

This section is designed to test your knowledge of selected topics in this issue of the journal. The correct answers are given at the foot of the page.

Self-assessment

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SINGLE BEST ANSWER

1 A 6-year-old Afro-Caribbean boy presents to accident and emergency with severe abdominal pain. The boy looks dehydrated and is cold. The parents give a history of a congenital disorder of haemoglobin structure that affects them too. The boy also complains of pain in long bones and on examination there is an enlarged spleen. His haemoglobin is 70 mg/l. Which of the following is true regarding the diagnosis and management of this condition?

- A. Haemoglobin analysis, by electrophoresis may confirm the diagnosis
- B. There is no proven pharmacological therapy available for this condition
- C. Immediate blood transfusion may prove detrimental
- D. This disease is inherited as an autosomal dominant condition
- E. There is no role for prophylactic antibiotics

2 A 70-year-old lady presents in accident and emergency with major bleeding. History reveals that she is on apixaban for a previous stroke. All emergency blood tests including clotting are ordered. Her renal function seems significantly impaired. Local policy for haemorrhage management has been initiated. Which of the following are true regarding the management of this patient?

- A. Prothrombin complex may be used as a direct antidote
- B. Bleeding can be reversed with a monoclonal antibody that binds and clears apixaban
- C. Intravenous vitamin K should be immediately administered
- D. Treatment may be guided by serum apixaban levels
- E. The patient can have significant anticoagulant on board and may have normal clotting screens

MULTIPLE CHOICE QUESTIONS

3 Anaesthesia for patients with sickle cell and other haemoglobinopathies

Which of the following are true regarding the clinical presentation of thalassaemia syndrome?

- A. Haemoglobin structure is normal
- B. The anaemia is described as hypochromic and macrocytic

- C. Inheritance follows an autosomal recessive pattern
- D. Symptoms typically can be noticed in the neonatal period
- E. Bone expansion secondary to extramedullary erythropoiesis may lead to a difficult airway

4 Risks of perioperative blood transfusions

Which of the following are true regarding the presentation and management of transfusion-associated circulatory overload (TACO)?

- A. TACO is defined as acute or worsening pulmonary oedema within 6 hours of transfusion
- B. This is a rare cause of transfusion-related major morbidity and mortality reported to SHOT in 2017
- C. TACO is much more common in patients who are morbidly obese
- D. Weight adjusted dosing should be used to guide the volume of transfusion administered
- E. Previous history of fluid overload secondary to renal disease is a risk factor for TACO.

5 Jehovah's Witnesses

Which of the following are true regarding the management of a Jehovah's Witness patient?

- A. A competent adult cannot change their mind once a decision is made prior to surgery
- B. In an emergency situation, where a child's life is at risk, it is acceptable to withhold life-saving treatment if it is against parental wishes
- C. Autologous predonation is acceptable and should be recommended
- D. Perfluorocarbons are licensed for use in the UK
- E. Hyperbaric oxygen therapy has been recommended in select cases postoperatively to optimise oxygenation

6 Physiology of haemostasis

Platelet structure and function

- A. The average lifespan is around 120 days
- B. α -granules within the platelets contain adenosine diphosphate (ADP)
- C. The platelet membrane contains numerous glycoproteins which serve as binding sites for various other molecules such as fibrinogen
- D. von Willebrand factor is secreted from within the platelet membrane
- E. Platelet aggregation is achieved by platelets cross linking at GPIIb/IIIa receptors on the platelet membrane

ANSWERS

1. Correct answer: A. Sickle cell disease (SCD) refers to a group of congenital disorders that lead to the formation of structurally abnormal haemoglobin molecules. SCD results from a mutation on chromosome 11, which causes an amino acid substitution – valine for glutamic acid – on the β -globin subunit of HbA leading to the production of haemoglobin S (HbS). SCD is an inherited autosomal recessive condition. Individuals who inherit this abnormal gene from both parents (homozygous HbSS) have a severe form of SCD, often referred to as sickle cell anaemia in which the majority of haemoglobin is HbS.

The vast majority of newborn babies with SCD are born in Africa, the Mediterranean, Middle East and parts of India; however, the number of patients with SCD is thought to be increasing in all countries. This results in vascular occlusion and haemolytic anaemia, two characteristic features of SCD.

Traditionally, it was thought that a trigger, such as hypoxia, led to sickling, precipitating further flow impairment, stasis and acidosis leading to a 'vicious cycle' of further sickling ending in ischaemia, infarction and end organ damage. Stress, hypothermia, alcohol abuse and dehydration are some other states, which favour stasis and promote sickling.

The Sickledex test may be used as a rapid screening tool to identify the presence of HbS; however, it is unable to distinguish between sickle cell trait and SCD. Haemoglobin analysis, by electrophoresis, remains necessary and is the gold standard to confirm the diagnosis.

Palpation for splenic size to encourage early presentation of splenic sequestration crisis has been suggested to be of benefit. Oral penicillin prophylaxis is started at the time of diagnosis. There is an increased susceptibility to infections by encapsulated bacteria and as such, fever in a patient with SCD should be urgently assessed by a medical professional, cultures taken and appropriate antibiotics started.

Hydroxycarbamide (hydroxyurea - HU) is currently the only approved pharmacological treatment for SCD. It is given as a once daily dose (15–35 mg/kg/day) and evidence supports its use to reduce the risk of acute pain and the acute chest syndrome as well as being effective in primary stroke prevention.

The aim of blood transfusion is to decrease the concentration of HbS to <30%. Blood transfusion can be used for both therapy and prophylaxis. Transfusion is readily used during sequestration crises, in multi-organ failure, and in severe sepsis. During acute painful crisis, anaemia should not be routinely treated with blood transfusion unless there is an Hb drop greater than 2 g/dl or deteriorating organ function. As a simple rule, transfusion can be given if Hb is <9 g/dl.

2. Correct answer: E. Direct oral anticoagulants (DOACs) that are currently used in clinical practice include anti-Xa drugs such as: Apixaban, rivaroxaban and edoxaban; and the direct anti-thrombin agent dabigatran. In patients experiencing major bleeding or life-threatening bleeding whilst taking a DOAC the following management is advised. Stop the DOAC. Manage the bleeding according to your local major haemorrhage policy, including the use of tranexamic acid. Take a full coagulation screen including PT, aPTT, fibrinogen, and thrombin time. A drug specific test can also be taken (i.e. a 'rivaroxaban level!'); not all hospitals are able to offer this service. Remember that a patient can have significant anticoagulant on board and have normal clotting screens, especially apixaban. Standard ROTEM tests (EXTEM/INTEM CT) can detect DOACs (dabigatran, edoxaban, rivaroxaban) at therapeutic levels, but appear insensitive to apixaban.

Treatment of a patient with significant bleeding who is taking an anti-Xa DOAC is more complex due to the lack of a direct antidote. Currently, practice in the UK includes the use of PCC (50 units/kg) which is given principally to improve thrombin generating capacity. There is no role for intravenous vitamin K in patients with bleeding secondary to DOACs.

3. Correct answers: A, C, E

4. Correct answers: A, D, E

5. Correct answer: E

6. Correct answers: C, E