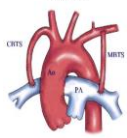


An Introduction to Adult Congenital Heart Disease

Dr Louise Coats
ACHD Consultant
Freeman Hospital, Newcastle upon Tyne Hospitals NHS Foundation Trust



Switching Arteries
Sidetracks Blood and
Oxygen to Otherwise
Starved Lungs

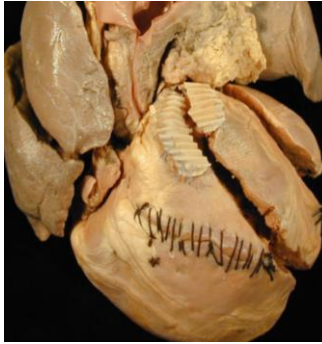


The "Blue" Infant's Blood Lacks What
Should Be in the Lung It Bypasses
By Sidetracking the Artery of the Arm
To the Lung, the Heart and the
Circulation is Restored

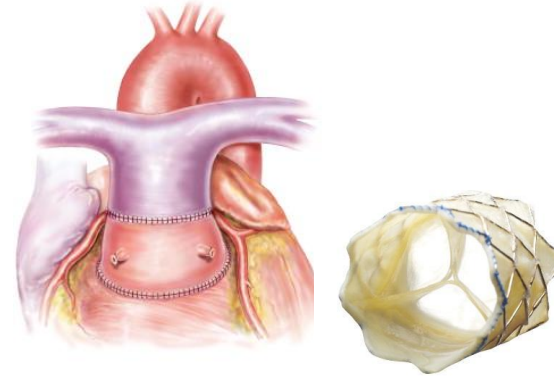
By Robert D. Potter
Medical Editor
MEDICAL progress means
greater safety and longer
life, and the child of our day
the world's great progress have
found in heart, more than any other
since history's earliest doctors



**Blalock
Taussig Shunt**
1944



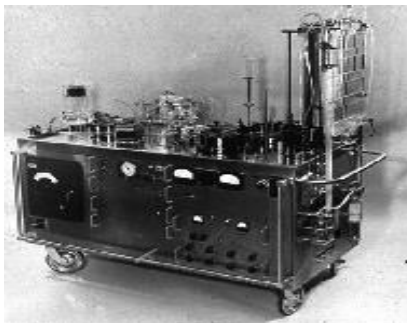
TOF Repair
Lillehei
1954



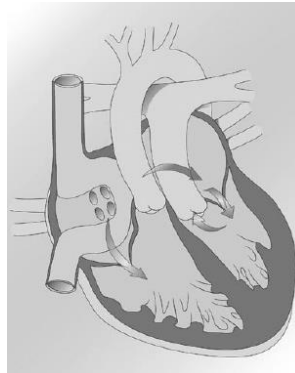
**Arterial Switch
(TGA)**
1976

TPV
2007

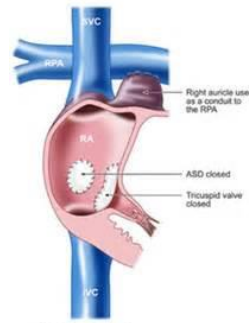
1953
**Cardiopulmonary
Bypass**



1959-64
Mustard/Senning (TGA)

