Learning pack: Haematology

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.

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New Diagnosis of Thalassaemia

Objectives

- Understand how thalassaemia is screened for in the UK
- Draw a pedigree
- Explain the inheritance of thalassaemia
- Explain the long-term implications of a diagnosis of thalassaemia to parents of a newly-diagnosed child

Activity 1: Group discussion (20-30 minutes)

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

1. What are the different types of thalassaemia?
2. How is thalassaemia screened for in the UK?
3. What happens if a patient has a positive screening test?

Activity 2: Work individually (10 minutes)

Review the short case introduction below:

Gina and Enzo have recently moved to the UK from Southern Italy and are planning on starting a family. They are aware that Gina will have some blood tests at her booking appointment when she becomes pregnant, but Enzo has a family history of beta thalassaemia and they are concerned about whether their child will get it. Enzo's grandmother had beta thalassaemia major, and Enzo's brother has it too. No one else in his family is affected to their knowledge.

1. Draw a pedigree chart for Enzo's family including his generation, his parents, and his grandparents

Activity 3: Work in pairs or threes (20-30 minutes)

Gina and Enzo undergo testing and find that they are both carriers of beta thalassaemia. Gina becomes pregnant, and declines any invasive testing during pregnancy. Shortly after her baby is delivered she asks to speak to the paediatric doctor regarding the chances of her child having beta thalassaemia, and what this would mean if they test positive.

1. Choose one person to play Gina. Explain to her the chances of her baby having beta thalassaemia, and how you will test for this.
2. Swap over the person playing Gina. Explain to her the longer-term implications of a diagnosis of beta thalassaemia major in her baby.

References

1. Types of thalassaemia: https://www.uclh.nhs.uk/OurServices/ServiceA-Z/Cancer/CBD/THAL/Pages/Home.aspx

2. UK screening for thalassaemia and pattern of inheritance: https://www.nhs.uk/conditions/pregnancy-and-baby/screening-sickle-cell-thalassaemia-pregnant/
3. Testing for thalassaemia: https://labtestsonline.org.uk/conditions/thalassaemia
4. Drawing pedigree charts (the basics!): https://www.youtube.com/watch?v=Gd09V2AkZv4
5. Implications of a diagnosis of beta thalassaemia: https://patient.info/allergies-blood-immune/thalassaemia-leaflet

Breaking bad news: positive G6PDD result from prolonged jaundice screen

Objectives

1. Understand the testing process for G6PD deficiency
2. Explain to a parent how the diagnosis is confirmed after a positive screen
3. Outline to a parent how G6PD deficiency is managed

Role play activity (30 minutes): pairs or threes

One or two of you will play parent(s), and the other will play the paediatric registrar.

Read the case introductions below:

Parent(s): Your son, Milo, is 3 weeks old. He was found to have jaundice on day 3. Although his bilirubin level has never crossed the phototherapy threshold, he still looks yellow so your midwife referred you to the Prolonged Jaundice clinic. He has been feeding well, gaining weight, and you have no other concerns about him. The doctor said he looked well, but yellow, and so did a set of blood tests. You have been called back into the hospital to discuss the results.

Doctor: You have completed a prolonged jaundice screen on Milo. This shows that his bilirubin level is below the treatment threshold at 77, but his G6PD activity result is <10% of normal. You plan to refer him for genetic testing to confirm a diagnosis of G6PD deficiency.

Resources

1. G6PDD Favism Association: https://www.g6pd.org/en/G6PDDeficiency.aspx
2. RCH Melbourne: https://www.rch.org.au/kidsinfo/fact_sheets/G6PD_deficiency/
3. Baby's First Test: https://www.babysfirsttest.org/newborn-screening/conditions/glucose-6-phosphate-dehydrogenase-deficiency
4. Lab tests online: https://labtestsonline.org.uk/tests/g6pd

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Counselling parents: Vitamin K

Objectives

1. Reflect on your experiences of discussing interventions with parents who do not initially agree with your proposed course of action
2. Explain the rationale for IM vitamin K
3. Gain confidence in approaching, and ending, difficult discussions

Group case-based discussion (45 minutes – 1 hour)

Case scenario: You are the neonatal SHO on call for labour ward. You receive a bleep from a midwife asking you to attend a forceps delivery for a term baby due to failure to progress. It has been an uncomplicated pregnancy, there are no risk factors for sepsis, and the baby is born in good condition with no resuscitation required.

