Interesting Case Studies

Judith Buckland, MBA, RDCS, FASE

CARDIOSERV
INSPIRING EXCELLENCE IN IMAGING

Interesting Case Studies

1. Dueling Myxomas
2. RV Thrombus in Transit
3. Double Chambered LV

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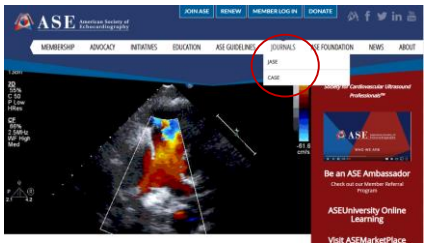
American Society of Echocardiography



Dueling Myxoma

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Outline

- Signs & Symptoms
- History
- CT / CTA
- Echo (TTE)
- Echo (TEE)
- Treatment
- Outcome

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Patient History

- 69-year-old man
- Hx ischemic CMO
- Afib (anticoagulated)
- CAD, PCI/prox LAD /2 years prior
- EF 25% - 30%
- Single-lead ICD
- Hyperlipidemia, DM, and distant history of pulmonary embolism.

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Signs and Symptoms

Presents with:

- Aphasia
- Right-sided weakness
- Right facial droop
- BP 130/90 mmHg
- Irregular rhythm with soft systolic murmur
- ECG - ventricular paced rhythm with underlying Afib

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Head CT / CTA

- Head CT - no pathology
- CTA - Acute occlusion - M2 branch of middle cerebral artery

Diagnosis: Acute stroke

Cause: Known Afib with patient non-compliance of anti-coagulation meds

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Treatment

- Not a candidate for tPA - hx anticoagulation
- Successful intra-arterial pharmacologic thrombolysis
  - **Intra-arterial thrombolysis** uses an endovascular microcatheter delivery system.
  - Catheter port is positioned immediately *within and adjacent to the thrombus*, and fibrinolytic agents are infused *directly into the clot*.
- Echo ordered

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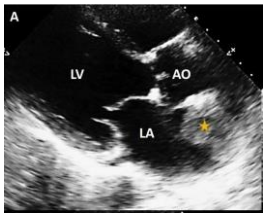
Echocardiogram (TTE)

- Chronic severely ↓ LVF
- Mildly ↓ RV function
- Mild - moderate TR
- LA mass (3.7-cm)
- Suggestive of myxoma



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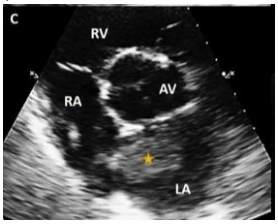
Echocardiogram (TTE)

- PSAX
- LA: Smooth echogenic mass



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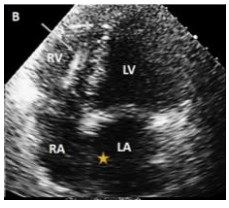
Echocardiogram (TTE)

- A4C– mass poorly visualized
- ICD wire seen in the RV



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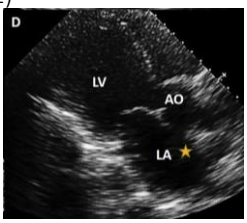
Echocardiogram (TTE)

- 3-chamber view
- Poorly visualized left atrial mass



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Next Step

- TLS – No definitive diagnosis of Myxoma
- Thrombus vs. tumor??
- Cardiac MRI contraindicated due to implanted defibrillator
- TEE was ordered

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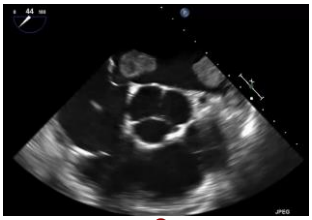
TEE



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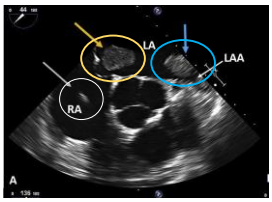
2D TEE Findings

