South Thames Sickle Cell & Thalassaemia Network

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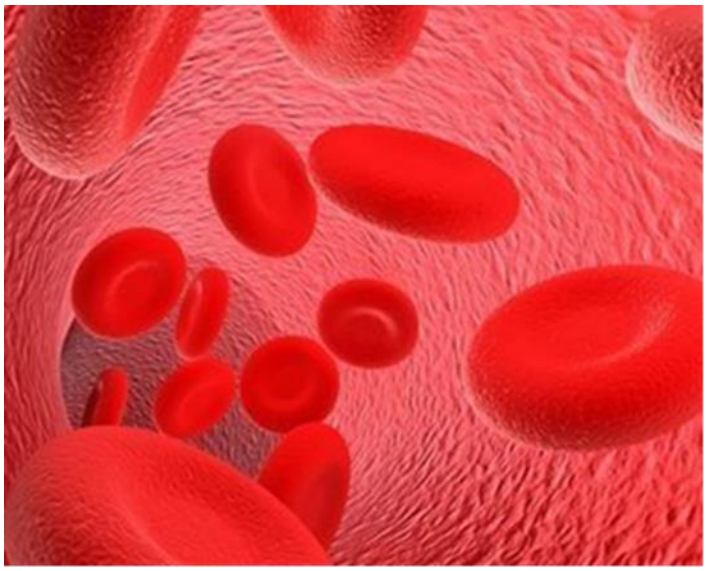
Paediatric and Adolescent Haemoglobinopathy Workbook



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AKNOWLEDGEMENTS
This Paediatric Haemoglobinopathy Workbook has been produced collaboratively with input from both Paediatric and Adult Haemoglobinopathy teams. It is intended for the training of nurses and allied healthcare professionals involved in the care of Haemoglobinopathy patients, and is based on published documents including the National Standards of Care and the Royal College of nursing (RCN) competencies 'Caring for people with Sickle Cell Disease and Thalassaemia syndromes', as well as relevant published guidelines and research.

We are grateful to all members of the Kings College Hospital Haemoglobinopathy teams, South Thames Sickle and Thalassaemia Network (STSTN) and members of the multi-disciplinary teams and the wider specialist teams including nephrologists, orthopaedic surgeons, cardiologists, hepatologists and obstetricians with whom we collaborate.

We also thank our training day sponsors ROALD DAHL and NOVARTIS.

We thank attendees for raising awareness, improving knowledge and skills and being part of working towards offering haemogloboinopathy patients the optimal care.

INTRODUCTION TO WORKBOOK
The Haemoglobinopathy Workbook is intended to illustrate key clinical issues and management strategies in the care of haemoglobinopathy patients. It is hoped that this will enable the widespread use of effective preventative and therapeutic interventions for adult haemoglobinopathy patients. Completion of the workbook and attendance at the study day is required for the successful completion of this haemoglobinopathy training event. You can contact your area Practice Development Educator to notify them of your successful completion so that they can update your training record

AIMS and OBJECTIVES
This workbook is designed for registered nurses, junior doctors and allied health professionals (AHP) who have some basic experience in caring for haemoglobinopathy patients (including Sickle Cell disease and Thalassaemia). We aim to promote high consistent standards of nursing and medical care. This workbook should be studied in combination with attendance at the training day.

The training day aims to update and build on your specialist knowledge of haemoglobinopathies. There will be an opportunity to discuss specific issues and queries with specialist haemoglobinopathy consultants, nursing staff and psychologists.

ACCREDITATION

This programme has been accredited by the RCN Centre for Professional Accreditation until September 4th 2018

Accreditation applies only to the educational content of the programme and does not apply to any product

RESOURCES, REFERENCES AND FURTHER READING

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4. Piel FB, Steinberg MH, Rees DC. Sickle Cell Disease. New England Journal of Medicine 2017 April 20;375 (16):1561-1573.

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10. Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK 3rd Edition, 2016. UK Thalassaemia Society.

11. STSTN South Thames Sickle and Thalassaemia network. Guidelines, policies, and leaflets created by Network. website http://www.ststn.co.uk/

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13. Thalassaemia International Federation (TIF) Guidelines http://www.thalassaemia.org.cy

14. Transcranial Doppler Scanning for Children with Sickle Cell Disease - Standards and Guidance, second edition. September 2016.

