The diagnosis and treatment of aortitis in Behcet's disease

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CONTROVERSES ET ACTUALITÉS EN CHIRURGIE VASCULAIRE

CONTROVERSIES & UPDATES IN VASCULAR SURGERY

JANUARY 25-27 2018

MARRIOTT RIVE GAUCHE & CONFERENCE CENTER

PARIS, FRANCE **WWW.CACVS.ORG**

I do not have any potential conflict of interest

Introduction

- Multisystemic vasculitis.
- It affects : - Mucocutaneous
  - Ocular

vous

ular

- Gastro-intestinal

Introduction

- First description : Hulusi Behçet 1937.
  Turkish dermatologist.

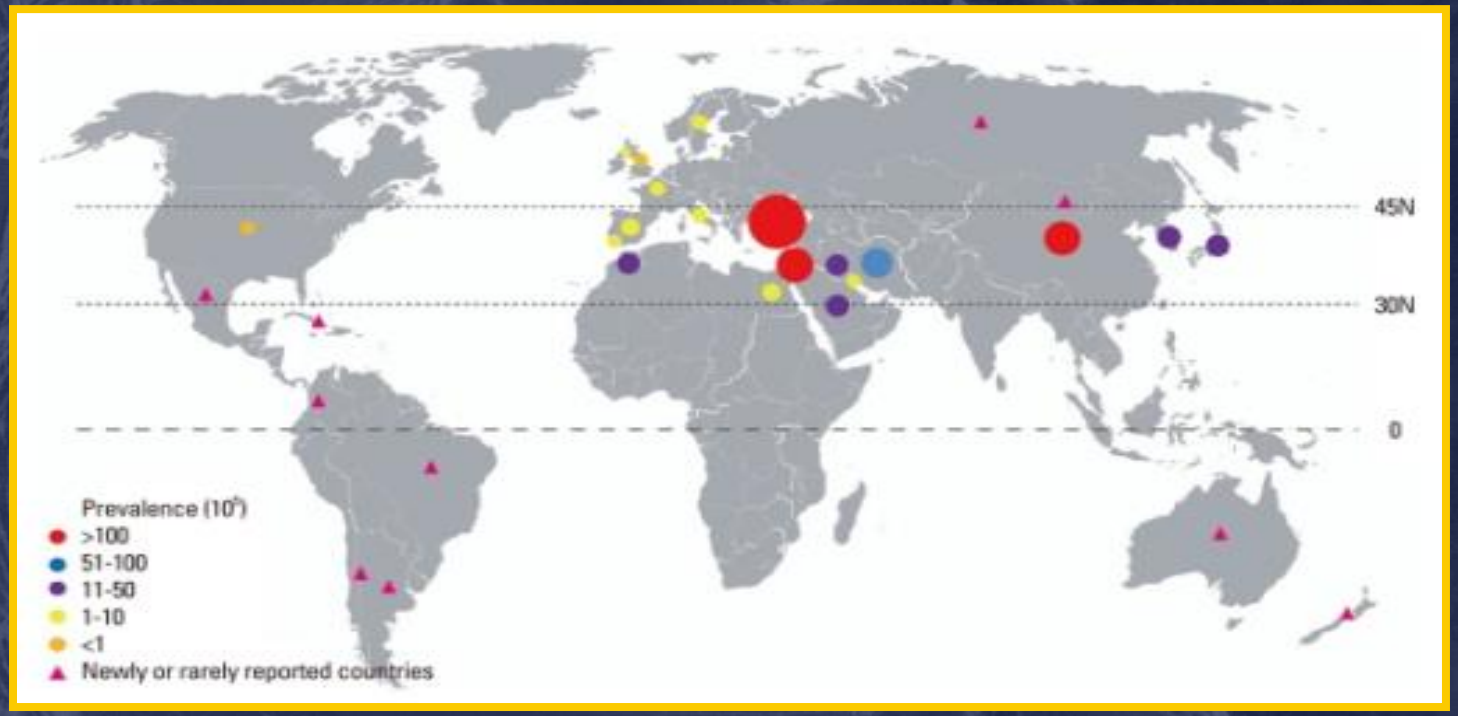
- Second or third decade of live .

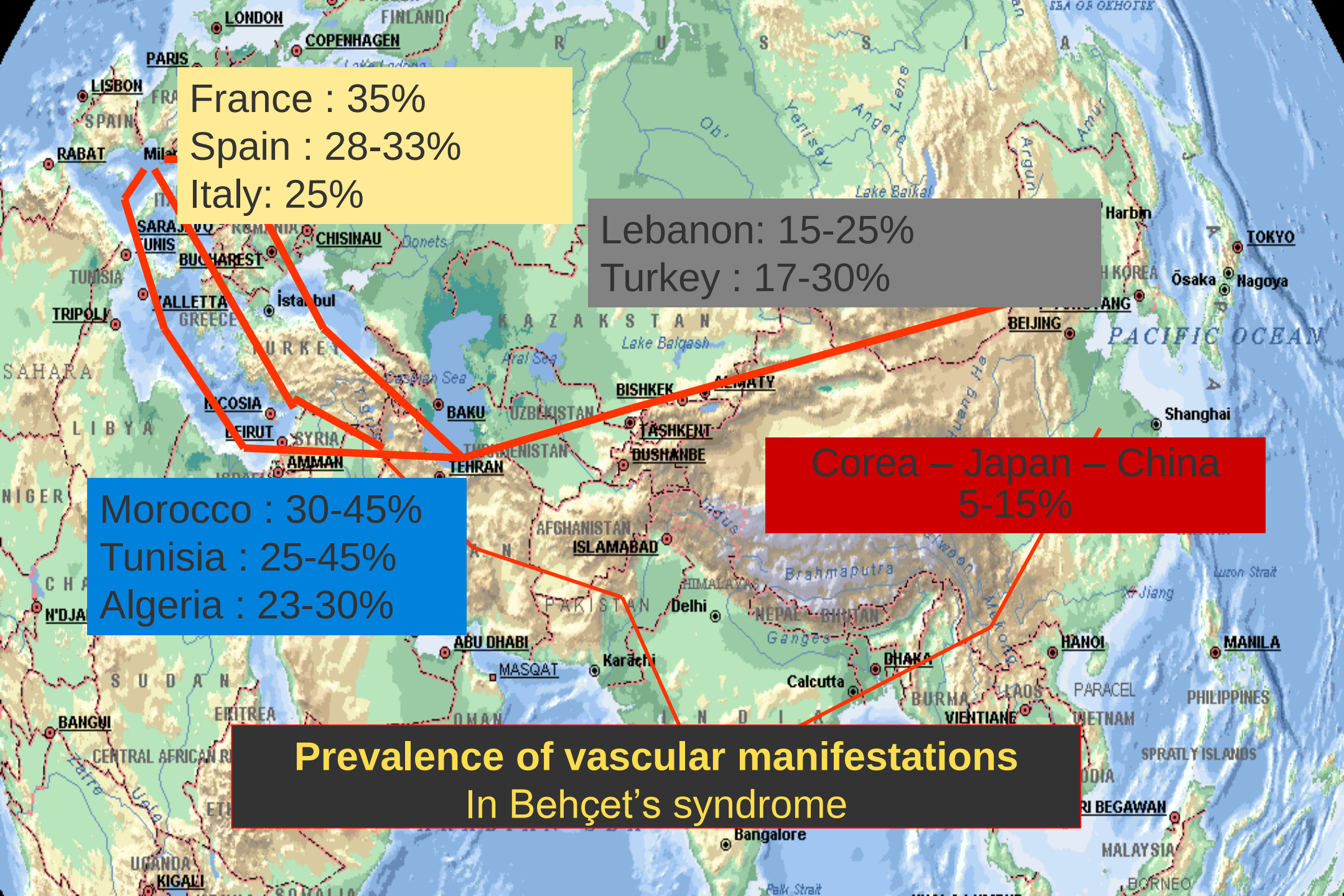
- Men : 4/1.

Geography

- Mediterraneen and asiatic countries.

Turkey :300/100000-Japan-Tunisia :10/100000




France : 35%
Spain : 28-33%
Italy: 25%
Lebanon: 15-25%
Turkey : 17-30%
Corea – Japan – China
5-15%
Morocco : 30-45%
Tunisia : 25-45%
Algeria : 23-30%
Prevalence of vascular manifestations
In Behçet's syndrome
LONDON
COPENHAGEN
PARIS
LISBON
RABAT
CHISINAU
BUCHAREST
TUNIS
TRIPOLI
İstanbul
BAKU
NICOSIA
BEIRUT
AMMAN
TEHRAN
BISHKEK
ALMATY
TASHKENT
DUSHANBE
BEIJING
Harbin
TOKYO
Ōsaka
Nagoya
Shanghai
ISLAMABAD
Delhi
ABU DHABI
MASQAT
Karachi
Calcutta
DHAKA
HANOI
MANILA
VIENTIANE
BANGUI
KIGALI
Bangalore
KAZAKSTAN
PACIFIC OCEAN
SAHARA
LIBYA
SUDAN
ERITREA
INDIA
PAKISTAN
AFGHANISTAN
UZBEKISTAN
TURKMENISTAN
GREECE
TURKEY
SYRIA
NEPAL
BURMA
LAOS
VIETNAM
PHILIPPINES
MALAYSIA
BORNEO
SPRATLY ISLANDS
Lake Baikal
Lake Balqash
Aral Sea
Caspian Sea
Ob'
Yenisey
Lena
Angara
Amur
Argun
Donets
Brahmaputra
Ganges
Indus
Mekong
Jiang
Luzon Strait
Palk Strait
HIMALAYAS
PARACEL

Diagnosis

Diagnostic Criteria for Behcet's Disease†

Criterion	Required features
Recurrent oral ulceration	Aphthous (idiopathic) ulceration, observed by physician or patient, with at least three episodes in any 12 month period
Plus any two of the following	
Recurrent genital ulceration	Aphthous ulceration or scarring, observed by physician or patient
Eye lesions	Anterior or posterior uveitis cells in vitreous in slit lamp examination; or retinal vasculitis documented by ophthalmologist
Skin lesions	Erythema nodosum-like lesions observed by physician or patient; papulopustular skin lesions or pseudofolliculitis with characteristic acneiform nodules observed by physician
Pathergy test	Interpreted at 24 to 48 hours by physician

†Adapted from International Study Group for Behcet's Disease. Criteria for diagnosis of Behcet's disease. Lancet 1990; 335:1078.

Diagnosis

- Pathergy test

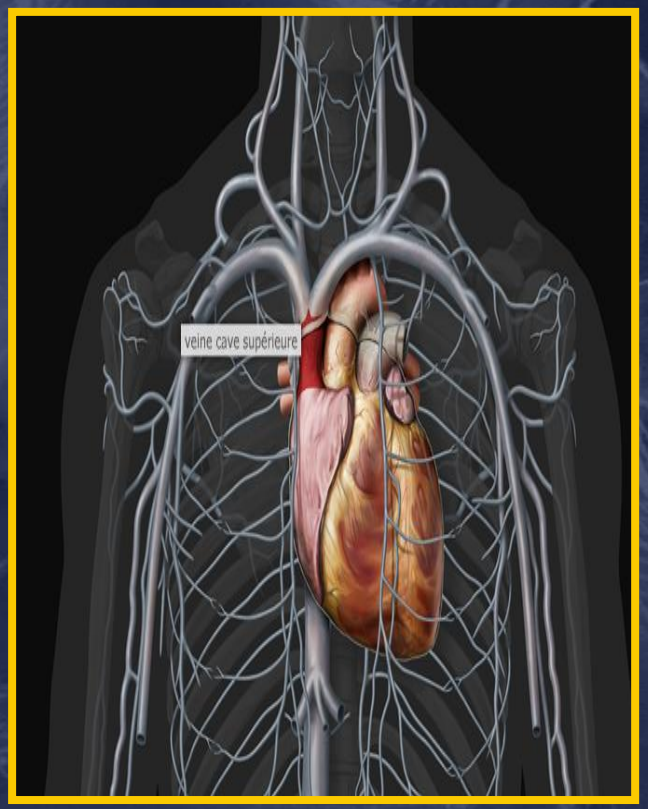
- Genetic predisposition : HLA BW51

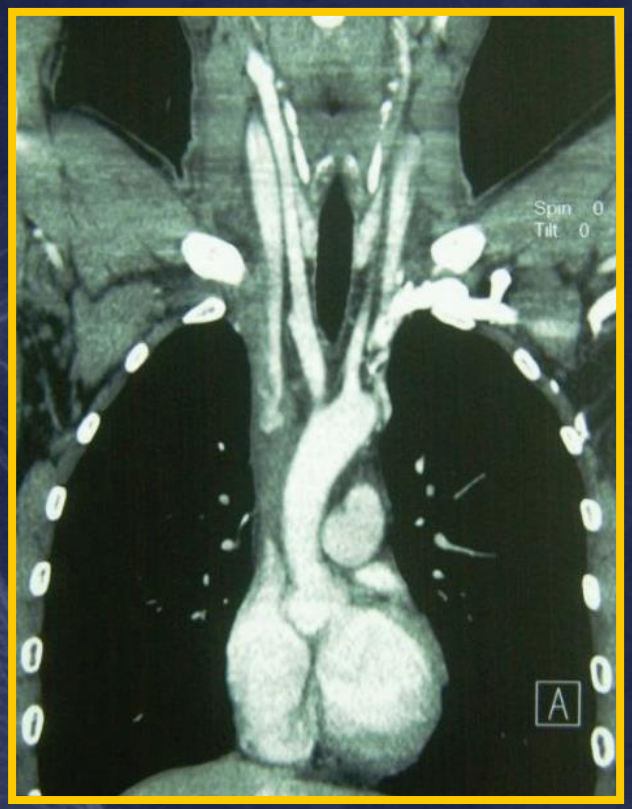
Clinical course of Behçet disease is characterized

by reemissions and exacerbations.

Vasculo-Behçet's

- 3,6-24 %. Venous +++





Vasculo-Behçet's

- Artery involvement : 1.5-2.2 %
  - Aneurysm : 65 %
  - Occlusion : 35 %
- Major cause of death because the risk of rupture .
- Interval betwen initial diagnosis of BD and diagnosis of vasculo-Behçet is 6-8 years.

Occlusion

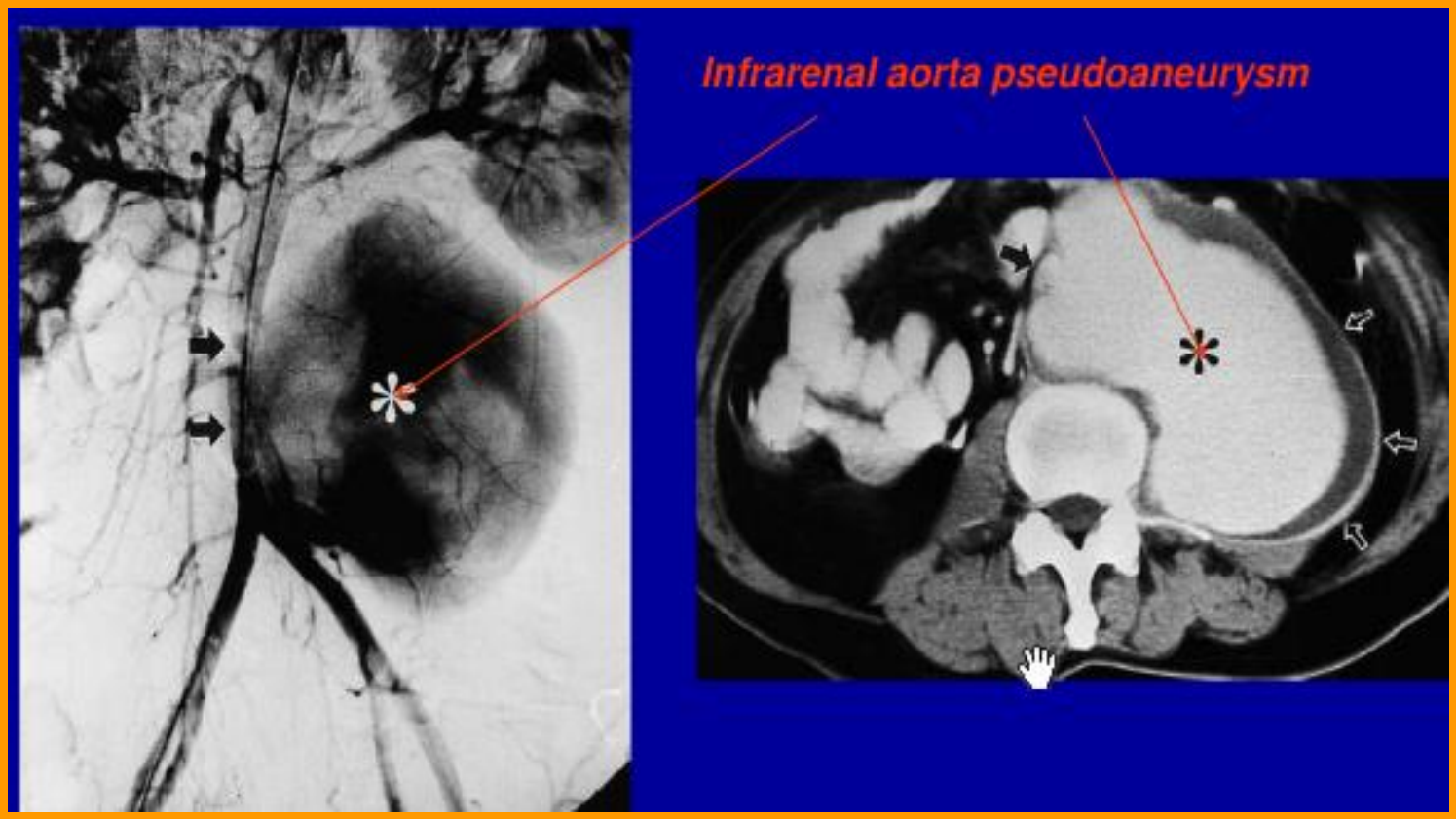
Aneurysms

First case of arterial involvement reported by Mishima in 1961.

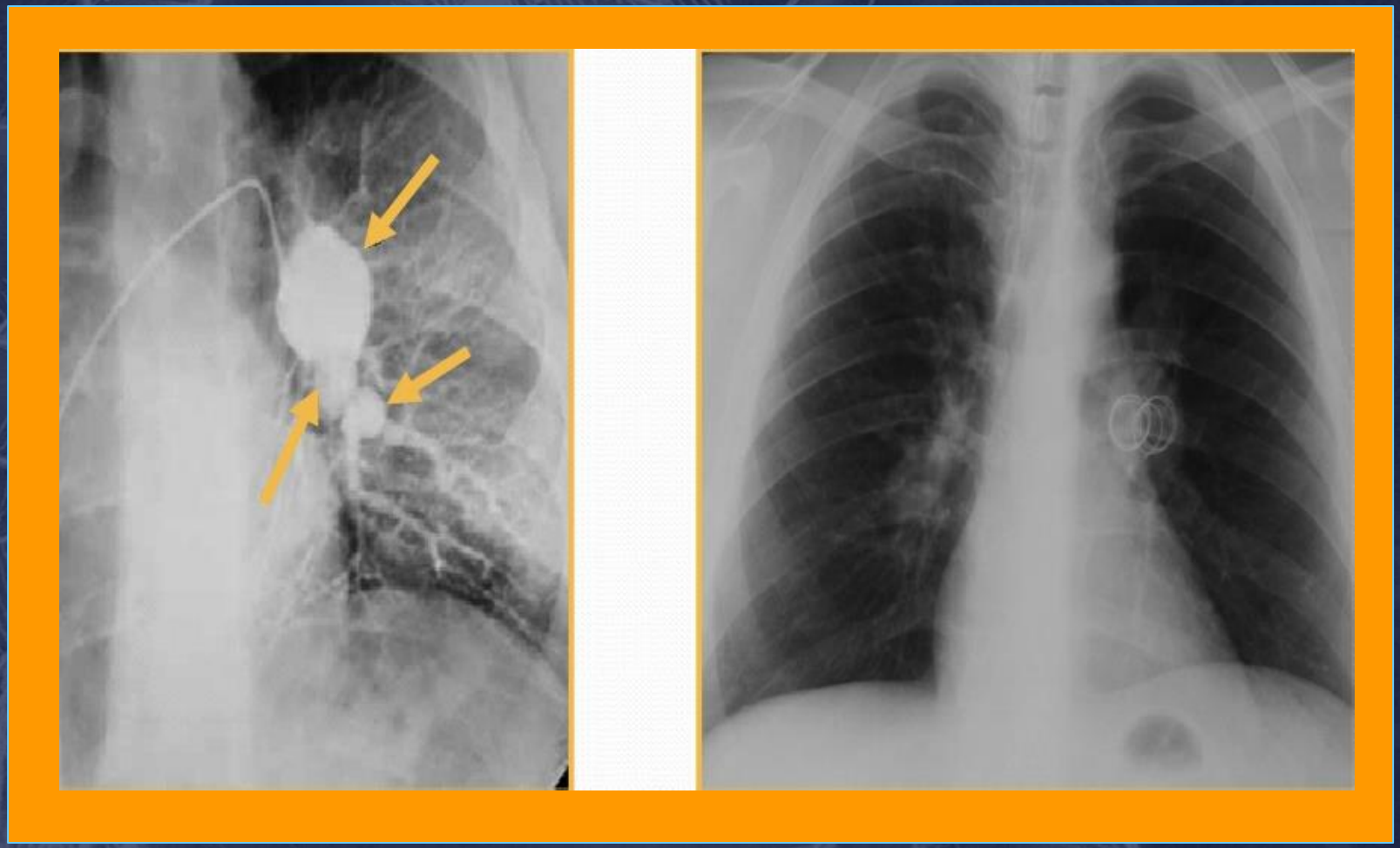
- Aorta.
- Pulmonary artery.
- Femoral.
- Others .

