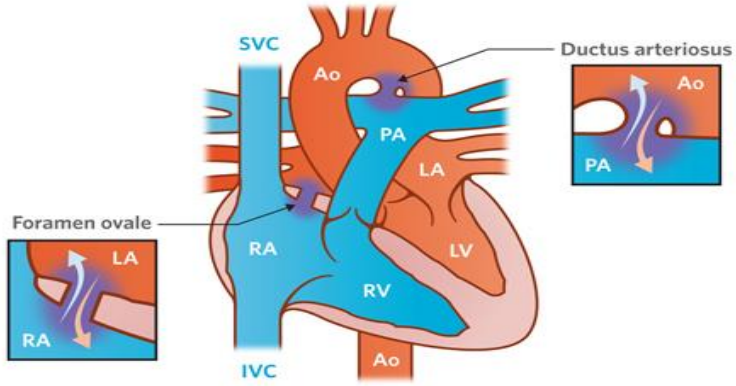
Congenital Heart Disease
On NICU
And the CHD Network



Rachel Sampson
Cardiac link nurse

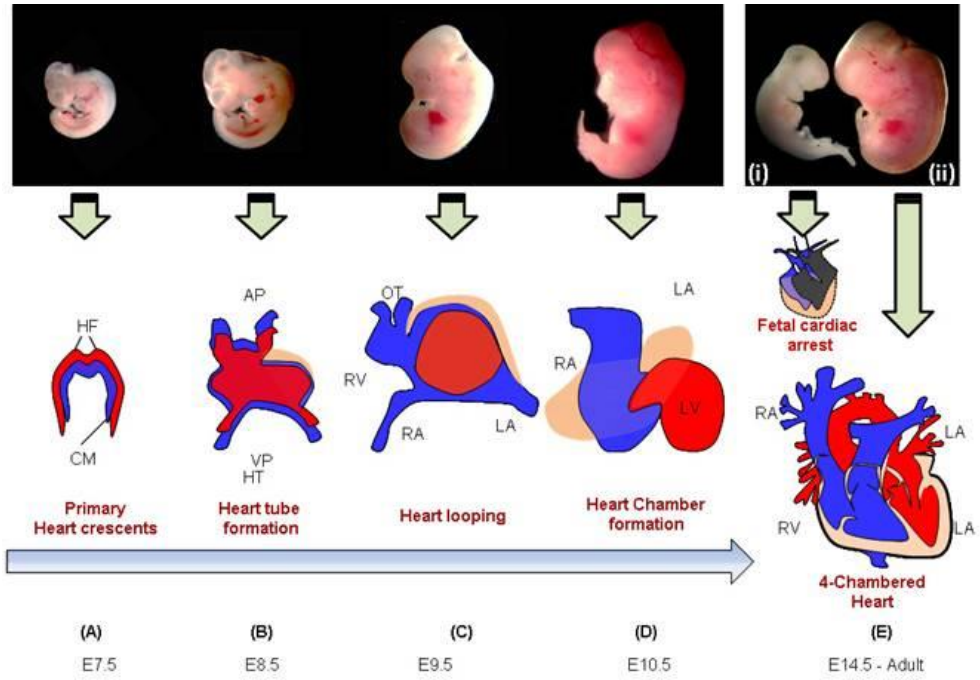
The role of the cardiac link nurse

Link Nurses are defined as qualified nurses, working in clinical areas within the Network who take responsibility for participating in the care of a patient (child or adult) with congenital heart disease. They will develop relationships with staff in the specialist centres, with the aim of improving communication channels and facilitating education of other staff within their area of work.

Link nurses are not specialist nurses but are nurses with a keen interest in patients with cardiac conditions

Congenital heart disease means that there is an abnormality of the heart which has been present since birth. It occurs in about 8 of every 1,000 babies born alive. About a third of these children have mild heart abnormalities and never require any treatment.

The heart is the first organ formed during organogenesis



Congenital heart disease occurs during the first 8 weeks of fetal development

CONGENITAL HEART DISEASE

tiny tickers
a better start for tiny hearts

CHD is when something goes wrong when a baby's heart is formed...

A baby is born in the UK with CHD roughly every 2 hours

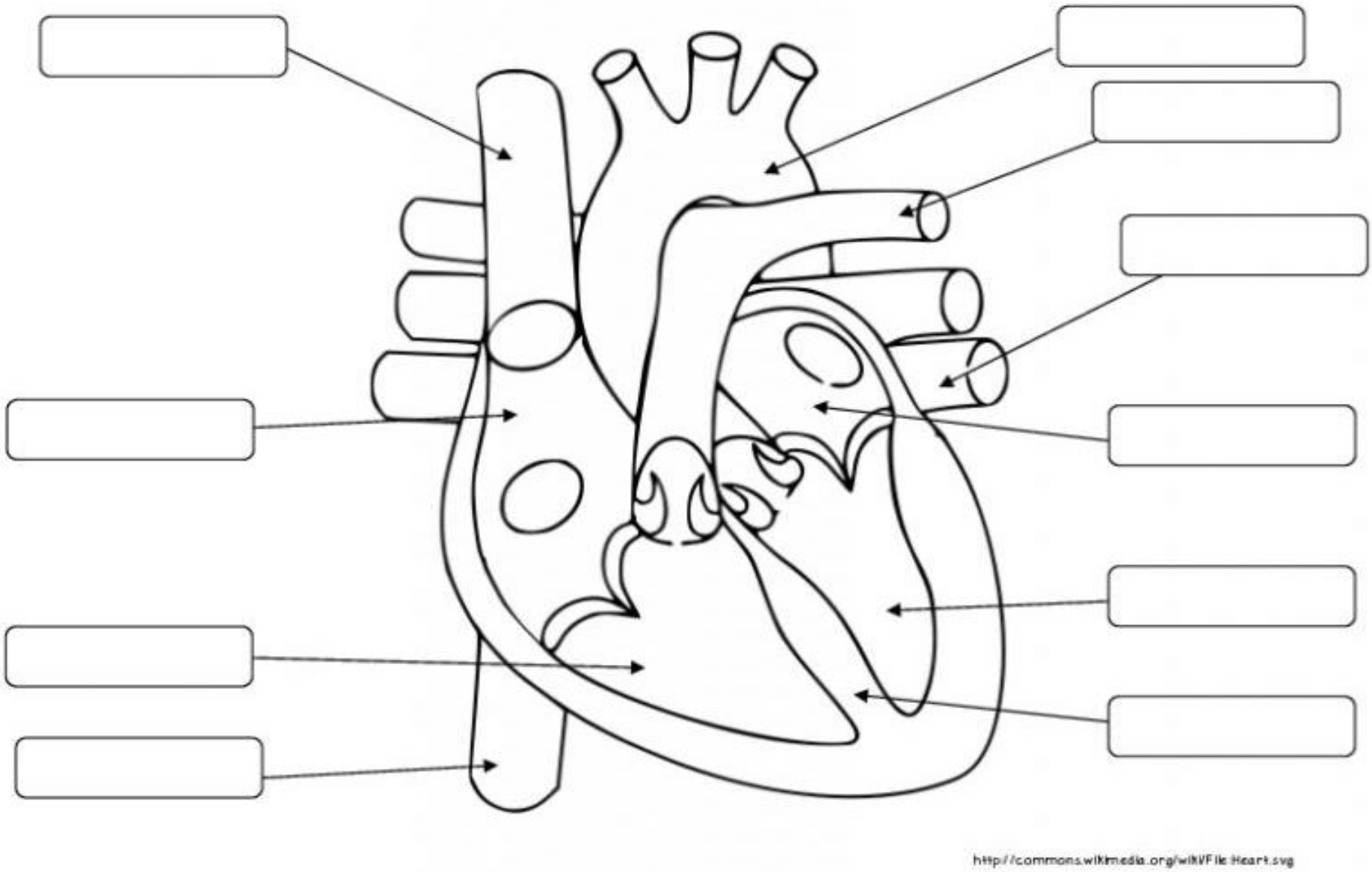
Up to 1 in every 125 babies has some sort of heart defect

