

Population, Ecology And Epidemiology of the Sickle Cell Disease in Orissa

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ABSTRACT The eco-biological diversity not only influences the quality of life, but also determines the disease pattern and morbidity. The congenial bioenvironmental conditions proliferate the healthy people. This study gives an account of the population, ecology, health status and distribution of the sickle cell disease in Orissa. The geographical distribution of the sickle cell disease entertains the geomedical diversity in the Eastern Coast region and Western part of Orissa. The genetic load of sickle cell anemia in Orissa may be eroded by limited population growth, preventing consanguineal marriages, providing better medical facilities and nutrition, improving health scenario by enhanced social development and proper health education to the poorest of the poor.

INTRODUCTION

The sickle cell haemoglobinopathy is becoming a major genetic and public health problem in the state of Orissa. It causes haemolytic anaemia, high morbidity, mortality and reproductive wastage and affects the general health, psycho-social behaviour, manpower and drains the economy of a large number of tribal as well as non-tribal people (Balgir, 1993). It is, therefore essential to check and prevent the growth and spread of such a deleterious disease which drains off the substantive economy of the state. The population of India is growing annually at a rate of 2.1% and the addition of unhealthy persons further compound burden on the national resources. At least one child with genetic defects is born in India in every 40 seconds and every year 5000 infants are born with hereditary anaemia (Verma, 1991).

The distribution of various human diseases

in space and time in relation to geographical and bioenvironmental variables is fascinating. The present study deals with the distribution of sickle cell disease in Orissa in relation to population and health status of the people.

PHYSIOGNOMY AND ENVIRONMENT

The state of Orissa may be broadly divided into four distinct geographical regions, namely, Northern plateau, Central river basin, Eastern hills, and Coastal plains, each with different characteristics. Cultivation of rice is the principal occupation of about 76% of the people. About 35.8% of the total geographical area of the state is under forests (55,785 sq. km). The climate of Eastern coast region is hot and humid with cool evening and night. The Western part of Orissa has hot and dry climate with high temperature, sometimes, rising upto 49°C. This region as a whole is drought prone with scanty rainfall. The majority of the people are poverty stricken and vulnerable to tropical diseases.

POPULATION

According to the Census reports of 1981 and 1991, the total population of Orissa State was 26,370,271 and 31,659,736 persons, respectively with annual growth rate of 2%. The Scheduled Caste (SC) and Scheduled Tribe (ST) constituted 3,865,543 (14.7%) and 5,915,067 (22.4%), respectively and 37.1% of the total population of the state in accordance with the 1981 report. However, according to 1991 Census, the SC and ST were

5,129,314 (16.2%) and 7,032,214 (22.2%), respectively with total 38.4% of the total population of Orissa.

A large number of general caste, SC and ST people have sickle cell haemoglobinopathy in Orissa (Balgir, 1992a). There are 60 million carriers and 1,20,000 sickle cell homozygotes are added every year in the World (WHO Report, 1983). There were 2,434,170 carriers and 1,21,375 sickle cell disease homozygotes among the tribes of India as per the calculations based on 1981 Census data (Rao, 1988).

In Orissa alone, there were 94,410 carriers and 1,726 homozygous for sickle cell disease as per the estimates based on 1981 Census data (Rao, 1988). We are of the opinion that these figures are under-estimates (Balgir, 1992a). According to our estimates, there were 3-4 million people suffering from sickle cell haemoglobinopathy based on 1981 Census data in Orissa. About 2-3 million belonged to general castes, three fourth of a million to SC and one-fourth of a million were ST as calculated for 1981 Census data (Balgir, 1992a).

The sickle cell disease is an autosomal inherited anomaly which means that both sexes are vulnerable. When the disease is inherited from one parent, we call it a sickle cell trait (heterozygote) and when both parents contribute, it is called a sickle cell disease. Marital consanguinity (marriage among blood relatives) increases the homozygosity in the population, leading to high prevalence of sickle cell disease. Consanguineous marriages, especially the cross-cousin are prevalent among the Agharia, Kulita, Chasa, Gaud, Khandayat, Dumal, Oilmen, Ganda, Ghasi, etc. in Orissa (Balgir, 1993).

SICKLE CELL AND MALARIA

Persons with sickle cell trait develop malaria less often and less severely than those without the trait. Sickle cell anaemia is most

frequent in regions with a high incidence of virulent form of *Plasmodium falciparum* infection (Balgir, 1992b). In heterozygotes, the erythrocytes parasitized by *Plasmodium falciparum* are more effectively removed from the circulation by phagocytosis than in normal individuals. The sickle cell trait confers some protection against falciparum malaria during critical period in early childhood between the loss of passively acquired maternal immunity and the development of active immunity (Serjeant, 1992). Similarly, the erythrocytes containing fetal haemoglobin are more resistant to infection by *Plasmodium falciparum* than those containing only adult haemoglobin.

