Long term outcome after coarctation repair

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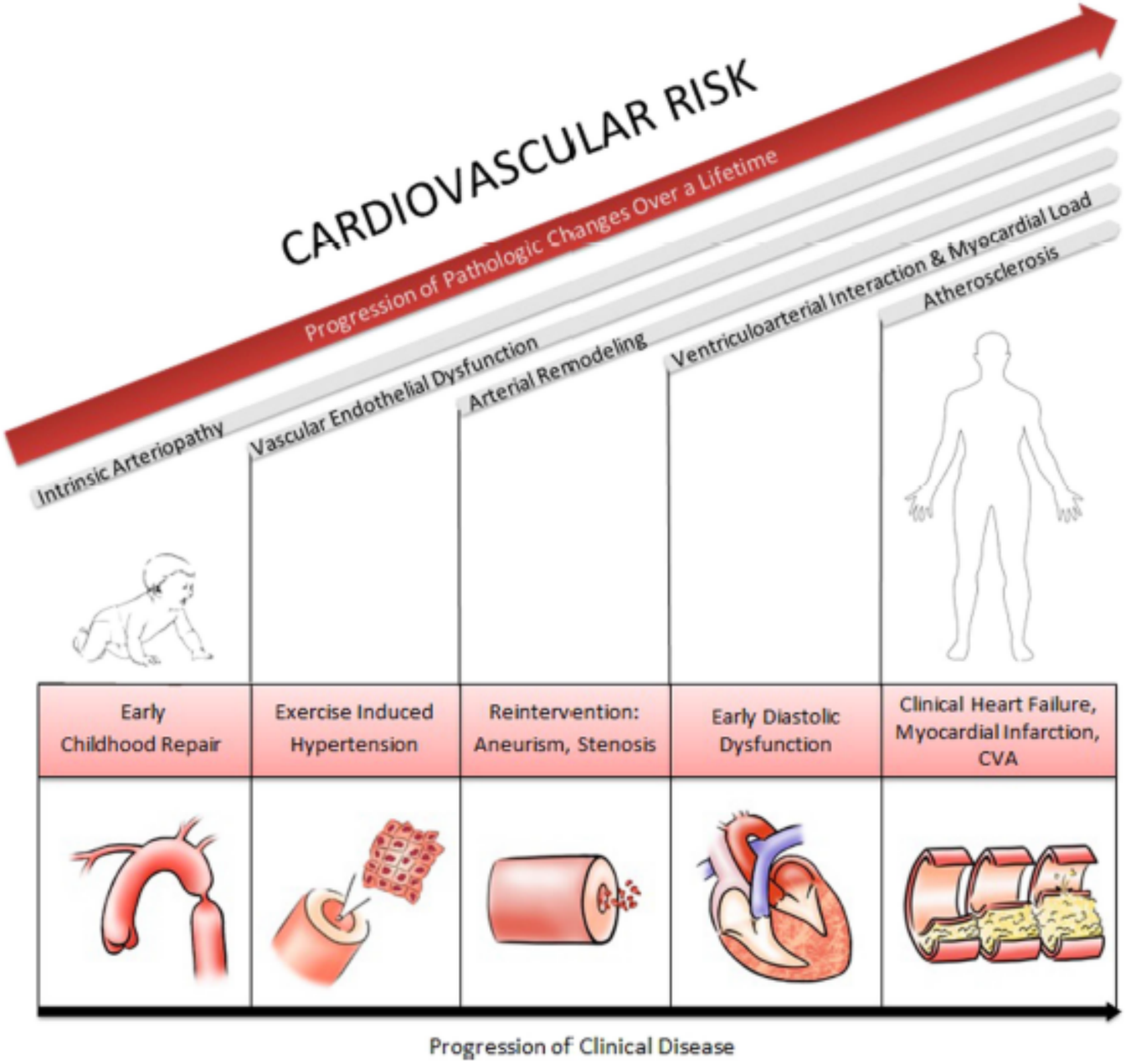
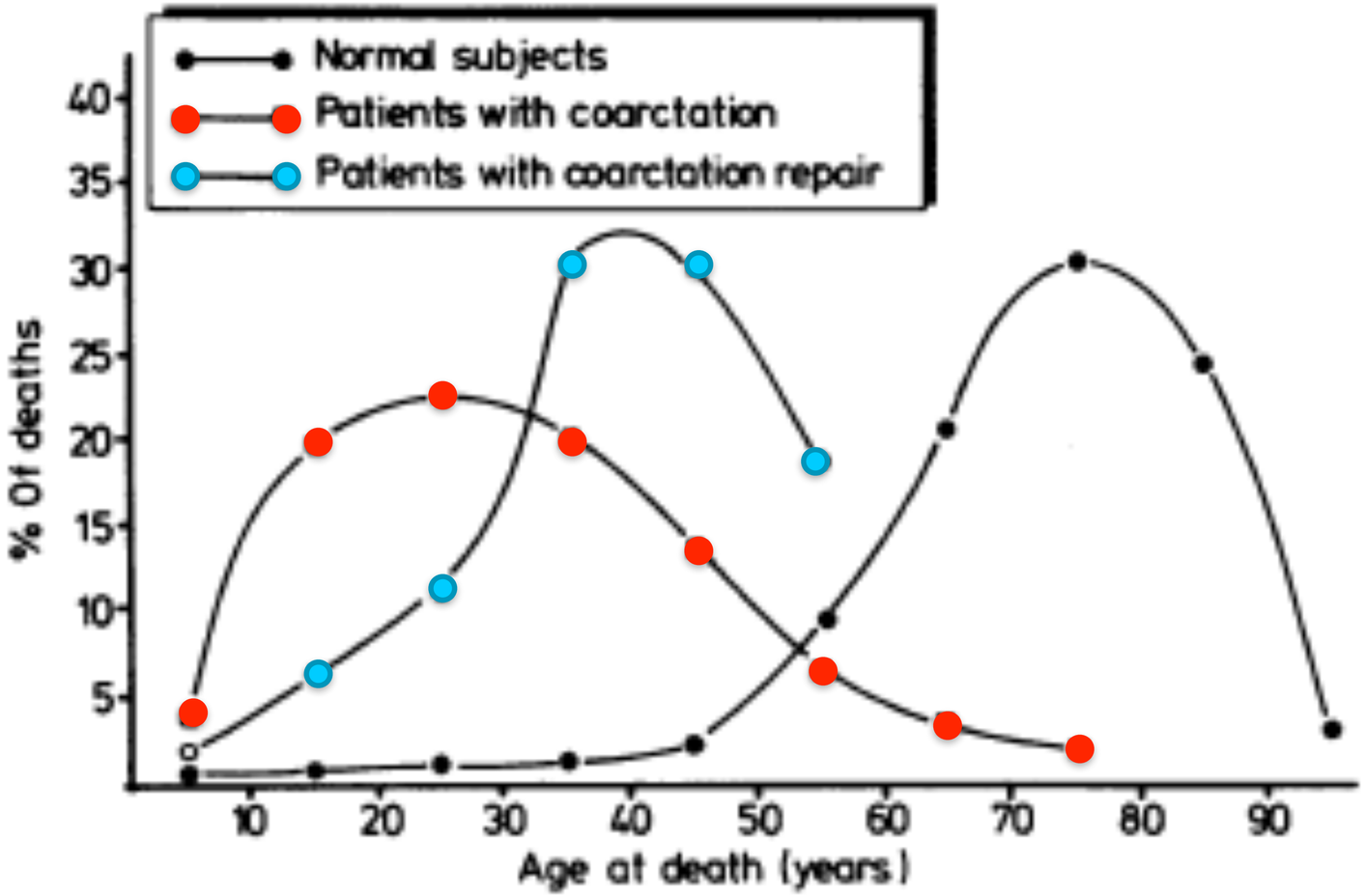
Maladies Cardiaques Héréditaires- CARDIOGEN



Bhatt AB et al. Heart 2015;101:1173-5

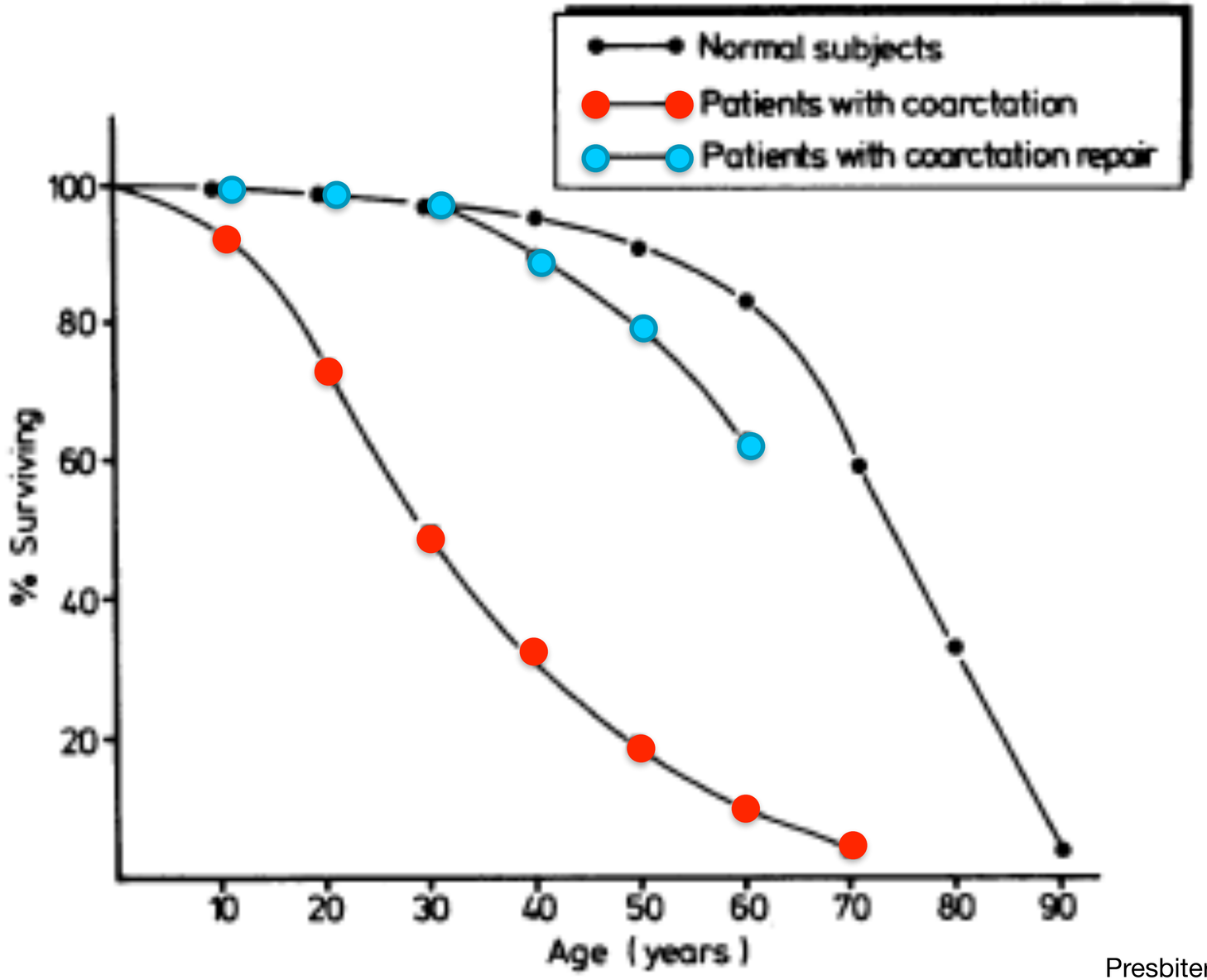
Distribution of death by age of subjects with **coarctation**, of subjects with **coarctation repair**, and of the **general population**



The curves on the left and right are taken from Campbell.
The middle curve shows the distribution of death in our patients with coarctation repair. These curves have been adjusted for age.

