

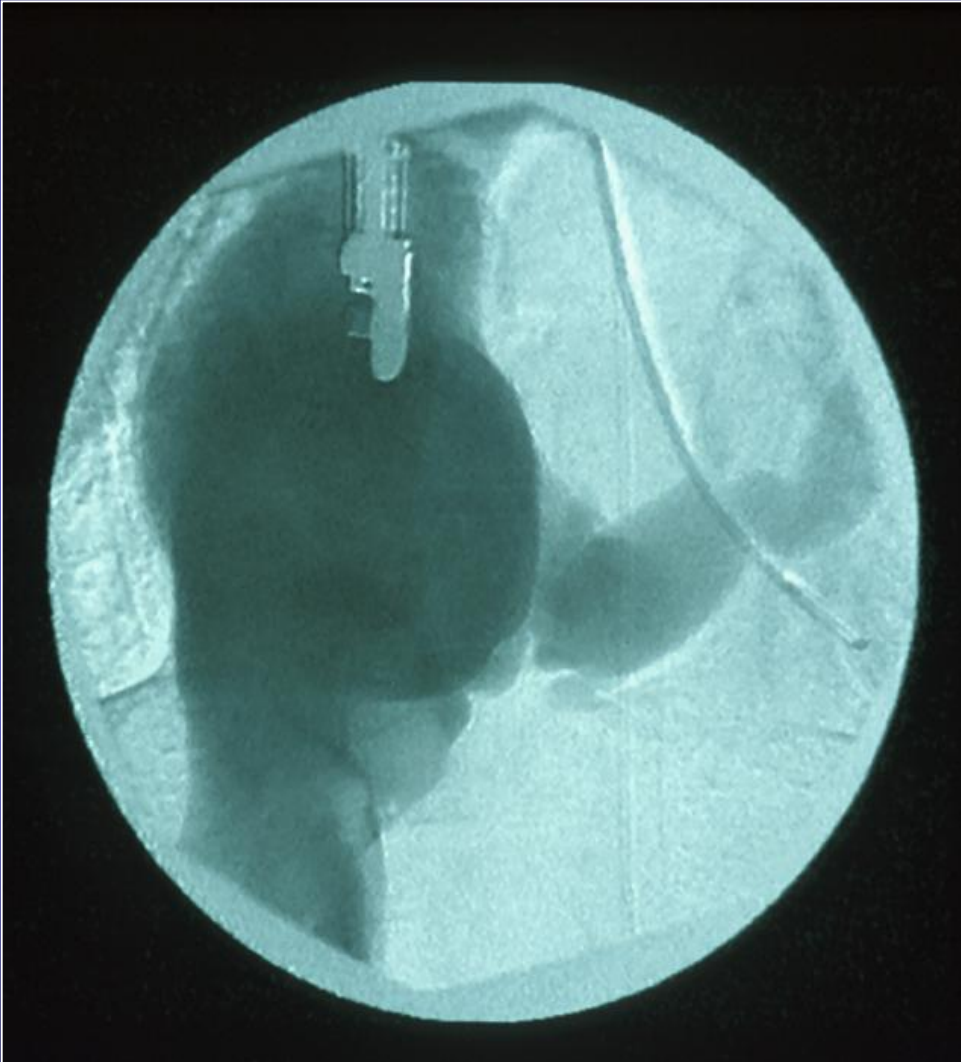


Children are not little adults



Adults are not big children

ACHD: Long Term Sequelae



Complex circulations with built in limitations

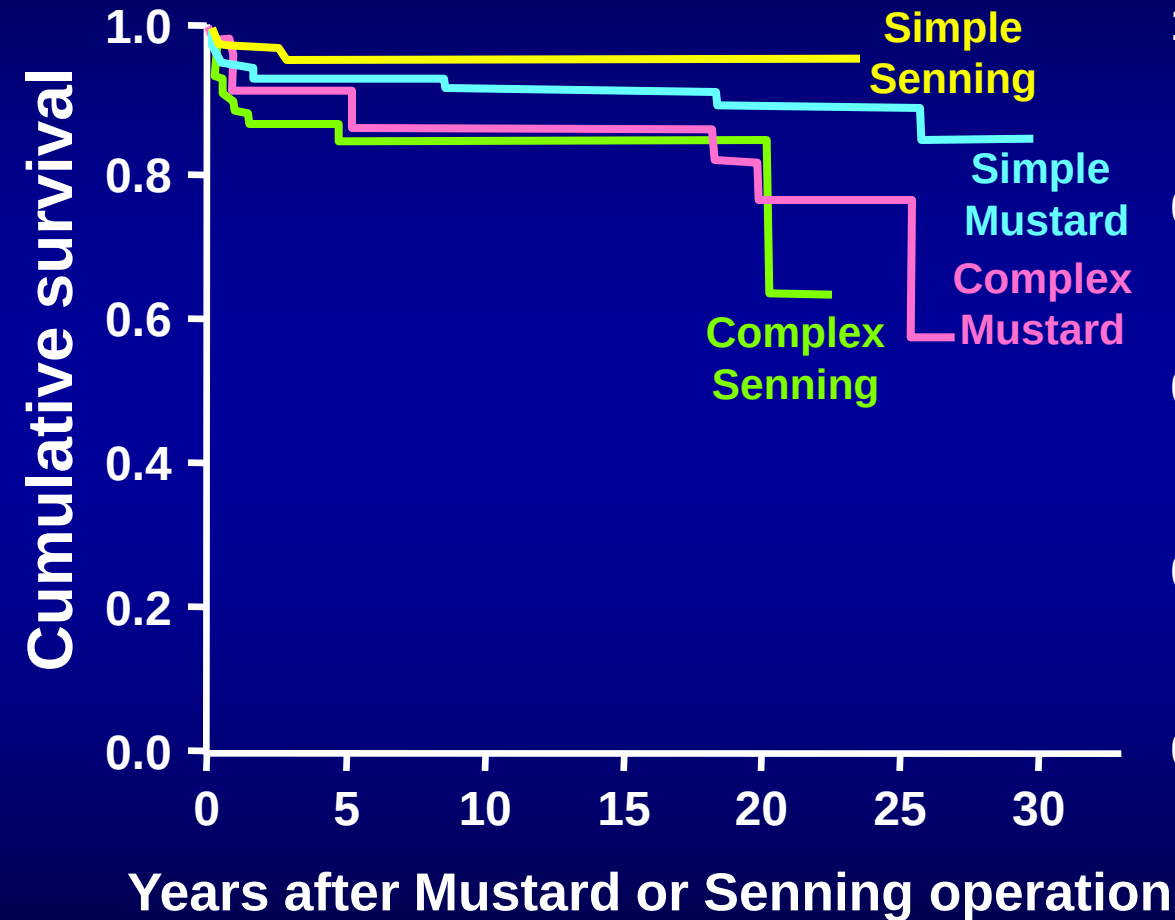
- **Development of arrhythmia and pump failure**

- **Late survival for many conditions still unknown**

- **Advances in imaging and treatments**

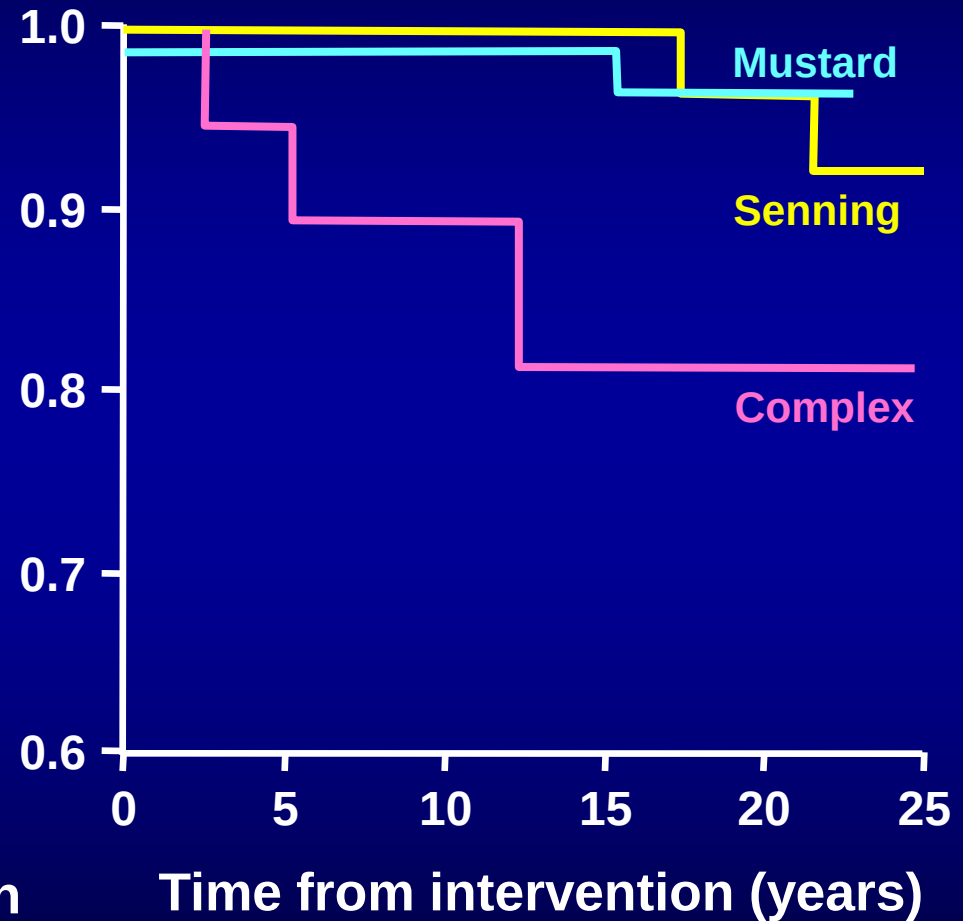
Outcome after IAR of TGA

339 pts - 124 M, 215 S



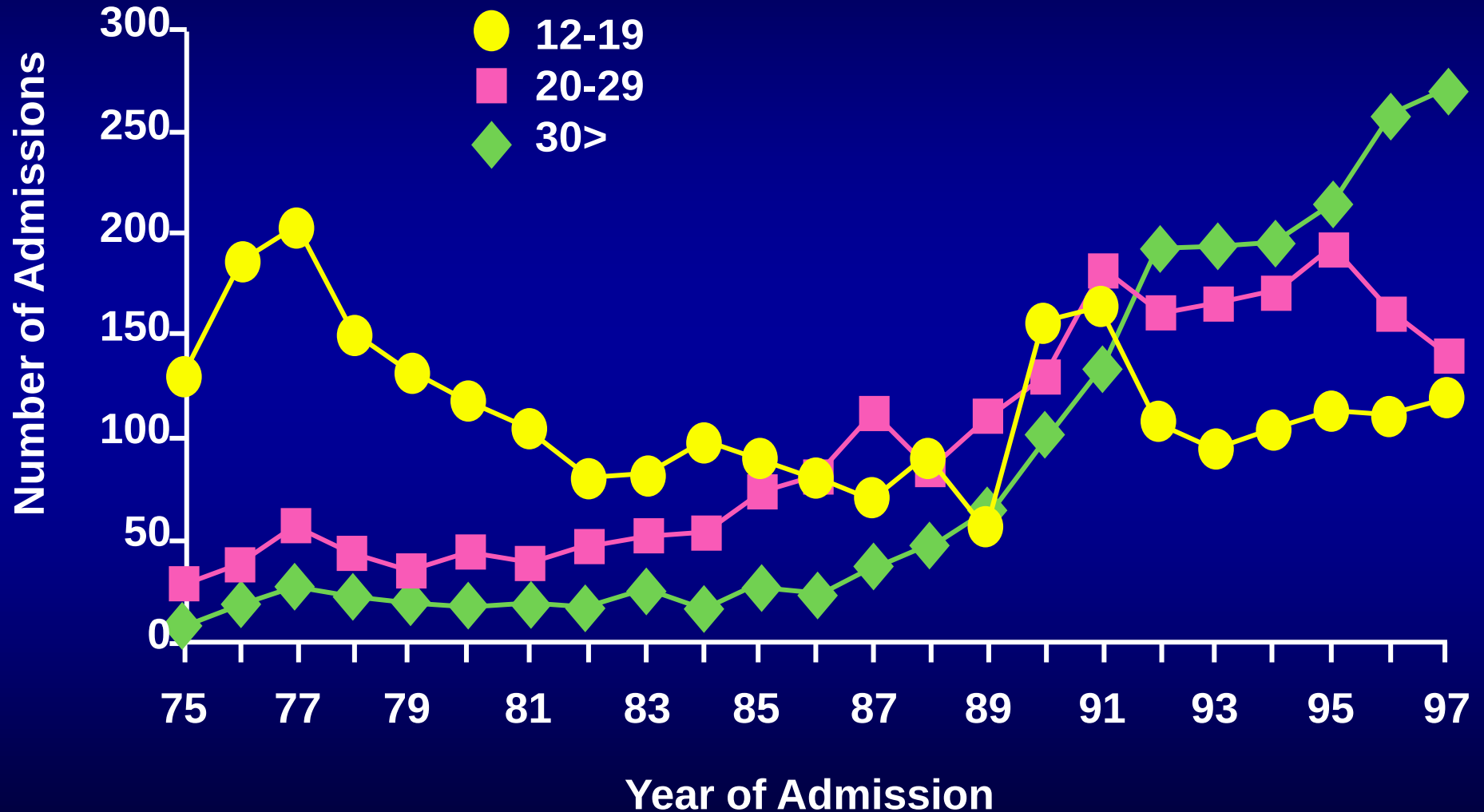
Moons Heart 2004;90:307-313

339 pts - 124 M, 215 S



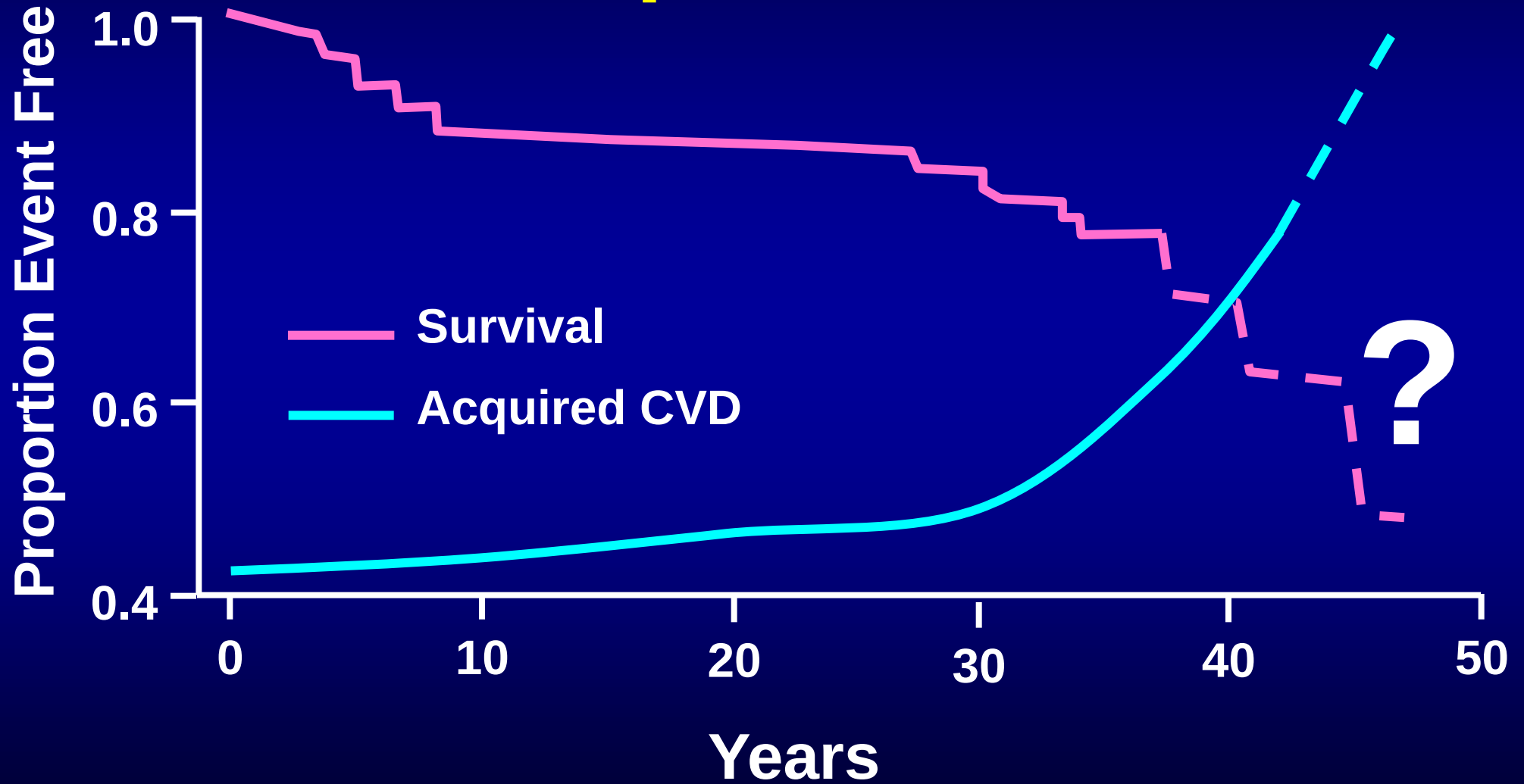
Dos Heart 2005;91:652-6565

Population is Getting Older

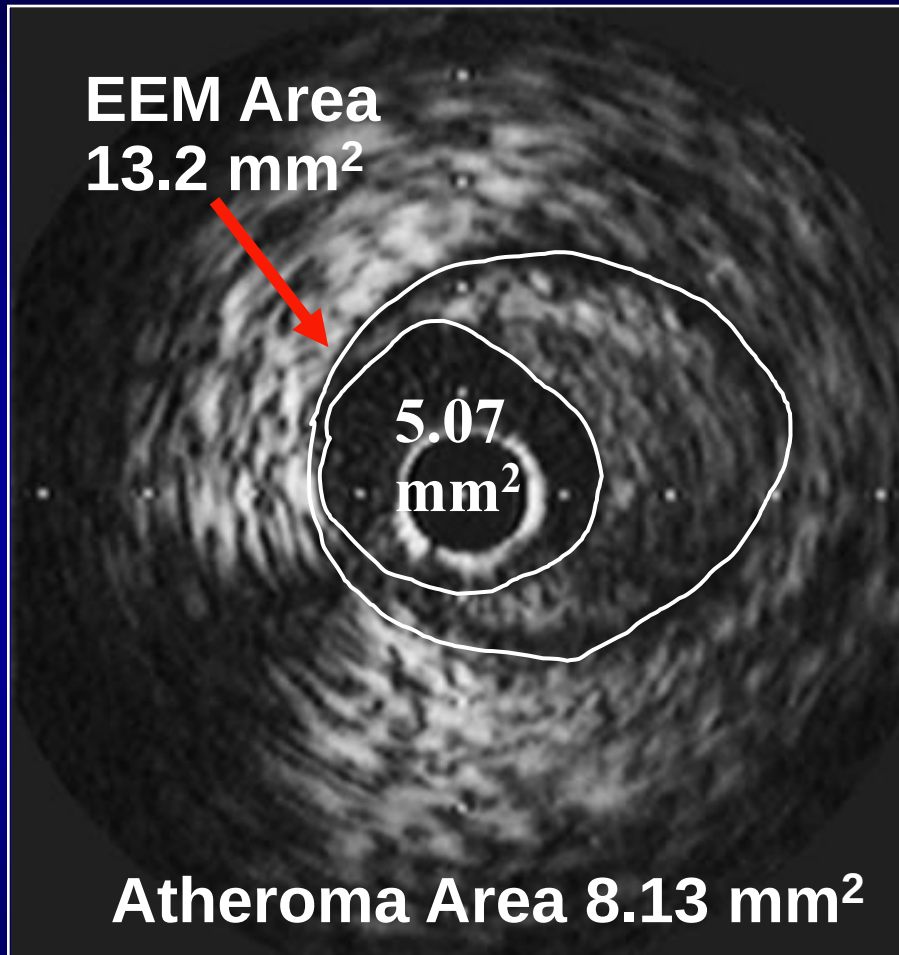


Statistics from Jane Somerville, London, UK.

