



Primary Tumors of the Heart

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Objectives

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



Background

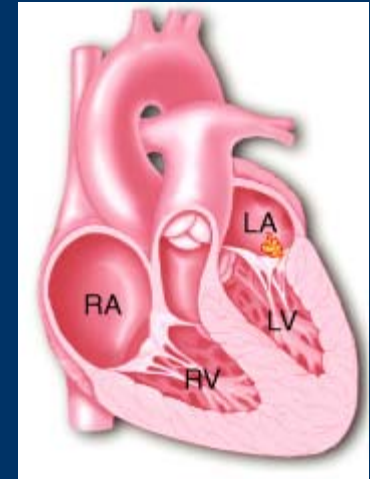
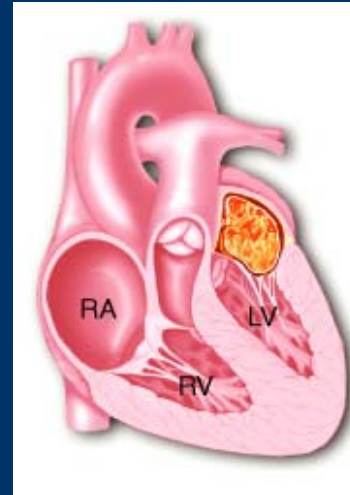
- Extremely rare (incidence 0.01%-0.02% in autopsy series)
- Often missed or misdiagnosed
- >75% benign



Clinical Presentation

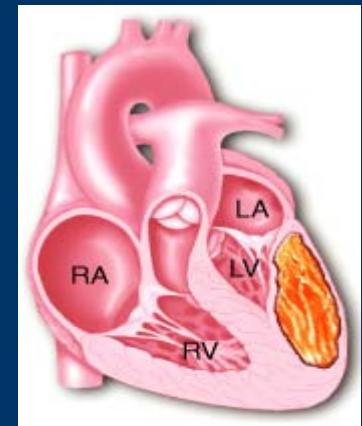
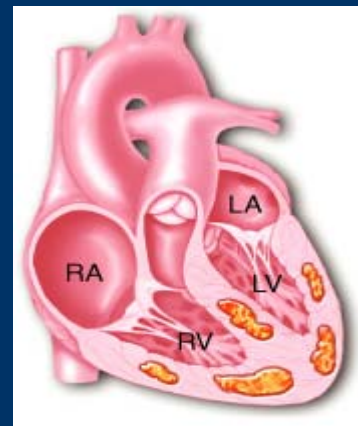
INTRACAVITARY

- 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 7th 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*

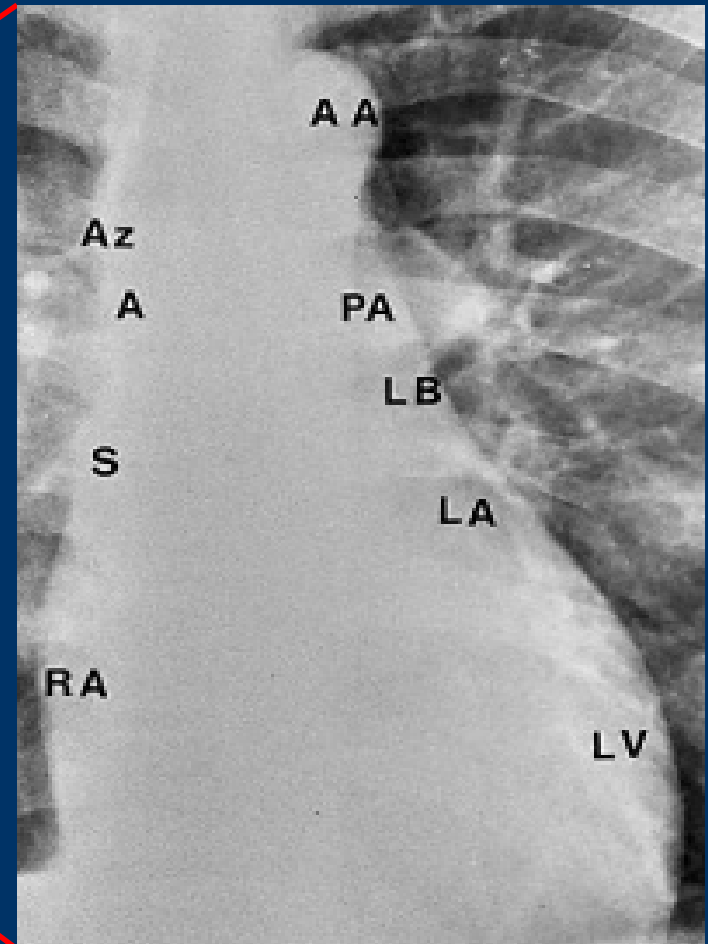
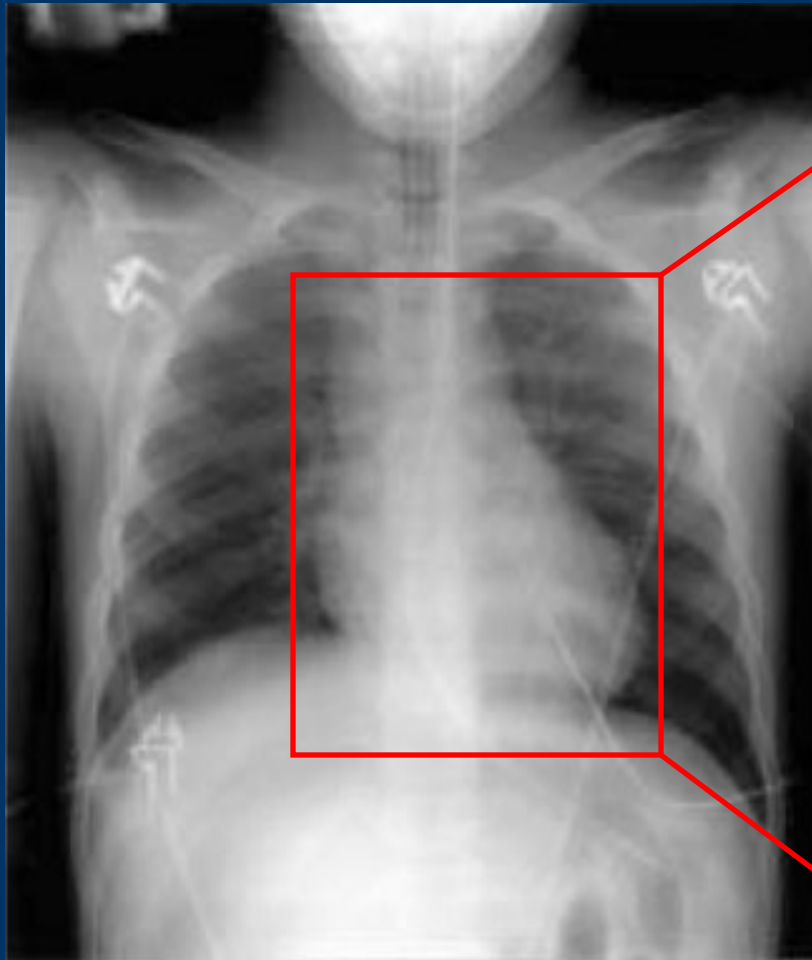