- Midesophageal short axis view
- Two smooth mobile masses consistent with myxomas
  1. Attached to the IAS *
  2. Attached overlying the orifice of the left atrial appendage (LAA) *
- Defibrillator wire in the right atrium



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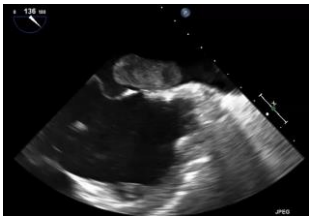
2D TEE



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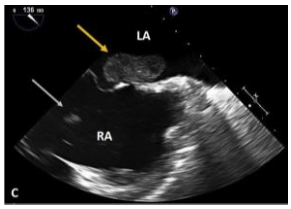
2D TEE Findings

- TEE Bicaval view
- Myxoma with typical insertion
- IAS - left atrial side
- Defibrillator wire in RA



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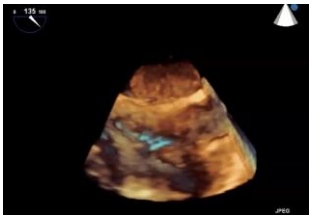
3D TEE



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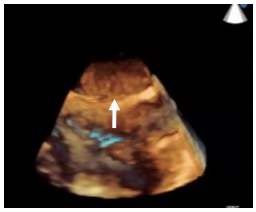
3D TEE Findings

- 3D TEE
- Myxoma attached to the IAS
- Visualization of the stalk.



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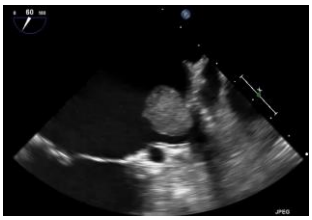
2D TEE



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2D TEE Finding

- Midesophageal view
- Smooth mass
- Perdunculated - characteristic stalk of a myxoma
- Attached near the left atrial appendage (LAA)
- Attachment point clearly identified
- Video: Rocking at attachment point above the left atrial appendage

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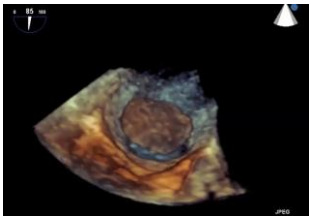
3D Imaging Performed



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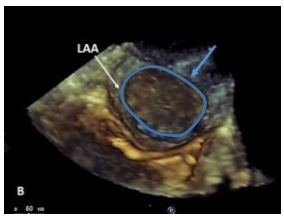
3D TEE Findings

- Myxoma overlying the opening of the LAA
- Smooth borders consistent with myxoma



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Treatment Plan

Patient was evaluated by cardiothoracic surgery due to:

- Distinctive characteristic findings of myxoma on TEE
- Location and embolic potential of the dual myxomas

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Surgery

- Successful operation
- Histopathology:
  - soft, tan-pink cut surface
  - cystic-appearing mass
- Post-op uneventful
- Successful discharge to a skilled nursing facility

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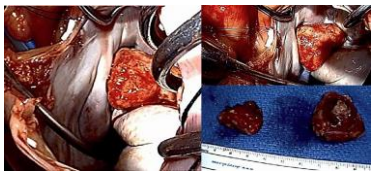
Myxoma – Surgical Images

- Intraoperative gross images
- Gross visualization of the two myxomas after surgical resection



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Post-Op Echo

- Post-op echo demonstrated successful resection of the myxomas
- EF of 25% - 30% (previously reported)
- 6 month f/u echo
  - No recurrence of left atrial masses
  - EF 25% - 30%

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Case Study Review

- Patient presented with signs of a stroke
- Echo & TEE - key diagnostic test in the management of stroke
- MRI was contraindicated due to the patient's implanted defibrillator
- Echo was critical for diagnosis and plan of care

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Case Study Review

- Initially, assumed the patient's acute stroke was due to non-compliance of anticoagulation meds
  - Afib
  - Elevated risk scores
- TEE revealed a cardiac mass - not one but two left atrial myxomas!

