

22 and 23 Novembre 2018

## M3C-Necker

Cardiologie Congénitale et Pédiatrique

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M3C Academy

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# Transposition of the great arteries

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## Polemics with the M3C staff

**Unité médico-chirurgicale de Cardiologie Congénitale et Pédiatrique  
Hôpital Universitaire Necker Enfants malades – APHP, Université Paris Descartes, Sorbonne Paris Cité  
IcarP Cardiology, Institut Hospitalo-Universitaire IMAGINE**

**Centre de Référence Maladies Rares  
Malformations Cardiaques Congénitales Complexes-M3C  
Centre de Référence Maladies Rares  
Maladies Cardiaques Héritaires- CARDIOGEN**





**« I want to terminate pregnancy after my foetus had been diagnosed with a simple TGA. »**



**« I want to terminate pregnancy after my foetus had been diagnosed with a TGA :**

**with VSD and pulmonary stenosis**

**with VSD and coarctation of the aorta**

**with intramural type C left coronary artery »**



**« I do not want my patient to be delivered in the reference center after I had diagnosed a foetal TGA with VSD in a pregnant woman referred to me. »**



**« I perform Rashkind procedure in every patient with TGA because I am convinced that it is necessary and also because I need to train myself. »**

# Droit au Rashkind partout et pour tous ?

- assurer une protection efficace de ces savoirs
- permettre aux communautés locales de tirer profit de l'exploitation de leurs savoirs traditionnels
- de ne pas restreindre la propension de ces savoirs à contribuer au progrès scientifique
- de veiller au partage des connaissances qui en peuvent en résulter
- des savoirs dynamiques et à des solutions pratiques en rapport avec l'environnement social contemporain de la collectivité
- des valeurs culturelles détenu(e)s collectivement [et leur] maîtrise n'est pas nécessairement aux mains d'individus qui se servent de savoirs fragmentés, mais aux mains de la communauté ou de la collectivité toute entière

**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 <sup>1</sup>**

**Balloon atrial septotomy **performed out-of-hours produced higher complication rates** as opposed to balloon atrial septotomy 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 <sup>3</sup>**

**Rashkind procedure was not associated with increased risk of necrotising enterocolitis, but was associated with nearly twice the risk of clinically recognized stroke (1% versus 0%,  $p = 0.046$ ) <sup>2</sup>**

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

**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
<b>Balloon septostomy</b>	<b>5</b>

**TABLE 3. Advanced Training—Recommended Minimum Case Numbers**


Total cardiac catheterizations	200
Interventional procedures	100
Type of intervention	
<b>Balloon septostomy</b>	<b>5</b>
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


## **Savoir faire et compétences techniques: référentiel métier de Cardiologue Congénitaliste et Pédiatrique français**

### *Cathétérisme cardiaque interventionnel*

50 (assistant ou en première ligne), avec **maitrise de l'atrioseptotomie de Rashkind** et de l'entraînement électrosystolique transitoire.

## Justification *a posteriori* du Transfert *in utero*

  
Transfert indiscutable  
n=899 (71,5%)

  
Transfert à justifier  
A posteriori  
N=359 (28,5%)

Motif du transfert	Total	Total %
Nécessité de Rashkind	344	27,3
Cardiopathie évolutive	272	21,6
DDP	107	8,5
DDS	93	7,4
Nécessité d'intervention	83	6,6
DDP possible	156	12,4
Complément diagnostique	94	7,5
Tolérance néonatale	51	4,1
DDS possible	35	2,8
Pas d'intérêt cardiaque	23	1,8
Total	1258	100,0