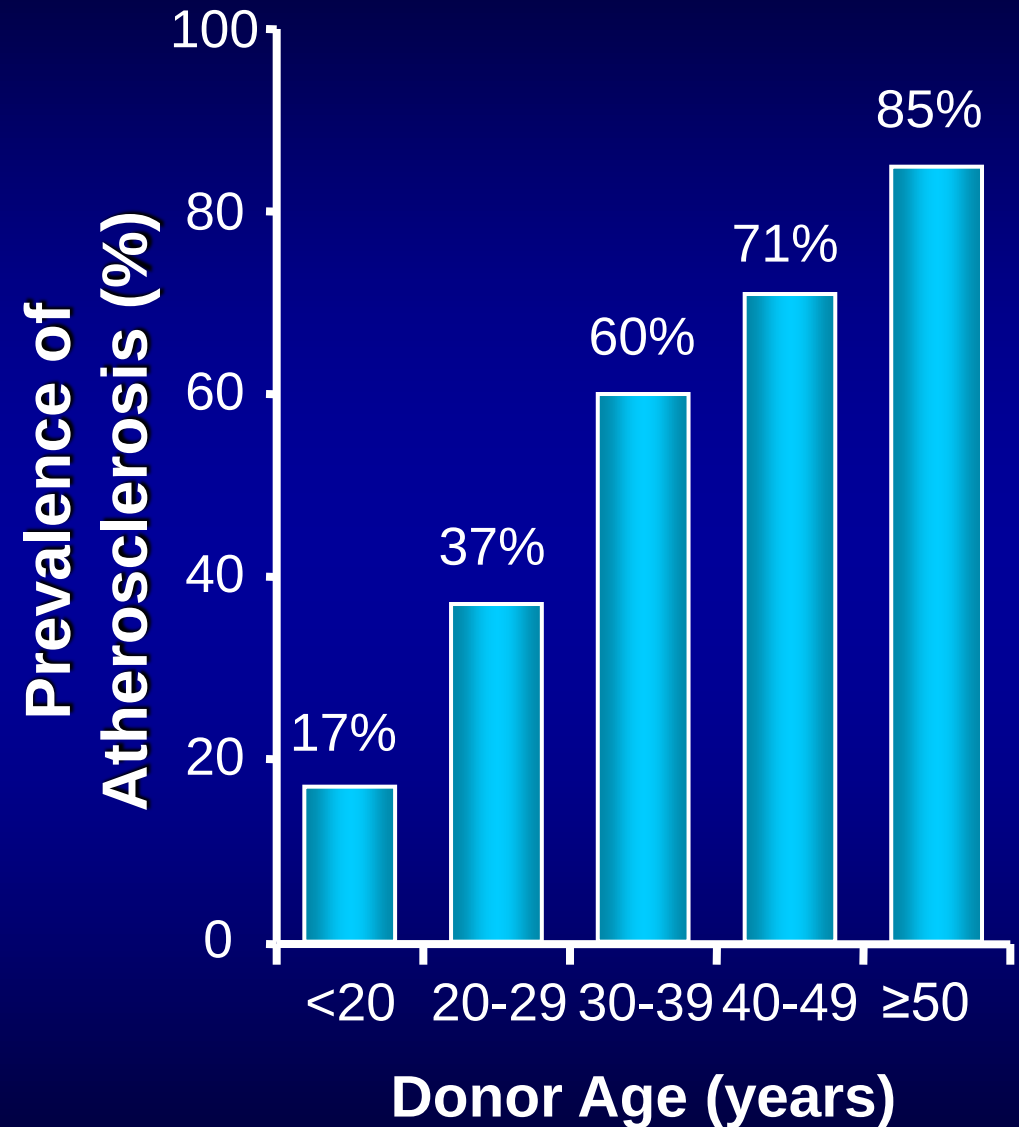
ACHD Outcome: Impact of acquired CVD



Prevalence of Atherosclerosis by Donor Age



32 Year Old Female

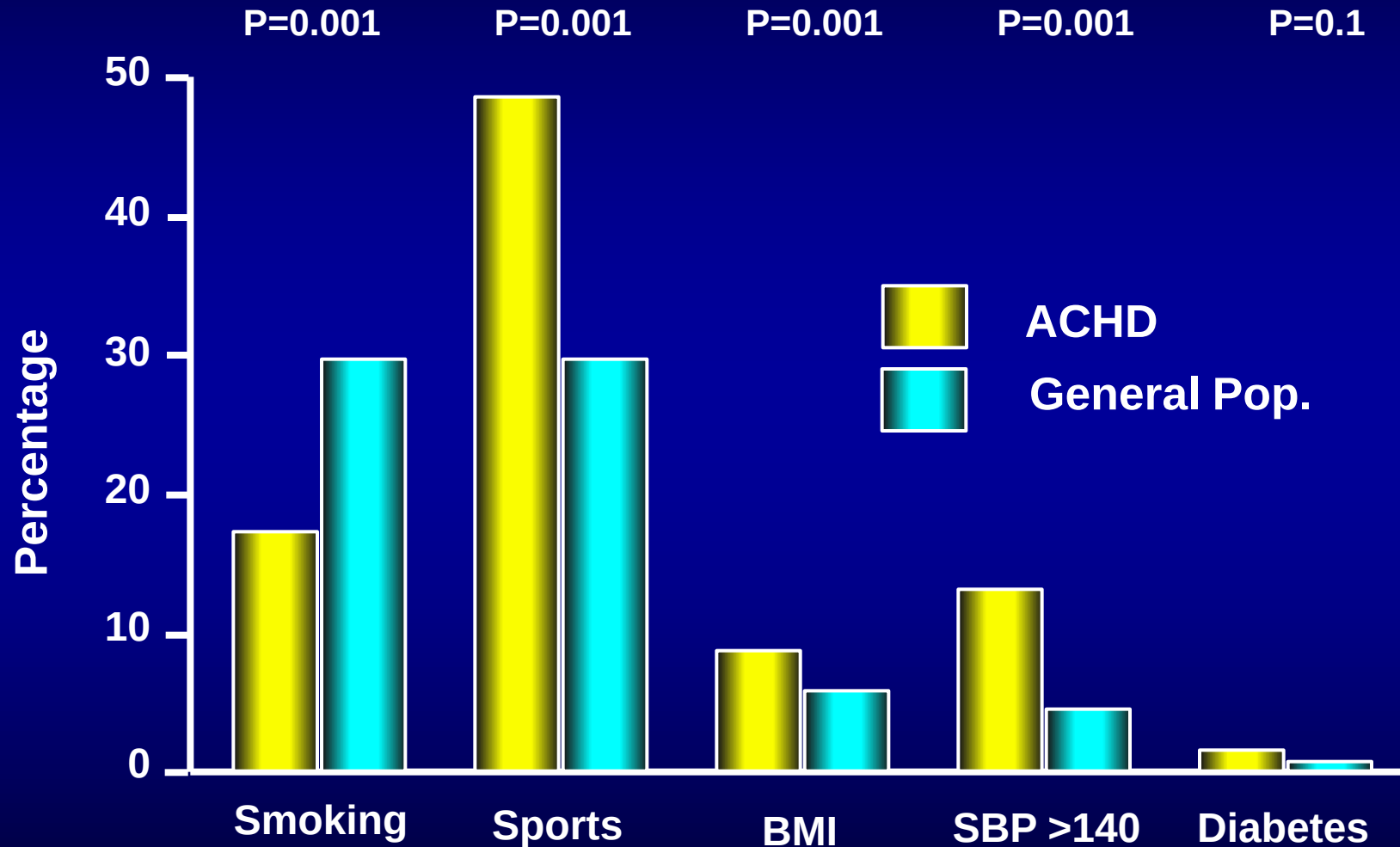


Impact of Acquired CVD in ACHD

- Are risk factors equally common in ACHD?
- What is impact of RFs in ACHD-more or less?
- Specific ACHD conditions which predispose to RFs eg BP
- Specific conditions more vulnerable to acquired heart disease?

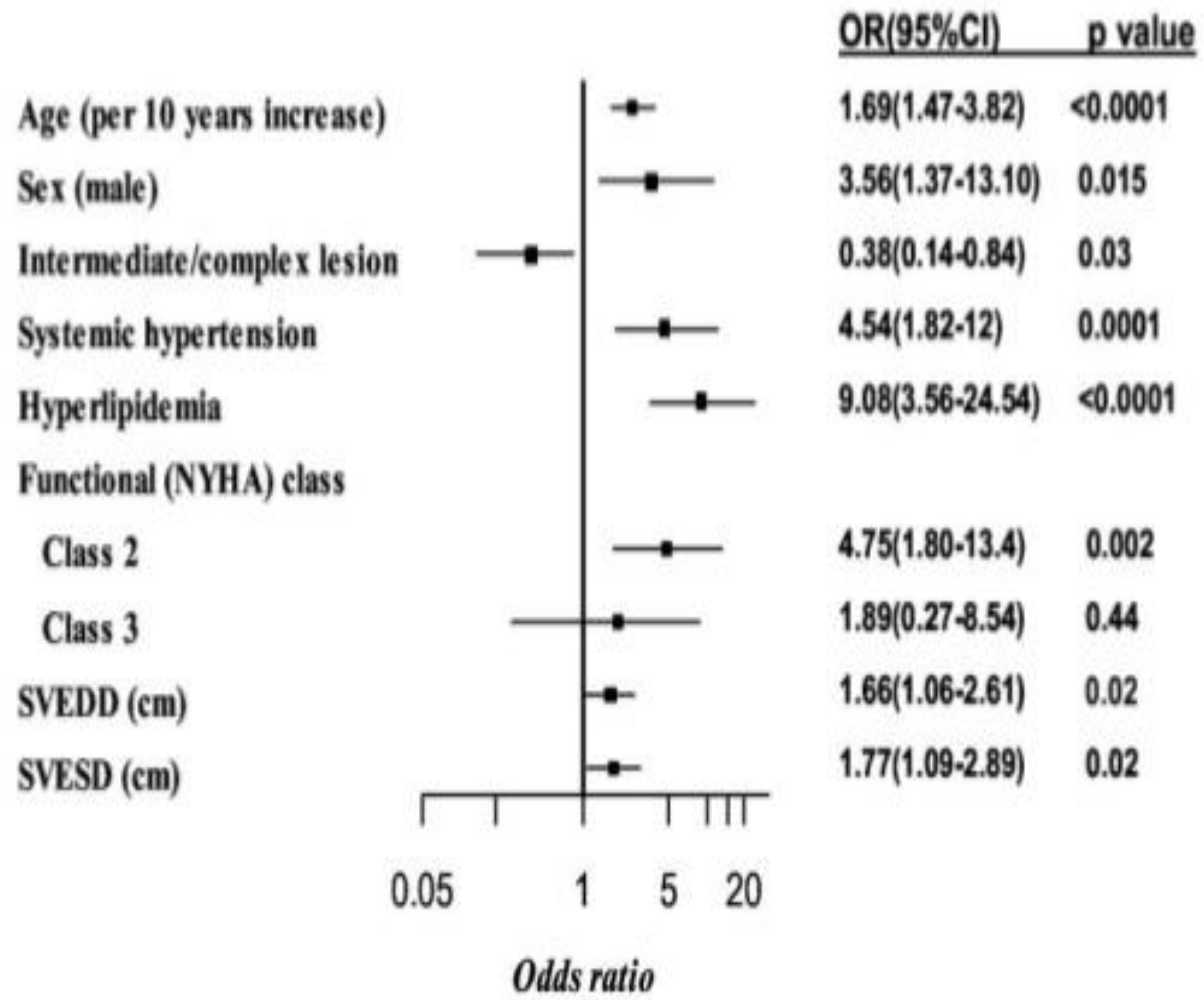
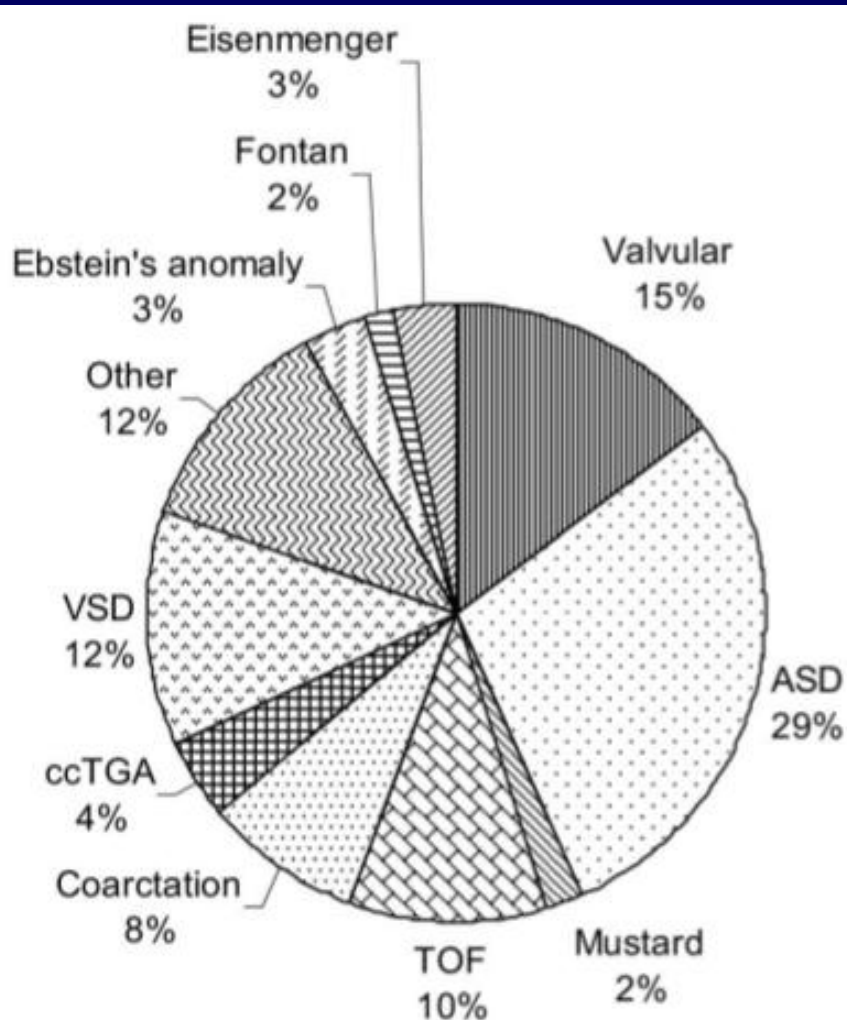
CVRFs in ACHD Population

