



# Primary Tumors of the Heart

Mitul B. Kadakia, HMS III

Gillian Lieberman, M.D.

Beth Israel Deaconess Medical Center





# Objectives

- Background & Patient Presentation
- Radiological Tests used to identify cardiac tumors
  - Indications, Advantages, & Disadvantages
  - Normal Scan and Anatomy
  - Our patient
  - Effect on differential



# Background

---

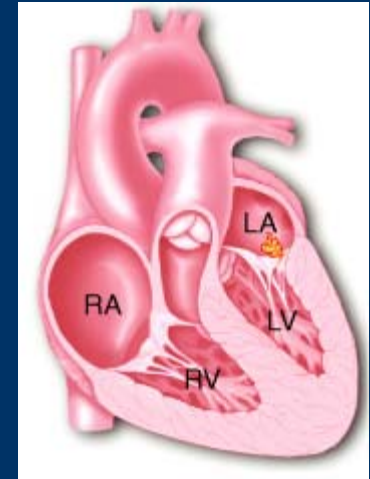
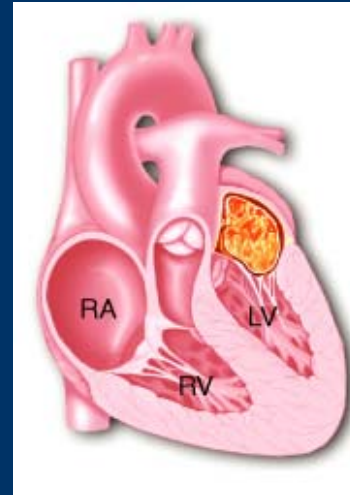
- Extremely rare (incidence 0.01%-0.02% in autopsy series)
- Often missed or misdiagnosed
- >75% benign



# Clinical Presentation

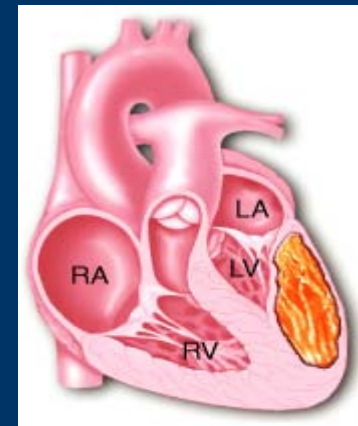
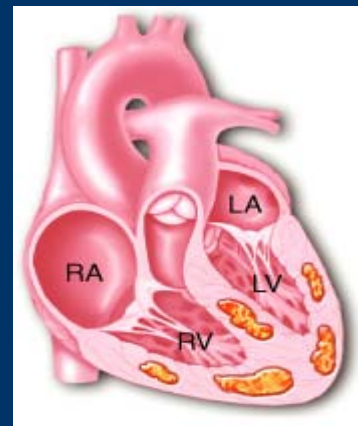
## INTRACAVITARY

- Heart Failure (CHF or low-output)
- Embolic event (PE, stroke)



## INTRAMURAL

- Heart Failure (systolic or diastolic dysfunction)
- Arrhythmia





# Patient C.H.

---

- 3 year old male with history of dyspnea and cyanosis.
- EKG notable for periods of supraventricular tachyarrhythmia



# Differential Diagnosis

- Valvular Heart Disease (Congenital, AR, AS, MR, MS)
- Hypertrophic cardiomyopathy
- Heart Failure
- Arrhythmia
- Ischemic Heart Disease
- Pulmonary Embolism
- Pulmonary HTN
- Pneumonia
- **Cardiac Mass**



# Differential of Cardiac Mass in Children

## Benign

<u>Tumor</u>	<u>General Appearance</u>
Rhabdo-Myoma (42%)	Multiple, small, Intramural masses
Fibroma (18%)	Intramural, large, Solid mass, ant. LV
Myxoma (17%)	Mobile, pedunculated LA > RA
Teratoma (12%)	Multiple tissue layers In pericardium

Braunwald, *Heart Disease 7<sup>th</sup> ed.*; Sparrow, *Radiographics 2005*

## Malignant

*Rhabdomyosarcoma (41%)*  
*Fibrosarcoma (18%)*

## Other

*Lipomatous Hypertrophy*    *Endocarditis*    *Cyst*  
*Thrombus*    *Metastatic CA*



# Menu of Tests

- Chest Roentgenogram
- Computerized Tomography
- Echocardiography
- Magnetic Resonance Imaging
- *Nuclear Medicine*
- *Angiography*



