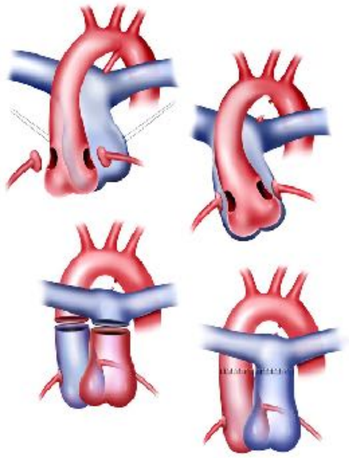
The problem of postoperative evaluation of coronary arteries anatomy after ASO for TGA



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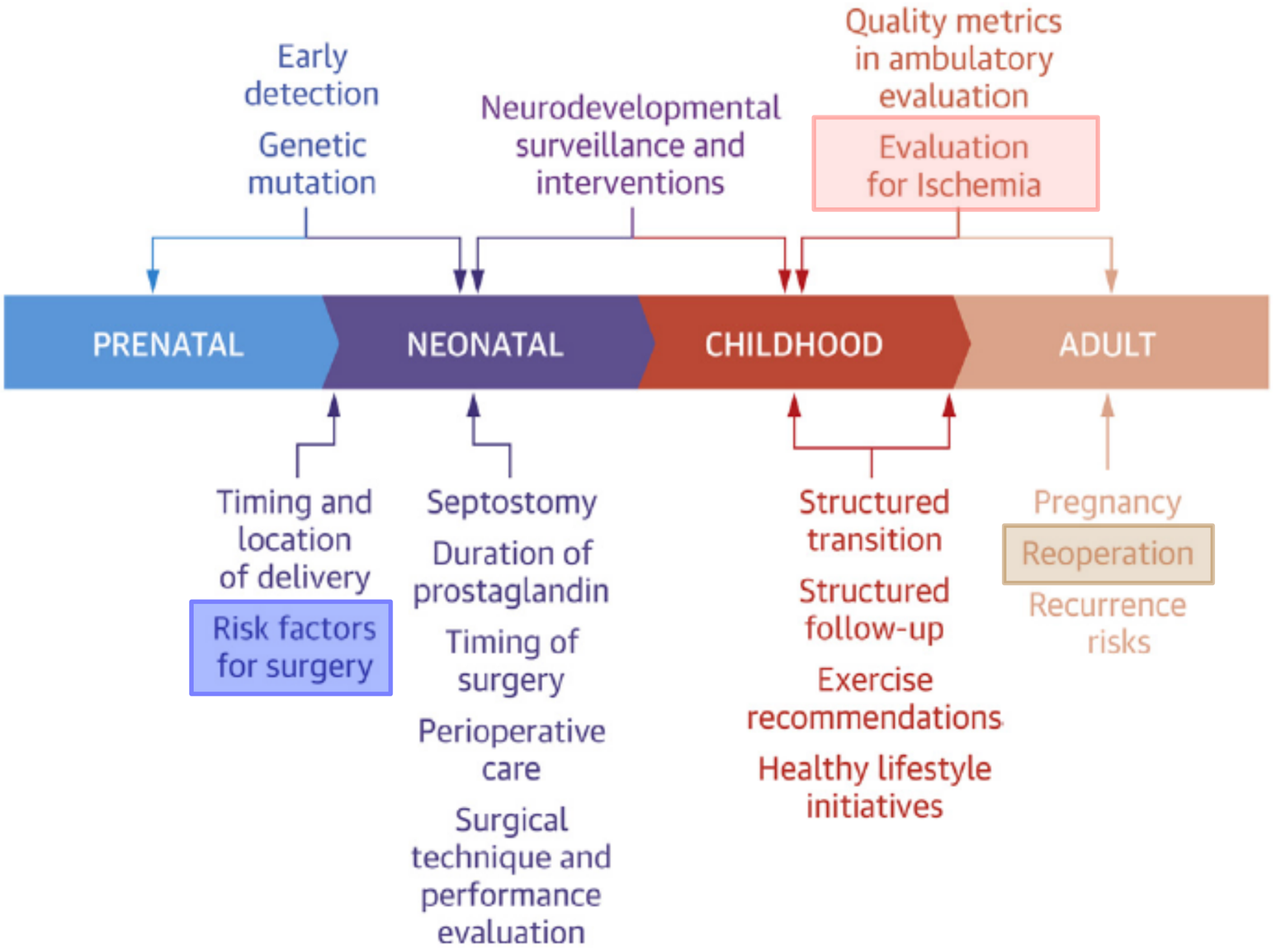
Centre de Référence Maladies Rares
Malformations Cardiaques Congénitales Complexes-M3C

Centre de Référence Maladies Rares
Maladies Cardiaques Héréditaires- CARDIOGEN

Hot topics in diagnosis, management and follow-up of patients with TGA



Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511

Life-long management of the ASO population poses several challenges

1) absence of current consensus regarding the appropriate interval and modality for surveillance imaging;

2) there is no defined management strategy when subclinical anatomic or physiologic abnormalities are identified;

3) symptoms attributable to potential complications, especially coronary obstruction, are rare and therefore practitioners must be vigilant for classic and atypical presentations;

4) the effects of acquired coronary artery disease superimposed on manipulated coronary arteries remain unknown.

Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511

Life-long management of the ASO population poses several challenges

The coronary arteries after transfer

1) symptoms attributable to potential complications, especially coronary obstruction, are rare and therefore practitioners must be vigilant for classic and atypical presentations;

What is the incidence of ischemic events after the ASO for TGA ?

When do they occur ? Do we know risk factors ?

2) absence of current consensus regarding the appropriate interval and modality for surveillance imaging;

Is systematic screening for coronary artery obstruction useful after the ASO for TGA ?

When ? How to screen ?

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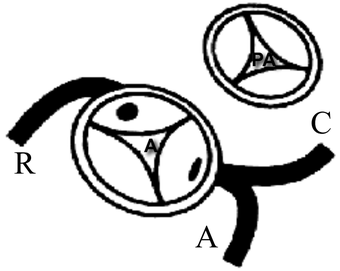
Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511

Big variations in coronary anatomy (origin, loops, epicardial, intramural)

Type A



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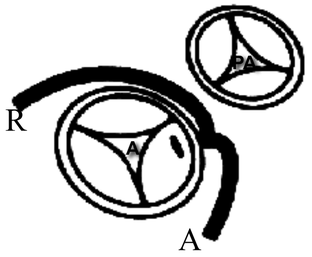

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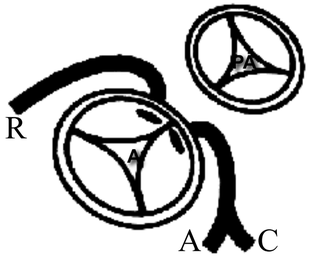
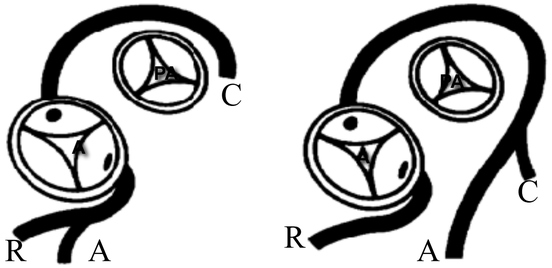
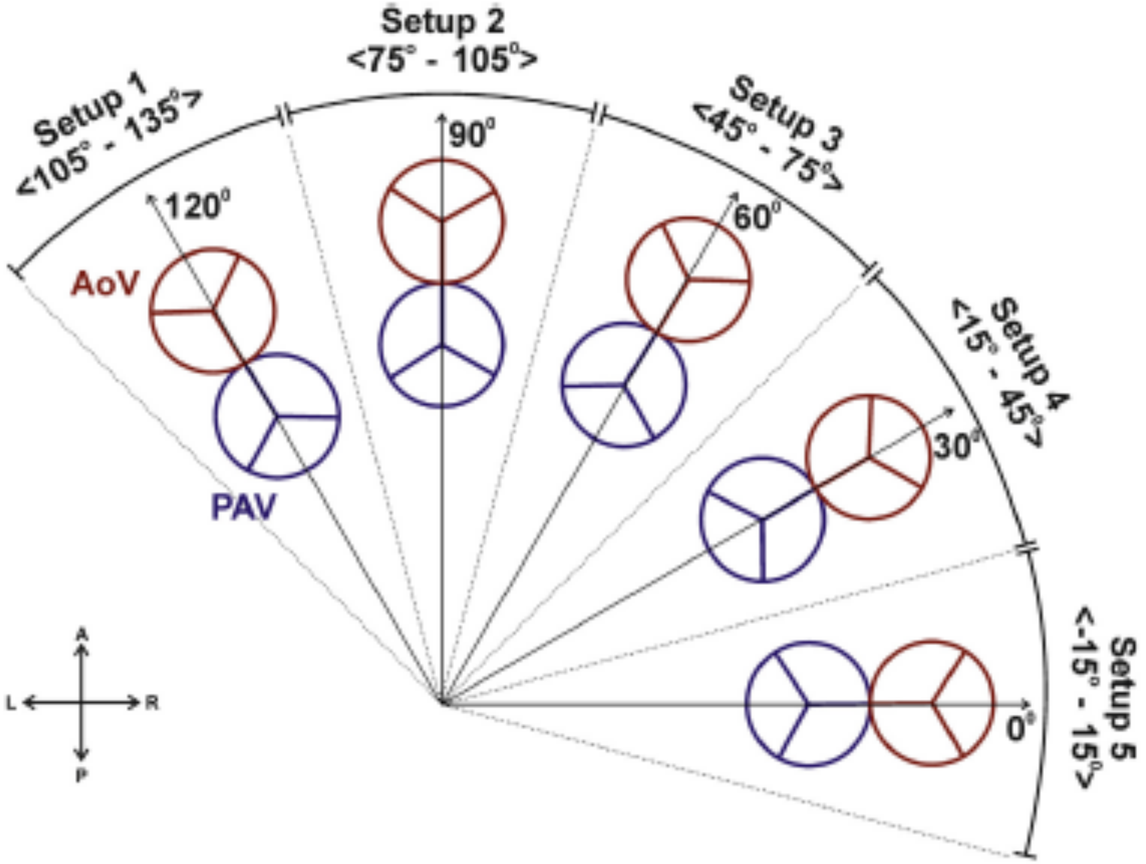


Type D

Type E

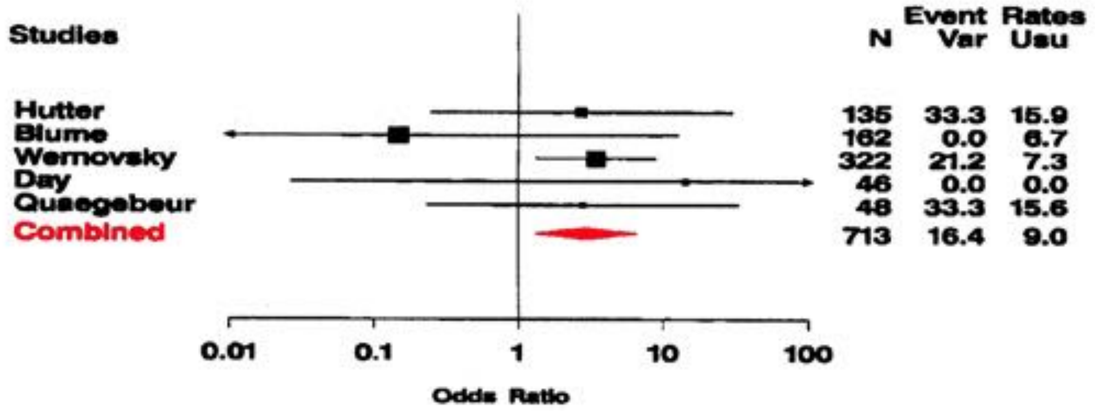




Coronary configuration	Setup 1 (n=83)	Setup 2 (n=246)	Setup 3 (n=263)	Setup 4 (n=58)	Setup 5 (n=65)
usual (n=474)	65 (78.3%)	198 (80.5%)	178 (67.7%)	21 (36.2%)	14 (21.5%)
anomalies (n=241)	18 (21.7%)	48 (19.5%)	85 (32.3%)	37 (63.8%)	51 (78.5%)