15. Guidelines on red cell transfusion in sickle cell disease. Part I: principles and laboratory aspects. Bernard A. Davis, Shubha Allard, Kate Ryan et al on behalf of the British Committee for Standards in Haematology. 7 November 2016.

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17. Vichinsky E. Consensus document for transfusion-related iron overload. Semin Hematol 2001;38:2-4

    http://www.b-s-h.org.uk/media/2640/transfusion_scd_01178_part_ii-1.pdf.

18. Learning Outcomes and Competencies in conjunction with the RCN Competencies, NHS Screening Programmes for Sickle Cell and Thalassaemia. Streetly, Tangayi and Anionwu. 2011

Sickle Cell Patients who present to Paediatric Accident and Emergency Department

1. Where can you find all Sickle cell policies?

2. Where are individualised Adolescent Sickle care plans found?

3. What observations should be carried out on arrival?

4. What blood tests should be taken on presentation?

5. What is the time frame that a Sickle Cell Patient should receive pain relief after presenting to A & E, and what history should be taken before giving the pain relief?

6. What condition should be considered with a sickle cell patient who presents with oxygen saturations of less than 94%, ⇧ Work of Breathing , Shortness of Breath, temp and history of Coryzal Symptoms**?**

7. What condition should be considered when a Sickle Cell Patient presents with a distended abdomen, pallor, lethargy Shortness of Breath, ⇧ Work of Breathing and lethargy?

8. What is a vaso-occlusive episode?

9. List some of the types of vaso-occlusive episodes?

10.What other complications may occur in children/ adolescents with sickle cell disease?

Admission of Paediatric and Adolescent Patients with Sickle Cell to the Ward

1. On admission of a Sickle Cell paediatric patient to the ward how often vital signs should be taken and what are the observations?

2. Which of the following would prompt you to escalate a deteriorating patient to the Sickle Cell Team?
a) Decreased level of consciousness

b) A drop in oxygen saturations of more than 2% from the patient's baseline

c) A drop in oxygen saturations of more than 4% from the patient's baseline +/- saturation of less than 94%

d) If triggering PEWS (Paediatric Early Warning Score)

3. When monitoring oxygen Saturations this should always be recorded and monitored in……..?

4. What are the key aspects of treatment for a patient admitted with painful crisis?

5. How often should pain score and observation be carried out for a patient admitted with a painful vaso-occlusive episode?

6. What are the key aspects of treatment for a patient admitted with headache untouched by pain relief, blurry vision, slurred speech and unilateral weakness?

7. What should you do if a patient appears over narcosed?

8. When can the monitoring of the patients vital signs be extended?

9. What is girdle syndrome and how is it treated? Most likely to be seen in older children

Treatment
1.
2.
3.
4.
5.
6.
7.
8.
9.

Management of Sickle Complications Acute Chest Syndrome (ACS) in Paediatric and Adolescent Sickle Cell Patients

1. What methods can you administer to try and prevent a patient admitted with a painful vaso-occlusive episode (painful crisis) from developing ACS?

a. Adequate pain relief
b. Incentive Spirometry
c. Continuous monitoring of vital signs
d. Antibiotics
e. Respiratory physiotherapy
f. All of the above

2. What methods of treatment can you provide for a patient who has confirmed ACS?

a. Review by Sickle Cell Team/Senior Nurses
b. Chest X-ray
c. Continuous monitoring of vital signs
d. Top up transfusion if steady state Hb has dropped by 20g/l or more
e. Exchange Transfusion (Apheresis) with direction from Sickle Cell consultant and registrar
f. Antibiotics
g. All of the above

Management of Sickle Complications Sequestration in Paediatric and Adolescent Sickle Cell Patients

1. What is sequestration and what are the symptoms?

2. What are the organs affected by sequestration?

3. What treatment can you administer for sequestration?

4. Some major indications that a Sickle Cell patient may require a splenectomy are?

5. Name two complications that can occur post splenectomy?

Management of Sickle Complications Management of Sickle Complications Cerebrovascular accidents /Stroke in Paediatric and Adolescent Sickle Cell Patients

1. Name types of stroke or cerebrovascular accident (CVA) that a Sickle cell patient can present with?

2. What are the signs of an overt stroke?

3. What treatment should be administered for a patient with suspected stroke immediately?

4. What treatment should be provided for Sickle Cell Disease patient post Stroke?

5. What are some of the stroke prevention methods that a comprehensive centre must provide for sickle cell patients?

6. What is the age range that children with Sickle Cell Disease should have Transcranial Doppler Scans?

Management of Sickle Complications Renal function in Paediatric and Adolescent Sickle Cell Patients

1.List some renal complications that a sickle patient may experience?

