Public Awareness of Glucose-6-Phosphate Dehydrogenase Deficiency in Sokoto

Jelani I¹, Garba N² and Raji A.Y³

¹Department of Chemical Pathology, Faculty of Medical Laboratory Sciences, Usmanu Danfodiyo University, Sokoto, Nigeria. ²Department of Medical Laboratory Sciences, Faculty of Allied Health Sciences, Bayero University, Kano, Nigeria. ³Department of Animal Science, Faculty of Agriculture, Bayero University, Kano, Nigeria.

*Corresponding author: jelannsaudia@yahoo.com

Received: 01.04.16; Accepted: 22.04.16; Published: 25.04.16

ABSTRACT

Background: Awareness of glucose-6-phosphate dehydrogenase (G-6-PD) deficiency is very vital because of its far reaching implications of mild chronic hemolytic episode in deficient individuals which could be triggered following ingestion of oxidant agents such as fava bean, oxidant drugs and infections. Aim: The aim of this study was to assess public awareness of G-6-PD deficiency with expectations towards reducing its public health burden through proper enlightenment and education in Sokoto. Methods: This is a cross sectional survey that was carried out among people living within Sokoto metropolis. A total of 500 respondents (307 males and 193 females) were included in the study. The data instrument used was self-/interviewer-administered questionnaire. The data obtained were analysed using SPSS software package (Version 20). Results: Two hundred and seventy (54%) of the participants have heard of G-6-PD deficiency and 240 (48%) recognize it as a blood related disease, only 48 (9.6%) are aware of their G-6PD status. Three hundred and eighteen (63.6%) knew that Fava bean ingestion can be a triggering factor for hemolysis in affected individuals. Males were more knowledgeable than their Female counterpart about the disease that affects them most. Conclusion: With poor awareness about G-6-PD deficiency among people in Sokoto, there is a great challenge of acute hemolysis in deficient individuals ignorantly treated with oxidant drugs. Therefore, stakeholders at various levels should introduce campaign programs in addressing the public health burden of this abnormality.

Key words: Public awareness, glucose-6-phosphate dehydrogenase, oxidant, haemolytic episode, oxidant drugs, fava bean

INTRODUCTION

Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency was first identified in American blacks in the course of studies of sensitivity to the haemolytic effect of primaquine. G-6-PD deficiency is an asymptomatic X-linked inherited condition which is almost unknown to the public. G-6-PD deficiency is produced by mutations in the G-6-PD gene, mostly point mutations which lead to enzyme instability and absent activity.
mutations and small deletions that causes structural defects in the enzyme. This gene is located near the telomeric region of the long arm of the X chromosome (band Xq28), close to the genes for coagulation Factor 8, color blindness and X-linked dyskeratosis congenita. In relation to the red blood cell, G-6-PD functions to reduce nicotinamide adenine dinucleotide phosphate (NADPH) while oxidizing glucose-6-phosphate. NADPH is needed for the production of reduced glutathione (GSH) which is important to defend the red cells against oxidant stress. G-6-PD deficiency is the most common human enzyme defect, being present in more than 400 million people worldwide. The highest prevalence is reported in Africa, Mediterranean Europe, South-East Asia, and Latin America. In Nigeria, 4-26% prevalence of G-6-PD deficiency was reported. Omisakin et al. reported 25.5% among blood donors in the South Western part of Nigeria.

The clinical manifestations of G-6-PD deficiency contribute to neonatal jaundice which is accompanied by hyperbilirubinaemia and puts many infants at risk of brain damage (kernicterus) within the first few days of life. Kernicterus can lead to hearing deficits, behavior problems, and permanent neurologic damage. During childhood, many children with G-6-PD deficiency are healthy until they are exposed to a pro-oxidant medication or chemical. Classically, anti-malarial drugs are strong oxidant agents and have substantial use in Sub-Saharan Africa. Additionally, exposure to the pro-oxidant naphthalene, the active ingredient in mothballs, is common among young children. In G-6-PD deficient children, pro-oxidant exposure can lead to a rapid imbalance in the redox status in red blood cells leading to haemolysis and resultant severe anaemia, heart failure, and even death if not recognized early.

