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Editorial

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Hemolytic Uremic Syndrome: Early Diagnosis and Follow-up in Unusual Presentations

Síndrome hemolítica urêmica: diagnóstico precoce e acompanhamento em apresentações incomuns

Síndrome hemolítico urémico: diagnóstico precoz y seguimiento en presentaciones inusuales

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Hemolytic uremic syndrome (HUS) is defined as the presence of the classic triad: microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury.[1] It can present in 90% of cases in the typical form, after a diarrheal episode associated with infection by *Shiga* toxin-producing *Escherichia coli*, or in the atypical form, associated with dysfunction of the alternative pathway of the complement system resulting from genetic conditions or secondary causes,[1, 2] such as that described in this editorial.

The initial approach to these cases requires a thorough clinical evaluation, observing the significant pallor presented by the child, with indication for serial blood transfusions, presence of petechiae and ecchymoses associated with thrombocytopenia and reduced urinary output and altered level of consciousness, related to the retention of nitrogenous waste products due to acute kidney injury, these data being confirmed by the results of laboratory tests.[3] At this point, hospitalization in an intensive care unit is necessary, initiating renal replacement therapy, preferably peritoneal dialysis, due to the lower risk of consumption of coagulation factors, and greater indication for platelet transfusion, which leads to an increase in thrombosis in the microcirculation and a worse prognosis.[1,2,3]

The most indicated laboratory tests to be requested in these cases are the complete blood count with platelet count, hemolysis markers such as schistocytes (peripheral blood smear), lactate dehydrogenase, haptoglobin, nitrogenous waste products, electrolytes, and dosage of the activity of ADAMTS13 and serum levels of the C3 fraction of the complement system, which, when reduced, suggests atypical HUS.

In HUS, anemia, thrombocytopenia, elevated lactate dehydrogenase, and reduced haptoglobin may be observed. ADAMTS13 activity below 10% indicates the presence of thrombotic thrombocytopenic purpura. Measurement of nitrogenous waste products and electrolytes is essential to assess renal dysfunction and indicate renal replacement therapy.[3,4,5]

The tetravalent meningococcal B conjugate vaccine (MenACW135Y) is recommended before the use of Eculizumab or Ravulizumab to protect against most meningococcal serotypes (at least 15 days before the start of therapy). Other vaccines are also indicated, such as Pn13, Pn23, Hib, and influenza, as well as updating the vaccination schedule, including booster doses.

Although the manufacturer recommends the use of antibiotics for only 15 days after vaccination, the use of prophylactic antibiotics (against meningococcal disease) is recommended while the patient is undergoing treatment with a C5 inhibitor, in case prior vaccination is not possible.[4]

The article entitled “Atypical post-staphylococcal hemolytic uremic syndrome: A pediatric case report in a tertiary hospital”, published in this issue, not only describes a rare case, fulfilling an essential educational role by alerting pediatricians, nephrologists and intensivists about an unusual etiology of HUS, but also contributes to the discussion about the prognosis and follow-up of these patients. Although HUS associated with staphylococcal infection may result in renal recovery, the risk of long-term sequelae – such as hypertension and chronic kidney disease – should not be underestimated.

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