Multiple aneurysms : 18 %.

Abdominal Aorta



Pulmonary artery

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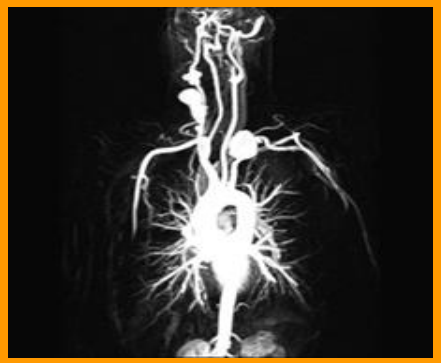
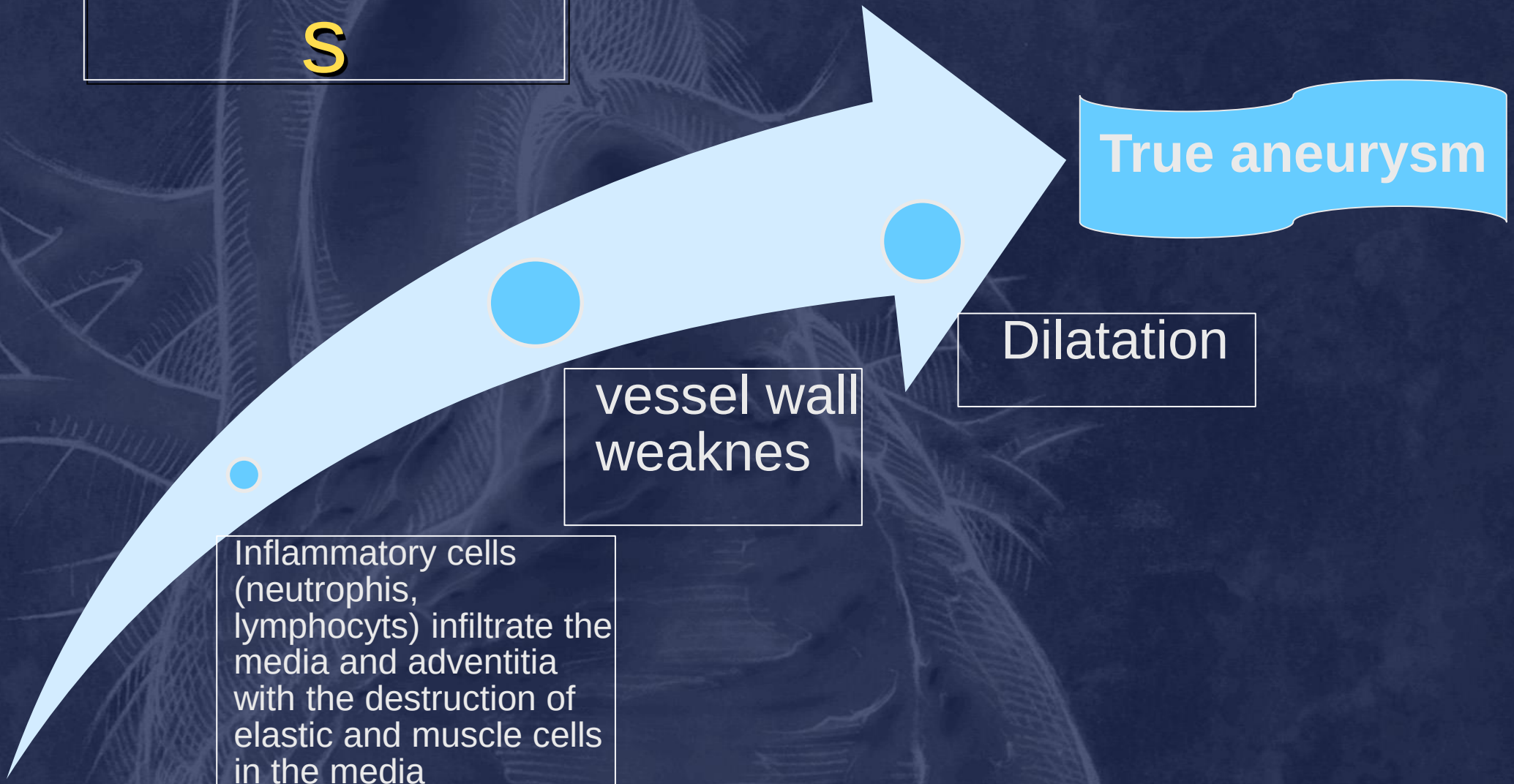
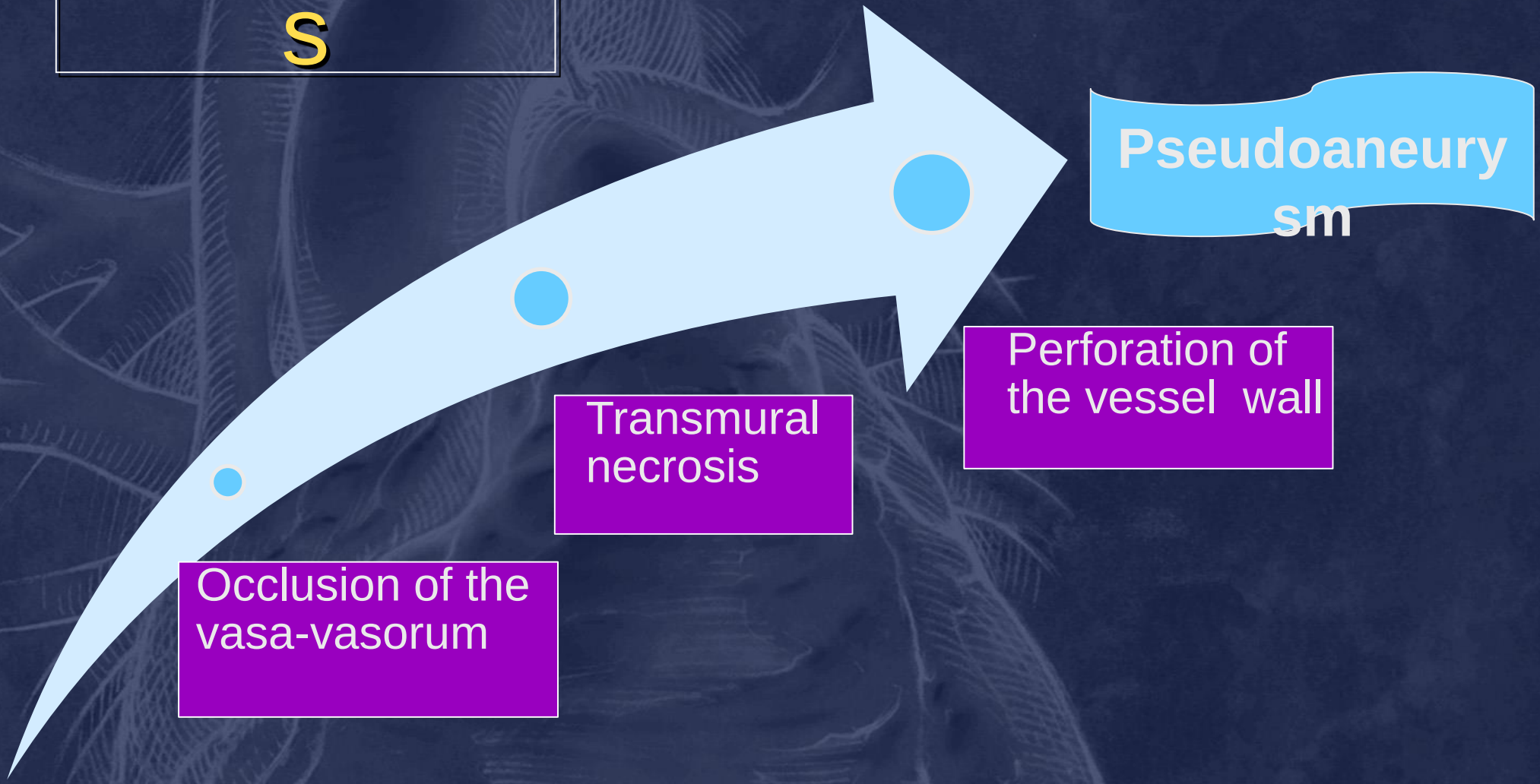
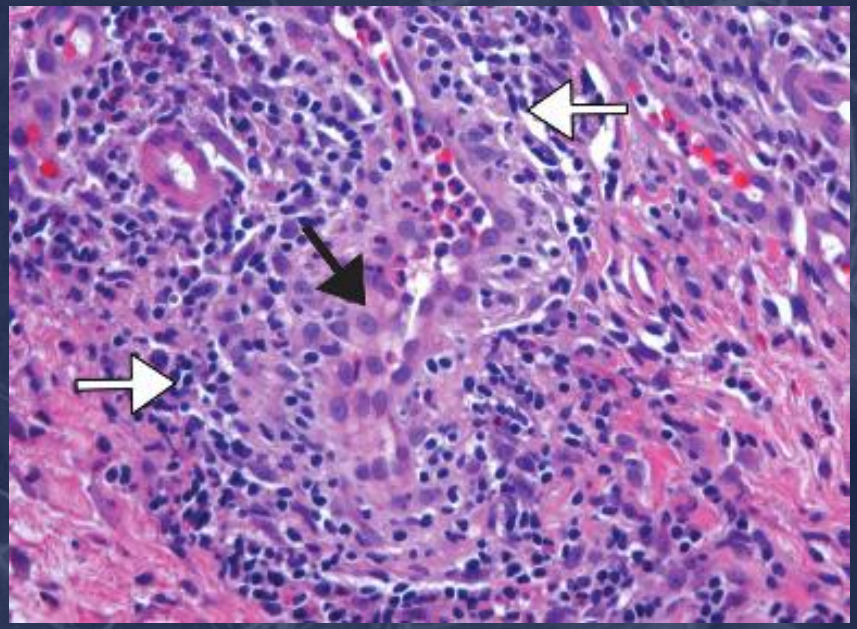


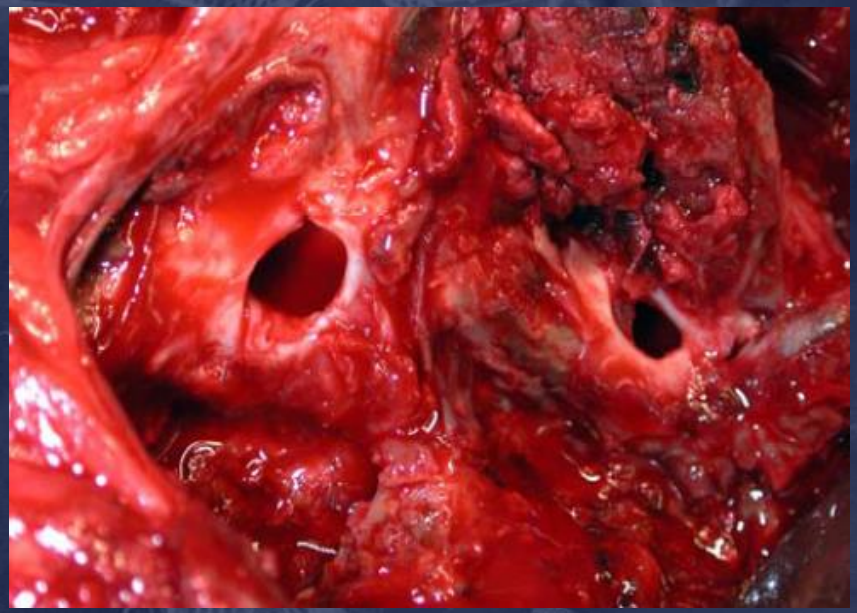
Pathogenis

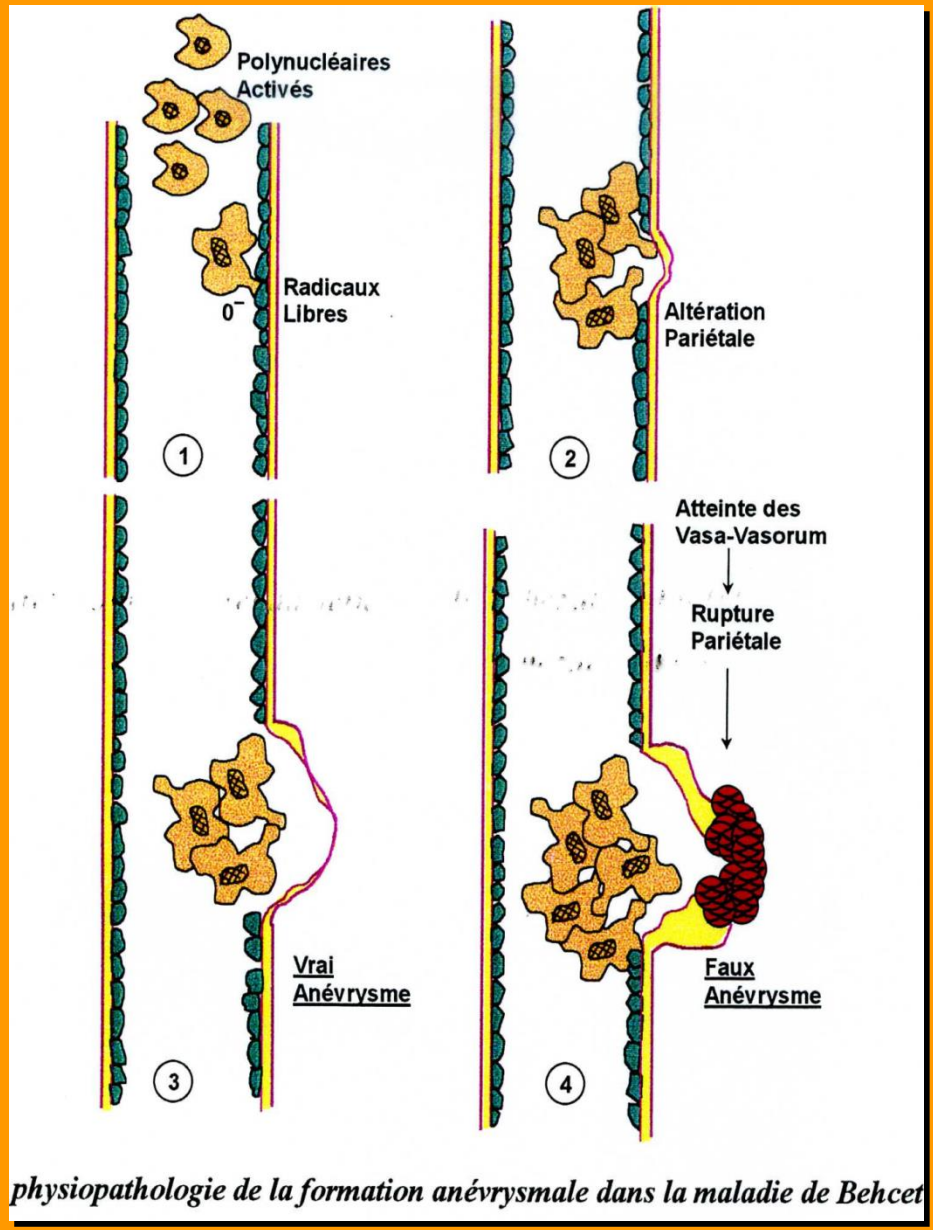

True aneurysm
Dilatation
vessel wall weaknes
Inflammatory cells (neutrophis, lymphocyts) infiltrate the media and adventitia with the destruction of elastic and muscle cells in the media

Pathogenis
s







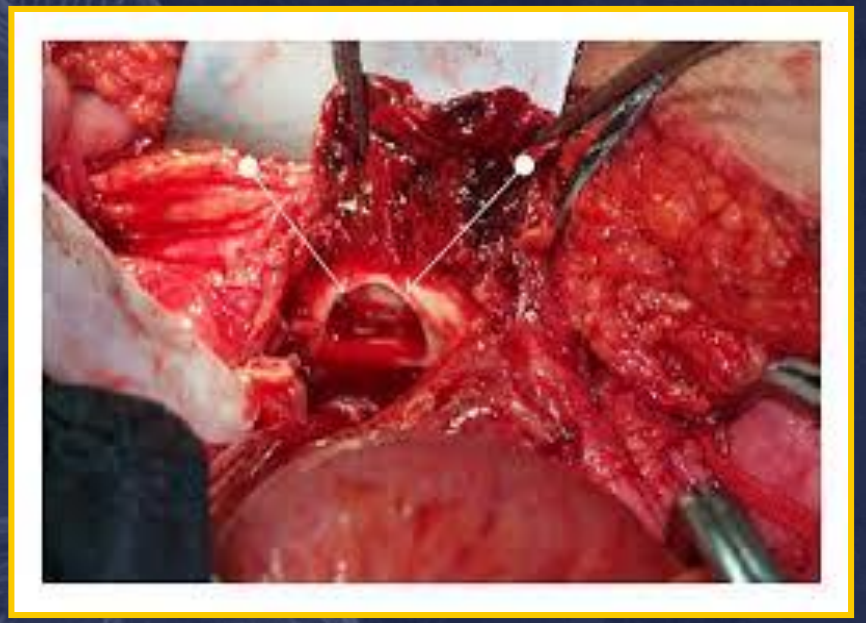


physiopathologie de la formation anévrysmale dans la maladie de Behcet

Clinical presentation

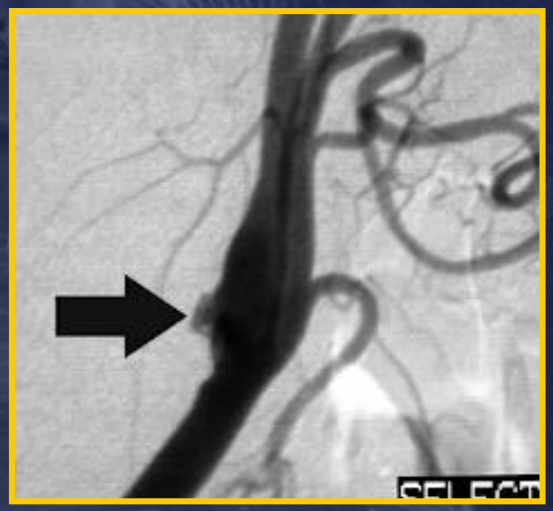
- Thoracic and abdominal aneurysms are discovered in the chronic stage with vague symptoms(back pain and abdominal disconfort).
- Complications :

  - Rupture : 60 %.
    - *Duodenum
    - *Vena cava
    - *Vertebral
  - -Retroperitoneal fibrosis .



