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REVIEW PAPER

# Brief review: Pulmonary artery aneurysms and pseudoaneurysms

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**Abstract**
The purpose of this review article is to provide a brief overview of pulmonary artery aneurysms and pseudoaneurysms, to discuss the classifications of these conditions, review the role of imaging and discuss management in affected patients.

**Keywords** Pulmonary artery aneurysms · Pulmonary artery pseudoaneurysms · Endovascular

## Brief overview

Pulmonary artery aneurysms (PAAs) and pulmonary artery pseudoaneurysms (PAPs) are rare but often lethal. The presentation of these vascular pathologies can range from a silent incidental finding on imaging to massive hemoptysis [1, 2]. Massive hemoptysis (MH), classically defined as the expectoration of more than 300 mL in 24 h, is associated with significant morbidity and mortality [2–6]. However, MH is present in only a minority of cases; more common symptoms include cough, dyspnea, and pleuritic chest pain, with symptomatic presentation dependent on the size and location of the aneurysmal vasculature [7, 8].

Workup of suspected PAA or PAP may consist of bronchoscopy, computed tomography angiogram (CTA), magnetic resonance angiogram (MRA), or catheter angiogram. Although these methods can help to identify the PAA or PAP, selective arteriography offers a means to perform endovascular intervention, in addition to diagnosis. Timely treatment is crucial given the risk of enlargement and rupture leading to death, which occurs in approximately 50% of untreated patients [9].

## Terminology

A true PAA involves all three layers of the vessel wall, whereas a PAP does not involve all three layers. Simply put, "aneurysmal" vascular anatomy refers to a degree of dilatation beyond the upper limit of the accepted range per vessel diameter. The dilatation can be saccular or more fusiform in shape [1, 10].

## Physiology

In normal physiology, the hydrostatic pressure within an artery is balanced by the tension applied by the arterial wall. By the law of Laplace ($T = P \times R/t$), these opposing pressures are directly proportional to the size, specifically the radius, of the artery [2, 11]. Histologic cross-sections of the elastic pulmonary artery and the distal muscular arterial branches reflect the adaptive changes in the media layer of the arterial wall that allow for adequate arterial wall tension via elastic fibers and layers of smooth muscle cells [12].

In an aneurysm, the functional radius increases in a structure that was not designed to accommodate higher pressures. This corresponds to the risk of enlargement over time seen in PAAs. When the tension required to oppose hydrostatic pressures exceeds the strength of the wall, the risk of rupture is greater, especially when there is nonuniform disruption in the arterial wall as seen in PAPs.

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## PAA/PAP classification

Pulmonary artery aneurysms were originally classified based on the presence of venous communication [2]. Other groups have since differentiated PAAs by location (proximal or distal), whereas others suggest that PAAs should be categorized based on underlying pathology and classified based on etiology [13]. Although venous involvement or location may affect flow dynamics and hence interventional approaches, classifying PAAs by etiology would separate aneurysms that may behave similarly. A more uniform classification system based on etiology would allow for more clinically meaningful reports to be generated, to be easily compared, and to be reviewed. An effort such as this could potentially lead to more accurate identification of optimal workup and therapeutic options based on underlying pathology.

A more recent classification of PAA (Table 1) delineates the at-risk populations by defining congenital and acquired etiologies [10]. The three most common causes of PAA are structural cardiac and vascular abnormalities, infection, and pulmonary hypertension. The common factor unifying most of these pathologies is weakening of the media layer [2, 11].

The leading causes of PAP are infection, trauma (especially iatrogenic injury), and malignancy [9]. PAP has often been reported in the context of Swan-Ganz catheter placement and less commonly in the context of chest tube placement [14]. For a point of reference, a prospective study of 500 Swan-Ganz catheterizations demonstrated a 0.2% complication rate for pulmonary artery rupture [15].

