London School of Paediatrics Cardiology Learning Pack

This learning pack can be used for local teaching and for individual reading and reflection. Several activities have been designed and they have been mapped to the RCPCH Progress Curriculum.

Feel free to use any or all of this pack in your department. If you wish, you can reflect on the learning activity and upload to you e-portfolio.

Comments/feedback to mehrengise.cooper@nhs.net.

Congenital cardiac disease: duct-dependant pulmonary circulations

Objectives:

- Understand how congenital heart disease is screened for, and counselled, antenatally
- Explain to the parents how you will manage the condition
- Communicate factually correct information effectively to the parents without jargon
- Recognise the impact on children and their families

Activity: Group discussion (10 minutes)

Facilitator: Consultant, Registrar
Participants: Level 1-3 Trainees

Instructions for facilitator: use the questions below to facilitate a group discussion

1. *What congenital cardiac lesions have a duct-dependant pulmonary circulation*
2. *How are congenital heart diseases screened for antenatally in the UK*
3. *What happens if congenital heart disease is suspected antenatally?*

Resources:

1. Congenital cardiac lesions with a duct-dependant pulmonary circulation https://www.uptodate.com/contents/identifying-newborns-with-critical-congenital-heart-disease?search=cyanotic%20congenital%20heart%20disease&source=search_result&selectedTitle=5~88&usage_type=default&display_rank=5#H1312102076
2. The 5 required views in the fetal cardiac protocol https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/749742/NHS_fetal_anomaly_screening_programme_handbook_FINAL1.2_18.10.18.pdf
3. The paediatric cardiac pathway.

https://www.england.nhs.uk/wp-content/uploads/2018/08/Congenital-heart-disease-standards-and-specifications.pdf

Congenital cardiac disease: duct-dependant pulmonary circulations

Activity 2: Communication simulation

Facilitator: Consultant, Registrar

Participants: Level 1,2 or 3 Trainees

Role Player: any

Case

You are working in the Paediatric Department in a District General Hospital. You will be talking to the mother of Toby, a 3 day old male infant who presented to A&E with cyanosis. The antenatal scans were unremarkable and Toby was discharged from hospital on Day 1 of life. He is currently in Paediatric A&E Resus and is on IV Dinoprostone. He has had an echocardiogram demonstrating a large ventricular septal defect with predominantly right-to-left flow, an overriding aorta, severe pulmonary stenosis and right ventricular hypertrophy. There is a patent ductus arteriosus with left-to-right flow. Your colleague has referred the patient to a Paediatric Cardiology centre and is on the phone to the Neonatal Transport Team.

Facilitator instructions:

Set up a communication scenario with the participant and the parent, following the timing of the MRCPH clinical Communication station.

Lead on the discussion and feedback following the conversation.

Participant instructions:

- *Talk to Toby's mother about the echo findings, transfer to a Paediatric Cardiology centre and next steps*

Role Player instructions:

- *Your antenatal scans were unremarkable and you were expecting a healthy baby*
- *You are very anxious that there is an underlying problem*
- *If not volunteered – ask if they have a diagram demonstrating the condition*
- *Enquire about the next steps for investigation and management*

Discussion and Feedback

Facilitate discussion on the following:*:*

- *Introduction and structured consultation*
- *Exploration of the mother's ideas, concerns and expectations*
- *Use of description and diagram of normal blood flow in the heart and the lungs, the blood flow in this condition, the rationale for Dinoprostone, the requirement for transfer to a Paediatric Cardiology centre, the initial steps that will occur at the Paediatric Cardiology centre and the likely requirement for surgical intervention.*
- *Avoidance of jargon*

Resources:

1. Tetralogy of Fallot.
http://www.pted.org/?id=tetralogyfallot1
2. Bailliard F, Anderson R. (2009). Tetralogy of Fallot. *Orphanet J Rare Dis*. 4(2)
3. Tetralogy of Fallot.
https://www.uptodate.com/contents/pathophysiology-clinical-features-and-diagnosis-of-tetralogy-of-fallot
4. NTS duct-dependant cardiac lesions.
https://london-nts.nhs.uk/wp-content/uploads/2020/09/NTS-Duct-Dependant-Cardiac-Lesions.pdf
5. Patient information leaflet on Tetralogy of Fallot.
https://www.bhf.org.uk/informationsupport/publications/children-and-young-people/understanding-your-childs-heart---tetralogy-of-fallot

Activity: Safe prescription of IV Dinoprostone

Dinoprostone is a commonly used medication in duct dependant cardiac lesions.

