Transposition of the great arteries in neonates

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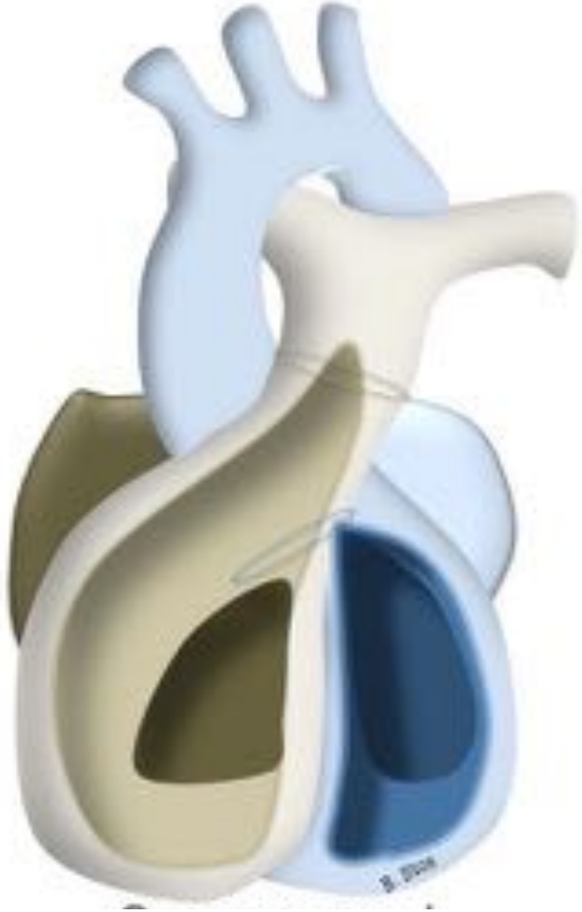
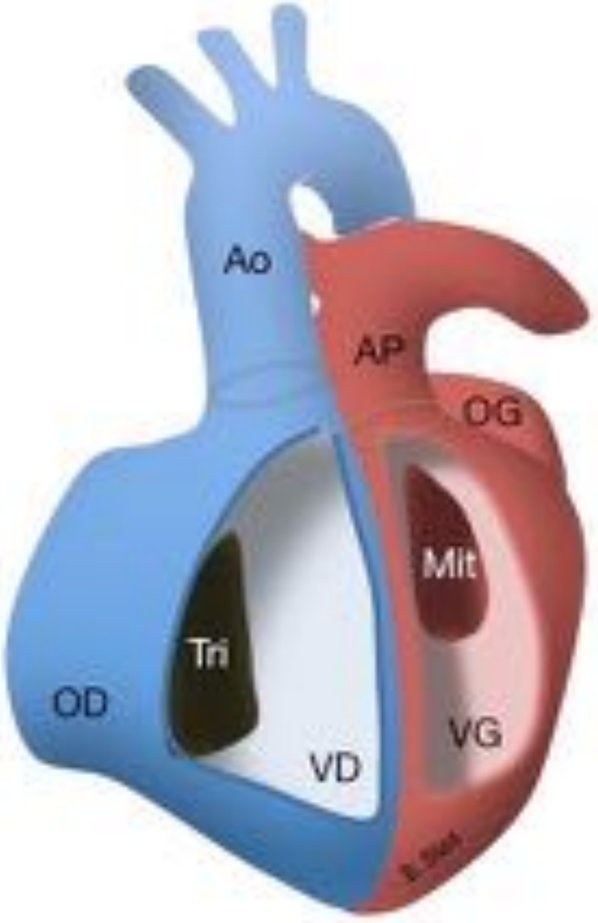
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

ANATOMY OF THE HEART WITH TGA

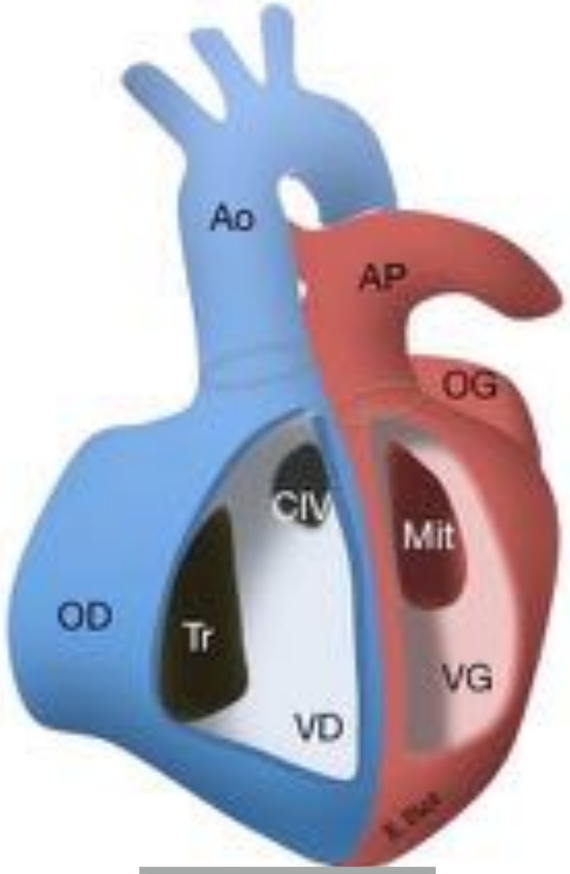


Normal heart

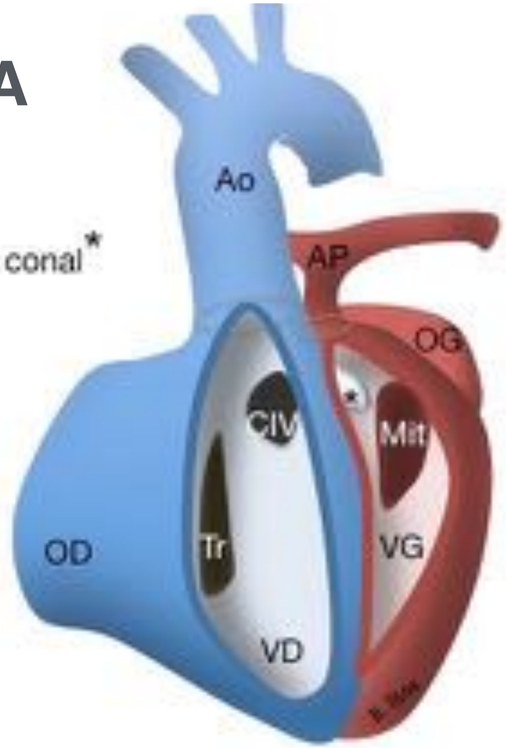


TGA

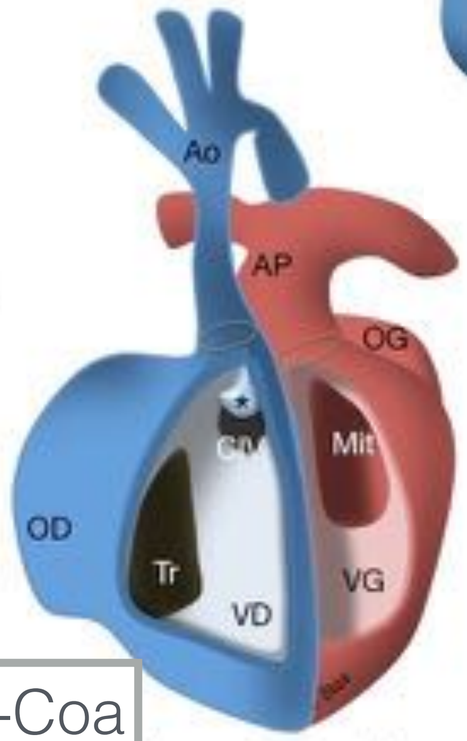
Other complex malpositions of the GA



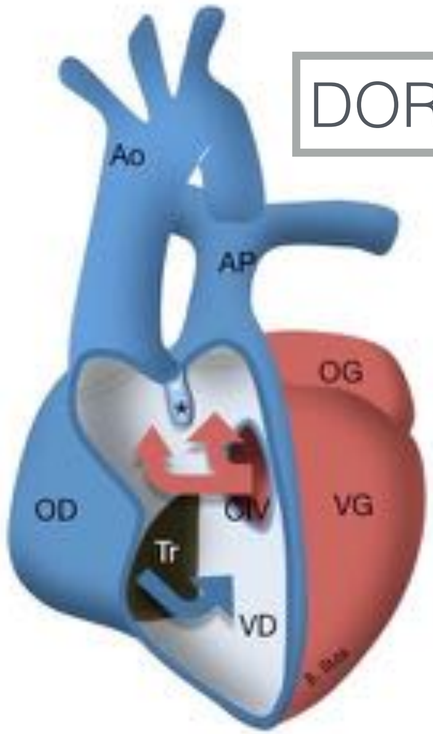
TGA-VSD



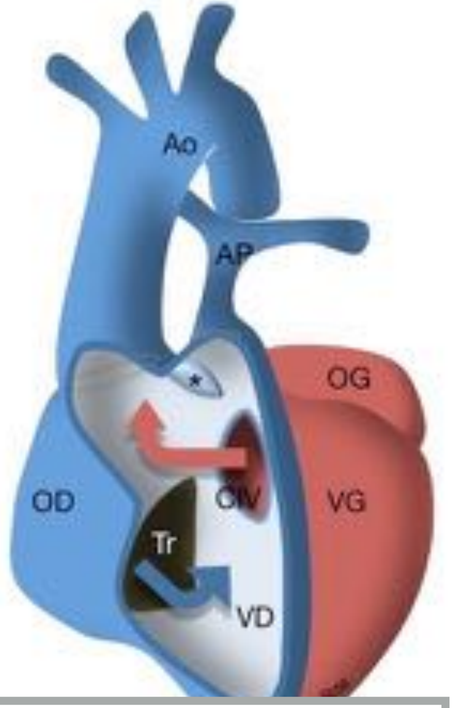
TGA-VSD-PS



TGA-VSD-Coa



DORV-Sub aortic VSD

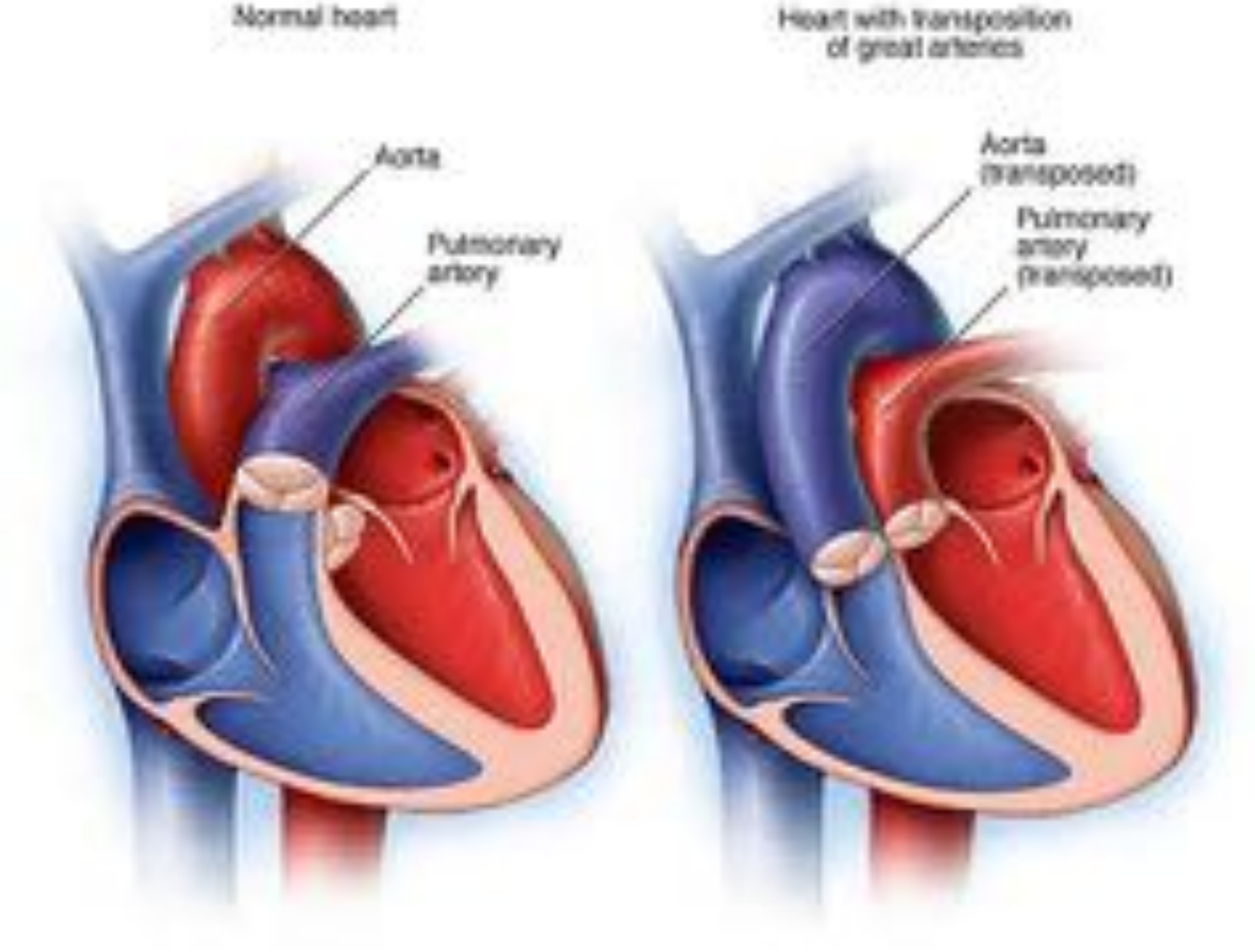


DORV-Sub aortic VSD-SP

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

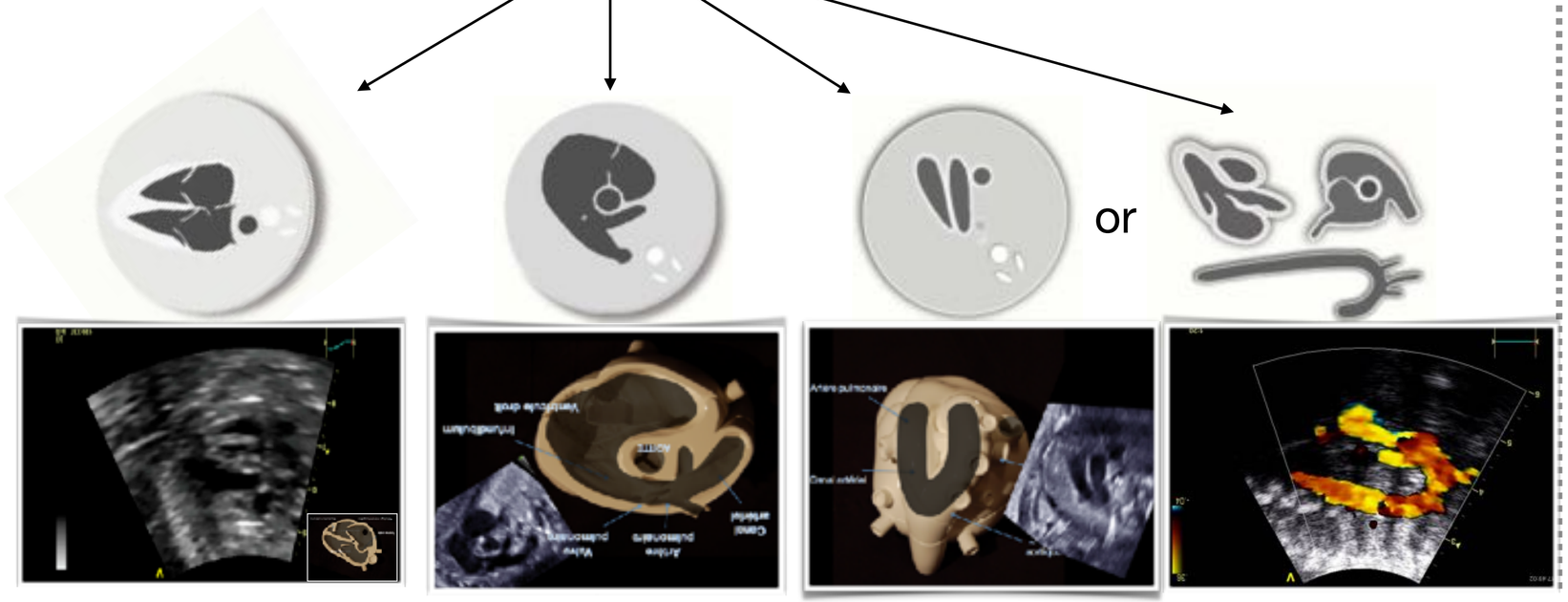
3 systematic foetal echographies - Level 1

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

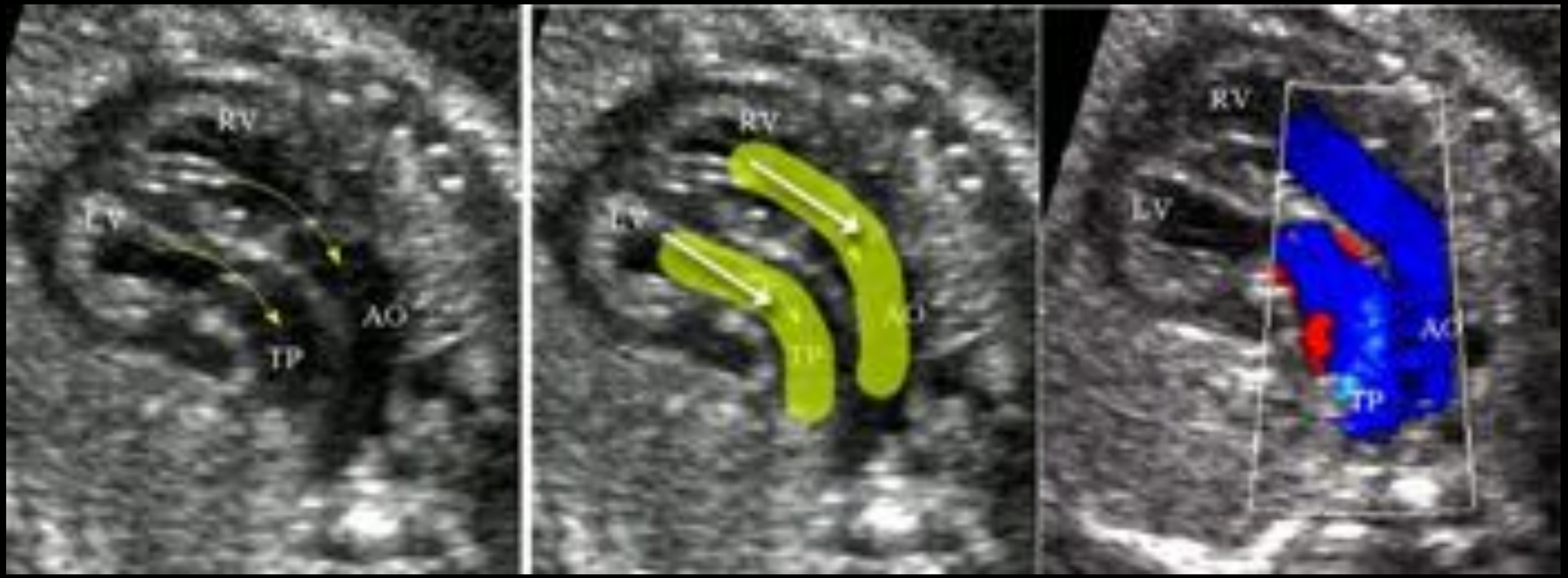
Preoperative mortality in TGA = 4-6%
(*vs./+)* Surgical mortality = 1-2%

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	48 800	
Normal	168	47	NS
Abnormal	60	31	NS
Postoperative mortality	30	0	<0.01
Hospital stay, d	30 ± 17	24 ± 11	<0.01

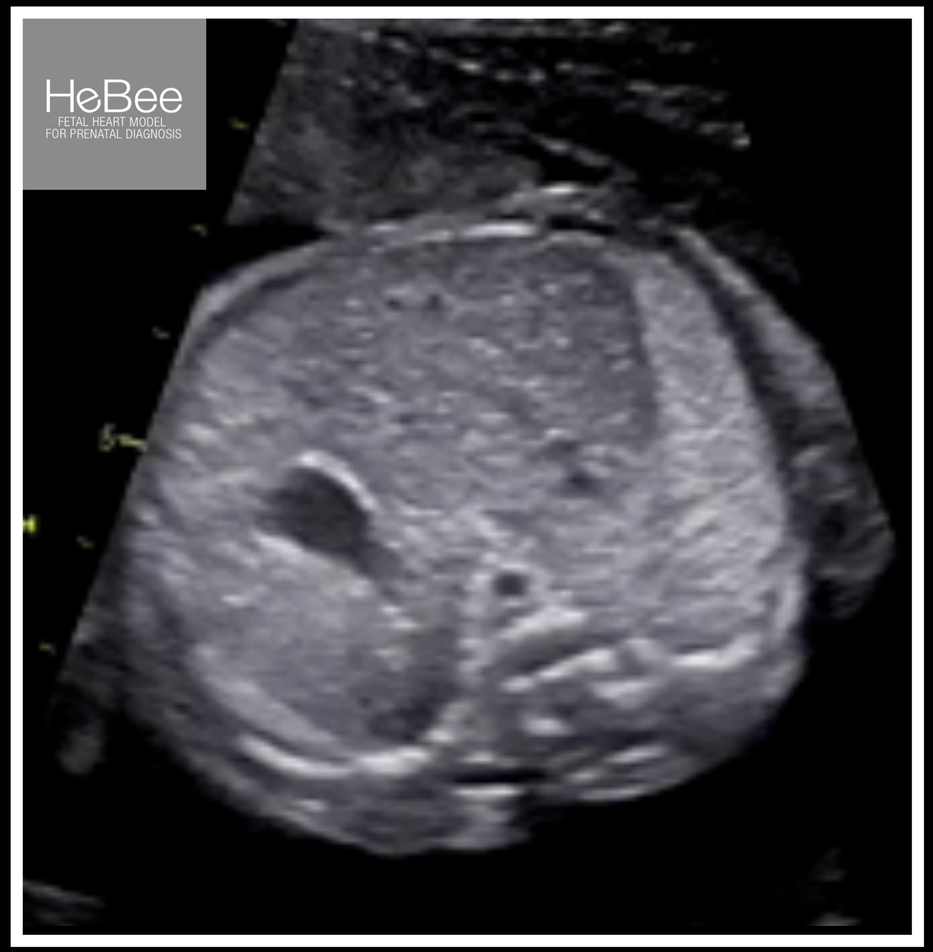
VSD indicates ventricular septal defect; CoA, coarctation; MOF, multiorgan failure; PGE, prostaglandin E; SAS, balloon atrioseptostomy; and ASO, arterial switch operation. Values are n (%).

