Introduction

Aneurysms and pseudoaneurysms are rare abnormalities of the pulmonary arteries. While their incidence is low, they represent potentially life-threatening conditions and can present a challenge for prompt diagnosis and treatment. An aneurysm of the pulmonary artery is defined as focal dilatation beyond maximum normal diameter (1). On computed tomography, a normal adult main pulmonary artery measures up to 29 mm in diameter and an interlobar pulmonary artery, 17 mm. A true aneurysm is defined as focal dilatation of an artery involving all three layers of the vascular wall—tunica intima, tunica media, and tunica adventitia. A pseudoaneurysm, by contrast, does not involve all three layers and thus poses a higher risk of rupture. A pulmonary artery pseudoaneurysm (PAPA) is a rare and potentially life-threatening disease characterized by focal saccular outpouching of a pulmonary artery representing a contained rupture of that artery (2). The mortality rate associated with the rupture of a pulmonary artery aneurysm (PAA) or PAPA has been reported from 50–100%; death is secondary to aspiration and asphyxia after intrapulmonary hemorrhage (3-6). PAA can also lead to dissection of the pulmonary artery and sudden cardiac death (2,7). Therefore, early diagnosis and treatment are critical for patient survival and optimal outcomes.

Clinical manifestations

Clinical manifestations for both PAA and PAPA are nonspecific and can be seen in many other conditions. In addition, patients with PAA and PAPA may remain asymptomatic (8). Reported symptoms include hemoptysis, shortness of breath, chest pain, palpitations or syncopal episodes (7-10). Additional symptoms attributed to extrinsic bronchial compression by a large PAA or PAPA may include cough, worsening dyspnea, cyanosis or pneumonia (10-12).

Etiology

PAA may be congenital or acquired. In an early autopsy study, the incidence of PAA was reported to be 1:14,000 with most occurring in the main pulmonary artery (13). PAA are more commonly associated with congenital
anomalies and PAPA are more commonly associated with acquired etiologies. Overall, most PAA and PAPA are caused by acquired etiologies such as trauma, iatrogenic injury, infection and Behcet’s disease (1). Congenital conditions associated with PAA and PAPA are listed in Table 1.

Congenital heart disease is the most common congenital anomaly associated with PAA (13-15). The postulated pathophysiology involves altered flow dynamics and increased hemodynamic sheer stress on the vessel wall, most commonly caused by left-to-right shunts (13-15). In decreasing order, the most common congenital cardiac anomalies associated with PAA include patent ductus arteriosus, ventricular septal defect, atrial septal defect, hypoplastic aortic valve and bicuspid aortic valve (13-15). Other congenital heart defects or valvular deficiencies such as pulmonary valve stenosis (Figure 1) have also been associated with PAA (1).

Another category of congenital conditions associated with PAA include diseases affecting connective tissue and vessels. Such conditions include Ehlers-Danlos syndrome, Marfan syndrome and cystic medial necrosis (9,16). As in arterial aneurysms throughout the body, pulmonary arteries and aneurysms are also subject to LaPlace’s law which states that arterial wall tension is proportional to the vessel radius at a given blood pressure. This suggests that, particularly in weakened arterial walls, larger aneurysmal arteries experience increased wall tension as they approach rupture.

As stated earlier, acquired etiologies of PAA and PAPA are more common than congenital etiologies. Acquired conditions of PAA and PAPA are listed in Table 2.

Acquired bacterial or fungal infections can cause mycotic pseudoaneurysms and less commonly aneurysms due to their ability to destroy or alter vessel walls. In patients with advanced syphilis, aneurysm formation most commonly occurs in the large pulmonary arteries with destruction occurring at the level of the vasa vasorum (17). Patients with advanced tuberculosis (TB) are at high risk of pseudoaneurysm formation in the intraparenchymal pulmonary arteries (18). These aneurysms known as Rasmussen aneurysms are peripheral PAs which occur due to erosion of a peripheral pulmonary artery branch (19).