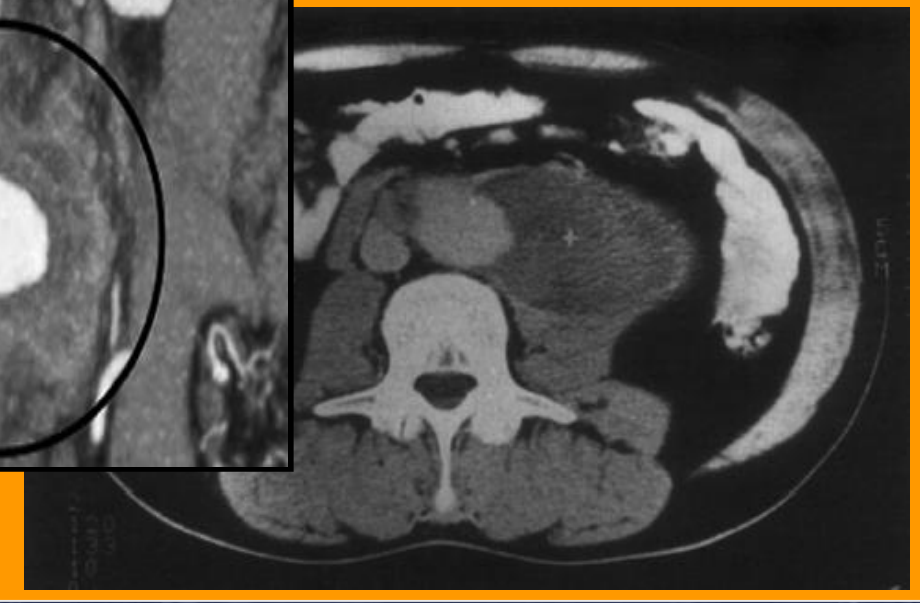
Explorations

- Duplex Sonography.
- Arteriography by arterial puncture must be avoided ⟶ intravenous digital substraction angiography.
- CT scan.
- MRA.

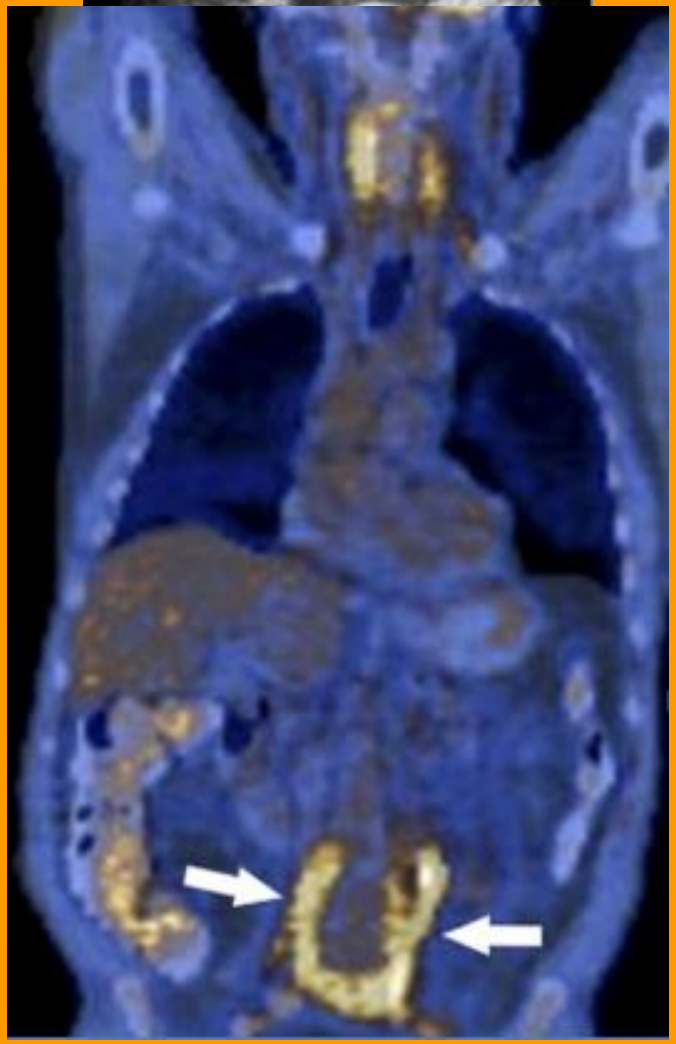


- PET scan (assessing inflammatory activity).

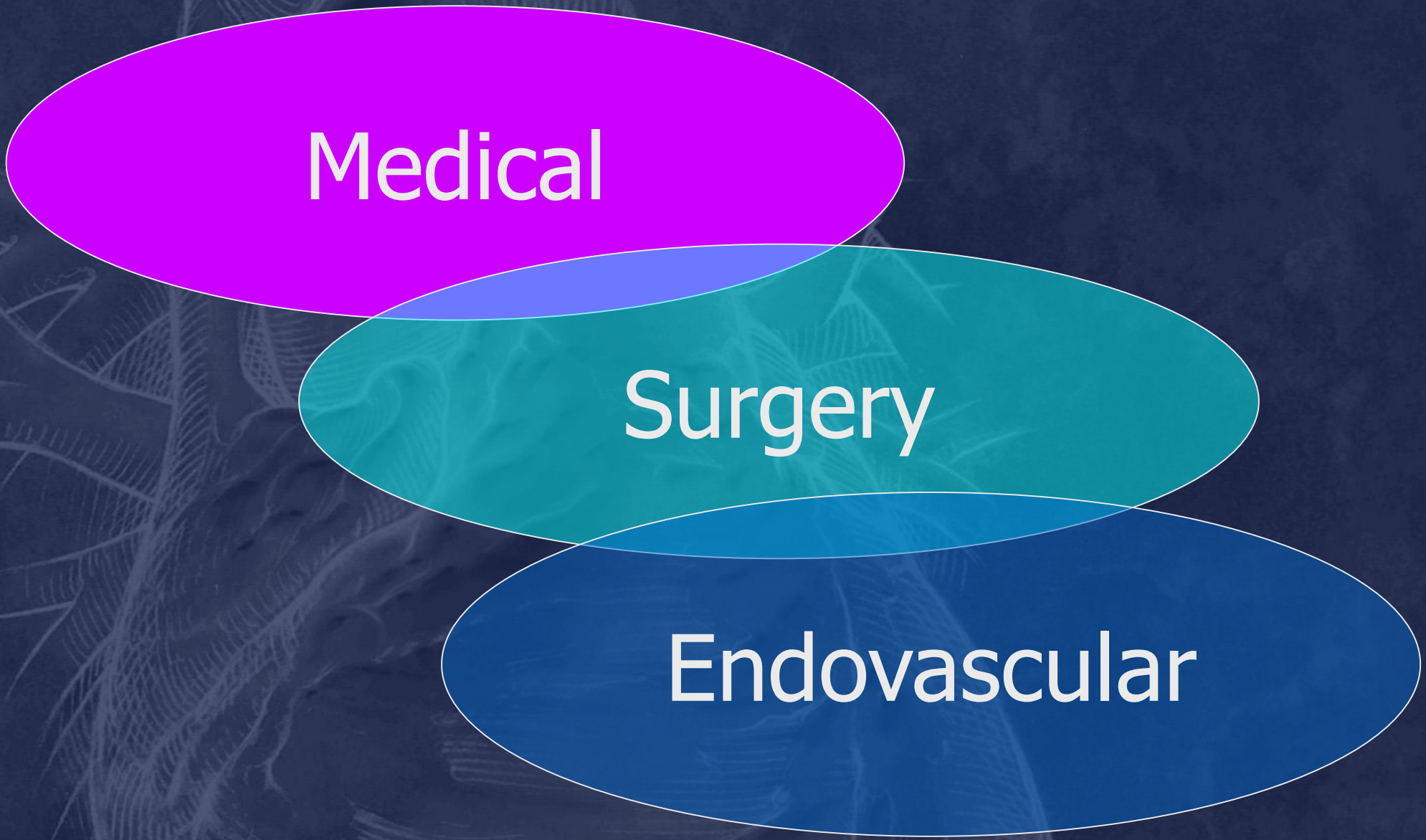

Spin:
Tilt: -9
F



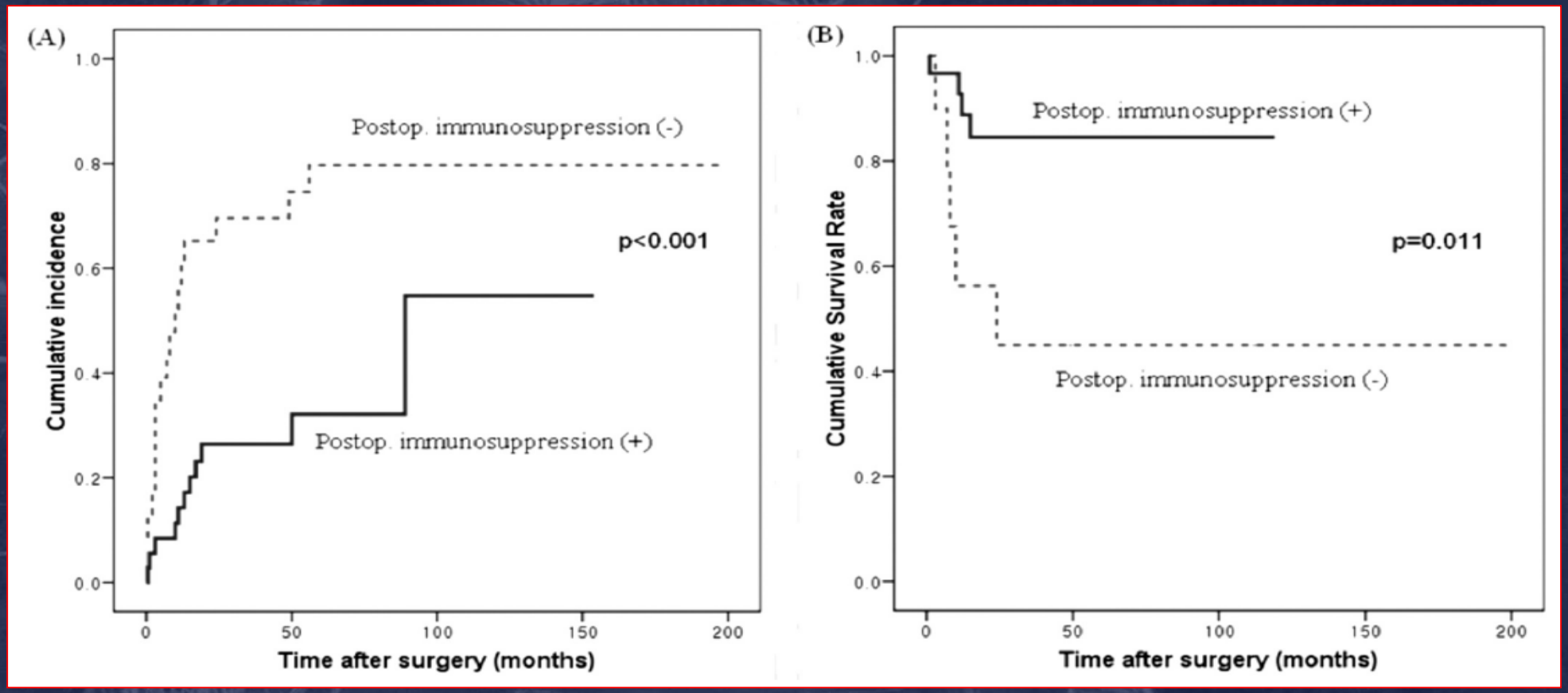

ROMB



Treatment



Medical treatment



Medical treatment

- Protocole EULAR 2008.

  Solumedrol and cyclosporin .

  Duration : 1 year (normalized CRP).

  Colchicin.

  Anti-platelet .

  Antivitamine K.

Surgery

- Challenging pathology for cardio-vascular surgeon.
- Aneurysm should be repared at the time of the detection because of the high risk of rupture .

Agressive approch.

Methods

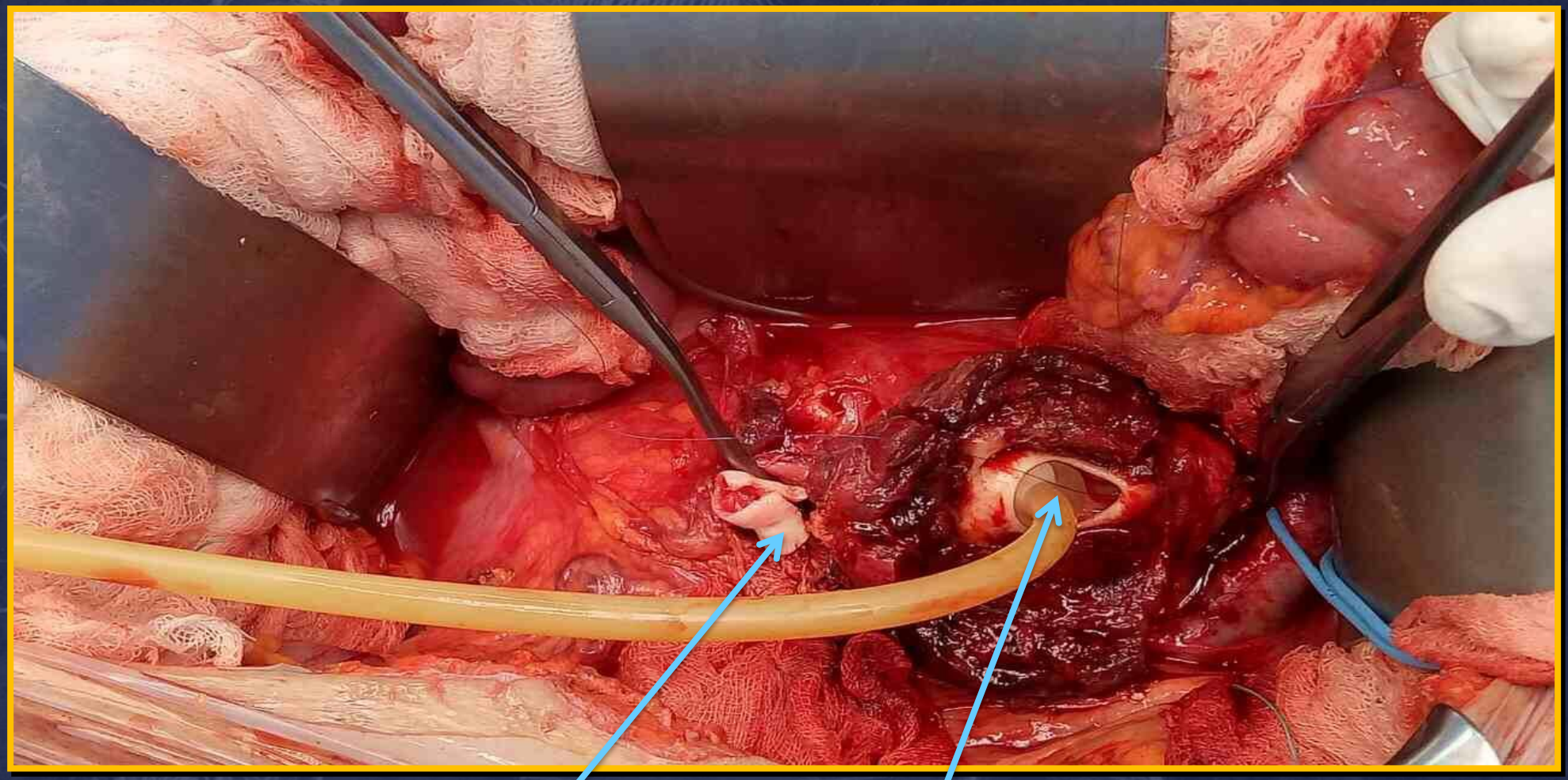
- Aneurysmorraphy.
- Simple patching :

  -smal defect.

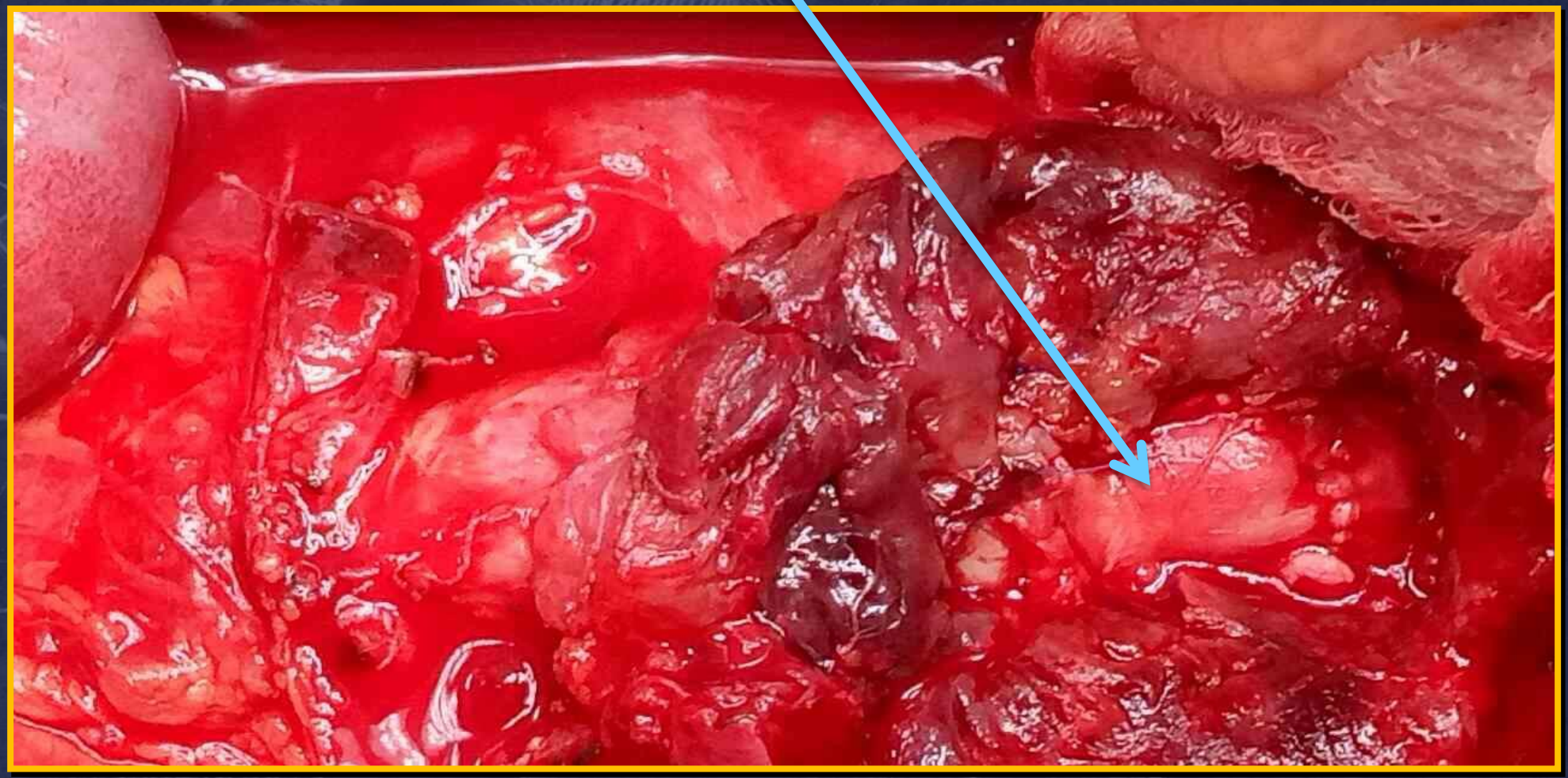
  -supra-renal aneurysm.

