Original Article

Blood Transfusion Rate in Congolese Patients with Sickle Cell Anemia

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ABSTRACT

Objective. The main objective of this study was to evaluate the rate of blood transfusion in African Sickle Cell Patients and the risks related to the use of total blood.

Methods. 186 sickle cell patients (95 males and 91 females) aged 0-21 years were regularly followed over a 3 years period in Katanga province, DR Congo. Indications for blood transfusion were mainly based on clinical criteria and Hb level (less than 5g% ml or a drop of 2g% under the steady state value). All the subjects, who were transfused, wer screened for hepatitis B surface antigen (HBs Ag) and Human Immune deficit Virus (HIV).

Results. Of 186 patients, 150 (80.6%) were transfused, and the average blood transfusion requirement was 0.4 units per patient-year. According to the age of first transfusion, 75.3% (113/150) of them were transfused before the 6th year of life; but the frequency of transfusions seemed to decline in children aged more than 13 years. The risk of HIV infection from blood transfusion was estimated at 1 per 37.1 units or 26 per 1000 blood units. The hepatitis B surface antigen was detected in 15 cases (10%) and HIV serology was positive in 17 patients (11.3%).

Conclusion. Because of the complications related to blood transfusions in Africa, efforts are needed in order to reduce the frequency of transfusions, by preventive measures (early diagnosis, malarial and penicillin-prophylaxis) and to use more rational indications. **[Indian J Pediatr 2007; 74 (8) : 735-738]** *E-mail : leon.tshilolo@gb-solution.cd*

Key words : *Congolese; Sickle cell anemia; Blood transfusion; HIV risk.*

In Africa, blood transfusions are frequently given to treat severe pediatric anemia associated mainly with malaria. Nearly all the transfusions are given within 24 h of admission and deaths takes place mainly on the first day of admission.[1,2] In some areas, as many as 19-47% of hospitalized children received transfusions; and because of a high HIV seropositivity rates among blood donors, blood transfusion is an important mode of HIV transmission among African children.[3]

Indication of blood transfusion in sickle cell patients depends on the clinical and biological data but varies between different areas. In many countries which have adopted modern medical practices, blood transfusion in patients with sickle cell disease (SCD) depends on rational indications (septic status, cerebral vascular crisis, pregnancy, acute chest syndrome, intolerable anemia, *etc.*) and consists mainly in the use of packed red cells[4,5] while in most African countries, severe symptomatic anemia is the most important and almost the only indication and total blood is mainly given.[6,7]

Democratic Republic of Congo (DRC) is, in spite of its rich natural resources, one of the poorest country in the World where no public medical assistance is organized and therefore a comprehensive care is cost effective. In DRC, most of the sickle cell patients bear mainly the Bantu (CAR) haplotype and develop a clinically severe form of sickle cell anemia (SCA) with a high mortality and morbidity. Acute anemia is very frequent in African sickle cell patients because of malaria, infections and other environmental conditions.[8]

Chronic transfusion program neither hydroxyurea treatment is not yet available in DRC.

In this report, we present our experience of blood transfusion in Congolese Sickle Cell patients suffering from acute symptomatic anemia and try to determine the

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frequency of transfusion and to establish its pattern and the main related problems.

MATERIAL AND METHODS

We studied 186 patients (95 males and 91 females) aged 0 - 21 yr with SCA in Katanga, a southeastern province of Congo (Zaire). Diagnosis of SCD was determined by standard laboratory procedures (electrophoresis hemoglobin) and family studies. Indications for blood transfusion were mainly based on clinical criteria and Hb level (less than 5g%ml or a drop of 2g% under the steady state value) during the course of severe anemia. All the subjects, who had received at least one transfusion, were screened for hepatitis B surface antigen (HBs Ag) and Human Immune deficit Virus (HIV). Hepatitis C test was not realized.

Clinical and biological parameters were recorded regularly for a period of 3 years. The values of biological and clinical parameters were subjected to statistical analysis using computer programme (SPSS10). The range for each parameters was calculated using mean ± standard deviation (SD) and comparisons were carried out using the students t-test of X^2 test, and p value less than 0.05 was considered statistically significant.