1976 patients (545 male, median age 26yrs) and 2550 controls



Risk Factors for Coronary Disease in ACHD

250 pts (53% men, 51yrs) 9.2% CAD

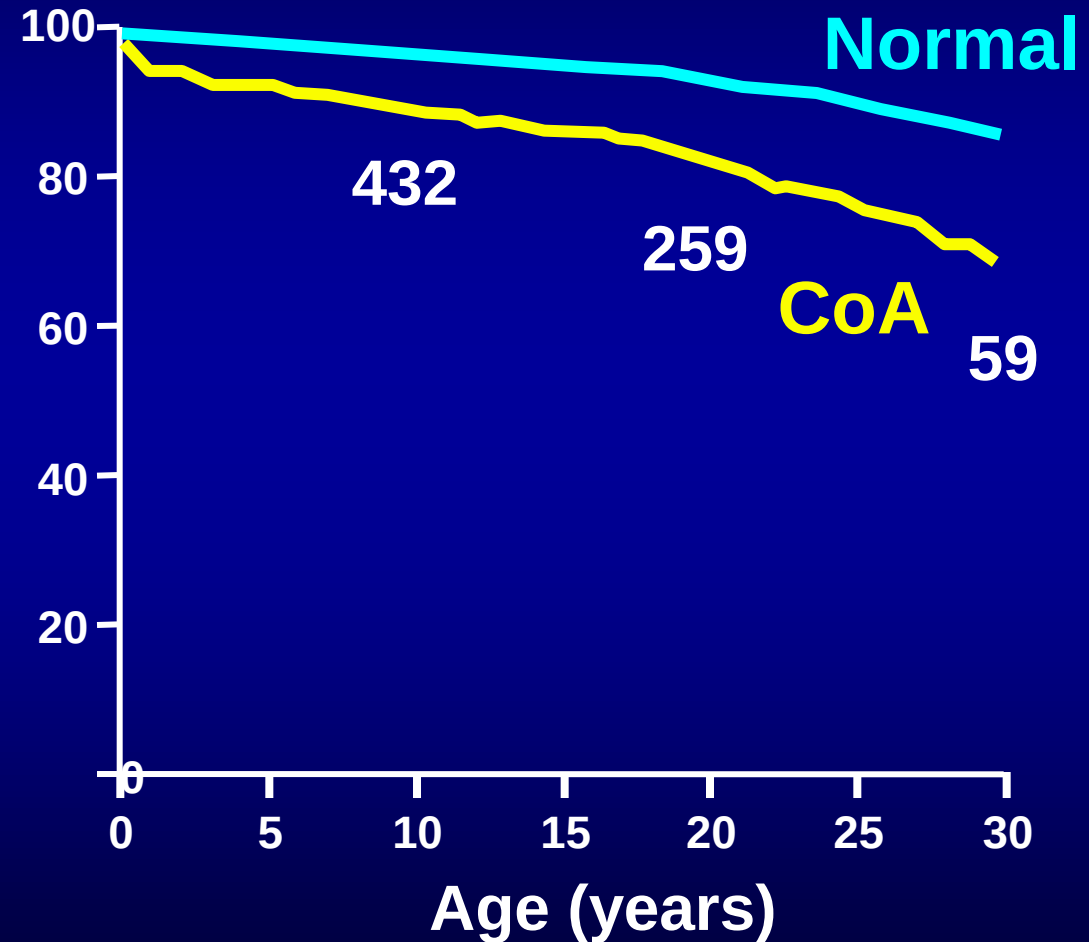


Giannakoulas AJC 2009; 103: 1445-1450

Coarctation – Long Term Outcome

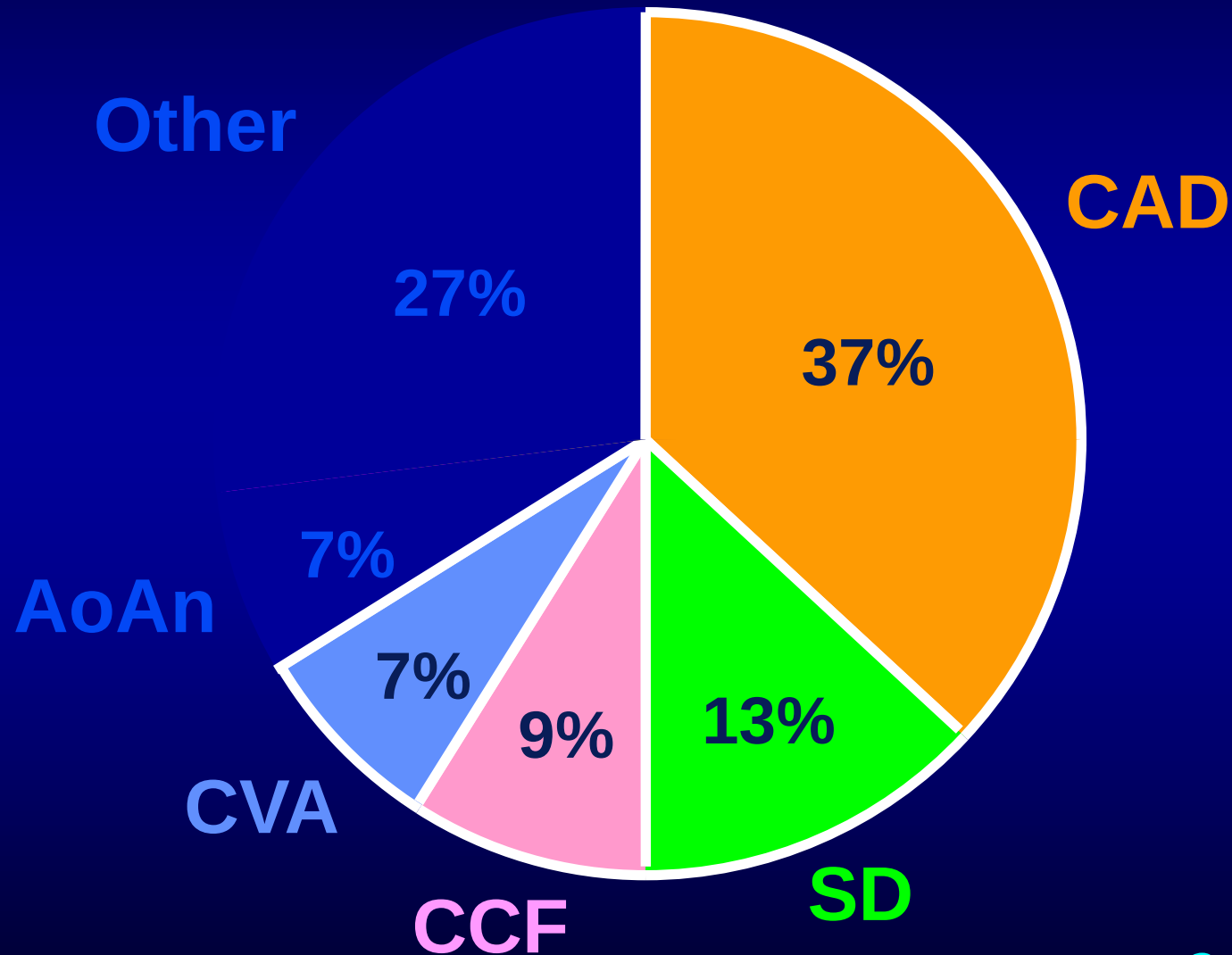


Mayo Clinic 1946-81 – 646 pts

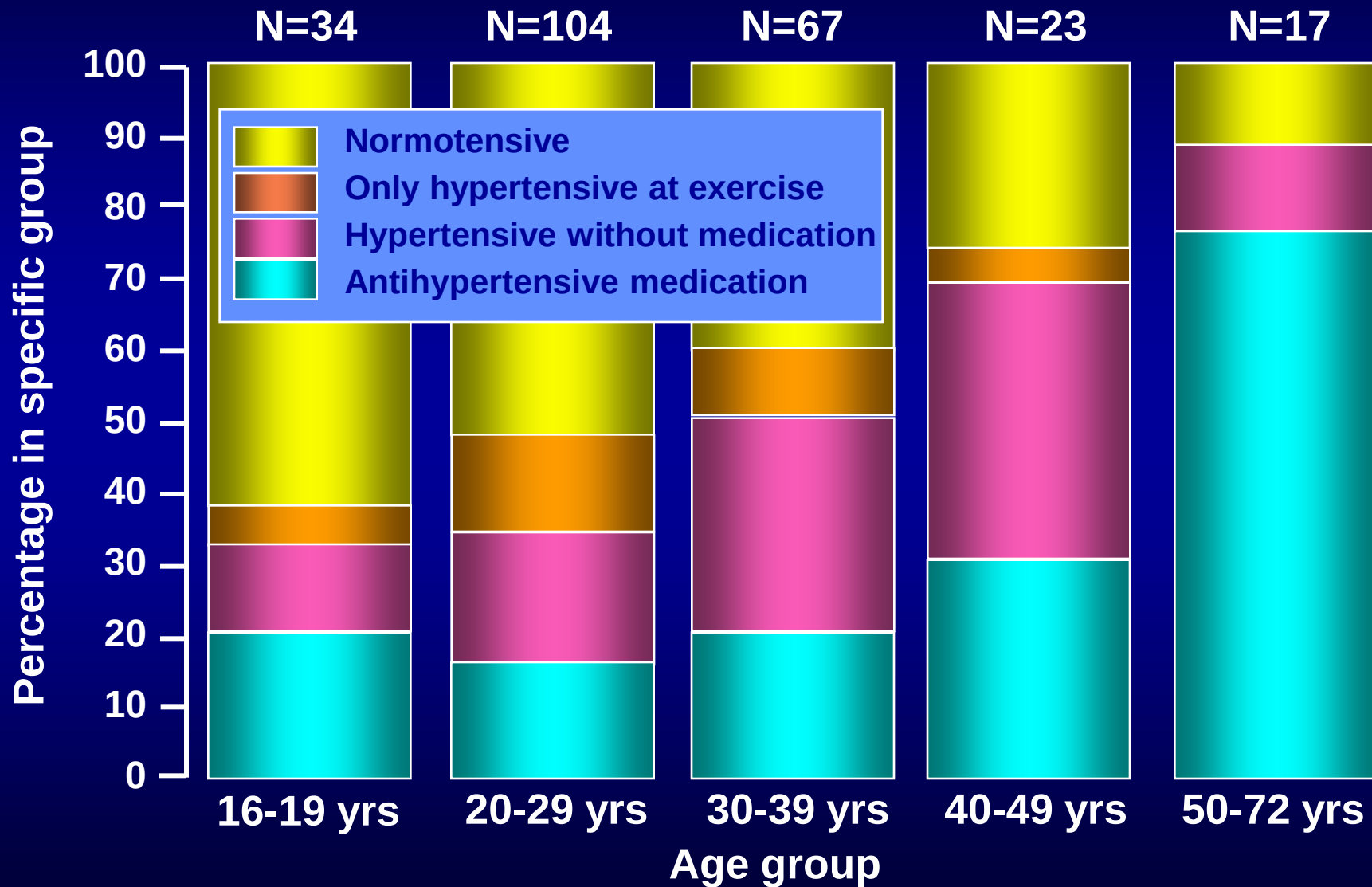


Cohen et al Circ 1989

Coarctation – Causes of Late Death

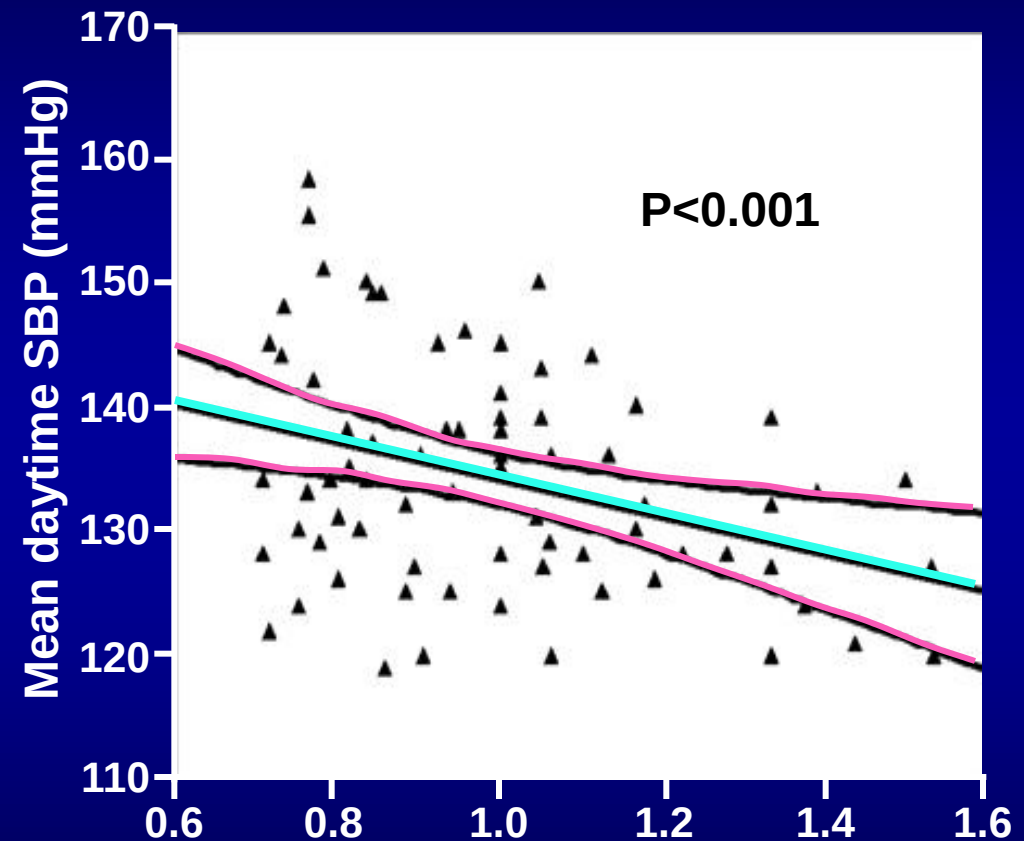
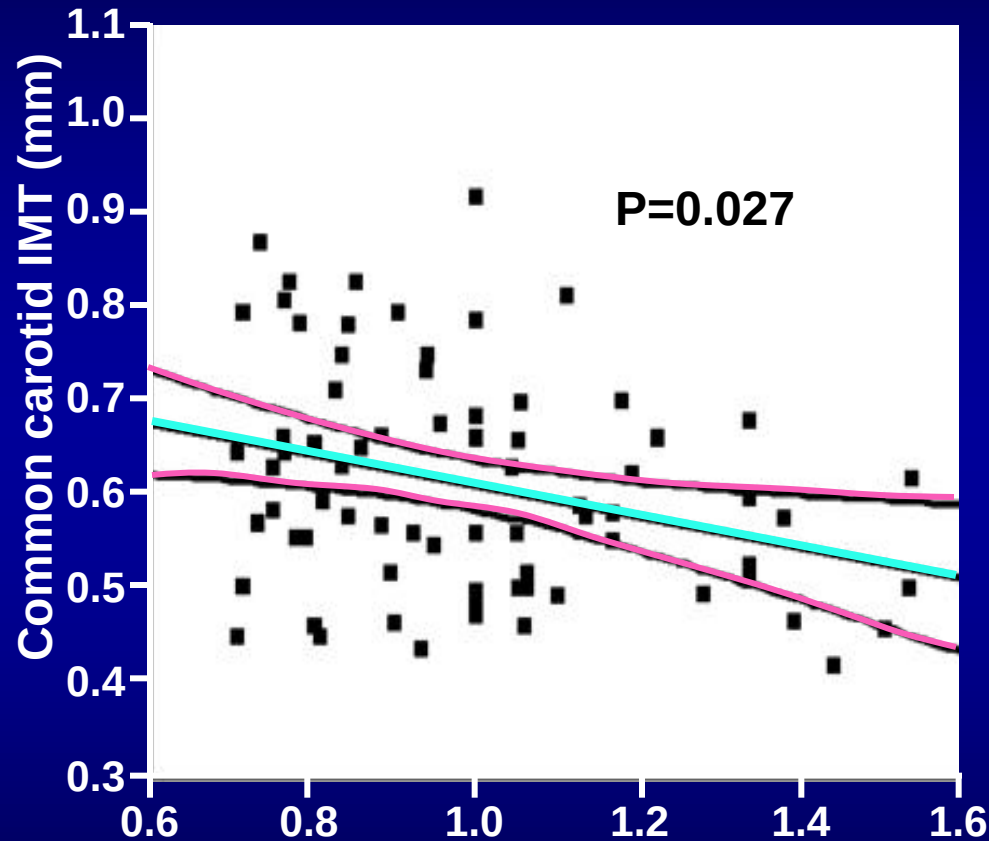


Prevalence of Hypertension after Coarctation Repair



Hager JTCS 2007; 134: 738-45

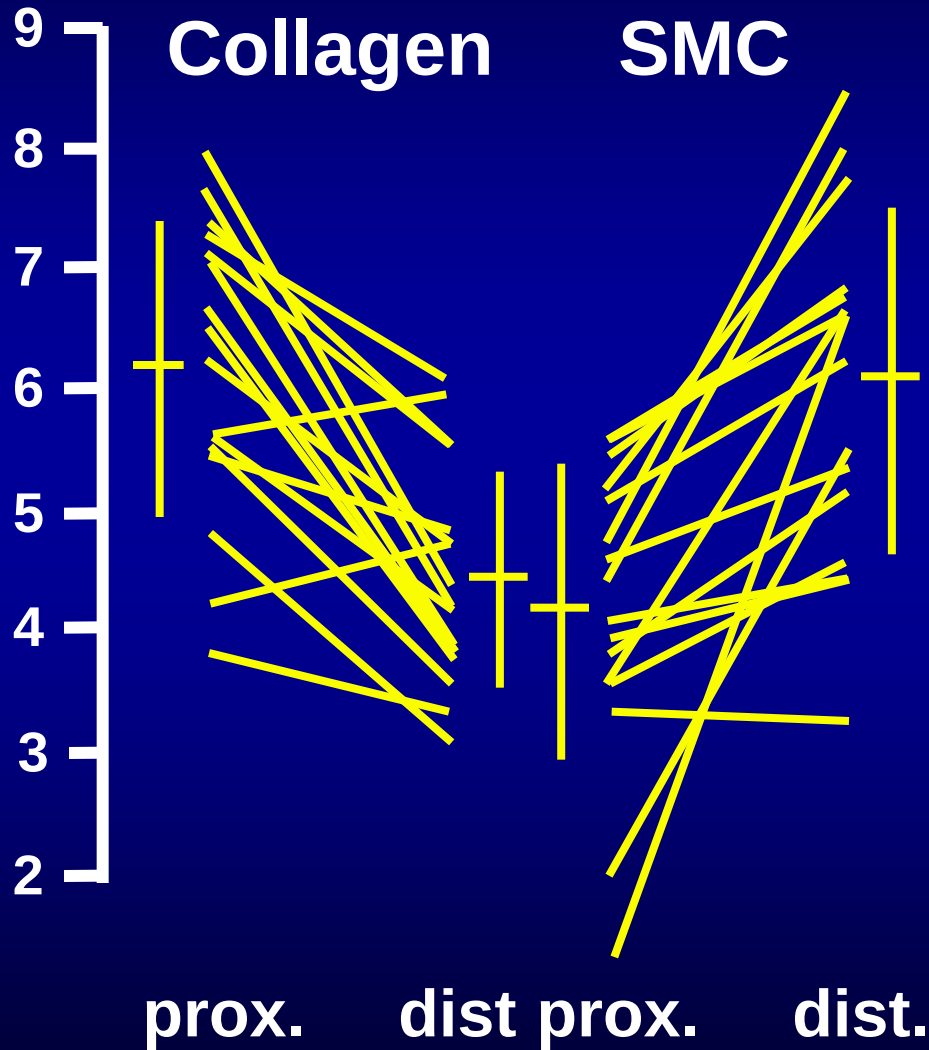
Mild Residual Coarctation and BP, Vascular Outcome