Chest Roentgenogram

- 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

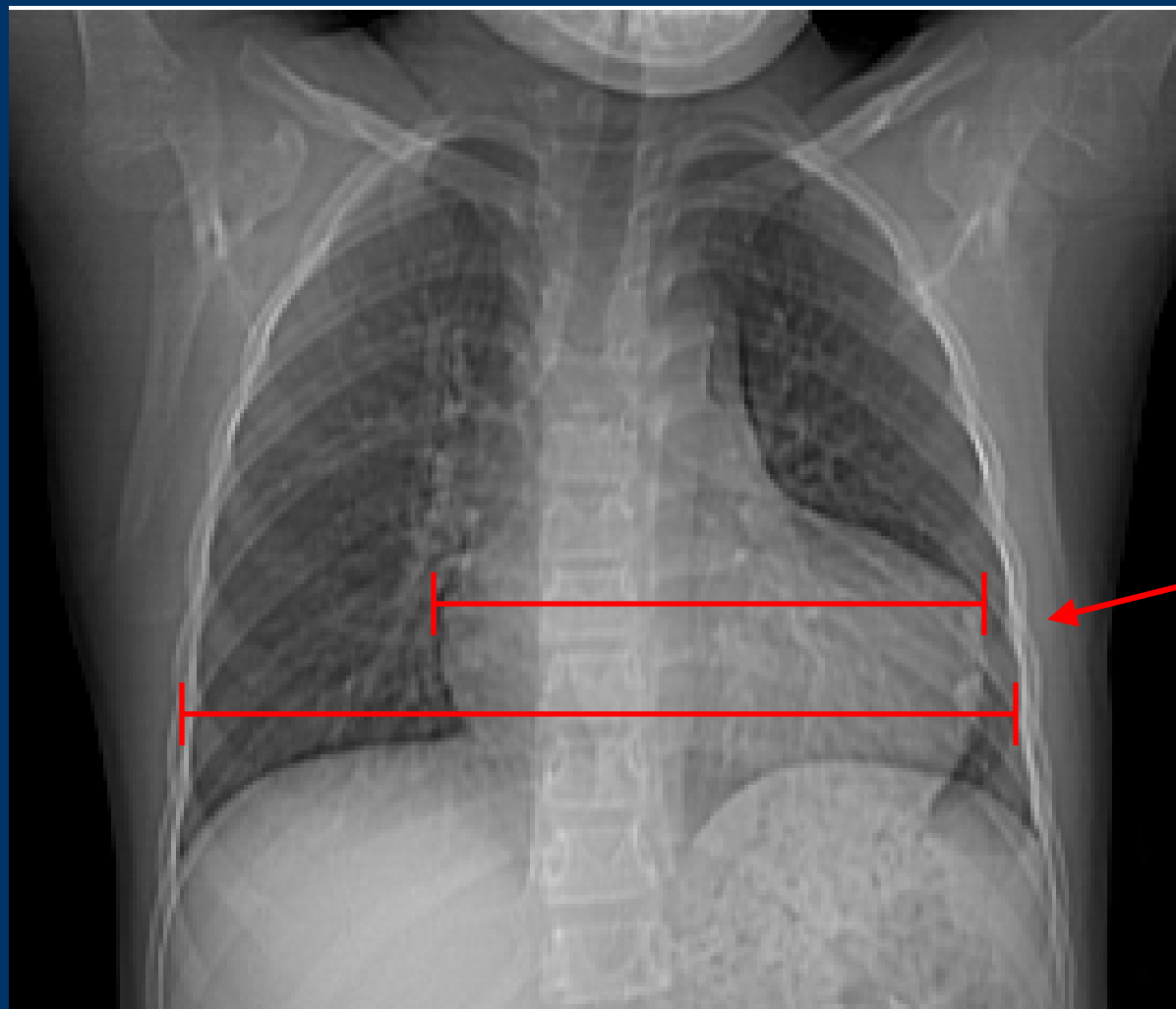


Normal Chest X-Ray





