23 Haemoglobin Disorders: Update

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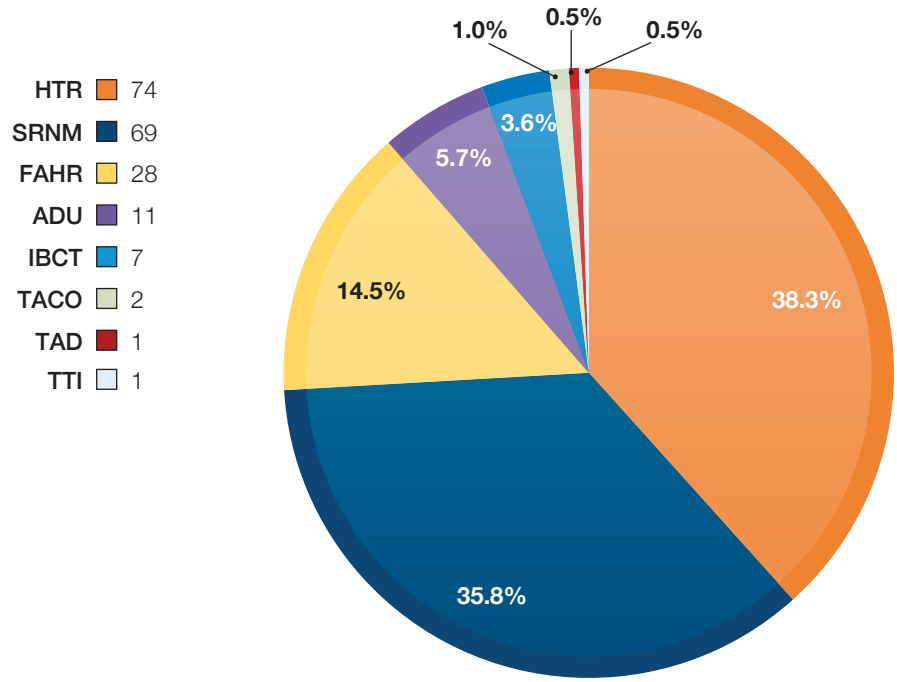
Forty-five incidents were reported. The majority were instances where the specific requirements were not met n=15, clinical errors in 4 and laboratory errors in 11, followed by haemolytic transfusion reactions n=14, 13/14 in patients with sickle cell disease (SCD). There were no deaths directly related to complications of transfusion; one woman with SCD died (Case 19.1 Chapter 19, Haemolytic Transfusion Reactions (HTR)) due to complications of SCD. The median age was 22, range 2 to 70 years; only 10 were over 40 years of age, in contrast to the median and ranges of the transfused population overall reported to SHOT: median 50 years, range 0 to 101.

Major morbidity occurred in 6 patients with SCD where transfusion was complicated by hyperhaemolysis. These are discussed in Chapter 19, Haemolytic Transfusion Reactions (HTR). There were 3 additional cases of major morbidity in febrile, acute and hypotensive reactions (FAHR) where the patients were admitted following their reactions.

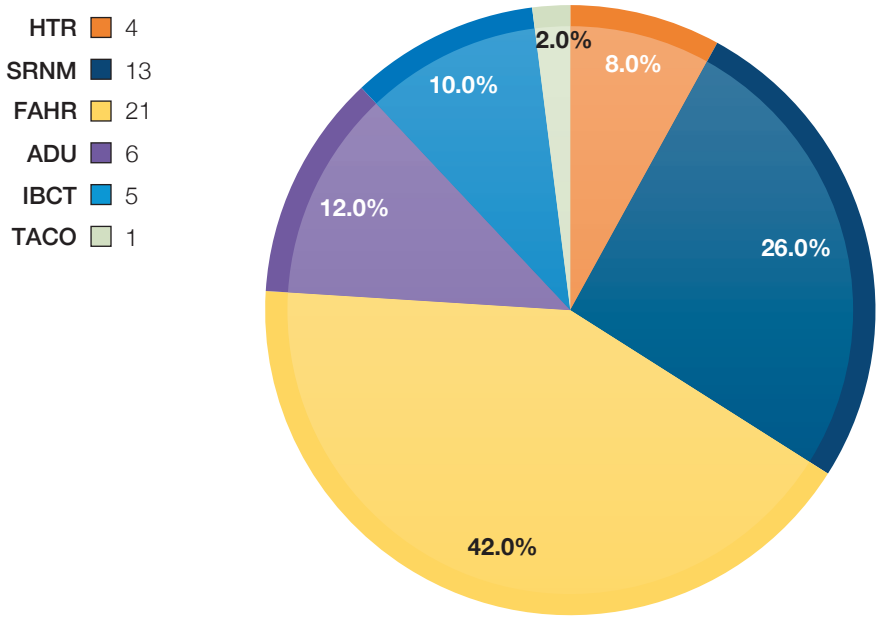
Figure 23.1: Cumulative data for adverse events in transfusion for patients with haemoglobin disorders 2010 to 2017

a) Sickle cell disease n=193



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

b) Thalassaemia n=50



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Careless practice n=4

Delay was reported for an exchange transfusion for a young pregnant woman with sickle cell disease but the clinical area did not inform the laboratory of the pregnancy so cytomegalovirus (CMV)-screened antigen-matched units had to be re-ordered. The patient had her day-case admission rearranged. The prescriber was a registrar in haematology.