2. How would you monitor renal function in a sickle patient?

Management of Sickle Complications Priapism in Paediatric and Adolescent Sickle Cell Patients

1. What is priapism?

2. How should priapism be treated?

Specific treatment in hospital setting:

3. What is the end result for a patient who has suffered with untreated priapism?

4. What advice would you provide to a patient experiencing stuttering priapism (short-lived painful erections of up to 30 minutes)?

Management of Sickle Complications Aplastic Crisis in Paediatric and Adolescent Sickle Cell Patients

1.Define briefly what an Aplastic crisis is ?

2. How will someone with aplastic crisis present?

3. What treatment can you administer for a patient presenting with Aplastic crisis?

Management of Sickle Complications Pain Management in Paediatric and Adolescent Sickle Cell Patients

1. How would you monitor paediatric patients pain scores Babies, Toddlers and Adolescents?

2. What type of pain relief can a paediatric patient receive when admitted with a painful crisis?

3. How often should pain score be carried out for patients admitted with painful crisis?

4. Where can you find the current guidelines for patients on a PCA /NCA?

5. What additional medications should be written on the 'PRN' side of the drug chart for Sickle Cell patients receiving PCA/NCA/Opioids?

6. What opiate **SHOULD NOT** be given to patients who present with Sickle Cell Disease? And for what reason?

7. What non-pharmacological treatments could be offered to an individual during a sickle cell crisis?

Management of Sickle Complications Blood Transfusion Management in Paediatric and Adolescent Sickle Cell Patients

1. Name some roles of blood in the human body?

2. Name the main blood groups?

3. What blood tests should patients undergo when they are on a regular top up transfusion programme?

4. What actions should you take if you suspect a delayed transfusion reaction could occur 10 to 14 days after transfusion.

5. Give two reasons why a Sickle Cell patient requires regular transfusions/exchange?

6.What blood tests should patients undergo pre-red-cell exchange?

7.What blood tests should patients undergo post red-cell exchange?

8. Where can the proforma for manual Sickle Cell exchange be found?

9. What are the national requirements for filtering blood for a Sickle cell patient?

10. How many days before a top/up or exchange can a sickle patient undergo a blood test?

11. What information is needed prior to an automated exchange?

a)Height
b)Weight
c)Pre- transfusion Haemoglobin level
d)Pre- transfusion Sickle percentage
e)Pre- transfusion Haematocrit level
f) Peripheral access/double lumen port
g) All of the above

12. What are the current national and trust requirements regarding monitoring of vital signs when receiving blood products?

13. What are the trust guidelines for checking patient blood unit before administration?

14. If a patient has a transfusion reaction when the unit of blood is started what would you do?

15. What are some of the complications of transfusions a SCD patient can experience?

16. What Urine test should be collected when patients are being regularly transfused?

17. What must you look for on a unit of blood for a sickle cell patient?

Blood provided for SCD patients should be:

HbS negative and, where possible, should be <7 days old for exchange transfusion (<10 days old for simple transfusion), but older blood may be given if the presence of red cell antibodies makes the provision of blood difficult

Blood should also be ABO-compatible, extended Rh- and Kell-matched units. If there are clinically significant red cell antibodies (current or historical) then the red cells selected should be negative for the corresponding antigens.

Transfusion to HbS <30% will prevent or reverse most acute sickle complications and significantly reduce long-term complications in chronically transfused patients. Baseline Hb and % HbS should be taken into consideration in setting target post-transfusion Hb in order to avoid **hyperviscosity**. In SCD patients with baseline Hb <90 g/l and not on regular transfusions, the **post-transfusion Hb should not exceed 100 g/l, particularly if %HbS is greater than 30%**. The post-transfusion Hb can be set at a higher target in chronically transfused patients or if %HbS is low, but should be individualised to each patient. Patients with high baseline Hb (>100 g/l) should not be transfused above their steady state Hb

18. When a patient on regular transfusions gets to a serum ferritin level of 1000 what medication should be started?

Blood transfusion in Sickle Cell Disease:
Consideration of sickle cell patients for transfusion, particularly long-term regimens, should weigh up the potential benefits against potential risks.

