Аортна Дисекация при Системен Лупус Еритематодес

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XIV
НАЦИОНАЛЕН КОНГРЕС ПО КАРДИОЛОГИЯ
2-5 ОКТОМВРИ 2014
ВАРНА, К.К. ЗЛАТНИ ПЯСЪЦИ

Представяне на пациента

- 42 годишна жена
- 1987 → ***Системен Лупус Еритематодес***

  ОБН → 2 месеца хемодиализа, плазмафереза, пулсова терапия с кортикостероиди и циклофосфамид- клинико-лабораторна ремисия
- 2008 → Влошаване на бъбречната функция

  Бъбречна биопсия- мезангиален лупусен нефрит, подобрение след терапия
- 1987-2014 системно лекувана и проследявана в Клиниката по нефрология

Придружаващи заболявания

- 09.2012 → Карцином на млечната жлеза, оперативно лечение, лъче- и химиотерапия, в ремисия
- Аваскуларна некроза и операция на десния медиален кондил на тибията и левия латерален глезен

Кардиологични проблеми

- 1999 → След II бременност и I нормално раждане → трайно повишени стойности на кръвното налягане (и на креатинина)

  TTE → Дилатация на асцендентната аорта с аортна регургитация

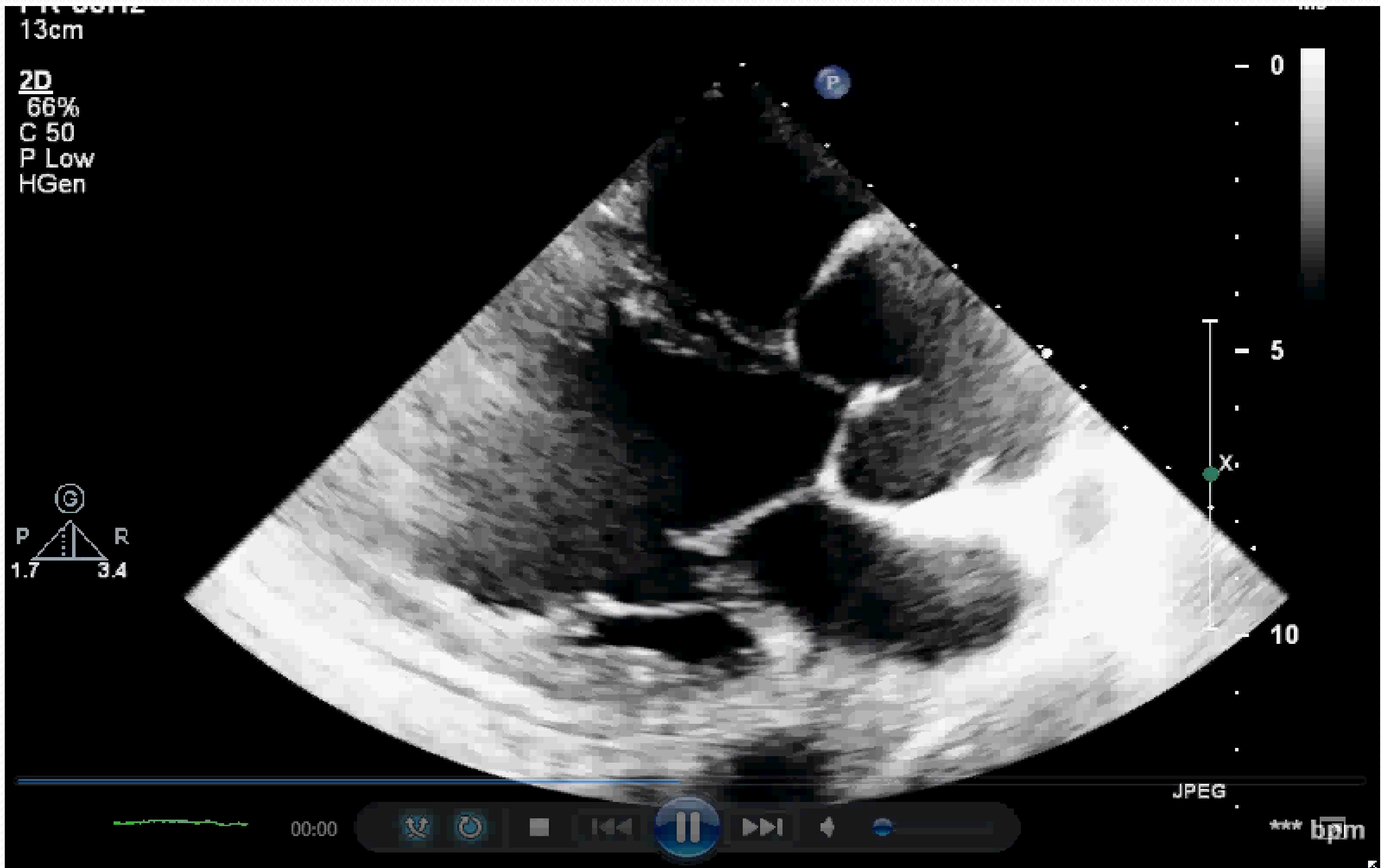
  Без оплаквания от гръдна болка/дискомфорт, без прояви на сърдечна недостатъчност

  Не е проследявана от кардиолог

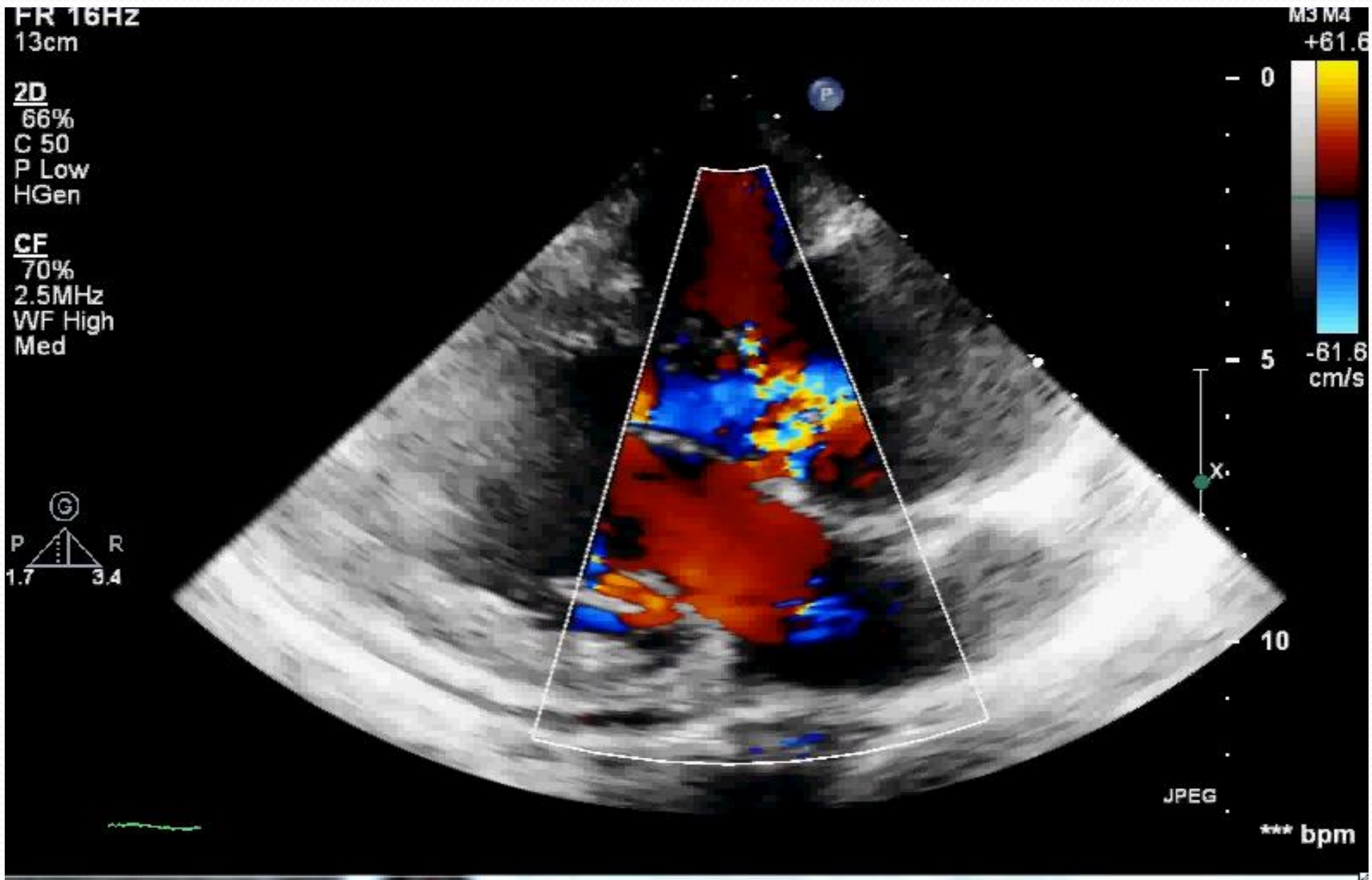
Хоспитализация

- 23.07.2014 постъпва в Клиниката по нефрология за оценка на бъбречната функция и лечение на лупусния нефрит
- АН ≈ 120/80 mm Hg, СЧ-65/мин.
- Лаб. изследвания: креа 239 мкмол/л, урея 15 ммол/л
Хб 113 г/л, Анти-ds ДНК ↑
- Терапия: ***Метилпреднизолон (пулс) 3 по 250 мг***
Циклофосфамид 400 мг
Небиволол таб. 5 мг, ½ таб. на ден
Триметазидин таб. 35 мг, 2 по 1 таб. на ден
Фолиева киселина таб. 0.4 мг, 1 таб. на ден
- Консултация с кардиолог