Presbitero P et al. Br Heart 1987;57:462-7

Percentage of subjects still alive at the end of each decade



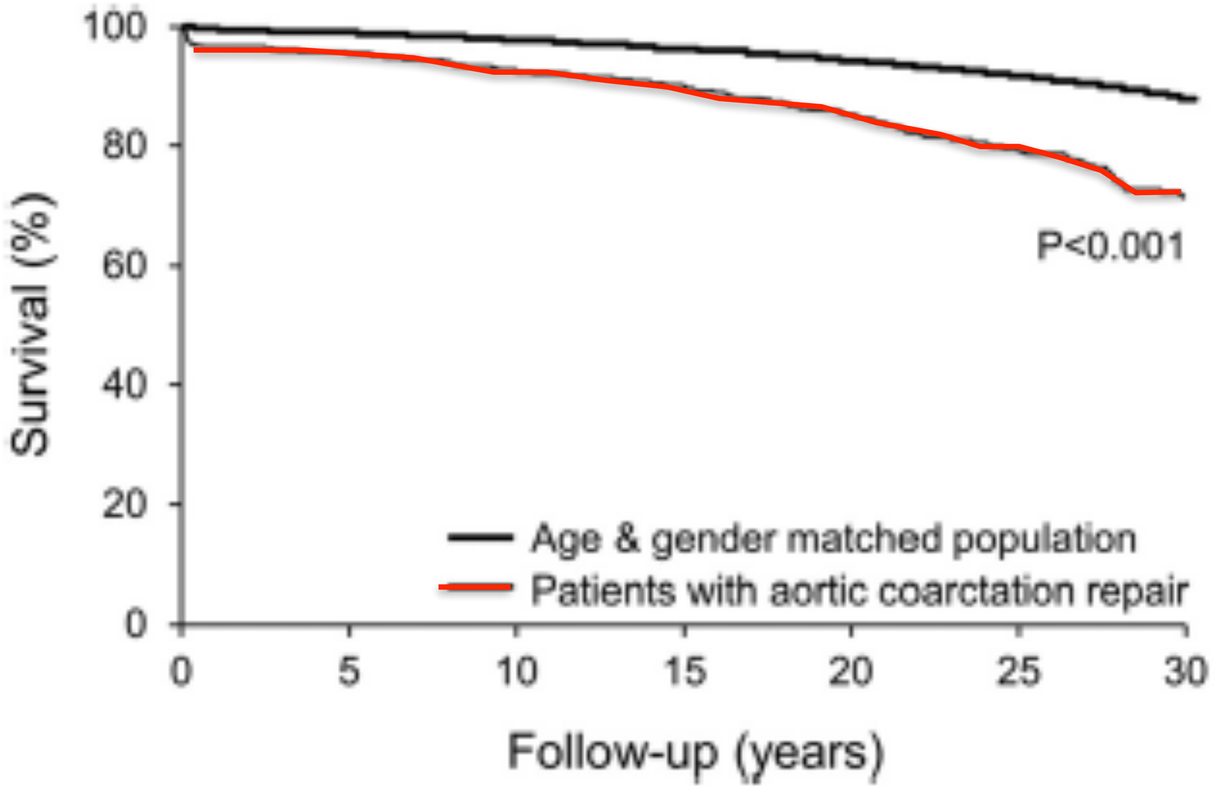
Presbitero P et al. Br Heart 1987;57:462-7

Overall survival after coarctation repair

Mean age at surgery 17.2 years

Table 1 Pre-Operative Characteristics (N = 819)

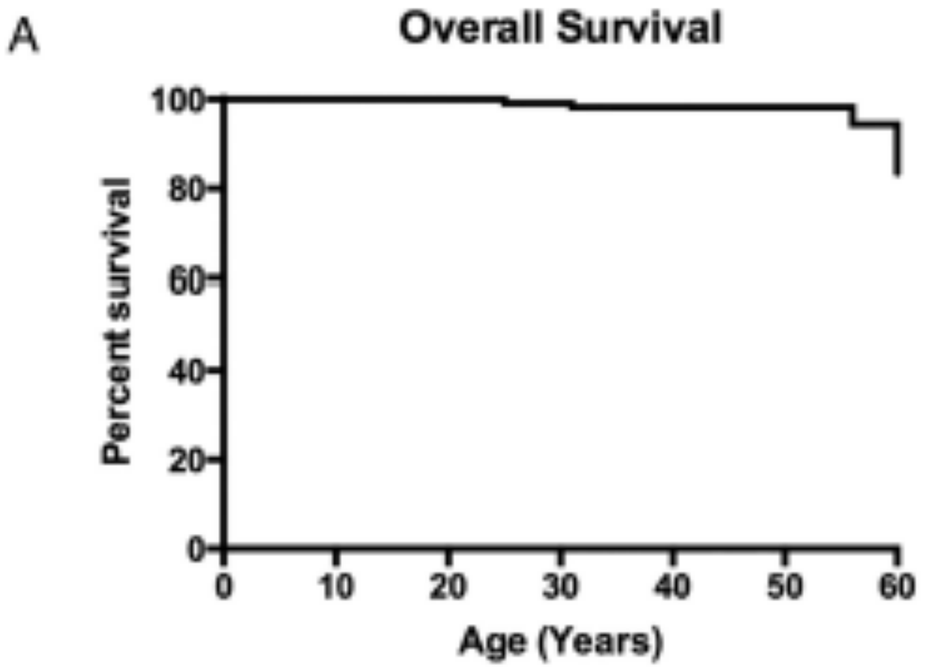
Characteristic	Value
Age at operation	
Mean ± SD, yrs	17.2 ± 13.6
Range	1 day to 72.2 yrs
Age group	
≤1 yr	116
>1–≤5 yrs	76
>5–≤10 yrs	123
>10–≤20 yrs	235
>20 yrs	269
Female	243 (30)
Pre-operation hypertension	683 (83)
NYHA class III or IV	32 (5)



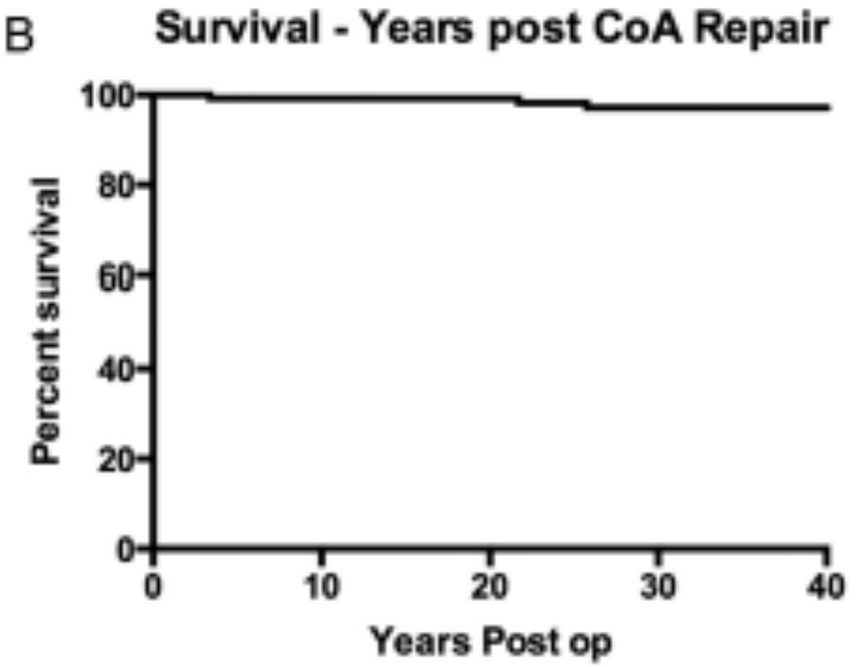
Brown ML et al. JACC 2013;62:1020-5

Kaplan–Meier curve showing long-term survival in patients with coarctation of the aorta (CoA) repair

Median age at surgery 60 months



Age (Years)	30	40	50	60
Patients at Risk	114	66	36	20
Cumulative Deaths	1	2	2	4



Years Post Repair	10	20	30	40
Patients at Risk	120	105	67	25
Cumulative Deaths	1	1	2	3

Choudhary P et al. Heart 2015;101:1190-95

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.

