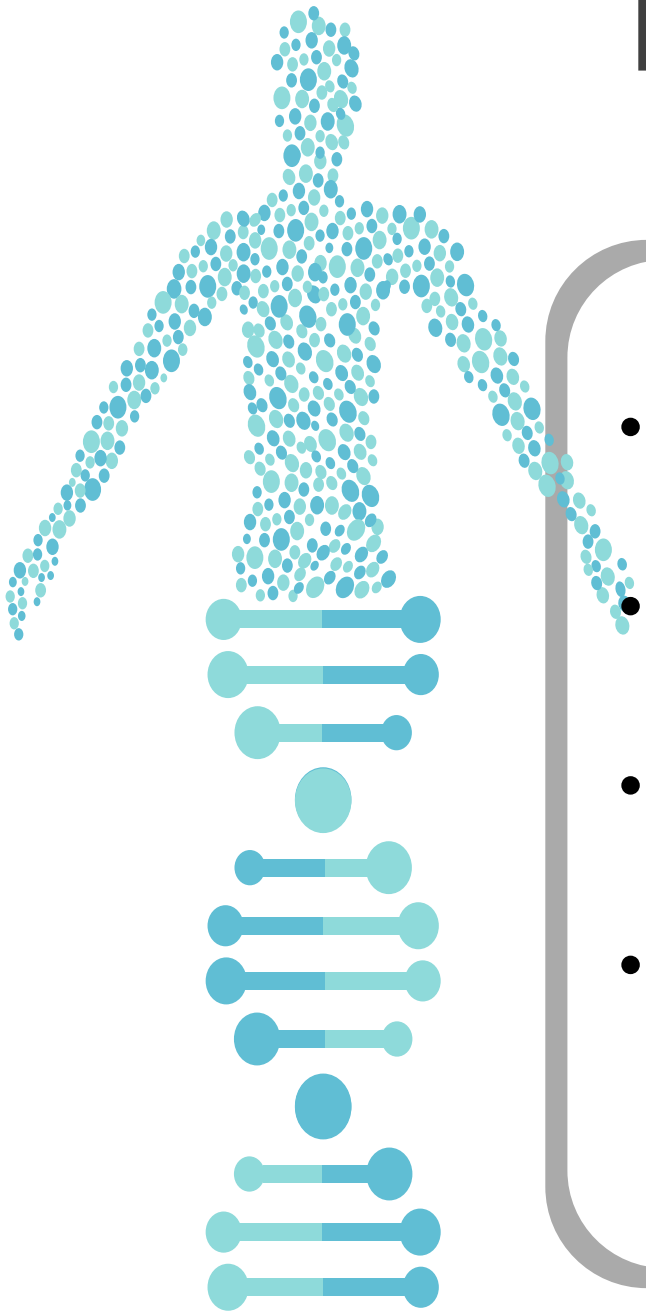
Unveiling the Enigma: Left Atrial Mass in Echocardiography

Mutaz Karameh M.D

Heart institute, Hadassah Medical Center, Hebrew university

N.V



- A 52-year-old male patient
- No past medical history other than active smoking.
- No family history of cardiovascular diseases.
- No chronic medications.

Clinical picture

- 6 months of general weakness, knee and ankle pain.
- No chest pain or shortness of breath.
- No fever, no night sweats, no loss of weight.

- Physical examinations - Clubbing.

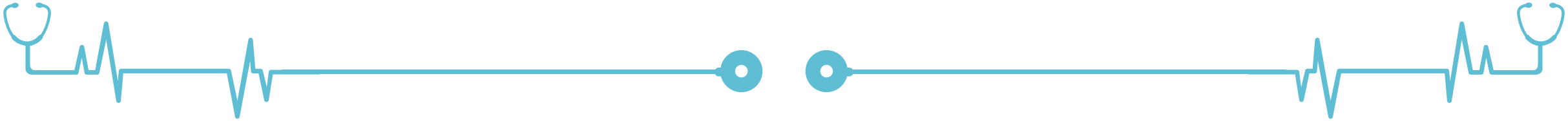
- Rheumatological workup – No findings.

- Labs - CRP 14 mg%.

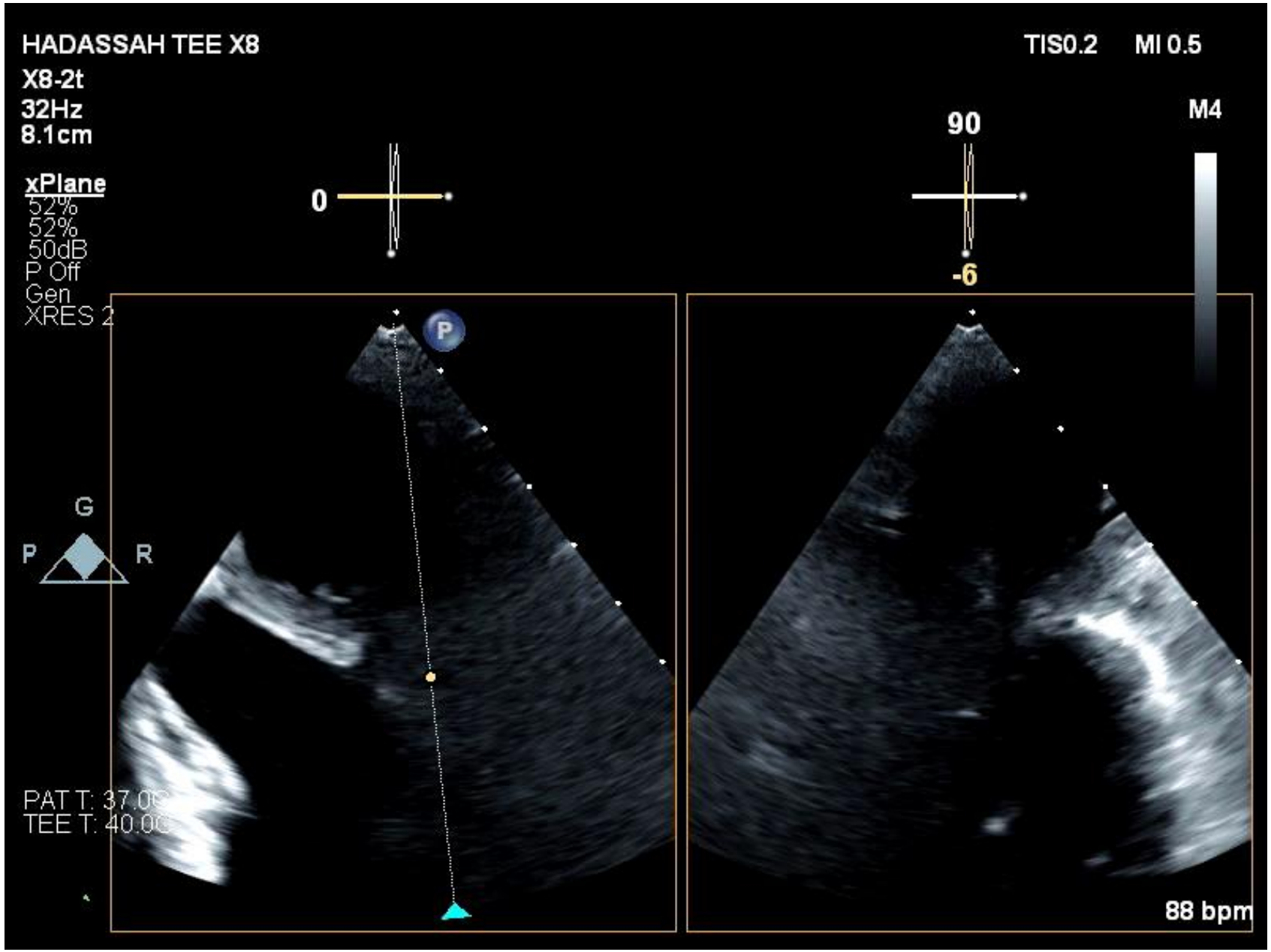
- Colonoscopy normal.