CHD is the most common congenital birth defect and causes up to 1 in 13 infant deaths

85% of babies born with CHD now survive into adulthood - that's more than ever before

Surgical survival rates in the UK are some of the best in the world - nearly 100% for some procedures

On average across the UK, nearly 50% of CHDs are spotted during pregnancy


http://commons.wikimedia.org/wiki/File:Heart.svg

SVC = superior vena cava
RA = right atrium
TV = tricuspid valve
RV = right ventricle
IVC = inferior vena cava
PV = pulmonary valve
PA = pulmonary artery
PVn = pulmonary vein
A = aorta
LA = left atrium
MV = mitral valve
LV = left ventricle
AV = aortic valve
S = **septum**

Key:

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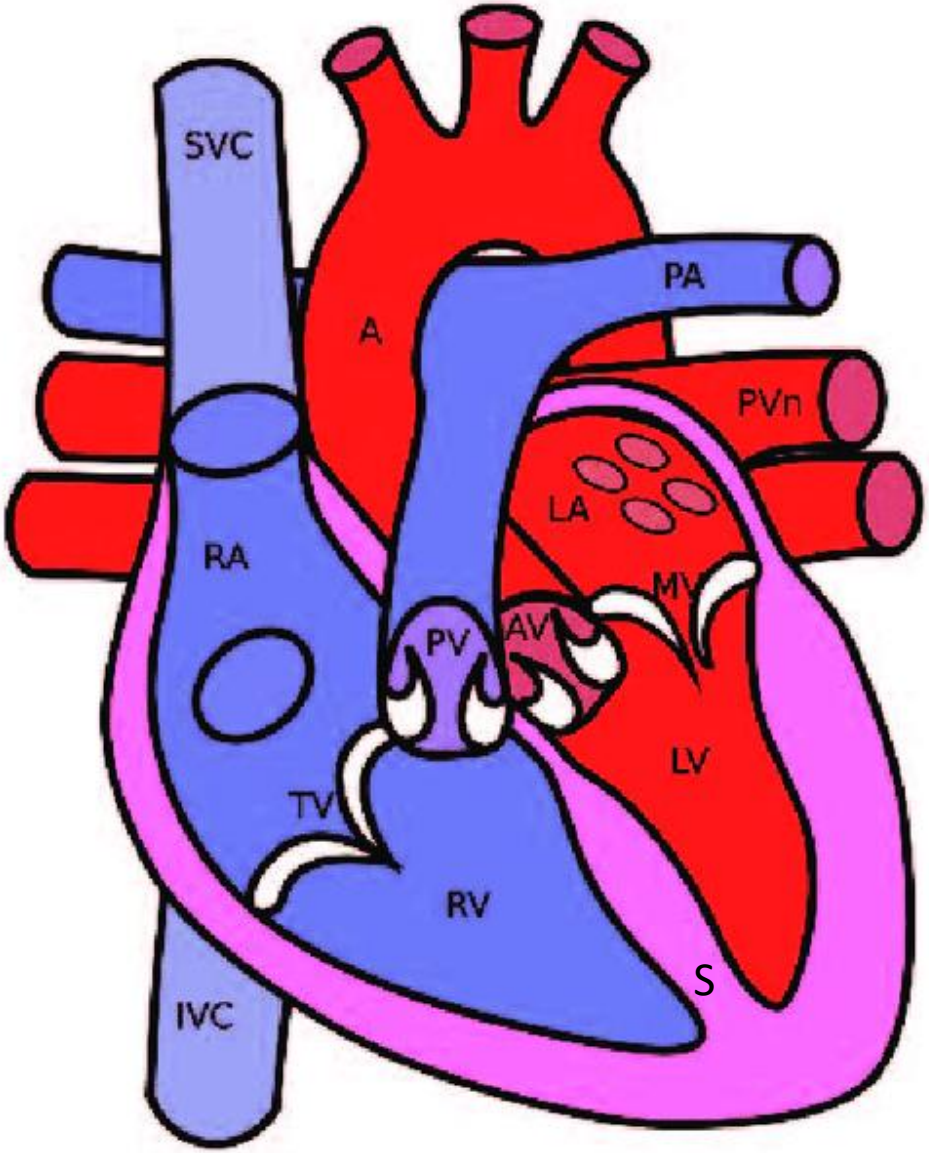


Adapted from a diagram by Dake and Eric Pearce

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%**

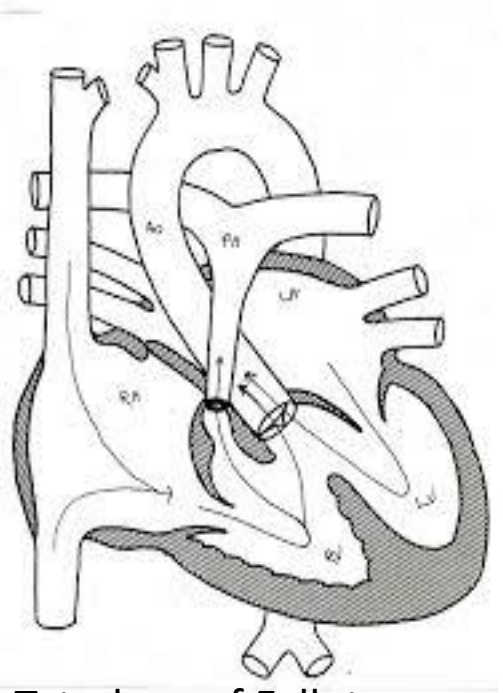
www.bhf.org

ESCcardiology

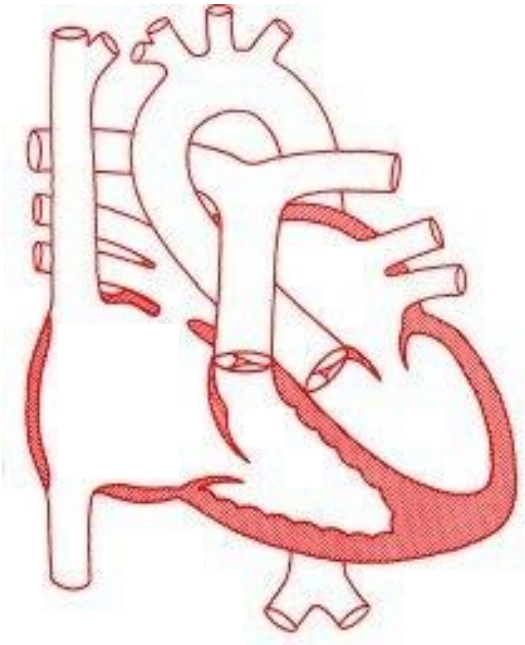
Can you guess the four common lesions?

