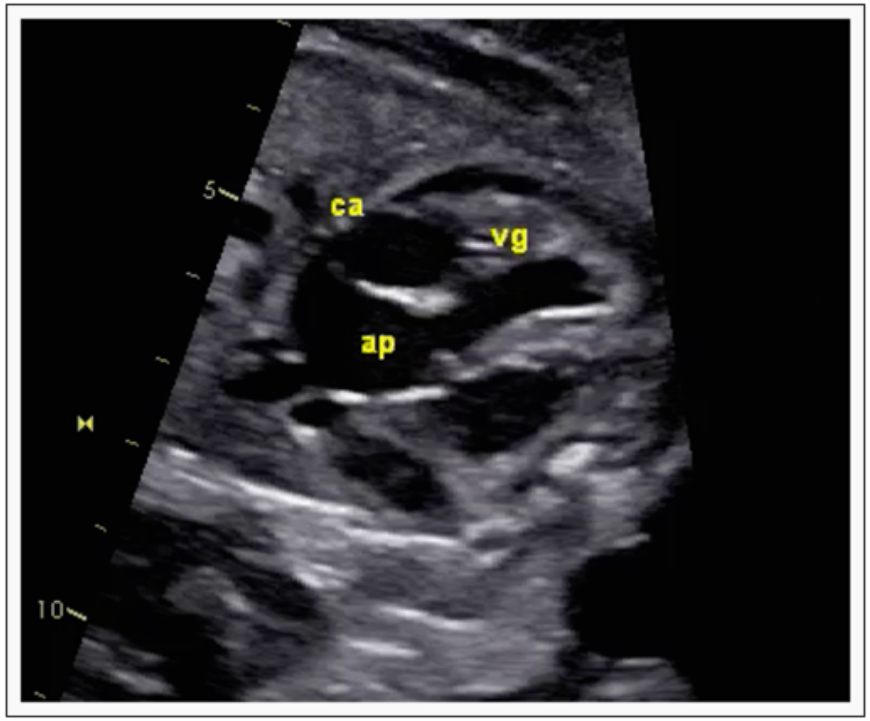
Transpositions des gros vaisseaux

Le point du vue du cardiopédiatre

Dr Daniela Laux, UE3C
M3C-HML-Centre des Malformations Congénitales Cardiaques complexes



Cardiopathies congénitales humaines	Fréquence	Incidence
Communication interventriculaire (CIV)	30%	1500
Communication interauriculaire (CIA)	8%	400
Sténose pulmonaire (SP)	7%	350
Persistance du canal artériel (PCA)	7%	350
Coarctation de l'aorte (CoA)	6%	300
Tétralogie de Fallot (T4F)	6%	300
Transposition des gros vaisseaux (TGV)	**5%**	**250**
Sténose aortique (SA)	5%	250
Canal atrioventriculaire (CAV)	4%	200
Atrésie pulmonaire à septum intact (APSI)	2%	100
Atrésie pulmonaire à septum ouvert (APSO)	2%	100
Atrésie tricuspide (AT)	2%	100
Tronc artériel commun (TAC)	2%	100
Retour veineux pulmonaire anormal (RVPA)	2%	100
Malpositions vasculaires (MV)	1%	50
Syndrome d'hypoplasie du cœur gauche (SHCG)	1%	50
Interruption de l'arc aortique (IAA)	1%	50
Ventricule unique (VU)	1%	50
Anomalie d'Ebstein	1%	50
Discordances AV et VA	1%	50
Autres	6%	300

Transposition des gros vaisseaux

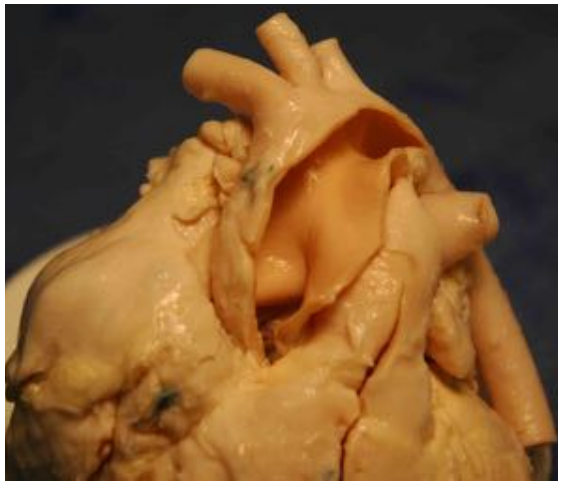
Définition:

- AP au dessus du ventricule G
- Aorte au dessus du ventricule D

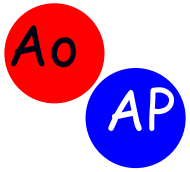
= discordance ventriculo-artérielle

- La TGV n'est qu'une des malpositions vasculaires qui incluent: TGV, VDDI, VGDI, malposition anatomiquement corrigée.

VX NORMOPOSES

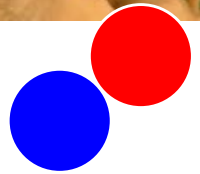


S = solitus

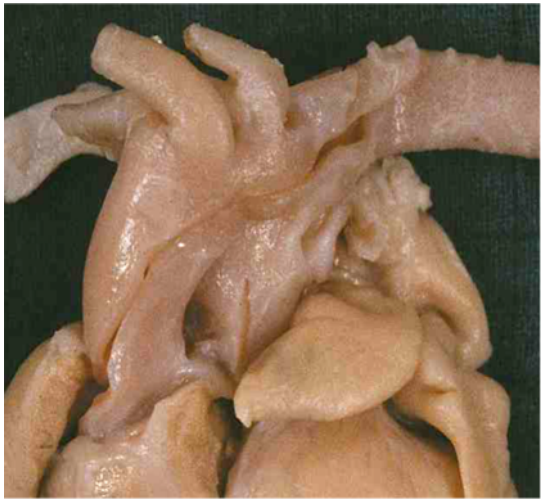


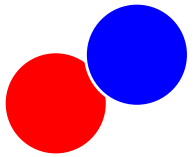
D L

I = inversus

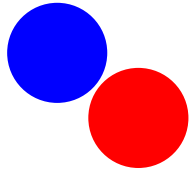


VX TRANSPOSES

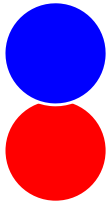




D-TGV



L-TGV

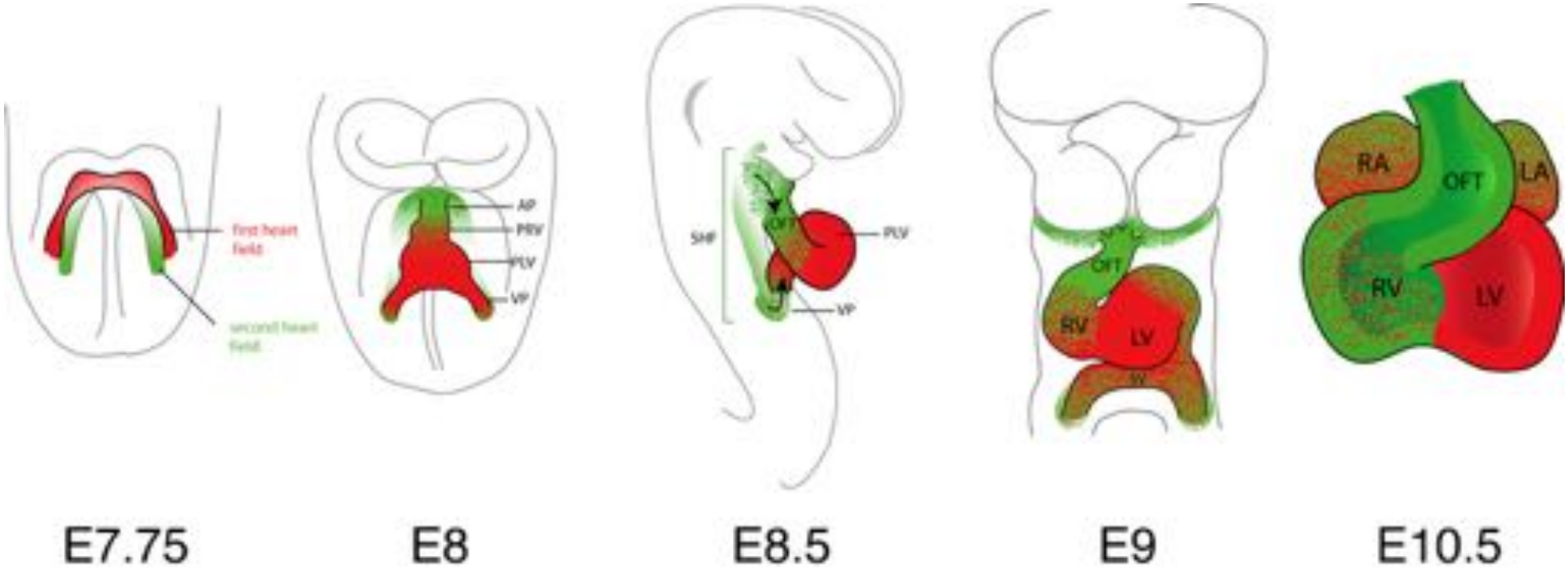


A = antéropostérieur

Courtesy L.Houyel

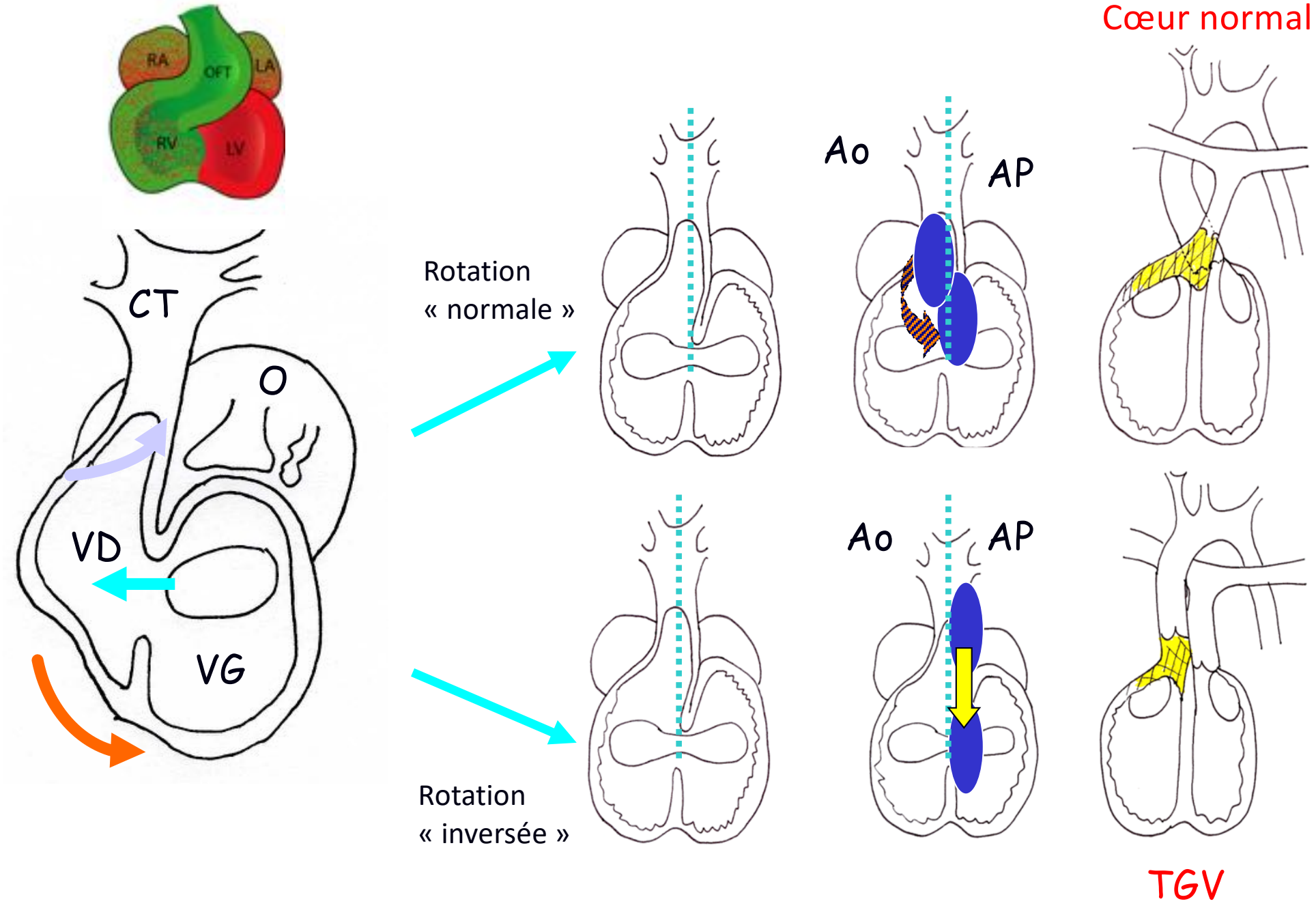
Rappel embryologique

Nouveau concept de morphogenèse : second champ cardiaque



Premier champ cardiaque (cardiac crescent) = 1st lineage

Second champ cardiaque (anterior heart field) = 2nd lineage


RA
OFT
LA
RV
LV
CT
O
VD
VG
Rotation
« normale »
Rotation
« inversée »
Ao
AP
Ao
AP
Cœur normal
TGV
Early looping
Convergence
Rotation
Courtesy L.Houyel

Types anatomiques: simples et complexes

- TGV « simple » – 60%: pas d'autre lésion associée
- TGV avec communication interventriculaire (CIV)
- TGV avec CIV et coarctation
- TGV avec CIV et sténose pulmonaire
- L-TGV (très rare)

- **Anomalies des valves AV:**
  - Fente mitrale et straddling mitral
  - Straddling tricuspide