High recurrence+++

Perforation of medial abdominal aorta



False aneurism of medial abdominal aorta: arterial patch

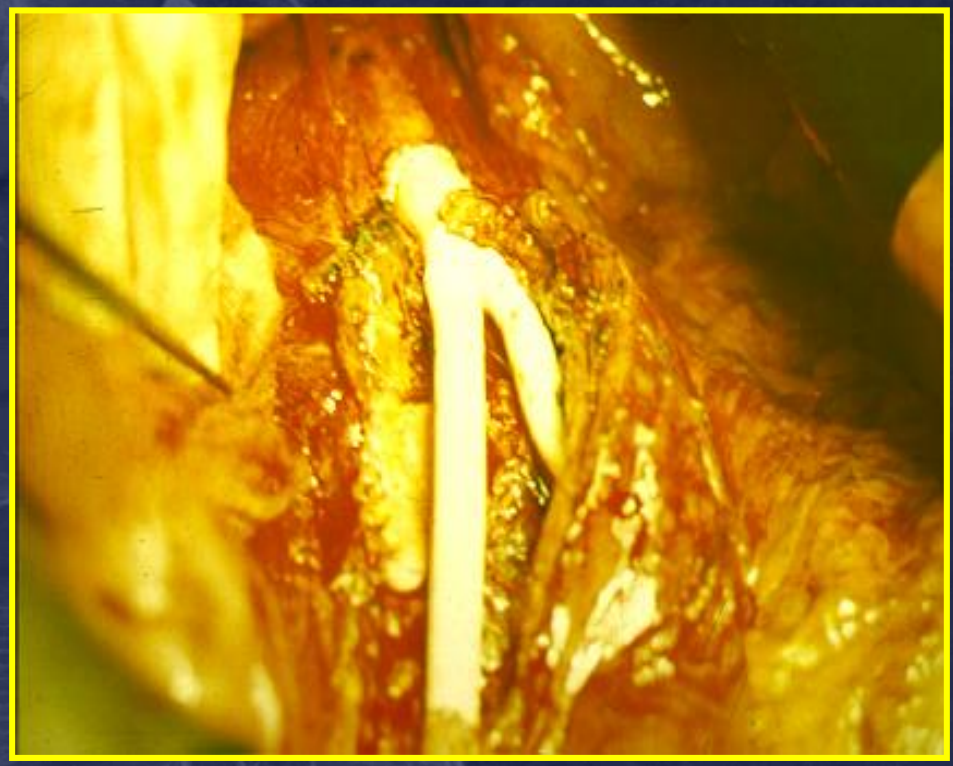


Methods

- Abdominal aorta tube graft :

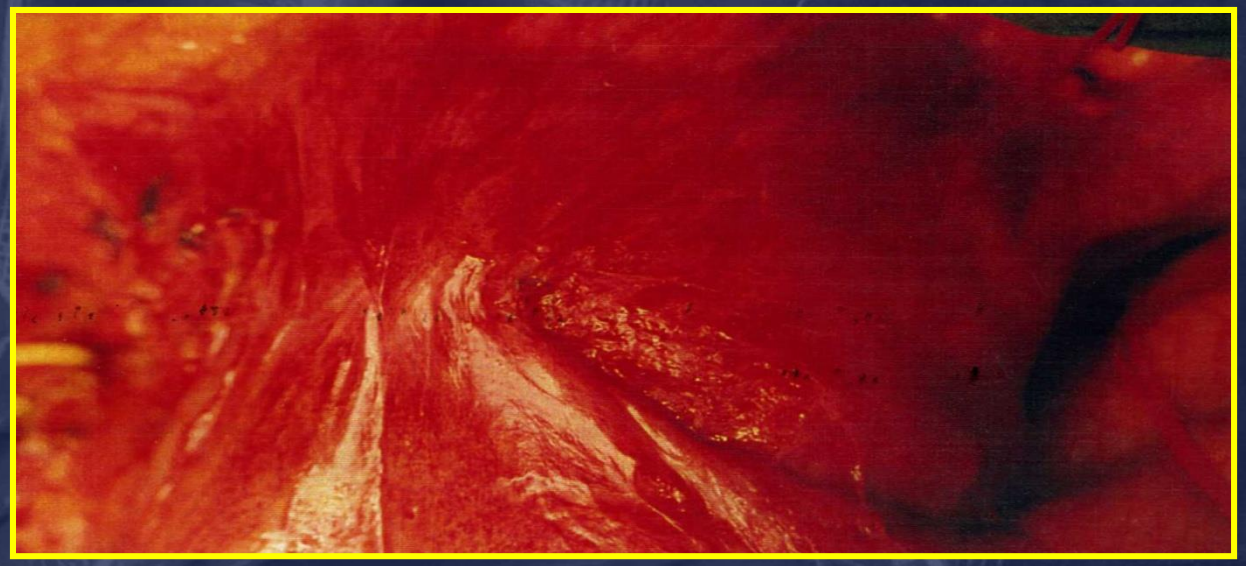
  PTFE- Dacron .

- Aorto-bi-iliac bypass.



Difficulties

- Periadventicite → difficulty of dissection.



- Fragile wall → dropping anastomotic → false aneurysm.
- Aortico-duodenal fistula .

Precautions

- Little traumatic clamping .
- Anastomosis on intact arteries far away from aneurysm.
- Suture support : patch-biologic glue.
- Wrapping with the aneurysm wall and omental flap .

Results

- Poor.
- Mortality :10-30%.
- Morbidity :

  - Occlusion .

  - Recurrence : 10-50 %.

  - Graft-enteric fistula :0.4-4 %.

Endovascular therapy

- First reported case : Vasseur in 1998.

Endovascular treatment of abdominal aneurysmal aortitis in Behçet's disease

Marc-Antoine Vasseur, MD, S. Haulon, MD, J.P. Beregi, MD, T. Le Tourneau, MD, A. Prat, MD, and H. Warembourgh, MD, *Lille, France*

Arterial complications of Behçet's disease are rare and affect mainly the aorta and iliac arteries. Perforation of the arterial wall is the most common lesion, predisposing to false aneurysm or rupture. Open surgical repair is difficult, and anastomotic false aneurysms often occur because of aortic wall fragility. We report here the case of using a bifurcated stent to treat aortoiliac false aneurysms in a 37-year-old patient. Endovascular repair could be an alternative treatment of aneurysmal manifestations in Behçet's disease.
(J Vasc Surg 1998;27:974-6.)

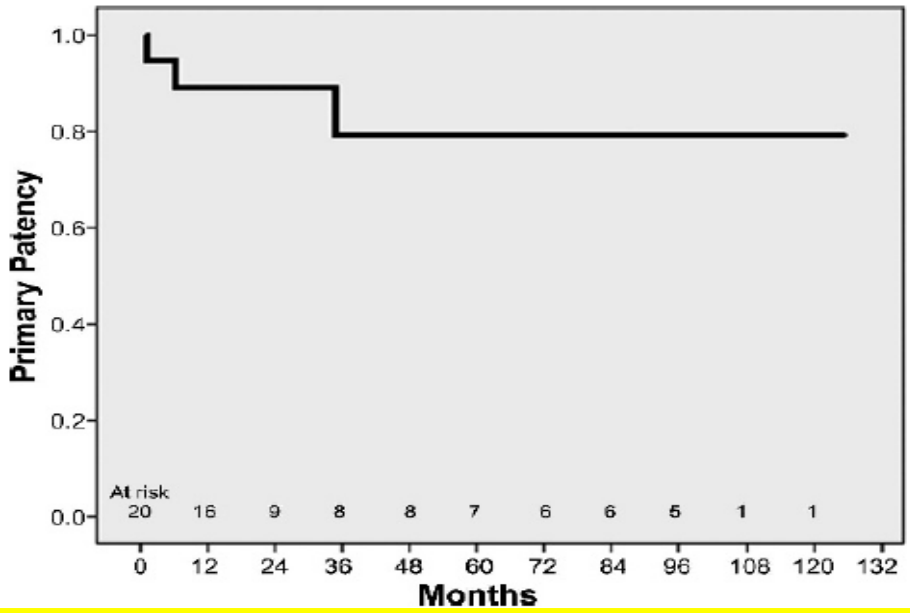
J ENDOVASC THER.
2009;16:631–636

◆ CLINICAL INVESTIGATION

Effectiveness and Safety of Endovascular Aneurysm Treatment in Patients With Vasculo-Behçet Disease

Won Ho Kim, MD; Donghoon Choi, MD, PhD; Jung-Sun Kim, MD, PhD; Young-Guk Ko, MD, PhD; Yangsoo Jang, MD, PhD; and Won Heum Shim, MD, PhD

Division of Cardiology, Yonsei Cardiovascular Hospital, Yonsei University College of Medicine, Seoul, Republic of Korea.

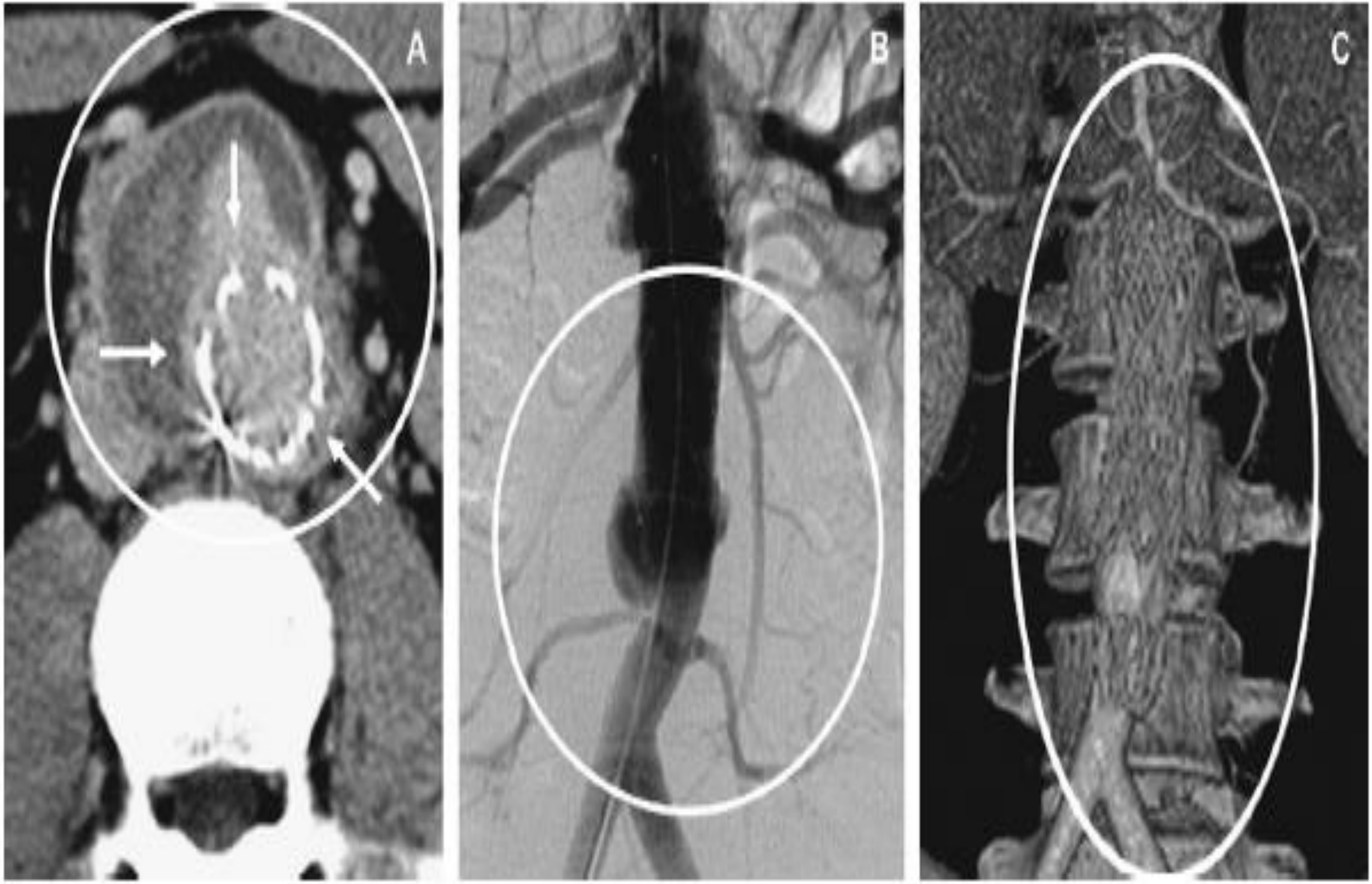


Advantages

- Lower mortality : 0,6-3,5 %.
- High technical succes rate.
- Complete regression of the aneurysm (3 months).

Complications

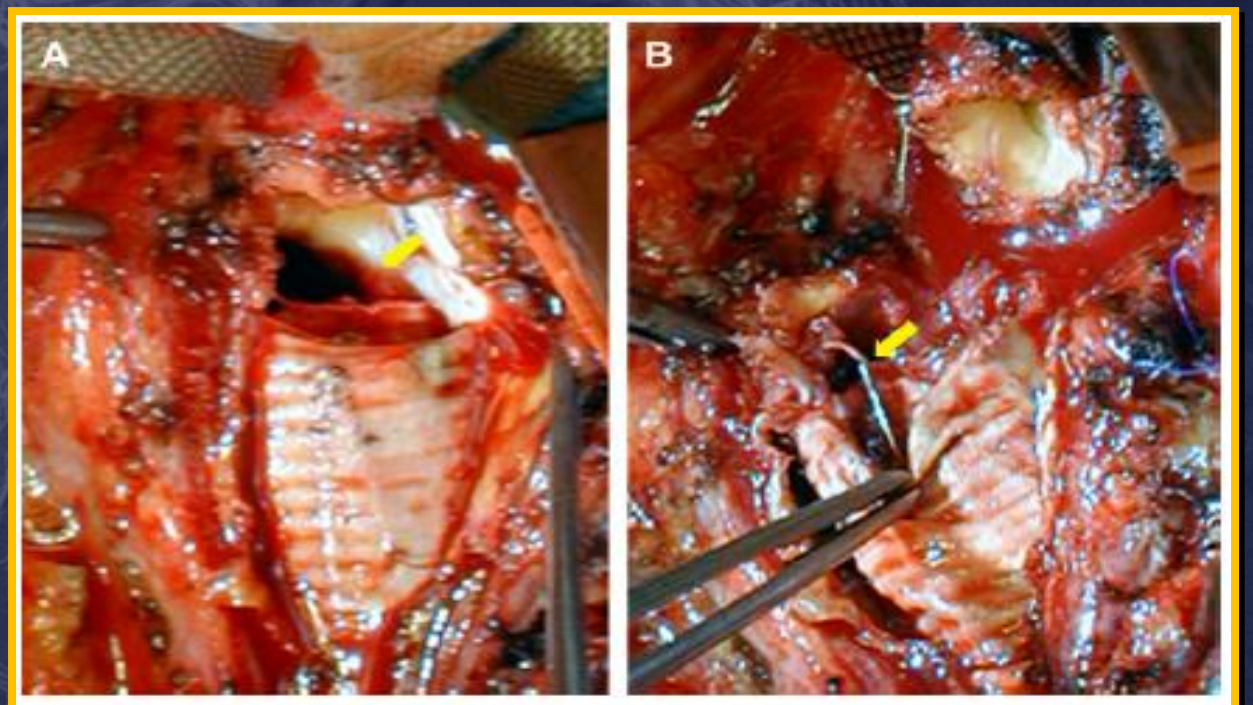
- Rate : 19 %.
- Endoleaks → rupture.
- Pseudoaneurysm at the puncture site.
- Recurrent aneurysm after EVAR
  (local mechanical stress of stent graft).


A
B
C

Successful Open Surgery for Recurrent Pseudo-aneurysm after Endovascular Aneurysm Repair in a Patient with Behçet's Disease

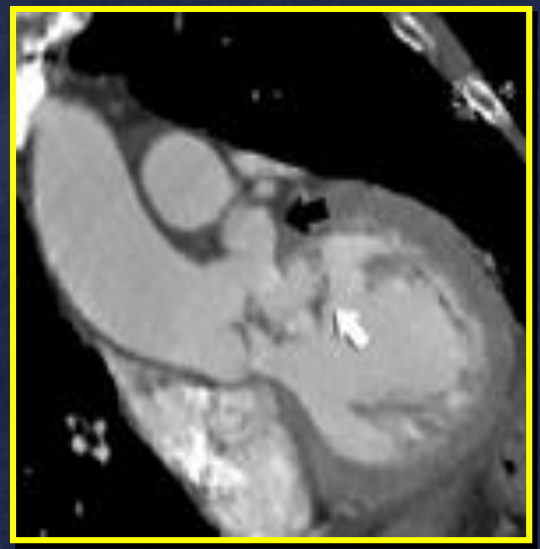
M. Nakai, S. Shimizu*, G. Kato, H. Mitsui, S. Sano

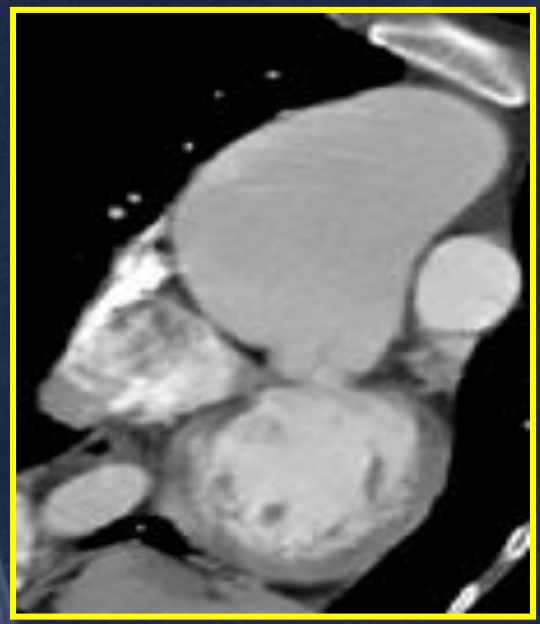
Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, 2-5-1 Shikata-cho, Kita-ku, Okayama 700-8558, Japan



Other aortic localization

- Ascending aorta .





Ruptured aneurysm of the sinus of Valsalva in a patient with Behcet's disease

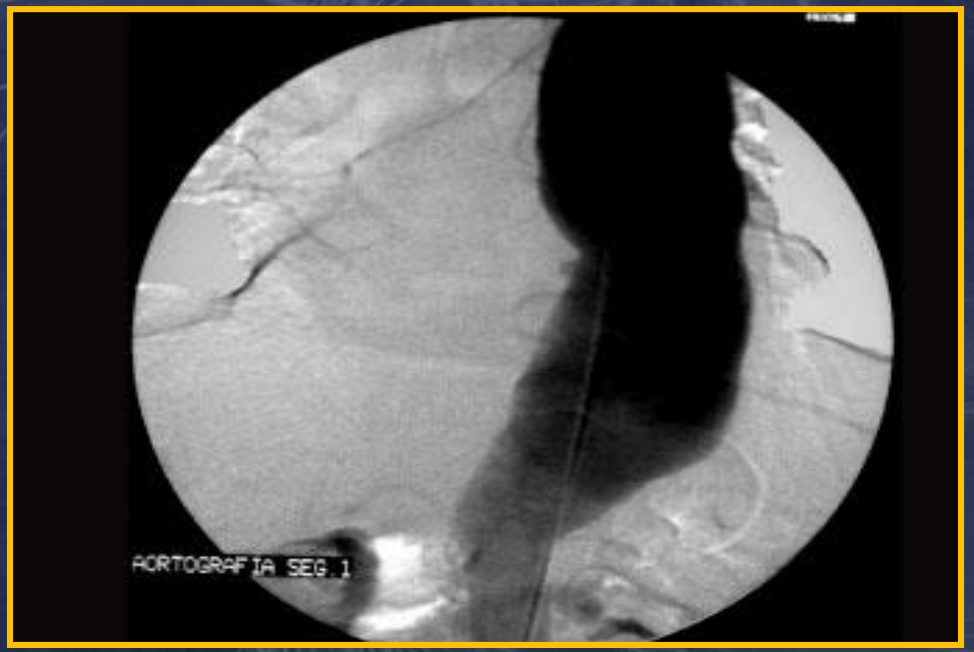
Kwang Kon Koh*[a], Ki Hoon Lee[a], Sam Soo Kim[a], Seung Chul Lee[b], Sung Hoon Jin[c], Seong Wook Cho[d]

[a]*Division of Cardiology,* [b]*Division of Dermatology,* [c]*Division of Thoracic Surgery, Department of Internal Medicine, Inha University Hospital and* [d]*Seoul National University Hospital, 3309-327 Taepyung-dong, Soojung-Ku, Sungnam-si, Kyunggi-do, Korea 461 192, South Korea*

Received 15 March 1994; revision accepted 5 July 1994

- Thoraco-abdominal aorta.