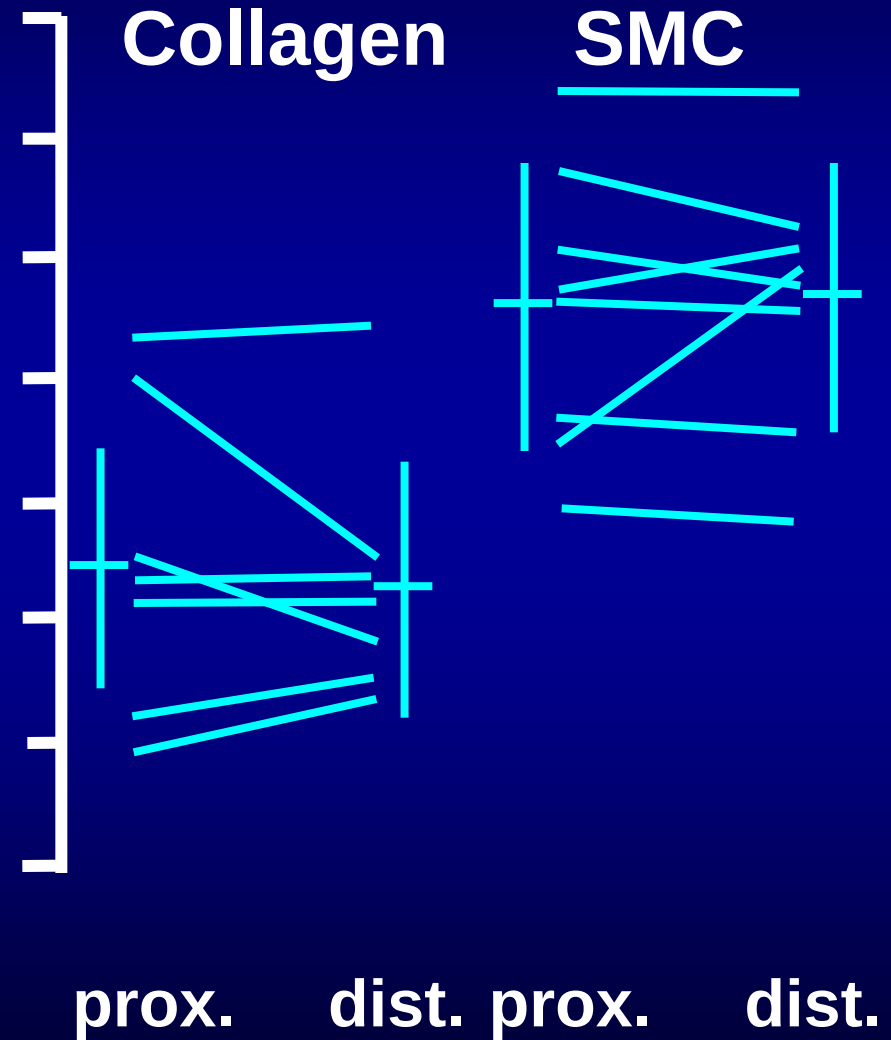
Site of repair / diaphragmatic aorta ratio

Aortic Structure in Coarctation

Coarctation (n = 8)

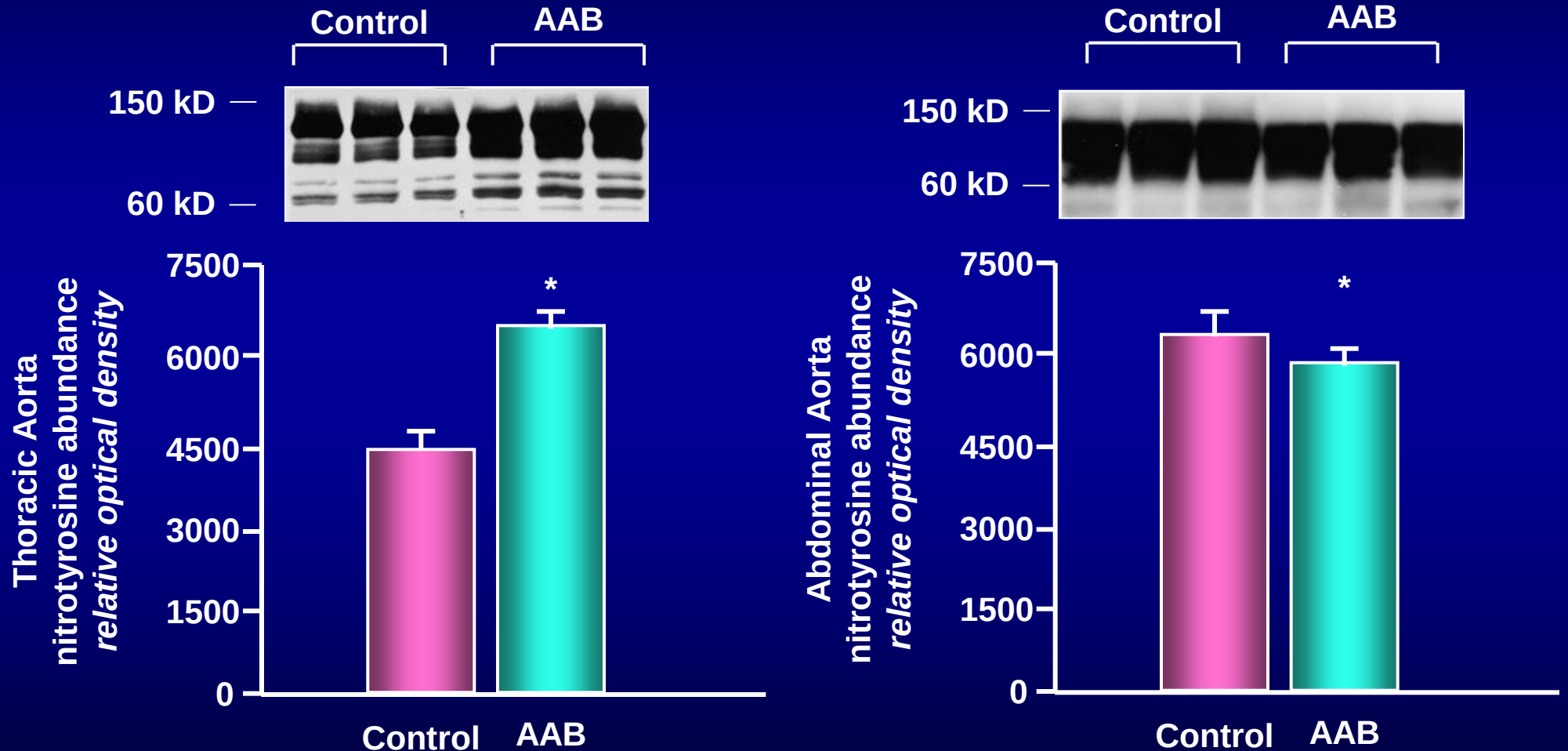


Controls (n = 8)



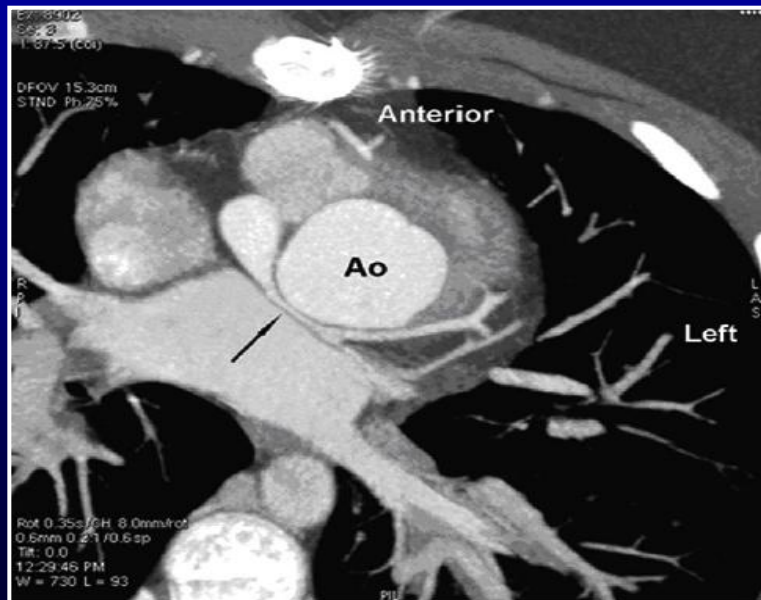
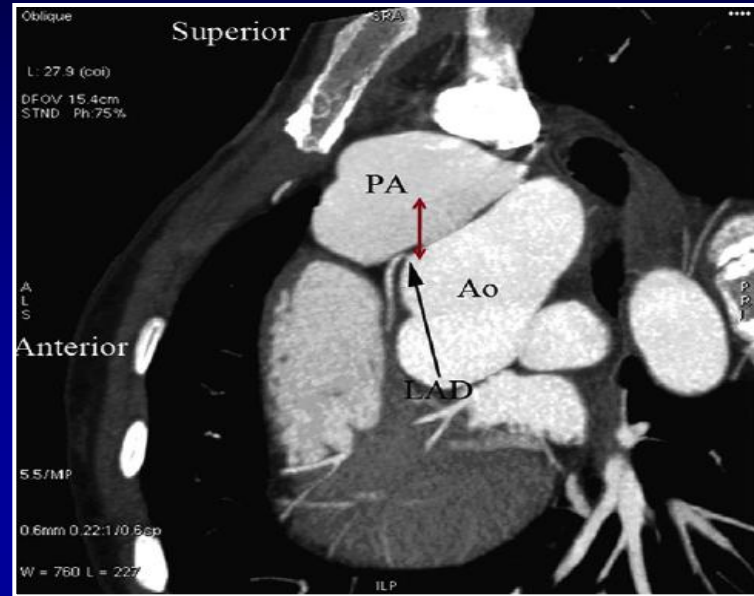
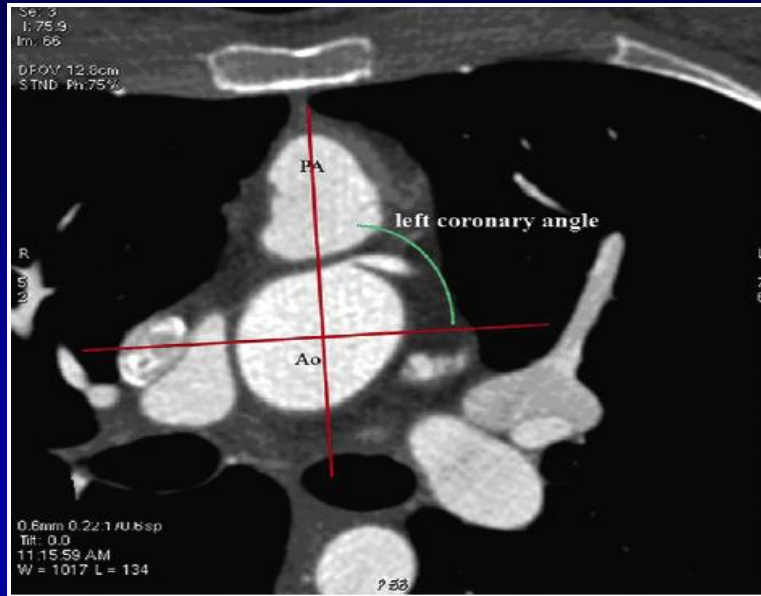
Oxidative Stress, Hypertension in Experimental Coarctation

12 male Sprague Dawley Rats



Barton et al Kidney Int. 2001; 60: 1083-1087

Coronary Complications after ASO: Vulnerability for Atherosclerosis and Myocardial Ischaemia



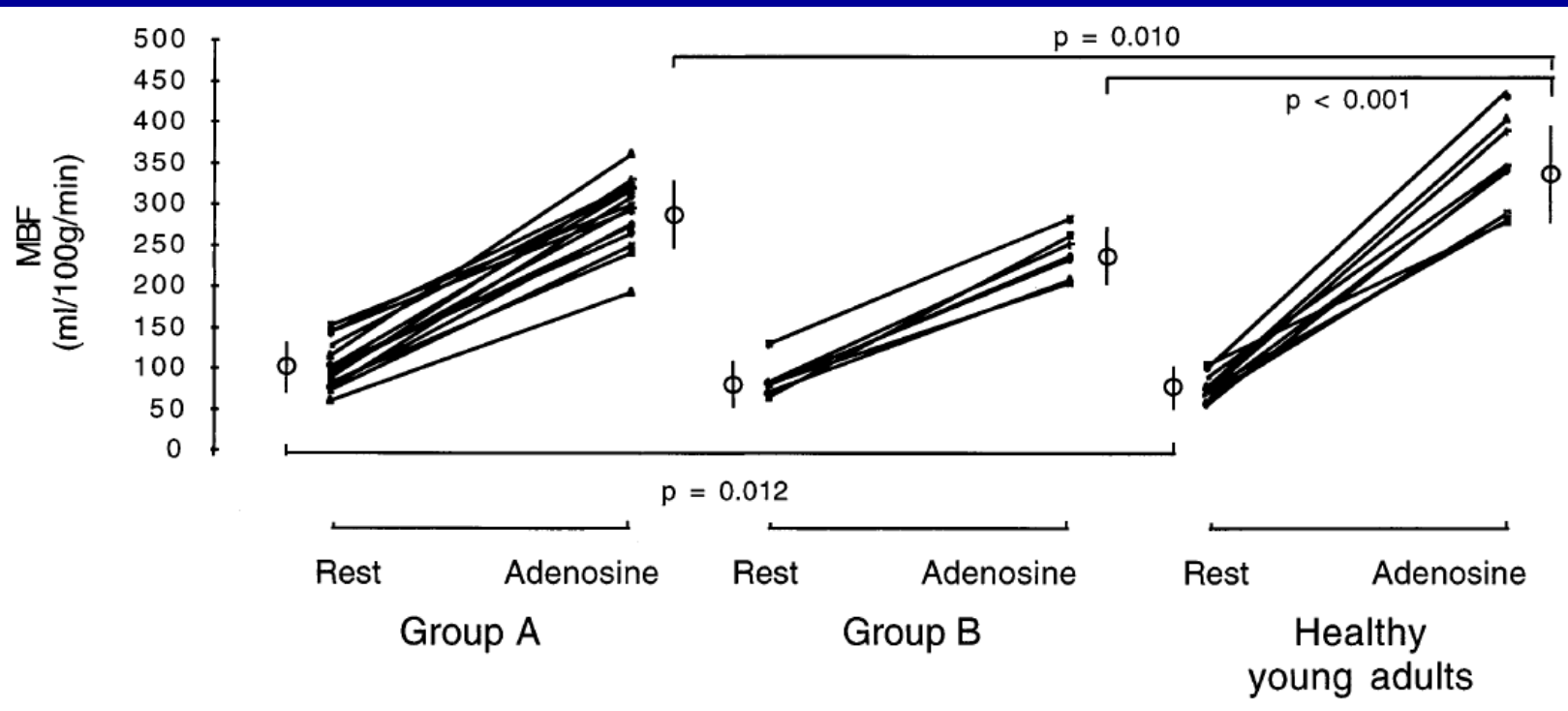
Myocardial Blood Flow and Coronary Flow Reserve Late After Anatomical Correction of Transposition of the Great Arteries

FRANK M. BENGEL, MD, MICHAEL HAUSER, MD,* CLAIRE S. DUVERNOY, MD,
ANDREAS KUEHN, MD,* SIBYLLE I. ZIEGLER, PhD, JENS C. STOLLFUSS, MD,
MAREIKE BECKMANN, MS,* URSULA SAUER, MD,* OTTO MUZIK, PhD†,
MARKUS SCHWAIGER, MD, JOHN HESS, MD*

Munich, Germany and Detroit, Michigan

JACC 1998; 32 :1955-1961

22 pts N-13 Ammonia PET 10yrs after ASO



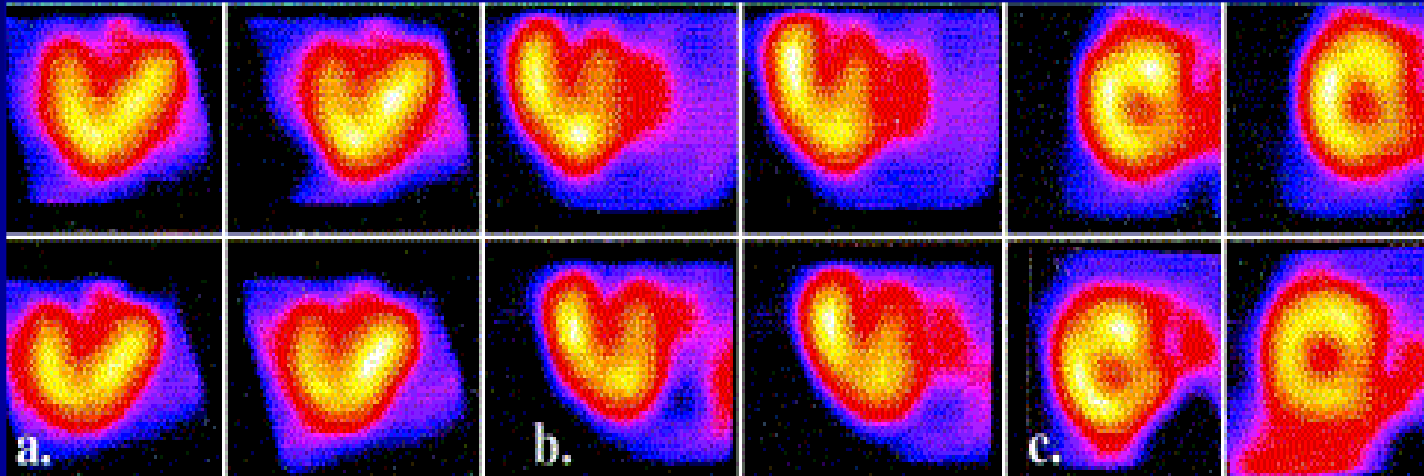
Why should the systemic RV fail?

