

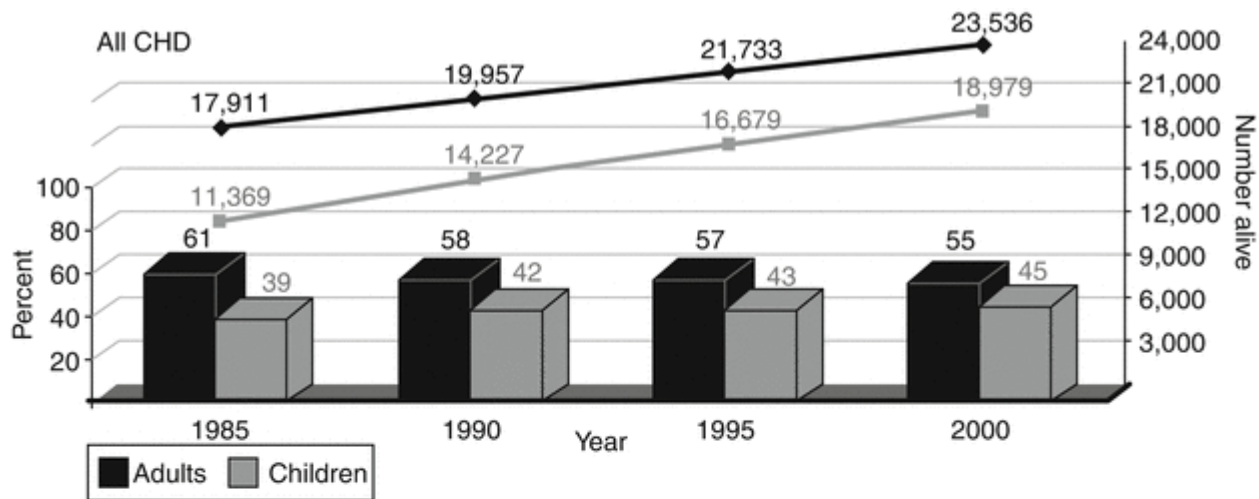
Adult Congenital Heart Disease An Overview

Sheena Vernon MSc
Lead Nurse CHD Network
Bristol Heart Institute

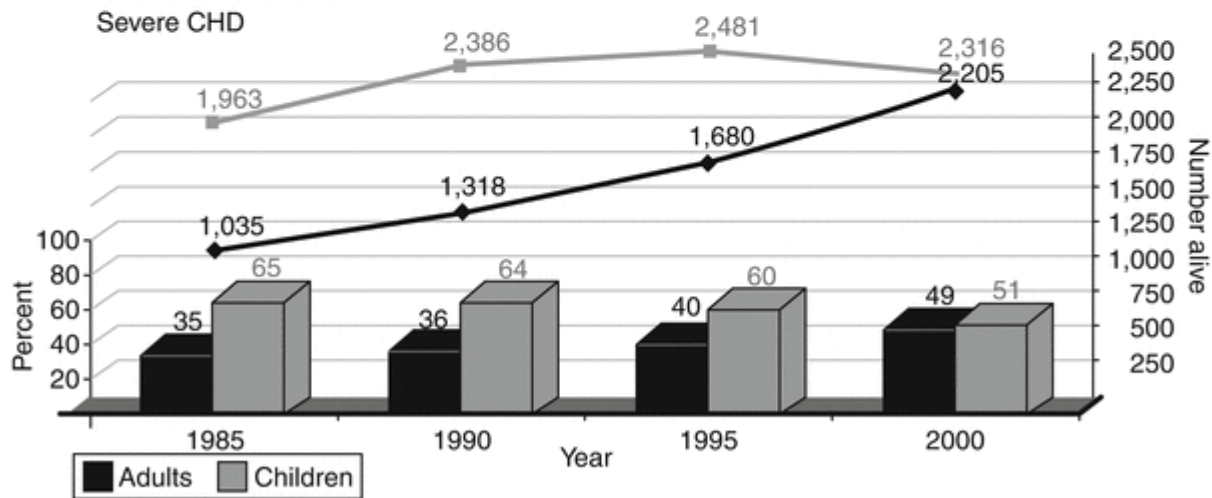
2020

POPULATION

- Incidence: 8 per 1000 live births.
- 40 yrs. ago mortality from untreated CHD was 60%- 70% over the age of 18 years.
- Success of cardiac surgery and cardiology in infancy improved life expectancy.
- 85% of CHD patients, including complex, rare and severe conditions will reach adulthood.
- **More** adults than children with CHD.



a



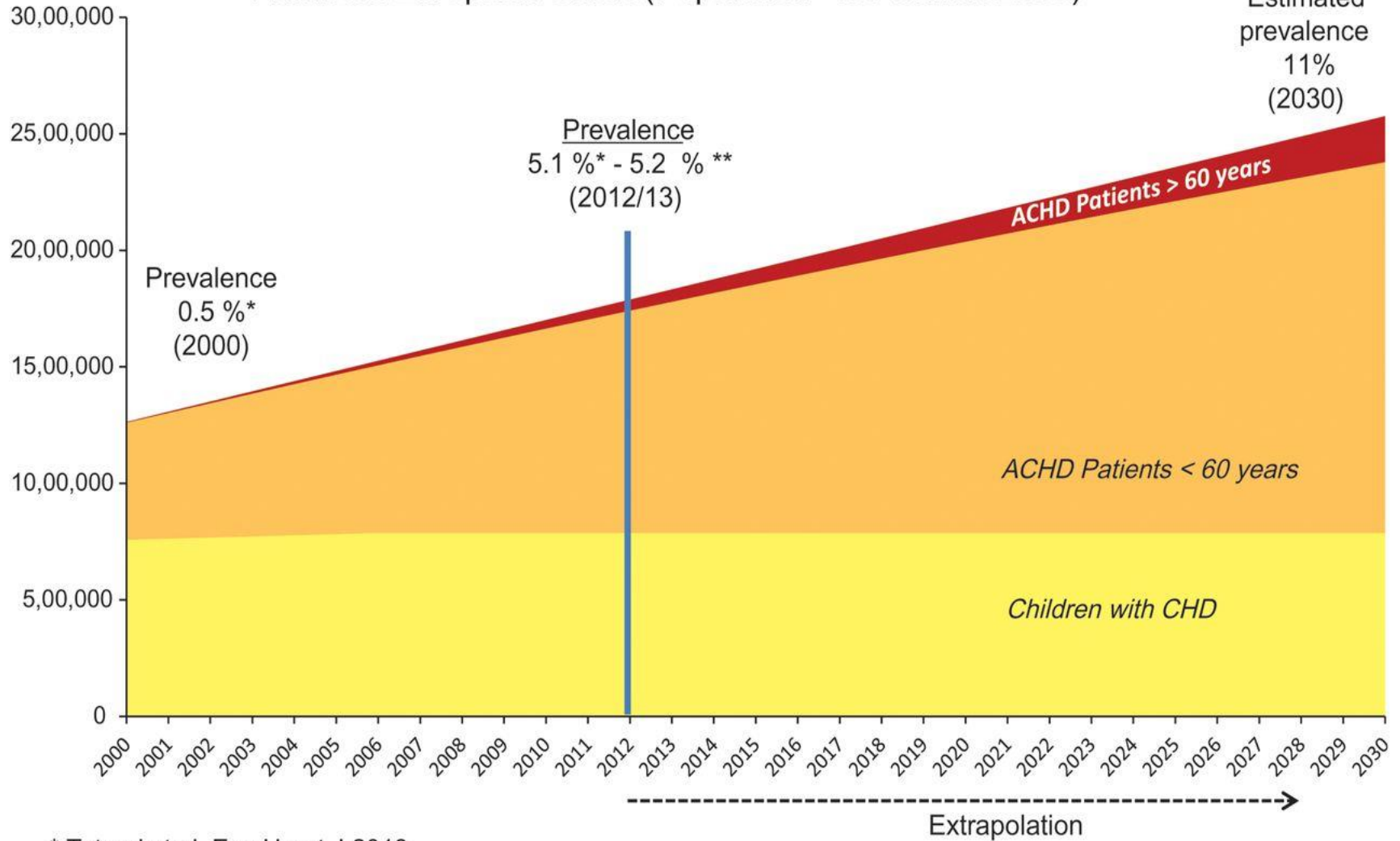
b

CHD = Congenital heart disease

From Marelli et al, J Amer Coll Card 2007

Numbers and proportion of adults and children with all CHD (a) and severe CHD (b) in 1985, 1990, and 2000 (From Marelli et al. (2007) J Am Coll Card)

Numbers European Union (Population 497 Mill. in 2008)



* Tutarel et al. Eur. Heart J 2013

** German Competence Network for Congenital Heart Disease (data on file)

Progress in Late Results Among Pediatric Cardiac Surgery Patients

A Population-Based 6-Decade Study With 98% Follow-Up

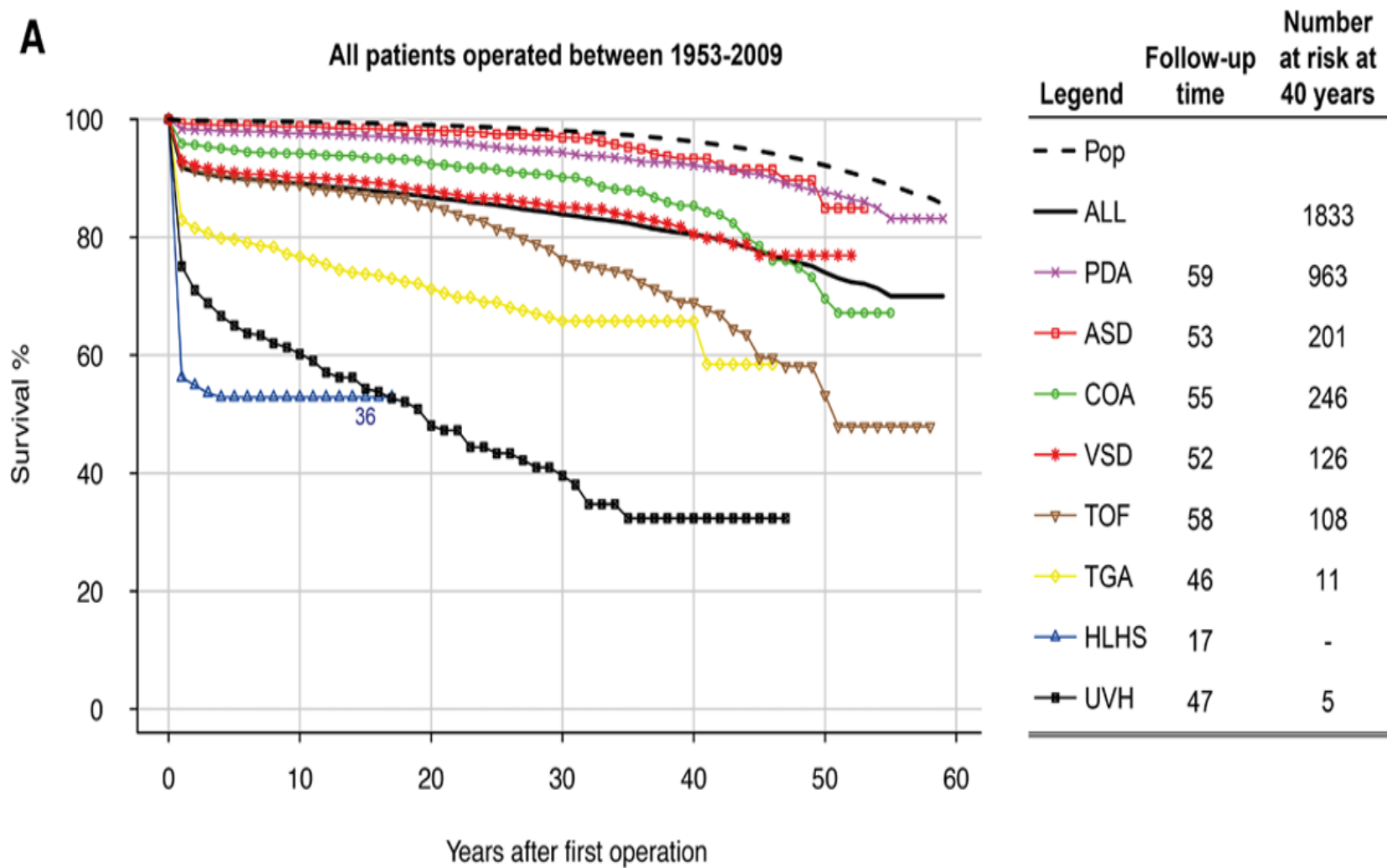
Alireza Raissadati, MD; Heta Nieminen, MD, PhD; Eero Jokinen, MD, PhD;
Heikki Sairanen, MD, PhD

