Glucose-6-Phosphate Dehydrogenase Deficiency in Neonatal Hyperbilirubinaemia in Maternity Hospital Kathmandu

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Introduction:

Glucose-6- Phosphate dehydrogenase (G-6-PD) is essential to maintain stability of red blood cells. Deficiency of this enzyme, an x-linked recessive disorder may manifest as spontaneous non spherocytic haemolytic anaemia, drug or infection induced haemolytic anaemia or haemolytic disease of the new born. Neonates with Hyperbilirubinaemia constitute a high risk group in whom the prevalence of G-6-PD deficiency is likely to be high. Prevalence of G-6-PD deficiency ranges widely in different parts of world and in different ethnic group. Neonatal jaundice is more common if infection occur or if oxidant drugs are given to the mother in late pregnancy or to the neonate. Infection can precipitate haemolysis at all ages. Drugs like antimalarials, sulphonamide, nitrofurantoin, aspirin, PAS, naphthelene and aniline dyes and broad bean can cause severe haemolysis.

Objective:

This study was undertaken to ascertain the role of G-6-PD deficiency in neonatal hyperbilirubinaemia in this hospital and to determine the need of routine screening of all neonates for G-6-PD deficiency.

Methodology

This study was conducted among neonates with hyperbilirubinaemia admitted to Maternity Hospital for 6 months from 1st Jestha to 30th Kartik 2056. Bilirubin level of all babies who developed clinically moderate jaundice either in post natal ward or SCBU/NICU was determined and all term babies with total serum bilirubin level of more than 12 mg/dl and all preterm with serum bilirubin exceeding 15 mg/dl were enrolled in the study, Babies born

outside the hospital and the babies who left the hospital before full investigation were done were excluded from the study. Maturity of those babies was determined by Parkin score. For G-6-PD enzyme assay blood was sent to laboratory and analyzed in the laboratory by Methaemoglobin reduction test. Blood was also sent for blood grouping, coomb's test and reticulocyte count. Reg. no, patients name, address, ethnic group, date of birth, birth weight, time of appearance of clinically moderate jaundice, time of maximum bilirubin level, hacmoglobin level, type of delivery, types of management of hyperbilirubinaemia and its outcome were entered in the structured questionnaire. For the neonates with positive Methaemoglobin reduction test, history of NNJ in previous child and its outcome was also noted. Parents of neonates with positive Methaemoglobin reduction test were also screened for G6PD deficiency. Data entry and statistical analysis were done partially manually and partially on computer (Window MS EXCEL).

Results '

of kernicterus

172 neonates with hyperbilirubinaemia were enrolled in the study. 11 (6.39%) were found to have deficiency of G6PD. The frequency of G6PD deficiency was similar in neonates of different ethnic group and gestational age. Maximum level of TSB and time of maximum level of TSB were similar in G-6-PD deficient and normal babies. Maximum total serum bilirubin did not exceed 20 mg% in deficient babies. But the haemoglobin level was lower and reticulocyte count were higher in G6PD deficient neonates. No positive history of maternal ingestion of oxidants drugs were found. Among the parents of G6PD deficient babies only one mother was found to have G6PD deficiency. All cases of NNJ improved with phototherapy alone and none of them required exchange transfusion. None of the babies developed features

Discussion.

About 13% of American black males and 2 of black female have G-6-PD deficiency.1 Italians, Greeks and other Mediterranean, Middle Eastern, oriental ethnic group also have high frequency ranging from 5-40% and upto 10% in African Negroes.2 In India G-6-PD deficiency ranges widely from a low as 0.2% to high as 19%.3 In a study conducted in South Indian referral hospital, 11.8% of newborns investigated for Hyperbilirubinaemia were found to have G-6-PD deficiency.4 Similar studies done in Chandigarh and Delhi in India and in Thailand12% with Hyperbilirubinaemia were G-6-PD deficienct.4 In this study of newborns investigated for Hyperbilirubinaemia 6.39% were found to have G-6 PD deficiecy. G6PD defeciency was found in 4.8% of neonates in one screening study conducted in 500 neonates in maternity hospital, Kathmandu in 1993.5 However there is no data from other parts of Nepal to indicate the extent to which G-6-PD deficiency contributes to neonatal hyperbilirubinacmia and the prevalence of G-6-PD in the country is not known. Since the actual prevalence of G6PD deficiency in the country is not known, routine screening of all newborn is not feasible or cost effective. Till the actual prevalence is known by screening test with larger sample size, we recommend to identify G6PD deficiency in all babies who develop significant NNJ so that appropriate counseling is provided to the family and potentially harmful drugs and foods are not given to these infants and their nursing mother.

