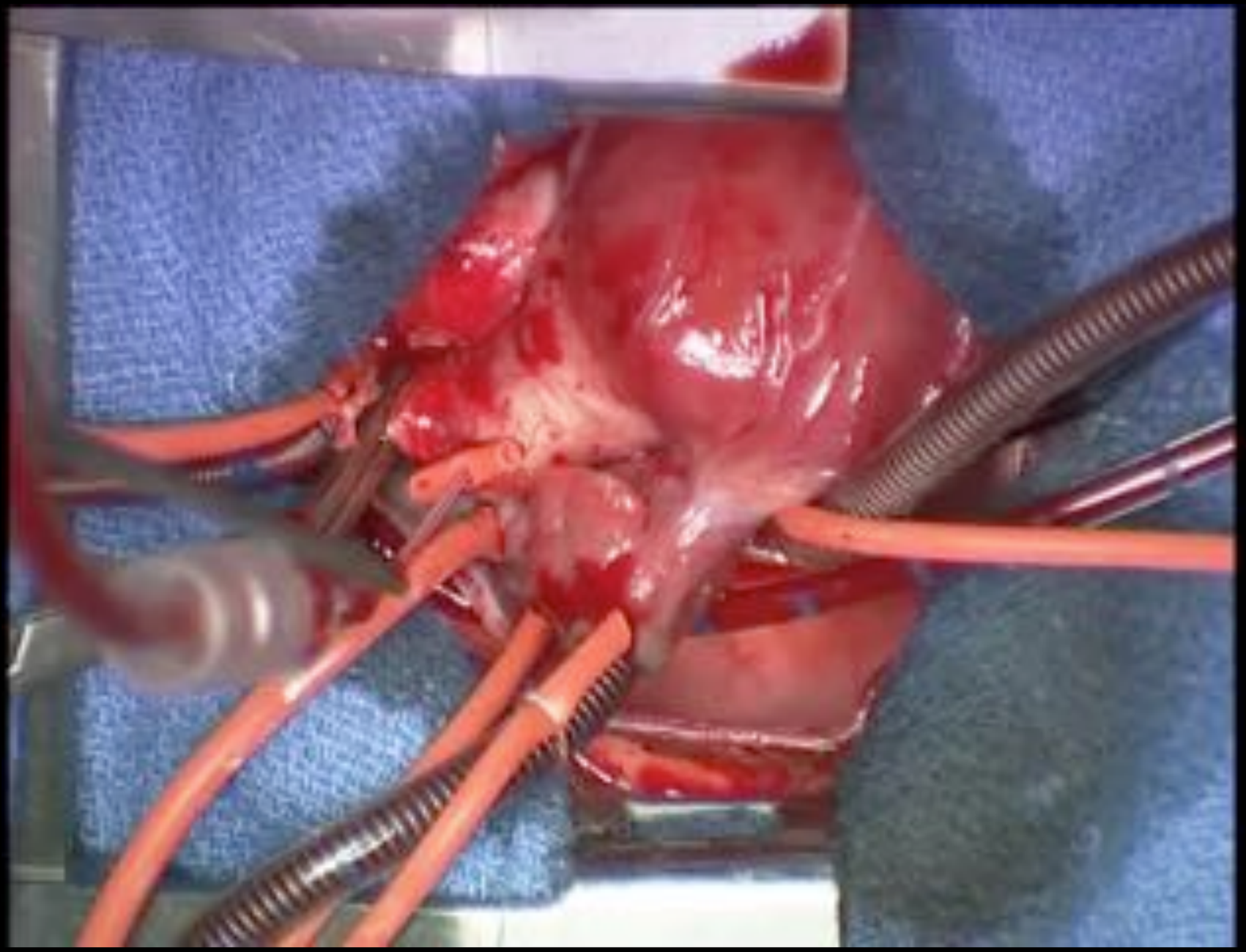


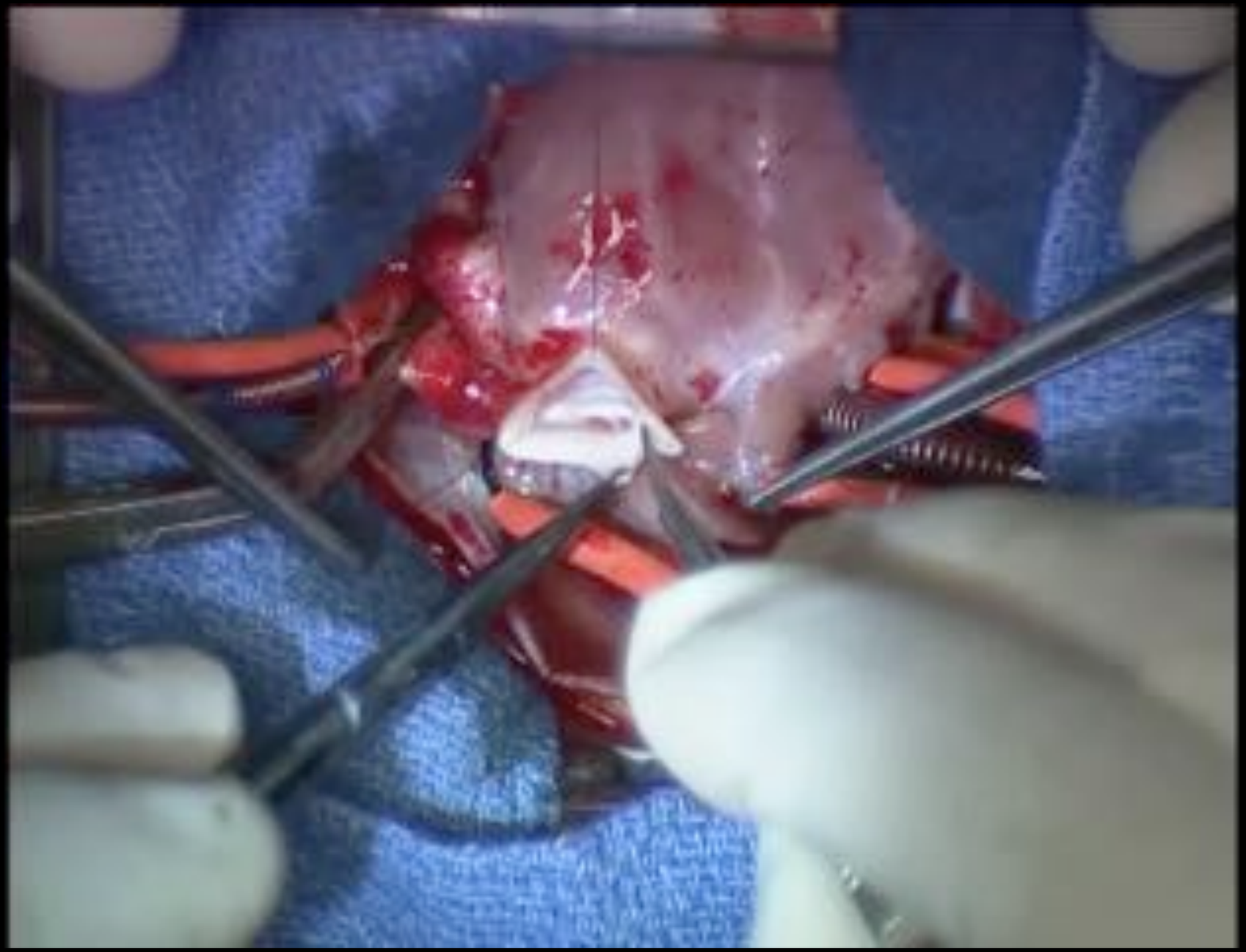
Yves Lecompte

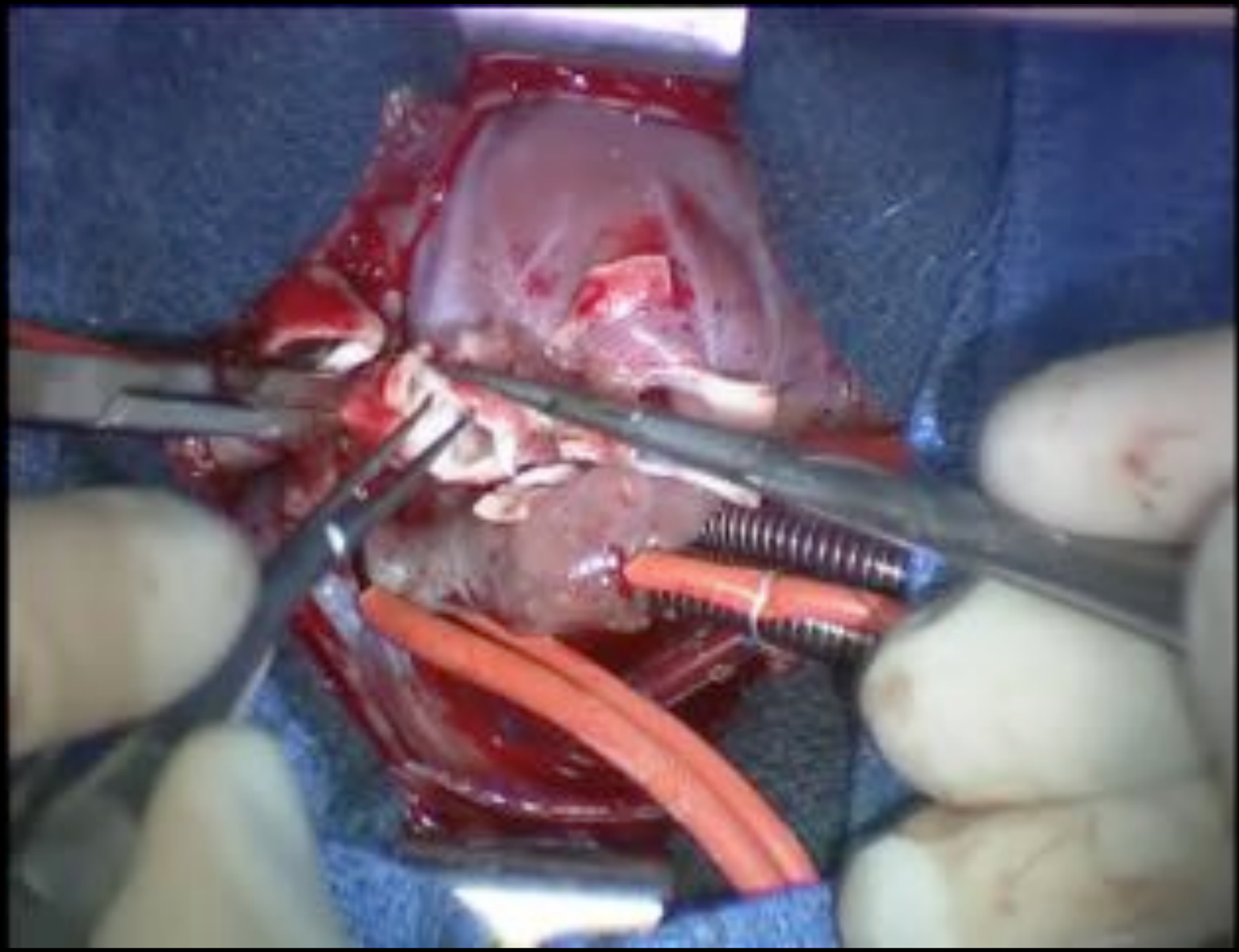


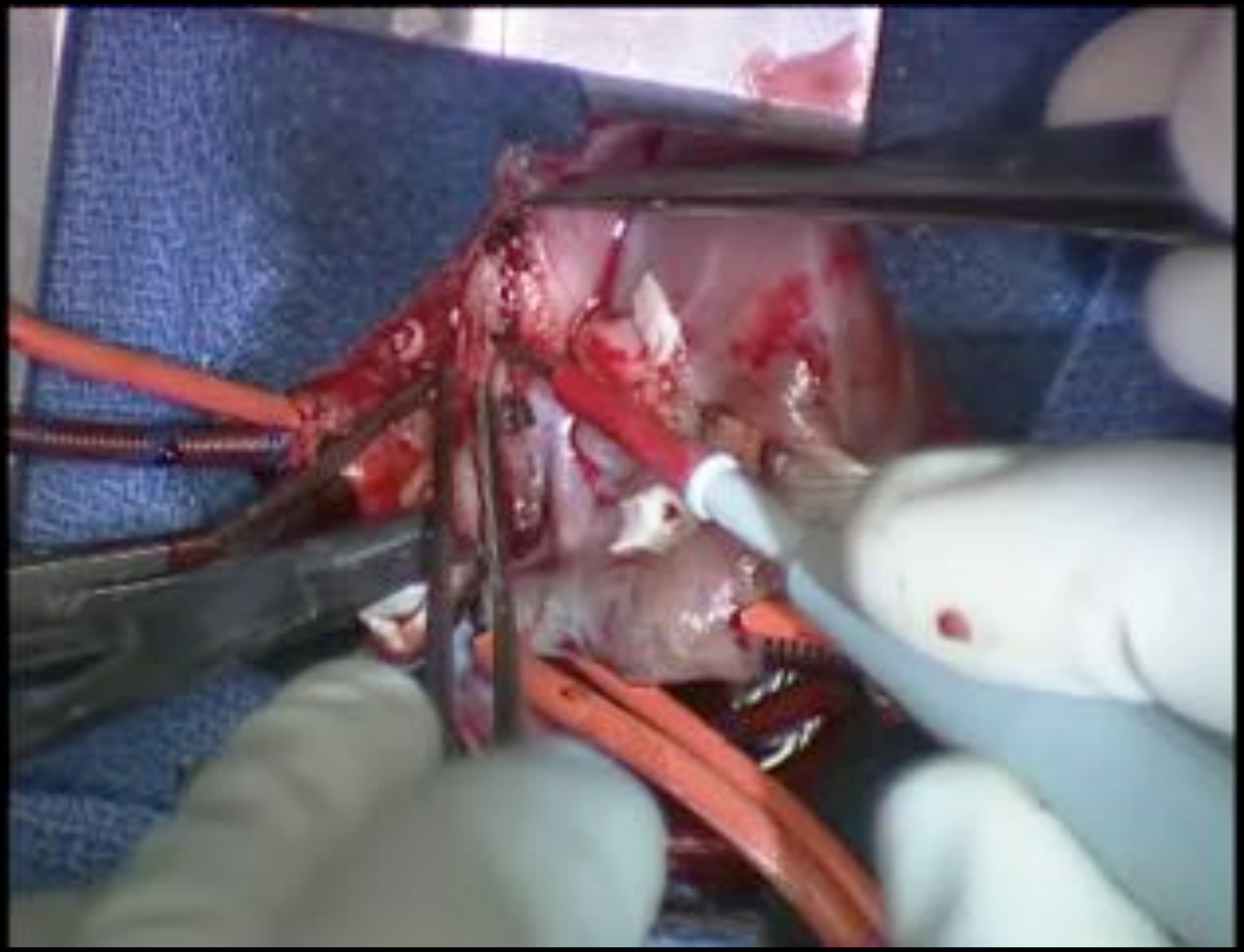


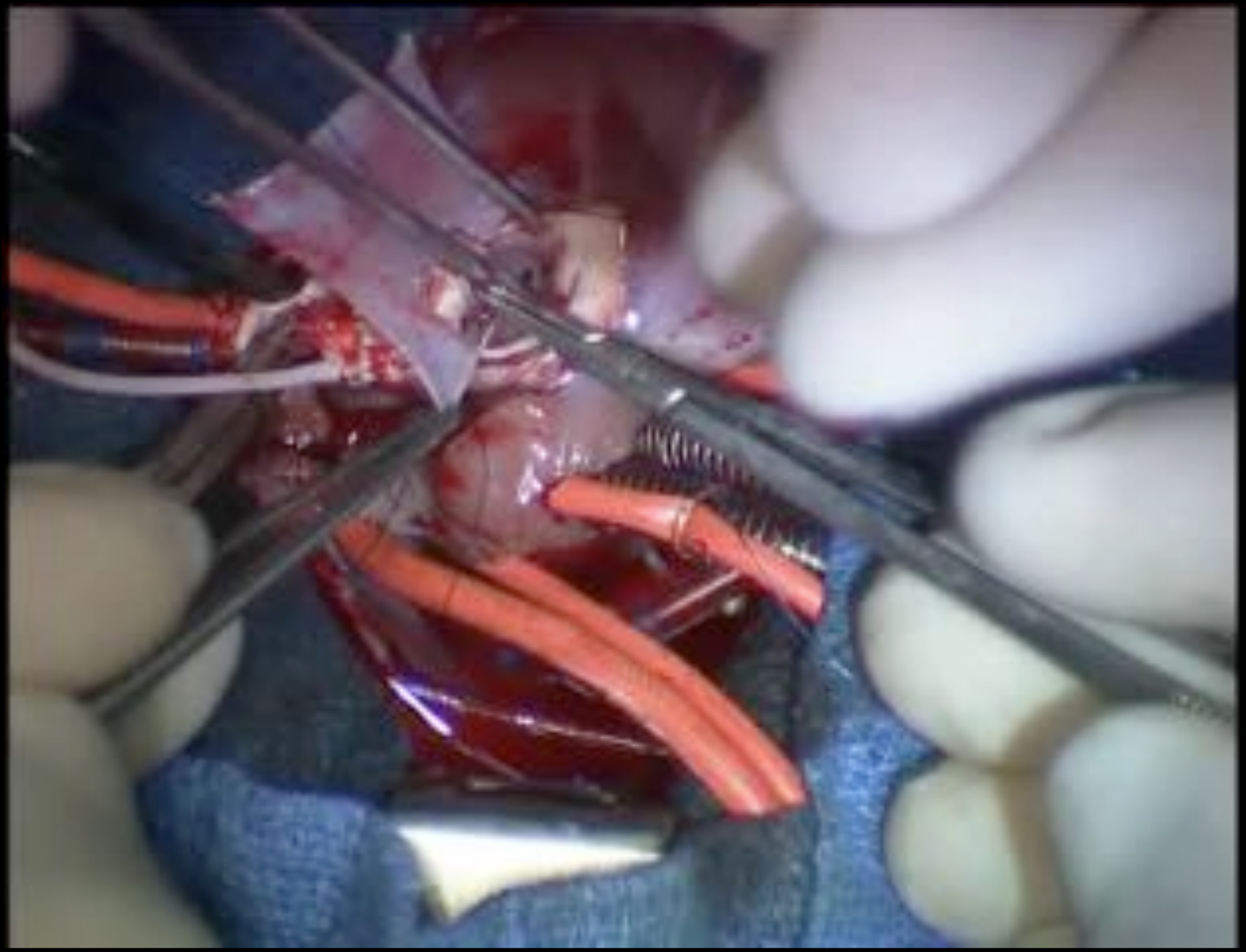


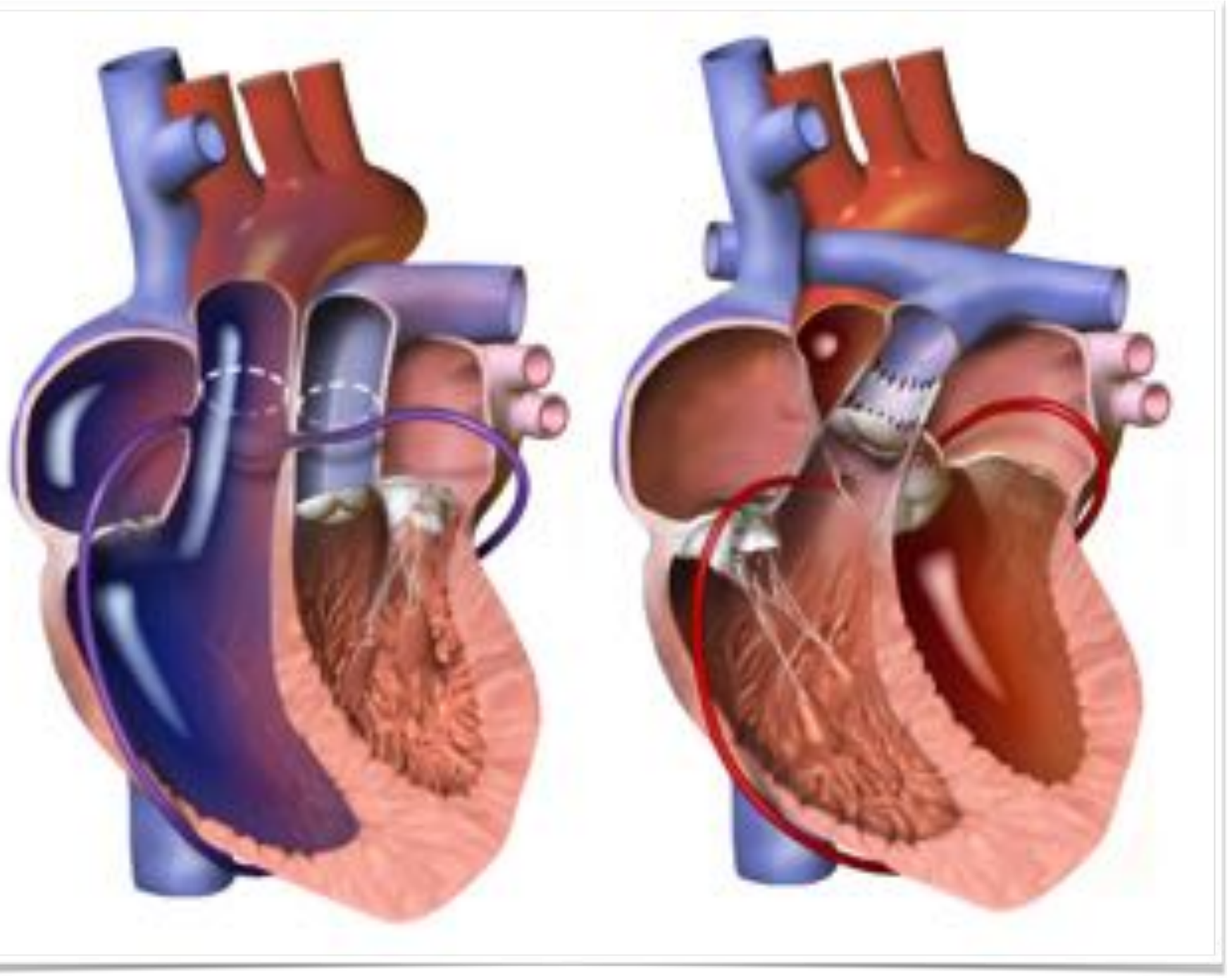




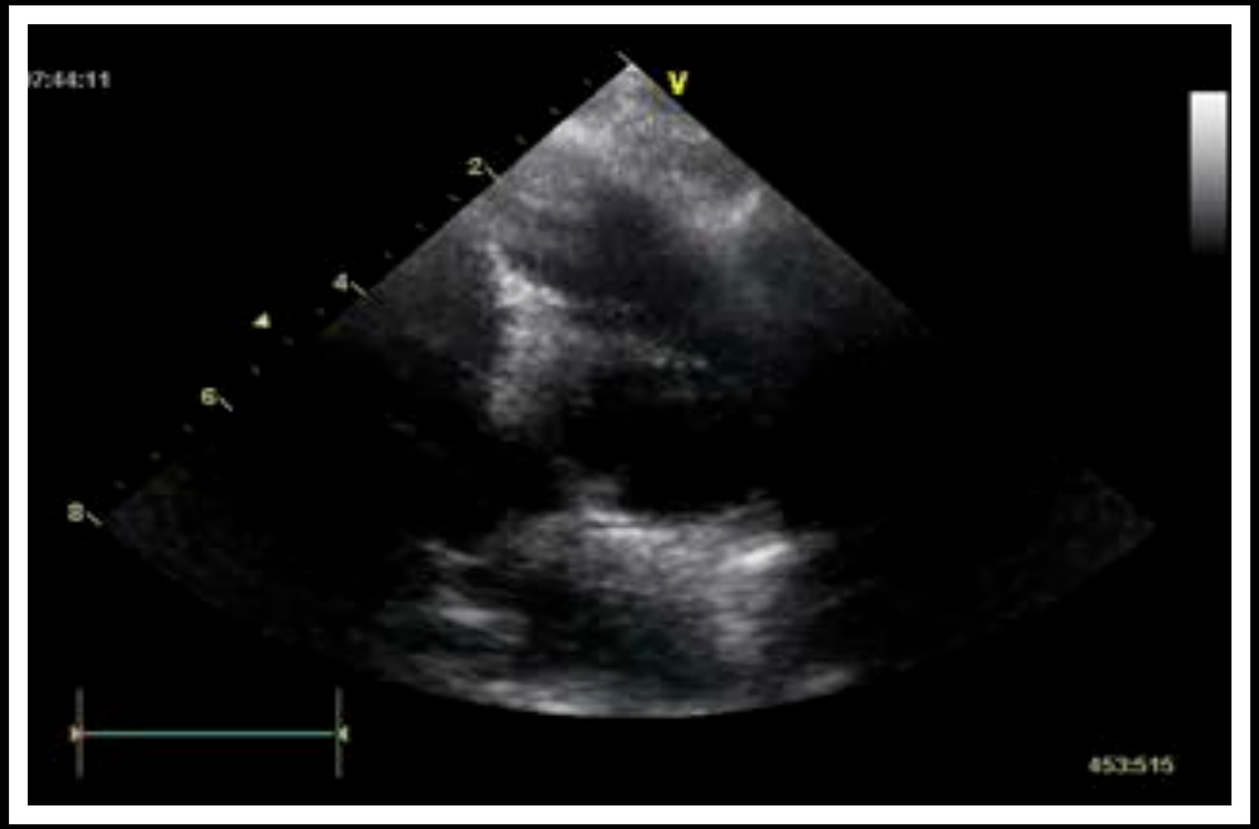


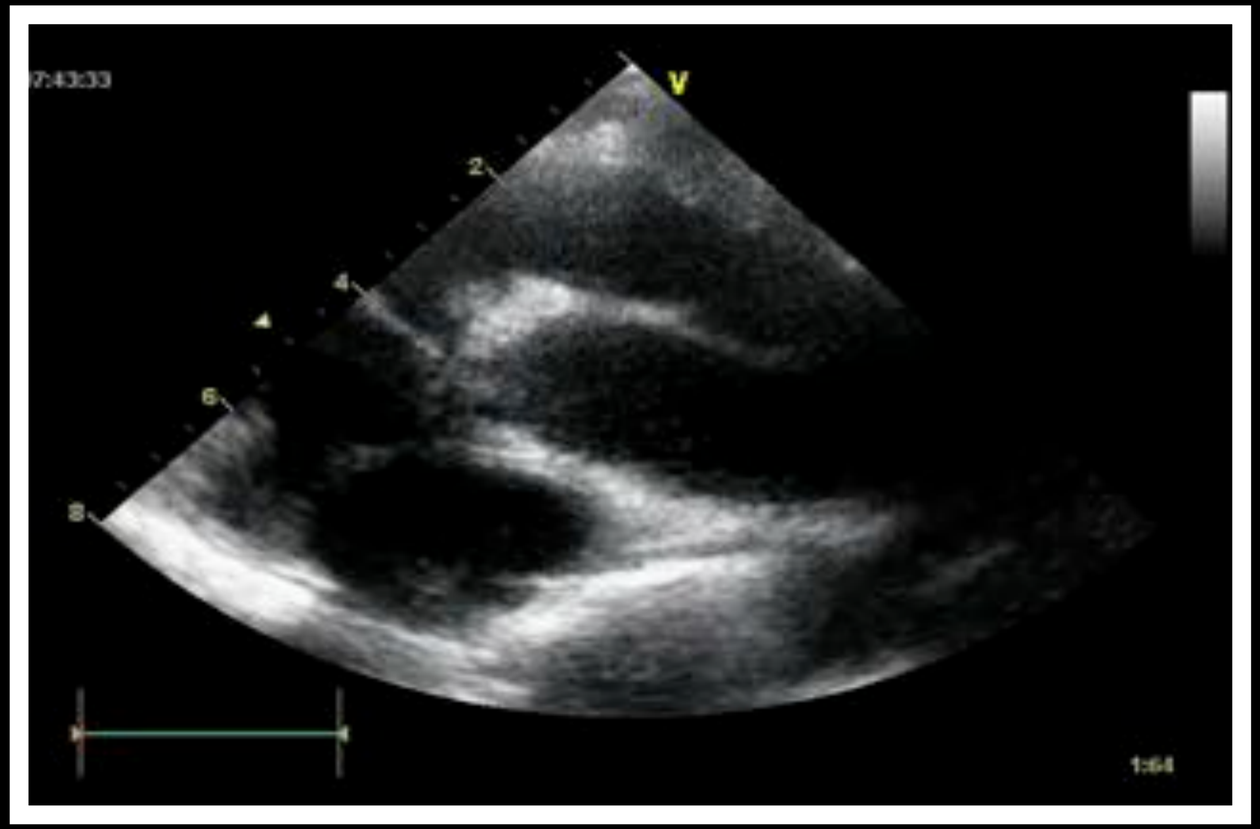




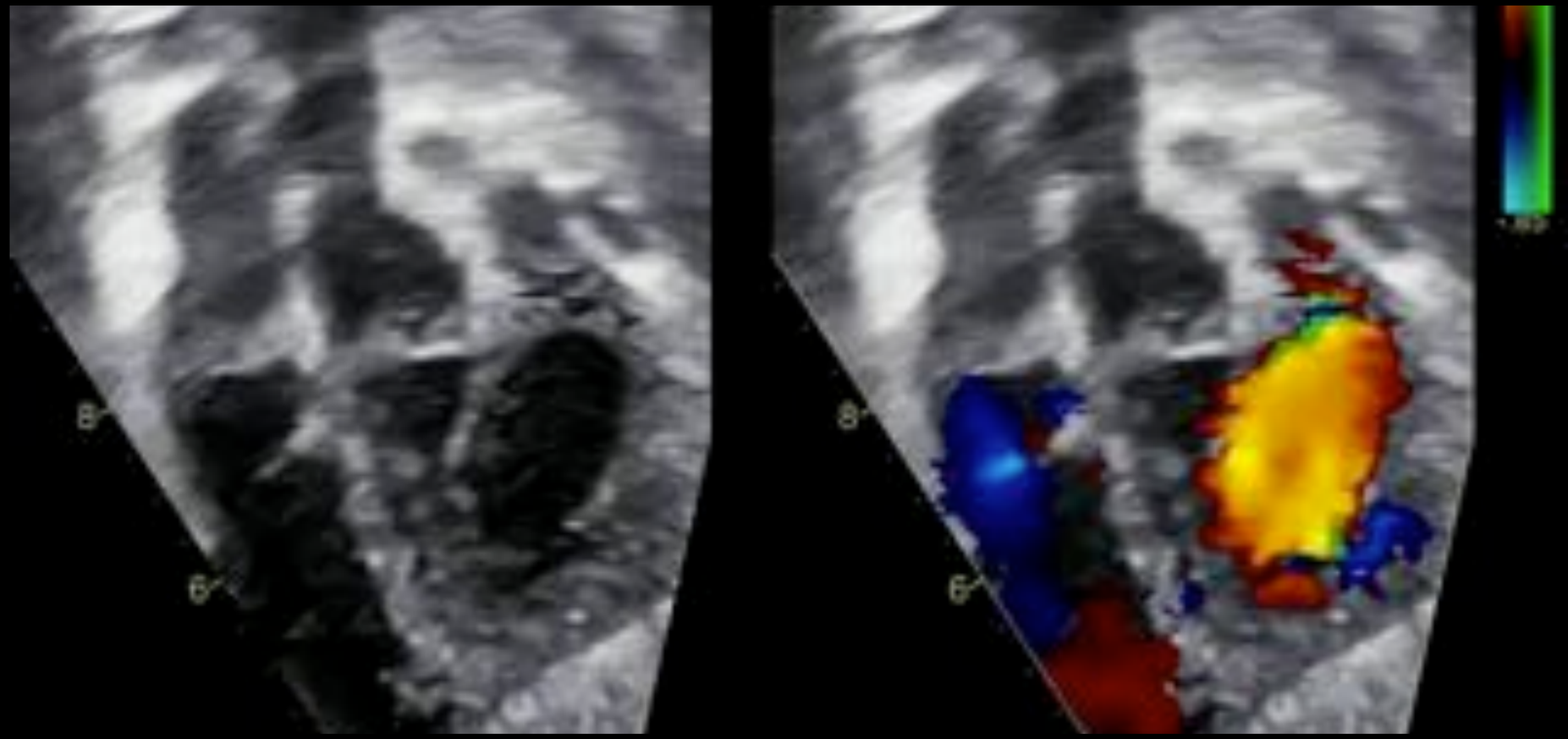


Outflow tracts after the arterial switch for TGA

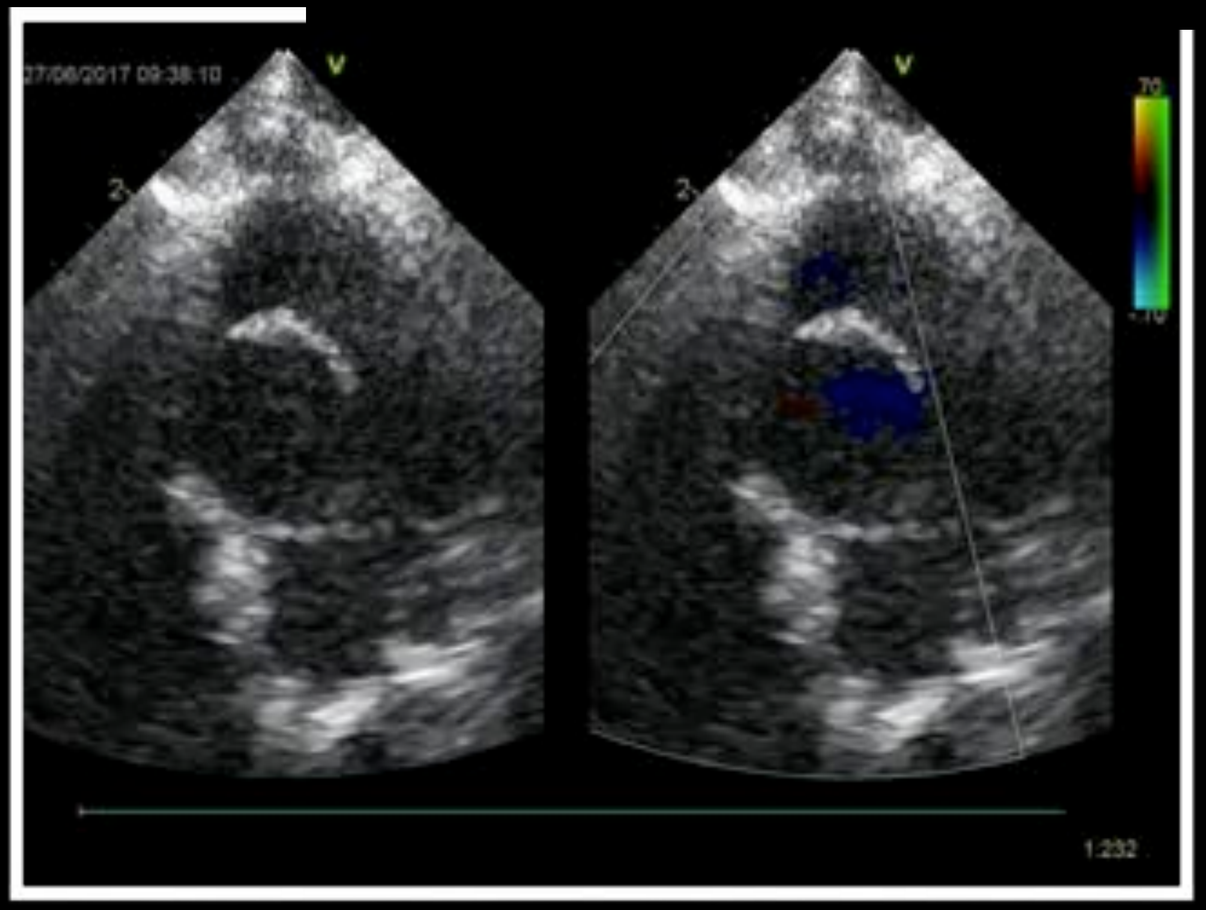


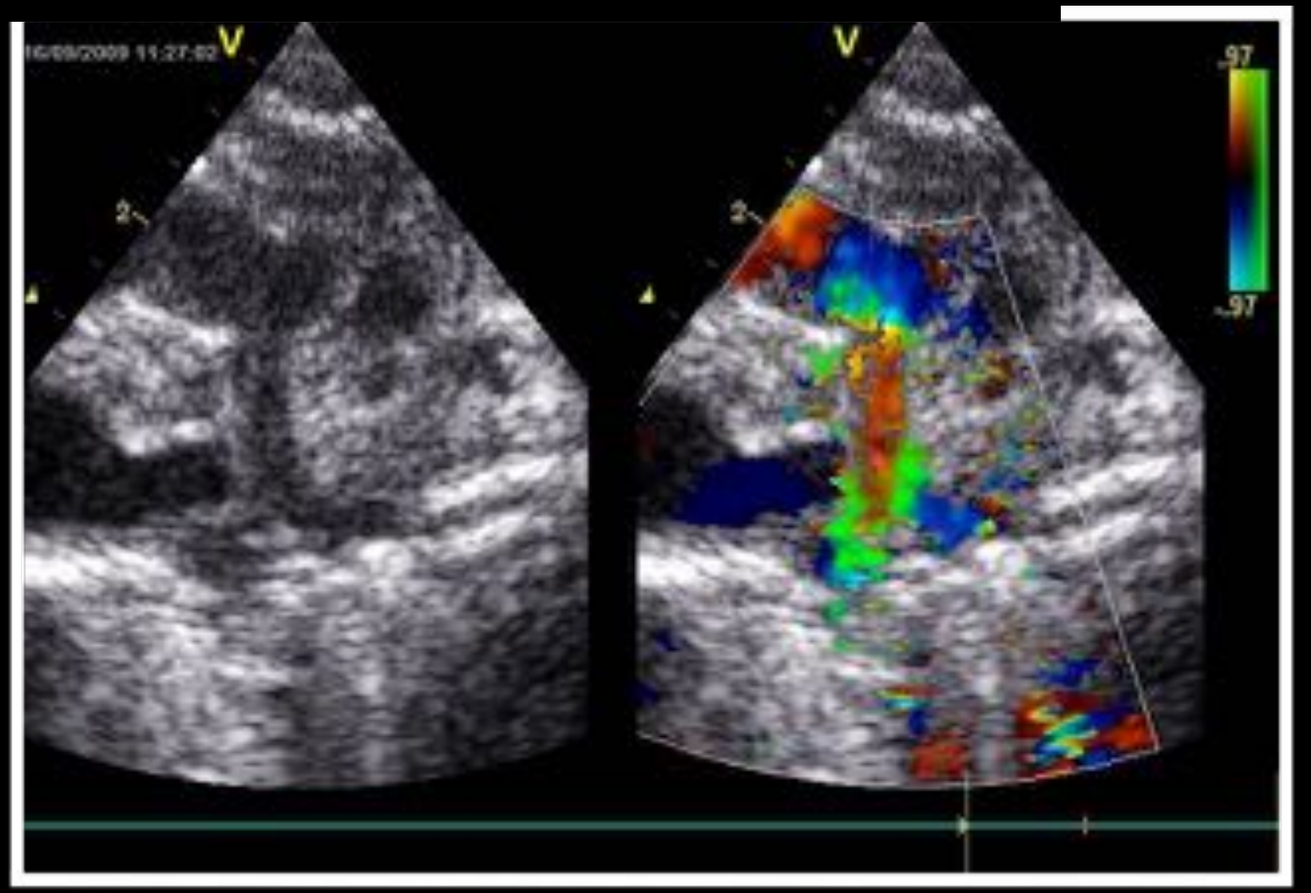


Outflow tracts after the arterial switch for TGA

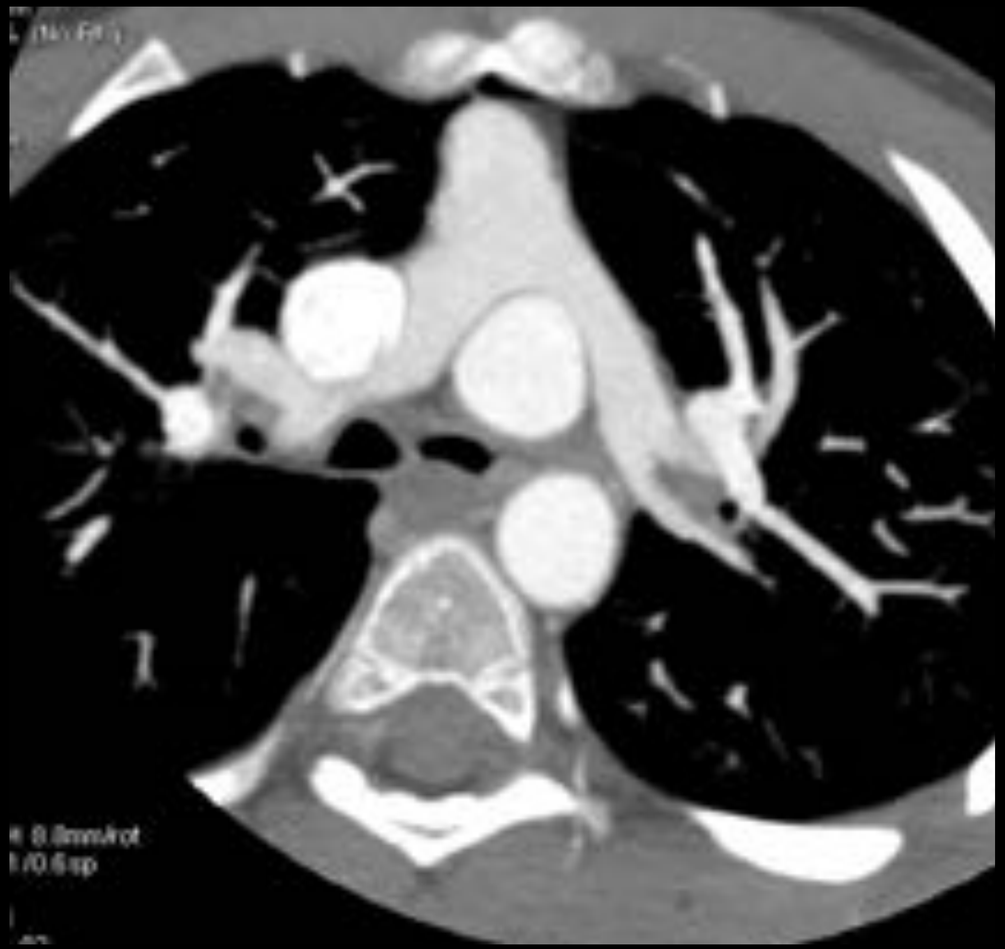


Outflow tracts after the arterial switch for TGA
The Lecompte manoeuver

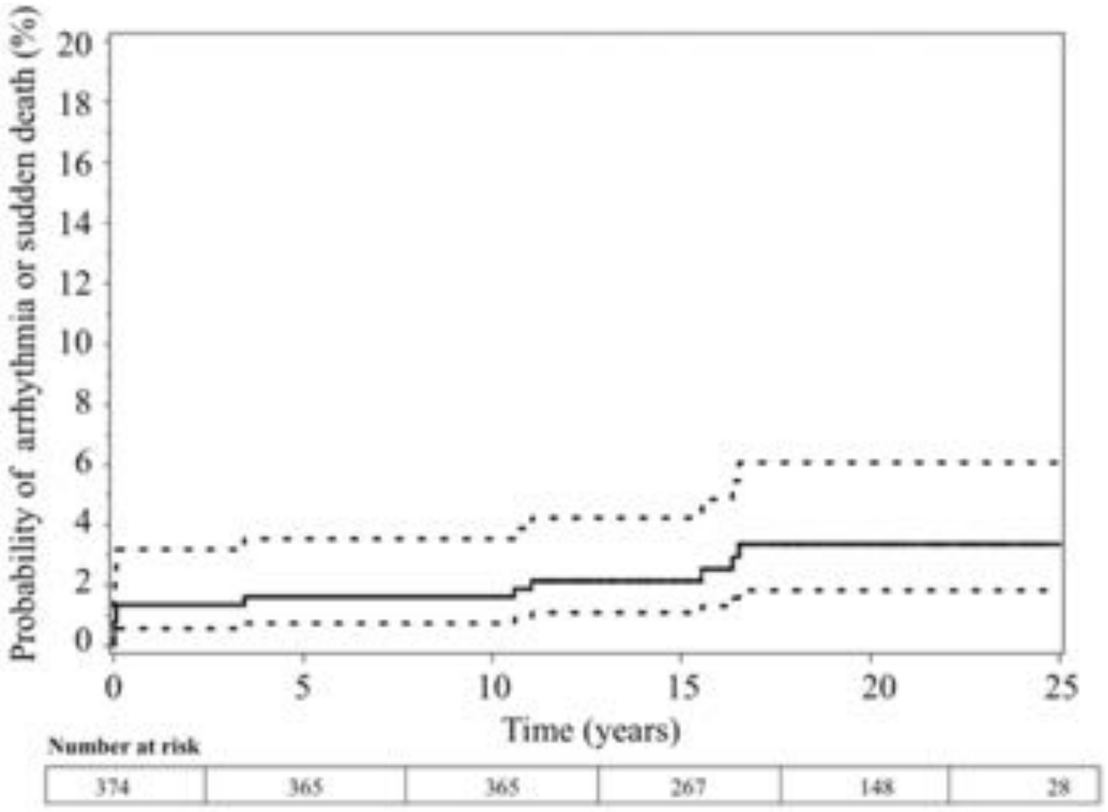




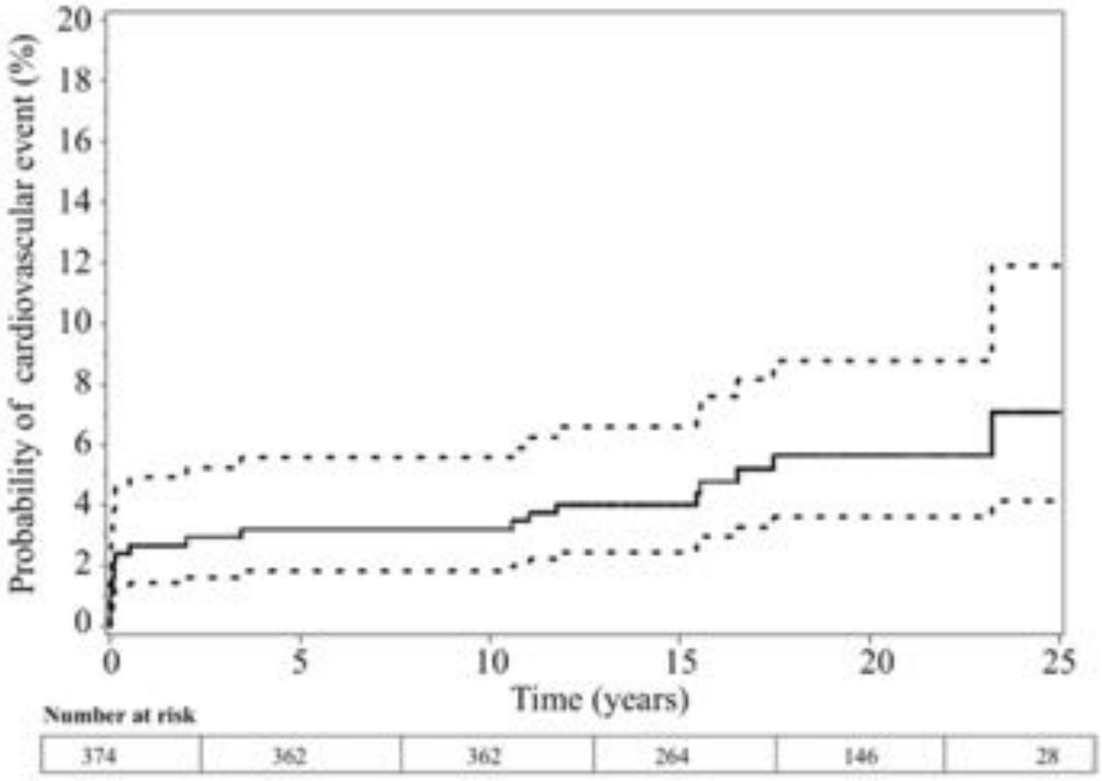
Outflow tracts after the arterial switch for TGA
The Lecompte manoeuver



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

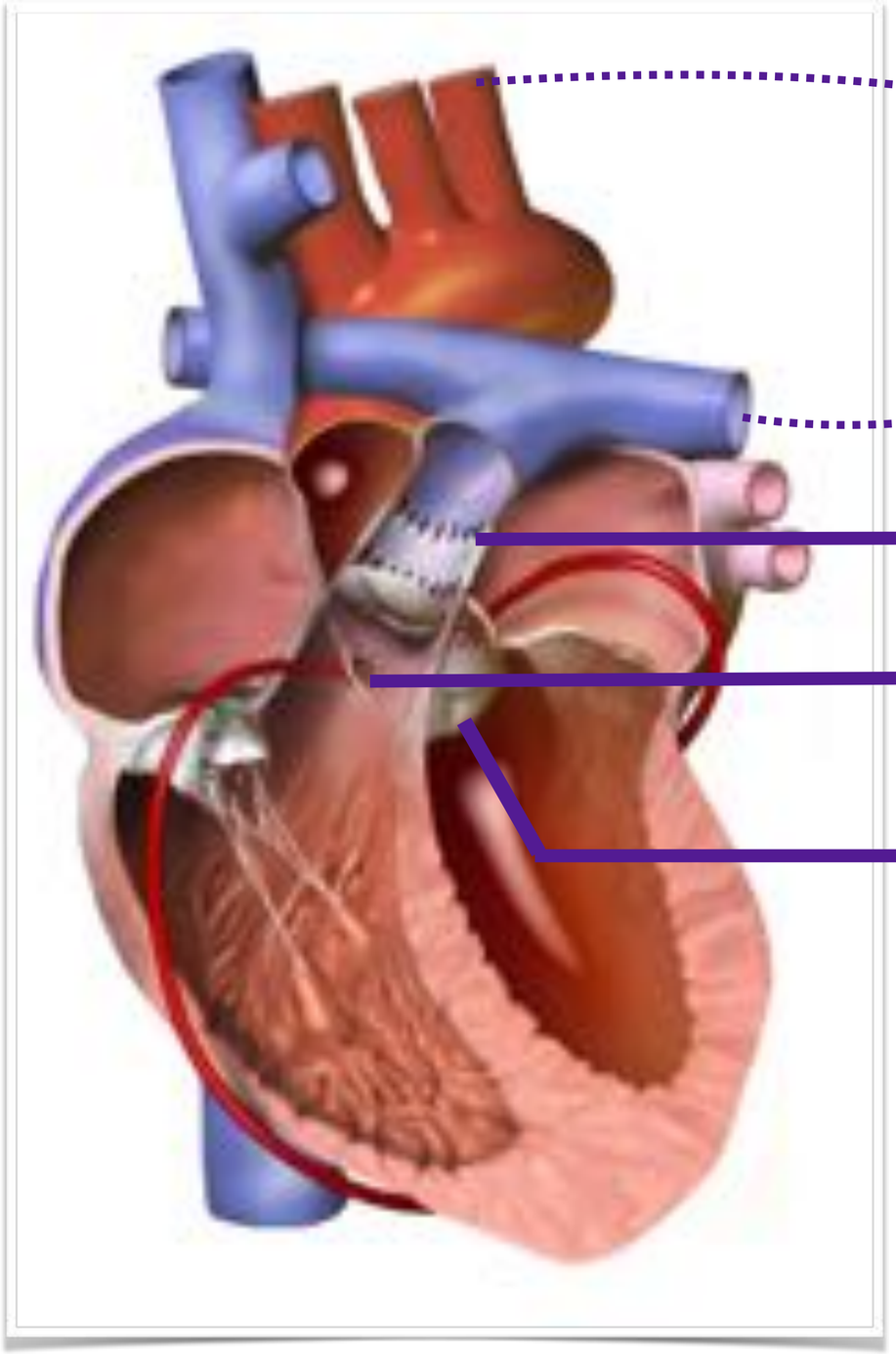
1) absence of current consensus regarding the appropriate interval and modality for surveillance imaging;

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

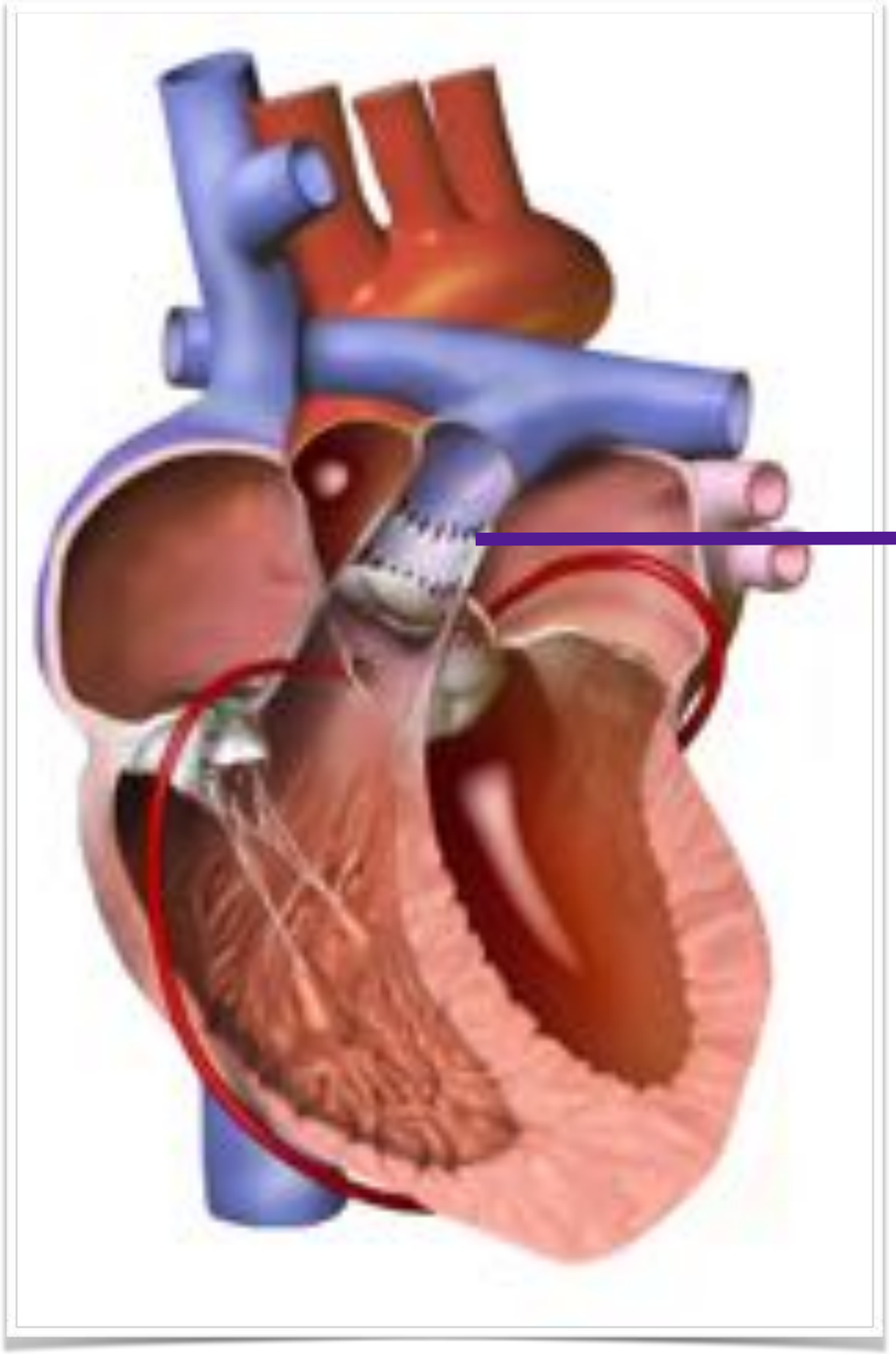
Coronary artery obstruction

Aortic valve regurgitation

Postoperative sequelae following the arterial switch operation

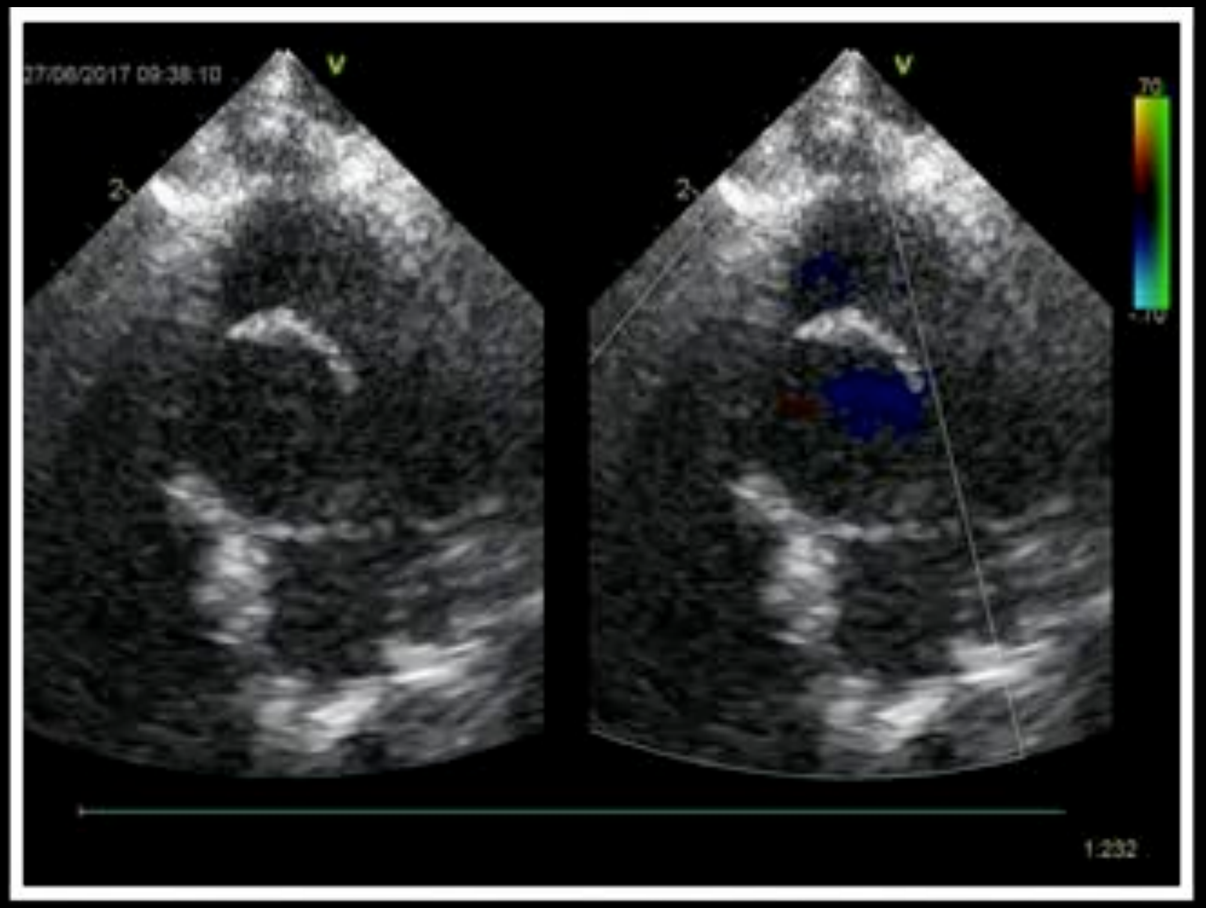
Prevalence of coronary lesions or potential coronary events

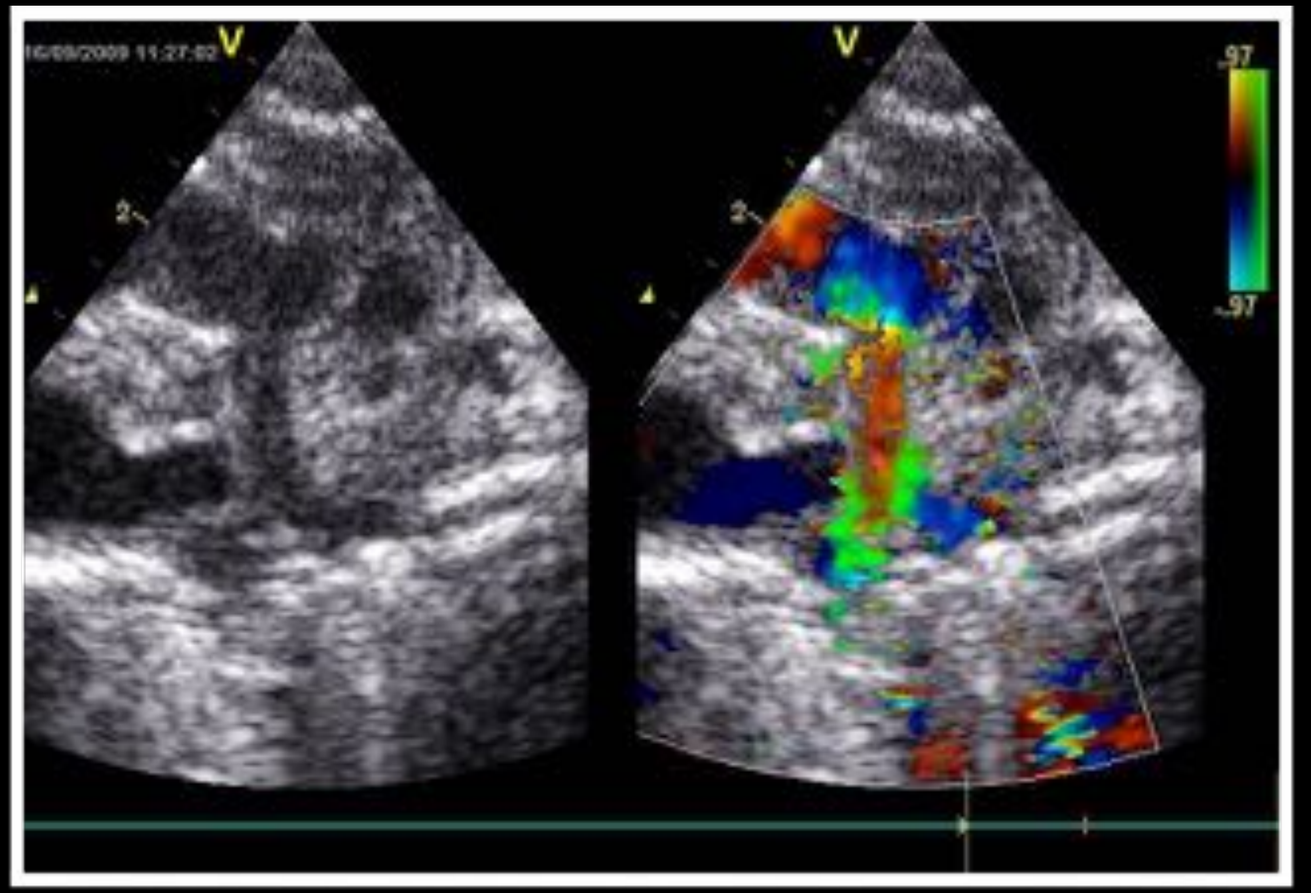
Long-Term Post-Operative Sequelae	Incidence
Supravalvular pulmonary stenosis*	~10%
Supravalvular aortic stenosis*	~5%
Neoaortic root dilation	Nearly universal
Neoaortic regurgitation	Most (moderate or severe in <10%)
Asymptomatic coronary occlusion	2%-7%
Sudden cardiac death	<1%
Arrhythmia	2%-10%
Aortic dissection or rupture	Unknown