Please colour the blood flow.

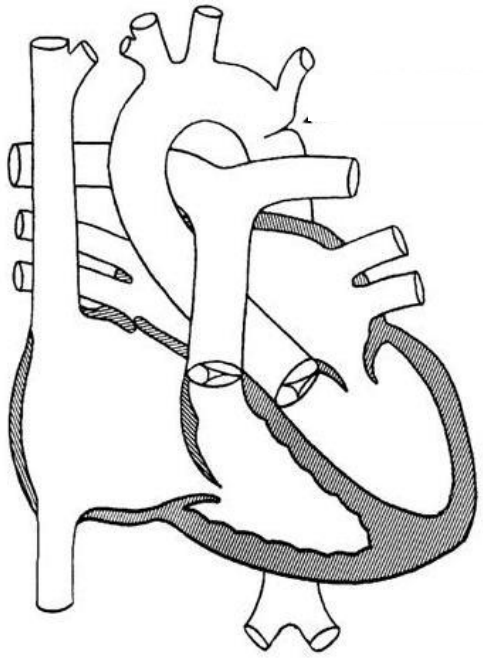
(Hint pressures in the left side of the heart are higher than the right side)



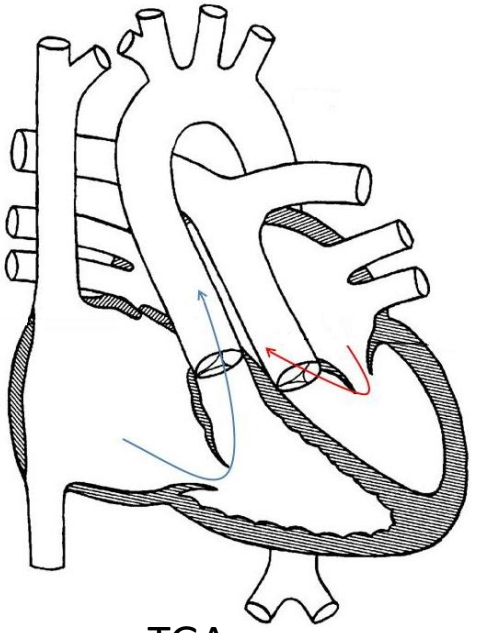
Tetralogy of Fallot



Atrial Septal Defect



Coarctation



TGA

Tetralogy of Fallot

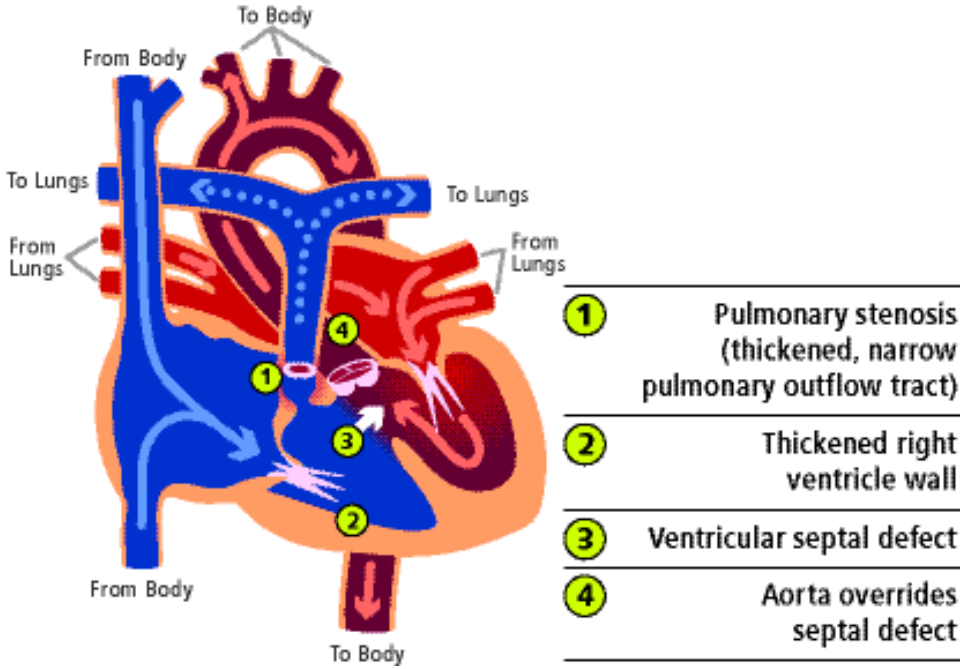
There are four things wrong with the heart (tetra = Greek for four)

1 A narrowing in the way out from the right side of the heart to the lungs (pulmonary stenosis)

2. The right ventricular muscle gets thickened as it has to work harder trying to push blood through the narrowed pulmonary valve. The greater the narrowing the more blue blood goes around the body. This leads to the baby looking dusky.

3. A hole in the partition between the two pumping chambers of the heart (ventricular septal defect)

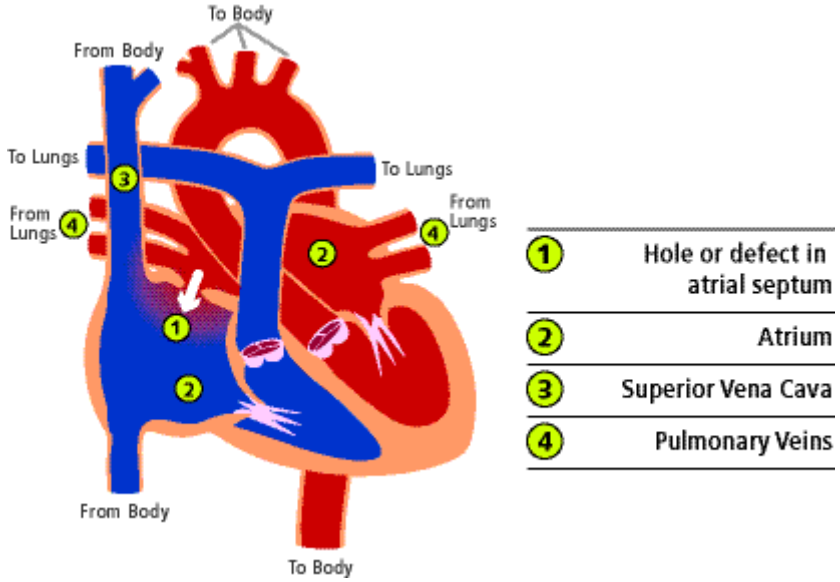
4 The aorta is over-riding the hole and receives blood from both ventricles therefore allowing some deoxygenated (blue) blood to go round the body.



