TOR VERGATA
UNIVERSITY OF ROME

70TH ESCVS CONGRESS
& 7TH IMAD MEETING
20 I 23 JUNE 2022

THE HUGHES-STOVIN SYNDROME: WHAT'S BEYOND PULMONARY ARTERY ANEURYSM.

Ascoli Marchetti A, Oddi FM, De Giorgi A, Ippoliti A.

Department of Vascular Surgery
Tor Vergata University, Rome

Friday October 5

07.30 Welcoming coffee

EPIDEMIOLOGY OF AAA
Frank A. Lederle

08.00 Aortic aneurysms: changes in epidemiology vs. changes in clinical practice,
Janet Powell

08.15 Gender differences in AAA,
Rebecka Hultgren

08.30 Pattern and associations of AAA enlargement,
Frank A. Lederle

08.45 What can we learn from the non-aneurysmal infrarenal aortic diameter?
Paul Norman

09.00 Talks selected from submitted abstracts

09.00 Increased mortality in subjects with small Abdominal Aortic Aneurysm (AAA) without previous history of cardiovascular disease compared to controls is associated with increased levels of hs-CRP and H-FABP,
Sehrabi Sarsouh

09.05 The prognosis of ruptured abdominal aortic aneurysms in Denmark 1994-2008,
Jes Lindholt

09.10 A comparative study of abdominal aortic aneurysm (AAA) in men and women,
Sehrabi Sarsouh

09.15 Procedures for ruptured abdominal aortic aneurysms decrease among 65-74 year old men in Sweden – but surprisingly not among the youngest,
Fredrik Lundgren

09.20 Morphological scanographic analysis of infrarenal aortic aneurysms: is there any predictive rupture factor except the maximal tranverse diameter?,
Jordane Herail

09.25 Discussion

09.45 Coffee break

10.15 Satellite symposium (see page 12)

GENETICS AND GENOMICS OF AAA
Helena Kuivaniemi

10.15 The German-Dutch-Australian AAA Expression Consortium,
Jonathan Golledge

10.30 Transcriptional genomics of AAA,
Gerard Tromp

10.45 LRP5 gene polymorphisms and AAA genetic susceptibility,
Betti Giusti

11.00 Discussion

PATHOPHYSIOLOGY AND BIOMARKERS OF AAA
Gillian Cockerill and Jean-Baptiste Michel

11.10 Identification of novel biomarkers of AAA by proteomic analysis,
Jose Luis Martin Ventura

11.25 Involvement of cell death in the pathophysiology of AAA,
Gillian Cockerill

11.40 On the potential increase of the oxidative stress status in patients with AAA,
Joel Pincemail

11.55 Role of signaling pathways in aortic aneurysms,
Koichi Yoshimura

12.10 Diabetes as a protective factor for development of abdominal aortic aneurysm disease,
Joost A. Van Herwaarden

12.25 Talks selected from submitted abstracts

12.25 Mechanisms for fibrinolysis of the intraluminal Thrombus of Abdominal Aortic Aneurysm,
Jesper Swedenborg

12.30 The biomarker mmp9 in open and evar patients treated for abdominal aortic aneurysm: personal experience and evidence from the literature,
Andrea Ascoli Marchetti

12.35 Limiting Perfusion following Intra-renal Abdominal Aortic Aneurysm Inner Version,
Marc A Bailey

12.40 Assessment of biomarkers and predictive models for abdominal aortic aneurysm growth,
Melina Nega de Cérniga

12.45 Increased levels of periostin in abdominal aortic aneurysms,
Osamu Yamashita

12.50 Study of the histological and biomechanical properties of fragments isolated from the anterior wall of abdominal aortic aneurysms,
Erasmo Simao Da Silva

JMV—Journal de Médecine Vasculaire (2017) 42, 21—28

ORIGINAL ARTICLE

EVAR and OPEN treatment of abdominal aortic aneurysm: What is the role of MMP-9 in the follow-up?☆

Traitement de l'anévrysme de l'aorte abdominale par endoprothèse ou chirurgie ouverte : quel est le rôle de la MMP-9 au cours du suivi ?