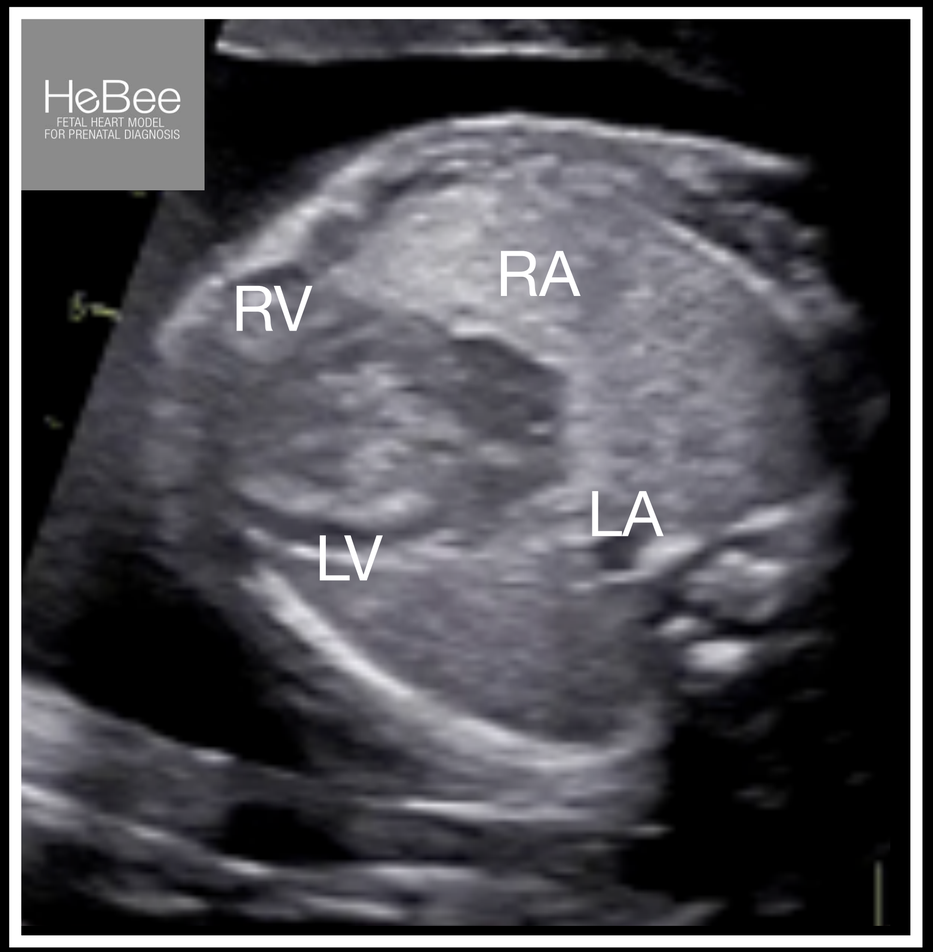
Bonnet D et al. Circulation 1997


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

TGA

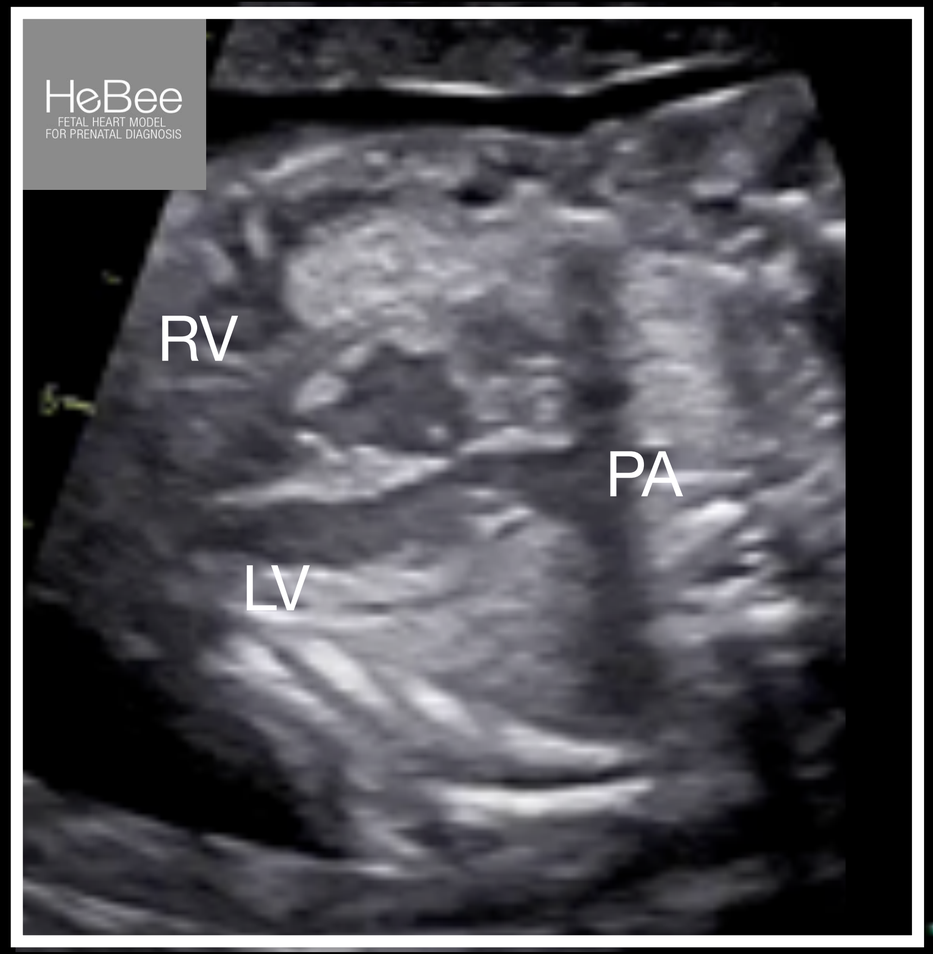
situs & 4 chamber view





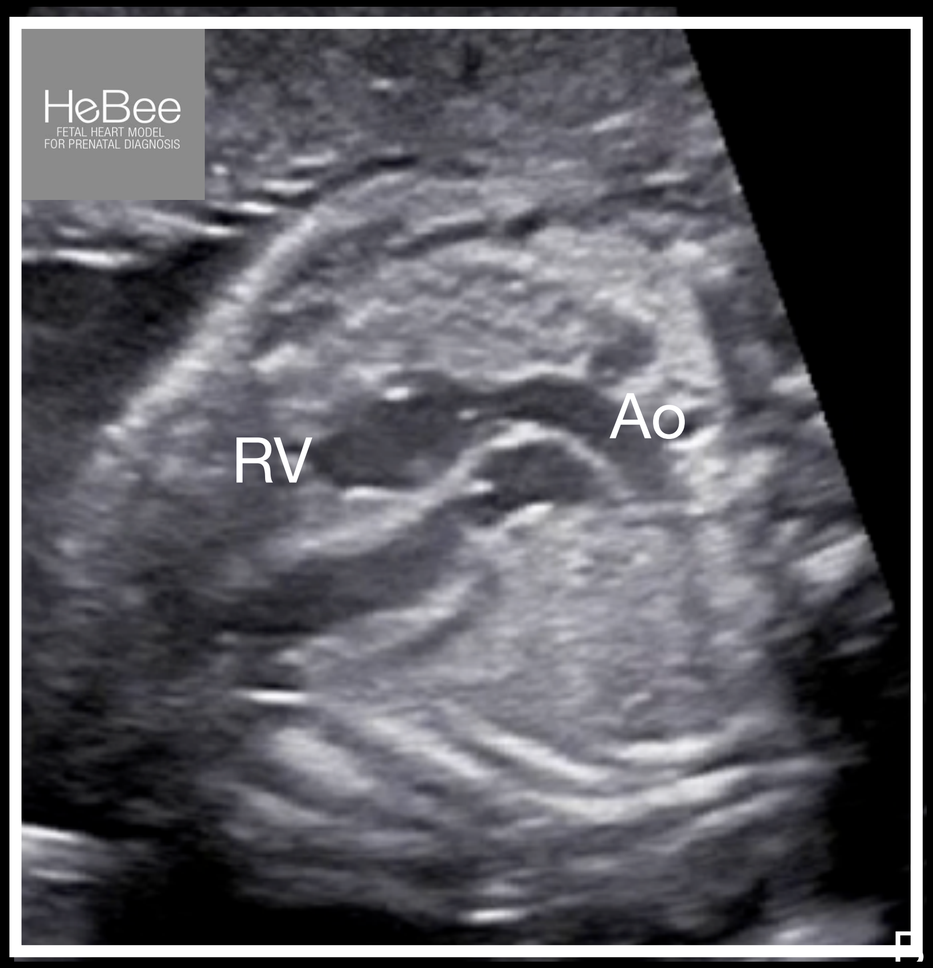
TGA

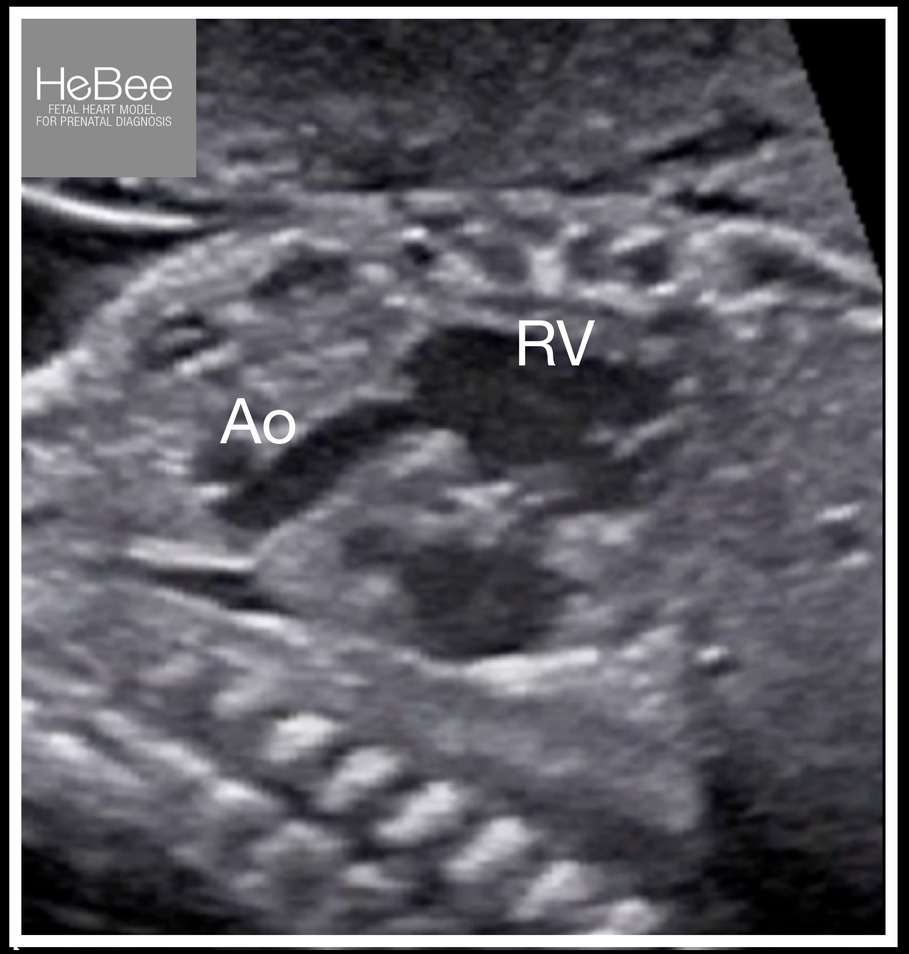
LVOT view



TGA

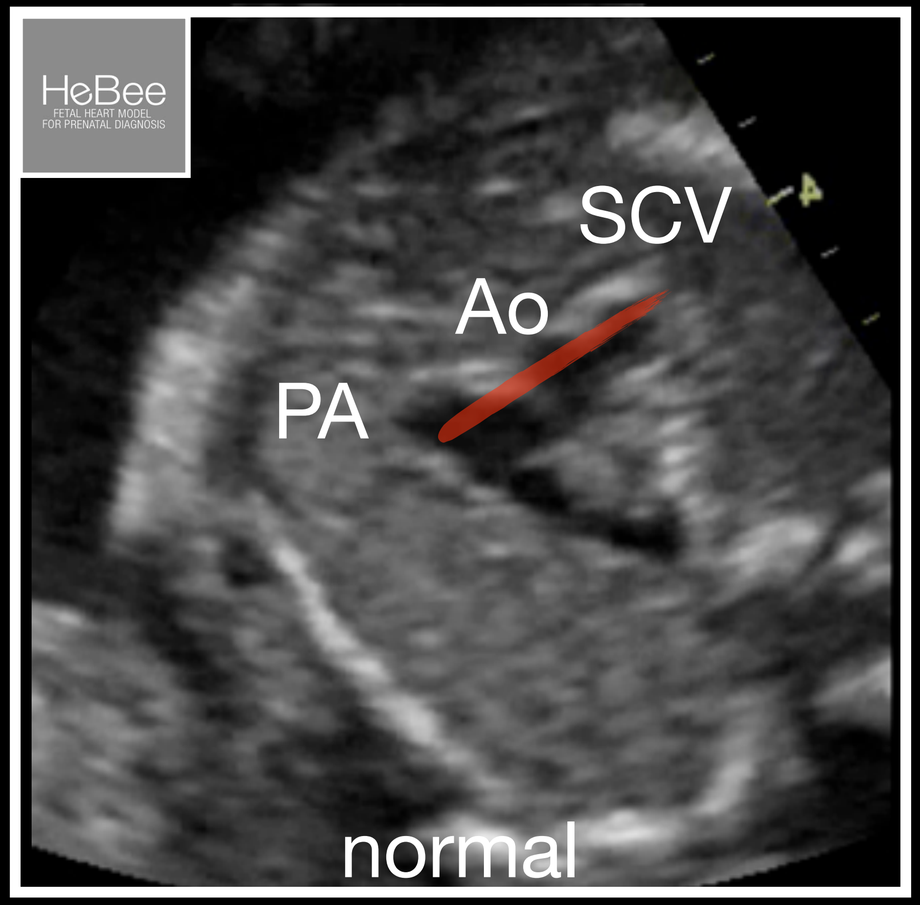
LVOT/RVOT view

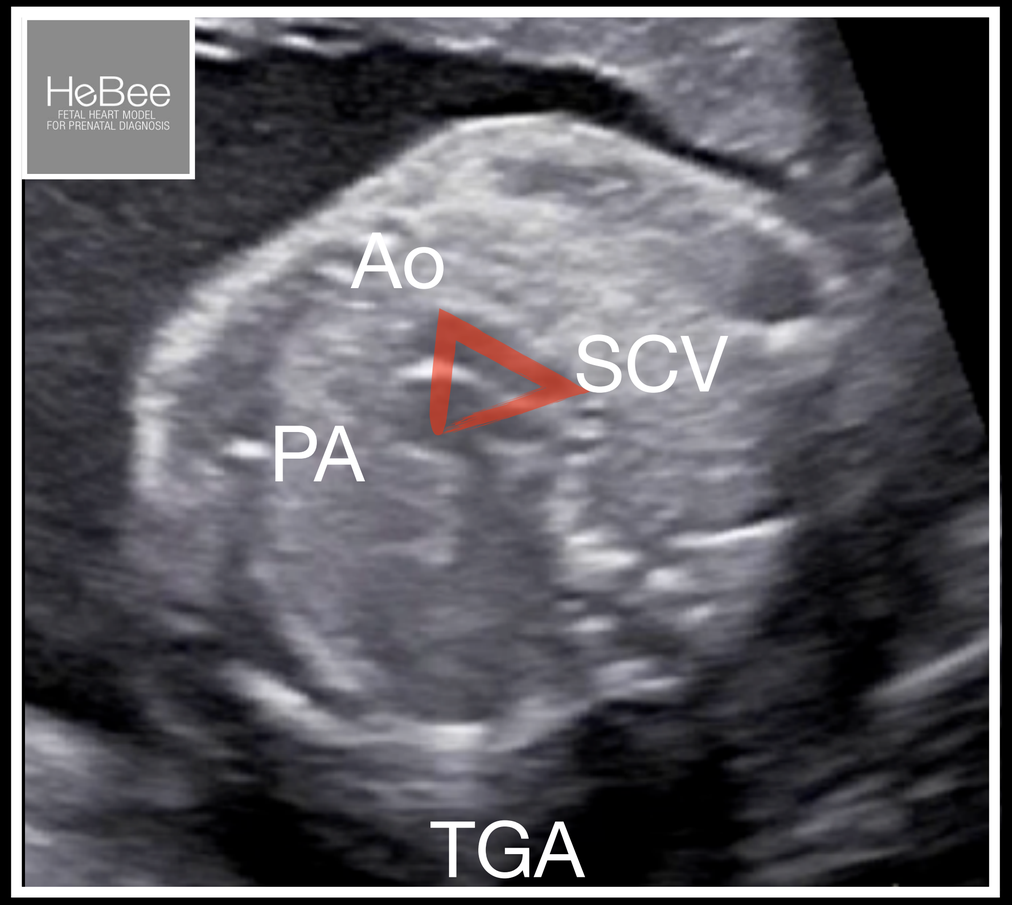




TGA

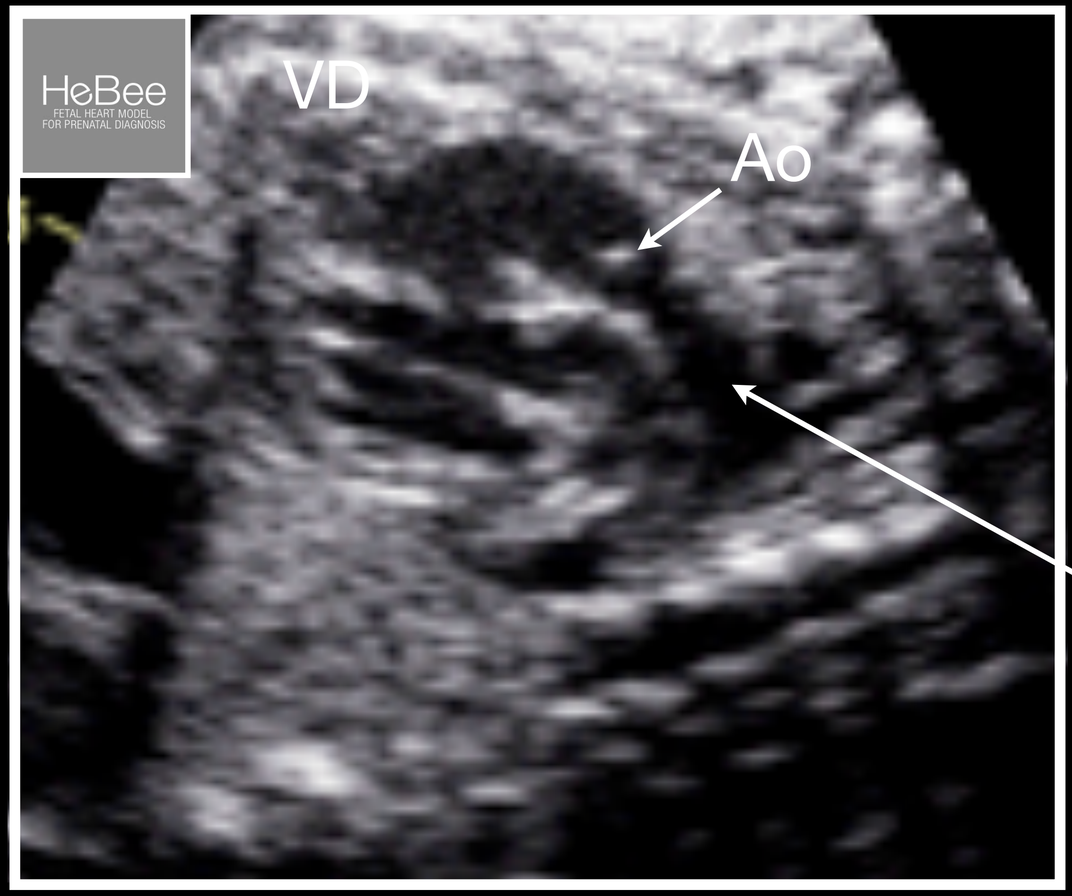
3 vessel view





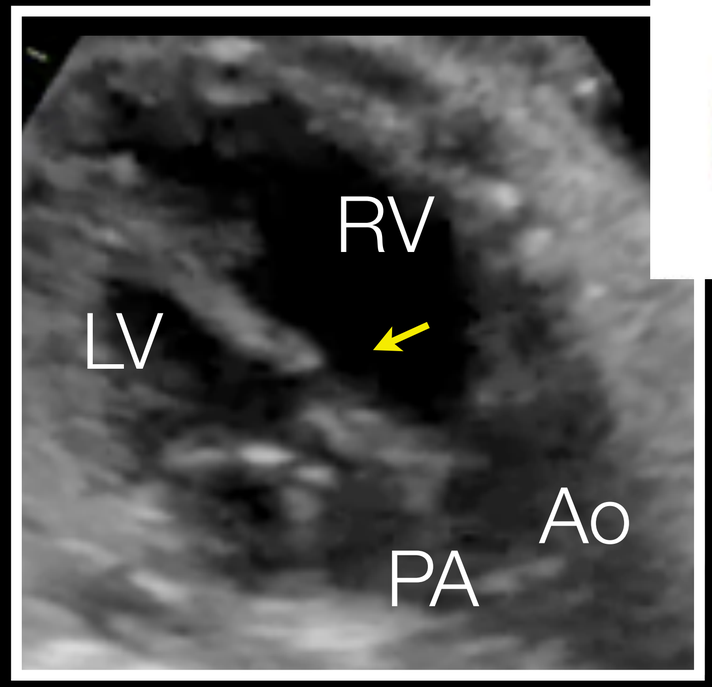
TGA

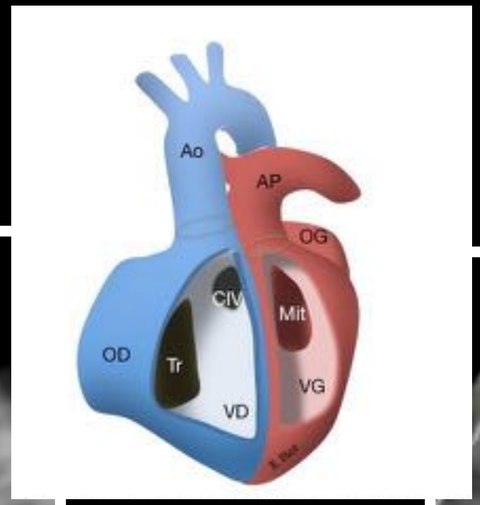
RVOT view

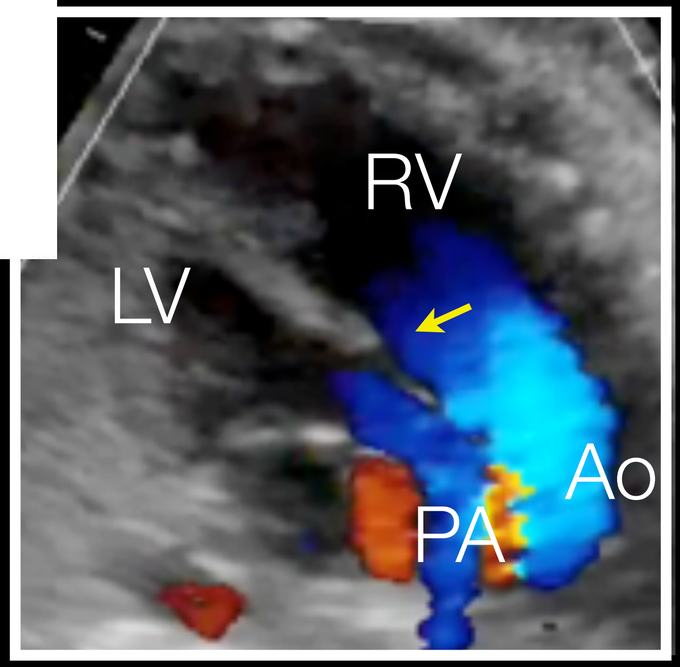


TGA

Complex forms



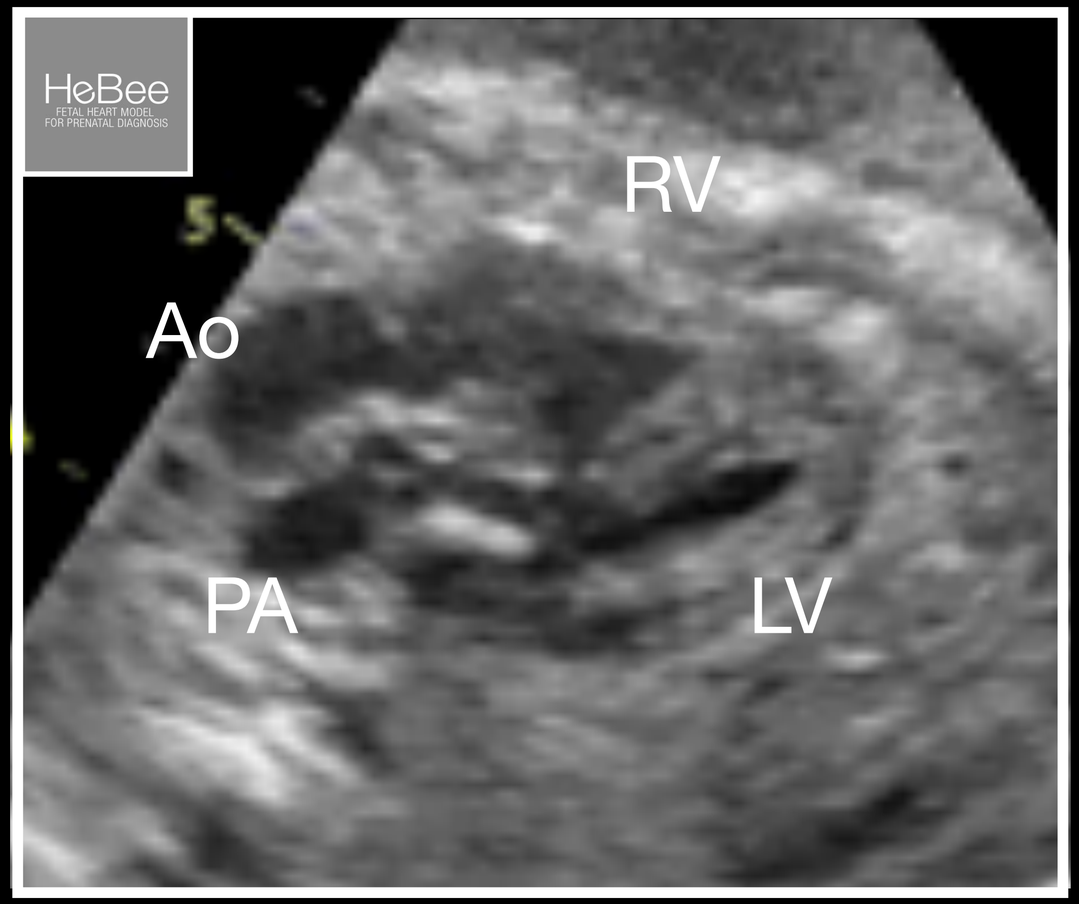




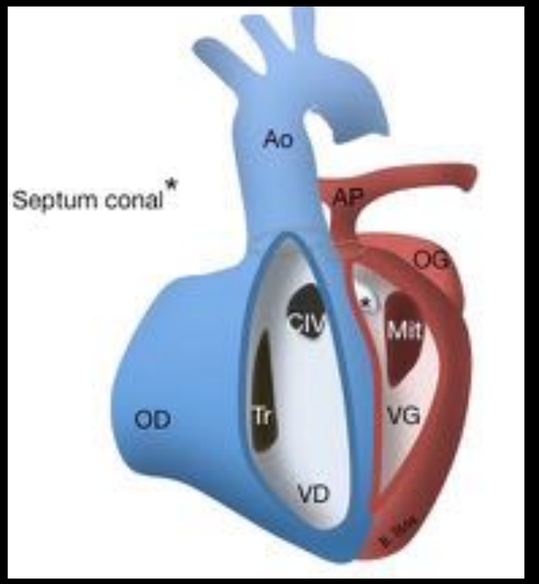
TGA-membranous VSD

TGA

Complex forms

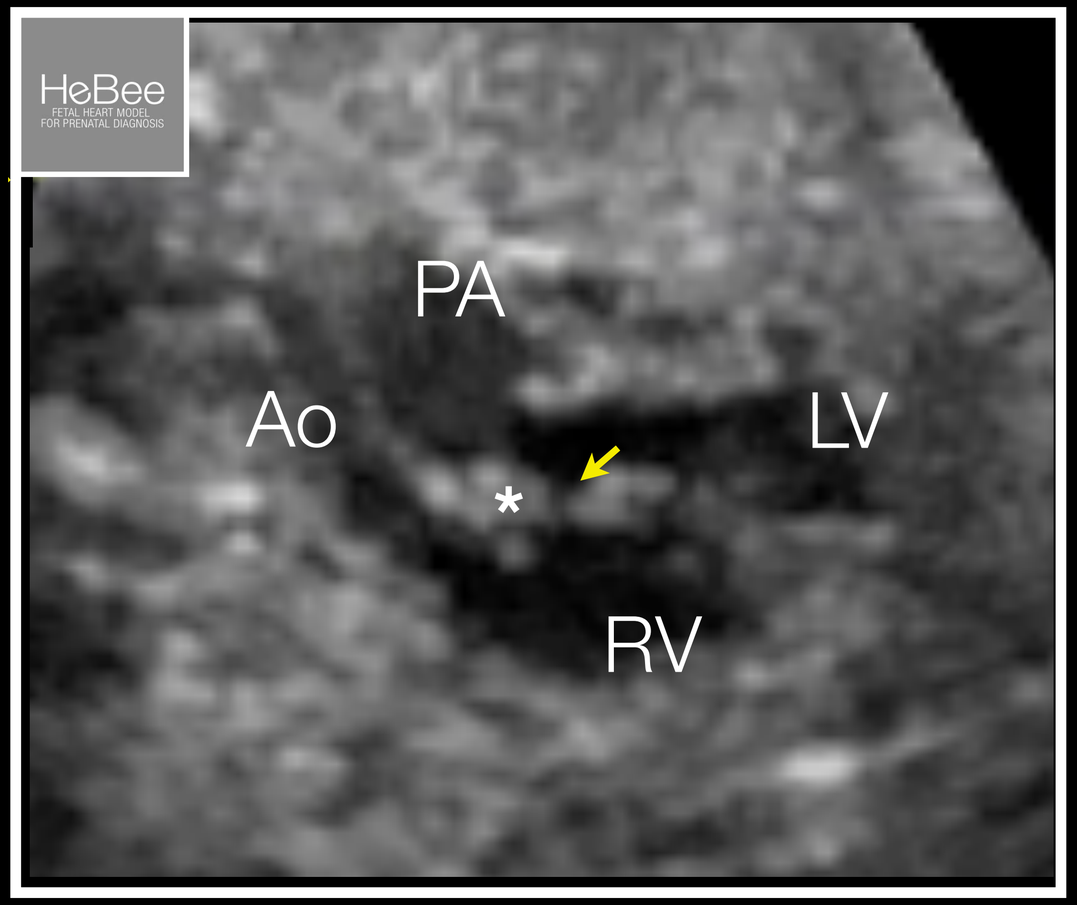


TGA-VSD-PS

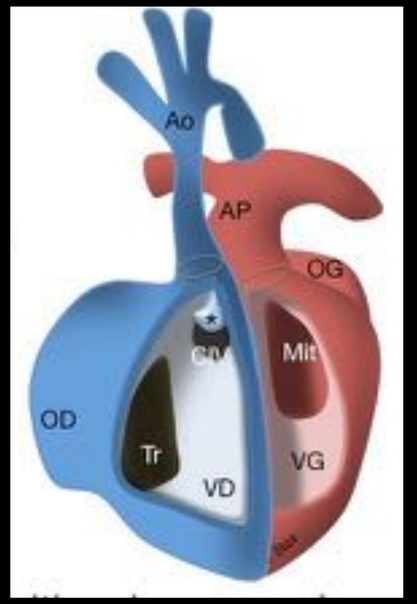


TGA

Complex forms

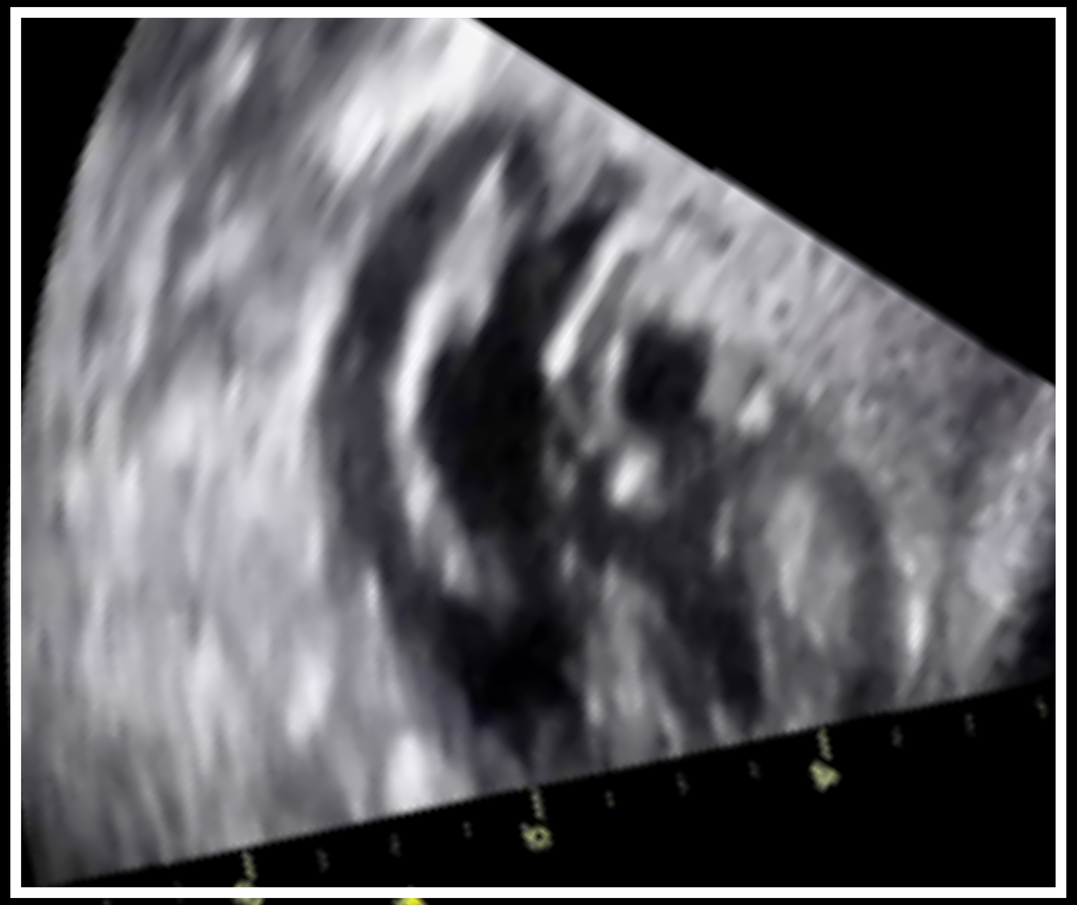


TGA-VSD-Coa

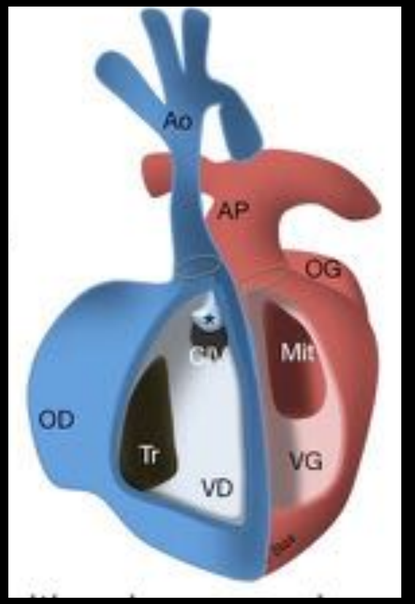


TGA

Complex forms

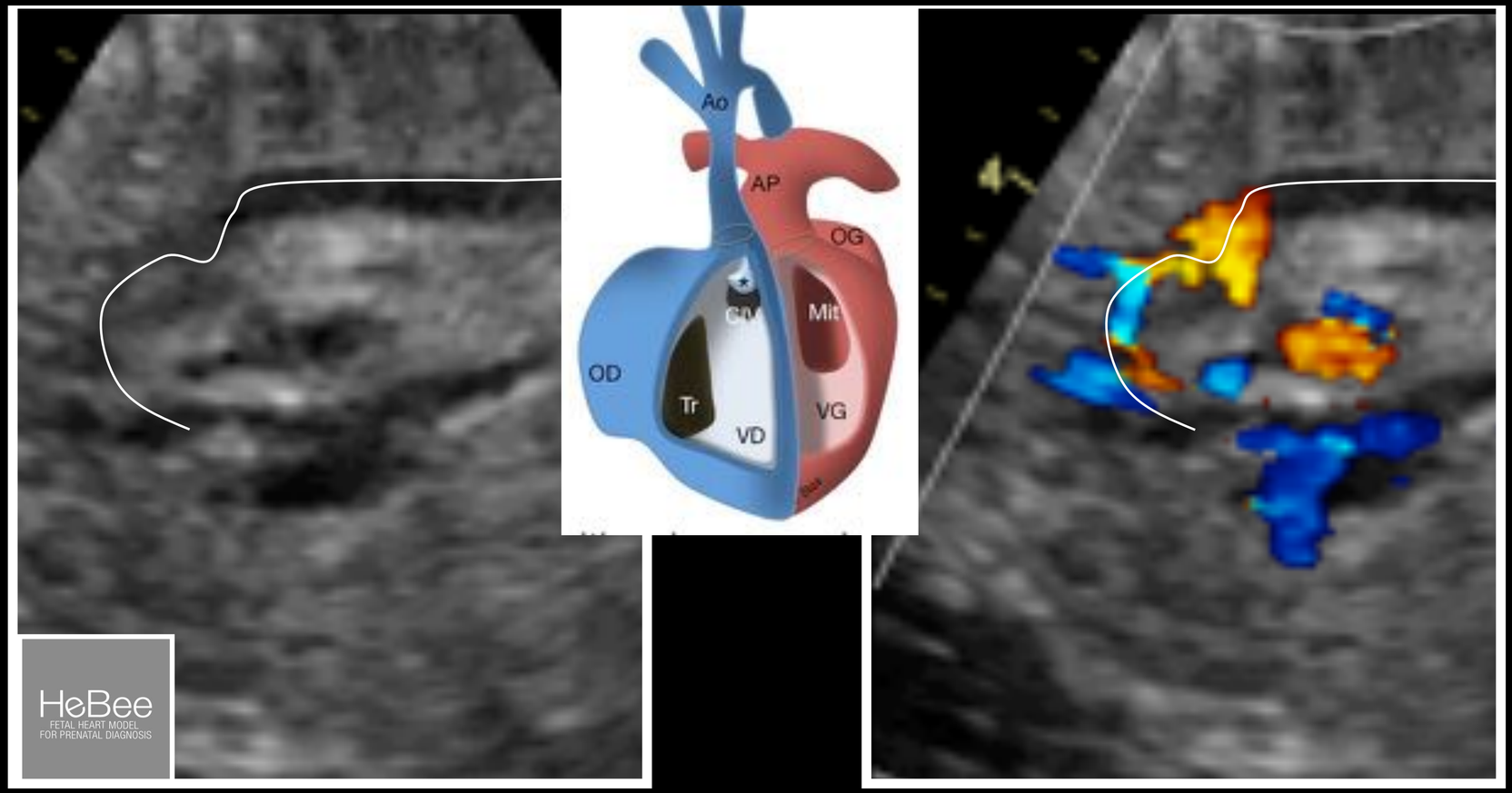


TGA-VSD-Coa



TGA

Complex forms



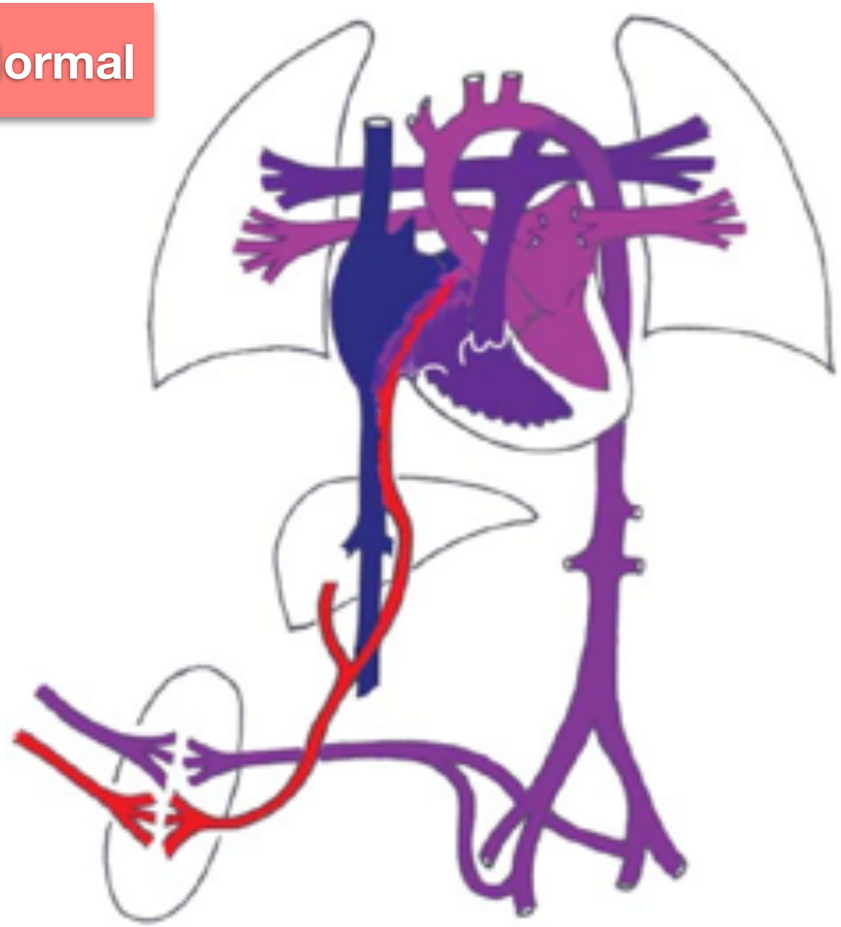