# 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

# Droit au Rashkind partout et pour tous

Very rare patients

< 1/an per pediatric cardiologist

Potentially the more severe patients

Delayed diagnosis, poor hemodynamic conditions, restrictive FO

Non trained physicians

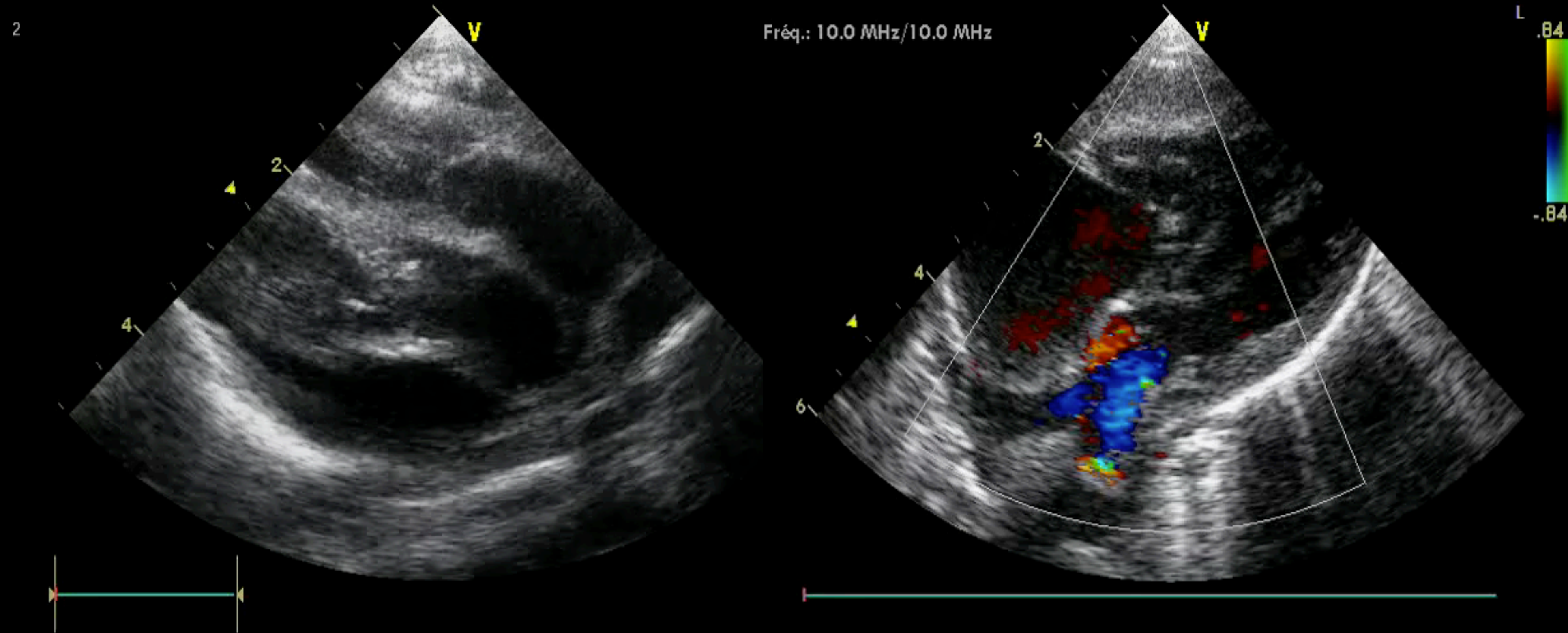
Non prepared cath lab

No back-up

Need for subsequent transport

# Cas clinique

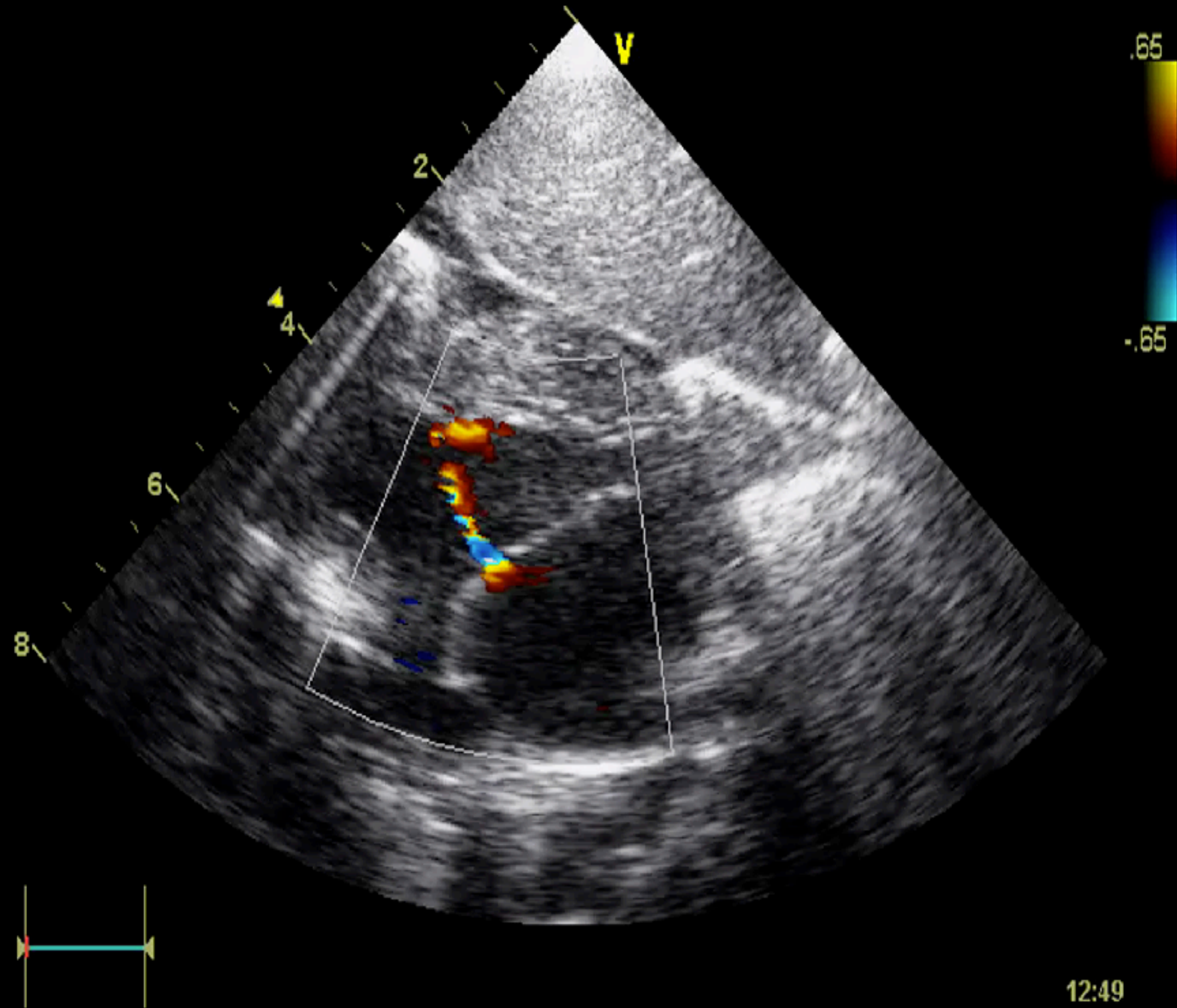
Vous accueillez un nouveau-né pour cyanose réfractaire.





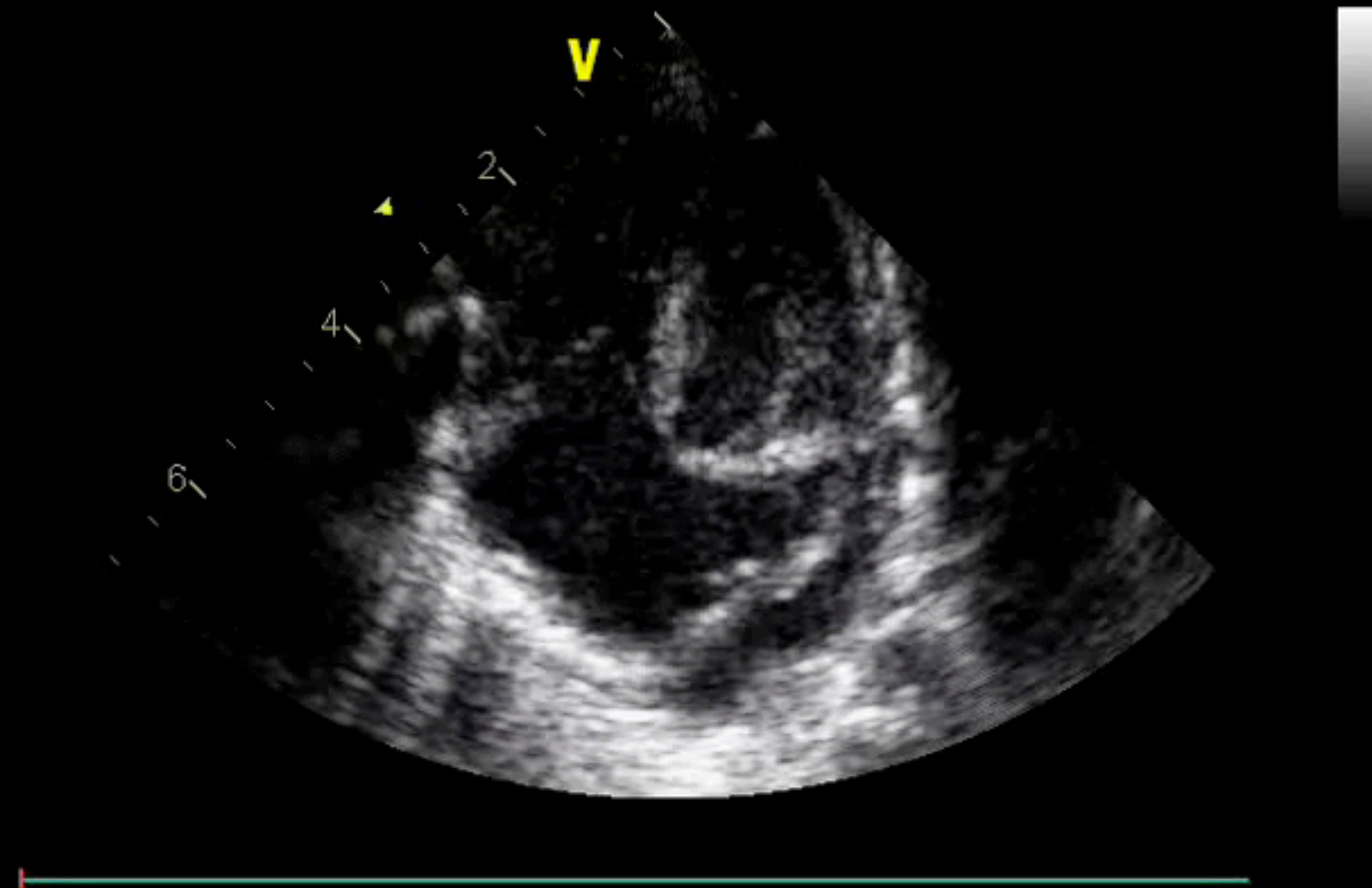
# Cas clinique

Il se dégrade rapidement avec une saturation à 60%, un teint gris et des signes d'acidose clinique.

