ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 08 | August - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

BURDEN OF SICKLE CELL ANAEMIA AND RELATED MULTIORGAN COMPLICATION AMONG PATIENTS COMING TO TERTIARY REFERAL CARE CENTRE – A RETROSPECTIVE CLINICAL STUDY

General Medicine

Dr. Nazia Ansari* Assistant Professor, Department of General Medicine CIMS, Bilaspur. *Corresponding Author

Dr. Vivekanand Assistant Professor, Department of General Medicine CIMS, Bilaspur.

Dr. Chandrahas Dhruw Associate Professor, Department of Radiation Oncology CIMS, Bilaspur.

Dr. Pankaj Tembhurnikar Professor, Department of General Medicine CIMS, Bilaspur.

ABSTRACT

Background: Sickle cell disease (SCD), also simply called sickle cell, is a group of haemoglobin-related blood disorders typically inherited. The most common type is known as sickle cell anaemia. It results in an abnormality in the oxygen-carrying protein haemoglobin found in cells. This leads to a rigid, sickle-like shape under certain circumstances. Problems in sickle cell disease typically begin around 5 to 6 months of age. A number of health problems may develop, such as attacks of pain (known as a **sickle cell crisis**) in joints, anaemia, swelling in the hands and feet, bacterial infections, dizziness and stroke. **Objective:** To identify the burden of sickle cell anaemia and related multiorgan complication (Sickle cell crisis) among patients coming to tertiary referral care hospital. **Method:** This retrospective clinical study involves 168 clinically and haematological proven cases of Sickle cell disease (SCD) registered in department of General Medicine CIMS, Bilaspur conducted during 1march 2023 to 1 march 2024. **Result:** Most of the patients were in the age group of 20-30 yrs, and least patients were from more than 70 yrs age group, SCA is predominant among males than in females in our study group, acute painful episodes are the most common complication being 17.26%, followed by severe anaemia 4.16%, stroke 1.78%, Acute painful episodes as complication is most common presentation in <20 year age group 7.14%, followed by in 20-30 years age group 6.54%, Solitary patient found in the age group of 30 -40 yrs of acute chest syndrome, Stroke was seen in 3 age groups <20, 20-30 and 30-40 with 0.6% frequency in each group. **Conclusion:** SCA is most commonly found in the age group of 20-30 yrs it is more dominant in males than females, acute painful episodes followed by severe anaemia were emerged as most common complications in our study group, and early diagnosis with keen awareness among people could help us to identify complications early and proper management.

KEYWORDS

Sickle cell disease (SCD), sickle cell crisis, stroke, acute painful episode.

