Original Article

Glucose – 6 – phosphate dehydrogenase deficiency In a group of Iraqi children

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Summary:

Background: *Glucose -6- phosphate deosphate dehydnogenase (G6PD) deficiency is a common problem in Iraqi as well as in tropical & subtropical region.*
Aim of study : *to study the epidemiological aspect of G6PD defiaiency among Iraqi children.*
Patients & Methods: *This cross sectional study was conducted in the Pediatric ward/AL – Kadhiymia Teaching Hospital during the season of Fava beans ingestion. Patients who were presented with acute attack of pallor were collected , history was taken and physical examination was done.*
Results : *Ninety seven cases were studied, males were affected more than female with a ratio of 3.85:1, the peak of age was between (1 – 5years), 76 cases ,(78.36%).Previous history of neonatal jaundice was found in 24 cases (24.74%), the acute hemolytic attack was precipitated by Fava beans ingestion in all of the cases (100 %) and pallor was the main presenting feature in all of them. The attack was very severe in 43 cases (44.32 %) with arrange of PCV between (1 – 9 %). History of recurrence was encountered in 11 cases (11.34 %), family history of G6PD deficiency was found in 22 cases (22.68%) .G6PD level was estimated two months later in 31 cases (31.95 %) only and were found to be deficient.*

J Fac Med Baghdad
2005; vol.47 No. 1
Received:June 2004
Accepted: Oct.2004

Introduction

Glucose – 6 – phosphate dehydrogenase(G6PD) deficiency is a common problem in Iraq as well as in tropical and subtropical region [1] .As a result of recent migrations G6PD had become widely spread in many other area of Europe and America[1] . The mode of inheritance is x – linked recessive [2] , although most of the affected individuals are asymptomatic , there is a risk of neonatal jaundice and acute hemolytic anemia triggered by infection and ingestion of certain drugs and broad beans [3] .Henna is one of he oxidant compound responsible for acute hemolytic attack [4] . Both extra and intravascular hemolysis are seen in G6PD deficiency [1].

Aim of the study:

Is to study the epidemiological aspect of G6PD deficiency among Iraqi children.

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Patients and method:

A cross sectional study was conducted in the Pediatric ward / AL- Kadhiymia Teaching Hospital during the season of Fava beans ingestion . Patients who were presented with acute attack of pallor and / or jaundice were collected . Information was taken including age , sex , history of Fava beans ingestion, preceded infection , acidosis, past history of neonatal jaundice , history of previous similar attack , family history of the disease . Physical examination was performed looking for pallor , jaundice , fever , presence or absence of hepatosplenomegaly .Complete blood picture and reticulocytes count were done with PCV estimation after blood transfusion . G6PD level was estimated two months after the acute attack using the methaemoglubin reduction test .

Result:

Total number collected was 97 cases , 77 cases (79.39 %) were males and 20 cases (20.61%) were females , male: female ratio equal to 3.85 : 1 . The peak age was between (1 – 5) years , 76 cases (78.36 %) as it is shown in(table – 1).Previous history of neonatal jaundice was found in 24 cases (78.36%) , 2 of them required exchange transfusion (8.33%), while the remaining 22cases were treated by phototherapy(91.67 %). All of the attacks were precipitated by Fava

beans ingestion (100%) , the attacks of hemolysis occur within 48 hours in 66 cases (68.05%) after the ingestion as it is shown in (table – 2).Non had previous history of infection, acidosis Pallor , jaundice and darkly colored urine were the main clinical manifestations as it is shown in (table – 3). The attack of hemolysis was very severe in 43 cases(44.32 %) with a pcv in the range of (1 – 9%)as it is shown in (table – 4) .Blood transfusion was required in all of the cases ,some of them were transfused twice in order to raise the hemoglobin to a safe level, pcv after blood transfusion is shown in(table – 5).Recurrent attacks of hemolysis were found in 11 cases only (11.34 %), most of them were above 10 years of age. Positive family history was detected in 22 cases only (22.68%). G6PD level was estimated two months later in 31 cases (31.95 %) by the methaemoglubin reduction test , all of them were deficient for the enzyme , the remaining patients did not come back for follow up.

Table -1 shows age and sex distribution

Age/ years	Males		Females	
	No.	%	No.	%
1 – 5	59	76.63	17	85
5 – 10	13	16.88	3	15
>10	5	6.49	o	/

Table – 2 shows the onset of hemolysis

Onset / days	No.	%
1	10	10.30
2	66	68.05
3	21	21.65

Table – 3- shows the clinical manifestation

Clinical features	No.	%
Pallor	97	100
Jaundice	97	100
Dark urine	97	100
Fever	36	37.11
Malaise	81	83.50
Abdominal pain	70	72.16

Table – 4 – shows PCV level before blood transfusion

PCV %	No.	%
1 – 9	43	44.32
10 – 19	46	47.43
20 - 29	8	8.25

Table – 5 shows PCV level after blood transfusion

PCV%	No.	%
25 -29	43	44.32
30 - 35	54	55.68

Discussion:

In this study males were affected more than females with a ratio of (3.85 :1) , previous study in Iraq gave a ratio of(6.5 :1) [5] , in Saudi Arabia the ratio was(7 :1) [6] , in Italy was(6.2:1)[7] , in Greece was (10:1) [8] .The predominance of male over female can be expressed by the mode of inheritance(sex linked) [2] .Female can express the disease when the number of x – chromosome carrying the abnormal gene is in excess of the number of chromosome carrying the normal gene which cause random x – inactivation in females and consequently variable expression of the enzyme [9] .