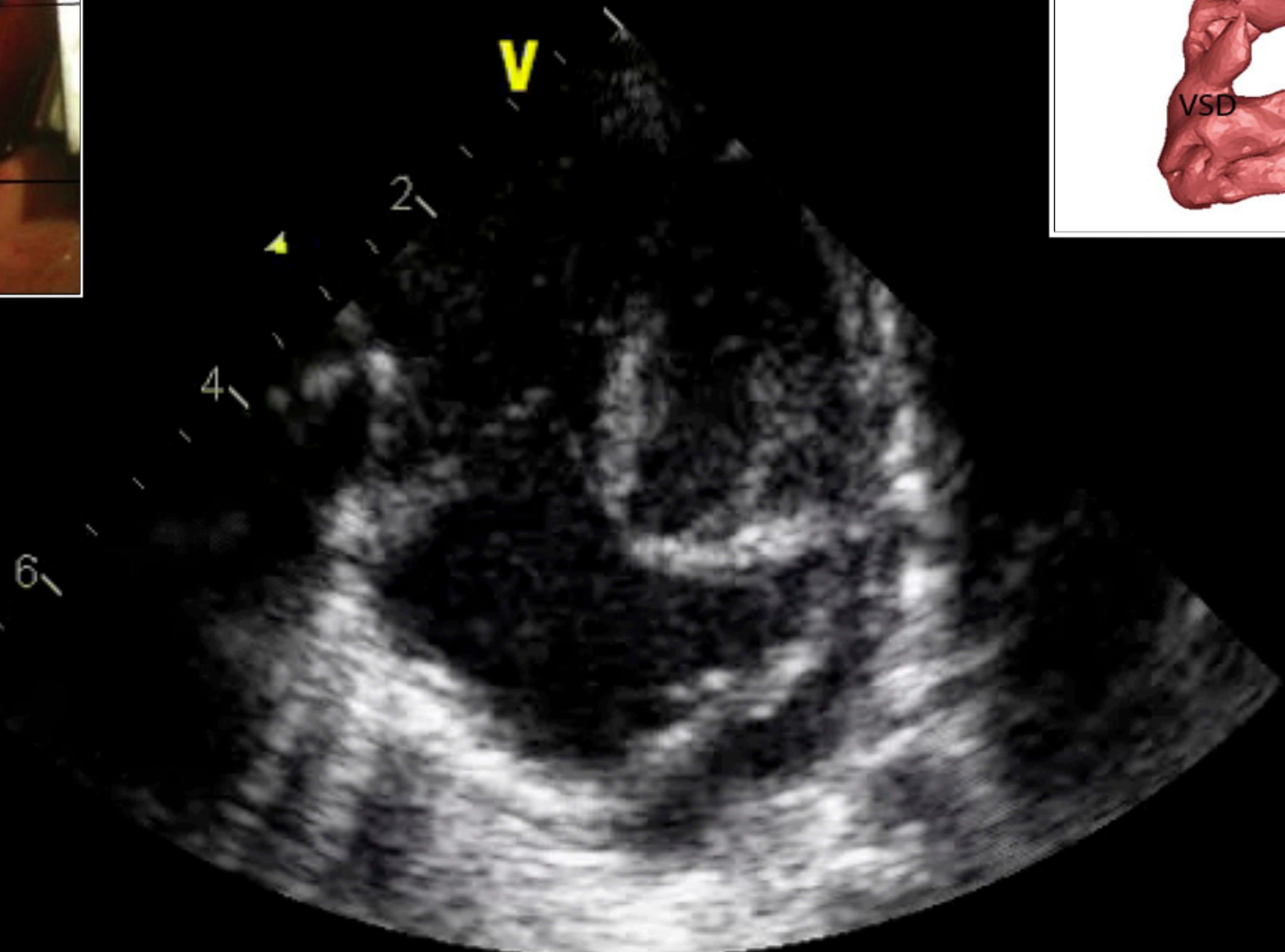
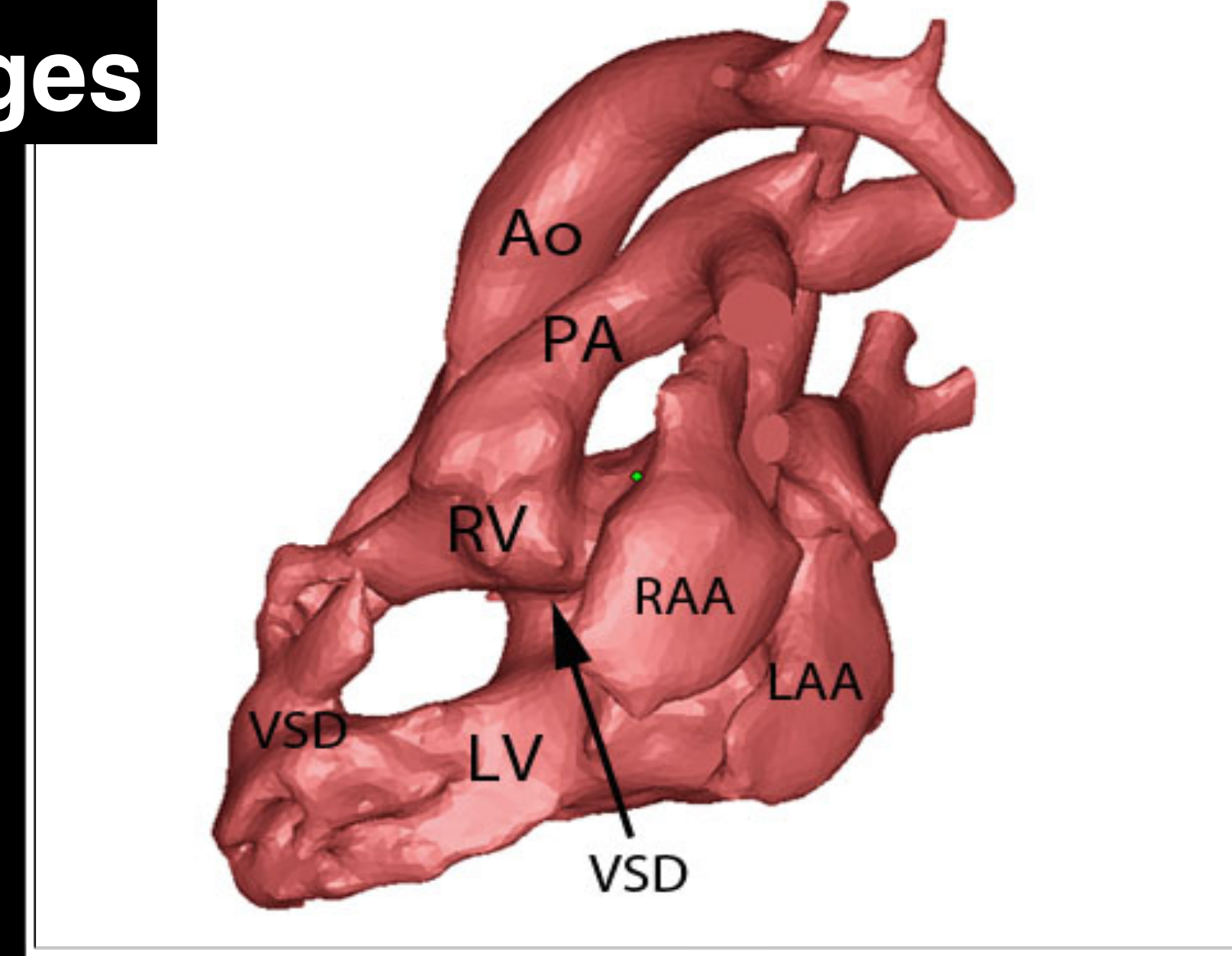
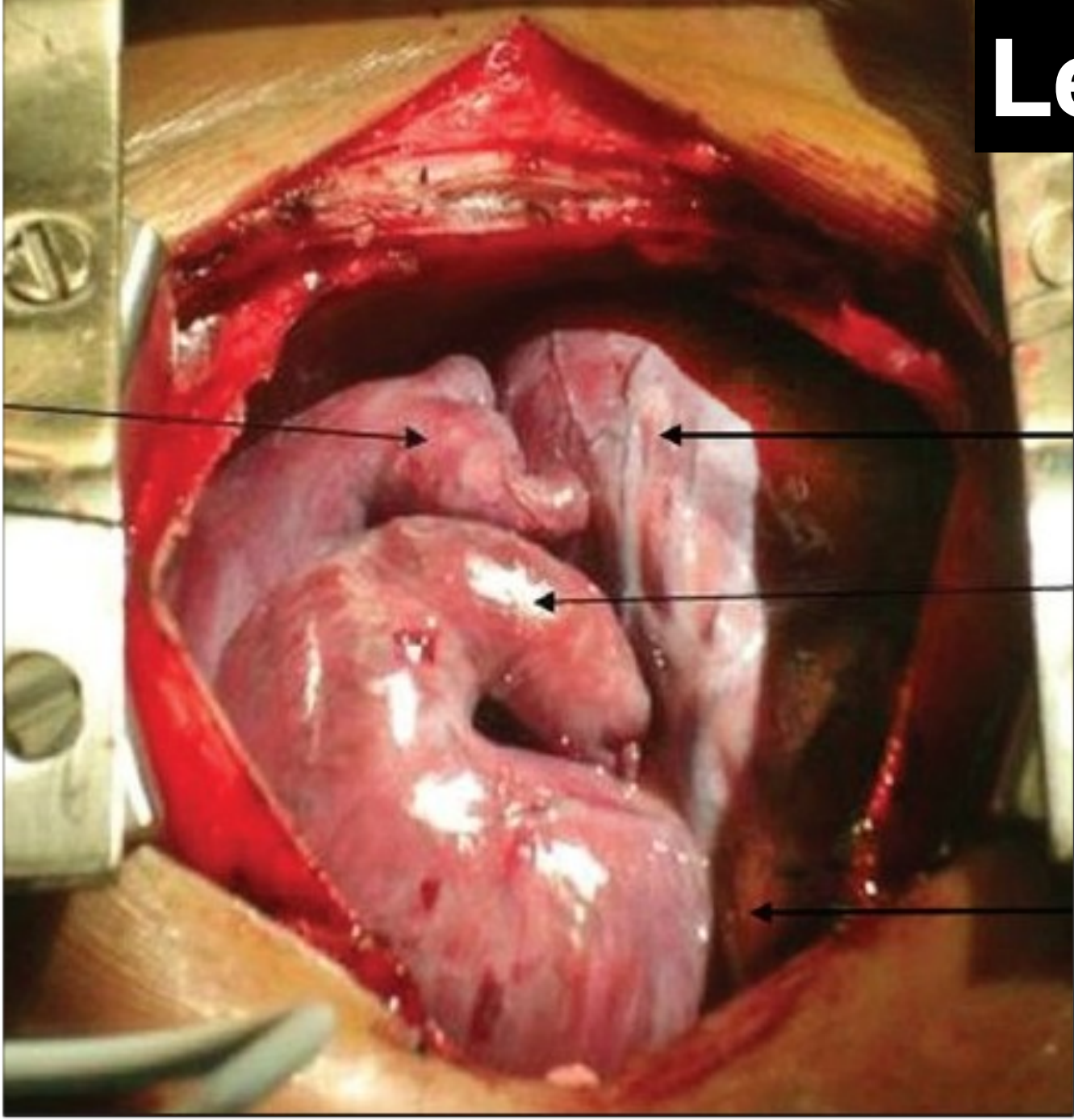