Diagnostic prénatal

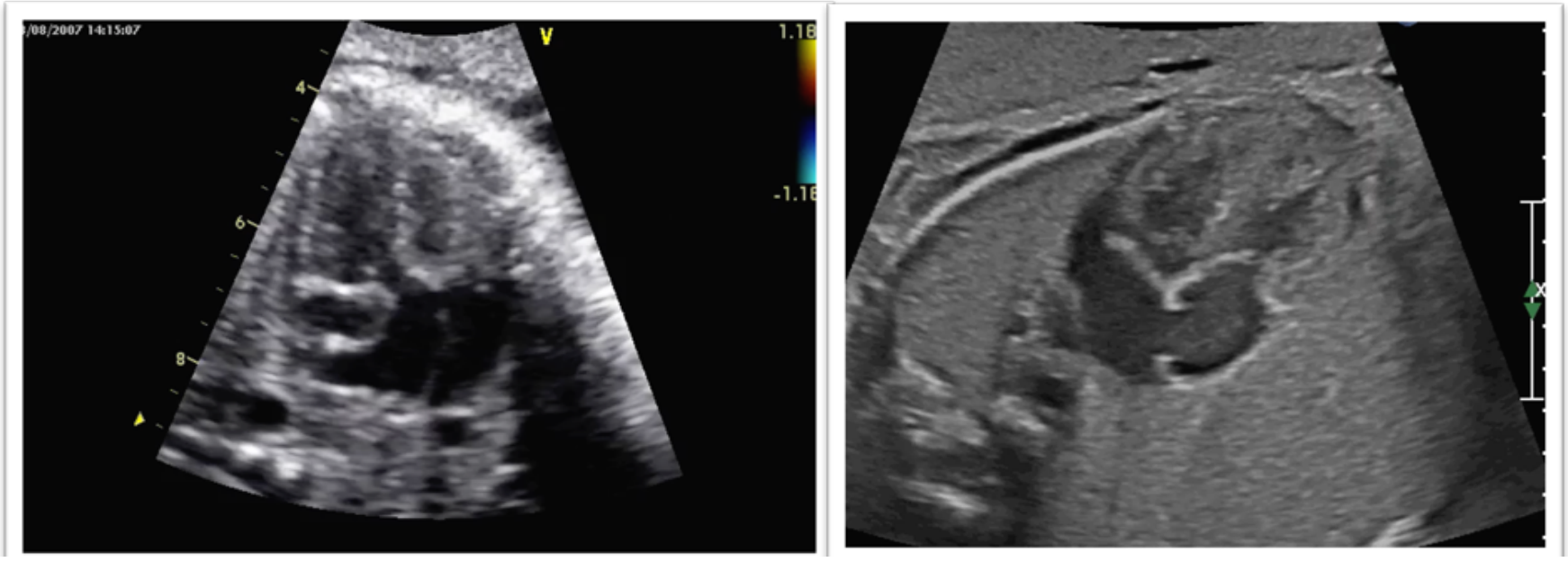
Table 3 - Prenatal diagnosis, pregnancy termination, perinatal and early neonatal mortality for selected (isolated) congenital heart anomalies - Paris Registry of Congenital Malformations, 1983-2000

Etude EPICARD

i) Transposition of Great Arteries

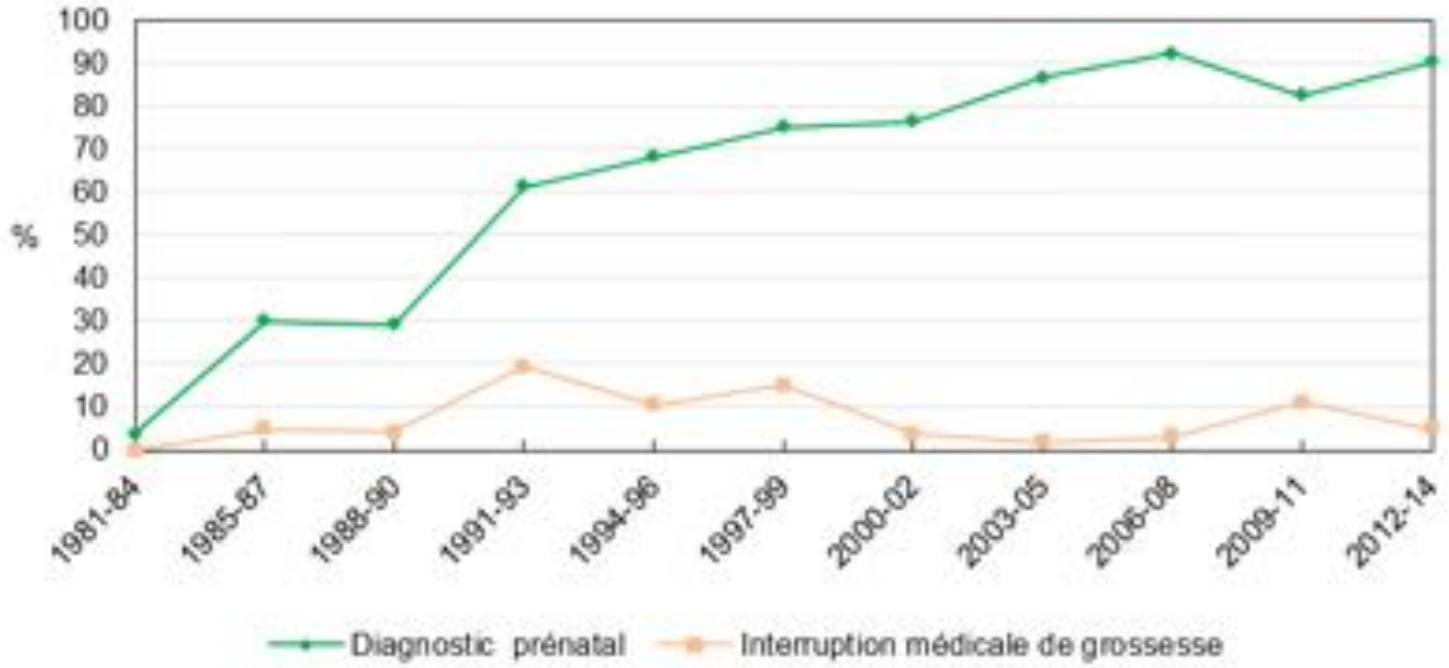
	1983 - 1988			1989 - 1994			1995 - 2000			
	N	%	95 % CI*	N	%	95 % CI*	N	%	95 % CI*	p*
Prenatal Diagnosis	16	12.5	1.6 - 38.3	27	48.1	28.7 - 68.1	40	72.5	56.1 - 85.4	0.001
Pregnancy Termination	17	0	0 - 19.5	27	7.4	0.9 - 24.3	40	0	0 - 8.8	0.62
First Week Mortality	16	18.8	4.0 - 45.6	24	8.3	1.0 - 27.0	39	2.6	0.1 - 13.5	0.04
Perinatal Mortality	17	23.5	6.8 - 49.9	25	12.0	2.5 - 31.2	40	5.0	0.6 - 16.9	0.02

Khoshnood B et al 2012



Evolution du diagnostic prénatal et IMG

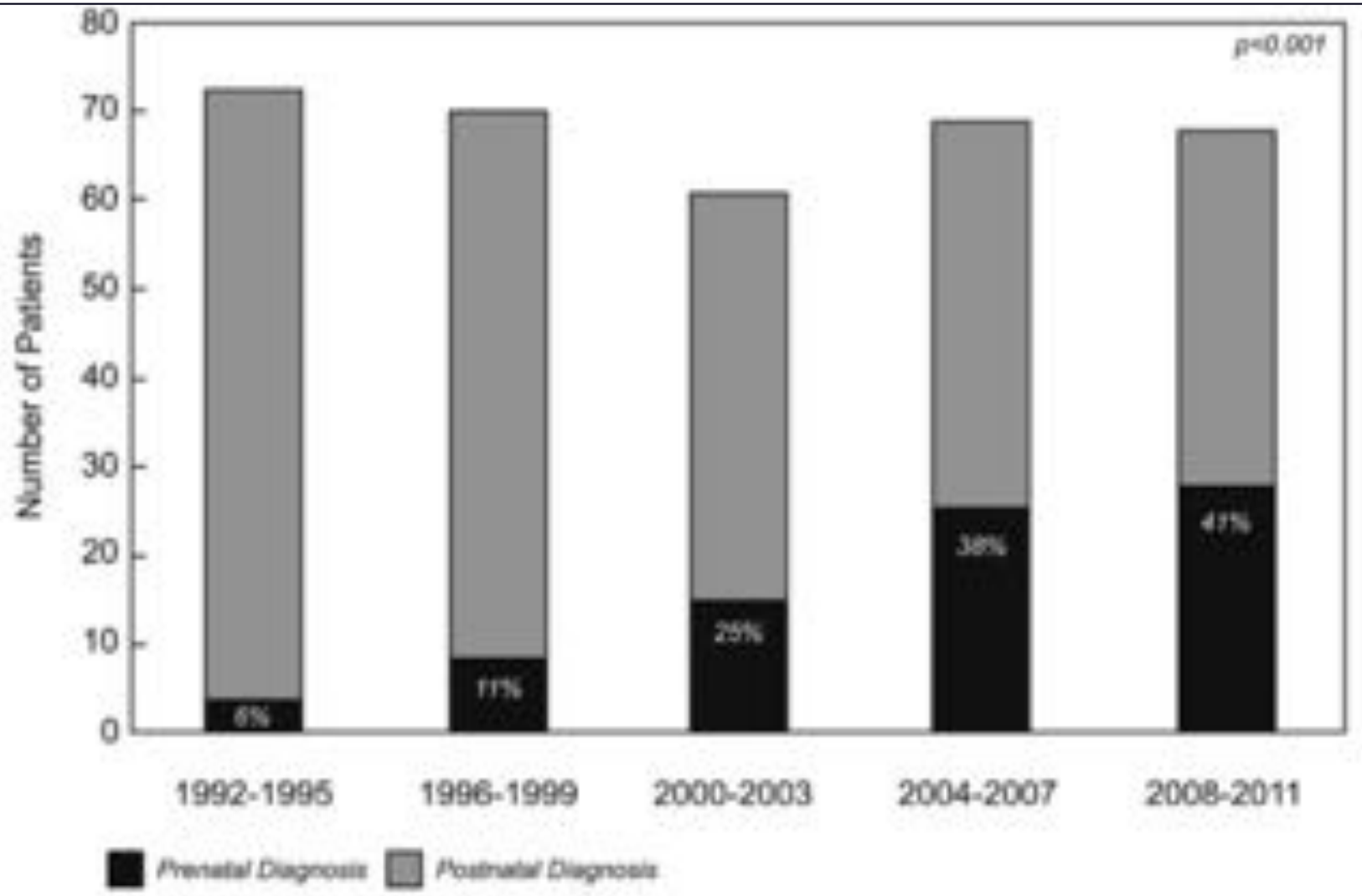
Transposition des gros vaisseaux (isolée)
Diagnostic prénatal et interruption médicale de grossesse



	1981-84	1985-87	1988-90	1991-93	1994-96	1997-99	2000-02	2003-05	2006-08	2009-11	2012-14
%DPN	3.8	30.0	29.2	61.3	68.1	75.0	76.5	86.5	92.2	82.5	90.3
% IMG	0.0	4.8	4.2	19.4	10.6	15.0	3.9	1.9	3.1	11.1	4.8

Naissances enregistrées à Paris de femmes domiciliées à Paris ou dans la Petite Couronne

Diagnostic prénatal ailleurs qu'en France



Escobar-Diaz et al. Ultrasoud Obst Gyn 2015

Detection of Transposition of the Great Arteries in Fetuses Reduces Neonatal Morbidity and Mortality

Damien Bonnet, MD; Anna Coltri, MD; Gianfranco Butera, MD; Laurent Fermont, MD; , MD

Comparison of Characteristics of Patients in the Prenatal and Postnatal Groups

	Postnatal Group	Prenatal Group	*P*
Isolated TGA	204	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±210	2.2±2.8	<0.01
Mechanical ventilation	95 (38)	12 (17.6)	<0.01
Metabolic acidosis ± MOF	56	8	<0.05
PGE_1 infusion	95	32	NS
BAS	168	54	NS
Preoperative mortality	15	0	<0.05
Coronary artery pattern	233 ASO	68 ASO	
Normal	168	47	NS
Abnormal	65	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_1, prostaglandin E_1; BAS, balloon atrioseptotomy; and ASO, arterial switch operation. Values are n (%).

Circulation. 1999;99:916-918

Impact postnatal du diagnostic prénatal (?)

- **Mortalité périopératoire**: études contradictoires
  - Amélioration globale des résultats concomitante de l'augmentation du DAN partout dans le monde

Bonnet et al. 1999, Khosnood at al. 2017

- **Morbidité périopératoire:**
  - Amélioration des délais opératoires, ventilation mécanique, acidose

Chakraborty et al. 2018, Cloete et al. 2019

- **Devenir neurodeveloppemental:**
  - Association positive DAN démontrée dans la TGV

Calderon 2012

PEC pratique en France

- DAN TGV -> CPDPN
- Confirmation par un cardiopédiatre expert
- Accompagnement du couple:
  information sur la pathologie/PEC/pronostic
- Soutien psychologique

- Accouchement déclenché en milieu spécialisé
- Cardiopédiatre sur place en SDN
- Evaluation postnatale hémodynamique immédiate
- Décision si Rashkind ou non

- Transfert en cardio/SI/ néona

Recommendation	Class[a]	Level[b]
It is recommended that the obstetric anomaly scan be performed at 18-22 weeks of gestation	I	C
To increase prenatal detection, it is recommended that outflow tract views, in addition to four-chamber views, be included in obstetric anomaly scans	I	C
It is recommended that the diagnosis be confirmed by a foetal cardiology specialist and that parental counselling should also be provided by a foetal cardiology specialist and other related health professionals (foetal medicine specialists, obstetricians, paediatric cardiac surgeons and neonatologists)	I	C
It is recommended that a detailed foetal anomaly scan be performed by a foetal medicine specialist	I	C
Because the risk for foetal karyotype abnormality is low in cases of TGA IVS, karyotyping may be considered on an individual basis where appropriate	IIb	C
After foetal diagnosis, follow-up to term is recommended for early detection of the development of high-risk features, which may require immediate intervention following delivery	I	C

Recommendation	Class[a]	Level[b]
It is recommended that delivery takes place at or near a tertiary-care paediatric cardiology and paediatric cardiac surgery centre	I	C
Vaginal delivery at term is recommended in most cases, whereas caesarean delivery is recommended when high-risk features are identified	I	C

[a]Class of recommendation.
[b]Level of evidence.
Recommendations for perinatal management

