Atypical Hemolytic Uremic Syndrome

Achieving positive patient outcomes with early diagnosis and appropriate management

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BACKGROUND: Atypical hemolytic uremic syndrome (aHUS), a condition found in adult and pediatric populations, can be idiopathic or acquired as a result of major systemic changes. aHUS presents with a wide array of symptoms that can be attributed to other less dangerous conditions. Because of its complex nature and rare occurrence, it is typically diagnosed in later stages and with multiple organ involvement.

OBJECTIVES: This article provides an overview of aHUS and available interventions.

METHODS: Current aHUS literature was reviewed, and implications for nursing care were identified.

FINDINGS: Early diagnosis is crucial to achieve positive patient outcomes. The difference in pathology among the different thrombotic microangiopathies and their appropriate management must be understood. Although aHUS requires a multidisciplinary approach, nurses play a crucial role in assessing disease progression and identifying possible complications.

Classification and Definition

aHUS falls under the category of thrombotic microangiopathy (TMA). TMA is a disease process in which endothelial damage within capillaries and arterioles results in inflammation and activation of coagulants, leading to the formation of lesions caused by platelet-rich thrombi (Riedl et al., 2014). Although an immune response is typically an appropriate reaction to cellular damage, in this particular microenvironment, the process becomes uncontrolled. Thrombus formation at the micro level can cause more damage to the vessels, creating a cycle of inflammation and coagulation (Afshar-Kharghan, 2008). The lack of blood flow to distal tissues leads to tissue ischemia and what engenders the various clinical manifestations of a TMA. This will continue as long as the underlying condition is left untreated.

TMAs are divided into two main categories: thrombotic thrombocytopenic purpura (TTP) and hemolytic uremic syndrome (HUS) (Cataland & Wu, 2014). The overlap of presenting symptoms and organ involvement has some literature referring to them as a combined TFP/HUS. However, the pathophysiology behind TFP differs from that of HUS in that TFP is caused by deficient serum levels of the ADAMTS13 protease (Loirat & Frémeaux-Bacchi, 2011).

KEYWORDS thrombotic microangiopathy; atypical hemolytic uremic syndrome; thrombotic thrombocytopenic purpura; cancer

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