ARCO AORTICO


AORTOGRAFIA SEG 1

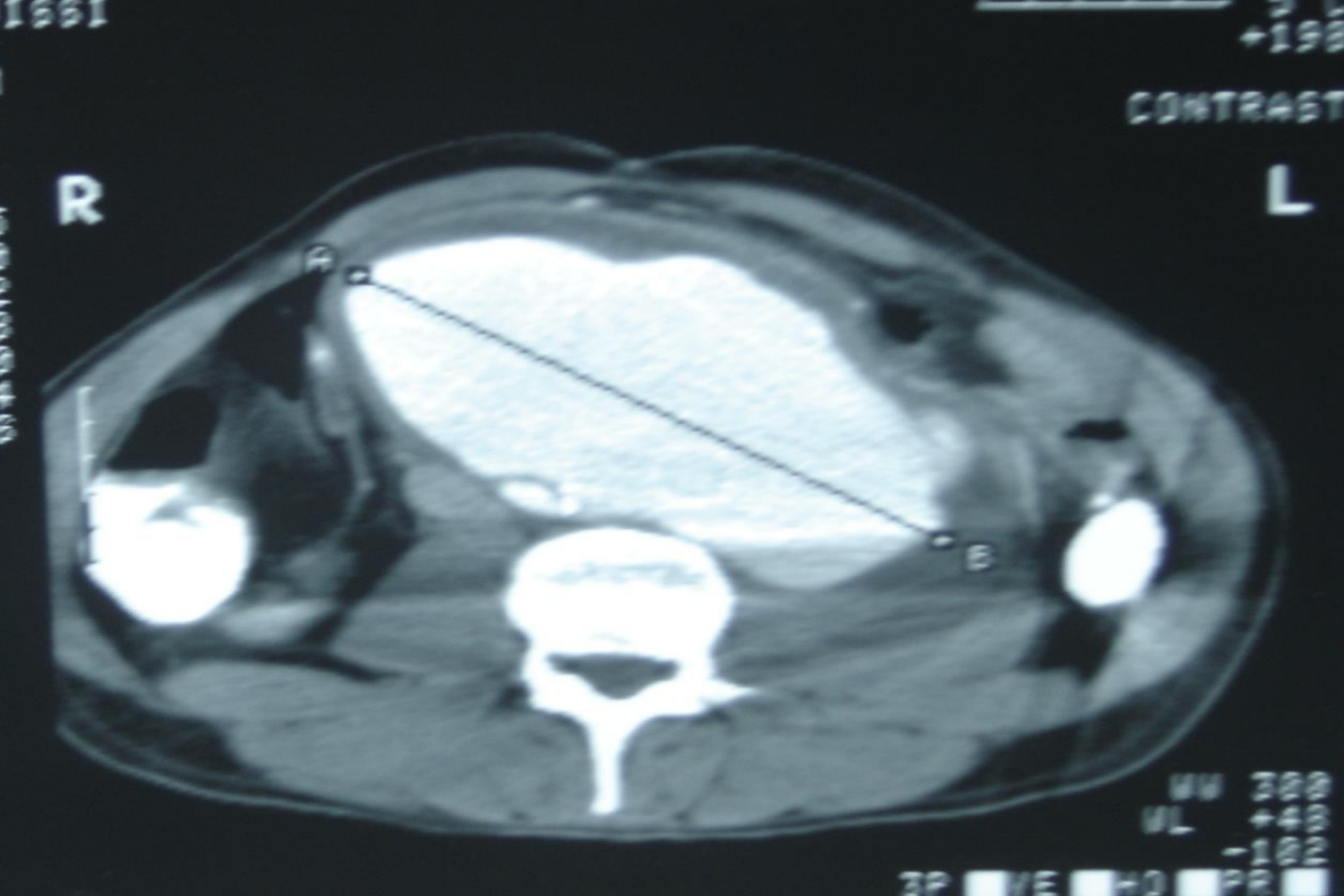

942
432
-78

Personal Study


1
2
3
4
5
6

Case 1

- M 46 y.
- Diagnosis BD : 10 y.
- Abdominal pain - Fievre.
- CRP 250.


R
L
A
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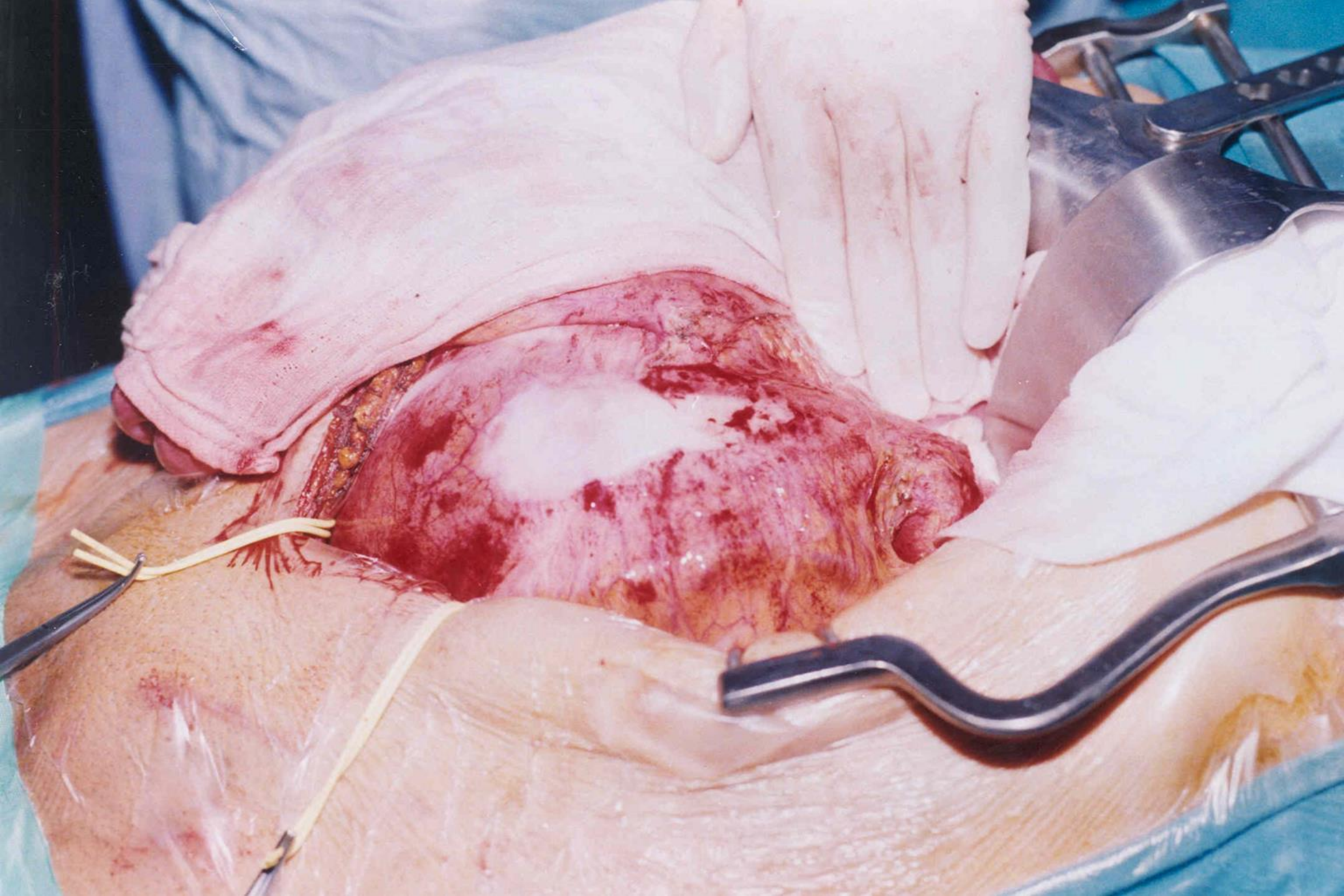
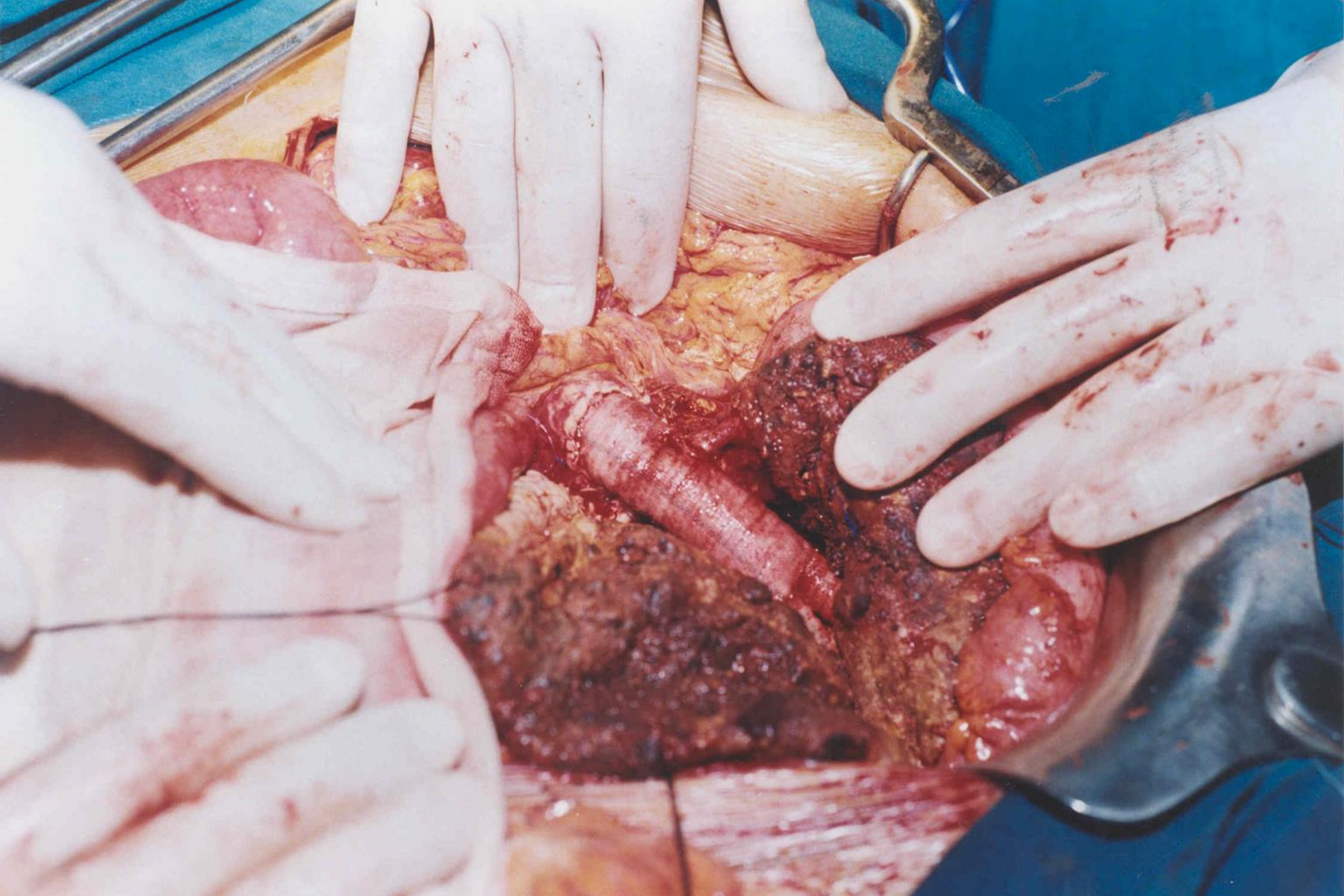
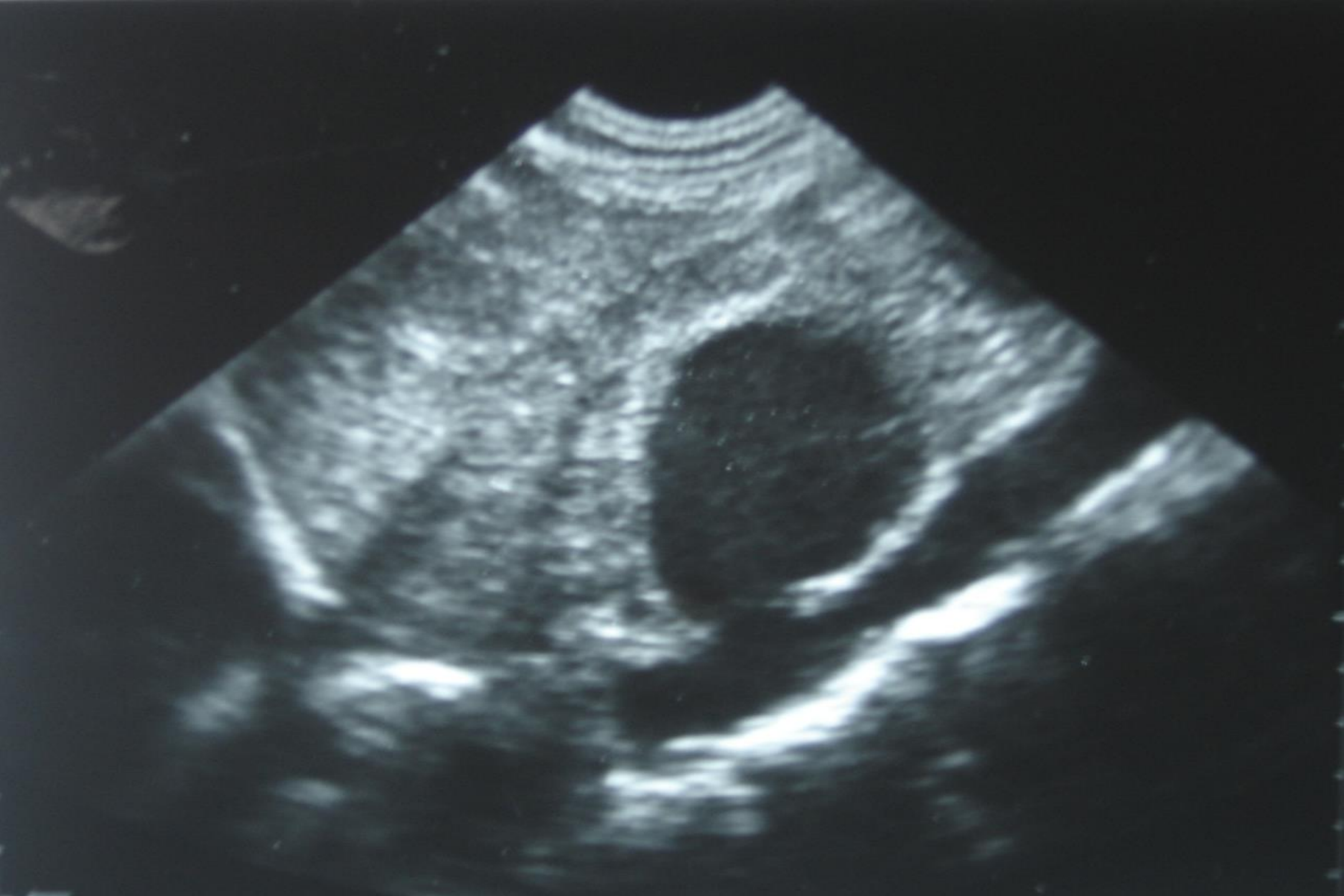
CONTRAST

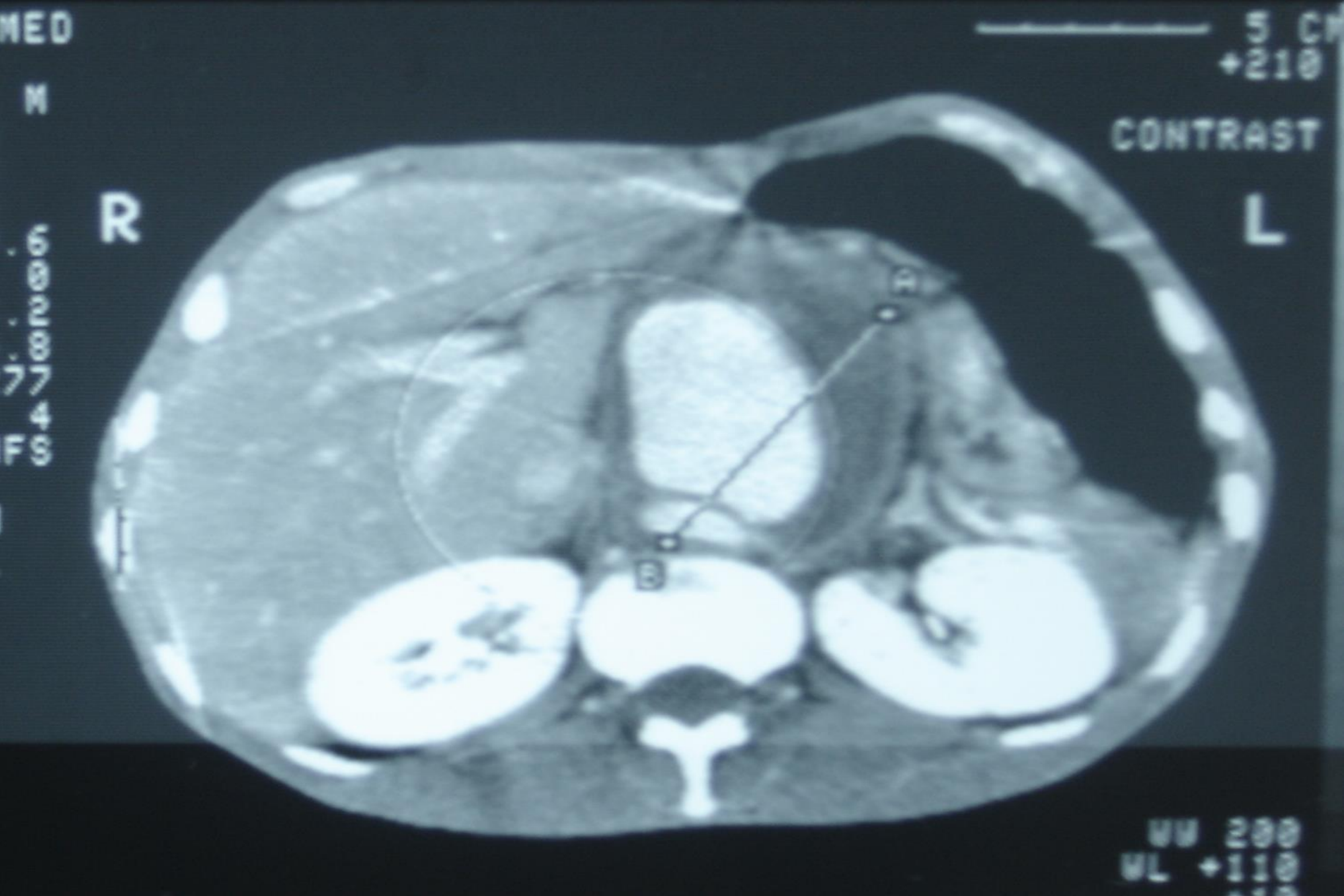


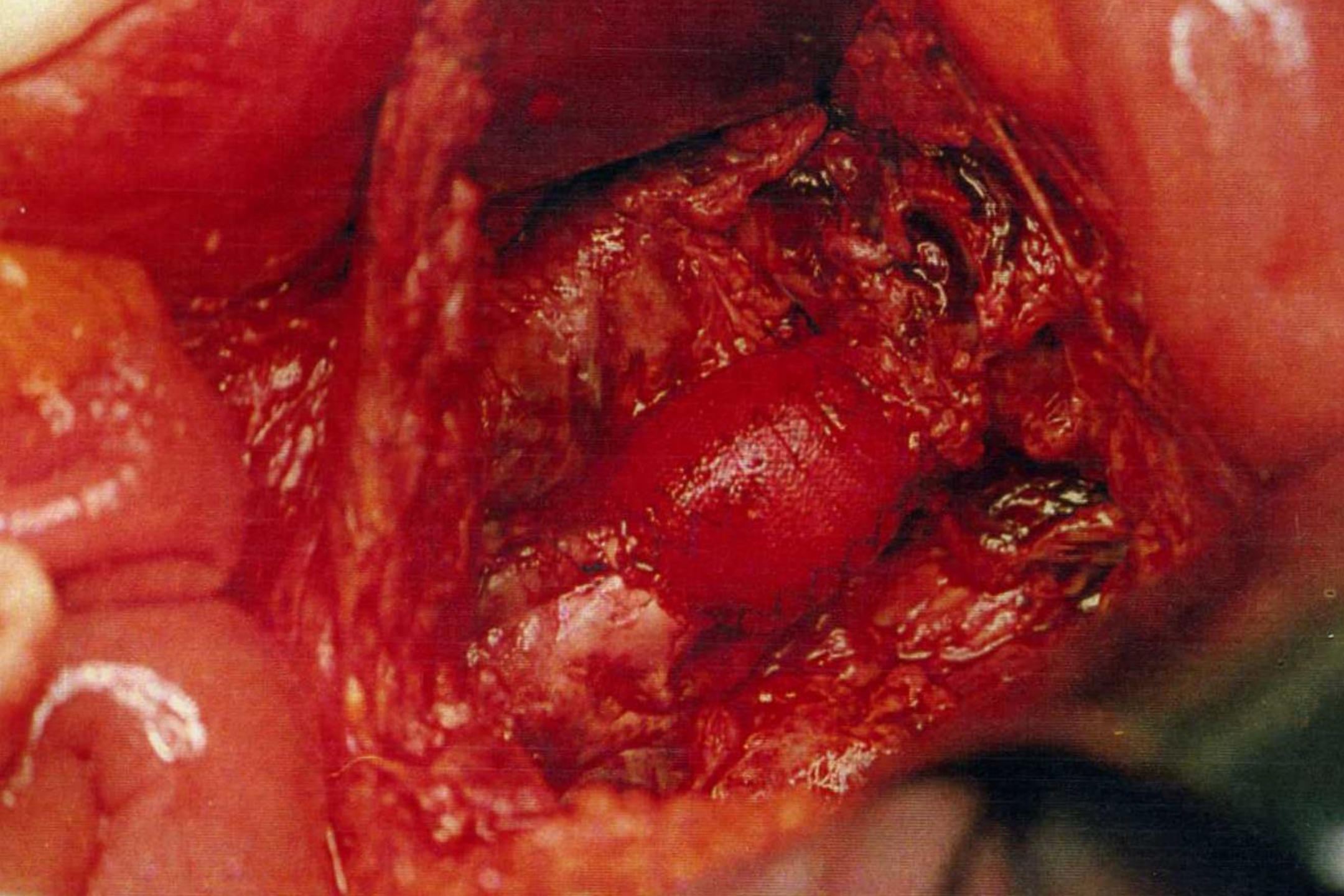


Case 2

- M 53 y.
- Diagnosis BD : 5 y.
- Abdominal pain .
- CRP 120.