TTE



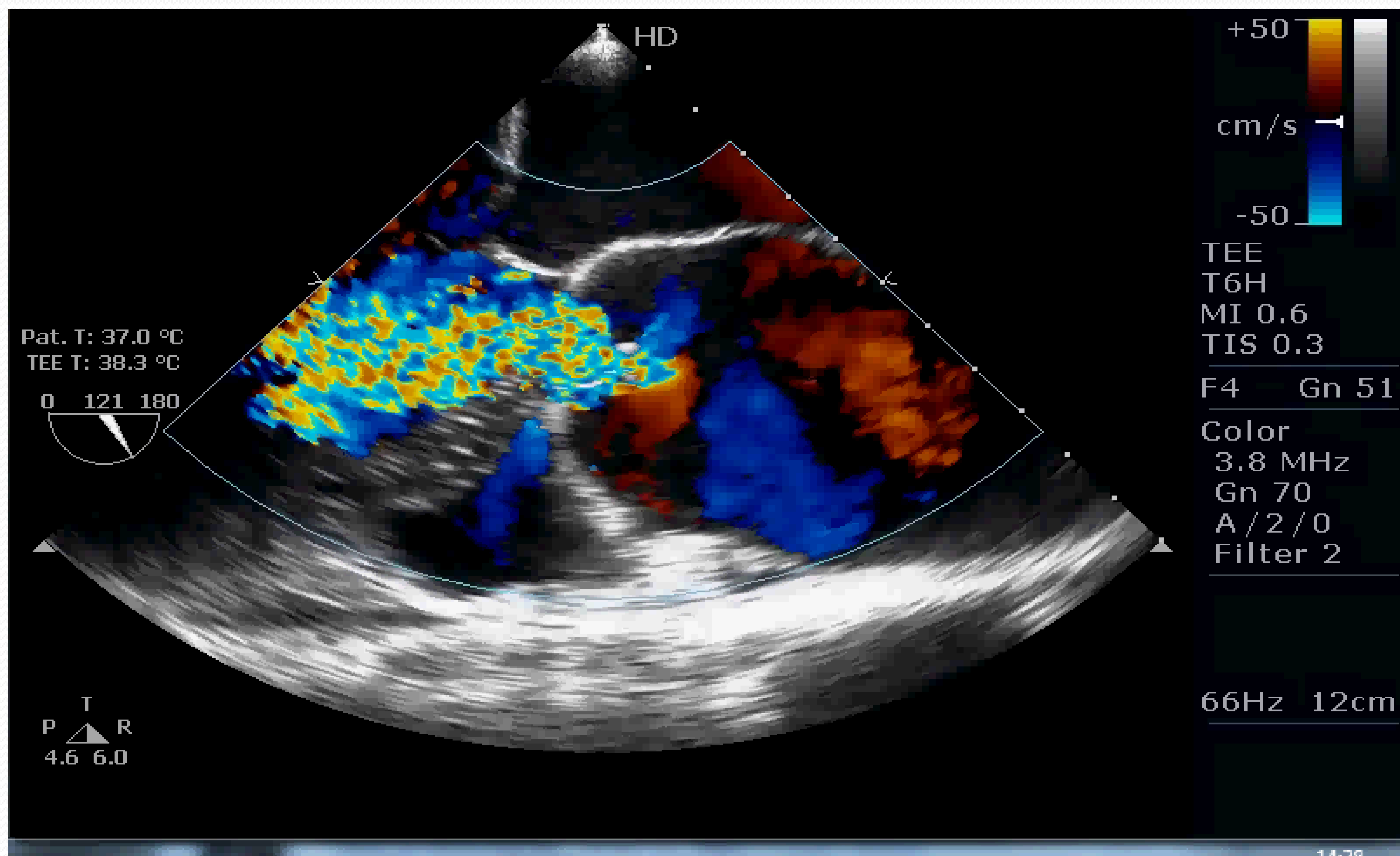
TTE



TEE

7/29/2014 PHILIPS
UMBAL Queen Joanna 10:20:09 AM
HD
Length = 5.19 cm
Pat. T: 37.0 °C
TEE T: 37.9 °C
0 121 180
TEE
T6H
MI 0.8
TIS 0.9
F4 Gn 51 5
232dB/C2 2
E/2/1
66Hz 14cm
T
P R
4.6 6.0
73
BPM

TEE



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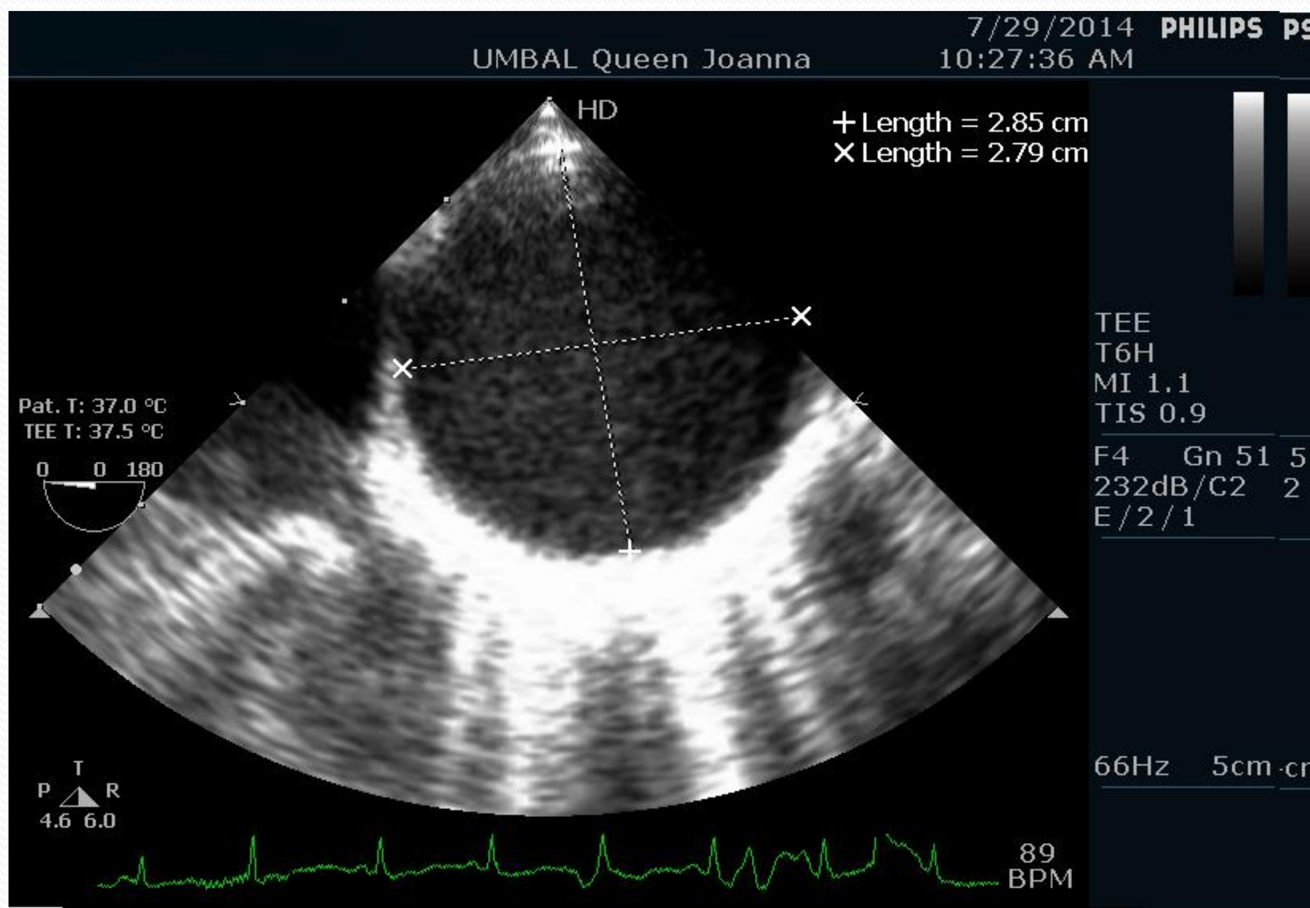


Как бихте постъпили?

