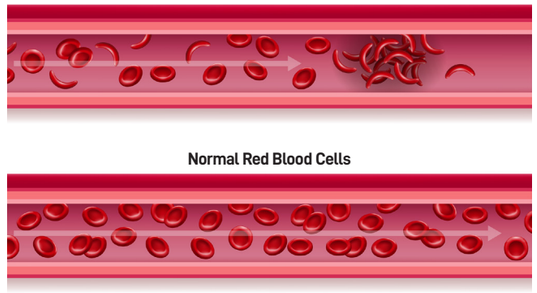
Blood Curdling

World Sickle Cell Day - Raising Awareness for a Genetic Disorder

Every year on 19 June, the world observes World Sickle Cell Day. This day is dedicated to raising awareness about sickle cell disease, a genetic disorder that affects millions of people globally.

Sickle cell disease (SCD) is a genetic disorder that affects the red blood cells. People with SCD have abnormal haemoglobin, which causes their red blood cells to become stiff, deformed and sticky. As a result, these cells cannot move easily through the blood vessels, leading to blockages and restricted blood flow. This can cause severe pain, damage to organs, and even death.

SCD is an inherited disorder, meaning that it is passed down from parents to their children. To develop SCD, a person must inherit two copies of the mutated haemoglobin gene, one from each parent. If a person inherits only one copy of the mutated gene, they may not develop SCD, but they may be carriers of the disease and may be at risk of passing it on to their children. SCD is more common in people of African descent, but it can also affect people from other ethnic backgrounds, including those of Middle Eastern, Hispanic and Mediterranean descent. In South Asia, the highest prevalence of the disease is in India, where over 20 million patients live with SCD.



In Singapore, the prevalence of SCD is relatively low compared to other countries. However, many people living with SCD in Singapore experience significant challenges because of the condition, particularly in managing pain and reducing complications.

DIAGNOSIS

There are many myths and misconceptions about SCD, which can cause difficulty in understanding the condition and finding appropriate care. For example, some people believe SCD only affects African Americans, or that it is contagious. These beliefs are untrue, and it is crucial to understand that SCD affects all races and ethnicities, and it is not contagious.

There are various ways to test for SCD, depending on the individual's age and the stage of the disease. Early diagnosis is essential, as it can allow for early intervention and management of the condition, which can improve life expectancy and quality of life.

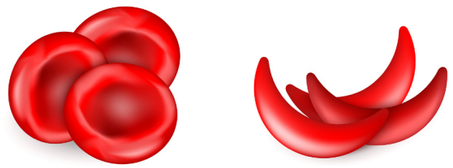
Raising awareness can help to reduce the stigma and discrimination faced by those with SCD. Many people with SCD face discrimination and prejudice due to misconceptions and stereotypes surrounding the disease. By educating the public about the disease, we can help to promote understanding and acceptance of those with SCD.

SCD is typically diagnosed through a blood test that looks for the presence of abnormal haemoglobin molecules. This test is often done as part of new-born screening in many countries, including Singapore. If a person is diagnosed with SCD, additional tests may be done to assess the severity of the disease and to look for any complications.

SYMPTOMS

One of the major challenges of SCD is that it is a "silent" disease that often goes unnoticed or misdiagnosed for years, leading to complications that could have been prevented if detected early. Some people with SCD may experience mild symptoms, while others may experience severe complications that can be life-threatening.

One of the most reported symptoms of SCD is severe chronic pain. This pain can affect many areas of the body and can be severe enough to interfere with daily activities, including school, work and social interactions. Pain management can be complex and challenging, often involving multiple medications

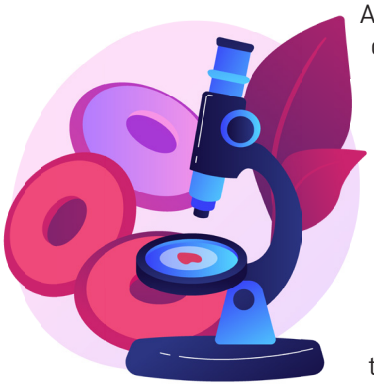


and regular hospital visits. Another significant issue in SCD is anaemia, a condition where the body does not have enough red blood cells to carry oxygen to the body's tissues. Anaemia can lead to fatigue and shortness of breath on exertion.

