CASO CLINICO

SOLACI 2015

PERU

Instituto Nacional Cardiovascular

LUCIO TORRES VILLACORTA

MEDICO RESIDENTE

CONTENIDO

- **CASO CLINICO**
- **EXAMENES AUXILIARES**
- **MARCO TEORICO**
- **DIAGNOSTICO**
- **MANEJO**
- **CONCLUSIONES**

CASO CLINICO

- **FILIACION**

- Nombre : DDA

- **Edad** : 35 años
- **Sexo** : Femenino
- **Natural** : Lima
- **Procedente** : Lima
- **Ocupacion** : Contadora

CASO CLINICO

- **ANTECEDENTES**

  - **FRCV** : Niega.
  - **CV** : CIA (2010)
  - **Otros** : Niega
  - **RAMs** : Niega
  - **Medicación** : Espironolactona 25mg/24h

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- **HISTORIA DE ENFERMEDAD**

  Referida a INCOR para manejo CIA

  Hace 5 años Disnea CF II.

  Hace 1 año Disnea CF III y **ANGINA CCS II**.

  Hace 6 meses ANGINA CCS III.

  Niega cianosis, sincope.

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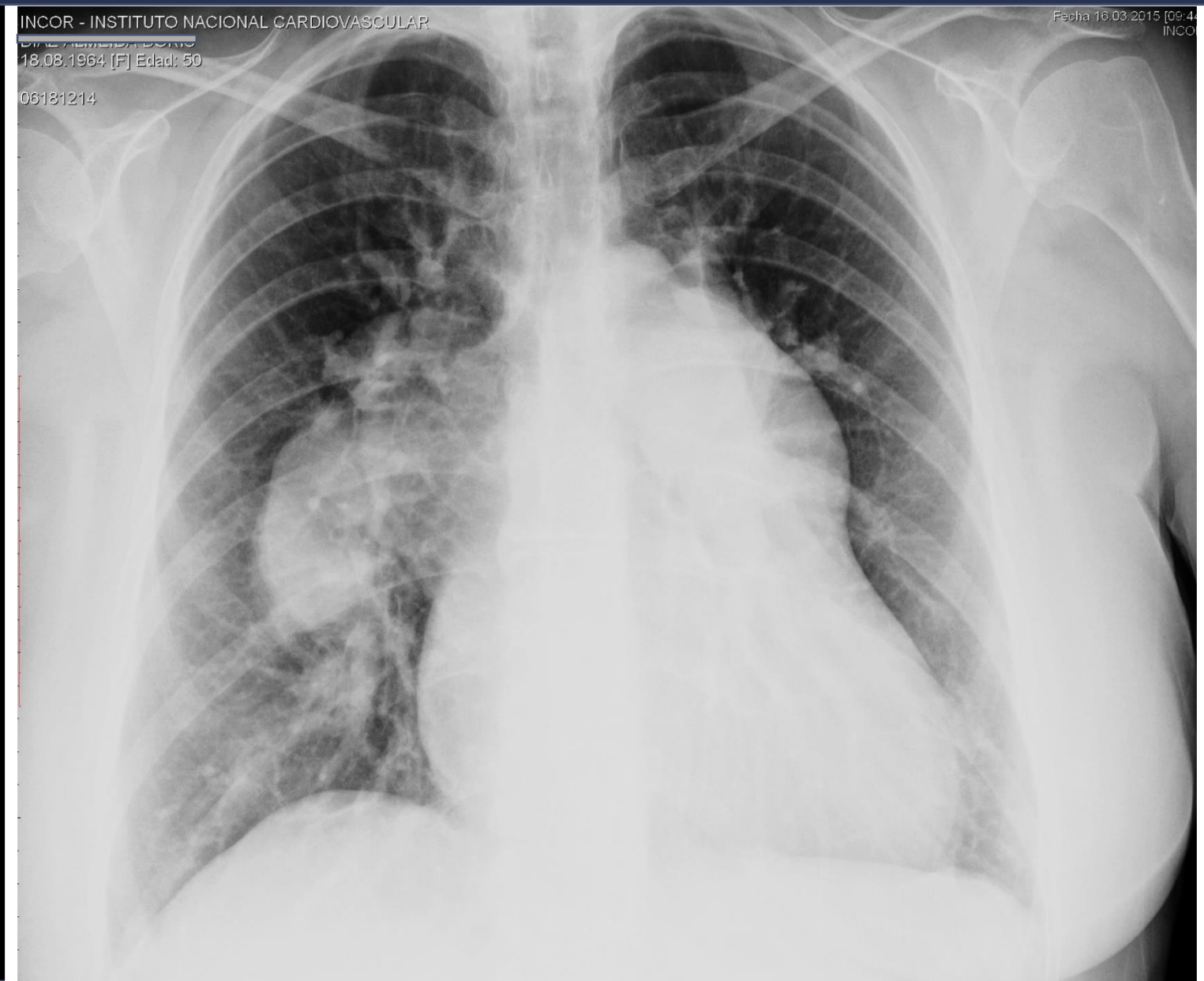
- **EXAMEN FISICO**

PA: 115/65 FC: 75lpm FR: 15 SatO2: 96%

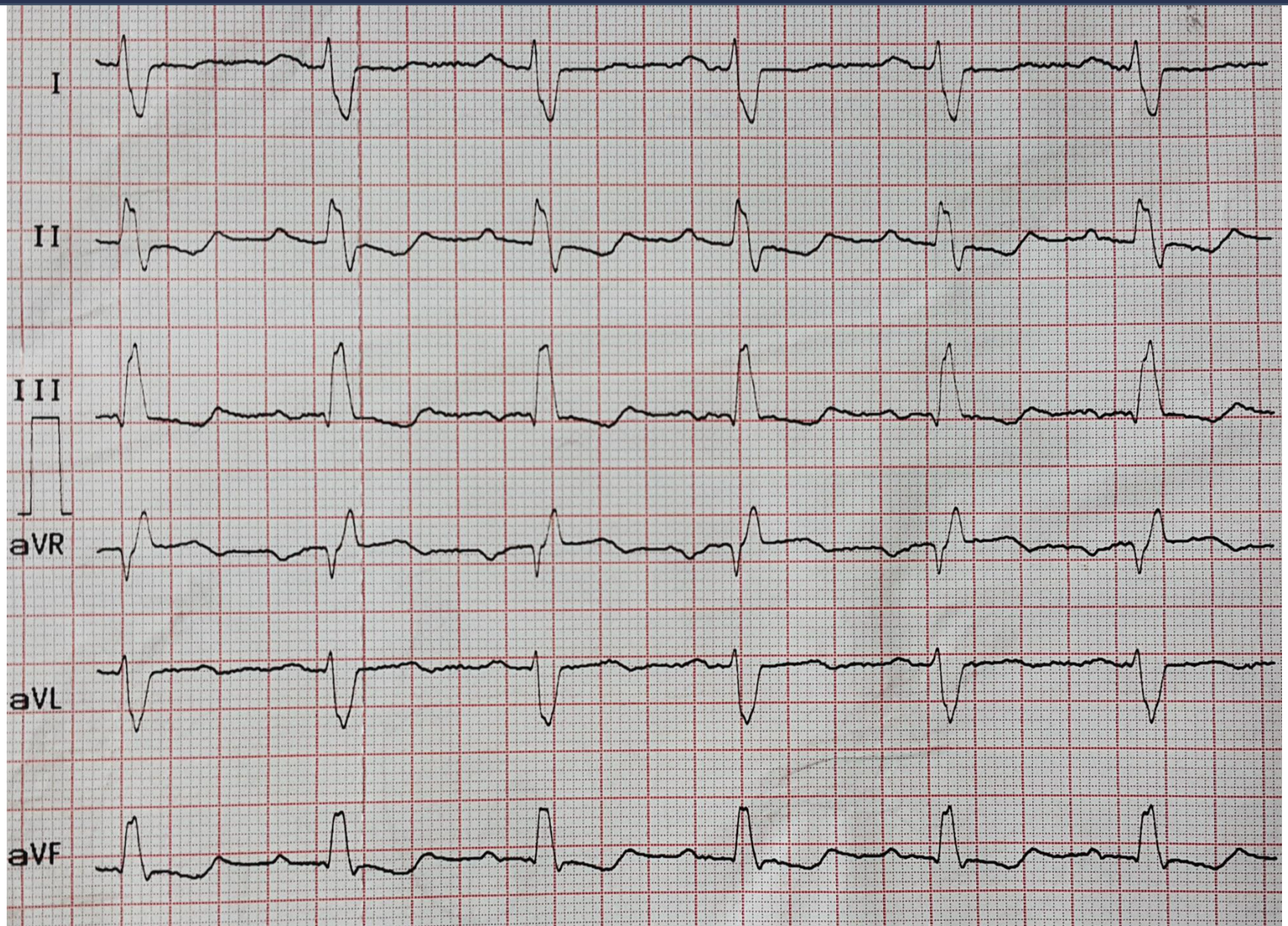
- No contributorio
- CV : RCR, StS FT III/IV , No S3, No IY.
- Pulsos distales conservados.
- No edemas. No cianosis.