### Table 1 Congenital conditions associated with pulmonary artery aneurysms and pseudoaneurysms

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>Heart defects</td>
<td>Persistent ductus arteriosus</td>
</tr>
<tr>
<td></td>
<td>Ventricular septal defects</td>
</tr>
<tr>
<td></td>
<td>Atrial septal defects</td>
</tr>
<tr>
<td></td>
<td>Hypoplastic aortic valve</td>
</tr>
<tr>
<td></td>
<td>Bicuspid aortic valve</td>
</tr>
<tr>
<td></td>
<td>Pulmonary valve stenosis</td>
</tr>
<tr>
<td></td>
<td>Pulmonary regurgitation</td>
</tr>
<tr>
<td></td>
<td>Absent pulmonary valve</td>
</tr>
<tr>
<td>Connective tissue abnormality</td>
<td>Ehlers-Danlos syndrome</td>
</tr>
<tr>
<td></td>
<td>Marfan syndrome</td>
</tr>
<tr>
<td></td>
<td>Cystic medial necrosis</td>
</tr>
</tbody>
</table>

Figure 1 Main pulmonary artery aneurysm secondary to pulmonary valve stenosis. Axial and sagittal CT images demonstrate a large 5.5 cm aneurysm (A,B, red arrows). This abnormality is not amenable to endovascular repair.
These Rasmussen aneurysms typically involve the upper lobes in the setting of reactivation tuberculosis (1). In the post-antibiotic era, syphilis and TB, once problematic and more prevalent pathogens, have seen a dramatic decrease in incidence (8) whereas the more common infectious agents are now taking the spotlight.

Other infectious entities which can lead to PAA or PAPA include pyogenic bacteria (1) and less commonly viral (20) and fungal infections. In the setting of pneumonia, destruction of the vessel wall occurs from the outer wall to inner lumen as a result of invasion from the neighboring consolidation (21). In endocarditis, typically seen in the setting of IV drug abuse, the patients are at risk for septic emboli which can cause aneurysms both centrally and peripherally (22)—a key finding on CT to aid diagnosis. The proposed mechanism by which aneurysms form in endocarditis is endovascular seeding of the lumen by septic emboli (14,22). It is therefore suggested that a PAPA secondary to endocarditis is not necessarily related to an adjacent lung consolidation, which may also be present concomitantly (8).

Pulmonary artery hypertension (PAH) has been widely implicated as a cause of PAA (9). The mechanism of aneurysm formation is similar to other areas of the body involving increased arterial wall stress (14). Numerous etiologies of PAH exist which can ultimately result in aneurysm formation. International consensus by the fifth World Symposium on Pulmonary Hypertension has classified these into five groups (23). These groups are listed in Table 3.

Pulmonary thromboembolic disease has been implicated as a cause of PAA or PAPA and two mechanisms have been

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Acquired conditions associated with pulmonary artery aneurysms and pseudoaneurysms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Etiology</td>
<td>Example</td>
</tr>
</tbody>
</table>
| Infectious | Syphilis  
Tuberculosis  
Pyogenic bacteria  
Endocarditis, septic embolism  
Bacterial, viral or fungal pneumonia |
| Vasculitis | Behcet’s disease  
Hughes-Stovin syndrome |
| Pulmonary arterial hypertension | Five groups (Table 3) |
| Chronic pulmonary embolism | – |
| Acute or chronic inflammatory lung disease | Bronchiectasis and pulmonary fibrosis (8)  
Interstitial lung diseases  
COPD |
| Neoplasm | Primary lung cancer  
Pulmonary metastasis |
| Iatrogenic | Cardiac surgery  
Chest tube placement  
Lung biopsy  
Pulmonary artery catheter placement  
Pulmonary artery angiography  
Lung resection  
Lung ablation  
Prior radiation to chest |
| Trauma | Gun shot  
Stab wound |
| Idiopathic | – |

