

Blood Pressure Indices in Children with Sickle Cell Disease, of Age 8-18 Yrs. at a Tertiary Care Centre in Chhattisgarh- A Cross Sectional Study

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ABSTRACT

BACKGROUND

Sickle cell disease is the commonest haemoglobinopathy. Anaemia leads to a hyperdynamic circulation, decreased systemic resistance, and sickle polymerization causing hyperviscosity, natriuresis and hyposthenuria that blunt the maximal plasma volume expansion interplay to affect blood pressure indices. Asian and African continents have a high share of sickle cell population, being historically malaria endemic areas. These populations have previously witnessed early death due to complications from sickle cell disease. With the recent advanced therapeutic approaches, sickle cell patients are surviving beyond adulthood. Increased survival has resulted in a new subset of sickle cell patients which is exposed to the effects of persistent haemolytic anaemia and the haemodynamic alterations. These effects have not been studied in sickle cell patients in the state of Chhattisgarh. Most of the studies conclude that sickle subjects have lower systolic, diastolic and mean blood pressure and higher pulse pressure.

METHODS

Participants were all sickle cell anaemia patients with SS pattern under the age group 8 to 18 yrs. from the Paediatric Department of the Pt J N M Medical College and associated Dr B R A M Hospital, Raipur and the controls were healthy age matched patients without any haemoglobinopathy. This study documented blood pressure indices - systolic blood pressure, diastolic blood pressure, mean blood pressure and pulse pressure in children with sickle cell disease and comparable controls presenting to a tertiary medical college hospital in Chhattisgarh.

RESULTS

The present cross sectional study reveals no difference in these blood pressure indices of systolic blood pressure, diastolic blood pressure, mean blood pressure and pulse pressure between sickle and control subjects. Data was tabulated for 50 patients of sickle cell disease and 50 healthy controls without any haemoglobinopathy. The study concluded that the mean systolic blood pressure in sickle cell patients and control group was 119.46 ± 8.32 & 117.14 ± 8.10 ($p=0.161$), the diastolic blood pressure was 73.42 ± 7.88 & 72.60 ± 6.62 ($p=0.575$), mean blood pressure was 137.71 ± 11.44 & 135.85 ± 9.91 ($p=0.385$) and the pulse pressure was 46.04 ± 10.13 & 44.52 ± 9.00 ($p=0.430$) respectively.

CONCLUSIONS

The blood pressure indices in sickle cell patients in India vary from those observed in other studies from other countries. It is possible that the sickle population in the present study may be having relative systemic hypertension that can be ascertained with future studies.

KEY WORDS

Sickle Cell Disease, Blood Pressure Indices, Systolic Blood Pressure, Diastolic Blood Pressure, Mean Blood Pressure

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BACKGROUND

Sickle Cell Disease (SCD) is the most prevalent inherited blood condition worldwide resulting from substitution of valine for glutamic acid at sixth position of beta globin of haemoglobin.^{1,2} Protection from fatal malaria has resulted in concentration of sickle cell prevalence in populations in the Asian and African continents. Sickle cell disease forms a large part and burden to the health care system in these two continents. ICMR study suggests that 20% of sickle cell disease patients die in the first two years of their life and an additional 30% fail to survive till adulthood³. Recent therapeutic approaches have enabled increased survival to adulthood and beyond.

Increased survival would translate into a new and different set of disease complications to be assessed and managed, where adult onset disease like hypertension may form a special subset with different needs. Polymerization of sickle haemoglobin alters the rheological properties of sickle cells to increase viscosity of the flowing blood. Increased viscosity and increased stroke volume and associated decrease in peripheral resistance interplay to determine the consequent blood pressure in sickle cell anaemia patient. Chronic anaemia in sickle cell disease increases cardiac output either by increasing stroke volume or tachycardia or both and resulting decrease in peripheral resistance is reflected in a lower blood pressure in sickle cell patients. Persistent haemolysis punctuated by intermittent vaso-occlusive episodes encountered in the life span of a sickle cell disease patient can lead to chronic vasculopathy. Majority of studies done reveals that the blood pressure in sickle cell anaemia patients is significantly lower than the controls.

