Efficacy of exchange blood transfusion in the management of priapism in patients with sickle cell disease in northeast Nigeria

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ABSTRACT

Background: Thirteen cases of priapism seen in adult patients with sickle cell disease (SCD) were treated by exchange blood transfusion (EBT) at the University of Maiduguri Teaching Hospital during a five-year period (1996-2000). Each patient was scheduled to receive a total of 12-unit exchange, but due to voluntary donor blood scarcity and differences in affordability of commercial donors only 1 (7.7%) patient could receive 10 units, 3 (23.1%) patients had 8 units each, and 9 (69.2%) patients received 7 units each.
Method: EBT was carried out manually in all cases and the mean number of units exchanged, the mean levels of Hb S and the mean levels of haematocrit were determined before the start of EBT (pre-EBT) and at the time of achieving complete detumescence.
Result: The exchange procedures were started with mean pre- EBT levels of 95% of Hb S and 0.24L/L of haematocrit; complete detumescence was achieved in all cases after an average of 6 units were exchanged by which time the mean Hb S level had fallen to 50.3% and the haematocrit had risen to a mean level of 0.28L/L.
Conclusion: EBT was successful in achieving complete detumescence and at much higher Hb S level (50.3%) than was earlier recommended. EBT is therefore effective in treating priapism in SCD and should be offered to such patients in order to avoid surgery, the risks of general anaesthesia in SCD and the possibility of postoperative impotence.

Keywords: Sickle cell disease, Priapism, Exchange blood transfusion.

Introduction

The modalities of red cell transfusion therapy have undergone considerable modification and refinement in the management of various clinical problems associated with sickle cell disease (SCD). The commonest modality of transfusion therapy most frequently employed in the clinical management of sickle cell disease is that of simple additive transfusion. [1] The main indication for this form of blood transfusion in SCD is the need to improve the oxygen-carrying capacity of the blood in conditions associated with worsening anaemia as may be encountered in hyperhaemolytic crises, acute splenic sequestration syndromes and aplastic crises. [1,2] However, additive transfusion in SCD must be carefully monitored in order to avoid the risk of jeopardizing cardiovascular function due to volume over-load, hence the need for cautious application of pre-transfusion diuretics and the preferential use of plasma depleted components such as plasma reduced blood or red cell concentrate in optimal additive solution (OAS). [3,4] Further more, it must be emphasized that simply raising the haematocrit level by additive transfusion without significantly reducing the percentage of hemoglobin S containing red cells may result in complications secondary to increased blood viscosity. [2] Transfusion must therefore be minimized and judiciously applied to restore steady state haematocrit level only, otherwise the benefit of increased oxygen-carrying capacity provided by simple additive transfusion can be offset by the adverse effect of increased viscosity. [2]

Another important but less commonly employed modality of transfusion therapy in SCD is Exchange Blood Transfusion (EBT), which is usually reserved for severe and life threatening clinical situations such as acute chest syndrome, cerebrovascular accidents and priapism. [1] The beneficial effect of EBT in SCD is due to improvement of microvascular perfusion as a result of decrease in the proportion of haemoglobin S containing red cells in the circulation. [2] The ideal types of red cell preparations preferred for EBT are the freshest possible units of whole blood or plasma reduced blood, [5,6] but not red cell concentrate in OAS the use of which may precipitate or aggravate pre-existing hypoproteinaemia because OAS is poor in protein. [3,4,7] Exchange blood transfusion can be performed manually or automatically with a cell separator, 3,4 the manual technique though laborious is the only available procedure in most developing countries including Nigeria.