Млада жена
СЛЕ с чести екзацербации
Лупусен нефрит с бъбречна недостатъчност
Дилатация на аортата с аортна регургитация
АХ, без прояви на СН

Как бихте постъпили?

- Образна диагностика
  - КТ
  - ЯМР
- Спешна кардиохирургия
- Планова кардиохирургия
- Наблюдение

30.07.2014

- 7.00 ч. силна болка в гърдите с ирадиация към гърба
- Запазена хемодинамика

(АН 130/80 mm Hg, СЧ- 73/мин, Сат-98%)

ЕКГ

HR 52/min
Intervals:
RR 1155 ms
P 114 ms
PR 132 ms
QRS 92 ms
QT 486 ms
QTC 454 ms

Axis:
P 42 °
QRS -13 °
T 73 °

P (II) 0.15 mV
S (V1) -1.47 mV
R (V5) 2.53 mV
Sokol. 4.45 mV

SINUS RHYTHM
LEFT ATRIAL ABNORMALITY
LEFTWARD AXIS
ST & T ABNORMALITY, CONSIDER
LATERAL ISCHEMIA OR LEFT VENTRICULAR

5.78

UNCONFIRME

Age:

cm / kg

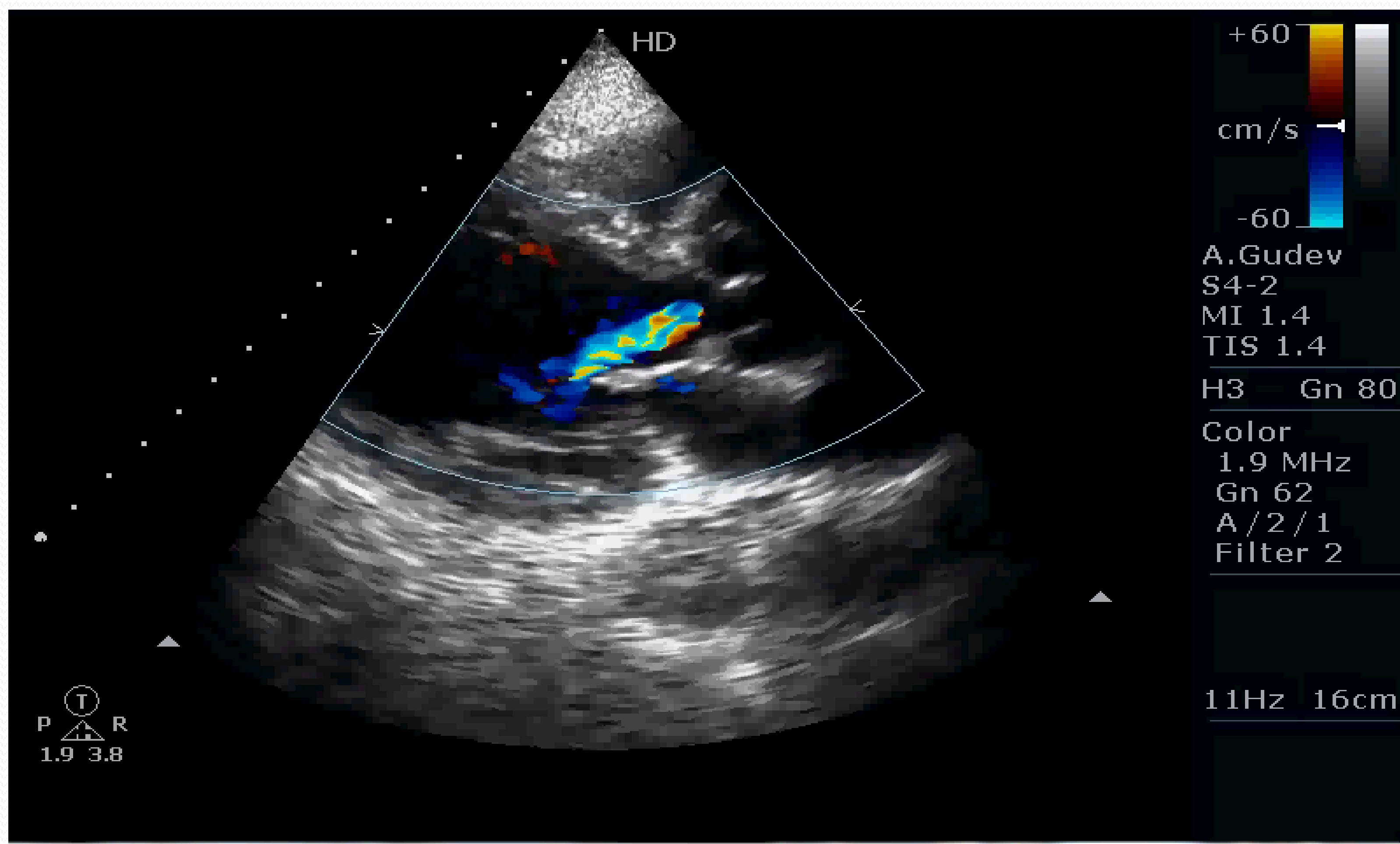
M / F

10 mm/mV

I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

25 mm/s 0.05-35Hz F50 SSF SBS We 30-JUL-14 07:04:55 UMBAL Ca Ica Yoanna - ISUL

TTE



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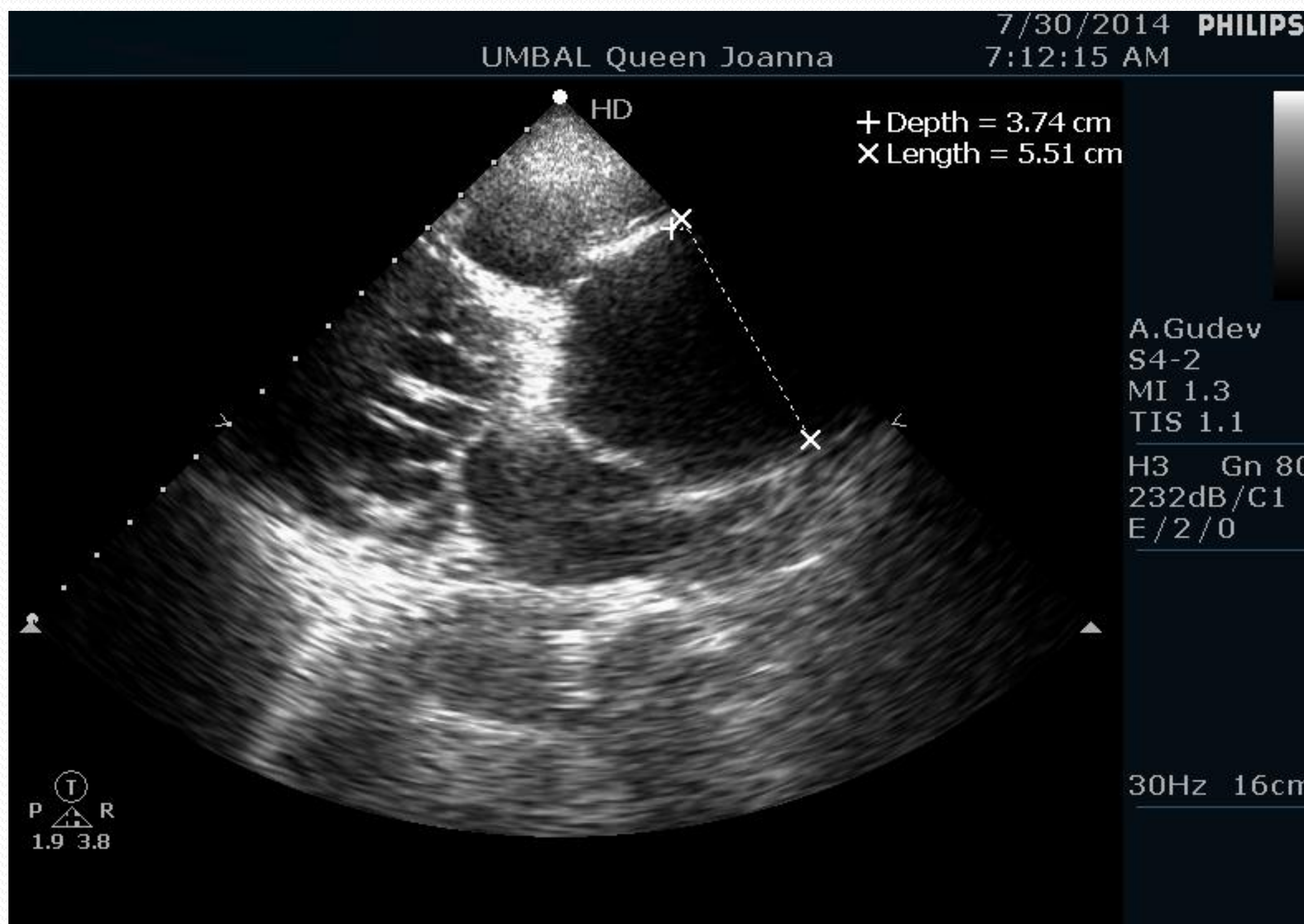