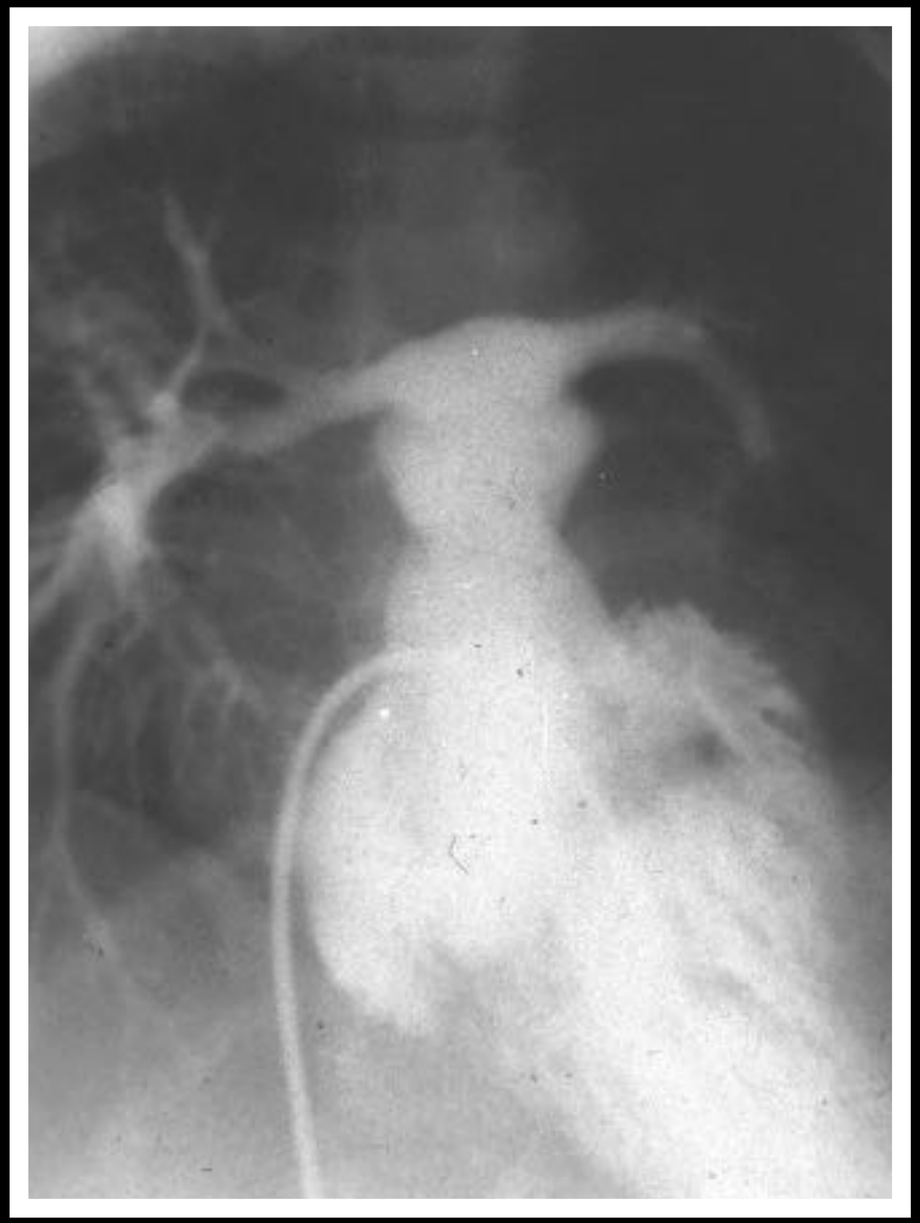


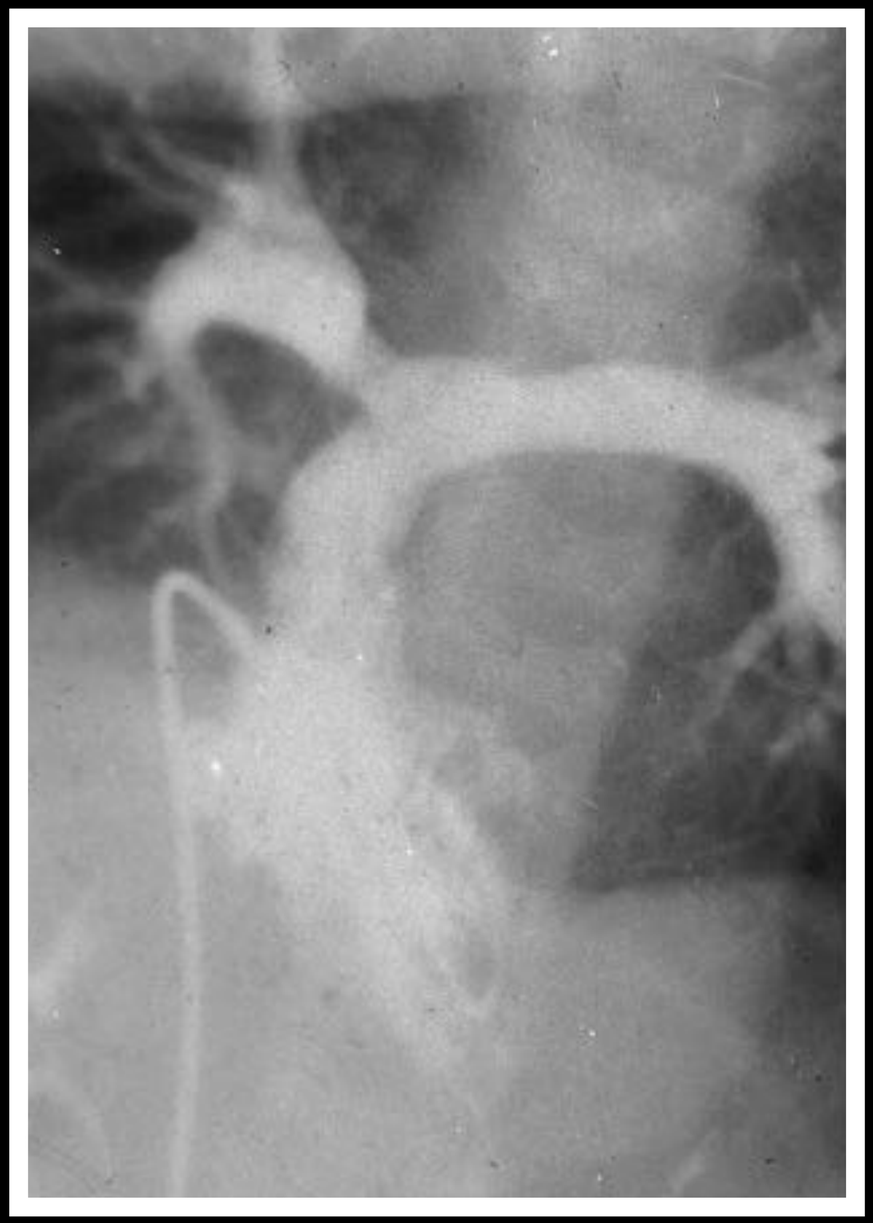
Pulmonary artery stenosis

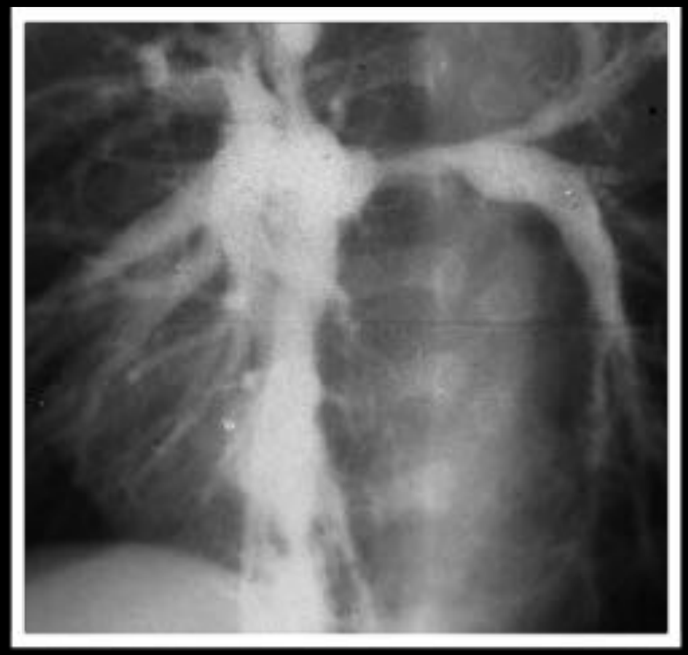
The Lecompte Manoeuver in ASO for TGA

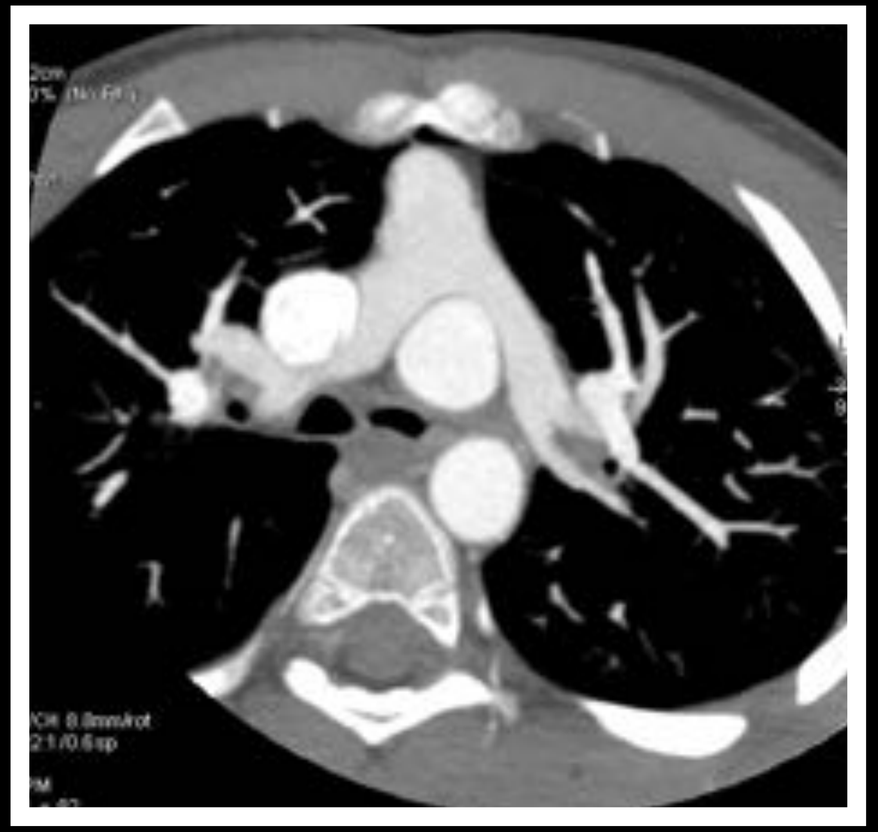


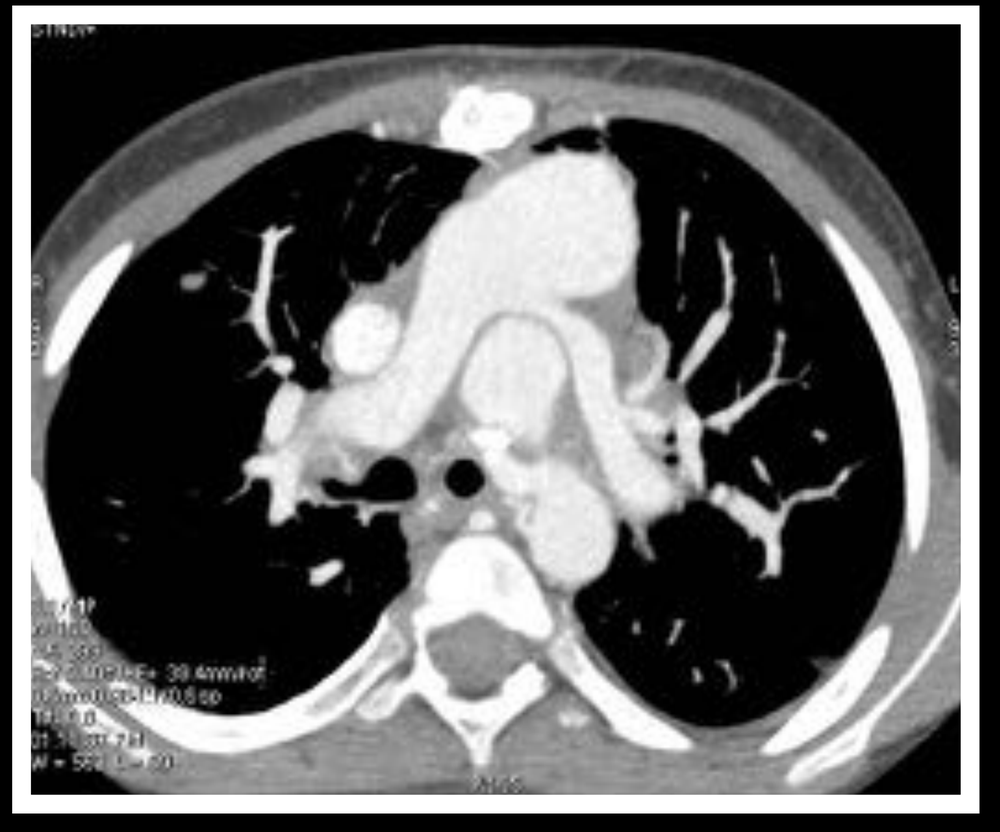


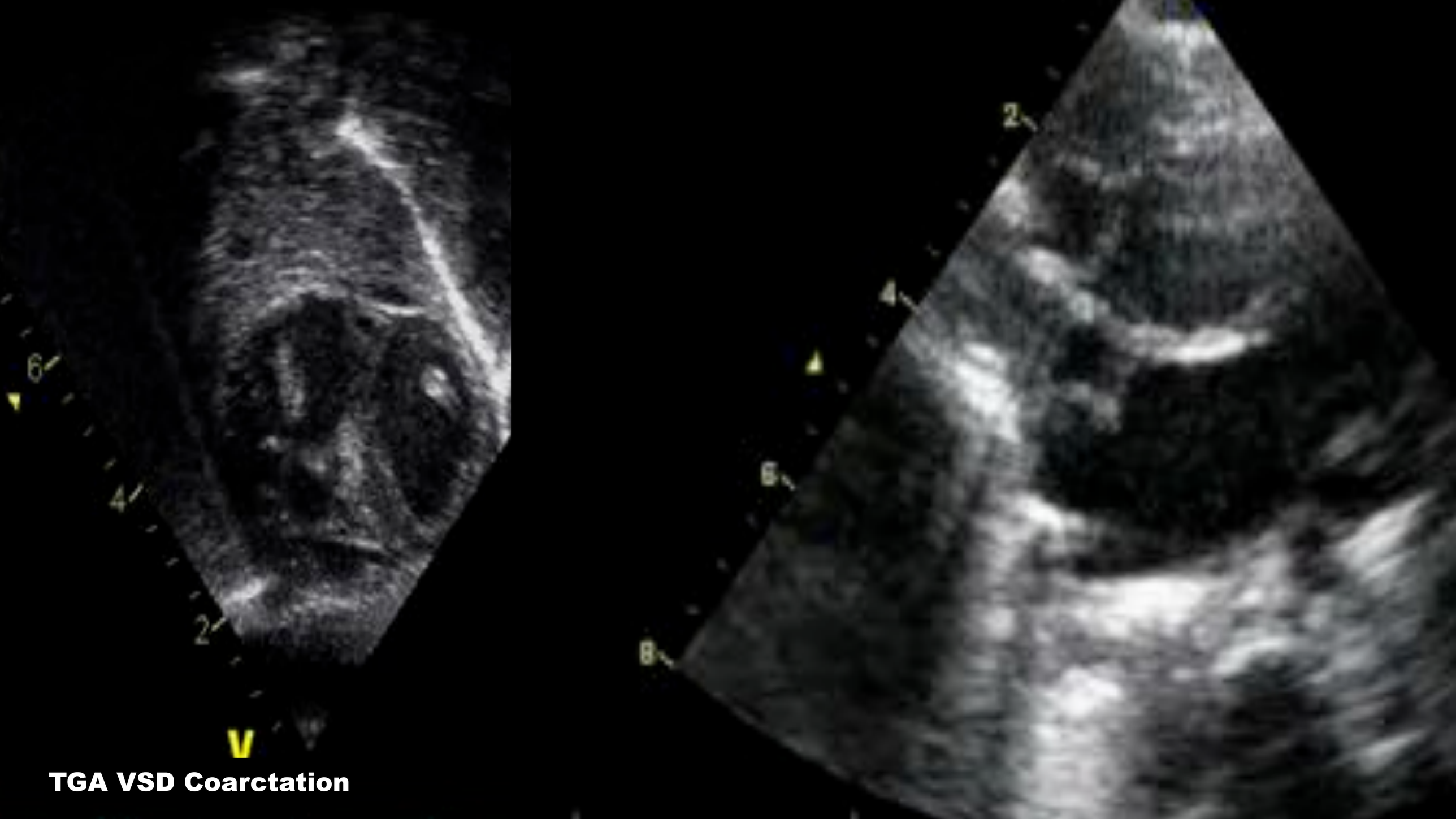




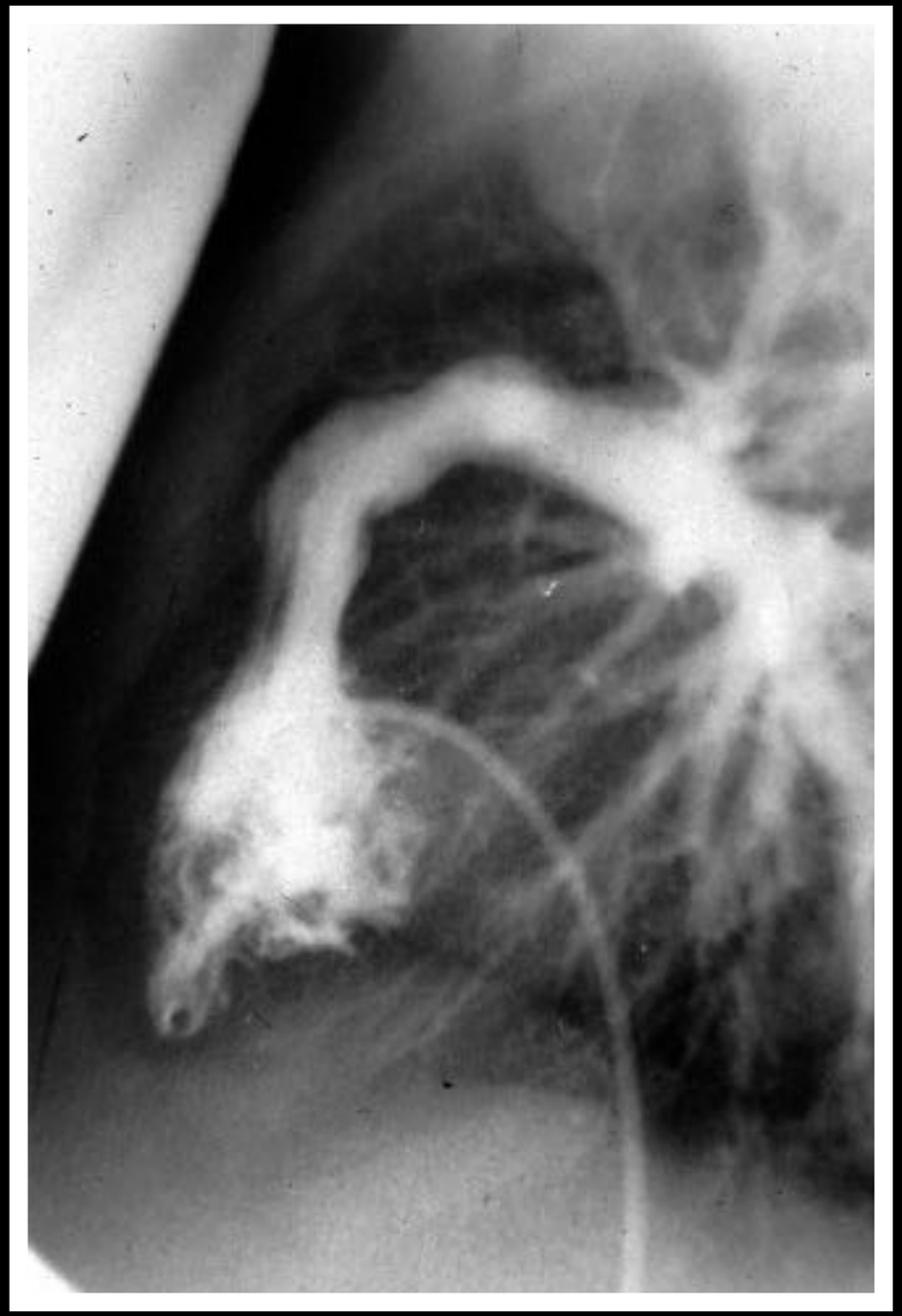


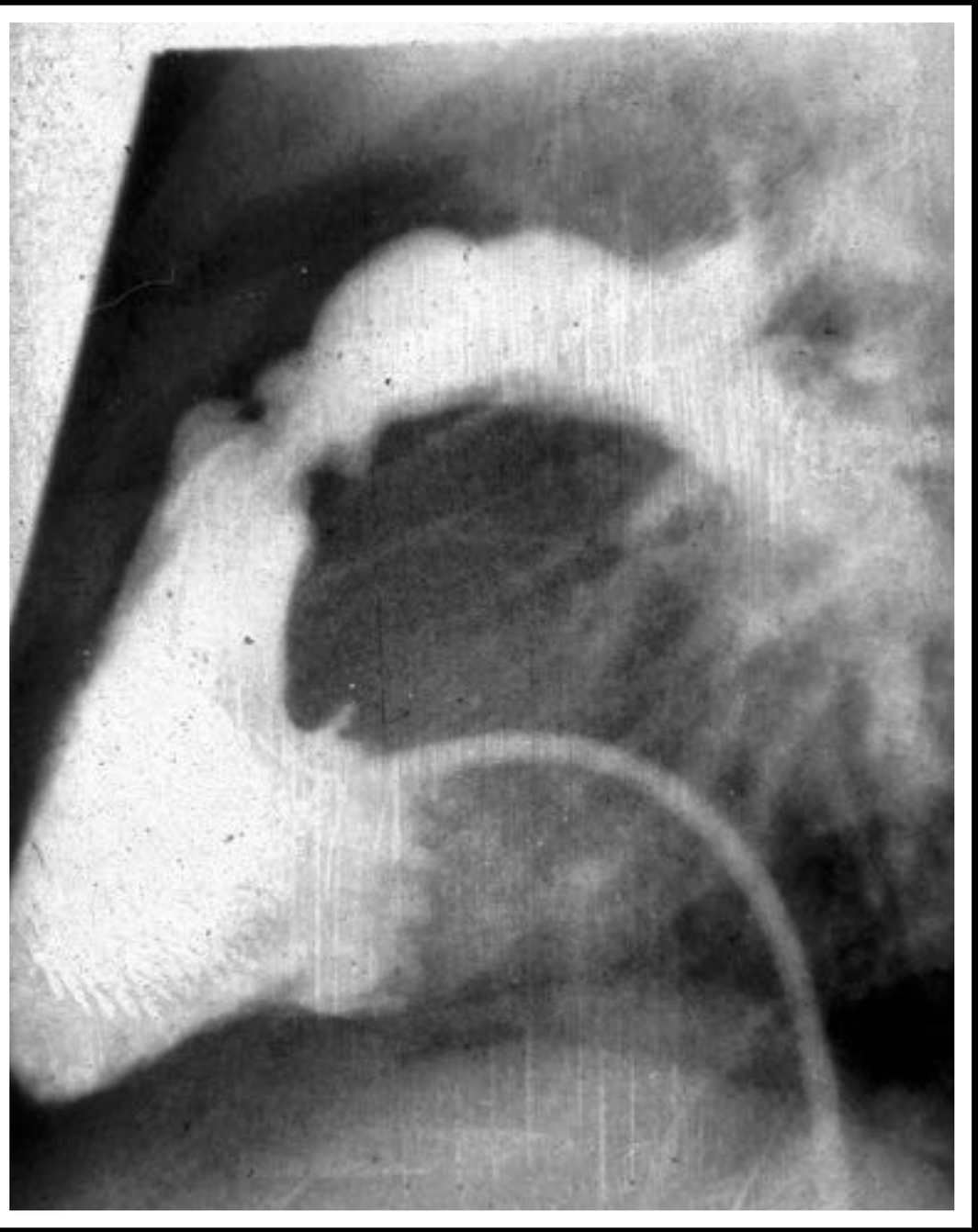


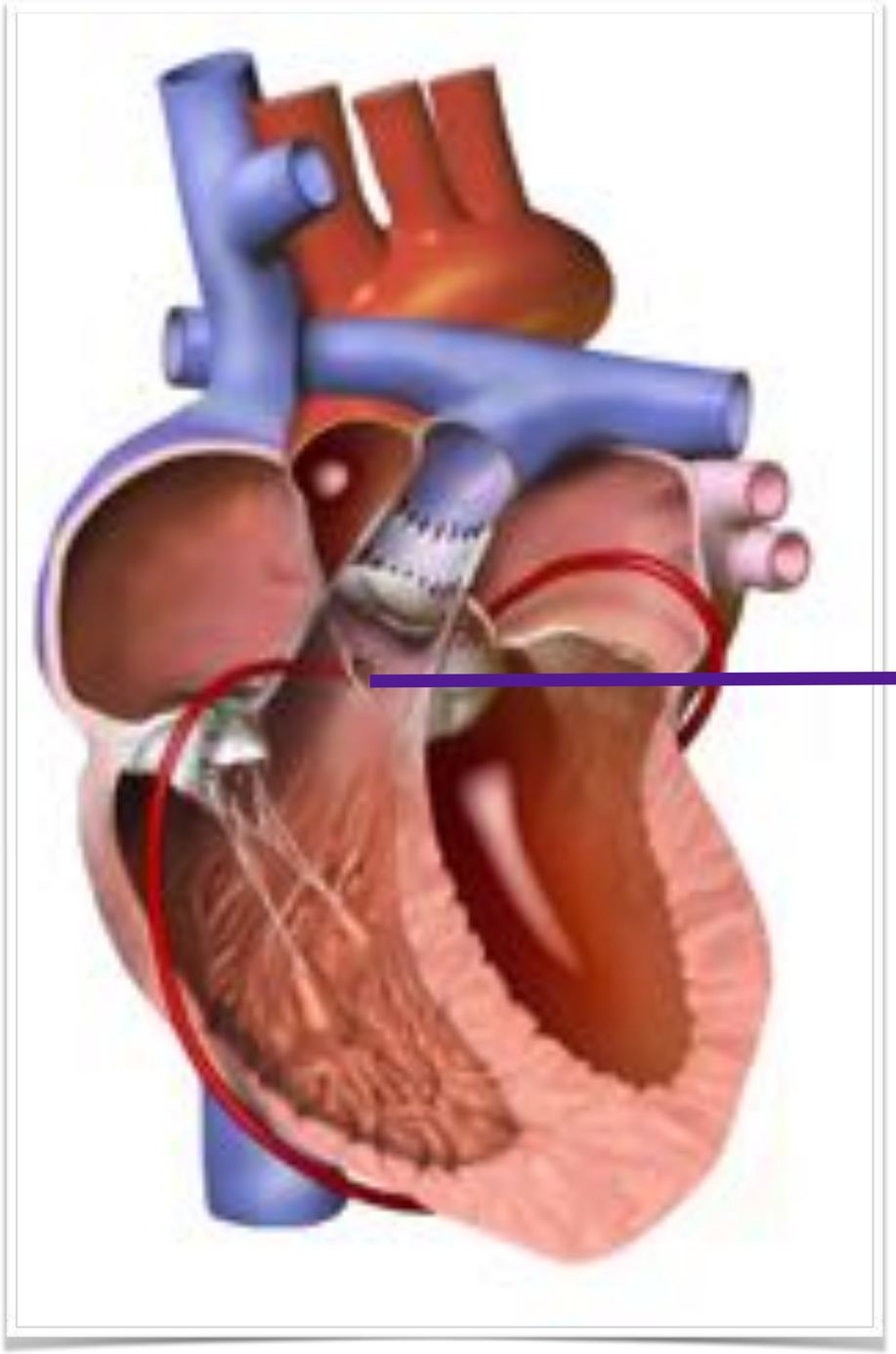




TGA VSD Coarctation







Coronary artery obstruction

Life-long management of the ASO population poses several challenges

The coronary arteries after transfer

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

What is the incidence of ischemic events after the ASO for TGA ?

When do they occur ? Do we know risk factors ?

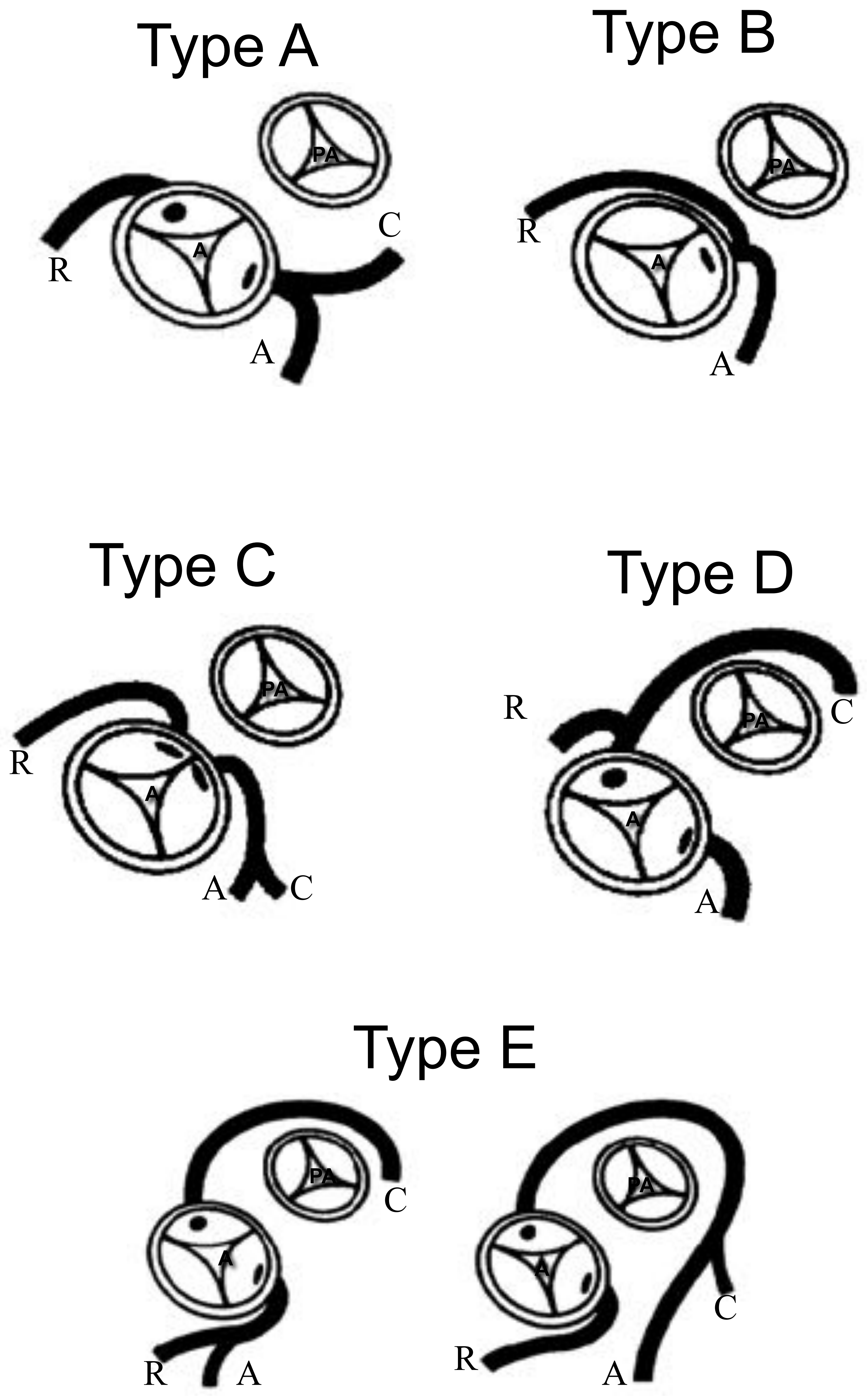
2) absence of current consensus regarding the appropriate interval and modality for surveillance imaging;

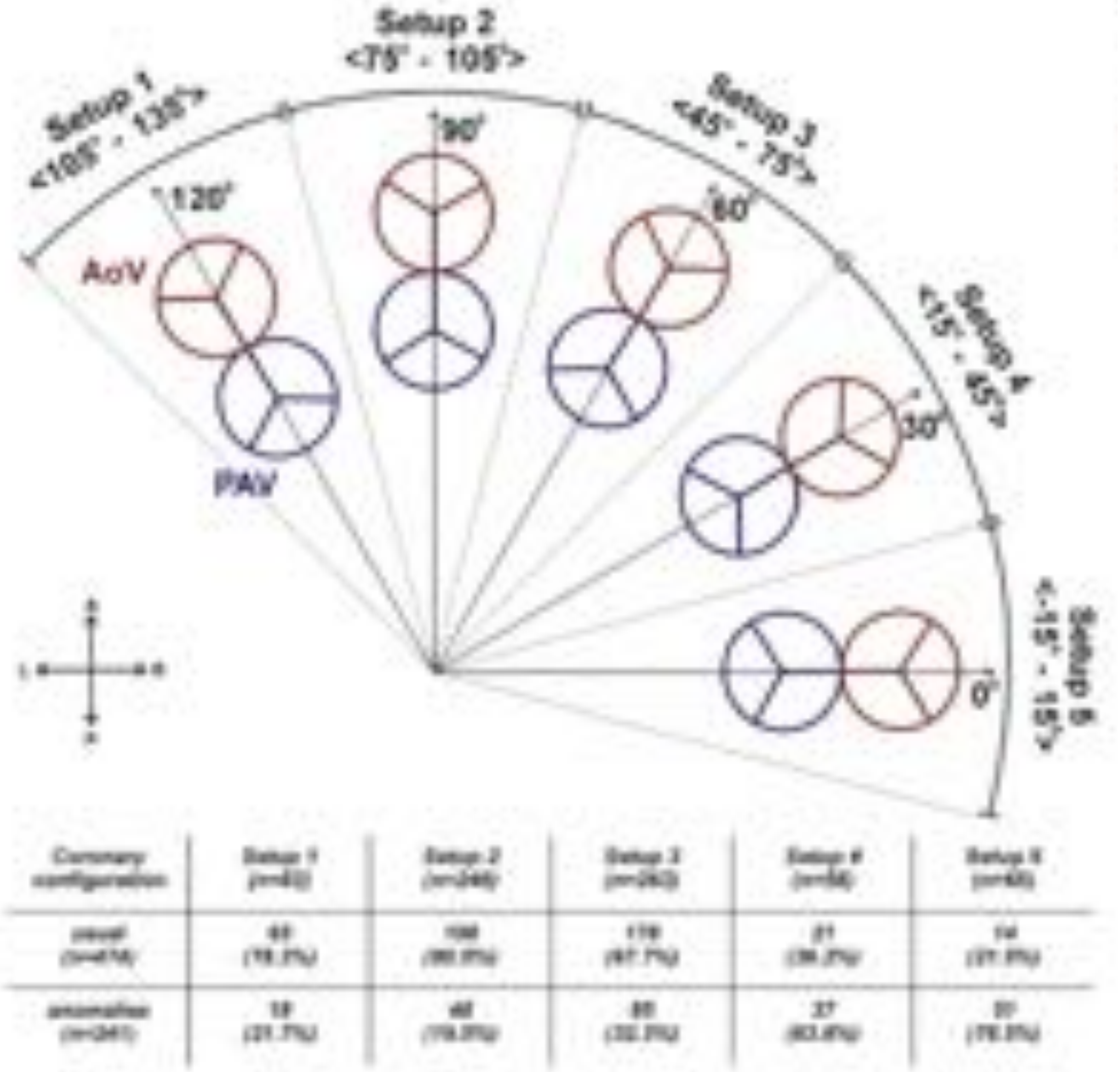
Is systematic screening for coronary artery obstruction useful after the ASO for TGA ?

When ? How to screen ?

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

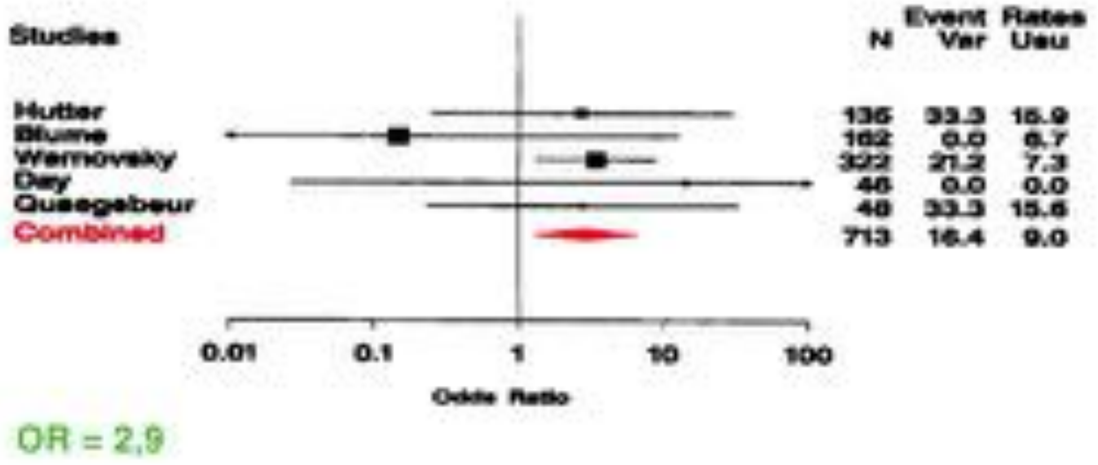
Big variations in coronary anatomy (origin, loops, epicardial, intramural)



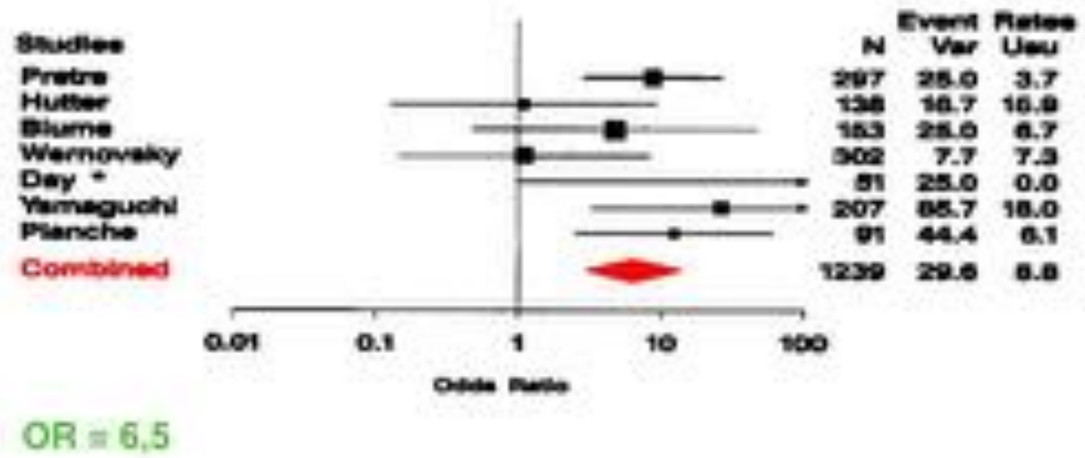


Coronary artery anatomy as a risk factor for early events

Single coronary artery



Intramural course



Pasquali, 2002

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

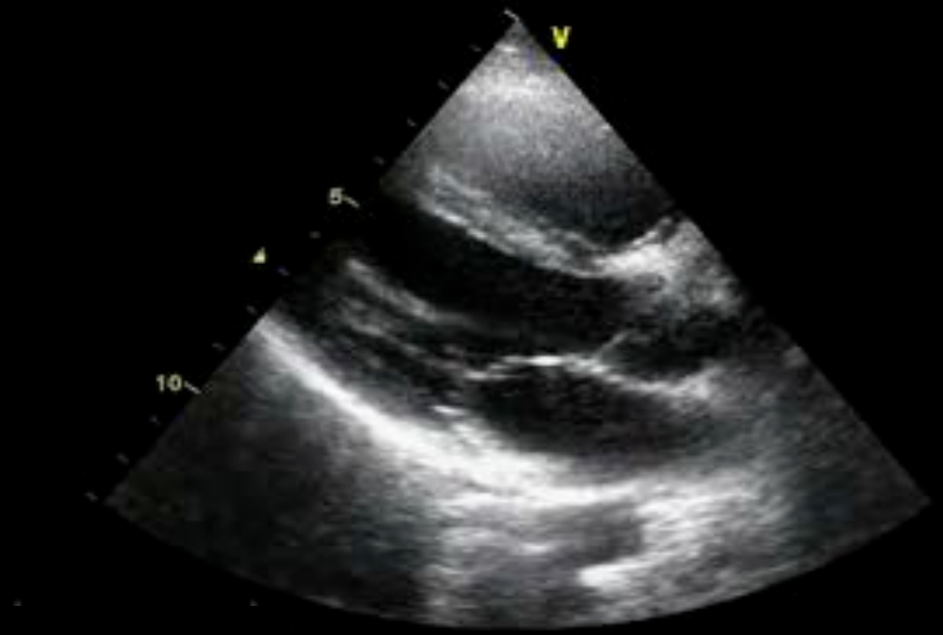
First Author (Ref. #), Year	Inclusive Years	N	% IVS	Early Survival for TGA IVS, %	5-Year Survival, %	10-Year Survival, %	Coronary Anatomic Risk Factors	Other Predictors of Early Mortality
Sarris (43), 2006*	1998-2000	613	70	97	NA	NA	Single coronary (univariate analysis only)	Open sternum
Lalezari (51), 2011	1977-2007	332	60.8	88.6	85.8†	85.2†	Not a risk factor for early mortality	Technical problems with coronary transfer
Fricke (85), 2012	1983-2009	618	64	98.2	98	97	Not a risk factor for early mortality	Weight <2.5 kg ECMO
Khairy (41), 2013	1983-1999	400	59.5	93.5†	NA	92.7†	Single right coronary artery	Post-operative heart failure
Cain (52), 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 (24), 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

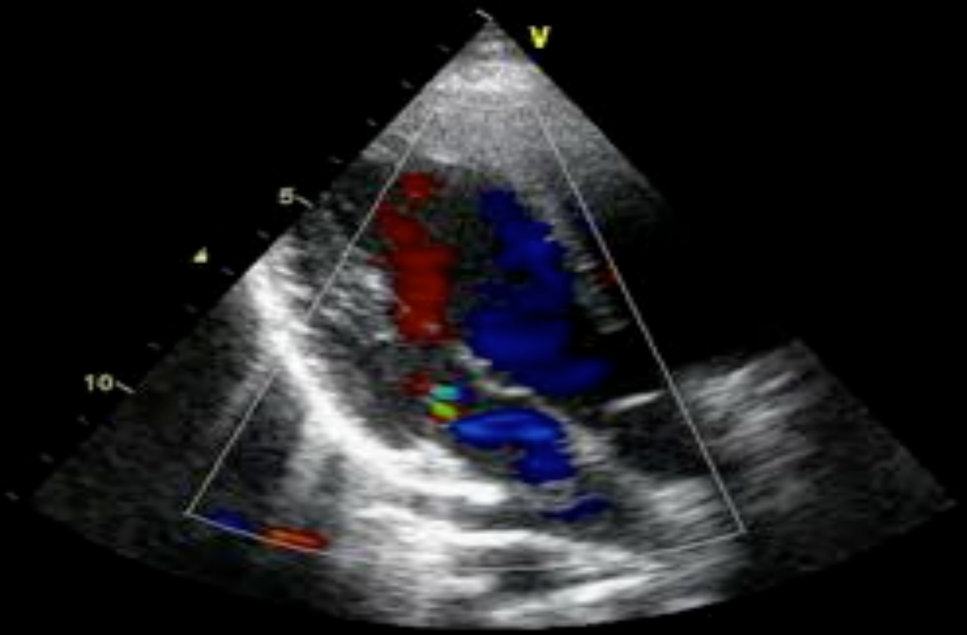
Postoperative sequelae following the arterial switch operation

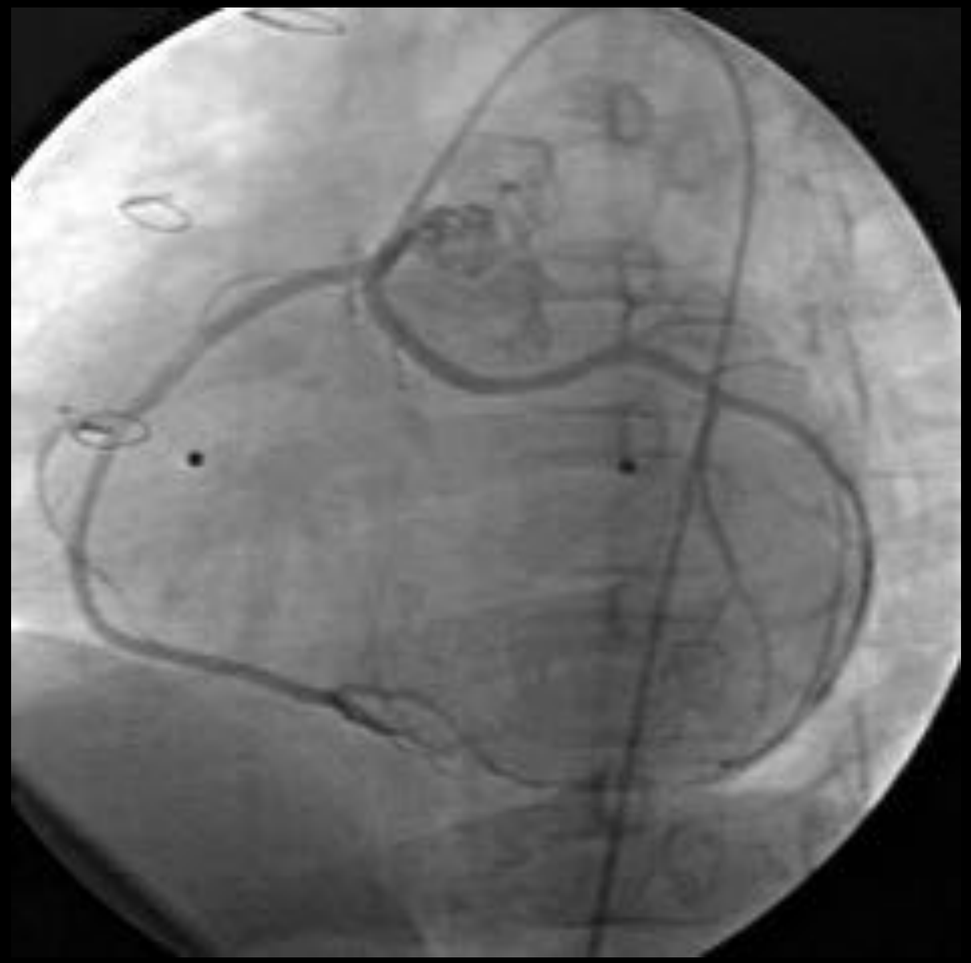
Prevalence of coronary lesions or potential coronary events

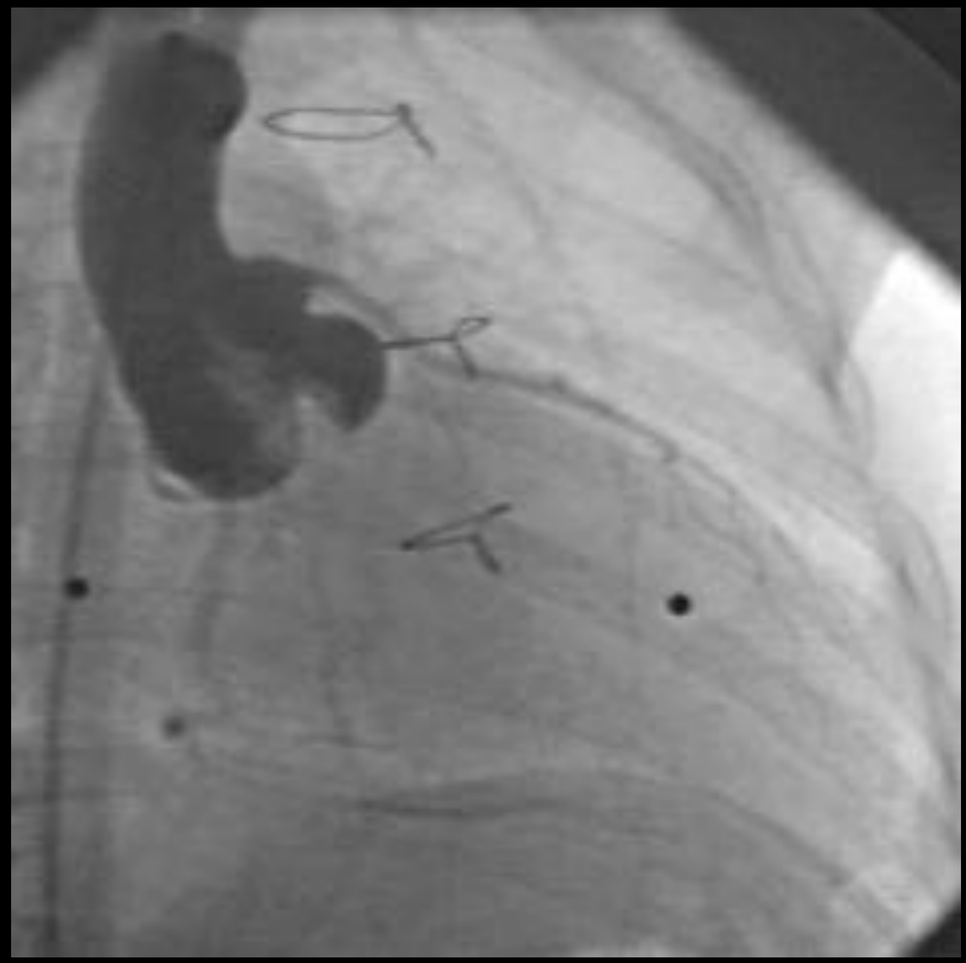
Long-Term Post-Operative Sequelae	Incidence
Supravalvular pulmonary stenosis*	~10%
Supravalvular aortic stenosis*	~5%
Neoaortic root dilation	Nearly universal
Neoaortic regurgitation	Most (moderate or severe in <10%)
Asymptomatic coronary occlusion	2%-7%
Sudden cardiac death	<1%
Arrhythmia	2%-10%
Aortic dissection or rupture	Unknown

Myocardial ischemia after the arterial switch for TGA





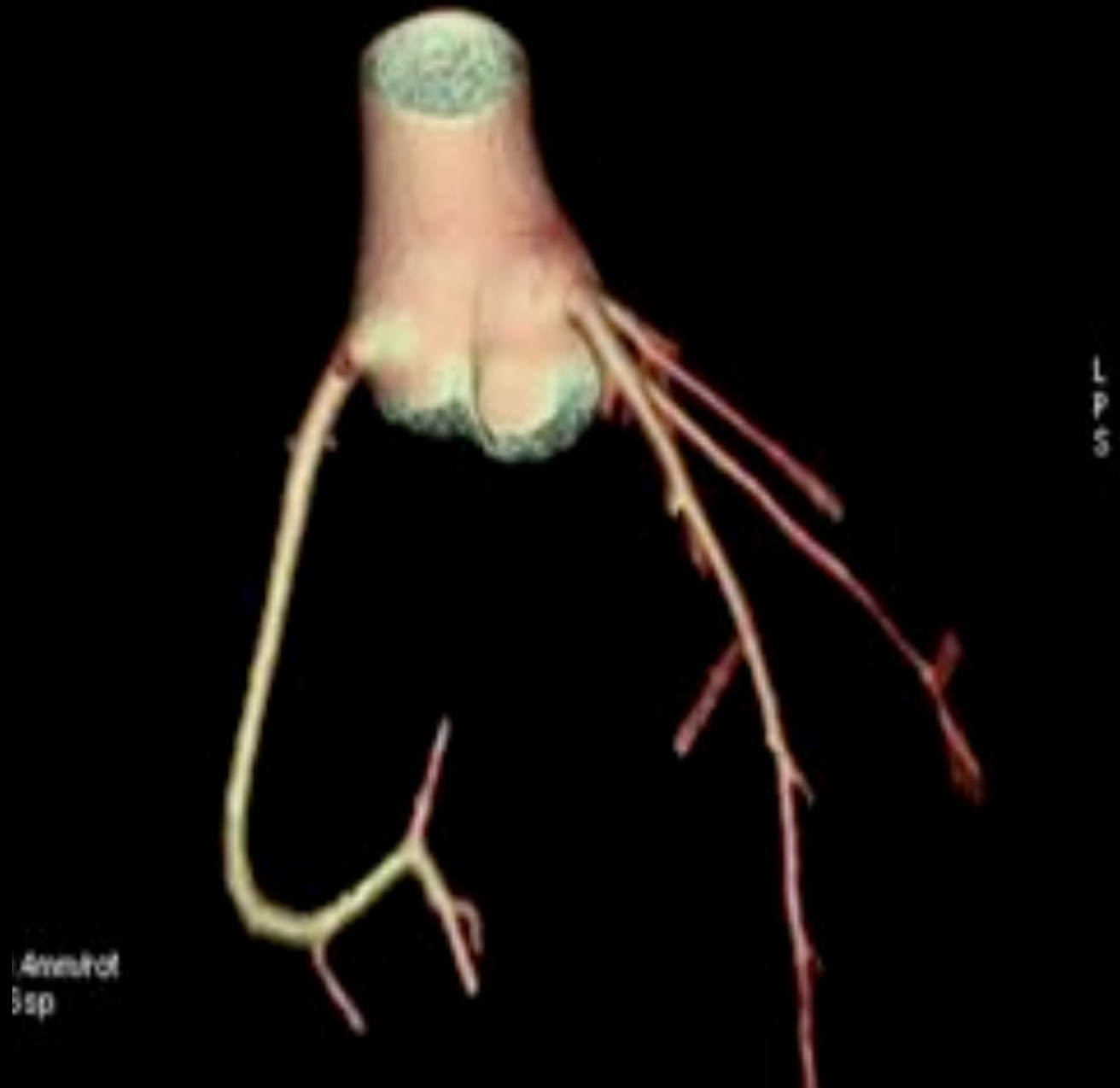




Type D coronary arteries after the arterial switch for TGA

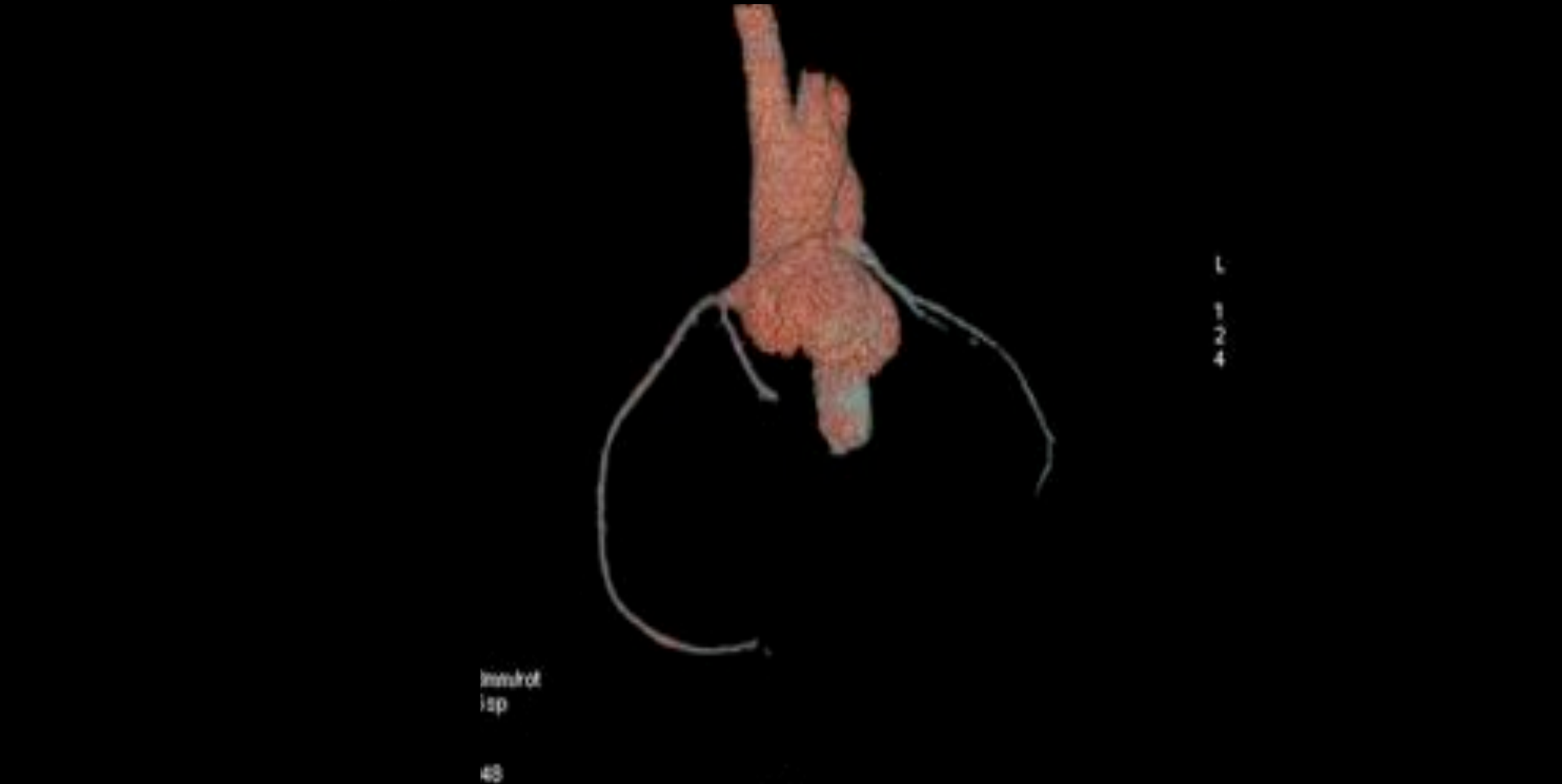
Coronary arteries after the arterial switch for TGA

High irradiation CT



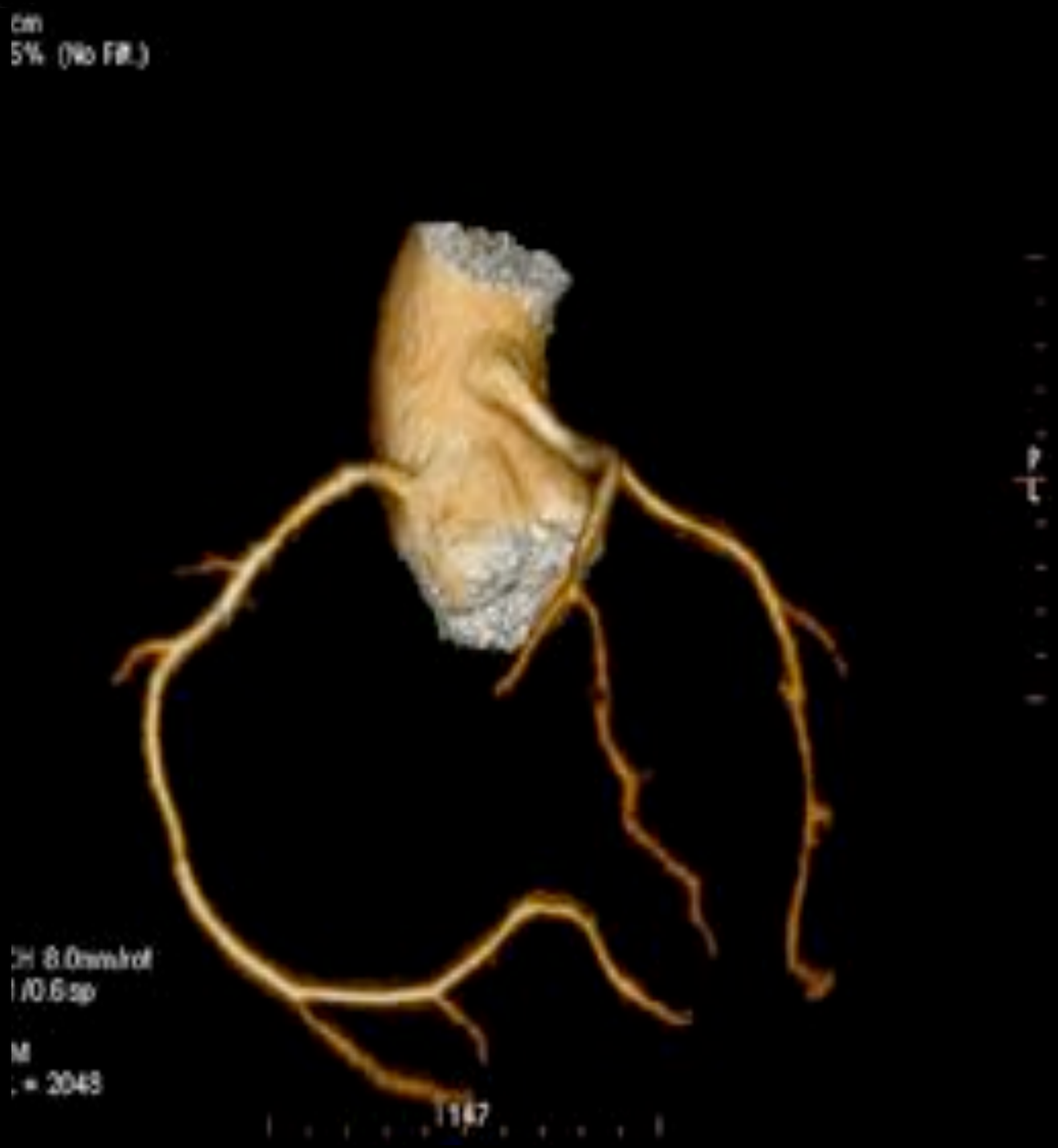
Coronary arteries after the arterial switch for TGA

Very low dose CT



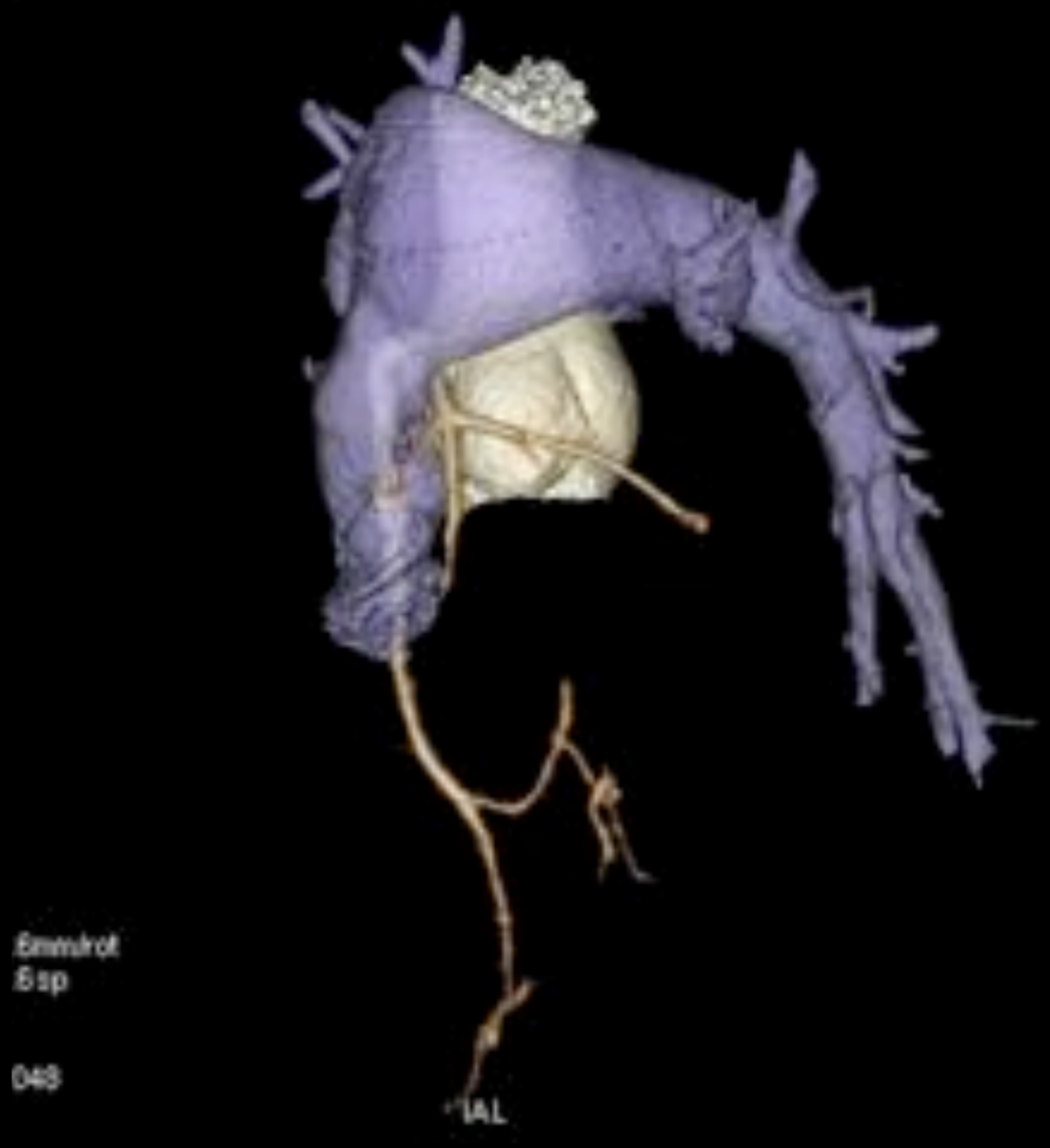
Coronary arteries after the arterial switch for TGA

High reimplantation of left CA

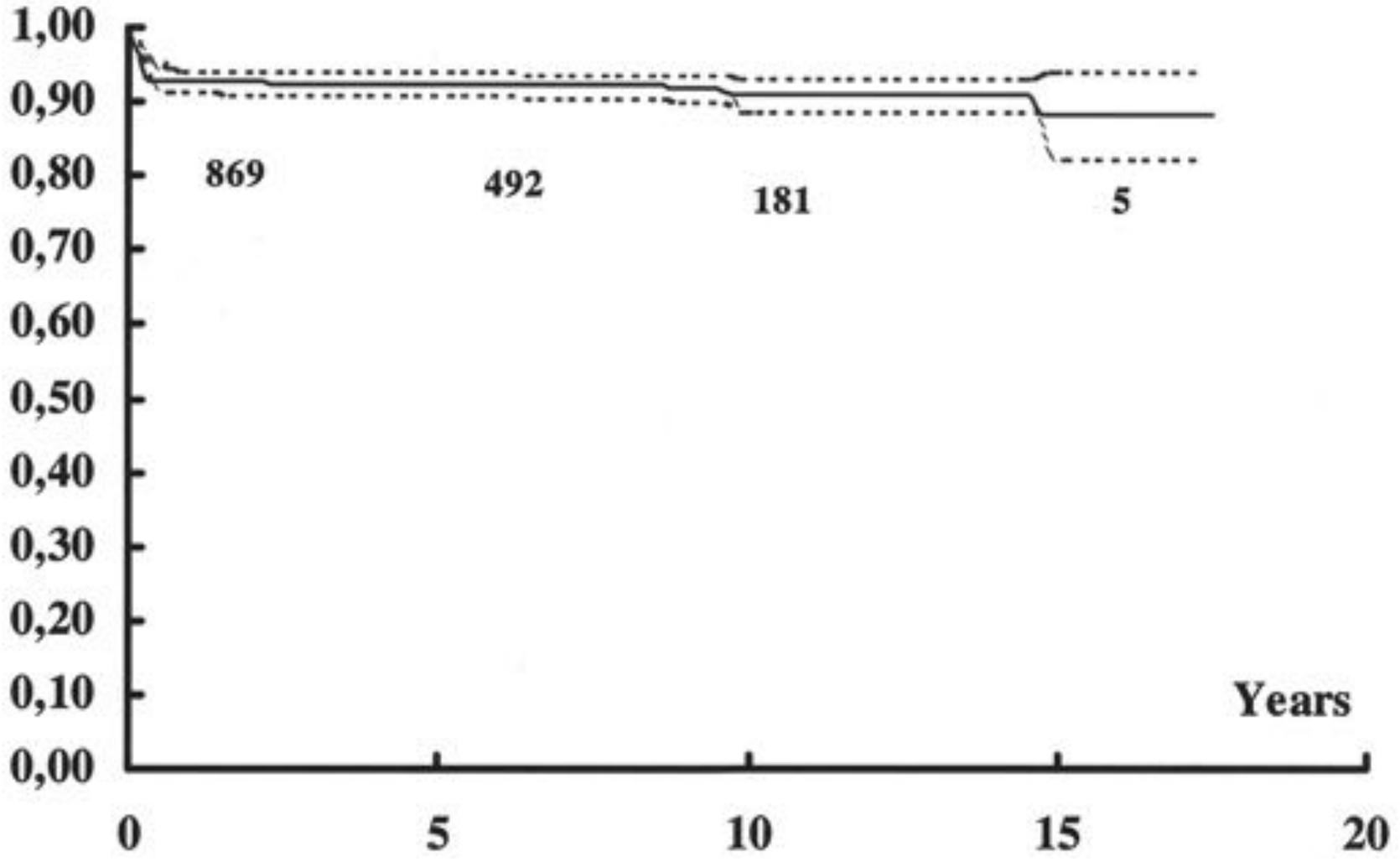


Coronary arteries after the arterial switch for TGA

Intervascular course fo left CA



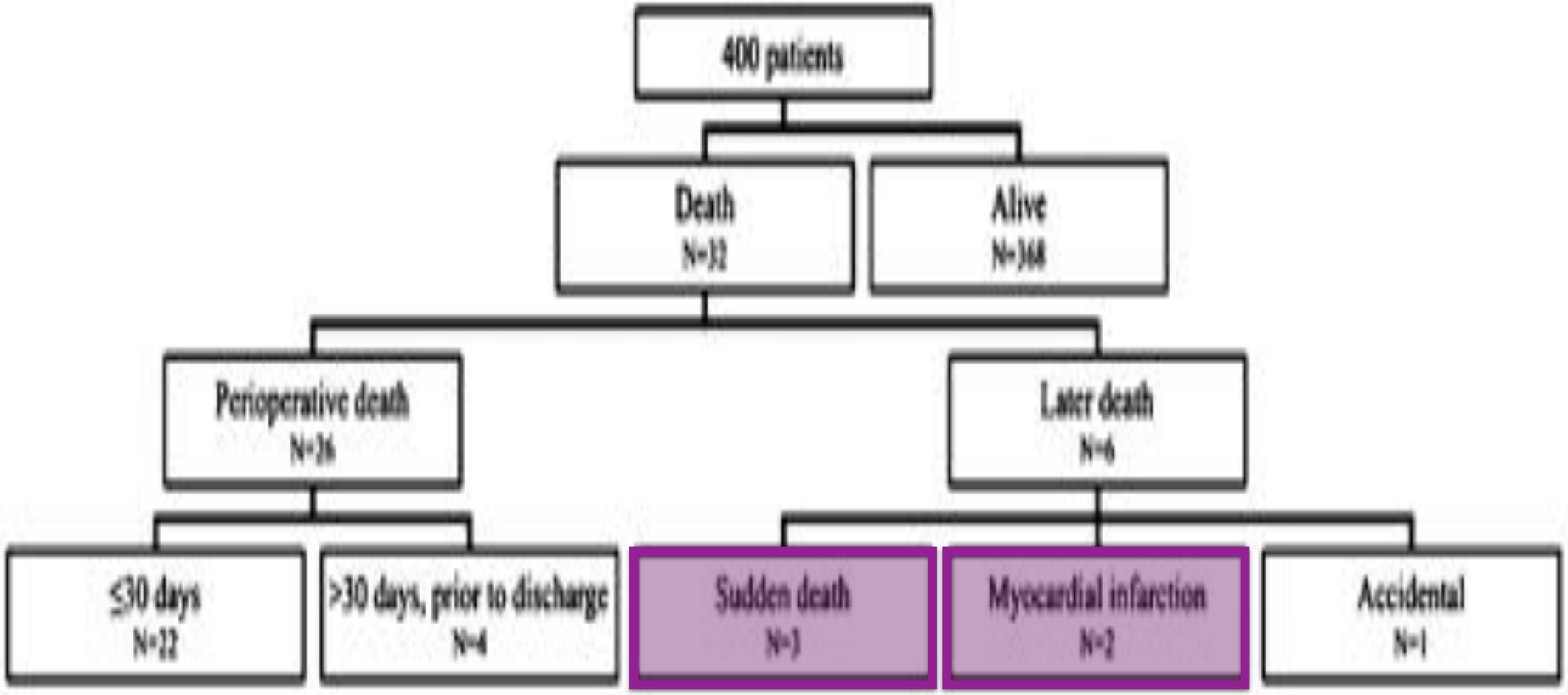
Actuarial survival free of coronary events for 1304 patients



Legendre A et al. Circulation. 2003;108:II-186-II-190

Postoperative sequelae following the arterial switch operation

Prevalence of coronary lesions or potential coronary events



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

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

Life-long management of the ASO population poses several challenges

The coronary arteries after transfer

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

What to do with abnormal coronary arteries in the absence of symptoms ?

