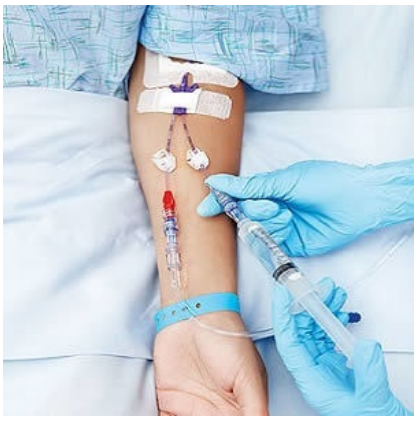
ICU
MANAGEMENT & PRACTICE

Expert Statements: Treatment of Critically Ill aHUS Patients



A review paper provides a summary of available data on the diagnosis and treatment strategies of atypical haemolytic uraemic syndrome (aHUS) in the ICU, with a view to enhancing the understanding of aHUS diagnosis and outcomes in patients managed in the ICU. The report was written by a panel of experts that met at an advisory board to discuss thrombotic microangiopathies in critically ill patients.

"Managing critically ill patients with aHUS requires basic skills that, in the absence of sufficient data from patients treated within the ICU, can be gleaned from an increasingly relevant literature outside the ICU. More data on critically ill patients with aHUS are needed to validate these conclusions within the ICU setting," concludes the report, which will appear in the journal CHEST.

aHUS is a rare but life-threatening condition that affects both children and adults. The annual incidence of aHUS is thought to be around 1–2 per million in adults. aHUS presents similarly to thrombotic thrombocytopenic purpura (TTP), and other causes or conditions with thrombotic microangiopathy (TMA) such as DIC or sepsis. TMAs are a group of disorders characterised by thrombocytopenia, microangiopathic haemolytic anaemia and organ dysfunction in which ischaemic organ injury can occur to the brain, kidneys, heart, pancreas, liver, lungs, eyes and skin.

Similarity in clinical presentation may hinder diagnosis and optimal treatment selection in the urgent setting within the ICU. Data on the clinical presentation and diagnosis of aHUS, particularly in the ICU setting, are limited by the small number of patients with this condition reported in ICU literature.

The critical nature of acute TMA means that a high proportion of patients may be admitted to the ICU at presentation. Owing to the severity of the progression of aHUS and other TMAs, a suspected diagnosis should be treated as a medical emergency, and initial supportive measures should be introduced with urgency. The British Committee for Standards in Haematology guidelines suggest that appropriate treatment should be initiated within 4–8 hours from diagnosis, as delays are associated with increased morbidity and mortality.

Eculizumab has been demonstrated to be effective and well tolerated in four prospective phase II trials in patients with aHUS. These trials were however not carried out in the critical care setting. In these trials, terminal complement inhibition with eculizumab was associated with inhibition of further TMA progression, increased platelet count and significant improvement in renal function.

Currently, there is no consensus on the diagnosis or treatment of aHUS for ICU specialists.

"Based on the paucity of adult aHUS cases overall and within the ICU, no specific recommendations

could be formally graded for the critical care setting. However, the expert panel recognises a core set of skills required by intensivists for diagnosing and managing patients with aHUS: recognising thrombotic microangiopathies, differentiating aHUS from related conditions, recognising involvement of other organ systems, understanding the pathophysiology of aHUS, knowing the diagnostic workup and relevant outcomes in critically ill aHUS patients, and knowing the standard of care for patients with aHUS based on available data and guidelines," the report says.

Source: CHEST
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