INTRODUCTION

Sickle cell disease is an inherited blood condition which is most common among people of African, Arabian and Indian origin. Sickle cell disease (SCD) affects approximately 5% of the world's population, and India has been the second highest country in the numbers of predicted SCD births. Despite the high burden in India, there is very few interventions dealing with the treatment and management of SCD are available. The most common form of SCD is sickle cell anemia (SCA), which arises in the presence of two copies of the haemoglobin S gene (HbS). HbS polymerizes when deoxygenated, yielding repetitive sickling episodes that lead to chronic haemolysis, vaso-occlusion, and eventually multiorgan dysfunction and death. Cerebrovascular disease is among the most common and debilitating complications of SCA, with 53% experiencing SCI by age 30 and 3.8% experiencing overt stroke by age 40 years. [1][2] Stroke is a devastating SCD complication with multiple detrimental long-term impacts and a high prevalence and recurrence rate. First, overt stroke has been associated with increased levels of health anxiety, which predicts lower levels of quality of life and higher levels of depression, general anxiety, and disability.[3] Second, cognitive impairment is significantly more common in individuals with SCD and ischemic stroke compared to those without a history of stroke. [4] Third, individuals with SCD and a history of end-organ damage such as stroke demonstrate higher rates of health care utilization, including more hospital days, emergency department visits, outpatient visits, lab tests, and outpatient pharmacy claims compared to those without end-organ damage. Another serious yet insidious cerebrovascular complication of SCD is silent cerebral infarct (SCI), which is generally defined as abnormal brain magnetic resonance imaging (MRI) findings in a patient with a normal neurologic exam and no history of overt stroke. [5] Individuals with compound heterozygous SCD are also at risk for SCIs with one study showing that 38% of those with $S\beta^+$ had evidence of silent infarction on brain imaging .[6] SCIs predict future overt stroke, progressive MRI abnormalities, and vascular stenosis.[7] Sickle cell disease (SCD) complications begin with the polymerization of sickle hemoglobin (HbS). Thus, SCD therapies are focused on preventing HbS production or reducing the circulating amount of HbS. Hydroxyurea treatment has become more widespread, whereas the number of evidence-based indications for erythrocyte transfusion is small. Hematopoietic stem cell transplant is a curative option for SCD but less than 25% of patients have a suitable donor. This article focuses on supportive and preventive care improvements and the benefits of hydroxyurea. The ribonucleotide diphosphate reductase inhibitor, hydroxyurea, is the recommended treatment for reducing VOCs in SCD. Its primary mechanism of action is through inducing production of fetal hemoglobin (HbF), though it also hydrates erythrocytes, reduces neutrophil and reticulocyte count and adhesion, reduces pro-inflammatory markers, and donates nitric oxide through the soluble guanylyl cyclase/cGMP pathway. [8]

Objective:-

To identify the burden of sickle cell anaemia and related multiorgan complication (Sickle cell crisis) among patients coming to tertiary referral care centre hospital.

MATERIALS AND METHODS

This retrospective clinical study involves 168 clinically and haematological proven cases of Sickle cell disease (SCD) registered in department of General Medicine CIMS, Bilaspur conducted during 1march 2023 to 1 march 2024.

Patient Inclusion Criteria

- Clinically and haematological proven cases of Sickle cell disease (SCD)
- All age group patients.
- ECOG performance score of 0 to 4.

Patient Exclusion Criteria

- All previously treated patients

RESULTS

Age

Most of the patients were in the age group of 20-30 yrs, and least patients were from more than 70 yrs age group, it shows that the young patients were suffered from SCA more than older patients.

Table 1 Age Interval Wise Distribution Of Patients

Age (yrs)	Frequency N = 168	%
<20	58	34.5

20 - 30	70	41.6
31 – 40	17	10.1
41 - 50	12	7.1
51 – 60	3	1.8
61 – 70	7	4.1
>70 yrs	1	0.6

Gender

58.9% patients were male and 41% were female. SCA is predominant among males than in females in our study group.

Table 2 Gender Wise Distribution Of Patients

Gender	Frequency	(%)
Female	69	41.0
Male	99	58.9
Total	168	

Complications:

Acute painful episodes are the most common complication being 17.2%, followed by severe anaemia 4.1%, stroke 1.7% and lastly splenic infarct and acute chest syndrome 0.6% each

Table 3 Various Complications With Frequency

Age	Frequency	%
Acute painful episodes	29	17.2
Acute chest syndrome	1	0.6
Severe anaemia	7	4.1
Splenic infarct	1	0.6
CVA (stroke)	3	1.7

Acute painful episodes

Acute painful episodes as complication is most common presentation in <20 year age group 7.1%, followed by in 20-30 years age group 6.5%, and only 3.2% in 30-40 years age group and lastly in 40-50 years age group 0.6%, similarly 0.6% in age group of 50-60 years.

Table 4: Age Wise Distribution In Acute Painful Episodes

Age	Frequency	%
<20	12	7.1
20-30	11	6.5
30-40	4	2.3
40-50	1	0.6
50-60	1	0.6
60-70	0	0
>70	0	0

Acute Chest Syndrome:

Solitary patient found in the age group of 30 -40 yrs

Table 5: Age Wise Details In Acute Chest Syndrome

Age	frequency	%
<20	0	
20-30	0	
30-40	1	0.6

Severe Anaemia:

Common in age group of 20-30 years 2.3%, followed by <20 years age group 1.19% and then 60-70 year age group 0.6%.