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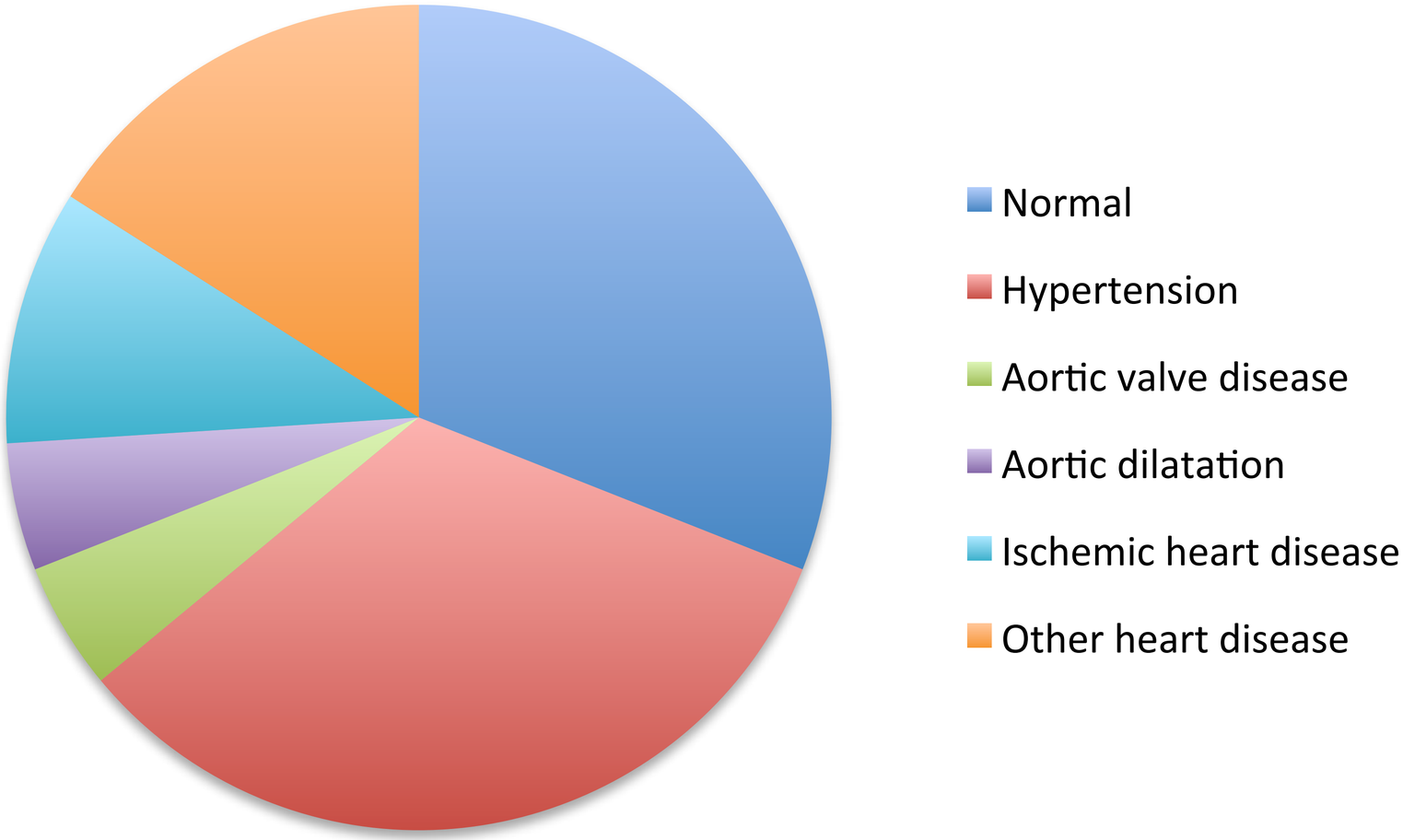
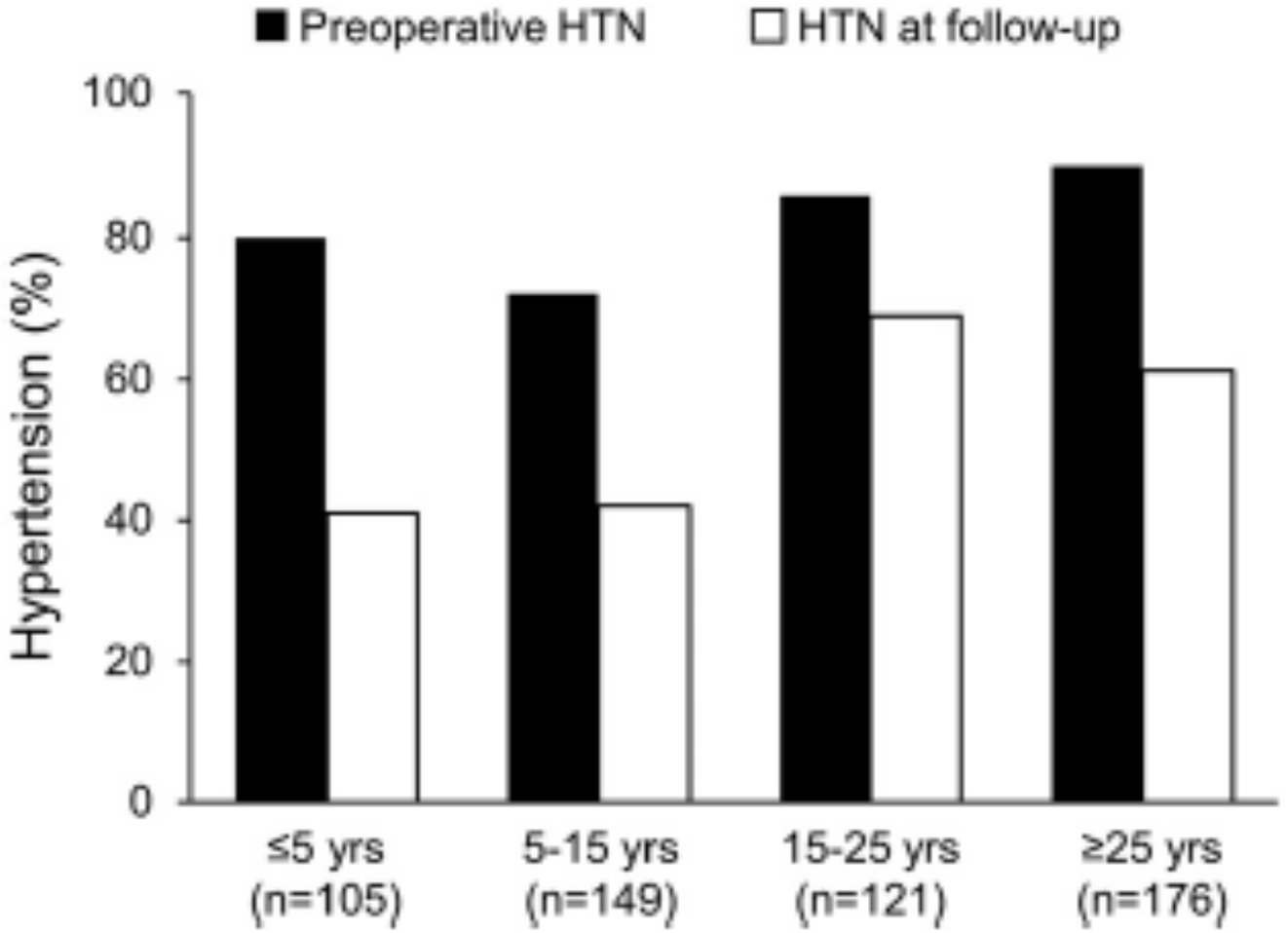
Cardiovascular morbidity in adults after coarctation repair



Stewart AB et al. Br Heart 1993;69:65-70

Comparison of hypertension pre- and postoperatively at various time intervals



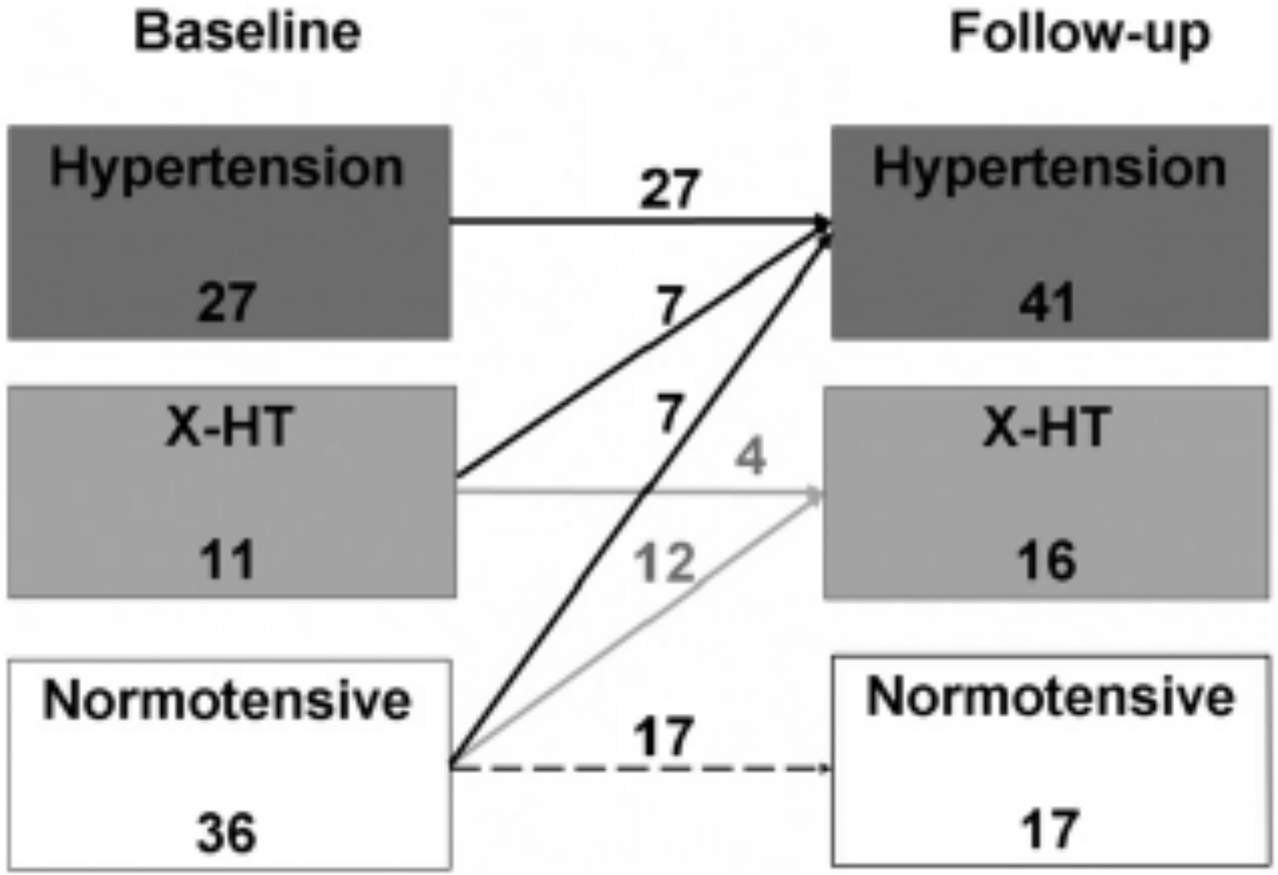
Brown ML et al. JACC 2013;62:1020-5

What type of HT ?