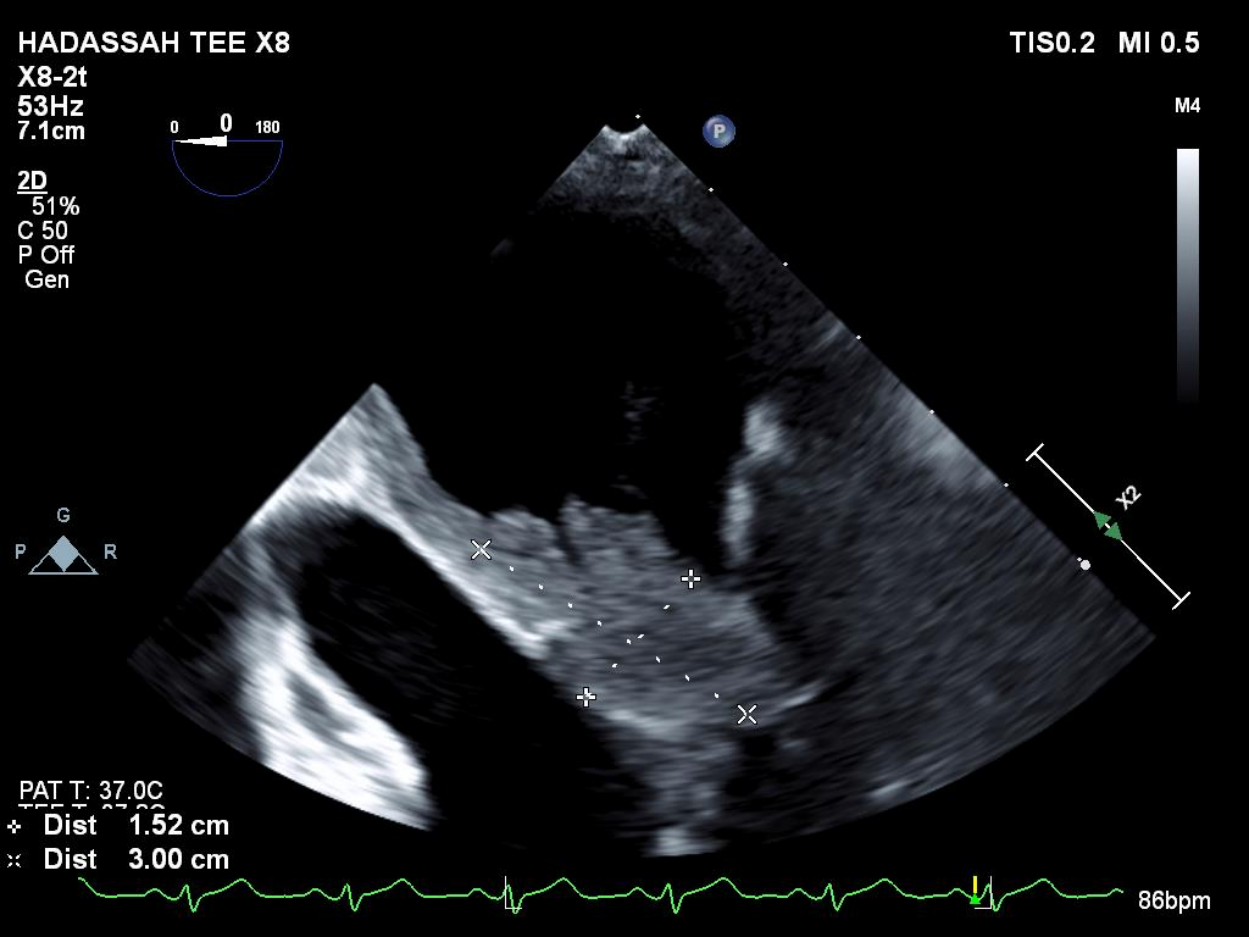
- CT chest abdominal and pelvis – normal ??

