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SICKLE CELL DISEASE- AN AYURVEDIC PERSPECTIVE

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ABSTRACT

The term sickle cell disease (SCD) describes a group of inherited red blood cell disorders. Sickle Cell Anemia results from an abnormal haemoglobin-S or sickle-cell haemoglobin. The molecular lesion in Hb-S is the substitution of valine for glutamic acid at the sixth residue of the beta chain(Hb-S6^{GLU-VA}).Sickle gene is found all over the world, particularly amongst people originated/migrated from Malaria endemic areas(tropical-sub tropical) of Africa & Asia. About 5,200 live births of Sickle Cell disease (SCD) each year is a major public health problem in India. Although SCD has been described in India in numerous ethnic groups, it is most prevalent. In Scheduled tribe population the prevalence of this disease is very high. Approximately 18 crores of the population of India comprises of the tribal community and among them about 1.8 crores and 14 lakh are expected to be sickle cell trait and sickle cell disease respectively. Complete cure of this disease is currently not available and the patients have to suffer from various complications, hence there is a need to design an Ayurvedic Protocol for the treatment/management of Sickle Cell Disease.

Key words: Hb.S, Pandu, R.B.C, S.C.D., Shodhan

INTRODUCTION

Sickle cell disease or Sickle cell anemia is a genetic disease seen amongst various tribal populations of India that affects the hemoglobin. Sickle cell anemia is the homozygous state for the Hb-S gene (Hb-SS). It is transmitted as an autosomal recessive disease.[1]According to a hypothesis, it is occurred due to natural mutation in Hemoglobin molecule to protect RBCs from malarial parasites by making them a little rigid, hence, preventing its entry and growth. Consequently Sickle Cell gene is mainly present amongst tribal group only, who originated from tropical regions or malaria prone regions. In this disease the shape of the red blood cells changes like a sickle and becomes elongated thereby sticking and occluding the micro-vessels leading to infarction.

Disease Review

SCA occurs due to inherited abnormal hemoglobin (Hb) gene, which produce Hb-S

(Hb-Sickle). The 20th nucleotide of the gene for the beta chain of hemoglobin is altered from the codon GAG to GTG. Thus, the 6th amino acid glutamic acid is substituted by valine—notated as an "E6V" mutation—and the protein is sufficiently altered to cause the sickle-cell disease.[2] This in turn alters one of the amino acids in the hemoglobin protein. The valine makes the hemoglobin molecules sticky; forming long fibers that alters the shape of the red blood cells, and this brings on an attack.

The diagnosis of SCA is only possible by carrying out a simple special blood test known as Sickling test on RBCs and further testing by either Hb electrophoresis or by HPLC technique to find out whether the person is heterozygous or homozygous.

Due to the presence of Hb-S and because of its abnormal characteristic, normal RBCs is converted into rigid-brittle Sickle shaped instead of soft round biconcave shape, which is the main cause of complication of Sickle Cell disease. The rigidity and abnormal shape reduce their ability to be propelled through tiny capillaries leading to the formation of entangled masses of cells in blood vessels. This obstructs the blood flow into organs, producing temporary or permanent organ dysfunction. Because of their abnormal shape, the spleen in the body destroys these RBCs, causing enlargement of spleen. Life span of RBC in SCD is less than 30 days instead of 90 to 120 days. Anemia results from the bone marrow's inability to produce enough blood cells to keep pace with the rate of destruction.

Table 1: Difference between Normal RBC & HbS.

Sr. No.	Normal Blood Cell	Sickle Cell
1	Doughnut shape, round	Sickle shape, crescent shape
2	Flexible	Stiff
3	Smooth	Sticky
4	Doesn't stick in the blood vessels	Stick in the blood vessels
5	90-120 days life span	15-20 days

SCA represents itself with the following common signs and symptoms

- Pallor
- Bone & Body ache
- Frequent jaundice
- Enlarged Spleen,
- Retarded Growth
- Frequent Infections
- Dactylitis (Hand-foot Syndrome)

EPIDEMIOLOGY

Sickle-cell disease occurs more commonly among people whose ancestors lived in tropical and sub-tropical sub-Saharan regions where malaria is common, carrying a single sickle-cell allele (trait) confers a selective advantage—in other words, being a heterozygote is advantageous.[3]

The highest frequency of Sickle Cell Disease is found in tropical regions, particularly sub-Saharan Africa, India and the Middle-East.[2] Migration of substantial populations from these high prevalence areas to low prevalence countries in Europe has dramatically increased in recent decades and in some European countries sickle cell disease has now overtaken more familiar genetic conditions such as hemophilia and cystic fibrosis.

Three quarters of total Sickle Cell Disease cases of the world occur in Africa. In several sections of Africa, the prevalence of Sickle Cell Trait (heterozygous) is as high as 30%. Although the SCD is most frequently found in Sub-Saharan Africa, it is also found in some parts of Sicily, Greece, Southern Turkey, all of which have areas in which malaria is endemic.

