Journal of American Science

Websites:
http://www.jofamericanscience.org
http://www.sciencepub.net

Emails:
editor@sciencepub.net
sciencepub@gmail.com

MARSLAND PRESS
Multidisciplinary Academic Journal Publisher

Effect of Transfusion of G6PD Deficient Blood to Children with B Thalassemia in Pediatric Department of Zagazig University Hospitals

Ehab Mahmoud Rasheed[1], Adel Sherif Ahmed[1], Ahmed Mohamed Gaballah[2], Dina Essam Moustafa[1]

[1] Pediatrics Department, Faculty of Medicine, Zagazig University, Egypt.
[2] Clinical Pathology Department, Faculty of Medicine, Zagazig University, Egypt.

Abstract: Background: B-thalassemia is the most common inherited single gene disorder worldwide, and glucose-6-phosphate dehydrogenase (G6PD) deficiency is the most common human enzyme deficiency. The goal of this study was to assess the prevalence of G6PD deficiency in transfused blood and its hazards if transfused to thalassemiac children. **Methods:** We examined 60 patients who were selected by random sampling. For diagnosis of thalassemia, complete blood count and hemoglobin electrophoresis were carried out and for G6PD deficiency, fluorescent spot test methods were used as a screening test. This study was cross sectional study which was done in Pediatric Hematology unit & Clinical Pathology Departments in Zagazig University Hospitals during period from July 2017 to August 2018. **Results:** This study included 60 patients with age ranged from one year to 12 years with mean ± SD age of the studied group is 6.81± 2.89 years, while sex distribution was 37male patients (61.7%) and 23 female patients (38.3%). Age of diagnosis of thalassemia was 7.12±1.5 month and ranged 6-11 months. Calculation of Z-score of G6PD in Blood bag it is (-1.56-3.27Unit/gram), as regard ROC curve for G6PD in Blood bag, Area under the curve =0.904, Cutoff level of G6PD≥ 7.19 U/g (Unit/gram) have sensitivity of 97.6%, and specificity of 57.9% in predicting sufficient increase of hemoglobin level after transfusion with high statistical significance. **Conclusion:** Whereas the frequency of b-thalassemia among patients is higher, the frequency of G6PD deficiency was not significantly different in the two populations. Prevalence of G6PD level in blood bag was found to be normal 48 bags (80%) and in deficient in the remaining 12 bags (20%).

[Ehab Mahmoud Rasheed, Adel Sherif Ahmed, Ahmed Mohamed Gaballah, Dina Essam Moustafa. **Effect of Transfusion of G6PD Deficient Blood to Children with B Thalassemia in Pediatric Department of Zagazig University Hospitals.** *J Am Sci* 2020;16(3):22-30]. ISSN 1545-1003 (print); ISSN 2375-7264 (online). http://www.jofamericanscience.org. 4. doi:10.7537/marsjas160320.04.

Key words: Thalassemia, 6-phosphogluconate dehydrogenase, Glucose-6-phosphate dehydrogenase, Complete blood count, Human immunodeficiency virus.

1. Introduction

Thalassemia is one of the most prevalent causes of chronic hemolysis worldwide and the commonest cause of chronic hemolysis in Egypt. It results from hemoglobinopathies, consisted of 2 main types (thalassemia major and thalassemia minor) and each type has many different subtypes (1).

Beta-thalassemia major (β-thalassemia) presents after 6 months of age with symptoms of chronic hemolysis as facial bone deformities, fatigue, growth failure, dyspnea, jaundice and hepato-splenomegaly (2).

Thalassemic patients need chronic transfusion regimen with suitable packed red blood cells (PRBCS). Therefore PRBCS must be healthy and devoid of any hemolytic liability before or during and after transfusion (2).

