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Prepaid Medicare Services Ltd

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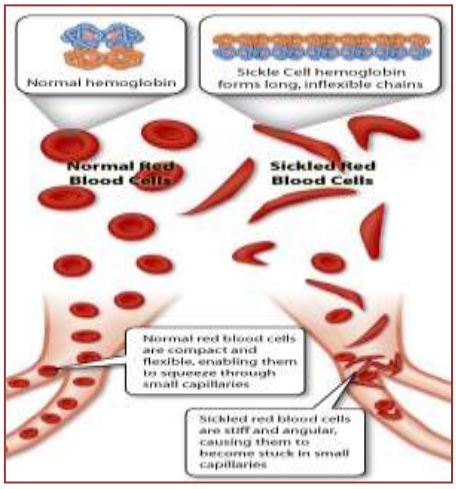
USEFUL LINKS

- **Sickle Cell Foundation in Nigeria**
- **WHO Factsheets on Sickle Cell Disease**

SICKLE CELL ANAEMIA



Sickle cell anemia is a disorder of the blood caused by an inherited abnormal hemoglobin (an oxygen-carrying protein within the red blood cells). The abnormal hemoglobin causes distorted (sickled) red blood cells. These red blood cells are fragile and prone to rupture causing anaemic conditions. The irregular sickled cells can also block blood vessels causing tissue and organ damage and pain.

Sickle cell disorder is by far the commonest inherited disorder in the world and three quarters of cases occur in Africa. In Nigeria, where it affects two out of every hundred children born, it causes suffering for innumerable patients and their families.

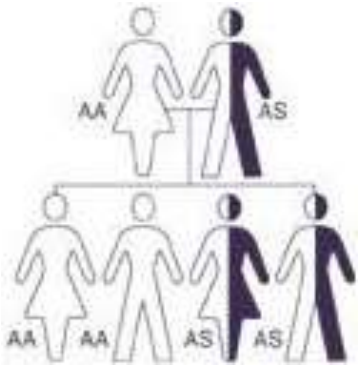
However, despite its importance, until now there has been no dedicated sickle cell center in Africa. This is partly because the very scale of the problem makes it difficult to see how to start.

Inheritance

Sickle-cell conditions are inherited from parents in much the same way as other physical and genetic traits. The types of haemoglobin a person makes in the red blood cells depend on what haemoglobin genes are inherited from his parents.

If one parent has sickle-cell anaemia (SS) and the other has sickle-cell trait (AS), there is a 50% chance of a child's having sickle-cell disease (SS) and a 50% chance of a child's having sickle-cell trait (AS).

When both parents have sickle-cell trait (AS), a child has a 25% chance (1 of 4) of sickle-cell disease (SS), as shown in the diagram.



How is sickle cell anemia diagnosed?

Sickle cell anemia is suggested when the abnormal sickle-shaped cells in the blood are identified under a microscope. Testing is typically performed on a smear of blood using a special low-oxygen preparation. Prenatal diagnosis (before birth) of sickle cell anemia is possible using amniocentesis or chorionic villus sampling. The sample obtained is then tested for DNA analysis of the fetal cells.

Management

Sickle-cell disease can be managed by simple procedures including:

- high fluid intake and a healthy diet
- folic acid supplementation
- pain medication
- vaccination and antibiotics for the prevention and treatment of infections
- a number of other therapeutic measures.

The most cost-effective strategy for reducing the burden of haemoglobin disorders is to complement disease management with prevention programmes. Inexpensive and reliable blood tests can identify couples at risk for having affected children. Such screening can be done before marriage or pregnancy, allowing couples to discuss the health of their family. Subsequent genetic counselling informs trait carriers of risks that the condition may be passed along to their children, the treatment needed, if affected by a haemoglobin disorder, and the possible options for the couple.

We welcome any comments, material or queries on this eNewsletter. Please contact us:

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