Case Presentation
Echo Boot Camp Board Review

Bobby Aertker, MD
THI Cardiology Fellow
Case #1

- 37 y.o. F with increasing symptoms of dyspnea on exertion and exercise intolerance. Occasional LE edema.
- PMHx: “Exercise induced asthma”
- She presented to an ER in her home town.
- Cardiac exam: irregularly irregular, tachycardic, HR 150’s
- EKG revealed evidence of atrial fibrillation with rapid ventricular response.
She was admitted to the hospital.

TTE at the OSH:
  - EF 23%.
  - LA and LV were dilated
  - Evidence of an indeterminate left atrial abnormality.

She was then referred to Houston for further cardiology workup.

She was discharged on sotalol, ramipril, and rivaroxaban.
- Her medication regimen was adjusted to metoprolol, losartan, digoxin, and rivaroxaban. She had adequate rate control with these meds.
- An echo was performed in clinic which showed the following...
LVEF 30%
A cardiac MRI was also performed.
• She was then referred to CV surgery for resection.
• Rate control regimen was uptitrated to good effect
• In the OR...
• Intra-op TEE...
3D TEE Images
• Uneventful post-operative course.
• Her atrial fibrillation completely resolved after resection of the cor triatriatum + Maze surgery.
• Her EF improved to ~47% within 2 months of post op follow up.
2 months post op echo
EDV was 182 ml on pre-op echo
• Now training for a triathlon.
• No further episodes of “asthma exacerbations.”
Cor triatriatum

- Numerous anatomic variations exist
  - Cor triatriatum sinistrum – left atrium divided into two chambers
  - Cor triatriatum dextrum – right atrium divided into two chambers (extremely uncommon)
- Rare
  - Accounts for ~0.1% of all congenital heart defects
Cor triatriatum

- Differing theories on embryogenesis but it may be the result of failure to incorporate the embryonic common pulmonary vein into the left atrium
- Can be isolated or seen in association with other defects
  - Persistent left SVC with unroofed coronary sinus, ASD, and PAPVR have been reported
Cor triatriatum physiology

- When the membrane is obstructive, venous flow is impeded → pulmonary edema and/or intraalveolar hemorrhage
  - Can affect both lungs or only one lung
- Most patients will manifest clinical symptoms in the first few years of life, but rarely, patients may present in adulthood
- Right heart enlargement and pulmonary hypertension may develop
- “Classic cath” will reveal pulmonary hypertension, elevated PCWP, and normal left atrial pressure
Treatment

- Surgical resection is indicated in patients with elevated PA pressures and/or symptoms related to the membrane.

- Typically, adults who present with previously undiagnosed cor triatriatum will not have obstructive gradients across the membrane.
Case #2

- 49 y.o. F with DM2 and a h/o cardiac surgery 7 years prior, presented with intermittent chest pain, fatigue, DOE, and presyncope. Symptoms occur at rest but are worsened with exertion.

- Several episodes of pre-syncope while driving.
• She underwent TTE but due to body habitus, the imaging windows were extremely limited.

• Given the severity of her symptoms, it was decided to proceed with LHC/RHC and TEE.
LHC

- LV 222/19-21
- Aorta 143/82, mean 109
- No obstructive CAD
- LV angiogram: Hyperdynamic LV with near complete obliteration of the cavity and narrowing visualized in the LVOT
RHC

- RA 12/9/11 (A/V/mean)
- RV 55/4-18
- PA 42/22, mean 33
- PCWP 23
Subvalvar aortic stenosis

- Can be due to fixed obstruction in the setting of subaortic stenosis or may be dynamic in the setting of hypertrophic obstructive cardiomyopathy.
- CW doppler interrogation through the LVOT:

Subaortic stenosis

- Anatomy can vary
  - Membrane
  - Fibromuscular ridge
  - Tunnel like obstruction
- Felt to be progressive with age
- Many develop associated aortic insufficiency due to high velocity jet damaging the aortic valve
Subaortic stenosis management

- Indications for surgical resection include degree of obstruction as measured by the gradient, degree of aortic insufficiency, and symptoms related to the obstruction.
- 20-30% can have recurrence after resection, with up to 20% of patients requiring reoperation.
Case #3

- 53 y.o. M with symptomatic severe aortic stenosis.
- Diagnosis of bicuspid aortic valve made in childhood
- Most recent echo:
  - AVA 1.0 cm²
  - AV peak gradient 98mm Hg, mean gradient 52 mm Hg
- Now becoming fatigued after strenuous exercise
TTE
• Referred for surgical aortic valve replacement
• Intra-op TEE showed...
Unicuspid aortic valve

