

TUMÖRER I HJÄRTAT/ Hjärttumörer

Entela Bollano

Överläkare

Kardiologkliniken /SU



2022 ESC Guidelines on cardio-oncology developed in collaboration with the European Hematology Association (EHA), the European Society for Therapeutic Radiology and Oncology (ESTRO) and the International Cardio-Oncology Society (IC-OS)

Developed by the task force on cardio-oncology of the European Society of Cardiology (ESC)

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ESC Clinical Practice Guidelines (CPG) Committee: listed in the Appendix.

ESC subspecialty communities having participated in the development of this document:

Associations: Association for Acute Cardiovascular Care (ACVCC), European Association of Cardiovascular Imaging (EACVI), European Association of Preventive Cardiology (EAPC), European Association of Percutaneous Cardiovascular Interventions (EAPCI), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

Councils: Council of Cardio-Oncology, Council on Hypertension, Council on Valvular Heart Disease.

Working Groups: Aorta and Peripheral Vascular Diseases, Cardiovascular Pharmacotherapy, e-Cardiology, Myocardial Function, Pulmonary Circulation and Right Ventricular Function, Thrombosis.

Patient Forum

JACC: CARDIOONCOLOGY

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VOL. 6, NO. 6, 2024

STATE-OF-THE-ART REVIEW

Multimodality Imaging in the Diagnostic Work-Up of Patients With Cardiac Masses

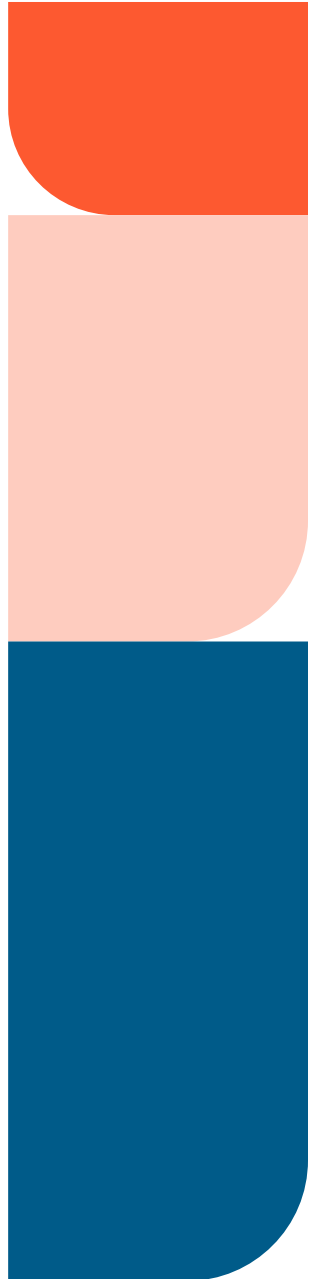
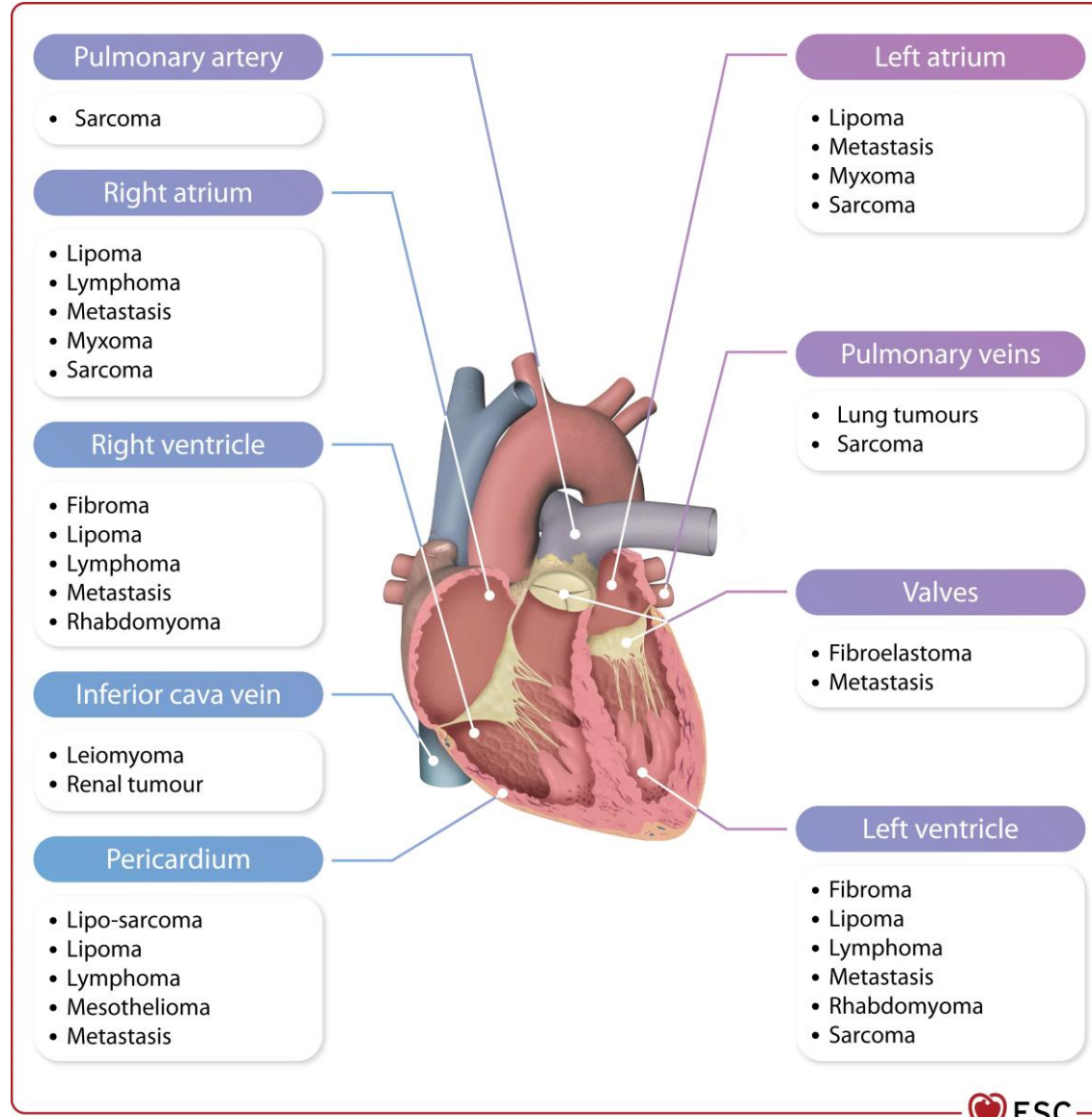


JACC: CardioOncology State-of-the-Art Review

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ABSTRACT

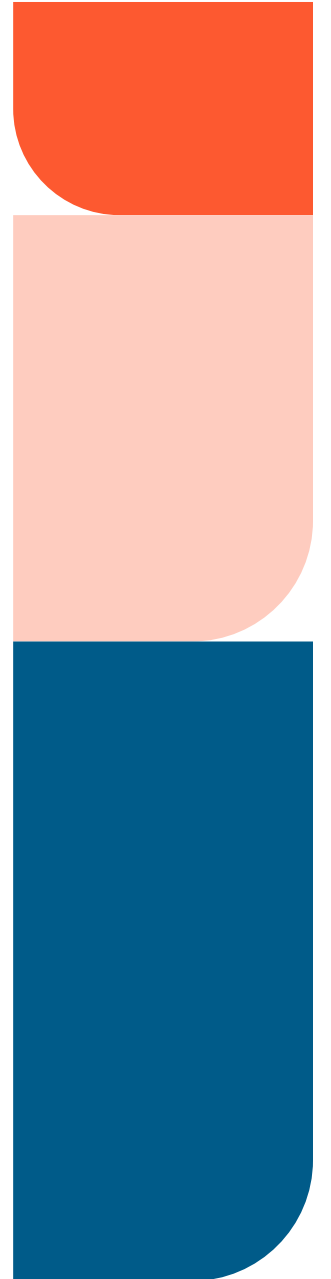
Cardiac masses encompass a diverse range of benign and malignant tumors as well as pseudotumors. Accurate histologic identification is essential for guiding appropriate treatment, yet the diagnostic process remains challenging. Although biopsy is traditionally the diagnostic gold standard, its invasive nature and associated risks limit its application. A noninvasive multimodality imaging approach has recently emerged as an alternative, but standardized protocols and supporting evidence are still lacking. Echocardiography is typically the initial imaging modality, with cardiac magnetic resonance recognized as the noninvasive diagnostic gold standard. Cardiac computed tomography provides complementary data to aid in diagnosis and management, while positron emission tomography serves as a third-level imaging option. This state-of-the-art review highlights the role of current multimodality imaging techniques in diagnosing and managing cardiac masses and explores future directions for their applications. (JACC CardioOncol. 2024;6:847-862) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).



