



70 Year Old Man with Dyspnea, Angina and T-wave Inversions on ECG *Case Presentation*

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70 Year Old Man With Dyspnea and Angina *2 Years Ago*

- Visual disturbance
- Dx of HCM, apical variant by echo

70 Year Old Man With Dyspnea and Angina *2 Years Ago*

- Progressive dyspnea and mild angina
- 80% RCA lesion was stented
- Small apical pouch on LV gram
- LVEDP 30 mmHg
- Systolic pressure in pouch 300 mmHg

70 Year Old Man With Dyspnea and Angina *2 Years Ago*

- Placed on beta blocker
- Referred to Mayo Clinic

68 Year Old Man With Progressive Dyspnea and Angina

Medications

- Metoprolol tartrate 100 mg qd
- Ezetimibe/simvastatin 10-40 mg qd
- Plavix 75 mg qd
- ASA 81 mg qd

68 Year Old Man With HCM, Apical Variant *Family Hx*

- Brother died in his 60s of "heart disease"
- Nephew died suddenly while running
- Another nephew diagnosed with HCM

68 Year Old Man With HCM, Apical Variant

Comorbidities

- CAD; s/p RCA stent
- Obstructive sleep apnea
- Hyperlipidemia



68 Year Old Man With HCM, Apical Variant

Echo

- HCM, apical variant; small apical pouch
- LVEF 70%
- Prominent, anteriorly-displaced anterolateral papillary muscle
- Grade 2/4 diastolic dysfunction
- RVSP 39 mmHg



68 Year Old Man With HCM, Apical Variant

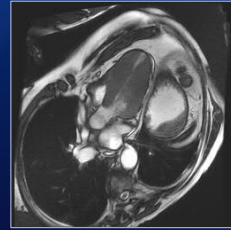
Cardiac MRI

- Apical variant HCM
- Small apical pouch
- Abnormal myocardial delayed enhancement involving inferior and lateral segments of pouch and small area of mid level anterior segment



68 Year Old Man With HCM, Apical Variant

Cardiac MRI



68 Year Old Man With HCM, Apical Variant

Holter

Mean Rate	1:27:20 AM - 2:00:00 PM	Standard Deviation	Rate Dependent Events	Pauses	C
Min	45 bpm	12.4112 HR	0 beats	Longest	0:00 min
Max	174 bpm	14.5854 HR	0 beats	Longest	0:00 min
Avg	70 bpm	14.5854 HR	0 beats		

Total Beats	4	Complex	0	Total Beats	100	Complex	0
% Beats	0.01	Totals	0 <td>% Beats</td> <td>0.17</td> <td>Complex</td> <td>0 </td>	% Beats	0.17	Complex	0
Events	4	Phonetic Runs	0 <td></td> <td></td> <td></td> <td></td>				

Min Rate	0 bpm	SVT/VT Runs	2	Max Rate	210.14 bpm
Avg Rate <td>0 bpm</td> <td>Max Rate <td>100 bpm</td> <td>Max Rate <td>111.85 bpm</td> </td></td>	0 bpm	Max Rate <td>100 bpm</td> <td>Max Rate <td>111.85 bpm</td> </td>	100 bpm	Max Rate <td>111.85 bpm</td>	111.85 bpm
Min Rate <td>0 bpm</td> <td>Max Rate <td>5 beats</td> <td>Max Rate <td>111.85 bpm</td> </td></td>	0 bpm	Max Rate <td>5 beats</td> <td>Max Rate <td>111.85 bpm</td> </td>	5 beats	Max Rate <td>111.85 bpm</td>	111.85 bpm
Max Rate <td>111.85 bpm</td> <td>Max SVT/VT <td>19 beats</td> <td>Max SVT/VT <td>300.06 bpm</td> </td></td>	111.85 bpm	Max SVT/VT <td>19 beats</td> <td>Max SVT/VT <td>300.06 bpm</td> </td>	19 beats	Max SVT/VT <td>300.06 bpm</td>	300.06 bpm
Max SVT/VT <td>0.3</td> <td>Max SVT/VT <td>6.2</td> <td></td> <td></td> </td>	0.3	Max SVT/VT <td>6.2</td> <td></td> <td></td>	6.2		
Max SVT/VT <td>0.1</td> <td>Max SVT/VT <td>1.7</td> <td></td> <td></td> </td>	0.1	Max SVT/VT <td>1.7</td> <td></td> <td></td>	1.7		

