STUDY OF NINE CASES OF AUTOMATED RED CELL EXCHANGE IN TERTIARY CARE CENTRE

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INTRODUCTION

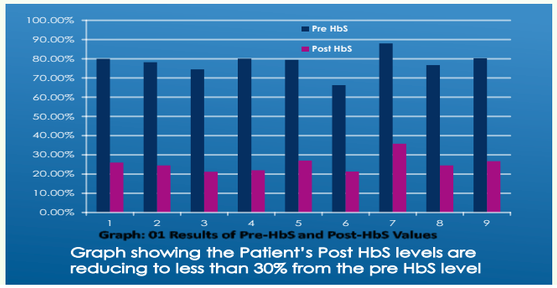
- Sickle Cell Disease is an inherited autosomal recessive blood disorder. It is a chronic disorder having qualitative defect in globin chain caused by a single mutation & substitution of valine with glutamic acid at sixth position in beta globin gene resulting in abnormal hemoglobin HbS.[1]
- Red cell transfusion increase the tissue oxygen delivery & decrease the proportion of sickle RBC in the circulation.so it is a key intervention in both the chronic as well as acute complication of sickle cell disease. Automated RCE is increasingly considered for other indications, like history of acute chest syndrome(ACS)& recurrent painful crises(RPC)[3]
- In Automated RCE, It is performed through an apheresis system. In which The removal of abnormal Red blood cells from the patient's blood & replacing them by normal donors red blood cells.
- This process requires specialist staff & equipment but can rapidly reduce the HbS level, avoiding iron loading , hence it is the recommended mode of delivering chronic transfusions for patient's with SCD.[3]

MATERIALS AND METHODS

- This Retrospective Study was conducted at tertiary care center of Baroda medical college & S.S.G Hospital , Gujarat in between 2021 to 2023.The Study analyzed the clinical outcome ,effectiveness & side effects of this procedure on 09 patients during the study period .

- **Automated red blood cell exchange:**
- Automated RCE was performed through double-lumen 16F catheter on apheresis machine Com.Tec (Fresenius Kabi, Germany) using the standard PL1kit(Fresenius Kabi) to perform all the 09 RCE procedures. The main goal of this procedure was to reducing hemoglobin S(HbS) levels by automated RCE in patients with sickle cell disease or its Variants is to 30%/under.
  So we readjust the less number of units Exchanged manually & decreases the likelihood of infectious disease & the risk of allo - immunization of blood cell antigens from exposure to more donor blood products.
- The machine has in-built software program (Version -04.03.08,Com.Tec)for performing RBC exchange. As part of pre-procedure requirements , demographic details of the patients along with hematologic parameters including hematocrit(HCT) and HbS concentration were entered in the software. The American Society for Apheresis (ASFA) guidelines 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 according to the patients body volume .
- The Blood Volume and HCT of each RBC bags were entered in the 'RBC calculator' of software for RBC exchange . The software predicted post procedure HCT as >30% and HbS as <30%. With this procedures, the patients vitals including pulse rate, blood pressure, oxygen saturation, and respiratory rate were monitored throughout the procedure.
- The packed red cell suspensions with additive solution(SAGM)using during aRCE were leukoreduced by pre storage leuko filtration , that were less than 7 days from the date of collection bags. This all units transfused were all negative for sickle cell using sickle solubility & HPLC and fully cross-match compatible units.
- The number & Volume of red cell units transfused was calculated based on the patients blood volume ,hemoglobin percentage, sickling cell percentage, target hemoglobin percentage. During the study period, the mean number of red cell units used per procedure was 4 to 6 units.
- The infusion rate of the citrate anti-coagulant is set at 0.6 ml/min when patient's first the program & then, if tolerated , increased to 0.8 ml/min and continuous intravenous calcium gluconate 10%(30 ml in 120 ml normal saline)infusion at the rate of 60ml/h was given to the patients during the procedure to prevent citrate effect. The procedure lasted 95 min and was completely uneventful.
- Table:1 Explain all details of automated RCE procedures of 09 patients