- Rarely diagnosed pre-operatively, more commonly diagnosed after surgical resection or during autopsies
- Most patients will present with aortic stenosis, however more than \( \frac{1}{4} \) of patients will also have aortic insufficiency
- Associated anomalies
  - Aortic root dilation and dissection
  - Coronary anomalies
  - PDA
  - Coarctation
- Mean age of presentation for AVR is younger for unicuspid aortic valves compared to both bi- and tri-cuspid aortic valves
Figure 1: Schematic representations of a unicommissural and an acommissural unicuspid aortic valve.
Case #4

- 57 y.o. M with a history of hyperlipidemia, GERD, and atrial fibrillation was seen by his cardiologist in Florida ~2 years ago.
- Echo at that time reportedly showed evidence of a secundum ASD and percutaneous closure was recommended.
• He then moved to Colorado and obtained a second opinion.
• Repeat imaging was performed and per the patient he was diagnosed with an ASD vs PFO.
• Closure was again recommended.
• Shortly thereafter he moved to Houston and established care with cardiology here.
• A TEE was then performed at our institution.
Echo images
Name the abnormality

- A. Secundum ASD
- B. Primum ASD
- C. Tunnel type PFO
- D. Sinus venosus ASD
- E. Other
Bubble study
Bubble study with valsalva
• A. Secundum ASD
• B. Primum ASD
• C. Tunnel type PFO
• D. Sinus venosus ASD
• E. Chiari network
• F. Eustachian valve
Right atrial variants

- The right sinus of venosus valve functions to direct embryonic oxygenated venous blood flow from the IVC into the foramen ovale and ultimately into the left atrium.
- This valve involutes during development.
Remnants of the right sinus venosus valve

- Incomplete resorption can result in a spectrum of anatomic variants
  - Eustachian valve
  - Thebesian valve
  - Cor triatriatum dexter
  - Chiari network
- Eustachian valve
  - Seen adjacent to the IVC
  - Remnant of the inferior portion of the right sinus venosus valve
  - Prevalence >50%
  - Size varies widely
  - Leaf like appearance and relatively immobile
  - Mean length ~1 cm
• Chiari network
  • Thin, weblike, reticulated group of fibers that originate from the Eustachian or Thebesian valve
  • It is the result of incomplete resorption of the right sinus venosus valve and septum spurium
  • Prevalence 2-15%
  • Mean length ~3 cm (generally larger than EV)
  • Highly mobile, whiplike movement
Cor triatriatum dexter

- Persistence of the right sinus venosus valve that divides the right atrium into two chambers
- Extremely rare (<0.025% of all congenital heart disease)
- Inserts onto the crista terminalis
• Thebesian valve
  • Seen at the orifice of the coronary sinus
  • Also a remnant of the inferior portion of the right sinus venosus valve
  • This valve covers <50% of the CS orifice in >60% of cases
  • Covers CS completely in 5% of cases
  • Rarely visualized by echo
Figure 2: Graphic showing anatomic variants of the right sinus valve remnants. (A) CT TV: persistent right sinus valve divides the right atrium, producing flow obstruction and directing a significant part of venous return through the PFO. (B) ON: reticulated network of fibers originating from the EV and connecting with different parts of the atrium. (C) Prominent EV: large membrane attached to the lower part of the atrial septum partially dividing the right atrium, but without flow obstruction. (D) Persistence of EV and TV as small rim-like vestigial (the most common morphology observed). SVC, Superior vena cava; TV, tricuspid valve.

(J Am Soc Echocardiogr 2016;29:183-94.)
Case #5

- 53 y.o. male with a history of CAD s/p STEMI and PCI to LAD, COPD, and metastatic cancer of unknown primary that was diagnosed by pericardial biopsy ~8 months ago.
- TTE images at time of initial diagnosis.
Baseline TTE performed 8 months prior to current presentation
• He then suffered a cardioembolic stroke.
• An echo was performed which showed new, severe mitral regurgitation and a TEE was requested for further evaluation of the mitral valve.
TEE images for new onset severe MR
Non-bacterial thrombotic endocarditis (NBTE)

• AKA marantic endocarditis

• When thrombi are large, some also use the terms verrucous endocarditis and Limans Sacks endocarditis

• Rare condition that is more commonly diagnosed via autopsy
• Variety of diseases that result in non-infectious lesions on the heart valves
  • Cancer (80%)
  • Lupus
  • Anti-phospholipid antibody syndrome
• Felt to be the result of endothelial damage in a patient with a prothrombotic state
• Sterile, platelet thrombi aggregate on heart valves
  • Aortic and mitral valves are more commonly involved
• Compared to infectious endocarditis, the vegetations in NBTE are much more easily dislodged resulting in thromboembolic events
Treatment consists of:

- Systemic anticoagulation
- Treatment of the underlying disease (cancer, etc)
- Surgery?
Thanks!