Clinico-Epidemiological Study of Patients Suffering from Sickle Cell Anaemia In a District Level Private Hospital of a Tribal District in South Gujarat

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Abstract:

Introduction: Sickle cell disorder is an autosomal recessive condition, in which inheritance of an affected gene from both parents results in a disorder. According to hospital based epidemiological survey, the prevalence of sickle cell gene is observed among tribal population, which accounts for 15 % of the total population of Gujarat and are distributed in various districts. **Objectives:** To study clinical profile, epidemiological characteristics and socio-demographic features of persons having sickle cell disease and sickle cell trait and establish relationships if any of the epidemiological profile with the clinical and socio-demographic features of the study subject. **Method:** The study was carried out in Janak Smarak hospital in Vyara Town, Tapi District. It was a descriptive cross-sectional study for a period of one year and minimum of 75 persons with sickle cell disease and 150 persons with sickle cell trait was taken for the study. **Results:**Proportion of females affected were more than that of the male. Majority of the participants belonged to Gamit caste and most of them were Hindus. Majority of the participants were married and it was significantly associated with Sickle cell status of the individuals. Majority of sickle cell disease patients had clinical manifestations of sickle cell crisis like pain in both upper and lower limbs; weakness, fever and body ache and sign of pallor. Nearly half of the sickle cell disease patients had a history of blood transfusion.**Conclusion:** Family history and predisposition happens to remain one of the most important predictors of the Sickle cell status among the population. Nearly more than half of the Sickle Cell Disease patients needed regular transfusion with Blood or any of its components. This points out the necessity of establishing these units in higher numbers in tribal areas.

Keywords: Blood transfusion, Sickle Cell Crisis, Sickle Cell Disease, Sickle Cell trait, Tribal Community

Introduction:

Sickle Cell Disease (SCD) is genetic blood disorder that affects the Haemoglobin in Red Blood Cell. Sickle cell disorder is an autosomal recessive condition, in which inheritance of an affected gene from both parents results in a disorder while inheritance of one abnormal gene results in a healthy carrier.[1] It is characterized by vaso-occlusive pain crises, risk for pneumococcal infections, acute chest syndrome, and stroke and organ failure and is associated with substantial morbidity and premature mortality.[2] The main reasons for mortality among sickle cell disease patients are infection, acute splenic sequestration, severe anaemia and haemolytic crisis.[1]

According to hospital based epidemiological survey, the prevalence of sickle cell gene is observed to be 0-18 percent in northern- eastern India, 0 to 33 percent in western India, 22.5 to 44.4 percent in central part of India and 1 to 40 percent in southern India and the gene frequency of Hb-S varies between 0.031- 0.41. [3]Another survey done by the Indian Red Cross Society in Gujarat, where nearly 1, 68,498 tribal individuals from 22 districts were screened, the overall prevalence of sickle cell carriers was found to be 11.37 per cent.[4, 5] Some studies have also documented high prevalence of sickle gene in various tribal communities of Gujarat that include Bhils and Dhodias of Panchmahal, Dublas, Naikas, Koli, Dhanka, Gamit, Vasava, Bariya, Varli, Vaghari, Kukna, Halpati, & Chaudhari. Tribal population accounts for 15 % of the total population of Gujarat and are distributed in various districts of the state such as Sabarkantha, Banaskantha, Panchmahal, Vadodara, Narmada,

Bharuch, Surat, Valsad, Dang and the surrounding UTs like Div-Daman. [6]In Tapi District of Gujarat, Sickle Cell Disease is more prevalent among the socio-economically disadvantaged and medically underserved communities.

The study was carried out in Janak Smarak Hospital, a trust managed hospital located in Vyara where all diagnostic and treatment facilities for this condition are available to know the epidemiological and clinical pattern of patients reported with this inherited disorder.

