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# Red blood cell exchange in sickle cell disease patient with multiple alloantibodies

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## Abstract:

There are several reports in medical literature about Red Cell Exchange (RCE) being routinely performed pre-operatively in sickle cell disease patients to provide immediate decrease in HbS concentration and prevent post-operative complications. We would like to present one such case of SCD who also had multiple allo-antibodies and had to undergo hemi-arthroplasty for avascular necrosis of head femur. Grouping and antibody screening was performed using column agglutination technique. 3-cell and 11-cell panel were used for antibody screening and identification, respectively. Automated RBC exchange was performed on apheresis machine Com.Tec using the standard PL1 kit (Fresenius Kabi, Germany). Multiple (anti-c, E) allo-antibodies were identified and successful pre-operative RCE was done with corresponding antigen-negative AHG compatible RBC units. Single RCE procedure reduced HbS concentration from 65% to 25%. The patient underwent uneventful hemi-arthroplasty and was discharged on post-operative day-7. Patient is on regular follow-up and continues to do well two months after the day of surgery. This is possibly the first case report from India, which illustrates successful automated RCE in a SCD patient with alloimmunization.

## Keywords:

Alloimmunization, red blood cell exchange, sickle cell disease, sickle hemoglobin

## Introduction

Alloimmunization is one of the most common complications of multiple transfusions in sickle cell disease (SCD) patients, and its incidence varies from 2% to 47% in various studies.<sup>[1-3]</sup> Other chronic complications in SCD patients are iron overload, vaso-occlusive events, recurrent pain crisis, and avascular necrosis of bone. In several of these complications and crisis, red blood cell (RBC) exchange is performed to provide immediate relief by rapid decrease in sickle hemoglobin (HbS) concentration and blood viscosity of patient. RBC exchange is also done before major

surgeries like joint replacement and has shown to result in fewer postoperative complications like acute chest syndrome.<sup>[4,5]</sup>

However, there seems to be a paucity of studies on preoperative red cell exchange (RCE) in SCD patient with alloimmunization. We would like to report a successful preoperative RCE in an alloimmunized SCD patient undergoing hemiarthroplasty.

## Case Report

A known homozygous SCD patient, 18-year-old male, presented with pain in the right hip and difficulty in walking as the principal complaint. After a thorough review, the patient was diagnosed with avascular necrosis of head femur and advised right hemiarthroplasty. Since

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the patient was known sickle cell homozygous, a hematological consultation including blood group and antibody screen was ordered.

### Blood grouping and antibody screen

This was performed on an automated platform AutoVue (Ortho-Clinical Diagnosis [OCD], USA) using column agglutination technique. Three-cell panel (Surgiscreen, OCD, USA) was used for antibody screen. Patient's blood group was found to be A RhD positive with positive antibody screen. Eleven-cell panel (Resolve Panel A, OCD, USA) was used to identify the specificity of the antibody. Initial results revealed anti-c alloantibody. However, additional 11-cell panel (Resolve Panel B, OCD, USA) also identified anti-E alloantibody. Presence of both these alloantibodies was further confirmed by absence of corresponding c and E antigen. Ten c and E antigen negative, anti-human globulin crossmatch compatible RBC units were identified from the inventory for possible RBC exchange.

### Red blood cell units

These RBC units were prepared from 450 ml whole blood collected in triple blood bag system 3F 63 ml CPD/100 ml SAG-M-PDS-V (Fresenius Kabi, Germany). All units were leucodepleted (BioR max, Fresenius Kabi, Germany) sickle negative and were <7 days from the date of donation.

### Automated red blood cell exchange

Very high HbS concentration (66%) warranted an RBC exchange before the patient could be taken up for surgery. Automated RBC exchange was performed through double-lumen 16F catheter on apheresis machine Com. Tec (Fresenius Kabi, Germany) using the standard PL1 kit (Fresenius Kabi, Germany).

The machine has in-built software program (Version-04.03.08, Com.Tec) for performing RBC exchange. As part of preprocedure requirements, demographic details of the patient along with

hematologic parameters including hematocrit (HCT) 35% and HbS concentration (66%) were entered in the software. The American Society for Apheresis (ASFA) guidelines<sup>[6]</sup> on apheresis state that RBC volume to be exchanged depends on target HbS level. With 100% RBC replacement and on the basis of target HbS level (<30%), the required RBC volume to be exchanged as calculated by the software was 2200 ml. Seven out of 10 RBC bags (total volume 2270 ml) identified were used for RBC exchange. Volume and HCT of each RBC bag was entered in the "RBC calculator" of software for RBC exchange. The software predicted postprocedure HCT as 35% and HbS as 28%.

