Cardio-Oncology Department and Research Center
Rajaie Cardiovascular Medical & Research Center
Tehran, Iran

CARDIOVASCULAR CARE IN PEDIATRIC PATIENTS WITH CARDIAC SARCOMA

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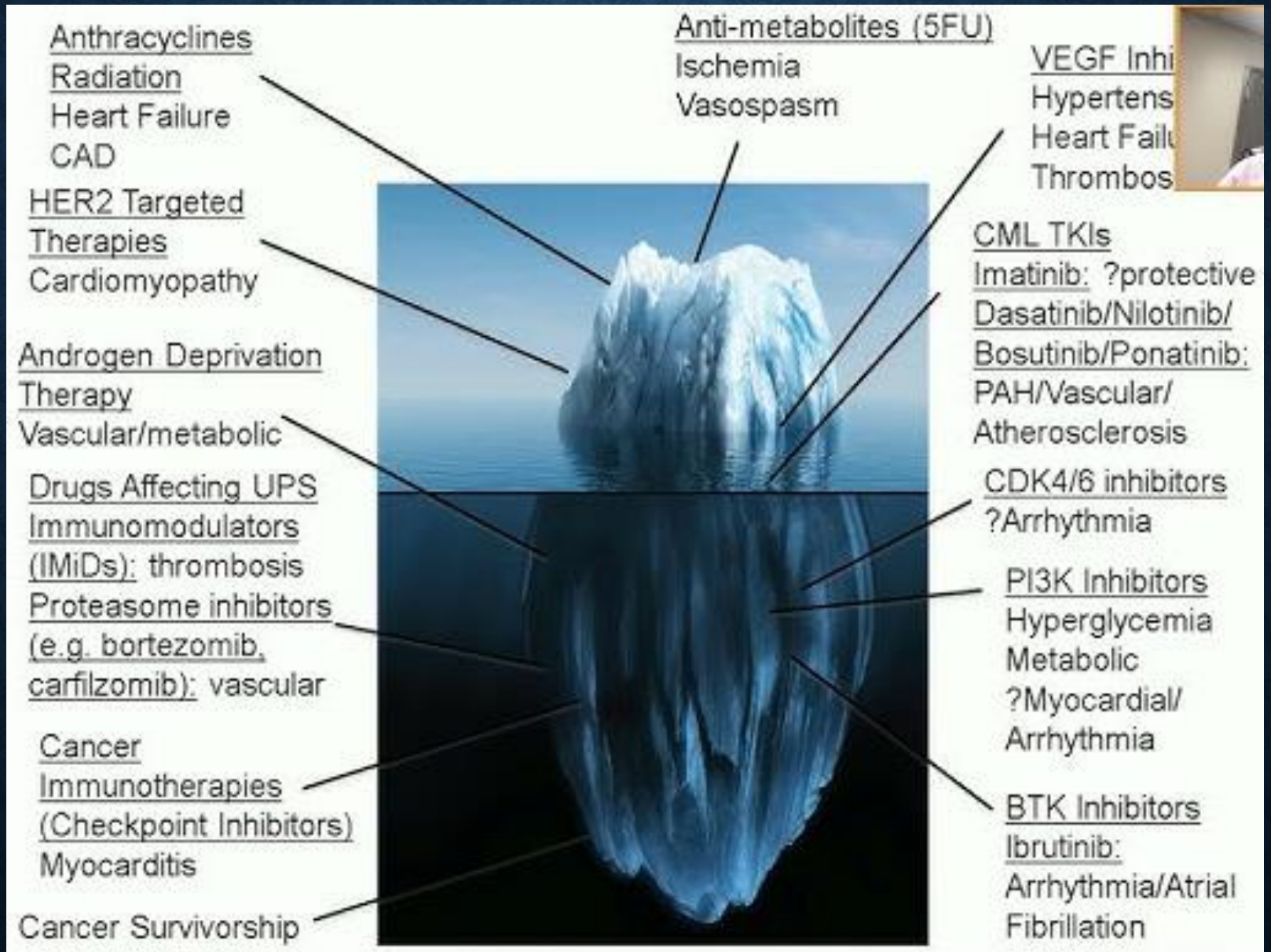
SYNOVIAL SARCOMA

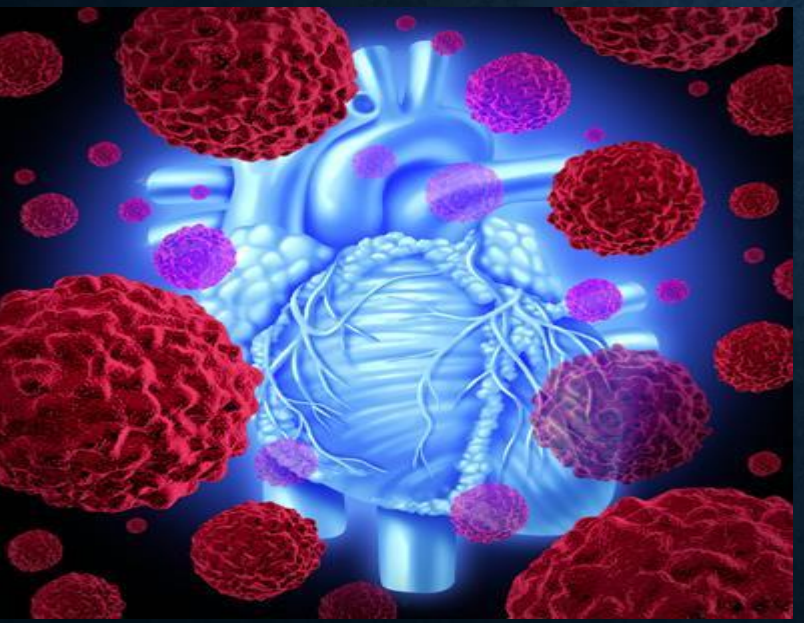
THE EVOLVING FIELD OF CARDIO-ONCOLOGY



Adapted from Moslehi, Cheng. *Science Translational Medicine*, 2013. Moslehi, *NEJM*. 2016

CARDIO-ONCOLOGY HAS EXPANDED OVER THE PAST 2 DECADES TO ADDRESS THE EVER-INCREASING ISSUE RELATED TO CARDIOVASCULAR DISEASE IN PATIENTS WITH CANCER AND SURVIVORS

Much of the focus of cardio-oncology has been on adult



Children represent 5% of new cancer diagnosis each year

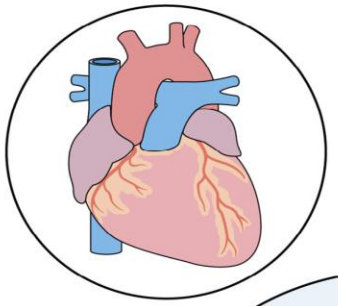
- high survival rate >85%
- decades of life expectancy after cancer treatment

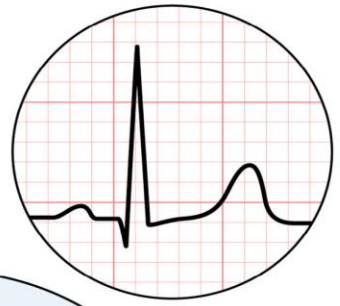
growing and aging populati on at risk of CTRCD

Left Ventricular Dysfunction
Anthracyclines, Alkylators, ABL targeting TKI, MEK inhibitors, Multi-tyrosine kinase inhibitors, CAR T-cell, Bi-specific antibodies, Radiotherapy

Pericardial Disease
Alkylators, Immune checkpoint inhibitors, Bi-specific antibodies, Radiotherapy

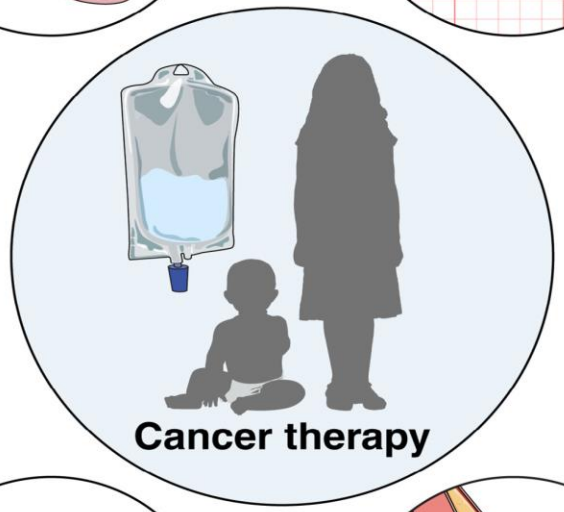
Valvular Disease
Radiotherapy

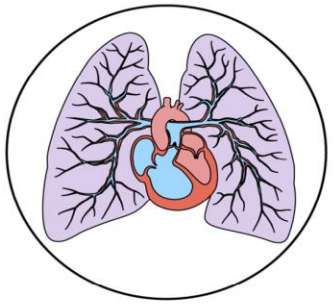




Arrhythmia, ECG changes
Anthracyclines, Alkylators, Arsenic, ABL targeting TKI, ALK inhibitors, BRAF kinase inhibitors, Multi-tyrosine kinase inhibitors, CAR T-cell, Bi-specific antibodies, Radiotherapy

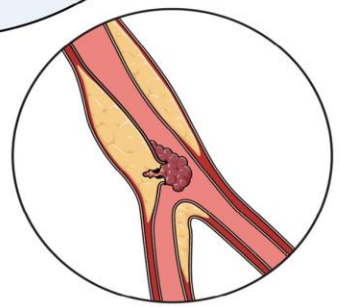
Metabolic Effects
ABL targeting TKI, ALK inhibitors, mTOR inhibitors





Pulmonary Hypertension and Fibrosis
Alkylators, Radiotherapy

Hypertension
ABL targeting TKI, ALK inhibitors, BRAF kinase inhibitors, MEK inhibitors, Multi-tyrosine kinase inhibitors, Anti-CD20, Immune checkpoint inhibitors



Vascular Thrombosis
ALK inhibitors, BRAF kinase inhibitors, MEK inhibitors, Multi-tyrosine kinase inhibitors

