Transpositions des gros vaisseaux

Dr Daniela Laux, UE3C-Paris et M3C-CCML

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

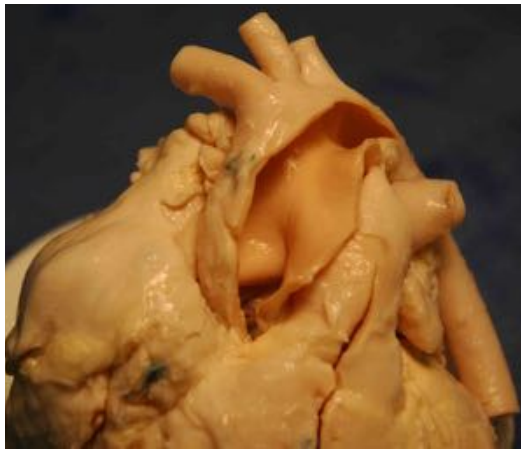
TGV - définition

- AP au dessus du ventricule G
- Aorte au dessus du ventricule D
- = discordance ventriculo-artérielle

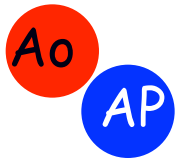
- La TGV n'est qu'une des malpositions vasculaires qui incluent:

  TGV, VDDI, VGDI, malposition anatomiquement corrigée

VX NORMOPOSES

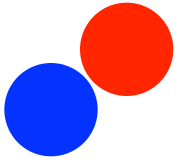


S = solitus

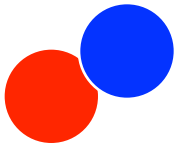


D L

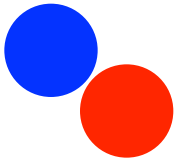
I = inversus



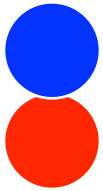
VX TRANSPOSES



D-TGV



L-TGV

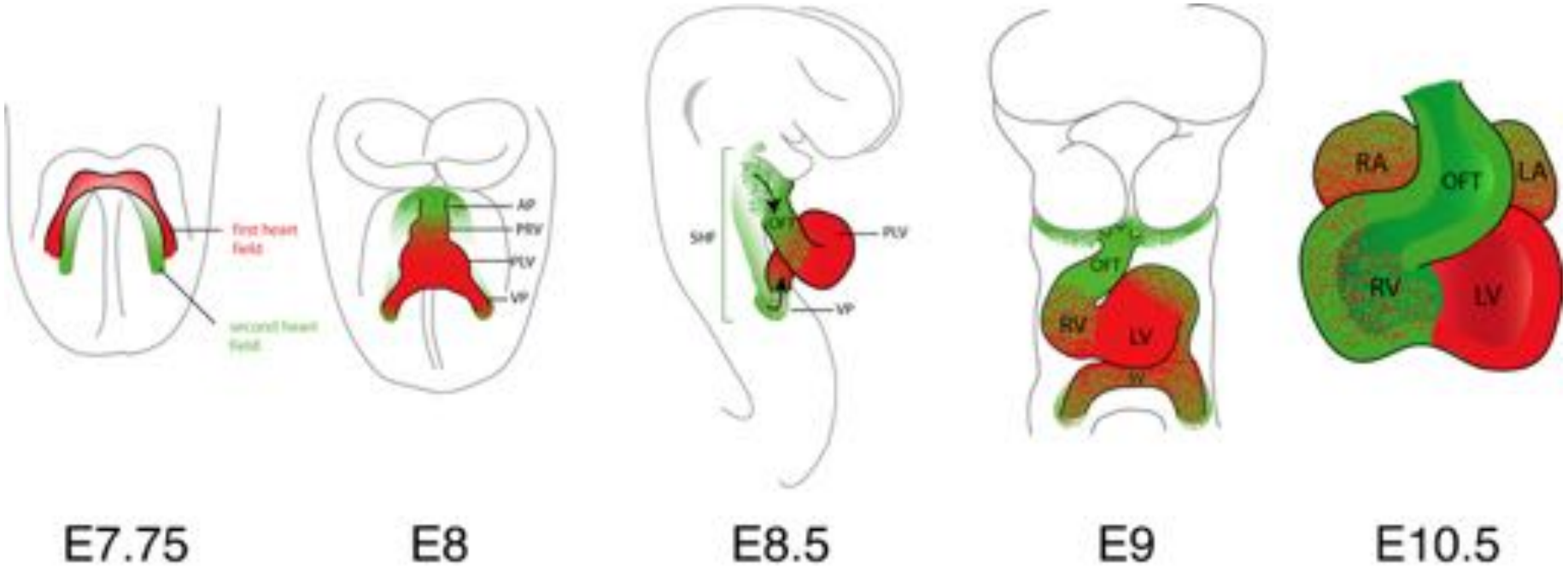


A = antéropostérieur

Courtesy L.Houyel

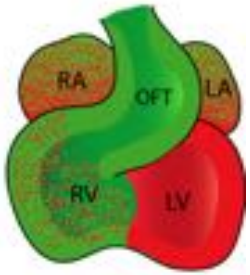
Rappel embryologique

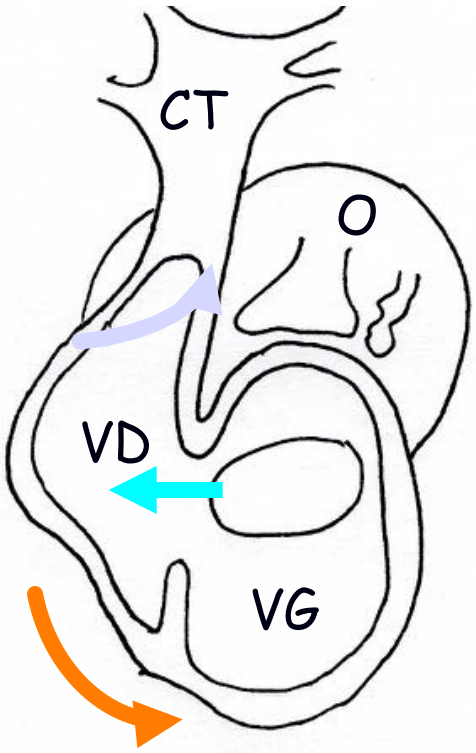
Nouveau concept de morphogenèse : second champ cardiaque



Premier champ cardiaque (cardiac crescent) = 1st lineage

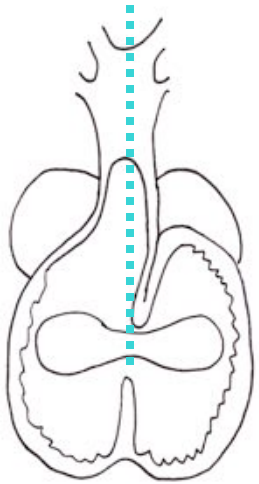
Second champ cardiaque (anterior heart field) = 2nd lineage

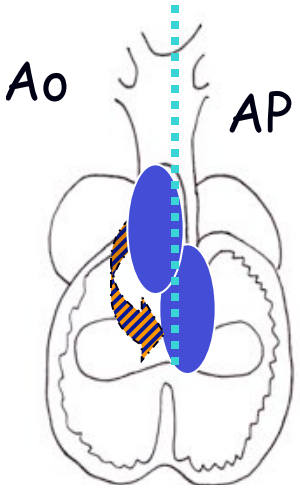


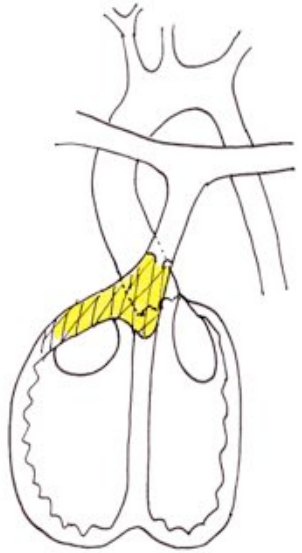


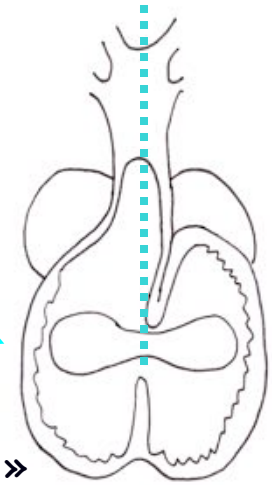
Rotation « normale »

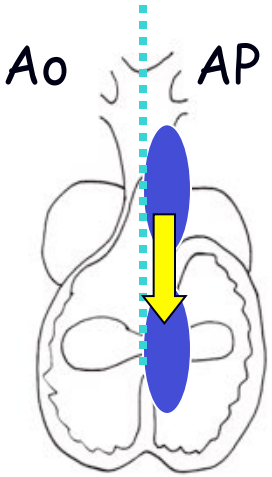
Rotation « inversée »

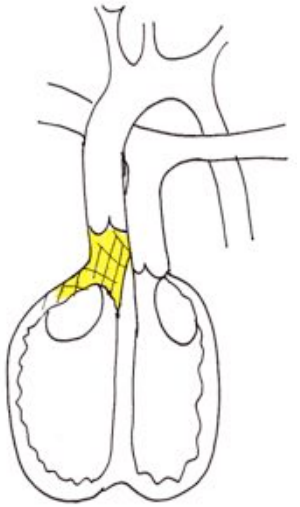












Cœur normal

TGV

Early looping — Convergence — Rotation

Courtesy L.Houyel

Types anatomiques: simples et complexes

- TGV simple – 60%: pas d'autre lésion associée
- TGV avec 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

