Bristol Royal Hospital For Children
Faculty of Children's Nurse Education

South Wales and South West Congenital Heart Disease Network

NHS University Hospitals Bristol and Weston NHS Foundation Trust

Paediatric Cardiac Workbook

Answers

Created by Carla Sims (2023)

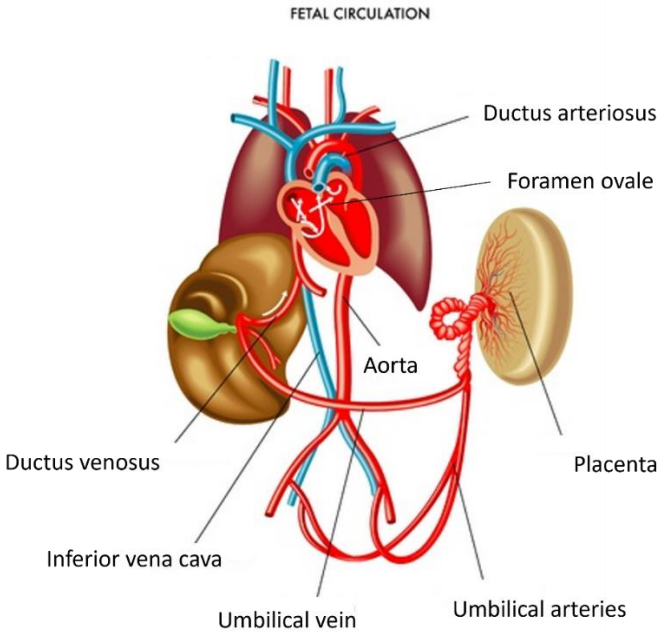
Senior Staff Nurse and Clinical Skills Facilitator for Dolphin Ward,

Bristol Royal Hospital for Children

With special thanks to

Alison Wood, Cardiac Nurse Educator, BRHC
Sheena Vernon, Lead Nurse, South Wales and South West Congenital Heart Disease Network

Foetal Circulation



Describe the changes which take place in the transition between foetal circulation and when the baby is born:

1. The umbilical cord is clamped and cut so the baby is no longer supplied with oxygen and nutrients via the placenta.
2. The pressure in the systemic circulation rises.
3. Reduction in maternal prostaglandins.
4. Oxygen saturations increase as the baby takes its first breath.
5. The ductus arteriosus closes.
6. The pressure in the pulmonary system decreases, encouraging more blood flow to the lungs and reduced pressures in the right side of the heart.
7. The increased pulmonary blood flow means there is more blood returning to the left atrium.
8. The changes in pressures between right and left side along with increased blood flow to the LA means that the foramen ovale flap is pushed closed. Blood no longer flows from RA to LA.
9. The ductus venosus closes so that blood passes through the liver before returning to the heart.

Maintaining the patency of the ductus arteriosus with Prostaglandins

What cardiac conditions can you think of that may require prostaglandin to maintain ductal patency after birth?

There are many answers that may apply here.

These can be divided into cardiac lesions that are duct-dependent for pulmonary circulation (pulmonary atresia with intact septum), systemic circulation (coarctation, hypoplastic left heart) or for mixing of the circulations (transposition of the great arteries)

What are the side-effects related to administration of dinoprostone (prostaglandin E2)?

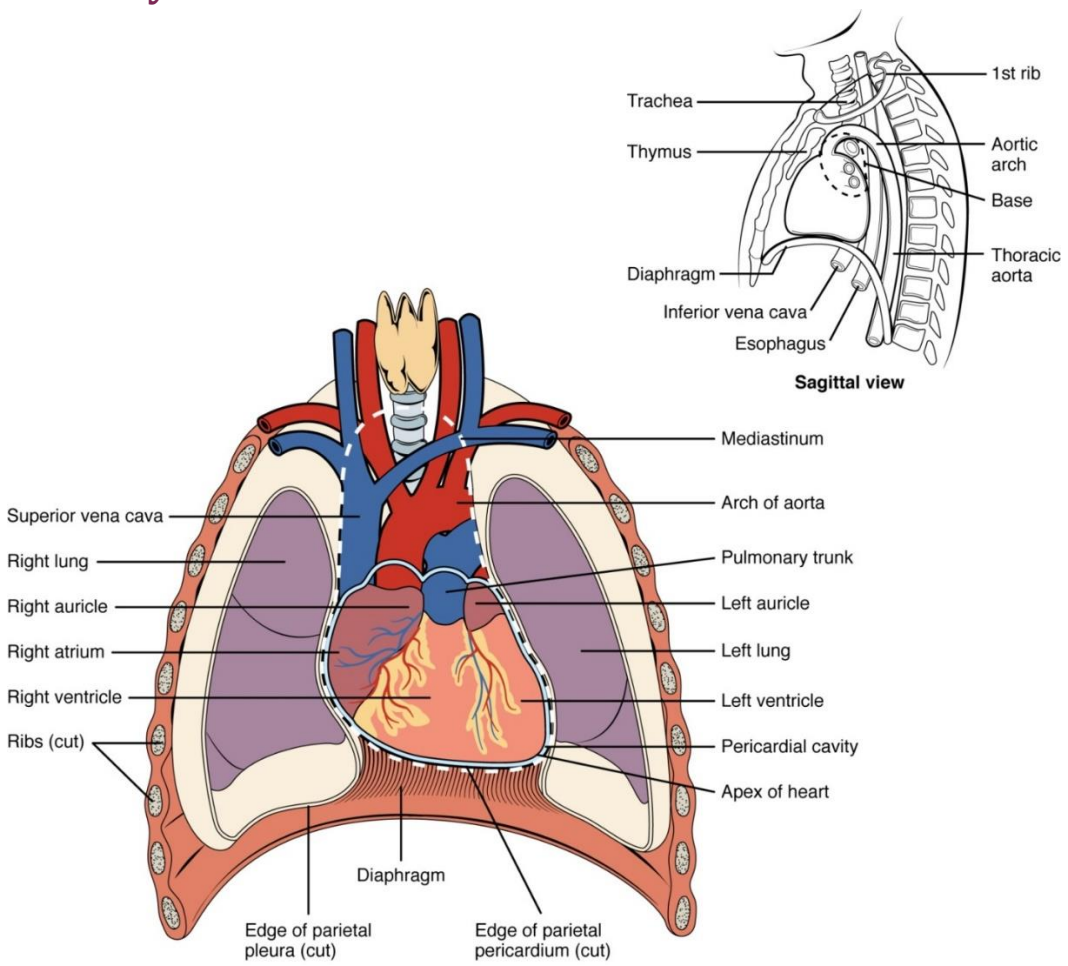
The most commonly seen side effect is apnoeas and this is more likely to occur upon commencing dinoprostone or with higher doses >20ng/kg/min