Coronary Artery Disease, Atherosclerosis
ABL targeting TKI, ALK inhibitors, Radiotherapy

Thomas D. Ryan. Circulation. Cardiovascular Toxicity in Patients Treated for Childhood Cancer: A Scientific Statement From the American Heart Association, Volume: 151, Issue: 15, Pages: e926-e943, DOI: (10.1161/CIR.0000000000001308)

Multimodal Imaging Atlas of Cardiac Masses

EDITED BY
AZIN ALIZADEHASL
MAJID MALEKI

ELSEVIER

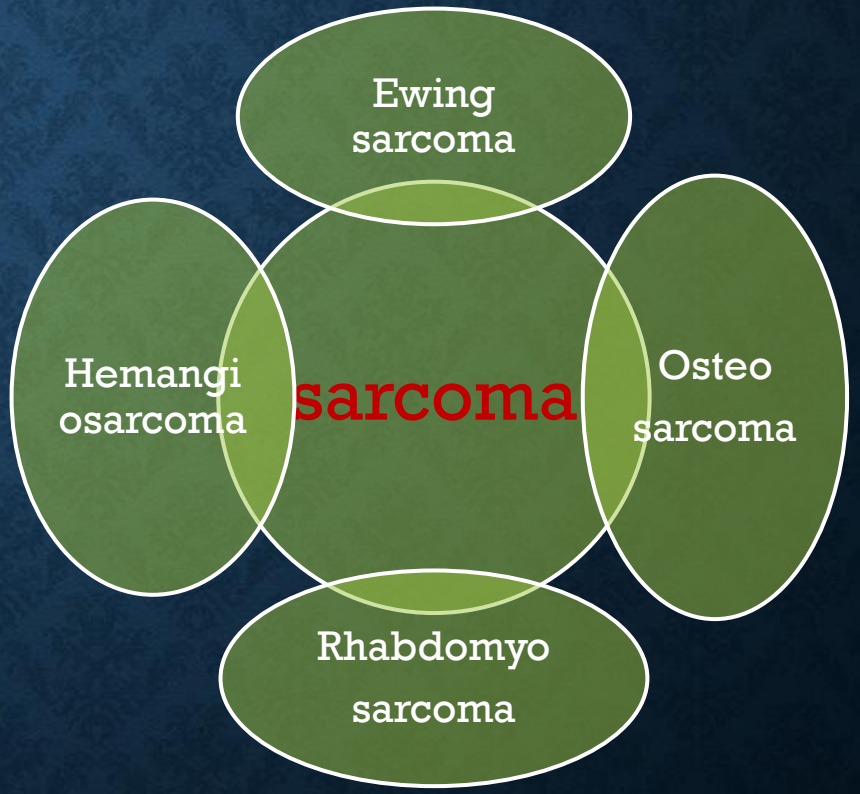
This mass is breaching anatomical boundries which makes malignancy as our first differential diagnosis, also there is other "**red flags" of malignicy,** just like the >5 cm diameter, the right heart localization , Pericardial effusion and involving the right AV groove.

Final diagnosis: "***Primary Cardiac AngioSarcoma***".

CARDIAC SARCOMA

- Cancer that develops in bone or soft tissue(muscle-tendons-connective tissue)

- Childhood sarcoma can be found anywhere in the body ,but most often in a child's arms , legs, chest and abdomen



PRIMARY CARDIAC SARCOMAS

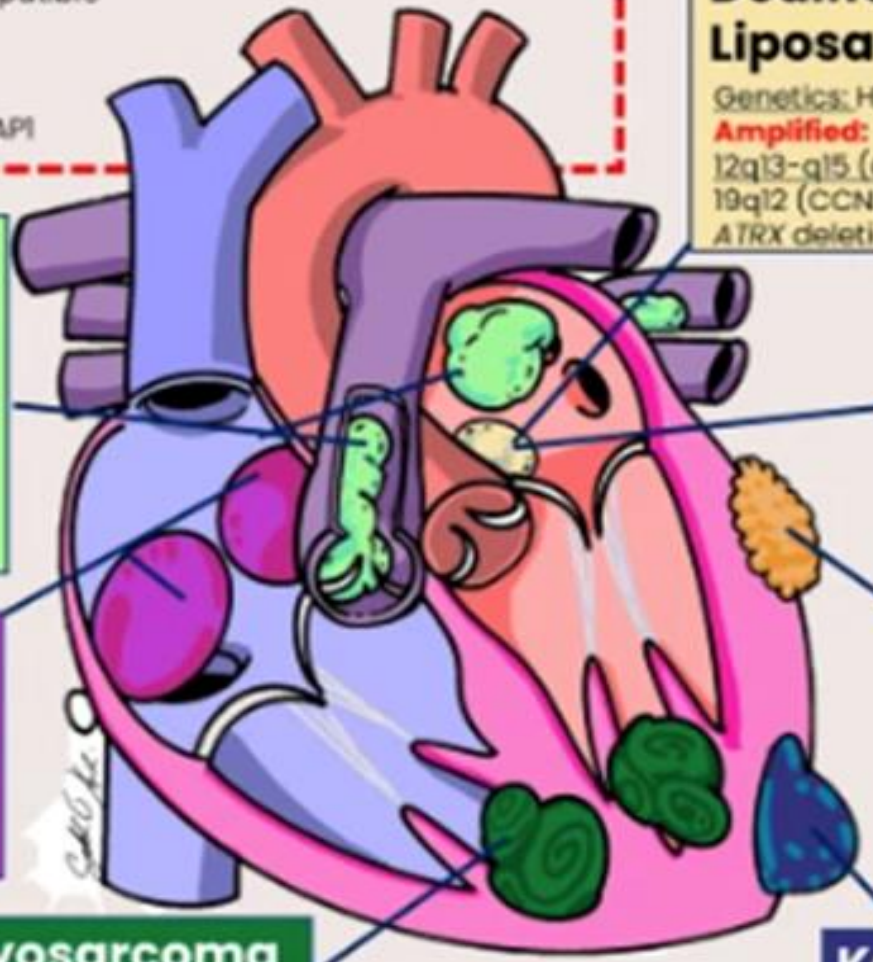
Undifferentiated Pleomorphic Sarcoma
(most common at Left Atrium)

Genetics of UPS (often compatible with Intimal Sarcoma)
Mutated or deleted:
TP53, RB1, ATRX or *TERT*
Amplified: 19q12 (CCNE1), YAP1

Dedifferentiated Liposarcoma
Genetics: High copy number, low TMB
Amplified:
12q13-q15 (*CDK4, HMGA2, MDM2*)
19q12 (CCNE1), 5p15 *TERT*
ATRX deletion (30%)

Intimal sarcoma
(Pulmonary trunk, artery, vein)

Genetics
Co-Amplified:
12q13-q15 (*CDK4, HMGA2, MDM2*)
& 4p12 (*PDGFRA, KIT, KDR*)
+/-
6p21 (*CCND3*) or 5p15 (*TERT*, 50%)



Leiomyosarcoma
Genetics
Mutated or deleted:
TP53, RB1, ATRX
(typically, no *TERT* or *MDM2*)

Synovial sarcoma
(Parietal pericardium)

SS18 (SYT) at 18q11
SS18::SSX1 - most common
SS18::SSX2, SS18::SSX4 or *SS18L1::SSX1*

Angiosarcoma
(Right heart, esp. AV sulcus)

Genetics: low TMB
Activating driver mutations:
POT1, PLCG1, KDR (VEGFR2), *FLT4* (VEGFR3)

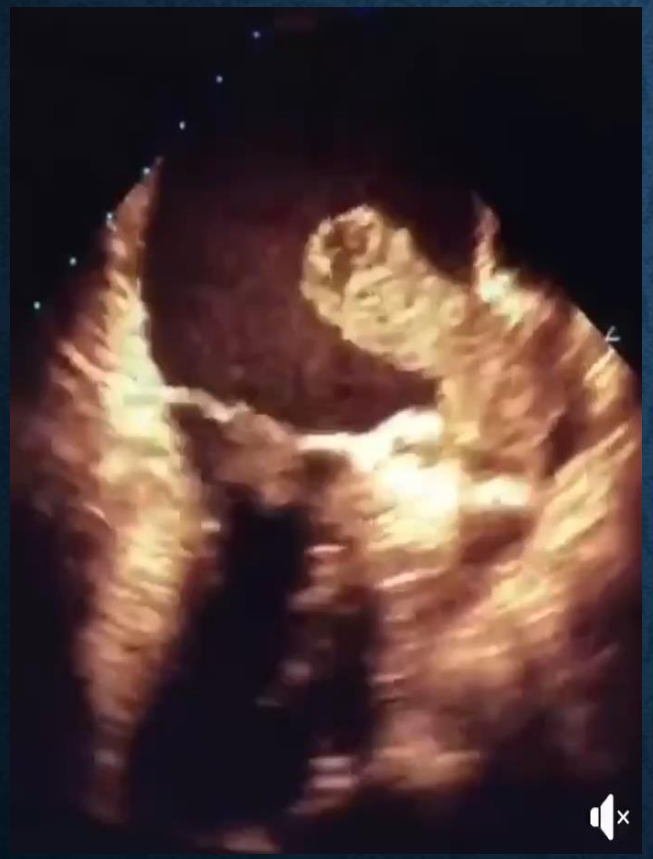
Rhabdomyosarcoma
(Non-specific sites)