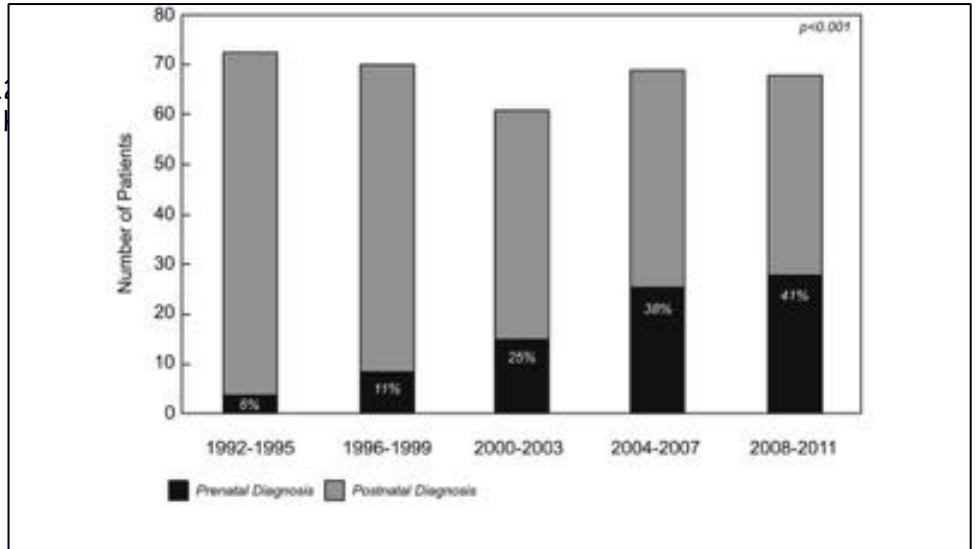
Diagnostic prénatal

Etude EPICARD

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

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



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

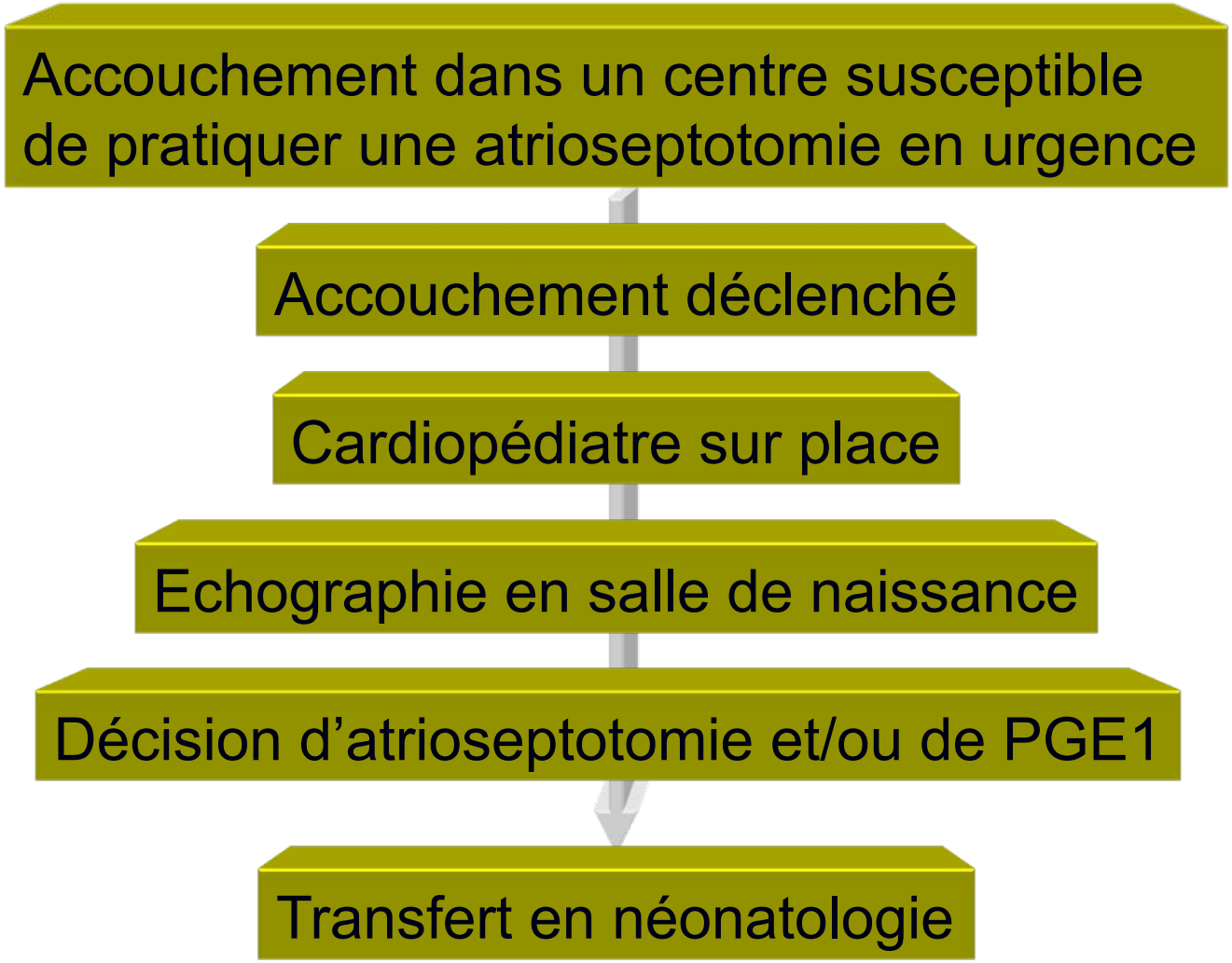
Damien Bonnet, MD; Anna Coltri, MD; Gianfranco Butera, MD; Laurent Fermont, MD;
, MD

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 (%).

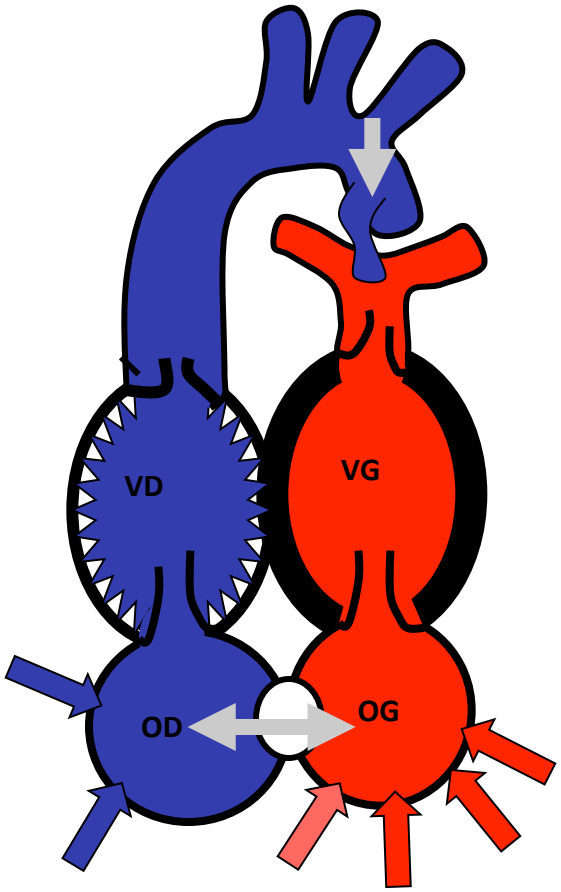
Circulation. 1999;99:916-918


Accouchement dans un centre susceptible de pratiquer une atrioseptotomie en urgence
Accouchement déclenché
Cardiopédiatre sur place
Echographie en salle de naissance
Décision d'atrioseptotomie et/ou de PGE1
Transfert en néonatologie

Clinique postnatal

Cyanose réfractaire en salle de naissance sans détresse respiratoire

Pathophysiologie TGV simple

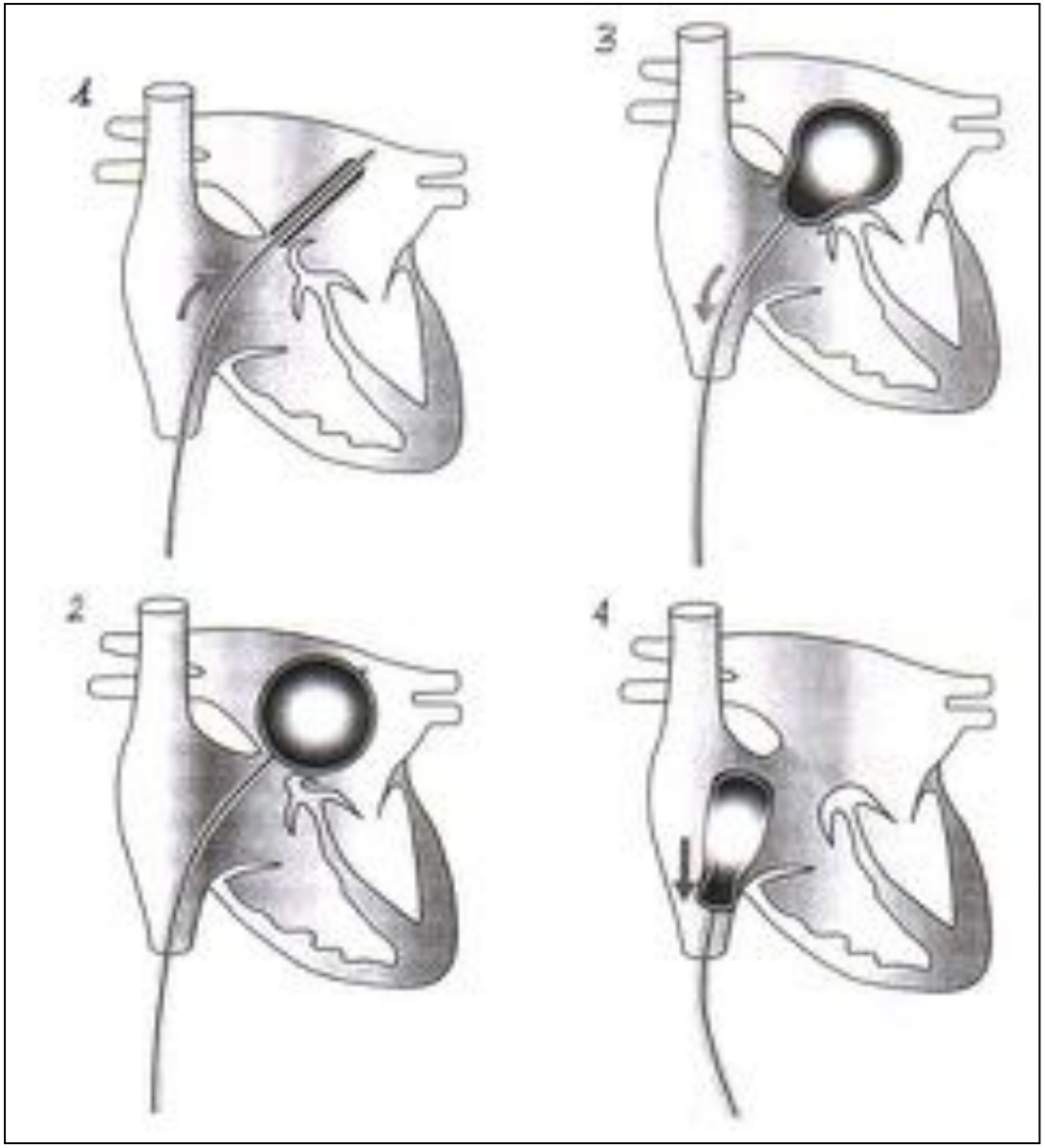


- Cardiopathie cyanogène car aorte nait du VD !
- **Circulation en parallèle**
- CA et FOP obligatoire pour un mixing efficace
- Risque d'OAP si FOP restrictif
- CA shunte Ao-AP à cause des résistances vasculaires
- FOP shunte G-D à cause des compliances ventriculaire