Immune Therapy: checkpoint inhibitors

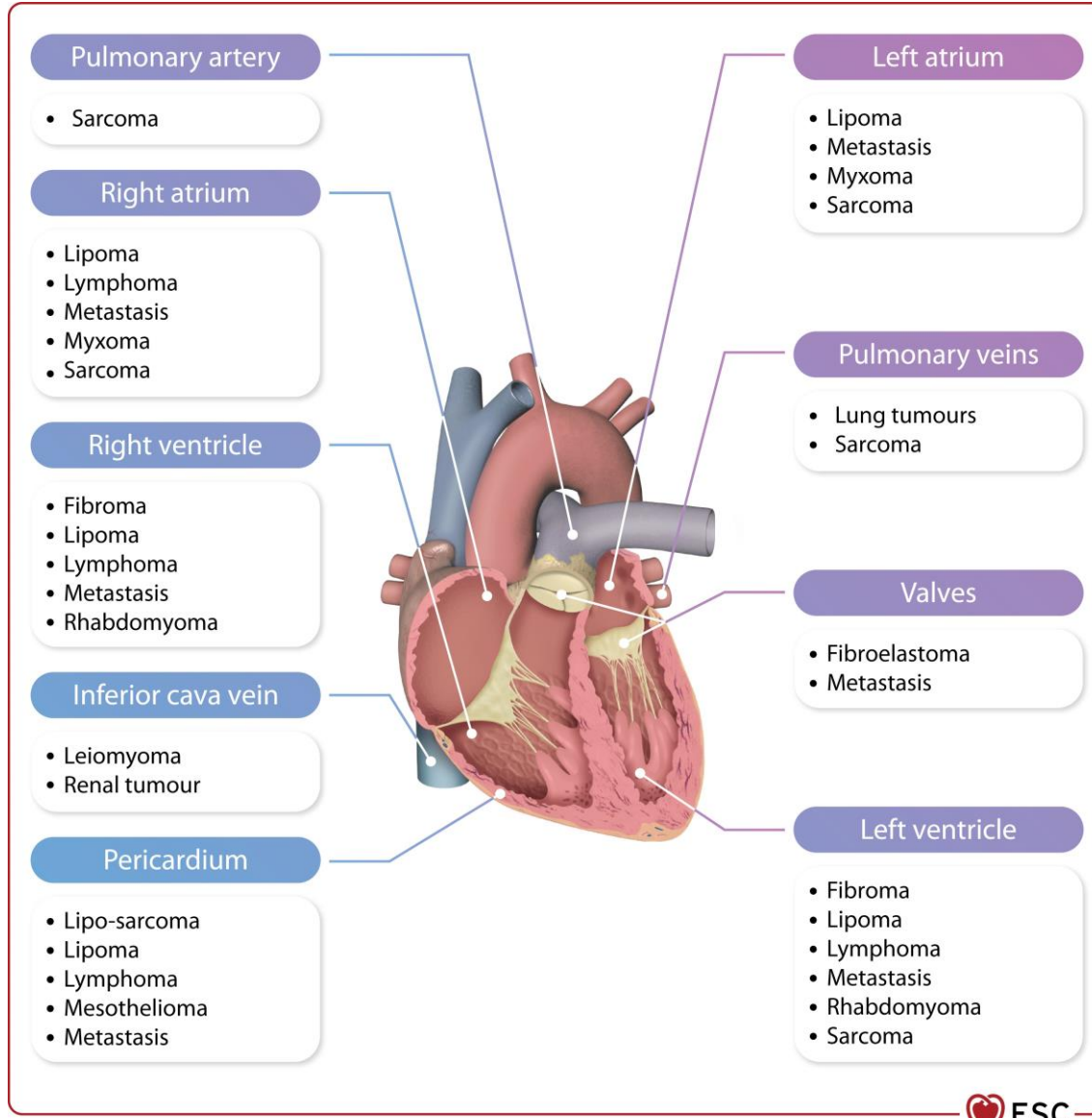
ICIs

- Revolution in multiple cancer and metastatic setting
 - Monoclonal antibodies block checkpoints in Ca cells, activate T cells to destroy Ca
 - CTLA-4 inhibitors: ipilimumab, tremelimumab
 - PD-1: nivolumab, pembroluzumab, cemiplimab
 - PD-L1: atezolizubam, avelumab, durvaluma
- Immune-related adverse events
 - Myocarditis: often early first 12 weeks, potentially life threatening
 - Arrhythmias, VT
 - Plaque ruptures, ischemia, MI, Takotsubo, Heart failure, pericarditis



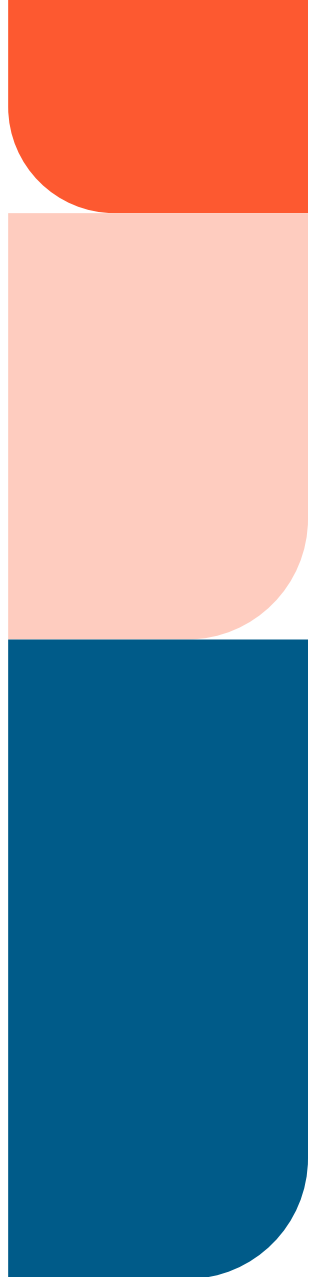
Immune Therapy: checkpoint inhibitors ICIs

Asymptomatic (increased troponins)	Suspect Myocarditis (stable patient)	Unstable patient (hypotension, arrhythmias, lower EF)
<ul style="list-style-type: none"> ✓ Hold ICI • Evaluate for signs and symptoms of CHF, ischemia, arrhythmias or myositis • Check EKG, CK, TNI serially • Echocardiography? • If normalized within 2 weeks and no cardiac abnormality, may reuse ICI • If not+ symptoms – Contact cardiology 	<ul style="list-style-type: none"> ➤ Consult cardiology • Rule out MI • Telemetry • ECHO /MRI • Biopsy?! • Diagnose: Myocarditis • Start Treatment: <ul style="list-style-type: none"> • Methylprednisolone 2mg/kg/da or 1 g IV bolus 3 days • If not improved in 24 hours or patient unstable move to CCU • If stable, taper over at least 4 weeks • Echo control; TNI, other treatment 	<ul style="list-style-type: none"> ➤ CCU • Specific treatment: <ul style="list-style-type: none"> • Methylprednisolone 1g IV and taper over 4-6 weeks • Adding ATG? Tacrolimus? MMF? Infliximab? • Extern PM (if needed) • Other treatments (inotropi; antiarrhythmics,antikoagulantia) • Contact to nearest Cardiac transplant Unit

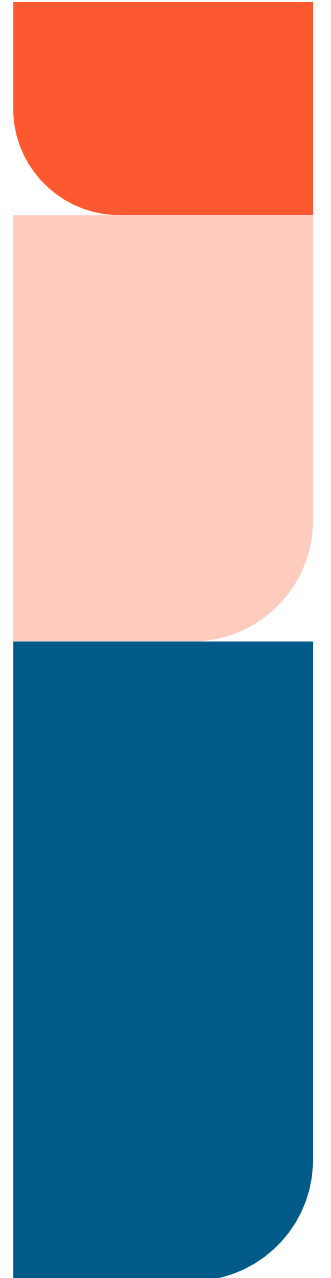


Hjärttumörer

- Primära hjärttumörer
 - Extremt sällsynta
 - Incidence < 0.1 procent
 - 90% är godartade
- Metastaser till hjärtat
 - 20 gånger vanligare
 - Hos 1 av 5 som dör pga cancer

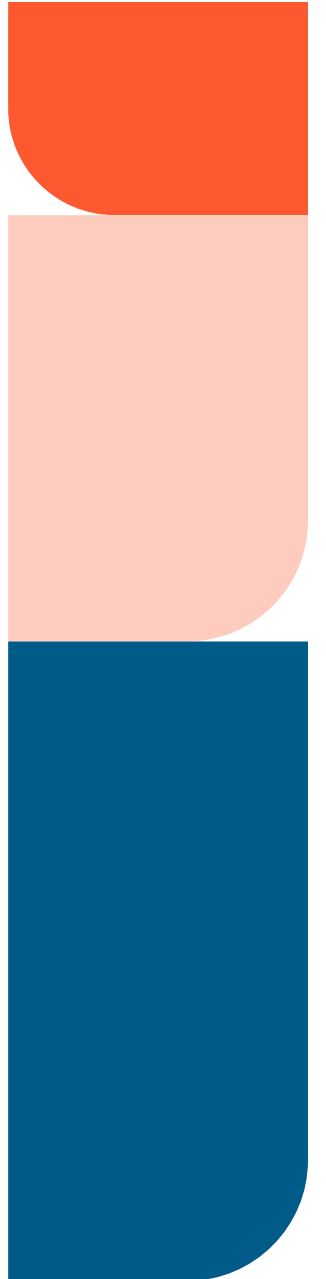


- Prevalens 0,01‰-0.1%
- 90% godartade
 - Av dessa utgör 80% av myxom i vä förmak
 - 15% i hö förmak
 - < 5% engagerar båda förmaken
 - Myxom > papillärt fibroelastom > lipom > rabdomyom > fibrom > hemangiom
- 10% maligna
 - Av dessa 75% sarkom
 - Angiosarkom > rabdomyosarkom > mesoteliom > fibrosarkom



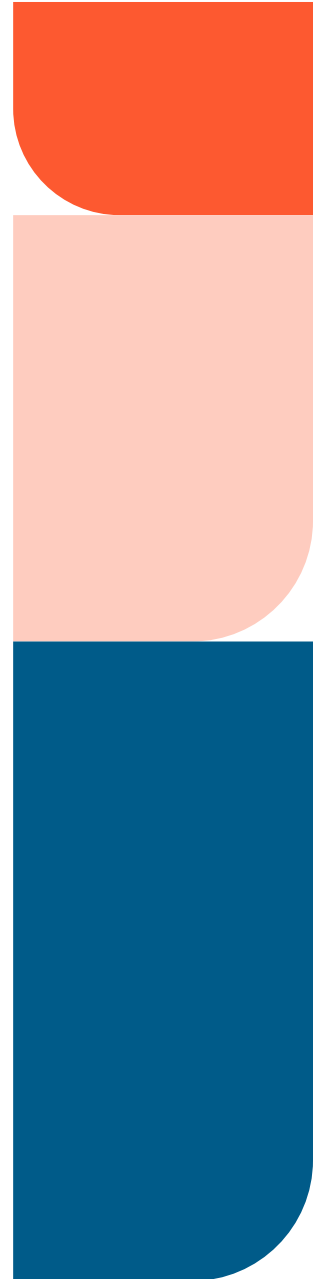
Symptom

- Embolier (vanligast hos tumörer i vän förmak, aortaklaff)
 - Neurologiska bortfall
 - Ischemi i extremiteter
- Flödesobstruktion
 - synkope
- Klaffpåverkan
 - Svikt, 2nd PAH etc
- Infiltrativ växt i myokard (svikt, arytmier, block, perikardvätska, infarkt)
- Infiltrativ växt i mediastinum, lunga etc (hosta, svalgpåverkan)
- Allmänsymtom (anemi, led/muskelvärk, viktnedgång, trötthet etc)