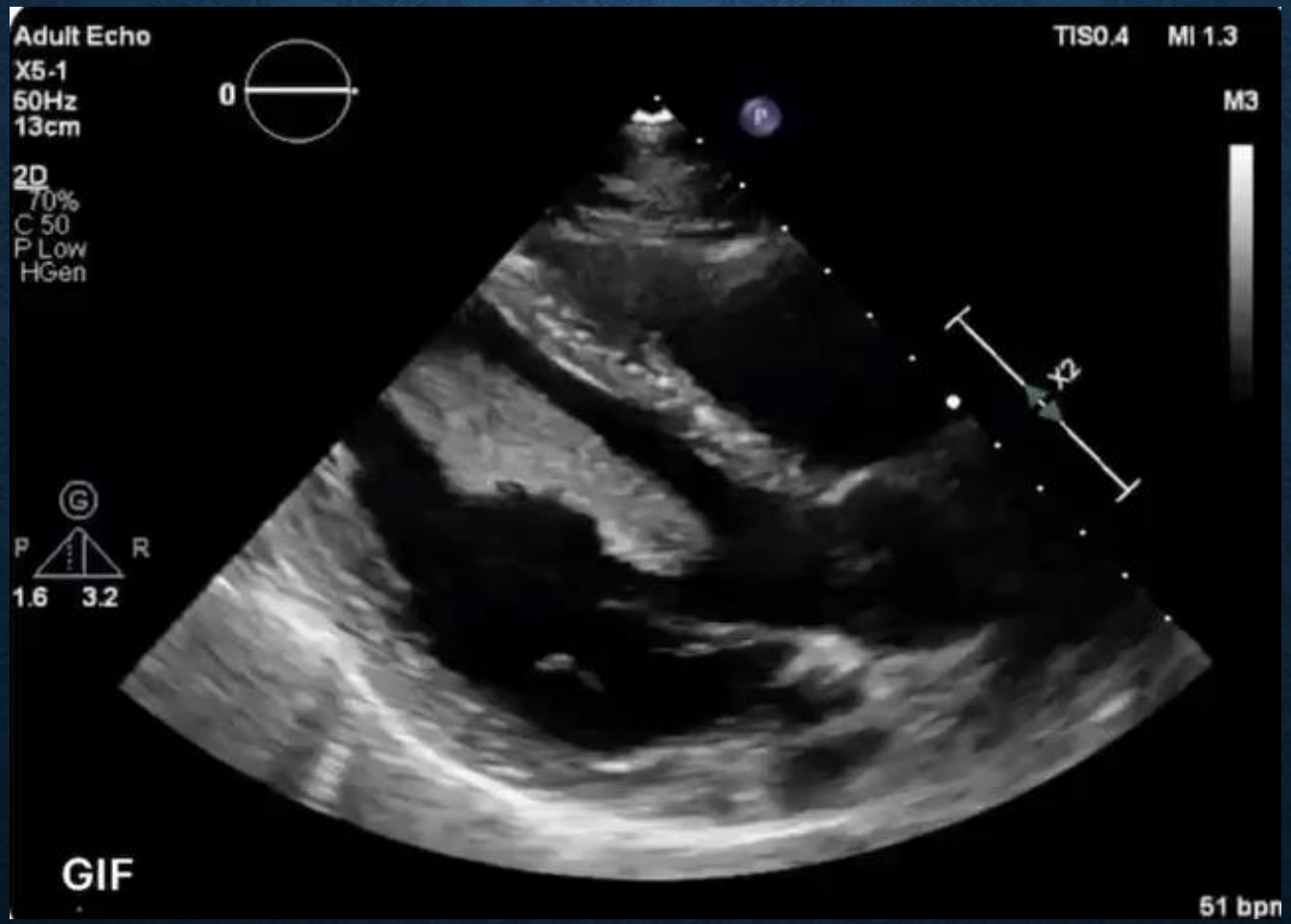
Genetics:
Embryonal - Ras (HRAS, NRAS, KRAS, NF1, FGFR4), PTEN, PIK3CA
Alveolar - *PAX3::FOXO1* or *PAX7::FOXO1*

Kaposi sarcoma
(Parietal pericardium)

Genetics: HHV8+
Viral tumors often lack driver mutations
Amplified
+/- 11q13 (*CCND1, FGF3, FGF19, FGF4*)

RHABDOMYOSARCOMA

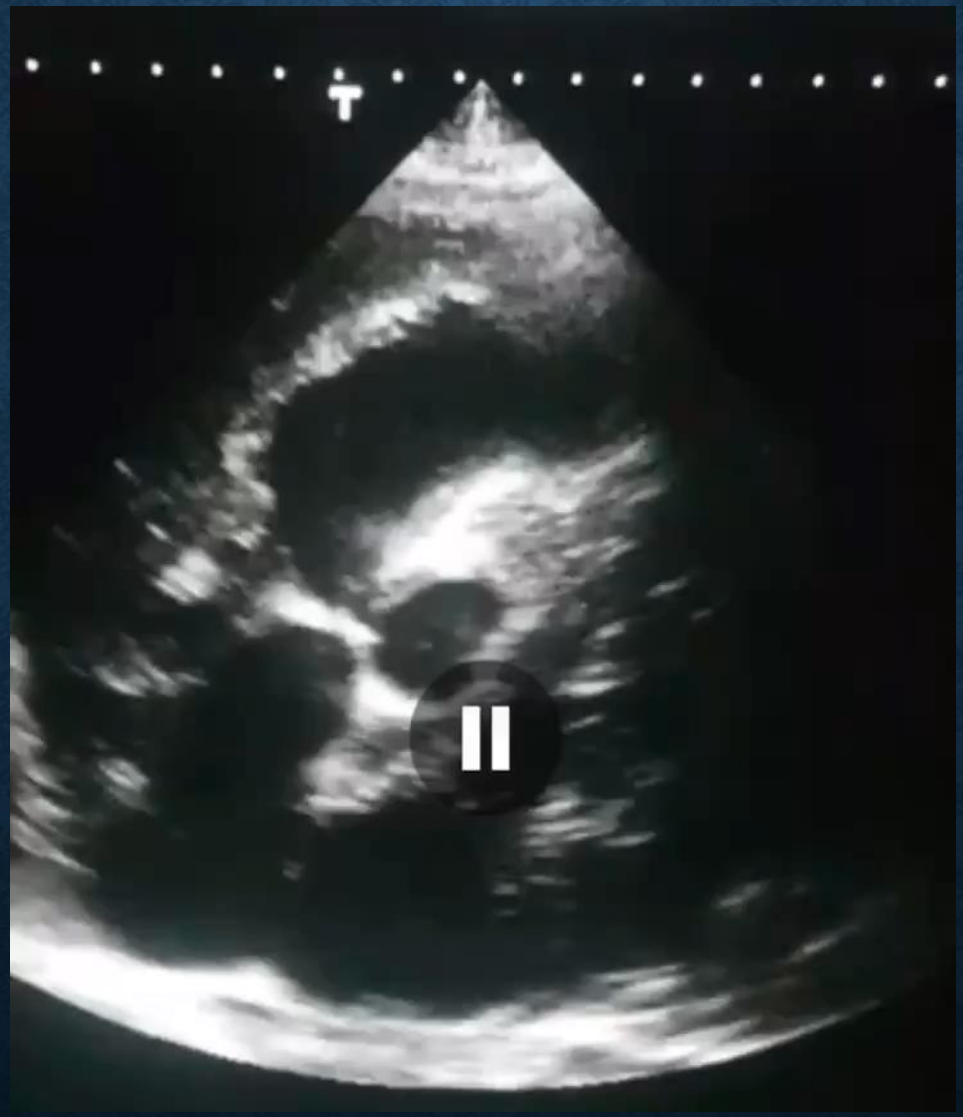



Adult Echo
X5-1
50Hz
13cm
2D
70%
C 50
P Low
HGen
TIS0.4
MI 1.3
M3
0
x2
G
P
R
1.6
3.2
GIF
51 bpm

Huge cardiac "Angiosarcoma"

LYPOSARCOMA !

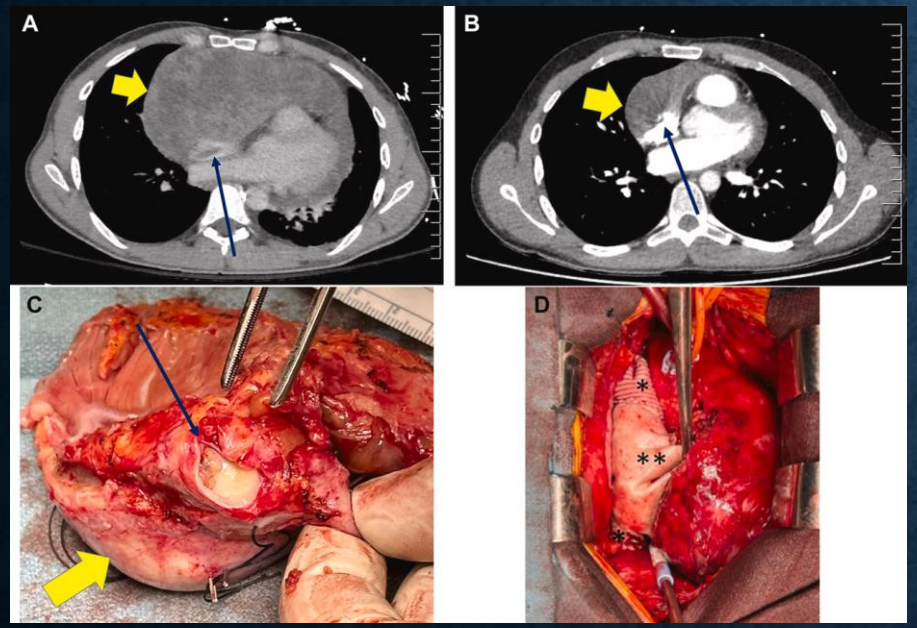
THE MASS IS ALONG RV FREE WALL



PRIMARY CARDIAC SARCOMA (PCS)

- Rare type of primary malignant tumor occurs in the heart (but 75% of malignant tumors)
- Most frequently diagnosed as Angiosarcoma,47 to 89% of pts present with metastasis at the time of diagnosis
- Rhabdomyosarcoma had best prognosis (20%) ,usually multifocal
- Aggressive (proliferate rapidly , locally invasive , high recurrence rate and high mortality rate)
- Median survival rate of 6 to 12 month