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Discussion: Myxoma

- Primary cardiac tumors are rare
- Myxomas account for almost half
- Myxomas are typically:
  - Pedunculated
  - Located in the LA
  - Vary in size
  - Usually only one tumor

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Discussion: Myxoma

- Clinical manifestations include cardioembolic events
- Other symptoms include:
  - Weight loss, fever, laboratory abnormalities such as anemia and elevated C-reactive protein
- Seen more in women

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Discussion: Myxoma

- Recommendation treatment - Surgical resection due to the risk of embolization
  - TEE (especially 3D) allows precise localization of the attachment point of these tumors, allowing improved operative planning
- Recurrence of tumor is rare
- The presence of two primary cardiac myxomas is a rare finding, most often seen in patients with Carney Complex

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Discussion: Carney Complex

- Rare endocrine disorder
- Characterized by multiple benign tumors, such as myxomas
- Spotty skin pigmentation
- Usually young men
- This patient (69M) did not have pigmented skin lesions, nor did he have evidence of any other endocrine tumors to suggest this diagnosis

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Conclusion

- Myxomas are the most common primary tumors of the heart, most commonly located in the left atrium
- The discovery of multiple myxomas is a rare finding
- This case study demonstrates **two left atrial** myxomas diagnosed on transesophageal echocardiography (TEE)

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End Case One

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RV Thrombus in Transit

Pulmonary Embolism

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Outline

- Signs & Symptoms
- History
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- Echo (TEE)
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- Outcome

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Signs and Symptoms

- 40F
- 3 wks progressive dyspnea on exertion
- Normal cardiopulmonary exam in ER
- No signs of volume overload
- Hemodynamically stable but *hypoxic*

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History

- HTN
- Obese
- Type 2 diabetes
- Unprovoked DVT (no known cause) and pulmonary emboli being treated with blood thinners
- Untreated pelvic mass of unknown etiology

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Chest CTA

- *Non-occlusive thrombi* in various branches of the right and left pulmonary arteries
- *Chronic* emboli with interval improvement
- She was started on heparin and 4L supplemental oxygen
- Successful reversal of hypoxemia

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Echocardiogram (TTE)

- Transthoracic echocardiography (TTE):
  - Mildly reduced RV function
  - RV fractional area change of 25%
  - TAPSE of 1.7 cm
  - New large RV mass suspicious for thrombus

TEE was ordered

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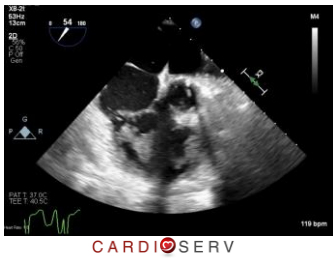
TEE: RV Inflow-Outflow View



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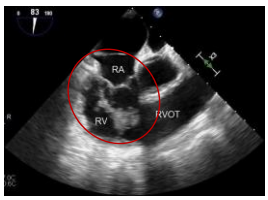
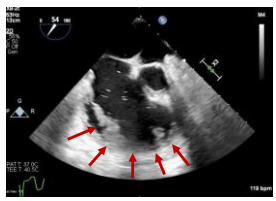
TEE: RV Inflow-Outflow View





Large RV multi-lobed thrombus

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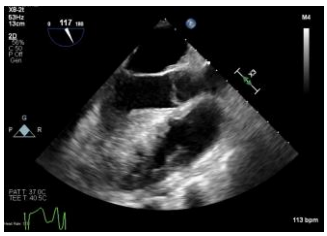
TEE: Long Axis View



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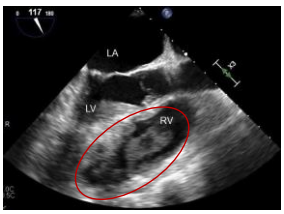
TEE: Long Axis View