Table 1. Age and Gender Distribution of Sickle Cell Patients.

Age	0-2	3-5	6-12	13-15	16-21	Total
M.	7	13	27	25	9	81
F.	7	8	25	13	16	69
Tot.	14	21	52	38	25	150

(F=females; M=males: n=150; age expressed in yr)
Note : Only a few number of children are aged less than 5 years because of a high mortality rate and the absence of a newborn screening program.

Table 2. Age Distribution of Patients at 1st and Last Transfusion.

Age	0-2	3-5	6 -12	13 -15	16-21	Unknown
1st Tr	94	19	9	1	0	27
Last Tr	38	25	41	9	7	20

(Tr.=Transfusions; n=150; age groups, expressed in yr)

RESULTS

Of the 186 patients with SCD, 150 (80.6%) of them were given blood transfusion (Table 1). According to the age of the first transfusion, we observed that the majority of patients (94/123) were transfused in the first two years of life. No data were available in 27 cases (Table 2). Five (3.3%) were transfused before 3 months of age, and one patient was transfused at 13 years and then received a total of 17 blood transfusions during a 2 year interval.

All of the patients (mean age 10 yr.) had received a total of 632 blood units (mean 4.2 range 1-22) and the average blood transfusion requirement was 0.4 units per patient per year (Fig. 1). When related to the age groups, the mean values of blood units were 2.2 in infants (0-2yr), 2.7 in pre-school years (3-5yr), 4.25 in school years (6-12yr), 5.12 in adolescents and young adults, respectively.

During a 3-yr follow-up period, 9 of the 150 patients were out of the study and 81(60%) were blood transfused. The free time interval between transfusions showed a mean value of 1 yr (SD: 0.9) in the 0-2 and 3-5 yr patients groups, 3.5 yr (SD: 3.2) in children aged 6-12 yr and 4 yr (SD: 4.3) in adolescents and young adults.

The frequency of transfusions and the need of blood

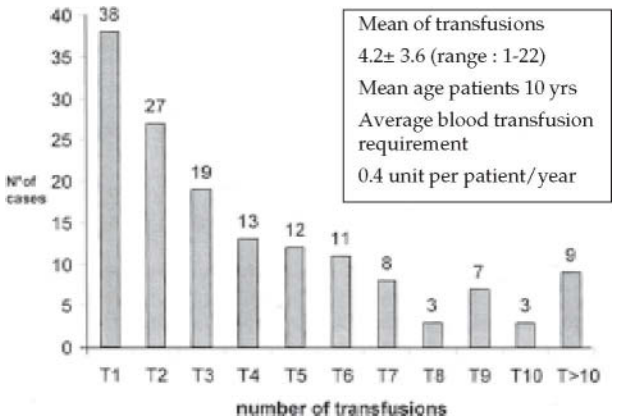

Fig. 1. Distribution of patients (n 150) according to the number of transfusions

Table 3. Sickle Cell Patients Related to the Transfusion Rate During a Three-Year Period of Follow-up (1990-93)

Age groups (yr)	0-2		3-5		6-12		13-21		Total (M)
No0 Transf.	M	F	M	F	M	F	M	F	
0	0	0	2	3	10	9	18	14	56 (30)
1	3	3	4	3	10	7	11	8	49 (28)
2	1	2	1	0	2	4	5	2	17 (9)
3	1	0	3	1	1	0	0	4	10 (5)
4	1	0	1	0	1	0	0	1	4 (2)
5	1	0	2	0	0	0	1	0	4 (4)
6 and more	1	0	0	0	0	0	0	0	1 (1)
total	8	5	13	7	24	20	35	29	141 (79)

(M: males, F : females); No transf: number of blood transfusions) 9 of the 150 patients were out of the study during this period.

