© Kamla-Raj 1998
PRINT: ISSN 0970-9274 ONLINE: 2456-6608
J. Hum. Ecol., 9(3): 281-284 (1998)
DOI: 10.31901/24566608.1998/09.03.13

Age, Sex and Seasonal Variations of Sickle Cell Disorder Cases in Orissa

B.P. Dash and R.K Das

Regional Medical Research Centre, Indian Council of Medical Research, Chandrasekharpur, Bhubaneswar 751 016, Orissa, India

KEY WORDS Sickle Cell Disorders. Age. Sex. Season. Orissa.

ABSTRACT A total of 451 sickle cell haemoglobinopathy cases aged from 4 months to 55 years were recorded in a clinic of Sickle Cell Research Centre (ICMR), Burla during the period from September 1991 to April 1993. Out of these, 442 cases (280 Males and 162 Females) were homozygous sickle cell disease or sickle cell- beta thalassaemia. One case of haemoglobin SD disease was encountered. Besides sickle cell disorders, cases like haemoglobin D trait (One case), haemoglobin E thalassaemia (One case) and 6 cases of Beta thalassaemia major were also encountered during the period under report. The magnitude of the problem is more in some general caste groups (Kulita, Chasa, Agharia, Gouda, etc.) than the Scheduled Castes and Scheduled Tribes. Regarding the seasonal attendance, it was found that more sickle cell disease cases attended the clinic in rainy months (June to September) than in Winter and Summer season.

INTRODUCTION

Several reports are available for the occurrence of homozygous sickle cell disease (Kar, 1991), sickle cell- beta thalassaemia (Kulozik et. al., 1991), haemoglobin SD disease (Kar,1991) and sickle cell alpha thalassaemia (Kulozik et al., 1988) in the state of Orissa. The clinical and haematological aspects of these disordrs have been well documented (Kar et al., 1986; Kulozik et al., 1988). These reports are based on the data collected during the field camps. Kar et. al. (1987) have reported the widespread distribution of sickle cell gene in Orissa based on sickling test of a large hospital sample in which the distinction between the heterozygotes and homozygous states has not been done. The homozygous sickle cell cases are also known as the sickle cell disease patients. These patients mainly suffer from anemia, mild jaundice, frequent pain attacks and require regular health check-up (Serjeant, 1985). As sickle cell gene is highly prevalent among different castes and tribal populations of Orissa, it is likely that, quite a good number of cases would have attended the local hospital which is falling in the sickle cell belt. So far, the magnitude of hospital load due to these genetic disorders has not been evaluated. Further, the pain crises of these cases vary from patient to patient and also related to seasonal variations in other countries, espcially in Africa and Jamaica (Serjeant, 1985). It is, therefore, worthwhile to have an idea on these aspects in the Indian setting. We have analysed data of a special clinic in a hospital which is centrally located in the sickle cell belt and caters to population of the states of Orissa and Madhya Pradesh.

MATERIAL AND METHODS

A total of 451 cases with different haemoglobinopathies attended the clinic of Sickle Cell Research Center (ICMR), V.S.S Medical College Hospital, Burla, Orissa during the period from September 1991 to April 1993. Data pertaining to age, sex, caste and monthly attendance of the cases were recorded in the register. The diagnosis was done by the wet sealed sickling test and performing the haemoglobin electrophoresis (Dash, 1978) both in alkaline (pH 8.6) and acidic medium (pH 6.2). HbA2 and HbF estimations of some of the selected cases and parents were also done for the identification of beta thalassaemia trait and patients following the standard procedure(Weatherall, 1983). As the parents of all the cases were not available, we have put together both the homozygous sickle cell disease and sickle cell - beta thalassaemia cases.

