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Full Length Research Paper

Prevalence of Sickle Cell Trait and Glucose 6 Phosphate Dehydrogenase Deficiency among Blood Donors in a Nigerian Tertiary Hospital

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ABSTRACT

Blood donation from sickle cell trait (SCT) and glucose-6-phosphate dehydrogenase (G6PD)-deficient donors might alter the quality of the donated blood during processing, storage or in the recipients' circulatory system. The aim of this study was to determine the prevalence of SCT and G6PD deficiency among blood donors in Jos University Teaching Hospital (JUTH). It also reviewed the benefits and risks of transfusing blood from these blood donors. This cross-sectional study was conducted on 130 blood samples obtained from blood donors that presented to JUTH blood bank during the period of March to June 2008. All samples were tested for Hb-S by alkaline cellulose acetate electrophoresis and for G6PD deficiency by quantitative spectrophotometer assay method. Out of the 130 blood donors, 27 (20.8%) were diagnosed for sickle cell trait, 26 (20%) for G6PD deficiency and 7(5.4%) for both conditions. We recommend the screening of all units for sickle cell trait and G6PD deficiency and to defer donations from donors with either of these conditions, unless if needed for special blood group compatibility, platelet apheresis or if these are likely to affect the blood bank inventory. If such blood is to be used, special precautions need to be undertaken to avoid complications in high-risk recipients.

Keywords: Sickle cell trait, G6PD deficiency, blood donors.

INTRODUCTION

The study of haemoglobin has been of major importance in Biology and Medicine. Haemoglobin and myoglobin were the first proteins to have their three-dimensional structures determined, and they have played a key role in understanding of the relationships between protein structure and function (Wainscoat, 1989).

The human haemoglobin molecule has four globin subunits, each covalently linked at a specific site to a

haeme group, consisting of an iron atom surrounded by a porphyrin ring. Haemoglobin, the main constituent of red blood cell cytosol is a metalloprotein that functions mainly in the transport of oxygen from the lungs to the tissues. Its function of oxygen transport is dependent on the ability of the ferrous iron to combine reversibly with molecular oxygen (Wainscoat, 1989).

Mutation in the genes coding for haemoglobin protein results in a group of inherited diseases known as haemoglobinopathies. Haemoglobin S is one of such

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haemoglobin proteins and the most common mutant haemoglobin variant, which is brought about by an autosomal structural single-point mutation. It is characterized by poor solubility in the deoxygenated state followed by polymerization leading to red blood cell (RBC) shape distortion, rigidity and eventually extravascular haemolysis.

People with one sickle haemoglobin gene and one normal haemoglobin gene known as sickle cell trait carriers are somewhat more resistant to malaria than people with two normal haemoglobin genes. In Nigeria, it has been estimated that about 50% of the population carry the sickle haemoglobin gene. However, the reported carrier rate of the sickle haemoglobin trait (AS) in southern Nigeria was 25% and 19-32.6% in northern Nigeria (Fleming *et al*, 1979; Walters & Lehman 1956; Jelliffe & Humphreys, 1952).

Glucose-6-phosphate dehydrogenase (G6PD) deficiency, the most common enzymopathy known to humans affects approximately 400 million people worldwide (Mason, 1996). This X-linked inherited disorder most commonly affects persons of African, Asian, Mediterranean, or Middle-Eastern descent. It is characterized by inability of RBCs to effectively counter oxidative stress, thereby exposing them to haemolysis. Persons with this condition are asymptomatic until they are exposed to oxidative stress.

Intrinsic red blood cell defects like G6PD deficiency and haemoglobinopathies might affect its survival, resistance to various stresses and/or interaction with other cells like leucocytes or endothelial cells. It is of high significance if such red blood cells are to be donated and transfused to recipients encountering a stressful event (Alabdulaah *et al*, 2010).

Among the inherited RBC disorders, G6PD deficiency and sickle cell trait share a number of features in common:

- Present in high frequency in many geographical areas and ethnic groups
- Usually asymptomatic and in stable conditions without alteration in haemoglobin levels, RBC count and indices, hence easily missed by routine full blood count and clinical history taken from an individual who has not been screened for them or experienced any acute haemolytic episode (Alabdulaah *et al*, 2010).

It is therefore, not uncommon to encounter such persons who are affected by the same conditions as prospective blood donors. Controversies regarding the quality of blood donated by these two groups during processing, storage or in the recipient's circulation exist, to the extent that some blood banks defer or reject individuals with these conditions (CBBS e-Network Forums).

In Nigeria, like many tropical and subtropical regions, there is high frequency of G6PD deficiency and sickle cell trait (Luzzatto and Gordon-Smith, 2001; Jelliffe & Humphreys, 1952). Presently, blood donors are not routinely screened for these conditions in most of our blood banks and blood transfusion service centres. Their detection and identification of such individuals is relied on the pre donation data. The potential alteration in the quality of the donated blood and the possibility of the risks of transfusing such blood to some recipients has spurred us to embark on this study. We therefore report the prevalence of sickle cell trait and G6PD deficiency among intending blood donors at the Jos University Teaching Hospital in Jos and also review the benefits and any risks of utilizing blood from these donors.