- TTE – echogenic structure in the left atrium



Transesophageal Echocardiogram

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X8-2t
53Hz
7.1cm
0 0 180
M4
2D
51%
C 50
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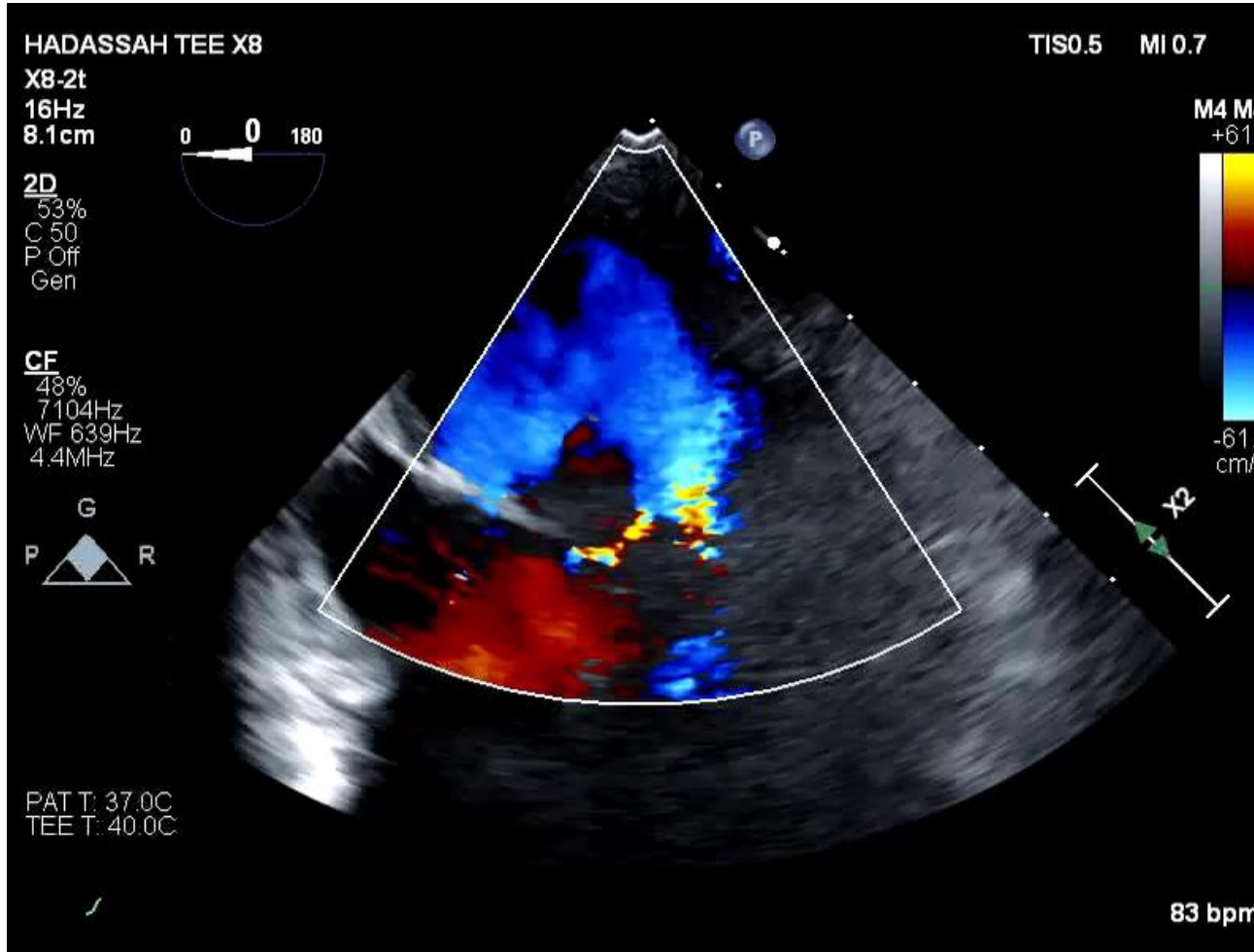
Gen
G
P R
X2
PAT T: 37.0C
Dist 1.52 cm
Dist 3.00 cm
86bpm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+61
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C 50
P Off
Gen
CF
48%
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WF 639Hz
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G
