

Transfusion Errors and Reactions in Patients with Haemoglobinopathies

Sickle cell disease (SCD) and thalassemia syndromes are complex haemoglobinopathies and although often grouped together have different clinical manifestations and treatment. Red cell transfusion has an important role in the management of haemoglobinopathies and is required intermittently or long term for many patients. Alloimmunisation rates are high, in part due to ethnic variations in blood groups between patients with haemoglobinopathies and the predominantly Caucasian blood donor population. Most alloantibodies are to D, C and K antigens resulting in progressive difficulty in providing compatible blood. Preventing alloimmunisation to the antigens of the Rh and Kell blood group systems appears to reduce the development of antibodies to other blood groups.

Specific requirements for transfusion in SCD and thalassaemia



All patients with SCD and thalassaemia should have an extended RBC phenotype using a pre-transfusion sample



To reduce the risk of alloimmunisation, all patients with sickle cell disease and thalassaemia should receive full Rh (CcEe) and Kell matched units



Patients with a history of one or more alloantibodies should receive antigen-negative units for the corresponding antibody

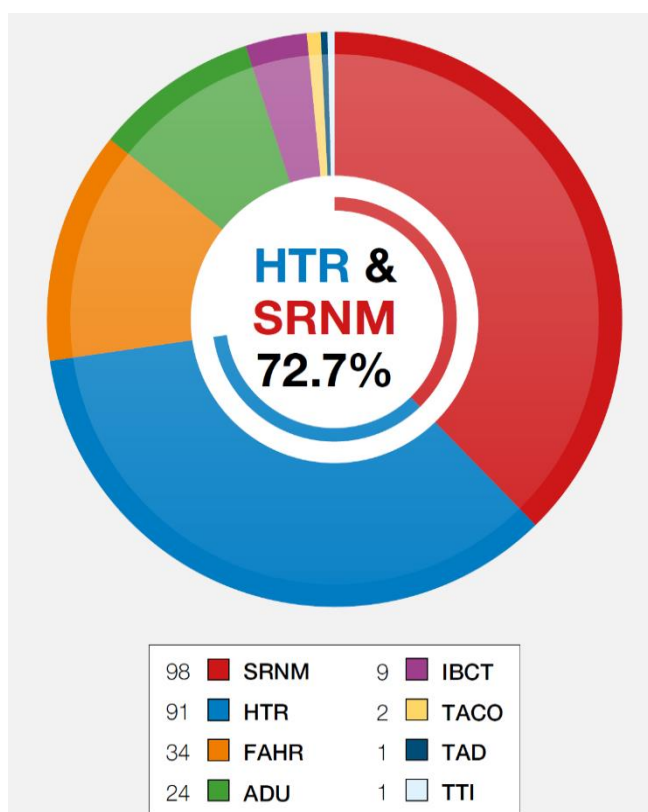


Reported reactions in SCD



The most frequent reported adverse events were specific requirements not met (SRNM) and haemolytic transfusion reactions (HTR), accounting for 37.7% and 35.0% respectively in cumulative data of 260 events from 2010-2019 (Figure 1)

Figure 1: Sickle cell disease 2010-2019 n=260



SCD patients are at higher risk of alloimmunisation and haemolytic transfusion reactions. This may be partly explained by ethnic variation of red cell antigens of donor and recipient



Post-transfusion hyperhaemolysis is considered a severe form of haemolytic transfusion reaction, characterized by the destruction of autologous red cells in addition to the transfused cells



The pathophysiology of hyperhaemolysis is poorly understood; proposed mechanisms include complement mediated haemolysis in association with the presence of alloantibodies and macrophage activation



Patients with a history of haemolytic transfusion reactions are at higher risk of recurrence and further transfusion must be carefully considered and specialist advice sought

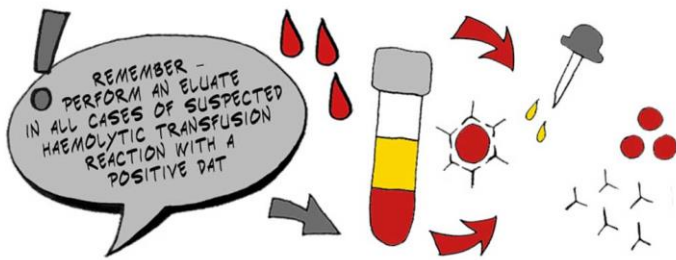
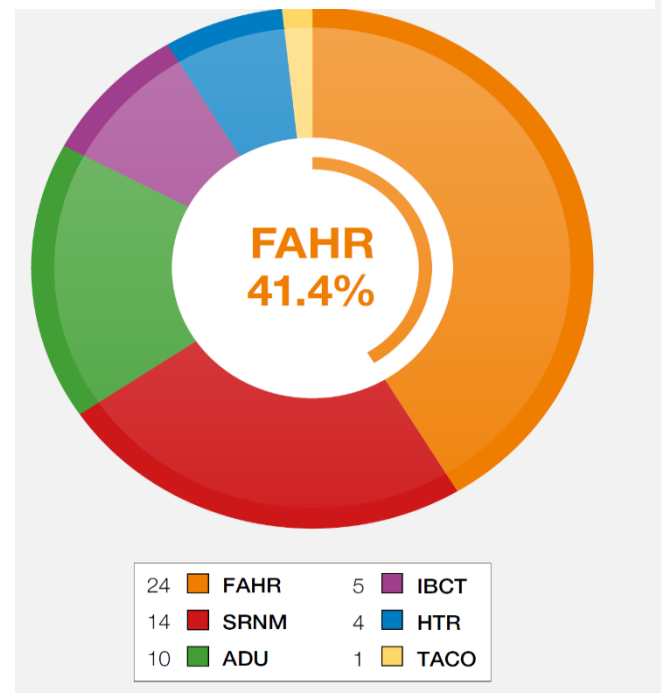
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Reported reactions in thalassaemia

The most commonly reported incidents were febrile, allergic and hypotensive reactions (FAHR) (41.4%) followed by specific requirements not met (24.1%) in cumulative data of 58 events from 2010-2019 (Figure 2).

Haemolytic transfusion reactions accounted for 4 (6.9%) of reported adverse events in thalassaemia patients.

Figure 2: Thalassaemia 2010-2019 n=58



Strategies to reduce the risk of adverse events in transfusion



One clear strategy to reduce the risk of adverse events is preventing unnecessary transfusion. Any decision to transfuse should be carefully considered in line with national guidance and following advice from a specialist haemoglobinopathy team



Procedures must be in place to ensure special requirements for blood products in patients with SCD and thalassaemia are provided - Many hospitals may not have a formal process for such requirements particularly in low prevalence areas



Ensuring haemoglobinopathy patients receive special requirements often relies on clinical teams communicating to the laboratory the diagnosis or need for special requirements and relies on laboratory awareness of special requirements. This should include confirming with national databases such as Sp-ICE in England if a patient has received transfusion elsewhere

Preventing unnecessary blood transfusion in haemoglobinopathy patients

Clinical team

Educate clinical teams about haemoglobinopathy and indications for transfusion
Transfusion history and confirmation of antibody history prior to transfusion
Communication with laboratory of patient details and specific requirements

Laboratory team

Ensure specific requirements can be highlighted on request form
System flags for haemoglobinopathy patients
Laboratory staff education on haemoglobinopathy

Patient

Empower patients to take an active role in managing and understanding their condition
Patients should carry 'special requirements' card detailing any antibodies or previous reactions
Contact details for specialist haemoglobinopathy team advice