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Sickle Cell Disease - A Case Report

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Abstract

Sickle cell disease (SCD) is a major healthcare and societal problem affecting millions of people worldwide. In the United States of America, it is the most common genetic disorder affecting more than 80,000 people/year; majority of which are the African Americans Arabian and Indian. It is a genetic blood disorder affecting the red blood cells. Sickle cell pain is the hallmark of SCD and is associated with a very high mortality and morbidity rates. Being a genetic abnormality, the complete eradication of the disease from the affected seems to be difficult. Genetic counselling during pregnancy being the prime preventive step, hematopoietic stem cell transplantation becomes the mainstay of treatment for complete eradication of the disease. However, it is not done very often because of the significant risks involved.

Keywords: Sickle cell disease, Sickle cell pain, Acute chest syndrome, Hemolytic anemia

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Introduction

Disasters interrupt progress, destroy the developmental outcomes and delay the growth leading to long-term effects. Disasters acquired or inborn creates more or less the same impact on human lives and the society around it. In this 4G world, there has been a paradigm shift of the word "DISASTER" from being restricted to outside human body to inside of it. Statistics depicting the increasing number of disasters as diseases inside the body, either due to easy detection by advanced medical technology or due to vigorous life style changes; poses a big question and needs further analysis.

The human body organization is so well formatted that the approximate 5 l of blood which is regularly pumped by the 300 g organ (heart) every second, continuously is able to reach every nook and hook of our body without fail. But at times, when there is an interruption to this flow of the blood; it leads to massive hemolytic disasters in the human body. One such inherited hemolytic disaster in the body is sickle cell anemia.[1-3]

Background

Sickle cell disease (SCD) was first described in the Nilgiri Hills of northern Tamil Nadu in 1952. The sickle cell gene is now widespread among people of the Deccan plateau of central India, north of Kerala and Tamil Nadu.[4] The coincidence of large tribal populations with the "sickle cell belt" of Central India and northern Kerala and Tamil Nadu has given rise to the assumption that tribal people are more prone to the HbS gene although this disease is widely distributed among tribal and non-tribal population.[8]

Incidence

SCD affects 20–25 million people globally and 50–80% of the infants with SCD in Africa die before they attain 5 years of age. It is estimated that around 240,000 children are born with SCD annually in the Sub-Saharan Africa.[7,8] In 2009, the United Nations general assembly recognized SCD as a global public health concern. Around 3/4th of the entire global sickle cell population belongs to Africa while only 10% of the SCD population belongs to the developing nations including India.[5,6]

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In India, SCD is mostly prevalent in the Central and Western parts of the country which has a concentrated tribal population of the country. Estimates state that there are about 5200 live births/year.[8,9] According to the Indian Council of Medical Research, around 20% children with SCD die before 2 years and 30% of them die before they reach adulthood.[3] Central India report severe disease (defined as >3 bone pain crises, >3 transfusions/year) in 30% children.[1]

India bears the largest tribal population globally and Odisha is one of the prominent states to harbor a huge percentage of tribes as well as the sickle cell burden in it. The effort to retain the culture and the mere existence of the tribe leads to the increased prevalence of marriages within the tribe which accounts for the increasing numbers of SCD population in it.[2] However, in India, the disease is largely under-documented.

Patterns of Disease and Inheritance

SCD is used to denote a group of genetic disorders in which there is a substitution of the amino acid glutamine by valine at position of 5 of the β- globin chain in the hemoglobin variant HbS. This mutation, under low oxygen concentrations causes a change in the shape of red blood cells (RBC), leading to the deformity and acquiring the shape of a sickle. Possession of a single HbS gene results in the generally harmless sickle cell trait (AS genotype) but inheritance of the HbS gene from both parents results in homozygous sickle cell (SS) disease that is often a severe condition destroying RBC rapidly and blocking flow in blood vessels with painful, serious complications.

There are several forms of sickle cells; sickle cell anemia, homozygous and more severe forms, HbS hemoglobinopathy and sickle cell thalassemia.