TABLE 1- Number, sex, maturity

TOTAL NO.	172		
MALE	82 (47.7%)	FEMALE	90 (52.3%)
TERM	136 (79%)	PRETERM	36 (21 %)

TABLE II - Methemoglobin positive cases (G6PD deficient)

TOTAL NO.	11(6.39%)		
MALE	5 (45.5%)	FEMALE	6 (54.5%)
TERM	8(5.8%)	PRETERM	3 (8.3%)

TABLE III- Ethnic group

G6PD NORMAL		G6PD DEFICIENT	
BRAHMIN	33(19.18 %)	2 (18.18%)	
CHHERTI	36 (20.93 %)	3 (27.27%)	
NEWAR	50 (29.06 %)	2 (18.18%)	
MAGAR	22 (12.79 %)	2 (18.18%)	
OTHER	31 (18.02%)	2 (18.18%)	

TABLE IV-TIME OF APPEARANCE OF CLINICALLY MODERATE JAUNDICE

TIME IN HOUR.	G6PD NORMAL	G6PD DEFICIENT
<24 HRS.	13 (8.07%)	2 (18.18%)
>24HRS<48 HRS	28 (17.4%)	3 (27.27 %)
>48 HRS<72 HRS	49 (30.43%)	5 (45.45 %)
>24 HRS<96 HRS	37 (22.98%)	1 (9.09 %)
>24 HRS<120 HRS	20 (12.42%)	0
>24 HRS<144 HRS	11 (6.83 %)	0
>24 HRS<168 HRS	3 (1.86 %)	0
MEAN / SD	49.54 HRS /23.33	73.62 HRS / 33.8

TABLE V-LEVEL OF MAX. TSB (TERM BABIES)

	G6PD NORMAL	G6PD DEFICIENT
12-15 Mg%	29 (22.64 %)	2 (25 %)
15.1-20 Mg %	82 (64 %)	6 (75 %)
> 20 Mg %	17 (13.28 %)	0
MEAN /SD	17.2/2.61	16.89/1.8

TABLE VI-LEVEL OF MAX. TSB (PRETERM BABIES)

	G6PD NORMAL	G6PD DEFICIENT
		<u> </u>
15-20 Mg %	28 (84.85 %)	3 (100 %)
> 20 Mg %	5 (15.15%)	o
MEAN/ SD	18.06 /1.75	16.67/1.53

TABLE VII- Haemoglobin level

	G6PD NORMAL	G6PD DEFICIENT
<10 gm	4(2.48%)	0
10.1-12 gm	12(7.45%)	4(36.36%)
12.1- 14gm	54(33.54%)	7 (63.63)%
14.1-16gm	- 60 (37.26%)	0
16.1 –18 gm	25(15.52%)	0
18.1 –20 gm	5 (3.1%)	o
> 20 gm	1 (0.62%)	0
Mean	14.77(+/- 1.71)	12.64 (+/-0.94)
SD	2.19	1,23

TABLE VIII- Reticulocyte count

	G6PD NORMAL	G6PD DEFICIENT
Upto 1 %	76 (47.2%)	1 (9.09%)
1.1- 3%	55 (34.16%)	2 (18.18%)
3.1% -6%	22 (13.64%)	5 (45.45%)
>6%	8 (4.96%)	3 (27.27%)
Mean	2.14 (+/- 1.19%)	4.04 (+/-1.78%
SD	1.86	2.31

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Study of C	5-6-P-D Deficiency
Name	Reg. No
Address	
Sex M F	
Pithnic Group Brahmin Chhetri Nev	var Magar Other
Date of birth and Time	Birth wt Kg
Type of delivery ND CS Forcep	Vaccum Preterm Twin Breech
Parity	
Parkin Score Matur	ity by Parking Score Wks
Time of appearance of Jaundice	Hrs.
Max .Bilirubin level	Time in Hour
Haemoglobin Level	when the bilirubin level is max.
Blood group of mother	Blood group of baby
Coombs test Positive Negative	
Methaemoglobin Reduction Test [Positive Negative
Type of Management of Hyperbilirubinaer	nia Phototherapy Exchange Transfusion
Outcome Cured NND LAMA	

To be filled up only if Methaemoglobin Reduction Test is Positive

	Yes No	
Yes No	Don't know	
Cured	NND	
r		
+	ve -ve	
The state of the s		
4-1	ve - ve	
	Cured +	

Signature

If yes, Name of drug