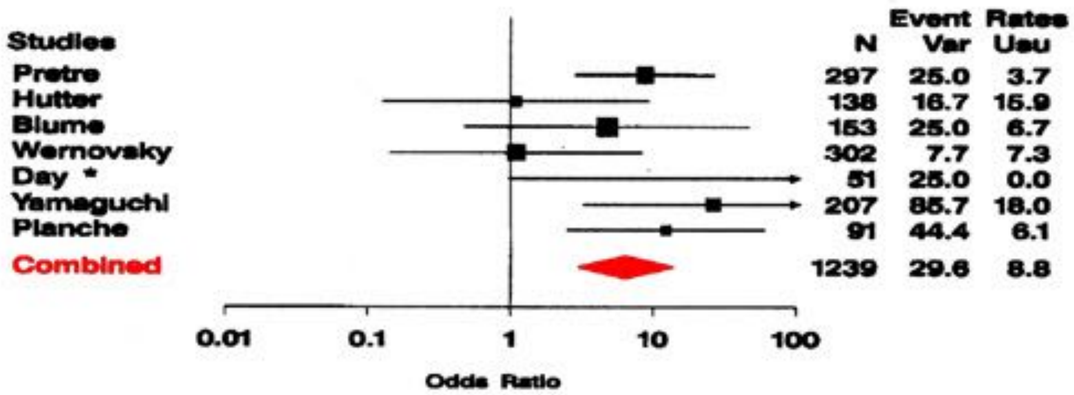
Coronary artery anatomy as a risk factor for early events

Single coronary artery



OR = 2,9

Intramural course



OR = 6,5

Pasquali, 2002

Outcomes and predictors of early mortality of the ASO for TGA with IVS

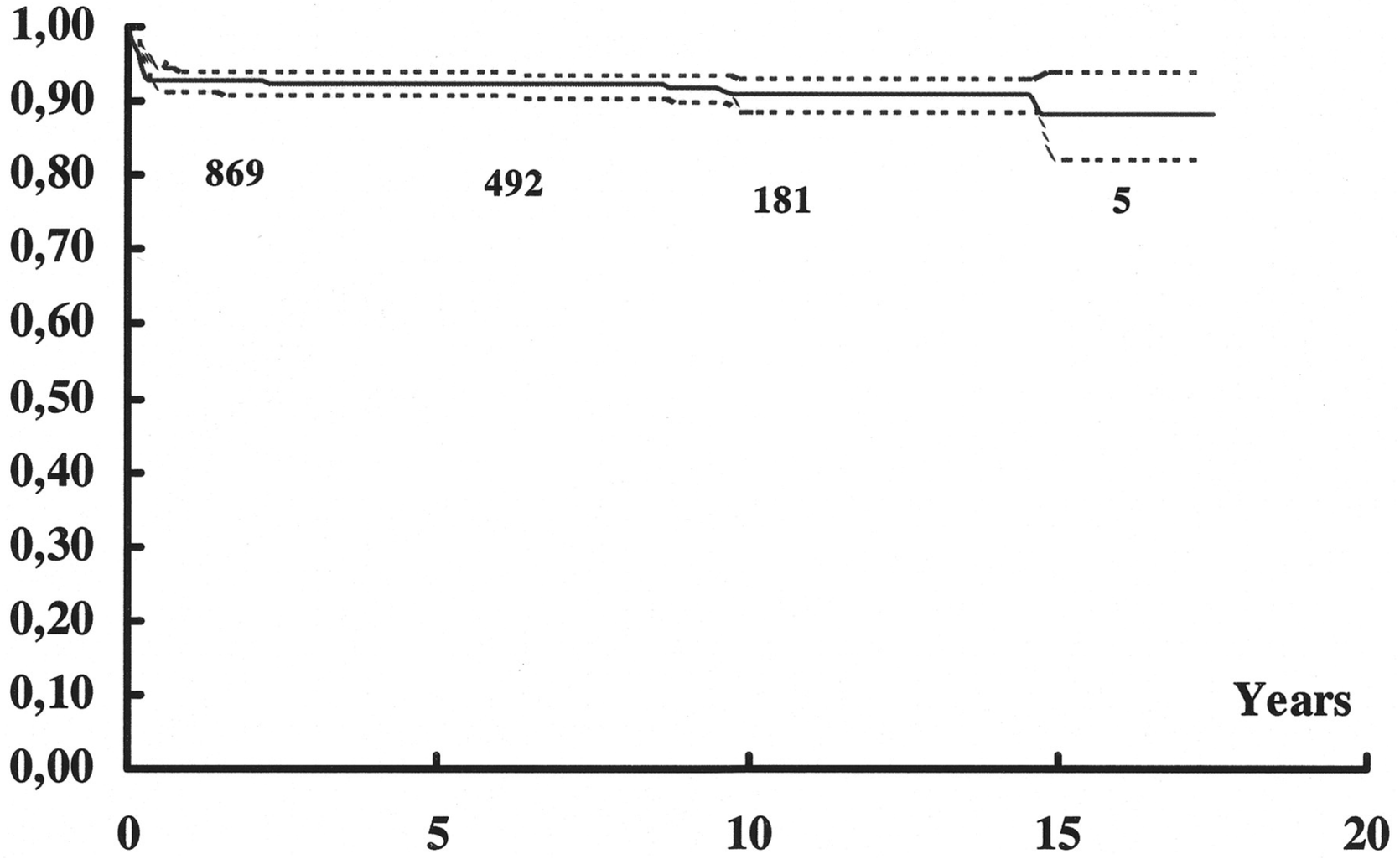
First Author (Ref. #), Year	Inclusive Years	N	% IVS	Early Survival for TGA IVS, %	5-Year Survival, %	10-Year Survival, %	Coronary Anatomic Risk Factors	Other Predictors of Early Mortality
Sarris (43), 2006*	1998-2000	613	70	97	NA	NA	Single coronary (univariate analysis only)	Open sternum
Lalezari (51), 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 (85), 2012	1983-2009	618	64	98.2	98	97	Not a risk factor for early mortality	Weight <2.5 kg ECMO
Khairy (41), 2013	1983-1999	400	59.5	93.5†	NA	92.7†	Single right coronary artery	Post-operative heart failure
Cain (52), 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 decreased resource utilization
Anderson (24), 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 decreased resource utilization

Postoperative sequelae following the arterial switch operation

Prevalence of coronary lesions or potential coronary events

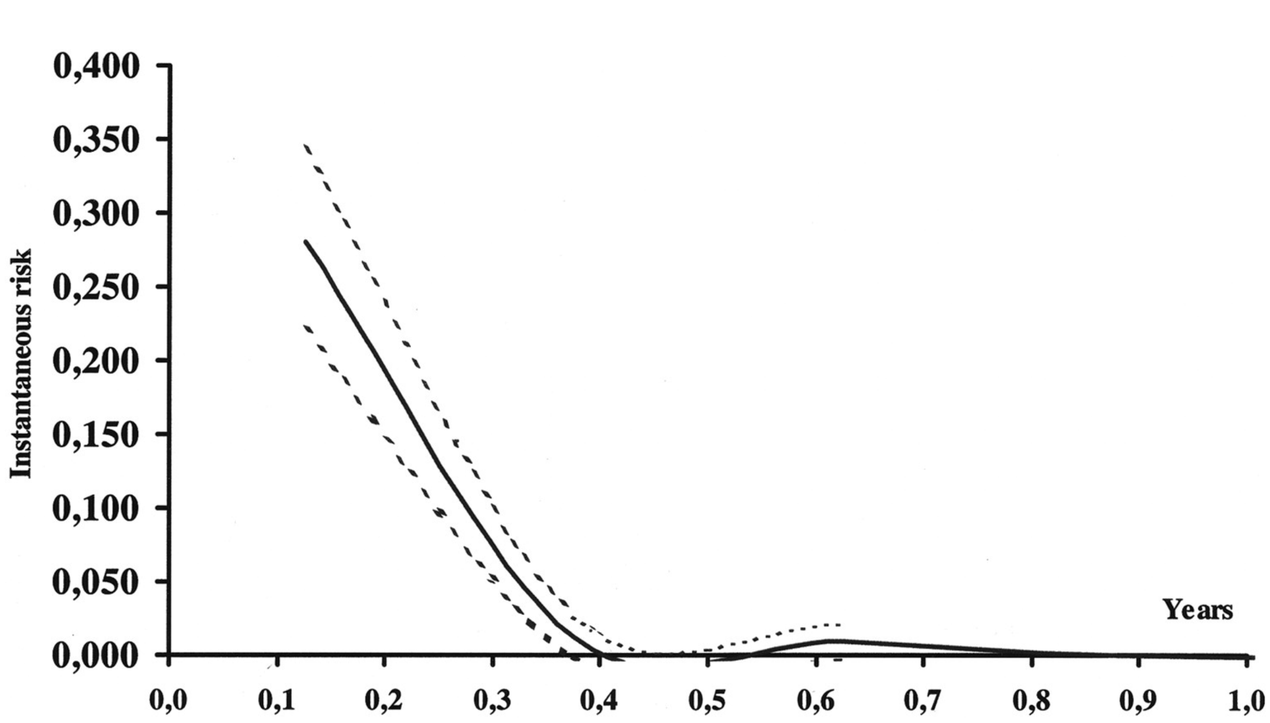
Long-Term Post-Operative Sequelae	Incidence
Supravalvular pulmonary stenosis*	~10%
Supravalvular aortic stenosis*	~5%
Neoaortic root dilation	Nearly universal
Neoaortic regurgitation	Most (moderate or severe in <10%)
Asymptomatic coronary occlusion	2%-7%
Sudden cardiac death	<1%
Arrhythmia	2%-10%
Aortic dissection or rupture	Unknown

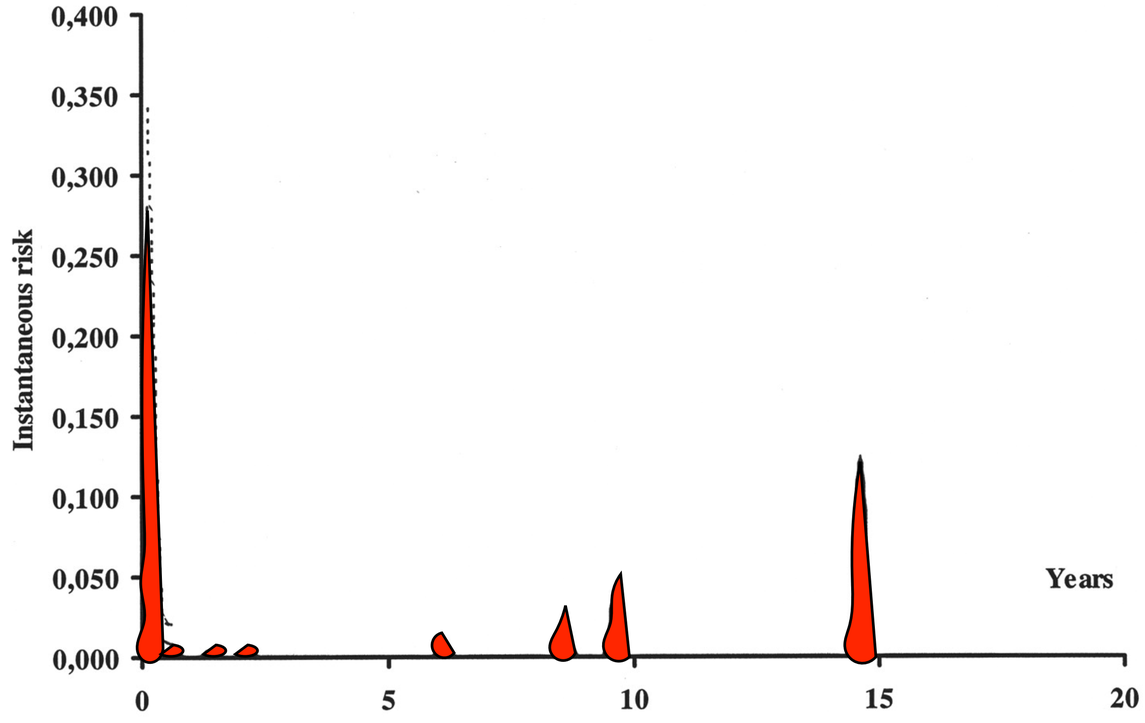
Actuarial survival free of coronary events for 1304 patients