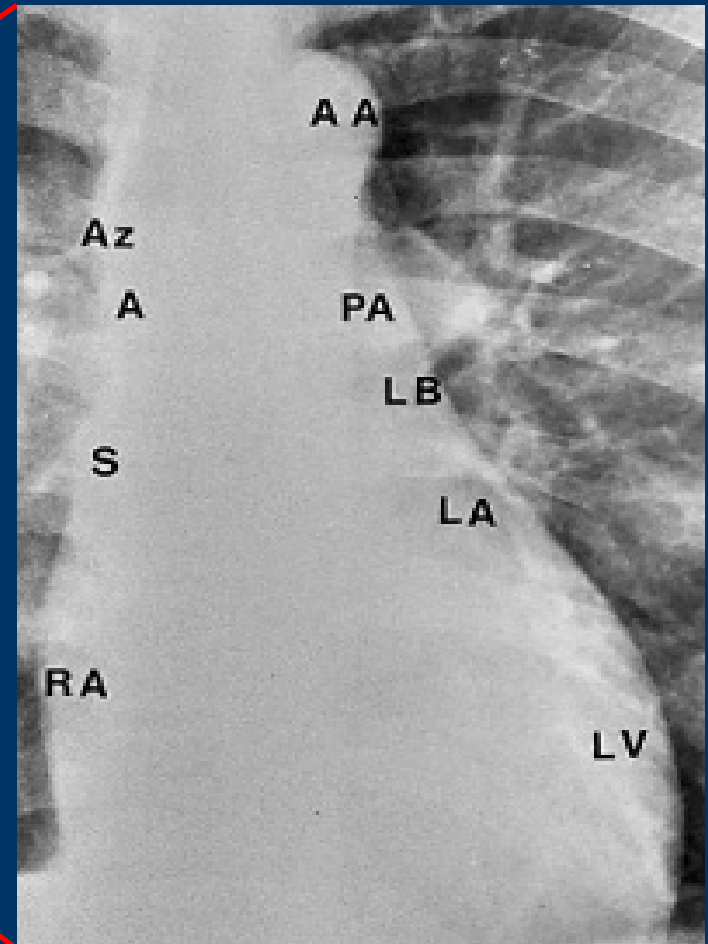
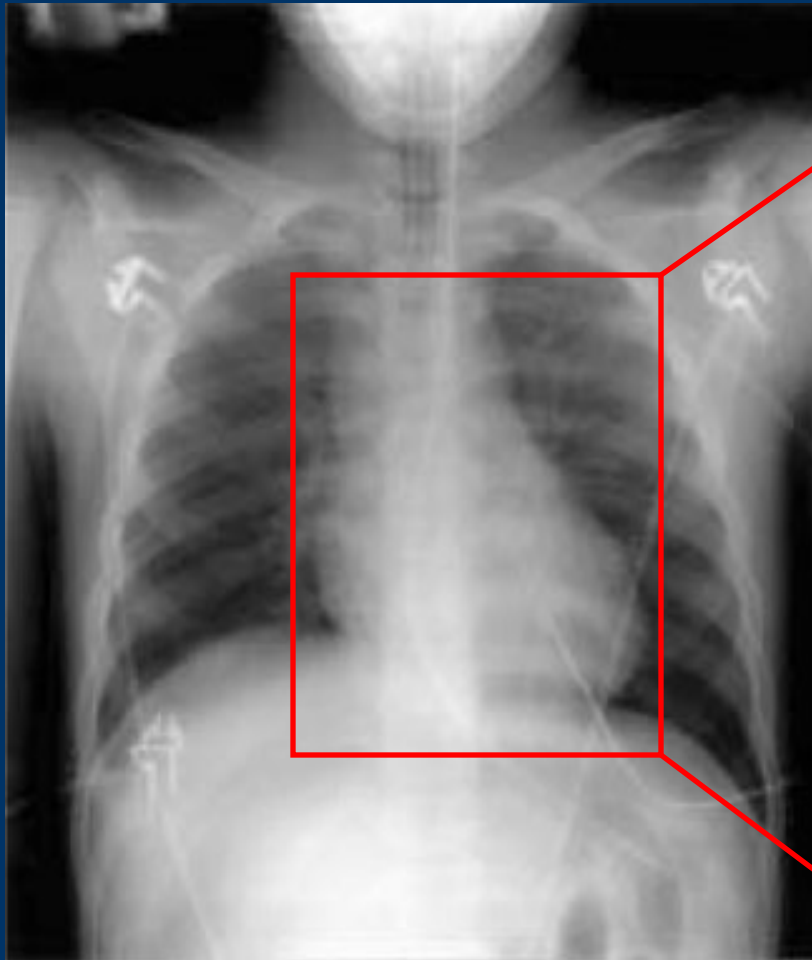


# Chest Roentgenogram

- Give first “hints” of tumor
- Can look for cardiac chamber enlargement, pericardial effusion, calcifications
- Associated pulmonary changes may be seen
- Limited information on tissue differentiation, morphology, location, mobility, infiltration