Priapism in SCD is caused by sickling episodes in the channels of the corpora cavernosa leading to stasis and protracted congestion of the penile erectile tissues resulting in painful erection. [8] Priapism is injurious to the erectile tissues, and if severe erection persists for more than a day, partial or complete impotence may occur as a result of corpora cavernosal fibrosis [8] Priapism is therefore a clinical emergency and should be treated with utmost urgency. The management of priapism depends on its severity and persistence. Mild cases of priapism are usually transient and generally tend to subside spontaneously, [8] but severe persistent cases will require active management, which may include modalities such as needle aspiration with intracorporeal adrenalin injection, EBT or surgical venous shunting. [8]

In our centre, at the University of Maiduguri Teaching Hospital (UMTH),

all cases of SCD with priapism are initially managed by needle aspiration with intracorporeal adrenalin injections followed by EBT if the aspiration procedure fails and patients are offered the surgical venous shunt procedure as a last resort if all of the earlier procedures failed to achieve detumescence. In this paper we present an assessment of the clinical efficacy of EBT in the management of priapism in patients with SCD as seen in the University of Maiduguri Teaching Hospital (UMTH), Maiduguri, North East Nigeria.

Patients and Methods

Thirteen consecutively admitted cases of priapism in patients with SCD, seen during a five-year period (1996-2000) were included in this report. All of the patients were adults (aged 19-28 years) with sickle cell anaemia (Hb SS) who were diagnosed and registered with the Haematology Clinic of the UMTH.

The patients were scheduled for EBT after needle aspiration with intracorporeal adrenalin injections had failed to achieve detumescence in each case. The initial plan in each case was to deliver a standard protocol of EBT for adult patients with SCD (4,6) in which the procedure is carried out iso-volaemically in four cycles in each of which three units are venesected and three units are transfused using two separate venous accesses to permit simultaneity of the procedures. The overall exchange protocol procedure for the four cycles would require 12 units of blood for each patient at the end of which the patient's Hb S level will be expected to drop to less than 20%. [4,6] However, because of the peculiarity of our environment with regards to voluntary donor blood scarcity and predominance of commercial donations, the actual number of units exchanged in each patient was determined by donor availability and patient affordability. In all cases the EBT was carried out wholly manually using the freshest possible whole blood units. The average length of time between the attack of priapism and the start of EBT was 11 hours.

In each case the levels of haematocrit were determined by manual micro centrifugation method as described by Dacie and Lewis [9] and percentages of Hb S were determined by automated Hb-electrophoresis and scan quantitation using 24-VISU Densitometer (Helena, France). The average number of units exchanged as well as the average values of haematocrit and Hb S were determined and recorded before the start of EBT and at the time of attaining complete detumescence among the 13 patients.

Result

A total of 13 SCD patients with priapism were treated with EBT. Only 1 (7.7%) patient received 10-unit exchange, 3 (23.1%) patients received 8-unit exchange each and 9 (69.2%) patients received 7-unit exchange each. Among the 13 treated patients the mean haematocrit level before initiating exchange transfusion (Pre-EBT) was 0.24L/L and the mean Hb S level was 95% as shown on Table 2. All of the 13 patients responded favourably and complete detumescence was attained after an average of only 6-unit exchange leading to a drop in the mean Hb S level to 50.3% and a rise in the mean haematocrit level to 0.28L/L (Table 2).

The average length of time from commencement of EBT to attainment of complete detumescence was 5 hours. All patients are registered and regularly seen in our clinic where they are followed up, in relation to erectile function, for a period of about 1 year (for those treated in 2000) to 4 years (for those treated in 1996) and all are found to have normal erectile functions.

Table 1: Changes in mean values of haematocrit and HbS before EBT and at detumescence in relation to the mean No. of units exchanged

