CIRA Case of the Month

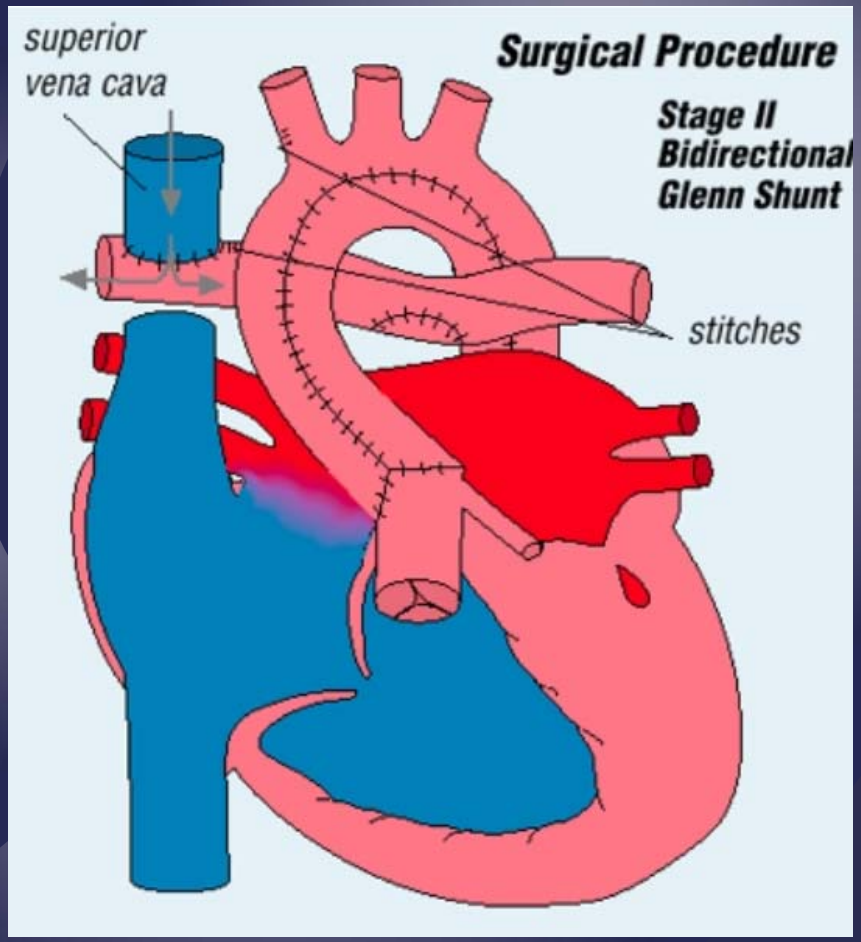
Case courtesy of Drs. N. Coffey, S. Ryan and A. Hadziomerovic
University of Ottawa, ON

Clinical History

- 23 year old male presenting with recurrent submassive and massive hemoptysis (since 2007)
- PMH:
  - Hypoplastic left ventricle
  - Double outlet right ventricle
  - Absent right PA
  - Hypoplastic left PA
  - IVC drains to right ventricle

Clinical History

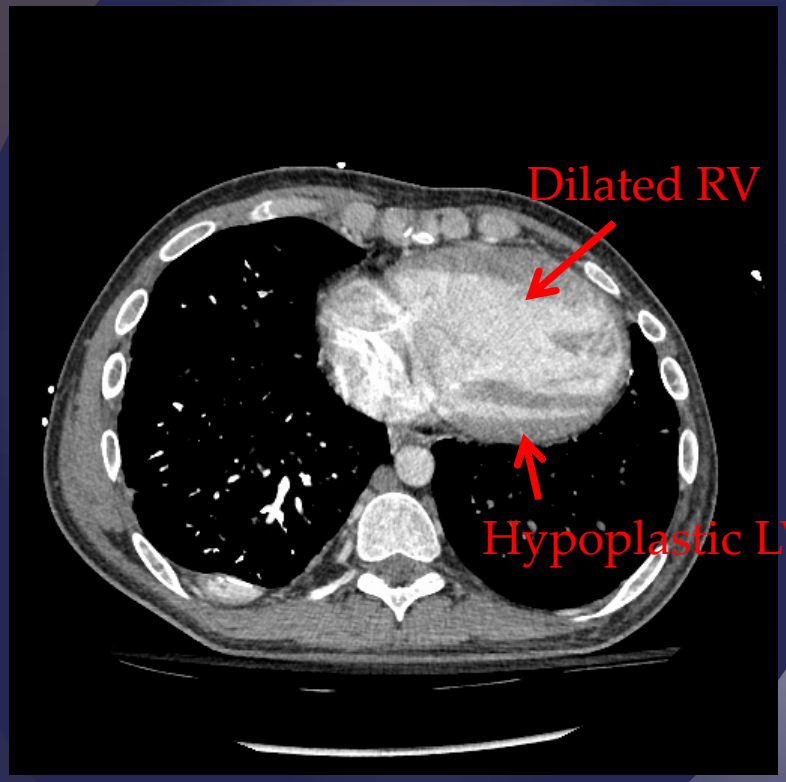
- Previous care all performed in the UK
- Remote left BT shunt (left SCA to LPA. Occluded? Not seen on CT or angio)
- Remote right Glenn shunt (SVC to right PA)