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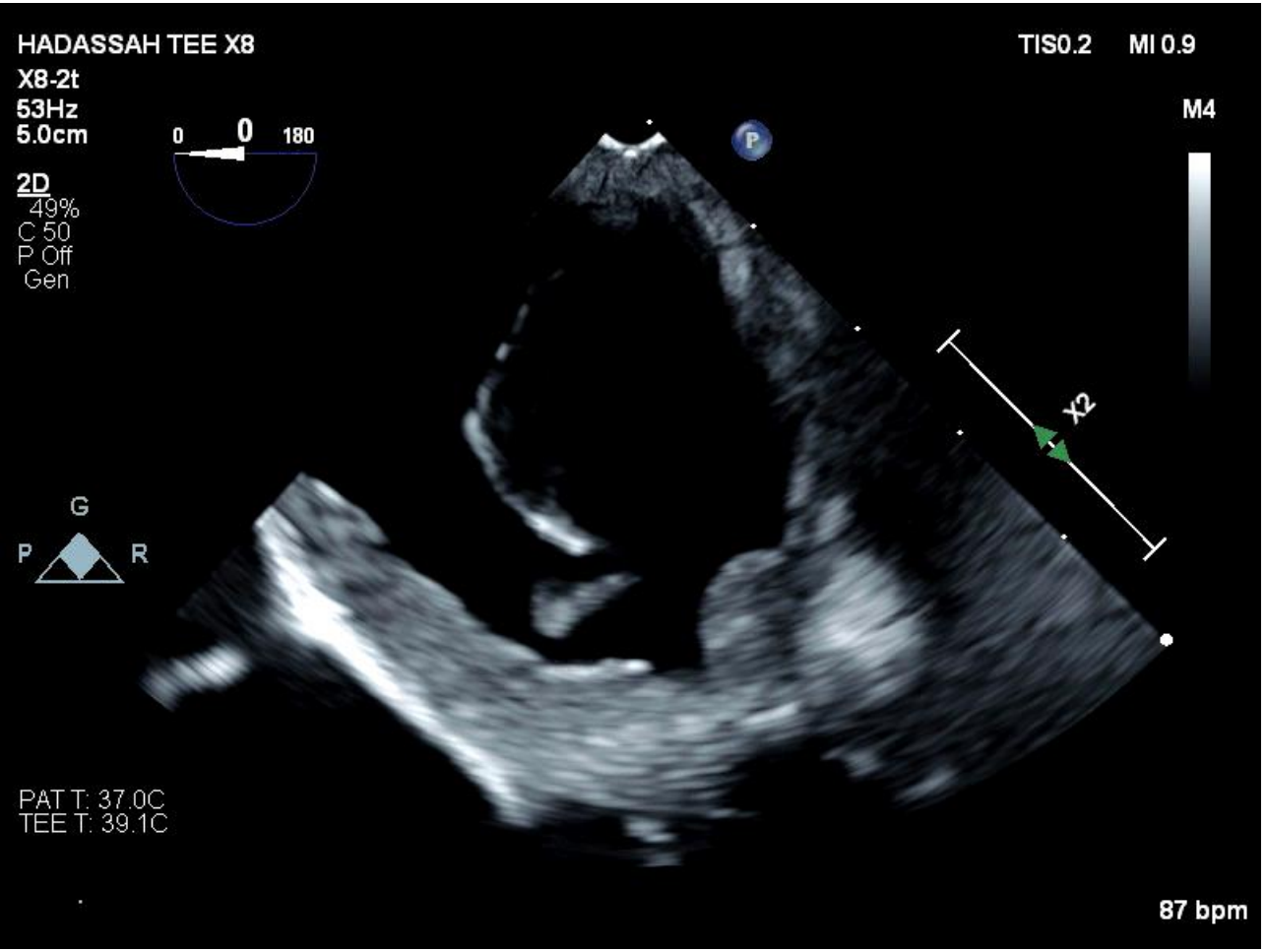
cm/
X2
PAT T: 37.0C
TEE T: 40.0C
83 bpm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X2
G
P R
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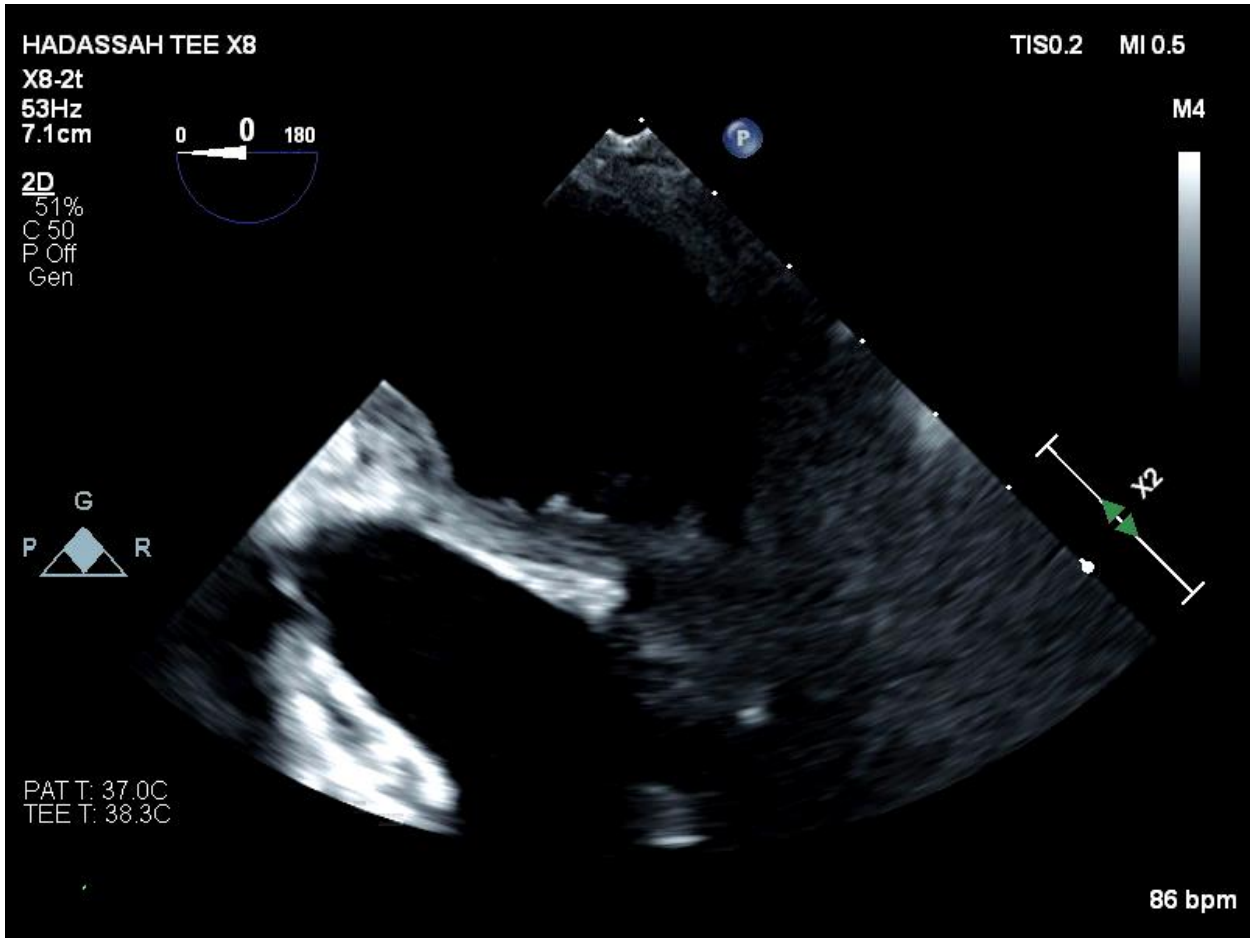
TEE T: 39.1C
87 bpm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M4
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X2
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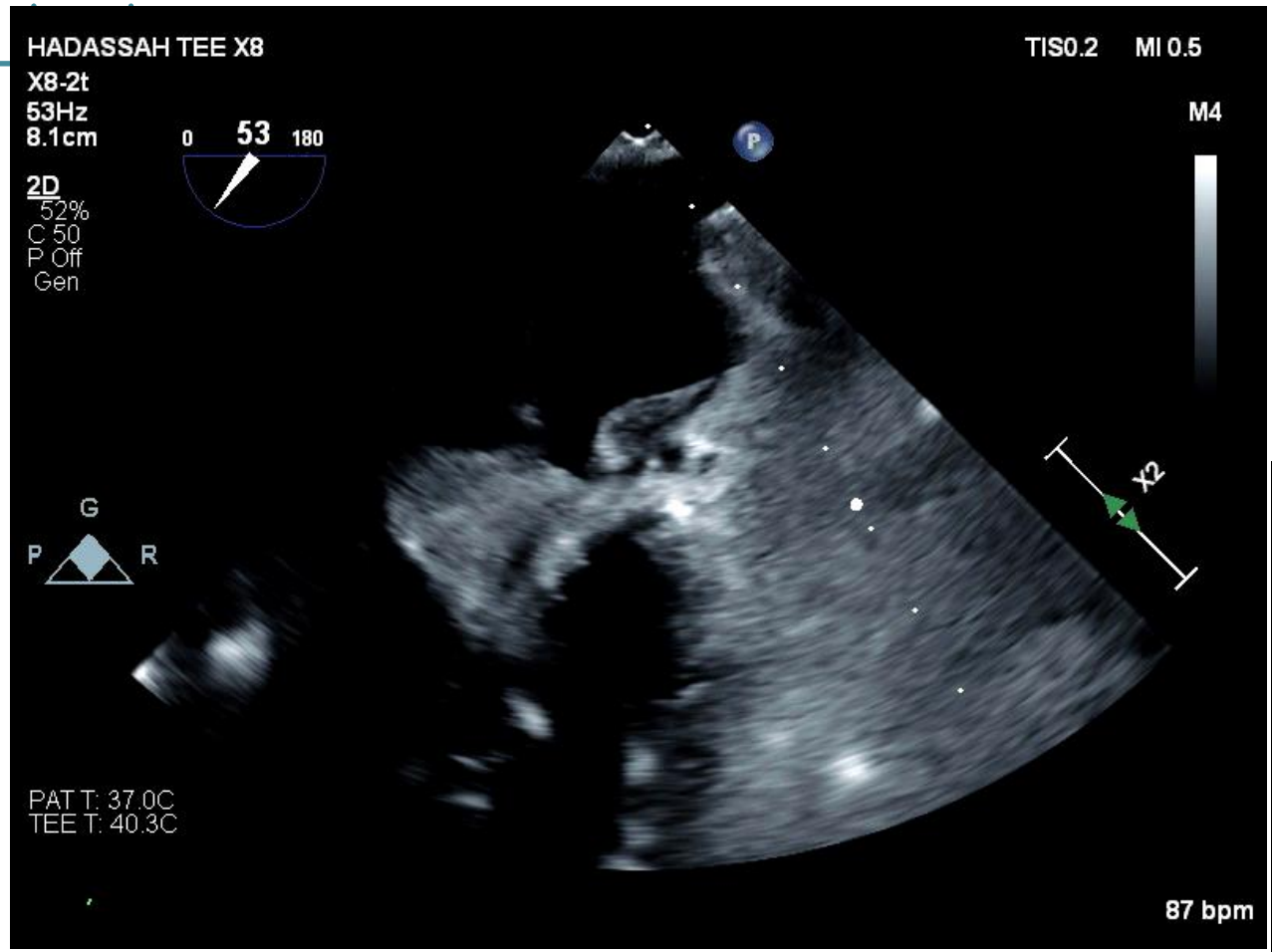
PAT T: 37.0C
TEE T: 38.3C