Circulation January 27, 2015

Raissadati et al Late Results After Pediatric Cardiac Surgery

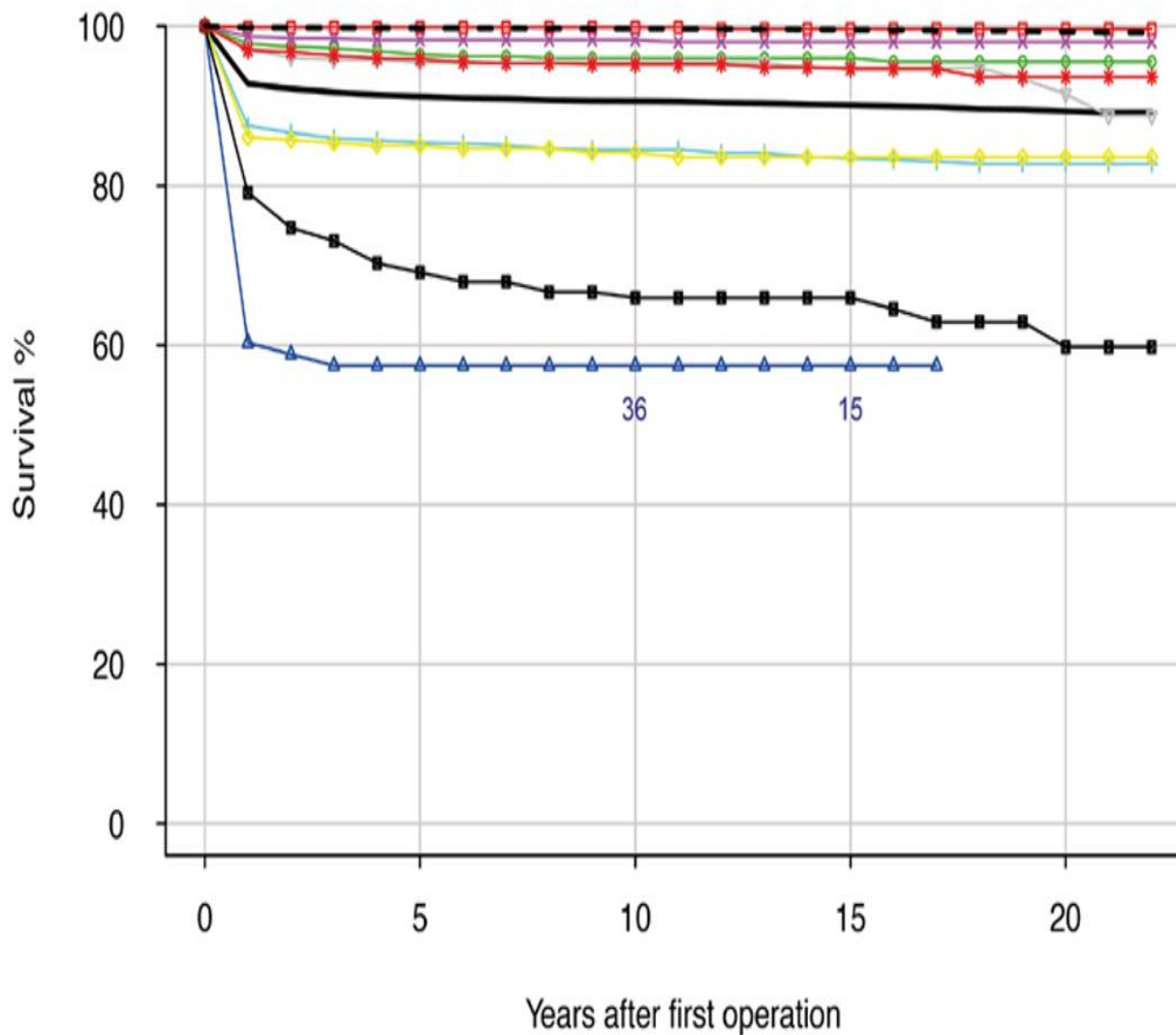
A

All patients operated between 1953-2009



B

All patients operated between 1990-2009



Legend	Follow-up time	Number at risk at 20 years
--- Pop		
— ALL		712
—× PDA	22	152
—□ ASD	22	140
—○ COA	22	68
—* VSD	22	116
—△ TOF	22	40
—◇ TGA	22	29
—△ HLHS	17	-
—■ UVH	22	15

Circulation January 27, 2015

Relative age

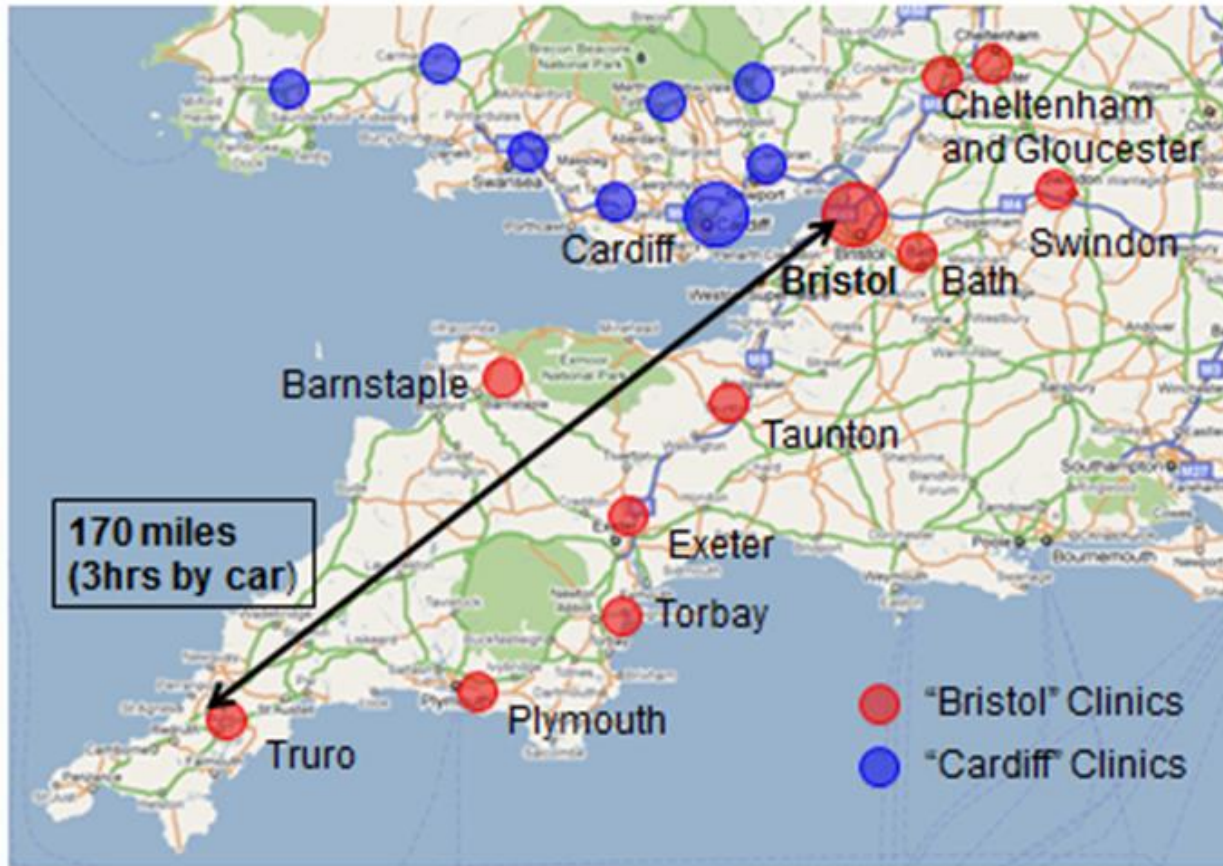
	Patient's age (years)										Age difference:
	20	25	30	35	40	45	50	55	60	65	
ASD	25	26	32	38	42	47	52	57	61	65	>40
Valvar disease	29	31	36	40	45	49	54	59	63	67	30-40
VSD	28	30	36	40	44	49	53	59	63	67	20-30
Aortic Coarctation	32	33	38	43	47	52	56	62	66	70	10-20
AVSD	33	34	39	44	48	52	57	62	66	70	5-10
Marfan syndrome	37	38	42	46	50	54	59	64	68	72	2-5
Tetralogy of Fallot	37	38	42	47	50	54	60	65	69	73	<2
Ebstein anomaly	42	43	47	51	54	59	63	68	72	76	>40
Systemic RV	46	48	51	55	59	63	67	72	76	80	>40
Eisenmenger syndrome	57	58	62	65	69	73	77	81	84	88	>40
Complex CHD	58	59	63	67	70	74	78	82	85	89	>40
Fontan	64	65	68	72	75	78	82	86	91	95	>40

Values present relative age adjusted for predicted 5-years mortality. Colors reflect the difference between relative and actual age. For example a 40 year old Fontan patient has a mortality rate that is comparable to that of a 75 year old individual without CHD.

SWSW POPULATION

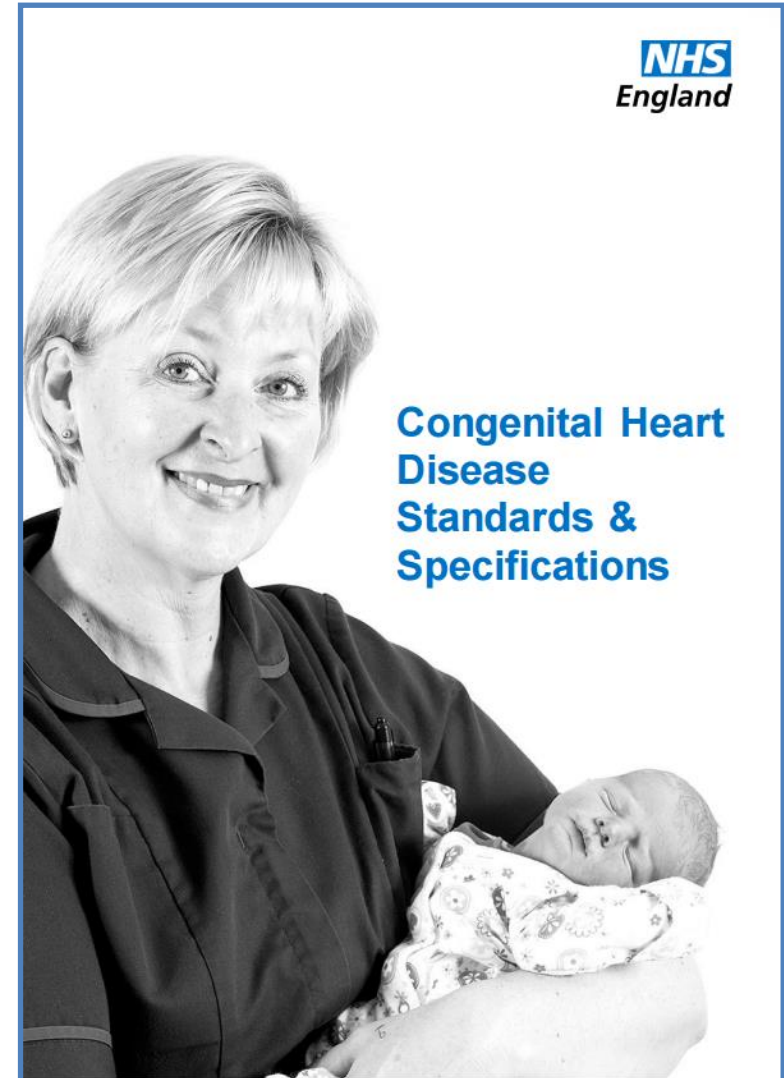
- 8,000 **Adults** South West
- 6,500 **Children** 135,000 adults and young people England
- In 2000 equal numbers of those alive with severe CHD were adults.

SWSW CHD Network



CHD STANDARDS 2016

- Section A: **The network approach**
- Section B: **Staffing and skills**
- Section C: **Facilities**
- Section D: **Interdependencies**
- Section E: **Training and education**
- Section F: **Organisation, audit**
- Section G: **Research**
- Section H: **Communication**
- Section I: **Transition**
- Section J: **Pregnancy contraception**
- Section K: **Fetal diagnosis**
- Section L: **Palliative care and bereavement**





Welcome to the Congenital Heart Disease Network South Wales and South West

We proudly support over 6,500 children and 8,000 adults with a congenital heart condition.

[Read More >](#)

[Babies and Children](#)

[Teenagers/Young](#)

[Adults](#)

Terminology

Grown-up Congenital Hearts
(GUCH)

Adult Congenital Heart
Disease (ACHD)

BRISTOL HEART INSTITUTE



OUTPATIENTS



ADULT CONGENITAL TEAM

- BHI Cardiologists x 5, Surgeons x 3
- Specialist registrar 3/4
- CNS x 3.6 (5)
- Cardiac obstetrics
- Consultant Radiologists TTE, TOE, CMRI, CT
- Anaesthetist CHD interest
- Level 2 Cardiff and South Wales team
- Level 3 clinics in 7 D.G.H's + Wales
- Barnstable, Cheltenham, Swindon, Taunton, Exeter, Torbay, Truro

Role of ACHD CNS? 342 26599

- In-patient and out patient issues
- Pre-assessment clinics
- Surgery, cardiology, medical admissions, arrhythmias, endocarditis, heart failure
- Learning disability work
- Pregnancy/contraception
- Teenage and young adult clinic
- End of life care
- Pulmonary hypertension
- Telephone Advice >3000 calls pa
- Write patient information
- Education to pts and staff



