

Original Research Article

Epidemiological and clinical study of sickle cell disease at tertiary care centre in Western India

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Received: 10 October 2019

Revised: 15 November 2019

Accepted: 20 November 2019

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ABSTRACT

Background: Sickle cell disorders are structural hemoglobinopathies, rendering red blood cells sickle shaped, less deformable and sticky leading to microvascular vaso-occlusion and premature red blood cells destruction which leads to varied clinical manifestations. It leads to lifelong morbidity thus affecting quality of life and contributes to early mortality thereby reducing the key national resources- the healthy workforce. This study was done to evaluate epidemiological and clinical profile of sickle cell disease attending the centre.

Methods: This study was cross-sectional, observational study conducted at tertiary care hospital in Gujarat. After taking ethical clearance patients were enrolled as per inclusion and exclusion criteria and epidemiological and clinical profile of sickle cell disease patients was studied.

Results: Mean age of sickle cell disease was 22.58 years. It was found in tribal communities of Gujarat like Rathwa, Baria, Tadvi etc. Commonest symptom was musculoskeletal pain (86.84%), followed by jaundice (71.05%), fever, dyspnoea, abdominal pain and chest pain. Most common systemic manifestation was pain crises (60.66%), followed by hemolytic anemia (31.15%), acute chest syndrome (30%), consolidation (11.67%), hepatopathy (10%) and avascular necrosis of hip. (6.56%).

Conclusions: Sickle cell disease is seen in younger patients. In Gujarat mainly tribal communities are affected. Major systemic manifestations of sickle cell disease include pain crisis followed by hemolytic crisis, acute chest syndrome, hepatopathy and AVN of hip.

Keywords: Acute chest syndrome, Epidemiology, Hemoglobinopathies, Haemolytic anemia, Pain crises, Sickle cell disease

INTRODUCTION

Hemoglobinopathies are disorders affecting the structure, function, or production of hemoglobin. These conditions are usually inherited and range in severity from asymptomatic laboratory abnormalities to death in utero. Different forms may present as hemolytic anemia, erythrocytosis, cyanosis, or vaso-occlusive stigmata.

Sickle cell disorders are structural hemoglobinopathies occurs when mutation alter the amino acid sequence of a

globin chain, altering the physiological properties of the variant hemoglobin and producing the characteristic clinical abnormalities. This variant hemoglobin polymerizes abnormally rendering red blood cell less deformable, sticky and altered shape like a sickle and abnormally adhere to endothelium of small venules.

These abnormalities provoke unpredictable episodes of microvascular vaso-occlusion and premature RBC destruction (hemolytic anemia). Prominent manifestations include episodes of ischemia pain and

ischemic malfunction or frank infarction in the spleen, central nervous system, bones, liver, kidneys and lungs.¹

In Gujarat, the Dhodia, Dubla, Gamit, and Naika tribes have a high prevalence of HbS (13-31%).² More recently very extensive population surveys have been done by the Indian Red Cross Society, Gujarat State Branch where 1,68,498 tribals from 22 districts were screened and the overall prevalence of sickle cell carriers was 11.37 per cent.³ National programme are being run in Gujarat to enlarge the epidemiological database, establishing special screening programmes for population at risk, impart genetic counselling and established special centres for treatments.

Aim of the study was to evaluate epidemiological and clinical profile of sickle cell disease attending the centre.

METHODS

It was a cross sectional, observational study. Study sample was taken from sixty patients admitted in medical wards of Shri Sayaji General hospital, Vadodara over a period from May 2009 to September 2011. Shri Sayaji General hospital, Vadodara covers a lot of patients from Vadodara city itself as well as large section of population from Vadodara district with higher tribal population from Chotaudaipur, Kawant, Naswadi, Dahod as well as the nearby district of Narmada, Bharuch, Panchmahal.

Inclusion criteria

- Patients who were found to have sickling test positive and had dominant sickle haemoglobin HbS with absent HbA or higher proportion of HbS than HbA and HbF on hemoglobin electrophoresis or,
- Patients who were known case of Sickle cell disease and related disorders and electrophoresis was done in past and admitted for different ailments in SSG hospital were included in the study.
- Patients who were more than 12 years of age.

Exclusion criteria

- Patients with sickle cell trait were excluded from this study.
- Patients less than 12 years of age.

Detailed clinical history and physical examination was done.

Investigations

Routine investigations

Routine investigations were carried out in all the individuals. This includes complete hemogram, peripheral smear examination, complete urinalysis, biochemical tests, ultrasonography of the abdomen, thin

peripheral smear, X ray chest and X ray of joints (as required) were done in all the patients.