Objectives:

- Safe prescribing practices
- Prescribing in duct-dependant cardiac lesions

Facilitator: Consultant, Registrar or paediatric pharmacist
Participant: Level 1-3 Trainee
Equipment: Fluid prescription chart, local guideline

Tasks:

1. Complete the Script Paediatric Safe Prescribing Cardiology module: https://www.safeprescriber.org/paediatric/
2. Prescribe IV Dinoprostone, at a rate of 20 nanograms/kg/minute, for a 3 day old male infant who weighs 3700g.
3. Explain to another team member (e.g. nursing staff, another doctor) the indications for Dinoprostone, it's mechanism of action and its side effects
4. Instruct the nurses on what monitoring is required for a child who is on IV Dinoprostone

Resources:

1. Dinoprostone. BNFc. https://bnfc.nice.org.uk/drug/dinoprostone.html
2. NTS Duct-dependant cardiac lesions. https://london-nts.nhs.uk/wp-content/uploads/2020/09/NTS-Duct-Dependant-Cardiac-Lesions.pdf

Activity- Group Discussion (10 minutes) – Discuss the impact of congenital heart disease

Facilitator: Consultant, Registrar

Participants: Level 1-3 Trainees

Instructions for facilitator:

Use the questions below to facilitate a group discussion

1. *What impact does congenital heart disease have upon childhood?*

   Consider prolonged hospital admissions, impact on infant feeding, missed school days in childhood, psychological impact etc

2. *Bring in experiences of congenital heart disease from participants*

3. *How can we support children and their families? Who can help from the MDT?*

Resources:

1. Nutrition and school.
http://www.pted.org/?id=list#1
2. Garcia Guerra G, Robertson CM, Alton GY, Joffe AR, Dinu IA, Nicholas D, Ross DB, Rebeyka IM; Western Canadian Complex Pediatric Therapies Follow-up Group. Quality of life 4 years after complex heart surgery in infancy. *J Thorac Cardiovasc Surg.* 2013 Feb;145(2):482-488.e2. doi: 10.1016/j.jtcvs.2012.03.050.
3. Latal B, Helfricht S, Fischer JE, Bauersfeld U, Landolt MA. Psychological adjustment and quality of life in children and adolescents following open-heart surgery for congenital heart disease: a systematic review. *BMC Pediatr.* 2009 Jan 22;9:6. doi: 10.1186/1471-2431-9-6.
4. Children with congenital heart disease – understanding your child's heart. https://www.bhf.org.uk/informationsupport/publications/children-and-young-people/dvd24-children-with-congenital-heart-disease---understanding-your-childs-heart
5. Teen Heart.
https://www.bhf.org.uk/informationsupport/support/children-and-young-people/teen-heartdont-delete
6. Preparing for surgery.
https://www.bhf.org.uk/informationsupport/support/children-and-young-people/preparing-for-surgery

Activity - Hypoxic Spells Group discussion

Objectives:

- Understand the pathophysiology of a hypoxic spell
- Understand the treatment of a hypoxic spell
- Explain to parents how to manage a hypoxic spell
- Communicate factually correct information effectively to the parents without jargon

Activity one: Group discussion (10 minutes)

Facilitator: Consultant, Registrar
Participants: Level 1-3 Trainees

Instructions for facilitator: use the questions below to facilitate a group discussion

1. *What clinical symptoms and signs characterise a hypoxic spell*
2. *Why do hypoxic spells occur*
3. *How are hypoxic spells treated*

Resources:

1. Pathophysiology, symptoms and management of hypoxic spells. Bailliard F, Anderson R. (2009). Tetralogy of Fallot. *Orphanet J Rare Dis*. 4(2)
2. Management of a hypoxic spell.
https://www.uptodate.com/contents/management-and-outcome-of-tetralogy-of-fallot?sectionName=Tet%20spells&search=tetralogy%20of%20fallot&topicRef=5769&anchor=H11096360&source=see_link#H11096360

Activity- Hypoxic Spells Communication station

Facilitator: Consultant, Registrar
Participants: Level 1-3 Trainees
Role Player: any

Case
You are working in the Paediatric Department in a District General Hospital. You will be talking to the mother of Alice, a 2 month old girl who presented to A&E with a hypoxic spell. At presentation, she was agitated, tachycardic, tachypnoeic and cyanotic. Alice was diagnosed with acyanotic Tetralogy of Fallot in the neonatal period. The hypoxic spell has been treated, Alice's Paediatric Cardiology team have been updated. Alice is now clinically well and medically fit for discharge.

Facilitator instructions:
Set up a communication scenario with the participant and the parent, following the timing of the MRCPH clinical Communication station. Lead on the discussion and feedback following the conversation.

Assess the following:

- *Introduction and structured consultation*
- *Exploration of the mother's ideas, concerns and expectations*
- *Explanation to include a description of the symptoms and signs of a hypoxic spell, contributing factors and how to minimise these, the use of positioning and comforting the infant, calling an ambulance immediately and to inform Alice's Paediatric Cardiology team if this is the first hypoxic spell she has had, or if the frequency of the hypoxic spells is increasing*
- *Avoidance of jargon*

Participant instructions:

- *Speak with Alice's mother about what hypoxic spells are, what factors may contribute to a hypoxic spell, how to treat a hypoxic spell at home, and when to bring Alice to A&E for treatment of the hypoxic spell.*

Role Player instructions:

- *You are pleased that Alice responded well to treatment and now appears well*
- *At the last Paediatric Cardiology clinic appointment, the doctor explained what to do if Alice is looking blue and crying a lot, but you have forgotten what was said*
- *You are anxious about what to do next time if this happens again*

Resources:

1. Patient information leaflet on hypoxic spells. https://alderhey.nhs.uk/application/files/8715/7546/8702/Cyanotic_Spells_Blue_Spells_in_Tetralogy_of_Fallot_-_PIAG_214.pdf

Activity: Group discussion (30 minutes)

Achieving pulmonary blood flow demystified

Objectives:

- Understand the different types of conduits and shunts used to achieve pulmonary blood flow in cardiac lesions with duct-dependant pulmonary blood flow
- Understand the physiological changes that occur in a pulmonary circulation that is dependant on a conduit or shunt, and how this may give rise to complications

Facilitator: Consultant, Registrar
Participants: Level 1-3 Trainees

Task:

- Discuss congenital cardiac disease- which cardiac anomalies cause cyanosis?
- Discuss the options available for achieving pulmonary blood flow in cardiac lesions with duct-dependant pulmonary blood flow
- Discuss the physiological complications that might arise in a pulmonary circulation that is dependant on a conduit or shunt.

Resources:

- Modified Blalock-Taussig shunt in Tetralogy of Fallot. http://www.pted.org/?id=doubleoutlet3
- Modified Blalock-Taussig shunt in Pulmonary Atresia. http://www.pted.org/?id=pulmonaryatresia3
- PDA stent in Pulmonary Atresia. https://www.uptodate.com/contents/pulmonary-atresia-with-intact-ventricular-septum-pa-ivs?search=pulmonary%20atresia&source=search_result&selectedTitle=1~150&usage_type=default&display_rank=1
- Blalock-Taussig shunt in Ebstein's anomaly. http://www.pted.org/?id=ebstein3
- Kiran U, Aggarwal S, Choudhary A, Uma B, Kapoor PM. The blalock and taussig shunt revisited. *Ann Card Anaesth*. 2017;20(3):323-330. doi:10.4103/aca.ACA_80_17
- Blalock-Taussig shunt, bidirectional Glenn repair and Fontan in Tricuspid Atresia. http://www.pted.org/?id=tricuspidatresia3
- Balloon valvuloplasty or transannular patch in Pulmonary Valve Stenosis. http://www.pted.org/?id=pulmonarystenosis3
- Küçük M, Özdemir R, Karaçelik M, Doksöz Ö, Karadeniz C, Yozgat Y, Meşe T, Sarıosmanoğlu N. Risk Factors for Thrombosis, Overshunting and Death in Infants after Modified Blalock-Taussig Shunt. Acta Cardiol Sin. 2016 May;32(3):337-42. doi: 10.6515/acs20150731a.
- Dirks V, Prêtre R, Knirsch W, Valsangiacomo Buechel ER, Seifert B, Schweiger M, Hübler M, Dave H. Modified Blalock Taussig shunt: a not-so-simple palliative procedure. Eur J Cardiothorac Surg. 2013 Dec;44(6):1096-102. doi: 10.1093/ejcts/ezt172.

· Agarwal A, Firdouse M, Brar N, et al. Incidence and Management of Thrombotic and Thromboembolic Complications Following the Superior Cavopulmonary Anastomosis Procedure: A Literature Review. *Clin Appl Thromb Hemost*. 2018;24(3):405-415. doi:10.1177/1076029617739702
· Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. *Heart* 2016;**102:**1081-1086.

Further reading:

· Park MK (2016). Park's The Pediatric Cardiology Handbook 5th ed. Philadelphia: Saunders.
· Illustrated field guide to congenital heart disease and repair. 2nd ed. Allen D. Everett and D. Scott Lim

Activity: Communication scenario - Sudden Arrhythmic Death Syndrome (SADS)

Duration
1hr

Objective
Understand causes and presentations of sudden arrhythmic death syndrome (SADS)

Number of people required
4 – Sabina, Sabina's mother, Registrar, Facilitator

Role
You are the paediatric registrar

Case
Sabina is a 15 year old girl who presents to the paediatric A and E at a DGH after tripping in her bedroom. She has hit her head needing 5 stitches which the ANP has kindly volunteered to stitch, but she has asked you to take the rest of the history and examination. Sabina is accompanied by her mother.

Instructions
Take the history from Sabina and her mother

Patient instructions
She's not really sure what has happened. Thinks she might have tripped over the new dog. Remembers being in her room and blood on her head and the dog was there and was licking some of the blood off the floor.

Parent instructions
Has once before had a funny turn at school when a fire alarm went off and was sent home. Past Hx Anxiety only. Only recent change in lifestyle is a new dog in the house which belongs to Sabina's older brother. This has been a problem because the neighbours complain about the dog's barking. Sabina's Mother heard the dog bark then a bang and when she went into the room Sabina was making some jerky movements for a few seconds and then woke up and was a little confused for a minute or so but then normal.

Activity: Flipped learning and brainstorm sudden arrhythmic death syndrome (SADS)

Following on from the communication scenario on the previous page,

Facilitator instructions for group brainstorm + flipped classroom

1. Establish differential diagnosis for cardiac syncope and what investigations would be best and when is the optimal time for these investigations

- Structural –CXR, ECG, echo, troponin, CK, BNP, ProBNP, genetics
  - Valve disease – Bicuspid valve with aortic Stenosis
  - Cardiomyopathy – HCM/DCM/Arrhythmogenic right ventricular cardiomyopathy
- Coronary – ECG, troponin, CK, echo
  - Anomalous left coronary from right coronary cusp
- Electrical
  - Conduction pathway – ECG, ep, genetics
    - WPW (very rare to get syncope) – only with atrial Fib
    - CHB
  - Channelopathies – ECG, genetics
    - Long QT
    - Brugada
    - CPVT