Chromosomal anomalies in fetal CHD

548 CHD-18.5%

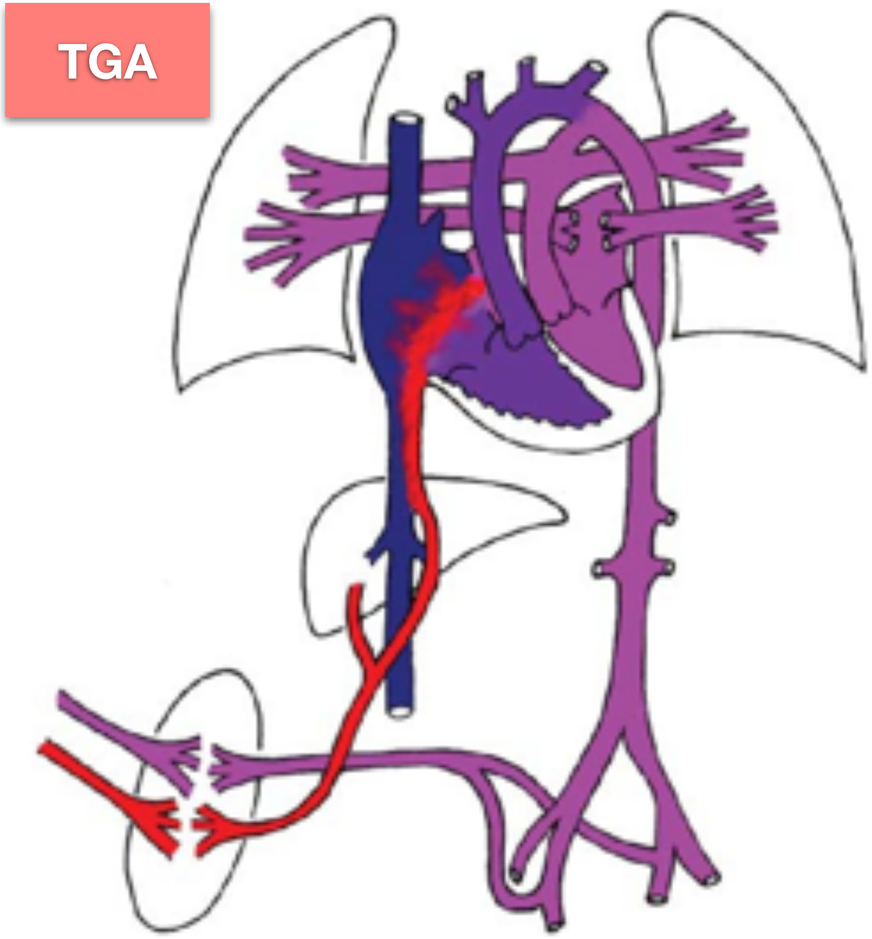
PA-IVS and PS	0	0
Left heart obstruction	12/130	9.2%
6 XO; 3 T18; 3 translocations		
Conotruncal defects	23/91	25%
20 del22q11; 1 T21; 2 translocations		
AVSD	32/68	47%
28 T21; 3 T18; 1 XXX		
VSD	12/74	16%
9 trisomies, 2 del22q11, 1 del5		
Transposition of the great vessels	0	0
DORV	7/38	18%
Univentricular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

Morphological and physiological consequences of the fetal circulation in TGA

Normal

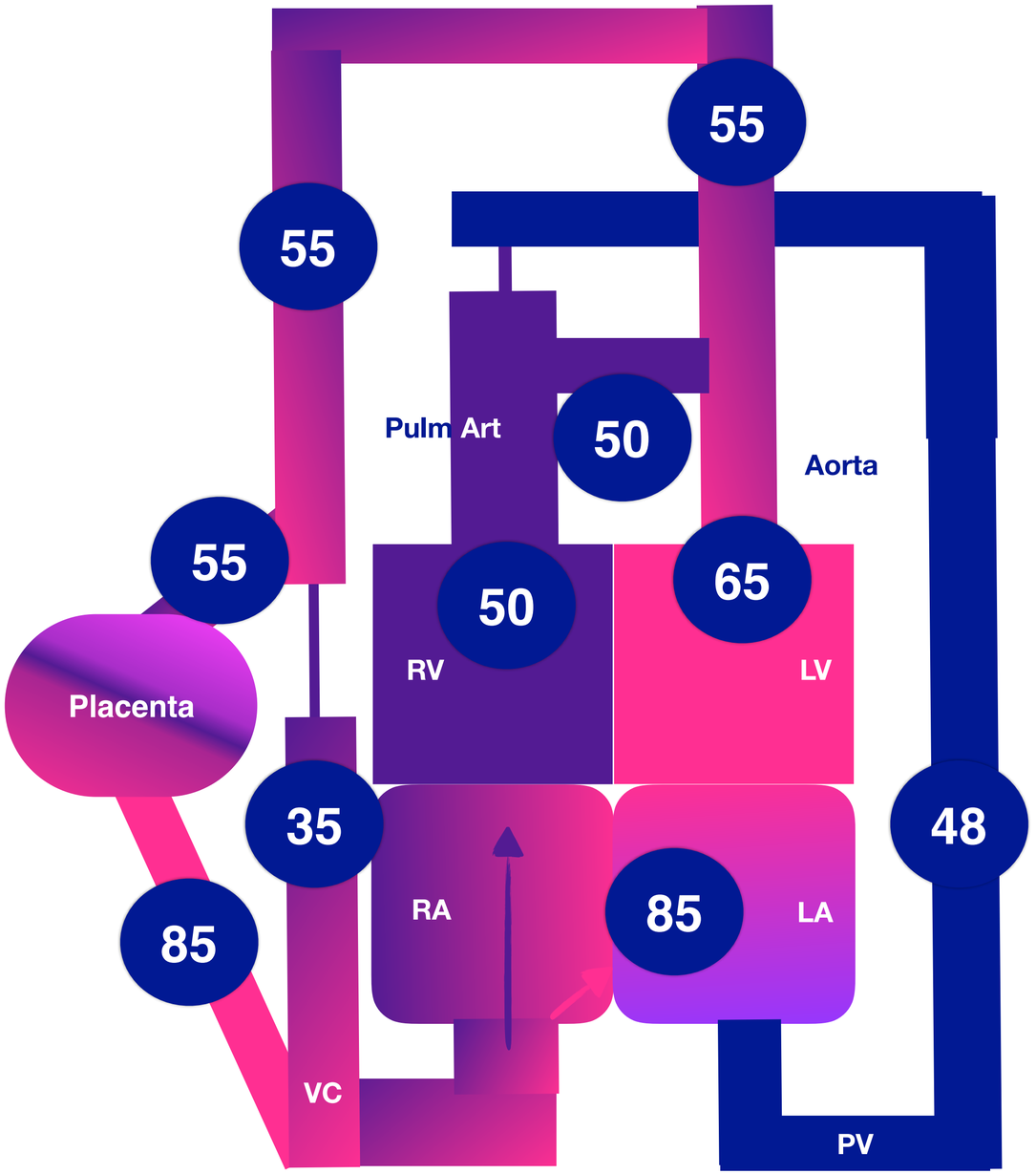


TGA

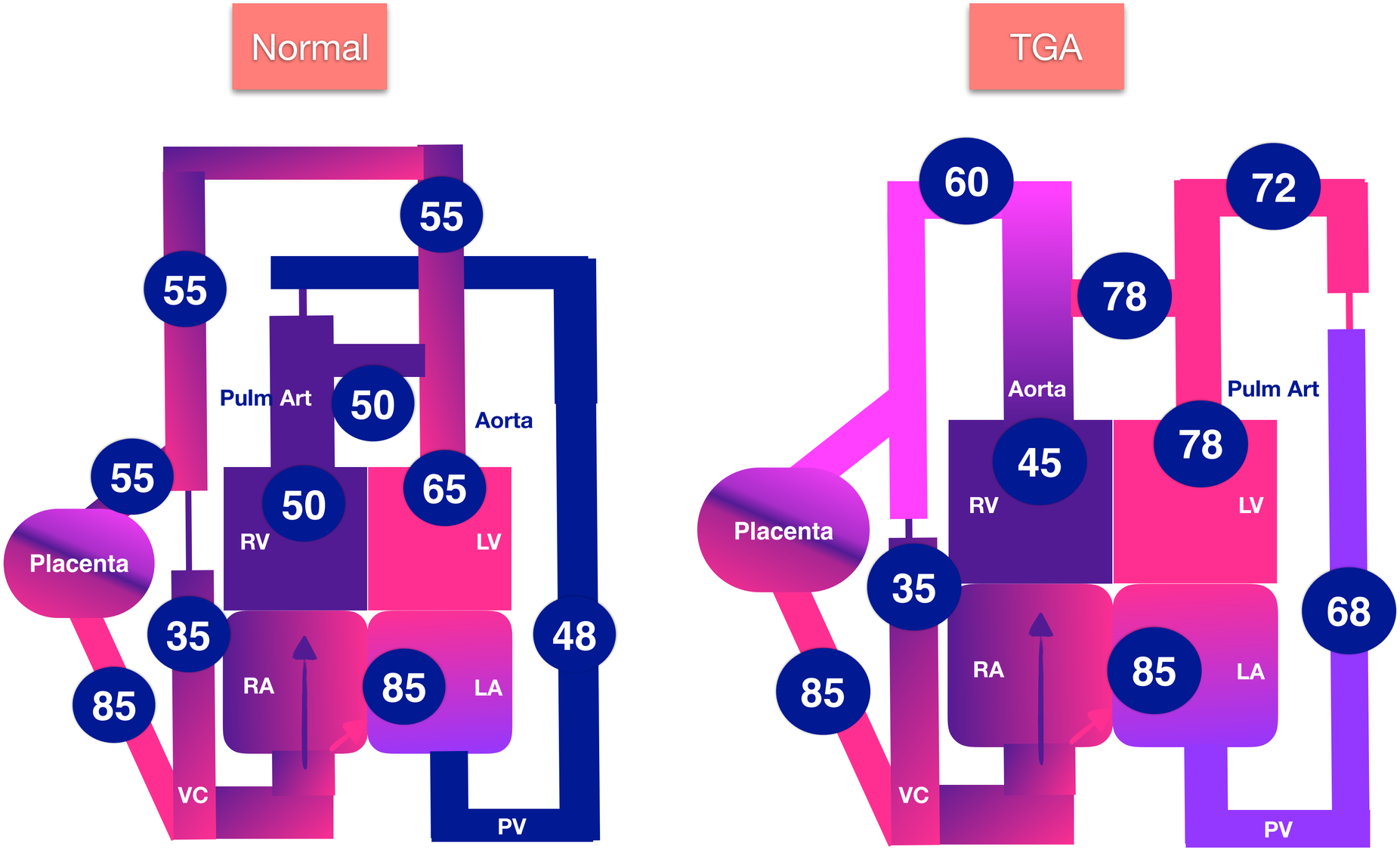


Sun L et al. Circulation. 2015;131:1313-1323.

Morphological and physiological consequences of the fetal circulation in TGA



Morphological and physiological consequences of the fetal circulation in TGA

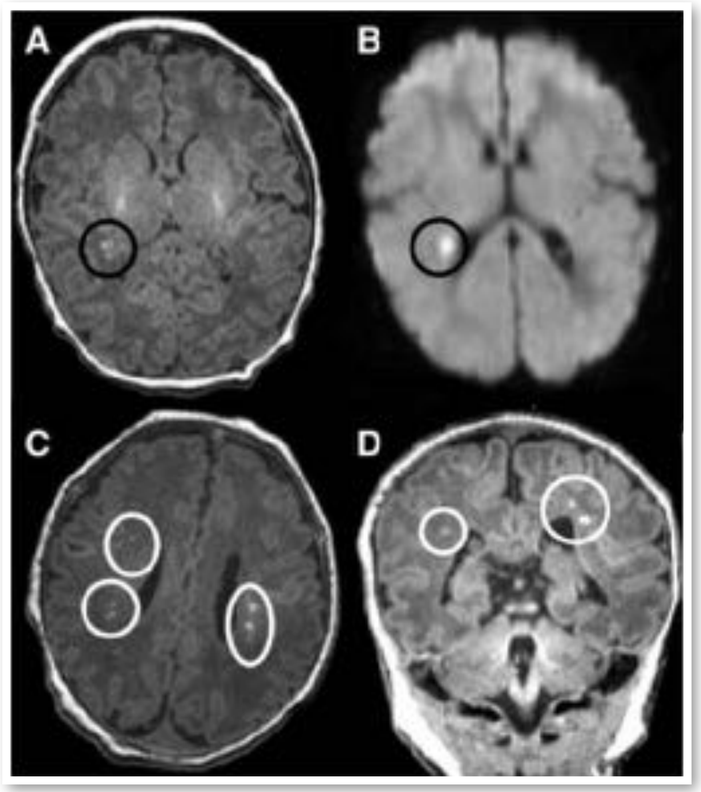


Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

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

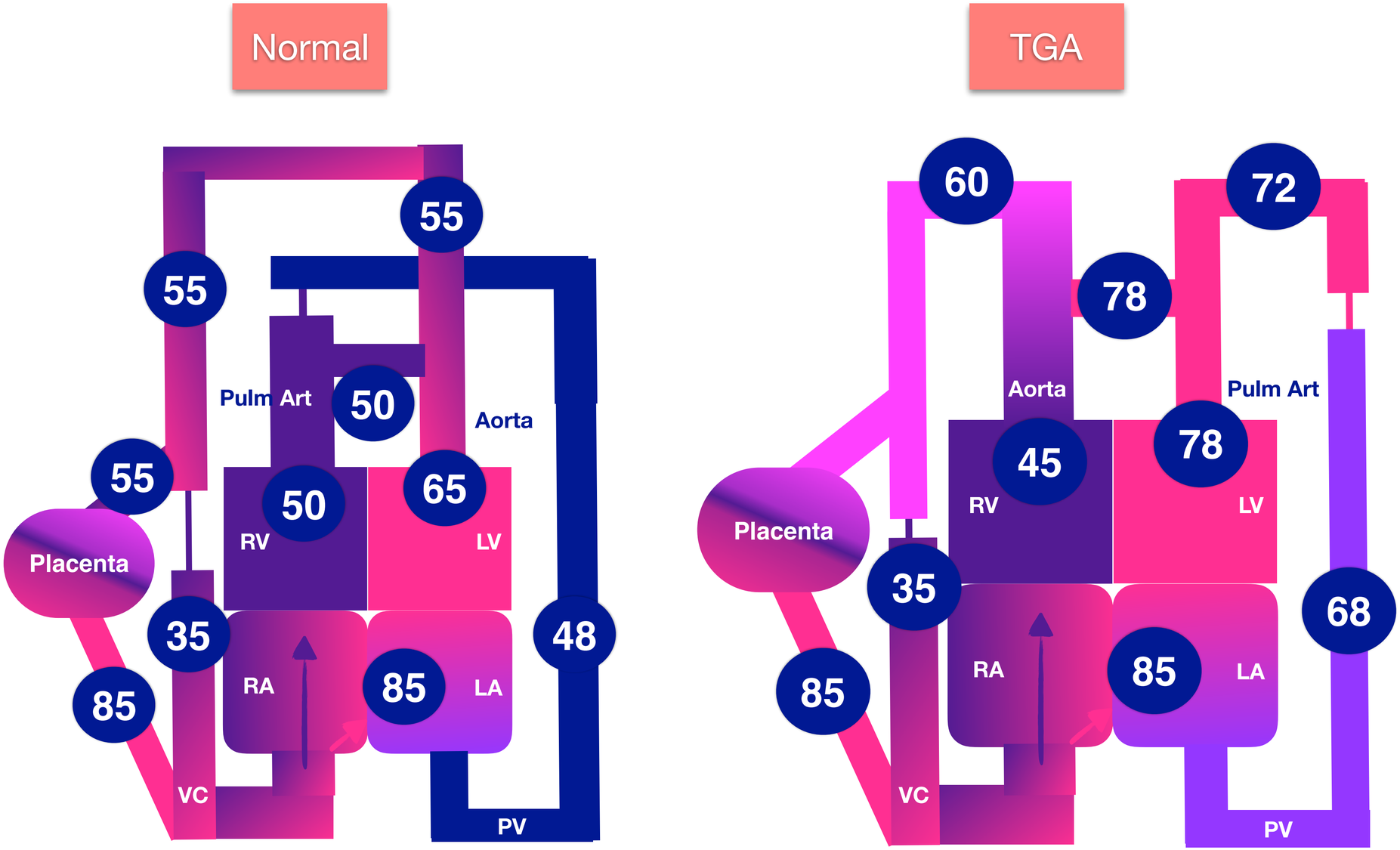
- **White matter lesions in** 30 to 40% of newborns with TGA (Miller et al., 2004; Licht et al., 2009)