- Large RV thrombus
- Multi-lobed
- Extending from TV to PV



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3D TEE

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3D TEE

- Large RV thrombus

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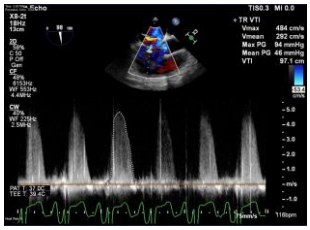
TEE: TR

- TR Moderate-Severe
- Peak pressure gradient 94 mmHg
- Mean pressure gradient 46 mmHg



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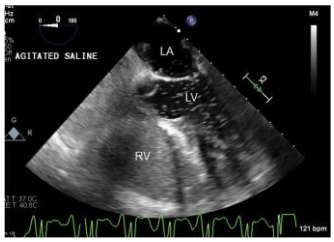
TEE: PFO

- 4 Chamber view
- Agitated saline
- PFO



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TEE: Other Findings

- There was no evidence of PV stenosis/regurgitation
- Estimated right atrial pressure of 8 mmHg
- RVSP 102 mmHg
- Mean PA pressure 54 mmHg

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Other Testing

- Lower Extremity Ultrasound
  - Chronic occlusive thrombi in various areas of the right lower leg
- Brain MRI
  - Multiple tiny, acute infarcts of the:
    - Bilateral cerebellar hemispheres
    - Bilateral parietal lobes
    - Left frontal lobe
- Neurological Exam
  - Normal neurological exam

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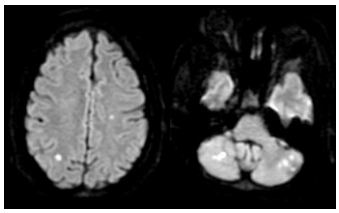
Brain MRI

- Multiple tiny, acute emboli
- Bilateral cerebellar hemispheres
- Parietal lobes
- Left frontal lobe



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Other Findings

- 12-cm ovarian mass found 1 year before admission
- Patient did not follow-up
- CT now shows 15 × 10 × 11cm cystic and solid right ovarian mass
- ↑ Cancer antigen-125
- Placed on a new blood thinner plus meds to ↓ PHTN

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Treatment

- IVC filter was placed to prevent any further emboli
- Repeat echo 5 days later:
  - Slight decrease in RV thrombus size
  - Improvement in RVSP to 55 mmHg
  - Mean PA pressure to 44 mmHg

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Initial Treatment Plan

- Initially considered: Emergent thrombolysis, surgery, and ECMO (heart-lung bypass support)
- None of these options were pursued because:
  - Patient was currently hemodynamically stable
  - Patient was improving on anticoagulation alone
- Patient was discharged with anticoagulant medication and plans for out-patient surgery for pelvic mass

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Pelvis Surgery

- 45 days later – pelvis surgery without any immediate complications:
  - Tumor debulking
  - Total hysterectomy
  - Removal of ovaries and fallopian tubes
- Pathology revealed cancer (adenocarcinoma)

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Intraoperative TEE

- Intraoperative TEE, at time of surgery showed a stable RV thrombus
- RVSP of 47 mmHg
- Mean PA pressure of 33 mmHg

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Post Op

- On the day after surgery, the patient was extubated successfully but subsequently developed progressive hypoxemia
- Chest x-ray showed clear lungs
- Chest CTA showed new pulmonary emboli:
  - Right upper lobe
  - Right middle lobe
  - Left lower lobe arteries

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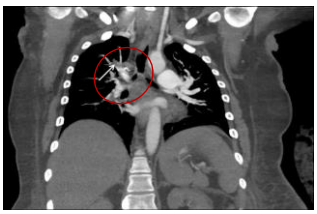
CTA

- New pulmonary emboli in the right upper lobe arteries (*arrow*), right middle lobe artery, and left lower lobe arteries



