49th Annual Conference of ISBTI

TRANSCON 2024

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21st - 23rd November, 2024

Ahmedabad

TRANSFUSION MANAGEMENT IN A TODDLER WITH SICKLE CELL ANEMIA FOR A MAJOR NEUROSURGERY PROCEDURE: A CASE REPORT

Event by: ISBTI (GUJARAT CHAPTER)

e POSTER ID- eP052

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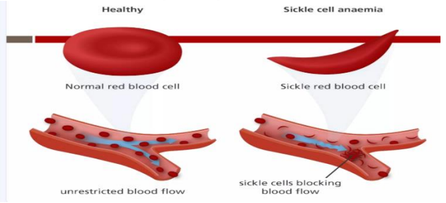
INTRODUCTION

Sickle cell disease (SCD) is an inherited red blood cell disorder. The typically round, healthy red blood cells that transport oxygen throughout the body become C-shaped (hence, the term sickle); these hard, sticky cells are short-lived, keeping red blood cells in short supply and interfere with healthy blood flow. Patients with SCD experience pain and other serious problems such as infection, acute chest syndrome, stroke, and skin ulcers that often convert into chronic nonhealing wounds. The challenge in treating patients with SCD is addressing the propensity toward infection without exacerbating pain. This case report describes the successful treatment of a female toddler with SCD in managing the blood requirements during a major neurosurgery for Lumbosacral Lipomyelocele.



CASE REPORT:

A 1yr 8month old female child with known case of sickle cell anaemia with Lumbosacral Lipomyelocele native of Orissa village, admitted for a corrective surgery under Neurosurgery unit. She was the first child, term AGA from normal vaginal delivery with birth weight 2.5 kg, uneventful other than, received phototherapy for 6 hrs in neonatal period. No previous hospital admissions or past transfusions. Mother and maternal grandmother known case of sickle cell disease.

At the age of 1 yr mother noticed gait abnormality when the baby started walking. On admission child weight: 9.5 kg, alert, moving all four limbs.

Her HbS was 72***%, Hb electrophoresis suggestive of homozygous for HbS with hereditary persistence of Fetal Hb.*** Sickling Test was Negative. Cardiac Echo was normal.

Blood group as O Rh positive, DAT, IAT - Negative, Auto-control-Neg

Parameter	Pre-transfusion	Post-transfusion	Post-0perative
Hb	8.9 g/dl	12.7	10.5
pcv	26	33.2	28
TC	11600	11500	10200
Plt count	3.51 L	2.66 L	2.8 L
T Bil/Direct	1.8/0.3	1.3/0.2	0.8/0.2
Sgot/sgpt	45/16	42/18	40/16
PT	17.2/ 14.1	16/14.2	16/14

Antibody screening & Identification- Negative

Extended Phenotyping of the child done was positive for s, P1, Fya, k.

So we decided to issue crossmatch compatible c, E, K negative Leucoreduced Fresh PRC units.

MANAGEMENT:

Selected c, E negative O rh positive donor from the Extended Phenotype Registry pool. We traced locally available 3 donors from our registry. Phlebotomy done screened and reserved the units and simple transfusion of the PRC unit in aliquots. Transfusion done in 10-15 ml/kg over 4 hrs. her Hb level improved, Hb S percentage reduced to 40% after 3 successive transfusions of 120 ml. Intra-0peratively 120 ml PRC transfused and the surgery was uneventful without any complications of hyper viscosity and acute ischaemic stroke. Child was discharged on the 7TH day

CONCLUSION:

A multidisciplinary approach and a team work which concluded in a successful major neurosurgical operation in a toddler with sickle cell disease. From our effort of Extended Phenotype Registry, we were able to do our task in a swift way and planned approach without any delay and concluded into a success story.