# Cas clinique

Au cours de la procédure, vous êtes troublés par l'anatomie des oreillettes. Quelle est votre interprétation ? Quel est le risque de cette anomalie ?



# Left juxtaposition of atrial appendages





**« In my institution there is no death after ASO and we have never seen any coronary artery anomaly, therefore we never screen patients for this type of problem. »**

## **The statistical issue in pediatric cardiology**

5% coronary obstructions

**80 ASO per year : 4 patients per year**

10 ASO per year : 1 patient every 2 years

60% asymptomatic : 1 patient every 4 years in the absence of screening

4 pediatric cardiologists : 1 patient every 16 years in the absence of screening



**L'expérience n'éclaire que la paume de celui qui en témoigne**  
***Confucius***

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>	Ref <sup>c</sup>
Routine annual follow-up visits should be considered and should include history taking, 12-lead ECG and echocardiograms	IIa	C	
CMR should be integrated in the routine evaluation of all postoperative patients with TGA with an individually tailored frequency	I	B	295
Routine 24-h Holter follow-up visits should be considered-annually for patients <5 years old and every second year for patients >5 years old	IIa	C	
Routine exercise testing at least every 3 years should be considered for patients >8 years old	IIa	C	
Routine invasive electrophysiology assessments are not indicated, because inducible ventricular tachycardia is not a predictor of future events in ASO patients	III	C	

ASO: arterial switch operation; CMR: cardiovascular magnetic resonance; ECG: electrocardiogram; TGA: transposition of the great arteries.

<sup>a</sup>Class of recommendation.

<sup>b</sup>Level of evidence.

<sup>c</sup>References.

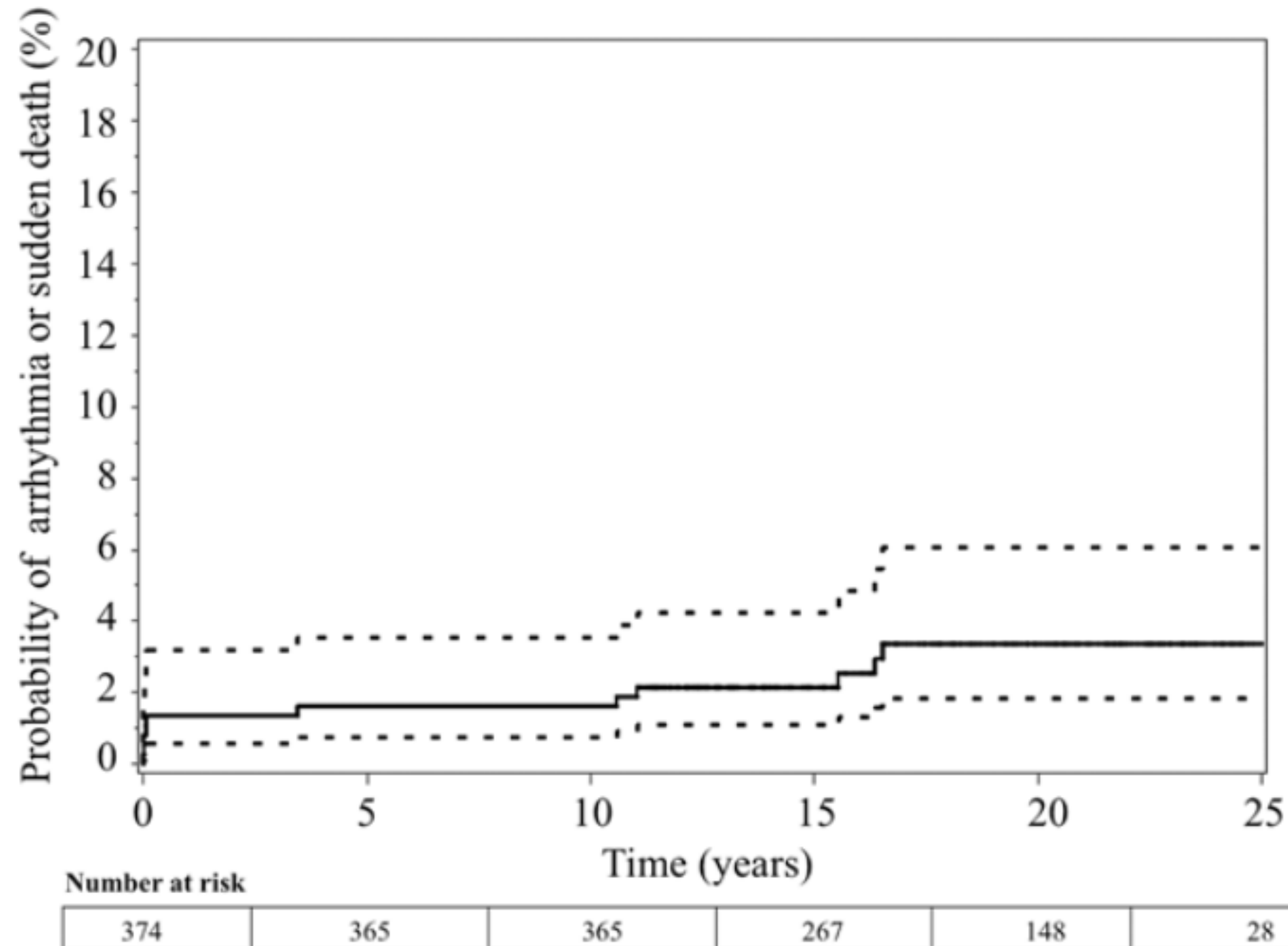
Recommendations for cardiology follow-up and arrhythmias



