Malpositions of the great arteries

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Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

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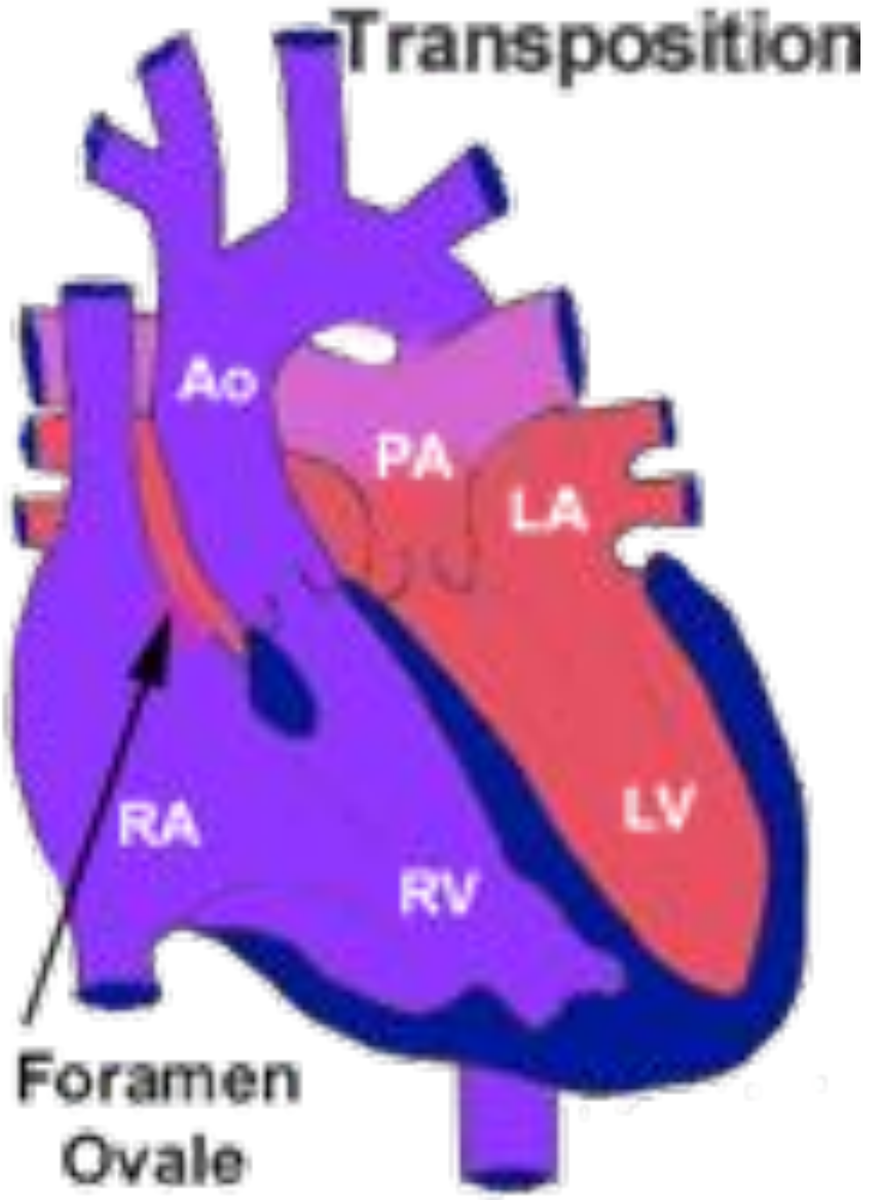
Maladies Cardiaques Héréditaires- CARDIOGEN


Transposition
Ao
PA
LA
LV
RA
RV
Foramen
Ovale

What causes transposition of the great arteries ?

Chromosomal anomalies in fetal CHD

548 CHD-18.5%

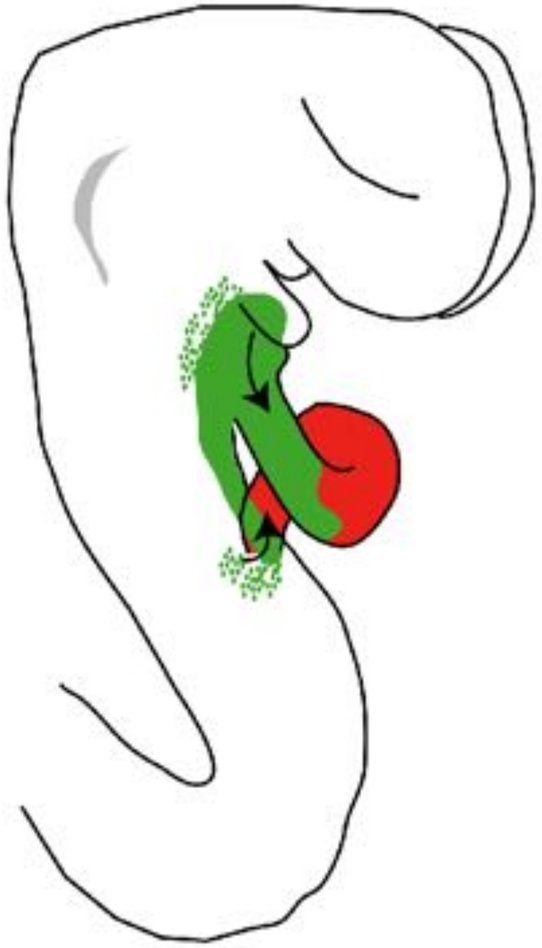
PA-IVS and PS	0	0
Left heart obstruction	12/130	9.2%
6 XO; 3 T18; 3 translocations		
Conotruncal defects	23/91	25%
20 del22q11; 1 T21; 2 translocations		
AVSD	32/68	47%
28 T21; 3 T18; 1 XXX		
VSD	12/74	16%
9 trisomies, 2 del22q11, 1 del5		
Transposition of the great vessels	0	0
DORV	7/38	18%

New paradigm for early cardiogenesis

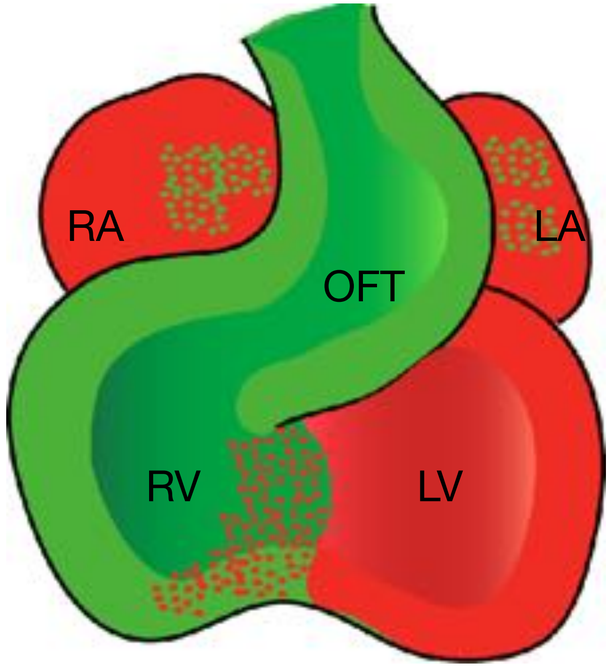
E7.5

Cardiac crescent
Second heart field

E8.5

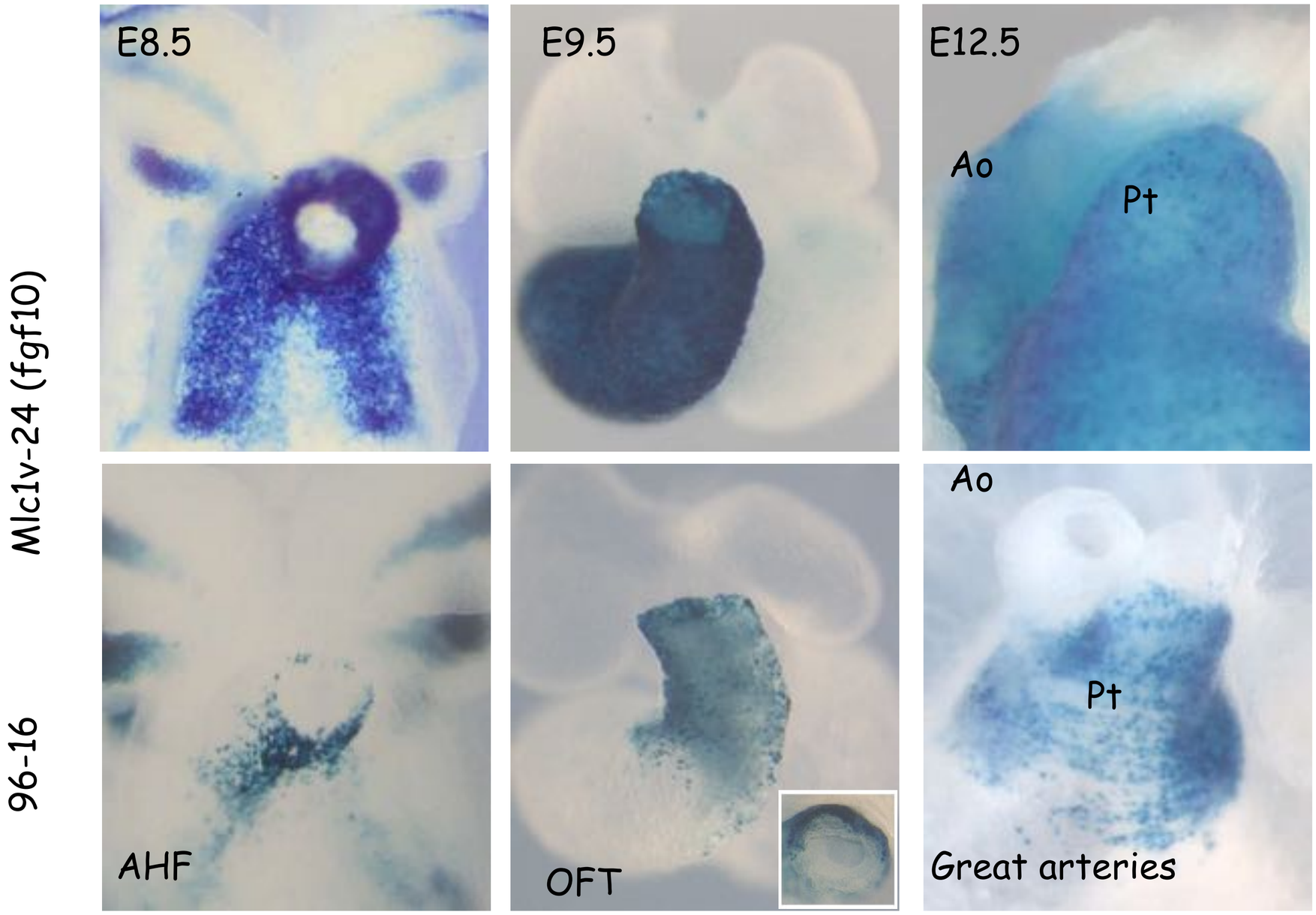


E10.5

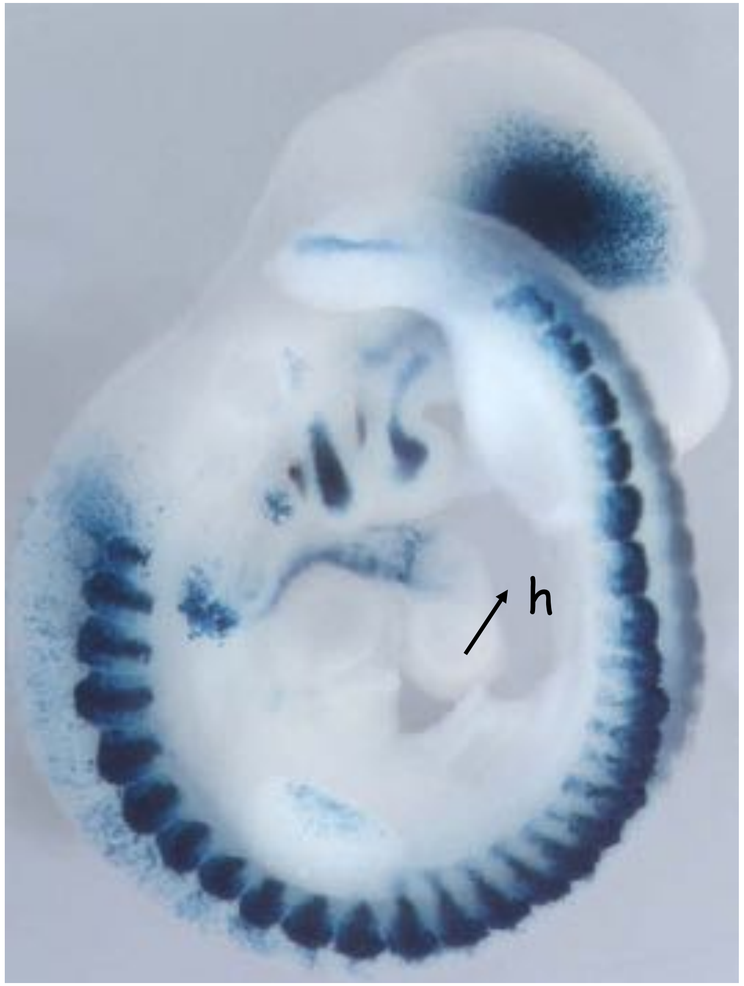


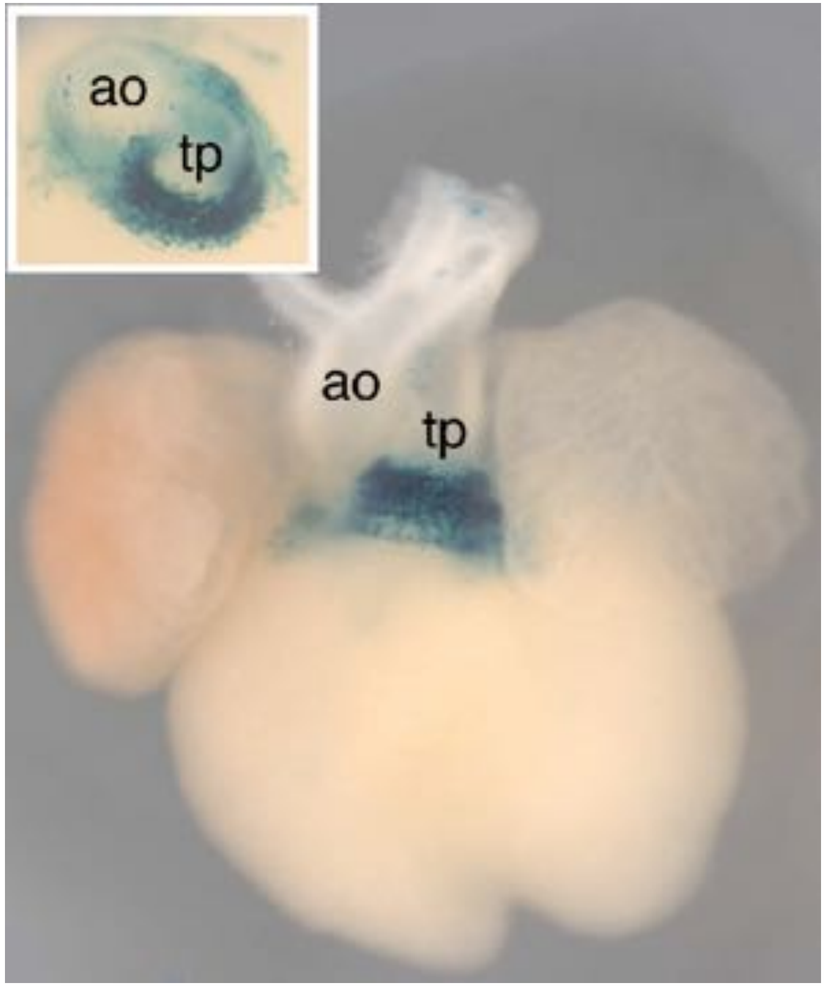
Left ventricle
Outflow tract
RA, LA, RV

Regionalization in the AHF and outflow tract

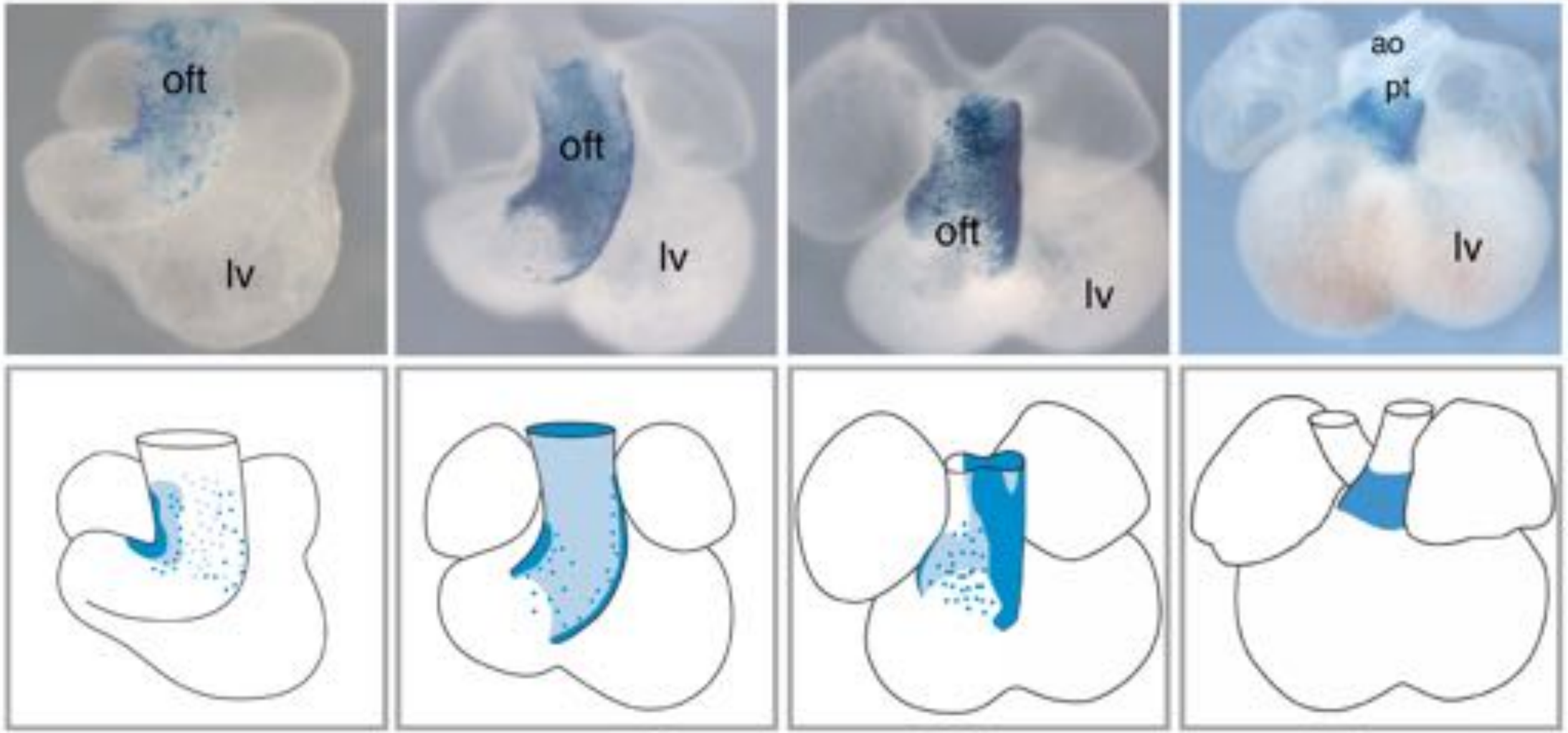


The 96-16 cardiosensor line provides a marker of myocardium at the base of the pulmonary trunk



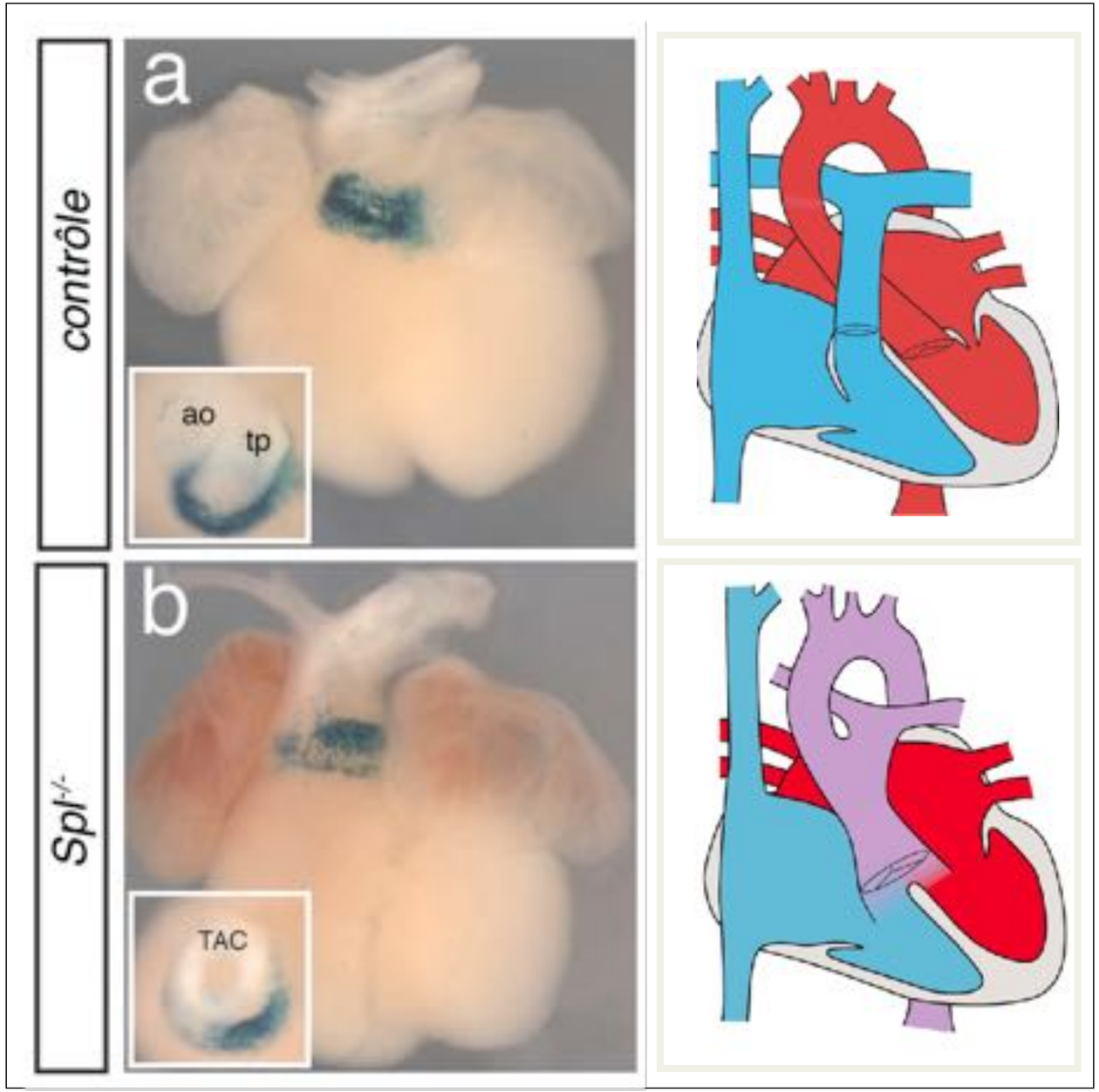


96-16 provides direct evidence for rotation of outflow tract myocardium



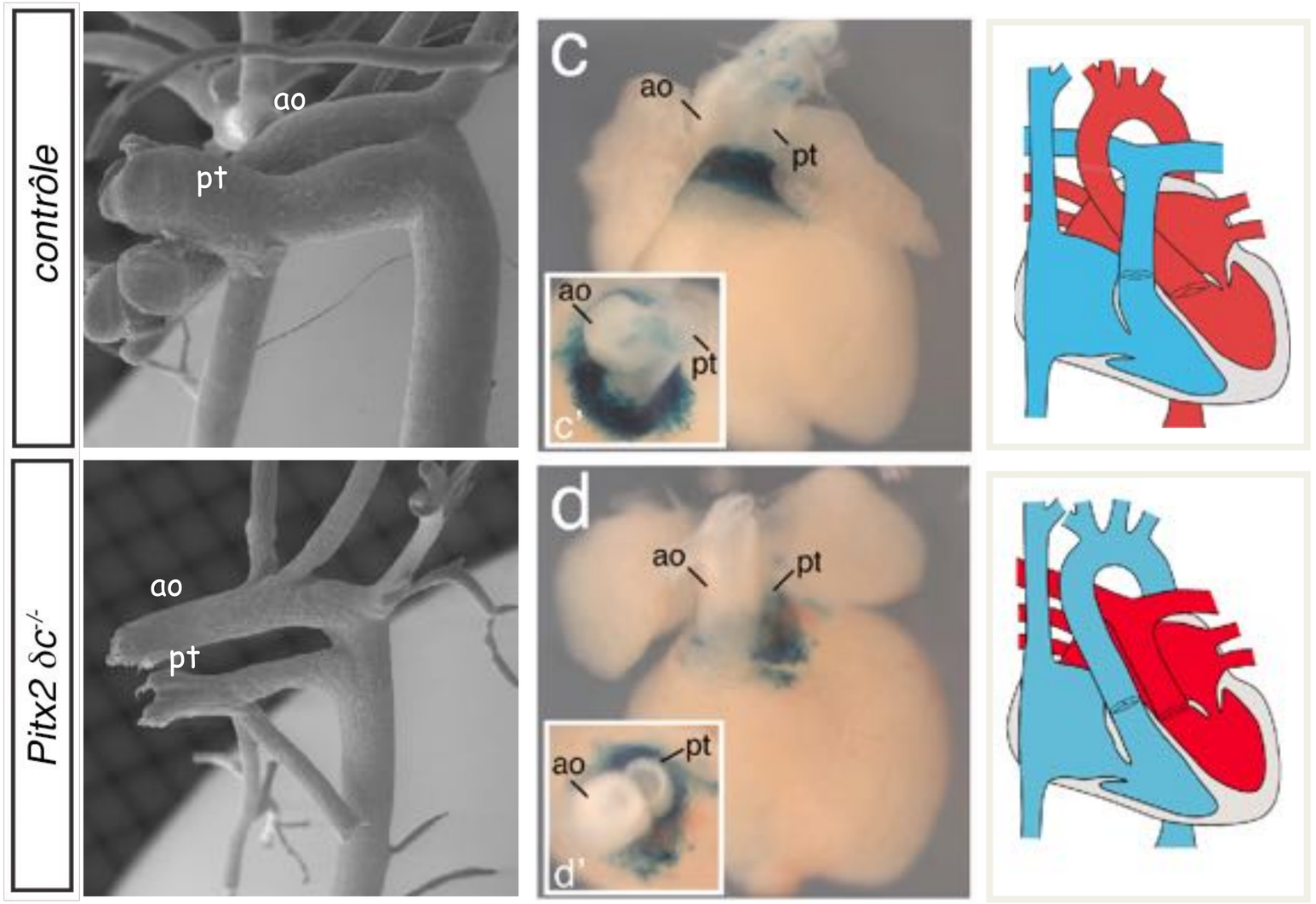
96-16 expression in Splotch heart with PTA

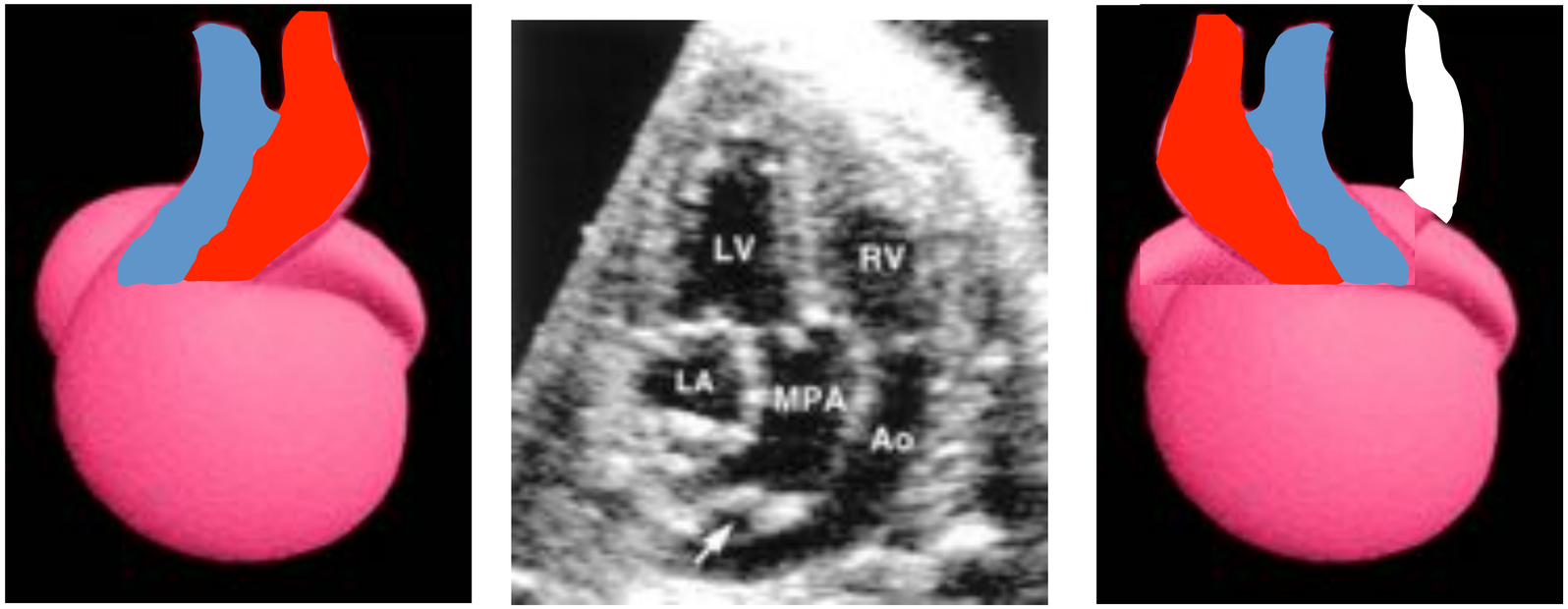
Persistent truncus arteriosus with a rotation defect
- in addition to a septation defect
- With abnormal neural crest cell migration

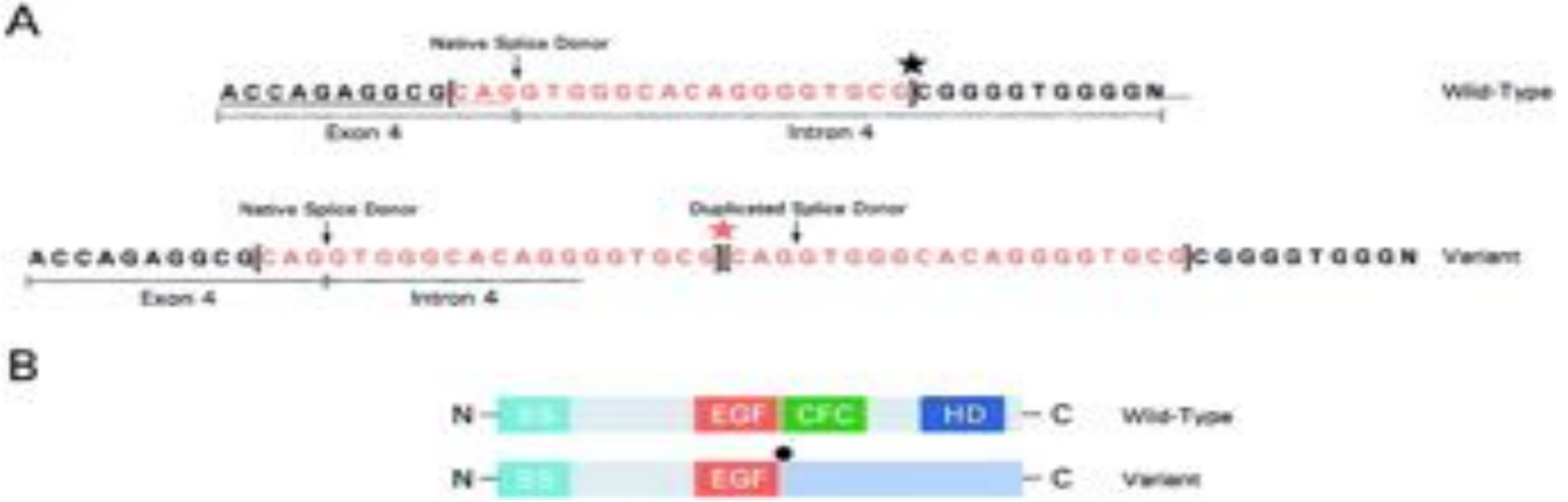


96-16 expression in Pitx2δc heart with TGA

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

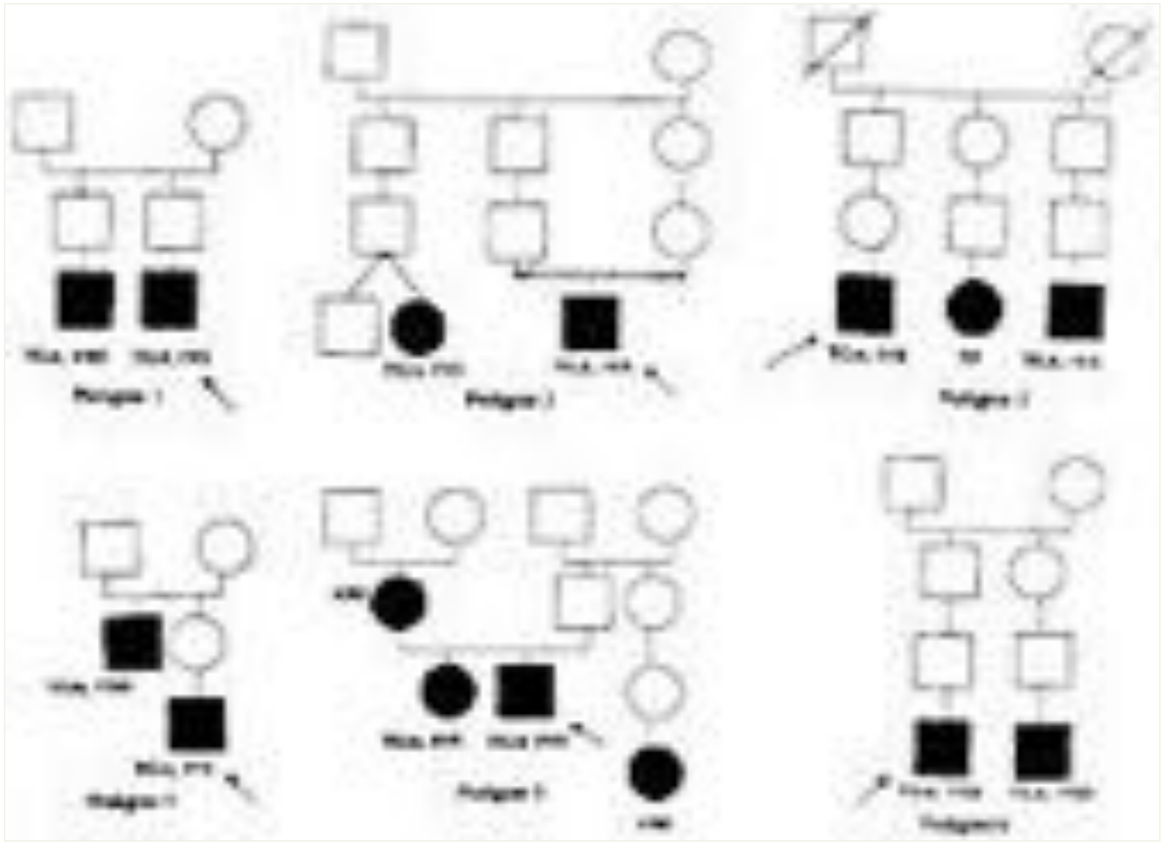




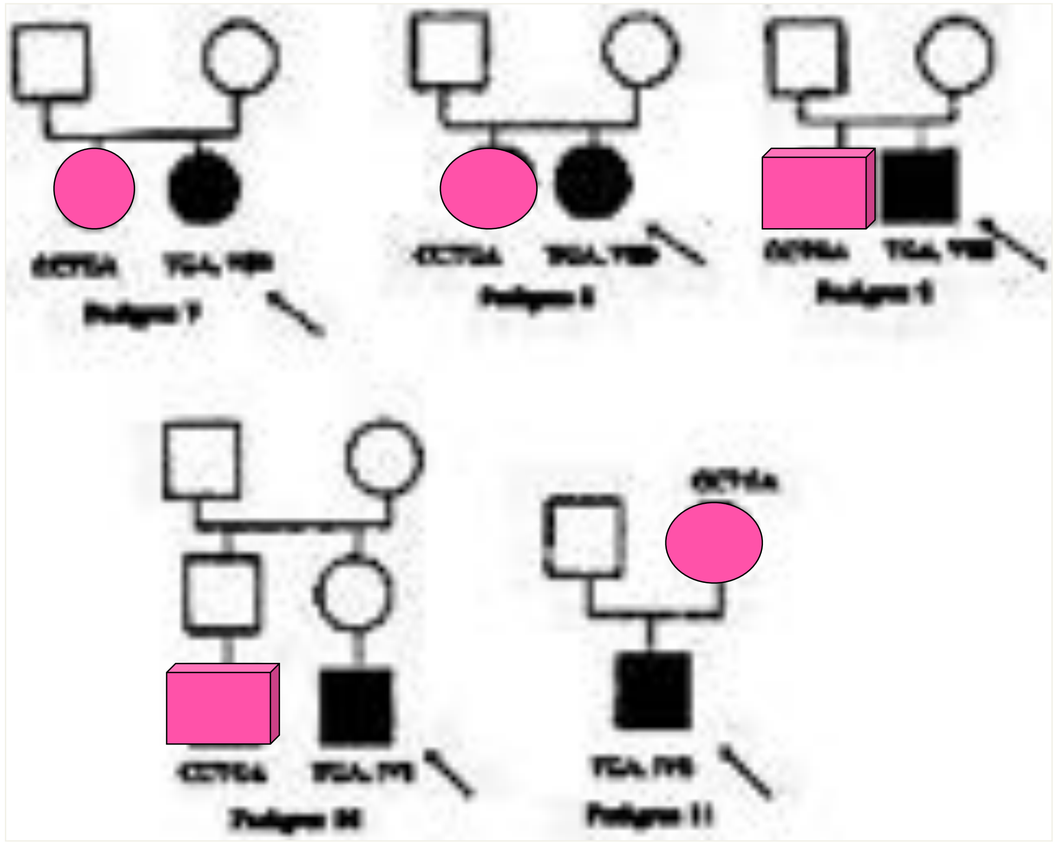


CFC1 mutations and TGA/DORV

ZIC3 mutations in TGA



TGV pedigrees



TGV (black) & DD (pink) pedigrees

Foetal TGA

The French system during pregnancy

3 systematic foetal echographies - Level 1

11 Weeks

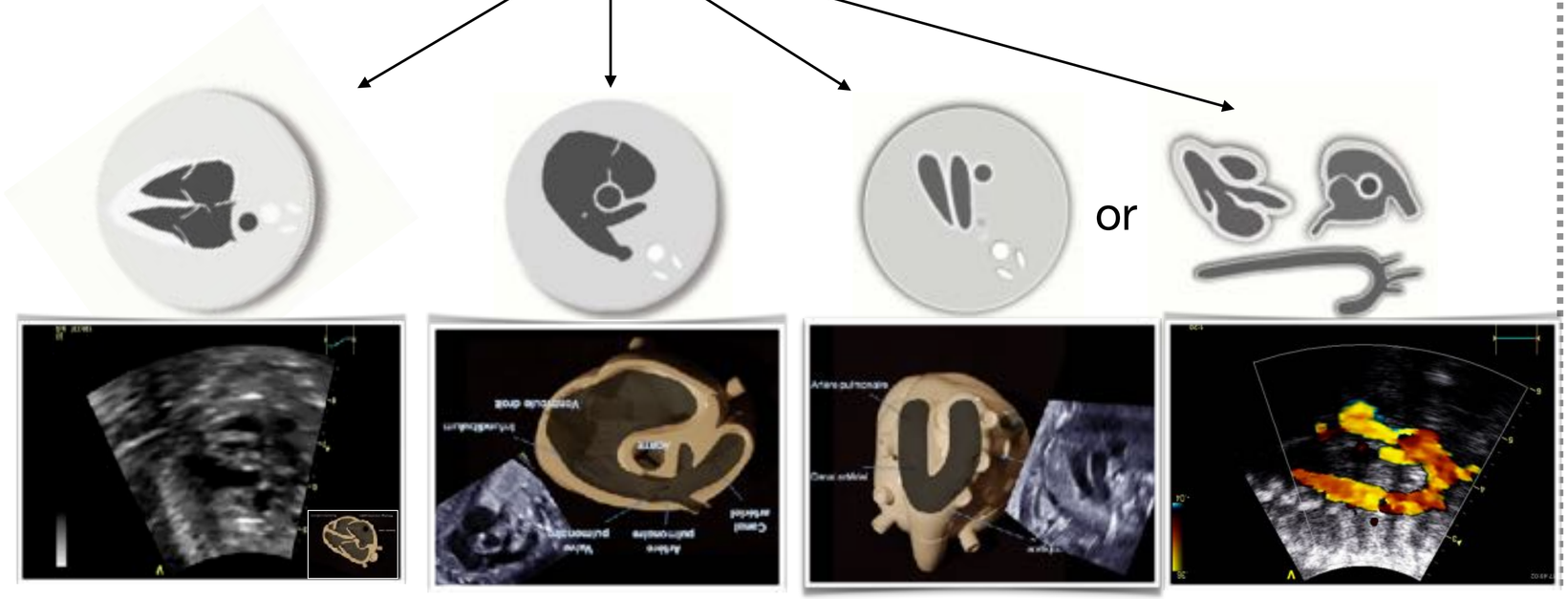
Nuchal translucency
Term
Number fetuses

18-22 Weeks

Morphology

32-34 Weeks

Growth
Malformations



In case of anomaly
or difficulty in assessing
normality

Level 2

Expert foetal
echography

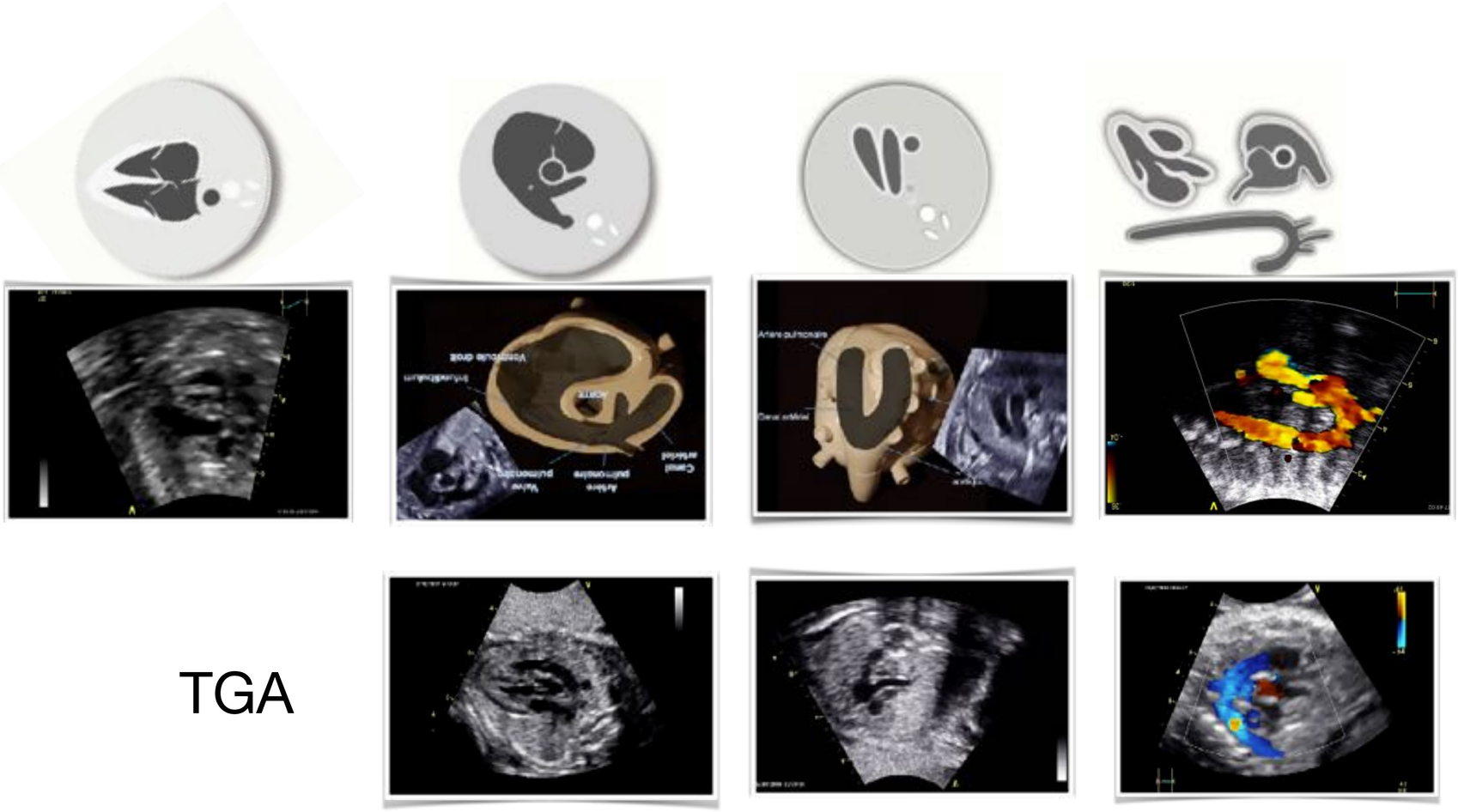
If heart anomaly is confirmed

↓

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

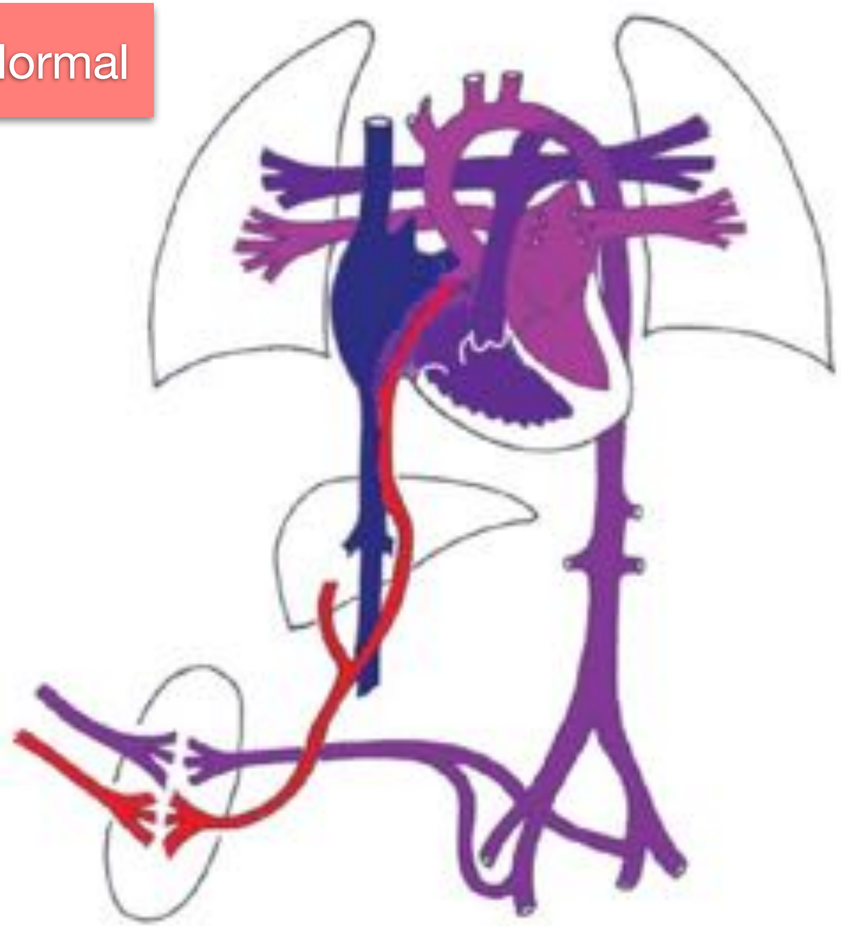
	Postnatal Group	Prenatal Group	*P*
Isolated TGA	304	57	NS
Associated defects	46	11	NS
VSD	31	8	NS
VSD + CoA	14	3	NS
CoA	1	1	NS
Age at admission, h	23 ± 216	2.2 ± 2.8	<0.01
Mechanical ventilation	95 (28)	12 (17.6)	<0.01
Metabolic acidosis + MOF	52	6	<0.05
PGE, infusion	95	22	NS
ISAS	168	54	NS
Preoperative mortality	15	0	<0.05
Coronary artery pattern	222 400	68 400	
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; NIC, balloon atrioseptostomy; and ASO, arterial switch operation. Values are n (%).

