2017 MID-ATLANTIC
CONFERENCE

7th ANNUAL CURRENT CONCEPTS IN
VASCULAR THERAPIES

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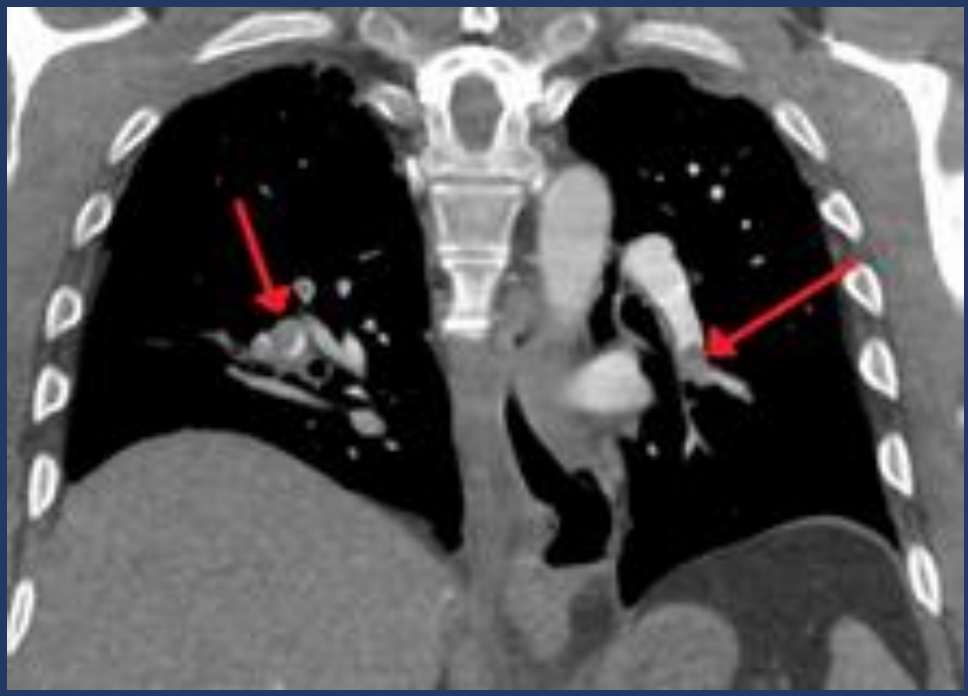
Phlegmasia Cerulea Dolens - A Limb Threatening Problem

Disclosures

Complications of DVT

- Early
  - Pulmonary Emboli
    - Life threatening “saddle embolus”
    - Incidence of PE is estimated to be approximately 60 to 70 per 100,000
    - PE is found in up to 50% of DVT patients (Silent PE)

A blood Clot in the Pulmonary Artery



Complications of DVT

- Early
  - What else?????

Phlegmasia Alba Dolens

Phlegmasia Cerulea Dolens

Venous Gangrene

Complications of DVT

- Late
  - PTS (Post Thrombotic Syndrome)
    - Leg swelling
    - Leg pain
    - Skin discoloration
    - Varicosities
    - Venous ulceration

Early Complications of DVT

Phlegmasia Alba Dolens → Phlegmasia Cerulea Dolens → Venous Gangrene

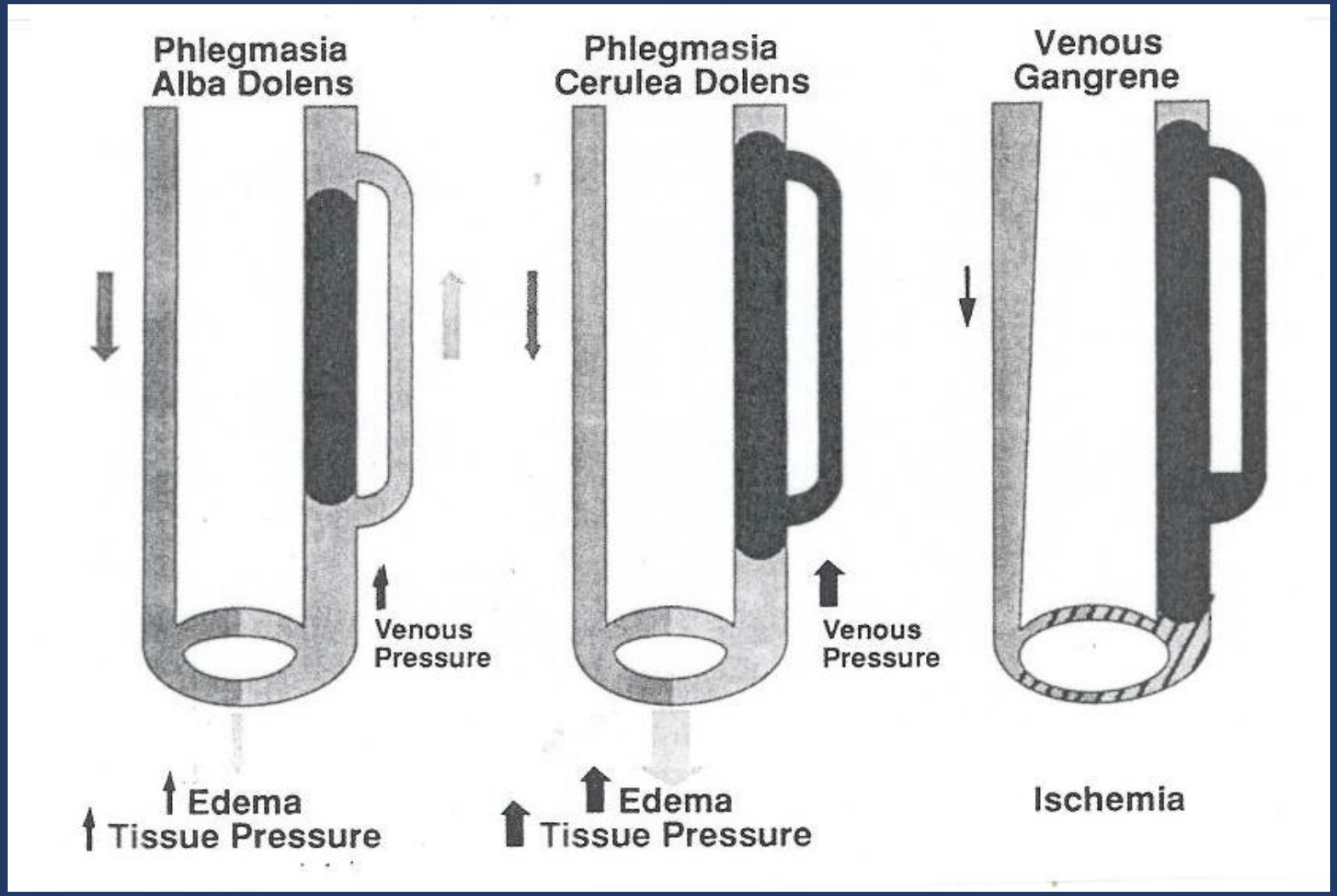
Plegmasia Alba Dolens

- Phlegmasia “Alba” Dolens
  - “Alba” means white (i.e. Albino)
  - Referred to as “Milk Leg or White Leg”
  - Historically seen in pregnant women (third trimester) or mothers who had just given birth
    - Compression of Lt iliac vein against the pelvic rim from an enlarged uterus
  - Presently it is due to venous occlusion (DVT)
    - 40% of patients with phlegmasia alba dolens have an underlying malignancy