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Repeat TTE

- Repeat TTE showed worsening right heart pressures
- RVSP of 91 mmHg and mean PA pressure of 56 mmHg
- Patient was restarted on anticoagulation (it had been stopped for surgery)

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Treatment

- Decision to proceed with thrombectomy , based on:
  - Worsening pressures
  - Continued risk for pulmonary and systemic emboli
  - Potential hemodynamic compromise

Surgery (sternotomy)

- Remove RV thrombus
- Close PFO

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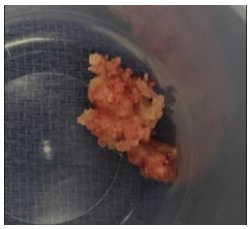
Surgical Finding

- RV mass- entwined in TV chordae
- Did not appear to be a thrombus
- Concerning for metastatic disease



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Post-op

- Pathology
  - Organizing thrombus without any malignant cells identified
- Postoperative TTE
  - No evidence of interatrial shunting
  - Showed improved RVSP 53 mmHg / mean PA pressure 41 mmHg
  - Attributed to debulking of the RV mass

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Post-op

- Post-op the patient remained hemodynamically stable and demonstrated good oxygen saturation on 3L of oxygen
- She was discharged with anticoagulants, sildenafil (↓ PHTN) and home oxygen
- After discharge:
  - Improved symptomatically
  - Remained clinically stable
  - Serial echo showed improving right heart pressures

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Follow-Up Echo

- 9 month F/U echo after thrombectomy
  - Patient was asymptomatic
  - No signs of recurring thrombus
  - TTE showed RVSP of 26 mmHg
  - Normal RV size
  - Mildly reduced RV function

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Discussion: Right Heart Thrombus

- Right heart thrombus
  - Uncommon condition (4%) of patients with pulmonary emboli
- It is often found in transit - originating from a systemic vein source
- May also form within the cardiac chambers - primary processes, such as Afib

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Discussion: Right Heart Thrombus

- Right heart thrombi are associated with significantly increased mortality
  - 27% - 45% with treatment
  - Near 100% in untreated patients
- About 98% of cases of right heart thrombi are associated with concurrent pulmonary emboli

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Discussion: Treatment

- The treatment options for RV thrombus include:
  - Anticoagulation
  - Thrombolysis
  - Surgical thrombectomy
- Overall, the existing literature has shown conflicting evidence, with no clear consensus on management recommendations
- Several studies have suggested a mortality benefit using a more aggressive approach with either thrombolysis or surgery

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Discussion: Therapeutic Dilemma

This case presented a particularly difficult and complex therapeutic dilemma:

- Severely elevated right heart pressures
- RV dysfunction
- Fragile clinical status
- High risk for hemodynamic instability
- PFO with resulting cerebral infarcts
- Pelvic malignancy

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Discussion: Therapeutic Dilemma

There was concern that either thrombolysis or surgery could cause the thrombus to dislodge, leading to hemodynamic compromise:

- Thrombolysis - could lead to hemorrhagic conversion of her strokes
- Immediate surgery - ***anticoagulation would have to be held*** in the setting of:
  - Significant pulmonary embolic burden
  - Large RV thrombus
  - Significant right heart strain

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Discussion: Therapeutic Dilemma

Initial Decision - anticoagulation alone:

- Potentially poor outcome of intervention
- Patient's initial improvement:
  - ↓ right heart pressures
  - ↓ thrombus size

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Discussion: Therapeutic Dilemma

Surgical decision:

- Patient developed recurrent acute pulmonary emboli
  - After anticoagulation was interrupted for pelvic surgery
- Patient had worsening right heart pressures
- Continued high risk for pulmonary and systemic emboli

Surgery:

- RV thrombus was surgically removed
- PFO was closed

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Case Summary

- Despite this successful outcome, the optimal treatment for right heart thrombi remains uncertain and warrants additional studies
- Until there is more definitive evidence, management decisions should be made on a case-by-case basis, with careful consideration of complicating factors

