

# Adult Congenital Heart Disease An Overview

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Lead Nurse CHD Network  
Bristol Heart Institute

2019



08.30 - 09.00	Registration and refreshments	Lecture Th 1
09.00 – 9.45	Overview of 8 common lesions	Sheena Vernon Lead Nurse
09.45 –10.30	Surgery for the single ventricle	Mr Andrew Parry
10.30 –11.00	Coffee	
11.00 –11.30	Red flags for general anaesthetic in complex congenital patients	Dr Amit Ranjan
11.30- 12.00	Pregnancy with Fontan’s circulation	Dr Stephanie Curtis
12.00- 12.30	Life threatening arrhythmia in Fontan patients	Dr Graham Stuart
12.30 –13.30	Lunch	
13.30 – 14.00	Fontan associated liver disease	Dr Radwa Bedair
14.00- 14.30	Patient story	Dr Andrew Shearn
14. 30- 15.00	Transferring complex patients to the adult services	Sheena Vernon RGN
15.00 – 15.15	Tea	
15.15 – 16.00	Interactive cases with group quiz	Caryl Evans RGN

# AIMS OF THIS TALK

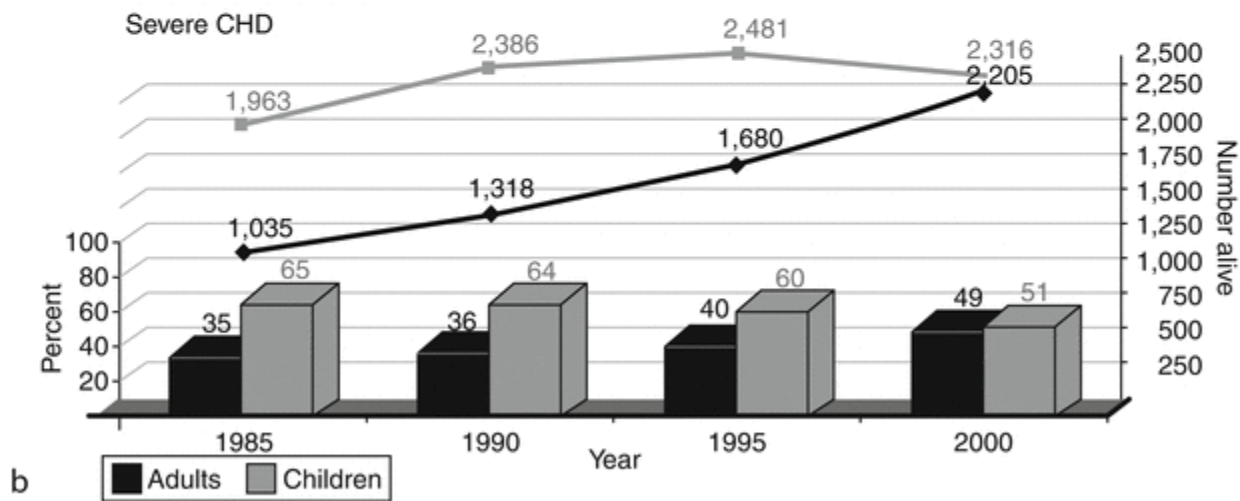
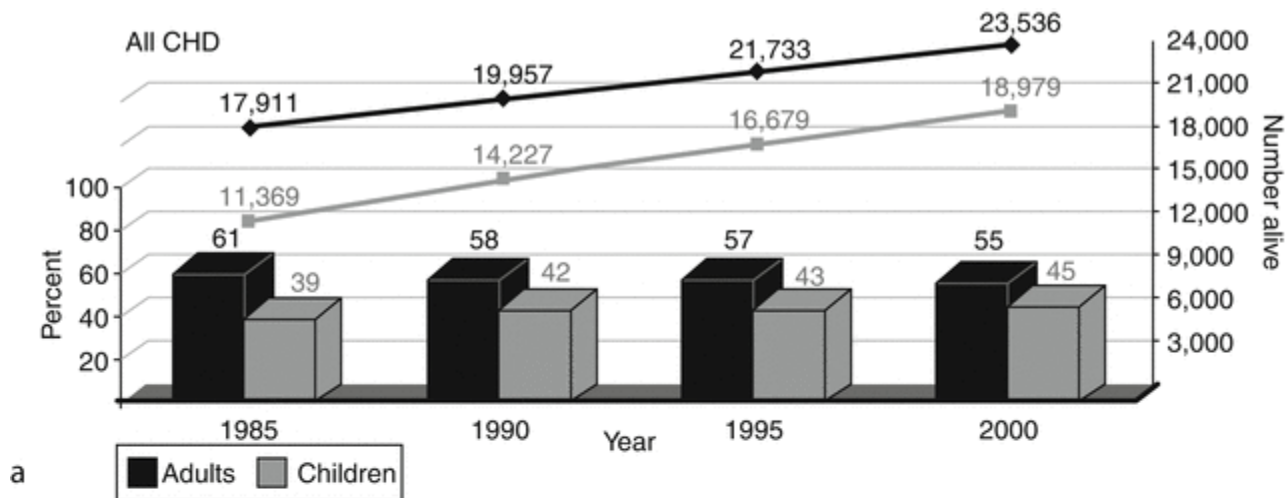
- Develop knowledge and skills.
- Develop insight into on-going needs of the patient group & changes in their condition.
- Develop ability to provide information on services suitable for individual patients and family members.
- Enable patients to participate in decisions regarding health & social issues that arise throughout life.

# OUTLINE

- Set the scene population
- Outline team
- What do we do
- Role of CNS
- Lesions
- Physical and psychological issues
- Guidelines

# POPULATION

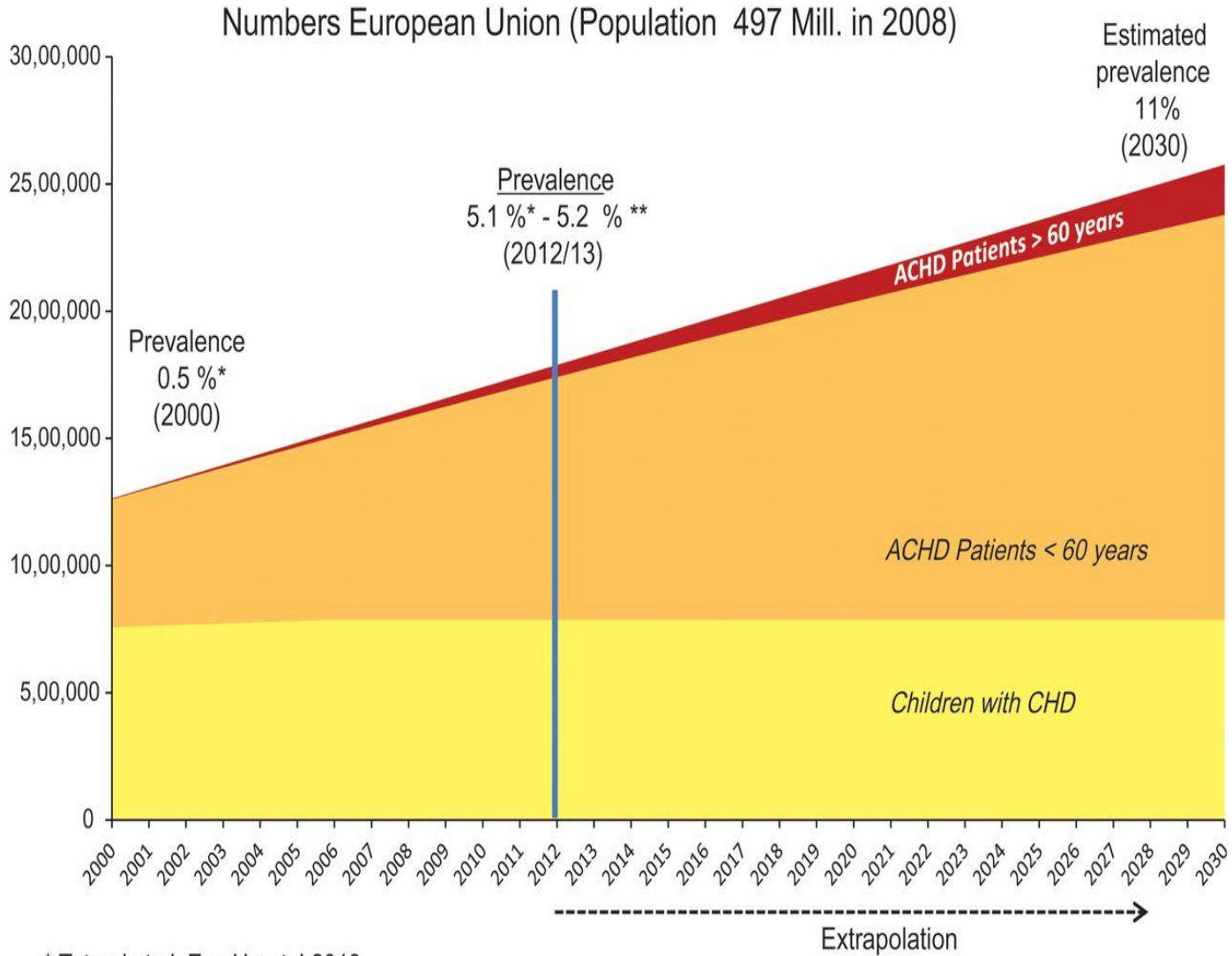
- Incidence: 8 per 1000 live births.
- 40 yrs. ago mortality from untreated CHD was 60%- 70% over the age of 18 years.
- Success of cardiac surgery and cardiology in infancy improved life expectancy.
- 85% of CHD patients, including complex, rare and severe conditions will reach adulthood.
- **More**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



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)*



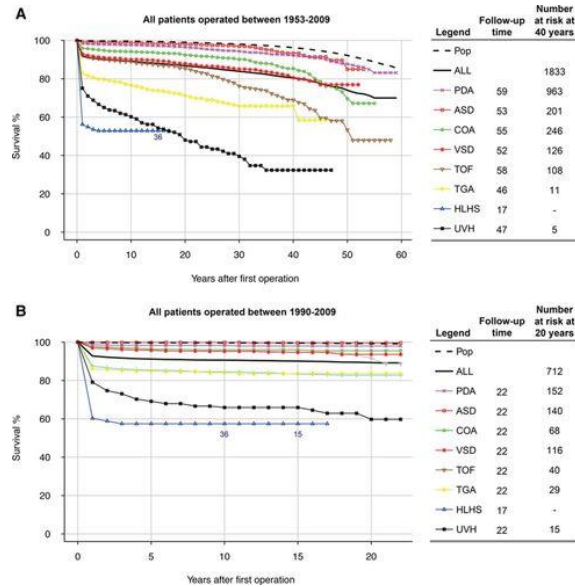
\* Tutarel et al. Eur. Heart J 2013

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





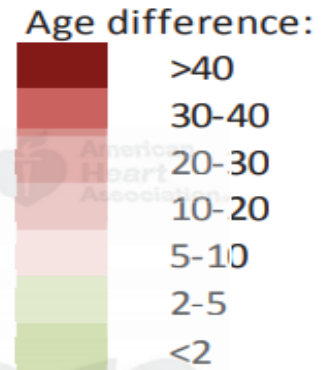
# Substantial improvements in outcome



Progress in Late Results Among Pediatric Cardiac Surgery Patients, Volume: 131, Issue: 4, Pages: 347-353, DOI: (10.1161/CIRCULATIONAHA.114.011190)