MATERIALS AND METHODS

This is a cross sectional prospective study carried out at the blood bank unit, department of Haematology and Blood Transfusion, Jos University Teaching Hospital (JUTH), Jos-Nigeria, between March 1 and June 30, 2008. Blood samples were obtained from one hundred and thirty (130) prospective blood donors who presented to JUTH blood bank during the period under study. They were aged 20 to 49 years. Blood samples were collected in ethylene diamine tetra-acetic acid (EDTA) bottles and then stored at 4°C in a refrigerator till required for processing. All blood samples so collected were analyzed within 48 hours of collection. The samples were analyzed for haemoglobin concentration, total leucocyte count and platelet count using standard haematologic techniques. The blood smears were prepared, and then followed by the investigations to detect haemoglobin S using cellulose acetate haemoglobin electrophoresis at alkaline pH of 8.9 and G6PD deficiency by quantitative spectrophotometer assay method. Statistical analyses were determined using statistical software epi-info computer package. Results were expressed as mean \pm SD and presented in tables.

RESULTS

A total of one hundred and thirty (130) intending blood donors were studied. They were aged between 20 and 49 years. One hundred and twenty seven (127) of them were males (97.7%), only three (3) were females (2.3%). The age and gender distribution of the subjects studied is shown in table 1. Majority of the blood donors (34.6%) were in the age bracket of 20 to 24 years and all males.

Out of the 130 blood donors, 27 (20.8%) were diagnosed for sickle cell trait, 26 (20%) for G6PD deficiency and 7(5.4%) for both conditions. All were males and Nigerians. Tables 2 and 3 show the distribution of the haemoglobin types and the G6PD status of the blood donors respectively. Their haemoglobin levels were normal, 153 ± 20 g/L, the total WBC count were normal, $5.2 \pm 1.2 \times 10^9$ /L, platelet count were normal, $187.5 \pm 37.5 \times 10^9$ /L and blood smears also appeared normal with a normocytic normochromic red blood cells morphology

Table 1:
Age and gender distribution of blood donors

Age (years)	Gender		Total no of subjects
	M	F	
20 – 24	45	0	45(34.6%)
25 – 29	31	1	32(24.6%)
30 – 34	26	1	27(20.8%)
35 – 39	7	1	8(6.2%)
40 – 44	12	0	12(9.2%)
45 – 49	6	0	6(4.6%)
Total	127	3	130(100)

Table 2:
Proportion of donors with sickle cell trait (SCT)

Haemoglobin type	n	percentage
Haemoglobin AS	27	20.8
Haemoglobin AA	103	79.2
Total	130	100

Table 3:
Proportion of donors with G6PD deficiency

G6PD status	n	Percentage
Deficient	26	20
Normal	104	80

DISCUSSION

Sickle cell trait (SCT) and G6PD deficiency both have high frequency of occurrence in Nigeria. These conditions are believed to provide some protection against malaria.

Sickle cell trait (SCT) has been considered a benign condition. It has high prevalence rate in many regions where malaria has been or presently is endemic. Nigeria being a malaria endemic region has high prevalence rate of sickle haemoglobin. The rate of occurrence of SCT in

Northern Nigeria was reported to be between 19-32.6% (Fleming et al, 1979).

In this study, the prevalence rate of SCT in our blood donor was 20.8%. This agrees with previous study carried out in the general population in Jos, the Plateau state capital (Egesie et al, 2003) and falls within the reported rate of occurrence of SCT in the Northern part of Nigeria (Fleming et al, 1979).

Red blood cells collected from SCT donors have been shown to frequently occlude white blood cells' reduction filters. The main cause of this filtration failure being attributed to haemoglobin polymerization (Stroncek et al, 2002 and 2004; Schuetz et al, 2004; Bryrre et al, 2003; Brandão et al, 2003).

In our blood bank and many other blood transfusion centres in Nigeria, routine screening of intending blood donors to detect the presence of sickle haemoglobin is not done yet. Nigeria is known to be one of the countries with the highest burden of sickle cell diseases (SCD), with a prevalence rate of sickle cell anaemia ranging from 1.5 to 3.1% in different parts of the country (Kaine and Udeozo 1981; Adewuyi and Akintunde 1987; Omotade et al, 1998). Furthermore, world health organisation (WHO) on the average, has estimated the prevalence of sickle cell anaemia to be about 20 per 1000 live births annually which translates into about 150 000 children born annually with sickle-cell anaemia in Nigeria, making her the country with the highest burden of sickle cell anaemia in the world (WHO, 2006). Many of the sufferers of sickle cell disease require blood transfusion as part of the therapeutic intervention in these patients. Often than not, the blood transfused would be from a SCT blood donor. A SCD patient receiving blood transfusion from a SCT carrier in emergency situation is certainly not being given the best supportive intervention as undesired effects are inevitable. Blood from SCT donor is more likely to increase the concentration of haemoglobin S in SCD patient instead of reducing it, the main aim of exchange blood transfusion in life threatening complications of SCD like acute chest syndrome and strokes (cerebrovascular accidents) among others. For this reason, there is an urgent need for formulation and implementation of policies that make it mandatory for all blood banks and transfusion centres to screen all intending blood donors for SCT with the view of adequately and appropriately identifying such units of donor blood and restriction placed on them for use in SCD patients.