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

Are there some « at risk » patients who would need systematic screening for myocardial ischemia ?

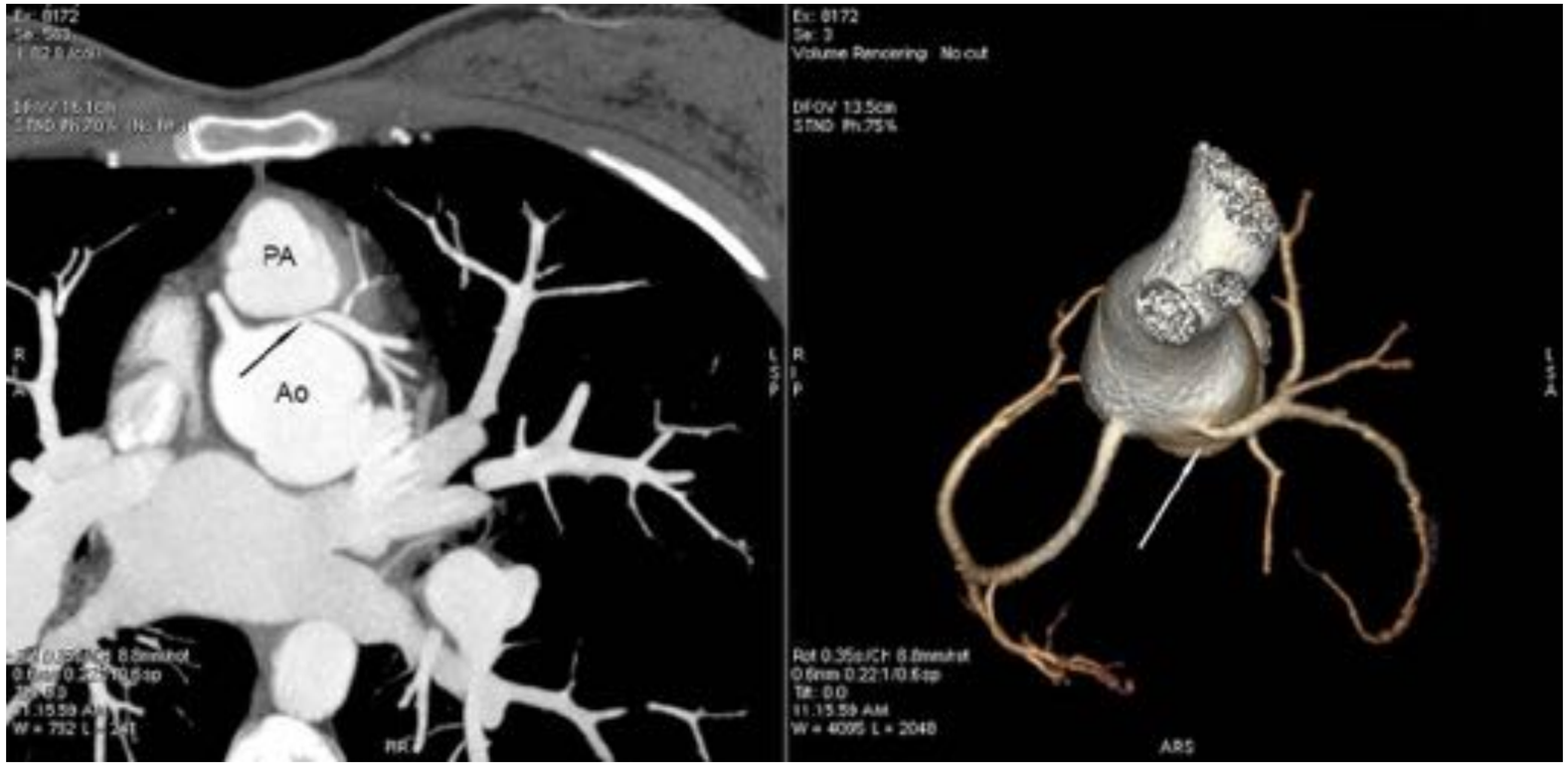
Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511

Parisian experience in systematic screening

1453 patients diagnosed with / or screened for coronary anomalies after ASO

78 coronary artery obstructions

Prevalence of asymptomatic coronary artery obstruction (> 30%) : 5.3%



Bonnet D et al. Heart 1996;76:274-9
Bonhoeffer P et al . J Am Coll Cardiol 1997;29:202-6
Szymczyk K et al. Pediatr Cardiol 2018;39:335-46

Parisian experience in systematic screening

1453 patients diagnosed with or screened for coronary anomalies after ASO

Circumstances of diagnosis of coronary artery obstruction

1) Potential coronary event : 29 (37%)

Clinical (chest pain, near-syncope or syncope, heart failure) : 6
ECG and/or echocardiographic signs of myocardial ischemia : 23

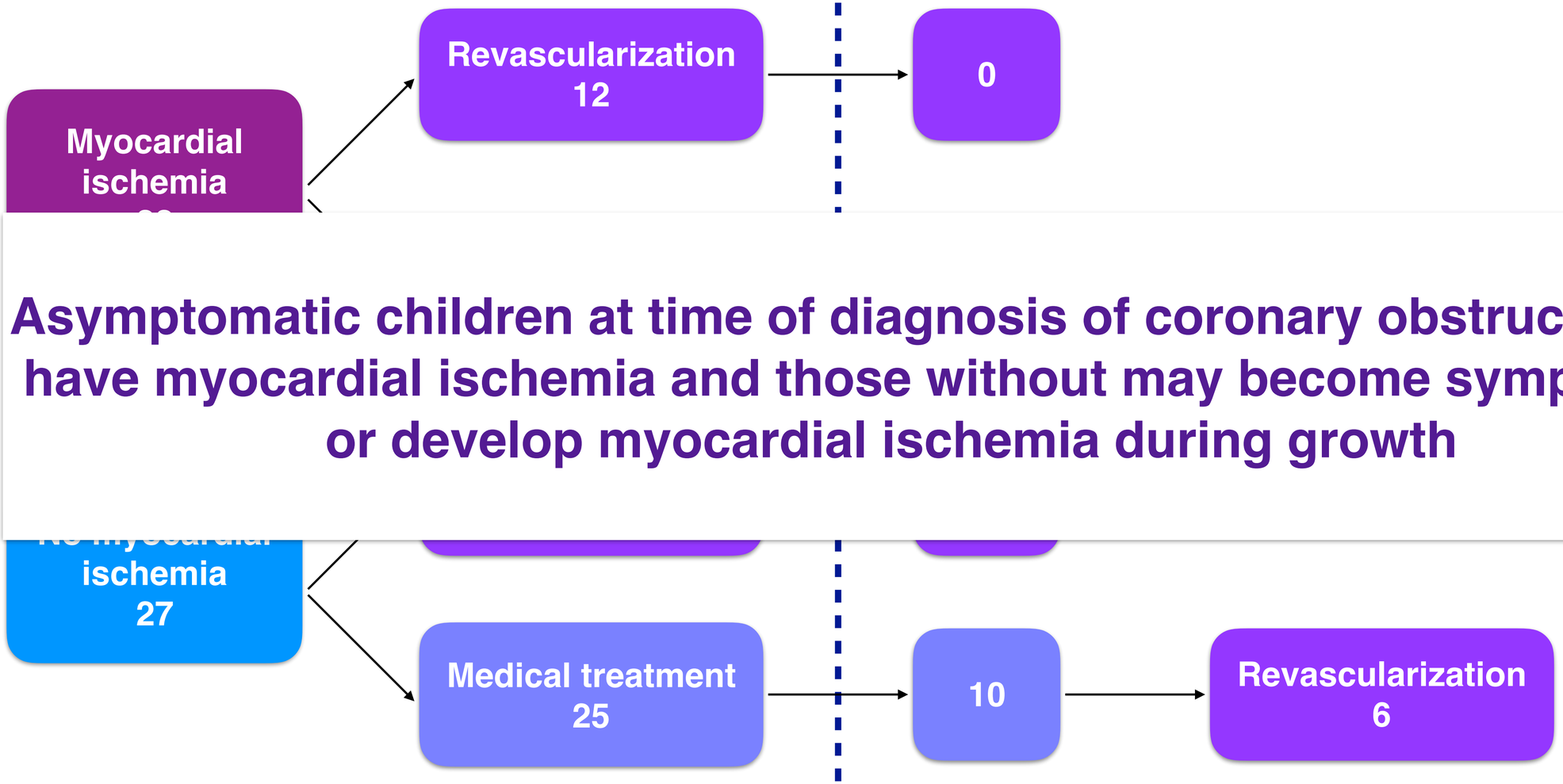
Age at potential coronary event < 6 months in 22 and > 6 years in 7

*Higher proportion of **Left main stem stenosis** compared to asymptomatic children and higher proportion of **severe stenosis > 50%** compared to mild stenosis and occlusion of the coronary artery*

2) Systematic screening in asymptomatic children : 49 (73%)

Parisian experience in systematic screening

49 asymptomatic patients with coronary obstruction



Asymptomatic children at time of diagnosis of coronary obstruction may have myocardial ischemia and those without may become symptomatic or develop myocardial ischemia during growth

PCE Median follow-up 6.8 years

Mechanisms of coronary artery complications after the ASO for TGA

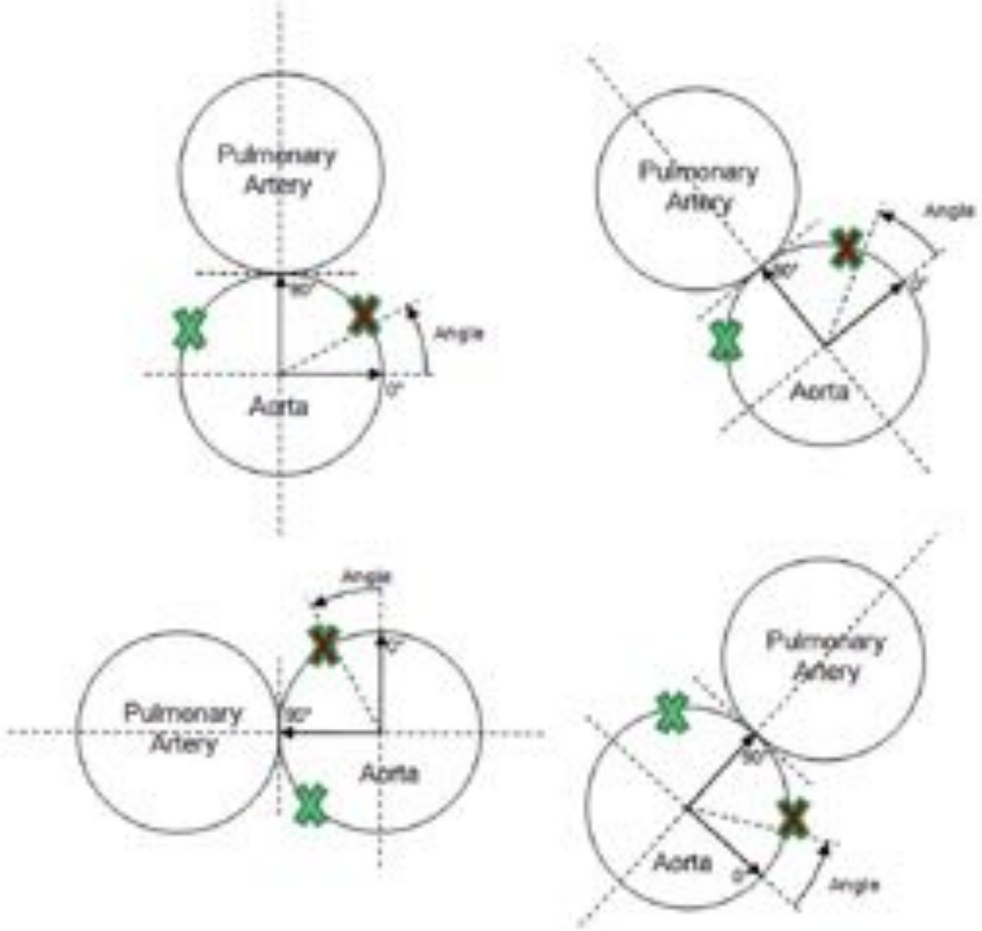


FIGURE 1. Determination of the coronary angles. The commonest positioning of the greats arteries and the coronaries after ASO are representing here. The coronary angles were determined as follows: (1) The aortopulmonary centerline that passes through the center of both the aorta and the pulmonary artery was drawn; (2) the perpendicular of the aortopulmonary centerline that passes through the center of the aorta is the reference line (0°); (3) the angle between the coronary ostia (*red cross*, left ostium; *green cross*, right ostium) and the reference line correspond to the coronary angle.

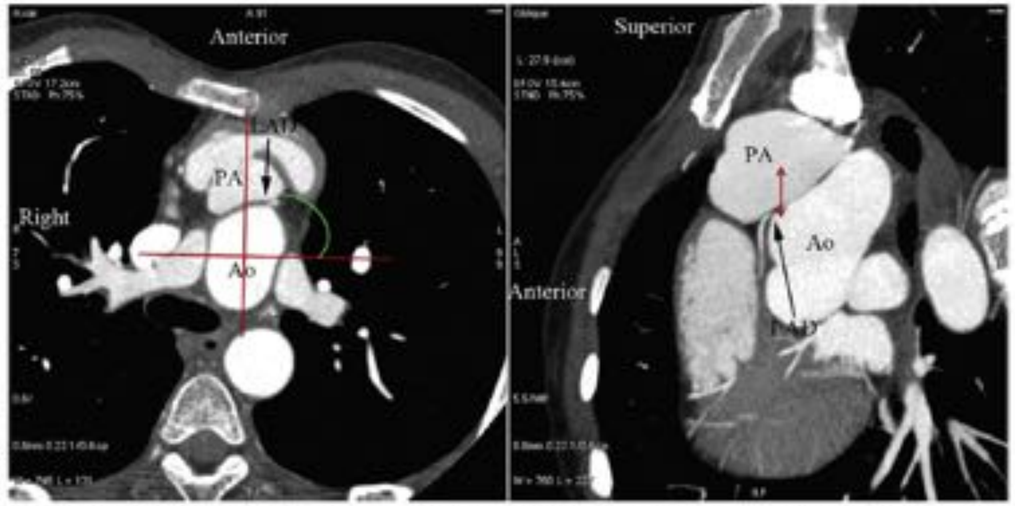


FIGURE 3. Tight stenosis of the LAD in the Yacoub type D. The reimplantation of the LAD (*black arrow* on the *left panel*) is to anterior as indicated by the left coronary angle almost equal to 90° (*green curve* on the *left panel*), and also too high above the left coronary sinus, as identified by the short coronary–pulmonary bifurcation distance (*red arrow* on the *right panel*) equal to 5 mm. *PA*, Pulmonary artery; *Ao*, aorta; *LAD*, left anterior descending artery.

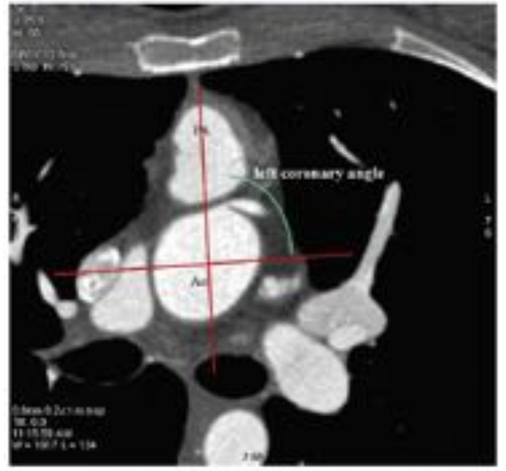


FIGURE 2. Significant stenosis of the ostium of the left coronary artery by anterior compression by the pulmonary artery. The reimplantation is too anterior, as confirmed by the left coronary angle (*green curve*) almost equal to 90°, corresponding to a reimplantation at 12 o' clock. *PA*, Pulmonary artery; *Ao*, aorta.

Ou P et al. JTCVS 2013;145:1263-9

Mechanisms of coronary artery complications after the ASO for TGA

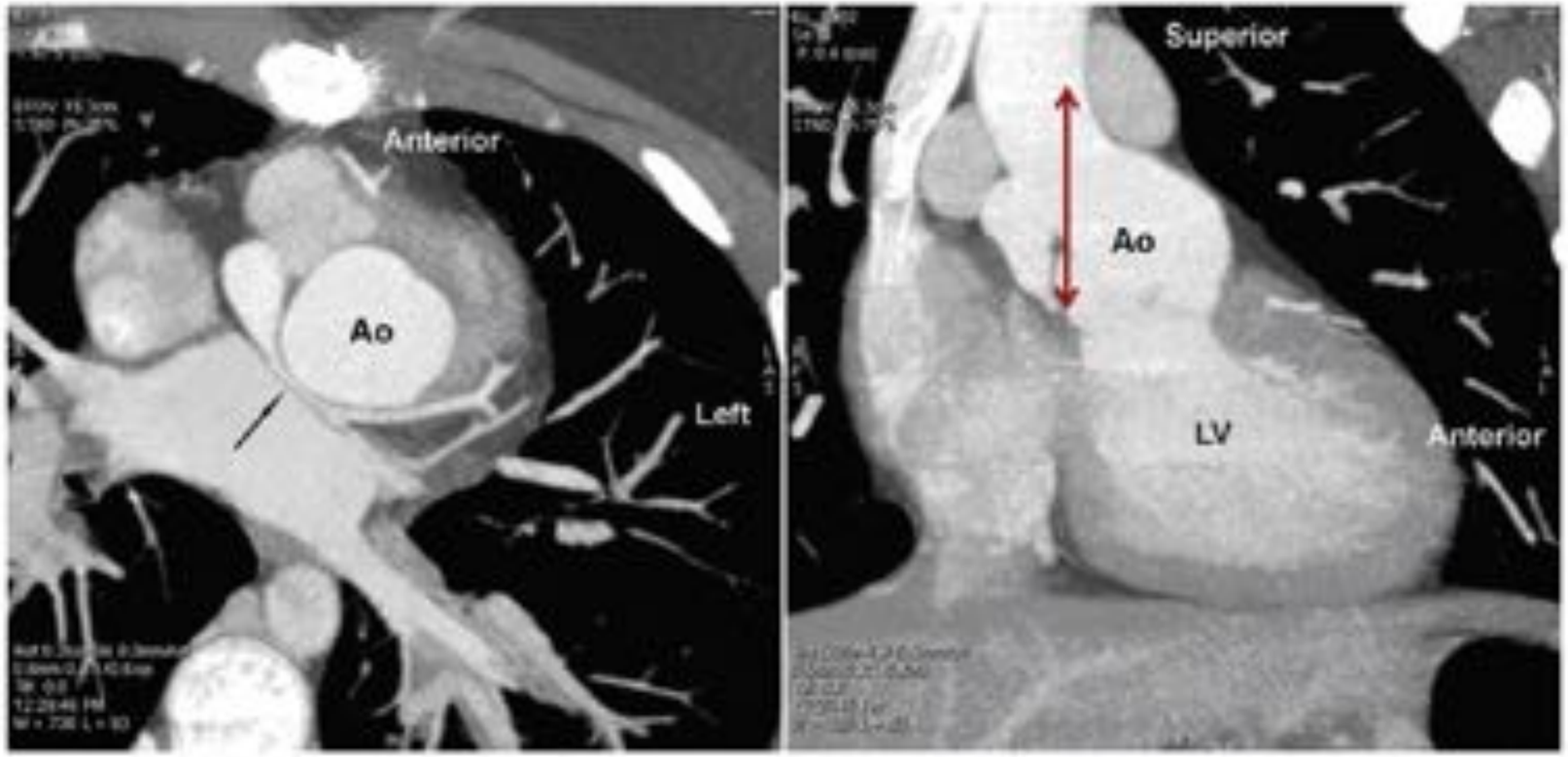


FIGURE 4. Mechanism of lesion of the circumflex artery in Yacoub type D. The reimplantation of the right ostium was too far under the right sinus, as evidenced by the long coronary–pulmonary artery distance (*red arrow* on the *right panel*). Thus, the course of the circumflex toward the left atrioventricular groove is increased, causing a stretching of the coronary artery (*black arrow* on the *left panel*). *Ao*, Aorta; *LV*, left ventricle.

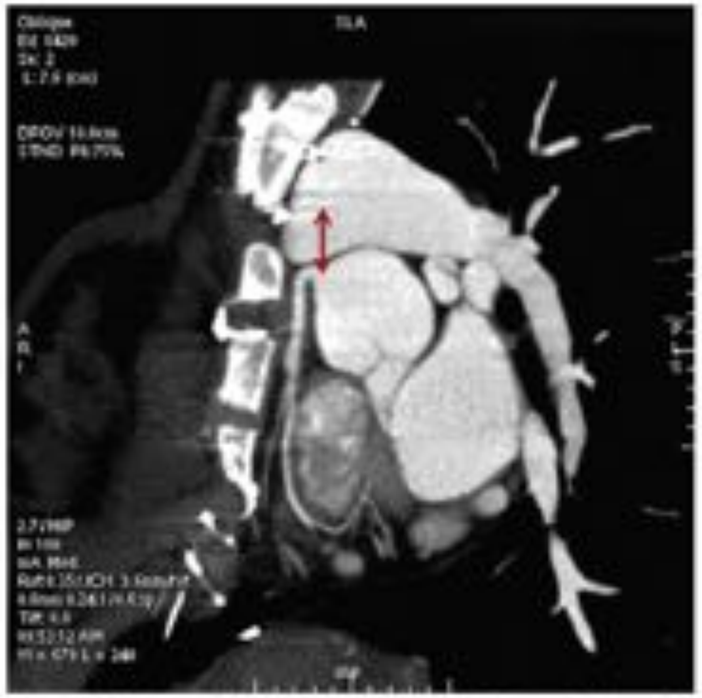


FIGURE 5. Lesion of the right coronary artery. The reimplantation of the right sinus is too high above the right sinus, as evidenced by the short coronary–pulmonary artery bifurcation distance (*red arrow*). The right ostium was then compressed by the pulmonary artery. This patient had a long "ulasty" of the ostium and proximal segment of the right coronary artery using saphenous vein. See the difference in caliber between the saphenous vein in the proximal segment and the native distal segment, which is thinner.

Ou P et al. JTCVS 2013;145:1263-9

Parisian experience in systematic RE-screening

Adolescents with previous normal coronary arteries at first screening

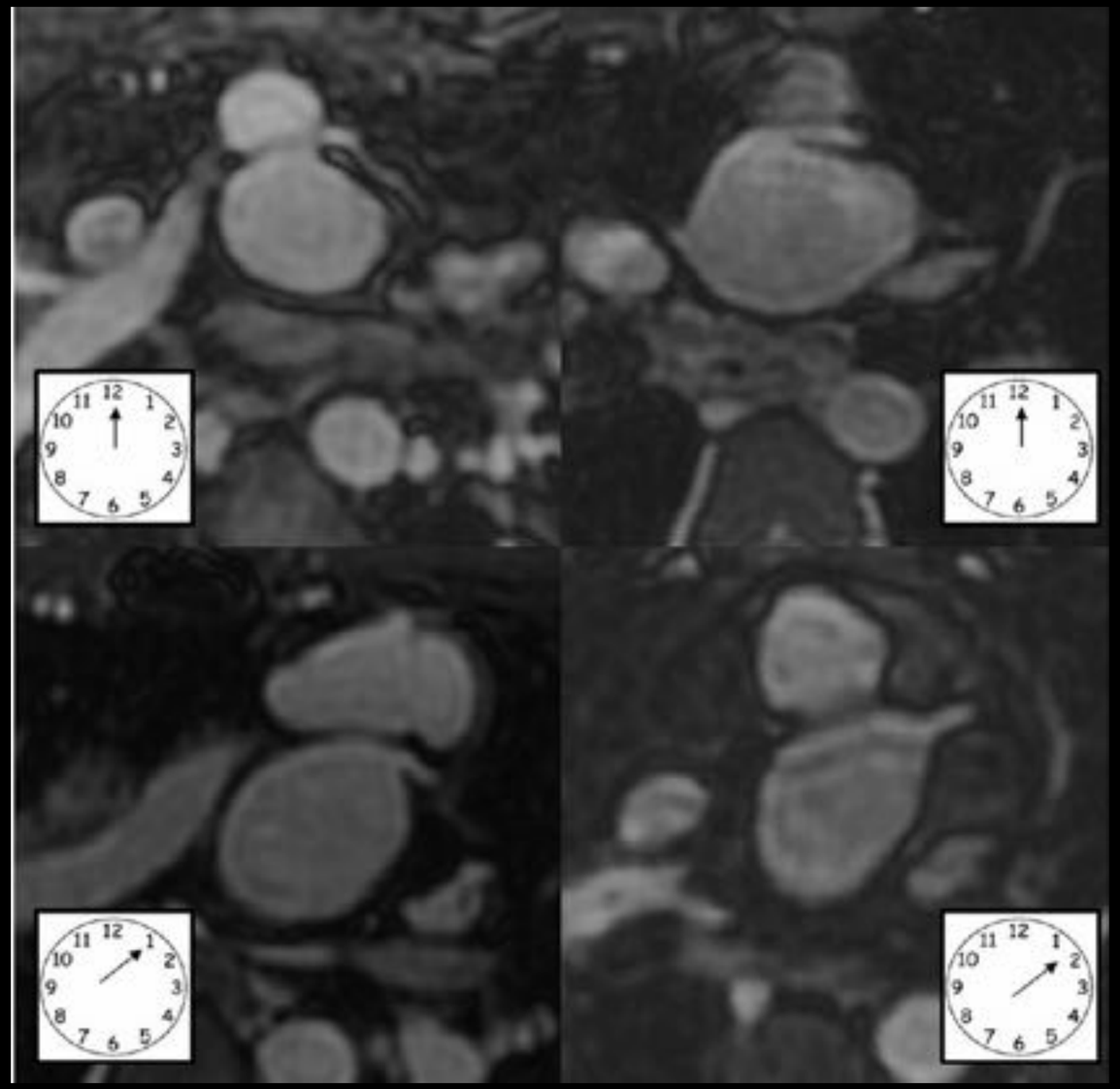
107 patients who had a normal coronary artery anatomy at first systematic screening (mean age 5.0 years, range 4-7 years).

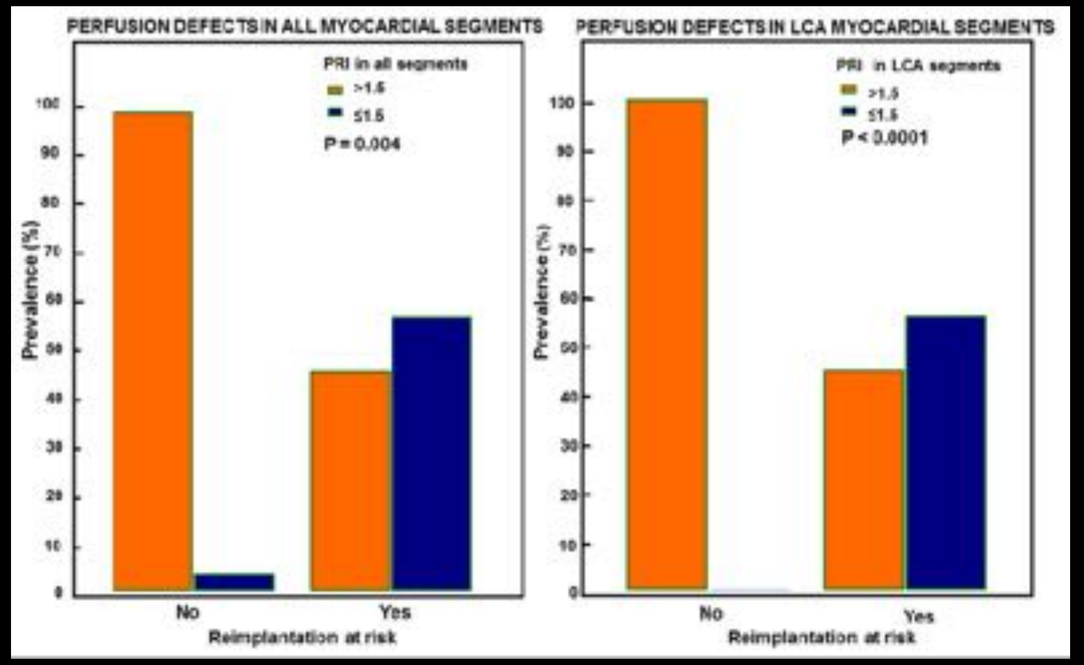
All patients had annual follow-up. All were all NYHA functional class I. None had symptoms of myocardial ischemia. None had ECG anomalies suggestive of myocardial ischemia. None of them had LV dysfunction (global or regional) or mitral regurgitation.

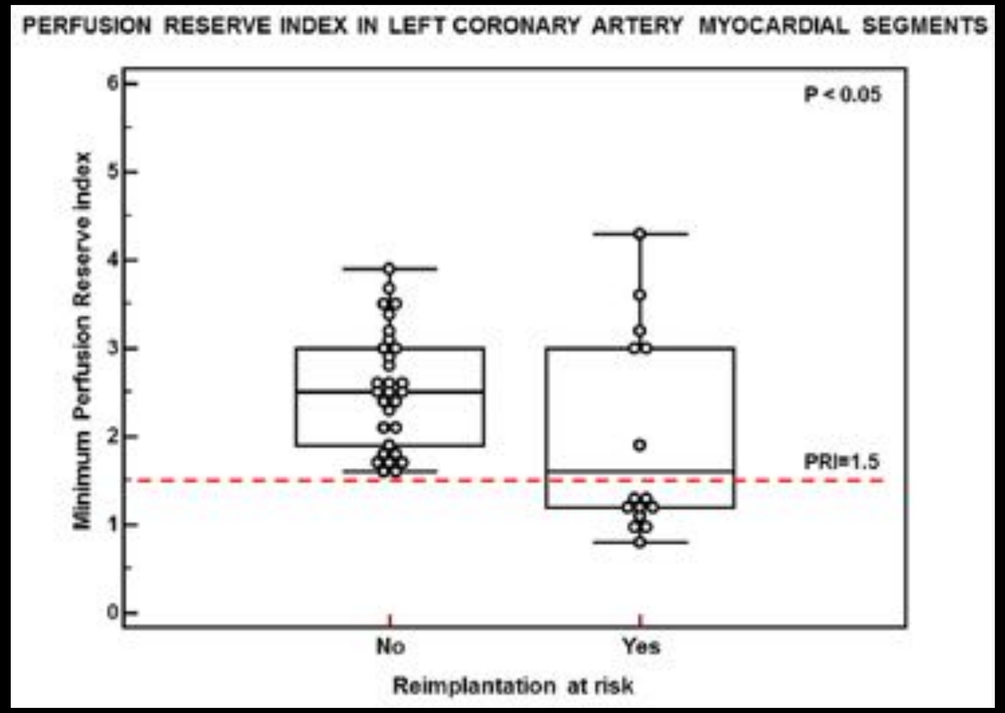
Only one/107 new obstruction of RCA (stenting)

No change in the position of the coronary ostium during growth

Raimondi F et al. JACC Imaging 2017

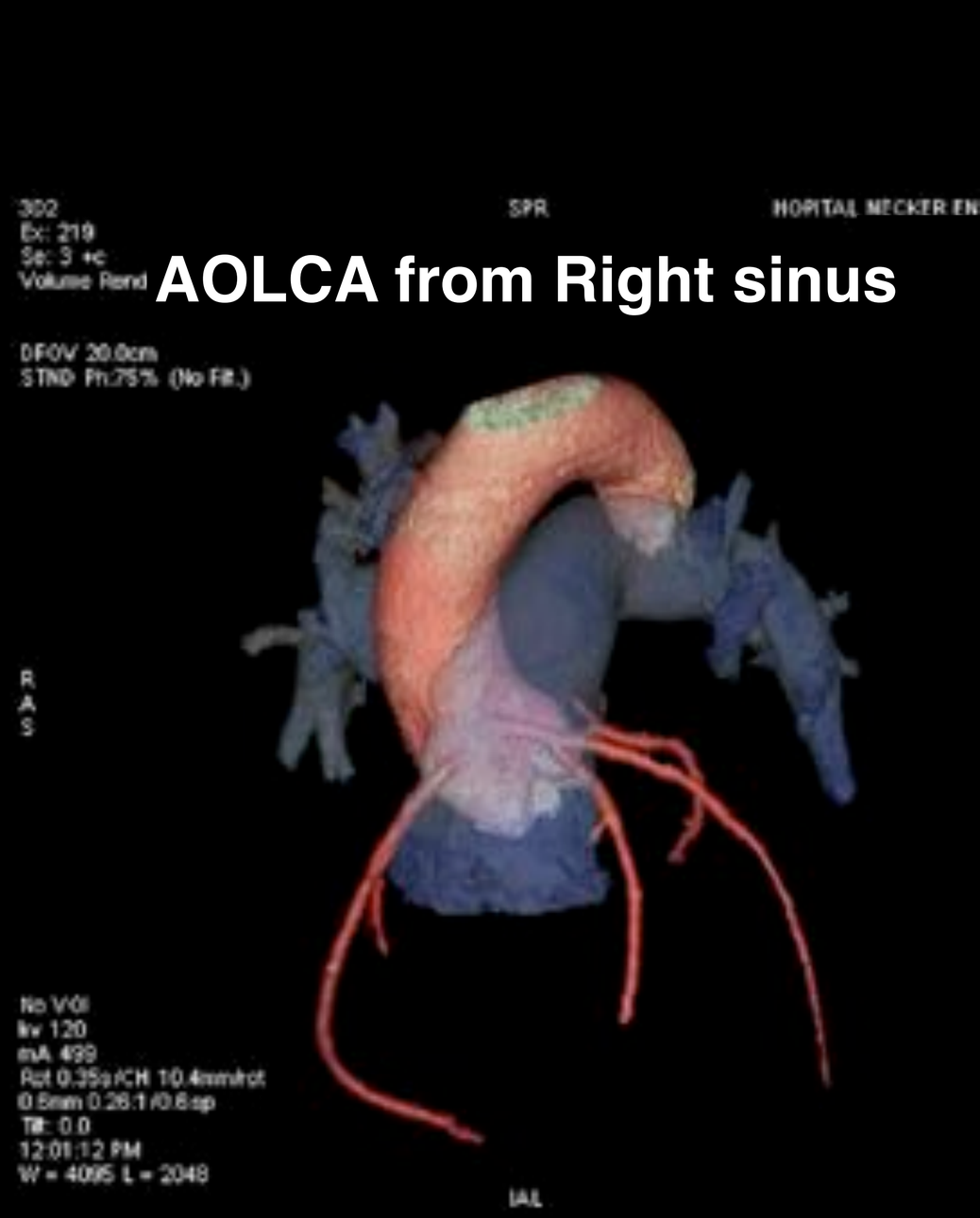


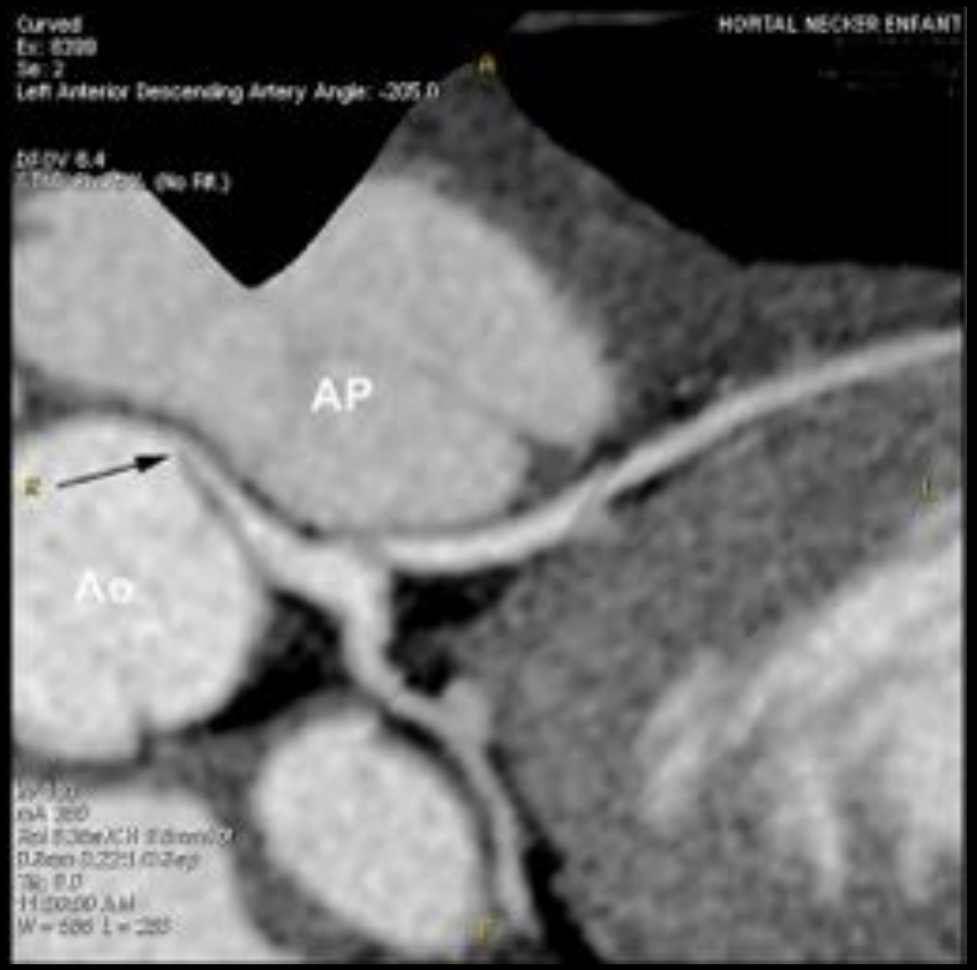


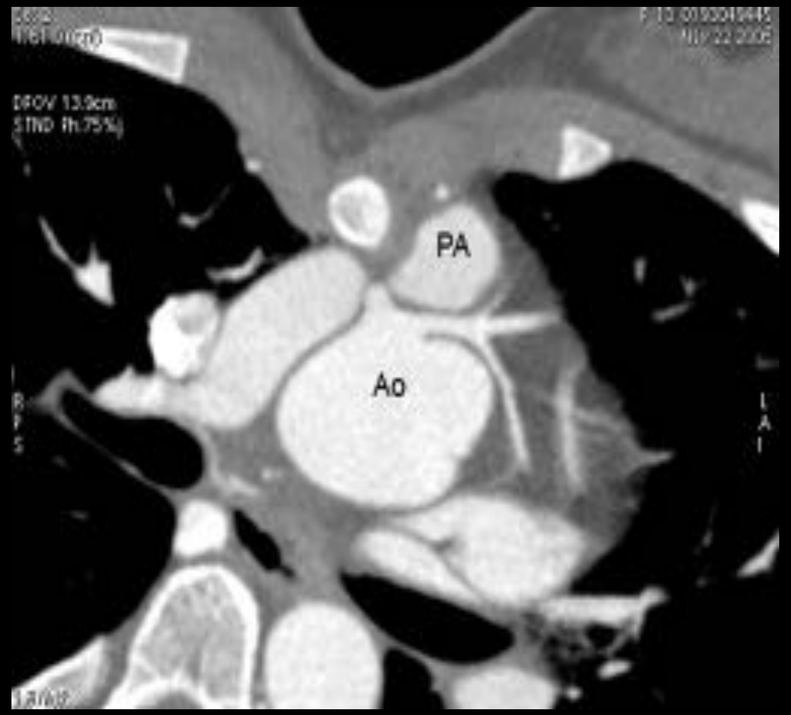


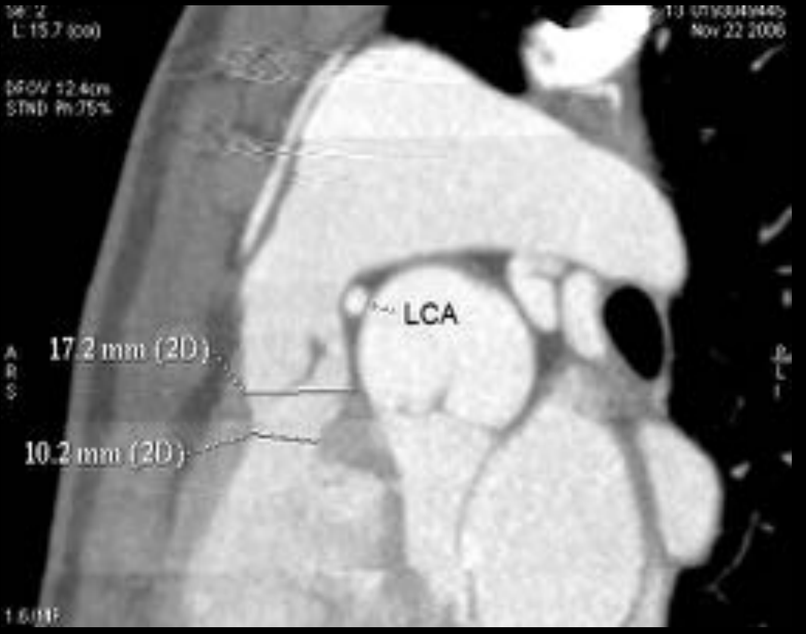
Raimondi F et al. JACC Imaging 2017


3D
Ex: 8420
Se: 2
Volume Rendering No cut
DFOV 18.9cm
STND Ph:75%
SPR
HOPITAL NECKER ENFANT
13 0193049445
POST ASO
A
R
S
No VOI
kv 100
mA Mod.
Rot 0.35s/CH 9.6mm/rot
0.6mm 0.24:1/0.6sp
Tilt: 0.0
09:53:12 AM
W = 4095 L = 2048
IAL

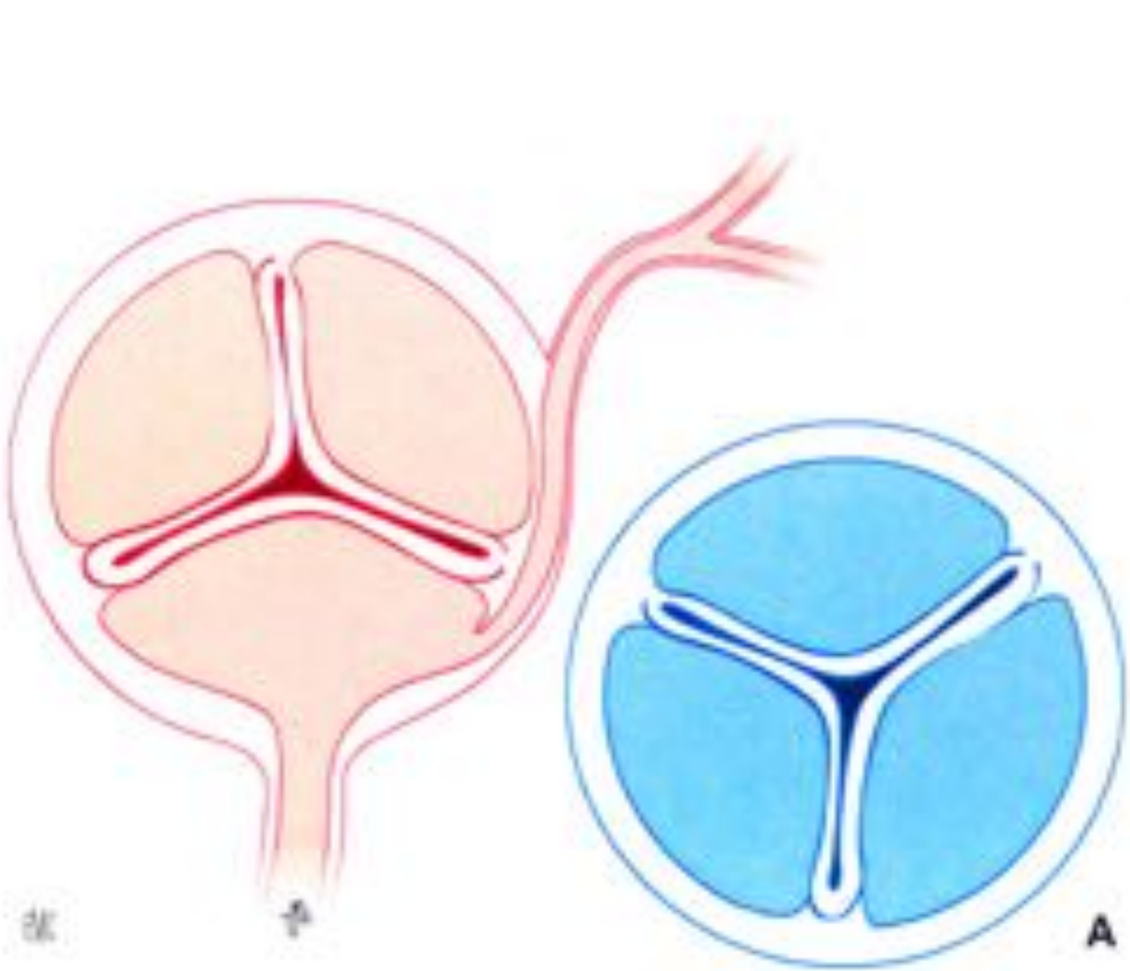

302
Ex: 219
Se: 3 +c
Volume Rend
SPR
HOPITAL NECKER EN
AOLCA from Right sinus
DFOV 20.0cm
STND Ph:75% (No Fit.)
R
A
S
No VOI
kv 120
mA 499
Rot 0.35s/CH 10.4mm/rot
0.6mm 0.26:1/0.6sp
Tilt: 0.0
12:01:12 PM
W = 4095 L = 2048
IAL


Curved
Ex: 6399
Se: 2
Left Anterior Descending Artery Angle: -205.0
HORTAL NECKER ENFANT
AP
Ao

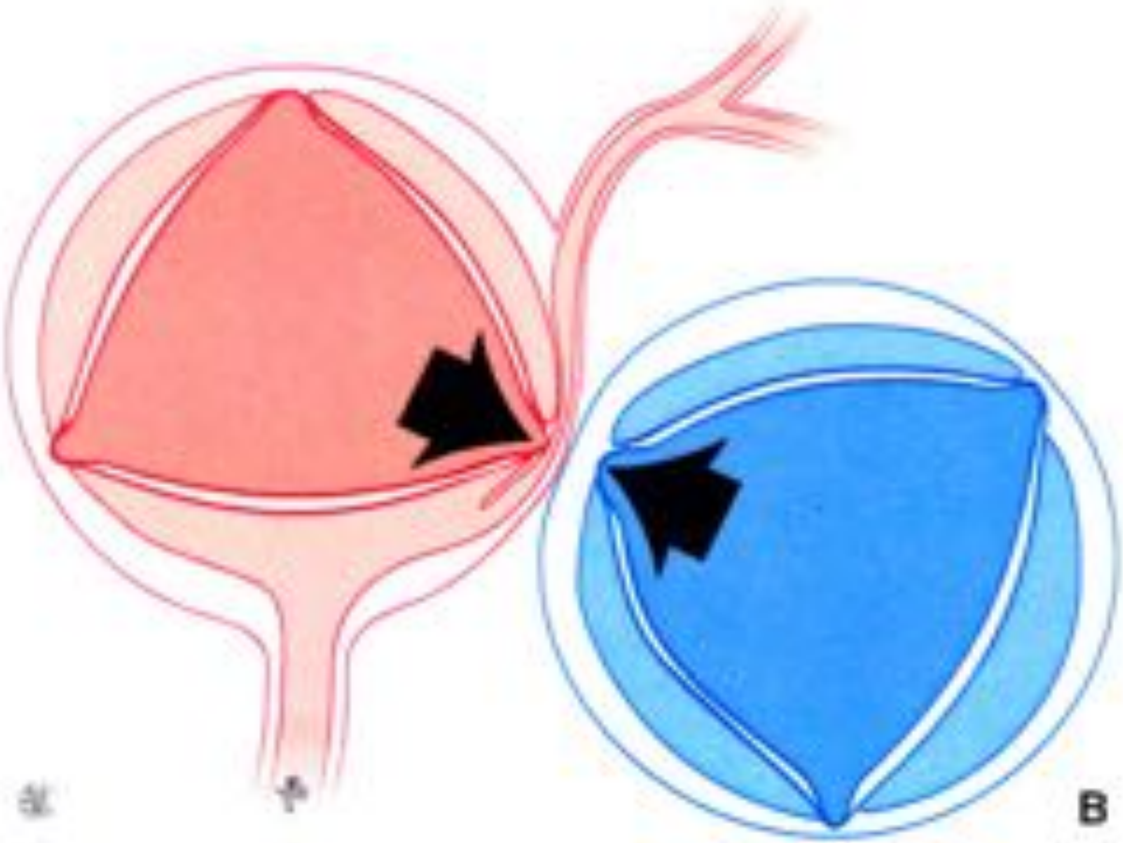

DFOV 13.9cm
STND fh75%
PA
Ao


L 15.7 (cor)
DFOV 12.4cm
STND fh75%
LCA
17.2 mm (2D)
10.2 mm (2D)

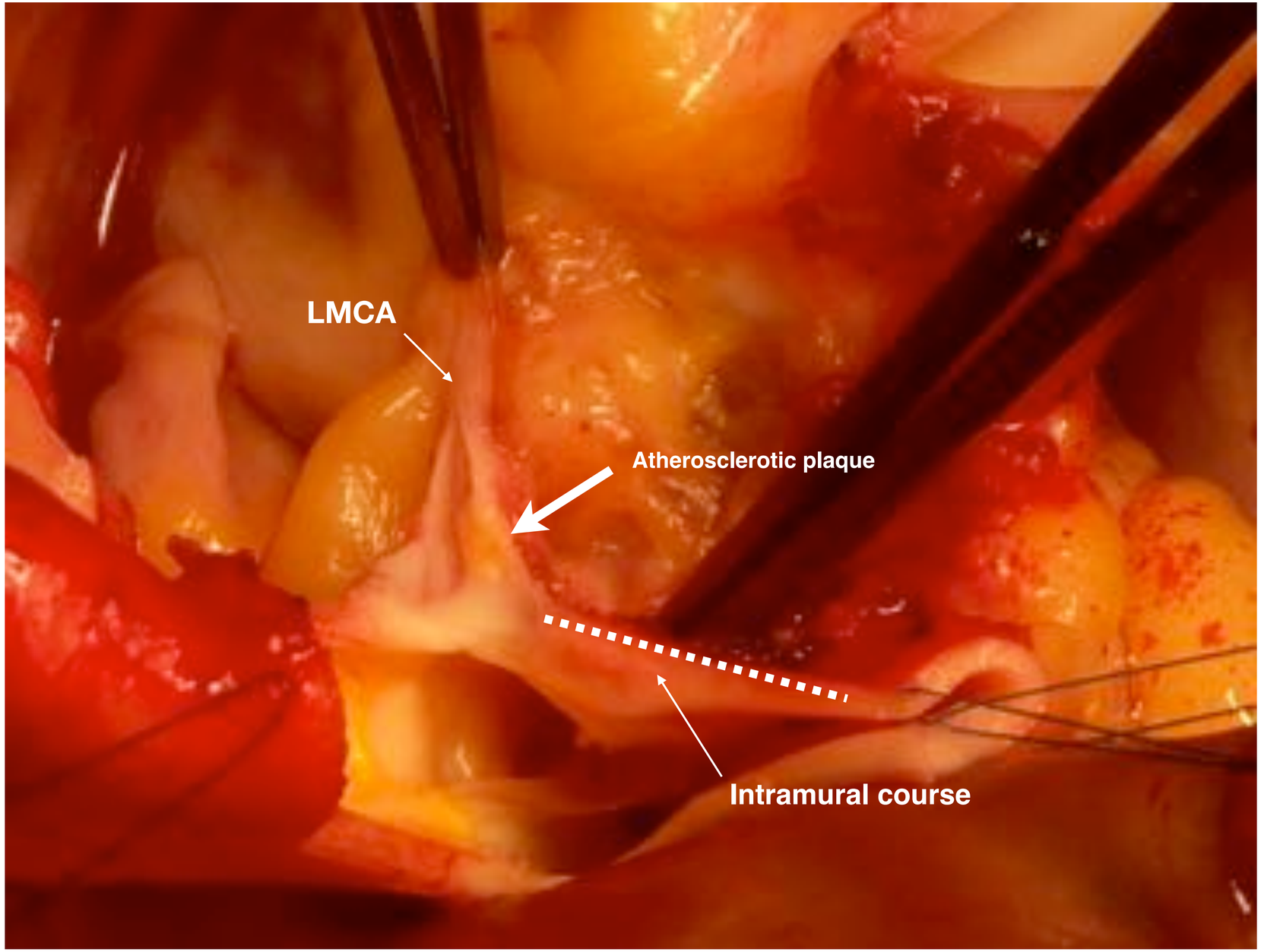
What will happen with neo-aortic root dilatation ?

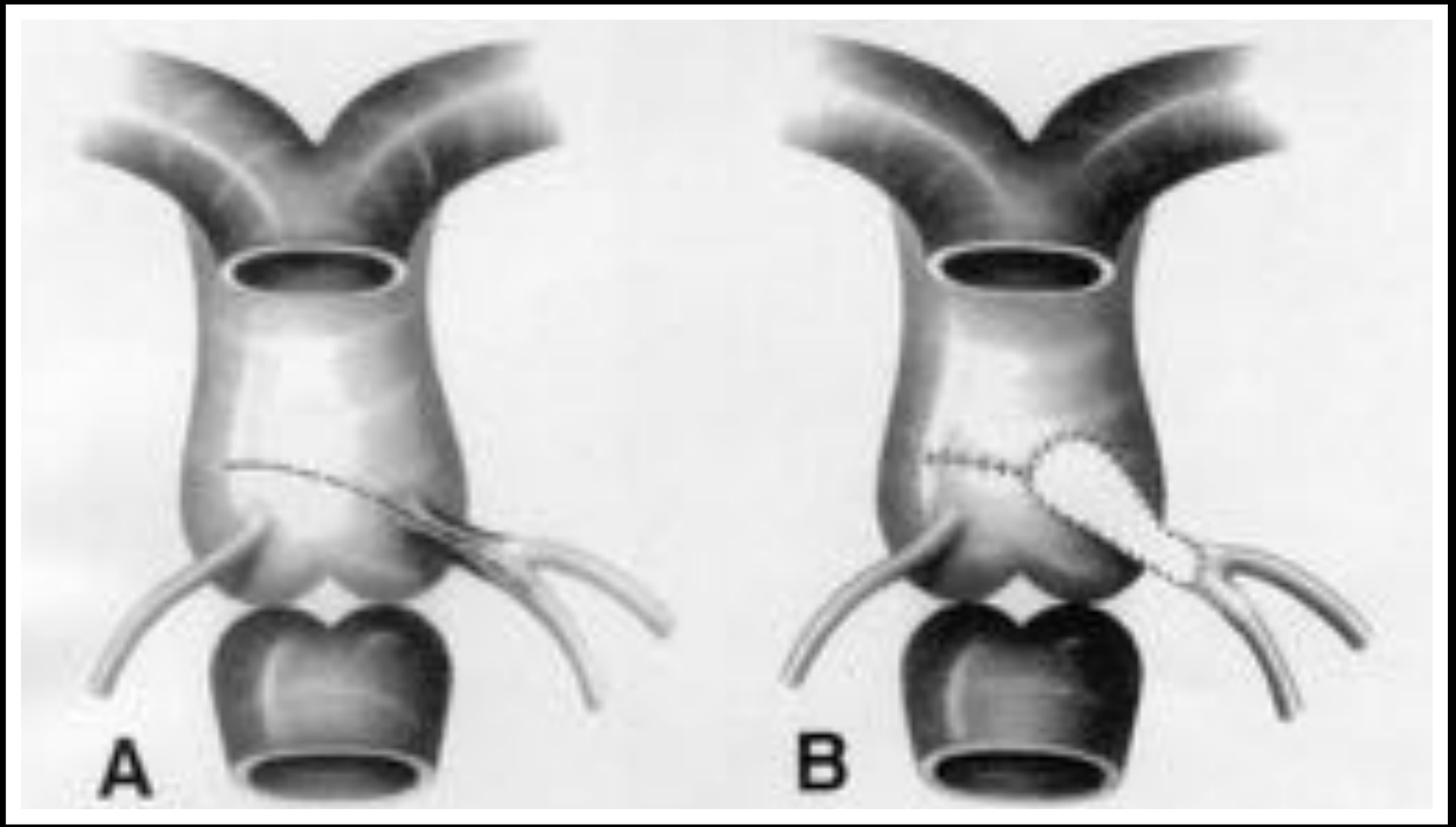


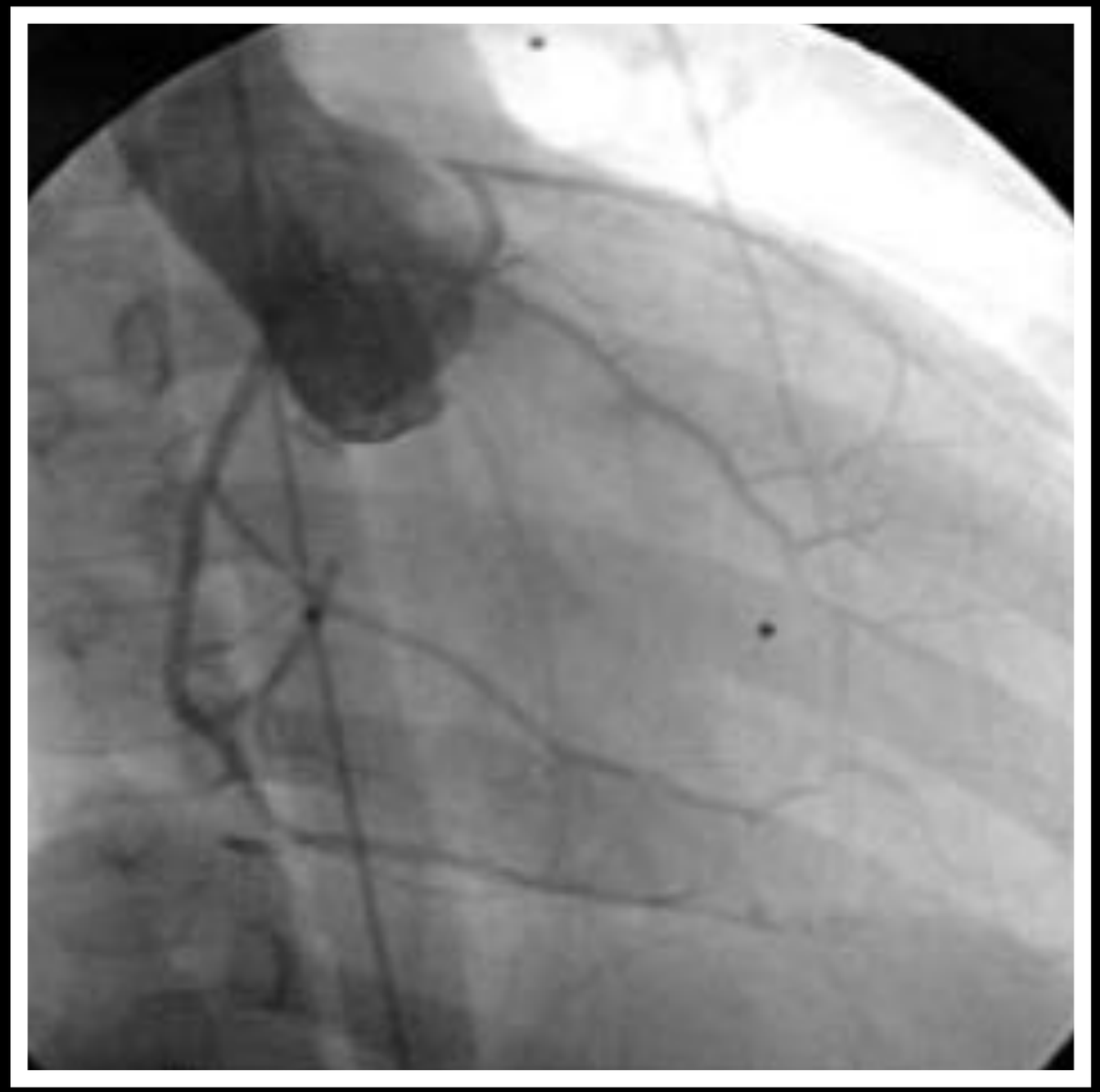
A

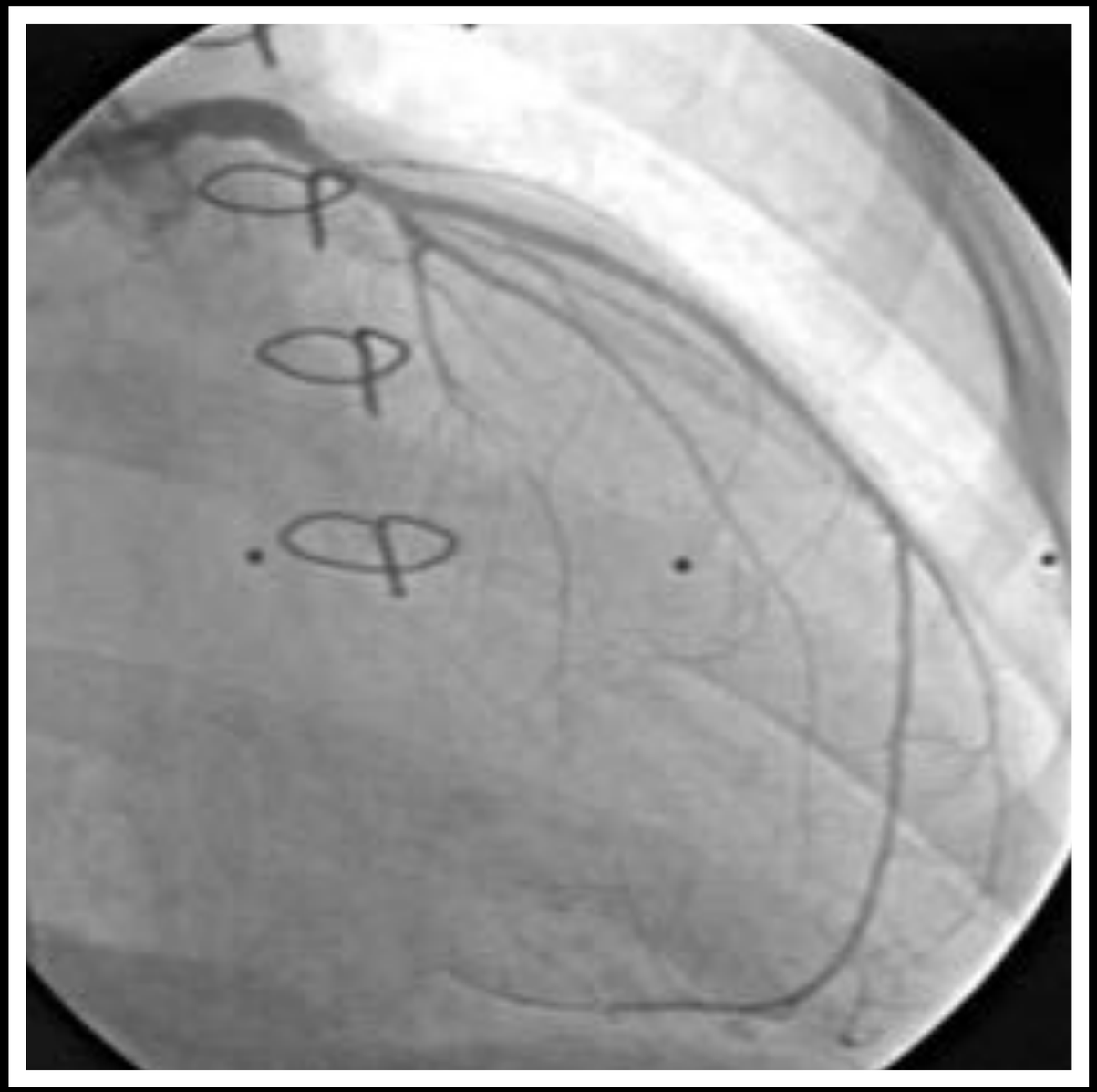


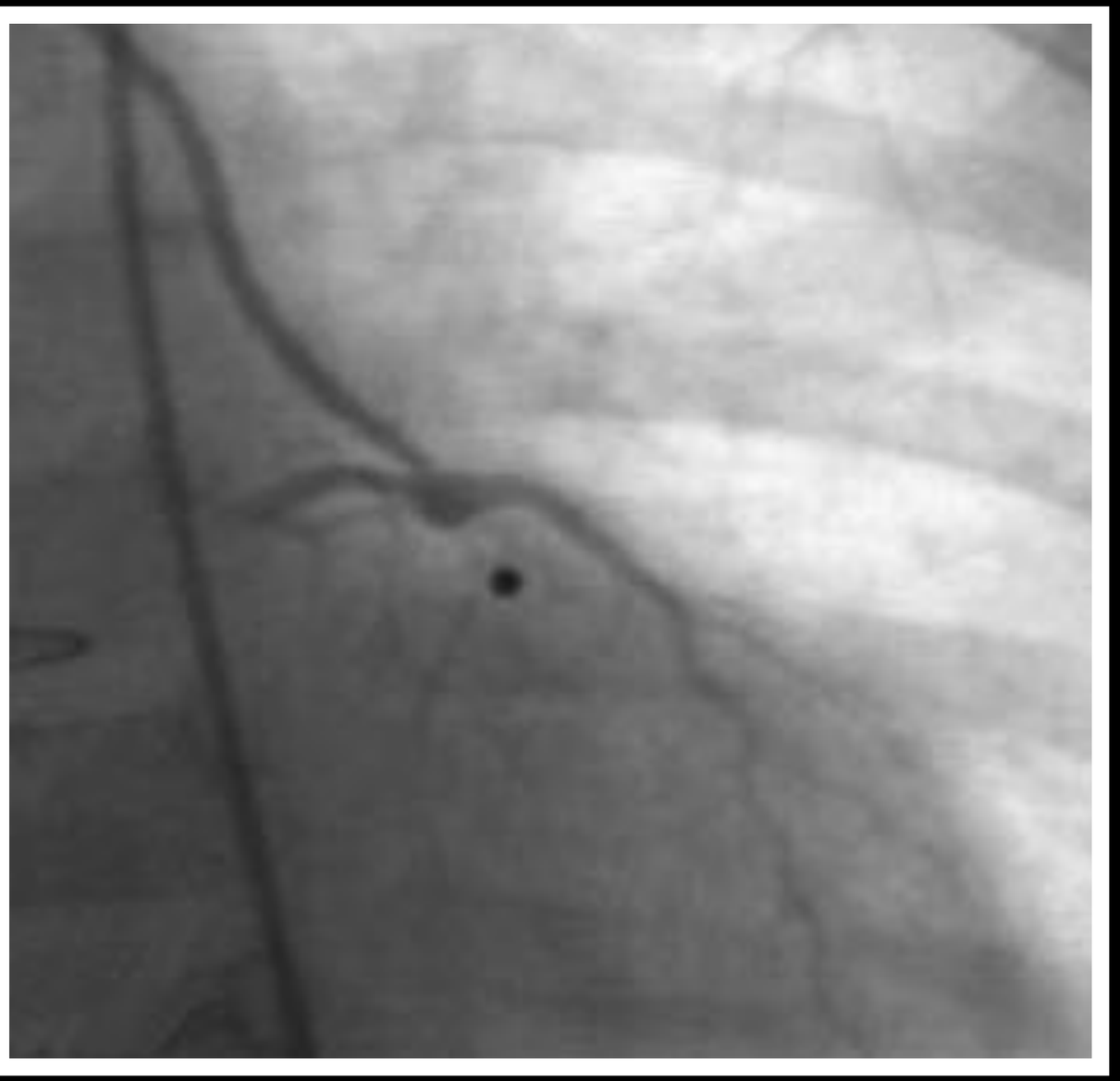
B

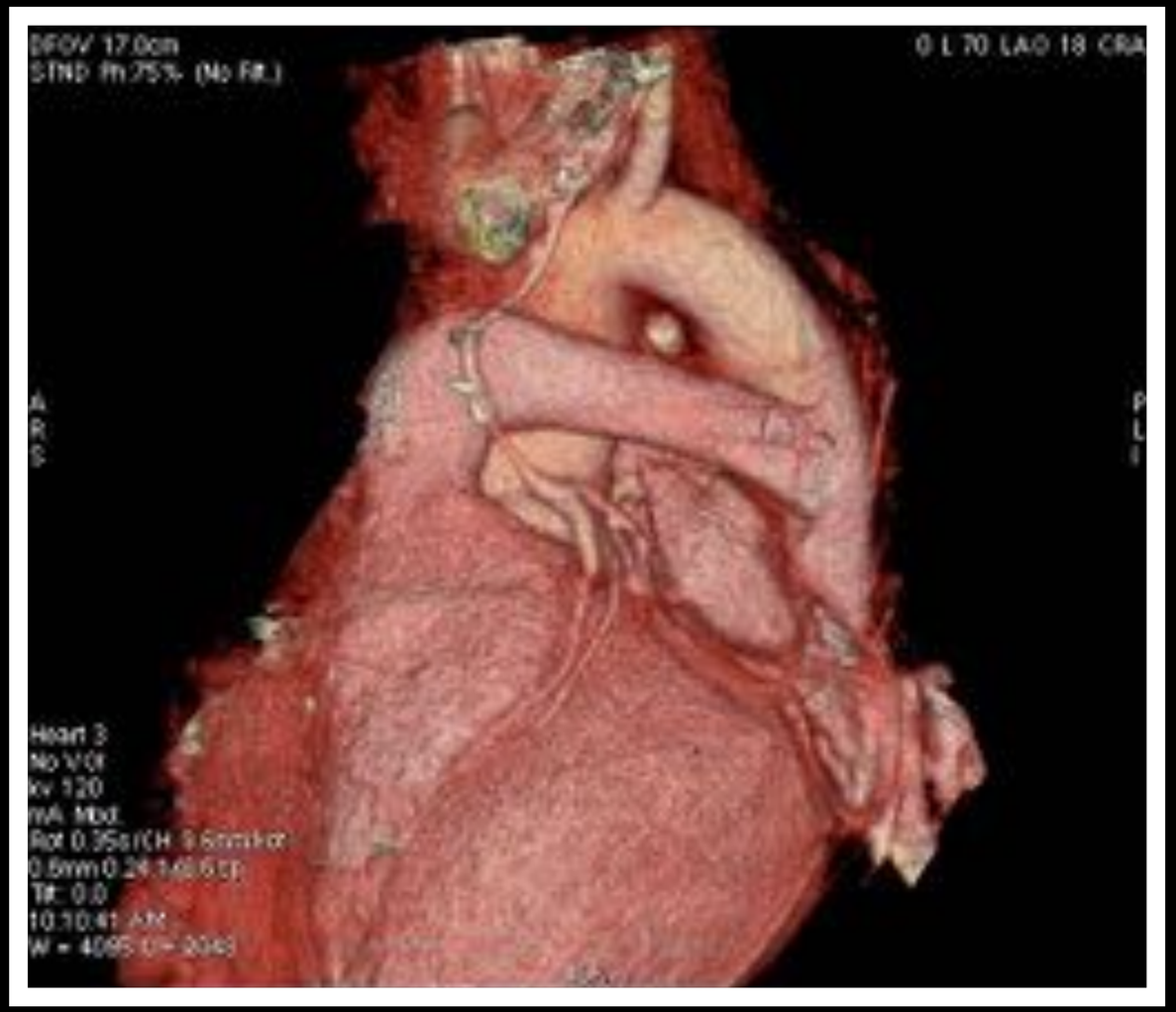

LMCA
Atherosclerotic plaque
Intramural course


A
B









Pontage mammaire interne-IVA post-switch

Conclusion Coronary arteries after ASO for TGA

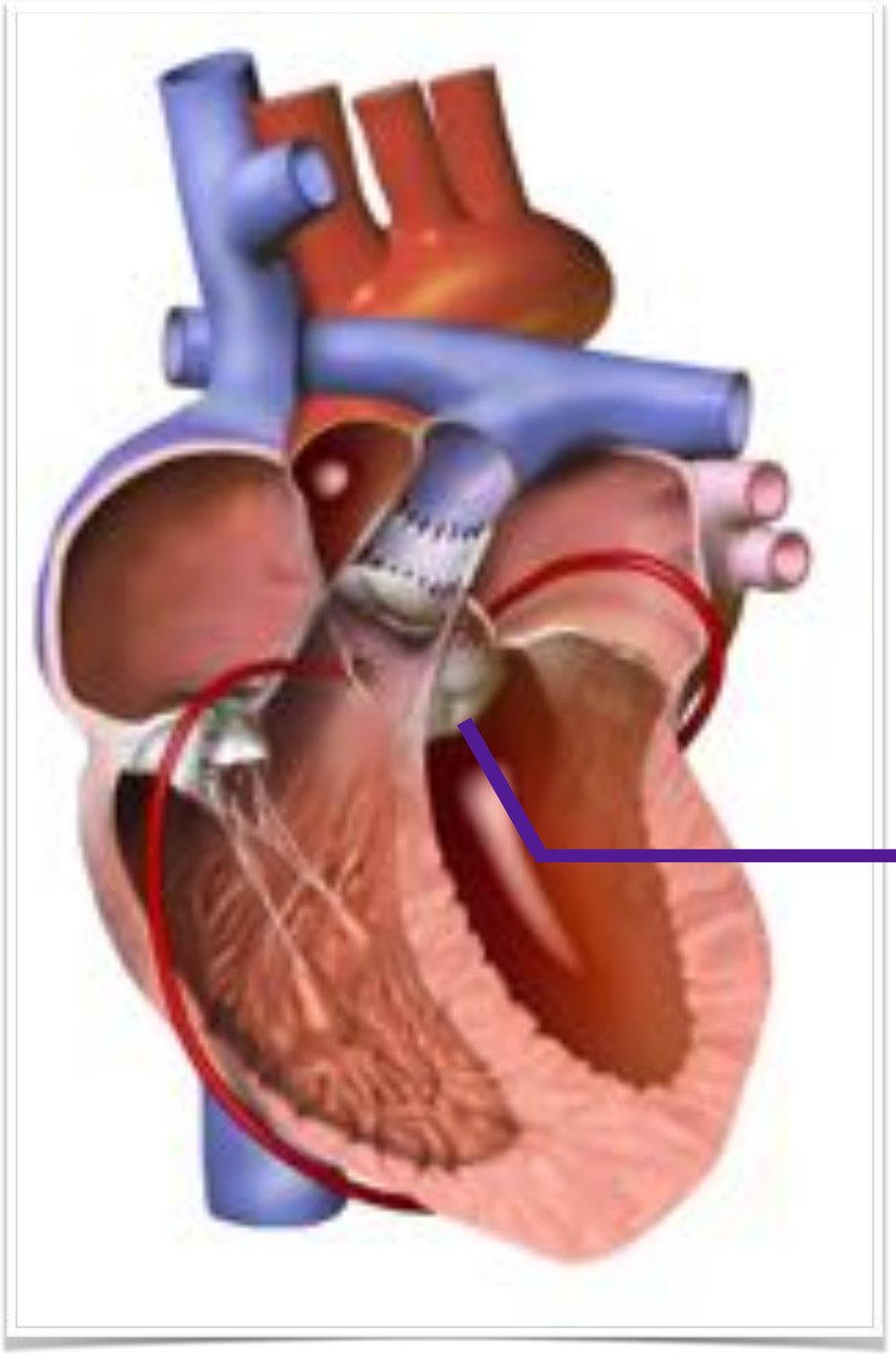
Systematic screening in ALL ASO patients is probably not efficient.