Cerebrovascular disease
Regular transfusion to maintain HbS <30% should be offered as initial treatment to children with SS or S/β^o thalassaemia aged 2–16 years judged to be at high risk for a first stroke on the basis of Transcranial Doppler ultrasonography (TCD).

Hydroxycarbamide treatment should be considered for the primary prevention of stroke in children with sickle cell anaemia and high TCD velocities but not severe Magnetic Resonance Angiography (MRA)-defined cerebral vasculopathy after an initial period of transfusions. The duration of the initial period of transfusion should be tailored to the individual patient but should be for a minimum of 1year; the transition to hydroxycarbamide should be done gradually and transfusion should be withdrawn after the hydroxycarbamide has been escalated to the maximum tolerated dose.

Regular transfusion to maintain HbS <30% effectively reduces the incidence of recurrence of cerebral infarction (defined as a stroke or a new or enlarged silent cerebral infarct) in children with sickle cell anaemia and S/β^o thalassaemia aged 5–15years. Treatment options including transfusion should be discussed with families of children who are found to have silent cerebral infarcts. Transfusion should be offered to children who are identified to be at greatest risk for recurrence of infarction after discussion of its benefits and risks.

Long-term transfusion to maintain HbS <30% is recommended for the prevention of recurrent ischaemic stroke due to sickle cell disease in both children and adults.
Adults or children who present with signs or symptoms suggestive of acute ischaemic stroke should be transfused without any delay to maintain HbS <30% pending further investigation. Those with confirmed stroke due to sickle cell disease should continue regular transfusions long-term.

Surgery
Preoperative transfusion is recommended for sickle cell disease (HbSS and HbSC) patients undergoing medium-risk surgery (e.g. abdominal, tonsillectomy, orthopaedic).
Transfusion is recommended for sickle cell patients of all genotypes requiring high-risk surgery (e.g. cardiovascular, brain).
All sickle cell patients with other genotypes undergoing surgery should be individually assessed, taking into account previous history and complexity of surgery, and a management plan should be formulated to include the need for transfusion.
Particular care should be taken to ensure that all aspects of perioperative care, including oxygenation, hydration, warmth and anaesthetic and surgical technique, are optimised in all sickle cell patients undergoing surgery.
For patients requiring emergency surgery, the urgency and complexity of the procedure should be taken into account in the timing of perioperative transfusion. Simple transfusion should be given preoperatively if Hb <90g/l provided this will not result in undue delay to surgery. If transfusion is likely to cause an unacceptable delay to surgery, it is reasonable to proceed to surgery while arranging to transfuse the patient intra- or post-operatively if necessary.

Acutely ill patients
Transfusion is recommended and may be life-saving in acute sickle complications such as splenic sequestration, hepatic sequestration, aplastic crisis and severe acute chest syndrome.
Transfusion should be considered in the unwell patient with acute multi-organ failure, mesenteric syndrome and patients with severe sepsis. Such cases should be discussed with the specialist haemoglobinopathy team (SHT).
Transfusion for other causes of acute anaemia requires individual assessment and should be discussed with the SHT. Transfusion may be given by simple transfusion (top up) or exchange depending on clinical severity under the guidance of the SHT.

Pregnancy
Prophylactic transfusion is not routinely required for sickle pregnancy, but should be considered for women with:

- previous or current medical, obstetric or fetal problems related to SCD
- women previously on hydroxycarbamide because of severe disease
- multiple pregnancy.

Women on long-term transfusions for stroke prevention or for amelioration of severe sickle complications should continue with regular transfusions throughout pregnancy.
Transfusion should be considered in women with worsening anaemia or those with acute SCD complications (acute chest syndrome, stroke etc.).

Amelioration of severe disease
In selected patients with severe disease, blood transfusion can be effective in ameliorating disease, resulting in reduction in hospital bed days. Hydroxycarbamide is recommended as first line treatment for prevention of recurrent acute chest syndrome or repeated painful episodes associated with chest syndrome. Regular transfusion should be considered for patients failing this treatment or for whom hydroxycarbamide is contraindicated or not acceptable.

Other indications
Transfusion is not recommended to treat steady state anaemia provided that Hb has not fallen over a period of time to symptomatic levels (e.g. with developing chronic kidney disease).
There is no evidence that transfusion shortens the duration of a painful crisis.