Legendre A et al. Circulation. 2003;108:II-186-II-190

Hazard function for coronary « events » for 1304 patients

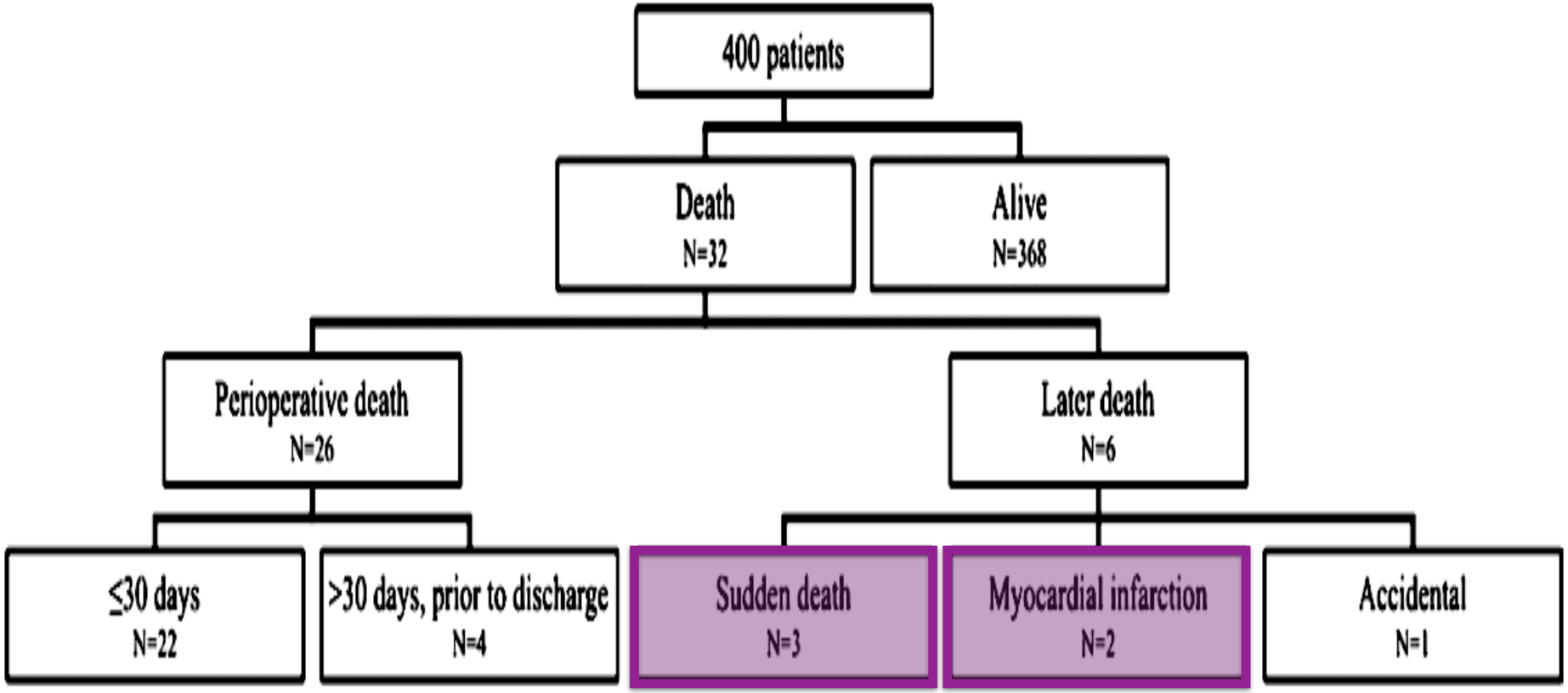




Legendre A et al. Circulation. 2003;108:II-186-II-190

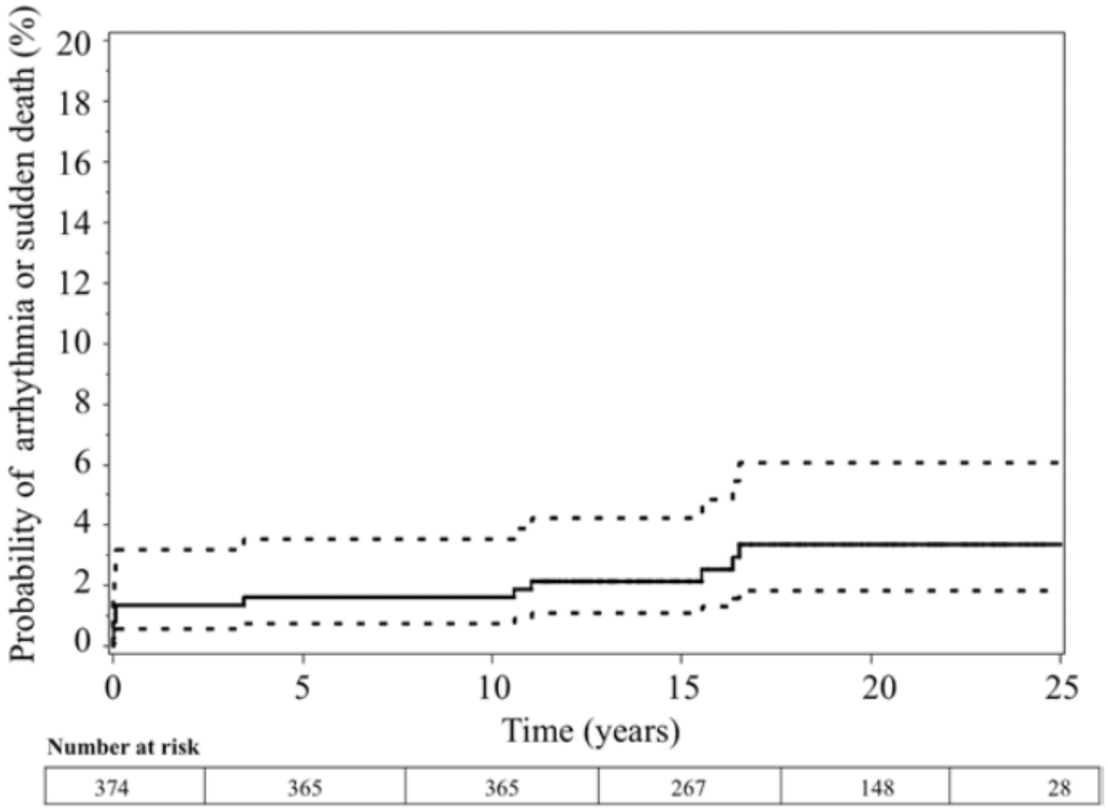
Postoperative sequelae following the arterial switch operation

Prevalence of coronary lesions or potential coronary events

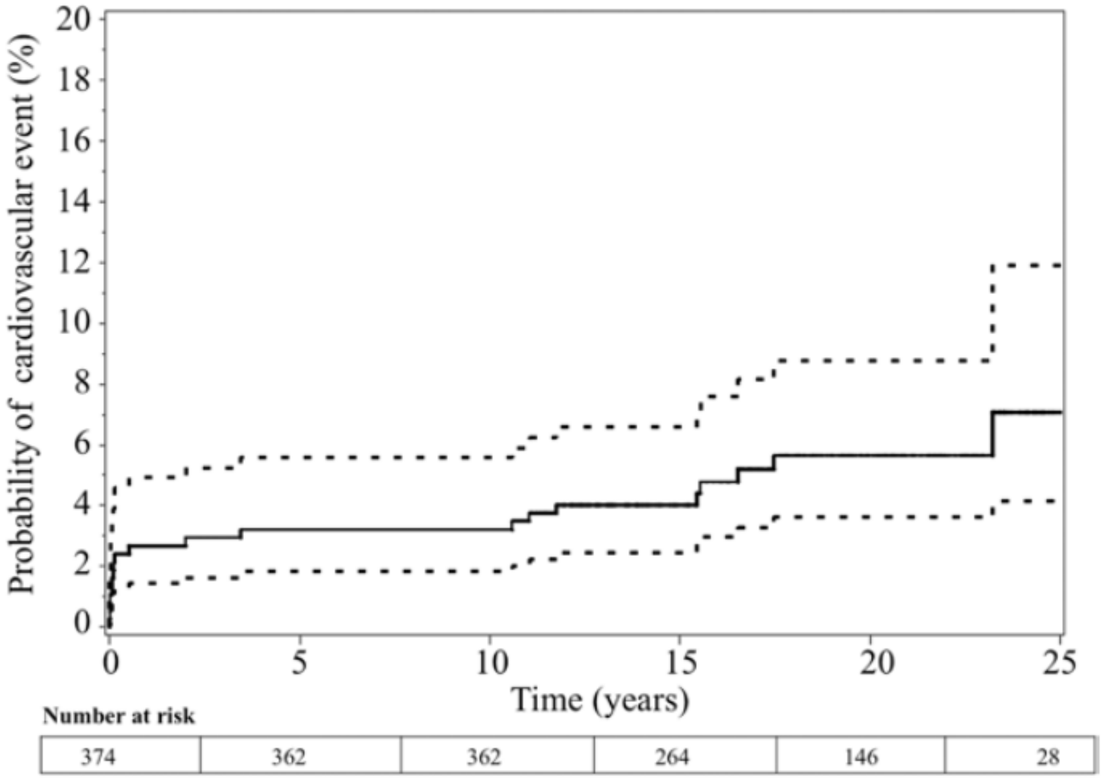


Khairy P et al. *Circulation*. 2013;127:331–339.

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.

Sudden death due to coronary artery lesions long-term after the arterial switch operation: a systematic review

- 52 studies : sudden death because of coronary complications in survivors after 5 years
- 8798 patients: 27 deaths > 5 years post-ASO (0.3%)
  - 10 were known with relevant residual lesions
  - 5 sudden death possibly from cardiac cause, no late death confirmed to be coronary related
- **Routine coronary imaging of asymptomatic single-stage ASO patients is not justified**

M3C

van Wijk SWH et al. Can J Cardiol 2017 33;1180-87

Life-long management of the ASO population poses several challenges

The coronary arteries after transfer

3) there is no defined management strategy when subclinical anatomic or physiologic abnormalities are identified;

What to do with abnormal coronary arteries in the absence of symptoms ?

4) the effects of acquired coronary artery disease superimposed on manipulated coronary arteries remain unknown.

Are there some « at risk » patients who would need systematic screening for myocardial ischemia ?