The WHO recommends population screening in regions where the prevalence of G-6-PD deficiency is 3–5% or more, but this has yet to become routine practice in many parts of Nigeria. Barriers to screening include cost, under-estimation of the public health impact of G-6-PD deficiency by the medical community, lack of awareness of G-6-PD deficiency among lay people and a paucity of guidelines regarding which high risk groups should be preferentially screened when general population screening is not possible. Screening test for the detection of G-6-PD deficiency is a pre-requisite test for oversea travelling, especially to Mediterranean countries where fava bean is been frequently cultivated and ingested. Also before primaquine therapy when travelling from a malaria endemic to non-malaria endemic region to achieve radical cure for the relapsing liver stages of Plasmodium vivax-hypnozoites which become dormant in infected hepatocytes.

Several diagnostic tests methods have been used to detect G-6-PD deficiency. The fluorescent spot test is simple and cheap and has a sensitivity of 100% in homozygotes and hemizygotes. A disadvantage of the test is that it only identifies individuals with less than 20% G-6-PD activity. This can yield false normal results in female heterozygotes and in deficient individuals who have experienced a recent episode of hemolysis due to the elevated G-6-PD activity in reticulocytes. The spot test is not well-suited for screening of neonates due to the presence of reticulocytosis. Cytochemical and spectrophotometric assays are available options. The spectrophotometric assay relies on the same principles as the fluorescent spot test. It also requires more training and equipment than the spot test. In the cytochemical assay, G-6-PD activity causes staining of individual erythrocytes by converting a water-soluble colorless compound into its insoluble dark-colored form through the production of NADPH. This test has a sensitivity of 85% in heterozygotes (compared to 32% and 11% for the fluorescent spot test and spectrophotometric assay respectively). The major setbacks of this test are that is it time-consuming, technically difficult and considerably more expensive than the fluorescent spot test.

Study by Aaron has shown that, G-6-PD deficiency is implicated in the pathogenesis of a number of common diseases like hypertension, diabetes mellitus and atherosclerosis. In addition, G-6-PD deficiency could lead to hyperglycemia, making more glucose available for the non-enzymatic production of advanced glycosylation end (AGE) products and cardiac dysfunction might also be aggravated by a deficiency in G-6-PD.
Association of G-6-PD deficiency with cancer, cataract formation and some other disorders has also been reported with controversial results.\textsuperscript{6,17-20} Cardiac dysfunction might also be aggravated by a deficiency in G-6-PD. As G-6-PD is a critical antioxidant enzyme, essential for maintenance of cytosolic redox status in cardiomyocytes, deficiencies may contribute to cardiac dysfunction through increased susceptibility to free radical injury and impairment of intracellular calcium transport.\textsuperscript{[21]} Complete deficiency of the enzyme in leukocytes was reported to be associated with chronic granulomatous disease due to neutrophil dysfunction.\textsuperscript{[22-24]}

G-6-PD deficiency is the most common genetic defect of red blood cells which cannot be reversed, but its awareness allows those affected and their parents to take steps to reduce the risk of acute hemolysis. Hence, this study was undertaken to assess public awareness of G-6-PD deficiency with expectations that would probably enhance it screening beforehand thereby reducing its implications particularly in this locality.

**RESULTS**

Table 1 shows the demographic characteristics of participants. Males constituted the majority being 61.4\% and 39.6\% were females respectively. Most respondents were within the age of 20 to 29 years. On marital status, more than two-third (370; 74\%), were single while less than one-third (130; 26\%) were married. Occupational status of respondents shows that (363; 72.6\%), (122; 24.4\%) and (15; 3\%) were students, civil servant and others respectively. Respondents of Hausa ethnicity made up the largest group (280; 56%) followed by those Yoruba descent (113; 22.6\%) and those of Igbo (34; 6.8\%) ethnicity. The results in figure 1 indicated that majority of the respondents are aware of the disease, and had recognized it as a blood disease. Similarly, their G-6-PD status was not known to them. Response to general knowledge questions about G-6-PD deficiency separated by gender is shown in figure 2. The results show that males are more knowledgeable of this disease which mainly affects them.