# Relative age

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

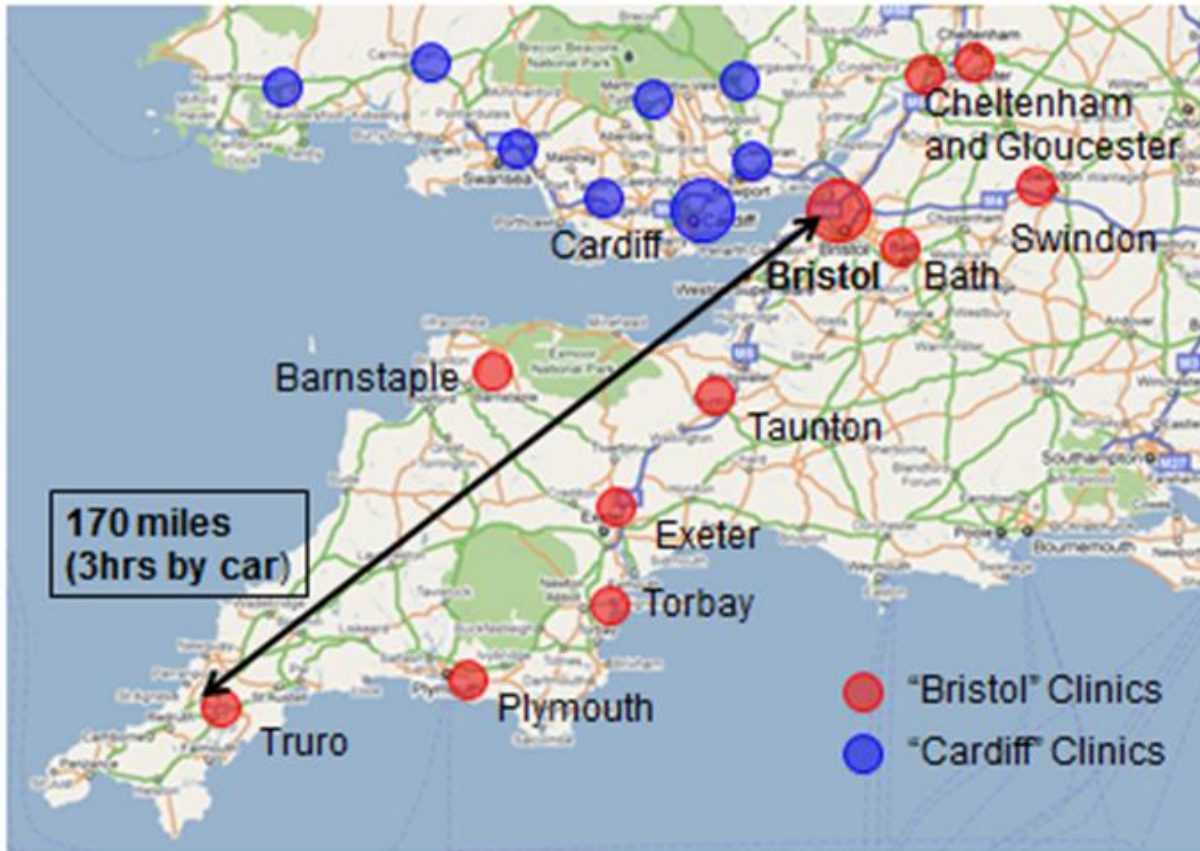


Values present relative age adjusted for predicted 5-years mortality. Colors reflect the difference between relative and actual age. For example a 40 year old Fontan patient has a mortality rate that is comparable to that of a 75 year old individual without CHD.

# CURRENT POPULATION

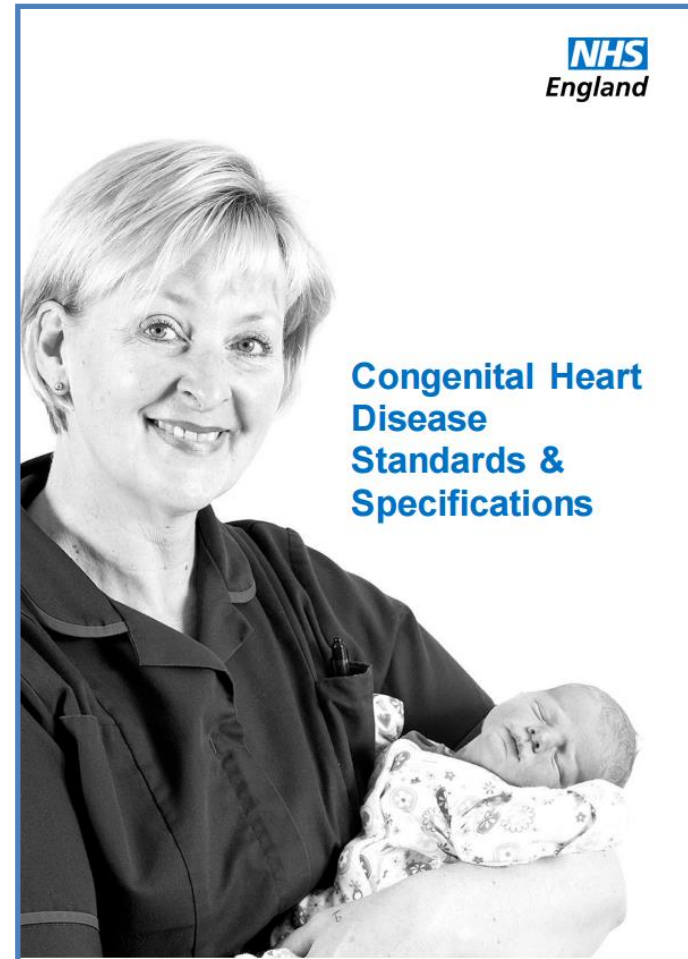
- 8,000 **Adults** South West
- 6,500 **Children** 135,000 adults and young people England
- In 2000 equal numbers of those alive with severe CHD were adults.

# CHD Network



# CHD STANDARDS

- 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, registrar x 2
- CNS x 3 (5)
- Obstetric team, 108 new pts
- Consultant Radiologists
- Anaesthetist CHD interest
- Peripheral clinics in 7 D.G.H's
- Barnstable, Cheltenham, Swindon, Taunton, Exeter, Torbay, Truro

# Role of ACHD CNS?

- 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 >2000 calls pa
- Write patient information
- Education to pts and staff



# 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%**

# PREDISPOSING FACTORS

- Maternal drugs e.g. anti-epileptics, lithium, alcohol
- Chromosomal Aberrations e.g. 1 in 700 Down's Syndrome. 40% D.S. have C.H.D. Turners/Williams Syndrome
- Environmental Factors e.g. Radiation
- Infection/Virus e.g. Rubella
- Maternal Conditions
- e.g. Diabetes



# Lesion information on all lesions on [www.swswchd.co.uk](http://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))