Atrial septal defect

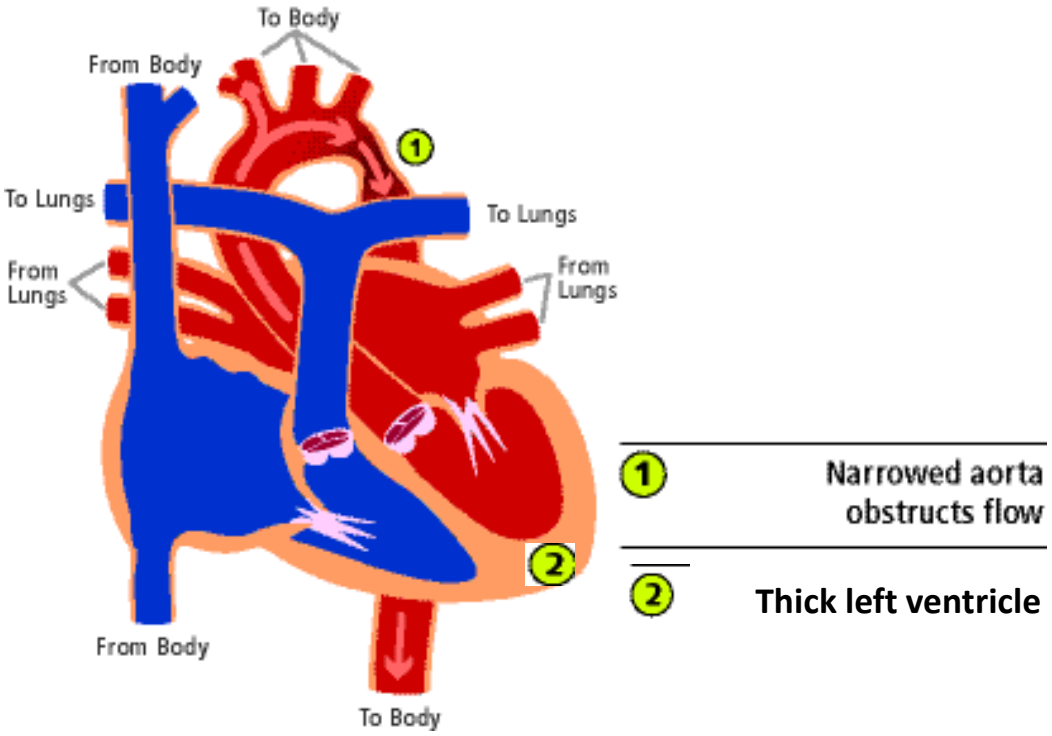
This is a hole (or defect) in the partition between the two filling parts of the heart (the atria).

Some of the red blood from the left side of the heart flows through the hole to the right side
and there is an increased amount of blood flowing to the lungs.



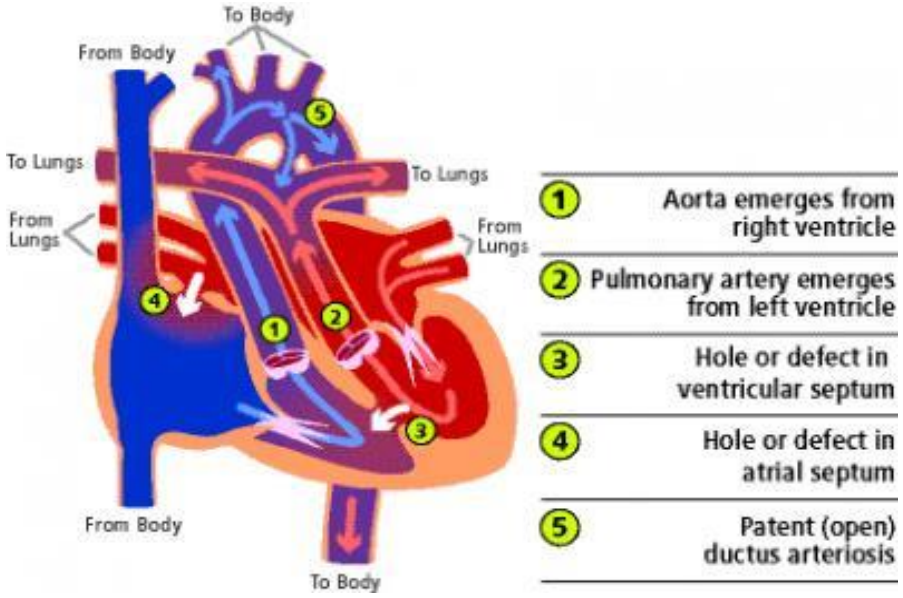
Coarctation of the aorta

This is a tight narrowing of the main artery (the aorta)
The left hand side of the heart has to work harder to pump blood away from the heart to the rest of the body



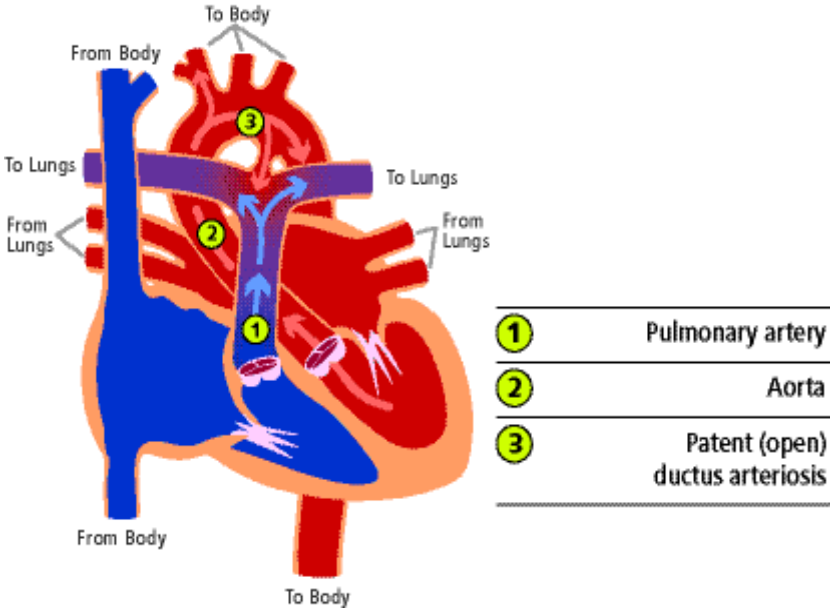
Transposition of the great arteries

The main arteries come from the wrong side of the heart. The aorta which goes round the body comes from the right sided pumping part (instead of the left) so blue blood is pumped round the body. The pulmonary artery which goes to the lungs comes from the left sided pumping part (instead of the right) and red blood goes again to the lungs. The baby can only survive if some of the red blood crosses over to join the blue blood to go round the body. The red blood can cross through a hole between the filling parts of the heart or between the pumping parts of the heart or through the ductus arteriosus.



