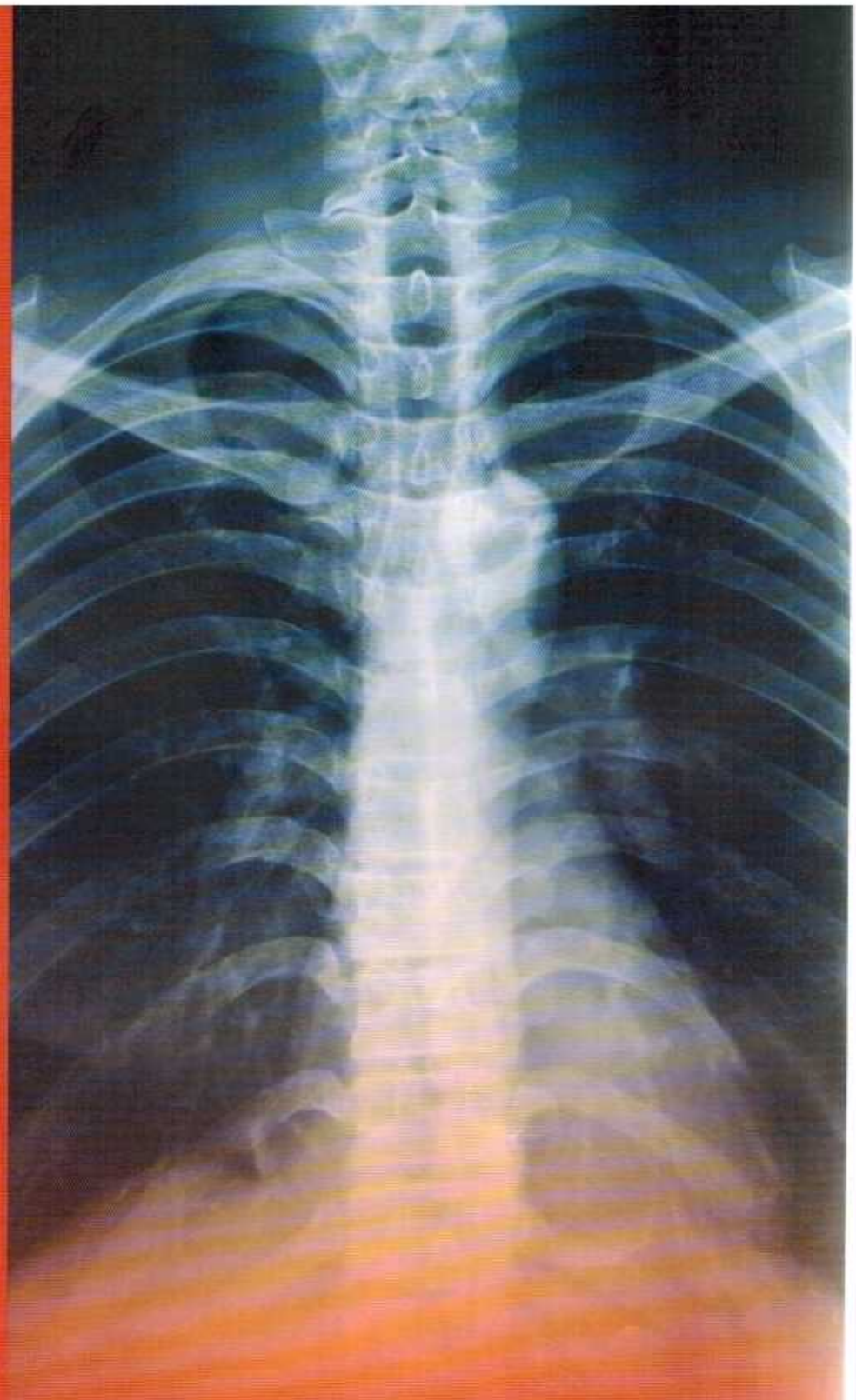
TRANSFUSION TODAY

Transfusion Today | Number 118, March 2019



Transfusion Risks and Complications

TRALI

Blooders Kids Project

Hyperhaemolysis

Basel Congress

ISBT

France Pirenne
President of the French Society of Blood Transfusion

ISBT ACADEMY

The first international seminar on delayed hemolytic transfusion reaction in Sickle Cell Disease

Delayed hemolytic transfusion reaction (DHTR) is the most dreaded complication of transfusion in sickle cell disease patients. Its frequency is underestimated, because the symptoms mimic vaso-occlusive crisis, and the underlying mechanism remains unclear. Alloimmunization is probably the leading cause of DHTR, but no antibodies are detectable in 30% of cases. There is currently no consensus concerning prevention and treatment, which depend on the underlying mechanism.

A meeting jointly supported by the International Society of Blood Transfusion (ISBT) and the French Society of Blood Transfusion (SFTS) on delayed hemolytic transfusion reaction (DHTR) in sickle cell disease took place December 17, 2018, in Creteil, France. This meeting was also organized with the support of Paris Est Créteil University, Labex GR-ex, Grand Paris Sud-Est Avenir, the Etablissement Français du Sang, under the aegis of the healthcare network for rare genetic diseases of red blood cells. This meeting was designed to consider all aspects of DHTR, bringing together specialists in the field, clinicians, scientists, and transfusion professionals, but also members of patient associations. More than 130 delegates from seven countries attended this international meeting.

Clinical, biological and therapeutic aspects were presented in the morning session. The definition of DHTR, including its most severe form, hyperhemolysis, was discussed. In SCD patients, DHTR has an incidence of about 4%, and accounts for 6% of all deaths. The clinical and biological presentation of DHTR was discussed in detail, together with its diagnosis (based on a nomogram using the post-transfusion HbA% and total Hb levels), prevention, and treatment. An analysis of French hemovigilance data for 2000 to 2016 was also presented, assessing the need to improve the recognition and declaration of this life-threatening reaction.

A number of cases, in adults and children, from France and from the US, were presented. These case reports demonstrated the difficulties of diagnosis and treatment decisions, but showed that eculizumab, an anti-C5 convertase antibody, probably stopped the hemolysis process efficiently in severe cases of hyperhemolysis. They also highlighted the importance of being aware of alloimmunization and DHTR history for the prevention of this syndrome.

The afternoon session was devoted to the pathophysiology of DHTR, and aimed to bring together the different pieces of a puzzle. The role of alloimmunization as the main trigger was discussed. Data from animal models and human studies have demonstrated the toxicity of the free heme released by hyperhemolysis to endothelial cells. The protective role of patrolling monocytes expressing high levels of heme oxidase-1, which scavenge endothelial cells injured by circulating heme, was also described. Finally, complement activation, through the binding of antibodies to microvesicles originating from red blood cells in the bloodstream, and to free heme, has shown this activation to be a major element of DHTR, providing evidence to a potential therapeutic role for complement inhibitors.

In conclusion, this meeting highlighted the need for more research in this field, and showed that the teams of clinicians and scientists working in this area are making progress towards understanding, preventing and managing this life-threatening condition more effectively.