Coronary events are rare but they do exist.

The risk factors might not be the initial coronary anatomy but the acquired coronary anatomy.

Patients with close relationship between great vessels and initial coronary course might be at risk for myocardial perfusion anomalies and potentially for late coronary events.

Should we image all patients to identify this « at risk » population ?



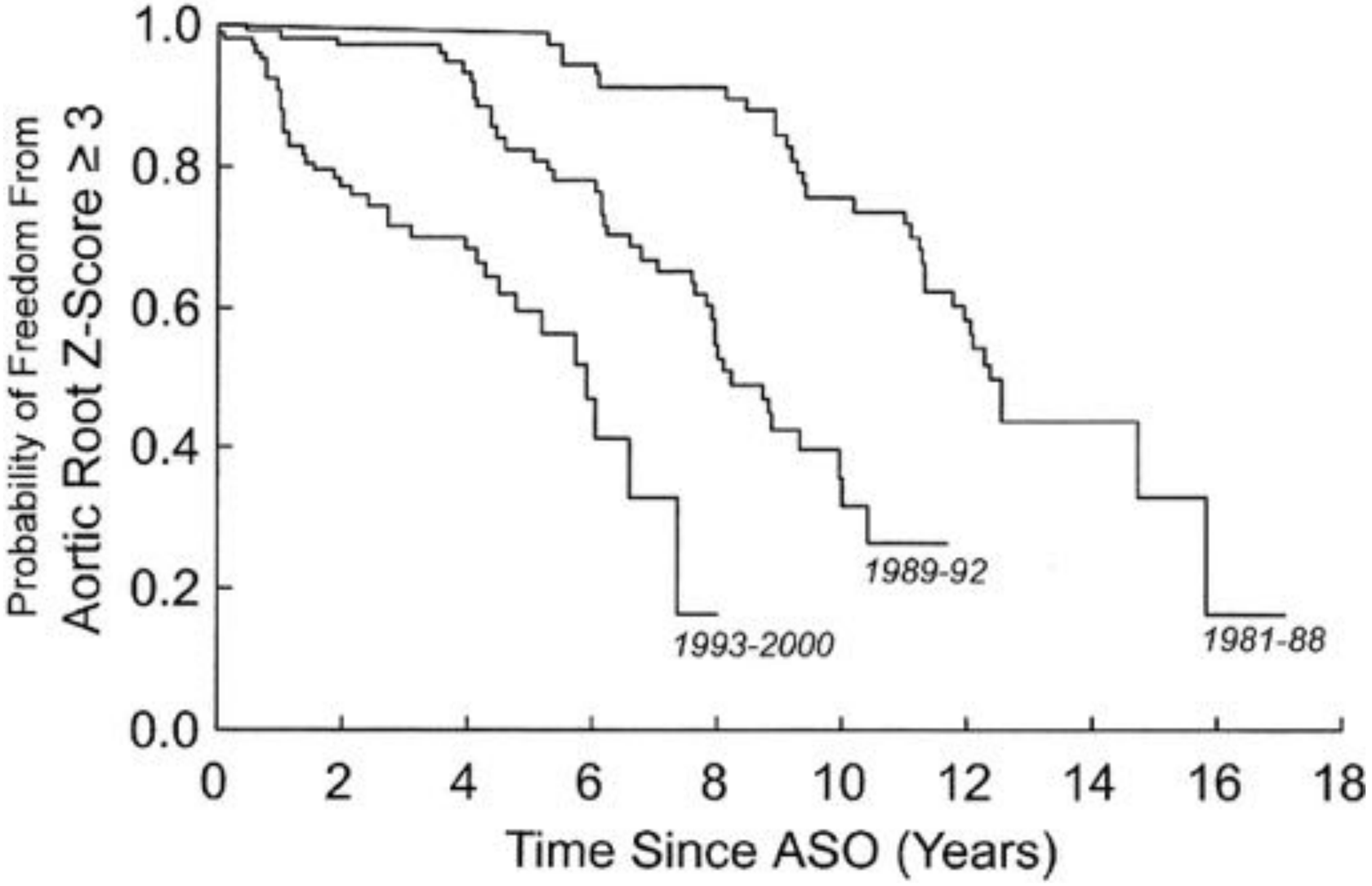
Aortic valve regurgitation

Aortic valve repair for AR after ASO ?

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

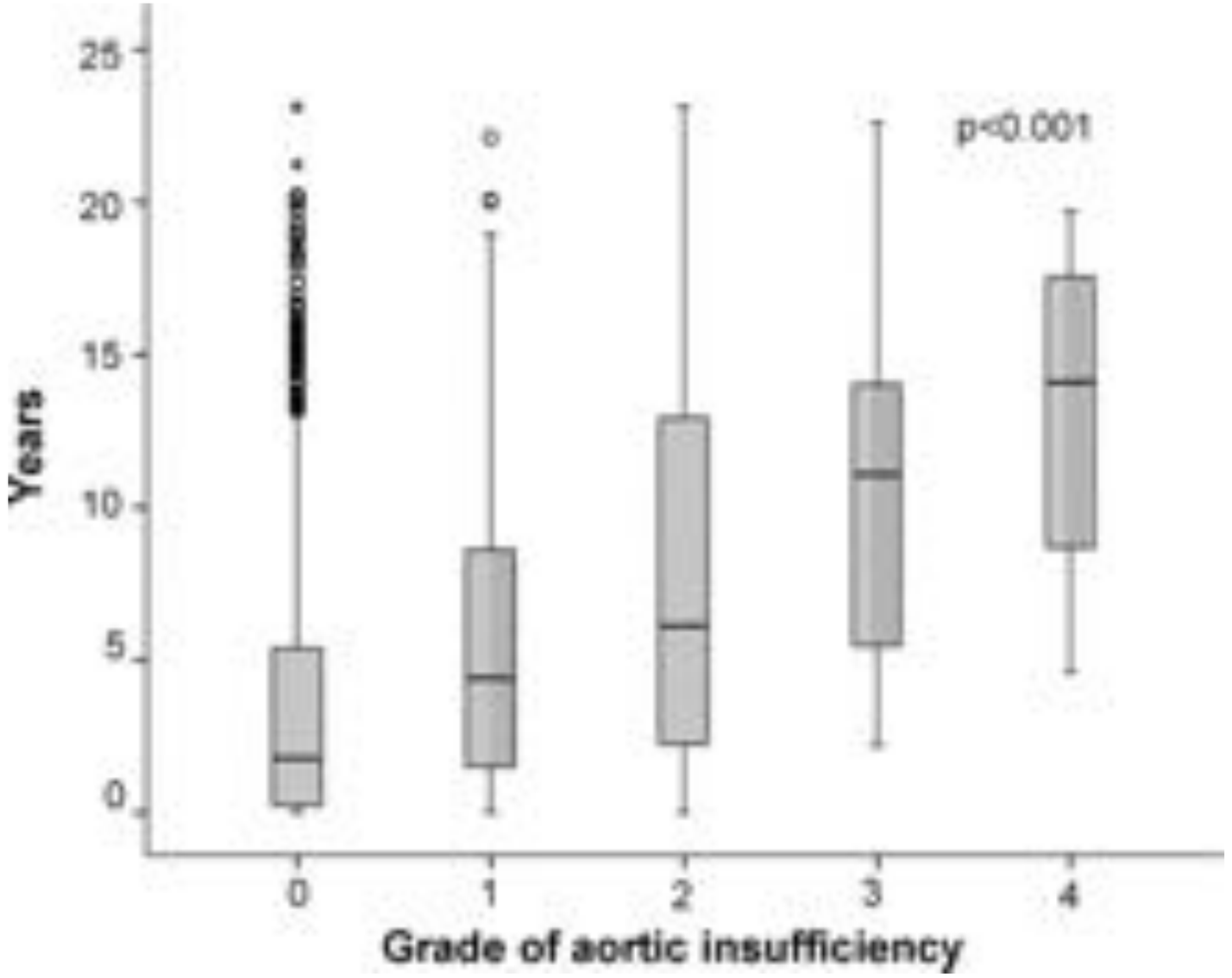
Freedom from aortic root dilation over time since ASO

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



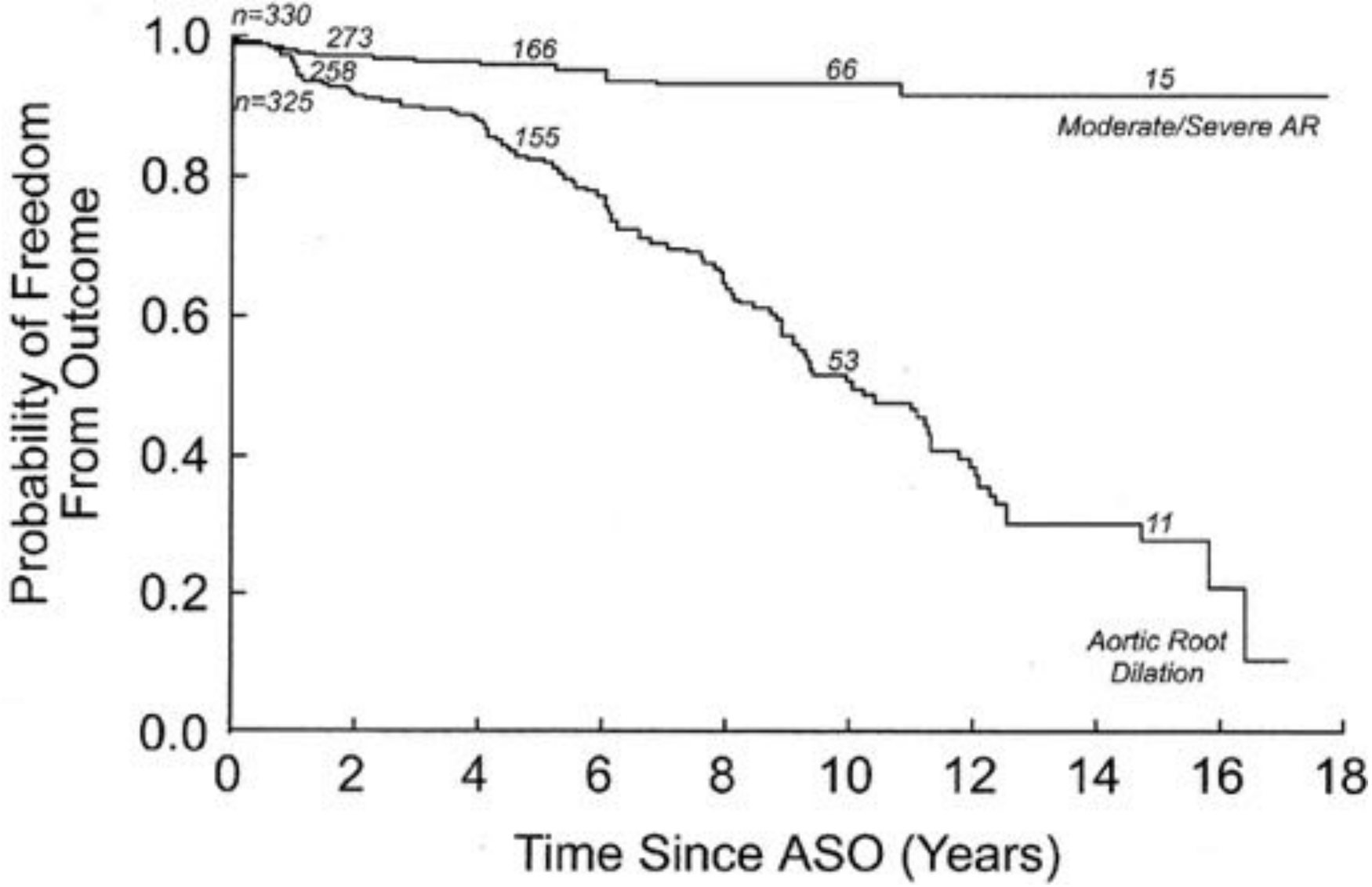
Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132

Development of Aortic Insufficiency over time

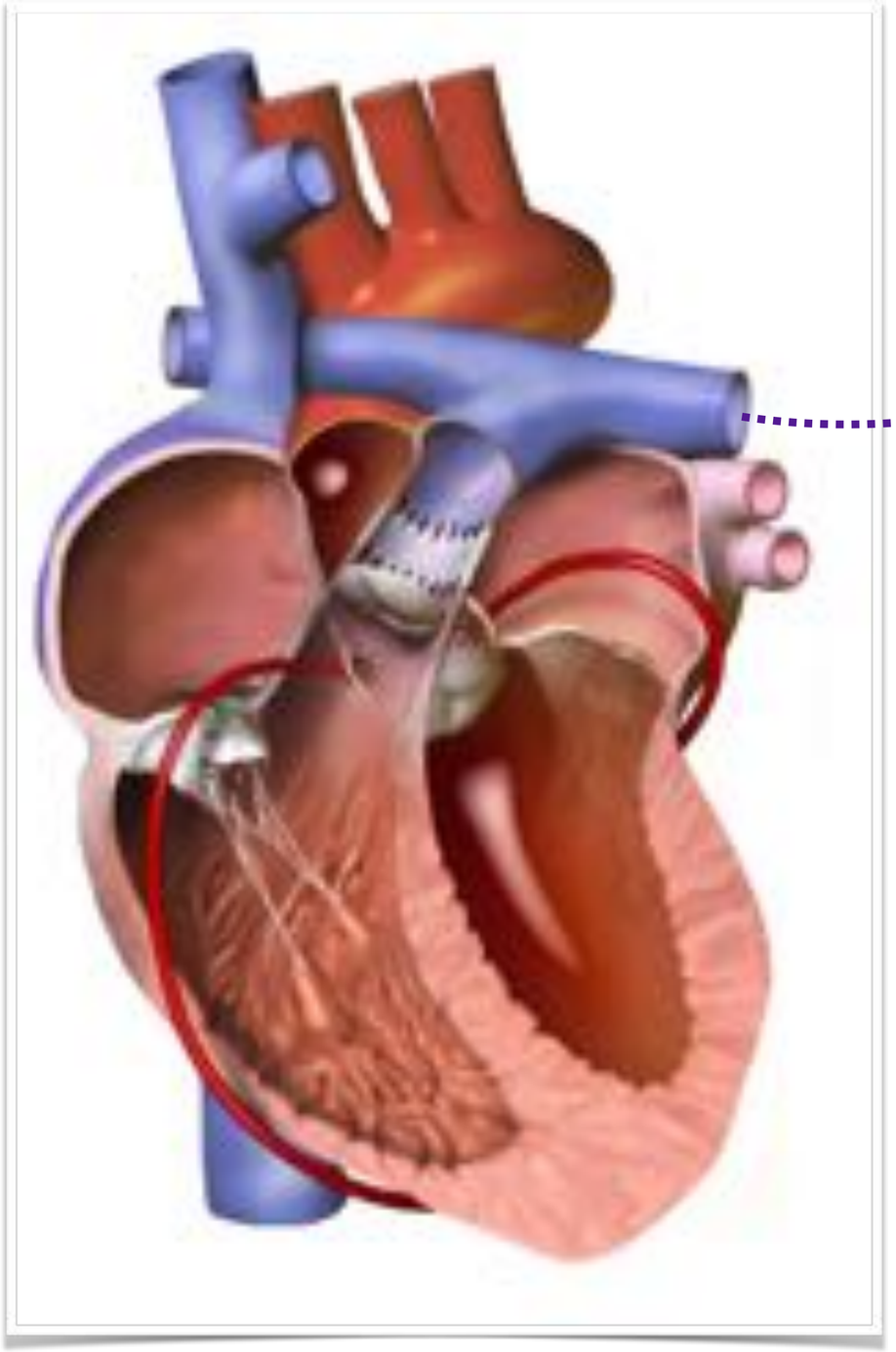


Lange R. et al.; Eur J Cardiothorac Surg 2008;34:711-717

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

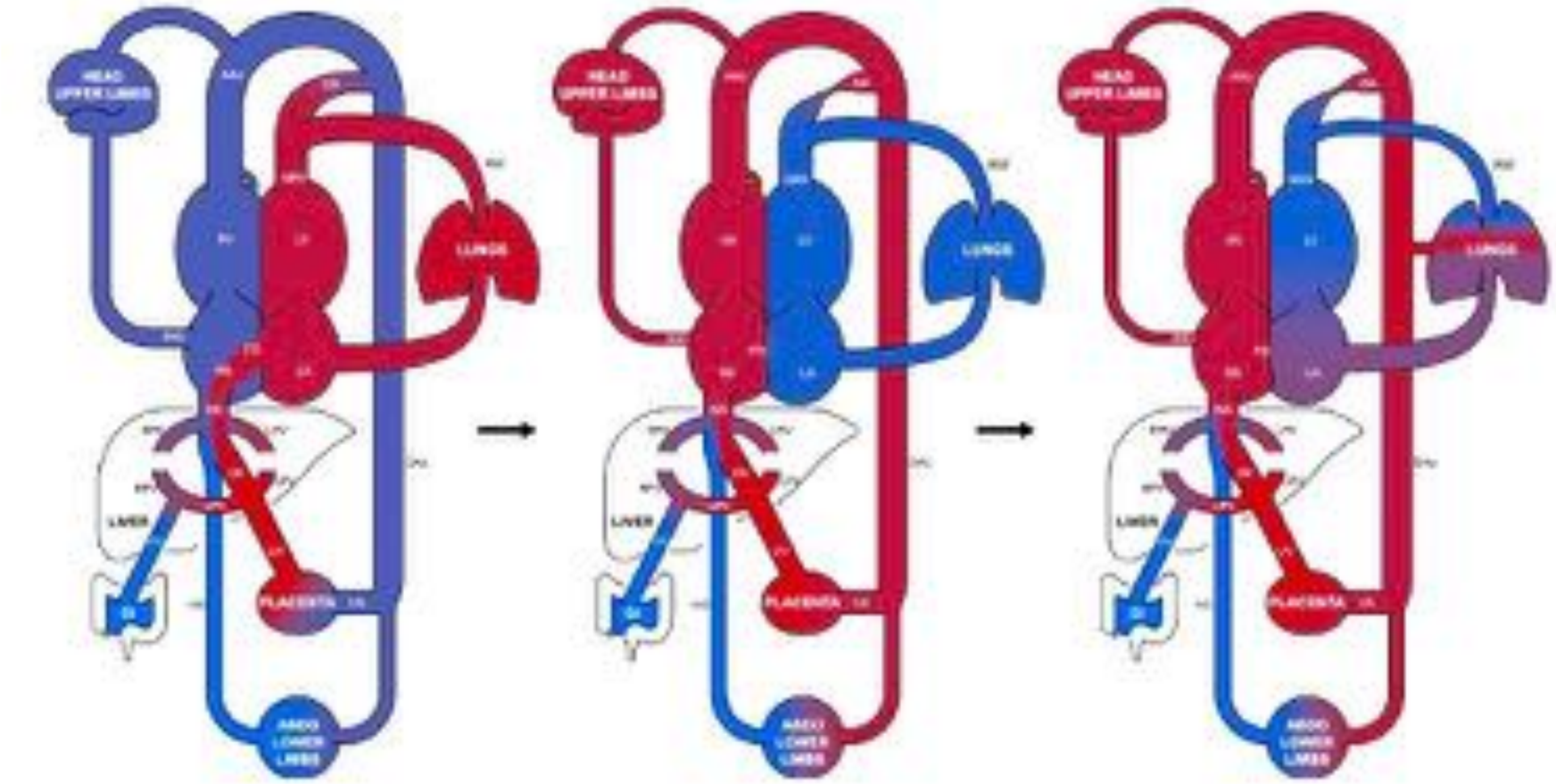


Schwartz, M. L. et al. Circulation 2004;110:II-128-II-132



Pulmonary arterial hypertension

Pulmonary circulation in fetal TGA



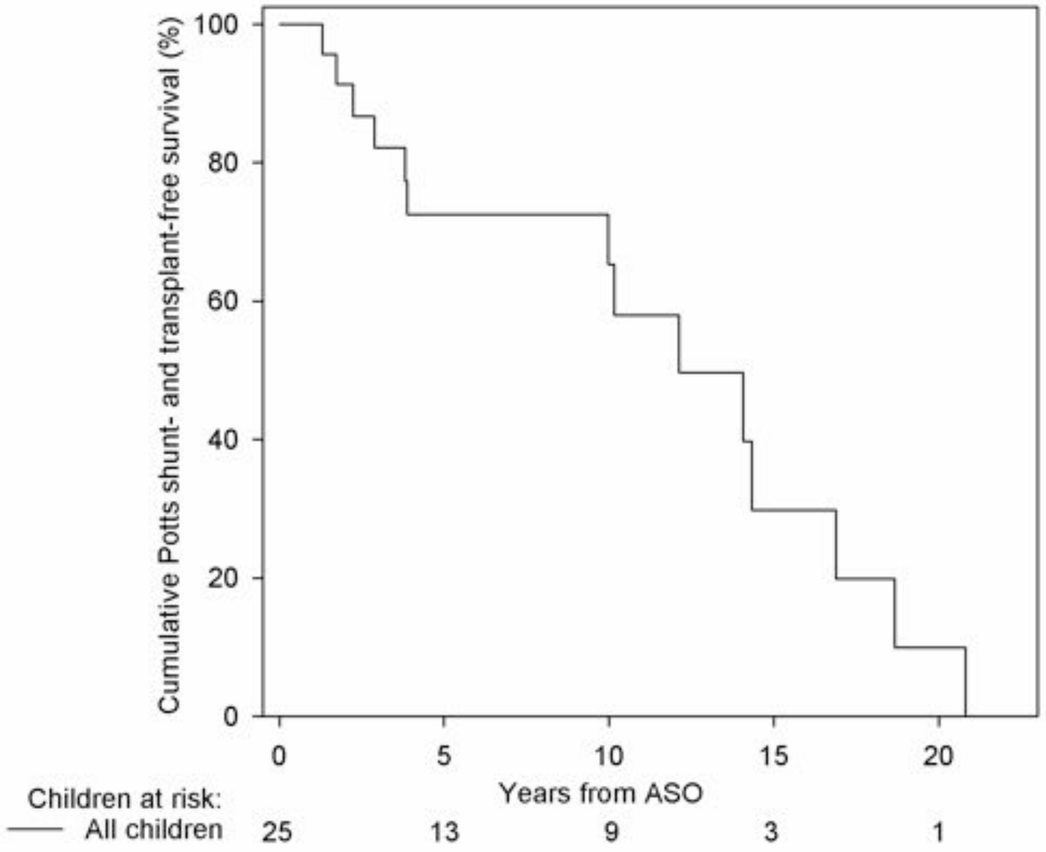
Initial increase in PBF due to vasodilatation with increased oxygen
Increased pulmonary venous return
Reduced size of the FO
Ductal constriction due to oxygen
Isolation of Pulmonary circulation
Increased PVR
Development of aorta-pulmonary collaterals

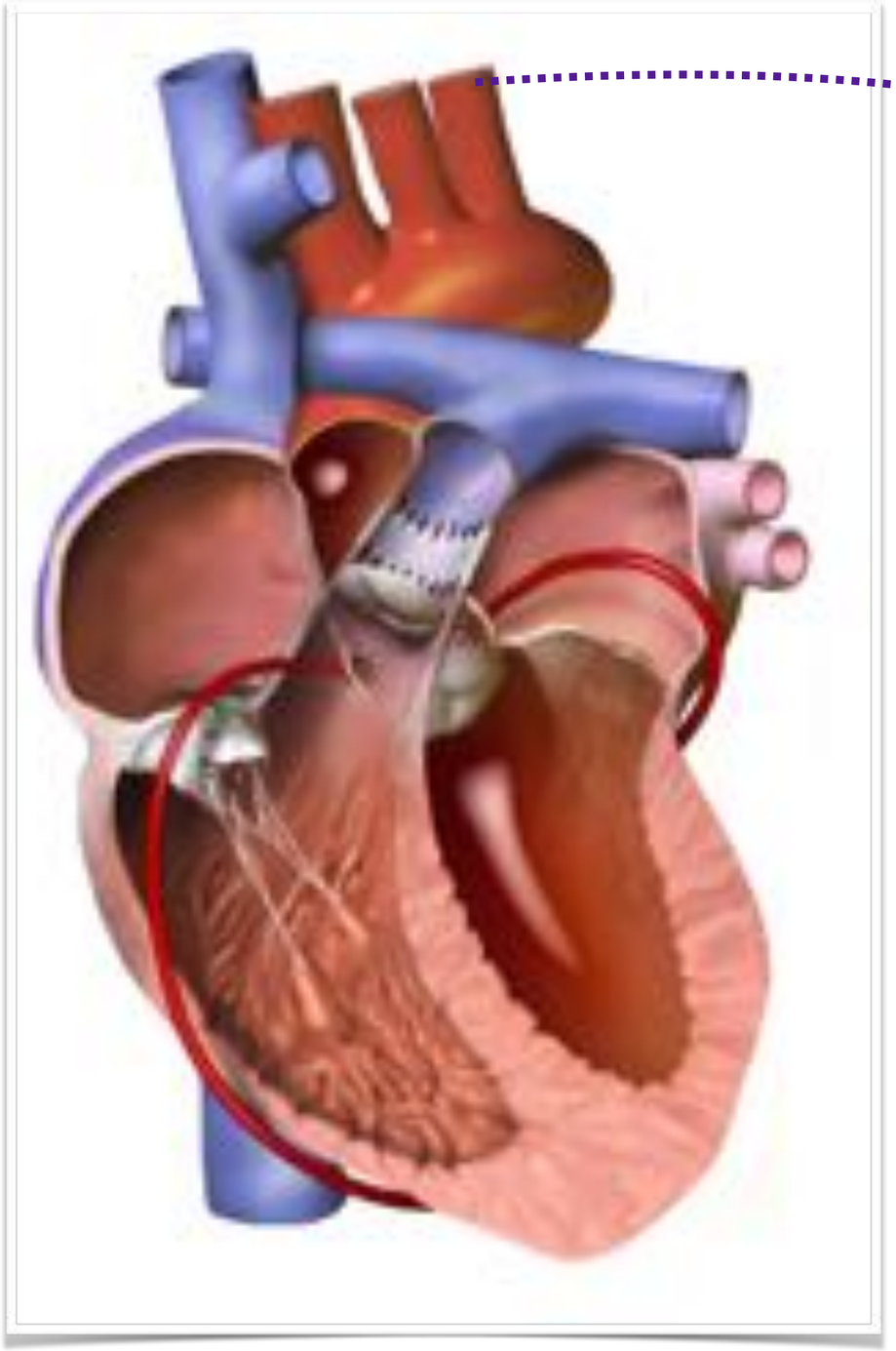
PAH characteristics after ASO for TGA

	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



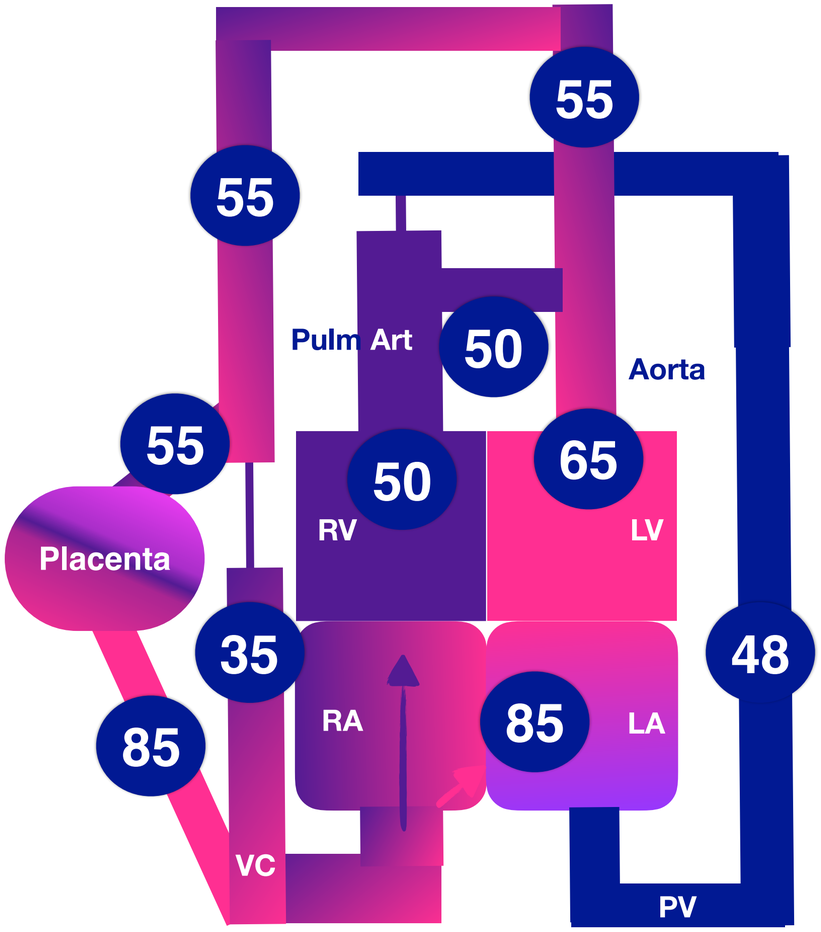


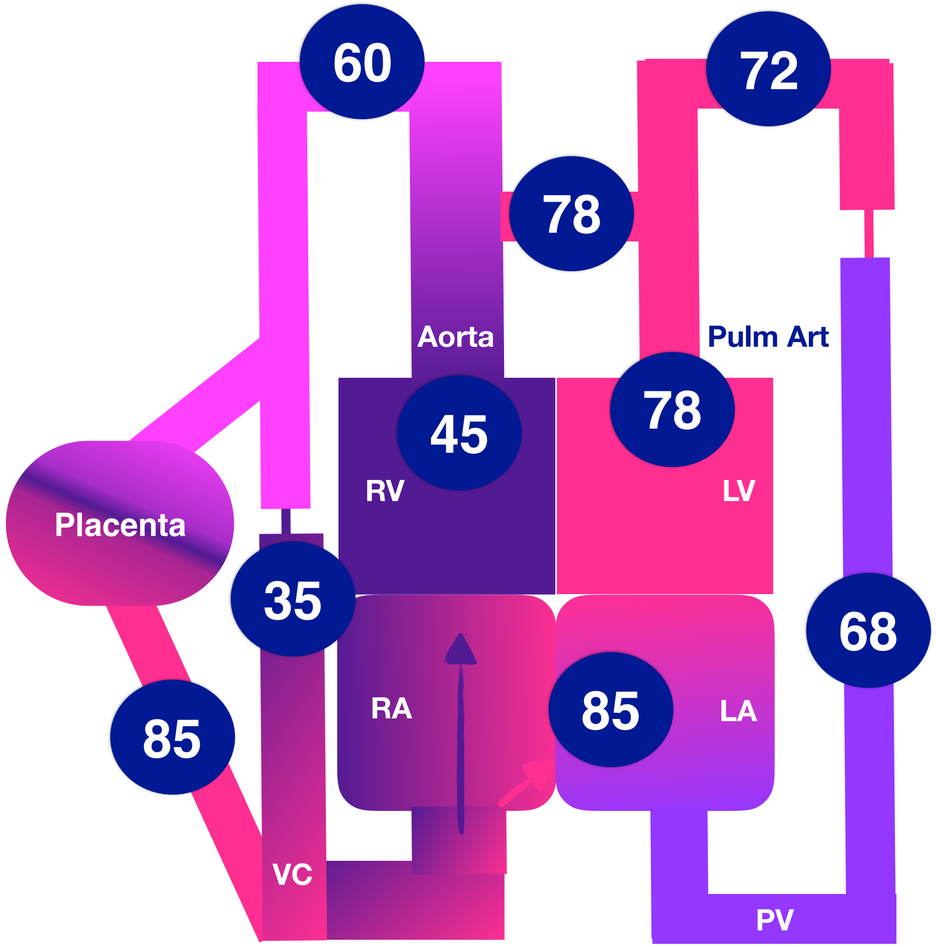
Neurodevelopmental outcomes

Susceptible Host

Fetal circulation and brain perfusion in TGA

Normal





TGA

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

Susceptible Host

Brain MRI anomalies in children with cyanotic congenital heart diseases

- **Brain immaturity in term newborns**
(Licht et al., 2009; Andropoulos et al., 2010)

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

- **Myelin maturation anomalies**
  Interhemisphere connection and frontal lobes
(Makki et al., 2013; Ibuki et al., 2012)

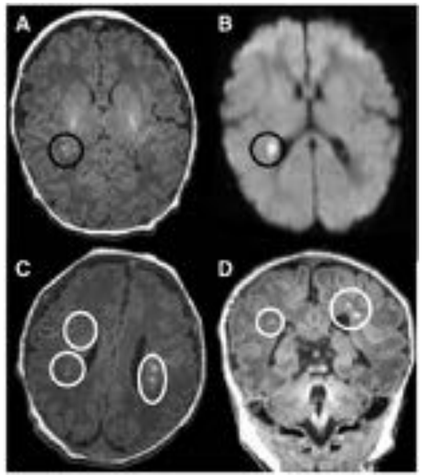


Fig. 1 White matter anomaly in TGA
(Petit et al., 2009)

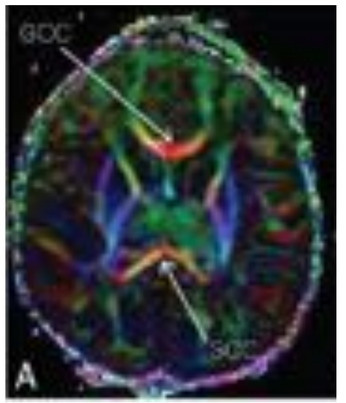
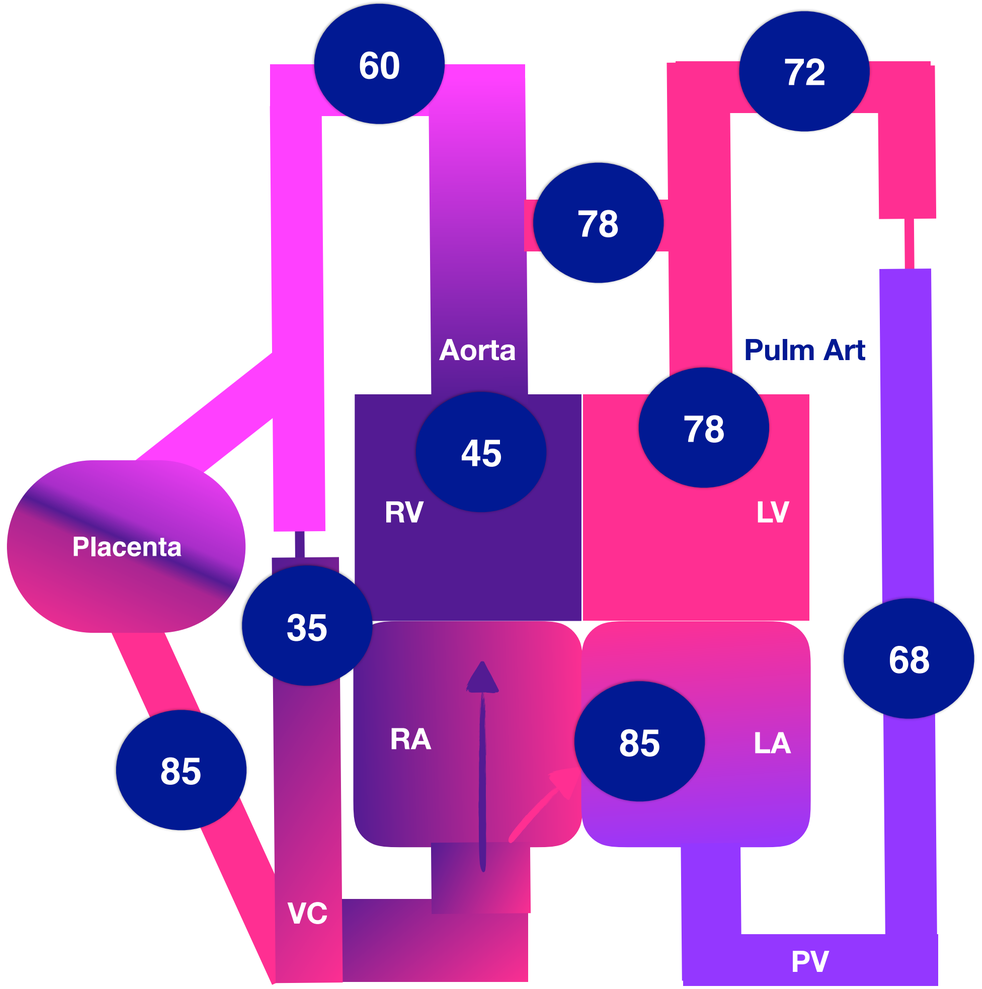
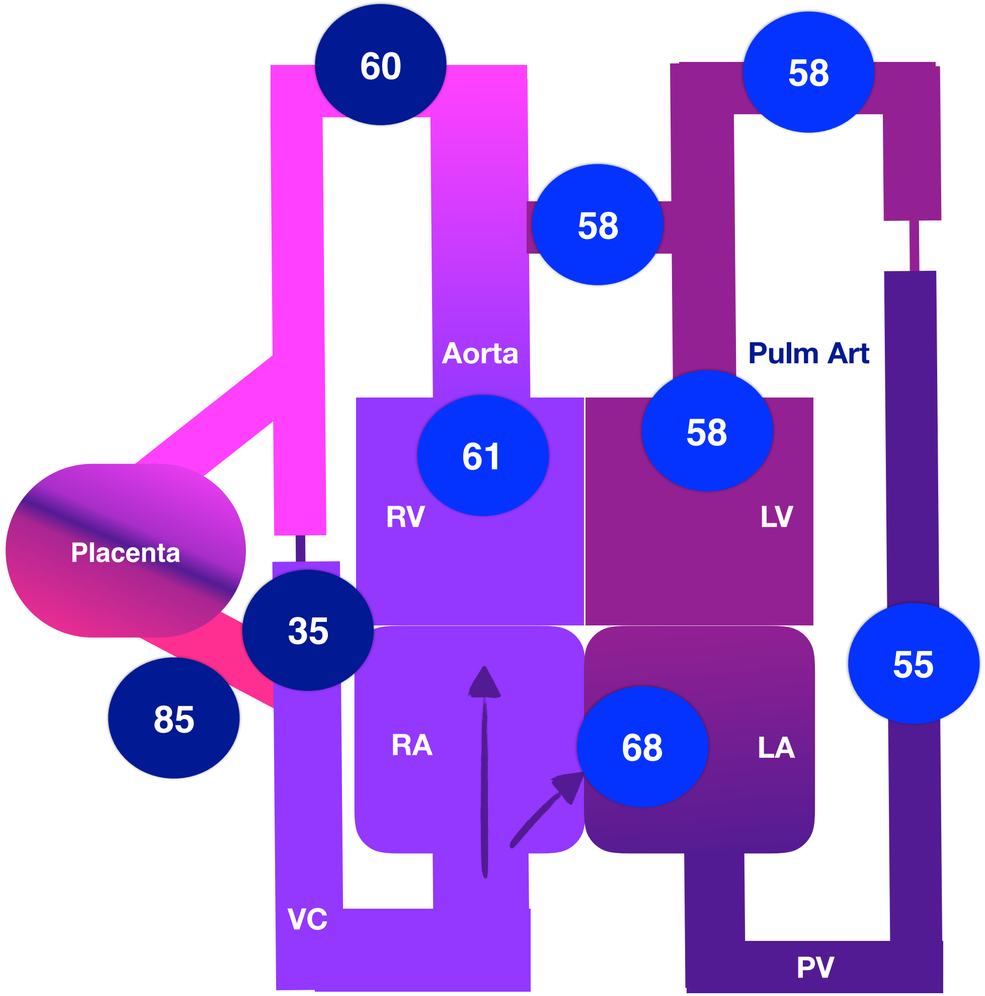


Fig. 2 DTI interhemisphere connection anomalies in complex CHD
(Makki et al., 2013)



TGA fetus



Closure of the ductus venous

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

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

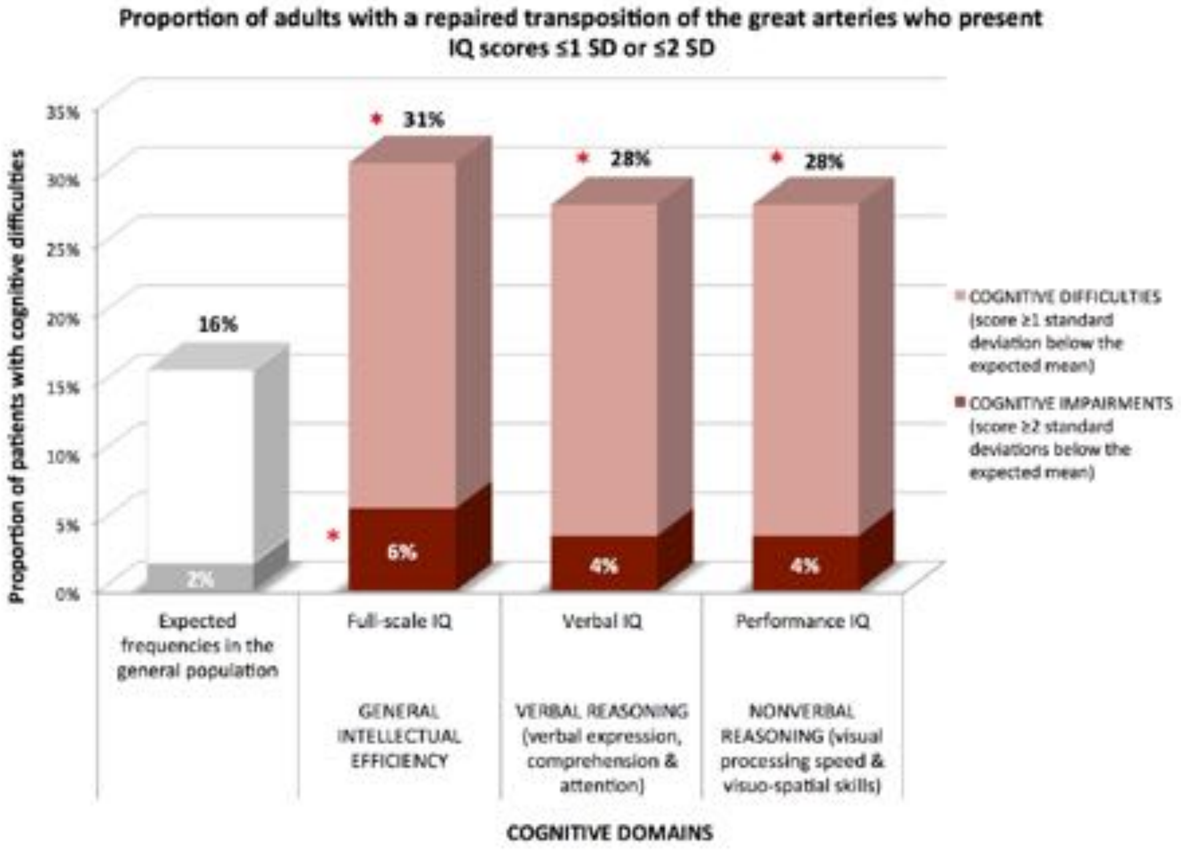
Calderon J et al. J Pediatr 2012

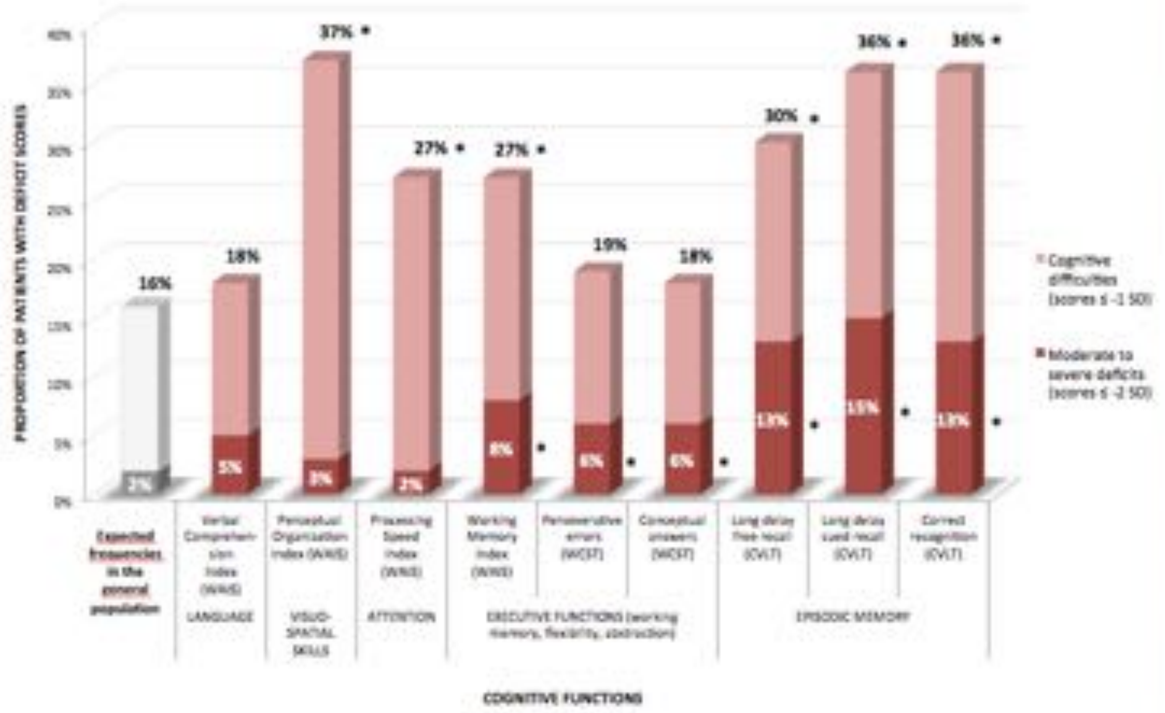
ToM tests in TGA vs controls
role of prenatal diagnosis

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

Neuropsychological and Psychiatric Issues in d-TGA and Suggested Interventions

Childhood

- Delays in psycho-motor development
- Visual-spatial deficits
- Executive functions deficits:
  - Self-regulation
  - Working Memory
  - Cognitive Flexibility
- Social cognition impairments:
  - Theory of Mind
- Behavioral difficulties

Adolescence

- Executive functions deficits:
  - Working Memory
  - Cognitive Flexibility
  - Planning
- High prevalence of Attention Deficit Hyperactivity Disorder (ADHD)
- Social cognition impairments
- Mental health issues:
  - Anxiety
  - Depression

Adulthood

- Reduced global cognitive functionning
- Poor executive functions:
  - Goal-directed behavior
  - Planning
  - Cognitive Flexibility
- Mental health issues:
  - Anxiety
  - Depression

Evidenced-based interventions with age-appropriate strategies:

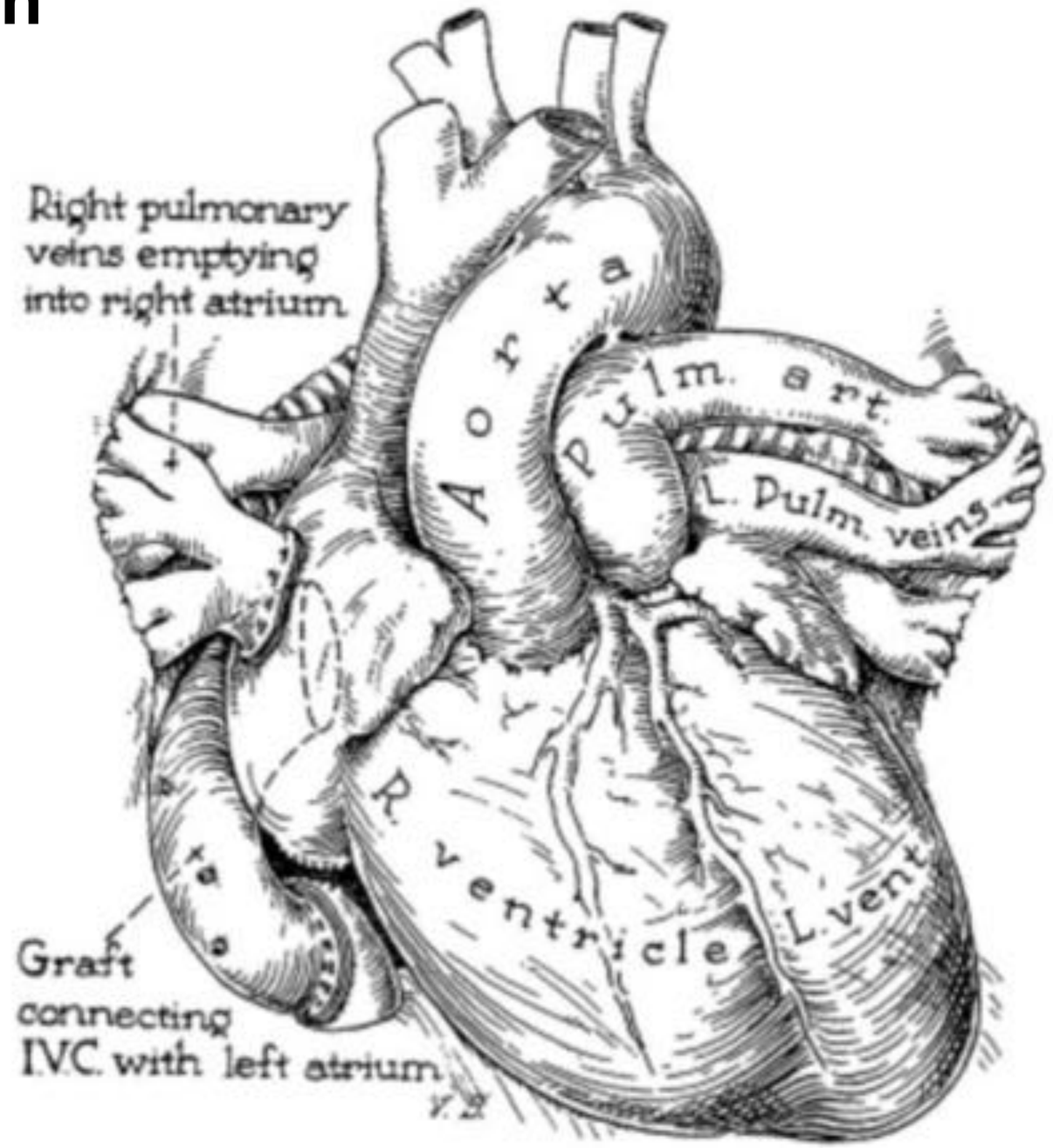
- ✓ Structured computerized training of specific impaired cognitive abilities (e.g., Working Memory training programs)
- ✓ Individual Cognitive Behavioral Therapy and/or group support therapy for mental health issues
- ✓ Mindfulness for better executive functioning and stress reduction
- ✓ Pharmacological treatment of ADHD (psycho-stimulant medication) and severe psychiatric disorders (anxiety and/or depression medication)