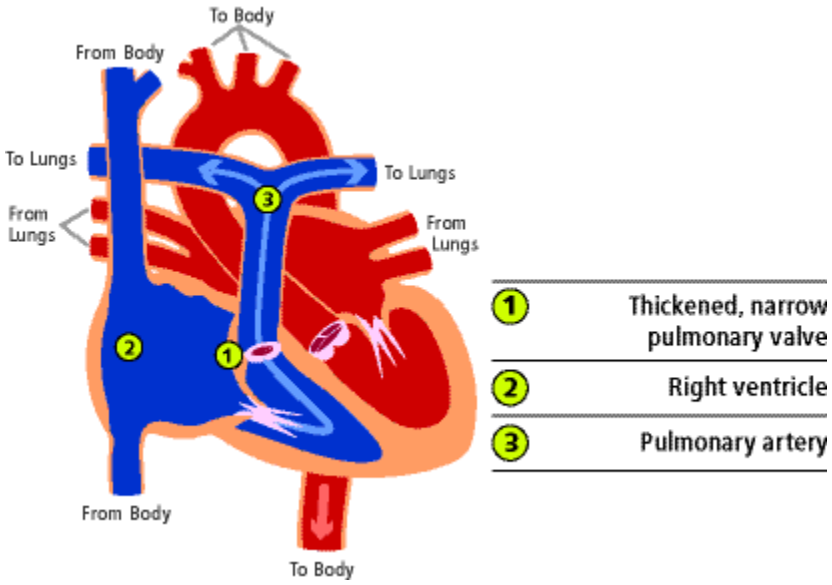
Persistent ductus arteriosus (PDA)

In the normal heart the ductus arteriosus (which is open in the infant before it is born) closes after birth. In some babies, however it remains open and allows blood to flow from the aorta (the main artery going around the body) to the lungs. The lungs receive an increased amount of blood and the heart has to do extra work.



Pulmonary stenosis

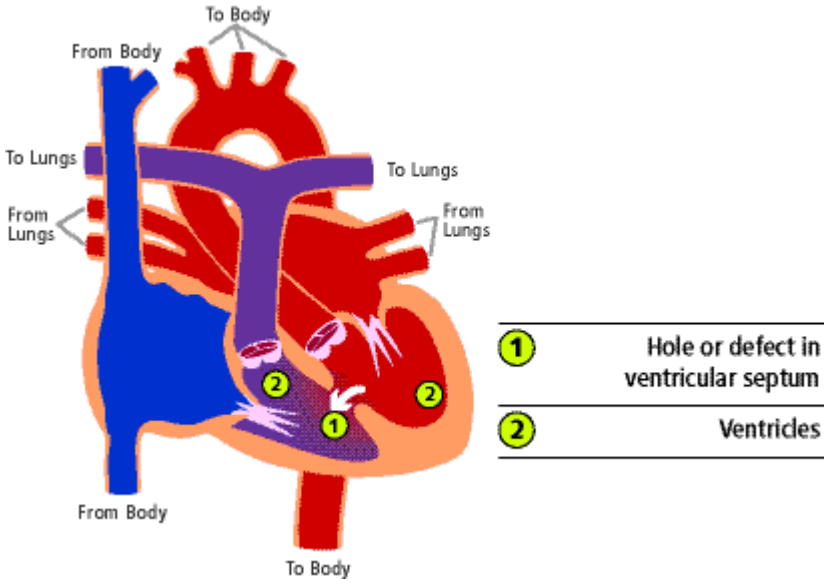
This is a narrowing of the valve in the artery going to the lungs. The pumping part of the heart on the right side (the right ventricle) has to work harder to force the blood through the narrowed area. The degree of narrowing may be mild, moderate or severe



Ventricular septal defect

This is a hole in the muscular partition between the two pumping chambers (ventricles) of the heart. Blood passes from the left sided chamber to the right sided chamber and as a result, the blood flow to the lungs is increased and the heart has to do more work than usual.

The blood pressure in the lung artery is usually much higher than normal.



Relative age

	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

Age difference:

- >40
- 30-40
- 20-30
- 10-20
- 5-10
- 2-5
- <2

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.

Cyanotic or Acyanotic lesions

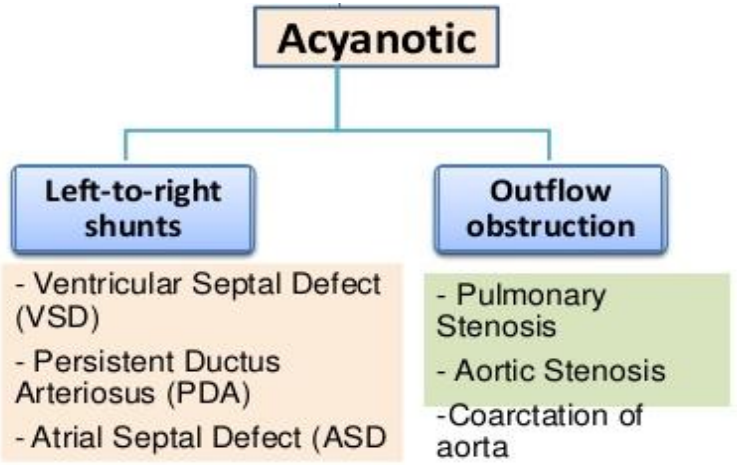
5 "T's"

Most common cyanotic lesions of the newborn

- Five • Total Anomalous Pulmonary Veins
- Four • Tetrology of Fallot
- Three • Tricuspid Atresia
- Two • Transposition
- One • Truncus Arteriosus

Aim for saturations 75 – 85%

Aim for normal saturations



Prostin dependant lesions

Causes vasodilation of the smooth muscle in the ductus, maintain patency

Left-Sided Obstruction (Systemic Circulation Impaired)	Right-Sided Obstruction (Pulmonary Circulation Impaired)
Hypoplastic left heart syndrome	Tetralogy of fallot with pulmonary atresia
Critical aortic stenosis	Pulmonary atresia
Coarctation of the aorta	Critical pulmonary stenosis
Interrupted aortic arch	Tricuspid atresia
	Severe Ebstein's anomaly
	Complete transposition of the great arteries with intact interventricular septum

- For lesions dependant on pulmonary blood flow, it improves oxygenation
- For lesions dependant on systemic blood flow improves blood pressure and perfusion

Prostin can be initiated before a definitive diagnosis is established

Nursing considerations

- Start with ABC

- Cardiac output
- ✓ Colour
- ✓ Temperature
- ✓ Perfusion/cap refill
- ✓ Blood pressure – use right arm
- ✓ Heart rate/ rhythm/ECG
- Nutrition
- Support/provide information to parents

Oxygen saturations – Pre and post ductal

- Right hand, either foot
- Not the left hand as this is influenced by the duct, studies have shown its closer to pre ductal saturations
- Target saturations 77-85% this is adequate to avoid tissue hypoxia
- If severe hypoxia **Do not hesitate administer oxygen**

