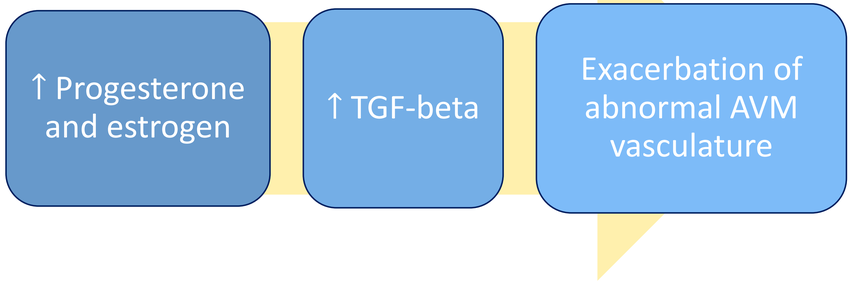
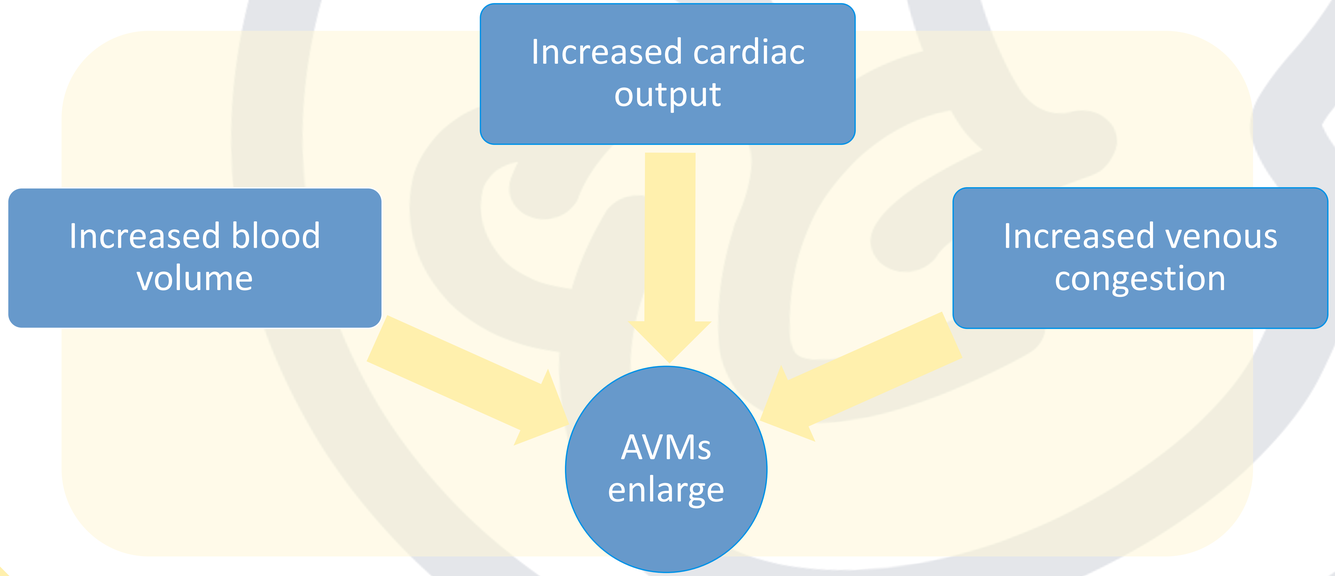
MANAGEMENT OF PULMONARY ARTERIOVENOUS MALFORMATIONS OF HEREDITARY HEMORRHAGIC TELANGIECTASIAS IN PREGNANCY