- ❑ 'Designed for a low afterload'
- ❑ Fibre arrangement
- ❑ Coronary supply
- ❑ Tricuspid valve

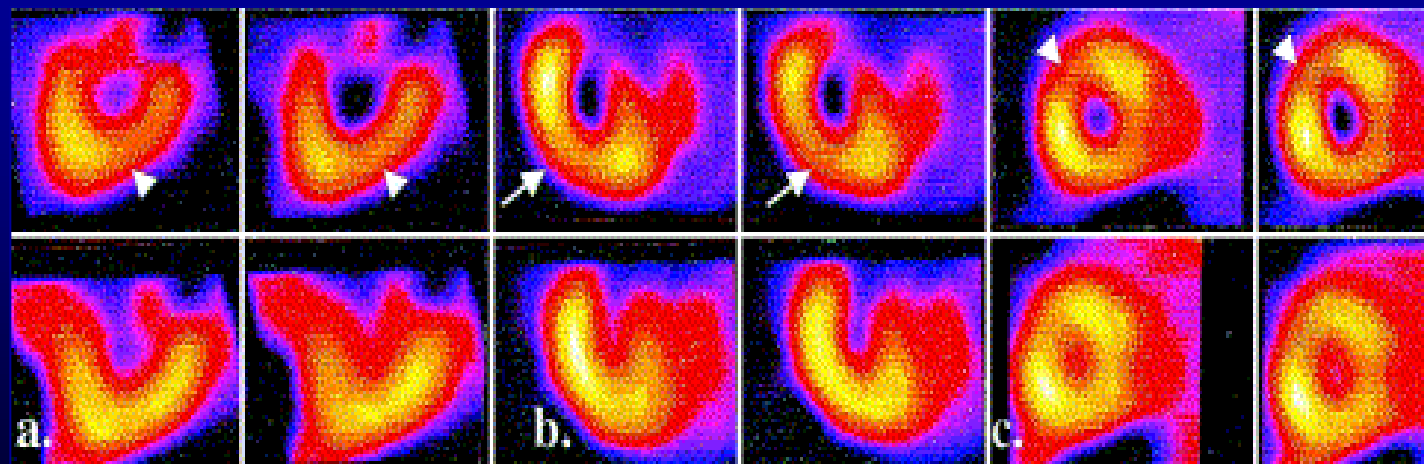


RV Dysfunction after IAR

RV perfusion 7-23 yrs post Mustard (n=61)



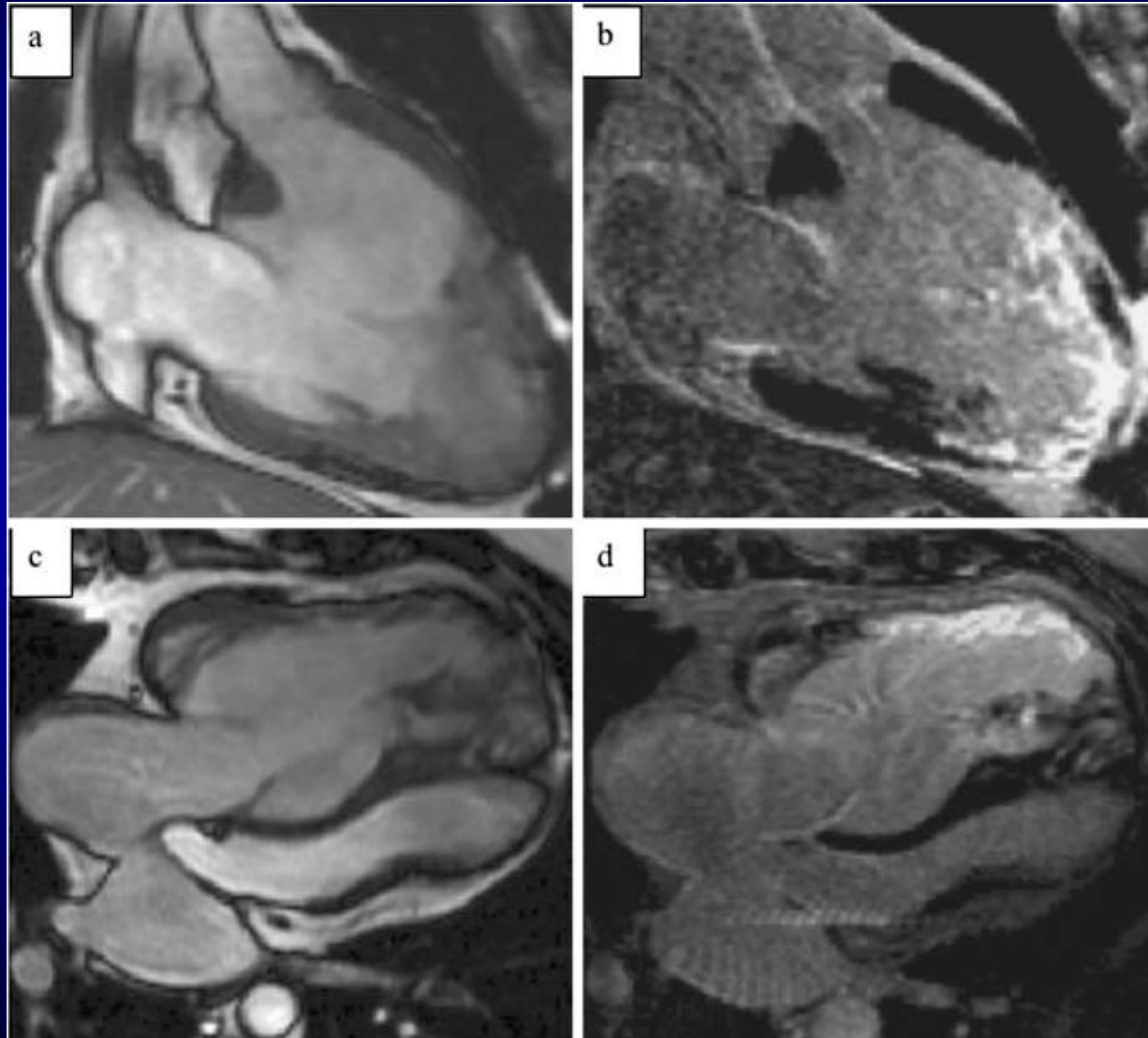
Normal



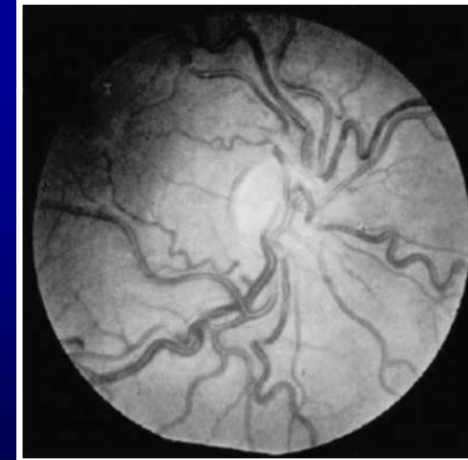
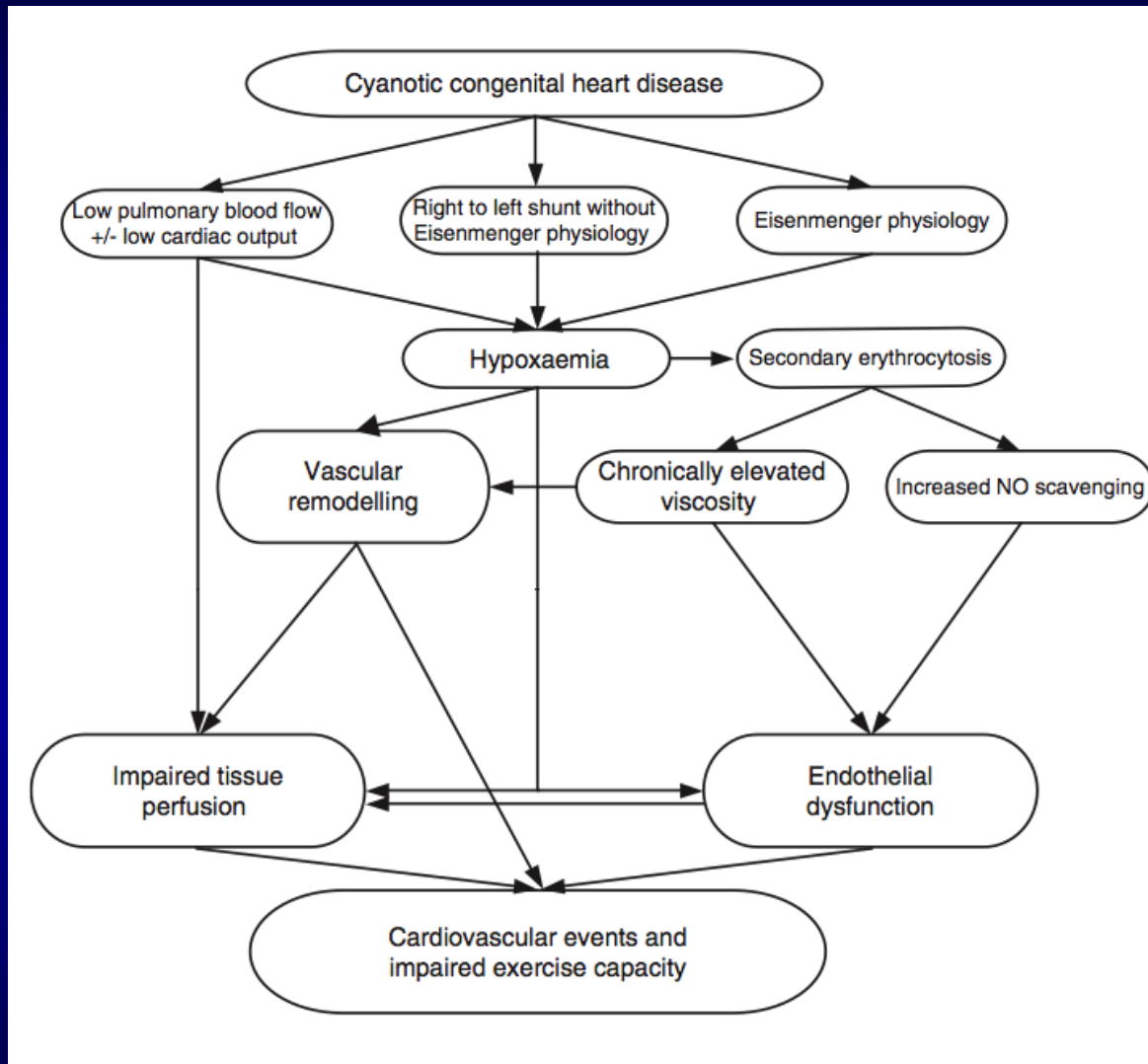
**Perfusion
Defects**

- 20 at rest
- 13 with exercise
- Correlated EF

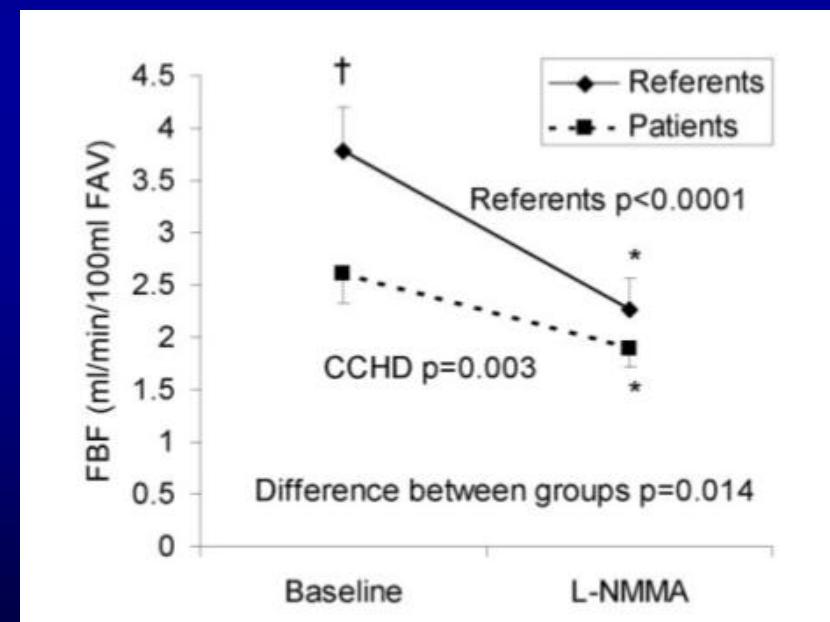
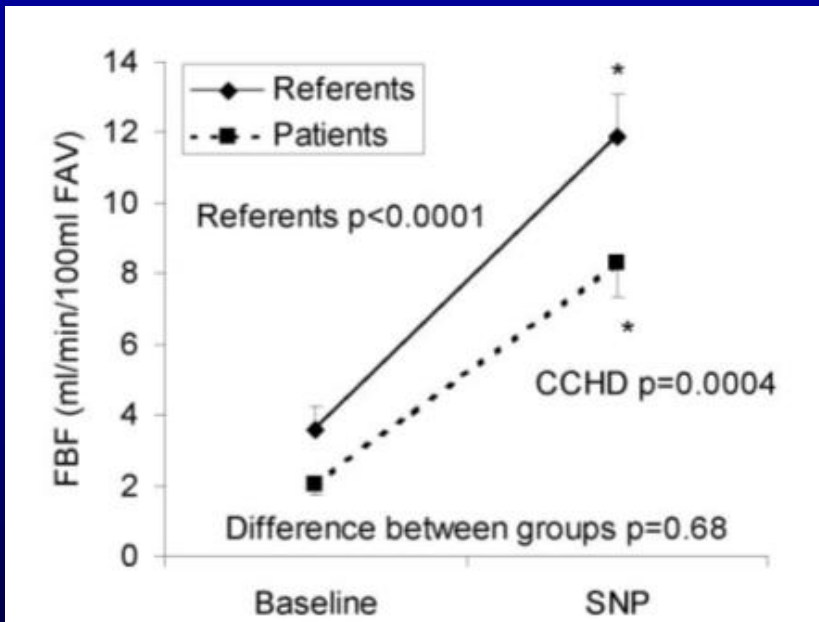
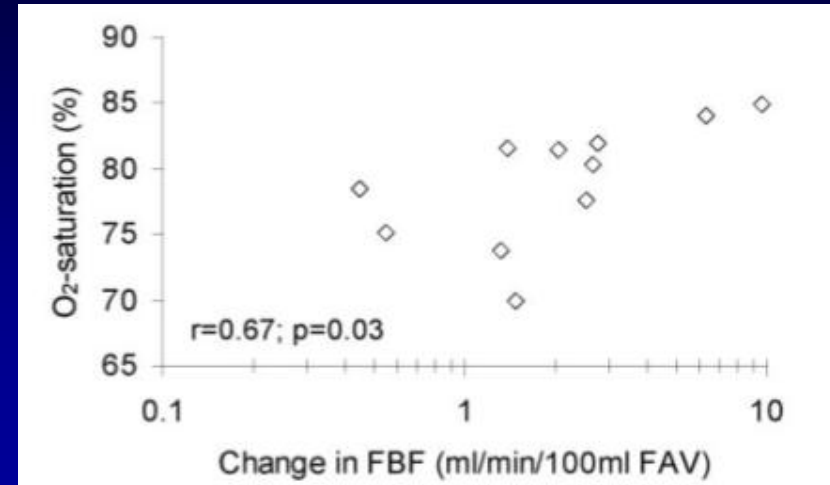
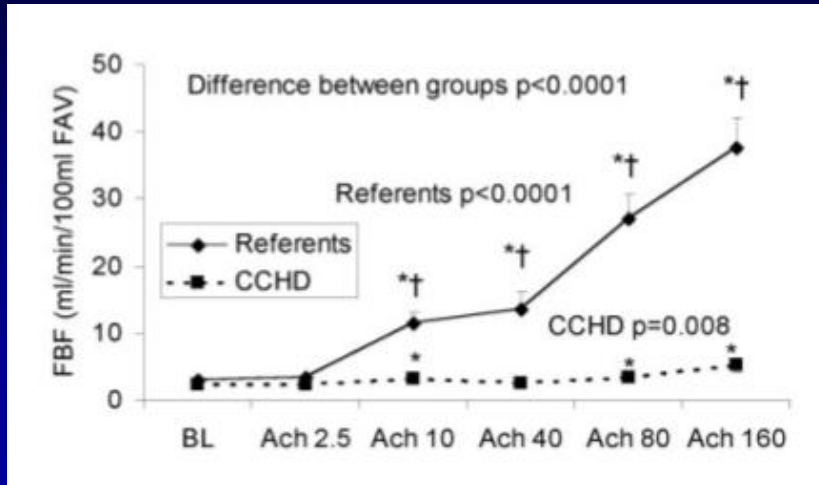
Late Gadolinium Enhancement of the Systemic RV