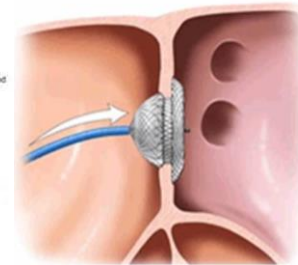


1971
Fontan Repair

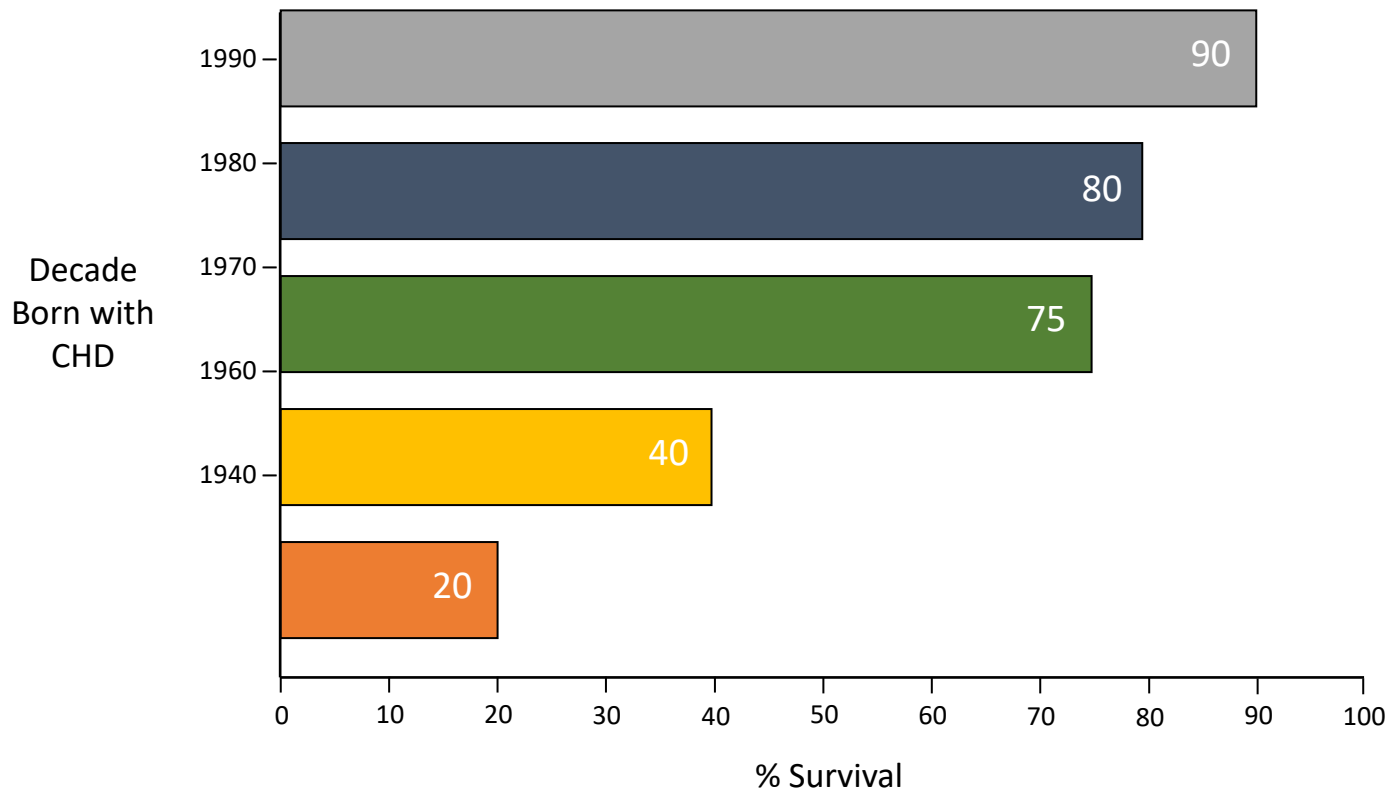


A. Classical Fontan

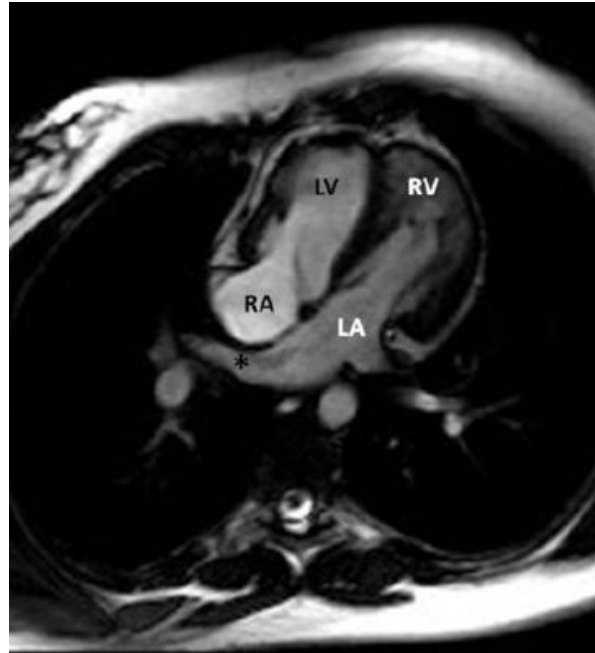
1989
ASD Device



Survival to 18 years of age with CHD



Sometimes diagnosed for first time in adult life.....



Increasing burden of disease

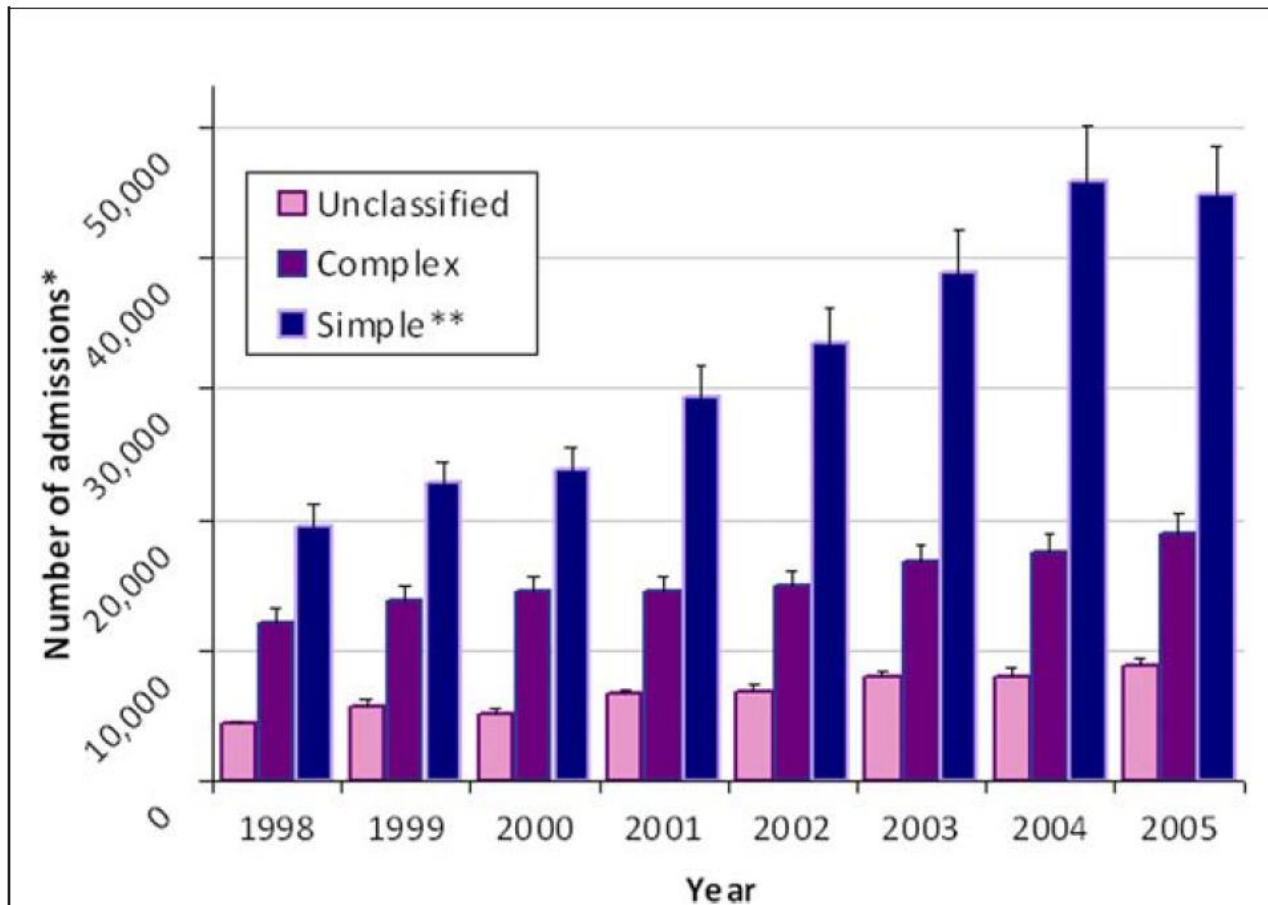


Figure 1

Annual Number of ACHD Admissions in the U.S. Categorized by Level of Defect Complexity

The Severity of ACHD

AP Classification

ANATOMY

I: Simple

Native: small ASD or VSD, mild PS

Repaired: PDA, secundum ASD, sinus venosus defect, VSD

II: Moderate Complexity

Ao-LV fistula, PAPVD, TAPVD, anomalous coronaries, PAVSD, CAVSD, congenital AV or MV disease, coarctation, Ebstein, RVOTO, primum ASD, >moderate PR/PS, branch PS, sinus of Valsava fistula/aneurysm, sub or supra-valvular AS, straddling AV valve