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

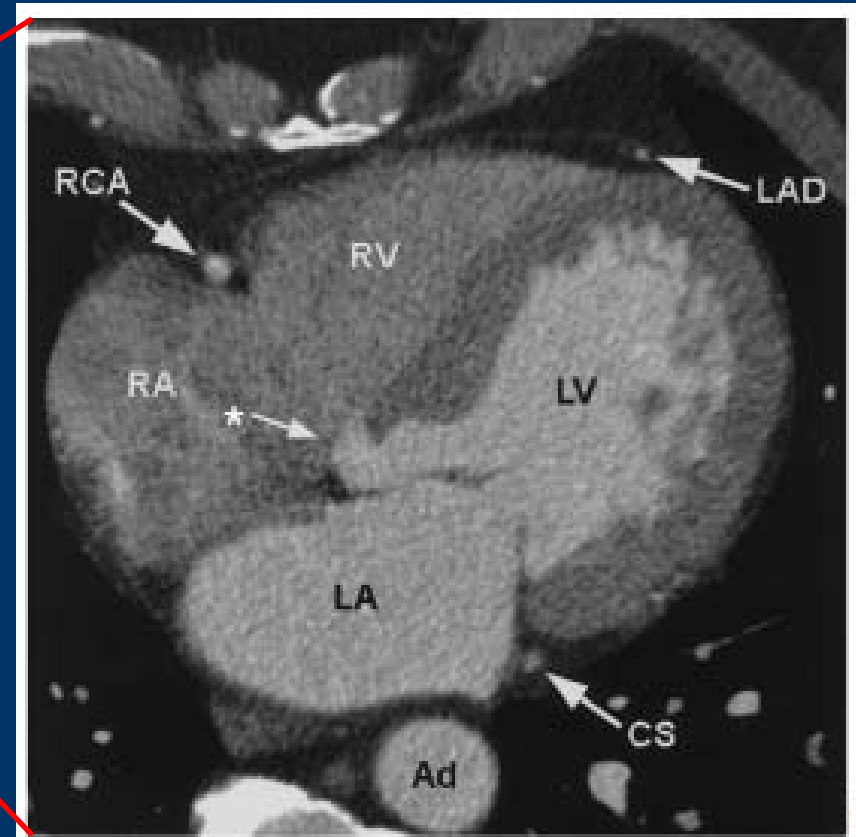
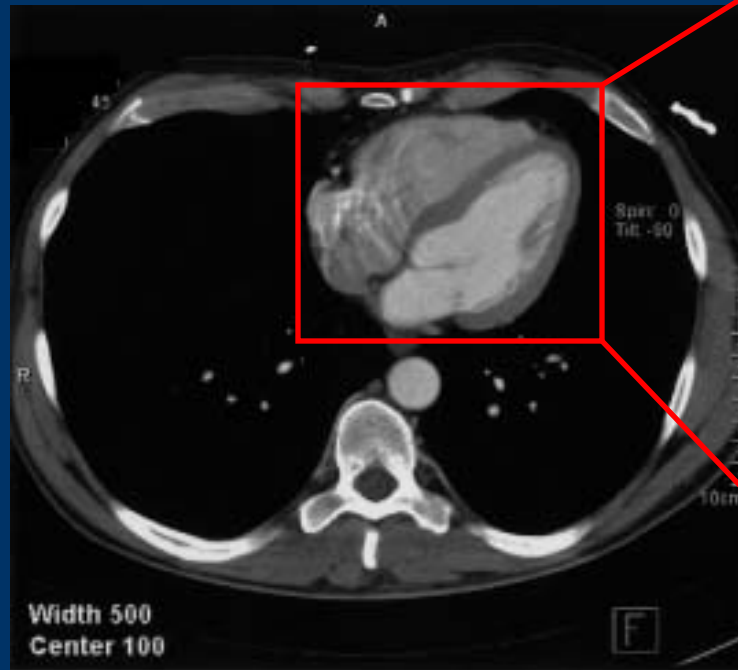


