Strengthening the Societal Pillars for effective management of sickle cell disease

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June 6, 2023

Abstract

Sickle cell disease (SCD) is a common inherited blood disorder of public health concern in India. Individuals with SCD may encounter stigma, discrimination, and misconceptions about their condition, leading to feelings of isolation and frustration. It is important to note that the psychological impact of SCD can vary among individuals. Some may develop resilience and coping strategies to manage the challenges, while others may require additional support from healthcare professionals, psychologists, or support groups to address their psychological needs. The multidisciplinary approach involving medical care, psychological support, and social interventions is crucial in improving the overall well-being of individuals living with sickle cell disease. While the clinical aspects of SCD are significant, it's also important to consider the psychological aspects that can affect individuals with the condition. In this context, there is a pressing need for societal support pillars which empower SCD patients and improves their quality of life. In this context, there is a pressing need for societal support pillars which empower SCD patients and improves their quality of life. The impact of societal support pillars on SCD patients is substantial. Strengthening healthcare systems, fostering robust social support networks, facilitating inclusive educational environments, and raising public awareness are integral components of comprehensive care for individuals living with SCD. All these pillars can significantly improve the quality of life and outcomes for SCD patients, ultimately leading to a more inclusive and supportive society.

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Conflict of Interest: None

Source of Funding: Nil

Abstract:

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Keywords : Sickle Cell Disease, Psychosocial illness, Support Systems, Stigma

Highlights

- Sickle Cell Disease patients often suffers from psychosocial illness.
- Societal support may improve the psychological well-being of SCD patients.
- Family, teachers, self-help group, traditional healers, genetic counsellors may act as support system.
- By Self-empowerment SCD patients may overcome with psychosocial illness.

Sickle cell disease (SCD) is a genetic disease caused by a mutation in the HBB gene (HBB:c.20A>T) located on chromosome 11 and affects the structure of hemoglobin, the protein that carries oxygen throughout the body. This abnormal hemoglobin polymerizes during the physiological stress and increased requirement of oxygen leading to change of shape of the red blood cell from round shape to sickle shape. Abnormal sickle shape erythrocyte alters the normal blood flow in small vessel leading to ischemia and reperfusion injury and acute painful crisis[1] .

It is estimated that 3,00,000 infants are born with SCD annually[2] , off which India contributes to approximately 44,000 birth of SCD[3] . The prevalence of SCD is higher in countries of sub-Saharan Africa, Middle East, central America and South East Asia. Three countries (India, Democratic Republic of Congo and Nigeria) shoulder half of the global burden of SCD across the globe and therefore SCD is a public health concern. In India, high prevalence of SCD has been observed in the underprivileged and marginalized populations (Schedule Tribes and Schedule Castes)[4].

Psychological problems of SCD

Apart from the clinical illness, SCD patients often suffer from mental illness in the form of stigma due to fear of non-acceptance by the community, fear education, livelihood and marriage (figure 1). Additionally, children's psychological health is negatively impacted by the disease's chronicity, hospital stays during the pain episodes, withdrawal from normal social environments, bullying at school and poor focus on academics which is upsetting for both children and their parents or guardians. This stigma may lead to psychological distress and disempowerment. Several studies show that depression among SCD patients is more susceptible as compared to the general population. In a PiSCES study, it was discovered that 6.5% of people with SCD have anxiety and 27.5% have depression. An individual with SCD, anxiety and depression were associated with regular episodes of pain which lead to poor physical and mental health. Therefore, regular monitoring of depression and anxiety in SCD patients along with their treatment is recommended[5] . Furthermore, caretakers of SCD patients (1-10 years) describe a wide range of difficulties in most categories, including everyday tasks, financial hardship, quality of life (QOL), depression and parent's adjustment related to child illness and therefore a mechanism for managing psychological issue pediatric SCD patients in needed[6] . A study from Nigeria shows that most participants believed that society had a poor perception and attitude towards SCD patients. In the adolescent group (14-18 years) it was found 23% of the population had com-

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plained of teasing and bullying in school for SCD students. Additionally, 55% of SCD patients of adolescent age were depressed and 88% were worried for their health condition[7] . Another study from Nigeria revealed that adolescents with SCD generally express emotional disturbance and misbehavior. Their psycho-social problems include limitations on career options and difficulty in finding a spouse.

Further, findings suggest that social workers ought to work in the health sector to assist adolescents with SCD during their health examination[8] . In a study from Brazzaville, it was found that in children/adolescent group 76.1% had disempowerment, 29.9% had anxiety and 5.5% had depression. Study suggested that stronger focus on education, behavior modification, and communication is necessary to improve the quality of care due to the prevalence of psychiatric illnesses in children and adolescents who live with SCD[9] . A previous study from India found that children with SCD have more psychological issues than children without SCD. Further findings suggested that establishing health care services for children with SCD, facilities for early detection and treatment of psychosocial disorders should be included[10] . Another study from India found that SCD patients had considerably lower health-related quality of life (HRQoL) than other chronic non-communicable disorders[11] .

