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Qualitative Exploration of Experiences of Tribal Patients with Sickle Cell Anemia in a Southern State of India

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ABSTRACT

Objective Sickle cell disease is a chronic and self-incapacitating disease intensely affecting patients and their families. The objective of the study is to study experiences and explore the needs of sickle cell anemic children and adolescents to design patient-centric services to enhance their quality of life. **Methods** The descriptive study was conducted in a southern state of India. Primary data collection was done with help of in-depth interviews of 6 to 19 years old patients diagnosed with sickle cell anemia for at least one year and their caregivers. **Results** The respondents associated the disease with frequent episodes of pain in the shoulders, knees, arms, and chest, fatigue, shortness of breath, suppressed hunger, and insomnia. The caregivers were aware of the precautions and treatment advice to the children, but the patients were partially aware of the disease and the triggering factors for the crisis. The patients reported low esteem, feelings of hopefulness, and being a burden on their families as a result of frequent pain, hospitalizations, loss of schooling, and employment which were a source of constant anxiety and depression in the young patients. **Discussion** SCD should be looked at through a bio-psychosocial lens as it emphasizes how biomedical aspects of the disease interact with family systems, socio-economic factors, and psychosocial aspects to produce guilt, shame, anxiety, and feeling of hopelessness that impede effective coping. Therefore, psychoeducational interventions, cognitive behavioral therapy, and non-pharmacological techniques must be included in the provision of comprehensive care to the patients.

INTRODUCTION

Sickle cell disease (SCD) is one of the most common monogenic disorders with an autosomal recessive inheritance. [1] It affects over two million people globally and is particularly common in Mediterranean countries. [2] The formation of long fibers within the RBCs causes a distorted sickle in those whose ancestors came from Sub-Saharan Africa, Saudi Arabia, India, and shape and eventually leads to increased hemolysis and vaso-occlusion of sickle red cells. The disruption of blood flow causes vascular occlusions, hemorrhages, infarctions, and ischemic necrosis of tissues and organs throughout the body. As a result, there may be several complications, including recurrent vaso-occlusive crises with stroke, splenic sequestration crisis, aplastic crisis, infections, bone damage, jaundice, leg ulcers, priapism, delayed growth, fatigue, and pain episodes. [3] Physiological complications for children include pneumococcal infections, meningitis, osteomyelitis, cerebral vascular infarction, and enuresis. The symptoms alleviate the quality of life of the patients. [4] Understanding the importance of sickle cell disease, the UN General Assembly, on December 18, 2008, adopted a resolution calling sickle cell anemia a public health problem and one of the world's foremost genetic diseases. [5]

Sickle Cell Disease in India

India, along with Nigeria and DR Congo, accounts for 50% of the world's SCD burden. [6] It is estimated that in India, there are 5,200 live births each year with SCD, which makes it a major public health problem. The first description of sickle hemoglobin in India was by Lehman and Cutbush in 1952 in the tribal populations in the Nilgiri hills in south India. [7] Dunlop and Mazumder also reported the presence of sickle hemoglobin in the tea garden workers of Assam who were migrant laborers from tribal groups in Bihar and Odisha. [8] Since then many population groups have been screened and the sickle cell gene is prevalent among the socio-economically disadvantaged ethnic groups of scheduled tribes, scheduled castes, and backward classes in India due to consanguineous marriages. [1], [9], [10], [13]

According to a survey by the Indian Council of Medical Research, 20% of children with the sickle disease in India die by the age of two, and 30% of children with SCD in tribal communities die before they reach adulthood [11], [12]. SCD has remained a neglected field of research in India and the magnitude of the problem has never been properly emphasized. This is

large because most of the reports spread a misconception that the sickle cell gene is confined only to tribal people and scheduled castes in India. The other reasons are the hereditary nature and no cure for the disease. [14]

The average life expectancy for individuals with SCD is estimated to be between 42 and 48 years of age [15]. But it was noted through analysis of secondary data that the average age of deaths in tribal population with SCD in the southern state of India was 24.24 years and the maximum reported deaths were between the age of 11 to 30 years of age during 2007 to 2017. This made it important to understand the issues faced by young sickle cell patients. The objective of the study was to explore the lived experiences of sickle cell anemic patients aged between 6 to 19 years in a southern state of India.

MATERIALS AND METHODS:

METHODS

It is a descriptive, cross-sectional study carried in a southern state of India. Primary data collection was done in September 2018 with help of in-depth interviews with patients aged between 6 and 19 years and diagnosed with sickle cell anemia for at least one year and their caregivers. An attempt was made to obtain a varied sample about age, time of diagnosis, and sex. The interviews were transcribed and codified and analyzed line by line to compare similarities and differences. This led to the creation of categories for experiences and needs of sick individuals with sickle cell disease. Secondary data regarding newborn screenings, outpatient treatment, hospitalization, blood transfusions, and deaths of the patients were analyzed through data in registers of the tribal hospital in the area.