Tests for diagnosis of sickle cell disease

Stained thin peripheral smear to see sickle shaped red blood cells which may occur in sickle cell disease and sickle cell β -thalassemia. The morphologic features of accelerated erythropoiesis like polychromatophilia, basophilic stippling, and normoblastosis were looked for.

Sickling test with sodium meta bisulphite test was done in all patients.

Hemoglobin electrophoresis was done on cellulose acetate membrane which is simple, rapid and sensitive method.

From history, clinical examination, hematological and biochemical investigation and radiological investigations major systemic manifestations were searched in our patients.

Data was entered and analysed using excel software. Descriptive statistics like frequency and percentage was applied.

RESULTS

In the present study total number of patients with sickle cell disease were 60. Amongst these total number of patients with sickle cell anemia were 38 and with sickle beta thalassemia were 22. In this study no other sickle cell syndromes could be found.

In this study maximum number of patients were in age group 12-30 years (51 patients-85%). But after 30 years of age only 8 patients of sickle cell disease were seen (13.33%) (Figure 1). Mean age of sickle cell anemia was 22.58 years. In this study total number of males with sickle cell anemia were 33(55%) and females were 27(44.26%) (Figure 2).

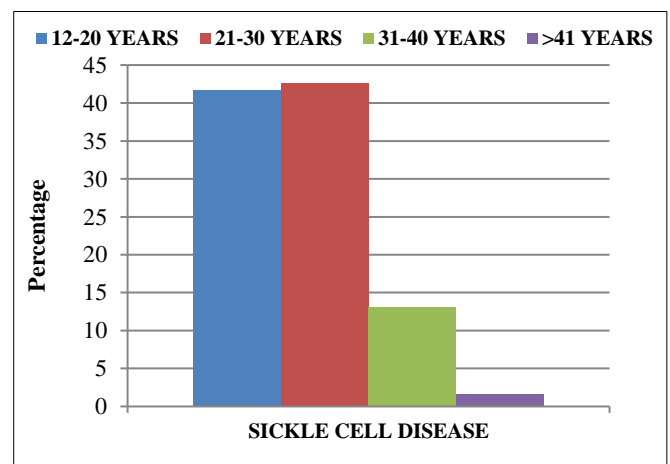


Figure 1: Age distribution in study population.

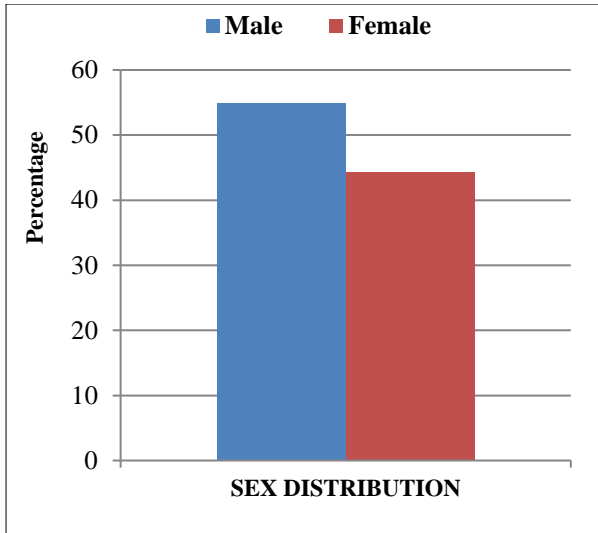


Figure 2: Sex distribution in study population.

Table 1 shows maximum number of patients belong to Rathwa caste (15 patients - 24.59%), followed by Bariya caste (12 patients - 19.67%), Tadvi caste (7 patients - 11.48%), Solanki caste (4.92%), Parmar caste (4.92%), Bhil caste (4.92%), Vasava caste (3.28%), Damor caste (3.28%), Muslim (1.64%). Maximum number of patients were from Vadodara, about 34 patients (56.67%), followed by 9 patients from Panchmahal (14.75%), 7 patients from Dahod (11.48%), 4 patients from Narmada (6.56%), 2 patients from Anand (3.28%), 2 patients from MP (3.28%) (Table 2).

Table 1: Ethnic group distribution.

Ethnic group	Total	Percentage (%)
Rathwa	15	24.59
Bariya	12	19.67
Tadvi	7	11.48
Soalnki	3	4.92
Vasava	2	3.28
Parmar	3	4.92
Vankar	0	0
Bhil	3	4.92
Muslim	1	1.64
Damor	2	3.28
Others	12	19.67

Table 2: Area distribution.

Area	Total	Percentage (%)
Vadodara	34	56.67
Panchmahal	9	14.75
Dahod	7	11.48
Narmada	4	6.56
Madhya Pradesh	2	3.28
Anand	2	3.28
Others	2	3.28
Total	60	100

Commonest symptom in patients with sickle cell disease of musculoskeletal pain (51 patients - 83.61%), followed by jaundice (41 patients - 67.21%), fever (40 patients - 65.57%), dyspnea (33 patients - 54.1%), abdominal pain (29 patients - 48.34%) and chest pain (19 patients - 31.67%) (Figure 3).