Physiologic reasons for lower systemic blood pressure are not known, however, different postulations include a medullary defect causing lower systolic blood pressure, hyperdynamic circulation due to anaemia causing systemic vasodilation, enhanced production of prostaglandins and nitric oxide and decreased vascular reactivity secondary to chronic vasculopathy. The effect of renal parenchymal and reno-vascular defects seen commonly in sickle cell disease patients on blood pressure also remains undetermined. Few studies in the past have addressed this issue with variable conclusions. Desai et al. found the systolic and diastolic blood pressures were not significantly different between sickle and control groups.⁴ In a study of 187 patients it was found that the mean blood pressure of 116/70 mmHg for sickle cell anaemia patient was significantly lower than comparable control group.⁵

Aneke in a study of 79 sickle patients found systolic blood pressure, mean and diastolic blood pressure to be significantly lower than mean diastolic blood pressure in control group⁶. However, the pulse pressure of sickle cell disease patients in comparison to the control group was significantly higher in the same study. This study aims to document blood pressure indices in children with sickle cell disease as compared to comparable control subjects presenting to a tertiary medical college hospital in Chhattisgarh.

METHODS

This is a cross sectional study conducted from January 2016 to September 2017. Participants were recruited from the Paediatric Department of the Pt J N M Medical College and associated Dr B R A M Hospital, Raipur. Controls were healthy age matched patients without any haemoglobinopathy. The study was approved by IEC and informed consent was obtained.

Inclusion Criteria

All sickle cell anaemia patients with SS pattern under the age group 8 to 18 yrs. were included in the study.

Exclusion Criteria

Sickle cell disease with known acquired heart disease and congenital heart disease, anaemia due to chronic illnesses, and thalassemia.

Sample Size

Sample Size for Unmatched Case-Control Study Assumptions.

Two-sided confidence level (1-alpha) = 95.

Power (% chance of detecting) = 80.

Ratio of Controls to Cases = 1.

Hypothetical proportion of controls with exposure = 40.

Hypothetical proportion of cases with exposure = 69.39.

Least extreme Odds Ratio to be detected (from pilot study): 3.40.

Fleiss Sample Size – Cases 48 & Controls 42.

Total sample size: 90.

Ten percent dropout or loss of follow up = 9.

Total final sample size = 99, rounded off to 100 with 50 cases and 50 control subjects to ease mathematical calculations.

Calculated from

<http://www.openepi.com/SampleSize/SSCC.htm>.

Sampling

All the demographic and clinical information was collected in the predesigned and pretested questionnaires. Blood pressure measurements were carried out by a trained medical social worker on a child sitting on a chair with arm resting on a table for support after 5 minutes rest period in a separate quiet room with one of the parents present by Watch BP HOME A (Microlife Corporation) as per 3 average reading automated protocol between 9.30 AM to 11.30 AM hours.

Statistical Analysis

Statistical t-test analysis was done using GNU PSPP version 1.2.0-g0fb4db. (<https://www.gnu.org/software/pspp/>).

Outcomes of the present study did not show normal distribution and were tested for significance by Kruskal-Wallis Test.

RESULTS

Data was tabulated for 50 patients of sickle cell disease and 50 healthy controls without any haemoglobinopathy. There were 27 male sickle cell patients (54%), and 23 female sickle cell patients (46%), while in the control group 31 males (62%) and 19 females (38%) respectively (chi-square, p=0.073) [Figure 1]. Age distribution had among sickle cell patients below 13 years, 20 subjects (40%), above 13 years, 30 subjects (60%) while in non-sickle cell control group, below 13 years, 22 subjects (44%), and above 13 years 28 subjects (56%) (Chi-square, p=0.099) [Figure 2]. The present study concluded that the mean systolic blood pressure in sickle cell patients and control group was 119.46 ± 8.32 & 117.14 ± 8.10 (p=0.161), the diastolic blood pressure was 73.42 ± 7.88 & 72.60 ± 6.62 (p=0.575), mean blood pressure was 137.71 ± 11.44 & 135.85 ± 9.91 (p=0.385) and the pulse pressure was 46.04 ± 10.13 & 44.52 ± 9.0 (p=0.430) respectively [Table -1]