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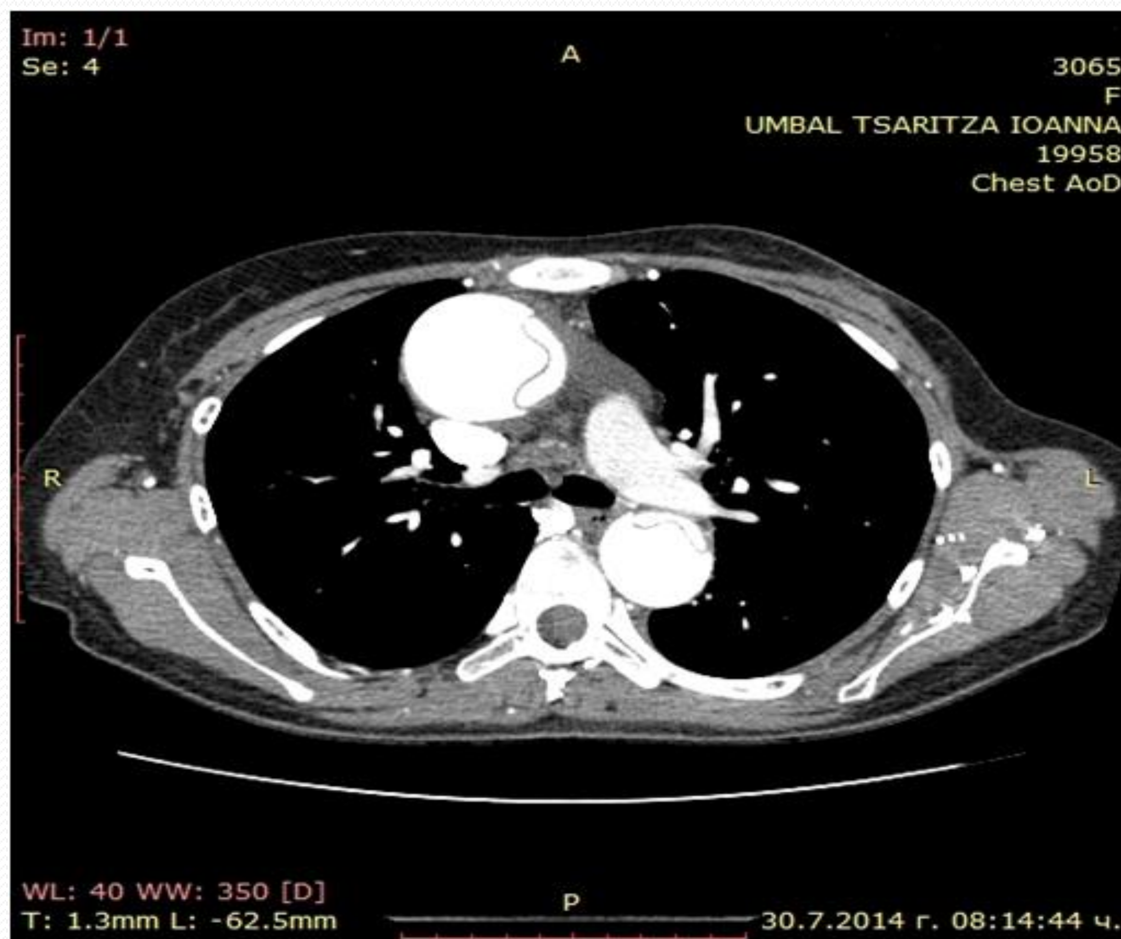

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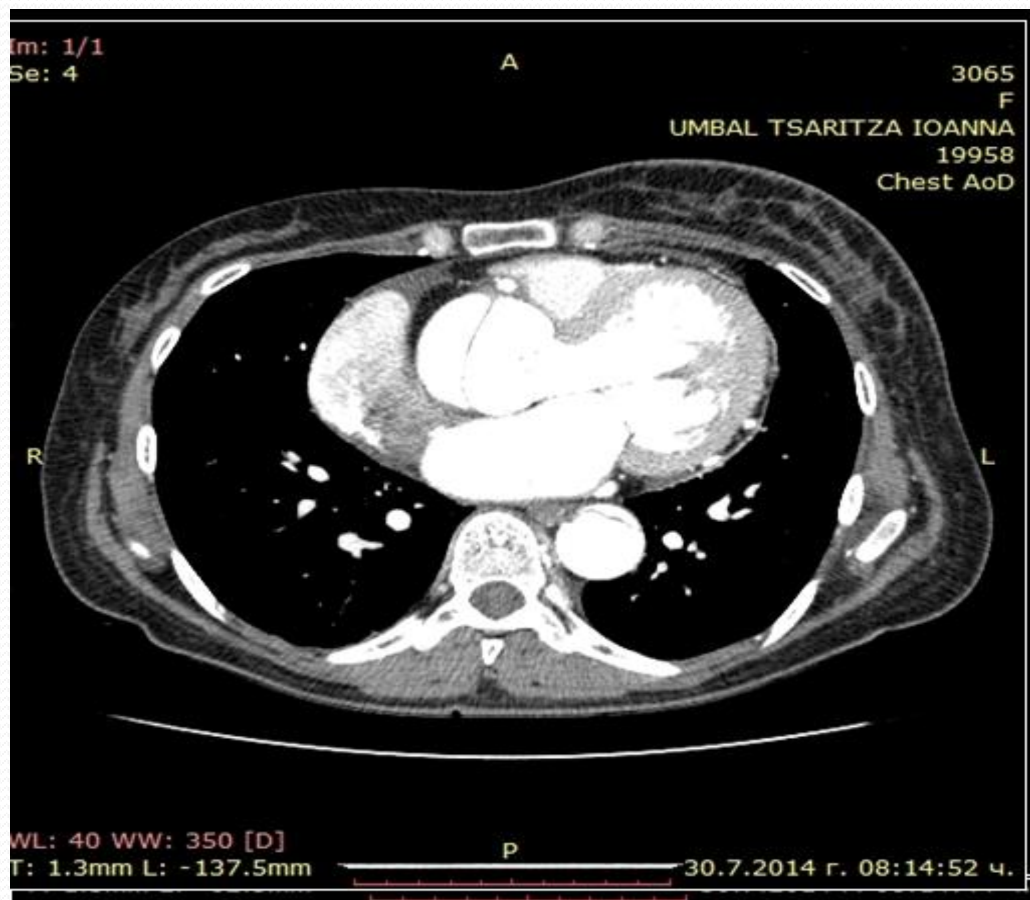

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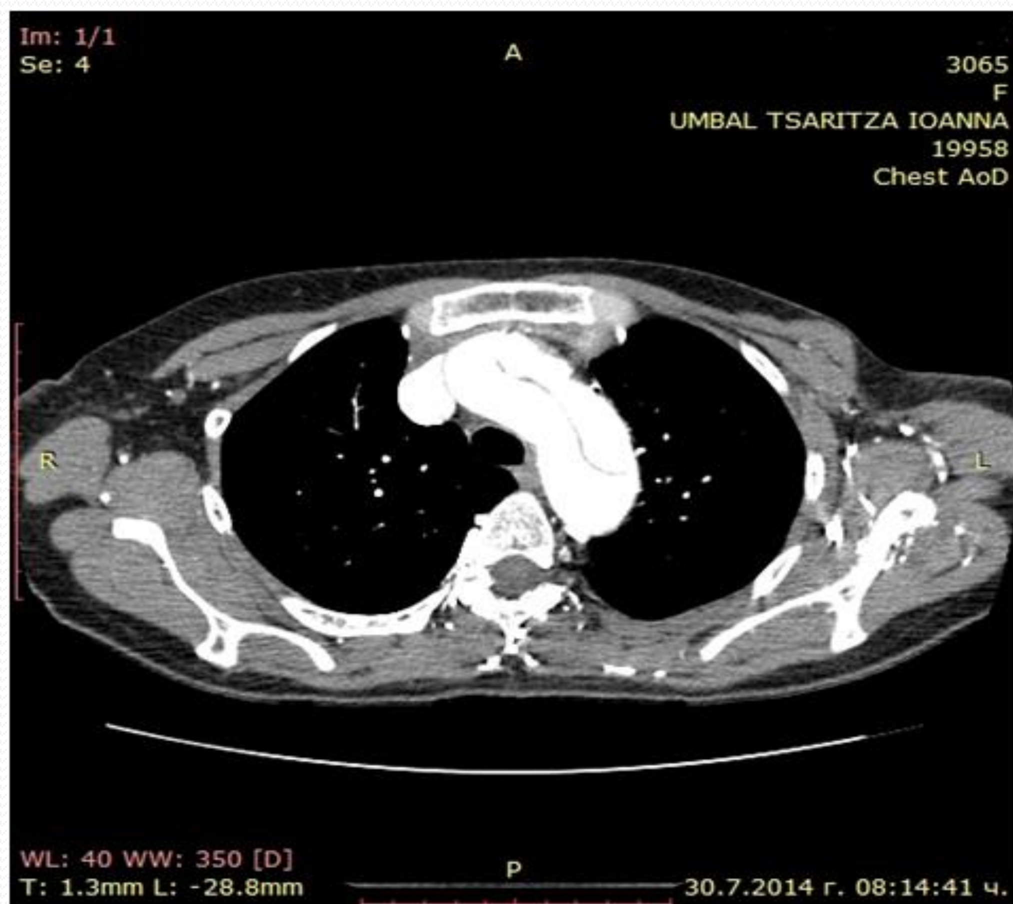