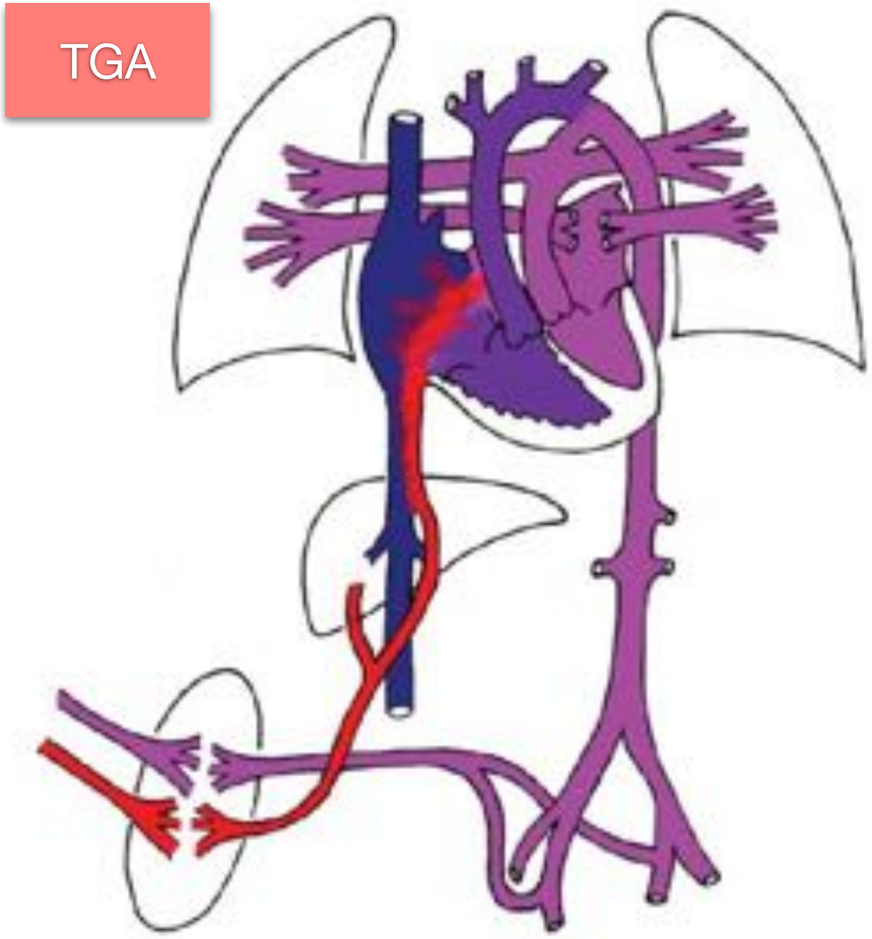
Bonnet D et al. Circulation 1997

Conséquences développementales de la répartition du débit sanguin foetal combiné

Normal

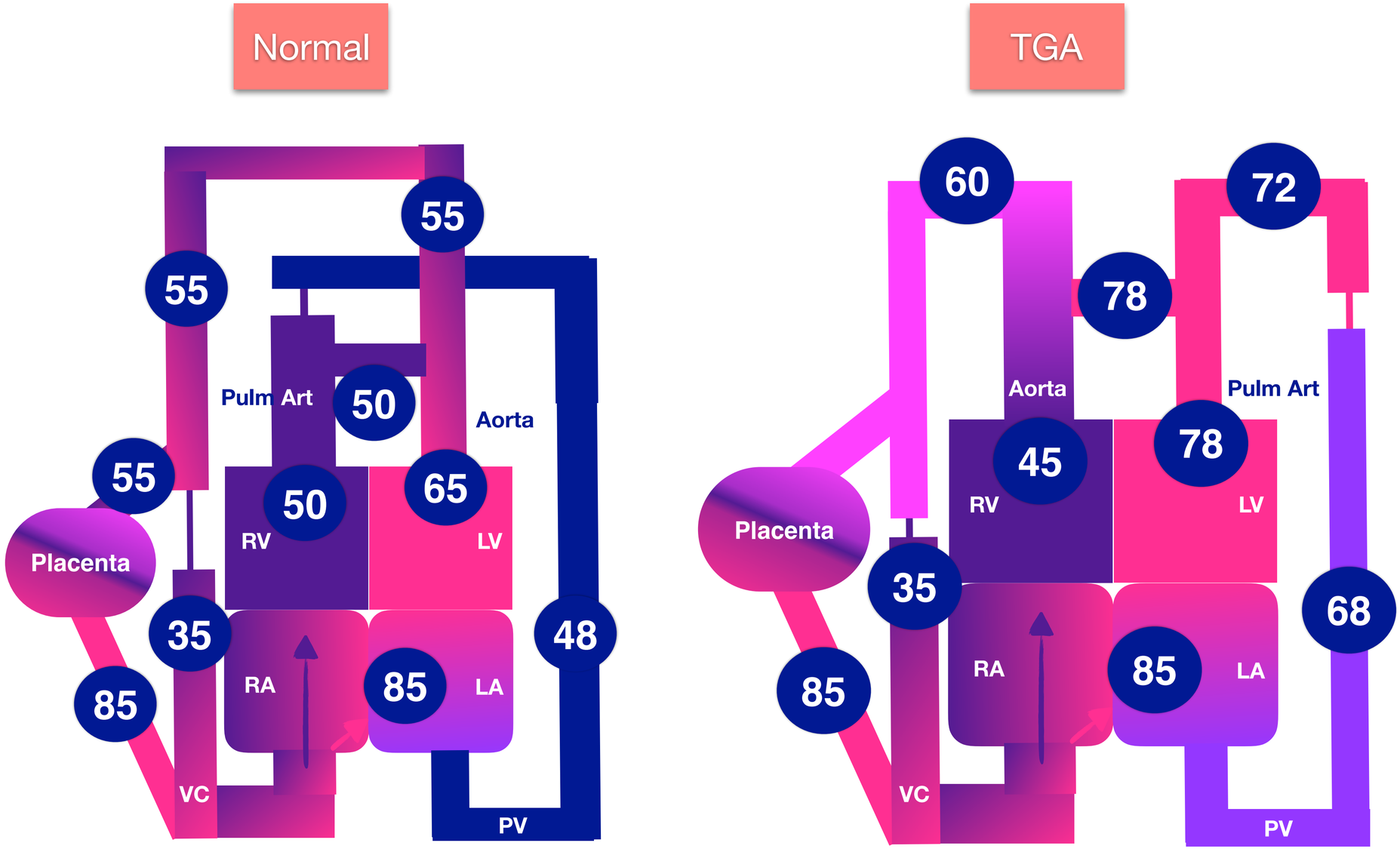


TGA



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

Conséquences développementales de la répartition du débit sanguin foetal combiné

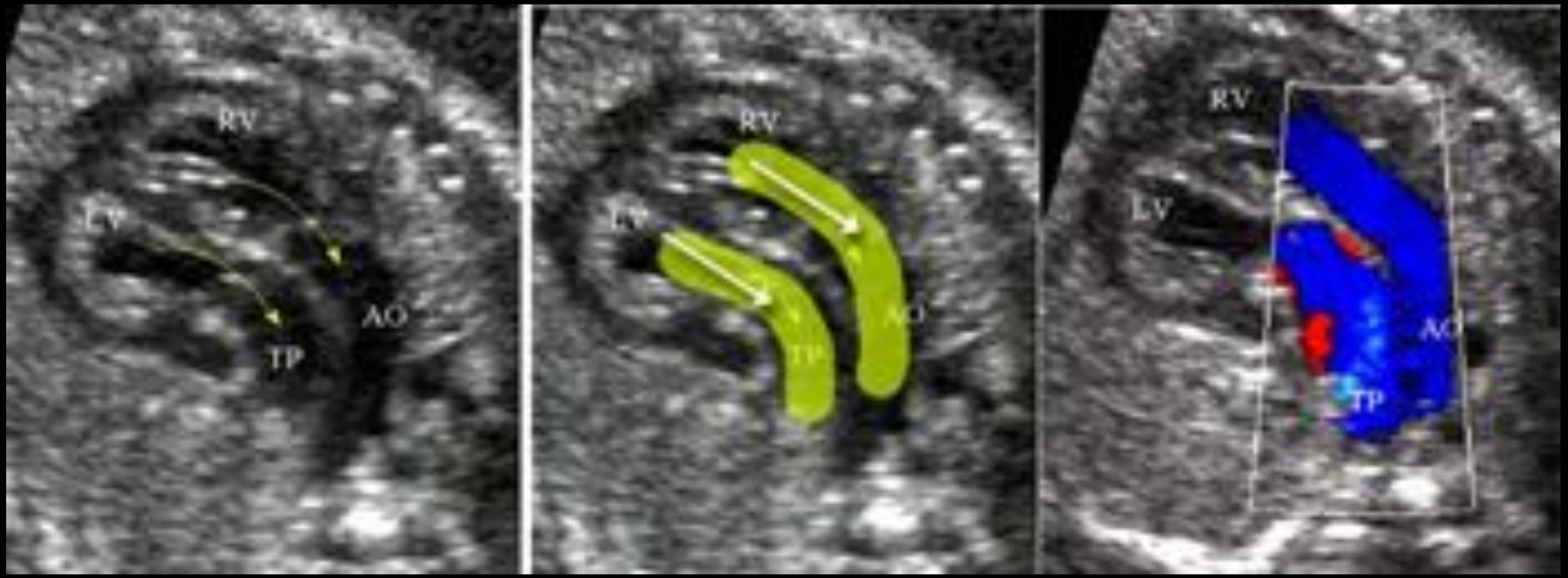


Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

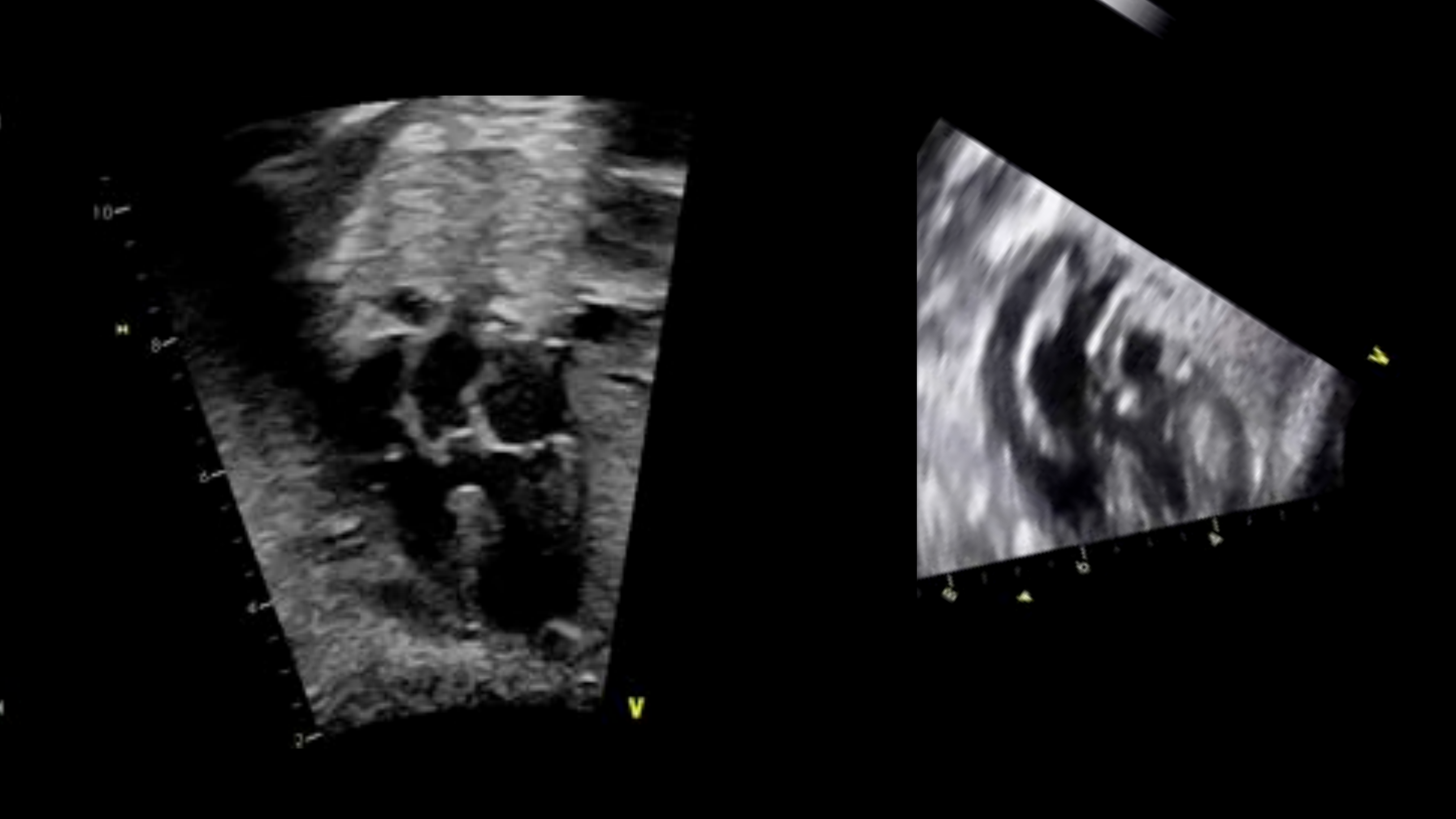
Prenatal diagnosis, pregnancy termination, perinatal and early neonatal mortality for selected (isolated) congenital heart anomalies

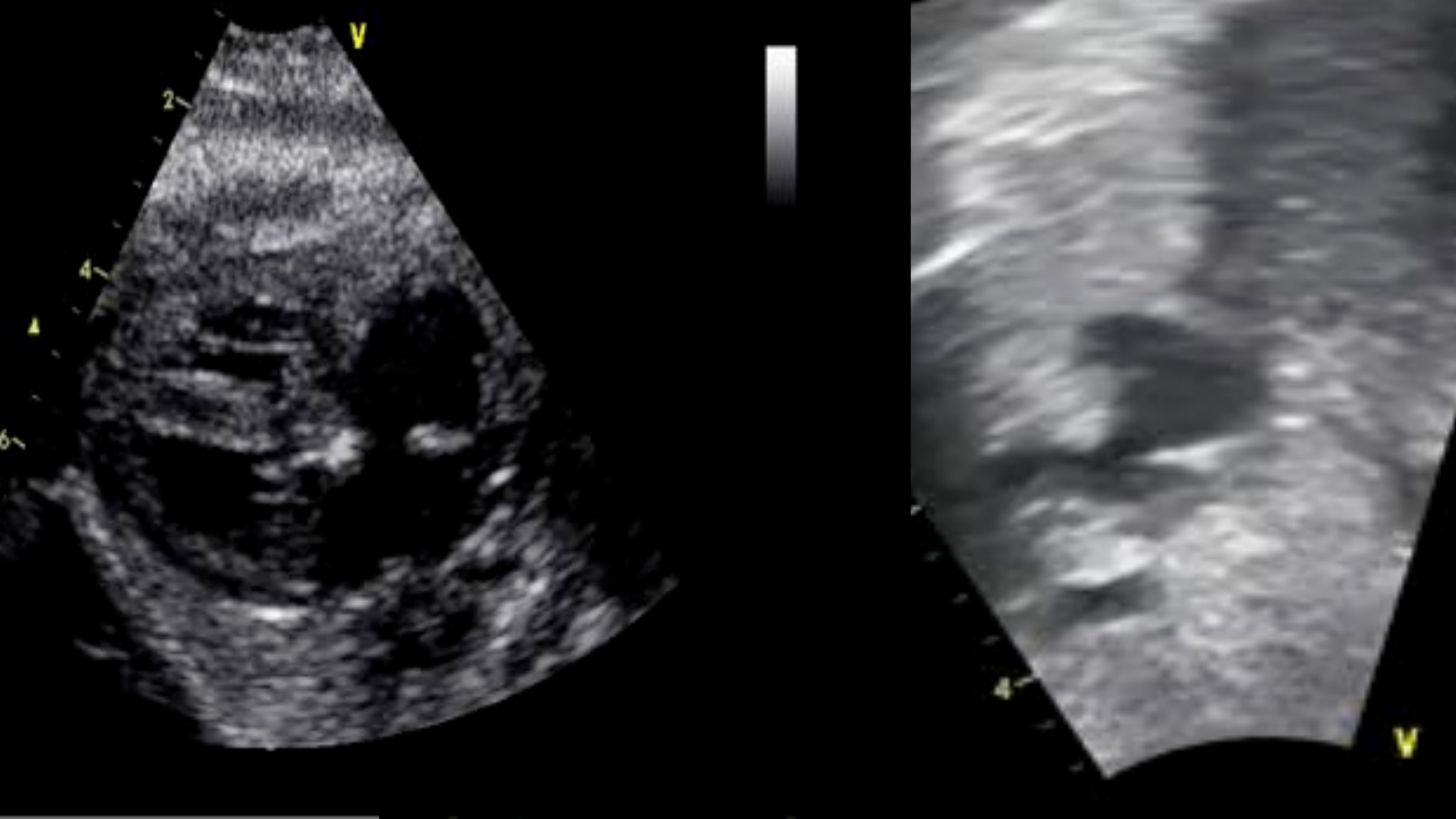
Paris Registry of Congenital Malformations, 1983-2000

	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


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

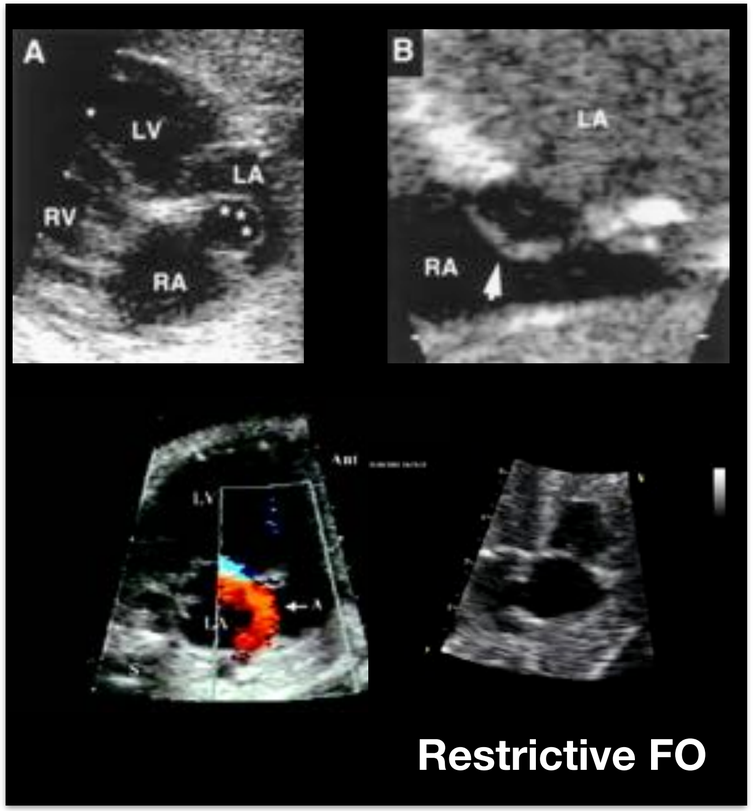

4
6
8



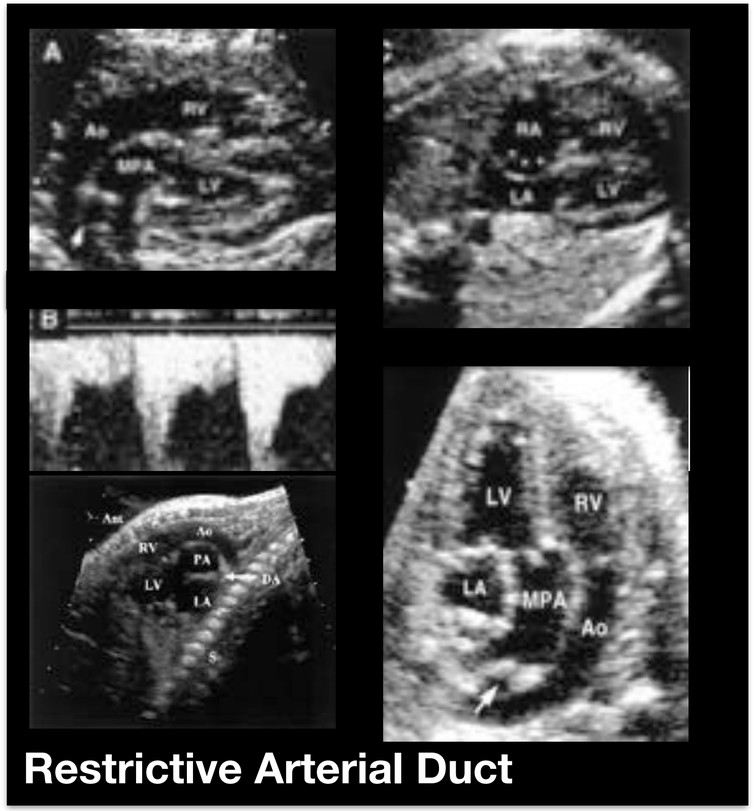

V
2
4
6
4
V

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

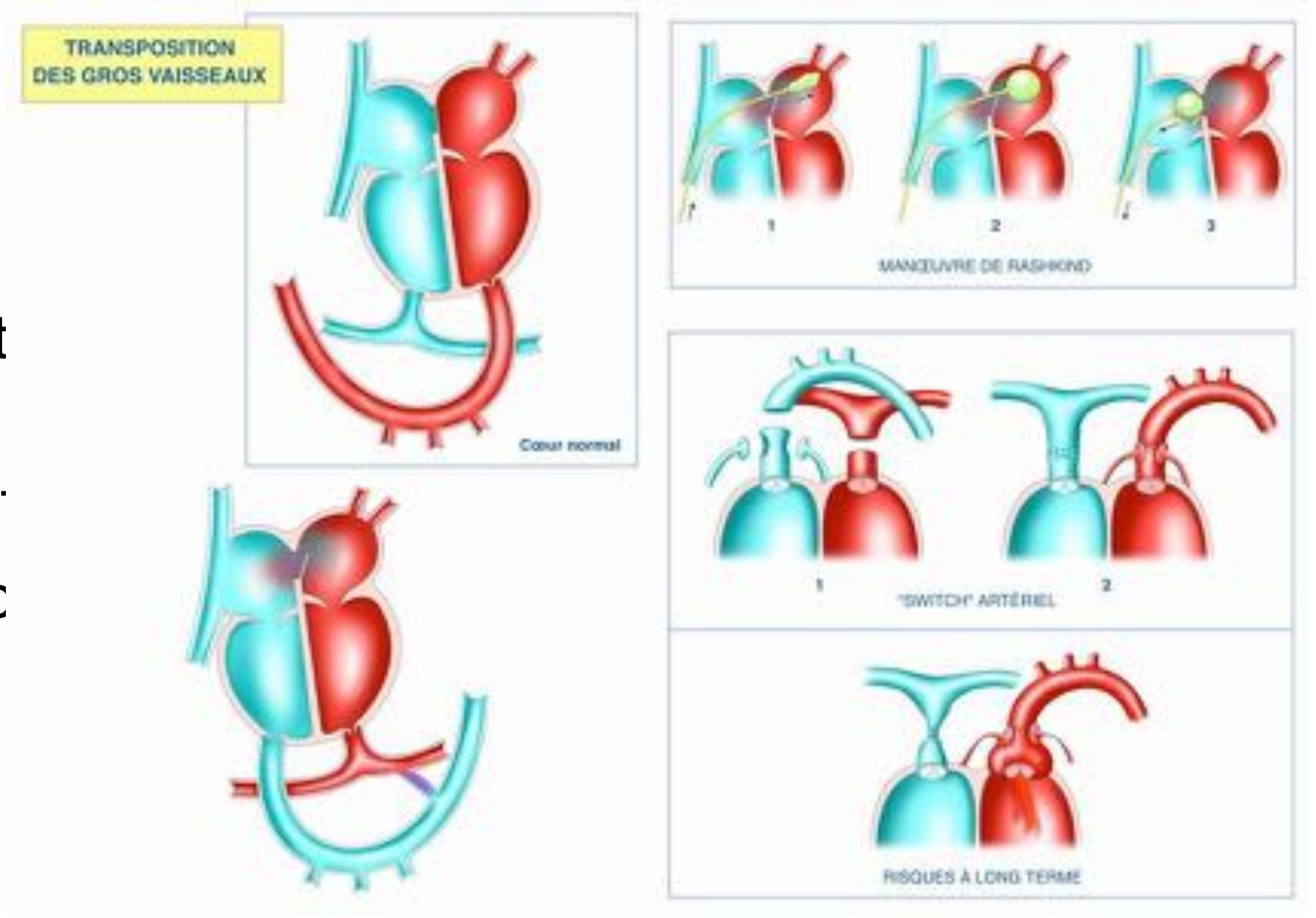
Finally, we do not care
All TGA are delivered on site with the same protocol

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 of transposition of the great arteries

Perinatal organization in Paris

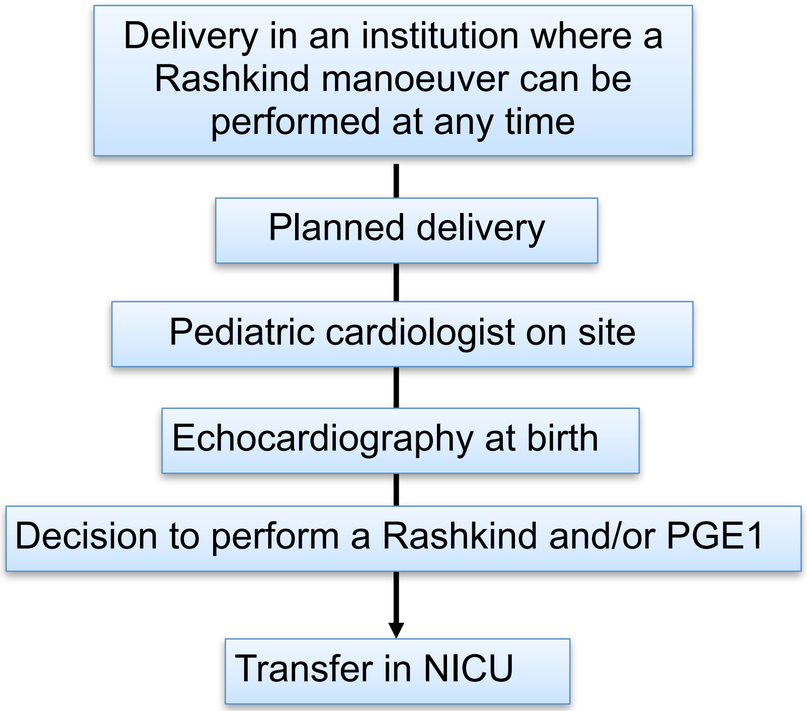
- Organisation of foetal cardiac growth surveillance
  - Foramen ovale and arterial duct
- *In utero* transfer organisation
- Organisation of perinatal management
- Prevention of early neonatal demise
- Prepare the parents to the future even
- Post-natal management and follow-up



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

Prenatal diagnosis of transposition of the great arteries

Perinatal organization



From 1992 to 217

717 prenatally diagnosed TGA (IVS or complex)

6 had congenitally corrected TGA

3 deaths immediately after birth in the delivery room

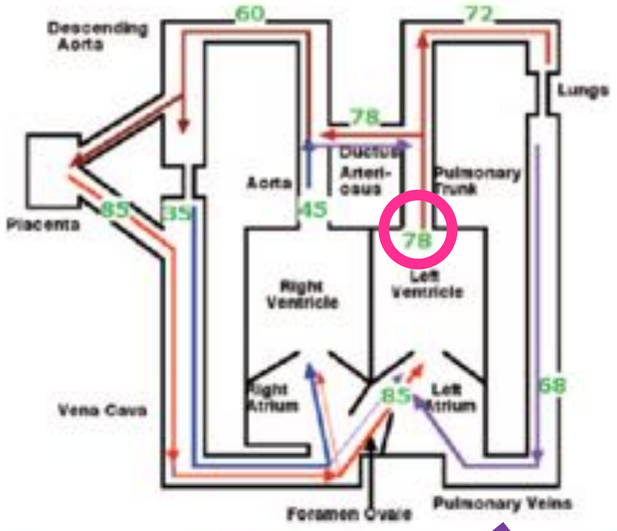
3 additional preoperative deaths

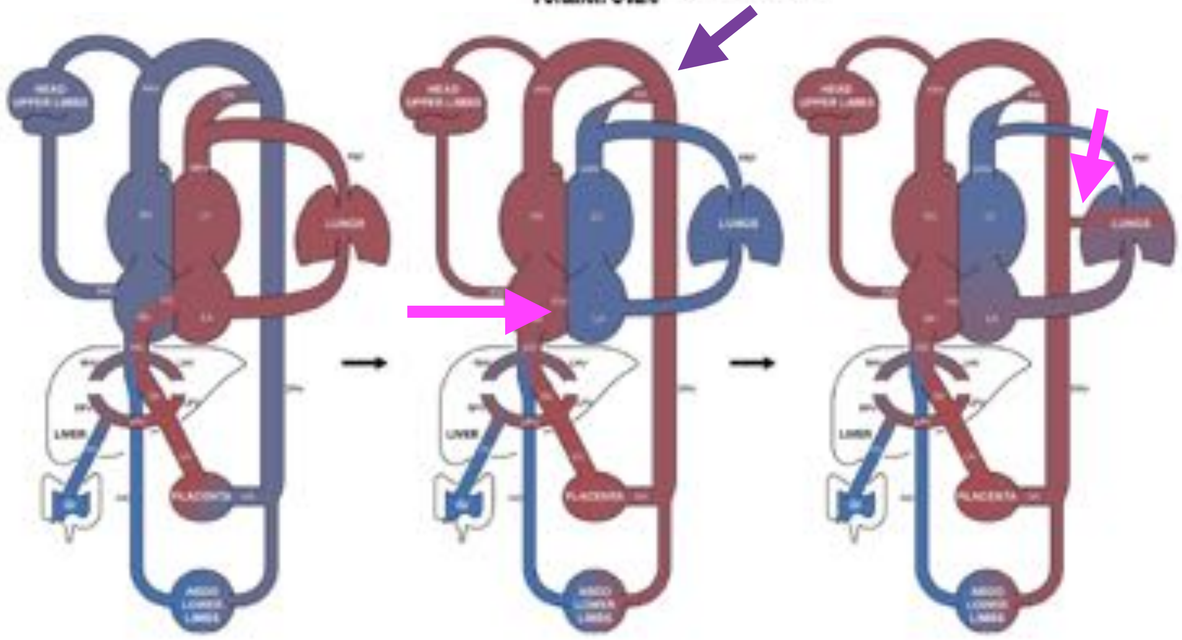
1 extra cardiac malformation in a CHARGE syndrome,
1 necrotizing enterocolitis
1 during the Rashkind procedure (perforation of the left atrium in left juxtaposition of the atrial appendages)

Surgical mortality 1.7 % : 693 survivors at discharge

Prenatal diagnosis of transposition of the great arteries

Fetal physiology - risk of increased PVR - Aorto-pulmonary collaterals





Initial increase in PBF

Reduced size of the FO

Ductal constriction

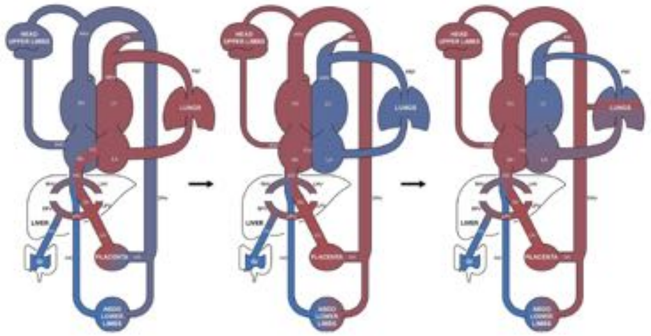
Isolation of Pulmonary circulation

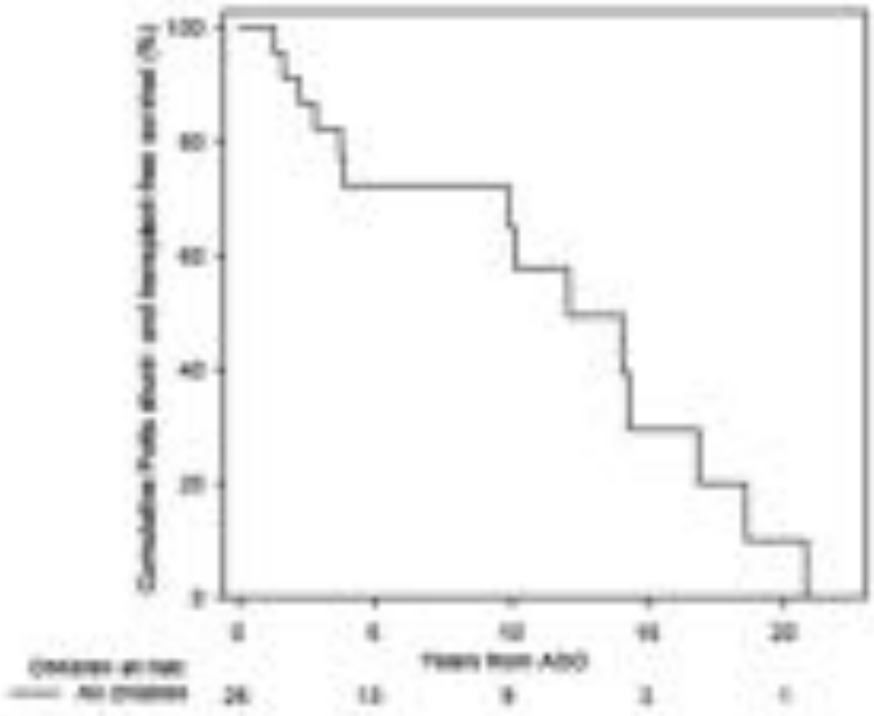
Increased PVR

Development of aorta-pulmonary collaterals

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

Pulmonary arterial hypertension in TGA



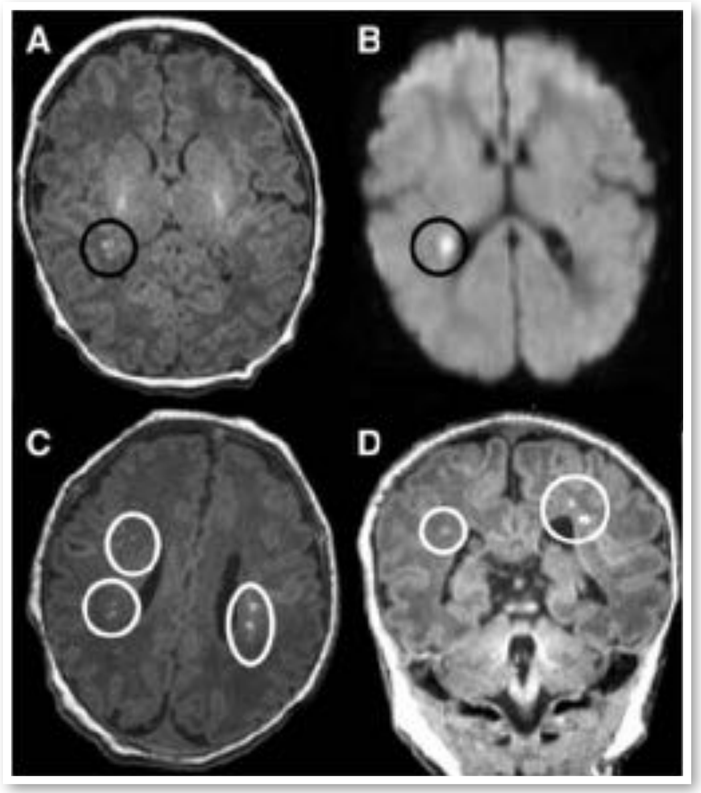


Zijlstra WM et al. Heart 2017

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

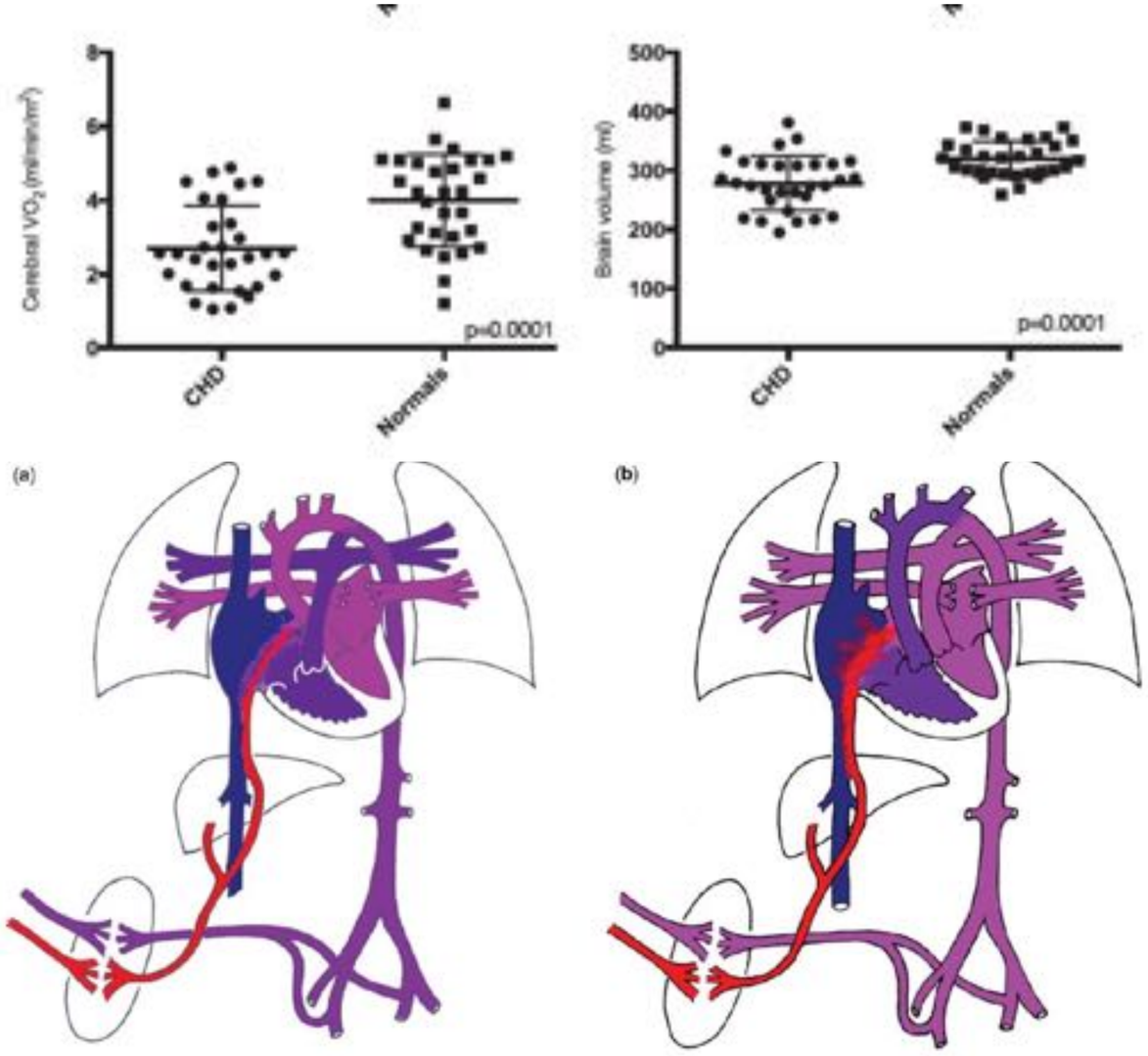
- **White matter lesions in** 30 to 40% of newborns with TGA (Miller et al., 2004; Licht et al., 2009)

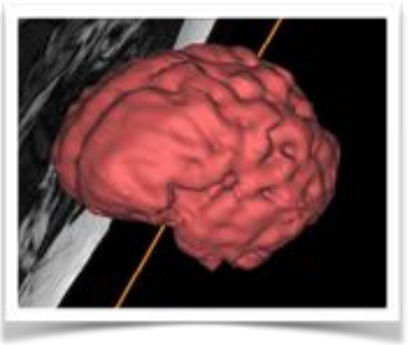
- Same type of anomalies but more severe in complex CHDs such as HLHS (Mahle et al., 2002).



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

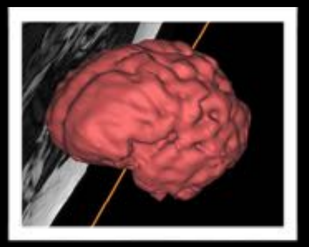




Mechanisms for reduced cerebral oxygenation and impaired brain growth in fetuses with CHD

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.

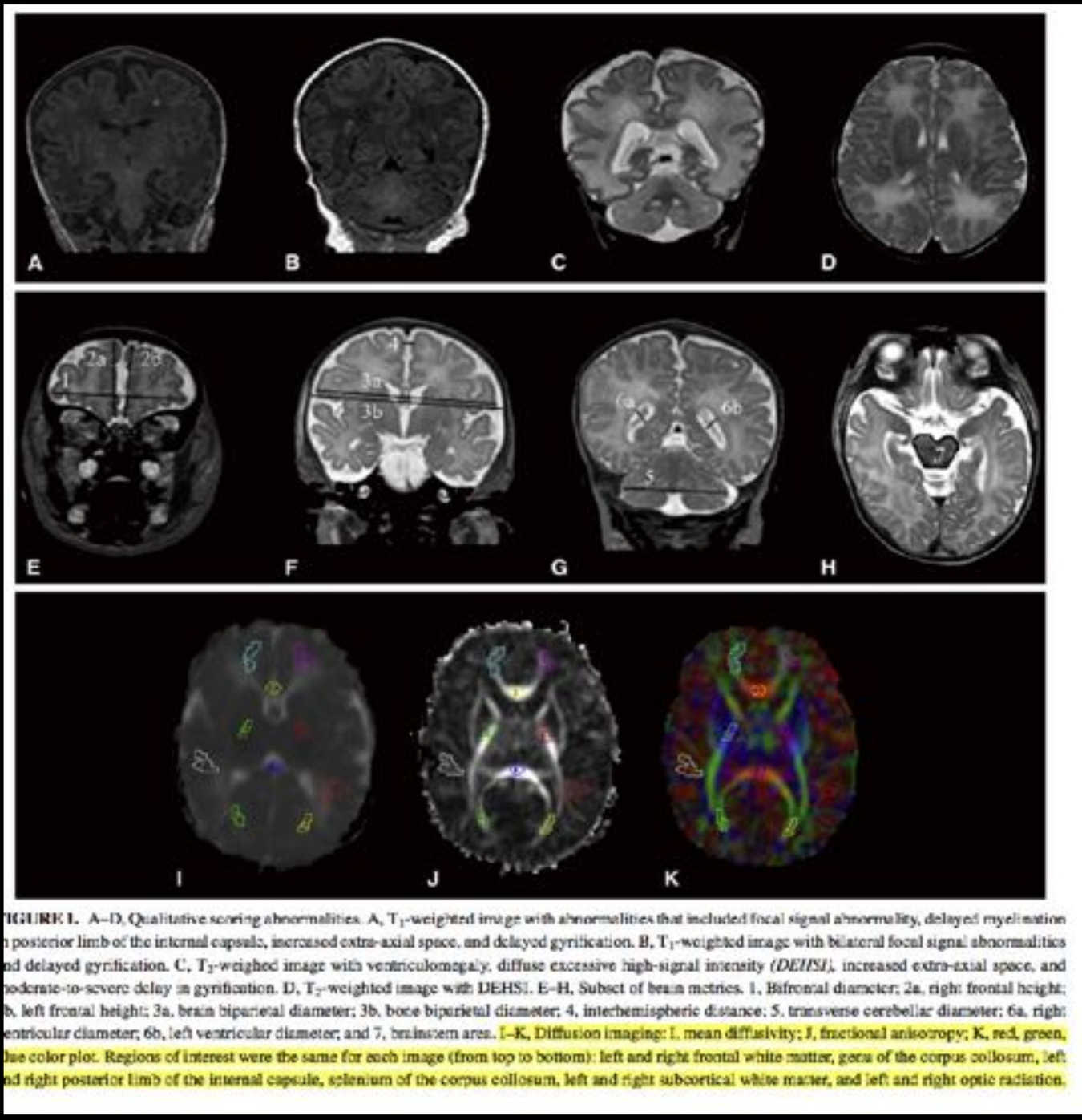


Brain dysmaturation observed in CHD appears to confer increased susceptibility to white matter injury in the perioperative period and neurodevelopmental deficits at 2 years.

The identification of fetal hypoxia as a potentially modifiable cause of delayed fetal brain development
may be clinically significant.

Oxygen saturations in the fetal sheep and human fetuses circulation can be augmented through increases in the oxygen concentration of maternal inhaled air.

Maternal hyperoxygenation could be a method to improve brain development in utero

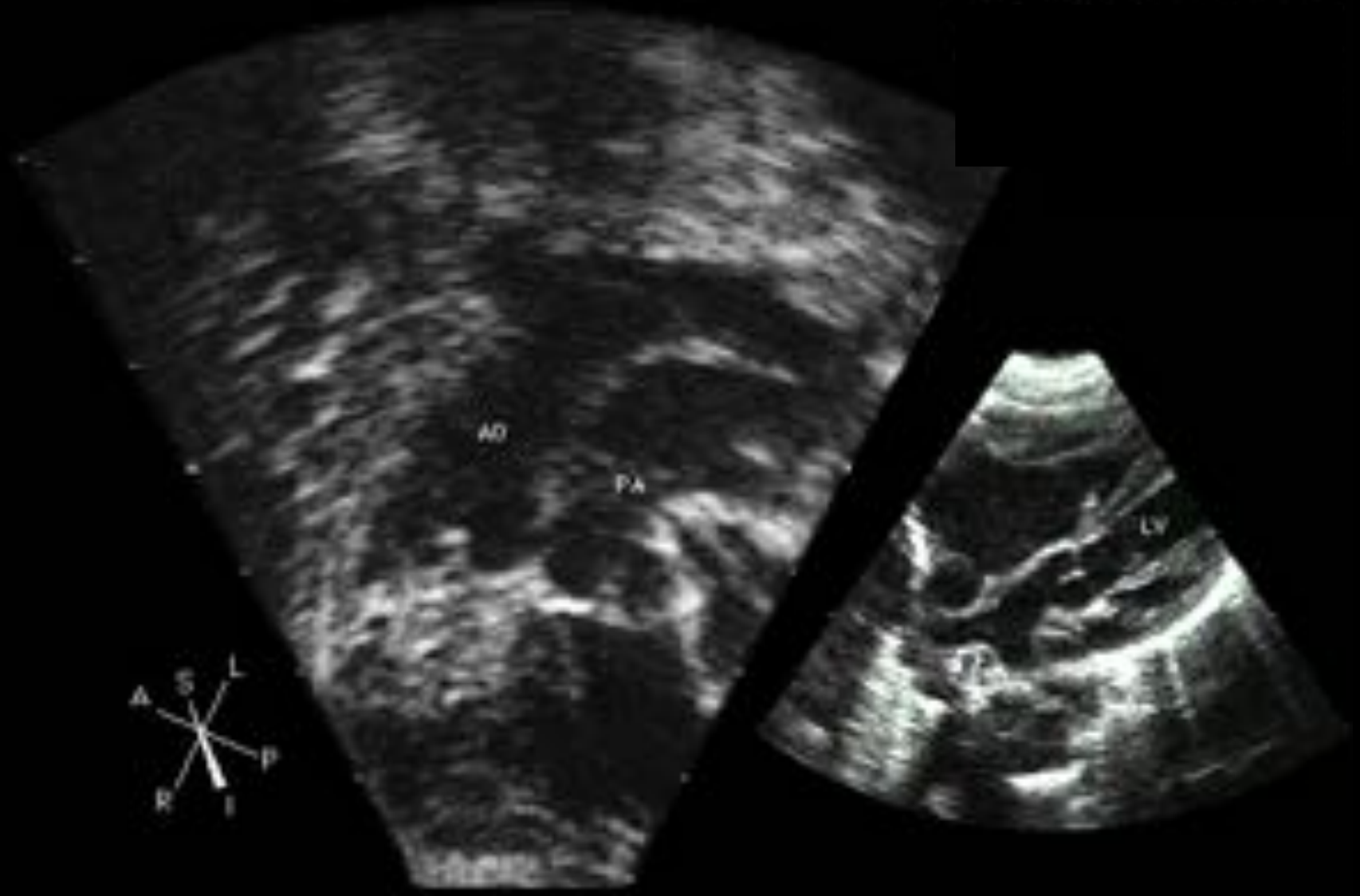


IGURE 1. A–D, Qualitative scoring abnormalities. A, T_1-weighted image with abnormalities that included focal signal abnormality, delayed myelination posterior limb of the internal capsule, increased extra-axial space, and delayed gyrification. B, T_1-weighted image with bilateral focal signal abnormalities nd delayed gyrification. C, T_2-weighed image with ventriculomegaly, diffuse excessive high-signal intensity *(DEHSI)*, increased extra-axial space, and oderate-to-severe delay in gyrification. D, T_2-weighted image with DEHSI. E–H, Subset of brain metrics. 1, Bifrontal diameter; 2a, right frontal height; b, left frontal height; 3a, brain biparietal diameter; 3b, bone biparietal diameter; 4, interhemispheric distance; 5, transverse cerebellar diameter; 6a, right entricular diameter; 6b, left ventricular diameter; and 7, brainstem area. I–K, Diffusion imaging: I, mean diffusivity; J, fractional anisotropy; K, red, green, lue color plot. Regions of interest were the same for each image (from top to bottom): left and right frontal white matter, genu of the corpus collosum, left nd right posterior limb of the internal capsule, splenium of the corpus collosum, left and right subcortical white matter, and left and right optic radiation.

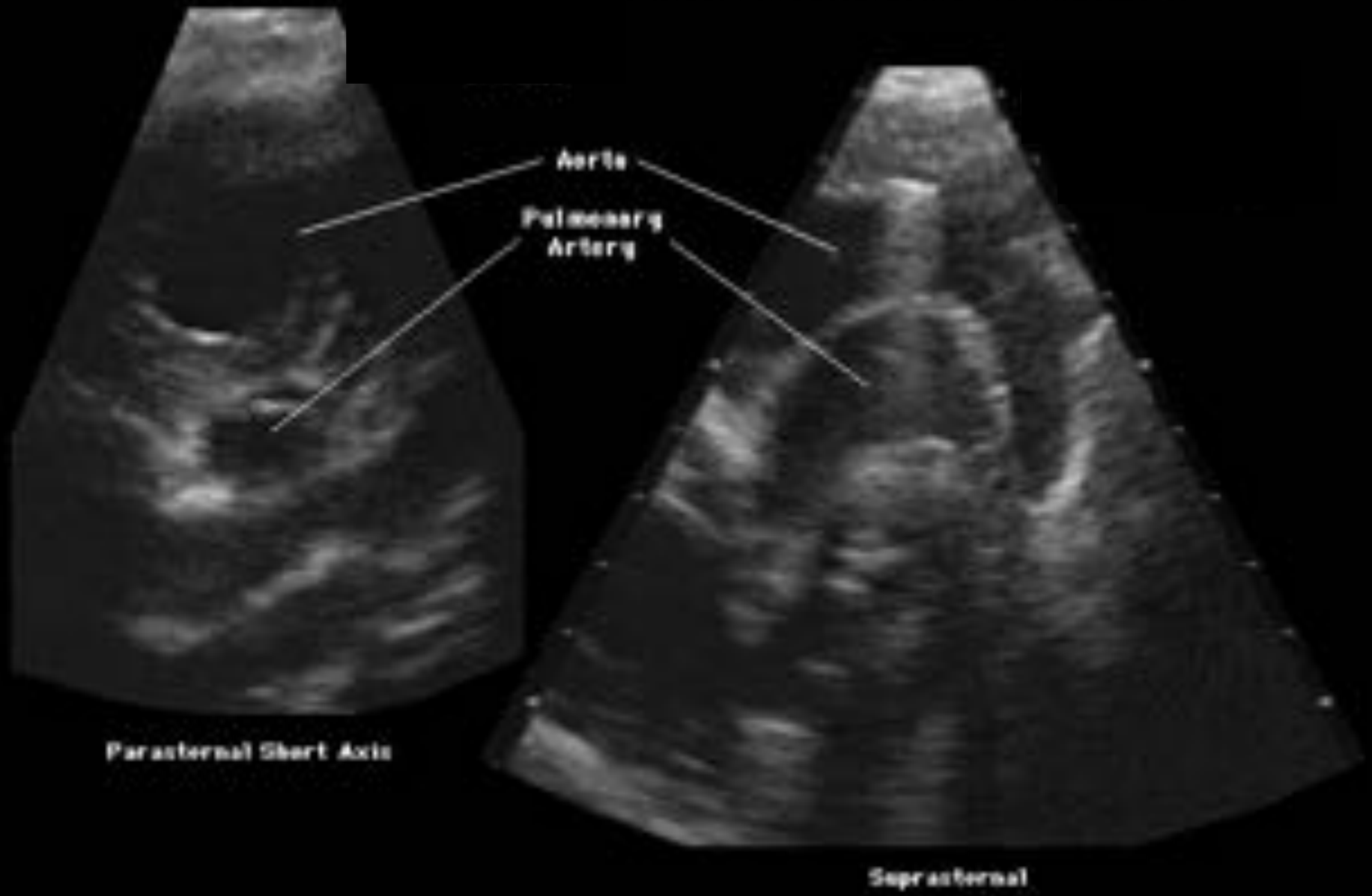
J Thorac Cardiovasc Surg 2012;143:1264-70

Neonatal diagnosis of TGA
Isolated cyanosis

d-Transposition of the Great Vessels, Subcostal View



Transposition of the Great Vessels, Vessel Anatomy




14/10/2010 12:09:24
V
2
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6
8


Pédiatrique
S12-4
44Hz
8.0cm
2D
G
P
R


V
2
4


V
AO
AP
OG
2
4
.49
-.49
115:171


V
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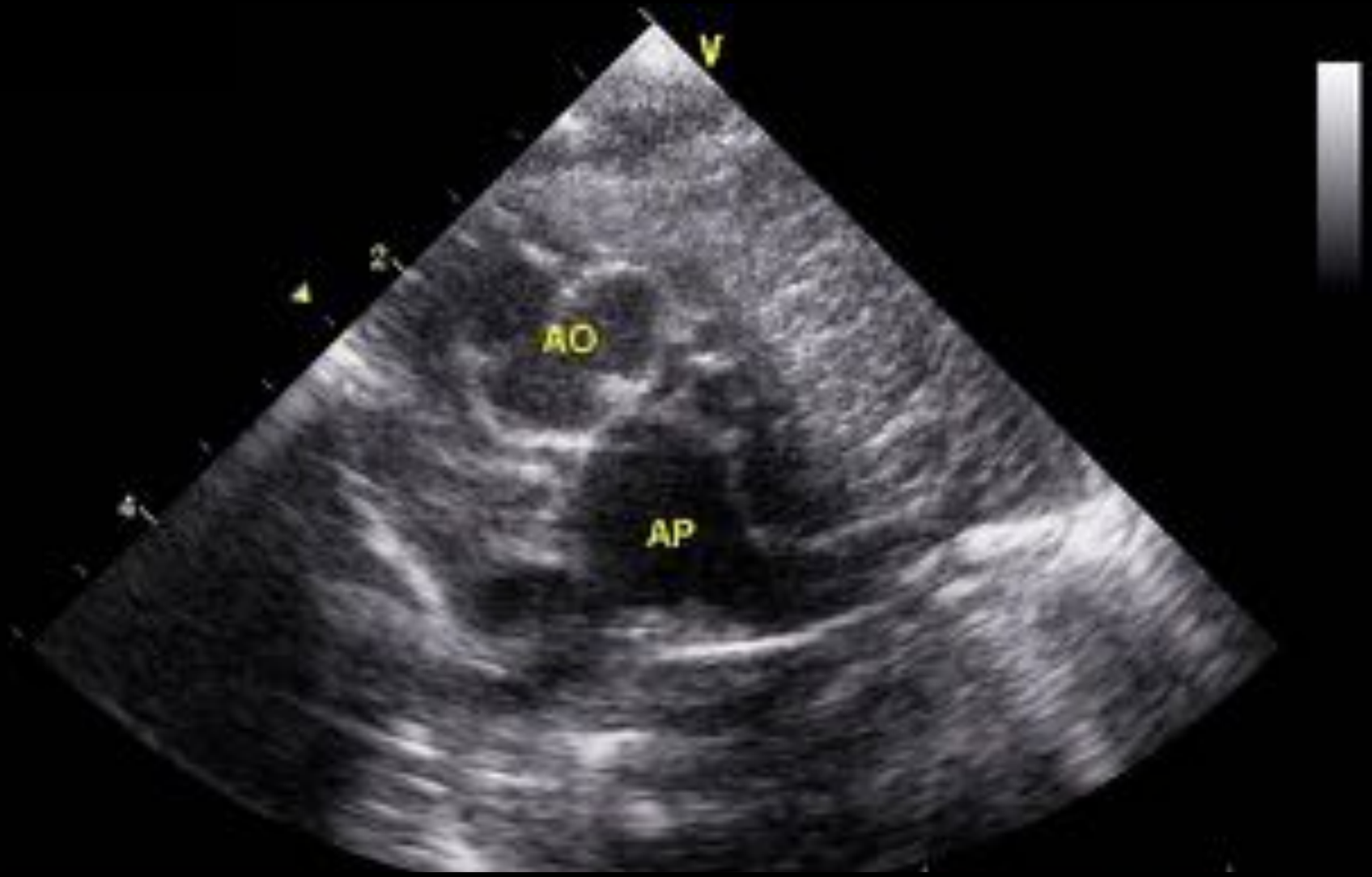
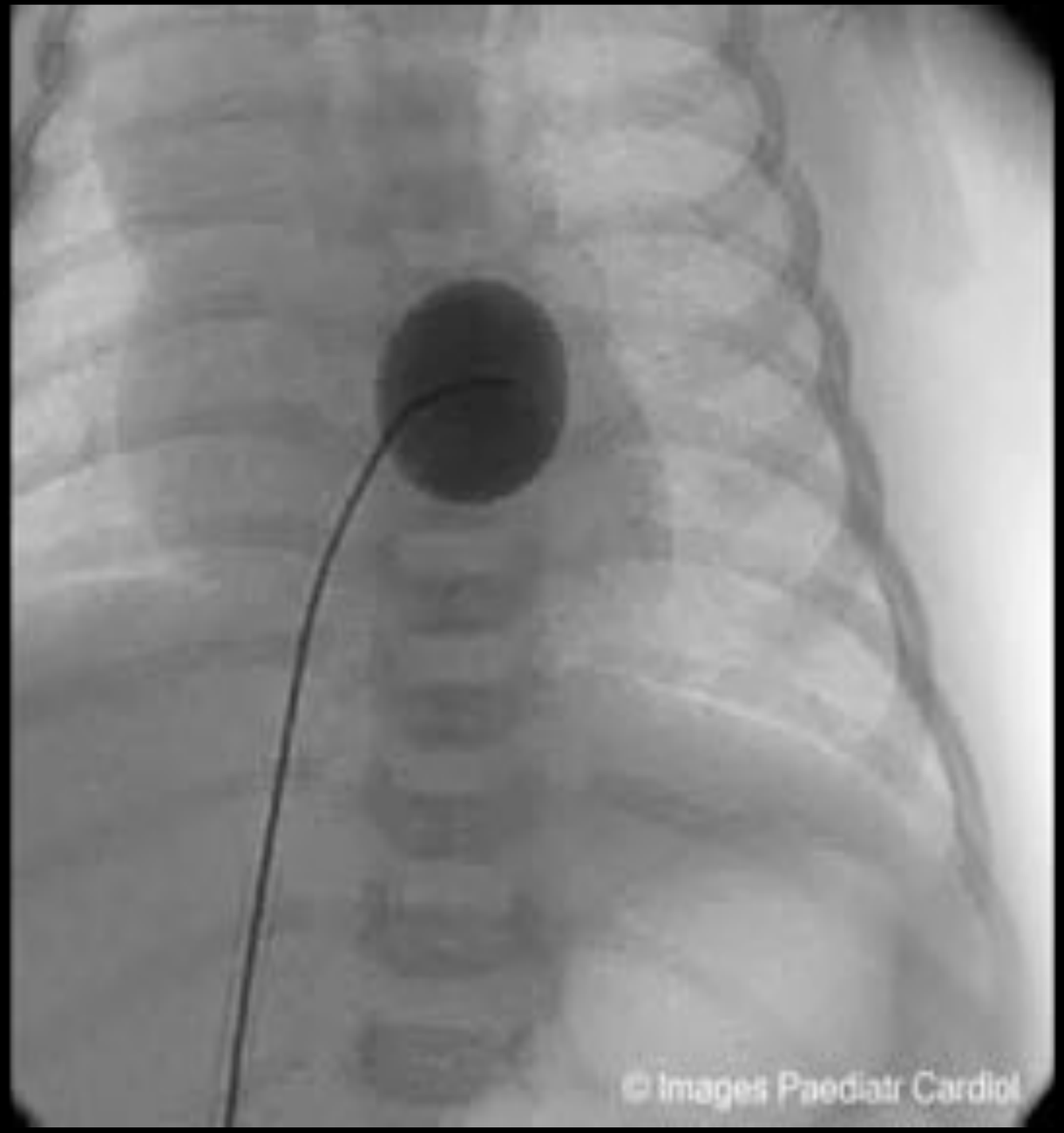
6


V
AO
AP

TGA with heart failure and restrictive PFO


© Images Paediatr Cardiol

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

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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

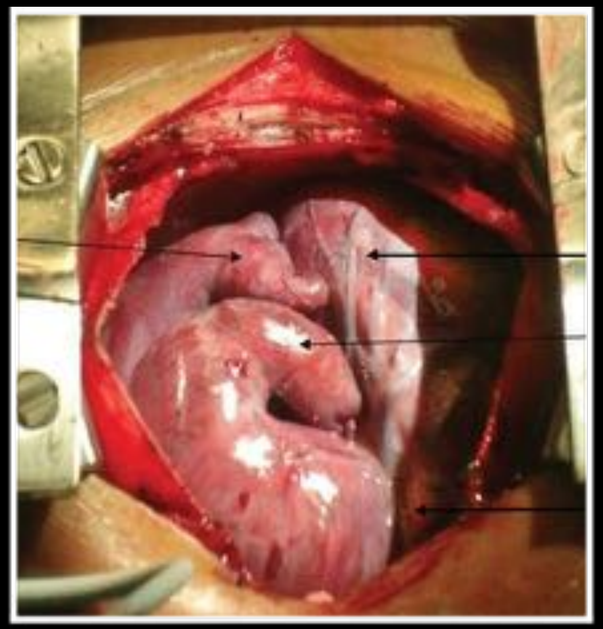
Incidence de la TGV = 0.15 pour mille naissances vivantes

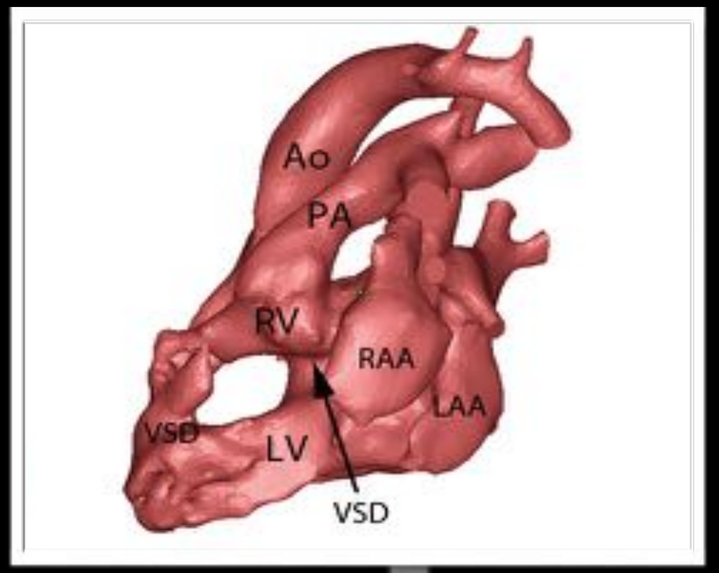
Soit environ 120 transpositions des gros vaisseaux / an

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*)

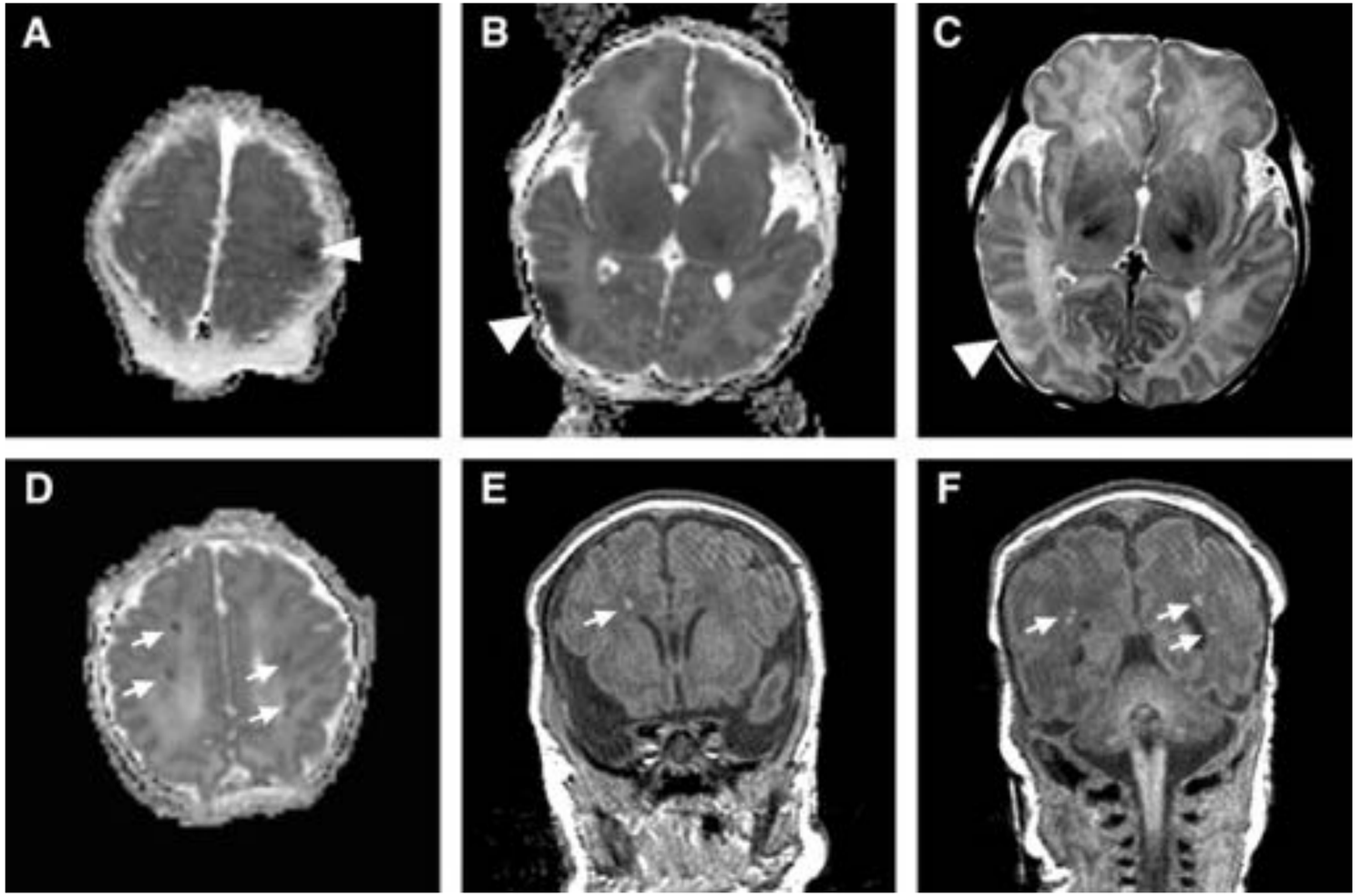
- -pour de nombreux centres moins de 1 Rashkind /an
- -pour de nombreux cardiopédiatres moins de 1 Rashkind /3 ans





Left juxtaposition of atrial appendages

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

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.