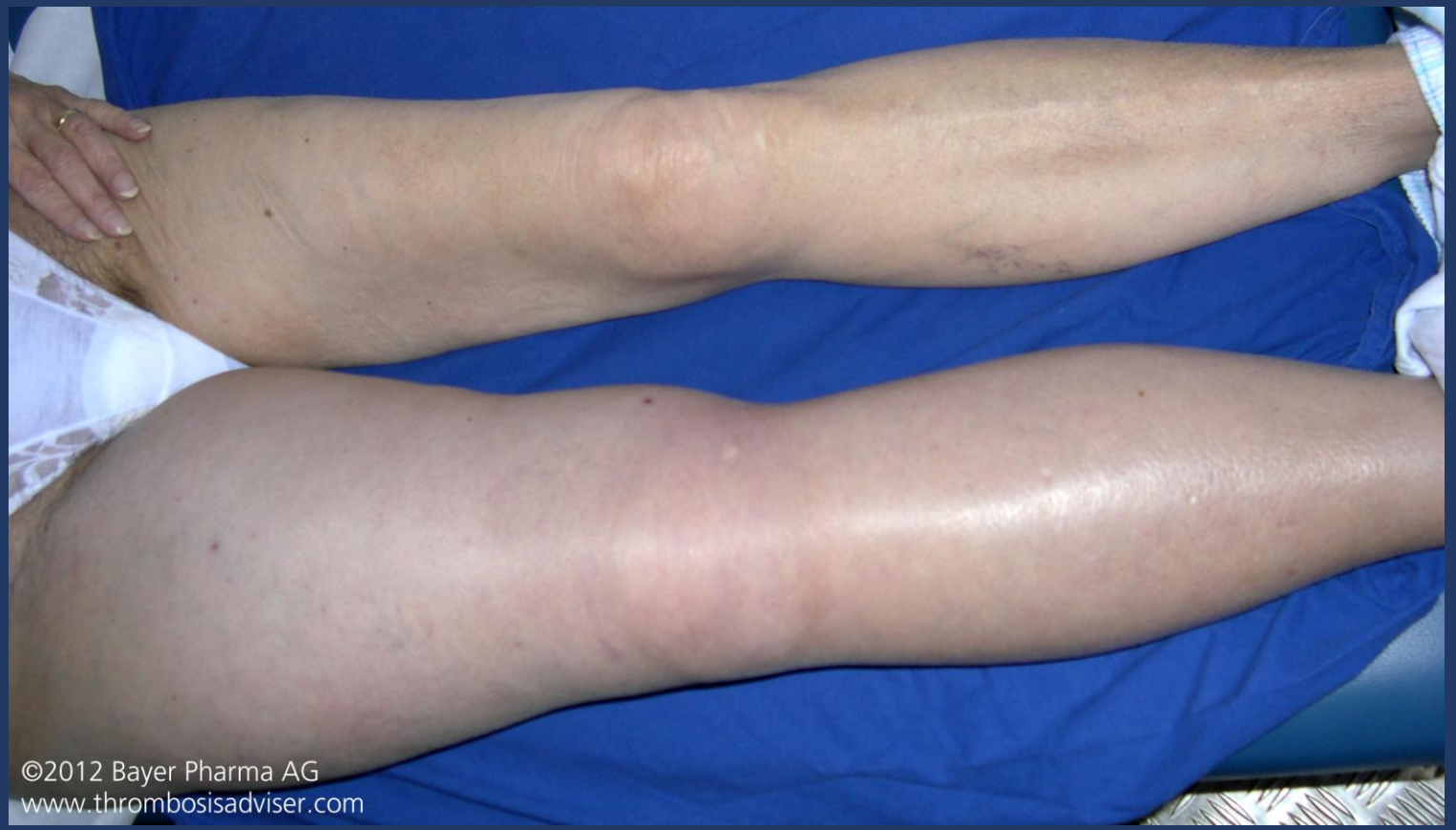
Pathophysiology

- Phlegmasia “Alba” Dolens
  - Thrombosis involving ONLY the major deep venous channels but SPARING the collateral veins
  - Venous drainage is decreased but still present via the superficial channels
  - At this stage the leg is painful, swollen and appears white (pale)
    - But no arterial compromise (either palpable or dopplerable pulses)

Pathophysiology



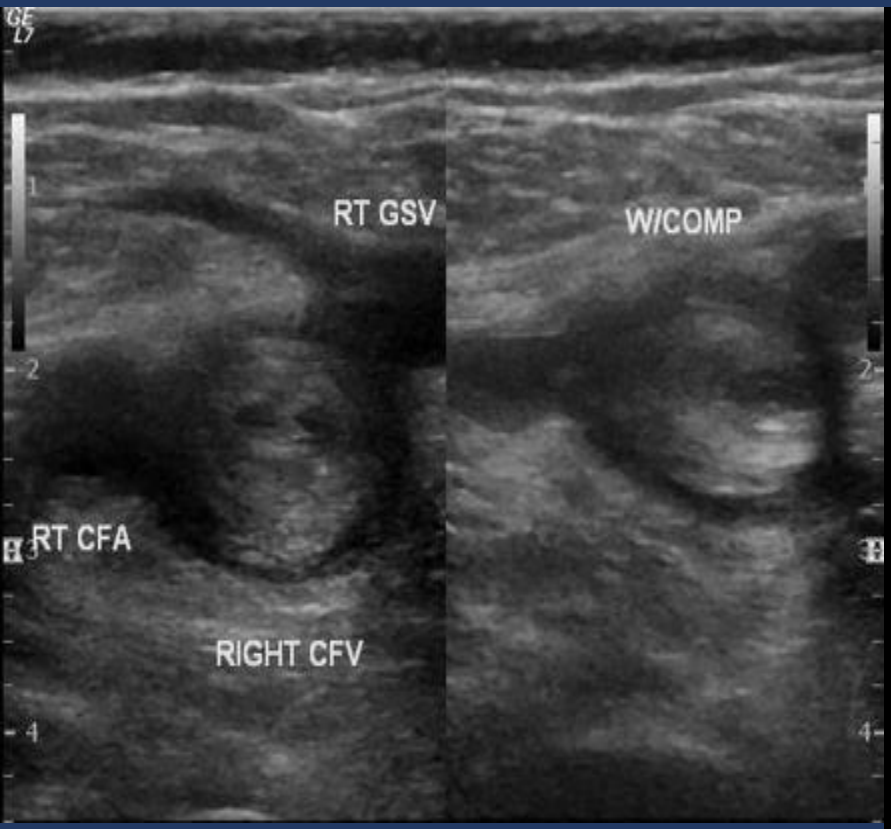
Phlegmasia Alba Dolens

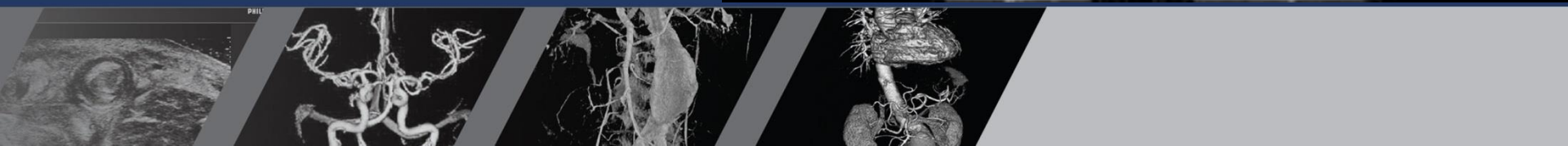


Diagnosis

- Phlegmasia “Alba” Dolens
  - Diagnosis is made with venous duplex
    - Identify Acute DVT
    - Location of DVT (proximal vs distal)
    - Extent of DVT
  - Contrast enchanced CT scan can be helpful
    - To identify thrombus is pelvic veins or IVC
    - Check for underlying malignancy