**DISCUSSION**

Nigeria is endemic for malaria and antimalarial drugs are routinely prescribed for most cases. This treatment can induce oxidative damage to the cells and in turn causes severe haemolysis in G-6-PD deficient individual. Awareness and knowledge of this genetic condition have tremendous health benefit in the management of deficient patients. In this study, participants showed poor knowledge to basic questions about G-6-PD deficiency, though many have heard and recognized it as blood related disease. However, their knowledge about the pattern of inheritance seems to be lacking. This finding is consistent with the report of Uzoegwu and Awah\textsuperscript{[25]} in Cameroon but at variance with the report of Shaikha and Amani\textsuperscript{[26]} in Bahrain. These differences may be due to lack of public enlightenment education at various level on the implications of G-6-PD deficiency and compulsory screening of the disease right from birth in most part of the developing countries.
Table 1: Demographic characteristics of participants

<table>
<thead>
<tr>
<th>Demographic Features</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>307</td>
<td>61.4</td>
</tr>
<tr>
<td>Female</td>
<td>193</td>
<td>39.6</td>
</tr>
<tr>
<td>Age (years)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10-19</td>
<td>44</td>
<td>8.8</td>
</tr>
<tr>
<td>20-29</td>
<td>354</td>
<td>70.8</td>
</tr>
<tr>
<td>30-39</td>
<td>73</td>
<td>14.6</td>
</tr>
<tr>
<td>40-49</td>
<td>23</td>
<td>4.6</td>
</tr>
<tr>
<td>50-59</td>
<td>6</td>
<td>1.2</td>
</tr>
<tr>
<td>Marital status</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Married</td>
<td>130</td>
<td>26.0</td>
</tr>
<tr>
<td>Single</td>
<td>370</td>
<td>74.0</td>
</tr>
<tr>
<td>Occupation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Civil servant</td>
<td>122</td>
<td>24.4</td>
</tr>
<tr>
<td>Students</td>
<td>363</td>
<td>72.6</td>
</tr>
<tr>
<td>Others</td>
<td>15</td>
<td>3.0</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hausa</td>
<td>280</td>
<td>56.0</td>
</tr>
<tr>
<td>Yoruba</td>
<td>113</td>
<td>22.6</td>
</tr>
<tr>
<td>Igbo</td>
<td>34</td>
<td>6.8</td>
</tr>
<tr>
<td>Others</td>
<td>73</td>
<td>14.6</td>
</tr>
</tbody>
</table>

n=number, %=percentage.

Figure 1: Response on general knowledge about G-6-PD deficiency

1=Have you heard of G-6-PD deficiency, 2=G-6-PD deficiency is a red cell disorder, 3=G-6-PD deficiency is an inherited disorder, 4=If a father is G-6-PD deficient and the mother is not, having a girl who is a carrier is possible, 5=Can I donate blood if am G-6-PD deficient, 6=Do you know your G-6-PD status, 7=Do you want know your status, 8=Can Fava beans trigger hemolytic episode due to G-6-PD deficiency, 9=Can antimalarial drugs trigger hemolytic episode due to G-6-PD deficiency, 10=Jaundice can be a sign of red blood cell destruction due to G-6-PD deficiency, 11=G-6-PD deficient individuals are less prone to severe malarial infection.
Jelani et al.: Public awareness of G-6-PD deficiency

Figure 2: Histogram showing; responses to general knowledge about G-6-PD deficiency in population of Sokoto metropolis separated by gender

M= male, F=female, (a)=G-6-PD deficiency is a red cell disorder, (b)= G-6-PD deficiency is an inherited disorder, (c)=If a father is G-6-PD deficient and the mother is not, having a girl who is a carrier is possible, (d)= Can I donate blood if am G-6-PD deficient, (e)= Can fava bean trigger hemolytic episode due to G-6-PD deficiency, (f)=Can antimalarial drugs trigger hemolytic episode due to G-6-PD deficiency, (g)= Jaundice can be a sign of red blood cell destruction due to G-6-PD deficiency, (h)= G-6-PD deficient individuals are less prone to severe malarial infection

