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Neurological Manifestations In Sickle Cell Disease (Scd) In Children

KEYWORDS: Sickle Cell Disease (SCD), Neurological manifestation, Homozygous 'SS', Heterozygous 'AS'.

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ABSTRACT ***Introduction:*** *Sickle cell disease (SCD) is autosomal recessive inherited condition characterized by presence of anomalous haemoglobin 'S' in the erythrocytes. Patients with SCD inherit an abnormal haemoglobin which becomes insoluble when deoxygenated and so distorts the red cells and cause tissue infarction1,2,3. The organs mainly affected are spleen, the bones, the kidney, the lung and the skin. But any organ may be involved and the brain is not exemption. Hence the study is taken up to find out neurological manifestations in SCD in children. Aims &* ***Objectives:*** *To find out prevalence, nature and outcome of neurological manifestations in SCD in children.* ***Material and Methods:*** *This prospective study was conducted in pediatrics department of tertiary care hospital from Oct 2000 to April 2002. The study group includes SCD patients admitted in pediatrics ward and patients attending SCD speciality clinics, who were electrophoretically confirmed for diagnosis of sickle cell haemoglobinopathy. A detail history, clinical examination and routine investigation were done in each case.* ***Observation and Results:*** *A total of 204 case of SCD who were electrophoretically confirmed for diagnosis of sickle cell haemoglobinopathy were enrolled during study period, out of which 120(58.82%) patients were homozygous "SS" and 84(41.17%) patients were heterozygous "AS" for SCD. Out of 120 patients of haemoglobin "SS" 16 patients had neurological symptoms while no patients of haemoglobin "AS" had any neurological symptoms. The mean age of presentation of neurological manifestations in the present study is 6.5 years, out of which 7 were boys and 9 were girls. The convulsion was most common symptom in 9(56.25%) followed by hemiparesis in 5(31.25%), monoparesis in 1(6.25%) and 1(6.25%) patient had quadriparesis. Out of 16 patients with neurological manifestation 12 patients (75%) had complete recovery, 1 patient (6.25%) had neurodeficit, 1 patient had transient ischaemic attack and 2 patients (12.5%) died. Conclusion: The neurological manifestations were more common in Homozygous "SS" patients compared to Heterozygous "AS" patients. The mean age of presentation of neurological manifestations in the present study is 6.5 years. The most common neurological presentation in SCD found in our study was convulsion followed by cranial nerve palsies, hemiparesis, monoparesis and quadriparesis.*

INTRODUCTION

Sickle cell disease (SCD) is autosomal recessive inherited condition characterized by presence of anomalous haemoglobin **'S'** in the erythrocytes. Patients with SCD inherit an abnormal haemoglobin which becomes insoluble when deoxygenated and so distorts the red cells and cause tissue infarction[1,2,3]. The organs mainly affected are spleen, the bones, the kidney, the lung and the skin. But any organ may be involved and the brain is not exemption. The most dramatic and debilitating complication of SCD is the occurrence of stroke. In children under age 10 years, the most common cause of stoke is cerebral infarction which typically presents with sign and symptoms of hemiparesis, monoparesis, hemianesthesia, visual field deficits, aphasia, cranial nerve palsies or acute change in behavior[4]. The pathological process of stroke in SCD includes occlusion of vessels mainly anterior cerebral arteries and middle cerebral arteries by deformed sickle red cell[4].

MATERIALS AND METHODS

This prospective study was conducted in pediatrics department of tertiary care hospital from Oct 2000 to April 2002. The study group includes SCD patients (either homozygous sickle cell anaemia or heterozygous sickle cell trait) admitted in pediatrics ward and patients attending SCD speciality clinics, who were electrophoretically confirmed for diagnosis of sickle cell haemoglobinopathy. A detail history, clinical examination and routine investigation were done in each case. A systematic neurological examination (higher function, cranial nerve, motor system, sensory system, meningeal sign) was done in every case. Investigation like Hb, peripheral smear, blood sugar, serum calcium, lumbar puncture with cerebrospinal fluid biochemistry and computerized tomography of brain was done in every case who found to have neurological involvement on history and clinical examination. The details of neurological examination were recorded in predesigned and pretested performa.