RESULTS AND DISCUSSION:

RESULTS

The sample consisted of seven girls aged between 6 years to 19 years and five boys aged between 7 years to 17 years, who had been diagnosed with SCD for the past one to eight years. All the respondents belonged to similar social and economic backgrounds, belonging to low-income families with parents working in the informal sector as daily laborers in tea estates or bakery shops. Two adolescents belonged to the single-parent family after their fathers had left

the family as a consequence of children being diagnosed with SS disease. The family size of the patients ranged from five to seven with patients having two to three siblings in the family. Three boys aged 7 years, 11 years, and 17 years and three girls aged 14 years, 17 years, and 19 years had dropped out of school.

The diagnosis of SCD among most of the patients occurred after episodes of acute pain, infections as cough and fever, fainting, and during pregnancy, while three respondents reported being diagnosed by active screening after their siblings were known to have SCD. In all the cases, the caregivers were aware of the precautions and treatment advice to the children, but the patients were partially aware of the disease and the triggering factors associated with the sickle cell crisis. They associated the disease with frequent episodes of pain in the shoulders, knees, arms, and chest, fatigue, and shortness of breath, and also complained about suppressed hunger and insomnia. The caregivers, as well as patients, appreciated the provision of subsidized care and prompt quality treatment by the tribal hospital under the sickle cell program. The respondents visit the tribal hospital in the area once in one or two months for regular blood tests and medicines, which are provided free of cost to them. Few of the patients admitted that they don't take medicines regularly despite knowing its necessity for their well-being, due to bad taste and being tired of eating medicines daily because it does not treat the disease. It was also noted that they have to get hospitalized one to six times a year due to severe pain, breathlessness, and chronic fever.

It was observed that most of the patients were quiet and started interactions after the second or third visit, while one of them refused to come out of his room during the researcher's visits to the home. The mother of this seven years' old boy said, *"He used to go to a government school but now he has dropped out for the past 2 months. Since then he keeps silent and doesn't speak a word out. And if anyone comes to the house he shuts the door."*

The patients reported low esteem, feelings of hopefulness as a result of frequent pain, hospitalizations, loss of schooling and employment which were the source of anxiety and depression in the young patients. In the fourth visit made to her by the researcher, a fourteen years' old shy tribal girl who has to be hospitalized frequently due to pain crisis said, *"I wanted to become a cop but I quit school after 6th standard due to increased episodes of pain. I don't like it, but I will have to get admitted again to the hospital after some time."*

SCD also implicated itself through episodes of fear of death, the guilt of being a burden on the family, loss of morale, and higher risk for marital dysfunction. The levels of stress and self-pity were so high that it left many of the children in tears while expressing their emotions in front of the researcher. An eight years' old boy said, *"I feel I am a burden on the family. My father left the house when I was diagnosed with the disease. My elder brother also has the disease. Due to me, a mother cries and has to work day and night."*

A nineteen years' old girl said, "*I was diagnosed when I got pregnant. My child got aborted and I was sent back to my parents. I can't sleep. I feel tired and restless all the time. I am hurting everyone."*

Very few patients were aware of the precautions to be taken such as taking more fluids, avoiding extreme temperatures, activities demanding physical exertion, too much exposure to Sun, emotional stress, smoking, getting regular vaccinations, and practicing good hygiene to prevent infections. The two patients who followed the precautions showed positive signs in education and reported very less episodes of crisis. When it was enquired about the triggering factors for pain episodes, most of the respondents were silent or answered that they had never pondered about it. They were given time to think over it, and it was noted that in subsequent visits by the researcher, most of them were able to identify risk factors that initiated intense pain. The risk factors identified were stress due to family issues, loss of schooling, and fear of unemployment, physical exertions after cycling or playing in the Sun, and cold temperature. These factors led to acute pain, which amplified stress for the patients and their families.

A fifteen years' old girl who aspired to become a doctor said, "*There are regular flights between my brother and sister, and sister is always crying. I don't like it, I am not able to cope up with that situation and there is no-one I can even discuss this with. Whenever I think about it, I suffer from intense pain."*

The patients were also enquired about the techniques they adopt for pain management. Though there was a strong preference for medicines, the patients also reported sitting quietly in a room, drinking warm water, praying, and lying down with closed eyes as strategies to fight back mild pain. In case of moderate to severe pain, all the patients seek hospital services.

DISCUSSION:

SCD is a chronic, degenerative, and self-incapacitating disease affecting patients and their families intensely and permanently. People in a vulnerable socioeconomic situation are more exposed to the psychosocial effects of the disease, and therefore, these individuals deserve special attention in this context. [16] The present study was carried out in children and adolescents belonging to ethnic, low socio-economic groups of a southern state of India. The purpose of the qualitative phenomenological study was to study the patient experiences of having the genetic disorder and to explore the needs of the patients.