Table 4. Clinical Data Related to the Number of Blood Transfusions

N°of patients											Total
	38	27	19	13	12	11	8	3	7	3	9	150 (100)
N° of transfusions	T1	T2	T3	T4	T5	T6	T7	T8	T9	T10	>T10	
Hepatomegaly	16	9	8	7	7	6	2	0	6	1	5	67 (44.6)
Splenomegaly	11	4	7	3	4	3	3	0	3	2	5	45 (30)
Epistaxis	9	6	3	3	4	3	4	1	4	1	2	40 (26.6)

No correlation was seen with the number of blood transfusions but the persistence of hepatomegaly was higher in patients who had received more than 8 blood transfusion (12/19 cases).

seemed to decline in children aged more than 13 yr. Fifteen patients (10%) needed repetitive blood transfusions over a very short period of time, not exceeding 10 days.

Table 3 displayed some peculiar clinical data in congolese sickle cell patients:

Splenomegaly and hepatomegaly were found in 45 (30 %) and 67 (44.6 %) patients, respectively. No correlation was seen with the number of blood transfusions but the persistence of hepatomegaly was higher in patients who had received more than 8 blood transfusions (12/19 cases). (Table 4). Forty patients (26.6%) had suffered at least one episode of epistaxis with a predominance of males (ratio 1.5/1). No specific cause of nose bleeding was identified.

The hepatitis B surface antigen was detected in 15 cases (10%) and HIV serology was positive in 17 patients (11.3%). Almost all the HIV positive subjects presented the HBs Ag in the serum. The prevalence of hepatitis B and HIV infection in blood donors was of 5 and 3.5% respectively. Hepatitis C test was not available.

Since all the patients had received 632 blood units and 17 of them were infected with the HIV agent, we estimated the risk of HIV infection from blood transfusion at 1 per 37.1 units or 26 per 1,000 blood units.

DISCUSSION

Sickle cell patients living in Congo are more susceptible to anemic crisis than those living in developed countries because of environmental (malnutrition, malaria, infections) and genetic factors (Bantu haplotype)[5,9]. While in developd countries, the indications for blood transfusion are variable[4] in our context, severe symptomatic anemia was the only cause. In addition, blood transfusions wre carried out very early in Congolese patients, even before diagnosis of SCD was established.[9, 10] The average blood transfusion requirement is close to those reported in other African studies[11] and the need for blood transfusion seems to reduce with age according to the free time interval between transfusions.

We are of the opinion that patients who needed repeated blood transfusions over a short time would have developed alloimmunisation or auto-antibodies in course of development of Delayed Haemolytic Transfusion Reaction (DHTR), as observed in Europe and America.[12,13] As the majority of donors were family members, we can exclude the hypothesis of racial differences between donors and recipients as an explanation for such a phenomenon. Furthermore studies are needed.

As reported in some African countries[14] many of our patients presented persistent splenomegaly past the age of 5 years (Adeodu *et al*[14]). Revealed that patients with splenomegaly suffered more from anemic crises than those without splenomegaly. Buchanan *et al.*[15] demonstrated that intensive transfusion therapy (with the haemoglobin S level maintained at less than 20%) was accompanied by increased splenic size and phagocytic function. In the present study, we did not find any correlation with either the need for blood transfusion or with the unit number of transfusion therapy. Epistaxis predominated in males but spared children under the age of 3 years. The cause of torrential rose bluding in SC patients has not yet determined even though a such phenomenon has been reported in other African patients.[10,11,16]

In spite of the lack of HIV and HBV status before transfusions, the risk of post-transfusion viral infections was high in our patients. Indeed, HIV seroprevalence in Katanga (Shaba) was high (5.4%).[17] We were not able to screen for hepatitis C virus and would hypothesize that hepatomegaly could be explained by hemochromatosis or chronic hepatitis C infection as demonstrated in a Nigerian study.[18]

CONCLUSION

Because of the complications related to total blood transfusions, efforts must be made in order to reduce the frequency of transfusions, by preventive measures (early diagnosis, malarial and penicillin prophylaxis) and the use of red blood packed units.

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