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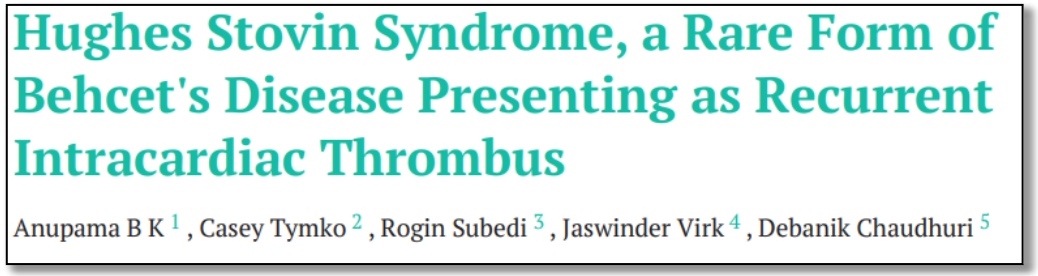
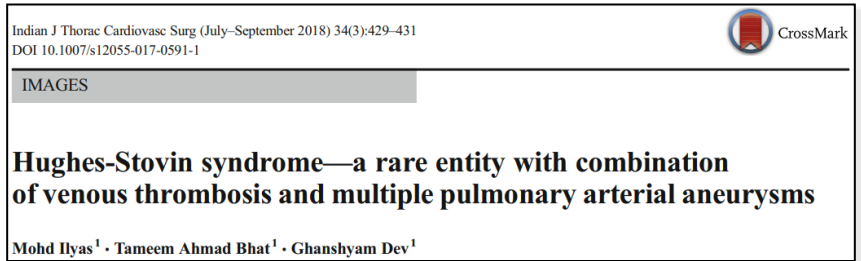
THE HUGHES-STOVIN SYNDROME

Hughes-Stovin syndrome is a rare entity.

The etiology of Hughes-Stovin syndrome is still unknown and the natural course of the illness is usually fatal; however, it is supposed to be a clinical variant manifestation of Behçet disease and it usually affects young men.





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ASSOCIATION WITH BEHCET

Similarities between the Behcet desease and Hughes-Stovin syndrome:

- Gender: mostly young males
- Common clinical manifestation: fever, arthralgia, thrombosis
- Pulmonary artery aneurysm association with thrombosis: HSS 100%, BD 80%
- Common histopathologic findings of PAA: -perivascular inflammation
  -arterial wall destruction
- Treatment of choice: cyclophosphamide, corticosteroids and azathioprine

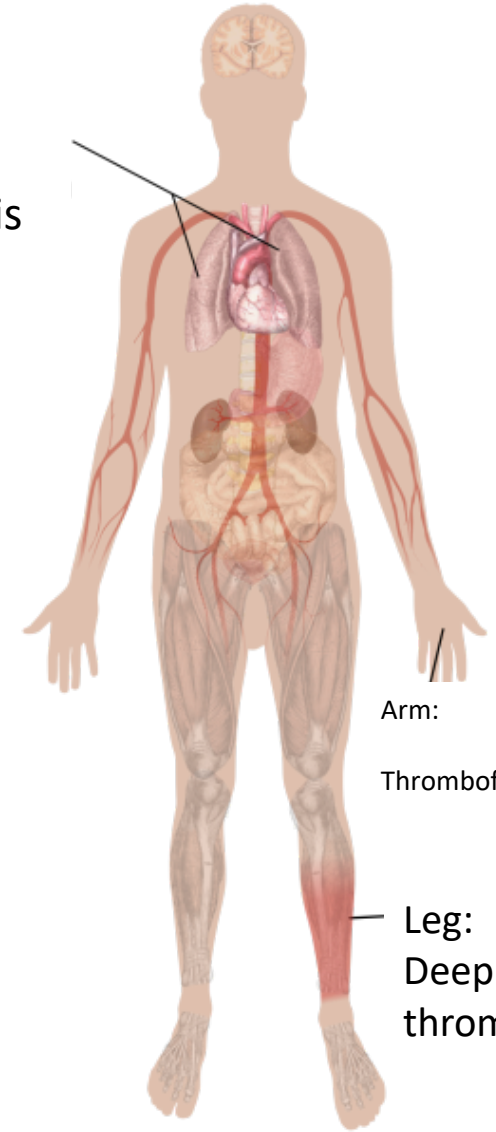
PATHOLOGICAL MANIFESTATIONS

The Hughes-Stovin is characterized by:

Multiple Pulmonary Artery Aneurysms

Peripheral Venous Thrombosis

Lung:
Embolism
Thrombosis
Aneurysm



Arm:

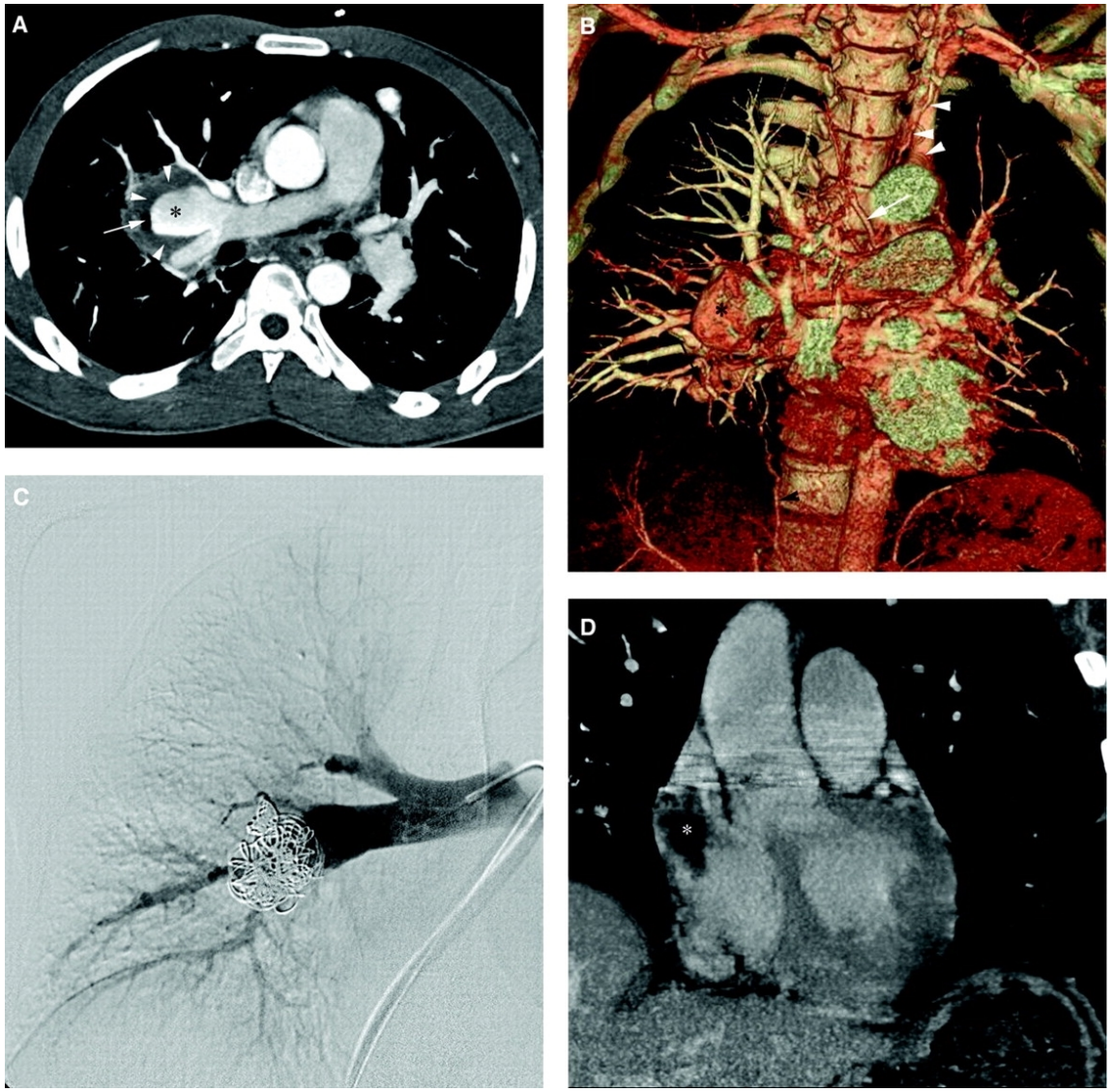
Thromboflebitis

Leg:
Deep venous thrombosis

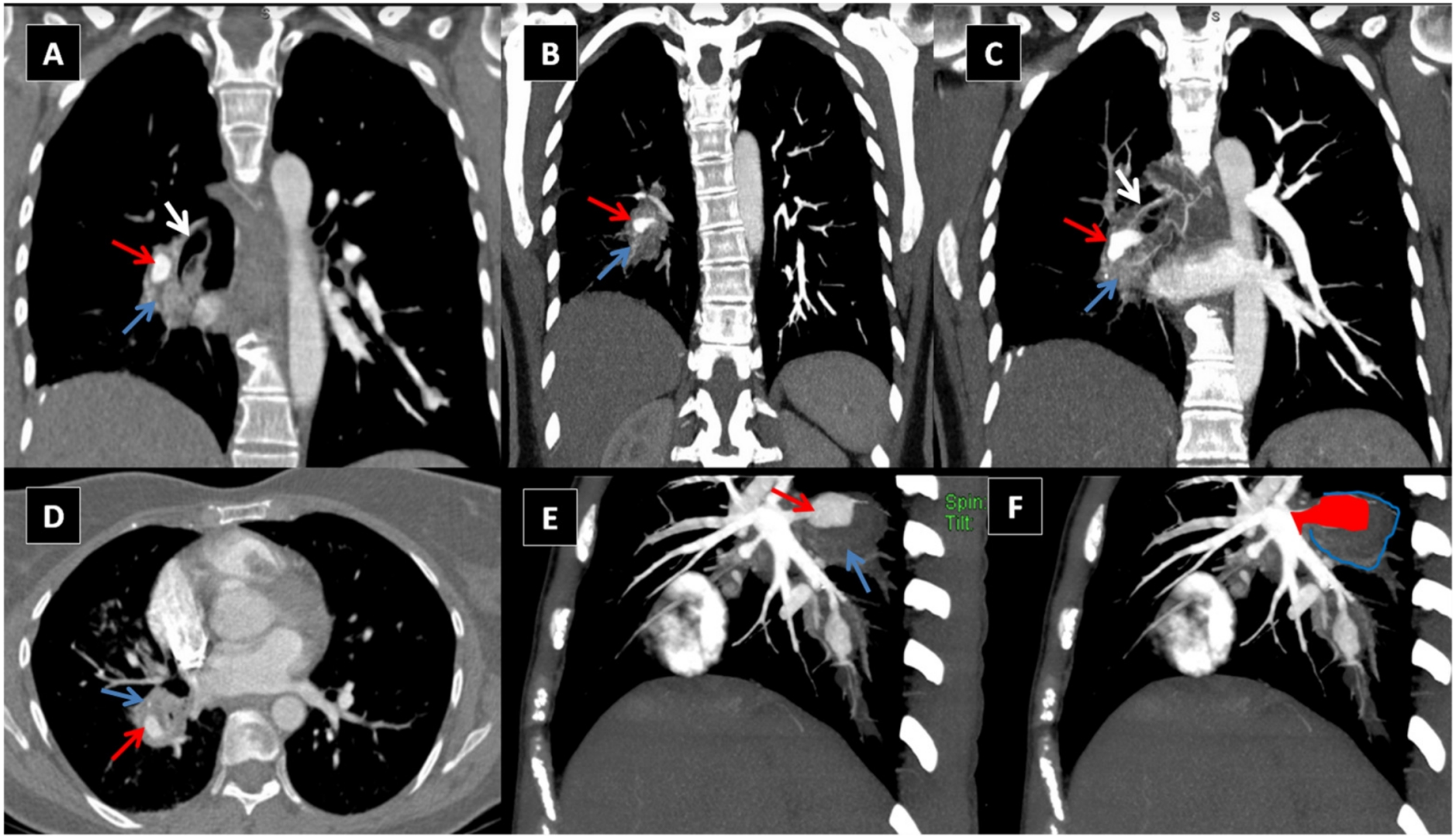
PULMONARY INVOLVEMENT

Typical pulmonary symptoms are:

- recurrent fever
- chills
- coughs
- fulminant haemoptysis
- dyspnea
- chest pain
- signs of pulmonary hypertension



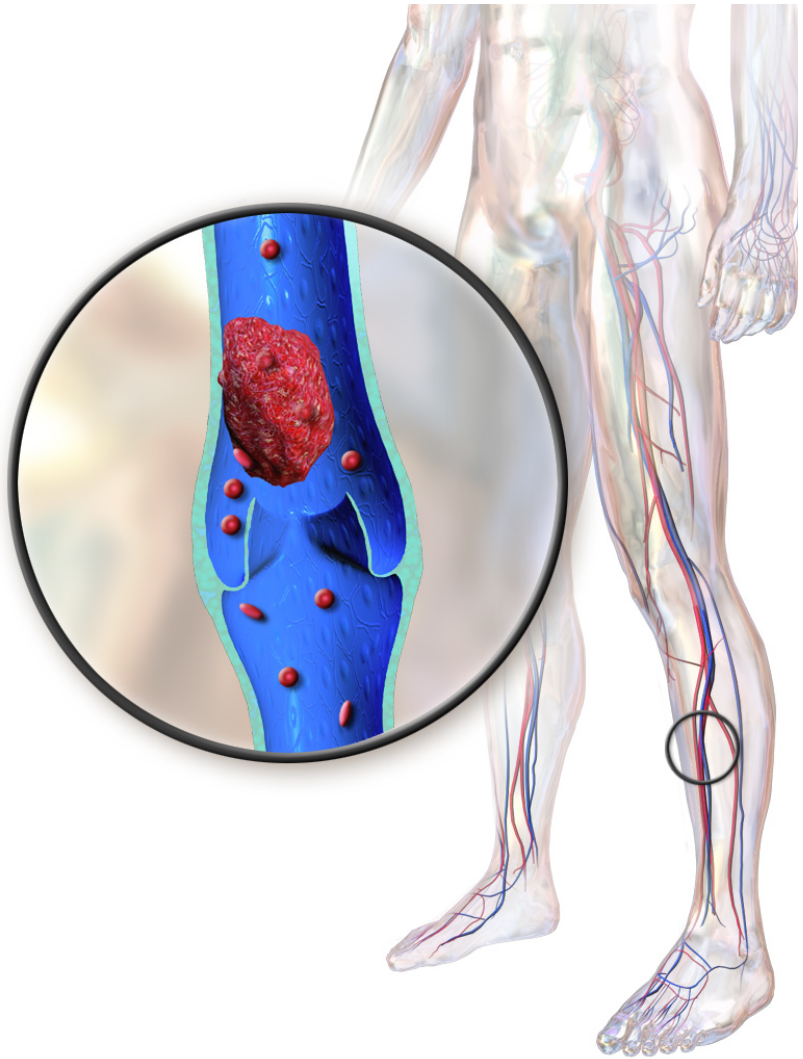
PULMONARY ARTERY ANEURYSMS



VESSEL INVOLVEMENT

Recurrent phlebitis commonly involves the large vessels resulting in thrombus formation. In general there is a thrombus formation predisposition affecting the peripheral veins. Thrombosis of the vena cava and of the right atrium has also been described.