Prise en charge médicale néonatale de la TGV

Rashkind

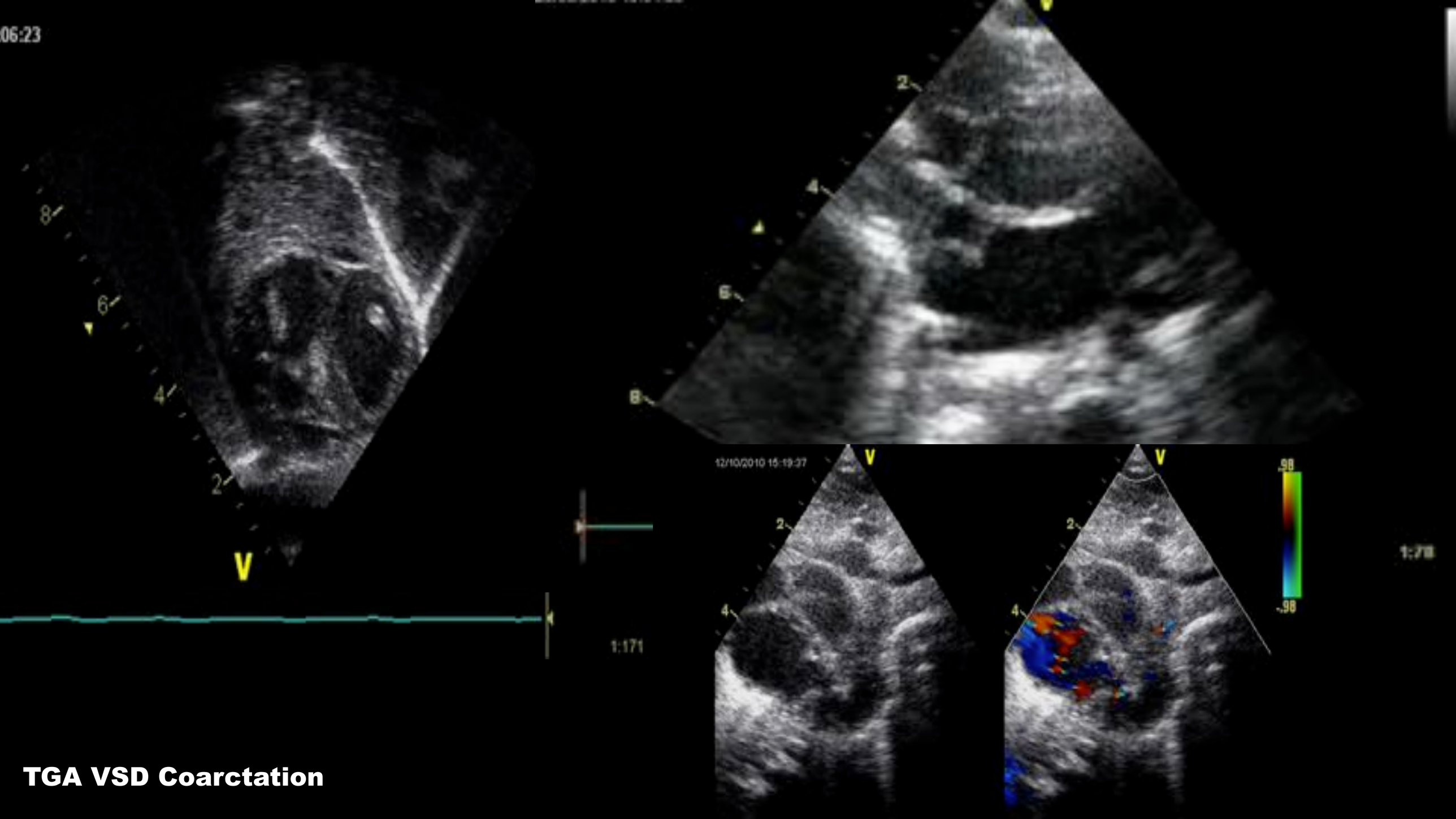
- Avantage : Mixing
- Inconvénient : décharge le VG

PGE1

- Avantage : précharge le VG
- Inconvénients : Effets secondaires

Evaluation échographique

1. Foramen ovale et canal artériel
2. Equilibre des ventricules
   Petit VD : risque de coarctation
   Petit VG : vérifier la voie pulmonaire
3. Anatomie de la valve mitrale
   Fente mitrale – insertions anormales
4. Anatomie des artères coronaires ?
5. Masse VG

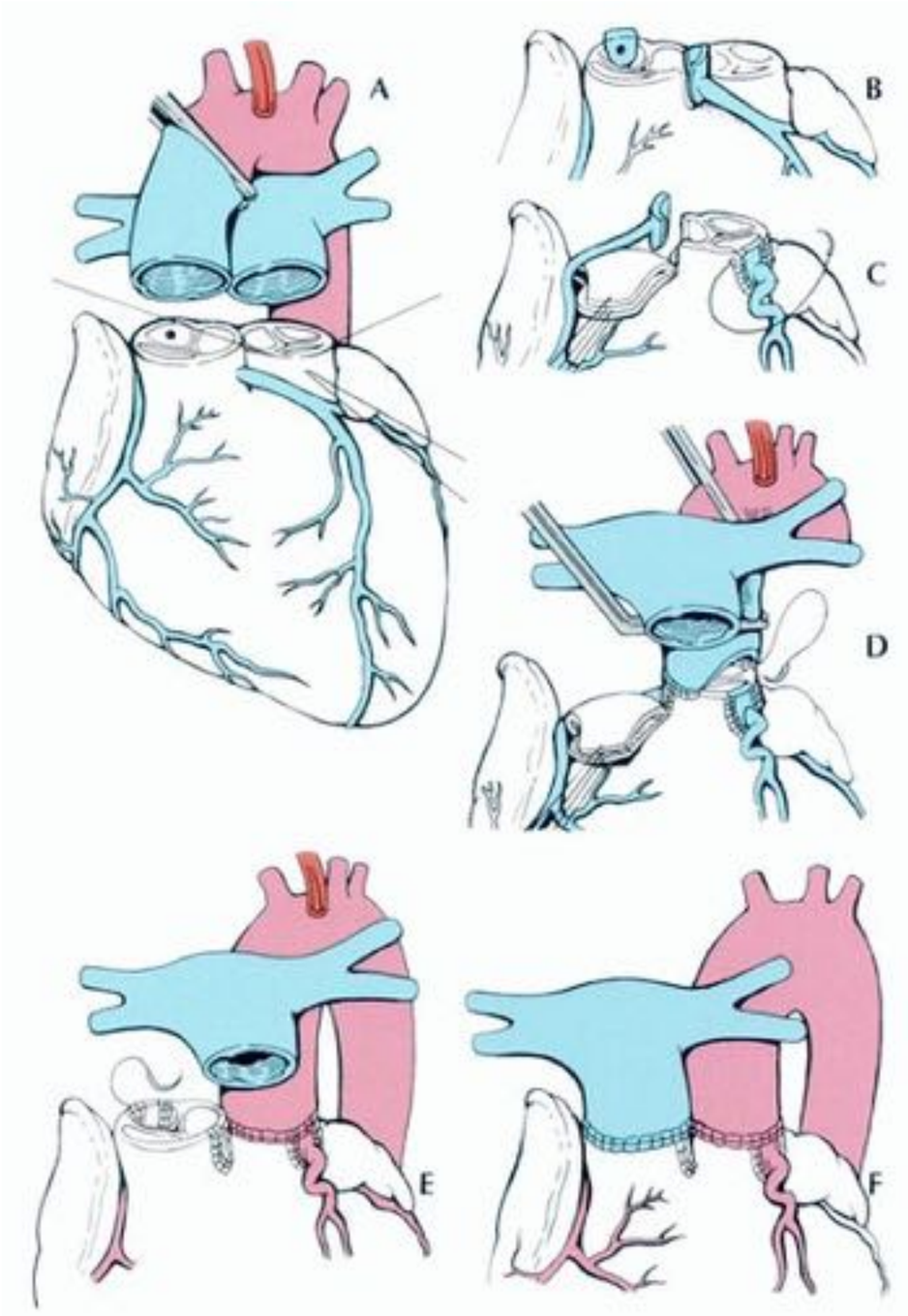


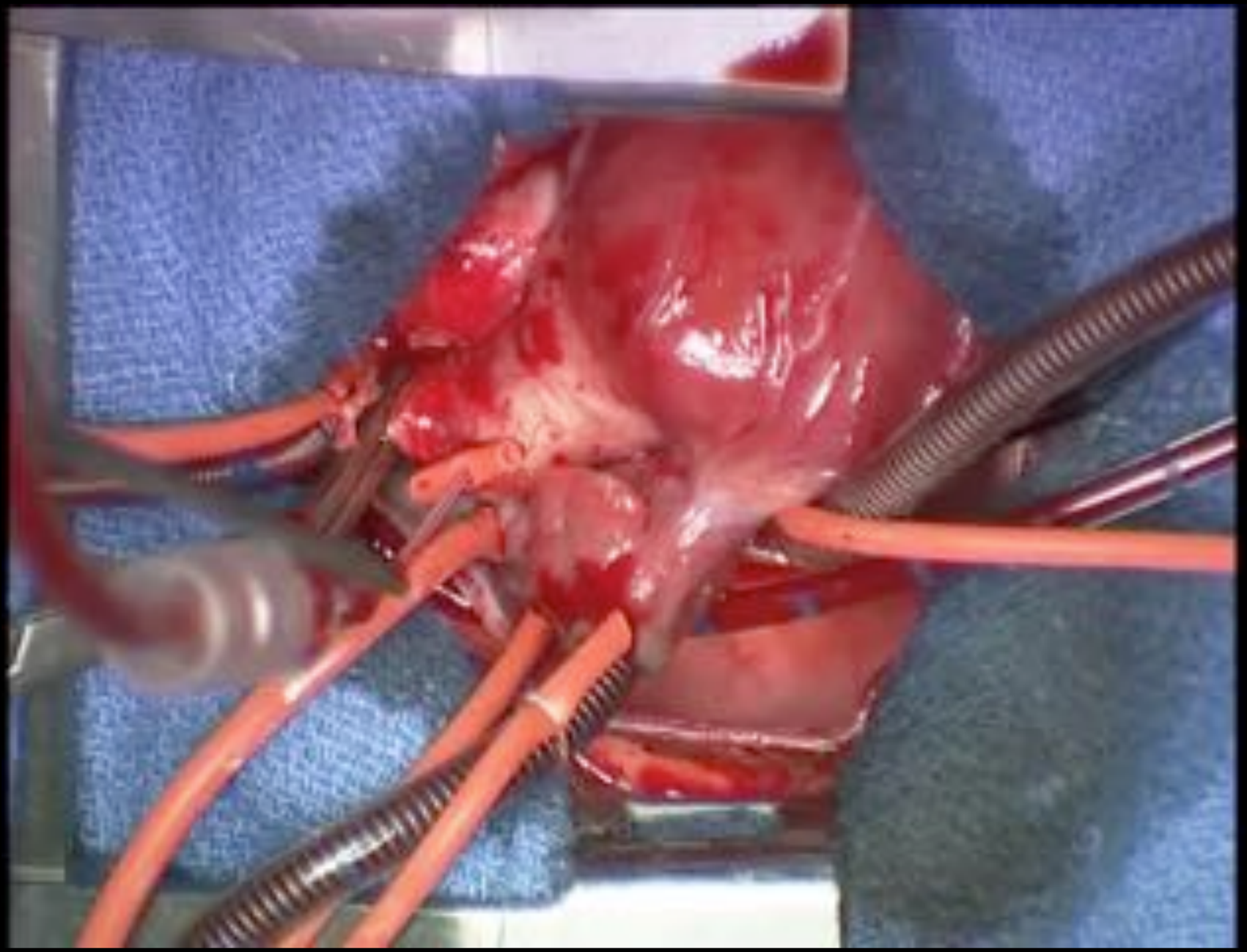
TGA VSD Coarctation

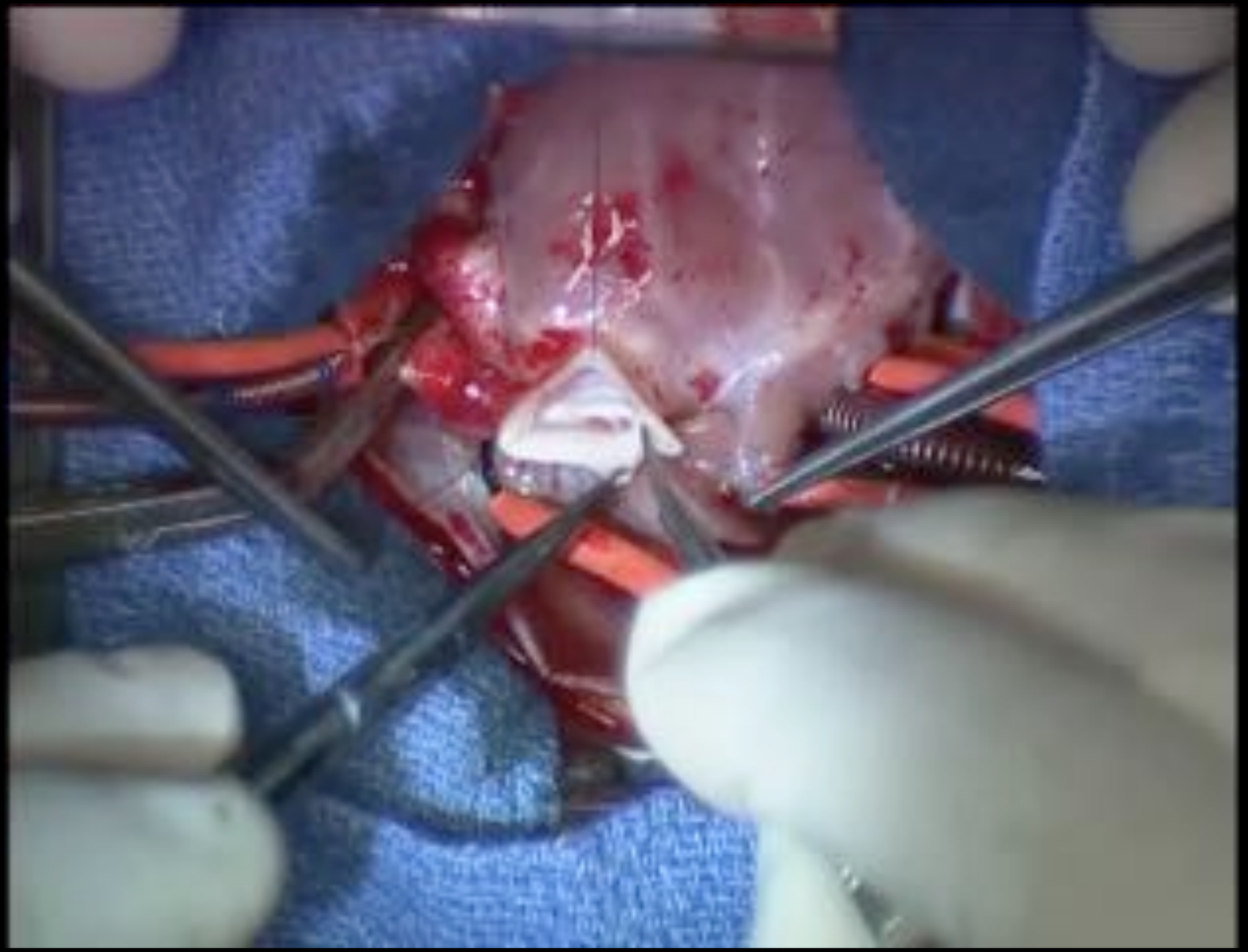
Mitral cleft

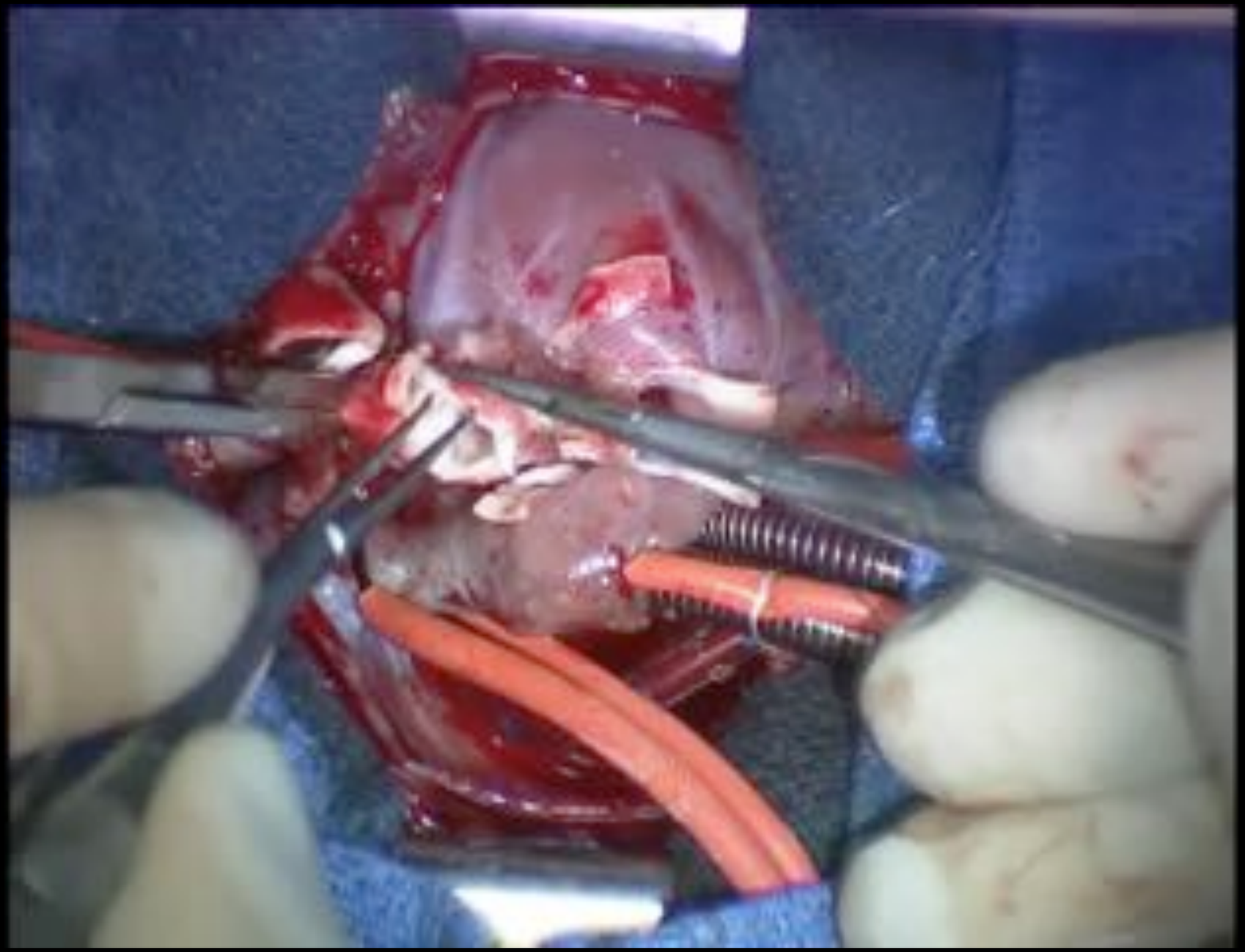
TGA left outflow tract obstruction

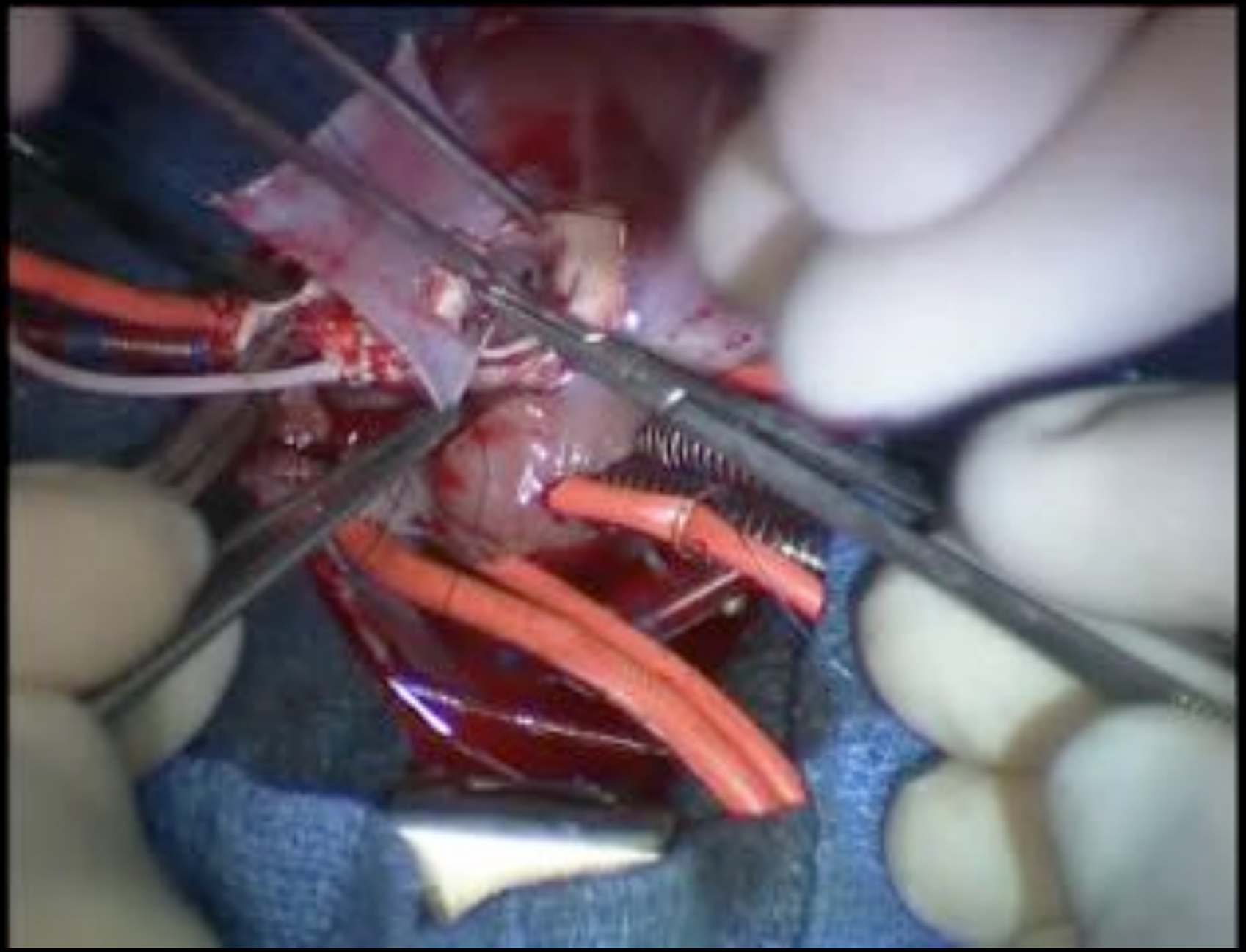
Arterial switch



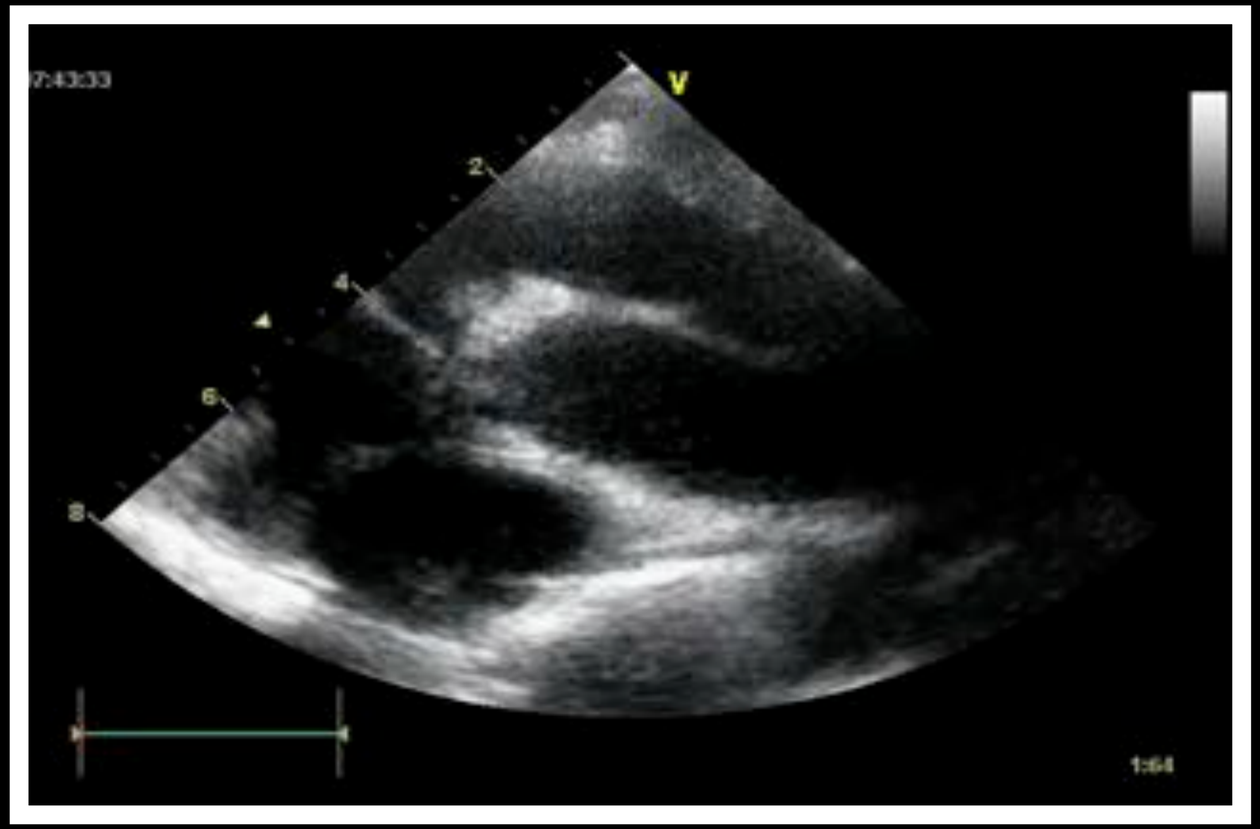


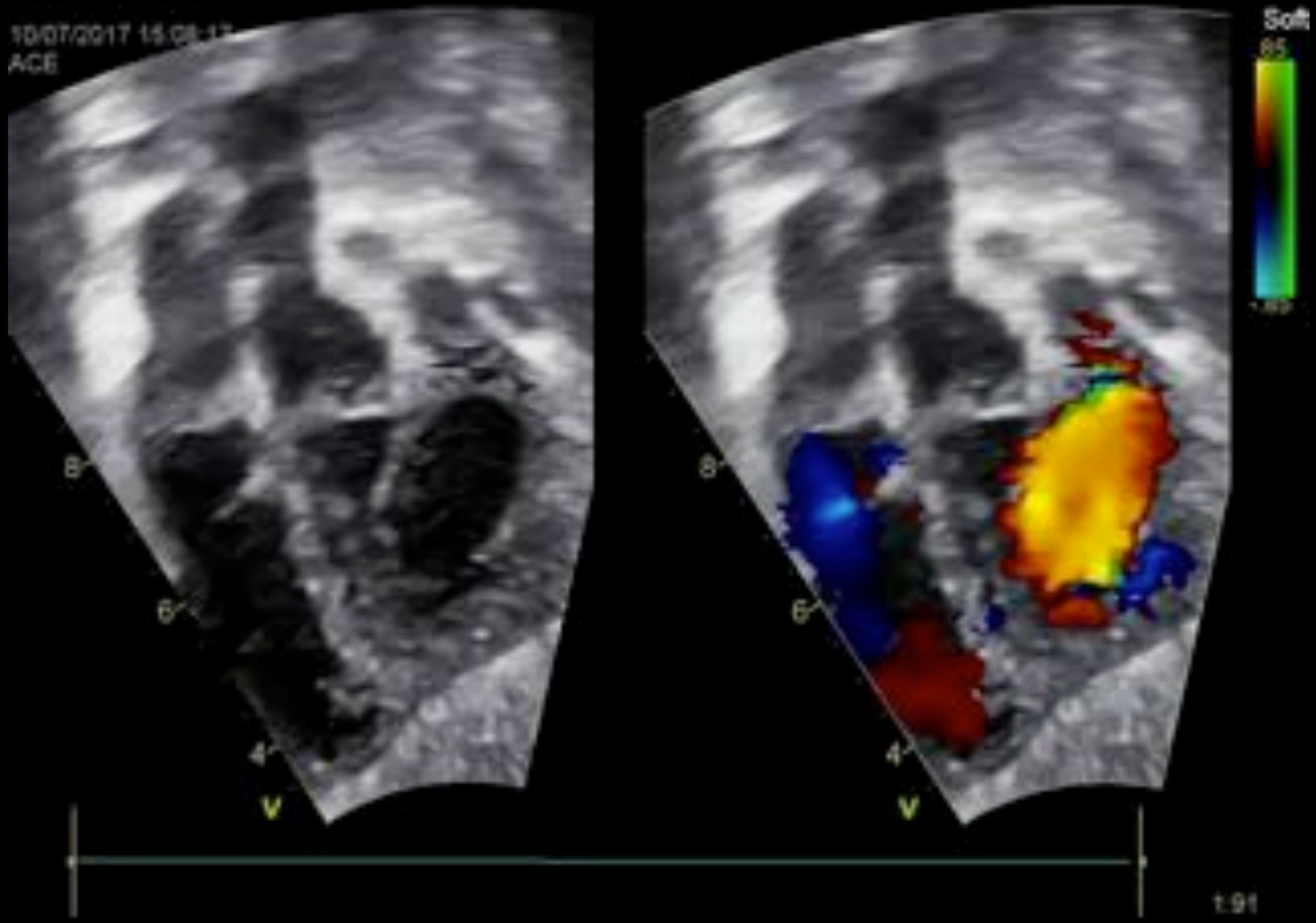




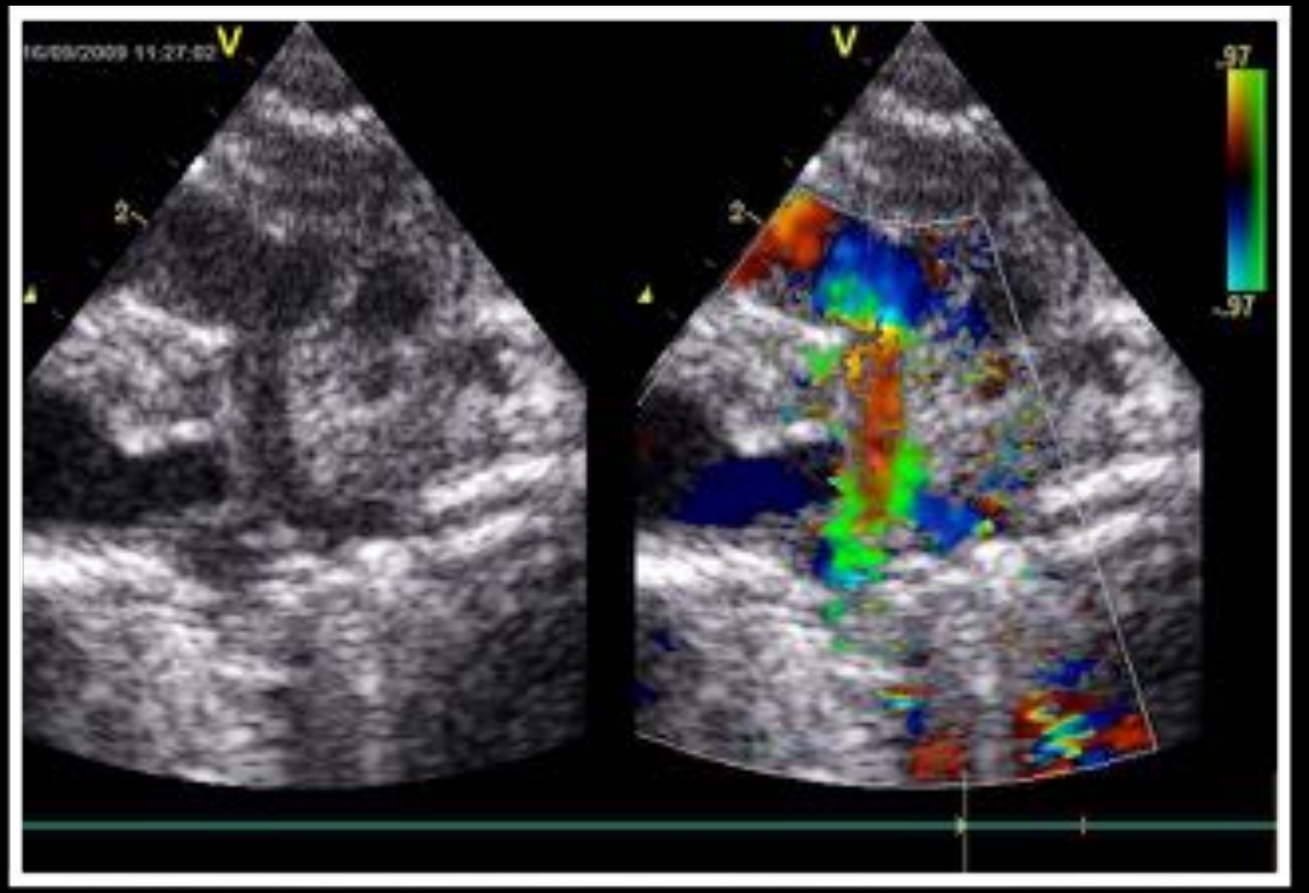


Aspect des voies d’éjection après le switch : Echo

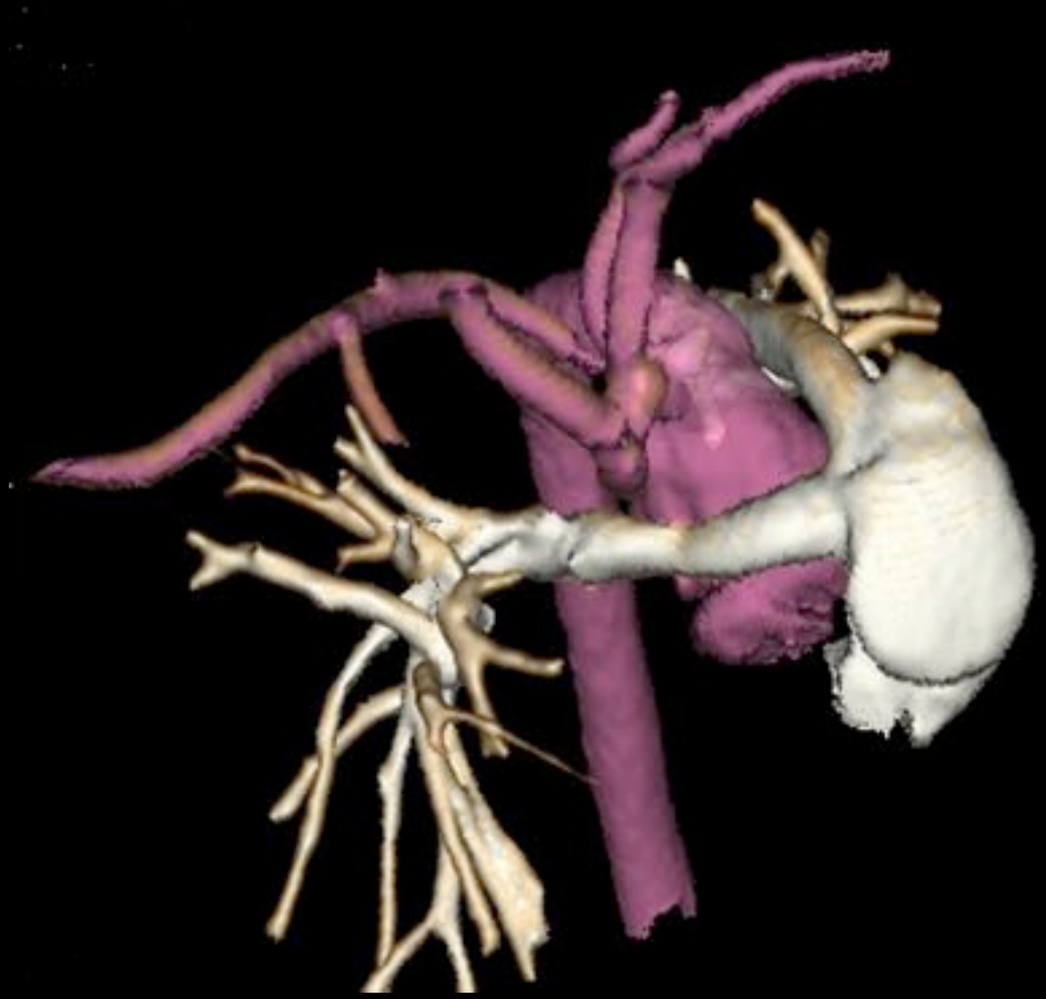



ACE
8
6
4
V
8
6
4
V
1:91

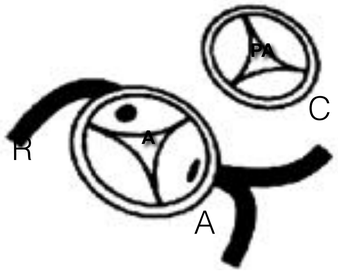

27/06/2017 09:38:10
1.232


16/09/2009 11:27:02
97
-97

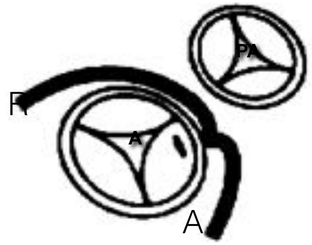
Aspect des voies d’éjection après le switch : CT



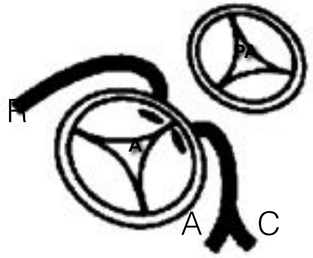
Type A



Type B

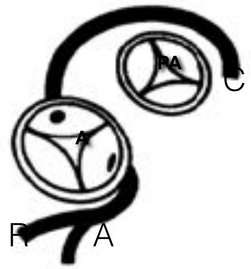


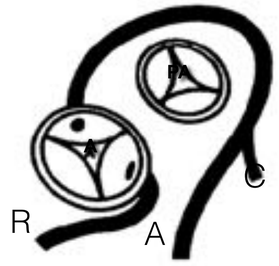
Type C

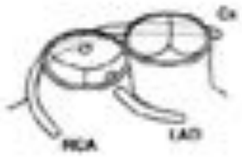
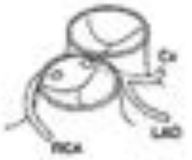


Type D

Type E



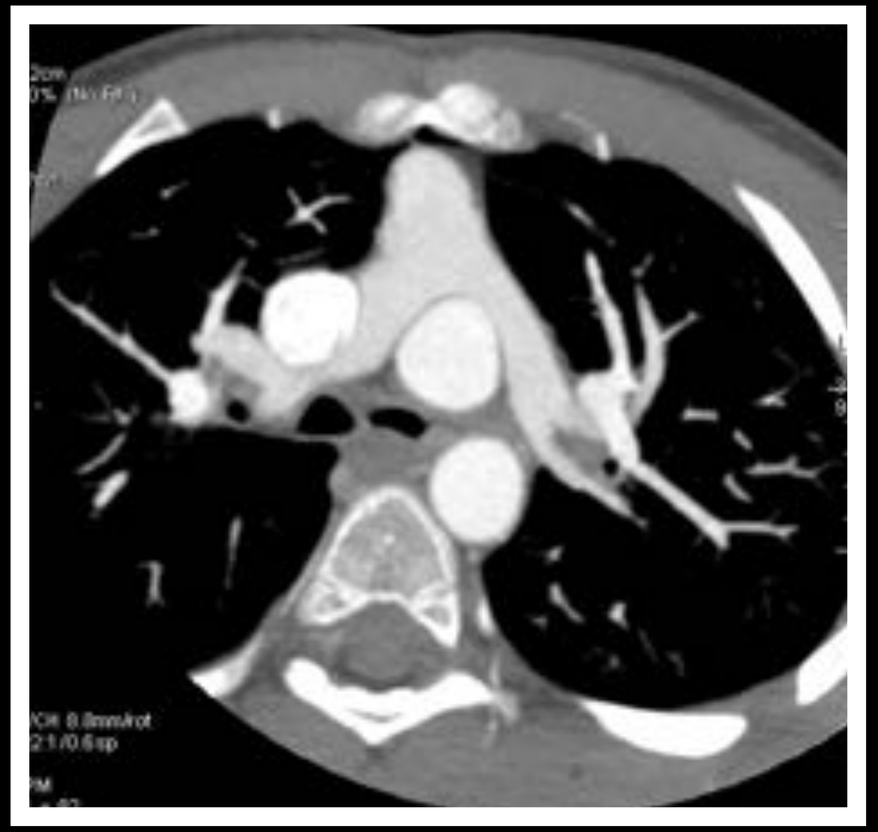


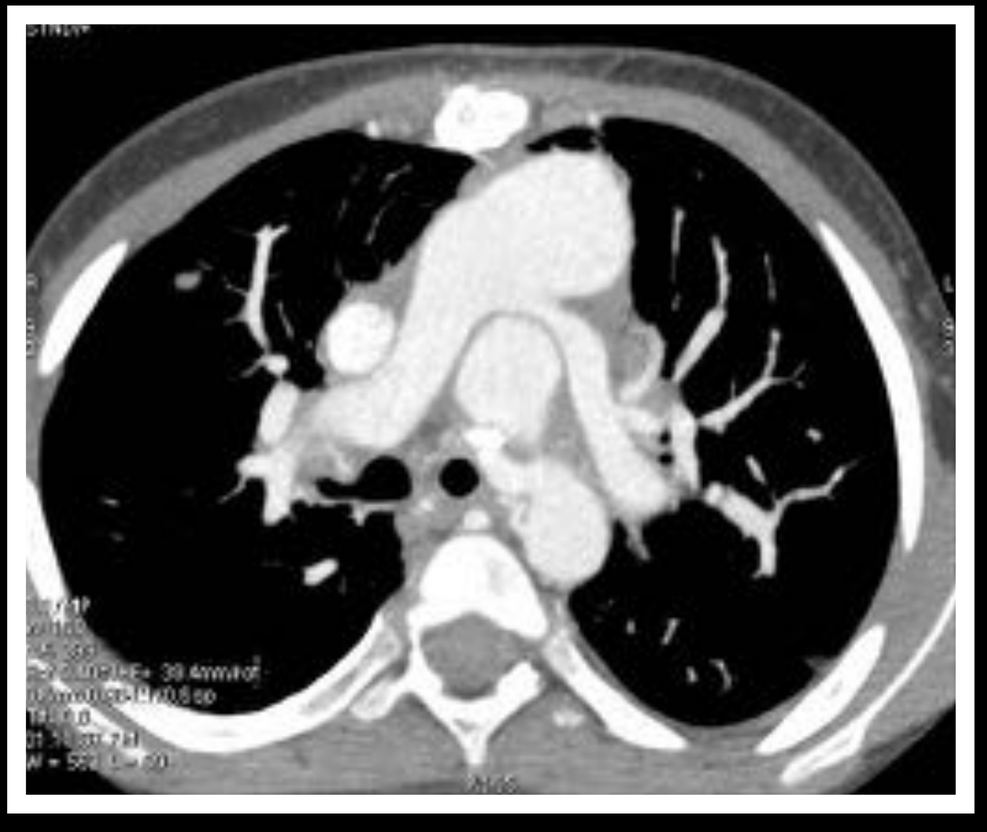
Habituel	Circonflexe de l'ACD	ACG unique	ACD unique	Inversée	ACD inversée et circonflexe	Coronaires Intramurales
						
66,9%	16,1%	1,7%	3,9%	2,4%	4,2%	3,2%

Complications du switch artériel

- Mortalité
  - 1 à 2%
  - Liée aux coronaires
- Morbidité
  - Artères pulmonaires
  - Coronaires
  - Aorte

Complications artérielles pulmonaires

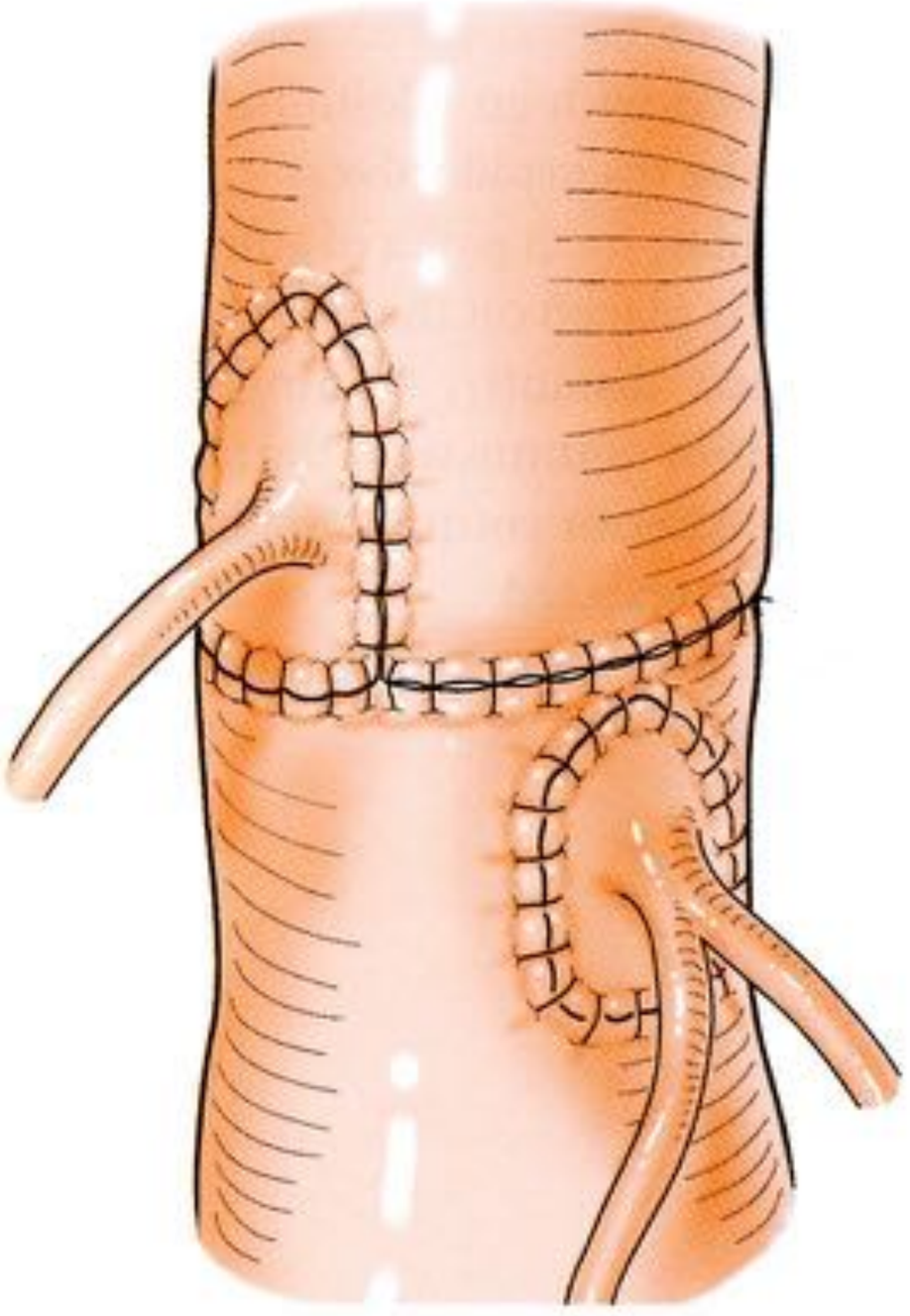


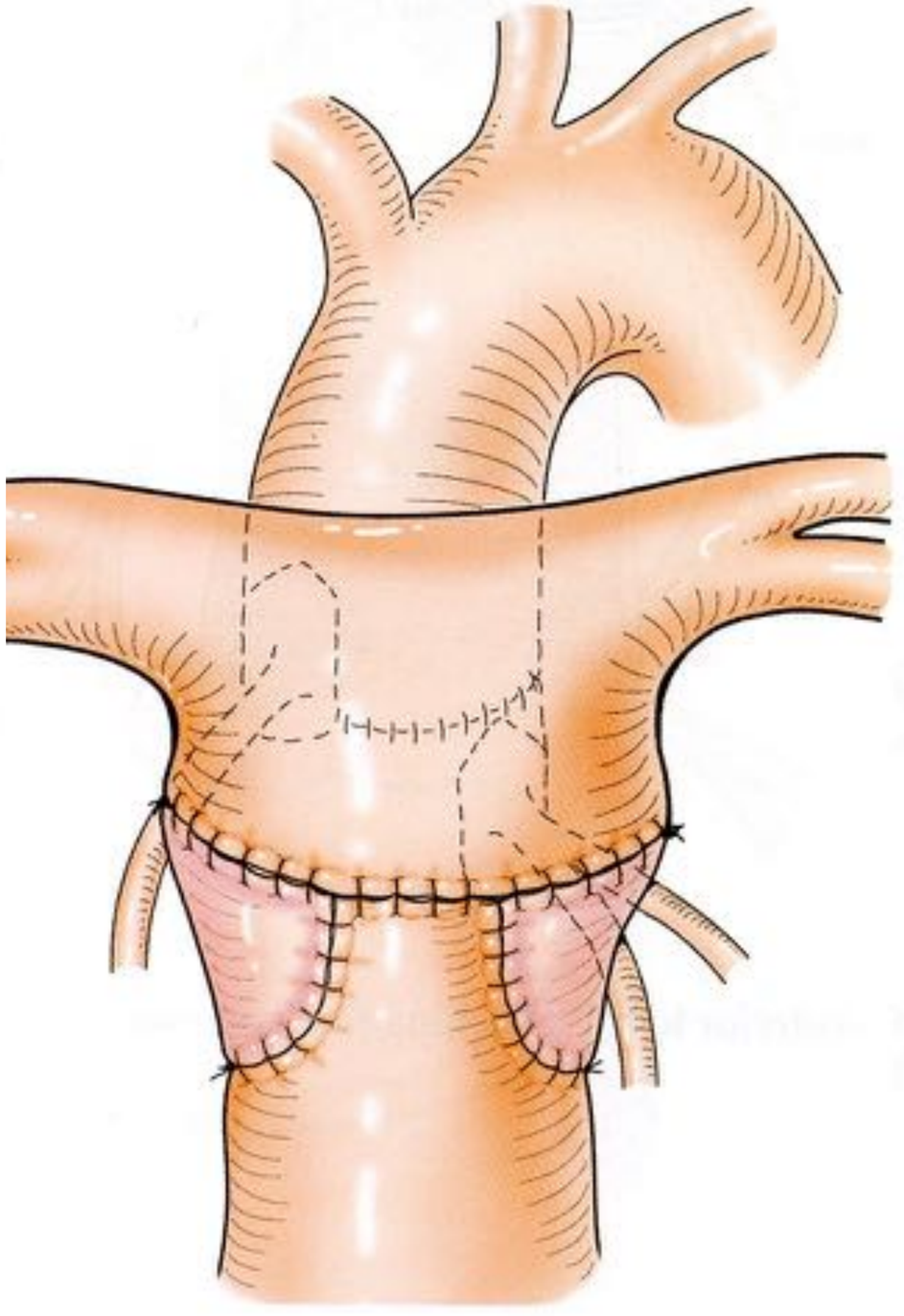


Les lésions coronaires après switch artériel

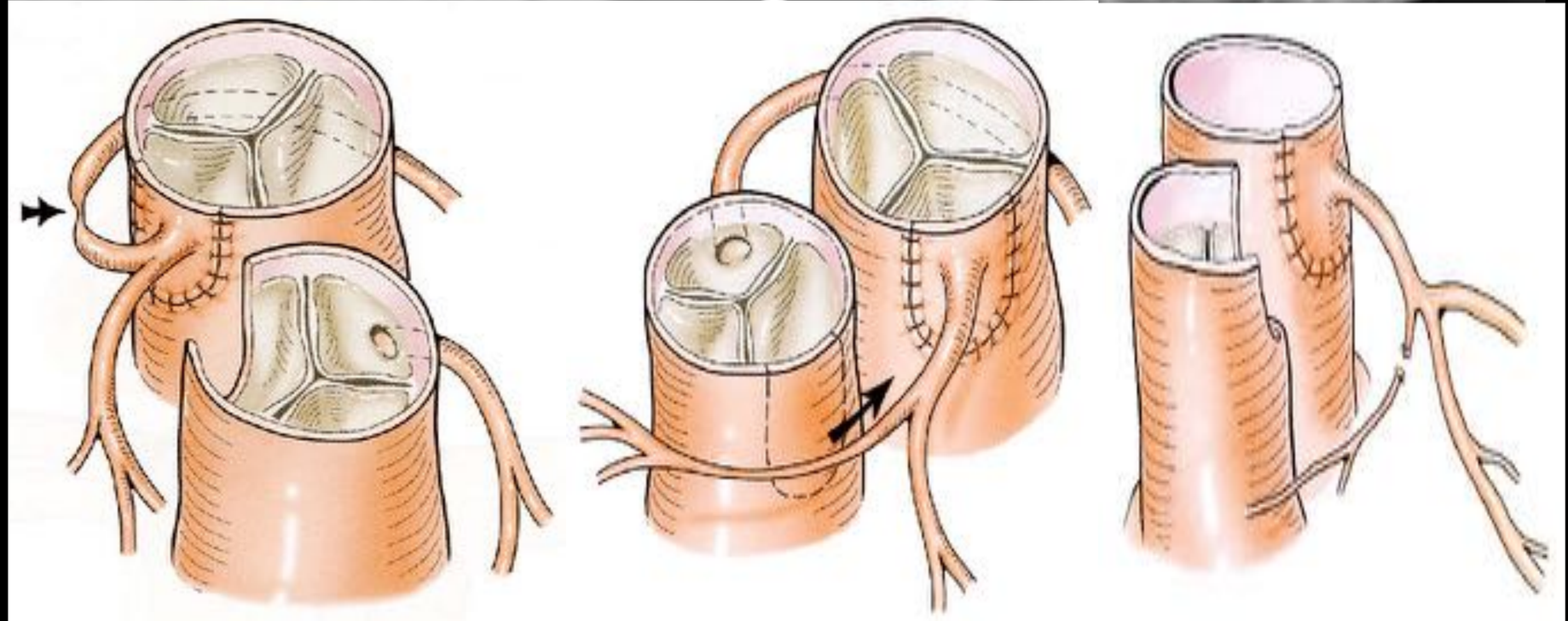
Comment les prévenir ?