Classification of PAPs based on underlying disease process has not been described in the literature. Some groups instead classify PAPs based on location or flow. A single-center retrospective study demonstrated that PAPs show a predilection for peripheral pulmonary arteries, as seen in Fig. 1c. This study also found that when multiple PAPs are observed, it is often in the clinical setting of infection or malignancy involving the lung or endocarditis [9]. Seeding of the arterial system with septic emboli is thought to occur directly via erosion of the arterial wall and development of the pseudoaneurysm [9].

More recently, an organized classification based on pattern of blood supply has been described, with PAPs divided into four categories (Types A–D; Table 2). Type A and B PAPs have patent feeding pulmonary arteries but are distinguished by the presence of substantial bronchopulmonary shunting visualized during selective angiography. Type B PAPs typically have reversed flow in the feeding pulmonary artery because of the bronchopulmonary shunt. Type C PAPs are characterized by bronchial and nonbronchial/systemic collateral blood supply with a degree of bronchopulmonary shunting rather than a singular feeding artery seen on arteriography. Type D PAPs are not visualized on angiography but can be seen on CTA, likely because of slow arterial flow; these are not typically treated by angiographic intervention or surgery [5]. The main benefit of this classification system is that it can drive management, particularly the technical aspects of maintaining angiographic control and establishing laminar flow.

**Table 1** Etiologies of pulmonary artery aneurysms

| Congenital | Acquired | Bronchial artery aneurysm | Pulmonary vein aneurysm |
|---|---|---|---|
| Valvular/postvalvular stenosis | Pulmonary arterial hypertension | Congenital (sequestration, pulmonary agenesis) | Isolated ("varix") |
| Increased pulmonary flow (left-to-right shunt), Eisenmenger syndrome | Cystic medial necrosis | Atherosclerosis | Associated with arteriovenous malformation (hereditary hemorrhagic telangiectasia) |
| Connective tissue abnormalities[a] | Vasculitis[b] | Vasculitis[d] | Mitral valve disease |
| | Infection[c] | Bronchiectasis | |
| | Endocarditis | Tuberculosis and atypical mycobacteria | |
| | Neoplasms | Sarcoidosis | |
| | Trauma | Post-traumatic | |
| | Iatrogenic | Hereditary hemorrhagic telangiectasis (Osler–Weber–Rendu disease) | |
| | Idiopathic | Idiopathic | |

[a]Marfan disease, Ehler–Danlos disease, Williams–Beuren syndrome, tuberous sclerosis

[b]Behçet disease, Hughes–Stovin syndrome, Takayasu arteritis

[c]Tuberculosis (Rasmussen aneurysm), pyogenic, fungal, syphilis, schistosomiasis