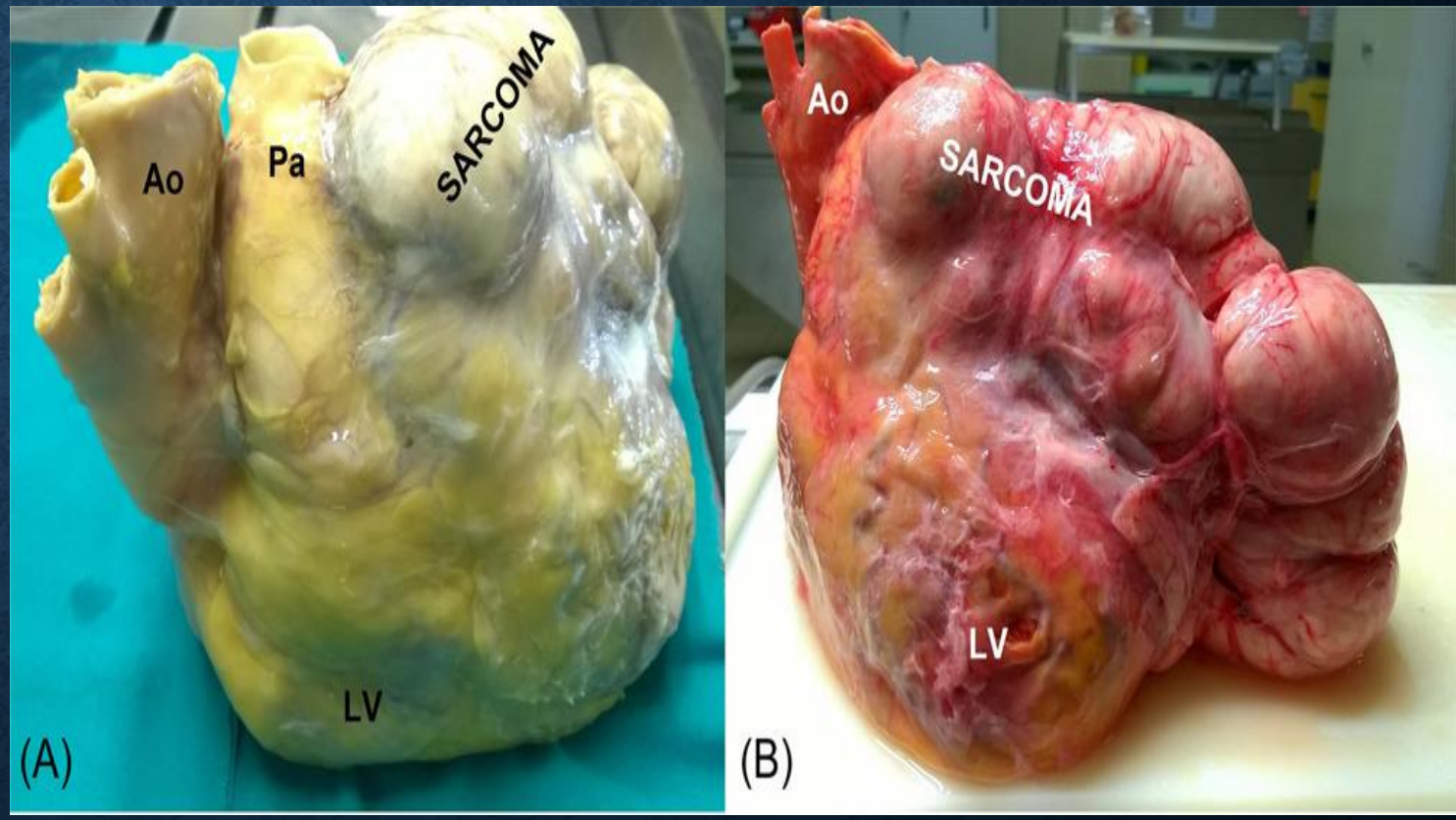
SIGNS AND SYMPTOMS



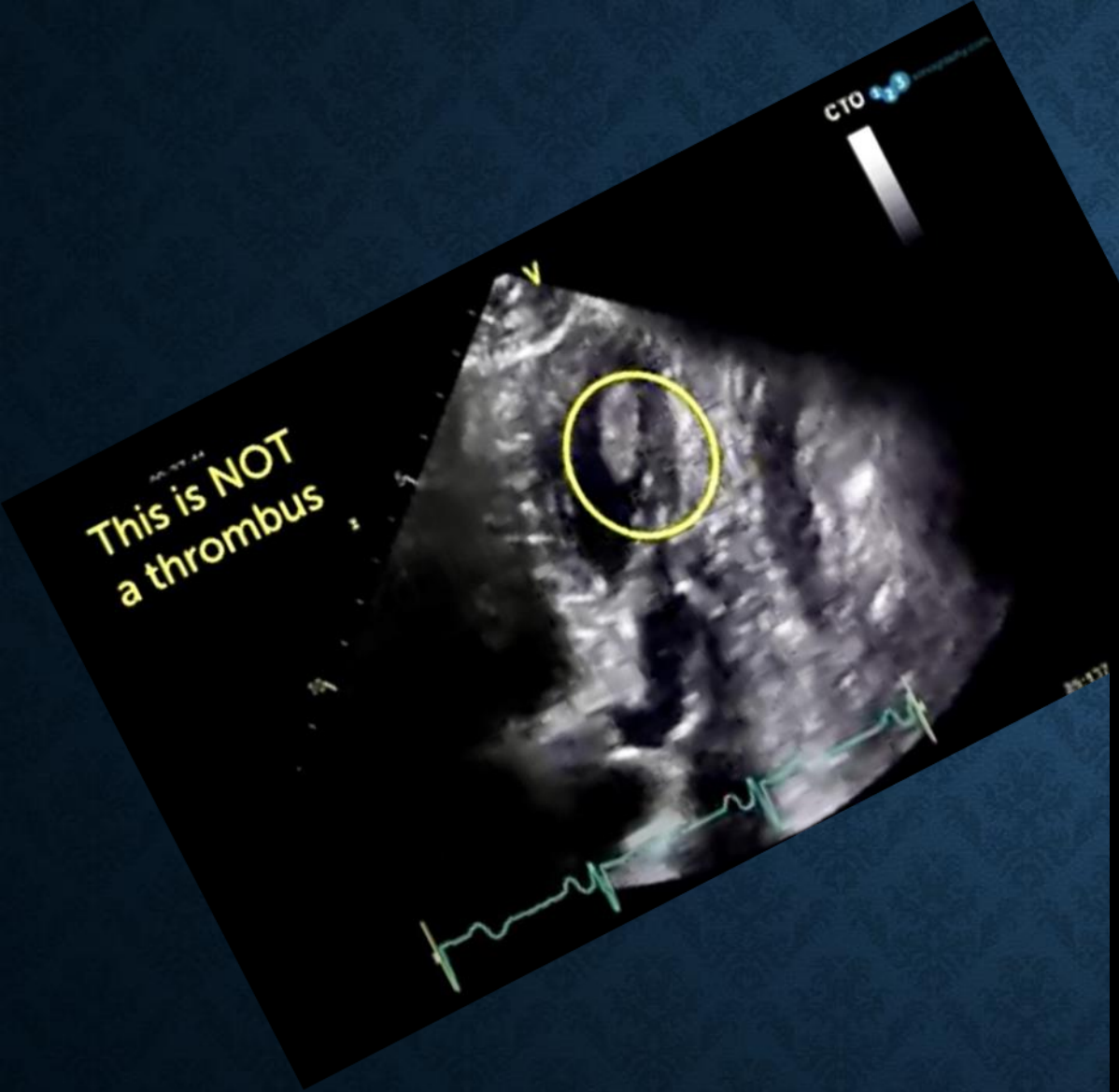
- Symptoms: depends on the location of the tumor
- Obstruction of inflow or outflow tract of RA (swelling of leg ,ankle ,abdomen , neck distension, pericardial effusion ,tamponade, emboly(stroke , limb ischemia), dyspnea, arrythmia, palpitation ,angina faint ,hemoptysis ,fever ,weakness , wight loss , Raynaud's phenomenon , enlargement of finger soft tissue

DIAGNOSIS

- Echocardiography
- ECG
- CT
- CMR
- CXR
- Cardiac catheterization
- Biopsy after excision
- Blood Tests



ANGIOSARCOMA



27 YEARS OLD MAN WITH LEIOMYOSARCOMA


10

23 YEARS OLD FEMALE COMPLAINTS OF CHEST PAIN & PROGRESSIVE SEVERE DYSPNEA 1 MONTH WITH SEVERE EOSINOPHILIA ON PERIPHERAL BLOOD SMEAR

ESOPHAGUS LEIOMYOSARCOMA

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ADULT: CARDIAC TUMORS: CASE REPORTS · Volume 16, P123-127, December 2022 · *Open Access*

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Resection of a synovial cell sarcoma by cardiac autotransplantation: A case report

