

# Spatio-Temporal Epidemiological Analyses of Sickle Cell Disorder of a Tribal Region in Odisha, India



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**Abstract:** Now sickle cell disease (SCD) is an important health issue in many countries. India has been ranked as the second worst affected country in terms of predicted SCD births. SCD is an abnormal of the hereditary haematological disorder. The study was including 57 number of SCD patients which belong to the tribal region. These patients experienced with several symptoms like fever, loss of appetite, abdominal distension, myalgia, pain in legs, jaundice and black stool. Female individuals were more prevalent to SCD in compare to male. The SCD patients with an average age of 12.99 ( $\pm 11.66$ ) years. There was found significant difference in between the male individuals at ( $p < 0.01$ ). In these patients the concentration of haemoglobin, iron and ferritin also varies. In ABO blood group O blood group more prevalent than other blood group. SCD often causes complications on many organs. As the prevalence is more common in children, this will affect the socioeconomic status of a nation. This becomes a major challenge for healthcare professionals. Proper diagnosis and awareness for SCD in susceptible region is necessary for the better management and control of this disease. A well-designed epidemiological analysis will be necessary for better treatment and prevention of this disease.

**Index Terms:** Complication, haemoglobin, SCD, socioeconomic, tribes.

## I. INTRODUCTION

SCD is now one of the lives threatening haematological disorder. According to the World Health Organization, annually three lakh to five lakh new born born with are having haemoglobin disorders and from these more than about twenty thousand individuals are SCD (Alenzi and AlShaya, 2019). The global burden of SCD is increasing and the annual number of affected newborns may be expected to increase approximately from 300,000 to more than 400,000 between 2010 and 2050. In Sub-Saharan Africa the incidence is more than other, but the highest allelic frequencies have been reported in Indian populations (Hockham et al., 2018). India has been ranked as the second worst affected country in terms of predicted SCD births (Piel et al., 2013). India has the

largest tribal populations globally. It is suggested that tribes of Nilgiri hills was the main source of sickle cell haemoglobin (HbS) and later it affected rest parts of India (Lalie et al., 1989). According to the latest census of India conducted in , tribal populations account for about one fourth of Indian population. In India, SCD is predominantly found amongst scheduled tribe (ST) and scheduled caste (SC) populations. These are the most socioeconomically backward and disadvantaged population subgroups in the country. The average frequency of HbS trait is about 20% in Chhattisgarh, Maharashtra, Madhya Pradesh, Odisha and Gujarat in India (Nagar and Raman, 2015). In Odisha, SCD is a serious concern about health status of the state (Colah et al., 2015).

SCD is caused by HbS (Ashley-Koch et al., 2000). It is caused due to the mutation in the molecular level of globins proteins (Akinbami et al., 2012). Globins are the proteins which present in the erythrocytes as a component of haemoglobin. In this disorder the thymine is replaced with adenine and it substitutes the glutamic acid with valine in globin protein. It occurs on the short arm of eleventh chromosome. This disorder is caused due to the formation of abnormal haemoglobin, called HbS. The life span of normal red blood cells (RBCs) reduces from 120 days to 20 days. Consequently it makes the normal RBCs to stiff, rigid and sickle cells. The primary pathophysiology is based on the polymerization of deoxyHbS with formation of long fibers within the RBCs and triggers the cascade of erythrocyte alterations (Damanhour, 2015). HbS polymerization is directly related various pathophysiological disorder. The pathophysiology of SCD is also characterized with multi organ failure, a periodic pain, inflammation, rapid infection, stroke and delayed growth with late sexual maturity (Lionne et al., 2012). It may be fatal without effective treatment within the first few years of incidence. It can cause morbidity both in acute and chronic condition (Piel et al., 2017).

During last decade, many institutions in India have made a remarkable survey to estimate the prevalence of SCD in different village up to state level (Colah et al., 2015).. Various data analyses related to sickle cells in India have been published previously (Patra et al., 2015). Currently the map accounting for the socio-demographic complexity of the Indian population especially in Koraput district of Odisha is inadequate. It should enable better assessment for treatment, prevention and also proper management.

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Here we analyse the data of recent survey of Koraput district at different hospitals level. It will present an evidenced map in Koraput district of Odisha, India with variation to the environmental condition. This work will provide an important public health resource for developing model of care at national and regional levels. Burden of SCD according to geographical distribution is essential for recent public health policies (Kato et al., 2018).

