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Understanding the landscape of sickle cell disease in India

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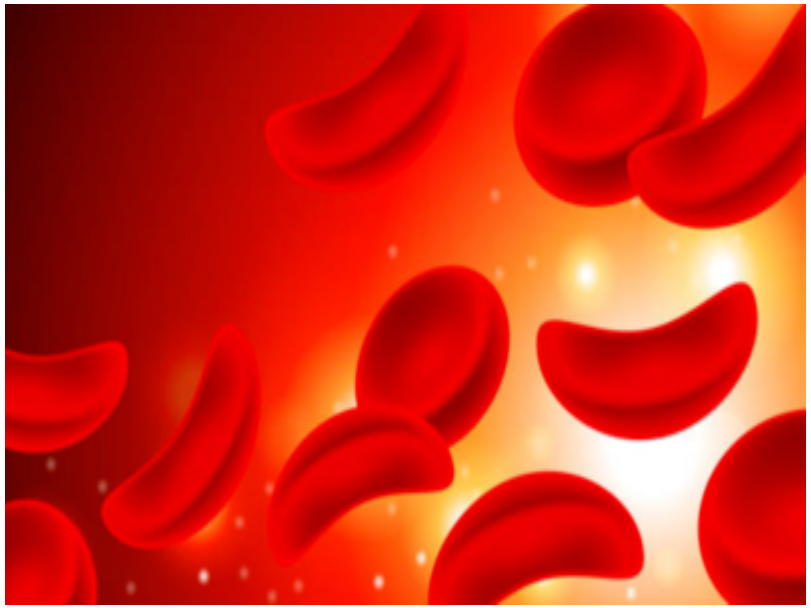

Sickle cell disease (SCD) is an inherited condition that affects the oxygen-carrying capacity of blood. It is most prevalent in Sub-Saharan Africa, the Middle East, India, and some parts of the Mediterranean. India is home to over 20 million patients with this condition, making it the country with the second-highest burden of the disease globally. This condition disproportionately affects marginalised and tribal communities, affecting 1 in 86 births.

Sickle cell disease is a major health concern

SCD is a significant public health concern in India, particularly among tribal populations. Normally, the RBCs that carry blood in the body are shaped like a doughnut without a hole in the centre. However, individuals with SCD have misshapen RBCs that are curved like a sickle. Besides affecting the amount of oxygen they can carry; such RBCs also tend to clump together and block the flow of blood. The symptoms of this condition are severe pain, difficulty in vision, growth delay in children, and frequent infections. Moreover, this condition can also be life-threatening and can cause several complications such as organ damage, gallstones, leg ulcers, deep vein thrombosis (formation of clots in deep veins), stroke and blindness.

It is highly encouraging to health workers that the Government of India plans to eliminate SCD by 2047. There are several ground realities that pose a considerable challenge to this gigantic task which is possible only through government machinery. The prevalence of the disease among marginalized and tribal communities impacts several outcome variables such as inadequate access to care, poverty, nutrition, low socioeconomic status, and home environment. The mortality related to sickle cell disease is also very high in certain regions with almost 20%

of children dying before they reach the age of two and 30% of children in tribal areas dying before they reach adulthood.2 There is also an acute lack of awareness among those affected and their families, resulting in many patients seeking medical aid only at advanced stage of the disease, once organ damage has set in. There is also a lack of awareness among medical practitioners as well has health care workers about SCD and its treatment, which may often delay the diagnosis.

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While screening is vital to ensure that patients receive care on time, it is equally important that they receive counselling to distinguish between those with SCD and those who are carriers of the disease (having only one copy of the sickle haemoglobin gene). Once diagnosed, there are several cost-effective strategies available to alter the prognosis of the condition such as immunization against pneumococcus, penicillin prophylaxis to prevent severe infections, hydroxyurea to reduce sickling and related complication and the availability of comprehensive care. The patient should be under the care of a multidisciplinary team which can hand hold the family through challenges that emerge at different age groups. In the absence of a formal referral pathway, many patients in the country are at the mercy of quacks. Many patient receive untested unsafe blood from professional blood donors thus resulting in transfusion related infections. India was found to have the greatest unmet need for blood units in the world.

Newer treatment options offer hope

The real world data on outcomes from sickle cell disease and its research remains fragmented and scarce. In this setting, new treatment options offer a glimmer of hope to these patients and their families.

Hydroxyurea is often used to prevent sickling of blood cells and complications arising due to SCD. Although not a new drug, the access remains poor in many communities and it is underutilized due to fear of side effects and inadequate awareness.2 New drugs that have come such as L Glutamine and crizanlizumab, suffer from either access and cost issues in India or poor data to support its use. In this setting, both bone marrow transplantation and gene therapy that are curative treatment options, offer the promise of a one-time cure from this chronic disabling disease. The access to bone marrow transplantation has improved significantly in the last decade in India for both children and adults. However, it still needs infusion of an allogeneic product thus posing risks for

graft vs host disease and long-term immune suppression. Gene therapy on the other hand, is performed by modification of patient's own stem cells, either by correcting the defective gene or improving hemoglobin F production. Newer drugs that have been effective in thalassemia and hemolytic anemias are also finding their way to sickle cell disease treatment armamentarium. These treatment options thus bring in a lot of hope to these communities affected by this painful disease which impacts quality of life of not only the patient but also the family.

No public health initiative can be impactful, unless we address the issues of screening and reducing the burden of the disease in the country. No country with such a disease load can treat its citizens with the best curative and supportive care options lifelong. Hence, the initiative of mass screening for carrier state and effort to eliminate the disease although ambitious is the only way ahead to address this disease. By bringing together multiple stake holders aligned to the same goal directed at raising awareness, improving screening, post screening counseling, education and capacity building at different levels we definitely become a formidable force to reckon with. Addressing the multifold challenges associated with the condition and adopting evidence-based interventions are necessary to reduce the burden of the disease and ensure that future generations can manage it effectively.

(Author: Dr. Nita Radhakrishnan, Additional Professor and HOD, Department of Pediatric Hematology-Oncology, Post Graduate Institute of Child Health, Noida)