86 bpm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8.1cm
0 53 180
M4
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52%
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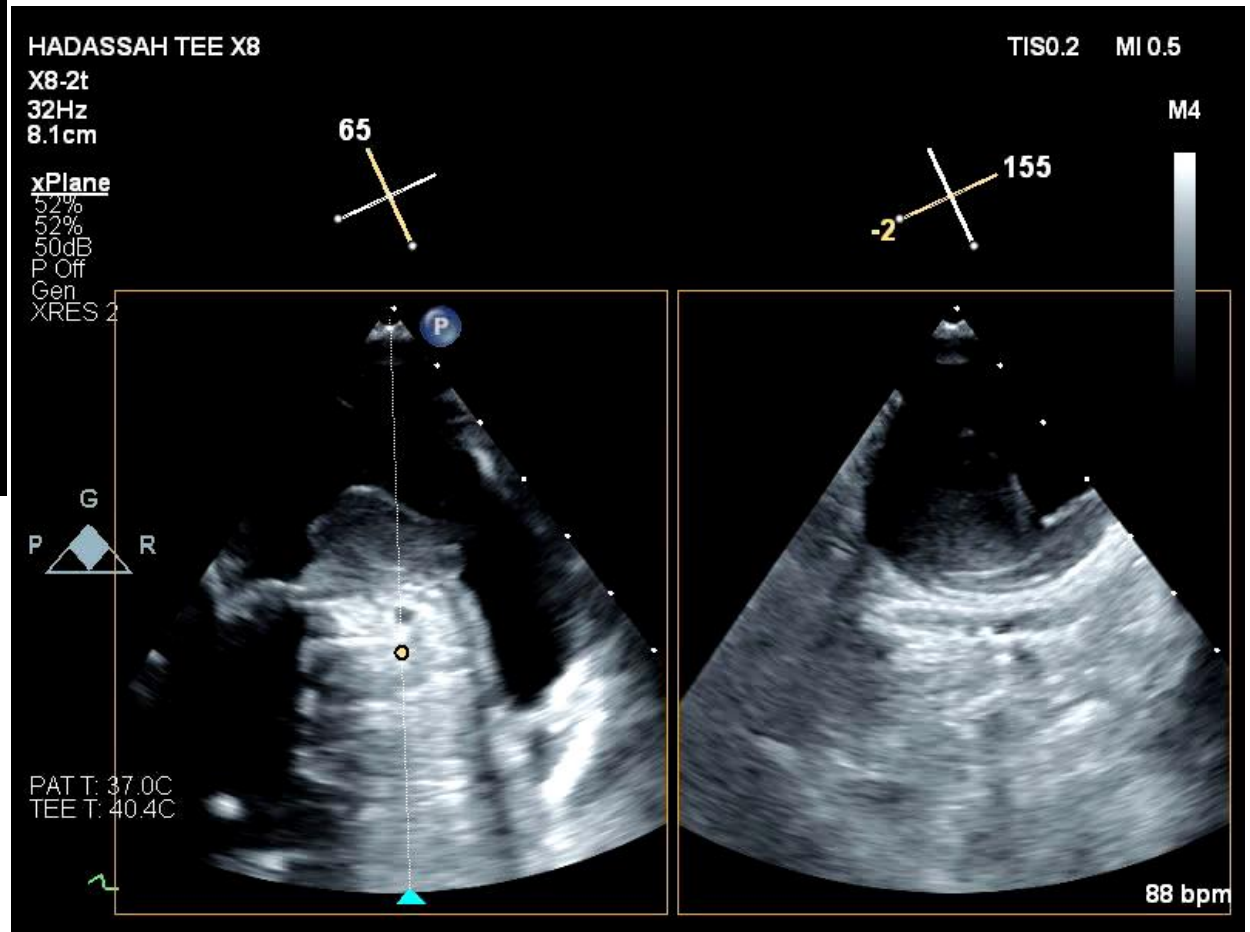
P R
PAT T: 37.0C
TEE T: 40.3C
87 bpm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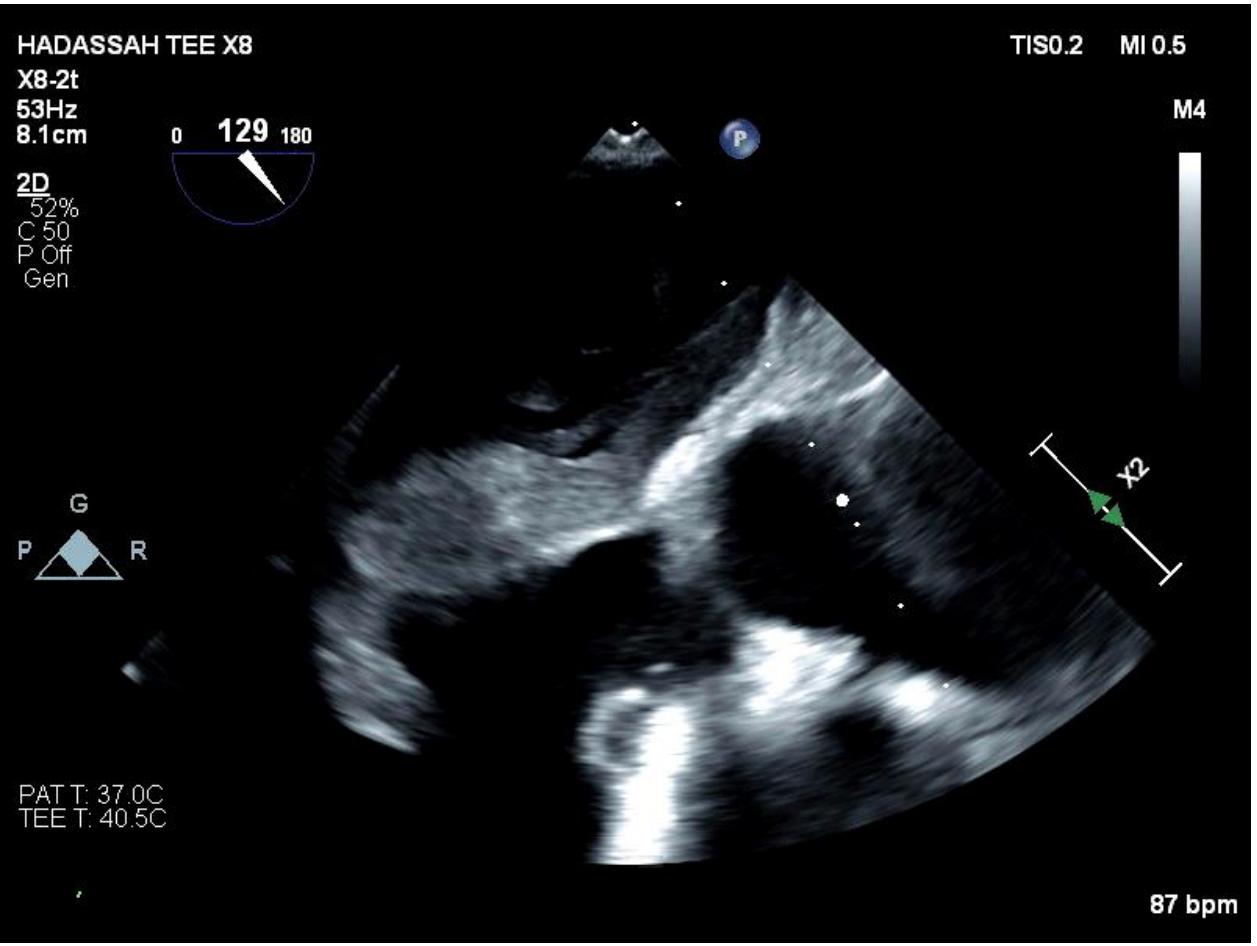
M4
65
155
-2
xPlane
52%
52%
50dB
P Off
Gen
XRES 2
G
P R
PAT T: 37.0C
TEE T: 40.4C
88 bpm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X8-2t
53Hz
8.1cm
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Gen