Kasmi L et al. Frontiers in Pediatrics 2017 in press

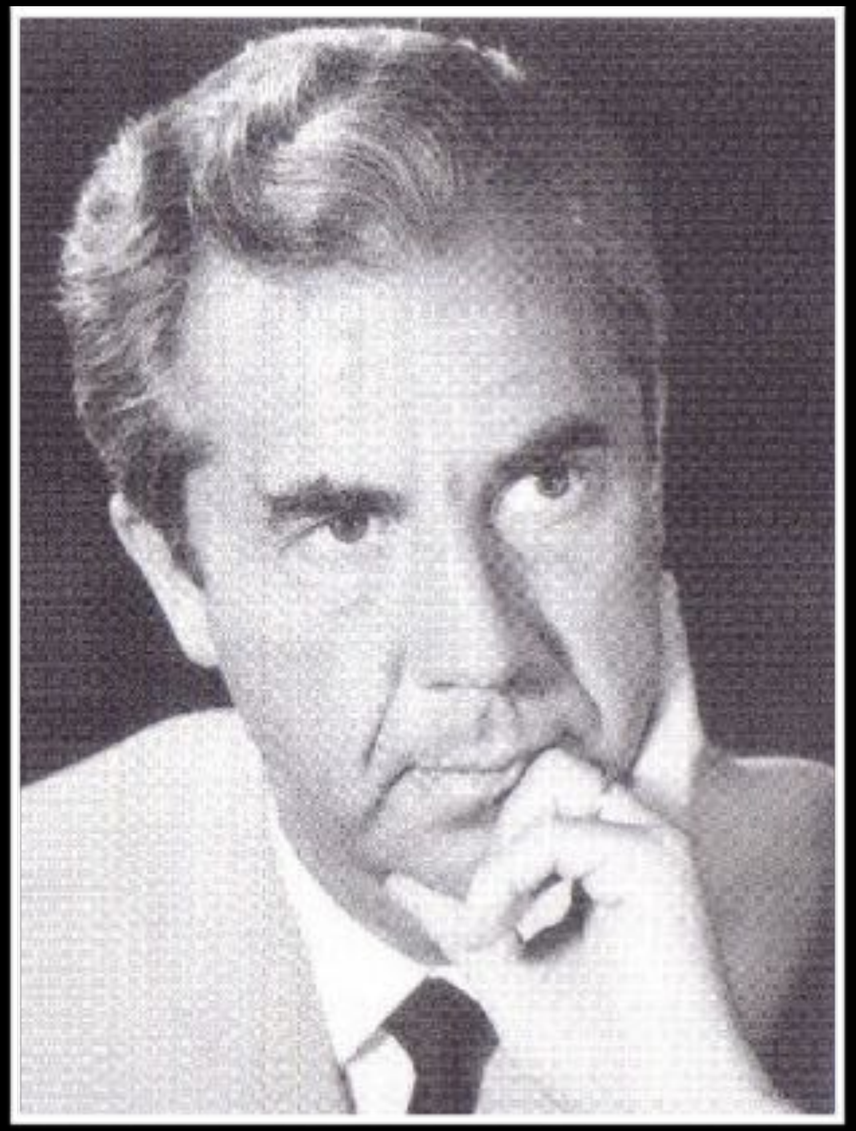
Atrial repair of TGA

The Senning and Mustard operations

Baffes operation



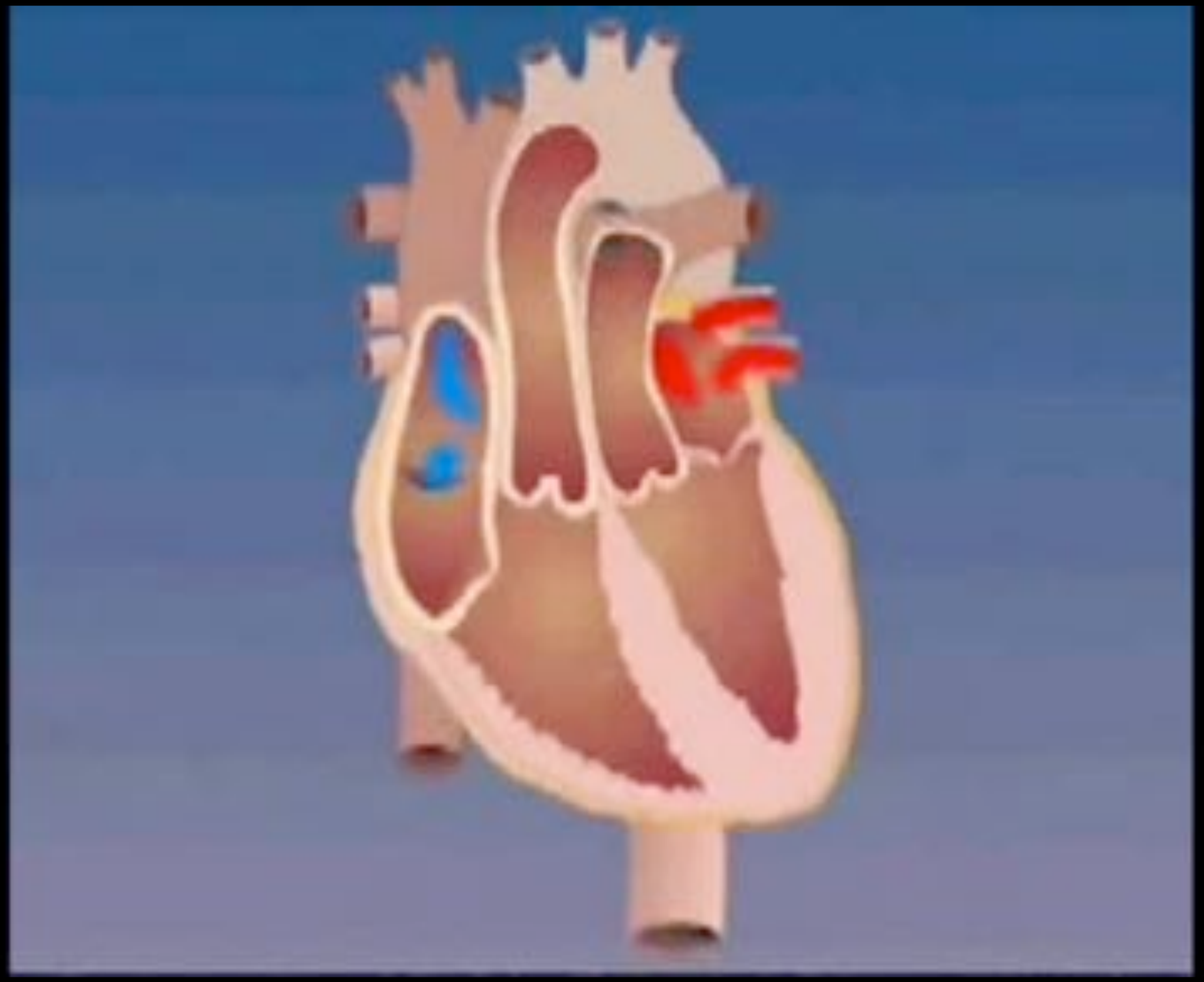
Atrial correction of TGA



Ake Senning

William Thornton Mustard

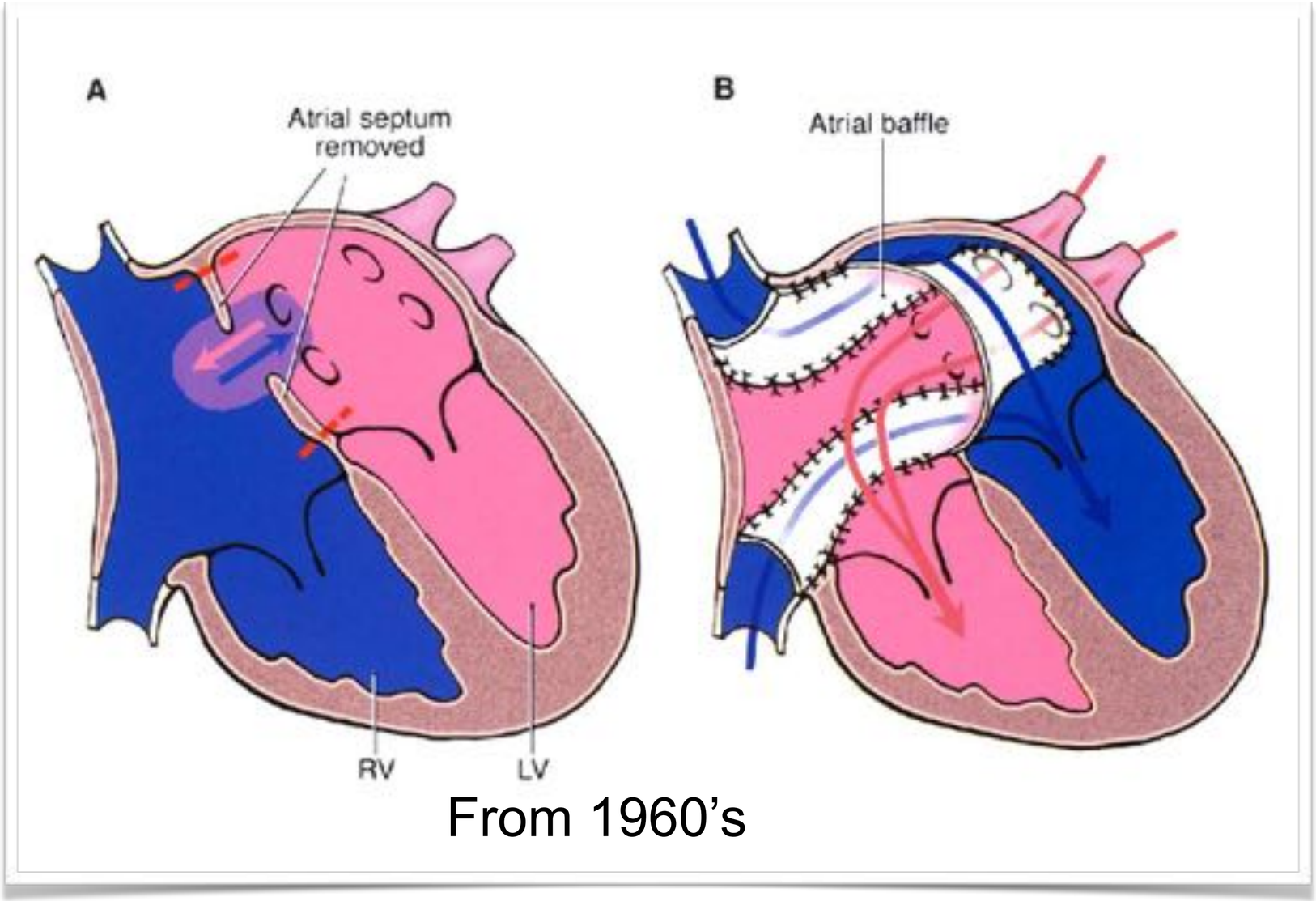
Mustard operation



Atrial switch

Mustard procedure: pericardium

Senning procedure: atrial tissue

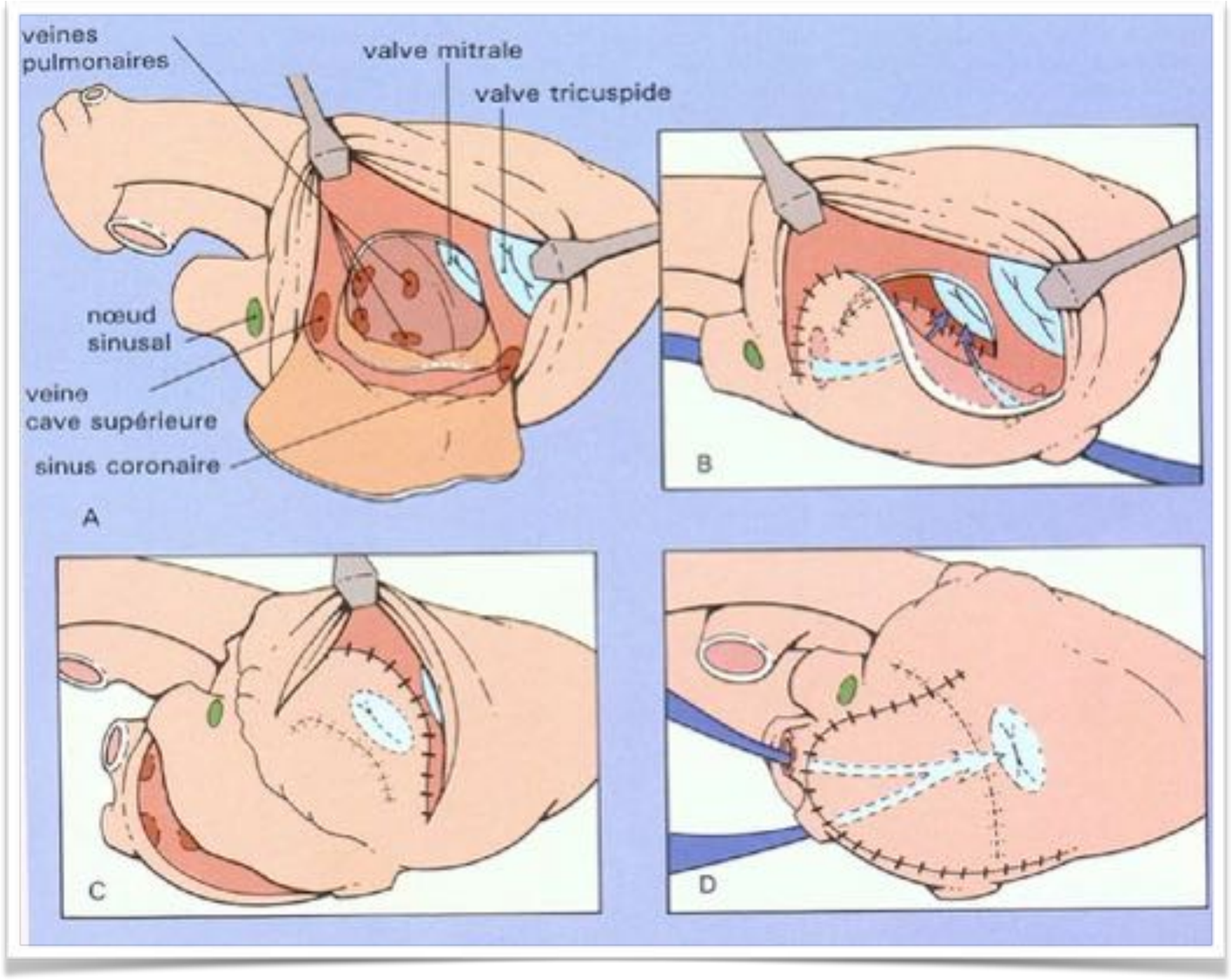


From 1960’s

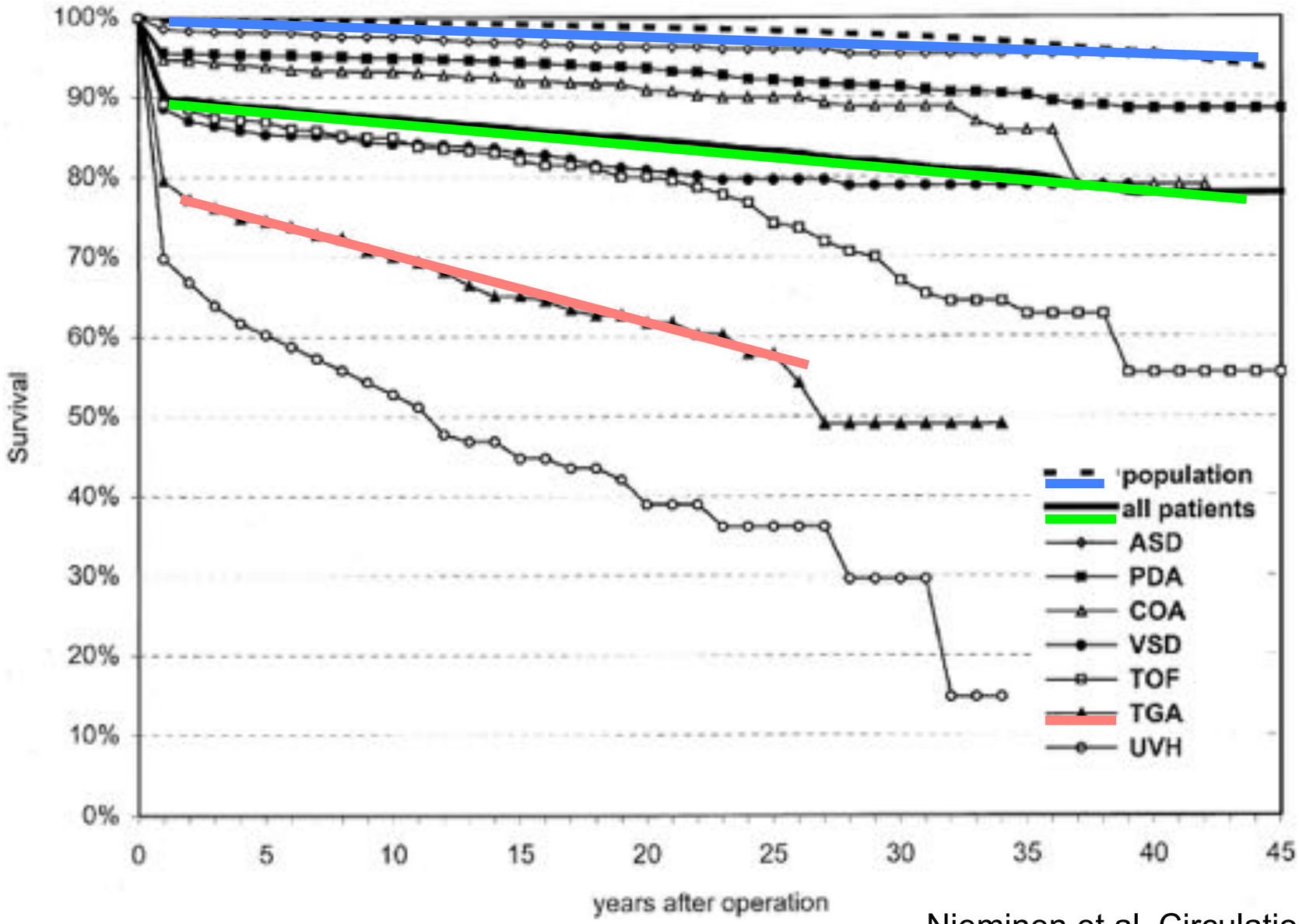
Atrial switch

Mustard procedure: pericardium

Senning procedure: atrial tissue

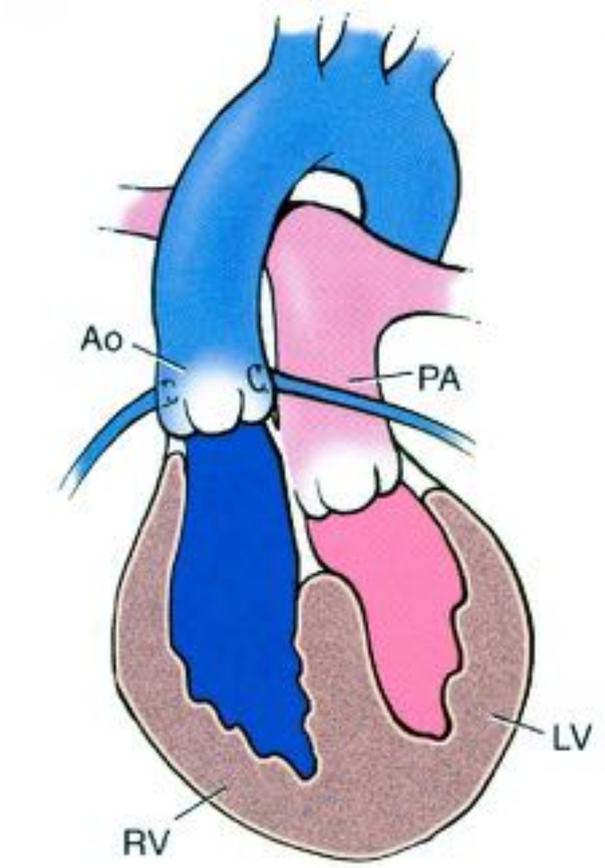


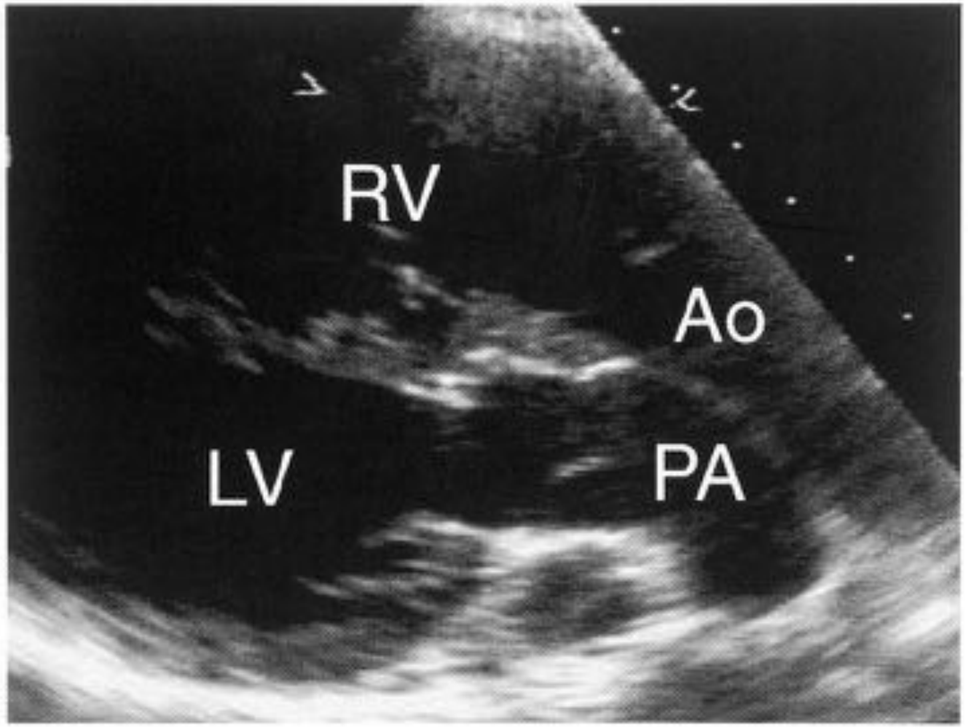
Survival of all patients, separate diagnostic groups, and general population



Nieminen et al, Circulation 2001

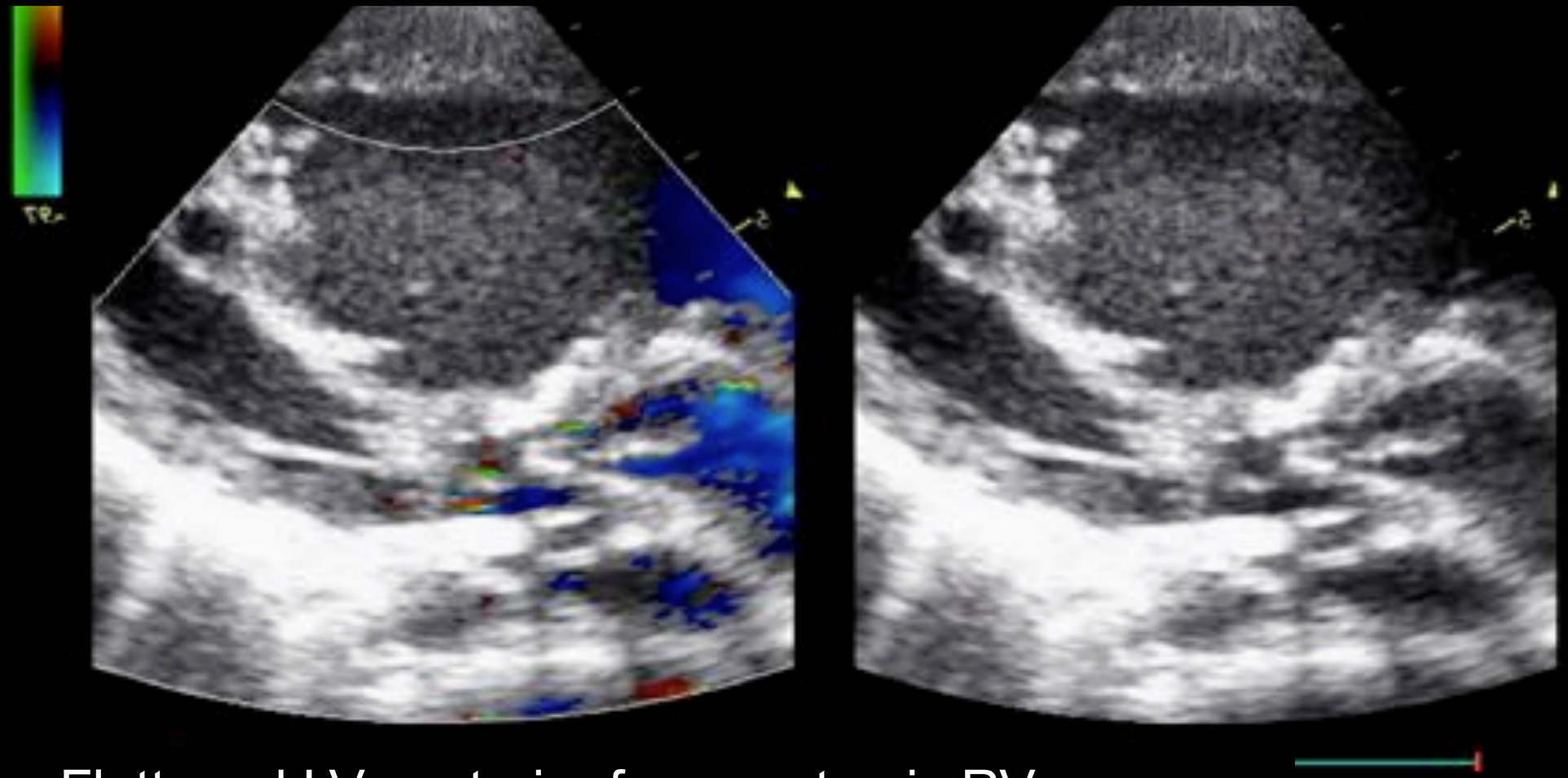
Echocardiogram long axis view





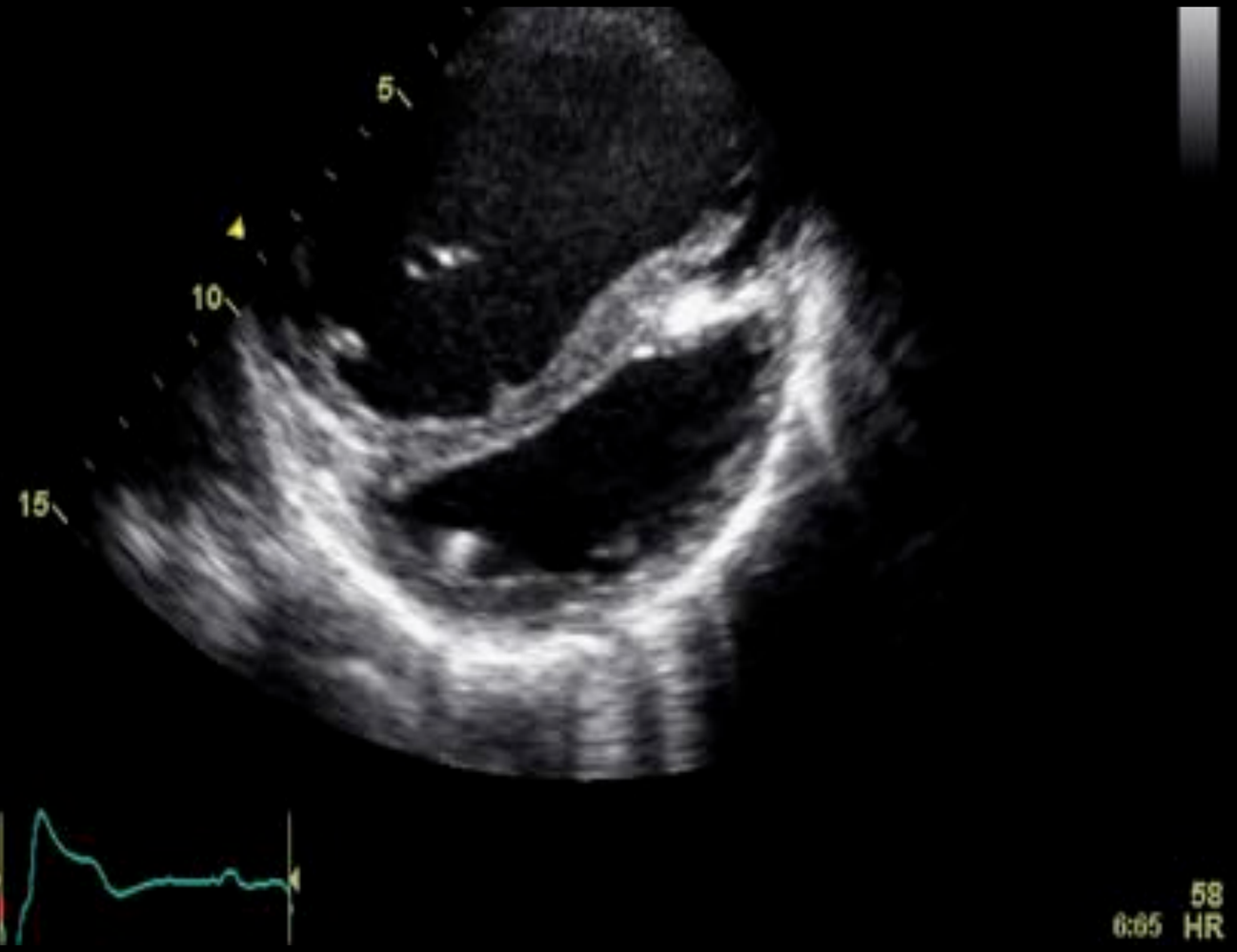
Aorta anterior, PA posterior

Long axis view after atrial correction of TGA



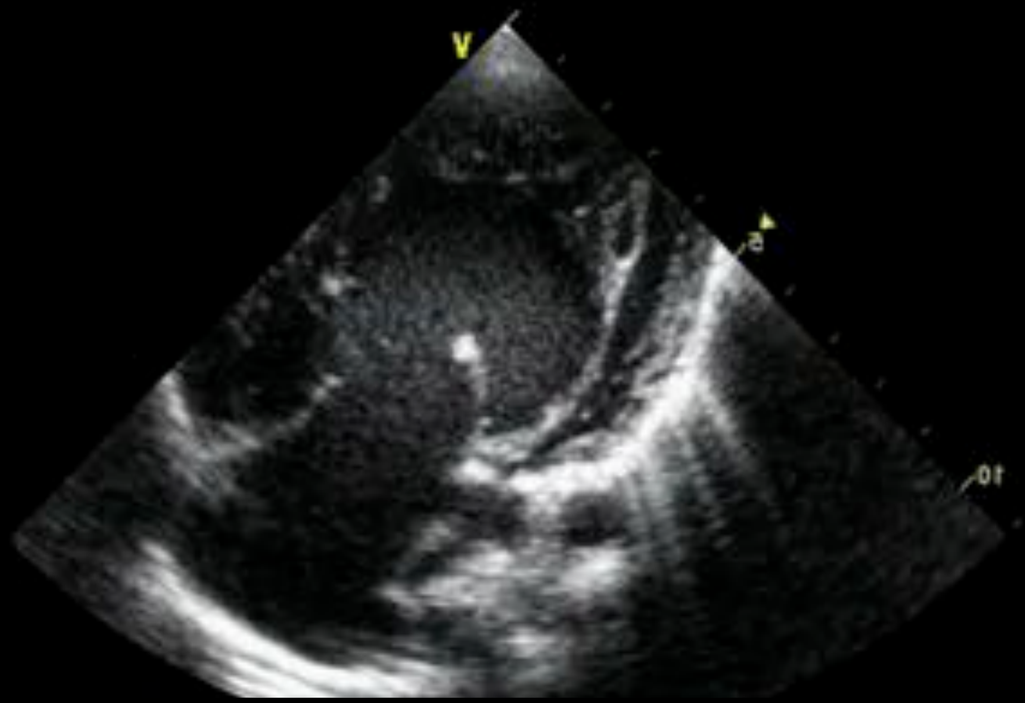
Flattened LV posterior from systemic RV

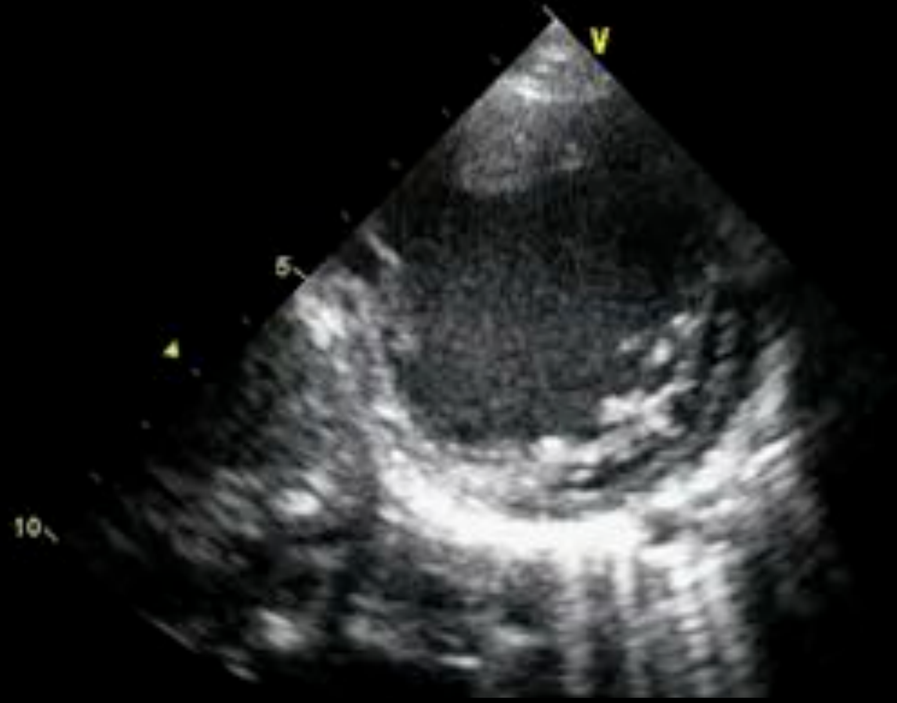
Short axis view after atrial correction of TGA



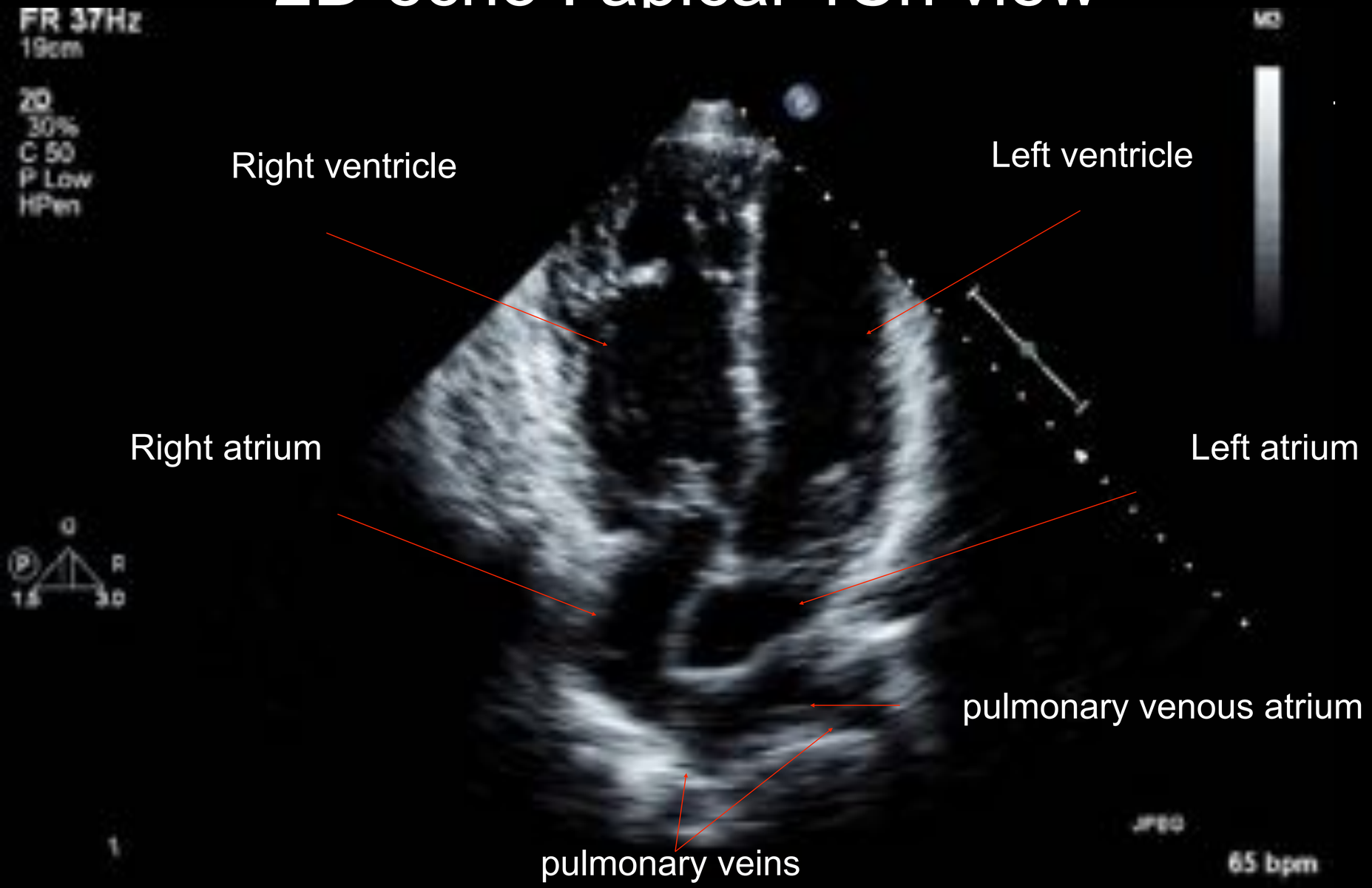
Flattened LV posterior from systemic RV

Right ventricle after atrial correction of TGA

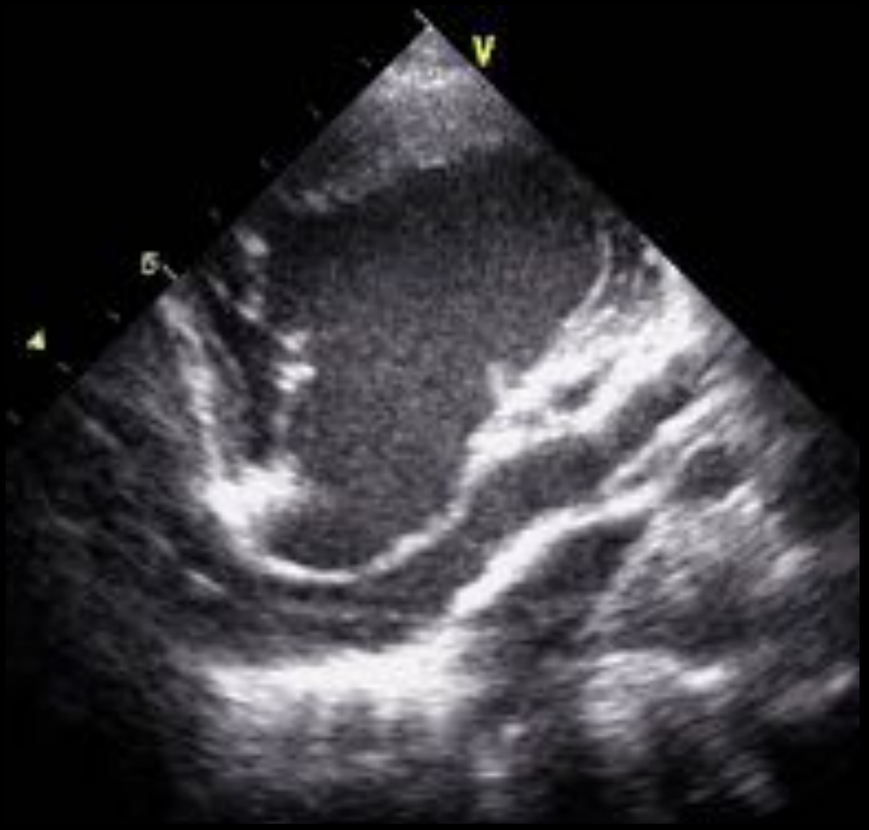




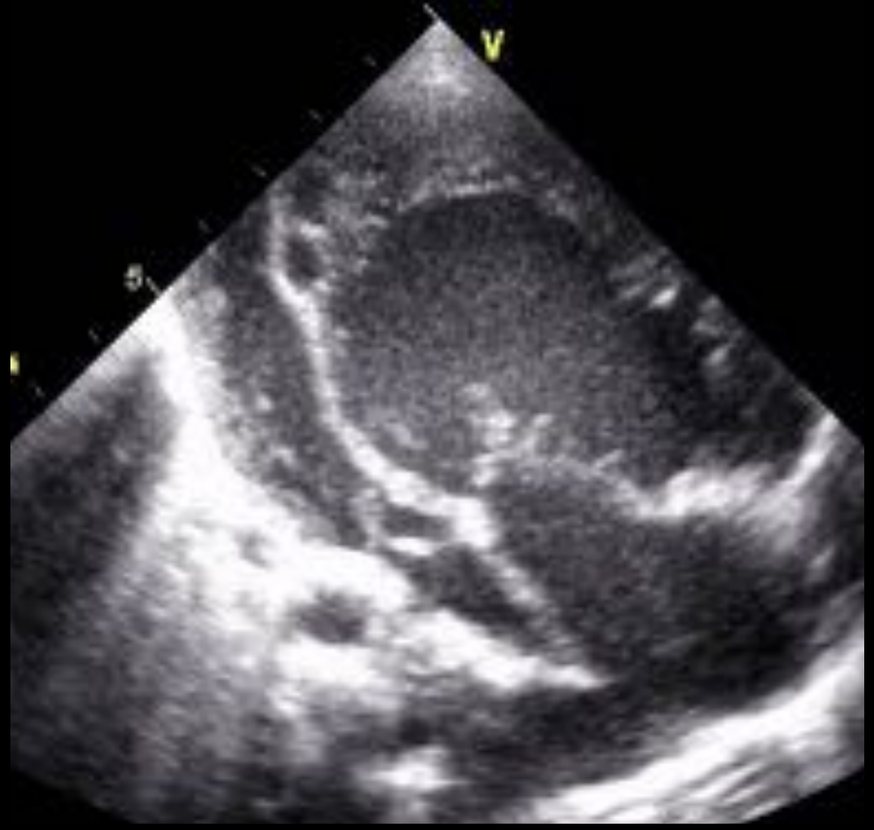
2D echo : apical 4Ch view



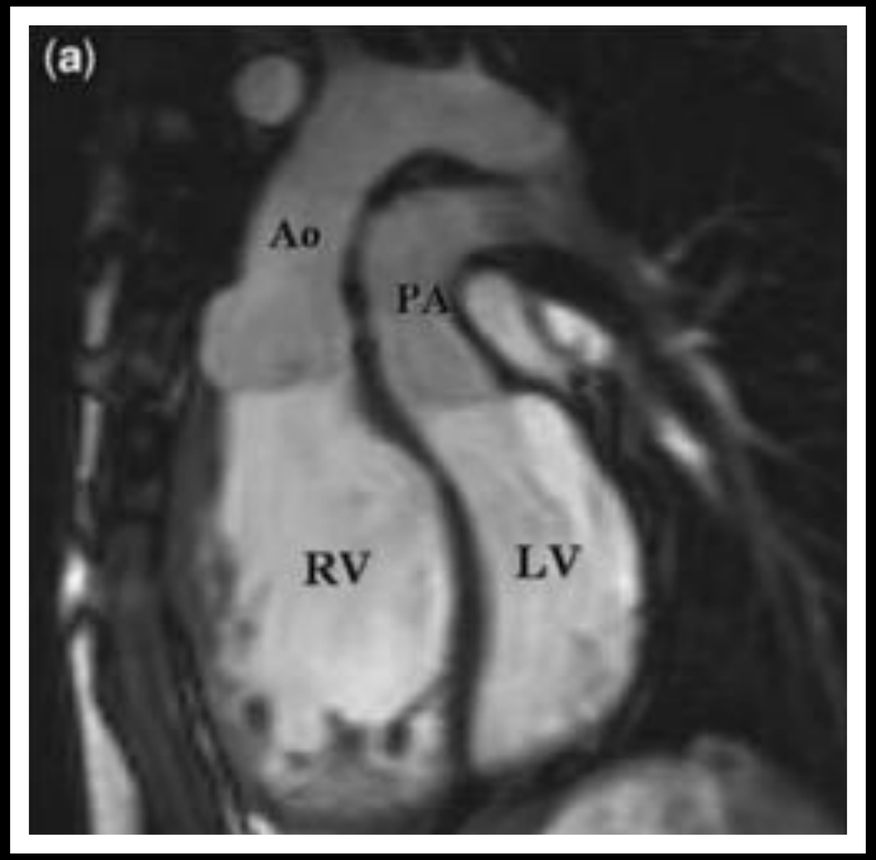
Caval baffle

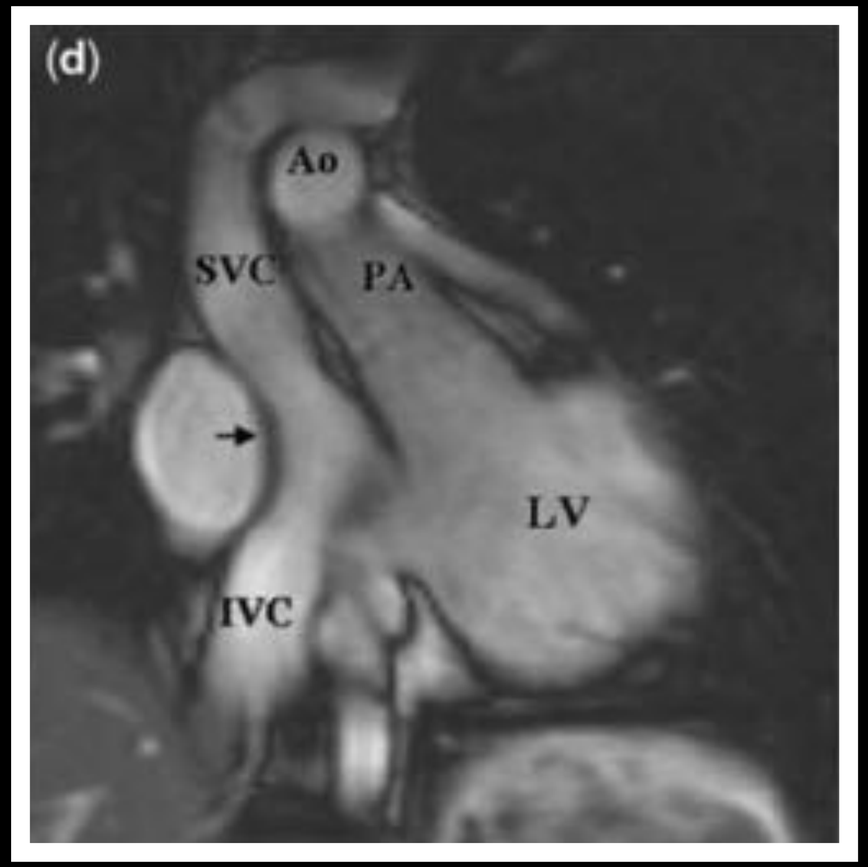


Pulmonary veins baffle



Arterial and atrial pathways in Mustard

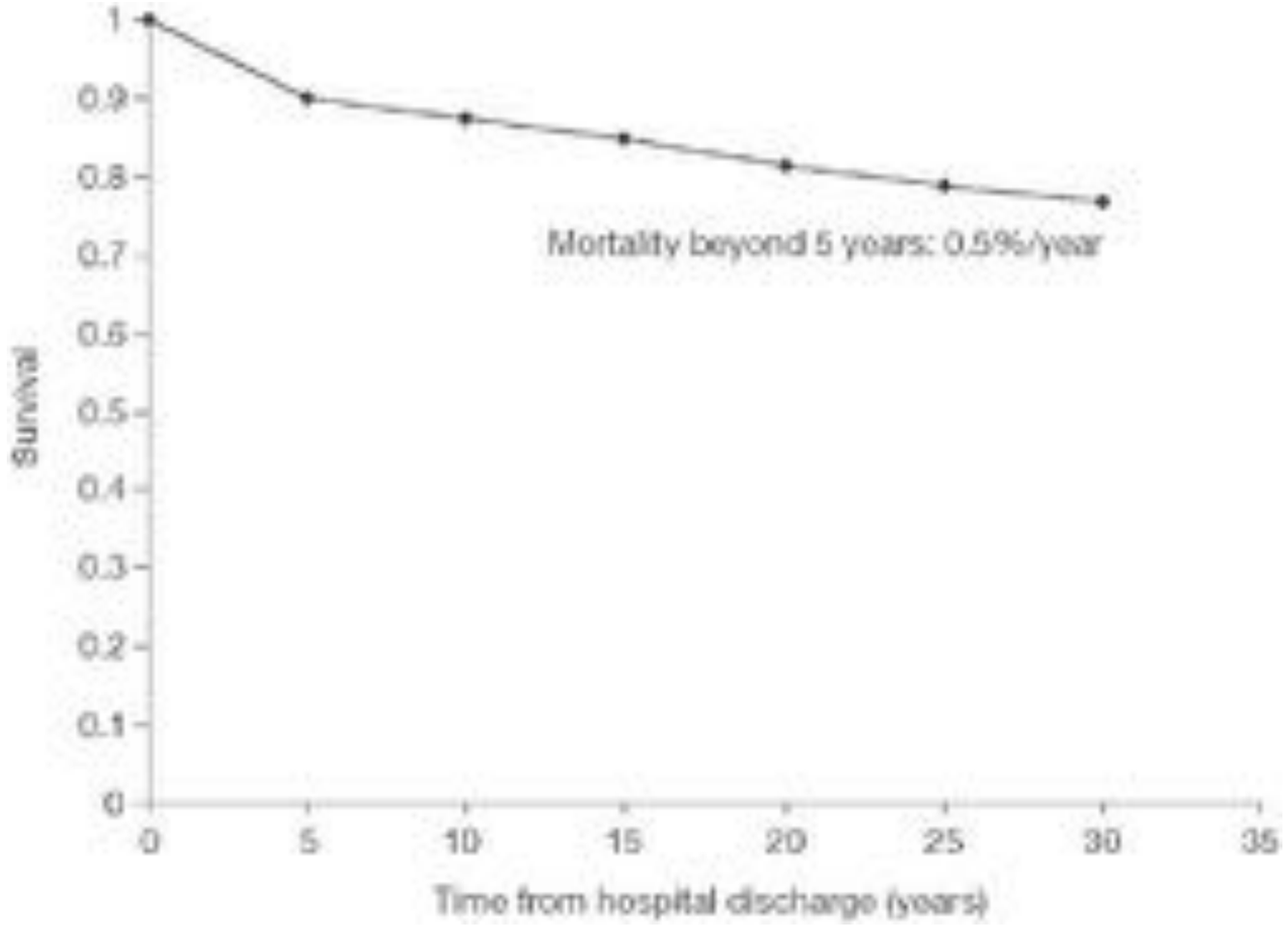




Johansson et al, Cardiol Young, 2010

Late complications after atrial switch

Early death



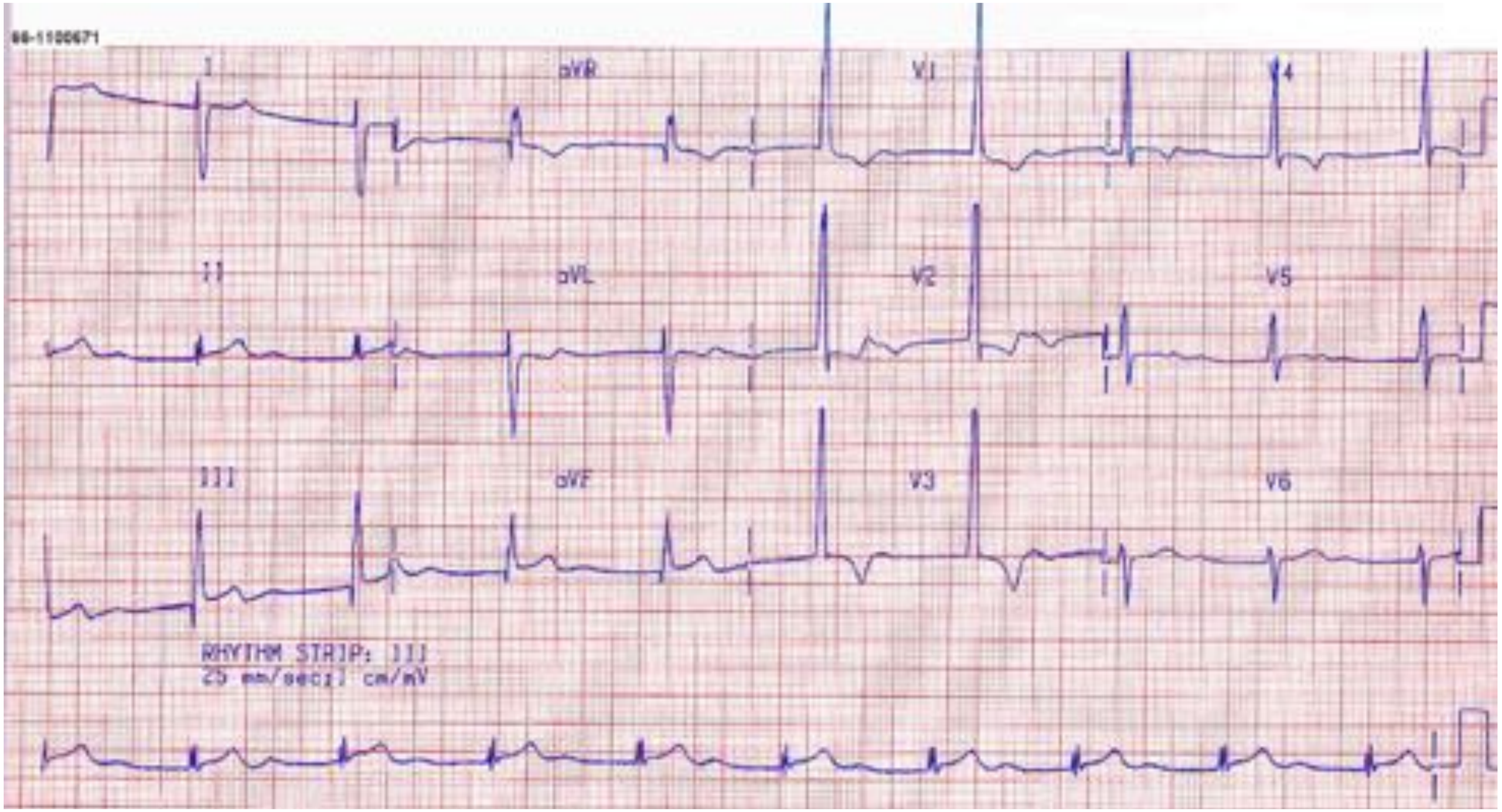
Love BA et al.Nat Clin Pract Cardiovasc Med 2008

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



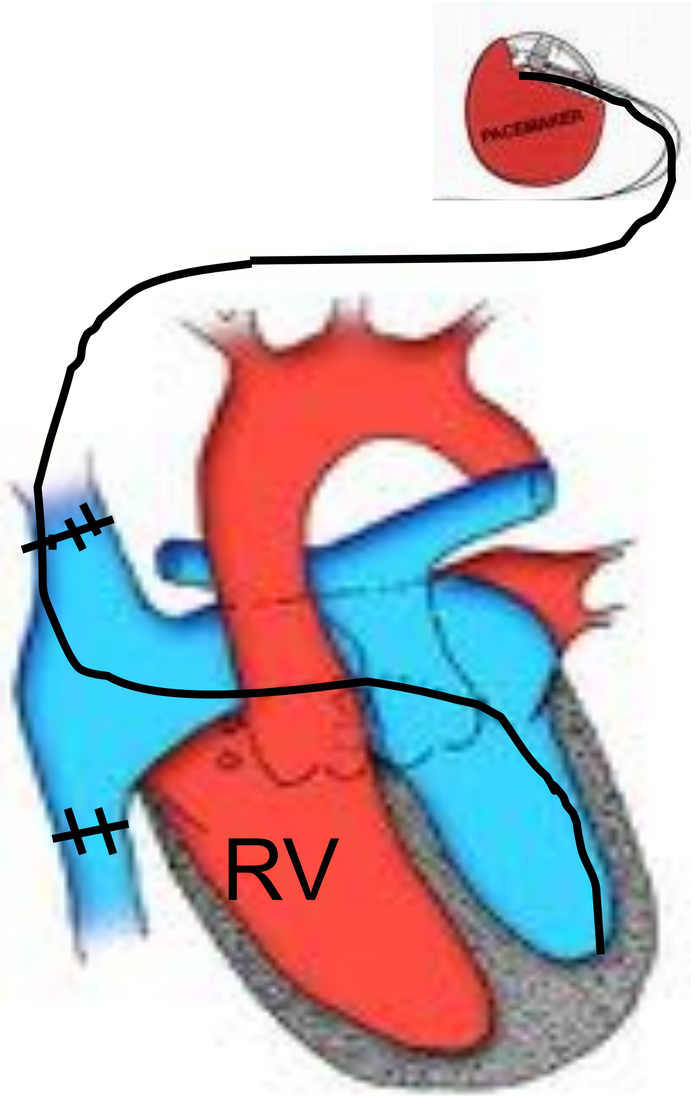
RVH

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:

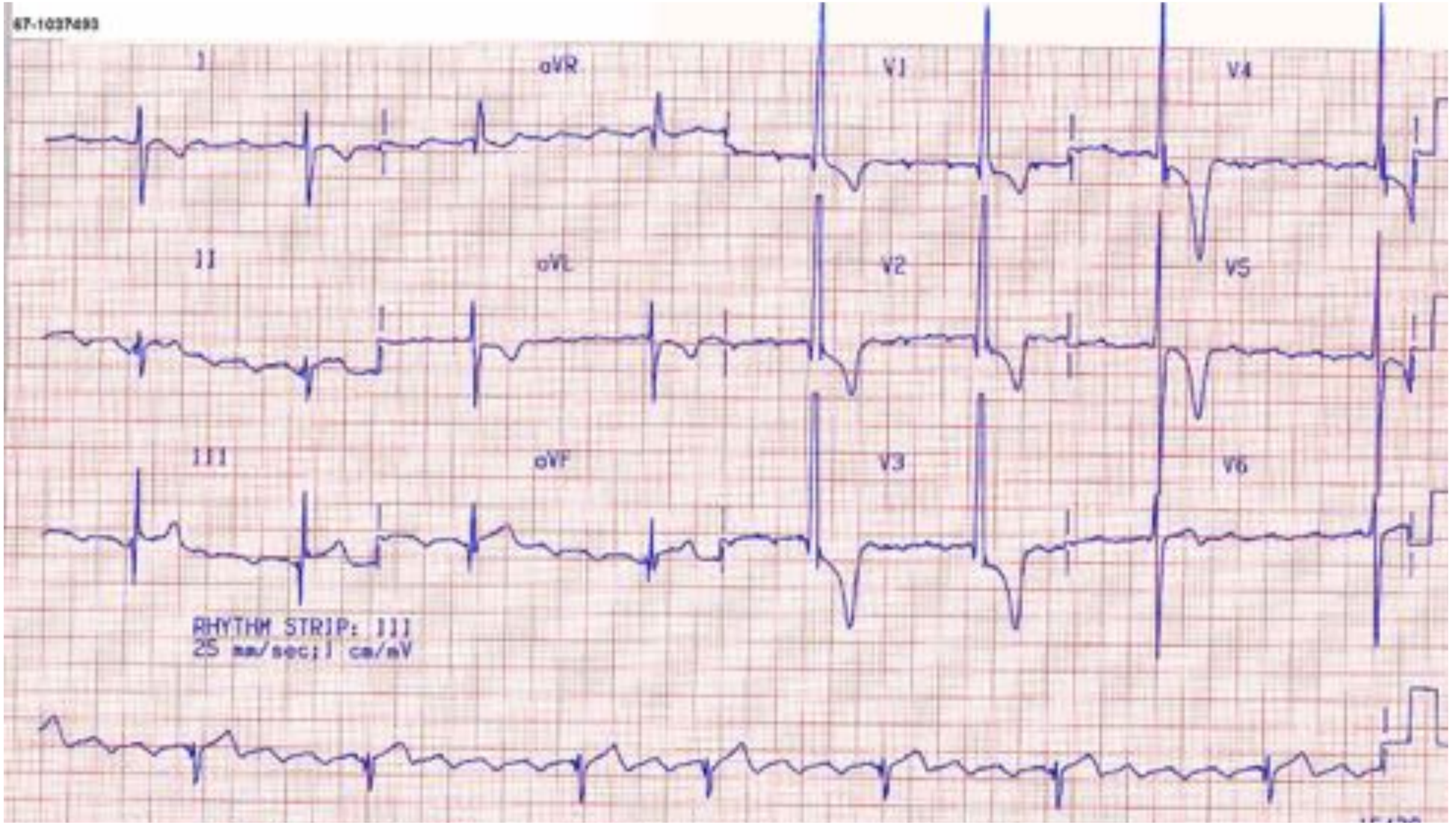
Ventricular lead will end up,

after some unusual loops,

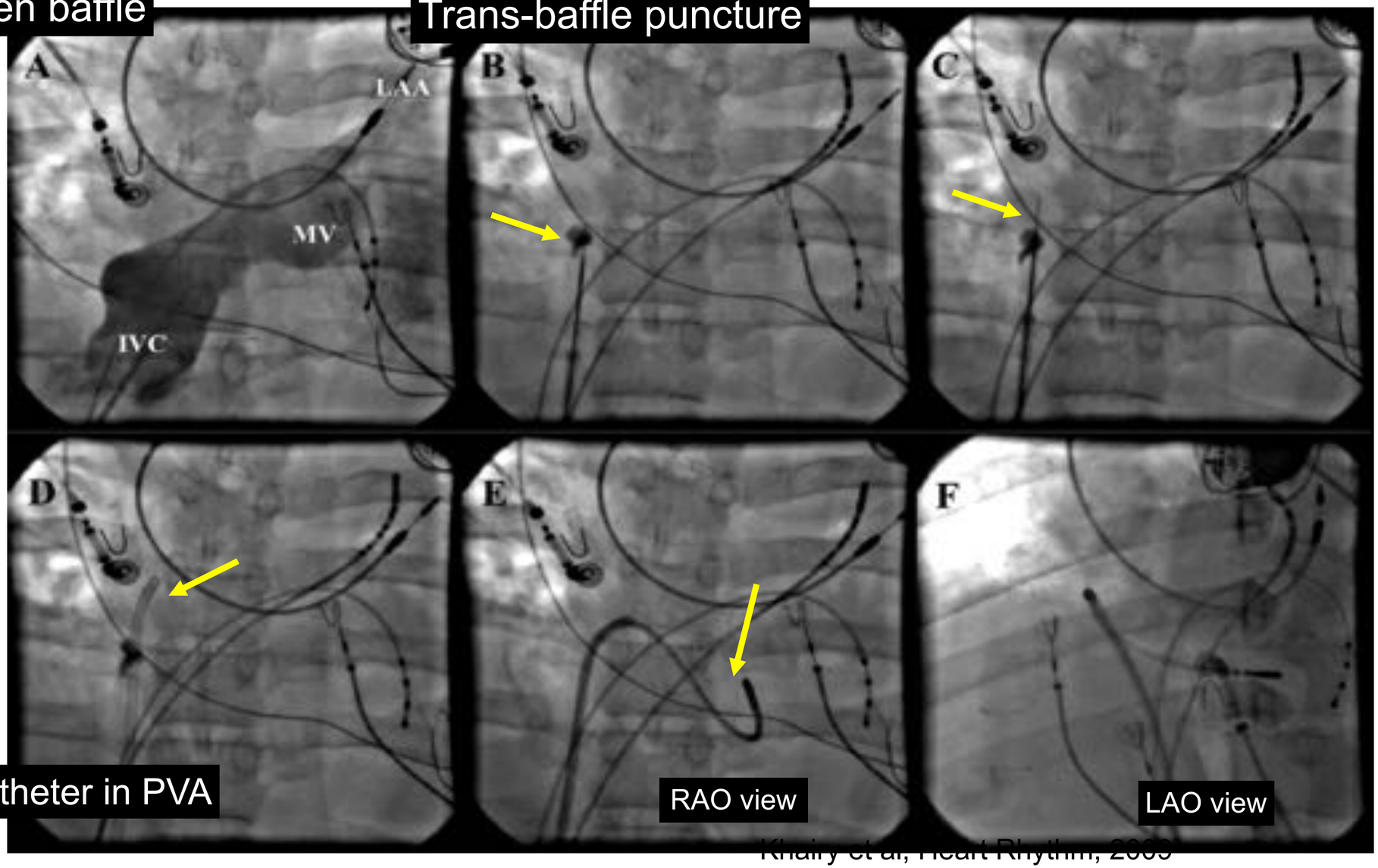
in a smooth-walled LV

Atrial flutter after atrial switch

Atrial arrhythmias may lead to hemodynamic instability and sudden death

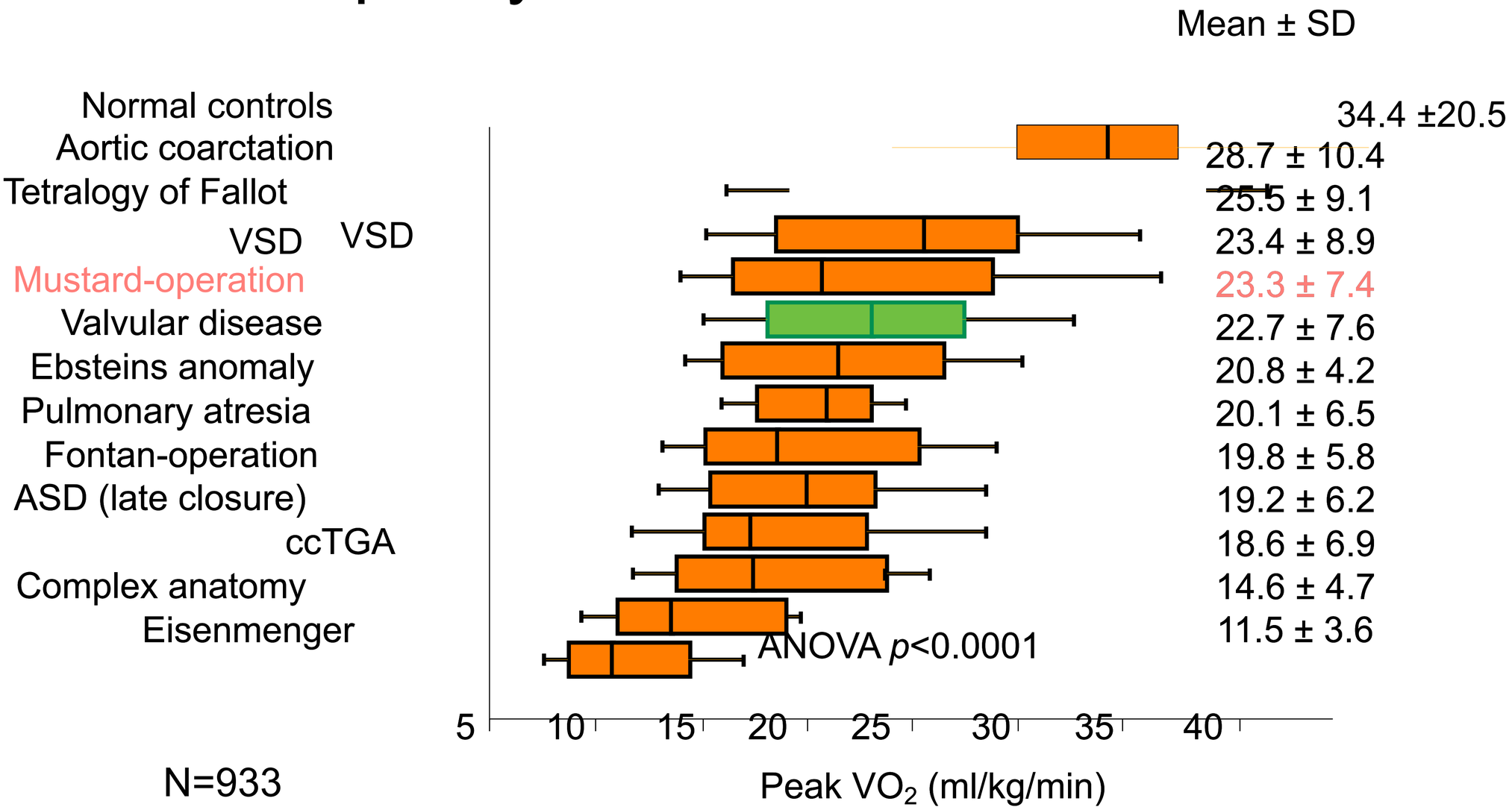


Transbaffle puncture in IART



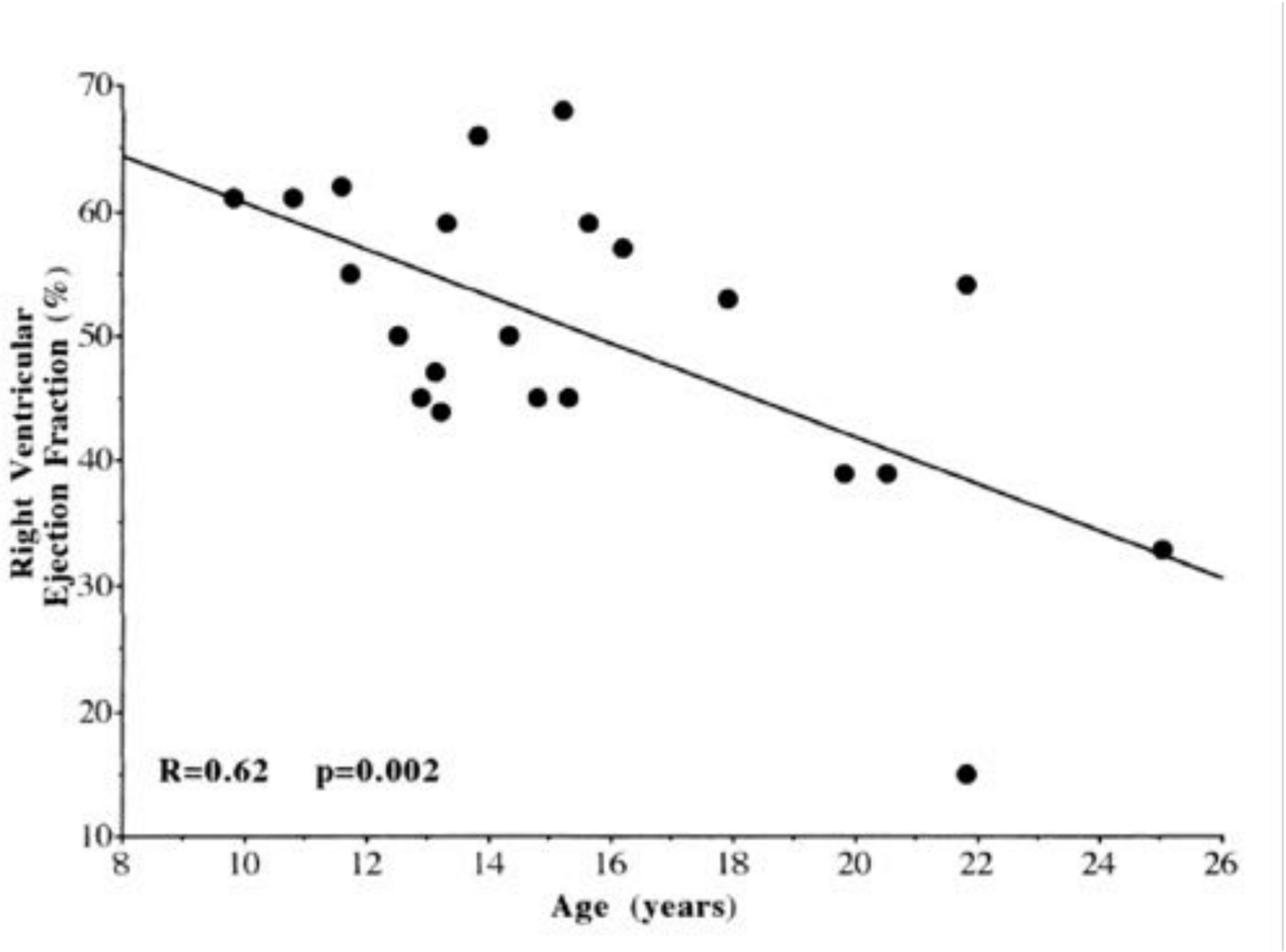
Khairy et al, Heart Rhythm, 2009

Exercise capacity in CHD



Diller GP, et al. Circulation 2005;

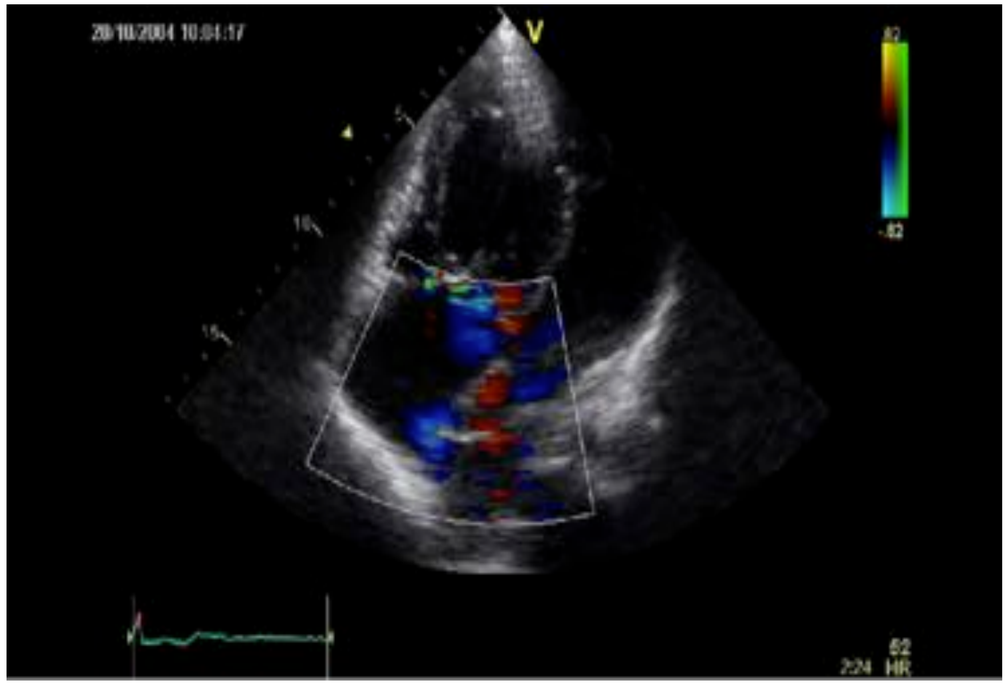
Decline of RV function after atrial switch



Milane et al. , JACC 2000.