Fever

Hypotension

Tachy- or brady-cardia

Cardiac Anatomy



OpenStax College (https://commons.wikimedia.org/wiki/File:2001_Heart_Position_in_ThoraxN.jpg)„2001 Heart Position in ThoraxN“, https://creativecommons.org/licenses/by/3.0/legalcode

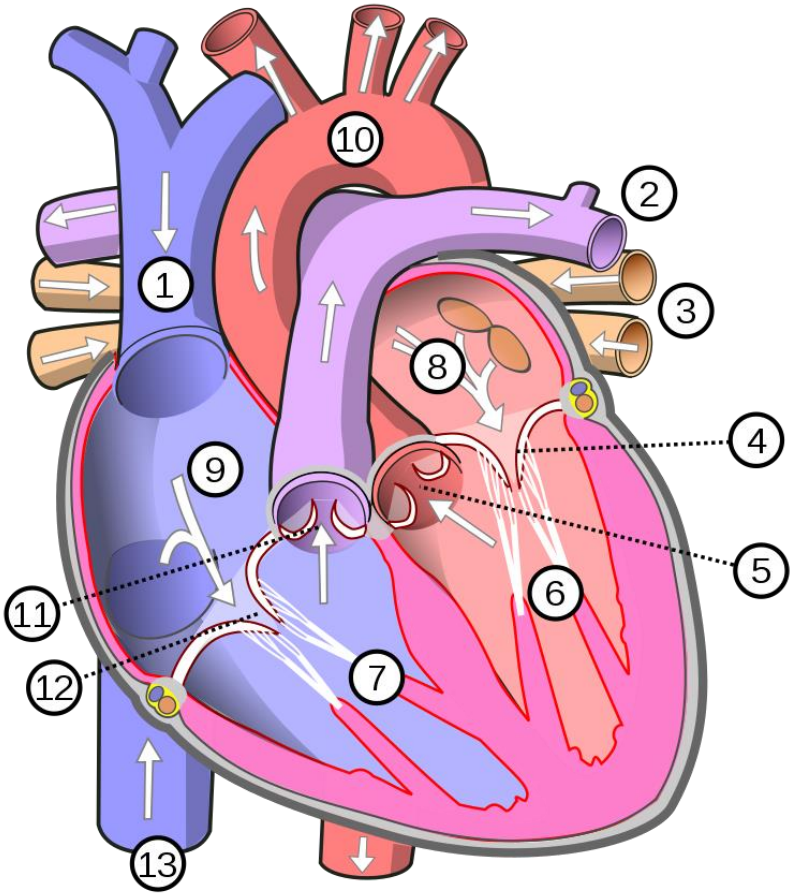
Consider the size of a newborn's heart. What impact does this have on cardiac surgical and interventional cardiology procedures?

Considerations: Valve size, conduit size, weight of the patient.

1. New-born's heart is the size of their fist – could be smaller than a walnut! Smaller size = more tricky to perform delicate procedures on blood vessels.
2. Tools may not be produced small enough. The devices to be inserted may not come in appropriate sizes – valves, conduits, occlusion devices etc.
3. If a device is successfully inserted into the patient then it means that they will have to have further surgery as they grow due to the device not growing with them and rapid growth phase particularly in first few years of life.

Structure of the Heart

Label this diagram of the heart and draw the direction of the blood flow. Identify whether the blood is oxygenated or deoxygenated.



1) Superior Vena Cava
2) Pulmonary Artery
3) Pulmonary veins
4) Mitral Valve
5) Aortic Valve
6) Left Ventricle
7) Right Ventricle
8) Left Atrium
9) Right Atrium
10) Aorta
11) Pulmonary Valve
12) Tricuspid Valve
13) Inferior Vena Cava

The Heart Valves

Valves prevent the back flow of blood within the heart. They separate the upper and lower chambers of the heart (atria and ventricles) and the ventricles from the great vessels.