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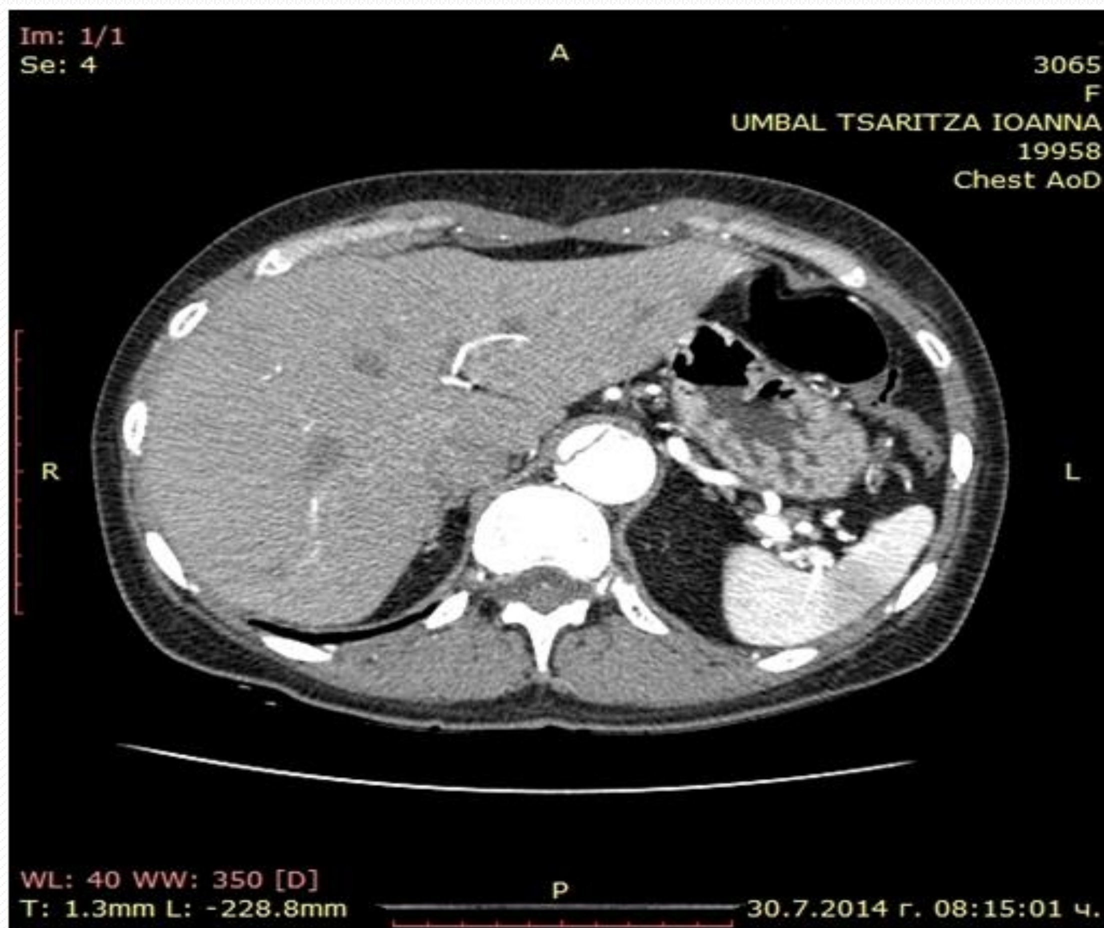
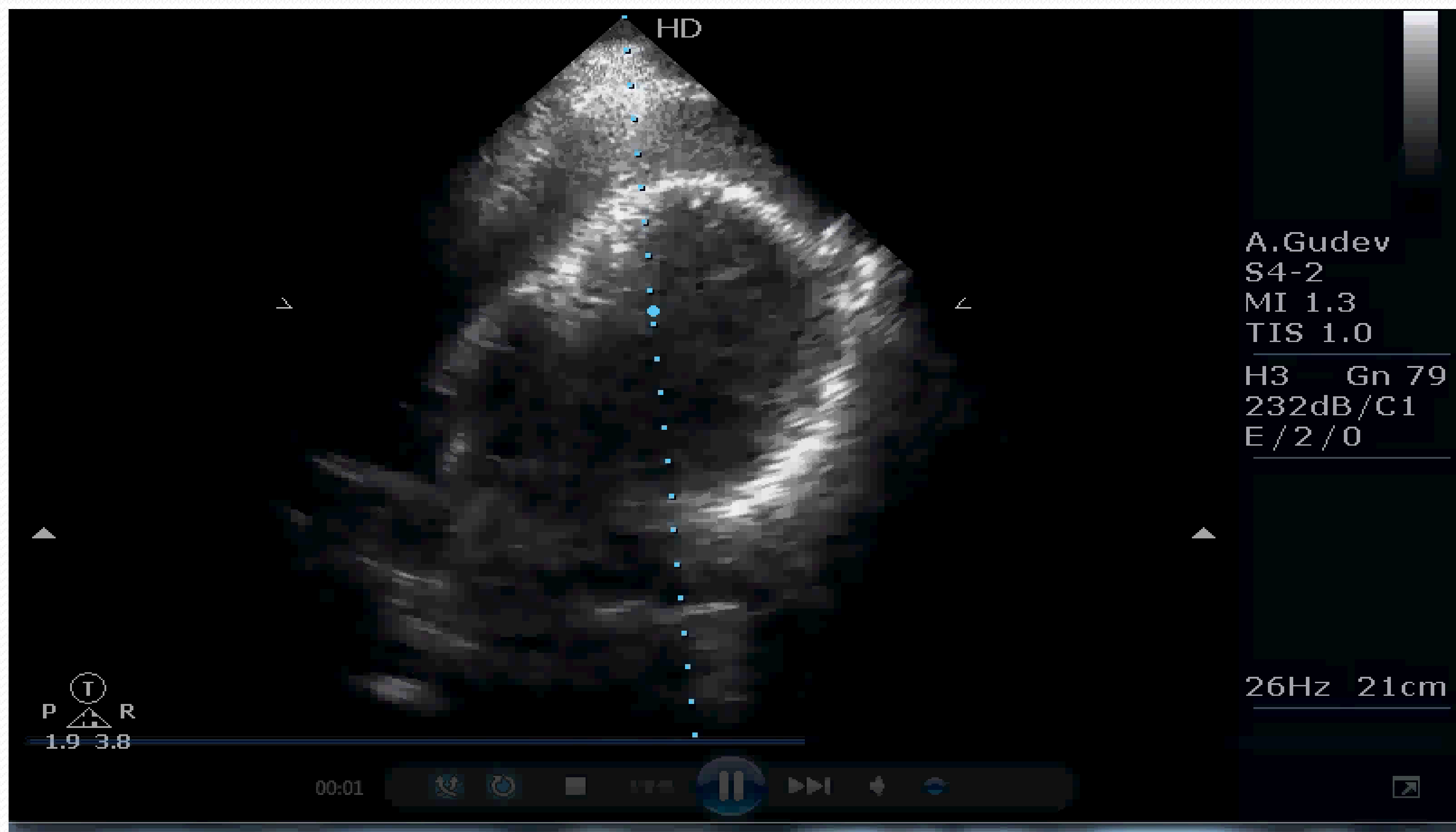