CT - Limitations

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

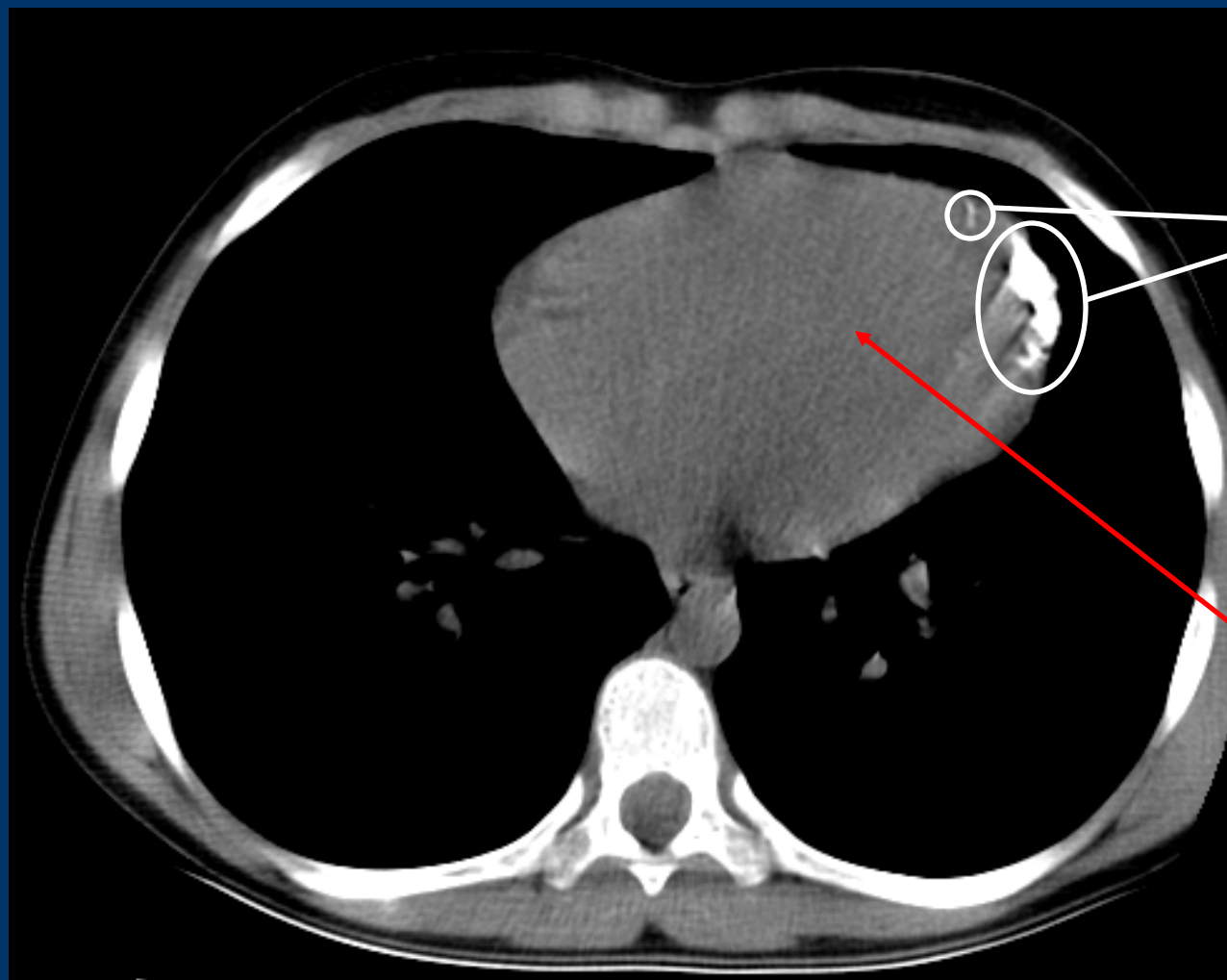


Computerized Tomography - Normals





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 7th 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

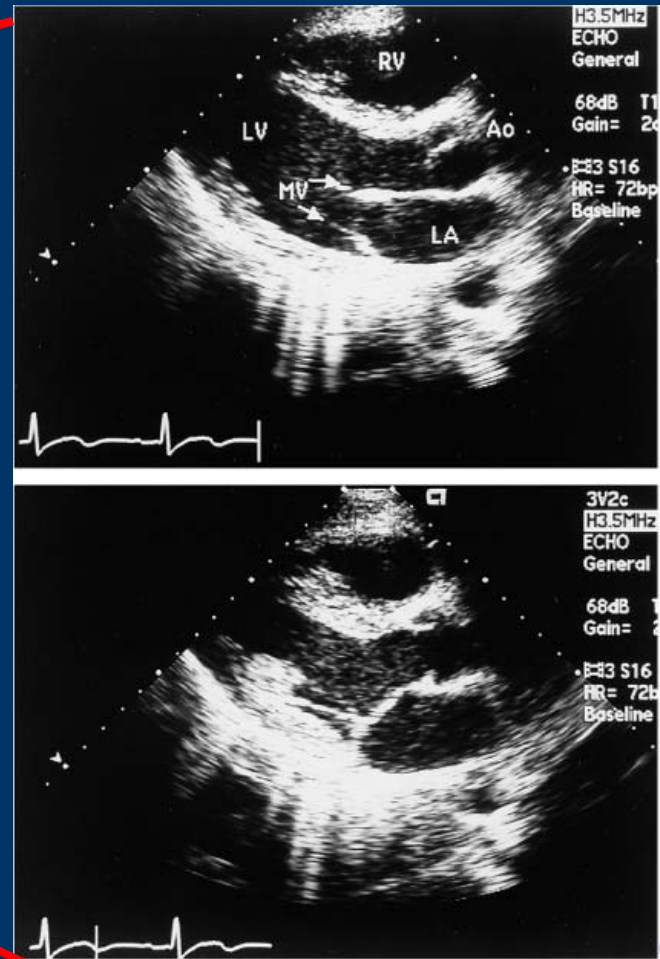
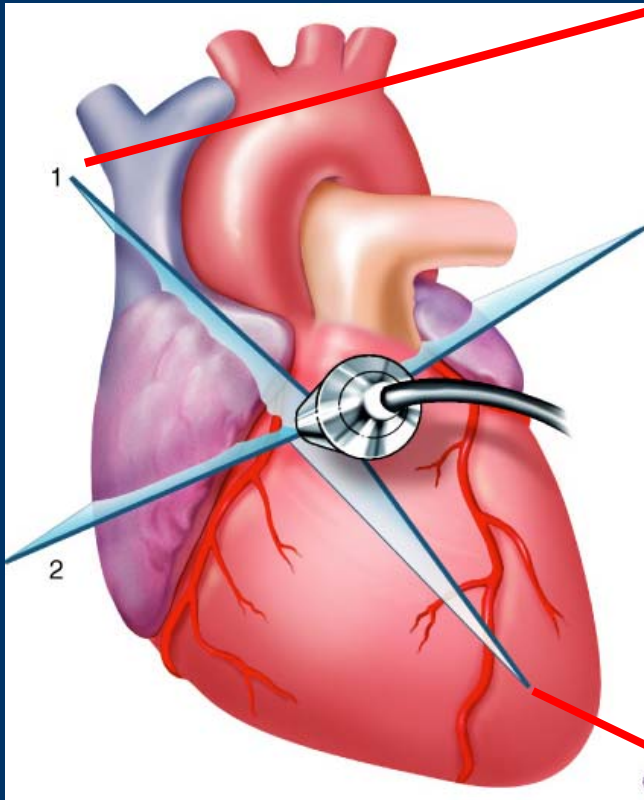