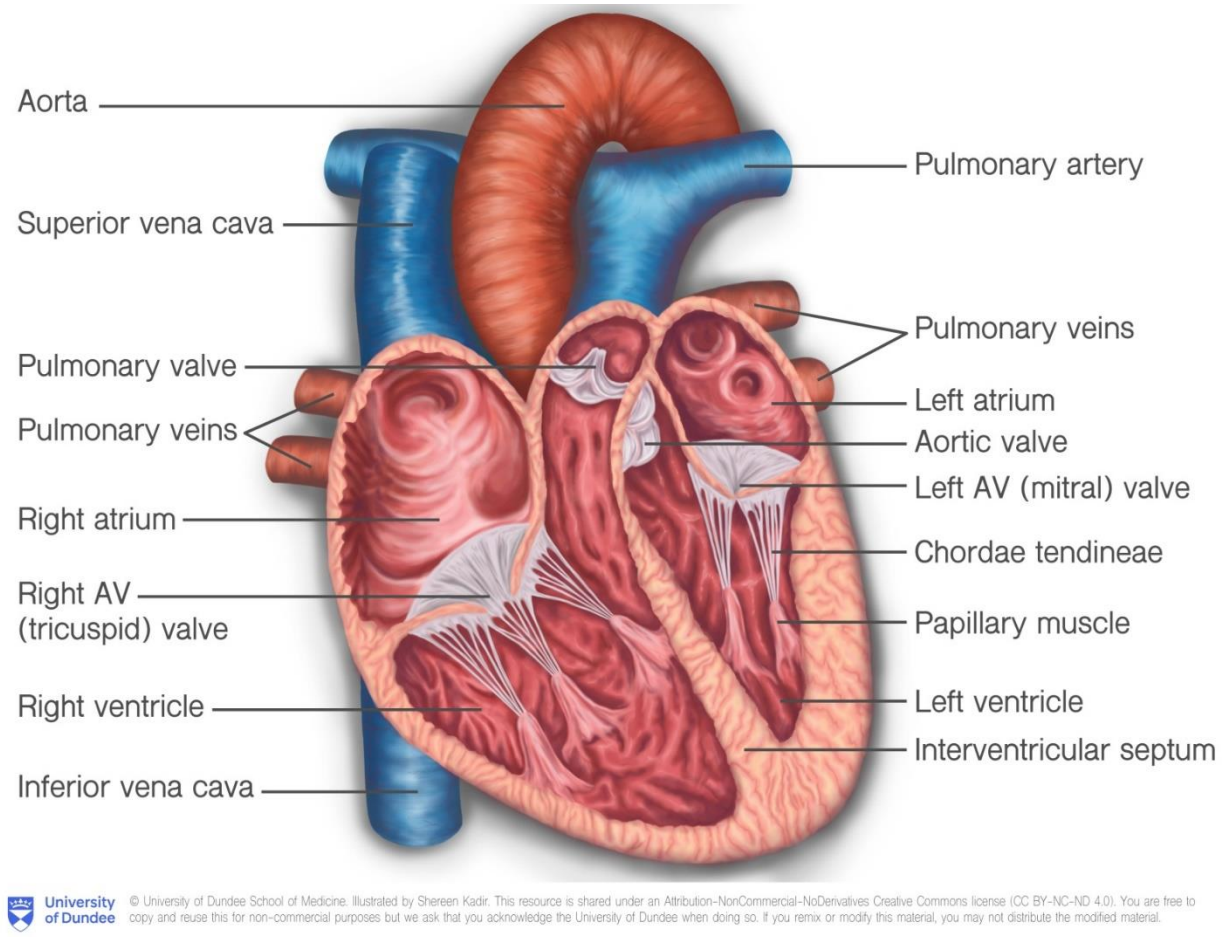
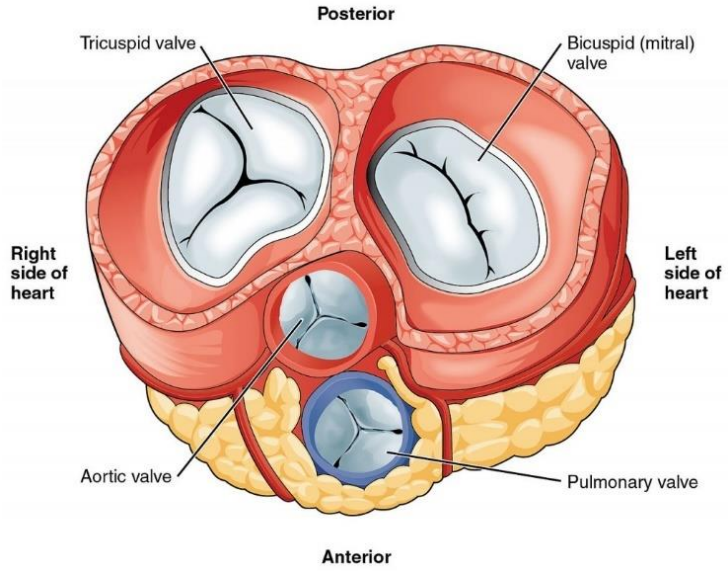


Image below shows the four valves of the heart visible with the atria and great vessels removed.



OpenStax College (https://commons.wikimedia.org/wiki/File:2011_Heart_Valves.jpg), „2011 Heart Valves", https://creativecommons.org/licenses/by/3.0/legalcode

Name the two atrioventricular valves.

Tricuspid (right side) and mitral or less-commonly bicuspid (left side)

How many leaflets does each have?

Tricuspid = 3 Mitral = 2

Name the two semilunar valves.

Pulmonary and aortic

How many leaflets does each have?

3

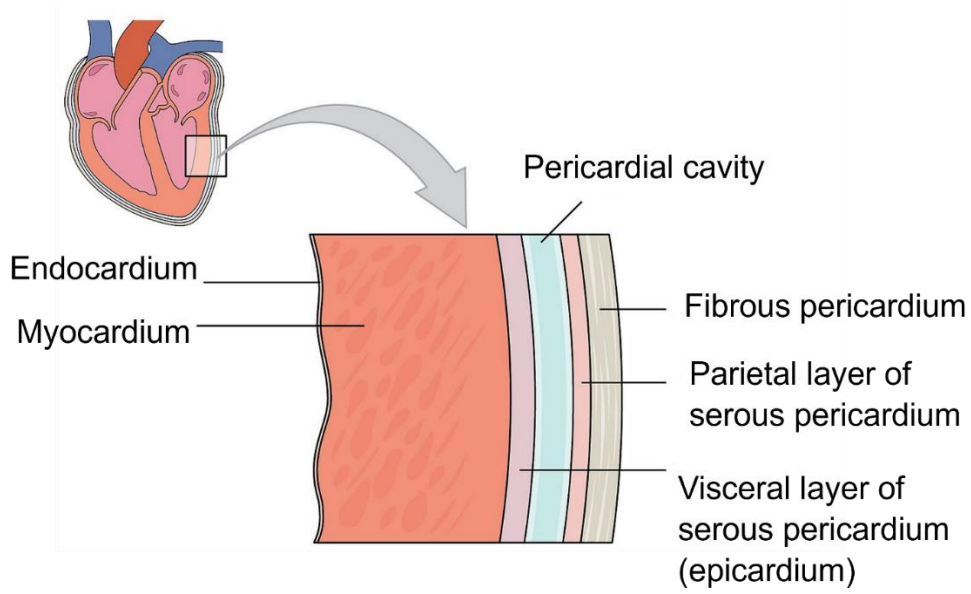
If the valves are 'incompetent', what does this mean and what is the consequence for blood flow?

