

# Frequency distribution of sickle cell anemia, sickle cell trait and sickle/beta-thalassemia among anemic patients in Saudi Arabia

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## Abstract

**Background:** Notwithstanding, the growing incidence of sickle cell hemoglobinopathies (SCH) such as sickle cell anemia (SCA) or sickle cell disease, sickle/beta-thalassemia; the exact prevalence remains obscure in Saudi Arabia. Hence, this study is an attempt to determine the frequency of SCA and sickle cell trait (SCT) among all anemic patients with SCH treated at the King Abdul-Aziz Medical City (KAMC), Riyadh, Saudi Arabia. Furthermore, the hemoglobin (Hb) S and other Hb patterns (Hb AS and Hb F) were also estimated in SCA and SCT patients. **Materials and Methods:** Results of Hb capillary electrophoresis performed on all patients with SCH from January 2011 to December 2013 were evaluated retrospectively. **Results:** Of a total of 3332 patient data analyzed, 307 were anemic patients (58% males and 42% females) with SCH. The sickling test showed all the patients to be positive. Hb electrophoresis revealed the incidence of 96.7%, 3.3%, and 0% of the patients suffered from SCA, SCT and sickle/beta-thalassemia, respectively. Patients with SCA had a higher level of Hb F and showed no crisis when compared with other SCA patients who had lower or no Hb F levels. **Conclusion:** SCA is relatively frequent among males (56.4%) than females out of all patients with SCH. The SCA incidence was more common (48.5%) among children, frequency of SCT among adult age group was 1.6%, while sickle/beta-thalassemia was 0%.

**Key words:** Fetal hemoglobin, sickle cell anemia, sickle cell hemoglobinopathies, sickle cell trait, sickle/beta-thalassemia

## INTRODUCTION

Anemia is a condition in which the hemoglobin (Hb) concentration level is reduced. Sickle cell anemia is a disease of the blood, which is caused by an inherited Hb S gene.<sup>[1]</sup> Normal Hb consists of: Hb A, Hb A2 and Hb F, while a person with SCH has different Hb pattern.<sup>[2]</sup> In SCA the red blood cells are sickle or boat-shape.<sup>[3]</sup> Sickle Hb S is produced as a result of replacement of glutamic

acid instead of valine in position number six of the  $\beta$  chain.<sup>[4,5]</sup> The abnormal red cells (sickle cells) can block small blood capillaries causing damage in tissues and organs, which leads to pain (crisis)<sup>[6-9]</sup> increased extravascular destruction of red blood cells<sup>[10]</sup> and increased risk of severe dehydration.<sup>[11]</sup> SCA affects all age groups globally<sup>[11]</sup> with demographic variations. Children who are born with

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SCA inherit it from their parents. Persons who have the SCT or have SCA can be carriers of Hb S.<sup>[12,13]</sup>

Here, we determined the frequency distribution of SCA and SCT among anemic patients attending at KAMC - Riyadh, Saudi Arabia to assess the distribution of the disease and identify carriers among the population.

## MATERIALS AND METHODS

A retrospective chart review study was conducted to determine the frequency of SCA and SCT among anemic Saudi Arabian patients. The study data were collected from hematology laboratory at KAMC, National Guard of Health Affairs Hospital complex in Riyadh, Saudi Arabia.

- Inclusion criteria: Male and female patients of all age groups who were diagnosed with anemia in the hematology laboratory from January 2011 to December 2013 were included.
- Exclusion criteria: Nonanemic patients or anemic patients with anemia other than SCH were excluded.

### Data collection methods, instruments used, measurements

A computer printout of demographic data and discharge clinical events/outcomes collected from records department for all episodes of hospital discharges that were coded for diagnosis of SCA or SCT were analyzed.

### Data analysis

Study variables were directly entered into SPSS software, version 20 (The International Business Machines Corporation, New York). A backup soft copy version, as well as a hard copy print, was dated, saved and secured after each data entry update.

Statistical analysis of study variables was performed using SPSS software version 20. SCA, SCT frequency and clinical outcomes at discharge were determined.

## RESULTS

Out of 3332 anemic patient's data analyzed, 307 were anemic patients with SCH. The results of Hb electrophoresis showed 297 (96.7%) SCA, 10 (3.3%) SCT and sick/beta-thalassemia patients (0%) [Figures 1 and 2]. Patients with SCA, who had high levels of Hb F did not have any crisis when compared with SCA patients with low or no Hb F levels [Table 1]. The SCA was highly frequent (56.4%) among males while among the females the incidence was 40.5% out of all patients with SCH [Tables 2 and 3]. In children, SCA was more commonly (48.5%) observed while SCT was only 1.0%. The frequency of SCA and SCT in adults was 36.25 and 1.6% respectively [Tables 4 and 5].

**Table 1: Frequency of patient's crisis with respect to Hb patterns**

Patients crisis	Hb patterns			Total
	Hb SS	Hb A + Hb S	Hb S + Hb F	
Yes	232	0	2	234
No	10	10	54	73
Total	241	10	56	307

Hb: Hemoglobin

**Table 2: Frequency of Hb patterns among anemic patients with SCH**

Hb patterns	Frequency	Percentage
Hb SS	241	78.5
Hb A + Hb S	10	3.3
Hb S + Hb F	56	18.2
Total	307	100

Hb: Hemoglobin, SCH: Sickle cell hemoglobinopathies

**Table 3: Frequency of SCH with respect to gender**

Gender	Type of SCH		Total
	SCA (%)	SCT (%)	
Male	173 (56.4)	5 (1.65)	178
Female	124 (40.3)	5 (1.65)	129
Total	297 (96.7)	10 (3.3)	307