G6PD deficiency is very prevalent in Nigeria with a prevalence rate ranging from 4 - 26% (Luzzatto, 2001; Ademowo, 2002). In the northern part of the country, a prevalence rate of 20% has been reported in the North-

Central region (Egesie et al, 2008) and 20.6% in the North-Eastern region (Allison et al, 1961). Our results in this study confirmed a high prevalence of SCT and G6PD deficiency. Majority of the blood donors in this study were males. This collaborates with studies from elsewhere in Nigeria (Muktar et al, 2005). A similar finding was obtained in a study from Riyadh, Saudi Arabia where 98.7% of the blood donors were males (Alabdulaah et al, 2010). Reasons thought to be responsible for this observed trend are those of socio-cultural belief that blood donation is an activity reserved only for men, lack of advocacy, and temporary deferral of female blood donors resulting in gender inhibition. Other reasons for not donating blood by women include fear of expected trauma (during and after the process), concern about their own health with regards to anaemia, a possible pregnancy, low body weight or a previous deferral for these same reasons, temporary ineligibility due to medical reasons related to low levels of iron, ailments or difficult veins and fear of adverse reaction were important barriers to giving blood among women (Bani and Giussani, 2010).

The use of G6PD deficient blood has been studied for simple therapeutic intervention such as exchange blood transfusions (EBT). It has been proposed that several biochemical changes and depletion in the antioxidant defence system occur on storage of G6PD deficient blood (Orlina et al, 1970; Józwick et al, 1997). It was observed in a study that preterm infants who received G6PD deficient blood developed haemolysis requiring further intervention by exchange transfusions (Mimouni et al, 1986). Lesser drop in post-EBT total serum bilirubin, prolongation of the duration of phototherapy, the need for repeat EBT and the occurrence of massive intravascular haemolysis have been reported in infants in whom G6PD deficient donor blood were used for EBT practice (Walker and Bowman, 1959; Shahriary et al, 1997; Sandip et al, 2009). These complications were reported in patients with normal G6PD activity receiving G6PD deficient blood. In areas with high prevalence of G6PD deficiency, more neonates with jaundice that might require EBT are likely to be G6PD deficient, hence the incidence and severity of these complications are expected to be higher.

The use of G6PD deficient blood in adults has not been reported to show significant adverse clinical consequences following transfusion except biochemical changes reflecting mild haemolysis in one study (McCurdy and Morse 1975; Shalev et al, 1993; Huang et al, 1998).

The potential risk of haemolytic complications associated with the use of G6PD deficient blood for transfusing some selected recipients may justify the

practice of deferral of G6PD deficient blood donors by some blood banks in areas where, screening for this deficiency is practised as part of pre-donation screening. This is however, not the situation in our locality and many other blood banks or transfusion service centres in Nigeria. The pre-donation screening for G6PD deficiency should form part of pre-donation screening of all intending blood donors in regions of high prevalence of G6PD deficiency like Nigeria, for the reason that use of such G6PD deficient blood in some selected patients might be attended with adverse clinical complications, so that those found to be deficient may not necessarily be deferred but could have their donor units appropriately labelled and such units restricted from use in high risk patients going through stressful events.

The prevalence of SCT and G6PD deficiency is high in our blood donors. Transfusion with G6PD- deficient blood carries a potential risk of haemolytic complications, especially if it is used for EBT in neonates. The blood donated by SCT donors apart from its undesired effects if transfused to SCD patients, also lead to white blood cell filtration failure. For these reasons, in high-prevalence areas for SCT and/or G6PD deficiency like Nigeria, we recommend the screening of all units for these conditions and those donors found with these conditions unless if needed for special blood group compatibility or platelet aphaeresis be deferred. Alternatively, if the blood bank inventory is likely to be affected, then donations can be accepted but G6PD-deficient blood should be labelled and should not be released to be transfused to a G6PD-deficient patient or be used in EBT in the paediatric age group particularly neonates. Similarly, SCT units need to be labelled too; such blood is better stored in bags that allow increased oxygen saturation and not used to transfuse SCD patients or in patients where white blood cell filtration is of great concern. In blood banks with limited resources where screening for SCT and G6PD deficiency is not feasible we advise that the units that are likely to be transfused to high risk recipients be screened for these conditions, particularly if single-unit transfusion is going to be undertaken.

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