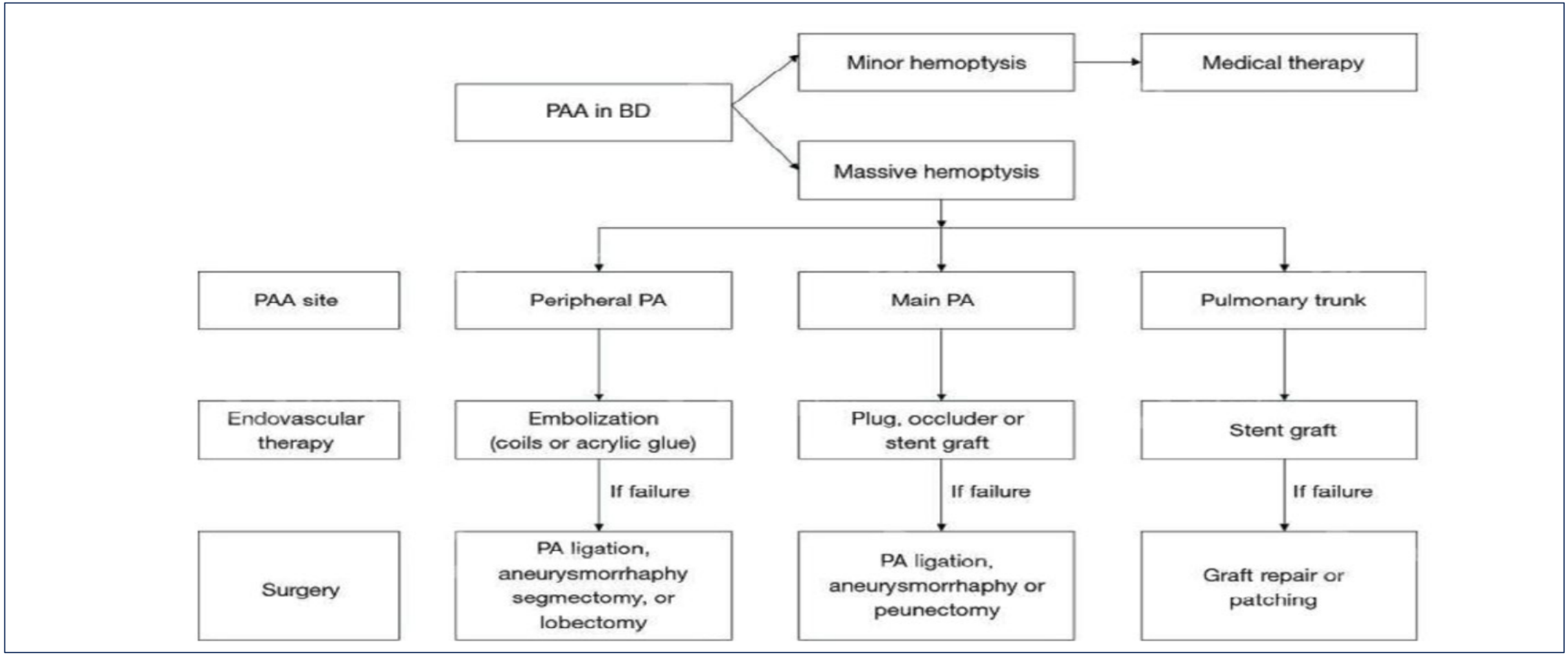
Histologic studies show destruction of the arterial wall and perivascular lymphomonocytic infiltration of capillaries and venules.



TREATMENT

- IMMUNOSUPPRESSION
- SURGICAL RESECTION OR ENDOVASCULAR TREATMENT OF HIGH-RISK LESIONS
  - Lobectomy, Segmentectomy
  - Pneumectomy
  - Ligature of carotid false aneurysm and posterior tibial artery
  - Exeresis of pulmonary mass
  - Repeated embolization of bronchial artery
  - Resection of pulmonary aneurysm
  - Lung transplantation
  - Embolization of pulmonary aneurism or bronchial artery
  - Chest drainage
  - Atrial mass removal
- THE USE OF ANTICOAGULANTS IS STILL DEBATED IN HSS

