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Sickle Cell Anemia in rural areas of Nandurbar District

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ABSTRACT:Sickle cell is a genetic or hereditary disorder, in which red blood cells (R.B.C) becomes sickle (oval) shape due to the deficiency of oxygen in blood. Sickle cell is of two different types i.e., carrier or patient it shows symptoms such as body ache, body pain, stroke, weakness, fatigue, infections, etc. the study suggested, that in the tribal populations or in the tribal areas like Nandurbar which is also known as a "tribal district" or "Adiwasi district" of Maharashtra state. There are more patients and carriers of the sickle cell anemia and sickle cell diseases as compare to any other districts of Maharashtra state. Because sickle cell is a major genetic disorder. During present study, the total number of sickle cell patients in Nandurbar district including its Talukas (Nandurbar, Taloda, Akkalkuwa, Dhadgaon, Shahada and Navapur) was counted from April 2009 to March 2015, the total number of carriers was found to be 1.95% while patients were found to be 0.18% of the total population.

KEYWORDS: Sickle cell anemia; sickle cell trait; Nandurbar district; Maharashtra state.

I. INTRODUCTION

[9, 10, 11, 17, 42, 56] Sickle cell anemia is a type of blood disorder in which normal red blood cells (RBC's) leads to a rigid sickle-like shape. [66, 67] It is a genetic disorder which is inherited from parents to offspring. There are two types in sickle cell anemia. The one that are actually patients of sickle cell disease and other are the carriers which carry the diseased state in a silent way also known as "silent disease" in which disease can cause to offspring but does not to a carrier (who carries it). [2, 3, 4, 8] It occurs when person inherits two abnormal copies of the haemoglobin gene from each parent the primary pathology is based on a polymerization of deoxy-HBS with formation of long fibres within the RBC's causing a distorted sickle shape which eventually leads to increased haemolysis and vaso-occular of sickle red cells. [15, 19, 21] The most common type of sickle cell disorder (SCD) is known as sickle cell anemia (SCA) it has an abnormality in the oxygen carrying protein haemoglobin found in red blood cells (RBC's) problems in sickle cell disease typically begin around 5 to 6 months of age, including anemia, swelling in the hands and feet's, bacterial infections, stroke, chest pain and fatigue long term pain may develop as people gets older. [3, 26, 27, 35] The average life expectancy in the sickle cell patients is between 40-60 years in this developed world. [3, 40, 41, 50] Sickle cell attack can be set off by temperature changes, excessive stress, high altitude as well as dehydration also. [6, 14, 51] Diagnosis of the sickle cell disease is by blood test and in many countries the test is carried at the birth of the baby. [3, 23, 43] Diagnosis may also possible during the pregnancy period.

[3, 45, 50] The care of the people with sickle cell anemia may include the prevention of infection with vaccination as well as with antibiotics, [3, 12, 13, 18, 38, 48] it may also include high fluid intake, by taking the folic acid supplementation and also by taking some pain medications other measures may include transfusion of blood or by a transplant of bone marrow cells. [17, 50, 53, 57] Signs and symptoms for the sickle cell anemia usually begin from early childhood its severity may vary from the person to person. [3] It may lead to various acute and chronic complications, several of which have a mortality rate also. In Maharashtra the sickle cell gene is widespread in all the eastern districts also known as khandesh in the satpuda ranges in the north and in some parts of Vidarbha and Marathwada. [3, 30, 37, 69] The tribal groups with a high prevalence of the disease are includes, Adivasi's, Bhils, Madias, Pawaras, Pardhans, and the Otkars. It has found that Shahada, Navapur, Akkalkuwa, Dhadgaon, Taloda including Nandurbar district has more than 3000 cases of sickle cell anemia. In this Dhadgaon and Nandurbar are on the top position with 0.36% and 0.27% of the disease and 3.65% and 1.30% of the carriers of the disease. Sickle cell diseases in

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which especially sickle cell anemia (SCA) can be manage by the folic acid and penicillin, malaria prevention Hydroxyurea administration, blood transfusion, bone marrow transplant, psychological therapies, etc. [3, 16, 45, 50, 56, 58] Recent research has found that umbilical cord blood transplant can potentially cure the condition, and also gene therapy started in 2014, when a sickle cell Mice was successfully treated using gene therapy.