Patient's vitals including pulse rate, blood pressure, oxygen saturation, and respiratory rate were monitored throughout the procedure. Continuous intravenous calcium gluconate 10% (30 ml in 120 ml normal saline) infusion at the rate of 60 ml/h was given to the patient during the procedure to prevent citrate effect. The procedure lasted 95 min and was completely uneventful. Table 1 compares various hematological parameters of the patient before and after the RBC exchange.

Patient follow-up: Patient underwent planned hemiarthroplasty of the right hip joint on the day following RBC exchange. Surgery was uneventful and patient was transfused 1 unit RBC (8<sup>th</sup> unit) during surgery and 1 unit (9<sup>th</sup> unit) later in postoperative Intensive Care Unit as replacement for blood loss. The clinical course of the patient was uneventful, and the patient was discharged on postoperative day 7. Currently, the patient is doing fine and is able to walk with own support after 8 weeks of follow-up.

## Discussion

This is possibly the first case report from India, of successful RBC exchange in an alloimmunised SCD patient. This was possible because authors' center had both the capability to detect and identify alloantibody(s)

**Table 1: Comparing various hematological parameters of the patient before and after the red blood cell exchange**

Hematologic parameters	Preprocedure	Postprocedure (predicted by software)	Postprocedure (actual)
Hb	12.5 gm/dl	Not predicted	12.5 gm/dl
Hematocrit	34.5%	34.5%	35%
MCV	101.8 fl	Not predicted	93.8 fl
MCH	36.9 pg	Not predicted	30.9 pg
MCHC	36.2%	Not predicted	33.1%
HbS	65.4%	28%	25.6%
RBC count	3.39 million/mm <sup>3</sup>	Not predicted	4.02 million/mm <sup>3</sup>
HbF	26.1%	Not predicted	10.3%
HbA	3.8%	Not predicted	51.7%
Absolute HbS	100%	42%	46.4%

RBC=Red blood cell, MCV=Mean cell volume, MCH=Mean cell hemoglobin, MCHC=Mean corpuscular hemoglobin concentration, Hb=Hemoglobin

and perform therapeutic RBC exchange. British Society for Hematology guidelines also underline this fact that a large referral center managing patients with SCD should have facilities and trained staff for automated exchange transfusion and should be able to provide red cells that are negative for corresponding antibodies if any clinically significant antibody is present.<sup>[7]</sup> Successful automated RCE resulted in smooth conduct of hemiarthroplasty and uneventful recovery of the patient. Although ASFA categorizes RCE, done as preoperative procedure for nonacute SCD, as category III; the evidence is Grade 2A (weak recommendation and high-quality evidence). The present case report also corroborates the fact that RCE works effectively in SCD.

Detection and identification of alloantibody in the recipient are of utmost importance. It is necessary to transfuse corresponding antigen negative blood as transfusion of antigen positive blood may cause a hemolytic transfusion reaction. Authors could arrange 10 RBC units that were corresponding antigens 'c' and 'E' negative for exchange and any intra- or post-operative requirement as their center was well equipped with such facilities. Specific reagent red cells, also known as select cells, play an important role in the confirmation of identified alloantibody and identification or elimination other clinically significant alloantibodies.<sup>[8]</sup> In the present report also, there was a common combination of two antibodies and it was with the help of select cells that underlying additional alloantibody (anti-E) was identified; only when select cells (E+ c- and E- c+) were used, and anti-E and anti-c were identified separately.

Role of automated RCE in preventing acute complications of SCD including recurrent stroke and resolution of priapism is well documented in the literature.<sup>[9]</sup> In SCD patient with very high HbS concentration awaiting surgery, performing RCE immediately achieves the target of high concentration of normal adult hemoglobin and thus providing adequate oxygen carrying capacity. This also helps in normal postoperative recovery of the patient as in the present case report.

Few observational studies suggest that RCE therapy is superior to simple transfusion in suspected stroke,<sup>[9]</sup> respiratory failure, and multiorgan failure.<sup>[10]</sup> However, multicenter cooperative studies performed by National Preoperative Transfusion in SCD Study Group have demonstrated that a simple transfusion approach in comparison to aggressive RCE transfusion approach provides equivalent outcomes, similar rates of major complications and fewer transfusion-related complications.<sup>[11-13]</sup> However, no randomized trials

analyzing the benefit of simple versus exchange transfusion for treating specific complications in SCD have been performed.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### Conclusion

This case illustrates successful automated RCE in a SCD patient with alloimmunization.

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### Conflicts of interest

There are no conflicts of interest.

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