PREVALENCE OF GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY AMONG HOSPITALIZED CHILDREN AT TERTIARY CARE HOSPITAL IN EASTERN PART OF NEPAL

Chandra Bhushan Jha*, Hem Sagar Rimal, Ram Bhakta Subedi, Surya Bahadur Parajuli

ABSTRACT

Introduction
Glucose 6 phosphate dehydrogenase (G6PD) deficiency is the most significant enzyme defect. It is an X linked inherited disorder that affects males and females are rarely affected by lyonization. Severe jaundice, anemia and hemolytic crisis following ingestion of fava beans and certain drugs are known to occur in children with G6PD deficiency. Therefore, routine neonatal and child screening programs to facilitate the identification and effective management of children with G6PD deficiency is paramount.

Objective
The main objective of this study is to find out the hospital prevalence of G6PD deficiency in hospitalized children at Birat Medical College teaching Hospital.

Methodology
This is a hospital based cross sectional study carried out in the Department of Paediatrics, Birat Medical College Teaching Hospital from 30th November 2020 to 30th May 2021. Three hundred children upto to ten years of age admied in department of pediatrics were included in this study. This study was performed on hospitalized children who were screened for G6PD deficiency. The test was carried out using the Randox G6PD quantitative in vitro test to determine the prevalence of G6PD deficiency among the admitted children upto 10 years of age. Data was analysed using SPSS version 16.

Result
This study was performed on 300 children, in which male babies (n=192; 64%) outnumbered the female babies (n=108; 36%). The majority of children were in the age group of < 1 years (n=131; 43.7%). The overall prevalence of G6PD deficiency was 9.3% of which 96.4% were moderately deficient while 3.6% was severely deficient. The highest proportion was noted in the age group of 1 to 5 years. The frequency of this disorder in males and females were 13.5% and 1.9% respectively which was statistically significant (p=0.001).

Conclusion
The present study confirms the high prevalence of G6PD deficiency in eastern region of Nepal. Therefore, we need to establish routine screening and educational programs in order to prevent grave complications in future.

KEYWORDS
Children, G6PD deficiency
INTRODUCTION

G6PD deficiency is a genetic disorder involving more than 400 million population worldwide.¹ It is an X linked inherited disorder and males are more commonly affected than the females.²

The main source of energy for the red cell is glucose, which is metabolized by two major routes; the hexose monophosphate (HMP) shunt and the glycolytic pathway.³ G6PD is a cytoplasmic enzyme in the pentose monophosphate pathway and catalyzes the conversion of nicotinamide adenine dinucleotide phosphate (NADP) to its reduced form, NADPH which helps to protect the red blood cells (RBC) from oxidative damage.⁴ Therefore, in patients who are deficient in G6PD, the RBCs are damaged and undergo lysis, leading to acute hemolysis. The term favism is used to describe hemolysis triggered by ingestion of fava beans in G6PD deficient individuals and it runs in families.⁵

Based on the percentage of G6PD enzyme activity, the deficiency is categorized as moderate (<30% activity) or severe (<10% activity). There are certain triggers in the form of foods (Fava beans), pollen inhalation, drugs (primaqine, chloramphenicol and aspirin) or chemicals (Henna, Naphthalene) or infections which lead to severe haemolysis in such a situation and urgent blood transfusion may be required.⁶

The enzyme G6PD deficiency contributes to neonatal jaundice which is accompanied by hyperbilirubinemia and leads infants at risk for kernicterus within the first few days of life. Many children with this enzyme deficiency are healthy during childhood and becomes symptomatic after exposure to a pro-oxidant medications such as anti-malarial drugs which lead to hemolysis and resultant severe anemia, heart failure, and even death if not recognized early.⁷

Therefore, screening and detection of G6PD deficiency helps in reducing such episodes through appropriate selection of treatment, patient counseling and abstinance from disease precipitating drugs.

