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# Bevacizumab in the treatment of hereditary hemorrhagic telangiectasia.

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INTRODUCTION: Hereditary hemorrhagic telangiectasia (HHT) is a rare multisystem vascular disorder characterized by epistaxis, mucocutaneous telangiectases and visceral arteriovenous malformations predisposing to shunting and hemorrhage. Angiogenesis has been implicated in the pathogenesis of HHT and therefore angiogenesis inhibitors appear to be the most promising agents. A literature search was performed to identify all articles reporting bevacizumab, a recombinant humanized monoclonal antibody that inhibits vascular endothelial growth factor (VEGF). We focused on the HHT pathogenesis, mechanism of action of the drug, its impact on the HHT symptoms and safety profile. AREAS COVERED: Systemic intravenous administration of bevacizumab improves the frequency and intensity of epistaxis, gastrointestinal (GI) bleeding episodes and liver arteriovenous malformations consequences. The safety profile of the systematic administration of the drug appears to be excellent with hypertension as the unique adverse effect reported so far. Its intranasal administration significantly decreases frequency and severity of nosebleeds and blood transfusion requirements. EXPERT OPINION: In the absence of randomized controlled trials in HHT, criteria of selecting patients and formal recommendations for treatment are lacking. For life-threatening epistaxis requiring blood transfusion, topical treatment with bevacizumab may be beneficial. Systemic treatment with bevacizumab is promising in symptomatic patients with organ involvement and life-threatening conditions.

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