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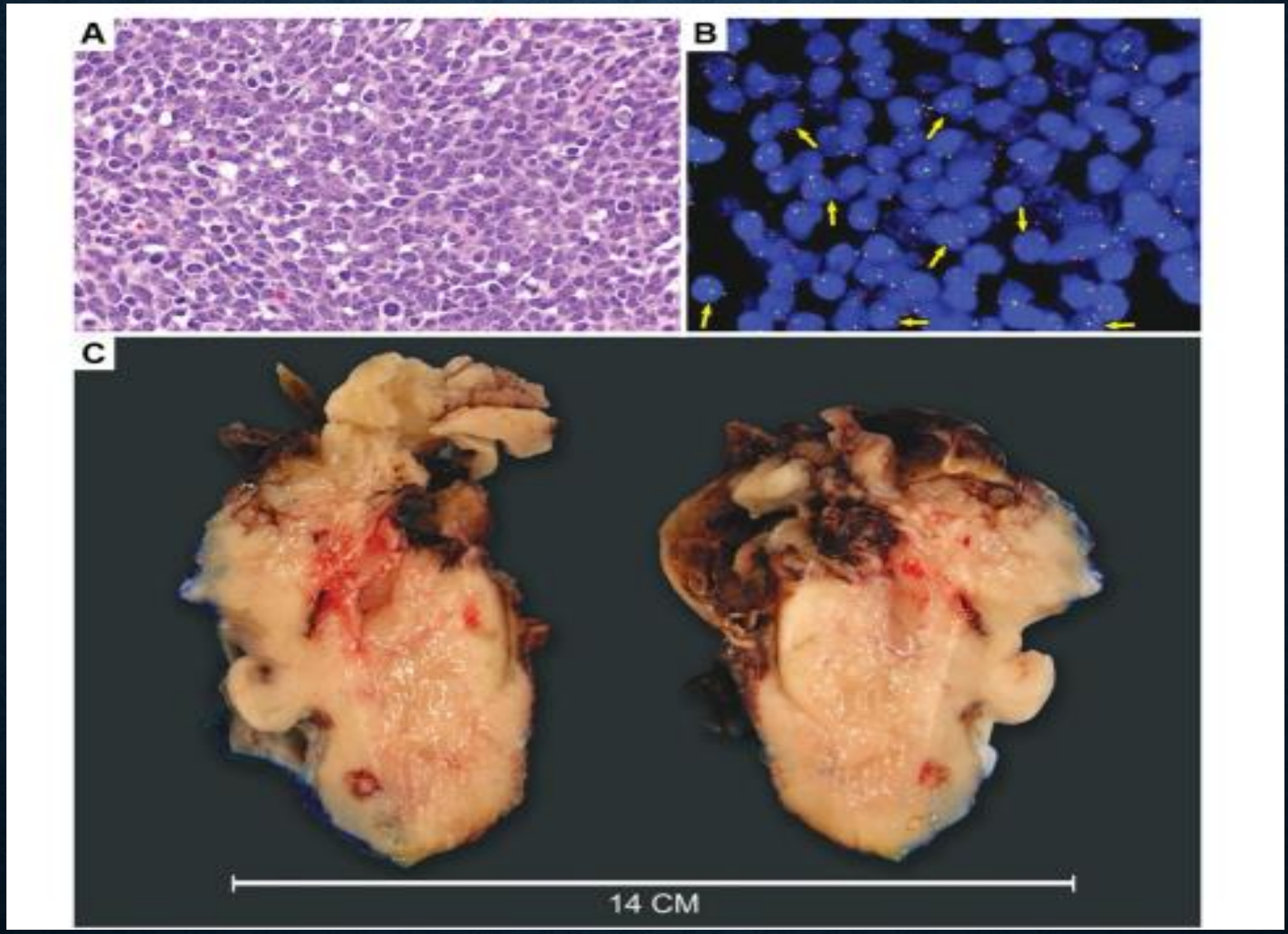
Patient with a large cardiac synovial sarcoma underwent a cardiac autotransplantation.

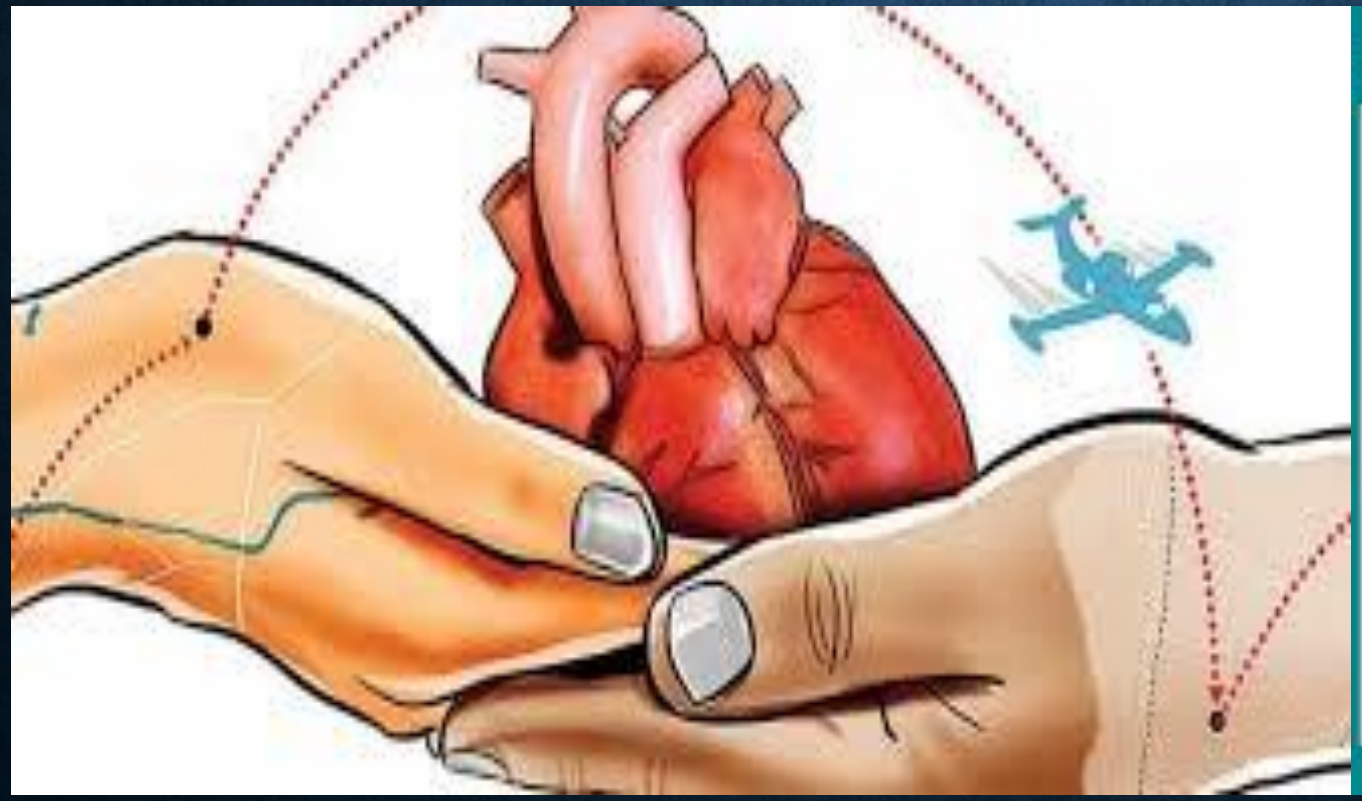
CENTRAL MESSAGE

Autotransplantation can be used as a technique to allow for safe tumor resection when a cardiac tumor involves both the left atrium and left ventricle.

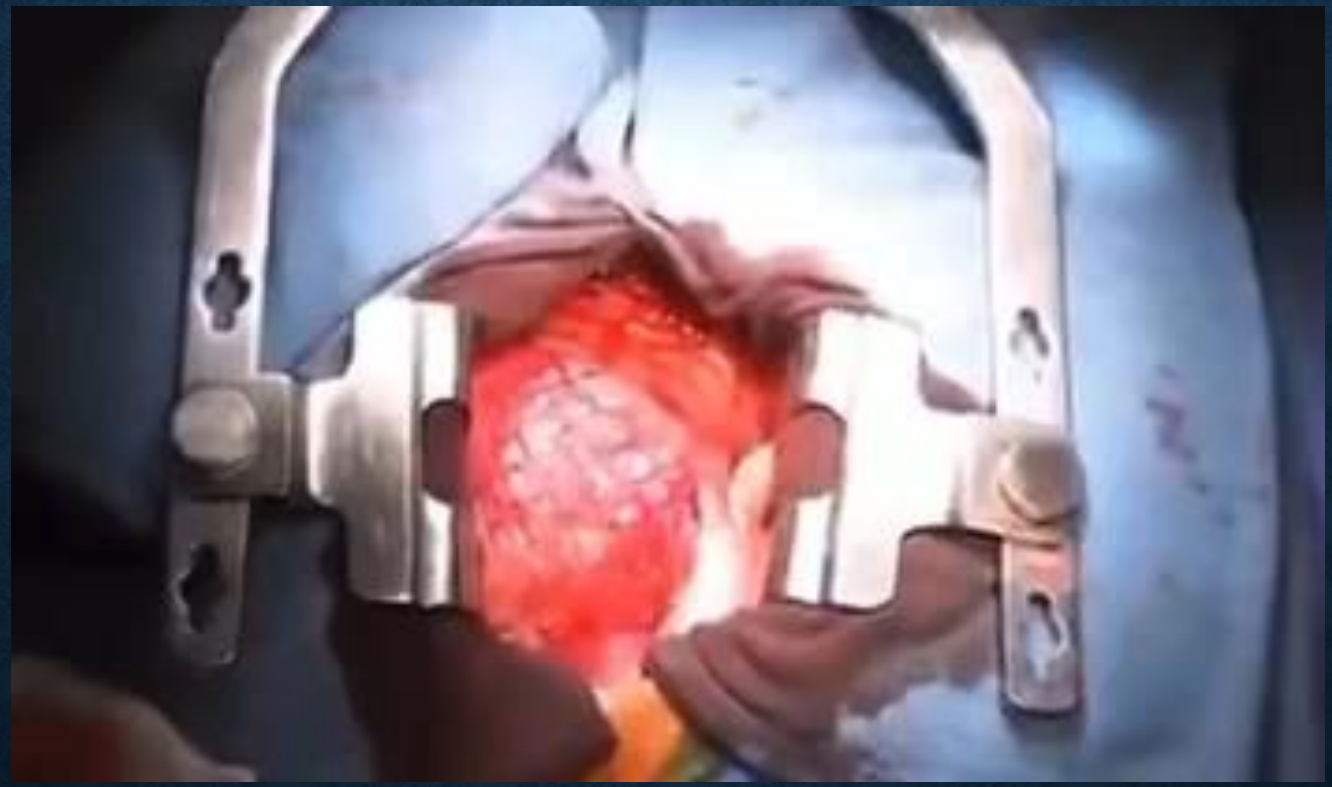
FIGURE 1. CT of the chest with intravenous contrast. A, CT angiography with intravenous contrast showing the 5.5- × 5.0-cm mass before surgery. B, CT of the chest with intravenous contrast taken after surgery. *CT*, Computed tomography.