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

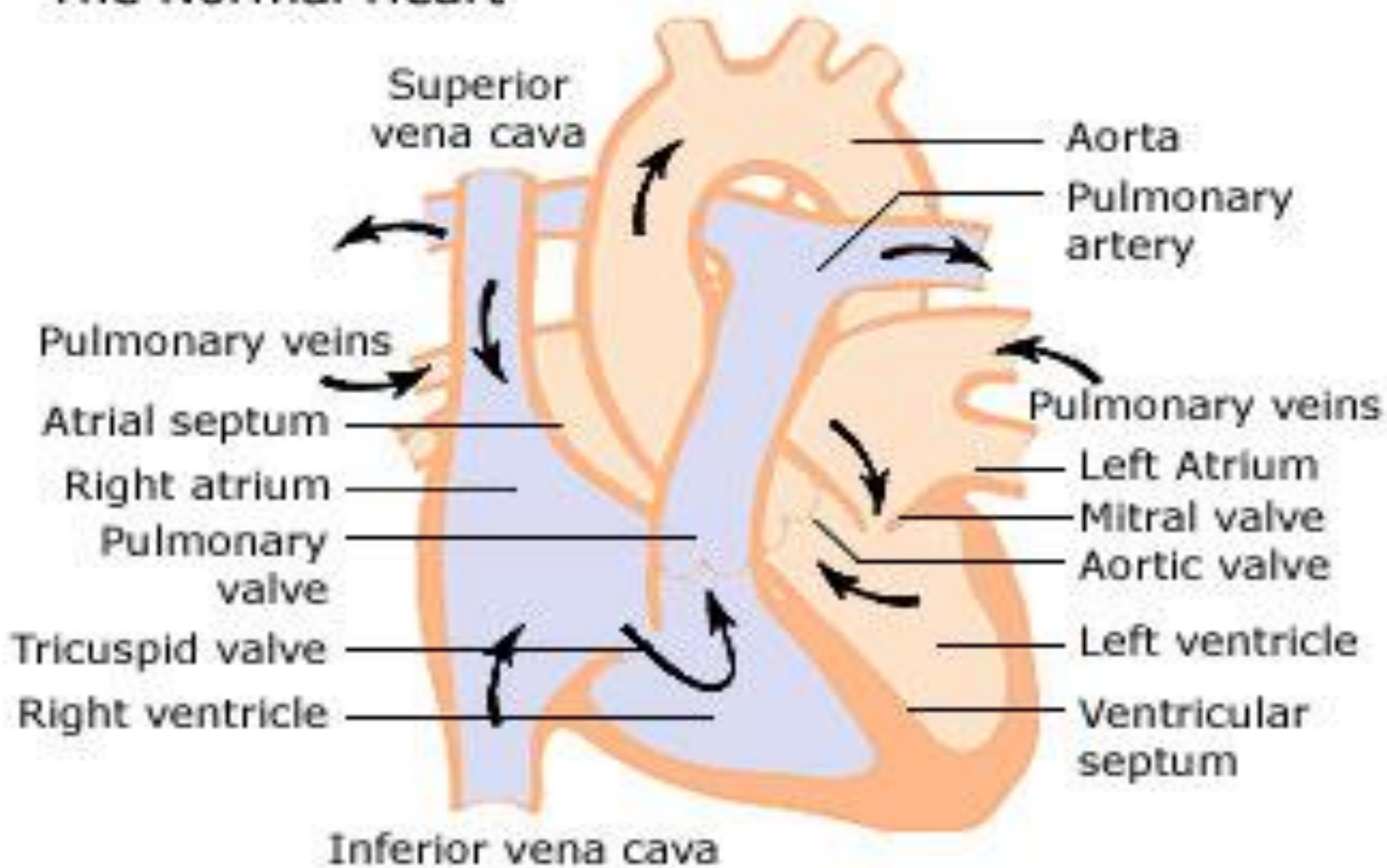
### GUIDANCE

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

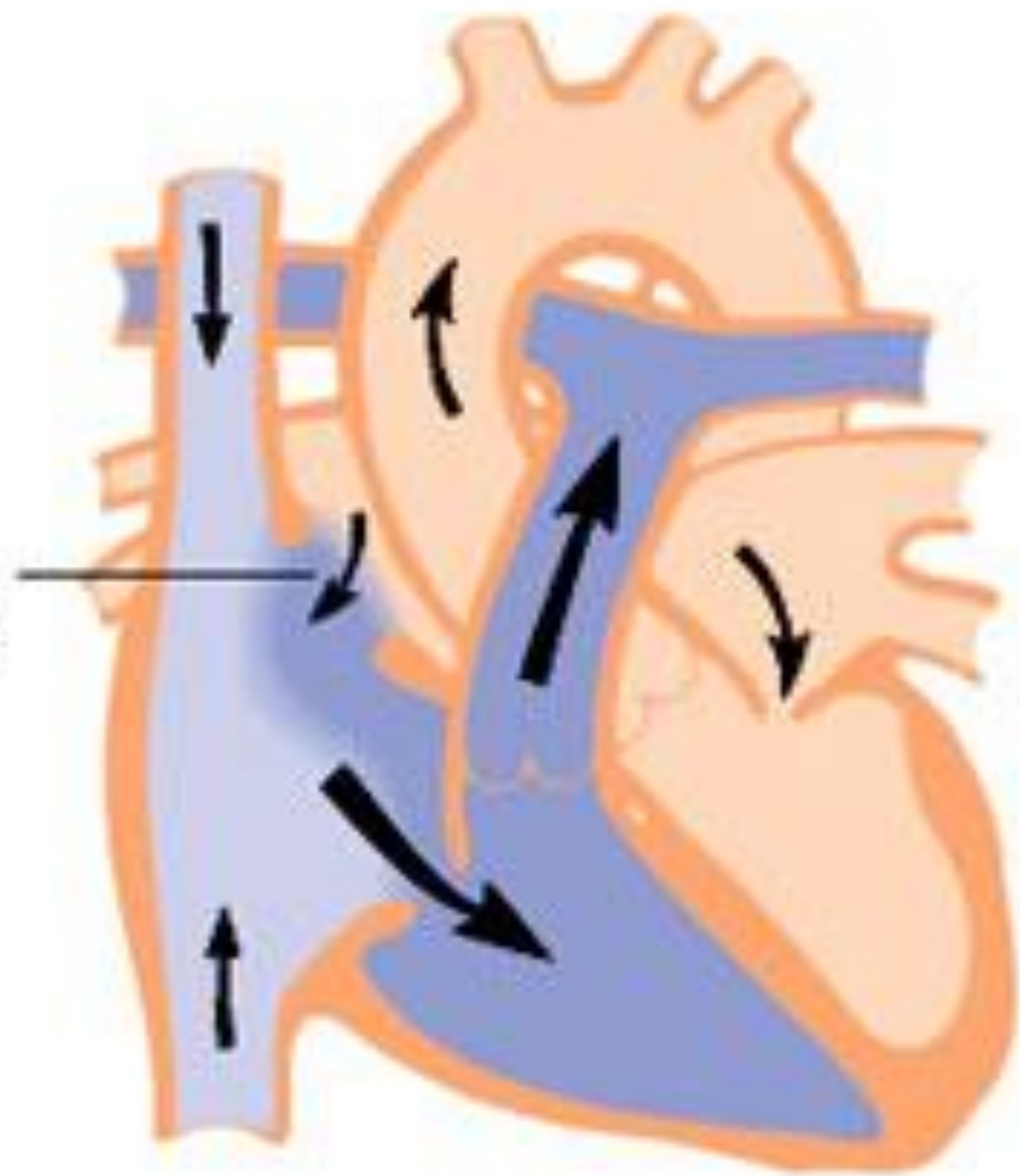
**Long-term complications:**



# The Normal Heart

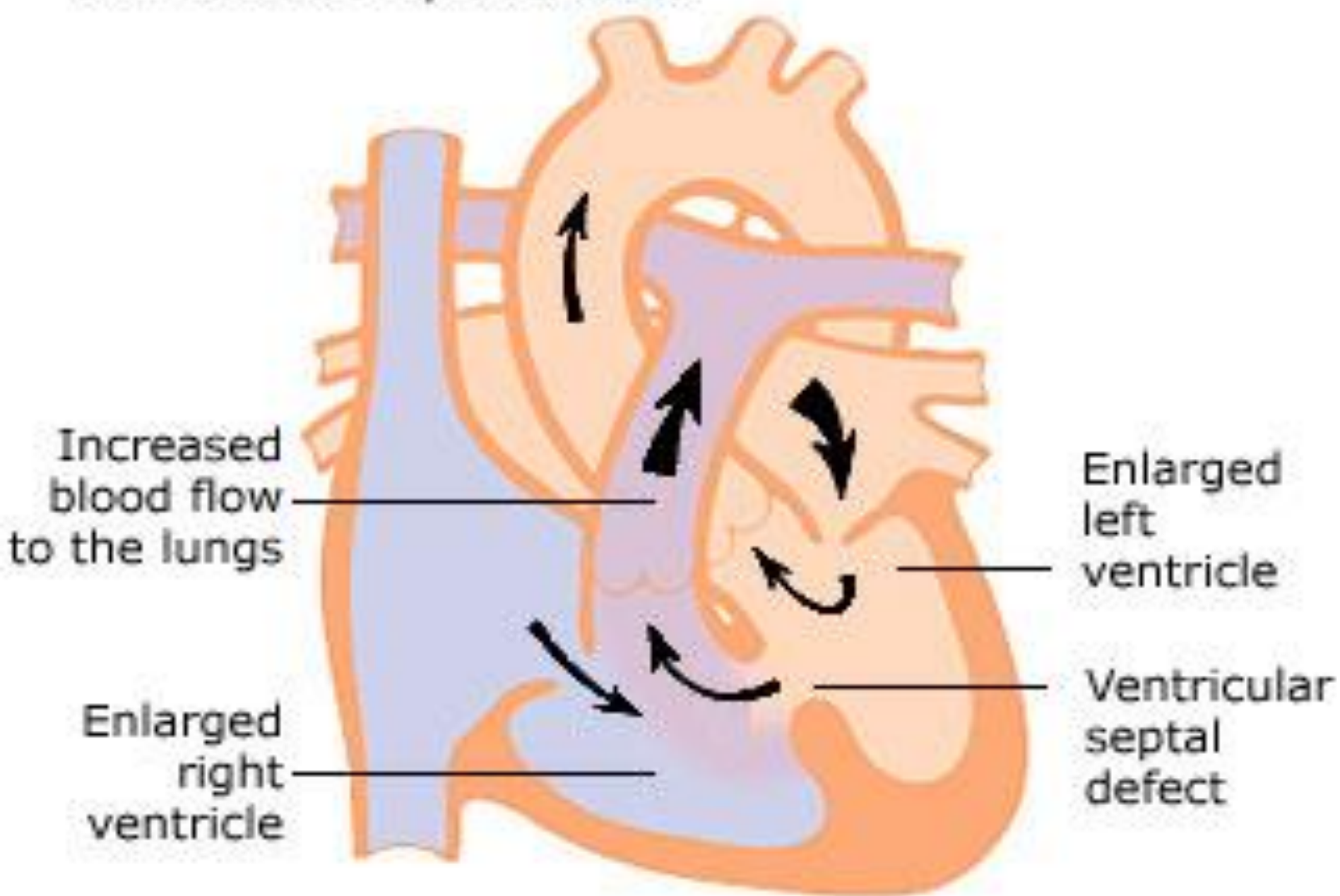


Atrial septal defect

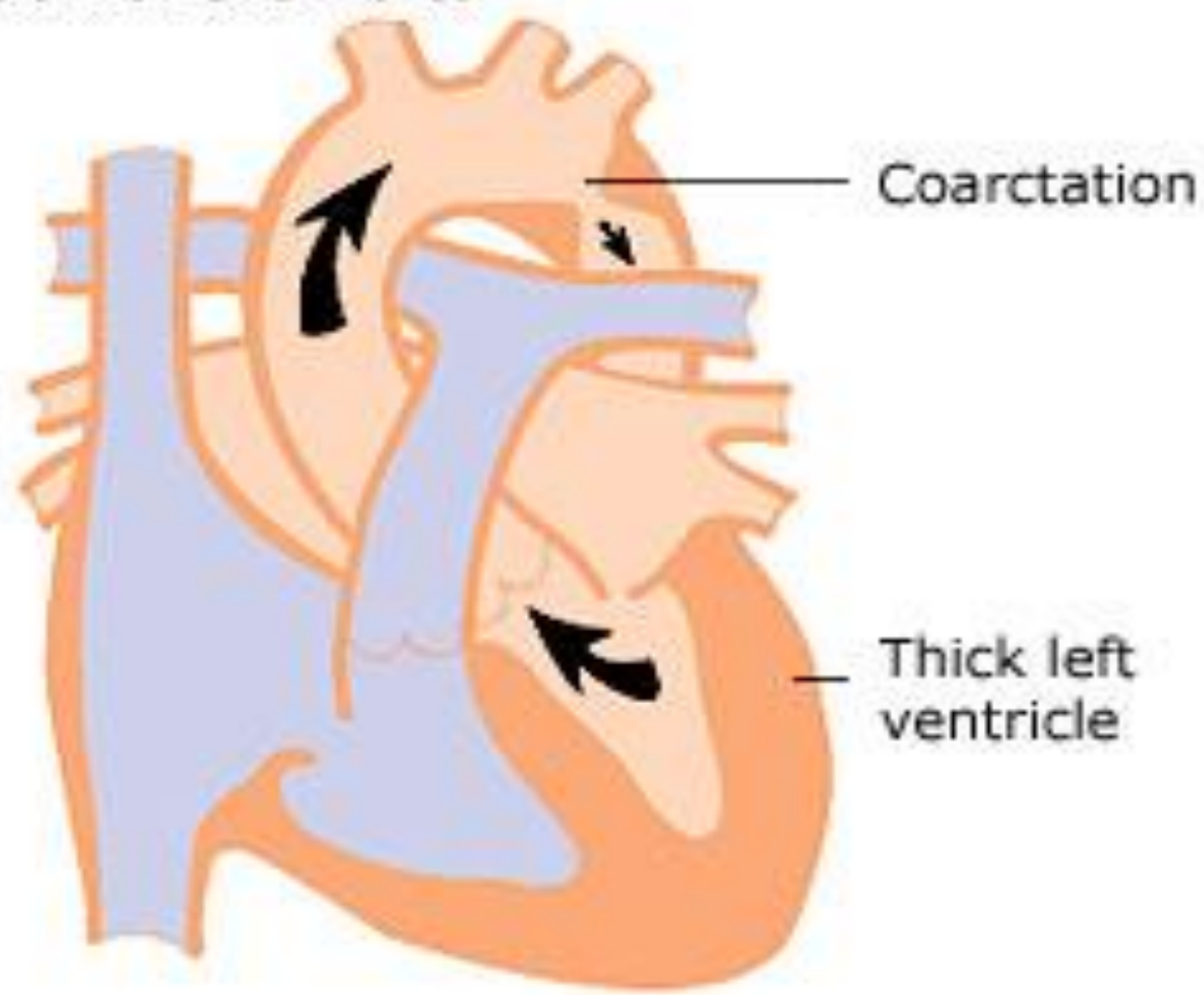




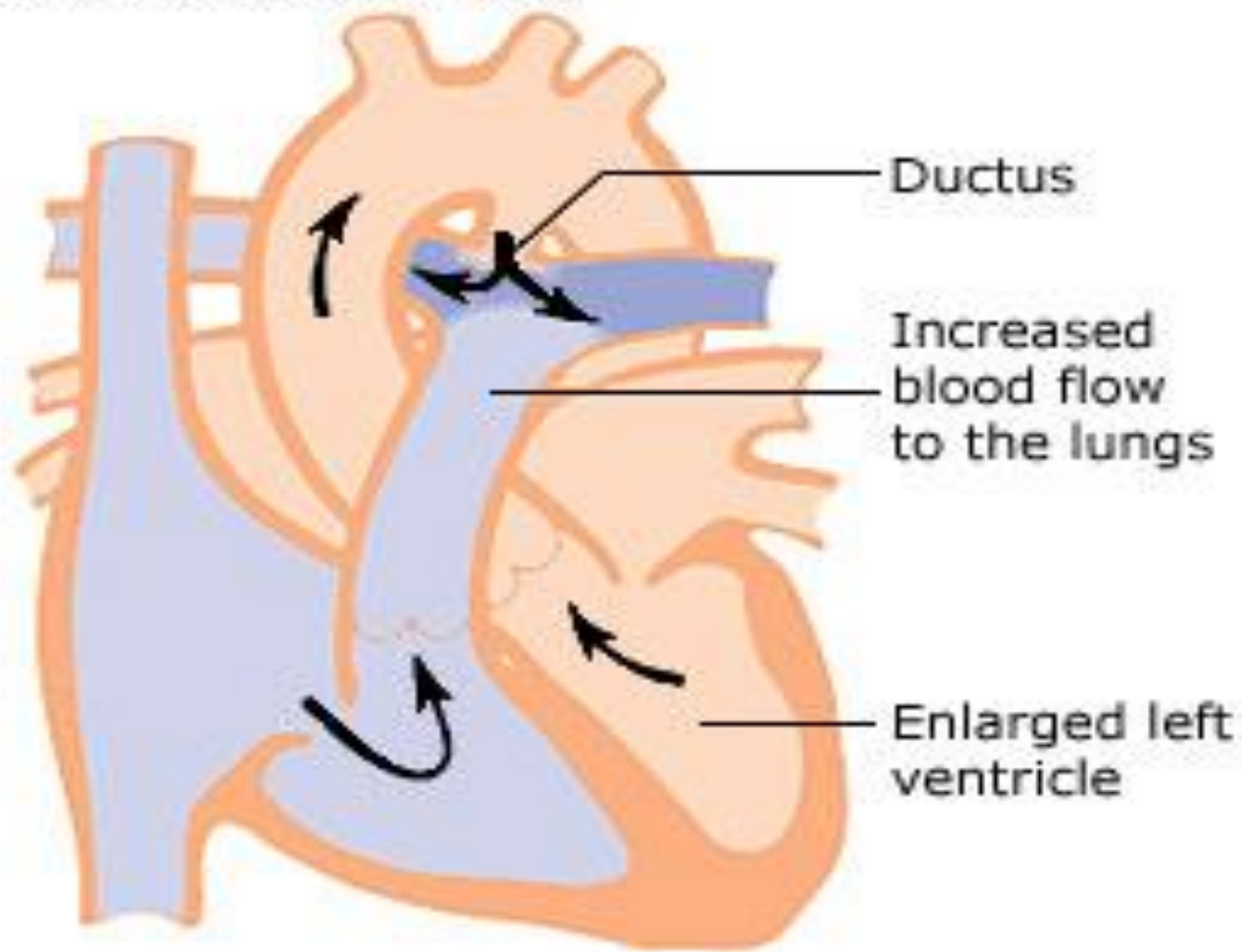
## Ventricular Septal Defect



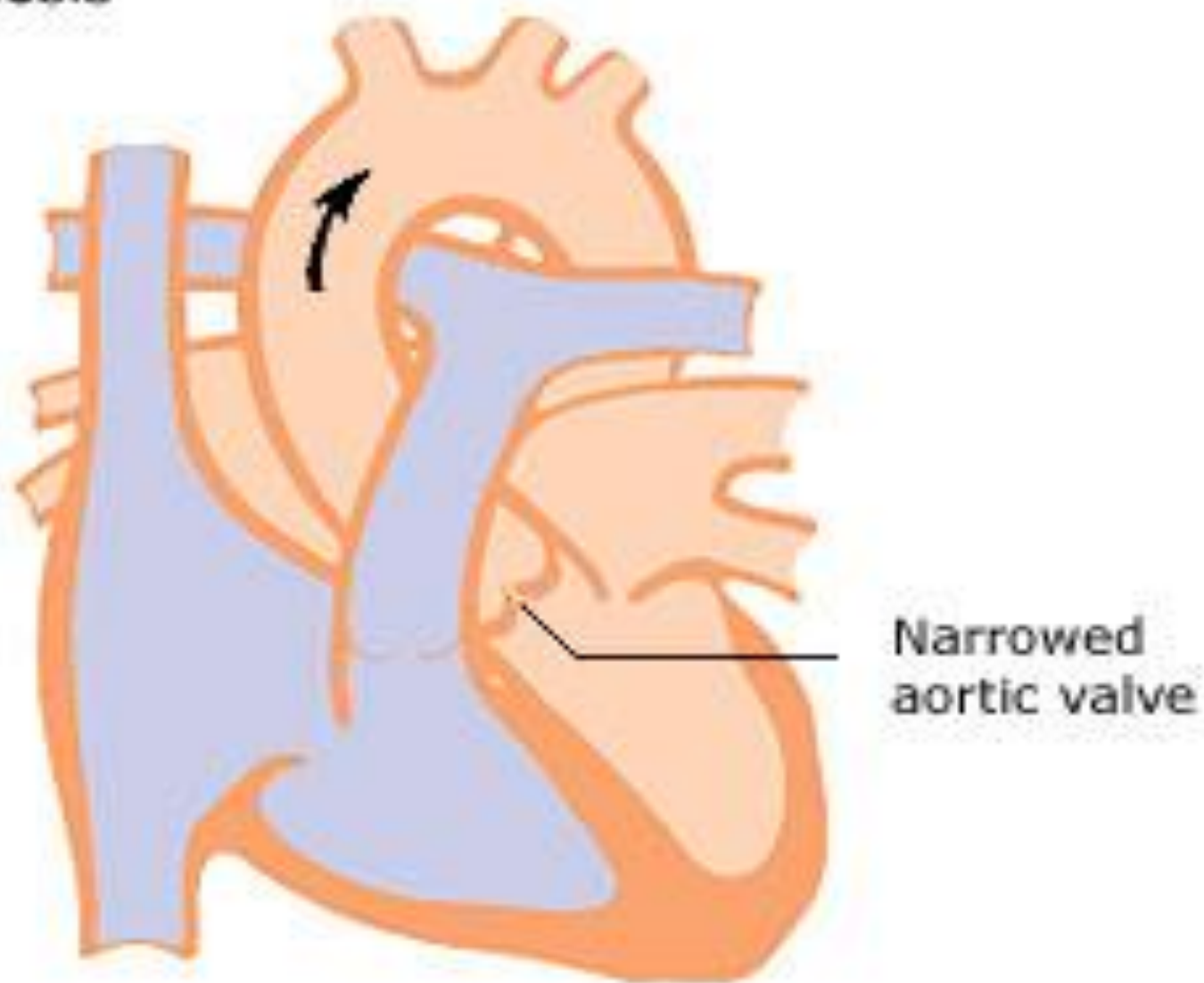
## Coarctation of the Aorta



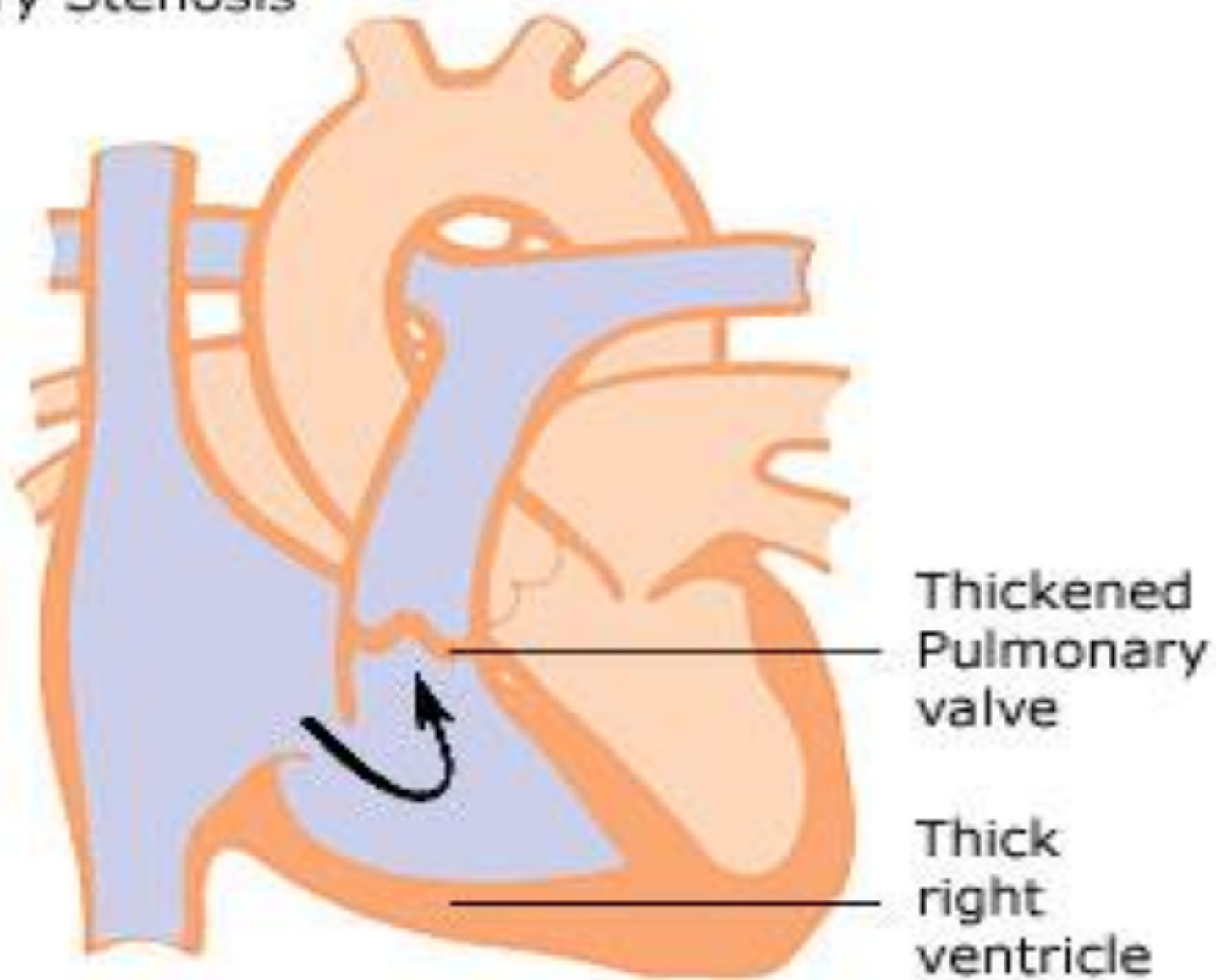
## Persistent Ductus Arteriosus



## Aortic stenosis

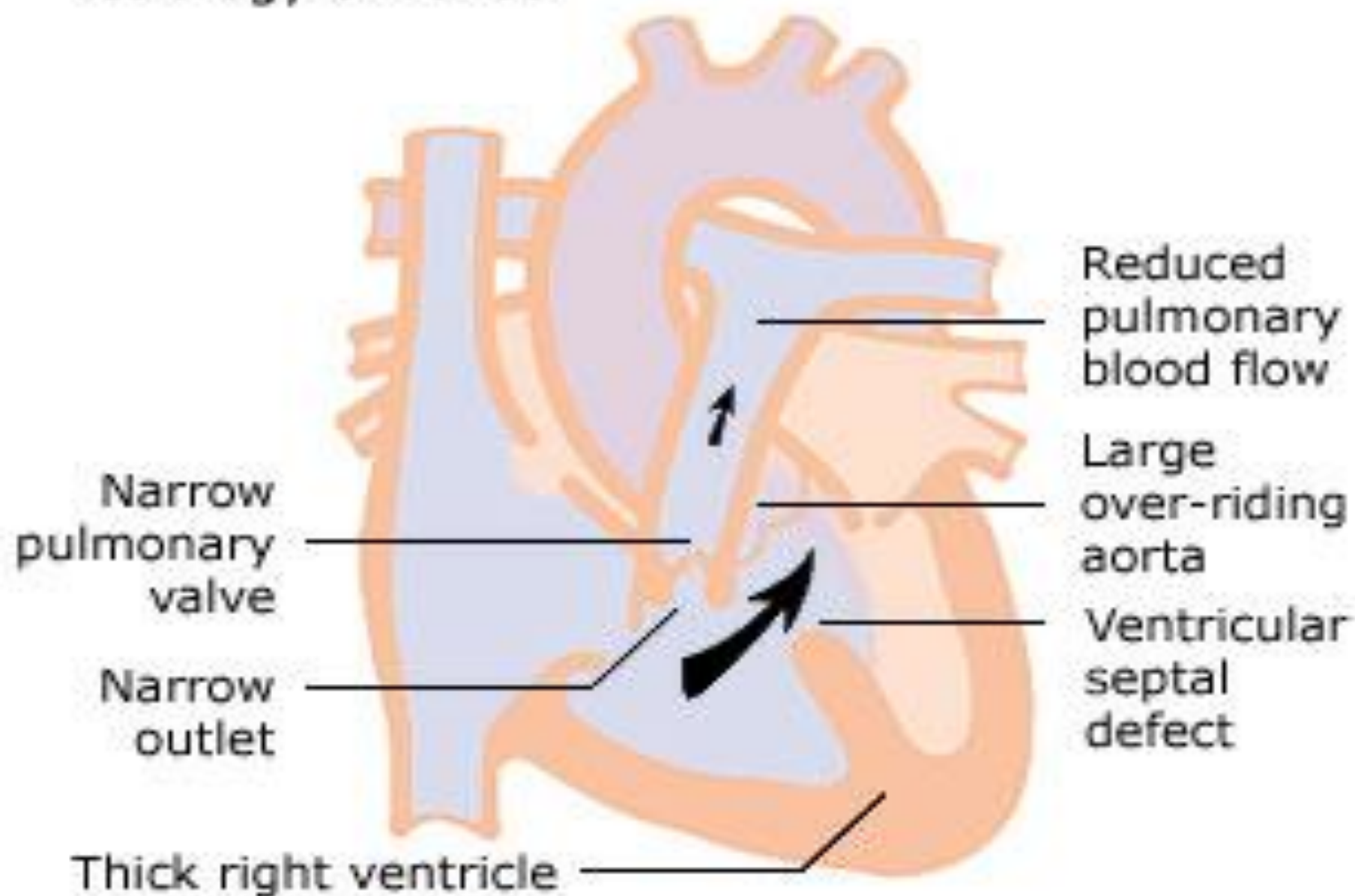


## Pulmonary Stenosis

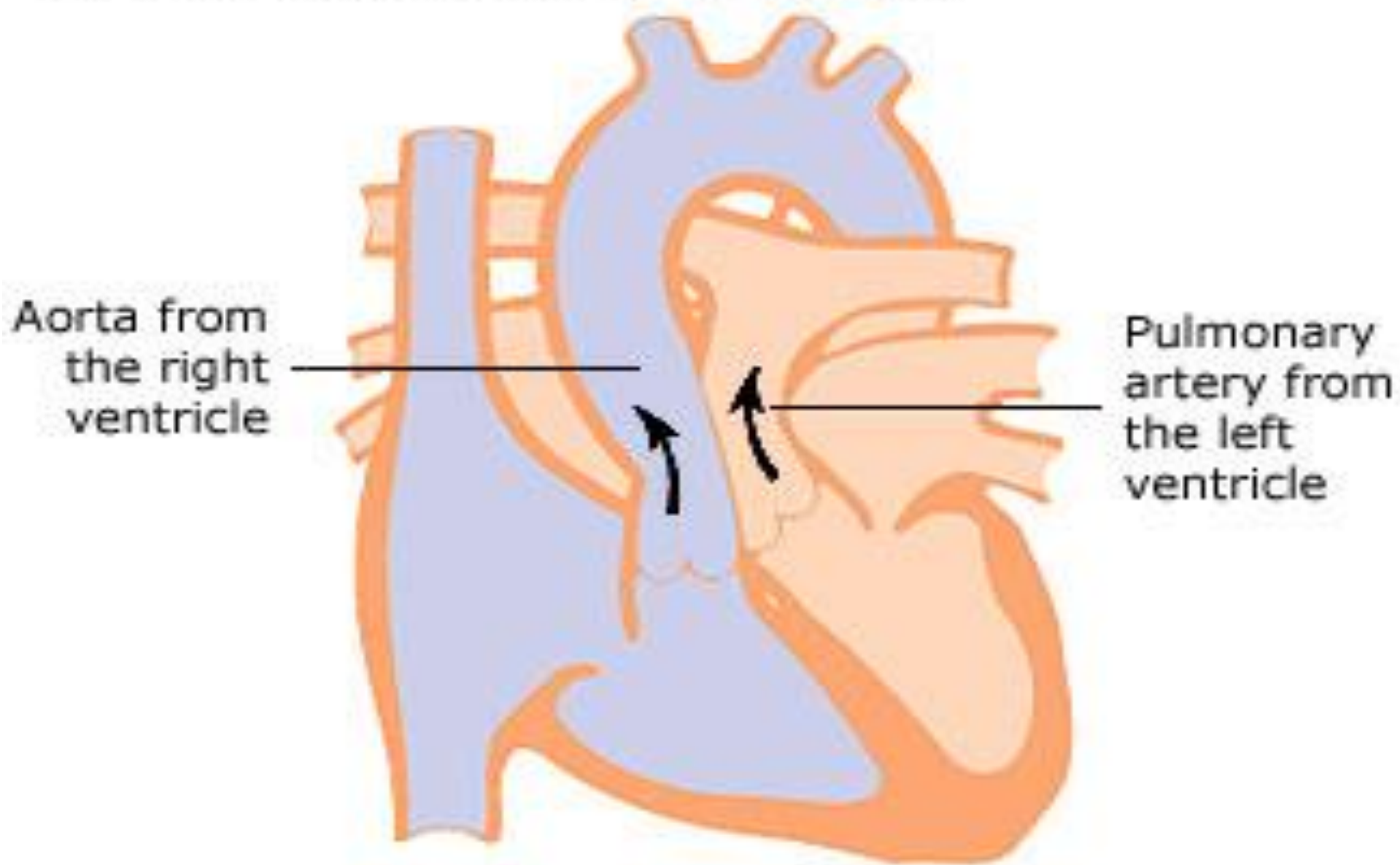




## Tetralogy of Fallot



## Transposition of the Great Arteries



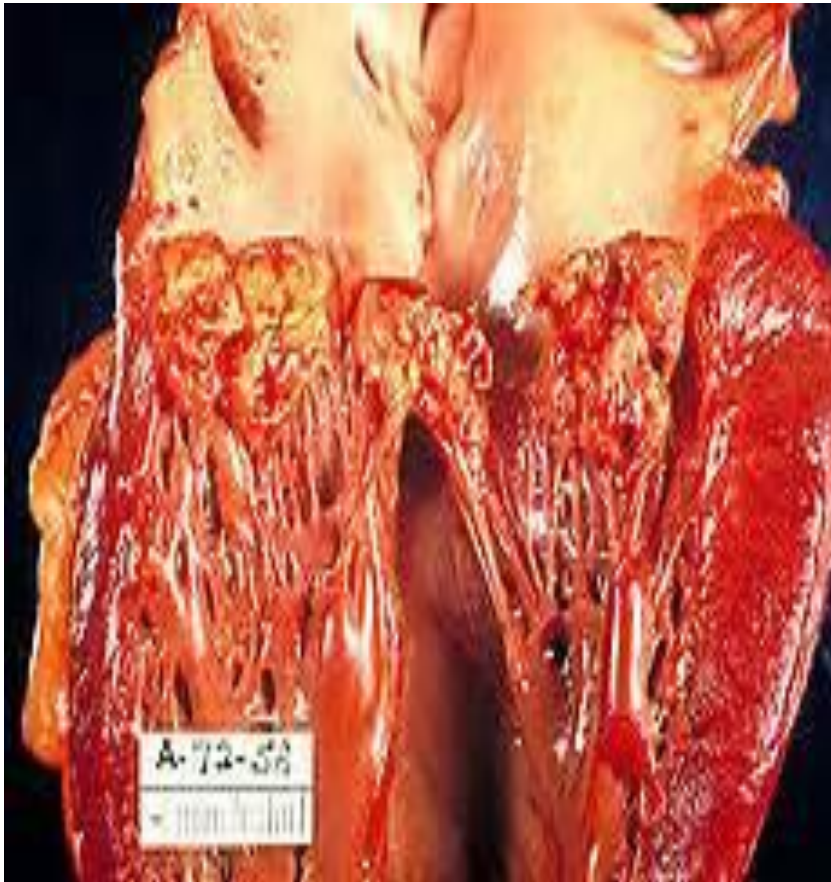
# PHYSICAL ASPECTS OF CARE

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