Unrepaired: sinus venosus defect, moderate/large secundum ASD/PDA, VSD with associated anomaly and/or >moderate shunt

Repaired: tetralogy of Fallot

III: Complex

Double-outlet ventricle, interrupted aortic arch, pulmonary atresia, transposition of great arteries, truncus arteriosus

Single ventricle (including double inlet left ventricle, tricuspid or mitral atresia, hypoplastic left heart), unrepaired or palliated (Fontan)

Cyanotic congenital heart defect (unrepaired or palliated)

Other abnormalities of atrioventricular and ventriculoarterial connection (crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

PHYSIOLOGY

A

NYHA I, No hemodynamic, anatomic or arrhythmic sequelae, Normal exercise capacity, renal/hepatic and pulmonary function

B

NYHA II, Mild hemodynamic sequelae (aortic or ventricular enlargement/dysfunction), Mild valvular disease, Trivial or small shunt, arrhythmia not requiring treatment, Objective cardiac limitation to exercise

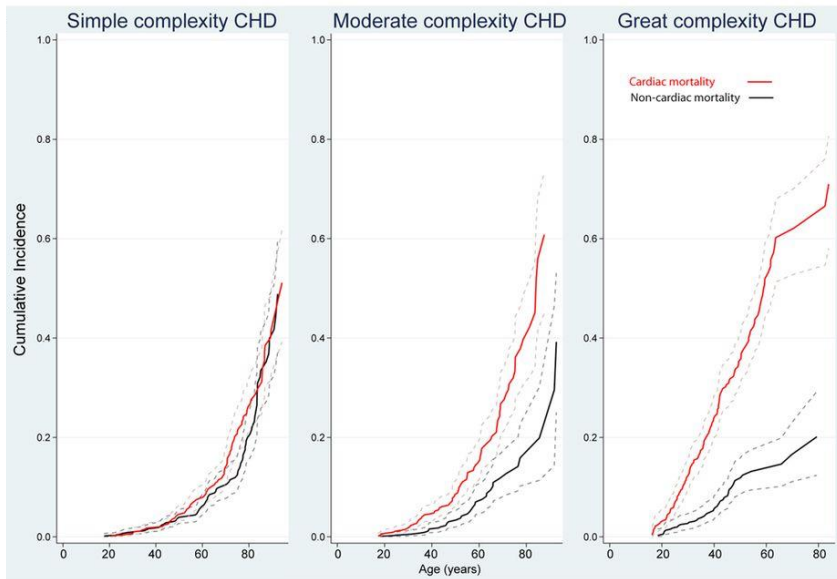
C

NYHA III, Significant valvular disease or ventricular dysfunction, moderate aortic enlargement, venous or arterial stenosis, mild/moderate hypoxemia/cyanosis, significant shunt, arrhythmias controlled with treatment, pulmonary hypertension (less than severe), end-organ dysfunction responsive to therapy

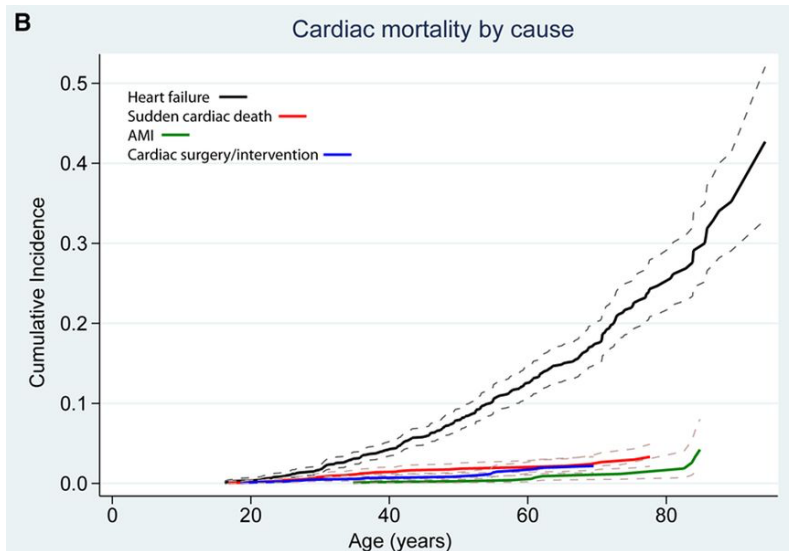
D

NYHA FC IV symptoms, severe aortic enlargement, Arrhythmias refractory to treatment, Severe hypoxemia (almost always associated with cyanosis), Severe pulmonary hypertension, Eisenmenger syndrome, Refractory end-organ dysfunction

Survival Prospects and Circumstances of Death



	Patient's age (years)									
	20	25	30	35	40	45	50	55	60	
ASD	25	26	32	38	42	47	52	57	61	
Valvar disease	29	31	36	40	45	49	54	59	63	
VSD	28	30	36	40	44	49	53	59	63	
Aortic Coarctation	32	33	38	43	47	52	56	62	66	
AVSD	33	34	39	44	48	52	57	62	66	
Marfan syndrome	37	38	42	46	50	54	59	64	68	
Tetralogy of Fallot	37	38	42	47	50	54	60	65	69	
Ebstein anomaly	42	43	47	51	54	59	63	68	72	
Systemic RV	46	48	51	55	59	63	67	72	76	
Eisenmenger syndrome	57	58	62	65	69	73	77	81	84	
Complex CHD	58	59	63	67	70	74	78	82	85	
Fontan	64	65	68	72	75	78	82	86	91	



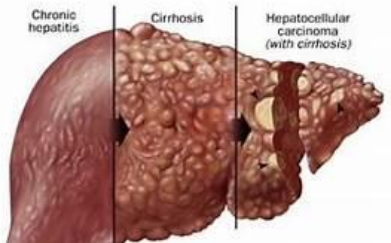
- Low complexity ACHD have a higher burden of CV risk factors and cardiac events ²
- 11-21% SCD <35 years due to ACHD ³