ЕМД – Перикардна тампонада



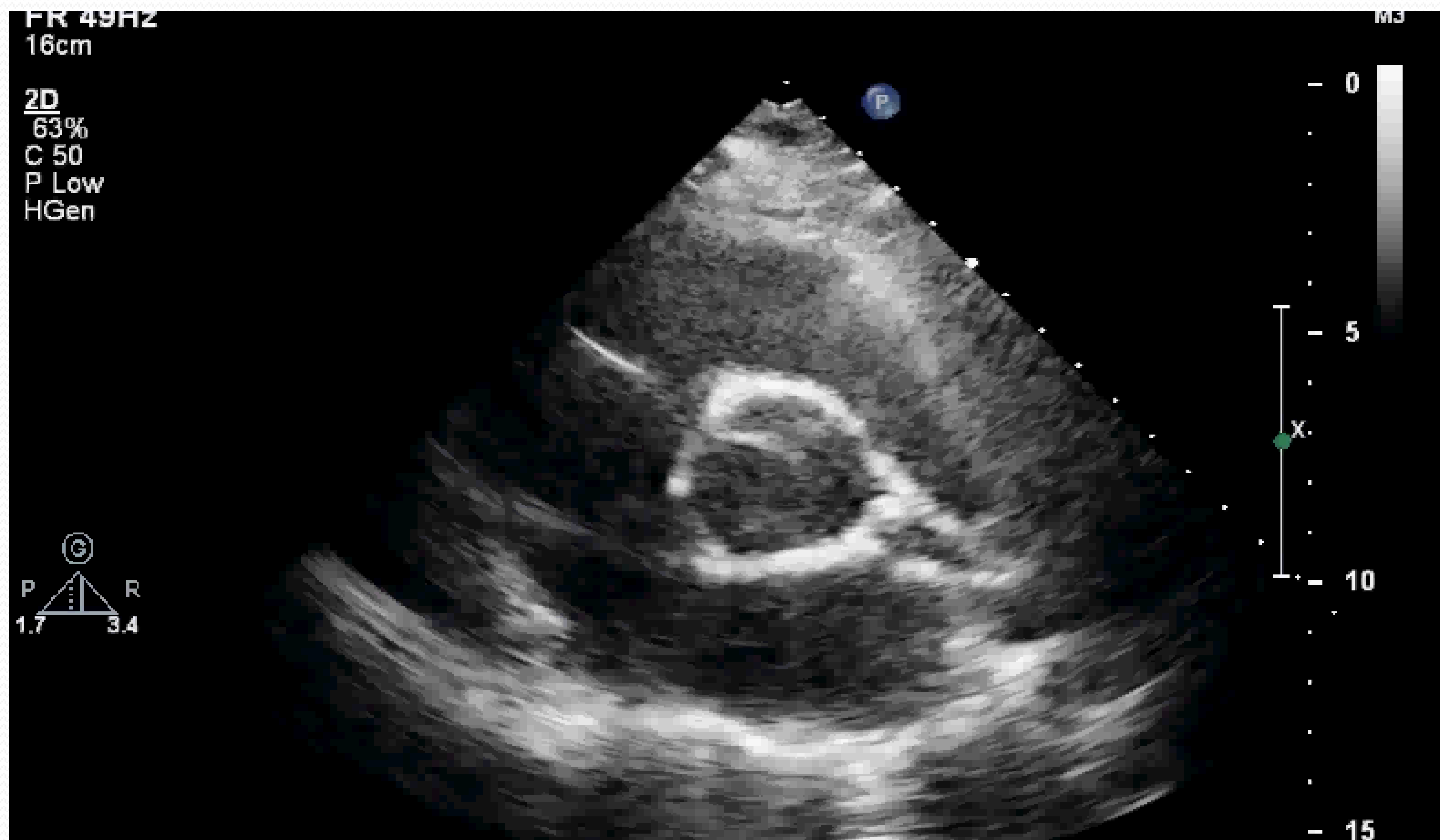
2 месеца по-късно...

- 41 годишна жена
- 2000- СЛЕ
- 2001- ПББ- мембранозна лупусна нефропатия- нефротичен синдром и нормални стойности на креатинин и урея-лечение с КС и цитостатици
- Вторична артериална хипертония
- Кортикостероид-индуциран захарен диабет

TTE

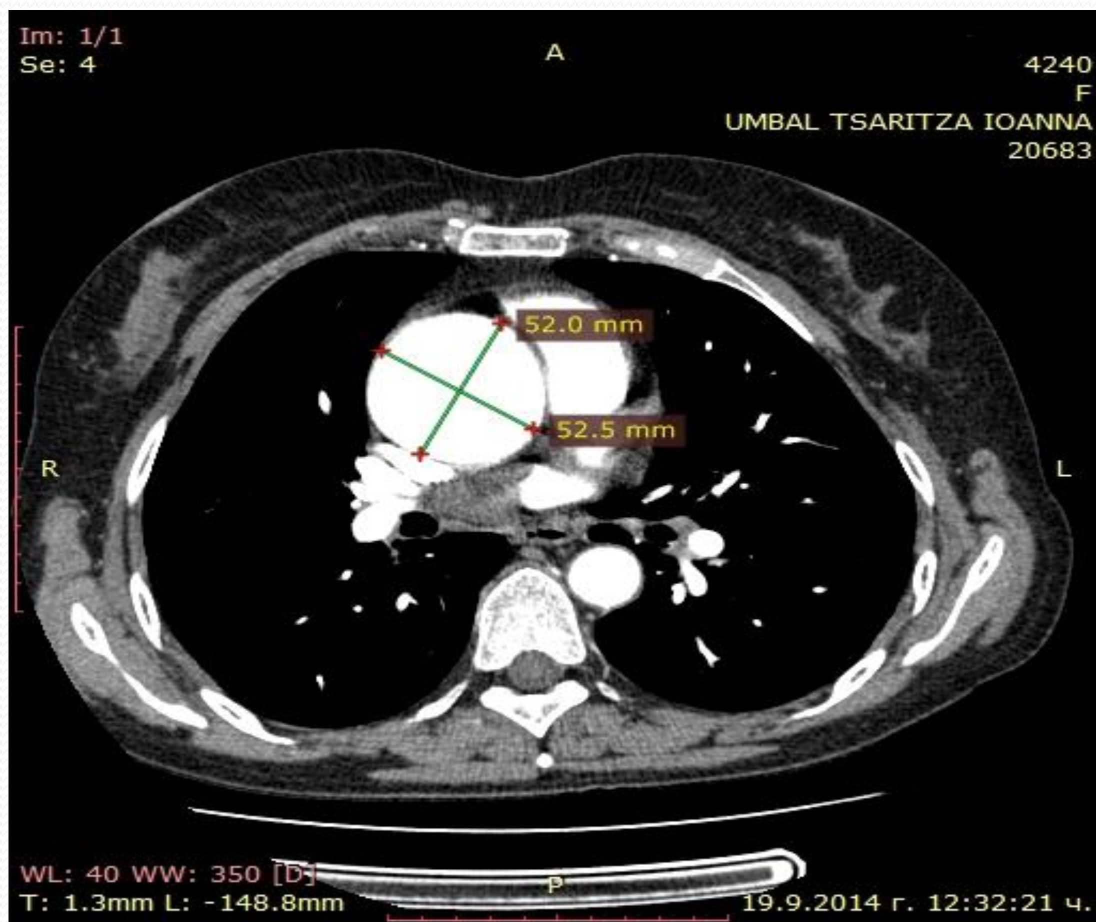
FR 49Hz
16cm
2D
63%
C 50
P Low
HGen
M3
P R
1.7 3.4
0
5
10
15
JPEG
*** bpm