- Exercise HT
- 24h Ambulatory Blood Pressure Monitoring
  - 60% abnormal pressure profile
  - 24% night HT in daytime normotensive patients
- Systolic HT with increased pulse pressure
- Resting HT

Lee MGY et al. J Thorac Cardiovasc Surg 2012;144:1110-8

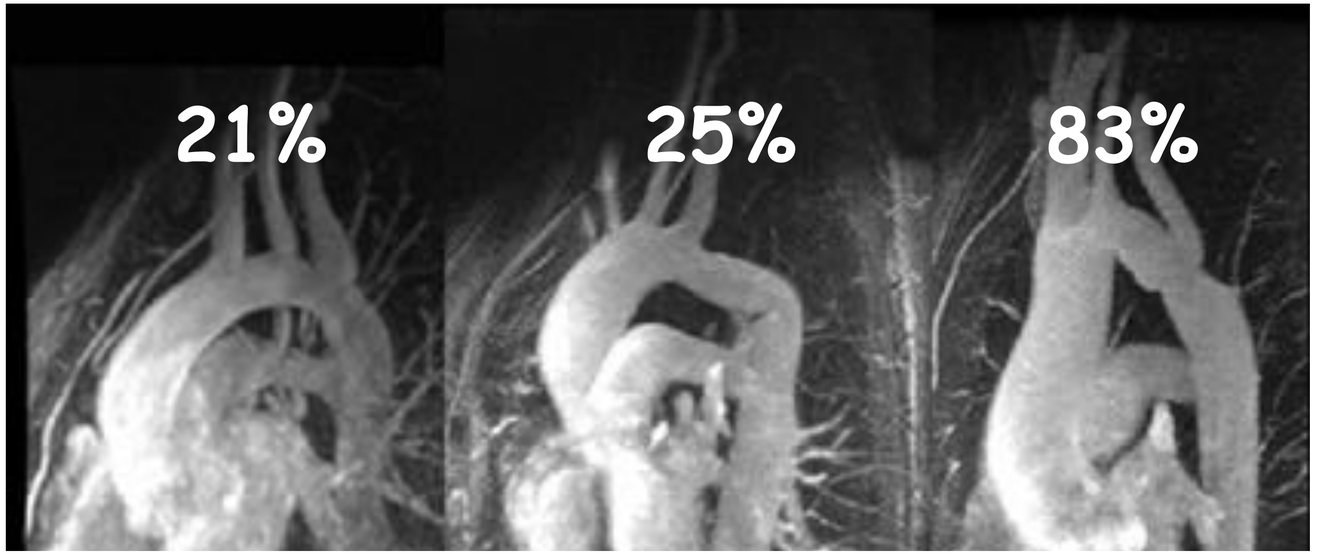
Exercise hypertension predicts late resting hypertension after coarctation repair



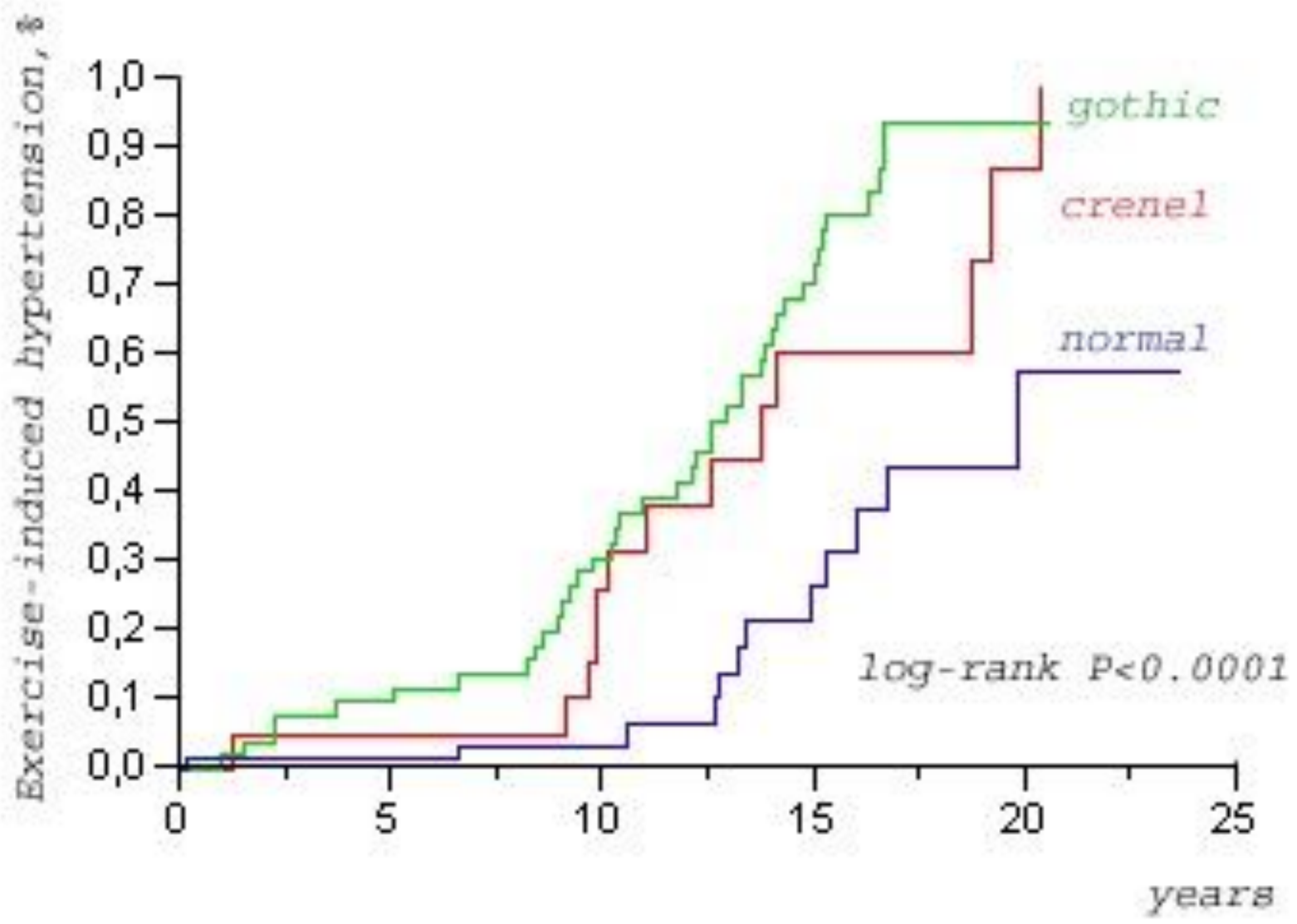
Baseline maximum exercise systolic BP was independently associated with the mean systolic BP at follow-up (p <0.005)

Luijendijk P et al. Am J Cardiol 2011;108:435-9

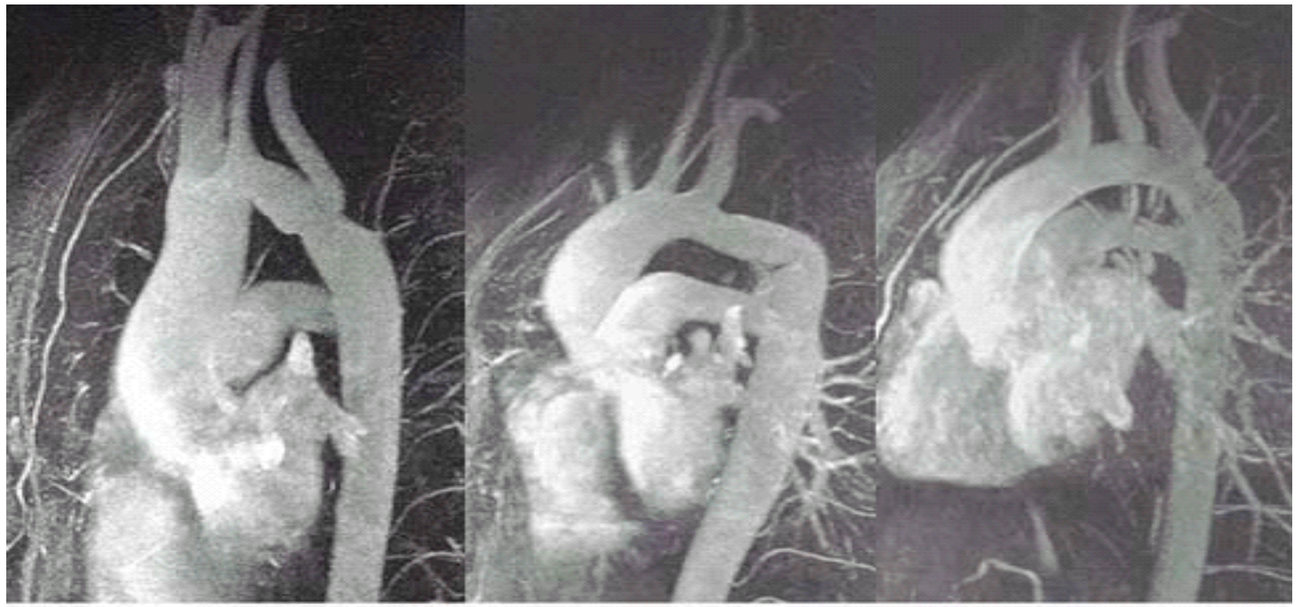
Correlation between aortic arch geometry and BP at exercise