# INFECTION

## ENDOCARDITIS



- Causes/risk?
- Diagnosis
- Bloods, TOE, ECG+ CXR
- Urine dip
- Treatment
- Complications
- Prophylaxis
- Nursing

# 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.
- Ablation, pacemaker or I.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 ♥ 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



# SURGICAL PROBLEMS

- 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
- 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

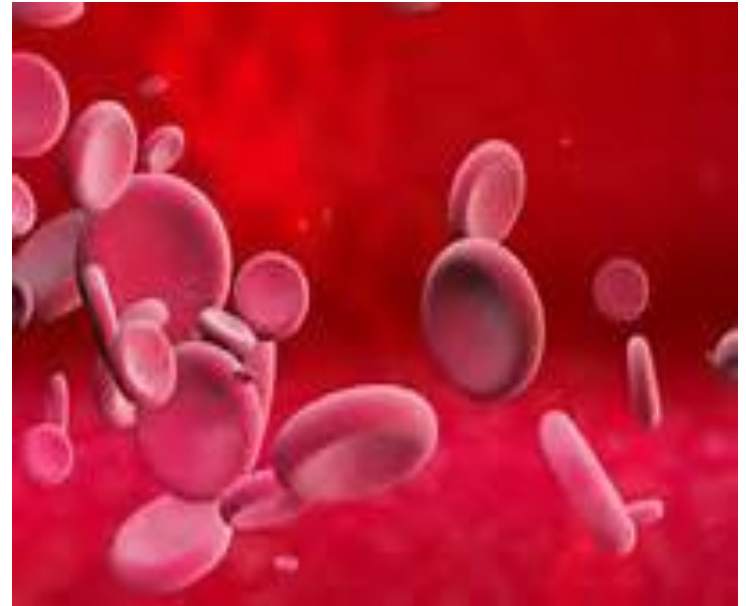
# CYANOSIS



# CYANOSIS

## Watch for.....

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

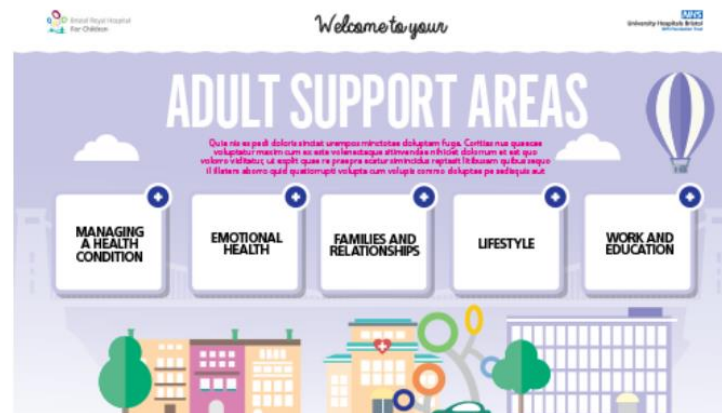


# EMERGENCIES

- 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](http://www.swswchd.co.uk)



Patients & Families

# Support

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

The screenshot shows a web browser window with the URL <https://www.sswchd.co.uk/en/page/patients-families>. The page features the logo for the South Wales and South West Congenital Heart Disease Network. A search bar is present with the text "What are you looking for?". There are also links for "Select Language" and "Resize text: A A A". The main navigation menu includes "Patients & Families", "Professionals", "About", "Hospitals", "Patient Pathways", "Research", "Charities", and "Contact Us". The "Patients & Families" dropdown menu is open, showing options: "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", and "Pregnancy". The background of the page is a dark purple color with a network diagram pattern.

[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](mailto:Sheena.Vernon@uhbristol.nhs.uk)).





# BACCNA

- 'British Adult Congenital Cardiac Nurses Association'