EXAMENES AUXILIARES

RADIOGRAFIA TORAX



ELECTROCARDIOGRAMA



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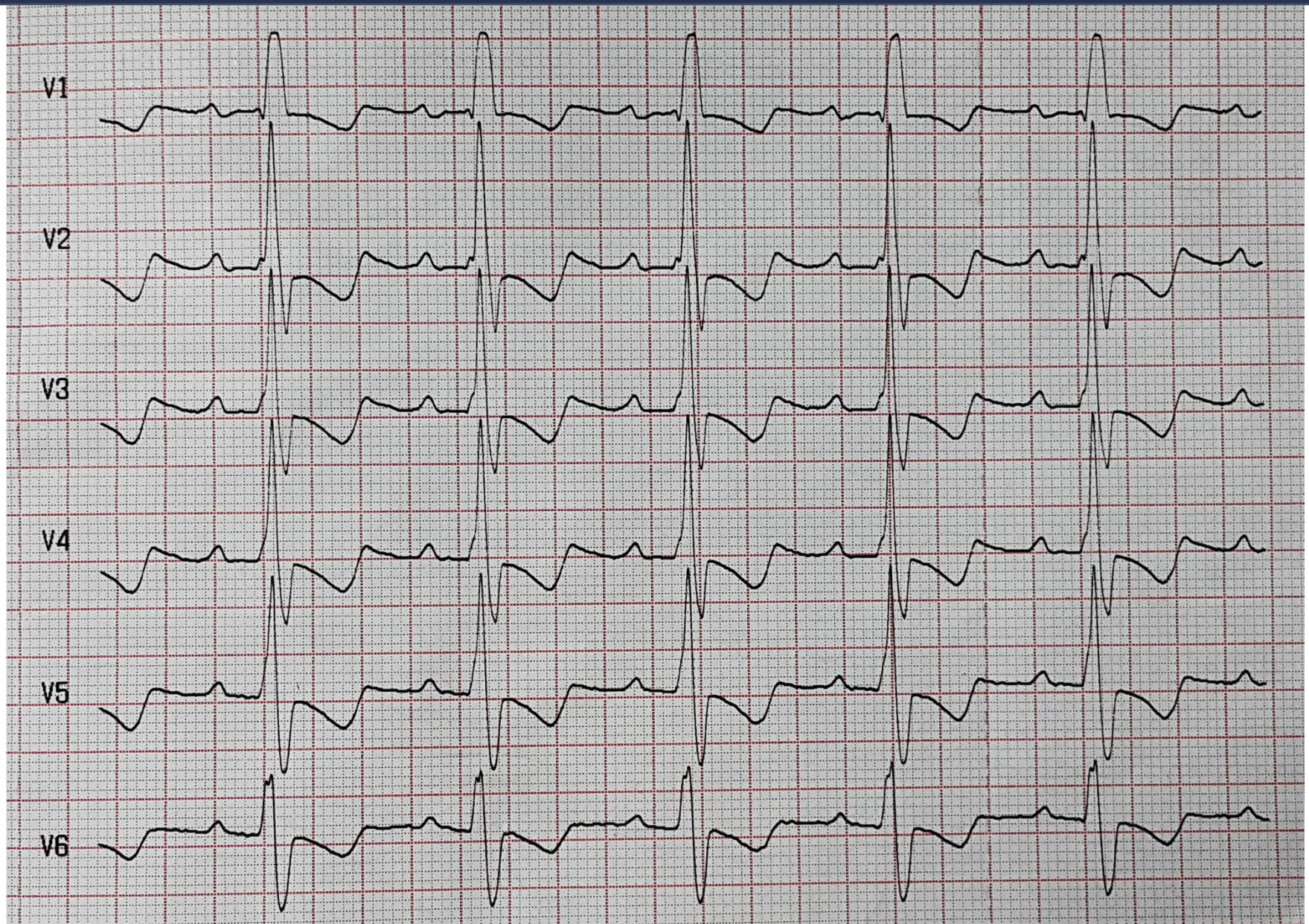
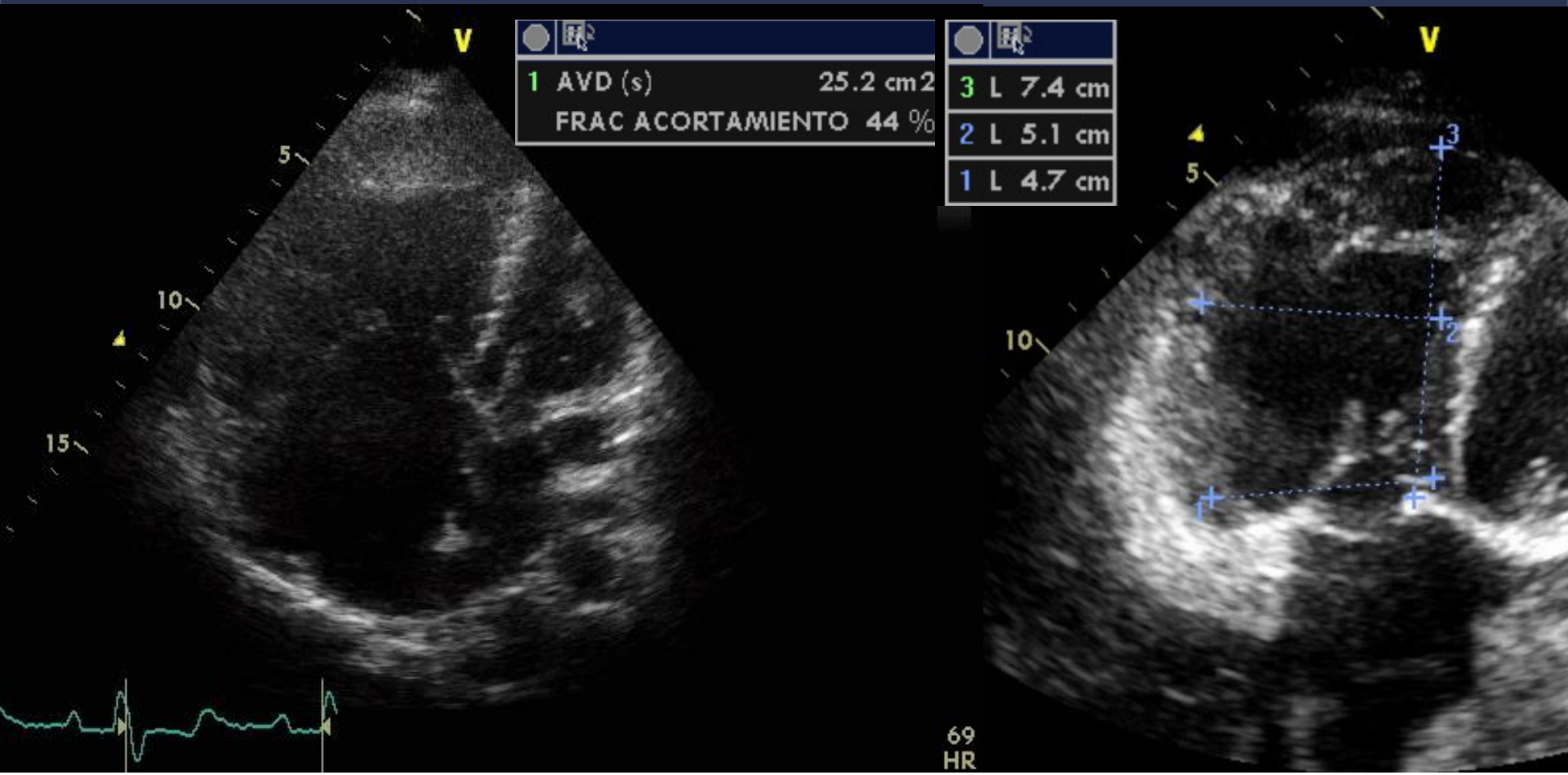
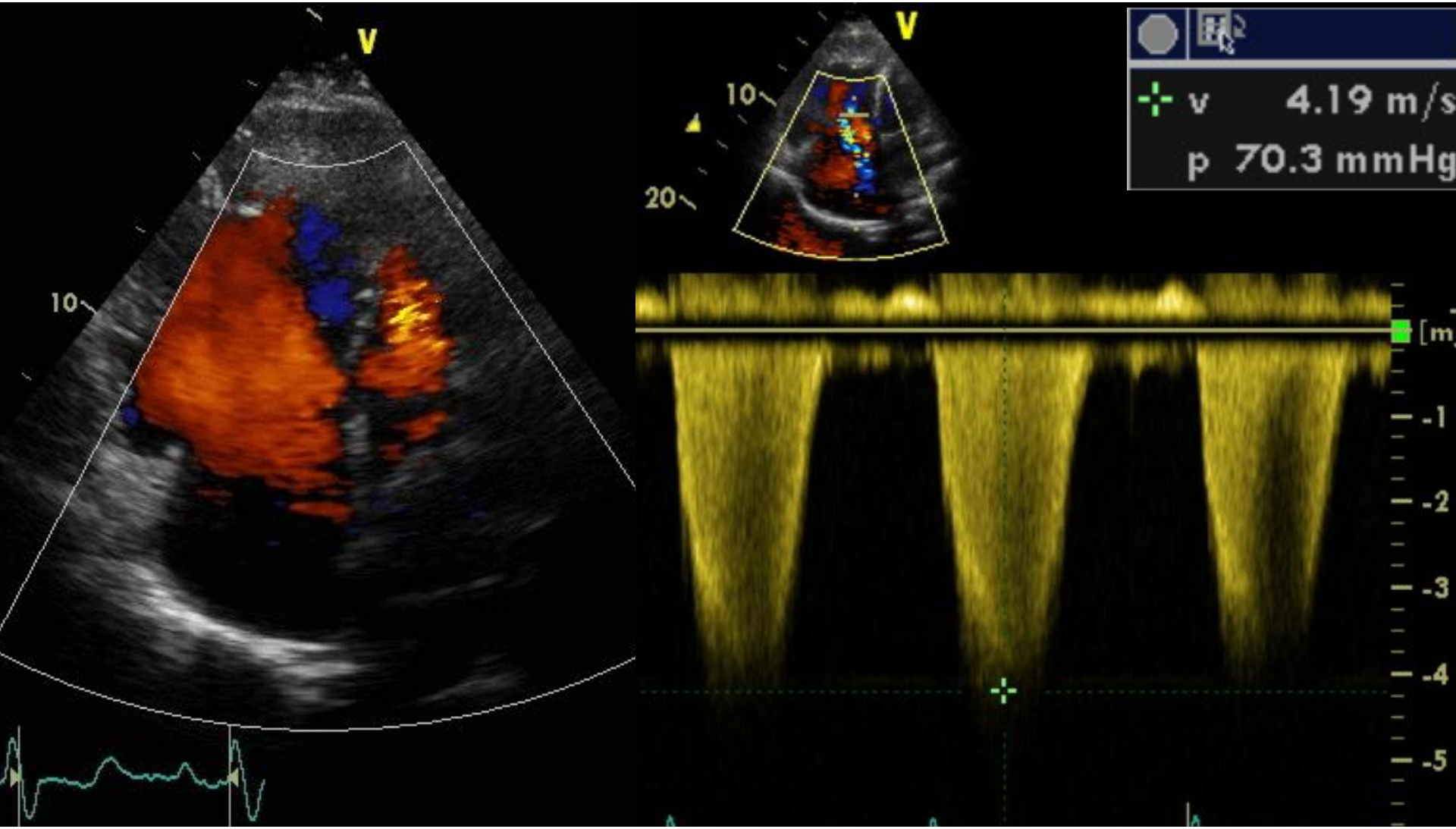


CASO CLINICO

ECOCARDIOGRAFIA TT

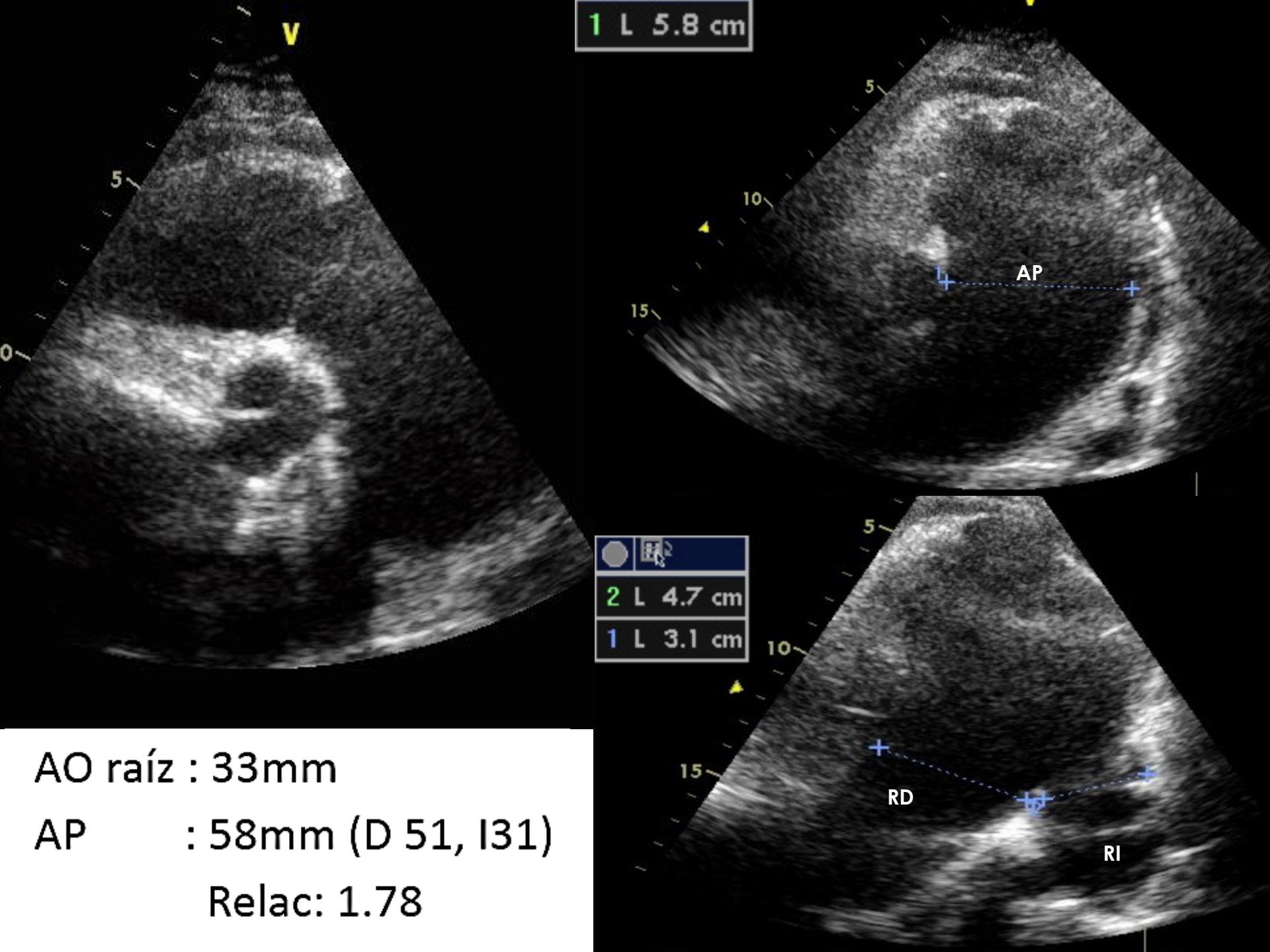


- FEVI : 60%
- AI : 46 (23)
- VID : 47
- AD : 24cm2 (88)
- VD : 47-51-74
- FAC : 44 %



IT severa VCI : 19 (>50%)

PSAP: 75mmHg (Grad VD/AD 70 + PAD 5)



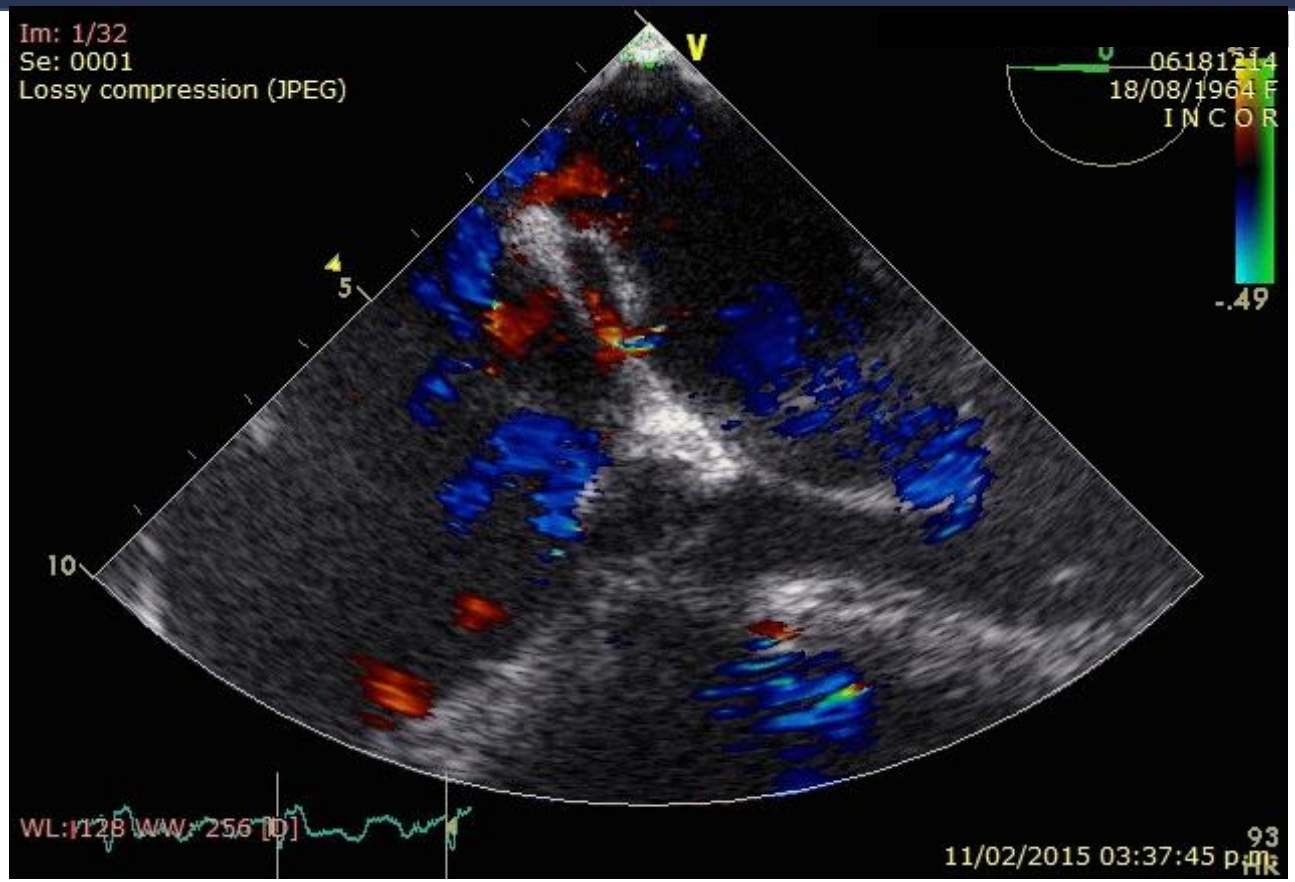
AO raíz : 33mm

AP : 58mm (D 51, I31)