Adult congenital heart disease nursing

RCN guidance on roles, career pathways and competence development

To support adult
standards

Guidelines from
RCN for nursing
published

PREDISPOSING FACTORS

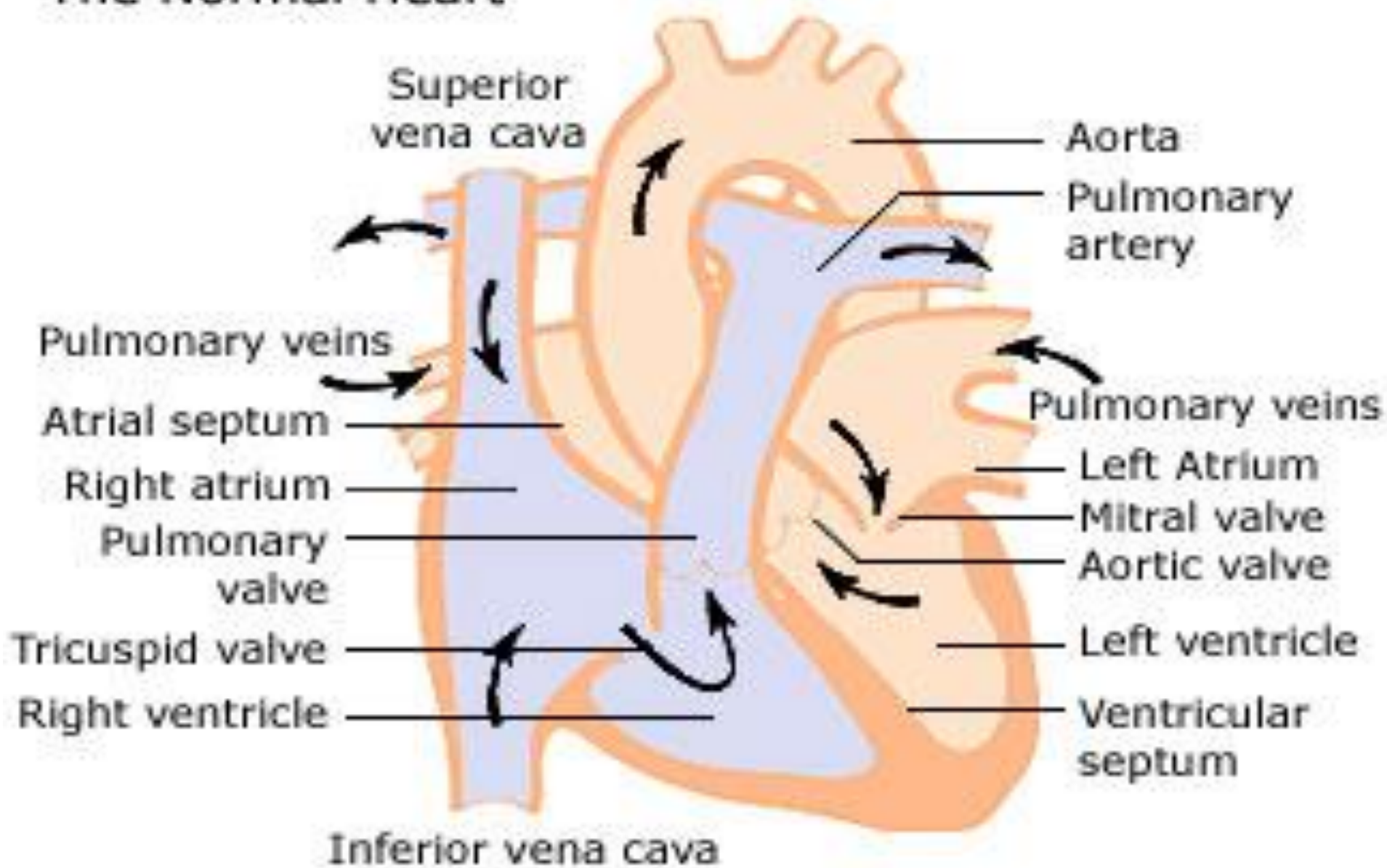
- Genetic 1 in 700 Down's Syndrome. 40% D.S. have C.H.D.
- 17 % CHD occurs in association with a syndrome
Turner's/Williams/Noonan's Syndrome
- Non Genetic
- Environmental Factors e.g. Radiation
- Infection/Virus e.g. Rubella
- Maternal Conditions e.g. Diabetes
- Maternal **drugs** e.g. anti-epileptics, lithium, alcohol



COMMON CONGENITAL HEART DEFECTS

- Atrial Septal Defect 10%
- Ventricular Septal Defect 30%
- Tetralogy of Fallots 6%
- Transposition of the Great Arteries 4%
- Coarctation of the Aorta 7%
- Patent Ductus Arteriosus 10%
- Aortic Stenosis 6%
- Pulmonary Stenosis 7%
- Other **20%**

The Normal Heart



Lesion information on all lesions on www.swswchd.co.uk

Professionals → Clinical information → adults



South Wales and South West
**Congenital Heart
Disease Network**

University Hospitals Bristol **NHS**
NHS Foundation Trust

Fontan Circulation (Total cavopulmonary circulation (TCPC))

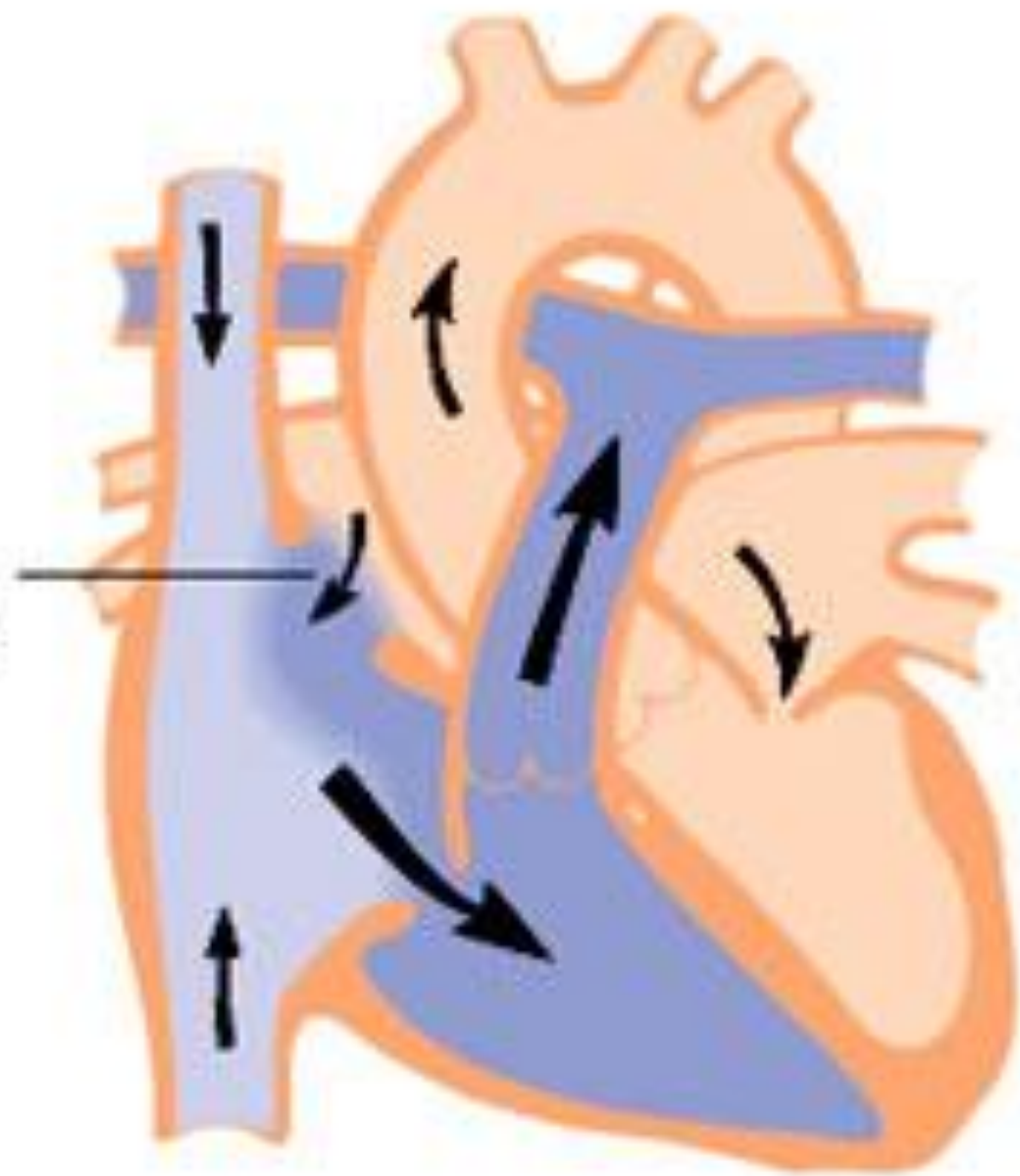
SETTING	South West England and South Wales
GUIDELINE FOR	Cardiology teams in South West England and South Wales hospitals
PATIENT GROUP	Adult patients with congenital heart disease

GUIDANCE

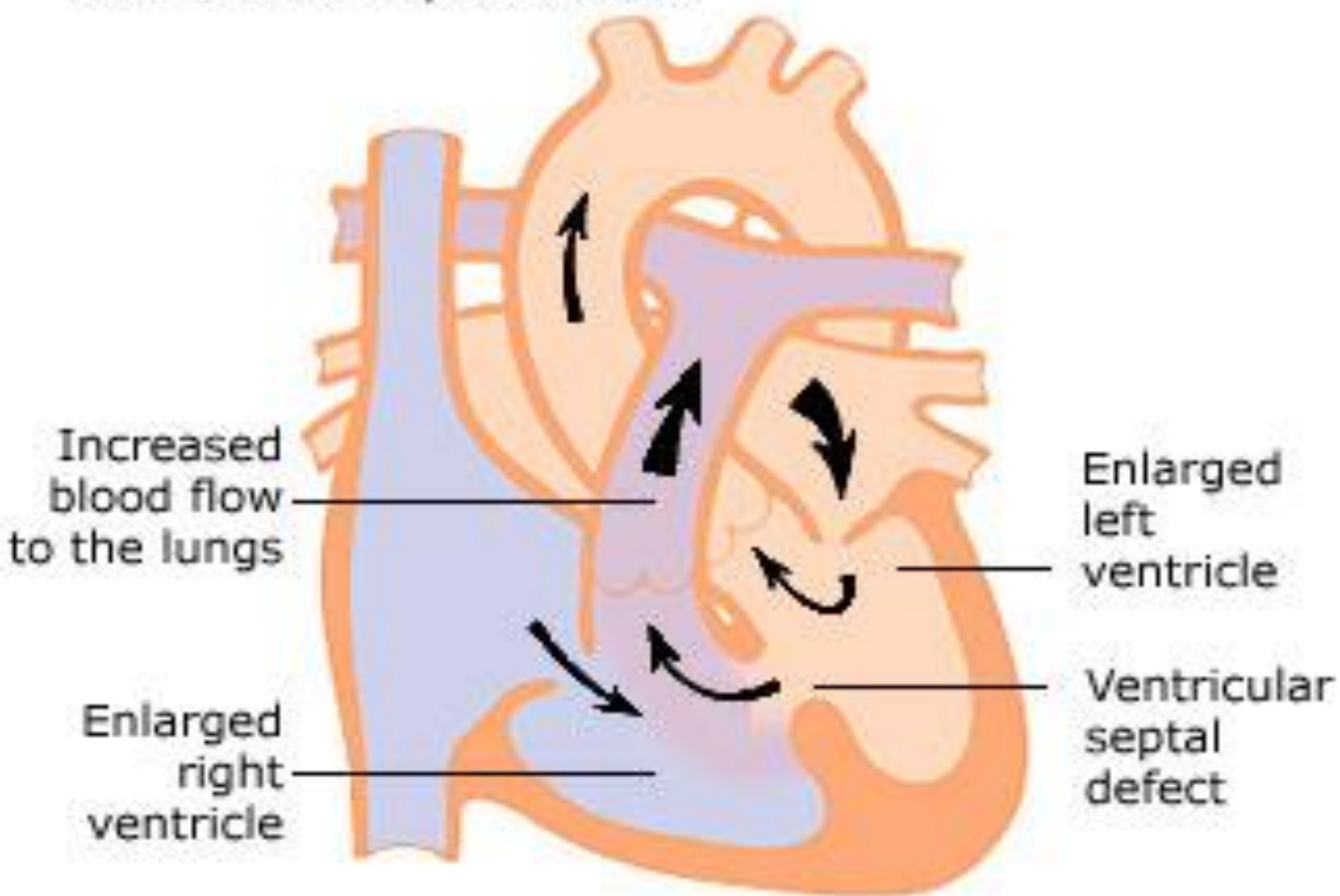
Follow-up:	annual
Associated lesions:	dependent on the underlying abnormality, note may be isomerism
Inheritance:	dependent on the underlying abnormality

Long-term complications:

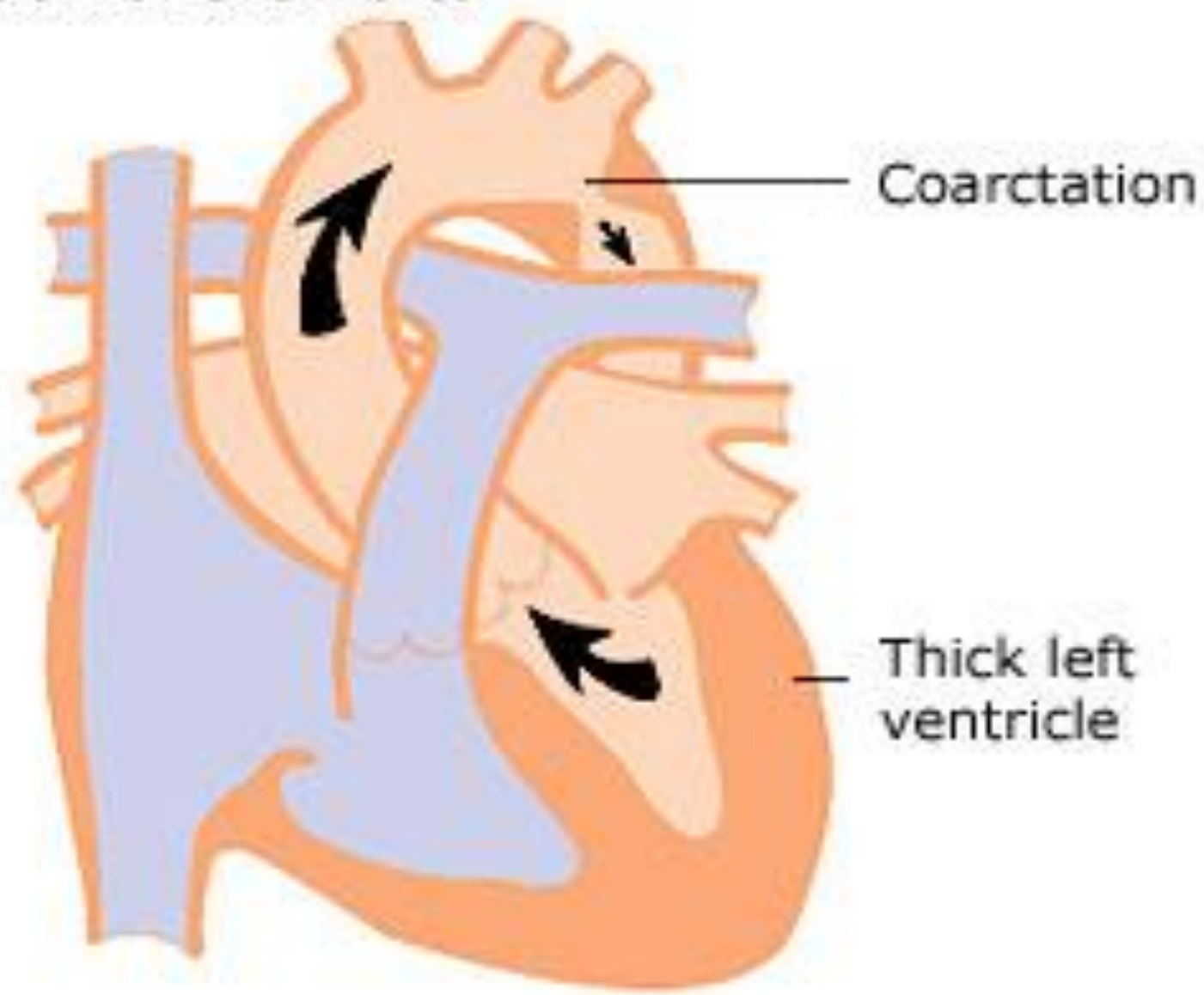
Atrial septal defect



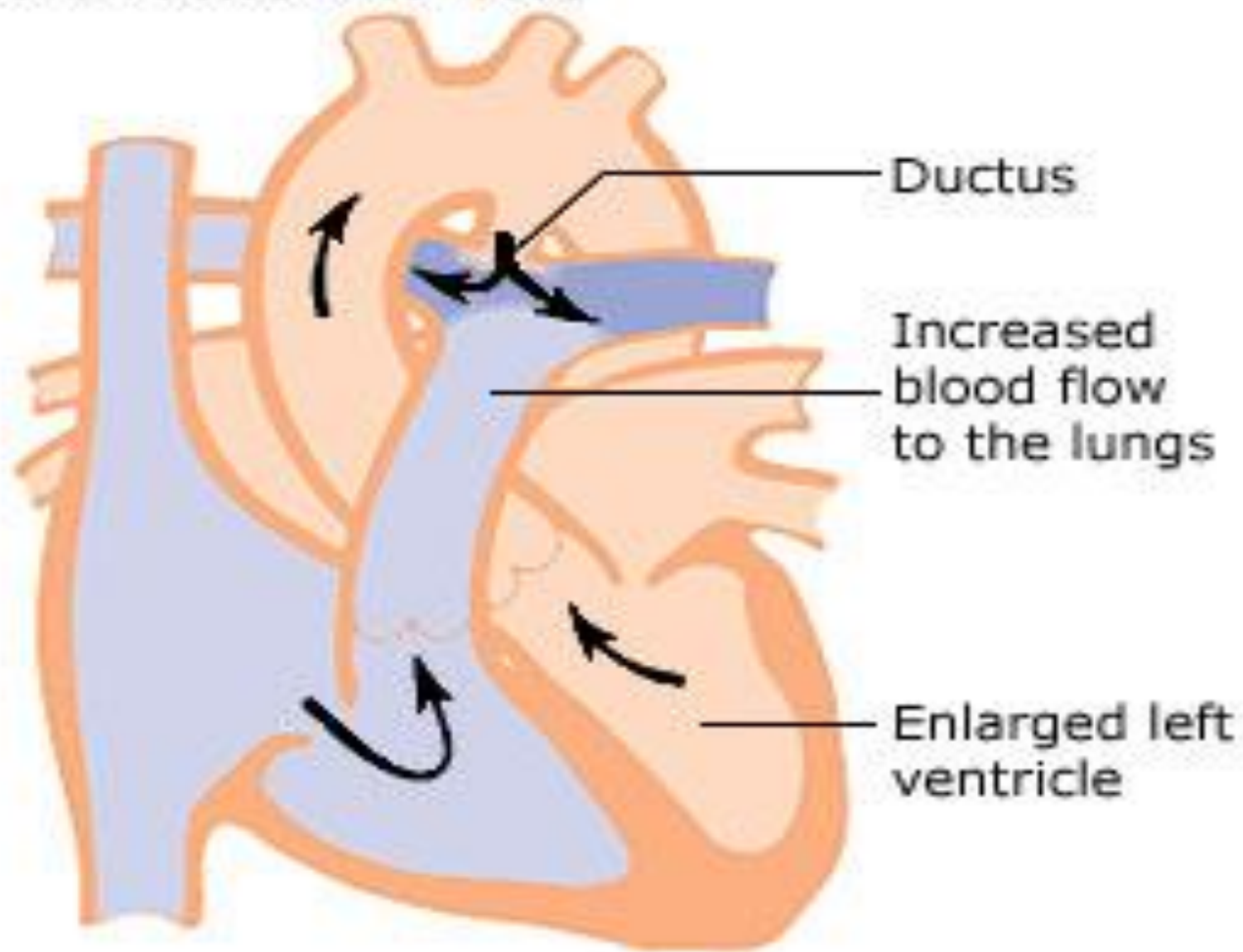
Ventricular Septal Defect



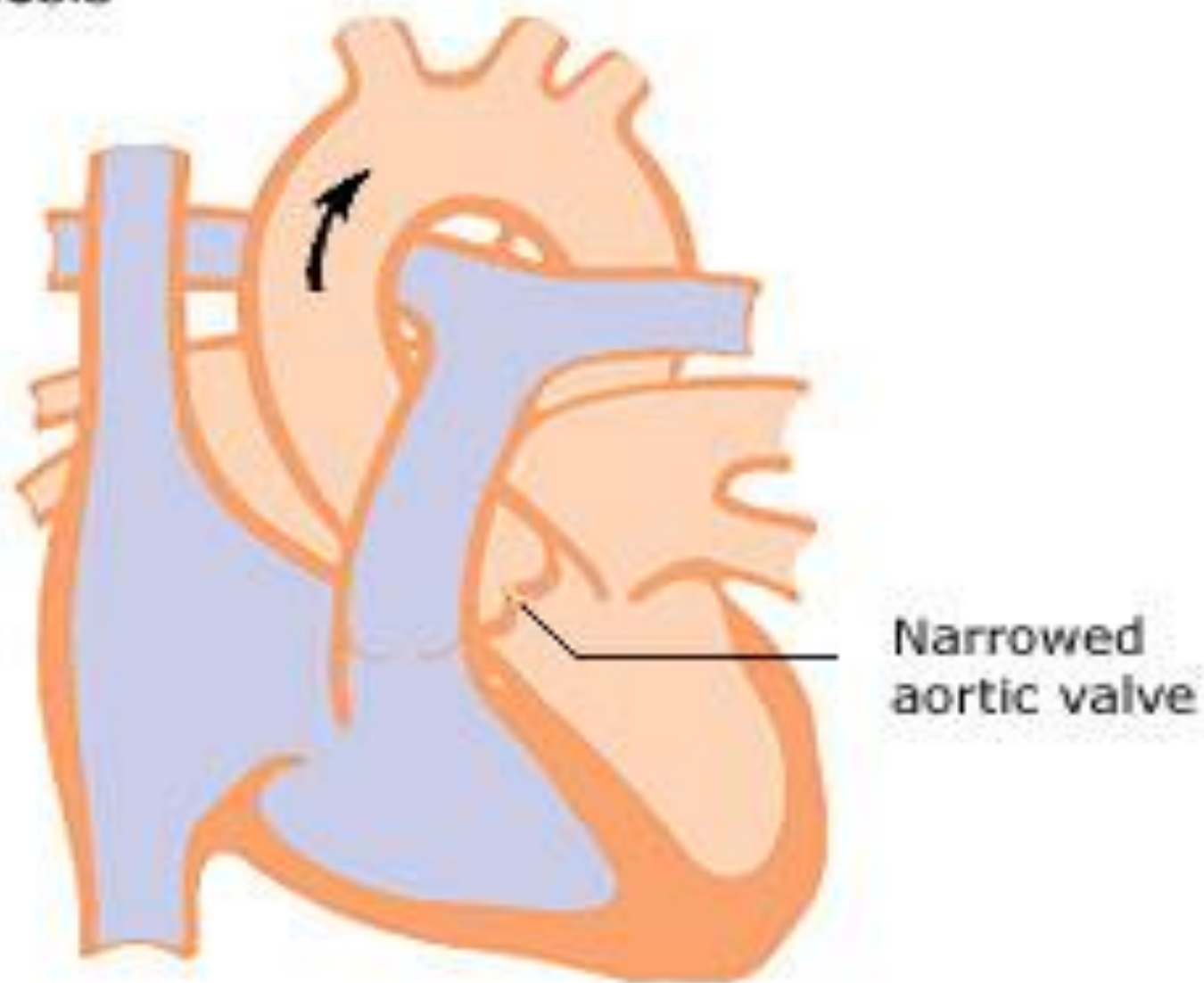
Coarctation of the Aorta



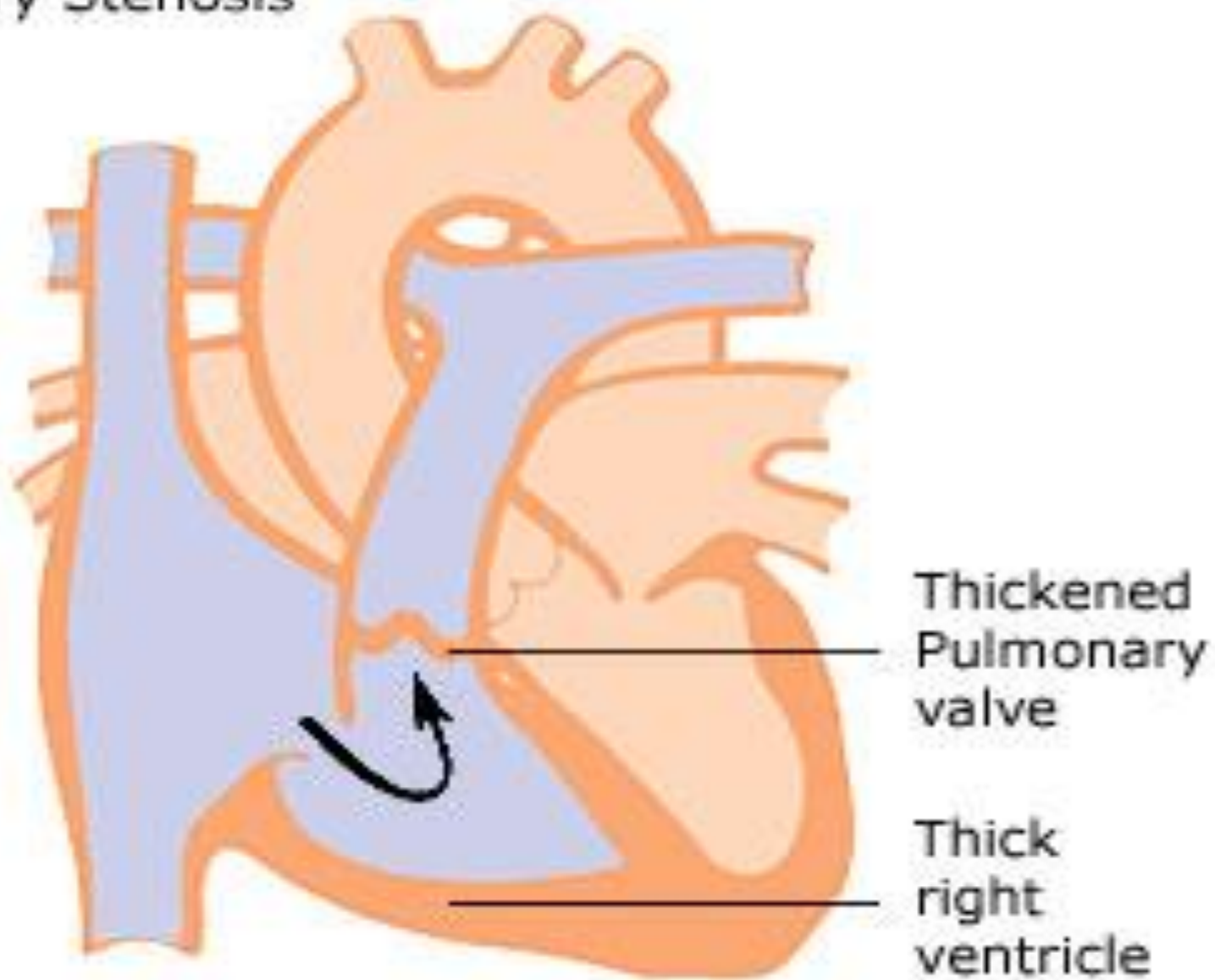
Persistent Ductus Arteriosus



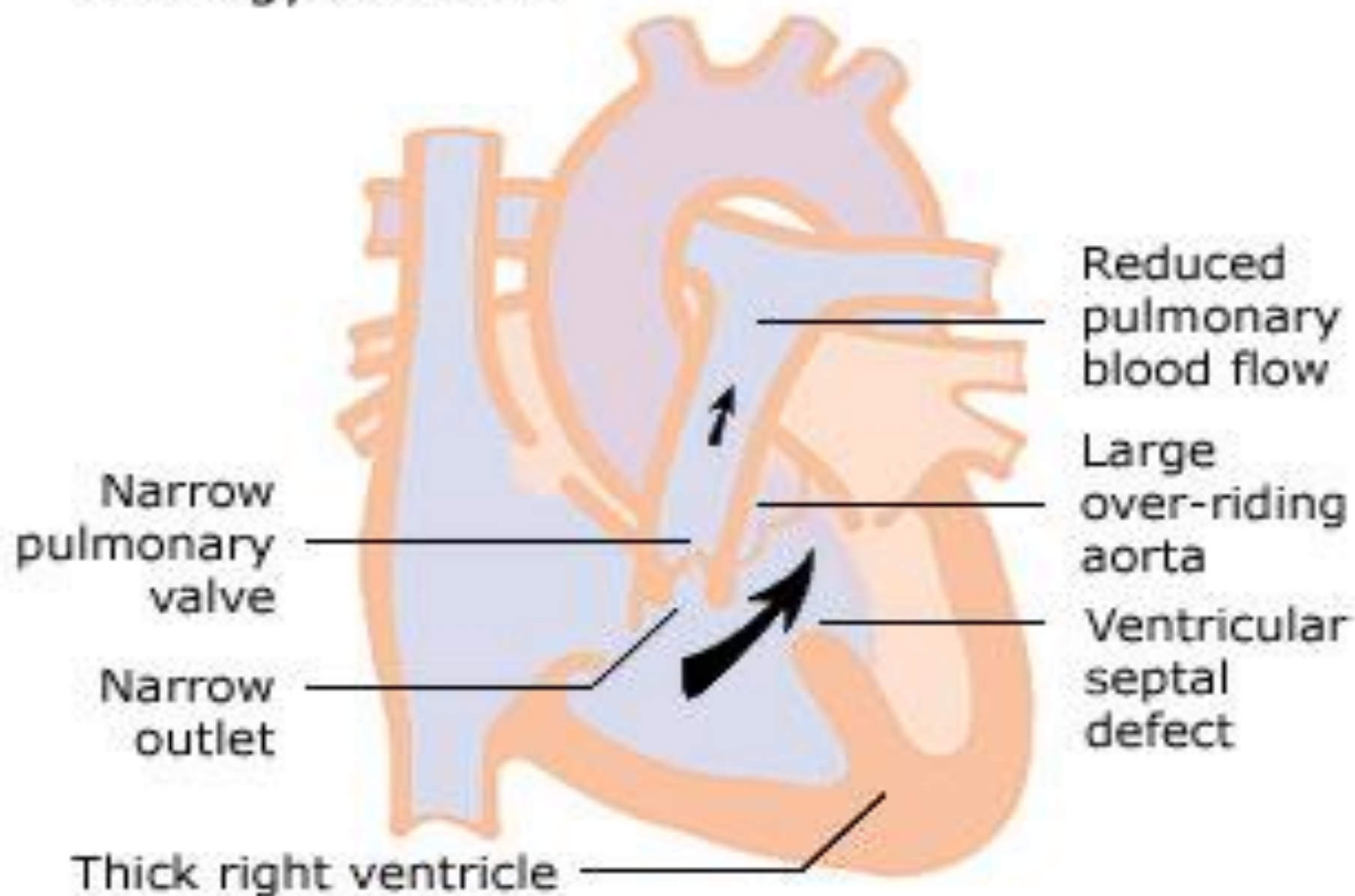
Aortic stenosis



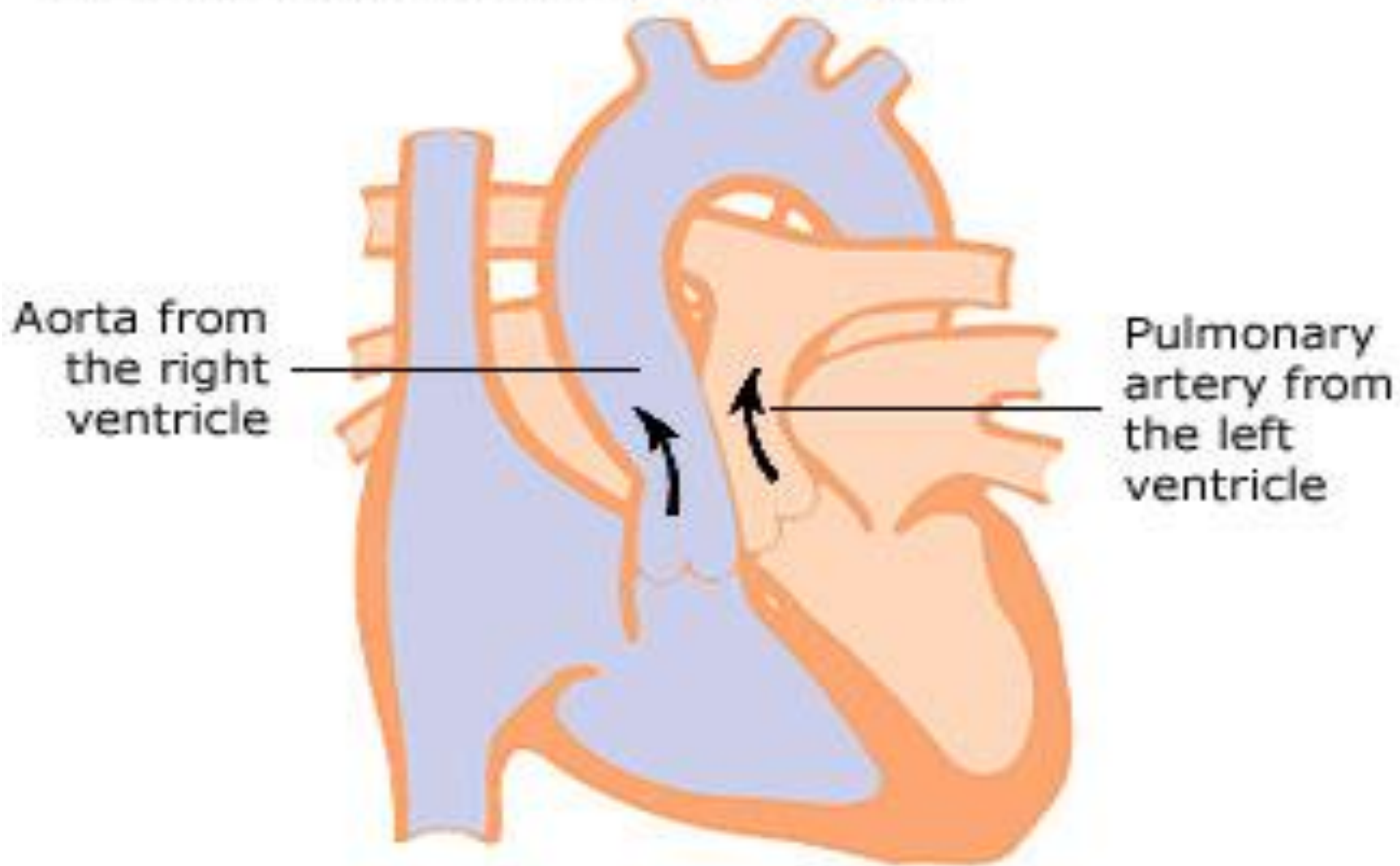
Pulmonary Stenosis



Tetralogy of Fallot



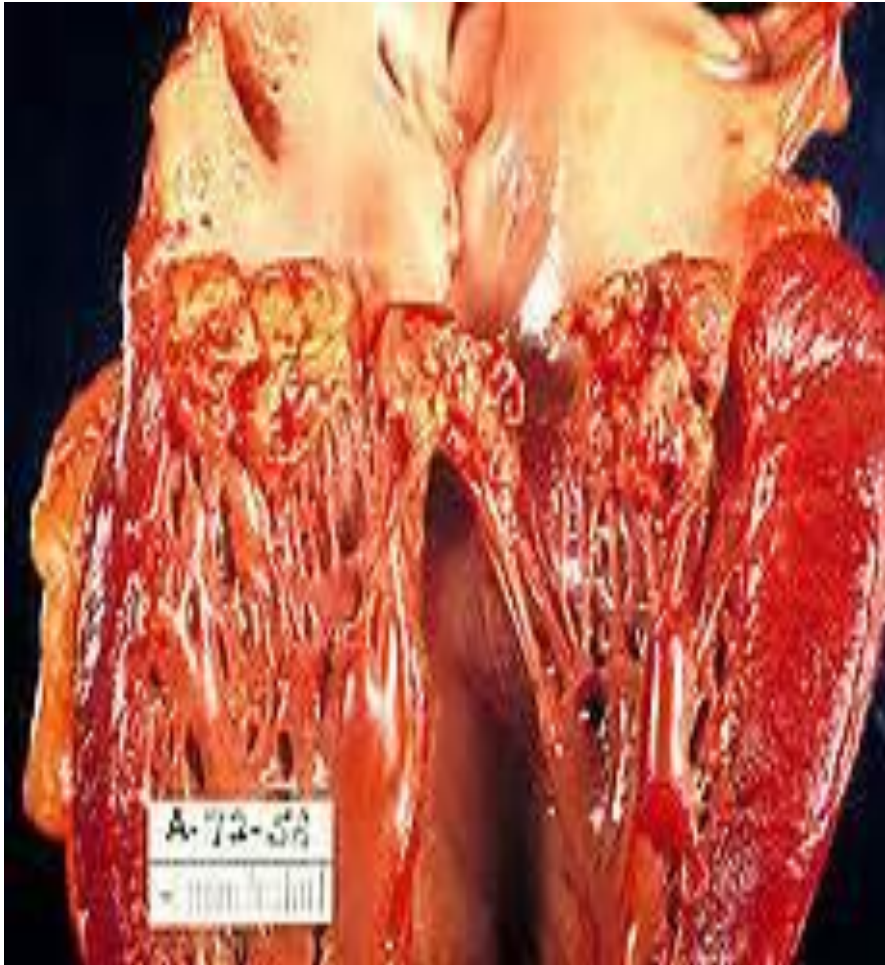
Transposition of the Great Arteries



MEDICAL ASPECTS OF CARE

- Infective endocarditis.
- Arrhythmias/heart failure.
- Surgery +/- re-operation – risks Intervention.
- Stroke.
- Cyanosis/Polycythaemia.
- Pregnancy/Contraception.
- Coronary Artery Disease.

ENDOCARDITIS



- Causes/risk?
- Clinical exam, stigmata, rash, murmur?
- Diagnosis
- Bloods,
- TOE, CXR, CT, CMR
- ECG
- Urine dip blood
- Treatment IV
- Complications
- Prophylaxis advice
- Nursing

ESC /AHA/ www.nice.org.uk

PIERCINGS



ARRHYTHMIAS