Clinical Manifestations

The clinical manifestations are varied and differ from every individual. Some of the common ones are as follows:

Vaso-occlusion	*Super-added infections*	*Jaundice*
Pulmonary infection	Anemia	Splenic sequestration
Musculoskeletal pain/damage	Heart disease	Stroke
Priapism in males	Renal failure, Leg ulcer	Retinopathy

Treatment

The treatment of a child with SCD is very individualized in nature. It depends on the age, the overall general health, and the severity of the disease being manifested by the individual child.[11]

Treatment modality may include the following as follows:

1. Analgesics used for pain crises.
2. Drinking plenty of water, daily 8–10 glasses. This helps prevent and treat pain crises. In some cases, intravenous (IV) fluids may be needed.
3. Blood transfusions to treat anemia, chronic pain, acute chest syndrome, and splenic sequestration and to prevent stroke.
4. Vaccines and antibiotics to prevent infections especially pulmonary which can make the child further immune-compromised.
5. Folic acid helps prevent severe anemia and prevent regular bouts of sickle cell crisis attacks.
6. Regular eye examinations to screen for retinopathy.
7. Hematopoietic stem cell transplantation. This is the "Gold standard" treatment for patients with SCD. However, the cost factor and the feasibility of doing it has been a great issue of concern.
8. Hydroxyurea to reduce the number of sickle cells in the blood. It reduces complications, painful episodes (crises), and hospital stays.

Prognosis

This disease being genetic in origin makes a permanent imprint in one's life. The chronicity of this disease affects their quality of life thus, lowering down their coping abilities. Therefore, it is essential to have adjustment process, counselling sessions to boost them up to accept this disease and to enhance their standards of living to have better and cheerful living experiences.[10-12]

Case Report

A 5-year-old boy, Master R, came to our (secondary level) hospital with the complaints of high-grade fever and cough since 2 days. On admission, his body temperature recorded 101.4°F, pulse rate- 98/mt., respiratory rate – 32/mt. and SPO_2-90%. He had chest retractions indicating signs of respiratory distress. As per his mother (primary care giver), her pregnancy and puerperal period was uneventful. The child had a normal neonatal period and had normal growth pattern. The child's parents had a non-consanguineous marriage.

On assessment, Master R. looked pale and had hepatosplenomegaly. On auscultation, adventitious breath sounds (rales) could be heard in bilateral lung fields and he looked icteric too. His blood investigations revealed the following:

- Hemoglobin- 6.8 g/dL
- C-reactive protein- 1.2 mg/dL.

Sickling Test-positive

Liver function tests- SGOT- 137 IU/mL and SGPT- 78 IU/mL.

He was further evaluated with hemoglobin electrophoresis which revealed the following:

HbS- 76.8% and HbF- 16.3%. This confirmed that the child has homozygous sickle cell anemia (HbSS). His chest X – ray showed bilateral consolidation in both the lungs. He was diagnosed as sickle cell anemia with splenic sequestration associated with acute chest syndrome. He was found negative for widal test and malaria.

Management of Master R

During his hospital stay, he was transfused three units of blood (125 ml each) in alternate days for 6 days. He was started on penicillin antibiotics (Injection Piperacillin/Tazobactam, a beta – lactamase inhibitor) to combat lung infection caused due to immune deficiency secondary to vaso-occlusive crisis. On the 2nd day after blood transfusion, he had one episode of fever (Temp. –101°F) which was managed by administering syr. Paracetamol 125 mg. He had a repeat chest X- ray on the 5th day of hospitalization which revealed marked reduction in the consolidation in the lungs. His vital signs recorded a body temperature of 98.8°F, Pulse rate- 86/mt. and respiratory rate- 32/mt. His condition improved during the hospitalization and he was discharged after 7 days of treatment in the hospital.

Nursing Management of Master R

"Nursing centers around the practice of caring and if it is effectively demonstrated and practiced, it will promote the development of health and individual and family growth"

On a nursing perspective, it is really challenging to provide comprehensive holistic care to chronic patients such as Master R living with SCD. It entails in itself numerous aspects which can be very well conferred in the ten carative factors based on Watson's theory of human caring. They are enumerated as follows:

1. Forming a humanistic – altruistic value system - Nurses gain rich knowledge on SCD and its management throughout their professional education. This helped the nurses in attending the actual and anticipated problems of Master R and render selfless, holistic care. Knowing about the chronicity of the disease, the nurses were able to pursue Master R as an individual with emotions and as a person who is going to live with SCD for his entire life.
2. Instilling faith and hope - Faith and hope was developed while caring for Master R. by appreciating the parents' efforts amidst of all odds since 5 years.

   The nurses notified the child's condition to the parents regularly, clarified all their doubts regarding treatment, diagnostic findings in an easy and understandable language. This developed good rapport between the nurses and the immediate caregivers which further helped in enhancing their trust. The patient being Christian by faith was encouraged to attend Morning Prayer in the hospital prayer hall along with his mother that may enrich the sense of well being by strengthening their belief.
3. Cultivating sensitivity to oneself and others- The nurses being aware of the chronicity of the disease, explained the mother about the nature of the disease in simple and understandable language. SCD being difficult to eradicate; the mother was advised about the measures to keep the crisis pain at bay. She

was advised on the regular administration of folic acid tablet 5 mg daily to her son, increased intake of water, avoidance of extremes of temperature and infection preventive measures. The nurses ensured open communication with the mother which allowed in building up a positive interpersonal relationship between the duos.