1 Diller et al. Circulation. 2015

2 Saha et al. Circulation 2019

3 Lynge et al. Circulation AE 2018

Extra-cardiac complications in ACHD



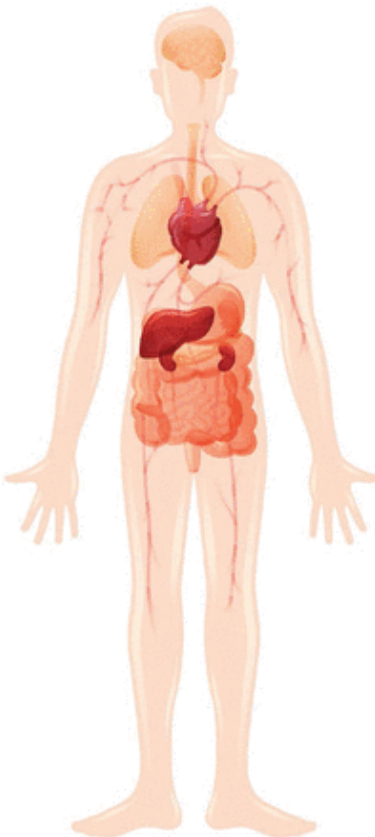
LUNG
 Restrictive lung disease
 Pulmonary hypertension
 Pulmonary hemorrhage
 Plastic bronchitis

LIVER
 Congestive hepatopathy
 Cardiac cirrhosis
 Fontan associated liver disease

**IMMUNOLOGY/
 INFECTIOUS DISEASE**
 Protein-losing enteropathy
 Infective endocarditis
 Pneumonia
 Brain abscess

HEMATOLOGY
 Secondary erythrocytosis/iron deficiency/hyperuricemia (Cyanotic CHD)
 Thromboembolism
 Anemia

ONCOLOGY
 Low-dose ionizing radiation and malignancy
 Hepatocellular carcinoma
 Age-appropriate cancer screening



PSYCHOSOCIAL
 Depression
 Anxiety
 Neurodevelopment deficits

ENDOCRINE
 Thyroid
 Calcium hemostasis/Bone health
 Obesity/Metabolic syndrome
 Diabetes
 Dyslipidemia

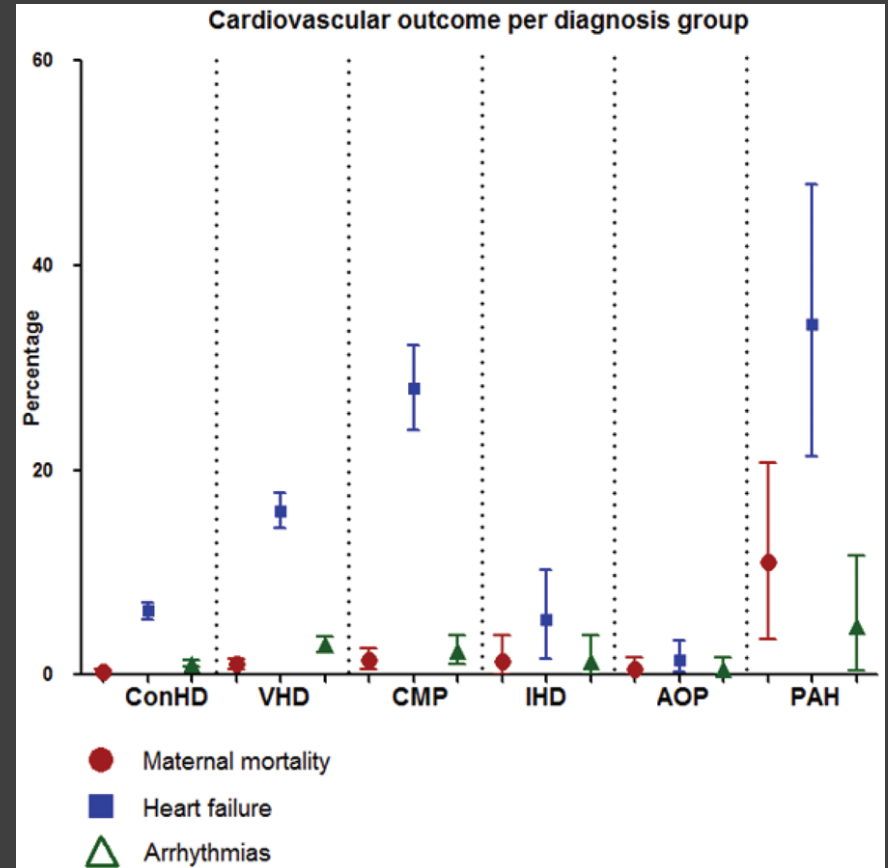
RENAL
 Chronic kidney disease
 Cardiorenal syndrome

VASCULAR
 Cerebrovascular disease
 Peripheral venous/arterial disease
 Aortopathy
 Endothelial dysfunction
 Hypertension



Pregnancy in women with congenital heart disease

- Majority tolerate pregnancy well
- Specialist care best provided by MDT
 - ACHD/heart failure cardiology, obstetrics
 - Haematology, neonatology, anaesthesia and genetics
 - Timely counselling essential
- Team should be involved early in pregnancy in order to plan antenatal care, including delivery and post-partum follow-up

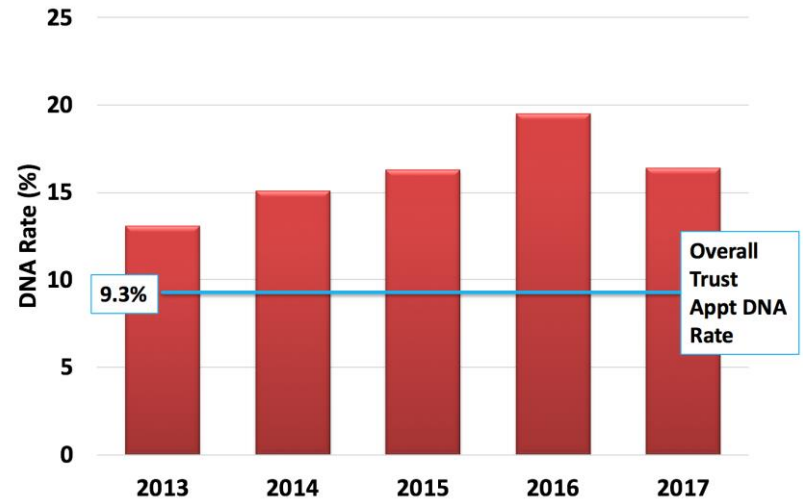
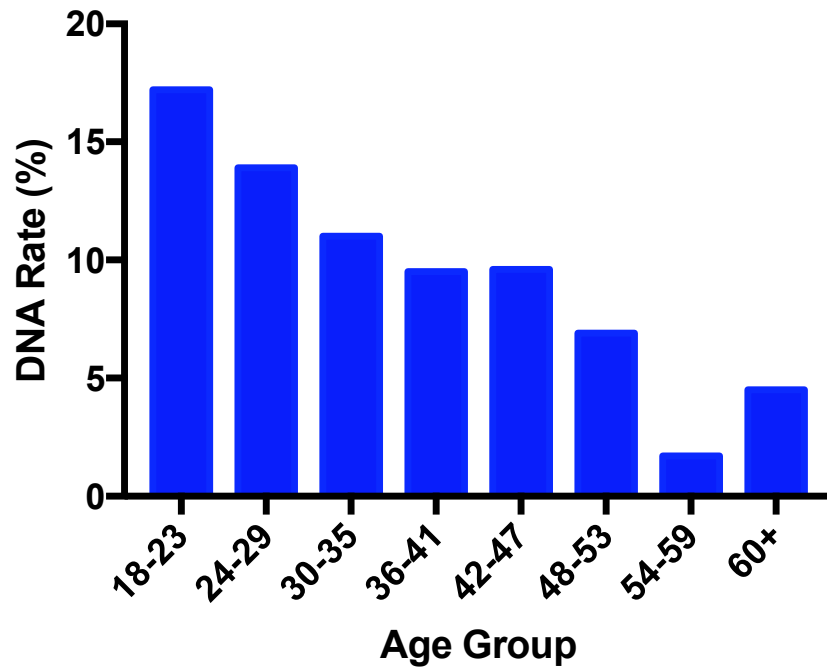


