G6PD DEFICIENCY AND THE MOST SEVER PRECIPITATING FACTOR IN BASRAH CITY[+]

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Abstract:

During two years period, 118 cases of infants and children under ten years of age with G6PD deficiency were diagnosed and managed in private medical clinic. When patients with G6PD deficiency ingested one of the precipitating factor like Ziziphus spina fruit *, broad bean, sun flower seeds and some drugs red blood cell hemolysis occur. The study aimed to find which of these factors more effective and severe in inducing RBCs hemolysis.

The ingestion of Ziziphus spina fruit, was detected in (51.7%) and was the most severe precipitating factor, followed by broad beans ingestion (30.5%)and ingestion of other foods and drugs (17.8) %

المستخلص

خلال سنتين من البحث تم تشخيص وعلاج ١١٨ حالة من الرضع والأطفال تحت سن عشرة سنوات من العمر مصابون بمرض نقص إنزيم G6PD في كريات الدم الحمر ، في عيادة خاصه للطب الباطني ، المرضى المصابون بنقص أنزيم G6PD عندما يتناولون أحد الأغذية التي تؤدي الى التكسر مثل النبق والباقلاء وحب زهرة الشمس وبعض الأدوية المعروفة يحصل انحلال كريات الدم الحمر ، في هذا العمل تم دراسة أي من هذه الأغذية أو الأدوية أكثر تأثيراً و خطورة في حصول التحلل.

وجد أن تناول النبق[**] في (٥١,٧%)من الحالات وكان أكثرها خطورة يليه تناول الباقلاء(٣٠,٥ %) ثم باقي المواد الغذائية والأدوية المعروفة طبياً (١٧,٨%).

Introduction:

G6PD is the first enzyme in the hexose mono phosphate shunt of Embden Myerhof glycolytic path way from which red cells derive most of their metabolic energy [1, 2, 3] The function of the hexose.mono phosphate shunt is to service the enzymes glutathione reducatase and glutathione peroxides, which protect the red cell against damage due to oxidation[1, 4, 5]. In the absence of G6PD this protective mechanism is crippled and certain foods and drugs in sufficient concentration can seriously injure the red cell.

G6PD deficiency is inherited as X- linked recessive disorder and has high frequency among African blacks, a similar deficiency occurs in Caucasian and Mongoloid races, where it is usually more sever. There has been a rise in the incidence of this disorder in other countries including Iraq specially Basrah city because of immigration [3,6,7].

Many foods and drugs in common use e.g Ziziphus spina fruit , Broad bean, seed of sun flower, some of antimalarials, sulfonamides, and chloramphenicol are capable of precipitating haemolysis in individuals with this defect.

G6PD deficiency is well known all over the world. Patients with G6PD deficiency normally enjoyed good health but are liable to haemolysis if any of incriminated drugs or foods are ingested [2, 10].

This research aimed to show which of incriminated foods or drugs are more effective, severe, and rapid in inducing hemolysis in Basrah city.

The study also attempted to investigate the relation between such prevalence and selected variables of age, sex, and seasonal pattern.

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Material and methods:

The research was conducted over two years period (December 1999-to November 2001) in private medical clinic. All cases presented with suspected jaundice were subjected for clinical examination and investigations .Blood specimens were collected and sent for hematological lab. for G6PD assay.

The diagnosis confirmed by estimating the G6PD activity of the red blood cells. A number of screening tests are also available e.g (1) the ascorbate cyanide test of Jacob and Jandl which monitor the whole glutathione regulating system of which G6PD is fundamental part ; (2) spot tests employing either the reduction of soluble tetra-zolium compounds to insoluble purple formazan or the fluorescence of NADPH which is abioproduct of G6PD activity .

A total of (118) cases of G6PD deficiency were diagnosed and followed during the period of the study. All cases were interrogated about ingestion of precipiting agents (foods or drug)

The amount of food and the dose of the drug and history of the same attack with the precipitating agent were also investigated.

Results

The number of patients with haemolytic jaundice due to G6PD deficiency precipitated by Ziziphus spina fruit were significantly higher (51.7%) than broad bean (30.5%) and others agents (17.8%) (Table 1) .