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End Case Two

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Double Chambered LV

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Outline

- Signs & Symptoms
- History
- ECG
- Echo (TTE)
- Echo (TEE)
- Angiogram
- Cardiac MRI
- Treatment/Outcome

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Signs and Symptoms

- 49M
- History of alcohol abuse
- Type 1 diabetes on insulin
- Sudden onset of CP & profound palpitations while exercising
- Called 911 – initial rhythm was monomorphic v-tach 280 beats/min
- Converted to sinus rhythm w/ bolus of amiodarone bolus

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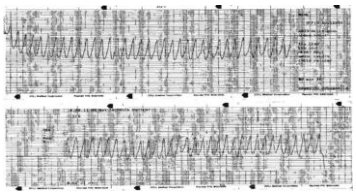
ECG in the field

- Monomorphic ventricular tachycardia at the time of presentation (EMS)



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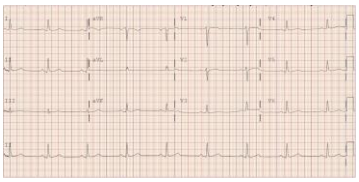
ECG

- Admission 12-lead ECG
- Normal sinus rhythm
- Normal limits, without ST-segment elevations or Q waves



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History

NO known history of:

- MI
- Cardiac trauma
- Illicit drug use
- Infection
- No family history of sudden cardiac death

Physical examination was unremarkable and subsequent normal ECGs

TTE Echo ordered

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TTE Echocardiogram

Transthoracic echo showed

- Unusual appearance to the LV
- Suspicious structure/attachment originating from LV apex
- Apical lateral wall - thinned & akinetic

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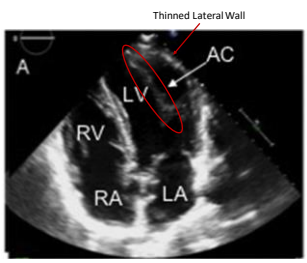
TTE Echo

- Unusual double-chambered appearance of the LV
- Accessory Chamber (AC)?
- Lateral wall - thinned and akinetic



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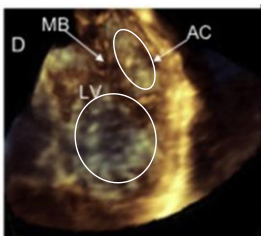
3D Echo

- 3D Echo
- Short Axis
- Double-chambered appearance of left ventricle



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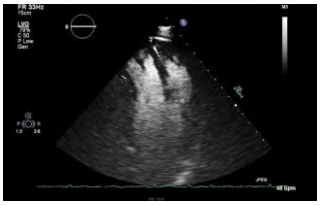
Echo Contrast



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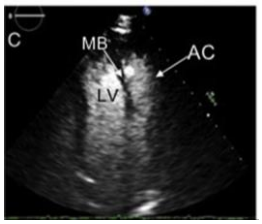
Echo Contrast

- Double-chambered LV
- Primary LV cavity w/ accessory chamber
- Subdivided by muscular ridge originating from apex
- Free unobstructed communication between the primary LV cavity & accessory chamber



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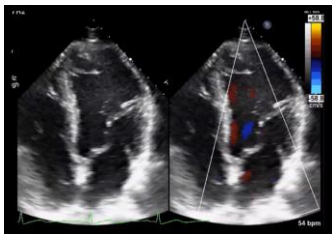
TTE ECHO Color



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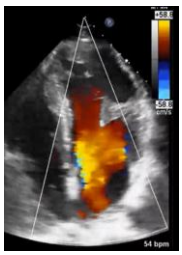
TTE Echo - Color

- Color demonstrates the absence of flow acceleration between the primary LV cavity and the accessory chamber
- Normally function MV without stenosis or regurgitation