Objectives of the study:

1. To study clinical profile of persons having sickle cell disease and sickle cell trait.
2. To study the epidemiological characteristics and Socio-demographic features of persons having sickle cell disease and sickle cell trait
3. Establish relationships if any of the epidemiological profile with the clinical and socio-demographic features of the study subjects

Method:

The study was carried out in Janak Smarak hospital in Vyara Town, Tapi District, Gujarat. It was a descriptive cross-sectional study between March 2014 to February 2015 i.e. for a period of one year. After getting permission from the institutional ethics committee of Sumandeep Vidhyapeeth, Piparia, persons having sickle cell disease or sickle cell trait (diagnosed at Janak Smarak Hospital, Vyara) were contacted to explain the purpose of the study and nature of his / her participation and to collect the basic personal and epidemiological information, clinical information like symptomatology, History of blood transfusion, Family history of Sickle cell Disease & Sickle cell Trait, after getting their written consent for participation in the study. Only those who consented for the study were included as subjects to collect the data further. Prior permission from the hospital authorities was also sought to carry out the study.

For estimation of sample size, a small pilot study was carried out between 1st April 2013 to 30th September 2013. During this period of 6 months, a total of 38 patients were diagnosed with Sickle cell Anemia and 164 individuals were diagnosed having sickle cell trait. This data was then compared with the past records of 3 years of the hospital and it was observed that the no. of sickle cell patients and traits who had reported to the hospital were almost similar in number, hence it was assumed that for a forthcoming period of one year a minimum of 75 persons with sickle cell disease and 150 persons with sickle cell trait would be available for study at the hospital. This was hence taken as the sample size for the study.

The study was carried out using a pre-designed and pre-tested questionnaire and the data was collected by directly interviewing the subjects and examining them.

The data collected was analyzed using MS Excel 2007 and suitable statistical tests were applied at 5% level of significance.

Results:

Table 1 suggests that proportion of females affected were more than that of their male counterparts. Majority of the participants belonged to Gamit caste and most of them were Hindus. Majority of the participants were married. Majority of subjects among both males and females were educated up to secondary with more proportion of illiterate among females was more than males. Most of the patients were from Social Class 1 and 2.

Table 2 shows that the prevalence of the SCD and Sickle Cell Trait (SCT) was higher in age groups above 18 yrs of age. However, the prevalence of both SCD & SCT was almost 30% in those aged <17 yrs. It was also noteworthy that the females were affected more than the males in both the groups. To assess the impact of Social Demographic variable with SCD & SCT, all of the socio demographic variables were analysed & association was sought using chi squared test which showed that out of all the variables, only marital status was significantly associated with Sickle cell status of the individuals. (X^2value= 17.68; $p<0.001$)

Table 3 shows that Majority of sickle cell Disease patients had clinical manifestations of sickle cell crisis like pain in both upper & lower limbs (58%); weakness (53%), fever (48%) and body ache (44.3%). Where as in sickle cell trait patients, it was

