

Spill the Beans: A Rare Case of a Late-Presenting Blue Rubber Bleb Nevus Syndrome

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Introduction

- Blue rubber bleb nevus syndrome (BRBNS), also known as Bean syndrome, is a rare genetic disorder with only about 200 cases documented in literature
- It involves the formation of venous malformations throughout various organ systems and is characterized by the development of blue nevi which are soft, elevated but compressible skin lesions often found on skin or mucous membranes
- Patients are often diagnosed at birth or in early childhood as they are born with a “dominant lesion” and develop further blue nevi as they age

Case Presentation

- An 84 year-old female with atrial fibrillation on Xarelto, hypertension, and hyperlipidemia presented with sudden onset lower abdominal pain and melanotic stools for the past three days
- On presentation, the patient was found to be tachycardic with a heart rate of 127 and was found to be hypotensive to 97/71mmHg
- While in the ED, the patient had additional episodes of witnessed melena
- Gastroenterology was consulted and the patient underwent an esophagogastroduodenoscopy (EGD) which showed multiple large vascular blebs in the esophagus and stomach concerning for BRBNS

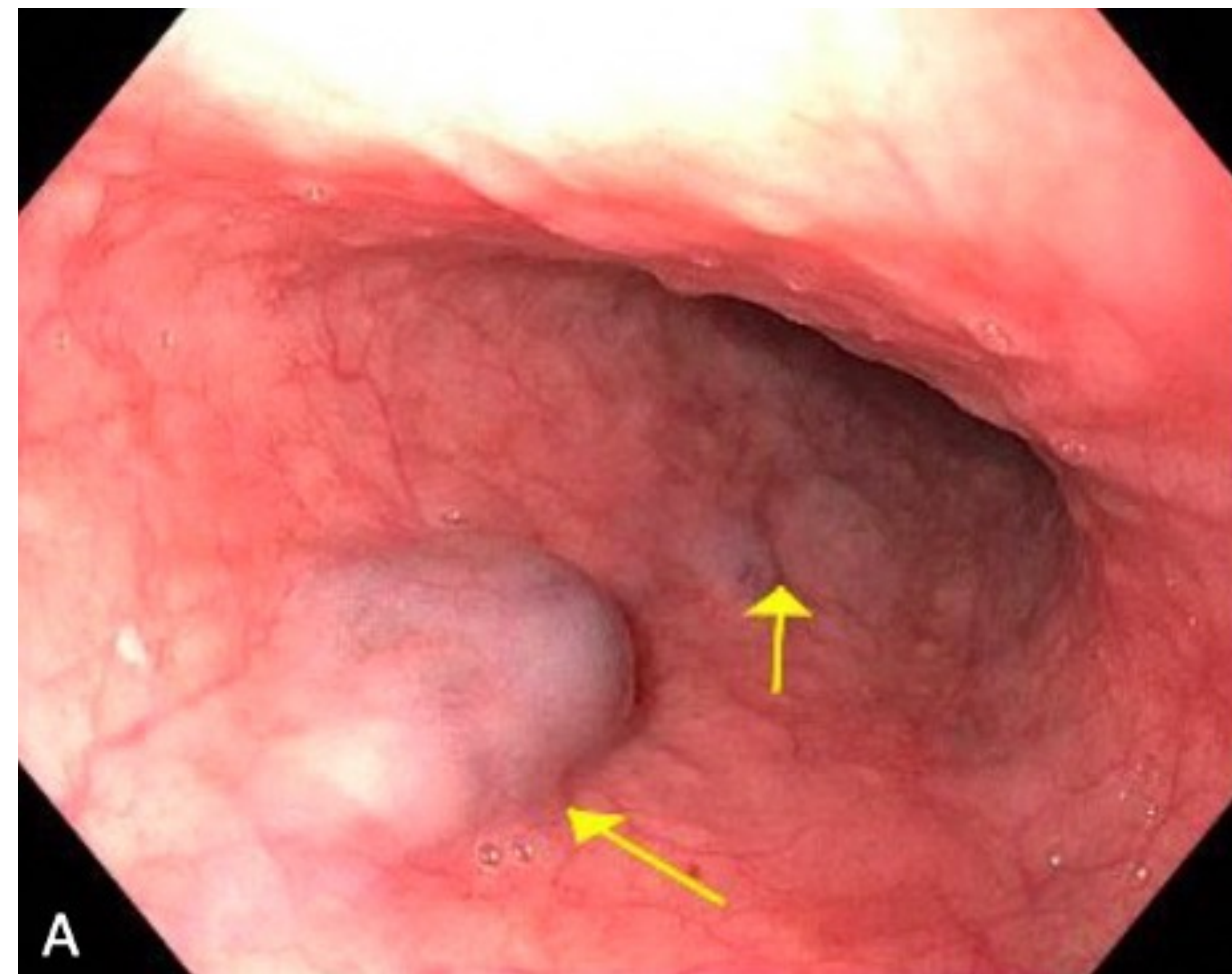
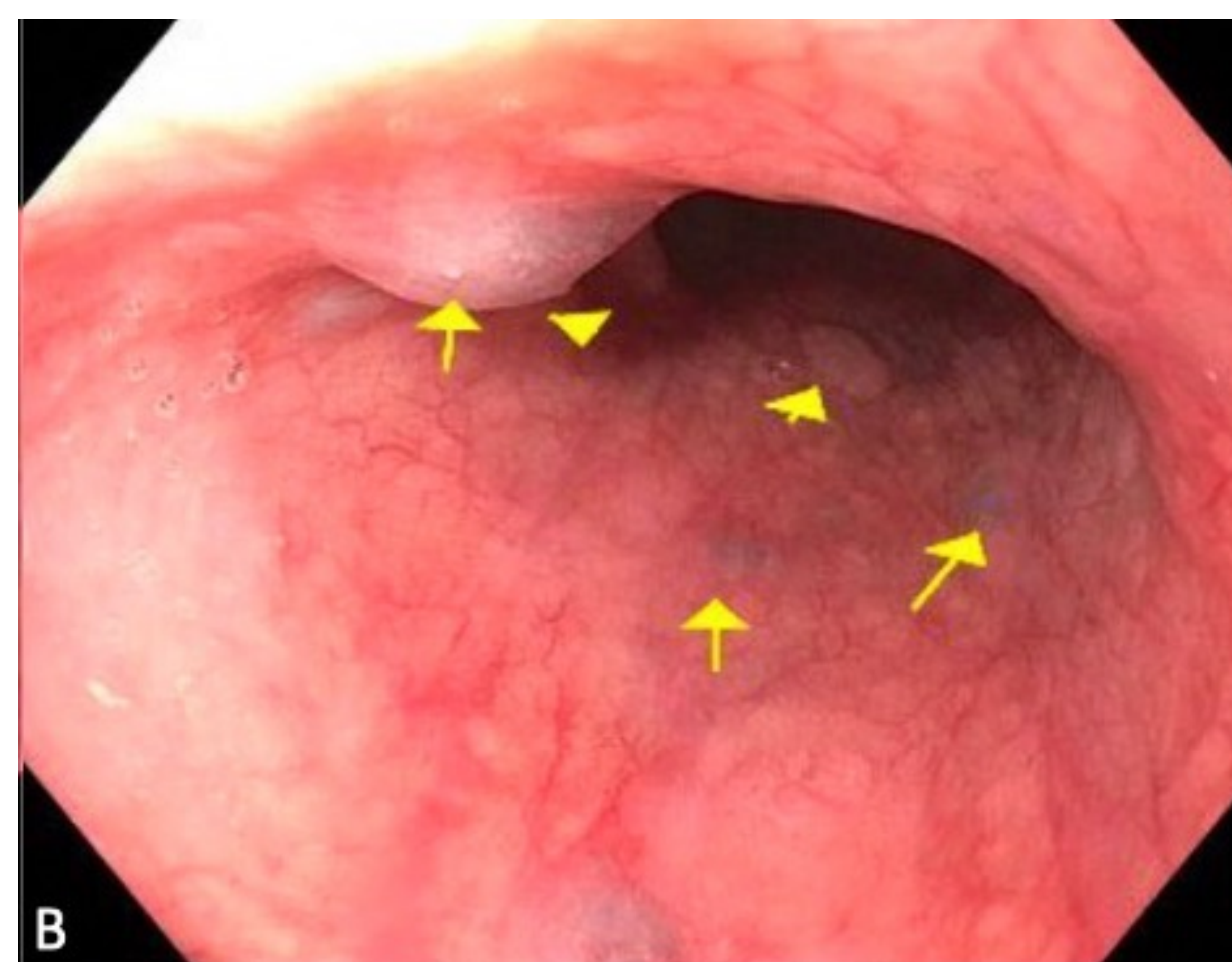