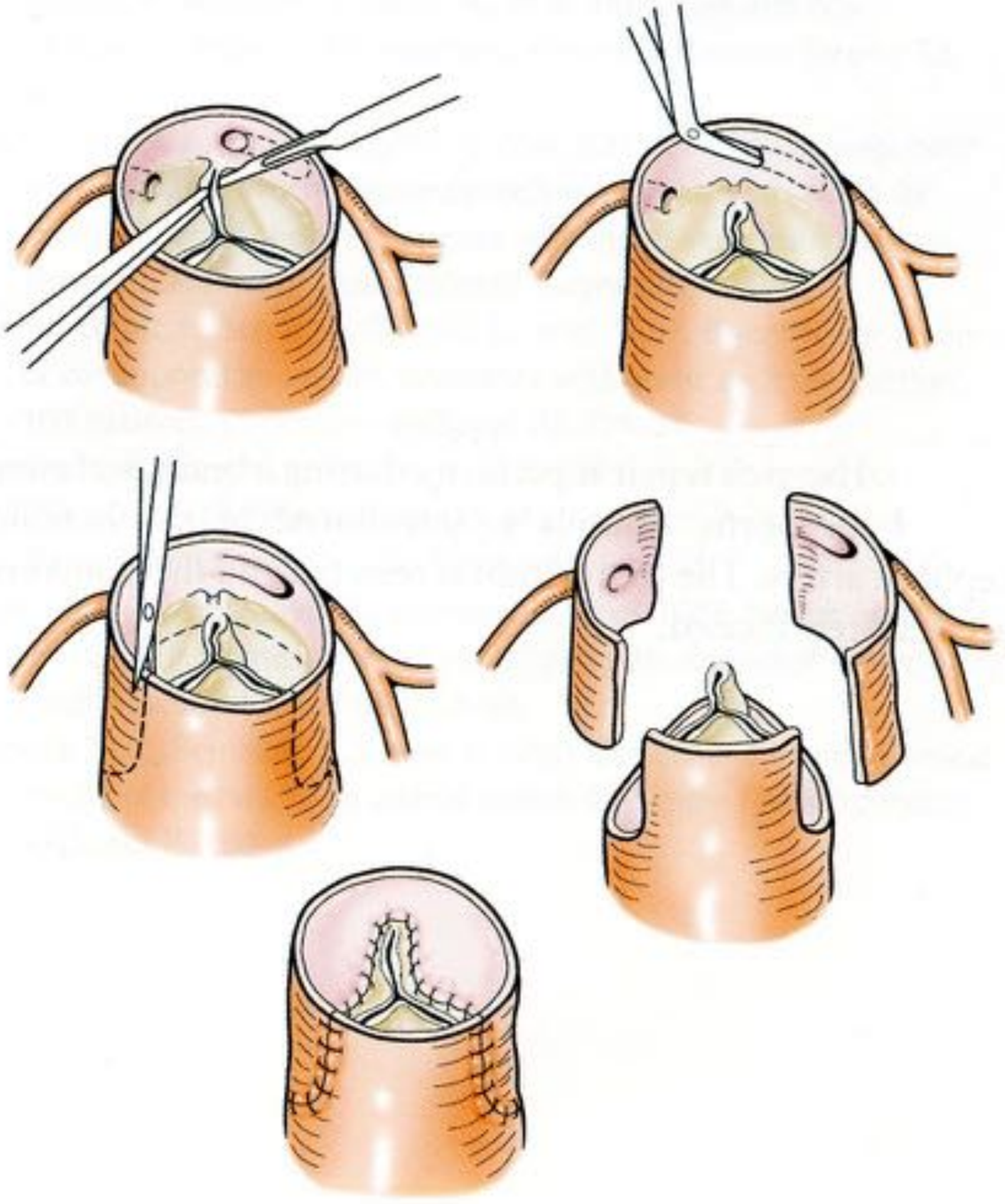
- Le transfert coronaire doit être parfait
  - rotation
  - distorsion
  - kinking
  - compression
  - étirement
  - lésion intimale
- Pas de technique standardisée





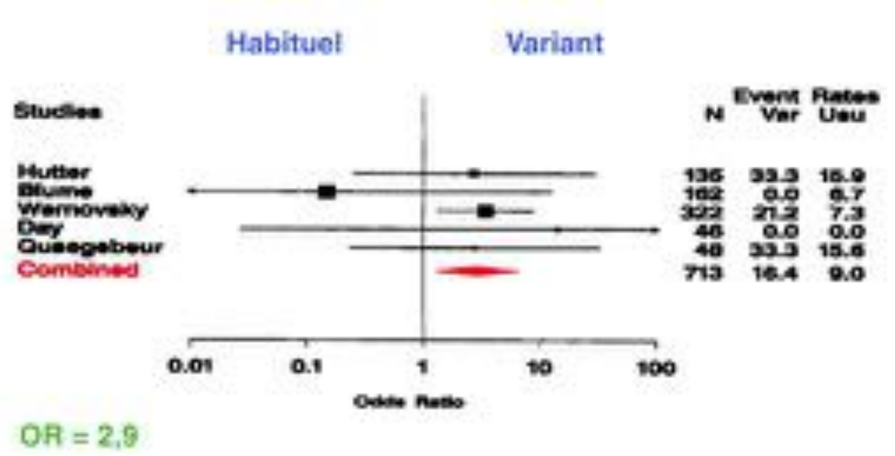

PA
Ao
Ao
PA



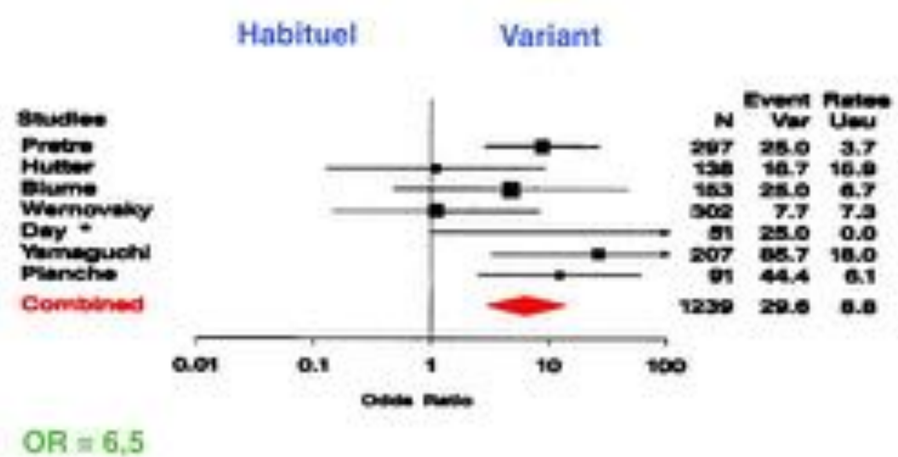


Influence de la distribution coronaire sur la mortalité après switch artériel

Coronaires uniques



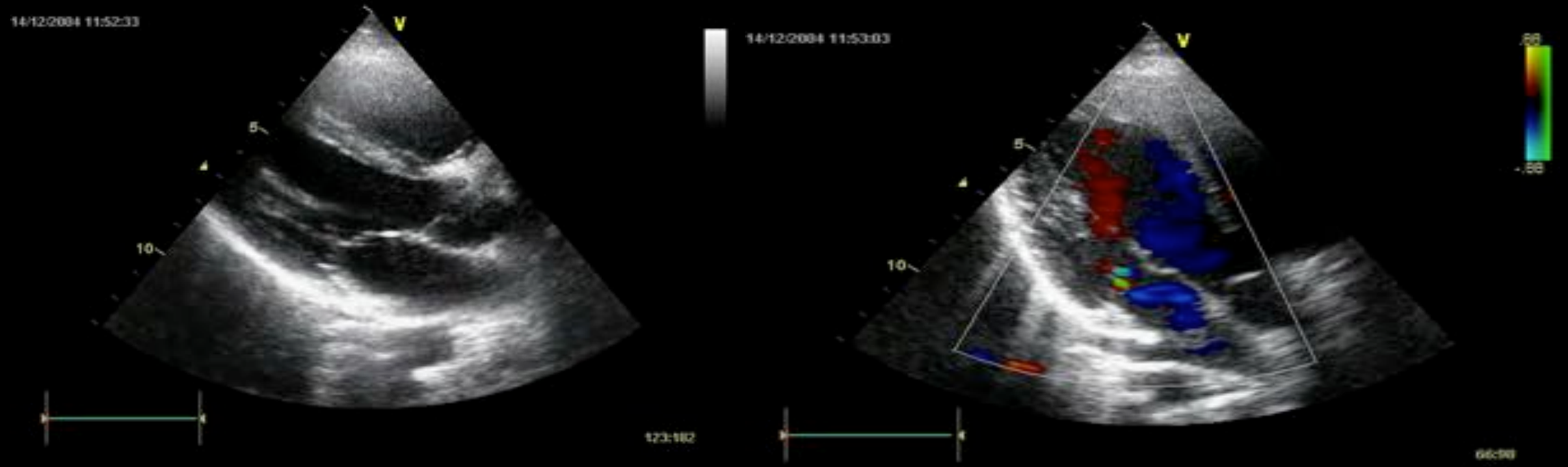
Coronaires intramurales

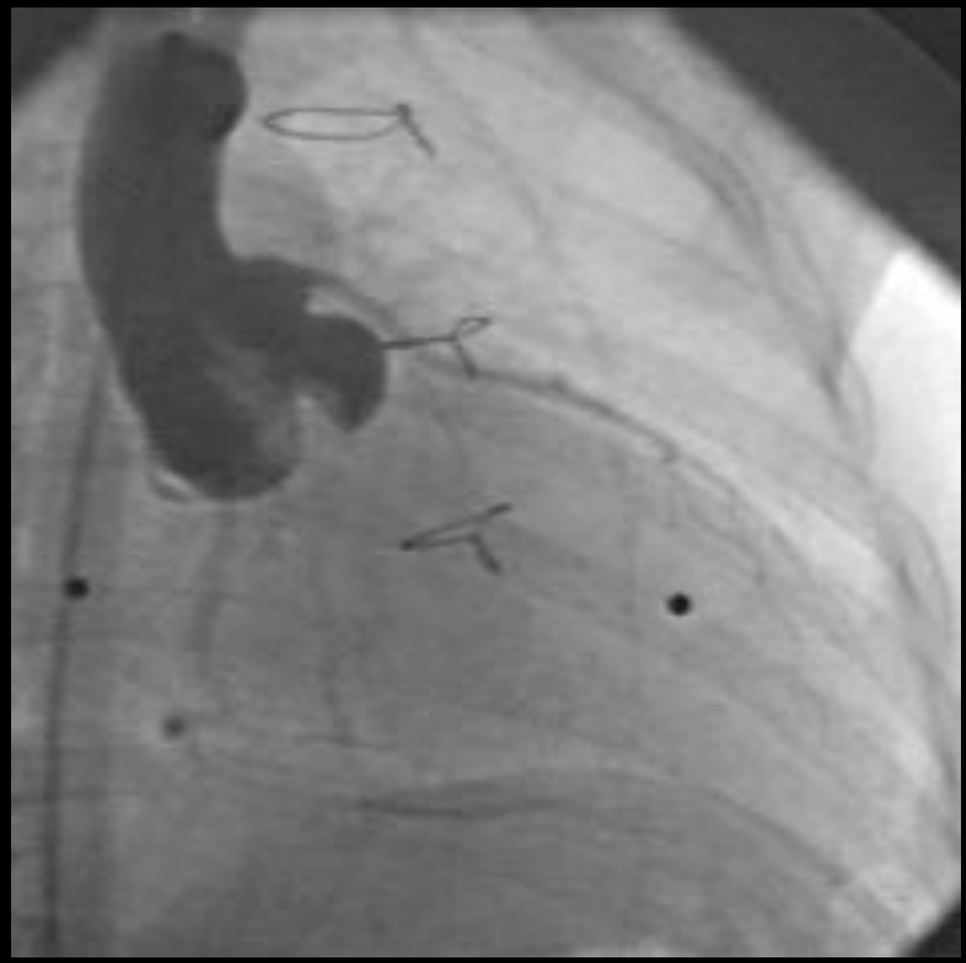


- Artères coronaires uniques et artères coronaires avec un trajet intramural restent à risque.
- La mortalité d' une distribution variante est deux fois élevée que celle d 'une distribution habituelle (OR 1,7).

Pasquali, 2002

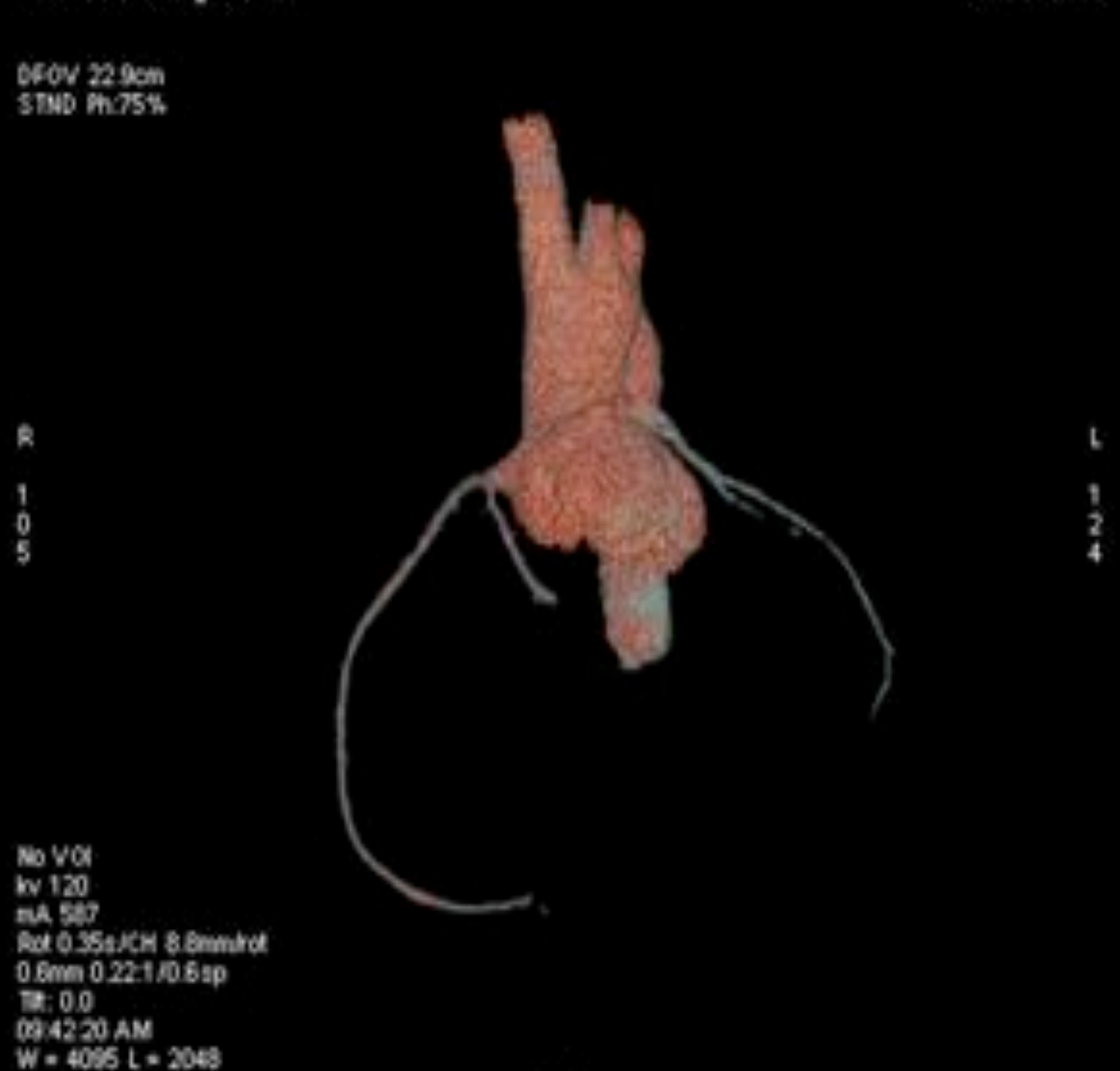
Ischémie myocardique TGV





Coronaires de type D


DFOV 13.4cm
STND Ph:75% (No Filt.)
R
A
I
L
P
S
No VOI
kv 120
mA Mod.
Rot 0.35s/CW 10.4mm/rot
0.6mm 0.26:1/0.6sp
Tilt: 0.0
12:25:34 PM

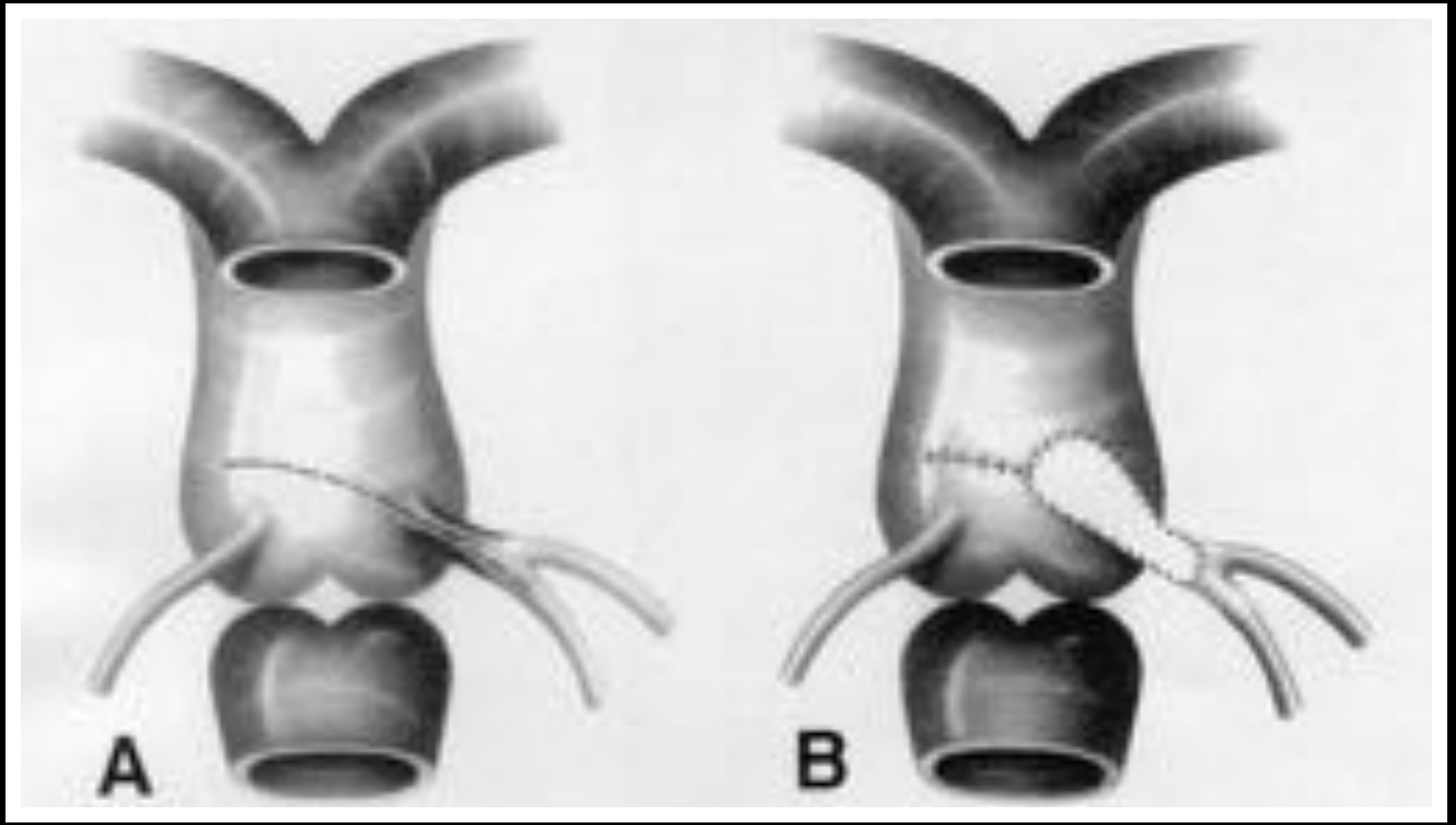

DFOV 22.9cm
STND Ph:75%
R 105
L 124
No VOI
kv 120
mA 587
Rot 0.35s/CH 8.8mm/rot
0.6mm 0.22:1/0.6sp
Tilt: 0.0
09:42:20 AM
W = 4095 L = 2048

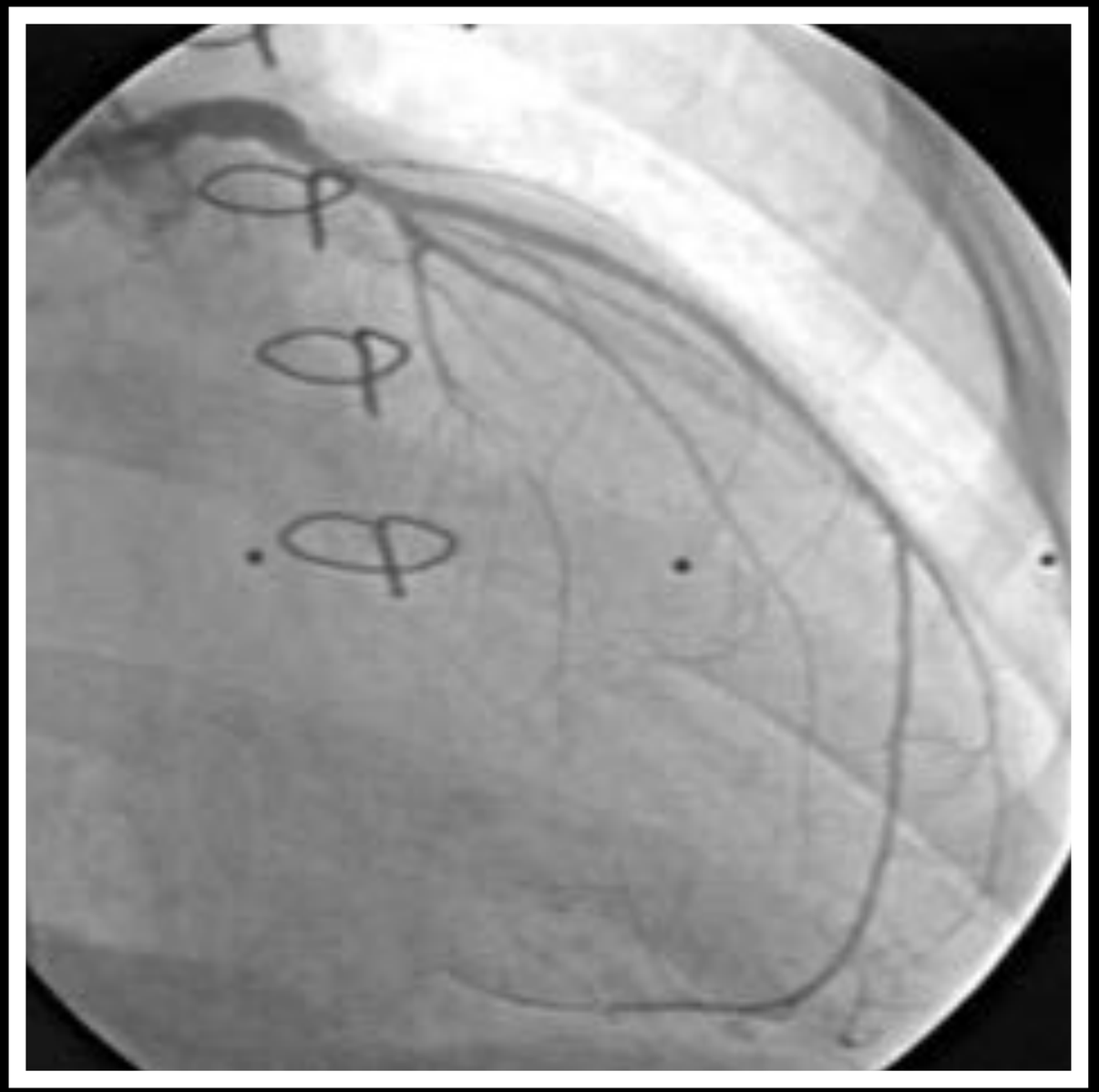

Ex: 32
Se: 2 +c
Volume Rendering No cut
DFOV 15.0cm
STND Rh:75% (No Filt.)
A
R
No VOI
kv 120
mA Mod.
Rot 0.35s/CH 8.0mm/rot
0.6mm 0.2:1/0.6 sp
Tilt: 0.0
10:03:33 AM
W = 4095 L = 2048
1147

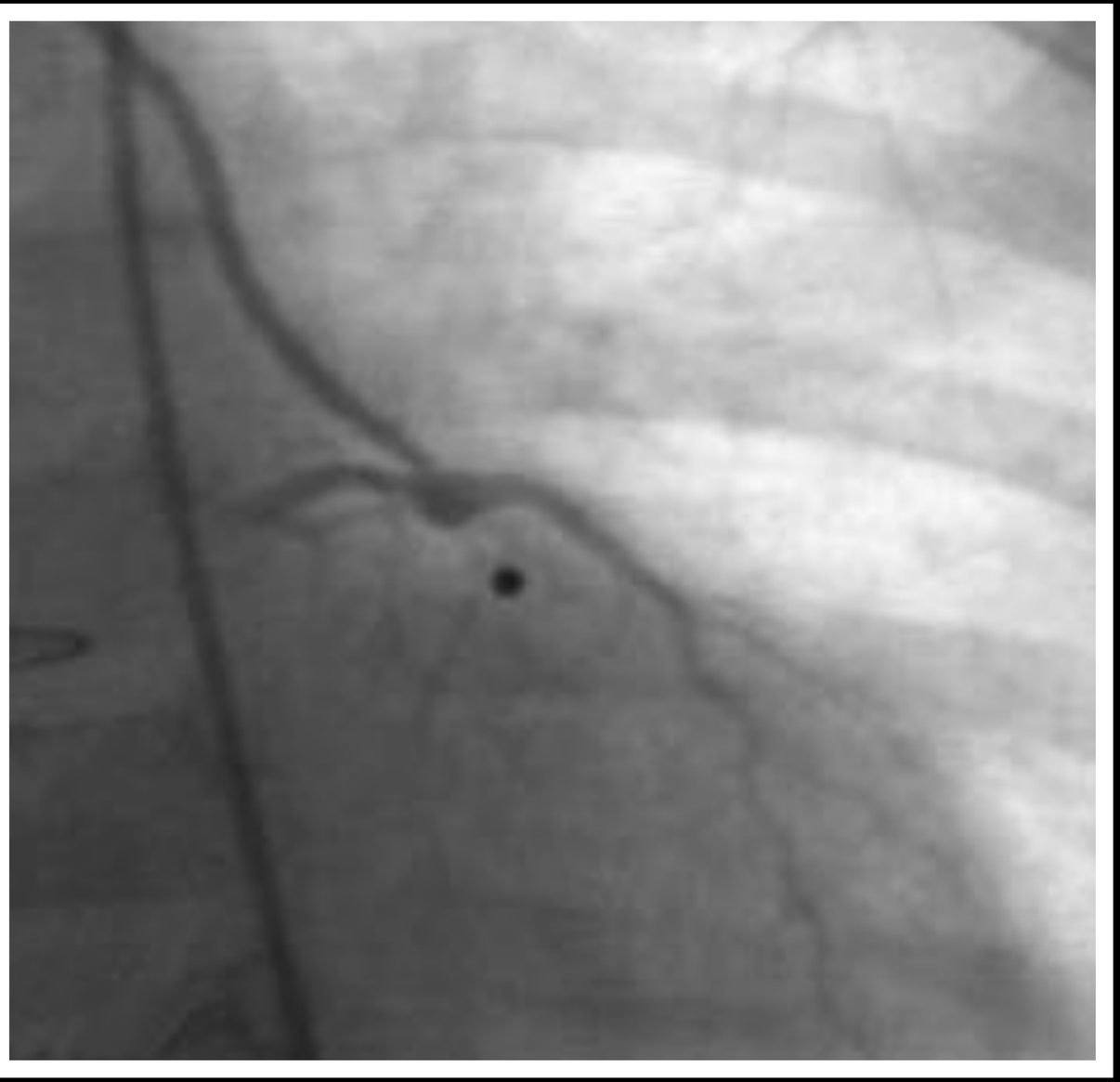

Ex: 8420
Se: 2
Volume Rendering No cut
DFOV 18.9cm
STND Ph:75%
13 0193049445
Nov 22 2006
A
R
S
F
L
I
No VOI
kv 100
mA Mod.
Rot 0.35s/CH 9.6mm/rot
0.6mm 0.24:1/0.6sp
Tilt: 0.0
09:53:12 AM
W = 4095 L = 2048
IAL

Que faire en cas de lésion coronaire ?

- Détecter l’ischémie myocardique
- Décider d’un traitement adapté à:
  - La présence d’une ischémie myocardique
  - La complexité de la lésion à revasculariser
  - L’âge et le poids de l’enfant


A
B





Pontage mammaire interne-IVA post-switch

Anévrisme de l’aorte ascendante – Insuffisance aortique

Qualité de vie et développement intellectuel

- QI normal
- Pas d’anomalie du développement intellectuel après 30 ans de recul
- Fonctions exécutives pouvant être altérées de façon modérée

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

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

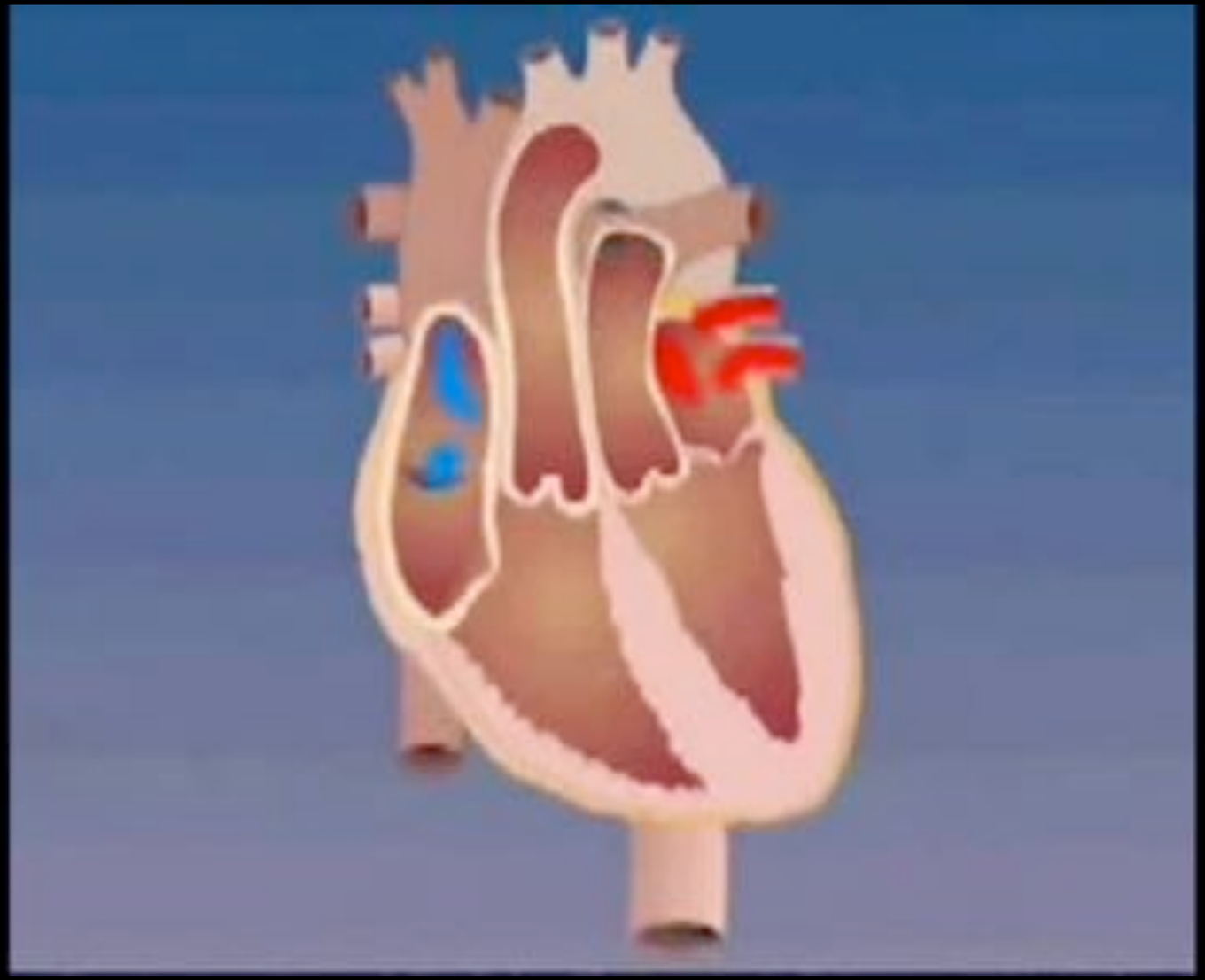
Calderon J, Bonnet D. J Pediatr 2012

Correction atriale de la TGV

Mustard operation

5-8 % of congenital heart disease

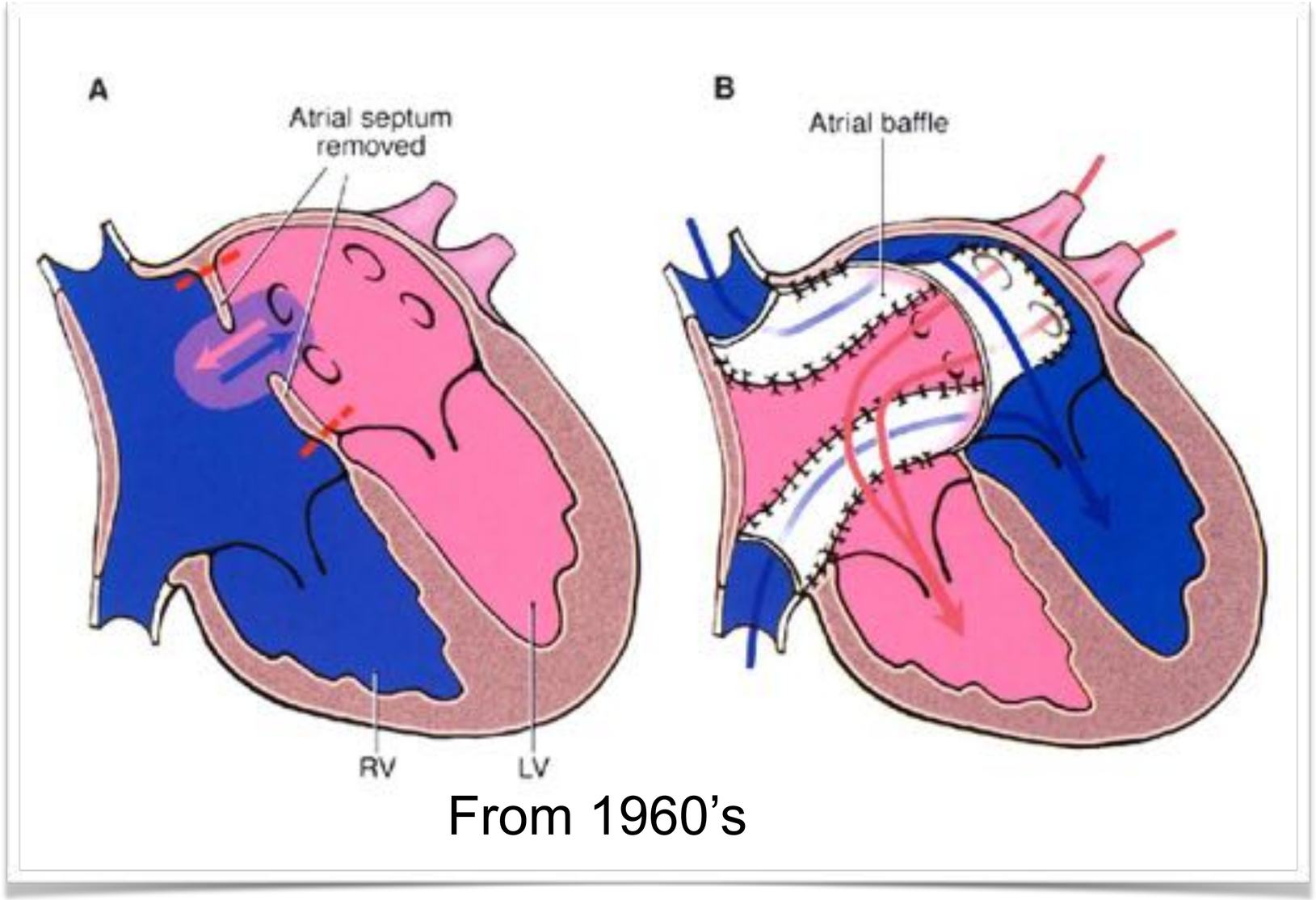
70% isolated TGA
30% complex (VSD & PS)



Atrial switch

Mustard procedure: pericard

Senning procedure: atrial tissue

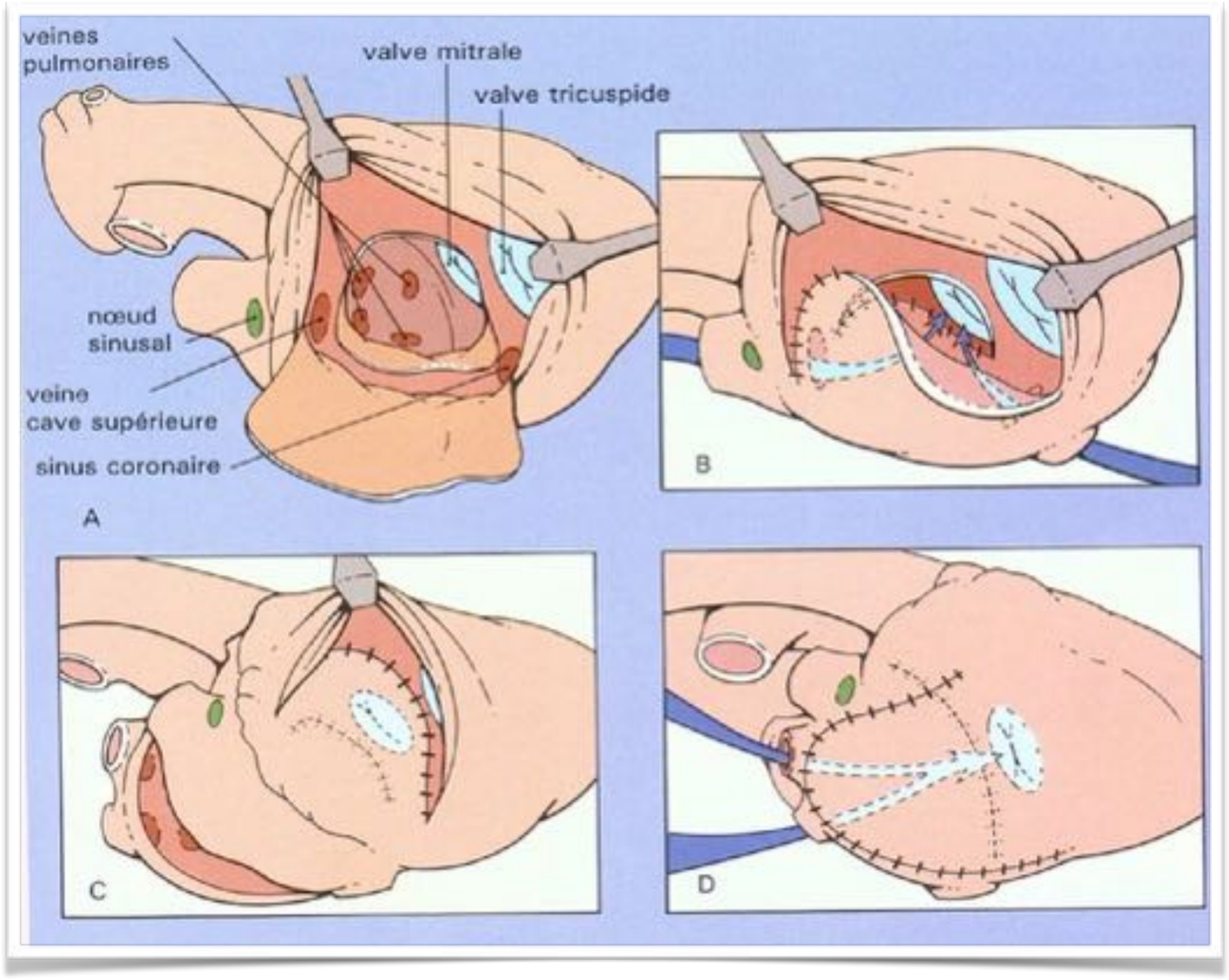


From 1960’s

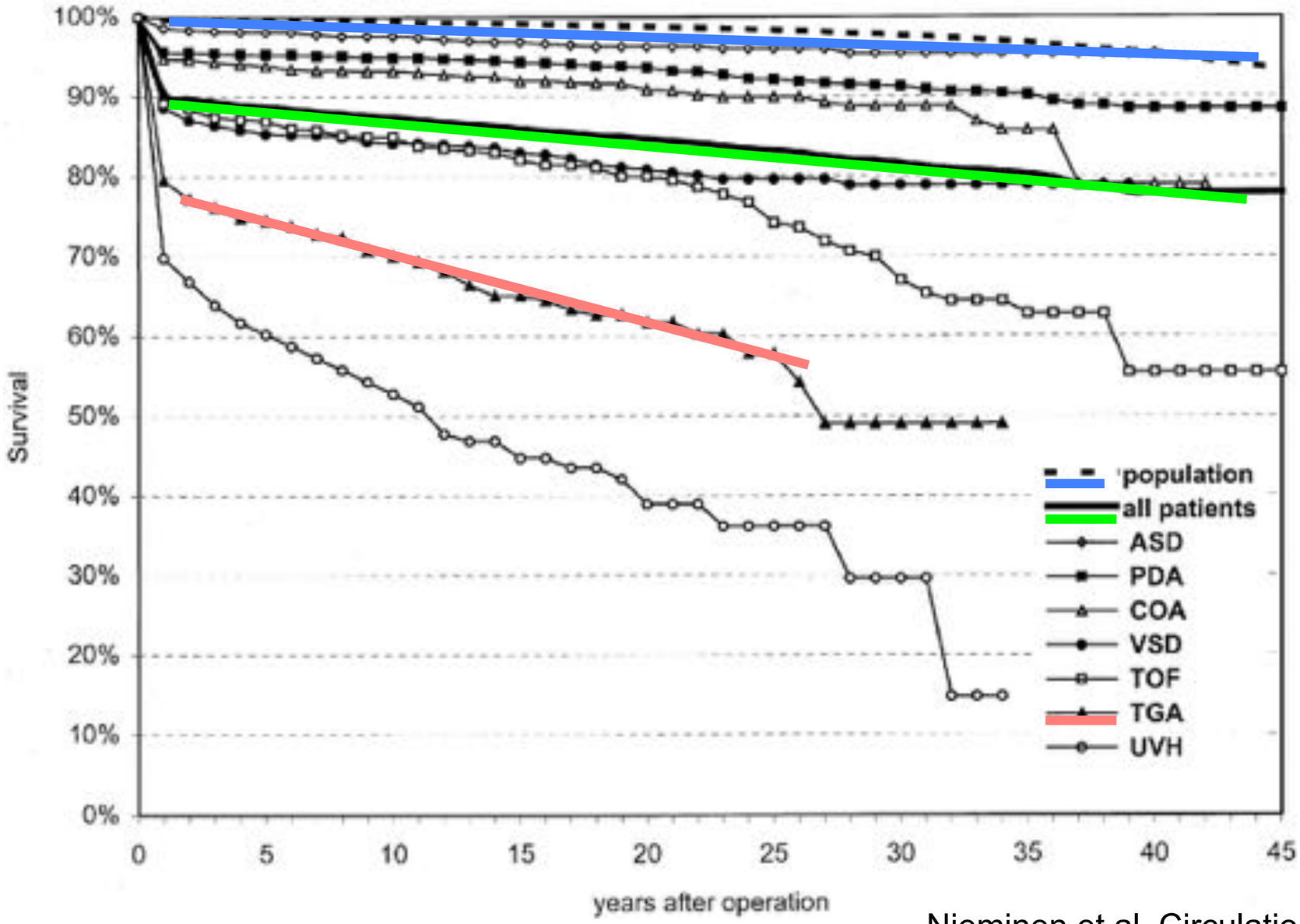
Atrial switch

Mustard procedure: pericardium

Senning procedure: atrial tissue

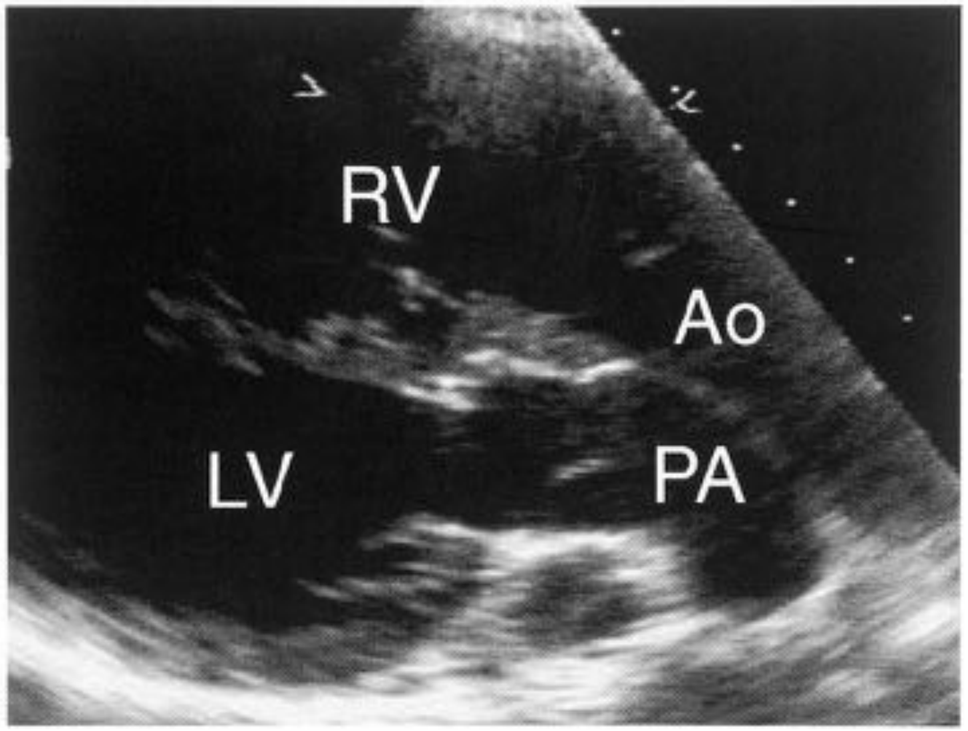


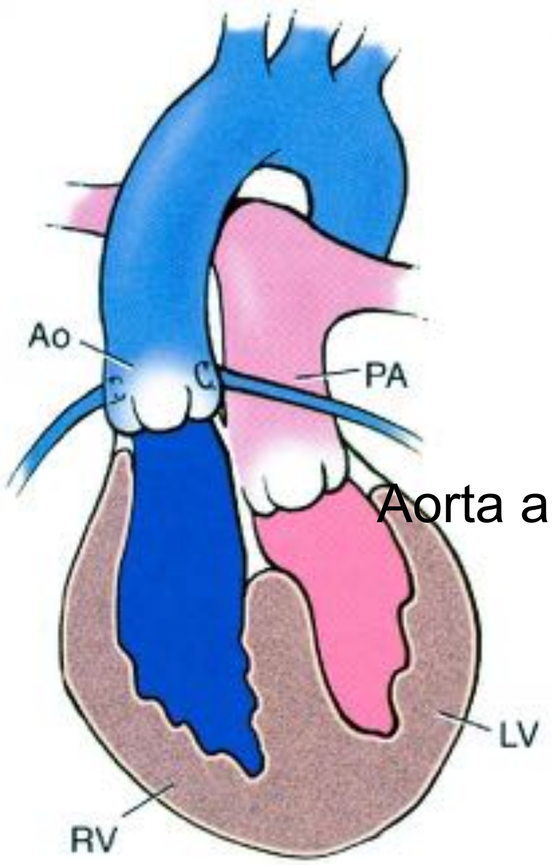
Survival of all patients, separate diagnostic groups, and general population