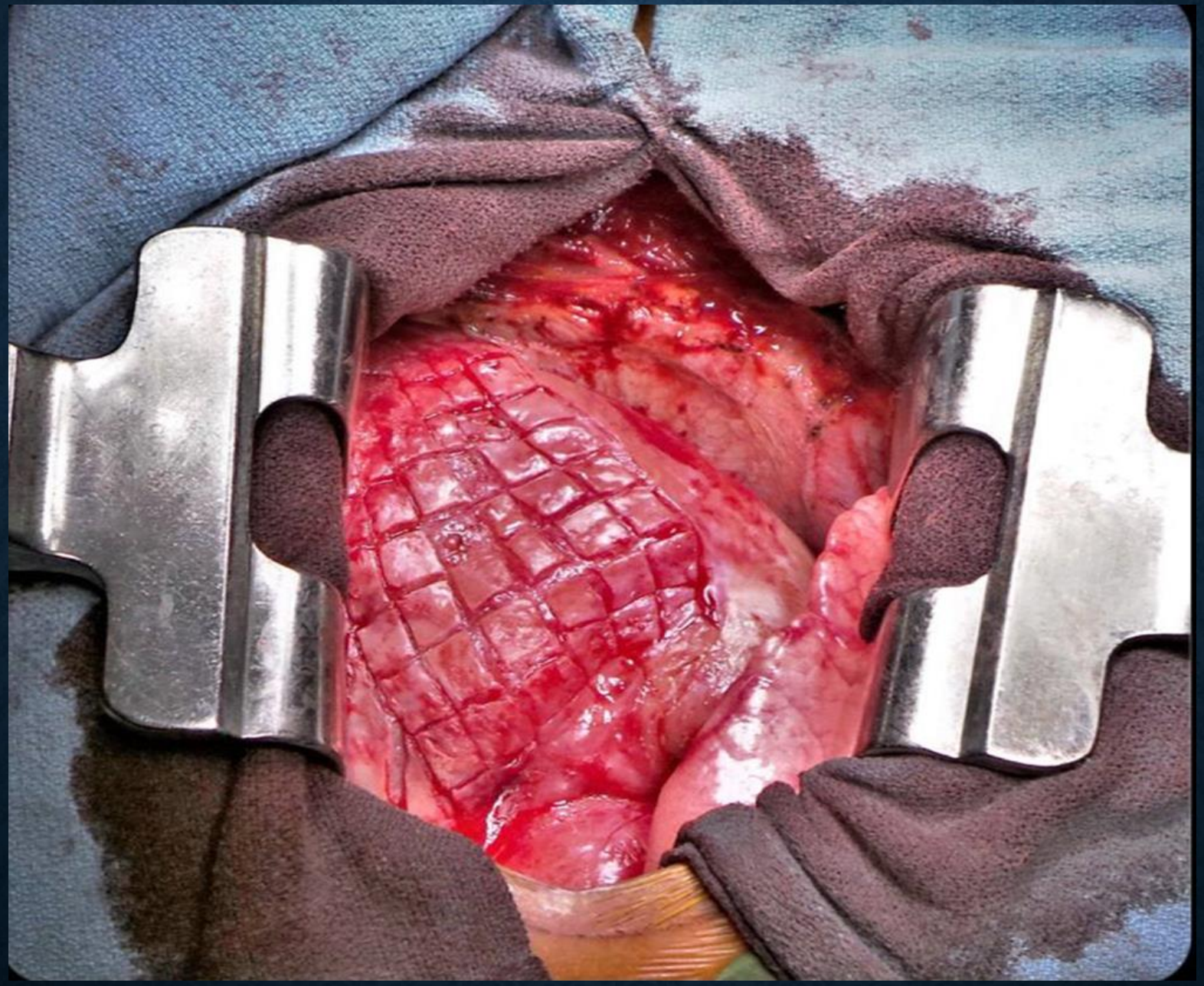

A
B
C
14 CM

23 / 280 HEART TRANSPLANTATION DUE TO CARDIOTOXICITY

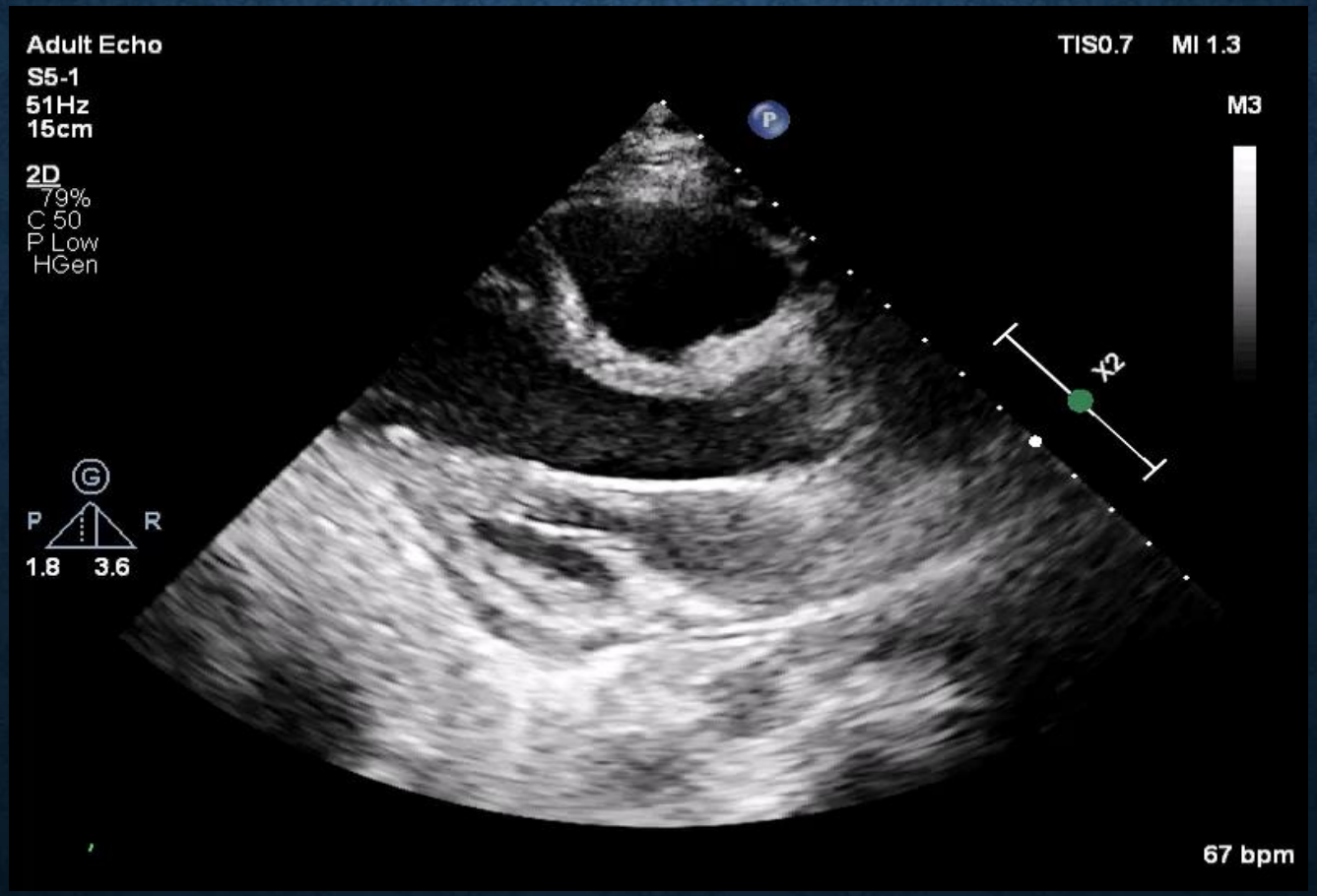


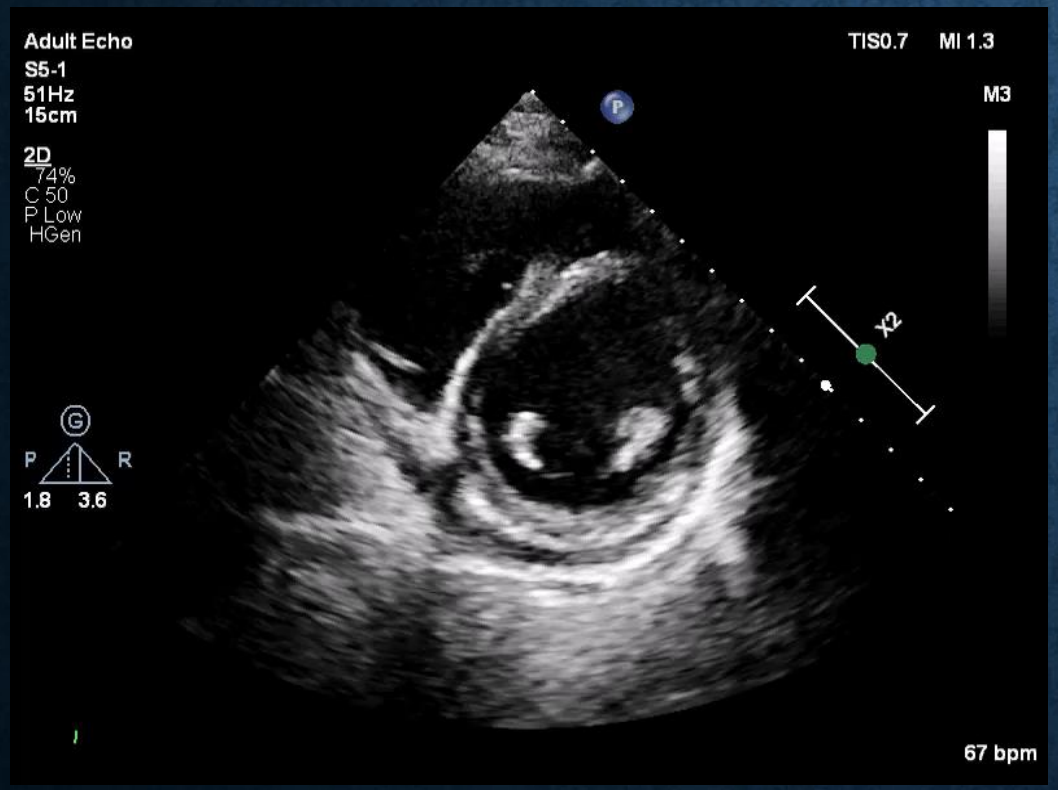
RADIATION INDUCED EFFUSIVE CP (OSTEOSARCOMA), 17 YEARS OLD MALE