The valve does not close fully after each beat

Allows back flow of blood

Leads to decreased cardiac output

Layers of the Heart



OpenStax College (https://commons.wikimedia.org/wiki/File:2004_Heart_Wall.jpg), „2004 Heart Wall", remove labels by Alison Wood, https://creativecommons.org/licenses/by/3.0/legalcode

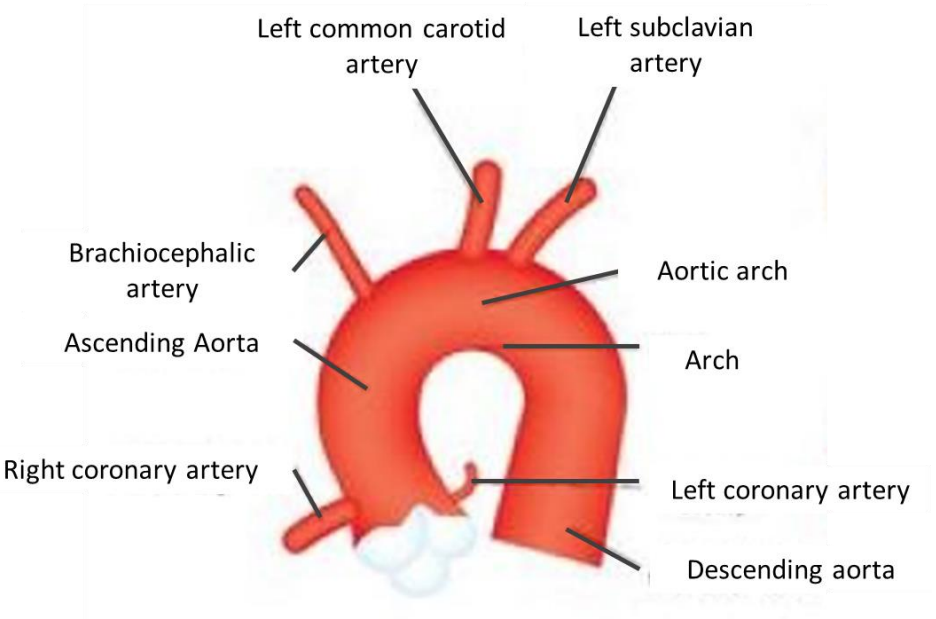
Label the layers of the heart using the following labels:
Pericardial cavity
Fibrous pericardium
Parietal layer of serous pericardium
Visceral layer of serous pericardium (epicardium)
Endocardium
Myocardium

What is important about the structure of each of the layers in relation to their function?

The outer layer of the heart (fibrous pericardium) is tough and inelastic. It protects the heart and restricts the amount of stretch that the muscle can be placed under. It also anchors the heart in its position in the chest cavity.

The parietal and visceral layers surround the pericardial cavity which contains a small amount of fluid which enables smooth frictionless movement of the heart muscle within its sac. An increase in this fluid will restrict the space available in the chambers inside the heart and may impact on cardiac output.

The myocardium is the muscular layer of the heart.
The endocardium is smooth and continuous with the valves and blood vessels and minimises risks of blood clots forming allowing blood to flow smoothly.



Major arteries Superior to the heart

Label the three vessels which arise from the aortic arch.

Where do the coronary arteries arise from?

The right and left coronary arteries arise from the ascending aorta and are connected behind the leaflets of the aortic valve.

What is the main difference when comparing coronary perfusion to systemic perfusion?

Coronary perfusion occurs during diastole (relaxation of the heart) whereas systemic perfusion occurs during systole

To where do the coronary veins drain?

The drain to the coronary sinus which empties into the right atrium

Conduction System of the Heart

Describe the pathway of the electrical impulse as it passes through the heart.

The impulse is initiated in the sino-atrial (SA) node. This is an area of specialised cells, situated at the right-side of the upper wall in the right atrium. These cells spontaneously generate electrical impulses which then are transmitted across both atria where they converge at the atrioventricular (AV) node.

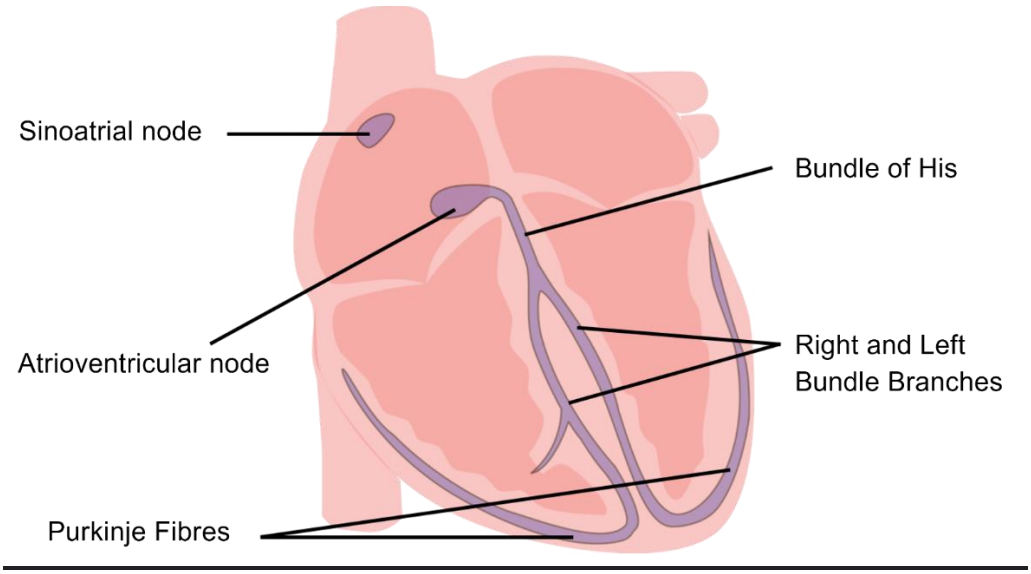
The AV node is located centrally on the front of the heart between the upper and lower chambers. There is a brief pause at this point before the impulse moves down the Bundle of His which sits over the septum. This divides into the left and right bundle branches, still moving down towards the apex of the heart, before spreading upwards and outwards across both ventricles through the Purkinje fibres.

