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A Study of Socio-Demographic Profile of Thalassemia Major Patients Under Treatment at A Tertiary Care Centre in Western Gujarat

KEYWORDS: Thalassemia, hemoglobinopathies, communities

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ABSTRACT *Thalassemia Major is one of the most commonly prevalent Hemoglobinopathies in the world, and especially in our part of the country. Earlier, Thalassemia was considered a part of pediatrics only, as only exceptional patient used to survive to adulthood, but now with advanced management protocols and multi-disciplinary approach, more and more patients are seen by physicians. This study aims to evaluate the socio-demographic profile of thalassemia major patients surviving into adulthood so as to orient the physicians and the general practitioners to adult thalassemia patients*

INTRODUCTION

Thalassemia is an autosomal recessive disease. As per WHO estimate, 4.5% of world populations are carriers of hemoglobinopathies. The largest concentration of thalassemia patients are seen in South East Asia, Sri Lanka, Bangladesh, North West India, Pakistan, Middle East Countries, Greece, and Italy. In South East Asia, there are 40 million carriers of thalassemia gene, 50% of which are in India alone. The mean prevalence of the carrier status in India is 3.3% (ranging from 1 to 17% in various communities). If a line is drawn between Mumbai and Kolkata on the Map of India, the region above the line has an incidence of 3 to 17%, whereas region below the line has incidence of less than 3%. It is estimated that every year approximately 1 00 000 children with thalassemia major are born all over the world. With the birth rate of 22.8/1000 in India, it is estimated that there are about 9000 to 10000 cases being added every year. The management of thalassemia major in a developing country poses a major challenge to the Government Health Services. Lack of resources and coordination hampers the treatment of this multidisciplinary problem in a variety of ways. Awareness and availability of antenatal diagnosis is limited. Thalassemia major has long remained in the domain of pediatricians but advances in medical science and improved health care have contributed to the longevity of life and its quality; to the extent that it now needs attention of the adult physicians too. As of today, most physicians do not have to encounter thalassemics as their patients. Moreover, knowledge and management issues relating to thalassemia are unknown to most physicians.

There is a severe resource data crunch at a time when information needs to be readily available to the physicians, the emphasis being on "evidence-based medicine."

This study included thalassemia major patients above the of 12 years (considered as adults), and 100 such patients (n=100) were enrolled who were under treatment at our hospital.

OBSERVATIONS:

The age distribution of the patients under study was as shown in Table-1. The age group composition of the sample showed a declining proportion with increasing age, thus showing a reduced life Expectancy in the adult Thalassemics. The proportion of patients above 20 years of age was very low (23%), with only 2 patients were above the age of 30 years, while the maximum (40%) comprised of 12-15 yrs age group, followed by 37% in the 16-20 years age group. Out of these, 60% were males, while females comprised 40% of the patients.

TABLE – 1
AGE DISTRIBUTION

AGE	Percent
12-15	40.00%
16-20	37.00%
21-25	17.00%
26-30	4.00%
>30	2.00%
Total	100.00%

As already known, thalassemia preferentially affects certain castes, so we studied the caste-wise distribution of these patients, Hindus comprising of 82%, while muslims were 18%. The detailed caste-wise distribution of these was as shown in Table-2.

TABLE – 2
CASTE-WISE DISTRIBUTION

CASTE	Percent
AAHIR	9.76%
BHANUSHALI	17.07%
BHARWAD	7.32%
BRAHMIN	2.44%
CHAMAR	6.10%
GANDHAR	1.22%
HARIJAN	19.51%
JAIN	1.22%
KODI	1.22%
LOHANA	1.22%
MEGHWAD	1.22%
PATEL	4.88%
PINJARA	1.22%
PRAJAPATI	1.22%
RAJPUT	4.88%

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SATVARA	13.41%
SINDHI	4.88%
TAMAR	1.22%
TOTAL	100.00%

From our knowledge of thalassemia, it is known that for a child to be a thalassemia major, both the parents need to be thalassemia Minors, and one of the many factors responsible for the selective affection of Thalassemia to a certain communities is the practice of Consanguineous Marriage. In our study, 19 patients had a history of consanguineous marriage, wherein the parents were among the first degree relatives. Also there is increased affection of Siblings among the Thalassemics, as a result of which, once a child is affected, the parents are advised to undergo prenatal diagnosis for subsequent pregnancies, if any. And we also studied the correlation of these two variables, that does consanguineous marriage increase the incidence of other sibling being affected. From the table it is evident that 19% of patients had the history of consanguineous marriage and 27% had the other sibling affected, and when the correlation among the two was found, of the 27 patients who had other sibling affected, 10 had history of consanguineous marriage, and of the 73 patients who did not have other sibling affected, the prevalence of consanguineous marriage was in 9 patients only. With a CHI SQUARE value of 7.8187 and a p-VALUE of 0.0051, *this correlation was found to be statistically significant.*

TABLE – 3
PREVALENCE OF CONSANGUINEOUS MARRIAGE AND SIBLING AFFECTION

CONSANGUINEOUS MARRIAGE	OTHER SIBLING AFFECTED					
	Yes	Percent	No	Percent	TOTAL	Percent
Yes	10	52.60%	9	47.40%	19	100.00%
No	17	21.00%	64	79.00%	81	100.00%
TOTAL	27	27.00%	73	73.00%	100	100.00%
CHI SQUARE = 7.8187, P VALUE = 0.0051						

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Because of the considerable morbidity imposed by thalassemia and the need for regular blood transfusions, the academic performance of these patients is severely impaired. The level of education among them was low as compared to the normal population, with 68% of patients having Primary or lesser Education, and only 4% of them having Graduation or above.

TABLE – 4
LEVEL OF EDUCATION

LEVEL OF EDUCATION	Percent
PRIMARY	68.00%
SECONDARY	28.00%
GRADUATE AND ABOVE	4.00%
Total	100.00%

Also, considering the annual family income (AFI), the prevalence of Thalassemia among the affluent class was seen to be low, with 84% of the patients falling into the group with AFI less than 5 lacs per annum. And only 4% of patients falling in the income group of above 10 lacs per annum.

CONCLUSIONS:

Thalassemia major patients contribute an increasing population of adult patientsseen by a physician, and thus proper knowledge regarding the various socio-demographic features help a long way in better management of these patients as well as for prevention of thalassemia by screening in high risk populations and premarital and/or prenatal counseling and screening.

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