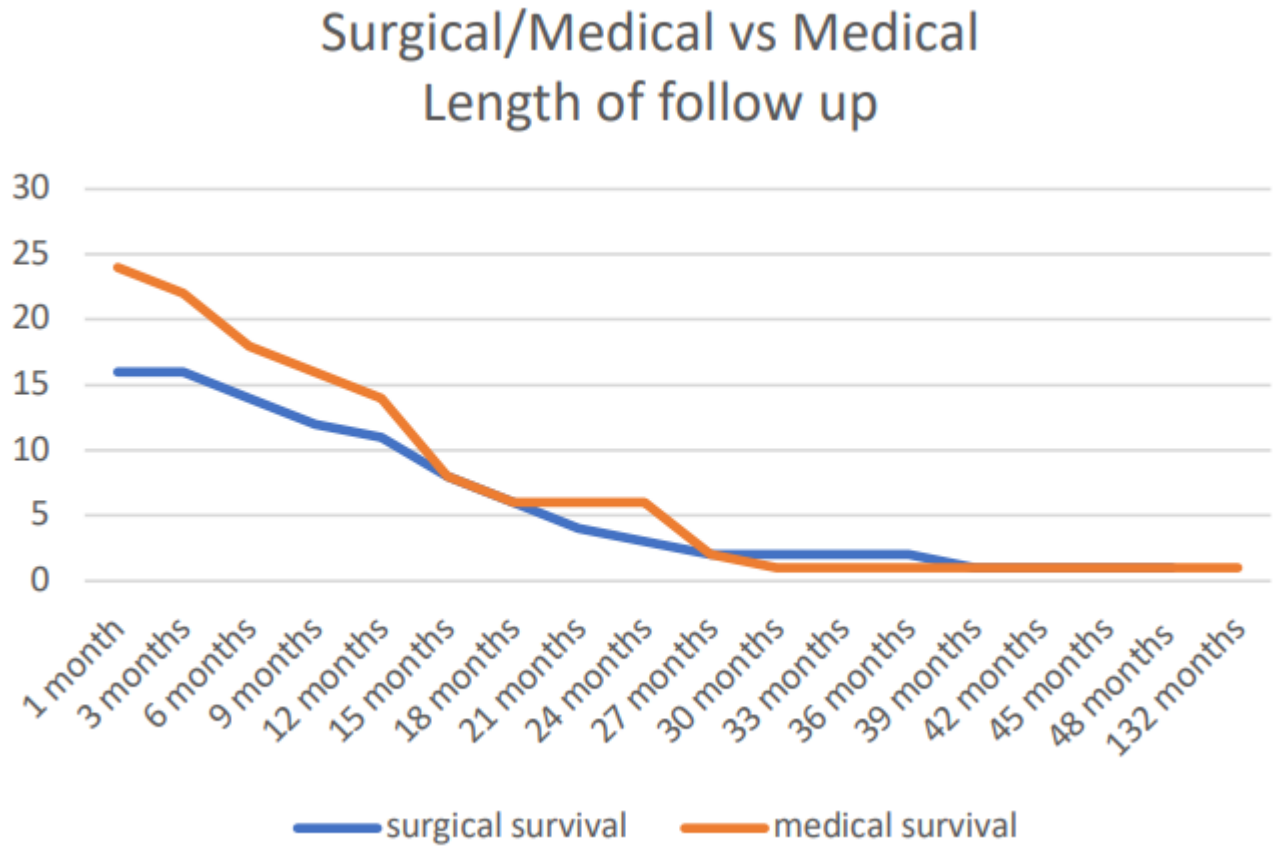
Managment protocol of PAA in BD according to degree of hemoptysis and presenting clinical condition: open surgery and endovascular treatment



Journal of Surgery 16:4; 2020 Abd Alkader

TREATMENT

Literature review of follow up length of patients after HSS diagnosis: surgical versus medical/surgical therapy.