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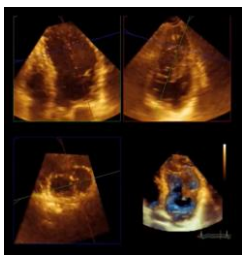
3D Echo



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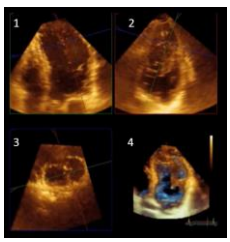
3D Echo

1. Dyskinetic lateral wall of the accessory chamber
2. Papillary muscles visualized
3. Cross sectional apex showing the subdivision of the LV
4. Color – no gradient between primary and accessory chamber



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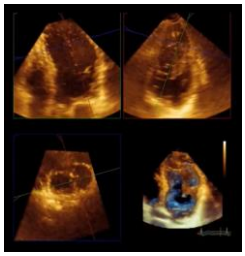
3D Echo



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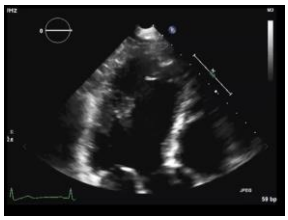
Echo – 3 Chamber



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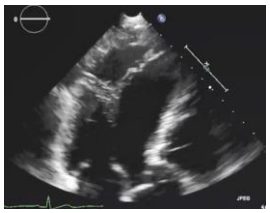
Echo 3-Chamber

- Subdivision of the left ventricle by an anomalous muscular ridge



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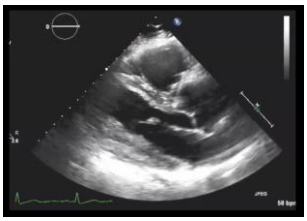
Parasternal Long Axis



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Parasternal Long Axis

- Structurally normal mitral valve.
- Contractility of the IVS and inferolateral walls were normal.

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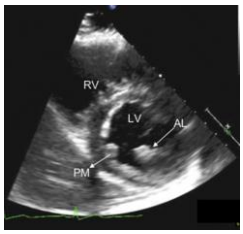
PSAX

- 2 distinct papillary muscles
- Anterolateral papillary muscle is dominant & larger than the posteromedial papillary muscle



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Papillary Muscle

- Distinct asymmetry of the anterolateral (AL) papillary muscle
- The AL papillary muscle was significantly larger than the posteromedial papillary muscle
- AL papillary muscle appeared to be attached to the questionable muscular ridge
- This anatomy likely falls within the wide spectrum of parachute-like mitral valve variants *without* the typical valvular dysfunction that is associated with a parachute mitral valve

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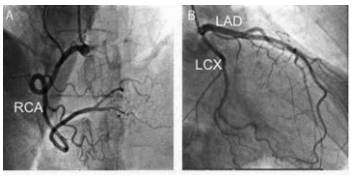
Angiogram

- Coronary angiogram did not demonstrate obstructive CAD



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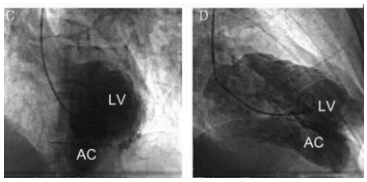
Angiogram

- Left ventriculography reveals a large LV outpouching lateral to the LV apex, consistent with the presence of an accessory chamber



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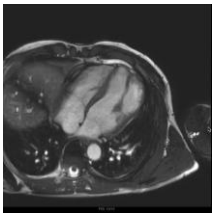
Cardiac MRI



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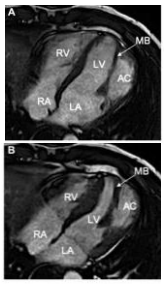
Cardiac MRI

- Presence of a large muscle band that subdivides the left ventricle into a primary and an accessory chamber. The wall of the accessory chamber is thin and akinetic
- EF 42% - supports congenital DCLV rather than an aneurysm or a pseudoaneurysm