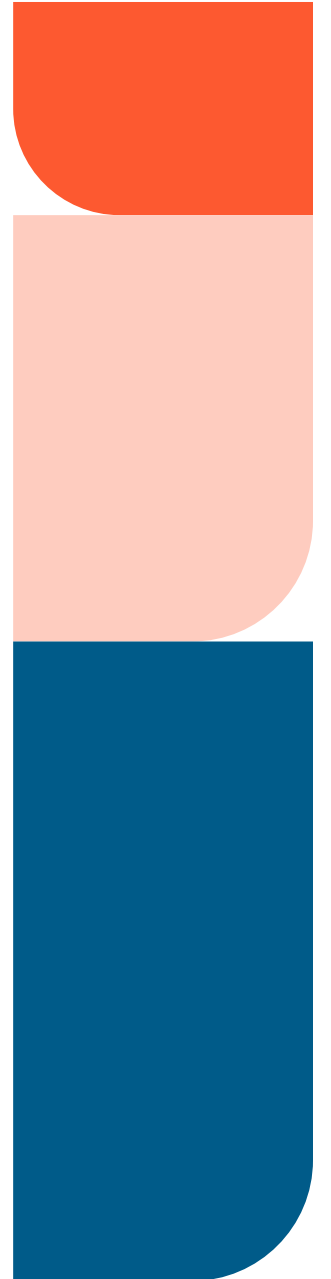
Vä förmakstumorer

- Påverkar blodflödet och mitralklaffen
- Liknande symtom som vid mitralklaffvitier
- Hjärtsvikt, 2nd PH (dyspne, orthopnea, paroxysmal nocturnal dyspne, lungödem, hosta, hemoptys, trötthet etc
- Kan vara lägesberoende
- Embolisering (neurologiska symtom som primära symtomet ~ 12%)
- Myxom vanligast



Hö förmak

- Flödesobstruerande liknande symtom som vid trikuspidalisstenos
- Klassiska symtom (trötthet, ödem, ascites, halsvenstas, leverförstoring)
- Lungembolisering
- Vid stigande tryck ev shuntning/embolisering via öppetstående foramen ovale

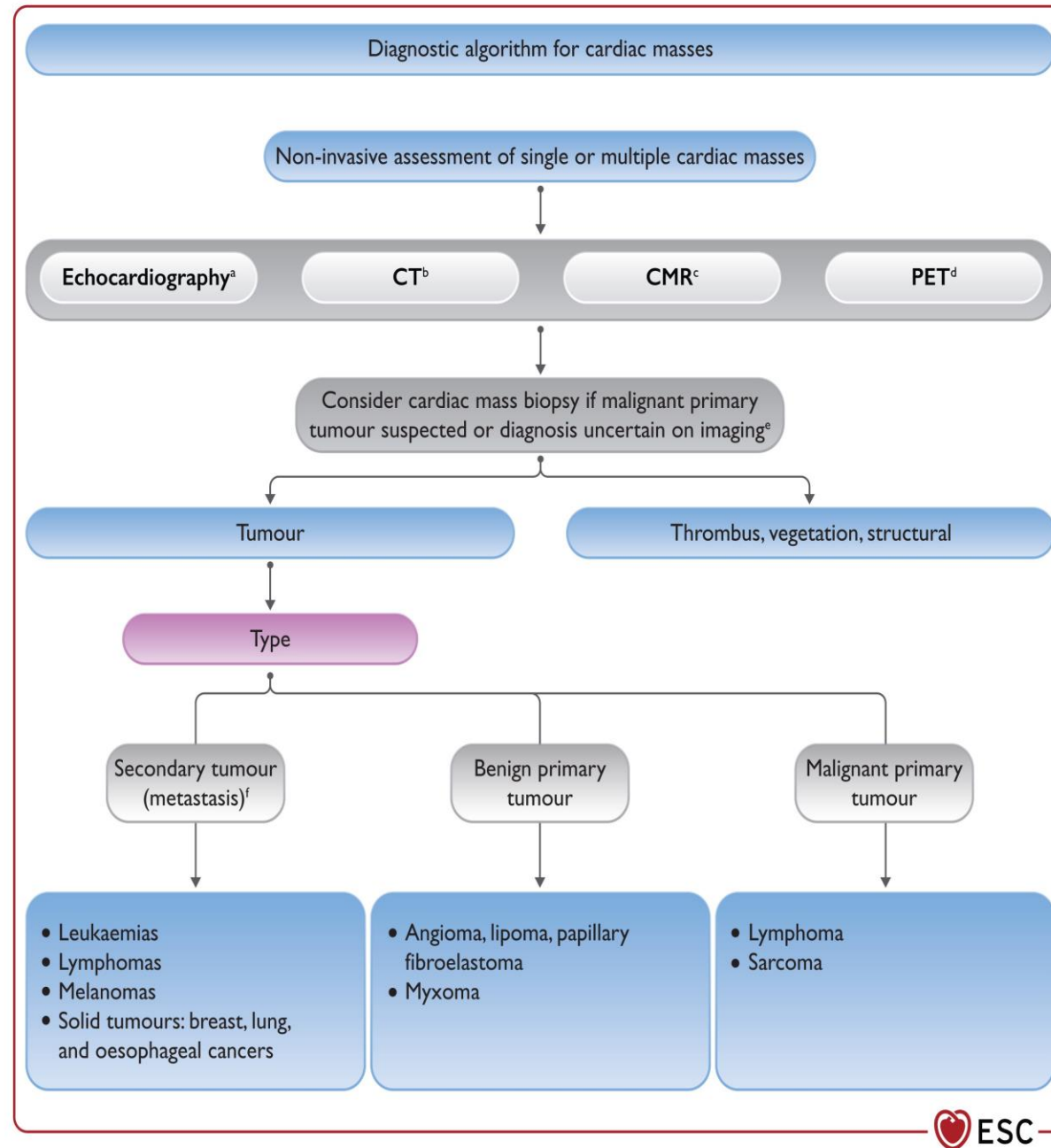


Hö kammare


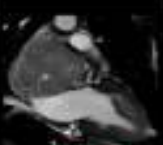







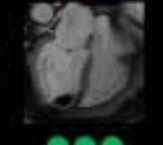
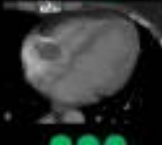



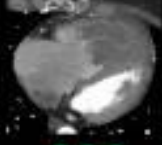





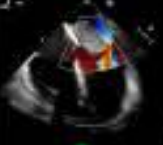

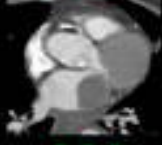
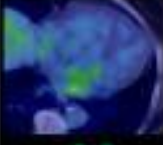
- HK-svikt (ödem, ascites, dyspne, synkope, plötslig död, halsvensstas, hepatosplenomegali)
- Diff diagnoser: pulmonalisstenos, restriktiv kardiomyopati, TI

Vä kammare

- Intramurala tumörer (arytmier, konduktionsstörningar)
- Intracavitära (VK-svikt, synkope, embolier)
 - Diff: tromb

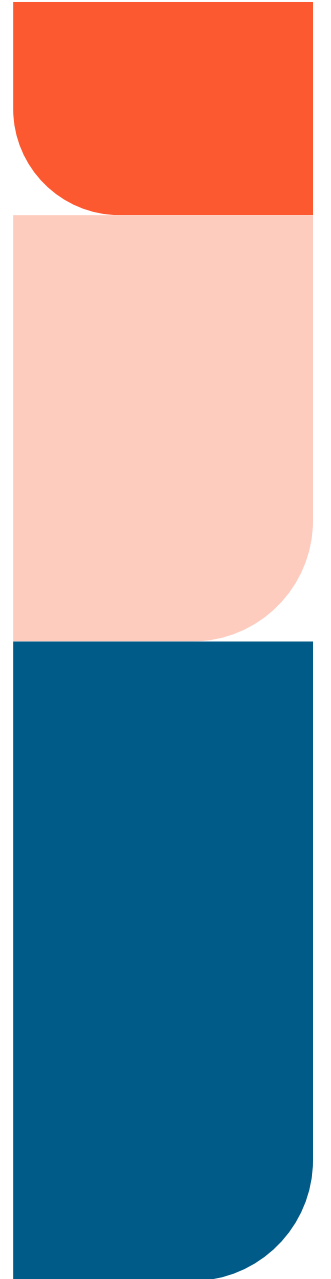


Diagnostic Accuracy of Multimodality Imaging in Characterizing Cardiac Masses

	ECHO	CMR	CT	PET
Detection and localization	 ●●●	 ●●●	 ●●●	
Identification of anatomical variants	 ●●●	 ●●●	 ●●	
Differentiate tumors from pseudotumors	 ●●	 ●●●●	 ●●●●	 ●
Detection of malignancy	 ●●	 ●●●●	 ●●●●	 ●●●●
Staging and guidance of treatment	 ●	 ●●	 ●●●●	 ●●●●
Pre-surgical planning	 ●	 ●●	 ●●●●	 ●●

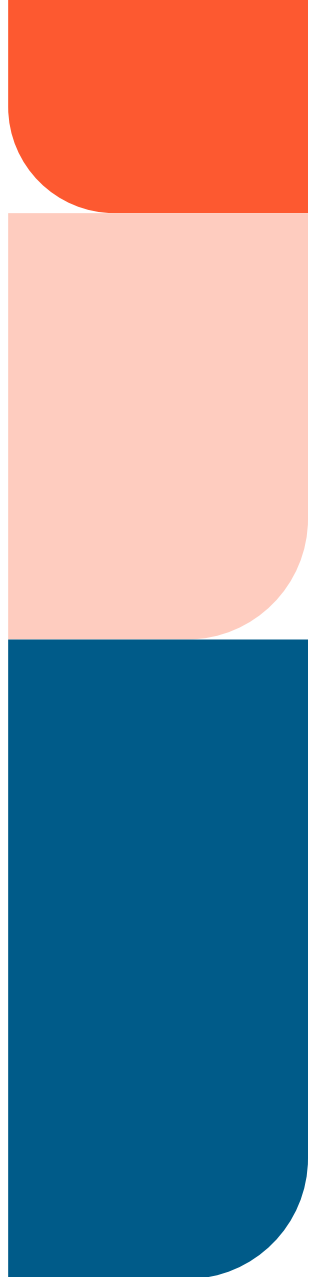
Utredning

- Primär/Sekundär
- TTE/TEE (var/storlek/hemodynamik)
- Röntgen (rtg-pulm, CT, MR, PET-CT)
 - Morfologi (stjälkade/bredbasiga/fast/fluktuerande)
 - MR: T1/T2 viktade sekvenser (tumörens kemiska mikromiljö)
 - PET-CT: utredning primär/2nd, staging; utvärdering av beh effekt
- Biopsi
 - Cytologi perikardvätska
- Coronarangio
 - Inför op
 - Engagemang coronarkärl, blodförsörjning epikardiella tumörer