- Operative procedures from the early years, scarring affecting the conducting pathway.
- A/F, atrial flutter signs of deterioration in patients with Fontans, Fallots, A.S, single ventricle hearts and right sided conduit
- Treatment return to S/R, anti-coagulate
- Risk of S.C.D.
- EOL discussion

ARRHYTHMIA

- Urgent cardioversion
- Mapping
- Catheter ablation and surgical approaches
- Pacing/ ICD
- Medication/side effects/pregnancy
- **Danger Fontans and Ebsteins ,TGA Mustards or Sennings flutter**
- SVT most common
- **VT** in AS + TOF

RIGHT SIDED HEART FAILURE

- Fontans
- PH
- Large ASD
- TV disease Ebsteins
- RV/PA stenosis eg Rastelli
- Mustards with baffle obstruction
- Systemic RV eg CCTGA, Mustards

RIGHT SIDED ♥ FAILURE

(Cor Pulmonale)

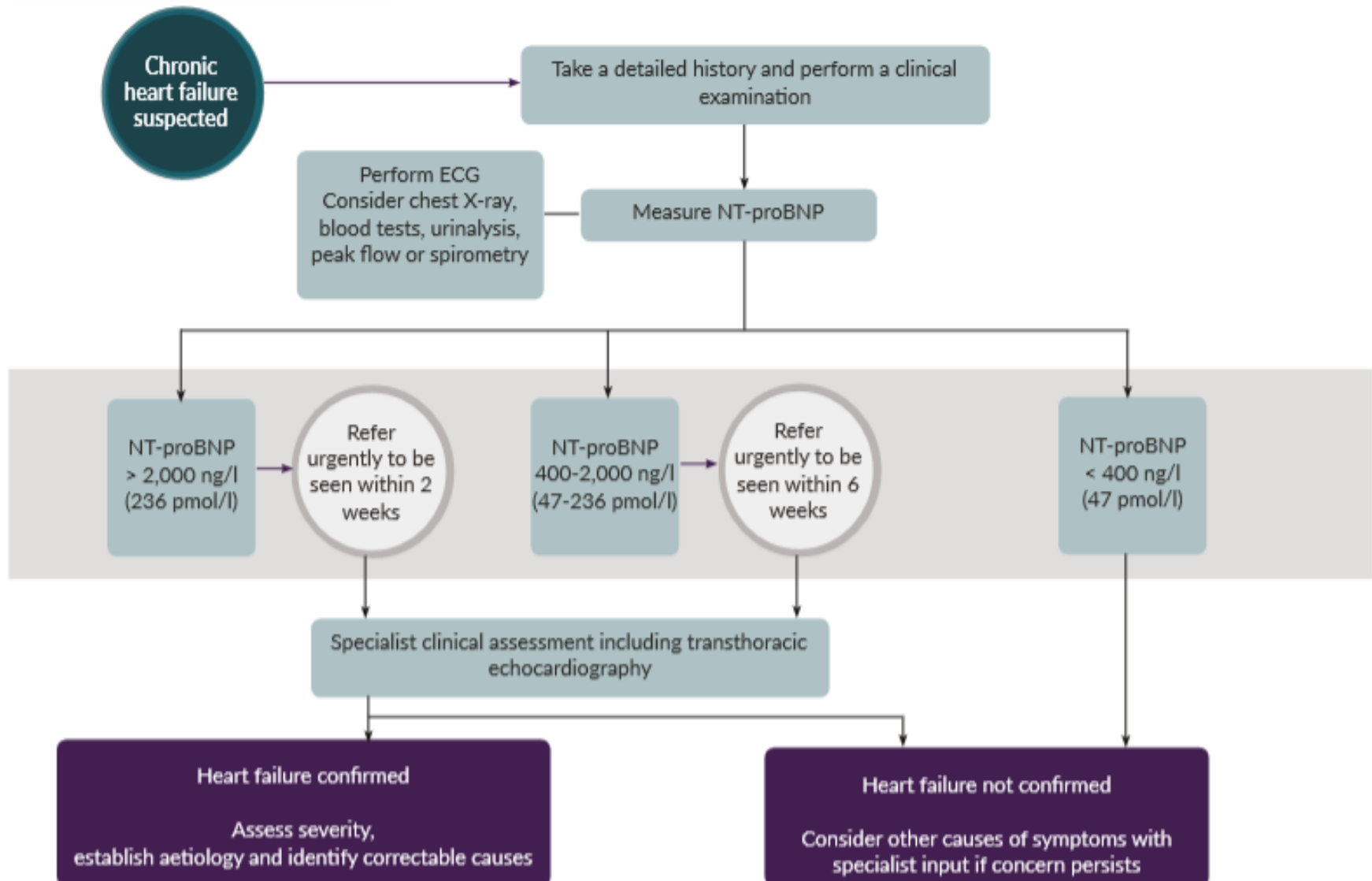
- Fatigue
 - ↑ Peripheral Venous Pressure
 - Ascites
 - Enlarged Liver & Spleen
 - May be secondary to chronic pulmonary problems
 - Distended Jugular Veins
 - Anorexia & Complaints of GI Distress
 - Weight Gain
 - Dependent Edema
- 