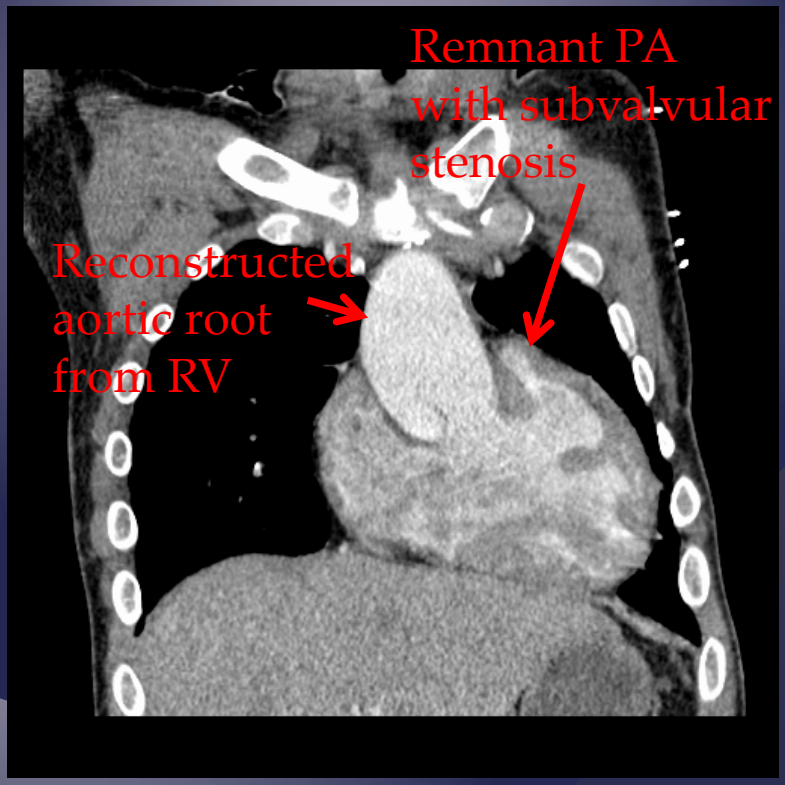


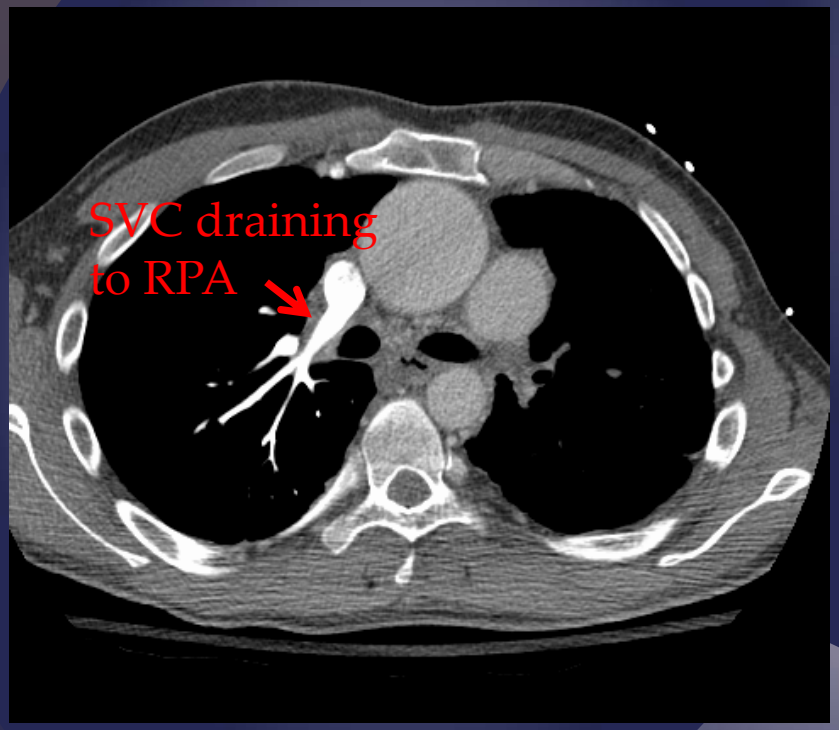
Hemoptysis History

- Initiated in 2007 with no episodes from 2012-2016
- Episodes now of up to 700 cc once or twice daily
- Multiple tranexamic acid and transfusion requirements for target Hgb > 180
- Baseline saturations in low 70's