Nieminen et al, Circulation 2001

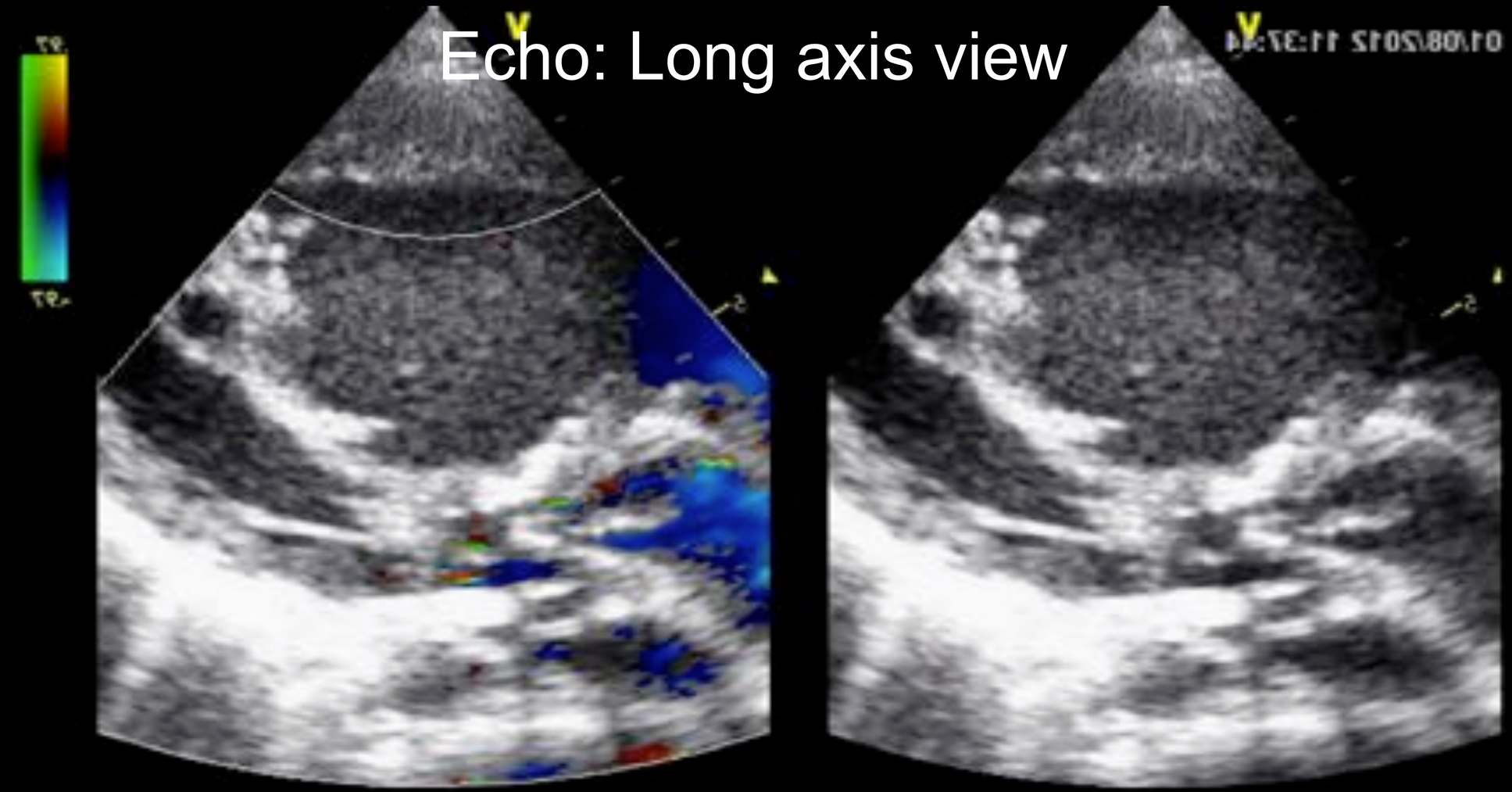
Echocardiogram long axis view





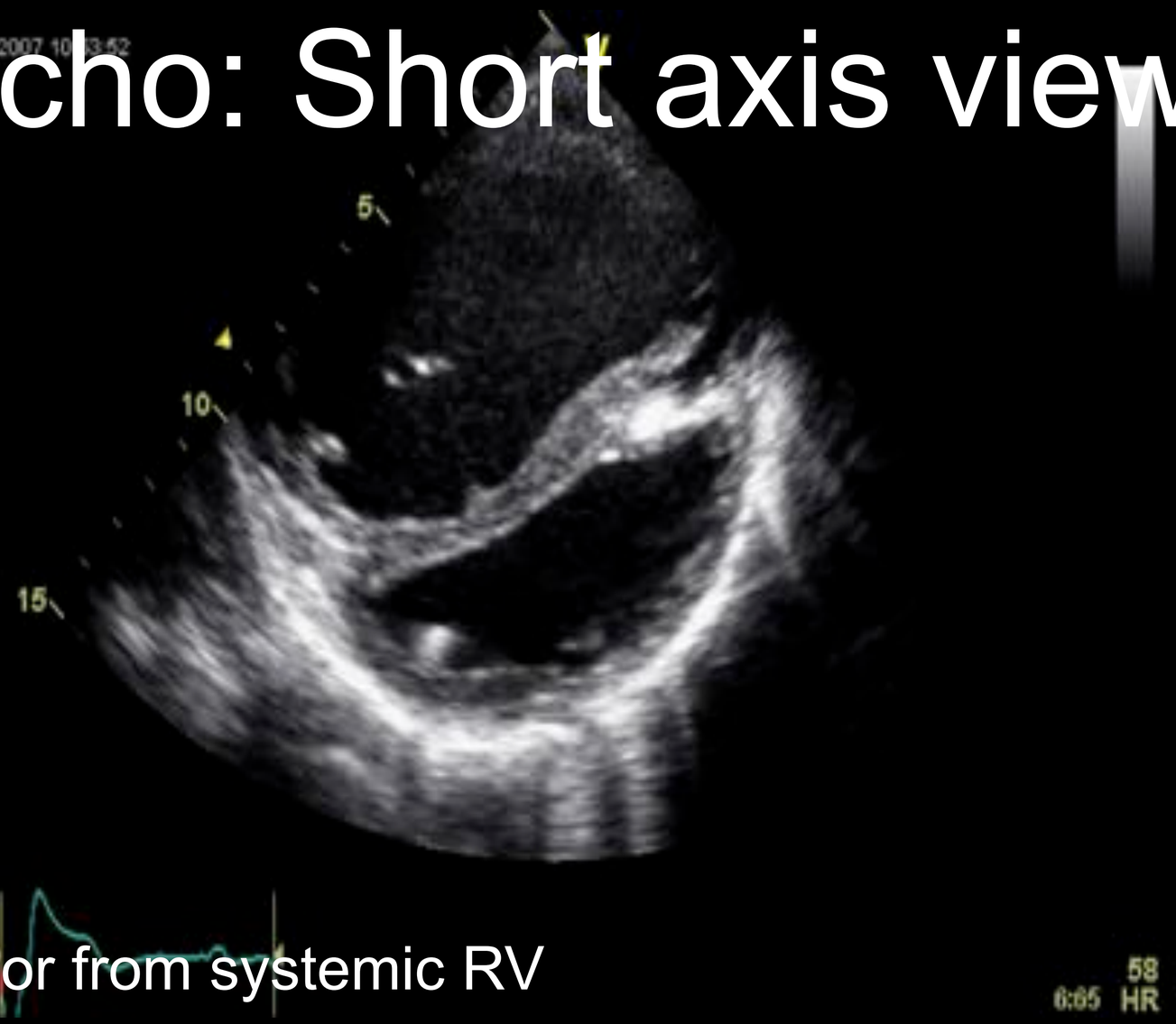
Aorta anterior, PA posterior

Echo: Long axis view

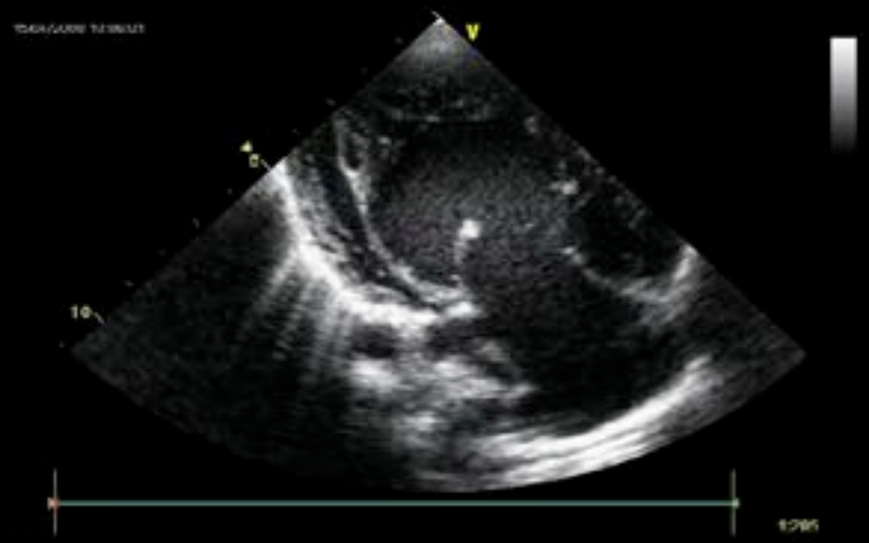


Flattened LV posterior from systemic RV

Echo: Short axis view

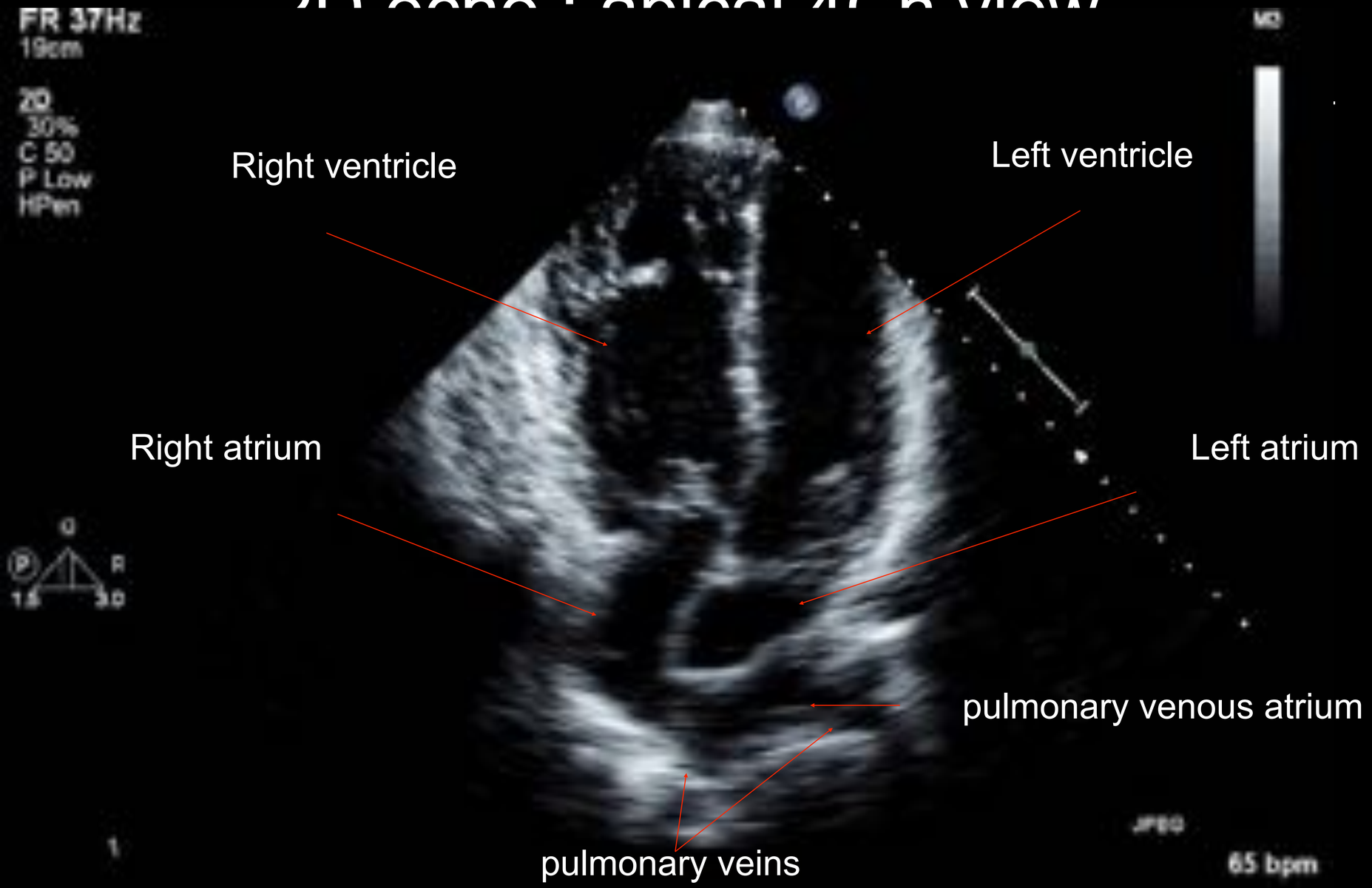


Flattened LV posterior from systemic RV

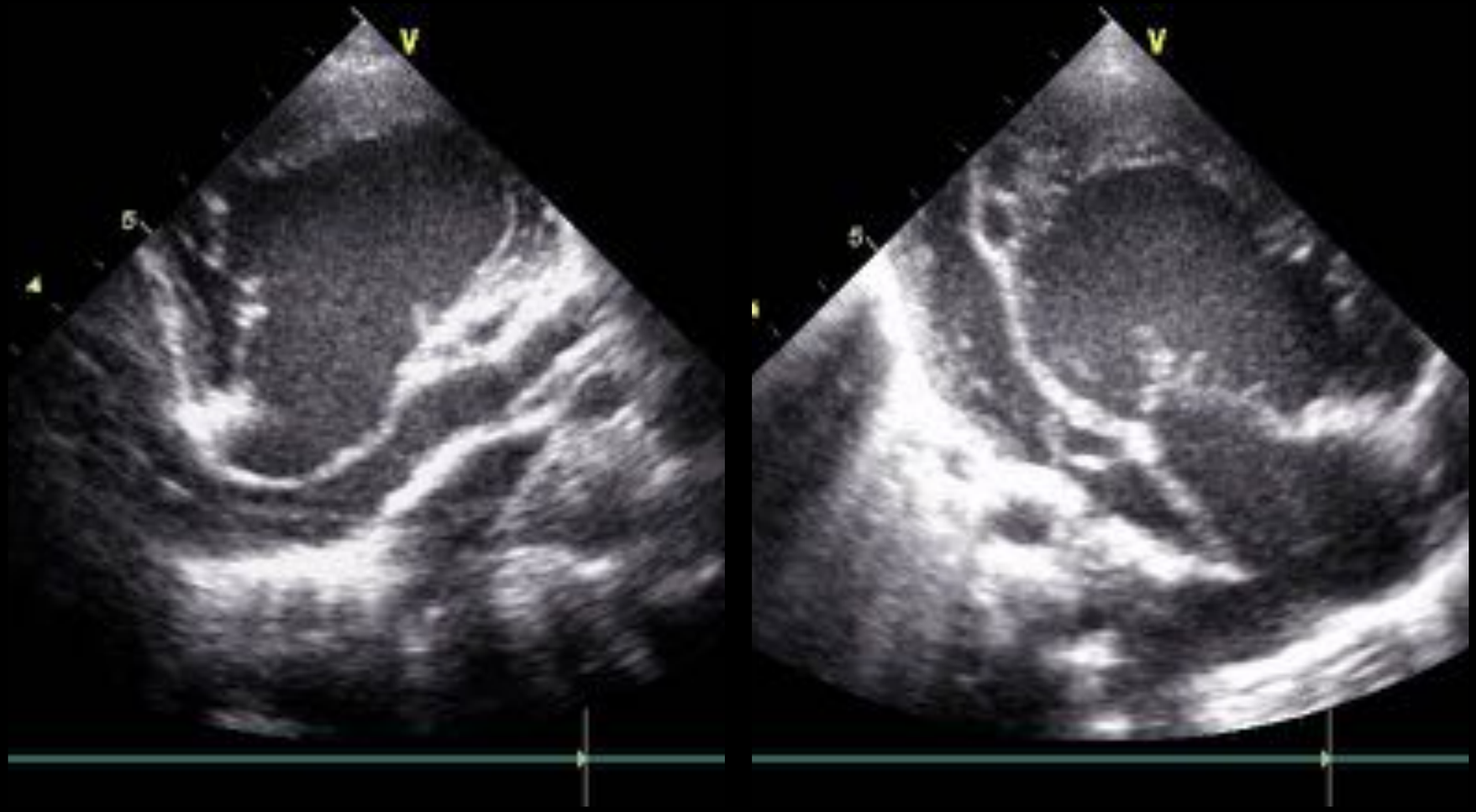


Senning in TGA

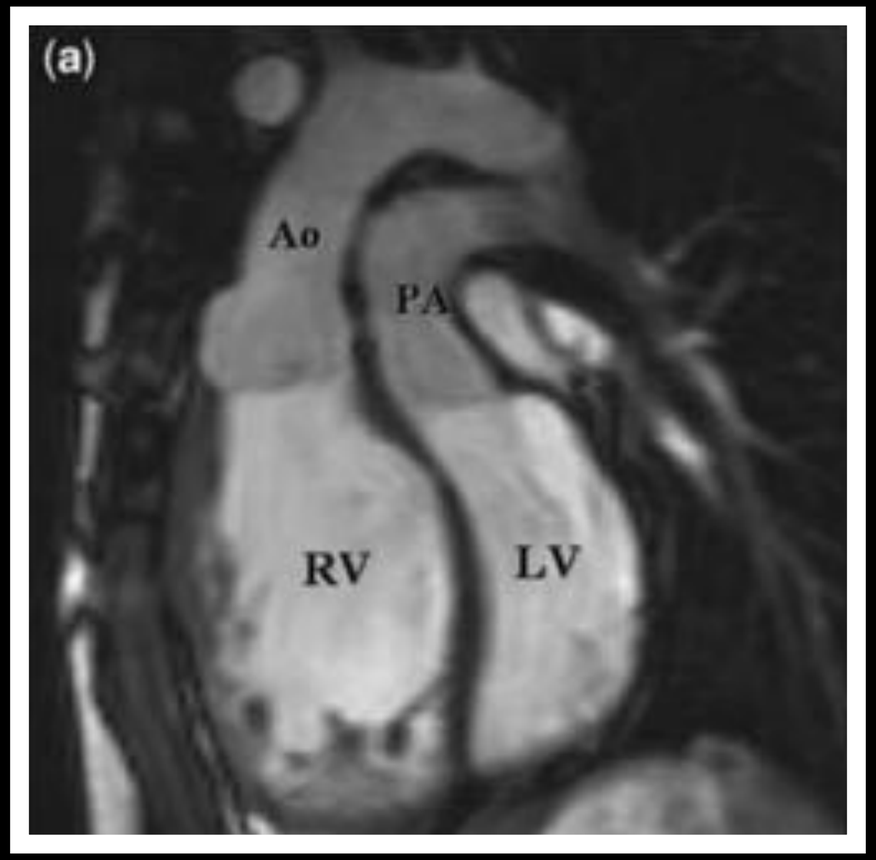
2D echo : apical 4Ch view

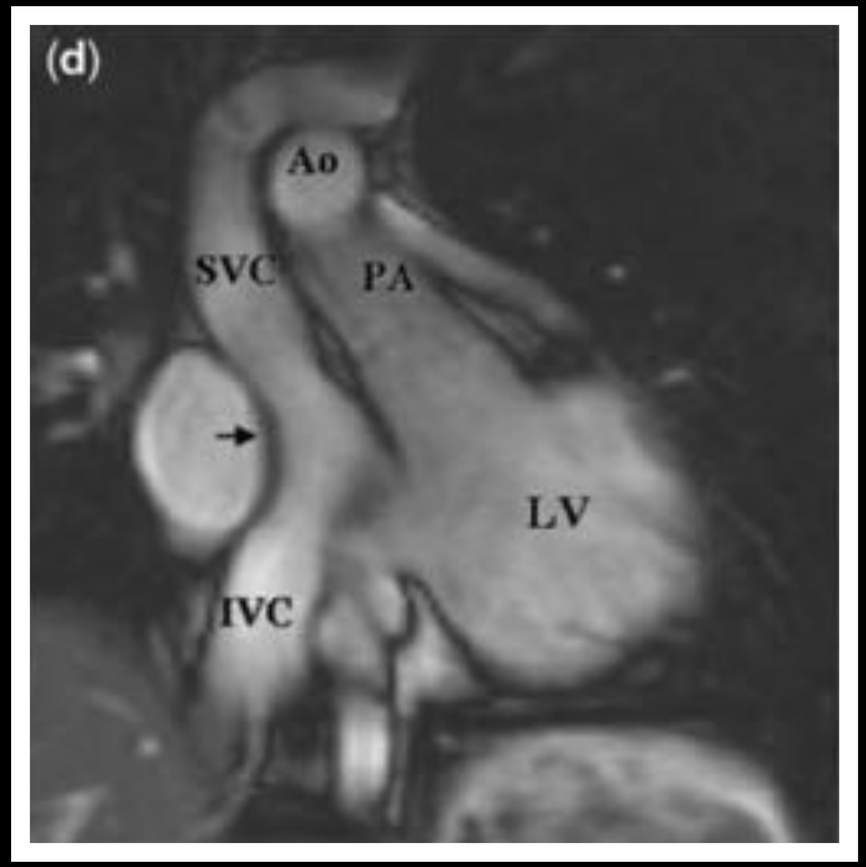


Caval bafflePulmonary veins baffle



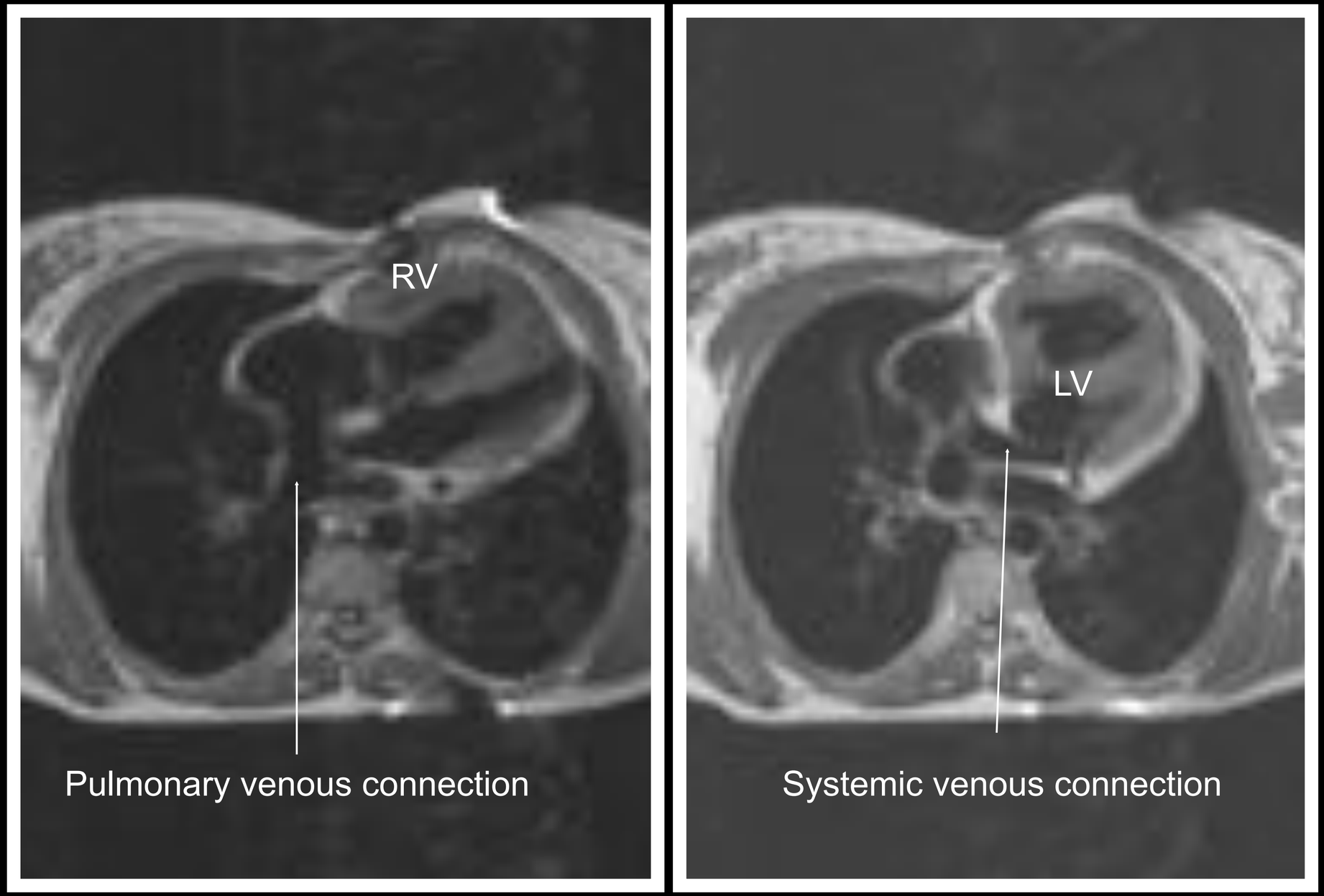
Arterial and atrial pathways in Mustard





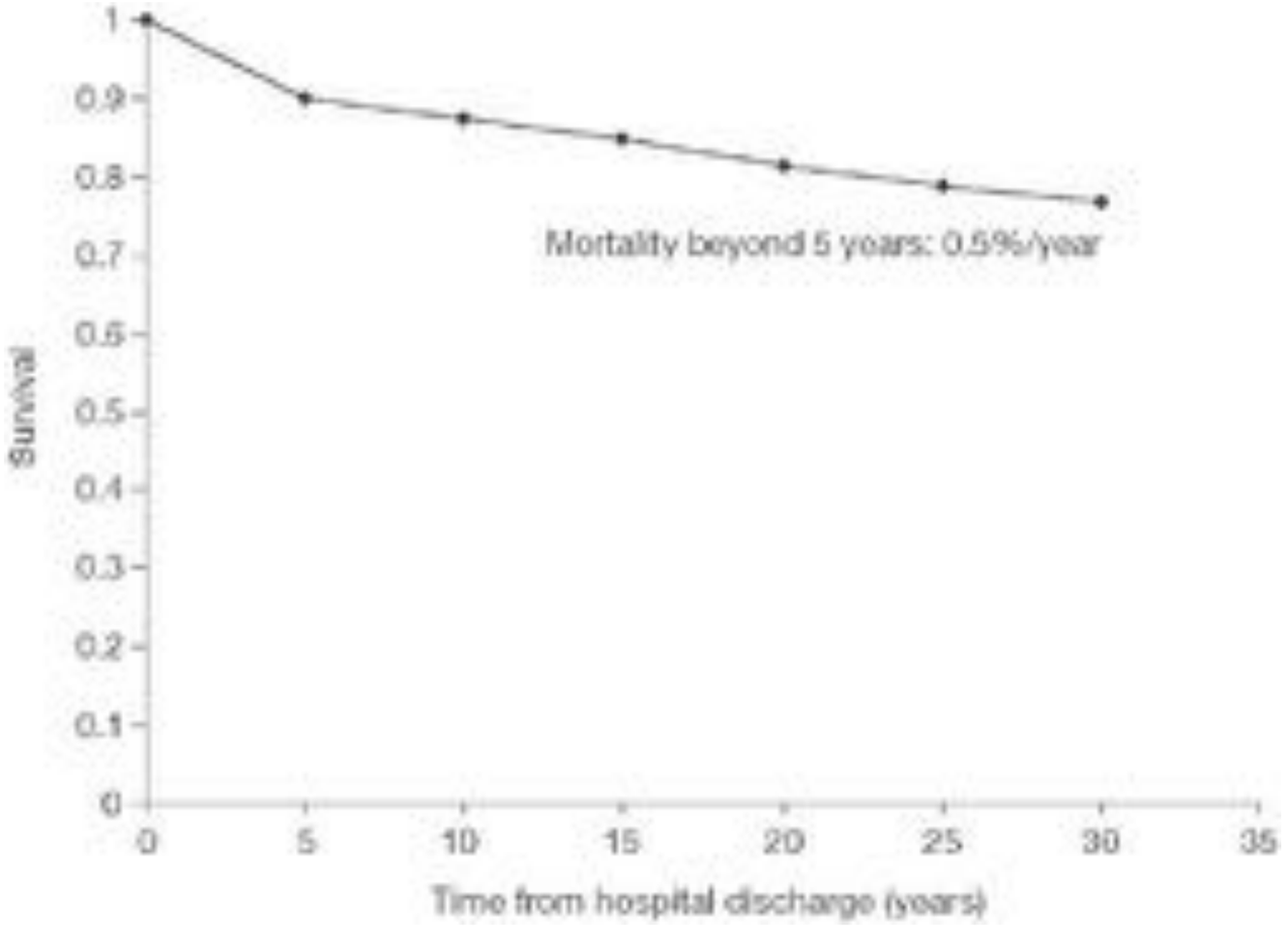
Johansson et al, Cardiol Young, 2010

MRI: Mustard procedure



Late complications after atrial switch

Early death



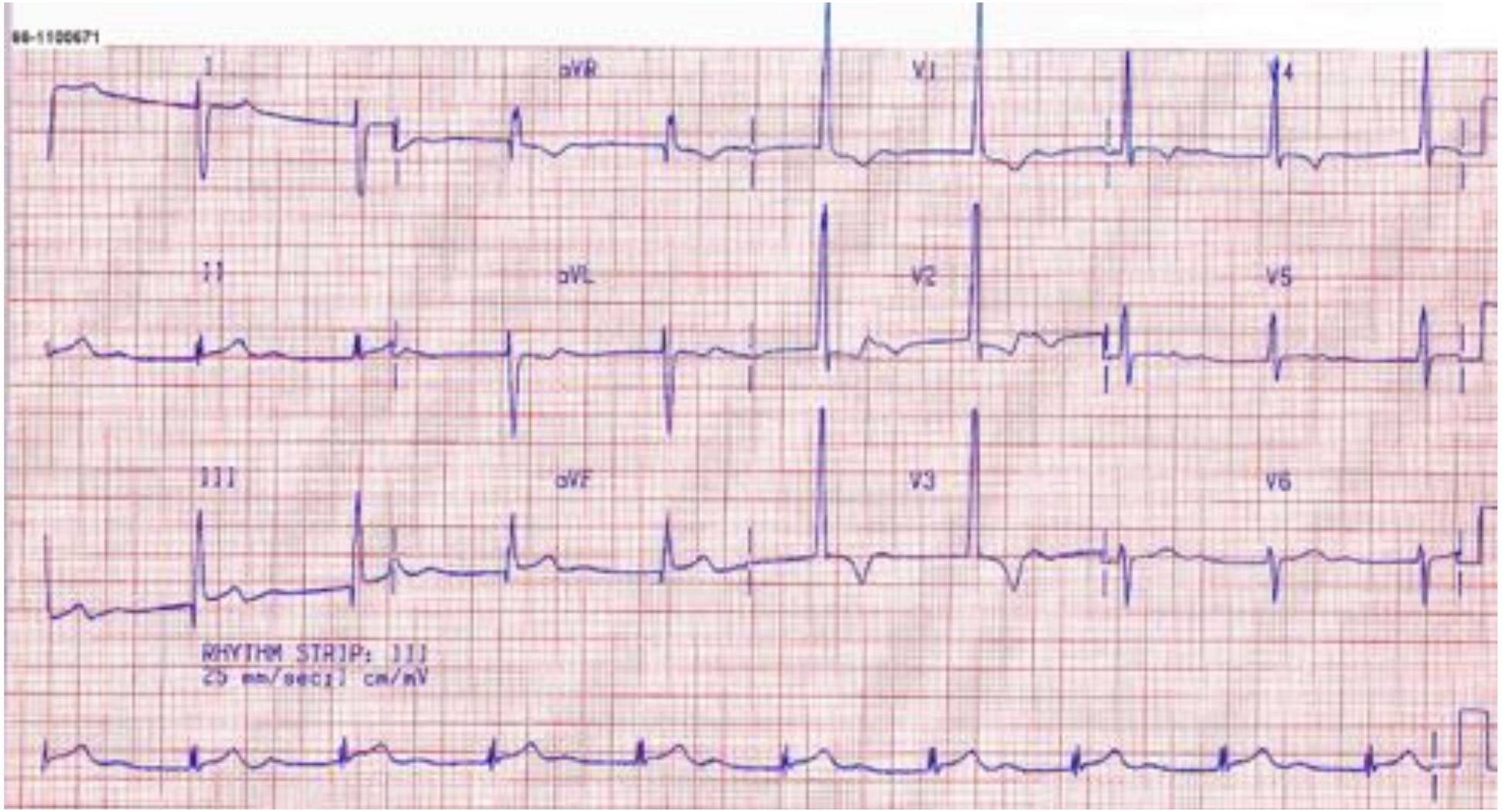
Love BA et al.Nat Clin Pract Cardiovasc Med 2008

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



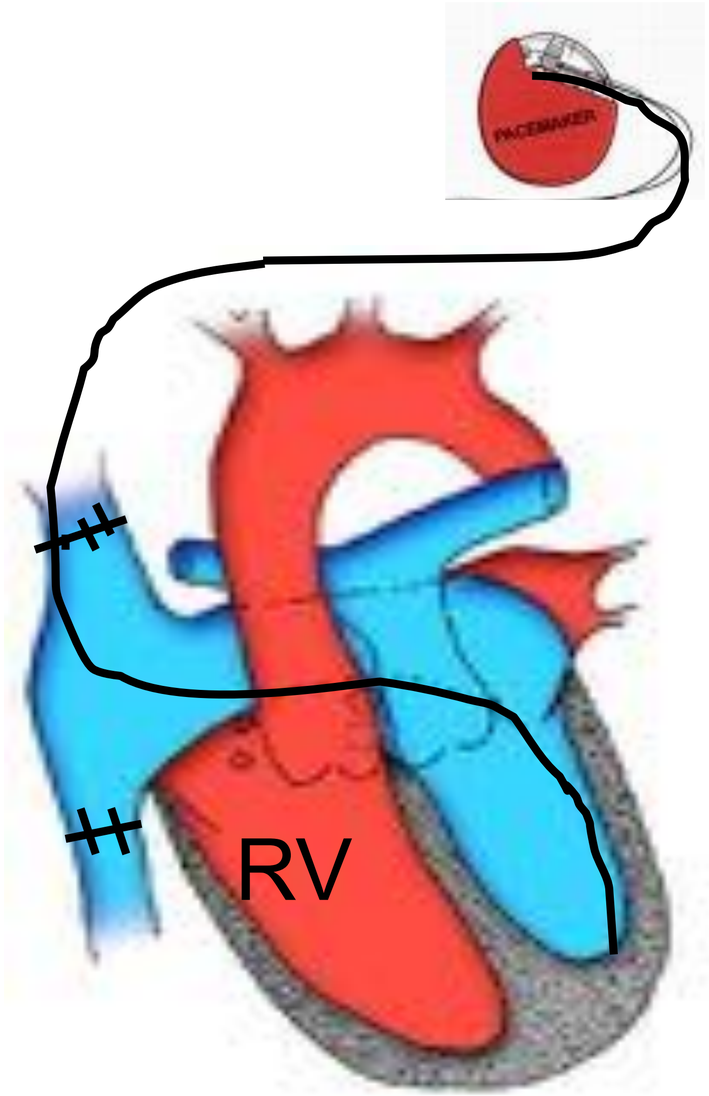
RVH

Right heart axis

Junctional rhythm

Only 40% of pts has SR at age 20

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



Be aware of altered venous connection:

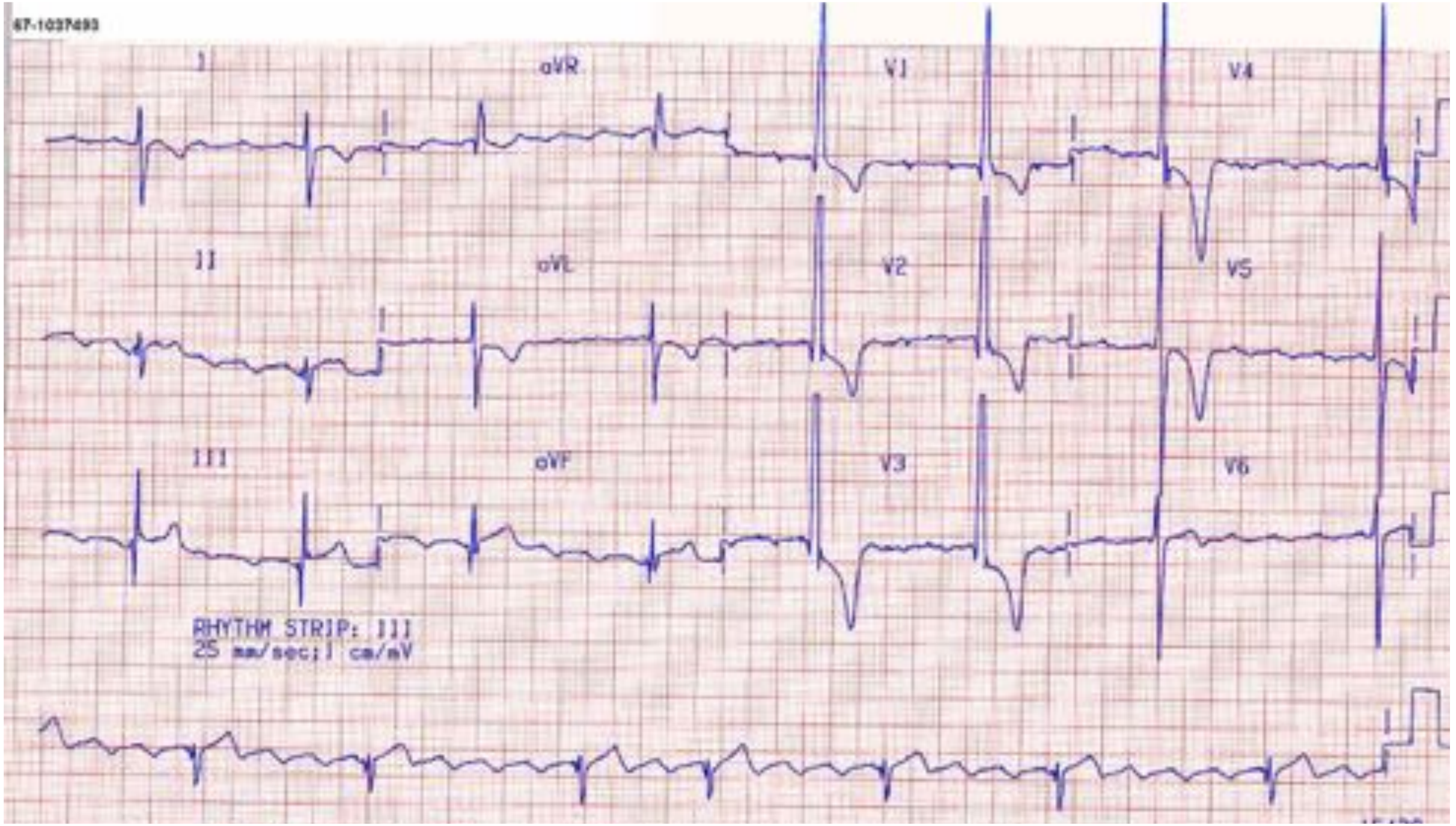
Ventricular lead will end up,

after some unusual loops,

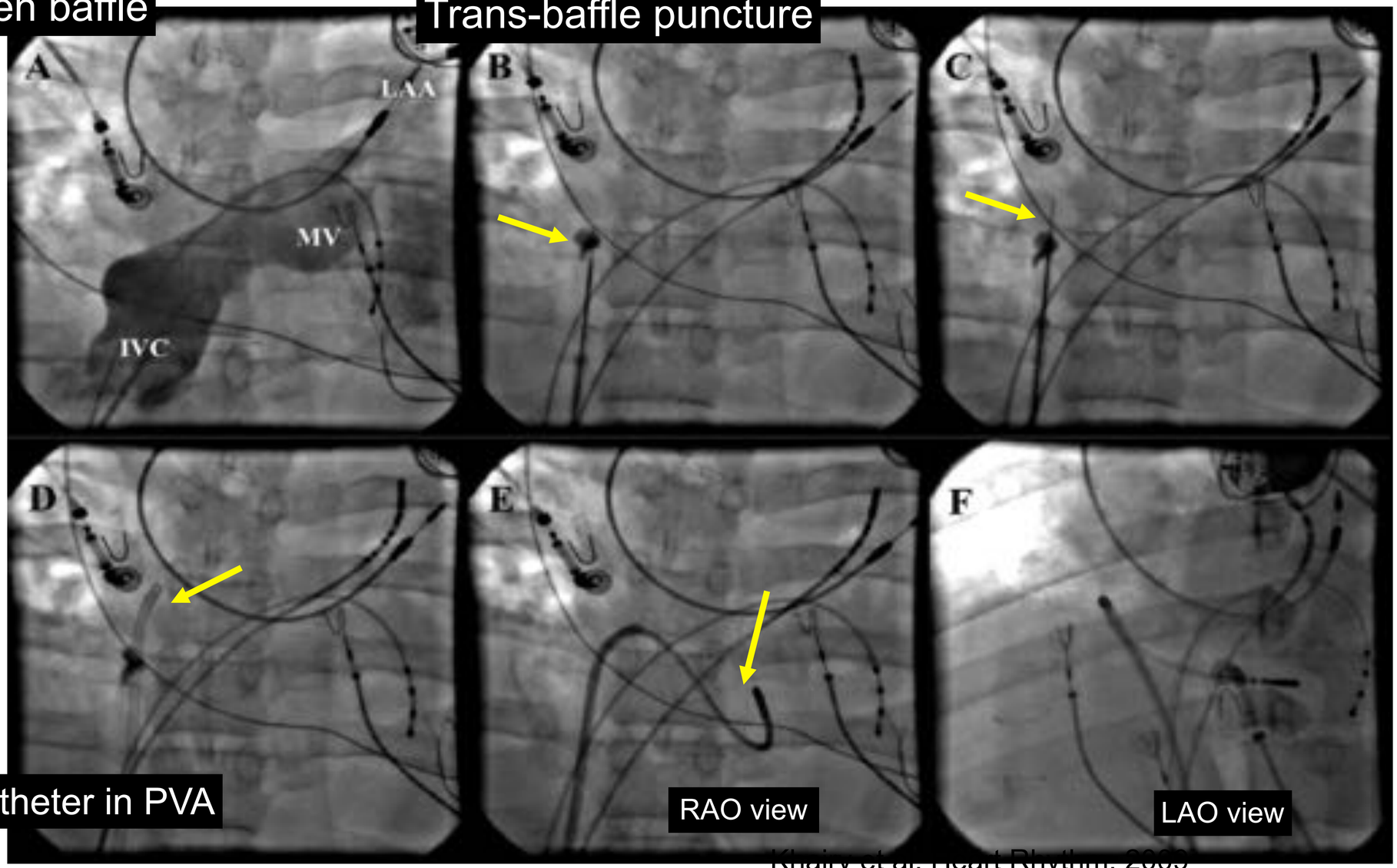
in a smooth-walled LV

Atrial flutter after atrial switch

Atrial arrhythmias may lead to hemodynamic instability and sudden death



Transbaffle puncture in IART

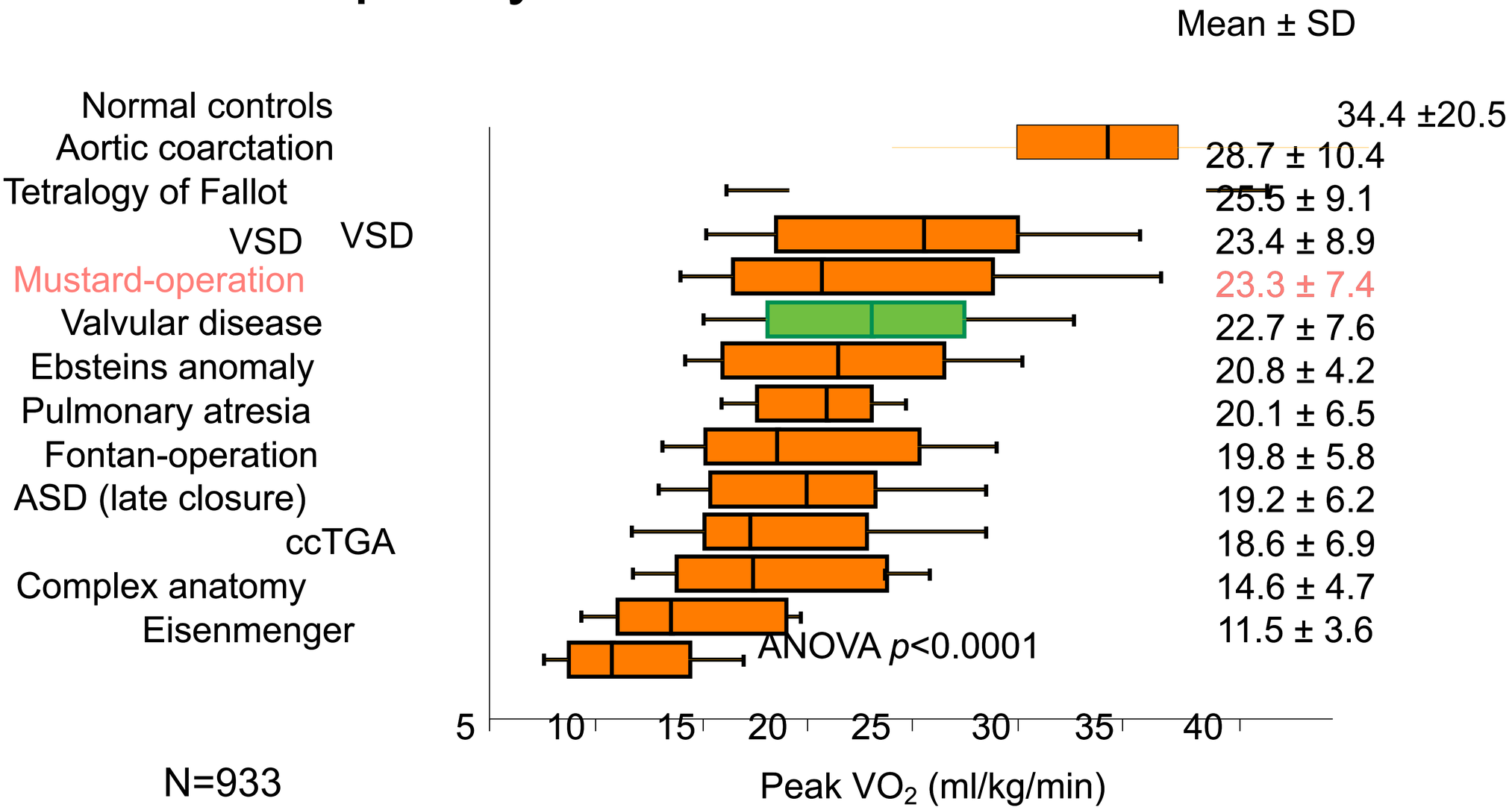


Khairy et al, Heart Rhythm, 2009

Late complications atrial switch

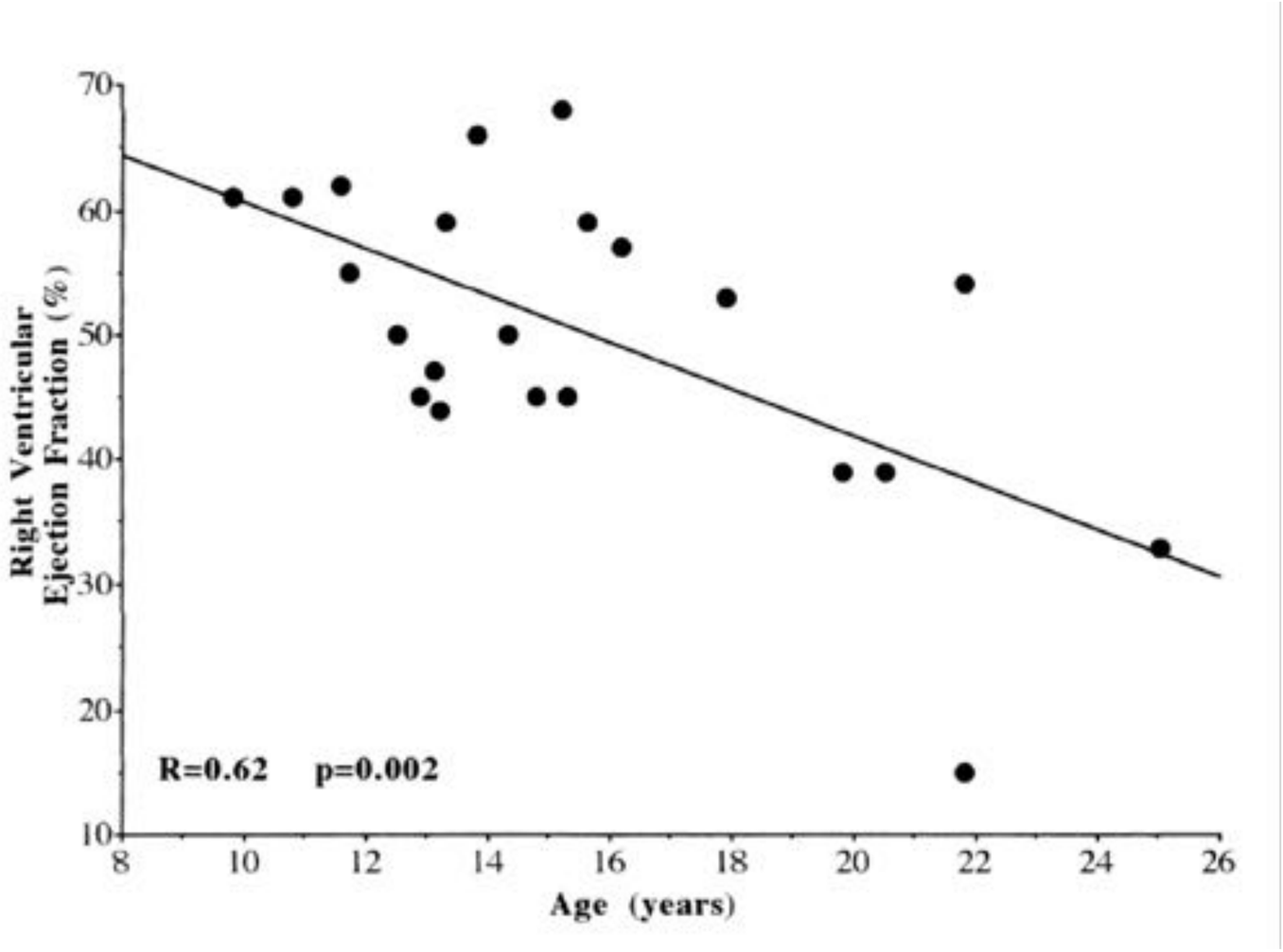
- Early death
- Arrhythmias
- Exercise capacity

Exercise capacity in CHD



Diller GP, et al. Circulation 2005;

Decline of RV function after atrial switch



Milane et al. , JACC 2000.

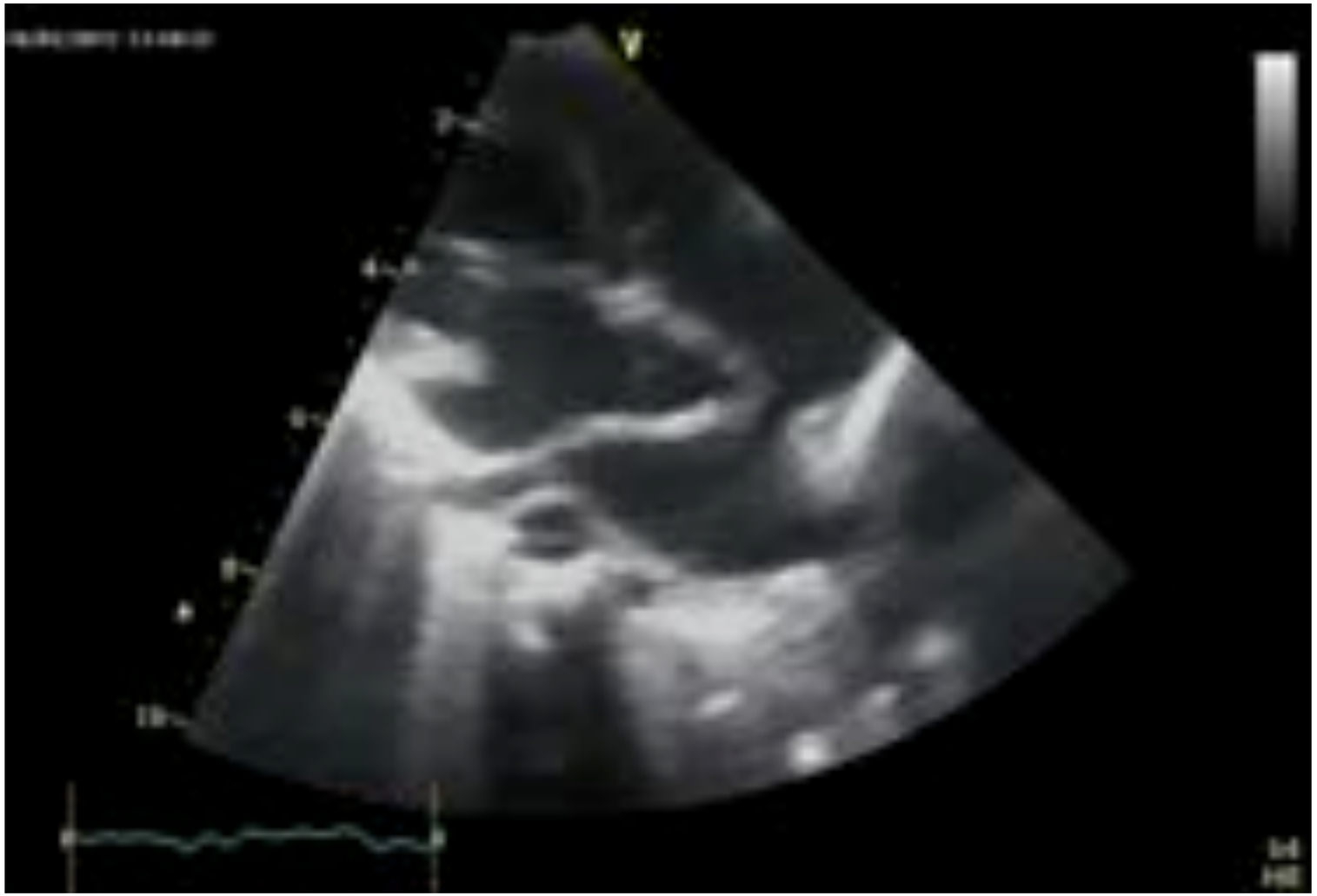
Late complications atrial switch

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

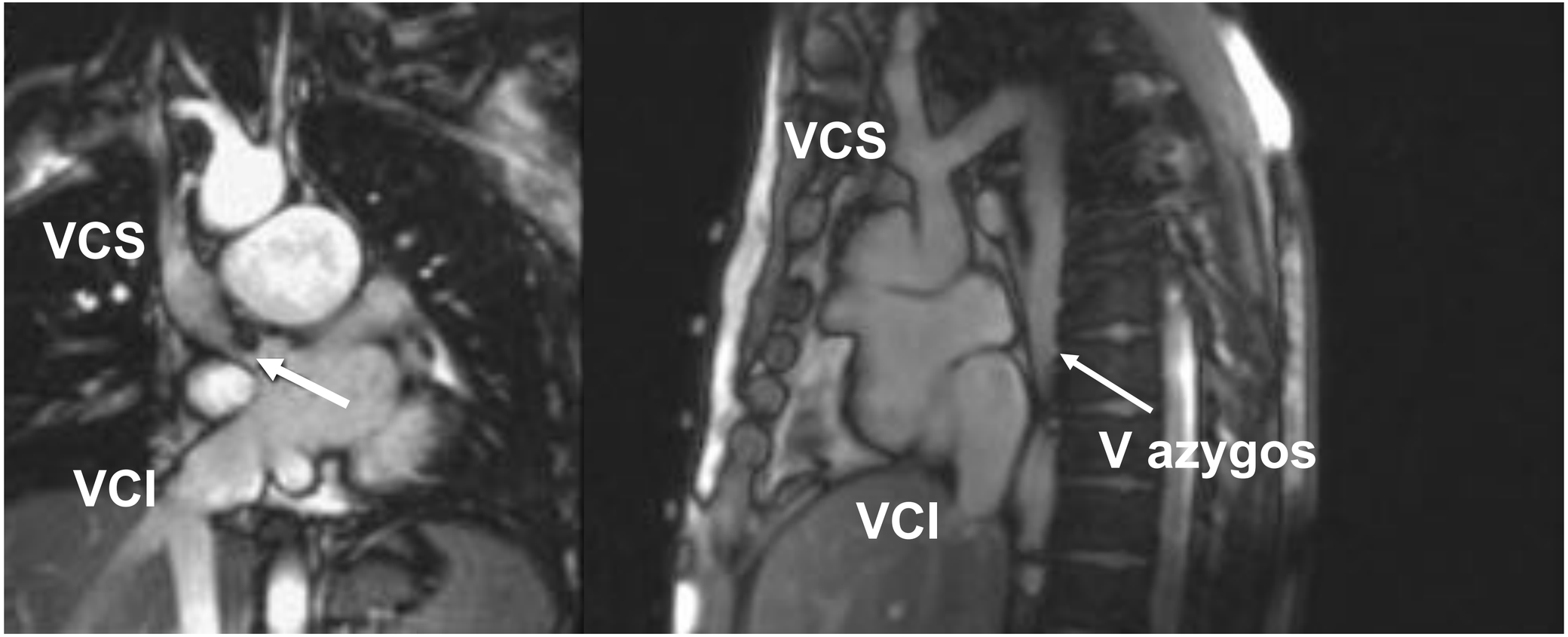
Abnormal septal configuration
RV dilation

Late complications atrial switch

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



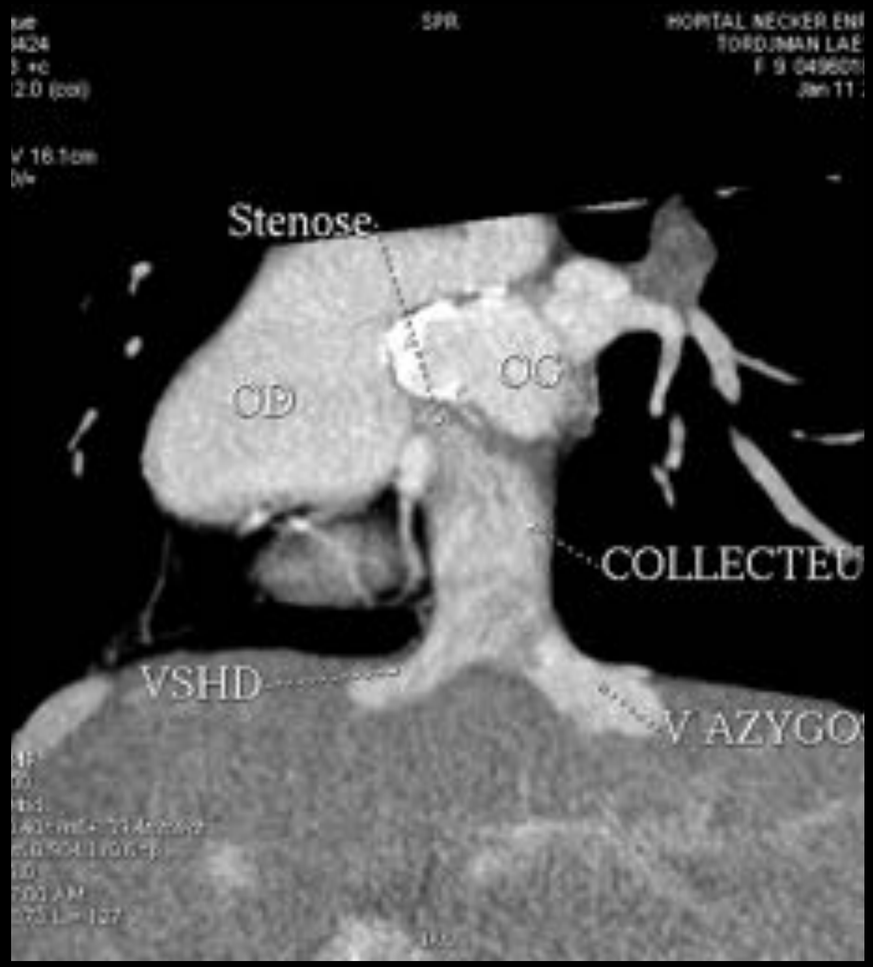
Baffle obstruction with increased azygos flow



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

obstruction Radiofrequency + Stent

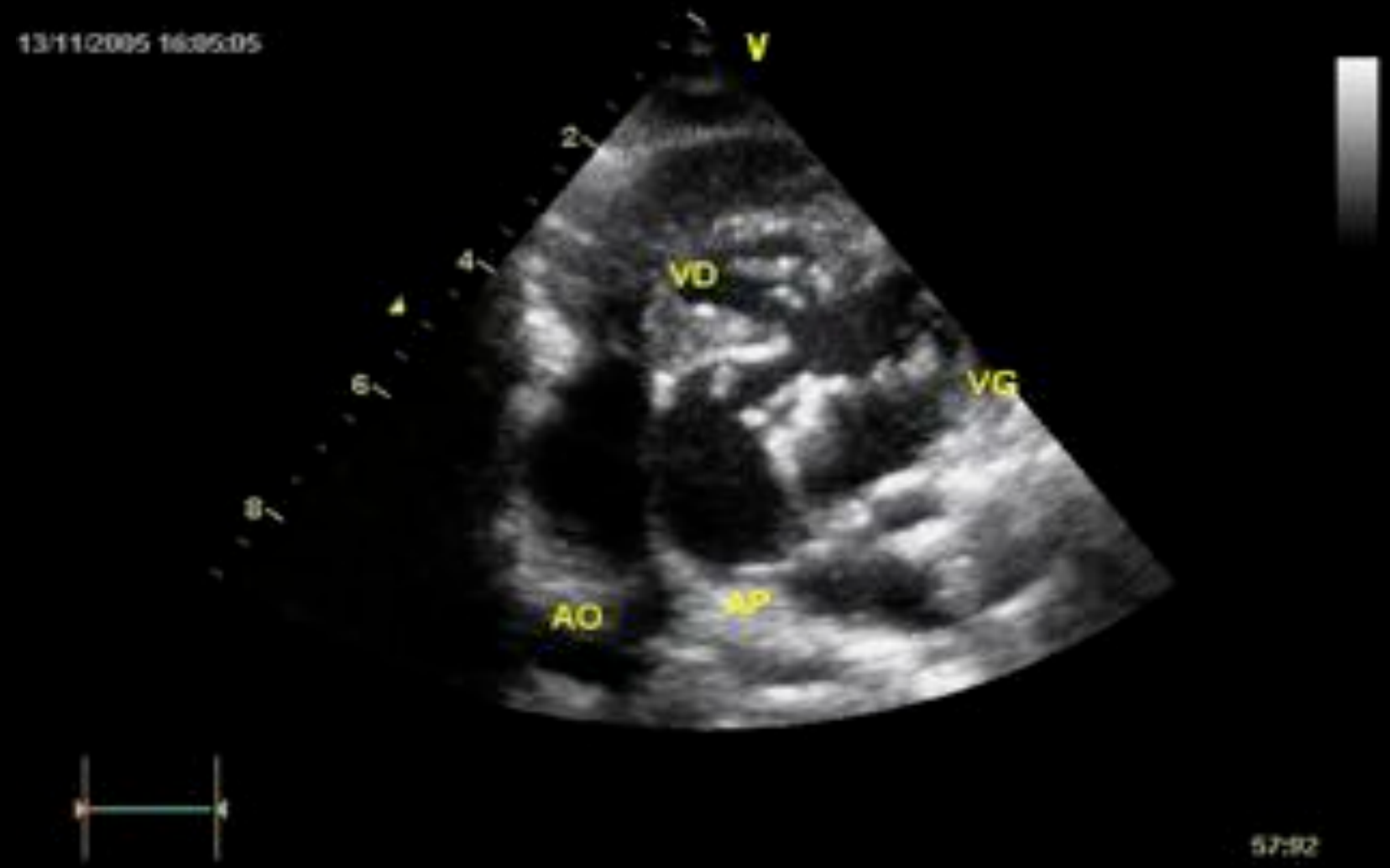
Love BA et al., Nat Clin Pract Cardiovasc Med 2008