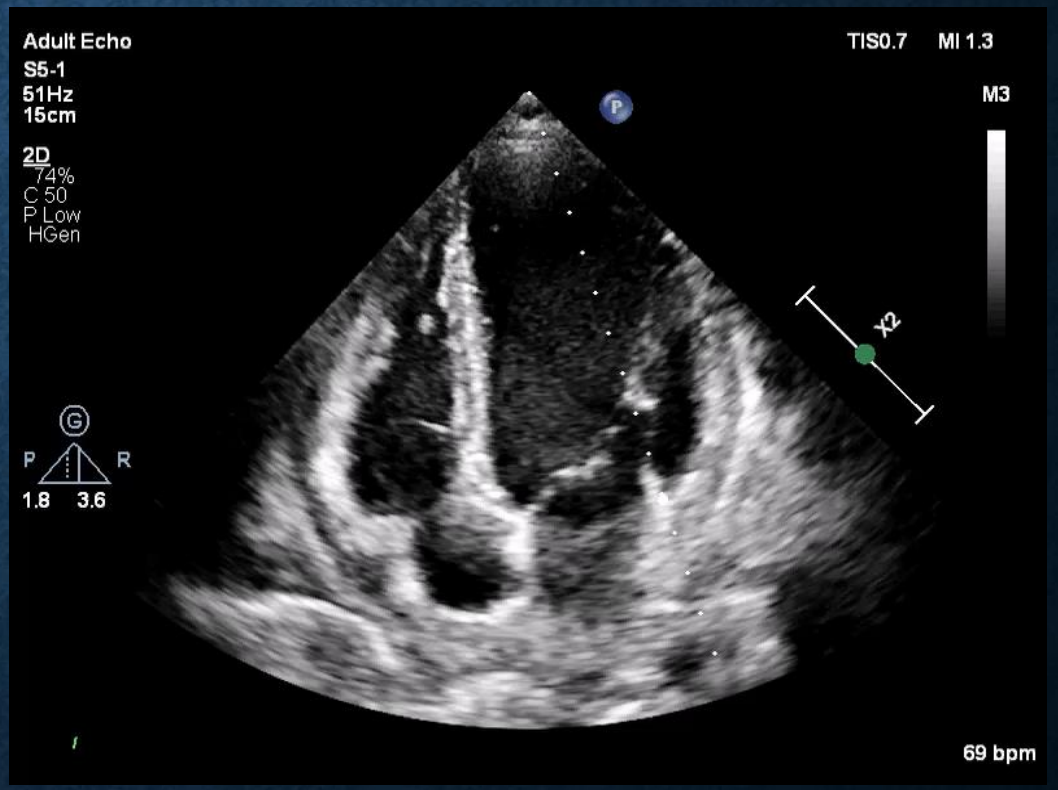


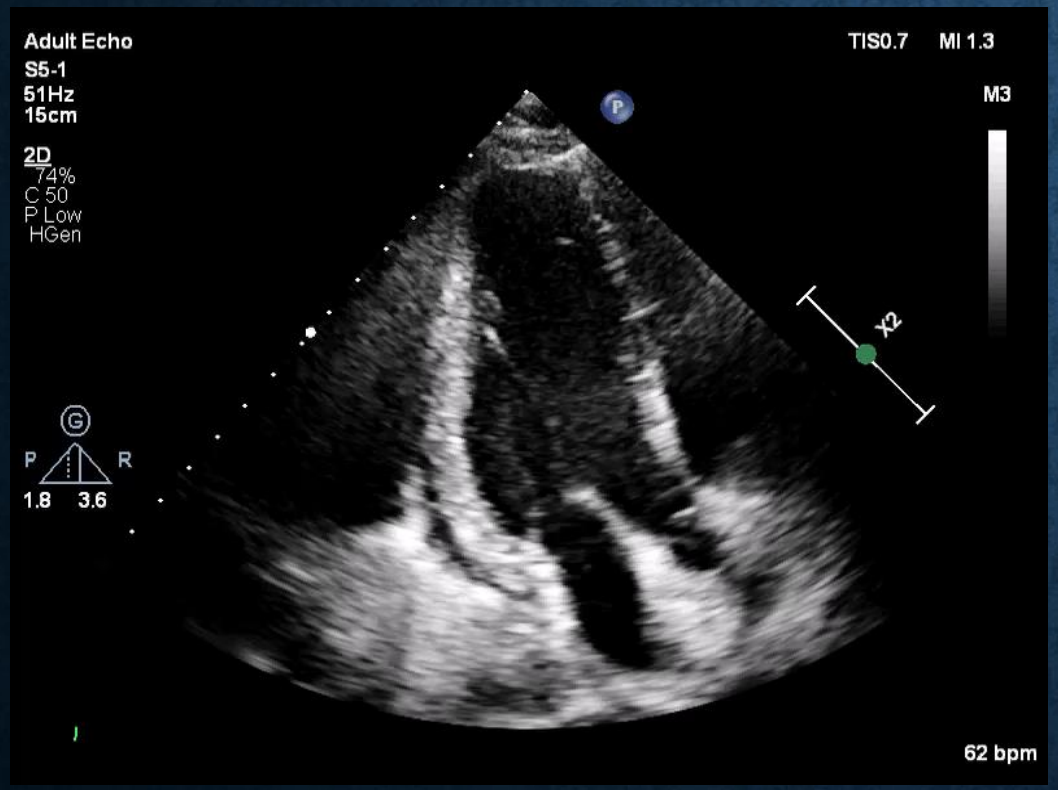


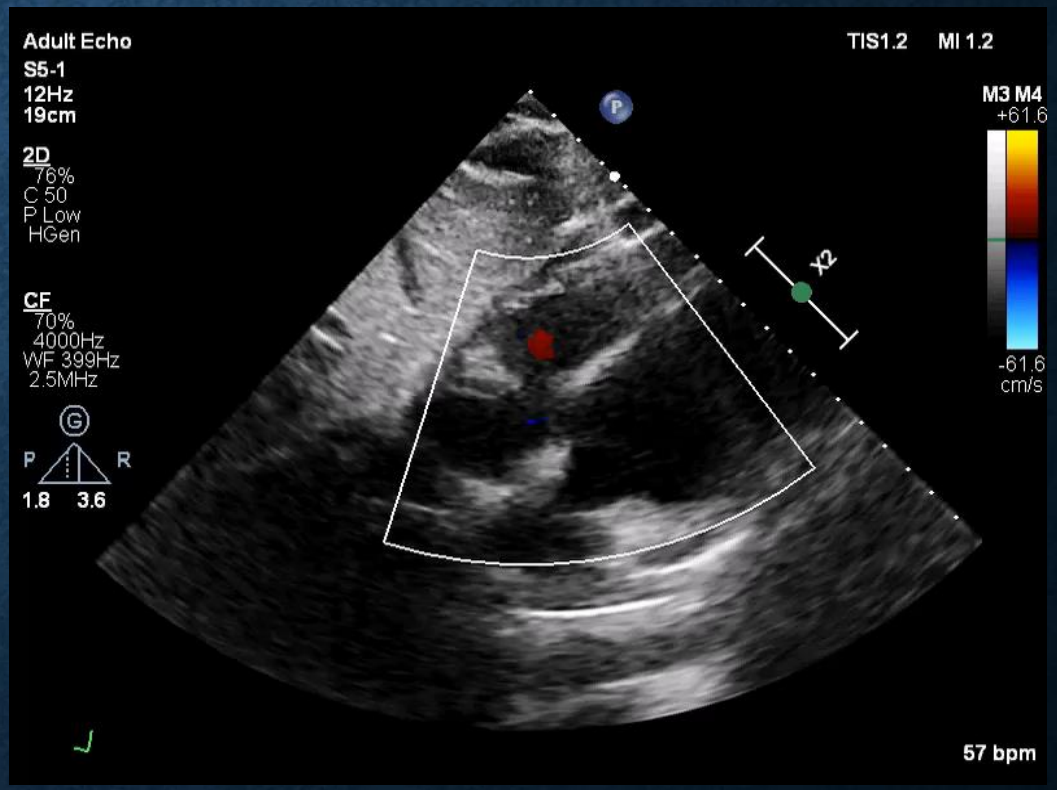
SOFT TISSUE SARCOMA




Adult Echo
S5-1
51Hz
15cm
2D
74%
C 50
P Low
HGen
TIS0.7
MI 1.3
M3
X2
P
R
1.8
3.6
67 bpm


Adult Echo
S5-1
51Hz
15cm
2D
74%
C 50
P Low
HGen
TIS0.7
MI 1.3
M3
X2
P
R
1.8
3.6
69 bpm