II. TRIBALS OF NANDURBAR

[3, 30, 34, 49, 52] Globally after Africa India is the country which has the largest concentration of tribal population. [30, 37, 60] Tribal's are basically considered as the early settlers in the country and are considered to be the original inhabitants. According to census of India 2011, the tribal population of Nandurbar is 65.53%. [37] The states of Madhya Pradesh, Maharashtra, Odisha, Gujarat, Rajasthan, Jharkhand, Chhattisgarh, Andhra Pradesh, west Bengal and Karnataka according for around 83% of the total schedule tribe population in the country. [3, 7] In Nandurbar major Adiwasi tribes are concentrated in the forests of satpuda mountain ranges and Basin of Narmada River. These contains mainly Bhils one of the dominant tribes in India whose original home is in the hilly country between Abu and Asirgarh from which they mostly spread over in the states of Madhya Pradesh, Rajasthan, Gujarat and Maharashtra. Bhils are mostly found in Satpuda Mountain Range. In the year 1871 The British Government declared them as a "Criminal Tribe". According to census of 2001, total population of Bhils in Nandurbar district is 7, 14,127. There is a myth among the Bhils of satpuda ranges that, during the stone age Beni-Hejah and Raja Pantha tied chickens to their legs and danced in the gav diwali festival because, in those days "ghungrus" (tiny bells) were yet to be invented therefore, it can broadly be said that the history of bills dates back to the Stone Age. [3] In Nandurbar district the major population of Adiwasi consists of Vasave, Vasava, Valvi, Padvi, Tadvi, Naik, Gavit and Gamit. These are the dominant clans of Bhils and not their sub-tribes. "PawaraBhils" or "Pawaras" mostly reside in the Satpuda Ranges and especially in the belt between the Tapi and Narmada Rivers. Their major population in Nandurbar district is concentrated in Akrani mahal (Dhadgaon), Shahada and some hilly parts of Taloda taluka. The kokani tribe has migrated from Konkan region of Maharashtra during the famine of 1396-1408. It is because of this they got the name Kokani are found in the Nasik, Thane, Dhule, and Nandurbar district. The Adiwasi have their customary personal laws which includes law relating to marriage (Betrothal, means of acquiring mat, forms of marriage and preferential mating), automatic dissolution of marriage, remarriage. [3] However, Reich et al concluded that "Several thousand years ago, the entire subcontinent underwent a period of massive intermarriage, shuffling its population's genetic deck. So thoroughly that it left dear traces even in the genomes of today's most isolated Tribes".

III. PREVALENCE OF SICKLE GENE IN TRIBAL COMMUNITIES IN MAHARASHTRA

[1, 3, 29, 30, 31, 37] In Maharashtra, the sickle gene is widespread in all the north western districts in Satpuda ranges and in the eastern districts also known as Vidarbha also it is slightly active in some part of Marathwada. [60, 61, 64, 68] The prevalence of sickle cell carriers in different tribes varies from 0-35 %. The tribal group with a high prevalence of HBS (20-35%) includes the Adiwasi, Bhil, Otkars, Pawara, Pardhans, Naik, Madia, Valvi, Vasave and Vasava. [6] It has also been estimated that districts like Nandurbar, Bhandara, Jalgaon, Nagpur, and Chandrapur would have more than 5000 cases of sickle cell anemia (from government survey 2015).

[61] In a large multicentre study, [3] where 15,200 individuals from 14 primitive tribes from Maharashtra, Gujarat, Tamil Nadu and Odisha were screened, the HBS allele frequency varied from 0.011 to 0.120 and β-Thalassemia allele frequency varied from 0.005 to 0.024. [3, 72] Associated with iron deficiency was seen in 26.2 percent of sickle heterozygote's as well as in 67.7 percent of sickle homozygous in this study. [60, 3]Kaur et al, have summarized the distribution of HBS in different tribal groups have been screened for HBS; there are still many gaps in our knowledge about the distribution of the HBS gene in tribal communities in India.