Sarris et al. City 2017

Diagnosis and Treatment of Fetal Cardiac Disease

A Scientific Statement From the American Heart Association

Table 19. Level of Care Assignment and Coordinating Action Plan

LOC	Definition	Example CHD	Delivery Recommendations	DR Recommendations
P	CHD in which palliative care is planned	CHD with severe/fatal chromosome abnormality or multisystem disease	Arrange for family support/palliative care services Normal delivery at local hospital	
1	CHD without predicted risk of hemodynamic instability in the DR or first days of life	VSD, AVSD, mild TOF	Arrange cardiology consultation or outpatient evaluation Normal delivery at local hospital	Routine DR care Neonatal evaluation
2	CHD with minimal risk of hemodynamic instability in DR but requiring postnatal catheterization/surgery	Ductal-dependent lesions, including HLHS, critical coarctation, severe AS, IAA, PA/IVS, severe TOF	Consider planned induction usually near term Delivery at hospital with neonatologist and accessible cardiology consultation	Neonatalogist in DR Routine DR care, initiate PGE if indicated Transport for catheterization/surgery
3	CHD with likely hemodynamic instability in DR requiring immediate specialty care for stabilization	d-TGA with concerning atrial septum primum (note: it is reasonable to consider all d-TGA fetuses without an ASD at risk) Uncontrolled arrhythmias CHB with heart failure	Planned induction at 38–39 wk; consider C/S if necessary to coordinate services Delivery at hospital that can execute rapid care, including necessary stabilizing/lifesaving procedures	Neonatologist and cardiac specialist in DR, including all necessary equipment Plan for intervention as indicated by diagnosis Plan for urgent transport if indicated
4	CHD with expected hemodynamic instability with placental separation requiring immediate catheterization/surgery in DR to improve chance of survival	HLHS/severely RFO or IAS d-TGA/severely RFO or IAS and abnormal DA Obstructed TAPVR Ebstein anomaly with hydrops TOF with APV and severe airway obstruction Uncontrolled arrhythmias with hydrops CHB with low ventricular rate, EFE, and/or hydrops	C/S in cardiac facility with necessary specialists in the DR usually at 38–39 wk	Specialized cardiac care team in DR Plan for intervention as indicated by diagnosis; may include catheterization, surgery, or ECMO
5	CHD in which cardiac transplantation is planned	HLHS/IAS, CHD including severe Ebstein anomaly, CHD, or cardiomyopathy with severe ventricular dysfunction	List after 35 wk of gestation C/S when heart is available	Specialized cardiac care team in DR

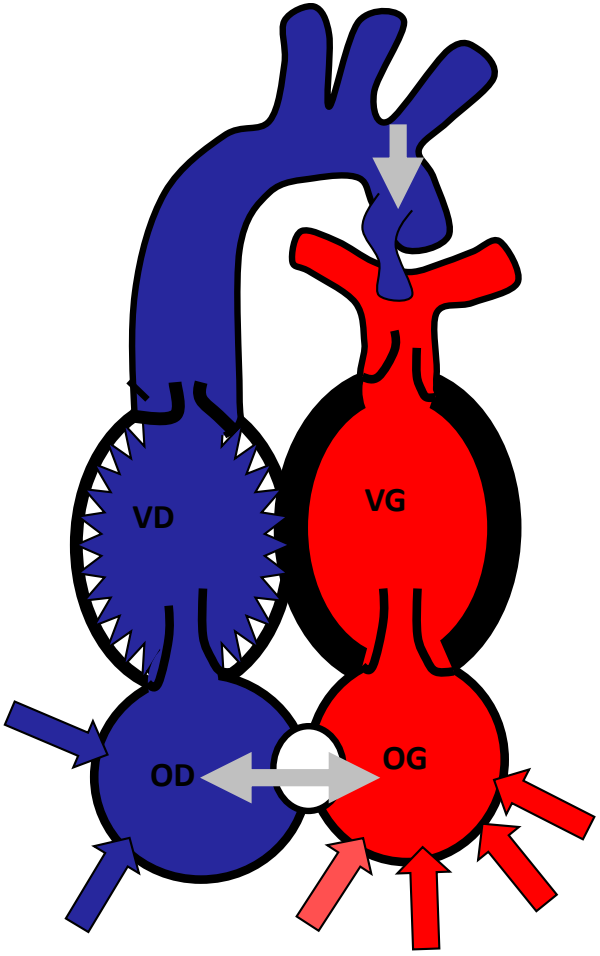
AHA 2014 Donofrio et al.

Clinique postnatal

Cyanose réfractaire en salle de naissance sans détresse respiratoire

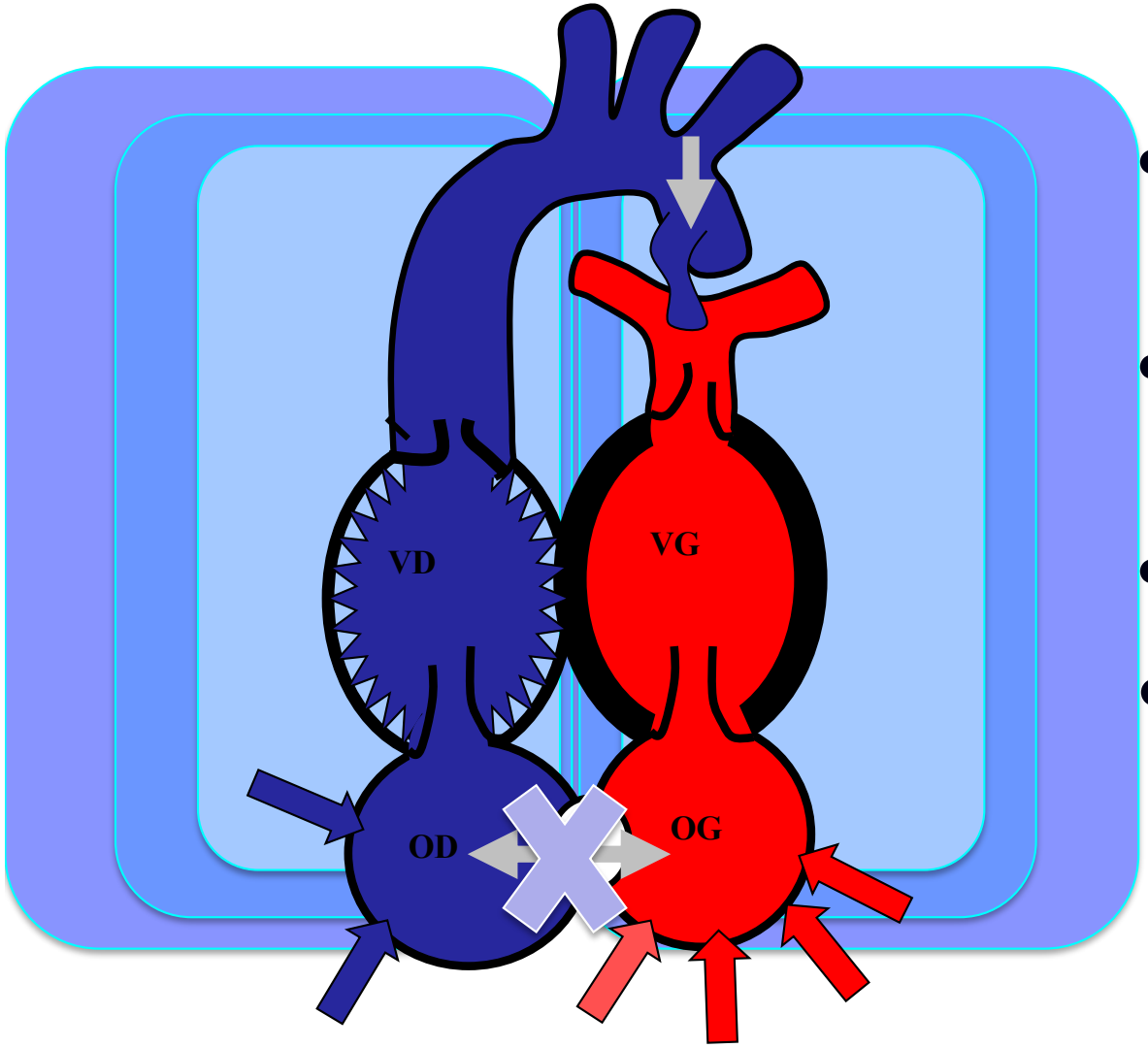
= TGV jusqu'à la preuve du contraire

Pathophysiologie TGV simple



- Cardiopathie cyanogène car aorte nait du VD !
- **Circulation en parallèle**
- CA et FOP obligatoire pour un mixing efficace
- CA shunte Ao-AP à cause des résistances vasculaires
- FOP shunte G-D à cause des compliances ventriculaire

Le FOP restrictif avant Rashkind



- FOP restrictif ou fermé= Œdème pulmonaire
- Majoration de la cyanose
- Majoration de l’acidose
- Mixing inefficace jusqu’au décès

Prise en charge médicale néonatale

- **Rashkind**
  - Mixing
  - Déprécharge le VG
- **PGE1**
  - Effets secondaires: apnée, douleur, fièvre
  - Précharge le VG
- Surveillance glycémies
- Surveillance alimentation entérale: risque d'entéropathie/entérocolite
- Risque théorique d'AVC en cas de KTC: VD-aorte-cerveau

Clinical guidelines for the management of patients with transposition of the great arteries with intact ventricular septum

The Task Force on Transposition of the Great Arteries of the European Association for Cardio-Thoracic Surgery (EACTS) and the Association for European Paediatric and Congenital Cardiology (AEPC)

Recommendation	Class[a]	Level[b]	Ref[c]
Neonatal pulse oximetry screening is crucial for timely diagnosis of TGA	I	C	51
Echocardiography is the modality of choice for diagnosing TGA postnatally and allows accurate evaluation of the coronary artery pattern and exclusion of other relevant malformations in most cases	I	B	49, 50
Performance of BAS should be considered, under echocardiographic guidance	IIa	B	52–54

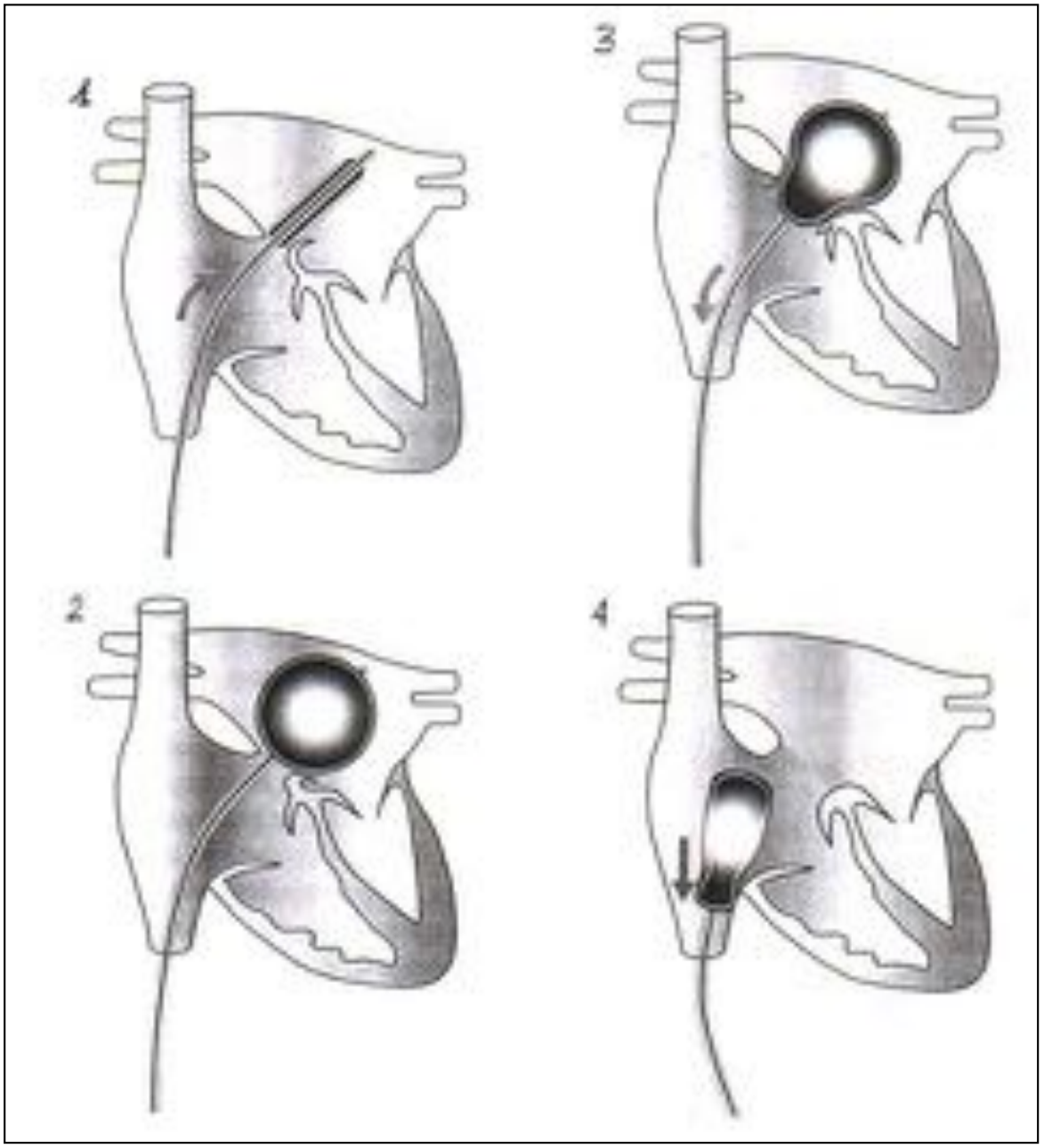
BAS: balloon atrial septostomy; TGA: transposition of the great arteries.
[a]Class of recommendation.
[b]Level of evidence.
[c]References
Recommendations for postnatal diagnosis

Recommendation	Class[a]	Level[b]
Immediately after birth, IV infusion of PGEI is recommended to maintain ductal patency until the comprehensive series of postnatal echocardiograms is complete and all sites of intercirculatory mixing have been evaluated	I	C
Avoidance of elective intubation of infants on PGE1 during transport is recommended. The decision to intubate prior to transport must be individualized	I	C
An individualized management strategy for low birth weight and premature infants is recommended, taking into account patient and institutional factors. Management options include primary repair as late as 3 months of age, late single-stage repair with postoperative VAD or ECLS support and two-stage repair	I	C
A primary ASO may be considered the preferred management strategy for low-birth-weight and premature infants and can be performed with acceptable but increased early and mid-term risk	IIb	C

ASO: arterial switch operation; ECLS: extracorporeal life support; IV: intravenous; PGE1: prostaglandin E1; VAD: ventricular assist device.
[a]Class of recommendation.
[b]Level of evidence.
Recommendations for perinatal management in a neonatal intensive care unit