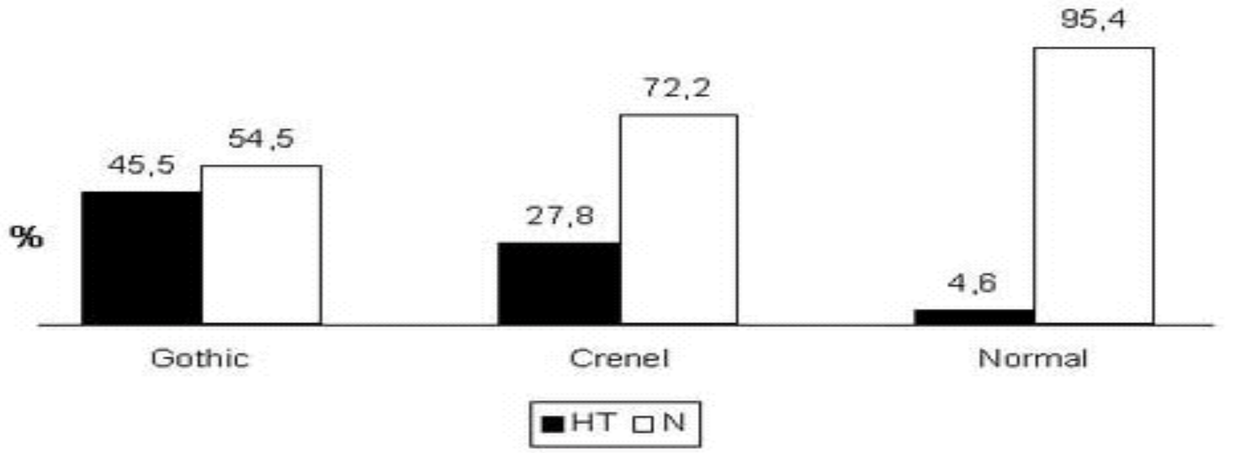


Exercise induced HT was correlated to the gothic arch form and to A/T

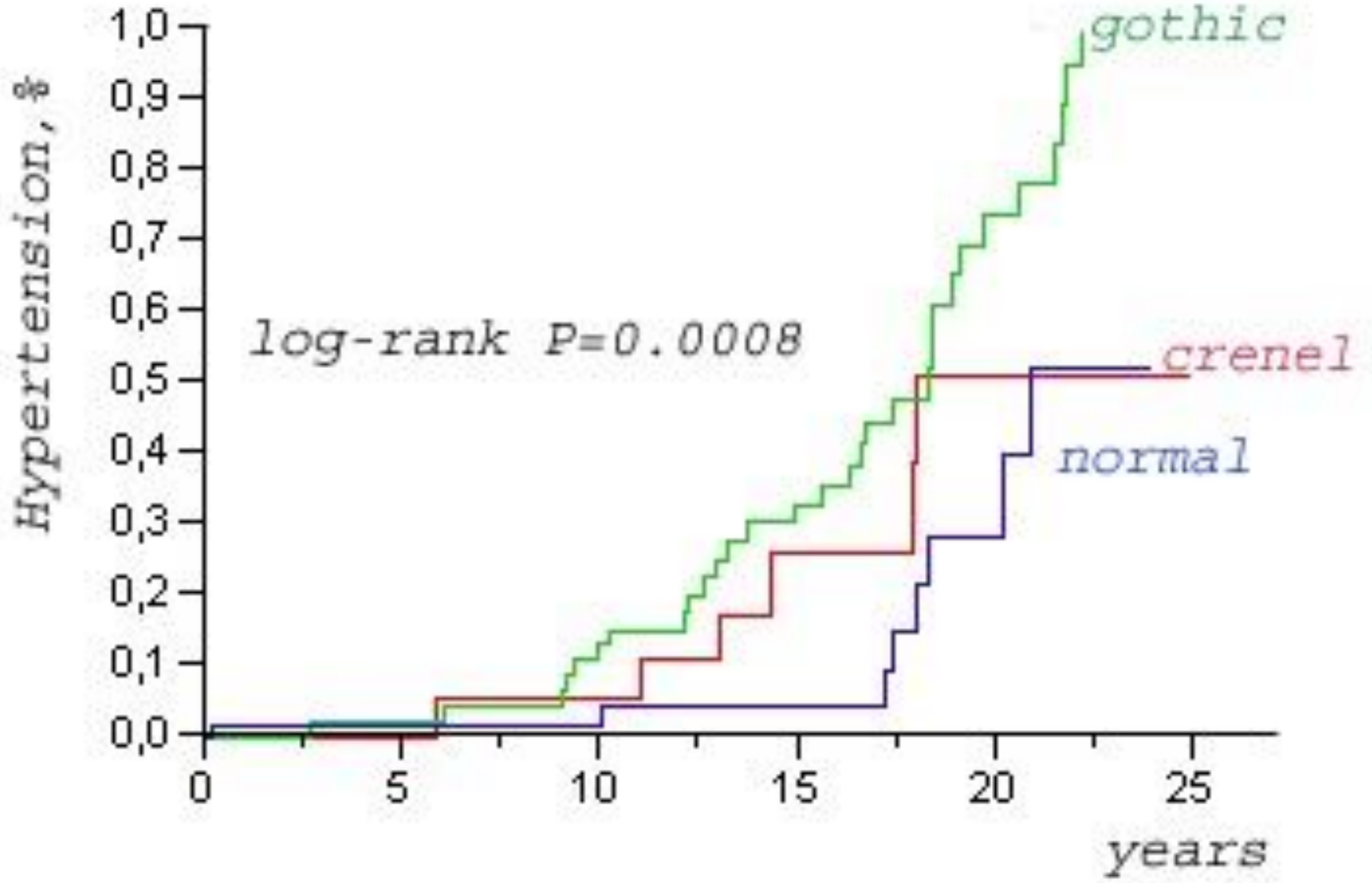

Exercise-induced hypertension, %
1,0
0,9
0,8
0,7
0,6
0,5
0,4
0,3
0,2
0,1
0,0
0
5
10
15
20
25
years
gothic
crenel
normal
log-rank P<0.0001

Aortic arch geometry and resting HT after CoA repair





Ou P. et al, Eur. Heart J. 2004


Hypertension, %
1,0
0,9
0,8
0,7
0,6
0,5
0,4
0,3
0,2
0,1
0,0
0
5
10
15
20
25
years
log-rank P=0.0008
gothic
crenel
normal

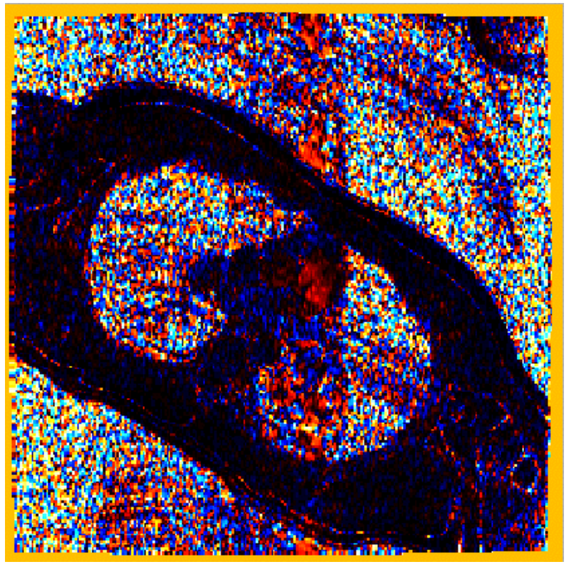
Aortic arch geometry and flow dynamics after CoA repair

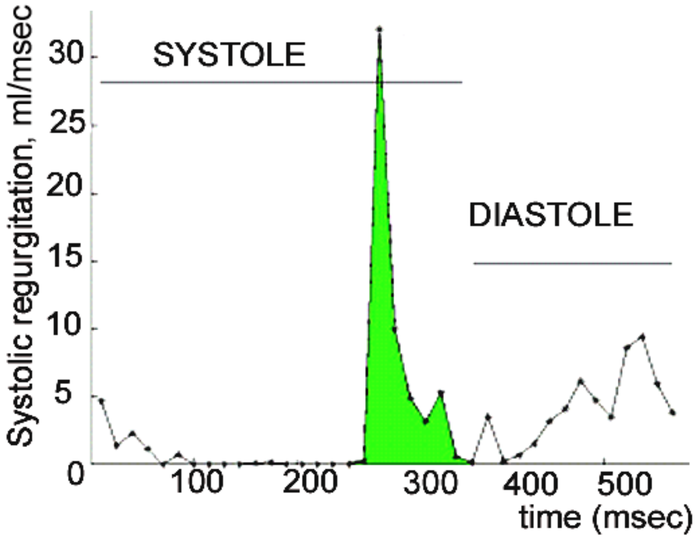
	GOTHIC	CRENEL	NORMAL	*P value*
PWV, (m/sec)	8.6 ±3.4	4.5 ±1.6	3.7 ±0.5	*<0.0001*
Systolic regurgitation (ml/msec)	14.4 ±5.3	7.9 ±1.3	5.3 ±1.9	*<0.001*