The midwife bleeps you again just after you leave asking you to come and speak to the baby's mother, Angela, because she has refused consent for intramuscular vitamin K.

Discuss as a group:

1. Why might Angela be refusing the injection?
2. How do you feel when you are asked to speak to families in these scenarios?
3. Have you ever felt like you have been in this situation and found it particularly difficult, or particularly easy? Why was that?

Case scenario: You attend and speak to Angela. She has 2 older children, both of whom did not have vitamin K after she read in the 1990s that the injection might be linked to leukaemia. Both of her previous children have grown up to be healthy with no issues. She also isn't convinced of the safety of the other ingredients in the injection and wants to know why her baby can't get all the vitamins she needs from her breast milk.

Discuss as a group:

1. How can you approach this discussion with Angela?
2. Are there any alternatives to the IM Vitamin K injection?
3. What are the important points to ensure she understands when making this decision?
4. How can you end the conversation with Angela if she still refuses the vitamin K?

Resources

1. GSTT Vitamin K leaflet: https://www.guysandstthomas.nhs.uk/resources/patient-information/maternity/vitamin-K-and-your-newborn-baby.pdf
2. NCT Information for Parents: https://www.nct.org.uk/labour-birth/after-your-baby-born/vitamin-k-and-newborns-what-you-need-know
3. ESPGHAN Position Paper: https://pubmed.ncbi.nlm.nih.gov/27050049/?from_term=vitamin+k+neonate&from_pos=4

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Management of suspected sickle cell acute chest syndrome

Objectives

1. Understand the pathophysiology of acute chest syndrome
2. Outline common signs of acute chest syndrome
3. Outline the management of acute chest syndrome
4. Recap your local guideline for management of acute chest syndrome
5. Explain to a parent, or older child, how you will manage their condition

Group discussion (1 hour)

Instructions for facilitator (consultant): Use the learning objectives above to guide a discussion regarding acute chest syndrome in children. Resources are listed below to provide information for trainees; they may want to read this information prior to attempting to answer the questions above.

Instructions for trainees: Use the resources below and your own knowledge and experiences to participate in the group discussion.

Resources

1. BSH Acute chest syndrome guideline: https://b-s-h.org.uk/guidelines/guidelines/management-of-acute-chest-syndrome-in-sickle-cell-disease/
2. European Medical Journal review paper: https://www.emjreviews.com/hematology/article/editors-pick-acute-chest-syndrome-in-children-with-sickle-cell-disease-current-perspectives-on-pathogenesis-and-treatment/

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Exchange transfusions

Objectives

1. Understand the indications for an exchange transfusion
2. Gather the necessary equipment and documents to undertake an exchange transfusion
3. Explain the step-by-step process
4. Outline possible complications and how to monitor for them
5. Recap your local guideline for exchange transfusions

Group discussion (1 hour)

Instructions for facilitator (consultant): Use the learning objectives above to guide a discussion regarding exchange transfusions. Larger groups may find it helpful to break into smaller groups, with more senior trainees supporting junior trainees to answer the above questions. Resources are listed below to provide information for trainees; they may want to read this information prior to attempting to answer the questions above.

Instructions for trainees: Use the resources below and your own knowledge and experiences to participate in the group discussion.

Resources

1. South Thames Sickle & Thalassaemia Network guideline (p4): https://www.ststn.co.uk/wp-content/uploads/2012/02/k.-transfusion-guideline-2010_TV.1.pdf
2. Neonatal Exchange Transfusion guideline with equipment list and information about complications: https://www.rch.org.au/uploadedFiles/Main/Content/neonatal_rch/EXCHANGE_TRANSFUSION.pdf
3. Evelina London information for parents and carers: https://www.evelinalondon.nhs.uk/resources/patient-information/exchange-transfusion-information-for-parents.pdf

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Transitioning to adult care

Objectives

1. Consider possible issues that may arise when a paediatric patient transitions to adult care
2. Reflect on your own experiences working with adolescent patients
3. Consider strategies to improve transition
4. Review current evidence regarding optimising transition

Group session (45 minutes – 1 hour): split into groups of 3-5 and work on the following questions

Discussion in groups: Consider long-term diagnoses such as sickle cell, thalassaemia, and other non-haematological conditions that require young people to be in frequent contact with specialist services.

1. What are some of the barriers to transition for young people with long-term conditions like sickle cell disease?
2. What might be some of the advantages of transitioning to adult care?
3. Think about older adolescent patients you have met with sickle cell disease, thalassaemia, cystic fibrosis, or any other long-term condition. How do you think they felt about transitioning to adult services, and why?