Table 1: Socio demographic profile of the subjects

Variable	Female (n=132)		Male (n=93)		Total (n=225)	
Age (Years)	**Number**	**Percentage (%)**	**Number**	**Percentage (%)**	**Number**	**Percentage (%)**
1 – 5	4	3.03	2	2.15	6	2.66
6-17	33	25	34	36.55	67	29.78
18-45	81	61.37	48	51.61	129	57.34
46- 100	14	10.60	9	9.67	23	10.22
Caste						
Gamit	99	75	66	70.96	156	69.33
Chaudhari	39	29.54	23	24.73	62	27.55
Others*	3	2.27	4	4.30	7	3.11
Marital Status						
Married	72	54.54	34	36.55	106	47.74
Unmarried	21	15.90	25	26.88	46	20.72
Widow/widower	4	3.03	0	0	4	1.77
NA(1-17 years of age)	35	26.51	34	36.55	69	30.66
Religion						
Hindu	123	93.18	86	92.47	209	92.88
Christian	9	6.81	7	7.52	16	7.11
Education						
Illiterate	25	18.93	7	7.52	32	14.22
Primary (1-7 standard)	32	24.24	33	35.48	65	28.88
Secondary (8-10 standard)	40	30.30	23	24.73	63	28
Higher Secondary (11-12 standard)	18	13.63	10	10.57	28	12.44
Diploma/ ITI / PTC	1	0.75	1	1.07	2	0.88
Graduate	12	9.09	15	16.12	27	12
PG/Diploma	2	1.51	2	2.15	4	1.77
NA	2	1.51	2	2.15	4	1.77
Occupation						
Student	53	40.15	50	53.76	103	45.77
Housewife	67	50.75	0	0	67	29.77
Farmer	6	4.54	24	25.80	30	13.33
Job	0	0	5	5.37	5	2.22
Teacher	5	3.78	5	5.37	10	4.44
ANM	1	0.75	0	0	1	0.44
Shopkeeper	1	0.75	0	0	1	0.44
Driver	0	0	1	1.07	1	0.44
NA**	2	1.51	2	2.15	2	0.88
Social Class* **						
CLASS -I	42	31.8	31	33.3	73	32.4
CLASS - II	77	58.3	50	53.8	127	56.4
CLASS - III	12	9.1	9	9.7	21	9.3
CLASS - IV	0	0	2	2.2	2	0.9
CLASS - V	1	0.8	1	1.1	2	0.9

* "Others" include Valvi and Kokani castes. ** NA suggest children less than 18 years of age (out of school)

*** Social class as per Modified B.G. Prasad`s Classification of 2015 (AICPI= Rs. 816/-)

Table 2: Association of various Socio-Demographic Variables with SCD & SCT status of the subjects

Variable	SCD (n=75)		SCT (n=150)		Chi –Square
Age (Years)	**Number**	**Percentage (%)**	**Number**	**Percentage (%)**	
1 – 5	3	4	4	3	χ^2 value=0.418, df=2, p=0.8112
6-17	21	28	46	31	
18-45	51	68	78	52	
46- 100	0	0	22	14	
Sex					
Female	41	55	91	61	χ^2 value=0.515, df =1, p=0.4728
Male	34	45	59	39	
Gamit	53	71	103	69	χ^2 value=0.094, df =1, p=0.7591
Chaudhari	22	29	42	28	
Others*	0	0	5	3	
Marital Status					
Married	27	36	79	53	χ^2 value=17.682, df =2, p=0.000145
Unmarried	29	39	17	11	
Widow/widower	0	0	4	3	
NA** (1-17 years of age)	19	25	50	33	
Religion					
Hindu	68	91	141	94	χ^2 value=0.036, df =1, p=0.8499
Christian	7	9	9	6	
Education					
Illiterate	9	12	23	15	χ^2 value=2.894, df =4, p=0.5757
Primary (1-7 standard)	24	32	41	27	
Secondary (8-10 standard)	17	23	46	31	
Higher Secondary (11-12 standard)	11	15	17	11	
Diploma/ ITI / PTC	0	0	2	1	
Graduate	12	16	15	10	
PG/Diploma	1	1	3	2	
NA	1	1	3	2	
Occupation					
Student	43	57	60	40	χ^2 value=6.738, DF=5, p=0.2408
Housewife	18	24	49	33	
Farmer	7	9	23	15	
Job	3	4	5	3	
Teacher	2	3	8	5	
ANM	1	1	0	0	
Shopkeeper	0	0	1	1	
Driver	0	0	1	1	
NA	1	1	3	2	
Social Class* **					
CLASS -I	20	27	53	35	χ^2 value=9.38, DF=3, p=0.0245
CLASS - II	43	57	84	56	
CLASS - III	8	11	13	9	
CLASS - IV	2	3	0	0	
CLASS - V	2	3	0	0	

* "Others" include Valvi and Kokani castes.

