SICKLE CELL ANEMIA

FREQUENTLY ASKED QUESTIONS

(FAQ)

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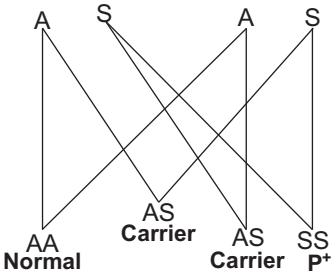
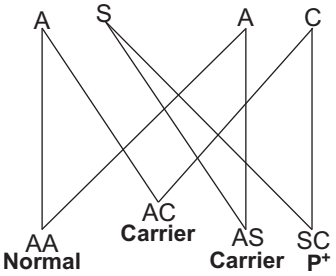
1. What is sickle cell anaemia? How many types do we have?

Sickle cell anemia is a state of chronic anaemia caused by a mutation in the gene sequence of the red blood cell. Glutamic acid is replaced by valine in the B6 position of the haemoglobin chain. This results in an abnormality which is inherited thus:





Carriers do not manifest the symptoms of the sickness because the presence of the dominant A gene suppresses the manifestation of the effects of the defective gene. Its only individuals with two defective genes SS or SC that manifest the symptoms of the illness.

2. Can Genotype AA mother and AA father give birth to an SS child?

No, its not possible because an SS child has to inherit defective gene from both father and mother. Since the parents do not carry the S gene they cannot transfer it to their offspring.

3. Can one get SS from blood transfusion?

No, SS is inherited from the DNA passed on from the parents. The blood cells transfused into a patient live out their life span and die without changing the inherited genetic code.

4. How early can SS be detected in a child?

SS can be detected in pregnancy or at delivery if there is a high index if suspicion which warrants the test. If these early tests are not done, then suspicion arises from about 8 months when the child develops hand and foot syndrome (Dactylitis). This usually occurs when fetal haemoglobin levels decrease after 6 months of age.

5. Why is SCA an African disease?

Its an African problem because the S gene protects against plasmodium falciparum infection which is prevalent in Africa in the sub-saharan region.

6. How an SCA be prevented?

SCA can be prevented if two people who carry the defective gene do not procreate.

7. Can Hbss marry Hbss?

Hbs can marry Hbs on the condition that they will not have

biological children so as not to produce more Hbss .

8. Does chronic leg ulcer ever heal?

It does with good management and adequate nutrition.

9. Can Hbss patient take soft drinks like Coca cola and Fanta?

Yes they can but in moderation because soft drinks are loaded with sugar. Too much sugar leads to diabetes which we don't want to add to the problems associated with SCA.

10. Why are Hbss patients always stubborn?

Any individual with chronic illness gets fed up once a while with all the do's and don'ts. Living with chronic pain is bound to affect one's personality one way or the other.

11. Is it true that all Hbss women die at child's birth?

No, this is not true, but they require specialist care during pregnancy and delivery and should not go for antenatal care in a primary health centre or maternity, rather look for a specialized centre to get the best outcome.

12. What should the Hbss woman eat and do during pregnancy please?

Hbss woman needs to eat nourishing foods with a good amount of fruits and vegetables incorporated into her diet. She should register for ante natal care as soon as she discovers that she's pregnant so she can have adequate supervision.

13. Hbss does not live beyond 40, true or false?

FALSE. With good care and good motivation by the patient, Hbss have lived up to 70 and above.

14. If hbss doesn't die by age 18 and 21, then they will live to age 40, True or False? *False*

15. What type of exercise Hbss do?

Physical exercises are not recommended due to increase oxygen demand, but board games would give recreation too.

16. What type of job/profession can Hbss do?

Any job that they have the flare/talent for which doesn't require too much physical activity.

17. Why do Hbss individuals have yellow eyes and big abdomen?

The yellow pigment which stains the white of the eyes is bilirubin, a bye product of red cell degeneration. The sickle

red cells in Hbss patients have a shorter life span than normal red cells. So they die early and a lot of bilirubin is spilled into the blood at a faster rate than the liver can clear up, it increases in the blood causing the yellow colour in the eyes and urine.

It is also increased when there's an infection which causes red cells to die rapidly and also when there's malaria infection because plasmodium parasites also destroy red cells.

Children with Hbss tend to have big abdomen when their liver and spleen are stressed and working harder to keep the body system in equilibrium. Liver and spleen get bigger when they're working harder.

18. Can Hbss patient take part in sporting activity?

It is not advisable because oxygen requirement is increased during sporting activity and Hbs is already in a state of chronic anemia.

19. What can an Hbss do if he has a painful erection at night when he can't come to hospital?

Apply ice pack, take a shower or take a walk.