- 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*

Morphological and physiological consequences of the fetal circulation in TGA



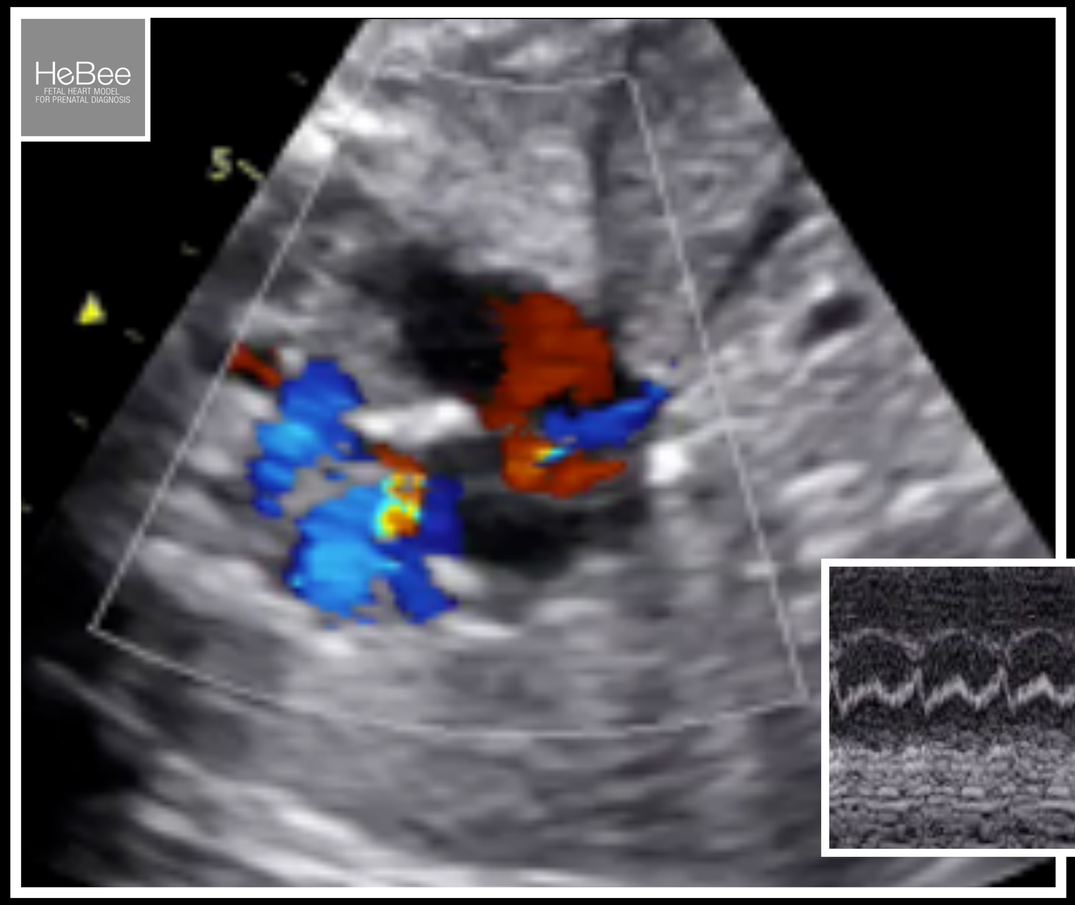
Rudolph A. Ped Res 2007;61:375-80
Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

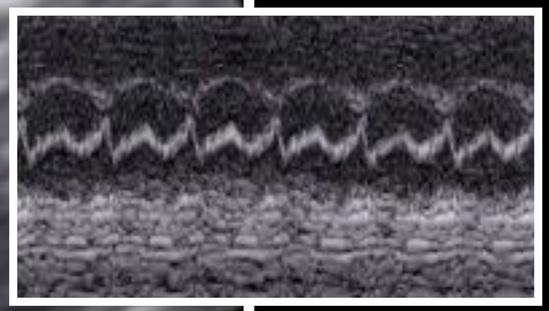
TGA

Foramen Ovale

TGA

Foramen Ovale



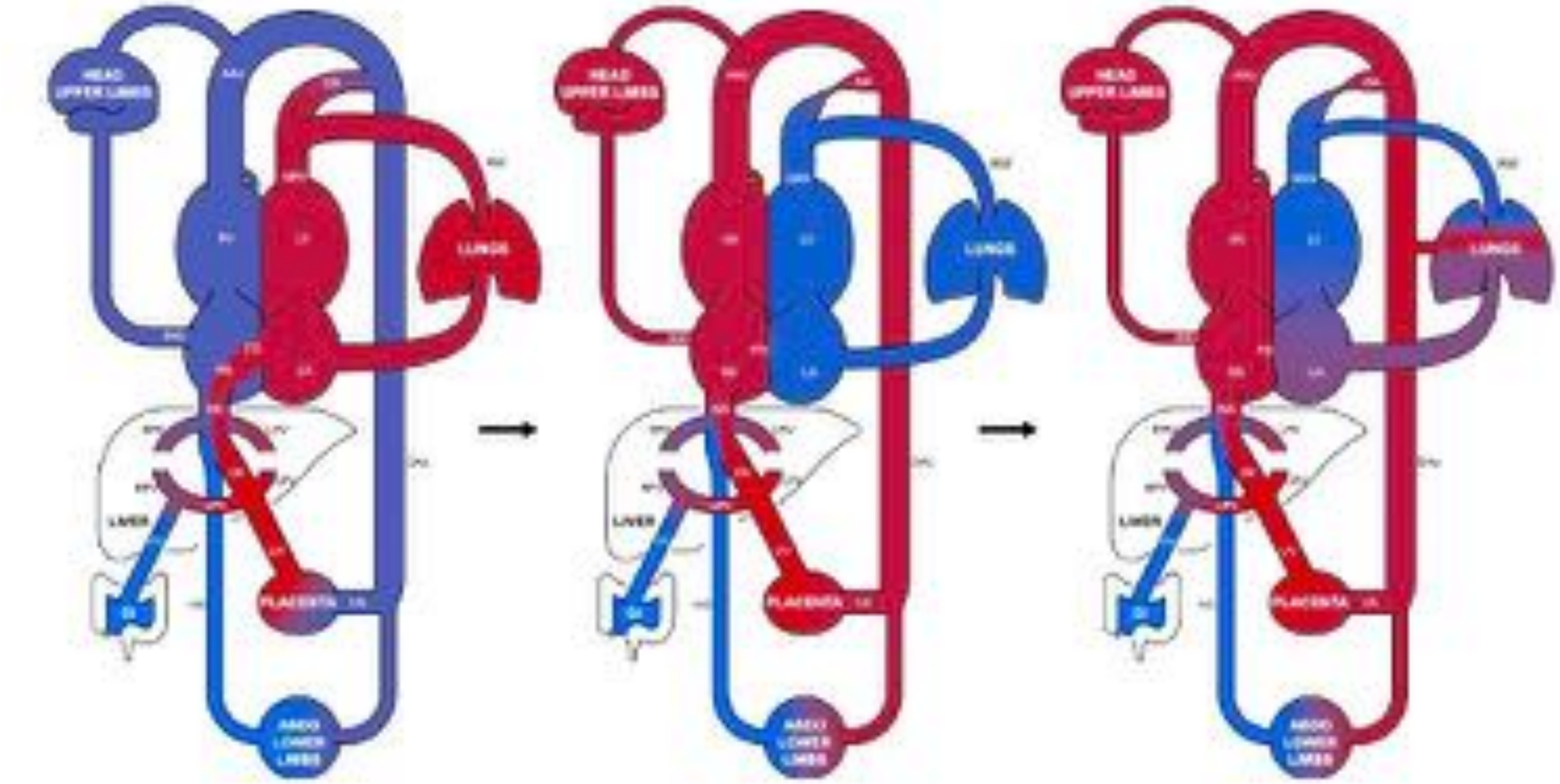


-Floppy Mb
-Small FO
-Small septal length
-pulmonary veins velocity > 0,41 m/s

TGA

Arterial duct

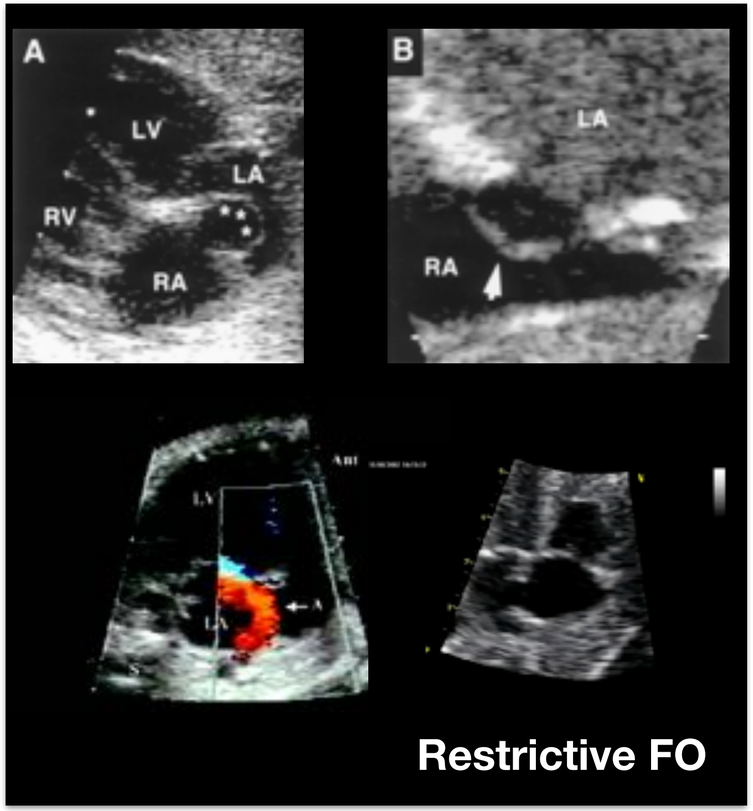
Morphological and physiological consequences of the fetal circulation in TGA

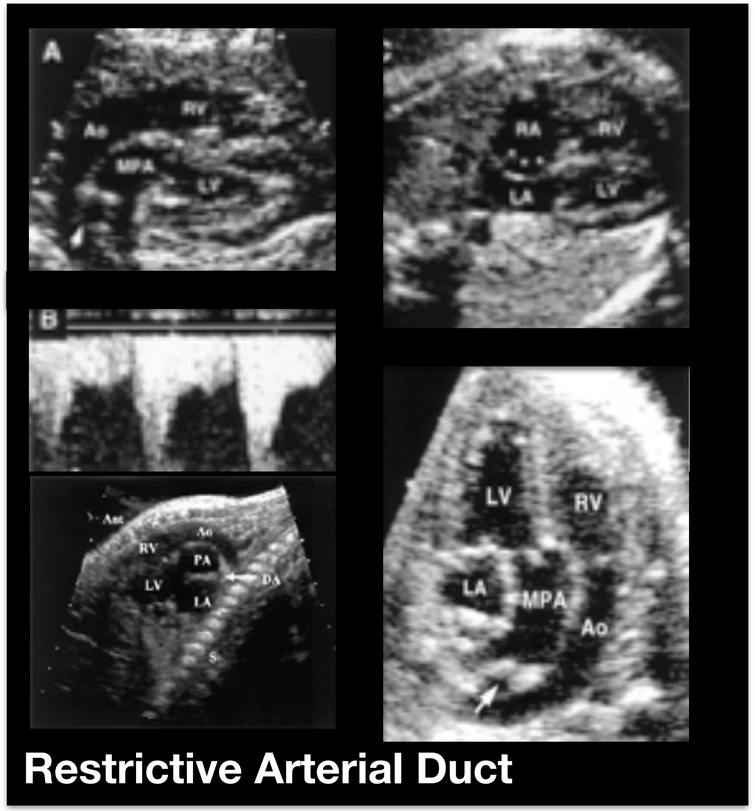


Initial increase in PBF due to vasodilatation with increased oxygen
Increased pulmonary venous return
Reduced size of the FO
Ductal constriction due to oxygen
Isolation of Pulmonary circulation
Increased PVR
Development of aorta-pulmonary collaterals

Prenatal diagnosis of transposition of the great arteries

Prevention of early neonatal demise





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

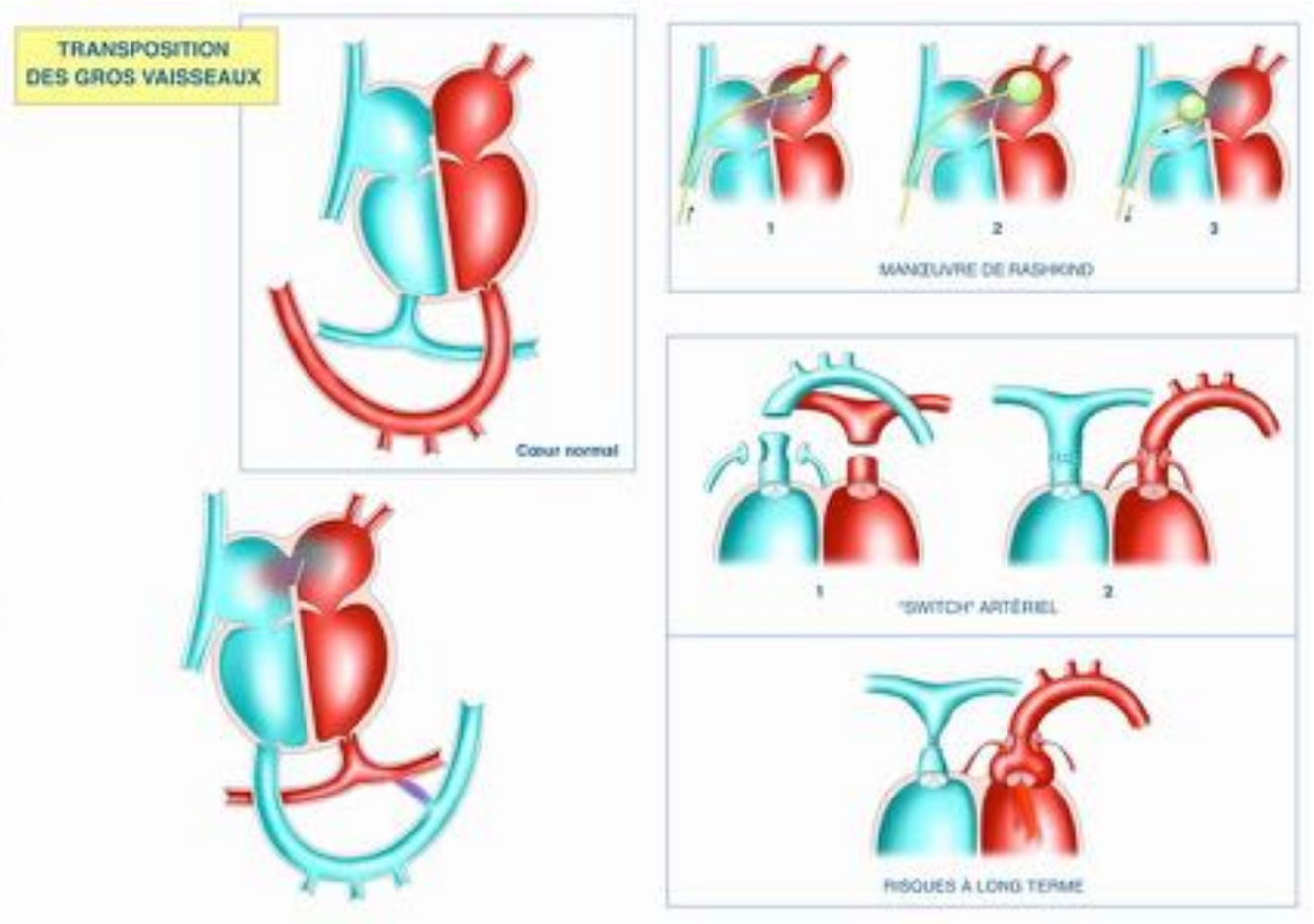
Paris Registry of Congenital Malformations, 1983-2000

	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

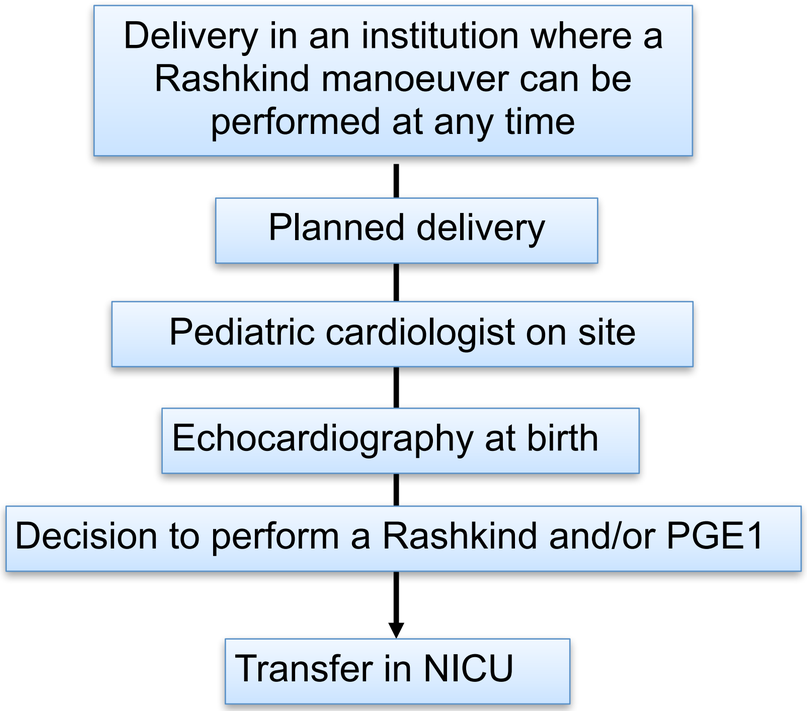
- Organisation of foetal cardiac growth surveillance
  - Foramen ovale and arterial duct
- *In utero* transfer organisation
- Organisation of perinatal management
- Prevention of early neonatal demise
- Prepare the parents to the future even
- Post-natal management and follow-up



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.

Prenatal diagnosis of transposition of the great arteries

Perinatal organization



From 1992 to 2017

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)

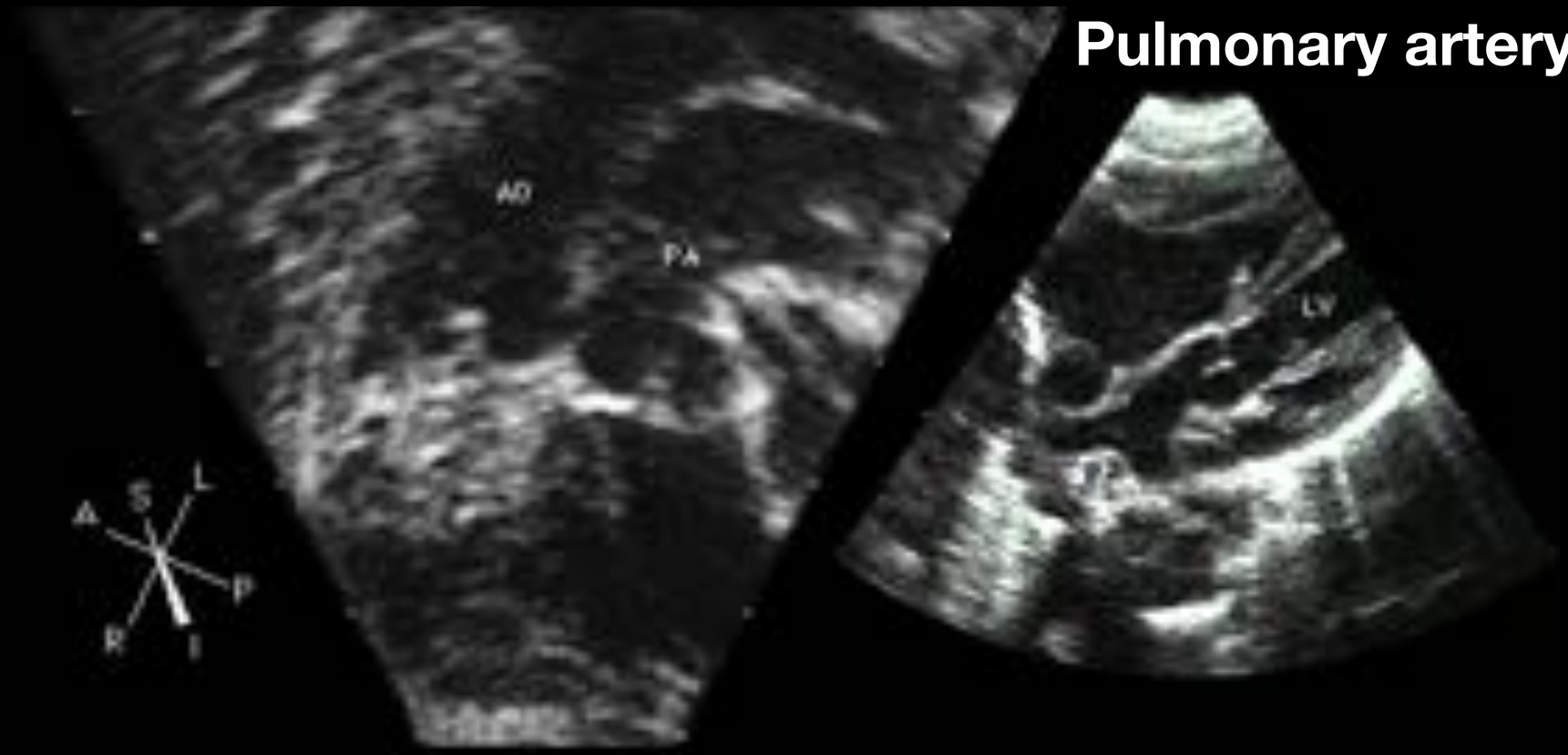
Surgical mortality 1.7 % : 693 survivors at discharge

