



Heart Failure Case

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70-Year-Old Female

- Seeks another opinion for dyspnea
- Paroxysmal atrial fibrillation – prior ablation
 - Residual atrial level shunt
- Family history of HHT, remote personal clinical diagnosis
 - No genetic testing or prior treatment
- Chronic anemia related to epistaxis and GI bleeding
 - IV iron every 2 weeks to maintain Hb 8 -10 (12 – 15)



Labs

- Hb 10.2 g/dL (12 – 15), HCT 32.1% (35 – 44)
- Ferritin 309 mcg/L (11-307)
- NT-ProBNP 1062 pg/mL
- Creatinine 1.0
- ECG – atrial fibrillation

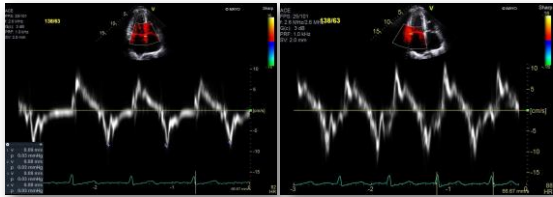


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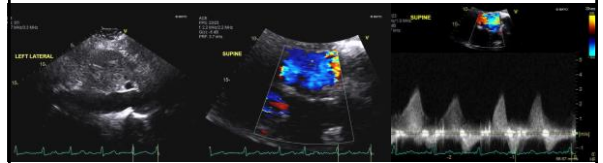
70-Year-Old Female with Dyspnea



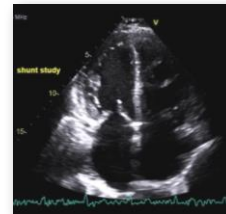
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Shunt Study

Echo Math

- Mild LV enlargement, EF 68%
 - CO 10 L/min, CI 6 L/min/m²
- Moderate RV enlargement
 - Calculated RVSP 79 mmHg
- Marked LAE
- Atrial level shunt

What do you suggest?

1. Repeat AF ablation
2. Close atrial level shunt
3. PH therapy
4. Transfusion
5. Something else

What is causing this patients symptoms?

1. Atrial level shunt
2. Atrial fibrillation
3. Idiopathic PH
4. Right heart failure
5. Other

Hereditary Hemorrhagic Telangiectasia



Hereditary hemorrhagic telangiectasia (HHT)

- Rare AD genetic disorder
 - 3 major types
 - 600 different HHT-causing mutations
- Abnormal blood vessels
 - Mucocutaneous telangiectasias cause epistaxis, GI bleeding, and iron-deficiency anemia
 - AV malformations (AVMs) commonly occur in pulmonary (50%), hepatic (30%), and cerebral (10%) circulations
 - Epistaxis usually earliest sign, often occurring in childhood

HHT – International Diagnostic Criteria

Curaçao diagnostic criteria

1. Spontaneous and recurrent epistaxis
 2. Multiple mucocutaneous telangiectasia at characteristic sites
 3. Visceral involvement (eg, GI telangiectasia; pulmonary, cerebral, or hepatic AVMs)
 4. First-degree relative with HHT
- Definite = 3+ criteria, Suspected = 2 criteria, Unlikely = 1 criterion
 - Often confirmed by gene testing

Hereditary hemorrhagic telangiectasia (HHT)

- Elevated plasma concentrations and tissue expression of vascular endothelial growth factor (VEGF) and transforming growth factor β (TGF- β)
 - TGF- β stimulates the production of VEGF, which plays a key role in angiogenesis

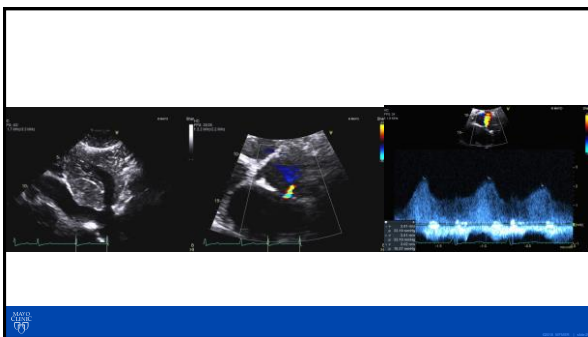
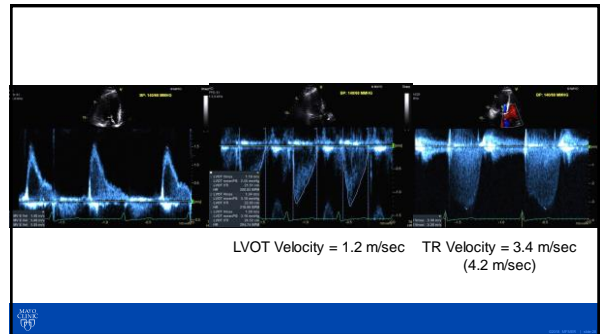
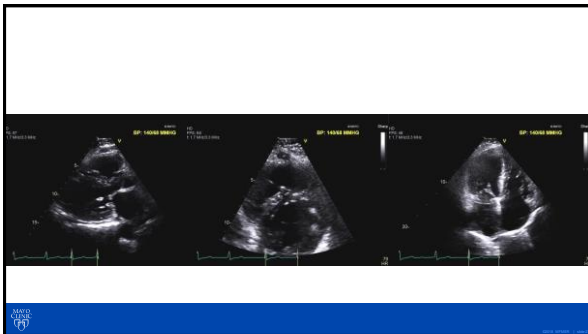
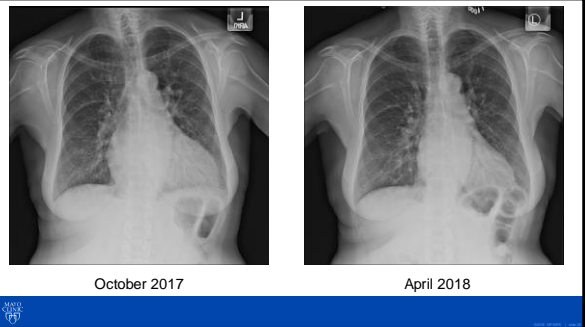
What to do?

- 34 HHT pts with refractory anemia
 - IV bevacizumab (Avastin) angiogenesis inhibitor monoclonal antibody over 4 yr period
 - Epistaxis severity score (ESS) were significantly reduced from 6.5 to 3.3 at one month of treatment
 - Transfusions decreased from 82% at baseline to 9% at end of study
 - Therapy was well tolerated

What Happened...

- Avastin – 5 mg/kg dose every 2 weeks (4 doses) followed by four doses at the same strength one month apart

Iyer VN et al: Mayo Clinic Proceedings 2018



Returned Six Months Later

- Marked improvement in epistaxis
- Cardiac output
 - 9.5 – 6 L/min
 - CI 3.43 L/min/m²
- No active CHF symptoms or dyspnea
- Hb 15.9 (prior 10.2 g)

Take Home Points – Heart Failure

- HHT can cause high output heart failure
 - Anemia, AVMs
- New treatments may improve clinical picture
- Unusual cause of reversible heart failure



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