| COPD, chronic obstructive pulmonary disease. |

<table>
<thead>
<tr>
<th>Table 3</th>
<th>Classification of PAH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Category</td>
<td>Etiology</td>
</tr>
</tbody>
</table>
| Group 1 | Idiopathic  
Heritable  
Drug and toxin-induced  
Association with connective tissue disease  
HIV infection  
Portal hypertension  
Congenital heart disease and schistosomiasis  
Pulmonary veno-occlusive disease and/or pulmonary capillary hemangiomatosis  
Persistent pulmonary hypertension of the newborn |
| Group 2 | Secondary to left heart disease |
| Group 3 | Secondary to lung diseases and/or hypoxia |
| Group 4 | Chronic thromboembolic pulmonary hypertension (CTEPH) |
| Group 5 | Unclear multifactorial mechanism |

PAH, pulmonary artery hypertension.
of acute or chronic inflammatory lung disease (38). In
chronic pulmonary embolism is another relatively common
cause of PAA. Aneurysms which arise from this condition
tend to be associated with mural thickening, webs or
intramural thrombi which can calcify (1).

PAPAs may develop as a result of primary lung cancer or
pulmonary metastatic disease. Though this phenomenon
is rare, multiple cases have been reported (1,8,25,26). The
proposed mechanism of PAPA formation involves direct
tumoral invasion and vessel wall erosion (25-27).

Another less common cause of PAA or PAPA is vasculitis.
Of the vasculitides, Behcet’s disease, has been reported to
be the most frequent cause of pulmonary aneurysms (28).
Behcet’s disease is a chronic multisystem vasculitis
hallmarked by oral and genital ulcers and uveitis (29). It is
most prevalent in the Middle East and Asia and may result
in aneurysms typically of the right lower lobe with recurrent
thrombosis and inflammation (30). It has been reported
that the apparent PAAs that arise from Behcet’s disease
are in fact PAPA and caused by complications of vasculitis
and transmural necrosis (31). Hughes-Stovin syndrome
is a rare condition which presents as a combination of
systemic venous thrombosis and pulmonary aneurysms (32)
and is suggested to be a variant of Bechet’s disease (32,33).

As in Behcet’s disease, arterial aneurysms form secondary
to obliterative endoarteritis of the vasa vasorum thereby
compromising the integrity of the vessel wall (32).
Therefore, both of these vasculitides are associated with
PAPA rather than PAA (32).

Iatrogenic and traumatic causes have increased in
incidence as a source of PAA and PAPA (Figures 2,3).
Numerous cases have been reported citing such causes as
Swan-Ganz catheter placement, cardiothoracic surgery,
chest tube placement, catheter-directed angiography, lung
biopsies and even percutaneous ablation (1,3,8,34-36).
The resulting pulmonary vascular injuries would more likely form
PAPA. PAPA formation has also been reported in cases of
penetrating trauma such as stab or gun-shot wounds (8,37).

Several other less common causes of PAA or PAPA have
been described. PAA or PAPAs have developed as a result of
acute or chronic inflammatory lung disease (38). In
their retrospective review, Chen et al observed one patient
who was noted to have a PAPA associated with traction
bronchiectasis in the setting of pulmonary fibrosis (8). The
exact mechanism of vascular injury has not been described.

Atherosclerosis has also been reported as a rare cause of
PAA as a degenerative vascular disease (14).

Idiopathic PAA is a rare and enigmatic diagnosis in the
absence of other causes. In order to better characterize this
classification, four pathological criteria have been defined: (I)
simple dilatation of the pulmonary trunk, with or without
involvement of the rest of the arterial tree; (II) absence of
abnormal intra- or extracardiac shunts; (III) absence of
chronic cardiac or pulmonary disease, either clinically or at
autopsy; (IV) absence of arterial disease, such as syphilis or
more than minimal atheromatosis or arteriolar sclerosis (39).