The main objective of this investigation was the gender ratio, age, haematological and biochemical variation in tribal community of Koraput district of Odisha in Indian patients with sickle cell anaemia.

**II. METHODS**

These investigations were based on 57 numbers of SCD patients from different hospitals and health centres of Koraput city. As it is a tribal rich region, here mainly local people come for the treatment of several haematological disorders. The study was conducted in different hospital and health centres of Koraput city, Odisha and analysed at Centurion University of technology and Management, Odisha from July, 2018 to February 2019.

During the study the patients were experienced with fever, loss of appetite, abdominal distension, myalgia, pain in legs, pallor and black stool etc. During this retrospective study, patient records were identified according to their diagnosis by the help of physician. A data collection sheet was developed to collect information from patient records. The sheet included socio-demographic data like age, gender, location and nationality, as well as variables related to clinical data related to laboratory findings, were tabulated and presented as mean±SD. Comparison of clinical presentation and laboratory findings by gender of the studied cases were statistically analysed independent t-test. The value  $p < 0.05$  was used as an indicator of statistically significant differences. An internet-based search through Pubmed web site was used to find prior related study and data were analyzed using the PAST statistical software (Chainy et al., 2008).

**III. RESULTS**

The study was including 57 number of SCD patients and these patients were experienced with several symptoms like fever, loss of appetite, abdominal distension, myalgia, pain in legs, jaundice and black stool. In this study, out of which 24 were males (42.11%) and 33 were females (57.89%). The average age of SCD patient was 12.99 (±11.66) year. For male the mean age was 10.95(±12.09) years and for female was 15.04(±11.08) years. On comparison with male and female age, it was found that no significant difference at ( $p < 0.05$ ), but significant difference was found in between the male individuals at ( $p < 0.01$ ). In case of female, the ages also showed difference highly significantly ( $p < 0.01$ ).

**A. Age and blood group analysis**

Patients were divided in different age group as shown in table 1, prevalence was more in between 0-10 year age group in both sexes and less number of individuals was found in more than 30 years age group.

**Table 1 Individuals at different age group**

| Age group in years | Numbers of Male | Numbers of Female |
|--------------------|-----------------|-------------------|
| 0-10               | 14              | 12                |
| 10-20              | 7               | 6                 |
| 20-30              | 2               | 8                 |
| 30-40              | 1               | 7                 |

At 0-10 year age group male was more prevalence as compared to female and in the age group of 20-30 years females were more affected than male. Due to limited therapeutic options for SCD, blood transfusions are very essential. During this study blood group was also considered in table-2 as one of the parameter for the study. These investigations showed that O blood group has the highest prevalence than other ABO blood groups in the SCD individuals. From ‘O’ blood group female individuals were more affected as compared to male and AB blood group were least affected. During the survey, the individuals with O,A, B and AB blood groups were affected and showed the incidence in decreasing order. This study shows very high prevalence of Blood group O and rare AB blood groups among SCD individual. High frequency of ‘O’ blood group are advantages in transfusion as universal blood donor (Nwabuko and Okoh, 2017).

**Table 2 individuals with ABO blood group in both male and female**

| Blood group | Number of males | Number of females |
|-------------|-----------------|-------------------|
| O           | 9               | 11                |
| A           | 7               | 9                 |
| B           | 3               | 2                 |
| AB          | 2               | 2                 |

**B. Haemoglobin Concentrationa**

As SCD is a haematological disorder related to haemoglobin (Ansari et al.,2018). The concentration of haemoglobin of normal male individual is about 14 mg/dL and female individual is about 12 mg/dL. In this study the patients were having low concentration of haemoglobin table-3. Low haemoglobin is one of the useful biomarker for SCD. Due to cerebral problem, it is related with strokes (Gueguen et al., 2014) and may cause early death (Andemariam et al., 2014).The mean concentration of haemoglobin of male was 6.11(±2.55) and mean concentration of haemoglobin in female was 6.89(±1.66). At ( $P < 0.05$ ) there was no significant difference in haemoglobin of both the genders.