[d]Behçet disease, Hughes–Stovin syndrome

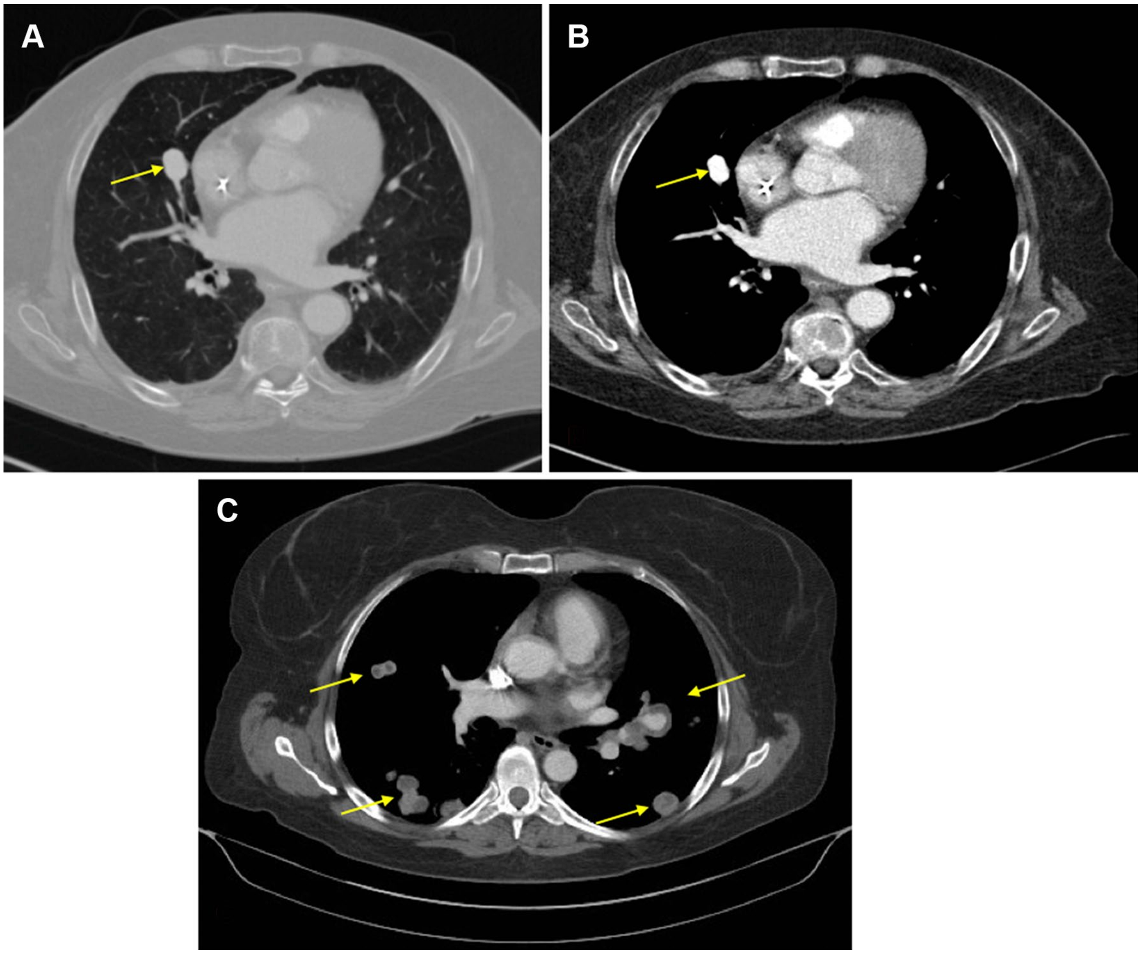


**Fig. 1** Pseudoaneurysms can be seen centrally and peripherally. **a**, **b** Central pseudoaneurysm secondary to recent right heart catheterization. **c** Peripheral pseudoaneurysms in the setting of malignancy

**Table 2** Pseudoaneurysm classification summary based on blood supply

| Category | Method of visualization | Presence of substantial bronchopulmonary shunting |
|---|---|---|
| Type A | Angiography | No |
| Type B | Angiography | Yes |
| Type C | Angiography | Yes |
| Type D | CTA | No |

## Role of imaging

Pulmonary artery aneurysms and PAPs are so rare that they are often not included in differential diagnoses and are frequently missed on initial imaging workup. A retrospective study showed that PAP was clinically suspected in only 23% of patients presenting with hemoptysis and in 13% of all patients with PAP. Radiologists did not report 46% of PAPs on initial contrast-enhanced CT scans even though these PAPs were visible in retrospect. Interestingly, a clinical history of hemoptysis did not significantly affect the detection rate for PAPs on initial CT scans [9].

For symptomatic patients, particularly those with hemoptysis, the American College of Radiology (ACR) Appropriateness Criteria noted that there is a conflict in the literature regarding bronchoscopy versus imaging and their role in workup in these pathologies and generally, in the setting of hemoptysis. The ACR specifically noted there were inconsistencies in the clinical approach to hemoptysis [16]. Reports have demonstrated that aneurysm size is underestimated with digital subtraction angiography (DSA), and so the use of spiral CT is recommended instead [17, 18]. CTA can provide information about the size, number, location, and extent of PAAs and PAPs. This information is essential for treatment planning.