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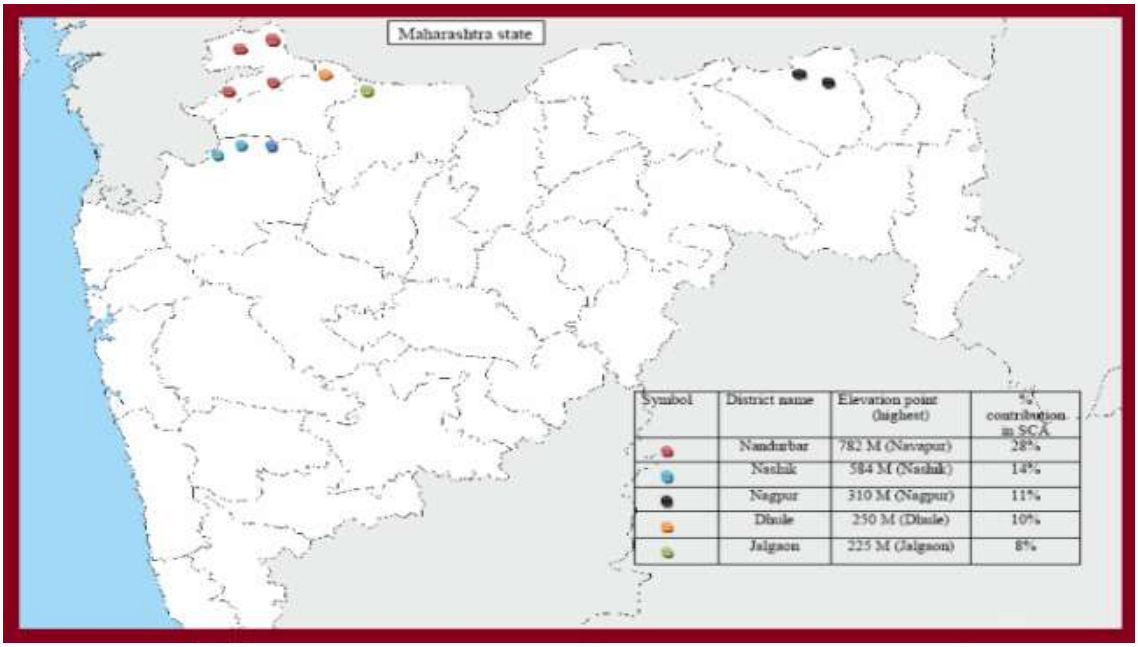
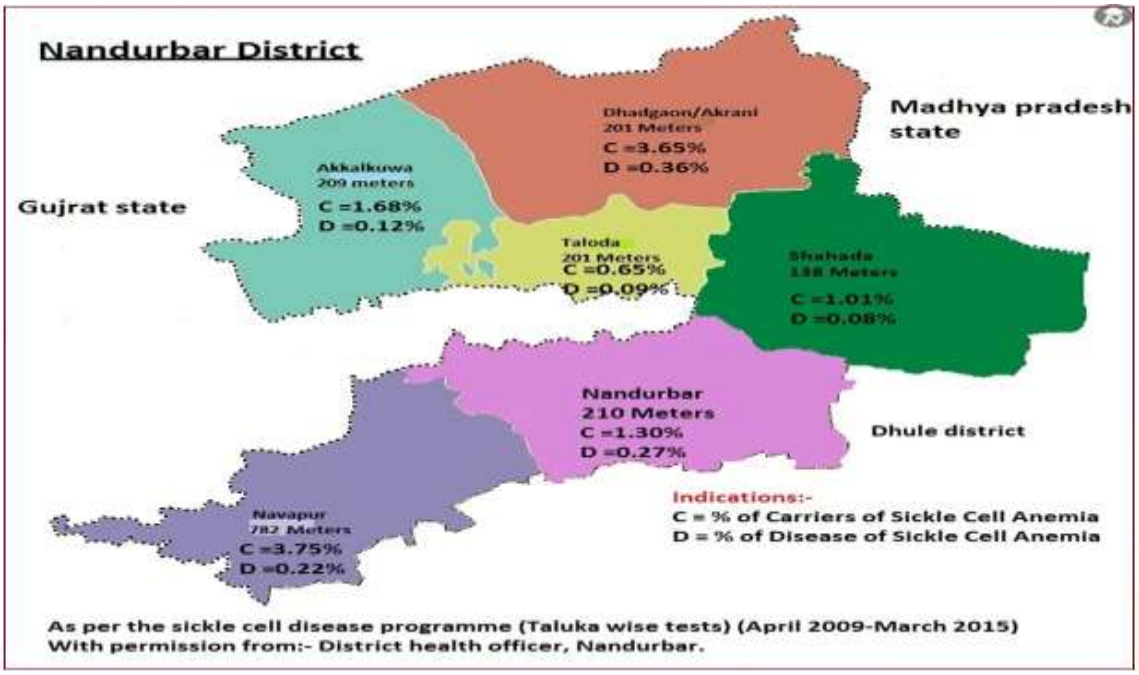
The figure shows the district wise distribution of sickle gene among tribal populations in Maharashtra.