Glucose 6 phosphate dehydrogenase (G6PD) is the enzyme responsible for catalyzing the first step in the pentose phosphate pathway (the hexose monophosphate shunt) to generate Nicotineamide Adenine Dinucleotide Phosphate (NADPH) which is subsequently utilized in processes that reduce hydrogen peroxide (H2O2) to water, ameliorating oxidative stress in red blood cells (RBCs). Approximately 400 million people worldwide are G6PD deficient making it the most common human enzyme deficiency (3).

Clinical manifestations of G6PD deficiency include neonatal jaundice and acute hemolytic anemia arising from the oxidative stresses on RBCs, as drug exposure, infections and refrigerated storage of the deficient blood which leads to decreased recovery in vivo after transfusion with several cases had manifested signs of hemolysis in vivo following transfusion of G6PD-deficient RBCs (4).

From previous reported cases, accelerated destruction of G6PD deficient RBCs, either in the storage bag or following transfusion may result in increased levels of extracellular hemoglobin.

Hemolysis-associated pathologies including smooth muscle dystonia, vasculopathy and endothelial dysfunction are believed to be caused by nitric oxide scavenging by cell-free hemoglobin (5).

Regarding RBCs transfusion, previous few studies have examined the activity of the RBCs G6PD activity during storage and after transfusion, which showed 35% decrease in the activity during the storage, also showed shorter half-life of G6PD deficient blood even in absence of any oxidative factors. In addition, the 24 hour post transfusion recovery of G6PD deficient RBCs following refrigerated storage is significantly less than that for normal RBCs, despite that, blood donors are not routinely screened for G6PD deficiency and the blood centers differ in their deferral of donors with known G6PD deficiency (6).

The World Health Organization (WHO) suggests that policies for the assessment of prospective donors should be developed in regions where there is a high incidence of RBC enzymopathies such as G6PD deficiency and inherited membrane defects. According to the WHO Guidelines on assessing donor suitability for blood donation, individuals with G6PD deficiency who do not have a history of hemolysis may be accepted for donation, however, blood from these donors should not be used for intrauterine transfusion, neonatal exchange transfusion, or for G6PD deficient patients. In addition the WHO recommends deferring G6PD-deficient donors with a history of hemolysis (7).

This study aimed to assess the prevalence of G6PD deficiency in transfused blood and its hazards if transfused to thalassemiac children.

2. Patients and Methods

Study design:

This study was cross sectional study which was done in Pediatric Hematology unit & Clinical Pathology Departments in Zagazig University Hospitals during period from July 2017 to August 2018.

Sample size:

Assuming to attendance 1000 of thalassemia patient and frequency of G6PD deficiency transfused 60%, sample was calculated by EPI-program to be 60 patients. At confidence level 95% and power test 80%.

Inclusion criteria:

- Age and sex: children age group from 6 months age up to 16 years old patients in both sexes.
- All patients presenting with thalassemia major children and need repeated blood transfusion.
- Approval was taken from the institutional review boaed (TRB) of faculty of medicine Zagazig University and from patients or there cares.

Exclusion criteria:

- Pediatric patients suffer from hemolysis due to other causes rather than thalassemia children and need blood transfusion.
- Refusal of patient or their cares to participle in the study.

Operational design:

Type of study:

Cross-sectional study.

Methods:

All cases enrolled in the study will be subjected to the following:

1) Detailed history taking from patients and their files with special emphasis on age, sex, residence, family history of thalassemia, age of diagnosis of it and ABO blood group. etc.

2) Through and complete clinical examination.

3) Detailed routine Laboratory investigations from patients files including:

a. CBC, reticlocytic count, electrophoresis and serum ferritin.

b. Liver function test (Total bilirubin, indirect bilirubin, AST and ALT).

c. Kidney function test (Urea and creatinine).

4) Period of storage (in days) was obtained from the blood bags before transfusion to patient.

5) G6PD was assessed by sampling the blood bags before transfusion.