After contraction, the heart muscle relaxes and prepares for the next beat. It is during relaxation (diastole) that the coronary arteries are filled and perfuse the heart muscle. This rapid electrical conduction enables a coordinated muscle contraction and smooth, effective passage of blood through and out of the heart.

On this diagram identify:

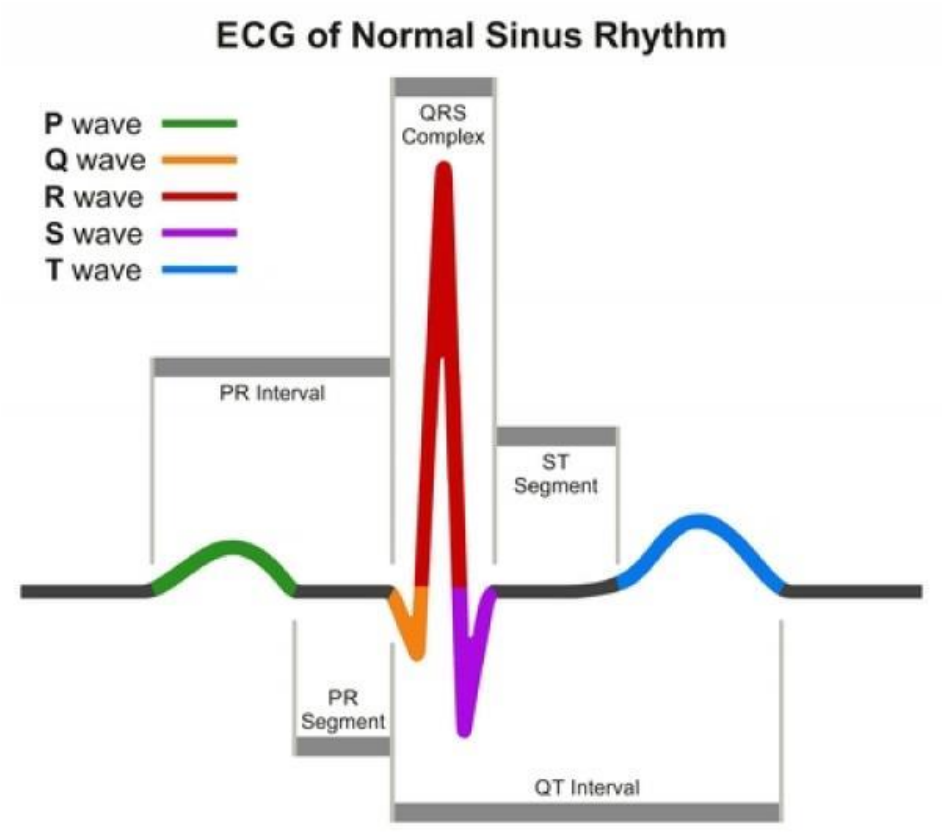
Sinoatrial node Atrioventricular node Bundle of His Purkinje Fibres