Behandling

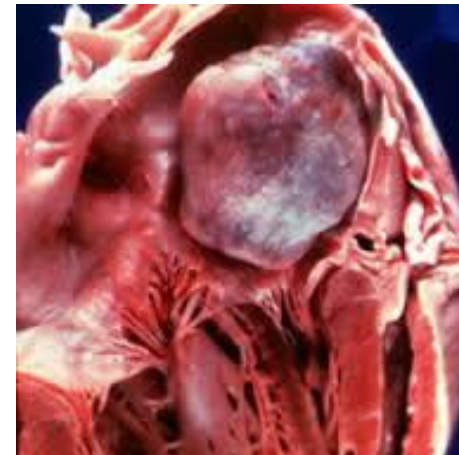
- Operation
 - Sternotomi vanligast
 - Myxom: semiakut ffa pga risk för emboliserin
 - Ofta kurativ vid myxom, recidiv extremt sällsynta
- Cytostatika ffa mot primärtumör
- Strålning



Myxom

- 80% av benigna tumörer
- Breddbasiga, "geléaktiga", från 1cm-15cm i diameter
- Embolier hos 35%, allmän sjd-känsla 35%,

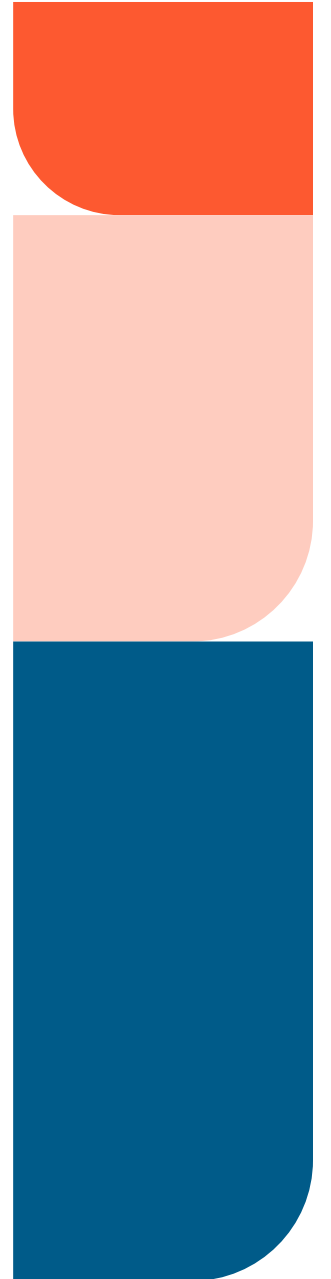
- Behandling: resektion
- 2-5% recidiv



Papillärt fibroelastom

- Näst vanligast primära hjärttumören
- 55% män, medelålder 60 år
- Stjälkade
- Storlek medel 9 mm (2-70mm)
- 80% engagerar klaffar
- Vanligaste primära symtomet: embolier (tumörvävnad/tromb)

Behandling: kirurgi alt exspektans beroende på embolirisk

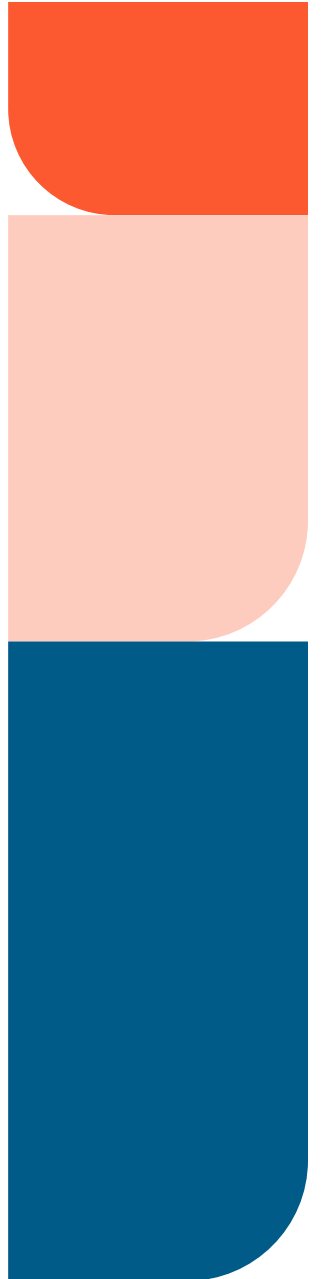


Rabdomyom

- Barn < 1år
- Ventrikulärt engagemang, försvinner ofta spontant och resektion är mycket sällan nödvändigt.

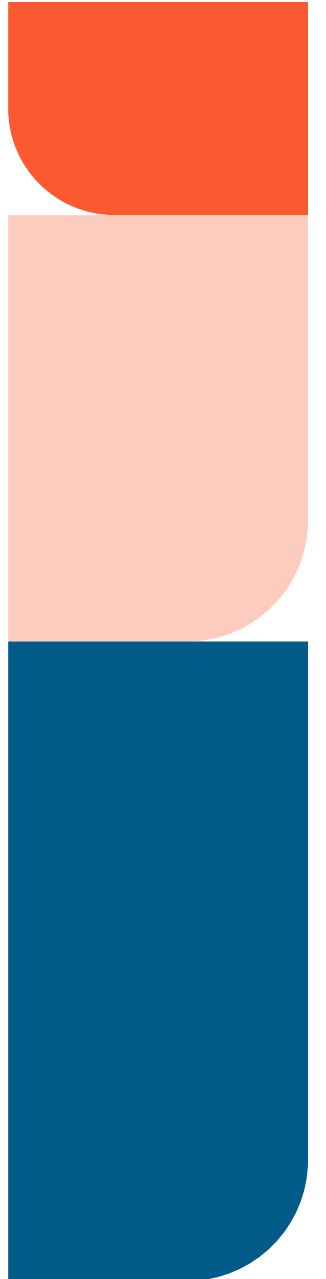
Fibrom

- Näst vanligast hos barn
- Ventrikulärt engagemang, blir ofta väldigt stora, resektion/trpl



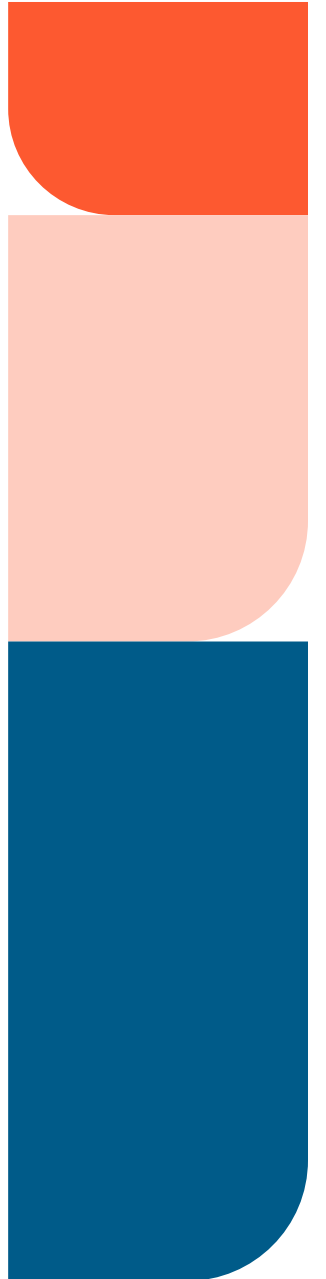
Sarkoma

- Maligna, 15% av primära hjärttumörer
- Angiosarkom > rabdomyosarkom > fibrosarkom > leiomyosarkom
- Dåliga resultat vid resektion
- Gjort försök till primär transplantation (!)
- Cytostatika, strålning
- Medelöverlevnad från diagnos < 1år

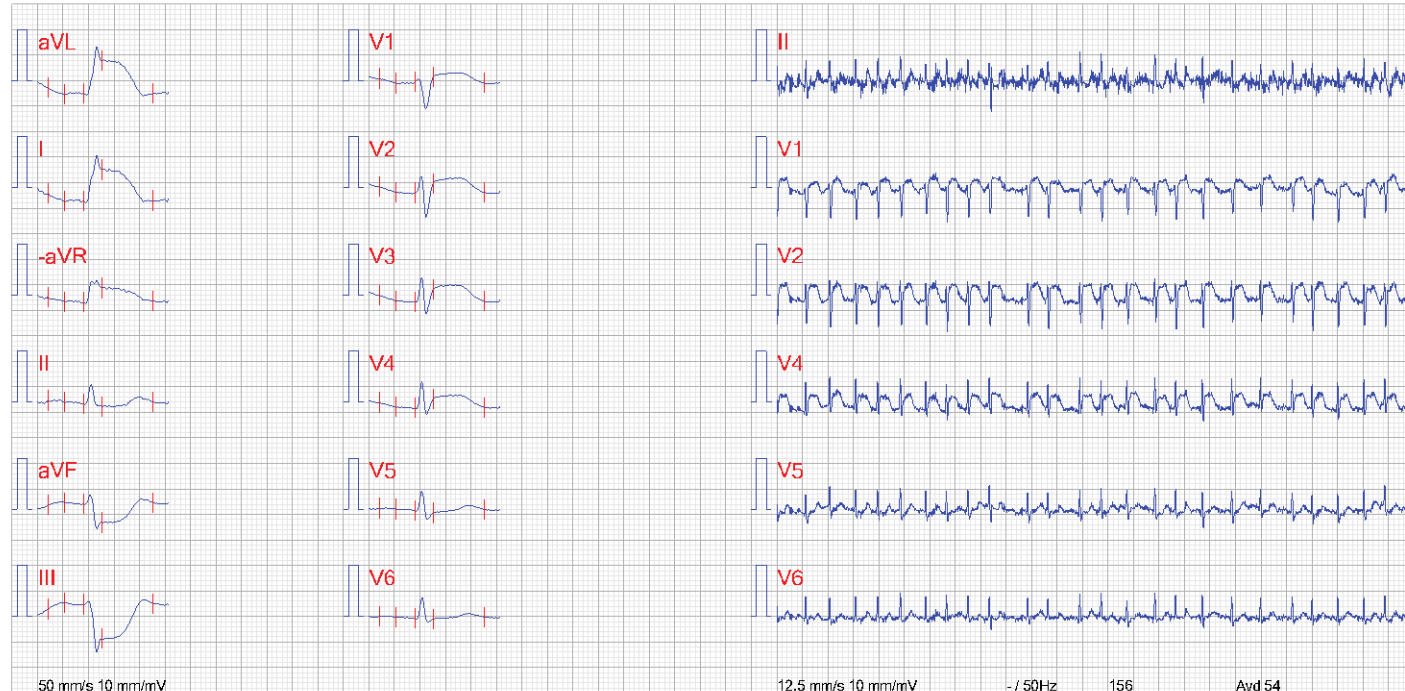


Metastaser i hjärta

- Melanom
- Lungcancer
- Bröstcancer
- Sarkom i mjukvävnad
- Njurcarcinom
- Esofaguscancer
- Levercancer
- Tyroideacancer