2 OUT OF 3 heart defects aren't picked up during pregnancy scans

1000 newborns leave UK hospitals each year with undetected major defects

Age- around 6 days of age

Low saturations despite oxygen

Reduced feeding

Not waking for feeds

Absent femoral pulse

Could this be cardiac

Overview of the CHD Network

South Wales and South West
Congenital Heart
Disease Network

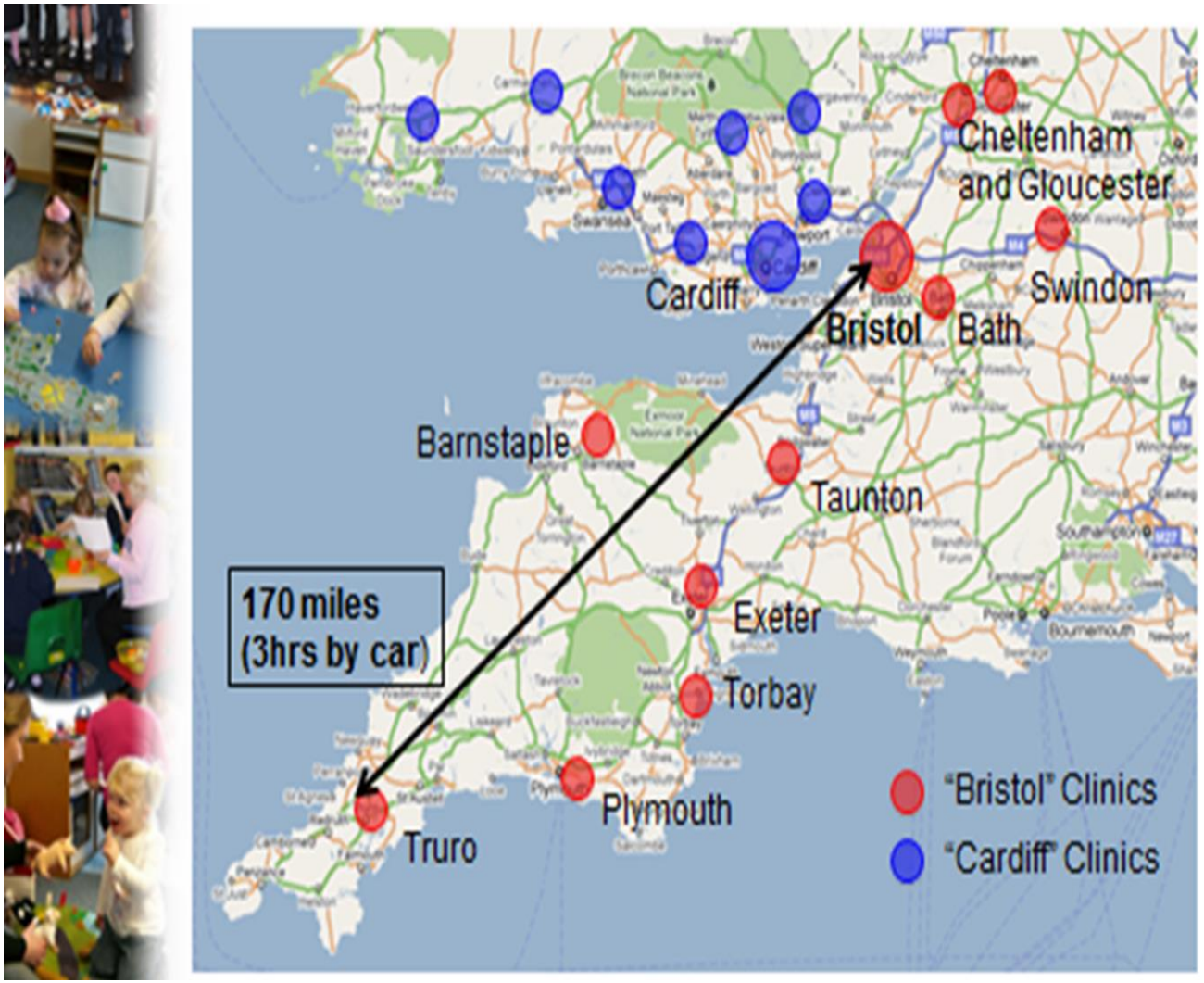
The South West and South Wales Congenital Heart Disease Network brings together all of the hospitals, clinicians, commissioners and patients involved with congenital heart disease services in the region in order to deliver the highest quality care and experience for patients.

CHD care is provided in a network ‘hub and spoke 'model

To ensure the right level of care is offered to the patients at the right time

- Level 1 specialist surgery and procedures (UH Bristol)
- Level 2 some procedures (UH wales in Cardiff)
- Level 3 outpatients (local hospitals)

The Network Region

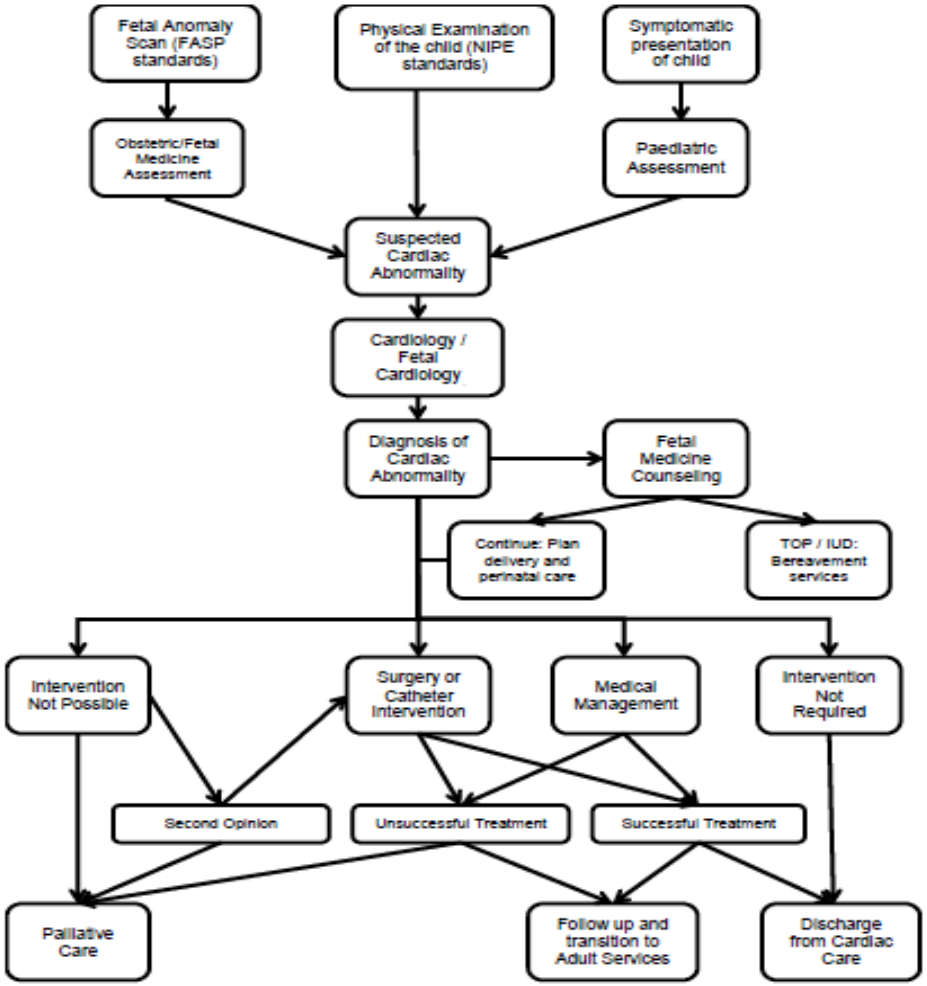


South Wales and South West Congenital Heart Disease Network

The 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, governance and audit
- Section G: Research
- Section H: Communication with patients
- Section I: Transition
- Section J: Pregnancy and contraception
- Section K: Foetal diagnosis
- Section L: Palliative care and bereavement

3.2.2 Patient care pathway



3.2.3 Referral

Patients enter the paediatric cardiac pathway through:

Standard	Paediatric	Implementation timescale
The following specialties or facilities **should ideally** be located on the same hospital site as Specialist Children's Surgical Centres. Consultants from the following services must be able to provide urgent telephone advice (call to advice within 30 **minutes**) and a visit or transfer of care within **four hours** if needed. The services must be experienced in caring for patients with congenital heart disease.		
D16(L1)	Paediatric Neurology.	Immediate
D17(L1)	Paediatric Respiratory Medicine.	Immediate
D18(L1)	Neonatology.	Immediate
D19(L1)	Clinical Haematology.	Immediate
D20(L1)	Infection control team experienced in the needs of paediatric cardiac surgery patients.	Immediate
D21(L1)	Paediatric Neurosurgery.	Immediate
D22(L1)	Child Psychiatry (with dedicated sessions and 24/7 on call).	Immediate
D23(L1)	Clinical biochemistry (including toxicology).	Immediate
D24(L1)	Pharmacy (with dedicated sessions for CHD and 24/7 on-call for urgent supply and advice).	Immediate
D25(L1)	Paediatric Endocrinology.	Immediate
D26(L1)	Paediatric Orthopaedics.	Immediate
D27(L1)	Plastic surgery.	Immediate

Network Activity

- 18 adult and 19 paediatric providers, covering level 1 (specialist surgical), 2 (specialist cardiology) and 3 (local centre) services
- Over 6,500 children and 8,000 adults with congenital heart problems
- 30 clinics per week, seeing more than 20,000 outpatient attendances pa
- 37 paediatricians with expertise in cardiology
- 17 adult cardiologists with specialist congenital interest
- Over 40 specialist and link nurses
- Over 425 heart operations

South Wales and South West Congenital Heart Disease Network

New Congenital Heart Disease Network website

Resources for patients, families and professionals

For patients and families:

- Patient information
- Getting support
- Understanding your CHD journey

For professionals:

- Guidelines
- Protocols
- Training and education

www.swswchd.co.uk

South Wales and South West Congenital Heart Disease Network

Paediatric Cardiac Clinical Nurse Specialists

Bristol Royal Hospital for Children

Are available to assist you with issues surrounding diagnosis, medications, lifestyle and school/nursery, as well as to provide emotional support for your family

Monday to Friday, 8am - 4pm

Cardiac nurse specialist call 0117 342 8286 / 0117 342 8578 or email cardiacnurses@uhbristol.nhs.uk

Hospital switchboard: 0117 923 0000 Bleep 3983

Support for Babies and Children

Click here to find out about the range of different support available if you care for a baby or child with a Congenital Heart condition.

Find out more ›

Support for Young People

Click here to find out about the range of different support available if you are a teenager or young person with a Congenital Heart condition.

Find out more ›

Support for Parents/Carers

Click here to find out about the range of different support available if you are a parent of a child with a Congenital Heart condition.

Find out more ›

Support for Adults

Click here to find out about the range of different support available if you are an adult with a Congenital Heart condition.

Find out more ›

South Wales and South West Congenital Heart Disease Network

Welcome to our training and education page. Here you will find information about forthcoming training events run by the network or hospitals within the network, as well as events run by other organisations that may be of interest. Our draft training and education strategy is available here.

2019 Network events

Consultant away day, Cardiff, 4th March 2019

South West Adult Congenital Heart Disease Training Programme, 18th March 2019, Bristol. For more information, click here.

The Welsh Paediatric Cardiovascular Network Spring meeting, 12th April 2019, Cardiff. For more information, click here.

Paediatric Cardiac Study Day for Community and Ward Nurses, 30th May 2019, Bristol. For more information, click here.

PEC study day, Bristol. 2nd July 2019. For for information, click here.

ACHD Study day, Bristol, 1st October 2019. For more information, click here.

2019 Other events

London Arrhythmia Summit 2019, 15th March. For more information, click here.

Paediatric Arrhythmia Study Day for Nurses, 25th March 2019, Bristol. For more information, click here.

Children's Cardiac Enhanced Care & Management course for nurses, 3rd, 10th, 16th, 23rd, 30th april, 8th, 17th, 28th May, Bristol. For more information, click here.

EuroGUCH, 5th-6th April 2019, Zagreb, Croatia. For more information, click here.

ACHD Study Days by Barts and GOSH, April - July 2019. For more information, click here.

Pulmonary Hypertension Education Day, 3rd May 2019, London. For more information, click here.

AEPC 2019, 15th-18th May 2019, Seville, Spain. For more information, click here.

Dementia and End of Life Care training, 4th July 2019, Bristol. For more information, click here.

Loss and Bereavement training, 10th July, Bristol. For more information, click here.

Paediatric Palliative Care Study Day, 16th July 2019, Bristol. For more information. click here.

BCCA Annual Meeting 2019, 19-20 November 2019, Gateshead. For more information. click here.

South Wales and South West Congenital Heart Disease Network

February 2019
Issue 5

Newsletter

Inside this issue

News from the Network Team

Welcome to our first newsletter of 2019 containing project updates, inspiring patient stories and an opportunity to introduce you to some new members of our team. We are delighted to welcome Cat McElvaney (Interim Network Manager) and Morwenna Bugg (Network Support Manager) to the Network Team.

We are sad to say goodbye to Caitlin Moss, who has worked as the Network Manager since the Networks initiation in April 2016. Caitlin has been appointed substantively to the role of Deputy Divisional Director for Women's and Children's Services in University Hospitals Bristol, following a short-notice secondment in October 2018. Caitlin has loved working with all of the Network's stakeholders across the region, and being part of the team from day one of the network being set-up. She is incredibly proud of what has been collectively achieved, and has mixed emotions about moving on. In her new role, Caitlin will remain responsible for children's networks from a University Hospitals Bristol perspective, so will continue to be involved in and supportive of the Network moving forward. .We would like to thank Caitlin for all her hard work, determination and commitment, which undoubtedly has been a major factor in the success of this Network. A process for the substantive recruitment to the Network Manager role will launch in the coming weeks, so watch this space!

2019 promises to be a busy year for us with an ambitious work programme. However, progress would not be possible without the fantastic collaboration of colleagues. This is exemplified by the truly excellent work from our Dental Team led by Mick Allen, Consultant in Special Care Dentistry and Rosemary Power, Consultant in Paediatric Dentistry. See page 2 for more details.