COMPLICATIONS

One of the most common complications of sickle cell anaemia is pain crises. These are episodes of severe pain that can last for hours or even days, and are caused by the sickle-shaped

red blood cells getting stuck in the small blood vessels. Pain crises can occur anywhere in the body, but are mostly felt in the bones, joints and abdomen.



Another complication of sickle cell anaemia is an increased risk of infections. The spleen, which helps to fight infections, is often damaged in people with sickle cell anaemia. This makes them more susceptible to infections, particularly those caused by bacteria like pneumonia.

SCD can also cause damage to organs, including the kidneys, liver and lungs. Over time, the sickle-shaped red blood cells can cause scarring and inflammation in these organs, which can lead to chronic health problems. In addition, people with sickle cell anaemia have an increased risk of stroke. The sickle-shaped red blood cells can block the blood vessels that supply the brain with oxygen, leading to a stroke.

Overall, SCD is a complex disorder that can cause a wide range of complications. However, with proper medical care and management, many people with SCD can live long and healthy lives.

TREATMENT

Individuals with SCD can benefit from working with a team of healthcare professionals to manage their condition successfully. This may include a haematologist, a pain specialist, physical therapist, and other specialists as needed. Additionally, it is essential to maintain a healthy lifestyle, including regular exercise, a balanced diet and adequate hydration.

While there is no cure for SCD, there are treatments available that can help to manage the symptoms and complications of the disease. The primary goal of treatment is to prevent and manage pain, which is often the most debilitating symptom of SCD.

Pain crises can be managed with pain medications such as opioids and nonsteroidal anti-inflammatory drugs (NSAIDs). In addition, hydration and rest can help to alleviate pain and prevent future crises. Another treatment for SCD is blood transfusions. Blood transfusions can help to increase the number of healthy red blood cells in the body, which can reduce the risk of complications such as stroke and organ damage. However, frequent blood transfusions can lead to iron overload, so people who receive transfusions may also need to undergo chelation therapy to remove excess iron from their bodies.

Bone marrow transplantation is another treatment option for SCD. This procedure involves replacing the patient's bone marrow with that of a donor who does not have SCD. Bone marrow transplantation can cure SCD in some cases, but it is a risky procedure that is typically reserved for people with severe symptoms.

Finally, people with SCD can benefit from ongoing medical care and management. This may include regular check-ups with a healthcare provider, vaccinations to prevent infections, and genetic counselling to help manage the risk of passing the disease on to future children.

In conclusion, while there is no cure for SCD, there are several treatments available that can help to manage its symptoms and complications. With proper medical care and management, many people with SCD can live long and healthy lives.

IMPORTANCE OF RAISING AWARENESS ABOUT SICKLE CELL DISEASE

Raising awareness about SCD is crucial for several reasons. Firstly, it can help to improve the diagnosis and treatment of the disease, especially in countries where the disease is not well-known. By increasing awareness among healthcare professionals and the public, more resources can be allocated for research and treatment of the disease.

Secondly, raising awareness can help to reduce the stigma and discrimination faced by those with SCD. Many people with SCD face discrimination and prejudice due to misconceptions and stereotypes surrounding the disease. By educating the public about the disease, we can help to promote understanding and acceptance of those with SCD.

Finally, raising awareness can help to improve the quality of life for those with SCD and their families. By providing information and resources about the disease, we can help those affected by SCD to better understand the disease, and manage its symptoms and complications. This can lead to improved health outcomes and a better quality of life for those with SCD. PRIME

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