Blue Rubber Bleb Nevus Syndrome (BRBNS) is a rare disorder characterized by widespread, protuberant venous malformations with the potential for life-threatening hemorrhage. The skin and GI tract are predominately affected, and GI bleeding is the most common complication. Case reports have demonstrated symptomatic lesions in the epidural, orbital, pleural, and pericardial spaces, as well as within solid organs, the GU tract, or elsewhere (1). Airway lesions have been reported, ranging from the oropharynx to the bronchus (2). This implies that peri-operative management of patients with BRBNS may be complex, yet there is a paucity of data on this subject. Our poster presentation aims to highlight the anesthetic implications of this rare disease.

A 14-yr-old male with BRBNS presented with two weeks of progressive dyspnea. He reported occasional bleeding from cutaneous lesions, but no stigmata of GI bleeding. Vital signs were consistent with compensated shock (tachycardia without hypotension), and workup revealed profound anemia, with a hemoglobin of 3.6 g/dL. Symptoms resolved after repeated packed red blood cell transfusions. The patient was scheduled for EGD, colonoscopy, and capsule endoscopy, which revealed friable lesions in the transverse colon. He was medically managed with iron supplementation and sirolimus.

BRBNS presents multiple challenges to the anesthesiologist. This patient presented with evidence of hemorrhagic shock requiring active resuscitation and transfusion. Large hemopericardium and hemothorax were ruled out based on clinical exam and chest radiograph. His airway was carefully assessed for vascular lesions, as lesions in the oral cavity and hypopharynx can lead to difficult direct laryngoscopy and/or airway bleeding. Fiberoptic intubation has been recommended as a preferred route of intubation to allow for visual inspection of the airway and to decrease the risk of traumatic hemorrhage (2). However, optimal airway management is not yet definitively established. This patient was uneventfully intubated with direct laryngoscopy, and no resistance was met advancing the endotracheal tube. When upper GI bleeding is evident, full-stomach precautions should be taken. Rapid-sequence induction and intubation may be challenging due to the risk of unrecognized airway lesions and bleeding with laryngoscopy. Awake fiberoptic intubation is a viable alternative, but offers significant challenges in the pediatric population. Finally, special attention should be paid to positioning and padding of the cutaneous lesions to avoid pressure and shearing, which may lead to rupture.

In summary, BRBNS offers unique challenges to the anesthesiologist, requiring a detailed understanding of the underlying pathophysiology in order to safely deliver general anesthesia.