Sr.No	Age-Sex/ Weight	Total Exchange of Volume (No of Bags) & DAT/IAT	Past Medical History	Presenting Symptoms	Clinical Course With Outcome
01.	20 yr,Female/43 kg	1500ml(05) Negative	Avascular necrosis, Recurrent VOC	Hip pain(VOC), Respi symptoms	Severe Favorable
02.	21 yr Male/50 kg	1400ml(04) Negative	Retro Sternal Chest Pain,Cardiac remodeling	VOC low oxygen saturation,B /L LL palsy,V1-V3(Sinus Tachycardia),Generalised body ache,Headache	Severe Favorable
03.	18 yr Male/35 kg	1600ml(04) Negative	Splenomegaly, Mild VOC	Generalise body ache, Joint pain, Mild pallor, Severe VOC	Severe Favorable
04.	22 yr Male/42 kg	1750ml(04) Negative	K/C/O SCD Since 17 yrs,Recurrent VOC	Lower Back Ache,Chest Pain, Right Shoulder pain	Severe Favorable
05.	22 yr Male/42 kg	1000ml(03) Negative	Hip pain,Respiratory symptoms	VOC,Fever,Lower back pain,Hip joint pain	Moderate Favorable
06.	46 yr Male/59 kg	1300ml(07) Negative	Breathing Difficulty,Minor VOC	Hemolytic crisis, Body-ache	Mild Favorable with discharge against medical advice
07.	32 yr Male/60 kg	868ml(05) Negative	Cardiomegaly,Acute chest syndrome	Generalise body ache, Yellowish discoloration on skin,VOC	Mild Favorable
08.	35 yr Male/42 kg	1400ml(05) Negative	Asthma,Respiratory &GI Symp.	Acute chest pain, Breathlessness	Severe Favorable
09.	25 yr Male.55 kg	1235ml(05) Negative	Acute chest syndrome,B/L Hip joint pain	Abdominal pain, Back pain,VOC	Severe Favorable



Graph: 01 Results of Pre-HbS and Post-HbS Values

Graph showing the Patient's Post HbS levels are reducing to less than 30% from the pre HbS level

RESULT

Nine patient's who underwent the RCE had the procedure done for sickle cell disease , among which 01 was female and 08 were males . All Nine patients were complains related to sickle cell disease as described in Table:1.According to the Age group of patients were between 18 to 45 years with a mean age of 26.77 years.

- **Haematological Parameters:**

The average initial HbS levels were between 73-85% and post RCE it was brought down to 22-35%.HbA levels were maintained between 60% to 75% with target hematocrit of approximately 34%.In all cases confirmation HbA% was done through HPLC. Target Hb% was achieved in a single sitting in all the cases. single sitting in all the cases.

- **Allo-immunization:** In a-RCE,none of these patients developed any new alloantibodies.

- **Recurrent Painful Crises:**
- All Responding patients showed earliest sign of a gradual and progressive reduction in the total number of days they had to attend hospital for pain management over the years, indicating that patients experience a faster and more significant improvement in

DISCUSSION

- RCE is an effective treatment modality that can be used safely in a large number of conditions . Though the efficacy is well defined in many conditions in clinical trials.
- The major advantage of RCE in SCD is its iron neutrality. Several studies have demonstrated stable iron levels in RCE. Treatment of patients of SCD with bone marrow necrosis and multi-organ failure is mainly supportive. RCE is effective in management of both acute and chronic complications of sickle cell disease.
- However , American society for apheresis advises RCE to be used as a first line Therapy. None of these patients showed the presence of any allo or auto antibodies during antibody type and screen except one sickle cell patient who had anti-D allo-antibody. Studies suggest that SCD patients may benefit from extended antigen matching,especially in patients who have already made allo-antibodies or patients with warm reacting auto-antibodies.
- Most of the RCE in the present study were uneventful except for few allergic and febrile reactions . None of the patients had any reactions due to citrate toxicity, as the anti-coagulant used during these procedures gets metabolized in the liver within minutes and the ratio of anti-coagulant used is generally 1:12,which is very low .

CONCLUSION

- RCE is safe and clinically effective procedure with minimal or no side effects when initiated at the right time by trained personnel's . In our experience , a-RCE is a safe intervention with very low alloimmunization rates and no risk of iron overloading ,also reducing the number of episodes of emergency hospital attendance. We have seen some very good responses from patients with recurrent painful crises, but it is clear that these responses build over time.
- Multicentre coordinated studies investigating improving the delivery of a-RCE , choosing hematological targets, and assessing clinical responses represent the desired next steps for the near future.

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