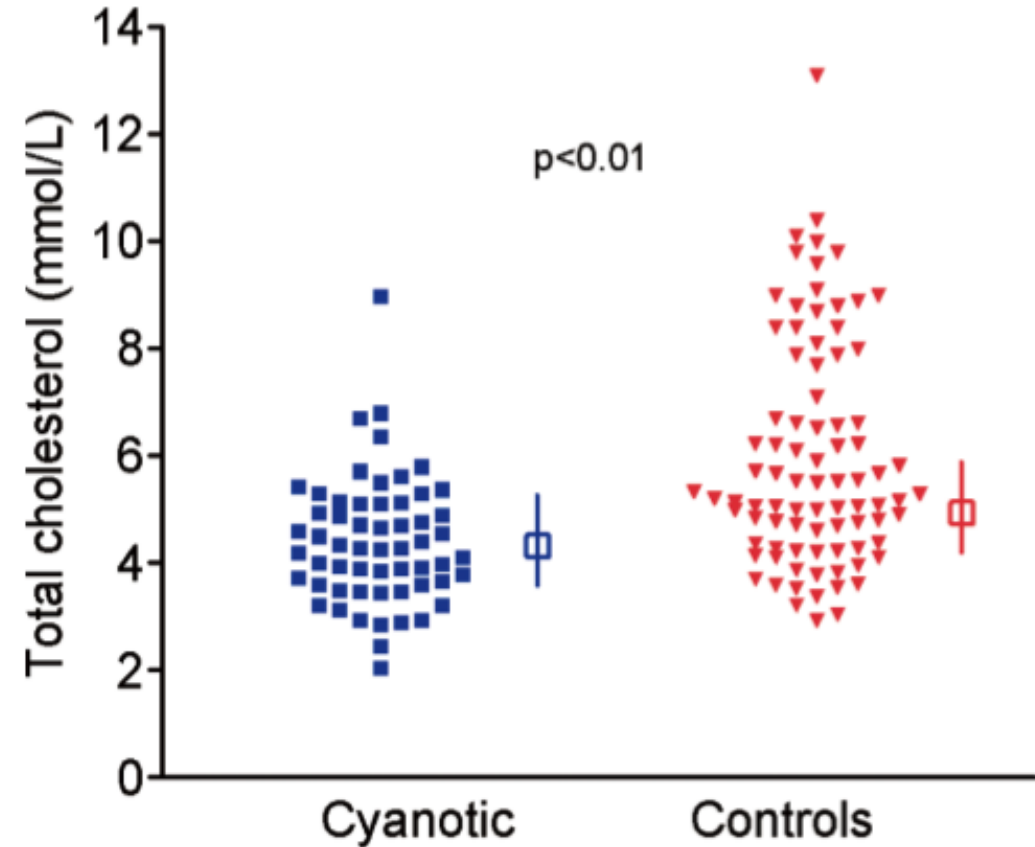
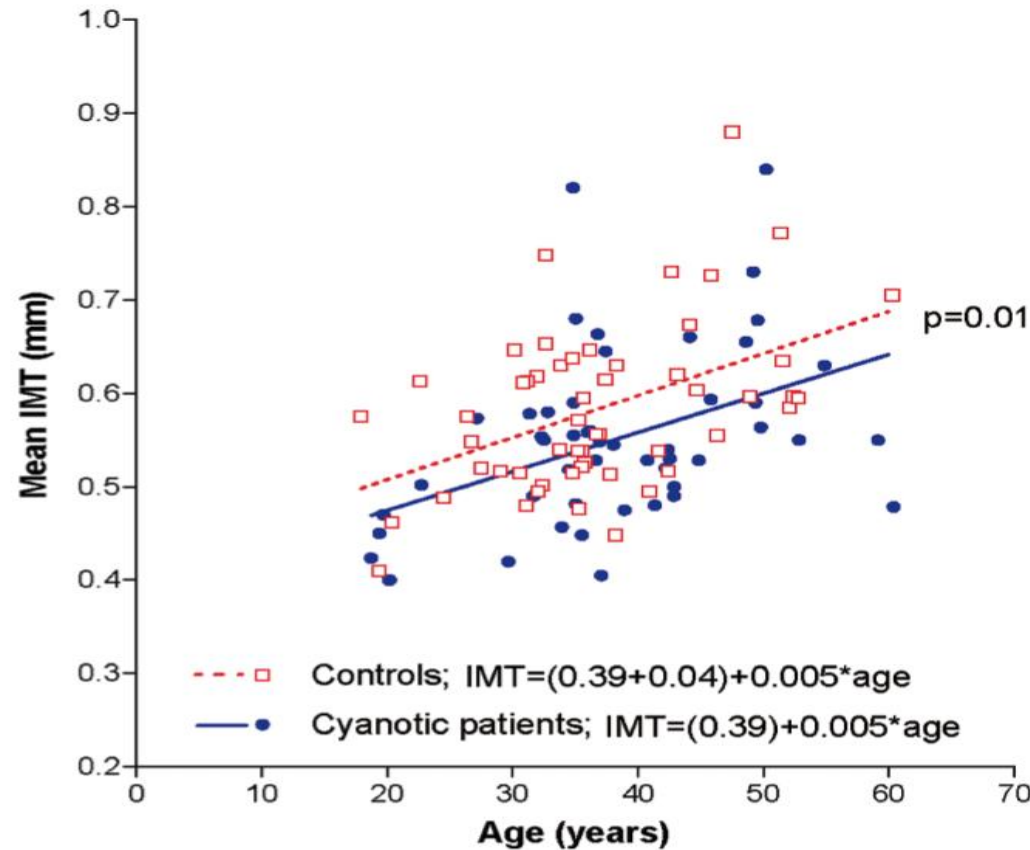
Vascular Function in Cyanotic CHD



Endothelial Dysfunction in Cyanotic CHD



Atherosclerosis in Cyanotic CHD



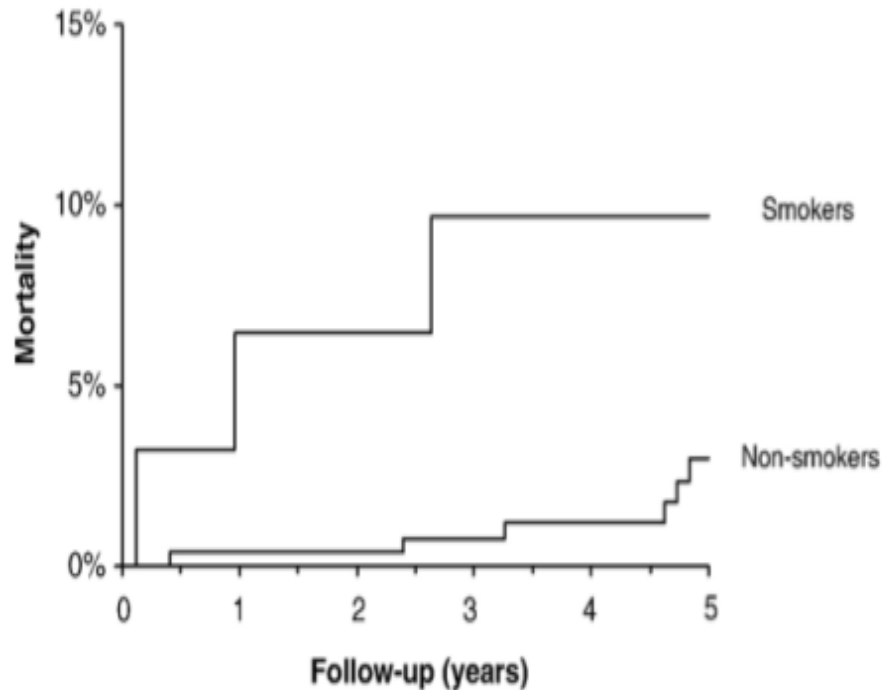
	Cyanotic	Controls	p
Cholesterol (mmol/l)	4.4	4.9	0.02
BP (mmHg)	113/71	124/76	0.01