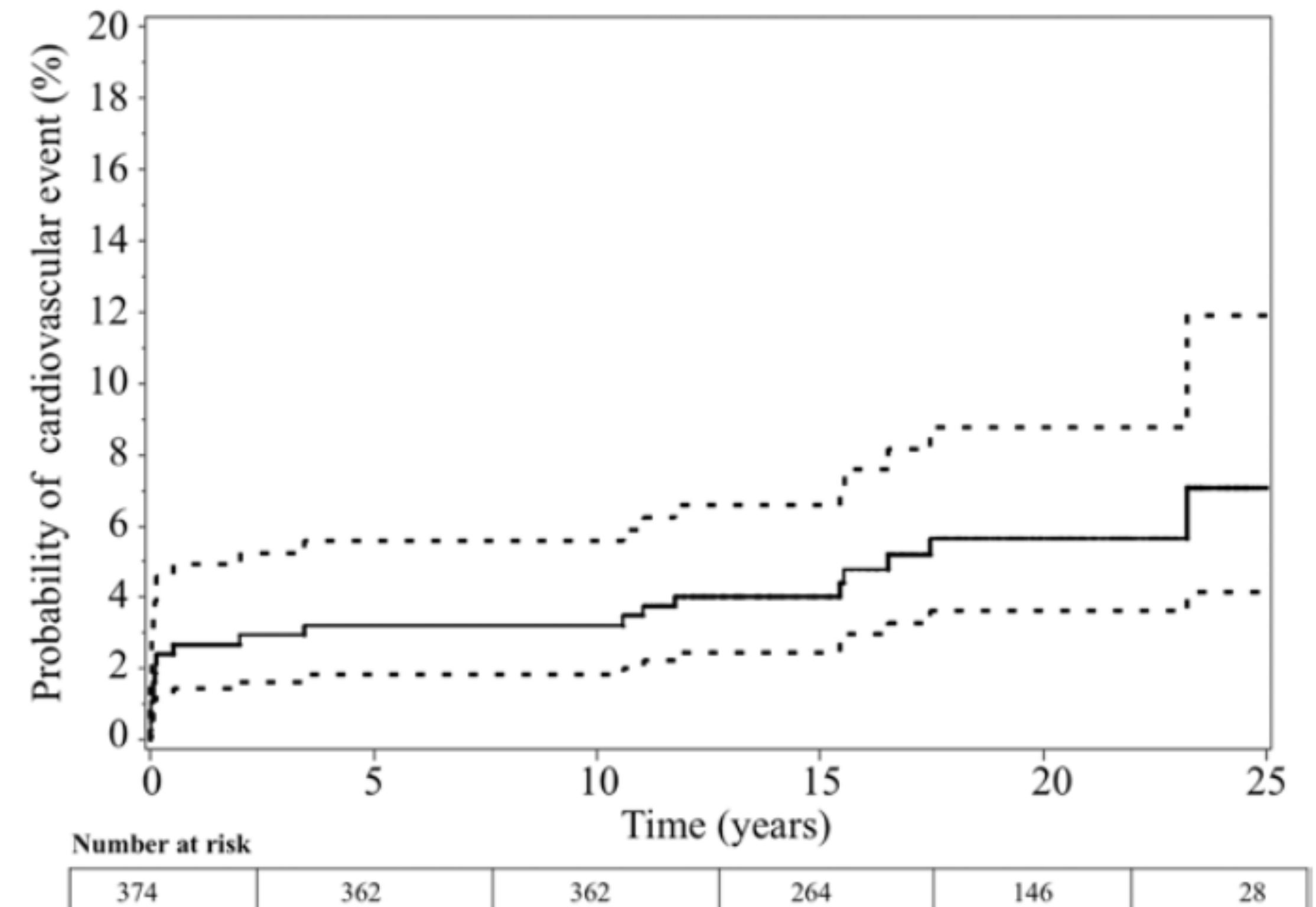
**« The risk of atherosclerotic lesions after the ASO is certainly low as myocardial infarction has not been described in adult patients. I think that it is useless to screen adult patients for myocardial ischemia after ASO for TGA. »**



# Cardiovascular events in the long term



Cumulative probability of arrhythmia or sudden death



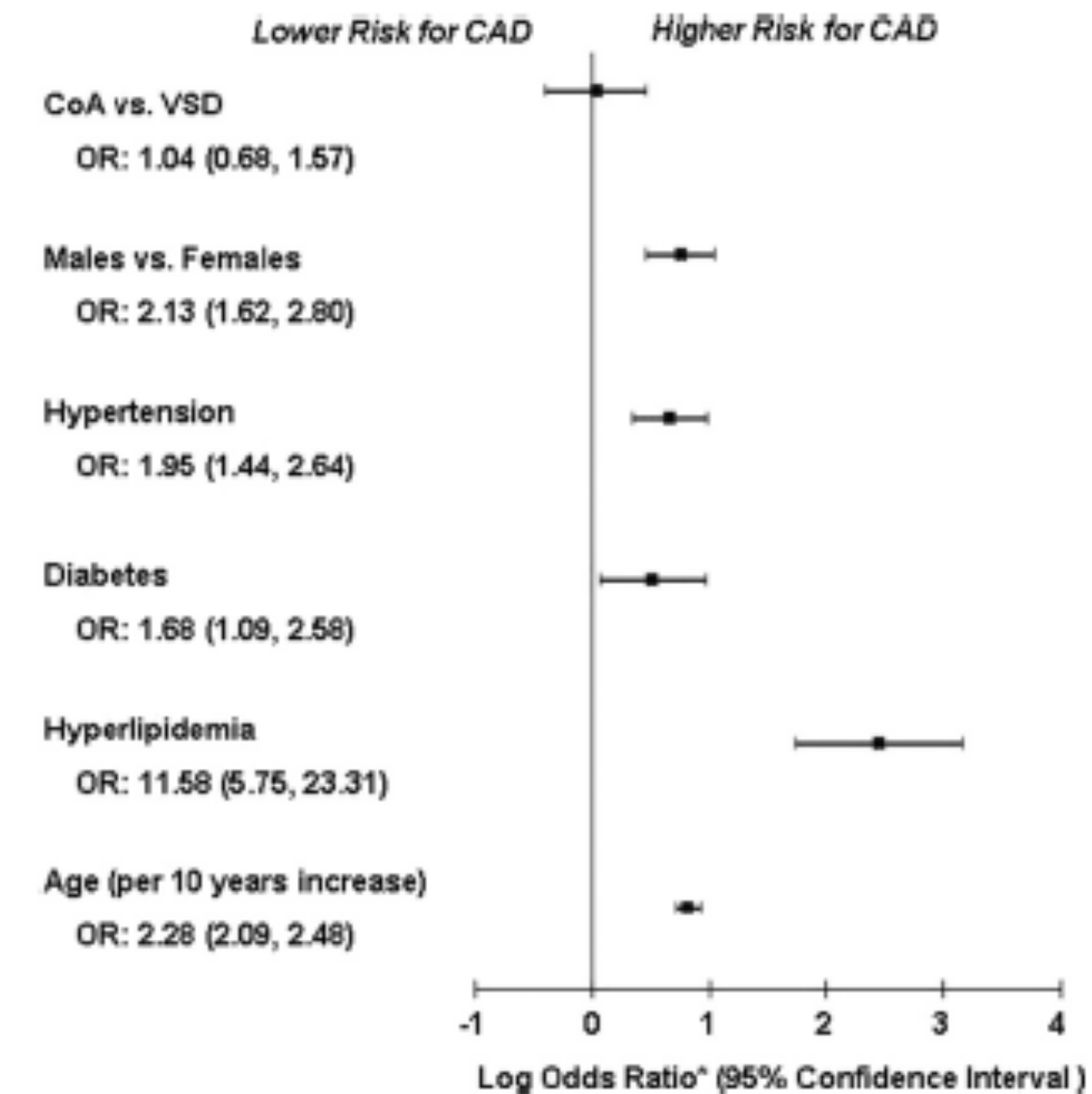
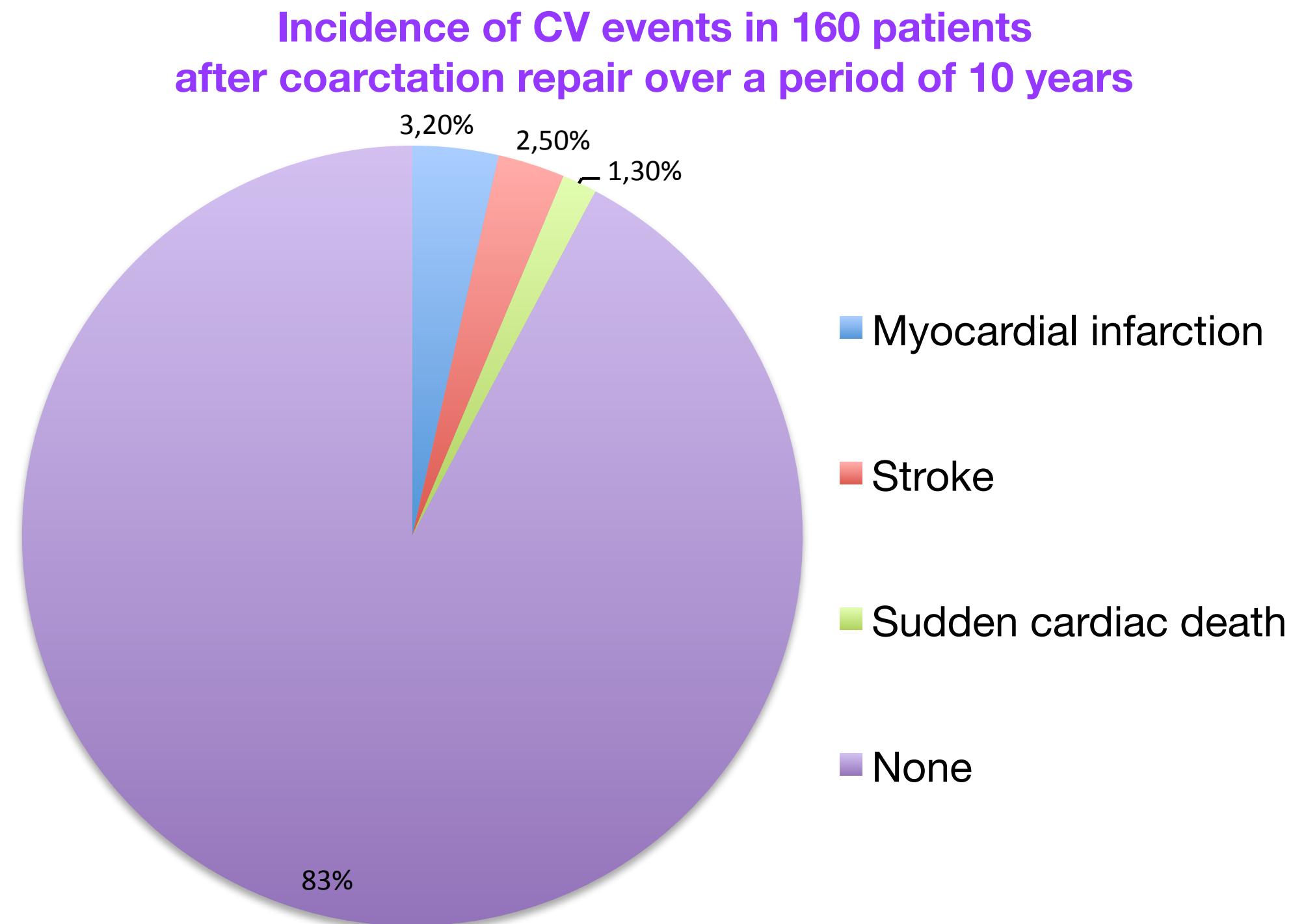
Cumulative probability of the combined cardiovascular outcome