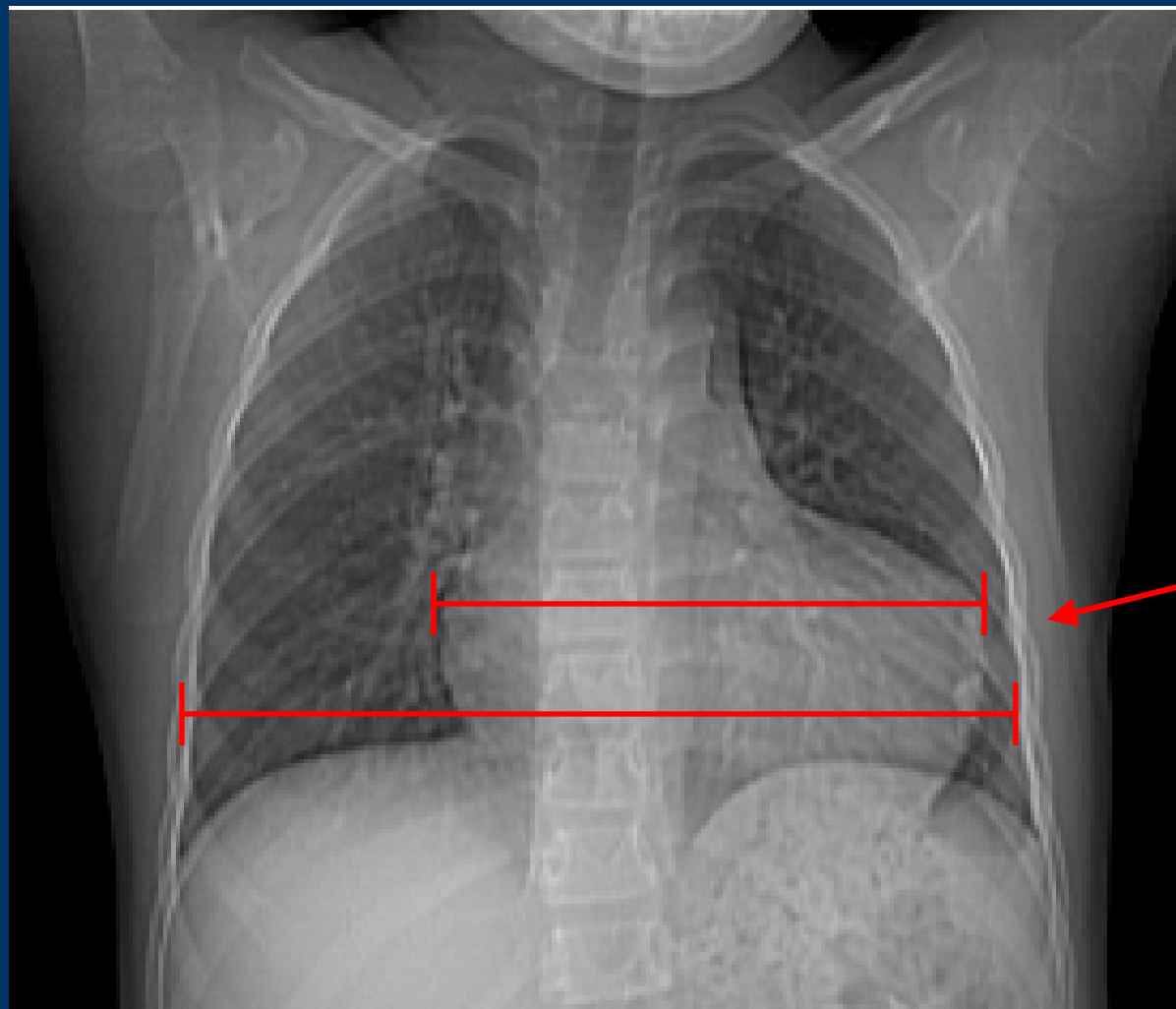


# Normal Chest X-Ray





# CT Scout – Patient C.H.



**Heart Diameter  
> 50% of thorax  
diameter**



# Computerized Tomography

- Can be used to assess size, location, myocardial infiltration, intraluminal invasion (with contrast), and effect on surrounding structures.
- Shows calcifications
- Views include mediastinum and surrounding structure
- Faster, Increased availability
- High Resolution



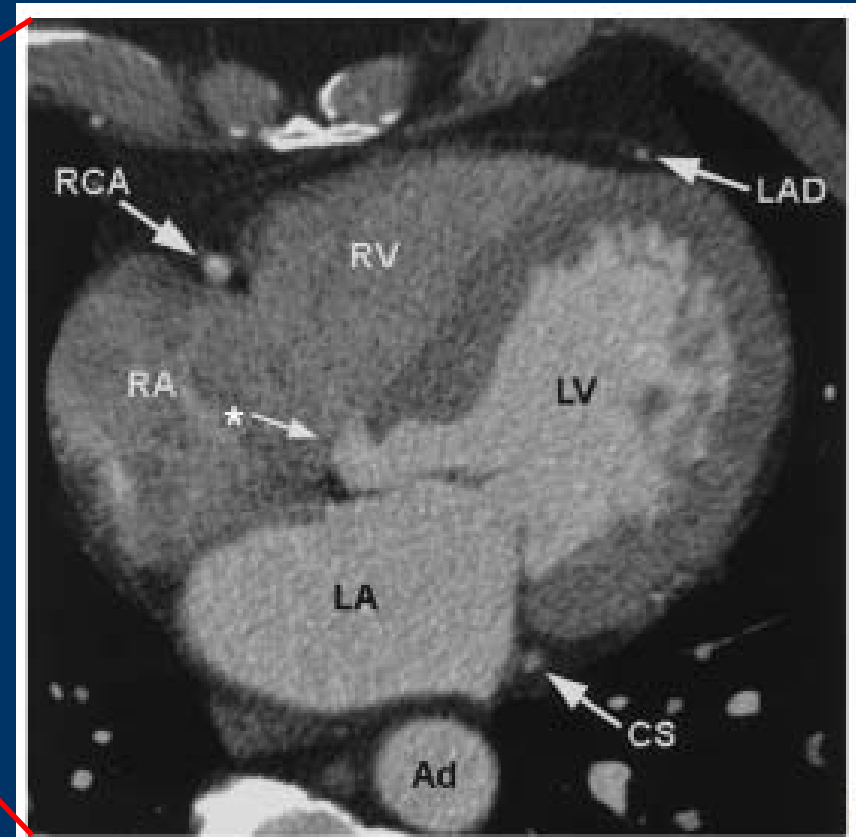
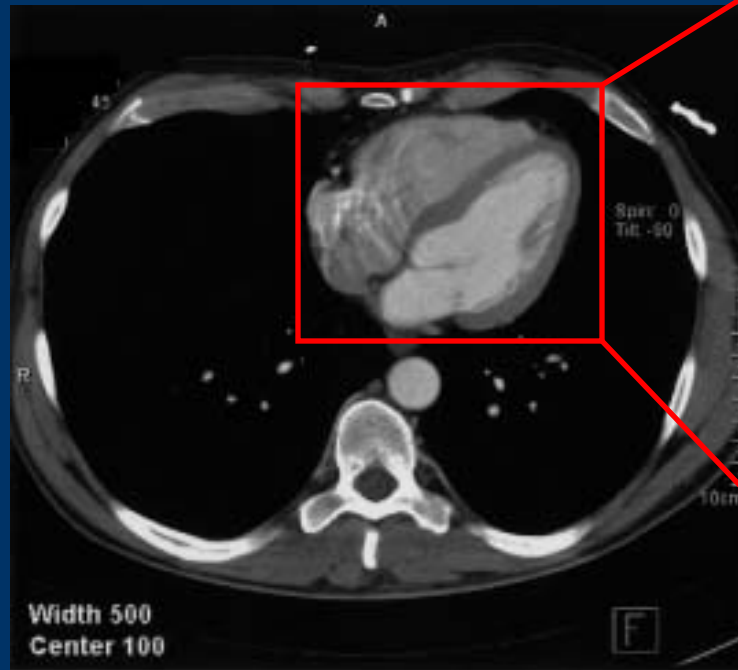
# CT - Limitations

---

- Recent advances have improved CT soft tissue discrimination, but still not as good as MR.
- Limited planes
- +Radiation

