Erythroexchange in sickle cell disease. A three-step procedure to remove more haemoglobin S

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Dear Sir,

Transfusion therapy is a cornerstone in the treatment of sickle cell anaemia as it improves tissue oxygenation (by increasing the absolute value of haemoglobin and reducing the proportion of haemoglobin S) as well as reducing blood viscosity; chronic transfusion therapy also leads to the suppression of the propensity of endogenous red blood cells to sickle.

The transfusion therapy is based mainly on red blood cell transfusions while experience with the use of erythrocyte exchange (EEX) in this disease is limited.

EEX, practised both as acute therapy and as periodic and scheduled treatment, is an "exchange of red blood cells" designed to remove large volumes of sickle cells quickly and replace them with normal red blood cells, without increasing blood viscosity or causing iron overload. The aim of EEX is to reduce and maintain the haemoglobin S <30%, while bringing the values of haemoglobin and haematocrit to approximately 10 g/dL (haematocrit <30%). Acute EEX is considered the treatment of choice in cases in which the levels of haemoglobin are ≥10 g/dL, especially in the treatment of stroke.

On the other hand, the purpose of periodic, programmed EEX is to maintain the values of haemoglobin S stably below 30-35%, avoiding iron overload, reducing accesses to hospital and thereby improving the quality of life of patients.

At the Blood Transfusion Service of St. Anna Hospital in Ferrara, we use EEX in the chronic treatment of patients with homozygous sickle cell disease. So far we have followed ten patients (4 males and 6 females), four of whom are children. Most of the patients are African or Albanian.

In the period between 2003 to 2013 we performed over 350 EEX procedures, which were carried out in 60-90 minutes with a continuous flow cell separator (Fresenius Kabi AG, Bad Homburg, Germany - COM.TEC®). The red blood cells used for the exchange were leucodepleted, extensively typed (for Rh, Kell, Jk, Fy) and resuspended in SAG-M preservative solution (maximum 5-7 days of storage) with a haematocrit of about 60%. The erythrocyte volume exchanged in each procedure was calculated using a modified method studied at our Transfusion Service to obtain greater removal of haemoglobin S. This method involves three steps carried out with the plasma exchange device of the cell separator.

Step 1 - Consisted of connecting, on the cell separator, the reinfusion line of red blood cells with the bag of the waste and removing a volume of patient's red blood cells calculated to reduce the haematocrit by 10-15%, using saline or human albumin solutions at 4% as replacement fluid.

Step 2 - Was isovolaemic exchange of patient's red blood cells with an equal volume of homologous erythrocytes (20 mL of red blood cells/kg weight).

Step 3 - Was to restore the connection of the reinfusion line of red blood cells with the patient and restore the haematocrit to the desired value by exchanging a volume of the patient's plasma with the appropriate volume of homologous red blood cells.

Compared to standard red cell exchange, the use of this procedure allows a greater percentage reduction of haemoglobin S (60% instead of 50%) using a smaller volume of homologous red blood cells (20 mL red blood cells/kg weight instead of 30 mL/kg weight).

Pre-exchange haemoglobin S levels (evaluated after 55-60 days) were always under 50-52%. The procedures were well tolerated and there were no adverse reactions; isovolaemic EEX also safeguarded our patients from the iron overload that can follow transfusion therapy.

Only one patient had an episode of erythrocyte alloimmunisation, with anti-M antibody specificity. From a clinical point of view, all the patients treated with this novel EEX are in durable symptomatic remission and none has dangerous complications of their disease. EEX is still a little-known procedure used in the acute or long-term treatment of sickle cell disease.

Our experience leads us to believe, partly in view of the increasing number of citizens of different ethnic groups (including those with high incidences of sickle cell disease) that live in our country, that EEX should be taken into account for the management of these patients, because of its high tolerability and effective reduction and control of haemoglobin S levels (with excellent control of symptoms and complications) without iron overload. Furthermore, the increased removal of
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haemoglobin S with the three-step procedure not only saves homologous red blood cells, but could also provide longer periods of therapeutic efficacy thereby reducing accesses to hospital and improving the patients' quality of life.

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References