Malaria not only causes early death but predisposes the individual to considerable ill health also. In a community in which malaria is endemic, those who are more resistant to infection, not only survive better, but will have a higher reproductive fitness (Balgir et al., 1995). The increased fertility of the heterozygotes balances the elimination of sickle cell genes through affected homozygotes (sickle cell anaemia).

DISTRIBUTION OF SICKLE CELL GENE

In Orissa, our (Balgir, 1994b) experience shows that the sickle cell haemoglobinopathy is more prevalent in Western Orissa (Table 1) especially in the districts of Sundargarh, Deogarh, Jharsuguda, Sambalpur, Bargarh, Bolangir, Kalahandi, Nowrangpur, Koraput, Phulbani and Dhenkanal than in the coastal districts, namely Mayurbhanj, Balasore, Cuttack, Khurda, Puri, Ganjam, etc. (Fig. 1). This situation is probably due to marital consanguinity, territorial proximity and endogamy, Caste/Tribe and class hierarchy, economy, geographical barriers (hills, forests, river, etc.), climatic variations and eco-biological diversity. Further, the sickle cell gene is not only prevalent in the tribal or scheduled



Fig. 1. Map of Orissa showing different districts

Table 1 : District-wise distribution of Sickle Cell Disease (HbSS) and Trait (HbAS) cases (in per cent) at the Sickle Clinic, V. S. S. Medical College Hospital, Burla in Orissa

District	Schedule Tribe		Schedule Caste		General Caste		Total	
	Hb AS	Hb SS	Hb AS	Hb SS	Hb AS	Hb SS	Hb AS	Hb SS
Balasore ¹	0.0	0.0	0.0	0.0	0.1	0.0	0.1	0.0
Bolangir ²	1.1	0.7	4.6	2.4	4.7	4.9	4.3	3.8
Cuttack ³	0.0	0.0	0.0	0.0	0.1	0.1	0.1	0.0
Dhenkanal ⁴	0.0	0.0	1.5	0.3	3.9	3.6	2.9	2.4
Ganjam ⁵	0.0	0.0	0.1	0.0	0.4	0.4	0.3	0.3
Kalahandi ⁶	0.4	0.0	1.2	0.5	0.8	0.6	0.8	0.5
Keonjhar	0.0	0.0	0.1	0.1	0.1	0.2	0.1	0.2
Kopraput ⁷	0.4	0.4	0.0	0.0	0.1	0.1	0.1	0.1
Mayurbhanj	0.0	0.4	0.1	0.1	0.0	0.1	0.0	0.1
Phulbani ⁸	0.0	0.4	0.9	0.3	0.5	0.3	0.6	0.3
Puri ⁹	0.0	0.0	0.0	0.0	0.1	0.0	0.0	0.0
Sambalpur ¹⁰	8.8	21.6	26.5	24.2	32.3	24.9	28.3	24.4
Sundargarh	0.4	1.5	0.6	0.5	3.2	2.2	2.2	1.7

1 includes Bhadrak; 2 includes Sonepur; 3 includes Jagatsingpur, Jajpur and Kendrapada; 4 includes Angul; 5 includes Gajapati; 6 includes Nawapara; 7 includes Malkangiri, Nawarangpur and Rayagada; 8 includes Boudh; 9 includes Khurda and Nayagarh; 10 includes Bargarh, Deogarh and Jharsuguda districts

caste populations of Orissa, but it has penetrated even into the general castes of Orissa (Balgir, 1994a,b,c, 1995).

Genetic load of sickle cell anemia is indeed very high affecting not only the health and physical performance but also poses a challenge to the health services. It is estimated that 1,86,096 cases of sickle cell anaemia are present in the Indian sub-continent (Bhasin et al., 1994). Each patient is not only burden on the health services but also affects normal family life. It is, therefore, essential to check the birth of abnormal individuals especially with sickle cell disease through prenatal diagnosis and therapeutic abortion. Such facilities are although meagre in India, but available which need to be extended to those regions where the prevalence of hereditary diseases such as sickle cell disease, G-6-PD deficiency, etc. is alarmingly high. Management of genetic disorders through preventive, promotive and curative methods is important. With improving environmental and socio-economic conditions, better public health, and medical facilities, effective malarial prophylaxis, and better nutrition, children with sickle cell disease can be better managed.

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