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Feature Articles

## Sickle Cell Disease In Children Challenges And Comprehensive Care

July 3, 2024

By Dr. M. Giripunja, Consultant-Haematology, Manipal Hospital, Mysuru

Sickle Cell Disease (SCD) is a genetic disorder that affects the haemoglobin and is a major challenge in affected children. This is an inherited condition that leads to formation of haemoglobin (haemoglobin S) which creates hyper-rigid sickle-shaped red blood cells. These malformed cells may obstruct the bloodstream causing many sicknesses. Meeting the needs of children with SCD entails a holistic and continuous care since one has to manage the above difficulties appropriately.

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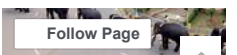
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#### Understanding the Challenges

**Pain Crises** – Vaso-occlusive crisis or pain crisis are sudden painful episodes, occurring when crescent shaped red blood cells stick to the walls of blood arteries. Some of the complications observed in children suffering from vaso-occlusive crisis, are traumatising experiences and may require hospitalisation for control of the pain. Pain crises may also cause organ damage.

**Infections** – SCD children are often susceptible to spleen problems, and hence prone to infections more than their healthy counterparts. The spleen which has a major role filtering bacteria out of the blood stream is mostly malfunctional in children with SCD, making them prone to infections particularly those caused by encapsulated bacteria such as Streptococcus pneumoniae and Haemophilus influenzae. Vaccination on regular basis, as well as prophylactic antibiotics to avert fatal infections.

**Chronic Anaemia** – Sickled red blood cells have shorter life-span. The dead RBS clog within circulation and therefore, the patient suffers from chronic hemolytic anemia. This condition is characterised by recurrent fatigue, pallor and poor physical growth or development.

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Dr. M. Giripunja

Anemia treatment is mainly blood transfusion and constant checking of the haemoglobin levels. Also, adequate intake of folic acid and other nutrients should be increased to allow formation of healthy red blood cells.

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Organ Damage – Chronic sickling leads to progressive damage to so many organs such as the spleen, liver, kidneys, and lungs. Such damages may result in complications such as acute chest syndrome, pulmonary hypertension, and chronic kidney disease. Prevention of stroke is also very important due to high incidence of cerebral vasculopathy that affects SCD patients.

Psychosocial Issues – SCD is a long-lasting condition, and the constant medical treatments that the child undergoes can result in mental health issues. The symptoms often lead to uneasy feelings, series of hospitalisation and physical restriction that consequently ends with anxiety, depressive thoughts and social withdrawal.

Counselling and mental health support is therefore important in tackling these concerns and teach children with SCD how to face the challenges they encounter each day.

Educational Challenges – Another common problem with SCD is that due to numerous doctor's visits and hospitalisations, children experience educational losses.

Moreover, with chronic anaemia, comes fatigue and cognitive impact. This, in turn, affects their learning and academic performance. So, the use of individualised education plans (IEPs) and support in school is crucial to make these children continue learning and achieve their goals at school.

To summarise, these specific challenges can be addressed by Comprehensive Care which includes, regular check-ups with haematologist to monitor the progression of SCD and identify any emerging complications, pain management, adequate hydration, relaxation techniques, prevention of infections by prophylactic medications and vaccination, nutritional support with proper supplements, psychological, and educational support.

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