

Learning Objectives

1. Describe a rare presentation of downhill esophageal varices
2. Review epidemiology and pathophysiology of disease
3. Discuss diagnostic evaluation, treatment options and surveillance

Case Presentation

HPI:

A 67-year-old female with atrial flutter presented with 3-weeks of melena and an elevated INR. She takes Warfarin and denies any recent dose adjustment. Outpatient labs revealed an INR >11, so she was instructed to go to the ER. She reported intermittent melena, fatigue, DOE and palpitations. She denied NSAID use but drinks 3 glasses of wine/night for ~20 years. She reported a previous diet mainly consisting of salads and fish but endorsed decreased po intake for ~2 months. She denied a personal history of malignancy or GI bleeding. She previously underwent a screening colonoscopy with removal of 2 benign polyps 5 years ago. On admission, she was afebrile and hemodynamically stable.

ROS:

Denied weight loss, chest/abdominal pain, cough, dysphagia, n/v, change in stool caliber or hematochezia.

Physical Exam:

Gen: Obese F in NAD
Cv: +3/6 LLSB holosystolic murmur
Abd: soft, ND, NTT, normoactive BS
Skin: no jaundice or bruising

Labs/Testing:

- Hgb 7.5 g/dL, INR 11.3, T. bili 1.9 mg/dL, LFT WNL
- **EGD** : large proximal EV, grade 1 distal EV, nodular gastric antral vascular ectasia (GAVE)
- **CT Chest**: No evidence of intrathoracic malignancy or SVC obstruction
- **CT AP**: cirrhotic morphology w/ portal HTN and splenomegaly
- **TTE**: moderate TR, RVSP 90 mmHg

Treatment:

- 5mg IV Vitamin K
- IV PPI BID
- Discontinuation of Warfarin on discharge

Diagnosis:

- Presumed upper GIB suspected due to nodular GAVE or mucosal oozing in the setting of dietary Vitamin K deficiency and supratherapeutic Warfarin dosing
- Downhill EV due to benign SVC obstruction (pulmonary HTN and TR)
- Decompensated cirrhosis likely cardiac vs. ETOH

Referral to Cardiology and Hepatology:

- RHC: mPAP >35 mmHg
- Plan for TJ liver bx with measurement of HVPG for transplant evaluation