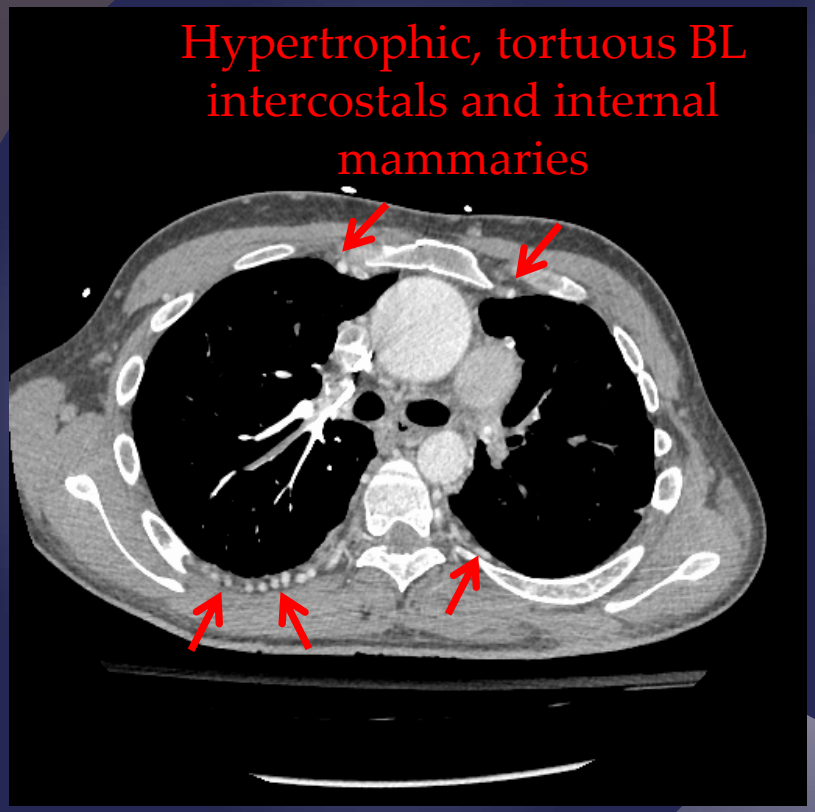
CTPA on admission

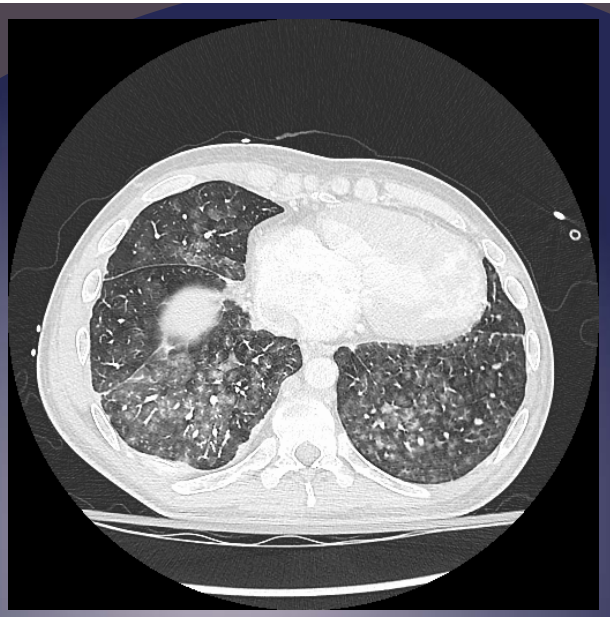


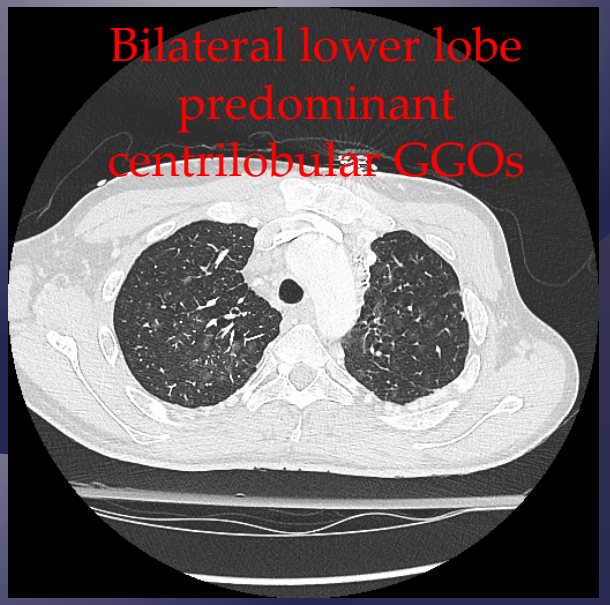




Single ventricle system
Severely hypoplastic left pulmonary arterial tree
No BT shunt identified


