Sickle cell disease: a lethal advantage

Sickle cells and societies

SUSAN RAE
Today though, with generally improved living conditions, combined with modern medicine, sicklers are living longer and often to maturity. So the S gene is probably not diminishing greatly, although it will gradually become more dilute and spread through an increasingly racially mixed population.

Sickle cell disease however remains incurable, and affects about 75,000 people in the USA alone. Fortunately, the last thirty years or so have seen great advances in the way it is both treated and managed.The centre within the Children's Hospital of Philadelphia is one of ten specialist units for sickle cell research and treatment in the United States. The centre cares for children with sickle cell disease from birth up to the age of 18, and they take a comprehensive approach to the care of their patients. This involves a multidisciplinary team to address both the medical and psycho-social problems that sicklers face.

KWAKU OHENE FREMPONG
The only preventive treatment we have deals with the infection. We know now that children with sickle cell disease, when given penicillin twice a day can actually be helped to prevent the occurrence of pneumococcal infections. These pneumococcal infections are the leading cause of death especially in the first three years of life. So that's one of the main treatments that we have in the young children. We also have to educate the family very well about the signs and symptoms of infection, because even with the penicillin prophylaxis, and even with vaccines that we give them, the children can still get sick from these infections and die from it. And one of the more important things that families can do is to understand this risk and to rush children who show signs of infection to the hospital so they can be treated.

STEPHANIE TRIBBLE
A lot of people don't really know what sickle cell is, unless you talk to them, explain it to them. They don't know what it is. Because I had to inform the school what sickle cell is, so they can look for different things - when Kevin goes in to crisis, or if he have headaches or get a high fever, to let them know that they have to alert me.

SUSAN RAE
As a result of improved treatment and care of sicklers in childhood, their life expectancy has increased dramatically in recent years.

ACTUALITY EXAMINATION
...do this side again. Right, very good. Can you put your arms up for me. Don't let me push this side down.

KIM SMITH-WHITLEY
Sickle cell disease now is a much different disease than it was forty years ago. Within the last forty years we've understood and realised the importance of penicillin prophylaxis. We've understood the importance of new born screening and early identification. Changes in surgical techniques have taken us into a new era. Whereas life expectancy forty years ago was in the twenty to thirty age range, and now we're in the forty age range, and I think that in the next ten years we'll realise that we're almost at the equivalent of a life expectancy for the average African American.

KWAKU OHENE FREMPONG
In the United States now, forty four out of the fifty states have added testing for sickle cell disease to their new born screening programmes. So this is a programme that we think is

very important, and we really believe that throughout the world where sickle cell disease is common, new born testing programmes should be implemented.

SUSAN RAE
Dr Ohene-Frempong and his team have helped to set up Africa's first new born screening programme, in his native country of Ghana. Based at the Komfo Anokye Teaching Hospital in Kumasi, the programme is modelled on the one in Philadelphia.

They hope to identify sicklers at birth and improve the treatment that is offered to them, as well as to find out more about the way the disease develops in African Children.

MERCY ESSEL AHUN
The idea was to pick them up very early. The children who'll be picked up in this study will be followed up. So that the natural history of the disease would be followed. Because so far in Africa what we've done is we've had sickle cell clinics where people come to the clinic, but we haven't had a cohort. Now we follow them through to find out what happens to them. Because not much is known about what happens to the children like the first five years of life.

We do screening both for new-born babies and then for babies at the well baby clinics. Because in Kumasi about 60% of the babies are delivered by a trained health worker. The others are not. So these people are picked up at the well baby clinic. And what we do is a heel prick and we impregnate it on to, the blood on to filter paper and send it for examination. So it's really quite simple.

SUSAN RAE
The blood samples are sent to labs at the Noguchi Institute in Accra for analysis. Haemoglobin is extracted from the dried blood on the filter paper and then analysed using a technique called electro phoresis. An electric current is passed through the samples, which makes the haemoglobin travel across a gel. The different types of haemoglobin travel different distances and can be compared against a control and identified.

MERCY ESSEL AHUN
We have screened about twenty seven thousand babies and we've had more than five hundred with sickle cell disease, so we've found out that about 2% have sickle cell disease.

SUSAN RAE
Babies who are diagnosed as having sickle cell disease are enrolled in the clinic at the Komfo Anokye Hospital - where they are put onto penicillin prophylaxis, and have their general health and development regularly monitored.