Figure 1A and 1B: Esophagogastroduodenoscopy (EGD) showing multiple large vascular blebs in the esophagus concerning for Blue rubber bleb nevus syndrome



Outcomes

- Due to concern for additional gastrointestinal bleeding events, the patient was deemed high-risk for further anticoagulation; Xarelto was discontinued
- Structural cardiology was consulted for a watchman device evaluation

Discussion

- Venous malformations related to BRBNS are commonly found on the skin and in the gastrointestinal tract
- Patients with BRBNS are at an increased risk of gastrointestinal bleeding and iron deficiency anemia as nevi can bleed spontaneously and can be life-threatening
- Treatment is usually supportive; however, surgical removal of blebs is an option
- Alternatives to anticoagulation are required in patients with BRBNS due to high-risk for bleeding

References

- Baigrie D, Rice AS, An IC. Blue Rubber Bleb Nevus Syndrome. [Updated 2022 Jul 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK541085/>
- Ugochi O. Ogu, Ghada Abusin, Rolla F. Abu-Arja, Janice M. Staber, "Successful Management of Blue Rubber Bleb Nevus Syndrome (BRBNS) with Sirolimus", Case Reports in Pediatrics, vol. 2018, Article ID 7654278, 5 pages, 2018. <https://doi.org/10.1155/2018/7654278>