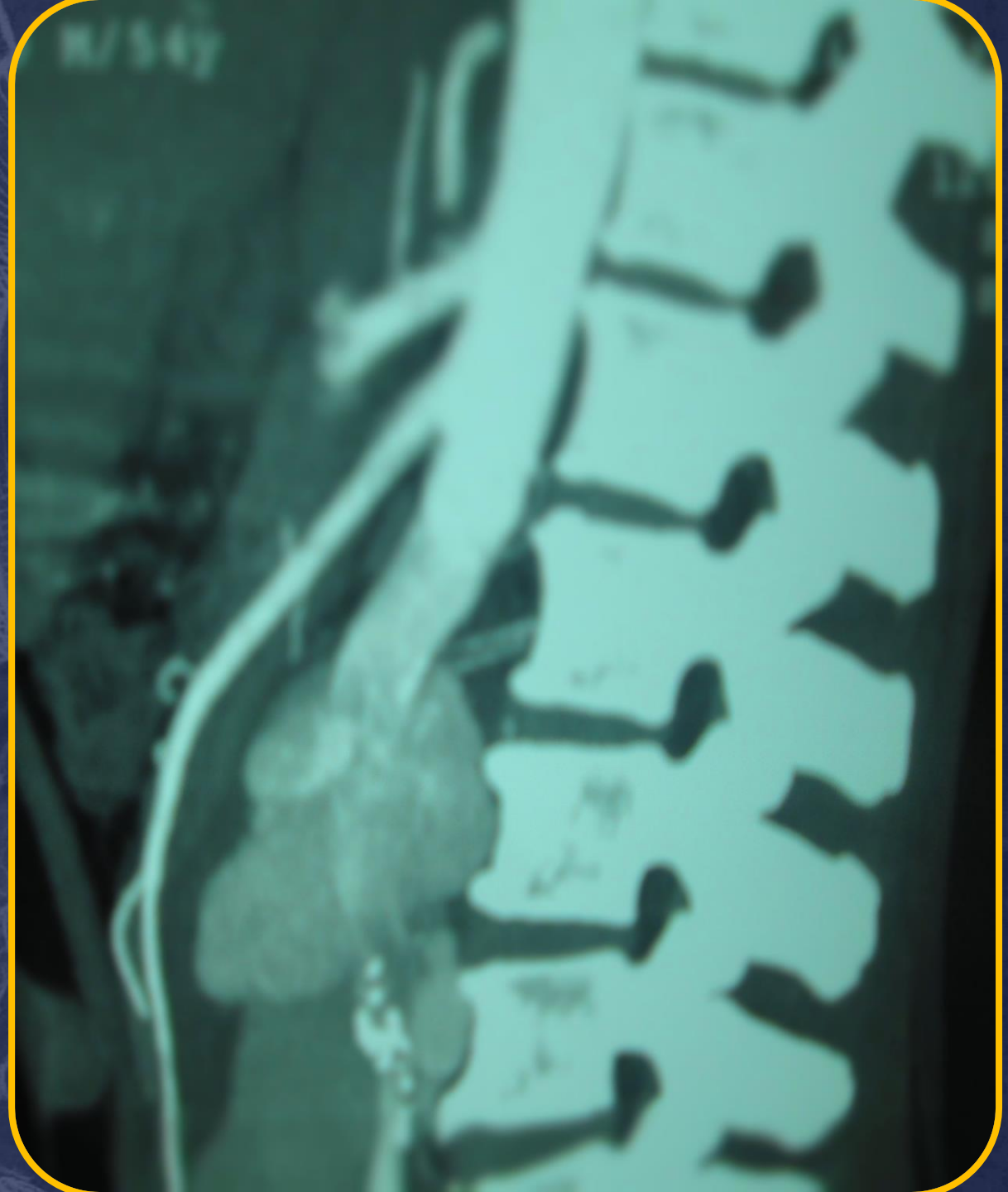

MED
M
R
L
5 CM
+210
CONTRAST
WW 200
WL +110

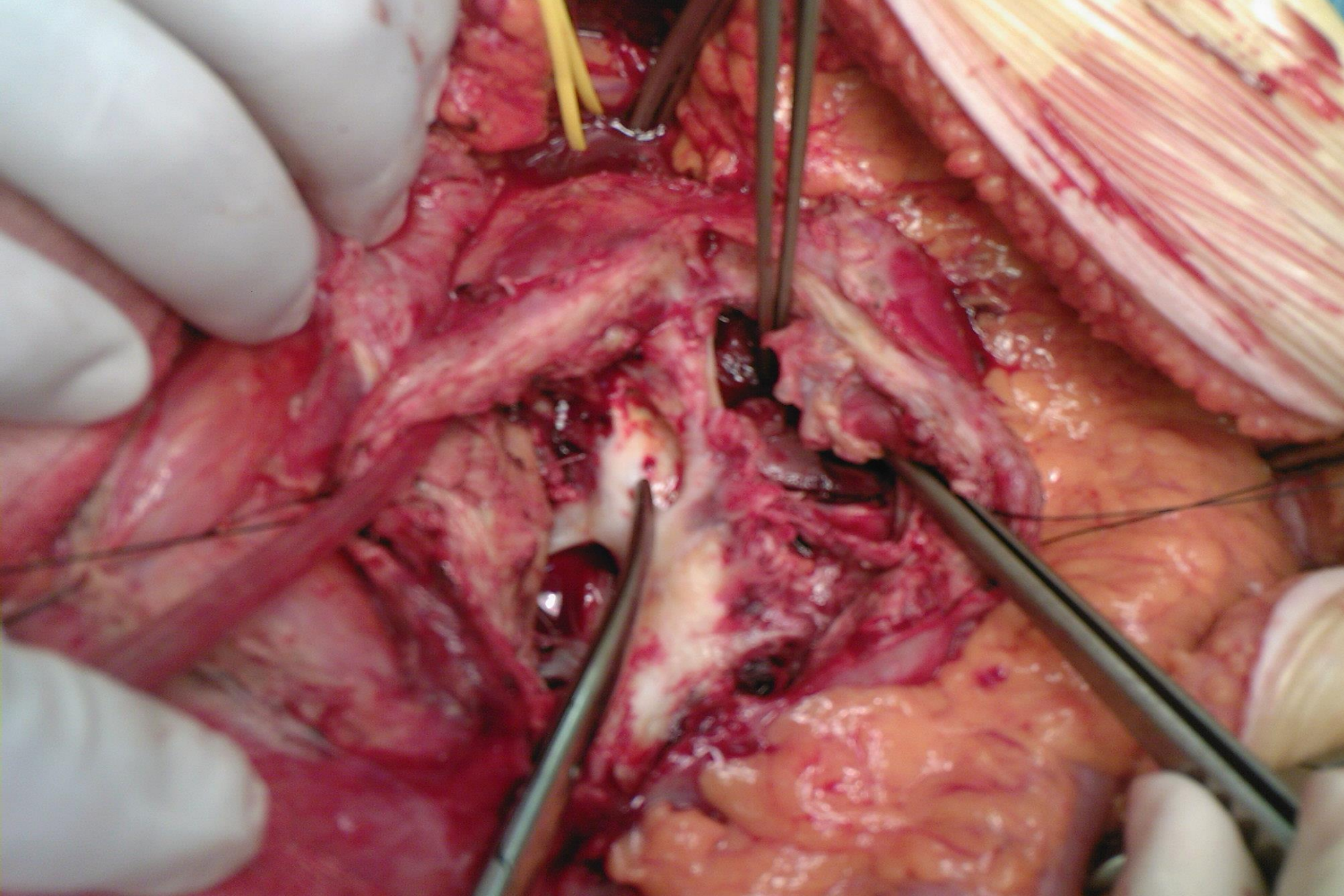


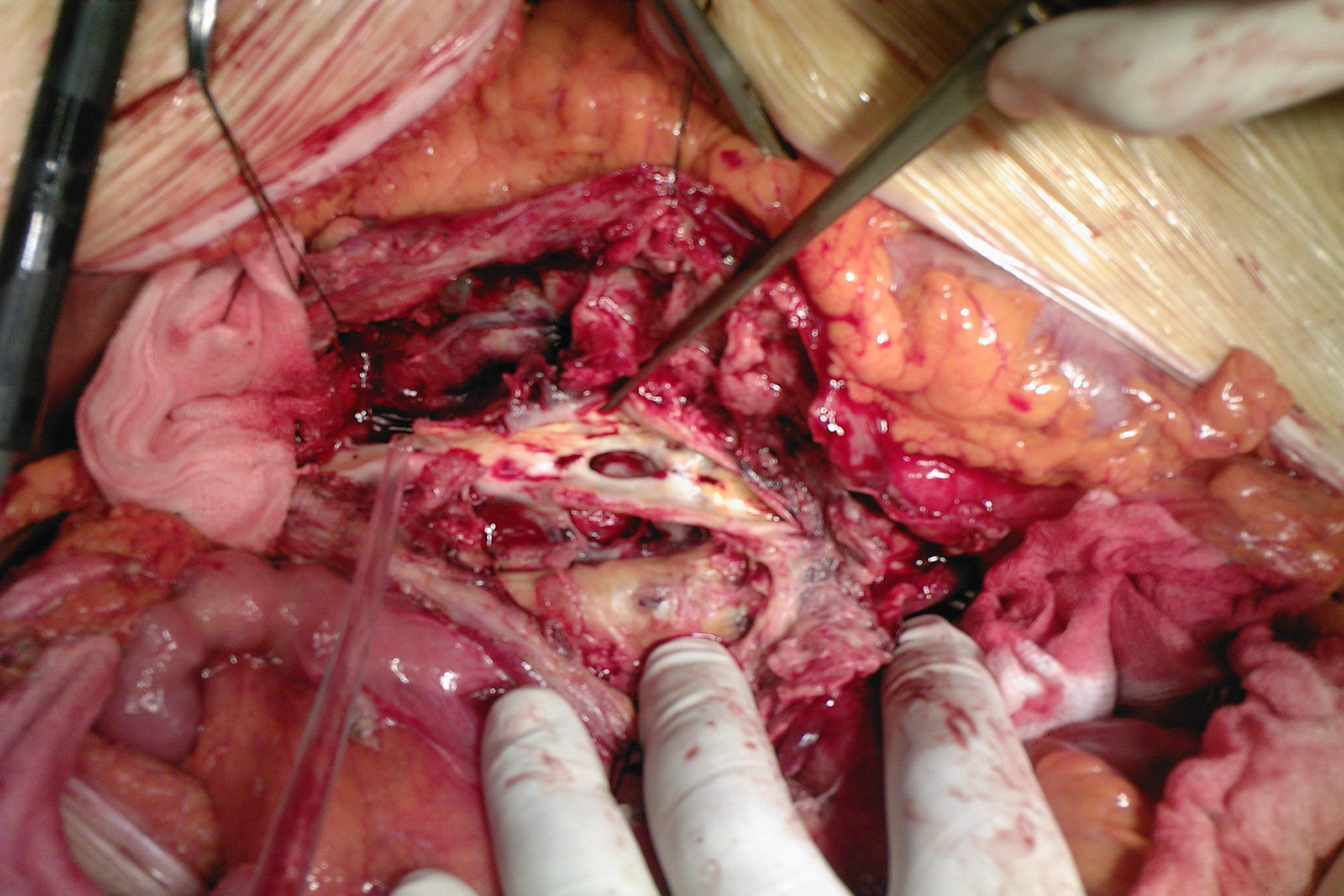

Ex 39886
Se 5 +c
Volume Rendering No cut
DFOV 13.5cm
SOFT
111.1 mm (3D)
57.7 mm (3D)

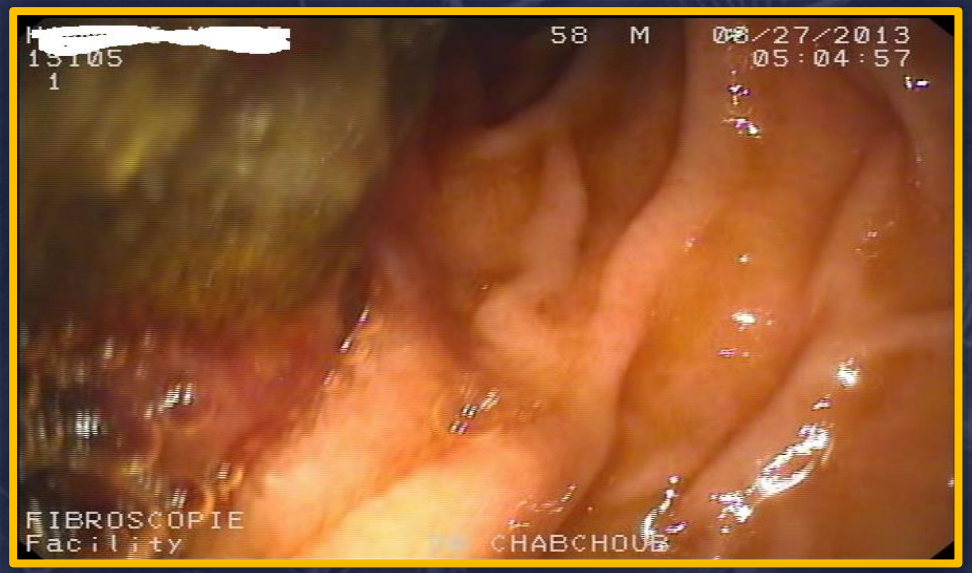
Case 3

- M 54 y.
- Diagnosis BD : 18 y.
- Abdominal pain.
- CRP 150.





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58 M 08/27/2013
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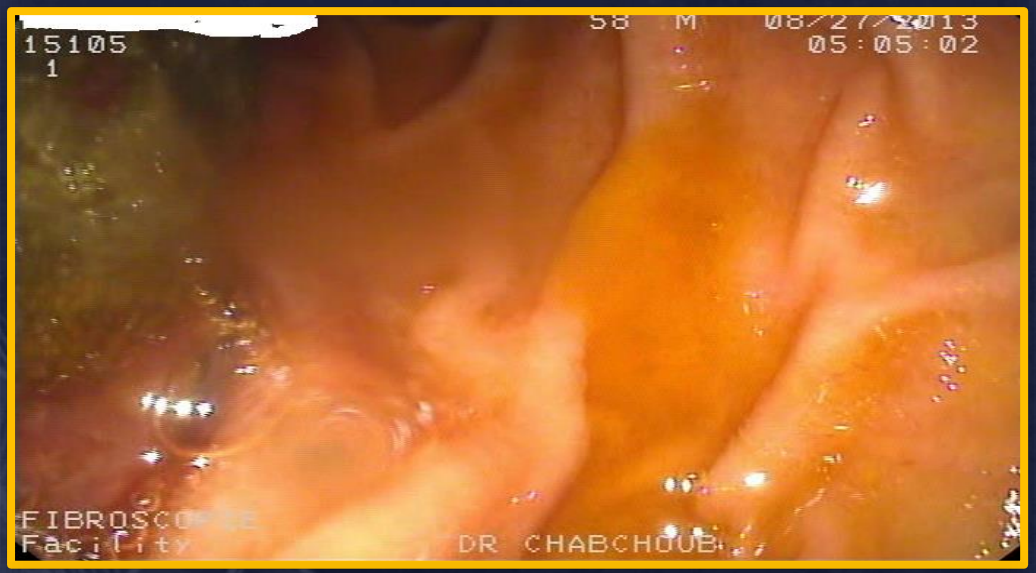
Facility
DR CHABCHOUB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58 M 08/27/2013
05:05:02
FIBROSCOPIE
Facility
DR CHABCHOUB