The haemolysis of red blood cells was more severe in Ziziphus spina fruit which led to admission of 9/61 (14.8%) of patients to the hospital, while in broad bean 3/36 (8.3%) of patients were admitted to the hospital but no patients needed hospital admission in the other agents (Table 2)

The onset of hemolysis after ingestion of the precipitating agent was more rapid after ingestion of Ziziphus spina fruit, (2- 4 days) than after ingestion of broad bean (3-7 days) and other agents (6- 10 days) (Table 3).

The seasonal distribution of G6PD is shown in (table 4) G6PD recovered through out the year, but the recovery rate was most frequent during spring 64 / 118(54.2%) and Autumn 25/118 (21.2 %) whereas it was less frequent during the remaining time of the year. G6PD deficiency was recovered in 83 / 118 (70. 3%) of male patient and 35/118(29.71%) of female (table 6), there was a statistical difference related to sex(X^2= 6.8)

G6PD deficiency was recovered in all age groups. The rate of recovery increased among higher age groups, in all precipitating agents which was around (7.6%) in up to two years age while it reached (31.4%) in (8-10)years age groups (table . 6) .

Table (1) G6PD deficiency in relation to precipitating factors

Precipitating factor	*No. of Cases*	*%*
Ziziphus spina	61	51.7%
Broad bean	36	30.5%
Others	21	17.8%

Table (2) No. of patient admitted to hospital in relation to precipitating factors.

Precipitating f.	**Total**	**No. of admission.**	**%**
Ziziphus spina	**61**	**9**	**14.8**
Broad beans	**36**	**3**	**8.3**
Others	**21**	**0**	**0**
Total	**118**	**12**	**10.2**

Table(3): Time of haemolysis onset in relation to precipitating factor.

Precipitating F.	*Time of haemolysis onset*
Ziziphus spina	2- 4 days
Broad beans	3- 7 days
Others	6-10 days

Table (4): Seasonal distribution of G6PD deficiency in relation to precipitating factor.

Season	*Ziziphus S. No. 61(%)*	*Broad b. No.36 (%)*	*No. of others No.21(%)*	*Total No.118(%)*
Winter	5 (8.2%)	5 (13.9%)	4 (19%)	14 (11.9%)
Spring	35 (57.41%)	17(47.2%)	12(57.1%)	64(54.2%)
Summer	1(1. 6%)	11(30. 6%)	3(14. 3%)	15(12.7%)
Autumn	20 (32.71%)	3 (8 . 3 %)	2 (9. 5%)	25 (21. 2%)

Table (5): Precipitation of haemolysis in relation to sex.

Precipitating F.	*No. Males(%)*	*No. females (%)*	*Total No.*
Ziziphus spina	47. (77%)	14 (23%)	61
Broad beans	24 (66. 6%)	12 (33. 4%)	36
Other	12 (57%)	9 (43 %)	21
Total	83 (70.3%)	35 (29.7%)	118

Table (6): Distribution of G6 PD deficiency in relation to age.

Age/ year	*Ziziphus S.*	*Broad beans*	*Other*	*Total %*
0-2	5 (8.2 %)	2 (5.5%)	2 (9.5 %)	9. (7.6%)
2-4	10 (16. 4%)	4 (11.1%)	2 (9. 5 %)	16 (13. 6%)
4-6	15 (24-6%)	9 (25%)	5 (23.8%)	29(24.6%)
6-8	13 (21.3%)	11(30.5%)	3 (14. 3%)	27 (22.9%)
8- 10	18 (29. 5%)	10 (27. 8%)	9 (42.8 %)	37 (31. 4%)

Discussion

G6PD deficiency was detected in 118 of jaundiced patients. Ziziphus spina fruit was one of the incriminated food that led to more hemolysis (51.5%) than in broad beans (30.5%) and other agents (17.8%). These results are higher than those reported in other studies [3,4,9], this might be due to higher consumption of Ziziphus spina fruit by children of Basrah city.

The incriminated foods or drugs which produce more rapid onset of haemolysis lead to sever anemia and more fatality.Ingestion of Ziziphus spina fruits lead to rapid onset of haemolysis due to rapid interference in the function of hexose mono phosphate shunt [2,5,6,8].

The seasonal prevalence of hemolysis after ingestion of these incriminated foods and the results related to sex were similar to that reported in scientific books [1, 5, 8].

There was predominant distribution rat of hemolysis among higher age groups in all incriminated foods and drugs, this seems slightly more than that reported in other studies [6, 9,10].

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