Relac: 1.78

CASO CLINICO

ECOCARDIOGRAFIA TE



- CIA tipo SENO VENOSO
  21mm x 20mm
- Shunt Izquierda a Derecha
- Dilatacion cavidades derechas.
- Insuficiencia tricuspidea severa

Table 8B Echocardiographic signs suggesting pulmonary hypertension used to assess the probability of pulmonary hypertension in addition to tricuspid regurgitation velocity measurement in *Table 8A*

A: The ventricles[a]	B: Pulmonary artery[a]	C: Inferior vena cava and right atrium[a]
Right ventricle/ left ventricle basal diameter ratio >1.0	Right ventricular outflow Doppler acceleration time <105 msec and/or midsystolic notching	Inferior cava diameter >21 mm with decreased inspiratory collapse (<50 % with a sniff or <20 % with quiet inspiration)
Flattening of the interventricular septum (left ventricular eccentricity index >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/sec	Right atrial area (end-systole) >18 cm^2
	PA diameter >25 mm	

AP diam : 59mm
VD/VI basal : 1.03
AD : 24cm2
Grad VD/AD : 7m/s

Table 8A Echocardiographic probability of pulmonary hypertension in symptomatic patients with a suspicion of pulmonary hypertension

Peak tricuspid regurgitation velocity (m/s)	Presence of other echo 'PH signs'[a]	Echocardiographic probability of pulmonary hypertension
≤2.8 or not measurable	No	Low
≤2.8 or not measurable	Yes	Intermediate
2.9–3.4	No	
2.9–3.4	Yes	High
>3.4	Not required	

ALTA PROBABILIDAD HTP

CASO CLINICO

Mujer **JOVEN**

Cardiopatía **CONGENITA** ACIANOTICA

ALTA PROBABILIDAD **HTP**

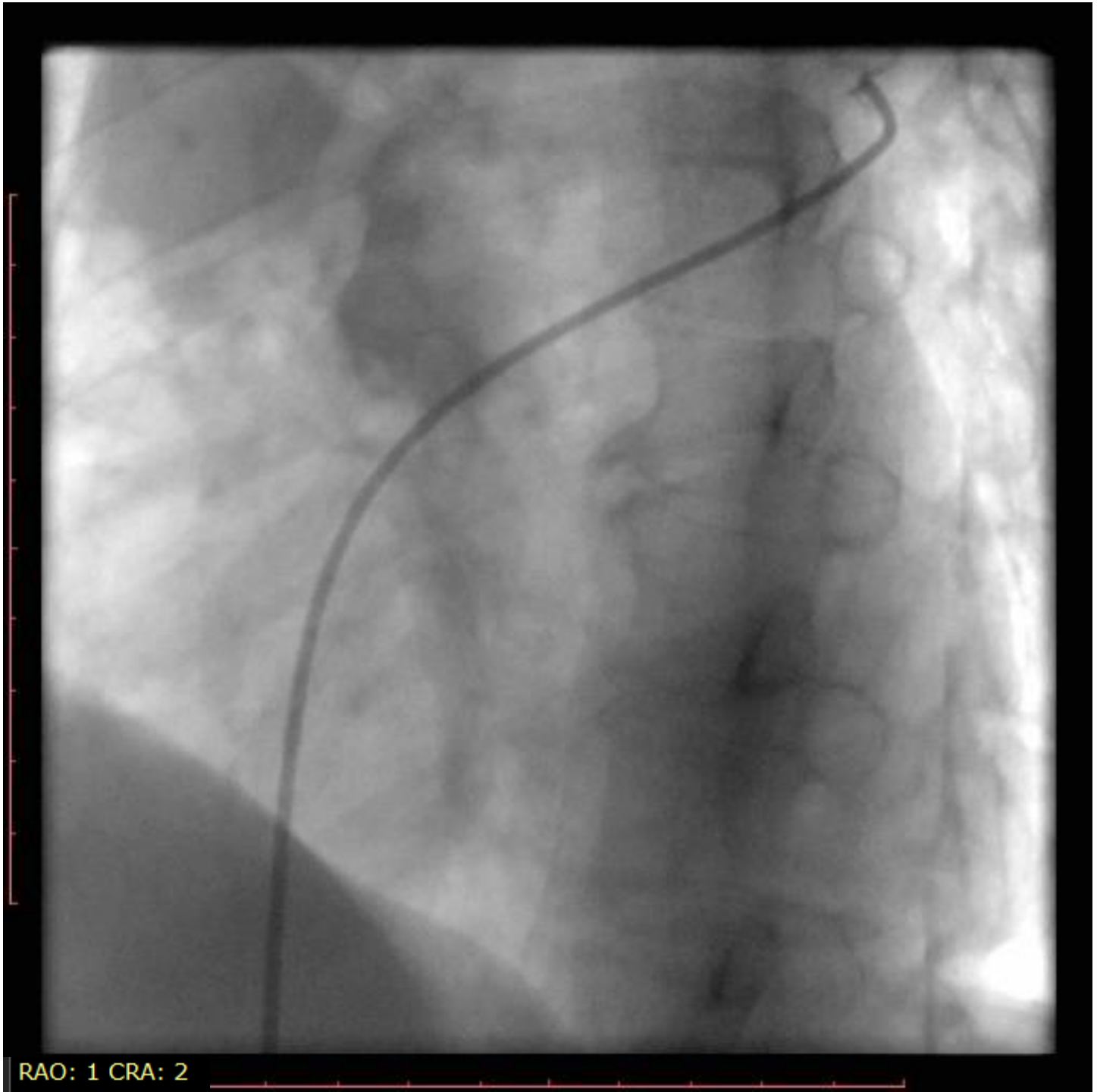
2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

Table 10 Recommendations for right heart catheterization in pulmonary hypertension

RHC is recommended to confirm the diagnosis of pulmonary arterial hypertension (group 1) and to support treatment decisions	I	C
RHC is recommended in patients with congenital cardiac shunts to support decisions on correction (Table 24)	I	C

CATETERISMO CARDIACO

DERECHO


RAO: 1 CRA: 2

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CATETERISMO DERECHO

Posición	PAS/PAD	Media mmHg	Contenido vol%	SatO2 %
VCS			13.75	78.6
VCI			13.64	79.2
AD		**19**	16.86	**89.9**
VD	100/18			
AP	100/27	**51**	15.59	83.1
CAP		**15**		
VI	151/18		17.82	95
Ao	151/93	117	18.01	96

CASO CLINICO

CATETERISMO DERECHO

CALCULOS HEMODINAMICOS			
Dif. AV SISTEMICA	4.26	RVS	23.19uW
Dif. AV PULMONAR	2.42	**RVP**	**4.6 uW**
GCS	**5.01**L/min	RVP/RVS	4.9
GCP	7.89L/min	Shunt I → D	3.52L/min
Relación flujo pulmonar/sistémico	**1.67**	IC	2.89

Table 4 Comprehensive clinical classification of pulmonary hypertension (updated from Simonneau et *al.*[5])

1. Pulmonary arterial hypertension

- 1.1 Idiopathic
- 1.2 Heritable
  - 1.2.1 BMPR2 mutation
  - 1.2.2 Other mutations
- 1.3 Drugs and toxins induced
- 1.4 Associated with:
  - 1.4.1 Connective tissue disease
  - 1.4.2 Human immunodeficiency virus (HIV) infection
  - 1.4.3 Portal hypertension
  - 1.4.4 Congenital heart disease
  - 1.4.5 Schistosomiasis

2. Pulmonary hypertension due to left heart disease

- 2.1 Left ventricular systolic dysfunction
- 2.2 Left ventricular diastolic dysfunction
- 2.3 Valvular disease
- 2.4 Congenital / acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
- 2.5 Congenital /acquired pulmonary veins stenosis

3. Pulmonary hypertension due to lung diseases and/or hypoxia

- 3.1 Chronic obstructive pulmonary disease
- 3.2 Interstitial lung disease
- 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
- 3.4 Sleep-disordered breathing
- 3.5 Alveolar hypoventilation disorders
- 3.6 Chronic exposure to high altitude
- 3.7 Developmental lung diseases (Web Table III)

4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions

- 4.1 Chronic thromboembolic pulmonary hypertension
- 4.2 Other pulmonary artery obstructions
  - 4.2.1 Angiosarcoma
  - 4.2.2 Other intravascular tumors
  - 4.2.3 Arteritis
  - 4.2.4 Congenital pulmonary arteries stenoses
  - 4.2.5 Parasites (hydatidosis)

5. Pulmonary hypertension with unclear and/or multifactorial mechanisms

- 5.1 Haematological disorders: chronic haemolytic anaemia, myeloproliferative disorders, splenectomy
- 5.2 Systemic disorders, sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis
- 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
- 5.4 Others: pulmonary tumoral thrombotic microangiopathy,

PAPm > 25 MMhG
PCP < 18
RVP > 3uw
C.C.C Shunt

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

2. Pulmonary arterial hypertension (PAH, group 1) is a clinical condition characterized by the presence of pre-capillary PH (Table 3) and pulmonary vascular resistance >3 Wood units, in the absence of other causes of pre-capillary PH such as PH due to lung diseases, chronic thromboembolic PH, or other rare diseases (Table 4). PAH includes different forms that share a similar clinical picture and virtually identical pathological changes of the lung microcirculation (Table 4).

HIPERTENSION PULMONAR

SINTOMAS

Survival in Patients With Idiopathic, Familial, and Anorexigen-Associated Pulmonary Arterial Hypertension in the Modern Management Era

Marc Humbert, MD, PhD; Olivier Sitbon, MD, PhD; Ari Chaouat, MD, PhD; Michèle Bertocchi, MD;

Registry on Primary Pulmonary hypertension of National Institutes of Health (NIH) 1981-85.

D´Alonzo, R.J.Barst. S.M. Ayres, et al.

Predicting survival in pulmonary arterial hypertension.

Registry to evaluate early and Long Term Pulmonary Arterial Hypertension Disease Management (REVEAL)

Circulation, Volume 122, 2010, pp. 164-172

ANGINA ?

DISNEA	**80 %**
FATIGA	19 %
SINCOPE	13 %

ANGINA
HIPERTENSION PULMONAR

- Síntoma descrito

- ETIOLOGIA NO DEFINIDA
  - Isquemia / Disfunción VD- VI
  - Anatomía coronaria

- Importancia :

Disfunción VI	**20%**
MUERTE SUBITA	**26%**

BRAUNWALD´S HEART DISEASE 10th – 2015
Pulmonary Hypertension 74; 1975 - 98

Patient No.	Sex	Age (years)	New York Heart Association Functional Class	Diagnosis	Mean Pulmonary Artery Pressure (mm Hg)	Pulmonary Trunk Diameter (mm)	Pulmonary Trunk to Aortic Diameter Ratio	Chest Pain	Left Coronary Artery Compression
1	F	29	III	Atrial septal defect	50	40	2.00	Yes	Yes
2	F	26	II	Atrial septal defect	64	51	1.70	Yes	Yes
3	F	54	III	Atrioventricular septal defect	38	68	2.62	Yes	Yes
4	F	43	III	Atrial septal defect	47	68	2.83	Yes	No
5	F	53	III	Idiopathic pulmonary arterial hypertension	56	37	1.28	Yes	No
6	F	38	II	Atrial septal defect					
7	F	24	IV	Idiopathic pulmonary arterial hypertension					
8	F	37	III	Idiopathic pulmonary arterial hypertension					
9	F	23	III	Atrial septal defect					
10	M	46	II	Idiopathic pulmonary arterial hypertension					
11	M	15	IV	Atrial septal defect					
12	F	18	II	Atrial septal defect					
13	F	38	III	Atrial septal defect					
14	F	39	III	Atrial septal defect					
15	F	47	II	Atrial septal defect					
16	M	62	II	Atrial septal defect					
17	F	55	III	Atrial septal defect					
18	F	60	II	Atrial septal defect					
19	F	81	II	Atrial septal defect					
20	F	66	III	Atrial septal defect					
21	F	53	II	Atrial septal defect					
22	M	86	IV	Atrial septal defect					
23	M	50	II	Atrial septal defect					
24	F	42	III	Atrial septal defect					
25	F	72	II	Atrial septal defect					
26	F	73	III	Atrial septal defect					
27	F	47	II	Atrial septal defect					
28	M	66	III	Atrial septal defect					
29	F	57	II	Atrial septal defect					
30	M	80	III	Atrial septal defect					
31	F	44	III	Atrial septal defect					
32	M	43	III	Aortic coarctation + persistent ductus arteriosus					
33	F	58	IV	Atrial septal defect					
34	F	53	III	Idiopathic pulmonary arterial hypertension					
35	M	47	III	Atrial septal defect	50	64	2.46	Yes	Yes
36	M	29	III	Atrial + ventricular septal defect	77	50	2.17	Yes	No

Am J Med. 2004;116:369–374.