6) Follow up of the cases after transfusion:

- Amount of transfusion needed (CC/kg of packed RBCs) and frequency of transfusion.
- Post-transfusion HB level and HCT after.
- Clinical scenario with any reported complications.
- Pre next-transfusion HB level and HCT.

G6PD level in blood bag:

Calculation of Z-score of G6PD in Blood bag it is (-1.56-3.27Unit/gram), as regard ROC curve for G6PD in Blood bag, Area under the curve =0.904, Cutoff level of G6PD≥ 7.19 U/g (Unit/gram) have sensitivity of 97.6%, and specificity of 57.9% in predicting sufficient increase of hemoglobin level after transfusion with high statistical significance.

- Normal G6PD ≥ 7.19 -18 U/g of hemoglobin.
- Deficient G6PD < 7.19 U/g.

Storage of blood bag was classified into (Garcia-Roa, et al.,2017):

- Storage blood bag < 20days.
- Storages blood bag ≥20days.

G6PD measurements:

Assay principle:

- G6PD assay was done on whole blood using cobas C311 chemistry analyser Roche diagnostics.

Sample Collection:

- 1ml of citrated blood from transfused bags collected on sterile plane tube and immediately transfused to the lab for analysis.
- Hemoglobin level in the sample was measured and G6PD value were calculated as U (enzyme/activity)/g hemoglobin.

Statistical analysis:

The collected data were analyzed by computer using Statistical Package of Social Services version 24 (SPSS), Data were represented in tables and graphs, Continuous Quantitative variables e.g. age were expressed as the mean ± SD & median (range), and categorical qualitative variables were expressed as absolute frequencies (number) & relative frequencies (percentage). Suitable statistical tests of significance were used after checked for normality. The results were considered statistically significant when the significant probability was less than 0.05 (P <0.05). P-value < 0.001 was considered highly statistically significant (HS), and P-value ≥ 0.05 was considered statistically insignificant (NS).

3. Result

This table (Table 1) shows that the mean age of the studied group is 6.81± 2.89 years, with a range from 1 to 12 years. About 62% of the studied group were males and 38 % were females.

This table (Table 2) shows that mean of age of diagnosis of thalassemia was 7.12±1.5 month and ranged 6-11 months, most of the studied group having negative family history of thalassemia (90 %), while 1/3 of them have splenomegaly (33.3 %), and only 15% of them had splenectomy, regarding blood grouping O+, B+ and AB+ are the most prevalent blood group (31.7%, 21.7% and 18.3%) respectively.

Table (1): Demographic data of the studied β-thalassemia patients (N=60).

Demographic data	Studied children (N=60)	
	No.	%
Age (years):		
Mean ± SD	6.81± 2.89	
Median (Range)	6.75(1-12)	
Sex:		
Male	37	62
Female	23	38

Table (2): Present history, clinical picture and blood group distribution of the studied β-thalassemia children:

Item	Studied children (N=60)	
	No.	%
Age of diagnosis of thalassemia (months):		
• Mean ± SD	7.12±1.5	
• Median (Range)	7(6-11)	
Family history of thalassemia:		
• Positive	6	10.0
• Negative	54	90.0
Splenomegaly:		
• Positive	20	33.3
• Negative	40	66.7
Splenectomy:		
• Positive	9	15.0
• Negative	51	85.0
ABO-blood group:		
• **A**+ve	10	16.7
• **A**-ve	3	5.0
• **B**+ve	13	21.7
• **B**-ve	1	1.7
• **AB**+ve	11	18.3
• **O**+ve	19	31.7
• **O**-ve	3	5.0

Table (3): Hemoglobin, hematocrit and serum ferritin level among the studied β– thalassemiachildren:

Item	Studied children (N=60)
Hemoglobin (gm/dl):	
▪ Mean ± SD	4.82 ± 1.01
▪ Median (Range)	5 (3-6.3)
HCT (%):	
▪ Mean ± SD	22.08 ± 2.26
▪ Median (Range)	22.3 (11.9-26)
Serum Ferritin level (mg/dl):	
▪ Mean ± SD	2579.4±1633.72
▪ Median (Range)	2012.5(600-7000)

This table (Table 4) shows that the mean of hemoglobin level among the studied patients was4.82 ± 1.01 gm/dl, with a range from (3-6.3), HCT level was 22.08 ± 2.26% with range from (11.9-26) and mean of serum ferritin level was 2579.4±1633.72 mg/dl, with a range from (600-7000).