Diagnosis





Treatment

- Phlegmasia “Alba” Dolens
  - Treatment
    - Admit to inpatient
    - Anticoagulation (IV heparin gtt)
    - Elevate affected limb
    - Vascular surgery consultation
      - Medical management vs. Endovascular venous lysis (CDT, mechanical thrombectomy, pharmacomechanical thrombectomy)

Phlegmasia Cerulea Dolens

- Phlegmasia “Cerulea” Dolens (PCD)
  - “Cerulea” means Blue
  - Referred to as “Painful Blue Leg”
- Rare condition that occurs in < 1% of DVT patients
- Up to 90% of patients with PCD have underlying malignancy (50% occult malignancy)

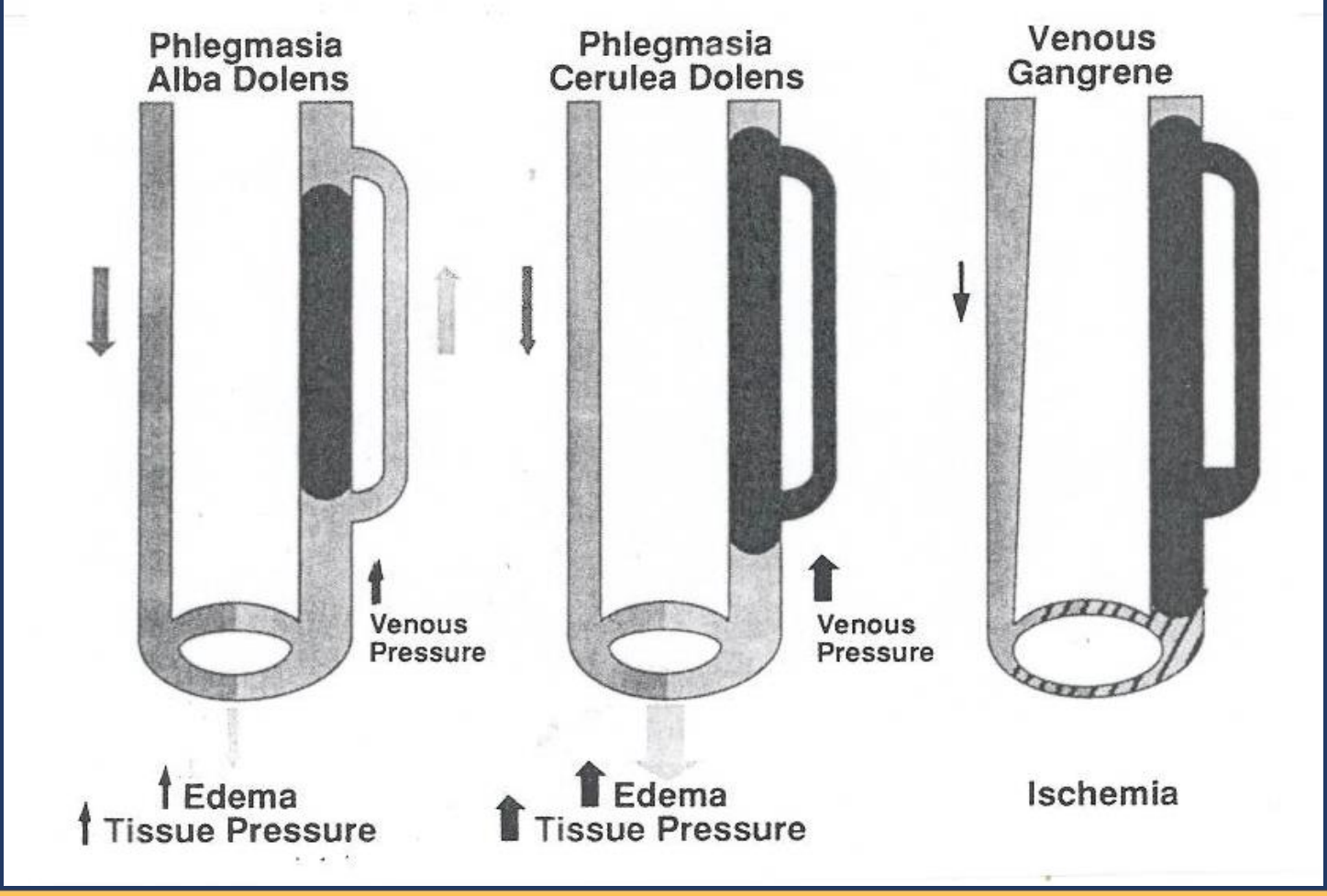
Phlegmasia Cerulea Dolens

- 50% of patients with PCD progress to Venous Gangrene
  - 30-50% Limb Amputation rate
  - Overall Mortality rate of 20-40%

