**CASE -Sickle Cell Anemia:**

AW, a 25-year-old Afro-Caribbean man, has been admitted to your ward with a sickle cell crisis. AW has a raised temperature and complains of severe pain in his limbs, chest and lower back. His regular medications are:

• Phenoxymethylpenicillin 500 mg twice a day

• Folic acid 5 mg daily.

AW's blood results are as follows:

WBC 13.1 x 109/L (4.0-11.0 x 109/L) RBC 3.5 x 109/L (3.8-4.8 x 109/L)

Hb 8.5 g/dL (12.0-15.0 g/dL) Hct 0.33 (0.36-0.46)

Bilirubin 45 micromol/L (3-17 micromol/L)

Blood film showed increased reticulocytes, sickle cells and presence of target cells.

**Questions:**

1. Which groups of people are at the highest risk of having sickle cell anaemia?
2. What is the cause of sickle cell anaemia?
3. What is a sickle cell crisis?
4. What situations may precipitate a sickle cell crisis? What do you think may have precipitated AW's crisis?
5. The doctor caring for AW asks your advice regarding analgesia. So far he has prescribed regular paracetamol and full-dose dihydrocodeine but AW is still in severe pain. What recommendations can you make regarding analgesia for AW?
6. Write your comment on AW's blood results.
7. What other acute management may be necessary for AW?
8. Why is AW taking phenoxymethylpenicillin and folic acid?
9. AW has frequent crises and is concerned because he is due to go on holiday shortly. What advice could you give AW to help reduce the risk of further crises?
10. What are the prognosis and long-term complications for patients with sickle cell anaemia?
11. Due to AW's frequent crises the medical team caring for him is considering initiating hydroxycarbamide (hydroxyurea). What evidence is there to support the use of hydroxycarbamide in the management of sickle cell anaemia?
12. How is hydroxycarbamide thought to work in the management of sickle cell anaemia?
13. The medical team ask what dose of hydroxycarbamide should be prescribed and what monitoring they should undertake. What do you advise?
14. How would you counsel AW regarding his new medication?
15. Hydroxycarbamide is not licensed for the management of sickle cell anaemia.

What implications does this have? What should you do?