In Asia, the prevalence of deficiency ranges from 6.0% to 15.8%.⁸

WHO recommends population screening in regions where the prevalence of G6PD deficiency is 3–5% or more, but this has yet to become routine pracce in Nepal. The barriers to screening include cost, under estimation of the public health impact of G6PD deficiency by the medical community, lack of awareness of G6PD deficiency among people and a paucity of guidelines regarding which high risk groups should be preferentially screened when general population screening is notpossible.⁹

There is a paucity of data on G6PD deficiency among children in the Koshi zone of Nepal. This study was specifically aimed at determining the prevalence of G6PD deficiency in children visiting the Pediatric unit of Birat Medical College and Teaching Hospital for pediatric related care. The research findings may be useful for the pediatrician to make comprehensive management plan at an early stage while dealing with children with G6PD deficiency. The main objective of this study is to determine the prevalence rate of G6PD deficiency in children so that the complications can be prevented.

METHODOLOGY

This is a hospital based cross sectional study carried out from 30th November 2020 to 30th May 2021 in the department of Pediatric, Birat Medical College and Teaching Hospital. Ethical clearance was obtained from the Institutional Review Committee (IRC) of the institute to carry out the study. The study was conducted on 300 children out of which 192 (64%) were males and 108 (36%) were females. The informed consent was taken from their parents. The parents of children unwilling to give consent, children older than 10 years of age and samples with incomplete data were excluded. In this study all the children admitted in pediatrics and neonatal ward up to the age of ten years were included. The ethylene diaminetetraacetic acid (EDTA)-anticoagulated blood was used for the screening of the subjects for G6PD deficiency using the Randox G6PD quantitative in vitro test. Red blood cell G6PD value of ≥2.9 U/gHb was regarded as normal. Children with red blood cell G6PD value of <2.9 U/gHb were regarded as deficient, while those with red blood cell G6PD value of <1.6 U/gHb were regarded as severely deficient.⁴ The data was recorded in a pre designed proforma and data analysis was done using statistical package of social science (SPSS) version 16. Numerical variables were reported in terms of mean and standard deviation. Categorical variables were reported in terms of numbers and percentages.

RESULTS

The present study constituted a total of 300 cases, in which male children (n=192; 64%) outnumbered the female children (n=108; 36%). The majority of children were in the age group of < 1 years (n=131; 43.7%) followed by 115 (38.3%) cases in the age group of 1 to 5 years and 54 (18%) cases in the age group of 6 to 10 years. (Table 1)

<table>
<thead>
<tr>
<th>Variables</th>
<th>Number</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Gender</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>192</td>
<td>64.0</td>
</tr>
<tr>
<td>Female</td>
<td>108</td>
<td>36.0</td>
</tr>
<tr>
<td><strong>Age</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 1 yrs</td>
<td>131</td>
<td>43.7</td>
</tr>
<tr>
<td>1-5 yrs</td>
<td>115</td>
<td>38.3</td>
</tr>
<tr>
<td>6-10 yrs</td>
<td>54</td>
<td>18.0</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>300</td>
<td>100.0</td>
</tr>
</tbody>
</table>

TABLE 1: Summary Statistics of VAS Score and Nirschl Scale
Figure 1 shows the percentage of children with G6PD level. Of the 300 children tested, 28(9.3%) were G6PD deficient and 272 (90.7%) cases had normal G6PD level. Out of 28 G6PD deficient children, twenty seven (96.4%) were moderately deficient while one (3.6%) was severely deficient.

Chi square value 11.16 p value= 0.001

The highest prevalence of G6PD deficiency occurred among children in the 1-5 years age group which was 53.6 % followed by 39.3% cases in the age group of less than 1 year and 7.1% cases in the age group of 6-10 years. (Table 3)

Table 2: Distribution of G6PD deficiency based on sex

<table>
<thead>
<tr>
<th>Gender</th>
<th>G6PD Level</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>≥2.9</td>
<td>166(86.5%)</td>
</tr>
<tr>
<td>Female</td>
<td>&lt;2.9</td>
<td>106(98.1%)</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>272(90.7%)</td>
</tr>
</tbody>
</table>

Table 3: Prevalence of G6PD deficiency in different age group

<table>
<thead>
<tr>
<th>Age group</th>
<th>G6PD level</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1 yrs</td>
<td>≥2.9</td>
<td>120(91.6%)</td>
</tr>
<tr>
<td>1-5 yrs</td>
<td>&lt;2.9</td>
<td>100(87.0%)</td>
</tr>
<tr>
<td>6-10 yrs</td>
<td></td>
<td>52(96.3%)</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>272(90.7%)</td>
</tr>
</tbody>
</table>

DISCUSSION

Glucose-6-phosphate dehydrogenase is the most common human enzyme deficiency worldwide. The clinical expression of G6PD deficiency encompasses a spectrum of disease severity related to the ability of red cells to generate NADPH.