Hypertrophic, tortuous BL intercostals and internal mammaries



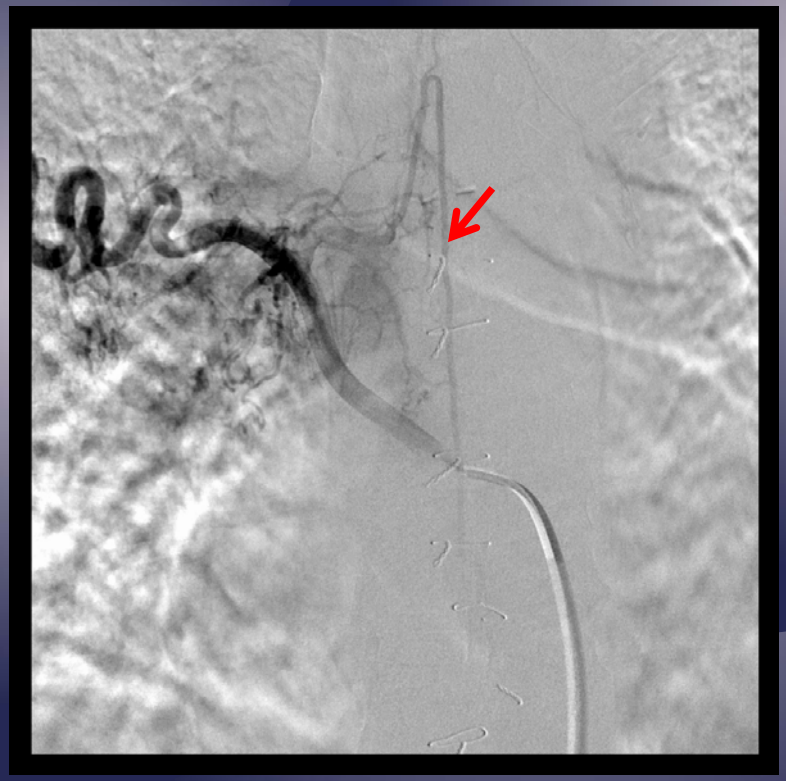

Bilateral lower lobe predominant centrilobular GGOs

Initial Angiogram

Initial Embolization Attempt

Initial Embolization Attempt

Initial Embolization Attempt



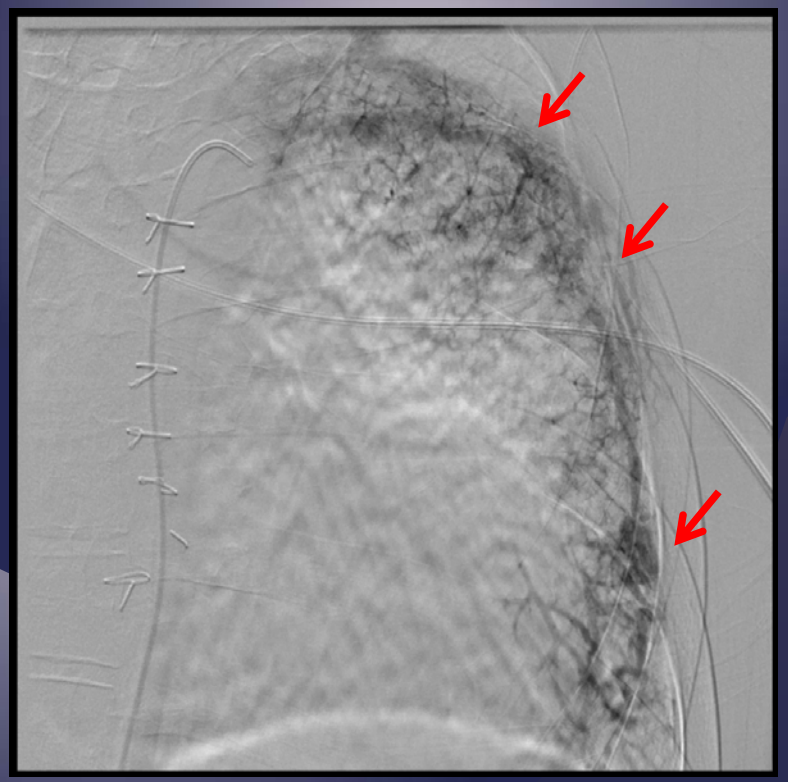