Fall1



50 mm/s 10 mm/mV

Datum:	2016-09-21 19:33:30	HF:	151 /min	P:	64 ms
Id:	[REDACTED]	BT:	-/- mmHg	PQ:	142 ms
Efternamn:	[REDACTED]	Längd:	- cm	QRS:	70 ms
Förnamn:	[REDACTED]	Vikt:	- kg	QT:	272 ms
Kön:	K	P-R-T-axlar:	5 / -15 / -21 °	QTc(H):	431 ms
Ålder:	74 år				
Föd.datum:	1942-05-03				
Operatör:					
Läkare:					

12,5 mm/s 10 mm/mV - / 50Hz 156 Avd.54

[REDACTED]

*** ÖVERVÄG AKUT STEMI ***
*** EXTREM TAIKYKARDI ***

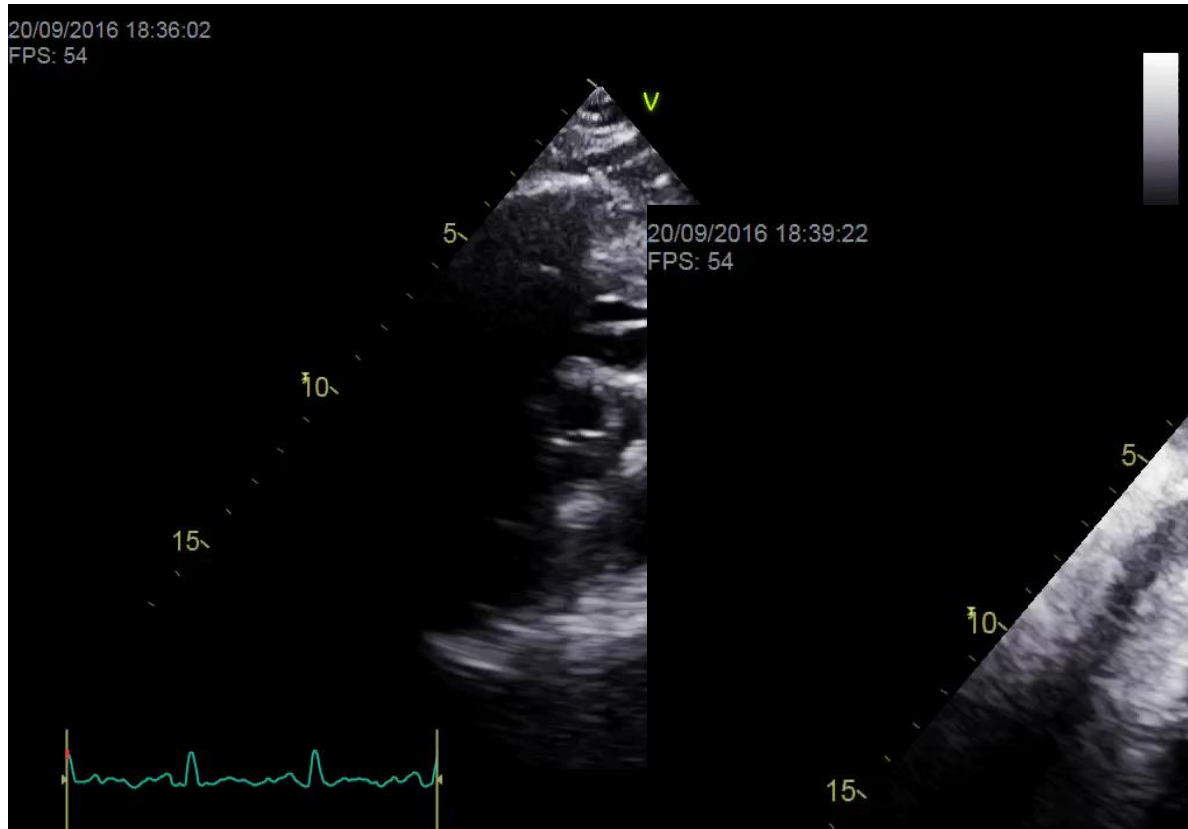
SINUSTAKYKARDI
VENTRIKULÄRA ES.
SUPRAVENTRIKULÄRA ES.

Utbredd ST-höjning.
MÖJLIG NYLIGEN GENOMGÅNGEN UTBREDD INFARKT.

ST-sänkning inom junctionområdet.
OSPECIFIKA ST-FÖRÄNDRINGAR.
FÖRÄNDRINGARNA KAN DELVIS VARA FREKVENSBETINGADE.

UCG

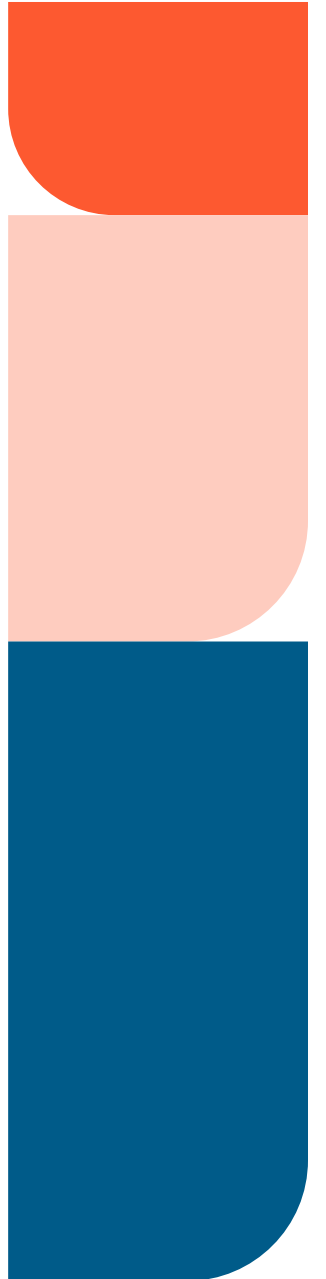


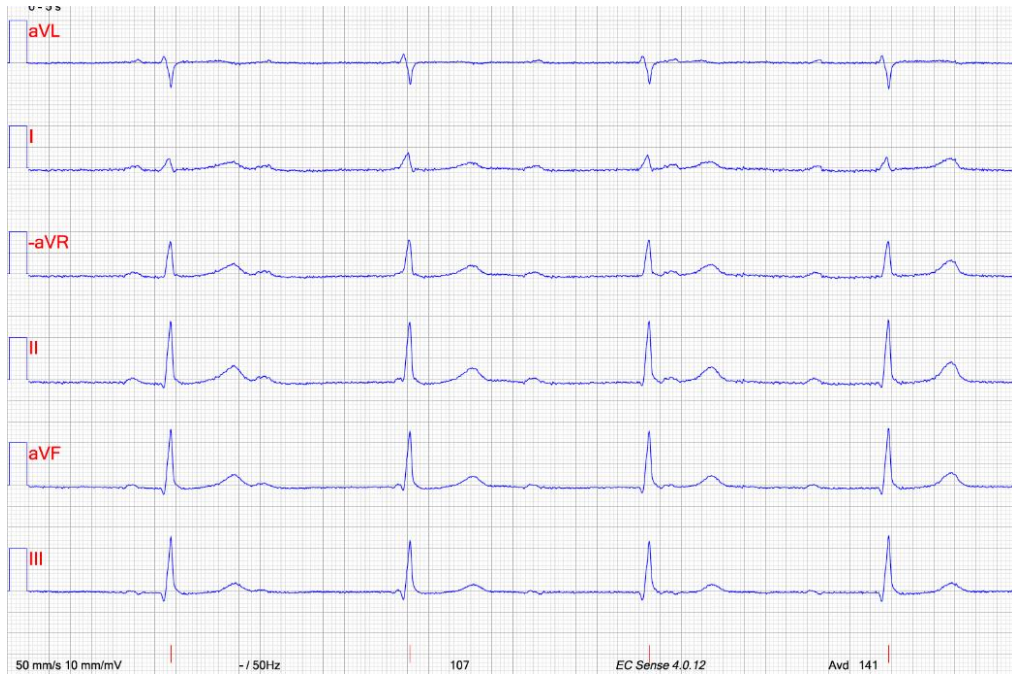


- Pat går Ad Mortum ngn vecka senare
- Troligtvis metastaser i hjärtat

Fall 2

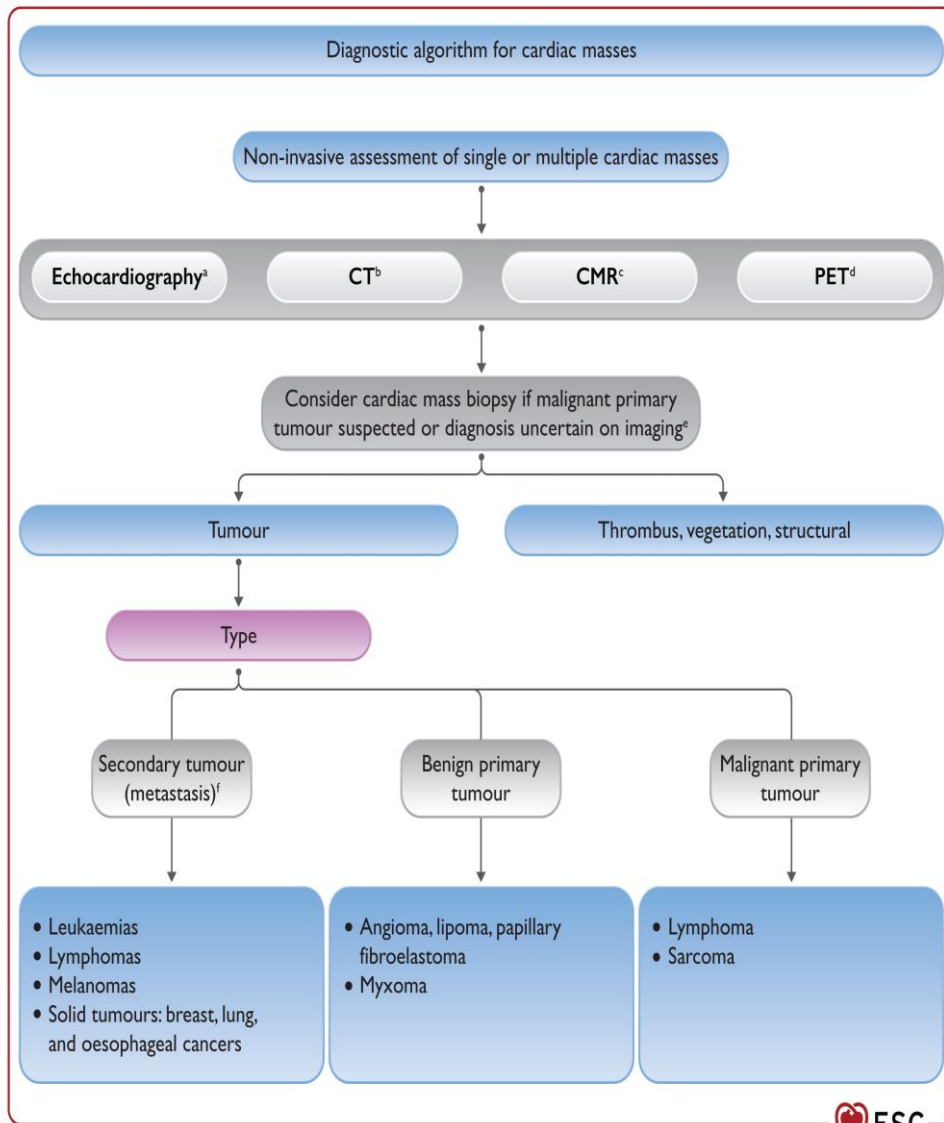
- 24 årig man Jan 2020
- Söker pga trötthet och allmän sjd känsla medicin
- Utredning visar anaplastisk storcelligt T-cellslymfom





UCG: Ingen perikardvätska. EF 60%, ingen hypokinesi. Normala slagvolymmer. Inga vitier av signifikans. Hö ua, PA-tryck ej stegrad. CVT < 5 mmHg





MR-hjärta:

Mjukdelsökning invid aortaroten som sträcker sig ner mot förmaksseptum, bakväggen av vä förmaket och basala delen av vä kammare.