***Elevations are as per government records**

Fig 1: -Maharashtra state with active sickle gene and percent contribution of districts.

Another figure shows different Talukas of Nandurbar and their contribution for sickle cell anemia with their elevation points.



***Elevations are as per government records**

Fig 2: -Nandurbar district with Elevation points and Percentage of carriers and diseased patients of Sickle Cell Anemia.

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IV. TECHNOLOGIES FOR SCREENING IN MAHARASHTRA

[6, 20, 54] Most of the early studies on epidemiology of sickle haemoglobin in different parts of the country use the sickling test or the solubility test and, in many reports, this was followed by the HB Electrophoresis to determine the phenotypes. [28, 47] But in recent years the Sophisticated Analytical Techniques such as High-Performance Liquid Chromatography (HPLC) analysis have been used in many large programmes to identify carriers of both sickle cell haemoglobin as well as β-Thalassemia. [54] Capillary Electrophoresis is a technique by which thin micros pore capillary is used for separation, this technique has also now been introduced in some government centres in Maharashtra such as King Edward Memorial Hospital (KEM Hospital) Mumbai and also in Non-Government Centres such as Apollo Spectra Hospital Mumbai, Tata Memorial Hospital Mumbai, Alexis Hospital For Blood Disorders and Medical College Nagpur, etc. [63] nonetheless even, the simple and cost-effective solubility test has been shown to have a sensitivity and specificity of 97.4 and 100 percent, respectively in comparison to High Performance Liquid Chromatography (HPLC) and could still serve as a good first line screen test for sickle haemoglobin in isolate and remote areas where other facilities are not easily available, like HPLC.

V. NEWBORN SCREENING FOR SICKLE CELL ANEMIA IN NANDURBAR

[5, 20] Newborn screening for sickle cell disorders in world was first offered in the New York programme in 1975 in the United States of America. Shortly after Robert Guthrie introduced dried blood specimen collection on filter paper. [20] Same as the New York Programme in 1974 in USA all the states initiated newborn screening and this has also been undertaken in many other countries where the sickle gene is common. Until today in Nandurbar district the screening test is only carried out in the Government Civil Hospital only there is not any Non-Governmental Organization or any private organization to carry out the screening in Nandurbar district. In Nandurbar district along with Newborn Screening, government has also introduced screening of β-Thalassemia and other Hemoglobinopathies follow up the care of birth cohorts over seven years, [46] it was introduced in the year 2012 in Nandurbar district of Maharashtra state, it has helped to understand the natural history of sickle cell disease and reduce early Morbidity and Mortality rate in Talukas of Nandurbar district.[59, 65] These have helped in other districts of Maharashtra states like Nagpur. [70] In Nagpur in Maharashtra, a targeted screening approach was used where 1162 babies of sickle heterozygous mothers were screened and 536 babies were Sickle Cell Carriers, 88 babies were Sickle Homozygous, 4 had Sickle β-Thalassemia and 2 has HBSD disease.