Methods of diagnosis

The mainstay of imaging for both detection and follow-
up of PAA and PAPA remains computed tomography
angiography (CTA). Given the nonspecific symptoms of
PAA and PAPA, CTA focus can be placed on either the
pulmonary arterial system or the aorta and bronchial artery
system. Multi-detector row CTA performed with bolus
tracking over the descending aorta, along with coronal and
sagittal multiplanar and maximum-intensity projection
(MIP) reformatted images, has been reported as an imaging
protocol for hemoptysis (40). Other CTA imaging protocols
call for bolus tracking over the main pulmonary artery (41)
to optimize pulmonary artery opacification. The advantage
of CTA is that it allows for the assessment of presence,
size, location and characteristics including saccular or
fusiform aneurysm type (42). Equally as important, it can
provide information regarding the underlying etiology of an
aneurysm in that additional clues may be found in the lungs,
heart or mediastinal structures. Where available, three-
dimensional and advanced post-image processing software
such as AquariusNET (TeraRecon, Foster City, CA, USA)
can prove to be extremely valuable in both detection as
well as treatment planning. For follow-up post-treatment
imaging, a non-contrast CT scan of the chest may
immediately precede a CTA to differentiate calcifications
or embolic materials from persistent contrast opacification.
Dual-energy CT (DECT) is an advanced CT technique
that allows for better material differentiation, such as
differentiating iodine from other hyperdense materials
and can also overcome some artifacts thus improving CT
pulmonary angiogram scan quality at relatively reduced
contrast and radiation dose (43).

Catheter-directed angiography has been considered the
gold-standard for diagnosis of PAA and PAPA. This allows
for the determination of the extent of vascular involvement
and an assessment of right-sided cardiac pressures (44). In addition, simultaneous endovascular treatments can be performed. Aside from being invasive, a major limitation of pulmonary angiography is that it does not yield information regarding extra-luminal structures, which can be essential to determine etiology. One solution may be cone-beam CT, which can provide additional benefits to the imaging arm and can be a very useful tool for planning appropriate endovascular therapy. The role of non-invasive imaging modalities such as CT and MRI, however, is expanding as the technology for image gathering and reconstruction continues to evolve (42).

Magnetic resonance imaging (MRI), while not as commonly used as CTA for evaluating the pulmonary arteries, is a viable alternative where CTA cannot be employed (i.e., allergy to iodinated contrast material, renal insufficiency). Use of MRI to evaluate the pulmonary arterial system has been documented and described (45,46). T1-weighted images have been shown to adequately demonstrate pseudoaneurysms. Fast spin echo (FSE) and gradient echo imaging are useful in the morphologic evaluation of pulmonary vasculature from the main pulmonary trunk to the subsegmental level (45). When possible, gadolinium-enhanced magnetic resonance
angiography improves visualization and evaluation of the subsegmental pulmonary artery branches (47). With regards to the pulmonary arterial system, one distinct advantage of MRI over CTA is that it can identify and elaborate on arterial wall thickening in connective tissue disease. Furthermore, it can provide information regarding blood flow direction in cases of post-stenotic dilatation due to pulmonary valve disease, which is important for early preventative action or treatment (1).

**Imaging findings**

CT appearances of PAA and PAPA can vary greatly in their number, location, morphology and associated findings. Differentiating PAA and PAPA may not be possible on imaging and requires correlation with clinical history and laboratory findings.