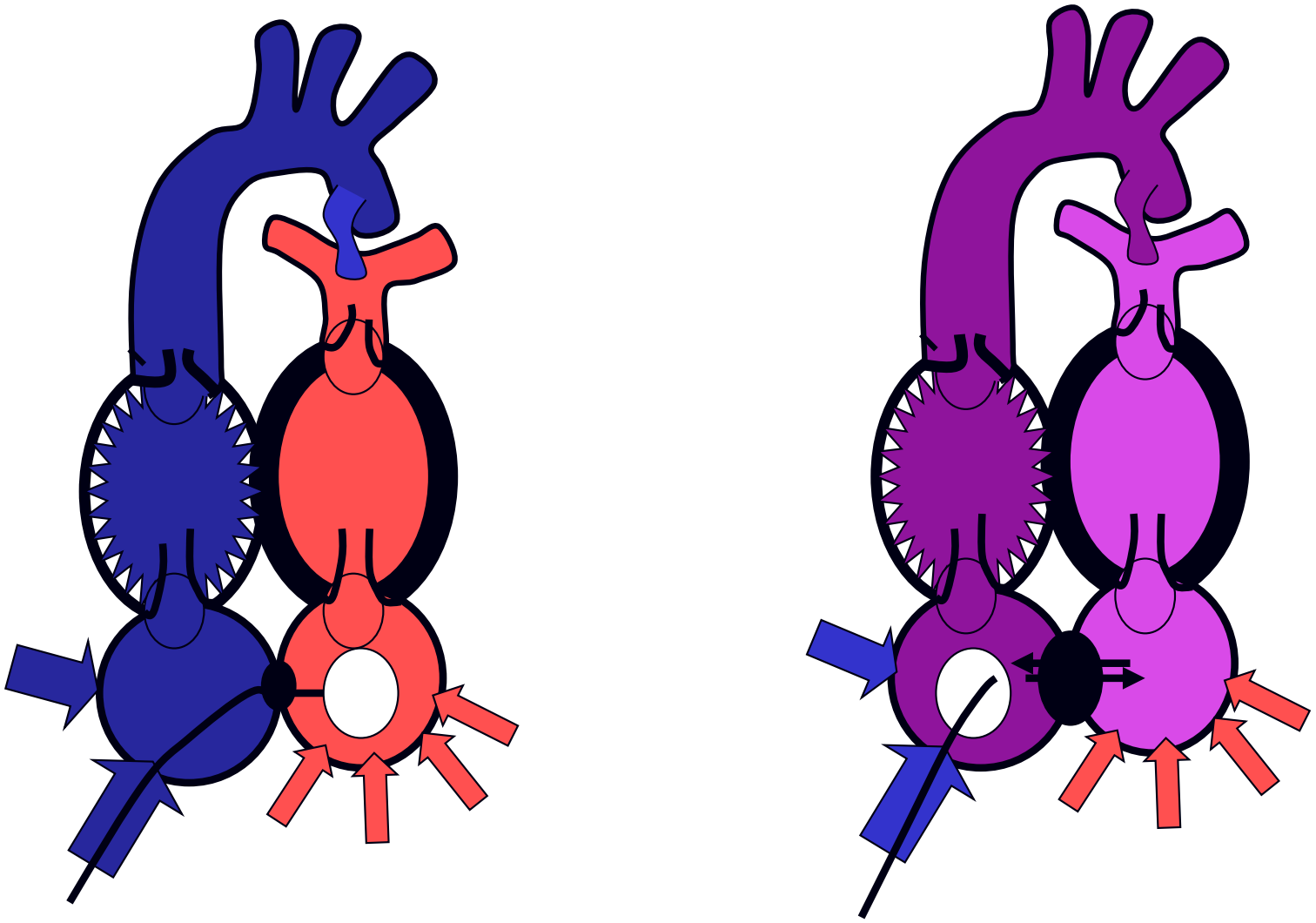
Sarris et al. City 2017

Manœuvre de Rashkind (1966)



Manœuvre de Rashkind : sous scopie ou écho

Rashkind: effet hémodynamique immédiat

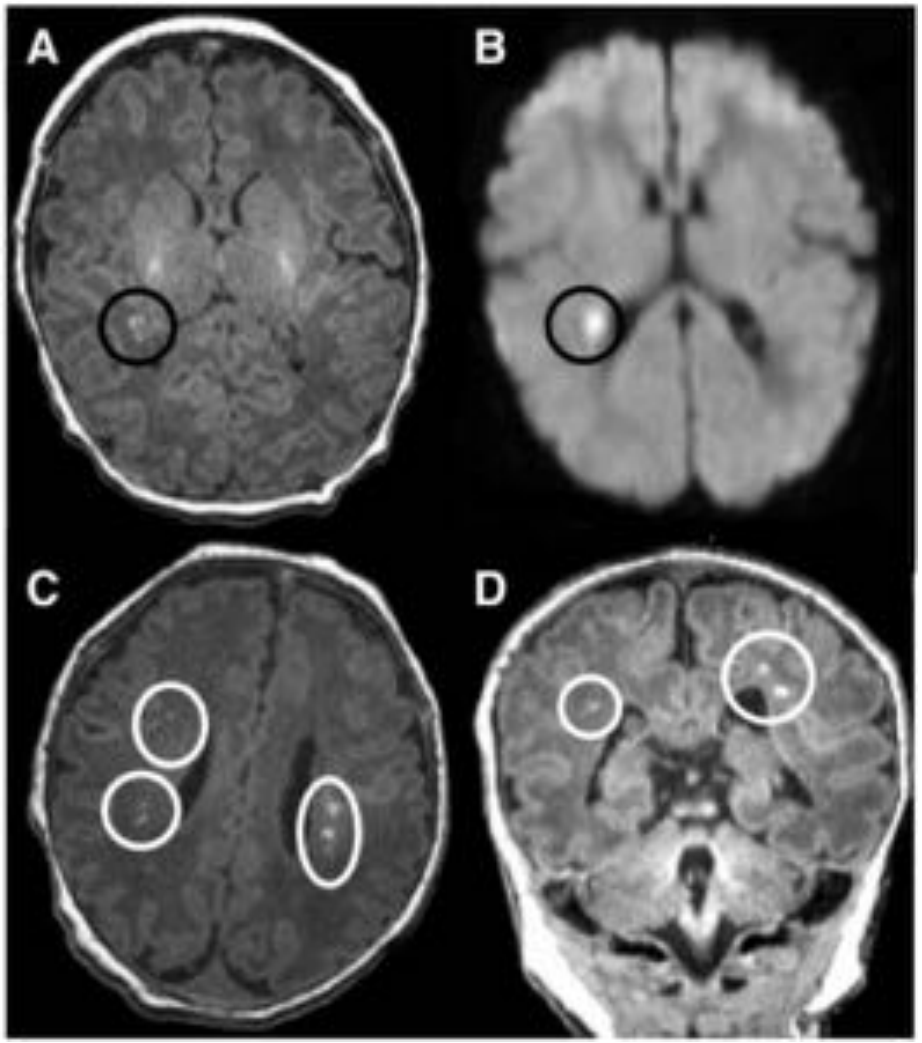


Procédure réalisée dans 70% à la naissance ou dans les premiers jours

Preoperative Brain Injury in Transposition of the Great Arteries Is Associated With Oxygenation and Time to Surgery, Not Balloon Atrial Septostomy

Petit et al. Circ 2009

N= 26 NN avec switch dont 14 avec Rashkind;
10/26 avaient une leucomalacie préopératoires



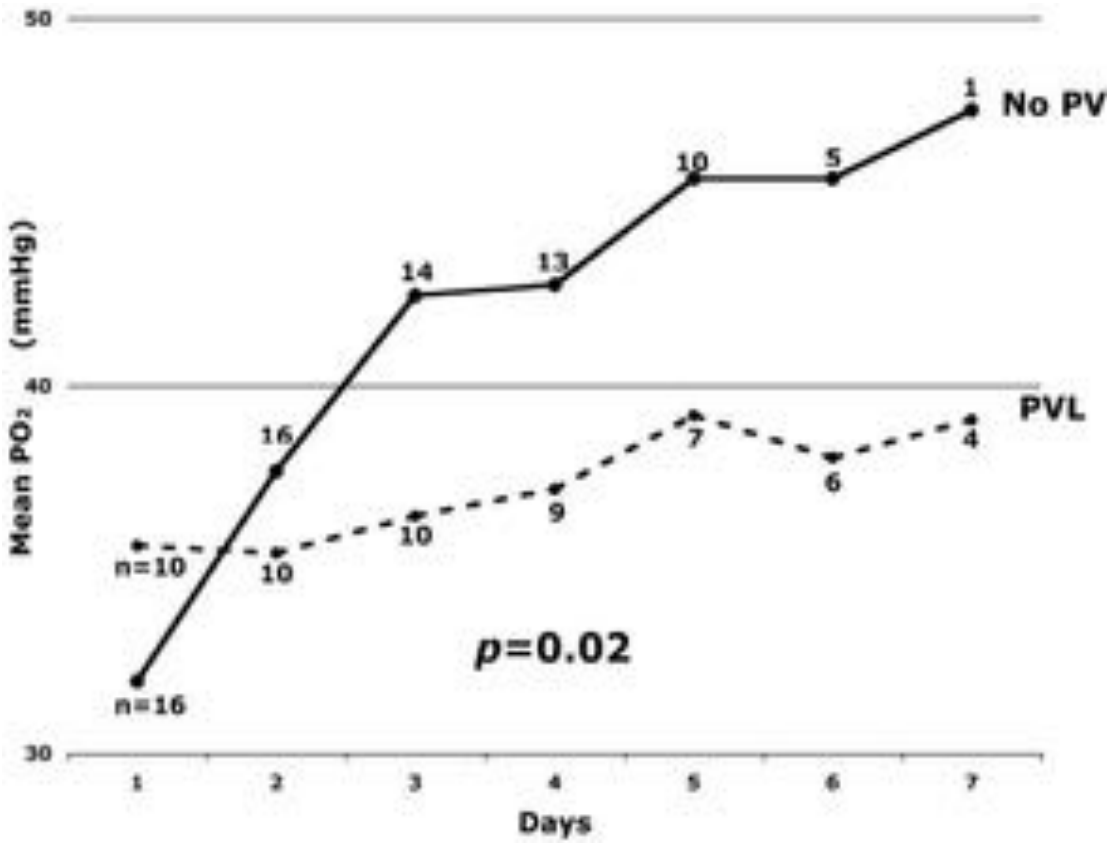


Figure 3.
A daily mean P_{O_2} was calculated for the PVL and no-PVL groups. Repeated-measures ANOVA demonstrated a significant difference in mean daily P_{O_2} between the PVL group (dashed line) and the no-PVL group (solid line; *P*=0.02). The PVL group never achieved a mean daily P_{O_2} >40 mm Hg.

Check liste écho pré op

- CIA large ou restrictive et CA ouvert ou fermé

- **Equilibre des ventricules**
  - Petit VD : risque de coarctation
  - Petit VG : vérifier la voie pulmonaire
- **Anatomie de la valve mitrale**
  - Fente non commissurale

- **Cardiopathies associées** (CIV, caorctation aortique)
- Valve pulmonaire (futur aortique)
- Discongruence aortopulmonaire ?
- Malalignement commissural ?
- Anatomie des artères coronaires ?

La question du VG dépréparé

- < 3 semaines: switch artériel
- > 3 semaines: évaluation forme du VG, présence CA/CIA/CIV, calcul de masse VG
- Deux stratégies
  - Switch avec ECMO postopératoire
  - Préparation du VG: cerclage + Blalock 7-10 jours puis switch
- Masse VG minimale : > 35 g/m2 calculée en TM

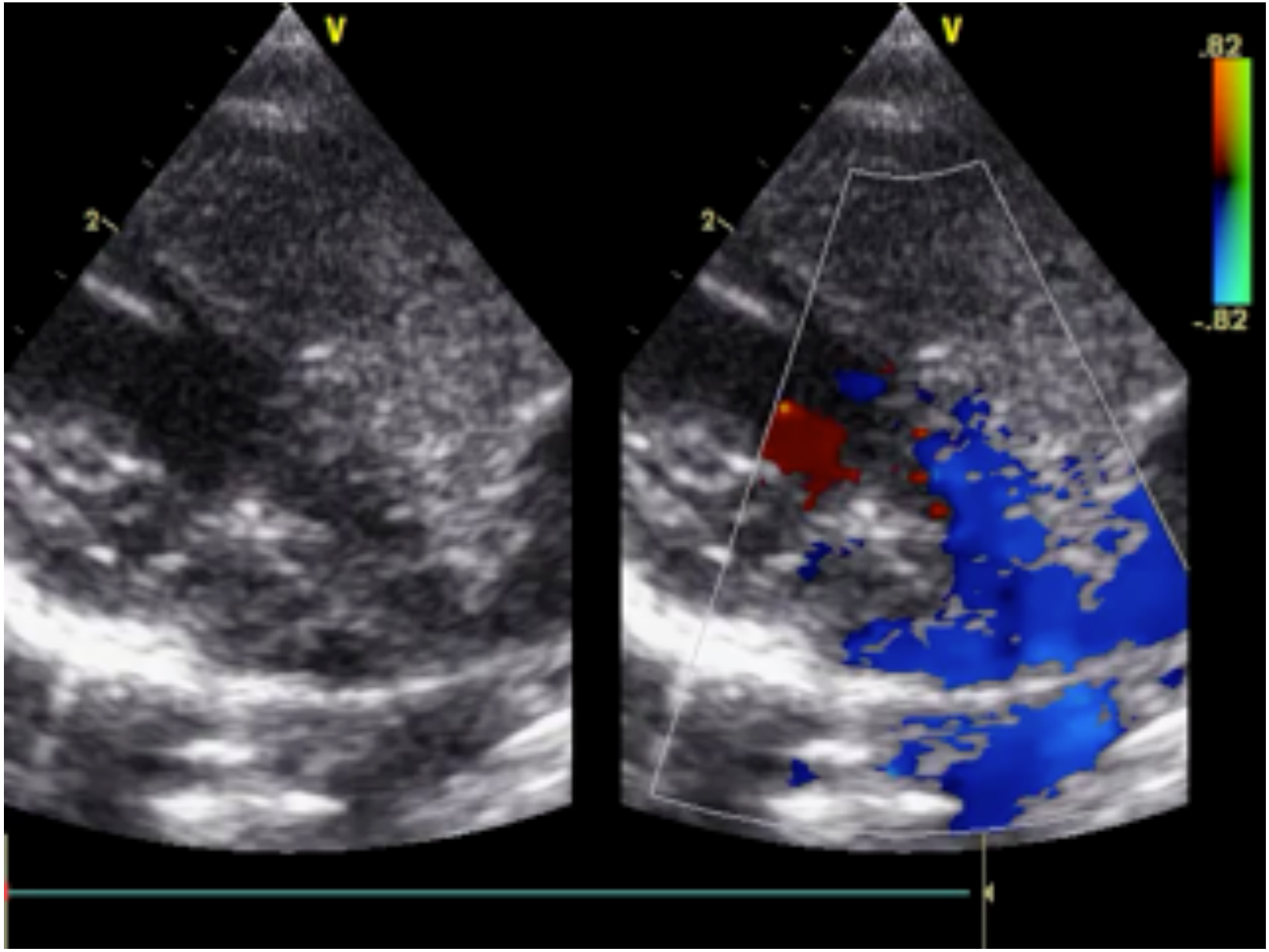
Table 1

LV Mass (ASE) = 1.04 * (LVED d + LVPW d + IVS d)^3 − LVED d^{3a}
Indexed LV Mass (G/m^2 = [0.8 * (LV Mass) to 0.6] / BSA