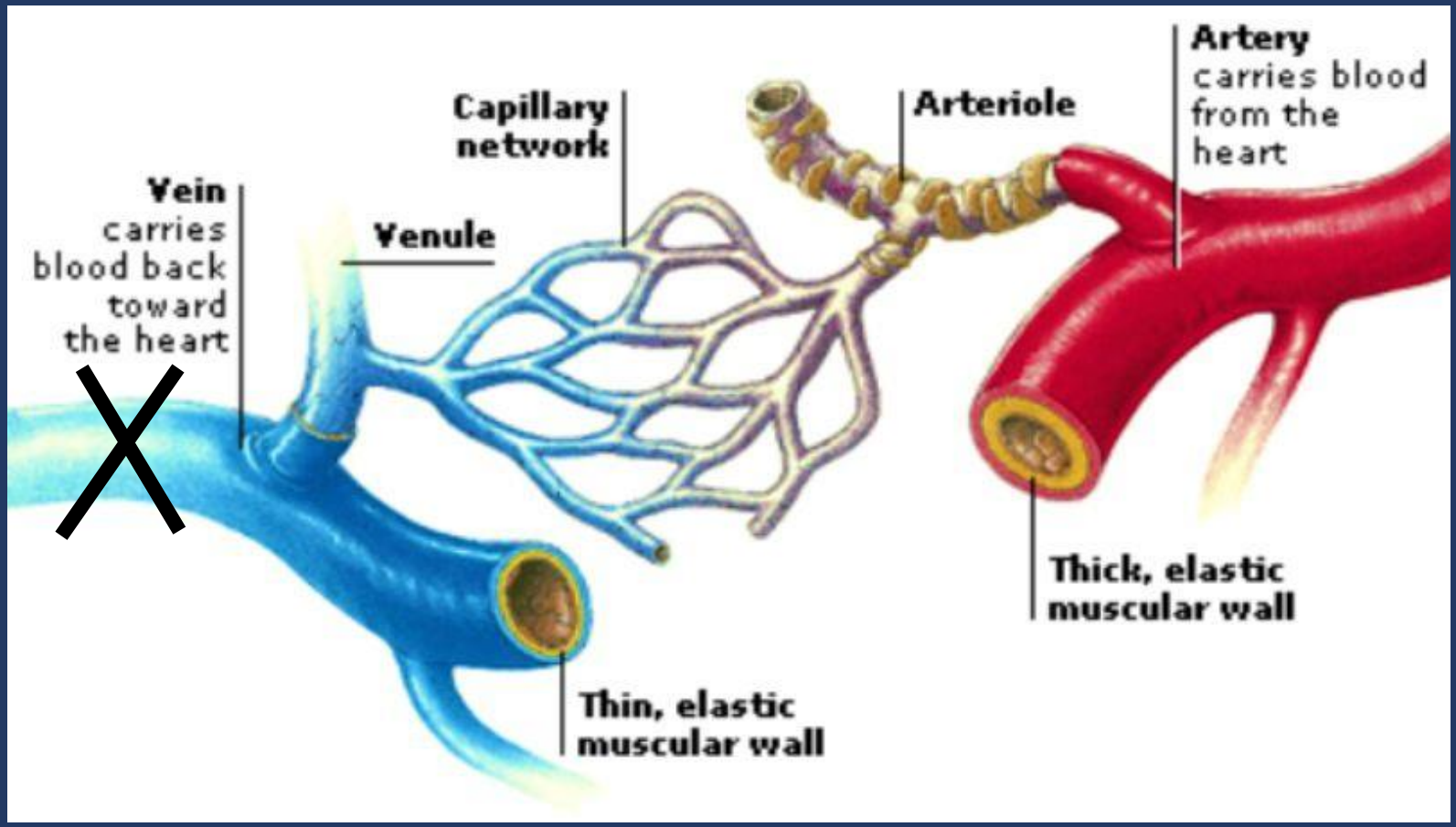
Pathophysiology

- Phlegmasia “Cerulea” Dolens
  - Thrombosis causing COMPLETE occlusion of venous drainage (deep and superficial system)
  - Leads to increase in capillary pressure
  - Leads to exudation of fluid into the interstitial space
  - Leads to skin blistering
- Characteristics of PCD
  - Pain, swelling and most importantly cyanosis “blue appearance”

Pathophysiology



Pathophysiology



Diagnosis

- Phlegmasia “Cerulea” Dolens
  - Based on clinical signs and symptoms
    - Painful, blue, swollen leg
  - Venous duplex to confirm the diagnosis and localize the thrombus (purpose of intervention)
  - Contrast enchanced CT to identify centrally located and pelvic thrombus

Plegmasia Cerulea Dolens

Treatment

- Phlegmasia “Cerulea” Dolens
  - THIS IS AN EMERGENCY!!
  - PATIENT IS GOING TO THE OR FOR SURGICAL VENOUS THROMBECTOMY
  - Fluid resuscitation
    - Patients are hypotensive and sometimes in shock due to fluid extravasation and loss of intravascular fluid
  - Anticoagulation
    - IV heparin bolus prior to OR

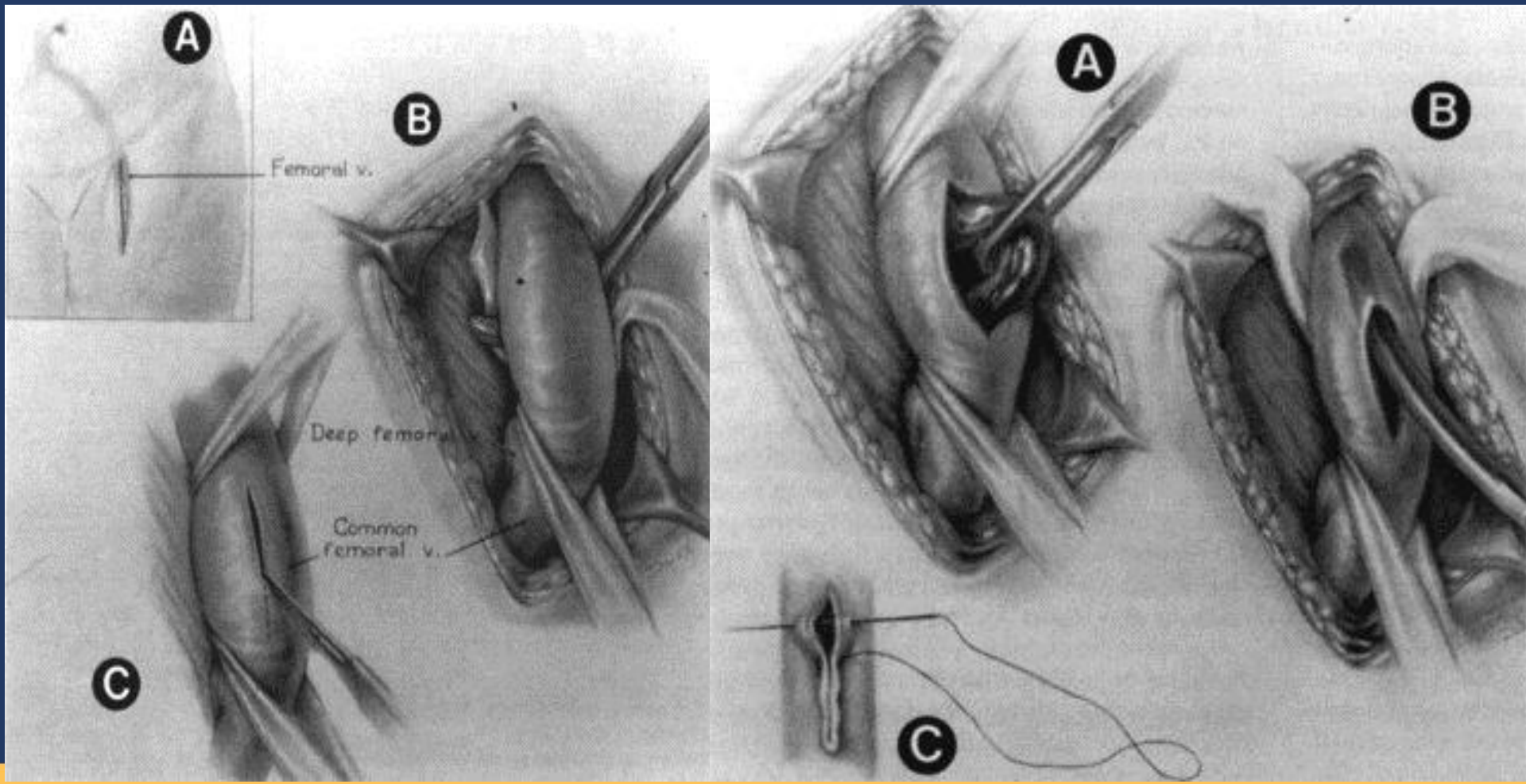
Treatment

- Phlegmasia “Cerulea” Dolens
  - Systemic tPA vs Catheter directed thrombolysis
    - Literature does not support either therapy as being successful
    - Time is tissue
  - Some surgeons recommend a brief (6 hours) of IV heparin gtt with profound leg elevation
    - If unsuccessful then will proceed with surgical venous thrombectomy