Prise en charge médicale néonatale de la TGV

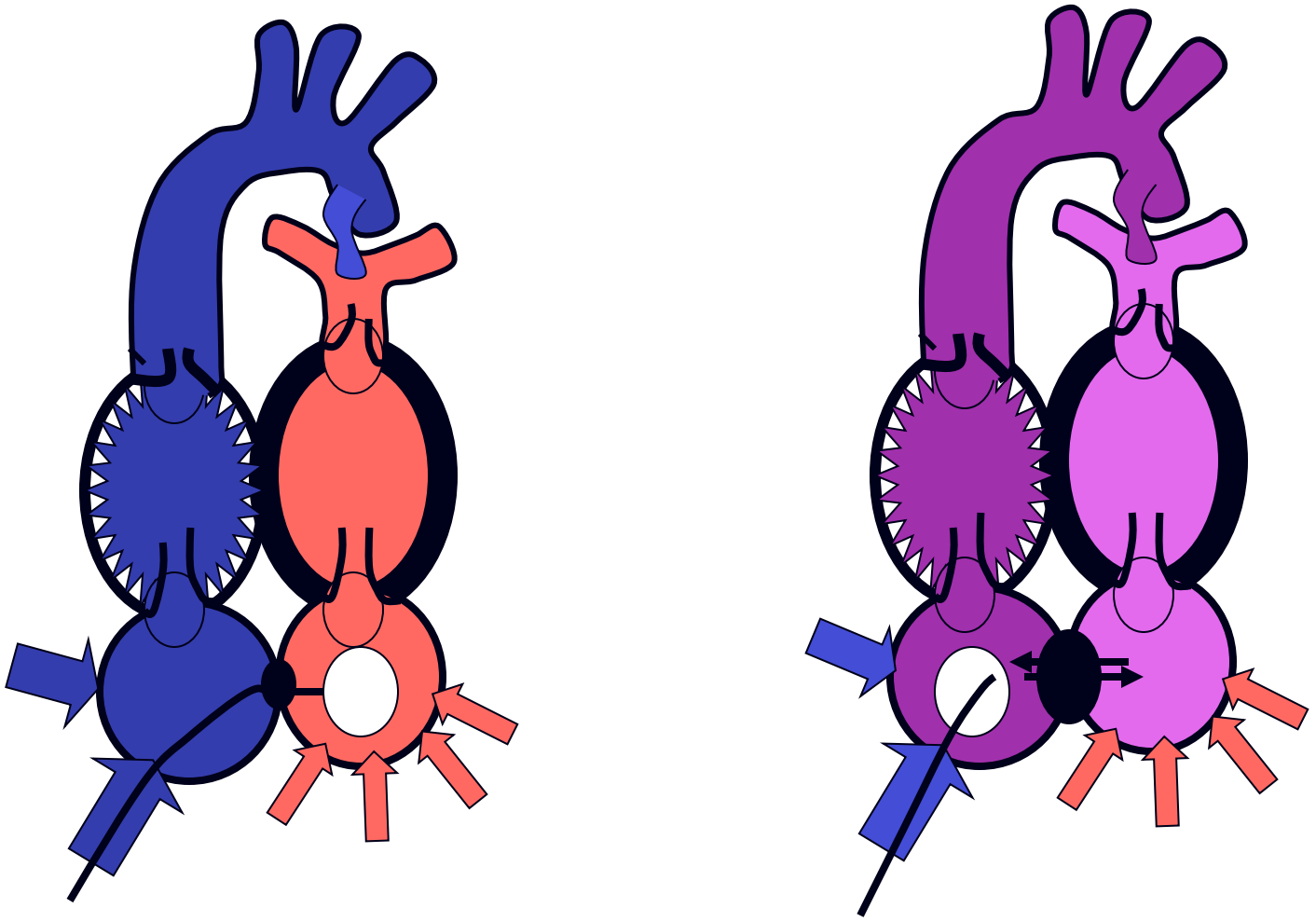
- **Rashkind**
  - Mixing
  - Dé-précharge le VG
- **PGE1**
  - Effets secondaires
  - Précharge le VG

Manœuvre de Rashkind (1966)



Manœuvre de Rashkind : sous scopie ou écho

Rashkind: effet hémodynamique immédiat

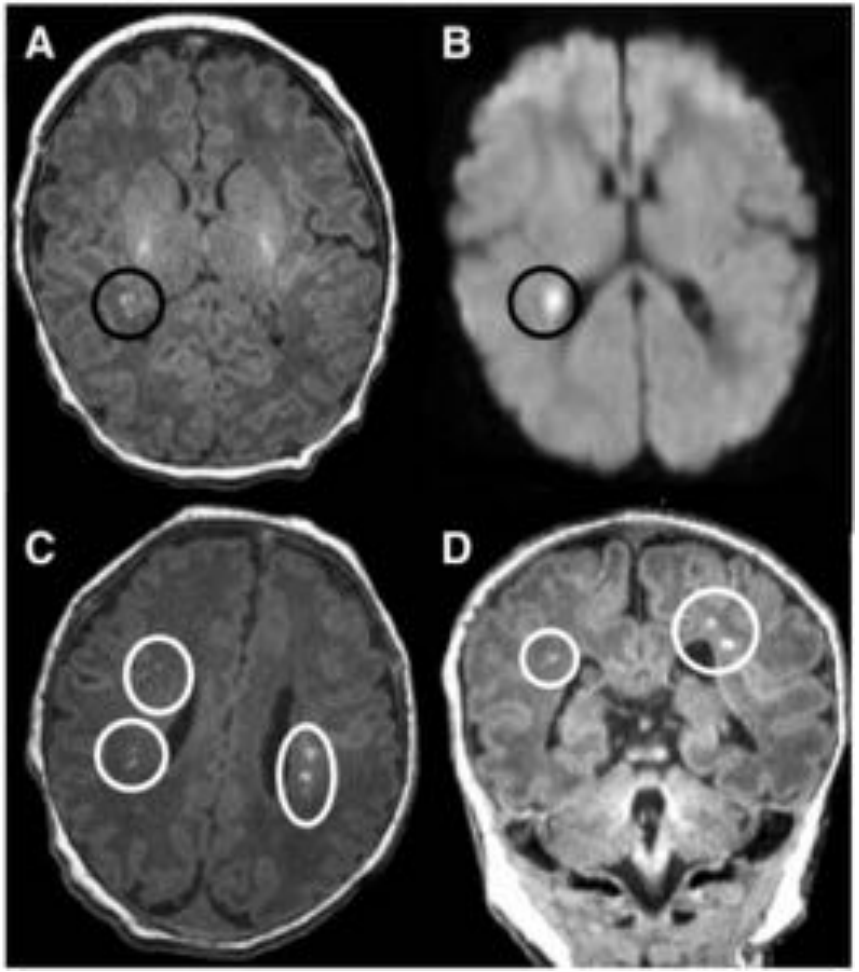


Procédure réalisée dans 70% à la naissance ou dans les premiers jours

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



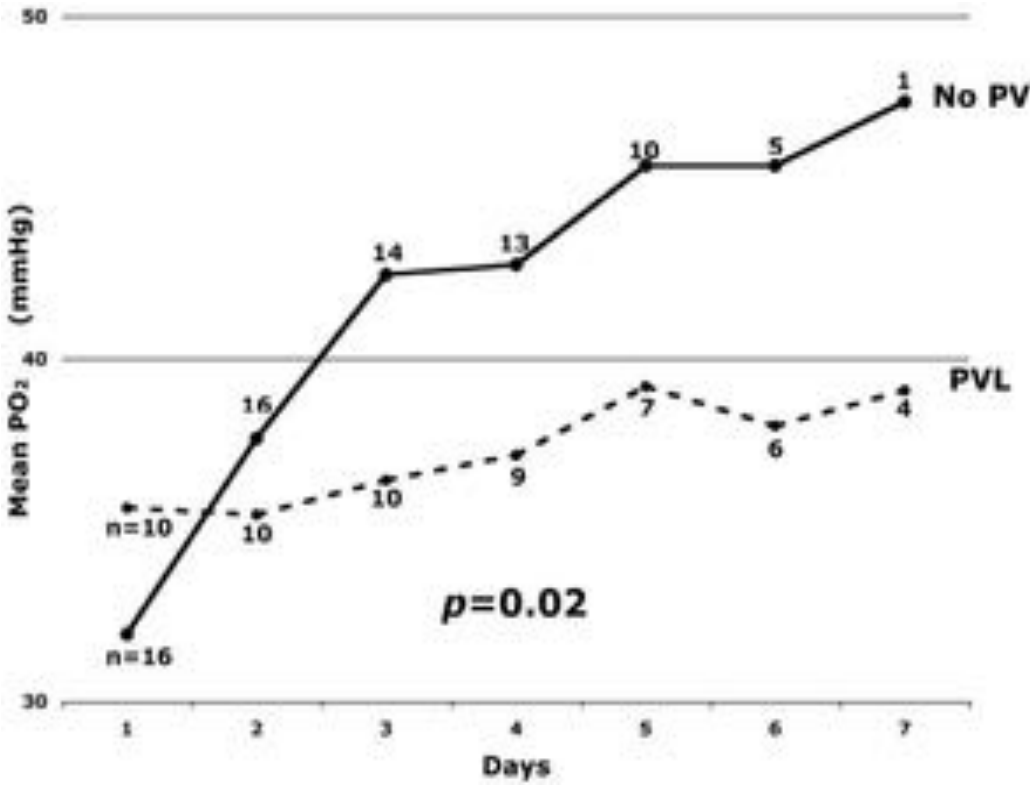


Figure 3.
A daily mean P_{O_2} was calculated for the PVL and no-PVL groups. Repeated-measures ANOVA demonstrated a significant difference in mean daily P_{O_2} between the PVL group (dashed line) and the no-PVL group (solid line; P=0.02). The PVL group never achieved a mean daily P_{O_2} >40 mm Hg.