Late complications atrial switch

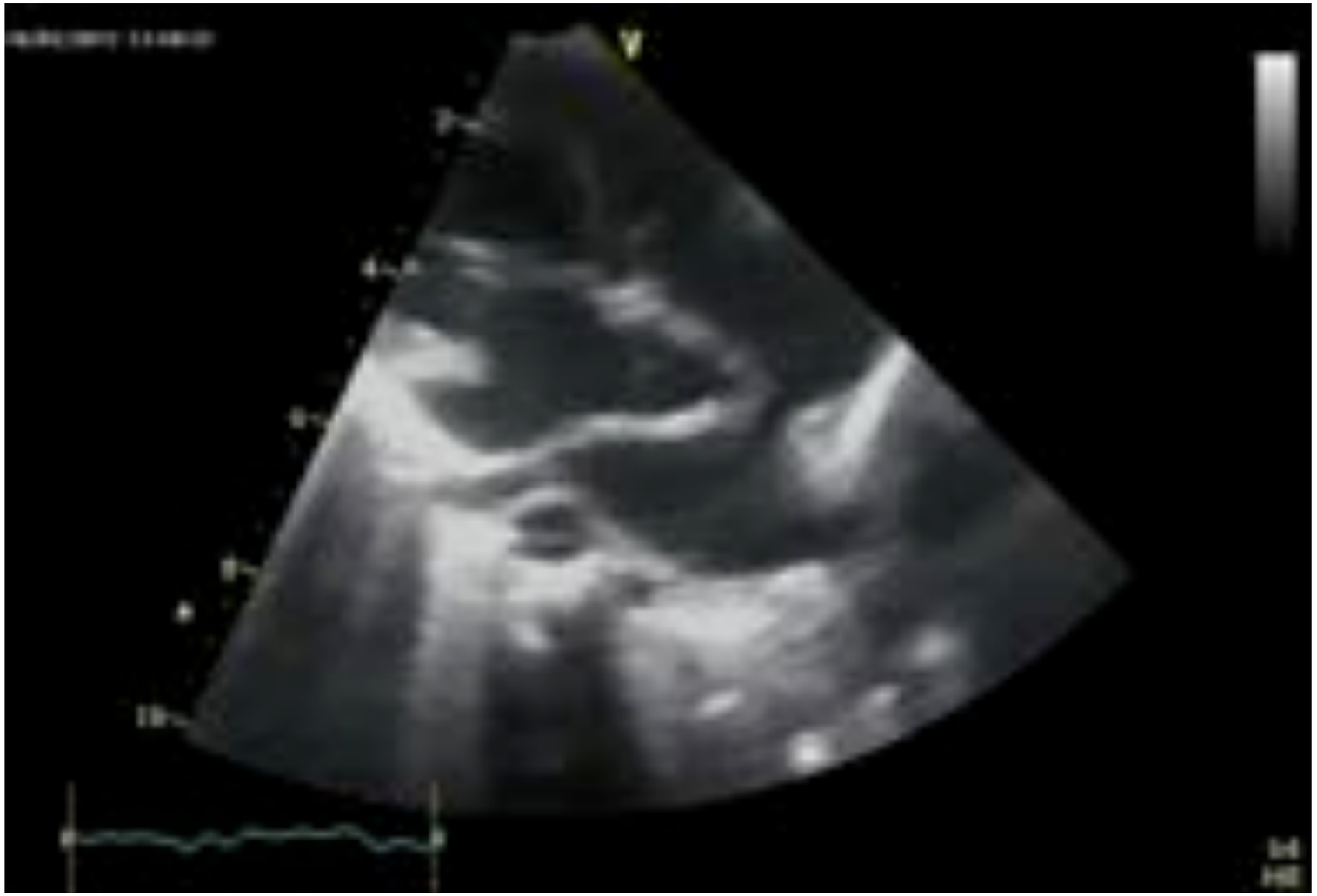
- Early death
- Arrhythmias
- Exercise capacity
- RV dysfunction ⬇
- Tricuspid regurgitation



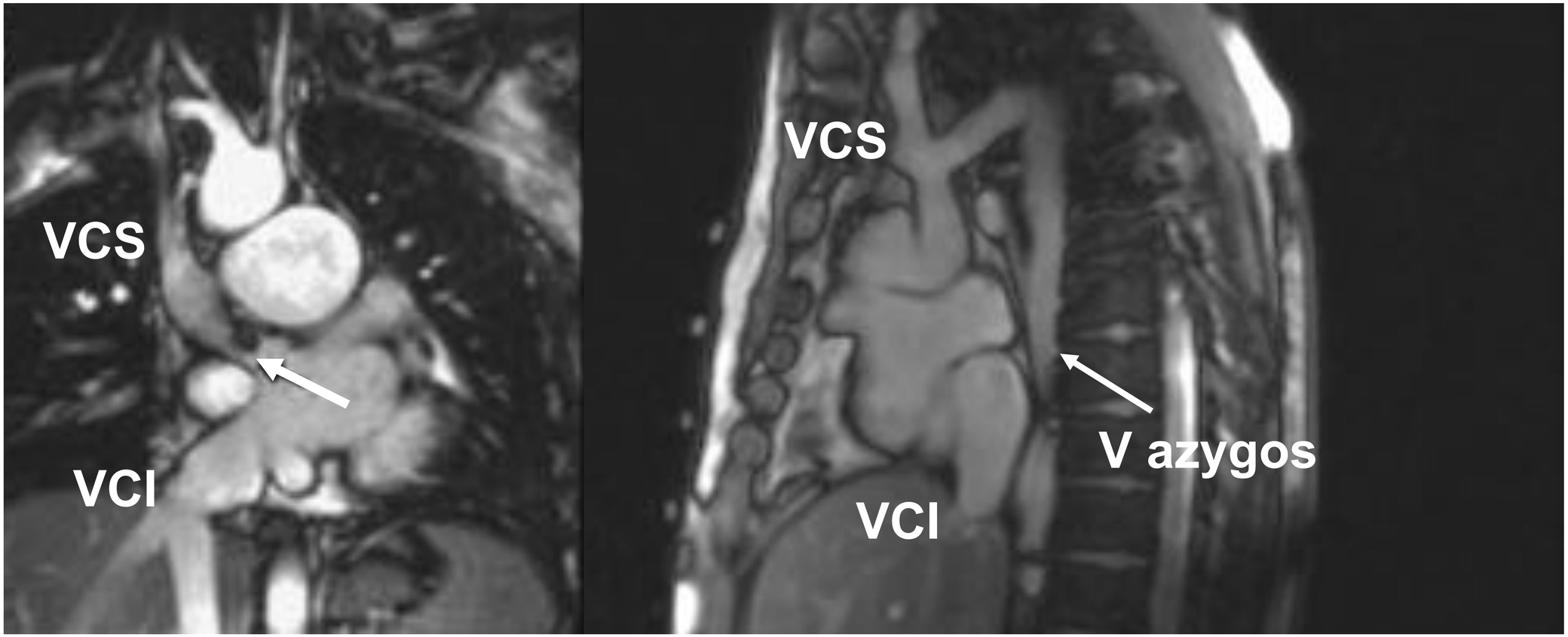
Abnormal septal configuration
RV dilation

Late complications atrial switch

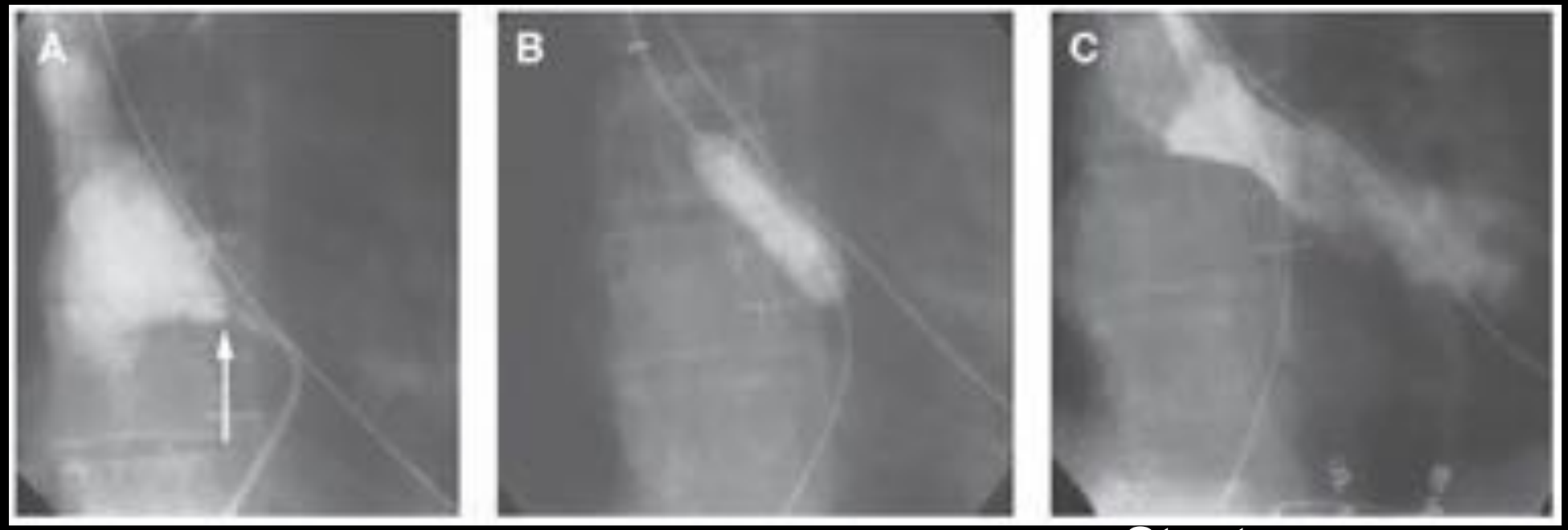
- Early death
- Arrhythmias
- Exercise capacity
- RV dysfunction
- Tricuspid regurgitation
- Baffle obstructions



Baffle obstruction with increased azygos flow

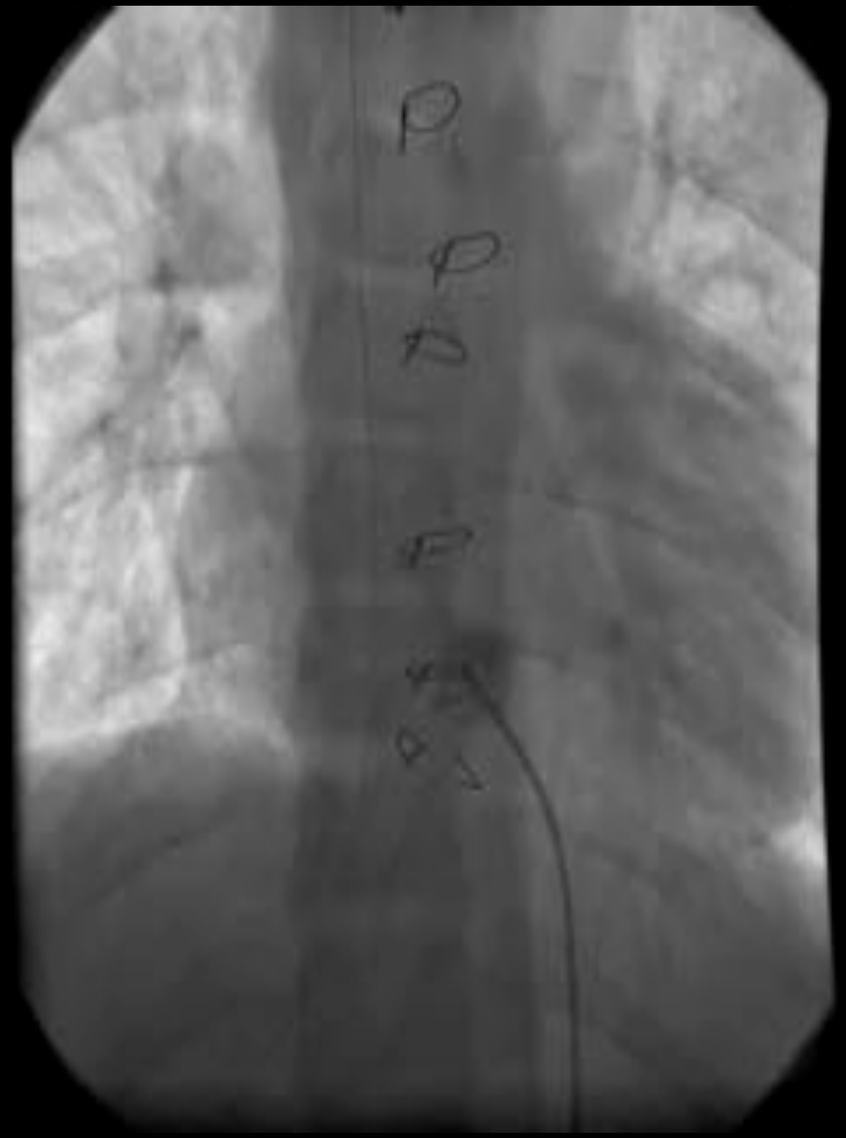


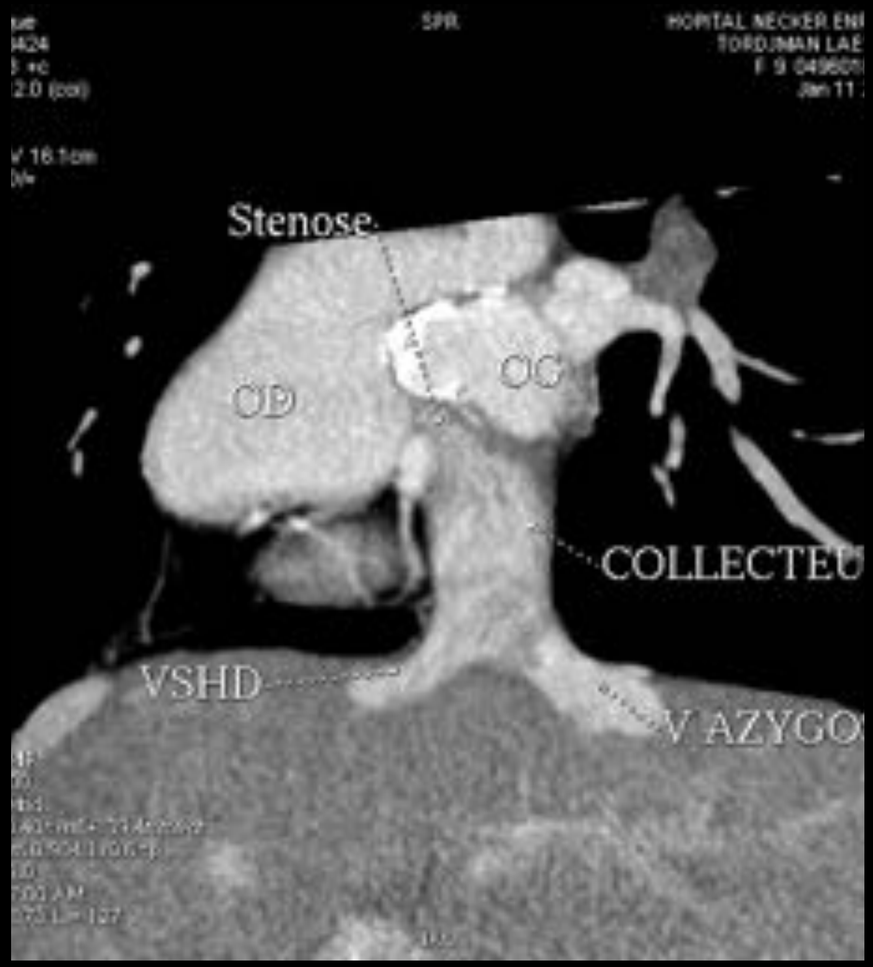
Superior baffle-limb stenosis in TGA after atrial switch and following PM implantation



obstruction Radiofrequency + Stent

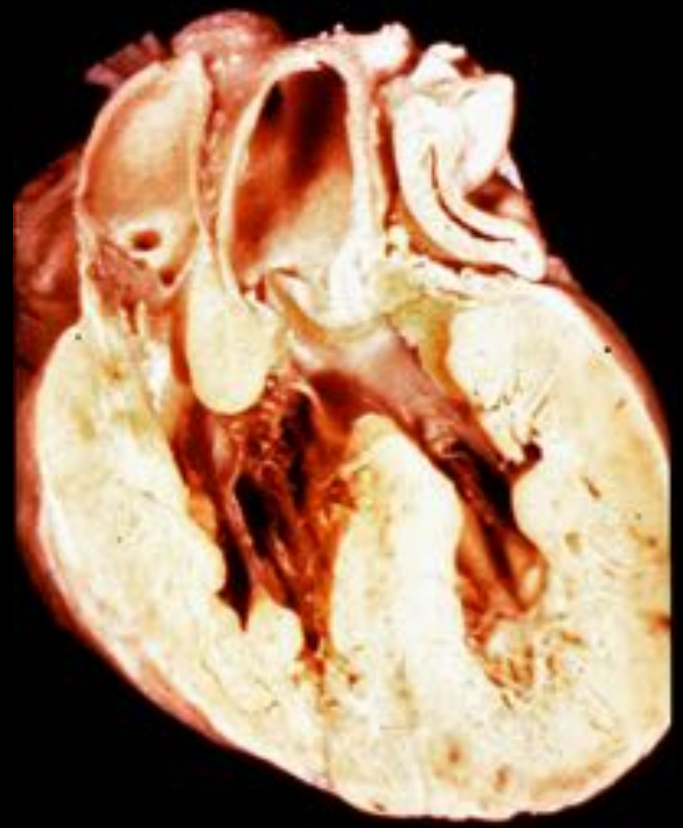
Love BA et al., Nat Clin Pract Cardiovasc Med 2008




SPR
HOPITAL NECKER ENI
TORDJMAN LAE
Jan 11
Stenose
OD
OG
COLLECTEU
VSHD
V AZYGO

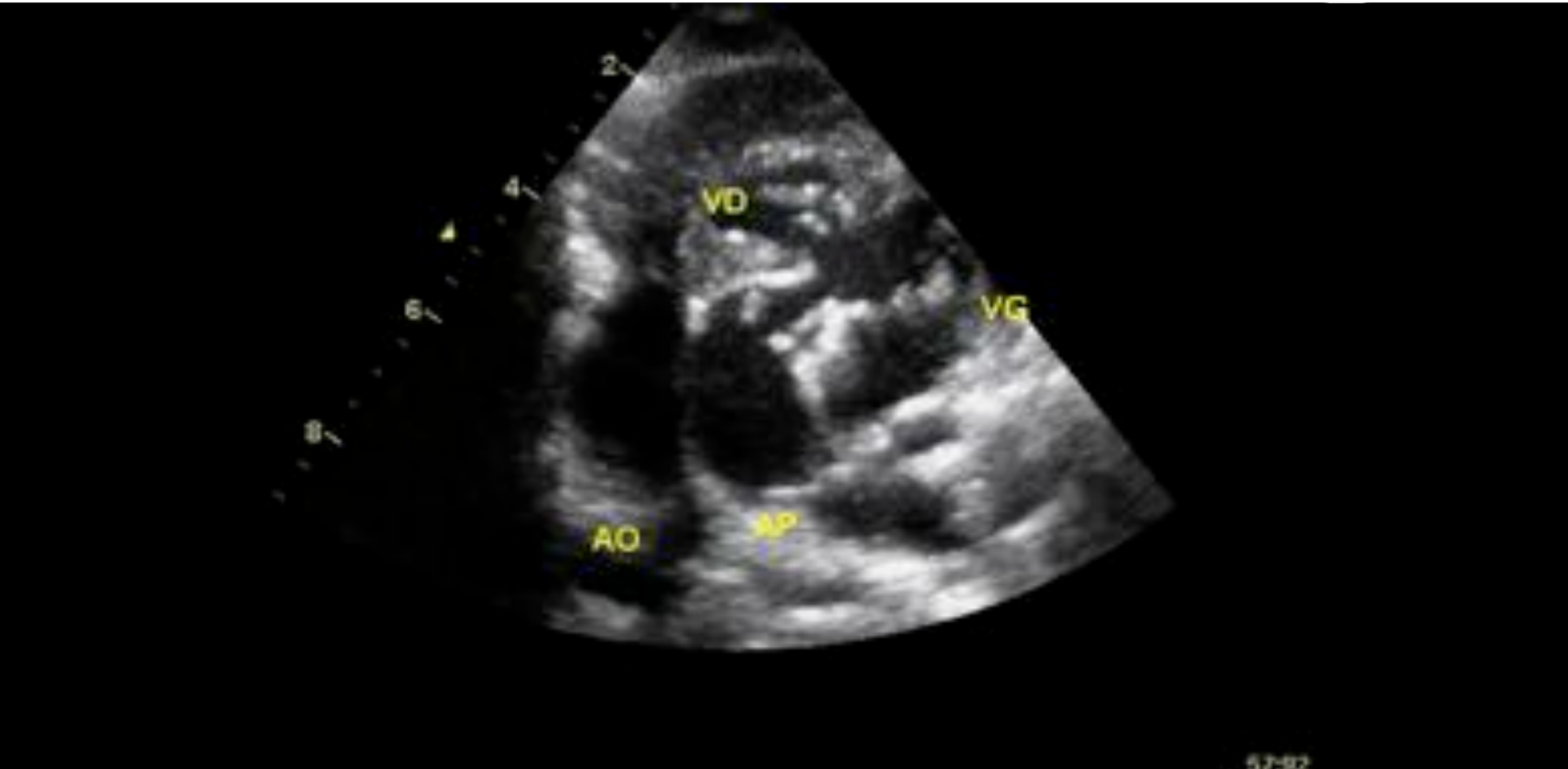
TGA

TGA - VSD - Pulmonary stenosis



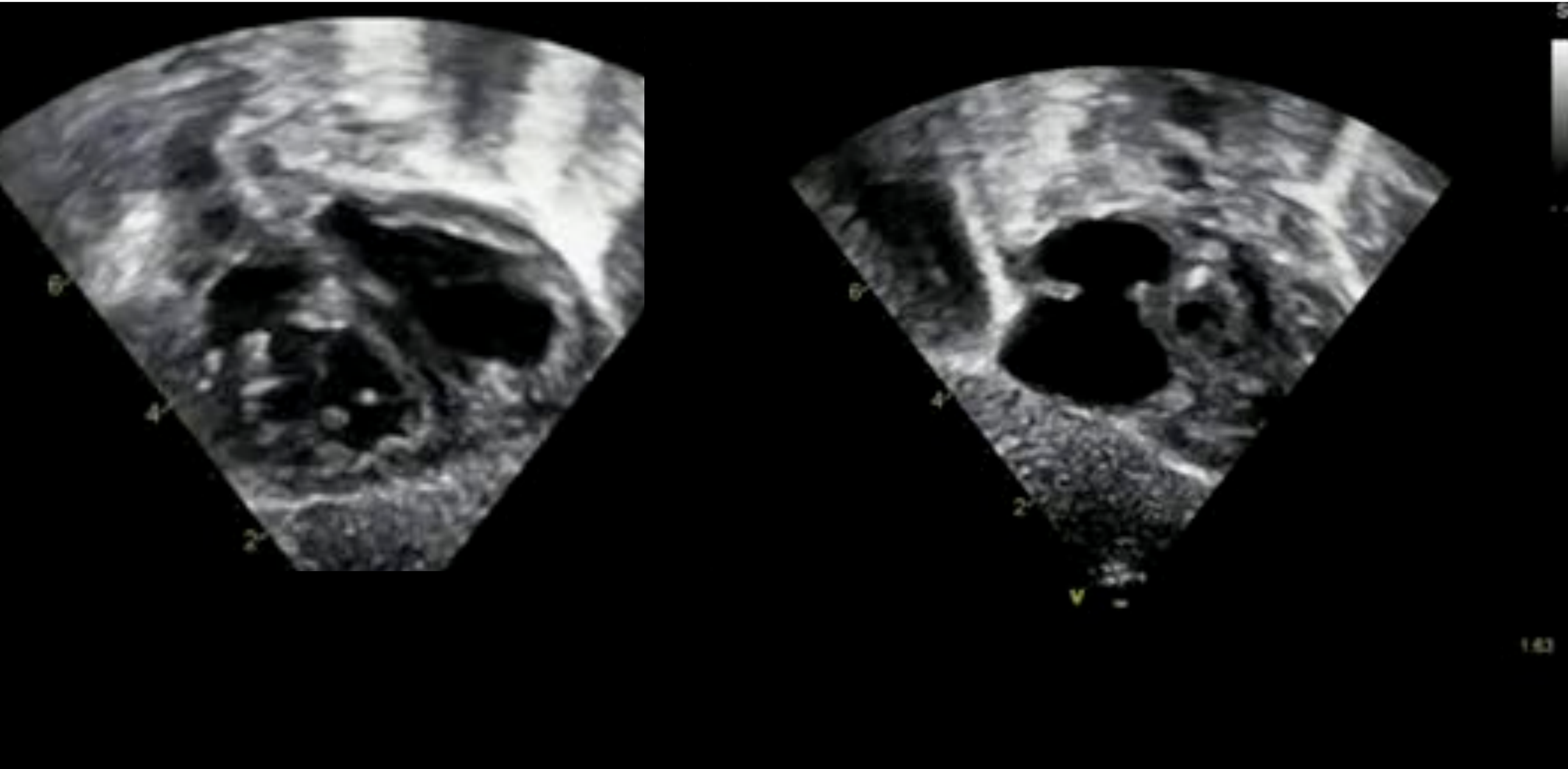
TGA

TGA - VSD - Pulmonary stenosis



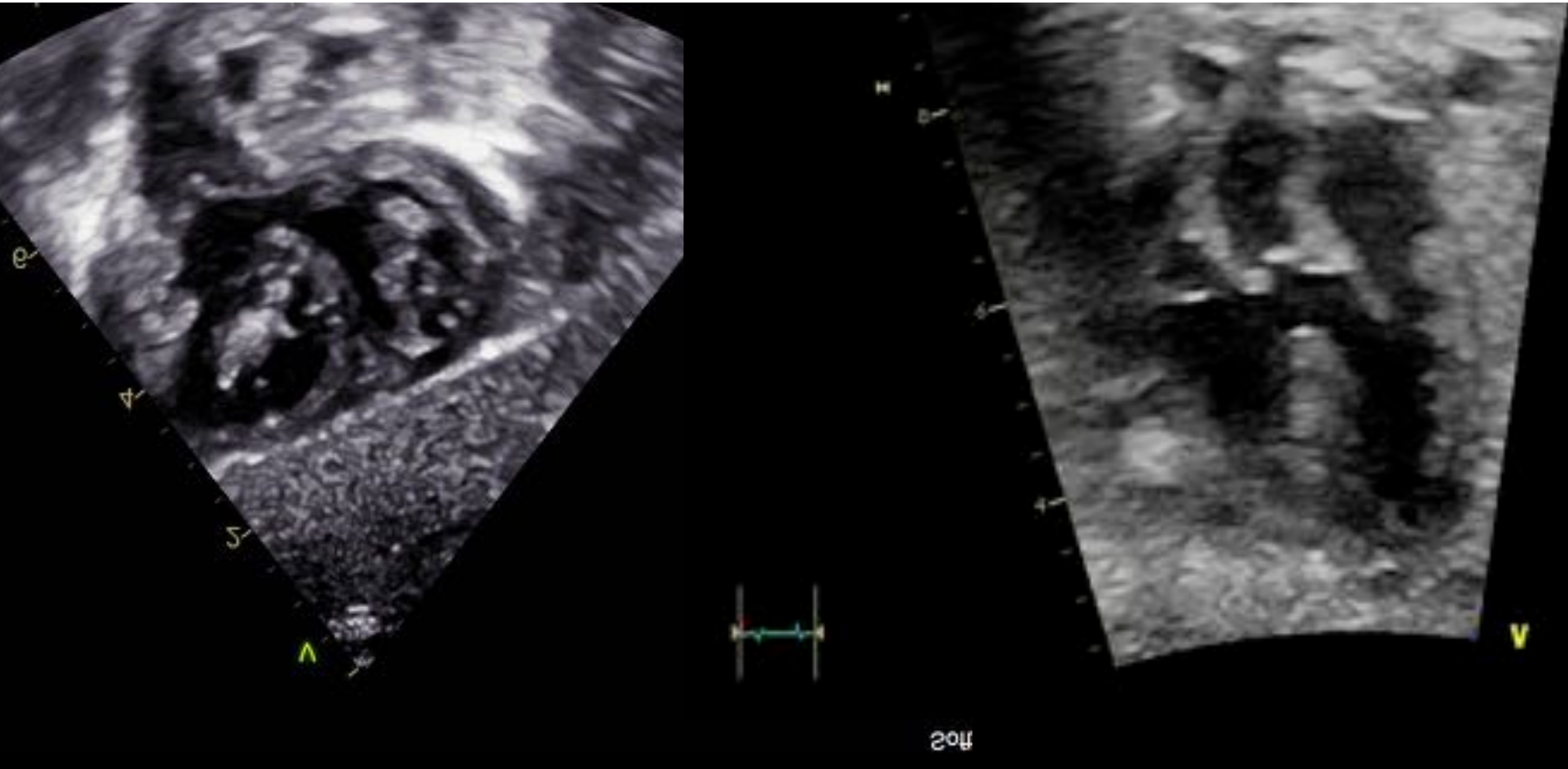
TGA

TGA - VSD - Pulmonary stenosis

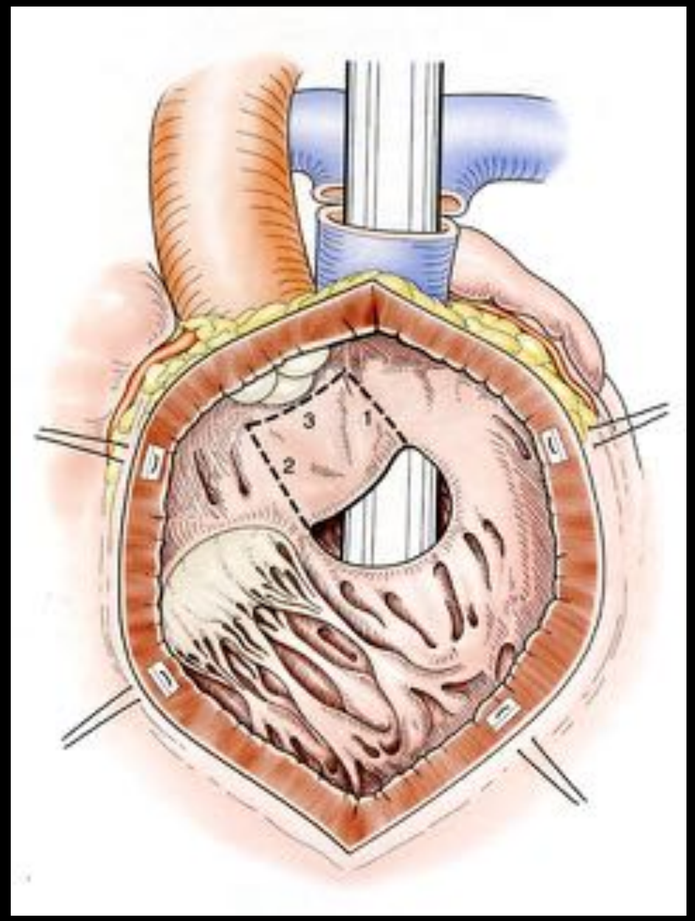


TGA

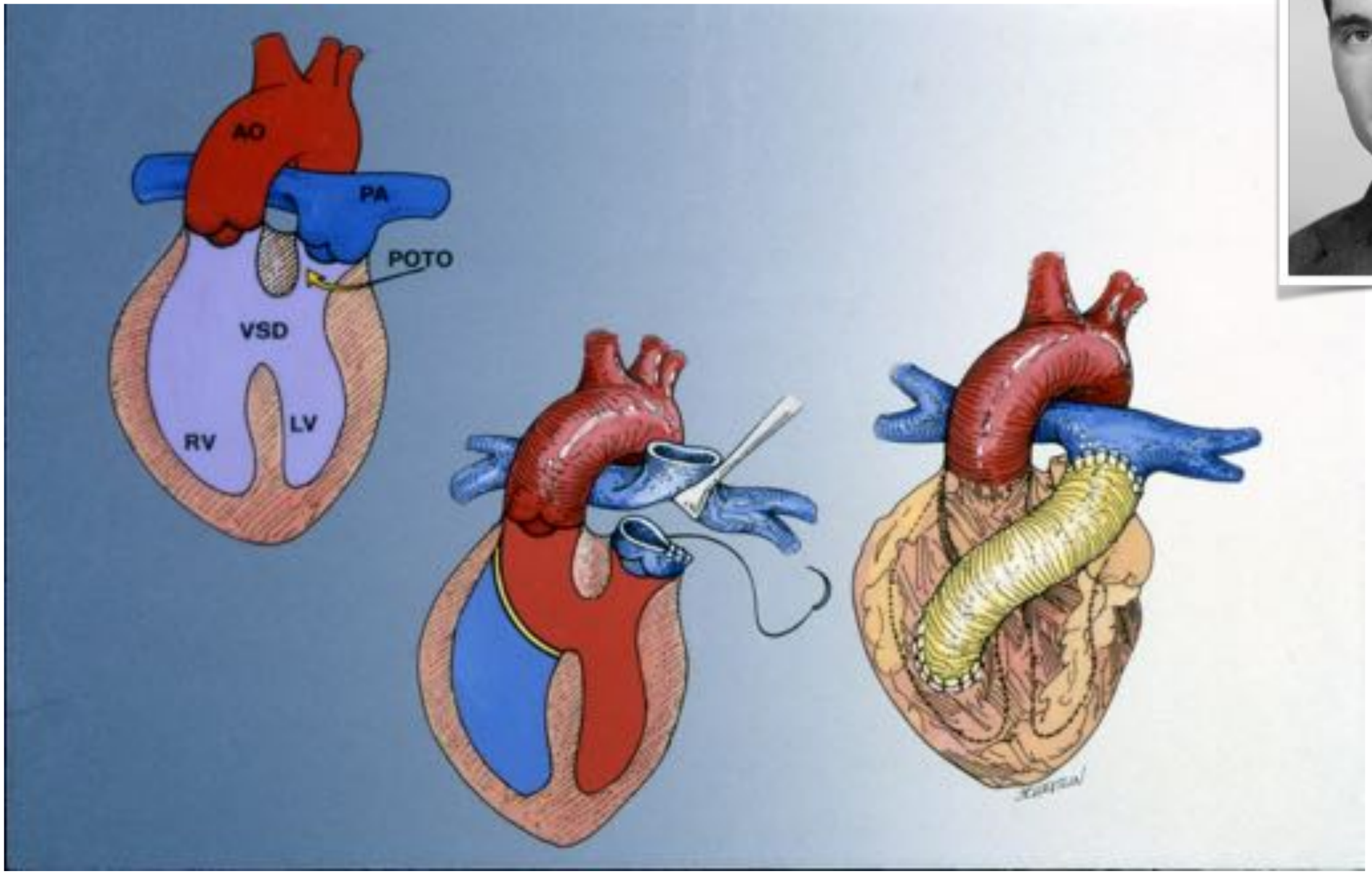
TGA - VSD - Pulmonary stenosis



The Rastelli operation

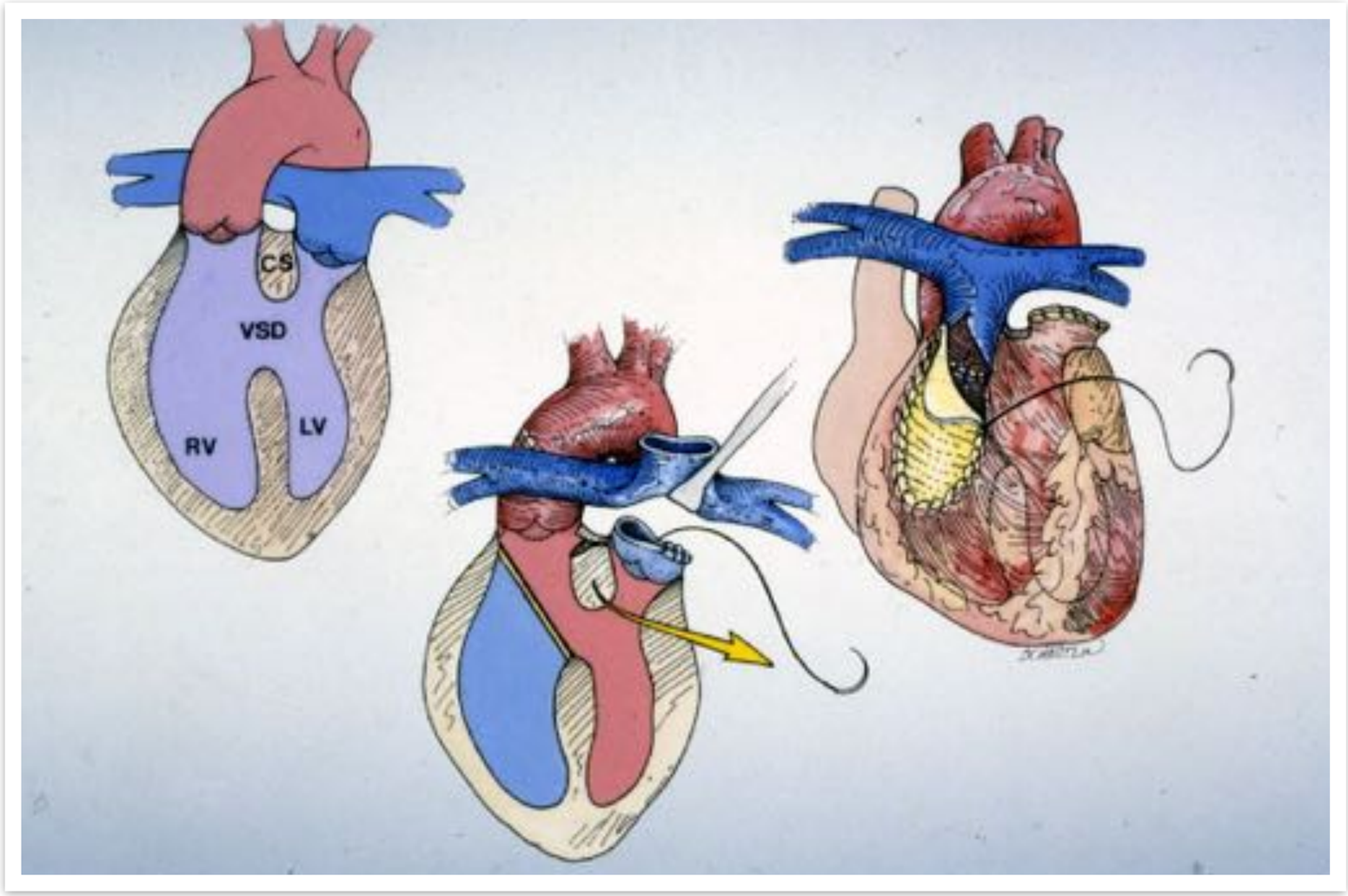


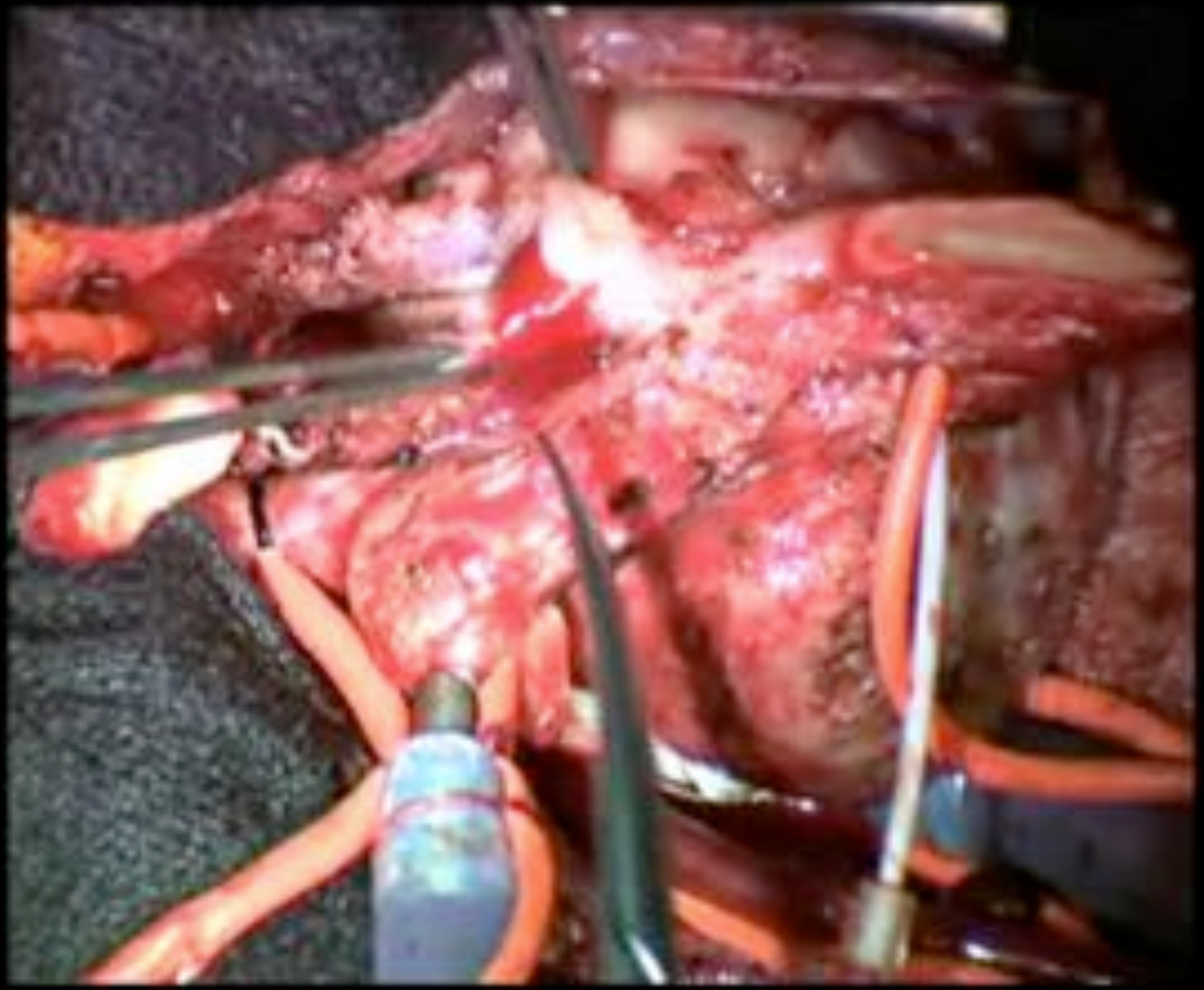
Rastelli procedure



CT after Rastelli operation

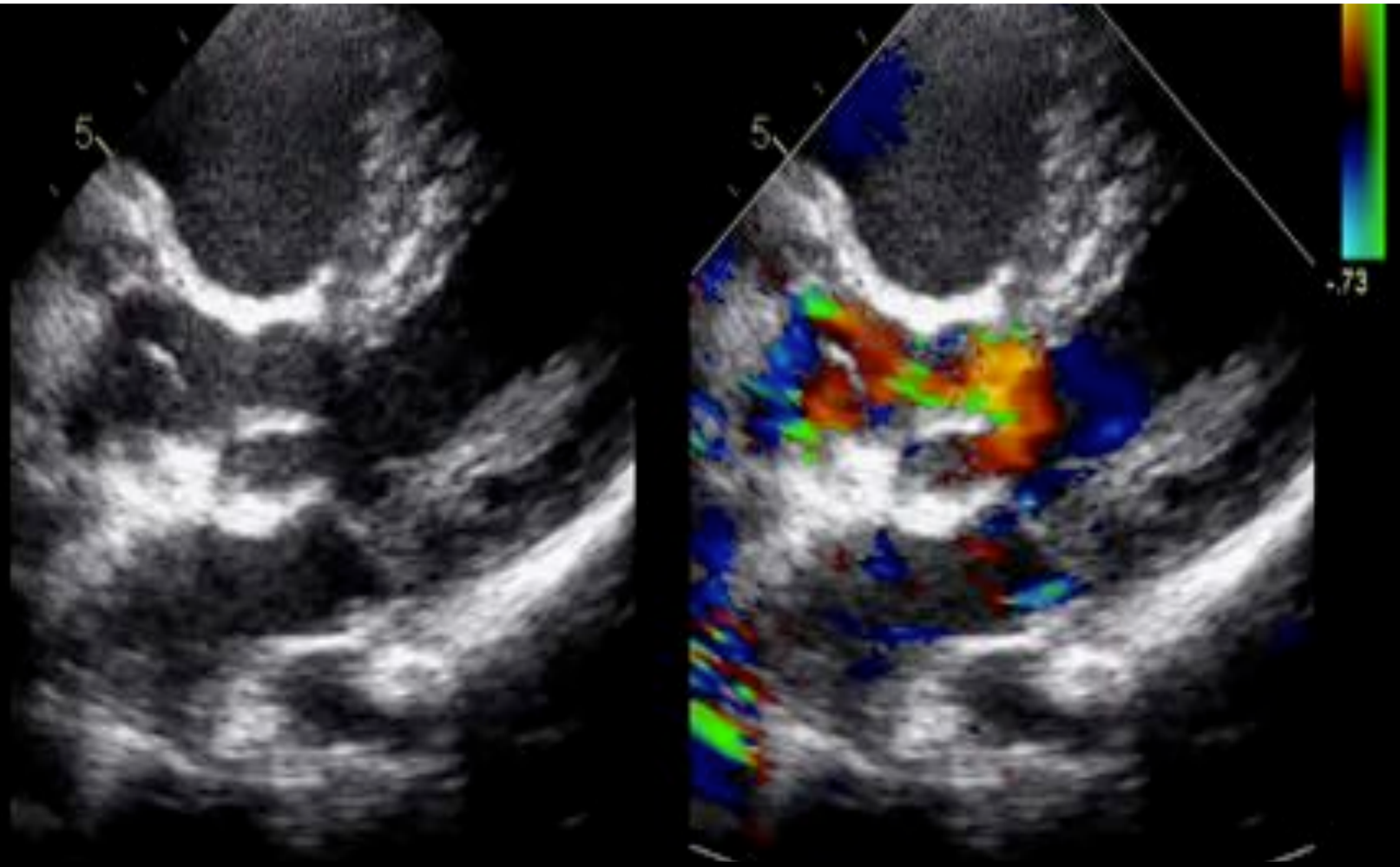
REV operation
(Réparation à l'Etage Ventriculaire)

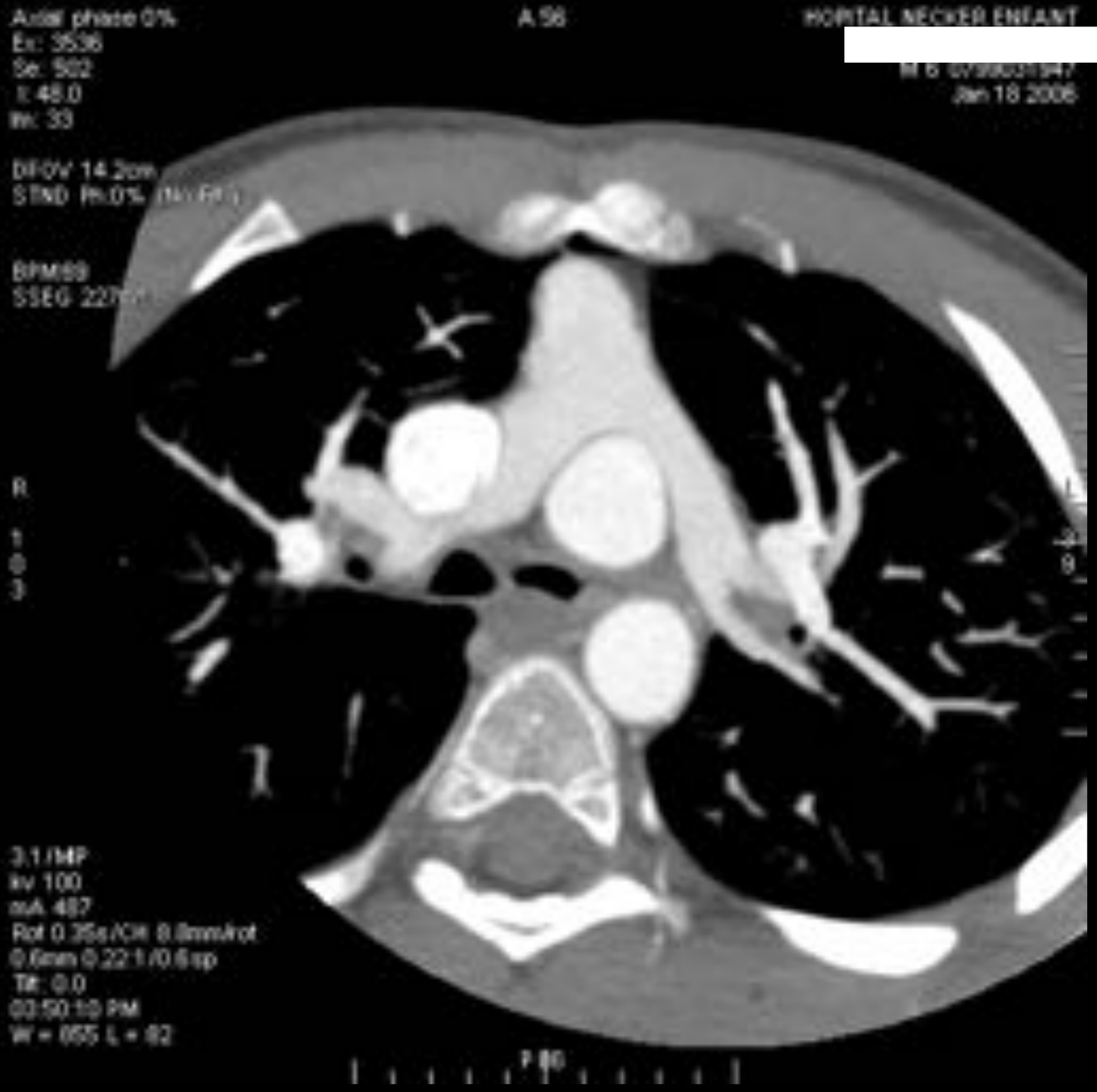




TGA

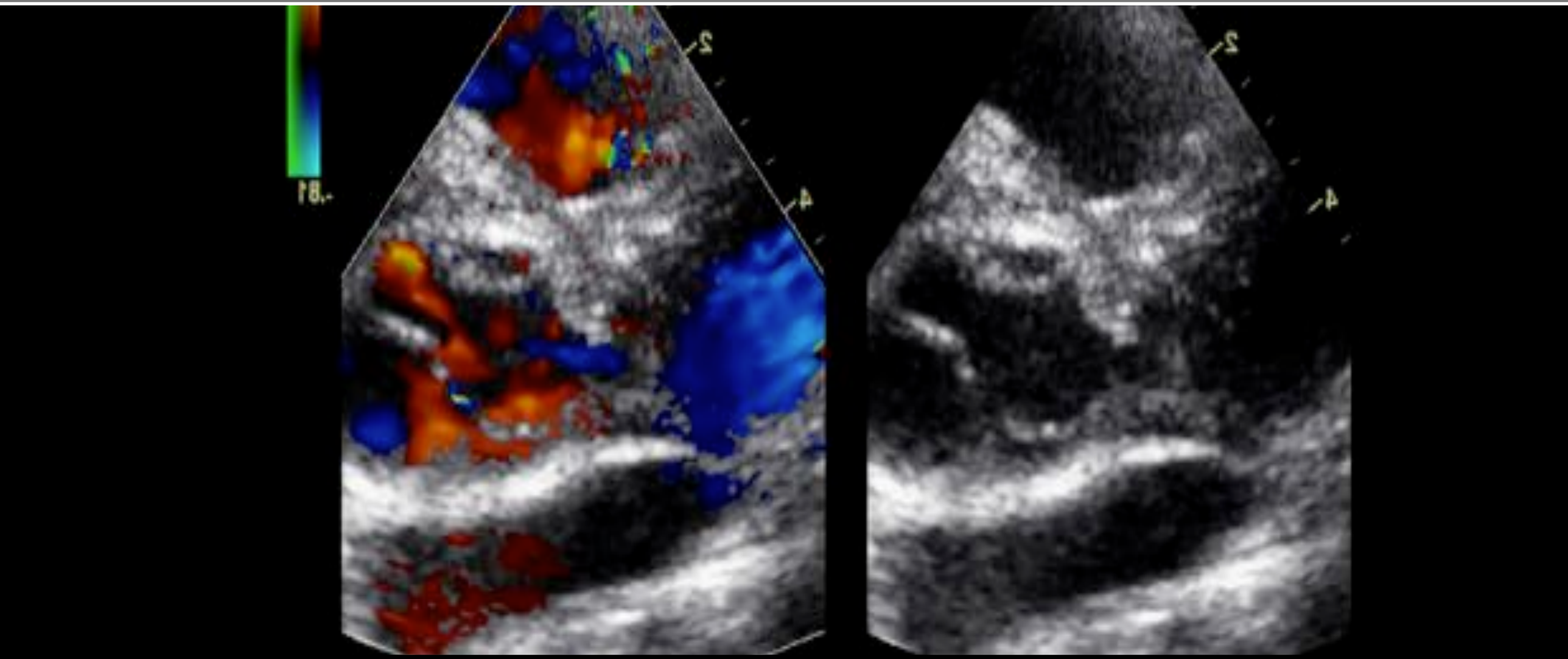
TGA - VSD - Pulmonary stenosis: tunnel from LV to aorta



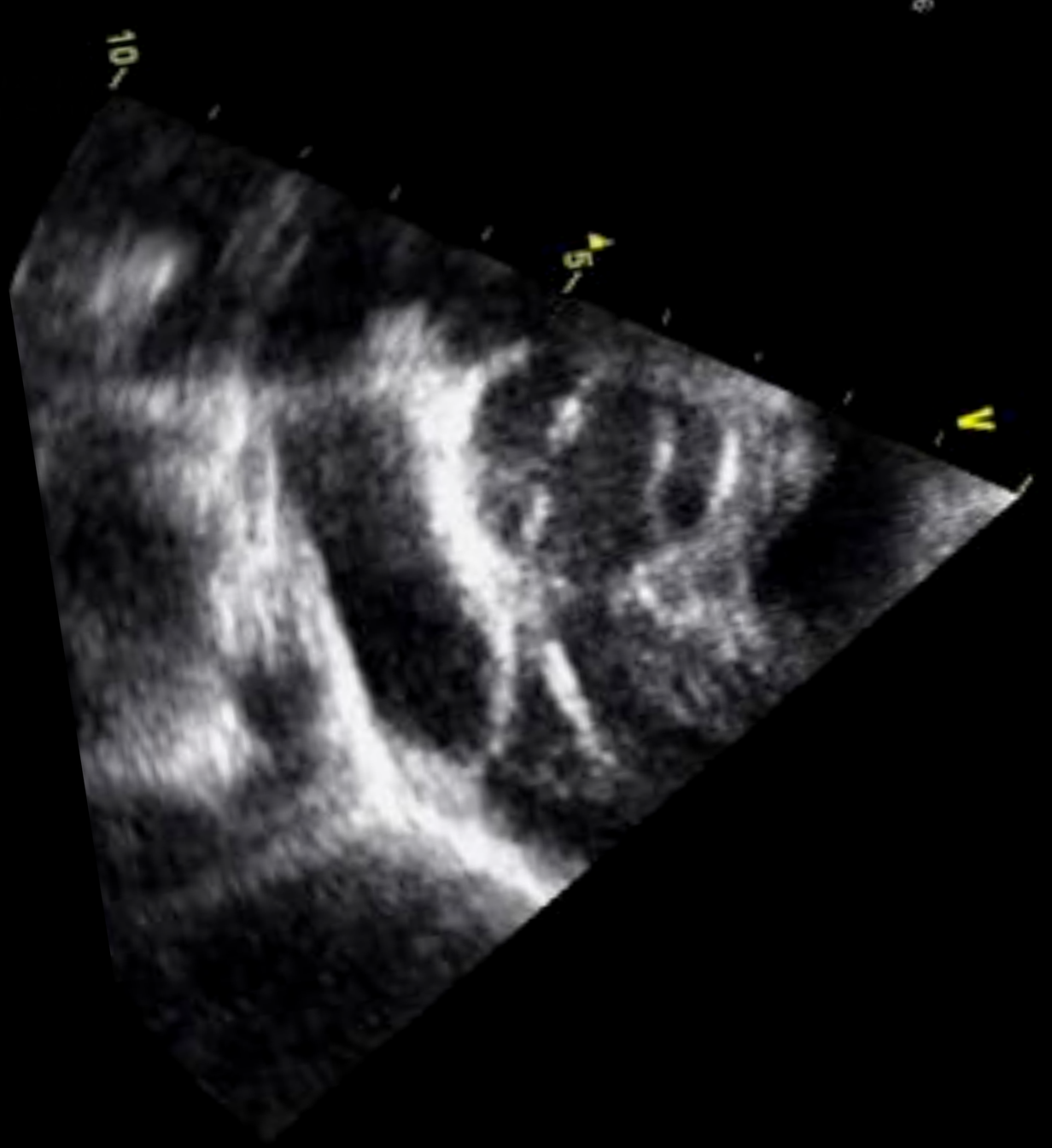

Axial phase 0%
Ex: 3536
Se: 502
I: 48.0
Im: 33
DFOV 14.2cm
STND Rk:0%
BPM:69
SSEG 227
R
1
0
3
3.1 MP
kv 100
mA 487
Rot 0.35s/CW 8.0mm/rot
0.6mm 0.22:1/0.6sp
Tilt: 0.0
03:50:10 PM
W = 655 L = 82
A 56
HOPITAL NECKER ENFANT
Jan 18 2006

TGA

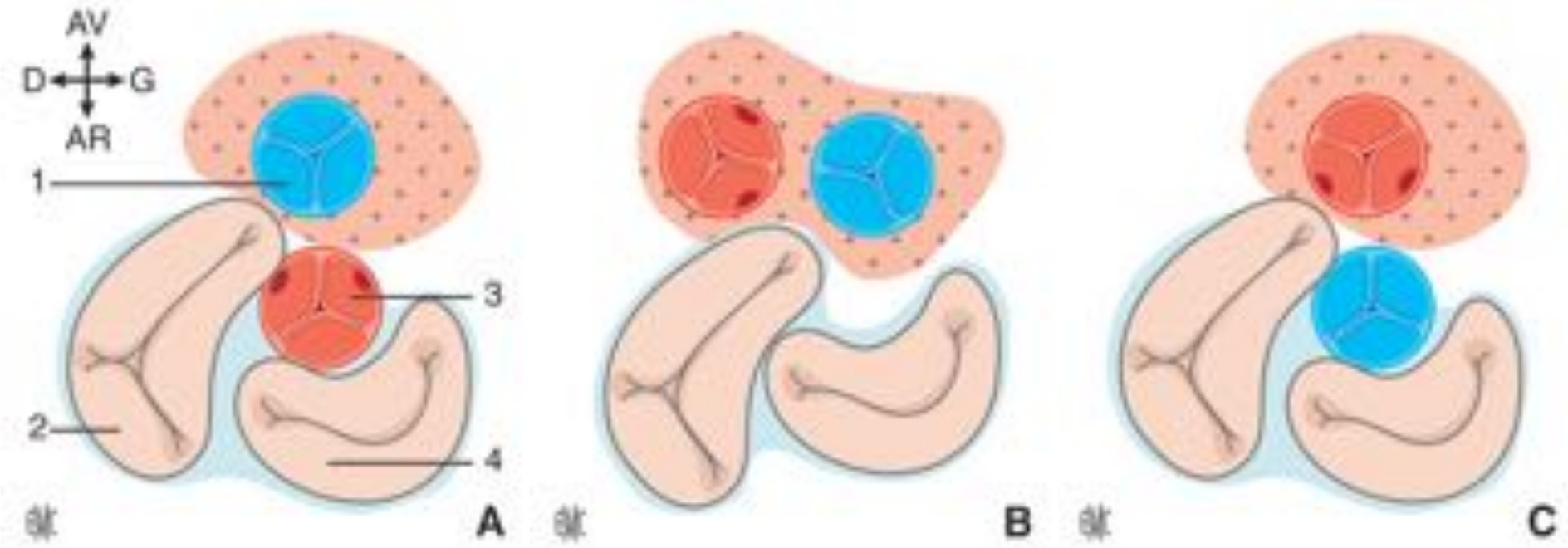
Sub-aortic obstruction after tunnel from LV to aorta



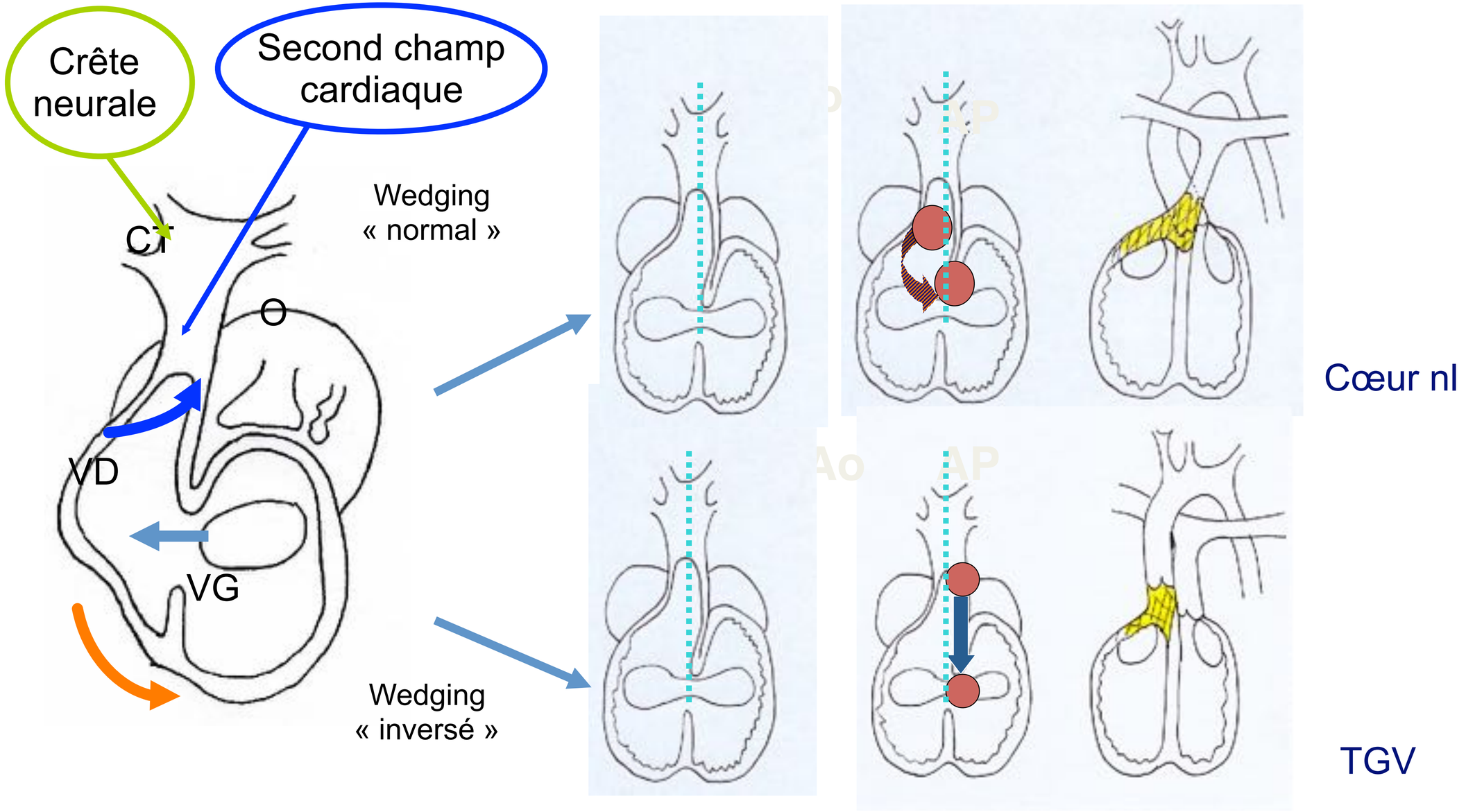
Rastelli/REV procedure
Sub aortic stenosis



Double outlet right ventricle



normal malpositions transposition



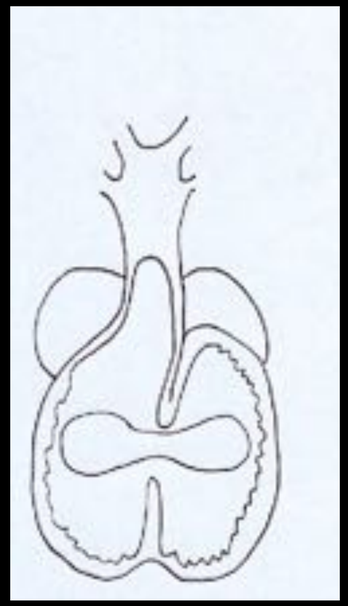
Early looping Convergence Wedging

3 groups of DORV (van Praagh)

- Groupe 1 : DORV with isolated anomalies of the outflow tracts
  « Late » DORV due to insufficient wedging
- Groupe 2 : DORV with outflow tracts anomalies + ventricles + AV valves
  « Early » DORV during « early looping »
- Groupe 3 : Looping anomalies
  DORV associated with heterotaxy