At this state, patient needs a support pillar for assuring good quality of life. Patients with SCD frequently experience psychological issues as a result of the effects of their pain and other symptoms on their daily life and how society views them. Both children and adults with SCD were shown to have psychological complications including ineffective pain management techniques, anxiety, sadness, neurocognitive impairment, and a poor quality of life due to functional limitations in day-to-day life. Additionally, one of the most significant causes of psychological stress for young people is marriage or finding a spouse. Without knowledge and awareness of sickle cell inheritance, it can be challenging to choose a spouse free of the disease or a characteristic, as well as to determine whether or not children will be born with the illness in the future. The overwhelming part is that patients with the condition (SCD) continue to face difficulties in managing the disease.

Over the course of a generation, medical care has significantly increased patients' quality of life and lifespan. However, the current situation regarding the recognition of the clinical implications of psychological complications and their management within a multidisciplinary context continues to be unsatisfactory[12] . There is a need for support pillars at different levels for coping with the disease. Support pillars are the system of social and psychological aspect that is directly linked with the patient capability and the family for coping-up the disease effectively by providing support.[13]The network of family, school teachers, self-help groups (SHG), traditional healers (in certain societies like the indigenous populations), genetic counselor and most importantly the affected person itself is crucial for the effective management of SCD. They all work together to make quality care at social and personal levels to make permeate choices for premarital and preconception counseling to have healthy children with accessibility of preventive measures run by the government.

1. FAMILY

Families of SCD individuals confront a variety of difficulties, from psychosocial issues to economic difficulties. The preliminary support pillar of SCD patients is their family. They provide physical, emotional as well as economic support to the patients. Proximities of family member and friends give SCD patients the feeling of security and confidence. Spending time with family enhances the benefit of treatment and motivates the patient for healthy habits. Family can help create a positive environment for recovery. Along with family support to the patients of SCD, family resilience is important firstly for examining family dynamics, enduring a crisis, making the most of opportunities for growth, and learning how to help others in similar circumstances. Family resilience is viewed as a process that develops over time as opposed to a quality. It opens a channel for identifying and developing crucial family strengths and resources that can lessen stress and susceptibility in high-risk situations, promote healing and growth after crises, and help families get through protracted adversity.[14]

There is need of education in terms of (i) knowledge of the child's health condition, (ii) common symptoms and care, and (iii) psycho-social problems and coping mechanism. Parents, especially mothers, have practical

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and psychological challenges when raising a child with SCD.[13]

Health education can impart mothers the knowledge and skills they need to reduce or eliminate the conditions that might lead to SCC, including as keeping the child warm, maintaining hydration, making the environment jovial, and engaging in lesser physical activity. This approach enables mothers to keep their independence while asserting control over the welfare of their children.[15]

In tribal context, the women play a significant role in the management of their natural, social, and economic resources. However, they continue to be behind because of traditional values. When compared to men, women have a higher social status and economic worth in tribal societies and women are considered more essential than their male counterparts in the upbringing of children, as men are typically involved in social and economic activities rather than caring for their families. In the tribal setting, women must be imparted health education since they play a vital role in the household. Most importantly, this health education must be provided in their own tongue so that they can comprehend SCD. Previous study has found that SCD patients experience a great sense of acceptance among the community from their families, friends, and the school support[17].

2. SCHOOL HEALTH PROGRAM

School health program is another aspect for creating awareness through information, education and communication (IEC).[18] The objective of the IEC at the school level is to generate awareness among students and teachers as well. Teachers may be cognizant about the symptoms and primary care of SCD patients. Since, teachers are the first point of contact for the child at school and therefore he/she can provide immediate care to a SCD child and therefore reducing morbidity. Awareness generation at community level also helps in preventing the stigma towards the disease. By spreading awareness about the psychological issues associated with SCD in government, private, and tribal ashram schools, we can foster a compassionate and inclusive environment that empowers students, supports their mental well-being, and ensures a brighter future for all.

3. SELF- HELP GROUP

In society several problems related to illiteracy, lack of skills, financial hardship, and false stigma related to diseases are present which need collective efforts. In this regard, self-help groups are an emerging social resource for improving healthcare facilities. Existing self-help groups can be included in the health system for awakening the community about SCD. They provide social and emotional support, decreases the feeling of isolation, provide practical knowledge to patient and families for better understanding of the disease. Self-help groups may overcome the discrimination associated with disease disability and enhance communication among the members of a family, professionals, nurses and counselors who deal with related problems. SCD affected family members can become a part of self –help groups as these groups are engaged in diverse set of activities including health care, education, microcredit, campaigning, and rehabilitation which facilitates empowerment among families. The belief behind self-help group is that the shared experience of a group is highly valuable in the promotion of understanding and healing mental health (venting out frustration stress and tension)[19]. Self –help groups act as a venting therapy for SCD affected families and patients too.