M3C

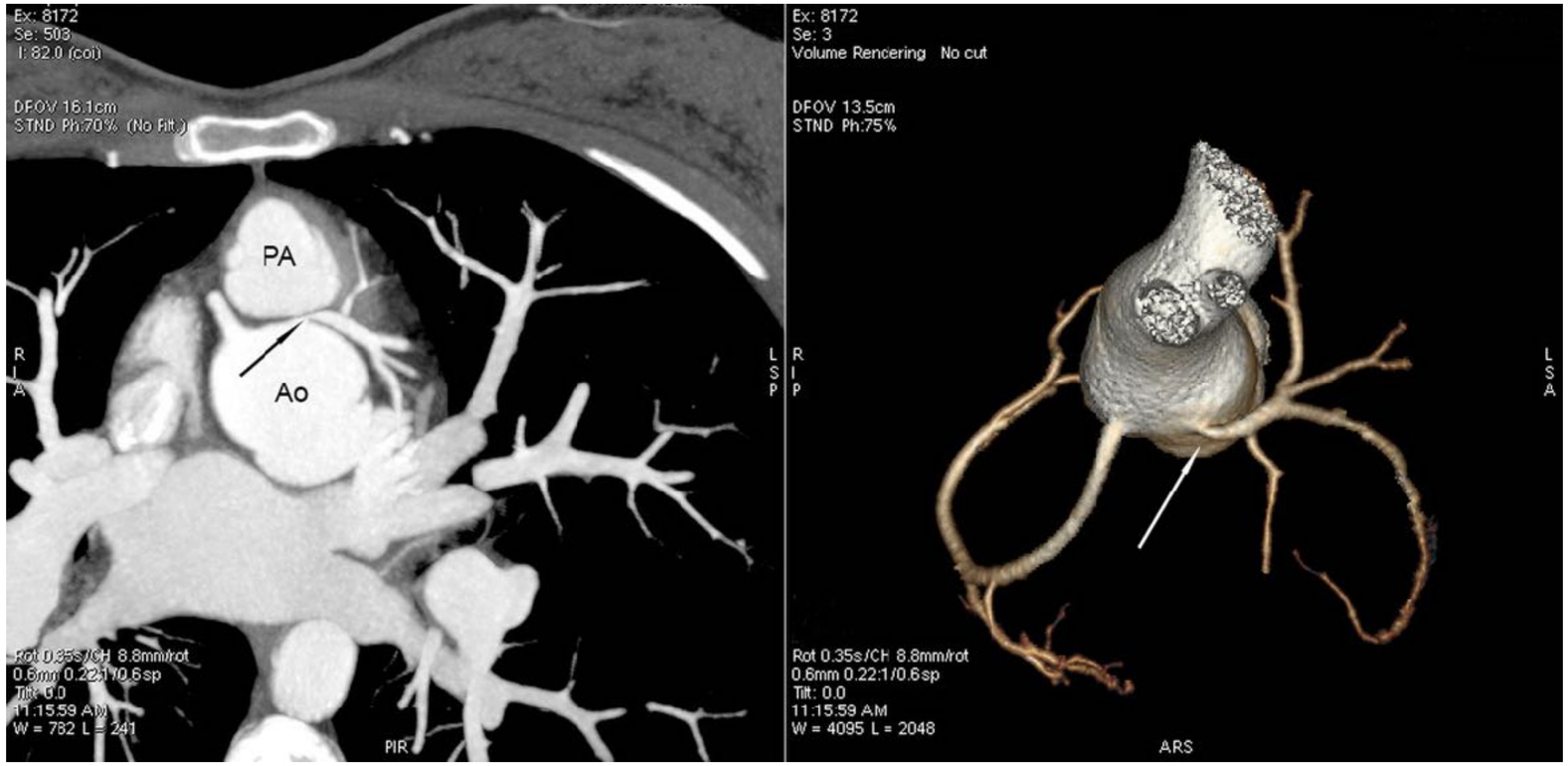
Villafane J et al. J Am Coll Cardiol 2014; 64: 498-511

Parisian experience in systematic screening

1453 patients diagnosed with / or screened for coronary anomalies after ASO

78 coronary artery obstructions

Prevalence of asymptomatic coronary artery obstruction (> 30%) : 5.3%



Bonnet D et al. Heart 1996;76:274-9
Bonhoeffer P et al . J Am Coll Cardiol 1997;29:202-6
Szymczyk K et al. Pediatr Cardiol 2018;39:335-46

Parisian experience in systematic screening

1453 patients diagnosed with or screened for coronary anomalies after ASO

Circumstances of diagnosis of coronary artery obstruction

1) Potential coronary event : 29 (37%)

Clinical (chest pain, near-syncope or syncope, heart failure) : 6
ECG and/or echocardiographic signs of myocardial ischemia : 23

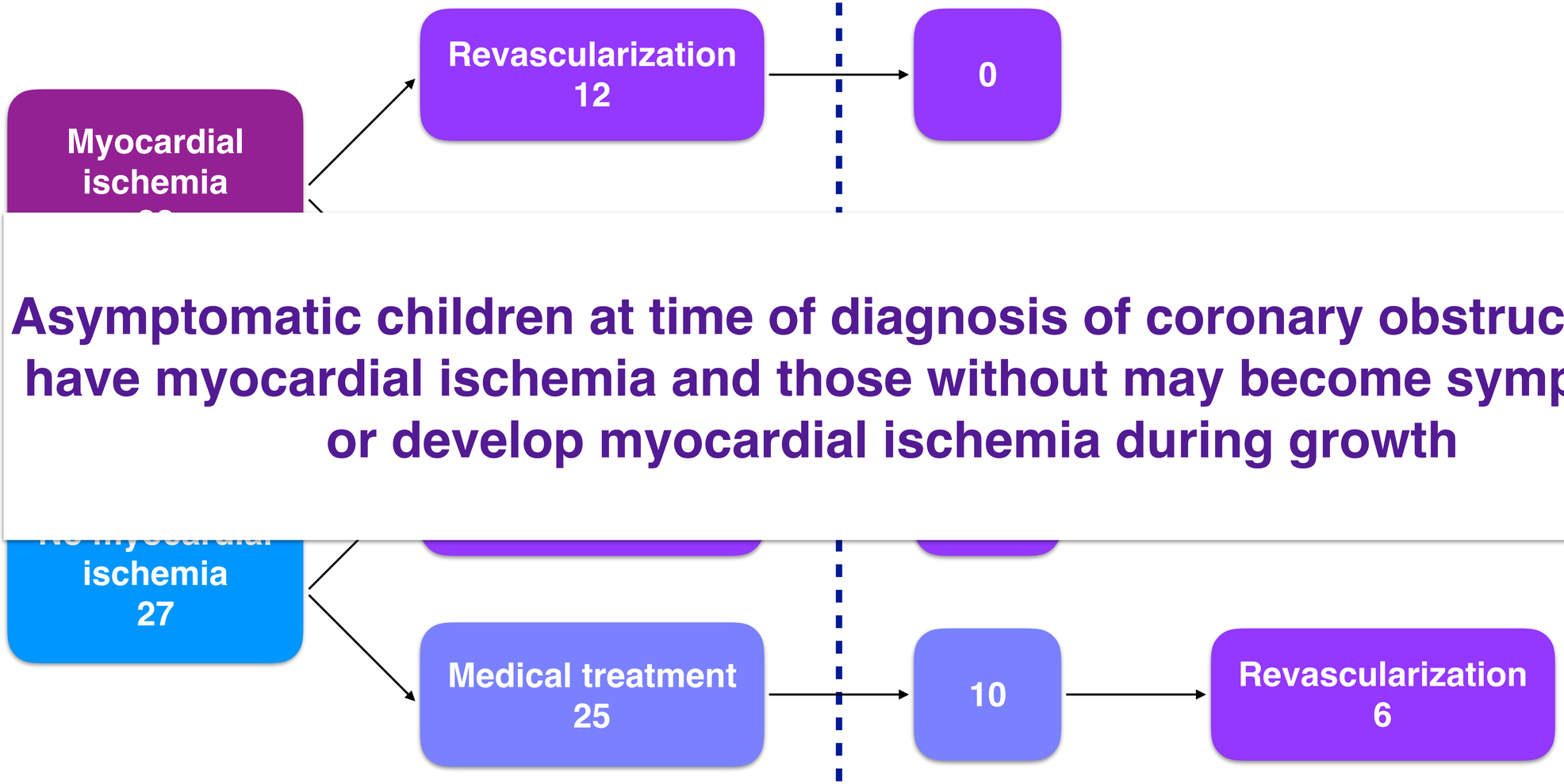
Age at potential coronary event < 6 months in 22 and > 6 years in 7

*Higher proportion of **Left main stem stenosis** compared to asymptomatic children and higher proportion of **severe stenosis > 50%** compared to mild stenosis and occlusion of the coronary artery*

2) Systematic screening in asymptomatic children : 49 (73%)

Parisian experience in systematic screening

49 asymptomatic patients with coronary obstruction



PCE Median follow-up 6.8 years

Mechanisms of coronary artery complications after the ASO for TGA

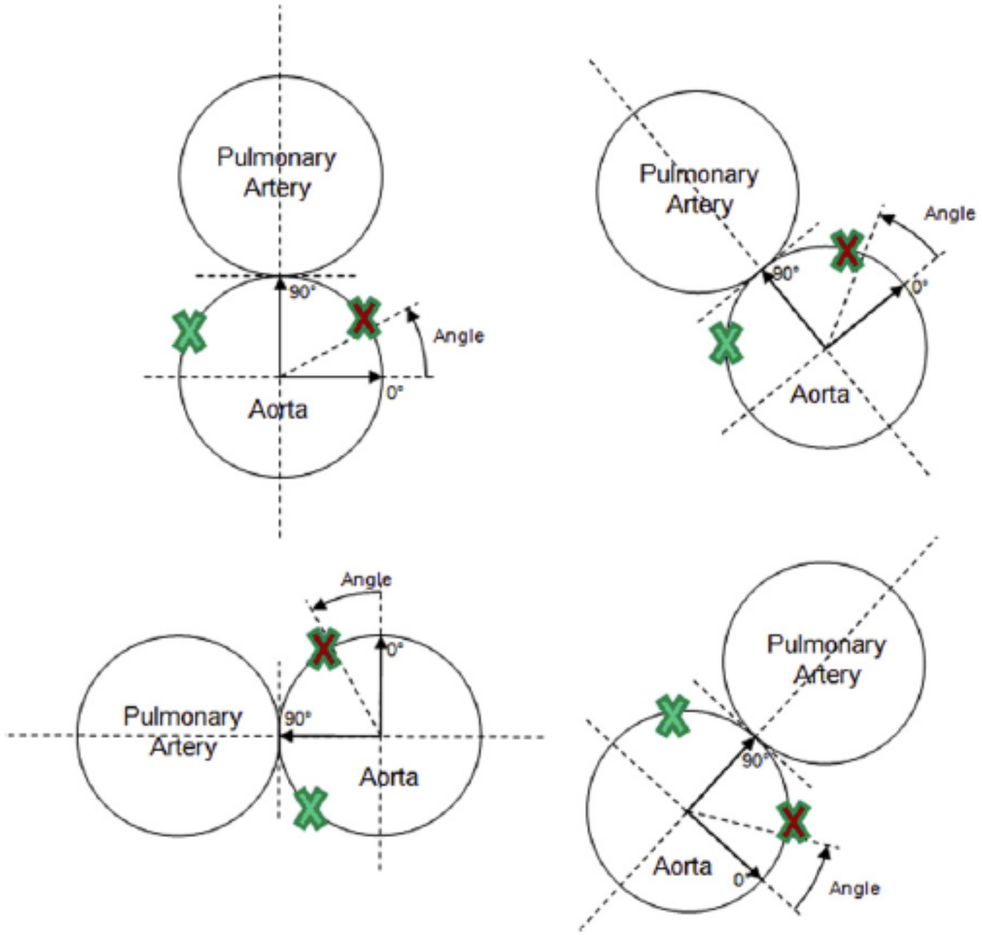


FIGURE 1. Determination of the coronary angles. The commonest positioning of the greats arteries and the coronaries after ASO are representing here. The coronary angles were determined as follows: (1) The aortopulmonary centerline that passes through the center of both the aorta and the pulmonary artery was drawn; (2) the perpendicular of the aortopulmonary centerline that passes through the center of the aorta is the reference line (0°); (3) the angle between the coronary ostia (*red cross,* left ostium; *green cross,* right ostium) and the reference line correspond to the coronary angle.