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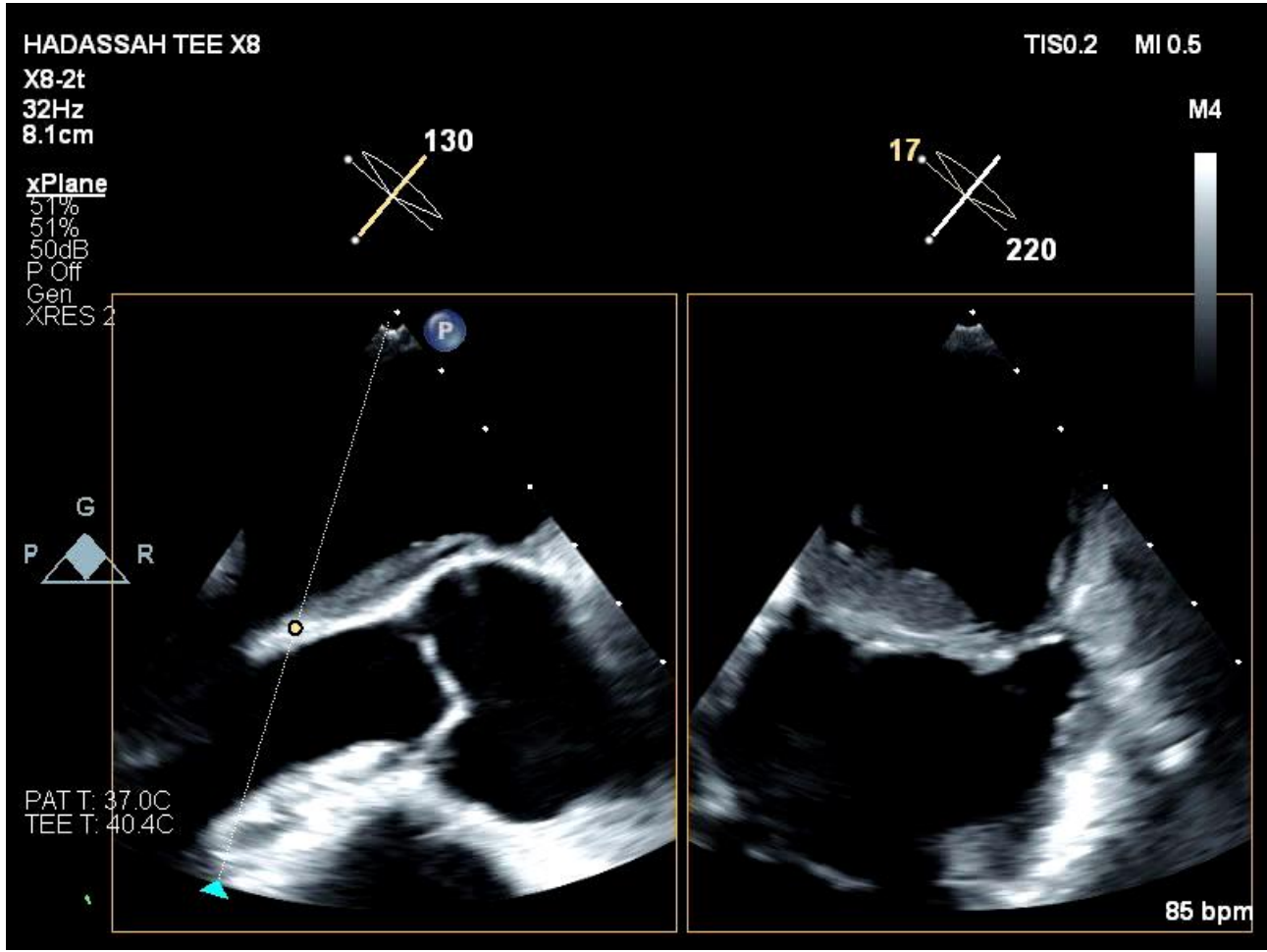
PAT T: 37.0C
TEE T: 40.5C
87 bpm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M4
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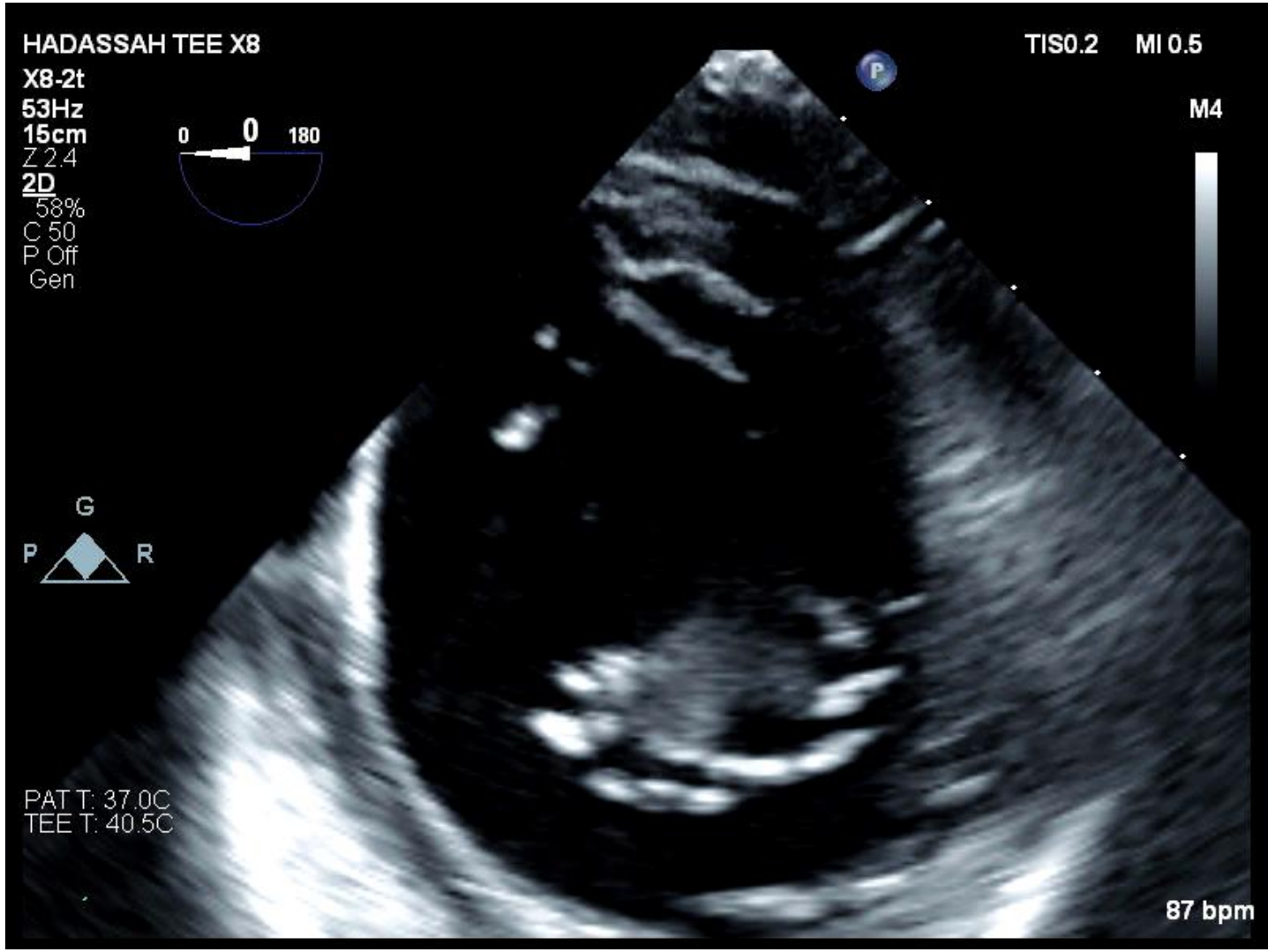
17
220
xPlane
51%
51%
50dB
P Off
Gen
XRES 2
G
P R
PAT T: 37.0C
TEE T: 40.4C
85 bpm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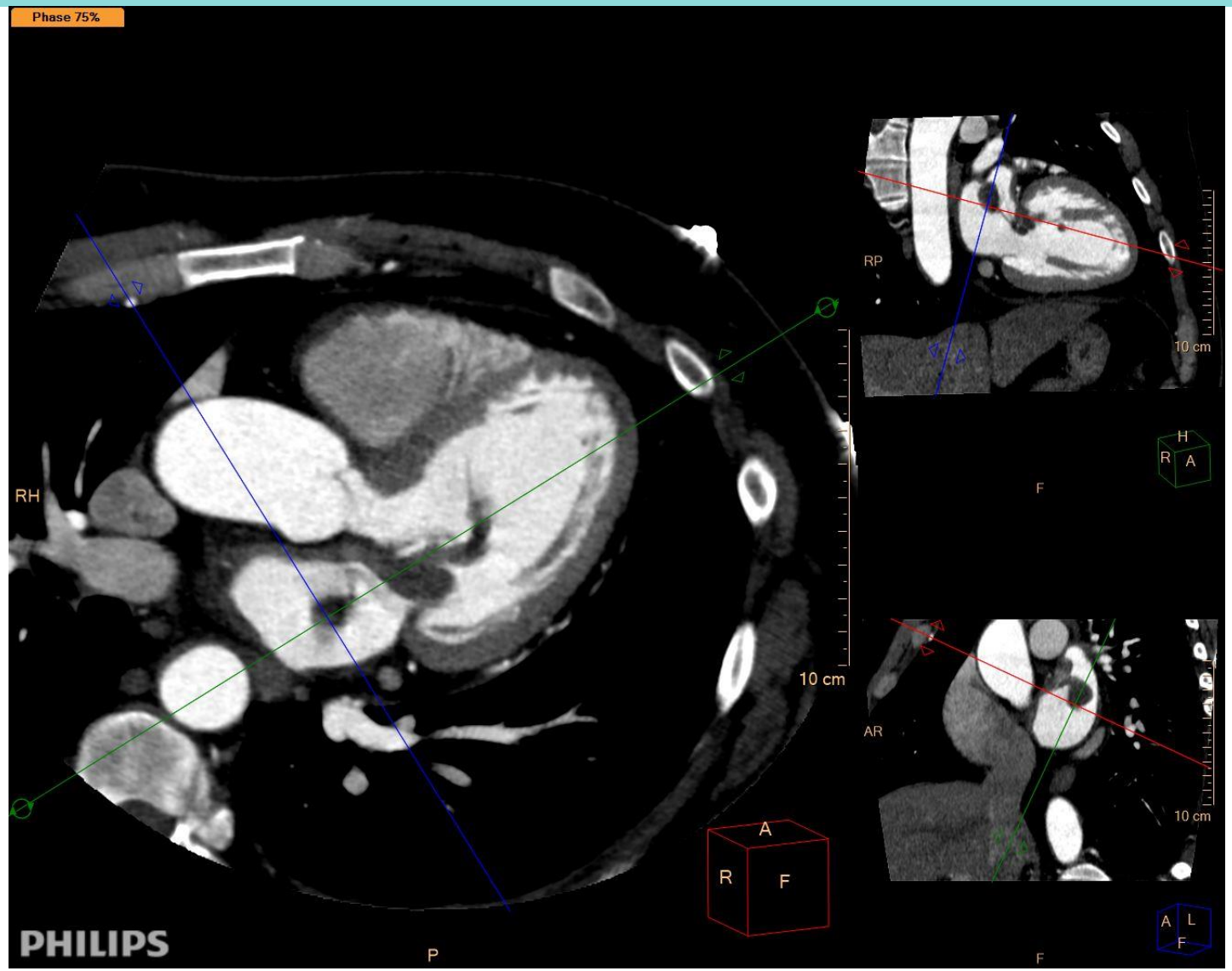
53Hz
15cm
Z 2.4
2D
58%
C 50
P Off
Gen
0
0
180
TIS0.2
MI 0.5
M4
G
P
R
PAT T: 37.0C
TEE T: 40.5C
87 bpm