** NA includes (1-17 years of age).

Table 3 : Clinical Manifestation at the time admission of sickle cell disease & sickle cell trait patient

Symptoms	SCD (n=75)		SCT (n=150)		Total (n=225)	
	Number	Percentage (%)	Number	Percentage (%)	Number	Percentage (%)
Weakness	40	53	31	21	71	32
Fever	36	48	47	31	83	37
Body ache/ Joint pain/ Back pain	33	44.3	110	74	156	69
Pain in both upper & lower limb	43	58	48	32	91	40.1
Breathlessness/ Chest pain/ Coughing	17	23	26	17	43	18
Giddiness/ Headache / Uneasiness	7	9.3	18	12	25	11
Abdominal pain/Vomiting	16	21	1	0.6	17	7.4
Signs						
Pallor	44	59	109	73	153	68
Icterus	10	13	20	13	30	13
Splenomegaly/						
Hepatomegaly	19	26	1	1	20	9
Oedema	2	3	20	13	22	10

*Many patients had multiple symptoms & Signs

seen that most of the patients had clinical manifestation of Body ache/Joint pain/ Back Pain 74 % followed by pain in both upper & lower limb 32%; fever 31% and weakness 21%.

Table also shows that, majority of both the Sickle Cell Disease and Sickle cell trait patients showed sign of pallor; followed by Splenomegaly/hepatomegaly in Sickle cell Disease patients & Icterus and Edema in the Sickle cell trait patients.

Figure 1: Family history of sickle cell disease & sickle cell trait among the patients

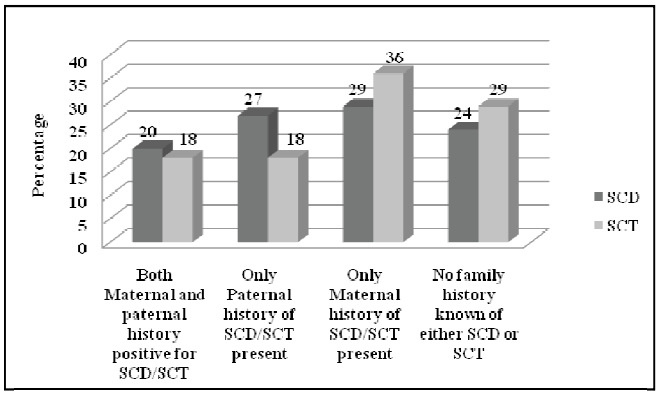
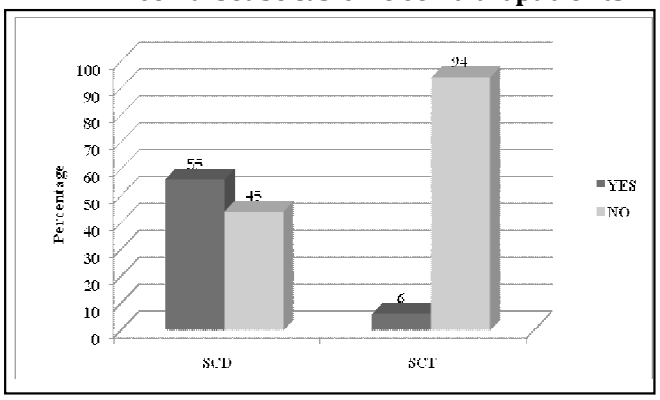


Figure 1 shows that nearly 30% of Sickle cell trait individuals and 25% of sickle cell disease patients had no family history of any sickle cell status, which implies that majority of them did have either a one parent or two parent history of sickle cell disease/trait. Nearly 20% had a two parent history and the remaining had only one parent history of SCD/trait. This clearly shows the family preponderance in the transmission of the sickle cell status.

Figure 2: History of Blood transfusion of sickle cell disease & sickle cell trait patients



It can be seen from Figure 2 that nearly half of the sickle cell disease patients and a very small proportion of sickle cell trait individuals had a history of blood transfusion. This clearly shows the importance of having blood storage centres and blood banks in more numbers in tribal districts as it can be a life-saving tool among the affected individuals. The requirement of blood transfusion among sickle cell disease patients was significantly higher as compared to the sickle cell trait patients and the same was statically significant using the chi squared test. ($\chi 2$ Test value=71.3, DF=1, $p<0.01$)

Discussion:

The present study was carried out during in Janak Smarak hospital of Vyara. A total of 225 participants, 75 of which were patients of Sickle Cell Disease & the remaining 150 had Sickle Cell Trait were included in the study. It was observed that proportion of females affected was more than that of their male counterparts which could have been because a higher sex ratio of Tapi district (1004 females per thousand males as per census 2011) [7]. A study conducted by Shrestha A et al [8] and Jain B et al [9] found that males were more affected than females.