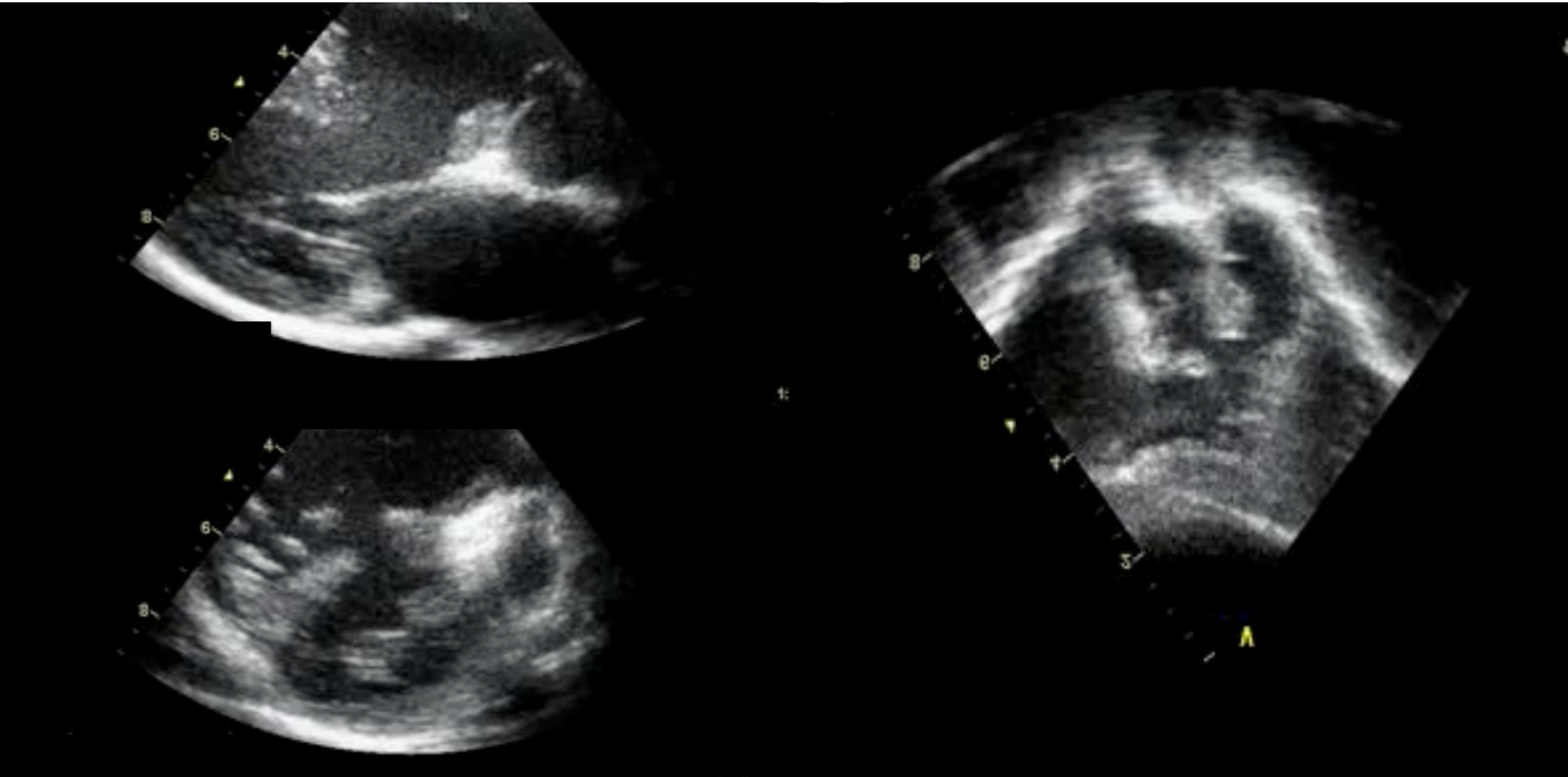
DORV

« Early » DORV

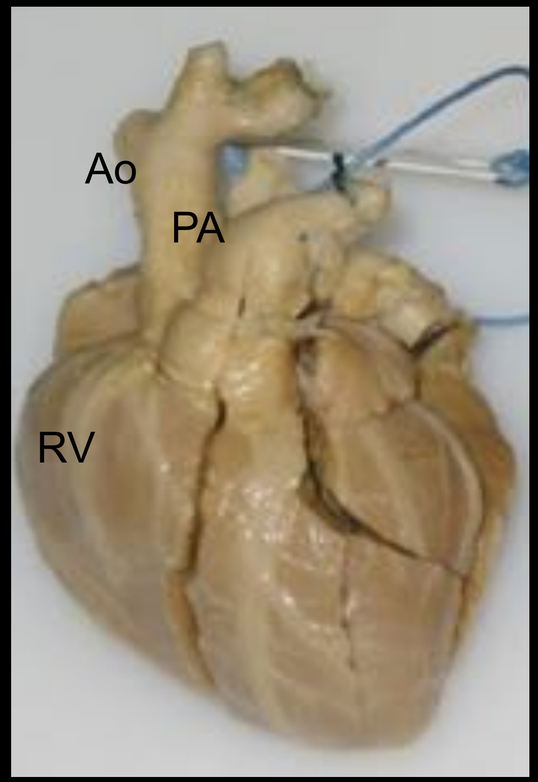


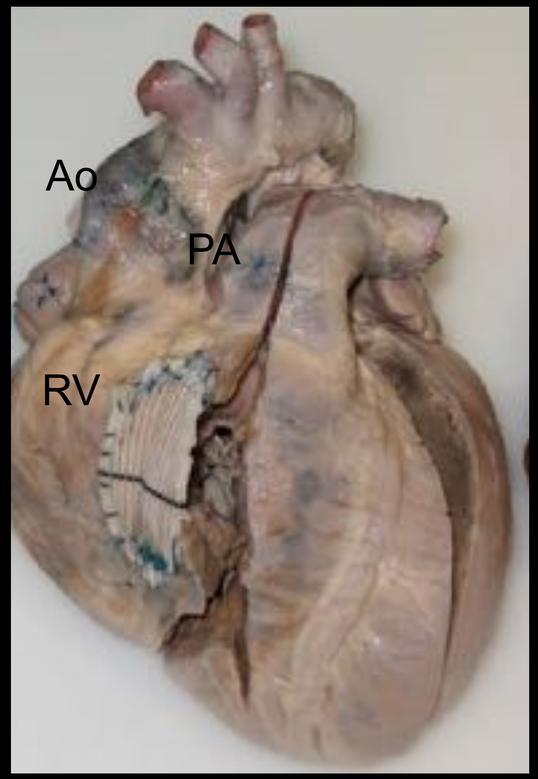
DORV

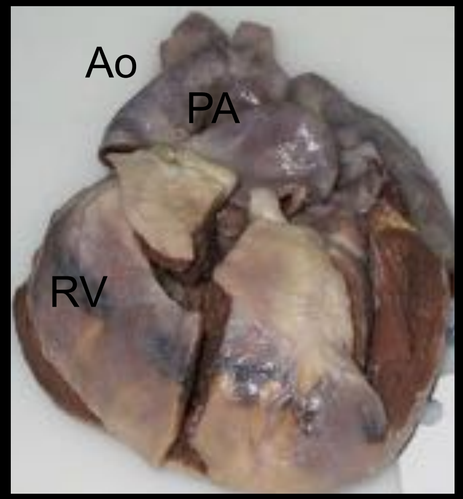
« Late » DORV

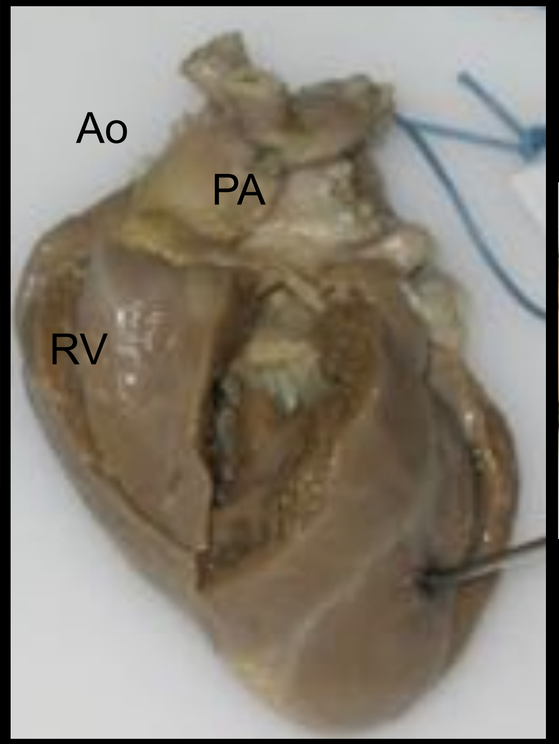


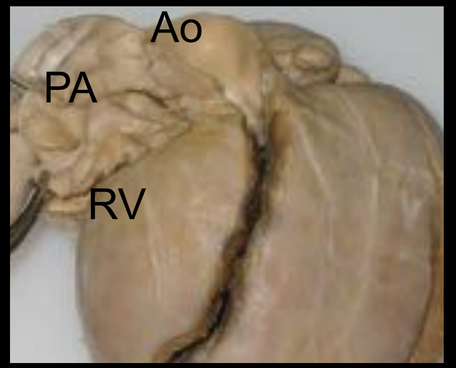
Relative position of the great vessels in DORV

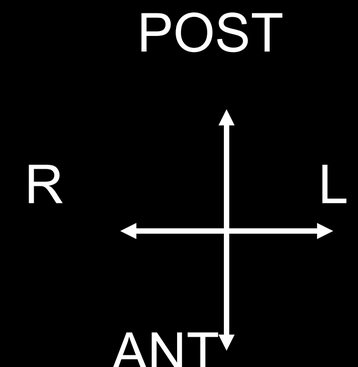




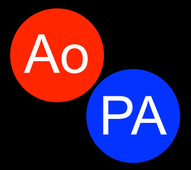


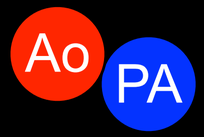


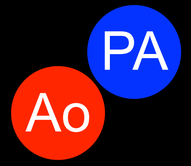


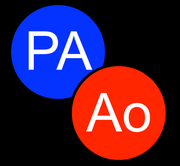


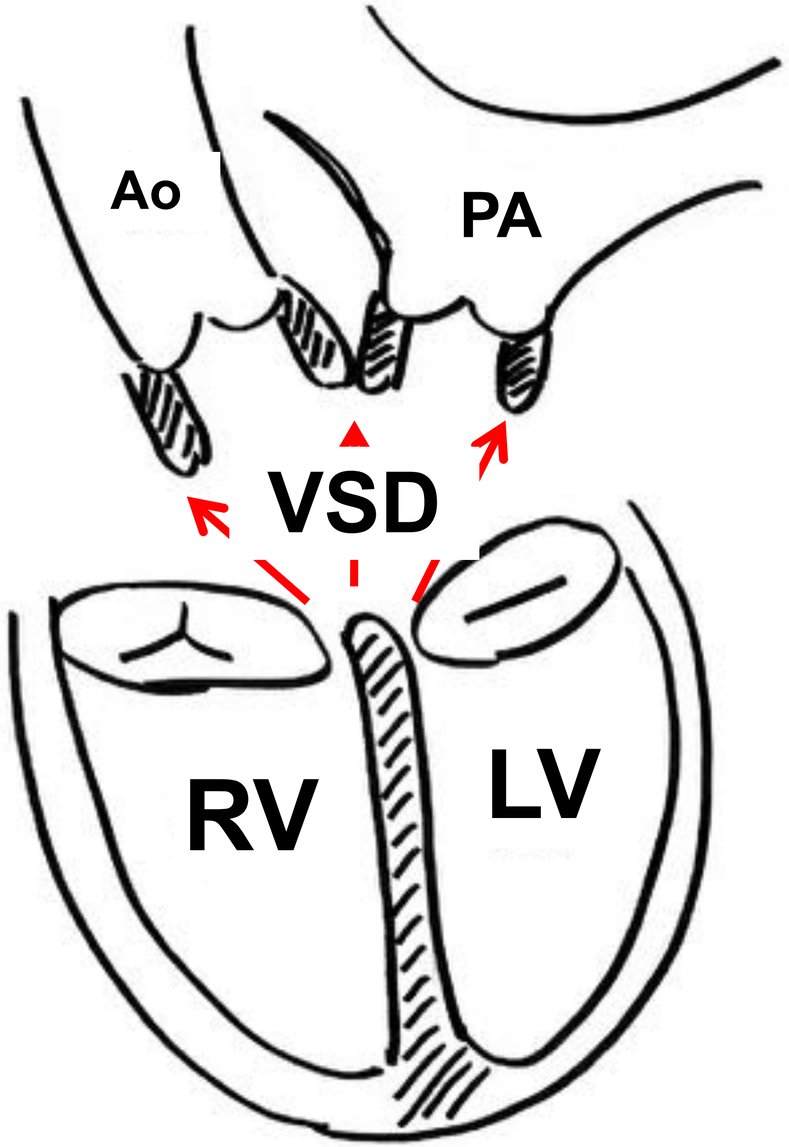
S,D,L



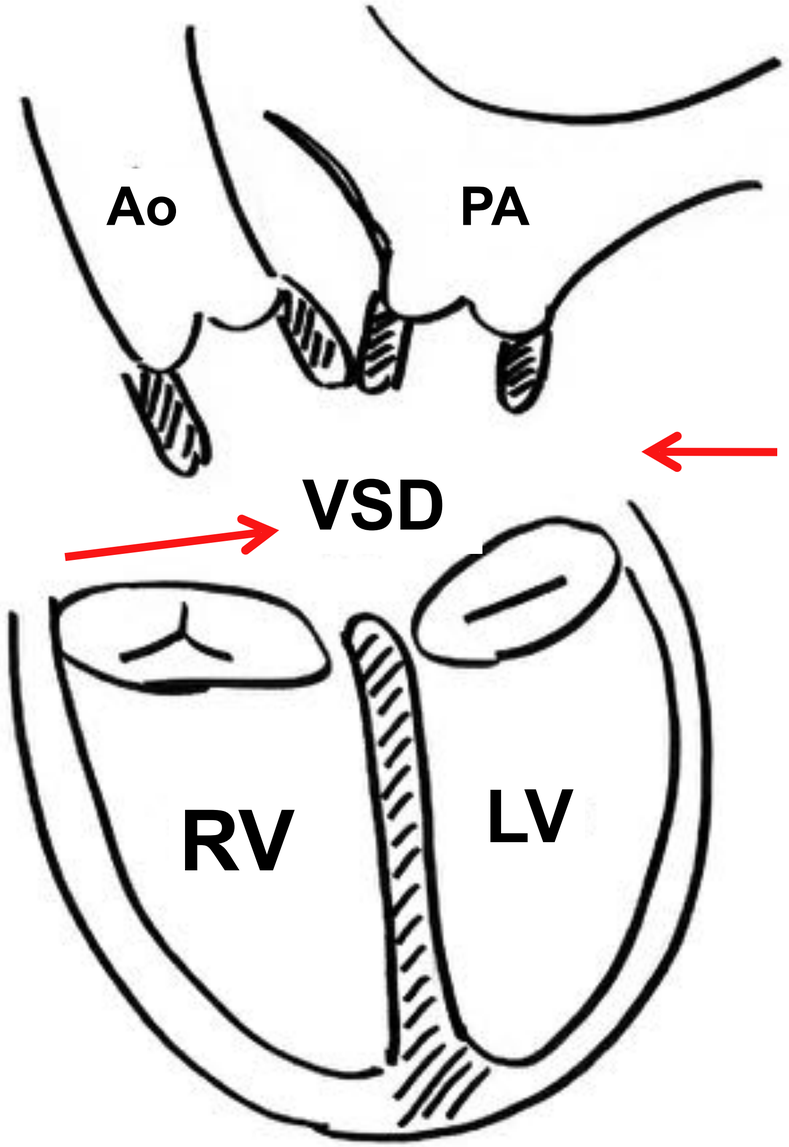




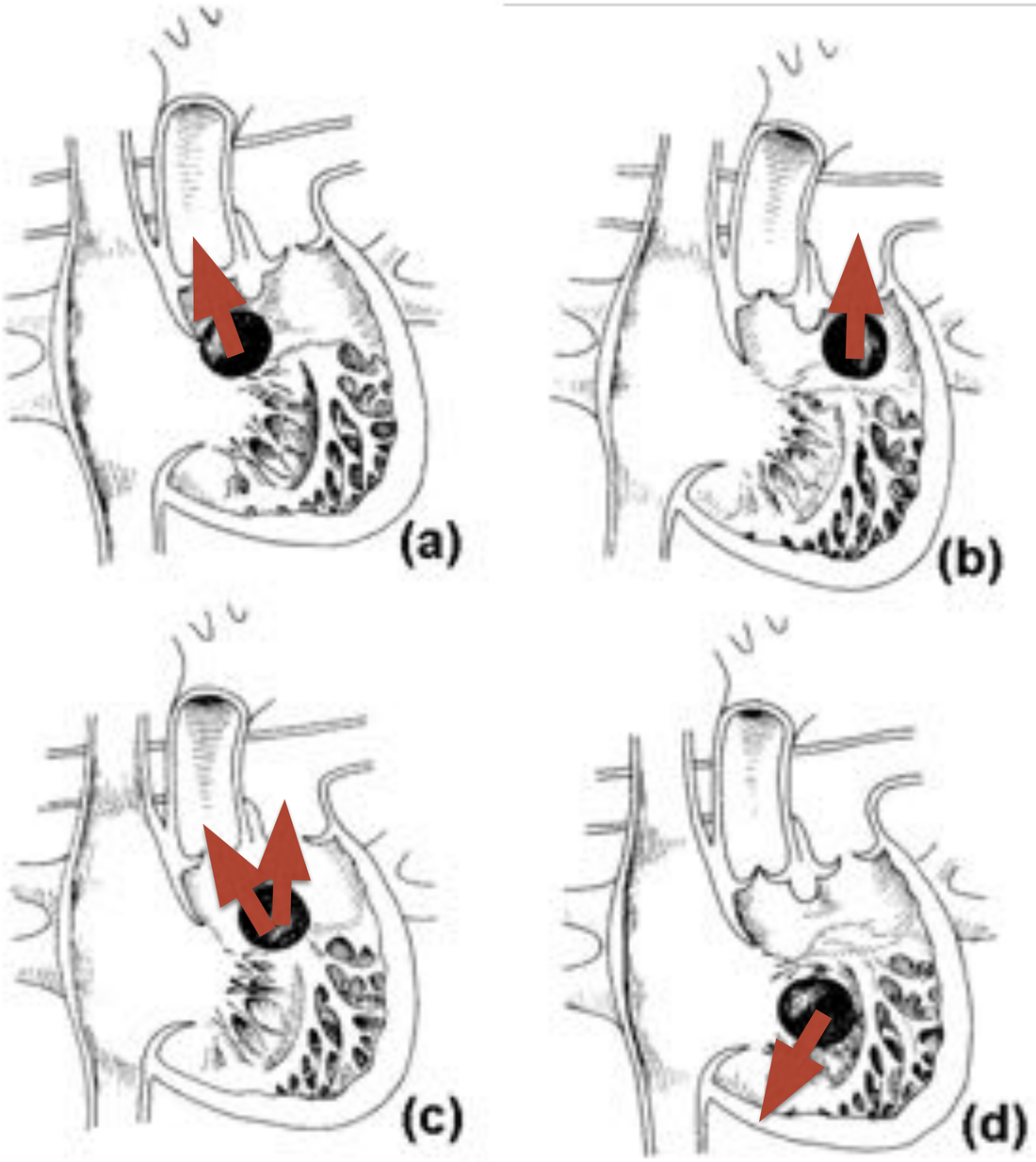



Ao
PA
VSD
RV
LV

VSD is (nearly) always cono-ventricular



Subarterial conus are extremely variable


(a)
(b)
(c)
(d)

For the surgeon : 3 simple questions

1. is biventricular repair possible ?

   if « YES »

2. is "anatomic" repair feasible ?

   if « NO »

3. which extra-anatomic repair is indicated ?

1. is biventricular repair possible ?

- Problems related to ventricles and AV valves
  - – size and function of ventricles
  - – anatomy of A-V valves
    - abnormal insertions on conal septum
    - straddling
    - malformation (stenosis/regurgitation)

- Problems related to VSD
  - – too large or multiple VSDs (swiss-cheese)
  - – too small and impossible to enlarge by resecting conal septum
  - – **non-conoventricular VSD (muscular)**

1. is biventricular repair possible ?

- biventricular repair is impossible
- biventricular repair is possible but hazardous
- univentricular pathway (Fontan) is indicated
  (< 20% of cases)

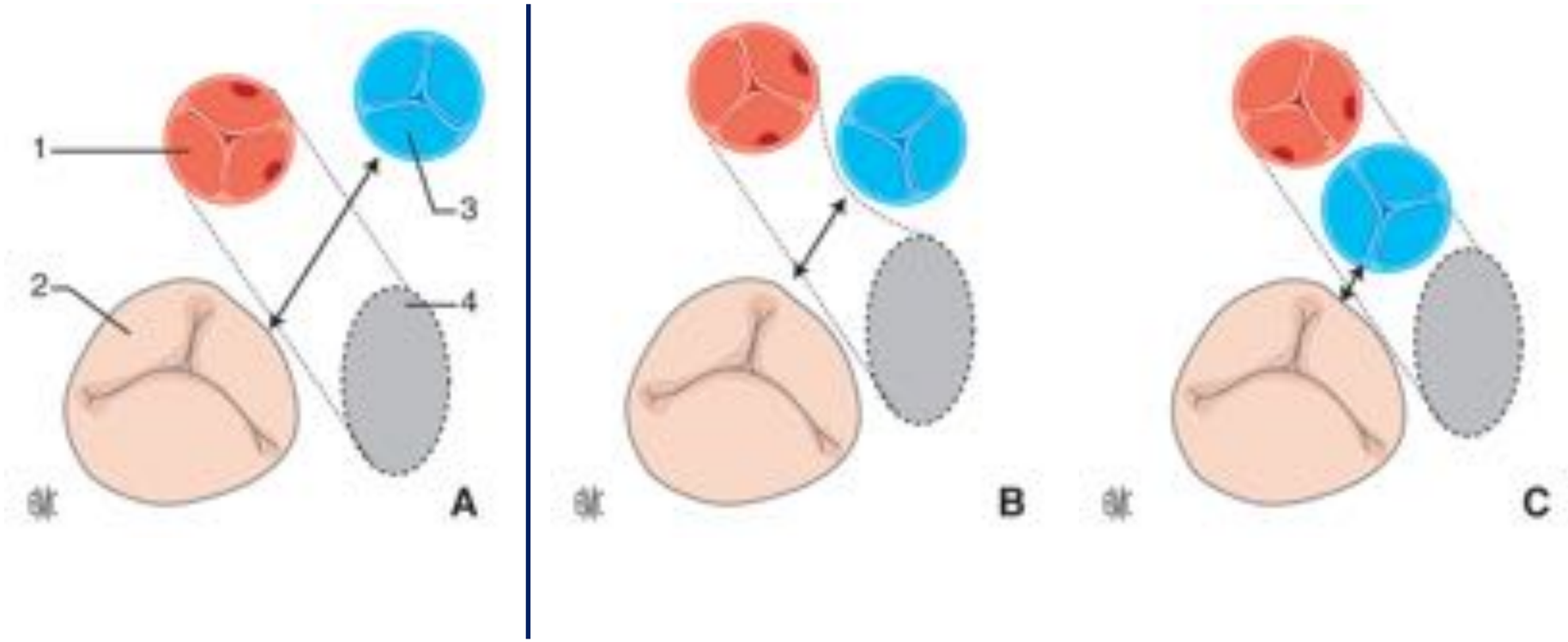
2. is "anatomic" repair feasible ?

. LV connected to Aorta
. RV connected to PA
. arterial valves in native position
. no extracardiac conduit

IntraVentricular Repair (IVR)

- VSD-type (no pulmonary stenosis)
- Fallot-type (pulmonary stenosis)

Determinant: tricuspid-to-pulmonary distance (lenght of subpulmonary conus)

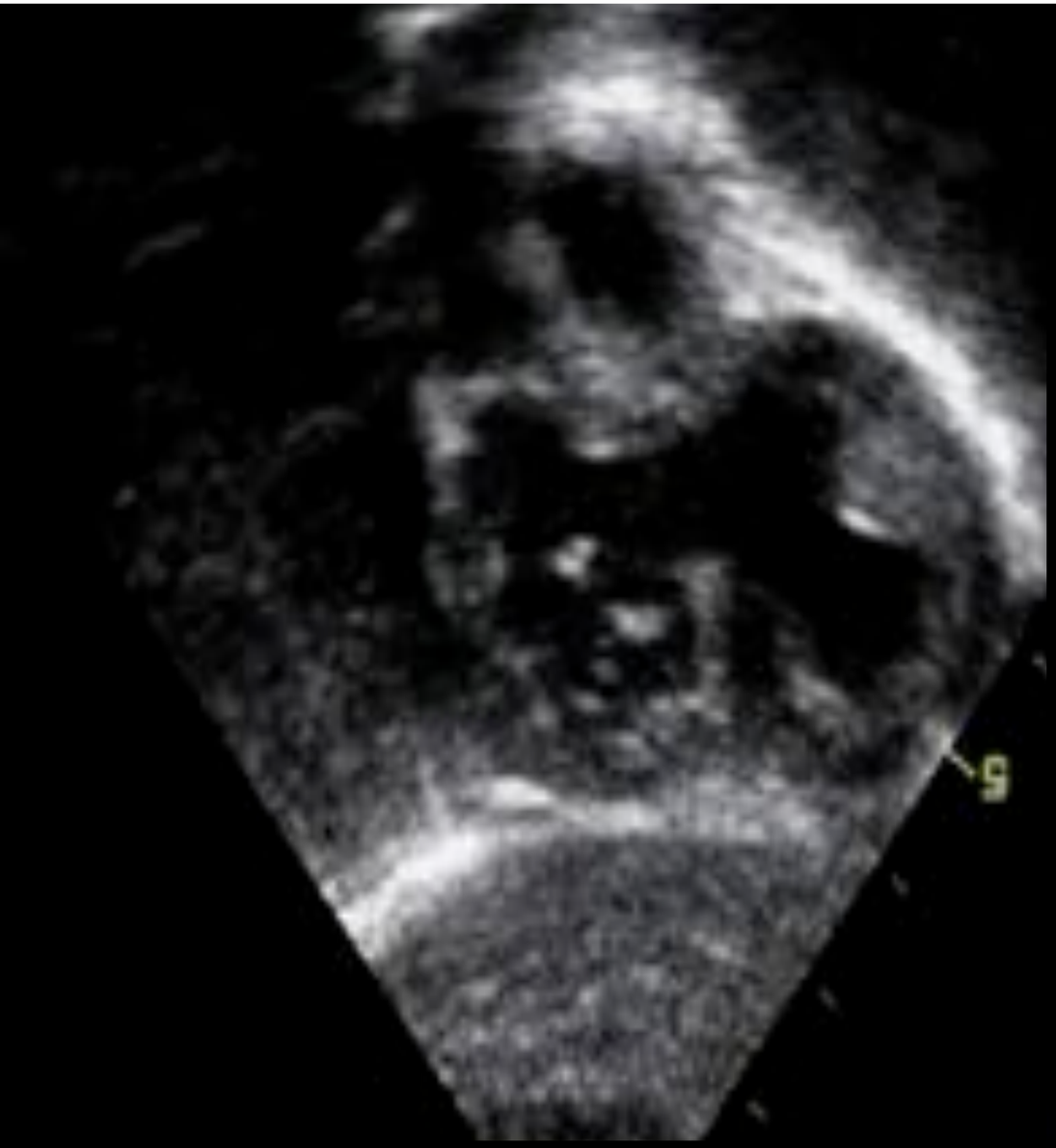


IVR possible

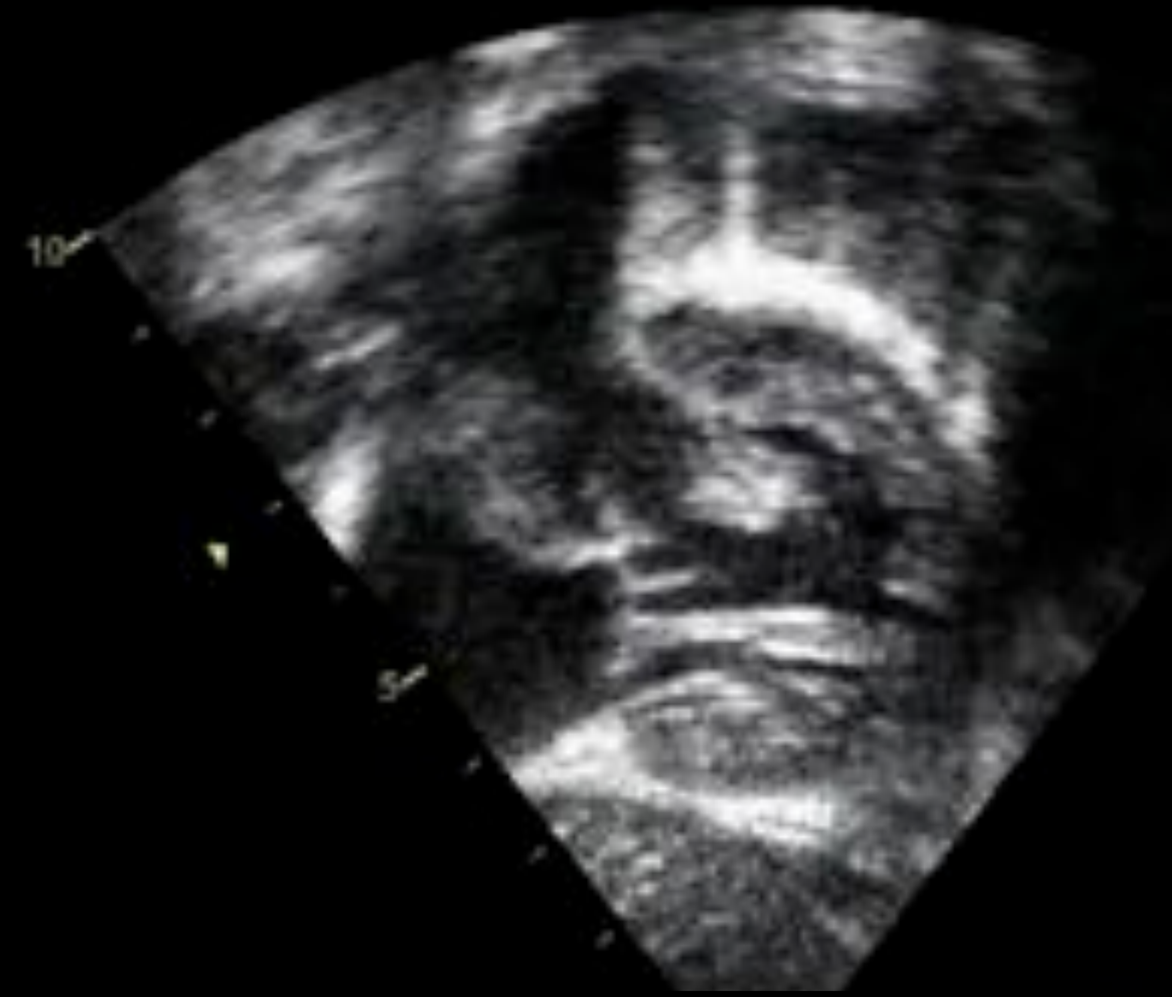
IVR impossible

DORV

« Late » DORV sub aortic VSD-Evaluation for IVR



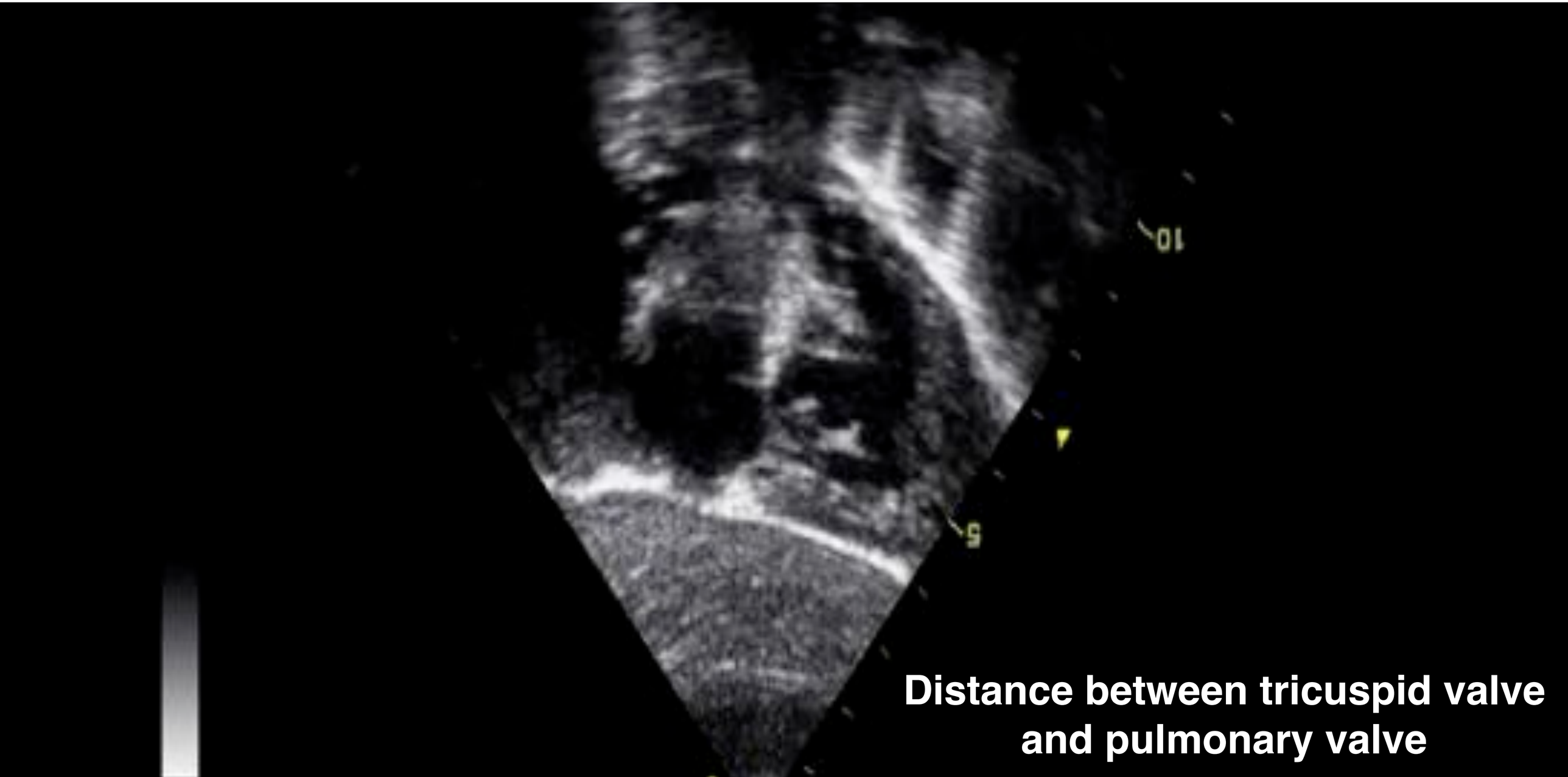
DORV sub-aortic VSD



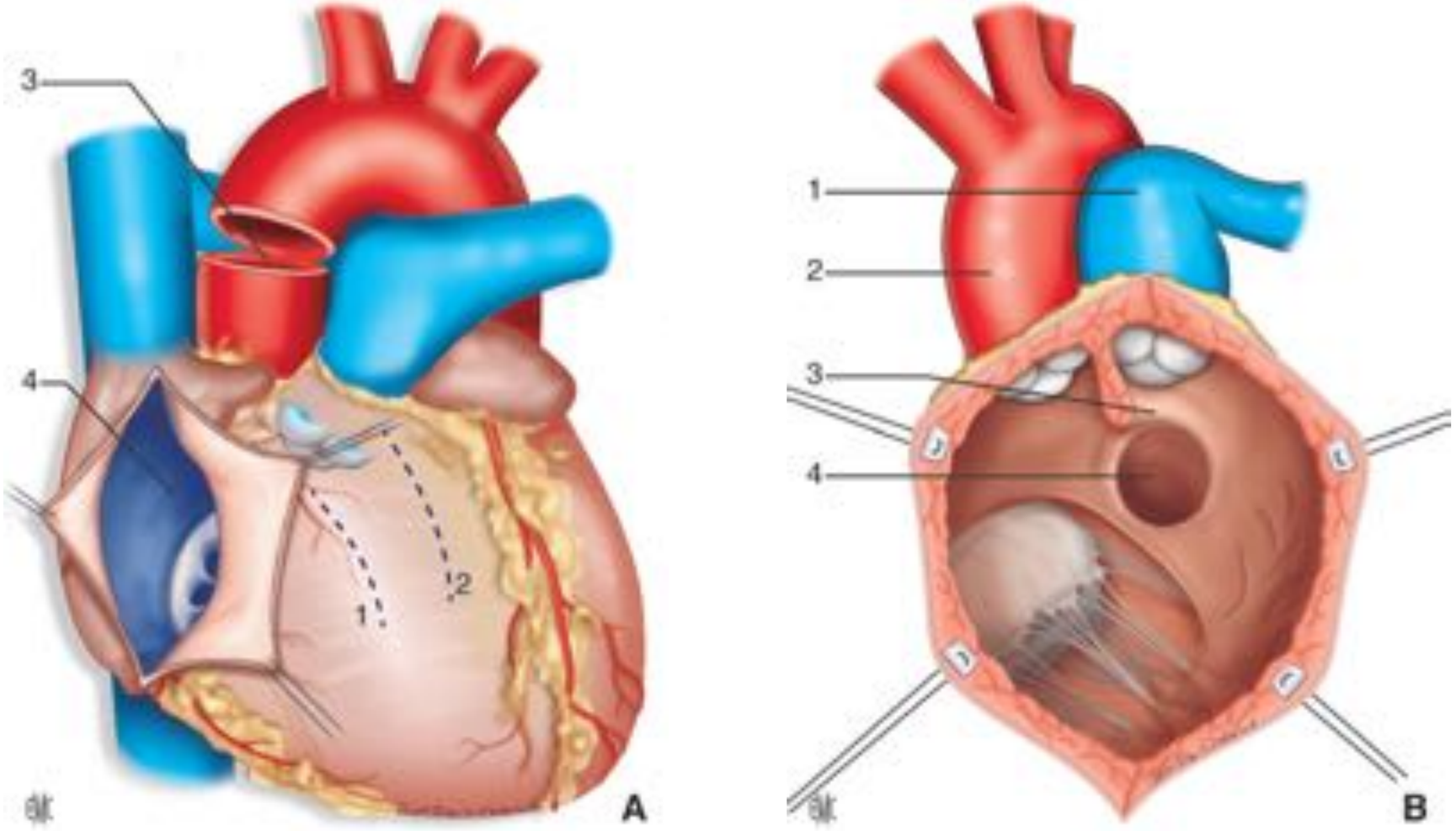
Distance between tricuspid valve and pulmonary valve

DORV

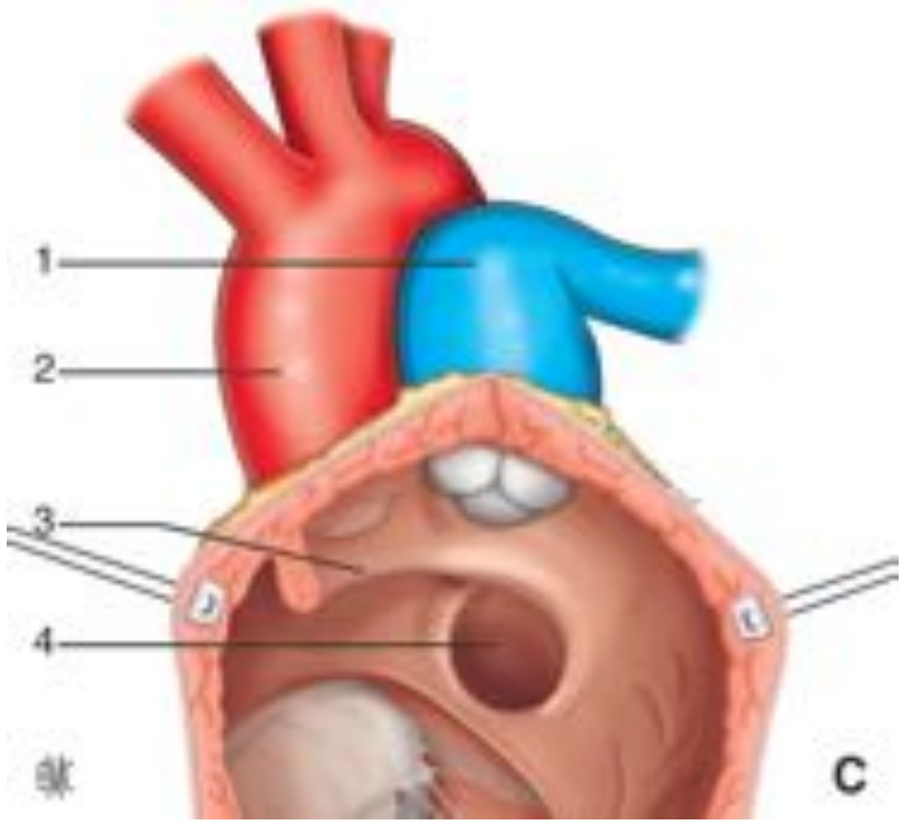
« Late » DORV sub aortic VSD-Evaluation for IVR

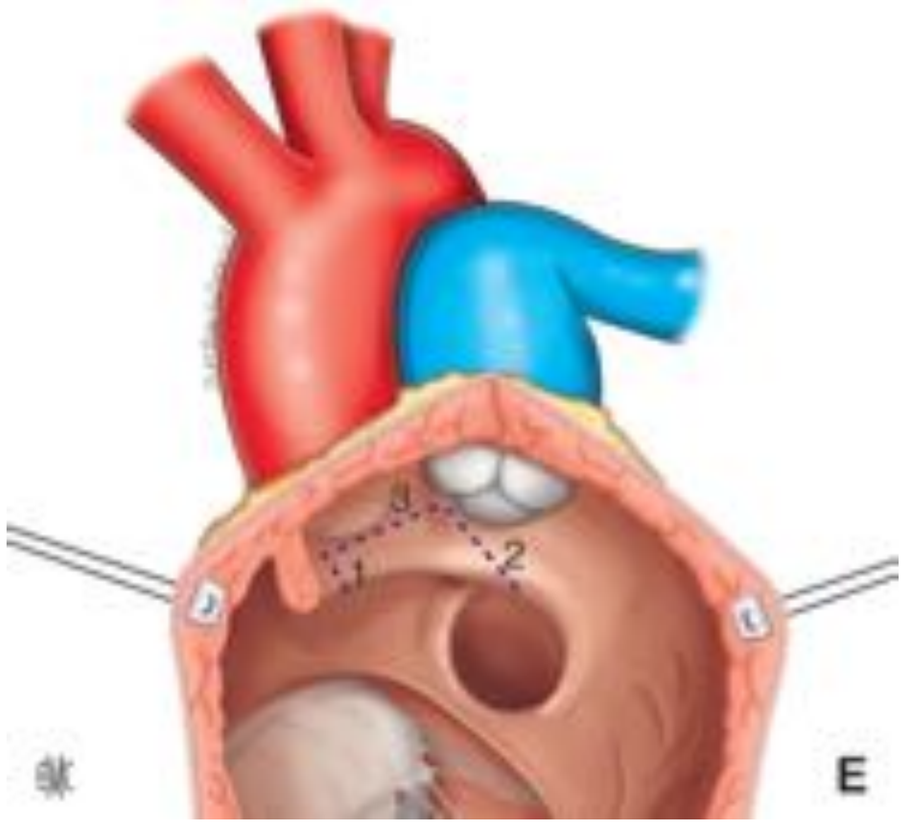


Intraventricular repair

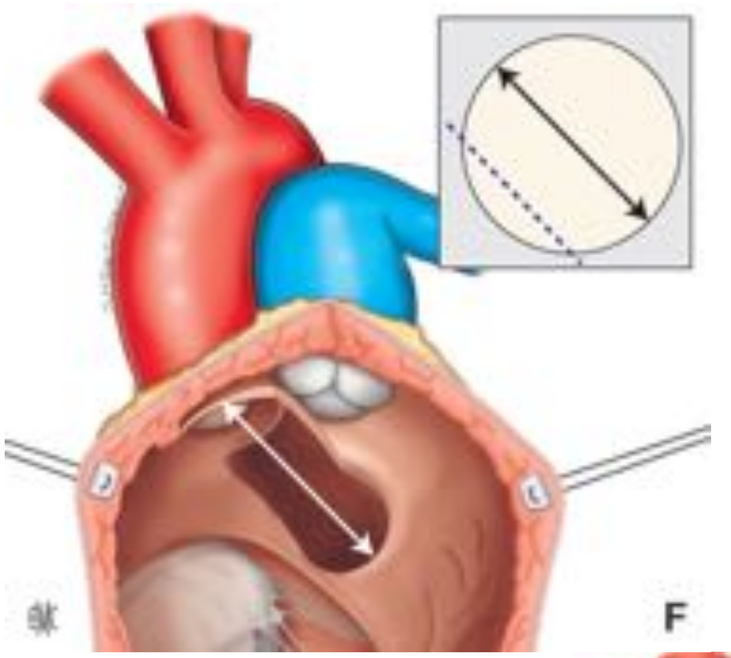


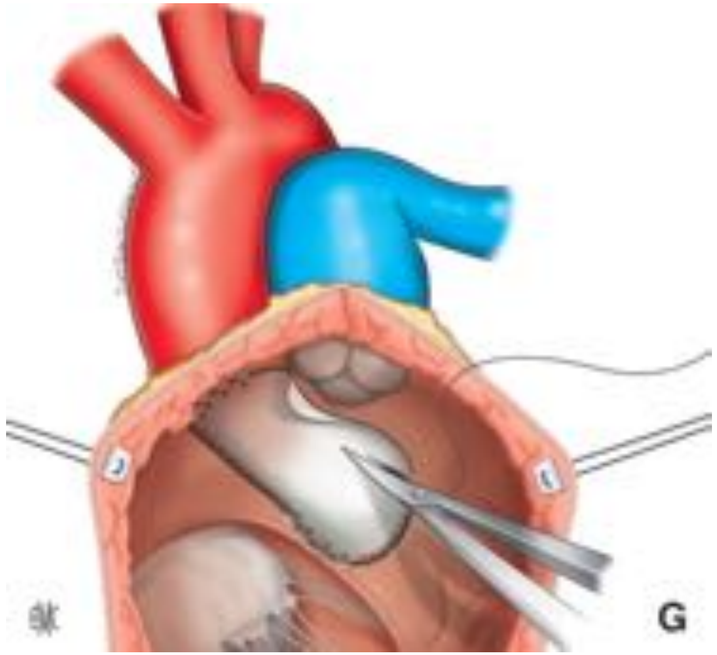
Intraventricular repair

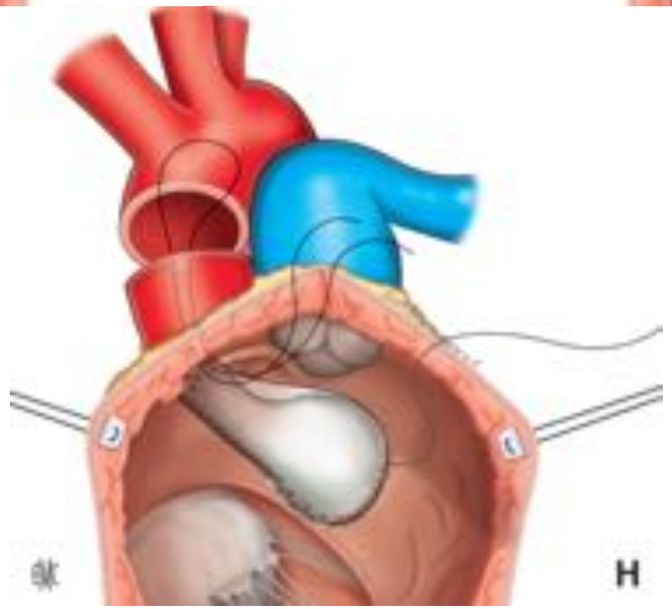




Intraventricular repair

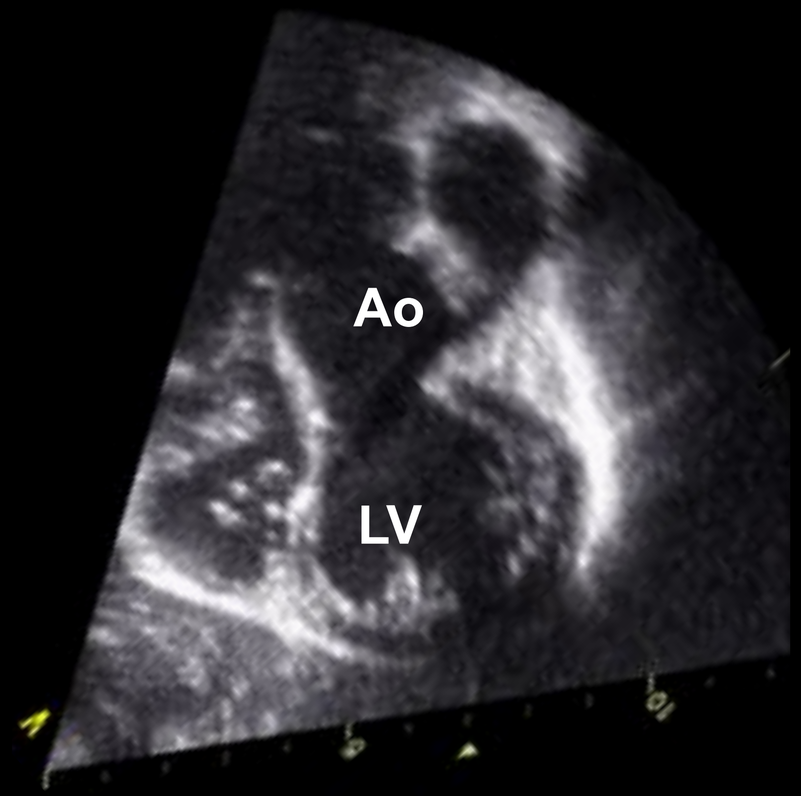


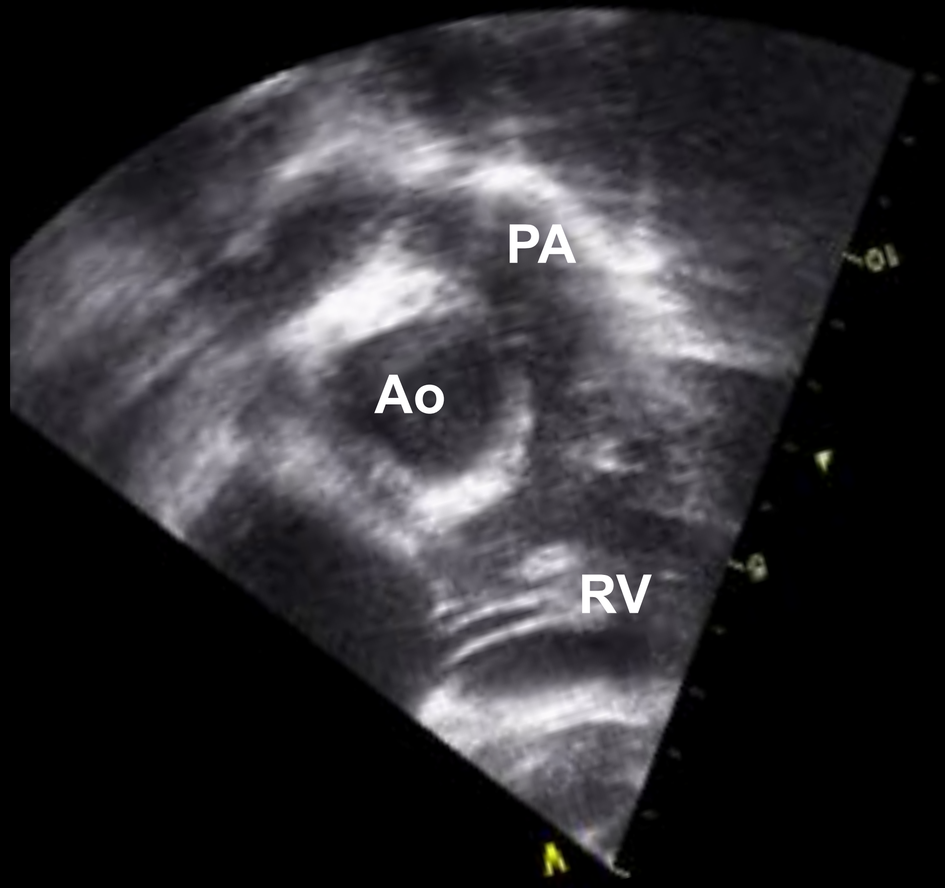




DORV

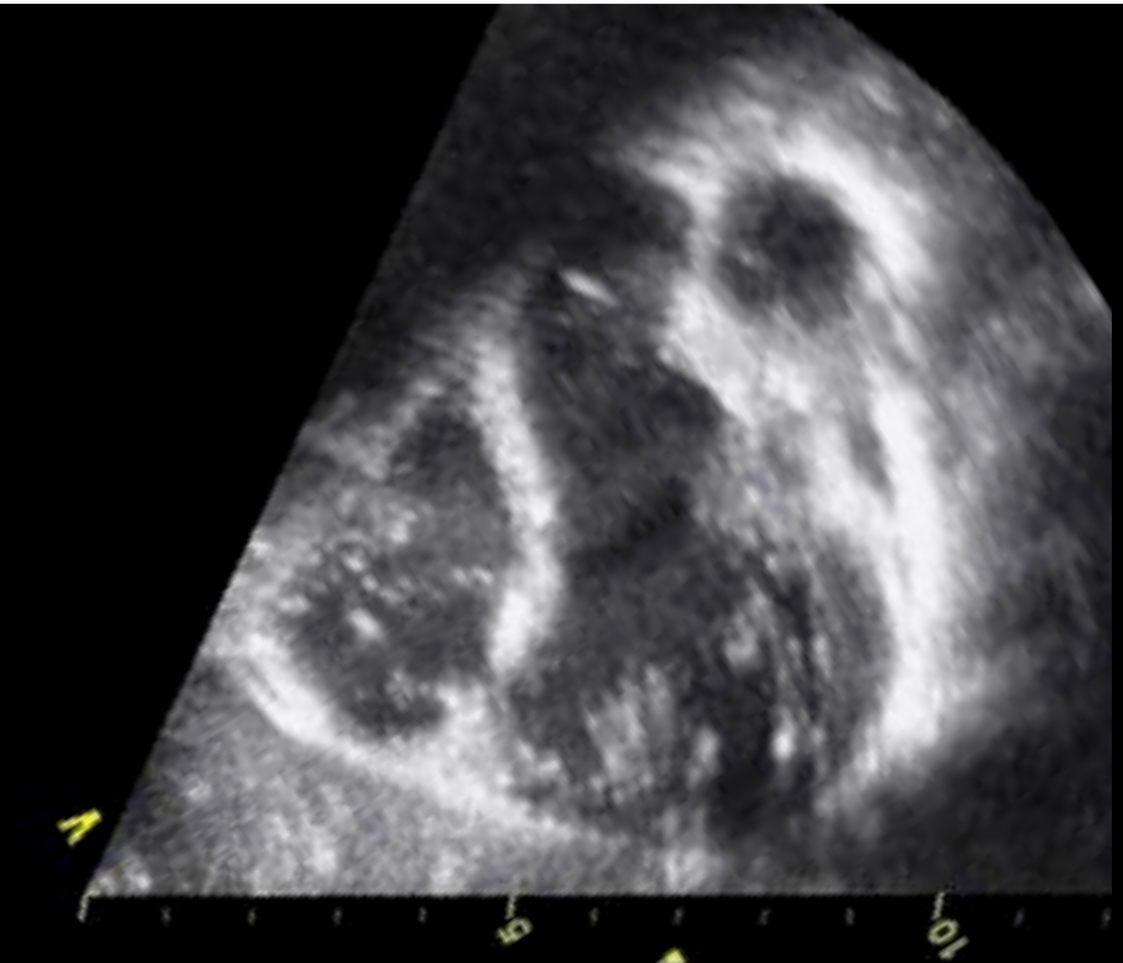
« Late » DORV sub aortic VSD-After IVR

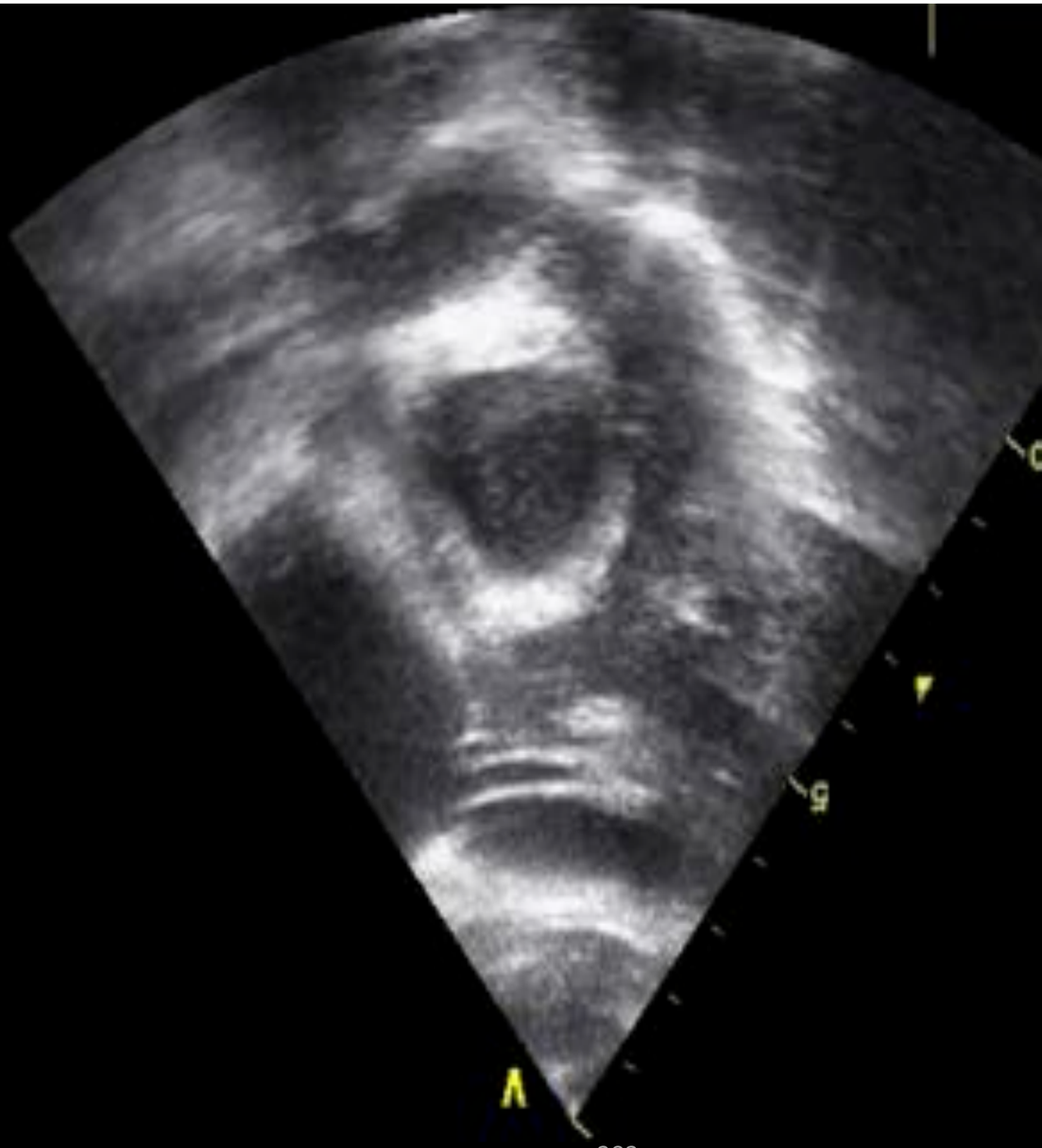




DORV

« Late » DORV sub aortic VSD-After IVR





2. is "anatomic" repair feasible ?

IVR may be possible but difficult :

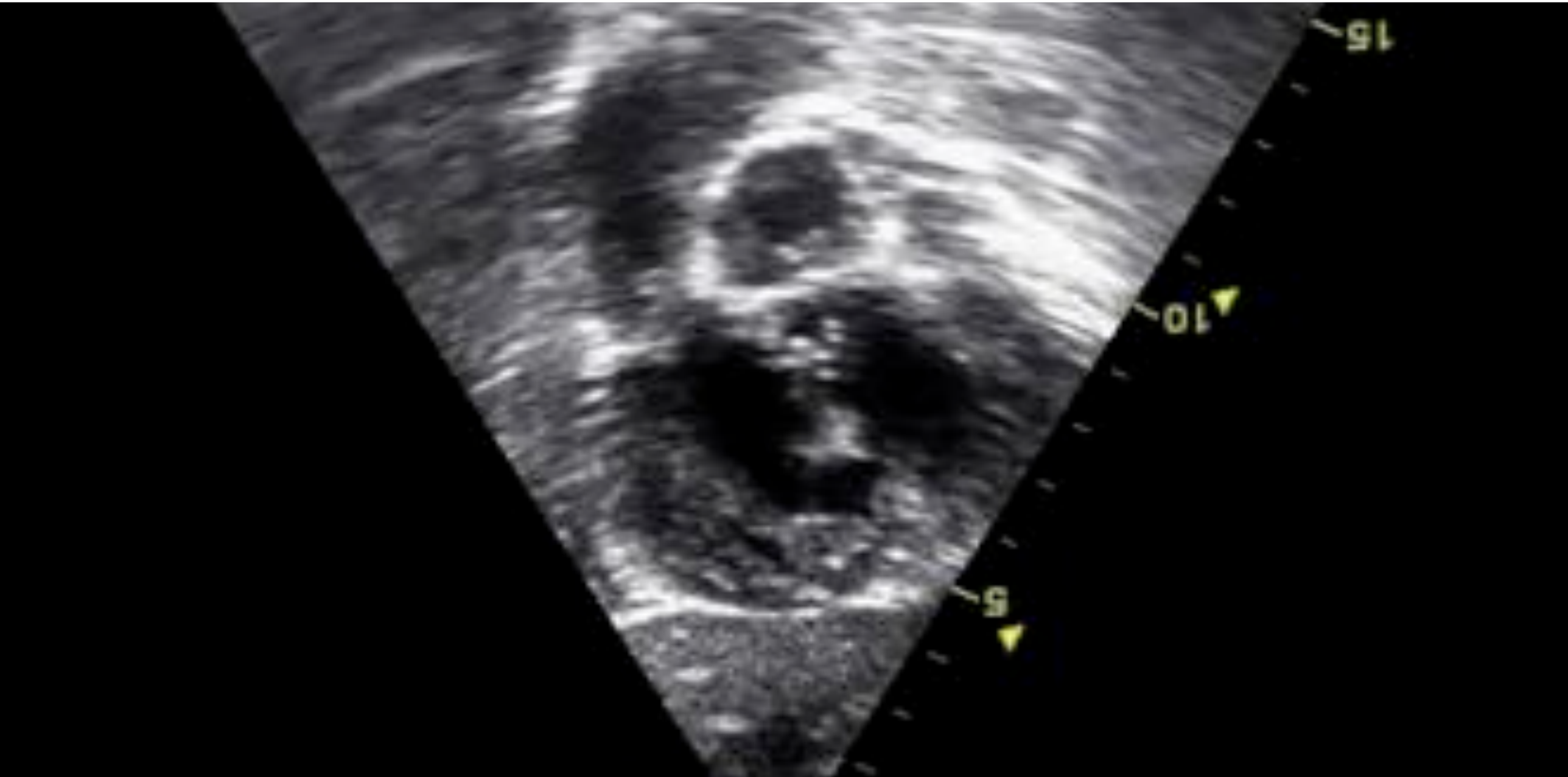
- **extensive abnormal insertions of tricuspid and/or mitral valve**

- **asymmetric subpulmonary conus**

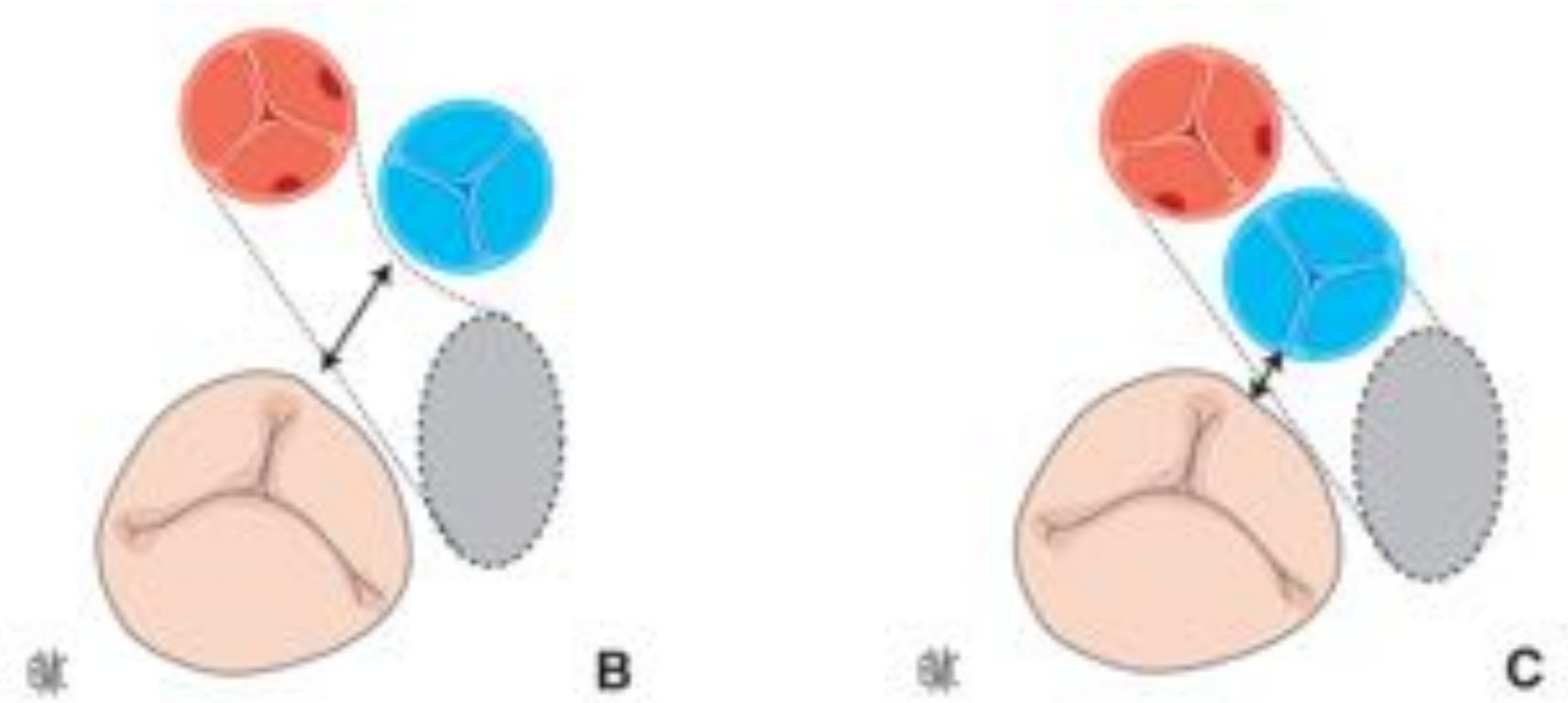
extra-anatomic repair may be preferable

DORV

« Late » DORV sub aortic VSD-Anormal tricuspid valve insertions



Tricuspid-to-pulmonary distance < Ao diameter



IVR impossible

3. which extra-anatomic repair is indicated ?

- when "anatomic" repair (IVR) is impossible

- determinant: pulmonary outflow tract
  (particularly pulmonary valve)
  - normal
  - very abnormal (stenotic)
  - mildly abnormal (good enough for pulmonary)

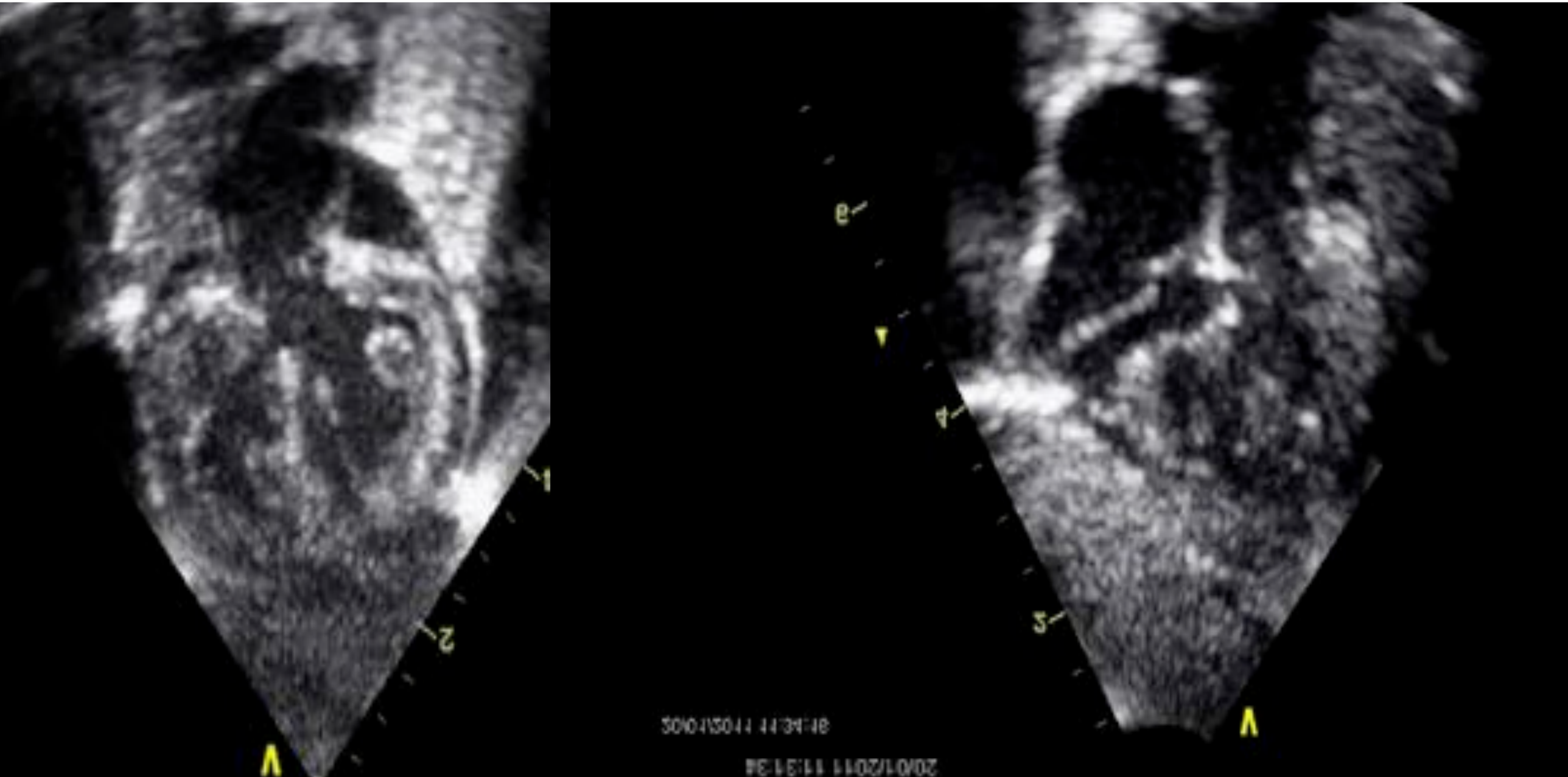
3. which extra-anatomic repair is indicated ?