Mjukdelsökningen mäter mellan aortaroot och vä förmak ca 1cm bred. Mjukdelsökningen i mellersta/apikala delen anterior av vä kammare 1x1,5cm med infiltrativ växt.

PET-CT:

*Ökat upptag i flertal **körtal** paraaortalt, vänster fossa supraklavikularis, halsen, lungor mm*

Ökat upptag intramuskulärt I illiacus, tonsiller
Ovanligt upptagsmönster i hjärtat med kraftigt förhöjt upptag apikalt i vä kammare, mellan förmak och aortaroten

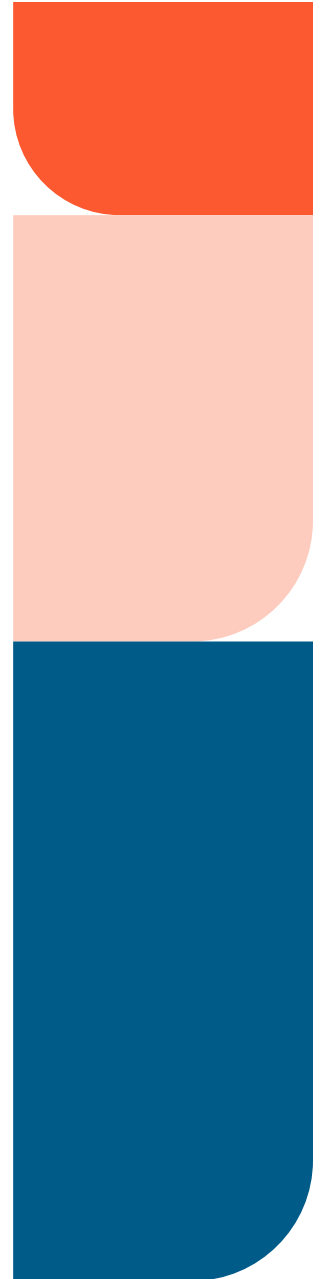
- MR-hjärta:
 - Mjukdelsökning invid aortaroten som sträcker sig ner mot förmaksseptum, bakväggen av vä förmaket och basala delen av vä kammare. Mjukdelsökningen mäter mellan aortaraot och vä förmak ca 1cm bred.
 - Mjukdelsökningen i mellersta/apikala delen anteriort av vä kammare 1x1,5cm med infiltrativ växt.

Behandling

AV-block III troligen pga ödem/tryck

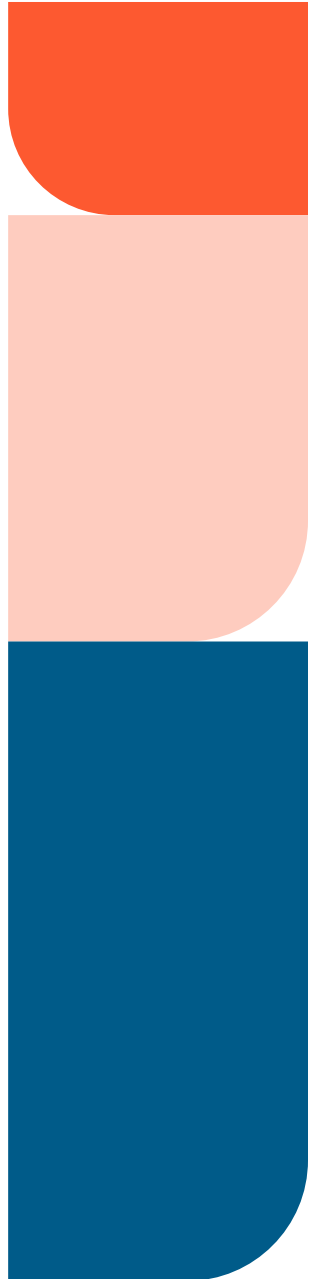
- Deltison + cellgifter (BV-CHEP)
- AV-block i regress 1 vecka senare

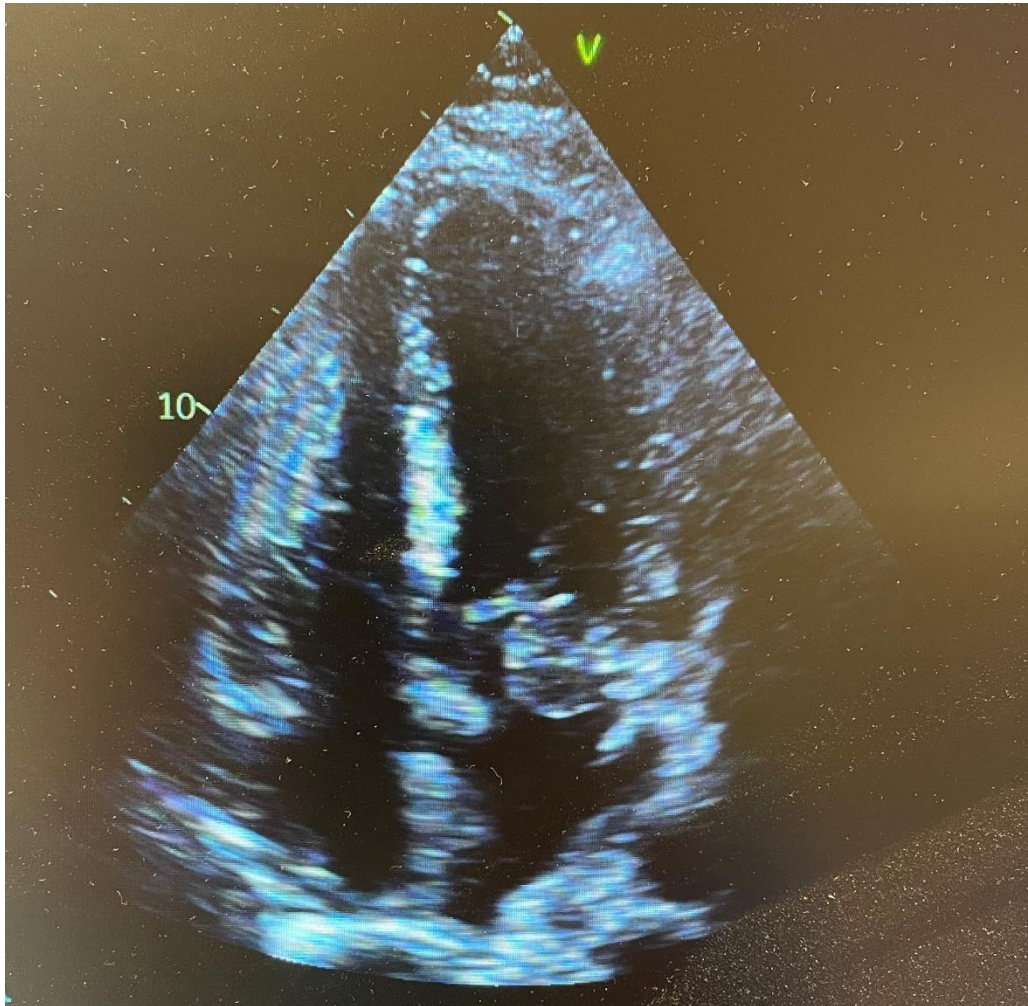
- Uppföljning via hematologen
 - ALK-positivt anaplastiskt storcelligt T-cellslymfom utbredd sjd stadium IV som behandlades med BV-CHEP 3 månader
 - NU "PET-negative"
 - Välmående



Uppföljning

- Rutinkontroll 13 månader senare
 - Välmående
 - Uppföljning med PET-CT: neg
 - CT-thorax/buk inga körtalar, däremot fynd i vä förmak
 - Vidare utredning med UCG





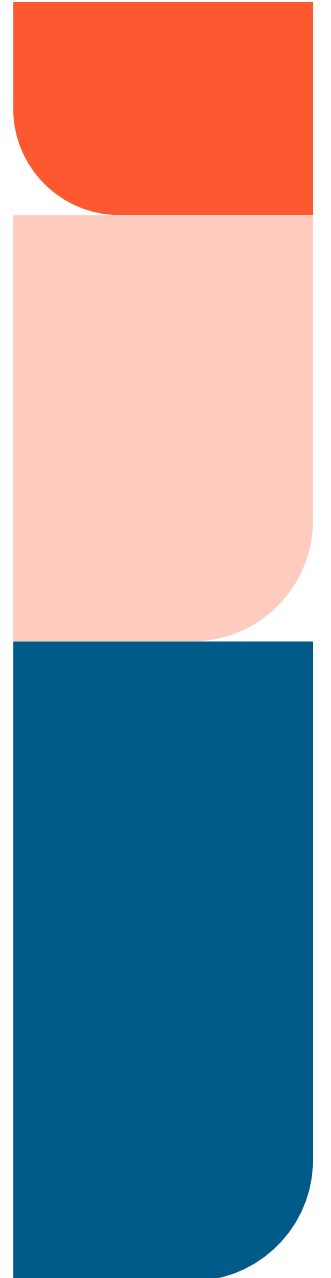
Rundad förändring vä förmak mätande
22x21mm

Progress sedan 1v

Förtjockning basalt vä kammare inferolateralt
mindre påtaglig jämfört med tidigare
undersökningar.