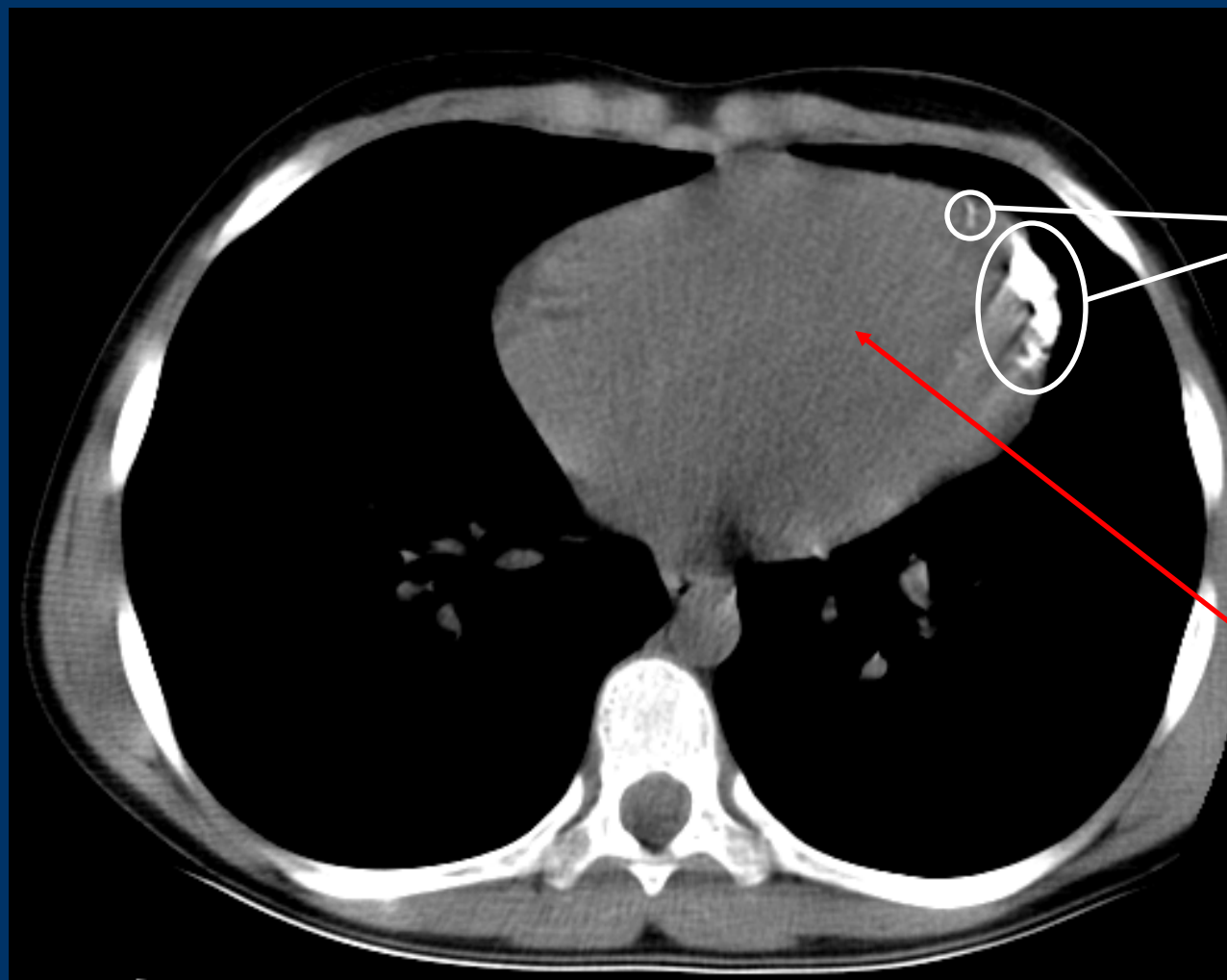


# Computerized Tomography - Normals





## Non-Contrast, Axial CT – Patient C.H.



**Abnormal  
Calcification**

**Homogeneous,  
soft tissue  
attenuation,  
enlargement of  
heart**

Courtesy Dr. Jason Handwerker,  
BIDMC/Children's



# Differential Diagnosis

- Valvular Heart Disease (Congenital, AR, AS, MR, MS)
- Hypertrophic cardiomyopathy
- Heart Failure
- Arrhythmia
- Ischemic Heart Disease
- Pulmonary Embolism
- Pulmonary HTN
- Pneumonia
- Cardiac Mass





# Which Mass is it?

## Benign

Braunwald, *Heart Disease 7<sup>th</sup> ed.*; Sparrow, *Radiographics 2005*

### Tumor

### General

### CT

Rhabdomyoma

Multiple, small,  
Intramural masses

Homogenous,  
Low attenuation



Fibroma

Intramural, large,  
Solid mass, ant. LV

Homogenous  
Low attenuation  
Calcifications



Myxoma

Mobile, pedunculated  
LA > RA

Heterogeneous  
Low attenuation  
Occasional Calc's



Teratoma

Multiple tissue layers  
In pericardium

Heterogeneous

## Malignant

*Rhabdomyosarcoma*  
*Fibrosarcoma*

## Other

*Lipomatous Hypertrophy*    *Endocarditis*    *Cyst*  
*Thrombus*    *Metastatic CA*



# Echocardiography

- Screening test of choice, portable, quick
- Can assess mobility, size, location, and attachments.
- Real-time images
- Through color doppler, can assess impact on myocardial blood flow.
- Transthoracic vs. Transesophageal