Hospital Sao Paulo
2002

TODOS **HTP** (36)

- CONGENITA
  - **CIA** (70 %)

ANGINA 26 (75%)

75% **MUJERES**

NYHA III (55%)

Coronary Artery Compression in Pulmonary Hypertension/Mesquita et al

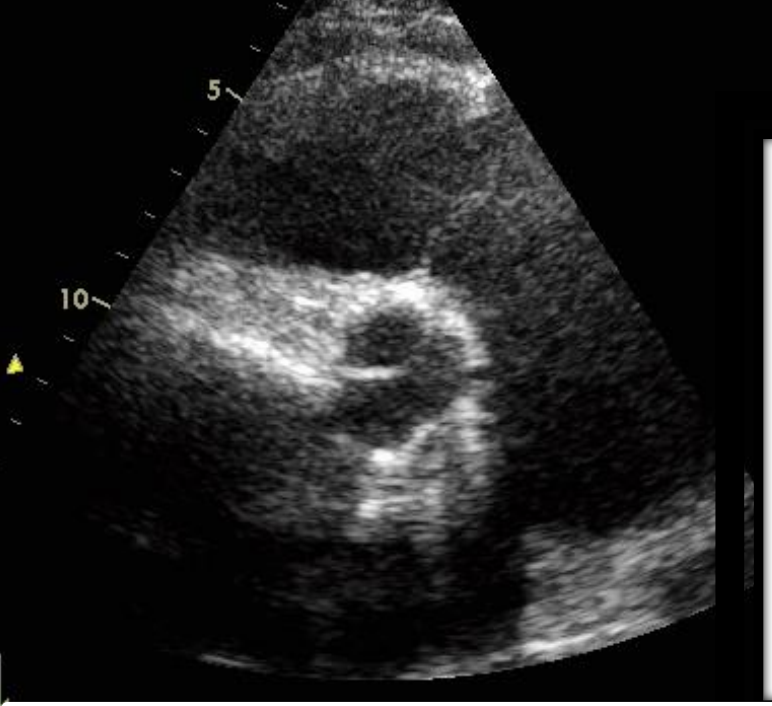
Table 2. Characteristics of Patients with or without Angina

Characteristic	Angina		*P* Value
	Absent	Present	
Men/women	3/7	7/19	0.99
Etiology of pulmonary hypertension (idiopathic/congenital heart disease)	2/8	7/19	0.99
Age (years)	61 ± 19	44 ± 15	0.005*
Mean pulmonary artery pressure (mm Hg)	41 ± 10	51 ± 14	0.06*
Pulmonary trunk diameter (mm)	34 ± 5	43 ± 13	0.01*
Pulmonary trunk to aortic diameter ratio	1.29 ± 0.22	1.66 ± 0.54	0.07

* Significant differences in multivariate analysis ($P < 0.005$).

CORONARIOGRAFIA DIAGNOSTICA

Coronary Artery Compression in Pulmonary Hypertension/Mesquita et al



AO raíz : 33mm

AP : 59mm (D 51, I31)

Relac: 1.78

VCI : 19 (>50%)

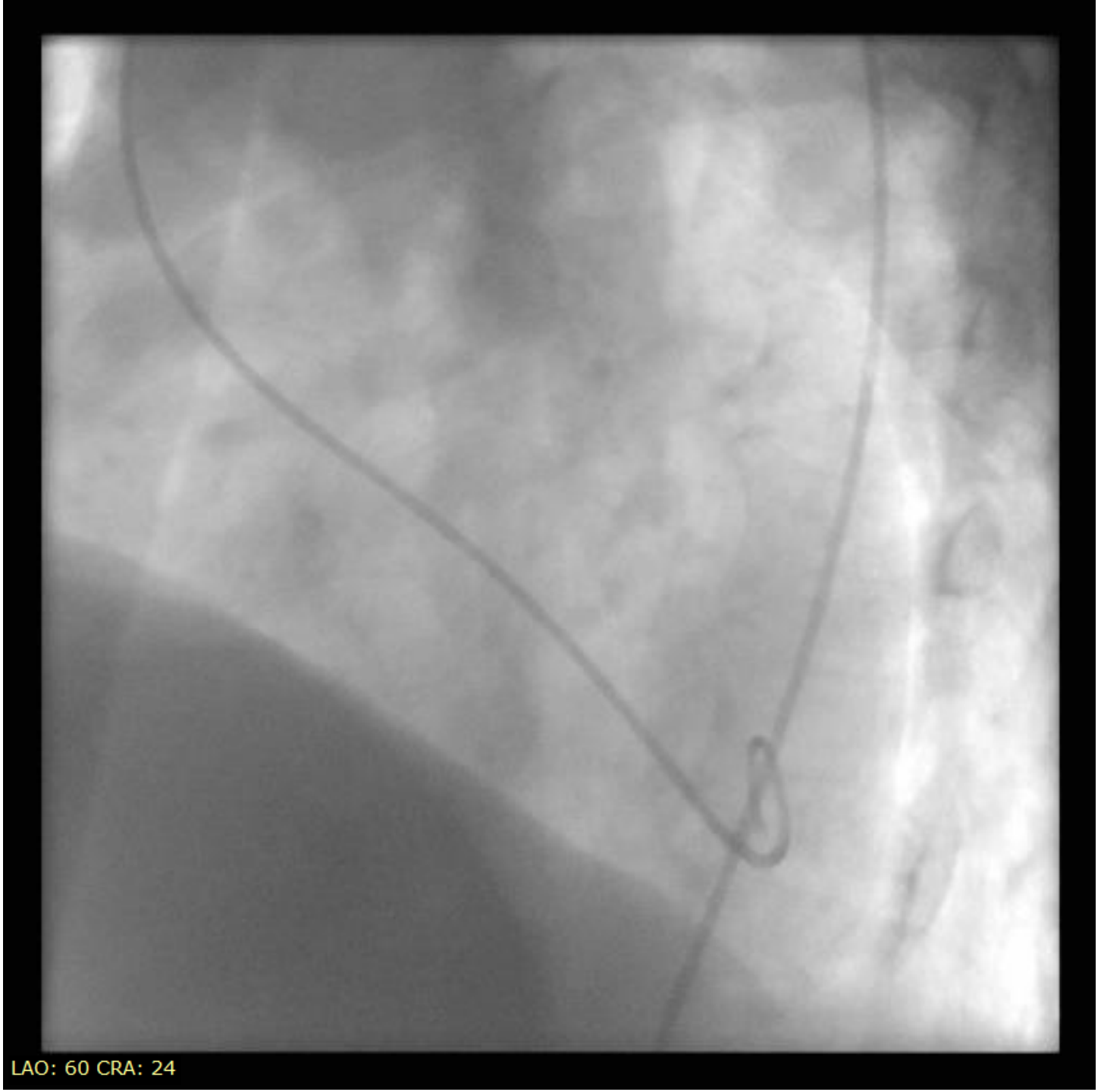
ALTA PROBABILIDAD
ESTENOSIS TCI
POR COMPRESION EXTRINSECA

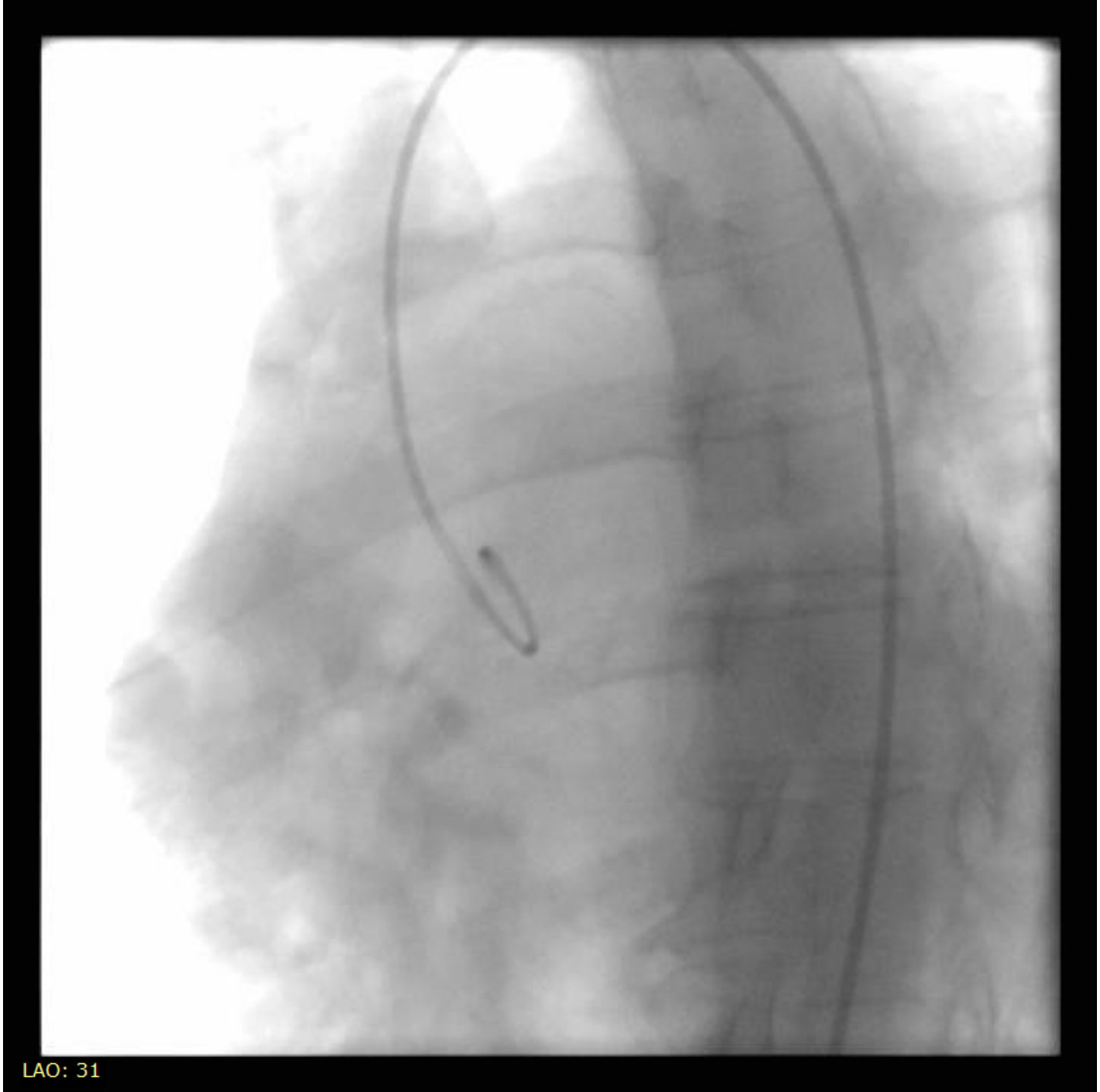
Diametro AP >= 40 (normal 25-30)