Initial Embolization Attempt

- No embolization performed
- Unable to gain access to right bronchial artery do to tiny tortuous collaterals arising from many intercostals

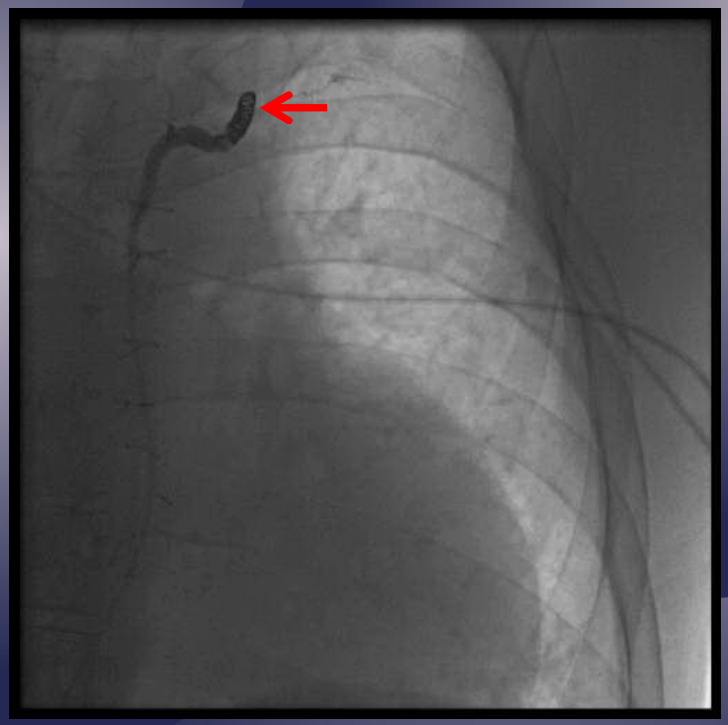
Clinical Course

- Recurrent Hemoptysis at one week
- Bronchoscopy/CT suggest diffuse bilateral alveolar hemorrhage

