Haemoglobin Disorders: Update 24

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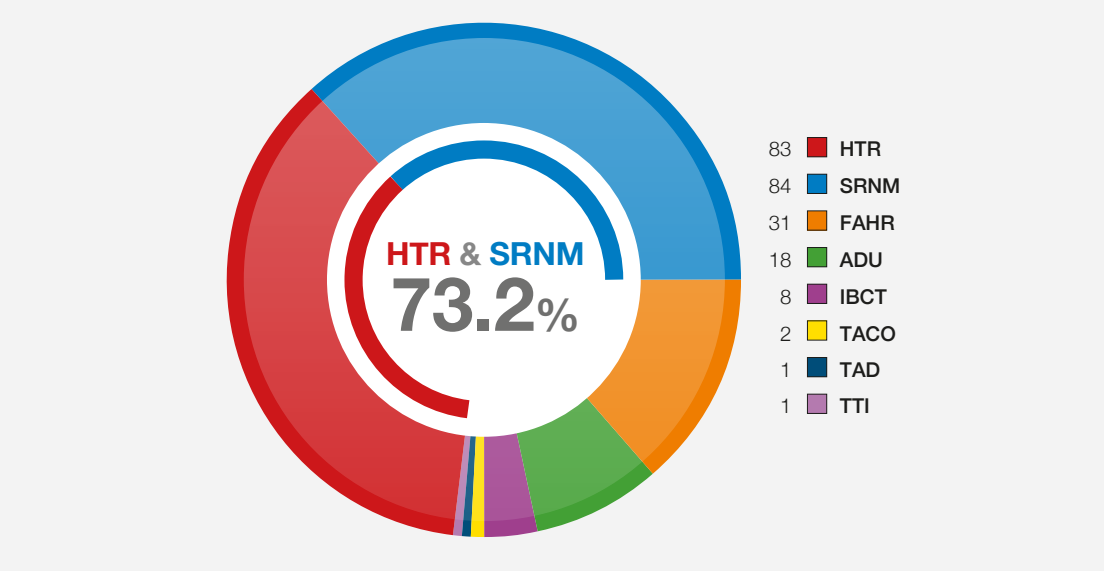
Thirty-nine incidents were reported in 2018 in patients with sickle cell disease (SCD) and transfusion-dependent thalassaemia (TDT). The most frequently reported incident was specific requirements not met, occurring in 16 cases. There were 9 cases of haemolytic transfusion reactions including 3 cases of hyperhaemolysis. There were no reported deaths directly related to complications of transfusion.

a) Sickle cell disease n=228



b) Thalassaemia n=52

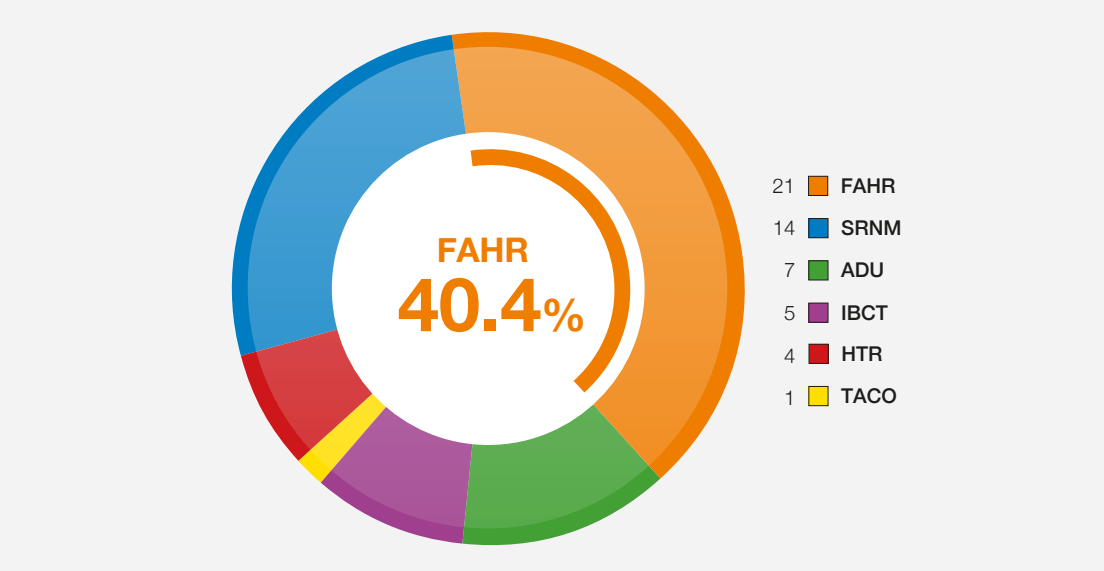


Figure 24.1: Cumulative data for adverse events in transfusion for patients with haemoglobin disorders 2010 to 2018

HTR=haemolytic transfusion reaction; SRNM=specific requirements not met; FAHR=febrile, allergic and hypotensive reaction; ADU=avoidable, delayed and under or overtransfusion; IBCT=incorrect blood component transfused; TACO=transfusion-associated circulatory overload; TAD=transfusion-associated dyspnoea; TTI=transfusion-transmitted infection

Careless practice n=8

An avoidable transfusion occurred in a young woman with SCD who had a haemoglobin (Hb) of 64g/L but no indication for transfusion. The general medical team referred to the generic hospital guidelines, which specified a Hb of less than 70g/L as the justification for transfusion. This guidance does not apply to patients with SCD and in this case the clinical haematology team was not consulted.

