Sickle cell disease: a lethal advantage

The history of the sickle-shaped cell

Vox Pop:
One of my friends died of sickle cell anaemia. She died when she was about eleven.

BENJIE HICKS
Nobody else but us people that have pain knows how it feels.

VOX POP
It's a disease among blacks only.

VOX POP
Basically we knew that she died of some type of blood disorder, because she was always skinny and she never played like the rest of us did.

BENJIE HICKS
It feels like somebody's hitting you with a hammer.

VOX POP
All of us don't have it, it's just every now and then it pops up.

SUSAN RAE
Sickle Cell disease is a condition with which most African Americans are familiar. It's an inherited blood disorder which although most common in people of African origin also affects populations around the Mediterranean, throughout the Arab world, and into the Indian sub continent. In today's multi racial societies, it is a condition of which everybody should be aware.

It's estimated that over a quarter of a million children are born with sickle cell disease world-wide every year.

Sickle Cell disease drastically reduces the life expectancy of sufferers, who experience recurrent bouts of crippling pain. The typical symptoms have been recognised for centuries in Africa, and are well known to traditional healers:

JEMIMA DENNIS-ANTWI
Traditionally I would say that most people are aware of the disease in the form of the way the signs and symptoms present. And they give names such as (African names), they have in Fanti they say (African names). So every tribe has its specific name it gives, and it all tends to describe the signs of the disease, especially the pain that they go through with the disease.

SALIFU ISSA
SUB TITLES: In my language we call it coblerwerema - it affects the bones and flesh - the pain is excruciating, like burning. The bones feel as though they are going to break in two.

SUSAN RAE
Africans were taken to the Americas as slaves from the 16th century onwards, but despite the traditional recognition of the condition in Africa, it seems to have escaped the notice of American physicians until the beginning of the 20th Century.

Even then it was first diagnosed, not in an African American, but in a West Indian Student: - Walter Clement Noel, whose family tomb still lies on his native island of Grenada.

It was by chance that Noel happened to consult James Herrick, a doctor with an interest in haematology - the study of blood.

KWAKU OHENE FREMPONG
A dental student from Graneda walked into a hospital in Chicago, and showed up with symptoms that baffled the doctors at that time. And it wasn't until the blood smear of that patient was looked at, before sickle cells were discovered.

SUSAN RAE
What Herrick noticed under the microscope were some unusually shaped blood cells. His discovery led to a search for the cause and explanation of the disease, which was to take nearly half a century.

KWAKU OHENE FREMPONG
Sickle cell disease is a very well known disease now. We understand very clearly that red blood cells of patients with sickle cell disease behave very differently from normal red blood cells. In normal red blood cells the chemical, the protein called haemoglobin which is the oxygen carrier in the blood, always remains as a nice solution in the red blood cell. This makes the red blood cell very soft, and able to squeeze through very very small capillaries in the body. Sickle cells unfortunately behave differently. When the sickle haemoglobin is carrying oxygen it behaves pretty much like normal haemoglobin, but when it gives up the oxygen that it's supposed to give up to the body, then the haemoglobin in there begins to gel, and this gel forces the cells to become distorted. They become distorted into these sickle cells as we call them, which also become very stiff, and these stiff cells then cannot pass through the tiny capillaries, so they tend to block them. So there are two basic problems that we see in sickle cell disease, one is that these cells block the flow of blood to different parts of the body and where ever this blockage occurs, tissue becomes inflamed and eventually the tissue may get damaged if oxygen supply is not restored.