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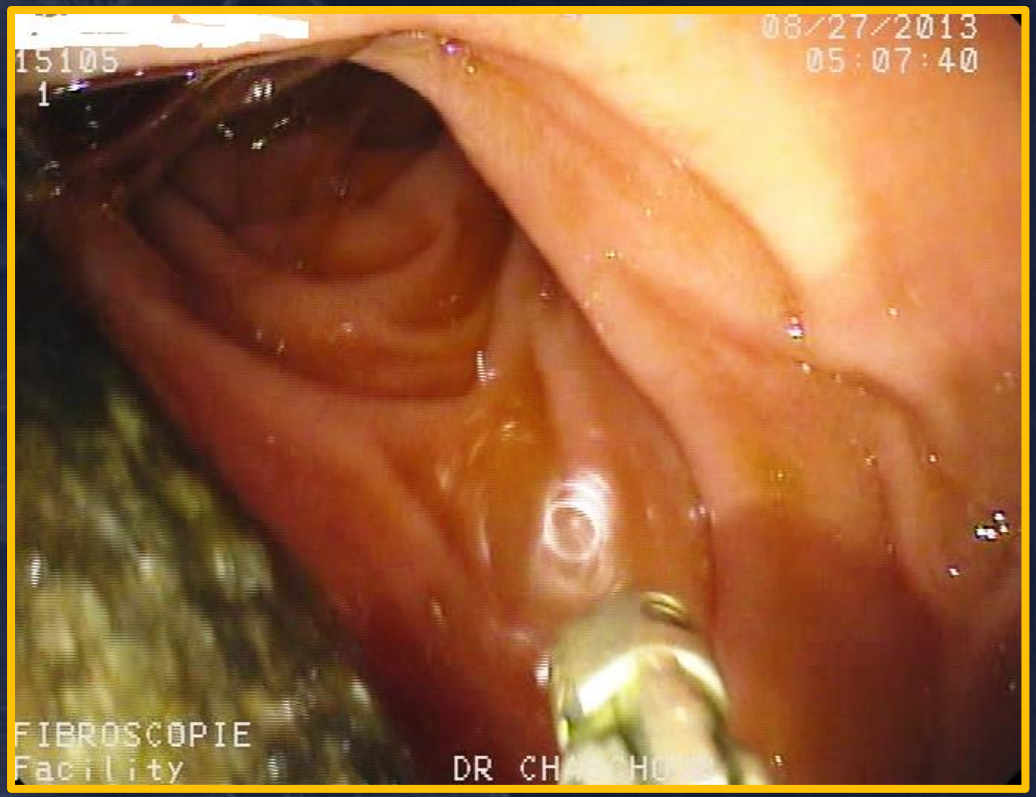
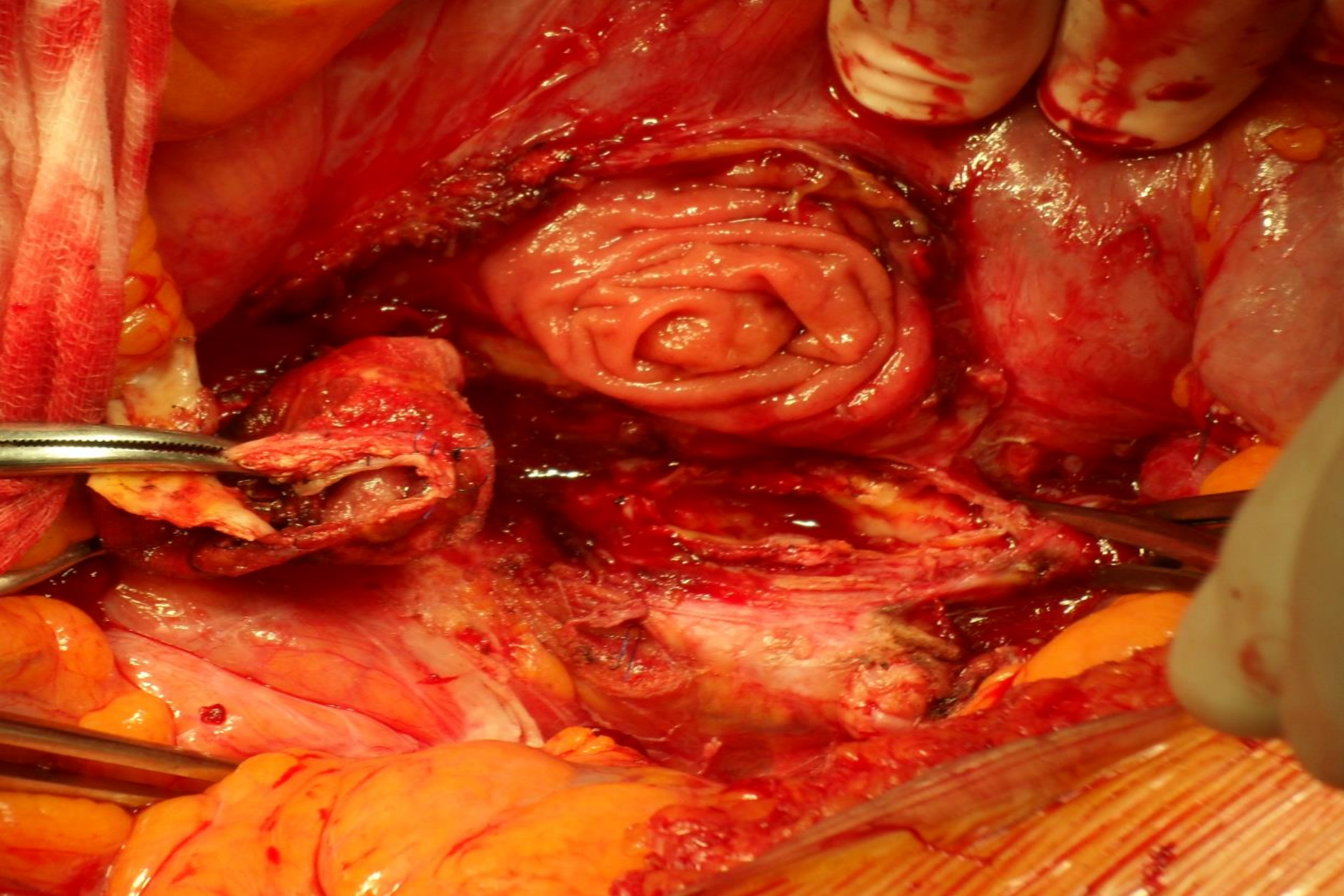
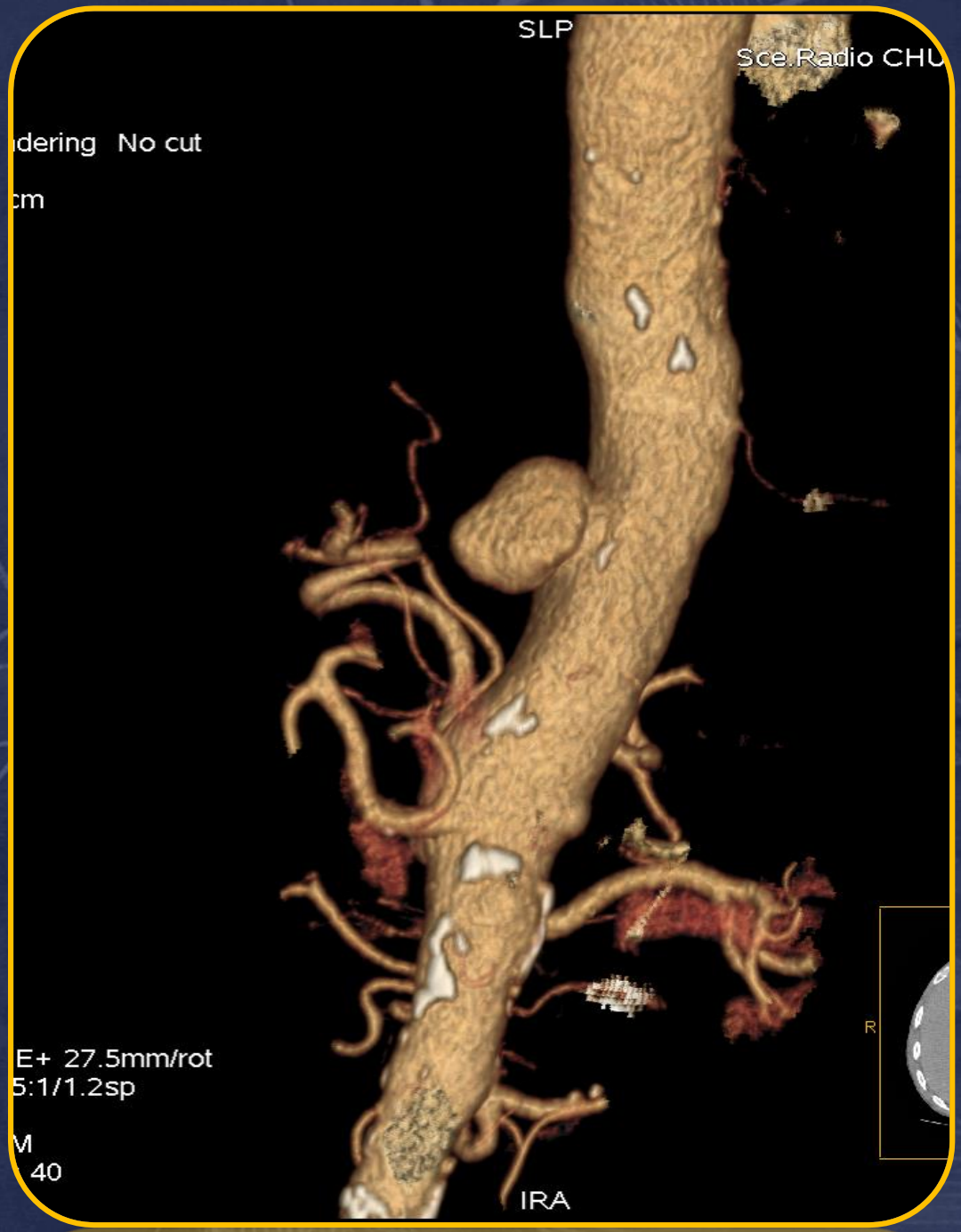
58 M 08/27/2013
05:06:30
FIBROSCOPIE
Facility
DR CHABCHOUB

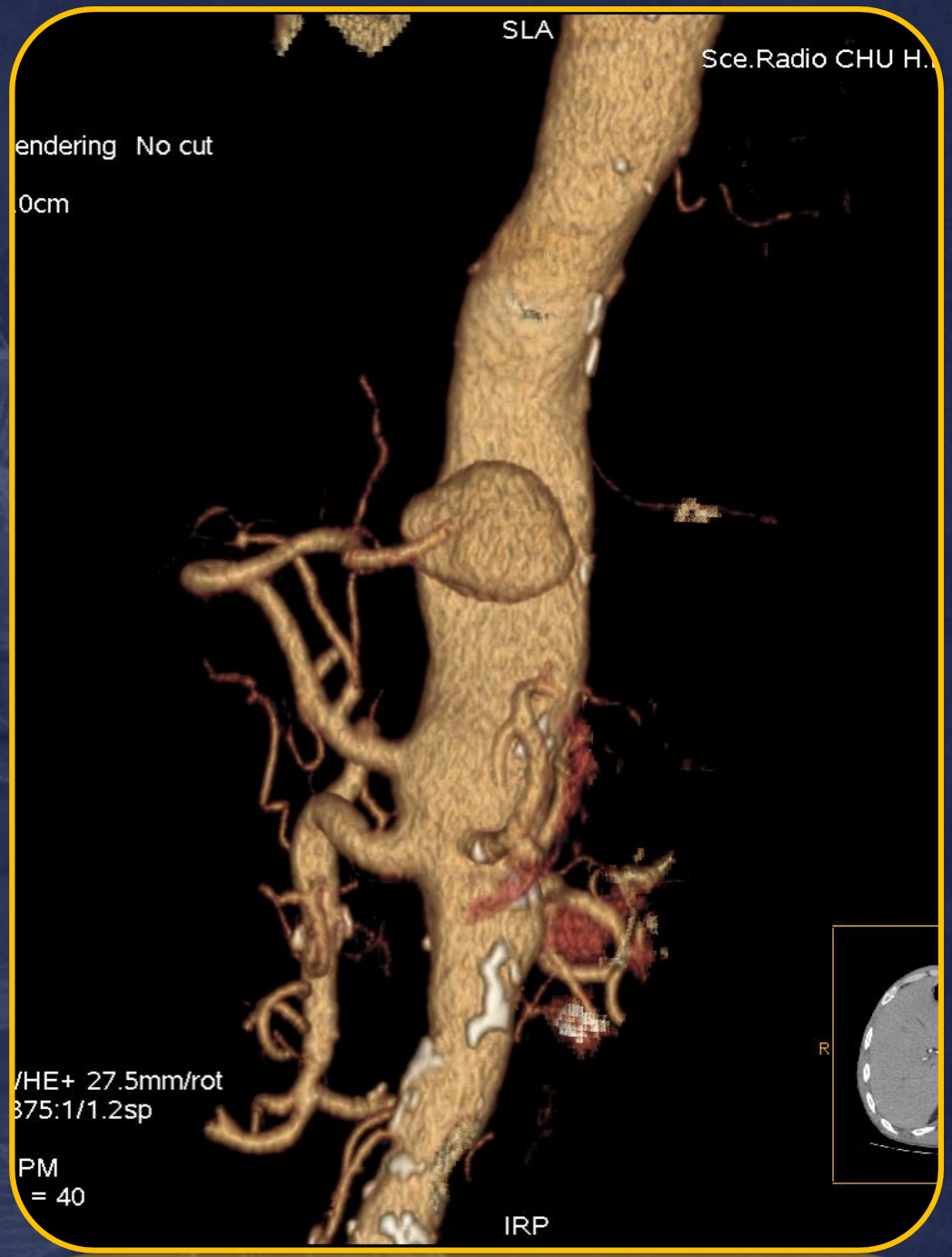

15105
1
08/27/2013
05:07:40
FIBROSCOPIE
Facility
DR CHABCHOUB

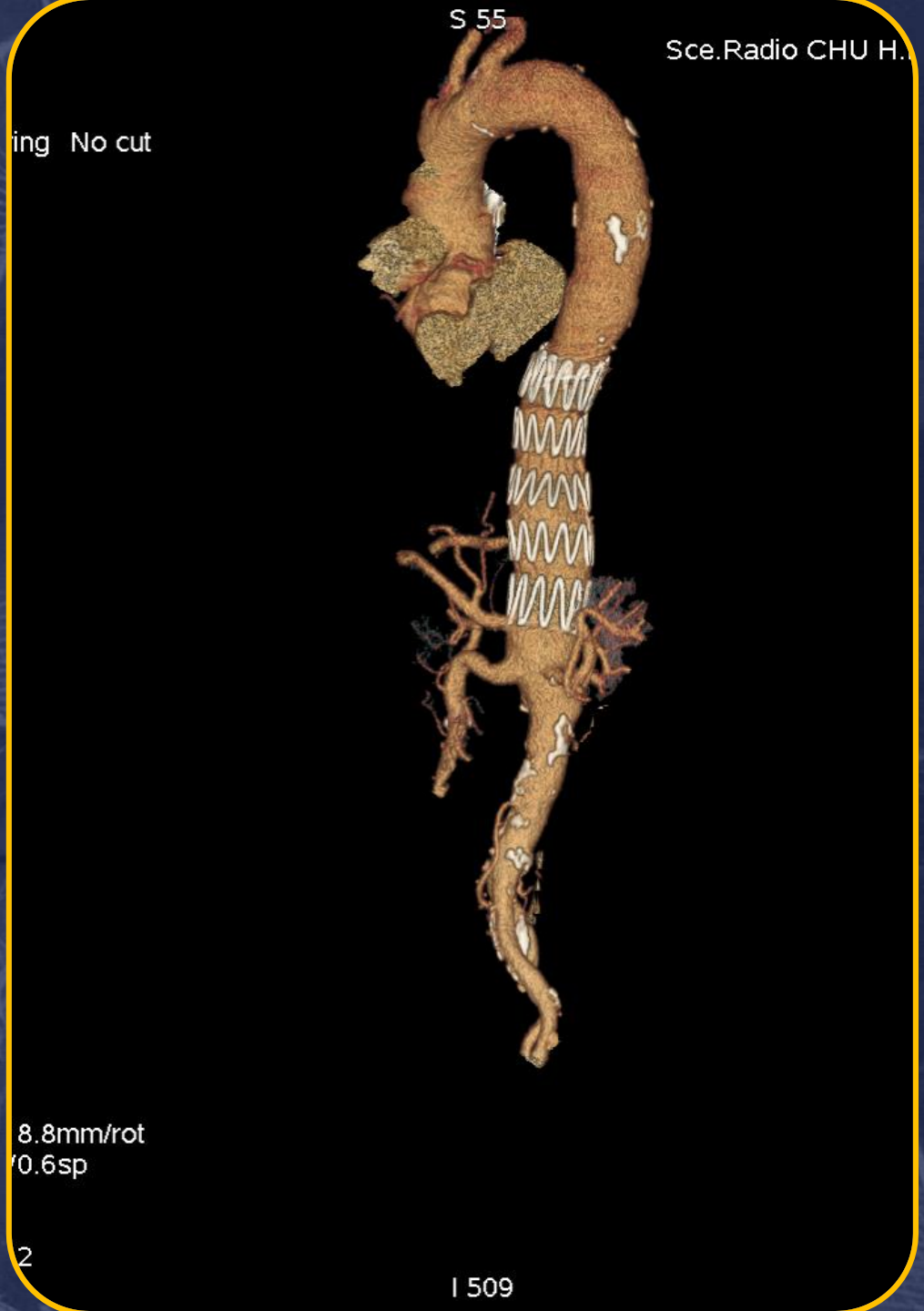


Case 4

- M 38 y.
- Diagnosis BD : 7y.
- Fortuitous.
- CRP 50.

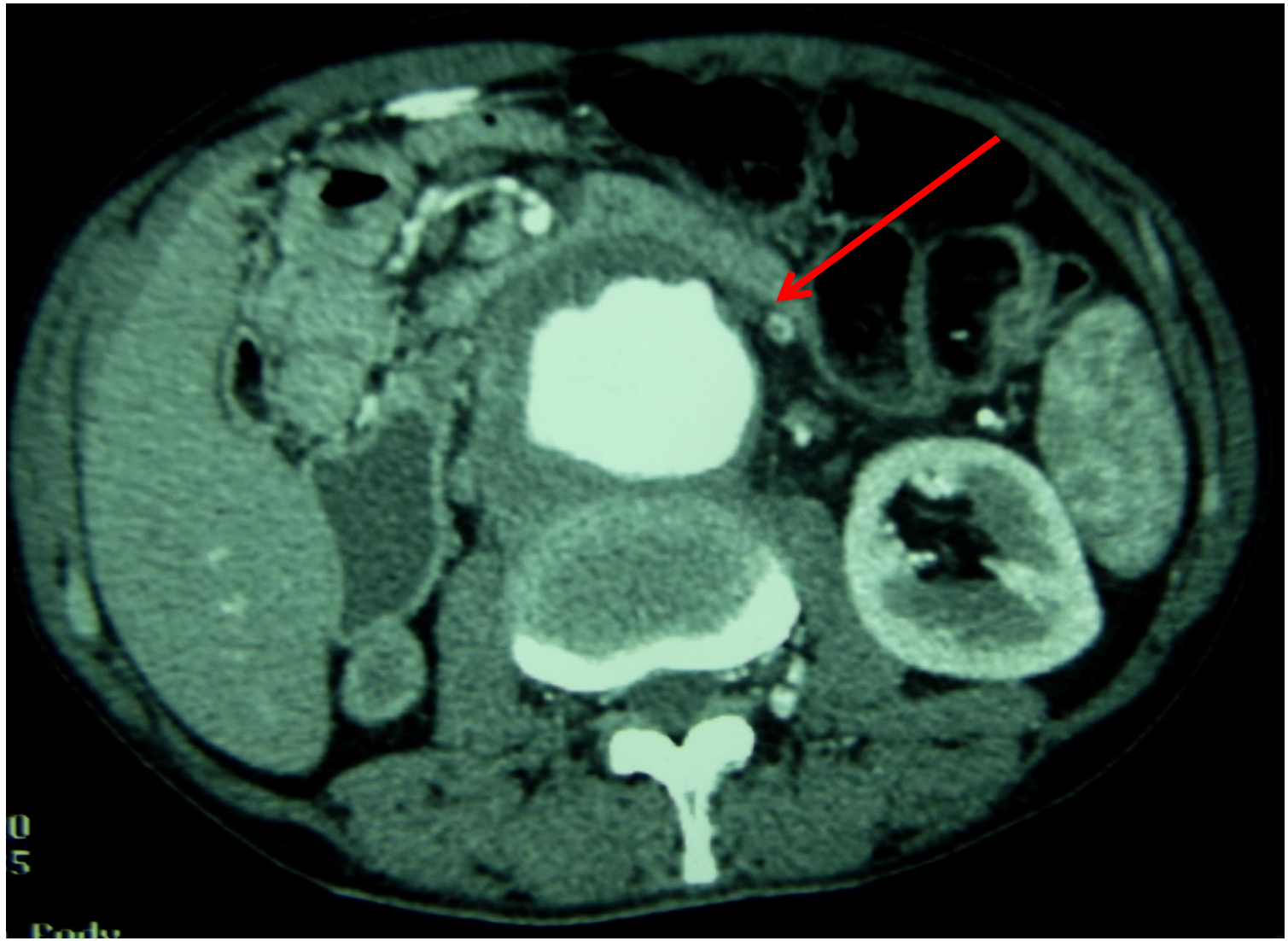

SLP
Sce.Radio CHU
dering No cut
cm
E+ 27.5mm/rot
5:1/1.2sp
M
40
IRA


SLA
Sce.Radio CHU H.
endering No cut
0cm
/HE+ 27.5mm/rot
375:1/1.2sp
PM
= 40
IRP


S 55
Sce.Radio CHU H.
ing No cut
8.8mm/rot
/0.6sp
2
I 509

Case 5

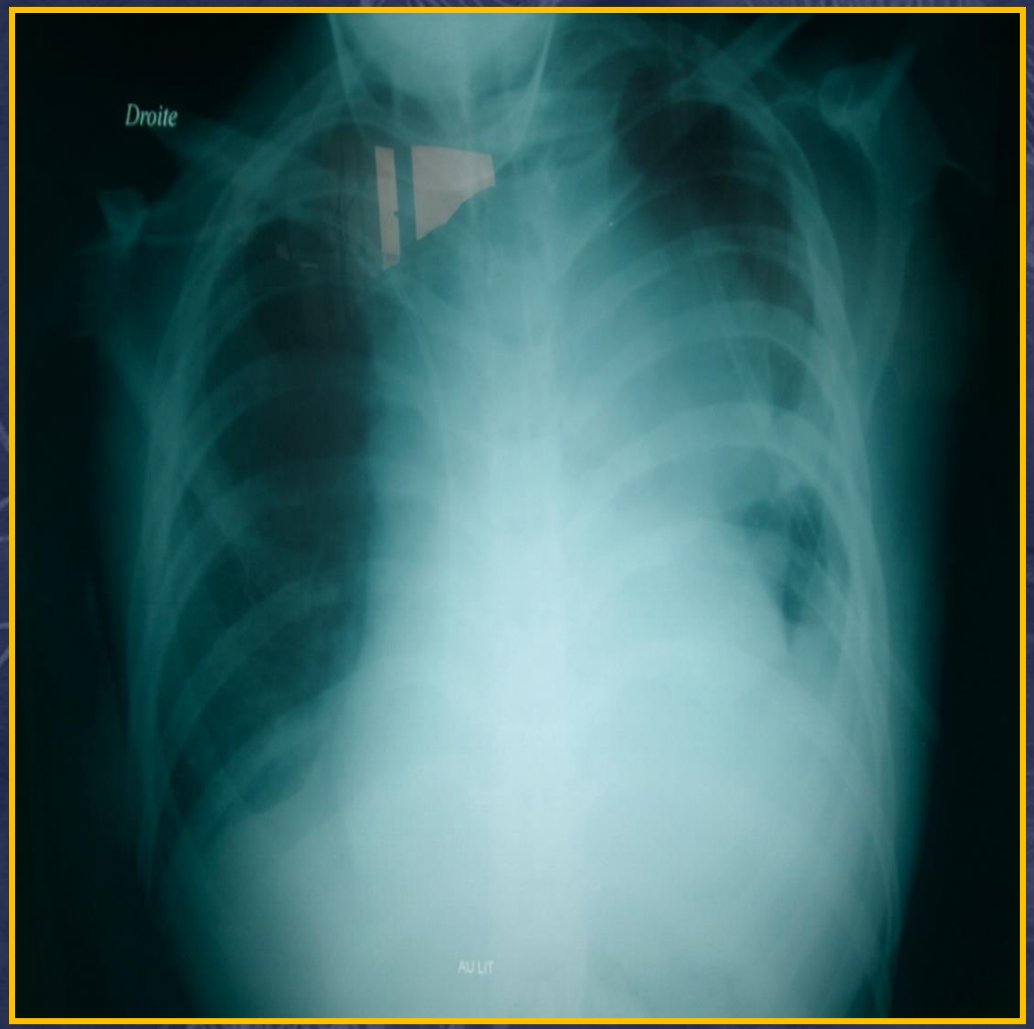
- M 47y.
- Diagnosis BD : 17y.
- Abdominal pain .
- CRP 90.