Neonatal diagnosis of TGA
Isolated cyanosis

TGA

Rapid diagnosis

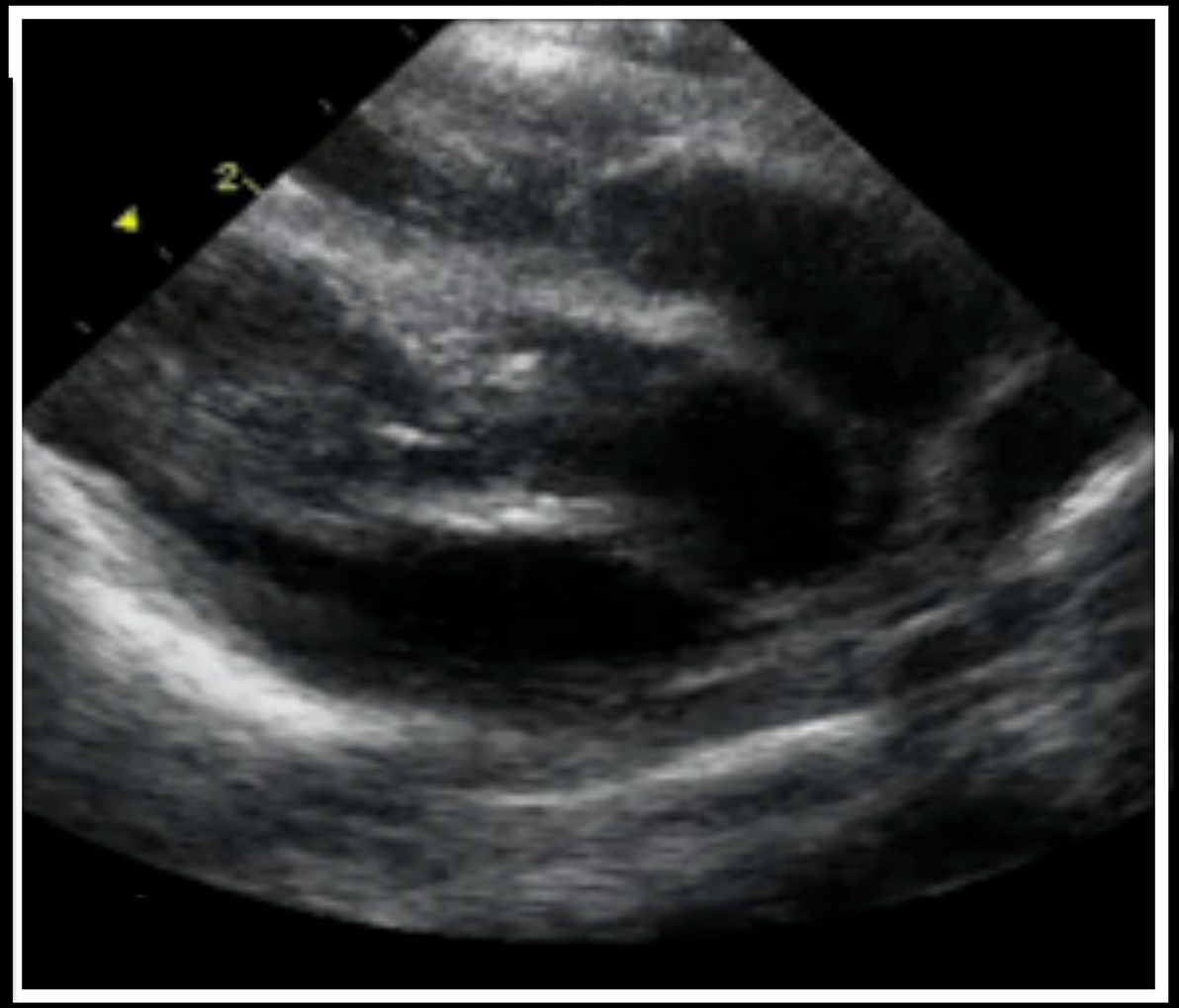
Parallel course of great vessels



Pulmonary artery from LV

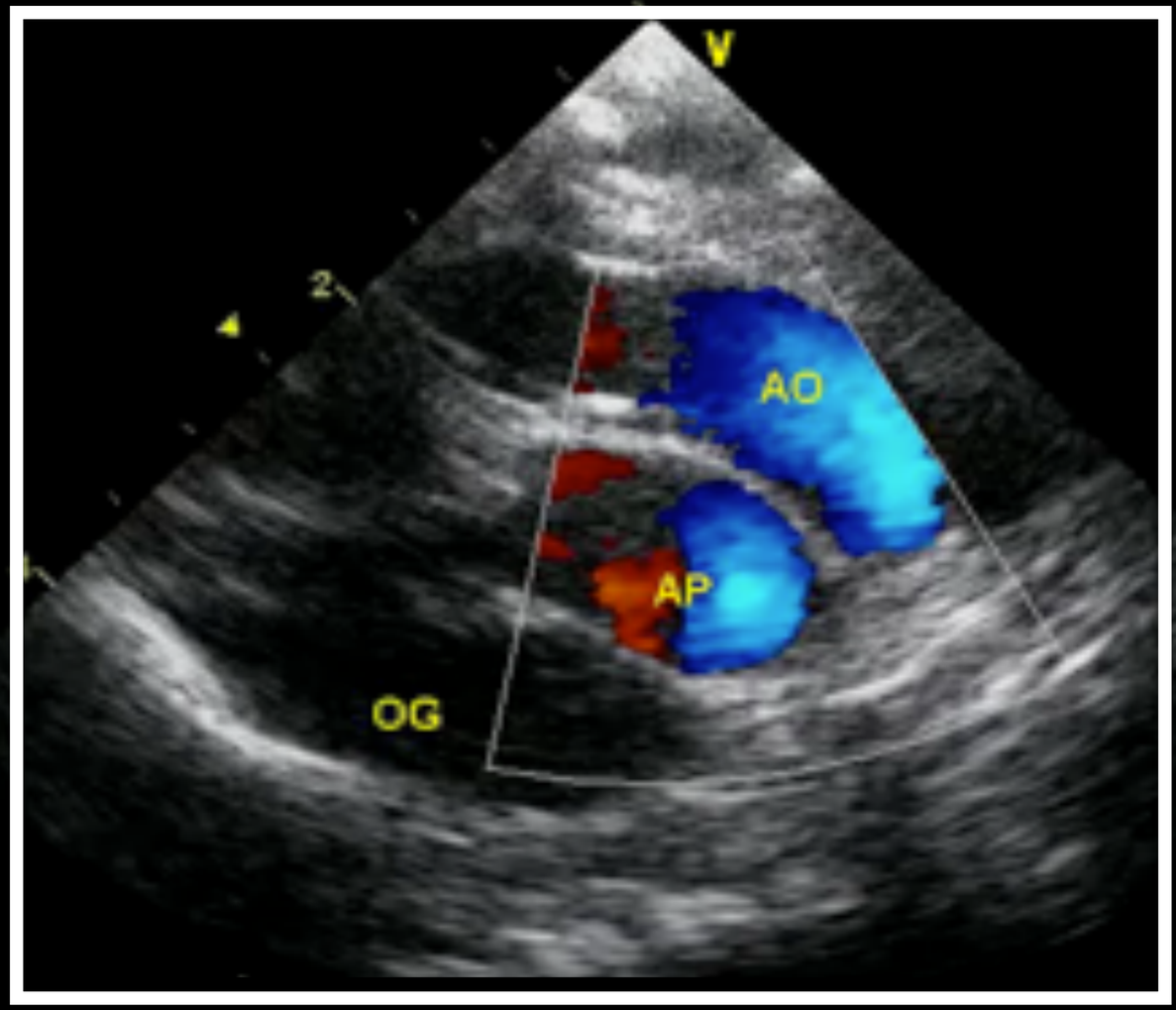
TGA

Rapid diagnosis



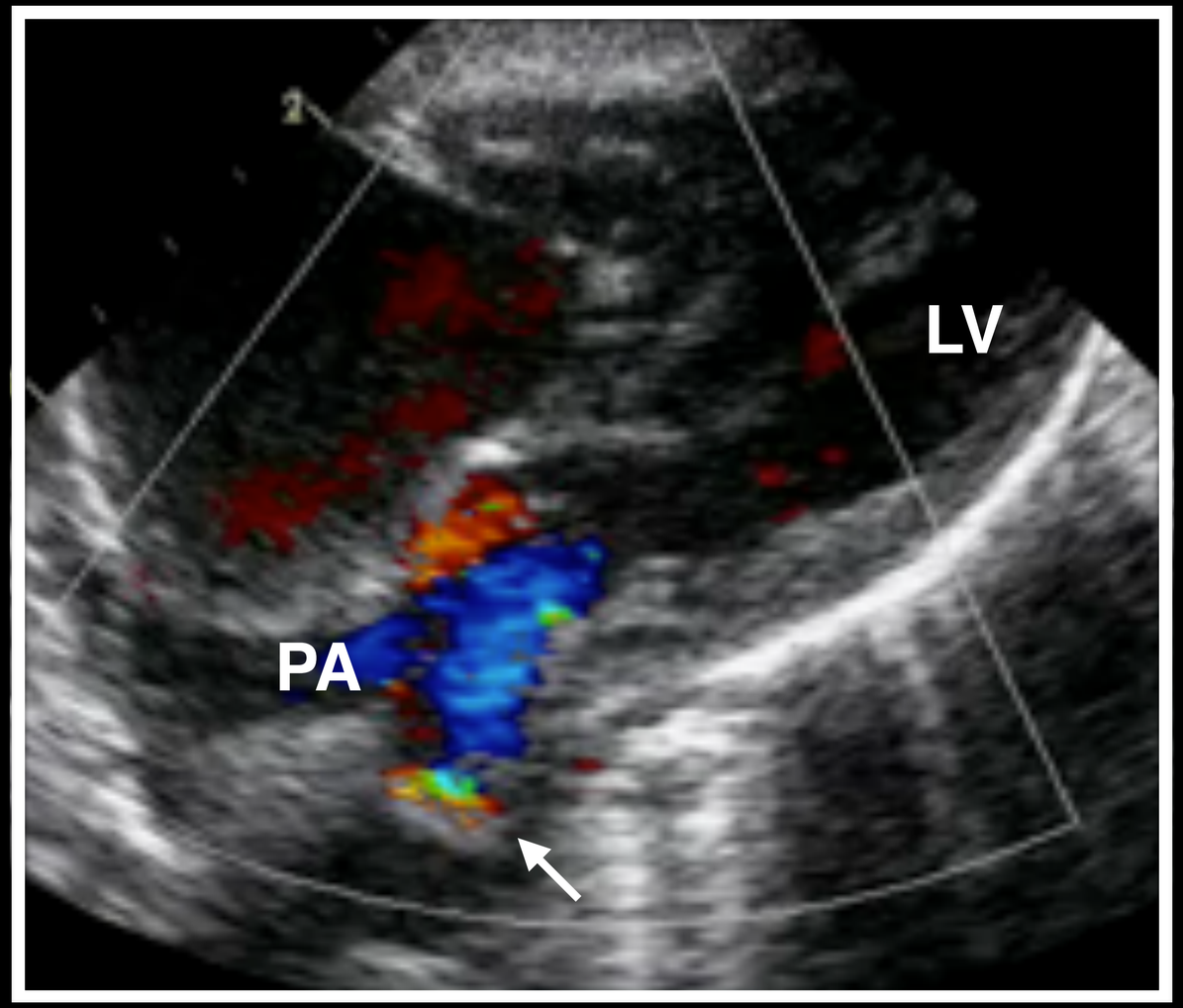
TGA

Rapid diagnosis



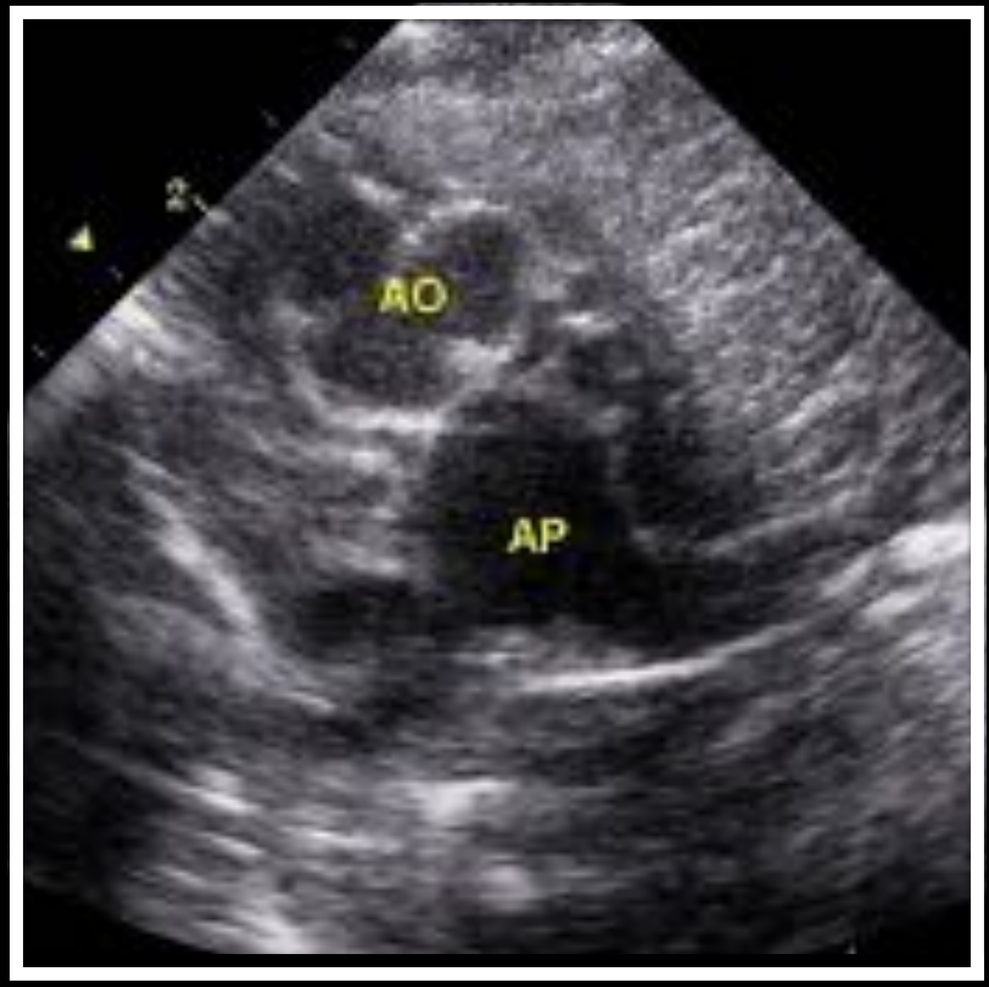
TGA

Rapid diagnosis



TGA

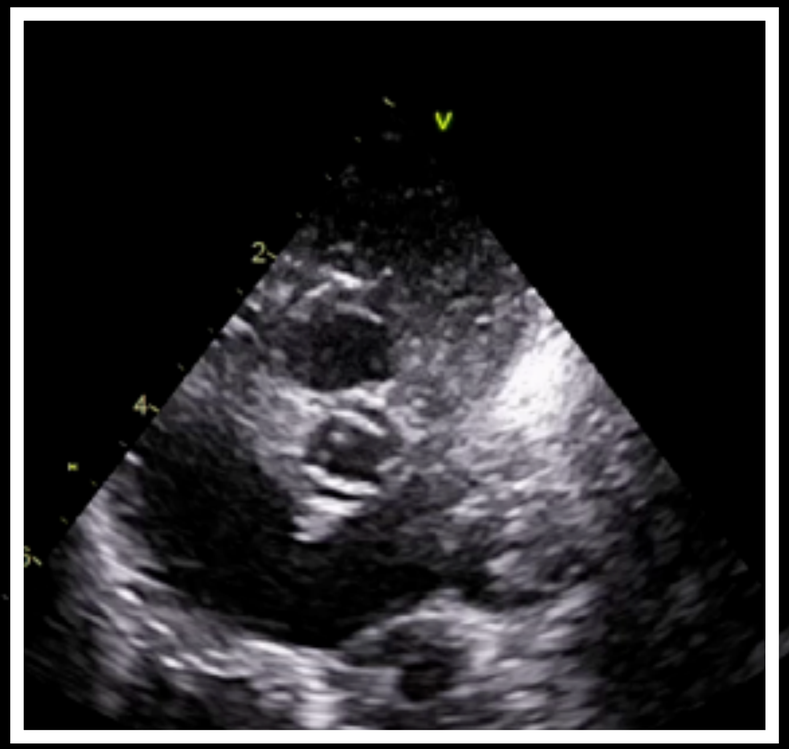
D-TGA

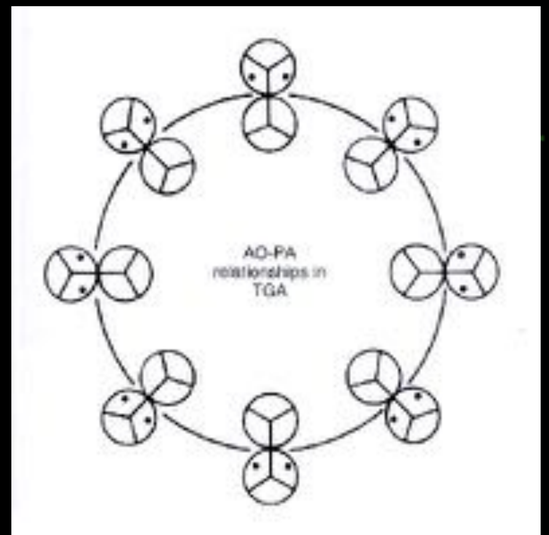


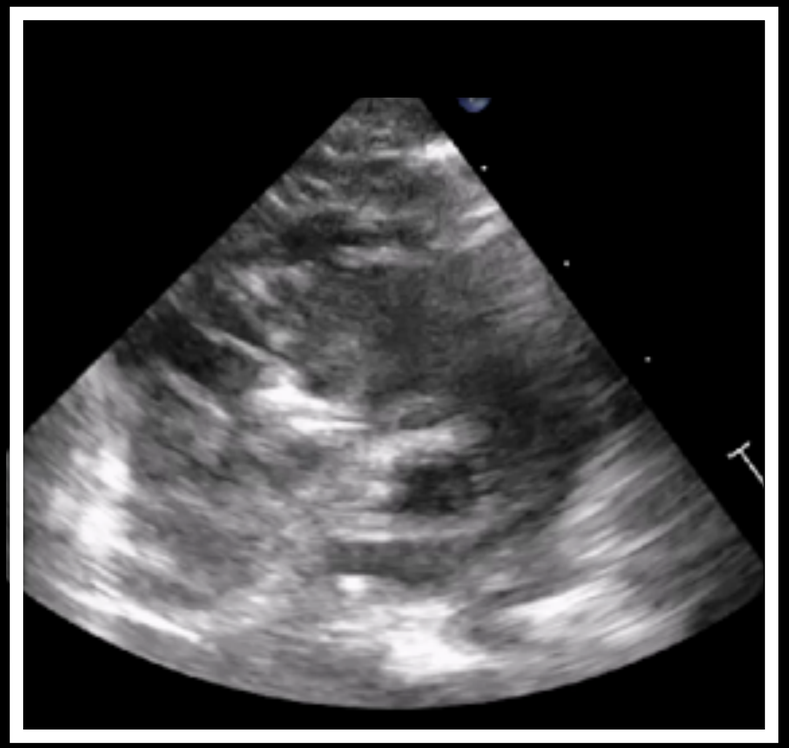
D-TGA

TGA

Aorto pulmonary vessels relationship and discrepancy



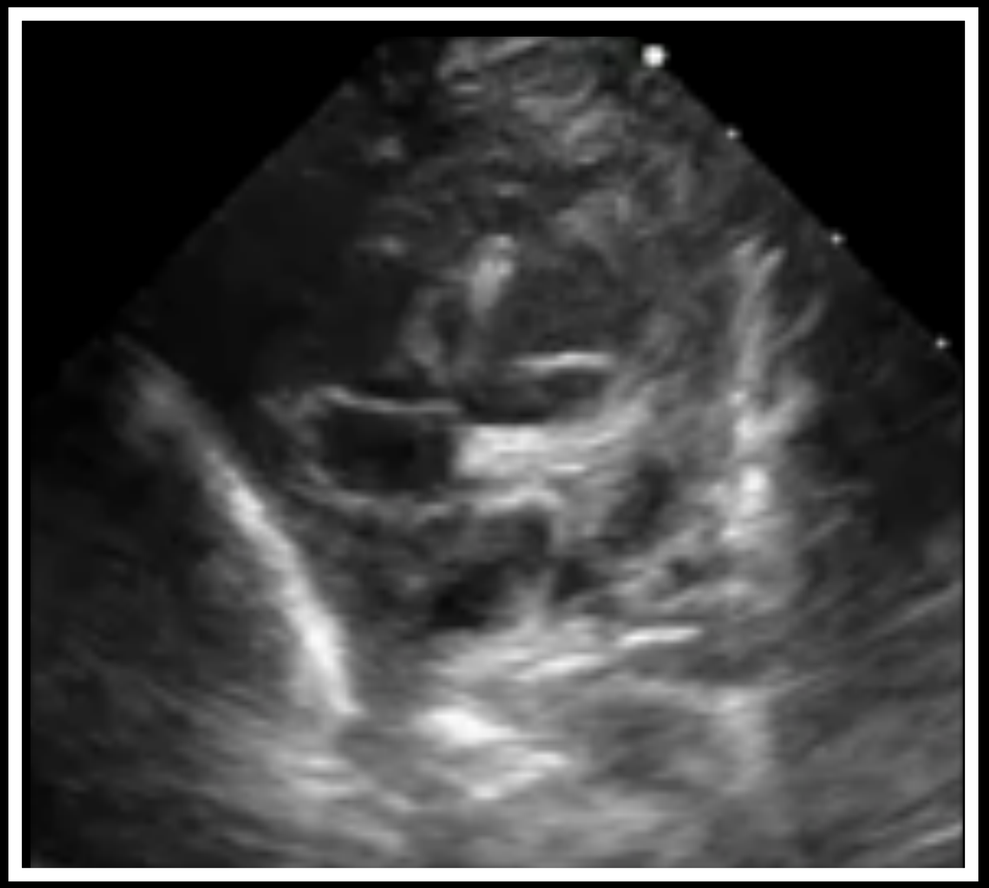




TGA

Relative position of the great vessels

L-TGA

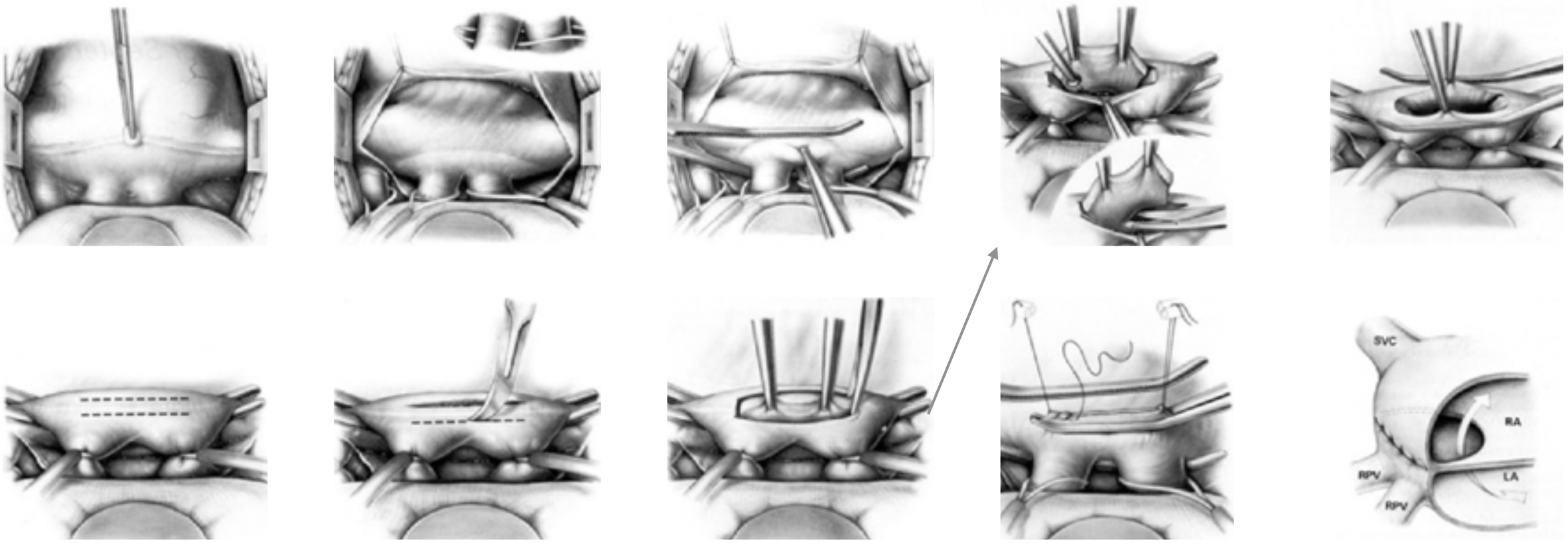


D-TGA with posterior aorta

TGA with heart failure and restrictive PFO

Alfred Blalock

Blalock-Hanlon procedure



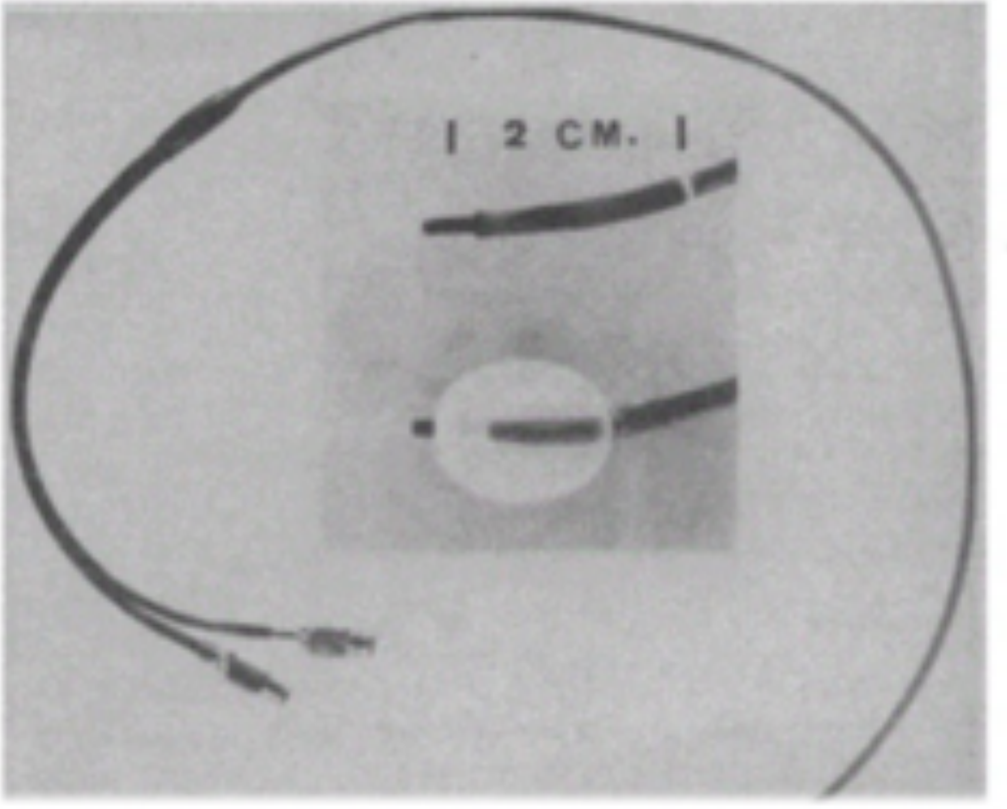
1950

Creation of an Atrial Septal Defect Without Thoracotomy

A Palliative Approach to Complete Transposition of the Great Arteries

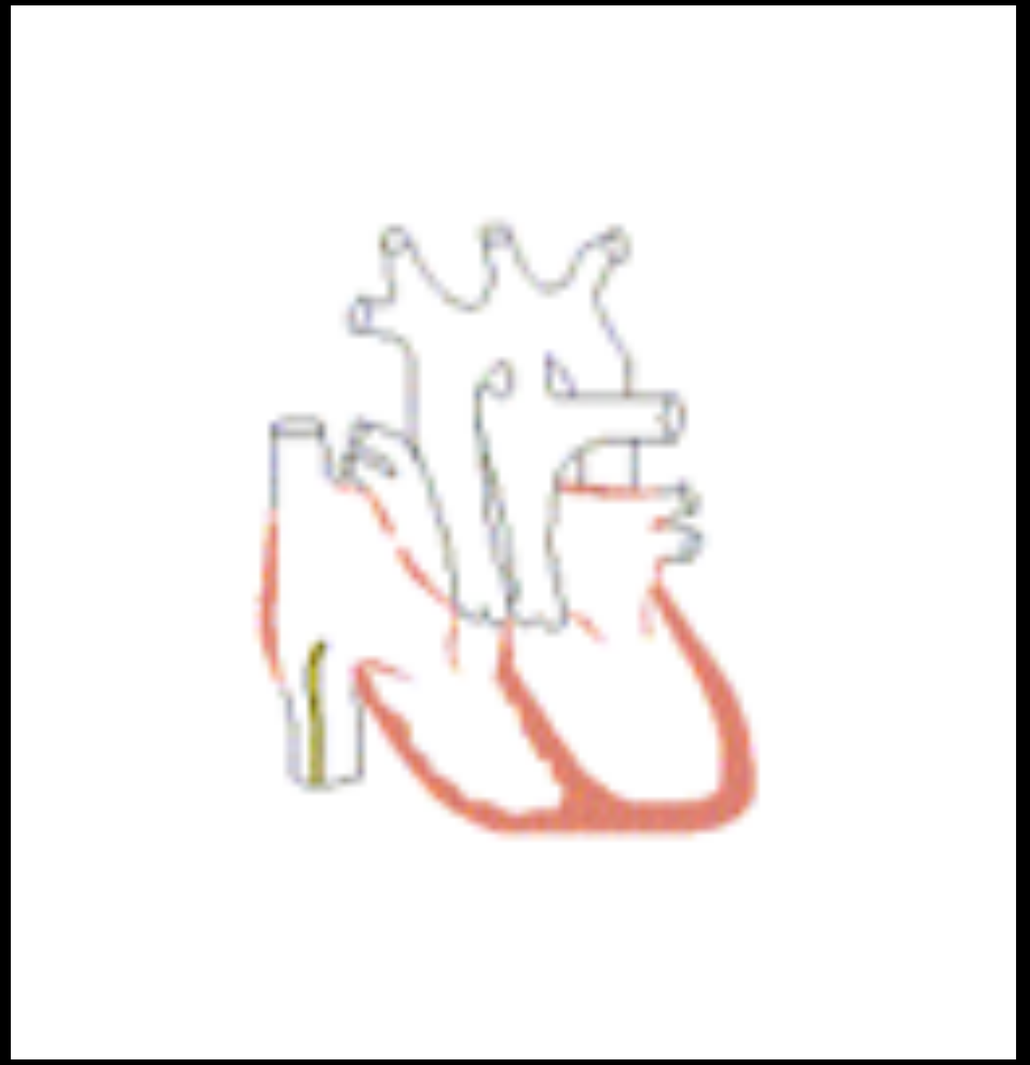
William J. Rashkind, MD, and William W. Miller, MD