Aortic arch geometry and flow dynamics after CoA repair

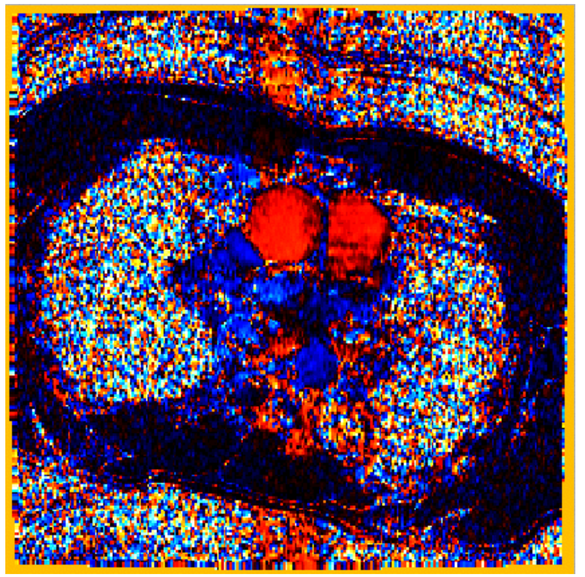
Flow dynamics in the ascending aorta

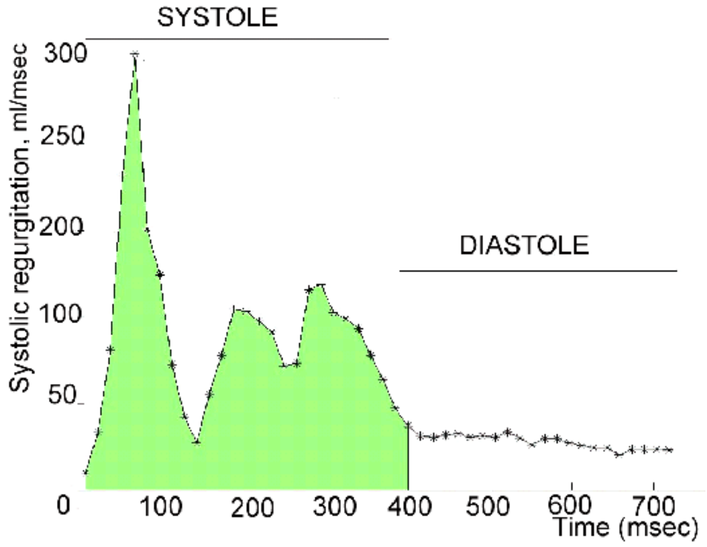
NORMAL

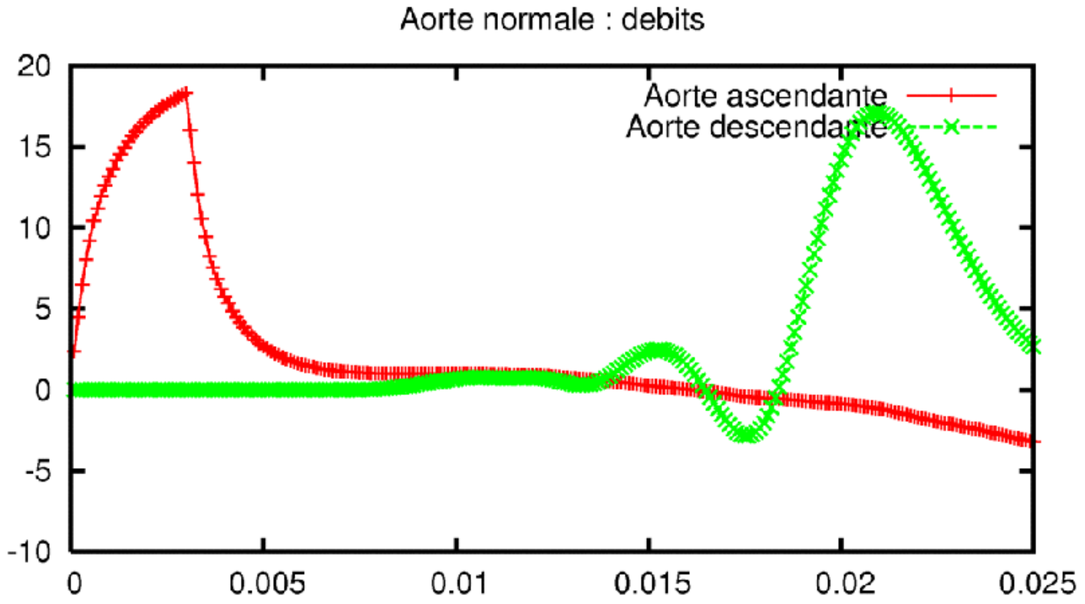


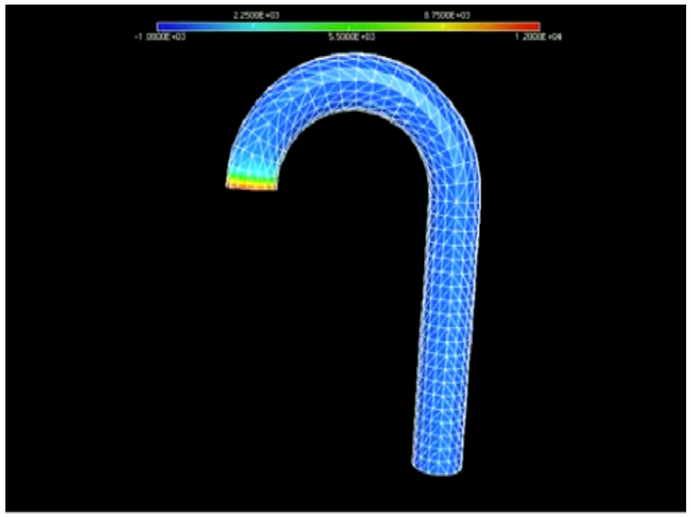


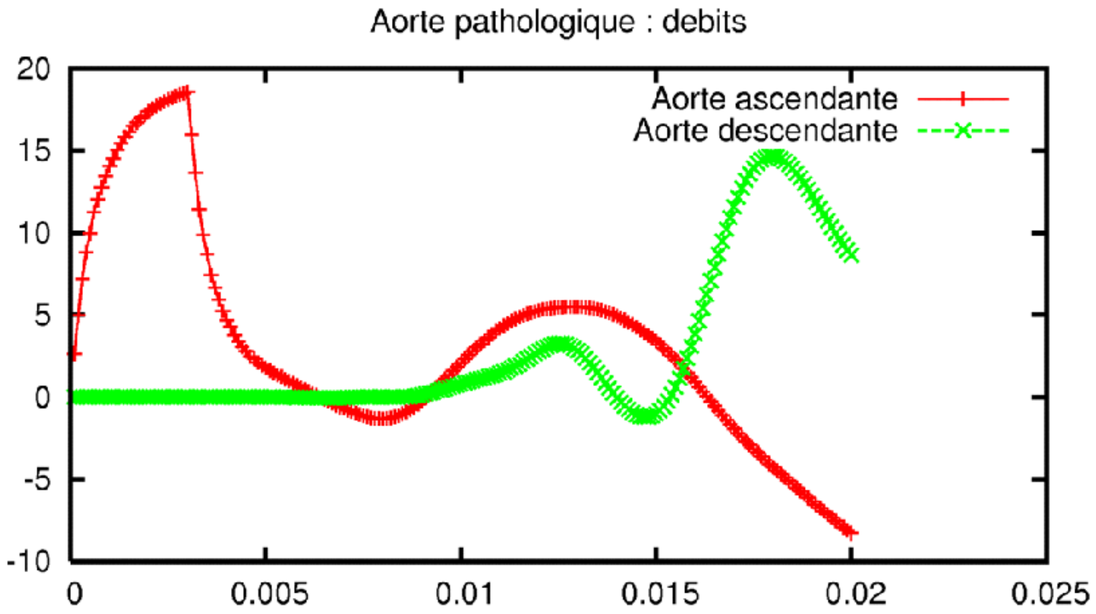
GOTHIC

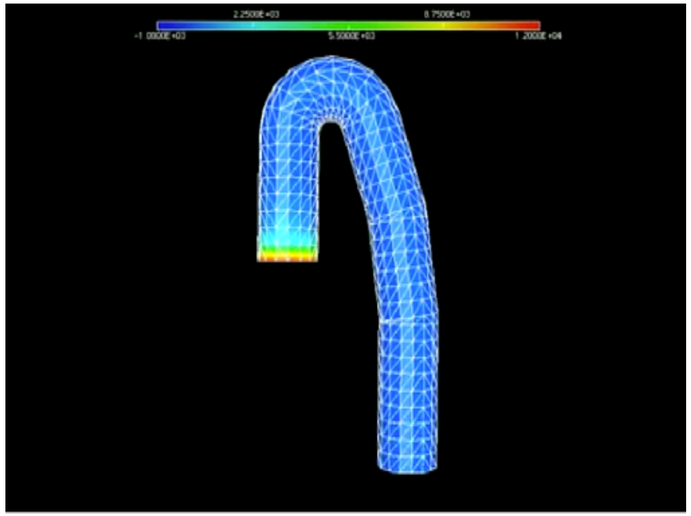




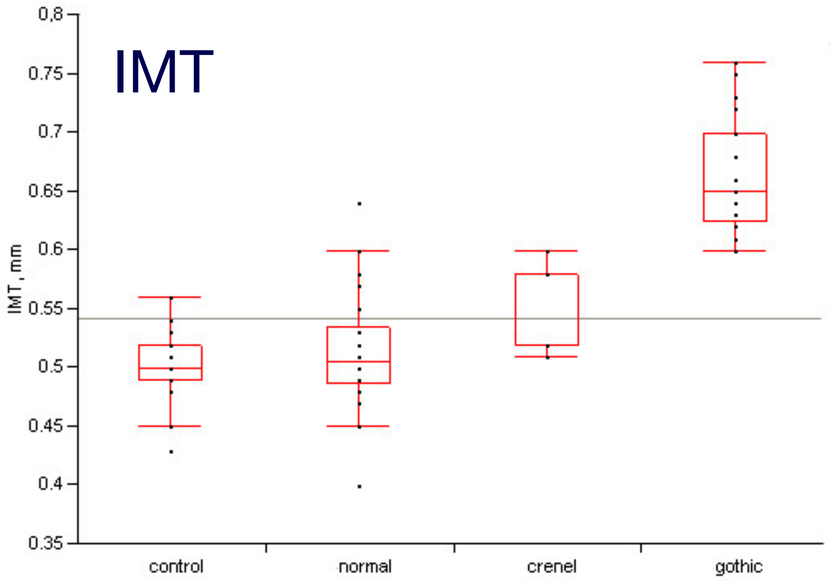


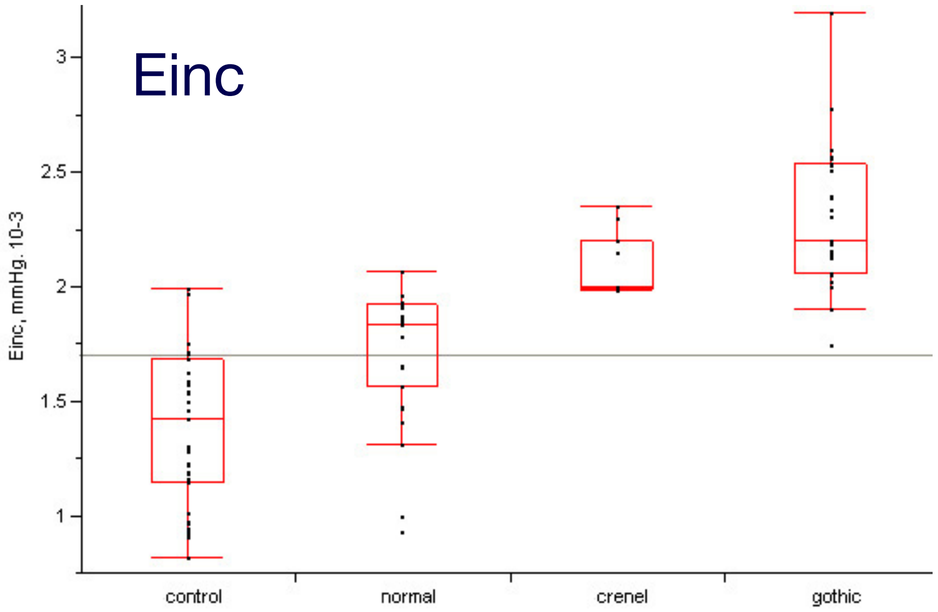


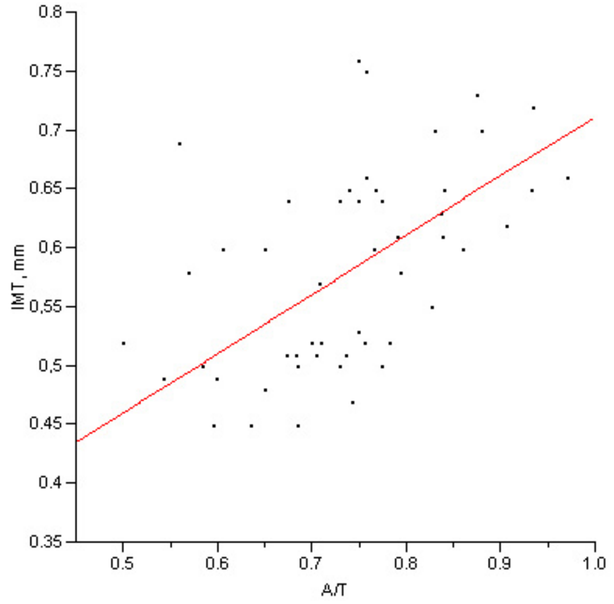


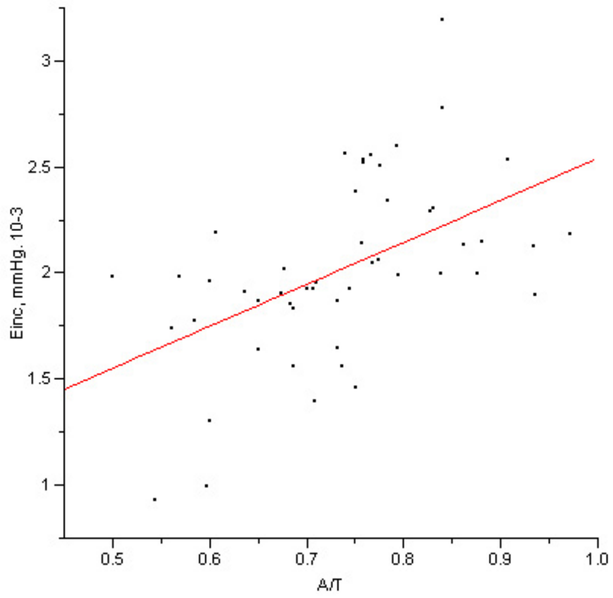


Szopos M et al. J Thorac Cardiovasc Surg. 2014;148:1572-82.









Ou P et al. J Am Coll Cardiol. 2007;49:883-90.
Ou P et al. J Thorac Cardiovasc Surg. 2008;135:62-8.

The significance of isolated, exercise-induced hypertension is a matter of debate.

- Exercise HT does not help to detect recoarctation.
- Exercise HT predicts resting HT.
- Exercise HT is associated with vascular remodeling and increased LV mass.
- Should exercise HT lead to early antihypertensive treatment ?

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 roel in the development of hypertension. The significance of isolated, exercise-induced hypertension is a matter of debate.