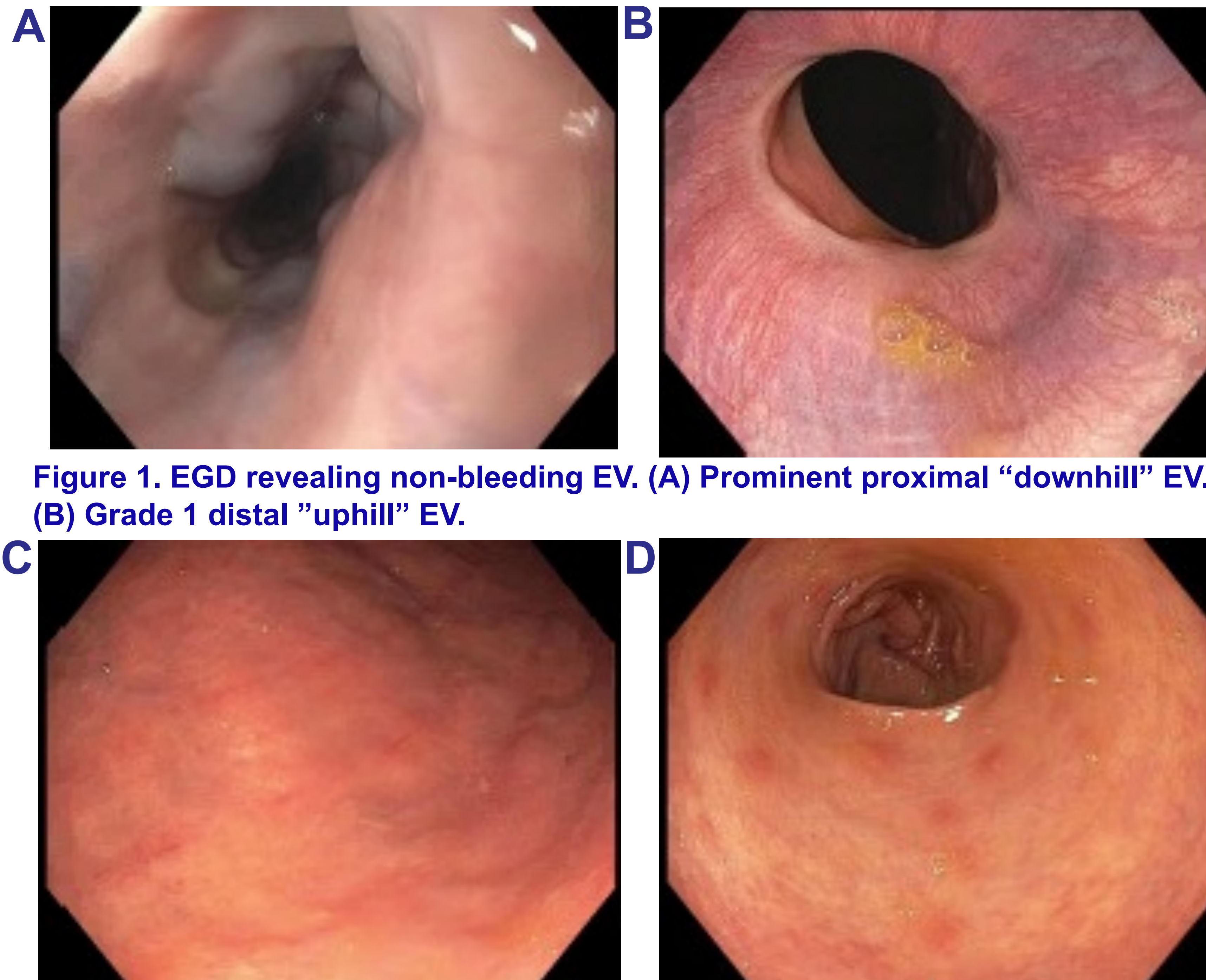


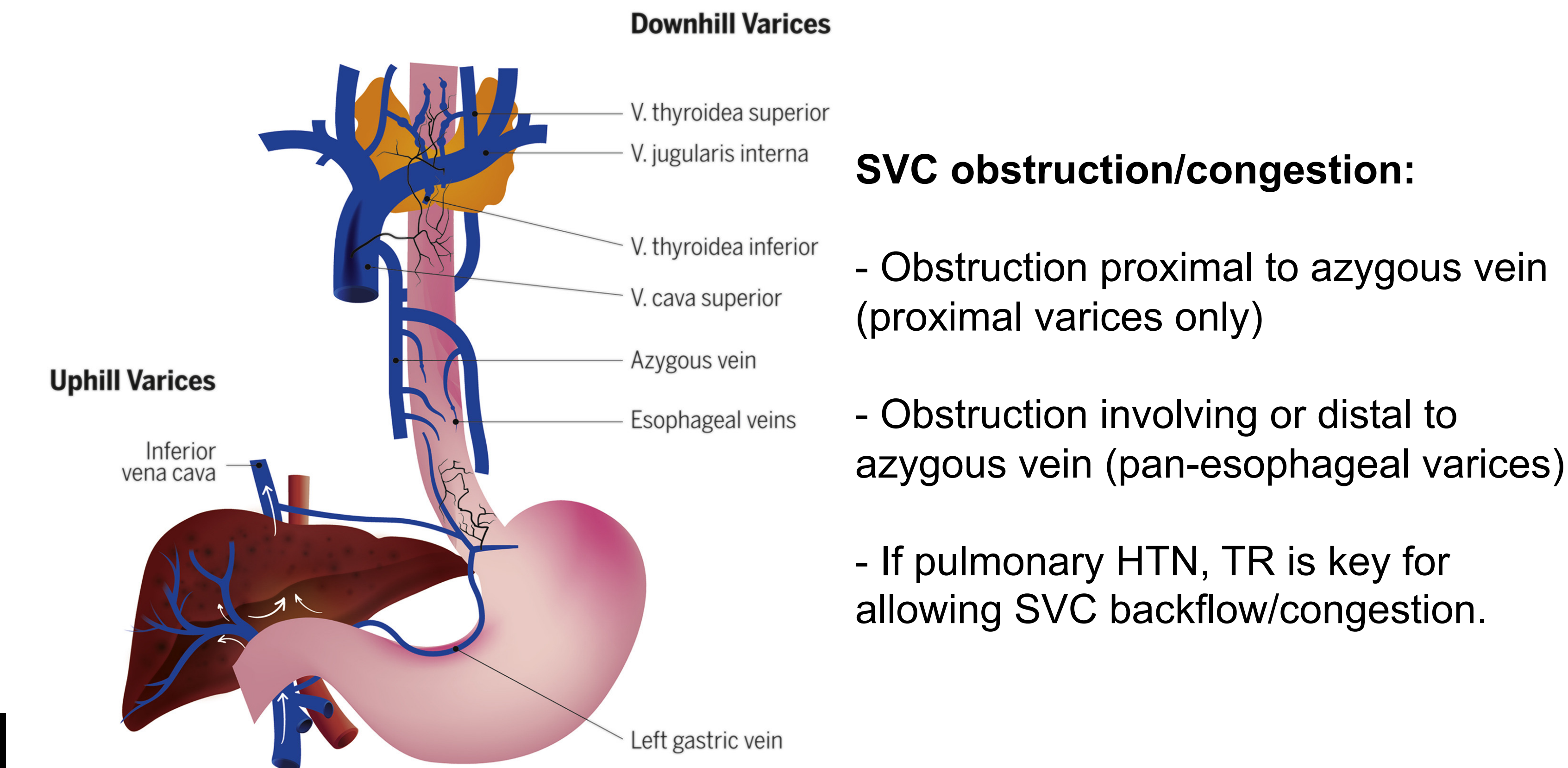
Figure 1. EGD revealing non-bleeding EV. (A) Prominent proximal "downhill" EV. (B) Grade 1 distal "uphill" EV.