Table 6: Age Wise Distribution Of Severe Anaemia

Age	frequency	%
<20	2	1.1
20-30	4	2.3
30-40	0	0
40-50	0	0
50-60	0	0
60-70	1	0.6
>70	0	0

Splenic infarct:

Single patient found in the age group of < 20 yrs

Table 7: Age Wise Distribution Of Splenic Infarct

Age	frequency	%
<20	1	0.6

Stroke:

Stroke was seen in 3 age groups <20, 20-30 and 30-40 with 0.6% frequency in each group

Table 8: Age Wise Distribution Of Stroke

Age	frequency	%
<20	1	0.6
20-30	1	0.6
30-40	1	0.6

Co-morbidities:

Hypertension is the most common associated co morbidity 2.97%,followed by diabetes mellitus and pulmonary TB 1.78% each, 1.19% of chronic alcoholism, congestive cardiac failure and HIV, 0.6% each of Rheumatic heart disease, pericardial effusion and chronic kidney disease

Table 9: Associated Co Morbidities

	frequency	%
Chronic alcoholic	2	1.1
HIV +	2	1.1
Hypertension	5	2.7
Diabetes mellitus	3	1.8
Chronic kidney disease	1	0.6
Pulmonary TB	3	1.8
Rheumatic heart disease	1	0.6
Pericardial effusion	1	0.6
Congestive cardiac failure	2	1.1

Death:

Acute painful episode is the most common complication associated with death 1.78%, followed by stroke 1.19%.

Table 10: Cause Of Death In Sickle Cell Disease Patients

Cause	frequency	%
CVA (Stroke)	2	1.19
Acute painful episodes	3	1.78

DISCUSSION

This retrospective clinical study involves 168 clinically and haematological proven cases of Sickle cell disease (SCD) registered in department of General Medicine CIMS, Bilaspur conducted during 1march 2023 to 1 march 2024. Most of the patients were in the age group of 20-30 yrs, and least patients were from more than 70 yrs age group, it shows that the young patients were suffered from SCA more than older patients. Our data is similar to the study done by ***A. V. Shrikhande et al.*** [9] where they found that Most of the patient in their study was in the age group of 15 to 35 yrs, 25.1%,and predominated by males i.e. 27(61.36%) compare to females results are similar to our study where we found that 58.9% patients were male and 41% were female. SCA is predominant among males than in females in our study group. In a longitudinal cohort survey of 308 adult patients with SCD in the US by ***McClish DK, Smith WR, Dahman BA, et al*** [10], the lower back and lower limbs (knee, shin, hip) were the most frequently reported pain sites leading to Acute painful episodes in our research also we found that most common crisis is Acute painful episodes being 17.2%, followed by severe anaemia 4.1%, stroke 1.7% and lastly splenic infarct and acute chest syndrome 0.6% each similar results were found by ***Al Juburi G et al*** [11] with estimates ranging from 50% from a recent analysis of hospital episode statistics in England to an older estimate of 95% from a single centre US retrospective cohort study by ***Ballas SK et al*** [12]

Acute chest syndrome is a combination of pulmonary infarction, infection, and fat embolism. It occurs as a result of sickling and ischaemia in deep tissues of the chest wall. Patients typically present with fever, acute respiratory signs and symptoms, such as cough, wheezing, dyspnoea, crepitations or chest pain, and consolidation on chest. In a US prospective cohort study of 1772 episodes of acute chest syndrome in 939 patients with SCD, 50% had a pain event in the two weeks preceding the event. Mortality rate was estimated to be 1% in children and 4% in adults by ***Vichinsky EP et al***. [13] in our study we depicted that a solitary patient suffered from the same. Stroke was seen in 3 age groups <20, 20-30 and 30-40 with 0.6% frequency in each group. The risk of stroke is enormous in SCD. Approximately 11% of SCD patients have clinically apparent strokes before the age of 20.That risk increases to 24% by the age of 45 by A child with SCD has a stroke risk that is 333 times greater than that of a healthy child without SCD or heart disease. . [14]

CONCLUSION

SCA is most commonly found in the age group of 20-30 yrs it is more dominant in males than females, acute painful episodes followed by

severe anemia were emerged as most common complications in our study group, and early diagnosis with keen awareness among people could help us to identify complications early and proper management. The central and state governments are now supporting the establishment of centres for the diagnosis of patients and comprehensive care.