Multidetector CT (MDCT) has been found to provide rapid, accurate assessment of the cause and consequences of hemorrhage into the airway; this information can also be used to guide management. Research has also suggested that contrast-enhanced MDCT is as accurate as bronchoscopy, with high sensitivity for detection of the location of extravasation and underlying pathology. Appropriately timed contrast during MDCT for the pulmonary circulation can be equivalent to a chest CTA. CTA requires peak arterial or venous enhancement, reconstructions/reformats, but these are not required. For the general workup of MH, the ACR Appropriateness Criteria ranked chest radiography and bronchial arteriography with or without embolization with 9 and 8, respectively. This indicates that both radiologic protocols are usually appropriate [16].

Below, is a summary of the advantages and disadvantages of the relevant imaging modalities used to further study patients with hemoptysis (Table 3). This table also includes numerical annotations for representative figures (Fig. 2a–d).

## Management

### Surgery vs endovascular approach

Previously, these pathologies were mainly surgically treated with aneurysmectomy, lobectomy, or ligation [11]. However, patient-specific limitations remain. Patients with a poor Eastern Cooperative Oncology Group status, tenuous medical state, or poor respiratory function may not be surgical candidates or may not be able to tolerate a further decrease in function from lobectomy. Surgical resection under general anesthesia also may not be possible in patients with MH without significant risk to the airway. Surgical resection is also not recommended for patients with severe pulmonary hypertension [5, 19–21]. Therefore, selective endovascular transcatheter embolization of the involved branch is the preferred alternative, as this treatment method preserves residual pulmonary function with fewer complications [22–24]. To date, no large studies have compared success rate of surgical versus endovascular management.

There are no official guidelines regarding which indications should be treated with surgical or endovascular approaches. In fact, there is conflicting information regarding how aneurysm size and location and degree of symptoms should be factored into these treatment decisions [11, 25]. Some centers may stabilize the aneurysm via endovascular approaches and if the risk of rebleeding is high, will later surgically resect [26]. Of note, there are no absolute contraindications to the endovascular approach.

### Endovascular management

Endovascular technique should be based on the location of the aneurysm and the ability to be superselective while minimizing the risk of rupture [25–28]. Specifically, the most common choice of access site, would be via femoral access. Access from the internal jugular vein or subclavian vein does not optimize “directional purchase” of the catheter system; radial and brachial access provide this purchase, but catheter management near the pulmonary trunk requires the difficult navigation of sharp angles. Therefore, femoral access is preferred. In the setting of a recent catheter-based procedure, use of a different access site should be considered to reduce the risk of thrombosis.

### Endovascular procedure

The most commonly described procedural technique for these patients uses a coaxial microcatheter system (Renegade, Boston Scientific, Natick, Mass; or Progreat, Terumo, Tokyo, Japan). The first steps usually involve localization via CTA and investigation via arteriography with intention to treat. A pigtail catheter is placed at the bifurcation of the pulmonary trunk to delineate the vascular anatomy. Because the goal is to restore laminar flow, embolic agents must be selected to eliminate aneurysmal pathology. Interestingly, there is no standardization or formal recommendation regarding the type of embolic agent to be used for this endovascular procedure. The two most commonly used agents are coils and glue. Other options include plugs, covered stents, and gelatin particles [5, 25, 29]. Covered stents are the alternative option when preserving distal blood flow

**Table 3** Advantages and disadvantages of imaging modalities for patient with hemoptysis

| Imaging modality | Pros | Cons |
|---|---|---|
| Radiography (Fig. 2a) | Minimal radiation | May not be detected, variable sensitivity |
| MDCT (Fig. 2b) | High sensitivity for detection of the location of extravasation and underlying pathology (comparable if not better than bronchoscopy) | Radiation; no opportunity for intervention |
| CTA | Identifies all aspects of an aneurysmal pathology that is meaningful in guiding treatment<br>Optimal for non-acute patients | Radiation; no opportunity for intervention<br>Can identify slow flow (Type D PAP) |
| Arteriography with intention for embolization (Fig. 2c, d) | Opportunity for intervention; immediate workup for patients with MH | Minimally invasive endovascular technique with radiation |