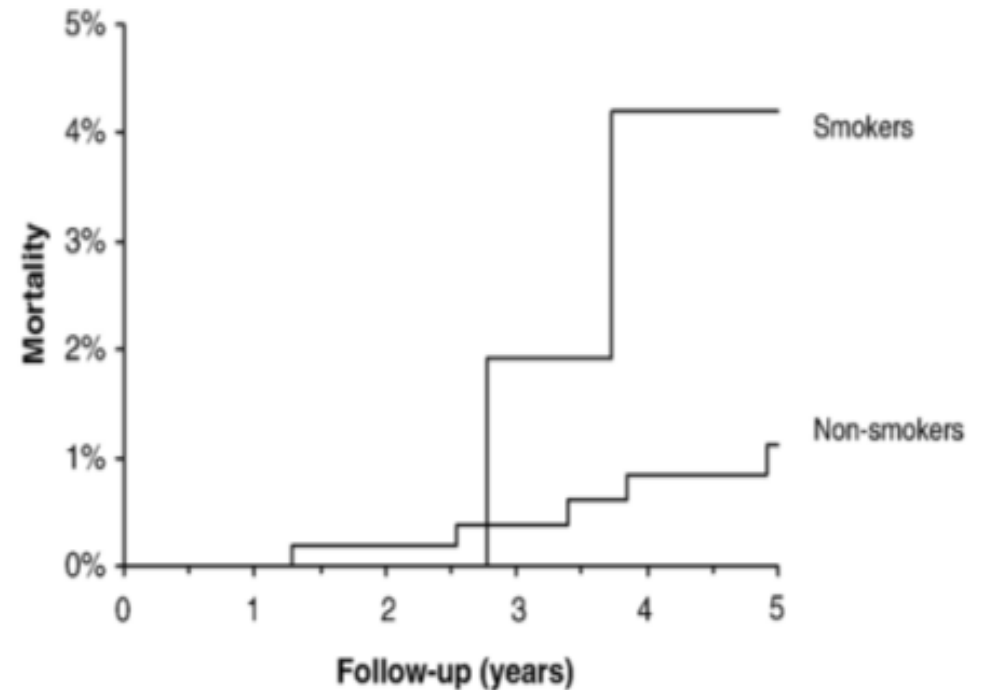
Smoking and Outcome in ACHD

3375 ACHDpts (aged 28yrs 9.3% smokers) Median FU 5.1yrs

Transposition of the Great Arteries

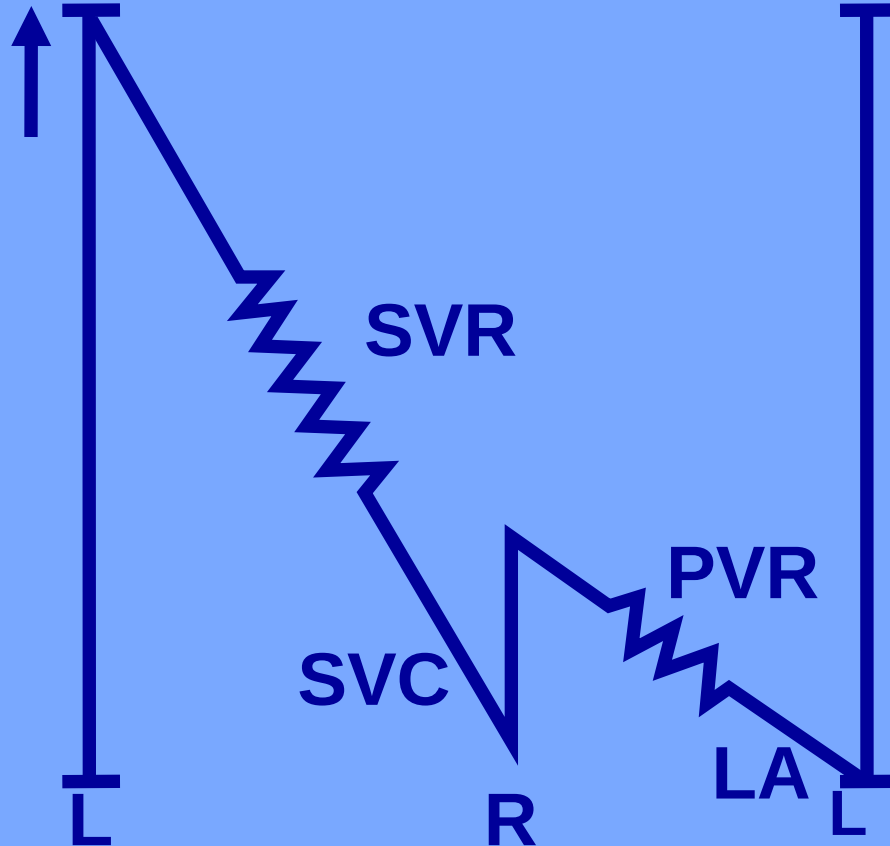


Tetralogy of Fallot

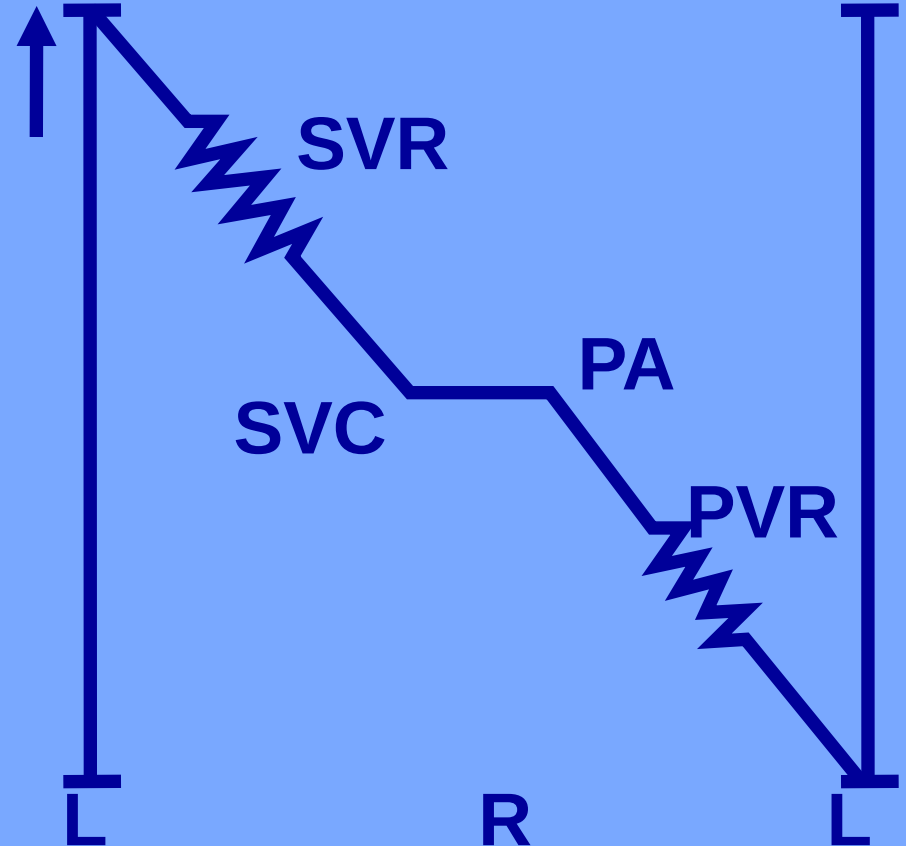


Fontan Circulation

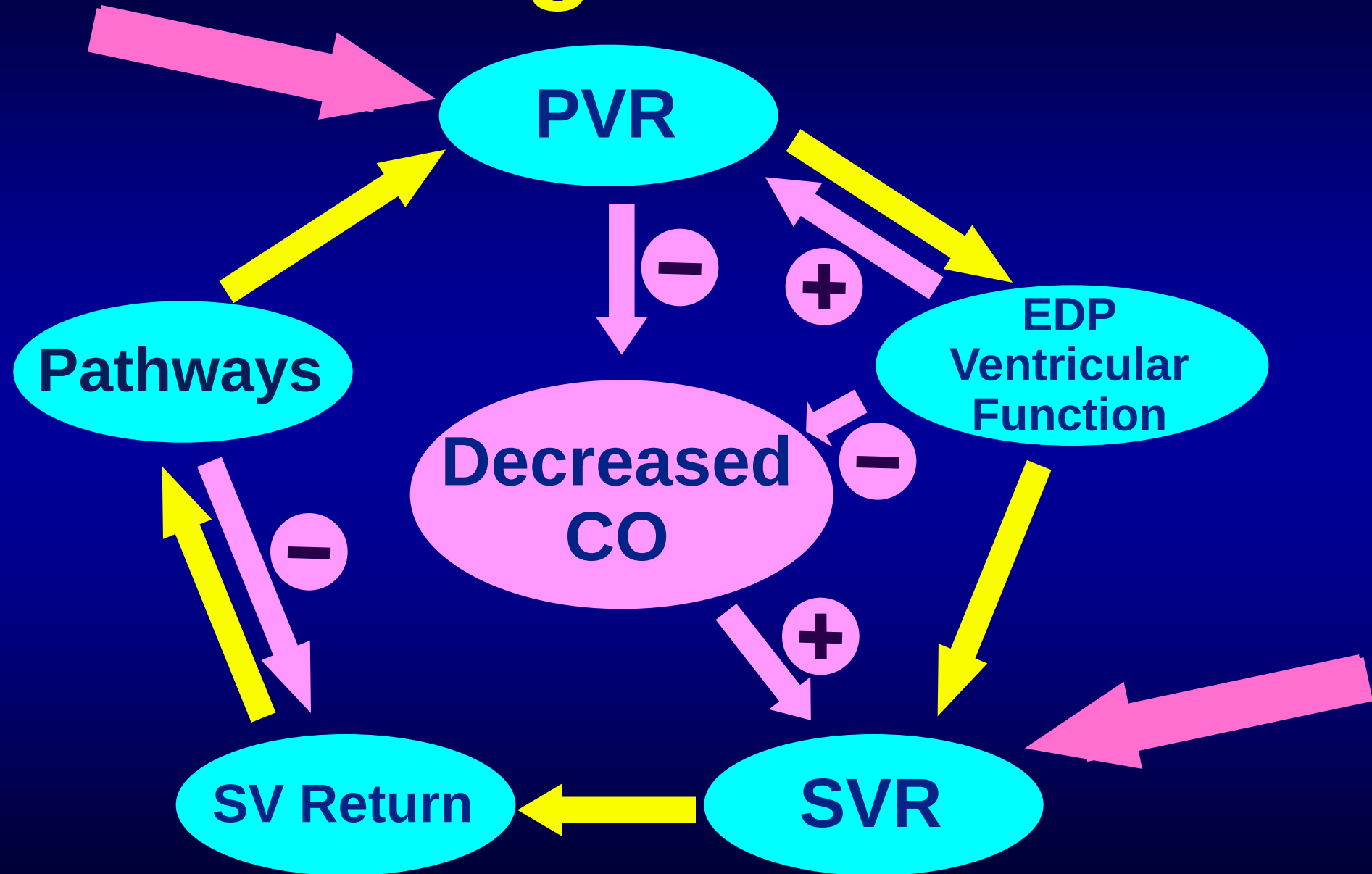
Pressure



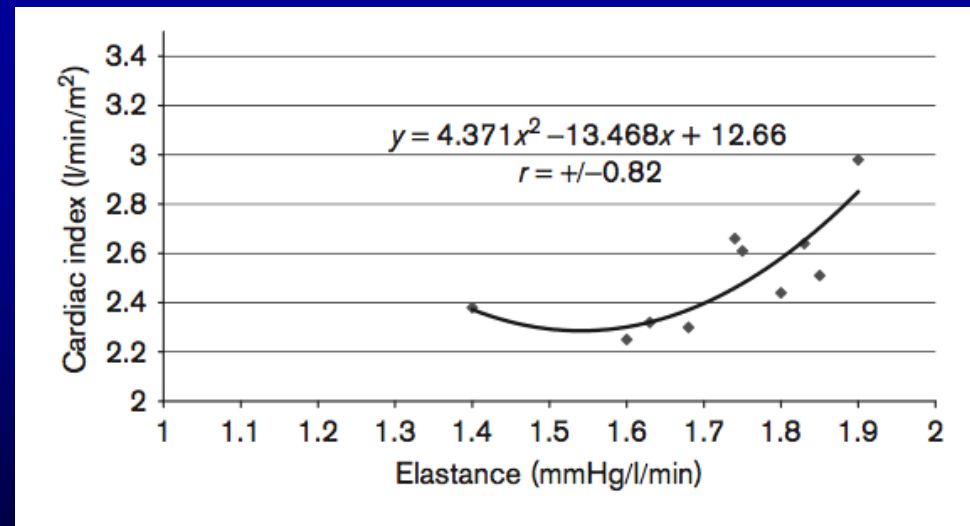
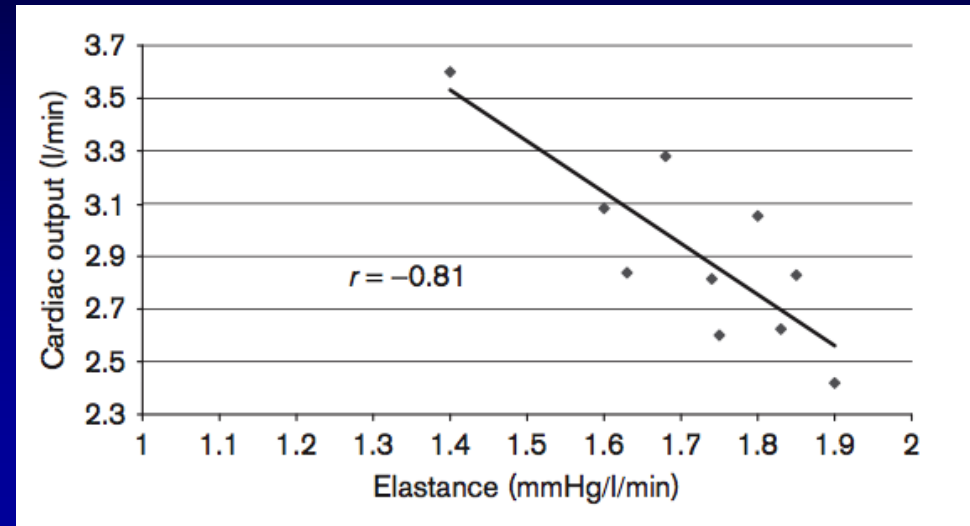
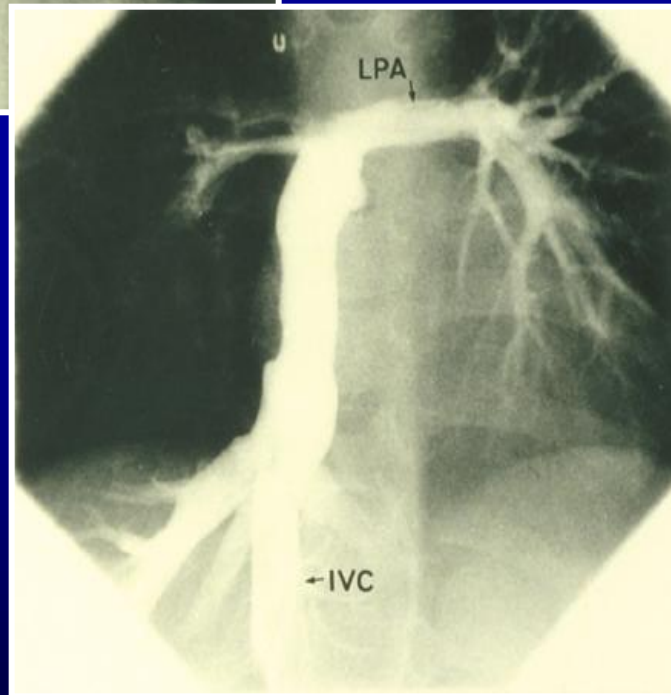
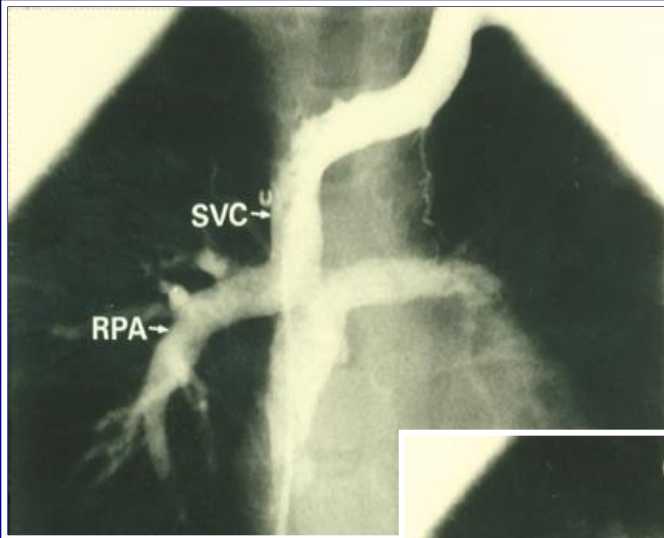
Pressure



Failing Fontan

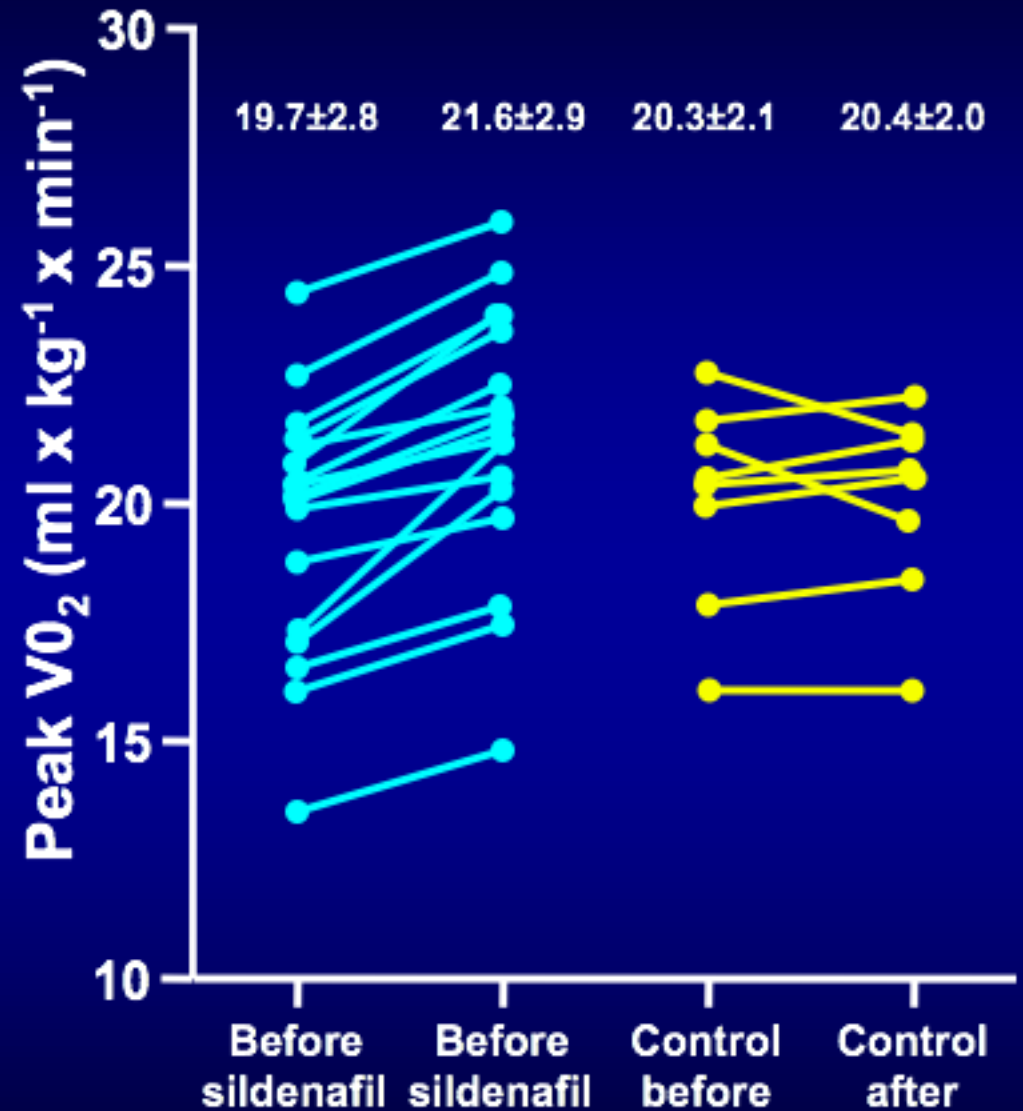


24 BP Monitoring After Fontan





Sildenafil in Fontan



Giardini EHJ 2008

Obesity in the Young

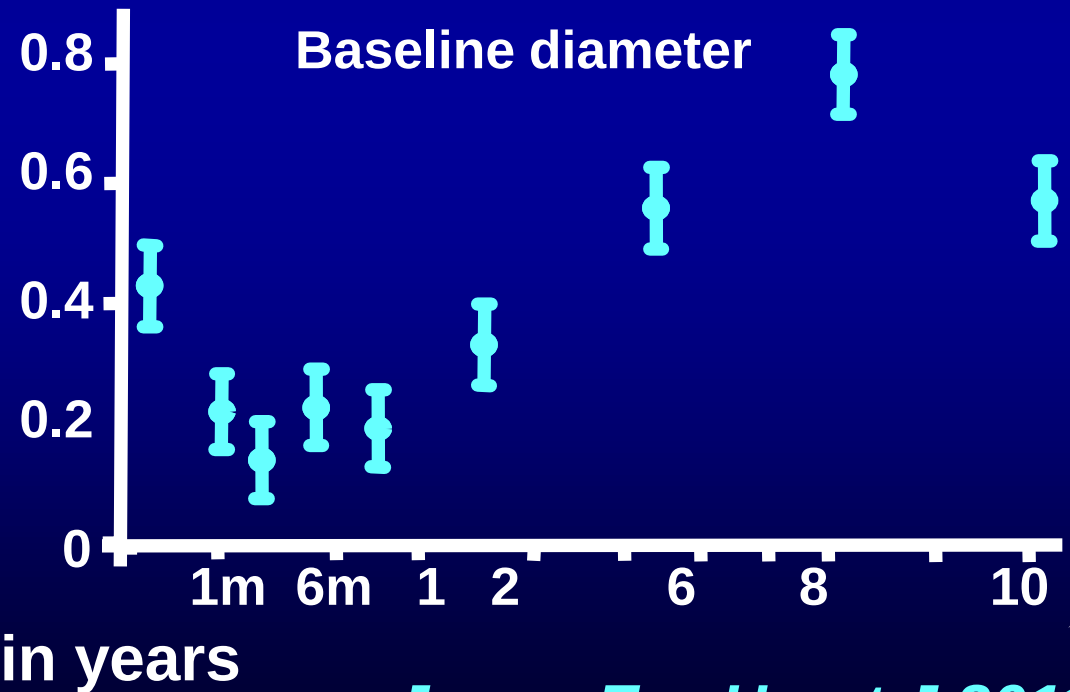
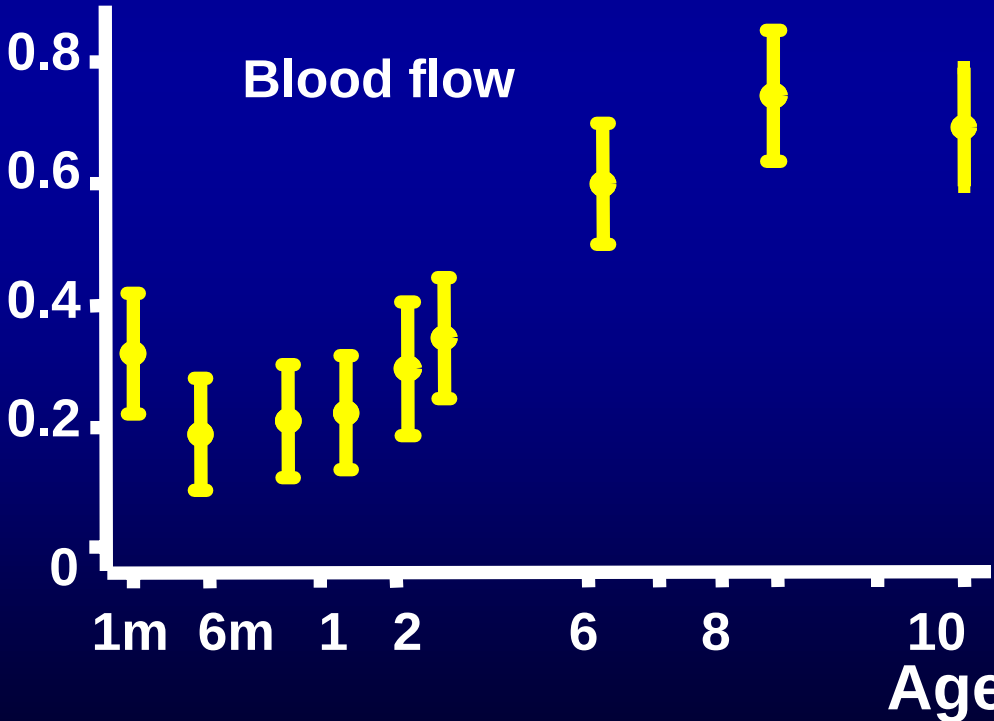


**“The commonest instruments
of suicide are a knife and fork”**

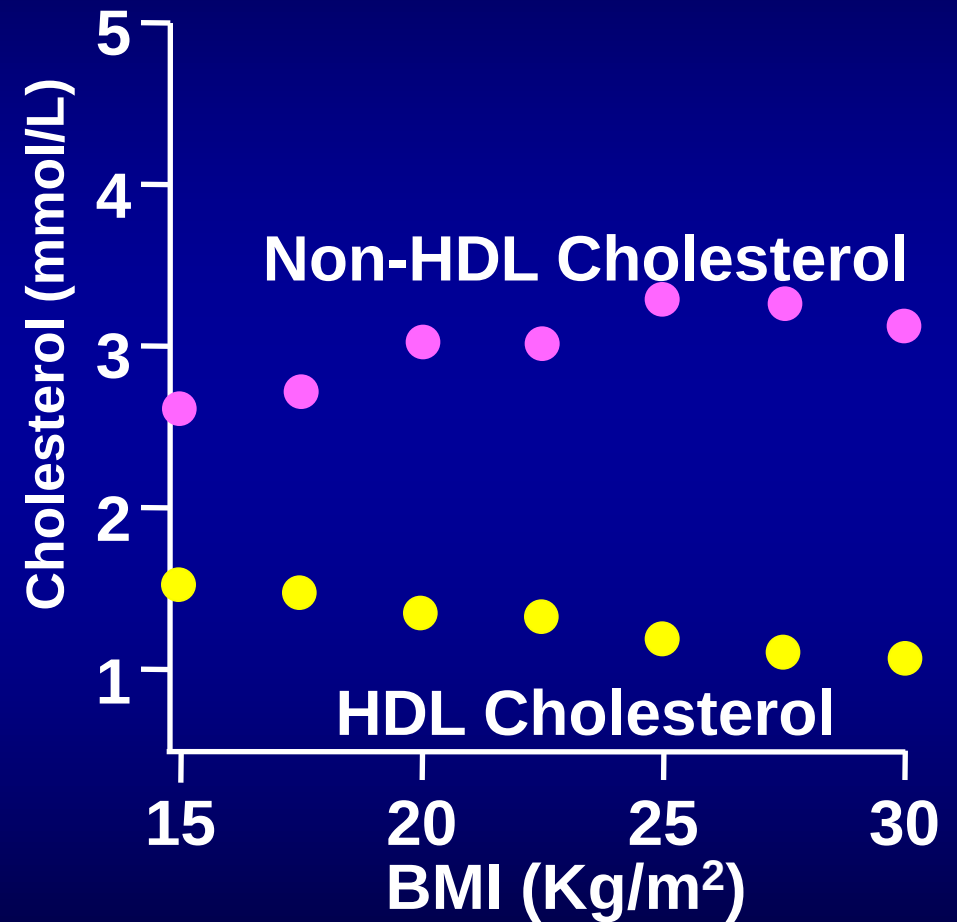
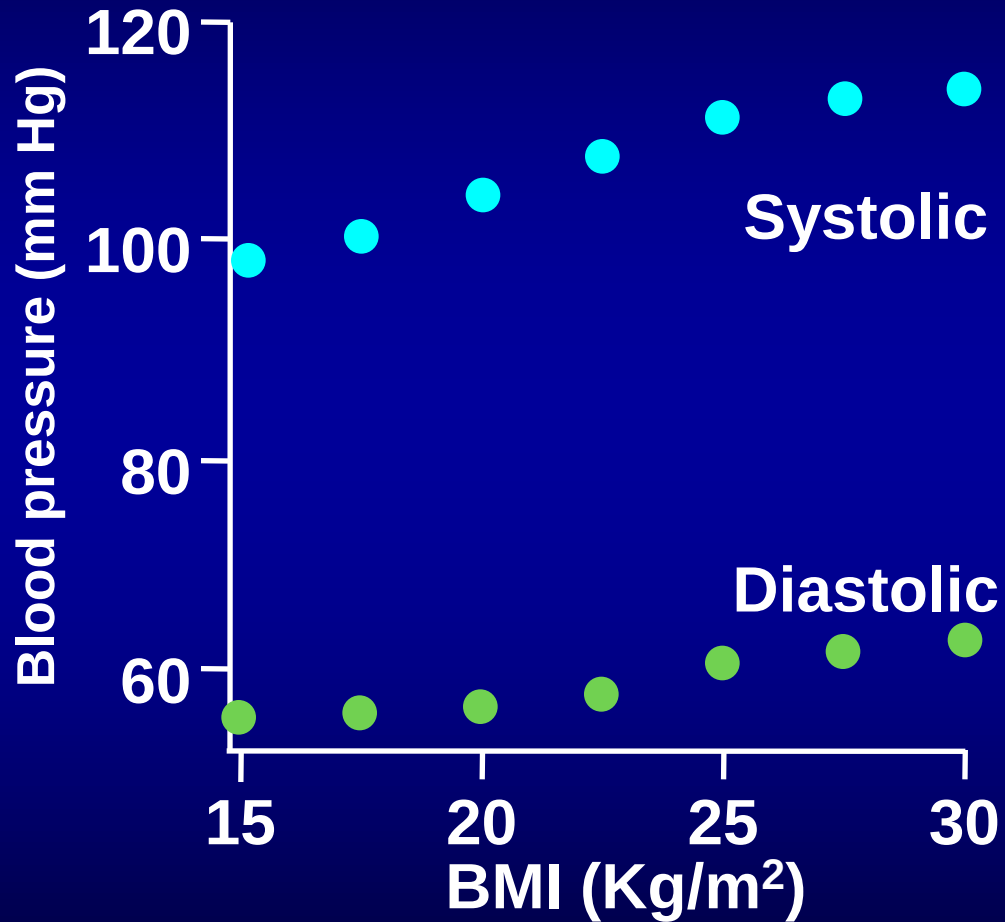
Martin Fischer