- when LVOT can be used as neoaortic
  - normal pulmonary valve
  - subvalvar area
    - . normal
    - . stenosis which can be relieved

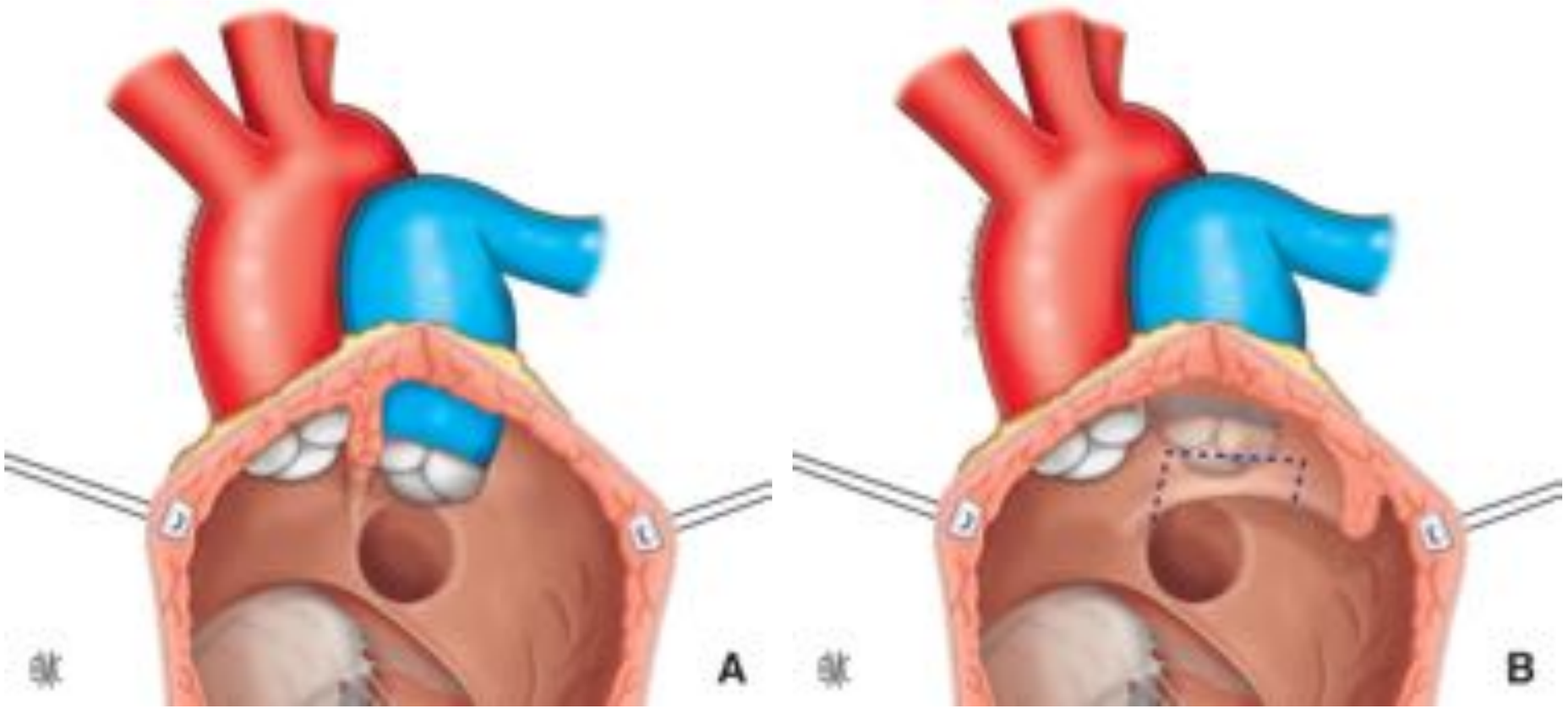
- **LV to PA connection + arterial switch**

DORV

« Late » DORV -Short Tricuspid-Pulmonary valve distance



LV-PA connection + arterial switch



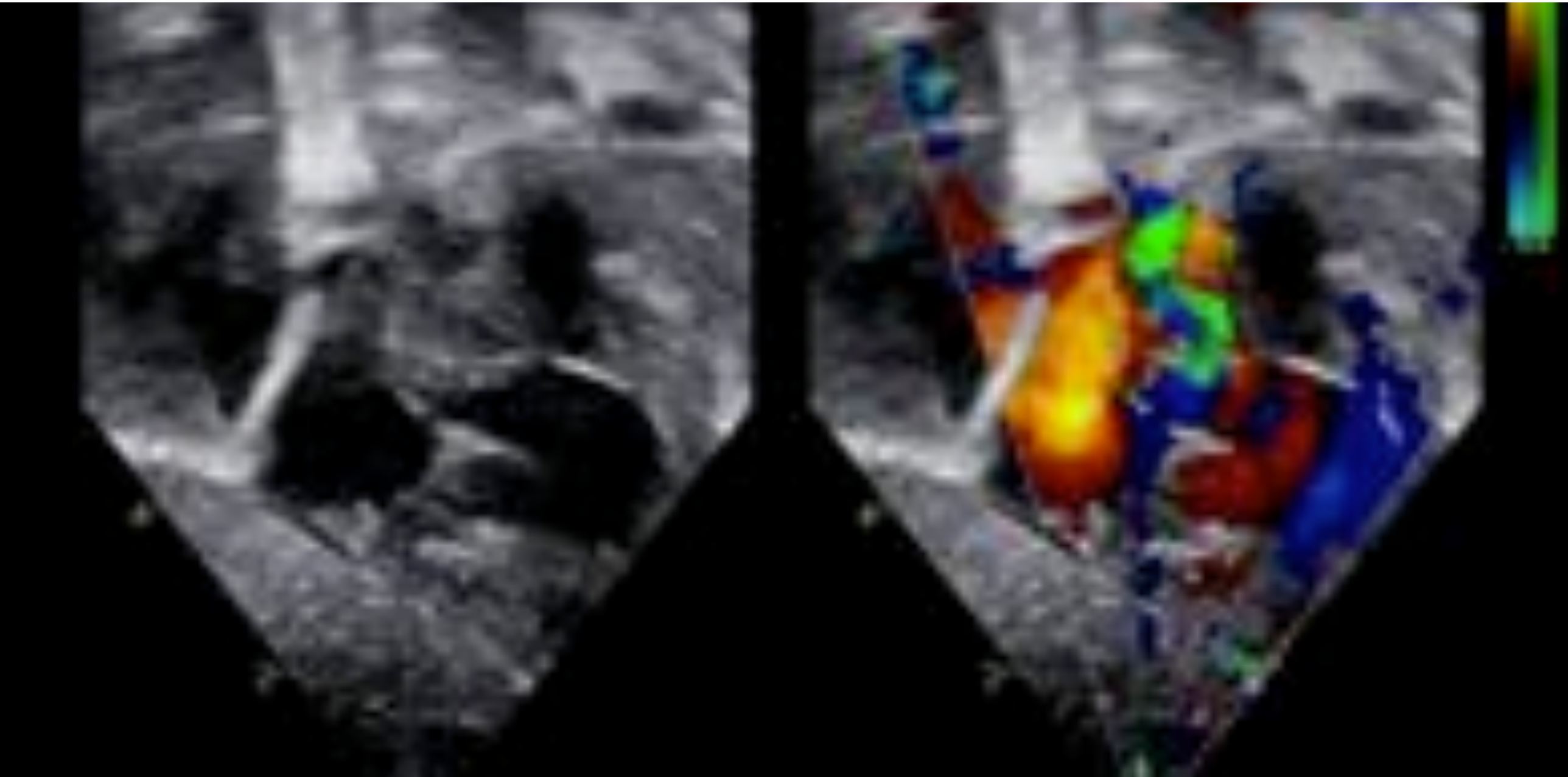
3. which extra-anatomic repair is indicated ?

- when LVOT cannot be used as neoaortic
  - severe valvar stenosis
  - non-resectable subvalvar obstruction

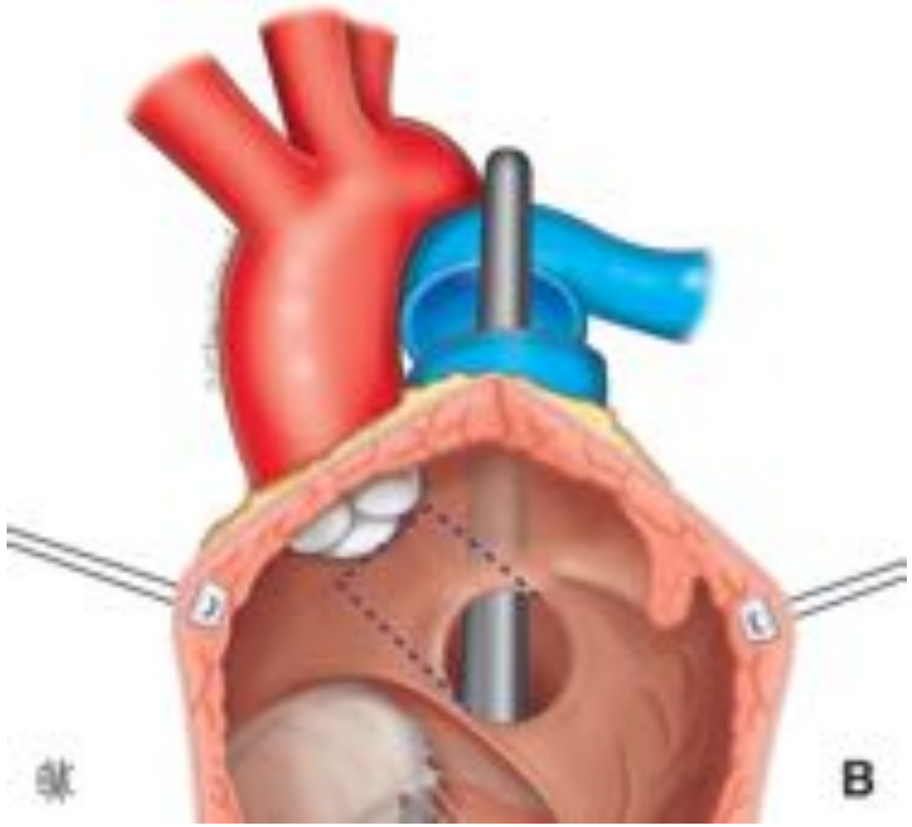
- **REV operation**
- **Bex-Nikaidoh operation**
- **Rastelli operation**

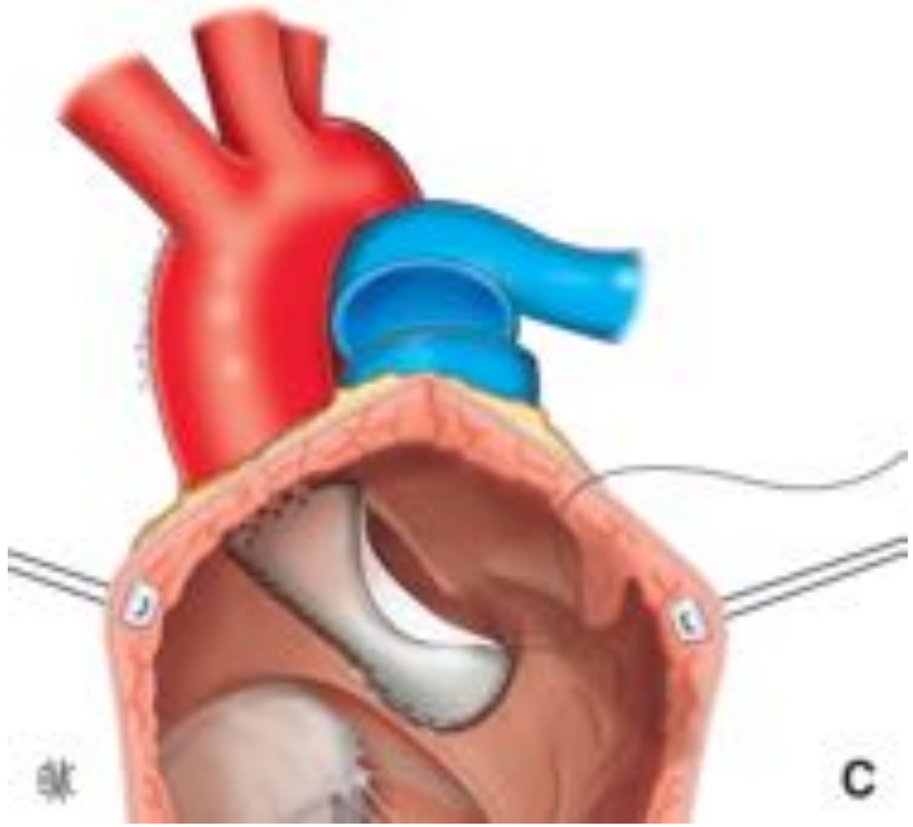
DORV

« Late » DORV -Short Tricuspid-Pulmonary valve distance-Severe subpulmonary stenosis

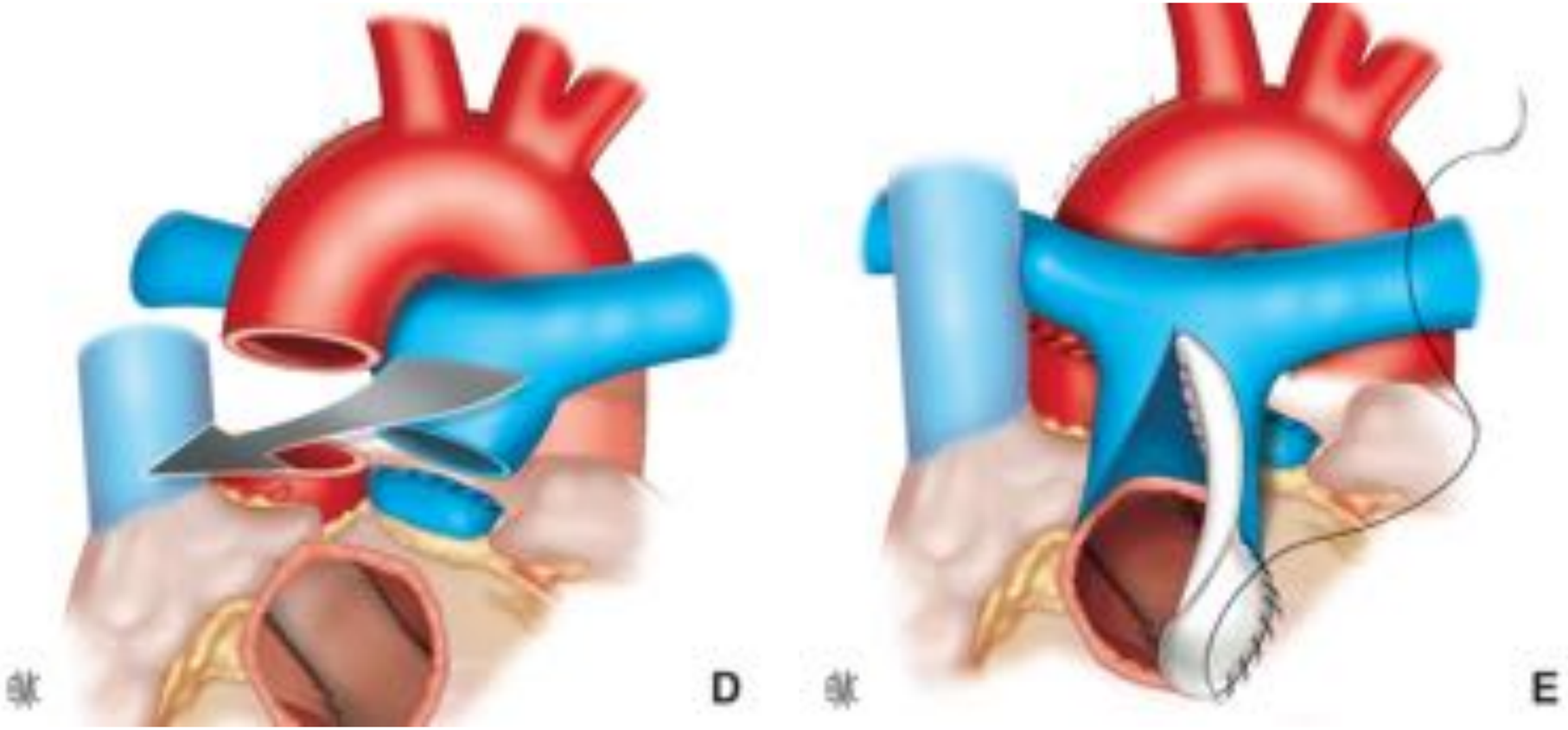


Extra-anatomic repair : REV

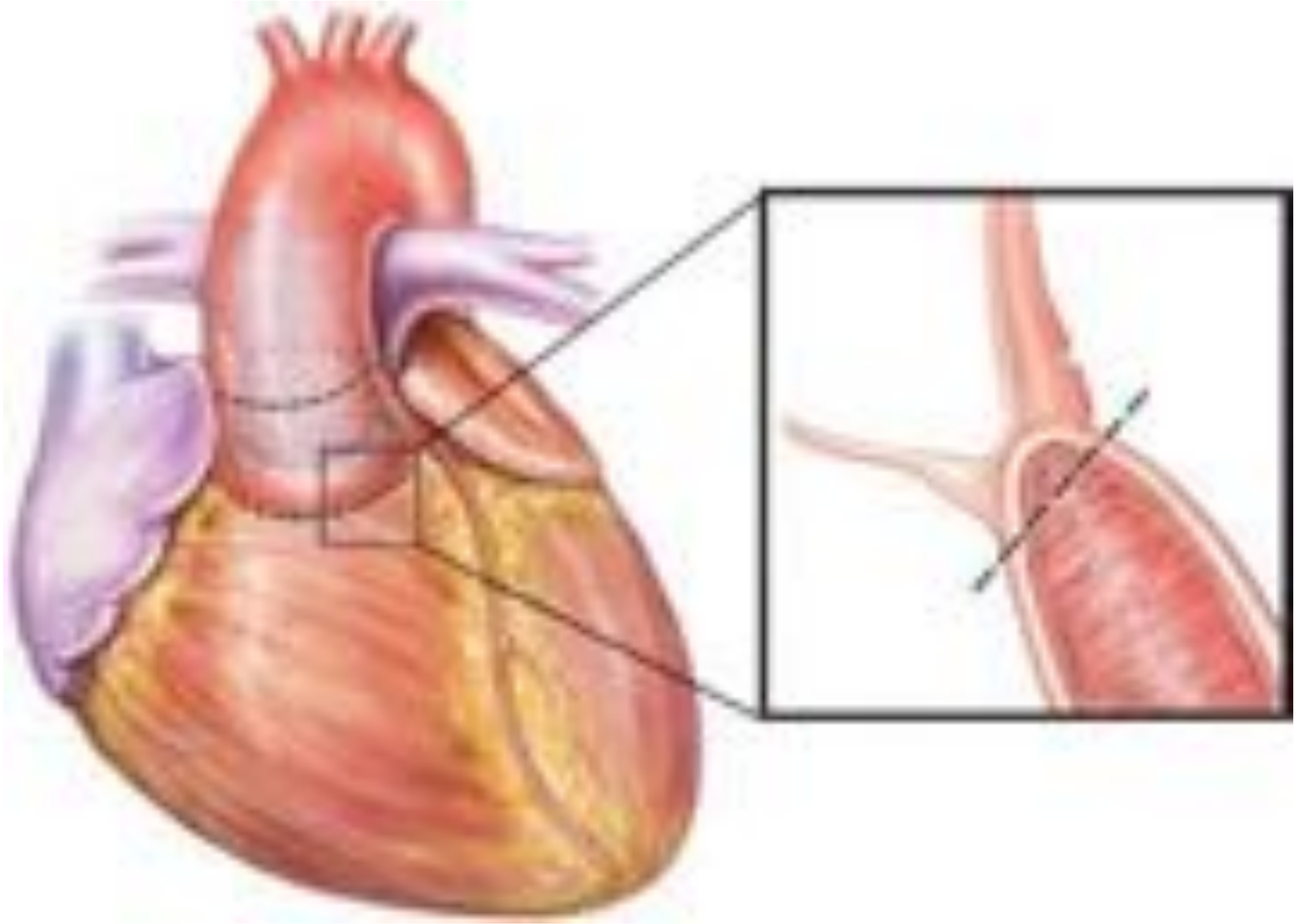




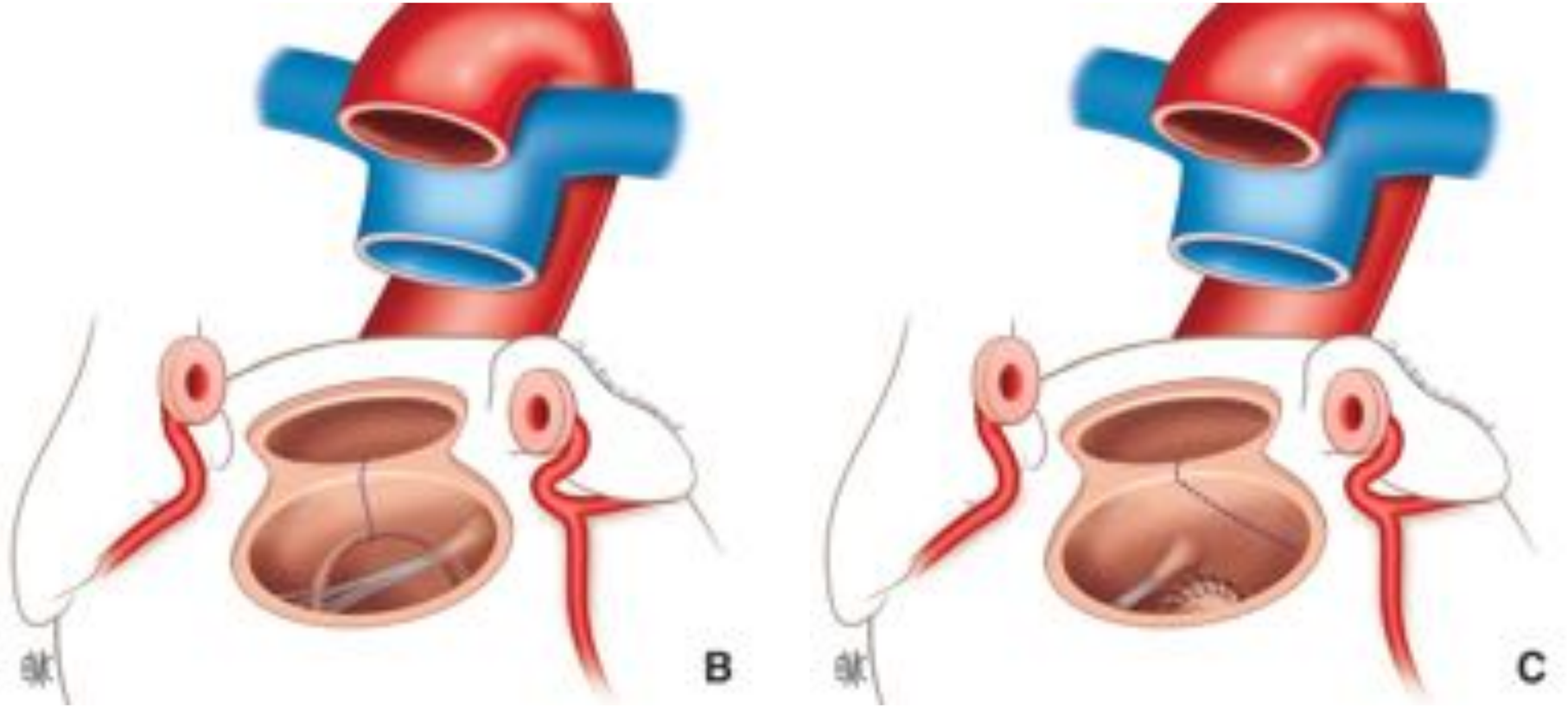
Extra-anatomic repair : REV



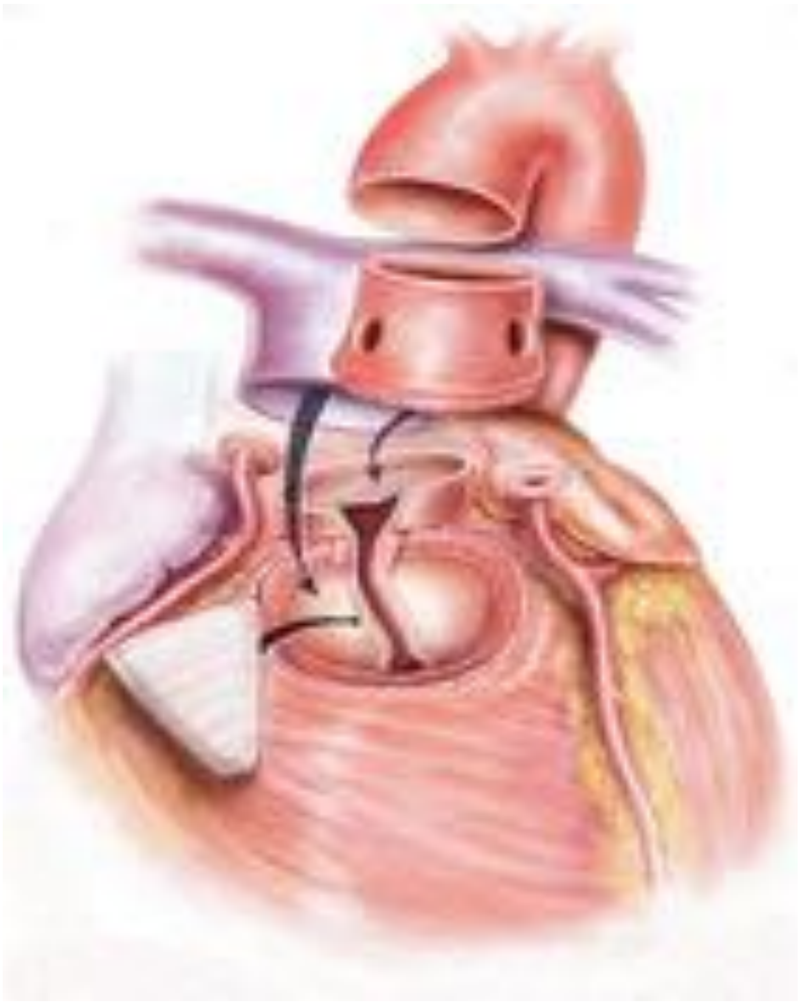
Extra-anatomic repair
Bex-Nikaidoh operation

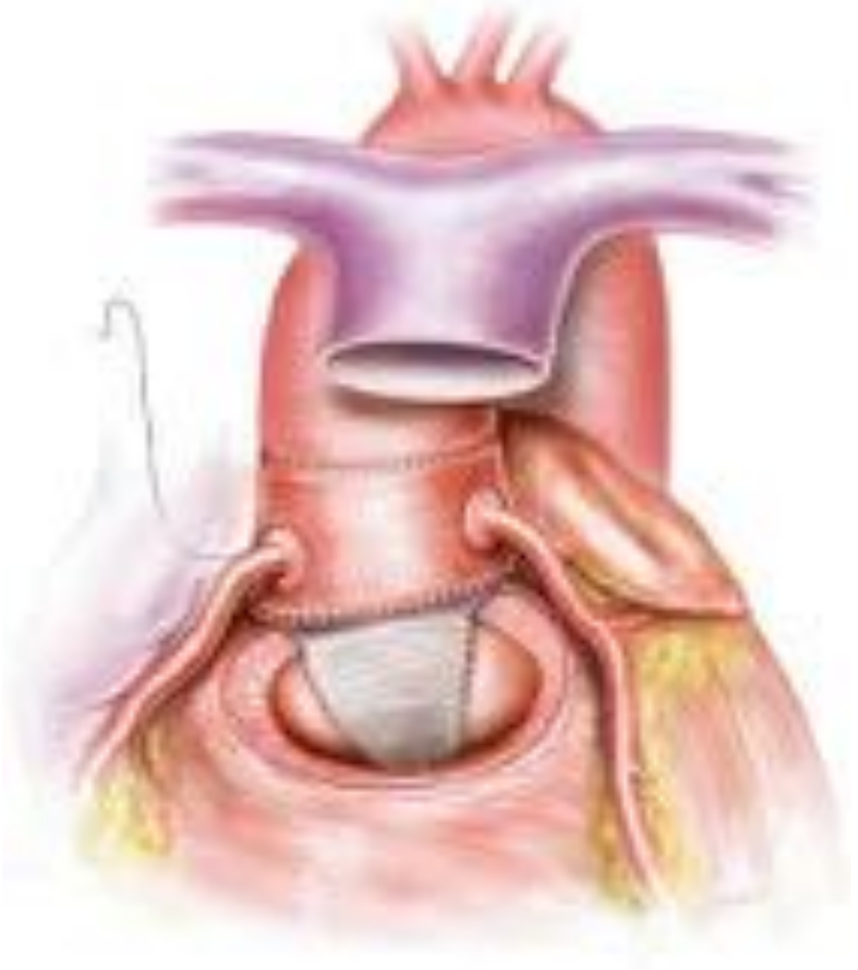


Extra-anatomic repair
Bex-Nikaidoh operation

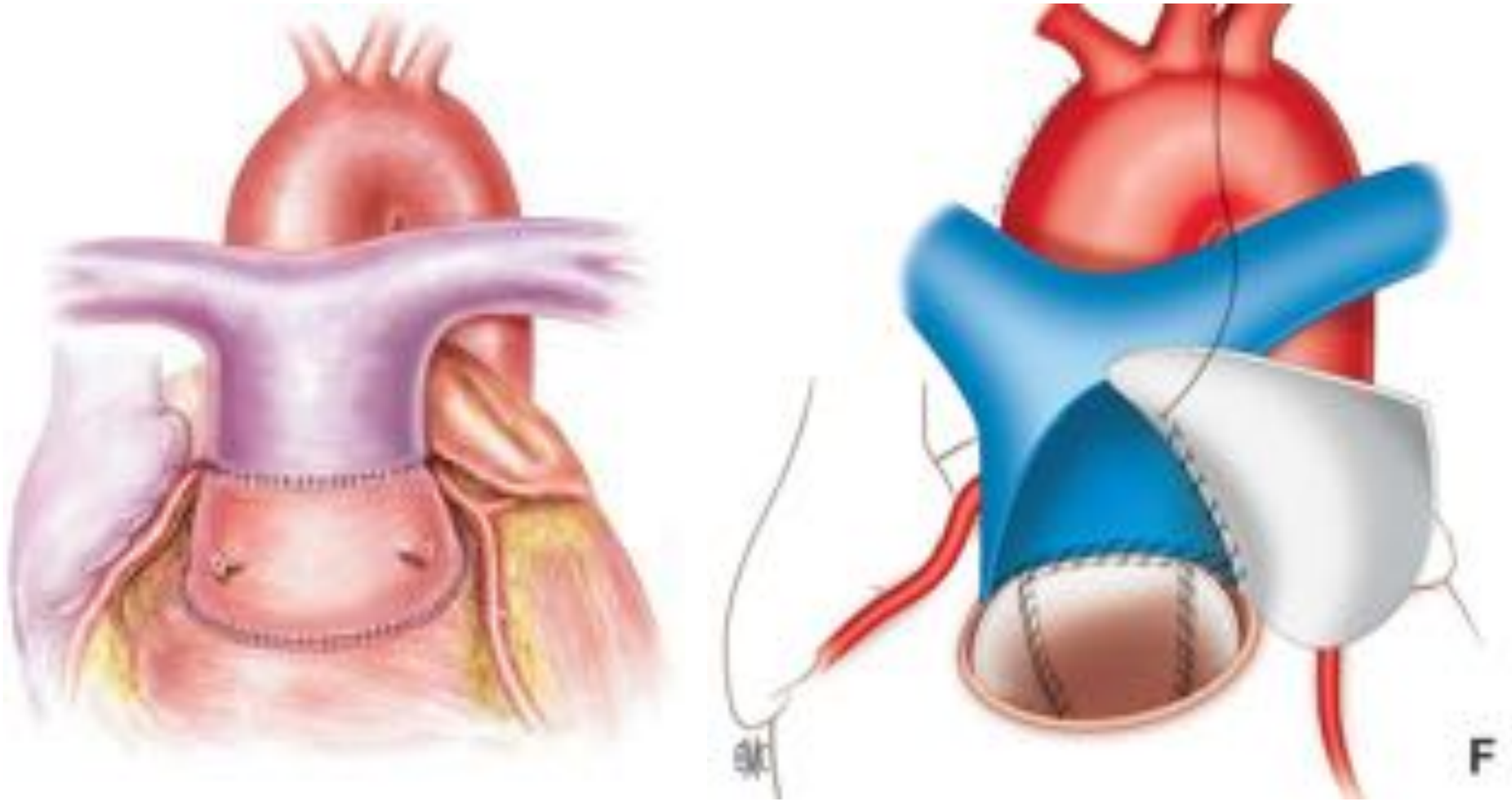


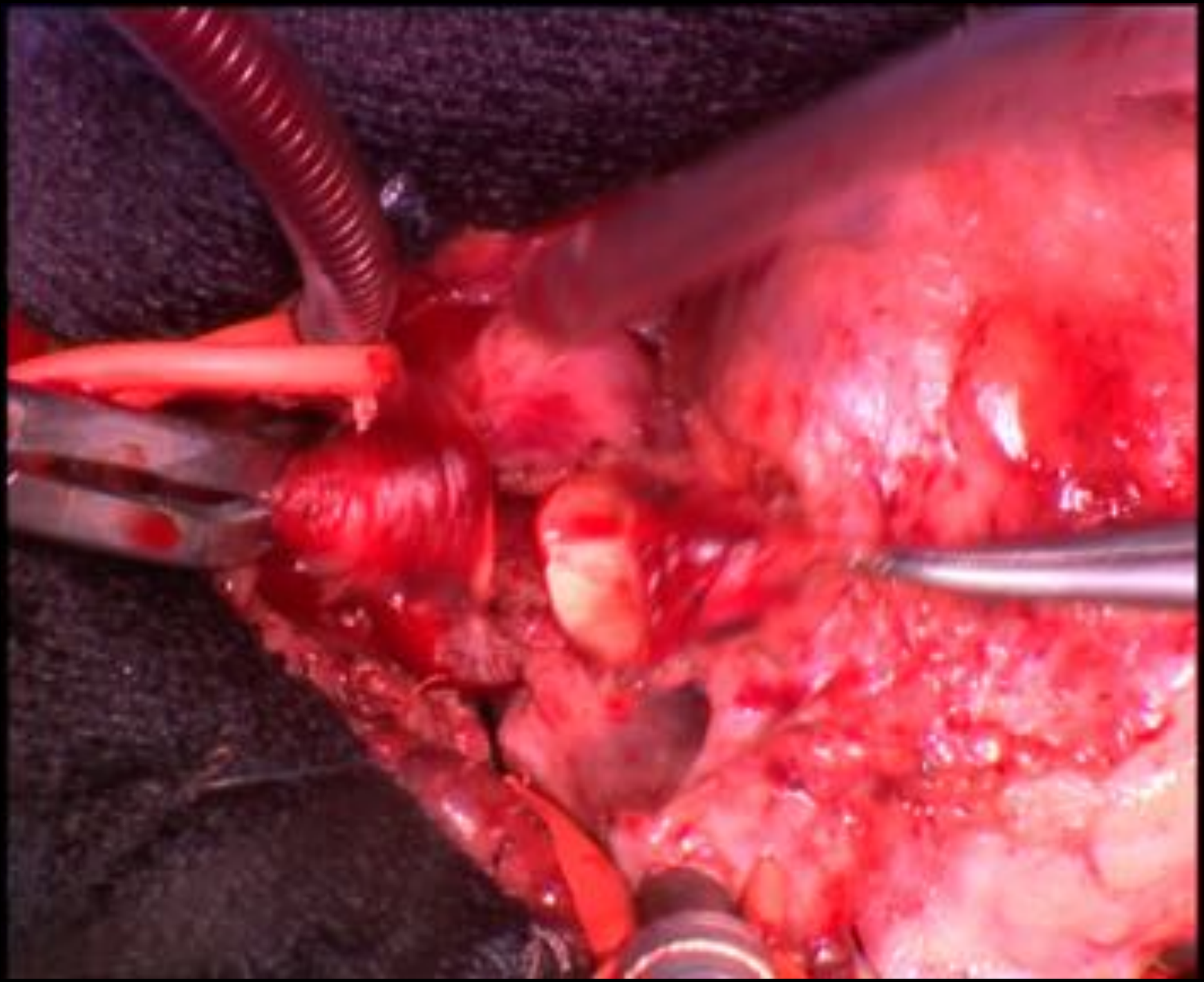
Extra-anatomic repair
Bex-Nikaidoh operation





Extra-anatomic repair
Bex-Nikaidoh operation





Indications for Bex-Nikaidoh operation

when REV is difficult / impossible

- abnormal insertions of mitral valve on conal septum
- extensive abnormal insertions of tricuspid valve
- remote VSD (inlet, muscular)
- absence of VSD

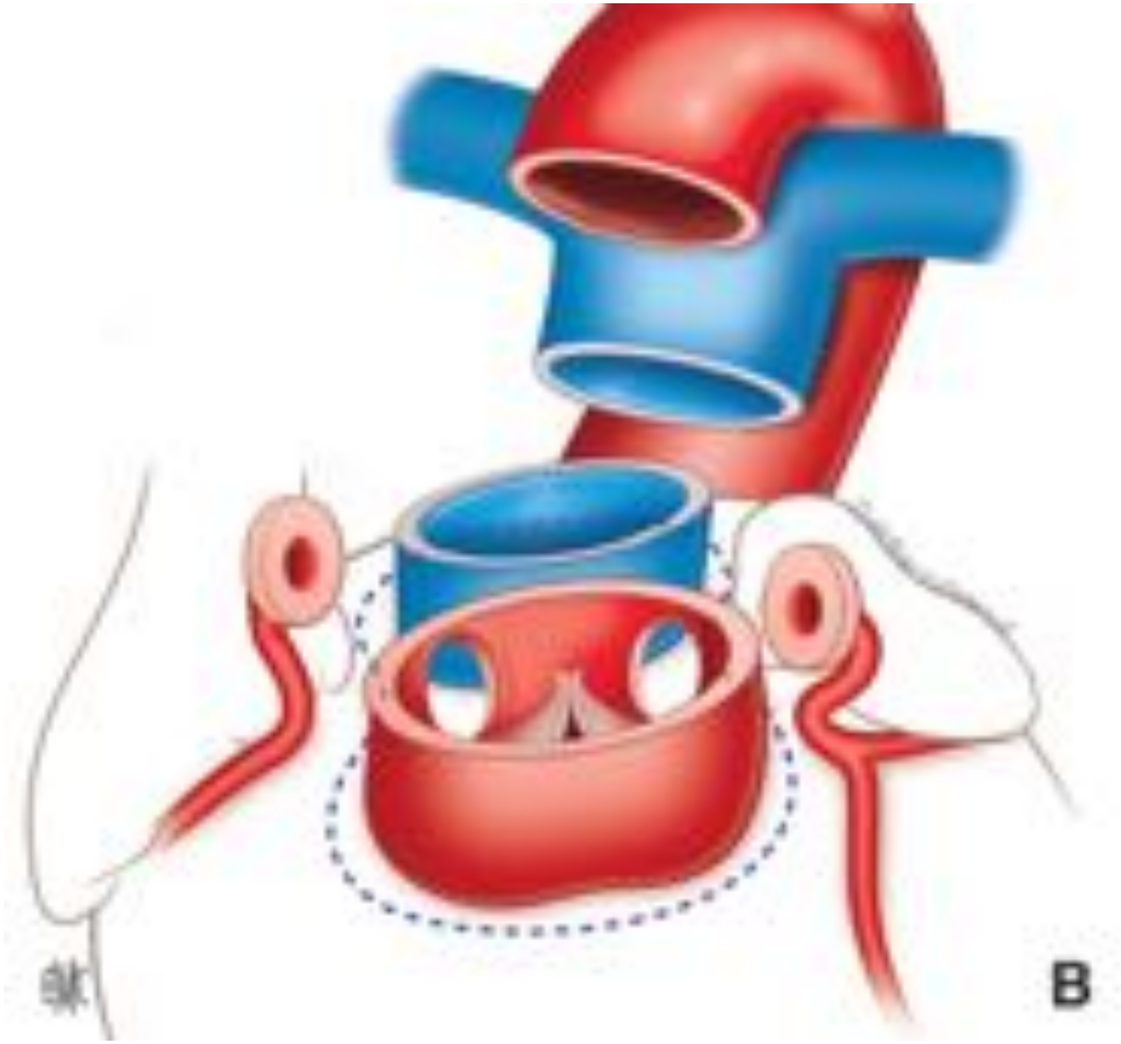
- coronary anatomy must allow Bex-Nikaidoh
  (no anterior loop)

3. which extra-anatomic repair is indicated ?

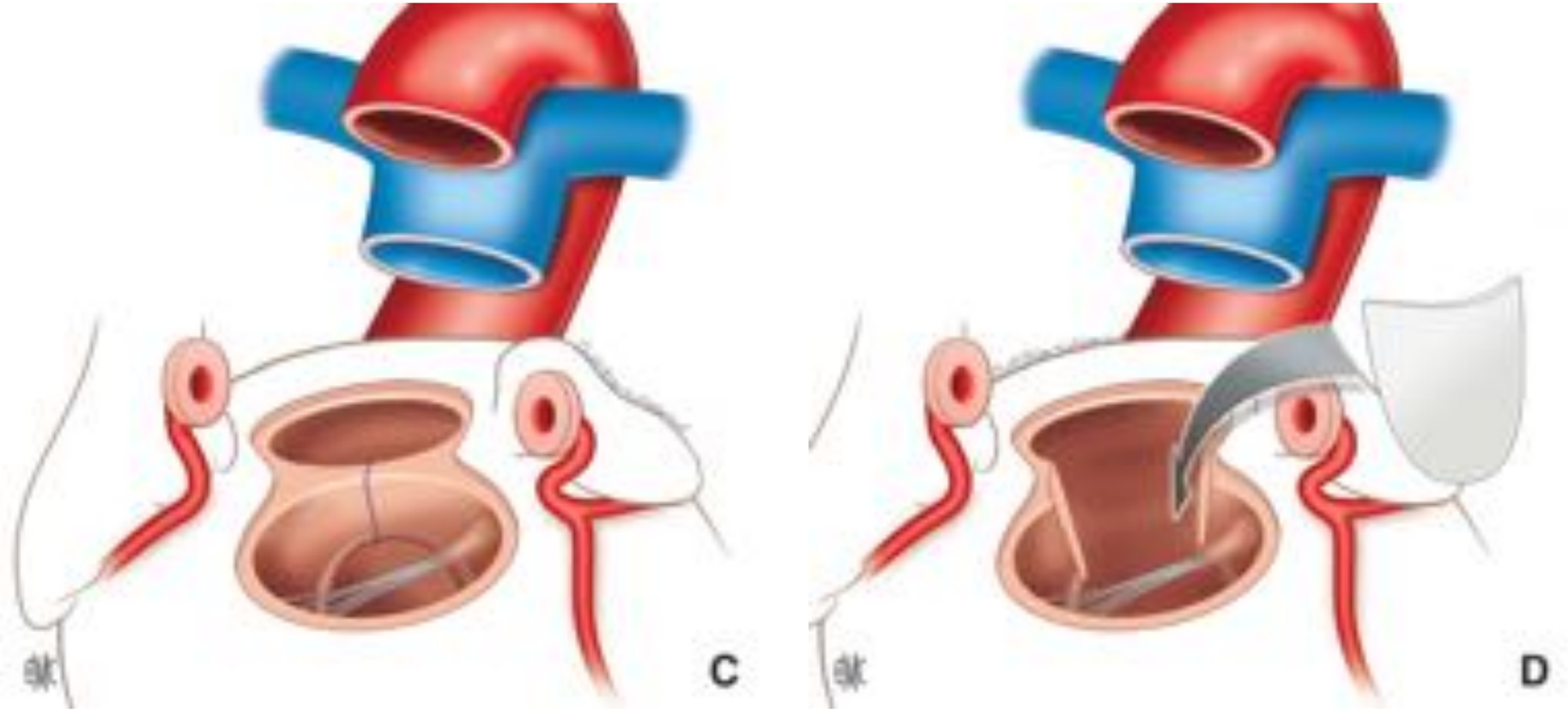
- when LVOT cannot be used as neoaortic
  but can be used as pulmonary
  - bicuspid pulmonary valve
  - mildly-dysplastic pulmonary valve

- **conotruncal rotation procedure**
  (if allowed by coronary anatomy)

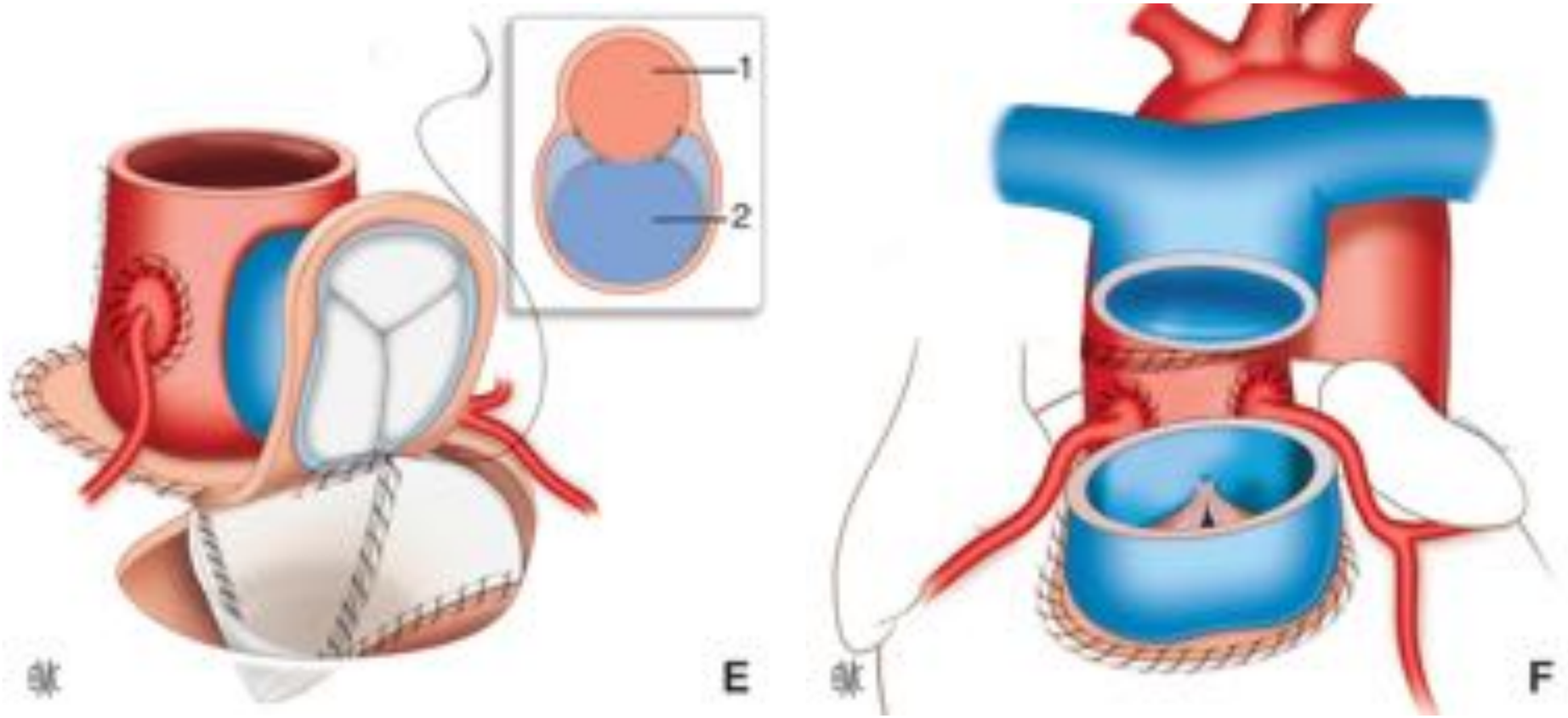
Conotruncal rotation procedure

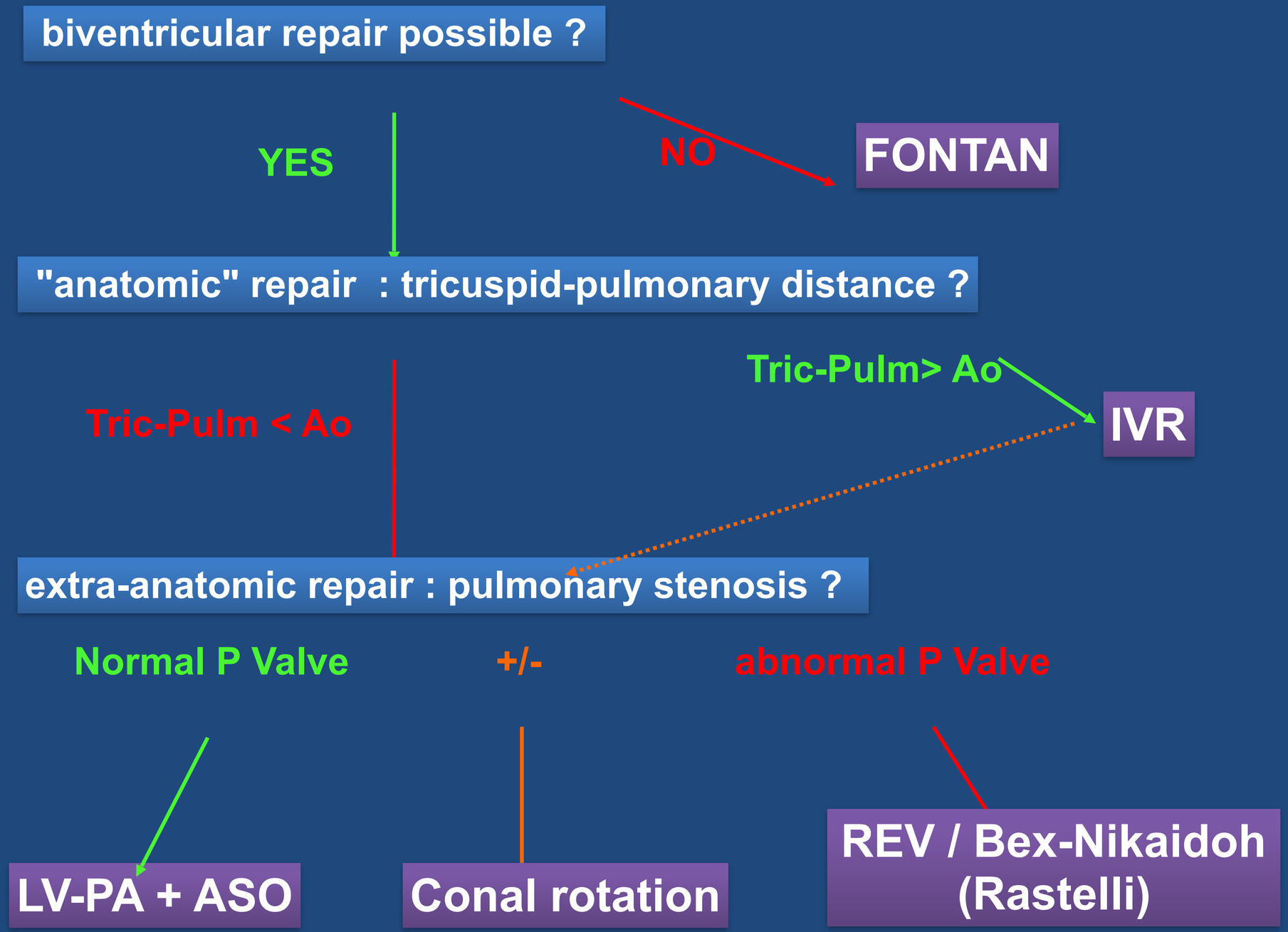


Conotruncal rotation procedure



Conotruncal rotation procedure




biventricular repair possible ?
YES
NO
FONTAN
"anatomic" repair : tricuspid-pulmonary distance ?
Tric-Pulm < Ao
Tric-Pulm> Ao
IVR
extra-anatomic repair : pulmonary stenosis ?
Normal P Valve
+/-
abnormal P Valve
LV-PA + ASO
Conal rotation
REV / Bex-Nikaidoh (Rastelli)

Cardiol Young 2007; 17: 26–34

ISSN 1047-9511
doi: 10.1017/S1047951106001326

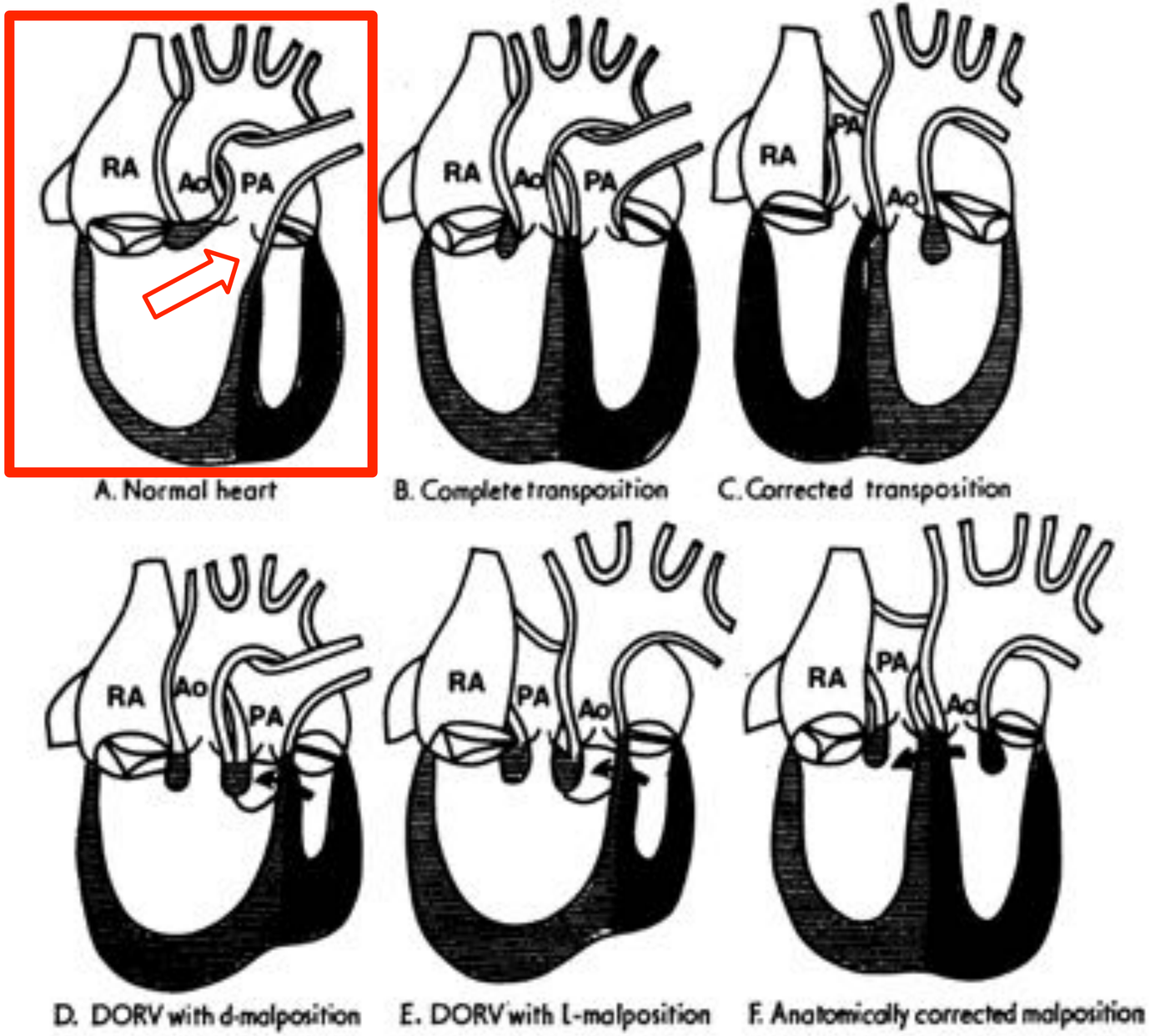
Continuing Medical Education

What is anatomically corrected malposition?

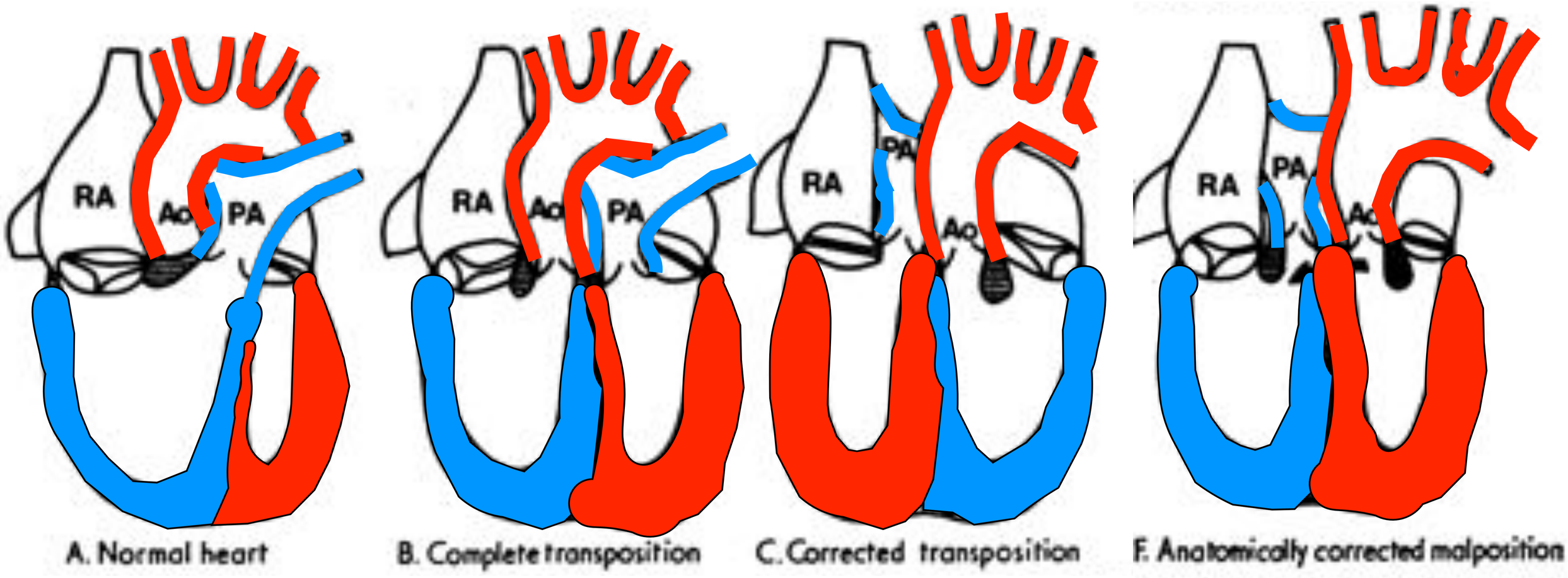
Alessandra Bernasconi,[1] Tiscar Cavalle-Garrido,[1] Don G. Perrin,[2] Robert H. Anderson[3]

[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»


RA
Ao
PA
A. Normal heart
B. Complete transposition
C. Corrected transposition
D. DORV with d-malposition
E. DORV with L-malposition
F. Anatomically corrected malposition

Anatomically corrected malposition of the great arteries

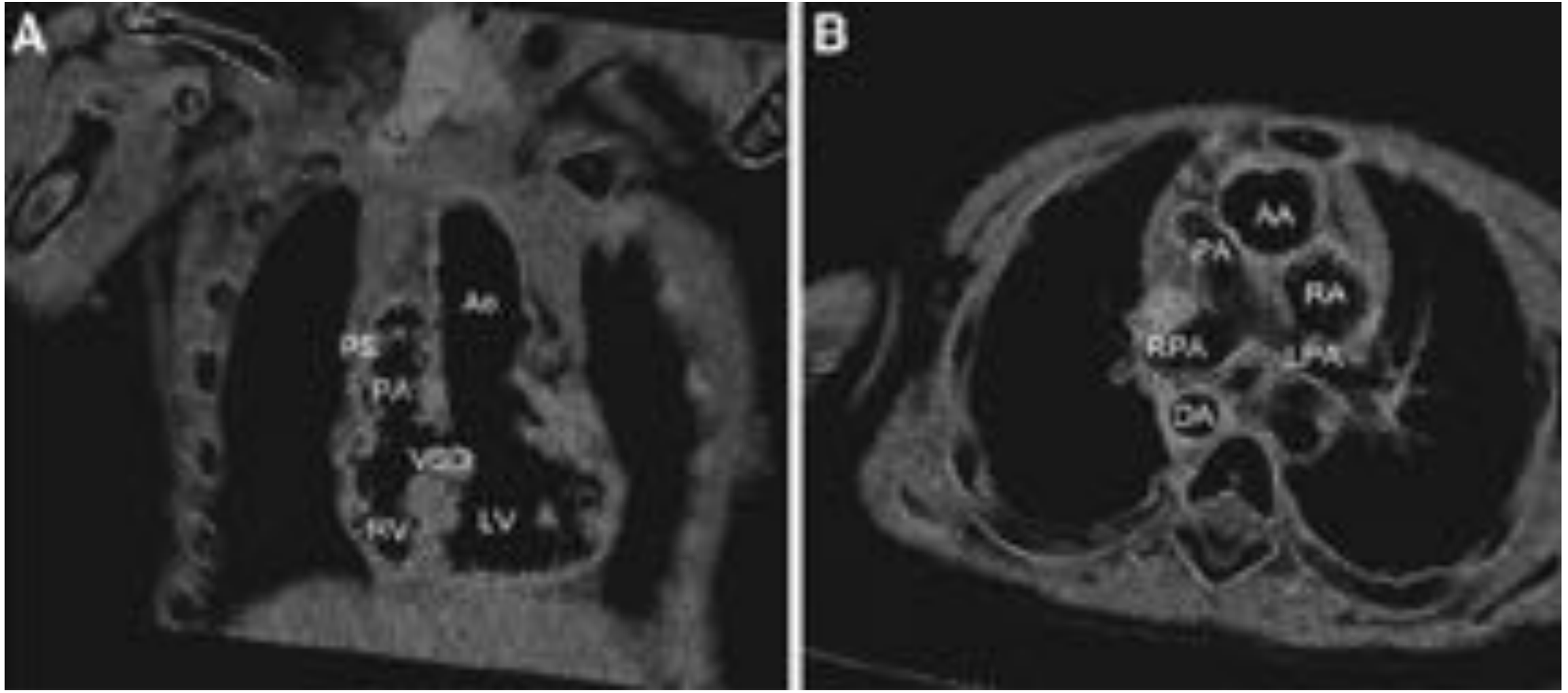


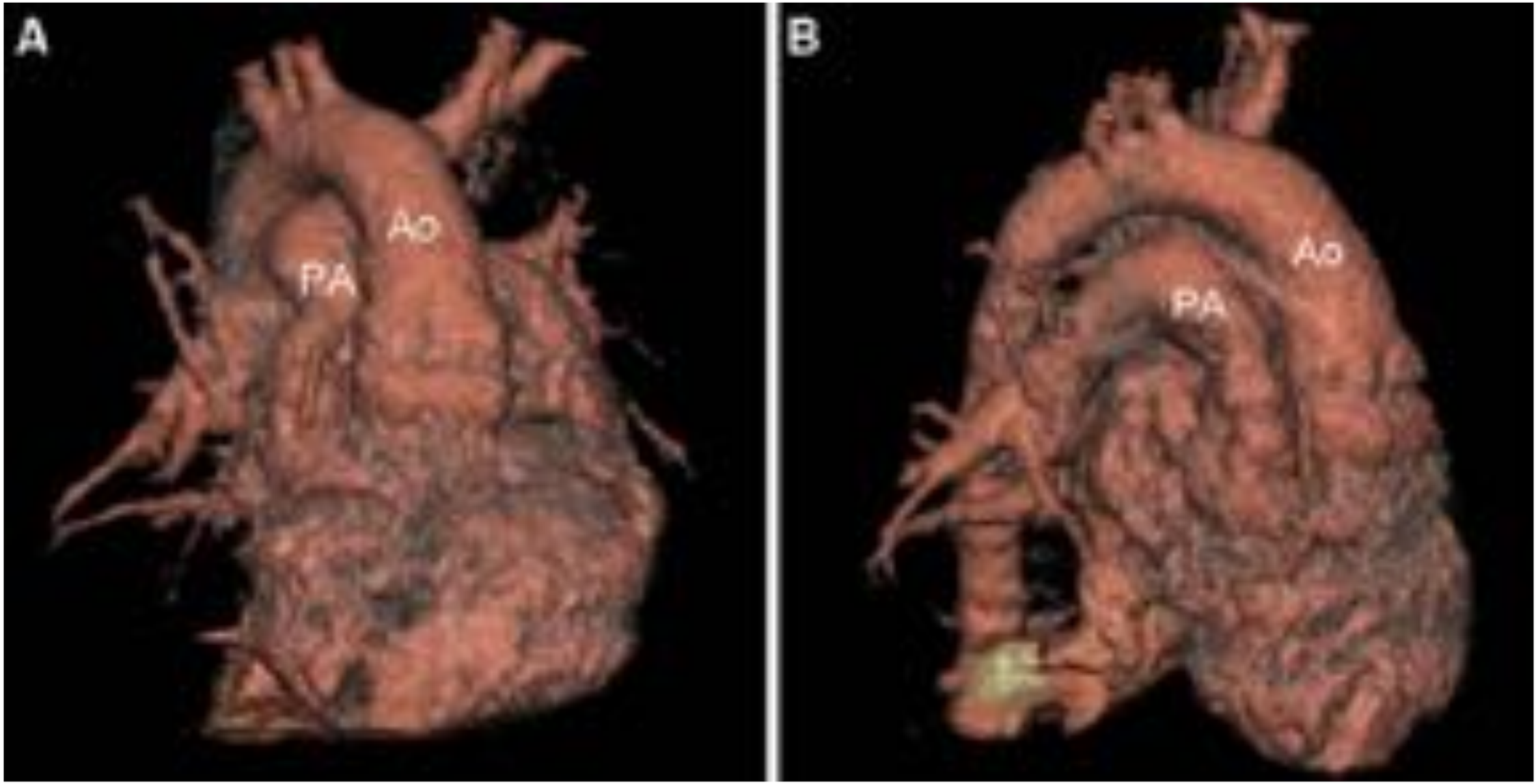
Pediatr Cardiol (2008) 29:467–468
DOI 10.1007/s00246-007-9080-4

IMAGES IN PEDIATRIC CARDIOLOGY

Anatomically Corrected Malposition of the Great Arteries

Ming-Ren Chen




A
Ao
PA
B
Ao
PA

Anatomically corrected malposition of the great arteries