

Innovations in Hematological research.

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Introduction

Hematology is the branch of medicine concerned with the study of the cause, prognosis, treatment, and prevention of diseases related to blood. It involves treating diseases that affect the production of blood and its components, such as blood cells, hemoglobin, blood proteins, bone marrow, platelets, blood vessels, spleen, and the mechanism of coagulation. Such diseases might include hemophilia, blood clots (thrombus), other bleeding disorders, and blood cancers such as leukemia, multiple myeloma, and lymphoma. The laboratory work that goes into the study of blood is frequently performed by a medical technologist or medical laboratory scientist.

Discussion

In volume 2, various aspects of hematology were discussed by the authors from different parts of the world. In the article Deivide Sousa Oliveira studied that “Plasmic score applicability for the diagnosis of thrombotic microangiopathy associated with ADAMTS13-acquired deficiency in a developing country” is a method to answer questions from 2011. Based on the pivotal validation work of the PLASMIC score, we tried to determine if the patients treated in our hospital had a correlation between ADAMTS13 activity levels and the clinical manifestation evaluated by those score. The analysis of ADAMTS13 activity was made by ELISA test that was collected in samples before the plasma exchange.

We found that in our population 100% (8 patients) with score above 6 had metalloproteinase activity below 10%. Comparison between the population with PLASMIC score above 6 and below 6 had important differences; the most important were median age (22 vs. 39.5), median days of plasma exchange (6.5 vs. 15.5), pregnancy (100% vs. 0%) and acute kidney injury (100% vs. 0%). In 50% of patients with PLASMIC score above 6 were finding ADAMTS13 inhibitors.

In addition we found that pregnancy, the need for early renal replacement therapy with dialysis and creatinine level above 2 mg/dL were independent factors associated with high levels of ADAMTS13 activity (p-value < 0.05). Patients without acquired deficiency of von Willebrand multimer cleavage metalloproteinase, more than half (75%) had an unfavorable outcome, remaining on dialysis after discharge or progressing to death. There is limitation in generalization our findings because the small number of patients.

The evolution of our understanding of TTP has been a milestone for Medicine, making it one of the most fatal diseases in a condition that, but when quickly diagnosed, has a high ability to be controlled. Use of the PLASMIC score is

vital and important, especially in developing countries, because of less access to specific tests. In this regard, the present study demonstrated that in our population the PLASMIC score can accurately differentiate patients with TTP and non-TTP [1].

Monica Pirastru et al, in their research article evaluated that The thalassemias, together with sickle cell disease (HBB: c. 20A>T), are the world's most common form of inherited anemia. The myriad manifestations of thalassemia result from the imbalanced synthesis of α -like and non- α -like globin chains and from the accumulation of unpaired counterpart.

Unpaired globin chains are unstable: they form intracellular aggregates which are insoluble and precipitate causing decreased deformability, membrane damage and selective removal of the damaged cell. Ineffective erythropoiesis and shortened red cell survival will lead to chronic anemia.

The β -thalassemias are characterized by a quantitative deficiency of β -globin chains underlain by a striking heterogeneity of molecular defects. Mutations that completely inactivate the β gene resulting in no β -globin production cause β^0 -thalassemia. Other mutations allow the production of some β globin but in markedly decreased amounts, and are classified as β^{+} -thalassemias.

The ability to perform DNA analysis has become an increasingly important requirement and the acquisition of such skills requires the development of suitable training programs involving hematologists, pediatricians, biologists and technical staff. A start has been made in developing and low-income countries and there is already considerable evidence that much more can still be done, including the further development of partnerships between countries where expertise in this field has been developed and countries where no such expertise exists.

References

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