|| ISSN(online): 2589-8698 || ISSN(print): 2589-868X ||
International Journal of Medical and Biomedical Studies
Available Online at www.ijmbs.info
NLM (National Library of Medicine ID: 101738825)
Index Copernicus Value 2019: 79.34
Original Research Article
Volume 5, Issue 10; October: 2021; Page No. 04-07

ANEMIA IN SICKLE CELL TRAIT

[1]Dr. Bushra Khanam (Associate Professor), [2]Dr. Srajit Singh (PG resident), [3]Dr Manish Kumar Saraf (PG resident), [4]Dr. Rudraksh Gautam (PG resident)

[1,2,3,4]Dept. of Medicine, Index Medical College Hospital & Research Centre, Indore

Article Info: Received 04 August 2021; Accepted 21 September 2021
DOI: https://doi.org/10.32553/ijmbs.v5i10.2230
Corresponding author: Dr. Srajit Singh
Conflict of interest: No conflict of interest.

Abstract

Background & Method: Subjects for the study were recruited from the rural comprehensive sickle cell program at Index Medical College Hospital & Research Centre, Indore. These patients were followed up with regularly at the comprehensive sickle cell clinic with all relevant clinical and laboratory data documented in detailed case report forms. Patients were advised to follow up on a monthly basis or as symptoms presented.

Result: In our study we found, maximum no of cases in age group of 01-15 (45.6%) followed by 16-30 (30.6%) In our study we found 11 males >18years & 19 males <18 years & 11 females >18 years & 05 females <18 years.

Conclusion: We provide for the first time detailed prevalence data of the βs gene among Rural Population ethnic groups in Central India. These population screening programs have uncovered previously undiagnosed cases, provided detailed information for population based disease counseling, prevention programs and comprehensive care programs. Over 5,000 individuals were identified with SCT and more than 1,000 individuals were identified with SCD through community-based and target high-risk screening. The population screening study shows how community-based screening can identify individuals who may not have otherwise sought care and guide individuals to available comprehensive care centers. The screening also helped identify specific sub-groups within the Rural Population that have higher prevalence rates of the βs gene. An estimated 1.1 million individuals in the state of Madhya Pradesh alone are expected to have SCT. Similar community-based screening programs need to be initiated throughout regions of India that have a high prevalence of Rural Population to determine the true prevalence of the βs gene in India.

In summary, there appeared to be a broad general knowledge of sickle cell disease in the identified. target samples, with many 1nteresting comparative responses to the attitudinal survey, but no statistically significant differences 1n black and white responses in western countries.

That all persons experiencing genetic disease have the option to choose genetic counseling and sufficient community education to know they have this choice. That all genetic counseling include supportive family therapy and education over a sufficient period of time so the family can integrate the information into their own life systems.

Keywords: Anemia, Sickle & Trait.
Study Designed: Observational Study

Introduction

The condition of having sickle cell disease will be defined as occurring in one or the other of the two following states: (1) sickle cell anemia-a severe, uncorrectable, and often fatal anemia with many clinical manifestations, and (2) sickle cell trait--a relatively benign condition with symptomatology occurring only under extraordinary circumstances.

Hemoglobin functions as the oxygen carrying substance of the blood, andis responsible. for the pigmentation of the erythrocytes or red blood cells. The condition of sickle cell anemia results from the presence of an abnormal hemoglobin, known as Hb-S, in the erythrocytes. The presence of this hemoglobin is hypothesized to have been an asset in protecting· its bearer from the effects of malaria in those countries where the occurrence of malaria is common.

The trait form of· sickle cell disease has no clinical manifestations except under extraordinary circumstances, during which the individual might experience a crisis similar to that of 'the anemia victims. Thus, symptomatology in sickle cell trait can be virtually obliterated by avoiding those circumstances which would precipitate a crisis conditional Sickle cell trait individuals may be incapacitated by

(1) Conditions of extreme hypoxia, such as flying in an unpressurized aircraft or with underwater swimming, (2) dur1ng anesthesia when anoxia has inadvertently occurred, (3) occurrence of occasional severe pneumonia, .and (4) extr.eme physical exercise.

When a child dies, the enigma of death strikes heavily at those who experience this tragedy. One of the greatest crises a family must face is the threat of loss through

death of one of its members. When it is a child whose death 1s imminent, the crisis is imbued with an additional dimension of emotion. The loss of this child is especially poignant when death occurs as the result of genetically acquired traits that lead to a disease 'for which there is no cure. Sickle cell anemia, because of its genetic origins, and due to the frequent youthfulness of its victims, is such a disease. The following study will address itself to this subject of sickle cell anemia--an unfortunate, life destroying human condition(5).

Material & Method

Subjects for the study were recruited from the rural comprehensive sickle cell program at Index Medical College Hospital & Research Centre, Indore.

These patients were followed up with regularly at the comprehensive sickle cell clinic with all relevant clinical and laboratory data documented in detailed case report forms. Patients were advised to follow up on a monthly basis or as symptoms presented. Patients included in this study received treatment and care, including for acute events, at the Index Medical College Hospital & Research Centre, Indore M.P. from duration Sept 2020 to June 2021.

For assessing demographic information, frequency and percentage were computed for categorical data. For continuous data the median was computed, data categorized into meaningful groups and the frequencies computed within each group. For clinical data each visit with a certain condition was counted to assess its overall frequency. Furthermore the number of patients who had the specific condition at least once during the study period as well as the range of occurrences of the condition within a person across the study population was compiled. IBM SPSS 20.0 & MS Excel were used.