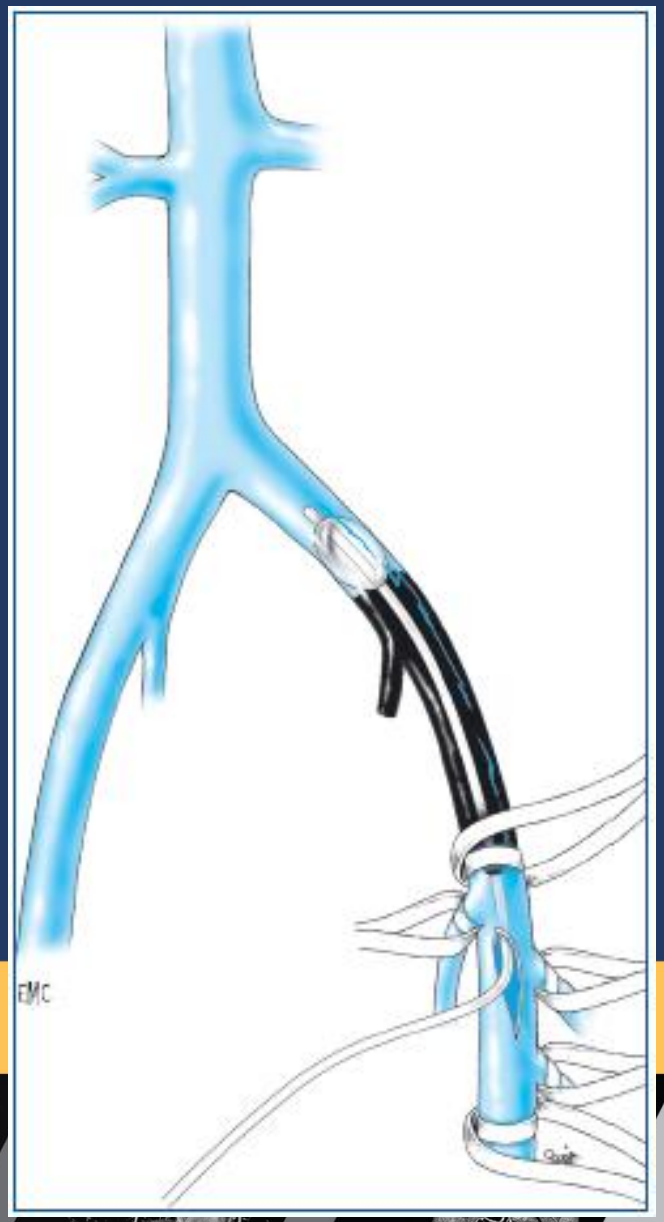
Surgical Treatment

- Phlegmasia “Cerulea” Dolens
  - Local, Regional or General anesthesia
  - Longitudinal groin incision to expose CFV, GSV and SFA
  - IV Heparin intra op if not given preop (Check ACT)
  - Venotomy to facilitate Fogarty catheter thrombectomy
    - American surgeons – place IVC filter (contralateral groin) before
    - European surgeons – no filter, perform thrombectomy with positive pressure ventilation or Valsalva maneuver

Surgical Treatment



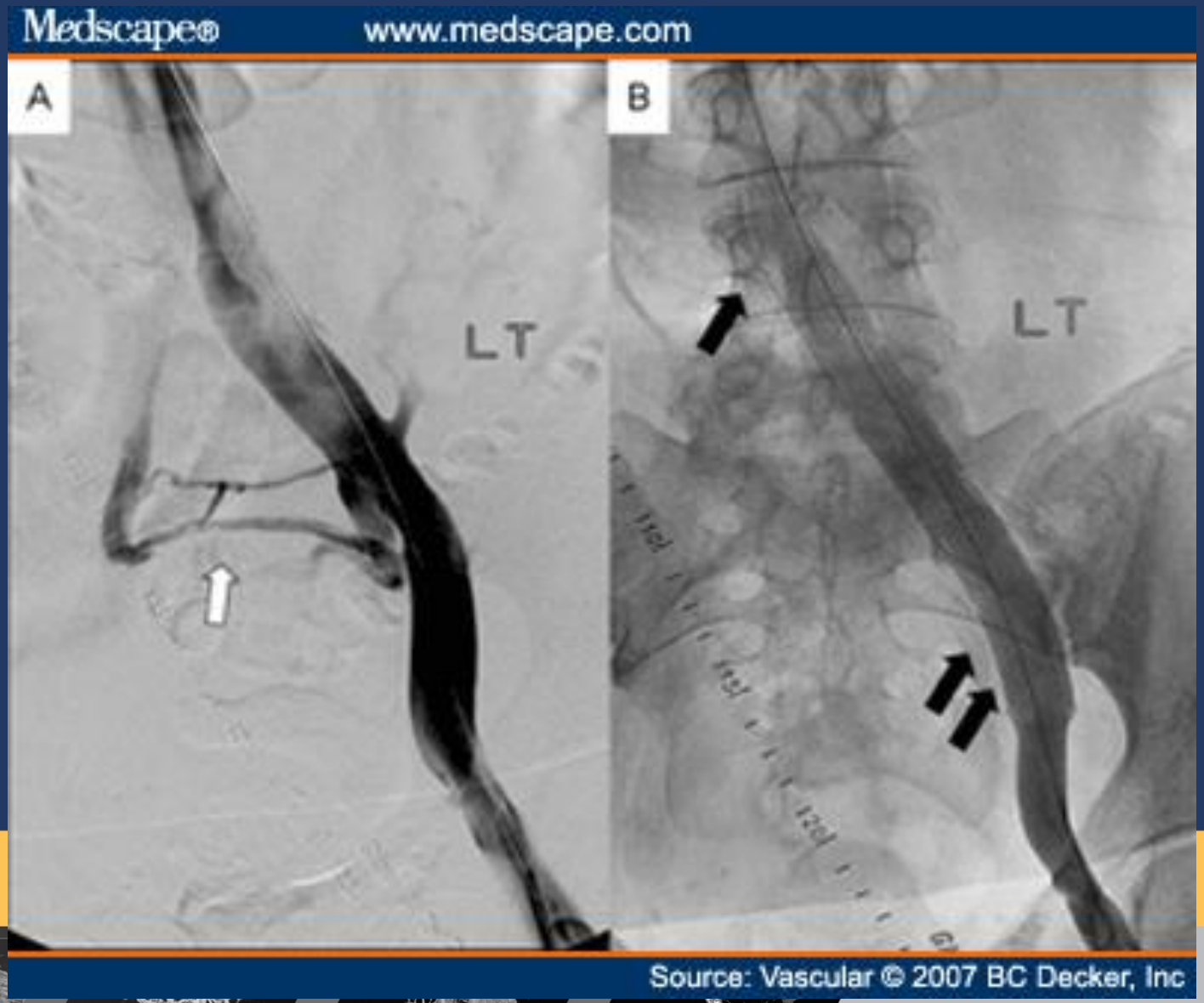
Surgical Treatment



Surgical Treatment

- Phlegmasia “Cerulea” Dolens
  - Place Fogarty balloon in Common Iliac vein
  - Pass suction catheter parallel to Fogarty and try to suction out the internal iliac vein thrombus
  - Must confirm iliac vein flow
    - MANDATORY VENOGRAM
  - If Iliac vein stenosis/compression (MTS) is noted then may need balloon angioplasty + stenting