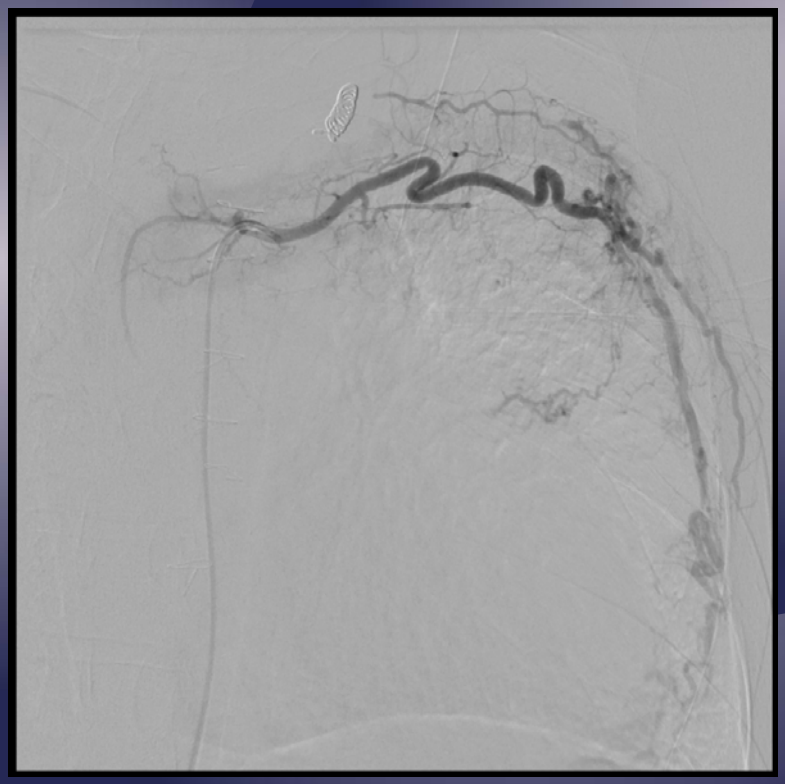
Second Embolization Attempt



Second Embolization Attempt



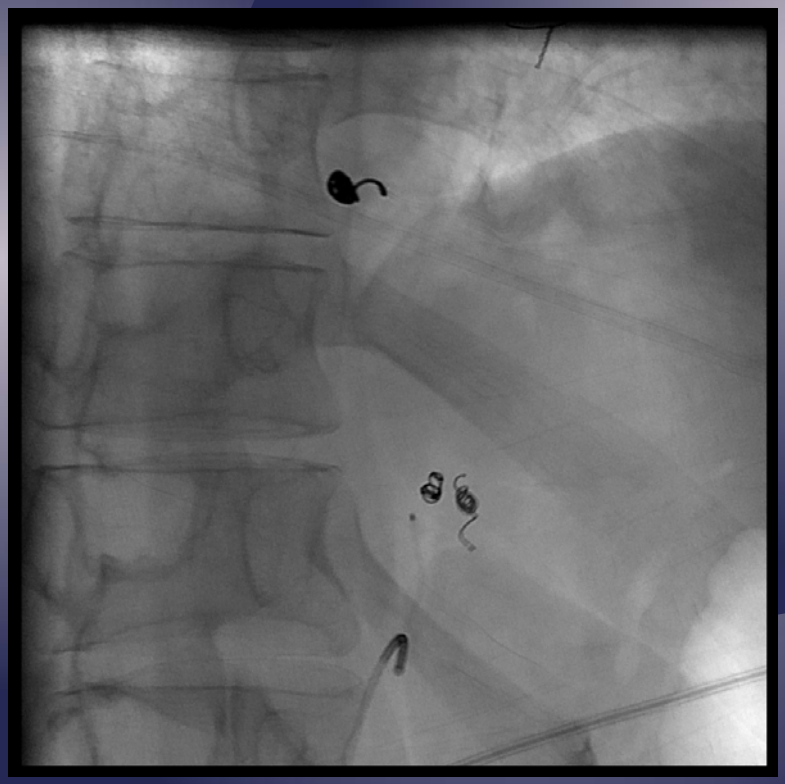
Second Embolization Attempt



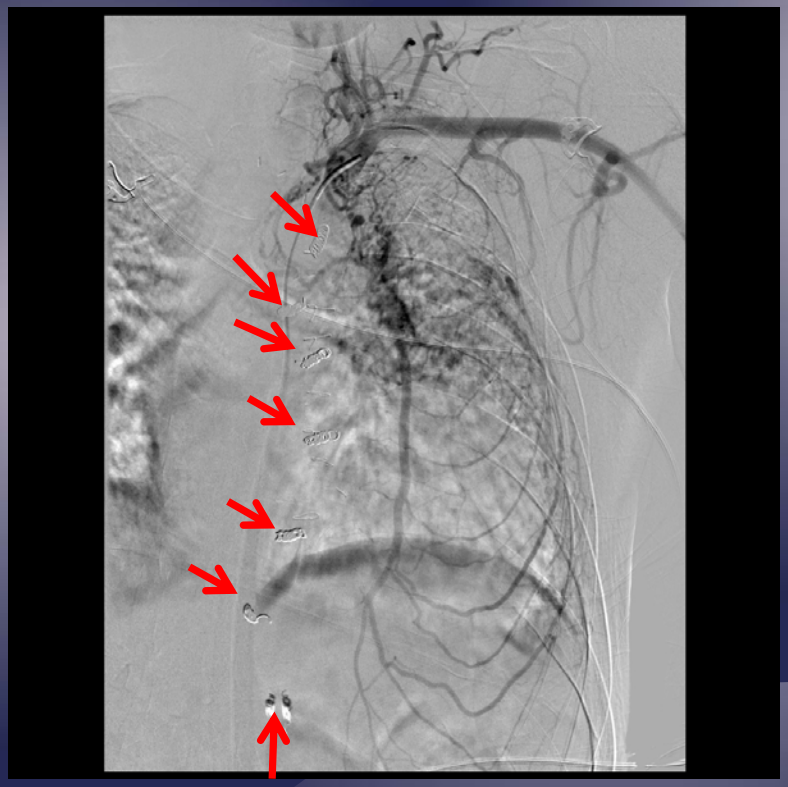
Second Embolization Attempt

Second Embolization Attempt

Second Embolization Attempt



Second Embolization Attempt



Second Embolization Attempt

- PVA embolization (700-1000 um) of left bronchial
- Coiled 6 proximal intercostals
- Coiled hypertrophied left gastric branches
- LIMA/thyrocervical trunk collaterals with moderate upper lobe parenchymal blush
  - Collateralizing intercostals
  - Not embolized

Considerations at this point

- Again unable to obtain access to main right bronchial artery
- ? worth embolizing multiple right intercostals
- Bronchial artery collateralized from many levels and likely to recruit more
- ? Risk to pulmonary function as bronchials/intercostals predominantly responsible for gas exchange in this patient

Ongoing Clinical Course