Lesion specific estimates

WHO 1	WHO II	WHO III	WHO IV
<p>Uncomplicated, small or mild PS, VSD, PDA</p> <p>MVP with no more than trivial MR</p> <p>Successfully repaired simple lesions, e.g. ASD, VSD, PDA, APVR</p> <p>Isolated ectopic beats</p>	<p>Unoperated ASD/VSD (if otherwise well and uncomplicated)</p> <p>Repaired ToF</p> <p>Most arrhythmias</p> <p>Mild LV impairment</p> <p>HCM</p> <p>Native or tissue valvular heart disease not considered WHO I or IV</p> <p>Marfan without root dilatation Aorta <45 mm associated with BAV Repaired coarctation</p>	<p>Mechanical valve</p> <p>Systemic RV</p> <p>Fontan</p> <p>Cyanotic heart disease</p> <p>Other complex congenital lesions</p> <p>Aortic dilatation 40–45 mm in Marfan 45–50 mm in aortic disease associated with bicuspid aortic valve</p>	<p>PAH of any cause</p> <p>Severe systemic LV dysfunction (LVEF<30%)</p> <p>Previous PPCMP with residual LV dysfunction</p> <p>Severe symptomatic AS, severe MS Native severe coarctation</p> <p>Marfan and root > 45 mm</p> <p>Fontan with complications</p>
	<p>These women need expert joint cardiac and obstetric preconception counselling, and care throughout the antenatal and peripartum period</p>		<p>Early contraception advice should be provided</p> <p>Termination should be discussed if pregnancy occurs</p>

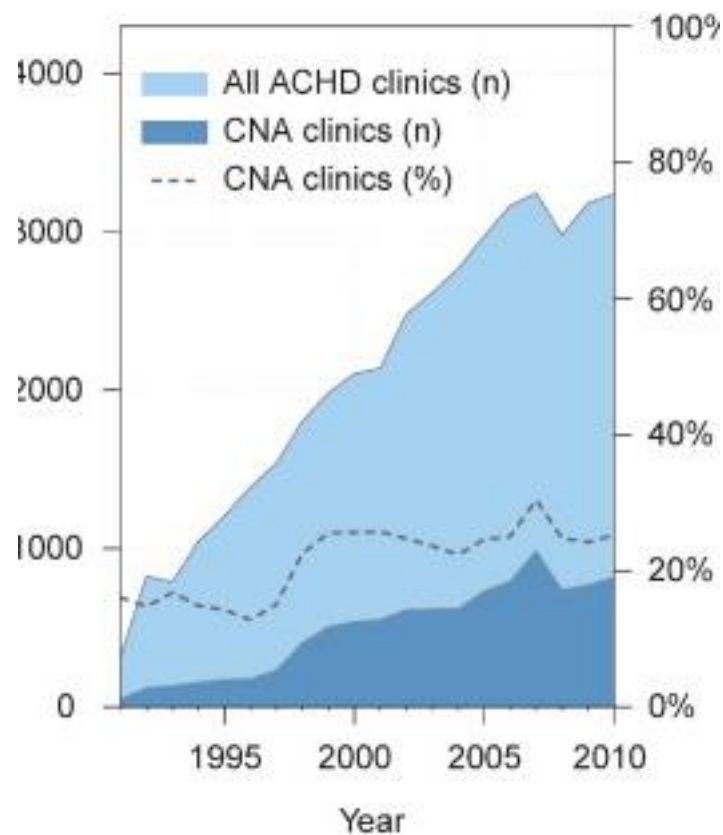
Non-Attendance

NUTH Data



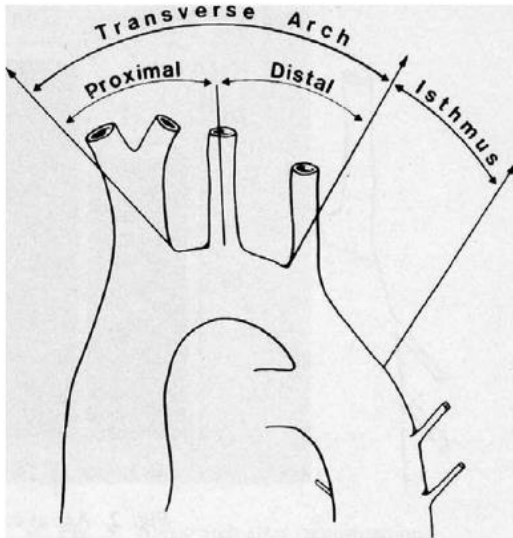
ACHD non-attendance associated with higher mortality

- **Risk factors**
- non-white ethnicity
- previously missed clinic appointments
- lower socioeconomic status

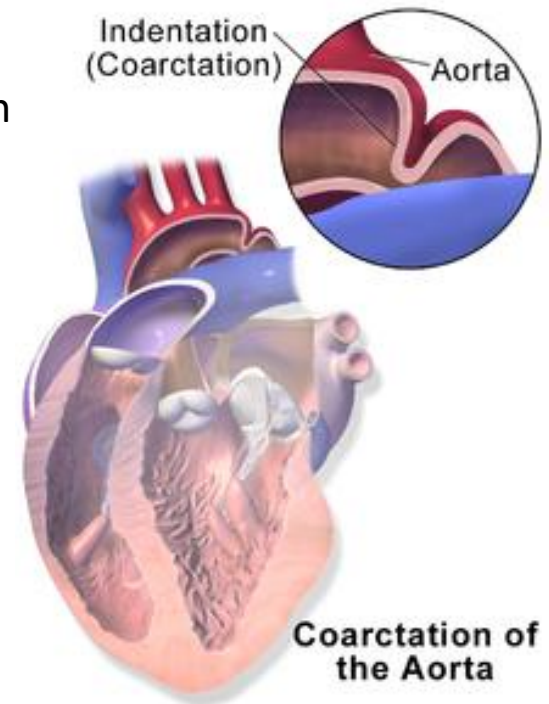


Coarctation of the Aorta

“constricted aortic segment comprising localized medial thickening, with some infolding of medial and neointimal tissue”