OBSERVATION & RESULTS

A total of 204 case of SCD who were electrophoretically confirmed for diagnosis of sickle cell haemoglobinopathy were enrolled during study period.

Table 1: Age, Sex and Haemoglobin Electrophoresis distribution

SR NO	Age Group (yrs)	Sex		Haemoglobin Electrophoresis		No of patients
		Male	Female	SS	SA	
1.	2-5	32	26	36	22	58(28.43%)
2.	6-8	26	25	25	26	51(25%)
3.	9-12	61	34	59	36	95(46.56%)

In present study 58 patients (28.43%) were in age group of 2-5 years, out of which 32 were male and 36 females. 51 patients (25%) were in age group of 6-8 years, out of

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which 26 were males and 25 females. 95 patients (46.56%) were in age group of 9-12 years, out of which 61 were males and 34 females. The above table also shows that 120 patients (58.83%) had homozygous and 84 patients (41.17%) had heterozygous sickle cell disease.(Table 1)

Table 2: Caste distribution

SR NO	Caste	No of patients(n=204)	Percentage(%)
1.	Buddhist	165	80
2.	Teli	8	3.92
3.	Kunbi	11	5.39
4.	Gonda	3	1.47
5.	Kosthi	3	1.47
6.	Gowari	1	0.49
7.	Sonar	2	0.98
8.	Thakur	1	0.49
9.	Muslim	10	4.90

In the present study out of 204 cases, 165 (80.88%) were from Buddhist community and less number of patients were from Kunbi, Teli, Gonda, Kosthi, Gowari, Sonar, Thakur and Muslim community.(Table 2)

Table 3: Type of crisis

SR NO	Type of Crisis	Haemoglobin 'SS' patients (%)	Haemoglobin 'SA' patients (%)
1	Vaso-occlusive	102(85%)	64(76.19%)
2	Sequestrtion	14(11.66%)	0(0%)
3	Haemolytic	36(30%)	2(2.38%)
4	Aplastic	0(0%)	0(0%)

Above table shows that the most common type of crisis in both Haemoglobin 'SS" and Haemoglobin'AS' patients was Vasoocclusive crisis followed by haemolytic and sequestration crisis. No patient had Aplastic crisis in the present study.(Table 3)

Table 4: General Examination

SR NO	Findings	No of patients	Percentage(%)
1.	Pallor	166	81.37
2.	Icterus	83	40.68
3.	Splenomegaly	94	46.07
4.	Hepatomegaly	88	43.13

In the present study 166 patients (81.37%) were pale, 83 patients (40.68%) had icterus, 94 patients (46.07%) had splenomegaly and 88 (43.13%) patients had hepatomegaly. (table 4)

Table 5: Neurological findings
a)Handedness

SR NO	Handedness	No of patients(n=204)	Percentage(%)
1	Right	197	96.56
2	Left	4	1.96
3	Not developed	3	1.47

In present study, most of the patients were right handed while 3 patients did not developed handedness.(Table 5a)

b) Higher Function

SR NO	Higher Function	No of patients	Percentage(%)
1	Normal	194	95.09
2	Stupurous	6	2.94
3	Comatose	3	1.47
4	Delirium, Delusion, Hallucination	0	0
5	Abnormal memory	0	0
6	Abnormality of speech and language	1	0.49

Table 5b shows that 6 patients presented as stupurous, 3 as comatose and 1 with aphasia.

c) Neurodeficit

SR NO	Neurodeficit	No of patients
1	Cranial nerve palsy	7
2	Monoparesis	1
3	Hemiparesis	5
4	Quadriparesis	1

Table 5c shows that 1 patient presented as monoparesis of left upper limb, 5 patients with hemiparesis and 1 with quadriparesis. Cranial nerve palsy was associated with monoparesis, hemiparesis and 1 with quadriparesis who had right 6 nerve palsy.

d) Neurological manifestations in the present study

SR NO	Neurological manifestations	Haemoglobin "SS"	Haemoglobin "AS"	Percentage(%)
1	Convulsion	9	0	4.41
2	Hemiparesis	5	0	2.45
3	Monoparesis	1	0	0.49
4	Quadriparesis	1	0	0.49