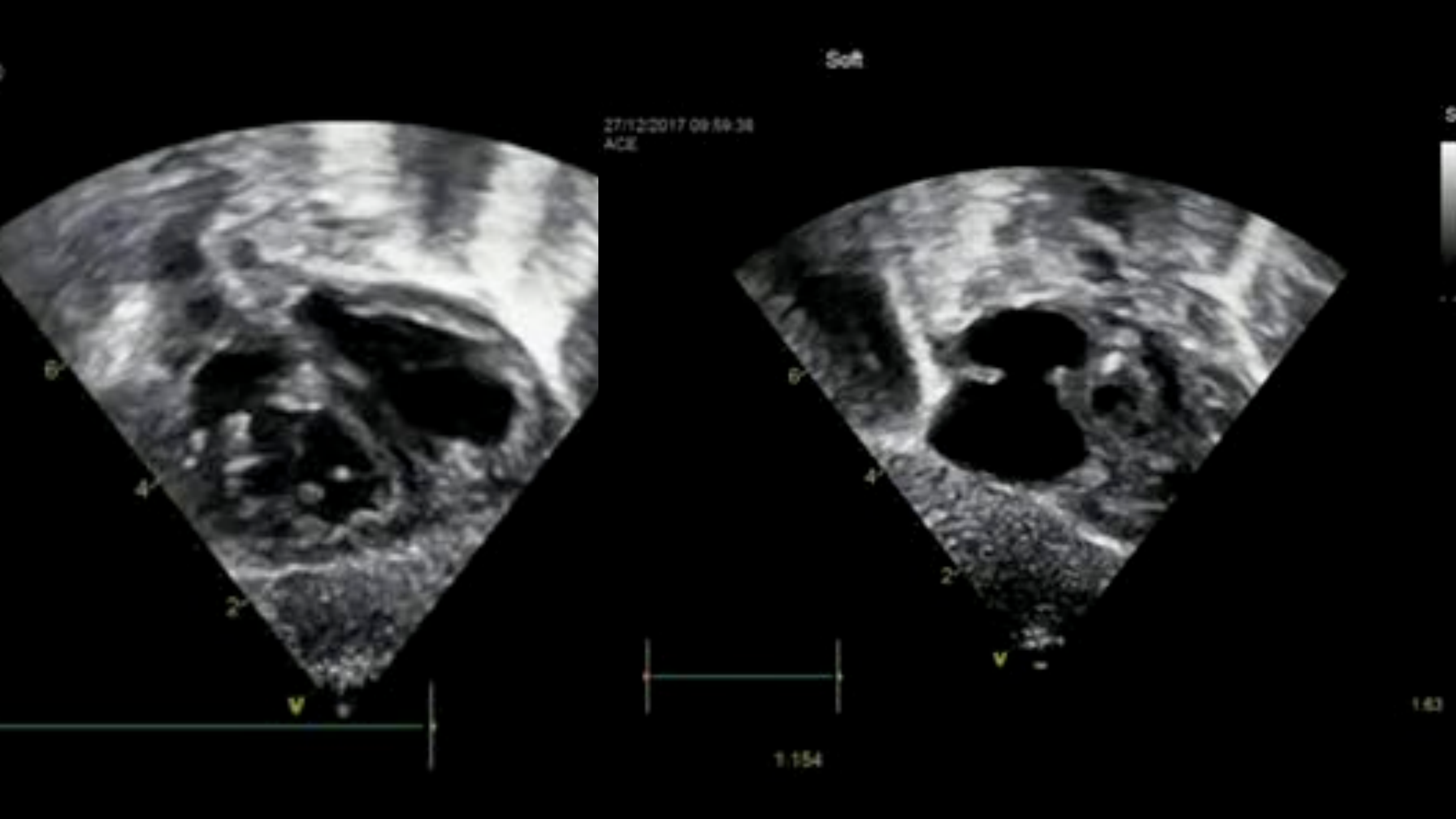

SPR
Stenose
OD
OG
COLLECTEU
VSHD
V AZYGO

Formes complexes

- L-TGV
- TGV + CIV
- TGV + Coarctation
- Anomalies des valves AV
  - Fente mitrale et straddling mitral
  - Straddling tricuspide

TGV + CIV + Sténose pulmonaire

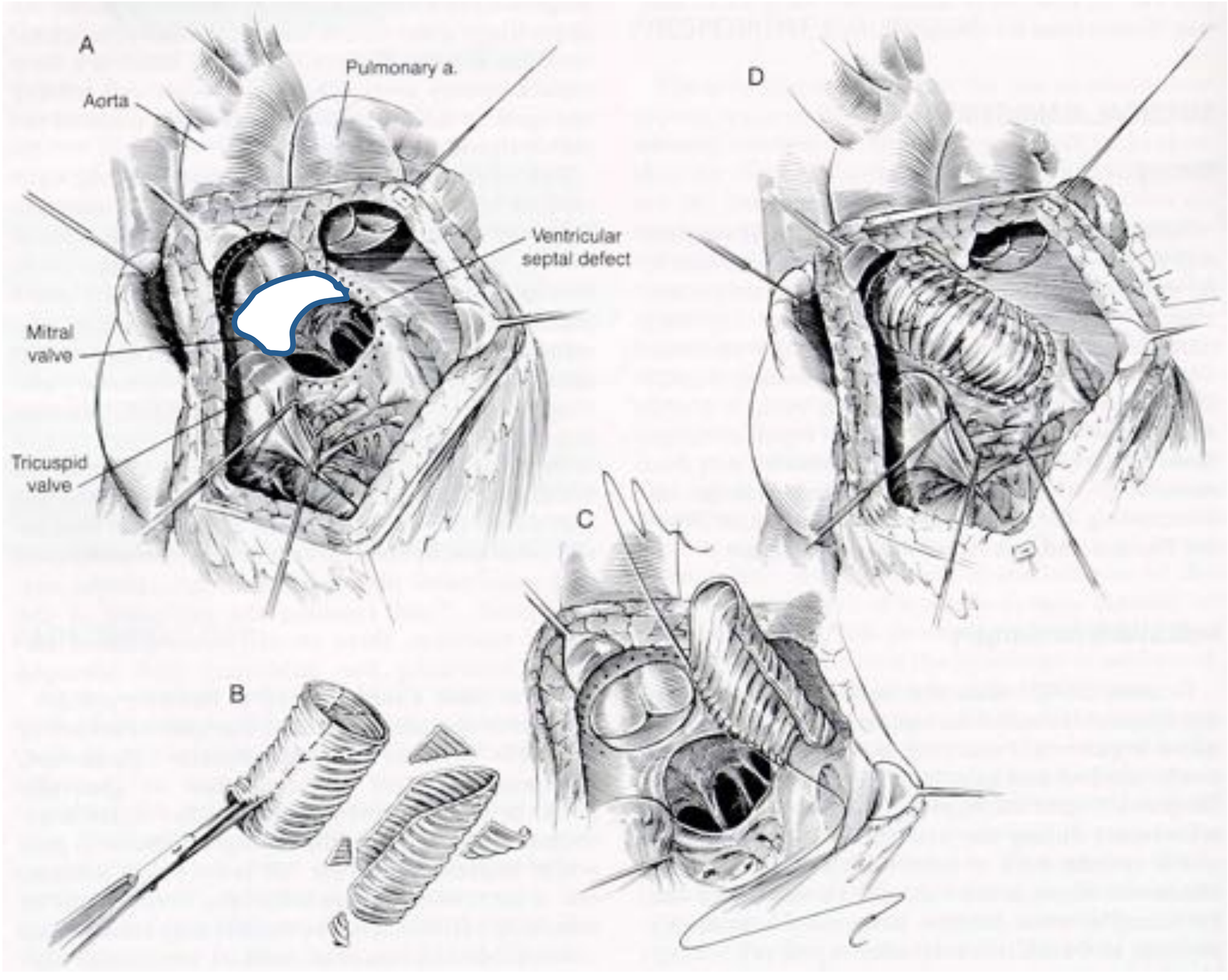

13/11/2005 16:05:05
V
2
4
6
8
VD
VG
AO
AP
57:92

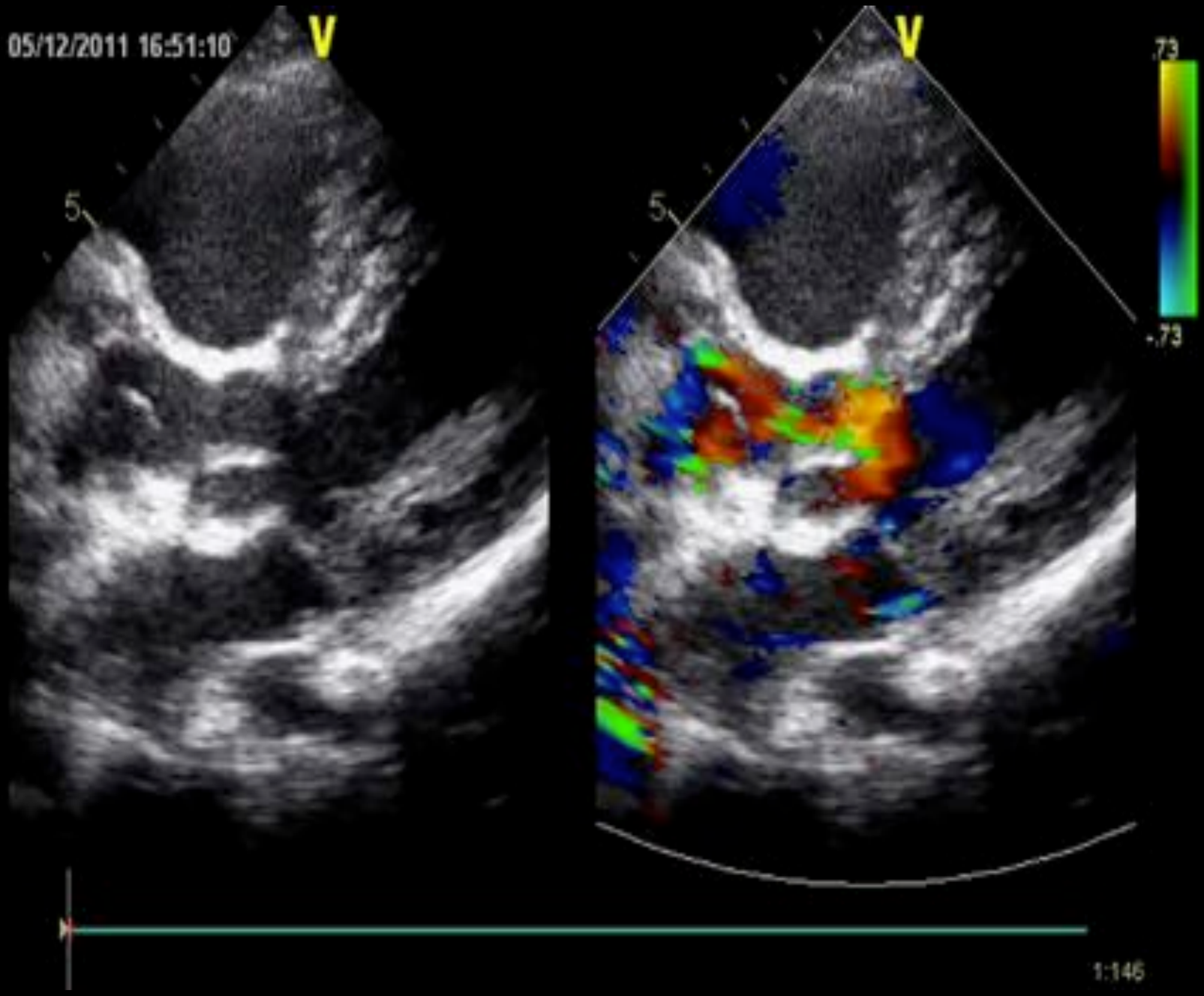

Soft
27/12/2017 09:59:38
ACE
6
4
2
V
1:154
143

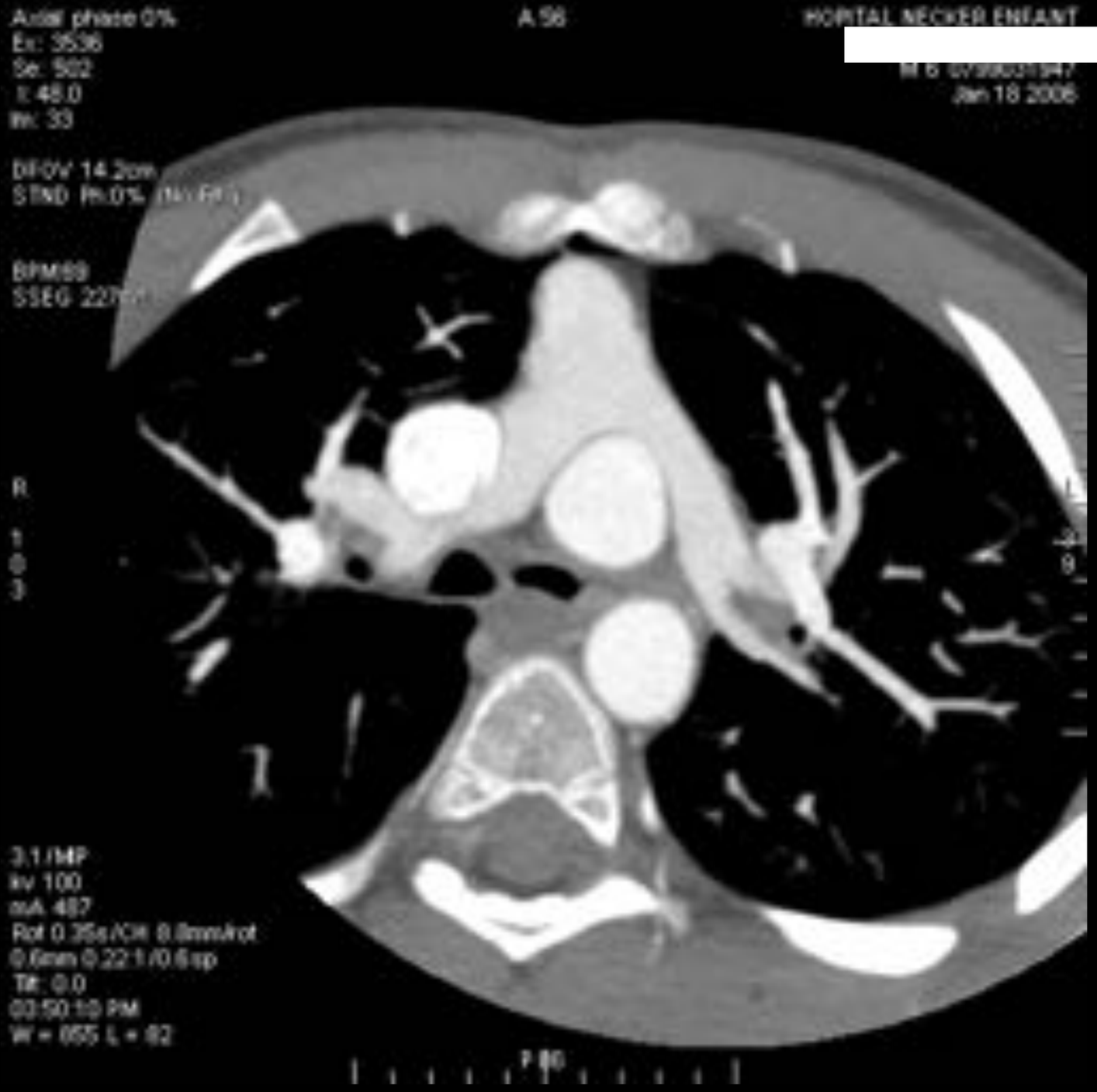
13:28:21

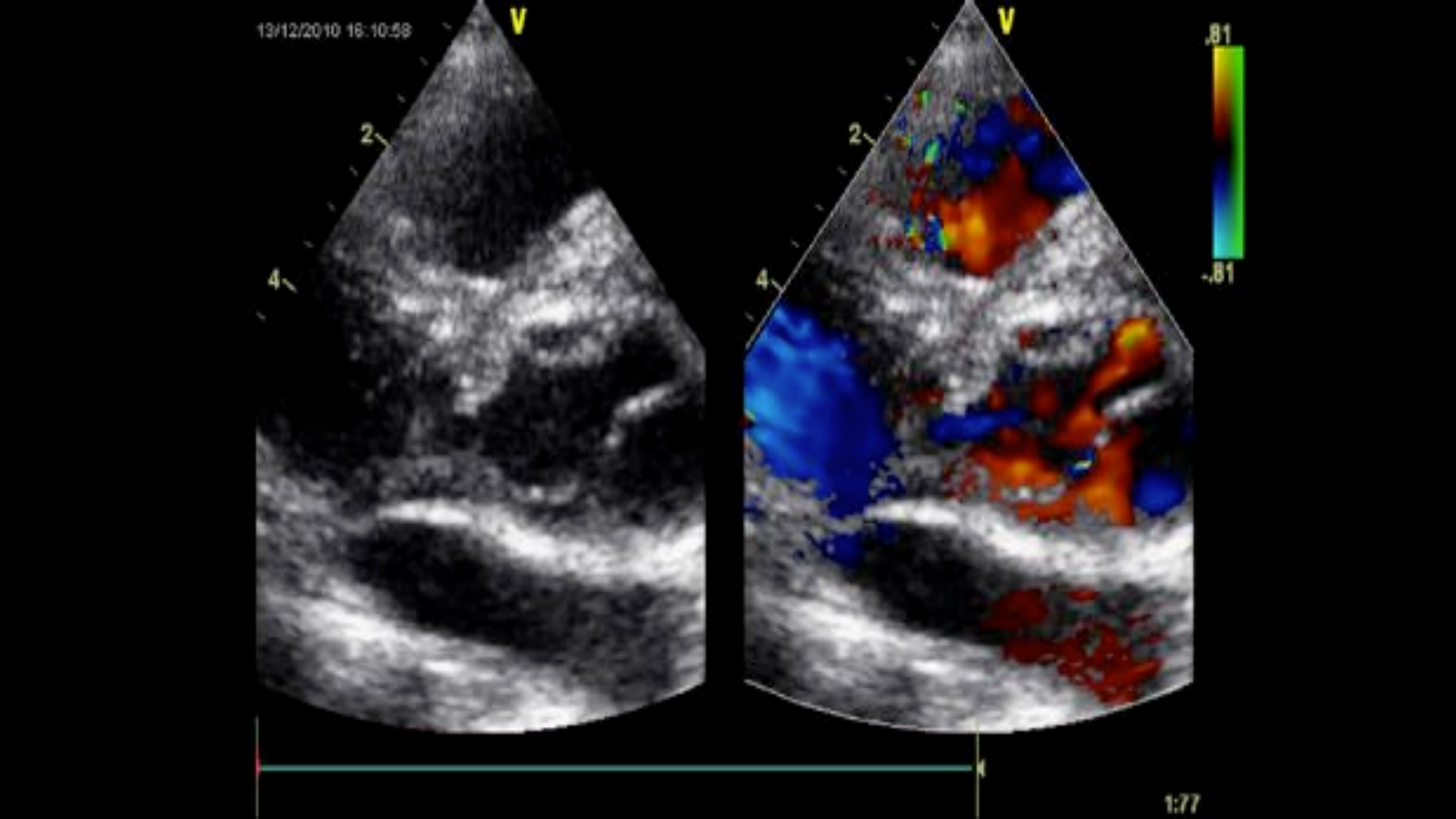
Clinique

- Cyanose + souffle
- Prise en charge néonatale
  - Rashkind souvent indiqué
  - Blalock risque de fixer les branches des AP
  - REV

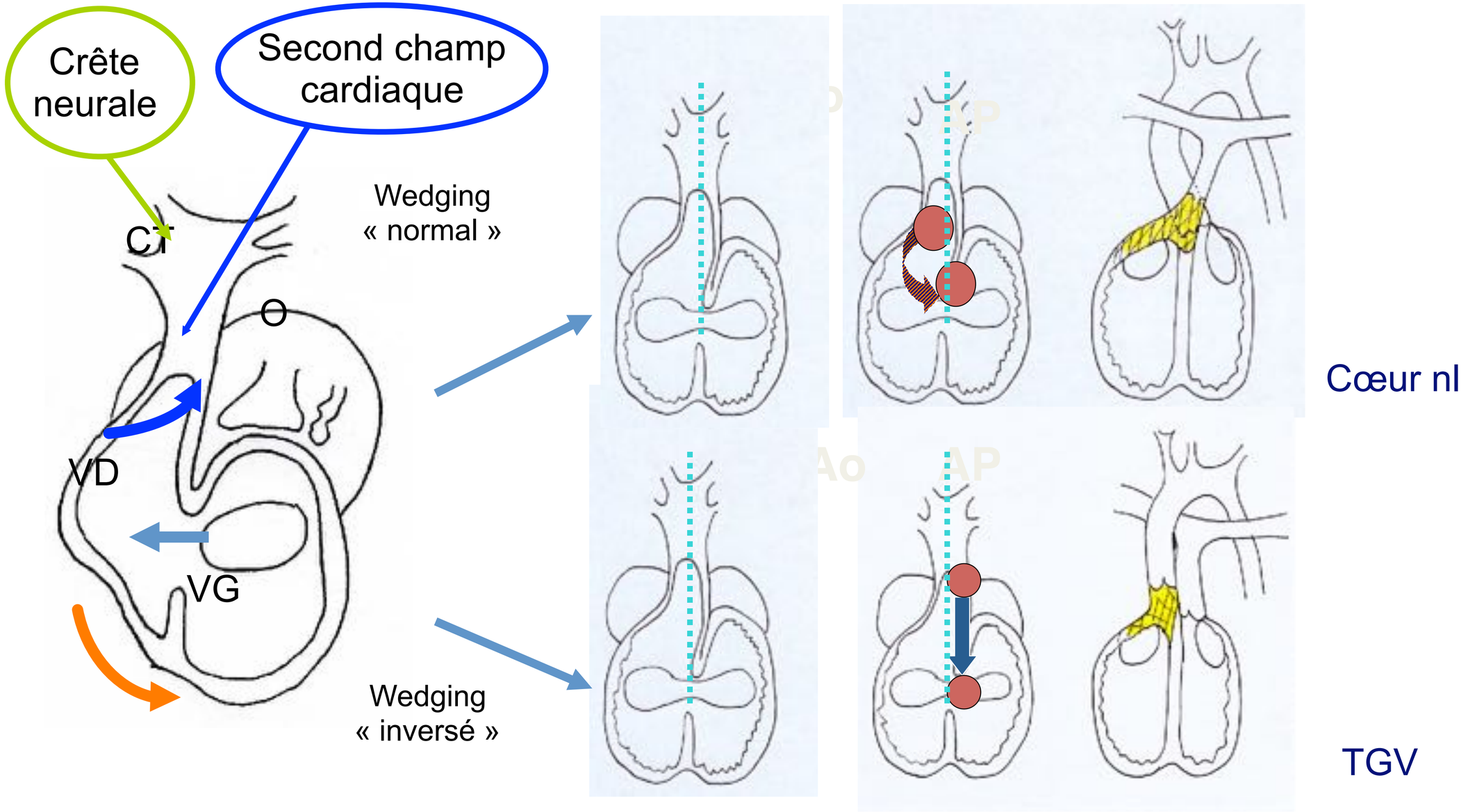

A
Pulmonary a.
Aorta
Ventricular septal defect
Mitral valve
Tricuspid valve
D
C
B


05/12/2011 16:51:10
V
5
V
5
.73
-.73
1:146


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


13/12/2010 16:10:58
V
2
4
V
2
4
.81
-.81
1:77

Ventricules droits à double issue

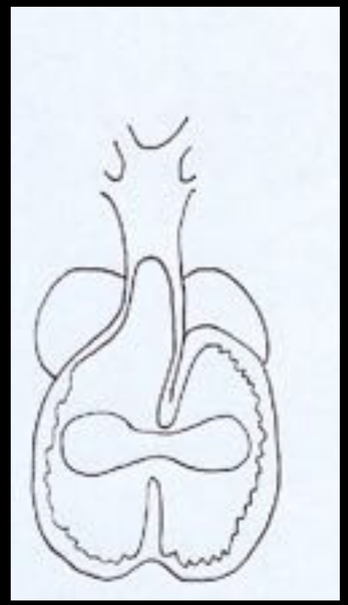


Early looping Convergence Wedging

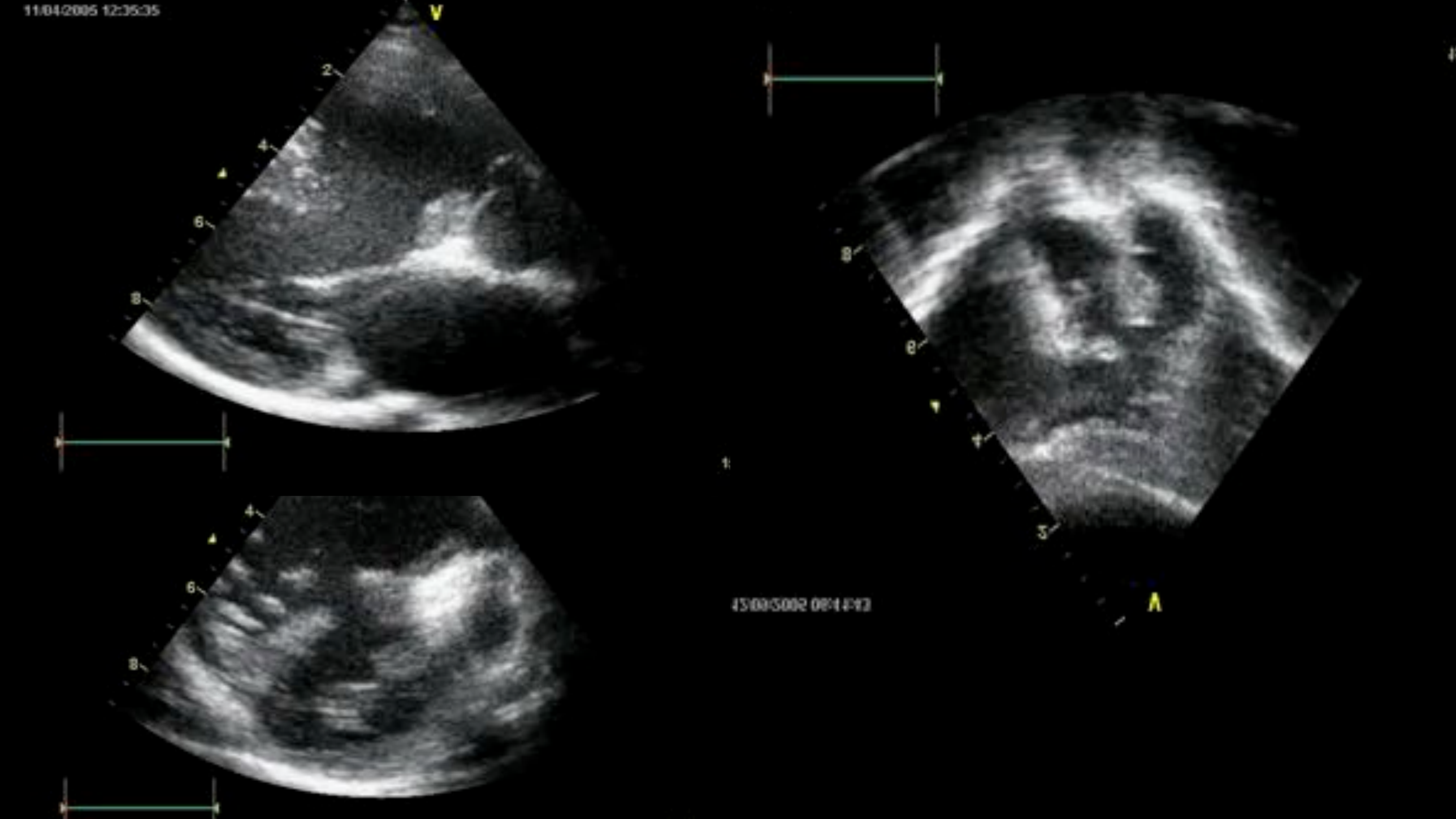
VDDI :mécanismes embryologiques
3 groupes (Van Praagh)

- Groupe 1 : VDDI avec anomalies seulement au niveau du conotruncus
  VDDI « tardifs » par insuffisance de wedging
- Groupe 2 : VDDI avec anomalies du conotruncus + des ventricules (VG) et des valves AV VDDI « précoces » au stade du « early looping »
- Groupe 3 : anomalies de la loop elle-même hétérotaxies

Plus l'anomalie survient tôt dans le développement,
plus la malformation est complexe

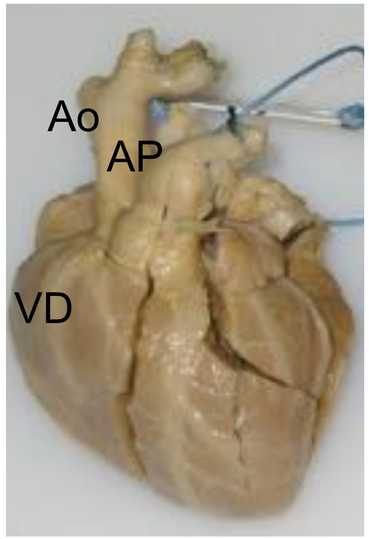


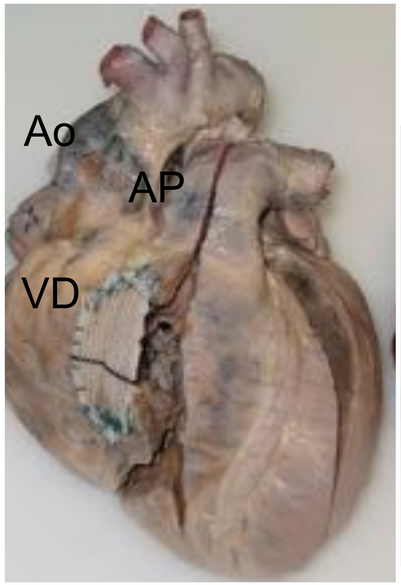
26/03/2008 10:31:32
6
4
2
V

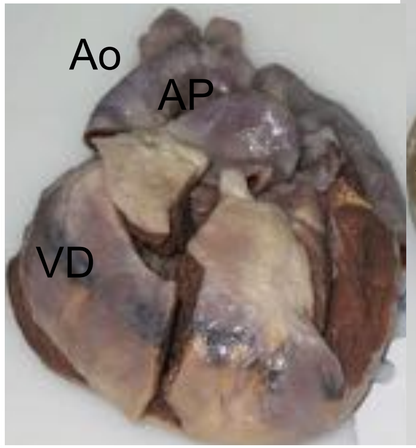

11/04/2005 12:35:35
V
2
4
6
8
4
6
8
8
6
4
3
A

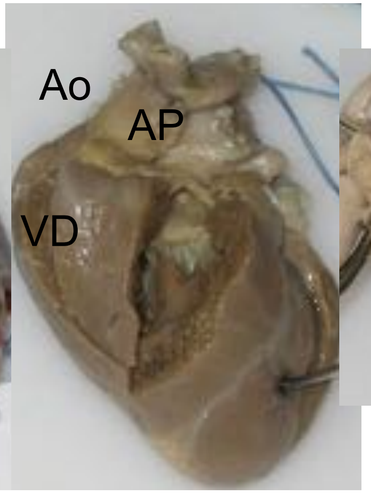
VDDI

- Le VDDI n’est qu’un des 4 types de malposition des gros vaisseaux :
  - TGV
  - VDDI
  - VGDI
  - Malposition anatomiquement corrigée des gros vaisseaux


Ao
AP
VD


Ao
AP
VD


Ao
AP
VD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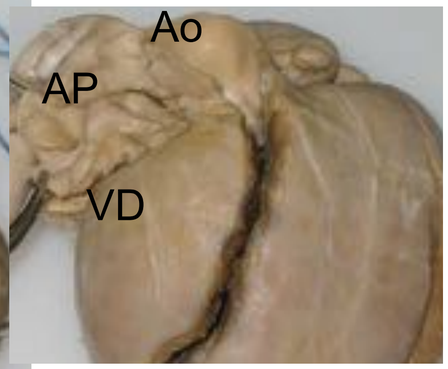
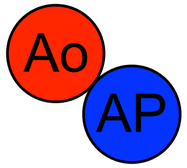
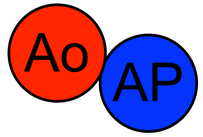
AP
VD


Ao
AP
VD

S,D,L


Ao
AP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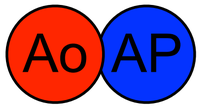
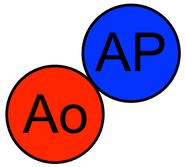
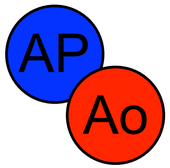
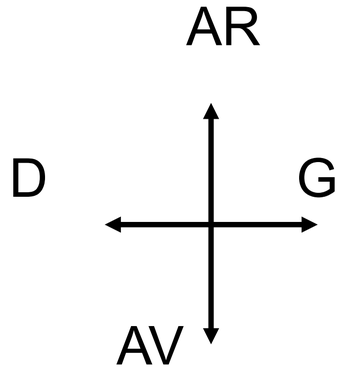
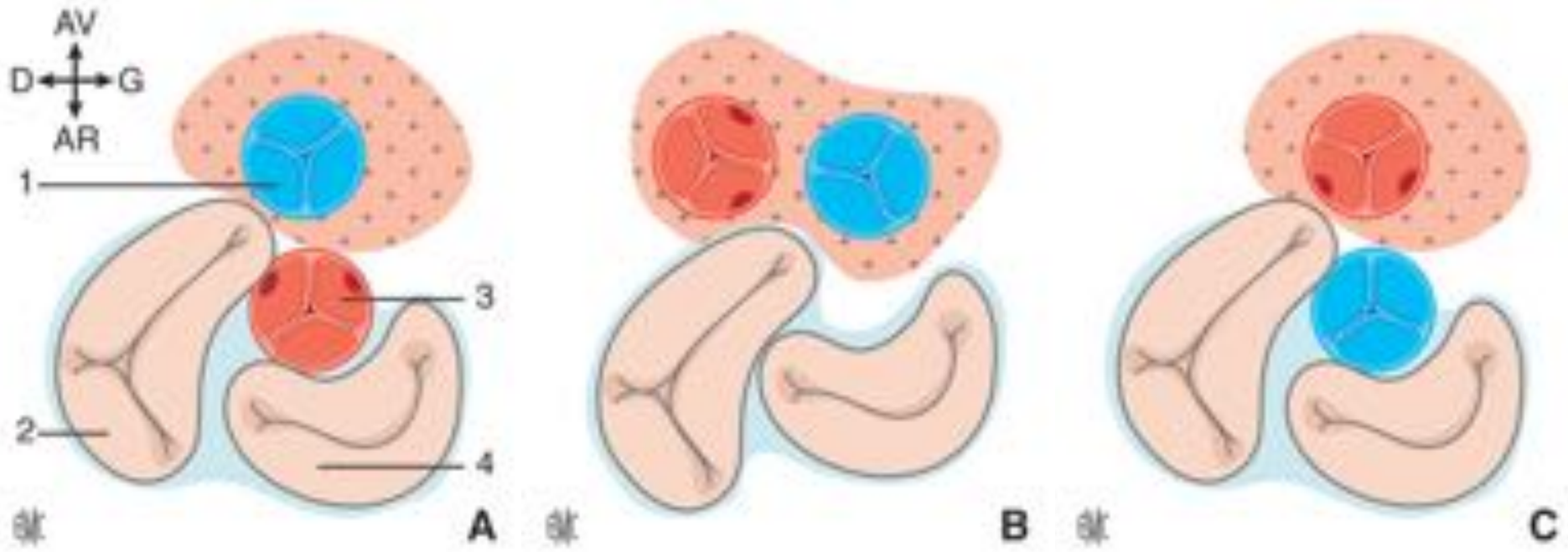
AP


Ao
AP

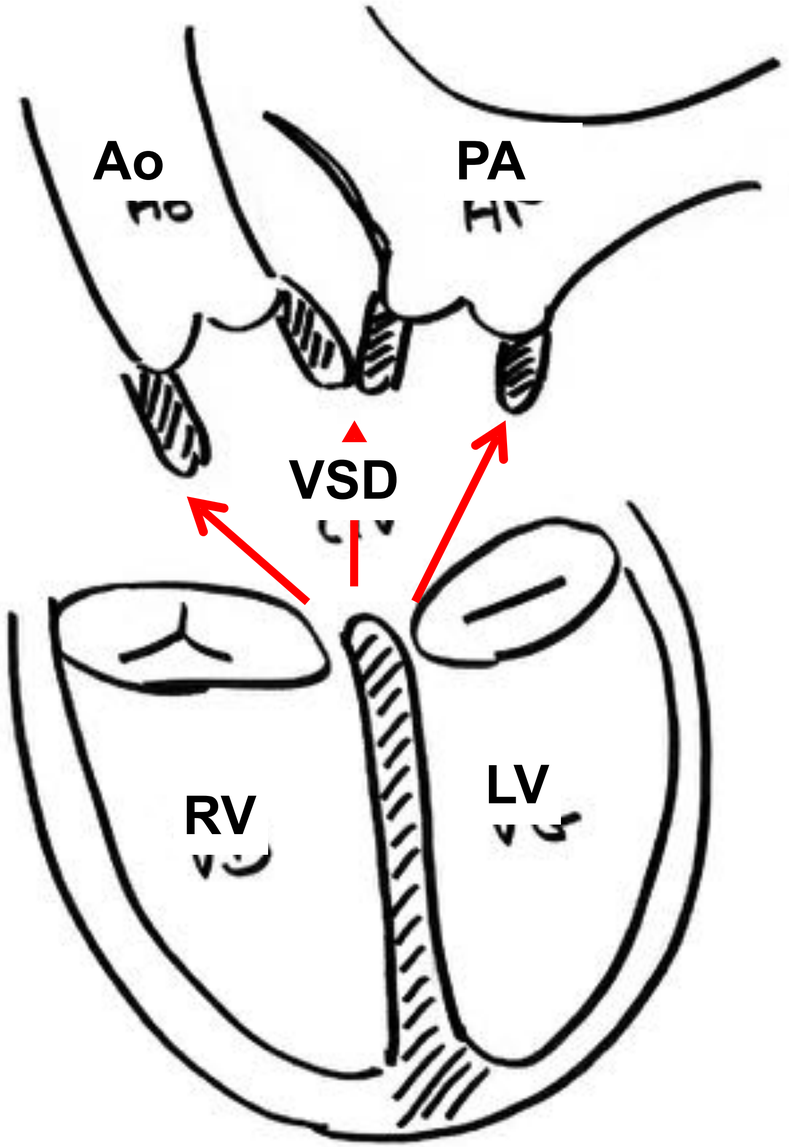

AP
Ao


AP
Ao

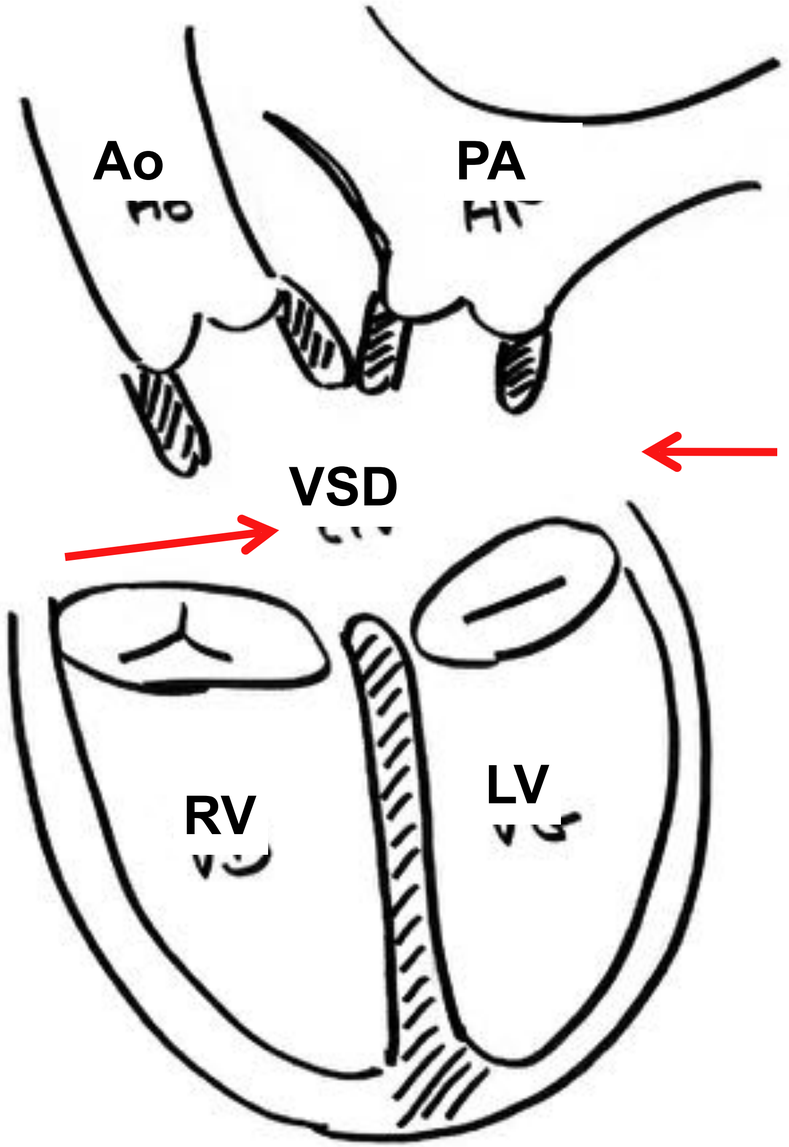

AR
D
G
AV



normal malpositions transposition


Ao
PA
VSD
RV
LV

VSD is (nearly) always cono-ventricular



Subarterial conuses are extremely variable

There are few more perplexing areas than the continuum of complete transposition of the great arteries, DORV and tetralogy of Fallot.

In the overall scheme of things, *the definition of congenital heart disease is less important than how well the malformation can be repaired.*

Robert Replogle 1985

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)

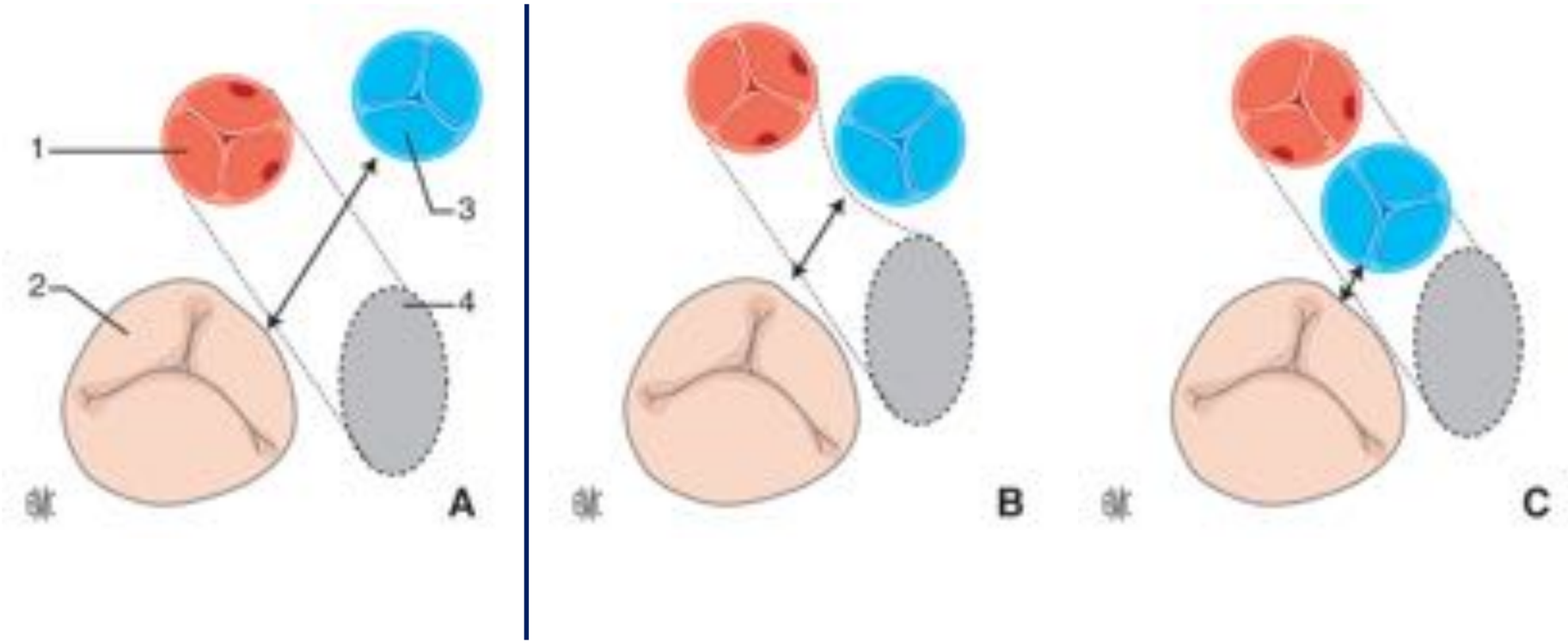
2. is "anatomic" repair feasible ?

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

IntraVentricular Repair (IVR)

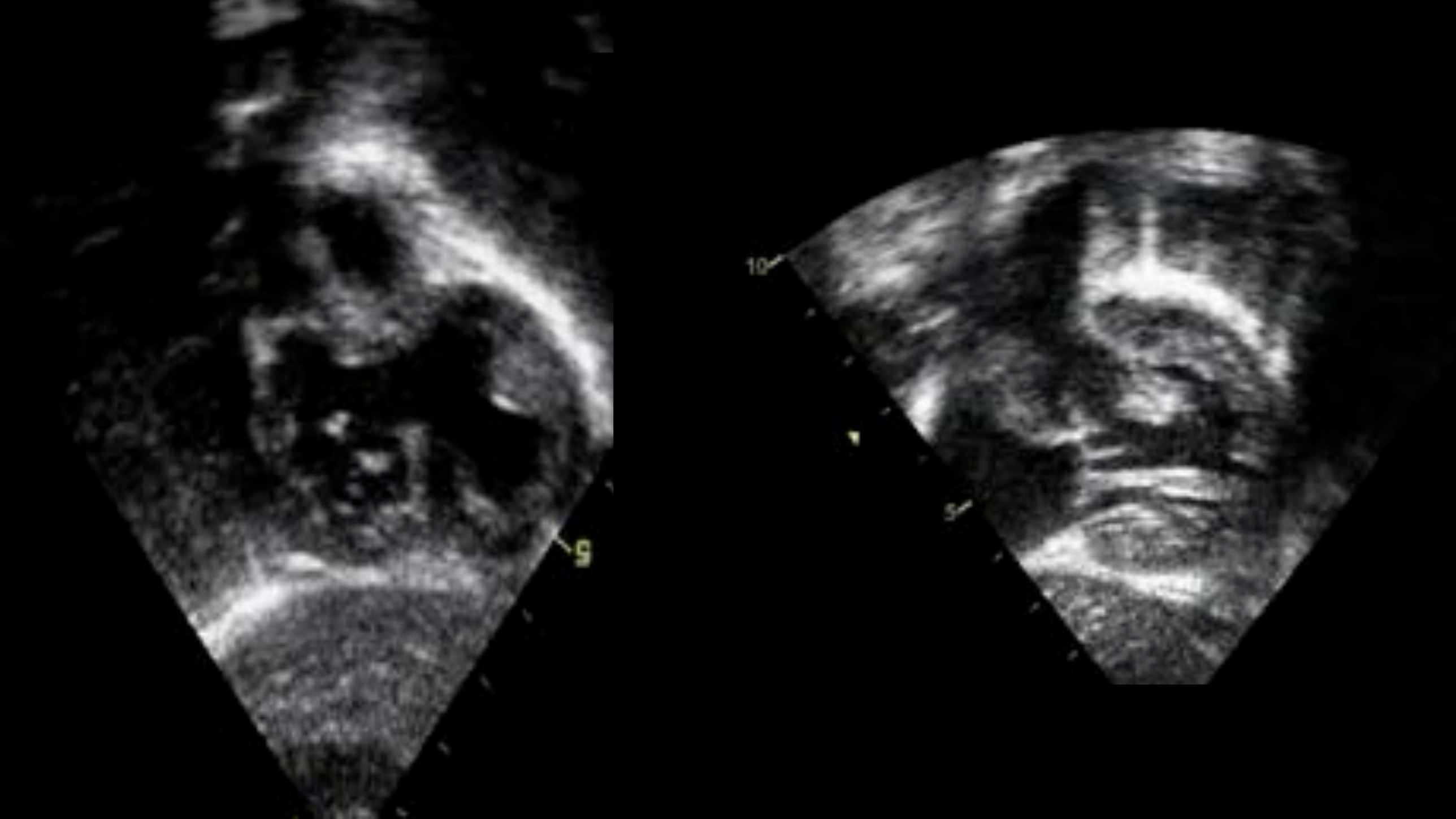
- VSD-type (no pulmonary stenosis)
- Fallot-type (pulmonary stenosis)

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



IVR possible

IVR impossible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10
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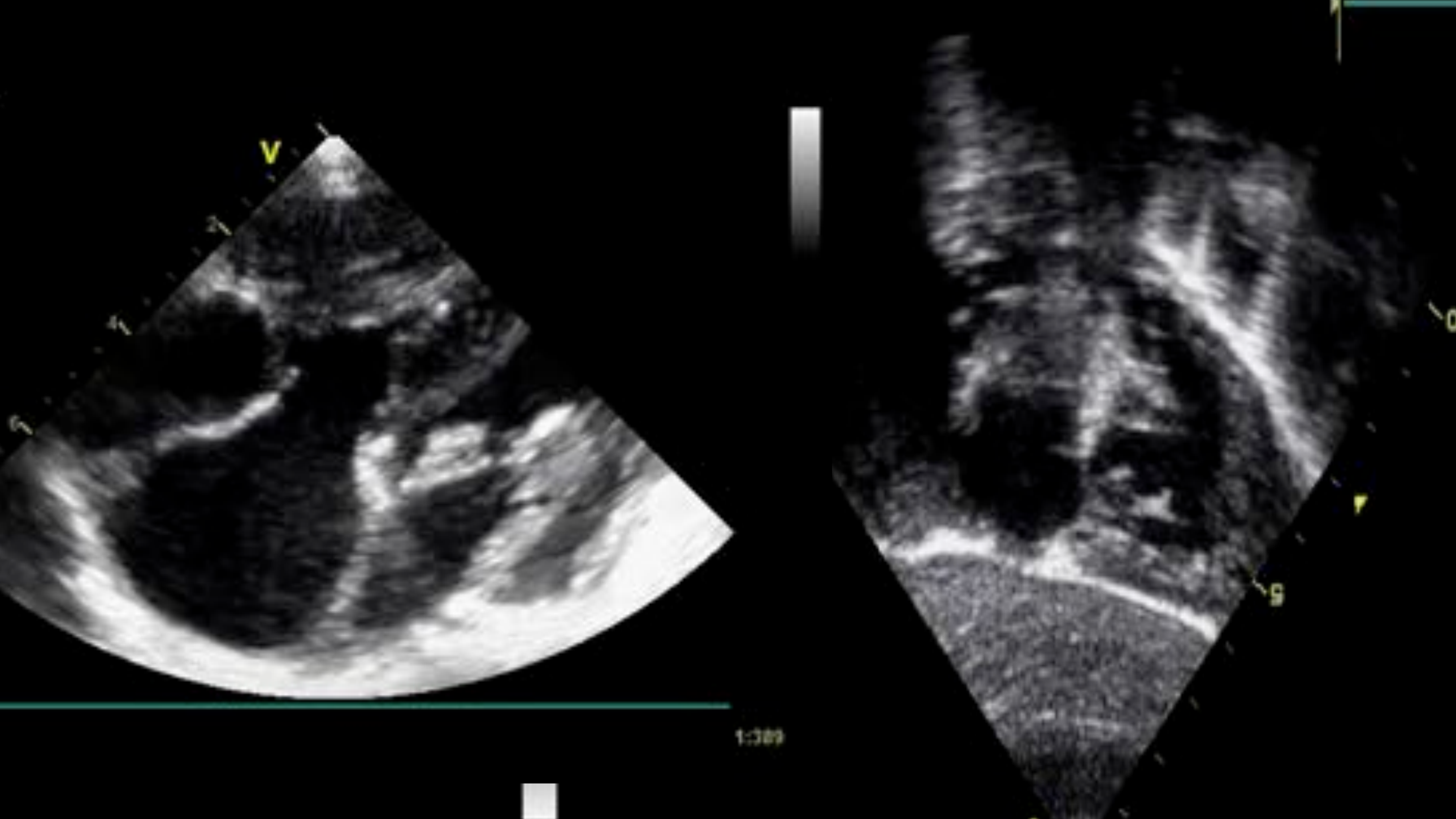
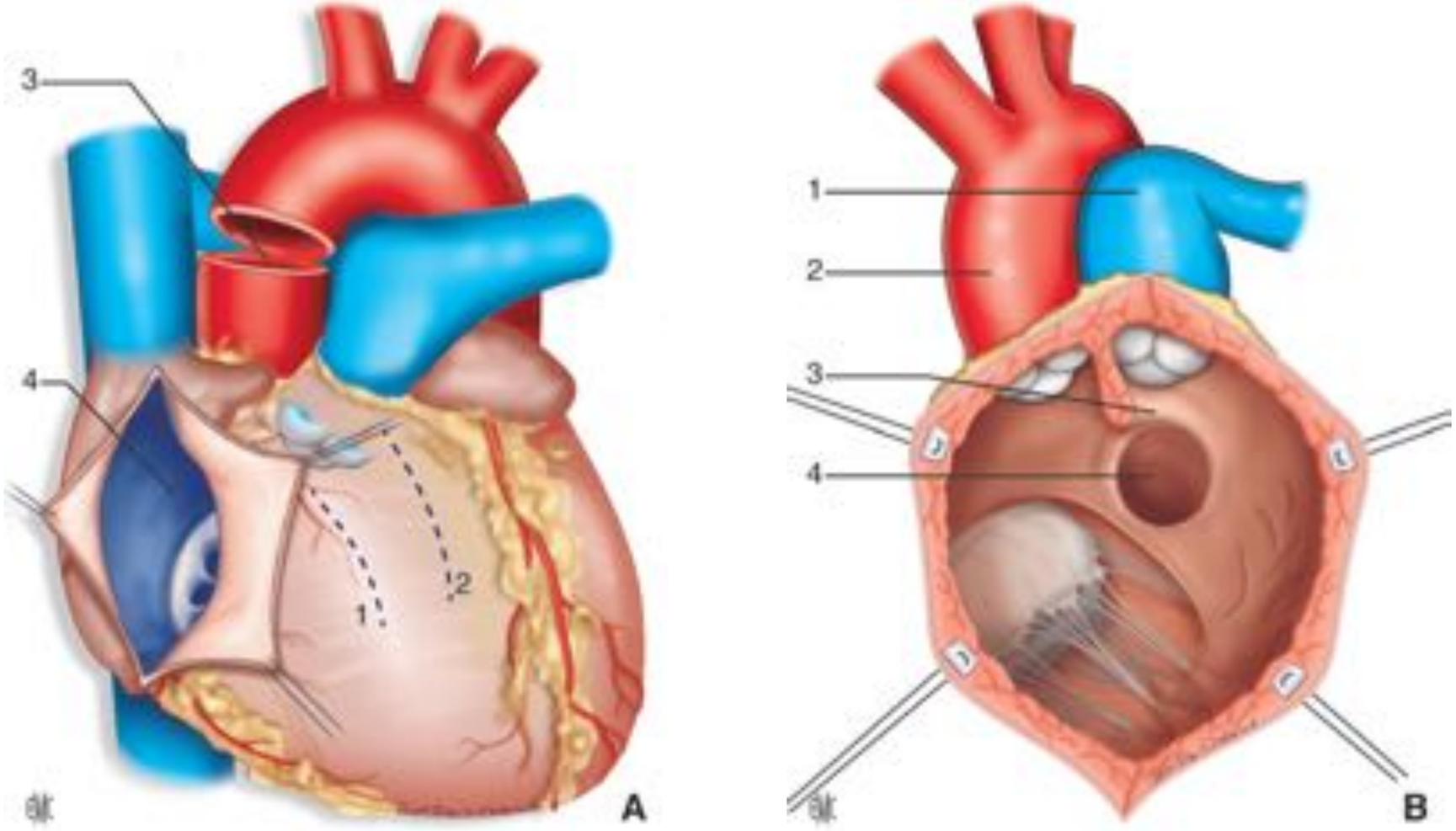
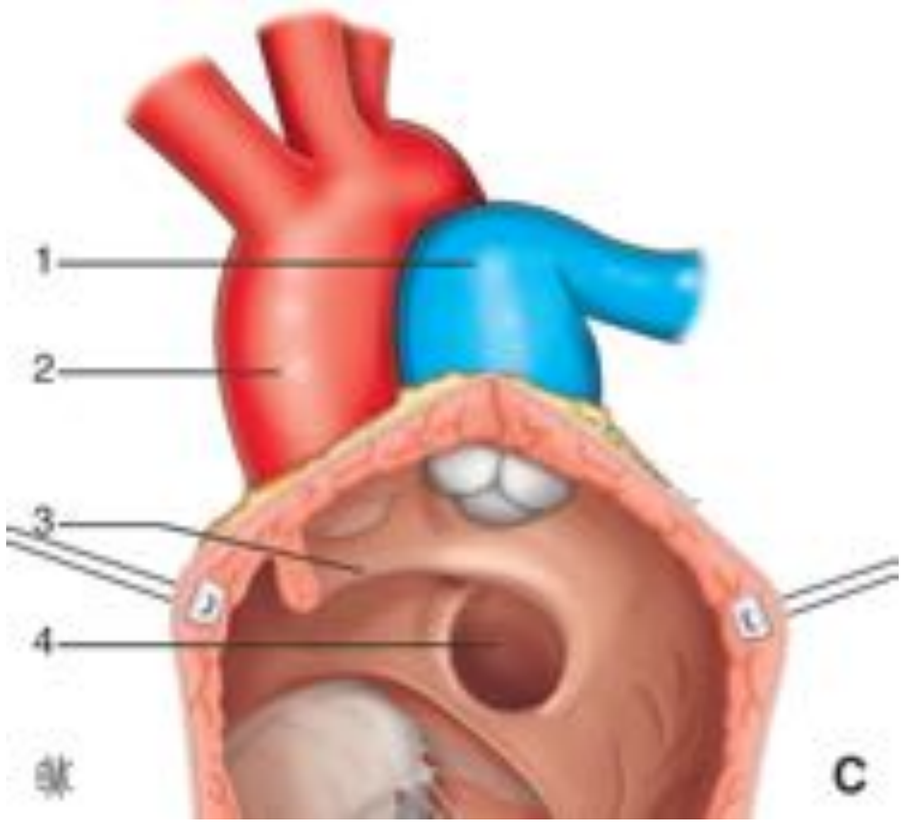
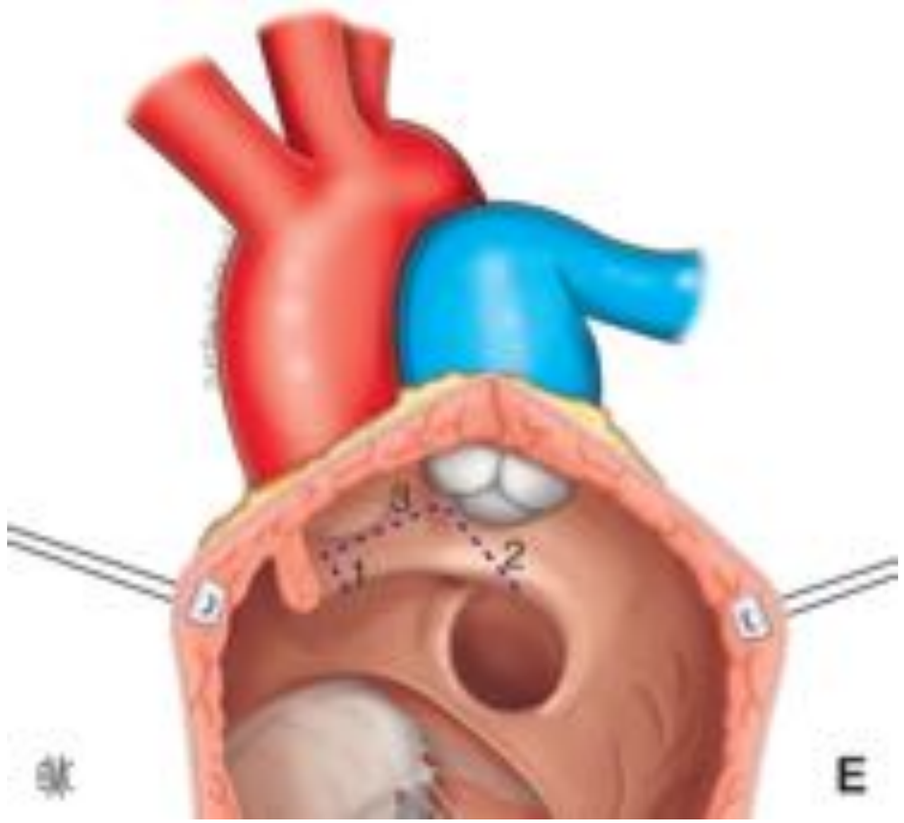

