Original Article

Sickle Cell Trait and Disease in Anaemic Patients, Visiting Health Care Centre, KFU, Al Hasa

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ABSTRACT

Objective: To determine the frequency of sickle cell trait and disease in anaemic patients of Al Hasa.
Study Design: Descriptive / cross-sectional study
Place and Duration of Study: This study was conducted at the At health Care Centre, King Faisal University, Al Hasa, from June to November 2016.
Materials and Methods: Anaemic patients, diagnosed by blood complete picture,were included in the study.Total 638 anaemic patients were investigated for sickle cell trait and disease, which consisted of 230 children (100 males and 130 females), and 408 adults (210 males and 198 females). The blood samples of these patients were analyzed by using both, the sickle cell screening kit and the haemoglobin electrophoresis.The data were analysd by using SPSS 18.
Results: 80 (12.5%) out of 638 investigated patients were positive for sickle cell trait, among which 32 were children (10 males and 22 females) and 48 were adults (26 males and 22 females). 20(3.1%) out of 638 investigated patients were positive for sickle cell disease, among which 6 were children (1 male and 5 females) and 14 were adults (10 males and 4 females).
Conclusion: We found a significant frequency of sickle cell trait and disease in anaemic patients of our region Al Hasa, which indicates the urgent need of screening and counselling programs in this region to prevent further transmission of this familial disorder to new generations.
Key Words: Anaemia, frequency, sickle cell disease, sickle cell trait

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INTRODUCTION

Sickle cell disorder is a group of inherited disorders of red blood cells which contain abnormal haemoglobin called sickle haemoglobin (Hb S).[1]Individuals who have both copies of abnormal haemoglobin genes (Hb SS) are termed as sickle cell disease (SCD) patients. Individuals who are affected with one copy of sickle haemoglobin and the other copy of normal haemoglobin (Hb AS) are termed as sickle cell carriers. The carrier state is often referred to as sickle cell trait (SCT).The SCD patients may present with diverse clinical manifestations, ranging from asymptomatic with mild anaemia to systemic illnesses which are caused due to haematological and vaso-occlusive events and infectious crises.[2, 3] SCD patients may also develop various life threatening systemic illnesses, such as, pulmonary hypertension, cardiac failure,[4] renal diseases[5]and skull bone infarction with epidural haematoma.[6]Many SCD patients suffer from leg ulcers[7]and acute & chronic bone pains, which result due to sickle cell crisis.[8]

Sickle cell disorder is considered as a major public health problem in certain parts of the world which include the Middle East, sub-Saharan Africa, India, Eastern coast of the America and Western Europe.[9] In the middle East, Kingdom of Saud Arabia (KSA) is well-known for its high prevalence of familial haemoglobinopathies, and the sickle cell disorderis one of those. It is prevelent throughout the KSA, being most common in the Eastern and Southern provinces.[10]The data on this disorder is scarce and back-dated due to very limited research in KSA. Due to lack of knowledge, many new generations are born with this disorder. Due to associated complicated clinical outcomes of this disorder, the families of the patients

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spend a huge amount of their earnings to the hospitals. Therefore, it is very important to know the updated prevalence of this disorder.

In the current study, we determined the frequency of this disorder in anaemic patients of Al Hasa, which is the largest city in Eastern province of KSA.These patients belonged to King Faisal University, which included the staff, students and their families.Hence, literate people and their families were included in this study. We chose the literate group to know the prevalence and awareness among these people, who would further make awareness and do counseling to the common people of this region to control the further transmission of this familial disorder to the new upcoming generations.

MATERIALS AND METHODS

This prospective study was conducted at Health Care Center of King Faisal University (KFU), Al Hasa city from June to November 2016. For ethical consideration, we obtained the ethical approval from ethical committee of KFU under the letter No. 9/31/89. The patients' consent was taken at the Health Care Center, KFU. The patients belonged to KFU, which included the staff, students and their families. Total 638 anaemic patients, diagnosed by performing blood complete picture (blood C.P) at Health Care Center of KFU were included in the study. Patients under the age of 14 years were considered children. The blood samples were analyzed first by using sickle cell screening test kit 'HbS-Solubility Screening' (Helena Bioscince Europe), and then confirmed by performing haemoglobin electrophoresis using BIO RAD System-D10.The controls, provided by the manufacturer, were run in parallel.