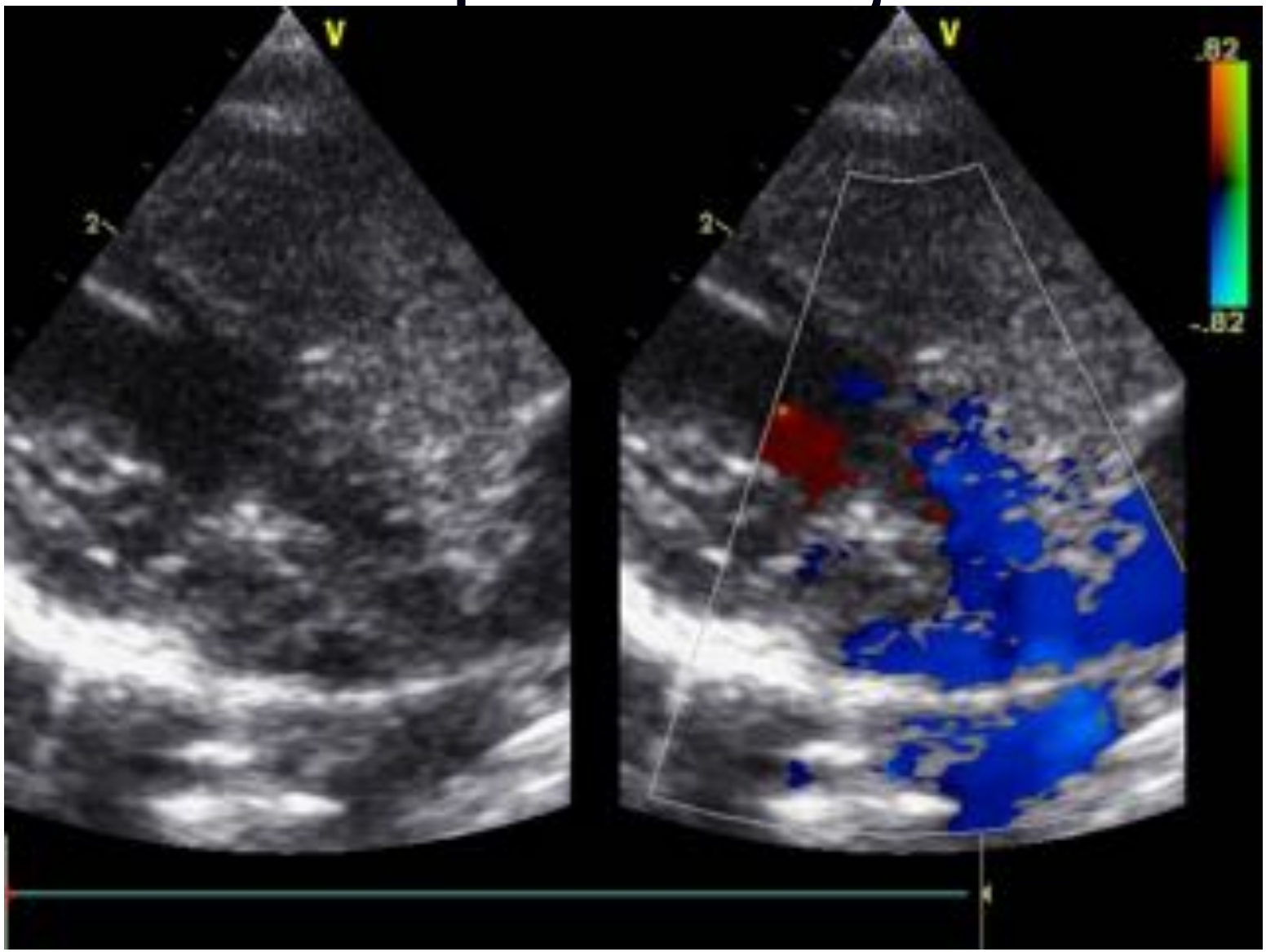
Check liste écho pré op

- CIA large ou restrictive et CA ouvert ou fermé

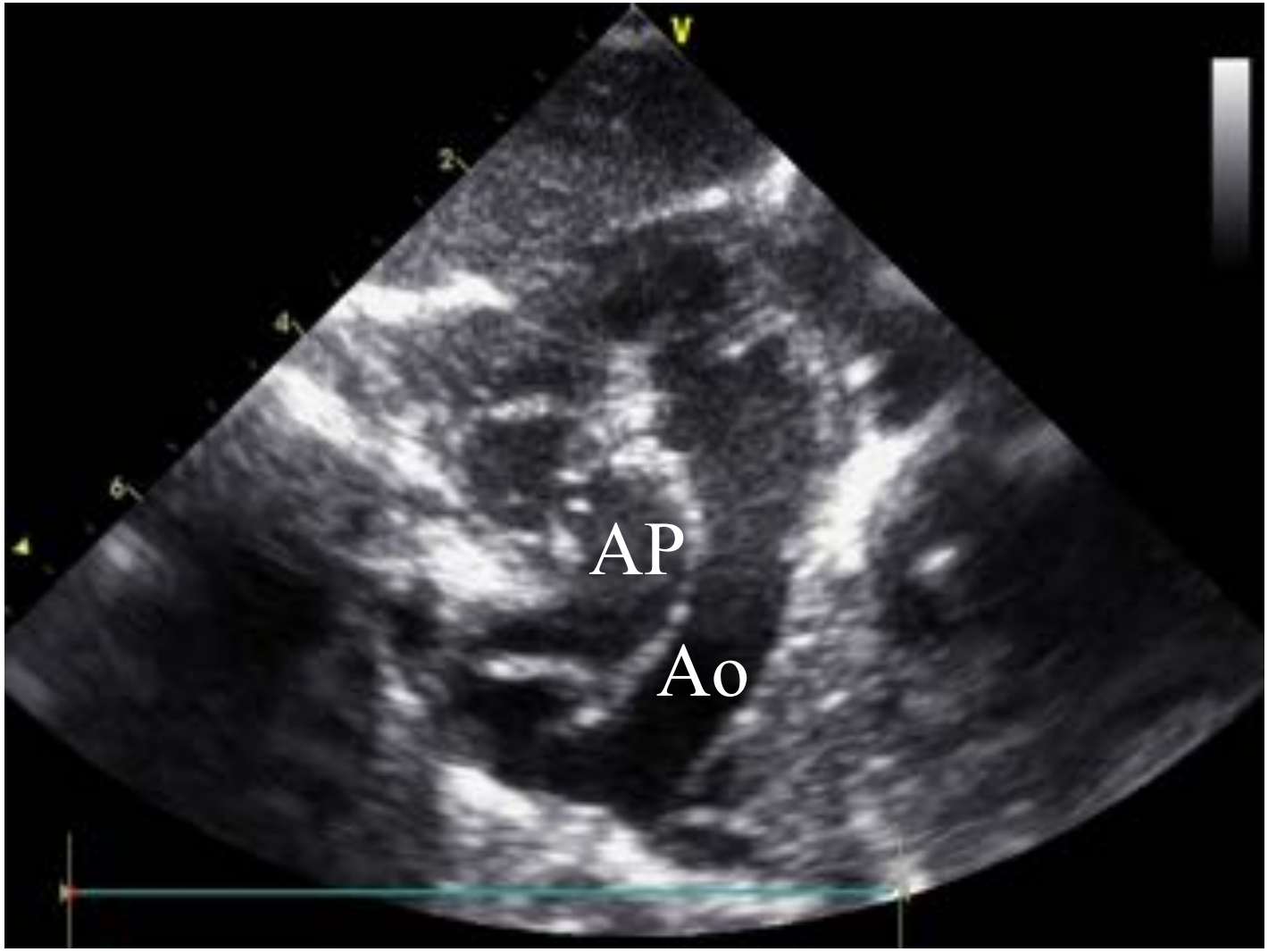
- **Equilibre des ventricules**
  - Petit VD : risque de coarctation
  - Petit VG : vérifier la voie pulmonaire
- **Anatomie de la valve mitrale**
  - Fente non commissurale