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- Attention is required for BAV, mitral valve disease, premature CAD, and berry aneurysm of the circle of Willis.

When to treat ?
How to treat residual or re-coarctation ?

Table 11 Indications for intervention in coarctation of the aorta

Indications	Class[a]	Level[b]
All patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention	I	C
Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention	IIa	C
Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention	IIb	C

[a]Class of recommendation.
[b]Level of evidence.
CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

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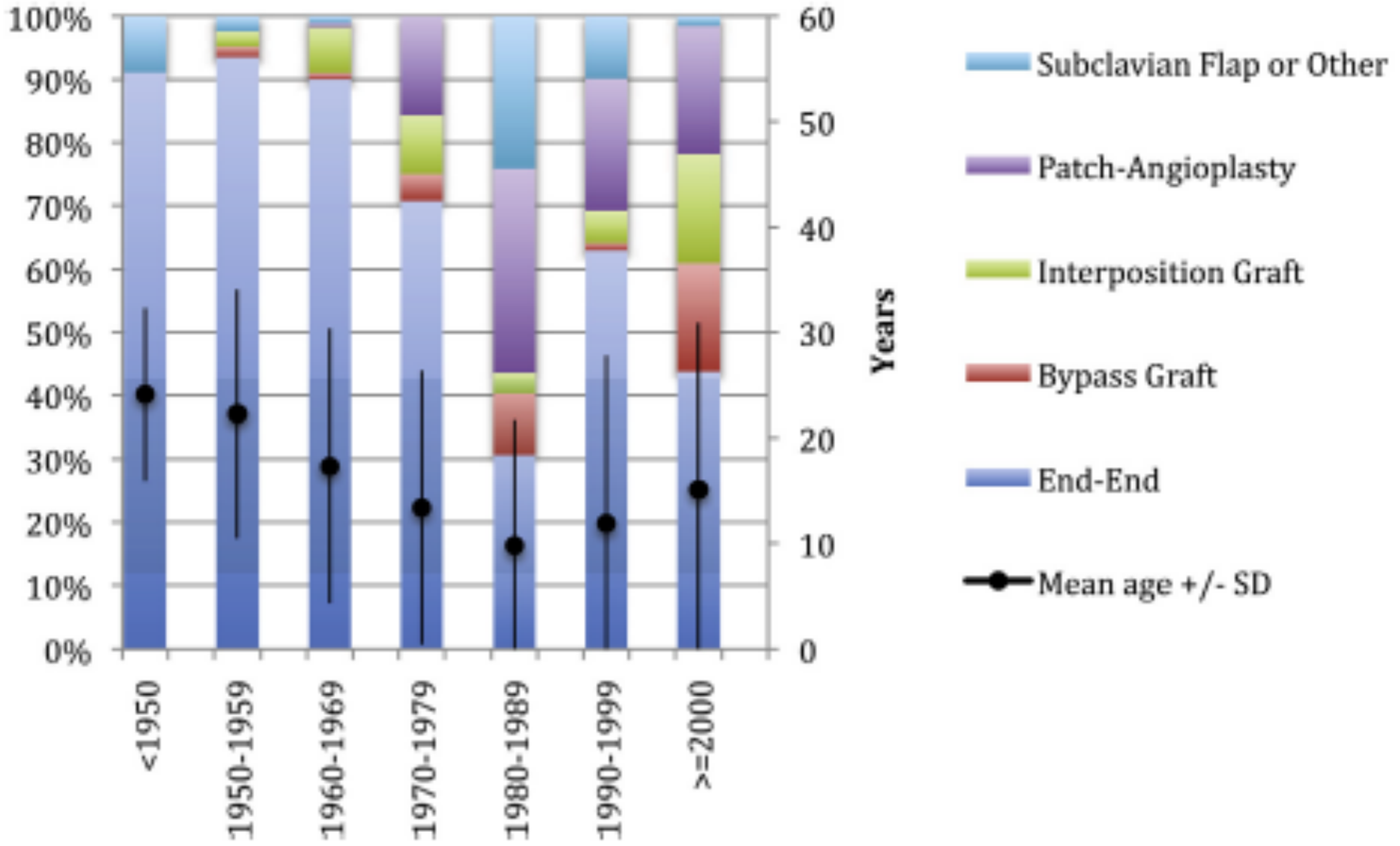
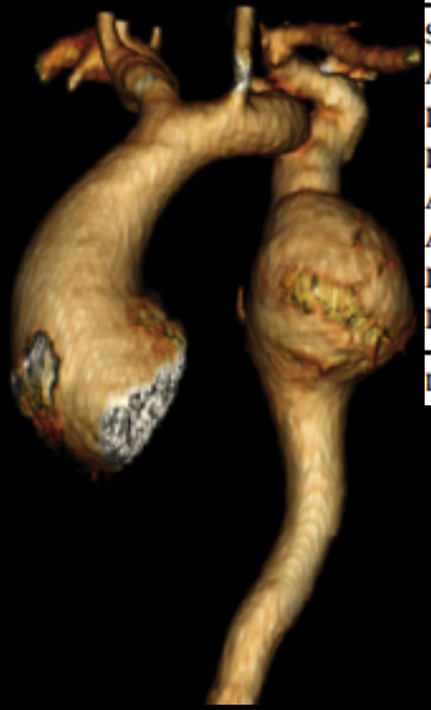
Type of repair by decade



Brown ML et al. JACC 2013;62:1020-5

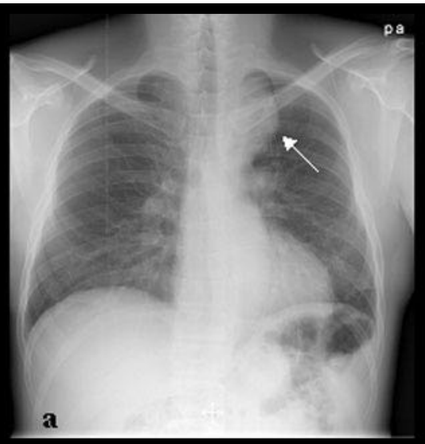
Aortic coarctation endocarditis

Aneurysm at the repair site



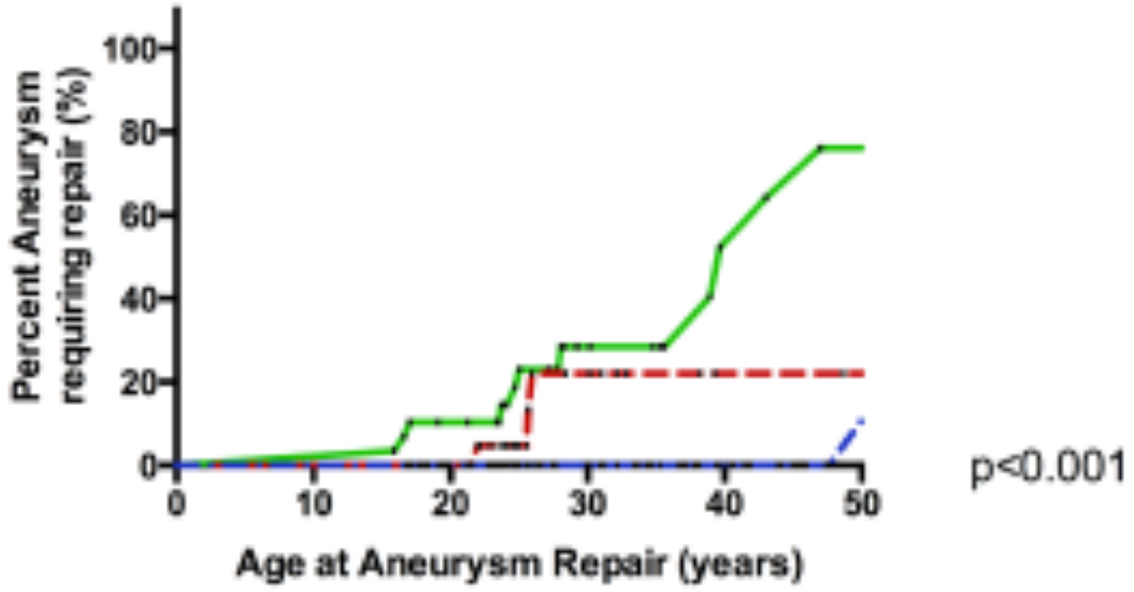
Location	Number (%)
Site of prior repair or DTA	42 (76.4%)
Ascending aorta	18 (32.7%)
Left subclavian artery	8 (14.5%)
Distal aortic arch	8 (14.5%)
Aortic root	6 (10.9%)
Abdominal aorta	1 (1.8%)
Iliac artery	1 (1.8%)
Innominate artery	1 (1.8%)

DTA = descending thoracic aorta.