Briefly, for screening, 2ml venous blood was collected inan EDTA bottle and mixed gently. From this sample, 20µl blood was taken in a test tube and 2ml reagent mixture was added and mixed well as per the kit manufacturer's instruction. The mixture was allowed to stand for 3-5 minutes and examined for turbidity. The positive and the negative controls were run in parallel with the test samples.A turbid reaction denoted a positive result (i.e., the lines on the provided viewing chart could not be seen when viewed through the sample tube). A negative result was obtained with a clear reaction (i.e., the lines on the viewing chart were clearly visible when viewed through the sample tube). Furthermore, to differentiate between SCDand SCT, the test tubes were centrifuged at 1200g without brake for 5 minutes.Red precipitate on top of yellow solution indicated homozygous HbS (sickle cell disease), while red precipitate on top of pink solution indicated heterozygous HbS (sickle cell trait).

For further confirmation of SCT and SCD, the positive samples were analysed by performing haemoglobin electrophoresis, using BIO RAD System-D10, which confirmed the final diagnosis of SCT and SCD.

The data were analysed by using SPSS 18.

RESULTS

Among total 638 investigatedanaemic patients, 80 (12.5%) were positive for SCT and 20 (3.1%) for SCD. Among those 80 positive patients for SCT, 32were children (10 males and 22 females), and 48 were adults (26 males and 22 females).From 20 positive patients for SCD, 6 were children (1 male and 5 females) and 14 were adults (10 males and 4 females).The prevalence of SCT andSCD in number of patients, based on age and gender groups is described in Figure 1.

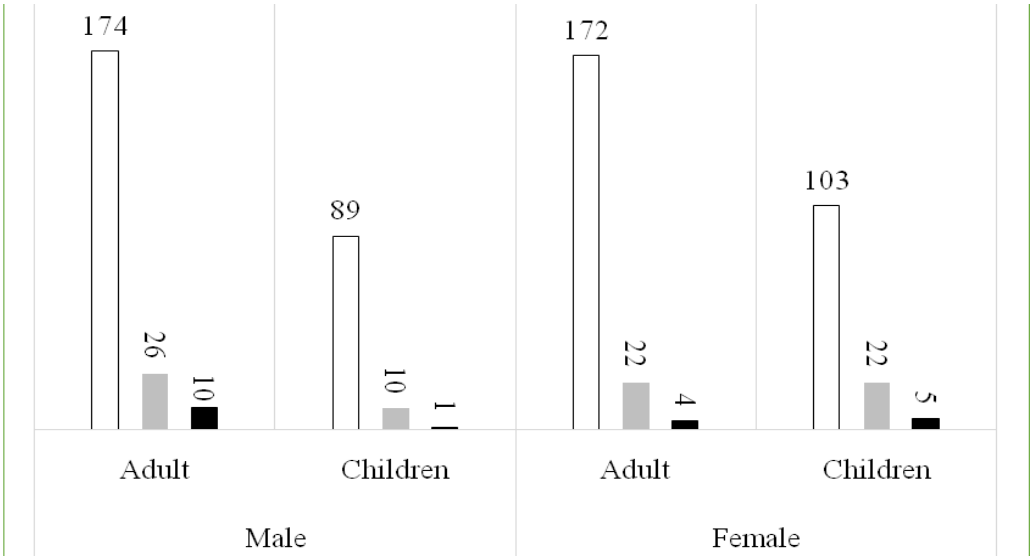

Figure No.1.Number of patients with sickle cell trait (SCT) and sickle cell disease (SCD), based on age and gender groups. Grey bar: SCT, Black bar: SCD, White bar: Negative for SCT & SCD.