PAA or PAPA can be seen anywhere along the pulmonary arterial tree. Depending on the etiology, PAA or PAPA may...
have a predilection for central or peripheral arteries. In one of the largest retrospective studies reviewing CTAs in patients with hemoptysis and PAPA, PAPAs showed a strong predilection for the peripheral pulmonary artery branches with 63% occurring in the periphery at the subsegmental pulmonary arterial level (8). The authors also noted that 83% of the PAPA discovered were observed to be solitary occurrences (8). Additional findings associated with PAA or PAPA largely depend on the underlying etiology. As suggested by Chen et al., hemorrhage surrounding a PAPA is a strong predictor of acute trauma (8). Multiple PAA or PAPA, were typically observed in the setting of endocarditis or lung metastases (8). In the pulmonary sequelae of endocarditis, a cavitary lesion may be identified adjacent to the PAPA or more peripherally. In pulmonary metastatic disease, a consolidation or mass would likely accompany a PAPA. Given that the majority of tumor emboli are microscopic, PAPAs typically involve the subsegmental pulmonary arteries and arterioles (42).

**Treatment**

Once a PAA or PAPA is diagnosed, the next challenge involves determining the appropriate treatment. As highlighted earlier, multiple etiologies exist for both entities and treatment must be tailored to the underlying cause or causes as well as choosing the least invasive procedure while achieving durable results.

Neither a consensus to treat PAA based on size criteria nor treatment guidelines delineating the roles of medical or procedural disciplines currently exist. PAA or PAPA can occur at various locations and take on different forms. In non-urgent, asymptomatic patients, conservative management of a PAA can be considered to address treatable underlying etiologies; the treatment of which is beyond the scope of this review. Medical treatment alone in a more complex case may be inadequate in preventing growth or rupture of a PAA or PAPA and therefore endovascular or surgical therapies may be indicated.

Several surgical techniques and procedures exist to treat PAAs. These include aneurysmorrhaphy, lobectomy, bilobectomy, aneurysmectomy and pneumonectomy (5). Surgical resection, however, carries high risks, especially in patients with severe pulmonary hypertension (48). When feasible, endovascular treatment can be offered as the first line therapy with the advantage of less morbidity and mortality. Although specific guidelines do not exist for endovascular versus surgical treatment of PAA, many case reports support first line consideration of endovascular treatment when feasible. Endovascular therapy may best serve saccular PAA or PAPA, both in the central and peripheral pulmonary arteries as suggested by published case reports. Alternatively, fusiform aneurysms of the peripheral pulmonary arteries may be treated endovascularly where pulmonary function and adequate reserve is available, similar to peripheral pulmonary arteriovenous malformations. Central fusiform aneurysms, however, require surgical management.

In treating PAA and PAPA, one important variable that should be addressed is a bronchopulmonary artery shunt with retrograde flow. In this event, a PAA or PAPA may not be demonstrated on pulmonary angiography due to systemic artery-to-pulmonary artery shunting but can be seen on bronchial or non-bronchial systemic angiograms (38). In these cases, embolization proximal to the PAA or PAPA from the pulmonary arteries may be insufficient.

**Coil embolization of arterial aneurysms or pseudoaneurysms** throughout the body is a widely accepted minimally invasive therapy (14,26,49). Intra-saccular embolization with coils has the advantage of preserving the pulmonary arteries distal to a PAA or PAPA while sparing pulmonary function (50) but should be performed with great caution due to the risk of rupture, especially in the case of PAPA. When intra-saccular embolization either carries an increased risk of rupture or is not feasible/incomplete, embolization proximal and distal to the PAA or PAPA neck can be performed (Figures 2-5). As mentioned earlier, proximal embolization alone may still allow perfusion of the aneurysm sac via bronchopulmonary anastomoses distal to a proximal embolization (51). For this reason, when embolizing the pulmonary artery supplying the PAA or PAPA, it is imperative to best rule-out a shunting phenomenon.

As an alternative to coils, the use of vascular plugs (Figure 6) in rare cases have been described (52,53). As with coil embolization, a bronchopulmonary shunt should be ruled out prior to performing a pulmonary artery embolization proximal to a PAA or PAPA.