CT



Pathology

Intimal Sarcoma

Hypercellular malignant tumor composed of partly spindled, partly epithelioid cells embedded in a fibromyxoid and fibroinflammatory background with scattered neutrophils and plasma cells. In some areas, the tumor cells show severe nuclear atypia and pleomorphism with multilobulation, large irregular nuclei, and prominent nucleoli.

No obvious heterologous elements are identified. The tumor is present at the edges of the specimen.

On immunostains, tumour cells are partly positive for CD34. They are negative for CAM5.2, AE1/AE3, S100, SOX10, SALL4, SMA, desmin, TFE, calretinin, D2-40, and CKIT. CD31 and ERG are positive in blood vessels within the tumor.

The KI67 is positive in about 10% of the cells.

FISH study showing MDM2 amplification

Intimal sarcoma (IS)

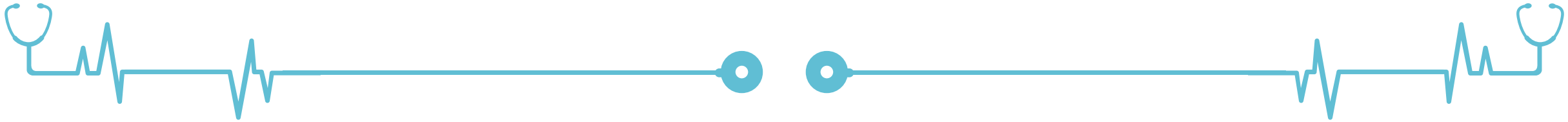
Introduction

- Primary cardiac tumors are rare, with an incidence of 0.001–0.03% based on autopsy results, of which approximately 25% are malignant
- 95% of these are sarcomas; the remaining 5% are lymphomas.
- Angiosarcomas are the most common type of primary cardiac sarcoma (about 37%) followed by undifferentiated sarcoma (24%), malignant fibrous histiocytoma (11–24%), leiomyosarcoma (8–9%).
- The rarest type of primary malignant cardiac sarcoma is intimal (spindle cell) sarcoma

Butany J, et al. Cardiac tumours: diagnosis and management. Lancet Oncol. 2005 Apr;6(4):219-28.

Introduction

- Malignant mesenchymal tumors that originate from the tunica intima of larger vessels, and they rarely involve the heart
- Very aggressive and have a poor prognosis. the recurrence rate is high
- Estimated median survival rate of only 3–13 months due to rapid growth and early metastasis

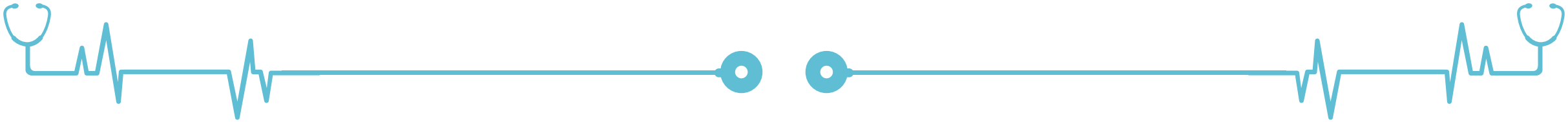


Clinical manifestation

- They usually present between the third and fifth decade of life.
- The most common symptoms are usually nonspecific, including dyspnea, chest or back pain, coughing, and hemoptysis.
- Additional symptoms such as weight loss, fever, or anemia further increase the suspicion of malignancy.

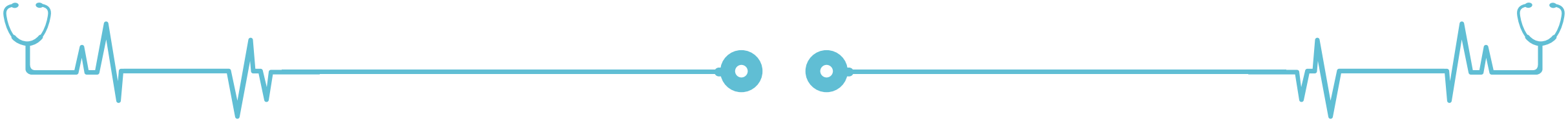
Diagnosis

A multimodal approach using various imaging modalities such as echocardiography, Computed Tomography (CT), PET-CT, and CMR to visualize the heart, thorax, and vascular structures, is the best method for rapid diagnosis of IS.



Diagnosis

- ✓ Transthoracic echocardiography and TEE are primarily used to determine the location of the mass, its relationship with surrounding tissues, and the presence of invasion.
- ✓ CT is more sensitive in assessing the size and extent of the mass, especially in identifying IS located in large arteries
- ✓ CMR enables accurate positioning of the tumor, assessment of the extent of involvement, evaluation of the functional impact, and tissue characterization of the lesion. CMR helps distinguish neoplastic masses from thrombi and provides appropriate visualization of vasculature and tissue edema
- ✓ FDG uptake on PET-CT is positive for malignant tumors such as IS and negative for thrombi.



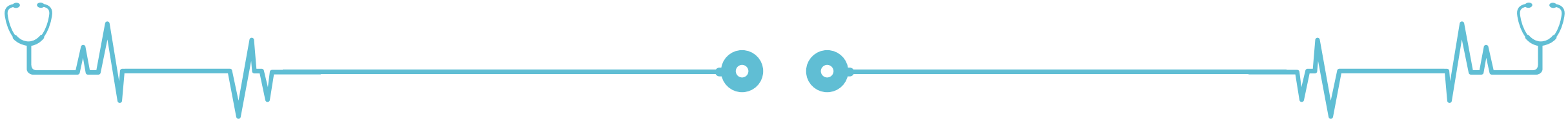
Diagnosis

The definitive diagnosis is made through pathology, with MDM2 gene amplification observed in the majority of the patients.

Management

There is no standard treatment for the management of patients with IS; it usually involves a multidisciplinary approach including surgery, radiation, chemotherapy, and targeted therapy.