Echocardiography - Limitations

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



TTE - Normal

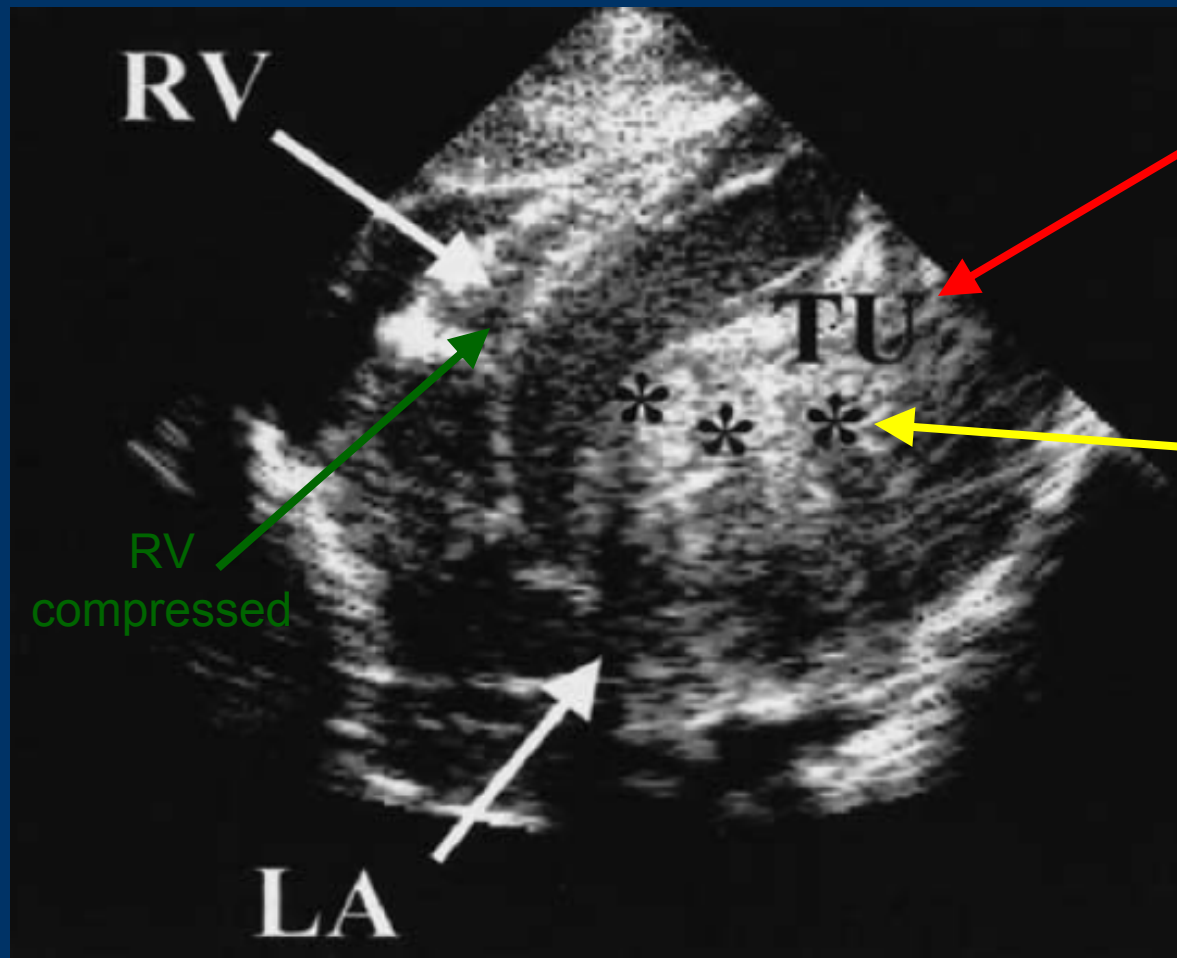


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TTE, 4 chamber view – Companion Patient #1



Hyperechogenic mass filling almost entire LV

Small central calcifications, with acoustic shadowing

Gutberlet, et al. *European Radiology* 2002



Differential Diagnosis

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- Cardiac Mass



Which Mass is it?