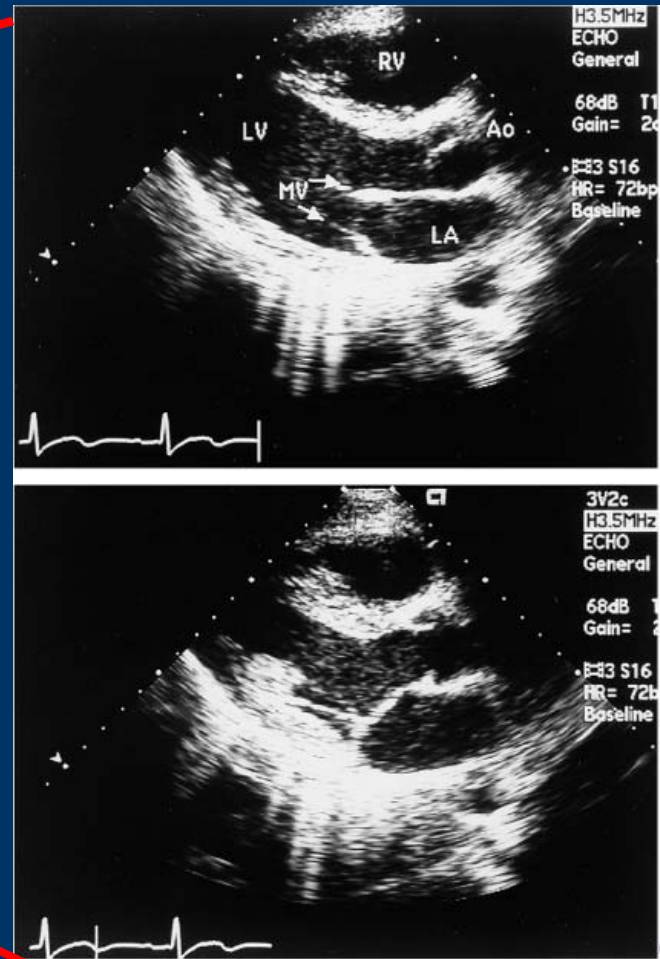
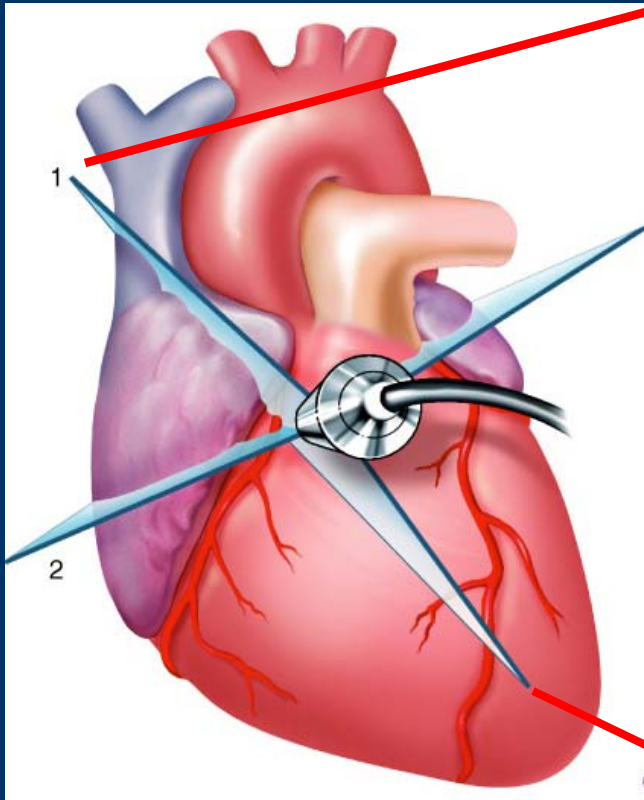


# Echocardiography - Limitations

- Limited by operator experience, body habitus
- Limited views
- Limited soft tissue characterization
- Invasive, with associated risks (TEE)



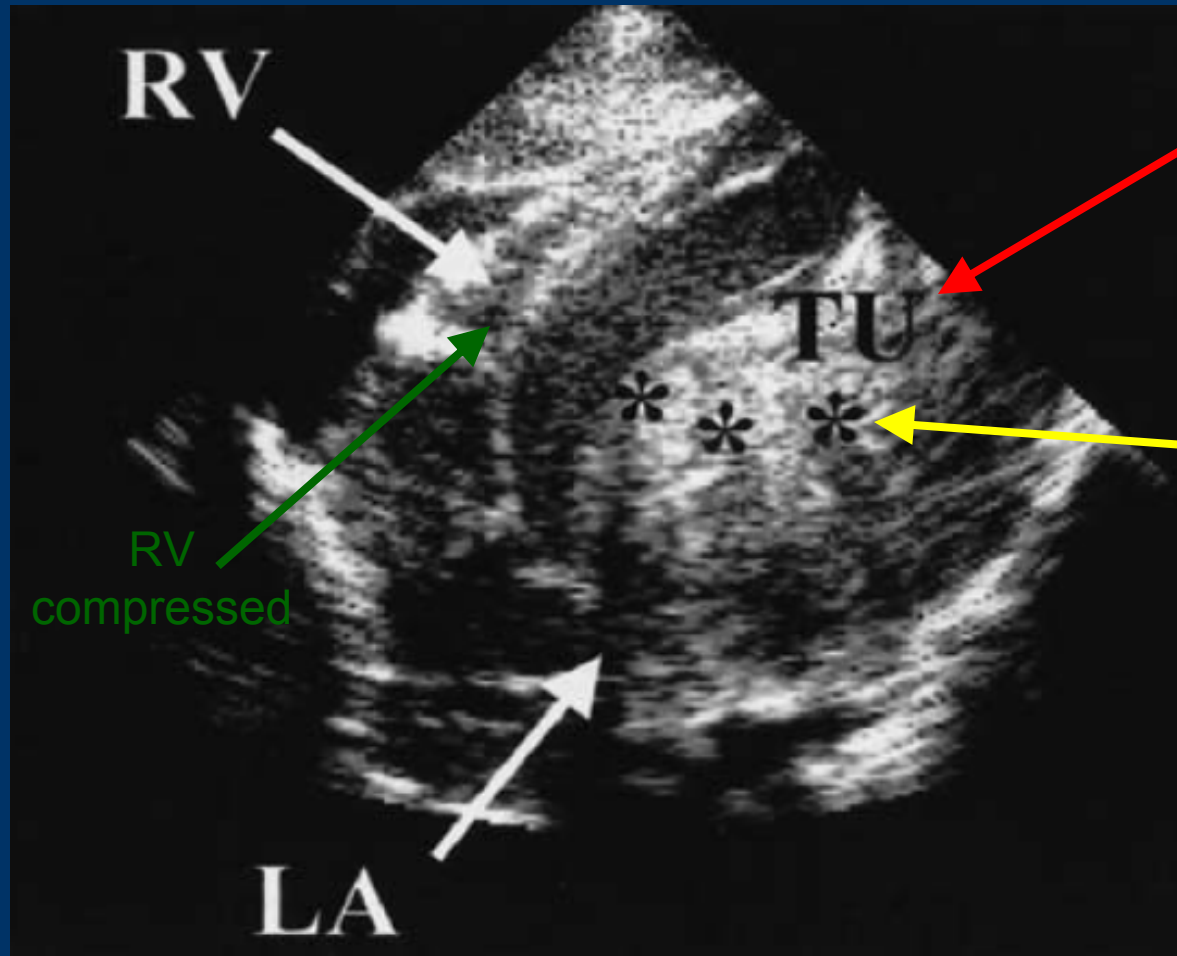
# TTE - Normal



DI  
AS  
TO  
LE  
  
SY  
ST  
O  
LE



# TTE, 4 chamber view – Companion Patient #1



Hyperechogenic mass filling almost entire LV

Small central calcifications, with acoustic shadowing

Gutberlet, et al. *European Radiology* 2002



# Differential Diagnosis

- Valvular Heart Disease (Congenital, AR, AS, MR, MS)
- Hypertrophic cardiomyopathy
- Endocarditis
- Heart Failure
- Arrhythmia
- Ischemic Heart Disease
- Pulmonary Embolism
- Pulmonary HTN
- Pneumonia
- Cardiac Mass