20. What is AVN and what can one due to reduce the pain?

AVN – is Avascular Necrosis of the head of femur. It happens when there's blockage of the single artery that supplies the head of femur by sickle cell agglutination which prevents supply of blood and oxygen to the head of femur. This then dies and crumbles, resulting in shortening of the limb due to the compressed femoral head. Analgesics, traction to relieve the pressure due to impaction at the hip joint help the pain a little, but total hip replacement surgery is the ultimate solution to AVN.

21. Can hbss join in religious annual fasting and prayer?

No, it is not advisable because of the dehydration associated with fasting. Dehydration triggers crisis. Irregular eating is unadvisable for Hbss.

22. What type of blood tonic should Hbss use?

I do not recommend any blood tonic. Use of blood tonic could lead to iron overload (polychromasia). Would rather encourage Hbss to eat nourishing foods with fruits and vegetables everyday. The body will absorb according to its needs and excess will be excreted rather than taking blood tonics.

Extra iron and folic acid will be required for pregnant Hbss though.

23. Who is more prone to crisis male or female Hbss Pt?

Female Hbss patients lose blood every month during the menstrual cycle making them more susceptible to anaemia, but young boys generally forget the limitation in exercise tending to play with their mates when they're not ill, so its balanced.

24. When is it right to get married?

When the individual is mentally and physically matured enough to handle the stress of living with another adult who is not from your family of origin.

25. What is the average life expectancy of Hbss patients?

It is the 5th decade of life like for any other Nigerian.

26. Is it true that Hbss patients are brilliant?

Yes, because what they lack in physical energy they make up by using their brains.

27. Can bone marrow transplant cure SCA? *Yes, it can*

28. Is it possible for Hbss patient not to have severe crisis all his/her life? *Yes it is possible.*

29. Why is it important for a child with SCA to be seen by a haematologist?

The haematologist is specialized in the care of SCA, so he's in a position to give the best to the Hbss child.

30. Can Hbss use enhancement drugs to improve stature etc?

Not advisable because of the side effects of the enhancing drugs.

31. How can stigmatization of SCA patients be eradicated?

By education of the public, also SCA patients coming together in groups or clubs which will help them to improve their self esteem and boost their confidence that they are not outcasts.

32. Fertility options if both partners are carriers

(a) They can choose to adopt rather procreate

(b) They could do Hbss screening during pregnancy and abort Hbss fetus and allow non SS pregnancies to mature.

33. How is daily life affected by SCA?

Daily life is affected by SCA because it's a constant battle to avoid trigger factors.

34. Is banana good for SCA patients?

Banana is a good source of energy but is high in potassium so it should be consumed in moderate quantity.

35. Are there local herbs available to heal SCA?

Yes, There are some herbs which patients claim have anti-sickling properties and also some which correct anaemia but documented studies to back up these claims are not readily available.

36. How can abuse of drugs be prevented among Hbs patients?

By education of Hbss patients and by legislature i.e. making it difficult to access addictive drugs over the counter.

37. What type of food is best for SCA patients?

Balanced diet in addition to fresh fruits and vegetable and pushing oral fluids so as to avoid dehydration.

38. What are the complications of SCA?

Physical features include prognatism, bossing of the skill failure to thrive (small statue). Sickle cell crisis can be mild in the form of bone pain, it could lead to stroke or death. SCA are also prone to pneumoccocal pneumonia and osteomyelitis from salmonella infectious.

Complications of sickle cell anaemia depend on where the clumping of sickle red cells and blockage of the artery occurs. This can happen in any organ of the body.

39. Is there a permanent solution to SCA?

Management is to take care of pain and correct anaemia. Avoid trigger factors so as to minimize the number of crisis. Permanent solution is for 2 partners who are carriers of the trait not to procreate. For those already born, they have the choice of taking drugs daily and bone marrow/stem cell transplant.

There's hope of gene editing also which could eradicate the S gene totally.

40. Priaprism leads to infertility. Is this true?

If treated on time does not affect fertility. If prolonged, can lead to tissue damage which can result in erectile dysfunction.

Note:

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About the AUTHOR

DR. (MRS.) BOSE OLA is a medical practitioner with over 30 years experience, ranging from Sickle Cell Management to running a successful private hospital. With a particular passion to treating patients with Sickle Cell Anemia, she ran the sickle cell club of Adeoyo from 1992 to 1996. Even though she specialised in Anaesthesia before retirement, she now runs an NGO for people living with sickle cell anemia called **Chrisbo Hb Champions Club**. The monthly meetings are held at Chrisbo Medical Centre, Akobo, Ibadan pending the acquisition of a permanent venue for the Champions' Club.

About the BOOK

These are a compilation of questions asked at our HB champions club meeting which started on the 8th of November 2020.We have put them together by the suggestion of one of my mentors in the person of **Dr. Tony Marinho** *(St Gregory's Diagnostic Center)* who believes that distributing this will have a wider reach and serve as a reminder for club members too.

The monthly meeting holds every second Saturday of the month at **Chrisbo Medical Centre, Akobo, Ibadan** by 10:00am prompt.

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