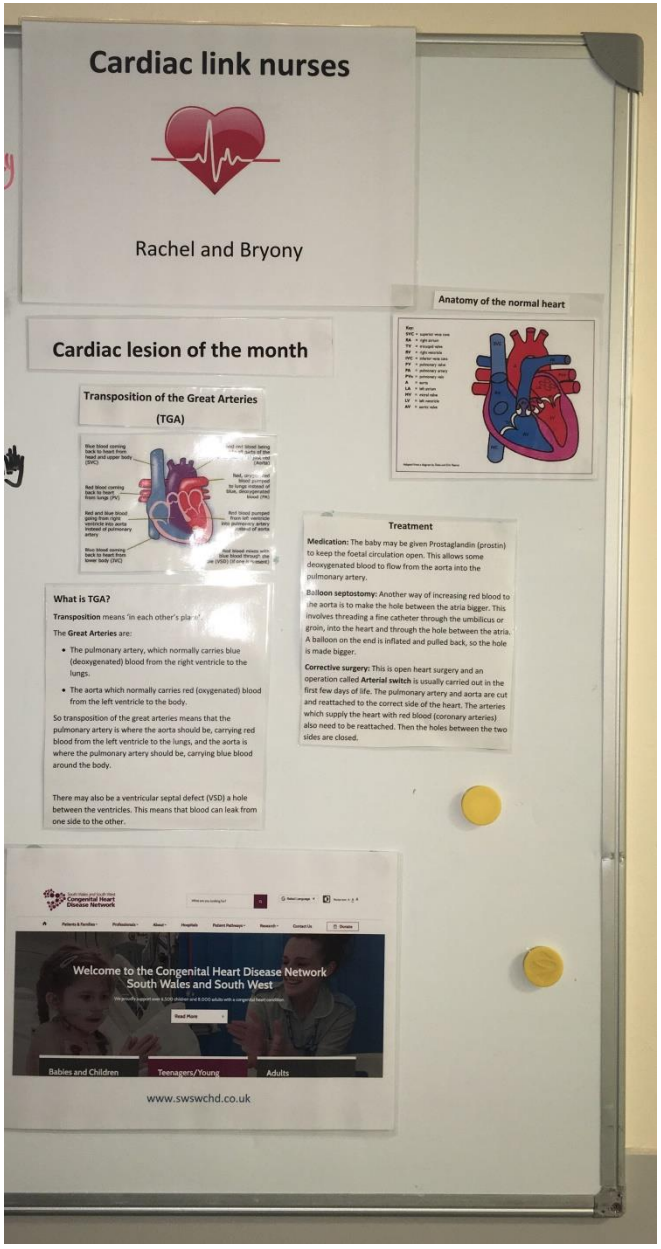
Finally, later this year NHS England's Quality Surveillance Team will conduct a programme of Peer Review visits to all Level 1 and Level 2 centres in the UK. Our Peer Review Visit is scheduled for the 10th-12th of June 2019. An assessment will be made against the published NHS England Standards for Congenital Heart Disease. It is clear that an assessment of CHD Networks, across the UK, will be an important part of these visits. We can be rightly proud of the collaboration and work and we have completed to date.

Meet Morwenna, Network Support Manager

Morwenna recently joined the Network Team and will be a great support in helping the network to deliver its ambitious work programme. Previously Morwenna worked at NHS Bristol, North Somerset and South Gloucestershire Clinical Commissioning Group after studying nutrition at university.

Key contacts

Clinical Director: Dr Andrew Tometzki
Andrew.Tometzki@uhbristol.nhs.uk
Lead Nurse: Sheena Vernon
Sheena.Vernon@UHBristol.nhs.uk
Interim Network Manager: Cat McElvaney
Cat.McElvaney@uhbristol.nhs.uk
Lead Psychologist: Ness Garratt
Vanessa.Garratt@uhbristol.nhs.uk
Support Manager: Morwenna Bugg
Morwenna.Bugg@uhbristol.nhs.uk



Cardiac information board

Found outside the seminar room

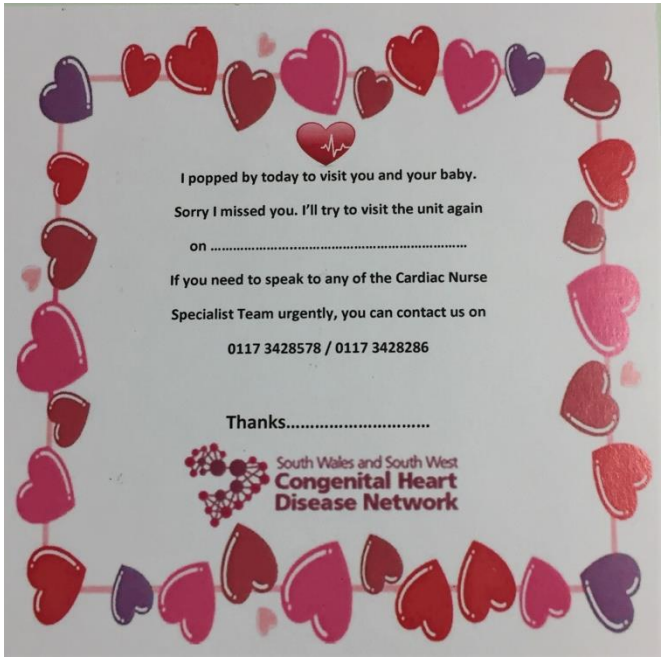
Every month will be updated with a new lesion

Any relevant study/ education days will be found on this board

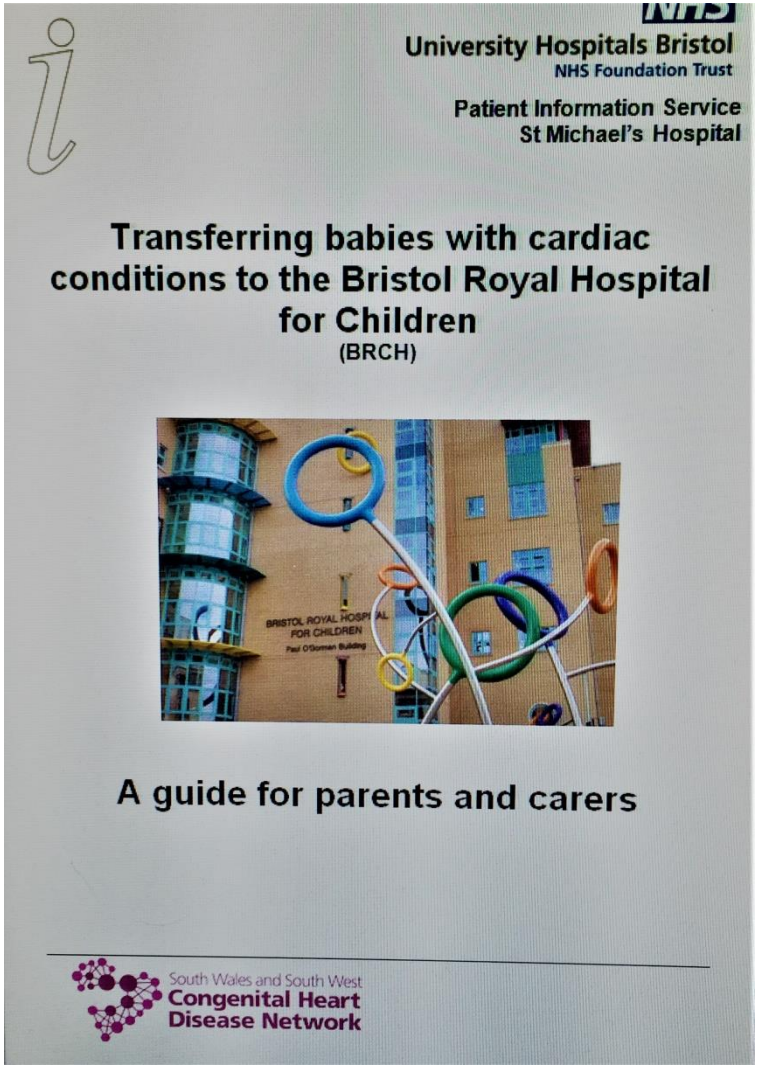
Network information will also be found here

Coming soon

Cardiac Nurse Specialist visit the unit on Mondays, if parents are not here please fill this card in and leave on babies bed space



Find them in the cot card draw behind main reception



Any Questions?

Ask Bryony