Surgical Treatment



Surgical Treatment

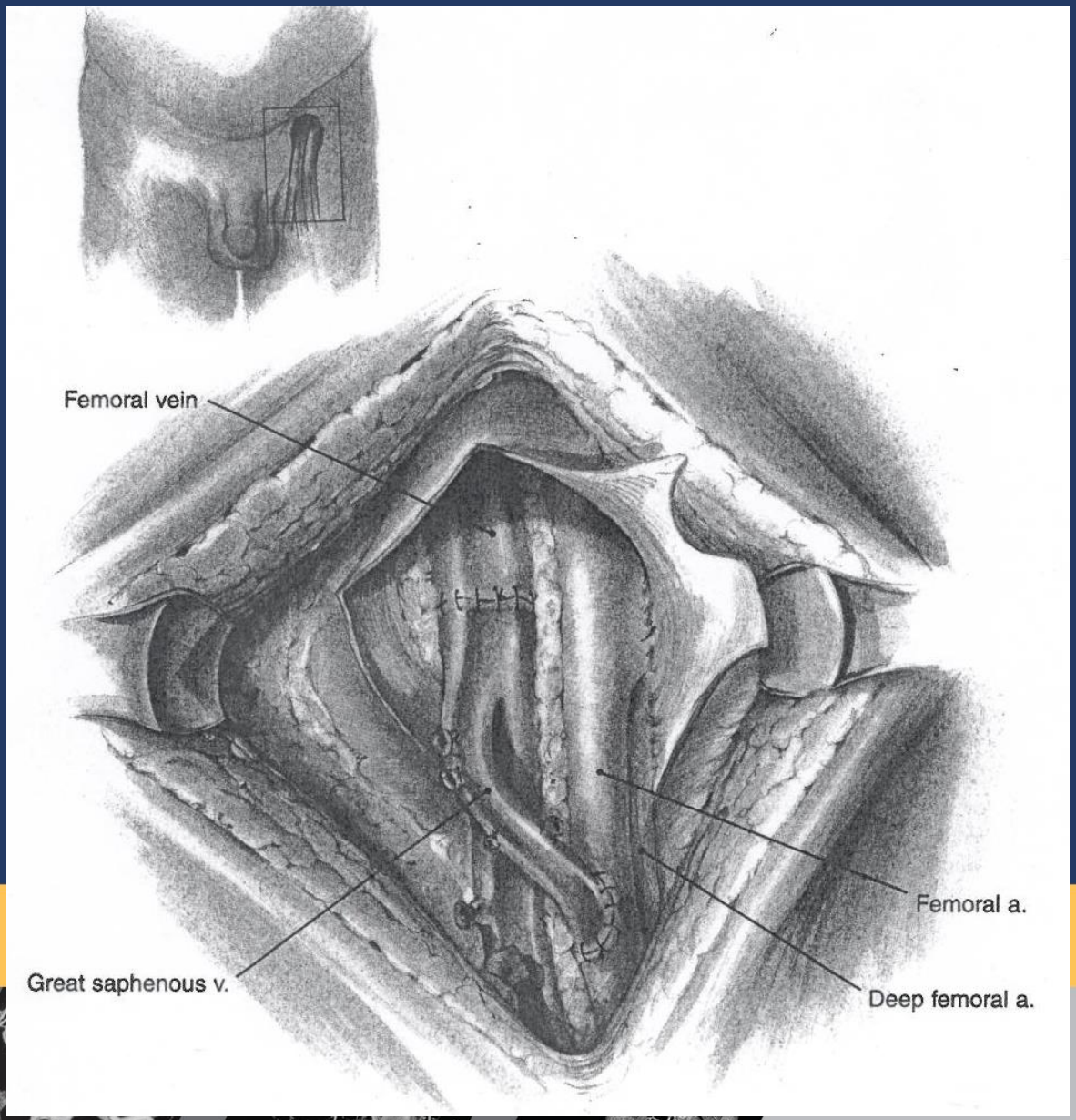
- Phlegmasia “Cerulea” Dolens
  - Thrombus in infrainguinal region is expressed manually using an Esmarch
  - Start wrapping at the base of the toes and proceed proximally to the groin incision
    - Passing fogarty distally will damage venous valves

Surgical Treatment

Surgical Treatment

- Phlegmasia “Cerulea” Dolens
  - Once the venous outflow is restored the venotomy is closed
  - An autogenous AVF is created in groin to increase iliac vein patency
    - GSV is divided and proximal end is anastomosed to SFA
  - Perform a 4 compartment lower leg fasciotomy

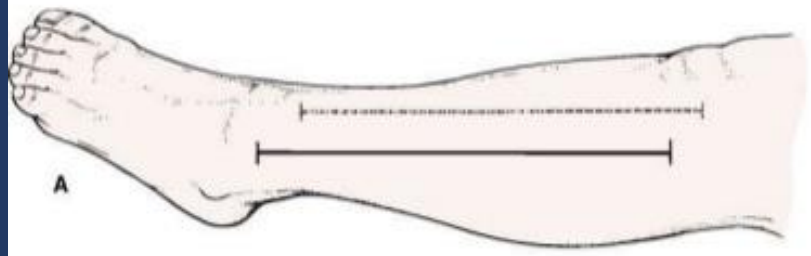
Surgical Treatment

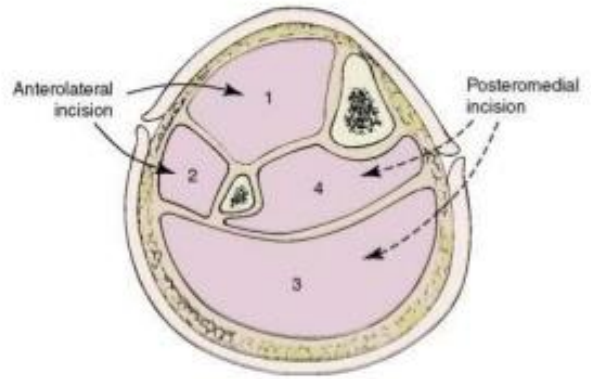


Surgical Treatment

DOUBLE-INCISION LEG FASCIOTOMY

Used to adequately decompress all four compartments





Surgical Treatment

- Phlegmasia “Cerulea” Dolens
  - What if there is thrombus extending into the IVC?
    - Transperitoneal incision
    - Expose IVC below renal veins to bifurcation
    - IVC is opened and the thrombus is removed