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

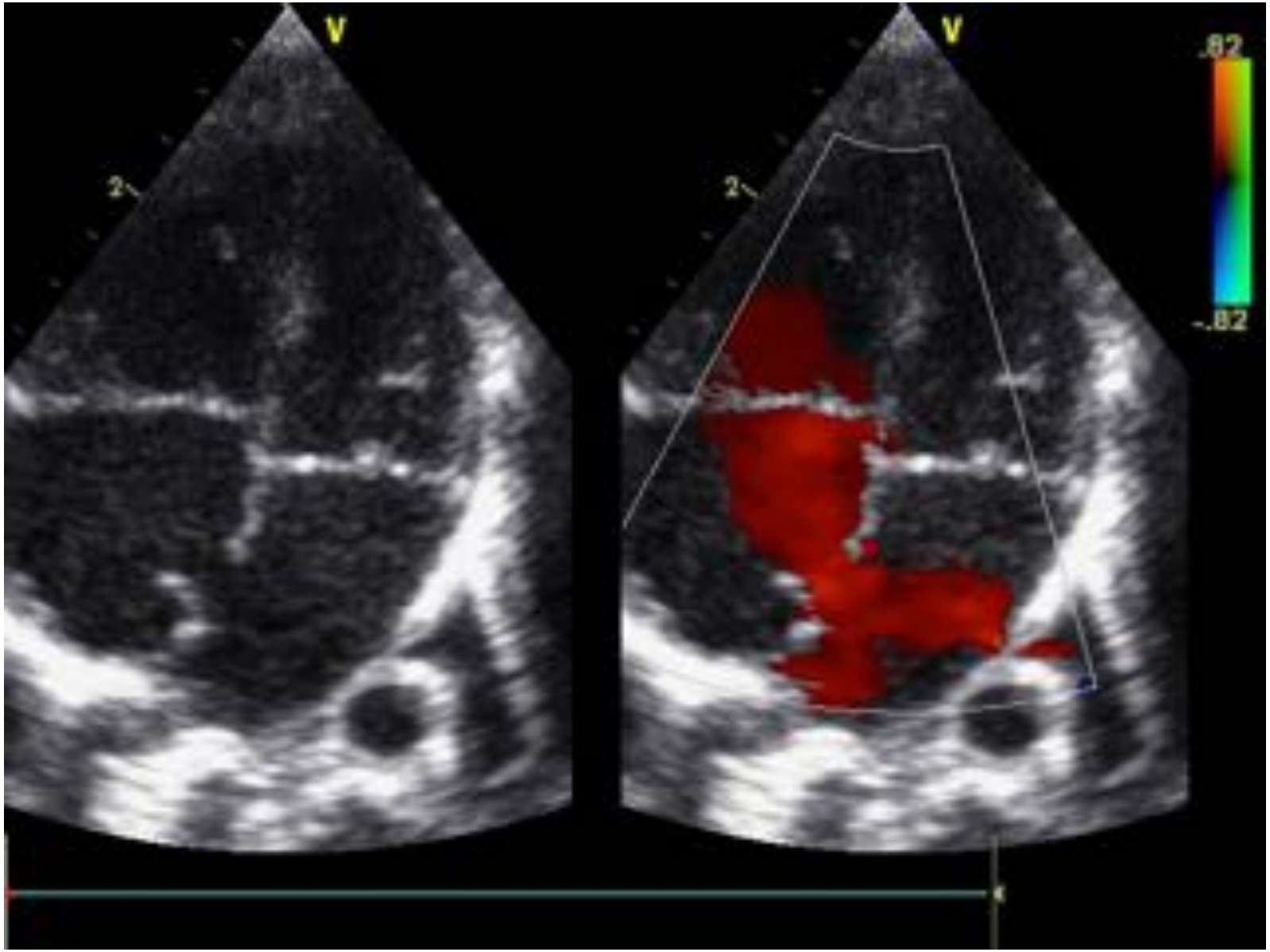
Vaisseaux parallels: grand axe



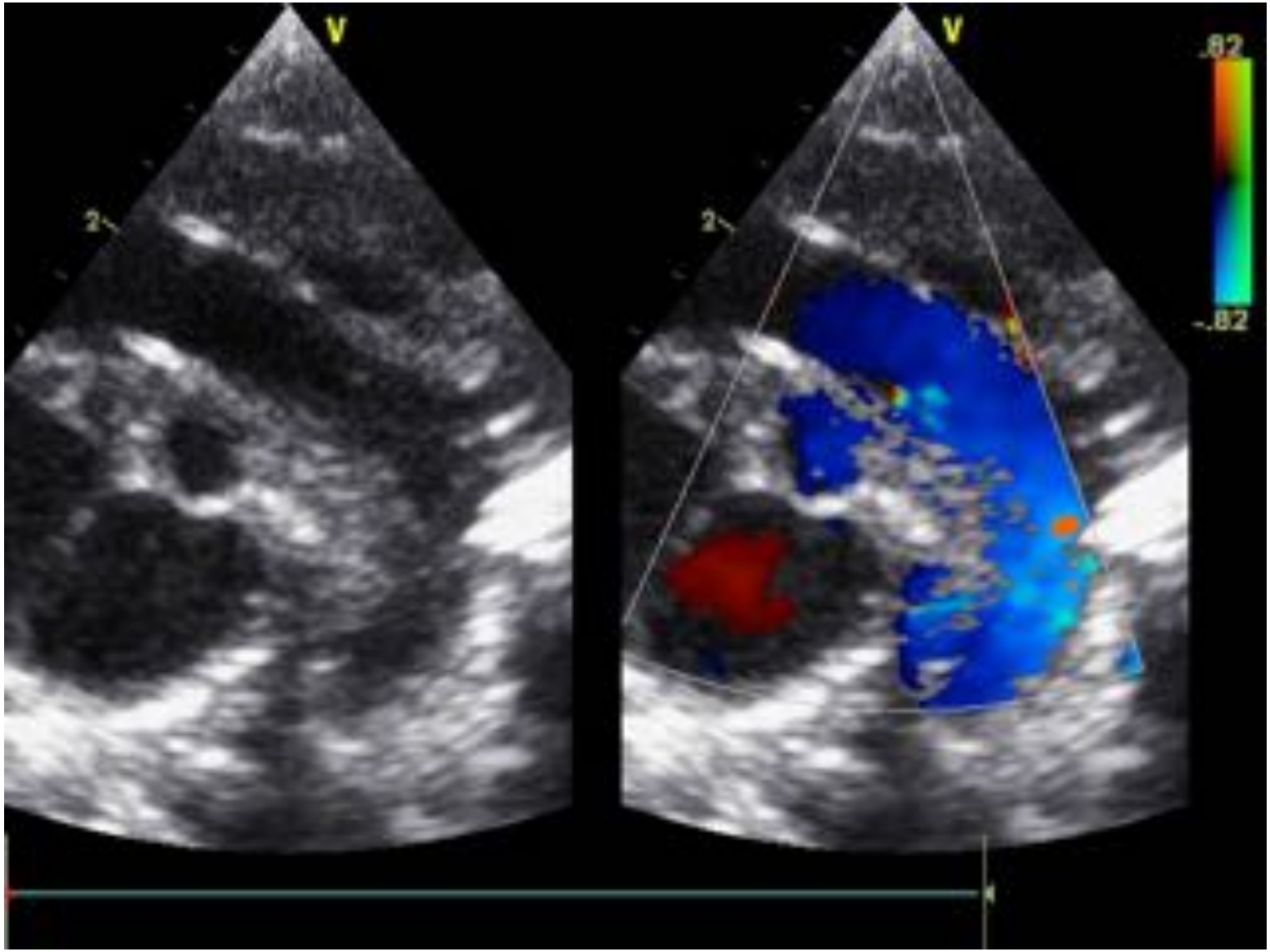
TGV en souscostal: vaisseaux parallèles



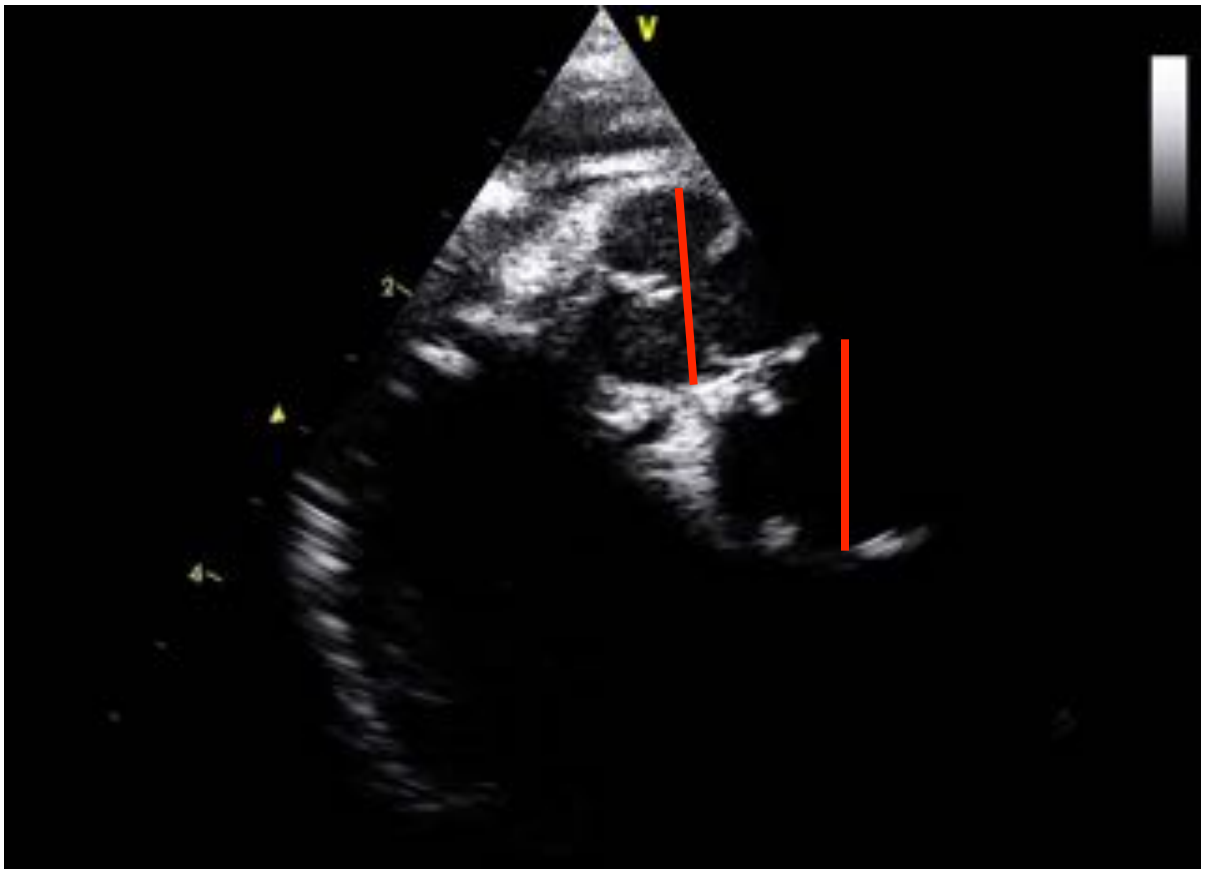
CIA post-RSK



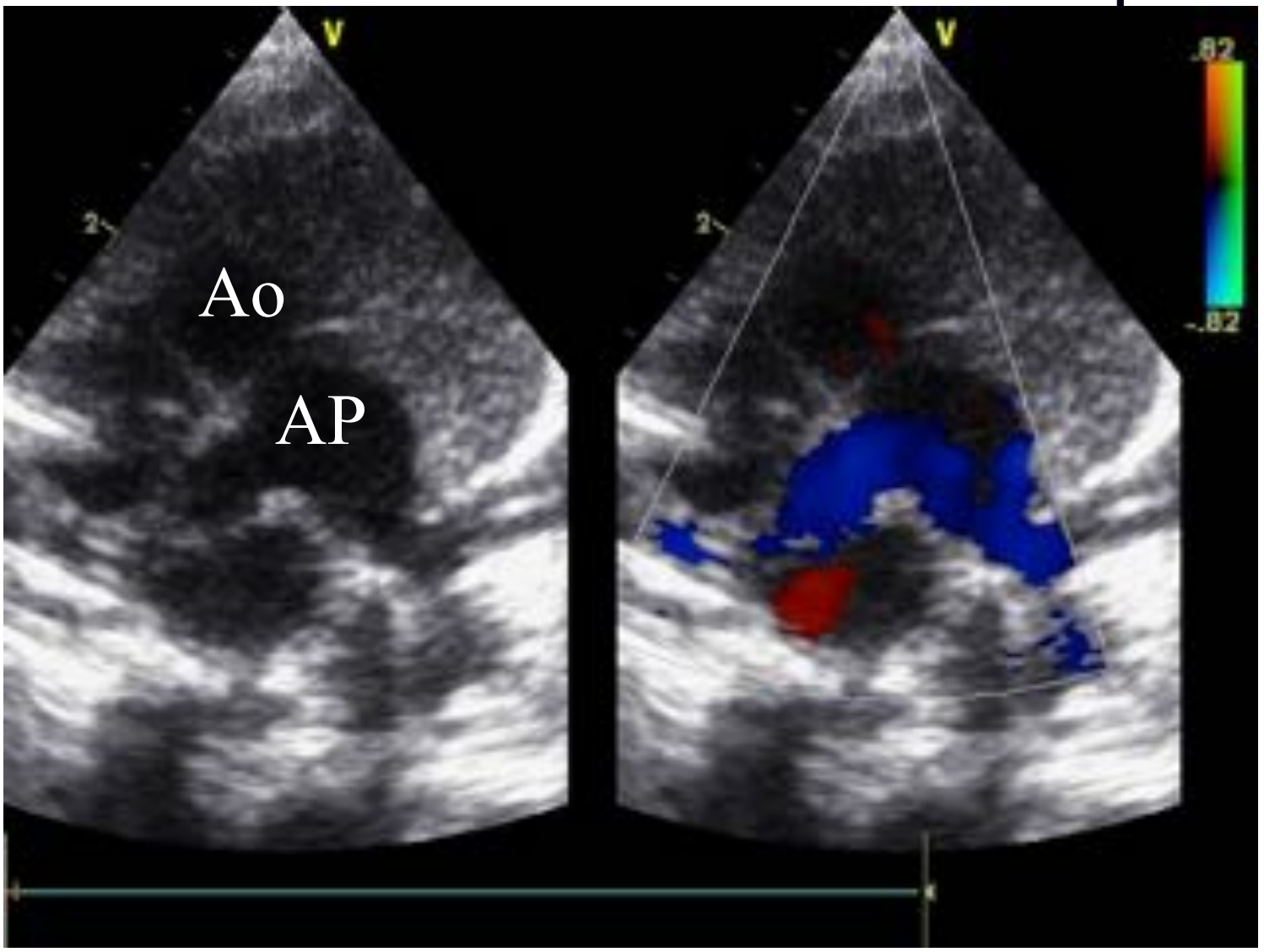
Canal artériel



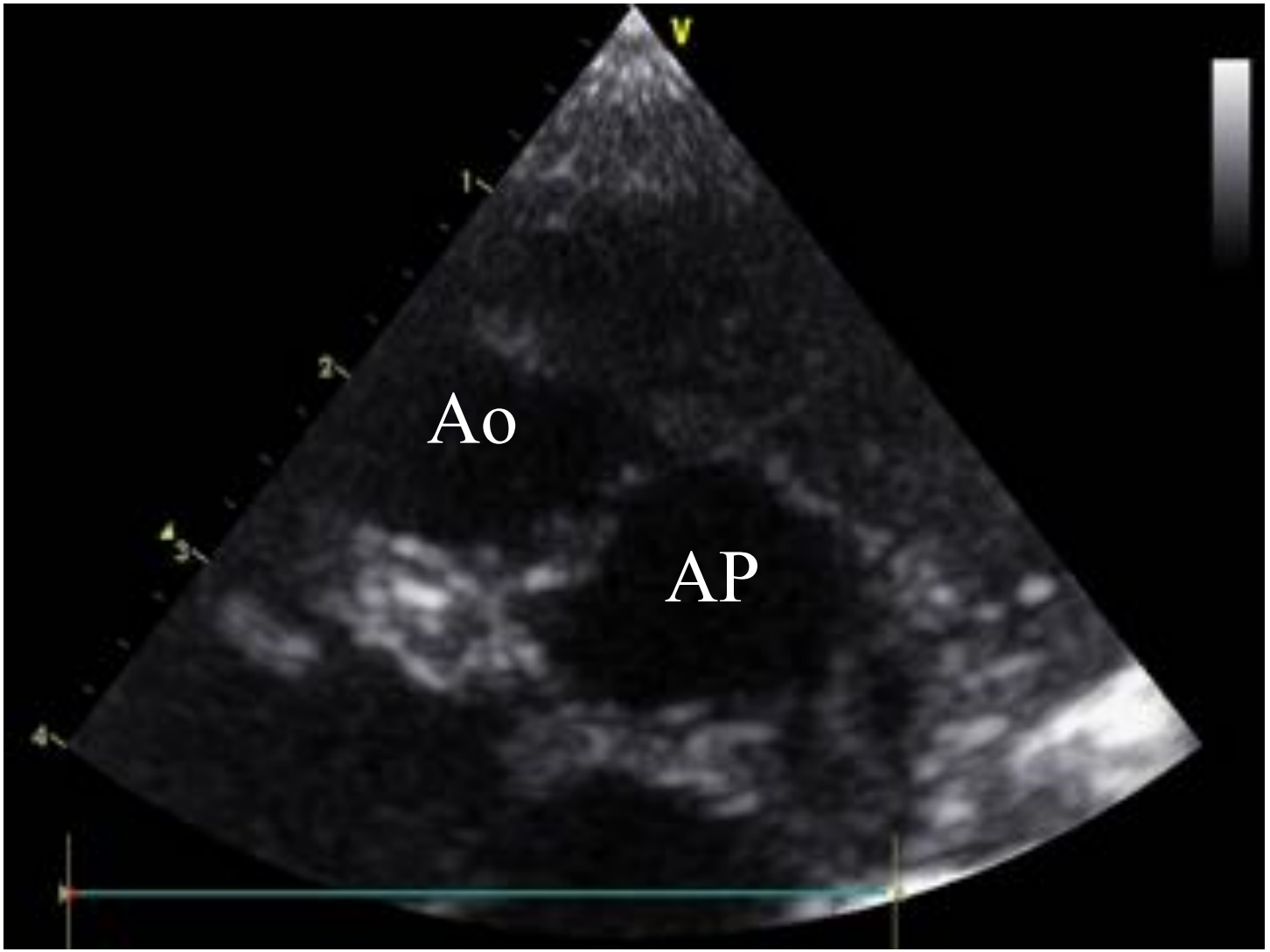
Congruence aortopulmonaire



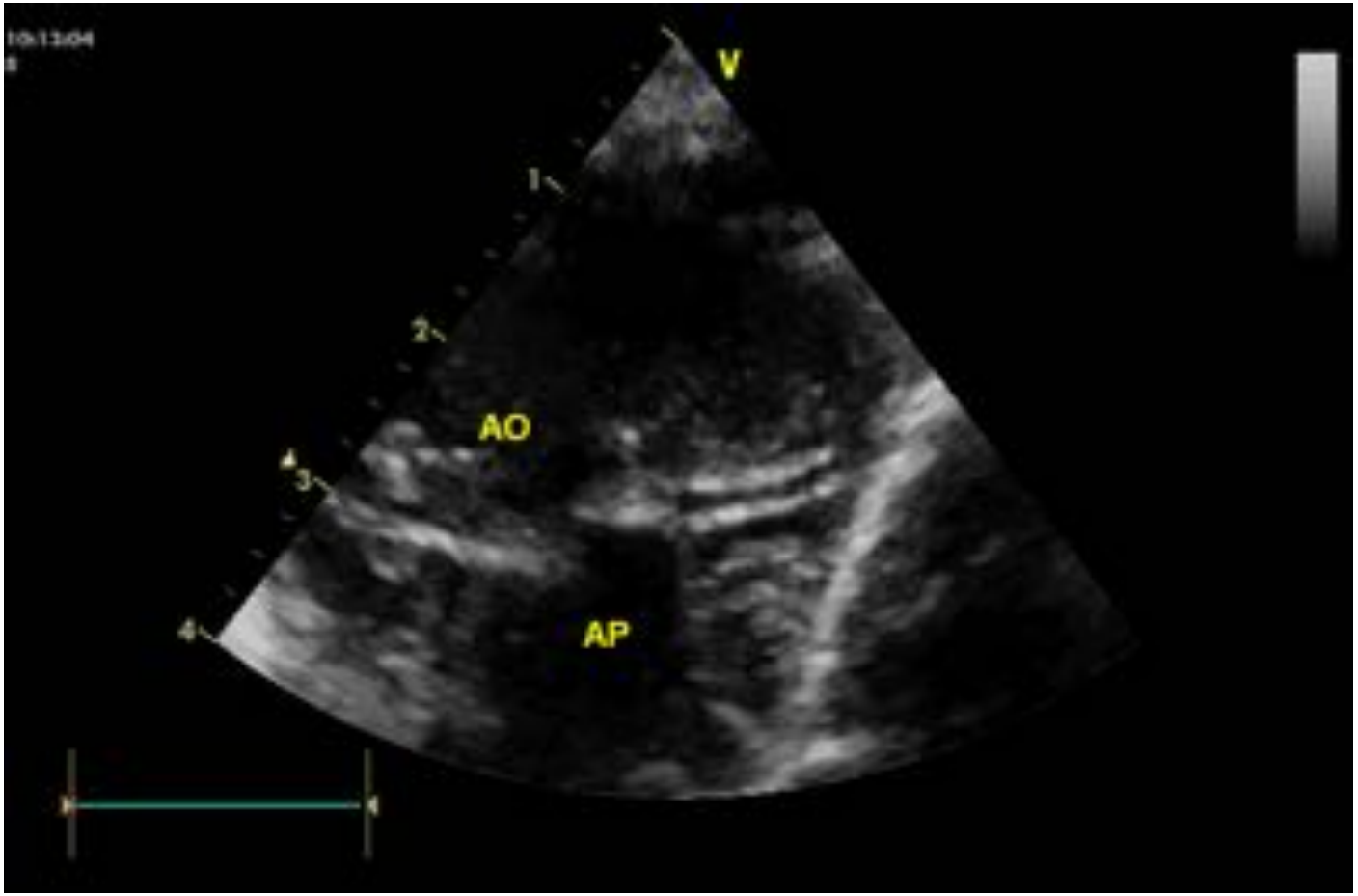
Position en D-TGV: aorte ant/AP post



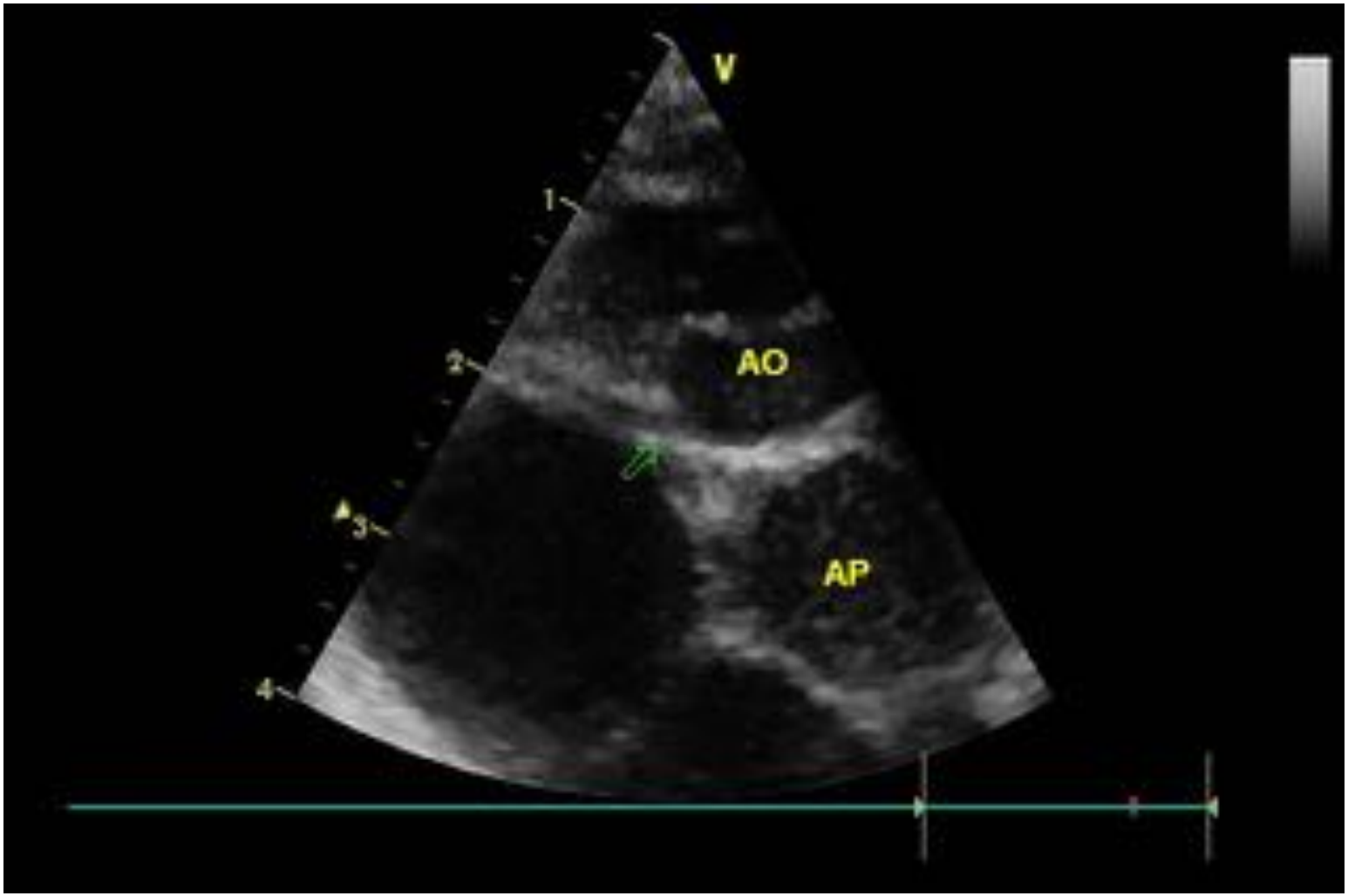
Alignement commissural



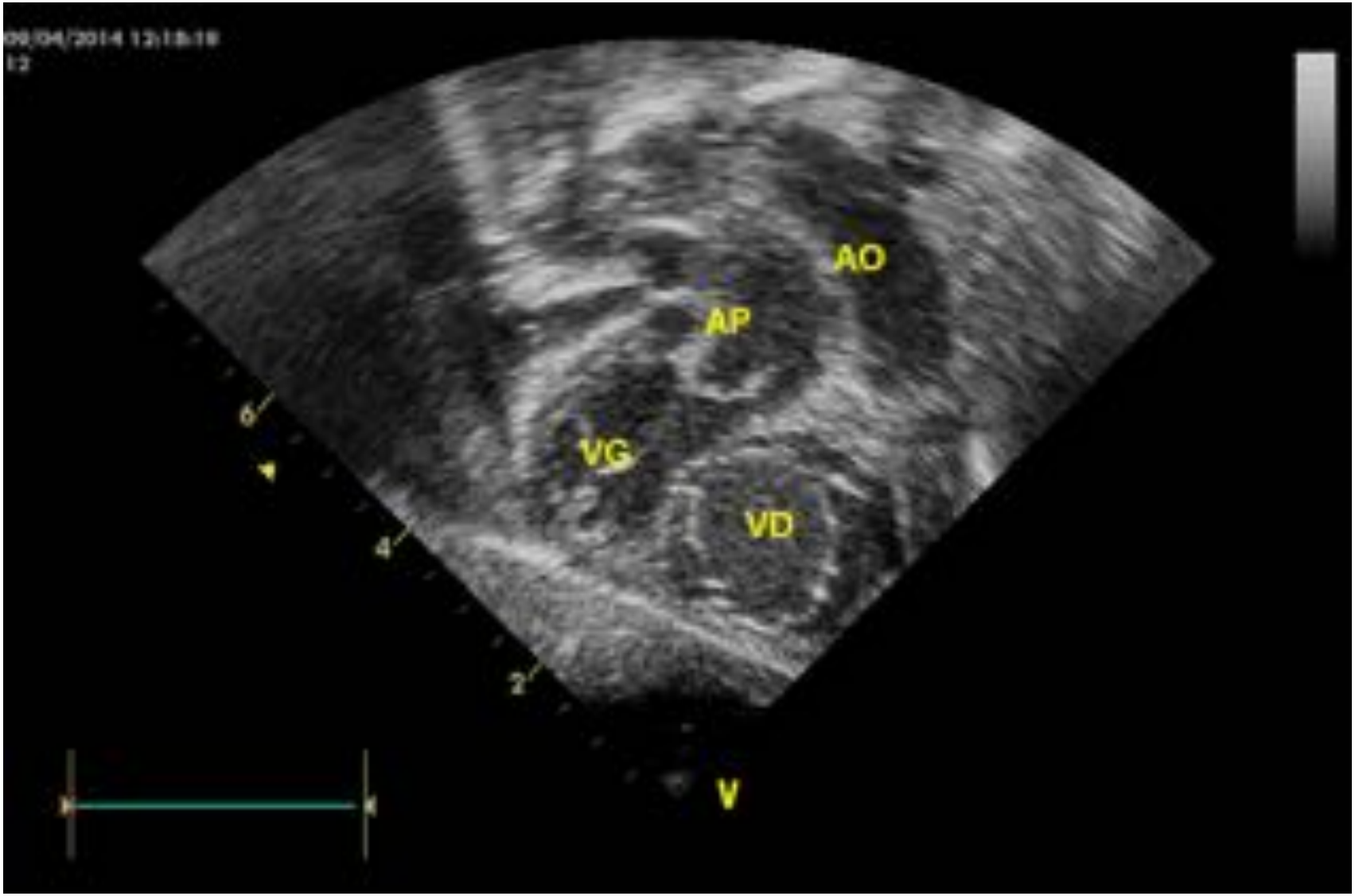