HEART FAILURE

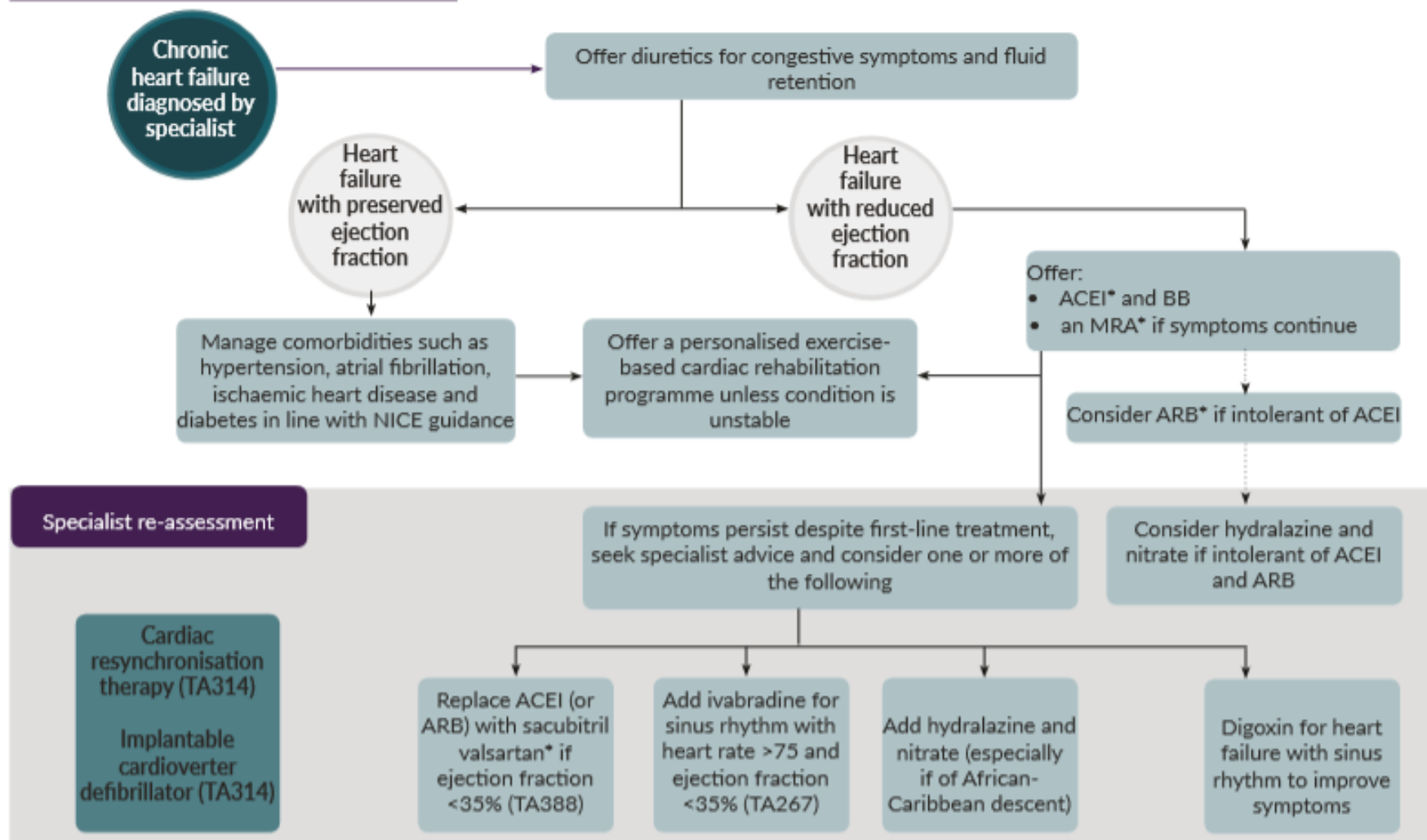
- Medication ACE-inhibitors, angiotensin receptor blockers (ARBs)
- Beta-blockers, aldosterone blockers (spironolactone or eplerenone)
- Diuretics, ivabradine, digoxin (occasionally)
- Fluid restriction, daily weight
- Lifestyle changes, smoking, diet, exercise, salt
- Devices, pacemakers, CRT, ICD
- Surgery, valve, LVAD, transplant



Chronic heart failure: diagnosis



Chronic heart failure: management



*Measure serum sodium, potassium and assess renal function before and after starting and after each dose increment. If eGFR is 30 to 45 ml/min/1.73 m², consider lower doses or slower titration of ACEI or ARBs, MRAs, sacubitril valsartan and digoxin.

SURGICAL ASPECTS OF CARE

- Risk of re-operation in this group
- Adhesions, bleeding, longer by-pass time
- Renal and liver function problems
- Arrhythmias
- Cyanosed patient will require a higher PCV.
- Higher filling pressures needed in some conditions FBC
- Pericardial and pleural effusions

SURGICAL EMERGENCIES



- Complications
- Bleeding, infection, fever, thrombosis, embolism, fluid overload, dehydration, PH, arrhythmias, ventricular dysfunction
- Early detection vital, aggressive management
- Pain control for catecholamine stress
- Avoid early discharge

CYANOSIS

- Cyanosis results from an increase in RBC as the body attempts to improve its oxygen carrying capacity
- Increased viscosity, thrombosis, stroke, embolus, PH
- Caution if NBM, IV fluids
- Caution with oxygen
- Venesection

3.4.8 Management of cyanotic patients

Cyanosis is caused by R–L shunt due to an anatomical communication between the systemic and pulmonary circulation at the atrial, ventricular, or arterial level. Cyanotic heart disease comprises a heterogeneous group of lesions with different underlying anatomy and pathophysiology: normal or restricted pulmonary blood flow in the presence of an obstruction across the pulmonary outflow tract or increased pulmonary blood flow in the absence of such an obstruction which, in some defects, may result in development of PAH and

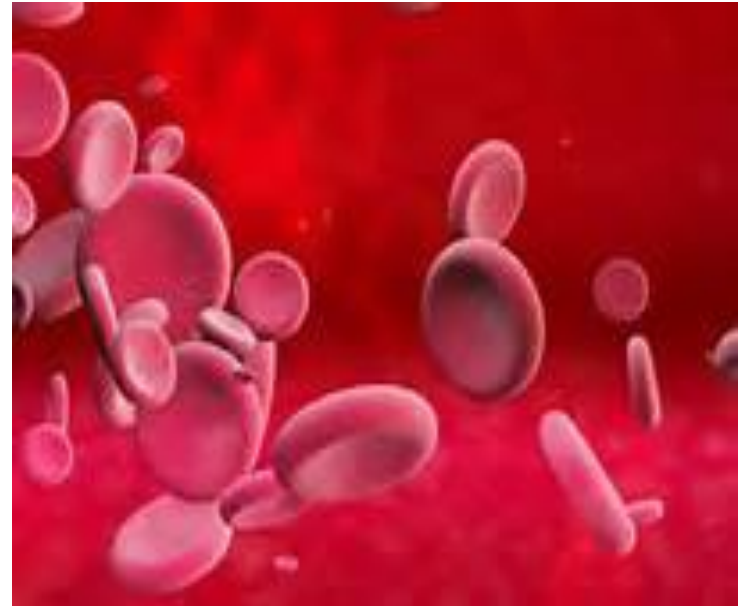
CYANOSIS



CYANOSIS

Watch for.....

- Sepsis, brain abscess
- Renal function
- Gout
- Gall stones
- Orthopaedic complications
- Skin, acne, I.E.
- Ferritin



Cyanotic Congenital Heart Disease

Pathophysiology

Eisenmenger syndrome: pulmonary vascular disease secondary to non-restrictive shunt

Shunt with normal or restricted blood flow to the lungs

Advanced Therapies

Pulmonary vasodilator therapy?

- Additional shunt / occlusion of shunts?
- Biventricular / univentricular repair?

Tests

Blood work (Box 3) Imaging (echocardiogram/MRI/CT) 6 Minute Walk Test

Complications

Hyperviscosity Symptoms

Thrombo-embolic complications

Bleeding complications (minor / major bleeding)

Gouty Arthritis

Infections (brain abscess?)

Renal dysfunction

Secondary erythrocytosis

Dehydration

Iron deficiency / anemia

Patient on any anticoagulants?

Phlebotomy

Rehydration

Iron Supplements

Preventive

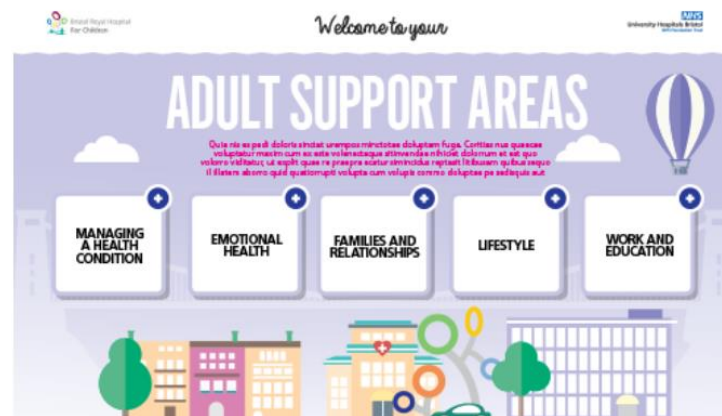
Annual Flu Shot Excellent oral hygiene Endocarditis prophylaxis Air filter No smoking/drugs
Pneumovax

EMERGENCIES Get help!

- Arrhythmia
- Surgery
- Cyanosis
- Infection
- Ht Failure
- Ischaemia
- Pregnancy
- Transplant

PSYCHOLOGY

- Anxiety about heart condition, prognosis
- Repeated hospital visits
- Risk taking behaviour
- Compliance
- Depression
- Phobia
- L.D.
- Toolkits on website for patients www.swswchd.co.uk



🏠 Patients & Families ▾

Support for you

- Clinical experience in Level 1, mentoring across the network
- Education/ Study pack-link nurse resources
- Annual and regional study days
- National group BACCNA/ BCCA



South Wales and South West
**Congenital Heart
Disease Network**

What are you looking for?

Select Language

Resize text: A A A

Home Patients & Families Professionals About Hospitals Patient Pathways Research Charities Contact Us

Clinical Information
Patient Support
Training & Education
Governance
Useful Resources
Audit
CHD ACHD nurses/Link nurses

Fetal
Children
Dentistry
Transition & Young People
Adults
Learning Difficulties
Palliative Care
Pregnancy

Families

Home | Patients & Families

This section of the website is dedicated to patients and their families/carers. Here we hope you will find lots of resources that are useful to you.

We have shared some **Patient Stories** for you to read. If you would like to add your story to our website, please **Contact Us**

The **Leaflets** section contains online versions of many leaflets relevant to a congenital heart condition that we hope you find useful. (If you are a professional with a new leaflet you'd like to add, please get in touch with Sheena.Vernon@uhbristol.nhs.uk).