Figure 2. EGD revealing nodular gastric antral vascular ectasia (GAVE). (C) "Watermelon stomach" from GAVE in the gastric body. (D) Nodular GAVE in the pyloric antrum.

Background

Epidemiology/Pathophysiology/Management:

- Male/Female 1:1
- Avg. age 68 years
- Represent 0.5% of all EV cases
- Develop by SVC obstruction resulting in retrograde venous blood flow through peri-esophageal collaterals located in the upper and middle esophagus.
- Most commonly caused by malignant SVC obstruction (~60%), iatrogenic (CVC, cardiac PM), thrombosis, COPD
- Also associated with other benign causes of SVC obstruction (ex. Behcet's disease, goiter, esophageal dysmotility)
- Rarely due to pulmonary arterial hypertension with concomitant tricuspid regurgitation
- Treatment involves correcting the underlying cause (ex. Surgical bypass, stenting, thrombolysis, chemotherapy, etc.)
- Endoscopic intervention reserved for emergent bleeding only:
 - Band ligation (preferred)
 - Sclerotherapy



SVC obstruction/congestion:

- Obstruction proximal to azygous vein (proximal varices only)
- Obstruction involving or distal to azygous vein (pan-esophageal varices)
- If pulmonary HTN, TR is key for allowing SVC backflow/congestion.

Discussion

Presentation/Diagnosis/Complications:

- Most commonly an asymptomatic incidental finding
- **Diagnosis:** EGD, video capsule endoscopy, CT/MRI angiography
- **Risk of hemorrhage:** extremely rare (0.1% of EV bleeds).
 - Increased risk w/ benign obstruction > malignancy related
 - ex. ESRD- central catheters/uremia, SVC syndrome- anticoagulant/antiplatelet use
- **Protective factors:**
 - submucosal location
 - less gastric acid exposure
 - no a/w coagulopathy or thrombocytopenia of cirrhosis
- **Surveillance:** If non-bleeding, likely no routine surveillance EGD needed
- **Complications of endoscopic intervention:** Bleeding, esophageal wall perforation w/ banding, stricture development w/ proximal esophageal banding, spinal infarction and pulmonary emboli w/ sclerotherapy

Conclusion

- Although rare, physicians must be mindful of downhill EV, especially in those with concern for UGIB and concomitant ESRD, hx CVC/cardiac PM, SVC syndrome, malignancy or severe pulmonary HTN + TR.
- If incidental downhill EV are found on EGD, it is crucial to undergo further w/u with radiographic imaging (ex. CTA, MRA), TTE or duplex US of neck vasculature and thyroid for malignancy r/o.
- Given the lack of reported cases, there are no guidelines for management and endoscopic surveillance. However, limited studies have provided evidence against routine surveillance in non-bleeding downhill EV.
- Endoscopic intervention via band ligation or sclerotherapy is high risk and reserved for emergent bleeding only.
- Likely more prevalent than stated in current literature, given asymptomatic presentation and may easily be missed without slow endoscopic retrieval through the middle and proximal esophagus.