V
1:389

Intraventricular repair

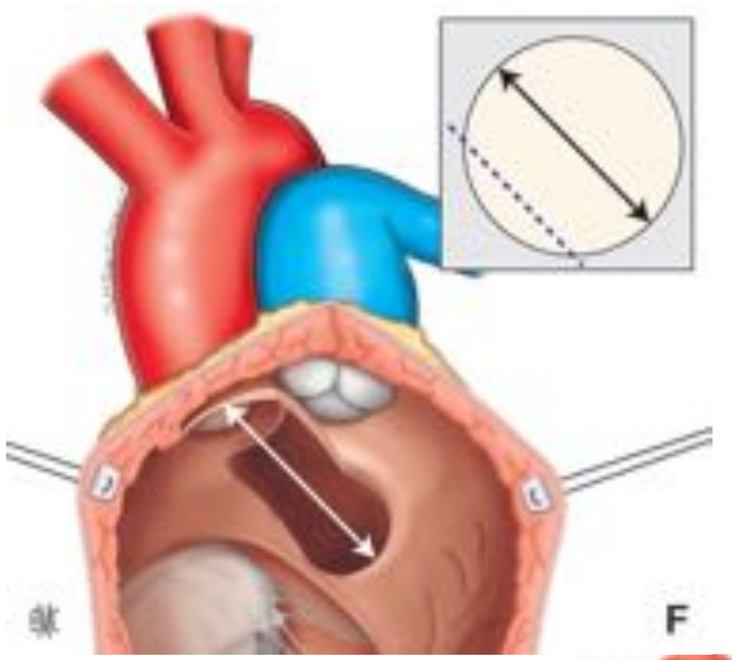


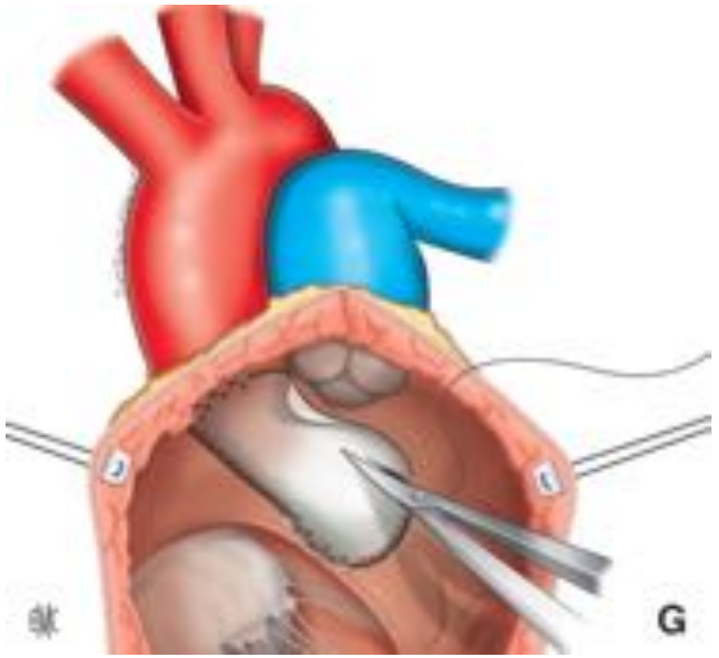
Intraventricular repair

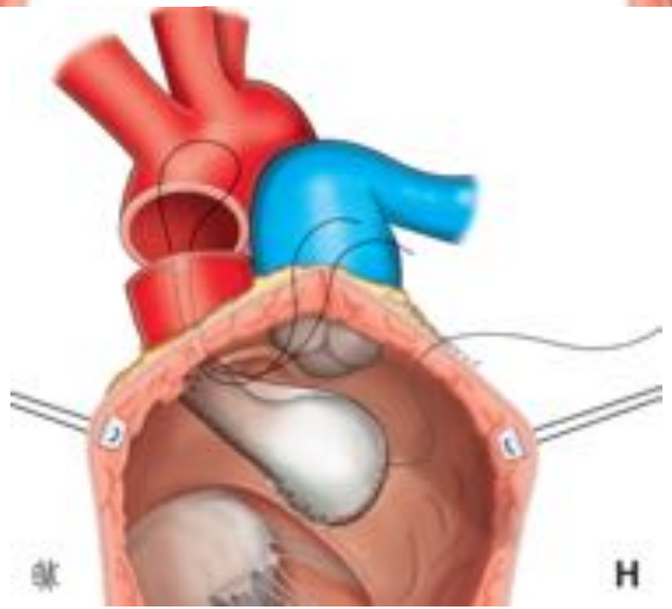




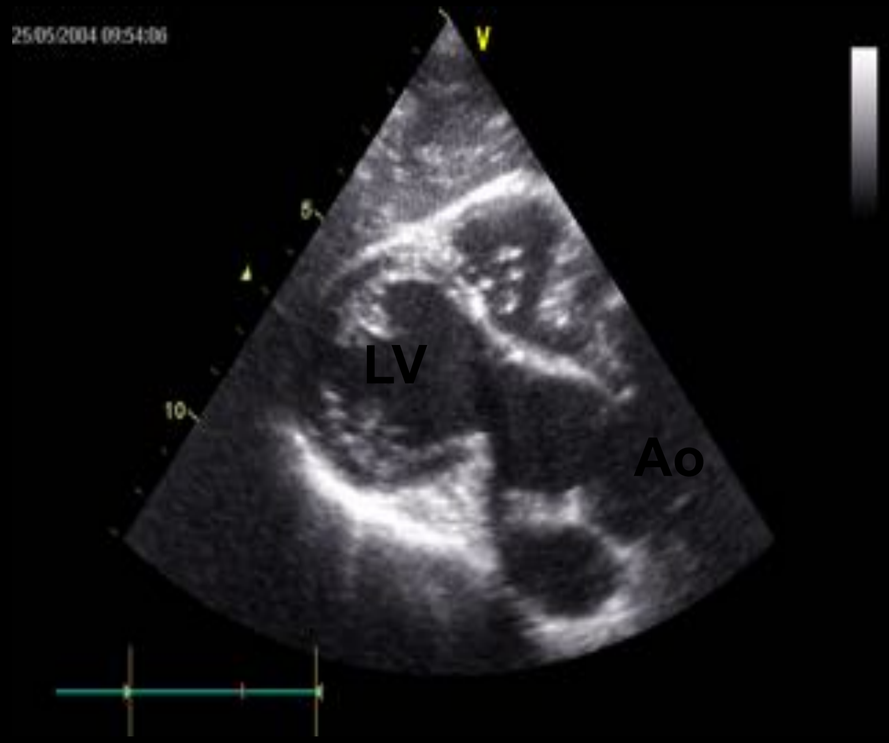
Intraventricular repair

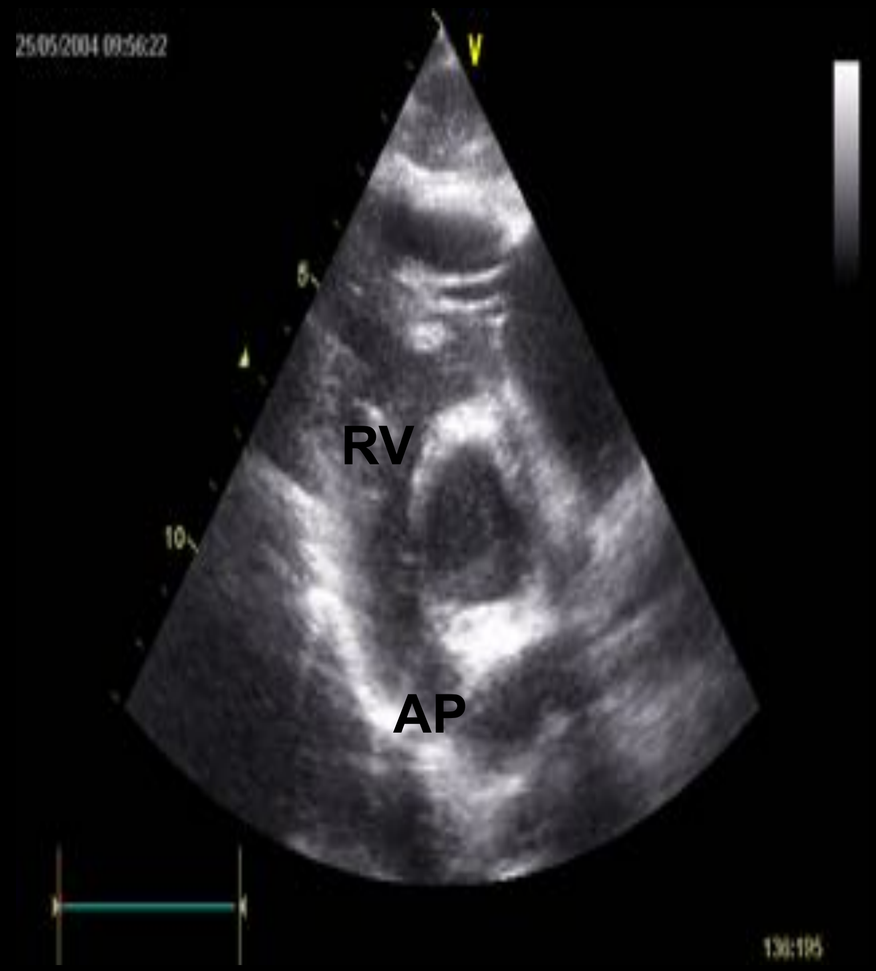






Intraventricular repair






25/05/2004 09:55:57
V
5
10
259:318


136:195
25/05/2004 09:56:22

2. is "anatomic" repair feasible ?

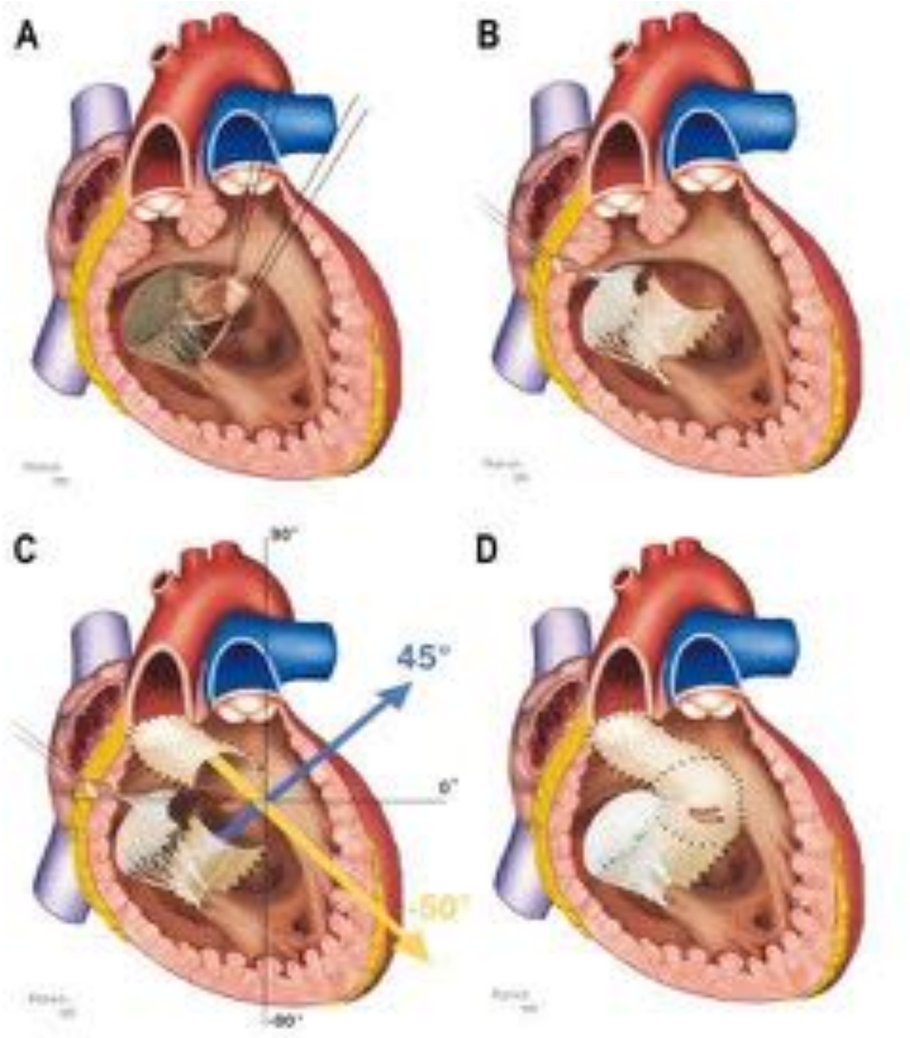
IVR may be possible but difficult :

. extensive abnormal insertions of tricuspid and/or mitral valve

. asymmetric subpulmonary conus

extra-anatomic repair may be preferable

Extensive insertions of tricuspid valve on conal septum



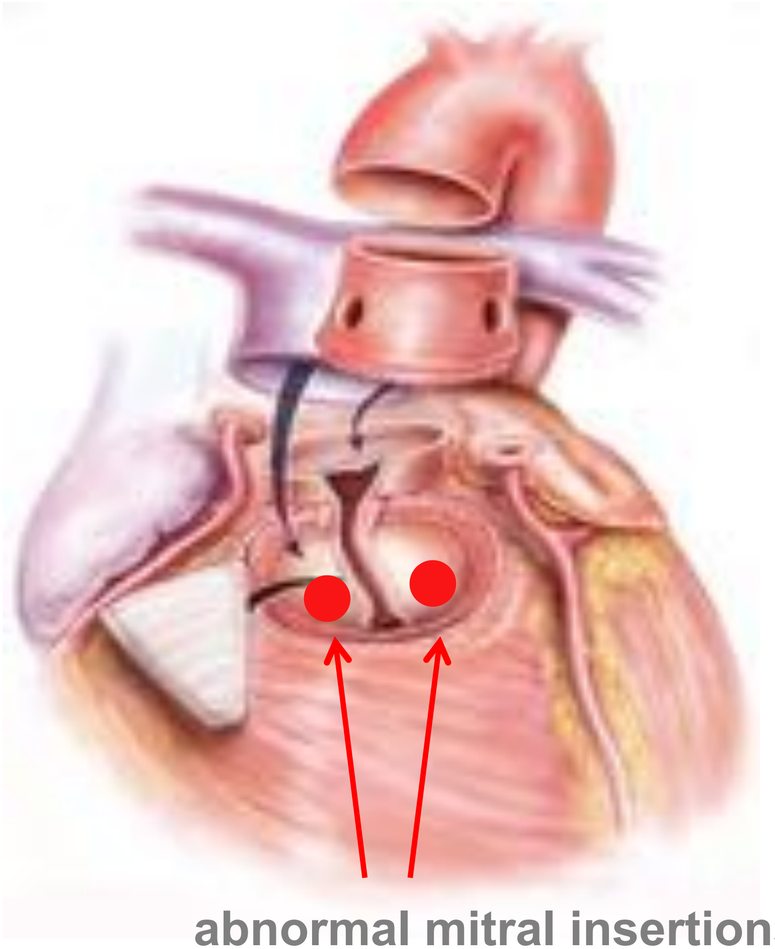
. LV-aorta tunnel is difficult

. extra-anatomic repair is preferable

- Bex-Nikaidoh
- cono-truncal rotation


5
10
15

Abnormal insertions of mitral valve on conal septum



abnormal mitral insertions

. rare

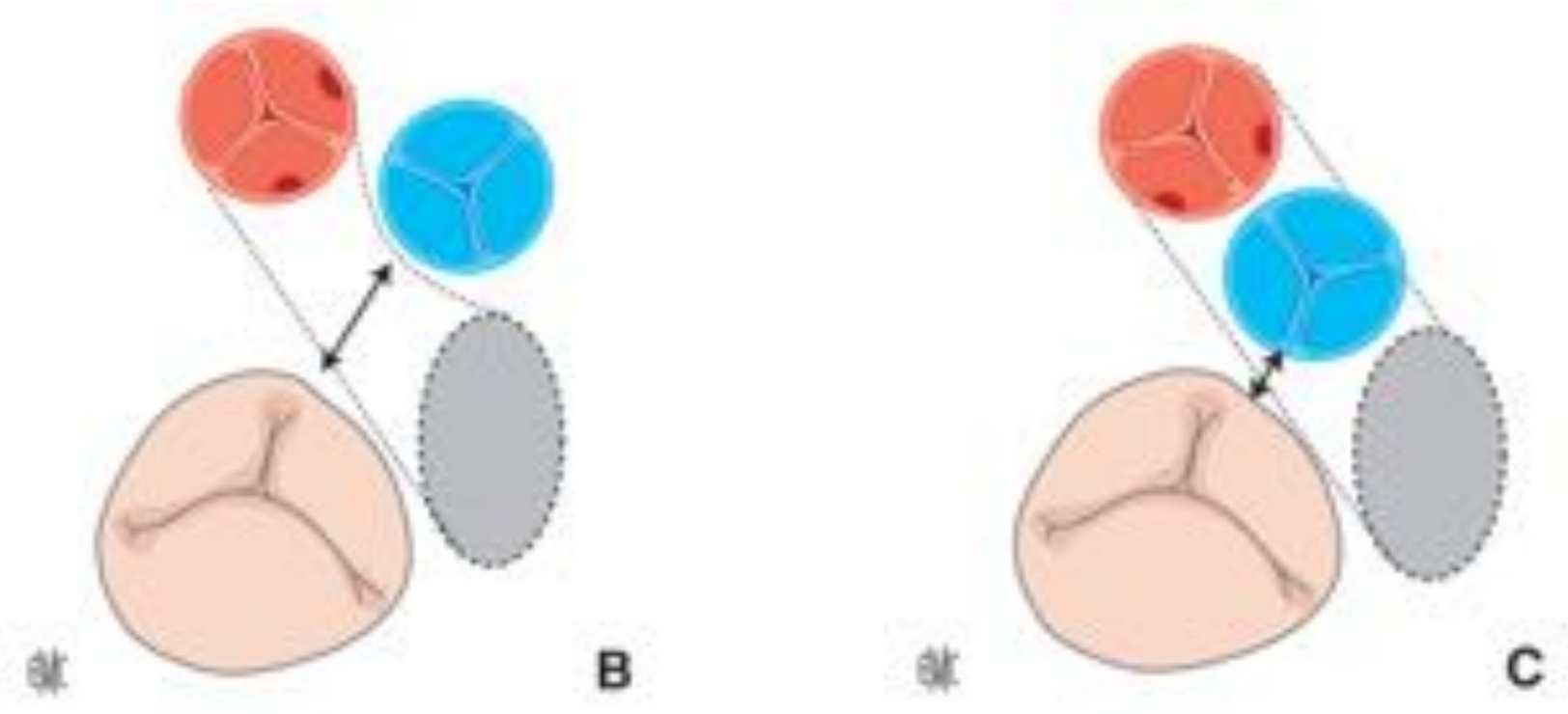
. difficult to manage

. resection of conal septum impossible

. Bex-Nikaidoh or cono-truncal rotation may be options in selected cases


V
V
1:204

Tricuspid-to-pulmonary distance < Ao diameter



IVR impossible

3. which extra-anatomic repair is indicated ?

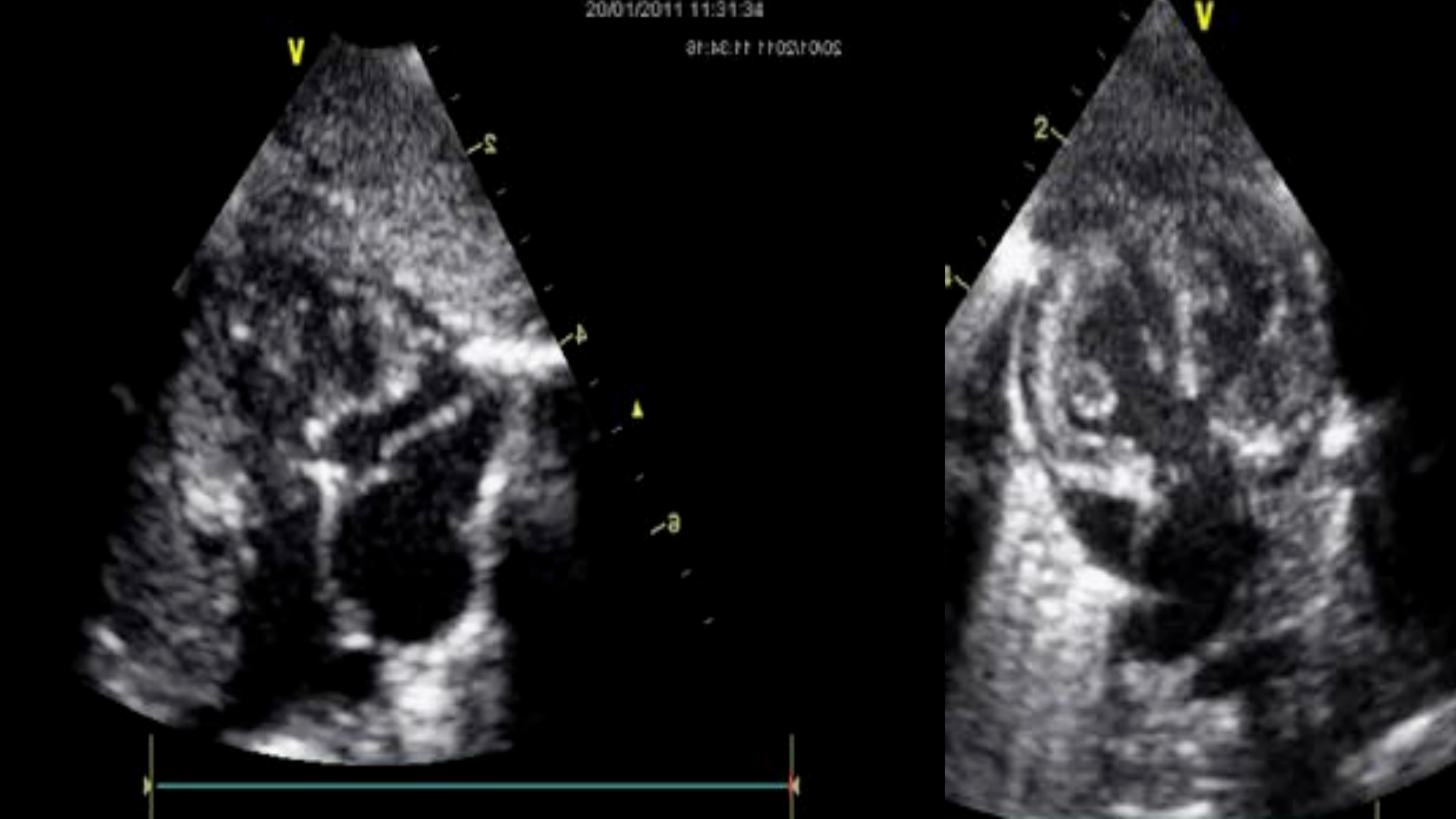
- when "anatomic" repair (IVR) is impossible

- determinant: pulmonary outflow tract
  (particularly pulmonary valve)
  - normal
  - very abnormal (stenotic)
  - midly 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**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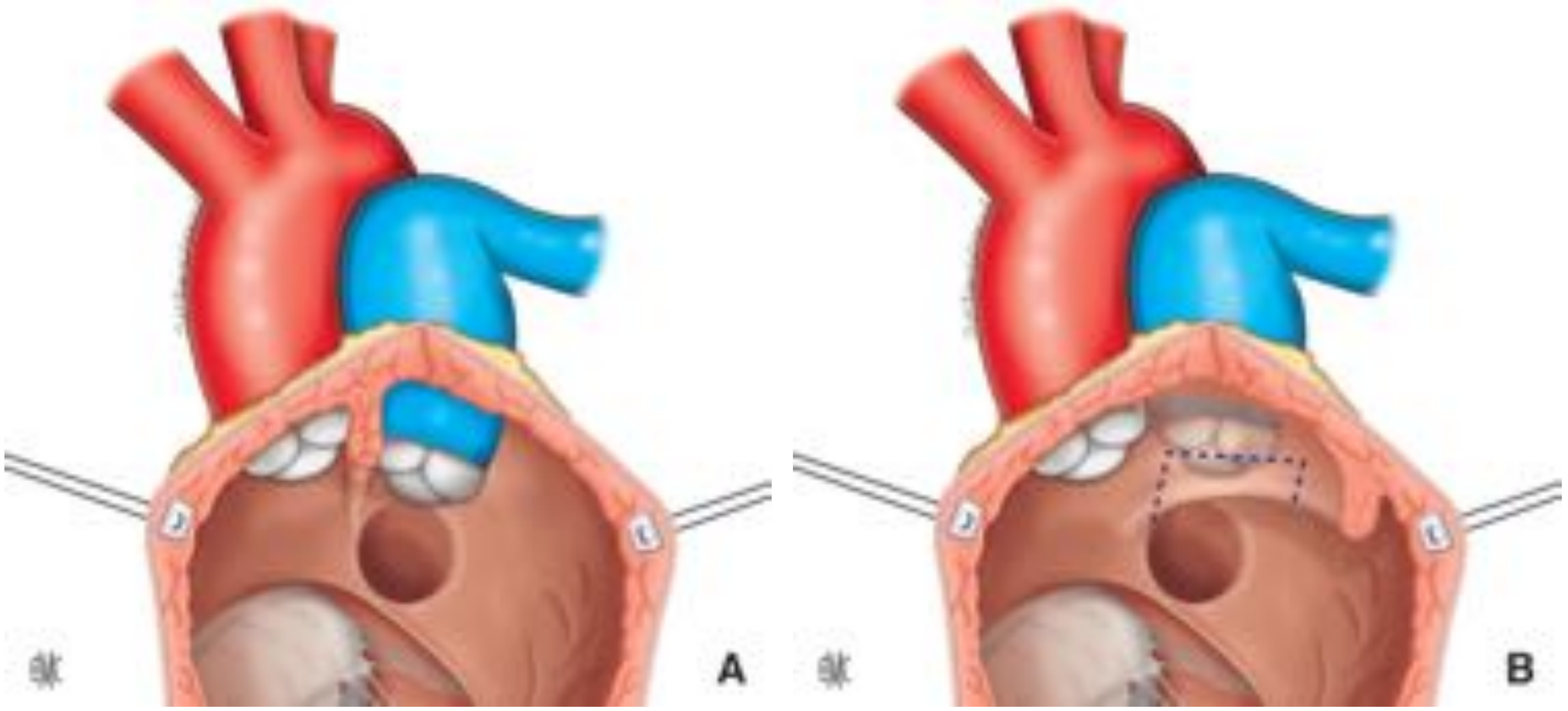
V
2
4
6
V
2

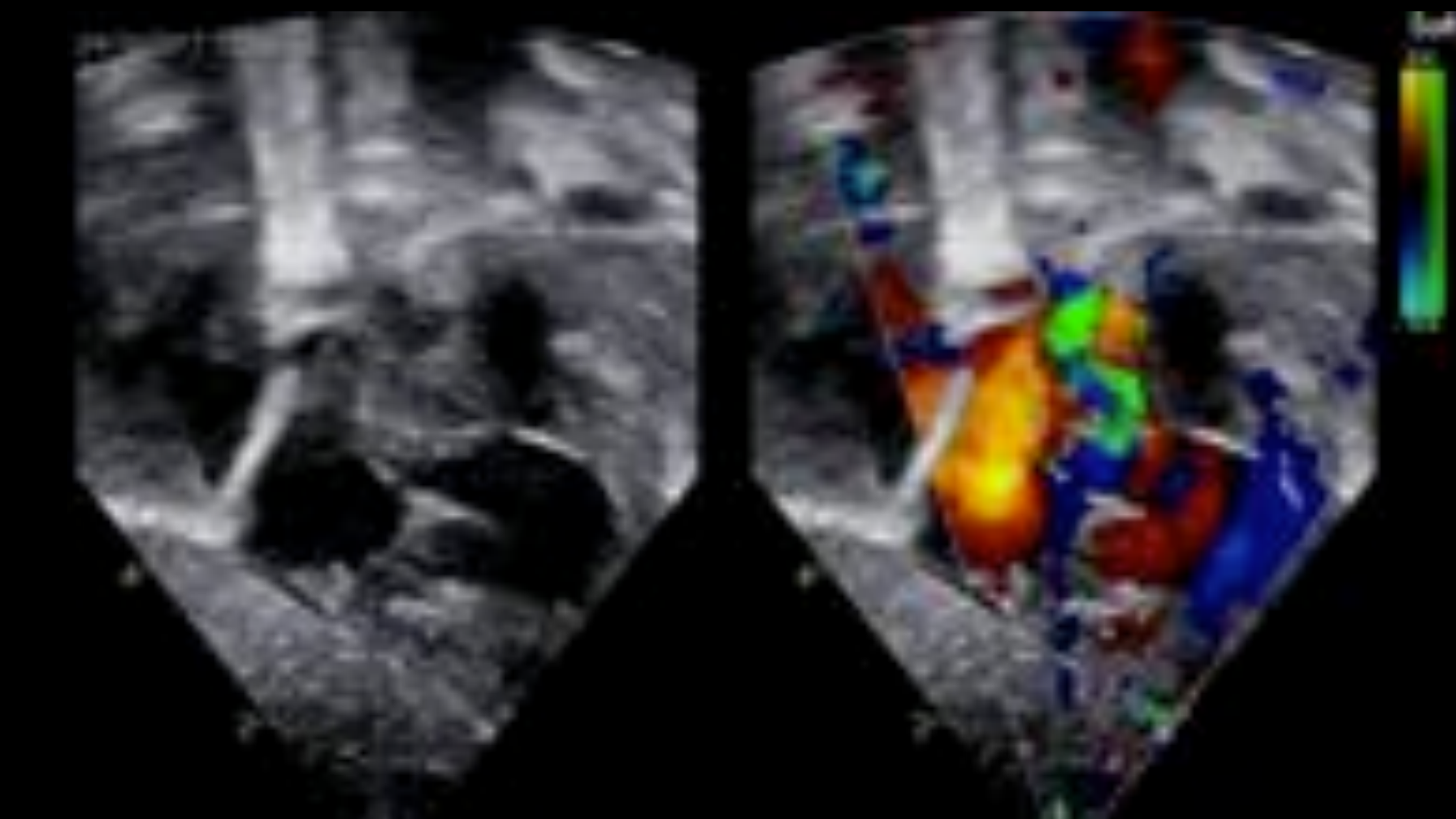
LV-PA connection + arterial switch



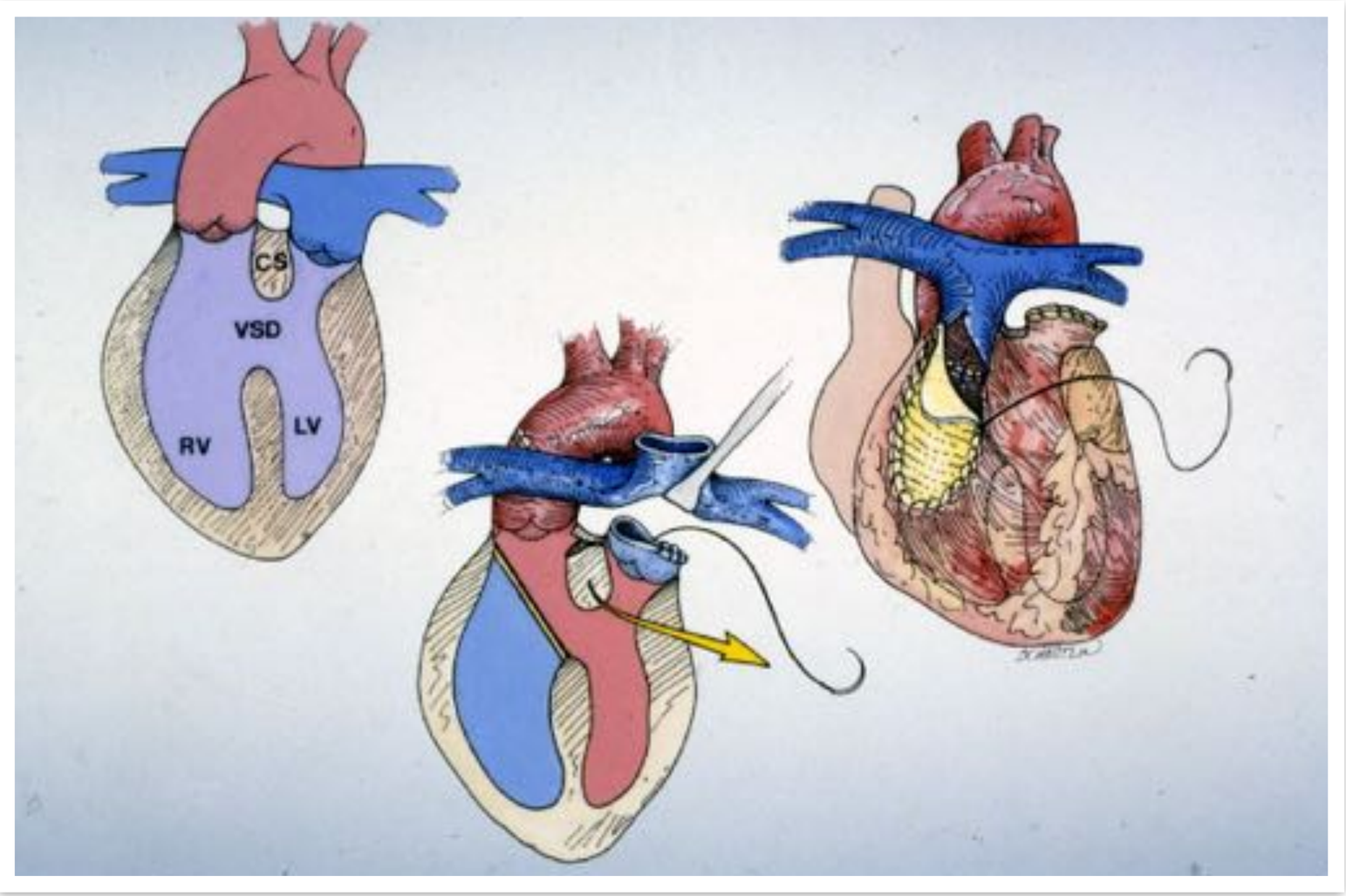
3. which extra-anatomic repair is indicated ?

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

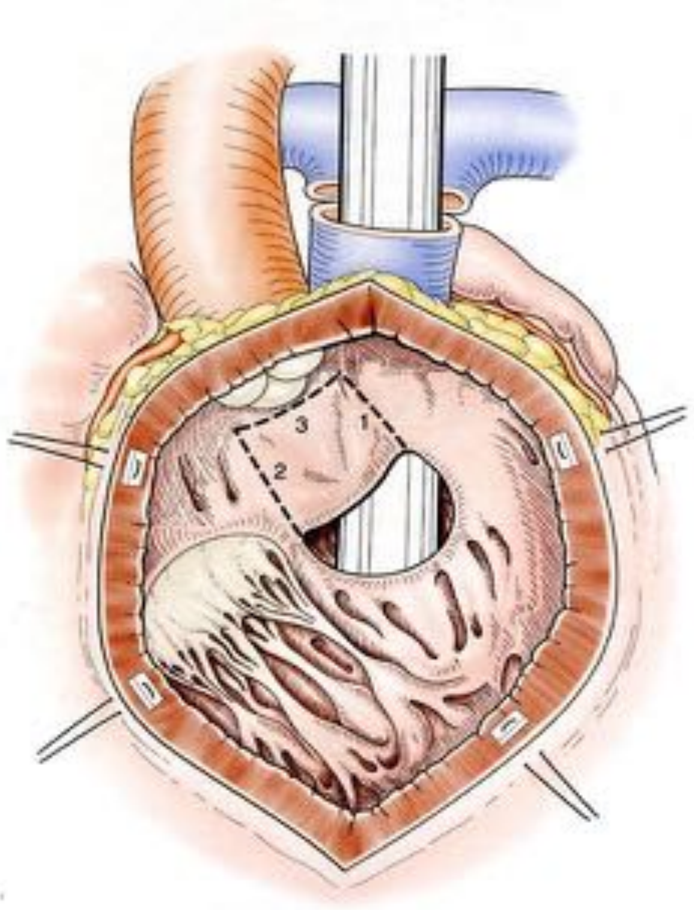
- **REV operation**
- **Bex-Nikaidoh operation**
- **Rastelli operation**



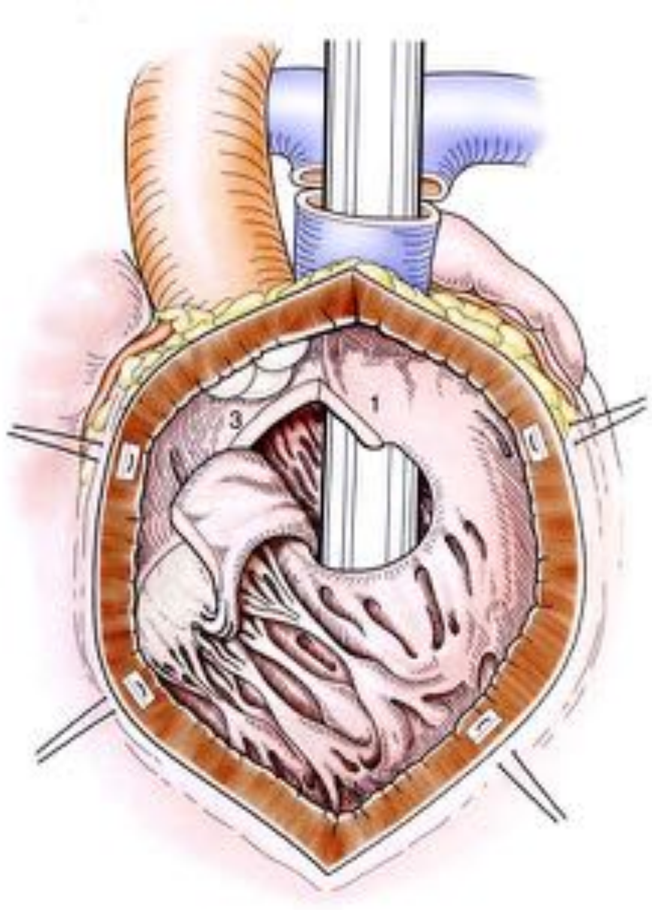
REV operation
(Réparation à l'Etage Ventriculaire)



Extra-anatomic repair : REV

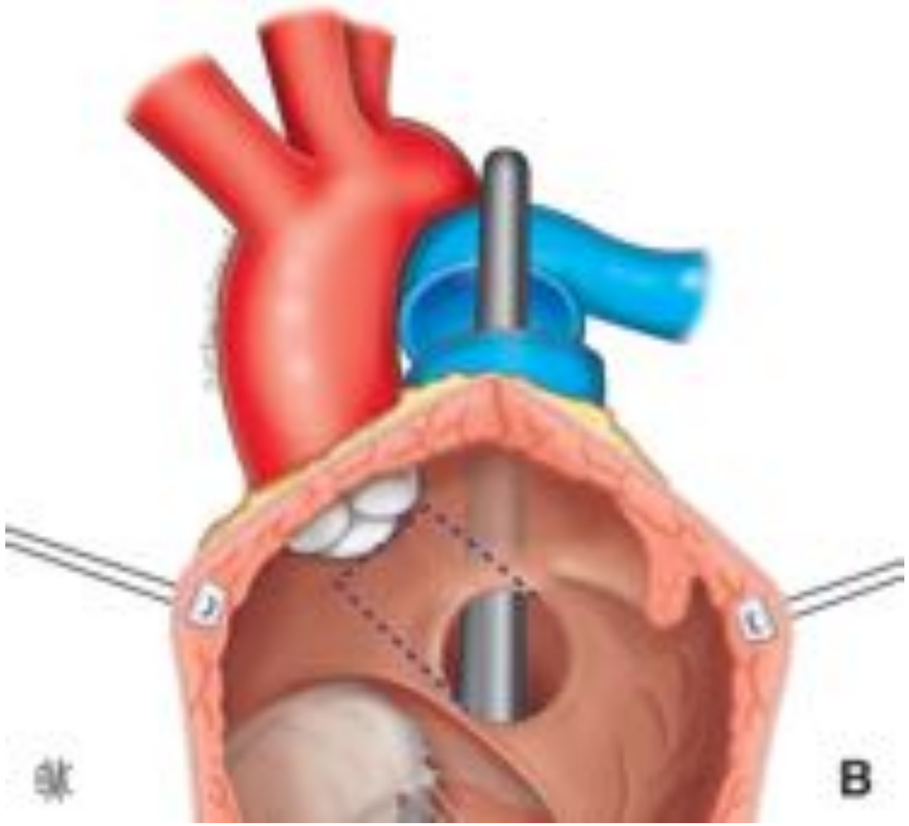


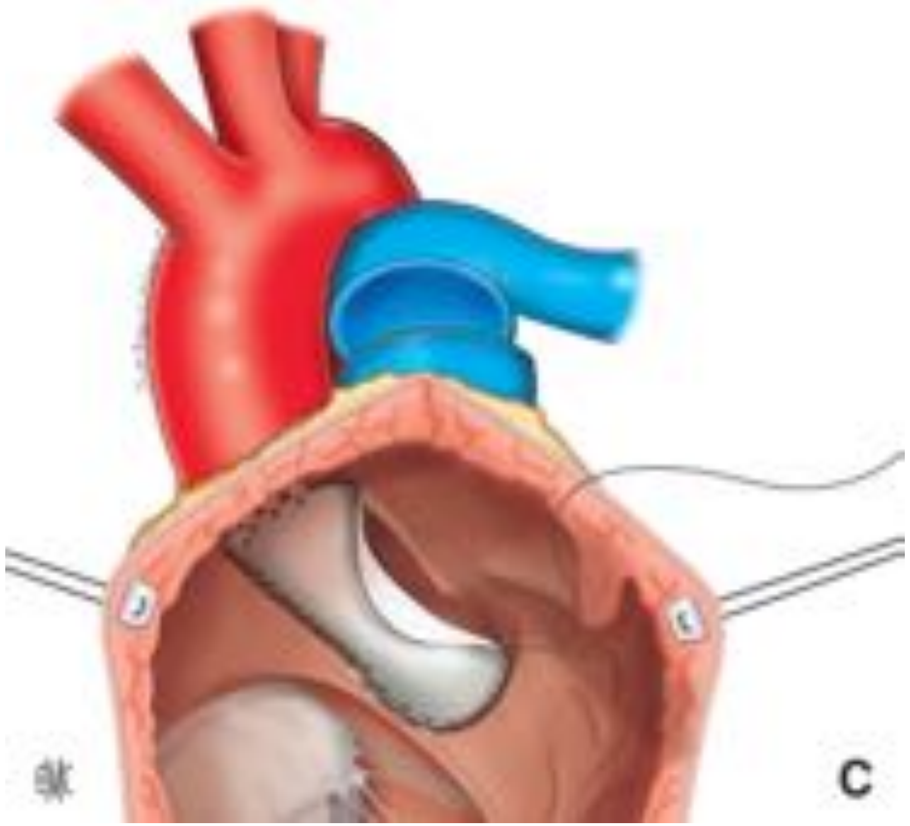
conal septum excision



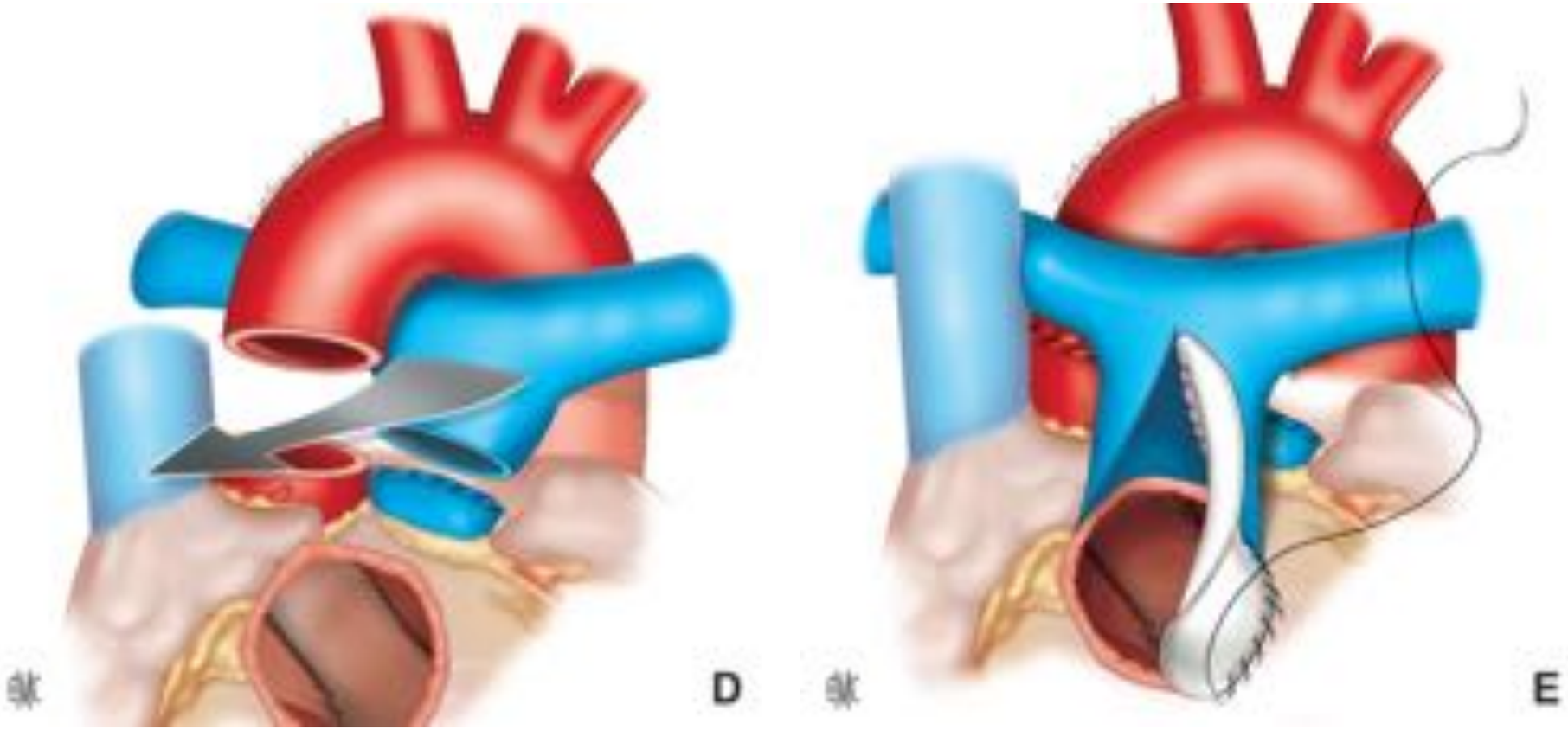
conal septum mobilization

Extra-anatomic repair : REV

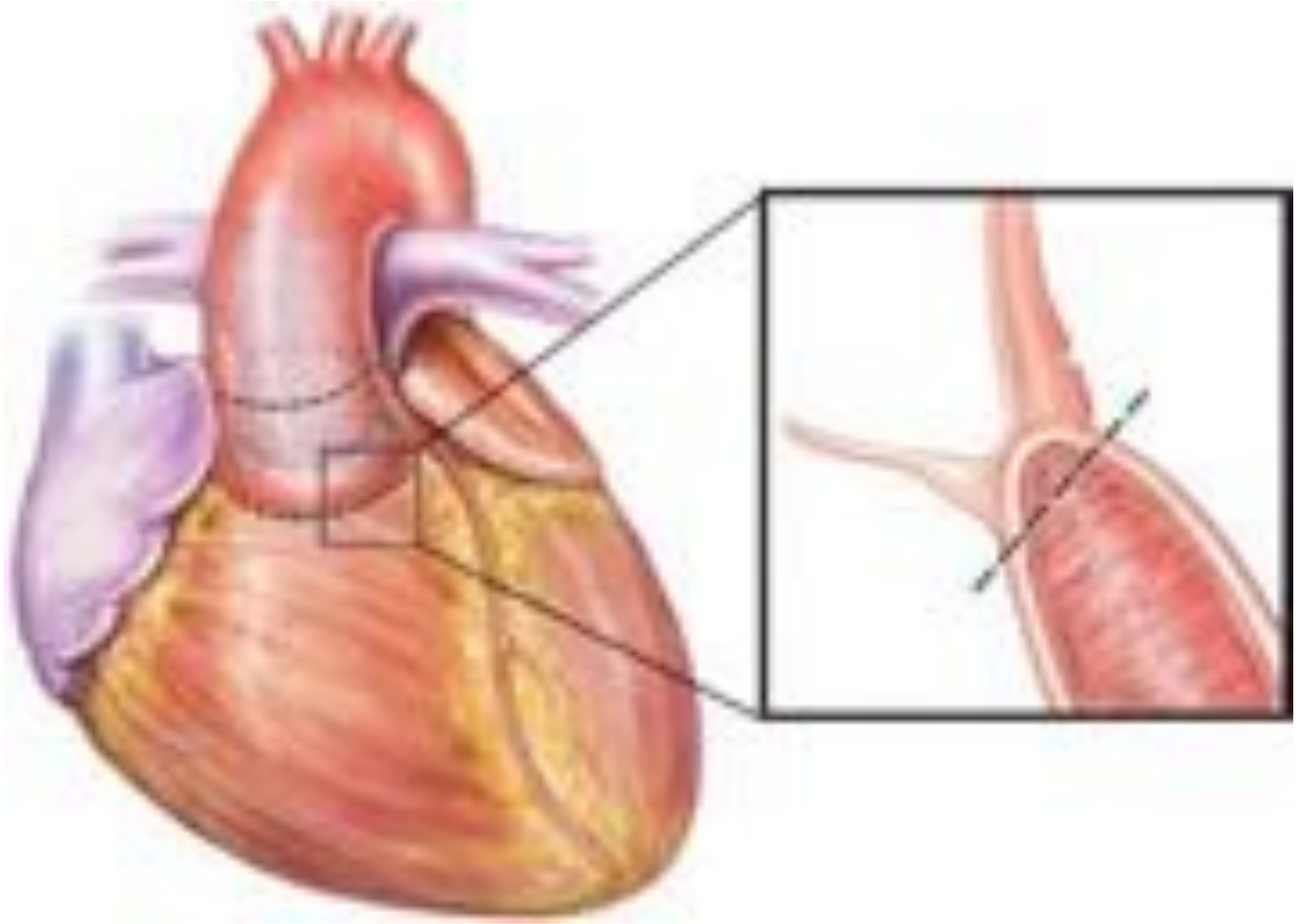




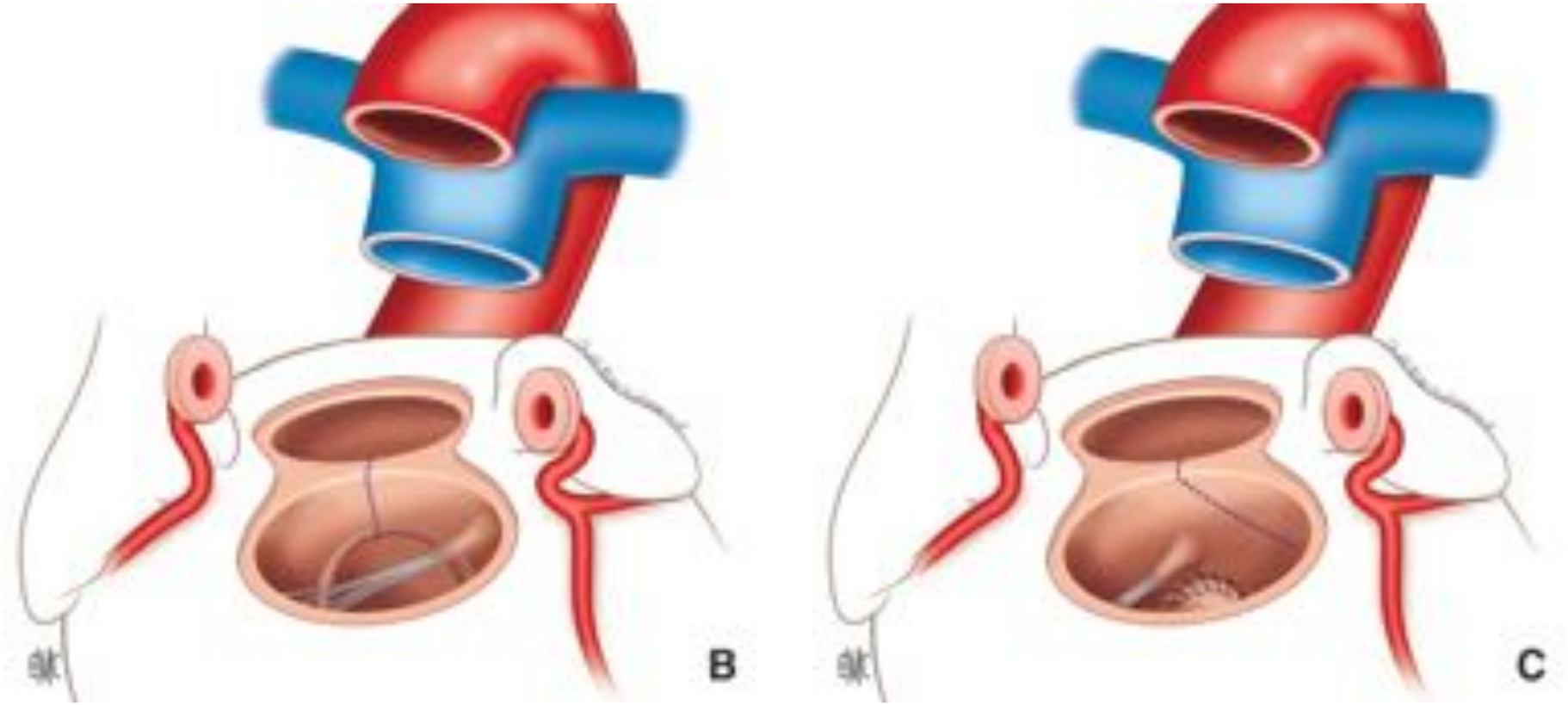
Extra-anatomic repair : REV



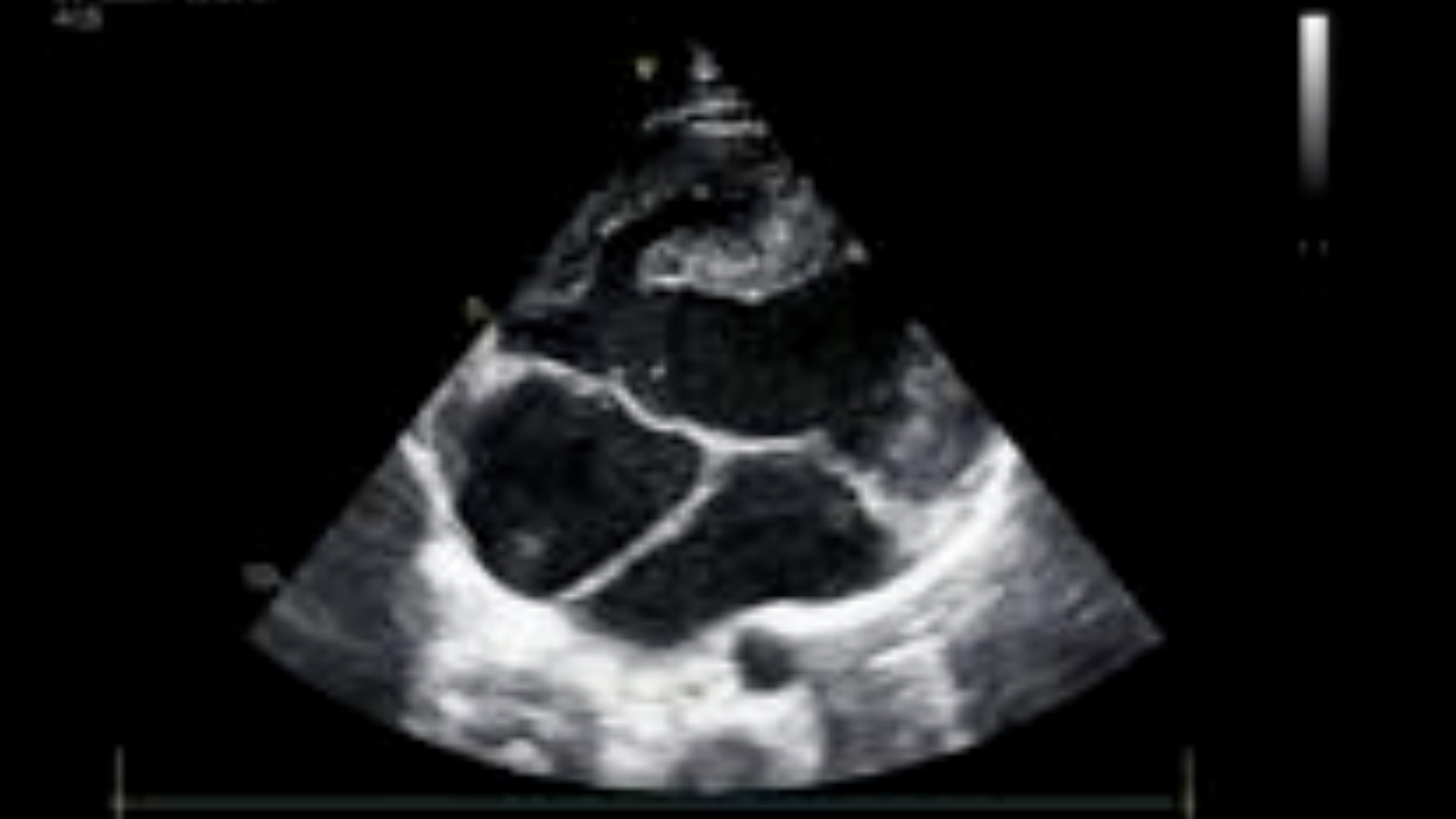
Extra-anatomic repair
Bex-Nikaidoh operation