2. **How can SADS (sudden arrhythmia death syndrome) present in ED?**

- Usually don't present with palpitations – this is more SVT
  - Syncope – most common – remember syncope in toddlers is not always breath holding – get an ECG
  - Presyncope – if very transient or conscious VT
  - Seizure-like – Tonic clonic movements are common in vasovagal and cardiac syncope and remember that fever is a trigger for Brugada so it might not be a simple febrile seizure
  - Trauma – syncope from trauma or trauma from syncope
  - Arrest

3. **Brainstorm meds that cause long QT**

    - www.crediblemeds.org – azithromycin, ondansetron most common
    - Reference ranges

4. **Microteaching session for colleagues/ flipped classroom**
    - Demonstrate to the F2 how to measure corrected QT with reference to some more challenging ECGs

Resources

PEM ED Paediatric Syncope Podcast Available at: https://www.pemed.org/blog/2014/12/21/pediatric-syncope-dfo-done-fell-out and https://www.pemed.org/blog/2015/4/6/pediatric-syncope-part-deuxstill-fell-out

Waddell-Smith K, Gow RM, Skinner JR. How to measure a QT interval. Med J Aust. 2017 Aug 7;207(3):107-110. doi: 10.5694/mja16.00442.

Crediblemeds Available at: https://crediblemeds.org/

Elayne Forbes. Cardiac Syncope, Don't Forget the Bubbles, 2015. Available at: https://doi.org/10.31440/DFTB.6610

Elayne Forbes. Syncope ECGs, Don't Forget the Bubbles, 2015. Available at: https://doi.org/10.31440/DFTB.6615

Activity: Low-fidelity simulation – The infant with heart failure

Duration
20 min

Objective
Understand the presentation, differential and management for acute heart failure in infants and young children

Facilitator
Consultant or registrar

Number of people required
5 – Jake, Jake's mother, Registrar, Resus Nurse, Facilitator

Role
You are the paediatric registrar in a DGH

Case
Jake is a 5 month old who presents with fever. He has a normal antenatal course with normal antenatal screening in the UK and a normal NIPE exam but saturations were not checked. He has had 4 days of a runny nose and fever but has become more breathless in the past day.

Instructions for Registrar Participant
Please assess Jake and manage appropriately

Further history to be provided by Jake's mother
On further history you find he has always been a poor feeder. He becomes breathless at the breast and sweaty and takes about an hour to take a feed. This has become worse in the past week where the feeds are only half-finished even after 1.5 hours. He has also not been putting on much weight though she is not sure about the centiles. The child has already been given some paracetamol 30 minutes prior to your assessment.

Initial assessment
Not toxic looking. Grossly small for age. Miserable. Cap refill 3-4 seconds centrally. Cool peripherally. Some mild increased WOB. HSDNM.

Chest – scattered wheeze and creps. Abdo SNT. 1-2cm hepatomegaly. Palpable femorals. No peripheral edema.

Sats 89-92% in air. RR 60. HR 170 regular. BP 75/45 in all 4 limbs. Febrile – 38.5 C

Initial management
Oxygen?
Intravenous access with bloods and gas?

The gas shows:
pH 7.2
pCO2 6.8
pO2 5.5
Hb 100
Bicarb 20
Lactate 3.8

Cultures and antibiotics?
Fluid bolus? How much and what fluid?
Xray?
When will you reassess?

Further assessment
30 minutes later
The capillary refill and cool peripheries has not changed. The sats are now 86%-88% in air and RR 75-80. The HR is 180 and the temperature is now 37.2C.
The gas shows the ph is 7.15 pCO2 6.5 pO2 5 Bicarb 18 Lactate 5.6

(If an Xray was ordered say – just had a covid patient and deep cleaning the room so not available yet)

Further management
A further fluid bolus?
When to reassess?
Mobile CXR is back online – cardiomegaly with pulmonary plethora

What is the management?

- Increased O2+/- facemask +/- ?NIV – CPAP vs BIPAP? What settings might you start at? Who would you talk to about this?

- Frusemide? as evidence of fluid overload – what dose and what route of administration? Ask to write it up on the drug chart
- Further fluid?
- Which inotrope? Ask to write dopamine on the drug chart
- ECG – what will you see?
- Call your local retrieval team (CATS/STRS ask you to call cardiology while they are organizing a conference call with the ECMO centre)
- Cardiology ask you to write up milrinone – write up milrinone
- Have conference call

Scenario ends after you have placed patient on NIV, given frusemide bolus, have written up milrinone and had conference call with an ECMO centre.