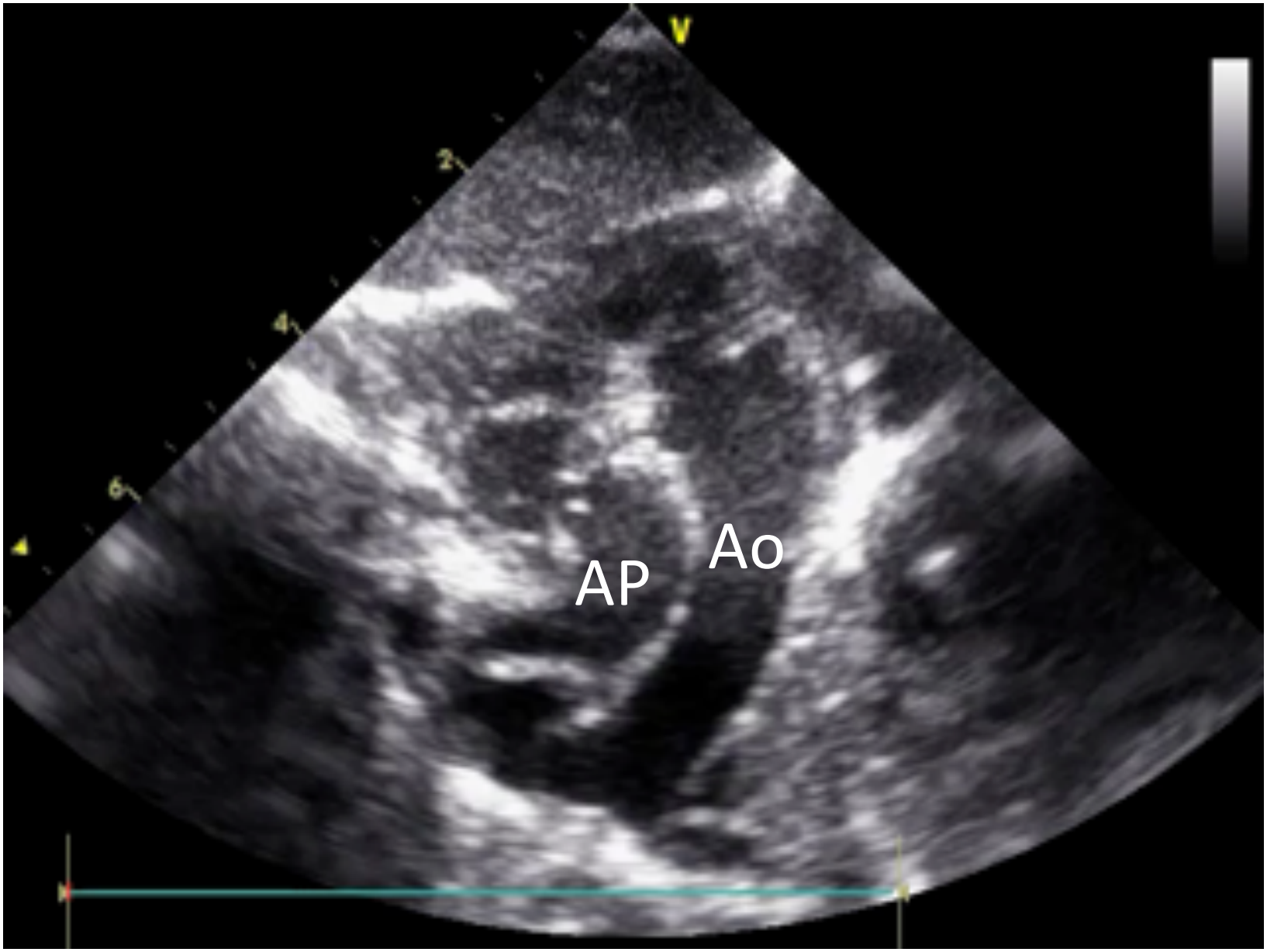
[a] LVED d, left ventricle end diastolic diameter; LVPW d, left ventricle posterior wall thickness; IVS d, left ventricle interventricular thickness; ASE, American Society of Echography.