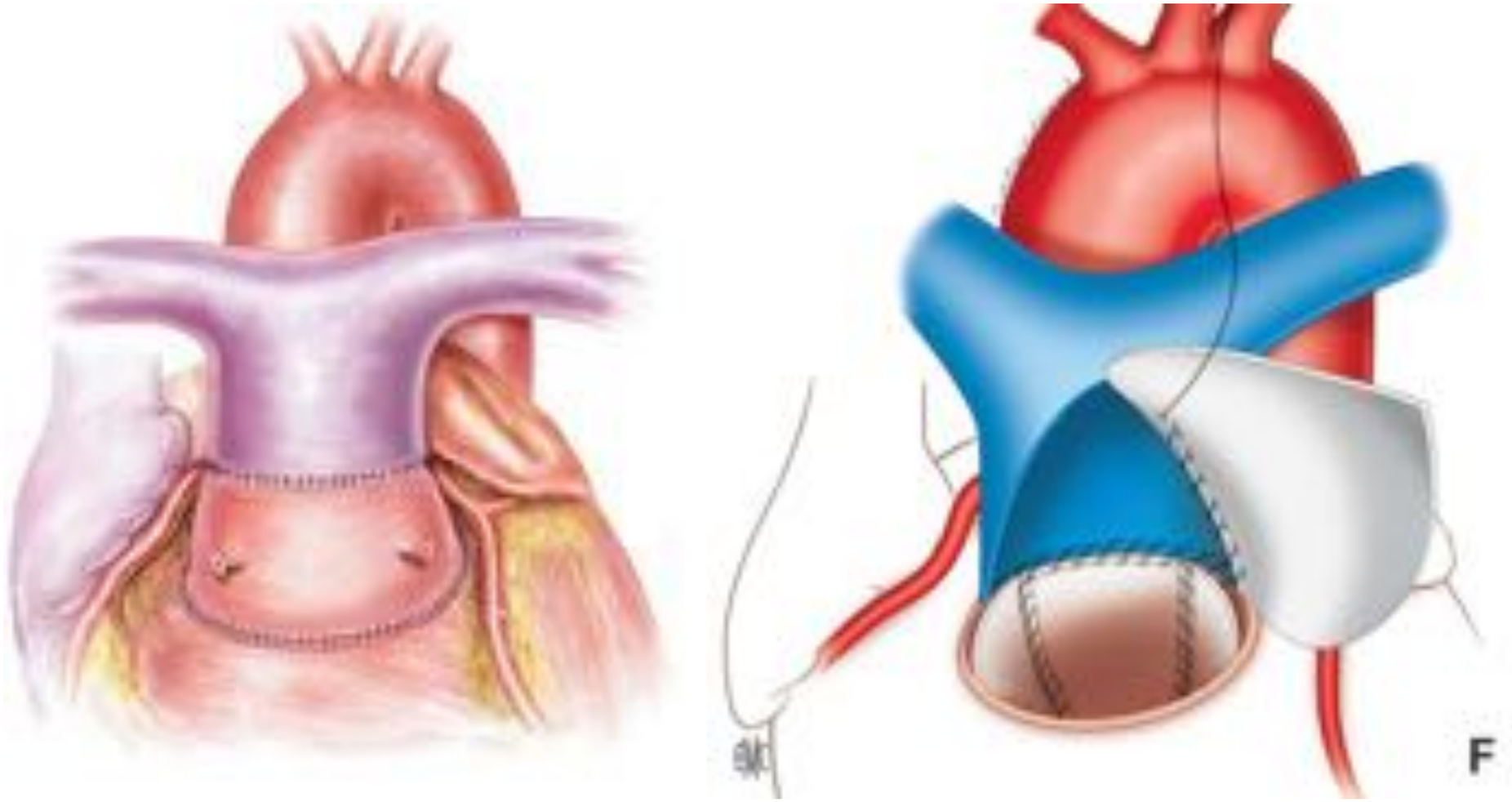
Extra-anatomic repair
Bex-Nikaidoh operation

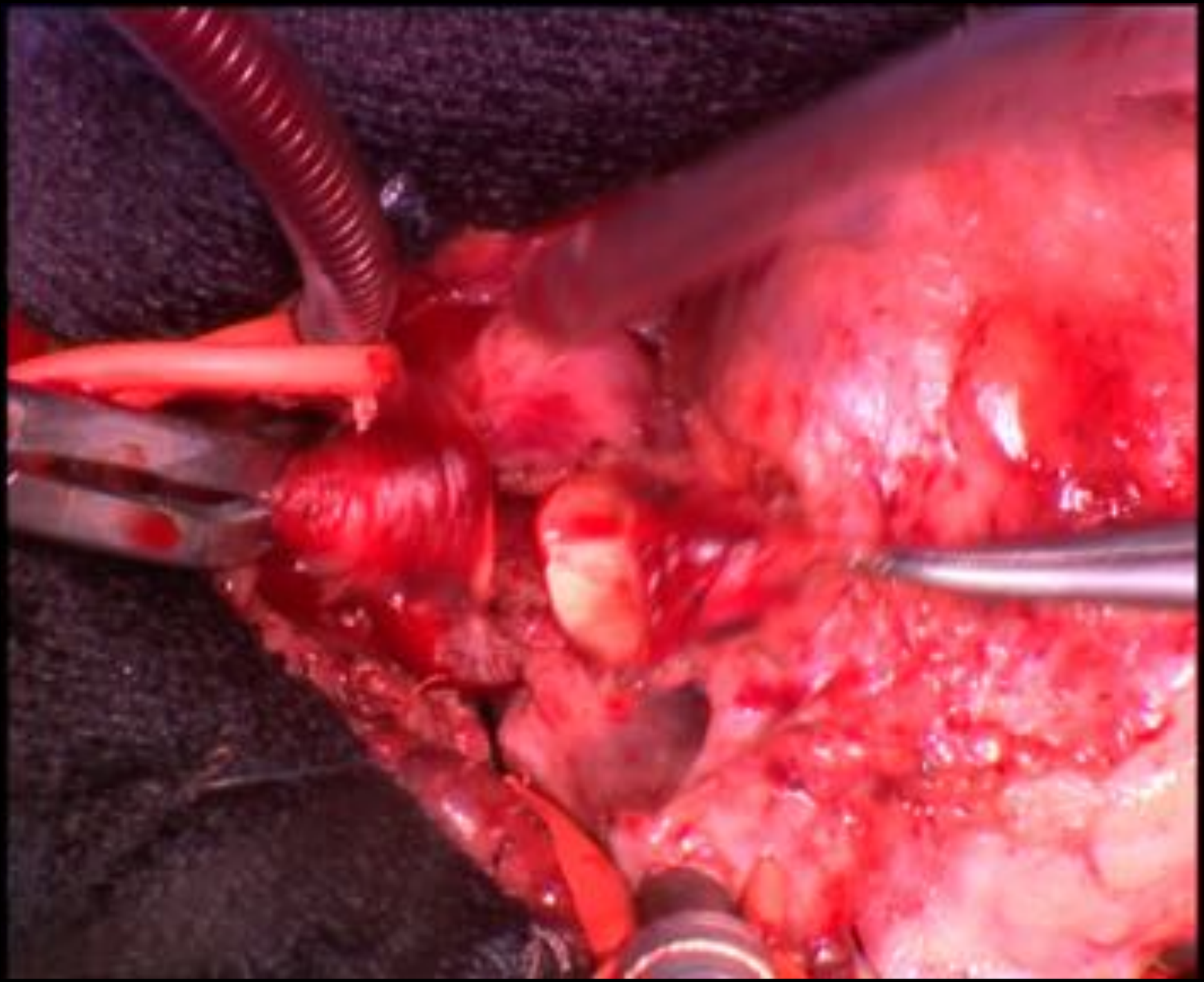


Extra-anatomic repair
Bex-Nikaidoh operation



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)

Indications for Rastelli operation

- inadequate pulmonary arteries (hypoplasia, stenosis)
- increased pulmonary vascular resistances

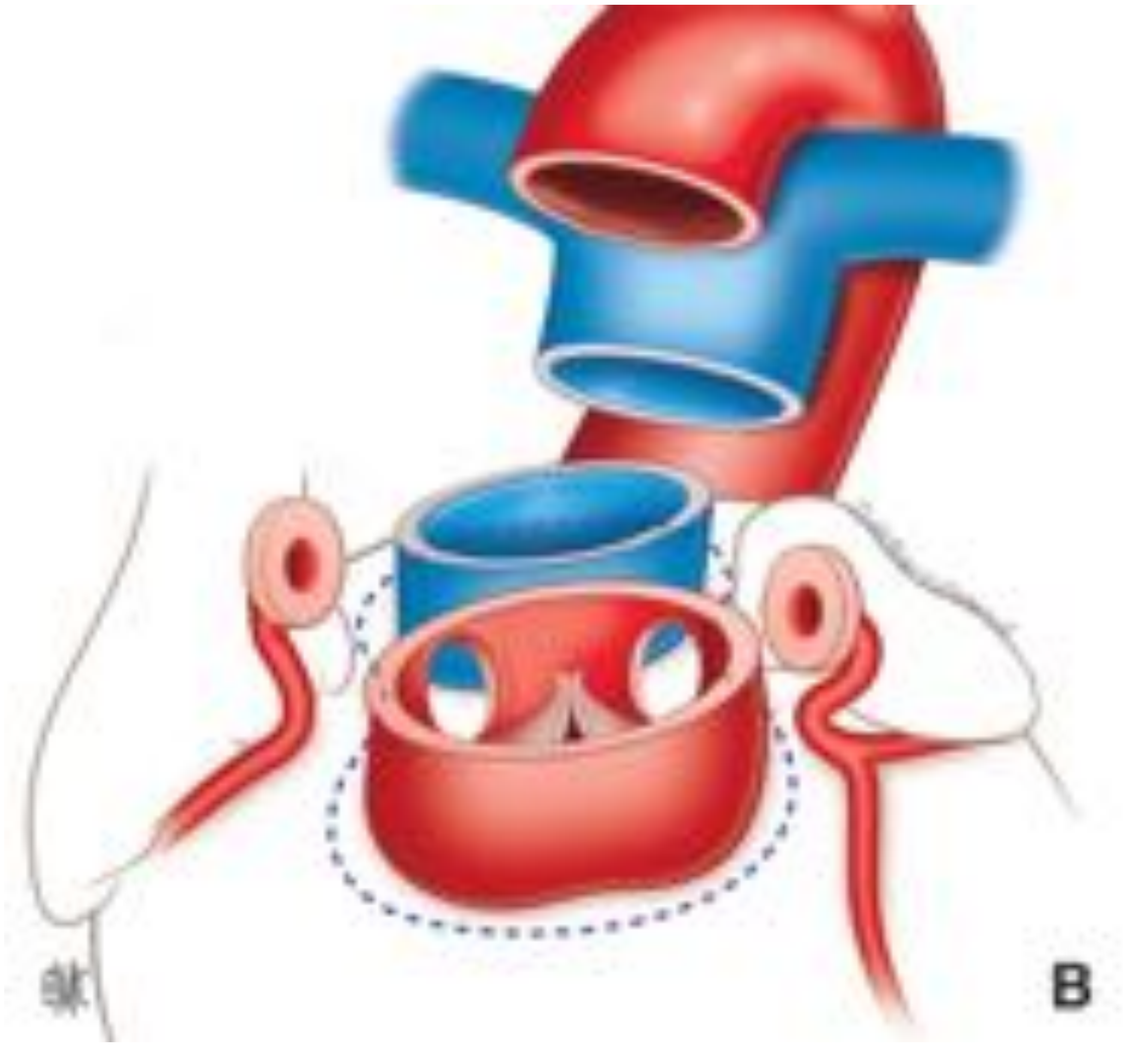
- very rare

3. which extra-anatomic repair is indicated ?

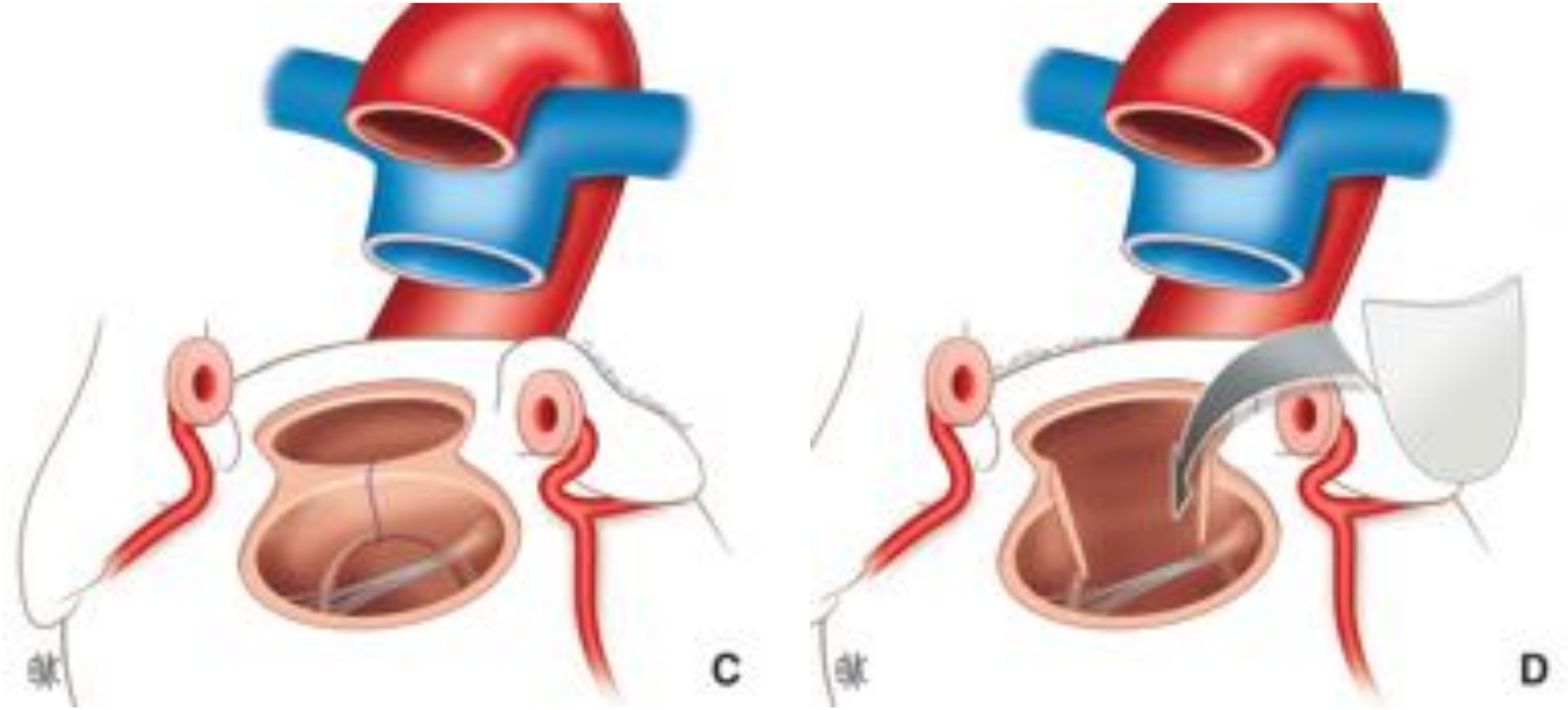
- when LVOT cannot be used as neoaortic
  but can be used as pulmonary
  - bicuspid pulmonary valve
  - midly-dysplastic pulmonary valve

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

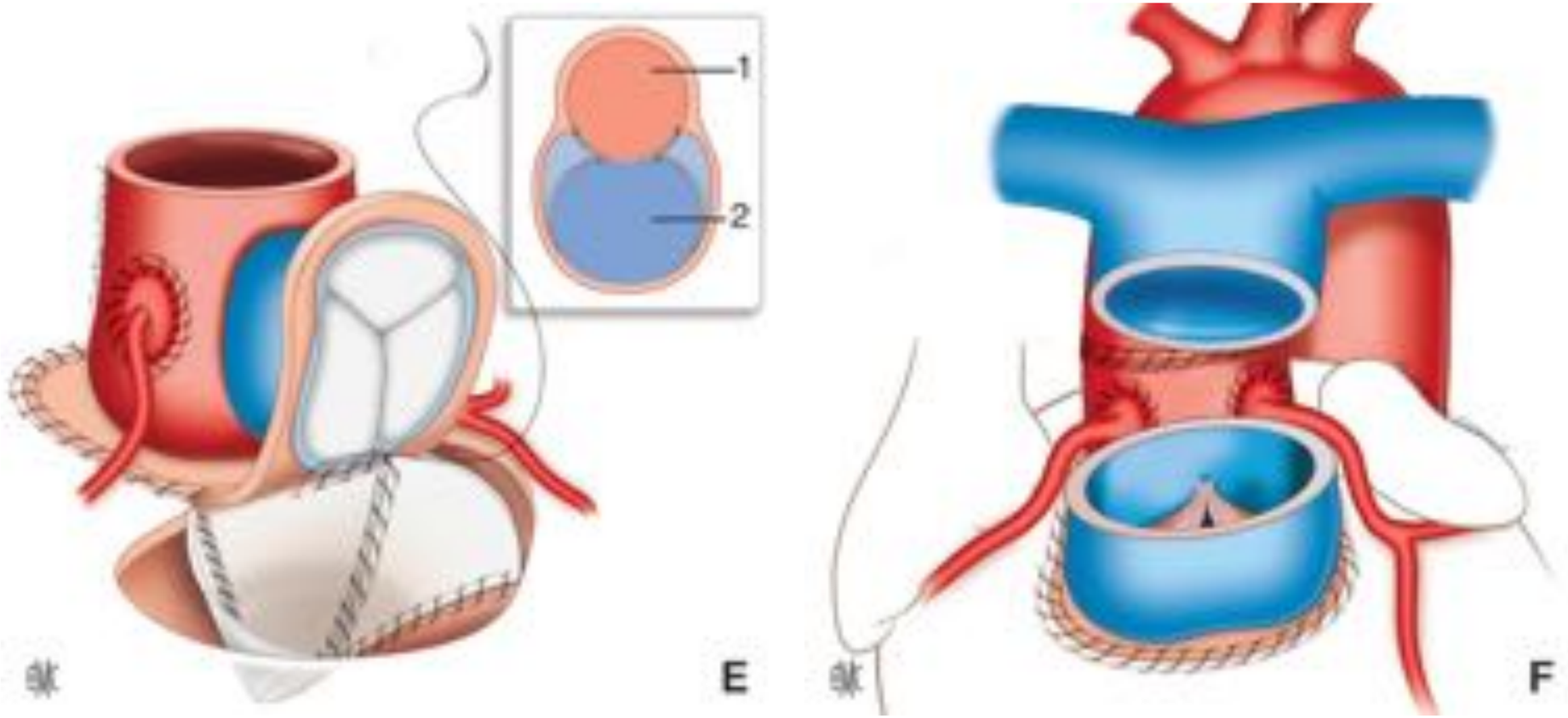
Conotruncal rotation procedure

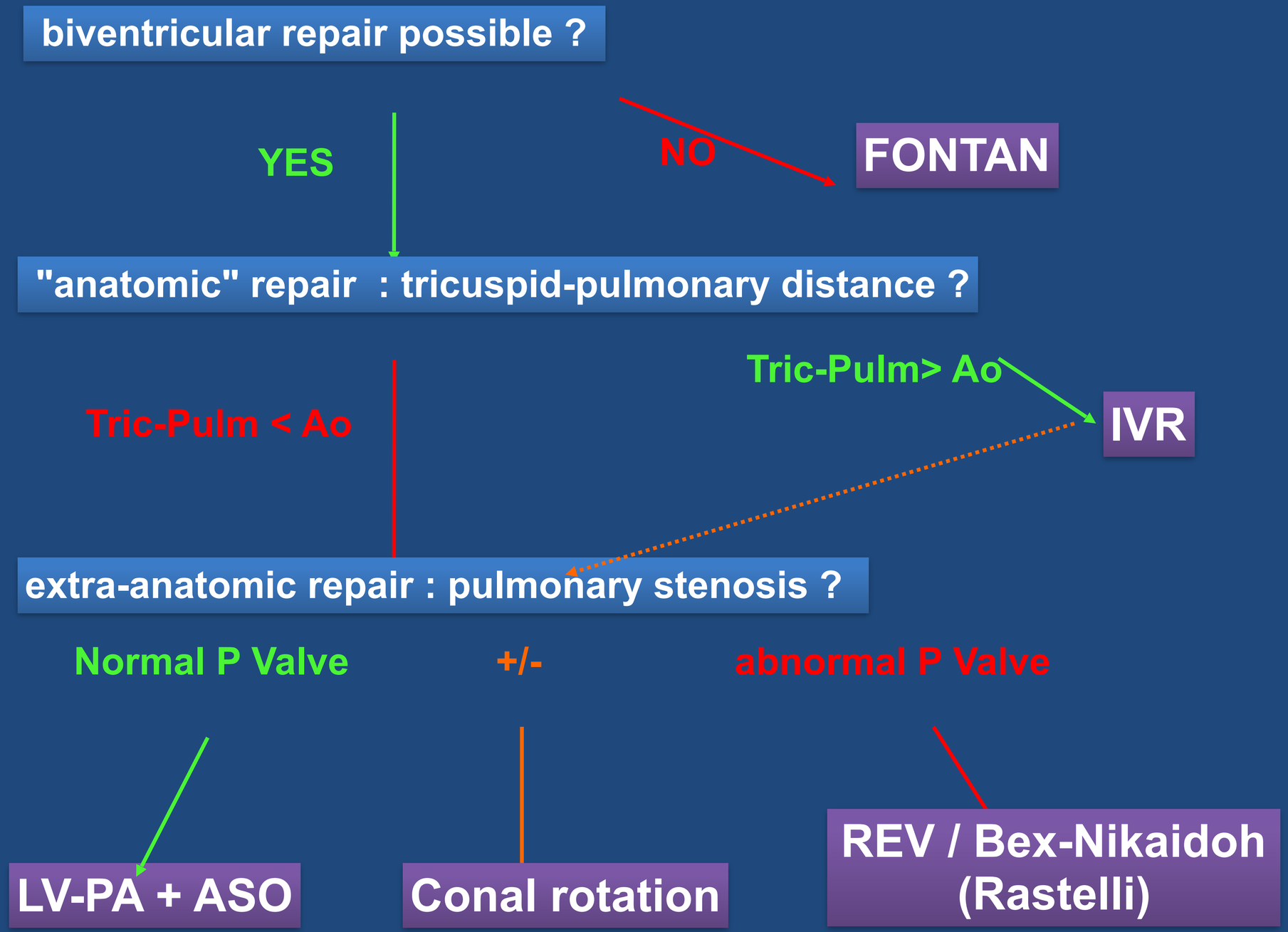


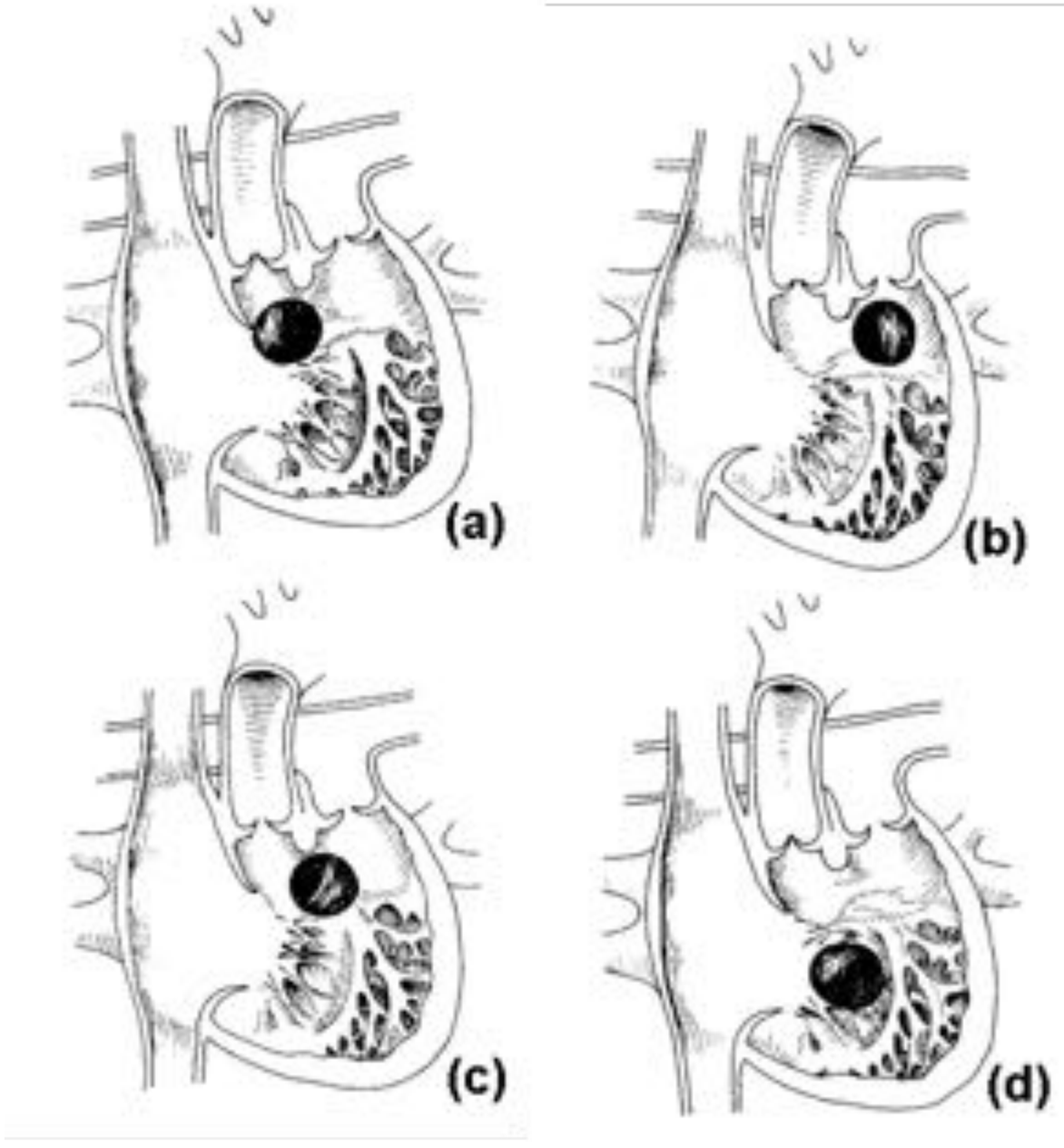
Conotruncal rotation procedure

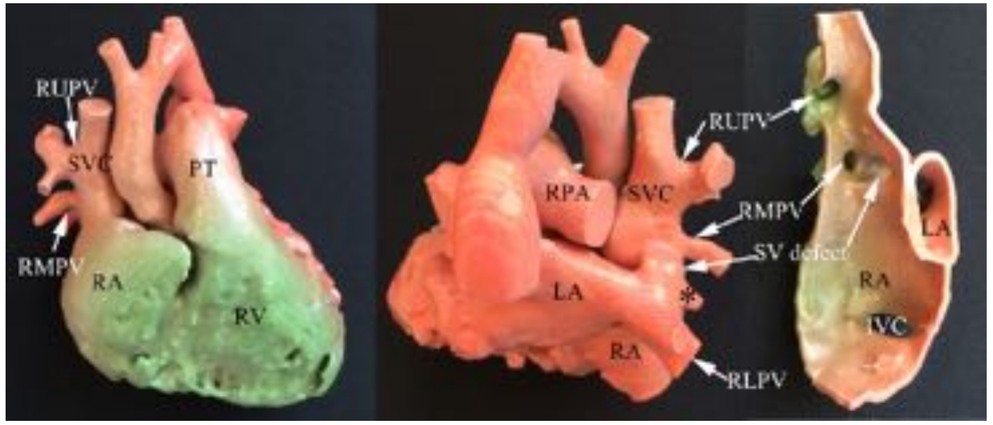


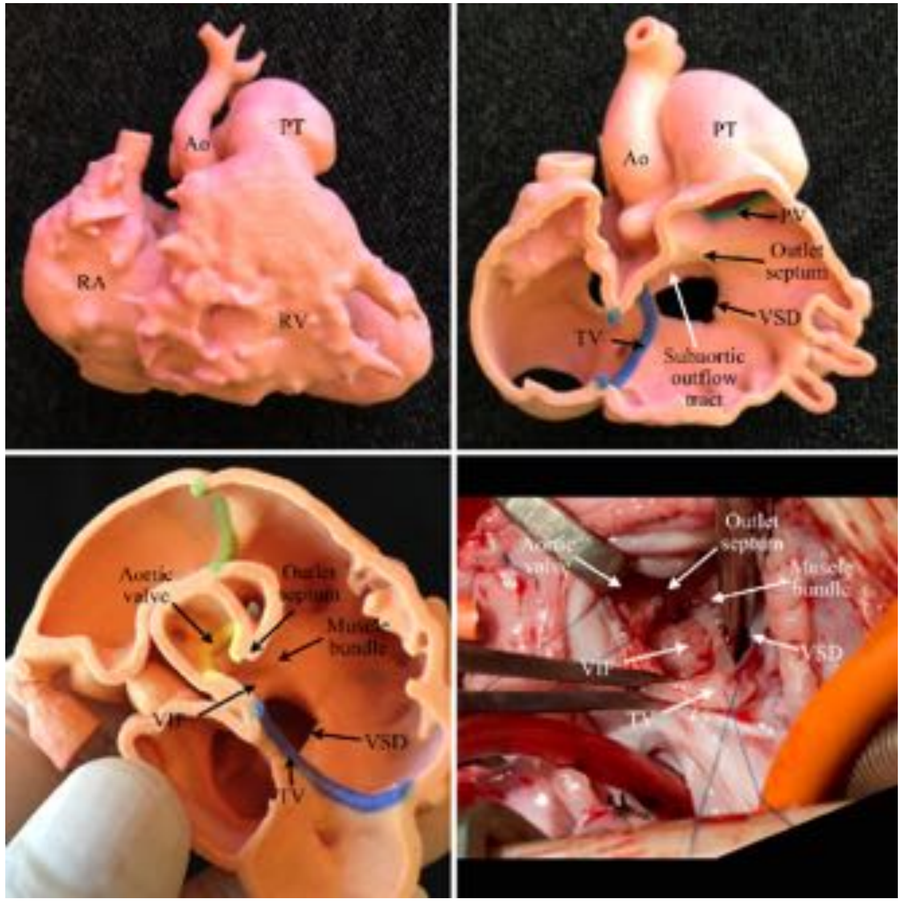
Conotruncal rotation procedure




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)


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


RUPV
SVC
PT
RMPV
RA
RV
RUPV
RPA
SVC
RMPV
SV defect
LA
RA
IVC
LA
RA
RLPV


Ao
PT
RA
RV
Ao
PT
PV
Outlet septum
VSD
TV
Subaortic outflow tract
Aortic valve
Outlet septum
Muscular bundle
VSD
TV
Outlet septum
Muscular bundle
VSD

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Continuing Medical Education

What is anatomically corrected malposition?

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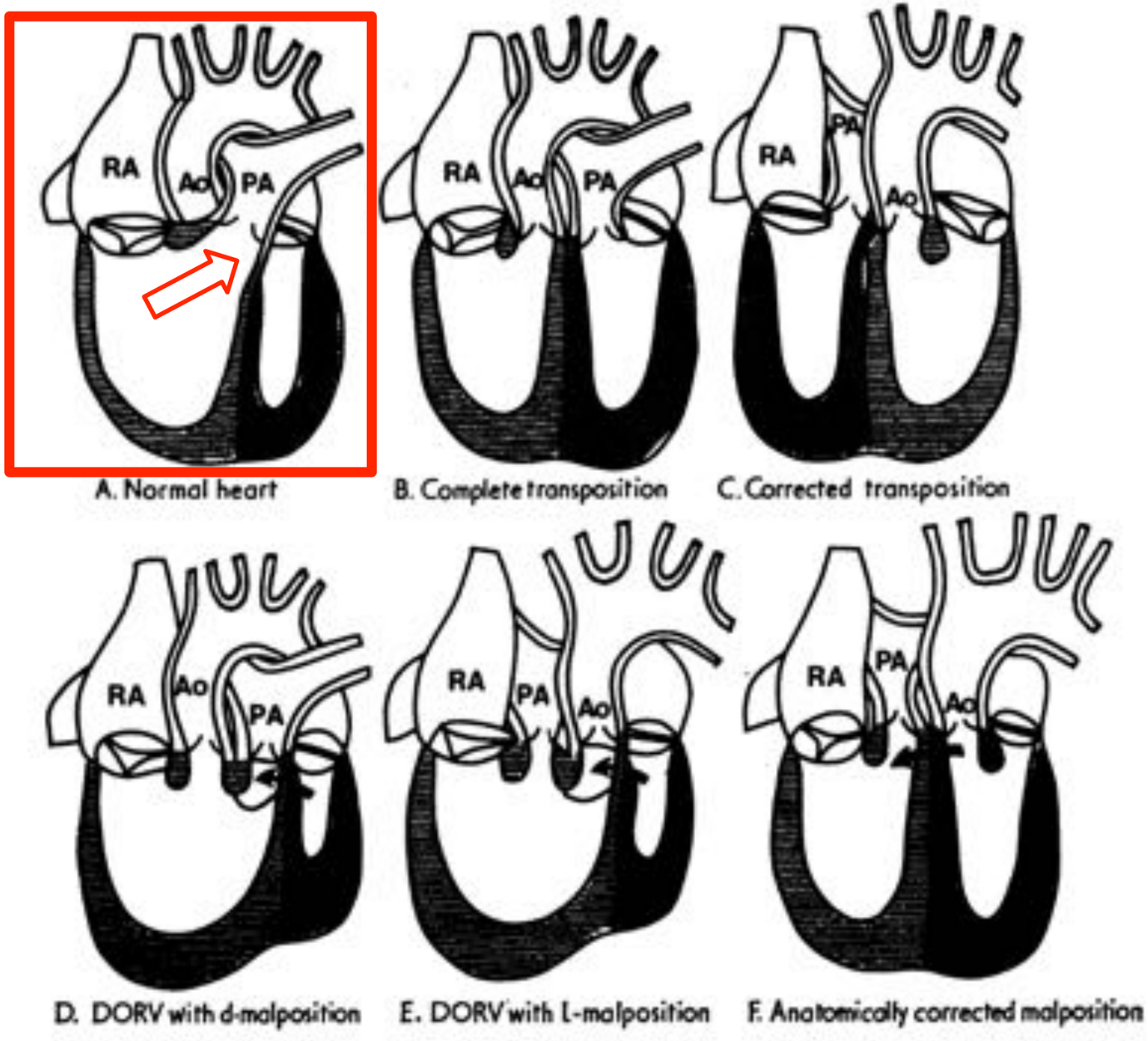
«It remains a fact, nonetheless, that many paediatric cardiologists and surgeons remain unaware of the significance of the malformation»

« On est un certain nombre à avoir fait l'impasse sur la question! »

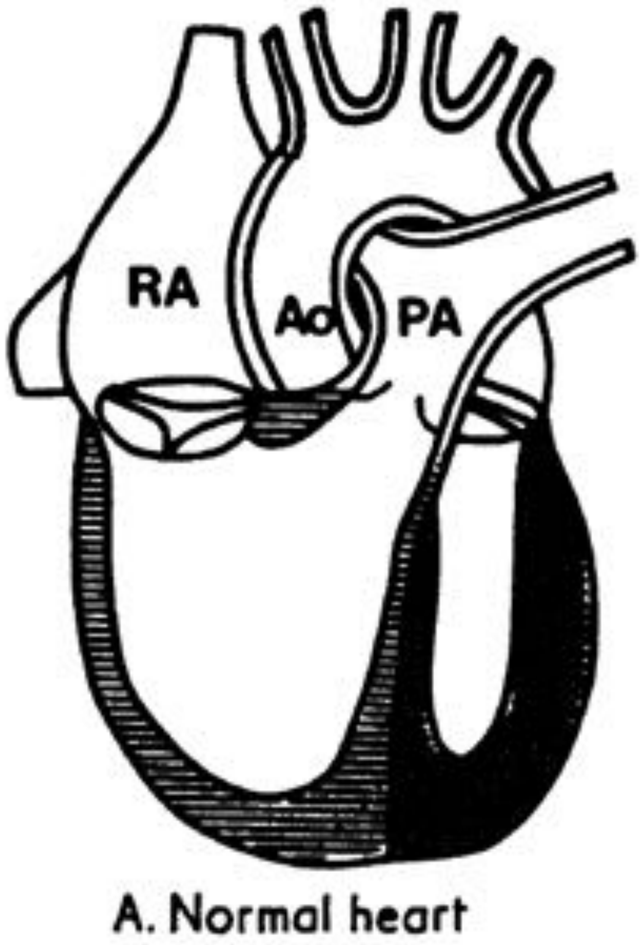
Jonction ventriculo-artérielle



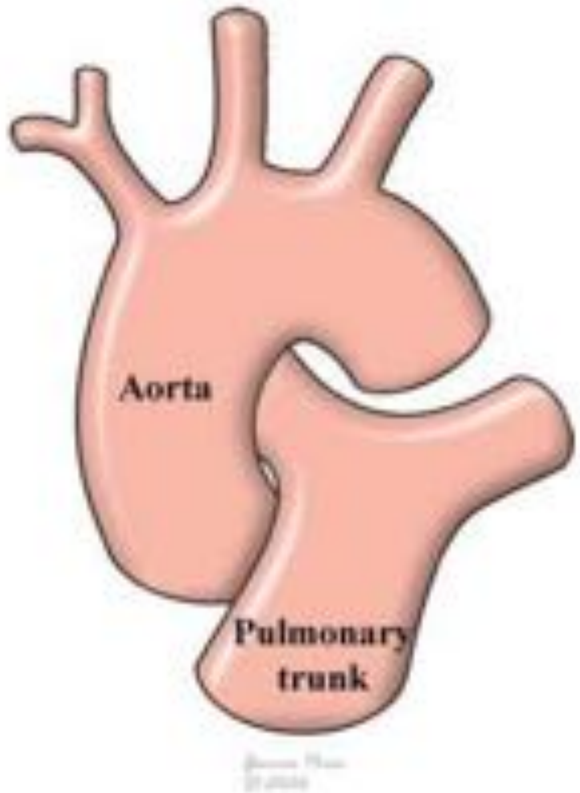
1. les gros vaisseaux sortent de leur ventricule respectif

CŒUR NORMAL

Situs solitus
Topologie ventriculaire: D loop

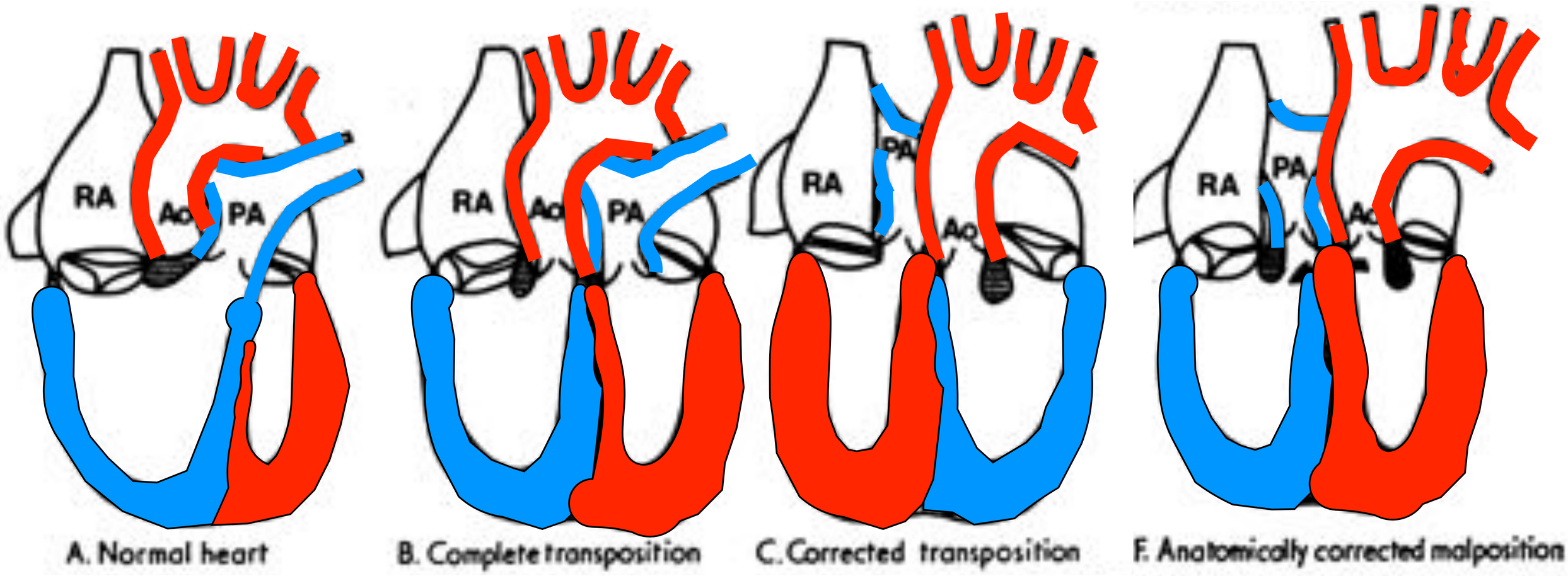


A. Normal heart



2. Relation spatiale particulière des deux gros vaisseaux entre eux:
Aorte postérieure et droite, AP croise antérieurement pour aller à gauche et en avant: aspect spiralé des 2 vx

Malposition anatomiquement corrigée



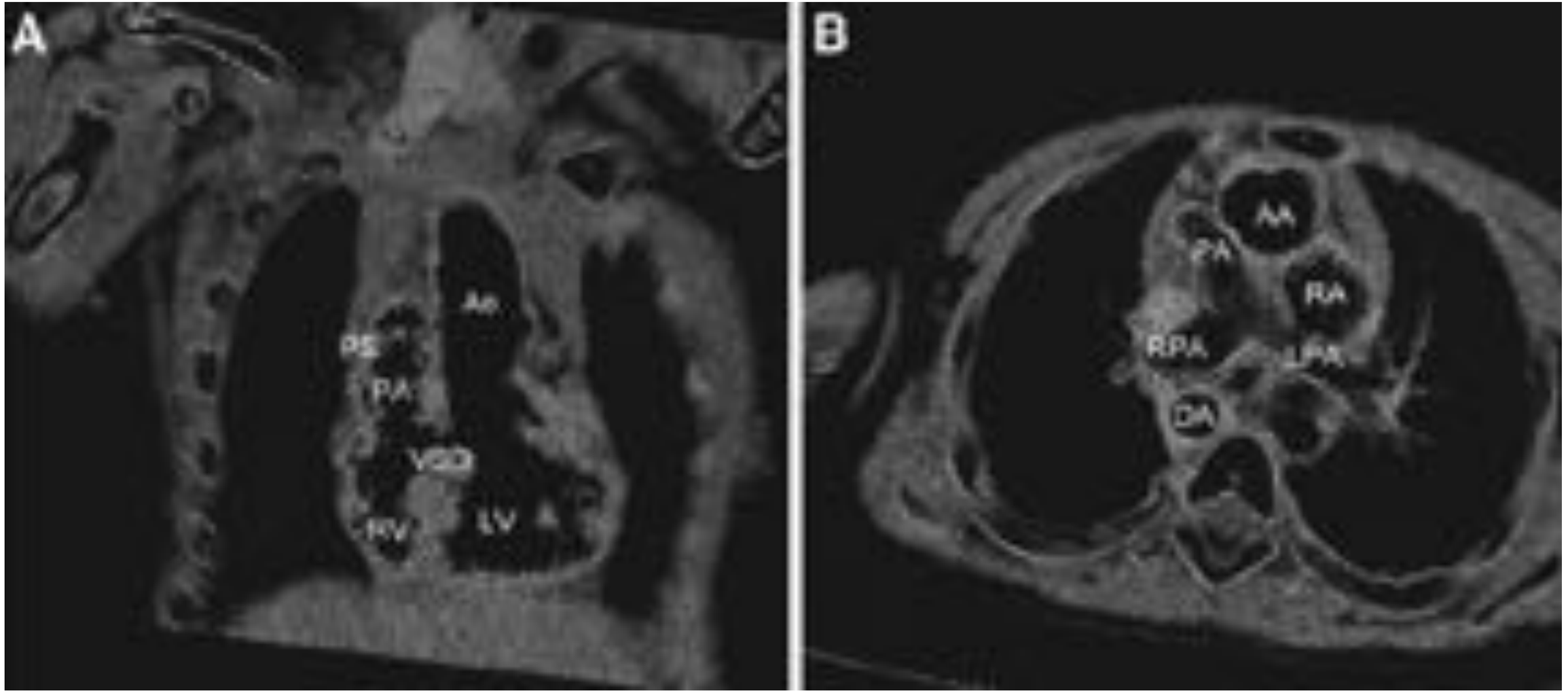
1. les gros vaisseaux sortent de leur ventricule respectif
2. Les 2 gros vx sortent parallèles des massifs ventriculaires, avec Aorte en avant et à gauche, l'AP à droite et postérieure

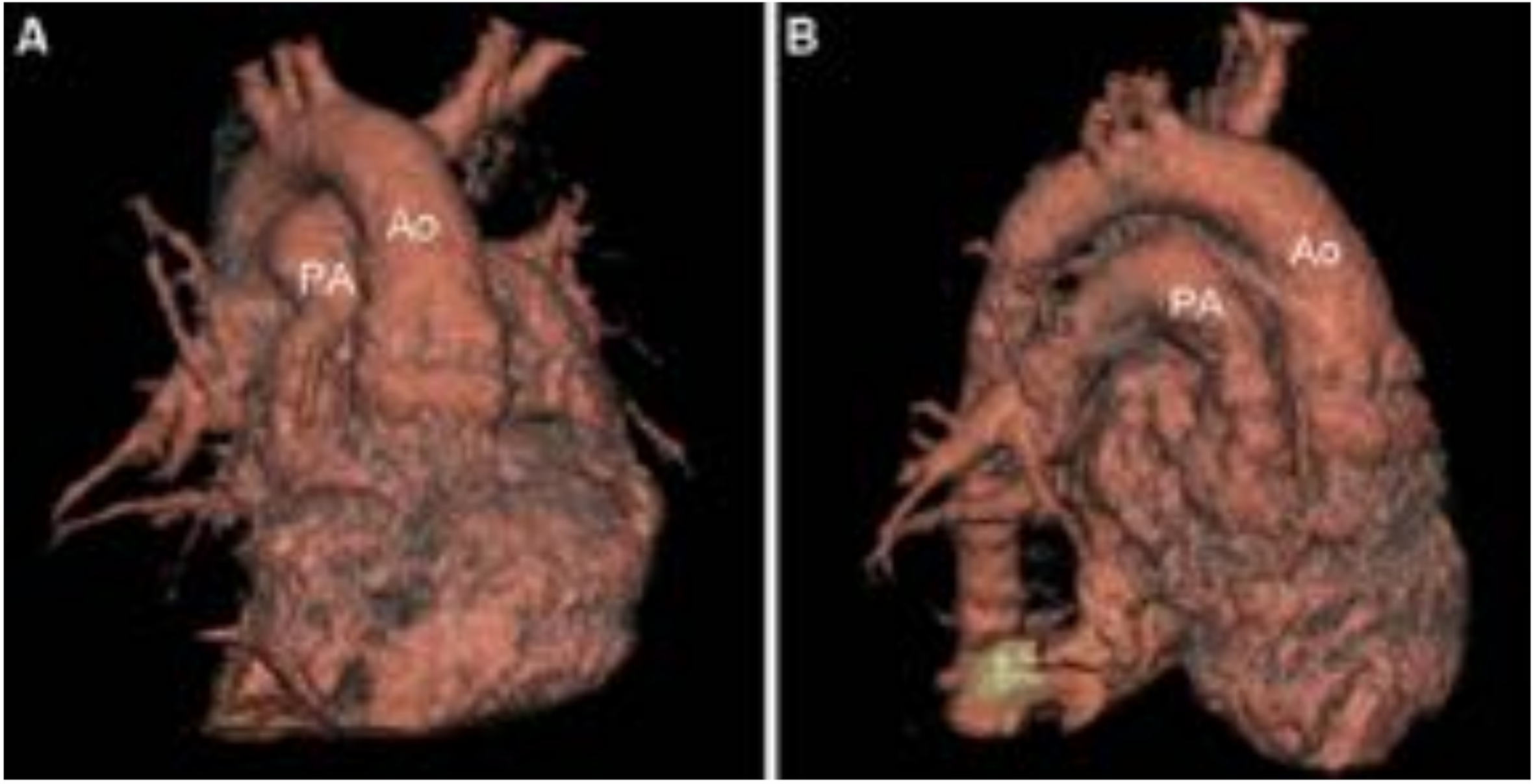
Pediatr Cardiol (2008) 29:467–468
DOI 10.1007/s00246-007-9080-4

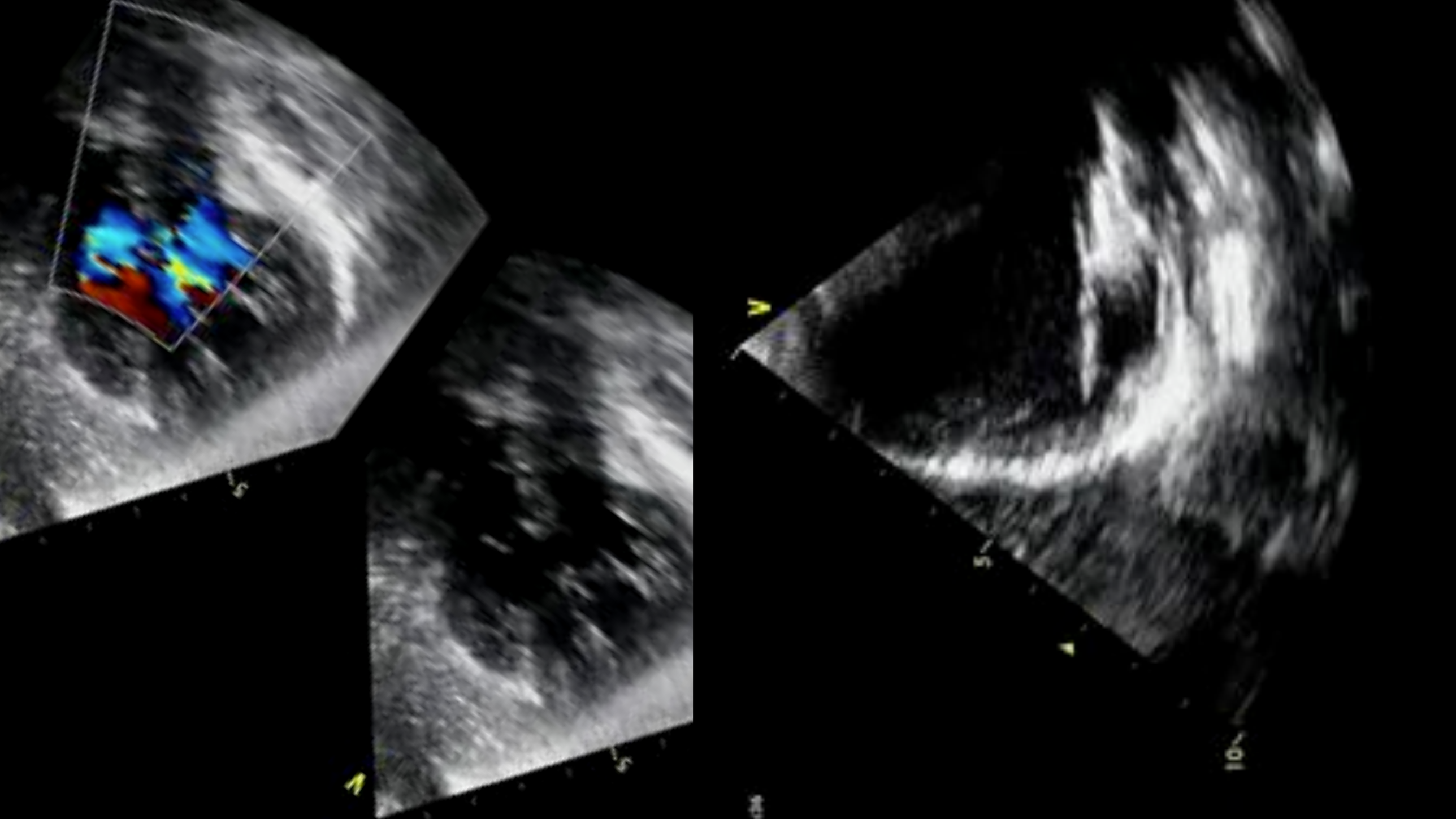
IMAGES IN PEDIATRIC CARDIOLOGY

Anatomically Corrected Malposition of the Great Arteries

Ming-Ren Chen




A
Ao
PA
B
Ao
PA




V
2
4
6
1:232
V
2
4
6

Double discordance