Feedback your discussions to the wider group.

Brainstorm session: Focusing on 2-3 of the barriers you identified, discuss how you could design a transition service that would address these. Who might be involved? Where might it take place?

Feedback your ideas to the wider group.

Individually: Review the evidence and suggestions in the resources below.

Discuss as a wider group: How do these suggestions compare to your ideas? Are there any surprises?

Resources

1. Health Service Journal: UK Multidisciplinary statement on optimising transition https://www.hsj.gr/medicine/optimising-the-transition-from-paediatric-to-adult-care-model-for-people-with-sickle-cell-disease-a-uk-multidisciplinary-consensu.php?aid=24486
2. Two videos from the CDC presenting young people's stories of transition (Kevin's Decision and Calvanay's Advice): https://www.cdc.gov/ncbddd/sicklecell/materials/video.html?s_cid=ncbddd_sc_v1_feat_2018_6&%20#su-p1

3. Sickle Cell Society Standards for Clinical Care of Adults with Sickle Cell Disease (p40): https://www.sicklecellsociety.org/wp-content/uploads/2018/05/Standards-for-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdf
4. South Thames Sickle & Thalassaemia Network/GSTT guideline: https://www.ststn.co.uk/wp-content/uploads/2012/02/Transition-Guidelines-GSTT.pdf

Simulation: Major Haemorrhage Protocol

Objectives

1. Understand when, and how, to activate a major haemorrhage protocol
2. List the people and products who become available to help when you active a major haemorrhage protocol
3. Gain confidence in managing a major haemorrhage

Simulation (45 minutes – 1 hour)

Instructions for facilitator: Use the case scenario from Don't Forget the Bubbles (link below) to lead a simulation with your trainees. Try to make this as realistic as possible in the current circumstances, and involve other members of the MDT who are available (particularly nursing staff). After the simulation, facilitate a debrief with your juniors and help them to answer their questions regarding major haemorrhage protocols.

Instructions for trainees: After the simulation, reflect on how it went. What could have gone better? What was done well? Discuss your questions regarding major haemorrhage protocols, and use the resources below alongside your local guidelines to answer them as a group.

Resources

1. Suggested simulation scenario + accompanying teaching material: https://dontforgetthebubbles.com/massive-transfusion-protocol/
2. JPAC guidelines: https://www.transfusionguidelines.org/transfusion-handbook/10-effective-transfusion-in-paediatric-practice/10-4-major-haemorrhage-in-infants-and-children
3. BJA review, including pathophysiology and definitions: https://academic.oup.com/bjaed/article/16/8/269/2364823

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Blood prescribing

Activity 1

Objectives

1. Review common errors in prescribing blood products for children
2. Recap local guidelines and local prescription charts
3. Understand how the chart is used by members of nursing staff

Group work (30-45 minutes)

Instructions for facilitator (consultant or senior nurse): Obtain some local prescription charts prior to the session. If possible, obtain information from your local blood bank as to what the most common prescribing errors are in your trust; if this is not possible, use the SHOT report below to insert some common errors into your charts. Distribute these amongst trainees, and invite nursing staff to attend the session. Use the charts to facilitate a group discussion using the questions below.

Instructions for trainees (and nursing staff): Review the prescription charts provided, and participate in the group discussion using the questions below.

Questions

1. What errors can you see in these prescription charts?
2. What are the most common errors that happen in the transfusion process nationally?
3. Who can collect a blood product in your clinical area?
4. What checks does the product have to go through before being administered?
5. How is a blood product administered?
6. What monitoring is doing during and after a transfusion, and by whom?

Resources

1. Serious Hazards of Transfusion 2018 Annual Report Summary https://www.shotuk.org/wp-content/uploads/myimages/130584.03-SHOT-REPORT-SUMMARY-2018_v6.pdf
2. JPAC Transfusion Handbook https://www.transfusionguidelines.org/transfusion-handbook
3. British Society for Haematology administration of blood products guideline: https://b-s-h.org.uk/guidelines/guidelines/administration-of-blood-components/
4. RCH Melbourne guideline: https://www.rch.org.au/clinicalguide/guideline_index/Blood_product_prescription/

Activity 2

Objectives:

1. Review recommendations from the Joint United Kingdom (UK) Blood Transfusion and Tissue Transplantation Services Professional Advisory Committee (JPAC) in Paediatric prescribing.