REFERENCES

1. K. Ohene-Frempong, S.J. Weiner, L.A. Sleeper, S.T. Miller, S. Embury, J.W. Moohr, *et al. Cerebrovascular accidents in sickle cell disease: rates and risk factors Blood, 91 (1998),* pp. 288-294, 10.1182/blood.V91.1.288.
2. M.R. DeBaun, L.C. Jordan, A.A. King, J. Schatz, E. Vichinsky, C.K. Fox, *et al.* American Society of Hematology 2020 guidelines for sickle cell disease: *prevention, diagnosis, and treatment of cerebrovascular disease in children and adults Blood Adv.,* 4 (2020), pp. 1554-1588.
3. P.R. Diamond, L. Dysch, J. Daniels Health Anxiety in Stroke Survivors: *A Cross-Sectional Study on the Prevalence of Health Anxiety in Stroke Survivors and its Impact on Quality of Life*
4. C. Maduakor, V. Alakbarzade, Y. Sammaraiee *The epidemiology of neurological complications in adults with sickle cell disease: a retrospective cohort study Front.* Neurol., 12 (2021), Article 744118.
5. M.R. Debaun, D. Armstrong, R.C. McKinstry, R.E. Ware, E. Vichinsky, F.J. Kirkham Silent cerebral infarcts: a review on a prevalent and progressive cause of neurologic injury in sickle cell anemia Blood, 119 (2012), pp. 4587-4596.
6. D.I. Zafeiriou, M. Prengler, N. Gombakis *Central nervous system abnormalities in asymptomatic young patients with Sbeta-thalassemia Ann. Neurol.,* 55 (2004), pp. 835-839.
7. S.T. Miller, E.A. Macklin, C.H. Pegelow *Silent infarction as a risk factor for overt stroke in children with sickle cell anemia: a report from the cooperative study of sickle cell disease* J. Pediatr., 139 (2001), pp. 385-390.
8. R.E. Ware, B.R. Davis, W.H. Schultz *Hydroxycarbamide versus chronic transfusion for maintenance of transcranial doppler flow velocities in children with sickle cell anaemia-TCD with transfusions changing to hydroxyurea (TWiTCH):* a multicentre, open-label, phase 3, non-inferiority trial Lancet, 387 (2016), pp. 661-670.
9. A. V. Shrikhande · A. A. Dani · J. R. Tijare · A. K. Agrawal. *Hematological profile of sickle cell disease in central India :* Indian J. Hematol. Blood Transfus 23(3–4):92–98
10. McClish DK, Smith WR, Dahman BA, et al *Pain site frequency and location in sickle cell disease: the PiSCES project.* Pain2009; 145:246-51.
11. Al Juburi G, Laverty AA, Green SA, et al. *Trends in hospital admissions for sickle cell disease in England,* 2001/02–2009/10. J Public Health2012; 34:570-6.
12. Ballas SK, Lusardi M. Hospital readmission for adult acute sickle cell painful episodes: frequency, etiology, and prognostic significance. Am J Hematol2005; 79:17-25.
13. Vichinsky EP, Styles LA, Colangelo LH, Wright EC, Castro O, Nickerson B, *Cooperative Study of Sickle Cell Disease Acute chest syndrome in sickle cell disease: clinical presentation and course.* Blood1997;89:1787-92.
14. Ohene-Frempong K, Weiner SJ *Cerebrovascular accidents in sickle cell disease: rates and risk factors.* Blood, 1998,vol.91 1 (pg.288-294)