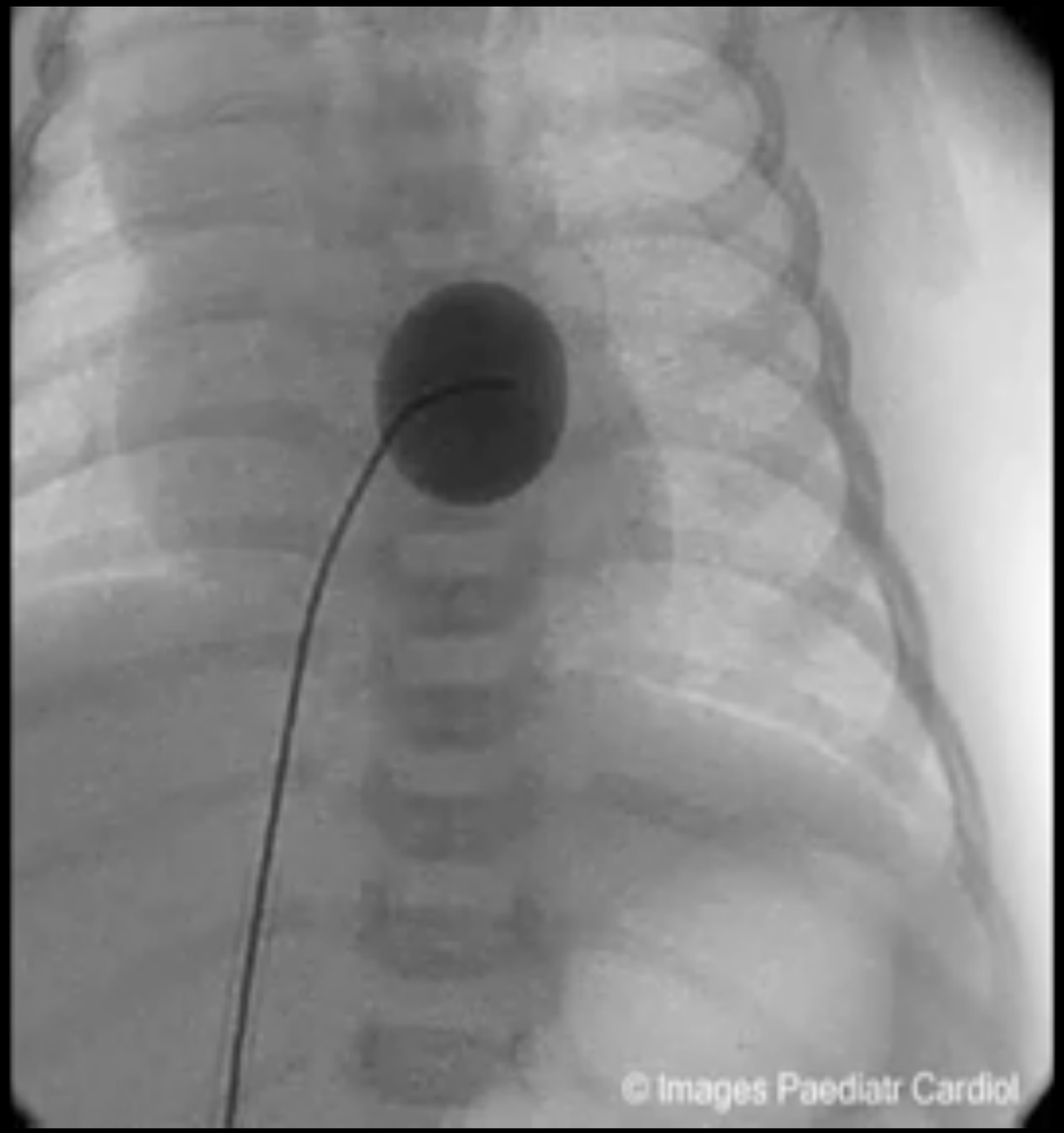
Transposition of the great vessels (TGV) occurs in approximately 20% of children who die with congenital heart disease.[1] With rare exceptions, patients with this lesion die in the first 6 months of life (50% within the first month). Approximately 40% of patients with TGV have an otherwise normal heart.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



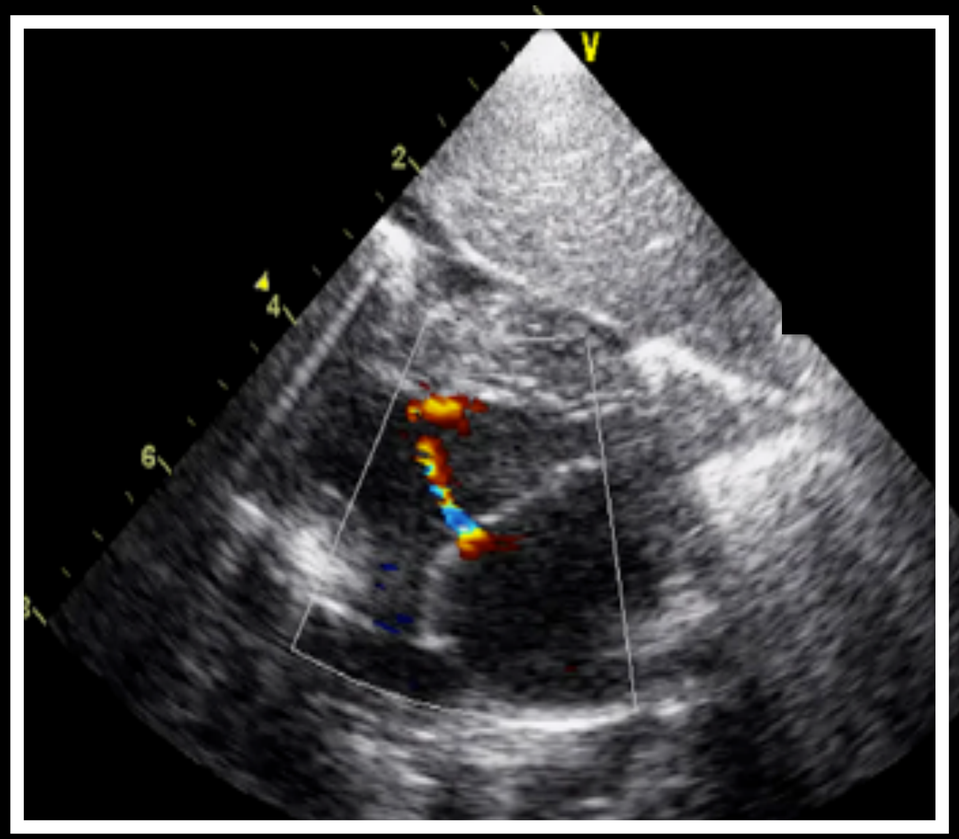
Rashkind WJ and Miller WW. JAMA 1966

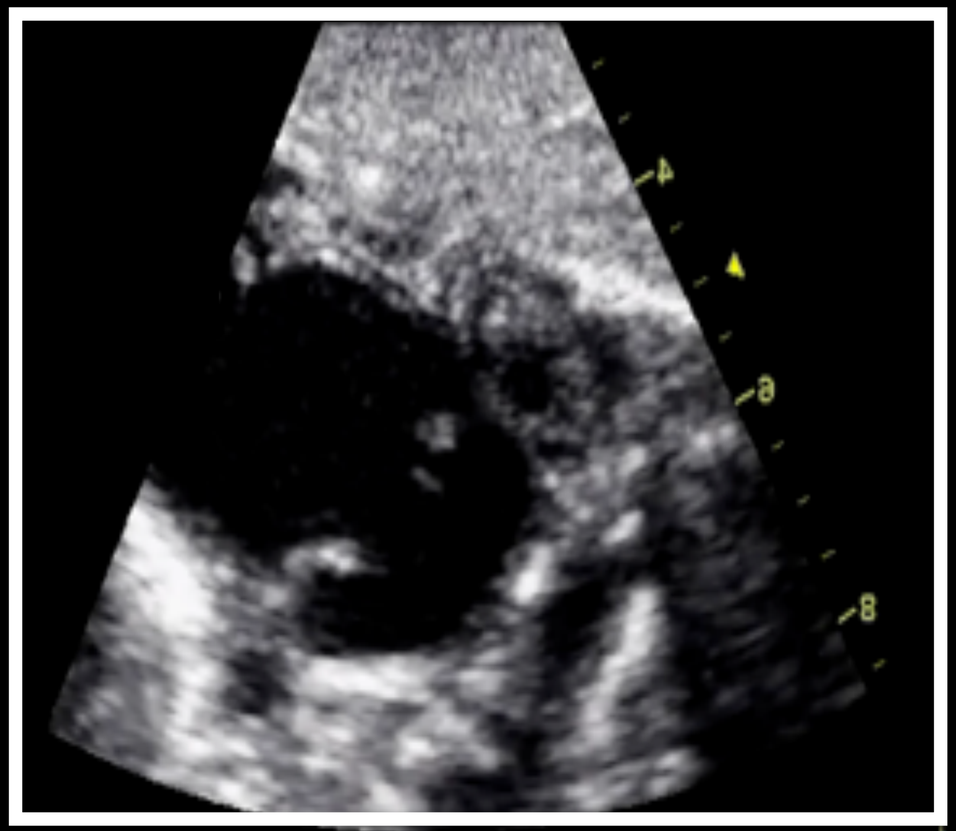
William Rashkind



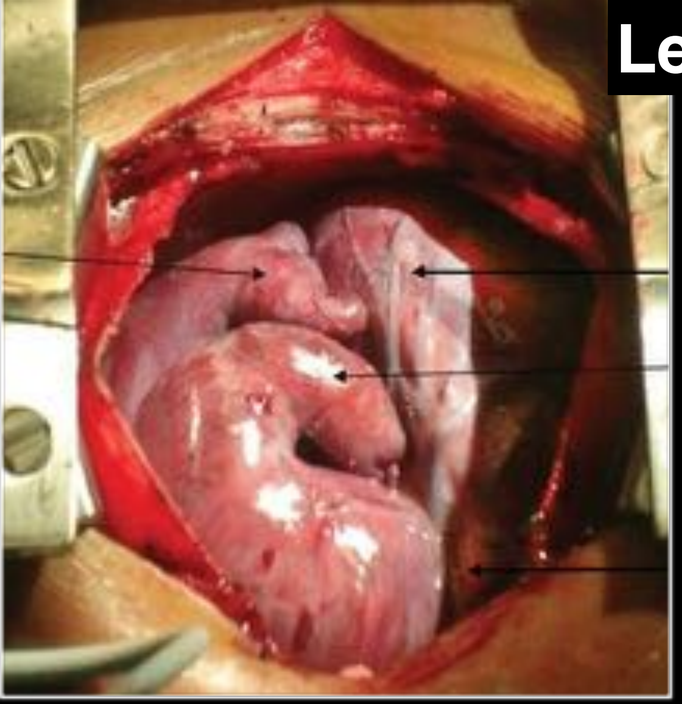

© Images Paediatr Cardiol

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

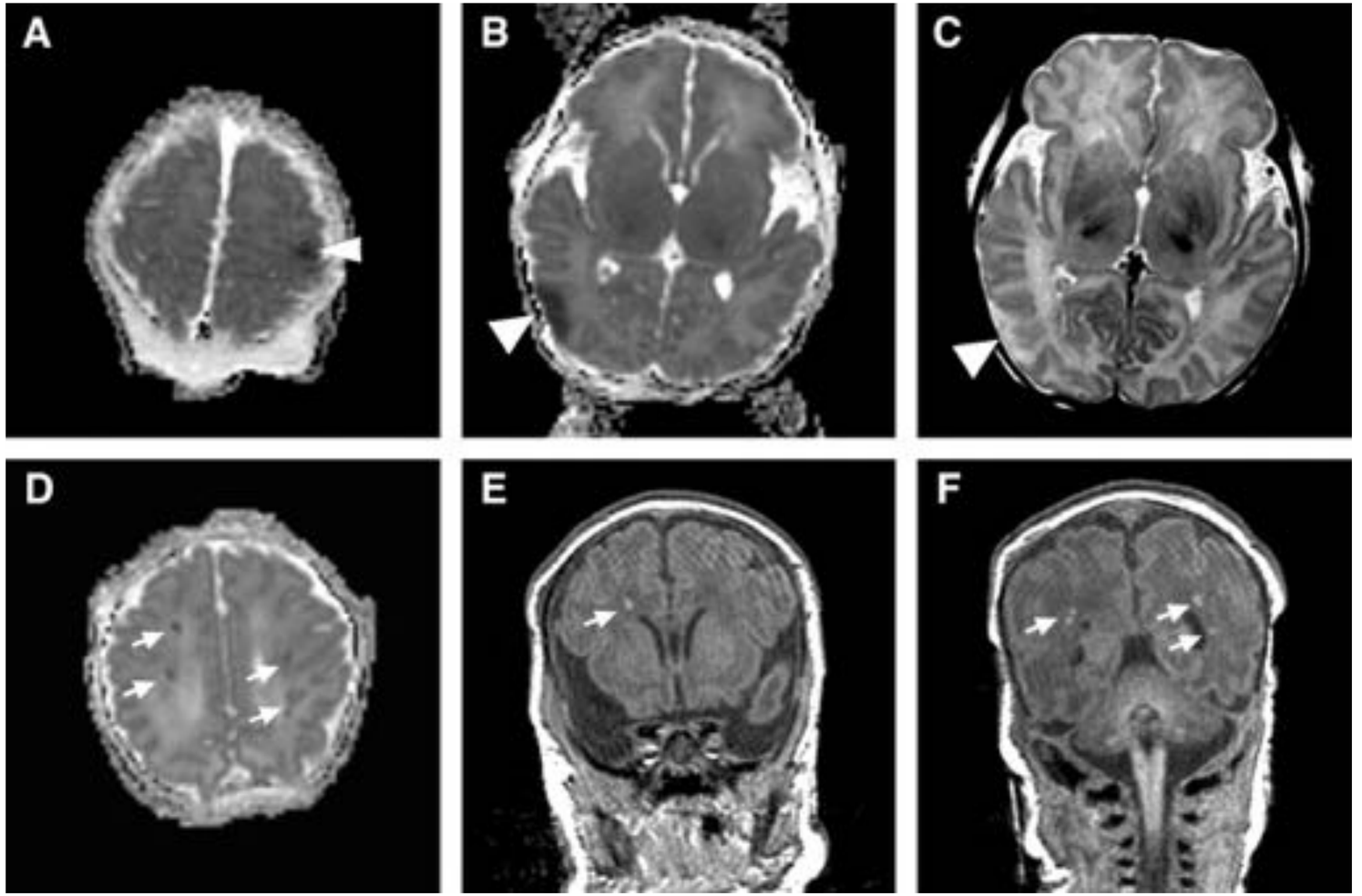
Soit environ 120 transpositions des gros vaisseaux / an

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.

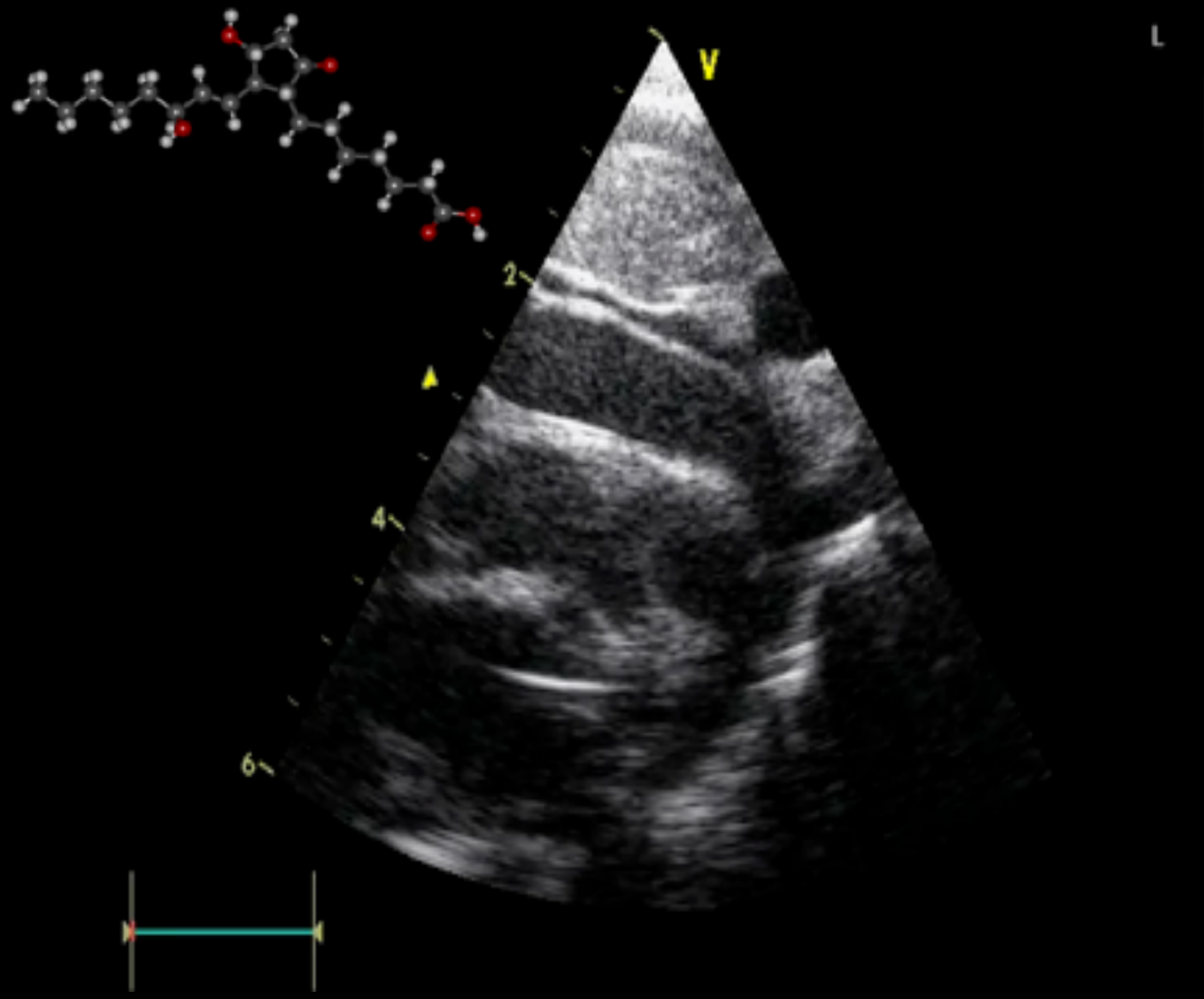
McQuillen P S et al. Circulation 2006;113:280-285

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;

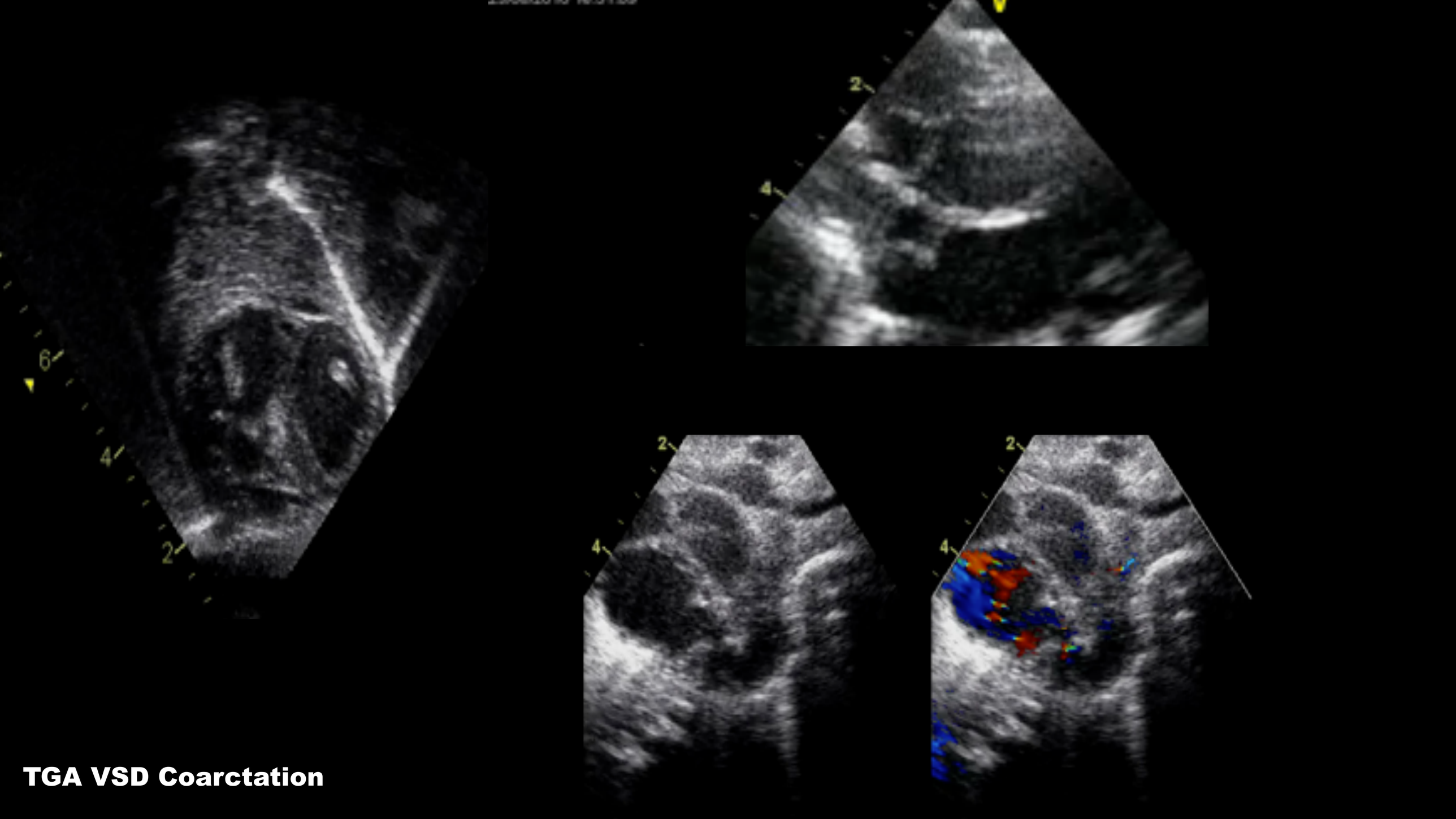
TABLE 5 Multivariate Analysis of Major Transport Complications

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	

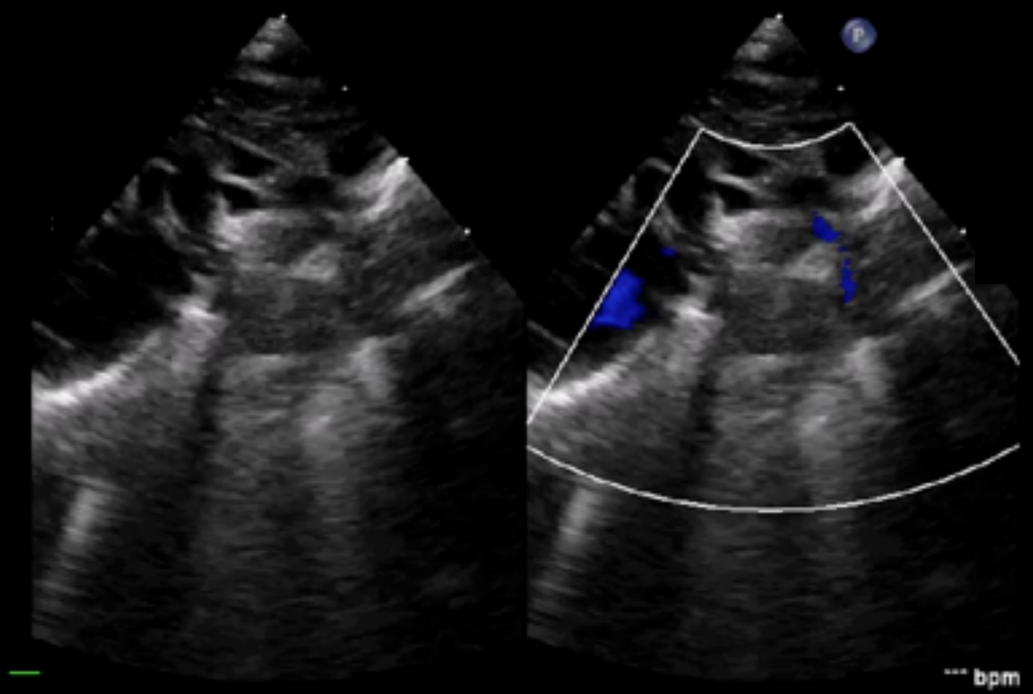
EGA indicates estimated gestational age; NS, not significant.