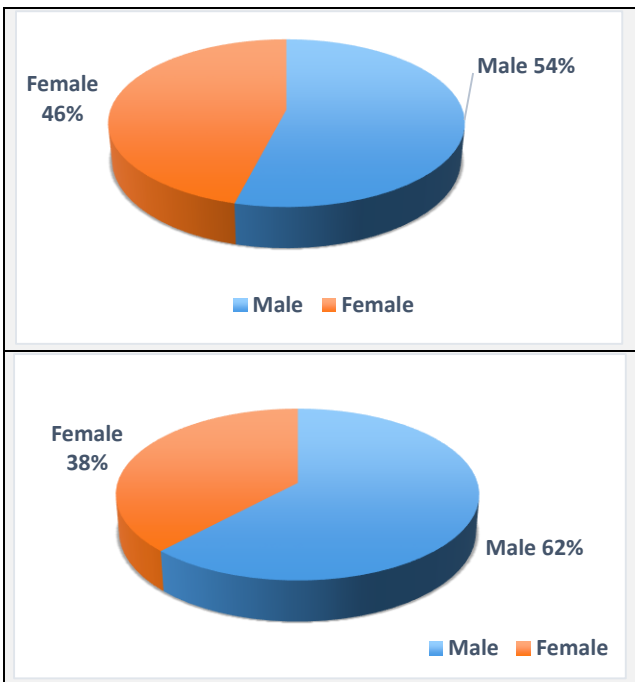


Figure 1. Sex Distribution - Sickle and Non Sickle Patients

Blood Pressure Indices	Sickle Cell Disease (n=50)	Non - Sickle Cell Disease (n=50)	p value (2-Tailed)
Systolic Blood Pressure	119.46 ± 8.32	117.14 ± 8.10	0.161
Diastolic Blood Pressure	73.42 ± 7.88	72.60 ± 6.62	0.575
Mean Blood Pressure	137.71 ± 11.44	135.85 ± 9.91	0.385
Pulse Pressure	46.04 ± 10.13	44.52 ± 9.00	0.430

Table 1. Blood Pressure indices in Sickle Cell Disease Patients and Controls

DISCUSSION

Blood pressure and chronological age follow linear relationship till adulthood. Systolic blood pressure remains below 100 mmHg till six years and reaches 120/80 mmHg by adulthood. In concordance to the present study, Desai et al. found the systolic (122.3 ± 21.7 mmHg vs 132 ± 17.6 mmHg, p=0.81) and diastolic blood pressures (68.3 ± 18 mmHg vs 174.9 ± 16.3 mmHg, p=0.30) to be not significantly different between sickle and control groups.⁴ However, Johnson in a

study of 187 patients found that the mean blood pressure of 116/70 mmHg for sickle cell anaemia patient was significantly lower than comparable control group.⁵ Aneke in a study of 79 sickle patients found systolic blood pressure mean of 115.52 ± 11.75 to be significantly (p=0.001) lower than mean 113.20 ± 7.94 for control group, and diastolic blood pressure was 62.59 ± 9.33 significantly lower (p = 0.003) than mean diastolic blood pressure of 75.40 ± 5.70 for control group⁶. However, the pulse pressure of 42.92 ± 10.91 in comparison to 37.8 ± 7.43 for the control group was significantly (p = 0.003) higher. Similarly, a study of 187 patients from Los Angeles, 64 adult patients from Jamaica, 81 adult patient from Netherlands, and studies from Jamaica, Turkey and Nigeria and 3317 subject in Cooperative study of sickle cell disease reached similar conclusions that blood pressure for sickle cell anaemia patient is significantly lower than controls.⁷ The underlying mechanisms were postulated by as the natriuresis and hyposthenuria that blunts the maximal plasma volume expansion and cause lower blood pressure.⁷ The present study found no significant difference in the blood pressure indices of systolic and diastolic blood pressure, mean blood pressure and pulse pressure, in discordance to all the above mentioned studies, except the study by Desai et al.⁴ The present study has a larger population in higher than 13 years age group that corresponds to more adult level blood pressures. Amma Benneh-Akwasi Kuma 2018⁸ from retrospective chart review of 1,000 adults with sickle cell disease at the Ghana Institute of Clinical Genetics have proposed a phenomenon termed relative systemic hypertension (RSH) with a systolic blood pressure range of 120–139 mmHg, and diastolic blood pressure range of 70–89 mmHg. It is possible that the sickle population in the present study may be having relative systemic hypertension that can be ascertained with future studies.

Limitations

The present study is limited by a small study population. There is less representation of cases below 13 years of age and none below 8 years of age.

CONCLUSIONS

The present study determines no significant difference in blood pressure indices between sickle cell and normal subjects. This is in contradiction to the majority of other studies. Further studies are needed to ascertain that the phenomenon of relative systemic hypertension underlies this difference in the present study.

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