4. TRADITIONAL HEALERS

As SCD is more prevalent in the tribal population, traditional healers can play an important role in improving health awareness programs, screening and treatment outcomes directly. Traditional healers are an integral part of the pluralistic healthcare system, and concomitantly in tribal cultures the traditional healer is often socially empowered. These healers typically use indigenous methods that go beyond the scope of contemporary medicine. The community can quickly reach these healers, and they are highly regarded. Thus, traditional healers, enjoying authority and faith in tribal community, and are crucial in prompting the people for accepting diagnosis, treatment and management of SCD.[18] The participation of traditional healers in the SCD project can be crucial, especially in tribal areas. Hence, it's a felt need that the services of traditional healers should be integrated with the program and the expertise of SCD should be shared with the healers.

5. GENETIC COUNSELOR

A genetic counselor is perhaps the most important support pillar for patients and their families. They are healthcare professionals who help patients to interpret results of genetic tests and negotiate decisions about reproduction and treatment. They not only impart awareness of the disease but also provide strength emotionally, psychologically, and in social behavior. They provide proper information regarding marriages and also give guidance to the ANC and PNC patients. As the availability of genetic counselor is not possible in every health care center, so training should be imparted to all the available health staff such as doctors, nurses, counselors for counseling patients with genetic diseases. The goal of genetic counseling is to help people make thoughtful, informed and balanced decisions about having children by educating them on how hereditary succession can affect the likelihood that certain genetic diseases, like SCD, will develop or recur in the future.[20]

6. SELF-EMPOWERMENT

The holy text of Bhagwat Gita emphasizes the importance of self as *"Man is made by his beliefs. As he believes, so he is."* [21] Thus, empowerment of the self is very important which involves educating the individual about his own health condition so that he is able to confidently make his own decisions and undertake self- care. The government took many initiatives for the self-empowerment of SCD patients such special guidelines in education system, social security *etc* .[22] . Person with SCD frequently experience episodes of extreme pain that necessitates hospitalization and restricts their everyday activities. The pain is often throbbing, lasts for hours to a week or longer, and tends to move over the body hence person (SCD patient) comes under the purview of physically disability as they are not able to work as much as they are required. In this scenario, skill development initiatives increase productivity and reduce mental stress leading to improved living standard of SCD patients. If these patients are trained in any skill then they can work from home and initiate small-scale businesses for their livelihood. Since students with SCD tend to have low self-esteem, building their skills is crucial for their overall development because it has a direct impact on how they perceive about themselves. In due course of time, they learn to balance their emotional and social lives with their physical health, which is crucial for becoming independent.

Conclusion

The disease elimination initiative is a major strategic and political challenge for governments, civil society, academia, and the private sector. The government has launched a number of SCD eradication initiatives, such as training, screening, data management, and financial help, but it now needs to develop a multi-sectoral strategy under a single umbrella with the shared objective of eliminating SCD by 2047. The psychosocial effects of SCD are substantial. Low self-esteem and a sense of hopelessness emerges in SCD patients due to ongoing discomfort (recurrent pain episodes), frequent hospital visits, a lack of professional possibilities, dread of an early death, financial load, and societal ignorance. Ultimately, these instances point to depression in patients. For reducing psychological issues among SCD patients there is a pressing need for including the six support pillars, as suggested here, in the road map of SCD elimination programs. This will create a strong network for awareness about disease, the treatment and monitoring of patients. The inclusion of support pillars in healthcare facilities and ecosystem would guarantee SCD patients a high quality of life because they serve as a tipping point for their psychological well-being. Efficient management of SCD requires combined approach of clinical parameters along with psychological parameters. These pillars offer a comprehensive approach that encompasses various aspects of support, education, and empowerment for individuals and families affected by SCD. The implementation of a comprehensive and multidisciplinary approach by the government is crucial for addressing the diverse needs of individuals affected by SCD. To ensure effective support, a well-planned infrastructure or roadmap should be established. By integrating these pillars, policymakers can create a supportive environment that addresses both the physical and psychological burdens associated with sickle cell disease. This holistic approach will not only improve the quality of life for individuals with the condition but also promote a more inclusive and compassionate society that values the well-being of all its members Just like a vibrant garden, the study of SCD has experienced remarkable growth, encompassing the intricate aspects of psychological factors within the management approach. With

a strong foundation established, it radiates a beacon of optimism for numerous individuals, spreading its therapeutic influence to those around the globe who endure the burdens of SCD "**The goal of the support pillars is to provide "happy, prolific and long life those who live with SCD"**

Ethic statement - Not applicable

Legend to figure:

Figure 1: SCD patients moving towards psychological complications leading poor quality of life

Figure 2: Role of support pillars for betterment of SCD patients

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Problem faced by SCD patients

Training And Awareness Campaigns