4. Developing helping, trusting, caring relationships - Master R' s mother was quite apprehensive regarding the care modality for her son. She belonging to a mediocore family was quite worried regarding the expenses and the quality of life of her son in relation to the disease. The nurses being the frontline caregivers of Master R., considering the financial state of the family, informed his mother about the availability of "free health services" by the government of Odisha for sickle cell patients. The nurses encouraged his mother to avail a "health card" for her son from nearby Government hospital which will enable her to obtain blood units from the blood banks free of cost in times of need for her son in future. All these enhanced the trusting relationship between the patient, family and the nurses providing care for Master R.
5. Promoting expression of positive and negative feelings - Nurses patiently and actively listened to the grievances of Master R's mother regarding manpower, time and finances. She being the sole caregiver for her son as her husband is working out-of-state was a big reason of concern. The nurses provided positive reinforcement for the efforts of mother and encouraged alternatives as per the available resources for caring of Master R. A non- judgemental approach was carried out throughout his hospitalization.

   The nurses developed good rapport with Master R. and his family members. They counselled them about the genetic origin of the disease and explained about the prognosis of the disease (that was explained to them already by the physician) in an easy and understandable manner.
6. Systematically using the scientific problem solving method for decision making - The nurses administered the prescribed antibiotics which helped in combating the lung infection of Master R. Furthermore, transfused 3 units of blood to Master R. during his hospitalization to increase his hemoglobin level which was decreased due to hemolysis. The nurses advised the mother to avoid the child to be involved in strenuous activity or any extremes of temperature as this would further aggravate the disease process.
7. Promoting interpersonal teaching - learning - The nurse taught Master R's mother about the clinical manifestations and the readily available health care services for patients living with SCD. Nurses reinforced repeatedly to Master R. And his mother the significance of more intake of fluid and the intake of folic acid and zinc supplements regularly to prevent further sickle cell crisis pain attacks. The nurses counselled the mother and motivated her to give Pneumococcal vaccine to Master R. to prevent pulmonary infections and to improve the quality of life.
8. Providing a supportive, protective or corrective mental, physical socio-cultural and spiritual environment-
   a. Master R. was nursed in fowler's position, administered asthalin nebulization q6 hourly to relieve bronchospasm. His mother was encouraged to maintain adequate hydration and good personal hygiene for Master R. to combat infection.
   b. Master R. being 5 years old was provided a bed with side rails to prevent the risk of injuries and falls. His peripheral IV line was assessed regularly to rule out thrombophlebitis proving to be the nidus of infection. Blood tests were done regularly to rule out the grade of hemolysis and its compensation and transfused units of RBC.
   c. Master R. being Christian by faith, the nurses intimated the chaplain of the hospital who visited him & his mother and provided spiritual support.
   d. Positive reinforcements were given to the mother for all the efforts she took to care for Master R. Figure 1 shows Jean Watson's ten carative factors and Table 1 depicts early complications in sickle cell disease.
9. Assisting with the gratification of human needs-
   a. Lower order biophysical needs- Master R. was administered nebulization q6h to reduce his pulmonary

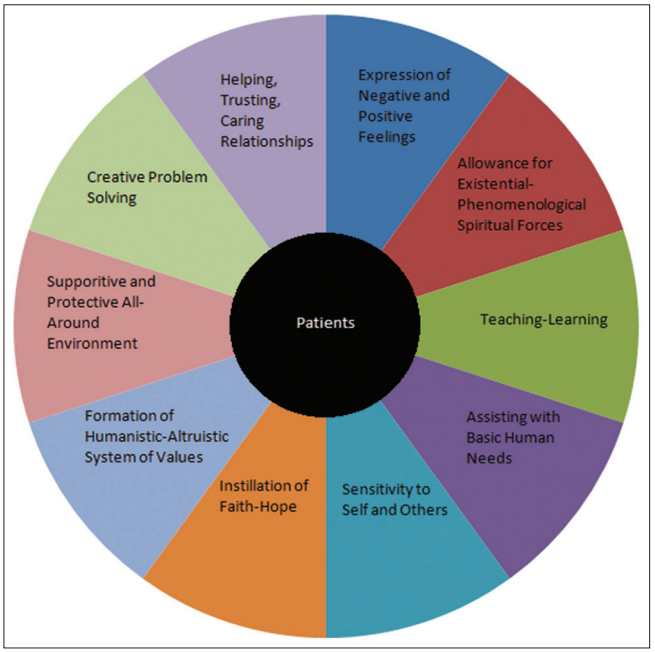

Figure 1: Jean Watson's ten carative factors

Table 1 : Early complications in sickle cell (SS) disease of Indian origin[2]