- Significantly decreased volume during hemoptysis (200-300cc) but continued daily occurrence with desaturations to 20's during episodes

Thoracic Covered Graft Placement?

- Discussed at rounds and with patient as option
- Nothing found in literature for cases of refractory hemoptysis
- Patient initially declined due to stated risk of paralysis and cardiovascular collapse due to occlusion of collaterals
- Recurrent large volume hemoptysis occurred requiring transfer to ICU
- Patient decided to proceed with thoracic endograft

Thoracic Covered Graft Placement

- Distal graft deployed first
- Immediate transient episode of hypotension
- Responded to pressor treatment after 5-10 minutes
- Proximal graft deployed without incident

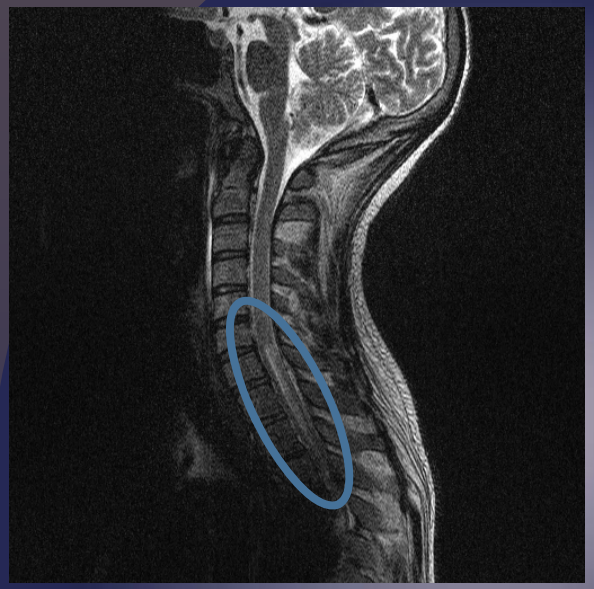
Post Operative Course

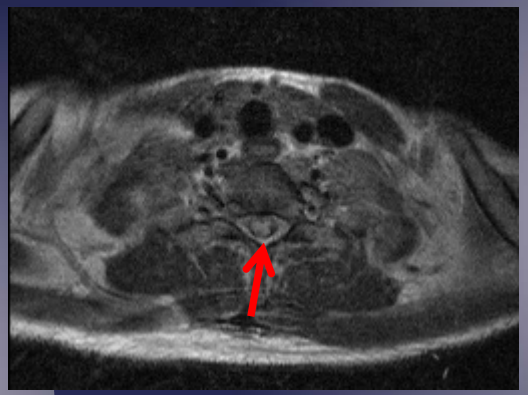
- 4 hours post op in ICU had significant cardiovascular decompensation with SPB of 50 for 30 mins
- Associated loss of movement in legs and decreasing LOC
- Lumbar drain inserted