Lacour-Gayet 2001
Sarrris et al. City 2017

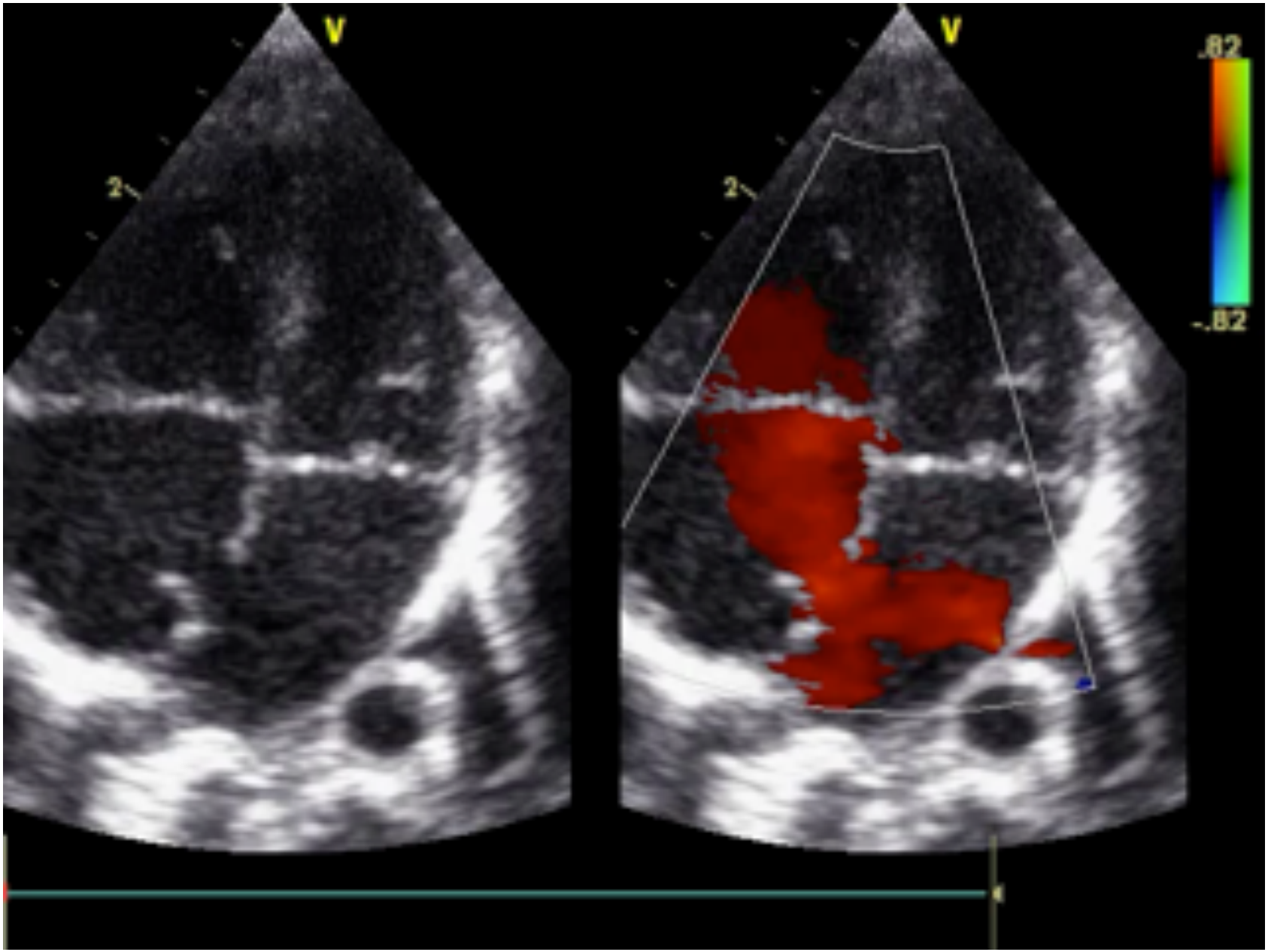
Vaisseaux parallèles: grand axe



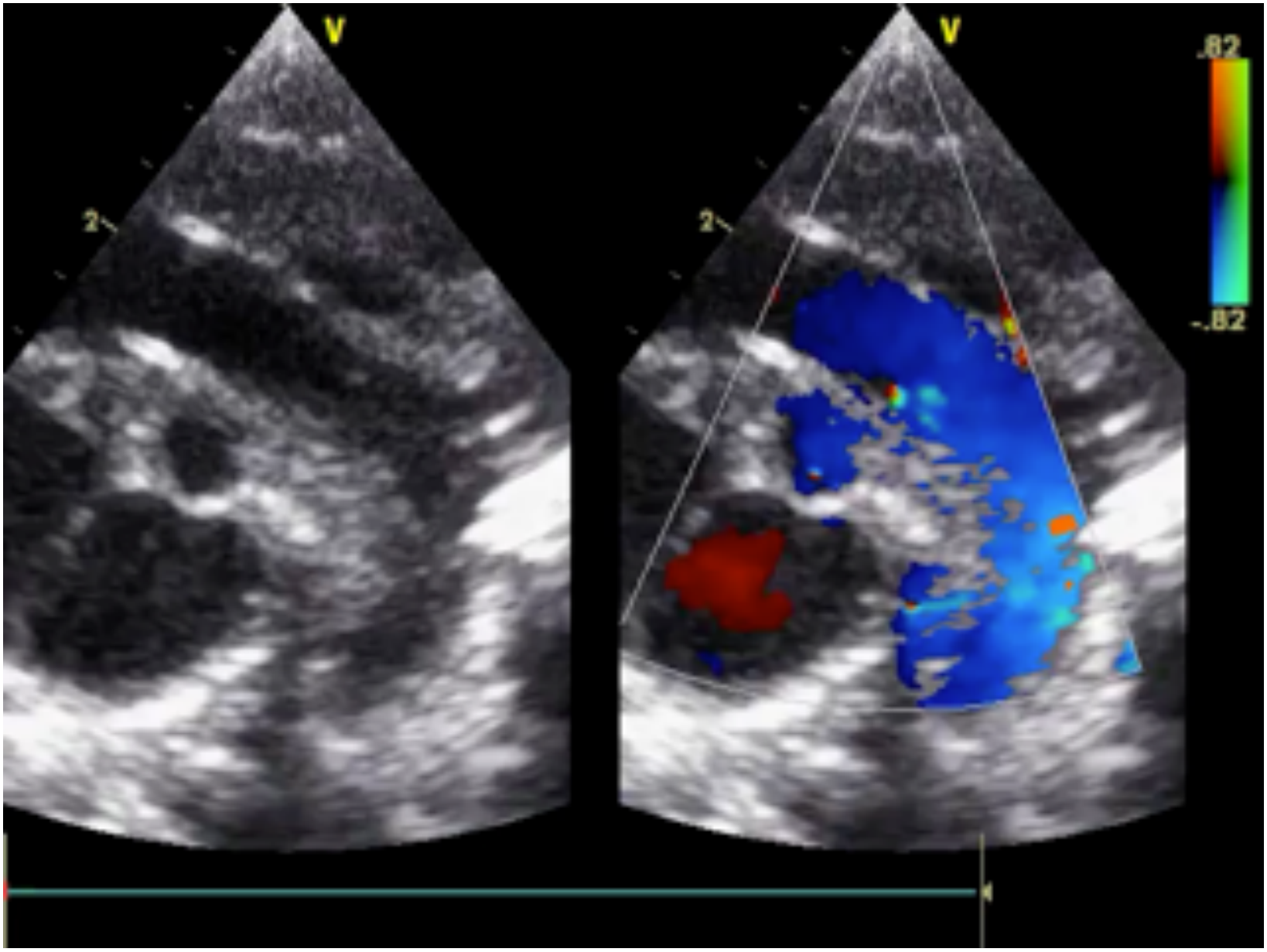
TGV en souscostal: vaisseaux parallèles



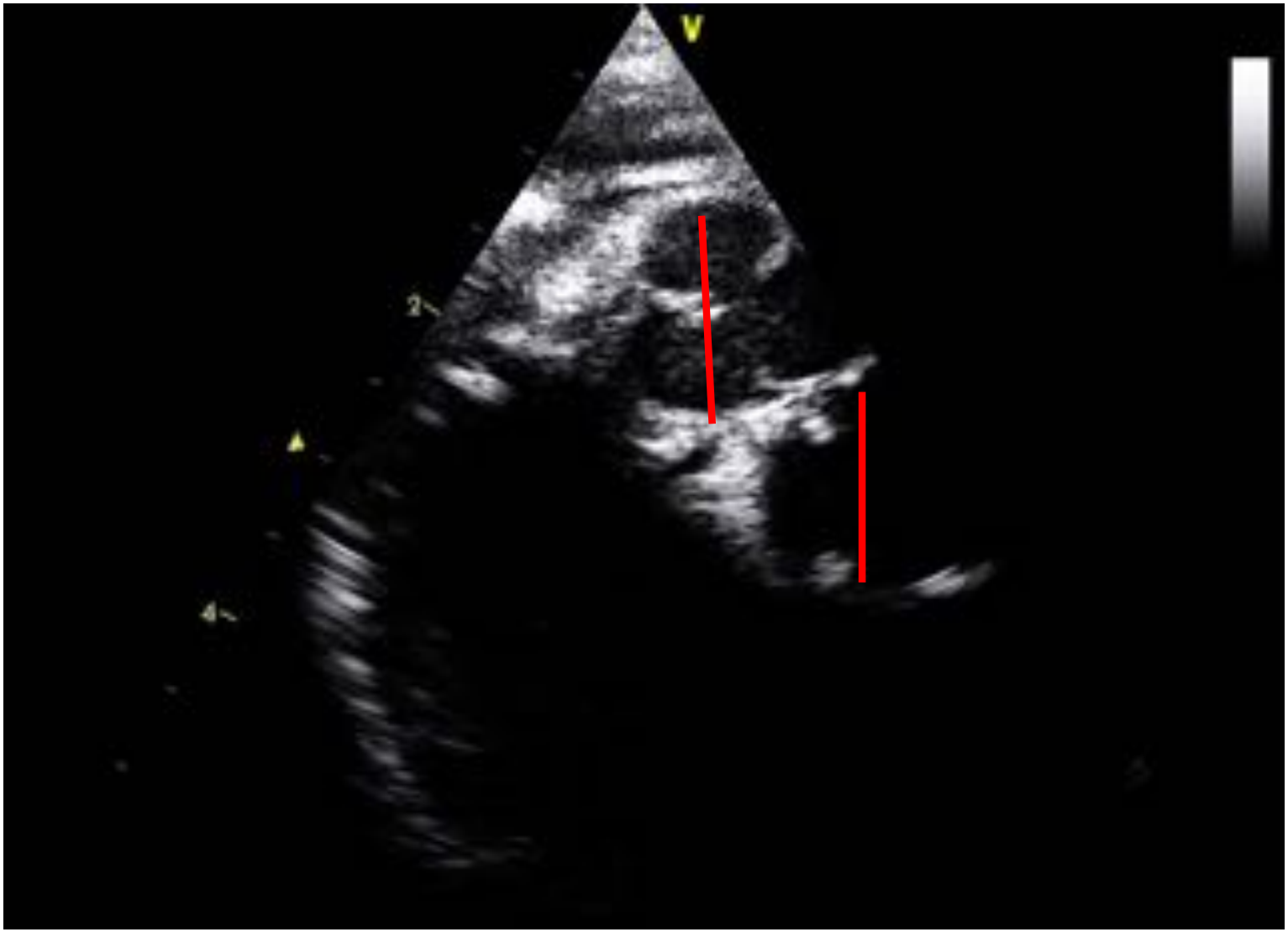
CIA post-RSK



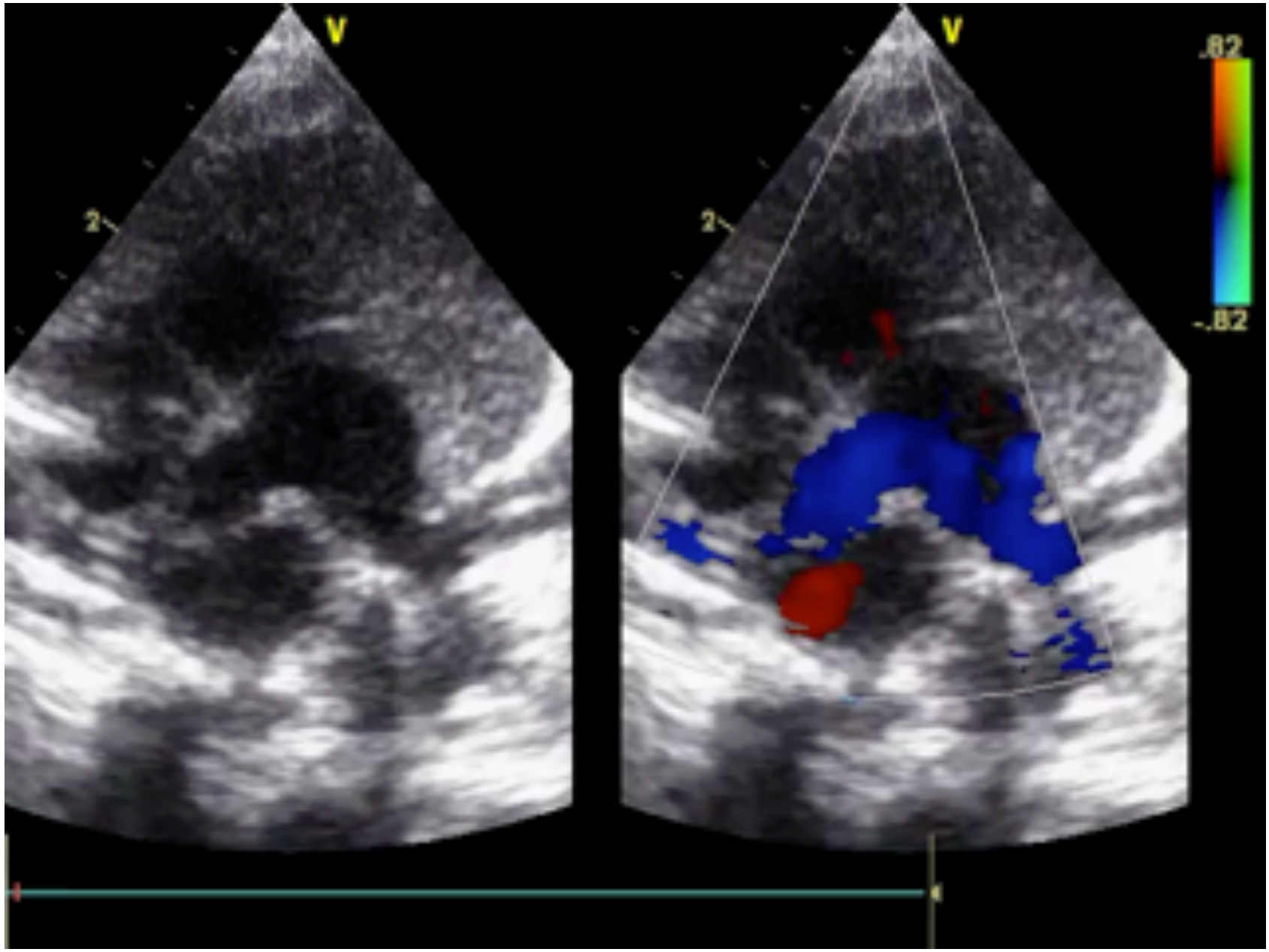
Canal artériel



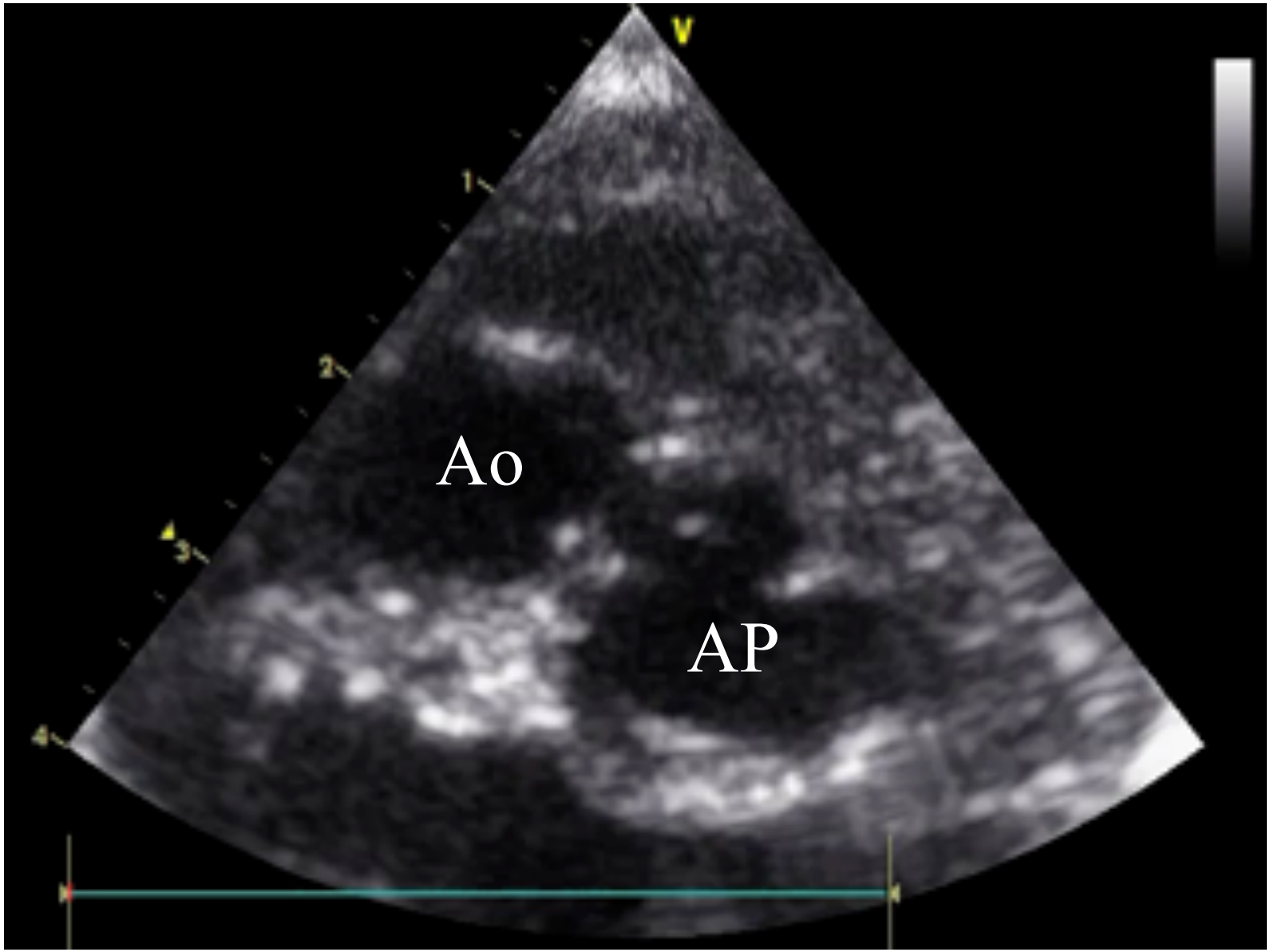
Congruence aortopulmonaire



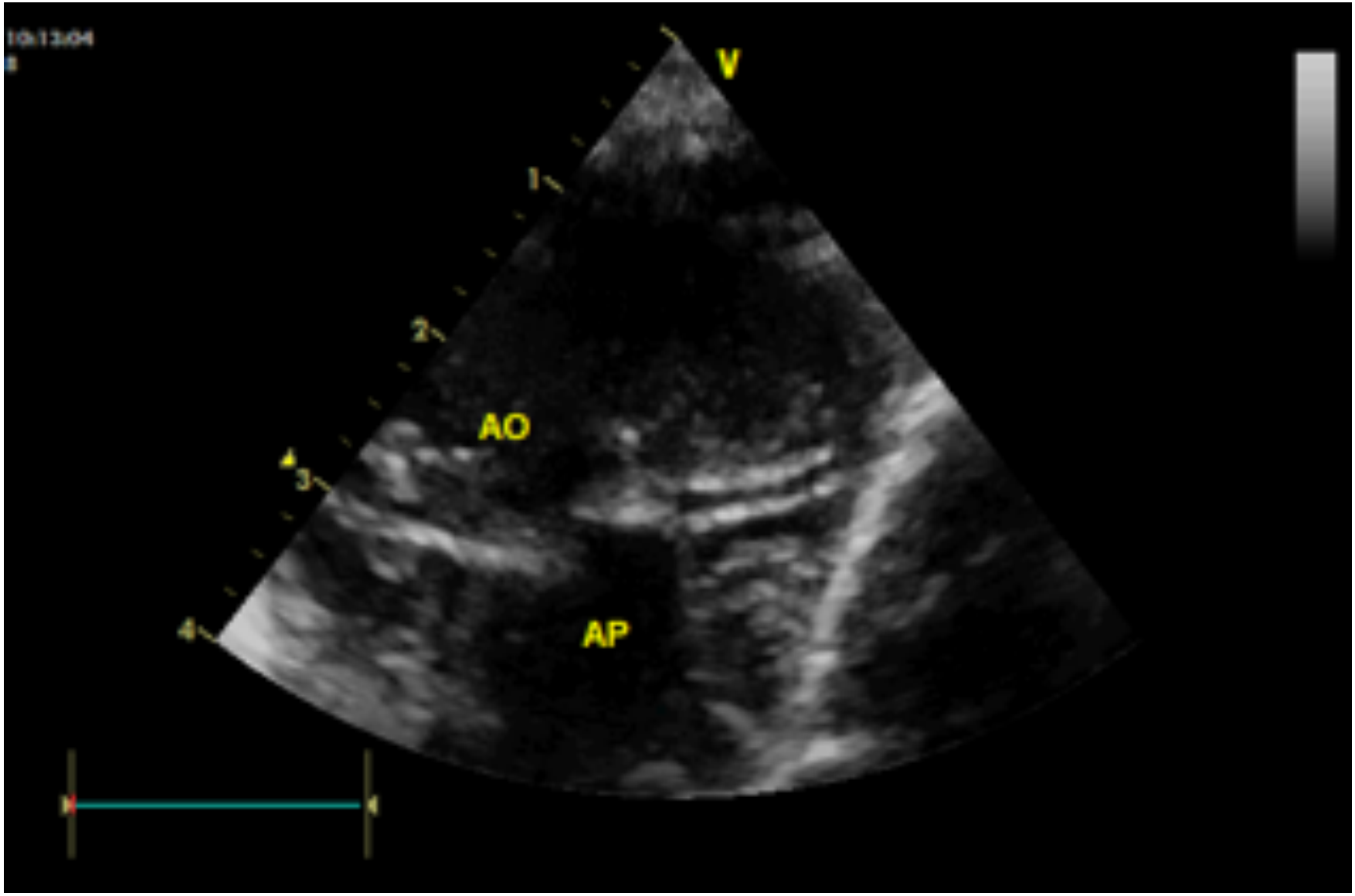
Position en D-TGV: aorte ant/AP post



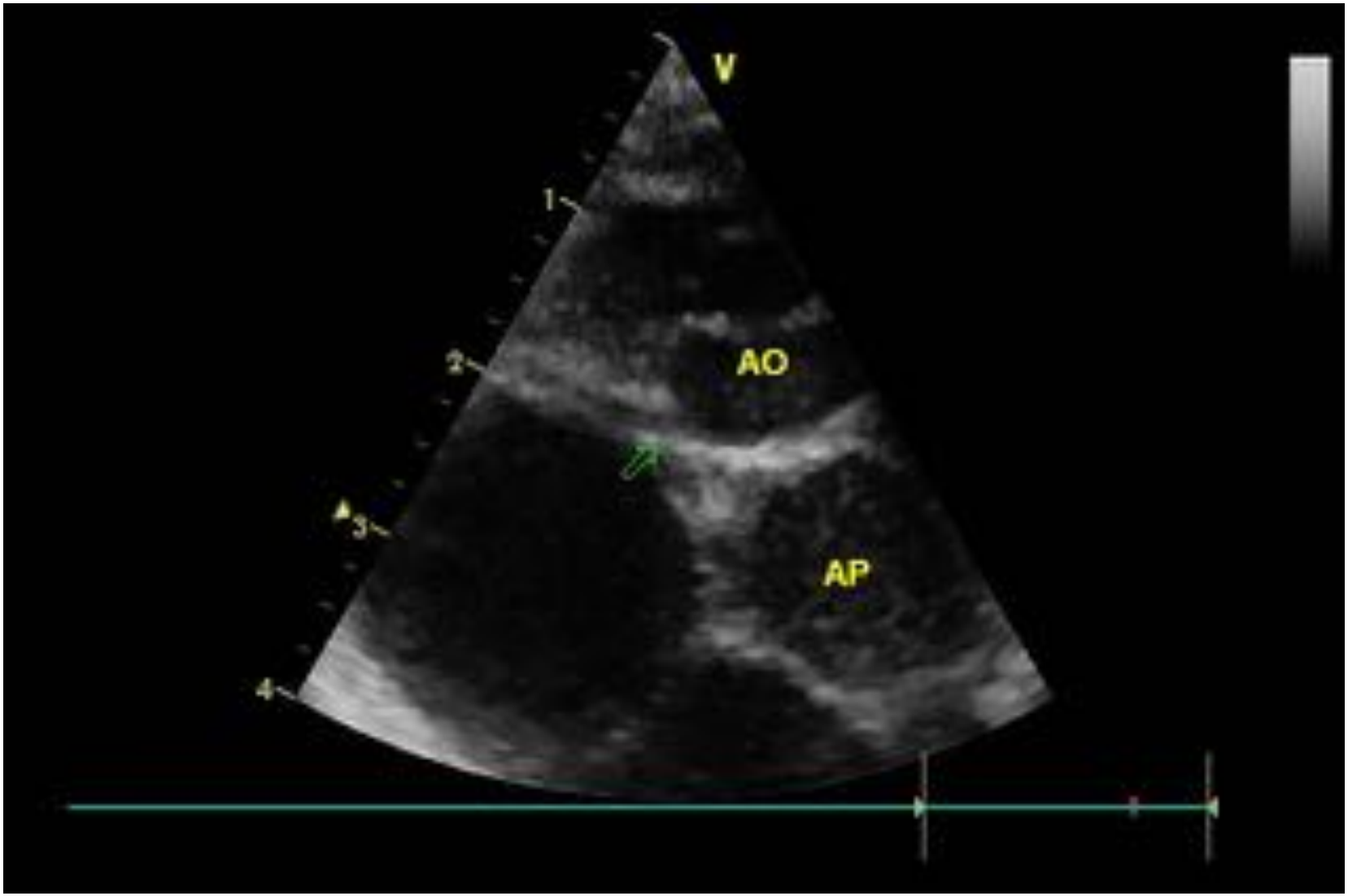
Alignement commissural



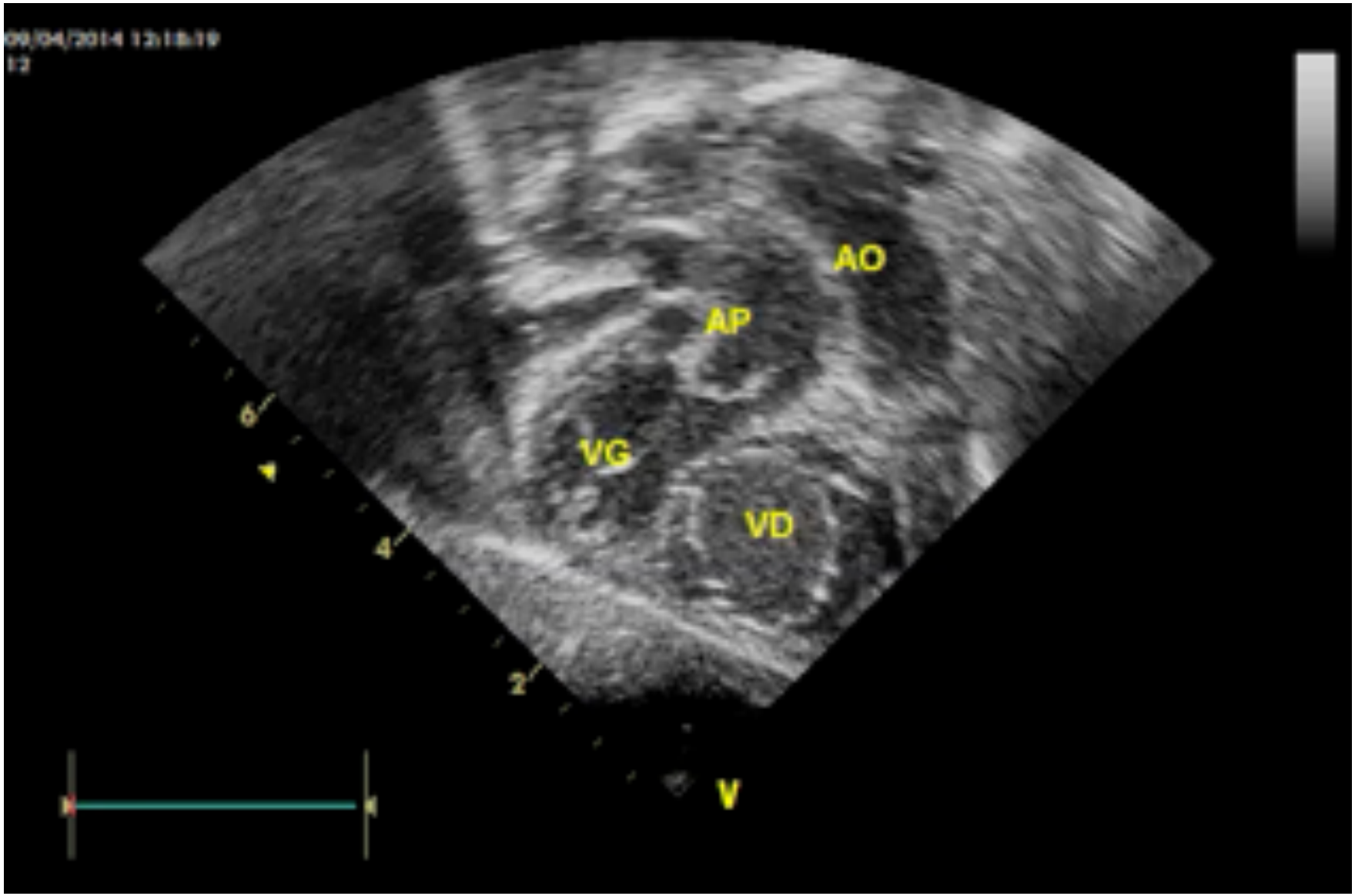
Coronaires Type A: CG



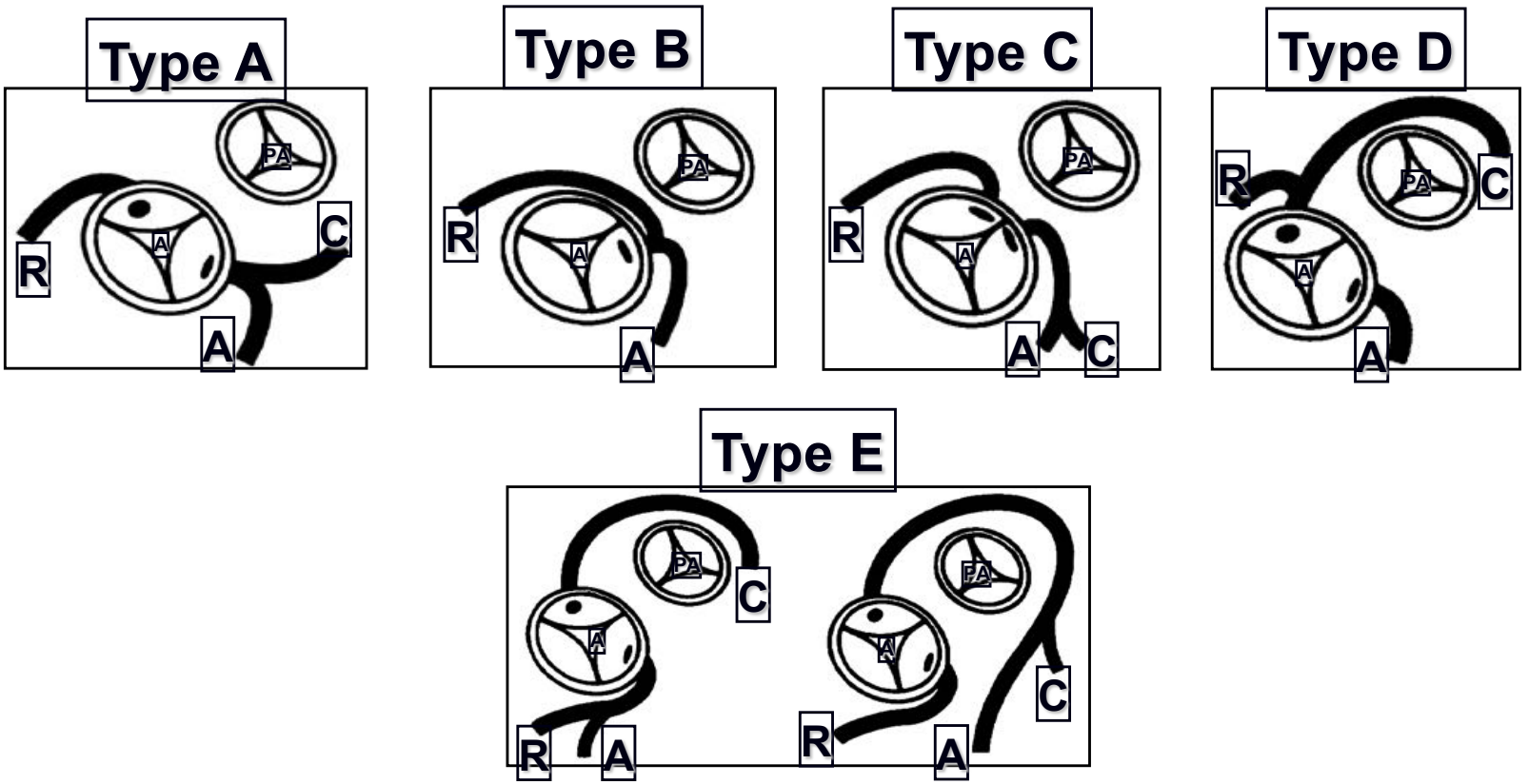
Coronaires Type A: CD

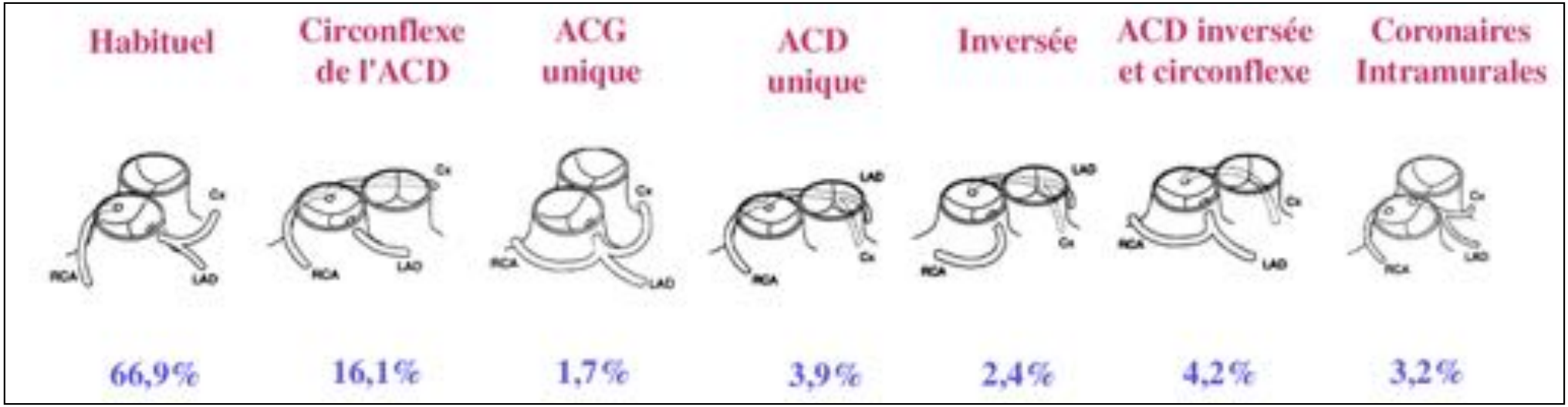


TGV avec CIV d'outlet



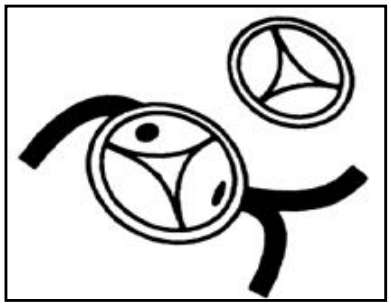
Classification de Yacoub