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

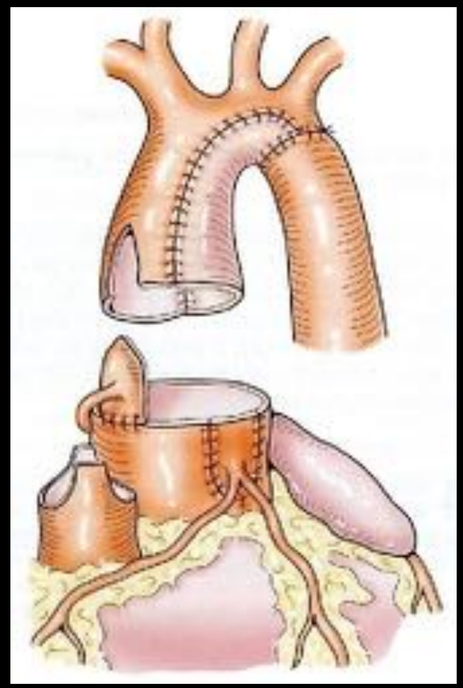

TGA VSD Coarctation

Aortic arch hypoplasia and coarctation

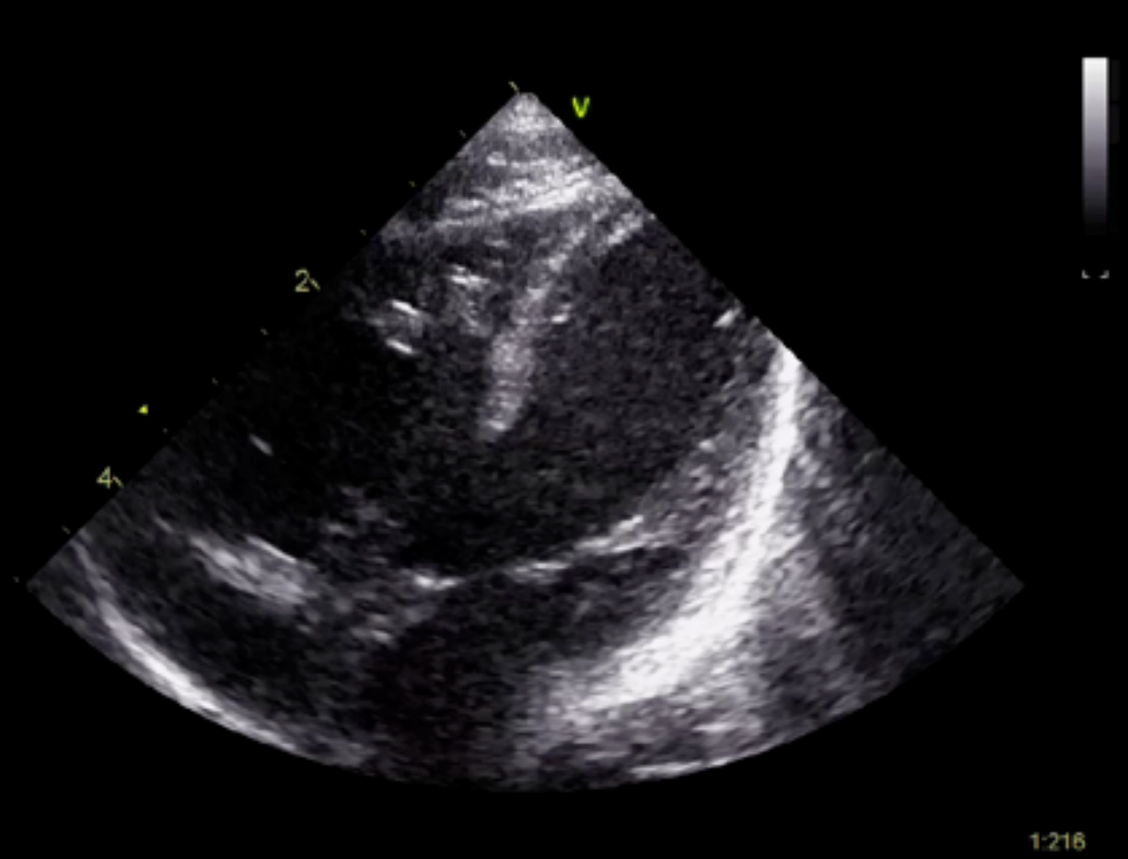


Localization and extension of the narrowed portion

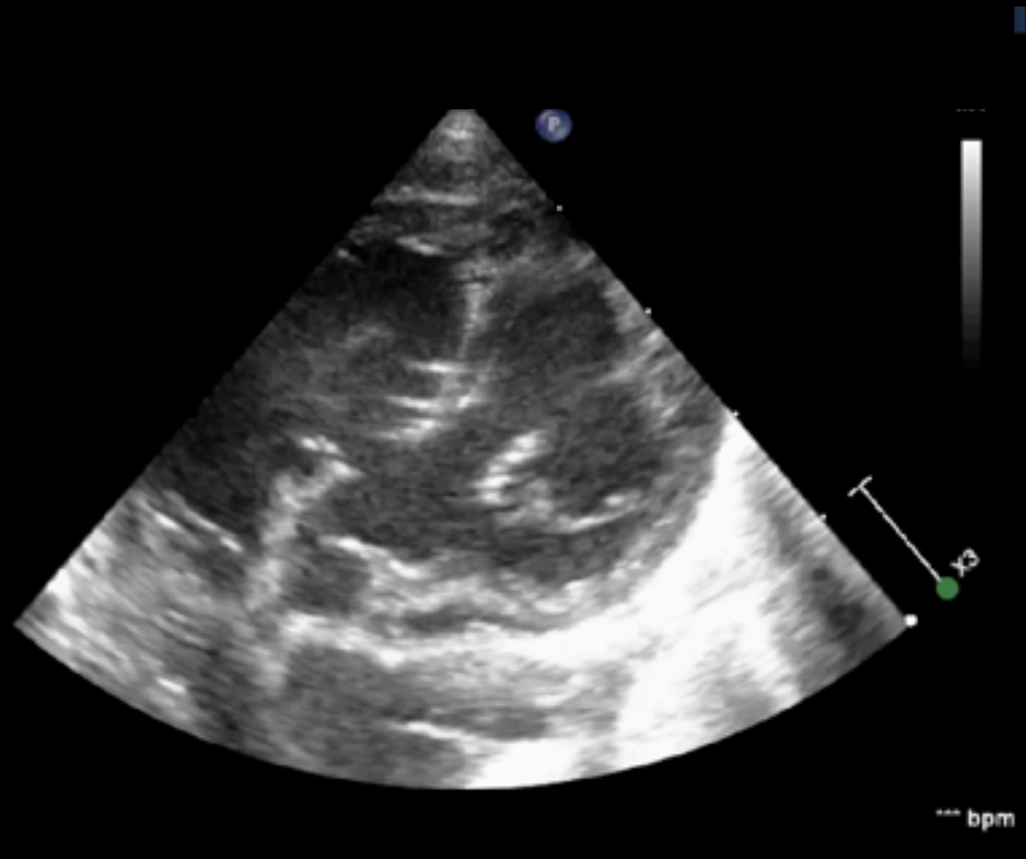
2 techniques: enlargement and extended end to end
=> Discrepancy between aortic and pulmonary roots



VSD: localization and size

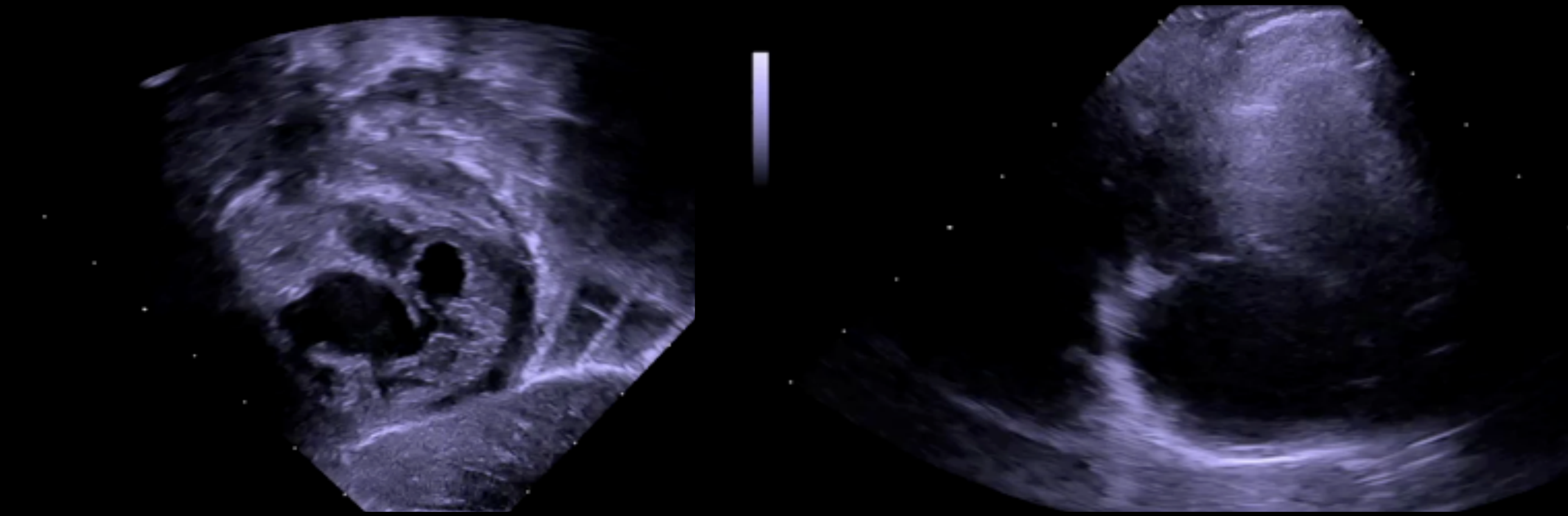


Inlet VSD



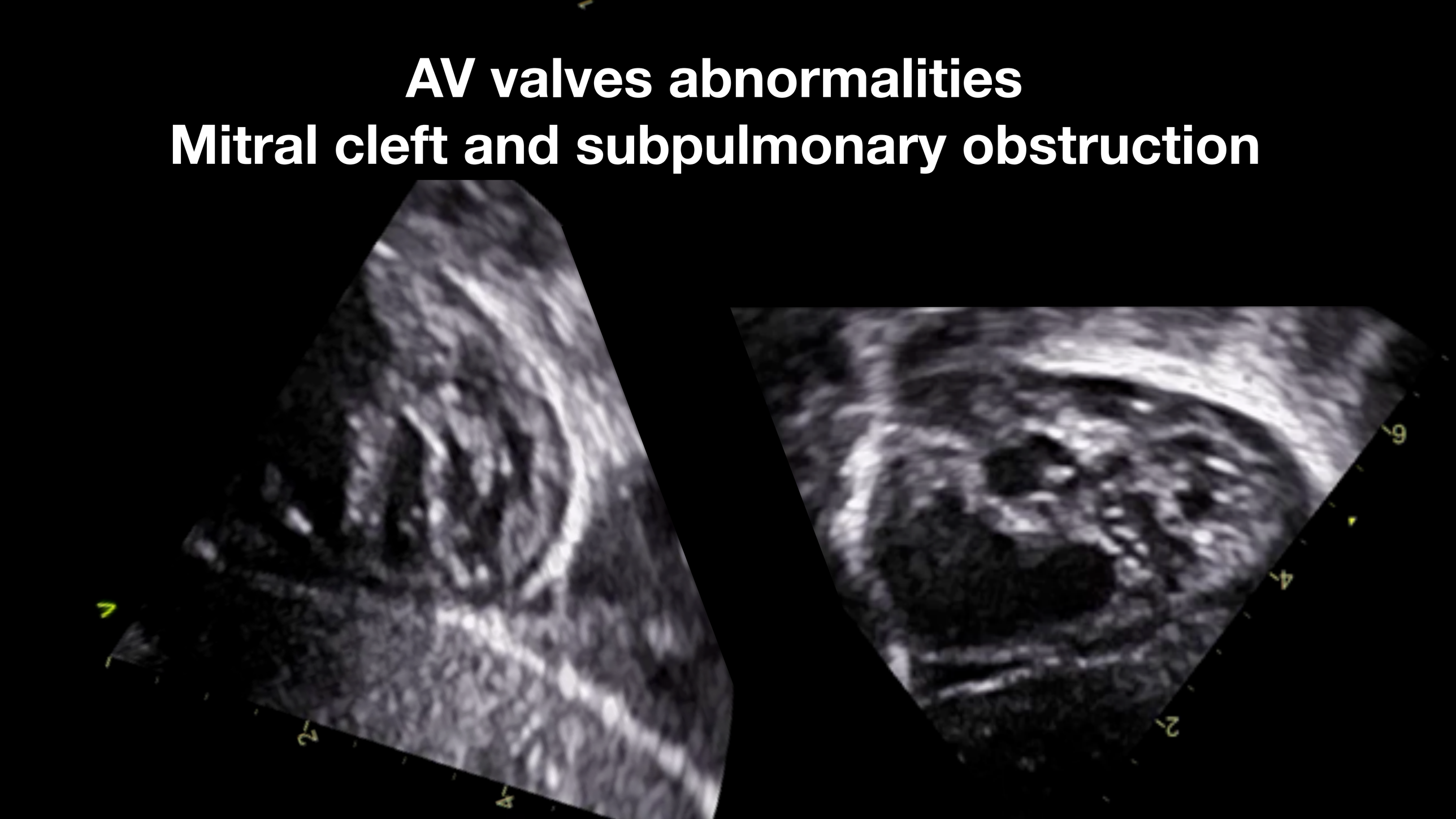
Outlet VSD

AV valves abnormalities
Straddling and over-riding



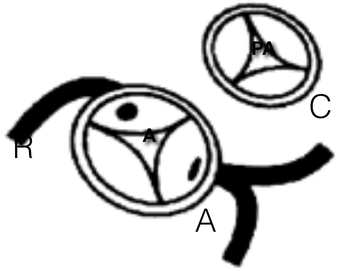
Straddling of tricuspid valve

AV valves abnormalities
Mitral cleft and subpulmonary obstruction

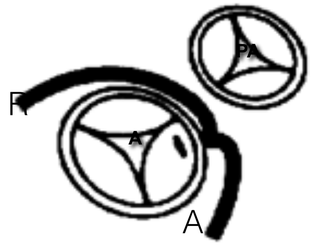


TGA left outflow tract obstruction

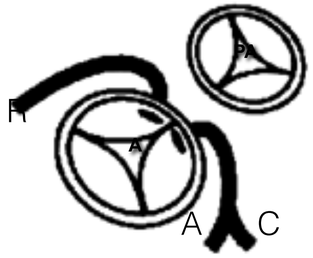
Type A



Type B

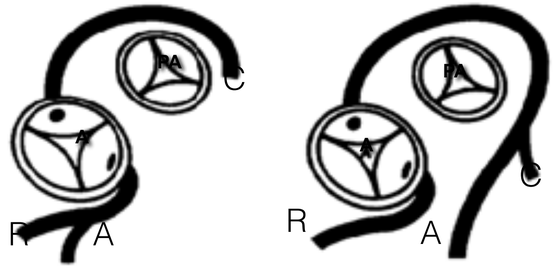


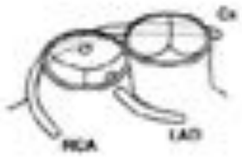
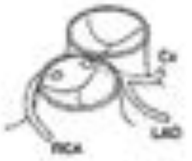
Type C

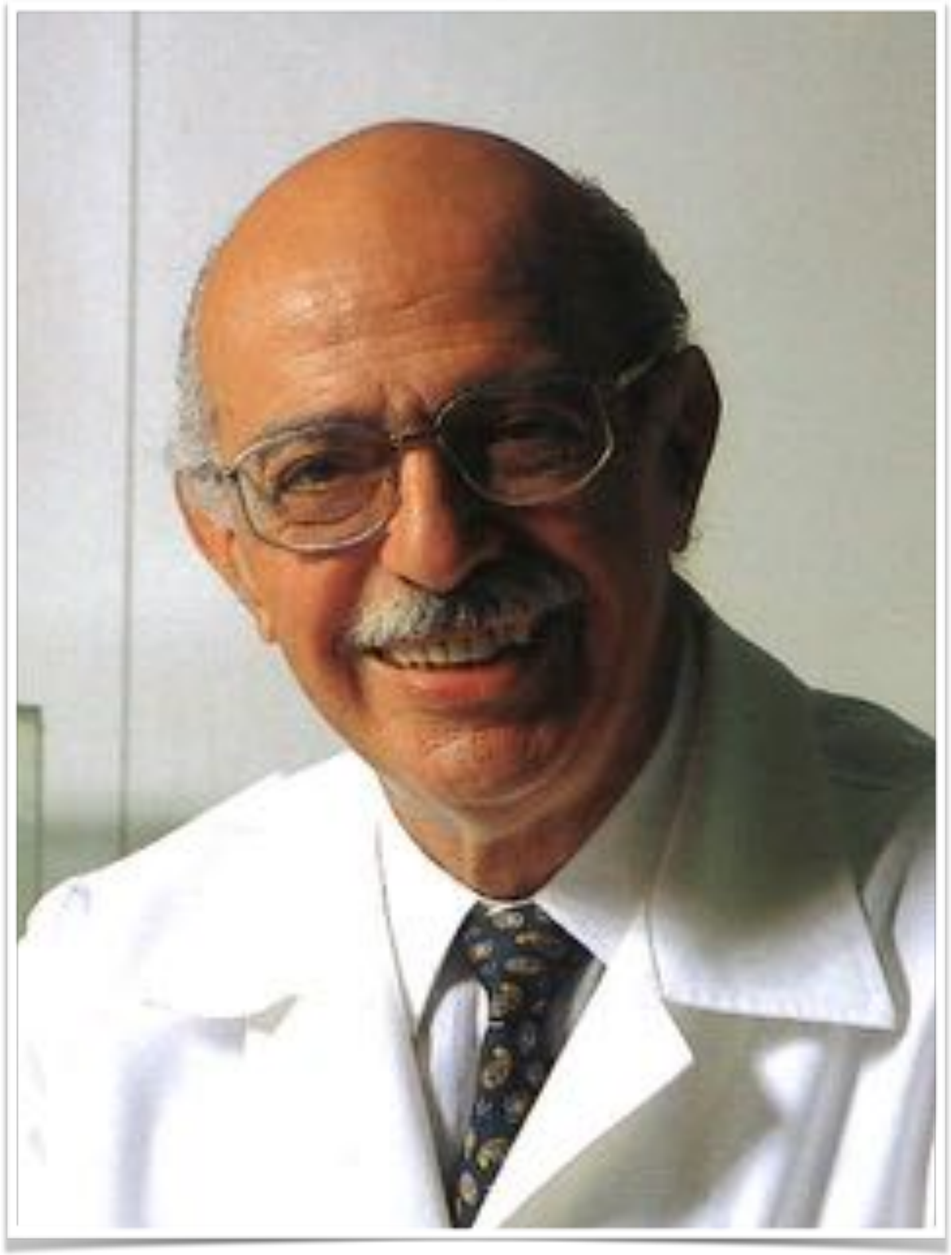


Type D

Type E

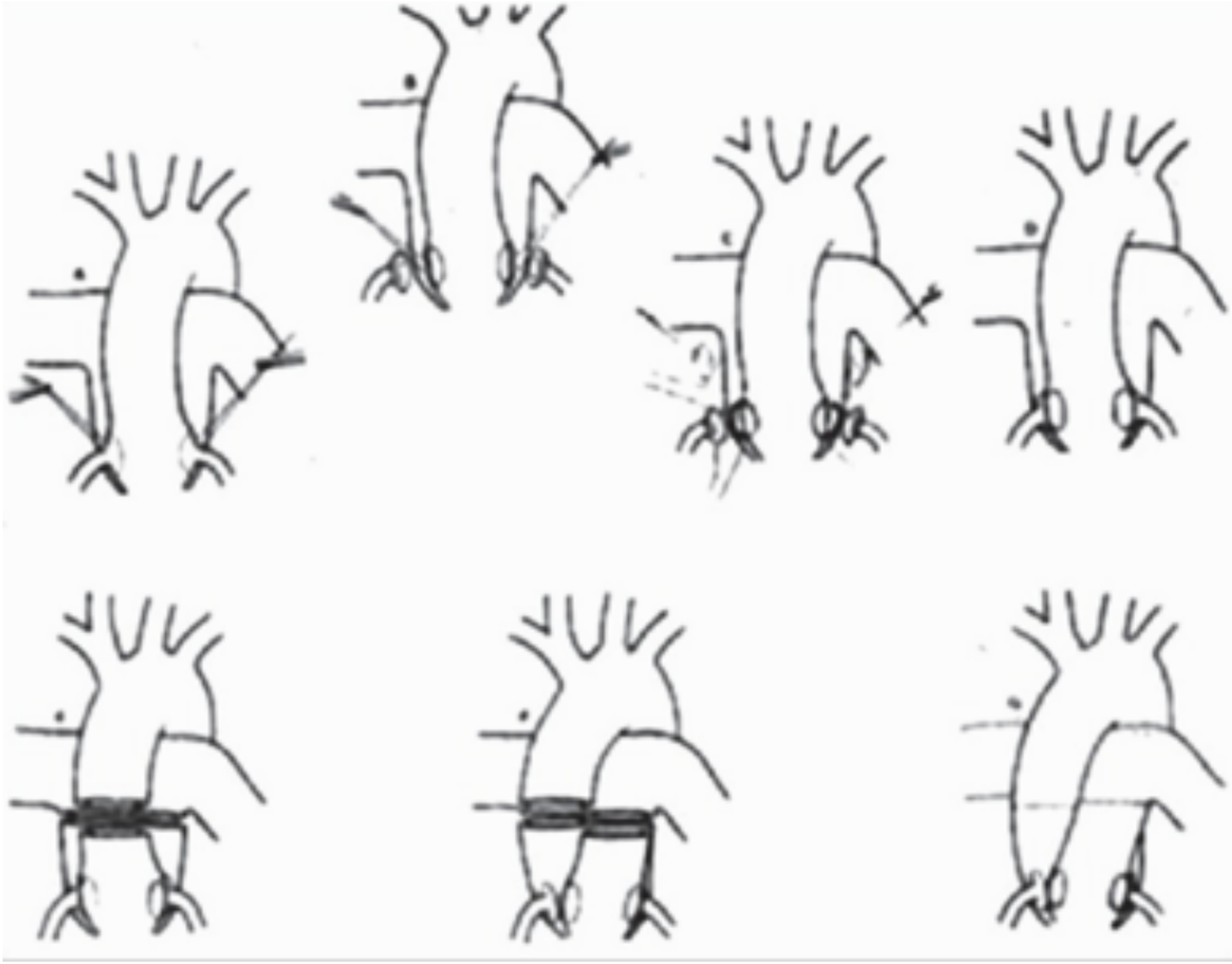


Habituel	Circonflexe de l'ACD	ACG unique	ACD unique	Inversée	ACD inversée et circonflexe	Coronaires Intramurales
						