# Which Mass is it?

## Benign

Braunwald, *Heart Disease 7<sup>th</sup> ed.*; Sparrow, *Radiographics 2005*

### Tumor

Rhabdomyoma

### General

**Multiple, small, Intramural masses**

### CT

Homogenous, Low attenuation ✓

### Echo

**Hyperechoic** ✓

Fibroma

**Intramural, large, Solid mass, ant. LV** ✓

Homogenous Low attenuation ✓  
Calcifications

**Hyperechoic** ✓

Myxoma

**Mobile, pedunculated LA > RA**

Heterogeneous Low attenuation ✓  
Occasional Calc's

**Heterogeneous**

Teratoma

**Multiple tissue layers In pericardium**

Heterogeneous

**Heterogeneous**

## Malignant

*Rhabdomyosarcoma*  
*Fibrosarcoma*

## Other

*Lipomatous Hypertrophy*    *Endocarditis*    *Cyst*  
*Thrombus*    *Metastatic CA*



# Magnetic Resonance Imaging

- Superior soft tissue characterization, multiple planes, no ionizing radiation
- Specialized sequences allow assessment of detailed morphological information, suppression of tissue (i.e. fat), demonstration of fluid, vascularity
- Views include mediastinum and surrounding structure
- Helpful in surgical planning
- 3-D imaging allows for evaluation of cardiac function, blood flow, and mobility of the mass





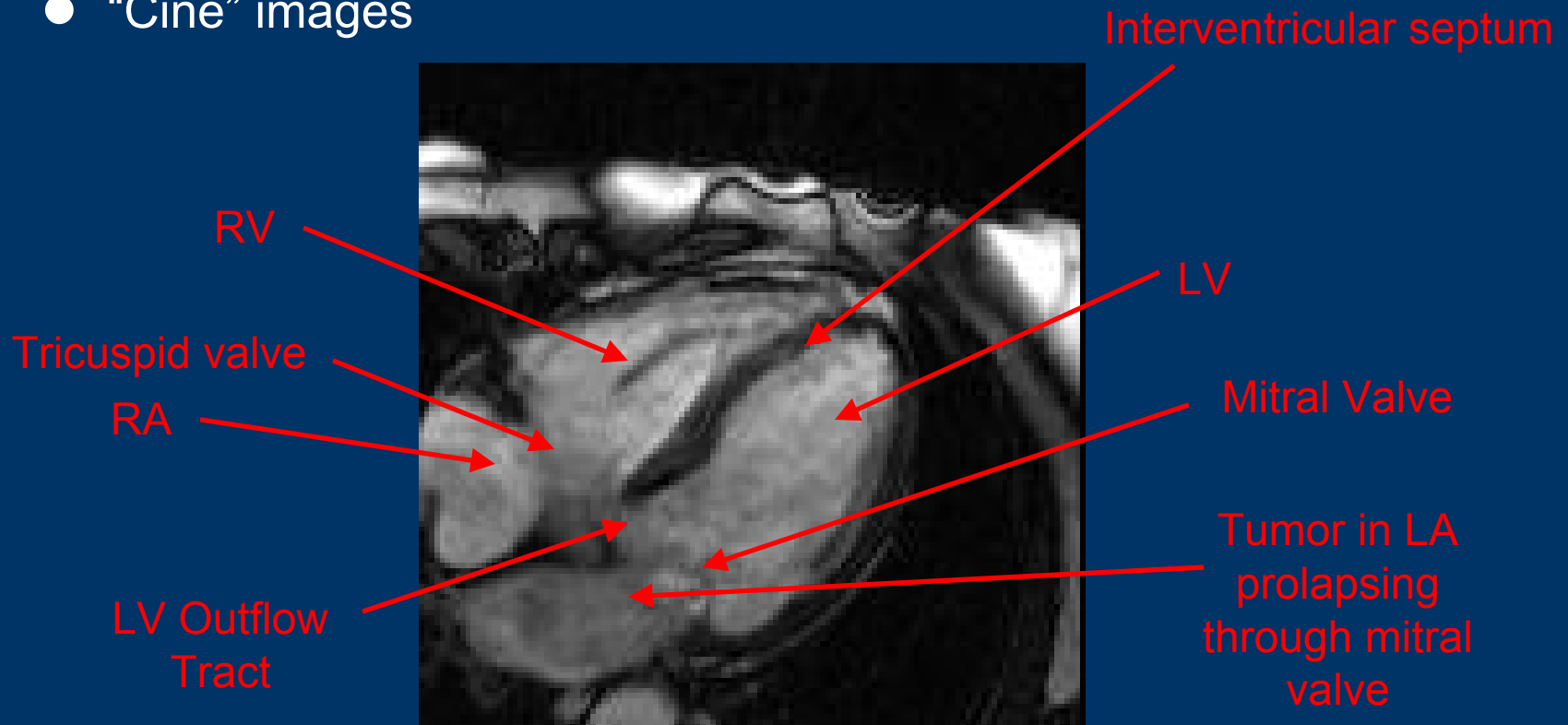
# MRI - Limitations

- Calcifications not seen
- Time consuming, Expensive
- Claustrophobia
- Requires EKG gating