Complication	*Intervention*	*Cost*	*Frequency (%)*	*Observations*
Pneumococcal septicemia	Pneumococcal prophylaxis with penicillin for 4 years, conjugate and regular vaccine.	High	Unknown	Not described
Acute splenic sequestration	Teaching parents to check spleen size at home	Low	Almost 10	Events occur, but natural history and optimal treatment is unknown
Chronic hypersplenism	Chronic transfusion or splenectomy	Moderate	Almost 5	Hyperplenism occurs and optimal management is needed
Stroke	Transfusion to prevent stroke, Trans-cranial Doppler (TCD)	Very high	Unknown	Stroke occurs, but natural history is unknown and TCD may not be justified

congestion and spasm. He was advised to take high protein and iron rich diet to boost his immunity, faster recovery and combat hemolysis. All prescribed medicines were administered to Master R. on time.

b. Higher order psychosocial needs- Master R's mother was counselled about the chronic nature of SCD. She acknowledged that it is a lifetime disease and her son has to live with it. She realized the significance of regular intake of folic acid and zinc supplements to prevent sickle cell crisis attacks. She also verbalized the importance of regular follow-up and frequent need for blood transfusions for her son.

10. Allowing for existential - phenomenological forces - The nurses while rendering care to Master R. were liberal enough to absorb the mixed emotions projected out by Master R's mother throughout the hospital stay. The nurses could comprehend and appreciate the severity of the disease and its management, the immense stress and burnout experienced by the immediate caregivers. Nurses gained rich knowledge and experience through care of Master R. A calm manner approach was practiced to answer and clarify all the doubts of Master R's mother. His mother, the primary caregiver was appreciated now and then for every effort taken to alleviate the pain and suffering of Master R. and progress toward optimum health.

Conclusion

SCD is an emerging public health concern. It being inherited disorder, its growth can be regulated by regular advanced antenatal counselling and screening. However, this seems to be costly and is being a problem that only could be seen as the tip of ice berg. However, the real challenge remains in what lies beneath!! There is a huge population both including adult and children living with SCD in our country and each of them have their own story of pain and suffering to narrate. As health care professionals, we must try to be sensitive to their needs, reach out to them and take every possible measure to make their lives a life with dignity and happiness.

References

1. Balgir RS, Sharma SK. Distribution of sickle cell haemoglobin in India. Indian J Hematol 1988;6:1-14.
2. Colah R, Mukherjee M, Ghosh K. Sickle cell disease in India. Curr Opin Hematol 2014;21:215-23.
3. Kleigmian K. Nelson Textbook of Pediatrics. 1st ed. Philadelphia, PA: Saunders; 2007. p. 2027-30.
4. Lehmann H, Cutbush M. Sickle-cell trait in southern India. Br Med J 1952;1:404-5.
5. Government of Gujarat. Sickle Cell Anemia Control Project. Ahmedabad: Government of Gujarat; 2012. Available from: https://doi.org//www.sickle-cell: gujarat.gov.in/sc_program.aspx. [Last accessed on 2021 Jan 03].
6. Mulumba LL, Wlson L. Sickle cell disease among children in Africa: An integreative literature review and global recommendations. Int J Afr Nurs Sci 2015;3:56-64.
7. Pedram M, Jaseeb K, Haghi S, Vafaie M. First presentation of sickle cell anemia in a 35 year old girl: case report. Iran Red Crescent Med J 2012;14:184-7.
8. Rao VR. Genetics and Epidemiology of sickle cell anemia in India. Indian J Med Sci 1988;42:218-22.
9. Deepak S, Yasobant S, Golechha M. Situational analysis of sickle cell disease in Gujarat, India. Indian J Community Med 2017;42:218-22.
10. Seeler RA, Shwiaki MZ. Acute Splenic Sequestration crisis in young children with sickle cell anemia. Clinical observation in 20 episodes in 14 children. Clin Pediatr (Phila) 1972;11:701-4.
11. Serjeant GR, Ghosh K, Patel J. Sickle cell disease in India: A perspective. Indian J Med Res 2016;143:21-4.
12. Wethers DL. Sickle cell disease in children: Part II. Diagnosis and treatment of major complications and recent advances in treatment. Am Fam Pysicians 2000;62:1309-14.