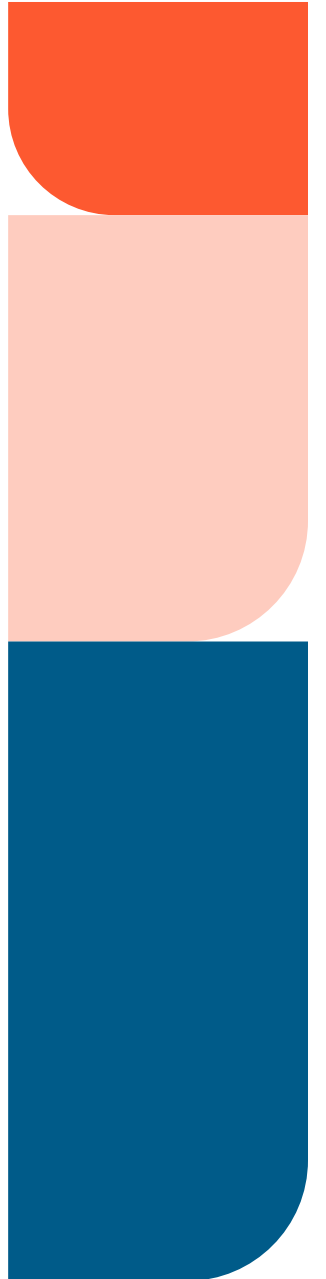
Vad göra?

- Biopsi
 - Vä förmak mot förändring x 2
 - Vä kammare inf/lat mot tidigare förtjockning
 - Normal myokard, inga tecken på tumor, inga tecken på inflammation
 - Indikation öppen thorax biopsi?
 - Vad göra?



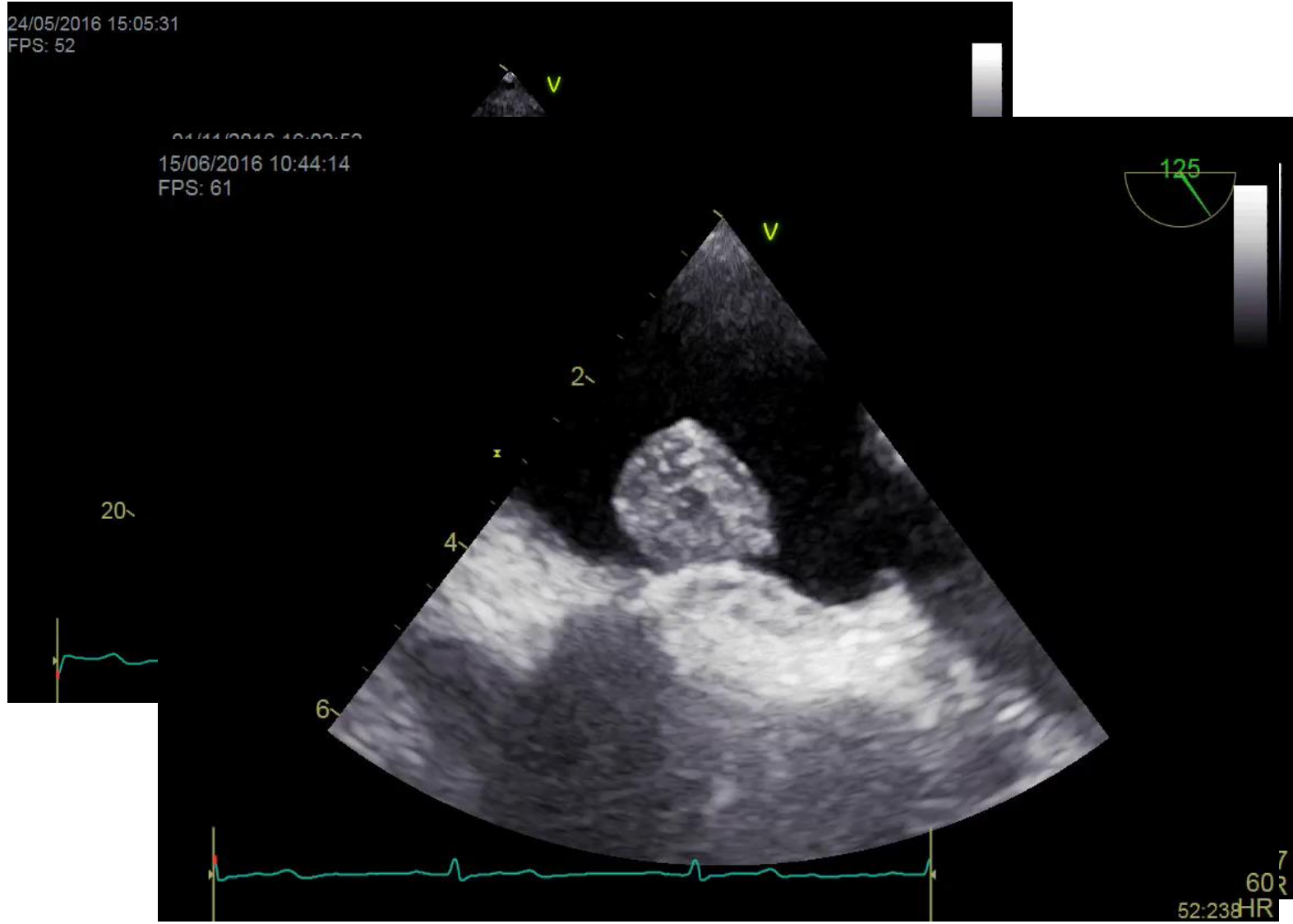
- NOAC

Total regress på 2 månader



Fall 3

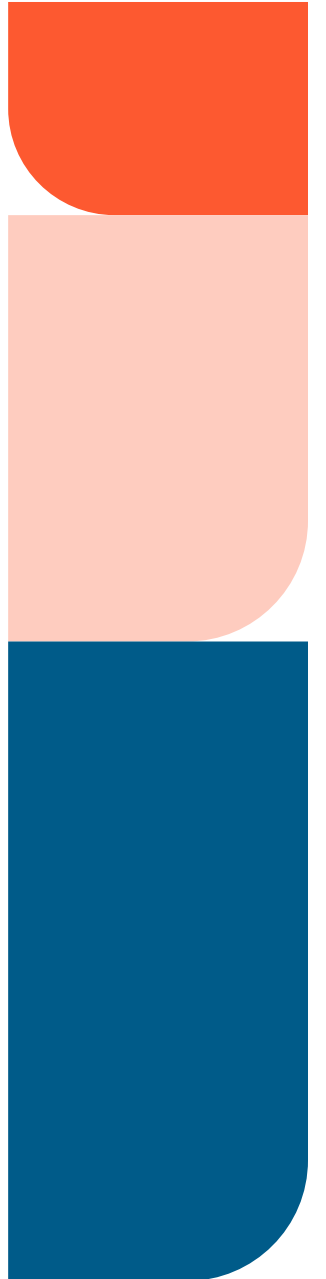
- 2004 söker akut pga centrala bröstsmärtor, hypotension
 - CT-aorta visar Typ A dissektion med stor AI
 - Akut operation med mekanisk klaff och kompositgraft
 - Waran.
-
- Välmående sedan dess, ffa kontakt med VC-läkare
 - Remiss UCG pga länge sedan förra undersökning, "kan vara bra att göra", symtomfri, välmående



Diagnos

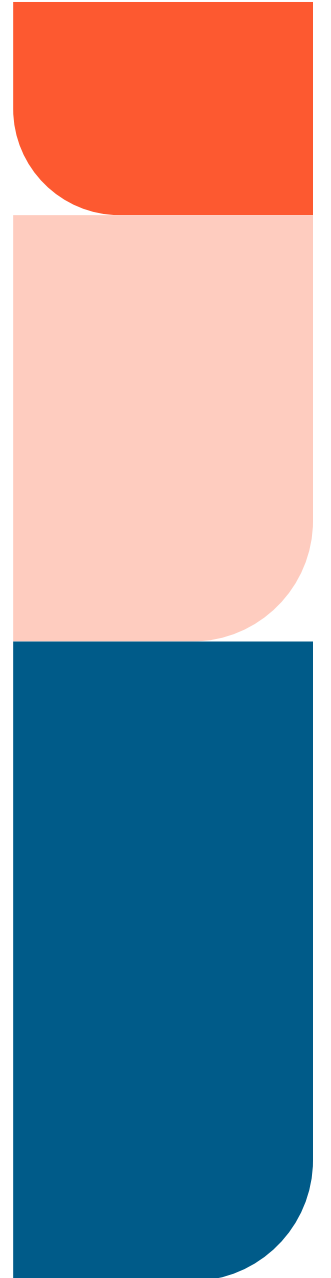
Tromb ej sannolikt, waran, terapeutiskt INR