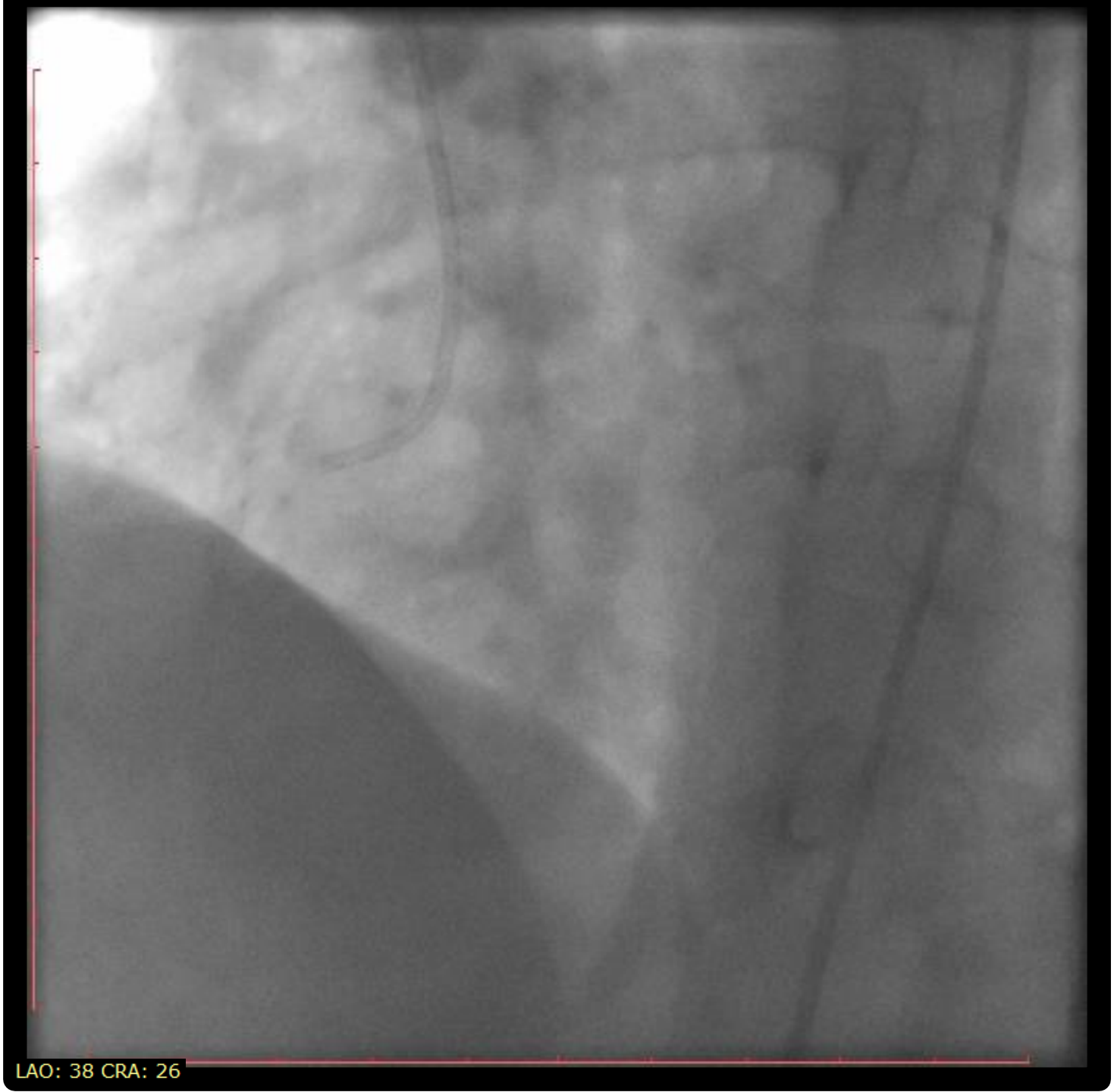
Rel AP / Ao >= 1.21 (normal 1)

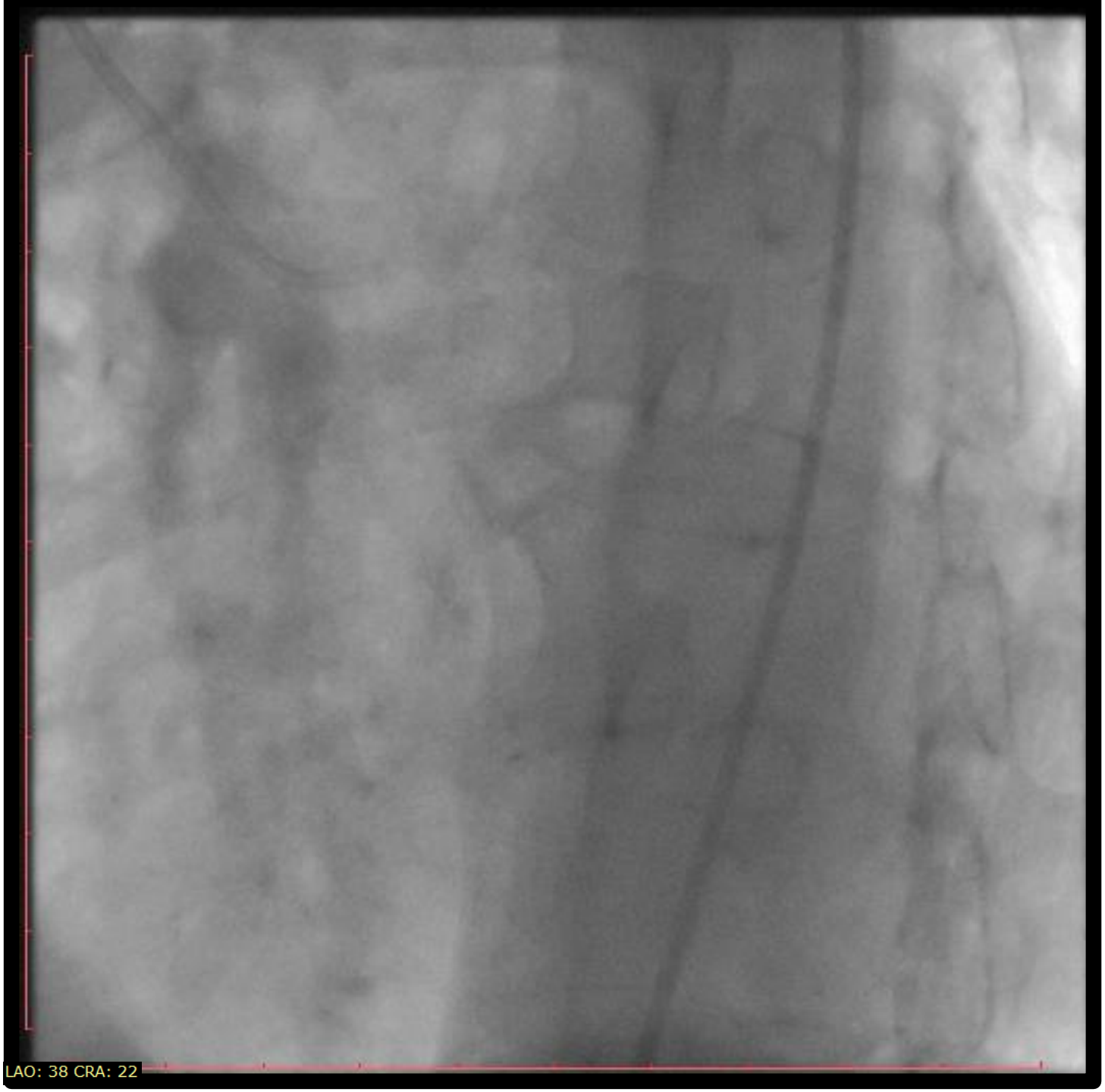
Coronary Artery Compression in Pulmonary Hypertension/Mesquita et al