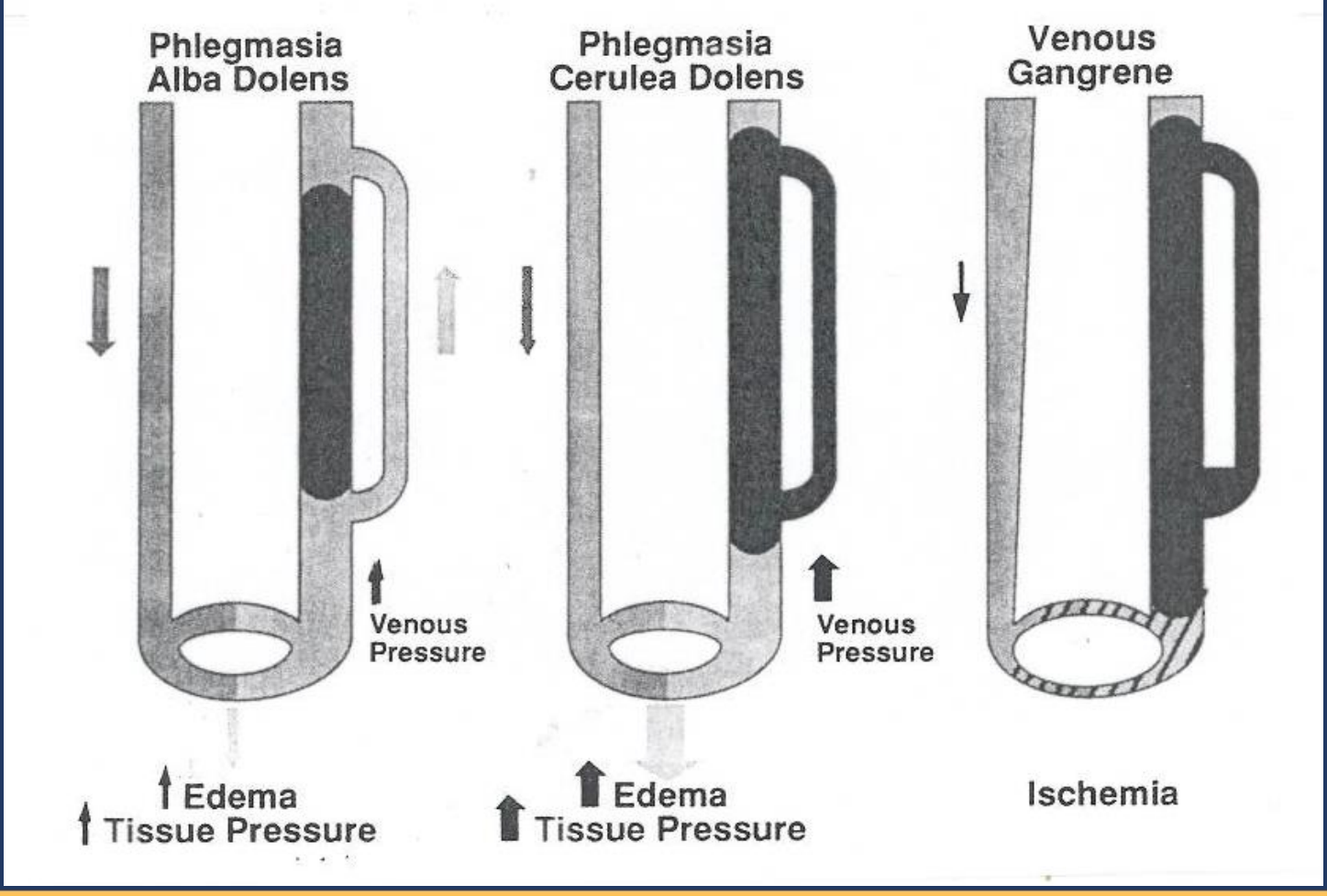
Post Operative Treatment

- Phlegmasia “Cerulea” Dolens
  - Continue IV heparin drip x 5 days
  - Then transition to oral anticoagulation (coumadin, xarelto, eliquis) x 6 months
  - Gradient Compression Stockings
  - Once swelling decreased, return to OR to close fasciotomy sites or heal with secondary intention
  - Ligate groin AVF in 6-12 weeks

Venous Gangrene

- Venous Gangrene
  - Massive iliofemoral or IVC occlusion with extensive vascular congestion and venous ischemia
  - Thrombosis causing COMPLETE occlusion of venous drainage (deep and superficial system) AND ARTERIAL COMPROMISE
  - 50% of phlegmasia cerulea dolens progress to venous gangrene

Pathophysiology



Venous Gangrene

- Clinical Features
  - Excruciating Limb pain
  - Severe edema
  - Blistering with fluid extravasation
  - Superficial gangrene and necrosis
  - No motor or sensory to foot
  - IRREVERSIBLE (Phlegmasia Alba and Cerulea Dolens are reversible)
  - Treatment is AMPUTATION

Venous Gangrene

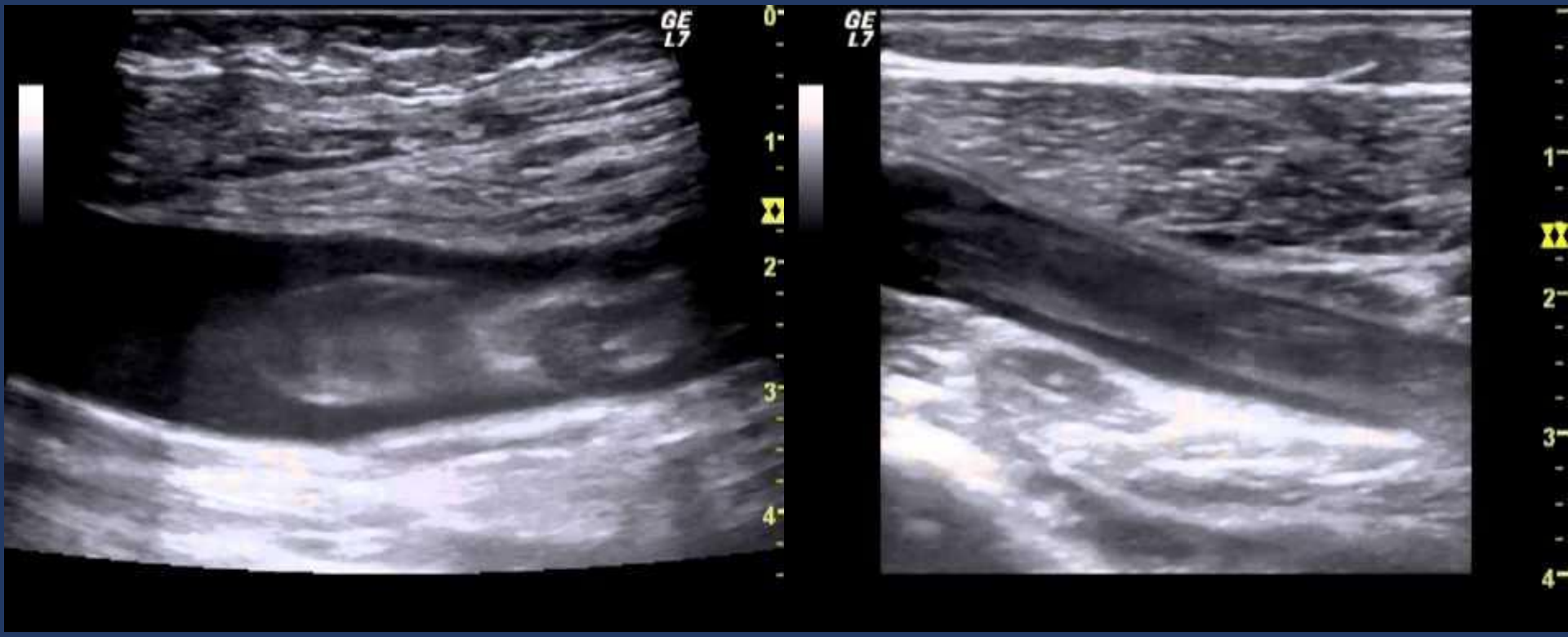
Case Study

- 75 year old male presents to ED with LLE pain and swelling x 1 day
- Denies CP or SOB
- Pt recently diagnosed with Stage IV Lung cancer and received 2 doses of chemotherapy via a mediport
- Vital signs are stable
- O/E – LLE is edematous but NO PHLEGMASIA, Pedal pulses are palpable