- Lokalisation
- Myxom –bredbasig, geléliknande
- Subakut operation

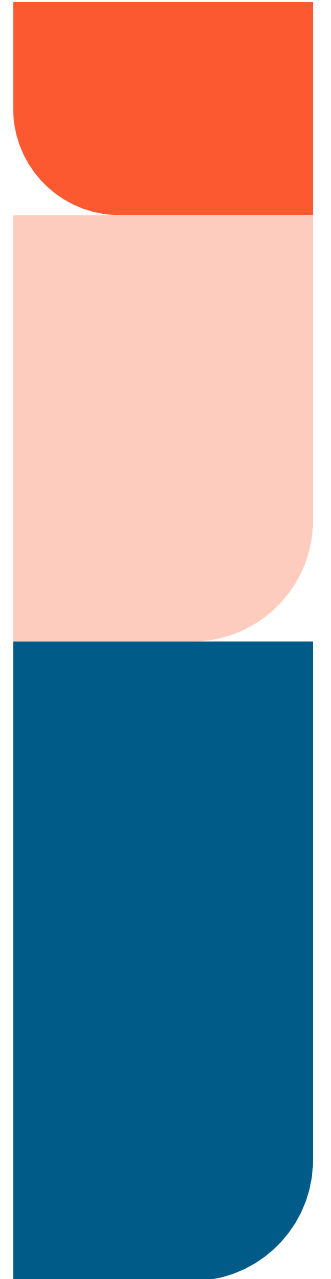


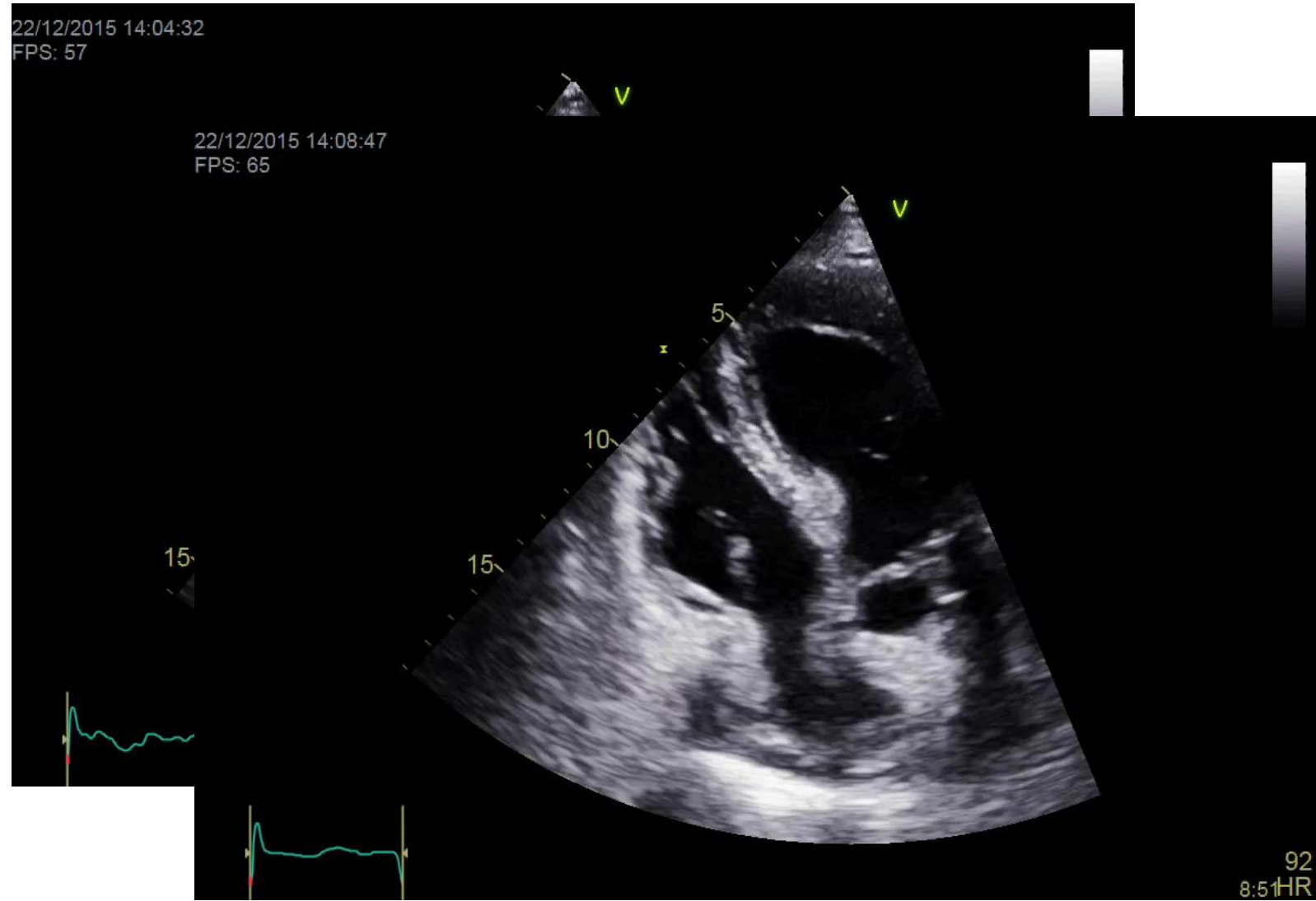
Fall 4

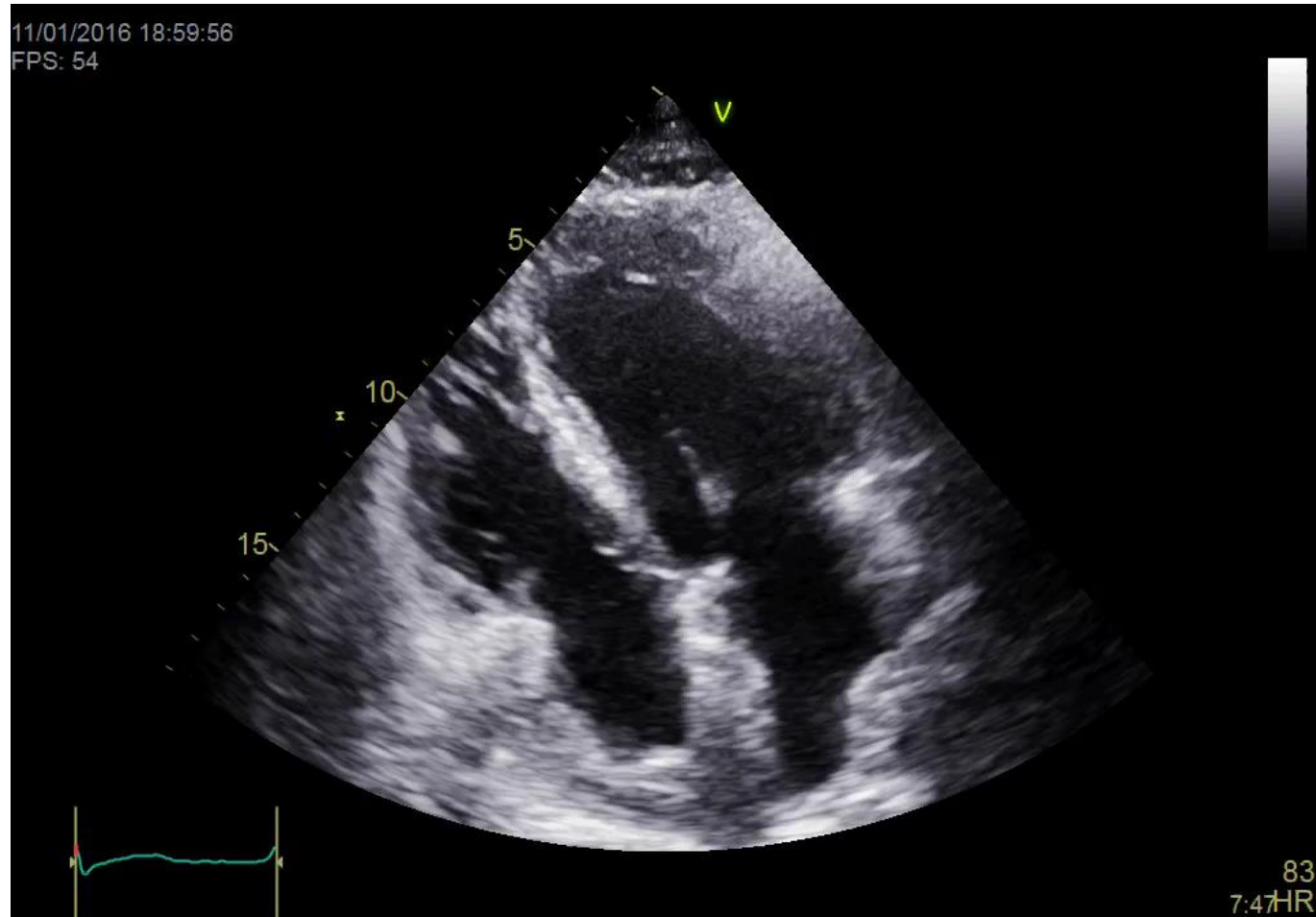
- Man 64år, tidigare väs frisk
- Ht 2013 tilltagande trötthet, söker VC
- Tolkas som utmattningssyndrom, sjukskriven (aldrig tidigare varit sjukskriven längre perioder.
- Progredierande trötthetsymtom, frekventa feberepisoder sommaren 2014
- Njurstensliknande smärtor Ht 2015, CT visar perirenal svullnad i fettet, förtjockad njurbäckenvägg. Tolkas som tubulointerstitiell nefrit.
- sommaren 2015 smärta i fotled – artrit , smärta knä - artros



- hösten 2015 nedsatt allmäntillstånd, dyspne, CRP 80, SR 45,
- CT-pleuravätska & perikardvätska
- Persisterande feber, utreds via reumatologen, ingen typisk systeminflammatorisk sjukdom, negativ RA, SLE etc
- Pga perikardvätska görs UCG

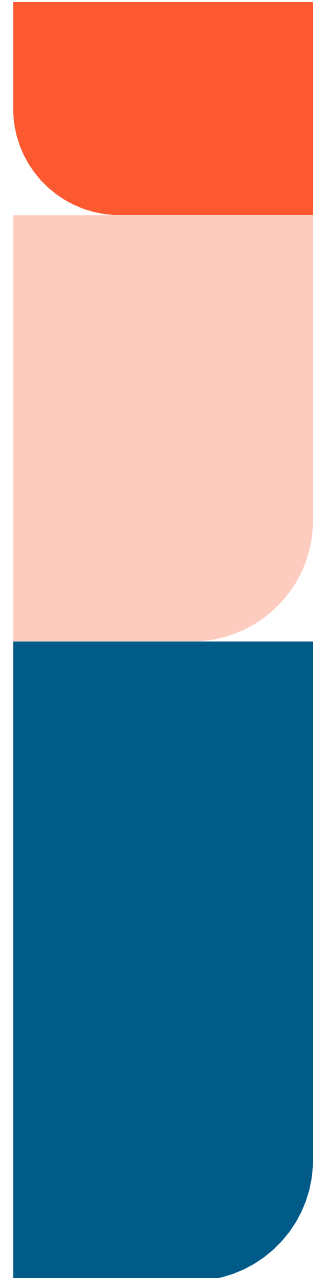




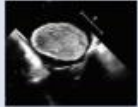
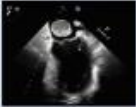







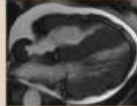


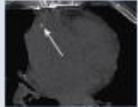


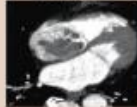

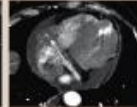

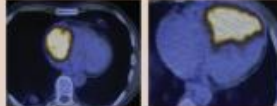









Utredning

- UCG/MR infiltrativt växande tumör: sarkom? angiosarkom?
- Hjärtbiopsi jan 2016 normala "myocyter, lindrig fibros"
- Öppen hjärtbiopsi mars 2016: "inflammatoriska celler IgG4 syndrom"
- PET-CT inflammatoriskt upptag hö maxillaris, pleura, hjärta, aorta retroperitonealt
- IgG4: lymfocytär infiltration med anrikning av IgG4-positiva plasmaceller och specifik fibros



CENTRAL ILLUSTRATION: Red Flags for Malignancy in Cardiac Masses Identified Through Multimodality Imaging

Multimodality Imaging in Cardiac Masses						
	Highly suspicious of a benign mass			Highly suspicious of a malignant mass		
Echo	Regular shape 	Pedunculated 	Left side 	Right side 	Sessile 	Pericardial effusion 
CMR	Homogeneous enhancement 	EGE - 	Regular margins 	Infiltration 	Cardiac mass LGE + 	Heterogeneous enhancement 
CT	Calcification 	Hypodense 	Regular margins 	Irregular margins 	Isodense 	Infiltration 
PET	Radiotracer uptake -/+ 			Radiotracer uptake ++/+++ 		
Assess the risk of cardiac mass embolism and hemodynamic impact, and patient clinical status						
Treatment Strategy: Which and When?						
	Consider anticoagulation* 	Surgery mass excision 	Follow-up 	Neoadjuvant chemotherapy 	Surgery 	Adjuvant chemotherapy 

Angeli F, et al. JACC CardioOncol. 2024;6(6):847-862.

TACK!