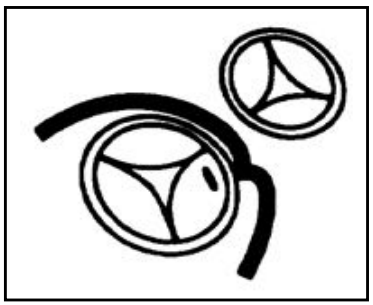
Anatomie coronaire

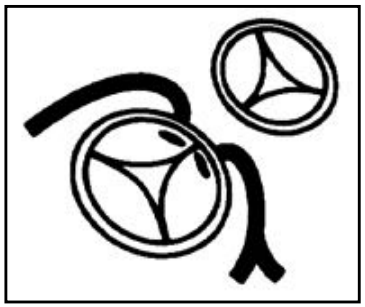
- « normale » : 60%



- boucle antérieure et/ou postérieure : 35%

- entre gros vaisseaux (intramurale) : 5%





Les lésions coronaires après switch artériel

Comment les détecter?

ECG et échographie (IM!!!)
Coroscanner si signe d'ischémie
Coronarographie si doute
Test d'ischémie (scintigraphie)
Coroscanner systématique à 5 ans

Que faire ?

Rappeler votre chirurgien…

Devenir

Mortalité postopératoire

- TGV simple: 1-5%
- TGV + CIV: 3-6%
- TGV+CIV+ Coa: 6-10%

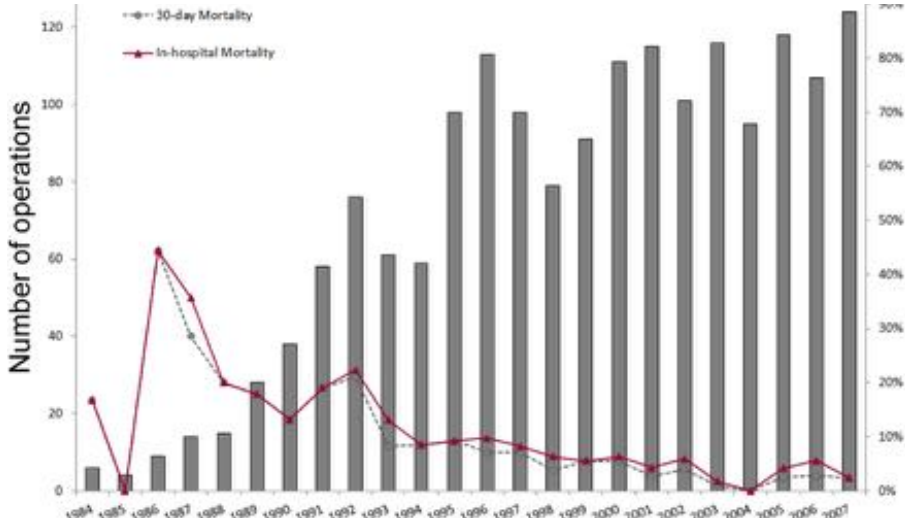
FdR de décès:
Anatomie coronaire complexe

Fricke et al. 2012
Villafane JACC 2014

TABLE 2

Outcome and predictors of early mortality [illegible] switch operation for TGA with IVS publications during the last decade.