**Table 3 Concentration of haemoglobin in both male and female patients**

| Sl no. | Concentration of Haemoglobin mg/dL |        |
|--------|------------------------------------|--------|
|        | male                               | Female |
| 1      | 8.5                                | 5.9    |
| 2      | 8.3                                | 7.7    |
| 3      | 3                                  | 7.9    |
| 4      | 9.2                                | 3.1    |
| 5      | 7.9                                | 8.2    |



|            |      |      |
|------------|------|------|
| 6          | 4.2  | 5.7  |
| 7          | 6.2  | 8.4  |
| 8          | 2.2  | 7.2  |
| 9          | 4    | 6.4  |
| 10         | 7.6  | 8.4  |
| Mean       | 6.11 | 6.89 |
| Std. error | 0.8  | 0.5  |
| Std. dev   | 2.55 | 1.66 |

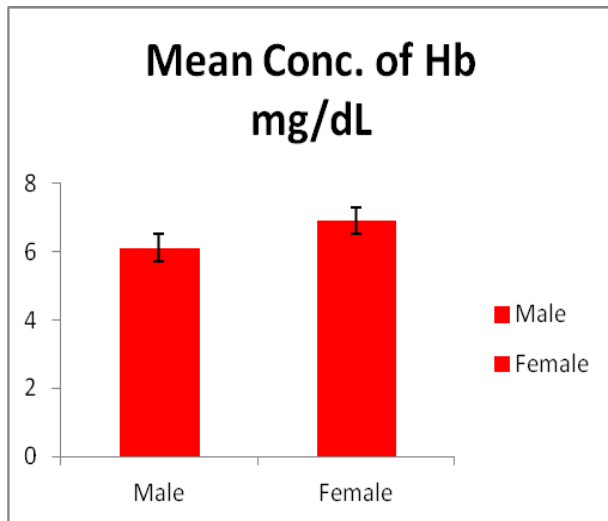


Fig.1. Haemoglobin concentration in both male and female in this study.

C. Iron and ferritin

Primarily iron stores in the form of ferritin and small amounts of ferritin are present in plasma. The plasma or serum ferritin is positively correlated with the total body iron. Anaemic patients generally need blood transfusions and that lead to iron overload. If untreated, accumulation of iron may enhance generation of free hydroxyl radicals that may lead to increase the morbidity and mortality (Mishra and Tiwari, 2013). In SCD the concentration of iron and ferritin also varies beyond the normal range. During the study, data of ten patients were represented with haemoglobin, iron and ferritin in table 4. Sickle cell disease patients experiences frequent pain and often cause haemolysis due to iron concentration. The concentration of Hb level is known to influence the blood viscosity in SCD patients. High serum ferritin concentration need to be deal cautiously in SCD patients with no history of chronic blood transfusion, as serum ferritin can be elevated in different medical conditions like fever, infection, inflammation, and hepatic dysfunction(Alsuliman et al.,2014).

Table 4 Individuals having different concentration of haemoglobin,iron and ferritin

| Sl Number | Hb(g/dL) | Iron (µg/dL) | Ferritin(ng/mL) |
|-----------|----------|--------------|-----------------|
| 1         | 7.8      | 100          | 160             |
| 2         | 8.4      | 58           | 40              |
| 3         | 7.2      | 30           | 18              |
| 4         | 6.4      | 42           | 35              |
| 5         | 7.9      | 89           | 110             |
| 6         | 6.8      | 48           | 22              |
| 7         | 6.7      | 30           | 50              |
| 8         | 6.5      | 68           | 100             |
| 9         | 6.1      | 80           | 600             |
| 10        | 7.2      | 32           | 20              |

IV. CONCLUSION

The development and understanding of this disease has been in gradual process since nineteenth century. SCD patients are more sensitive and unaware due to certain social and economic support. It is necessary to develop better management and socio-economic status of tribal people. It should be addressed by social awareness for creating utmost care of patients. SCD often cause complications on many organs. Long-term transfusion in SCD is a feared to be iron overload and this may lead to renal failure. It should be strongly emphasized at the clinical level to reduce complications of iron overload by minimising the use of non-transfusion therapy. Such strategies may help in reducing morbidity and mortality associated with disease.As the prevalence is more common in children, this will affect the socioeconomic and quality of life. Well-designed epidemiological surveys will be crucial to further assess the prevalence and burden of SCD in the regional as well as national. Necessary steps should be taken to improve outcomes by management strategies. In coming future, SCD looks more promising and hopeful research with social empowerment.

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work, membership, achievements, with photo that will be maximum 200-400 words.