The peak of age was between(1 -5) years which is in agreement of previous studies [6,7] ,the decline in the prevalence of the disease with age could be due to the avoidance of eating Fava beans and could point to an ill defined factors that manipulate the susceptibility to hemolysis in favism that decline or totally disappear as the child get older [10] .

Previous history of neonatal jaundice was found in (24.74%) of cases ,in a study done in Basrah G6PD deficiency was responsible for (46%) of neonatal jaundice [11] , in Jordan (21%) [12] ,in France (2.6%) [13] .

All of the attacks were acute(100%) and occur within 48 – 72 hours of Fava beans ingestion , non was caused by preceded infection or drug ingestion. In Jordan (86.1%) of the attacks were precipitated by Fava beans and (13.9%) were due to infection [14] ,in Turkey infection and drugs were the main offending agents [15] .

The pcv level gives an idea about the severity of hemolysis which was in the range of (1 -9%) and (10 – 19%) in the majority ,and some of the patients were transfused twice which indicate severe degree of hemolysis ,the same was noticed by Barwari in 1997 in Mousul [16] .In Jordanian study the hemoglobin level was between(6.4 -6.6) g/dl in most of the cases [14] .

Most of the cases presented for the first time , recurrence was seen in (11.34%), in a previous study done in Iraq recurrence encountered in (6.6%) [5] .

Family history was found in (22.28%) , In Jordan family history noticed in (31.47%) [15] . Absence of family history can be explained by new mutation [7] .

G6PD level was estimated two months later using the methaemoglubin test ,which is the test available at the when the research was done, all of the patients were deficient , this is a semi-quantitative method and meant to classify a sample simply as deficient or normal so every patient found to be deficient should be confirmed by spectrophotometric test [17] , when it is available.

References:

1- Lucio Luzzatto,Edward C.Gorden- Smith, The inherited hemolytic anemia: In post graduate hematology by A. V. Hoffbrand ,4th ed. ,ch.7 p.120 - 143 , 1999 ,Butter Worth Heinmann.

2- Behrmann PE ,Kliegman CM, Avin AM , Nelson text book of pediatric , 17th ed. ,ch. 420 , 2004,Philadelphia , W.B. Saunders company.

3- Mehta A ,Mason PJ ,Vulliamy TJ, glucose - 6- phosphate dehydrogenase deficiency ,Baillieres – Best – Pract – Res- Clin – Hematol,2000.Mar13(1) ,21 – 38.

4- William H. Zinkham ,Frank A. Oski, Henna :A potential cause of oxidative hemolysis and neonatal hyperbilirubinaemia ,pediatrics ,June ,vol.97 ,No.6 , 1996 ,25 – 27.

5- Sabri S., epidemiology of favism in Iraq ,A Thesis submitted for the degree of diploma in public health , Baghdad , 1975.

6- EL – Hazmi M.A.F.,Worsy A.S. glucose -6- phosphate dehydrogenase phenotyping in different region of Saudi Arabia ,A comparative assessment ,Saudi Medical J. ,vol.18(4) 1997, 393 -399.

7- Kattamis C.A. ,Kryriazakon M.,Chaides S. ,favism , clinical and biochemical data ,J.Med. Genet .,6, 1969 34- 41

8- Meloni T., Forteleoni N. , Dore A. ,Cutillo S., Favism and hemolytic anemia in glucose -6- phosphate dehydrogenase deficient subjects in North Sardinia ,Acta hematol ,70 ,1983 , 83 -90.

9- Sheth S. B. ,Hynicka W. Kazierad D. J. ,Davis H. , A (-) variant glucose -6- phosphate dehydrogenase deficiency in African women, abstract of the American society of hematology ,40th annual meeting ,Dec, 4 -8 ,1998 ,Miami Beach, Florida.

10- L . Luzzatto , D. Doper , membrane and enzyme abnormalities:I n practical hematology by John V. Dacie and S.M.Lewis 8th ed. ,ch. 13 ,1995, 225 – 235.

11- Lamia M. AL – Naama,Imad AL – Sadoon ,Menhal M. AL – Naama , neonatal jaundice and glucose -6- phosphate dehydrogenase deficiency in Basrah ,Annals of tropical pediatrics 1987 , 7 ,134 – 138.

12- Abu – Ekteisn F.,Daoud A., Pimawi H. Kakish K., Abu – Heija A. ,neonatal exchange transfusion , a Jordanian experience ,Ann. Trop. Pediatric , 2000 , Mar 20 (1) ,57 -60.

13- C. Badans , M. Leclaire ,J. Cllomb et al , Dēficiet en glucose -6- phosphate dēshydrogēnase et ictēre nēonatal , la press mēdical ,2001, 30 , 524 – 6 , Masson ,Paris.

14- Nawaf Momani , Awni Madani , Mufeed Hammoury ,et al , glucose -6- phosphate dehydrogenase deficiency in the north of Jordan , Iraqi medical j. , vol. 50 , No. 1, 2001 ,151 -158.

15- Kilink Y. ,Kumi M., hemolytic crisis due to glucose -6- phosphate dehydrogenase deficiency in the mid southern region of Turkey ,Acta pediatr. Scand. ,1990 , 79 (11) ,1075.

16- Barwari Z.A. M. ,Favism in Mousul , a clinico – hematological study , A thesis submitted to the Iraqi council of medical specialization in pediatrics , 1997 ,10 -20.

17- Luccio Luzzatto , glucose -6- phosphate dehydrogenase deficiency and hemolytic anemia : In Nathan and Oski ,Hematology of infancy and childhood ,5th ed. ,1998 , vol.1 , 704 – 762 , W.B. Saunders company.