Right and Left Bundle Branches

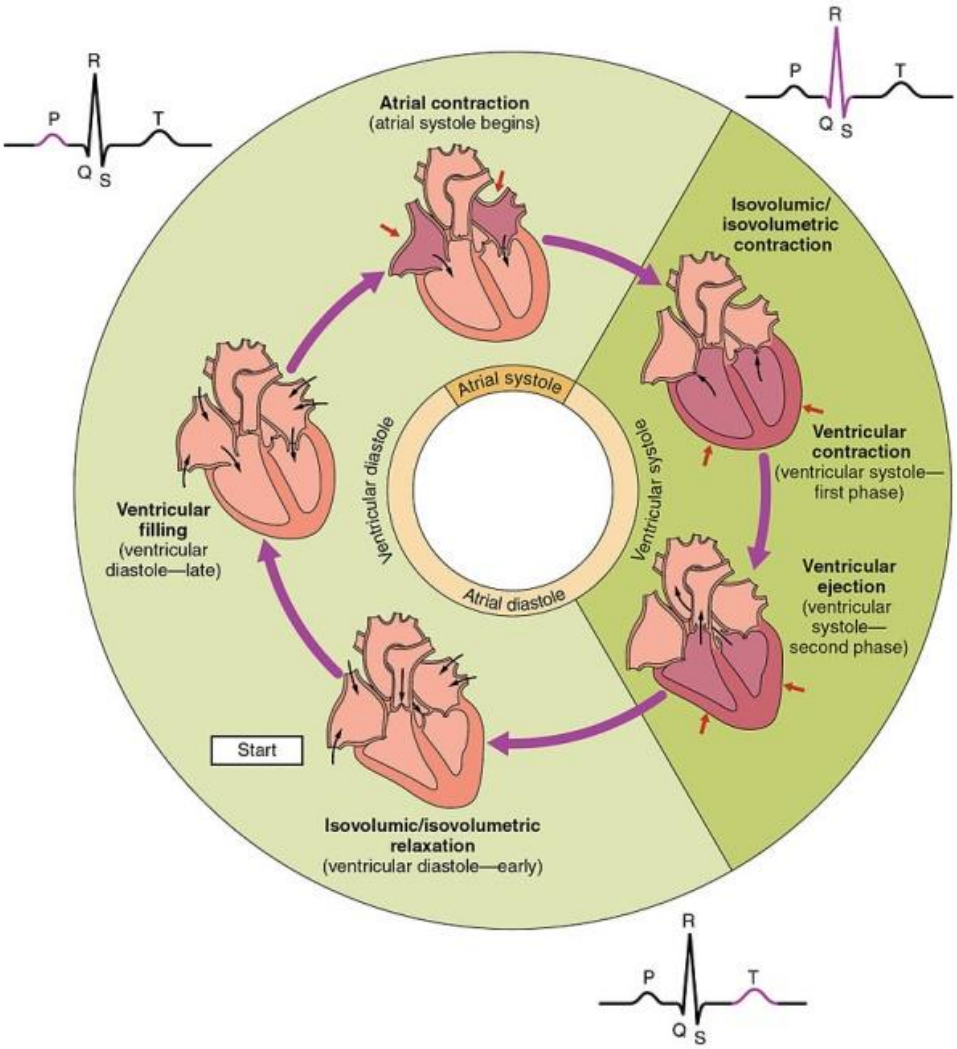


U Bhalraam (https://commons.wikimedia.org/wiki/File:Heart_vector_electrical_rest.png), added labels blank by Alison Wood, https://creativecommons.org/licenses/by-sa/4.0/legalcode

Electrocardiogram (ECG)



The Cardiac Cycle



OpenStax College (https://commons.wikimedia.org/wiki/File:2027_Phases_of_the_Cardiac_Cycle.jpg), „2027 Phases of the Cardiac Cycle", https://creativecommons.org/licenses/by/3.0/legalcode

Describe the activity within the heart at each stage in the cardiac cycle represented in the diagram above

The cardiac cycle is the activity of the human heart from the beginning of one heartbeat to the beginning of the next. It is typically spilt into 5 stages which encompass both systole (contraction of the heart) and diastole (relaxation of the heart muscle).

Stage 1 – Atrial contraction. The electrical impulse is initiated in the sino-atrial node and spreads across both atria. These cells 'depolarise' and cause the muscle to contract. The atrioventricular (AV) valves open. Blood is ejected from the atria into the ventricles through the atrioventricular nodes. Approximately 70% of ventricular filling occurs passively with only 30% occurring during atrial contraction. This can also be referred to as the "atrial kick".

Stage 2 – Isovolumetric ventricular contraction. The electrical impulse pauses briefly at the atrioventricular (AV) node to allow time for atria contraction to occur and blood to pass into the ventricles. The impulse then passes down the Bundle of His and right & left bundle branches before spreading up across the ventricles in the Purkinje fibres. This causes depolarisation of the ventricle muscle cells and elicits ventricular contraction. During isovolumetric contraction, the AV valves close (first heart sound), and the pressure is increasing within the ventricles, but the pressure is not yet high enough to open the semi-lunar pulmonary and aortic valves. The volume of blood in the ventricles remains constant during this brief phase.

Stage 3 – Ejection. The pulmonary and aortic valves open (AV valves remain closed) and the blood leaves the heart as it is pushed out of the ventricles into the arteries.

Stage 4 – Isovolumetric relaxation. The ventricular pressures drop below the pressures in the aorta and pulmonary artery and the semi-lunar valves close (second heart sound). This marks the beginning of diastole (ventricular relaxation) and all valves are closed at this point.

Stage 5 – Passive ventricular filling. Repolarisation occurs and the muscle of the ventricles relaxes. The AV valves open as the pressure in the ventricles drops below the pressure in the atria and this allows blood to flow passively from the atria to the ventricles. The heart is then ready to repeat the cycle.

Common Cardiac Conditions

VENTRICULAR SEPTAL DEFECT (VSD)

1.Describe the altered anatomy and blood flow of this defect	A hole in the septum between the two ventricles. Due to the higher pressures in the left side of the heart, blood flows from the left ventricle to the right ventricle and increases the blood flow to the lungs.
2. What symptoms might you expect to see in a patient if this is not repaired?	Breathlessness, pale, mottled, sweaty, tachycardia, poor feeding, sleepy.
3. What surgery/treatment is required?	Some smaller defects may close spontaneously. Open heart surgery using cardio-pulmonary bypass to repair the defect. Sometimes it is possible to close the defect via a cardiac catheter procedure.
4. What follow up/future treatment might they have?	Lifelong follow up recommended at specialist ACHD centres, 2-5 yearly intervals.

Atrial Septal Defect

1.Describe the altered anatomy and blood flow of this defect	A hole between the two upper chambers of the heart. Blood flows from the left atrium to the right atrium due to higher pressures on right. The gradient between the two side of the heart at this level is minimal so a smaller volume of blood is shunted than with a VSD but over time will still have effects of increased blood flow to the lungs
2. What symptoms might you expect to see in a patient if this is not repaired?	Unlikely to see symptoms as a baby but over time will see breathlessness, tachycardia, frequent respiratory infections etc
3. What surgery/treatment is required?	Cardiac catheter procedure to close the hole – a small tube is passed up through the femoral vein and/or artery into the heart and a closure device deployed to sit in the atrial septum. If unsuitable for catheter procedure the ASD may also be closed via open heart surgery with cardiopulmonary bypass
4. What follow up/future treatment might they have?	Patients repaired under 25 years of age who have no ongoing issues do not require ongoing follow-up, but should be warned of the risk of tachyarrhythmias

Coarctation of the Aorta

1.Describe the altered anatomy and blood flow of this defect	A narrowing of the aorta usually located at the point where the ductus arteriosus joins it. This prevents blood from flowing to the lower part of the body and abdominal organs.
2. What symptoms might you expect to see in a patient if this is not repaired?	Severity of obstruction can vary but patient may have circulatory compromise and shock. Pre-ductal sats may be normal and post-ductal much lower with poor perfusion on sats trace. Absent femoral pulses. 4 limb BP shows higher blood pressure to upper body (right arm in particular)
3. What surgery/treatment is required?	Surgery via intercostal incision without the need for cardiopulmonary bypass. Further procedure may be carried out via cardiac catheter to balloon the area of narrowing or insertion of a stent.
4. What follow up/future treatment might they have?	Regular follow up every two years at specialist ACHD centre to observe for high BP, recurrence of narrowing or aneurysm formation.