Since sickle cell patients require lifelong care with regular medications and frequent hospitalizations, the disease comes with a lot of expenses, and the pain crisis is often accentuated by the financial burden to the families. [16] The present study found that the provision of subsidized quality care under the sickle cell program run by the tribal hospital in the area has increased the utilization of healthcare services and considerably reduced the financial burden of disease on the families. The newborn screenings have helped adopt early interventions against the disease, which has consequently curbed child deaths under five years due to the disease complications. The adoption of evidence-based measures against the disease by the tribal hospital such as early identification of SCD by neonatal screening, the prompt establishment of preventive measures with prophylactic penicillin, immunizations, and therapeutic interventions, such as transfusions and hydroxyurea [17] have greatly improved the outcomes for SCD patients in the Southern state of India.

The study revealed that pain associated with SCD and other complications hurt the physical, social, emotional, psychological, and spiritual domains of patients and their caregivers. The pain was found to be the most distressing aspect among the implications of SCD for the patients and was the major reason for hospitalizations in the study. This also led to feelings of depression and hopelessness, which further complicated the management of the disease. This was found to be in congruence with findings of various studies [18], [19] where the pain was the major reason for life disruption for the SCD patients. The patients also complained of lack of hunger, insomnia, fatigue, functional limitations, emotional effects, and anxiety associated with both the disorder. This was similar to what was noted by researchers in other studies. [20], [21], [22]

The major finding of the study was problems of social adjustment in children and adolescents with SCD. The illness-related stress had a serious impact on an individual's relationships, schooling, aspirations regarding employment and marriage. In this context, the study revealed that few respondents had dropped out of school due to frequent hospitalizations, which has been a reason for constant stress for them. Similar findings have been asserted in the literature that many children with SCD are underachievers in school [23] due to regular school absenteeism [24] and this results in school dropout which hurts the ability to cope with SCD. Stigma was expressed by patients in form of disgrace, the burden to the family, self-pity, and being different from their peers. Reifsnider et al. reported similar effects of stigma related to SCD. [25] Lewis et al found that individuals who have chronic illnesses such as SCD are three to four times more likely to develop various mental health disorders. [26] Noll et al and Palermo et al asserted that SCD patients are at risk of maladjustment to life in several functional areas including emotional and behavioral problems, poor self-concept and interpersonal functioning, and limited physical abilities. [27], [28] It has also been found that SCD patients with severe anxiety report a significantly higher proportion of vaso-occlusive crisis [29]. Thus, it is important to tackle this aspect of the disease for better outcomes and quality of life for the patients.

The study observed that these repeated interruptions in life coupled with depression affected pain coping skills and self-care management by the patients. The pain coping strategies adopted by the patients were mainly pharmacological and there was a serious deficit of self-care management due to hopelessness and negative assumptions of never being able to lead a normal life. It was consistent with the findings of various researches [22], [30]. It was noted that the majority of patients were preoccupied with their illness and had poor quality of life despite the availability of quality healthcare services.

It is also known that health education and counseling about the disease to patients as well as their families is of critical importance. In the study, it was found that all the children and adolescents had incomplete information about the disease as well as the precautionary measures they must take. They also lacked the motivation to identify the triggering factors for crisis and pain as well as to adapt the precautions in their daily routine mainly due to incomplete awareness about the disease. Previous qualitative researches have identified that children with SCD desire open and honest communication from parents about their condition [31]. Honest communication that

provides children with realistic expectations about their future is a factor that may help them cope with the uncertainty of the disease and promote quality of life in children with SCD.

RECOMMENDATIONS:

It is recommended that psychological interventions should be incorporated into protocols for the management of patients with SCD and offered as standard care to improve their quality of life. Psychoeducational interventions primarily focus on improving the knowledge and understanding of the patients regarding the illness, and at the same time providing psychological support for better coping with the condition. SCD peer groups can the formed at the village level for constant support and to identify issues and concerns of the patients. Refresher training should be conducted to improve the counseling skills of the medical staff (nurses and doctors) and the school staff as they are the most frequent contacts with the patients as well as their families.

Cognitive-behavioral therapy techniques help patients manage the problems through positive change in their beliefs, attitudes, and behaviors and must be adopted to assist the patients to deal with negative emotions and conflicting thoughts. Various breathing exercises like the 4-7-8 breathing technique increases the lung capacity as well as help lower the stress levels during episodes of moderate pain. The use of non-pharmacological interventions like massage, deep heat, distraction techniques along with psychosocial support can boost the patients to tackle the complicated disease by modifying dysfunctional thoughts. Sickle cell patients are averse to overexertion and often are not able to do traditional jobs. So they should be provided vocational training related to jobs that do not require physical exertion for long hours. This will help create a strong social identity for the patients, alleviate the feeling of being a burden on the family, provide a purpose of living to the patients as well as empower them to live independently.

CONCLUSION:

SCD should be looked at through a bio-psychosocial lens as this emphasizes how biomedical aspects of the disease interact with family systems, socio-economic factors, and psychosocial aspects to produce guilt, shame, anxiety, and feeling of hopelessness that impede effective coping. The treatment of the disease should not be viewed narrowly as equivalent to sick care and there should be an equilibrium between the needs of the patients and the interventions for the

disease. Effective interventions must be designed to encompass all the factors that are run parallel to the disease and improve the quality of life of the patients.

CONFLICT OF INTEREST:

The author has no conflicts of interest associated with the material presented in this paper.

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