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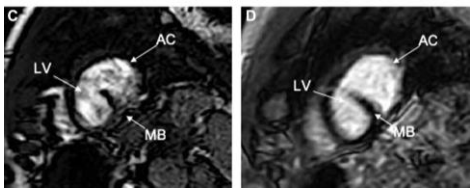
Cardiac MRI



No late gadolinium enhancement to suggest scar from prior ischemic injury

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TTE Findings

- LV cavity subdivided by a vertical tissue ridge in the center of the LV
- This suspected cavity has a similar thickness and appearance to myocardial tissue in both 2D and 3D
- There was no evidence of structural narrowing at the site of communication, rather a large visible gap by 2D & color Doppler
- Structurally normal MV with mild MR
- Lateral wall - thinned and akinetic
- No other wall motion abnormalities identified
- Global LVF mildly reduced, EF 45%

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Differential Diagnosis

Echo findings seemed consistent with DCLV, differential diagnosis of:

- Pseudoaneurysm at a prior silent myocardial rupture site
- True aneurysm
- Parachute Mitral Valve

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Differential Diagnosis: Pseudoaneurysm

- **Pseudoaneurysms**
  - Do not contain all three layers of cardiac tissue
  - Have a narrow neck
  - May exhibit paradoxical movement during systole
- ***DCLV contain all layers of cardiac tissue*** that typically contract synchronously with the rest of the ventricle

****Important** - Pseudoaneurysm typically requires urgent surgical management!

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Differential Diagnosis

Parachute mitral valve

- Parachute mitral valve can also have a similar appearance, but two distinct papillary muscles were identified that received chordae tendineae that appeared to be of equal lengths from both leaflets
- The mitral valve opened normally in a symmetric manner, without any stenosis and with minimal regurgitation

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Case Study Review

- This is the case of a man who presented with V-Tach diagnosed with Double Chamber Left Ventricle
- Echo and Cath suggested DCLV, but cardiac MRI was diagnostic

MRI Finding

- The absence of late uptake enhancement indicated a lack of a scar tissue which ruled out a prior silent MI
- No signs of fibrosis, ruled out trauma or infarction
- MRI showed thinned wall consisted of myocardial tissue, which excluded the possibility of a pseudoaneurysm

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Case Study Review

- Surgical resection was deferred given the close proximity of the defect to the papillary muscle and submitral apparatus
  - ↑ risk for complication that could lead to MV replacement in a young patient who otherwise had a fairly normal mitral valve
- Patient received implantable cardioverter-defibrillator (ICD) for secondary prevention of sudden cardiac death
- The patient had a recurrence of V-tach 1 year later and his ICD successfully converted him

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Conclusion

Double-chambered left ventricle (DCLV) is a rare congenital defect (0.04% to 0.42%)

- Characterized by division of LV chamber into two chambers by abnormal muscular tissue
- Generally asymptomatic without complications
- Can present with life-threatening arrhythmias

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Conclusion

- DCLV should be suspected by echocardiography in the presence of:
  - Unusual aneurysmal appearance
  - Subdivision of the LV by an anomalous muscular ridge
- The echo diagnosis is aided by the use of contrast and 3D
- MRI is vital to help rule out differential diagnosis
- The anomalous muscle ridge can cause the development of VTach

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Thank you!

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Slides / Hand-out available at

www.cardioserv.net/MCSS

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References

- Myxoma: https://www.cvcasejournal.com/article/S2468-6441(18)30190-7/fulltext
- Pulm Emb: https://www.cvcasejournal.com/article/S2468-6441(19)30014-9/fulltext *Right Ventricular Thrombus in Transit: Raising the Stakes in the Management of Pulmonary Embolism. Everett Lo, MD, Shudhanshu Aluhetti, MD, Jonathan M. Wong, MD, Leyla Delic, MD, Glenn Egria, MD, Andrew Rosenblatt, MD. CASE.* DOI: 10.1016/j.case.2019.05.006

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