There were 7 reports of delayed transfusion. In 2 similar cases there was a delay in emergency transfusion for acute chest syndrome in SCD. In both cases the patient was transferred across sites and due to inadequate handover and careless practice the first patient did not receive the transfusion until the following day and for the second there was a 15-hour delay but no serious adverse outcome occurred.

In a further 5 cases there was a delay in planned transfusions for SCD (4) and TDT (1). In 3 cases the patient required admission to hospital to complete the transfusion.

Specific requirements not met n=16

Clinical causes n=5

In 4 cases of SCD the clinicians did not inform the laboratory of the diagnosis of SCD therefore patients did not receive extended phenotype-matched blood. One of these patients was also pregnant and did not receive cytomegalovirus (CMV)-screened units.

Laboratory causes n=11

The common problem in most cases was the laboratory not acting on the clinical information on the request stating the diagnosis of SCD or TDT. Two patients with SCD subsequently developed Rh antibodies (anti-C and anti-e).

A young man with SCD was having a planned exchange transfusion. The clinical team were not aware the patient had a history of previous antibodies (anti-M and C^w) and also stated that the patient had not previously been transfused. The laboratory did not check Specialist Services electronic reporting using Sunquest's Integrated Clinical Environment (Sp-ICE), which had a record of the previous antibodies, and so the patient received M-positive units. Subsequent antibody screening did detect anti-M; there was no serious adverse outcome.

Wrong transfusion n=1

A child with SCD attending for a routine transfusion was given the identification band of a second child also attending for transfusion. The nurse gave the first child the unit of blood prepared for the second child; the nurse did not use positive identification and the child answered yes to the wrong name or was distracted. The error was discovered during observations after the transfusion of one unit (50mL). Both children were group O D-positive but the child was C-, e- and the unit was C+, e+. The child also had an anti-Kp^a however the unit transfused was Kp^a-negative. There was no reported serious adverse outcome or new antibody development.

Febrile, allergic and hypotensive reactions n=3

There were 3 transfusion reactions reported in patients with SCD requiring discontinuation of transfusion but no serious adverse outcomes.

Haemolytic transfusion reactions n=9

All 9 patients had a diagnosis of SCD (7 female, 2 male).

There were 2 cases of acute haemolytic transfusion reactions both occurring in pregnant women after urgent transfusion. In 1 case the patient had a history of multiple alloantibodies (anti-A1, -M, -S, -Jk^a, -Le^a, -Le^b). She received two units of blood matched for all clinically significant antibodies; one unit was Le^a-positive and one unit Le^b-positive and with both units she developed intravascular haemolysis.

Subsequent Le^a-negative and Le^b-negative units were given without event. This is an interesting case in which antibodies usually deemed not clinically significant and cold-reacting were thought to have caused a reaction.

There were 7 delayed haemolytic transfusion reactions reported, 4 of which occurred after urgent transfusion; 1 of these patients was pregnant and another transfused in the early postpartum period.

Three of the delayed transfusion reactions were deemed to be cases of hyperhaemolysis. These were all non-pregnant female patients, and 2 of these reactions occurred following urgent transfusion. None of these patients had a history of alloantibodies.

Near miss n=2

A young male with SCD was having a planned transfusion but units selected were not HbS-negative; this was noticed by the nurse before the units were given.

Commentary

Red cell transfusions for patients with haemoglobinopathies can be life-saving but there are important risks in this unique group of patients including an increased risk of alloimmunisation and hyperhaemolysis.

It is important to recognise a delayed haemolytic transfusion reaction early so these patients can be managed appropriately with consideration of temporarily withholding further transfusions when safe to do so, to avoid further haemolysis (Pirenne and Yazdanbakhsh 2018).

Revised standards for the clinical care of adults with SCD in the United Kingdom (UK) were published in May 2018 (Sickle Cell Society 2018). These can be downloaded from the Sickle Cell Society website.

Recommendations

- Each transfusion for a patient with sickle cell disease (SCD) should be clearly indicated in line with British Society for Haematology (BSH) guidance (BSH Davis et al. 2016) and must be authorised by the haematology team

Action: Consultant Haematologists and Hospital Transfusion Teams

- All hospital transfusion laboratories should have a robust system to ensure that a haemoglobinopathy diagnosis is highlighted on the blood request form so that mandatory specific requirements are not missed. This is imperative to reduce the risk of alloimmunisation, which can have serious implications for these patients
- Any history of red cell antibodies must be sought out. In England this should include accessing Specialist Services electronic reporting using Sunquest's Integrated Clinical Environment (Sp-ICE) to check for any historical antibodies. The presence of currently undetectable historical antibodies increases the risk of delayed haemolytic transfusion reactions according to many studies (Narbey et al. 2017)

Action: HTT and Transfusion Laboratory Managers

References

BSH Davis B, Allard S, Qureshi A, et al. (2016) Guidelines on red cell transfusion in sickle cell disease. Part I: principles and laboratory aspects. *Br J Haematol* 2016;**176(2)**:179-191.

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Narbey D, Habibi A, Chadebech P, et al. (2017) Incidence and predictive score for delayed hemolytic transfusion reaction in adults with sickle cell disease. *Am J Hematol* 2017;**92(12)**:1340-1348.

Pirenne F and Yazdanbakhsh K. (2018) How I safely transfuse patients with sickle-cell disease and manage delayed hemolytic transfusion reactions. *Blood* 2018;**131(25)**:2773-2781.

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