Transfusion is not recommended in uncomplicated painful crises but should be considered if there is a substantial drop in Hb from baseline (e.g. >20 g/l or to Hb <50 g/l), haemodynamic compromise or concern about impending critical organ complications.
The benefit of transfusion to relieve established acute priapism has not been shown in randomised controlled trials. Many patients require a shunt or drainage procedure under general anaesthesia, which may require a transfusion. Such cases should be discussed with the SHT.
Transfusion has been shown to reduce the incidence of symptomatic avascular necrosis in children receiving regular transfusions to maintain HbS <30% for prevention of recurrence of cerebral infarction. However, there is no consensus on the use of transfusion for the sole purpose of preventing this complication in routine practice.
Where transfusion is considered for indications where there is insufficient evidence for its benefit (e.g. leg ulcers, pulmonary hypertension, end stage renal or liver disease, progressive sickle cell retinopathy), a full risk-benefit assessment should be carried out in liaison with the SHT and each case should be considered on its own merits.

Investigation of a haemolytic transfusion reaction:

1) Document evidence of haemolysis.
Check haemoglobin concentration and review blood film

Check bilirubin, lactate dehydrogenase and reticulocyte count

Check urine for haemoglobinuria and if positive and hyperhaemolysis suspected, consider serial high performance liquid chromatography analysis of the urine

(2) Serological testing on pre-transfusion and post-transfusion blood samples.
Repeat ABO/Rh D typing

Check antibody screen on both samples

Red cell units transfused within 12–24 h should be crossmatched against both the pre- and post-reaction samples

Check the direct antiglobulin test (DAT). A positive DAT may be encountered as part of an investigation

(3) If antibody screen positive
Determine the specificity of the antibody (ies) – (antibody investigations may demonstrate a new alloantibody or antibodies in a patient with a delayed haemolytic transfusion reaction)

If the specificity of the red cell antibody is not clearly determined the sample should be sent to a red cell reference laboratory

(4) If DAT positive
Prepare an eluate to test for the presence of specific alloantibodies

Even if no new red cell alloantibody is detected in post-transfusion sample as above, but DAT is positive, red cell eluate studies should be undertaken

(5) Selection of red cell units for further transfusion
Carefully consider the need for further transfusion with consultant input and discussion with the National Blood Service if complex transfusion requirements

Select ABO extended Rh and K matched units negative for the relevant antigen(s) to which there are current or historical antibodies

Undertake serological crossmatch to check compatibility; **electronic issue should not be used**
If the identity of the new alloantibody is in doubt despite further specialist testing, consider providing extended antigen matched blood (if serological phenotyping cannot be used because of the presence of transfused donor red blood cells, the sample should be sent to an appropriate reference laboratory for molecular red cell genotyping)

Any adverse events or reactions related to transfusion should be appropriately investigated and reported to local risk management systems and to National Haemovigilance Schemes (e.g. Serious Hazards Of Transfusion – SHOT).

Management of Sickle Complications Central Venous Access in Paediatric and Adolescent Sickle Cell Patients

1. What are the types of ports that a sickle patient may have insitu?

2. Who can access ports?

3. How should ports be accessed?

4. What solution should a double lumen vortex port be locked with?

5. How often does the port-a-cath need to be flushed and locked?

6. What are other forms of access for a patient who is in need of top up transfusion or exchange?

Management of Sickle Complications Long Term Management of Paediatric and Adolescent Sickle Cell Patients

1. Which of the following may alleviate long term complications of sickle cell?

a) Hydroxycarbamide
b) Red cell exchange transfusion
c) Bone marrow transplantation
d) All of the above

2. In accordance to the National Standards for Haemoglobinopathies how soon after birth should a patient be seen by a comprehensive centre/ haemoglobinopathy Consultant?

3. What are the precautionary methods used to assist with stroke prevention for children between ages 2-16 years of age?

4. What is the lifelong prophylaxis treatment that a sickle cell patient should be taking from the age of 6weeks old?

National guidelines recommend the following:

- All routine vaccines, including live vaccines such as measles, mumps and rubella (MMR) can be given safely to children or adults with an absent or dysfunctional spleen.
- Asplenia or hyposplenism is not a contra-indication for live vaccinations prior to travel (eg, yellow fever and live oral typhoid vaccine).
- Re-immunisation with Pneumovax of hyposplenic/asplenic patients is currently recommended every five years. Re-immunisation in these patients may be made on the basis of antibody levels.
- Influenza vaccination - annual influenza vaccination is recommended after 6 months of age.