Impressions and Findings

- The basic rhythm was sinus with rates ranging from 45-174 bpm.
- There were 4 PVCs noted.
- There were 4 complex runs of 1-3 beats in the form of supraventricular tachycardia (SVT). The maximum SVT rate was 100 bpm.
- ST analysis was not done due to hyperlipidemic cardiomyopathy.
- The patient reported "bunches of beats" at that time the rate was 174 bpm.



68 Year Old Man With HCM, Apical Variant

TMET

Final ECG Impressions

Maximal test
 Stress ECG is abnormal but non-diagnostic for ischemia
 Exercise capacity is average
 Normal blood pressure response to exercise
 Heart rate response limited due to medications
 Abnormal heart rate recovery
 No arrhythmias noted
 No chest pain reported during test

Final VO2 Impressions

Below average peak VO2
 Maximal effort
 Cardiac output impairment
 Pulmonary response normal

Prognosis Based on Stress Test: Intermediate CV Risk

Comment: Below normal VO2 peak with plateau consistent with cardiac limitation



68 Year Old Man With Dyspnea and Angina Despite Med Rx

- Tapered off metoprolol
- Placed on verapamil
- Developed side effects
- Verapamil discontinued and metoprolol restarted

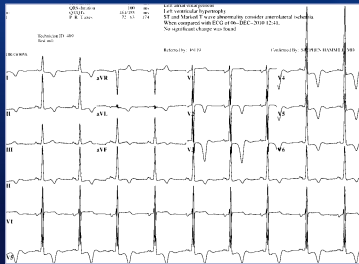


70 Year Old Man With Dyspnea and Angina Despite Med Rx

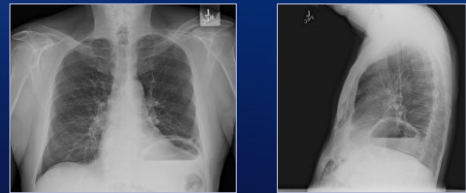
- Returned to Mayo Clinic
- Progressive dyspnea for 6 months
- Angina if he walks through dyspnea
- Symptoms worse after eating
- No palpitations
- No syncope/near-syncope



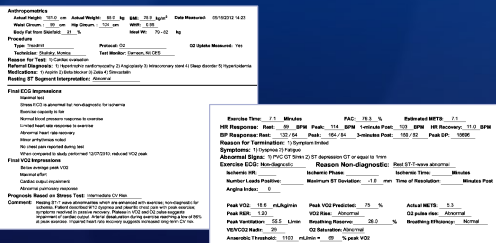
70 Year Old Man With Dyspnea and Angina ECG



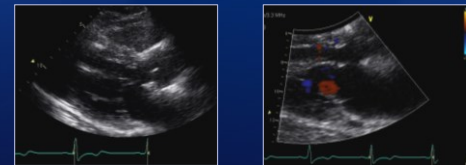
73 Year Old Man With Dyspnea and Angina Chest X-ray



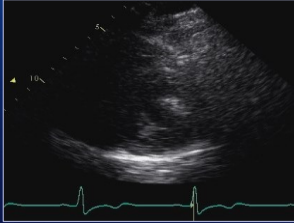
70 Year Old Man With Dyspnea and Angina Despite Med Rx O2 Consumption TMET



