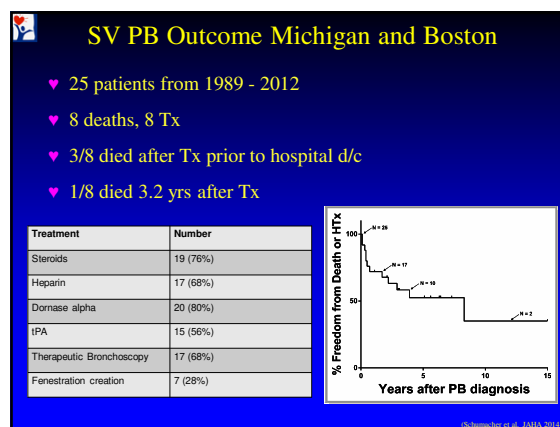


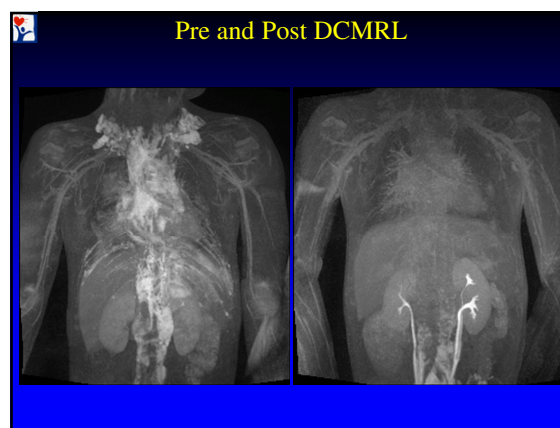
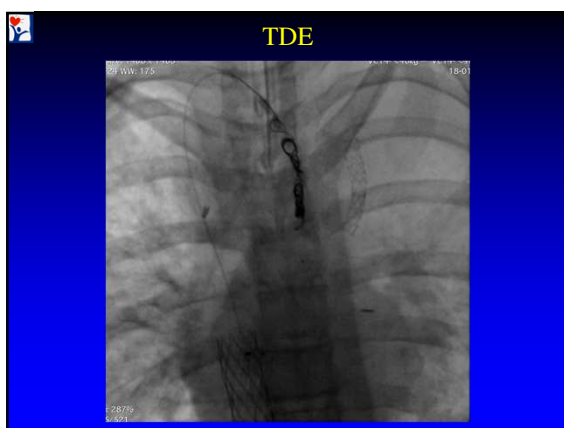
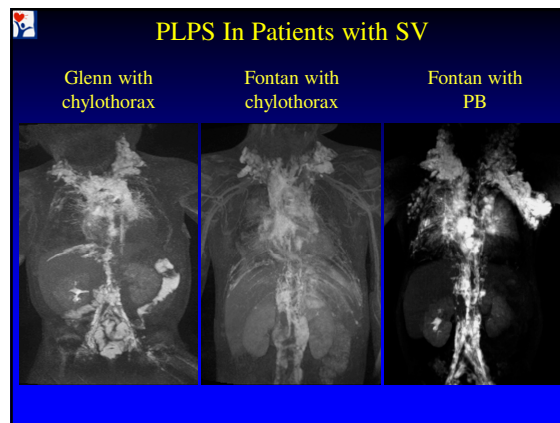
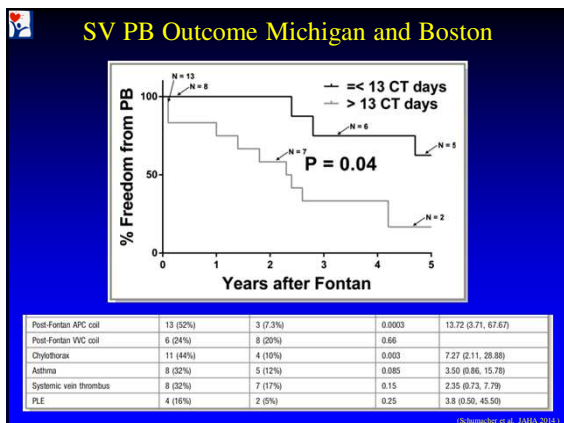
PB Treatment and Outcome CHOP Last 10 Years