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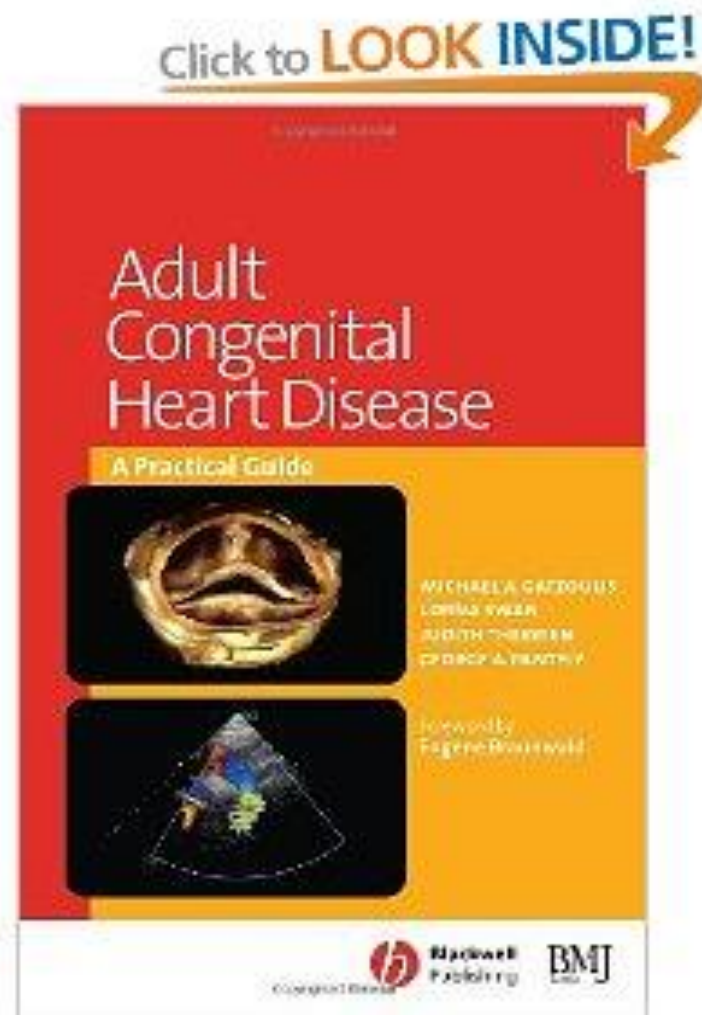
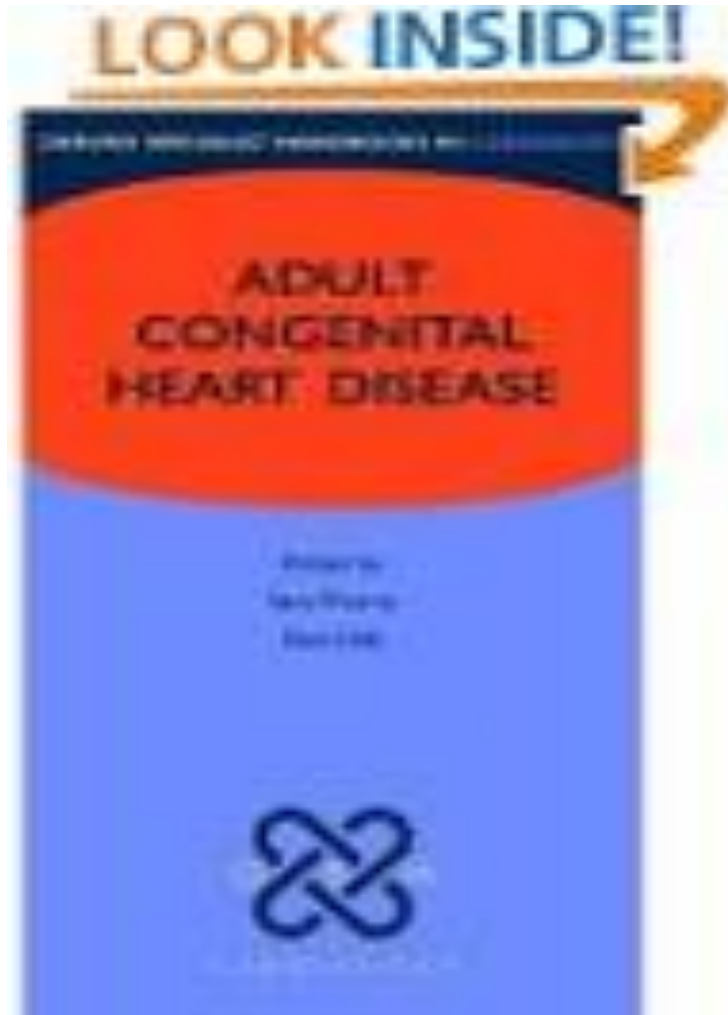
[Adults](#)

Adult congenital heart disease nursing

RCN guidance on roles, career pathways and competence development

To support adult
standards
guidelines from
RCN for nursing
published

READING



THANK YOU!

Questions?

LOOKING AHEAD

- 
- A photograph of a paved road with two yellow arrows painted on it, pointing towards the horizon. The sky is filled with large, dramatic clouds, and the sun is shining brightly from behind the clouds, creating a lens flare effect. The overall mood is one of hope and looking forward.
- **Congenital networks**
 - **Support & encourage patients to lead as normal a life as their condition allows.**

MARFANS SYNDROME

- Tall and slender build
- Disproportionately long arms, legs and fingers
- Breastbone that protrudes outward or dips inward
- High, arched palate and crowded teeth
- Heart murmurs
- Extreme near-sightedness
- Abnormally curved spine
- Flat feet



MARFANS SYNDROME

- Connective tissue disorder, the heart (aortic dissection), eyes (dislocated lenses) and skeleton (scoliosis)
- Affects 1 in 5,000 births
- Reduced life expectancy in many patients
- **Cardiac manifestations** such as aortic dissection, aortic regurgitation and heart failure
- Cardiac surgery for abnormalities of the aorta
- Beta blockers
- *www.marfan.org.uk*

GUIDELINES



Adult Congenital Heart Disease

A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)

THE NETWORK APPROACH

sets out: how networks will work

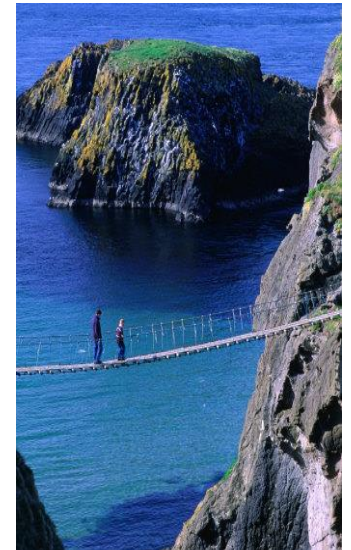
new/changing: clear leadership (clinical and professional);
cardiology (non-surgical) centres' participation in networks;
second opinions and referrals

- **Challenge** : communication between local, cardiology and surgical centres
- ACHD CNS from SSC or SCS provide support, education and a link to network opd and ward staff
- Local link nurse in local centre/cardiac CNS +ACHD

TRANSITION

- **sets out** seamless pathway of care to adult services
- **new/changing:** young people to be seen at least once at transition by a specialist with ACHD expertise; clear care plans/transition passports agreed; respecting particular needs of young people with ***learning disabilities*** and their carers.

- **Challenge:**
- Big numbers
- Young adult clinics, individual time + CNS time
- Letters of introduction to patients
- In-patient and out-patient support
- Appropriate information
- Avoid loss of F/up



Pregnancy

- Pre-pregnancy counselling for moderate to severe lesions & also:
- High risk, PH, severe Left sided lesions, Aortic root dilatation, cyanosis, ejection fraction less than 40%, mechanical valves.
- Care with ACE inhibitors, angiotensin11 receptors blockers and Amiodarone.

PALLIATIVE CARE AND BEREAVEMENT

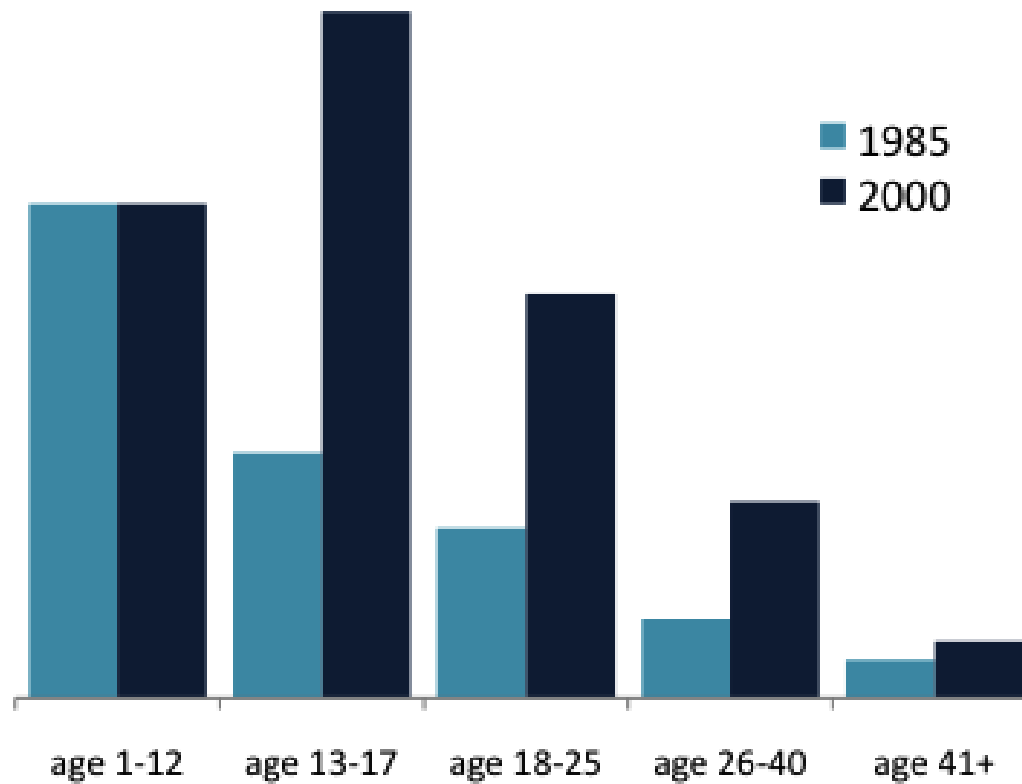
sets out: how to provide support at end of life and how to manage communication with families around the end of life

new/changing: all new

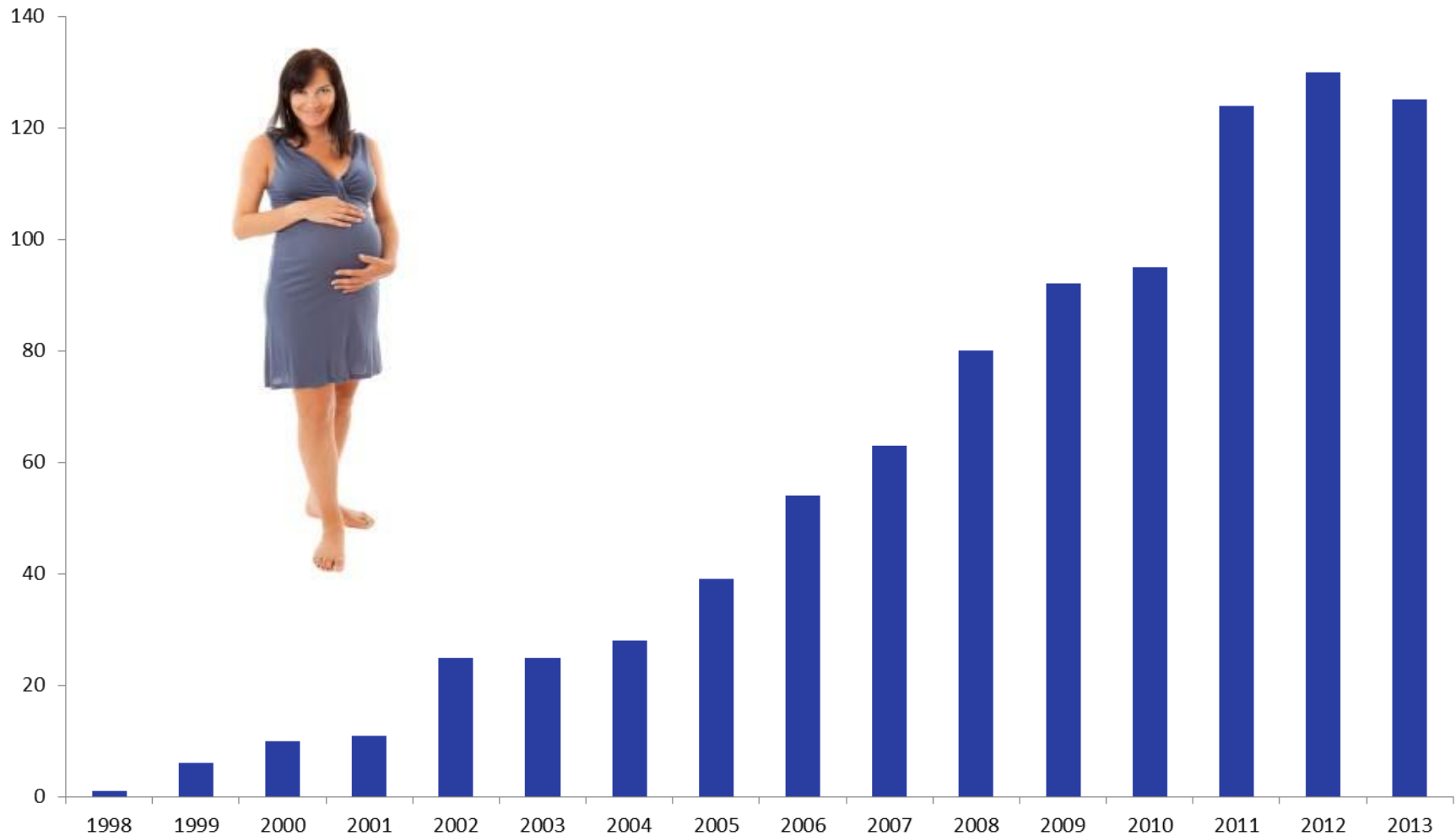
- **Challenge** : difficult conversations, patient, parents spouse, family and children
- Intense telephone advice
- End of life pathway
- Palliative care teams
- GP support



49% adults in 2000



130 new **pregnant** referrals in 2013



Charities

- Newsletter / leaflets
- Telephone help line
- Support groups/mental health
- Financial support
- Workshops / conferences
- Web Sites
- BHF Lifestyle advice