RESULTS

During the study period, 443 cases were suffering from sickle cell disorder. Out of these,

one is haemoglobin SD disease (a 10 year old Agharia boy from Raigarh in Madhya Pradesh) and the other 442 cases were homozygous sickle cell disease and sickle cell-beta thalassaemia. Out of 451 cases, one haemoglobin D trait, one haemoglobin E thalassaemia and 6 beta thalassaemia cases were also recorded (Table 1).

Table 1: Distribution of haemoglobinopathy cases

Haemoglobinopathies	*Male*	*Female*	*Total*
Total No. of cases recorded	284	167	451
No. of Sickle cell disorder cases recorded (Homozygous SCD+ Sickle cell Beta thalassaemia cases)	280	162	442
Haemoglobin SD case	1	-	1
Haemoglobin E-thalassaemia	1	-	1
Haemoglobin D trait	-	1	1
Beta thalassaemia cases	2	4	6

The age and sex distribution of 442 cases is given in table 2. There were 280 males and 162 females and more than 90% of total cases were between the age groups of 0-30 years

Table 2 : Age and sex wise distribution of sickle cell disorder cases

Age groups (in years)	*Male*		*Female*	
	No.	*%*	*No.*	*%*
<10	120	42.9	66	40.8
10-19	81	28.9	54	33.3
20-29	56	20.0	33	20.4
30-39	15	5.4	8	4.9
40-49	6	2.1	1	0.6
>50	2	0.7	0	0.0
Total	280	100.0	162	100.0

Data for the number of patients attended the clinic during different months and during different seasons are shown in figure 1. It is evident from the graph that more cases attended the clinic in the rainy season (June-September) than in the winter (October- January) and summer (February-May) months.

Out of 442 sickle cell disorder cases, 300 patients attended the different out-patient departments (OPDs) and 142 cases got admitted to different wards (indoor) of V.S.S. Medical College Hospital, Burla. During the study period, 82 and 55 cases were admitted in Medicine and Paediatric wards respectively, while 2 cases were referred from Surgery and one each from ENT, Orthopaedic and Urology wards (Table 3).

Table 3 : Hospital attendance of sickle cell disorder cases

	Patients	*No.*	*%*
(A)	Outdoor patients	300	67.9
(B)	Indoor Admissions	142	32.1
	a. Medicine	82	18.6
	b. Paediatric	55	12.4
	c. Others (Surgery, ENT, Urology)	5	1.1

Caste-wise distribution of sickle cell cases is given in table 4. The majority of them belonged to general caste (80.5%) followed by Scheduled Caste (16.7%) and Scheduled Tribe (2.7%).

Table 4 : Caste-wise distribution of sickle cell disorder cases

General Caste		*Scheduled Caste*		*Scheduled Tribe*	
Name	*No.*	*Name*	*No.*	*Name*	*No.*
Kulita	100	Ganda	49	Gond	7
Chasa	82	Ghasi	12	Munda	3
Agharia	47	Pana	8	Bhuyan	1
Gouda	34	Damba	1	Paroja	1
Teli	31				
Dumal	15				
Kshetriya	9				
Khandayat	7				
Others	35				
Total	360	Total	70	Total	12
Grand Total	442		-		-

DISCUSSION

From the present study, it is evident that the sickle cell disorder is the most common haemoglobinopathy in Orissa. During the period from September 1991 to April 1993, we have recorded 6 cases of beta thalassaemia patients. Haemoglobin SD disease and haemoglobin E-thalassaemia cases were from the state of Madhya Pradesh and Bihar, respectively. The presence of sickle cell gene in Madhya Pradesh and Bihar is well known (Balgir and Sharma, 1988; Dash, 1994). The presence of HbD and HbE is rare in these states. Since the inter-caste marriages and migration of population between different neighbouring states are taking place, it is possible to detect these abnormal cases in different states of India.

It is interesting to note that about 90 % of the total cases who attended the clinic were

Fig. 1. Hospital attendance of sickle cell disease cases

between 0 to 30 years of age. Only 23 males and 9 females were above 30 years of age. This could be due to early death of the patients (Kar, 1991), elimination of SCD cases from that locality or due to less morbidity in older patients (Serjeant, 1985).