VI. CARE OF SICKLE CELL DISEASE CASES IN TRIBAL REGIONS OF NANDURBAR

[24, 33] Nandurbar basically is a tribal district; it is a part of khandesh. Nandurbar has six Talukas (Sub-Districts) which has many of villages as Akkalkuwa has 192 villages, Shahada has 181 villages, Akrani(Dhadgaon) has 163 villages, Navapur has 157 villages, Nandurbar has 150 villages and Taloda has 92 villages basically, delivering healthcare to tribal populations is a challenge and a village based model has been described in Bardoli in Gujarat where an outreach programme is being undertaken with the help of a mobile clinical unit and a local villager has also been given basic health care training to regularly visit and monitor sickle cell disease patients and send those with significant complications to the hospital coordinating the programme.Similarly, in Nandurbar district also a mobile clinical unit is active to give the better care for the patients of sickle cell diseases. These mobile units basically in Maharashtra are known as “ASHA” organization is a sub part of the Civil Hospital mobile unit regulated by the Government Hospital of the district. It regulates the polio dose camps, sickle cell care programmes, hygiene training camps, first aid camps and medicine distribution camps in the isolated areas of Nandurbar, because Nandurbar is on a high altitude and this is a tough challenge to treat the patients who are from isolated areas mainly Tribal’s. Care such as the iron and folic acid tablets are given to the patients freely by the Government Hospital as well as “ASHA” organization. Monthly camps are organized by the district hospital. It has also been demonstrated in remote tribal population from Dhanora, Subtibane, Shehi, Bhavani pada, Gokhalpada, Umachh, Molgi, Daab in Nandurbar district where 69 percent of the 106 patients had at least one annual comprehensive clinical visit or visit by the mobile clinical unit and 0.10% painful episodes per year were reported and this study were also reported premature deaths in

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16 patients at a median of average age of 26 years mainly females due to acute chest syndrome, sepsis, severe anemia, stroke, mesenteric infarction or sudden unexplained death. In this study it was estimated that females are higher rate in sickle cell anemia disease and around 96% of the patients to this study was of Tribal Casts or are Tribal's. [25, 62, 71] A prospective study from Odisha showed that neonatal outcomes such as low birth weight, prenatal mortality rate, admissions to the neonatal care unit, intrauterine growth retardation and preterm births were significantly higher in sickle cell anemia mother with successful pregnancies being achieved in 84.44 percent of cases. [32, 43, 44] There was no maternal mortality; however, there were five intrauterine fatal deaths and one early neonatal death. There is a data made from the Sickle Cell Disease Control Programme block wise total test it was conducted in April 2009 to March 2015.

SICKLE CELL DISEASE CONTROL PROGRAMME (block wise total test) [APRIL 2009 to MARCH 2015]										
Block	Total Population	Total Solubility Test	Positive	Electro phoresis	Carrier	Disease	Normal	% Of Solubility	% Of Disease	% Of Carrier
Nandurbar	231132	159359	8065	8065	3007	631	4427	69	0.27	1.30
Shahada	342724	147700	5337	5337	3469	304	1564	43	0.08	1.01
Navapur	246812	189998	15415	15415	9257	566	5592	77	0.22	3.75
Akkalkuwa	333483	203727	8810	8810	5625	408	2777	61	0.12	1.68
Dhadgaon	253139	203691	13592	13592	9248	928	3416	80	0.36	3.65
Taloda	241005	105630	3671	3671	1572	234	1865	44	0.09	0.65
TOTAL	**1648295**	**1161743**	**54890**	**54890**	**32178**	**3071**	**19641**	**62**	**0.18**	**1.95**

Table1: -Sickle Cell Disease Control Programme (April 2009 to March 2015)

VII. ELEVATION CONCEPT

[22, 36, 39, 49] Previously, it was claimed that the Tribals which are very prevalent to malaria or the Tribals from Sub-Saharan and African Sub Continent have the chances of Sickle Cell Diseases or Blood Related Disorders, but in this theory which we named as the "Elevation concept" we have claimed that by studying the areas of Nandurbar, Nashik, Dhule, Jalgaon and Nagpur. It was estimated that "As the Higher Elevation level/Higher Altitude level increases the diseasedstate alsoincreases" this is because when the Elevation level increases from place to place the rate of atmospheric oxygen level decreases and which leads to the blood with a rigid sickle like shape due to inadequate oxygen supply and leads to the diseases like sickle cell anemia or other sickle cell disorders. It is common in Nandurbar district among the Tribals because they live on the top hilly areas such as Satpuda mountain ranges.