Results

Table 1: Age Distribution

S. No.	Age Group	No.	Percentage
1	**01-15**	**21**	**45.6**
2	**16-30**	**14**	**30.6**
3	**31-45**	**08**	**17.3**
4	**More than 45**	**03**	**06.5**

In our study we found, maximum no of cases in age group of 01-15 (45.6%) followed by 16-30 (30.6%)

Table 2: Gender Distribution

S. No.	Gender	No.	Percentage
1	Male	30	65.3
2	Female	16	34.7

In our study we found 11MALES >18YEARS & 19 MALES <18 YEARS& 11 FEMALES >18 YEARS & 05 FEMALES <18 YEARS.

Table 3: Hb

	Hb	HbA2	HbS	HbF	HbA
No.	46				
Mean	6.8370	3.0130	27.1543	1.9848	67.6152
Std. Error of Mean	0.39882	0.17467	1.46037	.65561	1.70037
Std. Deviation	2.70492	1.18464	9.90474	4.44657	11.53250
Minimum	1.90	1.70	4.60	0.30	33.80
Maximum	13.70	6.80	57.80	31.30	92.50

Table 4: MCV-MCH-MCHC

	MCV	MCH	MCHC
No.	46		
Mean	68.9696	21.3587	30.9696
Std. Error of Mean	1.78399	.65811	.49778
Std. Deviation	12.09962	4.46351	3.37612
Minimum	41.70	11.30	24.00
Maximum	95.10	31.40	40.00

Discussion

Our study for the first time describes the results of community based screening for the prevalence of the βs gene in different Rural Population within Central India. The practice of endogamy in India provides the rationale for the screening of individual populations to better understand the distribution of the βs gene and guide counseling and awareness programs.

Large scale screening as described in this study has been the foundation for regional disease control programs (6). While mass screening has provided detailed descriptions for the Rural Population in previous studies, such detailed descriptions are as yet unavailable for the Rural Population.

In dealing with a subject area as involved as sickle cell anemia, with its many medical, psychological, and sociological factors, one is faced with the need to selectively discriminate among vast quantities of associated literatures in an attempt to choose a review relevant to the subject area. Since a complete review of all literature was impossible within the scope of this study, the authors established certain criteria for literature selection, and a three-fold direction of inquiry was identified as being 'most appropriate to the needs of this study(7).

Sickle cell disease, in both its forms, is a genetically acquired condition. Inherited as an autosomal recessive, pattern, sickle cell anemia is the homogygous state of the abnormal hemoglobin S gene. The mating of two persons with the sickle cell trait would imply a 25 per cent probability that each offspring will inherit two abnormal genes (Hb-s),8 25 per cent probability that the offspring will inherit two normal genes, and a 50 per cent probability that the offspring will inherit one normal gene and one abnormal gene(8). In other words, based on statistical calculation using Mendelian model, a family consisting of four off spring will produce one offspring having sickle cell anemia, one offspring that is free of any sickle cell gene, and two offspring having one normal, and one abnormal gene

One, is most impressed in searching the literature for topics dealing with sickle cell anemia, by the dearth of material(9). Most literature approaching any other than a highly technical, medical model of sickle cell anemia, is practically nonexistent, and for that small amount that does exist, the publish dates are quite recent, usually within the last ten to fifteen years.

Conclusion

We provide for the first time detailed prevalence data of the βs gene among Rural Population ethnic groups in Central India. These population screening programs have uncovered previously undiagnosed cases, provided detailed information for population based disease counseling, prevention programs and comprehensive care programs. Over 5,000 individuals were identified with SCT and more than 1,000 individuals were identified with SCD through community-based and target high-risk screening. The population screening study shows how community-based screening can identify individuals who may not have otherwise sought care and guide individuals to available comprehensive care centers. The screening also helped identify specific sub-groups within the Rural Population that have higher prevalence rates of the βs gene. An estimated 1.1 million individuals in the state of Madhya Pradesh alone are expected to have SCT. Similar community-based screening programs need to be initiated throughout regions of India that have a high prevalence of Rural Population to determine the true prevalence of the βs gene in India.

In summary, there appeared to be a broad general knowledge of sickle cell disease in the identified. target samples, with many 1nteresting comparative responses to the attitudinal survey, but no statistically significant differences 1n black and white responses in western countries.

That all persons experiencing genetic disease have the option to choose genetic counseling and sufficient community education to know they have this choice. That all genetic counseling include supportive family therapy and education over a sufficient period of time so the family can integrate the information into their own life systems.

References

1. Balgir, R., Epidemiology, Population Health Genetics and Phenotypic Diversity of Sickle Cell Disease in India. The Internet Journal of Biological Anthropology, 2007. 1(2).
2. Shukla, R.N. and B.R. Solanki, Sickle Cell Trait in Central India. Lancet, 1985. 1: p. 297-298.
3. Kate, S.L. and D.P. Lingojwar, Epidemiology of Sickle Cell Disorder in the State of Maharashtra. . International Journal of Human Genetics, 2002. 2(3): p. 161-167.
4. Weatherall, D.J., The inherited diseases of hemoglobin are an emerging global health burden. Blood, 2010. 115(22): p. 4331-6.
5. Balgir, R.S., Spectrum of hemoglobinopathies in the state of Orissa, India: a ten years cohort study. J Assoc Physicians India. , 2005. 53: p. 1021-1026.
6. Embury, S.H., et al., Sickle Cell Disease: Basic Principles and Clinical Practice. 1994.
7. Eaton, W.A. and J. Hofrichter, Sickle cell hemoglobin polymerization. Adv Protein Chem, 1990. 40: p. 63-279.

8. Costanzo, V.L., Sickle Cell Trait Counseling for Student Athletes, in Human Genetics 2011, University of Pittsburgh.

9. Kaur, M., G.P. Das, and I.C. Verma, Sickle cell trait & disease among tribal communities in Orissa, Madhya Pradesh & Kerala. Indian J Med Res, 1997. 105: p. 111-6.