OSCAR Nottingham
Sickle Cell Support Service

Sickle Cell Disorder and Thalassaemia Major

Sickle Cell Disorder and Thalassaemia Major are serious and dangerous inherited conditions that affect the red blood cells in people from areas where malaria is or was common. Therefore, they are most likely to occur in people with family backgrounds from:

- Africa
- The Caribbean
- Mediterranean countries
- The Middle East
- Parts of Asia

Children can inherit the conditions from their parents when both parents have Sickle Cell or Thalassaemia Trait, which developed as defences against malaria. The traits themselves are not illnesses nor can they develop into one. Frequently people can live their entire lives without knowing they have one.

It is possible for those with Sickle Cell Trait to sometimes exhibit symptoms but they tend to be less severe and a connection between Sickle Cell Trait and the symptoms is often not made by medical professionals.

Sickle Cell Disorder

Sufferers are most likely to have family backgrounds from Africa, the Caribbean, Middle East and India.

Ordinarily red blood cells are round or doughnut shaped and flexible. In those with Sickle Cell Disorder (also known as Sickle Cell Anaemia), red blood cells can collapse to become

sickle or crescent shaped and become rigid. This causes them to clump and become stuck in blood vessels. When they do so they cause enormously painful episodes known as a 'crisis'. These can last hours, days or even weeks and months.

Because during Sickle Cell Crises blood flow is restricted, tissue and internal organs are starved of oxygen, causing long term damage. Strokes are also very common, particularly in children.

> *"You can be in crisis for a day, two days, a week, a month. I've had crisis and its affected everything. I've had pneumonia in my liver, I've had gallstones, I've had blood clots in my brain and nearly died because of it.*
>
> *I've had a baby and I've been through labour and I'm not lying to you when I say having a crisis is worse than being in labour. "*
>
> *An OSCAR client*

Crises can be triggered by:

- Dehydration
- Stress
- Extremes of temperature and damp conditions
- Altitude
- Extreme exertion

Trying to reduce the frequency and severity of crises is vital. The goals are to relieve pain, prevent infections and damage to organs. This is done by:

- Food supplements
- Keeping hydrated
- Antibiotics
- Pain medication
- Healthy lifestyle with welfare needs addressed

In addition to crises, other symptoms of Sickle Cell Disorder include:

- Fatigue due to reduced oxygen
- Anaemia
- Eye disease

- Acute chest syndrome (a common cause of death)
- Vulnerability to infections

Thalassaemia Major

Is found mostly in people with family backgrounds from India, Pakistan, other Asian countries and many Mediterranean, North African and Middle Eastern countries.

People with the condition cannot make red blood cells and those that are made contain very little haemoglobin (iron).

Symptoms of Thalassaemia Major include:

- Growth delay and poor feeding in babies
- Fatigue
- Weakness
- Shortness of Breath
- Jaundice

OSCAR Nottingham

OSCAR Nottingham was started by a group of concerned parents of young Sickle Cell sufferers, who struggled to find information, overcome their many difficulties, and find people who could understand their needs. It became a Registered Charity in 1983 and currently its aim is to provide non-medical support to those with Sickle Cell Disorder, Thalassaemia Major and their families.

From February 2014 OSCAR Nottingham has been running a pilot project running until 31st July 2015 funded by NHS Nottingham City Clinical Commissioning Group designed to:

- Help prevent recurrent hospital admission for Sickle Cell crisis by supporting people affected by Sickle Cell Disorder through social and welfare interventions that help minimise symptoms.

- Minimise the suffering that Sickle Cell and Thalassaemia have on their lives and those of their families and carers, therefore reducing further the demand on health provision.

- Raise awareness amongst those groups most affected of the hereditary aspect of Sickle Cell and Thalassaemia. People with sickle cell or Thalassaemia trait may show no symptoms, but there is a 25% chance of them becoming the parent of a child with Sickle Cell Disorder or Thalassaemia Major respectively if their partner also has the trait.

- Raise awareness of screening and the genetic counselling available amongst affected communities and to encourage increased participation from these groups in screening.

This project brings together support for individual's health issues and support for social and welfare issues. We know this a is significant project since research has shown that Sickle Cell is now one of the commonest reasons for admission to hospital and has the highest rate of multiple admissions for individual patients.[1]

OSCAR also runs the Wellbeing and Health for You (WHY) project. This is open to everyone from the community and is free to users. Health activities are held periodically around the city in schools premises, community organisations and churches such as:

- Healthy eating
- Martial arts
- Belly Dancing
- Cancer awareness
- Food Intolerance
- Yoga
- Support to quit smoking

[1] S Lucas, D Mason, M Mason and D Weyman, *'A Sickle Crisis? A report of the National Confidential Enquiry into Patient Outcome and Death'*, NCEPOD, 2008, p. 7