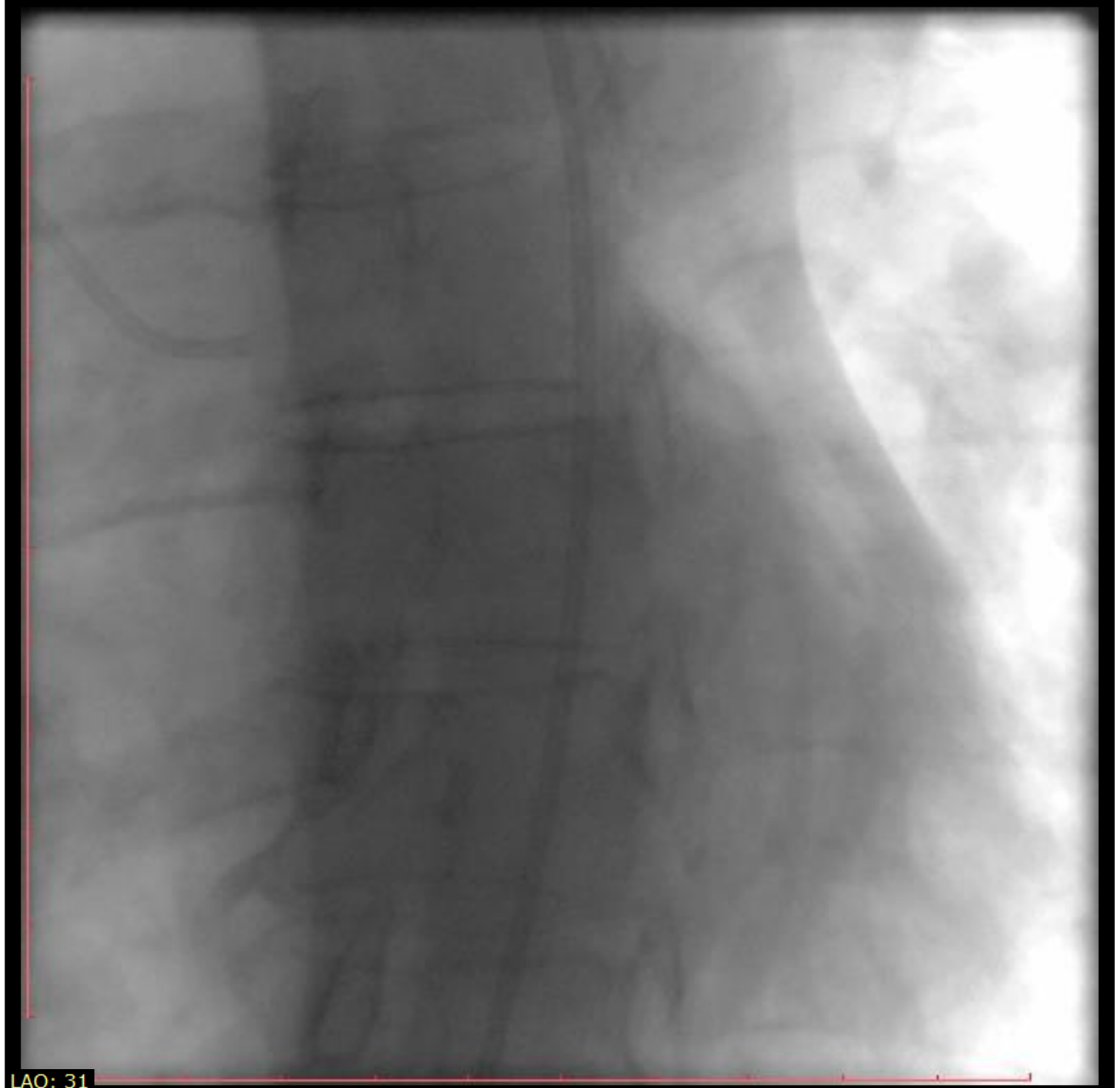
CATETERISMO CARDIACO IZQUIERDO


LAO: 60 CRA: 24


LAO: 31


LAO: 38 CRA: 26


LAO: 38 CRA: 22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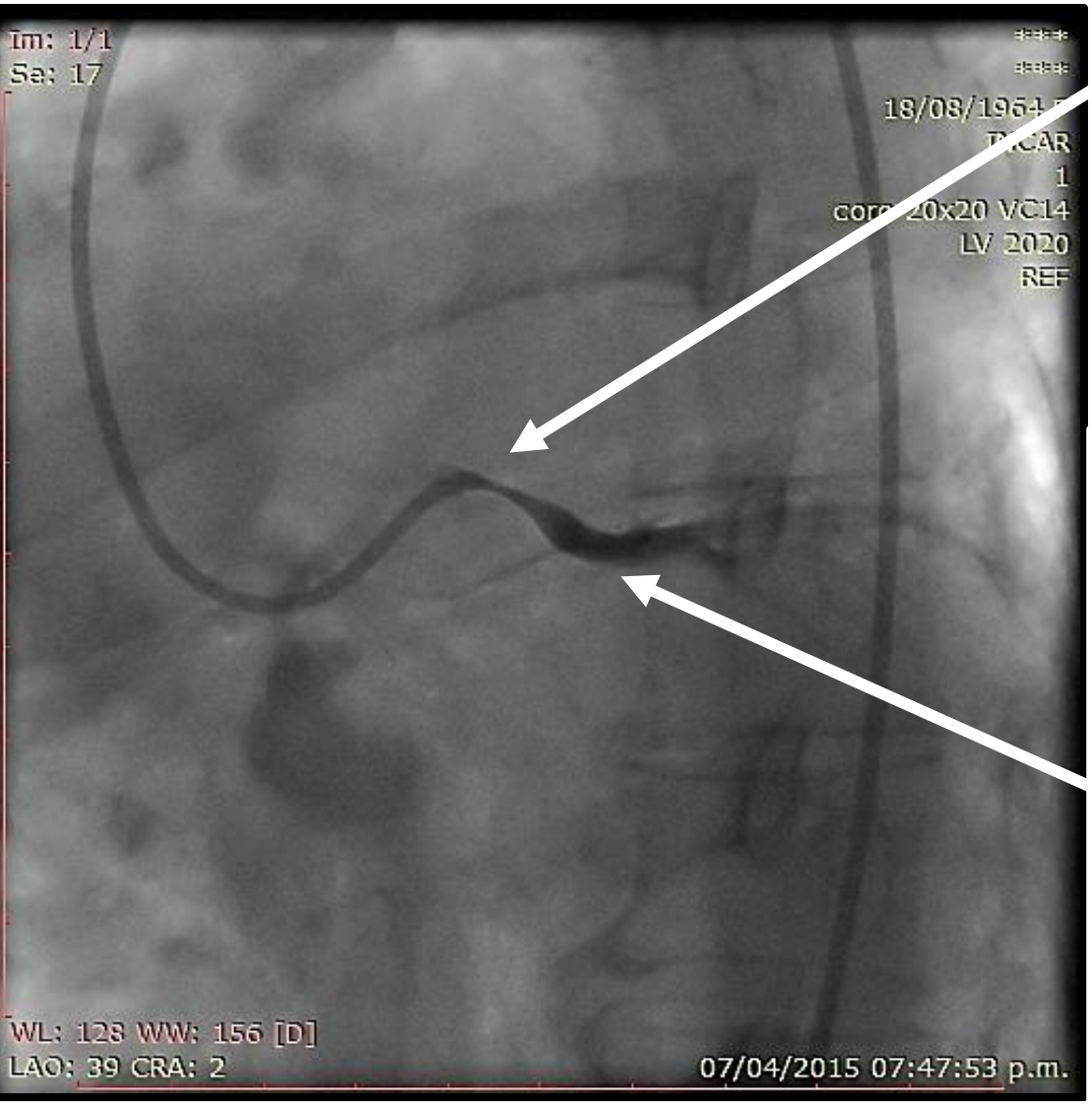
IVUS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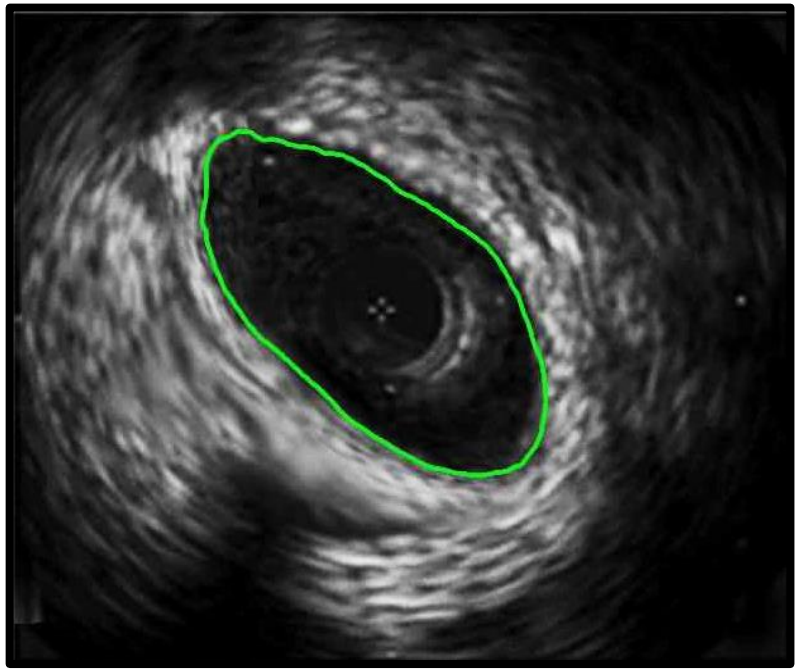
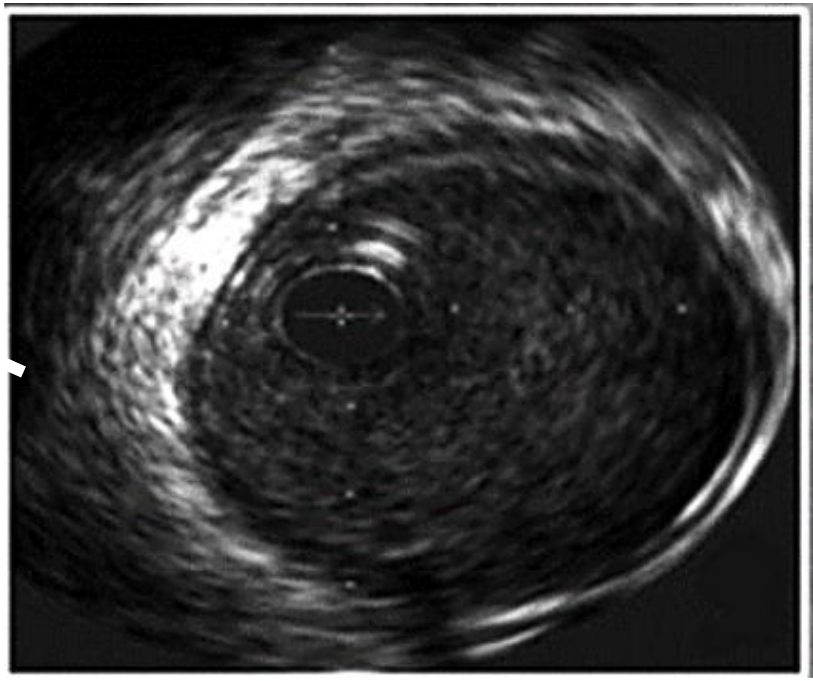




ESTENOSIS TCI
por COMPRESION EXTRINSECA
de ARTERIA PULMONAR?

HIPERTENSION PULMONAR
COMPRESION DE TCI

- DESCRITO DESDE **1956** por Corday et al.
  "Etiologia de IC en pacientes con HTP"
- Primera publicación: **1987**
- ESTADISTICAS
  - MAYOR **ASOCIACION** :

HAP 1 **C.CONGENITA** **CIA** 8 - 44%

- Causa **REVERSIBLE** de angina en HTP.

Paç et al. / International Journal of Cardiology 118 (2007) e41–e43

Coronary arteriographic findings in the patients with atrial septal defect and pulmonary hypertension (ASD +PH) – compression of left main coronary artery by pulmonary trunk.

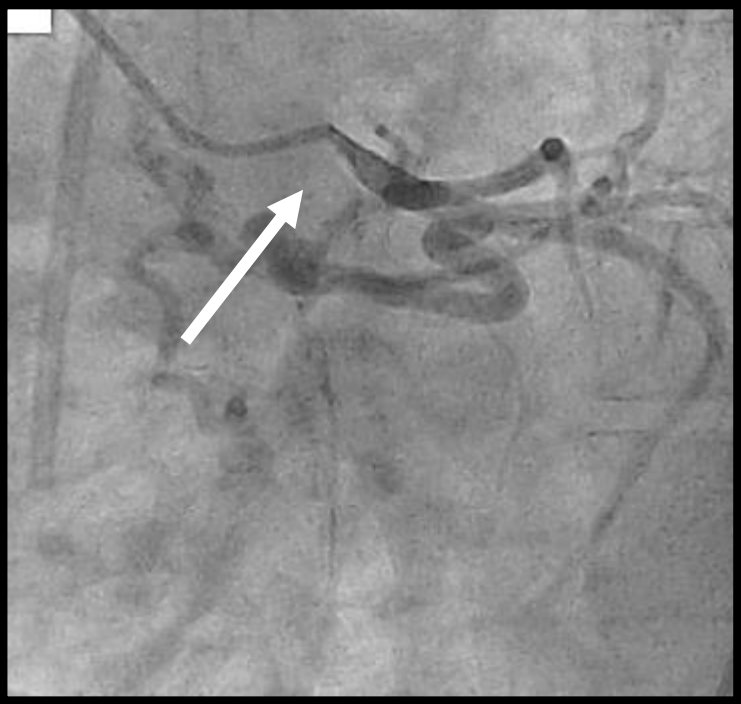
- **CATETERISMO** de 38 pacientes con CIA (15 – 62ª)
- **HTP** : 16 pacientes (**42%**)

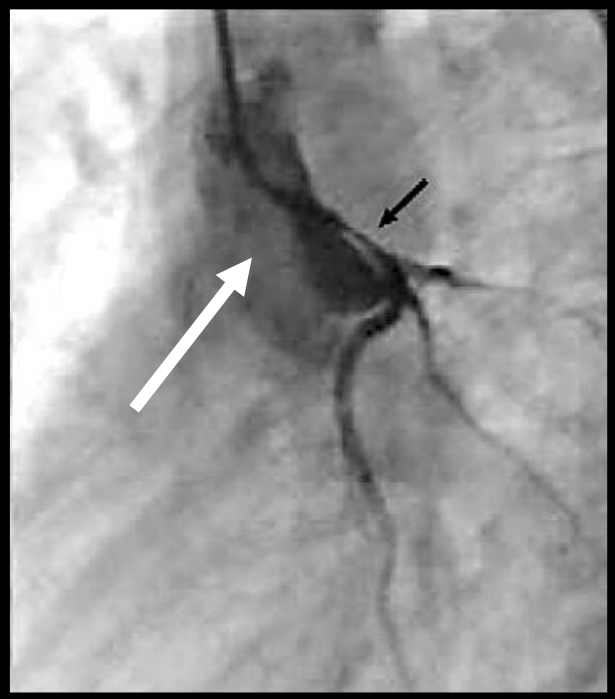
18% : **COMPRESION EXTRINSECA** por AP

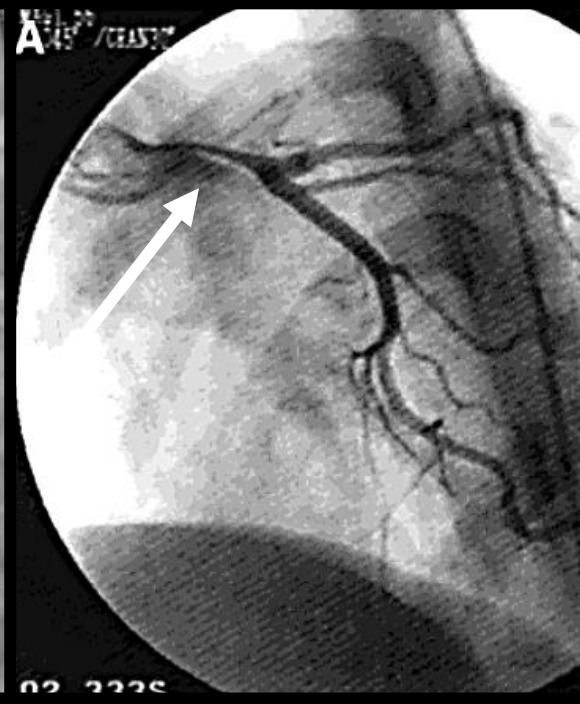
Table 3. Characteristics of patients with or without Left coronary artery compression

	LMCA		
	Abnormal	Normal	
mPAP (mmHg)	43.6 +/- 17.3	27.1 +/- 5.5	p< 0.01

Kokyu To Junkan. 1989 Jun;37(6):649-55





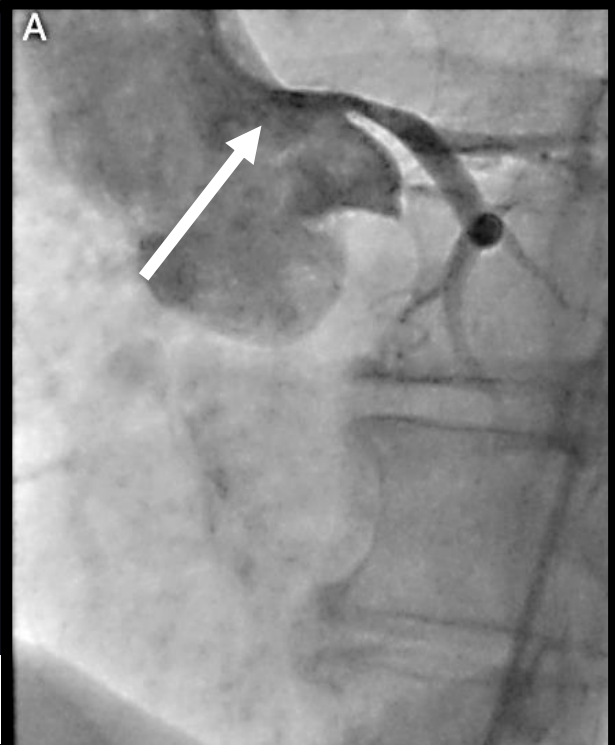


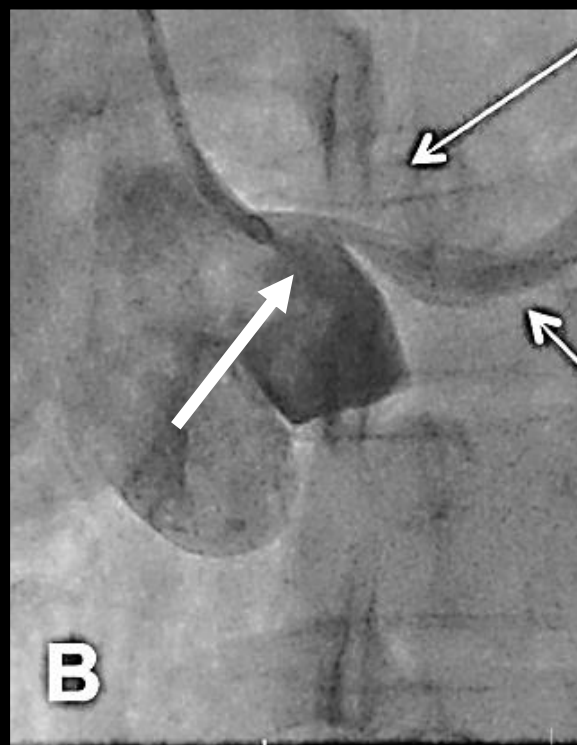
HALLAZGOS ANGIOGRAFICOS

OAL (>20°)