Stent-assisted coil embolization is a modified form of coil embolization whereby embolization of an aneurysm is performed through the interstice of a bare-metal stent to maintain vascular patency. Though this technique has not been described in the pulmonary arterial system, it is a viable treatment option which has been described and accepted in other areas of the body for wide-necked aneurysms (54-56) and in many cases, can be applied
Figure 4 Necrotizing pneumonia causing a left lower lobe pulmonary artery pseudoaneurysm. Axial and coronal CT images demonstrate a slightly dilated subsegmental left lower lobe pulmonary artery branch in a region of consolidation (A,B, red arrows). Angiographic images from a distal segmental pulmonary artery branch of the left lower lobe demonstrating two focal fusiform aneurysms (C, red arrow) of a distal subsegmental branch which were embolized with coils (D).

safely in the pulmonary arteries. A distinct advantage of this technique is the preservation of flow distal to the aneurysm sac.

Although uncommon, the use of stent grafts in the pulmonary arteries has been reported; effectively excluding a pseudoaneurysm sac (57-59) while maintaining vascular patency. Rare cases of successful glue embolization of PAPA with n-butyl cyanoacrylate (NBCA) have also been described in the literature (31,60,61). A novel technique of transcatheter embolization via the pulmonary artery with an NBCA and iodized oil mixture while employing balloon occlusion has been described to address the presence of a bronchopulmonary shunt (38).

A single earlier case of balloon embolization of a mycotic PAA has also been reported (62).

Although endovascular treatments of PAA and PAPA carry fewer inherent risks compared to surgery, they maintain similar risks to other endovascular embolization procedures throughout the body. These risks include contrast induced nephropathy, non-target embolization, arterial dissection, arterial thrombosis and partial or complete end-organ infarction.
Figure 5 Pulmonary artery pseudoaneurysm after lung nodule biopsy. Axial CT image demonstrates a spiculated lung nodule of the right upper lobe (A, yellow arrow). Axial and coronal CT post-biopsy images demonstrate a small pseudoaneurysm arising from a right upper lobe subsegmental pulmonary artery (B,C, red arrows). Angiogram from a distal posterior segment right upper lobe pulmonary artery demonstrates a small focal saccular pseudoaneurysm arising from a subsegmental branch (D, red arrow) which was embolized with coils (E).

Conclusions

Aneurysms and pseudoaneurysms of the pulmonary arteries are rare entities and often not considered in many clinical situations. CT pulmonary angiogram is the imaging modality of choice for diagnosis of PAAs and pseudoaneurysms. Currently, there is no large study comparing endovascular and surgical treatments. Additionally, consensus guidelines for treatment by either means have also not been established. As many case reports demonstrate, however, when indicated, endovascular therapy is the favored first-line treatment considering its decreased risk profile where treatment by coil embolization or other embolic devices is feasible. Before any treatment is
Figure 6 Multiple, bilateral pulmonary artery pseudoaneurysms from pulmonary artery vasculitis (Hugh-Stovin Syndrome) causing massive hemoptysis. Coronal CT image demonstrates several pseudoaneurysms of the right lung (A). Angiogram from the right main pulmonary artery confirmed the presence of multiple pulmonary artery pseudoaneurysms (B). Embolization of four segmental pulmonary arteries supplying four pseudoaneurysms with Amplatzer type II and type IV vascular plugs (C, red arrows) with successful exclusion of the pseudoaneurysms. Resultant peripheral perfusion defects of the right lower lobe are identified.

performed, however, managing these patients should be a collaborative multidisciplinary effort between the intensivist, pulmonologist, interventional radiologist, thoracic surgeon, and when applicable, the anesthesiologist. Given the lack of consensus treatment guidelines, a multidisciplinary plan of action is necessary to increase survival while minimizing procedure-related morbidities and mortalities.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

References

40. Bruzzi JF, Rémy-Jardin M, Delhaye D, et al. Multi-
Detector Row CT of Hemoptysis. Radiographics 2006;26:3-22.