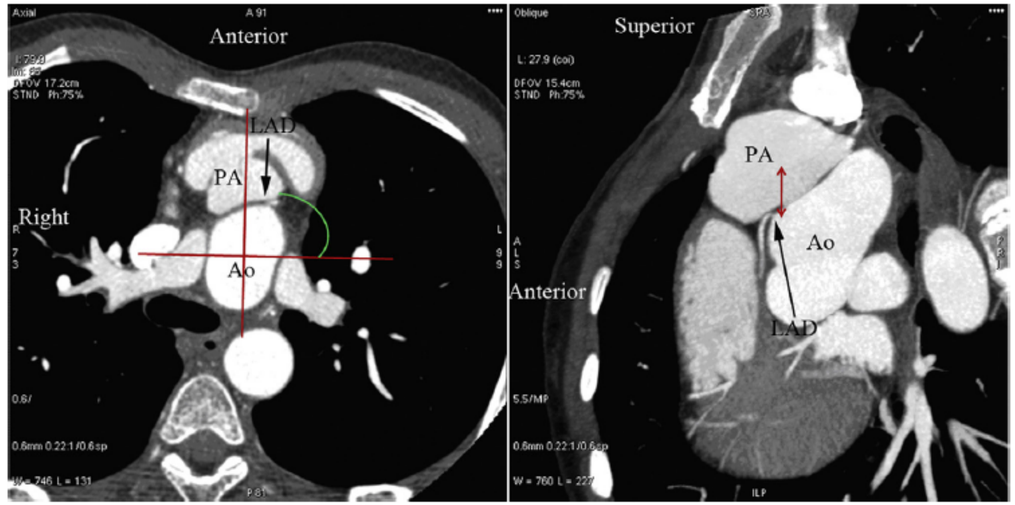


FIGURE 3. Tight stenosis of the LAD in the Yacoub type D. The reimplantation of the LAD (*black arrow* on the *left panel*) is to anterior as indicated by the left coronary angle almost equal to 90° (*green curve* on the *left panel*), and also too high above the left coronary sinus, as identified by the short coronary–pulmonary bifurcation distance (*red arrow* on the *right panel*) equal to 5 mm. *PA,* Pulmonary artery; *Ao,* aorta; *LAD,* left anterior descending artery.

FIGURE 2. Significant stenosis of the ostium of the left coronary artery by anterior compression by the pulmonary artery. The reimplantation is too anterior, as confirmed by the left coronary angle *(green curve)* almost equal to 90°, corresponding to a reimplantation at 12 o' clock. *PA,* Pulmonary artery; *Ao,* aorta.

Ou P et al. JTCVS 2013;145:1263-9

Mechanisms of coronary artery complications after the ASO for TGA

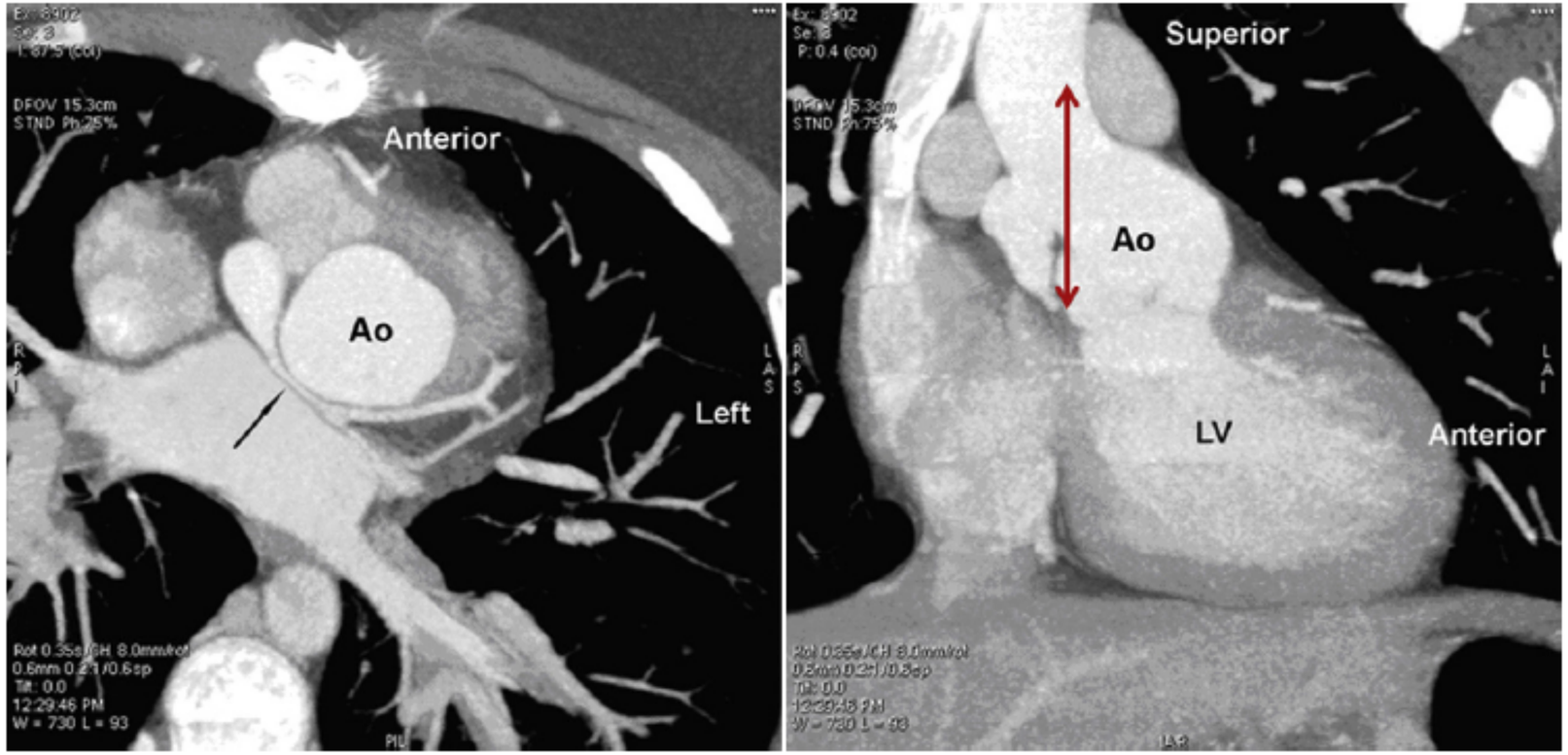


FIGURE 4. Mechanism of lesion of the circumflex artery in Yacoub type D. The reimplantation of the right ostium was too far under the right sinus, as evidenced by the long coronary–pulmonary artery distance (*red arrow* on the *right panel*). Thus, the course of the circumflex toward the left atrioventricular groove is increased, causing a stretching of the coronary artery (*black arrow* on the *left panel*). *Ao,* Aorta; *LV,* left ventricle.

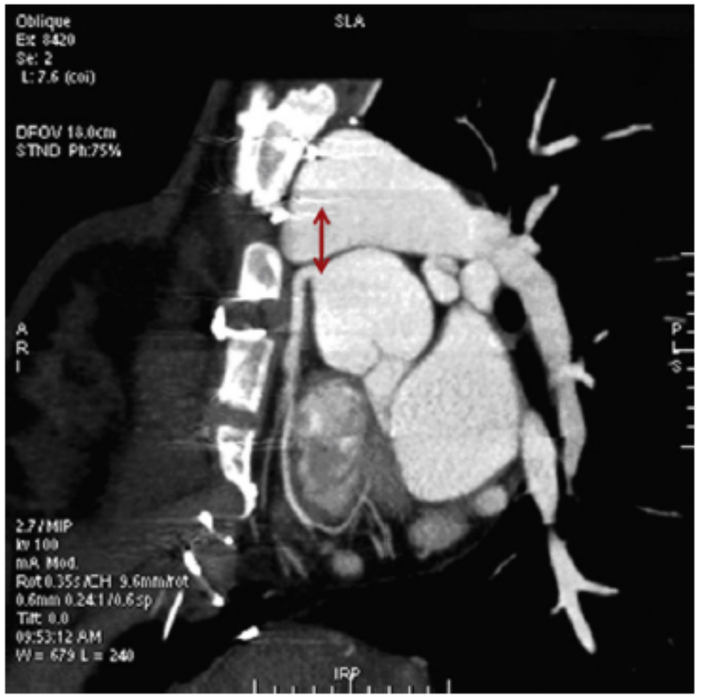


FIGURE 5. Lesion of the right coronary artery. The reimplantation of the right sinus is too high above the right sinus, as evidenced by the short coronary–pulmonary artery bifurcation distance *(red arrow)*. The right ostium was then compressed by the pulmonary artery. This patient had a long ''olasty'' of the ostium and proximal segment of the right coronary artery using saphenous vein. See the difference in caliber between the saphenous vein in the proximal segment and the native distal segment, which is thinner.

Ou P et al. JTCVS 2013;145:1263-9

Parisian experience in systematic RE-screening

Adolescents with previous normal coronary arteries at first screening

107 patients who had a normal coronary artery anatomy at first systematic screening (mean age 5.0 years, range 4-7 years).

All patients had annual follow-up. All were all NYHA functional class I. None had symptoms of myocardial ischemia. None had ECG anomalies suggestive of myocardial ischemia. None of them had LV dysfunction (global or regional) or mitral regurgitation.

Only one/107 new obstruction of RCA (stenting)

No change in the position of the coronary ostium during growth

Raimondi F et al. JACC Imaging 2017

Parisian experience in systematic RE-screening

Adolescents with previous normal coronary arteries at first screening

No qualitative perfusion defect was found.