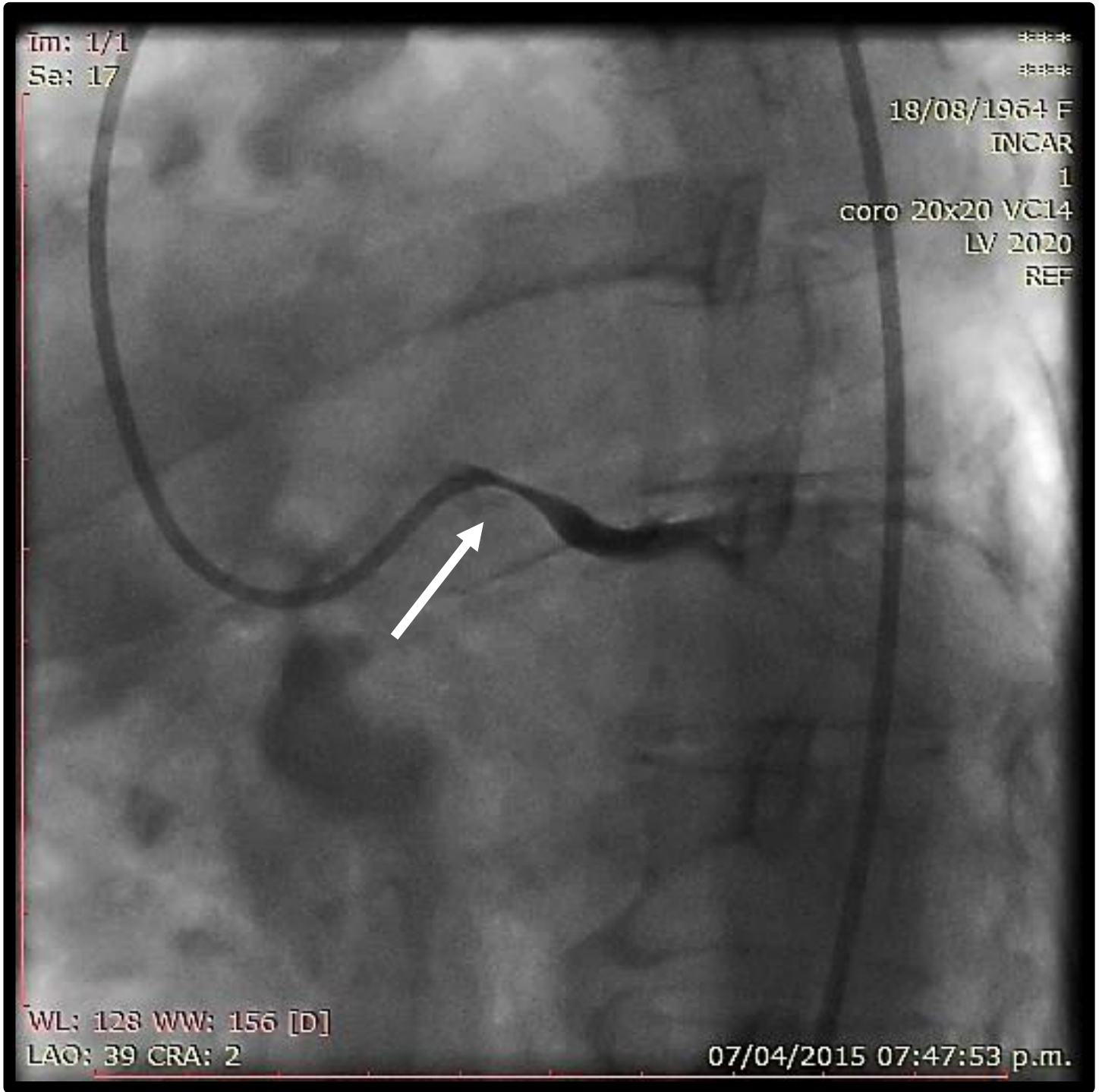
OSTIUM

DINAMICA





Kokyu To Junkan. 1989 Jun;37(6):649-55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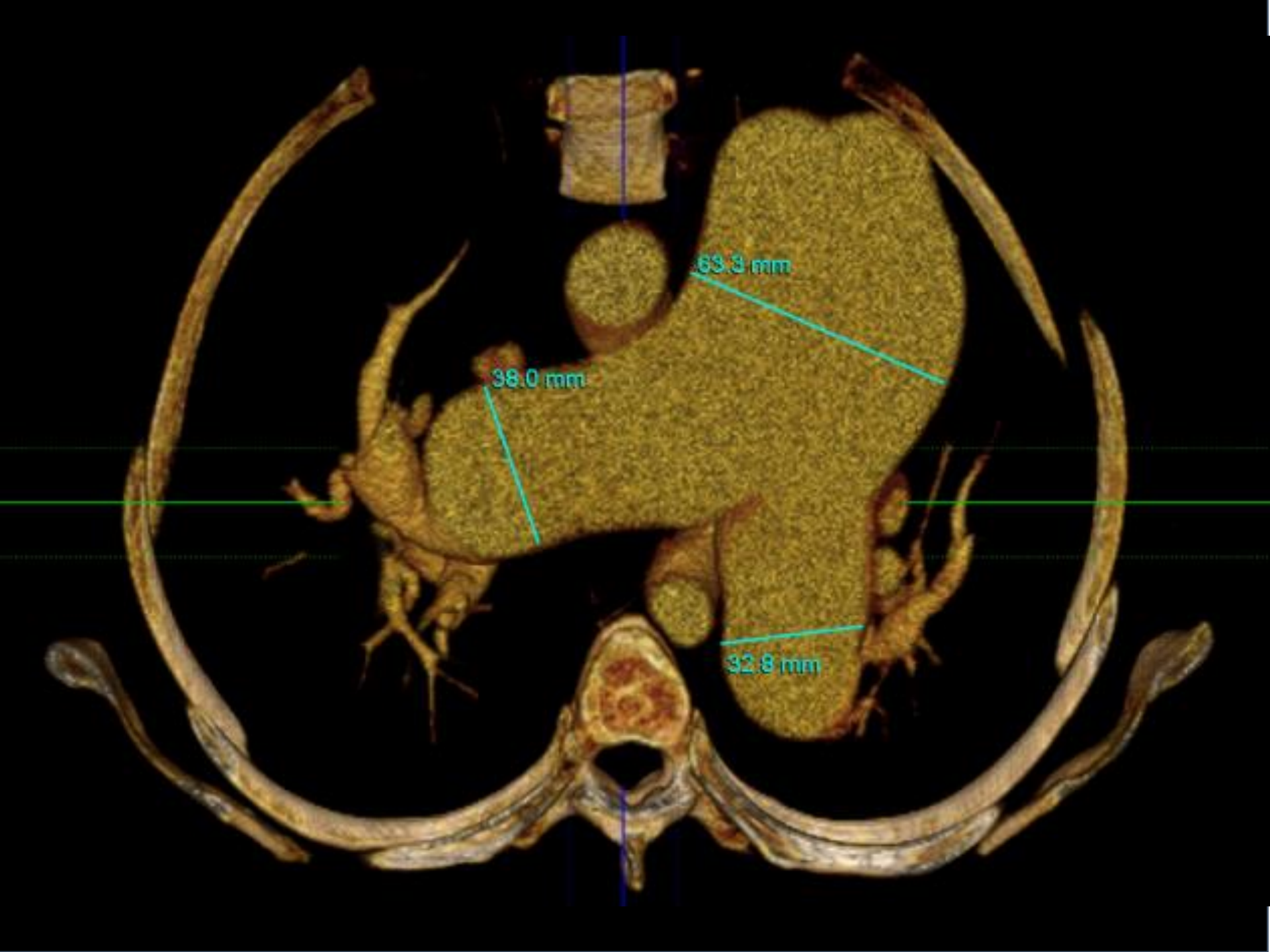
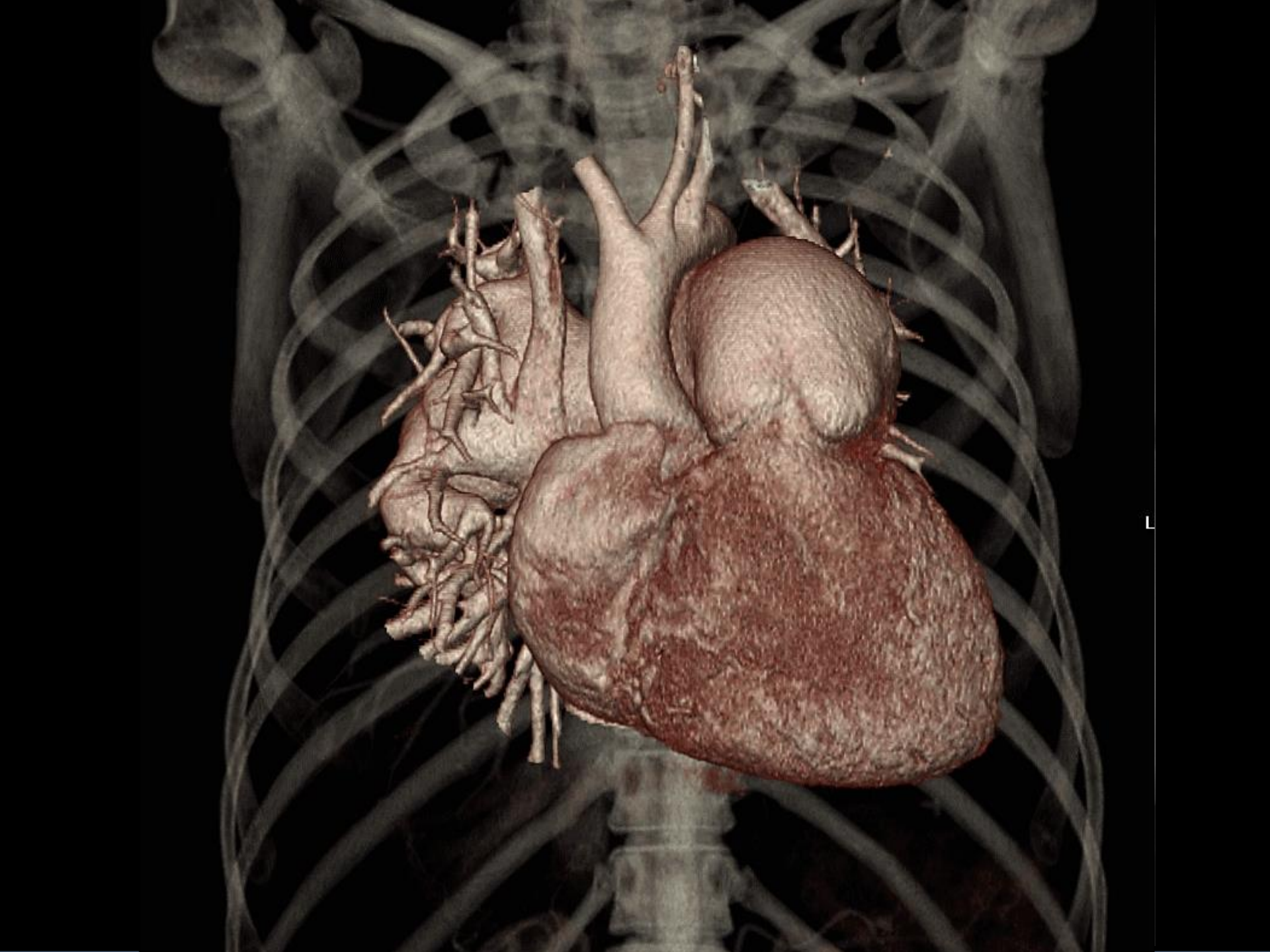
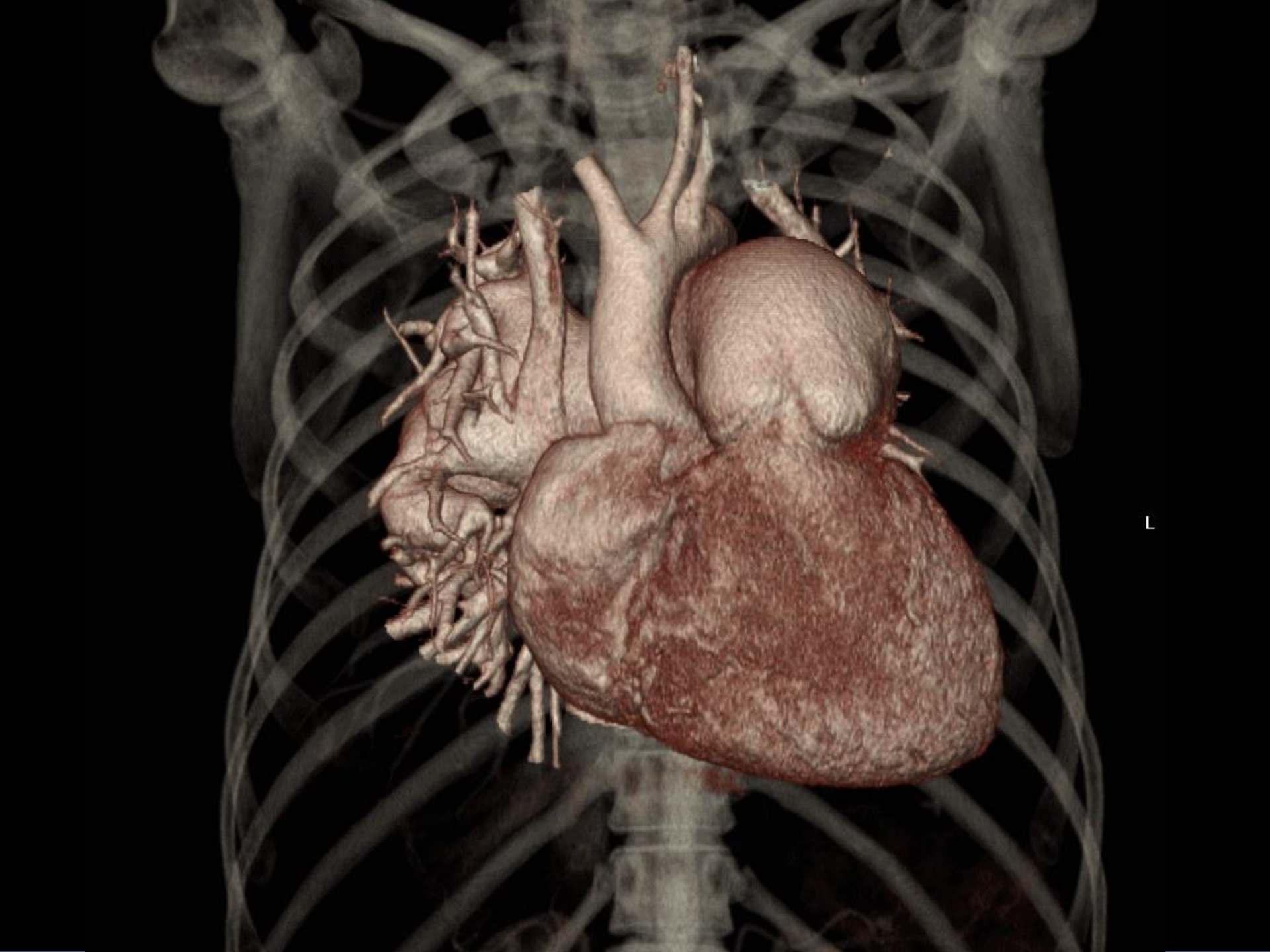
LAO: 39 CRA: 2
07/04/2015 07:47:53 p.m.