- Twice a year

- Agenda and networking



- BCCA November 19<sup>th</sup>-20<sup>th</sup> Newcastle

# CHD STANDARDS

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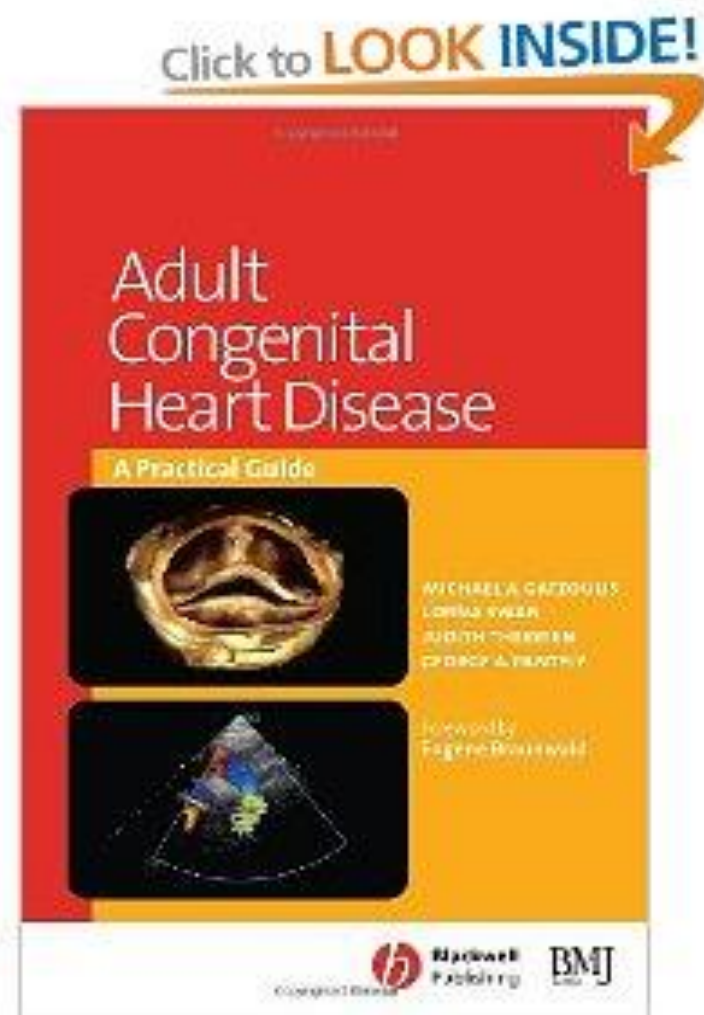
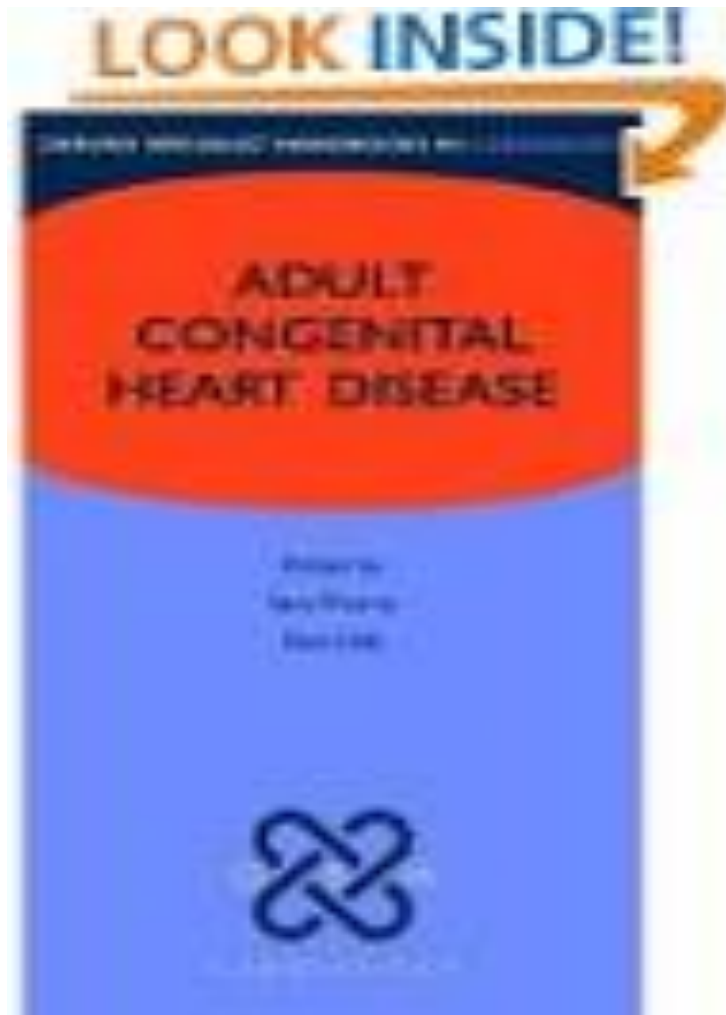


## Adult congenital heart disease nursing

*RCN guidance on roles, career pathways and competence development*

To support adult  
standards  
guidelines from  
RCN for nursing  
published

# READING



# ESC Guidelines

European Heart Journal Advance Access published August 27, 2010



European Heart Journal  
doi:10.1093/eurheartj/ehq319

ESC GUIDELINES



## ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

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The disclosure forms of the authors and reviewers are available on the ESC website [www.escardio.org/guidelines](http://www.escardio.org/guidelines)

ESC entities having participated in the development of this document:

**Associations:** European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA), European Association of Echocardiography (EAE)  
**Councils:** Cardiology Practice, Council on Primary Care, Cardiovascular Imaging, Cardiovascular Nursing and Allied Professions (CCNAP)  
**Working Groups:** Grown-up Congenital Heart Disease, Pulmonary Circulation and Right Ventricular Function, Valvular Heart Disease, Cardiovascular Surgery, Thrombosis, Acute Cardiac Care

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European Heart Journal Advance Access published August 29, 2010



European Heart Journal  
doi:10.1093/eurheartj/ehq319

ESC GUIDELINES



## 2015 ESC Guidelines for the management of infective endocarditis

The Task Force for the Management of Infective Endocarditis of the European Society of Cardiology (ESC)

Endorsed by: European Association for Cardio-Thoracic Surgery (EACTS), the European Association of Nuclear Medicine (EANM)

**Authors/Task Force Members:** Gilbert Habib<sup>a</sup> (Chairperson) (France), Patrizio Lancellotti<sup>a</sup> (co-Chairperson) (Belgium), Manuel J. Antunes (Portugal), Maria Grazia Bongiorno (Italy), Jean-Paul Casalta (France), Francesco Del Zotti (Italy), Raluca Dulgheru (Belgium), Gebrine El Khoury (Belgium), Paola Anna Erba<sup>a</sup> (Italy), Bernard Lheru (France), Jose M. Miro<sup>b</sup> (Spain), Barbara J. Mulder (The Netherlands), Edyta Plonska-Gosciniak (Poland), Susanna Price (UK), Jolien Roos-Hesselink (The Netherlands), Ulrika Snygg-Martin (Sweden), Franck Thuny (France), Pilar Tornos Mas (Spain), Isidre Vilacosta (Spain), and Jose Luis Zamorano (Spain)

**Document Reviewers:** Cetin Erol (CPG Review Coordinator) (Turkey), Petros Nihoyannopoulos (CPG Review Coordinator) (UK), Victor Aboyans (France), Stefan Agevall (Norway), George Athanassopoulos (Greece), Saide Aytekin (Turkey), Werner Benzer (Austria), Hector Bueno (Spain), Lidewij Broekhuizen (The Netherlands), Scipione Careri (Italy), Bernard Cosyns (Belgium), Julie De Backer (Belgium), Michele De Bonis (Italy), Konstantinos Dimopoulos (UK), Erwan Donat (France), Heinz Drexel (Austria), Frank Arnold Flachslampf (Sweden), Roger Hall (UK), Sigrun Halvorsen (Norway), Bruno Heinz<sup>a</sup> (France), Paulus Kirchhof (UK/Germany)

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ESC Committees for Practice Guidelines (CPG) and National Cardiac Societies document reviewers listed in the Appendix

ESC entities having participated in the development of this document:

**ESC Associations:** Acute Cardiovascular Care Association (ACCA), European Association for Cardiovascular Prevention & Rehabilitation (EACPR), European Association of Cardiovascular Imaging (EACVI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA)

**ESC Councils:** Council for Cardiology Practice (CCP), Council on Cardiovascular Nursing and Allied Professions (CCNAP), Council on Cardiovascular Primary Care (CCPC)

**ESC Working Groups:** Cardiovascular Pharmacotherapy, Cardiovascular Surgery, Grown-up Congenital Heart Disease, Myocardial and Pericardial Diseases, Pulmonary Circulation and Right Ventricular Function, Thrombosis, Valvular Heart Disease

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ESC Guidelines for the management of infective endocarditis

# GUIDELINES

- European Society of Cardiology's guidelines on the “Management of Grown Up Congenital Heart Disease” 2010
- The British Cardiac Society Working Party on Grown-up congenital heart disease (GUCH). September 2002.
- The 32nd Bethesda conference: Care of the Adult with Congenital Heart Disease JACC Vol 37, 2001.
- The Canadian Cardiovascular Society's Consensus Conference update 2001 update.



# LOOKING AHEAD

- 
- **Congenital networks**
  - **Support & encourage patients to lead as normal a life as their condition allows.**

**THANK YOU!**

Questions?





# 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](http://www.marfan.org.uk)*



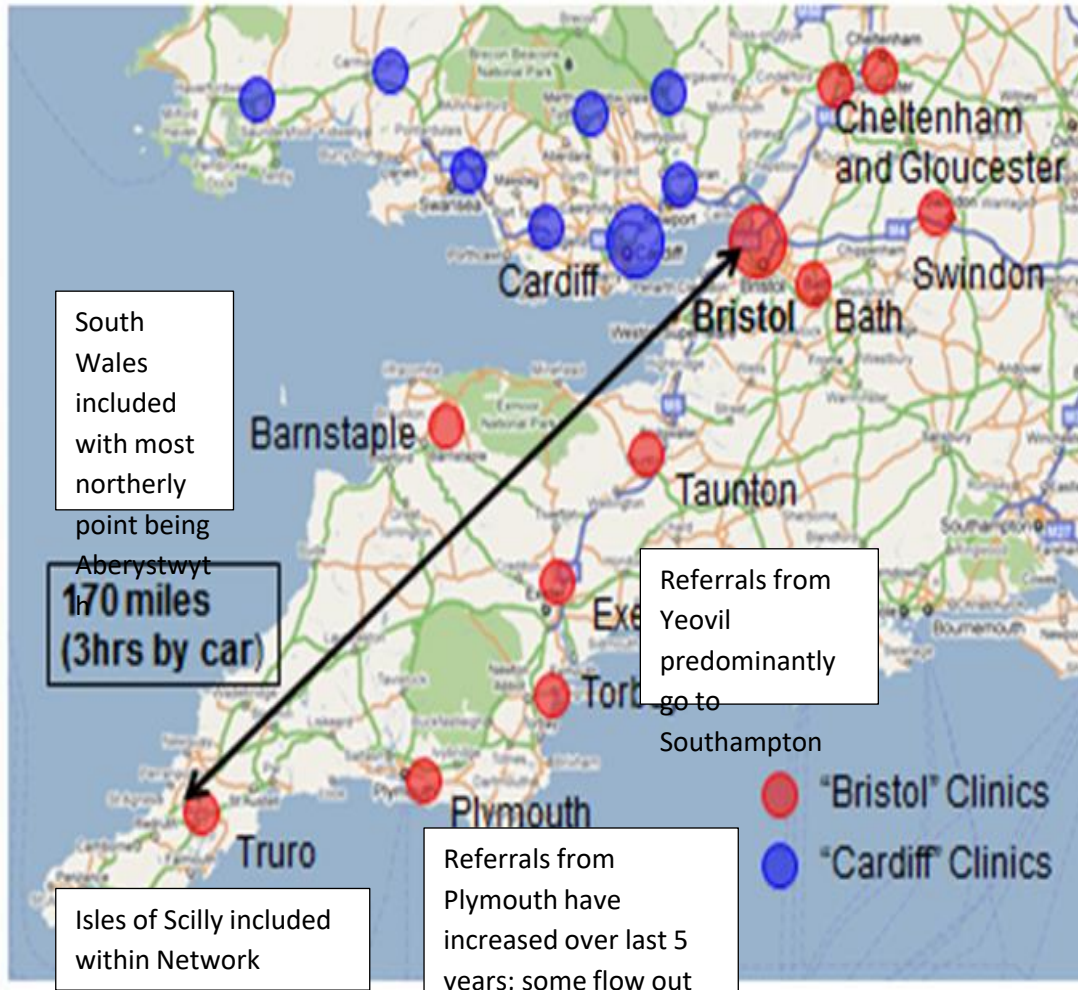
# GUIDELINES



## Adult Congenital Heart Disease

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

# Team & Geography of the Network



**Sheena Vernon,**  
Lead Nurse



**Caitlin Moss**  
Network Manager



**Dr Andrew Tometzki,**  
Clinical Director

Equity of access

Seamless care

Meeting national standards

Continual improvement

of Network to Patient voice

# CHD STANDARDS

- 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](#)



# THE NETWORK APPROACH

**sets out:** how networks will work

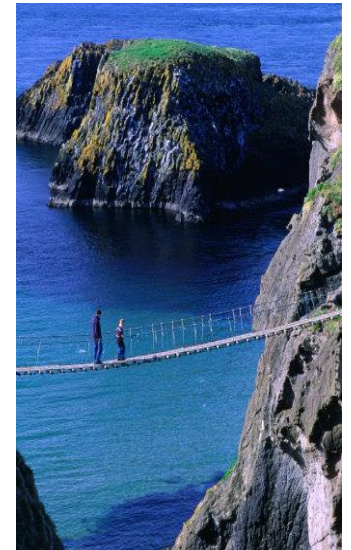
**new/changing:** clear leadership (clinical and professional);  
cardiology (non-surgical) centres' participation in networks;  
second opinions and referrals

- **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.