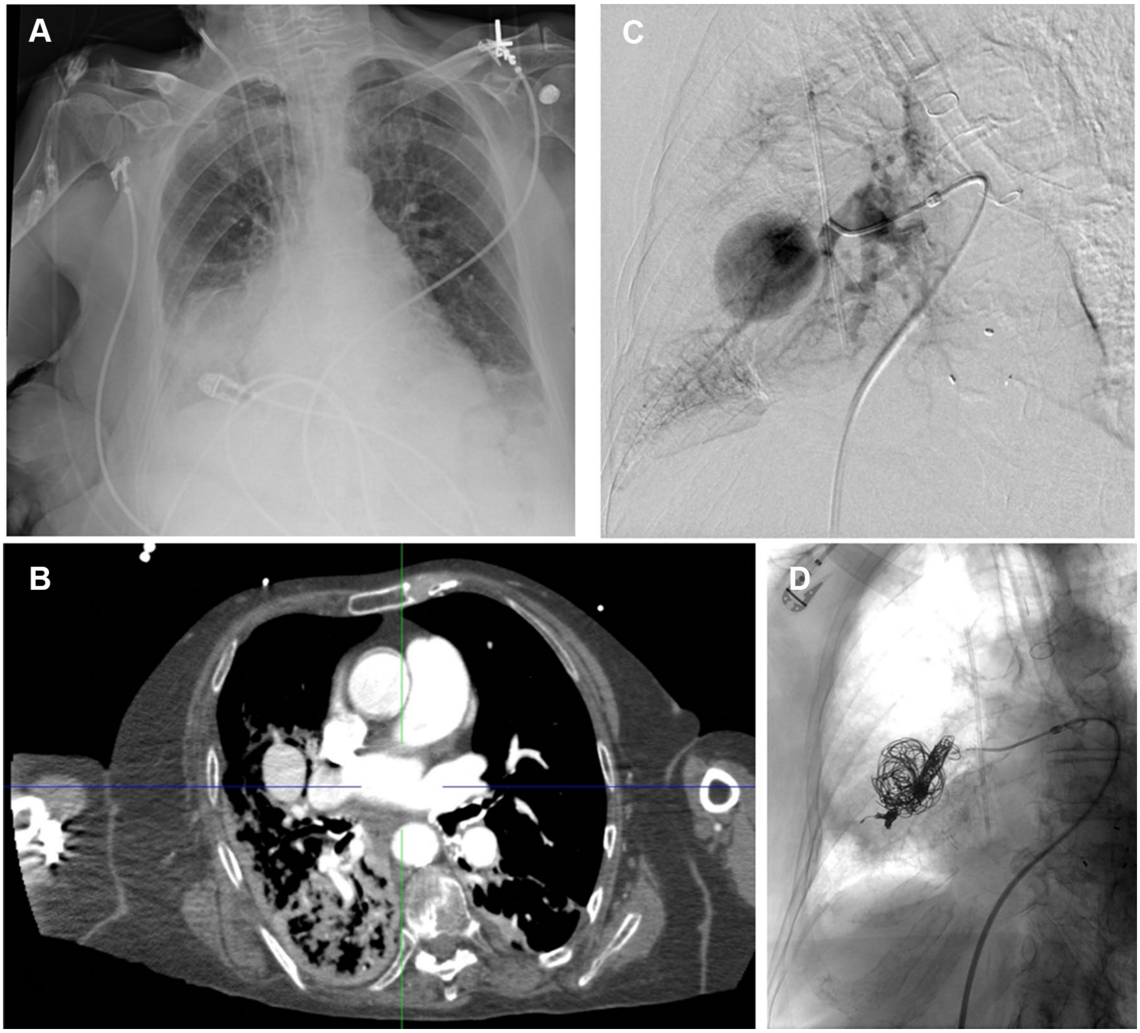


**Fig. 2** Imaging of a 75-year old woman with pulmonary aneurysm that necessitated coil embolization. **a** Chest X-ray does not clearly demonstrate the aneurysm. **b** CT with contrast clearly demonstrates aneurysm with blood supply. **c** Angiography provides a clear understanding of the vascular anatomy. **d** Aneurysm post-embolization with coils

is desired (Fig. 3). In other words, the selection of embolic agent is a regional choice and does not seem to correlate with underlying pathology; no consensus has been reached regarding optimal agent choice.

