

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.

KEYWORDS

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

DIGITAL OBJECT IDENTIFIER

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ATYPICAL HEMOLYTIC UREMIC SYNDROME (aHUS) IS A RARE CONDITION that is difficult to diagnose and a challenge to manage because of its highly variable clinical manifestations. aHUS can be seen in the adult and pediatric population. Literature suggests that aHUS develops in patients who have major systemic changes, such as those with cancer (Nester & Thomas, 2012). Patients with a malignancy—whether a solid tumor or hematologic malignancy—have a variety of changes occur systemically as a result of their disease process. For example, some tumors can cause direct tissue damage and others may release cellular markers that can affect the host in various ways. Aside from cancer, aHUS has been associated with underlying complement dysfunctions; some bacterial and viral infections; certain pharmaceutical drugs, such as those for cancer treatment; and even pregnancy (Kavanagh, Goodship, & Richards, 2006). In an age in which evidence-based practice is the guiding principle to nursing care, nurses must understand this condition to provide quality care for this complex patient population.

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 is 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 TTP/HUS. However, the pathophysiology behind TTP differs from that of HUS in that TTP is caused by deficient serum levels of the ADAMTS13 protease (Loirat & Frémeaux-Bacchi, 2011).