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-S6GLU-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
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—noted 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.

### Table 1: Difference between Normal RBC & HbS.

<table>
<thead>
<tr>
<th>Sr. No.</th>
<th>Normal Blood Cell</th>
<th>Sickle Cell</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Doughnut shape, round</td>
<td>Sickle shape, crescent shape</td>
</tr>
<tr>
<td>2</td>
<td>Flexible</td>
<td>Stiff</td>
</tr>
<tr>
<td>3</td>
<td>Smooth</td>
<td>Sticky</td>
</tr>
<tr>
<td>4</td>
<td>Doesn’t stick in the blood vessels</td>
<td>Stick in the blood vessels</td>
</tr>
<tr>
<td>5</td>
<td>90-120 days life span</td>
<td>15-20 days</td>
</tr>
</tbody>
</table>

**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]

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

### 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.
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

Also the importance of virechana is mentioned by Acharya Charak as above. Hence shodhan treatment can help in the management of above condition.

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 hepatoprotective 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.
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 lifestyle of such patients will definitely improve.

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