Table (4): Laboratory data among the studied β-thalassemiachildren:

Item	Studied children (N=60)
Liver function tests:	
Total Bilirubin (mg/dl):	
▪ Mean ± SD	2.03±0.09
▪ Median (Range)	1.65(1-3)
Direct Bilirubin (mg/dl):	
▪ Mean ± SD	0.10±0.07
▪ Median (Range)	0.10(0.01-0.27)
ALT (I. U):	
▪ Mean ± SD	61±6.49
▪ Median (Range)	65.5(29-89)
AST (I. U):	
▪ Mean ± SD	61.99±26.10
▪ Median (Range)	49.5(20-100)
Kidney function tests:	
Urea (mg/dl):	
▪ Mean ± SD	26.65±5.58
▪ Median (Range)	25.5(20-44)
Creatinine (mg/dl):	
▪ Mean ± SD	0.75±0.10
▪ Median (Range)	0.7(0.6-1.0)

This table (Table 5) shows that the mean of total bilirubin among the studied group is 2.03±0.09 mg/dl with a range from (1-3) and mean of direct bilirubin is 0.10±0.07 mg/dl with range (0.01-0.27), while mean of ALT is 61±6.49 I. U with range (29 -89) and mean of AST is 61.99±26.10 I. U with range (20-100). The mean of urea is 26.65±5.58 mg/dl with range (20-44) and mean of creatinine is 0.75±0.10 mg/dl with a range from (0.6-1.0).

Table (5): Characters of transfused blood among the studied β-thalassemiachildren:

Item	Studied children (N=60)
Amount of blood Transfusion /ml:	
▪ Mean ± SD	235.33 ± 71.7
▪ Median (Range)	250 (100-350)
Blood bag Storage (days):	
▪ Mean ± SD	15.17 ± 8.56
▪ Median (Range)	11(5-40)
G6PD in Blood bag U/g:	
▪ Mean ± SD	13.95±5.73
▪ Median (Range)	14.87(5-32.69)

This table (Table 6) shows that amount of blood transfusion in the studied beta-thalassemia children ranged from (100-350 ml) and it's mean is 235.33 ± 71.7 ml, mean storage duration of transfused blood is15.17 ± 8.56 days, with a range from (5-40) days, while mean of G6PD in Blood bag is 13.95±5.73 U/g with a range (5-32.69).

Table (6): Frequency of blood transfusion among the studied β-thalassemia children:

Frequency of transfusion	Studied children (N=60)	
	No.	%
• Each 2 weeks	13	21.7
• Each 3 weeks	13	21.7
• Each 25 days	1	1.7
• Each 4 weeks	33	55.0

This table (Table 7) shows that more than half of beta-thalassemia children receive blood transfusion every 4 week (55%).

Table (7): Increase of HB*after blood transfusion among the studied β – thalassemia children:

Item	Studied children (N=60)	
	No.	%
• Sufficient **	41	68.3
• Insufficient ***	19	31.7

This table (Table 8) shows that there is insufficient increase of HB after blood transfusion in about 1/3 of the studied patients (31.7 %).

Table (8): Prevalence of G6PD deficiency in blood bags:

G6PD deficiency (Unit/gram of hemoglobin).	Blood bags (N=60)	
	No.	%
• Deficient G6PD <7.19U/g	12	20.0
• Normal G6PD ≥7.19 U/g	48	80.0

This table (Table 9) shows that there is deficiency of G6PD below 7.19 cutoff level in 20% of the studied blood bags.

