Sickle Cell Disease

What is it?

Sickle cell disease is a genetic blood disorder characterised by abnormal haemoglobin, the protein in red blood cells that carries oxygen throughout the body. In individuals with sickle cell disease, the red blood cells become crescent-shaped (resembling a sickle) instead of their normal disc shape.

These abnormal cells can block blood flow, leading to pain, organ damage, and other serious complications.

Prevalence

Sickle cell disease is most common among people of African, Mediterranean, Middle Eastern, and Indian descent. It also occurs in people with ancestry from South and Central America, the Caribbean, and parts of Asia.

Diagnosis

Sickle cell disease is typically diagnosed through blood tests, including haemoglobin electrophoresis and genetic testing.

Life management

Individuals with sickle cell disease can take steps to manage their condition and improve their quality of life, including:

- Staying hydrated
- Avoiding extreme temperatures
- Getting regular medical check-ups
- Taking folic acid supplements
- Avoiding activities that can trigger crises, such as high-altitude or high-intensity exercise.

Employers need to acknowledge that employees diagnosed with sickle cell disease might require time off for treatment and medical appointments and allow flexibility with working hours, the tasks needed, and the working environment. If employees cannot work because of their illness, they might be entitled to statutory or company sick pay and should speak to their employers.

Genetic inheritance

Sickle cell disease is inherited when a person receives two abnormal copies of the haemoglobin gene, one from each parent. Individuals with one abnormal gene copy may have sickle cell trait, which usually doesn't cause symptoms but can be passed on to offspring.

Symptoms

Symptoms of sickle cell disease can vary in severity and may include:

- Fatigue
- Jaundice (yellowing of the skin and eyes)
- Pain episodes (crises), often in the bones, chest, abdomen, and joints
- Swelling in the hands and feet
- Frequent infections
- Delayed growth and development in children.

Complications

Sickle cell disease can lead to various complications, including:

- Acute chest syndrome - a condition similar to pneumonia that can be life-threatening
- Stroke
- Organ damage, particularly to the spleen, liver, kidneys, and eyes
- Leg ulcers
- Priapism (painful, prolonged erection).

Treatment

While there is no cure for sickle cell disease, treatment aims to manage symptoms and prevent complications. Treatment options may include:

- **Pain management** - using medications and other techniques to alleviate pain during crises
- **Hydroxyurea** - a medication that can help reduce the frequency and severity of pain episodes
- **Blood transfusions** - to increase the number of healthy red blood cells
- **Bone marrow or stem cell transplant** - this procedure may offer a cure in select cases.

References: Sickle Cell Society, NHS, NICE guidance.