

Management of Life-Threatening Haemolytic Anaemia in an Adolescent Jehovah's Witness

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Clinical History

16y Jehovah's Witness
 Known diagnosis of Primary Immunodeficiency,
 Previously treated with Steroids +/- IVIG
 Emergency Presentation

- abdo pain,
- jaundice,
- pallor
- hypotensive,
- tachycardic,
- Hb 50g/dl, nadir of 40g/dl

Established exacerbation of AIHA
 Advance Directive already in place which included

- No Primary Blood products
- IVIG acceptable
- Supported by parents

Management

Immediate

- IV Immunoglobulin- 1gm/kg/d for 2 days
- IV Methylprednisolone -1gm/d for 5 days

Transfer to ITU

Intense counselling of patient and family to accept blood transfusion

Trust Guidelines of Blood-sparing Management

- Erythropoietin stimulating agent (ESA), Erythropoietin
- IV Iron
- IM Cobalamin
- Folic acid

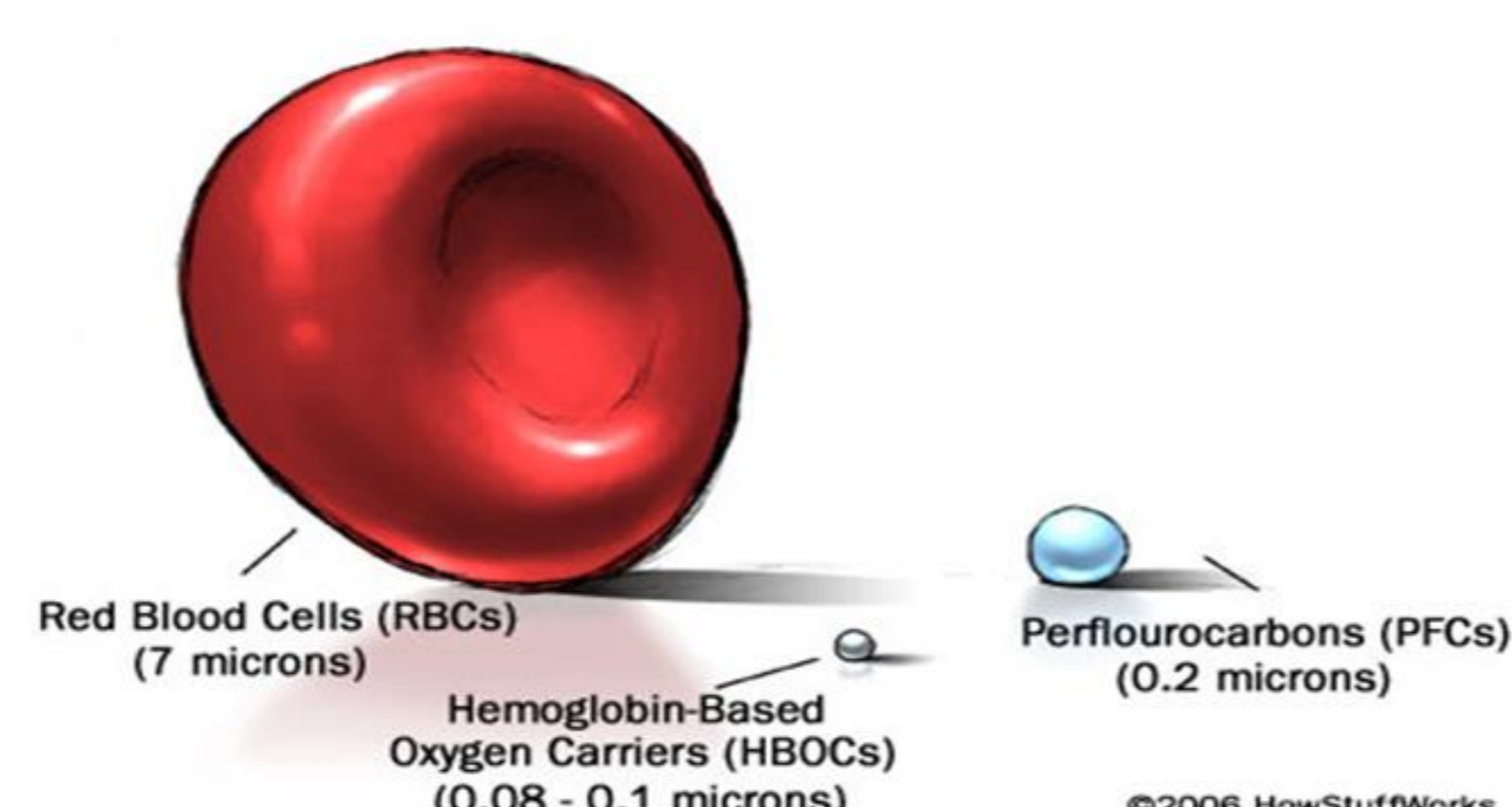
Minimal Phlebotomy

Alternatives Considered

Haemoglobin-based Oxygen carriers

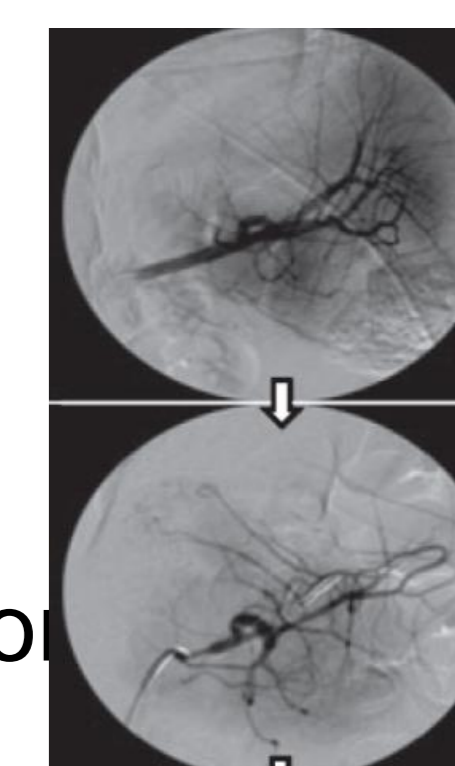
- purified, cross-linked acellular bovine Hb in a modified Ringer's solution
- Same oxygen carrying ability and dissociation
- No cross-matching required, long shelf life
- Approved for use in South Africa
- Not FDA approved
- Used as a "bridge"
- Half life is 19h,
- 1 Unit increases Hb by 0.63g/dL cf to PRBC unit increase of 0.87g/dL

- Concerns
- limited evidence base
 - safety profile
 - no previous use in Scotland



Splenic arterial embolization

- Selective embolization of superior splenic artery with microcatheter and a 6mm coil inserted
- Necrosis of superior lateral portion of spleen
- It can take about 8 days for a response
- Used in un-responsive disease which is not suitable for splenectomy
- Risk of post infarction syndrome



Intravenous Rituximab

- Delayed response

Elective intubation to decrease cardiac output

- Patient fully conscious
- Distress to family and staff

Transition to Adults

- Management of such profound anaemia without the use of blood products has now set a precedence for adult setting
- Structured approach will aid future management in any future relapses
- Advance directive will need to be renewed regularly throughout adulthood
- What role may the unproven aids of haemoglobin carriers play
- He and his family remain grateful for the care he received and this will be valuable in future relationships with healthcare professionals

References

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2. NHS GGC guidelines for the management of Jehovah's witness patients
3. Transfusions of Polymerized Bovine Hemoglobin in a Patient with Severe Autoimmune Hemolytic Anemia, Mullon J, Giacoppe G, Clagett C, Mcune D, and Dillard T. NEJM June 1, 2000
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Acknowledgments

All staff involved in this case and the patient who is keen to have his story heard