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Case Report

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Therapeutic Red cell exchange in Sickle Cell Disease: A series of four cases from a single centre in a developing country

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Abstract

Sickle Cell disease is associated with sickled red blood cells which could induce vascular changes leading to vaso-occlusion. Patients experience repeated painful crises, chest infections ,stroke or priapism causing great morbidity and organ dysfunction .Red cell exchange is one modality of treatment which can be used to alleviate the symptoms .We describe a four case series, all of them known cases of sickle cell anaemia presenting to the ER with symptoms of crises .Automated Red cell exchange was performed as an emergency measure to rapidly reduce the load of HbS and alleviate the symptoms in all four patients.

Keywords: sickle cell crisis ,Therapeutic Red cell exchange, cell separator

Introduction

Red Cell Exchange (RCE) is one of the treatment options for acute sickle cell crisis recommended by the American Society for Apheresis (Padmanabhan A, et al 2019). The common goal of red cell exchange as a treatment modality would be reduce the Haemoglobin S (HbS) levels to less than 30% and a final haematocrit not exceeding 30% (Robertson D. Davenport 2014).

Approved indications for RCE are severe acute chest crisis, Acute stroke, acute hepatic sequestration, severe intrahepatic cholestasis, recurrent severe painful crisis, and conditions requiring reduction of HbS prior to major surgeries (Padmanabhan A, et al 2019). Red cell exchange can be done by an automated method using a cell separator or by a manually performed

procedure, the former is considered faster and more efficient than the latter (Anubhav Gupta et al 2020). The cell separator, when used, calculates the volume of red cells to be exchanged based on a number of parameters (Staley E 2019). We report four cases of sickle cell crisis, who reported to the Emergency and were successfully managed with an automated RCE.

Case series report

Four cases, previously diagnosed as sickle cell anemia homozygous for HBSS were on hydroxyurea, presented to the ER of the hospital. Two patients presented with fever and breathlessness. They were noted to have crepitations and rhonchi with a patch of pneumonitic changes on CT chest in the upper lobe of the lungs. These two patients settled immediately clinically after RCE and Intra venous antibiotics and also showed radiological clearance of chest.

Another patient presented with complaints of severe headache with weakness in the left upper limb who settled immediately after a RCE. One patient presented with severe conjugated hyperbilirubinemia with a bilirubin 18 mg/dl and pain in right hypochondrium with no obstructive feature on CT abdomen ,considered as sickle related intrahepatic cholestasis and recovered completely post RCE. Red cell exchange was performed using the Spectra Optia Apheresis system (Terumo BCT Lakewood, Colorado, USA). Based on the body weight, height, gender, age, initial and final hematocrit as well as average replacement fluid hematocrit and the fluid balance of the patient, the cell separator calculates the volume to be exchanged. A central venous catheter placed in the internal jugular vein was used as the venous access for the RCE. We needed to reserve two to seven cross match compatible red cell units pre procedure which again depended on the weight of the patient and the volume to be exchanged. The Acid Citrate Dextrose (ACD) to Blood ratio was set at 1: 14. The exchanged fluid used was compatible leukodepleted packed Red Blood Cells with an average haematocrit of 57%.The end point for HbS reduction was set at 30% on the cell separator .No adverse effects were seen during the procedure.

The clinical and laboratory parameters of the four patients is depicted in Table I

Table I Clinical and laboratory parameters of patients who underwent automated Red Cell Exchange

S. No	Age in years	Gender	Indication	Pre hb gm/dl	Pre HCT%	Pre reticulocyte Count %	Pre HbS %	Post hb gm/dl	Post hct %	Post Retic count %	Post HbS%
1	21	m	Intrahepatic cholestasis	9.8	29	3	80	10.7	30	1.5	24
2	13	m	Impending stroke	8.1	24	2	79	10.8	31	1	28
3	11	f	Acute chest syndrome	10.4	31	1.5	78	10.6	32	1	20
4	2	m	Acute chest syndrome	11	33	2	78	11.1	33	1	28

The details of apheresis is shown in Table II

Table II: parameters of apheresis

S. Bo	Weight In kg	Duration in minutes	Processed blood Vol (ml)	Exchanged Vol (ml)	Transfused Vol (ml)	Hct of Donor blood %	% reduction
1	48	129	4216	2120	1935	55	70
2	30	111	2905	1574	1338	58	64.5
3	32	90	2549	1394	1113	60	74.3
4	11	117	1322	689	551	55	64

Discussion

Red cell exchange has been used in sickle cell disease commonly for severe acute chest crisis, acute stroke and for recurrent painful crisis with prolonged hospitalizations (Georg Stussi et al 2019) Automated Red cell exchange carries many advantages such as euvolemic procedure and quicker and effective reduction in the sickle cell burden compared to manual procedure. Since the target HbS and haematocrit are controlled there is no risk of hyper viscosity with blood transfusions. The extra corporeal volume during the procedure is also calculated by the cell separator which is 185 ml (Spectra Optia). In centres with apheresis units for stem cell collections these procedures can be adopted easily with automated software and self-taught programme and personnel training. This is a life saving procedure in acute settings especially in developing or underdeveloped countries with huge sickle cell disease burden with poor disease phenotype. We believe the benefit overweighs the disadvantages if used for appropriate indications .The reduction of HbS is also very fast and all our patients had an average reduction of 68.2% in approximately 112 minutes for one red cell exchange procedure. The average haematocrit of donor blood that was used for the RCE was 57%.None of our patients had any adverse effects during the procedure either due to the procedure or due to the blood transfusions. Our four patients had not developed red cell antibodies and compatible allogenic blood with ABO, Kell and Rh phenotype match and leukodepleted blood was used during the red cell exchange as advised by the British Committee of Standards in Haematology (BSCH) (Bernard A. Davis et al 2019)

Conclusion

Therapeutic Red Cell Exchange is a safe procedure and can be used in patients with sickle cell crisis and offers great benefits to patients by alleviating their symptoms coupled with the laboratory evidence of a faster reduction of HbS than manual exchange and is a safe option even in developing countries with skilled personnel.

References

1. Padmanabhan A, Connelly-Smith L, Aqui N, Balogun RA, Klingel R, Meyer E. Guidelines on the Use of Therapeutic Apheresis in Clinical Practice - Evidence-Based Approach from the Writing Committee of the American Society for Apheresis: The Eighth Special Issue.2019. J ClinApher. Jun; 34(3): 171–354
2. Davenport RD, Therapeutic Apheresis. 2014 .Chapter 26 Technical Manual 18th edition Bethesda, Maryland 20814-2749.645-662
3. Gupta A, Chaudhary K, Kaushik R Role of Emergency Automated Red Cell Exchange in Sickle Cell Crisis: A Case Report 2020.Clin Med Insights Case Rep. 3; doi:.1177/ 1179547620970200. eCollection 2020
4. Staley E, Hoang ST, Liu H, Pham HP. A brief review of common mathematical calculations in therapeutic apheresis.2019. J Clin Apher. Oct; 34(5): 607–12.

5. Stussi G Buser A, Holbro A. Red Blood Cells: Exchange, Transfuse or deplete .2019 Review Article: Transfus Med Hemother 2019;46:407–416
6. Spectra Optia.2015. Terumo BCT Apheresis System Operator's manual.
7. Bernard A. Davis, Allard S, Qureshi A,. Porter JB, Pancham S, Win N, Cho G, Ryanon K behalf of the British Committee for Standards in Haematology .Guidelines on red cell transfusion in sickle cell disease. Part I: principles and laboratory aspects.2017.British Journal of Haematology, 179–191

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