Table (9): Correlation between G6PD in Blood bag, duration of blood bag storage, hemoglobin after transfusion, HCT After transfusion and hemoglobin pre-next transfusion among the studied β_ thalassemia children:

Variable	Correlation co-efficient (r)			
	G6PD in Blood bag	Blood bag Storage (day)	HB after transfusion	HCT After transfusion
G6PD in Blood bag.	1.000			
Blood bag Storage (day)	**-.624****	1.000		
HB after transfusion	.212	**-.377****	1.000	
HCT after transfusion	.247	**-.354****	**.927****	1.000
HB pre-Next transfusion	**.306***	-.238	**.749****	**.780****

**Correlation is significant at the 0.05 level

There is significant negative correlation between G6PD in blood bag level and duration of blood bag storage (r =-0.624, P-value >0.05), also there is significant negative correlation between duration of blood bag storage and hemoglobin after transfusion, HCT after transfusion. There is significant positive correlation between hemoglobin pre-next transfusion, hemoglobin after transfusion, HCT after transfusion and fresh blood transfusion, which means that hemoglobin level in the pre-next transfusion will be high if G6PD in Blood bag level is high (fresh blood used) (Table 10).

Table (10): Relation between G6PD deficiency in blood bags and insufficient increase of Hb level after blood transfusion among the studied β-thalassemia children:

Increase in HB	G6PD in the studied blood bags (N=60)				chi- square test	P- value
	Deficient G6PD <7.19 (N=12)		Normal G6PD ≥7.19 (N=48)			
	No.	%	No.	%		
Sufficient	1	8.3	40	83.3	fisher's exact	0.000* (HS)
Insufficient	11	91.7	8	16.7		

Among deficient blood bags (G6PD below 7.19 cutoff level), only 8.3 % have sufficient increase of hemoglobin, while after transfusion of blood bags with normal level of G6PD ≥ 7.19, most of patients (83.3%) have sufficient increase of HB level, with high statistical significance (Table 11).

Table (11): Relation between G6PD deficiency in blood bags, insufficient increase of Hb level and duration of blood bag storage among the studied β-thalassemia children:

Item	Duration of blood bags storage (days)				chi- square test	P- value
	Less than 20 days (N=41)		More than 20 days (N=19)			
	No.	%	No.	%		
Insufficient increase of Hb level:						
Sufficient	40	97.6	1	5.3	fisher's exact	0.000* (HS)
Insufficient	1	2.4	18	94.7		
G6PD status:						
Deficient G6PD <7.19	1	2.4	11	57.9	fisher's exact	0.000* (HS)
Normal G6PD ≥7.19	40	97.6	8	42.1		

This table (Table 12) shows that among blood bags stored for less than 20 days, only 2.4 % have sufficient increase of hemoglobin, while after transfusion of blood bags stored more than 20 days, most of patients (94.7%) have insufficient increase of Hb level, with high statistical significance. As regard G6PD among blood bags stored for less than 20 days, only 2.4 % have deficient G6PD <7.19, while after transfusion of blood bags stored more than 20 days, about half of blood bags (57.9%) have deficient G6PD <7.19, with high statistical significance.

Table (12): Hemoglobin and HCT and duration of blood bag storage among the studied β-thalassemia children:

Item	Duration of blood bags storage (days)		MWT	P-value
	Less than 2days (N=41)	More than 20 days (N=19)		
Hemoglobin:				
Mean ± SD	9.37± 0.74	8.26 ± 0.99	-6.740	0.000* (HS)
Median (Range)	9.5(7.8-11)	8.1(5.9-10)		
HCT:				
Mean ± SD	31.37±3.33	26.54±3.49	-5.997	0.000* (HS)
Median (Range)	32(24-36.2)	25.2(22-34.2)		

The mean of hemoglobin level among the studied children after blood transfusion of blood bag stored for less than 20 days is statistically higher than mean of hemoglobin level among the studied children after blood transfusion of blood bag stored for more than 20 days (9.37± 0.74) Vs (8.26 ± 0.99) respectively. Also, there is insufficient increase in HB and HCT level after blood transfusion of blood bag stored for more than 20 days in the studied thalassemia children.