ANGIOTOMOGRAFIA


63.3 mm
38.0 mm
32.8 mm

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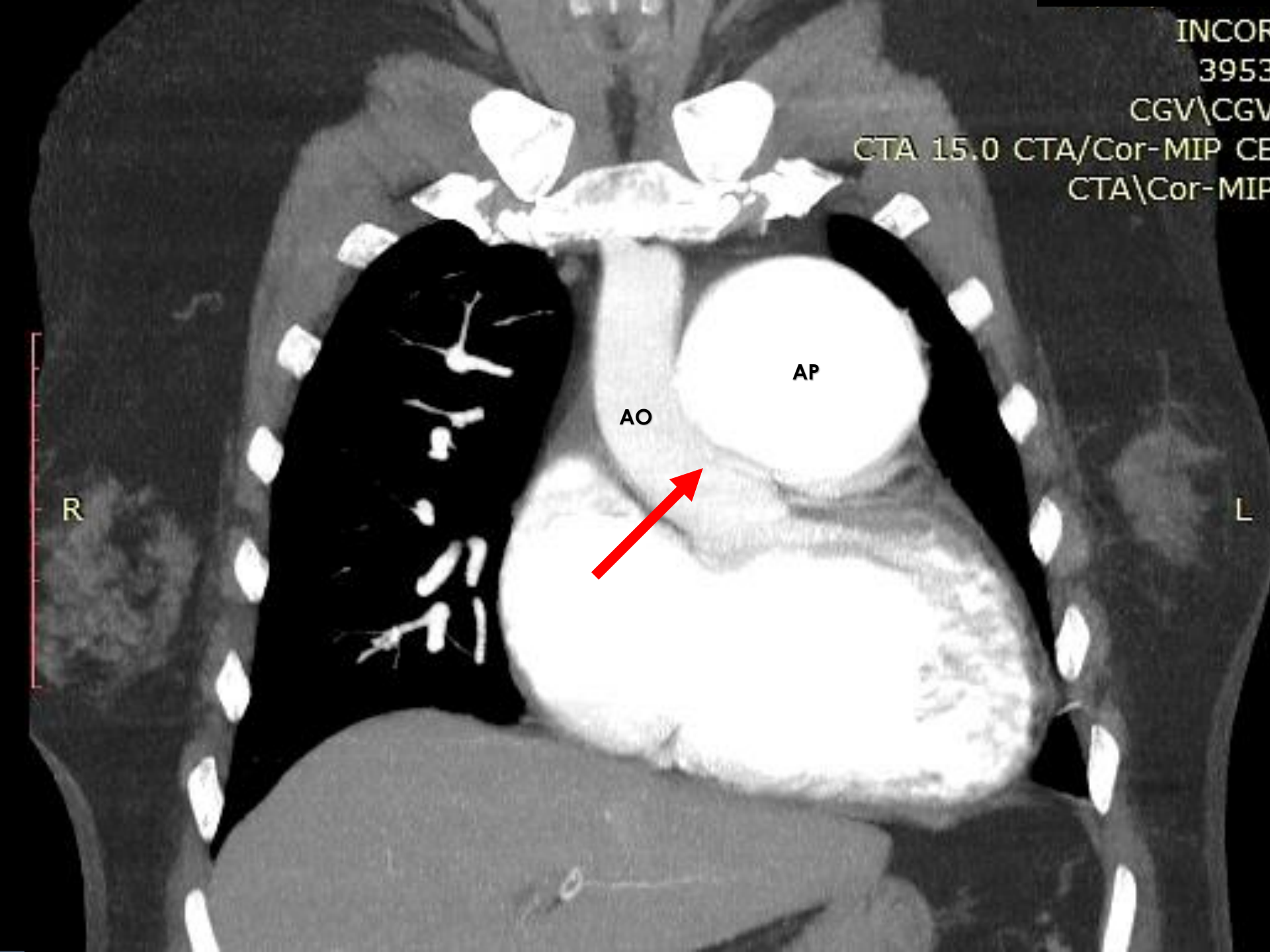
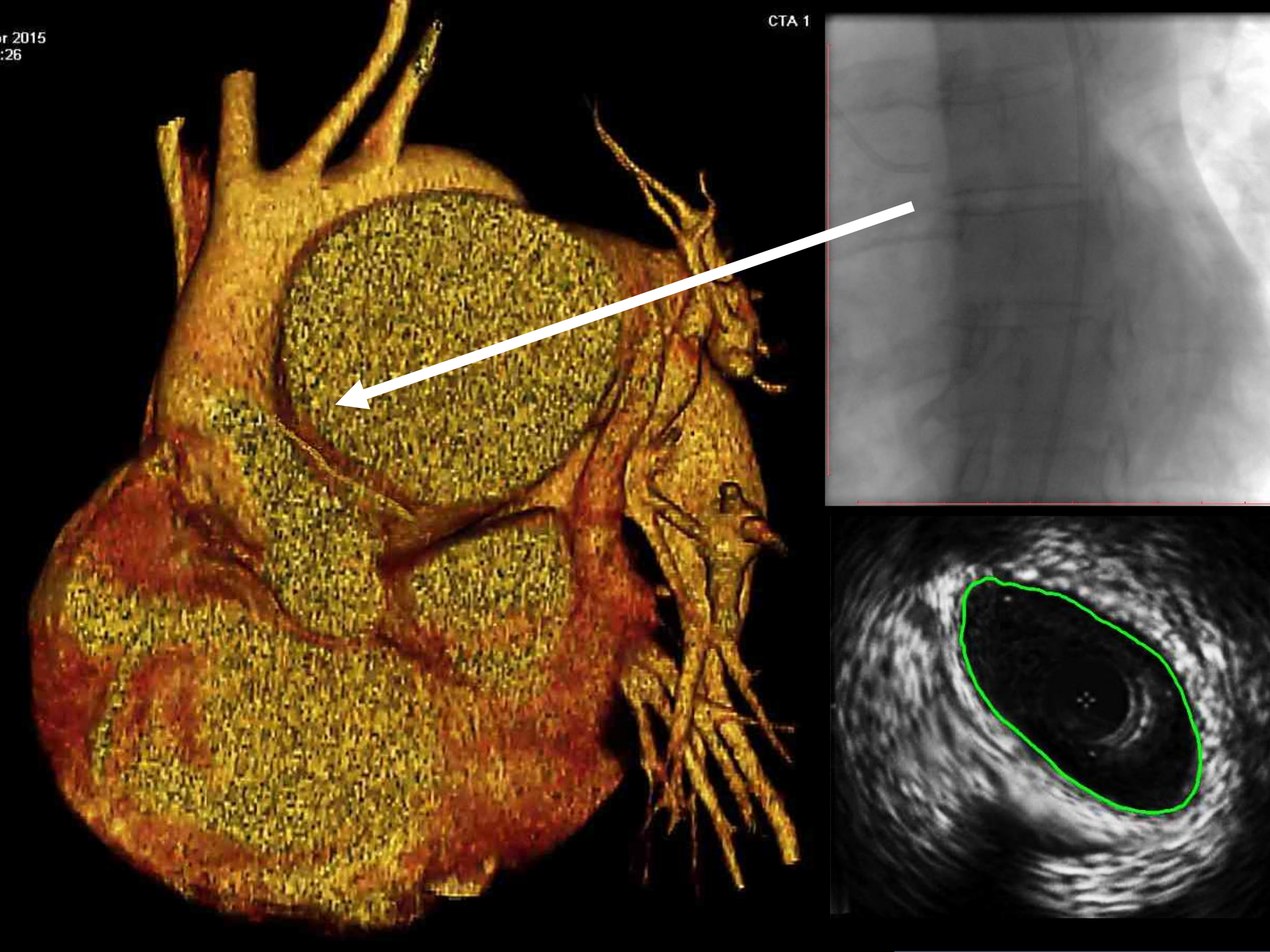

INCOR
3953
CGV\CGV
CTA 15.0 CTA/Cor-MIP CE
CTA\Cor-MIP
AP
AO
R
L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:26
CTA 1

COMPRESION EXTRINSECA DE TCI POR ARTERIA PULMONAR

MANEJO

- **CIERRE DE DEFECTO** congénito
- **REVASCULARIZACION**

  QUIRURGICA vs **PERCUTANEA**

  - Mejoria SINTOMATICA y clase funcional.

Mitsudo K, Fujino T, Matsunaga K, et al., Kokyu To Junkan 1989;37:649–55.
Gomez Varela S, Montes Orbe PM, Rev Esp Cardiol 2004;57:695–8.
Fujiwara K. Naito et al. J Thorac Cardiovasc Surg 1992; 104:449-52

COMPRESION EXTRINSECA DE TCI POR ARTERIA PULMONAR

MANEJO

INTERVENCIONISMO

- Primer stent en el 2001 : RICH buenos resultados
- TCI no protegido : ***MORBILIDAD***
- DES vs BMS (< MACE y Rvm)

Literature review of case reports of external LMCA compression due to dilated pulmonary artery causing ischemic symptoms.

Author/year	No. of PTS	Sex	Age (years)	Etiology	Symptoms	LMCA Stenosis (%)	Treatment	Follow up	Outcome
Rich et al[11]	2	F	71, 53	IPAH	Dyspnea, Angina, Syncope	90, 85	BMS	1	Alive, improved EF, free of symptoms
Varela et al[13]	1	F	31	IPAH	Dyspnea	80	DES	6	Alive, improved condition
Dubois et al[14]	1	F	51	ASD+PAH	Dyspnea, Angina	Severe ostial	DES	3	Alive, improved condition
Dodd et al[15]									
Lindsey et al									
Ginghina et									
Caldera et al									
Vaseghi et al									ree
Kothandam et al	1	M	58	PDA+PAH	Angina, Dyspnea, Heart failure	Severe ostial	1 BMS	6	Alive, relief of angina

Eficacia COMPARADA a largo plazo no establecida

K. Andjelkovic et al. / Heart & Lung 42 (2013) 382–386

CASO CLINICO

- MANEJO **QUIRURGICO**
  - CIERRE DEL DSA (PARCHE PPB)
  - RVM : Arteria Mamaria interna a DA
    Arteria Mamaria derecha a Mg
  - Plastia tricuspidea con banda 29

EVOLUCION ECOCARDIOGRAFICA

	INGRESO		PO 2	PO 45	BE CONTINUED...
AD	24cm2			24cm2	
AI	43mm	C	37mm	23cm2	
VD	48-51-74	I	36mm	45-50-73	
VID	47mm	R	35mm	43	
FEVI	60%	U	63%	65	
TAPSE		G	13		
FAC VD	35%	I	28%	36%	
PSAP	75mmHg	A			
APm			49mmHg	39mmHg	
AP (mm)	59mm		52mm	**50mm**	
	IT severa		IT leve	IT leve	
CIA			No shunts	No shunts	

CONCLUSIONES

1. La compresion extrinseca de TCI es una causa **reversible** de dolor toracico en HTP.
2. Si bien es infrecuente, debe sospecharse en pacientes con **HTP** asociados a **angina** con diametros de **AP >40mm** y Rel. **AP/ Ao >1.21**
3. Ante la sospecha: realizar **angiografia** coronaria
4. **Revascularizacion** mejora **sintomas** y CF.
5. Potencial reduccion de **mortalidad** en pacientes con HP.

GRACIAS por su atención