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# Low risk of cardiopathy in indian sickle cell anemia patients

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## ABSTRACT

Lipid metabolism alteration causes some serious complications in humans; especially cardiac related complications. Sickle cell patients have variable clinical pathophysiology where few researchers suggested the abnormal cholesterol metabolism in sickle cell patients. Here we present the lipid metabolism and cardiac related complications in Indian sickle cell patients. Our cases were sickle homozygous who were diagnosed by HPLC and lipid profiling done by Beckman auto analyzer. T test applied for statistical analysis. All the lipid variable were significantly low in sicklers (p-value <0.05). Studies conclude the lipid metabolism is adequate and Indian sickle cell patients had lesser risk of cardiac related complications.



## KEYWORDS

Cholesterol;
HDL;
LDL;
Sickle cell disease.

## INTRODUCTION

Cholesterol level is determined by lipid metabolism which is in turn influenced by heredity, diet, liver, kidney, thyroid and other endocrine organ functions. The metabolism of lipids may be altered in sickle cell anemia and β-thalassemia patients. Sickle cell disease (SCD) is characterized by red cell rigidity, compromised perfusion, tissue infarction and chronic haemolysis. Lipid levels in sickle-cell disease associated with haemolytic severity, vascular dysfunction and pulmonary hypertension[1]. The chronic hemolytic anemia experienced by SCD patients leads to adverse effects on oxygen transport by the blood and to a decrease in oxygen availability for peripheral tissues. Limited tissue oxygen availability has the potential to modify events of intracellular metabolism and thus alter lipid homeostasis[2]. Pulmonary arterial hypertension (PAH) has emerged as a major complication and independent risk factor for sudden death among adults with SCD[3]. Several biomarkers have been associated with SCD clinical prognosis. Researchers have found a complex network of associations among laboratory analyses and clinical events predicting a probably risk of death[4-6]. Total cholesterol levels including high density lipoprotein cholesterol (HDL-C), low-density lipoprotein cholesterol (LDL-C), very low-density lipoprotein cholesterol (VLDL-C) and triglycerides may play potential biomarker ion in sickle cell disease. The presence of pulmonary hypertension was shown to be associated with several laboratory test alterations[7]. Recent study has also demonstrated the important role of the

apolipoprotein pathway and its association with endothelial dysfunction in SCD patients with pulmonary hypertension[7]. Elevated cholesterol levels increase the risk of arteriosclerosis and coronary heart disease[8]. There is a paucity of data to Indian sickle cell patient and cardiac risk, thus our aim was to investigate the level of cholesterol and risk of cardiac vascular disease in Indian sickle cell anemia patients.

## MATERIAL AND METHOD

Subjects were sickle cell anaemia patients; attending the Outpatient department; All India Institute of Medical Sciences (AIIMS). About 5 ml blood samples collected from patients as well as controls after taken signed consent, and study approved by institutional ethical committee. Diagnosis of patients was done by high performance liquid chromatography (HPLC-Bio-Rad-Variant™Bio Rad, CA, USA) while Complete blood count and red cell indices were measured by automated cell analyzer (SYSMEX K-4500, Kobe Japan). Lipid profiling of sickle cell patients done by Beckman CX-9 auto analyzer using Randox diagnostic kit. Blood serum used to analysis of lipid profile. Statistical analysis was performed on GraphPad software (version 3.06). t test used to compares the means of two groups while p-value <0.05 was considered statistically significant.

## RESULT AND DISCUSSION

Lipid profiling of 53 sickle cell anaemia patients done who were recruited from outpatient department haematology. Patient had 34 male and 19 female with mean age of 11.2±7.5year. Age and sex mathhed seventy control (42 male and 28 female with mean age 11.3±6.4) had been characterized to compare the lipid profile of patients. Mean value of LDL-C (75.3±14.7), HDL-C(30.2±8.6) and troglyceride (65.5±13.4) were significantly low in sickle homozygous patients while mean value of VLDL-C (17.4±4.2) were closer to controle value, however it was statisticaly significant. Details of lipid variables are given in TABLE - 1. Hypocholesterolemia has been described in SCD patients with significantly decreased LDL-C and HDL-C[1, 9-13]. and has been also described as a potential biomarker for SCD clinical severity[14]. Triglyceride rich

**TABLE - 1 :**

| | Mean ±SD | | |
|---|---|---|---|
| **Cholesterol** | **HbSS** | **Control** | **P-value** |
| | N=53 | N=70 | |
| LDL-C | 75.3±14.7 | 95.6±18.3 | <0.001 |
| HDL-C | 30.2±8.6 | 51.3±7.2 | <0.001 |
| VLDL-C | 17.4±4.2 | 19.3±5.8 | 0.045 |
| Triglycerides | 65.5±13.4 | 81.2±15.6 | <0.001 |

VLDL-C particles availability may play an important role in lipid oxidization in SCD patients. It has been suggested that VLDL-C is an important factor for atherosclerosis development[15]. The increase of triglycerides probably contributes to an increase in the hepatic production of VLDL-C, increasing the number of receptors for LDL-C that is extensively metabolized, decreasing its serum levels. However the role of cholesterol and triglycerides and the regulation of assembly and production of VLDL-C are poorly understood[16, 17]. In our cases of sickle homozygous, the level of HDL-C and LDL-C were significantly low (p-value<0.001). Although VLDL-C level were closer to the control value (p-value 0.045) while triglyceride significantly low in sickle cell anemia patients (p-value<0.001). Levels of HDL-C are an important cardiovascular risk factor, and HDL-C and apoA-I have been shown to decrease lesions and improve vascular reactivity in animal models of atherosclerosis and in humans; these changes may be due to the reduction of oxidized lipids and the enhancement of reverse cholesterol transport[18]. el-Hazmi[19,20]. et.al. well documented the low level of cholesterol in SCD compare to controls and found a positive correlation between the hemoglobin level and the level of cholesterol. Since increase in cholesterol level increases the incidence of arteriosclerosis, coronary heart diseases, cholestasis and xanthomatosis, the patients with sickle cell anemia may be at an advantage in having a protection against these disorders. Patients with sickle-cell anemia exhibit pro-oxidative metabolic perturbations. Due to chronic oxidative stress, plasma low-density lipoprotein (LDL) from patients with sickle-cell anemia is more susceptible to oxidation. A study report significantly low level of plasma cholesterol and HDL-C level and significantly high level of triglyceride in sickle cell patients with increased LDL-C. Increased LDL susceptibility to oxidation could be a marker of oxidant

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stress and vasculopathy in patients with sickle-cell anemia[21]. Various study report the alter lipid metabolism and significantly low level of total cholesterol in sickle cell patients[20, 22-27]. Out of 53 patients; none of the sicklers clinically present cardiac complications and all the lipid variables within the normal range with significantly lower than controls. These observations suggest the lipid metabolism in Indian sickle cell patients is appropriate and had lower risk of cardiopathy in sicklers.

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