One of the salient feature of the present analysis is that, more number of sickle cell disease (SCD) cases were recorded in rainy and winter than in the summer season. This could be due to the cloudy and cold weather which is known to be one of the precipitating factor for pain crisis in SCD patients (Serjeant, 1994). Further, more males attended the clinic though both sexes are equally vulnerable. This could be due to partly bias towards females and partly more awareness among the male individuals because of socio-cultural reasons or people are more consious about the health of male members only.

Distribution of cases in different caste categories shows that more cases belonged to general caste namely, Kulita, Chasa, Agharia, Gouda, Teli, etc. Likewise, more cases were from Scheduled Caste Populations, like Ganda, Ghasia, etc. than the Scheduled Tribes. Based on this analysis, it is clear that sickle gene is widely distributed in some selected caste groups. This could be due to endogamy and consanguineous marriages among them (Balgir, 1995).

Regarding hospital attendance of these patients, it was seen that more cases were referred from different OPDs than from different wards. Out of indoor cases, more were from Medicine and Paediatric wards of the hospital. Only 5 cases were referred from other indoor wards. The main clinical symptoms of these patients were anemia, mild jaundice, repeated vaso-occlusive crises, haepato-splenomegaly (Kar et al., 1987; Kar, 1991) and other complications which require surgical or special intervention were rare.

ACKNOWLEDGEMENTS

We are very much thankful to Dr. K. Satyanarayana, Director, RMRC (ICMR), Bhubaneswar for providing the necessary facilities for this study.

REFERENCES

Balgir, R.S. and Sharma, S.K. : Distribution of sickle cell haemoglobin in India. *Ind. J. Haematol.*, **6**:1-14 (1988).

Balgir, R.S. : Population, ecology and epidemiology of the sickle cell disease in Orissa. *J.Hum. Ecol.*, **6** : 273-276 (1995)

Dash, B.P.: *Study of Glucose Metabolism in Sickle Haemoglobin Containing Human Red BloodCells*. Ph.D. Thesis, Sambalpur University, Burla (1994).

Dash,S. : A quick and simple method for routine haemoglobin electrophoresis. *Ind. J. Pathol. Microbiol.*, **21**: 839-841, (1978)..

Kar, B.C, Satpathy, R.K., Kulozik, A.E., Kulozik, M., Sirr, S. Serjeant, B.E., and Serjeant, G.R.: Sickle cell disease in Orissa state, India. *Lancet*, **22** : 1198-2001(1986).

Kar, B.C., Devi, S., Dash, K.C. and Das, M.: The sickle cell gene is widespread in India. *Trans. Royal. Soci. Trop. Med. Hygie.*, **81** : 273-275(1987).

Kar, B.C. : Sickle cell disease in India. *J.A.P.I.*, **39**: 954-960 (1991).

Kulozik, A.E., Kar, B.C., Serjeant, G.R., Serjeant, B.E., and Weatherall, D.J.: The molecular basis of α thalassaemia in India. Its interaction with the sickle cell gene. *Blood*, **71** : 467-472 (1988).

Kulozik, A.E., Bail, S. : Kar, B.C., Serjeant, B.E., and Serjeant, G.R.: Sickle cell-B+ thalassaemia in Orissa state, India. *Brit. J.Haematol.*, **77** : 215-220 (1991).

Serjeant, G.R. : *Sickle Cell Disease*, Oxford University press, New York (1985)

Serjeant, G.R., Charles, D.E.C., Rosemary, L., Joanne. M., Atul, S., and Peter, W.T. : The painful crisis of homozygous sickle cell disease : Clinical features. *Brit. J. Haematol.*, **87**: 586-591(1994).

Weatherall, D.J.: *The Thalassaemias. Methods in Haematology* **Vol.6**, Churchill Livingstone, NewYork (1993).