Table 2: Prevalence of Sickle Cell gene (%) in Some States of India [5]

Sr.No.	State	Prevalence	Sr.No.	State	Prevalence
1	Madhya Pradesh	0-48.5	6	Gujarat	0-30.0
2	Maharashtra	0-45.4	7	Kerala	0-29.7
3	Tamil Nadu	0-35.3	8	Karnataka	0-25.0
4	Andhra Pradesh	0-34.6	9	Orissa	0-12.4
5	Uttar Pradesh	0-32.6	10	West Bengal	0-01.1

Major Complications of Sickle Cell Disease

- Infections
- Pulmonary hypertension (increased pressure in the arteries of the lungs)
- Stroke
- Anemia
- Kidney problems
- Priapism (prolonged and painful erections)
- Liver problems
- Gallbladder disease
- Spleen damage
- Bone and joint problems
- Leg sores and ulcers
- Eye damage in the retina
- Acute Pain (sickle cell or vaso-occlusive) Crisis
- Chronic Pain
- Severe Anemia

Line of Treatment

General line of treatment includes

- Treating Anemia
- Preventing Infection
- Treating and Preventing Vaso occlusive crisis
- Management of complications
- Hematopoietic stem cell Transplantation

But the above treatment may lead to complications like Hemolysis, Iron overload, infection.

DISCUSSION

- **Ayurvedic Perspective**

The symptoms of Sickle cell anemia mainly resemble to that of the Pandu roga mentioned in Ayurvedic classics.

स पाण्डुरोग इत्युक्तः तस्य लिङ्गं भविष्यतः
हृदयस्पन्दनं रौक्ष्यं स्वेदाभावः श्रमस्तथा १२

संभूतेऽस्मिन् भवेत्सर्वः कर्णक्ष्वेडी हतानलः
दुर्बलः सदनोऽन्नद्विट् श्रमध्वमनिपीडितः १३

गात्रशूलज्वरश्वासगौरवारुचिमान्नरः
मृदितैरिव गात्रैश्च पीडितोन्मथितैरिव १४

शूनाक्षिकूटो हरितः शीर्णलोमा हतप्रभः
कोपनः शिशिरद्वेषी निद्रालुः क्षीवनोऽल्पवाक् १५

पिण्डिकोद्वेष्टकटचूरुपादस्फुरणसादनानि च
भवन्त्यारोहणायासैर्विशेषश्चास्य वक्ष्यते १६ [6]

But the *nidan* of *Pandu roga* doesn't correlate at all to that of the Sickle cell Anemia. Sickle cell anemia as we have known is a hereditary disease. Hence we have to find its origin in *Bija dusti roga*. But the *samprapti* of sickle cell disease (vaso occlusion) can be correlated to that of *mrutika bhakshanaj pandu* as in both the type of *storto-dusti* is *sanga*. Hence the management of vaso occlusion found in sickle cell disease can be treated by following the treatment of *mrutika bhakshanaj pandu*. Also in the *samanya chikitsa* of *pandu*, acharya charak mentions

तत्र पाण्डवामयी स्निग्धस्तीक्ष्णैरूर्ध्वानुलोमिकैः
संशोध्यो मृदुभिस्तिक्तैः कामली तु विरेचनैः ४०

[7]

Here importance is given to shodhan treatment.

कुर्याच्छोणितरोगेषु रक्तपित्तहरीं क्रियाम्
विरेकमुपवासं च स्रावणं शोणितस्य च १८

[8]

Also the importance of *virechana* is mentioned by Acharya Charak as above.

Hence *shodhan* treatment can help in the management of above condition.

निपातयेच्छरीरात्तु मृत्तिकां भक्षितां भिषक् ११७

युक्तिज्ञः शोधनैस्तीक्ष्णैः प्रसमीक्ष्य बलाबलम्
शुद्धकायस्य सर्पींषि बलाधानानि योजयेत् ११८

[9]

Apart from this treatment spleen is destroyed due to occlusion as well as spleenomegaly occurs as it has to carry out the process of haemolysis. spleen contains half a million immune cells, so when spleen gets destroyed immunity is lowered, hence prophylactic antibiotic are given, hence *rasayan chikitsa* as well as hepato-spleeno protective drugs like *sharpunkh*, *rohitaka, kumara, punanarva* should be thought upon and management of the complications should be done accordingly.

Rakta basti can be given in *rakta kshaya*.

तदेव दर्भमृदितं रक्तं बस्तिं प्रदापयेत्
श्यामाकाश्मर्यबदरीदूर्वोशीरैः शृतं पयः

[10]

Hence, management of Sickle cell disease can be done by planning the above treatment. There is a need to prepare such a Protocol of Ayurvedic treatment in managing SCD. And if research work is done on such patients, then the life style of such patients will definitely improve.

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