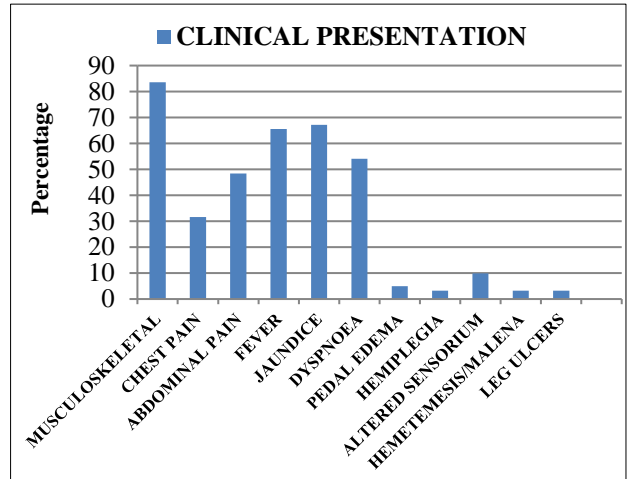


Figure 3: Clinical presentation of study population.

Past history of admission was positive in 51 patients (85%) overall, while 9 patients (15%) had no history of previous admission to hospital. Overall 48 patients (80%) were admitted for 1-4 times and 3 patients (5%) admitted for 5-10 times (Table 3).

Past history of pain crises was seen in 44 patients (73.33%) overall. Past history of jaundice was seen in 38 patients (63.33%) overall, while 19 patients (31.67%) had a history of blood transfusion (Table 3).

From history, clinical examination, hematological and biochemical investigation and radiological investigations major systemic manifestations were searched in patients.

Figure 4 shows most common systemic manifestation was pain crises (37 patients - 60.66%), followed by hemolytic anemia (19 patients - 31.15%), acute chest syndrome (18 patients- 30%), consolidation (7 patients - 11.67%), hepatopathy (6 patients - 10%), avascular necrosis of hip (4 patients - 6.56%).

Table 3: Number of admissions in past, past history of jaundice and blood transfusion.

Past history	Total	Percentage(%)
Past admission per year		
None	9	15
1-4 Times	48	80
5-10 Times	3	5
Past history of		
Pain crisis	44	73.33
Jaundice	38	63.33
Blood transfusion	19	31.67

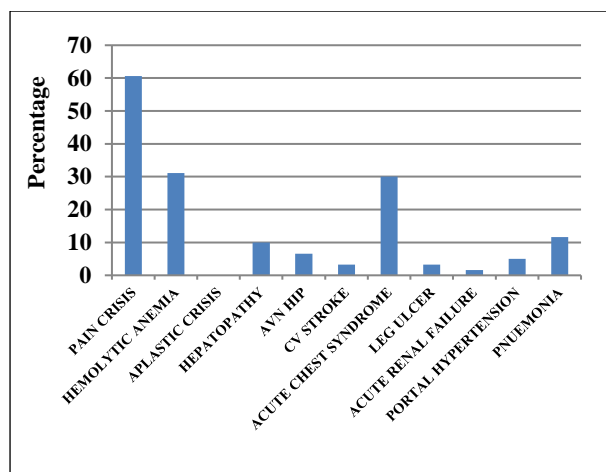


Figure 4: Analysis of major systemic manifestations in study population.

DISCUSSION

The haemoglobinopathies are inherited disorders of blood and are one of the major public health problems in India. These diseases are making important demands on health resources in developing countries. Present study was done to evaluate epidemiological and clinical profile of patients with sickle cell disease.

In present study mean age of patients overall was 22.58 years and maximum number of patients were in age group 12-30 years (51 patients-85%).

Study by Ibadapo in Nigeria comprising of adolescents and adults, the mean age was 20.5 years.⁴ Kar et al BC observed that the majority of sickle cell disease die at the age of 20 years.⁵

S. Diop et al, studied 108 patients of homozygous sickle cell disease in Senegal, showed a mean age of 27 years (20-51years). In their study 20-29 years were represented by 67.5% of patients, 30-39 years by 26.9% and 5.6% were above 40 years of age.⁶

The mean age of death is rising in Western countries because of improved awareness, better socioeconomic conditions, good standards of living and easy access to medical care and knowledge of the disease. But in India, because of lack of awareness, high child mortality still exists.

Male preponderance was observed. Ibadapao found male to female ratio of 1.5 in Nigerian adults.⁴ On chromosome Xp22, there is gene for the HbF production, so females have more HbF, which ameliorates sickling and many clinical features. In all studies comprising of later age groups, there is a female preponderance. While males have a greater preponderance for painful crisis at age of 15 to 20 years, which is not seen in females.