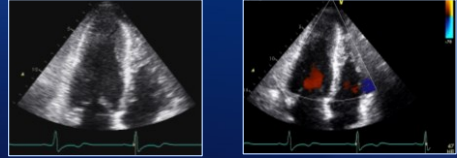
70 Year Old Man With Dyspnea and Angina



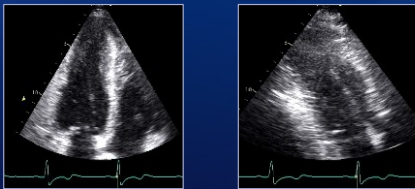
70 Year Old Man With Dyspnea and Angina



70 Year Old Man With Dyspnea and Angina



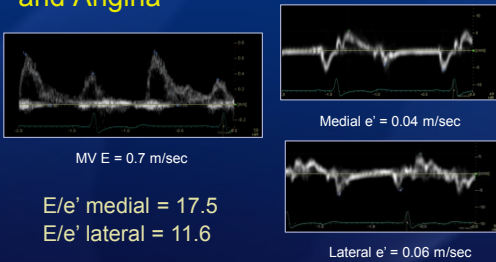
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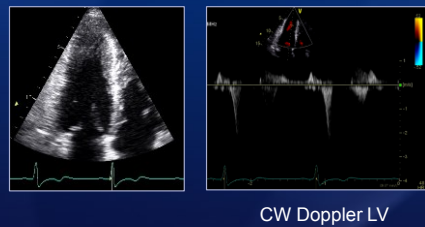
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70 Year Old Man With Dyspnea and Angina

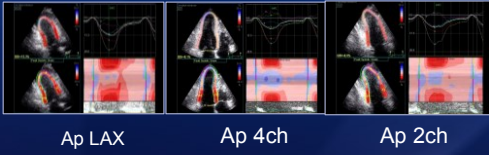


70 Year Old Man With Dyspnea and Angina



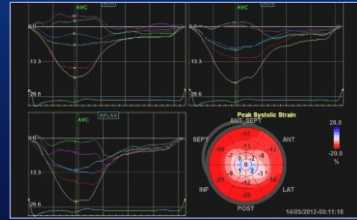
70 Year Old Man With Dyspnea and Angina

LV Longitudinal Systolic Strain



70 Year Old Man With Apical HCM

LV Longitudinal Systolic Strain



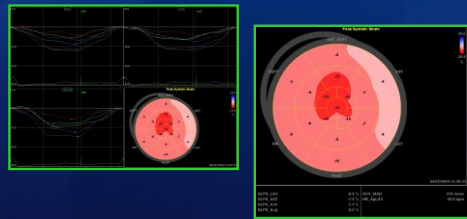
70 Year Old Man With Apical HCM

LV Longitudinal Systolic Strain

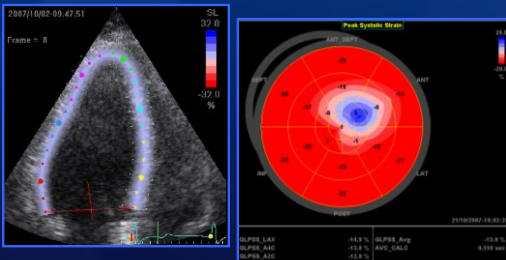


Advanced Cardiac Amyloid

2D Strain



Apical Hypertrophic Cardiomyopathy



What Happened to the Apical Pouch?

- 1) Spontaneously resolved
- 2) Obliterated by thrombus
- 3) Other

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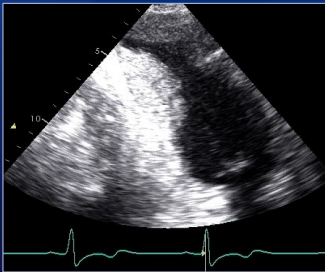


70 Year Old Man With Apical HCM

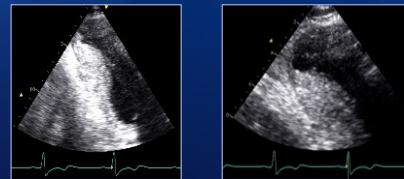
Additional Echo Images *From Lower Interspace*