Hypoplastic Left Heart Syndrome (HLHS)

1.Describe the altered anatomy and blood flow of this defect	The left ventricle and aorta have failed to develop normally and are extremely small. Blood flow is dependent on a connection between the atria to facilitate blood flow around the body. Duct-dependent lesion – requires dinoprostone infusion to maintain patency of the ductus arteriosus
2. What symptoms might you expect to see in a patient if this is not repaired?	This condition requires immediate treatment and surgery in the first 2 weeks of life to ensure survival. All treatment is palliative as normal heart and function can never be restored.
3. What surgery/treatment is required (in each stage)?	Initial operation aims to create a new aorta and provide some blood flow to the lungs (Norwood stage 1). This is carried out in the first two weeks of life. At 4-6 months the patient will have a Glenn shunt (SVC is connected to the pulmonary artery). The third major operation will be the Fontans (IVC connected to the pulmonary artery). The patient will then have a univentricular circulation – the blood flows from the body directly to the lungs before flowing back to the heart. This circulation is dependent on healthy lungs and low-pressure pulmonary blood flow.
4. What follow up/future treatment might they have?	Life-long care as this is a palliative procedure. Patient will be on lifelong anti-coagulation management. Likely to have reduced exercise tolerance and at risk of heart failure, arrhythmias and liver disease.

Contacts based at Bristol Royal Hospital for Children:

Sheena Vernon, Lead Nurse Congenital Heart Disease Network: Sheena.Vernon@uhbw.nhs.uk

Jess Hughes, Lead Nurse Congenital Heart Disease Network: Jessica.Hughes@uhbw.nhs.uk

Lisa Patten, Cardiac Nurse Specialist: 0117 3428286

Zoe Trotman, Ward Manager Dolphin Ward: 0117 3428332

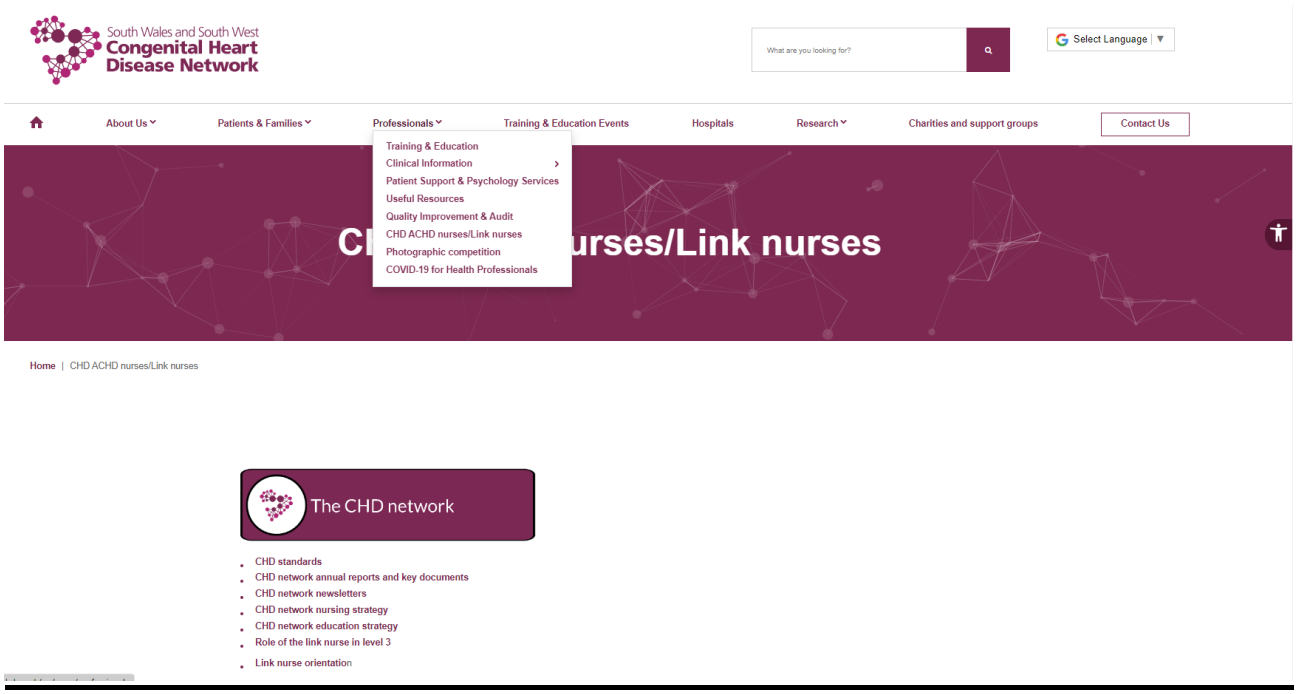
Alison Wood, Cardiac Educator: alison.wood@uhbw.nhs.uk

Carla Sims and Suzanne Conner, Clinical Support Facilitator: 0117 3428332

Faculty of Children's Nurse Education: FacultyOfChildrensNurseEducation@uhbw.nhs.uk

Network Website

- Type in the website: **www.swswchd.co.uk**



- Once on the website: hold curser over the professionals tab which will bring up 8 other tabs
- Click on CHD/ACHD Nurses/Link Nurses

Reference List

Assadi, R. (2013) Conduction System of the Heart. [Online] Available from: http://emedicine.medscape.com/article/1922987-overview [Accessed 27th July 2020]

Electrical Activity of the Heart. Available from: http://163.178.103.176/Tema1G/Grupos1/GermanT1/GATP13/E7.htm [Accessed 27th July 2020]

Peate, I. and Gormley-Fleming, E. (2019). *Fundamentals of Children's Anatomy and Physiology*. Somerset: Wiley.

Waugh, A. (2014). *Ross and Wilson anatomy and physiology in health and illness*. Edinburgh: Churchill Livingstone.