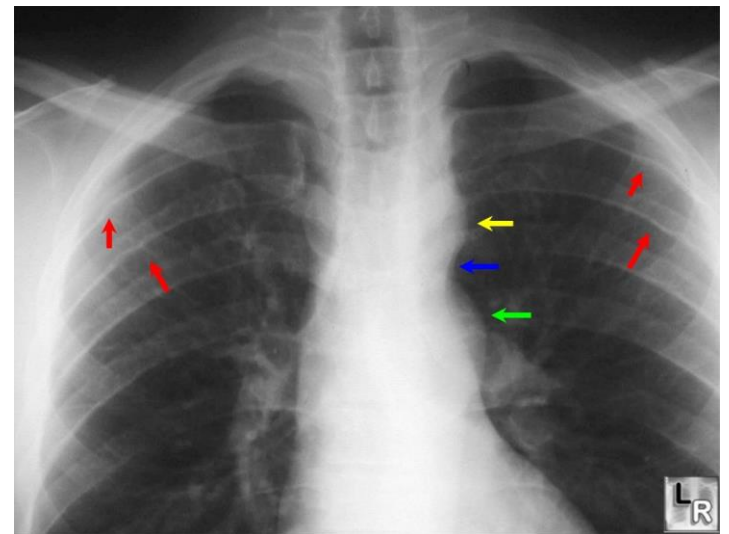
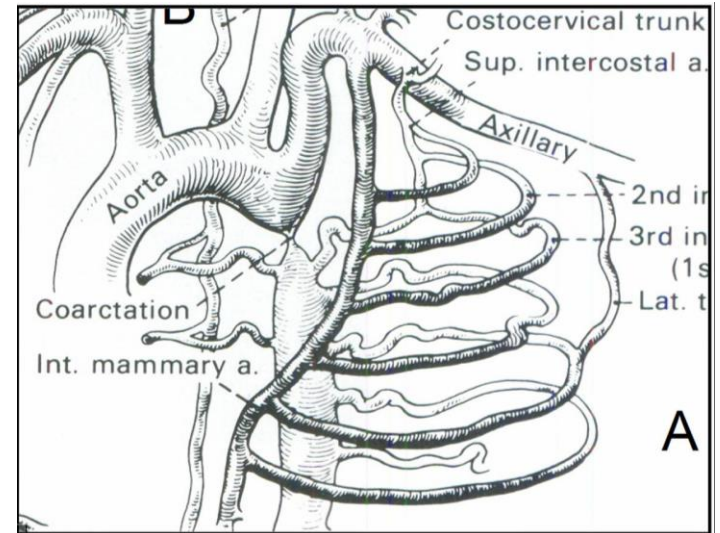
- Juxtaductal
- Just distal to left subclavian
- Shelf like or membranous
- Discrete or long segment
- Heterogeneous



Part of a generalized arteriopathy

Coarctation of the Aorta

- 5-8% CHD (30% simple, 30%VSD, 40% complex)
- Bimodal presentation
 - Early life: CHF
 - Late life: \uparrow BP, murmur, headache, epistaxis, ICH
- BAV (50%)
- Turners (X0) 15-20%
- Association with berry aneurysms (10%, \uparrow age)



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

Options for treatment

1. Percutaneous

a) Balloon Angioplasty

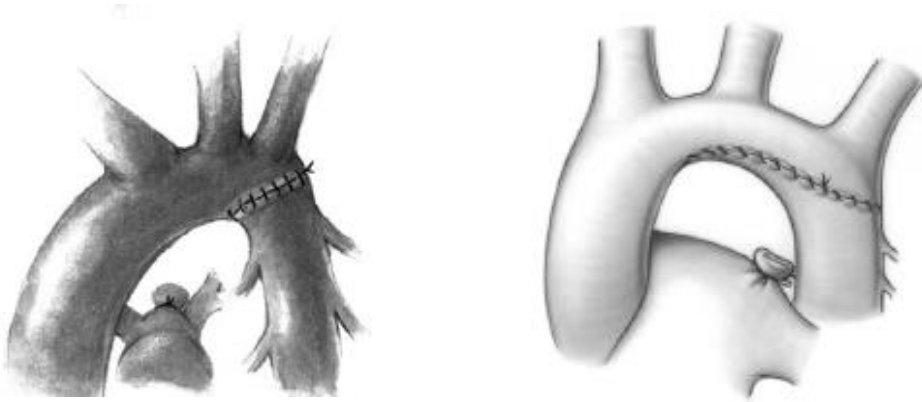
b) Stent

Treatment of choice in adult native CoA with appropriate anatomy

2. Surgical

Indications for Intervention

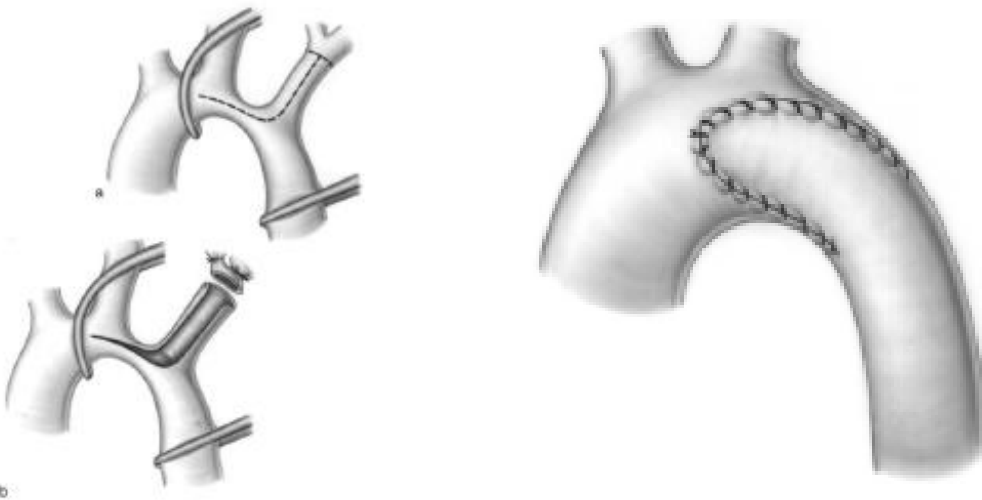
End to End Anastomosis



Pro: excise ductal tissue,
hypoplastic segment

Cons: circumferential scar,
anastomosis tension

Reverse Subclavian Flap Technique



Pro: Tension free
anastomosis, less dissection,
growth, partial scar

Cons: residual ductal tissue,
arch hypoplasia, limb growth

Natural history of coarctation of the aorta

Maurice Campbell

From the Cardiac Department, Guy's Hospital, London S.E.1, and the Institute of Cardiology, London W.1