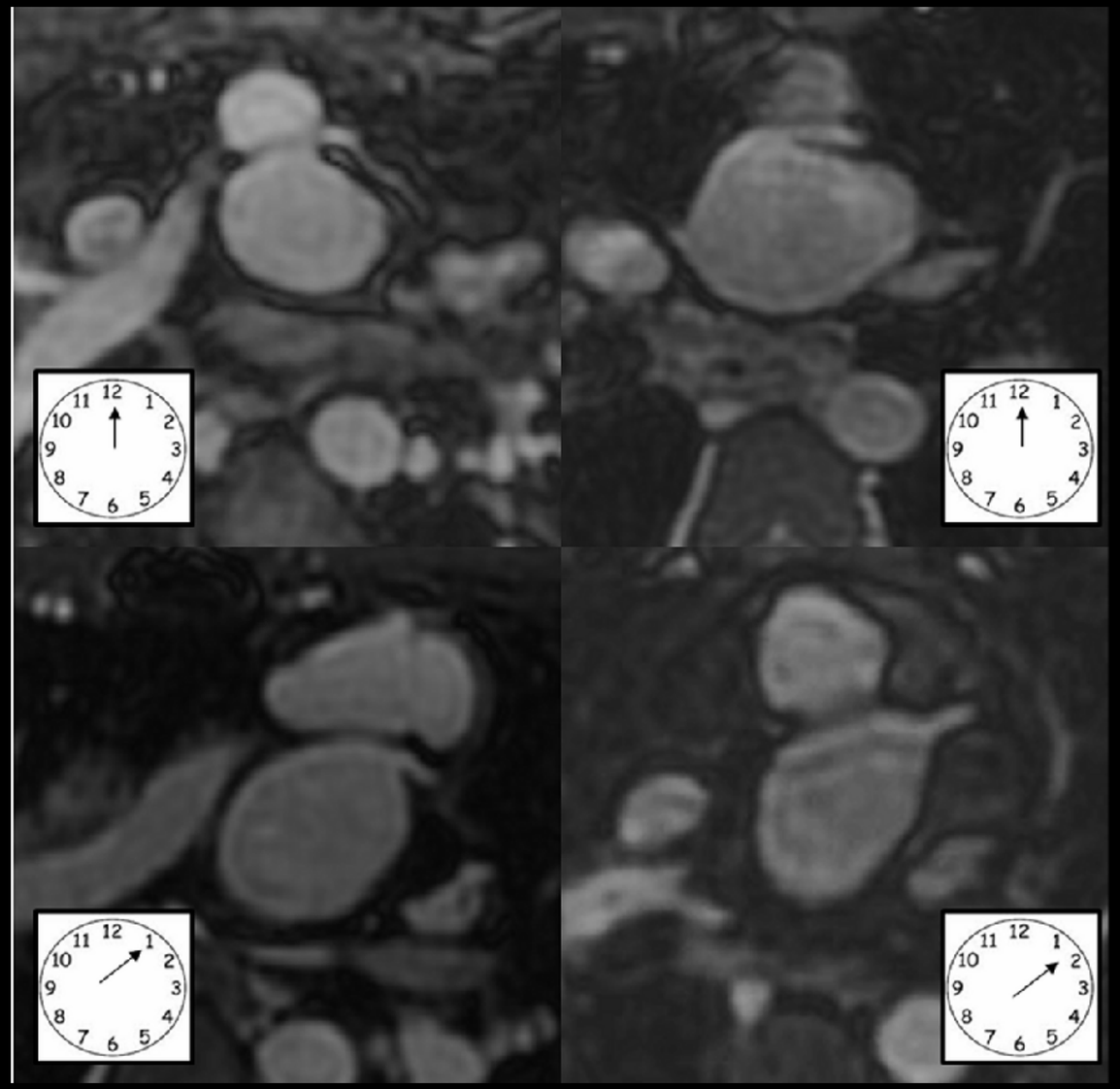
No LGE was found.

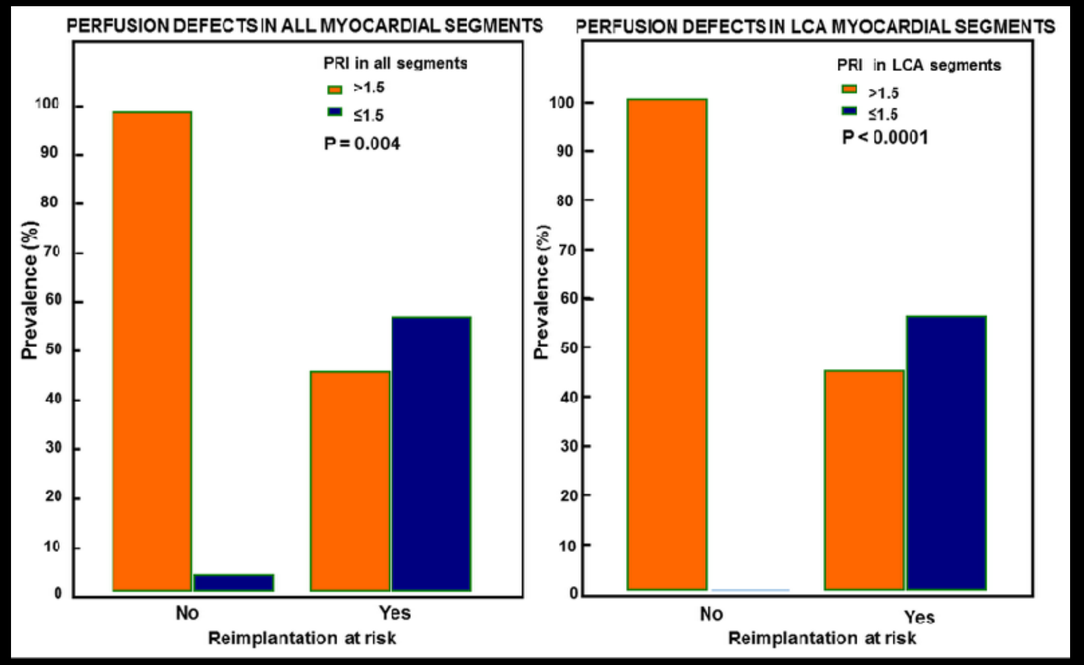
12% of patients had semi-quantitative perfusion defects (PRI <1.5 in at least 2 contiguous myocardial segments)

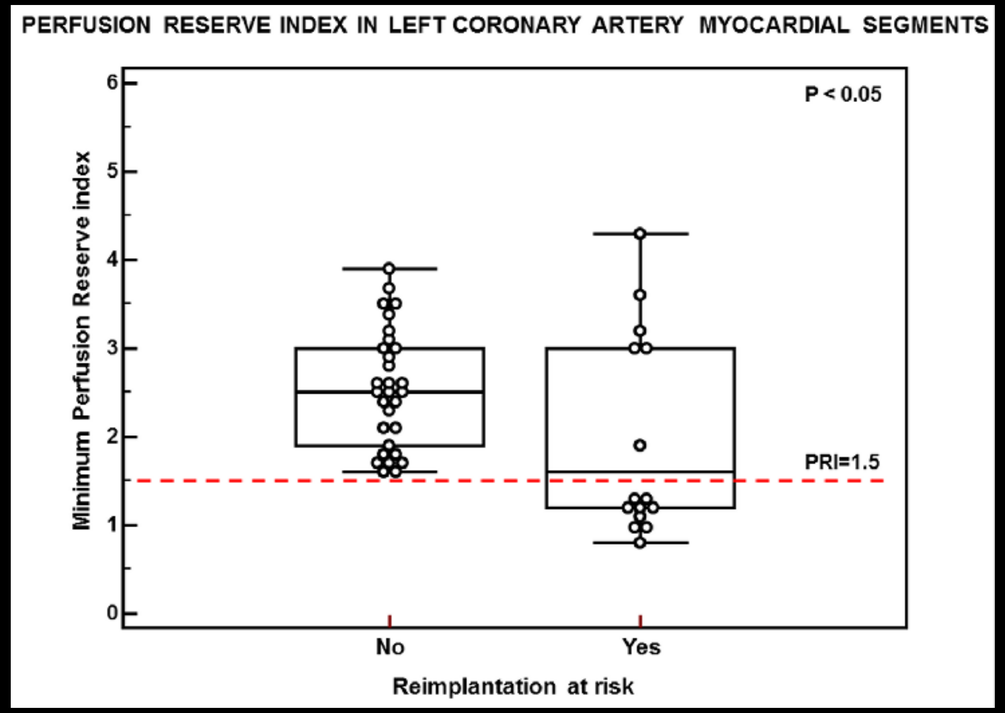
In 12 patients, the left CA was reimplanted in clock position between 12 and 1 o'clock. These patients had more frequent perfusion abnormality than patients with other sites of reimplantation (7/12 vs 0/45; p=0.0001).

The 12 patients with left CA reimplanted in clock position 12-1 o'clock had significantly lower PRI in myocardial segments irrigated by left CA myocardial compared to patients with other sites of reimplantation (2.0±1 vs 2.7±1, p<0.05).

Raimondi F et al. JACC Imaging 2017







Raimondi F et al. JACC Imaging 2017

3D
Ex: 8420
Se: 2
Volume Rendering No cut

SPR

HOPITAL NECKER ENFANT
****
F 13 0193049445

DFOV 18.9cm
STND Ph:75%

POST ASO

A
R
S

No VOI
kv 100
mA Mod.
Rot 0.35s/CH 9.6mm/rot
0.6mm 0.24:1/0.6sp
Tilt: 0.0
09:53:12 AM
W = 4095 L = 2048

IAL

3D2
Ex: 219
Se: 3 +c
Volume Rend

SPR

HOPITAL NECKER EN

AOLCA from Right sinus

DFOV 20.0cm
STND Ph:75% (No Filt.)

R
A
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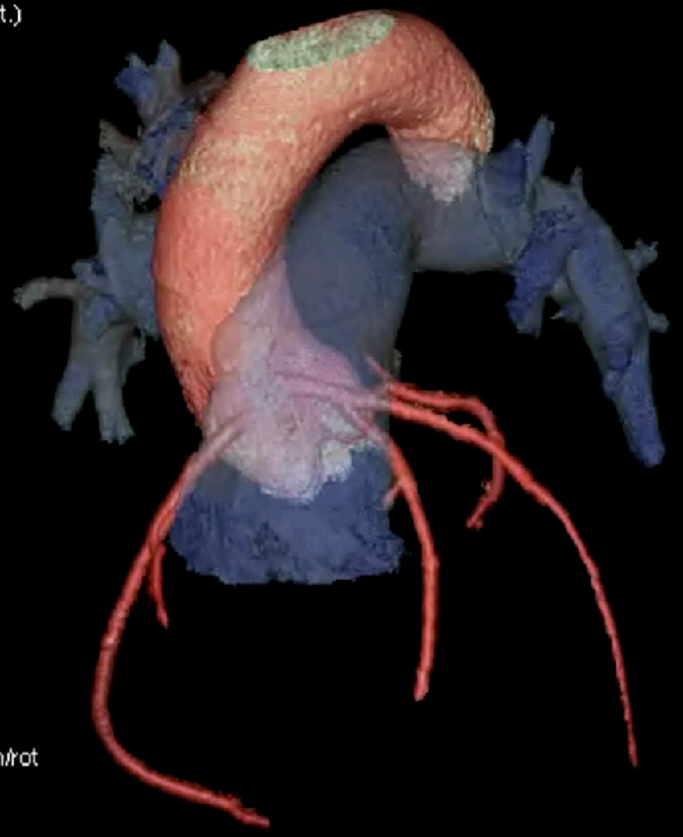