Benign

Braunwald, *Heart Disease 7th ed.*; Sparrow, *Radiographics 2005*

<u>Tumor</u>	<u>General</u>	<u>CT</u>	<u>Echo</u>
Rhabdomyoma	Multiple, small, Intramural masses	Homogenous, Low attenuation ✓	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



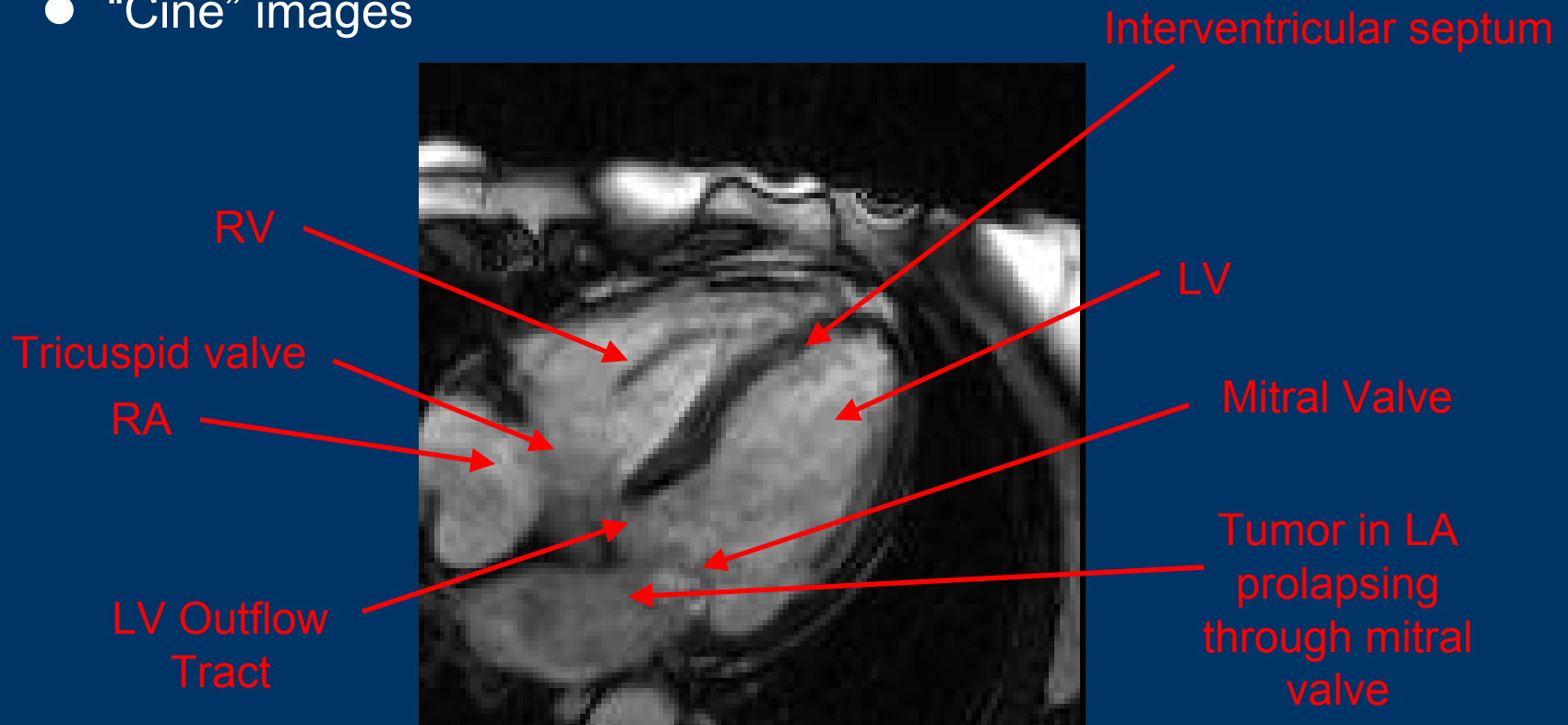
MRI - Limitations

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



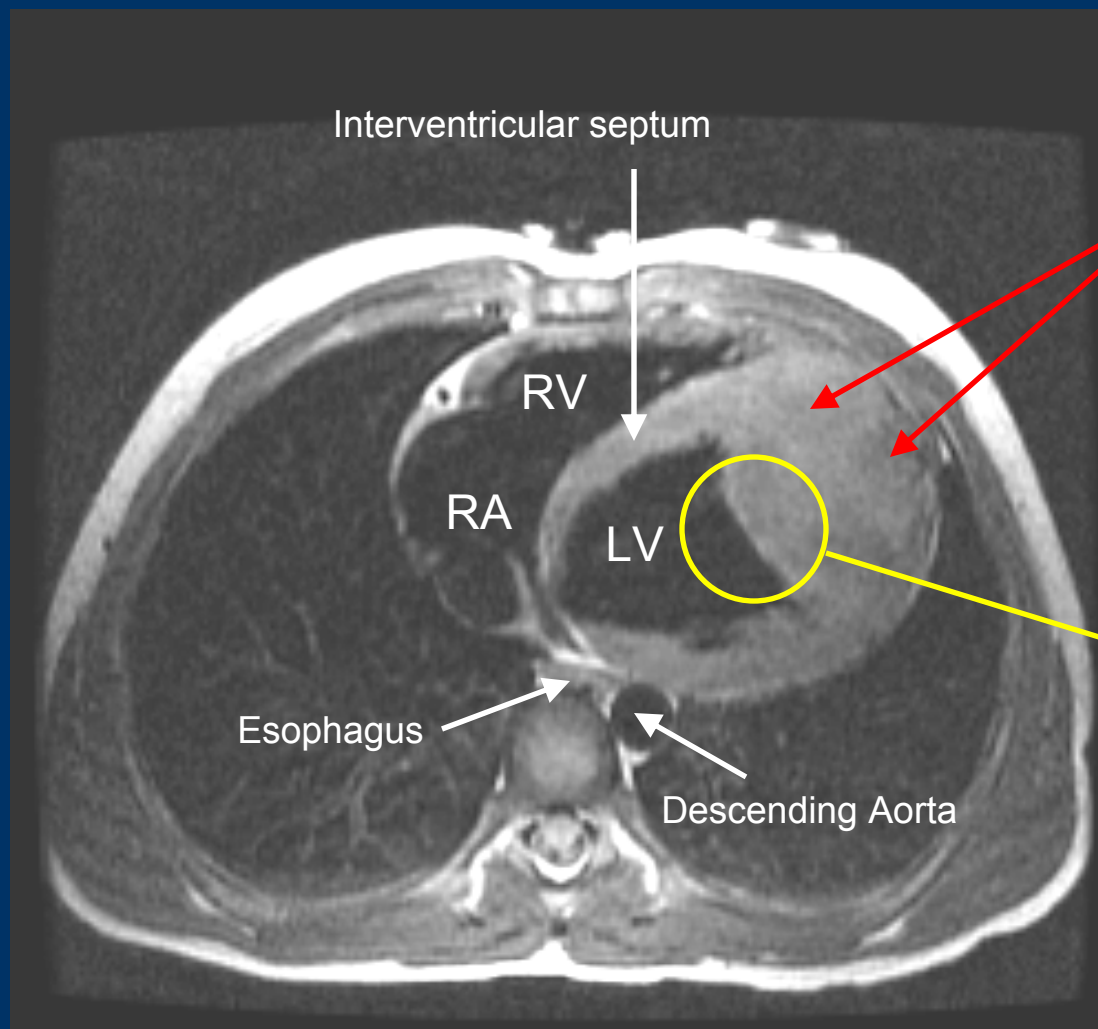
Cardiac MRI

- 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.

