Don't Forget The Bubbles

SICKLE CELL DISEASE

Learners Guide

Author **Beatrice Zanetti**
(Edits by the DFTB Team)
fellows@dontforgetthebubbles.com

PRE-READING FOR LEARNERS

To prepare for this session, learners could read:

https://www.stemlynsblog.org/nice-faces-the-sickle/

https://first10em.com/acute-chest-syndrome-sickle-cell-disease/

https://pedemmorsels.com/sickle-cell-disease-fever/

https://emergencymedicinecases.com/video/sickle-cell-disease/

For more information, this is a link
http://sickleoptions.org/en_US/video-library/
It has many useful videos for patients and doctors about sickle cell disease, management and communication scenarios.

CASE 1 (20 MINUTES): SALMONELLA OSTEOMYELITIS

15-month-old Kenyan boy presented to AE with right hand swelling.
He had a temperature of 38.5 °C and he was crying inconsolably.
Mother said that hand swelling episodes might have happened before when he was living with grandparents in Kenya. The patient was born in Mombasa; his mother moved to UK 6 months ago while he remained with his grandparents until recently, when he arrived in the UK.
On clinical examination, he had marked swelling over the proximal right ring finger plus diffuse swelling and erythema of the dorsum of the hand.

Investigations:
Bloods showed Hb of 8 g/L, White cell count 13x109/L, Platelets 570 x109/L, CRP 35mg/L.
Blood film showed sickle cells and further sample was sent for confirmation of diagnosis.
Blood culture is pending.
XR imaging showed a patchy area of lucency with periostitis and soft tissue swelling of metacarpals or metatarsals region.

Patient was started on intravenous antibiotics (ceftriaxone 50mg/kg OD) and was taken to theatre for exploration. In theatre, the patient was noted to have purulent fluid within the subperiosteal space of the right 4th proximal phalanx. Fluid culture yielded growth of Non-typhoidal Salmonella enterica.

1. Why is important to know whether this child has sickle cell disease?
What key elements from history taking would support the diagnosis of sickle cells?

2. How is sickle cell disease diagnosed?

3. What could be the differential diagnosis?

4. Why are sickle cell patients more susceptible to severe infections?

5. What strategies can be implemented to prevent infections in the asplenic patient?

CASE 2 (20 MINUTES) ACUTE PAINFUL CRISIS AND PRIAPISM

10-year-old boy with known SCA. He presents to ED due to severe pain in the legs. At home, he had 24 hours of alternating paracetamol and ibuprofen but pain was so unbearable that parents presented to hospital in the middle of the night. Patient does not have fever. Parents are also worried since child has had 3 hours of priapism but he is very shy about it.

1. How would you evaluate this child's pain?
2. What medications would you give to control pain at home?
3. What medications would you give to control pain in hospital?
4. How would you treat priapism?

ADVANCED CASE 1 (30 MINUTES) ACUTE CHEST SYNDROME

A 6-year-old girl from Saudi Arabia was referred by her General Practitioner to the local Emergency Department. She complained of cough and runny nose for 3 days. Past medical history was positive for sickle cell anaemia. She had had several episodes of acute pain crisis in the past. She attends sickle cell clinics regularly. She is on folic acid 5mg OD PO, penicillin V 250mg BD PO. For pain crisis, she has PRN paracetamol, ibuprofen and dihydrocodeine. She also suffers from asthma occasionally and she is on salbutamol inhalers as required.

Vitals: Temperature 39.5, HR 100 beats per minute, Respiratory Rate 35, saturations 90% at room air. Blood pressure 95/60 mg/Hg.

On examination, she has pallor, no jaundice. On auscultation, there is bilateral wheeze. On abdominal palpation, spleen is enlarged.

You order some investigations including a Chest CXR

1.What features can you see in the chest XR that will help you to diagnose the patient?

2.What would be the appropriate acute management?

3.Would you consider this patient for blood transfusion?

4.What is the role of hydroxycarbamide?

ADVANCED CASE 2 (30 MINUTES): STROKE IN SICKLE CELL DISEASE PATIENT

An 8-year-boy from Nigeria presented to ED with an acute right sided hemiplegia. He is known to have sickle cells and he is under the care of haematology department. He has been on holidays for 2 months in Nigeria and has not been compliant with his usual treatment. On examination, he looks pale and you can fell the spleen on abdominal palpation.

1. How common is cerebro-vascular disease in sickle cell patients?
2. What would be the acute management of this child?
3. What is the role of transcranial dopplers in primary prevention of strokes?
4. What strategies are useful for secondary stroke prevention?
5. In the above case, what could be the cause of the pallor (anaemia)?

QUIZ QUESTIONS: (10 MINUTES)

Question 1.

Which of the following statements is FALSE concerning Sickle Cell Anaemia?

A: It is a recessive autosomal inherited blood disorder
B: The sickle shape of the cell is due to an abnormality in haemoglobin
C: Symptoms are never seen in children
D: Fatigue is a common symptom

Question 2.

Why is sickle cell disease more common near the equator (Africa, Caribean countries)?

A. Sickle cell trait is a genetic mutation that provides protection against malaria
B. Sickle cell disease is a genetic mutation that provides protection against malaria
C. Sickle cell disease is a genetic mutation that provides protection against trypanosomiasis
D. Sickle cell trait is a genetic mutation that provides protection against trypanosomiasis

Question 3.

What infections are sickle cell patients more at risk of?

A. Pneumococcal sepsis
B. Rotavirus gastroenteritis and dehydration
C. Recurrent UTIs
D. Mycoplasma pneumonia

Question 4.

Transfusions in sickle cell patients are indicated in patients with:

A. Any surgical procedures
B. Acute fulminant priapism unresponsive to treatment
C. Asymptomatic functional asplenia
D. Moderate acute chest syndrome without hypoxia

Question 5.

What are specific treatments for sickle cell disease?

A. Regular blood transfusions
B. Hydroxycarbamide
C. Bone marrow transplant
D. All of the above

Question 6.

In an outpatient sickle cell visit, what would you be a priority to assess?

A. Haemoglobin A1C level
B. Saturation of oxygen
C. Reflexes
D. Vaccination history

INFOGRAPHICS (2 minutes)

1. Think about sickle cell disease in patients who come from or whose ancestors come from Africa, Caribbean, Middle east and Mediterranean regions

2. Sickle cell patients are high risk for serious bacterial infections bcause they have functional asplenia

3. Never underestimate pain in Sickle cell patients: evaluate pain with an adequate pain scale and treat it aggressively

4. A person with sickle cell disease can have a good quality of life by getting regular check-ups and following treatments prescribed by doctors

5. Be aware of potential life-threatening SCD complications, such as chest crises, severe infections or cerebrovascular disease. If in doubt, consult with a local haematologist regarding sickle cell patient management

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Guidelines:

From http://www.sickkids.ca/pdfs/haematology-oncology/8217-sicklecell-guidelines2006.pdf

Acute Chest Syndrome or Pneumonia : Guidelines for Management in Children with Sickle Cell Disease Acute Chest Syndrome or Pneumonia : Guidelines for Management in Children with Sickle Cell Disease. 2018; 1–7.

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https://emergencymedicinecases.com/sickle-cell-disease-management-rapid-reviews-video/

https://sicklecellanemianews.com/ketamine/

https://www.rcpch.ac.uk/sites/default/files/2019-04/20160203%20Key%20Recommendations%2008.04.19.pdf

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