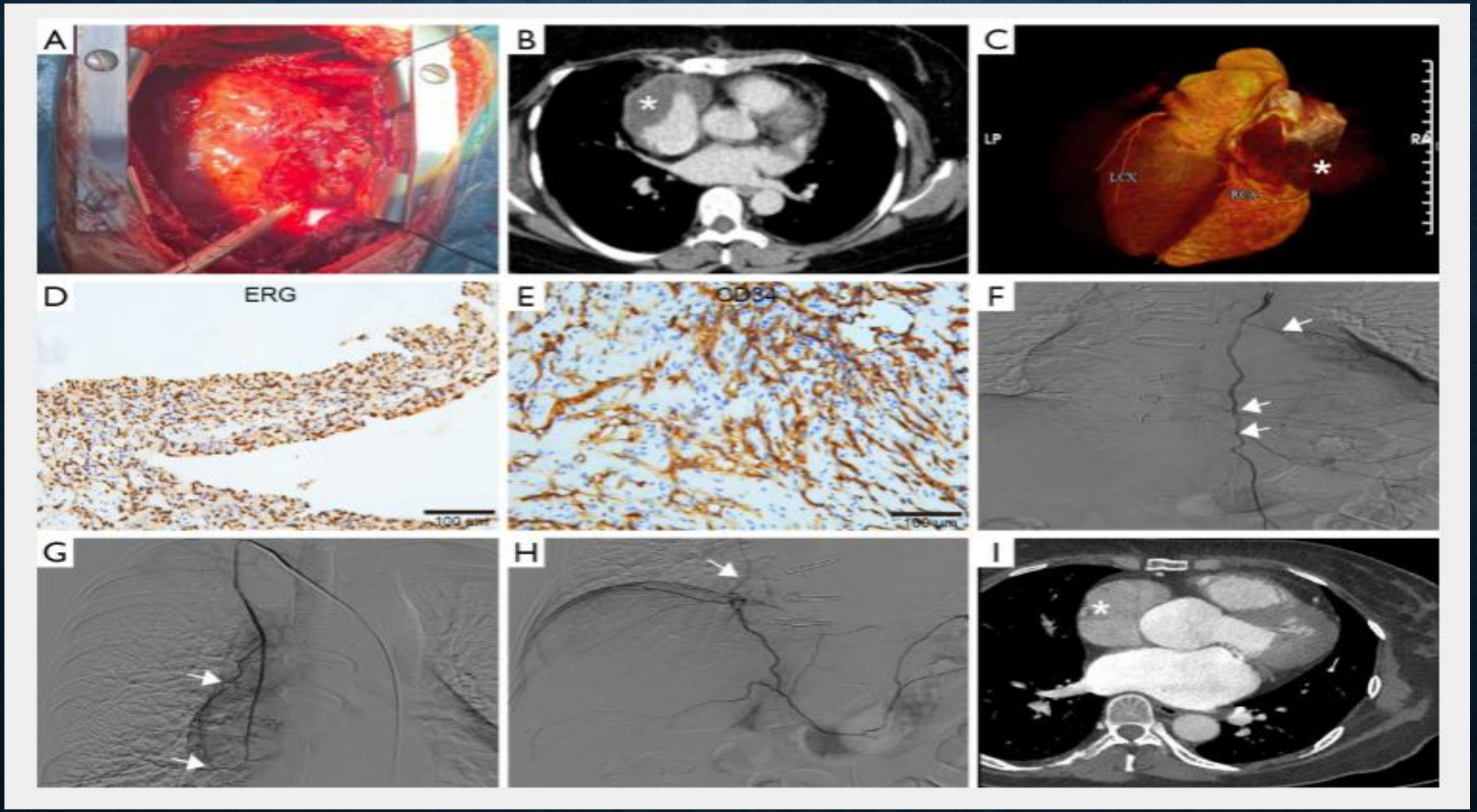

Adult Echo
S5-1
51Hz
15cm
2D
74%
C 50
P Low
HGen
TIS0.7
MI 1.3
M3
X2
P
R
1.8
3.6
62 bpm


Adult Echo
S5-1
12Hz
19cm
2D
76%
C 50
P Low
HGen
CF
70%
4000Hz
WF 399Hz
2.5MHz
TIS1.2
MI 1.2
M3 M4
+61.6
-61.6
cm/s
X2
P
R
1.8
3.6
57 bpm

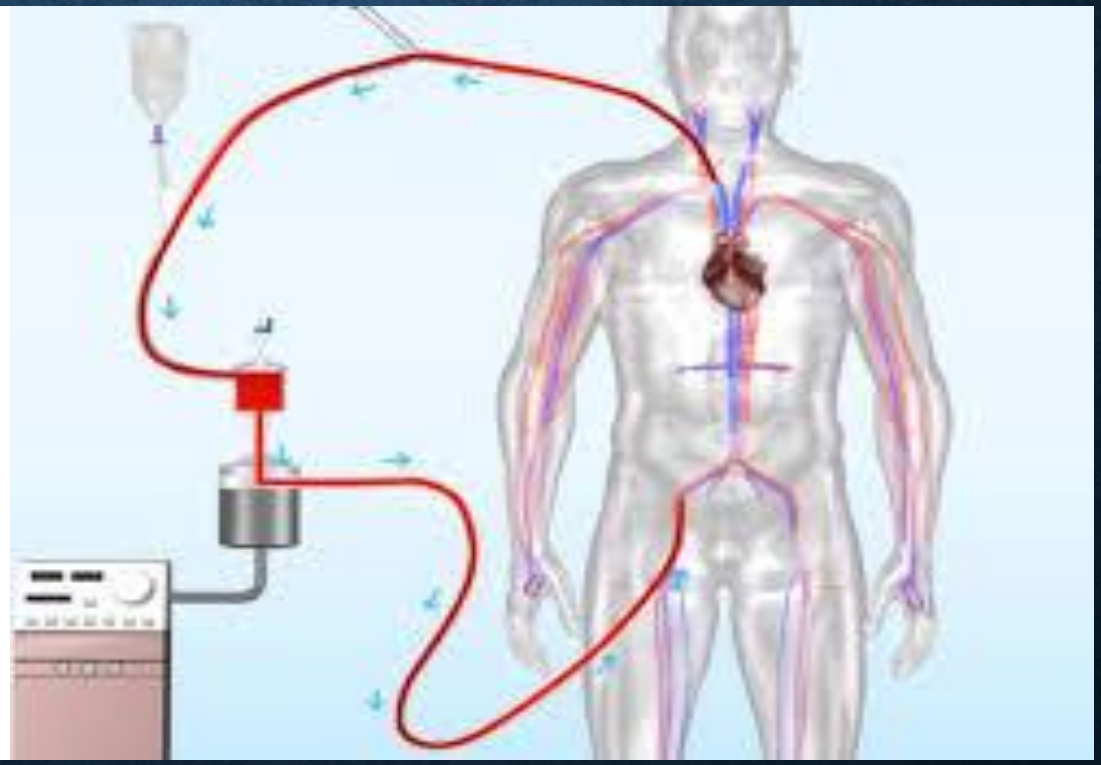
SARCOMA TREATMENT

- surgery(corner stone of therapy)
- Chemotherapy(Adriamycin and or Ifosfamide …)
- Radiotherapy
- Targeted therapy
- Immunotherapy

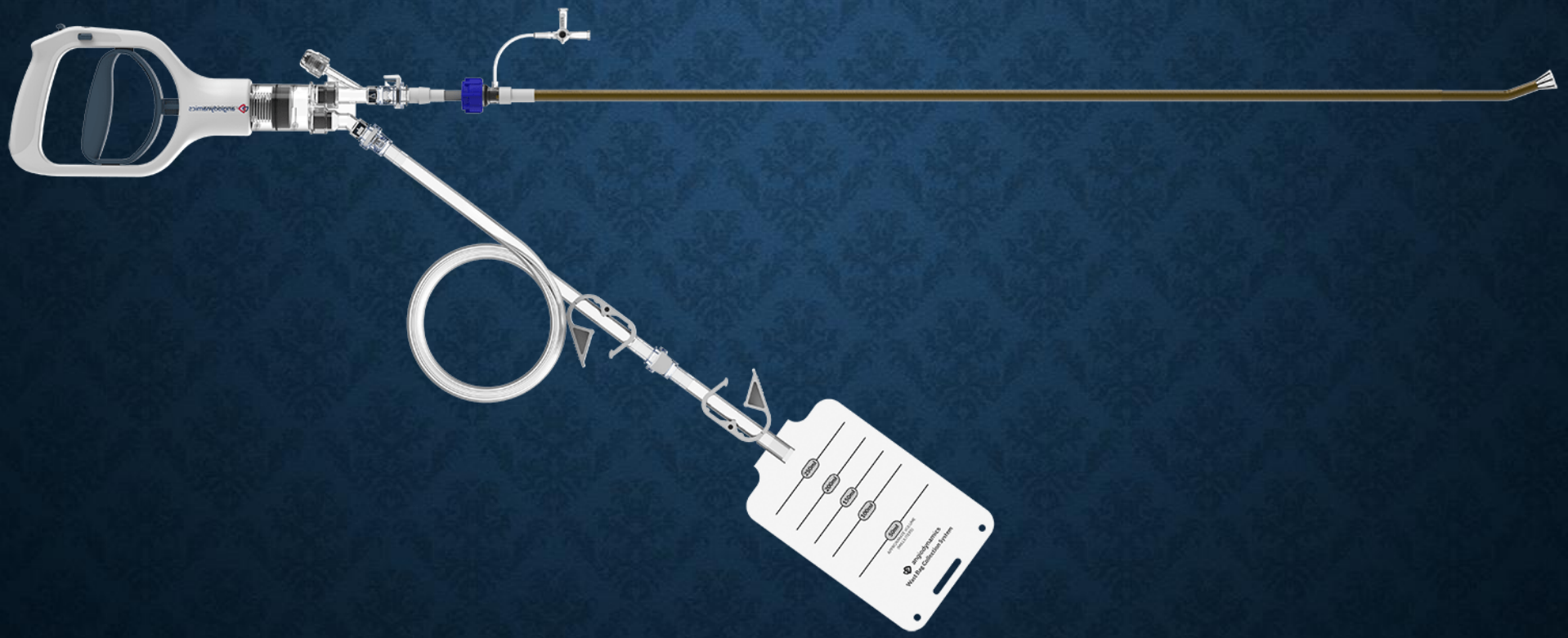
TRANSARTERIAL CHEMOEMBOLIZATION FOR INOPERABLE PRIMARY CARDIAC ANGIOSARCOMA



ANGIOVAC DEVICE



ALFA-VAC SUCTION DEVICE FOR CARDIAC MASSES

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