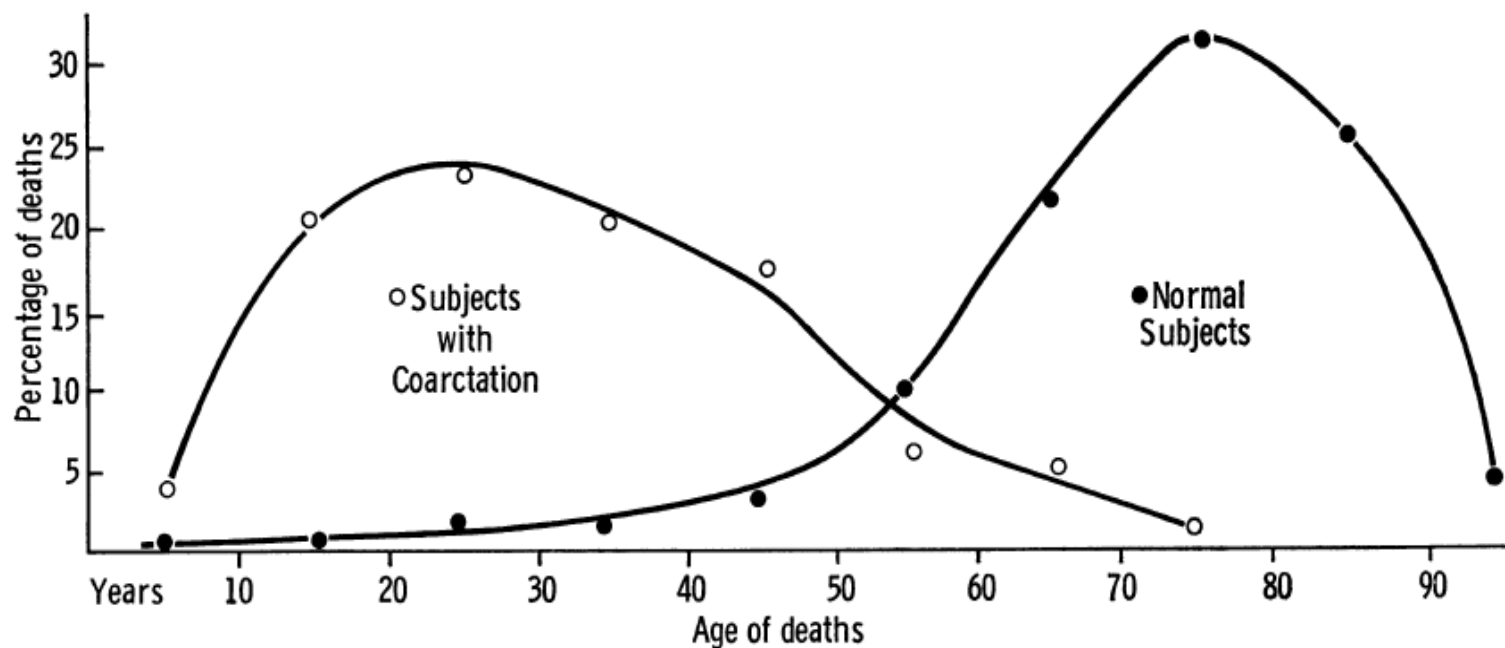
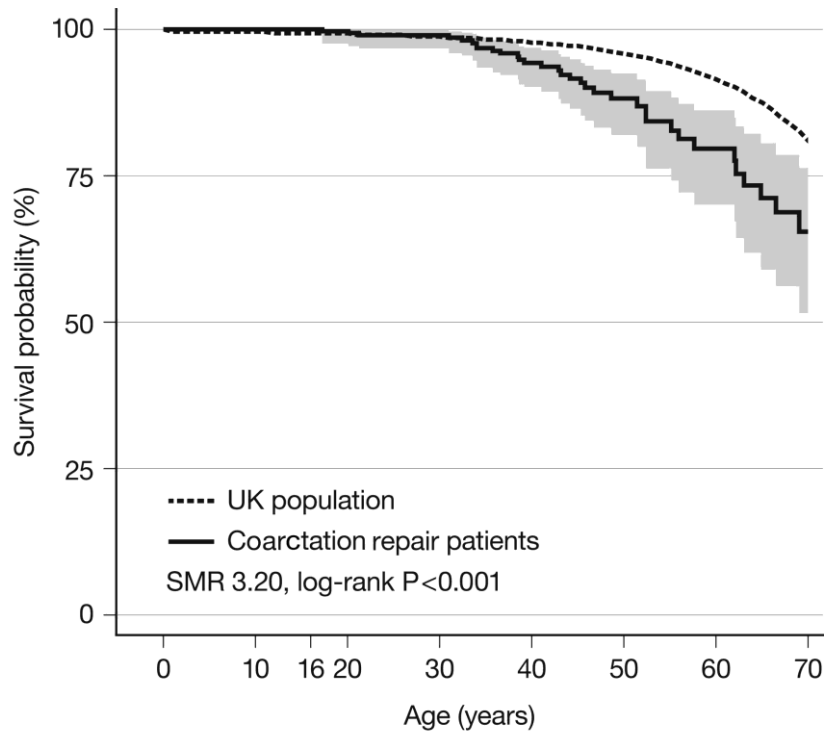
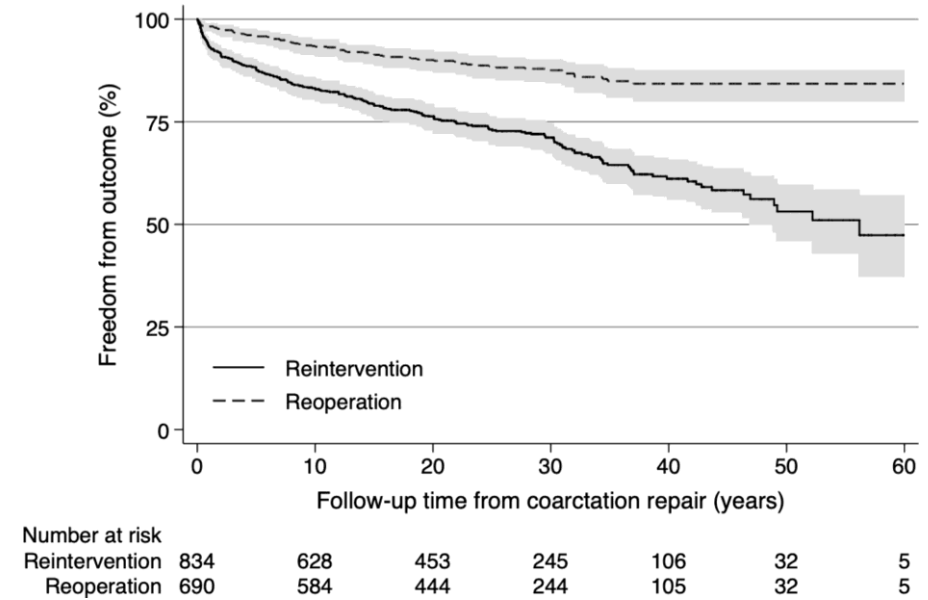