Case Study

- Blood work is normal
- LLE venous duplex to r/o DVT
- Venous duplex demonstrated acute femoropopliteal and tibial occlusive DVT, no extension into external iliac vein

Case Study



Case Study

- What do you do?
- Patient was discharged from ED with Xarelto (starter pack) x 21 days
- Instructed to follow up with vascular surgery in the next week
- Keep LLE elevated

Case Study

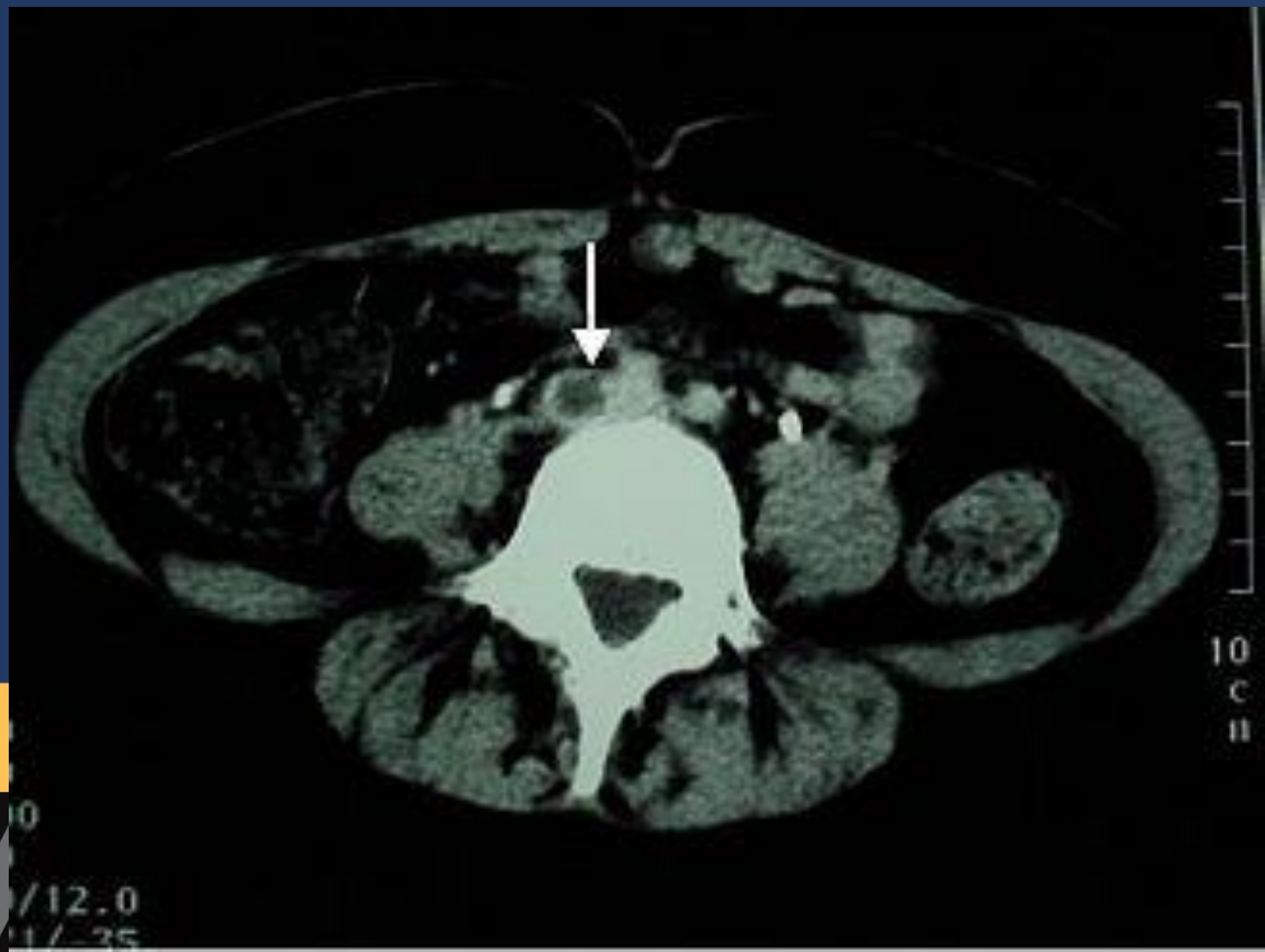
- Patient returns to ED 2 days later complaining of worsening pain and swelling
- A different ED physician evaluates patient
- O/E – LLE is much more swollen and appears white
- Repeat LLE venous duplex is ordered and now demonstrates Acute Left external iliac vein, femoral vein, popliteal vein and tibial vein DVT
- Vascular surgery consult obtained

Case Study

- Vascular surgery evaluates patient and feels that patient has developed Phlegmasia Alba Dolens
- Pt is admitted to ICU
- Initiate IV heparin gtt
- CT A/P with contrast to better evaluate IVC and pelvic veins
- Keep LLE elevated

Case Study

- No IVC clot
- Clot extends to the Lt CIV



Case Study

- Discuss options of medical management (IV Heparin vs. Endovascular thrombolysis/thrombectomy)
  - Risk of bleeding from venous lysis is high due to stage IV lung cancer
- Decision made to continue IV heparin and perform serial exams with the understanding that if this progress to phlegmasia cerulean dolens then patient will need surgery

Case Study

- Next morning the LLE pain is worse and now has spots of bluish purplish areas (new)

Case Study

- Pt taken to OR for surgical venous thrombectomy
  - Able to clear iliac vein clot
  - Venogram was normal
  - Able to express infrainguinal clot
  - AVF created in left groin
  - LLE prophylactic fasciotomy performed
- Post op patient continued on IV heparin gtt

Case Study

- Over the course of next few days, the LLE swelling, pain and color improved
- Fasciotomy sites closed on POD #4
- Pt given GCS
- Discharged on coumadin x 6 months

Take Home Points

- Phlegmasia “Alba” Dolens – “White” leg
  - Medical management (IV Heparin) vs Endovascular therapy
  - Check for Malignancy (40%)
- Phlegmasia “Cerulea” Dolens – “Blue” leg
  - Limb threatening emergency
  - Mandatory open intervention
  - Check for Malignancy (90%)

Take Home Points

- Venous Gangrene – “Game Over”
  - Amputation

Thank You