ALSPAC: Weight & Haemodynamic Changes

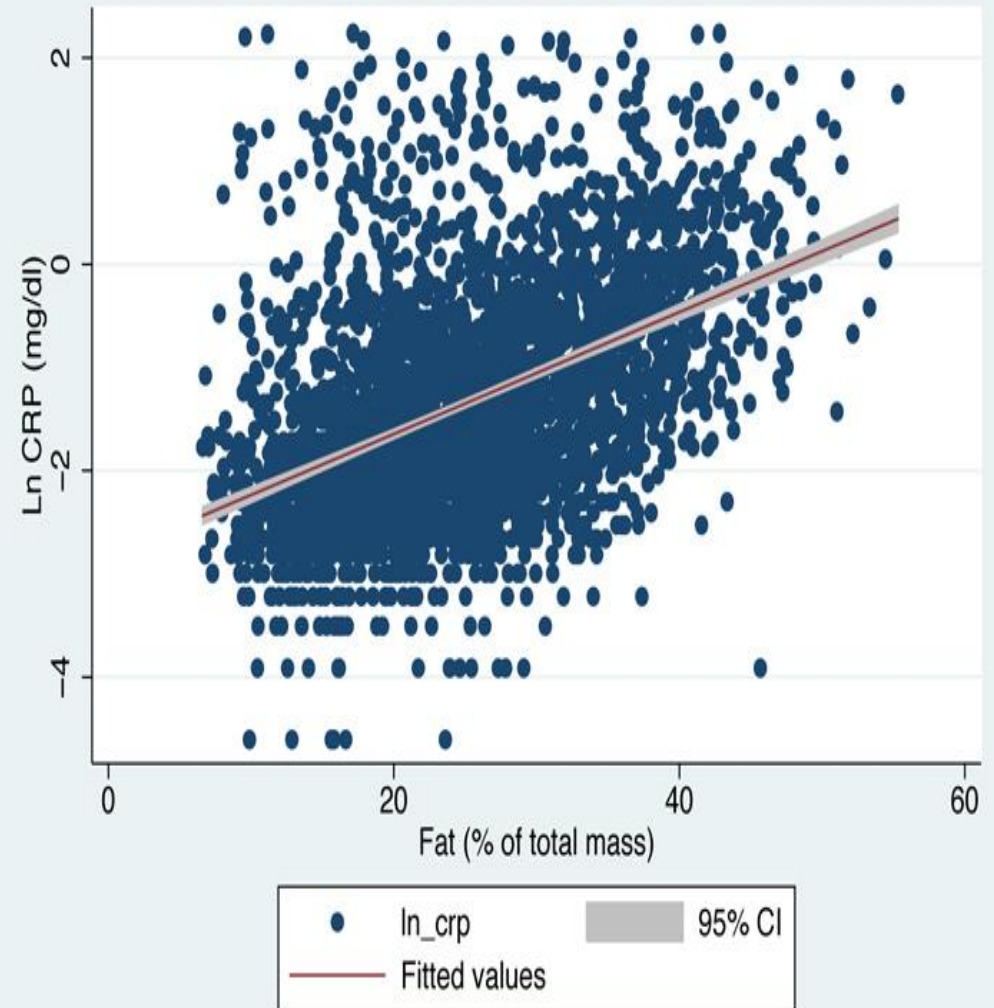
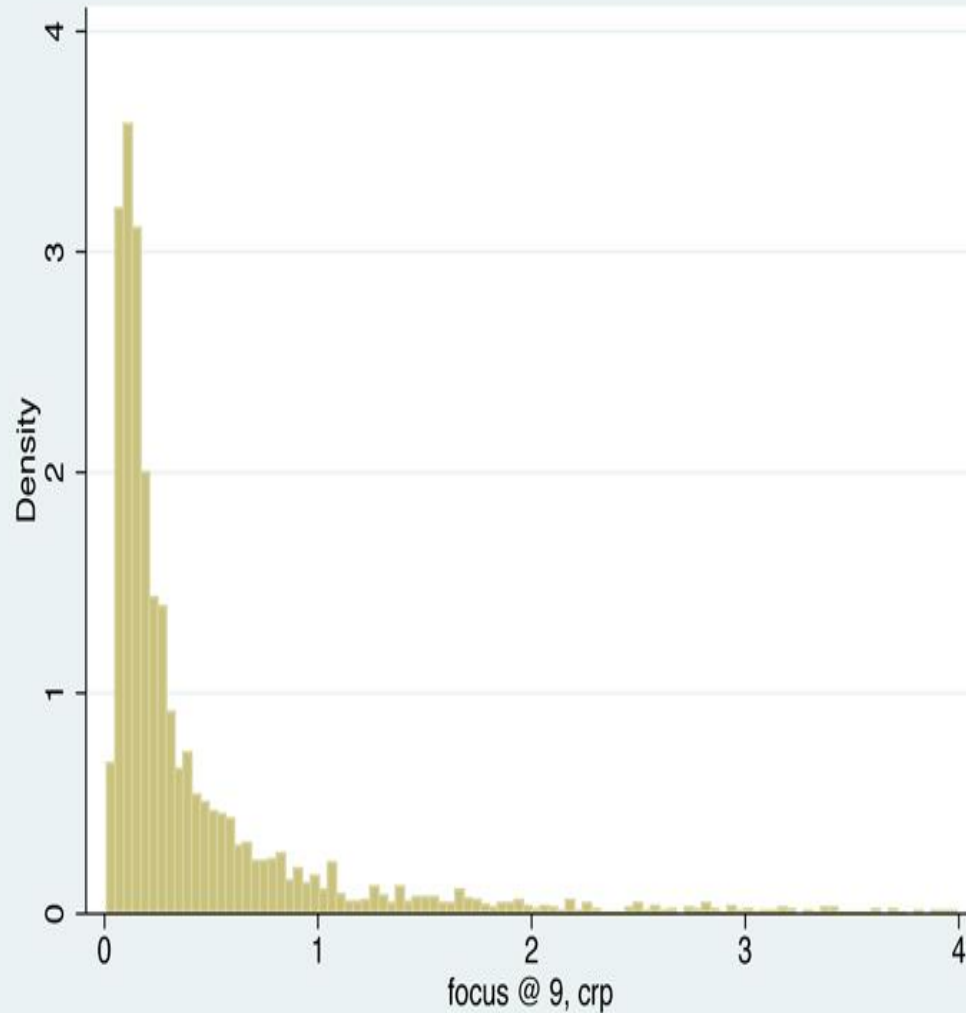
	lean	overweight	obese	P value
Heart rate	72(11)	73(11)	75(12)	<0.001
b.diameter	2.6(0.4)	2.8(0.4)	3.0(0.6)	<0.001
Blood flow	60.4(24.1)	71.7(28.4)	82.0(35.9)	<0.001



ALSPAC: Vascular Risk Factors vs BMI



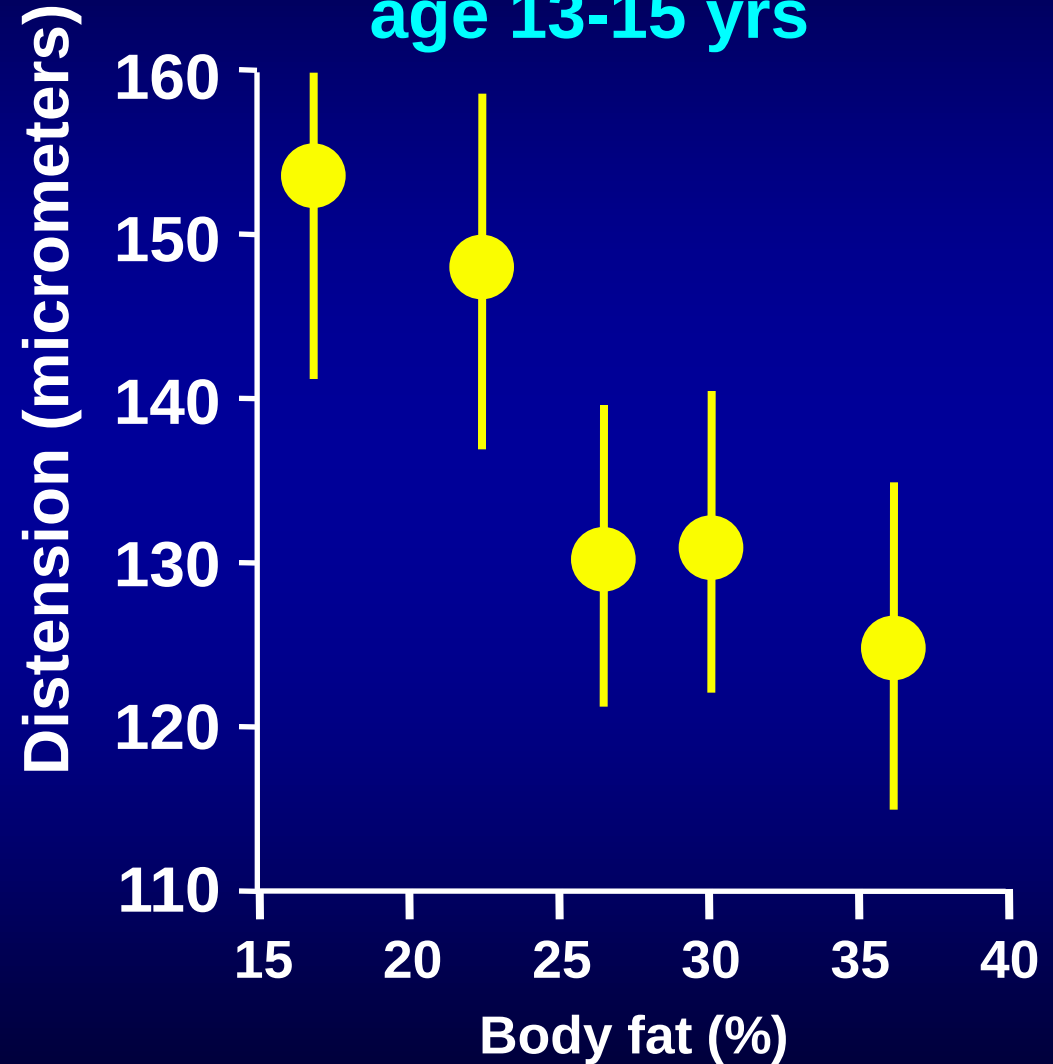
ALSPAC: CRP and Adiposity



Obesity and Arterial Distensibility

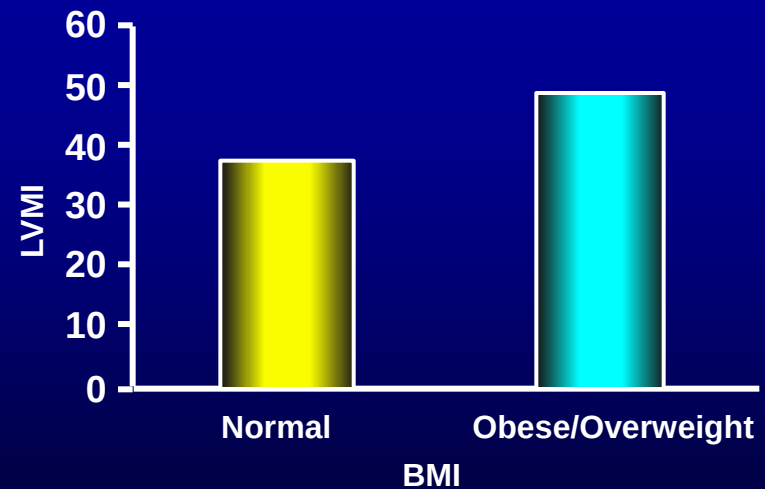
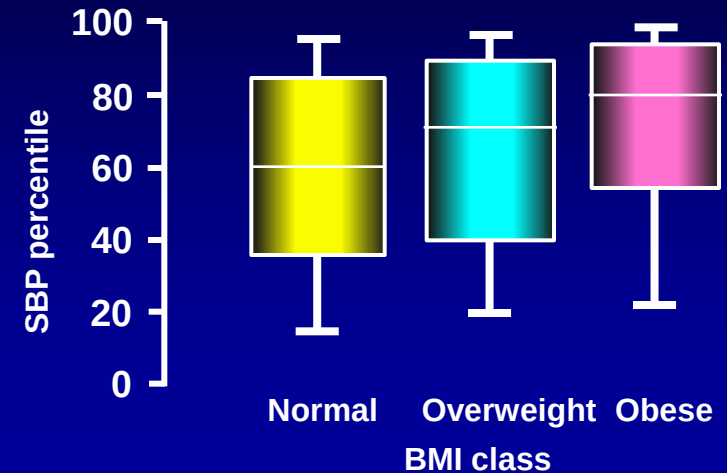


471 children (249 boys, 222 girls)
age 13-15 yrs



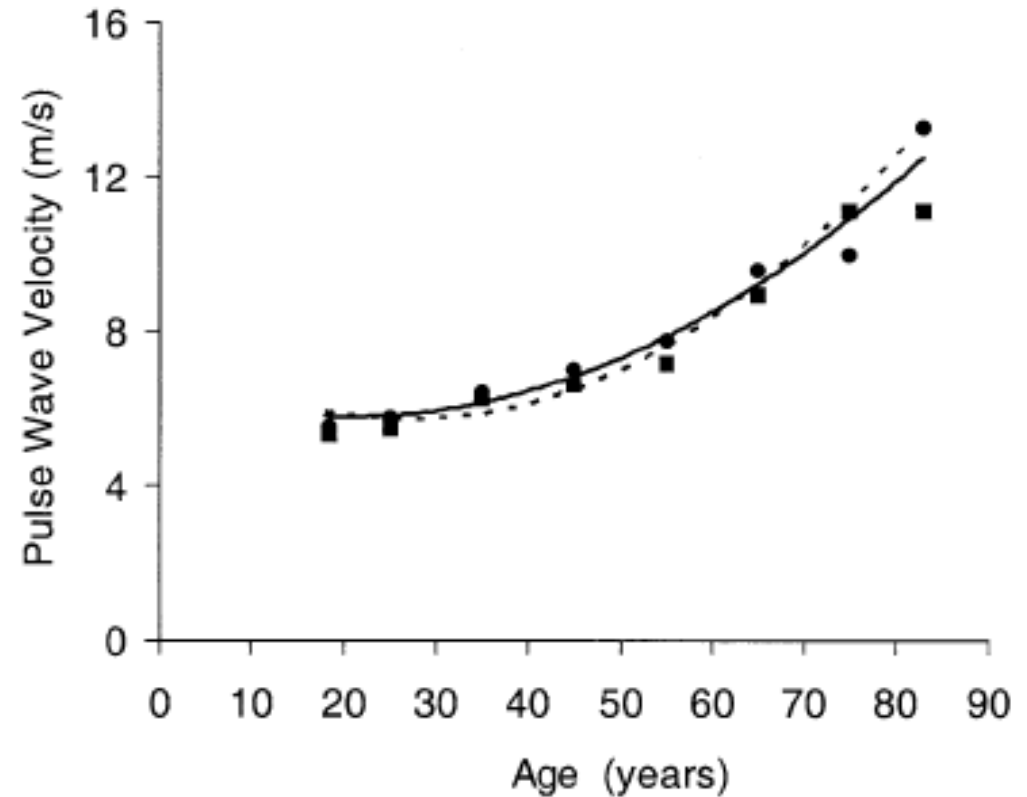
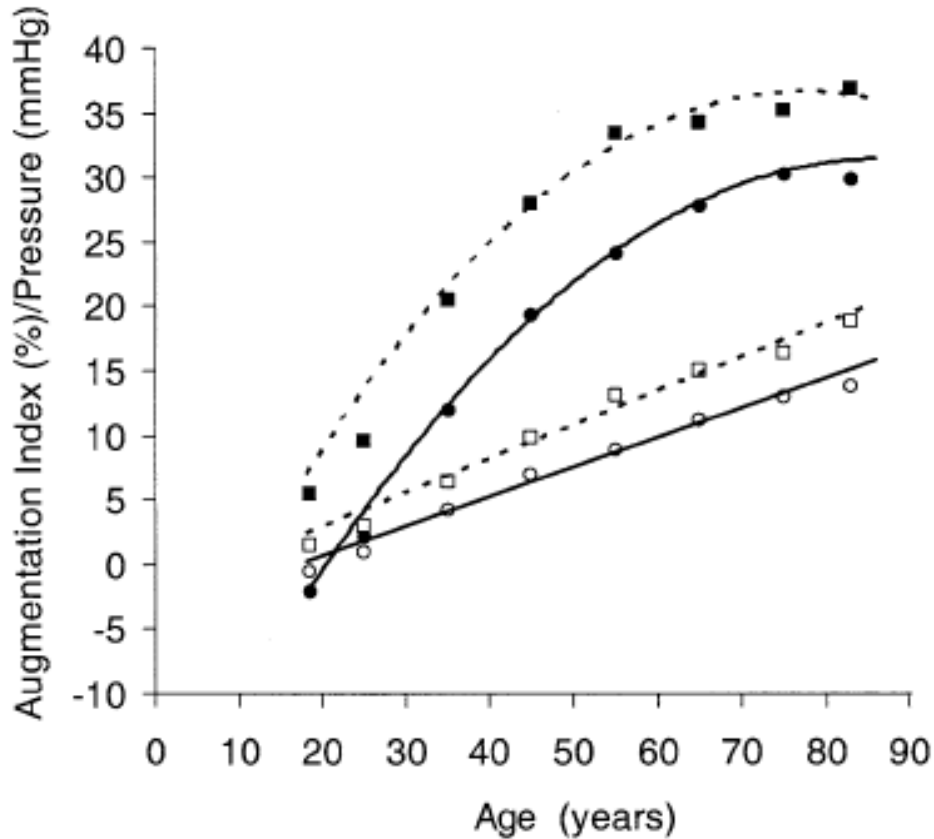
Singhal Circ 2002 106 (15) 1919-24

Cardiovascular Impact of Obesity in ACHD



Cohen Eur J Paediatr 2012; 171: 1145-1150

Ageing and Systemic Vasculature



Systemic arteries stiffen with age from as early as teenage years

Arterial Structure and Function After Recovery From the Metabolic Syndrome

The Cardiovascular Risk in Young Finns Study

Juha Koskinen, BM; Costan G. Magnussen, BHM; Leena Taittonen, MD, PhD; Leena Räsänen, PhD; Vera Mikkilä, PhD; Tomi Laitinen, MD, PhD; Tapani Rönnemaa, MD, PhD; Mika Kähönen, MD, PhD; Jorma S.A. Viikari, MD, PhD; Olli T. Raitakari, MD, PhD; Markus Juonala, MD, PhD

Conclusion—Recovery from the MetS was associated with positive effects on vascular properties during a 6-year follow-up period of young adults. (*Circulation*. 2010;121:392-400.)

**“A man is as old
as his arteries”**



Dr Thomas Sydenham (1624-89)

Emerging Threats for Complex GUCH

- **Ageing**
- **Diabetes**
- **Hypertension**
- **Lung Disease**
- **CAD**