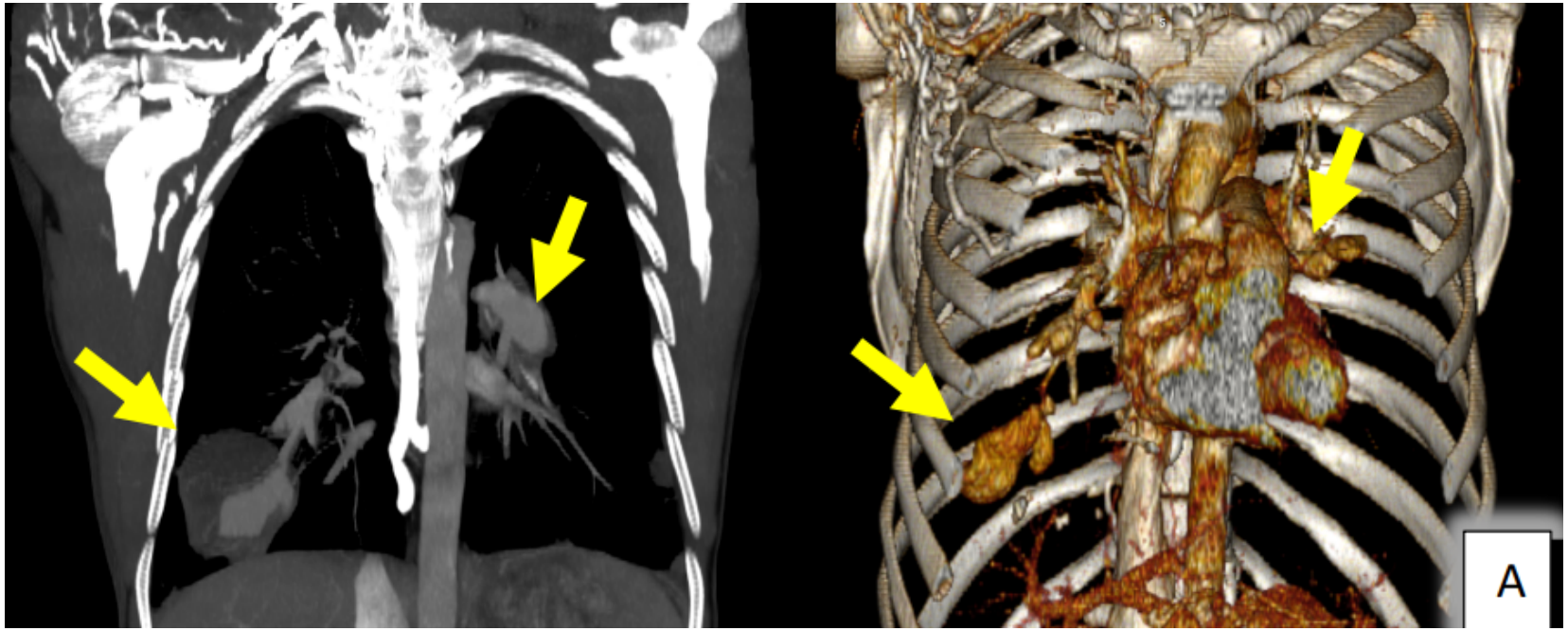
CASE

- 33-year-old male
- Hemoptysis
- shortness of breath
- pain in the left lower limb

- CT/DUS → superior vena cava parietal thrombosis, left external iliac and femoral veins thrombosis

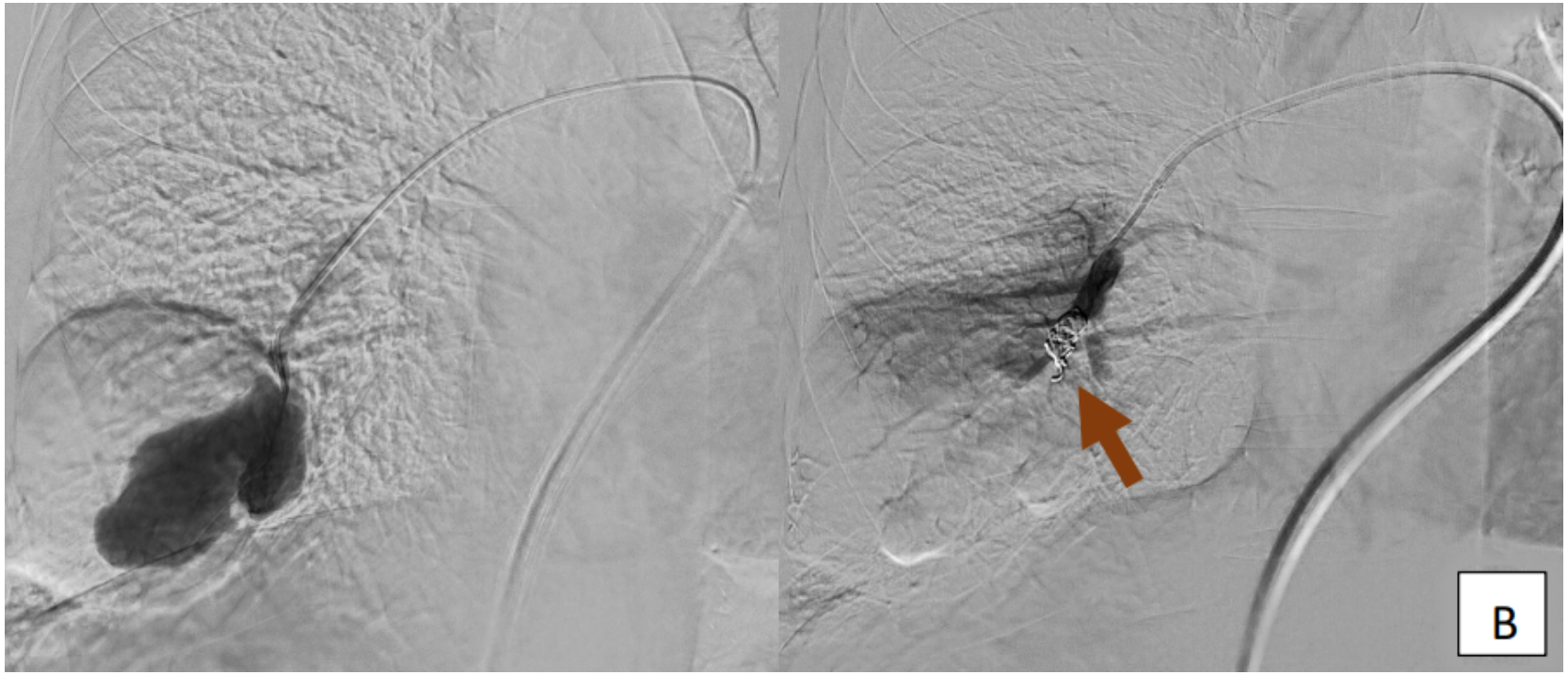
CASE

Preoperative image CT MIP and SSD reconstruction showing pulmonary aneurysms: patent right great aneurysm and little left aneurysm