Christine Ha MD, Katherine Bartz MD, Peter Yeh MD, Stephanie Lim MD

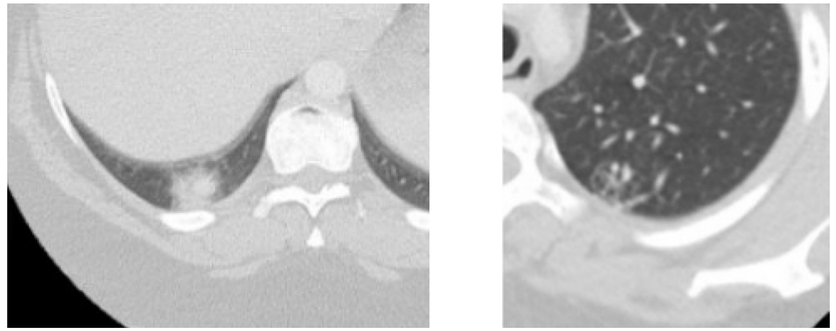
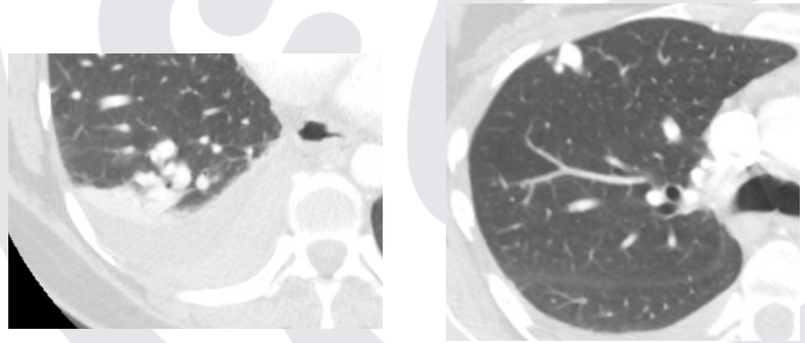
Background:

- Autosomal dominant
- Characterized by:
  - Mucocutaneous telangiectasias
  - AVMs of pulmonary, hepatic, cerebral circulations
- Most common complication
  - hemorrhage or stroke from untreated pulmonary AVMs





Case Series:

Past medical history	33 yo G2P1001 at 27w4d with obesity, gHTN	32 yo G1P0 at 27w1d otherwise healthy with recent PNA
Presentation	Gurgling sensation and single episode of hemoptysis	Acute hypoxic respiratory failure requiring HFNC
Diagnostics	**CTA chest:** Posterior basal RLL suspected AVM with adjacent hazy opacity suggesting interstitial congestion vs hemorrhage; also smaller similar finding in LUL 	**Thoracentesis:** chest tube 400 mL dark heme **TTE:** concerning for intracardiac shunt. **CTA chest:** numerous pulmonary AVM involving all lobes 
Treatment	- Urgent IR embolization of bleeding RLL AVM - POD4: Embolization of nonbleeding LUL AVM	- Urgent IR embolization for RUL and RLL targets - POD7: Embolization of nonbleeding L AVMs c/b epistaxis and reintubation
Follow up	- Stable postoperative ICU stay for monitoring - Discharged on POD7	- Epistaxis resolved, extubated POD1 - Chest tube removed and discharged POD11 - Future embolizations of remaining PAVMs planned

Discussion:

- Parturients with HHT should be screened for PAVMs:
  - TTE with bubble study
  - Low-dose non-contrast CT chest in second trimester
- PAVMs may lead to:
  - Hemoptysis
  - Hemothorax
  - Paradoxical embolic stroke
  - pHTN
  - High-output cardiac failure
- Asymptomatic PAVMs should be treated preconception or in second trimester
  - IR embolization includes risks of fetal anesthetic and limited radiation exposure
  - Current guidelines recommend treatment of PAVMs given high risk of maternal complications
- Patients with unknown PAVM status or untreated PAVMs require:
  - proximal air filters
  - prophylactic antibiotics

References:

1. GIM. 2011;13(7):607-616.
2. Can J Anaesth. 2009;56:374-384.
3. Ann Intern Med. 2020;173(12):989-1001.