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Managing Post Transfusion Hyperhemolysis in E-β-Thalassemia With Multiple Red Cell Alloantibodies and Subsequent Support Without Transfusion - A Case Study

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INTRODUCTION

Hyperhemolysis syndrome is a rare and life threatening condition characterized by:

- Destruction of autologous and transfused RBCs
- Low post transfusion Hemoglon level than pre transfusion
- High LDH
- High reticulocyte count

CASE

- A 43-year-old female a c/o E-βThalassemia (P_{3+0}) with a history of infrequent PRBC transfusions commencing from peripartum period (2-3 units/year for past 22 years) experienced increased transfusion requirements (1 unit/week) over last 9 months.
- She was admitted under TM clinic as she was deteriorating due to hemolytic transfusion reaction.
- Later she was **diagnosed as Hyperhemolysis syndrome** and a successful management of HHS done at along with E-β Thalassemias at TM clinic.

Clinical Features

- Pallor,icterus, high colored urine
- Fever with pain in back,legs
- Hepatosplenomegaly (L-8 cm, Spleen-25 cm Clinically)
- Shortness of breath

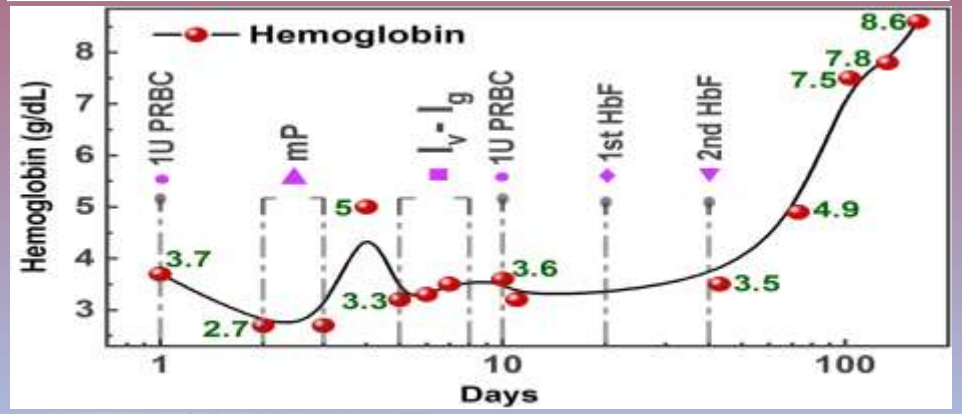
❑ IH WORK UP

- On physical appearance plasma was hemolysed, icteric.
- Saline control- positive, **Direct Antiglobulin test- positive (1+ in AHG phase)**
- Auto control- negative
- **Antibody screening:** On 3 cell panel was pan reactive
- Antibody identification (11 cell panel): was positive with different strength.
- After differential adsorption the **alloantibodies found were against c, E, Jkb,M Antigen**
- **Phenotype:** R1R1,(K-k+),(Jka+ Jkb-), (M-N+)

INVESTIGATIONS

LAB INVESTIGATIONS	1ST WEEK	2ND WEEK	3RD WEEK	1ST FOLLOW UP	2ND FOLLOW UP	3RD FOLLOW UP	4TH FOLLOW UP
LDH(U/L)	2965	1052	825	802	666	339	333
TB/DB(mg/dl)	6.2/2.4	4.1/1.8	2.3/0.9	2.2/0.9	2.2/0.8	1.9/0.8	1.8/0.8
ALT/AST(U/L)	166/64	70/38	76/66	74/64	35/40	51/48	35/40
Ur/Cr(mg/dl)	144/1.5	86/0.9	71/0.9	43/0.8	35/0.5	33/0.5	27/0.36

SPLEEN	25 cm	25 cm	25 cm	22 cm	22 cm	19 cm	17 cm
LIVER	8 cm	8cm	8 cm	6 cm	6 cm	4 cm	4 cm



- **REFERENCES:Anderson**
- https://doi.org/10.1182/blood-2021-152788
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MANAGEMENT

❑ **To manage Hyperhemolysis syndrome:**

- Inj. Methylprednisolone (500 mg) for 2 days
- IV-IG (2g/kg BW)
- O2 support and management for raised Ur/Cr

❑ For subsequent transfusion free support and Hemoglobin incremenmt:

- HbF inducers (Hydroxyurea-500 mg OD, Thalidomode-50 mg ODHS)
- oral corticosteroids (tapered dose as per body weight)

RESULT

- Patient was discharged with single HbF inducer.(thalidomide) and oral corticosteroids along with anti-heart failure measures. One month later Hb remains same.
- Second HbF inducer, hydroxyurea was introduced. Subsequent follow-up showed:

1st follow up	2nd follow up	3rd follow up	4th follow up
4.9 g/dl	7.5 g/dl	7.8 g/dl	8.6 g/dl

DISCUSSION

Inj. Methyl Prednisolone and IV Immuniglobulin played role in management of hyperhemolysis syndrome. Increased transfusion demand was reduced with single followed by dual HbF inducer as per response. Response criteria were set on the basis of improved pre-transfusion hemoglobin (Baseline hemoglobin increment atleast 1.5g/dL), reduced transfusion requirement and organomegaly.

CONCLUSION

- Non transfusion measure like HbF inducer can help to bypass complicated transfusion situation. The use of HbF inducers played a crucial role in stabilizing hemoglobin levels and in enhancing the patient's quality of life.
- Transfusion Medicine care showed a prospective initiative to facilitate a comprehensive approach for TDT hemoglobinopathies.