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This Concept is explained by the given graph.

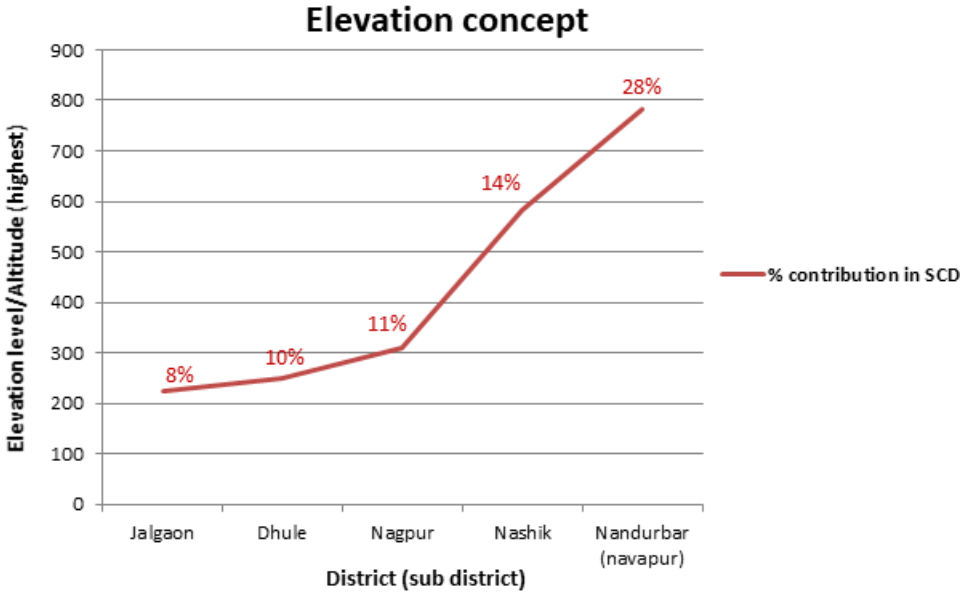

Fig 3: - The Elevation Concept

VIII. KEY MESSAGES

- Tribals are on a higher risk to Sickle Cell Diseases.
- Oxygen level should be maintained in the body.
- Blood Transfusion must be done by the age of 2 years.
- Mortality and Morbidity is highest below the age of 10 years.

IX. CONCLUSION

Our study suggests that in parts of eastern Maharashtra, the frequency of sickle cell trait is quite high irrespective of caste and creeds. The study further reveals that the urban and rural parts of Nandurbar district have more frequency of sickle cell anemia. Thus, for maintaining good health in the tribal areas, the steps have to be taken to restrict the further spread of sickle cell trait by proper management, treatment and creating awareness, a proper management are also involved screening of people with sickle cell trait and positive results needs to be treated with better treatment promptly and rigorously severe pain should be considered as a medical emergency that prompts timely and aggressive management until the pain is tolerable. Genetic counsellors should have to be focused on premarital screening positive patients should be informed about their implications and about their family planning. Until now, there is no specific treatment for the individuals with sickle cell disease; however, there are many prophylactic measures that help to limit the factors that may trigger sickling crisis episodes and complications capacitating the basic healthcare centres should be made a priority, since simple interventions, such as neonatal screening, immunization and education can have a great impact on public health. The most important thing is awareness among the peoples, especially the Tribals, prevalence of sickle cell anemia would be avoided through increasing awareness of the peoples by many effective ways. It can be created by inclusion of relevant information in the school curricular has been adopted, articles can also be published in newspapers and television sets, but in tribal community few peoples use newspapers and television sets, at that condition government can ask help from various non-government organizations and regional political leaders for their help and support to create awareness among the tribal population. It has also been concluded that the disease is common among tribal population and the Tribals which live on a top elevated areas such as Nandurbar and its sub-districts (Talukas) which have a high elevation in Maharashtra state. Due to high elevation supply of oxygen to blood (RBC) does not completes, so the shape of red blood cells (RBC) turns to sickle or rigid shape, as given in “elevation theory” so, to overcome these effective measures are to be taken.

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