White, S. (2014) The Heart Wall. [Online] Available from: http://teachmeanatomy.info/thorax/organs/heart/heart-wall/ [Accessed 7th January 2019]

Yen Ho, S. (2013) Anatomy of the Heart. *Textbook of Cardiology.* [Online] Available from: http://www.textbookofcardiology.org/wiki/Anatomy_of_the_Heart [Accessed 27th July 2020]

www.nottingham.ac.uk Cardiology Teaching Package. Available from: http://www.nottingham.ac.uk/nursing/practice/resources/cardiology/function/sinus_tachycardia.php [Accessed 27th July 2020]

Key Resources

Key Resources - Literature

- Aaronson, P.I., Ward, J.P.T. and Connolly, M.J. (2013) The Cardiovascular System at a Glance (4th Ed.). Chichester: Wiley-Blackwell.
- Cockett, A and Day, H. (Eds.) (2010) Children's High Dependency Nursing. Chichester: Wiley-Blackwell.
- Dixon, M., Crawford, D., Teasdale, D. and Murphy, J. (Eds.) (2009) Nursing the Highly Dependent Child or Infant: A Manual of Care. Chichester: Wiley- Blackwell.
    - Everett,A and Lim,S (2011)Illustrated Field Guide to Congenital Heart Disease and Repair(3rd Ed).Scientific Software Solutions.
- Gatzoulis, M.A., Swam, L., Therrien, J., Pantley, G.A. and Braunwald, E. (2005) Adult Congeital heart Disease: A Practical Guide. Oxford: Blackwell.
- Gleason, M.M., Rychik, J. and Shaddy, R.E. (2012) Pediatric Practice: Cardiology. New York: McGraw Hill.
- Hazinski, M.F. (2012) Nursing Care of the Critically Ill Child (3rd Ed.). London: Elsevier Mosby.
- Klabunde, R.E. (2012) Cardiovascular Physiology Concepts (2nd Ed). Philadelphia: Wolters Kluwer Health.
- Lissauer, T. and Clayden, G (2011) Illustrated Textbook of Paediatrics (4th Edition). Edinburgh, UK: Mosby.
- MacGregor, J. (2008) Introduction to the Anatomy and Physiology of Children. London, UK: Routledge.
- Macqueen, S., Bruce, A.B., and Gibson, F. (Ed.) (2012) The Great Ormond Street Hospital Manual of Children's Nursing Practices. Wiley-Blackwell: Oxford.
- Mavroudis, C., Backer, C. and Idriss, R.F. (2013) Pediatric Cardiac Surgery (4th Ed). Wiley-Blackwell.
- Miall, L. Rudolf, M. and Smith, D. (2012) Paediatrics at a Glance (3rd Ed). Oxford, UK: Blackwell Publishing.
- Munoz, R., Morell, V., da Cruz, E. and Vetterly, C.G. (Eds) (2010) Critical Care of Children with Heart Disease. Springer.
- Park, M.K. (2014) Park's Pediatric Cardiology for Practitioners (6th Ed). Philadelphia: Mosby Elsevier.
- Samuels, M., and Wieteska, S. (Ed.) (2011) Advanced Paediatric Life Support (5th Ed.). Oxford: Wiley-Blackwell.
- Thorne, S. and Clift, P. (Eds) (2009) Adult Congenital Heart Disease. Oxford University Press.

Key Resources - Journals

- American Heart Journal
- Archives of Diseases in Childhood
- British Journal of Nursing
- Chest
- Circulation
- European Heart Journal
- European Journal of Heart Failure
- Evidence-based child health
- Evidence Based Nursing
- Heart
- Intensive and Critical Care Nursing
- Issues in Comprehensive Paediatric Nursing
- Journal of Child Health Care
- Journal of Clinical Nursing
- Journal of Advanced Nursing
- Journal of Paediatrics
- Journal of Paediatrics and Child Health
- Journal of Paediatric Nursing
- Nursing in Critical Care
- Paediatrics and child health
- Paediatric Clinics of North America
- Paediatric Nursing

Key Resources - Websites

- http://www.dh.gov.uk
- http://mmcts.oxfordjournals.org
- http://www.nice.org.uk
- http://www.nmc-uk.org
- http://www.rcn.org.uk
- http://www.resus.org.uk
- http://www.swswchd.co.uk
- http://www.pted.org/

Cardiac Charities

- British Heart Foundation: https://www.bhf.org.uk/informationsupport/conditions/congenital-heart-disease

- Heart Heroes: https://heartheroes.co.uk

- Little Heart Matters: https://www.lhm.org.uk

- Youth at Heart: https://www.youthatheart.co.uk

Cardiac Education Courses

There are cardiac modules available through the Faculty of Children's Nurse Education, based at Bristol Royal Hospital for Children, which you may wish to undertake to increase your knowledge of cardiac anomalies. These are led by the Cardiac Nurse Educator, Alison Wood, who is an experienced cardiac ward nurse.

Children's Cardiac Nursing 1 course covers foundation knowledge of normal cardiac anatomy and physiology, ECG and arrhythmias, as well as considering the common cardiac defects and the care pathways for these patients. This course would be suitable for registered paediatric nurses working regularly with children with infants and children with congenital heart conditions.

Children's Cardiac Nursing 2 is an advanced course aimed at developing a deeper level of understanding of heart conditions including some of the less common defects. This course would suit paediatric nurses working within either a cardiac speciality area or a Paediatric Intensive Care Unit which frequently provides care for children with heart conditions.

For more details you can email directly (alison.wood@uhbw.nhs.uk) or for course dates and availability contact the course administrator (FacultyOfChildrensNurseEducation@uhbw.nhs.uk).

Feedback on our Cardiac Workbook

We would love feedback on our work book. Please email carla.sims@uhbw.nhs.uk with any suggestions of improvements/material you would like adding that would be useful

Copyright

We hope you have enhanced your Paediatric Cardiac Knowledge by completing our Cardiac Workbook

Thank you