Facilitator instructions: Debrief discussion

1. What is the differential diagnosis for acute heart failure in infants

- Myocarditis
- Cardiomyopathy
- Undiagnosed CHD – eg TAPVD
- Coronary abnormalities - Most commonly ALCAPA
- Secondary to arrhythmia

2. A keen SHO raises the possibility of PIMS-TS as a cause of acute heart failure and asks whether this would change the management

- What are the cardiac manifestations of PIMS-TS?
- What lab tests will you now send if you suspect PIMS-TS?
- What extra-management options should now be considered?

Reference:
Evelina Guideline: Paediatric Critical Care: PIMS-TS Paediatric Multisystem Inflammatory Syndrome temporally associated with SARS-CoV2. 2020. Available at: https://www.evelinalondon.nhs.uk/resources/our-services/hospital/south-thames-retrieval-service/pims-ts-paediatric-multisystem-inflammatory-syndrome-temporally-associated-with-sars-cov2.pdf

Activity Case based discussion- The older child with ST segment changes +/- troponin rise

Duration
20 minutes

Objective
To demonstrate the differential diagnosis and investigation pathway for ST segment changes and troponin elevation

Facilitator- Consultant or registrar

Participants- Level 1-3 Trainees

Number of people
3 – Samuel, registrar, facilitator

Role
Paediatric registrar at DGH

Case

Samuel is a 15 yr old boy who presents with a day of chest discomfort and feels like he can't take a full breath. He has been having his regular salbutamol on and off the whole day but doesn't think it's making much of a difference.

He has a past history of asthma. Also when he was 3 years old he had fever and red eyes and he received an intravenous medication made from blood products from a donor but it was not a transfusion. He and his mother cannot remember much more about it.

Instructions
Please examine Samuel and initiate a management plan

Assessment

Well looking. Grossly normal height and weight. Cap refill 2 seconds centrally. Warm peripherally. No mild increase WOB. HSDNM. Chest clear. Abdo SNT. no hepatomegaly. Palpable femorals. No peripheral oedema.

Sats 99% in air. RR 25. HR 120 reg . BP 110/60 in all 4 limbs. Afebrile

Initial management
CXR? – no pneumothorax
ECG? – showed some ST segment elevation on the anterior chest leads
Troponin? You discuss with the consultant on call – they think it's probably just pericarditis but given the history of Kawasaki – they ask you to send a troponin.

The troponin is 100.

Activity- Communication simulation

Speak to Cardiology

You speak to cardiology who suggest keeping overnight, serial troponin levels and CK and serial ECGs a proBNP - and an echo if it can be done but otherwise this can probably be done as an outpatient.

Discussion around some causes of:

- High troponin and ST segment changes on ECG
    - Perimyocarditis
    - Acute heart failure
    - Coronary abnormalities
        - Kawasaki aneurysm
        - Anomalous left coronary artery from right coronary cusp

- **High troponin alone without ST segment changes**
    - Can be cardiac as above but also non-cardiac eg. PE, sepsis, tachyarrhythmia, sympathomimetic like Beta agonists eg Salbutamol

Activity- Communication simulation

Communication - Explain to Samuel what troponin is and what ST segment changes are with the use of a diagram.

Communication – Explain to the 4th year medical student how to discern between high-take off, pericarditis ST changes, and ischaemia on ECG

Activity- Safe prescribing

Drug prescribing – Discuss the considerations around how to treat Samuel's pericarditis and write up prescriptions for treatment options (Write up NSAIDs but consider whether this will impact Samuel's asthma)

Resources

Jordan Evans, Megan Thomas, Amos Wong and Jeff Morgan. Ten 'not to be missed' paediatric ECGs, Don't Forget the Bubbles, 2020. Available at: https://doi.org/10.31440/DFTB.29306