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Typical or Atypical Hemolytic Uremic Syndrome and the Use of Eculizumab: 4 Illustrative Cases.

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Abstract

Typical hemolytic uremic syndrome (HUS) in children is caused mostly by Escherichia coli 0157:H7 in our country. Atypical HUS (aHUS) causes include Streptococcus pneumoniae, methyl malonic aciduria, deficiency of ADAMST 13, and genetic or acquired disorder of the complement. Treatment of HUS relies on supportive measures while treatment of aHUS includes plasmapheresis and specific treatments. Recently, eculizumab has been proposed for the treatment of aHUS and many clinicians now believe that eculizumab should be the first-line standard of care. The purpose of this article is to illustrate the difficulties in the diagnostic process of HUS and therefore the subsequent problem to promptly choose the appropriate treatment. To date, workup of HUS continues to take many days leaving the clinicians with a choice between several therapeutic options. With the emergence of eculizumab, it becomes crucial to find faster diagnostic tools and to adapt HUS treatment protocols. We reported here clinical cases where eculizumab use was probably not appropriate once the correct diagnosis of typical HUS was made and cases where it would have been useful because of the late diagnosis of aHUS.

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