4. Discussion

Thalassemias are genetic disorders with paternal inheritance, which are prevalent world widely, causing 25,000 deaths in 2013 with highest rates in the Mediterranean area (Italy, Greece, Turkey, West Asia, North Africa, South Asian and Southeast Asia). The β-thalassemia major is the most severe form and the affected children are dependent on regular blood transfusions for survival **(8).**

β-thalassemia is the most common genetically inherited hemoglobin disorder in Egypt with carrier rate varying from 5.3 to more than 9%**(9).**

G6PD-deficient RBCs if exposed to oxidative stresses such as storage for a long period, it will lead to hemolysis in vivo following transfusion of this RBCs. The question here is it safe to transfuse RBCs from G6PD-deficient donors. However, information regarding possible adverse effects of transfusing G6PD-deficient RBCs remains limited. Therefore, blood donors still are not routinely screened for G6PD deficiency and no rules in blood centers regarding deferral of known G6PD-deficient donors **(10).**

In patients with chronic anaemia secondary to congenital or acquired haematologic diseases like sickle cell disease (SCD), thalassaemia, myelodysplasia or aplastic anaemia, it is acknowledged that the transfusion of "young" (fresh or cryopreserved) RBC is more profitable, mostly because the period until the next transfusion is usually more prolonged **(11).**

Most of the studied group having positive family history of β -thalassemia (90 %). Same result were assessed for study in 2016, when researcher found a positive family history (72%) on his included participants **(12).**

The current study assessed the splenomegally among the studied patient. One third of them have splenomegaly (33.3 %), and only 15% of them had splenectomy. Same result were assessed in study in 2008, when researcher found an enlargement of spleen as a complication of thalassemic patient **(13).**

The current study evaluate the blood grouping among the studied patient O+. B+ and AB+ are the most prevalent blood group (31.7%, 21.7% and 18.3%) respectively. Same result were assessed in study in 2017, when researcher found on analysis, that the most common blood group affected by B-thalassemia is O+ve **(14).**

The current study assessed the total bilirubin serum levels among the studied patients. The results ranged from (1 to 3mg/dl), indicating a mild rise in their serum bilirubin levels. Same results were revealed clearly in a study in 2015, when the researcher found an increase in serum levels of bilirubin among the whole thalassemic patients who were included in the study **(15).**

Another vital organ functions was handled in this study. Kidney function tests were assessed in thalassemic patients and results showed that, urea ranged between 20 and 44mg/dl with mean value of urea 26.65±5.58 mg /dl and creatinine ranged between 0.6 and 1.0 mg /dl with mean value0.75±0.10mg /dl, indicating normal renal functions in those patients, results that match with the study done in Turkey, in which the researcher evaluated kidney functions in thalassemic patients, found normal kidney functions, with urea ranged between 6 and 21 with mean ± SD 11.9±3.3 and creatinine ranged between 0.3 and 0.8 with mean ± SD 0.4±0.1 revealing no significant correlation between control or thalassemic patient**,** results that also matched with a recent study done by

N. azar et al., which reveled tubular and glomerular functions appear to be well preserved in all variants of β-thalassemia after assessment of renal tubular and glomerular function of 72 patients with β-thalassemia **(16)**.