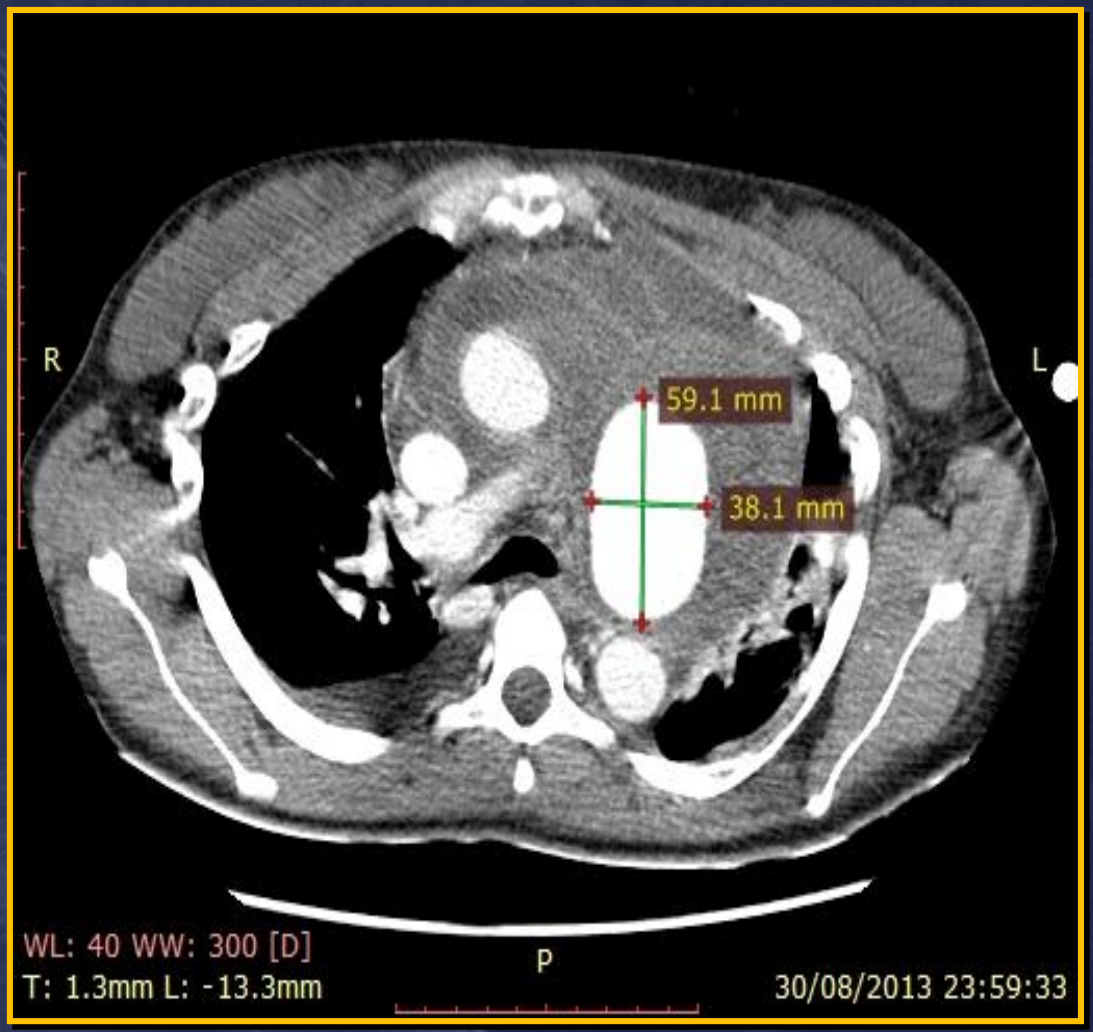

0
5

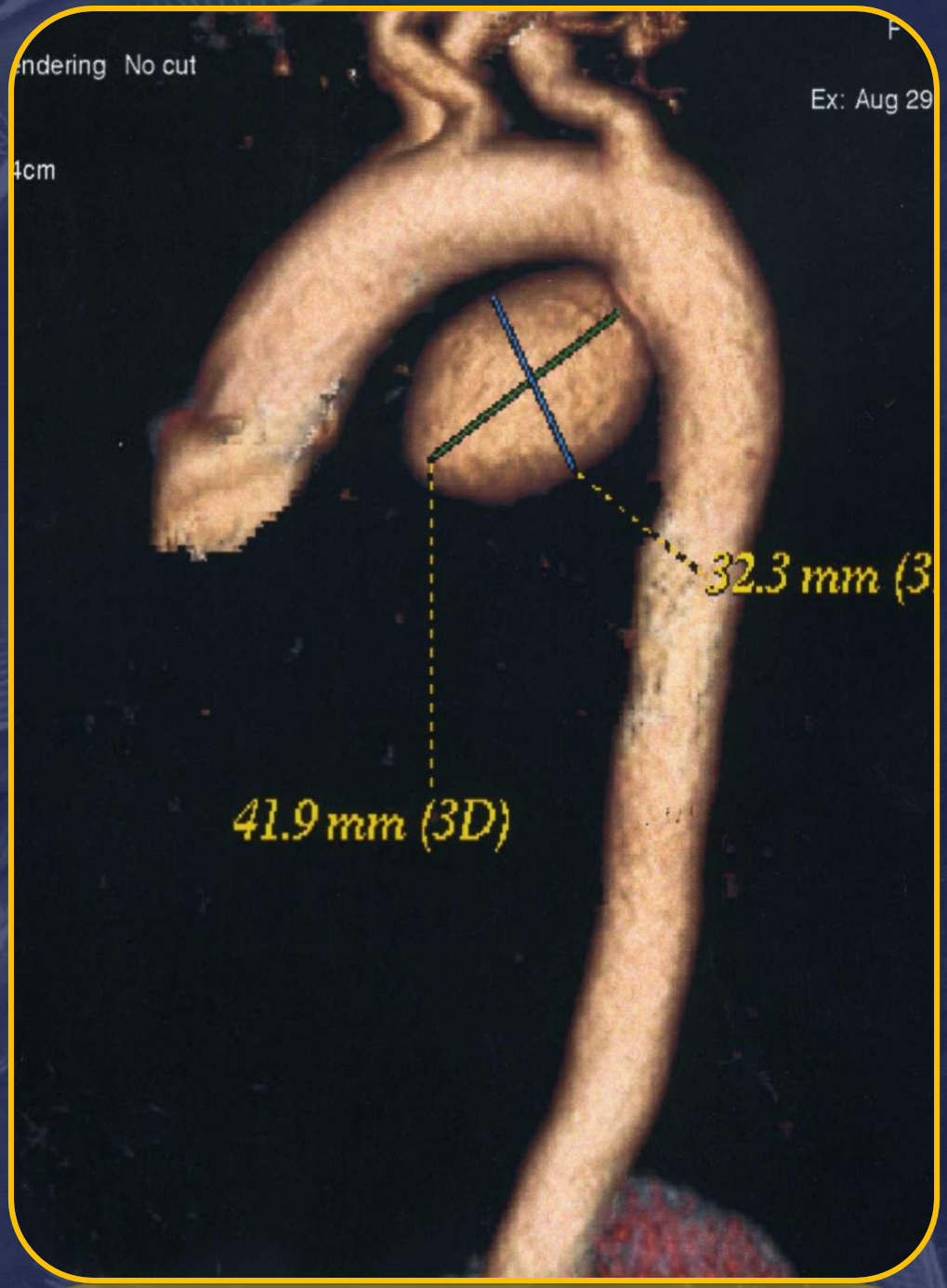
Case 6

- W 27y.
- Diagnosis BD : 0 y .
- Thoracic pain-Choc .
- CRP 237.

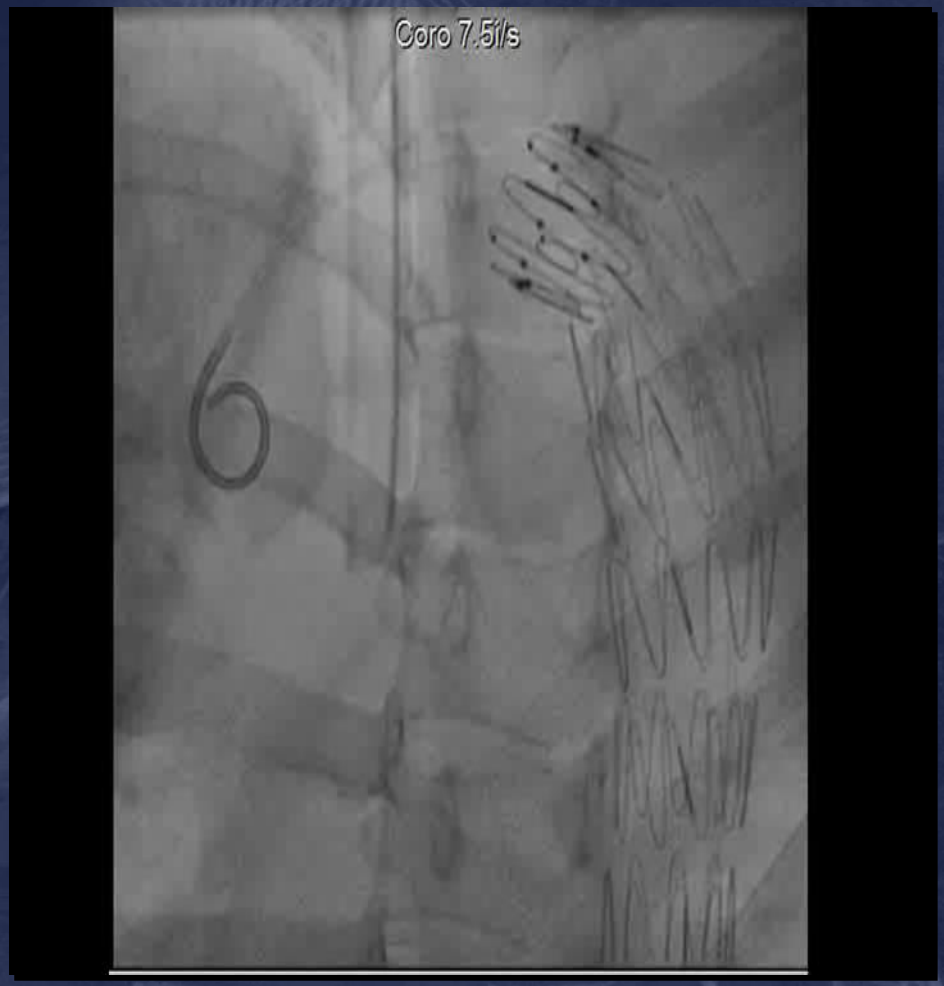

CENTRE DIMAGERIE ME... - OPE - Adult Heart TMSE
9:32:15 AM
73.4
73.4
cm/s
0
5
10
15
T
MI (1.2)
2DG
100
DR
60
CG
33
PRF
4.2k
Filter
6
5S2
T3.6
CF 2.2
21 fps
DICOM PRT.
180
IP5

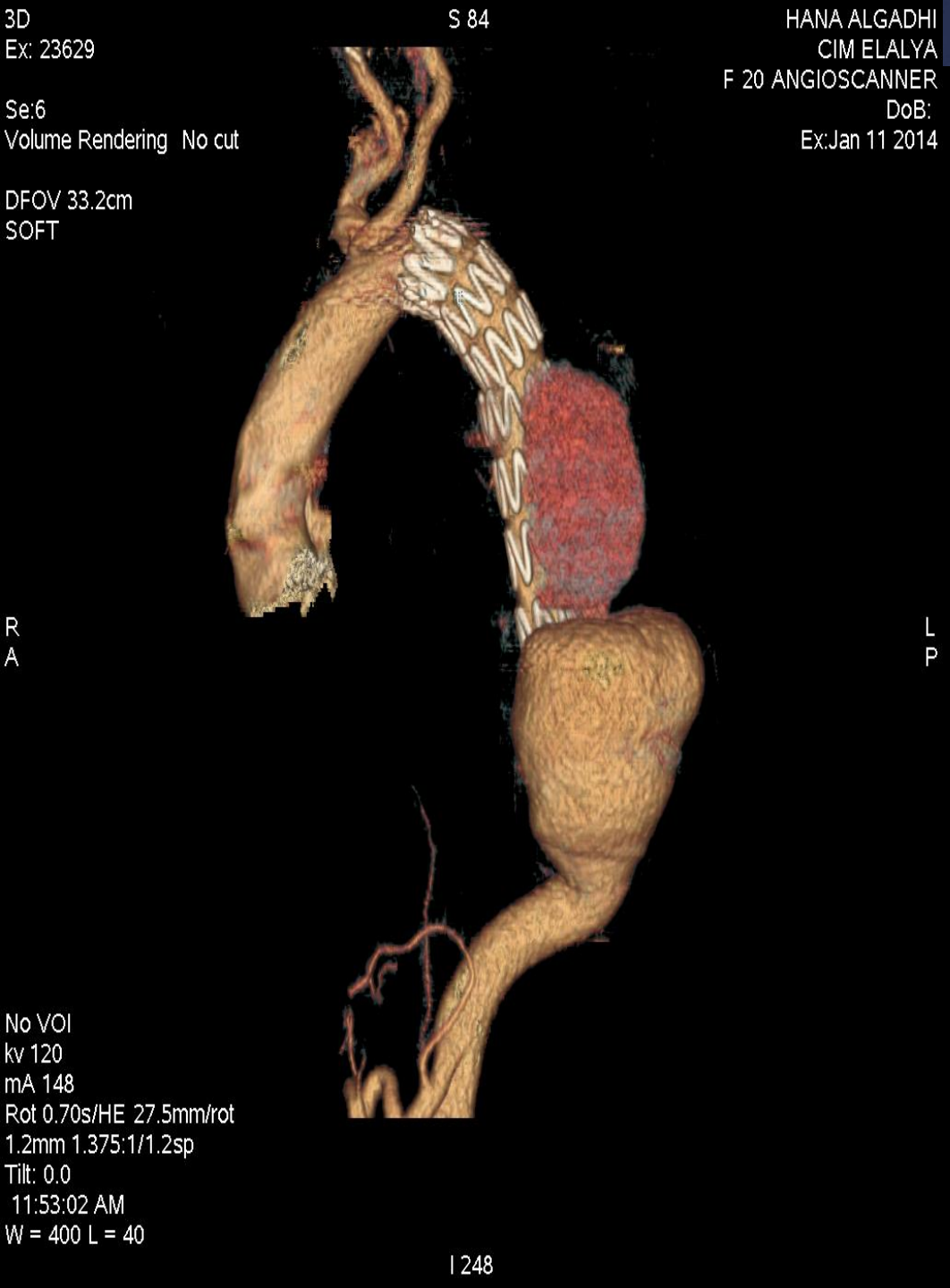

Droite

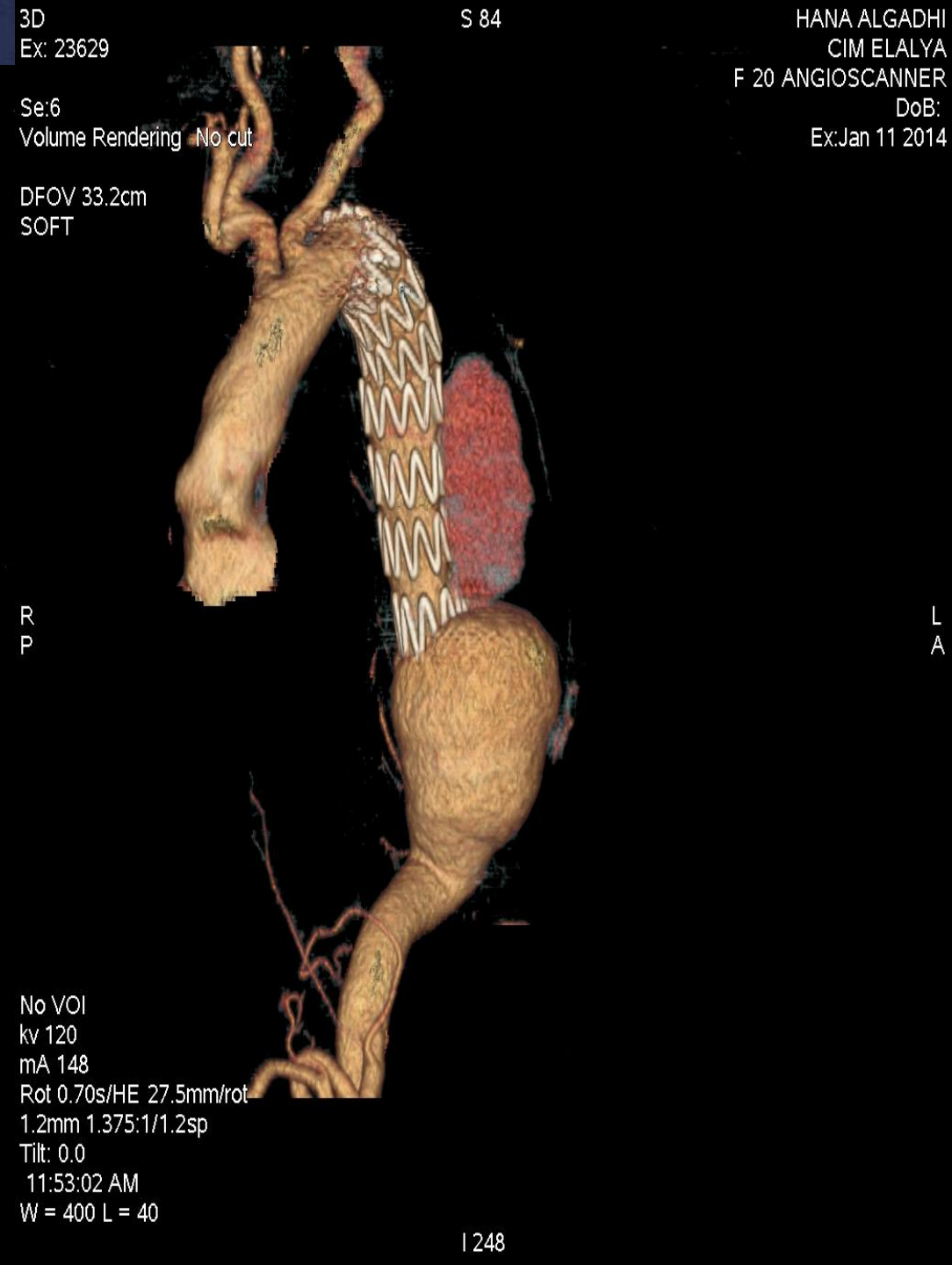

R
L
59.1 mm
38.1 mm
P
WL: 40 WW: 300 [D]
T: 1.3mm L: -13.3mm
30/08/2013 23:59:33


ndering No cut
Ex: Aug 29
4cm
32.3 mm (3
41.9 mm (3D)

Technique




3D
S 84
Ex: 23629
Se:6
Volume Rendering No cut
DFOV 33.2cm
SOFT
HANA ALGADHI
CIM ELALYA
F 20 ANGIOSCANNER
DoB:
Ex:Jan 11 2014
R
A
L
P
No VOI
kv 120
mA 148
Rot 0.70s/HE 27.5mm/rot
1.2mm 1.375:1/1.2sp
Tilt: 0.0
11:53:02 AM
W = 400 L = 40
I 248


3D
S 84
Ex: 23629
Se:6
Volume Rendering No cut
DFOV 33.2cm
SOFT
HANA ALGADHI
CIM ELALYA
F 20 ANGIOSCANNER
DoB:
Ex:Jan 11 2014
R
P
L
A
No VOI
kv 120
mA 148
Rot 0.70s/HE 27.5mm/rot
1.2mm 1.375:1/1.2sp
Tilt: 0.0
11:53:02 AM
W = 400 L = 40
I 248

Conclusion

- Rare.
- Aortitis serious.
- Surgery difficult.
- Endovascular +++
- Recurrence .
- Medical treatment .

Thank You