Identification and Management

Sinusoidal obstruction syndrome/veno-occlusive disease related to hematopoietic stem cell transplantation

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BACKGROUND: Sinusoidal obstruction syndrome (SOS), also called hepatic veno-occlusive disease (VOD), is a potentially life-threatening complication of hematopoietic stem cell transplantation (HSCT) that affects about 1 in 7 patients undergoing this procedure. SOS/VOD is caused by the conditioning regimens administered prior to HSCT; in some cases, SOS/VOD results from chemotherapy alone. SOS/VOD usually develops within three weeks following HSCT; however, it can have later onset.

OBJECTIVES: Clearly understanding how SOS/VOD develops may support prompt detection and treatment when the condition arises.

METHODS: Research on identification and management of SOS/VOD is summarized, and data from clinical trials are reviewed.

FINDINGS: This article describes the syndrome, risk factors, signs and symptoms, and appropriate supportive care and treatment. The authors also offer some practical tips for detecting SOS/VOD and providing patient care, as well as the latest information on treating and preventing this condition.

KEYWORDS
sinusoidal obstruction syndrome; veno-occlusive disease; defibrotide

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SINUSOIDAL OBSTRUCTION SYNDROME/VENO-OCCLUSIVE DISEASE (SOS/VOD) is an unpredictable, potentially life-threatening complication of conditioning regimens for allogeneic or autologous hematopoietic stem cell transplantation (HSCT), or of chemotherapy alone. Its complex pathophysiology encompasses endothelial cell activation and damage (Carreras & Díaz-Ricart, 2011). Clinical characteristics of SOS/VOD include painful hepatomegaly, jaundice, fluid retention, rapid weight gain, and ascites (Bearman, 1995; DeLeve, Shulman, & McDonald, 2002; Kumar, DeLeve, Kamath, & Tefferi, 2003). Severe SOS/VOD, developing in about 20%–40% of SOS/VOD cases in patients receiving allogeneic transplantation, is typically characterized by the presence of multiorgan dysfunction (MOD), sometimes called multiorgan failure, involving renal and/or respiratory dysfunction, and may be associated with greater than 80% mortality (Coppell et al., 2010). Some mild cases may require only vigilance and supportive care; others can progress unpredictably, and a comprehensive response that includes pharmacotherapy is indicated, particularly for moderate to severe cases.

The incidence of SOS/VOD post-HSCT was estimated as 13.7% (range = 0%–62%) in a meta-analysis of 135 studies from 1979–2007 involving about 25,000 patients with HSCT (Coppell et al., 2010). Increasing use of reduced-intensity conditioning (RIC) regimens may have reduced SOS/VOD risk during recent years (Carreras et al., 2011); however, SOS/VOD occurs post-RIC, with one institution reporting an 8.8% incidence in patients receiving allogeneic transplantation (Tsirigotis et al., 2014). Regarding SOS/VOD after chemotherapy alone, incidence in one study was 11% (15 of 139 participants) (Kantarjian et al., 2016).

Pathophysiology

HSCT conditioning may trigger a potentially rapid pathophysiologic cascade leading to SOS/VOD. Toxic metabolites of conditioning agents may activate and damage endothelial cells lining hepatic sinusoids (Carreras & Díaz-Ricart, 2011). This activation leads to loss of vascular integrity, transformation of endothelial cells from antithrombotic to prothrombotic (Hunt & Jurd, 1998), and release of inflammatory cytokines (Carreras & Díaz-Ricart, 2011).