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0.6mm 0.26:1/0.6sp
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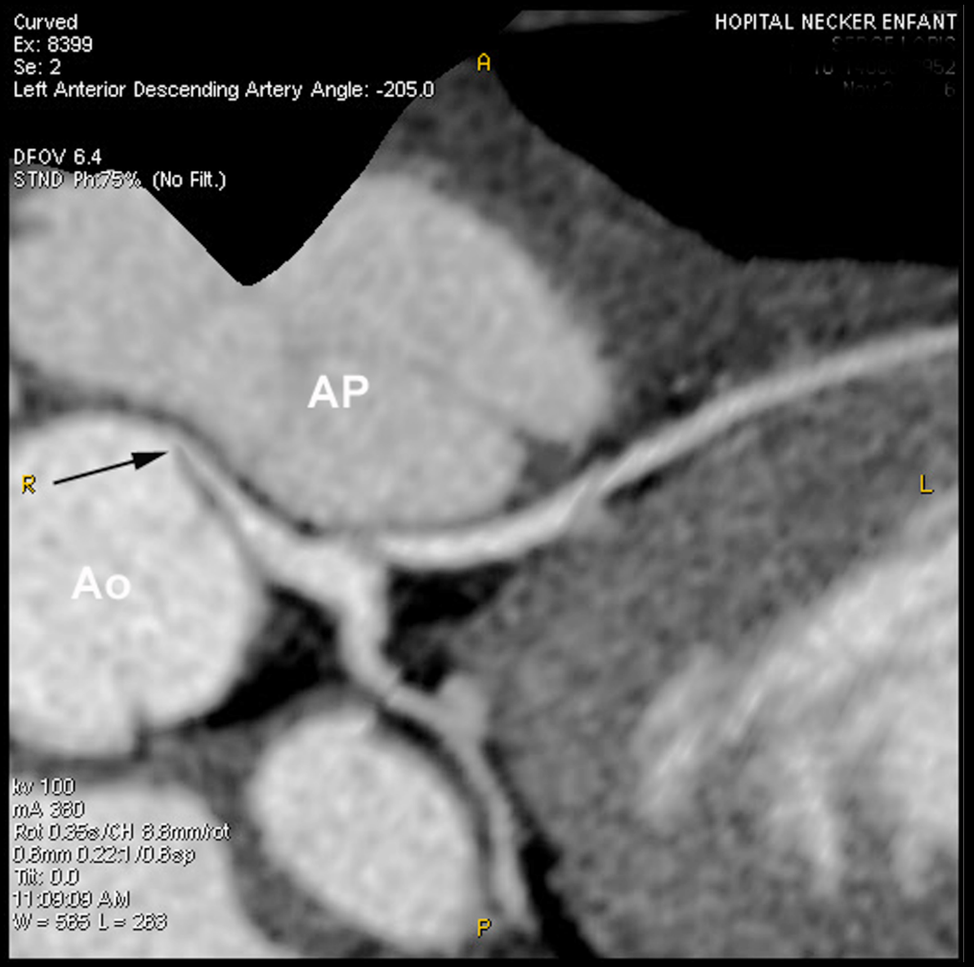
12:01:12 PM
W = 4095 L = 2048

IAL


Curved
Ex: 8399
Se: 2
Left Anterior Descending Artery Angle: -205.0
HOPITAL NECKER ENFANT
A
DFOV 6.4
STND Ph:75% (No Filt.)
AP
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L
Ao
kv 100
mA 380
Rot 0.35s/CH 6.8mm/rot
0.6mm 0.22:1 /0.6sp
Tilt: 0.0
11:09:09 AM
W = 585 L = 263
P


Se: 2
I: 61.0 (coi)
P 13 0193049445
Nov 22 2006
DFOV 13.9cm
STND Ph:75%
PA
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S
L
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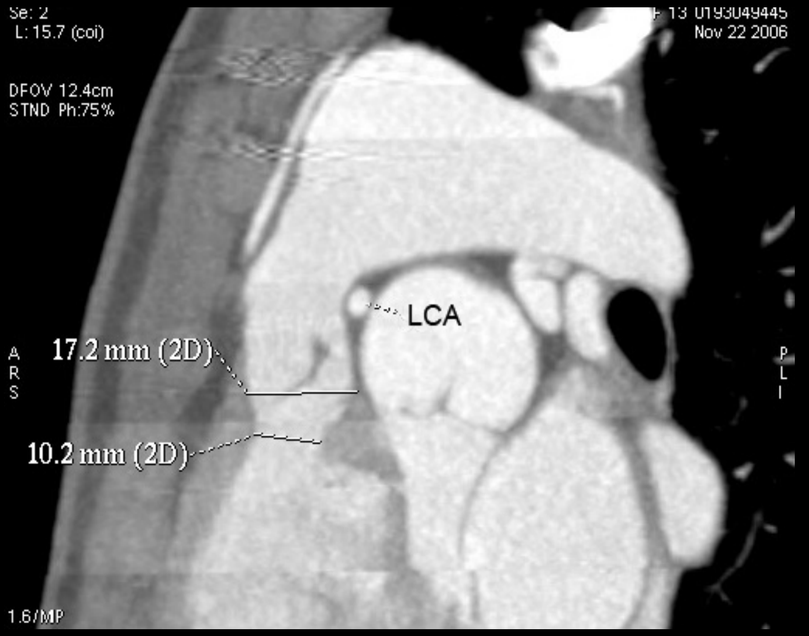
1.6/MP


Se: 2
L: 15.7 (coi)
P 13 0193049445
Nov 22 2006
DFOV 12.4cm
STND Ph:75%
LCA
17.2 mm (2D)
10.2 mm (2D)
A
R
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P
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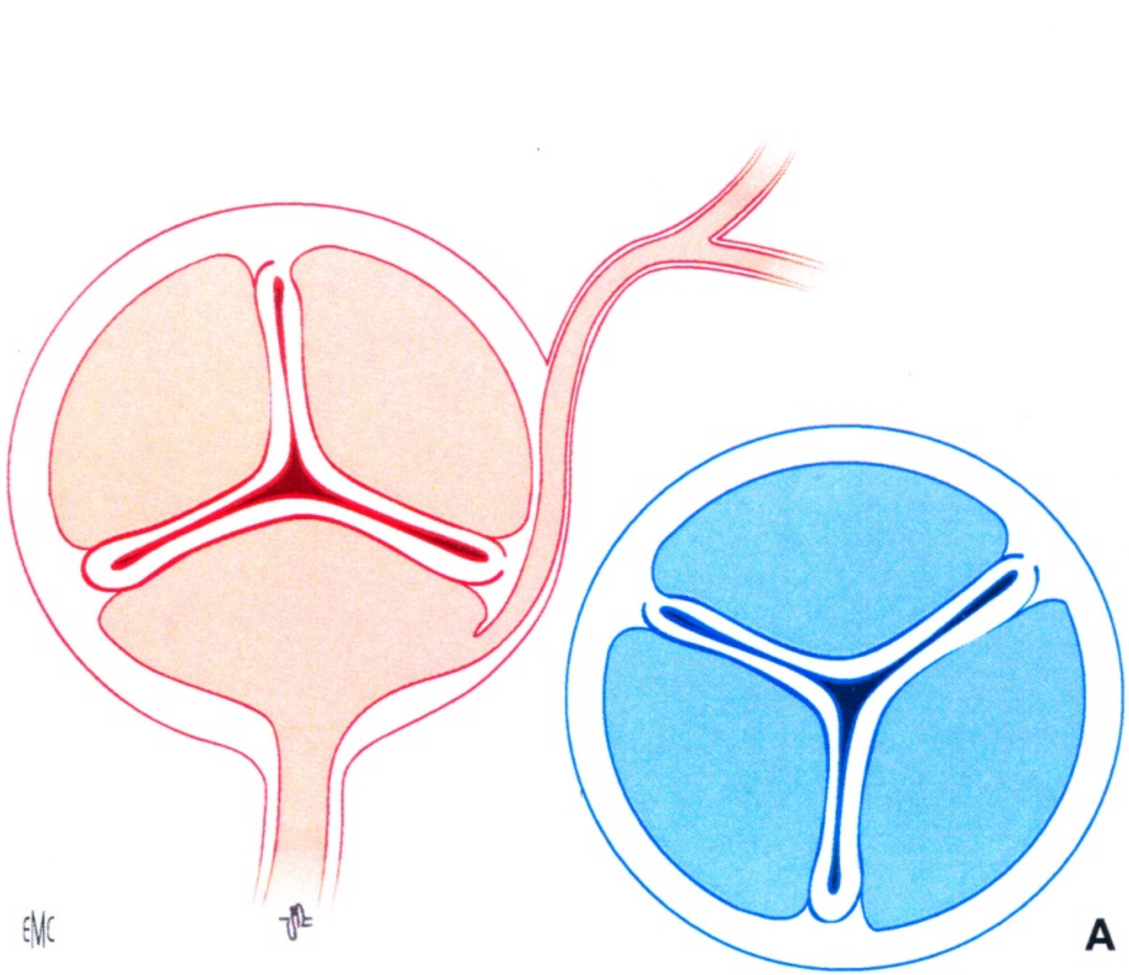
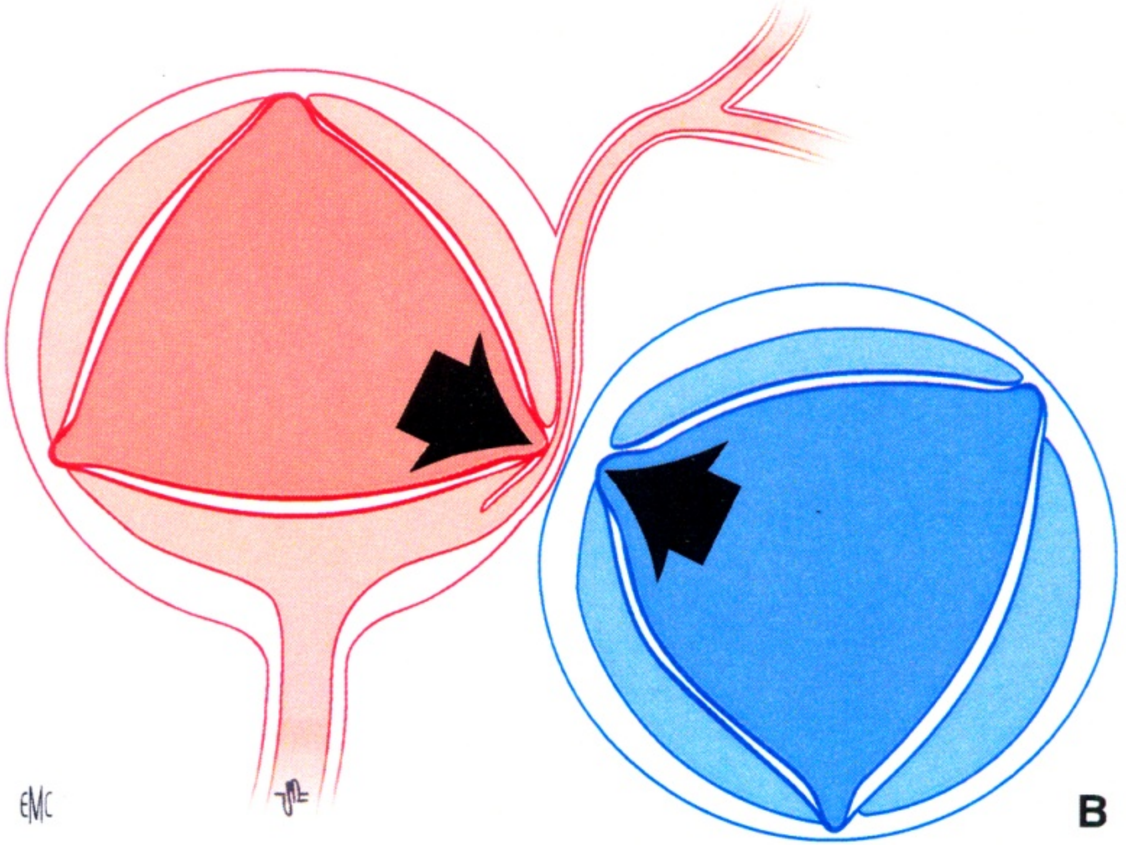
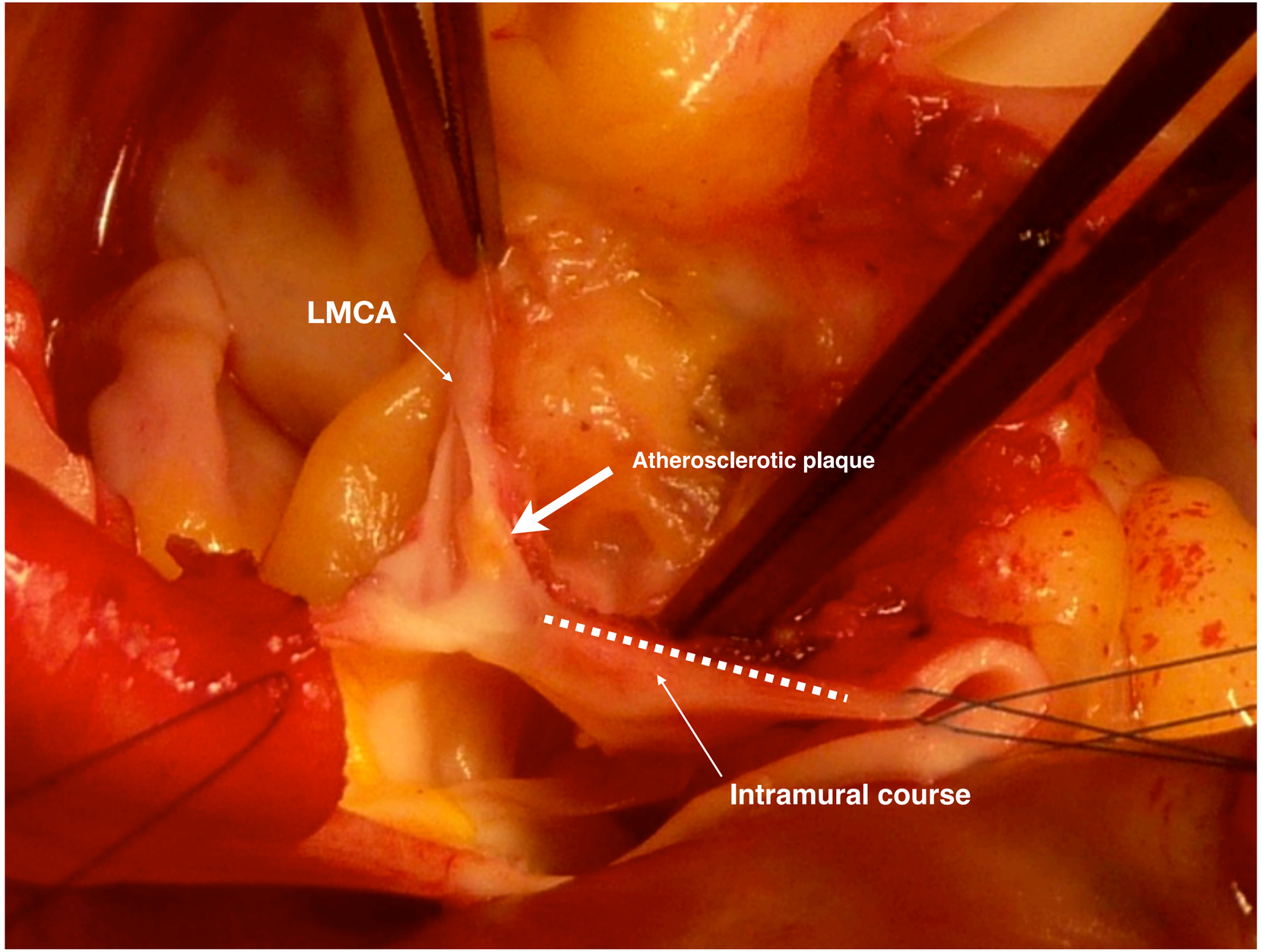
1.6/MP