70 Year Old Man With Apical HCM



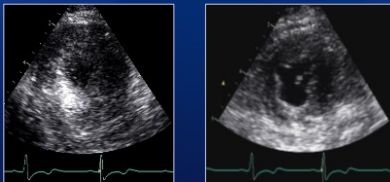
70 Year Old Man With Apical HCM



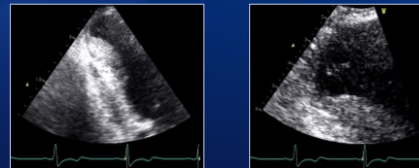
Apical Pouch



70 Year Old Man With Apical HCM



70 Year Old Man With Apical HCM



Apical Pouch



HCM, Apical Variant Apical Pouch

- 10-20% with ApHCM may develop apical aneurysm/diverticulum
- Frequently have normal epicardial coronary arteries
- Stress nuclear testing often demonstrates apical ischemia



HCM, Apical Variant Apical Pouch

- Increased risk of systemic emboli
- Increased incidence of sustained ventricular arrhythmias



Apical myectomy: A new surgical technique for management of severely symptomatic patients with apical hypertrophic cardiomyopathy

Hartzell V. Schaff, MD,^a Morgan L. Brown, MD,^a Joseph A. Dearani, MD,^a Martin D. Abel, MD,^b Steve R. Ommen, MD,^c Paul Sorajja, MD,^a A. Jamil Tajik, MD,^d and Rick A. Nishimura, MD^e

Schaff H; *Journal Thor and CV Surgery* 139
March 2010



70 Year Old Man With Dyspnea and Angina Despite Med Rx ECHO

- HCM, apical variant; small apical pouch
- Small LV cavity; EF 67%
- Grade 2/4 diastolic dysfunction
- Mild LA enlargement
- RVSP 54 mmHg



70 Year Old Man With Dyspnea and Angina Despite Med Rx Coronary Angio

- Right dominant
- Normal left main
- 20% mid LAD
- 30% distal cx
- 20% mid RCA



70 Year Old Man With Dyspnea and Angina Despite Med Rx Surgical Findings

- Apex of heart completely obliterated by hypertrophy
- Small apical pouch
- Endocardial scar in region of apposition of lateral wall and pap muscles with septum
- Anomalous pap muscle originating from septum and coursing to AML, inserting near free edge
- No LVOT obstruction



70 Year Old Man With Dyspnea and Angina Despite Med Rx *Cardiac Surgery*

- Apical ventriculotomy over pouch
- Through this incision removed muscle from septum and shaved both pap muscles
- Closed apical incision
- Inspected basal ventricular septum via aortotomy; no obstruction
- Closed aortotomy

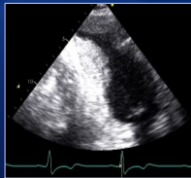


70 Year Old Man Status Post Apical Myectomy *Post-op Course*

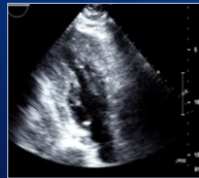
- Extubated evening of surgery
- Transferred to progressive care unit next day
- Left pleural effusion; 570 cc straw-colored fluid at centesis
- Afib; Rx with amiodorone
- In SR at dismissal day 7



70 Year Old Man With Apical HCM



Pre-op



Post-op



70 Year Old Man With HCM, Apical Variant

- There is about 30% chance of identifying causative genetic mutation for patients with apical variant
- Genetic testing for this patient did not identify any mutations of clinical significance



68 Year Old Man With HCM, Apical Variant

Recommendations for Screening of Relatives ECG and Echo

- Every 5 years for first and second degree relatives not participating in athletics (annually if participating in athletics)
- Annually for adolescent first degree relatives
- Every 2-3 years for children; annually after onset of puberty
- Screen nieces, nephews and grandchildren before starting competitive athletics



Mayo Clinic Locations