A greater proportion of respondents did not know whether deficient individual could donate blood or not. Conversely, transfusion with G-6-PD-deficient blood carries a potential risk of haemolytic complications, especially if it is used for exchanged blood transfusion in neonates.[27] Similarly, the use of G-6-PD-deficient blood has been studied for simple and exchange transfusions.[28] It has been proposed that several biochemical changes and depletion in the antioxidant defense system occur on storage of G-6-PD-deficient blood.[28] This is likely the reasons why authors reveal the necessity of including G-6-PD testing in the blood donors screening criteria since the ultimate goal of transfusion is to benefit the recipients.[29]

In the present study, almost all the respondents among which are medical personnel and students were ignorant of their G-6-PD status. As at the time of the study, there are two major hospitals in the town, neither of which routinely screen for G-6-PD deficiency which may contribute to this. The respondents showed fair knowledge of the symptoms of G-6-PD deficiency. This can be attributed to the relatively mild clinical nature of G-6-PD deficiency. No specific studies have assessed the knowledge of G-6-PD deficiency within the country to compare our results with.

CONCLUSION

We found a poor level of awareness of G-6-PD deficiency in our study. Therefore, we recommend public enlightenment programs towards ensuring G-6-PD screening beforehand to avert acute haemolytic episode which may occur in deficient individuals ignorantly treated with oxidants agents.
ACKNOWLEDGEMENTS

The authors thank all the participants of this study.

REFERENCES


doi: http://dx.doi.org/10.14194/ijmbr.5.1.7

How to cite this article: Jelani I, Garba N and Raji A.Y. Public Awareness of Glucose-6-Phosphate Dehydrogenase Deficiency in Sokoto. Int J Med Biomed Res 2016;5(1):50-56

Conflict of Interest: None declared
Submit your next manuscript to any of our journals that is the best fit for your research

Reasons to publish your manuscript with Michael Joanna Publications:
- User-friendly online submission
- Rigorous, constructive and unbiased peer-review
- No space constraints or coloured figure charges
- Immediate publication on acceptance
- Authors retain copyright
- Inclusion in AJOL, CAS, CNKI, DOAJ, EBSCO, Google Scholar, and J-Gate
- Unlimited and wide readership
- Member of COPE and CrossRef

Editorial Director
Professor Sofola A. Olusoga,
Department of Physiology,
University of Lagos,
Nigeria.
Tel: +234(0) 7093848134
Email: enquiry@michaeljoanna.com
www.michaeljoanna.com

International Journal of Medicine and Biomedical Research
Scope: IJMBR publishes cutting edge studies in medical sciences
Editor-in-Chief: Sofola A. Olusoga, MBBS, PhD, FAS
Deputy Editor: Lehr J. Eric, MD, PhD, FRCS
URL: www.ijmbr.com
E-mail: editor@ijmbr.com
Pissn: 2277-0941, eISSN: 2315-5019

International Journal of Ethnomedicine and Pharmacognosy
Scope: IJEP publishes novel findings on the use of complementary and alternative medicine in the management of diseases
Editor-in-Chief: Dickson A. Rita, B.Pharm, GCAP, PhD ,MPSGh, MCPA
Deputy Editor: Kuete V., PhD
URL: www.ijepharma.com
E-mail: editor@ijepharma.com
Pissn: 2437-1262, eISSN: 2437-1254

International Journal of Infectious and Tropical Diseases
Scope: IJITD publishes interesting findings on infectious and tropical diseases of public health importance
Editor-in-Chief: Yang Z., PhD
Deputy Editor: Liping L.P., MD, PhD
URL: www.ijitd.com
E-mail: editor@ijitd.com
Pissn: 2384-6607, eISSN: 2384-6585