CoaguChek machines



- INR test
- www.roche-diagnostic.co.uk
- www.coagucheck.co.uk

Charities

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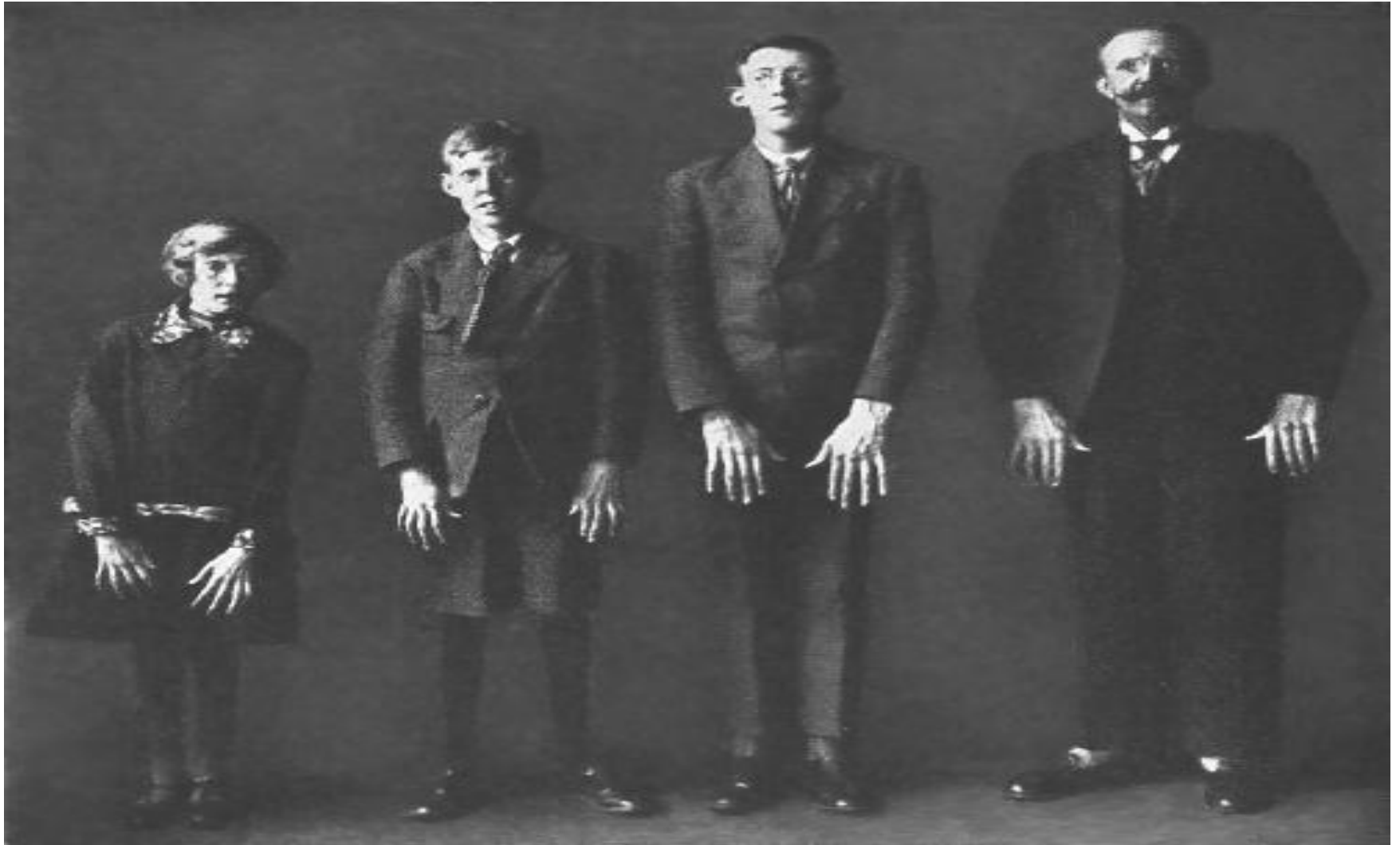
PATIENT PHONE CALLS

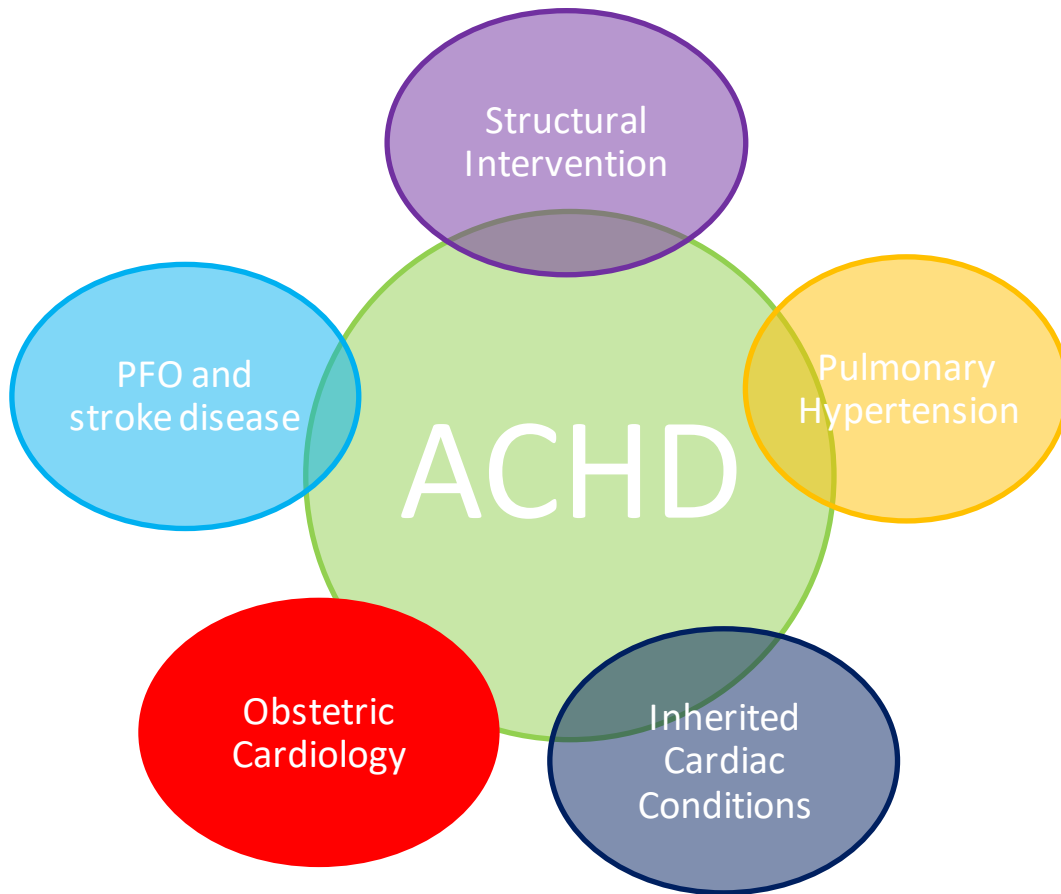
- 2,000 calls pa admission, surgery, intervention, pregnancy, learning disability, TYA. Advice for HC professionals.
- Support, bereavement.
- Long haul flights/ travel.
- Employment issues/benefits.
- Managing Warfarin – INR – Coagu check.
- Tel. Pre-op.

Piercings



MARFANS SYNDROME





Learning Disabilities

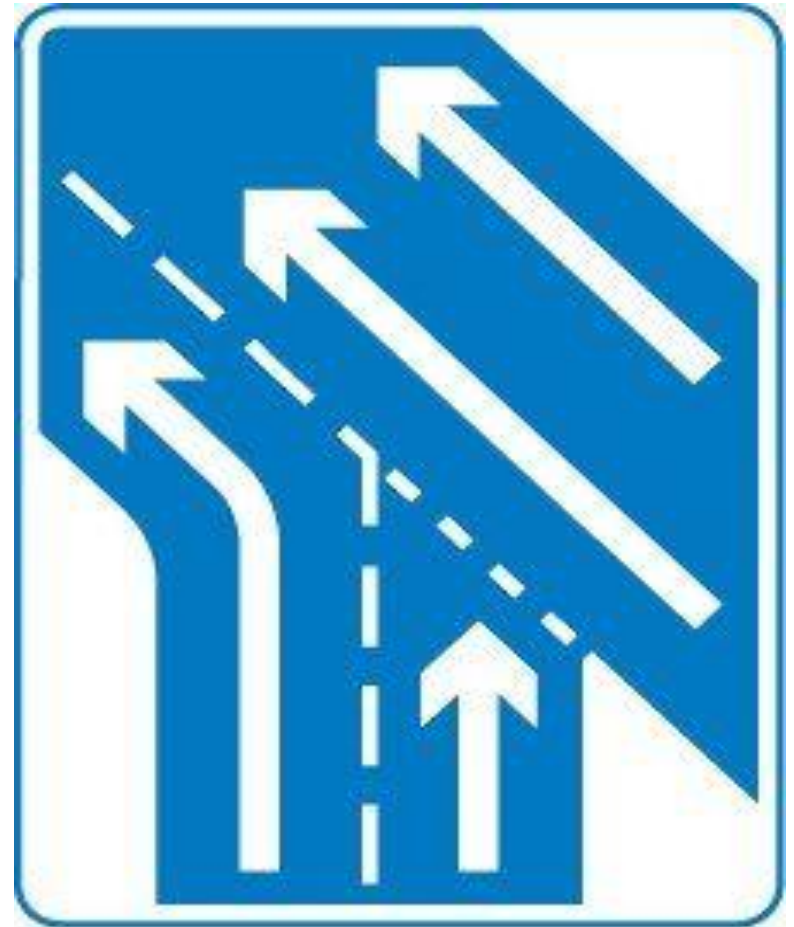
- Increasing numbers of patients having procedures and treatment
- 1 in 700 born with Downs, 40% will have CHD
- Time consuming
- Support for patient, family, CLDT and carers
- Capacity to consent? Best interest meetings?
- Appropriate communication

NURSING TEAM OF THE YEAR 2014



Lifestyle issue

- Outline of population
- Diet, alcohol, smoking and drugs
- Endocarditis
- Exercise
- Sex, pregnancy and contraception
- Extreme sport
- Risk taking
- Travel
- Support



Arrhythmias

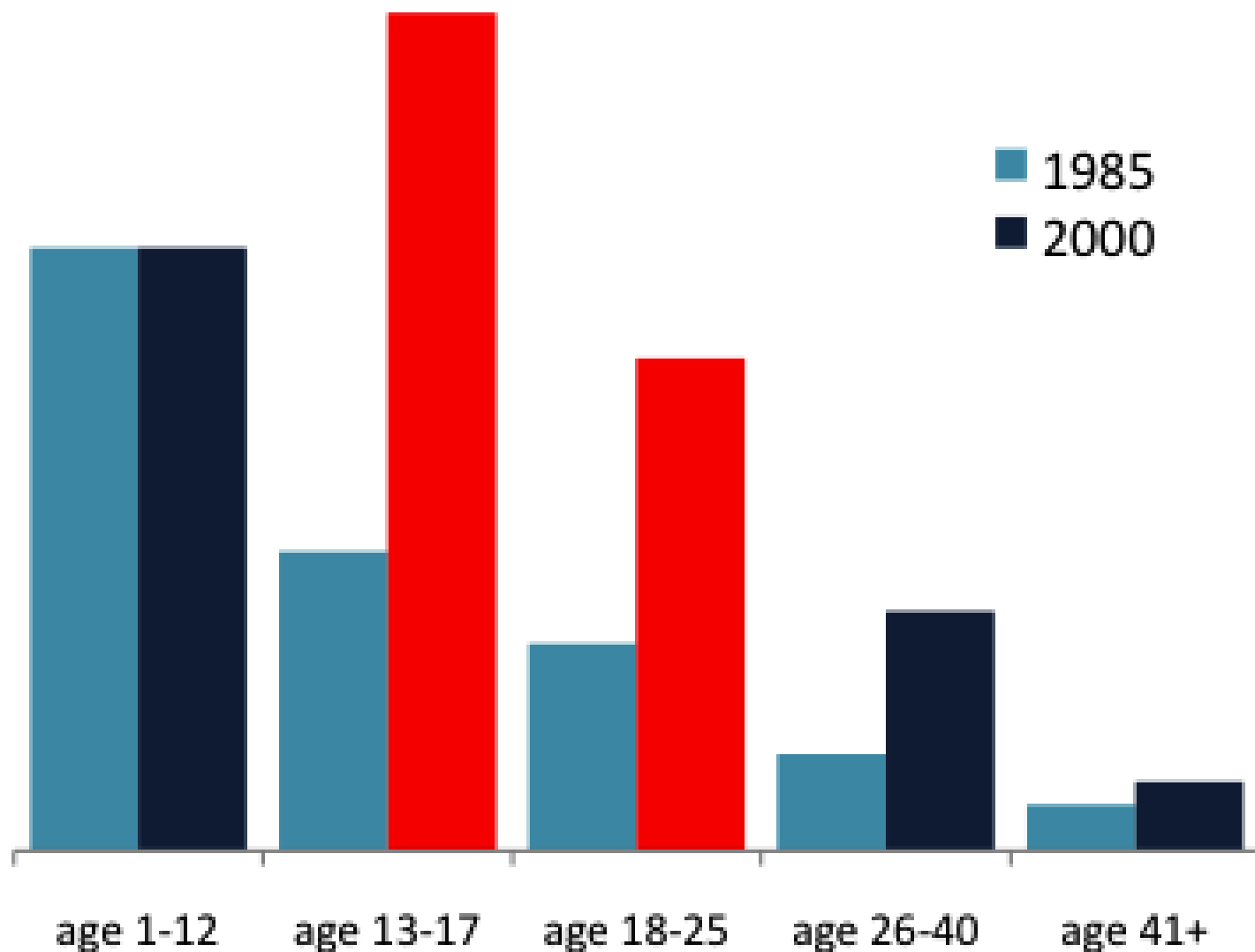
- Causes
- Precipitating factors
- Deterioration
- Treatment
- Structural v Electrical
- Haemodynamics
- SVT most common
- **VT** in AS + TOF



ADVICE LINE



number of 13-25 year olds **increased x 3**



2007-2014