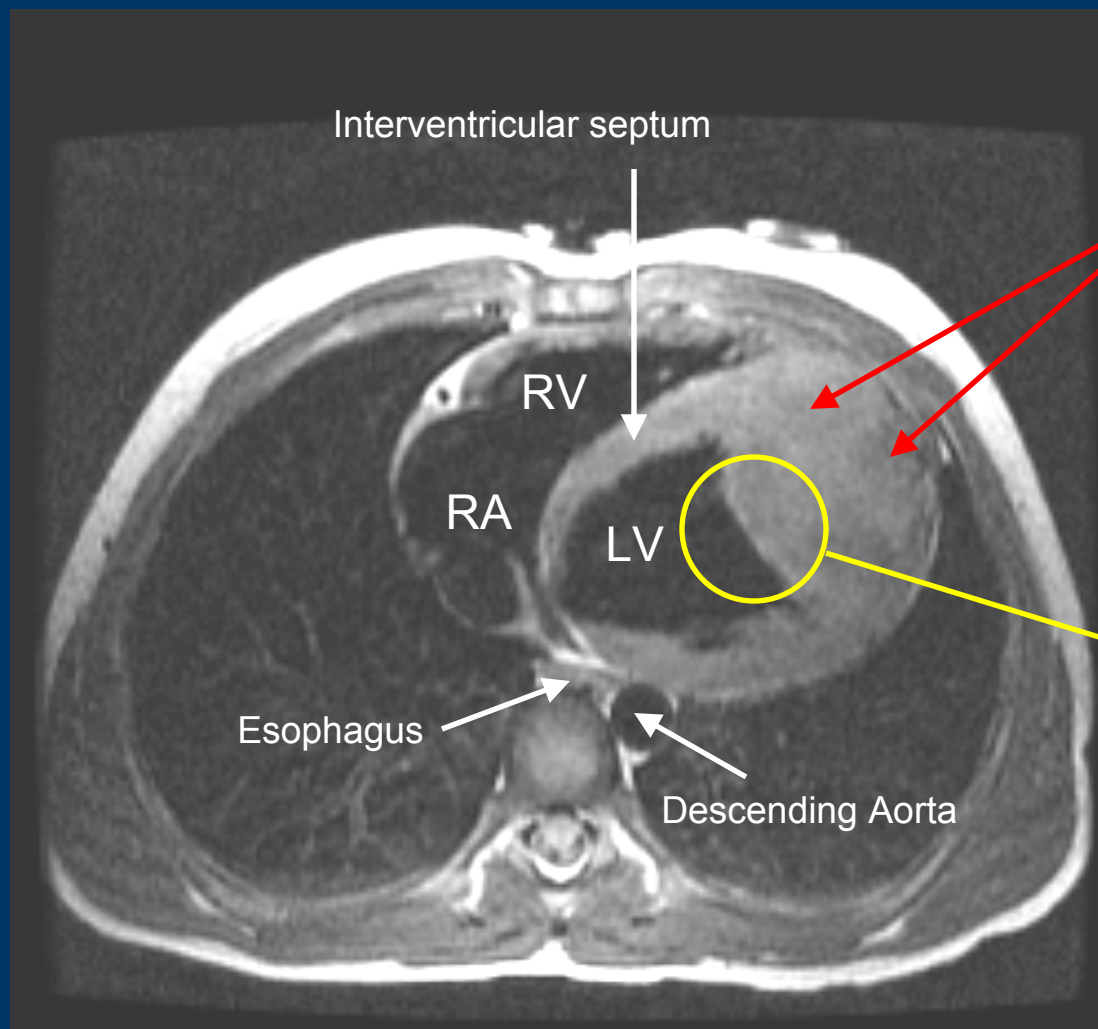
# Cardiac MRI

- Gated to “R” wave
- “Black-Blood” and “Bright-Blood” images
- “Cine” images





# Gadolinium-Enhanced, T1 Weighted Double IR, Fast Spin Echo (“Black Blood”) – Patient C.H.



Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration

Endocardium



## Gradient Echo (“Bright Blood”) – Patient C.H.

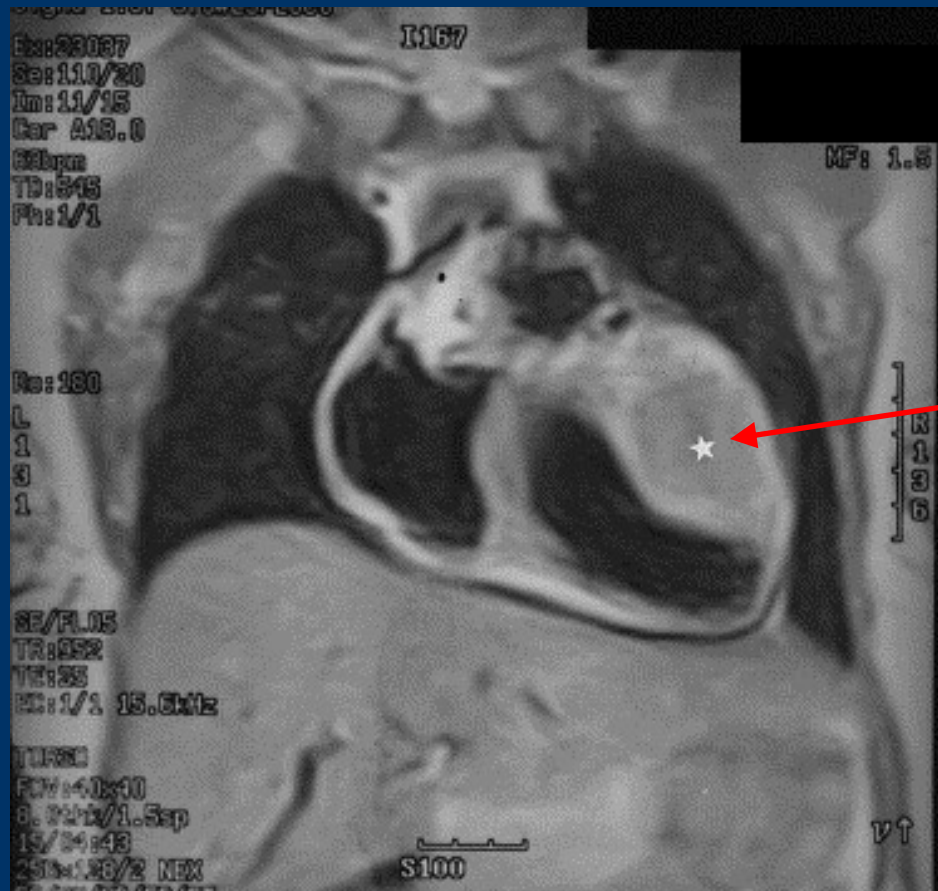


Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration

Endocardium



# Coronal MRI, Spin Echo T1 weighted – Companion Patient #2



Well-defined, homogenous, isointense mass arising from anterior left ventricular wall