Preventza O et al. Ann Thorac Surg 2013;95:1960-7

Incidence of descending aortic aneurysms requiring repair



p<0.001

Patients at risk	Total	Legend	20 years	30 years	40 years	50 years
End-to-end repair	43		36	21	13	7
Subclavian flap aortoplasty	28		25	7	3	2
Patch Aortoplasty	31		25	9	5	3

Choudhary P et al. Heart 2015;101:1190-95

Aneurysm of the ascending aorta

32% of patients with aortic root dilatation
21% with aneurysm of ascending aorta
74% had Bicuspid aortic valve

Padang R et al. Heart, Lung and Circulation 2014;23: 332–338

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Common cardiovascular anomalies associated with coarctation

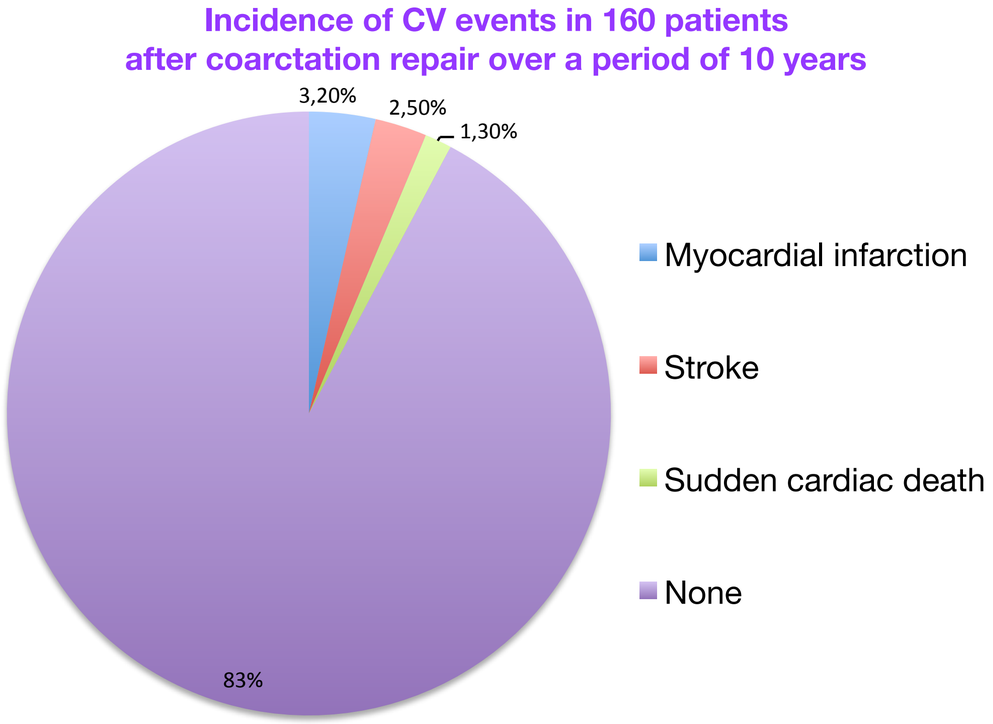
Anomaly	No. of patients	% ($n = 500$ unless otherwise specified)
Bicuspid aortic valve (including functional bicuspid valve)	268	59.6 ($n = 449$)
Arch hypoplasia	71	14.2
Ventricular septal defect	64	12.8
Mitral valve abnormalities	41	8.2
Mitral regurgitation	20	
Mitral valve prolapsed	6	
Mitral stenosis	4	
Double-orifice mitral valve	3	
Parachute mitral valve	2	
Imperforate mitral valve (associated with Shone complex)	2	
Patent ductus arteriosus	36	7.2
Subaortic stenosis	28	5.6
Other arch anomalies	21	4.2
Aberrant right subclavian artery	8	
Aberrant left subclavian artery	1	
Bovine pattern	7	
Other arch anomalies	5	
Left superior vena cava	21	4.2
Atrial septal defect	10	2

High proportion of Sievers type 0 BAV

Teo LLS et al. Pediatr Cardiol 2011;32:1120-7

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.



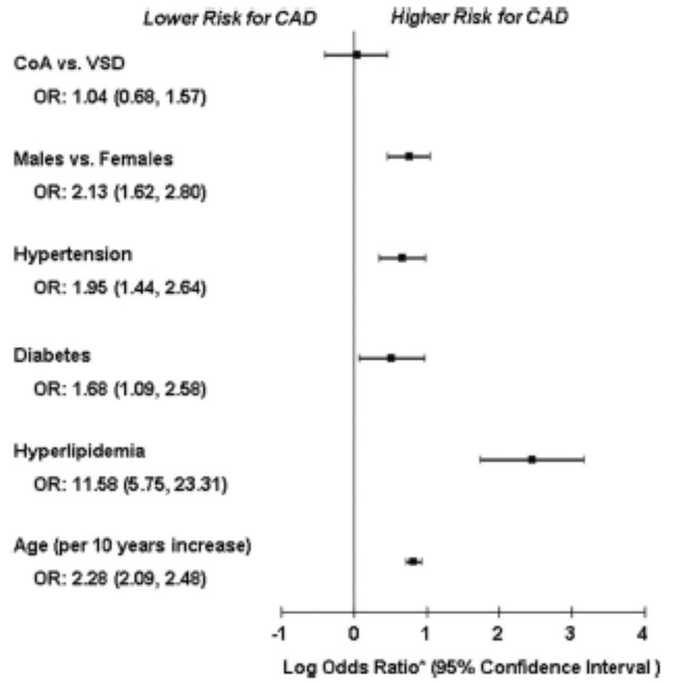


Figure 2. Multivariable analysis of the effect of aortic coarctation (CoA) vs ventricular septal defect (VSD) on coronary artery disease (CAD; nested case-control sample). OR indicates odds ratio.

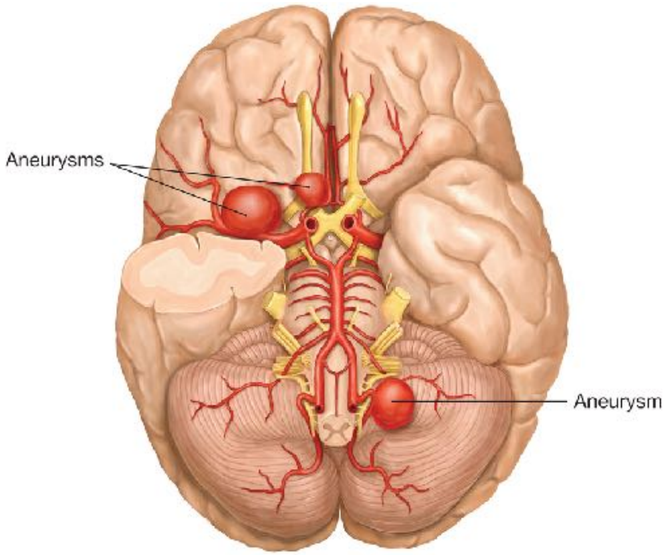
Luijendijk P et al. Int J Cardiol 2014;176:776-81
Roifman I et al. Circulation 2012;126:16-21

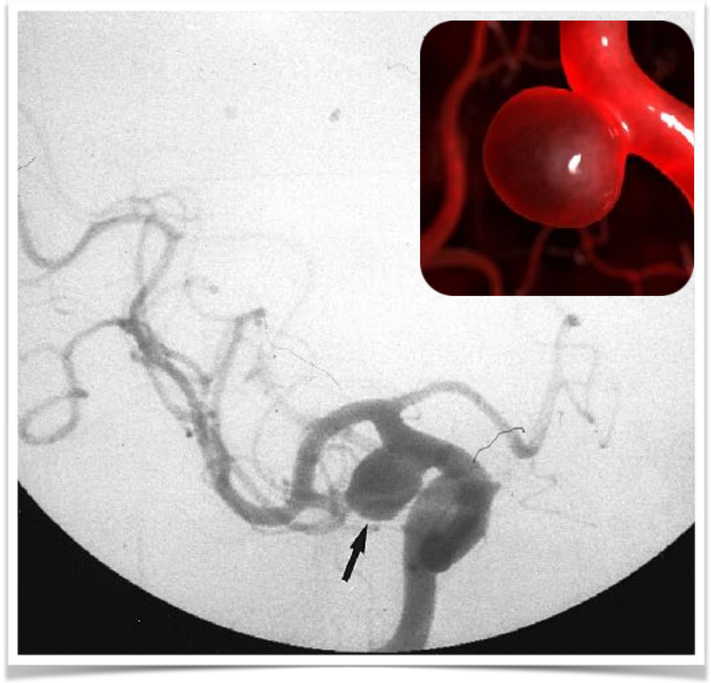
Berry aneurysms of the circle of Willis and coarctation

Controversial prevalence in patients with Coarctation

5-18% in a MRI study in adults (mean age 42 years)[1]

0% in a MRI study in adolescents (mean age 16 years) [2]





1-Connolly HM, et al. Mayo Clin Proc 2003;78:1491-9.
2-Donti A et al. Am J Cardiol 2015;116:630-3

Curtis SL et al. AJNR 2012
Cook SC et al. CHD 2013
Pickard et al. JAHA 2018
Stout et al. JACC 2018

Tailored treatment and follow-up

- Optimize aortic geometry during repair
  - to prevent/delay systemic HT
- Detect systemic hypertension
- MRI best available tool for aortic anatomy-geometry, aortic function, LV mass and function
- Intra-cardiac surveillance : echocardiography
- Prevent CV events by minimizing usual CV risk factors
- Pending questions:
  - Reinforce monitoring of patients with exercise HT ?
  - Reinforce follow-up in patients with gothic geometry, vascular dysfunction, associated BAV…
  - Screening for aortic aneurysms in patients with patch aortoplasty
  - Screening for berry aneurysms in patients operated at an older age and in hypertensive patients

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