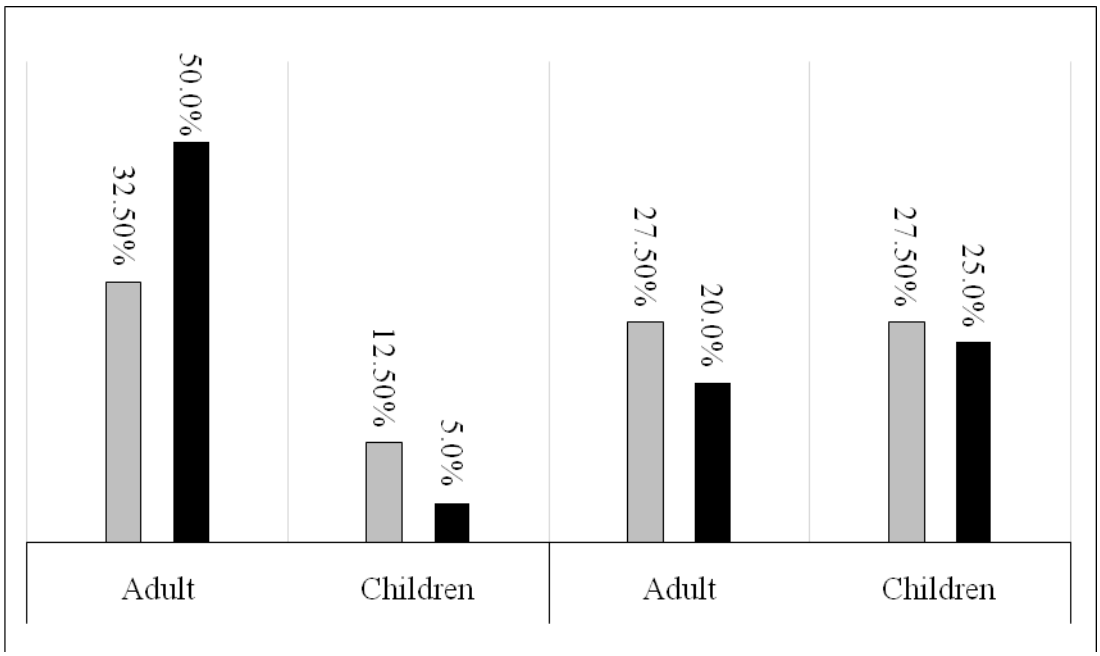

Figure No.2: Percentage of patients with sickle cell trait (SCT) and sickle cell disease (SCD) among individual groups. Grey bar: SCT, Black bar: SCD.s

The prevalence of SCT and SCD in percentage among individual group is described in Figure 2. Thus, we had 4 groups, i.e., female children, female adults, male children and male adults. The highest prevalence of SCD was found among male adults, which was 10 among 210 total male adults (4.7%). The second most common group was female children, which was 5 among total 130 children (3.8%). The third common group was female adults, which was 4 among total 198 female adults (2%), and the fourth group was male children which was 1 among total 100 male children (1%).

DISCUSSION

In this study, we investigated the frequency of SCD and SCT amonganaemic patients who belonged to the literate people and their families of KFU, Al Hasa. It was quite surprising that the frequency of this disorder was high among even literate people. We determined significant prequency of this disorder, i.e., 3.1% SCD and 12.5% SCT among our investigatedanaemic patients. Overall, we did not find a significant differencein the frequency among individual group, based on age and gender.This is justifiable with the fact that this is an autosomal disorder which affects the both genders equally and the patients may be diagnosed at early or later stage of life. Similarly, another study also reported no significant difference in the requency of this disorder among various groups.[11, 12]Saudi Arabia is a large country with approximately 32 million population, living in an area of 2,149,690 km^2. The data on this disorder among Saudi population is scattered and back dated. Al-Qurashi[11] reported a prevalence of SCD in five main regions of Saudi Arabia, which was 0.06% in Central, 0.12% in Western, 1.45% in Eastern, 0.0% in Northern, and 0.24% in Southernregions.Another study in KSA also showed almost the same prevalence for SCD (0.26%) and for SCT (4.2%).[13]Ziad Ahmed[14]reported overall0.2% prevalence of SCD and 4.3% of SCT in KSA.An old data of 1998 from Al Hasa region,reported by Nasserullah[15]showed considerable difference inprevalence of this disorder among neonates. He reported the prevalence of 2.35% for SCD and 28.21% for SCT in Qatif city, and 1.08% for SCD and 20.02% for SCT in Al Hasa city.A study on the screening of sickle cell disorderin 2004 showed higher prevalence of this disorder in our region than the other regions of KSA, which was 1.2% for SCD and 17% for SCT.[16]

On the basis of general prevalence, our results slightly differ from other studies conducted in Al Hasa region. As we chose a restricted group of literate people and their families, it was surprising that the prevalence of this disorder was even higher.Various factors may be possible for this, for example, lack of awareness,large family size and consanguineous marriages as many of the patients gave this history. These factors were previously described as the possible causes of sickle cell disorder in KSA.[17]Although our study was conducted on a relatively smaller number of patients in comparison to the other largesample-size studies, this prevalence is highly significant because only the literate people and their families were included and these people visited the Health Care Center just in 2016. Therefore, our updated findngs indicate that there is a

an urgent need to organize the awareness, screening and genetic counseling programs of this disorder not only among the literate people, but also in whole Al Hasaregion so that further transmission of this inherited disorder to the next generations can be avoided.

CONCLUSION

We found a significant frequency of sickle cell disorder among literate people and their families. Our researchrecommends theurgent need of awareness, screening and counselling programs of this disorder on higher level in Al Hasa region in order to prevent the further transmission of this disorder to the next generations.

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Final Approval of version:	Hussain Mohammed Alhiwaishil

Conflict of Interest: The study has no conflict of interest to declare by any author.

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