Approach to cardiac mass



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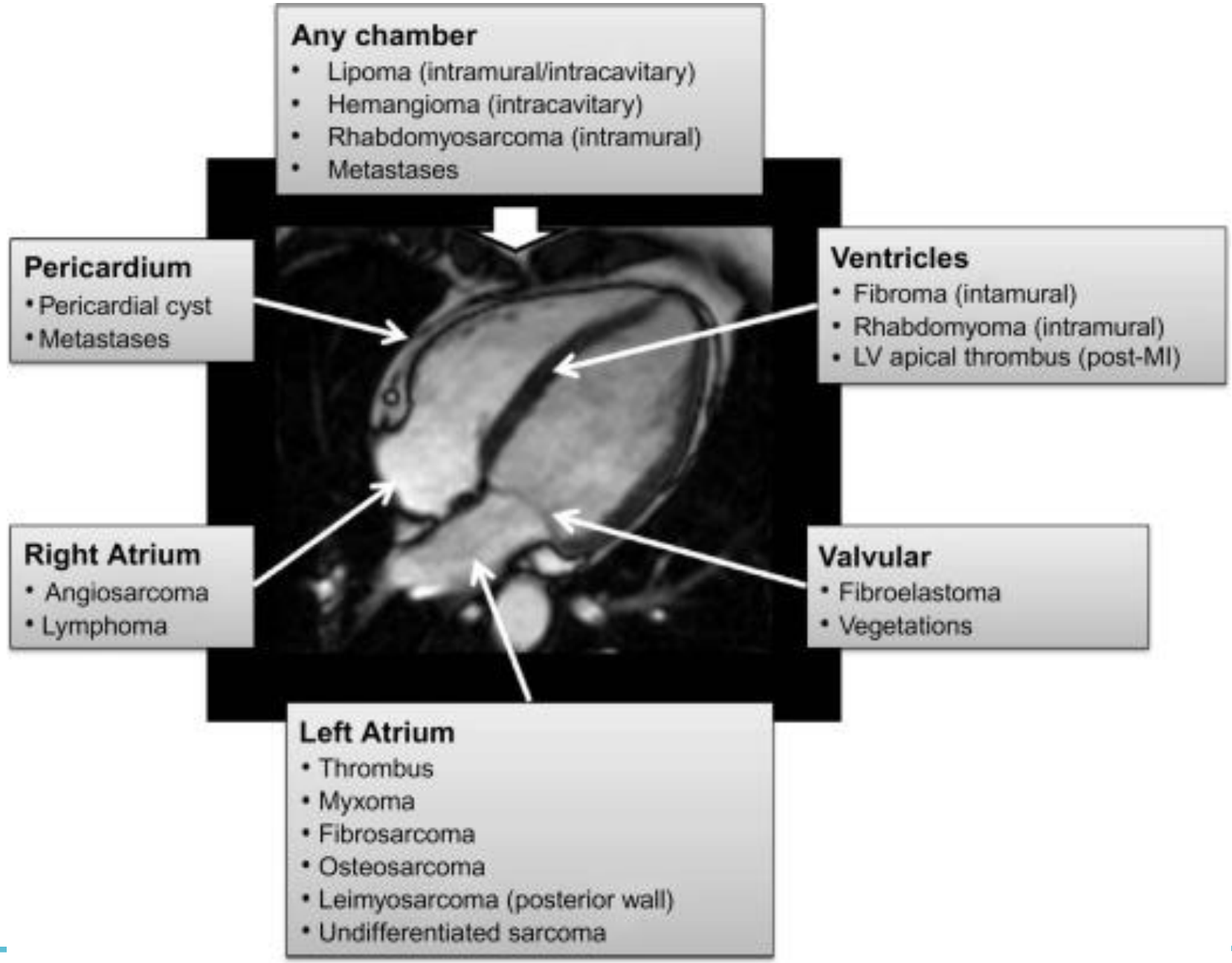

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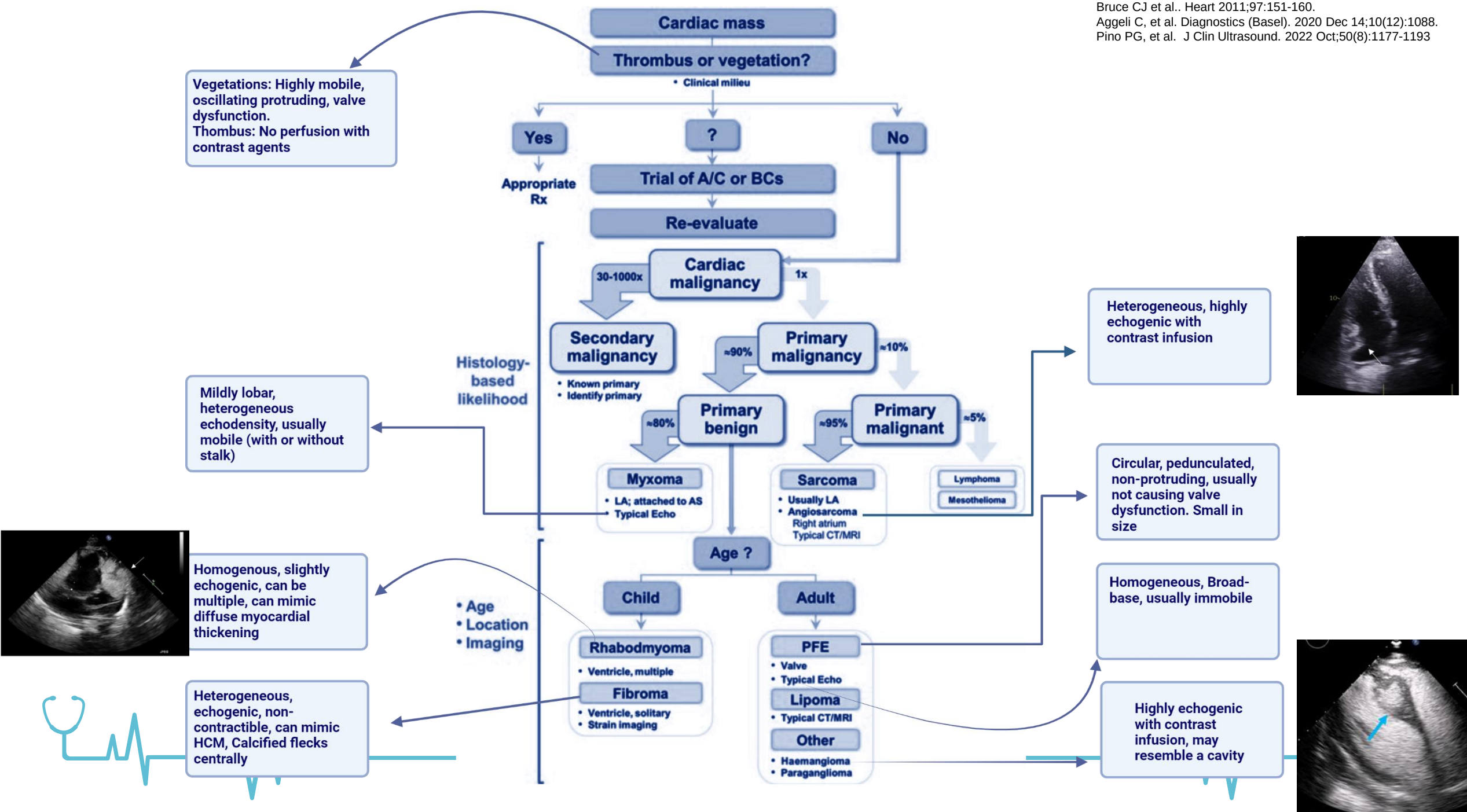

	Age of presentation	Common location	Echo features
Benign tumors			
Myxoma	• Early (familial) or middle adulthood	• LA, atrial septum, any other site	• Mildly lobar, heterogeneous echodensity, usually mobile (with or without stalk)
Fibroelastoma	• Middle or late adulthood	• Cardiac valves	• Circular, pedunculated, non-protruding, usually not causing valve dysfunction. Small in size
Lipoma	• Adulthood	• Left ventricle, any other site	• Homogeneous, Broad-base, usually immobile
Rhabdomyoma	• Infancy or early childhood	• Ventricles, atrioventricular valves	• Homogenous, slightly echogenic, can be multiple, can mimic diffuse myocardial thickening
Fibroma	• Early childhood	• Intraventricular septum, ventricles	• Heterogeneous, echogenic, non-contractible, can mimic HCM, Calcified flecks centrally
Hemangioma	• Any age	• Any other site	• Highly echogenic with contrast infusion, may resemble a cavity
Malignant primary heart tumors			
Angiosarcoma	• Early and middle adulthood	• RA, pericardium	• Heterogeneous, highly echogenic with contrast infusion
Rhabomyosarcoma	• Childhood, early adulthood	• Ventricles, any other site	• Normal-high echodensity
Lymphoma	• Adulthood	• RA, any other site	• Any size and shape
intimal sarcoma	• Adulthood	• PA, LA and RA	• Heterogeneous
Non-neoplasmatic heart masses			
Clots	• Adulthood	• LAA, LV apex	• Acute: Low echodensity Chronic: High echodensity. No perfusion with contrast agents
Vegetation	• Adulthood	• Cardiac valves	• Highly mobile, oscillating protruding, valve dysfunction.
Non-neoplasmatic calcified masses	• Adulthood	• Usually posterior mitral annulus	• Very high echodensity

Thank You