What will happen with neo-aortic root dilatation ?






LMCA
Atherosclerotic plaque
Intramural course

Who are the patients at risk after coronary transfer in ASO?

Type B and intramural course : early mortality

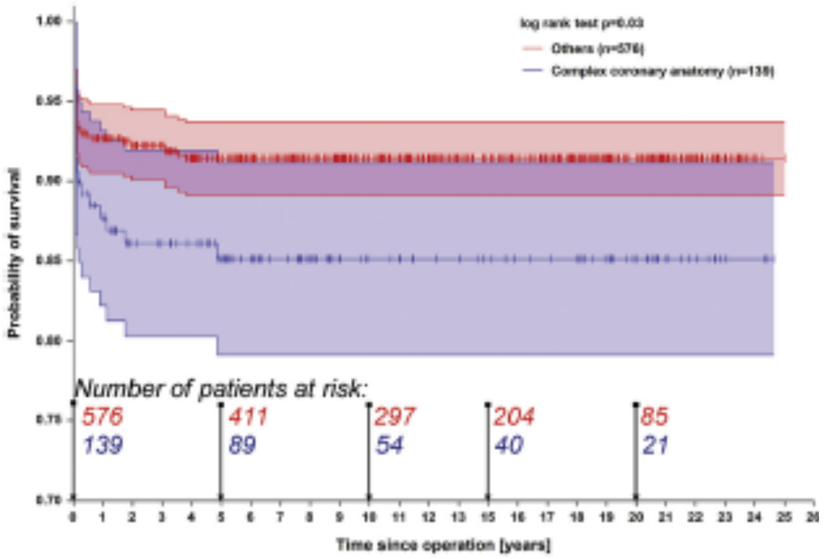
Anterior preimplantation of LCA : higher risk of obstruction, lower PRI

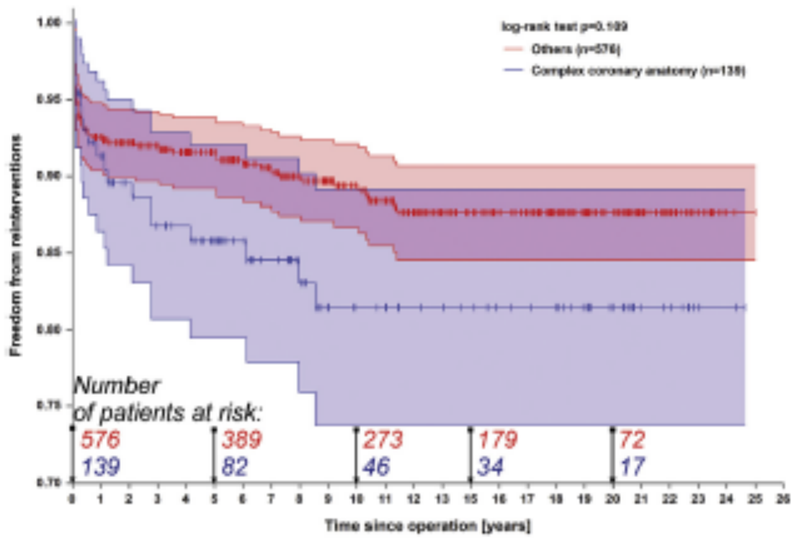
The objective of imaging coronary arteries in TGA is NOT ONLY detection of obstruction but screening for patients who are at risk of late coronary events

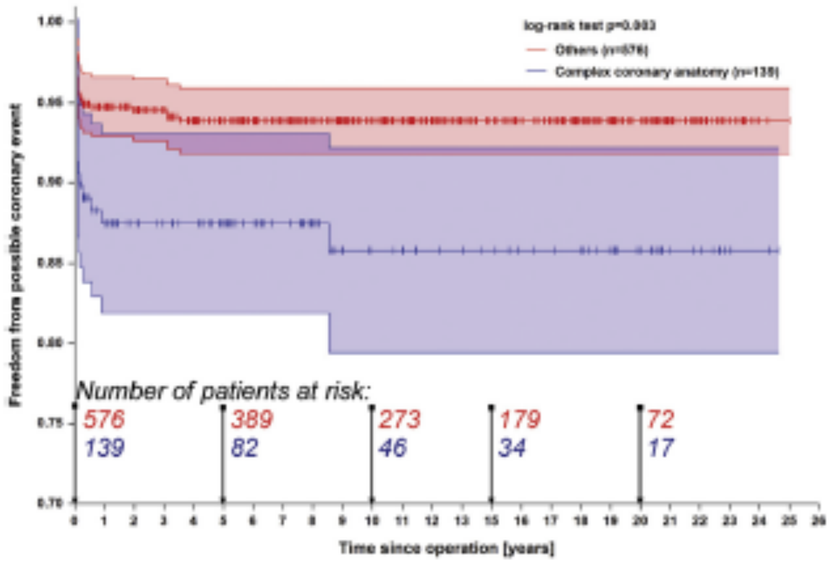
Batteux C et al. In preparation

Are there patients « at risk » for late coronary events ?

Actuarial KM according to the complexity of coronary anatomy after the ASO

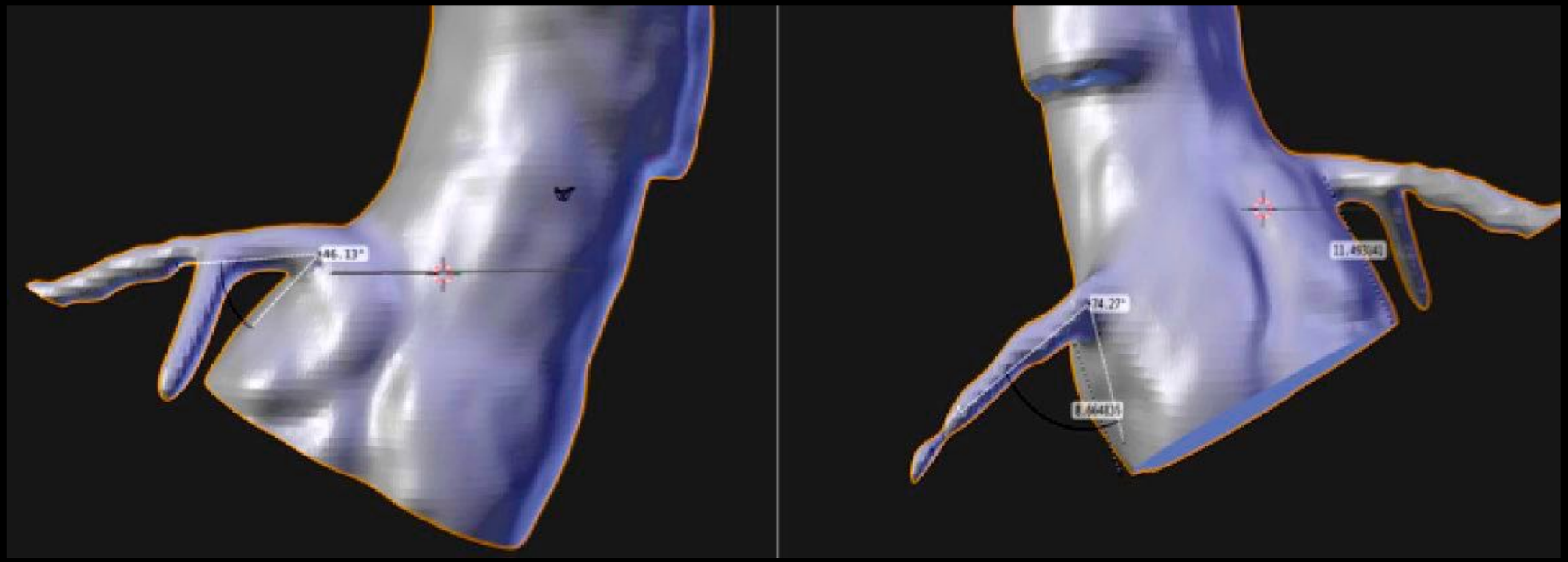






Survival **Re-operation** **Potential coronary events**

Angle of LCA with aortic wall



Batteux C et al. in preparation

Recommendation	Class[a]	Level[b]	Ref[c]
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
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 transluminar 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.
[a]Class of recommendation.
[b]Level of evidence.
[c]References.
Recommendations for reoperations for residual or recurrent coronary lesions

Sarris GE et al. CiTY 2017

Conclusion

Systematic screening in ALL ASO patients is probably not efficient.

Coronary events are rare but they do exist.

The risk factors for late events is not only the initial coronary anatomy but also the acquired coronary anatomy.

Patients with close relationship between great vessels and initial coronary course might be at risk for myocardial perfusion anomalies and potentially for late coronary events.

Should we image all patients to identify this « at risk » population ?

Thank you