## Choice of embolic material

Selective transcatheter embolization of feeding arteries can be performed with steel- or platinum-based coils. Tornado coils and/or coils with attached cotton strands can also be used. Most operators use a controlled coil detachment technique. It is not uncommon to use multiple coils to fill a saccular aneurysm. However, if the case is emergent and the risk of rupture is high, stasis may be achieved more quickly with use of an endovascular plug (Amplatzer, AGA Medical, Plymouth MN; or Microvascular Plug, Covidien, Dublin, Ireland), as a plug is larger than a coil and can fill a saccular area with a single deployment. Of note, if the aneurysm is more fusiform in shape, a covered stent may be more appropriate [5, 24, 29].

Another popular option is the use of *n*-butyl cyanoacrylate (NBCA) glue for embolization. This material, which is nonradiopaque, provides efficient embolization. The glue is a liquid monomeric formula that becomes a solid long-chain polymer on contact with anion substances. When using this embolic agent, the operator must work quickly and generate an ion-free environment within the catheter system. Combining the acrylic glue with ethiodized oil helps to facilitate the speed and flow of the material as it traverses the catheter system and is placed into the aneurysmal sac. Operators may find that adding tantalum to make the glue radiopaque increases their confidence in procedural success. Histologically, this has been shown to trigger a more intense inflammatory reaction from surrounding tissues than other