	Pre-EBT	Detume-scence
No. of units	0	6
Haematocrit (l/l)	0.24	0.28
% HbS	95	50.3

Discussion

The treatment of life threatening and severe conditions, including priapism, in adult patients with SCD requires an effective EBT with standard protocols [4,6] aimed at significant reduction in the level of Hb S to less than 20%, which will require removal and transfusion of at least 12 units of blood corresponding to about 1.25 times the patient's blood volume. [4,6] Lowering the Hb S level to less than 20% of the total haemoglobin in the blood would bring about sufficient reduction in blood viscosity and result in decreased intravascular sickling with a concomitant improvement in clinical status. [10] These protocols are very effective and convenient ways of delivering efficacious transfusion therapy for SCD patients in Europe and America because of the existence of established National Transfusion Services in those countries as a result of which voluntary donor blood is always readily and abundantly available. However, the situation in Nigeria is hardly ever that easy because of the lack of an established National Transfusion Service leading to severe scarcity of voluntary donor blood and predominance of costly commercial donors on the donor panels. [11 - 13] It is therefore not surprising that none of our patients in this report could afford up to 12 units of blood, infact only 1 (7.7%) patient could afford 10 units, while 3 (23.1%) patients could only afford 8 units of blood each and the majority of our subjects consisting of 9 (69.2%) patients could barely afford up to 7 units of blood each. The pattern revealed that fewer patients were able to afford larger number of units of blood required for the EBT, further more, no patient could afford the standard EBT protocol requirement of 12 units of blood. This is a reflection of the compromised socio-economic status of our SCD patients, the majority of who are generally dependent on parents and relatives for their expenses. SCD, like other chronic diseases, creates a significant economic burden on family members who may become exhausted and unwilling to give adequate financial assistance, this being more likely under the current high cost of living in the country. Further more, in our experience in this environment, family members of SCD patients are becoming increasingly unwilling to donate blood for their patients as a result of increasing awareness about HIV and the fear of being tested for the infection and the stigmatization that may follow

should the test result turn out to be positive. [11] These factors make it increasingly difficult to offer an effective EBT therapy in our environment. It must be emphasized that despite these difficulties only blood that has been tested and screened negative for hepatitis B surface antigen and HIV antibodies is acceptable for transfusion. [5]

Despite our limitations with regards blood availability, we have been able to achieve success in all of our 13 patients in whom we were able to attain complete detumescence after an average of 6-unit exchange, which brought the mean Hb S level down from a mean pre-EBT value of 95% to 50.3% as seen on table 1. This result is at variance with reports from Europe and America suggesting that the clinical efficacy of EBT in inhibiting and reversing intravascular sickling would significantly occur when Hb S levels have been reduced to 20-30%. [2, 4, 6, 14] Therefore, our results have revealed greater efficacy of EBT in the treatment of priapism in SCD than would be expected based on earlier reports; this would suggest that significant reduction in blood viscosity could occur at relatively higher levels of Hb S in SCD patients living in tropical countries such as Nigeria as compared with such patients who are resident in temperate countries of Europe and America. The reason for this disparity may be related to differences in climatic conditions in which the relatively higher tropical temperatures may favourably effect the rheological properties of circulating blood leading to lower viscosity than would be expected under conditions of low temperatures of the temperate countries. However, a well-organized comparative rheological studies between SCD patients resident in the tropics and their counterparts living in the temperate regions of the world will be needed to clarify these observations.

It can also be seen from table 1 that the EBT procedure has resulted in a rise in haematocrit value from a mean pre-EBT level of 0.24L/L to 0.28L/L after 6-unit exchange at the instance of detumescence. This is thought to be due to the fact that the blood removed from the patient with SCD always had a lower haematocrit than the donor blood being transfused. Hence, the EBT procedure leads to progressive net gain in haematocrit after each cycle of exchange. [4] The overall effect of the EBT procedure is that of a beneficial rise in haematocrit leading to improved oxygen delivery accompanied by improved tissue perfusion as a result of falling levels of Hb S and decrease in blood viscosity. [2,10] These changes eventually lead to inhibition and reversal of intravascular sickling processes resulting in clinical improvement. [2,10]

Conclusion

This study would suggest that EBT in SCD patients with priapism is efficacious in achieving detumescence at much higher haematocrit levels than was expected. Clinicians should therefore, despite voluntary donor blood scarcity in our environment, encourage affected patients and their relatives to bear the burden of safe blood procurement for EBT in order to avoid surgical intervention, which entails risks including that of general anesthesia in SCD as well as that of possible post operative impotence which can be psychologically devastating for the patients and their spouses. The urgent need for an effective National Transfusion Service to ameli-

orate the current nation wide voluntary donor blood scarcity cannot be over emphasised.

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