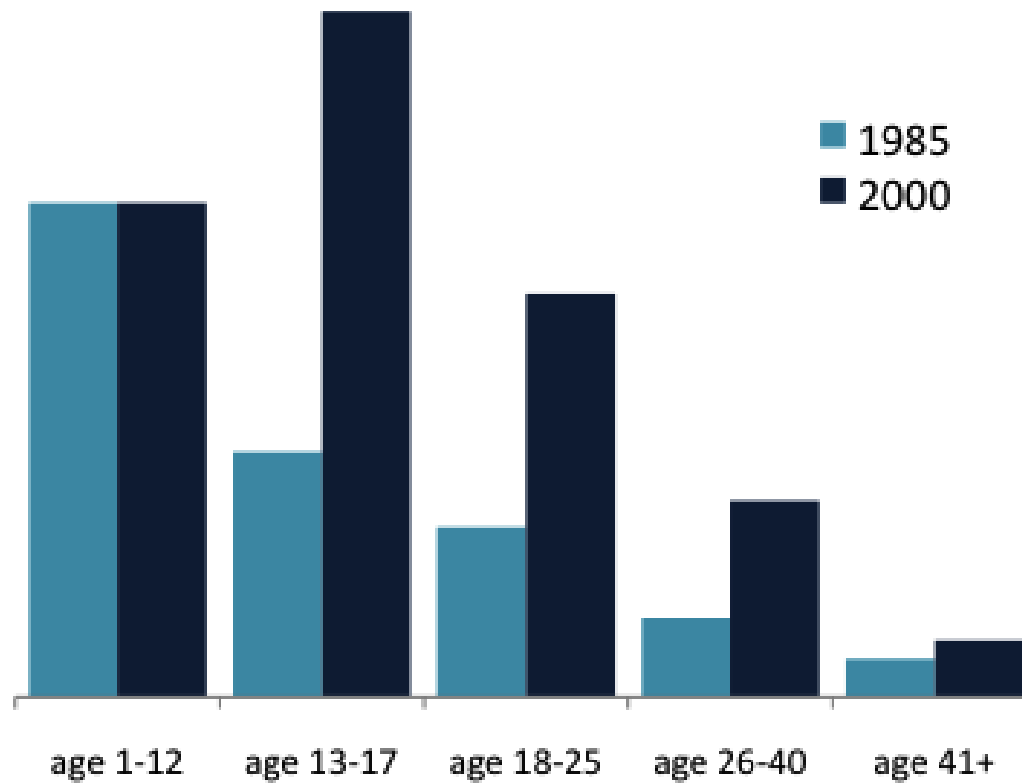
# PALLIATIVE CARE AND BEREAVEMENT

**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

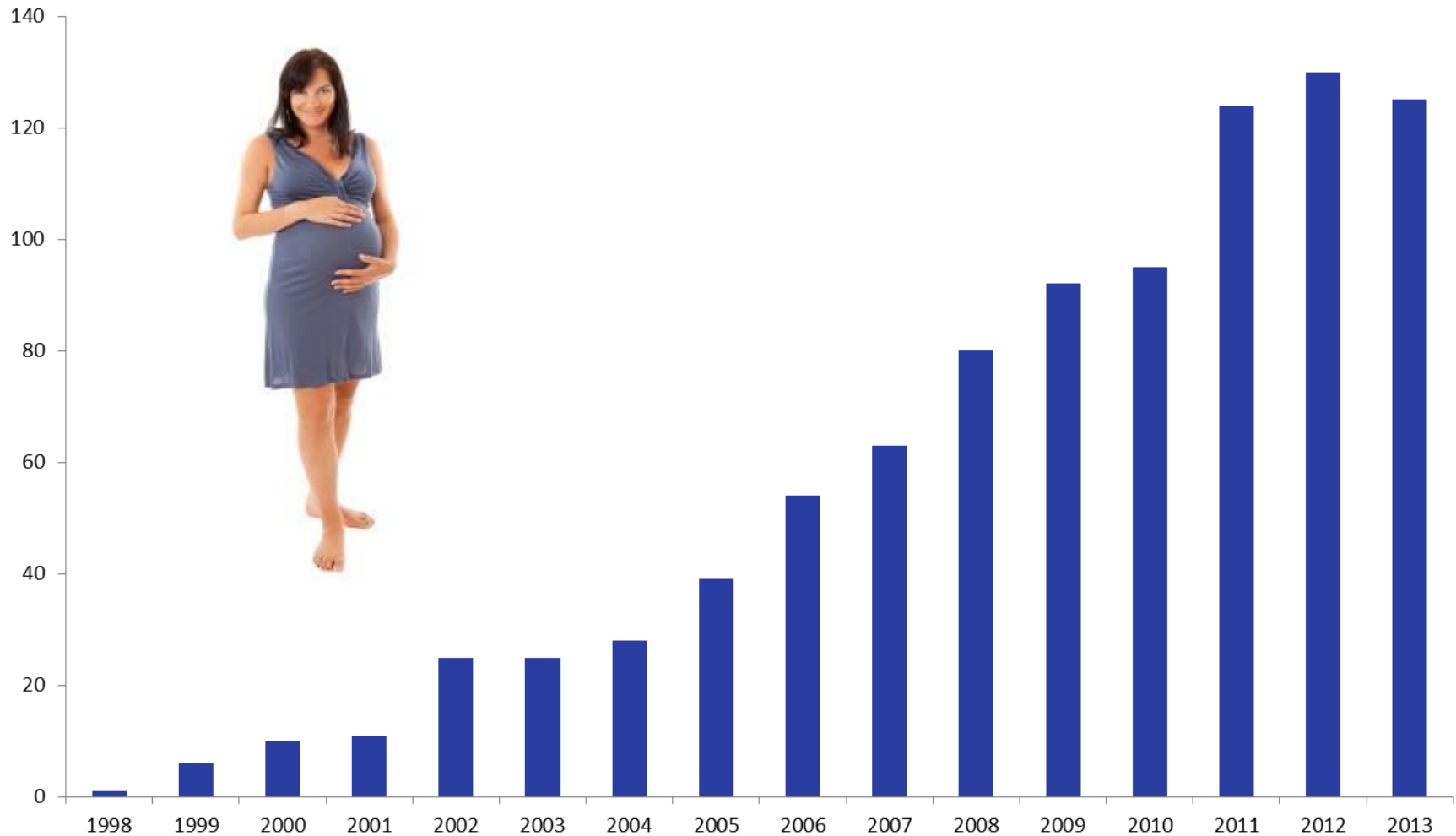
- **Challenge** : difficult conversations, patient, parents spouse, family and children
- Intense telephone advice
- End of life pathway
- Palliative care teams
- GP support



# 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

The  
Somerville  
Foundation

The logo for The Somerville Foundation features the word "Somerville" in a large, black, serif font. The letter "o" is replaced by a stylized heart shape, with the left half of the heart in blue and the right half in red. Above the "merville" part of "Somerville" is the word "The" in a smaller, black, serif font. Below "Somerville" is the word "Foundation" in a black, serif font.

# CoaguChek machines



- INR test
- [www.roche-diagnostic.co.uk](http://www.roche-diagnostic.co.uk)
- [www.coagucheck.co.uk](http://www.coagucheck.co.uk)

# Advice Line





# 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.

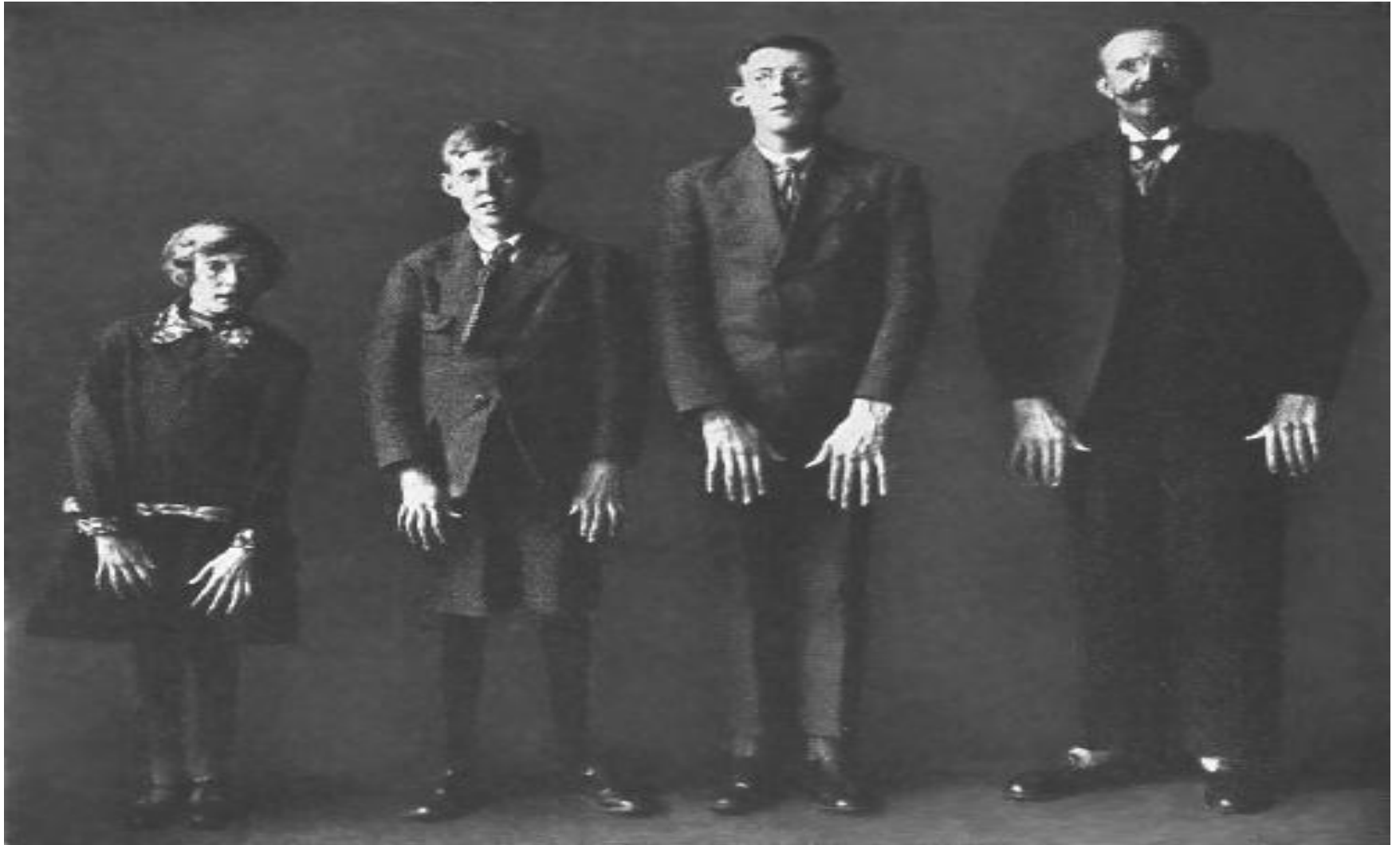
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# Piercings