Residua, sequelae, and complications are listed below  
ESC Guidelines 2010

- Arterial hypertension *at rest or during exercise* is common, even after successful treatment, and it is an important risk factor for premature CAD, ventricular dysfunction, and rupture of aortic or cerebral aneurysms. *The geometry of the arch may play a role in the development of hypertension. The significance of isolated, exercise-induced hypertension* is a matter of debate.
- Recurring or residual CoA may induce or aggravate systemic arterial hypertension and its consequences
- Aneurysms of the ascending aorta or at the intervention site present a risk of rupture and death. Patch repair are at particular risk of repair site aneurysms and should be imaged on a regular basis.
- **Attention is required for BAV, mitral valve disease, premature CAD, and berry aneurysm of the circle of Willis.**

# Risk of premature CAD after coarctation repair

CoA did not independently predict for the development of CAD (OR, 1.04; 95% CI, 0.68 –1.57) or premature CAD (OR for CoA versus ventricular septal defect, 1.44; 95% CI, 0.79 –2.64) after adjustment for other cardiovascular risk factors.



**Figure 2.** Multivariable analysis of the effect of aortic coarctation (CoA) vs ventricular septal defect (VSD) on coronary artery disease (CAD; nested case-control sample). OR indicates odds ratio.

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>	Ref <sup>c</sup>
Diagnostic suggestions			
Selective coronary angiography or multislice CT angiography after an ASO, possibly complemented by myocardial perfusion imaging using MRI or thallium-201 scintigraphy:			
• Is indicated in the presence of electrocardiographic signs, echocardiographic signs, or both, that are suggestive of myocardial ischaemia at any time after the operation;	I	C	293, 389
• Should be considered in the presence of unusual coronary patterns (single orifice, coronary arteries coursing between the great arteries) or intraoperative difficulties in coronary transfer, usually during the first postoperative year.	IIa	C	293, 389

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>	Ref <sup>c</sup>
Indications for late reoperation			
Reoperation is indicated in the event of late coronary insufficiency demonstrated with myocardial imaging	I	C	390, 399, 400
Revascularization may be considered in the absence of evident myocardial ischaemia but in the presence of demonstrated coronary obstruction	IIb	C	293
Suggested treatment			
Coronary (ostial) patch angioplasty is indicated for proximal discrete obstruction	I	C	293, 321, 383, 389
Internal mammary artery grafting should be considered for more distal lesions, long and complete occlusions of the main stem or residual obstruction after primary surgical arterioplasty	IIa	C	173, 293, 354, 389, 401
Coronary (ostial) patch angioplasty and concomitant internal mammary artery grafting are not indicated	III	C	173, 354
Percutaneous transluminal coronary angioplasty, with or without coronary stent implantation, may be considered, preferably after failing primary surgical arterioplasty	IIb	C	382, 397, 398

ASO: arterial switch operation; CT: computed tomography; MRI: magnetic resonance imaging.

<sup>a</sup>Class of recommendation.

<sup>b</sup>Level of evidence.

<sup>c</sup>References.

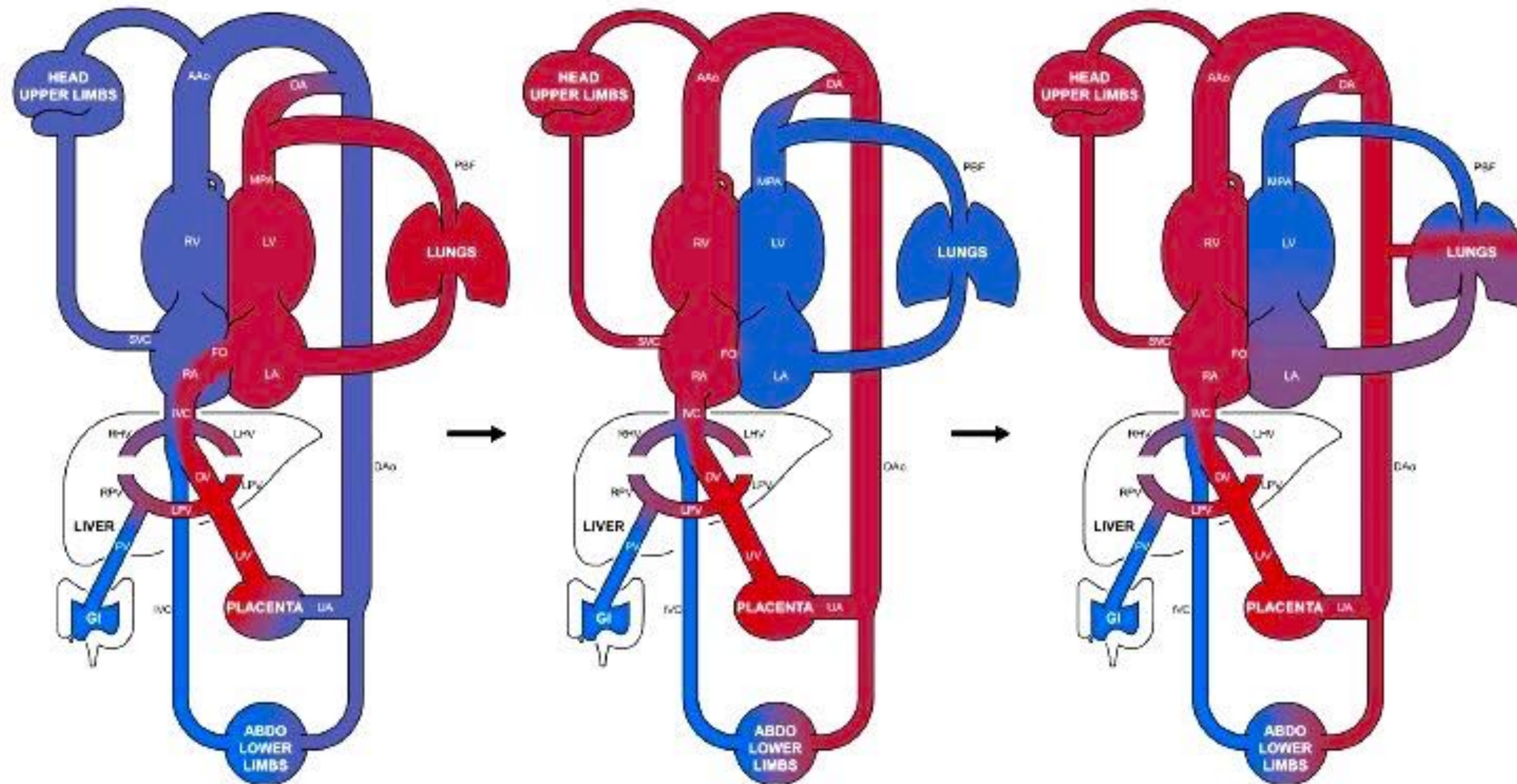
Recommendations for reoperations for residual or recurrent coronary lesions