2. Practice prescribing in example cases (use local charts and guidelines where available).

Case 1:
Annie is an ex 24+0 week, now 29+6 corrected gestation, neonate who is being ventilated after a diagnosis of necrotising enterocolitis that is being managed conservatively as present. Her Haemoglobin is 70g/l and Platelets are 15 X 10^9/L. Her clotting screen is normal. Per rectal bleeding has been noted with bloody and bilious aspirates.

- Which blood products might you need to prescribe?
- What quantity?
- Are there any special considerations?
- Prescribe these on your local drug charts

Resource

- JPAC Neonatal Transfusion Guidelines: https://www.transfusionguidelines.org/transfusion-handbook/10-effective-transfusion-in-paediatric-practice/10-2-neonatal-transfusion.pdf

Case 2
You are in A&E. A 9 year-old child has been brought by ambulance after being hit by a car. He has multiple injuries and is bleeding heavily. The team has given multiple saline boluses and emergency O negative blood. They have requested further blood products to be given.

- What blood products might need to be given (other than packed red cells)?
- Please discuss in your groups what doses of Fresh Frozen Plasma, Platelets and Cryoprecipitate you might want to prescribe?
- Prescribe these on your local drug charts

The patient has continued to bleed, despite packed RBCs, FFP, platelets and cryoprecipitate.

- Are there any drugs commonly used as adjusts that need to be considered?

Resources

- JPAC Major Haemorrhage Guidelines: https://www.transfusionguidelines.org/transfusion-handbook/10-effective-transfusion-in-paediatric-practice/10-4-major-haemorrhage-in-infants-and-children.pdf
- Southampton and Oxford Retrieval Guide to Major Haemorrhage: https://www.sort.nhs.uk/Media/Guidelines/Haemorrhage-quick-reference-guide.pdf

Prescribing chelation therapy

Objectives

1. Understand the indications for chelation therapy
2. Gain confidence in prescribing chelation therapy
3. Explain the potential side effects of chelators
4. Be aware of monitoring requirements for patients receiving chelation therapy

Group discussion (30-45 minutes)

Instructions for facilitator (consultant or pharmacist): Use the questions below and the resources at the bottom of this section to facilitate a group discussion with trainees. Ask trainees to break into smaller groups to practice explaining the potential risks of chelation therapy to families.

Instructions for trainees: Use the resources below, and your own knowledge and experience, to answer the questions listed as a group.

Questions

1. What are the indications for iron chelation therapy in children with sickle cell disease, thalassaemia, or other conditions?
2. What are the options for chelating agents?
3. Where could you find information about how to prescribe these?
4. What are the potential side effects of chelators?
5. How should chelation therapy be monitored?

Resources

1. BJH: Current approach to iron chelation in children https://onlinelibrary.wiley.com/doi/pdf/10.1111/bjh.12825
2. Whittington Iron Chelation guideline www.whittington.nhs.uk%2Fdocument.ashx%3Fid%3D6171&usg=AOvVaw3MqeDXSVg_FS_6QQdklaBJ
3. UCSF Benioff Children's Hospital Oakland Thalassaemia Centre information page on thalassaemia and chelation (useful explanations): https://thalassemia.com/treatment-chelation.aspx#gsc.tab=0

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Improving efficiency of acute sickle cell painful crisis pathways

Objectives

1. Familiarise yourself with your local pathway for treating acute sickle cell painful crises
2. Reflect on your experience of treating patients with acute painful crises
3. Design a QIP to improve your local pathway, or an audit to examine its effectiveness

Group session

Break into groups of 2-3. Find your local guideline for treating children presenting with acute sickle cell painful crises, including the guidelines for A+E and the ward (if you have separate ones). Review these in your groups, alongside the resources below, and discuss the following questions.

Questions

1. What in these guidelines works well?
2. What parts of the guideline are difficult to implement in practice?
3. Describe to each other the experiences you've had of treating patients in A+E and/or on the wards with acute painful crises. What went well? What could be improved?
4. How might you assess the performance of your current guideline or local practice?
5. What interventions might you make to improve your local guideline or local practice?

Resources

1. NICE guideline for management of acute painful sickle cell episode in hospital: https://www.nice.org.uk/guidance/cg143/evidence/full-guideline-pdf-186634333
2. Sickle Cell Society recommendations for clinical care (p72)https://www.sicklecellsociety.org/wp-content/uploads/2019/06/SCD-in-Childhood_draft-for-review.pdf

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