# Which Mass is it?

## Benign

<u>Tumor</u>	<u>General</u>	<u>CT</u>	<u>Echo</u>	<u>MRI</u>
Rhabdomyoma	Multiple, small, Intramural masses	Homogenous, Low attenuation ✓	Hyperechoic ✓	T1 – Isointense ✓ T2 - Hyperintense
Fibroma	Intramural, large, Solid mass, ant. LV ✓	Homogenous Low attenuation Calcifications ✓	Hyperechoic ✓	T1 – Isointense ✓ T2 - Hypointense
Myxoma	Mobile, pedunculated LA > RA	Heterogeneous Low attenuation Occasional Calc's ✓	Heterogeneous	Heterogeneous T1 – Isointense ✓ T2 - Hyperintense
Teratoma	Multiple tissue layers In pericardium	Heterogeneous	Heterogeneous	Heterogeneous

Braunwald, *Heart Disease 7<sup>th</sup> ed.*; Sparrow, *Radiographics 2005*

## Malignant

*Rhabdomyosarcoma*  
*Fibrosarcoma*

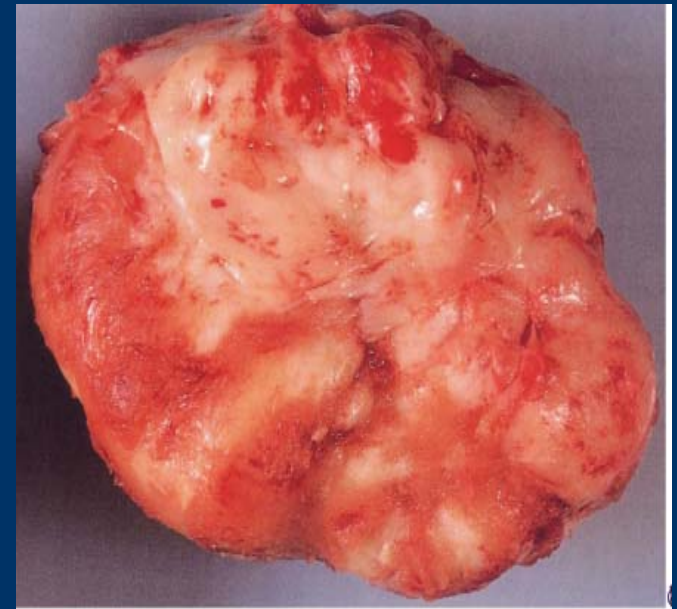
## Other

*Lipomatous Hypertrophy*    *Endocarditis*    *Cyst*  
*Thrombus*    *Metastatic CA*



# Cardiac Fibroma

- Benign connective tissue tumor, arises from fibroblasts
- Children <10 years of age
- 3-10 cm. Most commonly in anterior free wall of LV. Can be in posterior LV or RV.
- Increased incidence in Gorlin Syndrome (nevroid basal cell CA, medulloblastomas, fibrous histiocytomas, skin and skeletal abnormalities)
- Treatment – Surgical excision





# Summary of Radiological Tests

<u>Test</u>	<u>Value</u>	<u>Limitations</u>
CXR	Heart enlargement Lung findings	Location, Mobility, Infiltration, Morphology, Tissue Diff.
CT	Calcifications, size, location, infiltration, faster, ↑resolution	Soft tissue differentiation, planes Radiation
Echo	Real-time, good screen, mobility, attachments, size, motion, location	Invasive (TEE), operator-based, Soft tissue differentiation, views
MRI	Soft tissue differentiation, morphology, location, size, infiltration, surgical planning, diff. planes, tissue suppression	Time-consuming, calcifications not seen, EKG gating





# Summary

- Cardiac tumors are rare, but important not to miss
- Be aware of other, more common cardiac masses that may mimic tumors
- Radiological tools can allow characterization of cardiac masses and tumors with high degree of certainty
- Each test is complementary and tells a part of the story



# Acknowledgments

- Dr. Gillian Lieberman
- Pamela Lepkowski
- Larry Barbaras, our webmaster
- Dr. Jason Handwerker
- Dr. Muneeb Ahmed
- Dr. Jesse Wei
- Dr. Kamlesh Dave
- Fellow students



# References

- Araoz P, SL Mulvagh, HD Tazelaar, PR Julsrud, JF Breen. CT and MR Imaging of Benign Primary Cardiac Neoplasms with Echocardiographic Correlation. *Radiographics* 2000; 20:1303-1319.
- Braunwald, Eugene. *Heart Disease*, 7<sup>th</sup> edition. Philadelphia: Elsevier, 2005
- Busch U, C Kampmann, R Meyer, KH Sandring, G Hausdorf, W Konertz. Removal of a Giant Cardiac Fibroma from a 4-year-old-child. *Texas Heart Institute Journal* 1995; 22:261-4.
- Carlaps J, J. Montiel, J. Reig, J. Boldu, F Carreras. Complete surgical excision of a huge left ventricular fibroma. *Journal of Thoracic and Cardiovascular Surgery* 2005; 129:1443-1444.
- Grebenc ML, ML Rosado de Christenson, AP Burke, CE Green, JR Galvin. Primary Cardiac and Pericardial Neoplasms: Radiologic-Pathologic Correlation. *Radiographics* 2000; 20:1073-1103.
- Gutberlet M, H Abdul-Khaliq, B Stiller, U Schubert, G Stoltenburg-Didinger, PE Lange, R Hetzer, R Felix. Giant fibroma in the left ventricle of an infant: imaging findings in magnetic resonance imaging, echocardiography and angiography. *European Radiology* 2002; 12:S143-8.
- Ipek G, D Mansuroglu, A Omeroglu, SN Omeroglu, I Mataraci. Cardiac Fibroma. *The Annals of Thoracic Surgery* 2005; 79:1786.
- Novelline, Robert. *Squire's Fundamentals of Radiology*, 6<sup>th</sup> ed. Cambridge: Harvard, 2004
- Pooley, Robert A. AAPM/RSNA Physics Tutorial for Residents: Fundamentals of MR Imaging. *Radiographics* 2005; 25:1087-1099.
- Shapiro, LM. Cardiac tumours: diagnosis and management. *Heart* 2001; 85:218-222.
- Sparrow PJ, JB Kurian, TR Jones, MU Sivanathan. MR Imaging of Cardiac Tumors. *Radiographics* 2005; 25:1255-76.
- Zarroug, Abdalla E., Penny Stavlo, Greg A. Kays, David A. Rodeberg, Christopher Moir. Accidental Burials in Sand: A Potentially Fatal Summertime Hazard. *Mayo Clinic Proceedings* 2004; 79:774-776.