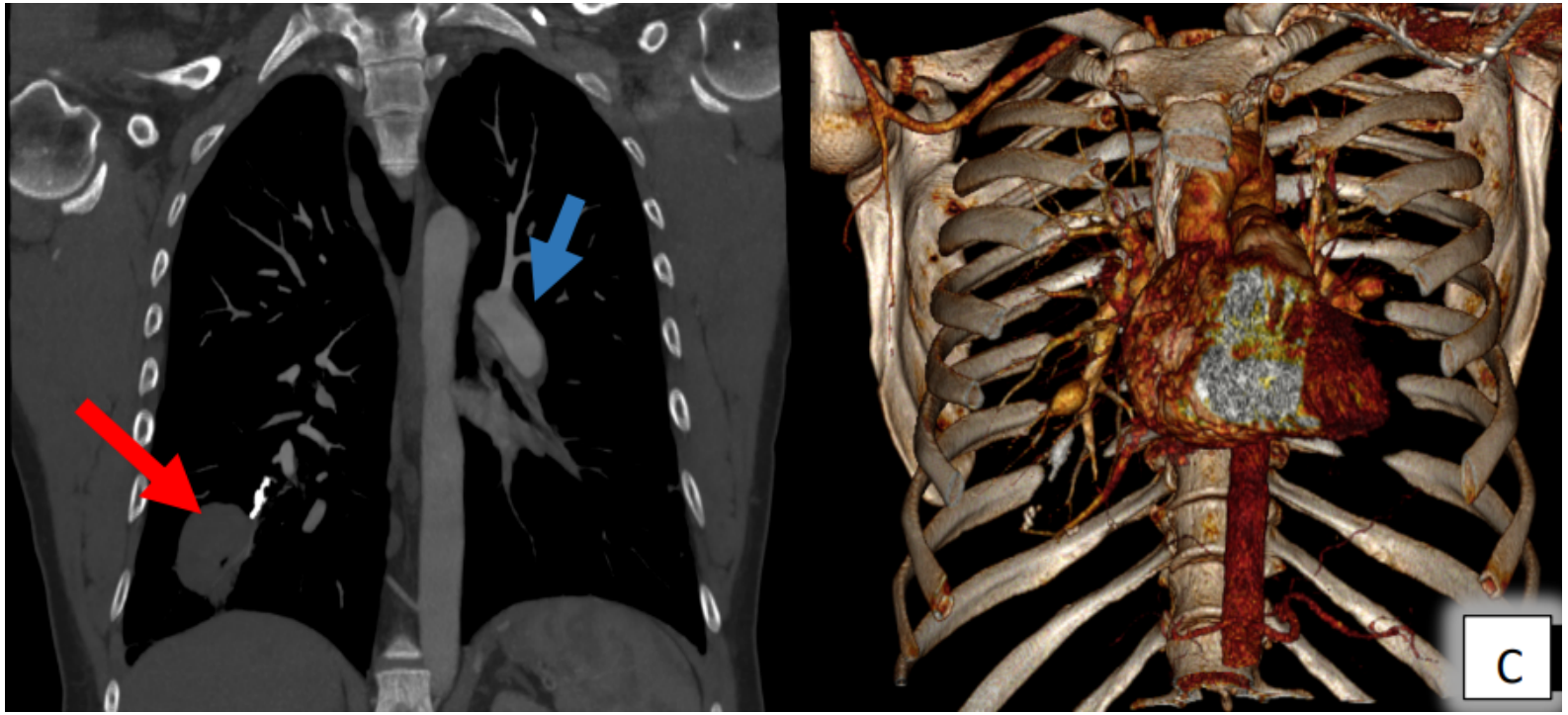
CASE

Intraoperative selective angiography confirming the non-ruptured aneurysm and successful coil embolization with aneurysm exclusion previous implantation of a vena cava filter.



CASE

Postoperative image CT MIP and SSD reconstruction showing complete right aneurysm exclusion and unmodified left aneurysm



CONCLUSION

- The frequency of this syndrome is rare, but its consequences can be fatal.
- The possibility of verifying a genetic correlation, also linked to Bechet's disease, may be represent the challenge of the future.
- New therapeutic options make treatment less invasive.

Thank you

- Vascular surgery: Fabio Massimo Oddi, Federico Pennetta, Alice de Giorgi and Arnaldo Ippoliti
- Interventional radiology: Renato Argirò, Daniele Morosetti
- Cardiac surgery: Lorella Belvivere
- Reumatology: Barbara Kroegel, Paola Triggianese, Elisabetta Greco
- Genetic: Federica Sangiuolo
- Internal medicine: Ilaria Coccia, Manfredi Teasuro