66,9%	16,1%	1,7%	3,9%	2,4%	4,2%	3,2%



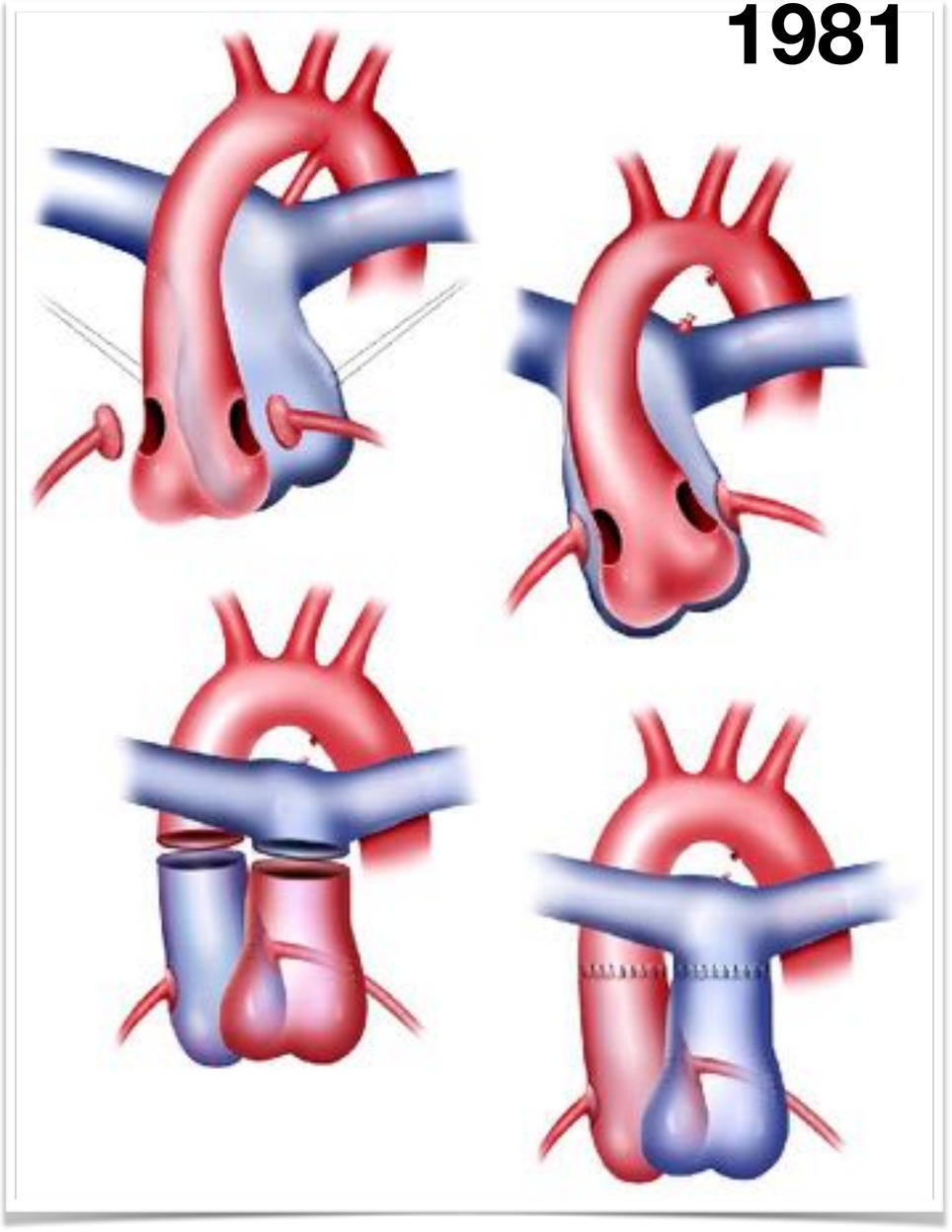
Adib Domingo Jatene

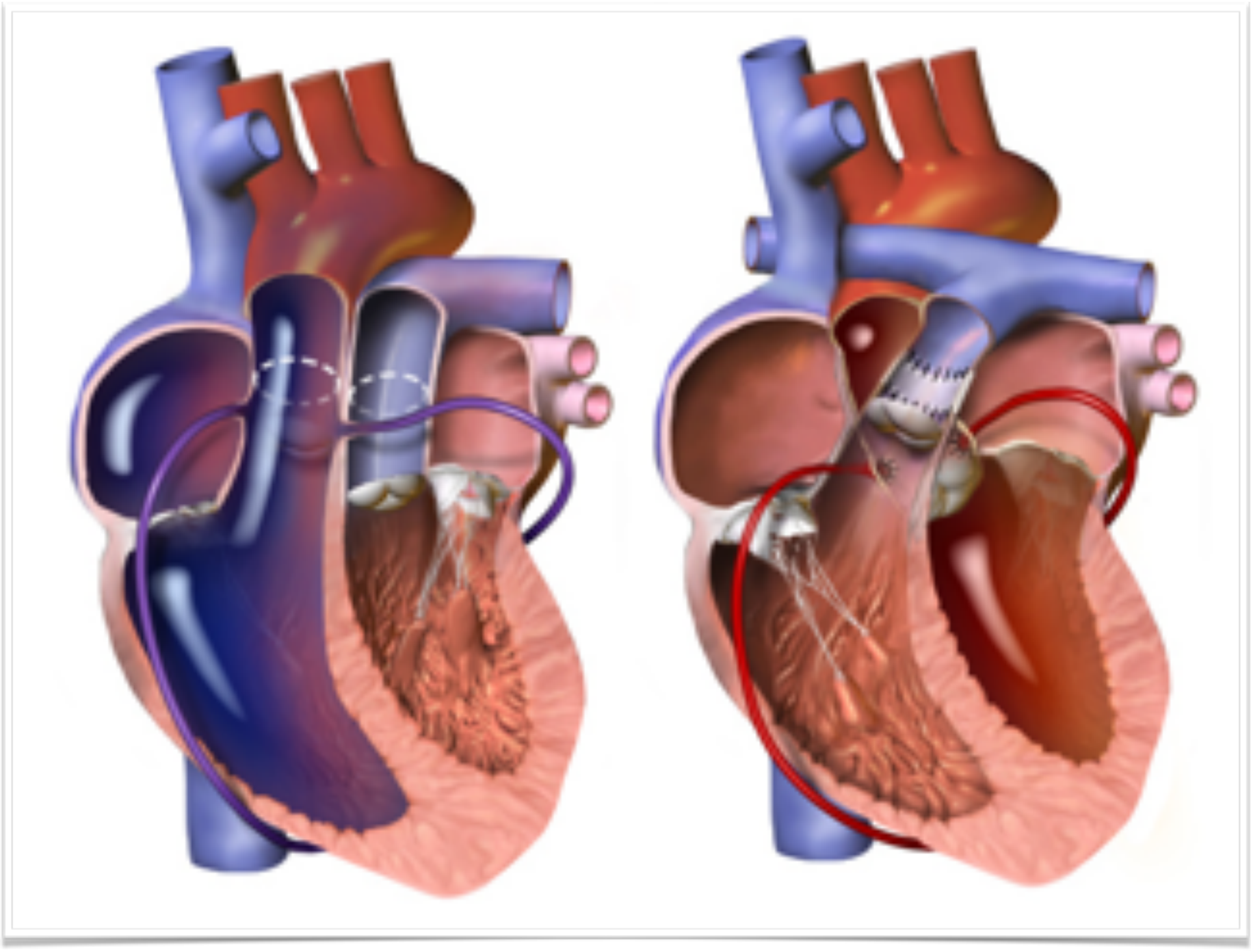
1975



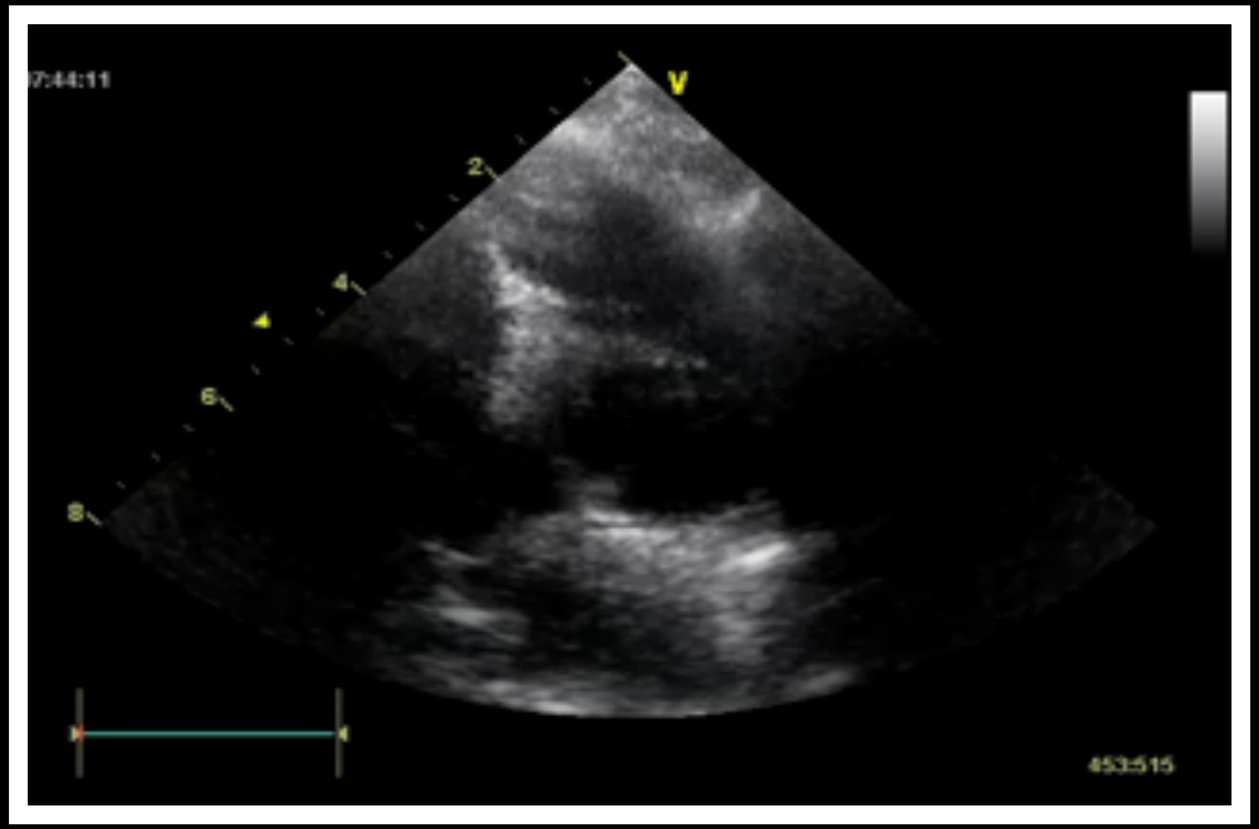
Yves Lecompte

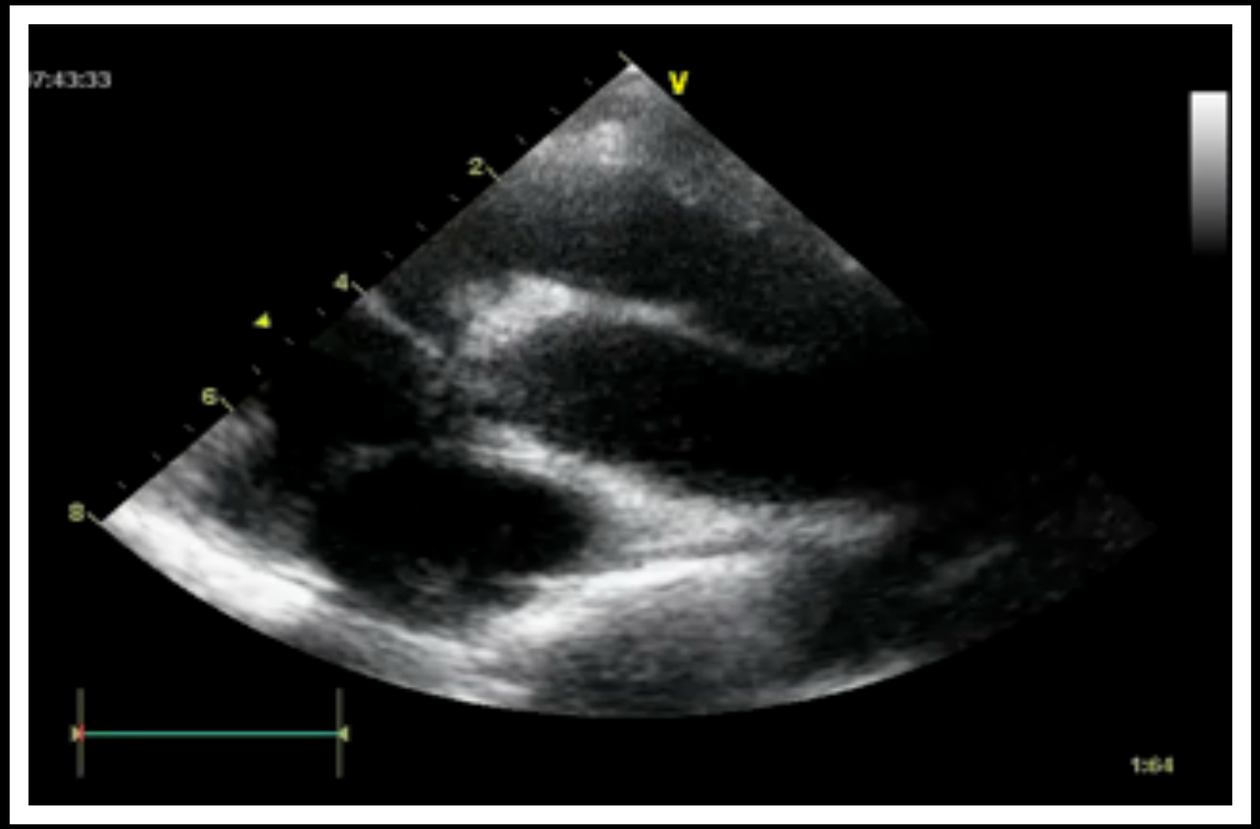
1981



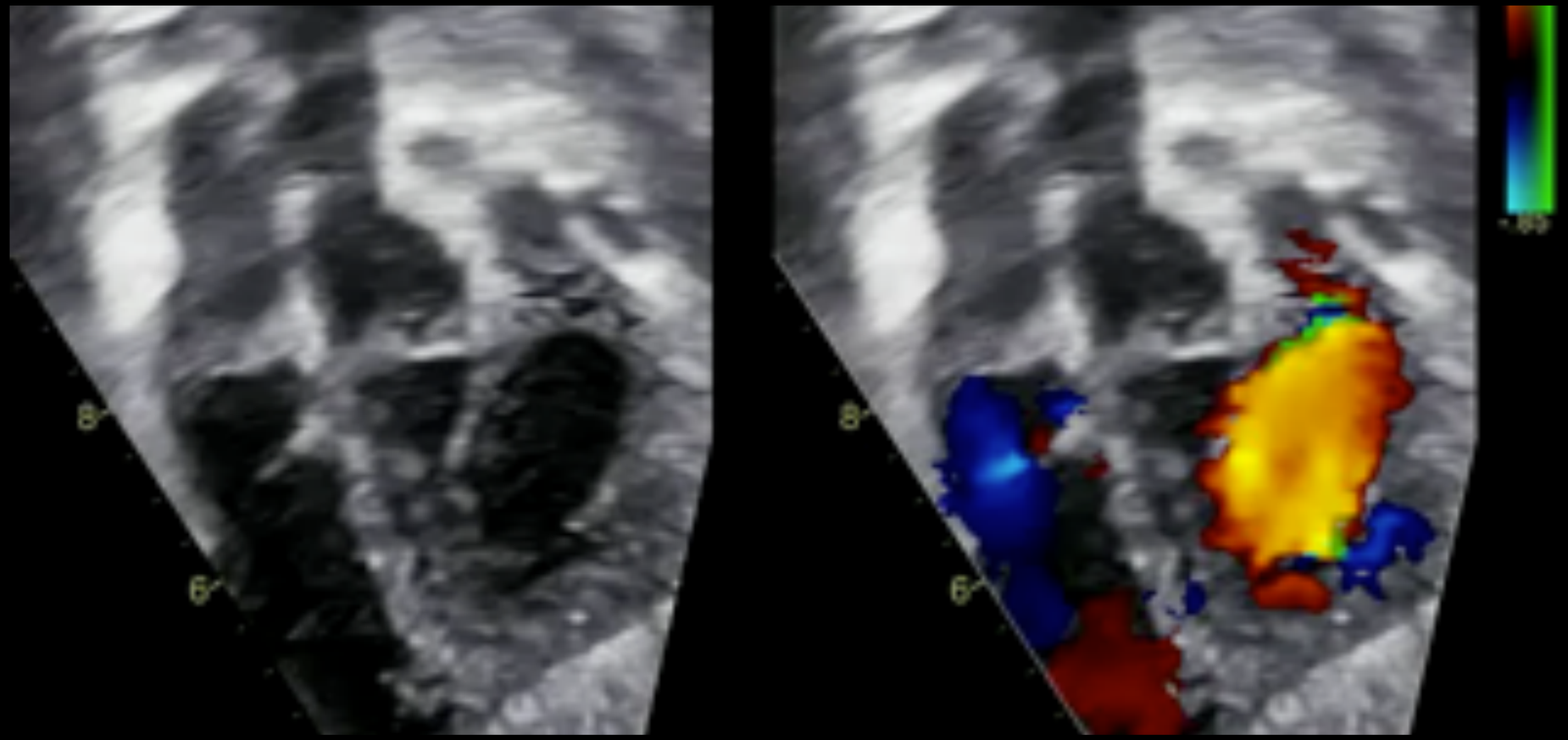


Outflow tracts after the arterial switch for TGA

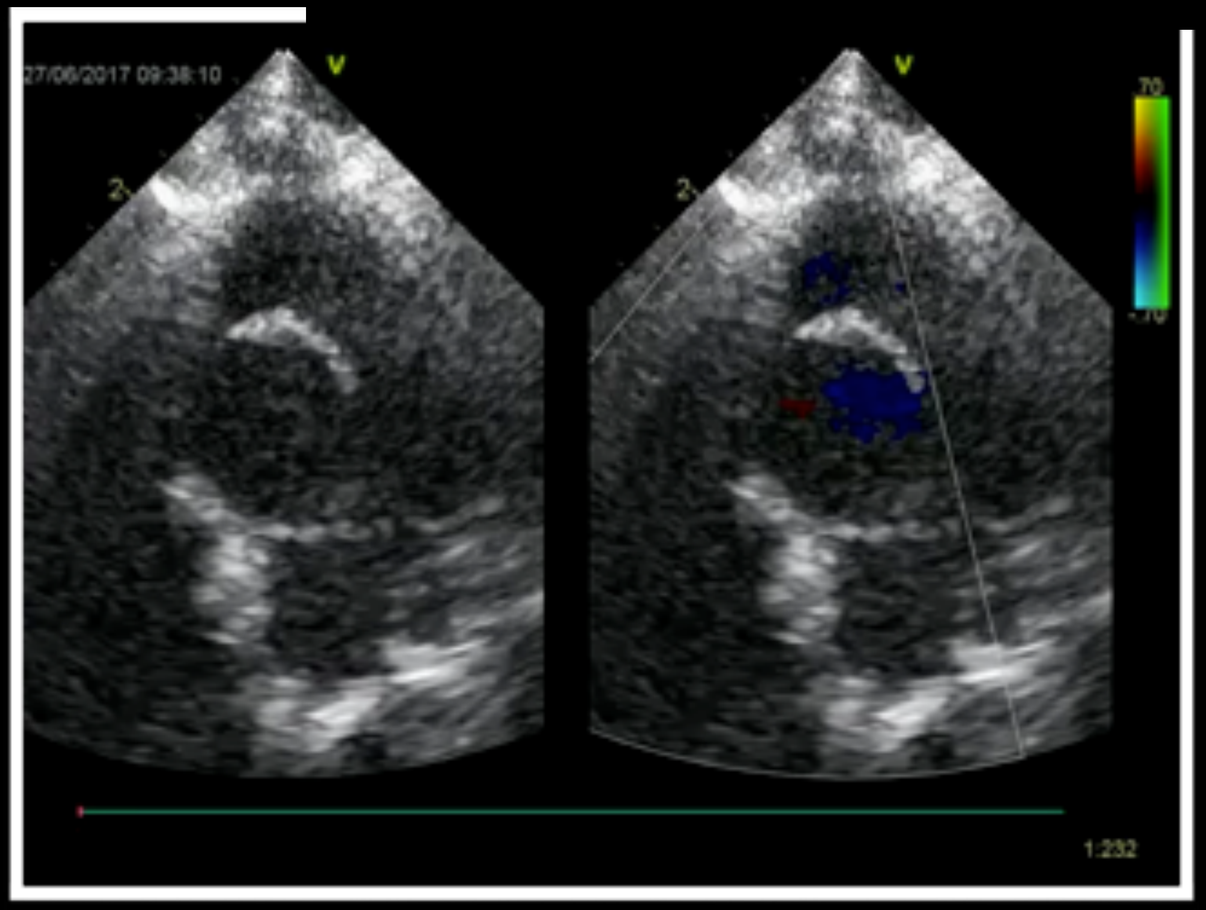


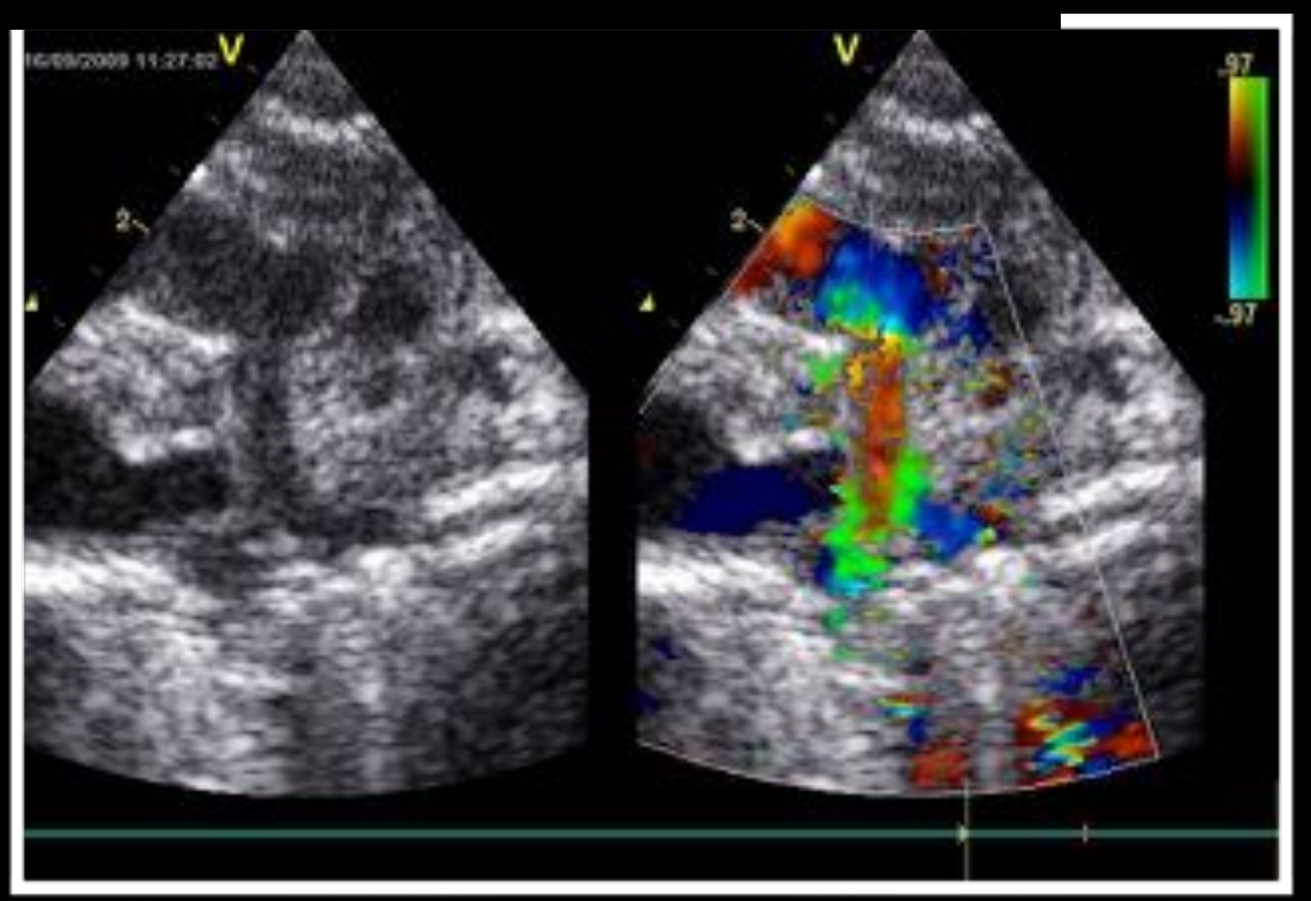


Outflow tracts after the arterial switch for TGA

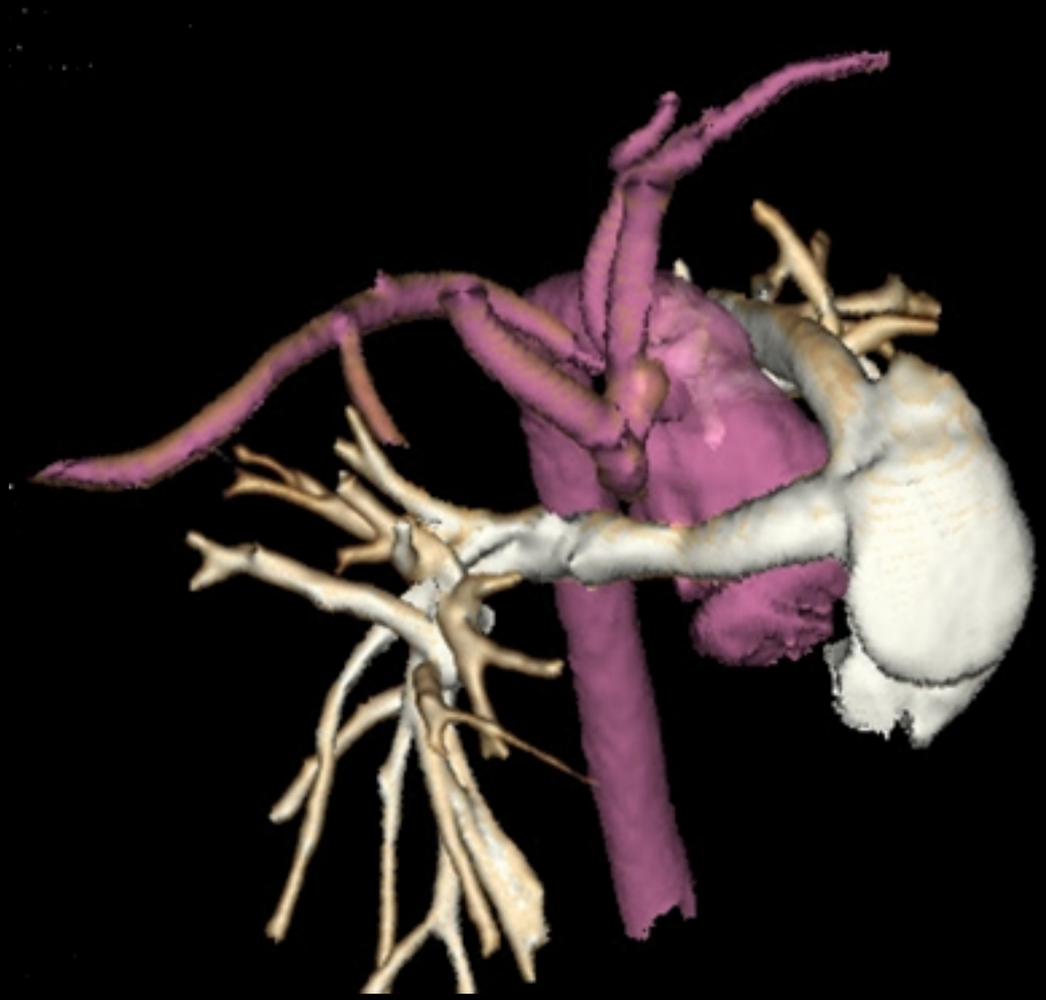


Outflow tracts after the arterial switch for TGA
The Lecompte manoeuver

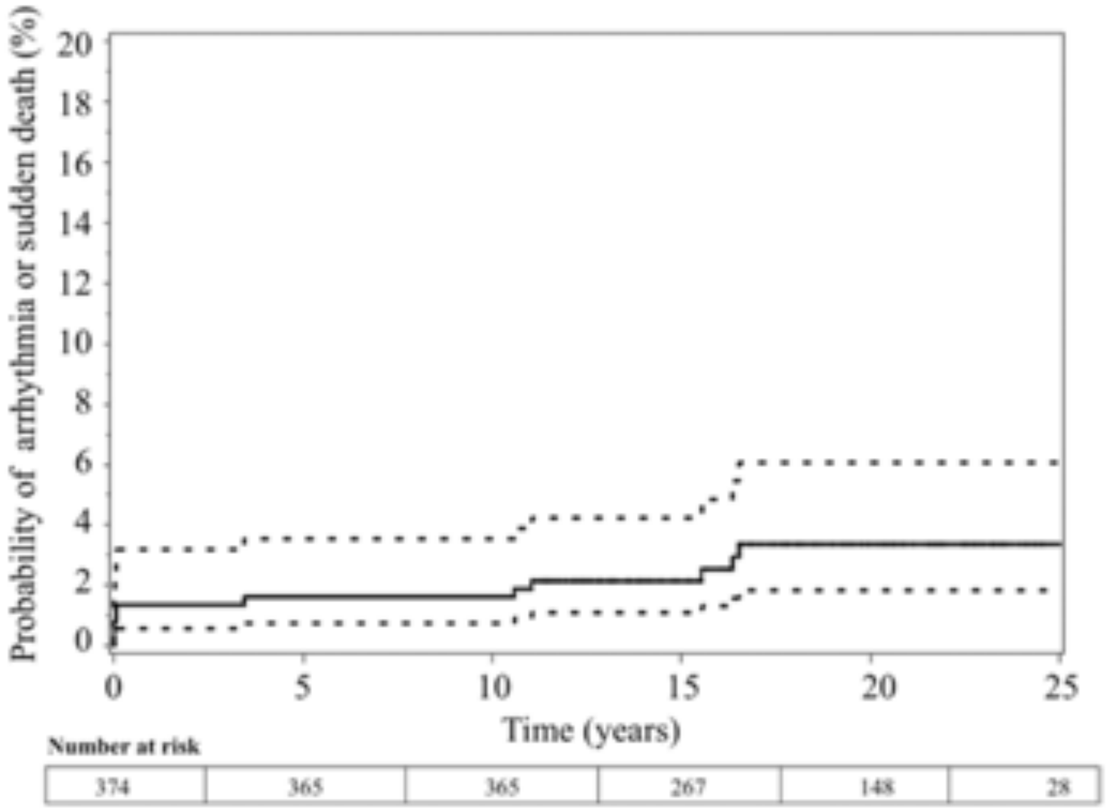




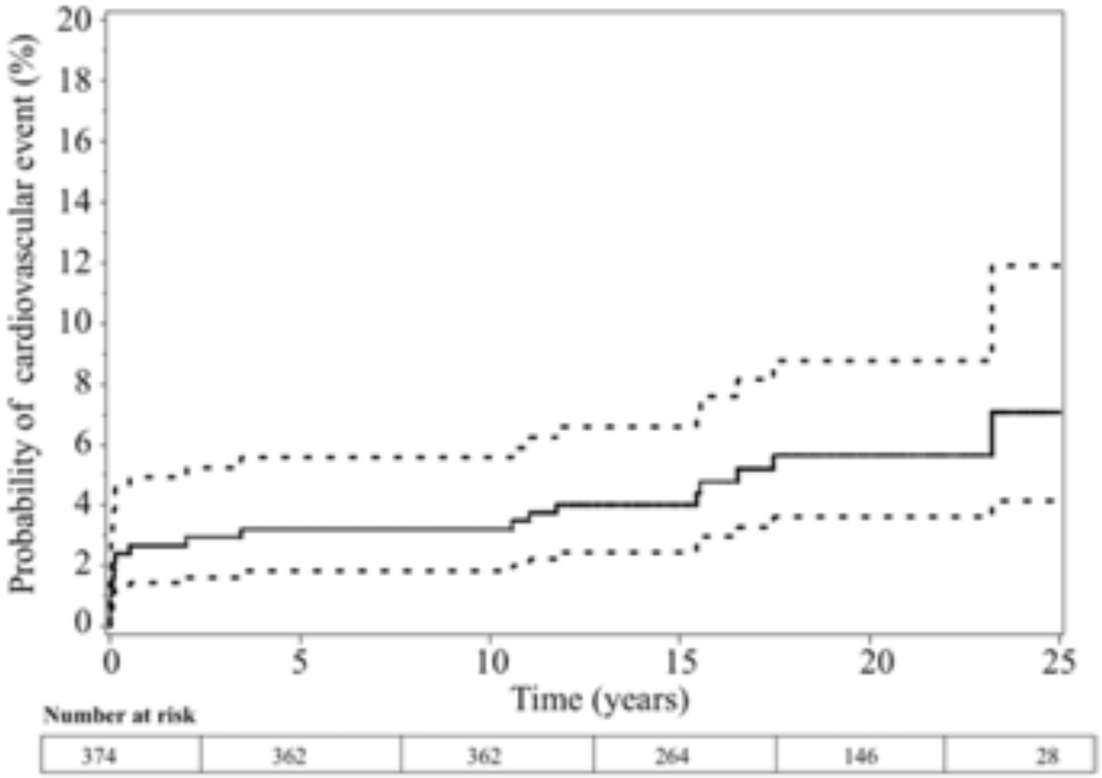
Outflow tracts after the arterial switch for TGA
The Lecompte manoeuver



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.