Lifelong prophylactic antibiotics

These are recommended in patients at high risk of pneumococcal infections and the antibiotics of choice are oral phenoxymethylpenicillin or macrolides. Patients developing infection, despite measures, must be given systemic antibiotics and admitted urgently to hospital.

- Risk factors for high risk in hyposplenism include:
    - Age <16 years or >50 years.
    - Poor response to pneumococcal vaccination.
    - Previous invasive pneumococcal illness.

- Use phenoxymethylpenicillin (adult 250-500 mg bd - although 500 mg od may be more realistic if compliance is a particular problem), amoxicillin (adult 250-500 mg daily), erythromycin (adult 250-500 mg daily) orally.
- Consider recommending that the patient take a full therapeutic dose of antibiotics if they develop infective symptoms such as pyrexia, malaise, shivering, etc and that they seek medical advice immediately.
- Allowing patients to have a reserve supply of antibiotics at home or on holiday may also seem appropriate.
- If not deemed to be high-risk then the pros and cons of taking lifelong antibiotic prophylaxis need to be discussed with each individual patient.

The British Committee for Standards in Haematology recommends the following:

- Patients should be given written information and carry a card to alert health professionals to the risk of overwhelming infection. Patients should wear an alert bracelet or pendant.
- Patients should be aware of the potential risks of overseas travel, particularly with regard to malaria and unusual infections - e.g., those resulting from animal bites.
- Patient records should be clearly labelled to indicate the underlying risk of infection. Vaccination and re-vaccination status should be clearly and adequately documented.

Management of Sickle Complications Peri-operative Care Paediatric and Adolescent Sickle Cell Patients

1. Before any planned surgical procedure an individual with sickle cell should have the following pre-requisites:

2. What is the current national standard guideline and trust recommendation on antibiotics for individuals with sickle cell and those without a functioning spleen?

3. Where can you find the haemoglobinopathy antibiotic guideline?

Pre-operative outpatient review with a haematology consultant with a specialist interest in haemoglobinopathies for peri-operative planning will include documented decisions about:

- Red cell transfusion (Exchange transfusion or top up to Hb of 10g/l) with blood fully matched for ABO, full-Rhesus phenotype (Cc/D/Ee), and K1 antigen, plus any other red cell allo-antigens present.
- Pain relief
- Hydration/fluid balance
- Oxygenation (including hyperoxygenation at induction of anaesthesia and careful monitoring of peri-operative oxygenation by pulse oximetry), incentive spirometry, chest physiotherapy, and consideration of prophylactic CPAP for 24 hours particularly post major abdominal and thoracic surgery
- Prophylactic antibiotics and infection screening and early management of any post-operative infections (hyposplenism)
- Thromboprophylaxis
- Temperature regulation (including consideration of warm air blankets, warmed IV fluids and adjustment of temperature in theatre)
- Avoidance of tourniquets and red cell salvage (neither suitable in sickle patients)
- Discharge planning and follow up.

Management of Thalassaemia Complications in Paediatric and Adolescent Thalassaemia

Thalassaemia

1. Name some types of thalassaemia

2. The major clinical consequences of the pathophysiology of thalassaemia are severe, life-threatening anaemia and severe bone marrow hyperactivity. These result in

3. List some complications of β-Thalassaemia Major and intermedia

4. How can you identify a person's pre-transfusion trough haemoglobin level?

5. Name at least three co-morbidities or conditions associated with Thalassaemia.

6. Individuals with thalassaemia should have what blood test prior to every transfusion?

7. How many days in advance of top up transfusion can a patient with Thalassaemia have pre-blood tests?

8. Thalassaemia patients who have special blood requirement where will these details be found where?

9. Urine should be tested monthly for:

10. Thalassaemia patients receiving blood transfusions should have what tests yearly?

Management of Transfusion related Iron Overload in Sickle Cell Patients

1. Untreated iron overload can lead to:

2. Side effects of Iron Chelation

Deferoxamine (Desferal)

Deferasirox (EXJADE)

Deferiprone

3. What is the serum ferritin Level that you start treating with chelators?

a) 800

b) 1000

c) 1200

d) 1020

4. What monitoring tests should be performed yearly to monitor iron overload for patients having regular blood transfusions?

All Practice Development Nurses Can email Eleanor Baggley: 0203 299 5102
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For a copy of the answer book.

Thank you

Please note: The RCN **cannot confirm competence of any practitioner**.