Coronaires Type A: CG



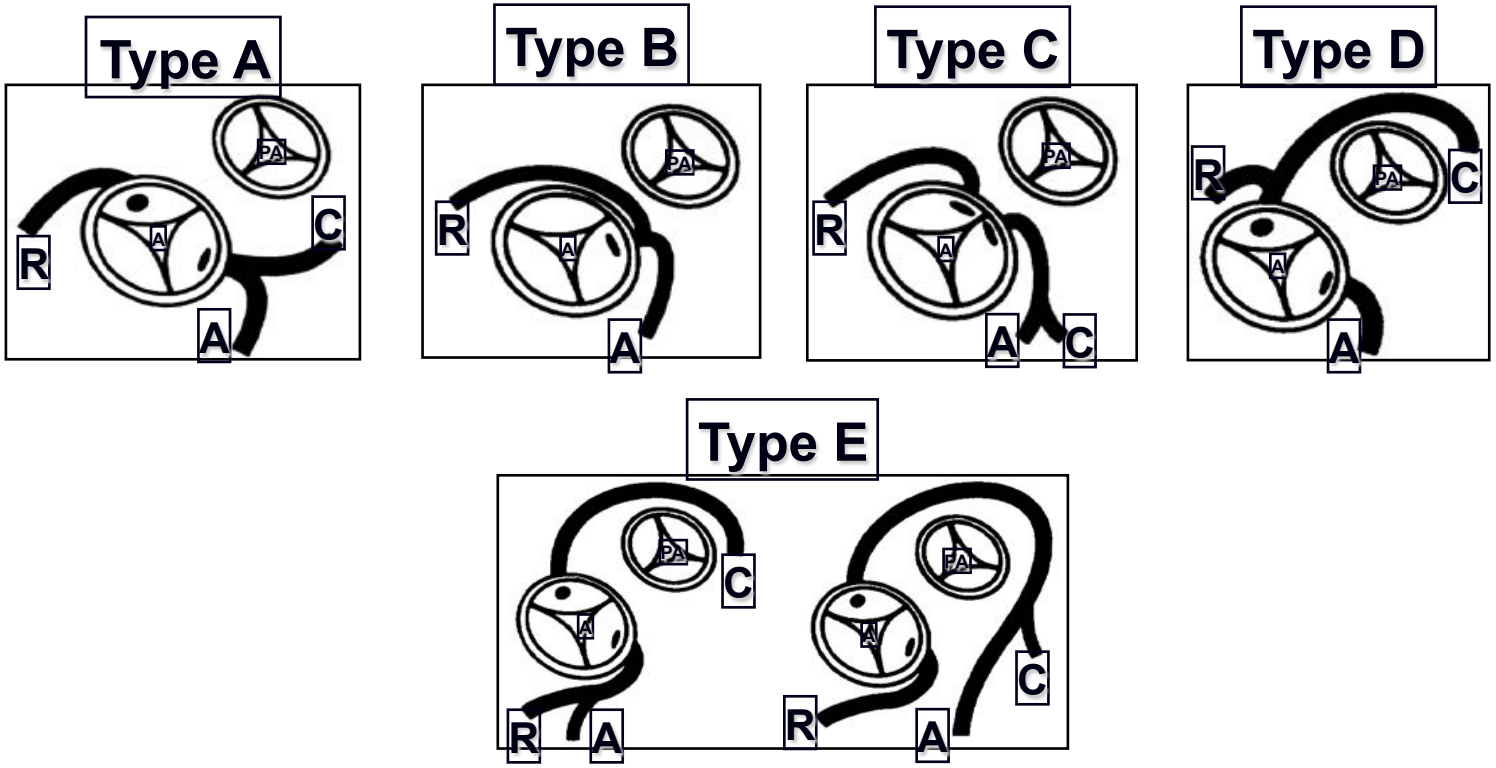
Coronaires Type A: CD

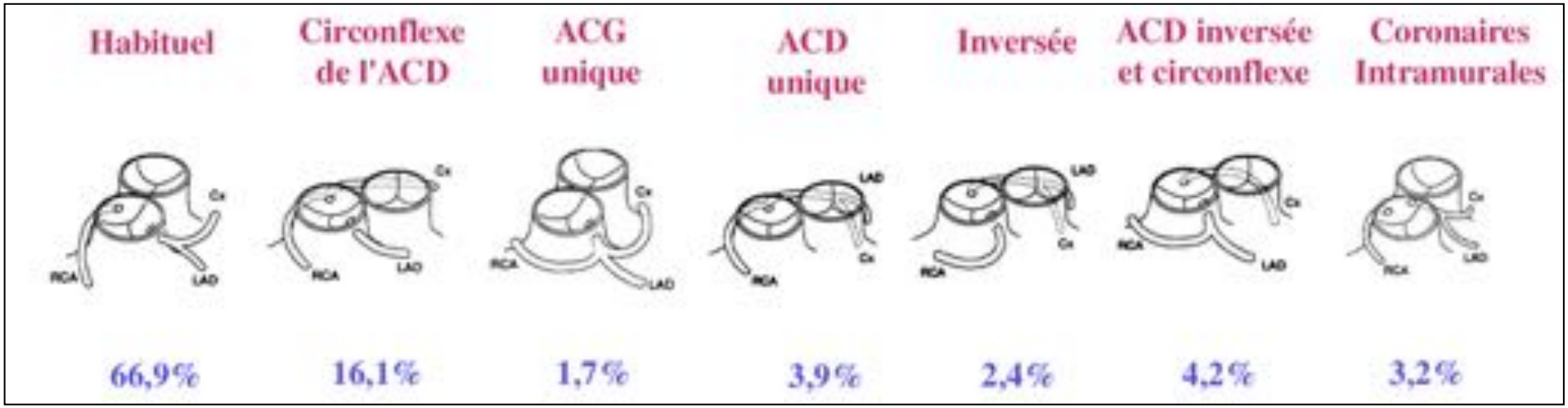


TGV avec CIV d'outlet



Classification de Yacoub





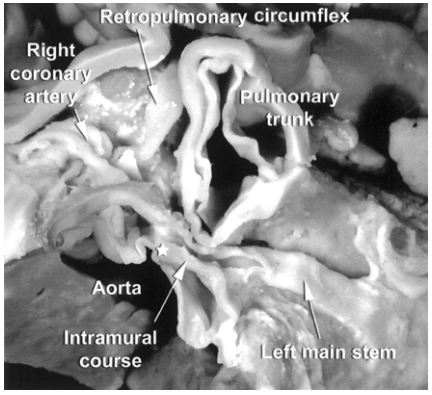
Anatomie coronaire

- « normale » : 60%
- boucle antérieure et/ou postérieure : 35%
- entre gros vaisseaux (intramurale) : 5%

Complications du switch artériel

- **Mortalité**
  - 1 à 2%
  - Liée aux coronaires

- **Morbidité** (importance du suivi)
  - 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



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

Outcomes of the Arterial Switch Operation for Transposition of the Great Arteries: 25 Years of Experience

Tyson A. Fricke, BMedSc, Yves d'Udekem, MD, PhD, Malcolm Richardson, MBBS,

Table 2. Early Mortality

Era	Overall (Deaths/No. Patients)	TGA-IVS	TGA-VSD	TGA-AAO
1983–1989	2/129	1/69	1/49	0/11
	1.6%	1.4%	2.0%	0%
1990–1999	9/266	3/189	3/64	3/13
	3.4%	1.6%	4.7%	23.1%
2000–2009	6/223	3/139	3/70	0/14
	2.6%	2.2%	4.3%	0%
1983–2009	17/618	7/397	7/183	3/38
	2.8%	1.8%	3.8%	7.9%

AAO = aortic arch obstruction; IVS = intact ventricular septum; TGA = transposition of great arteries; VSD = ventricular septal defect.