In this study maximum number of patients belong to Rathwa caste (15 patients - 24.59%), followed by Bariya caste (12 patients - 19.67%), Tadvi caste (7 patients - 11.48%), Solanki caste (4.92%), Parmar caste (4.92%), Bhil caste (4.92%), Vasava caste (3.28%), Damor caste (3.28%), Muslim (1.64%).

According to ICMR, Bhils (29%), Dhodia (31%), Dubla (9.5%), Naikas (31%), Gamits (31%), Kolis (20.5%), Dhanakas (20%), vasavas (27.6%), Bariyas (28%), Chaudhary (28%), Tadvi (27%) and Rathwas (28.4%) are among the major tribes having Sickle Cell problem in Gujarat. Kar et al, BC report that the sickle gene is widely prevalent in India. It has penetrated different castes and communities in India, despite of its origin from the tribal belt.⁵

Vadodara dominance in this study could be accounted by the fact that patients from tribal area are now moving to nearby cities for better job opportunities and health care facilities. Tribal accounts 15% of the total population of Gujarat and distributed in various districts Sabarkantha, Banaskantha, Panchmahal, Vadodara, Narmada, Bharuch, Surat, Valsad, Dang and Div-Daman.³

In this study commonest symptom in patients with sickle cell anemia was musculoskeletal pain, followed by jaundice. In a study by Ibadapo et al, in Nigeria, includes adolescents and adults, pain was noted in 57.5% patients.⁴ Gupta and Yadav, found that 40% had severe pain in bones and joints, while 36% had recurrent fever. Abdominal pain was seen in 16% of patients.⁷

Past history of pain crisis was seen in 73.33% patients overall. Past history of jaundice was seen in 63.33% patients overall, while 31.67% patients had a history of blood transfusion. Diop et al, study in Senegal, patients were more than 20 years of age, 96.3% had more than one vaso-occlusive event per year, and 26.9% had received a blood transfusion.⁶

In this study the most common systemic manifestation was pain crises (37 patients - 60.66%), followed by hemolytic anemia (19 patients - 31.15%), acute chest syndrome (18 patients- 30%), consolidation (7 patients - 11.67%), hepatopathy (6 patients - 10%), avascular necrosis of hip (4 patients - 6.56%) In a study of 123 patients of SCA by Serjeant GR et al, in Jamaica, 83% had painful crises, 24% had pneumonia, 8% had priapism and 12% had aplastic crisis.⁸

Acute chest syndrome was seen in 30% in study population. A study by Castro O et al, shows risk factors for acute chest syndrome, which is associated with high hemoglobin level, low HbF levels, early age (maximum prevalence in age group 2-4 years and lesser in adults). Castro et al shows the presence of ACS is an independent risk factor for early mortality even when corrected for by pain crisis.⁹

In this study hepatopathy was seen in 10% of patients. Because of sustained increase in heme catabolism, pigmentary gallstones in sickle cell disease is high, which also increase episodes of cholecystitis and cholelithiasis.

Haber Kern CM et al, showed a progressive age-related increase in prevalence of gall stones from 12% in 2-4 years to 43% in 15-18 years age group.¹⁰

CV stroke was seen in 3.28% patients in this study. In study by Platt OS, on mortality in sickle cell disease, they followed 3764 patients and reported CV stroke in 22% of patients. Patients who died in this study had no overt organic failure but died during an acute episode of stroke, painful crises, or chest syndrome.¹¹

Avascular necrosis of hip is seen in total 4 patients (6.56%). In a study of AVN in adults Kuwaitis by Marouf R et al, 48.6% had varying degrees of femoral head avascular necrosis.¹²

CONCLUSION

Sickle cell disease was seen in younger patients. In Gujarat mainly tribal communities are affected like Rahwa, Bariya, Tadvi, Solanki, Bhil, Vasava and Damor. Commonest symptom found was musculoskeletal pain, followed by jaundice, fever, dyspnea, abdominal pain and chest pain. Sickle cell disease patients required past, recurrent admissions with past history of admission positive in 85% of patients. Overall 48 patients (80%) were admitted for 1-4 times and 3 patients (5%) admitted for 5-10 times.

Major systemic manifestations of sickle cell disease were pain crisis followed by hemolytic crisis, acute chest syndrome, hepatopathy and AVN of hip.

Past history of pain crises was seen in 73.33% of patients, past history of jaundice was seen in 63.33% while 31.67% patients had a history of blood transfusion.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Patel VP, Gandhi AU, Shah C. Epidemiological and clinical study of sickle cell disease at tertiary care centre in Western India. Int J Adv Med 2020;7:97-101.