SCA: Sickle cell anemia, SCT: Sickle cell trait, SCH: Sickle cell hemoglobinopathies

**Table 4: Frequency of Hb patterns among SCA and SCT patients**

Types of SCH	Hb patterns			Total
	Hb SS (%)	Hb A + Hb S (%)	Hb S + Hb F (%)	
SCA	241 (81)	0 (0.0)	56 (19)	297
SCT	0 (0.0)	10 (100)	0 (0.0)	10
Total	241	10	56	307

Hb: Hemoglobin, SCA: Sickle cell anemia, SCT: Sickle cell trait, SCH: Sickle cell hemoglobinopathies

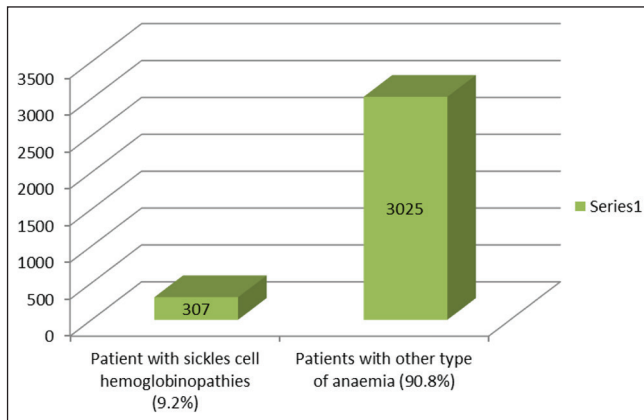
**Table 5: Frequency of SCA and SCT with respect to age groups**

Age	Types of SCH		Total
	SCA (%)	SCT (%)	
Child	149 (48.5)	3 (1.0)	152
Youth	37 (12.0)	2 (0.7)	39
Adult	111 (36.2)	5 (1.6)	116
Total	297 (96.7)	10 (3.3)	307

SCA: Sickle cell anemia, SCT: Sickle cell trait, SCH: Sickle cell hemoglobinopathies

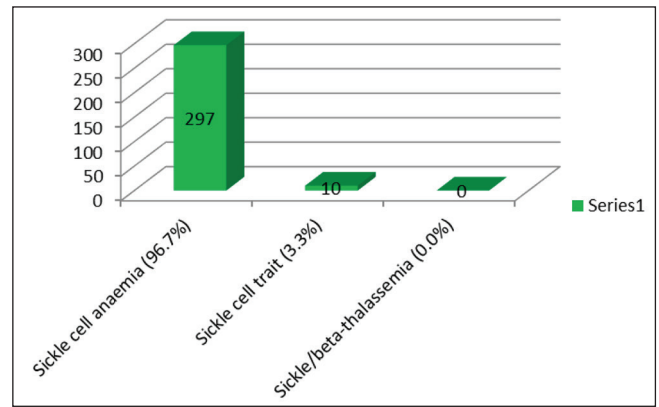
## DISCUSSION

Sickle cell hemoglobinopathies involve SCA, the SCT, and sickle/beta-thalassemia. SCA and SCT are most common forms of Hb defect (Hb S gene), which are inherited by children from parents.<sup>[14,15]</sup> SCH is one of the common health problems globally and in Saudi Arabia, where the gene frequency of this disease is highly prevalent.<sup>[16-19]</sup> In



**Figure 1:** Frequency distribution of patients with hemoglobinopathies among anemic patients

our study, we focused on SCA, SCT and the Hb patterns in each type of hemoglobinopathies in addition to age and gender of patients. The study group included 307 anemic patients with SCH, the high frequency of SCA among SCH Saudi patients with homozygous sickle cell (Hb SS) and less frequent SCT with the heterozygous sickle cell (Hb AS). These proportions are consistent with previous reports.<sup>[20,21]</sup> However, results of our study showed a high frequency of SCA (Hb SS) and SCT (Hb AS) among Saudi Arabian SCH patients. The high frequency of homozygous sickle cell (Hb SS) and heterozygous sickle cell (Hb AS) cases among the Saudi Arabian SCH population reflect the high frequency of Hb S gene, which may be due to consanguineous marriage, which is a common tribal tradition practice in this population.<sup>[22,23]</sup> Additionally mild cases of SCA were also observed among adult patients. Some patients with SCA never had symptoms or crisis due to the presence of fetal Hb in high level.<sup>[24]</sup> Fetal Hb seems to protect the patients from disease severity and crisis. In our study, the majority of the SCA patients with (Hb SS) pattern had a severe crisis, and they tended to be more symptomatic. In contrast, the majority of SCA patients had (Hb S + Hb F) and these patients did not suffer from any crisis due to the presence of fetal Hb in an appropriate concentration.<sup>[25,26]</sup> The level of fetal Hb in the blood circulation is very essential as they seem to protect the red blood cells from becoming sickle shape and hence prevent them from blocking the blood flow, especially in small capillaries.<sup>[26]</sup> We conclude that SCA is the most common type of hereditary anemia among the Saudi Arabian population. The distribution of Hb S among Saudi population illustrate that Hb AS predominates among family. High level of fetal Hb protects from disease crisis. The level of Hb S among individuals with SCA is gender and age independent. We recommend that family screening of the SCA and SCT is necessary to identify sickle cell carriers and should be extended to all areas, which have the high frequency of Hb S, and premarriage investigation should be considered as a routine investigation.



**Figure 2:** Frequency of sickle cell anemia, sickle cell trait and sickle/beta-thalassemia among patients with sickle cell hemoglobinopathies

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### Conflicts of interest

There are no conflicts of interest.

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