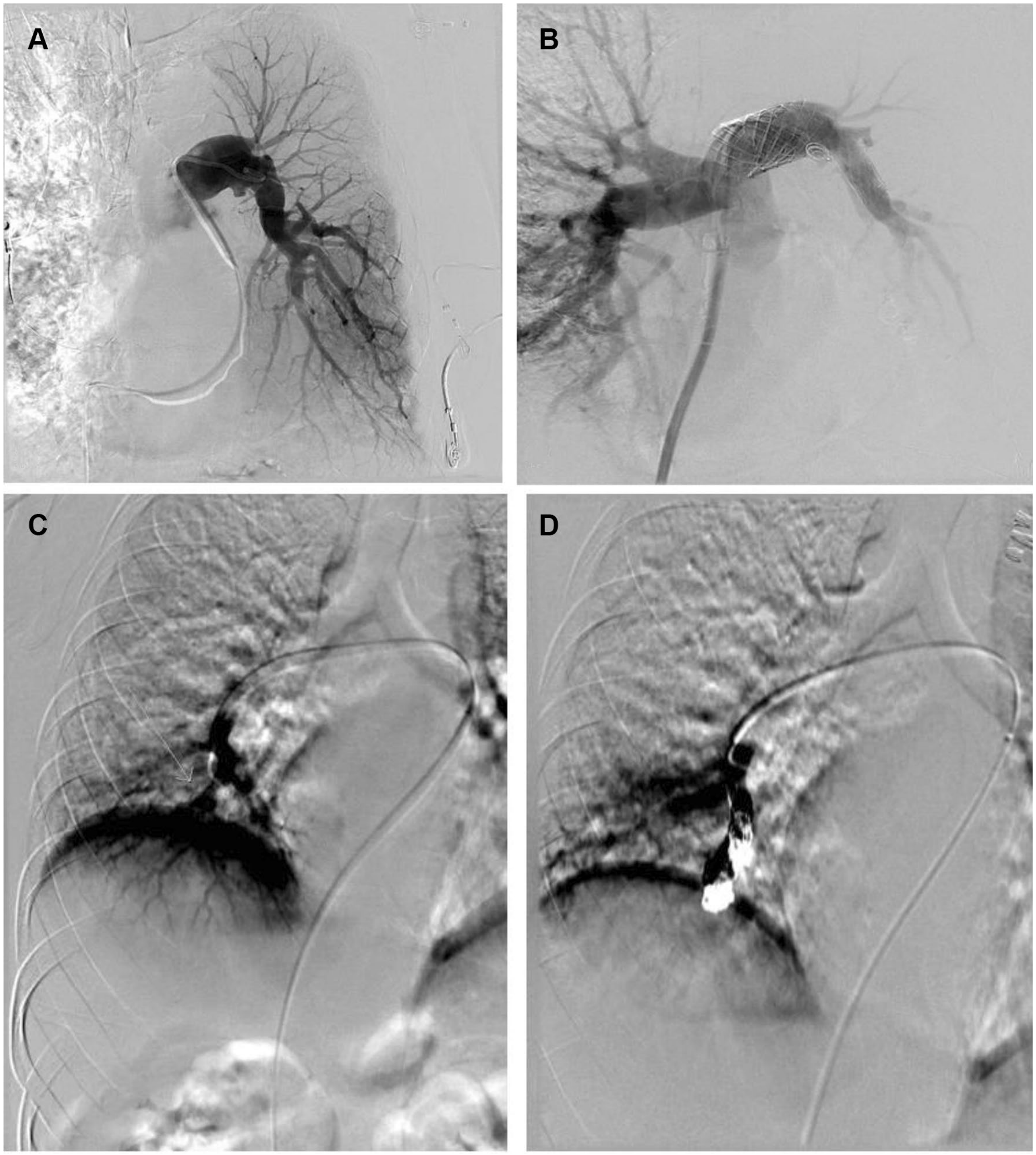


**Fig. 3** Examples of pre- and post- treatment using covered wall stent (**a, b**) and coil embolization (**c, d**)

embolic agents. The challenge of using this agent is timing of polymerization: if it is too quick, the agent will polymerize in the catheter system; if it is too slow, the agent can embolize distally [29].

Embolic agents are first deployed more distally to block outflow and are then delivered more proximally as the catheter system is pulled back, per standard embolization technique. Achieving full occlusion of distal flow before traveling proximally is essential. To minimize distal ischemia, transient balloon occlusion across the neck of the aneurysm may be considered. The classic alternative is the use of a scaffolding technique in which a larger coil is placed distal to the aneurysmal sac as an anchor that allows for smaller coils to be packed more proximally, later to be sealed with another larger coil [29].

## Complications of endovascular treatment

In terms of complications and subsequent management, it is difficult to quantify conversion-to-open rates. Given the high mortality of these pathologies, a failure in endovascular technique may not always be able to proceed to a surgical approach. The risks of recanalization and migration are generally related to the choice of embolic agent; the presence of complications, degree of postembolization syndrome, and length of hospital stay are more dependent on the underlying disease process than on the technical success of the procedure.

### Follow-up, post-treatment

Confirmation of effective treatment can be performed immediately after embolization with arteriography or venography. The use of noninterventional follow-up with CTA after the procedure depends on the size, location, and stability of the aneurysm. These factors also determine the timeframe for the initial follow-up, which can occur before discharge or even up to 3 months after the procedure. Studies have reported a high re-intervention rate (approximately 50%) in these patients [24]. Overall, reports of procedural success rates for surgery versus angiographic techniques have been mixed; however, multiple studies have shown control and successful stabilization of PAA and PAP, with endovascular interventions.

## Conclusion

Given the high mortality rates associated with PAA and PAP, early diagnosis and treatment of these conditions are essential. Endovascular intervention has been repeatedly shown to be a well-tolerated alternative to surgical management. Ultimately, if an intervention is desired, a conversation involving interventional radiologists, vascular surgeons, and/or cardiothoracic surgeons is warranted to weigh physician experience and skillsets for the safety of the patient.

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### Compliance with ethical standards

**Conflict of interest** There is no conflict of interest or industry support related to this project.

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