**« The only patient of mine who died after ASO developed pulmonary hypertension after two years of age and subsequently died of right heart failure. Probably coincidental association. »**

*Mortality is not an end-point*

# Fetal interventions in fetus with TGA

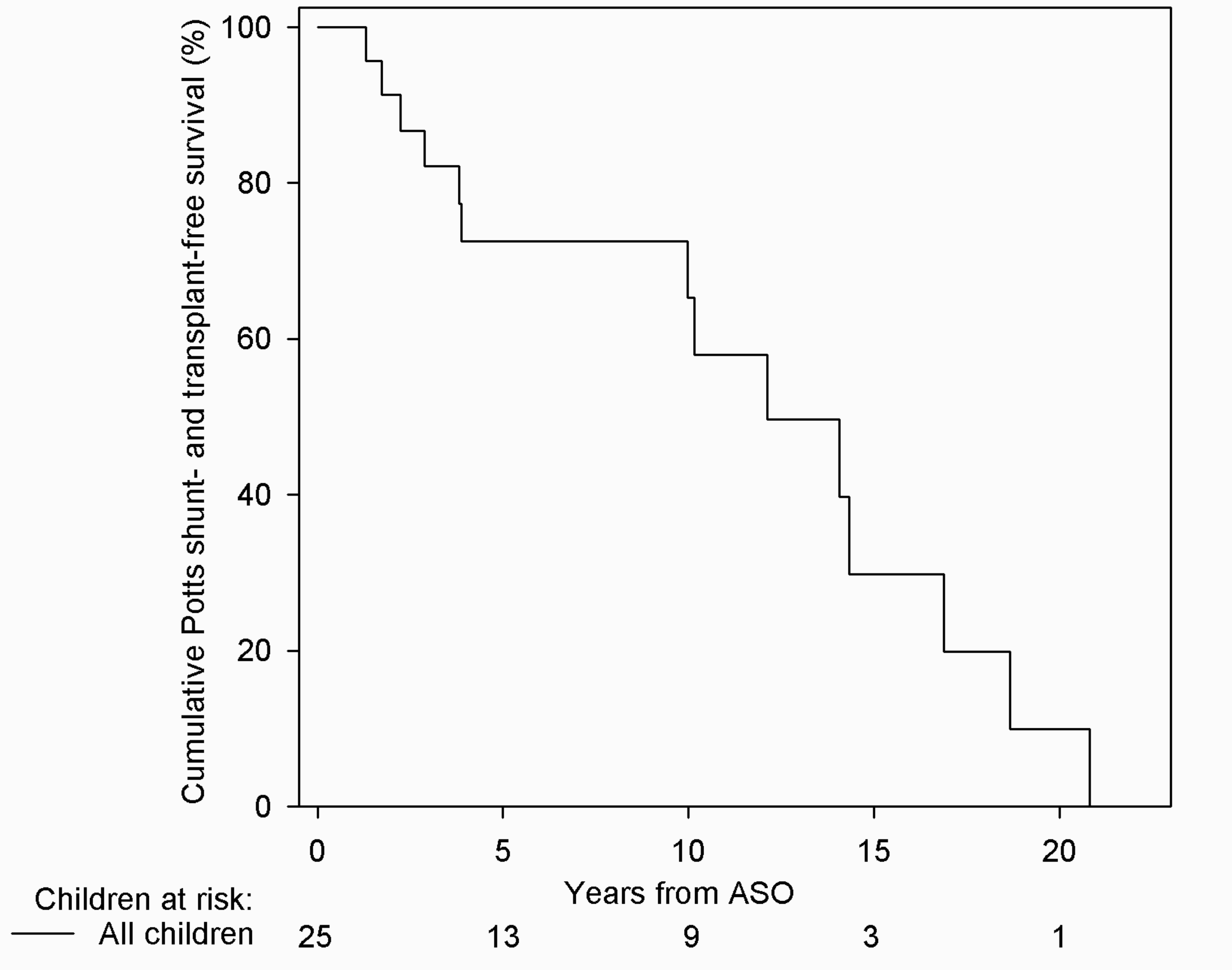


- Initial increase in PBF due to vasodilatation with increased oxygen
- Increased pulmonary venous return
- Reduced size of the FO
- Ductal constriction due to oxygen
- Isolation of Pulmonary circulation
- Increased PVR
- Development of aorta-pulmonary collaterals

# PAH characteristics after ASO for TGA

	Value
Age first PAH detection (months)	4 (1, 25)
PAH detection within one year after ASO	16 (64)
Age first detection (months)	2 (1, 4)
PAH detection more than one year after	9 (36)
Age first detection (months)	40 (25, 93)
Age first RHC (months)	10 (2, 30)
mPAP (mmHg)	48 (37, 55)
mSAP (mmHg)	62 (51, 65)
mPAP/mSAP	0.82 (0.71, 1.07)
mPCWP (mmHg)	10 (8, 12)
PVRi (WU.m <sup>2</sup> )	11.5 (8.7, 13.0)
PAH therapy at endpoint	
CCB monotherapy	1 (4)
PAH-targeted mono therapy	6 (24)
PAH-targeted dual therapy	8 (32)
PAH-targeted triple therapy	10 (40)

# Survival in PAH after ASO for TGA

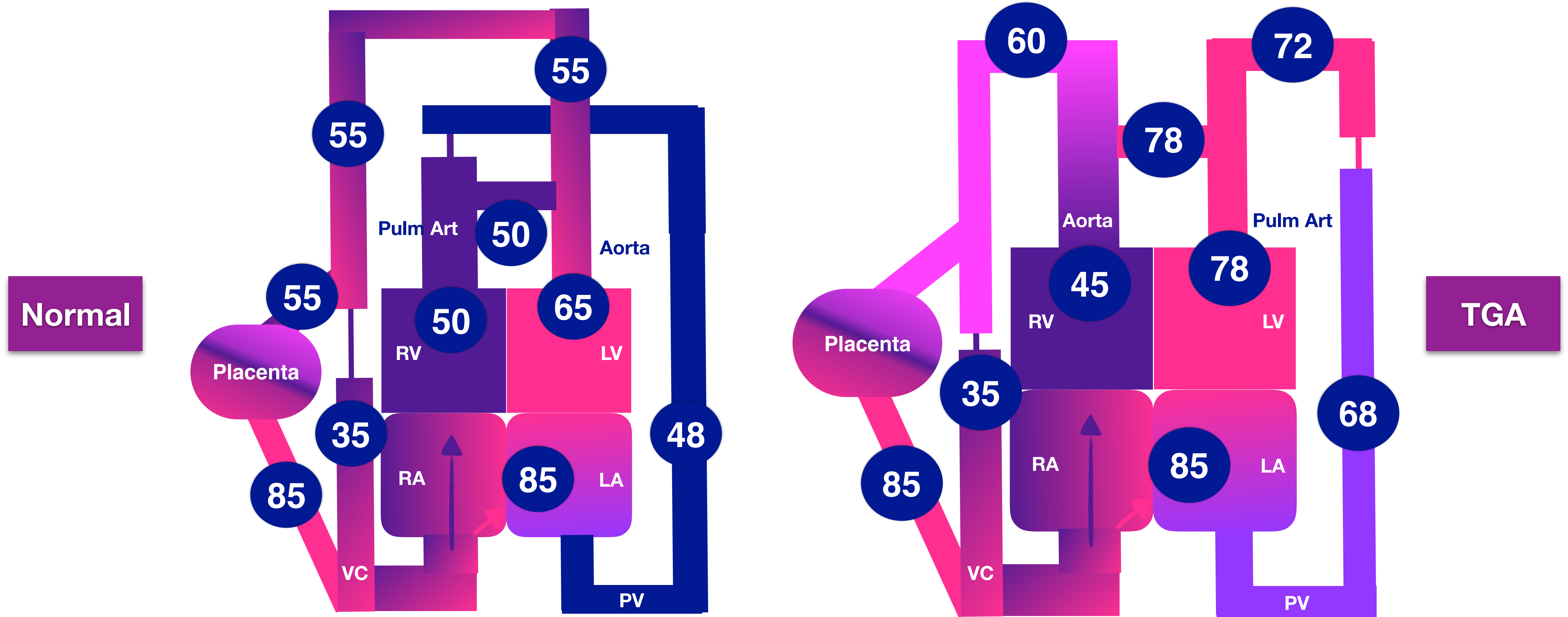




**« I never talk of neuro-developmental outcomes to parents of a foetus or a child with TGA because they may decide to go for termination or be afraid of mental retardation. My role of physician is also to protect them. »**