Frequency of transfusion of patients included in this study was variable ranging from every two weeks to every four weeks, with the majority receiving blood transfusion every 4 weeks (55%), this result matched with a study done in 2010 which revealed the frequency of transfusion in thalassemia major is every two to four weeks **(17).**

Our result showed that, prevalence of G6PD deficiency in the blood bags was12 bags (20%) from 60 blood bags with normal remaining blood bags (80%). Previous result in line with study in India in which, the G6PD enzyme in the blood bags before transfusion were examined. Of the 114 transfusion blood bags used across patients, 100bags (87.7%) had G6PD-sufficient RBCs and 14 bags (12.3%) had G6PD-deficient red cells **(18).**

Our study also showed that, there was counter relationship between duration of blood bags storage and G6PD level in blood bags, blood bags stored for more than 20 days have deficient G6PD level, blood bags stored less than 20 days have normal G6PD level. Same results guaranteed by **Emamghorashi et al., (19)** who found that, there was statistically significant difference between storage of blood bags < 20 days and storage in blood bags>20 days regarding G6PD level**.**

The current results revealed a statistically significant difference between the stored blood bags< 20 days and stored blood bags > 20 days regarding the hemoglobin and hematocrit rise in patients after transfusion of blood bags (P. value= 0.000*), same matched data by **Emamghorashi et al., (19)** which showed higher hemoglobin levels post transfusion of blood bags stored for less than 20 days than the mild hemoglobin rise among children post transfusion of blood bags stored for more than 20 days (9.37+- 0.74) Vs (8.2+- 0.99) respectively. Also, there is a high significant decrease in HCT level after blood transfusion of blood bags stored for more than 20 days in the studied children**.** Another matched results in a study in2007 which revealed the complications associated with receiving old blood bags: insufficient rise in hemoglobin (55.9%), hemoglobinuria (35.3%) and rise in bilirubin (8.8%), which were significantly higher than the recent one **(20).**

This study showed that regarding insufficient increase in Hb (as a complication after transfusion of G6PD deficient blood), there was statistically significant difference between normal G6PD blood bags and G6PD deficient blood regarding sufficient increase in hemoglobin > 2g /dl as insufficient increase in Hb, was present in 8 cases (16.7%) in children received normal G6PD blood in comparison to 11 cases (91.7%) of children received G6PD deficient blood, P.value = 0.000*), this conclusion agrees with Iranian study by **Nabavizadeh and Anushiravani, (20)** who found that an insufficient increase in HB was present as a complication after transfusion of G6PD deficient blood in 19 (55.9%) of the studies cases.

Also our study showed that, hemoglobin level pre-next transfusions among both groups (who had G6PD normal-blood and who had G6PD deficient – blood), there was statistically significant difference regarding Hb level among both groups, the same result was revealed by **Zekavat et al., (18)** which there was statistically significant difference between both groups regarding Hb level pre-next transfusions.

Conclusion

- Prevalence of G6PD level in blood bag was found to be normal 48 bags (80%) and in deficient in the remaining 12 bags (20%).
- There was high statistically significant difference between normal G6PD in blood bags and G6PD deficient blood regarding insufficient increase of hemoglobin.
- There was high statistically significant difference between storage in blood bag <20 days and storage of blood bags ≥20days regarding to G6PD level in blood bags.
- There was high statistically significant difference between storage in blood bag <20 days and storage of blood bags ≥20days regarding to hemoglobin (P-value=0.000 and hematocrit of patient after transfusion of blood bags.

We recommend the following:

- Transfusion of fresh blood (less than 20 days storage) to any patient need repeated blood transfusion like thalassemic patients.
- There is a general consensus to apply a transfusion strategy of 'young' RBCs on thalassemic patients that require transfusion, simply because they reduce the number of RBC units needed, thus decreasing general transfusion- related risks.
- Replication the study in other areas and among large sample.

References

1. Giardina P, Forget B, Hoffman R, et al (2008): Hematology Basic Principles and Practice. 5th ed. Philadelphia, PA: Elsevier Churchill Livingstone; chapter 41.
2. DeBaun M, Frei-Jones M, Vichinsky E, et al (2011): Nelson Textbook of Pediatrics. 19th ed.