FIG. 1 *The distribution of deaths by age, excluding deaths in the first year of life. In coarctation on the left and in normal subjects on the right, there is relatively little overlapping. For coarctation, the more rounded curve from reported necropsies is shown but the flatter curve from my calculations from deaths of patients under observation is similar (see Fig. 2).*

Long-term mortality and cardiovascular burden for adult survivors of coarctation of the aorta

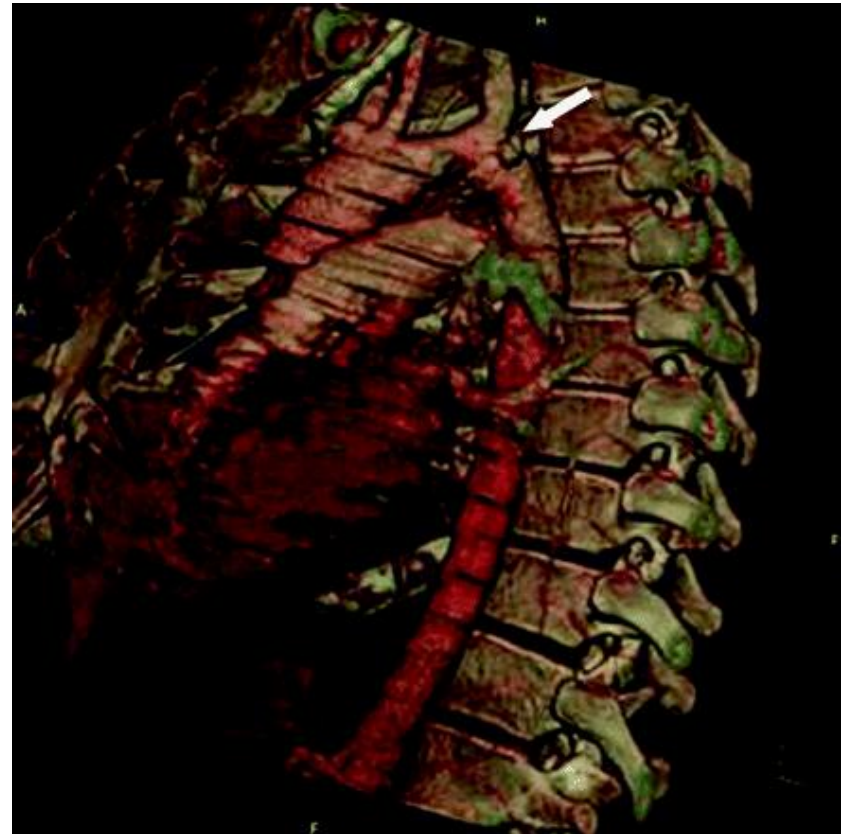


Number at risk	0	0	297	286	236	161	80	42	18
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Survivors of repair: fixed not cured

- Recoarctation
- Late aneurysm (rupture)
- **Hypertension**
- Accelerated coronary disease
- Premature cerebrovascular disease
- High incidence of BAV → AVR



Require long term, interval 3D imaging to screen for aneurysm or re-coarctation

The Pregnant Mum with Coarctation

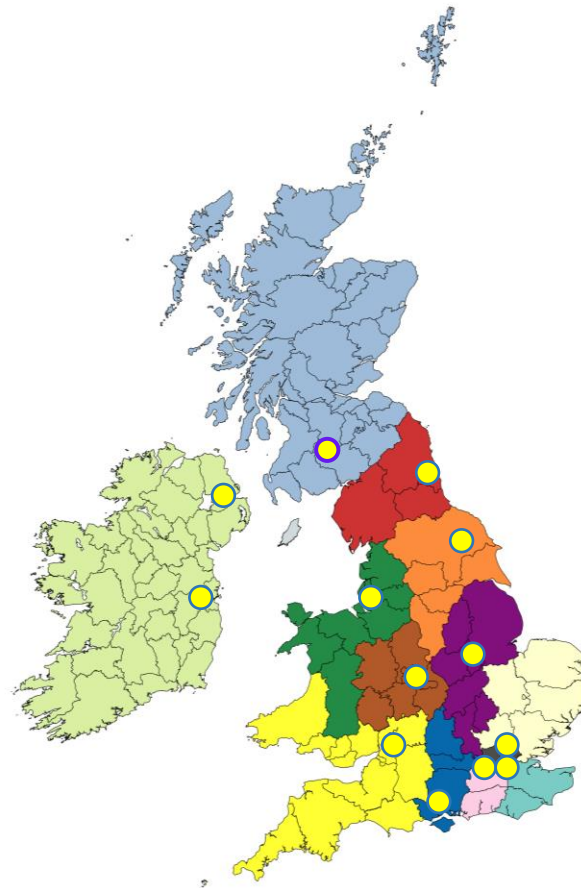
- Higher miscarriage rate
- Placental insufficiency
- Higher rates of hypertensive disorders (x4)
- Higher rate of LSCS (Limit the second stage)



The Wider Network



The CHD Operational Delivery Networks



Thank You



NENC-CHDN

North East and North Cumbria Congenital Heart Disease Network

@NENC_CHDN (Twitter)

@NENC.CHDN (Facebook)