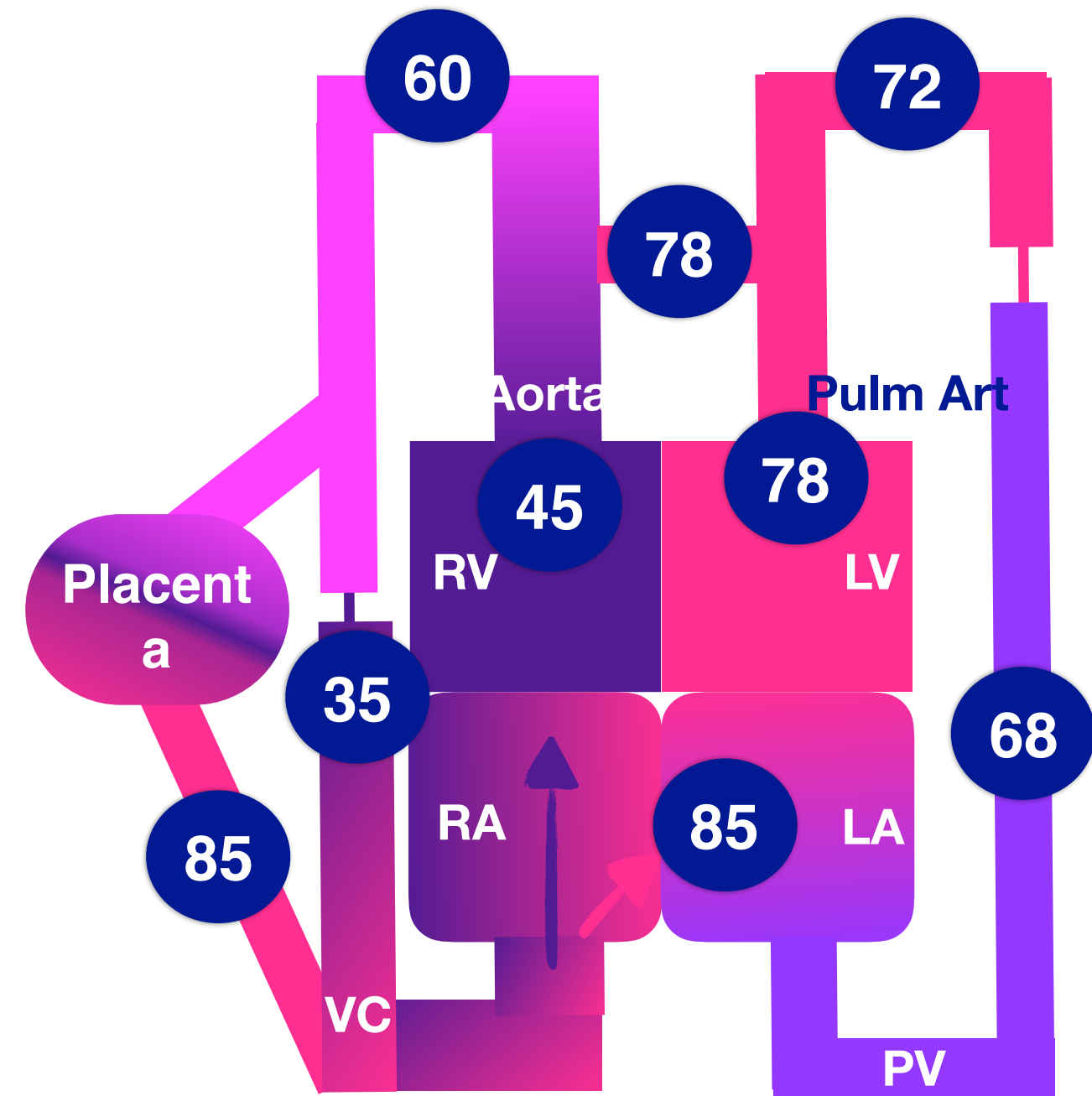
# Fetal circulation and brain perfusion in TGA



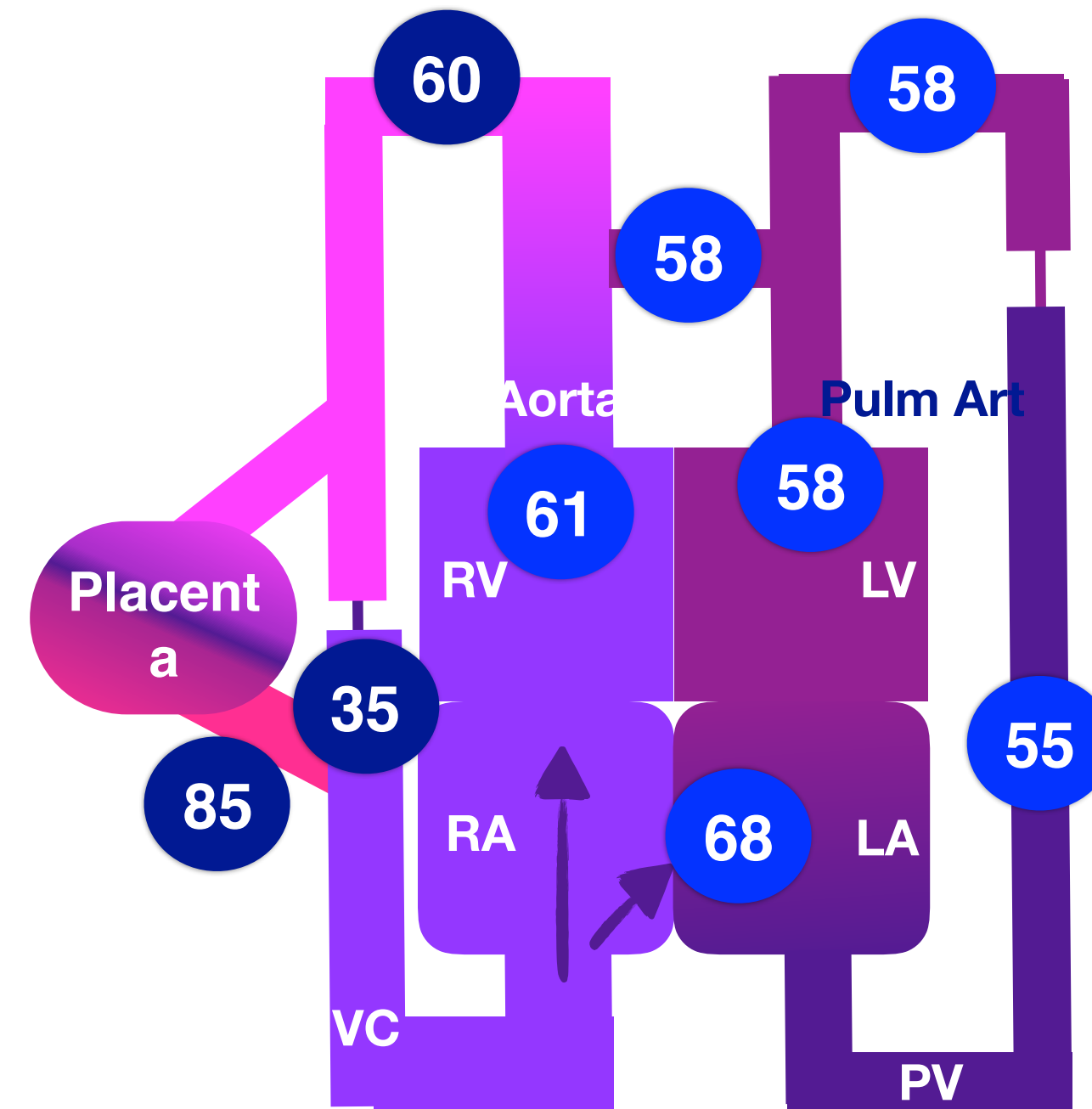
Rudolph A. Ped Res 2007;61:375-80  
 Prsa M et al. Circ Cardiovas Imaging 2014;7:663-70

*Mortality is not an end-point*

# Fetal interventions in fetus with TGA



**TGA fetus**



**Closure of the ductus venosus**