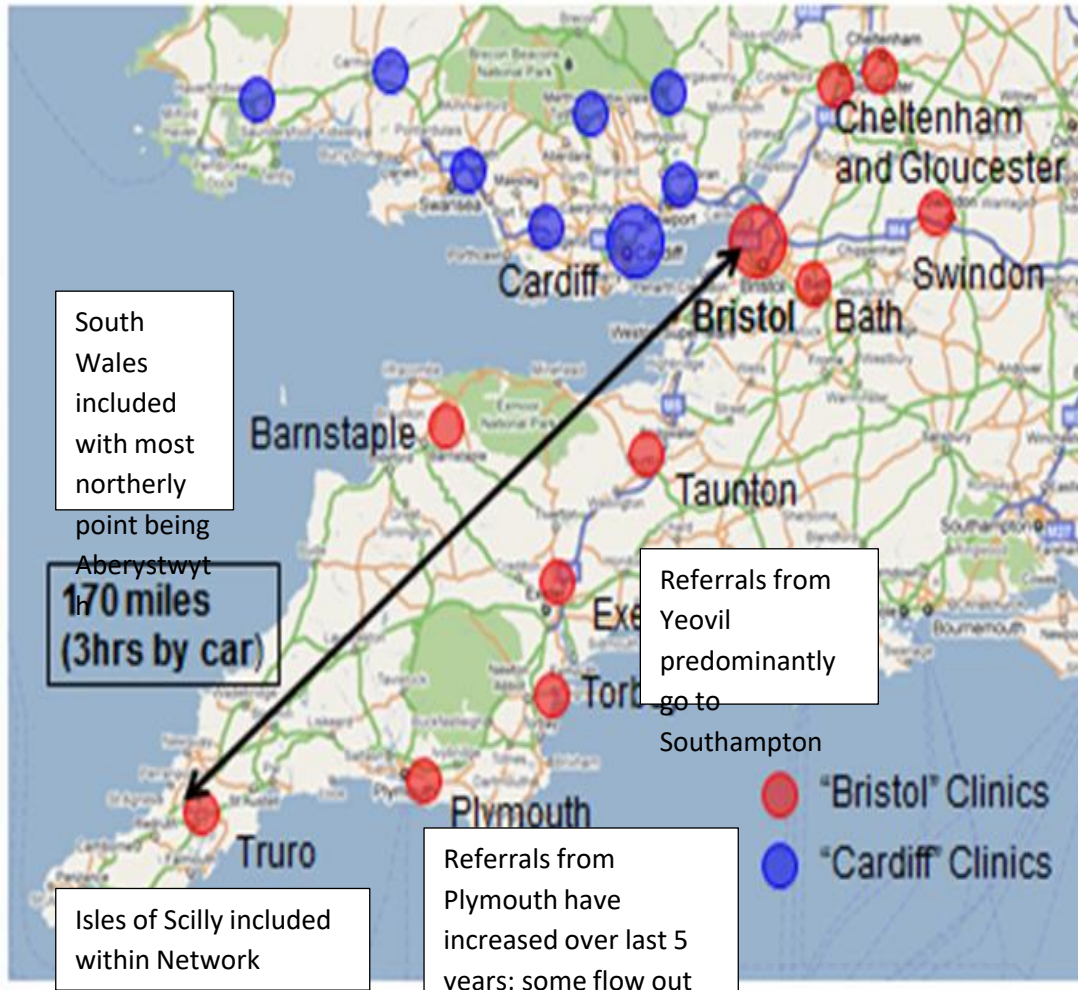


# MARFANS SYNDROME





# Team & Geography of the Network



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Lead Nurse



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Equity of access

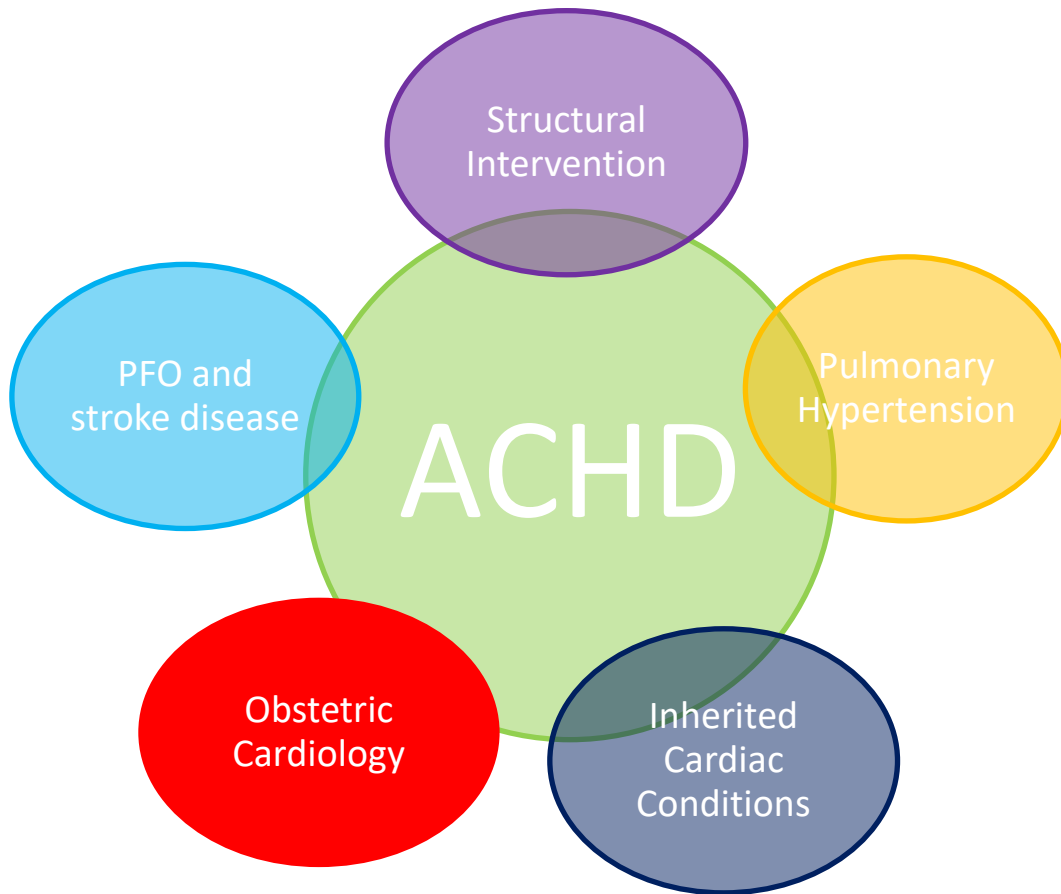
Seamless care

Meeting national standards

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# Learning Disabilities

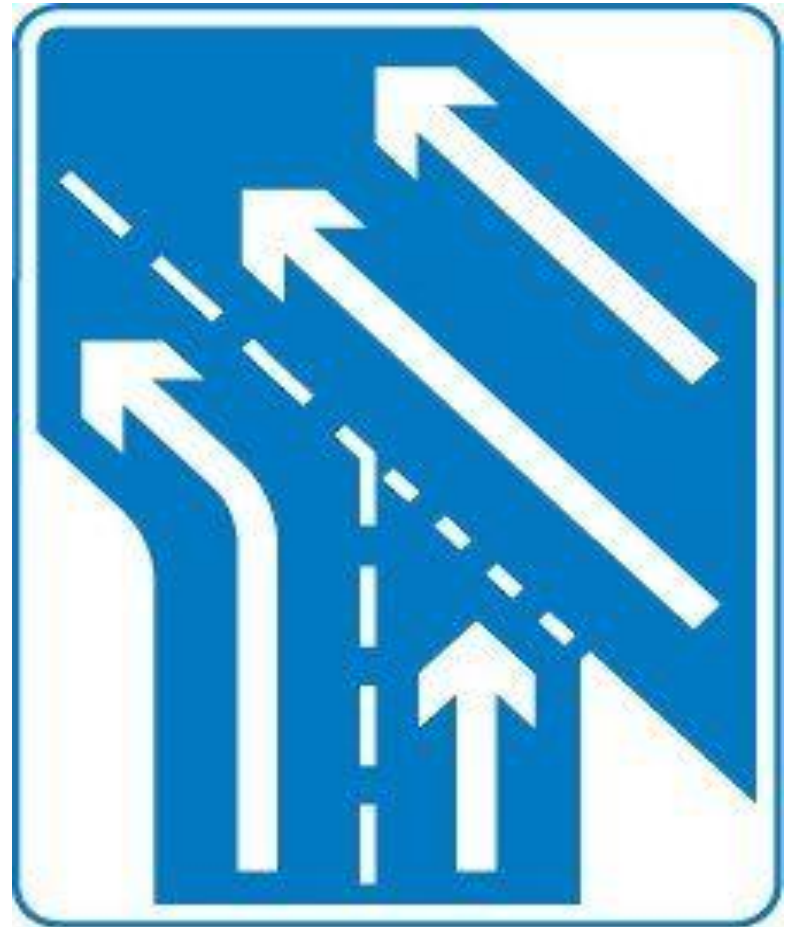
- Increasing numbers of patients having procedures and treatment
- 1 in 700 born with Downs, 40% will have CHD
- Time consuming
- Support for patient, family, CLDT and carers
- Capacity to consent? Best interest meetings?
- Appropriate communication

# 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

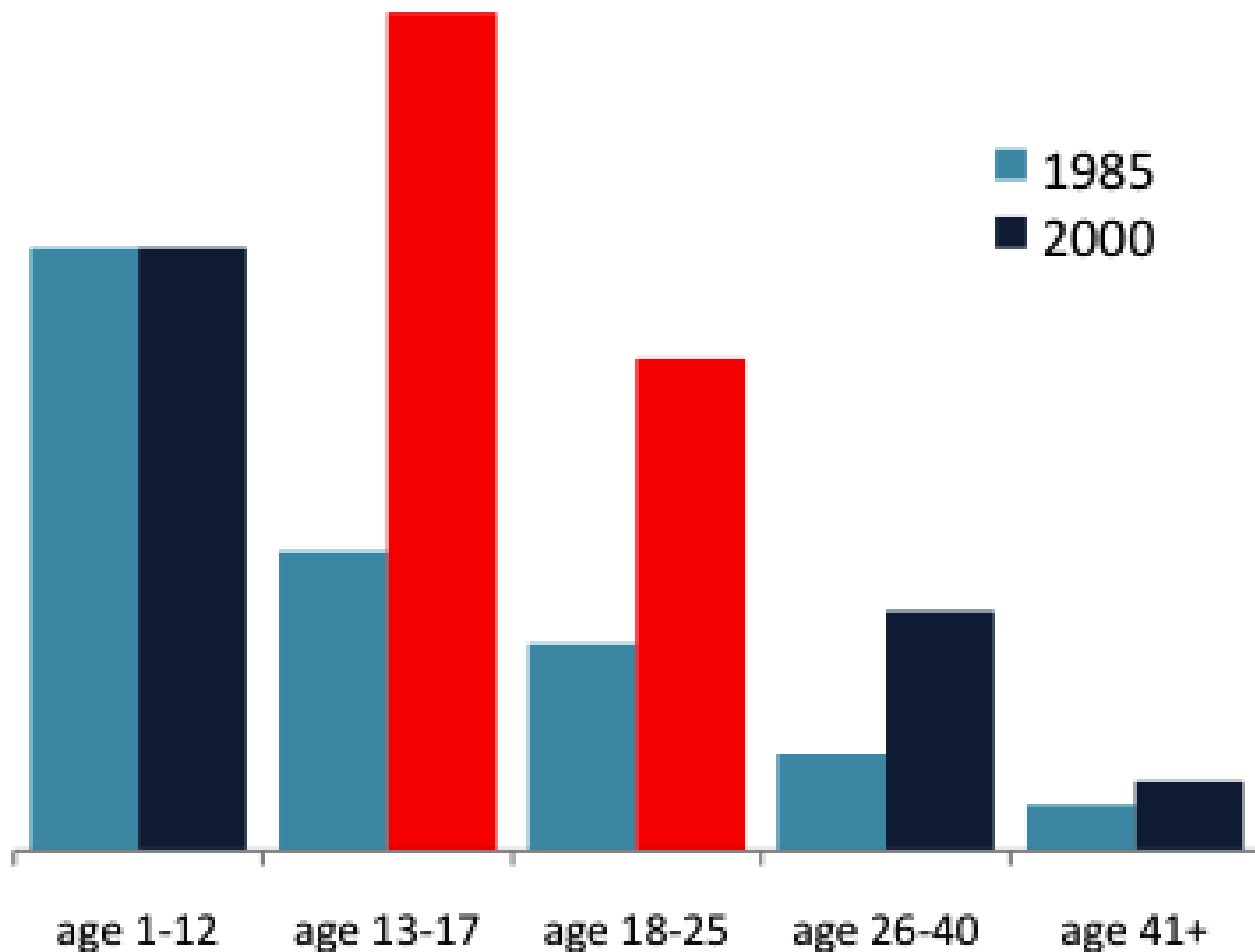


# ADVICE LINE





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



# 2007-2014