In the present study, neurological manifestations were seen in patients with haemoglobin "SS" only. Out of 204 patients with sickle cell disease 16 patients had neurological manifestations i,e, 7.84% patients had neurological manifestation, 9 patients(56.25%) presented with convulsion, 5(31.25%) patients had hemiparesis, 1(6.25%) patient had monoparesis and 1(6.25%) patient presented with quadriparesis.(Table 5d)

Table 6: Investigation
CT Scan

SR NO	CT findings
1	Infarction of left frontoparietal region supplied by middle cerebral artery
2	Diffuse cerebral atrophy with communicating hydrocephalus
3	Infarction of cerebral cortex supplied by both anterior cerebral artery with cerebral edema
4	Infarction of frontoparietal cerebral cortex supplied by right middle cerebral artery
5	Infarction in the territory of left middle cerebral artery
6	Minimal cerebral edema

In the present study computed tomography was done in all cases presented with neurological manifestations except 1 patient, whose general condition precluded shifting the patient for CT scan. Out of 15 CT Scans 6 showed abnormalities and 9 were normal.(Table 6a)

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a) **CSF Findings:** Lumbar puncture was done in all 16 cases presented with neurological manifestations and found to be normal.
b) **EEG:** In the present study, EEG was done in 9 patients presented with neurological manifestation. Out of these, only 1 was abnormal while rests were normal

Table 7: Outcome in SCD with neurological manifestations

SR NO	Outcome	Percentage(n=16)
1	Complete recovery	12
2	Permanent deficit	1
3	Transient ischemic attack	1
4	Death	2

In present study out of 16 patients with neurological manifestation, 2 patient died, 2 had neurological deficit while 12 had complete recovery.(Table 7)

DISCUSSION

A total of 204 case of SCD who were electrophoretically confirmed for diagnosis of sickle cell haemoglobinopathy were enrolled during study period. Out of 120 patients of haemoglobin "SS" 16 patients had neurological symptoms while no patients of haemoglobin "AS" had any neurological symptoms. Thus the **Prevalence** of neurological manifestation for Homozygous "SS" and Heterozygous "AS" patient of SCD are 13.33% and 0% respectively. The overall prevalence of neurological manifestations in SCD disease in children is 7.8%. In study done by **Amayo et al[5] (1992)** and **Kirkham et al[6] (2001)**, prevalence of neurological manifestations in SCD in children found was 5% and 20% respectively. Similarly **Balkaran et al[7] (1991)** in his study found the prevalence of neurological manifestations of 7.8% in SCD in children which correlate with our study. The mean age of presentation of neurological manifestations in the present study is 6.5 years, out of which 7 were boys and 9 were girls. In the study of **Balkaran et al [7] (1991)** stroke occurred in 17 of 310 childrens with haemoglobin at a median age of 6 years 3 months, out of which 12 were boys and 5 were girls. Thus the observation regarding age of presentation of neurological manifestation tallies with that of Balkaran. Out of 16 patients with neurological manifestations 9 patients presented as convulsion, 5 patients presented with hemiparesis, 1 with monoparesis and 1 with quadriparesis. **Balkaran et al[7] (1991)** in his study found that out of 17 children with neurological manifestation, 14 had hemiparesis, 1 had ptosis and 2 had subarachnoid hemorrhage. **Amayo et al[5] (1992)** in his study found that out of 18 patients 12 patients had hemiparesis and 6 patient had convulsion. Our present study correlates with above two studies. In present study out of 16 patients with neurological manifestation, 2 patient died, 2 had neurological deficit while 12 had complete recovery. In study of **Balkaran et al [7](1991)** 6 patients of 17 died due to neurological complication.

CONCLUSION

The neurological manifestations were more common in Homozygous "SS" patients compared to Heterozygous "AS" patients. The mean age of presentation of neurological manifestations in the present study is 6.5 years. Among Community, Buddhist was most commonly affected and vasoocclusive crisis was most common presentation. The most common neurological presentation in SCD found in our study was convulsion followed by cranial nerve palsies, hemiparesis, monoparesis and quadriparesis.

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