TTE

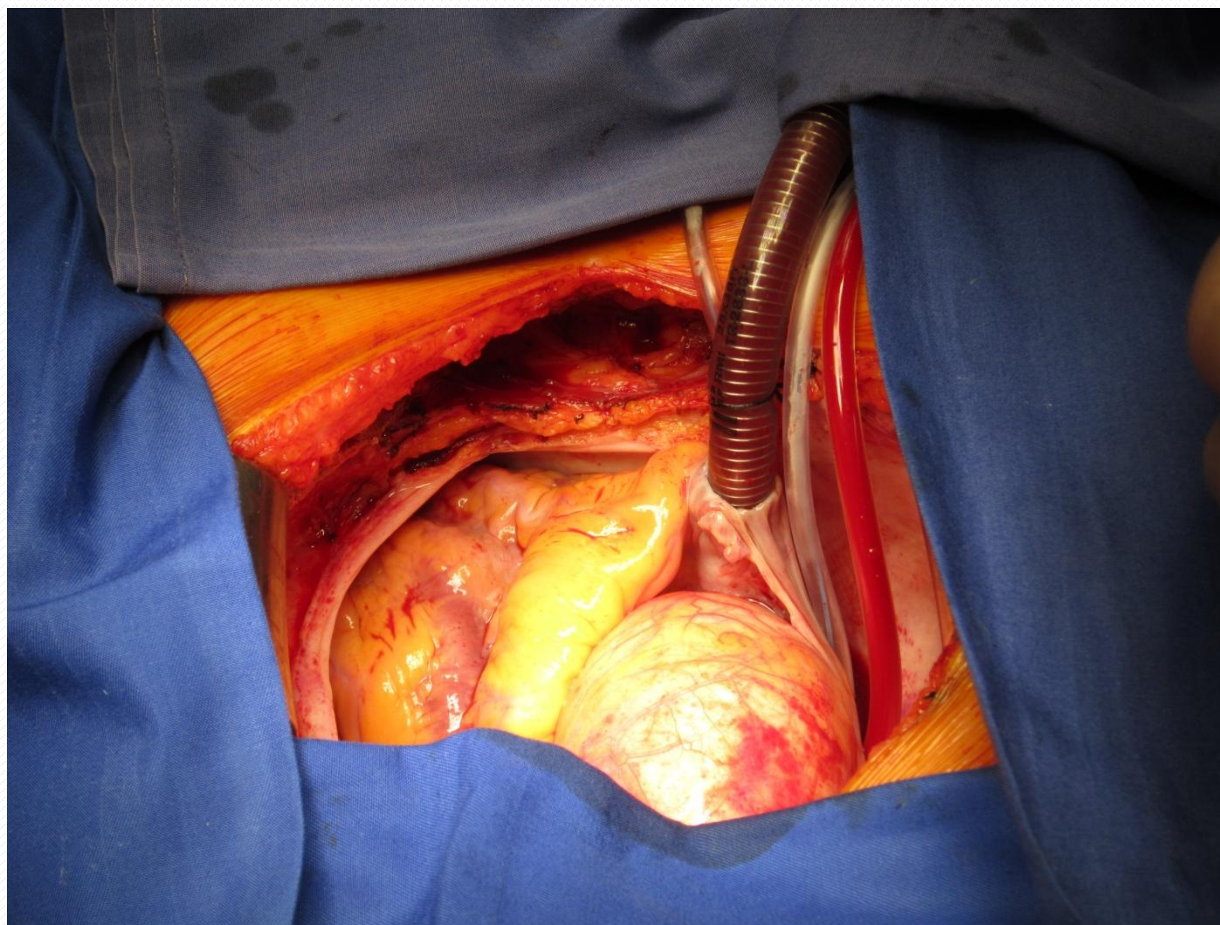


TTE

КТ



Интраоперативна снимка



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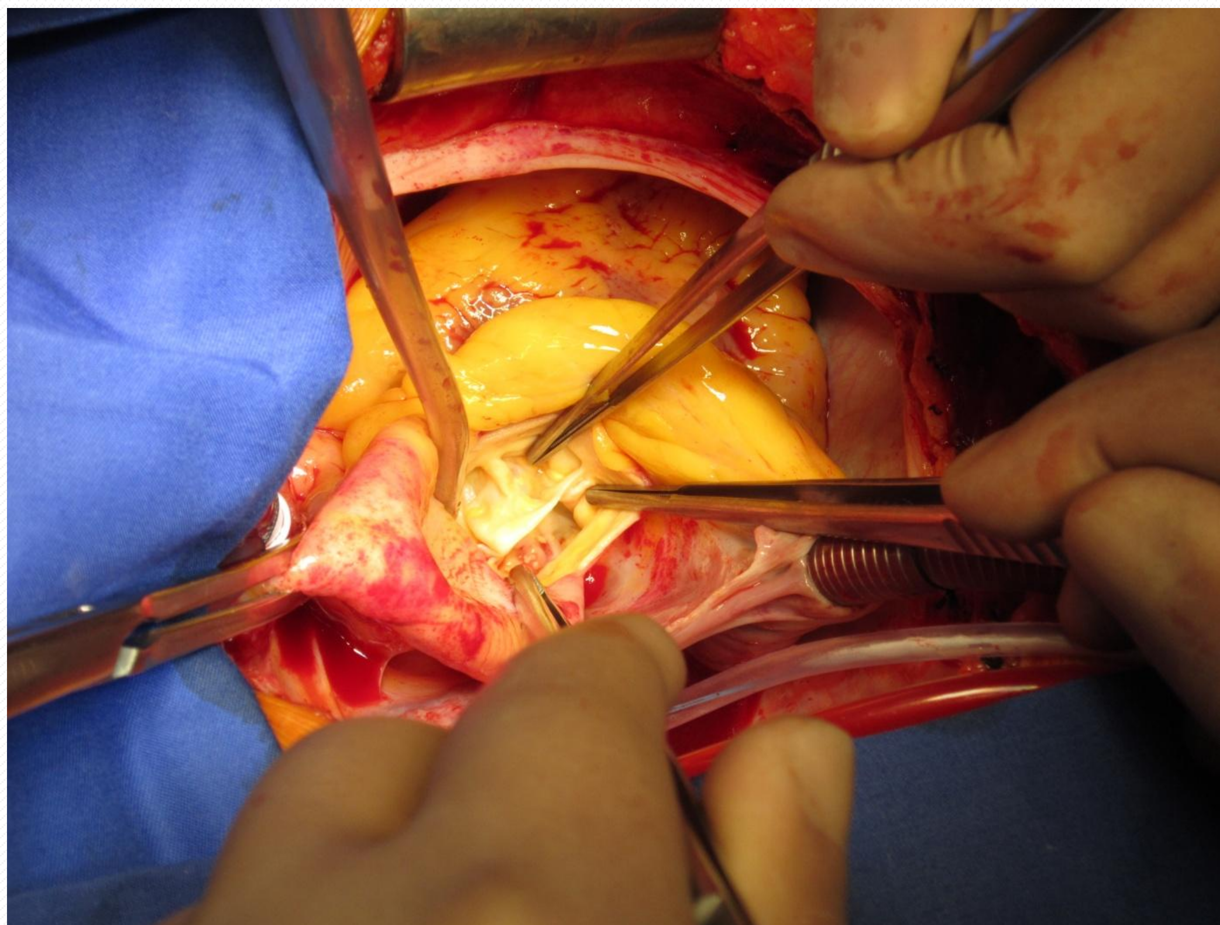

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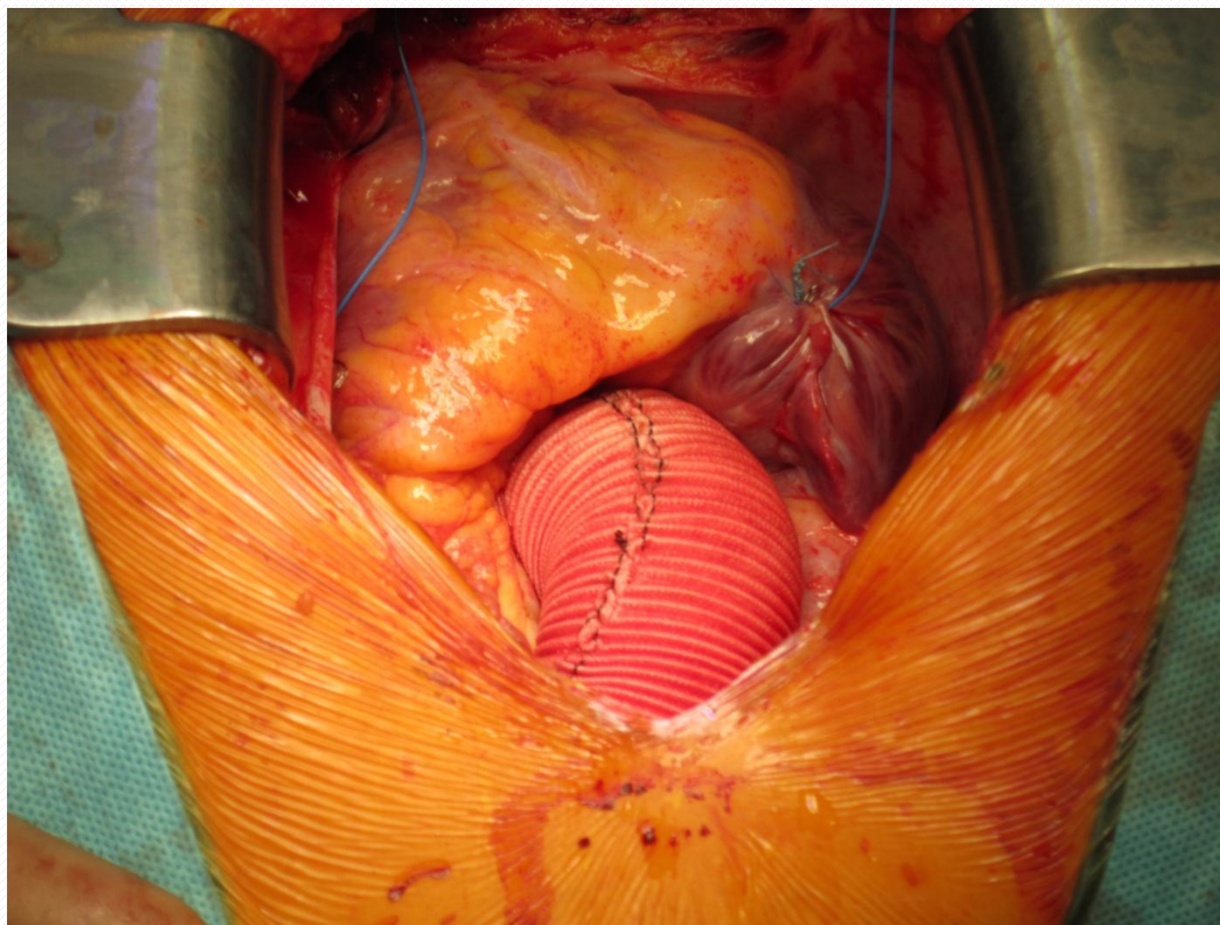

2014 ESC Guidelines on the diagnosis and treatment of aortic diseases

Recommendations	Class	Level
Surgery is indicated in patients who have aortic root aneurysm, with maximal aortic diameter ≥ 50 mm for patients with Marfan syndrome	I	C
Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameters: • ≥ 45 mm for patients with Marfan syndrome with risk factors • ≥ 50 mm for patients with bicuspid valve with risk factors • ≥ 55 mm for other patients with no elastopathy	I	C
Lower thresholds for intervention may be considered according to body surface area in patients of small stature or in the case of rapid progression, aortic valve regurgitation, planned pregnancy, and patient's preference	IIb	C

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Arq Bras Cardiol. 1992 Aug;59(2):127-30.