In our study, 192(64%) were male children and 108 (36%) were female children which is similar to the study done by Albagshi et al and Isaac et al who reported 56.5% and 65.3% male children respectively.

In this study, out of 28 G6PD deficient children, 96% were moderately deficient while 4% were severely deficient. This is similar to the study done by Isaac et al where 70.2% cases were moderately deficient while 29.4% were severely deficient.

In our study, male children were found to have a higher incidence (13.5%) compared with females (1.9%) which is similar to the study conducted by William et al, Isaac et al and Hassan et al where the percentage of male children were 24.1%, 22.1% and 13.79% respectively. However, in a study conducted by al-Abdulkareem et al, the female population had a higher rate of G6PD deficiency (13.5%). This further reconfirms the natural history of G6PD deficiency of being an X linked recessive disorder, as well as the fact that only male hemizygotes and female homozygotes are the individuals most often affected. However, females can also be clinically affected because of the skewed lyonization of the X chromosomes.

In this study, the overall prevalence of G6PD deficiency was 9.3%. Our finding is consistent with earlier study conducted in Nepal by Gautam et al who reported 11% cases of G6PD deficiency. Similarly, a study conducted by India by Kumar et al, in China by Fu et al and in Iran by Moosazadeh et al showed an overall burden of 8.5%, 7.28% and 6.7% respectively.

WHO recommends population screening in regions where the prevalence of G6PD deficiency is 3–5% or more, but this has yet to become routine practice in many parts of Nepal. The barriers to screening include cost, underestimation of the public health impact of G6PD deficiency by the medical community, lack of awareness of G6PD deficiency among people and a paucity of guidelines regarding which high risk groups should be preferentially screened when general population screening is not possible.

As a significant proportion of G6PD deficient people were reported in our study and as G6PD deficient children are highly vulnerable to life threatening hemolysis, screening tests implementation and educational programs are warranted. Newborn screening (NBS) program needs to be implemented to determine the true prevalence prospectively. This program will, not only determine the prevalence of G6PD deficiency, but will also plan for future monitoring for jaundice, to prevent acute encephalopathy from hyperbilirubinemia encephalopathy, and consequent observation.
for future hemolytic episodes.

The management of G6PD enzyme deficiency should include avoidance of drugs and foods that predispose to hemolysis, provision of safe red cell transfusion to manage acute hemolysis in acutely affected children, and facility of dialysis services to treat acute renal failure.

CONCLUSION

The prevalence of G6PD deficiency was higher among children in the age group of 1 to 5 years with male preponderance. WHO recommends population screening in regions where the prevalence of G6PD deficiency is 3–5% or more, but this has yet to become routine practice in Nepal. The barriers to screening include cost, under estimation of the public health impact of G6PD deficiency by the medical community, lack of awareness of G6PD deficiency among people and a paucity of guidelines regarding which high risk groups should be preferentially screened when general population screening is not possible. There is a need for the routine screening and educational programs for G6PD deficiency in our setting to prevent grave complications in future. Further analysis of risk factors of drugs, food and family history should be done.

RECOMMENDATION

In the view of higher prevalence of G6PD deficiency in the children of 1 to 5 years age group, routine screening and education program on a regular basis needs to be considered.

LIMITATIONS OF THE STUDY

An important limitation to our study is the small number of patients and shorter duration. Our study reflects data from one center only and may not represent that of other centers across the country. Hence, multicenter trials would be necessary to determine the prevalence of G6PD deficiency in children. Also genetic analysis was not done in our study.

ACKNOWLEDGEMENT

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CONFLICT OF INTEREST

None

FINANCIAL DISCLOSURE

None

REFERENCES