Majority of participants belong to Gamit caste, followed by Chaudhari caste because of a higher population of these communities residing in the district (Data of Census 2011)[7] The present study revealed that marital status was significantly associated with occurrence of SCD & SCT status. This can lead to higher chances of future transmission of sickle cell in the progeny due to higher prevalence of consanguineous marriages in the tribal communities. A study by M Kamble et al [10] also describes about history of consanguinity marriage; out of which (8.2%) had sickle cell disease and (5.2%) had sickle cell trait. It is hence extremely necessary to carry out genetic counselling after knowing sickle cell status among potential couples before marriage to avoid the further transmission in the successive generations.

Present study showed that over all literacy rate was 84% while 14% patients were illiterate, which was almost similar to the census data for the state and the district. A study by Gustafson SL et al [11] described that the a higher the level of knowledge of Sickle Cell Disease, there is better acceptance of genetic counselling and testing as was primarily found among the high school graduates. The more educated people are the better is the access to health services and more is the acceptance of IEC related to the disease.

Current study showed that the clinical manifestations of fever & pain in both upper & lower limb (hand foot syndrome) was among the most common symptoms in the study population A study by K Swarnkar [11] also showed similar results. Therefore, it is prudent that the affected individuals must be made aware of these alarming symptoms requiring admission and immediate treatment so that their lives can be saved. Our study revealed that pallor was the commonest sign in both SCD and SCT. A study by Patra Pradeep et al [12] and a study by K Swarnkar [11] also show that pallor was common sign in both SCD & SCT trait patients.

Current study also observed that about 29% of the Sickle Cell Disease patients & 36 % of the Sickle Cell Trait patients had a positive maternal history of SCD/SCT.A Study by Warade J et al [13] also showed a positive family history for either sickle cell disease or sickle cell trait and thus how showed the impact of family history over the occurrence of disease. The present study revealed that majority of sickle cell disease & 6% of sickle cell trait patients had history of blood transfusion. The requirement of blood transfusion is extremely critical and must be planned at tribal areas in secondary and tertiary care hospitals to ensure higher quality of life and better treatment facilities for the affected.

Conclusion:

It can be concluded from the study that younger generation of the community especially females were more affected with both sickle cell disease and trait. Most common clinical manifestations among the subjects include Fever & Pain in both upper & lower limb (hand foot syndrome) and also the joints. Family history and predisposition happens to remain one of the most important predictors of the Sickle cell status among the population thereby reiterating the need of regular Genetic Counseling at hospitals (Both Govt and Private) in Tribal Areas of the state. Nearly more

than half of the Sickle Cell Disease patients needed regular transfusion with Blood or any of its components. This points out the necessity of establishing these units in higher numbers in tribal areas.

Recommendations:

It is a well-known fact that sickle cell disease is an incurable genetic disease and hence prevention of the disorder stays as the mainstay of its control in the community settings. It is necessary to educate the tribal communities about the disease & its prevention. Endogamy marriages among the sickle cell trait patients increase the chances of the occurrence of the disease as the subsequent generation has a higher chance of contracting sickle cell disease and thus emphasizes the need of regular genetic counselling sessions in hospitals and in the community to ensure that more and more people can be protected from the disease and its dangerous repercussions.

Studies of similar types especially in tribal areas can help in generating and strengthening evidence for availability of special services to the affected including Genetic counselling, screening and treatment services including availability of blood units on a priority basis.

Declaration:

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Conflict of Interest: Nil

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