[Aortic dissection associated with systemic lupus erythematosus].

[Article in Portuguese]

Souza AJ[1], Tarasoutchi F, Cardoso LF, Pommerantzeff PM, Grinberg M.

Author information

Abstract

A 33-year-old female patient, with a 4-year history of hypertension plus a 3-year history of systemic lupus erythematosus, who had been taking high dosages of corticosteroids, has shown repetitive respiratory infections and congestive heart failure for the past 8 months. Angiocardiography confirmed the diagnosis of aortic insufficiency with aneurysmatic dilation of Valsalva's posterior sinus, ascending aorta of normal diameter and normal coronary arteries. Aortic dissection causing aortic insufficiency due to collapse of aortic leaflets was spotted during the surgery and was corrected by a bovine pericardial tube and suspension of aortic valve. The postoperative (PO) period was complicated by left-sided seizures followed by left hemiparesis and respiratory infection. She was discharged on the 25th PO day with mild left hemiparesis and in functional class I (NYHA), using medicines. We emphasize the need to consider the diagnosis of aortic dissection in patients with systemic lupus erythematosus and aortic insufficiency, specially in those who have a history of systemic arterial hypertension and long-term corticosteroid therapy.

Japanese Heart Journal
Japanese Heart Journal Association
ONLINE ISSN: 1348-673X PRINT ISSN: 0021-4868
(As of October 02, 2014) Registered articles: 4,854

Article | Volume/Issue/Page | DOI/JOI
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Japanese Heart Journal
Vol. 43 (2002) No. 5 P 567-571
DOI http://dx.doi.org/10.1536/jhj.43.567 JOI JST.JSTAGE/jhj/43.567
Case Reports

Language: English
Previous Article

Case Reports

Acute Type A Aortic Dissection in a Patient with Systemic Lupus Erythematosus

Shigeaki Aoyagi[1], Hidetoshi Akashi[1], Hiroyuki Otsuka[1], Hideki Sakashita[1], Teiji Okazaki[1], Kei-ichiro Tayama[1]

1) Department of Surgery, Kurume University School of Medicine

Released 2002/10/24

Keywords: Systemic lupus erythematosus, Aortic dissection, Steroid therapy

Full Text PDF [152K]

Abstracts | References(14) | Cited-By(10)

The case of a 27-year-old Japanese woman with type A acute aortic dissection who had been diagnosed with systemic lupus erythematosus (SLE) is presented. The patient also had aortic regurgitation due to non-infective endocarditis and systemic hypertension, and had been maintained on steroid therapy for 15 years. Her twin sister was also diagnosed with SLE. The patient was admitted to emergency due to severe back pain. A chest x-ray showed enlargement of the upper mediastinum. Echocardiography revealed a thickened and deformed aortic valve with aortic regurgitation and dissection of the ascending aorta, but pericardial effusion was not found. Computed tomography demonstrated aortic dissection extending from the ascending aorta to the abdominal aorta. Graft replacement of the ascending aorta and proximal aortic arch was performed under hypothermic circulatory arrest with retrograde cerebral perfusion. The patient recovered uneventfully. Aortic dissection complicated with SLE is extremely rare, and this is only the 15th case reported in the English or Japanese literature.

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SLE

Aortic Dissection Complicated with Hemothorax in an Adolescent Patient with Systemic Lupus Erythematosus: Case Report and Review of Literature

Hsin-Yi Wei, MD, Hung-Tao Chung, MD, Chang-Teng Wu, MD, and Jing-Long Huang, MD

Objectives: To report a young patient with systemic lupus erythematosus (SLE) complicated by aortic dissection. The relevant literature on the association of SLE and aortic dissection is reviewed.
Methods: We describe an adolescent patient with childhood-onset SLE diagnosed aortic dissection with presentation of hemothorax. The literature review was performed by a PubMed search using the keywords systemic lupus erythematosus (SLE), aortic dissection, hemothorax, and carotid intima-media thickness (CIMT).
Results: A 17-year-old male was admitted to the hospital for severe abdominal pain. His past medical history included childhood-onset SLE complicated with lupus nephritis. Acute aortic dissection complicated with hemothorax was diagnosed and he died despite medical therapy. The accelerated CIMT progression of our patient, 0.14 mm in 20 months, might suggest ongoing premature atherosclerotic changes in the aortic wall. On reviewing the English literature, 21 cases of aortic dissection in SLE have been analyzed and discussed. Patients younger than 40 years of age, hypertension, and long-term steroid use are common features of aortic dissection in SLE patients. The possible pathogenesis of aortic dissection in SLE includes atherosclerosis, degeneration, and vasculitis. The widely accepted CIMT measured by ultrasound could be a potential diagnostic tool to access the risk of cardiovascular events.
Conclusions: Aortic dissection is a rare complication of SLE, but could take place in an adolescent patient with childhood-onset disease. It is important to include aortic dissection as a differential diagnosis in SLE patients with unexplained sharp abdominal, chest, or back pain.
© 2011 Elsevier Inc. All rights reserved. Semin Arthritis Rheum 41:12-18

Keywords: systemic lupus erythematosus, childhood onset, aortic dissection, hemothorax, carotid intima-media thickness

Table 1 Reported Cases of Aortic Dissection in Patients with Systemic Lupus Erythematosus

No.	Author	Age/ Sex Patients	Duration of SLE (yr)	Steroid (yr)	Hypertension	Medication/ Surgery	Histological Findings Vasculitis/ Degeneration	Pathogenesis of Aortic Dissection	Outcome
1	Walts et al (25)	34/M	22	22	+	Medication	−/+	Atherosclerosis	Died
2	Pazirandeh et al (22)	38/F	2.5	2.5	+	Medication	−/+	Degeneration	Died
3	Yoshimoto et al (26)	28/F	11	2	+	Medication	−/−	Atherosclerosis	Died
4	Souza et al (27)	33/F	3	3	+	Surgery	NA	NA	Alive
5	Dugo et al (5)	30/F	0.9	0.9	+	Medication	NA	NA (vasculitis?)	Alive
6	Sclair et al (28)	30/F	10	2	+	Medication	NA	NA	Died
7	Guard et al (7)	31/F	9	9	+	Medication	+/−	Vasculitis	Died
8	Ohge et al (29)	55/F	11	11	NA	Surgery	NA	NA	Alive
9	Sueda et al (30)	52/F	26	21	+	Surgery	−/−	Atherosclerosis	Alive
10	Lam et al (31)	66/F	1	1	+	Medication	−/−	Atherosclerosis	Died
11	Khan et al (32)	38/M	23	23	NA	Surgery	NA	NA	Alive
12	Hussain et al (33)	40/M	22	22	+	Surgery	−/+	Degeneration/ undetermined necrosis	Alive
13	Choi et al (34)	28/F	5	5	+	Surgery	−/+	Degeneration	Alive
14	Wang et al (24)	36/M	0.9	0.9	NA	Medication	+/−	Vasculitis/ undetermined necrosis	Died
15	Motomura et al (35)	45/F	10	9	+	Surgery	NA	NA	Alive
16	Kunihara et al (36)	52/F	11	10	+	Surgery	NA	NA	Alive
17	Aoyagi et al (37)	27/F	15	15	+	Surgery	NA	NA	Alive
18	Murata et al (38)	43/M	2	No	+	Surgery	NA	NA	Alive
19	Arul Rajamurugan et al (39)	42/F	0.58	0.58	+	Medication	NA	NA	Died
20	Sato et al (40)	61/F	13	13	No	Medication	−/+	Atherosclerosis	Died
21	Wei et al (present)	17/M	7	7	+	Medication	NA	NA (atherosclerosis?)	Died

NA, not applicable.

Изводи

- Аортна дилатация- изключително рядко при СЛЕ
- Патогенеза- СЛЕ васкулит
- Васкулит- дегенерация и фиброза на медията на аортната стена с образуване на аневризми
- Бързо прогресираща дилатация на аневризмите
- Връзка между аневризма на аортата и лечението с кортикостероиди
- Всички пациенти със СЛЕ- скрининг за дилатация на аортата
- Критерии за интервенция на аортата при СЛЕ ???