An avoidable transfusion occurred in a thalassaemia patient on regular transfusions because the pre-transfusion haemoglobin (Hb) 116g/L, was not reviewed until the second unit was in progress. The authoriser had written up three units to be transfused 'because this was the amount she usually received'.

A child with SCD was overtransfused due to transcription of the wrong patient weight. Another child had a wrong transfusion rate set up (to give the total volume in 1 hour).

Specific requirements not met n=15

Clinical causes n=4

In all 4 instances (all SCD) the clinicians did not inform the laboratory, in 3 cases that these were patients with SCD and in the other that the patient was pregnant. In 2/3 cases there were serial errors that could have been detected including the correct diagnosis on subsequent requests that the laboratory failed to notice. The pregnant patient received 26 CMV-unscreened units in a series of exchange transfusions. Although 'CMV-screened' was included on some requests the reporter noted that their laboratory information system was not able to flag CMV requirements for pregnancy in addition to the requirements for SCD.

Laboratory causes n=11

A young child in sickle crisis developed an alloantibody after receiving a unit crossmatched by the Blood Service which was not matched for the extended Rh phenotype despite full phenotype information available from the red cell immunohaematology (RCI) laboratory report.

A woman in her 60s with SCD and subarachnoid haemorrhage was admitted to a hospital where she was not previously known. Another hospital was contacted for further information; she was known to

have three historical antibodies not currently detectable but there was a verbal communication error resulting in transfusion of inappropriate antigen-positive red cells. The information was also available from Specialist Services Electronic Reporting using Sunquest ICE (Sp-ICE) but the biomedical scientist (BMS) did not have access to this as she had not been competency-assessed and this incident occurred overnight. Shortage of resources was cited. As a result of this incident all BMS staff now have access to Sp-ICE, and any antibodies identified there can be added to the laboratory information management system (LIMS).

The remaining incidents were mostly caused by failure to note the clinical information, failure to search for historical transfusion results, or simple errors in red cell selection. The majority showed more than one error with a maximum of six in one case, i.e. there were other opportunities to detect the first error (failure to note the SCD on the request form in a patient new to that hospital where transfusion was urgent).

Wrong transfusions n=2

A child with beta thalassaemia was transfused with the wrong unit. Two nurses checked the unit against the prescription and medical records. The blood was run through a giving set and then the nurses repeated the procedure for another unit destined for a second patient. They then took one unit to the bedside, checking the name and date of birth against the prescription. The transfusion was connected and begun before one of the nurses checked the unit label and realised this was the wrong patient. Fortunately, although ABO-non-identical (patient group A, unit group O), this was not an incompatible transfusion.

This was a basic failure to perform the procedure correctly. The unit must be checked with the patient identification (ID), not the prescription. This was a serious error. As a result, all day-unit staff were retrained and the two staff involved were prevented from performing the bedside checks until competency had been re-assessed.

A child with SCD was identified as having a D-variant but this was not entered properly into the LIMS so that 17 D-positive components were given for elective transfusions. As a result, the laboratory checked the extended genotype file for any similar cases, updated flags and all BMS staff are to be given access to Sp-ICE.

Febrile/acute allergic/hypotensive transfusion reactions n=8

(7 SCD, 1 beta thalassaemia)

The age range was 6-34 years. Four of the 7 patients with SCD were on exchange transfusion programmes. After the reaction a child under 10 years of age with complex antibodies was changed from an exchange programme to top ups.

Haemolytic transfusion reactions n=14

Thirteen of these were in patients with SCD, 1 with beta thalassaemia. One woman in her 40s died related to complications of SCD following caesarean section (Case 19.1 in Chapter 19, Haemolytic Transfusion Reactions (HTR)). She also had hyperhaemolysis.

Near miss n=2

A unit of red cells was recalled by the specialist laboratory as it was not Jk^b-negative. Communication was poor but the unit was changed to a compatible one prior to transfusion to a woman in her 20s with SCD.

The second case was a teenager with SCD in whom excessive transfusion was avoided in a sickle crisis by rechecking the unexpectedly low Hb result and adjusting the authorised volume.

Commentary

People with SCD are at risk of serious transfusion complications including haemolysis resulting in death (Chubar and Bisharat 2017). To reduce the risk of alloimmunisation patients should have a full red cell phenotype recorded at diagnosis and in England can be fully genotyped including Rh variants which enables more appropriate red cell selection (Rees et al. 2018). Every transfusion should have a clear indication (Chinchilla Langeber et al. 2018), and good communication with the transfusion laboratory is essential. Delayed haemolytic transfusion reactions with hyperhaemolysis are serious and difficult to manage (Pirenne et al. 2017). Patients should be monitored for early recognition of this complication so that urgent measures can be taken to manage it. Confusion is caused by the similarity between features of haemolytic reactions and the symptoms of SCD complications.

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

References

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