Mortalité globale du switch artériel: 2,8%

D-TRANSPOSITION OF THE GREAT ARTERIES: Hot Topics in the Current Era of the Arterial Switch Operation

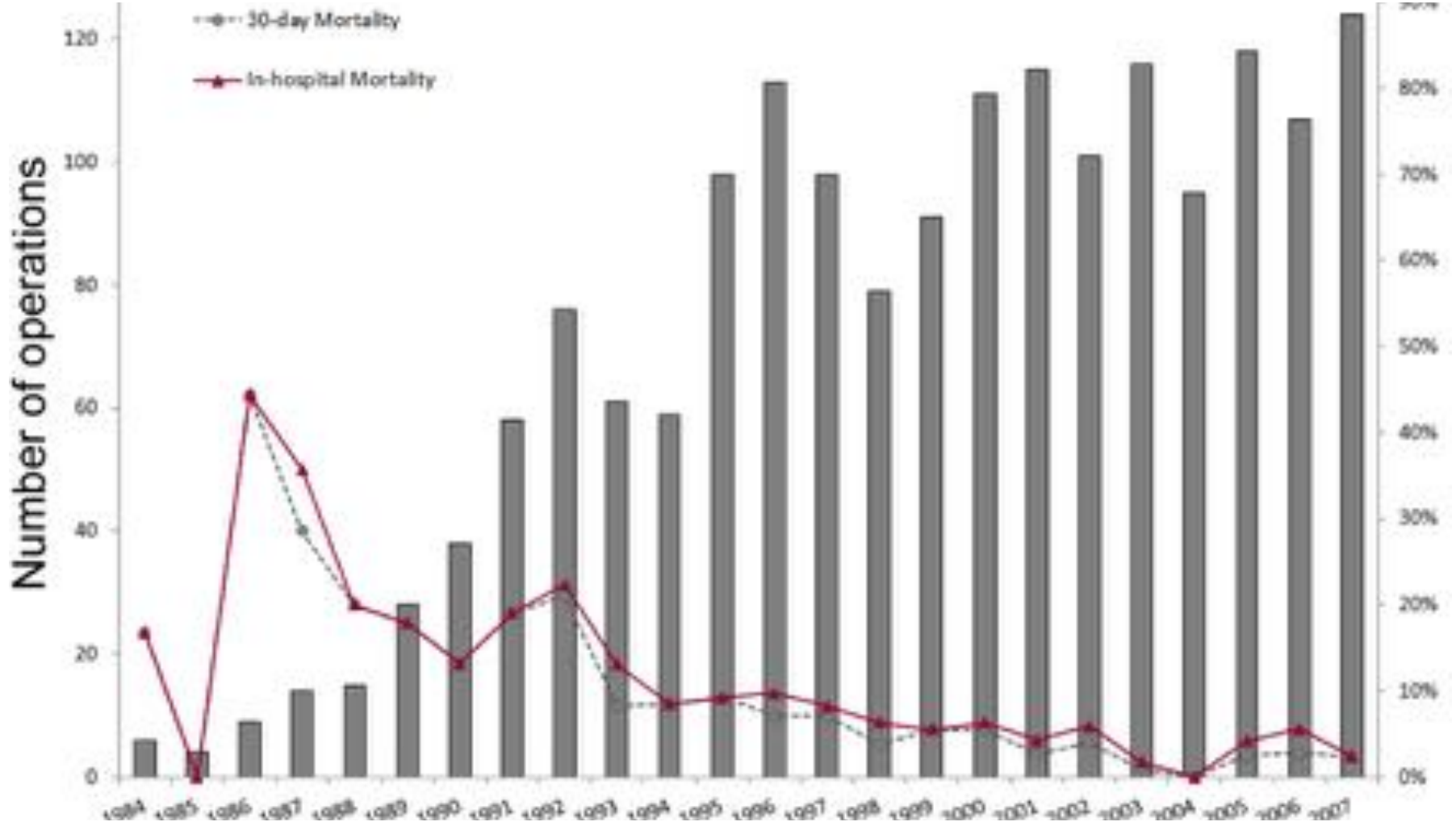
TABLE 2

Outcome and predictors of early mortality of the arterial 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

D-TRANSPOSITION OF THE GREAT ARTERIES: Hot Topics in the Current Era of the Arterial Switch Operation



Données du **Pediatric Cardiac Care Consortium**: 45 centres aux USA; n=1700 patients depuis 1984
2,9% de mortalité dans les derniers 5 ans

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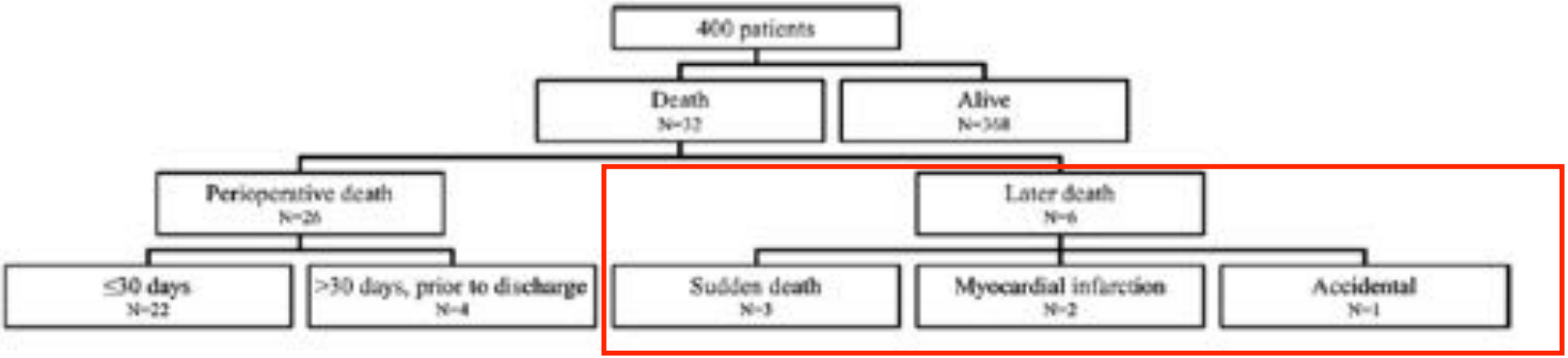
Cardiovascular Outcomes After the Arterial Switch Operation for D-Transposition of the Great Arteries

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

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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)

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Executive function and theory of mind in school-aged children after neonatal corrective cardiac surgery for transposition of the great arteries

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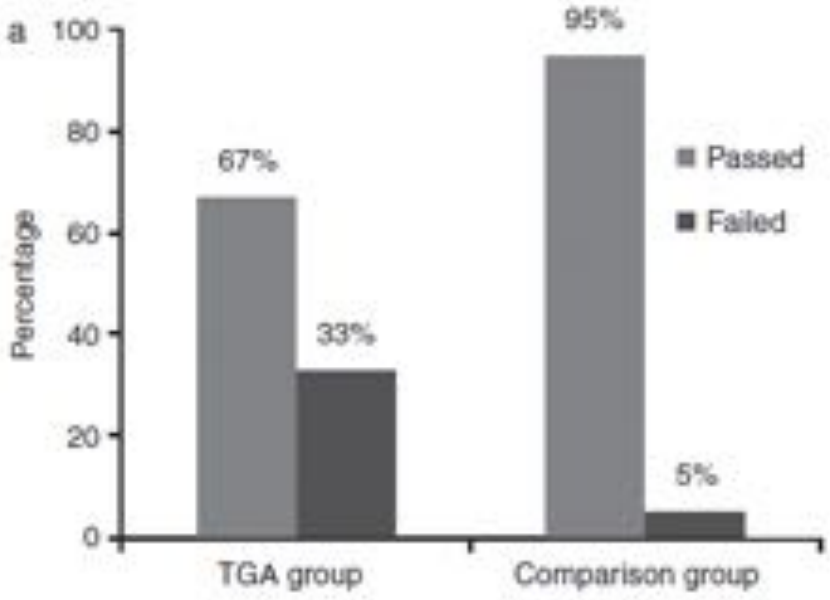

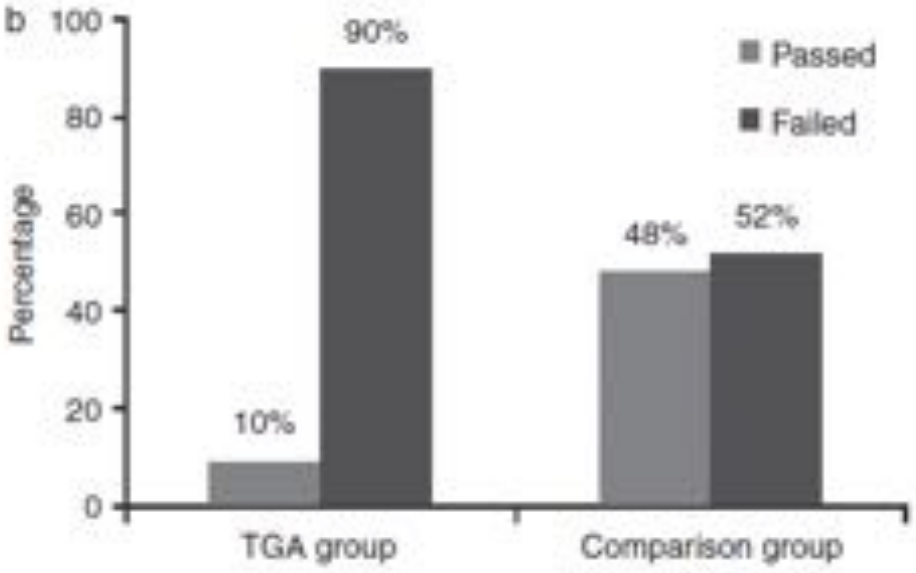

Figure 1: Performance of childen with transposition of the great arteries (TGA) and of comparison individuals on theory of mind tasks. (a) First-order false belief task (level 1). (b) Second-order false belief task (level 2).

n=21 pts vs n=21 ctr
Mean IQ 113 normal