Table 2 Treatment strategies and current symptoms

ID	Medical management	Catheter-based interventions	Surgical interventions	Heart transplant	Current status
1	Aspirin, statins, inhaled tPA		Prosthetic mitral valve replacement and resection of sub-A2	Y	Resolution of symptoms off tPA for > 3 years post-OHT; recent recurrence of casts in the setting of recurrent rejection and elevated CrP
2	Aspirin, statins, inhaled tPA	Balloon dilation of fenestration		Y	Asymptomatic; off tPA for 3 years since OHT
3	Aspirin, statins, inhaled tPA, carvedilol	Creation and stenting of fenestration		Y	Asymptomatic; off tPA for 2 years since OHT
4	Aspirin, statins, inhaled tPA, carvedilol	Creation and balloon dilation of fenestration, tPA stenting		N	Asymptomatic for 1.5 years; never required tPA
5	Aspirin, statins, inhaled tPA	Balloon dilation of LSVG and RPA stents		N	Recurrence of casts, ventricular dysfunction and PLE; continues on outpatient tPA
6	Aspirin, statins, inhaled tPA	Balloon dilation and stenting of fenestration, RPA dilation		N	Only small casts since fenestration stented; continues on outpatient tPA
7	Aspirin, statins, inhaled tPA		Resection of small septum and unroofing of conus above	N	Asymptomatic for > 2.5 years since surgical procedure; tPA weaned off
8	Aspirin, statins, inhaled tPA			N	Rare, episodic casts in the setting of viral infections; tPA weaned off
9	Aspirin, statins, inhaled tPA, bosentan	Balloon dilation and stenting of fenestration, dilation of LTV stent, embolization of APCs		N	Extremely rare casts; marked decrease in cast burden after addition of bosentan; continues on outpatient tPA
10	Aspirin, statins, inhaled tPA, carvedilol	Stenting of VVC and fenestration	Foram revision and patch augmentation of VVC	N	Asymptomatic for 3 months since foram revision; continues on outpatient tPA
11	Aspirin, statins, inhaled tPA	Coil embolization of VVC; bosentan; glue embolization of TD lymphatic collaterals		N	No casts with resolution of chronic cough after lymphatic intervention; tPA weaned off
12	Aspirin, statins, inhaled tPA, bosentan, carvedilol	Arterio-venous shunt strategy to take down to Glenn physiology		N	Very small rare casts; marked decrease in cast burden after addition of bosentan; continues on outpatient tPA
13	Chronic oral steroids, aspirin, statins, inhaled tPA, bosentan, carvedilol, digoxin			N	Rare casts; marked decrease in cast burden since addition of bosentan and carvedilol; continues on outpatient tPA
14	Aspirin, statins, inhaled tPA			N	Fewer casts since recently starting outpatient tPA

(Arbuckle et al. Ann. NY Acad. Sci. 2014)





- ### PB Outcomes
- ♥ Total 96 patients with PB seen
 - ♥ 2/96 without lymphatic etiology
 - ♥ 20 patients with idiopathic PB
 - ♥ 98% procedural success – TD cannulation
 - ♥ ~ 95% of PB with CHD significant improvement of symptoms
 - ♥ ~ 80% of PB with CHD cessation of casting
 - ♥ Majority treated with TDE and are type II
 - ♥ 2 patients treated with stent graft
 - ♥ F/U 4.5 years

- ### PB Outcome
- ♥ 55 patients with Fontan physiology and PB
 - ♥ 10/55 (18%) had concurrent PLE
 - ♥ 7/55 (14%) required a repeat lymphatic intervention
 - Contraindication for MRI
 - Left sided paravertebral networks
 - ♥ 3/55 (6%) developed new PLE after PB intervention
 - ♥ 2/55 (4%) had resolution of PLE symptoms after procedure
 - ♥ 3/55 (6%) with liver as source

PB Complications

- ♥ Stroke (~3%)
 - 1 Fontan patient – Glue
 - 1 Glenn patient - Glue
 - 1 patient with TOF - Lipiodol
- ♥ SIRS post procedure
 - 25% in first 24 hours
- ♥ Pancreatitis - 3 patients
- ♥ Abdominal pain ~ 100%
- ♥ Bleeding - 1 patient 3 days after starting anti coagulation
- ♥ Death 2 patients
 - 1 patient due to hemoptysis
 - 1 patient due to aspiration and MOSF

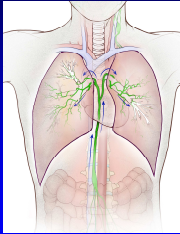
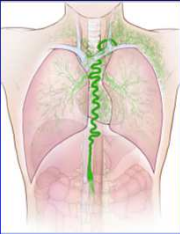

Stroke Prevention Measures

- ♥ Coagulation labs on admission
- ♥ FFP prior to procedure in case of deficits
- ♥ V-V collateral embolization or balloon occlusion
- ♥ Temporary fenestration occlusion
- ♥ Pre and post procedure head fluoroscopy
- ♥ Post procedure neuro exams
- ♥ Post procedure anticoagulation

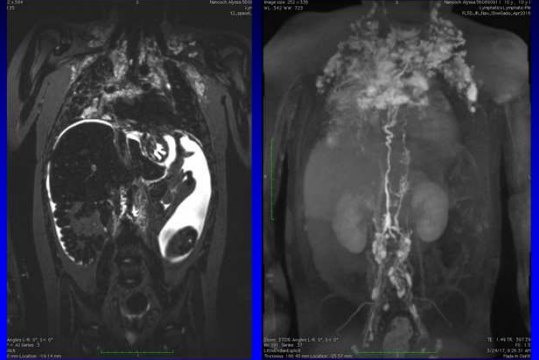
PB Cast and BAL Findings

- ♥ 55 Patients from March 2014 – May 2017
- ♥ 42 BAL analyzed for viruses by PCR:
 - Overall 16.7% positive
 - 4 for rhinovirus, 1 for both parainfluenza 3 and rhinovirus, 1 for adenovirus and 1 human metapneumovirus
- ♥ 54 BAL for Mycobacteria and Fungal
 - 0% mycobacteria
 - 9.3% fungal - either candida albicans or aspergillus fumigatus
- ♥ 56 BAL for bacterial Cx
 - 12.5% positive
 - H. Flu, M. Catarrhalis, S. Aureous, P. Aeruginosa
- ♥ 15 Casts for pathology
 - 14 mixed cellular
 - 1 Proteinaceous

PB Special Considerations

PB	PB + PLE or Ascites	Hepatopulmonary connections
		

10 yo TA s/p Fontan with Ascites and PB



Contrast Lymphangiogram and SLDE