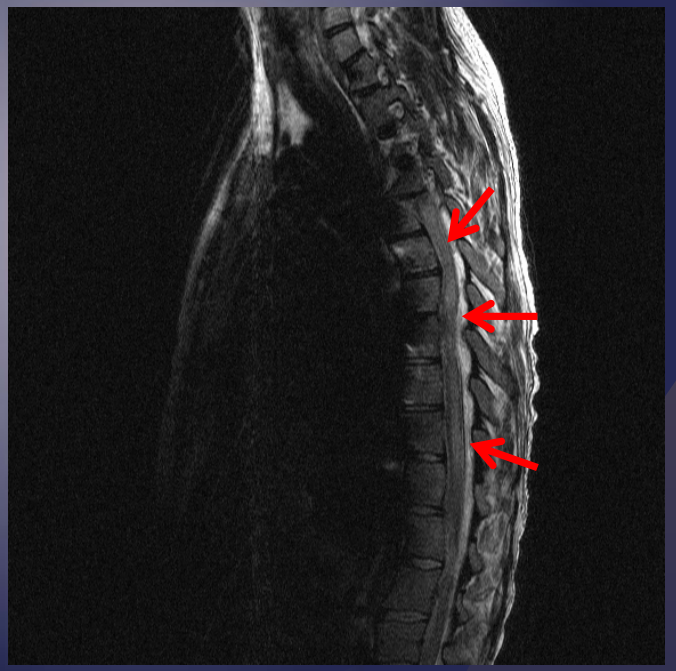
Post Operative Course

- Neuro exam (POD #4): Upper Limbs: normal
- Lower limbs: Power 0/5, absent pin prick below T4/T5
- Downgoing Babinski and brisk LL reflexes
- Abdominal and anal reflexes absent
- Urinary catheter remained in situ and had required manual fecal disimpaction

MRI Spine







Central T2 hyperintensity from C7-T11
? Cord ischemia related to prolonged hypotension

Long term follow up

- Large bilateral DVT's post op
- Rivaroxiban anticoagulation
- No further hemoptysis
- Readmitted from rehab with pneumonia
- PEA arrest NYD and passed away 11 months post op

TEVAR related Spinal Cord Ischemia

- Overall incidence: 3-4%
- Increased risk with length of graft/number of grafts deployed/coverage of left SCA
- Previous AAA repair increases risk to 12-14%
- SC perfusion pressure =

  spinal collateral network pressure – CSF pressure

TEVAR related Spinal Cord Ischemia- Preventative Measures

- Augmentation of collateral network pressure
  - Aggressive management of peri and post op hypotension
- CSF drainage
  - No consensus regarding routine use

Points to discuss

- Transplant
- More aggressive initial embolization?
- Staged grafting?
- Prophylactic lumbar drain? (High risk for SCI due to baseline hypoxia)
- Steroids for spinal cord protection

References

Oh THT, Wang TKM, Ramming J, Ramanathan T. First elective thoracic endovascular aortic repair to treat hemoptysis due to bronchiectasis. *Am J Respir Crit Care Med*. 2013;188(4):517-519. doi:10.1164/rccm.201210-1823CR.

Sidhu M, Wieseler K, Burdick TR, Shaw DWW. Bronchial Artery Embolization for Hemoptysis. *Semin Intervent Radiol*. 2008;25(3):310-318. doi:10.1055/s-0028-1085931.

Tom LM, Palevsky HI, Holsclaw DS, et al. Recurrent bleeding, survival, and longitudinal pulmonary function following bronchial artery embolization for hemoptysis in a U.S. adult population. *J Vasc Interv Radiol*. 2015;26(12):1806-1813. doi:10.1016/j.jvir.2015.08.019.