Author, year	Inclusive years	n	% IVS	Early Survival For TGA IVS %	5 Year Survival%	10 Year Survival%	Coronary anatomic risk factors	Other predictors of early mortality
Sarris, 2006*	1998–2000	613	70	97	NA	NA	Single coronary (univariate analysis only)	Open sternum
Lalezari, 2011	1977–2007	332	60.8	88.6	85.8†	85.2†	Not a risk factor for early mortality	Technical problems with coronary transfer
Fricke, 2012	1983–2009	618	64	98.2	98	97	Not a risk factor for early mortality	Weight < 2.5 kg ECMO
Khairy, 2013	1983–1999	400	59.5	93.5†	NA	92.7†	Single right coronary artery	Postoperative heart failure
Cain, 2014	2000–2011	70	100	98.6	NA	NA	None identified	No predictors of early mortality but earlier repair < 4 days of age was associated with decrease resource utilization
Anderson, 2014	2003–2012	140	75	98.6	NA	NA	None identified	No predictors of early mortality but earlier repair < 4 days of age was associated with decrease resource utilization

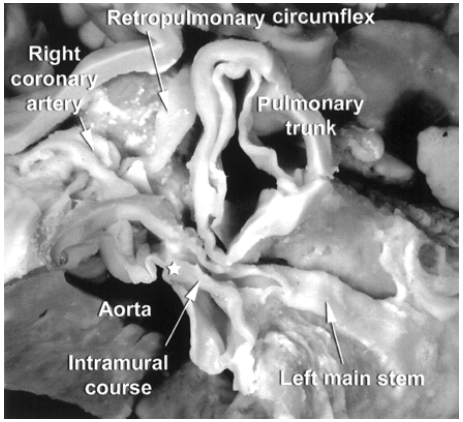


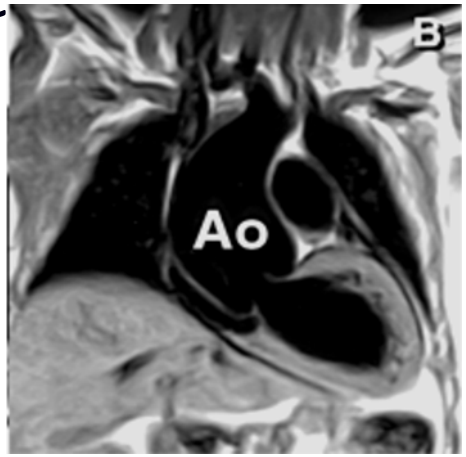
Villafane JACC 2014

Morbidité du switch artériel

- **Coronaires**
  - Coronaire intra-murale
  - Coronaire unique
- Voie droite (patch et Lecompte)
- IA sur néo-valve aortique, dilation Ao

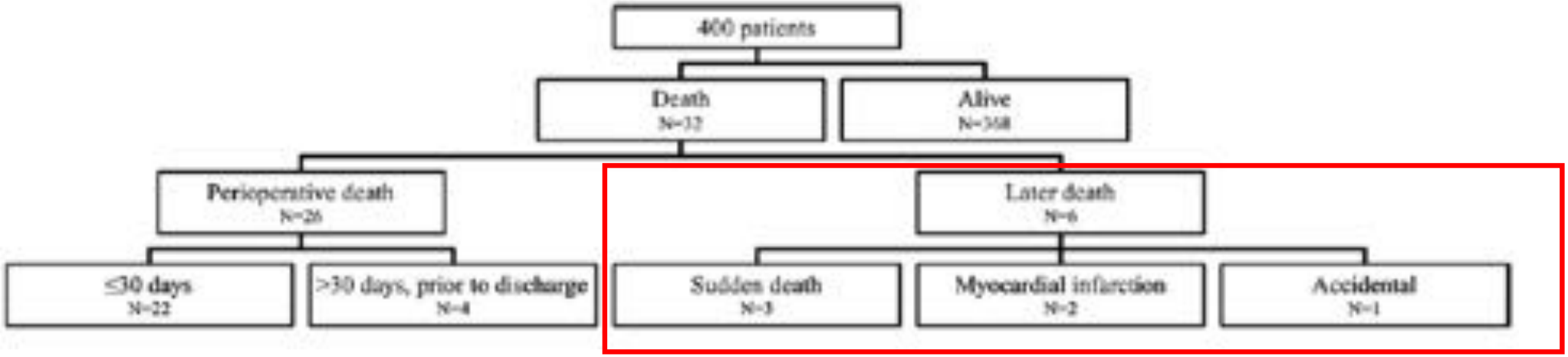
- **HTAP primitive:**
  - 1/200 TGV soit 100 fois plus fréquente que dans la population générale
  - Étiologie inconnue
  - Traitement médical……Potts





Cardiovascular Outcomes After the Arterial Switch Operation for D-Transposition of the Great Arteries

Paul Khairy, MD, PhD; Mathieu Clair, MD; Susan M. Fernandes, MHP, PA-C; Elizabeth D. Blume, MD; Andrew J. Powell, MD; Jane W. Newburger, MD, MPH; Michael J. Landzberg, MD; John E. Mayer Jr, MD



Switch artériel entre 1988-1999

Circulation. 2013;127:331-339

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Functional capacity	
New York Heart Association functional class, n (%)	
Class I	290 (97.3)
Class II	8 (2.7)
Class III or IV	0 (0)
Peak heart rate, bpm	180±18
Peak percent heart rate predicted, %	90.7±7.0
Heart rate reserve, bpm	101±21
Chronotropic index, %	83.9±10.9
Respiratory exchange ratio (RER)	1.16±0.09
Peak oxygen uptake, mL/kg/min	35.1±7.6
Percent maximum predicted peak oxygen uptake, %	86.1±15.1

Recognized comorbidities	
Coronary artery disease, n (%)	19 (5.2)
Hypertension, n (%)	12 (3.3)
Pulmonary stenosis	
Present, n (%)	171 (62.2)
Gradient in patients with pulmonary stenosis, mm Hg	25±17
At least moderate pulmonary stenosis, n (%)	28 (10.3)
Neoaortic stenosis	
Present, n (%)	37 (11.9)
Gradient in patients with neoaortic stenosis, mm Hg	19±7
At least moderate aortic stenosis, n (%)	10 (3.2)

Circulation. 2013;127:331-339

Associations Between Age at Arterial Switch Operation, Brain Growth, and Development in Infants With Transposition of the Great Arteries

Lim et al

Circulation. 2019;139:2728–2738. DOI: 10.1161/CIRCULATIONAHA.118.037495

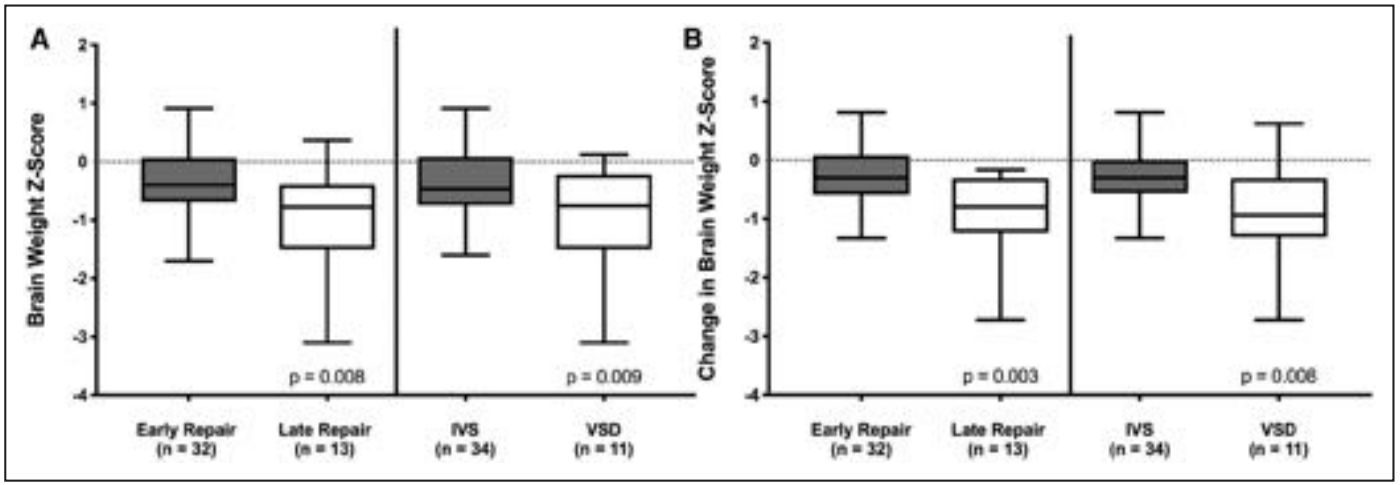

Figure 3. Brain weight *z* score comparisons.
A, Postoperative brain weight *z* score comparison in the early and late repair groups and in those with TGA/IVS and TGA/VSD. **B**, Change in brain weight *z* score between pre- and postoperative scans in early and late repair groups and in those with TGA/IVS and TGA/VSD. IVS indicates intact ventricular septum; TGA, transposition of the great arteries; and VSD, ventricular septal defect.

Table 3. Bayley-III Composite Scores

Composite Score	Early Repair	Late Repair	*P* Value
Cognitive	106±9	100±10	0.13
Language	98±13	90±20	0.50
Motor	104±13	102±12	0.76

Bayley-III scores in early vs late repair groups.

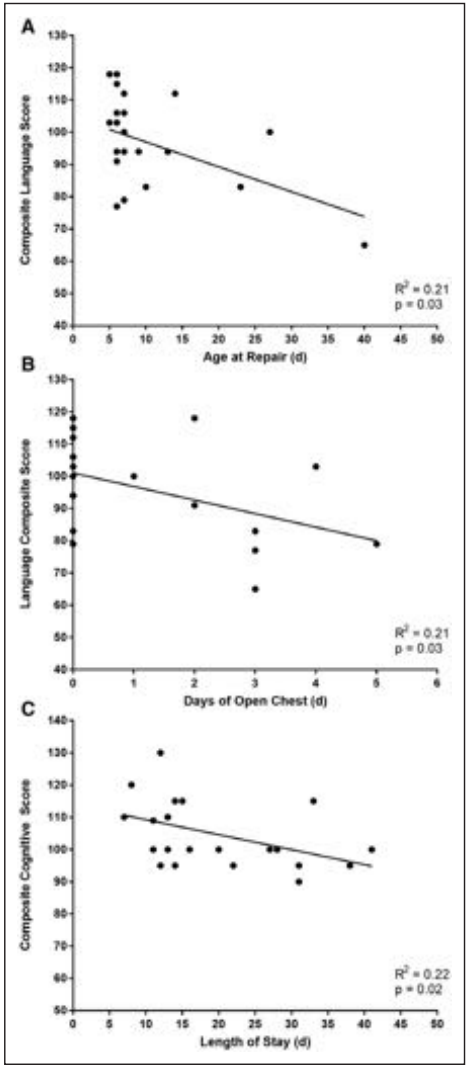

Figure 5. Linear regression of Bayley-III scores with clinical predictor variables.
A and **B**, Composite language scores were negatively correlated with age at surgery (**A**) and days of open chest (**B**). **C**, Composite cognitive scores were negatively correlated with length of stay.

Devenir neurodevelopmental

- TGV= une des cardiopathies le mieux exploré en neuropsychologie
- **Intelligence (sub)normale (IQ-testing)**
- Altération des fonctions cognitives supérieures: « theory of mind », visualisation temporo-spatiale etc.
- ADHS >> population de contrôle

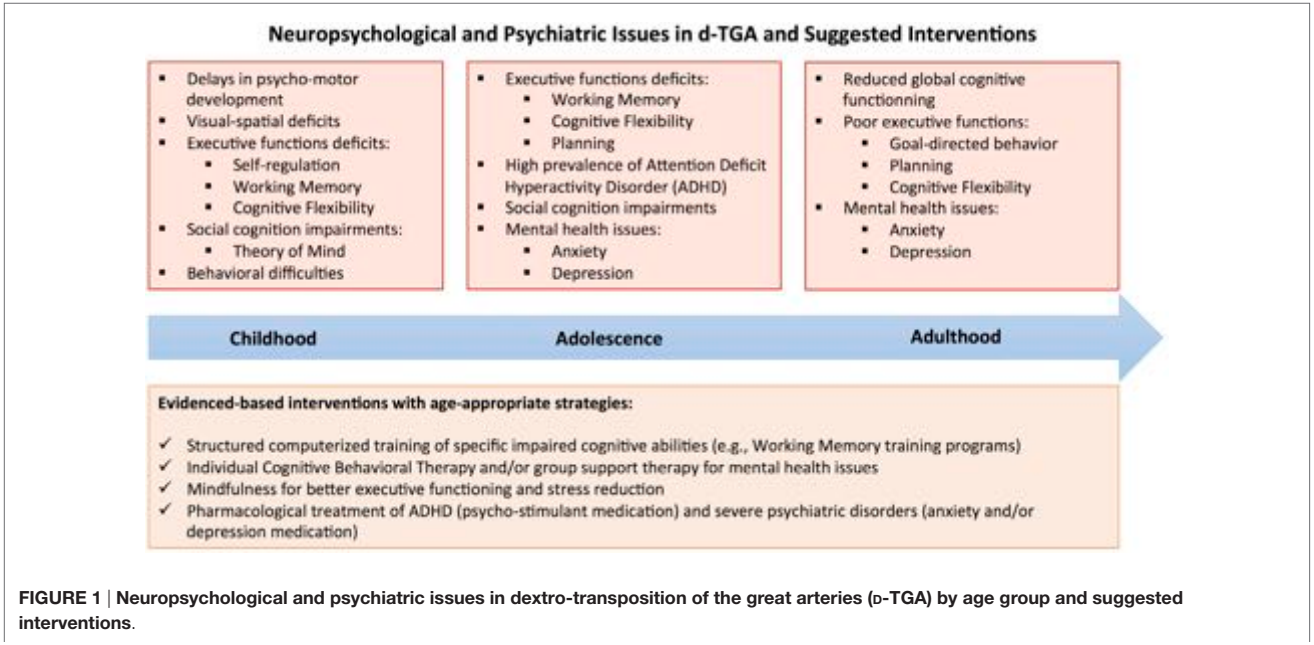

FIGURE 1 | **Neuropsychological and psychiatric issues in dextro-transposition of the great arteries (D-TGA) by age group and suggested interventions.**

Kalfa et al. 2017
Kasmi et al 2017
Calderon et al. 2010
Calderon et al. 2012
Calderon et al. 2014