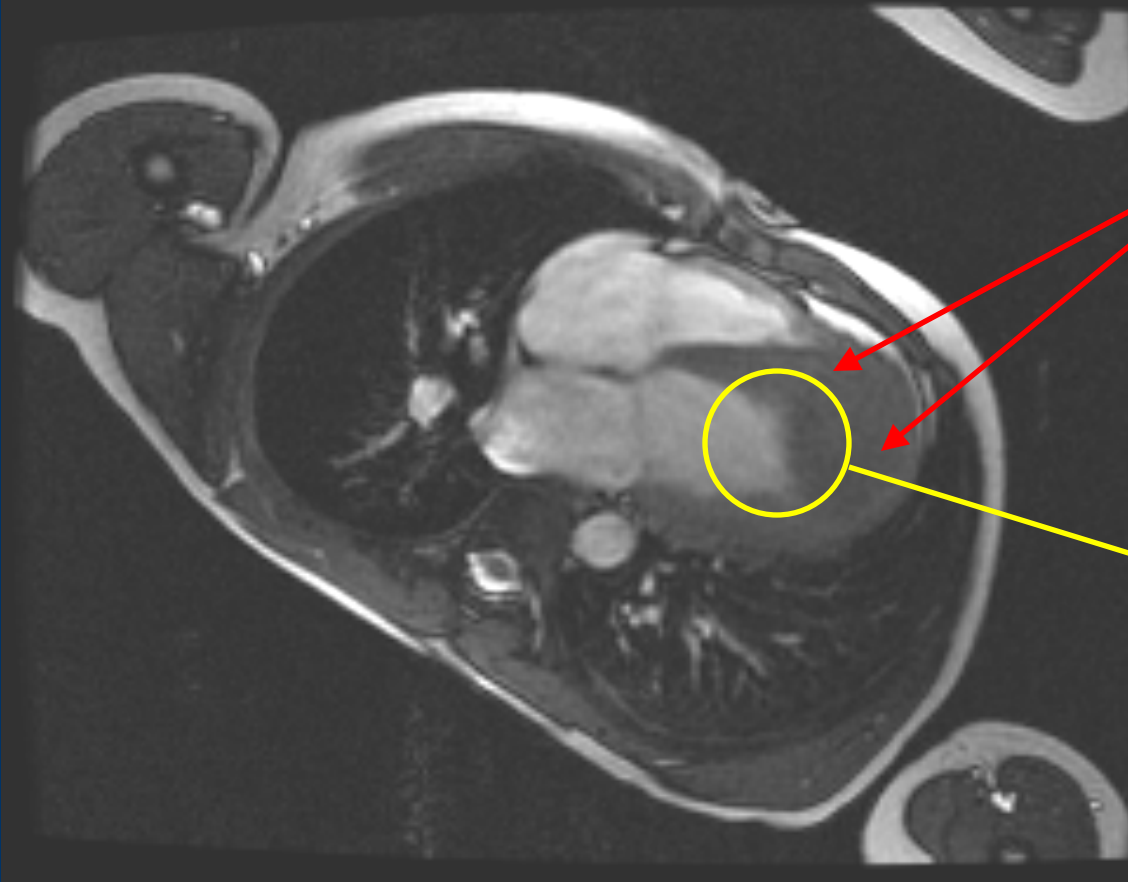


Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration

Endocardium



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

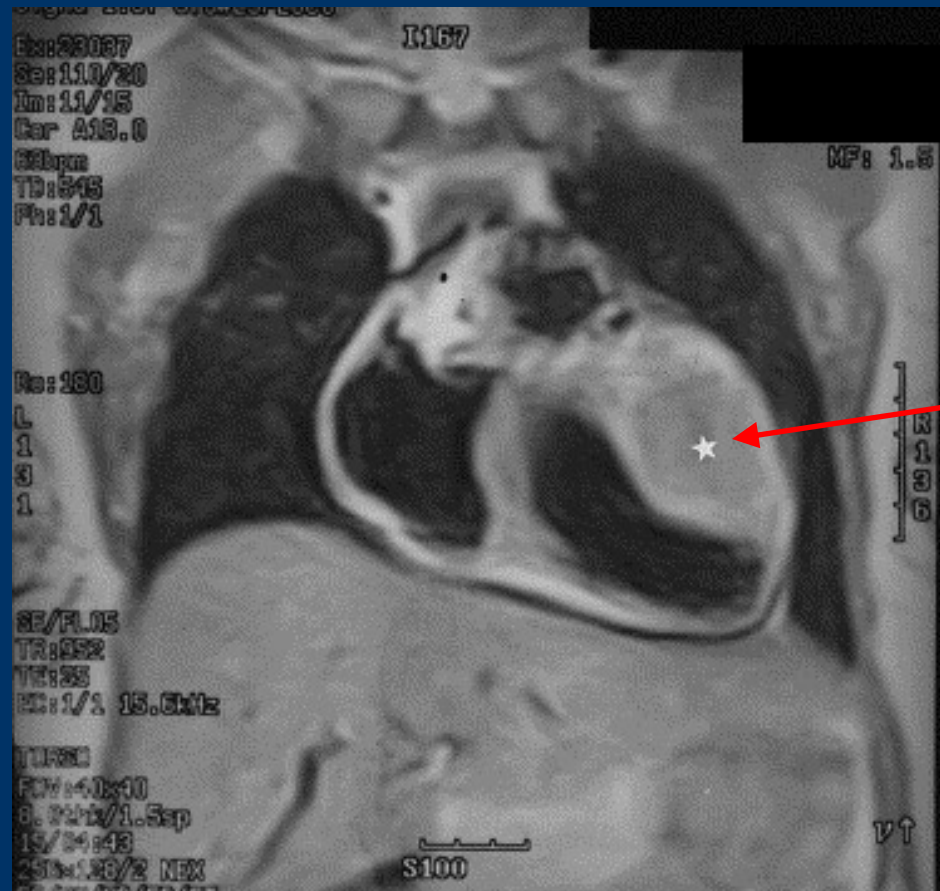


Large, homogenous, isointense mass arising from apex of left ventricle. Well-defined, sharp borders with no pericardial infiltration

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 7th ed.*; Sparrow, *Radiographics 2005*

Malignant

Rhabdomyosarcoma
Fibrosarcoma

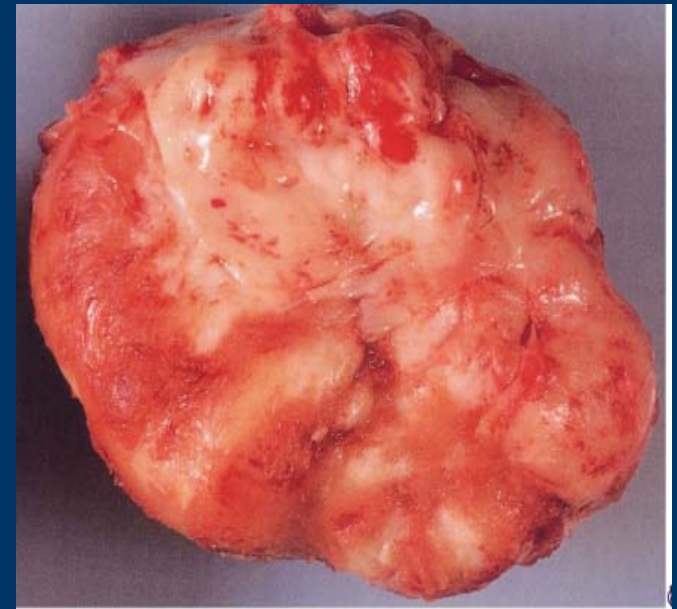
Other

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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



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