Philadelphia, PA: Saunders Elsevier; 2011: chapter 456.
3. Richard O, Jeffrey S, HuyP, et al (2013): Glucose-6-Phosphate Dehydrogenase-Deficiency in Transfusion Medicine: The Unknown Risks. Vox Sang; 105(4): 271–282.
4. Andre M, Renzaho A, Eliette H and Michael P (2014): Should Blood Donors Be Routinely Screened for Glucose-6-Phosphate Dehydrogenase Deficiency? A Systematic Review of Clinical Studies Focusing on Patients Transfused With Glucose-6-Phosphate Dehydrogenase–Deficient Red Cells. Transfusion Medicine Reviews; 28: 7–17.
5. Rother R, Bell L, Hillmen P, et al (2005): The clinical sequelae of intravascular hemolysis and extracellular plasma hemoglobin: a novel mechanism of human disease. JAMA; 293:1653–62.
6. Korgun D, Bilmen S and Yesilkaya A (2001): Alterations in the erythrocyte antioxidant system of blood stored in blood bags. Res Commun Mol Pathol Pharmacol;109: 357-363.
7. Stainsby D, Dhingra N, James V, et al (2012): Blood donor selection: guidelines on assessing donor suitability for blood donation. World Health Organization; Geneva, Switzerland.
8. Sharma D. C, Arya A, Kishor P, et al (2017): Overview on thalassemias: are view article. Medico Research Chronicles, 4 (3): 325-337.
9. AL-Beshlawy and youssry (2009): Prevention of hemoglob-inopathies in Egypt. Hemoglobin; 33(1):14-20.
10. Francis RO, Jhang J, Hendrickson JE, et al (2013): Frequency of glucose-6-phosphate dehydrogenase deficient packed red blood cell units in a metropolitan transfusion service. Transfusion; 53(3):606-611.
11. Chaebech P, de Menorval MA, Bodivit G, et al (2016): Evidence of benefits from using fresh and cryopreserved blood to transfuse patients with acute sickle cell disease. Transfusion; 56:1730-8.
12. Ashwin P Patel, Prakash H Parmar, Rupesh B Patel, et al (2016): Factors Influencing Beta-thalassemia Awareness in Western India. Ntl J Community Med; 7(3):193-19.
13. Cianciulli P (2008): Treatment of iron overload in thalassemia. Pediatric Endocrinology Reviews: PER; 6 (1):208-213.
14. Sinha PA, Mulkutkar SH, Bhavani JB, et al (2017): Original Research Article Study of distribution of ABO blood groups in ß-thalassemia patients. International Journal of Research in Medical Sciences Sinha; 5(8):3479-3483.
15. Sarai N, Rathore M, Bohra VD, et al (2015): Diagnostic significance of Liver and Renal function tests (LFT & RFT) in Iron overload in patients with β Thalassemia major. International Journal of Clinical Biochemistry and Research: 2(1):27-32 27.
16. Azar N, Leblond V, Ouzegdouh M, et al (2017): A transition from using multi-step procedures to a fully integrated system for performing extracorporeal photopheresis: A comparison of costs and efficiencies. J Clin Apher;32:474–478.
17. Galanello and Cao. A (2011): Alpha-thalassemia. Genetics in Medicine; 13(2): p 83-88.
18. Zekavat OR, Karimi M and Rahmanian F (2009): Is red cell from an otherwise healthy G6PD deficient donor efficient for transfusion to fauvism patient? Indian J Hematol Blood Transfus; 25:23-6.
19. Emamghorashi F, Hoshmand F and Moh-tashamifar A (2010): Screening foe glucose-6-phosphate dehydrogenase deficiency in blood donors. Hematology; 15: 122-124.
20. Nabavizadeh SH, Anushiravani A (2007): The prevelance of G6PD deficiency in blood transfusion recipients. Hematology;12(1):85-88.

2/26/2020