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# Reperfusion of Complex Pulmonary Arteriovenous Malformations After Embolization: Report of Three Cases

Pascal Lacombe,[1] Christine Lagrange,[1] Mostafa El Hajjam,[1] Thierry Chinet,[1] Jean-Pierre Pelage[2]

[1]Department of Radiology, Hopital Ambroise Paré, Boulogne, France
[2]Department of Respiratory Medicine, Hopital Ambroise Paré, Boulogne, France

## Abstract

The purpose of this report is to discuss the different mechanisms of reperfusion of pulmonary arteriovenous malformations (PAVMs) after embolization. Transcatheter embolotherapy is currently the first-line treatment of PAVMs to prevent neurologic complications or pulmonary hemorrhage. Initial good results can be expected but we report three cases of reperfusion of complex large PAVMs after coil embolization. After adequate embolization, reperfusion of PAVMs may occur by several mechanisms including recanalization of embolized arteries, recruitment of normal arterial branches, growth or enlargment and development of a systemic arterial supply.

**Key words:** Arteriovenous malformations, pulmonary—Arteriovenous malformations, therapeutic embolization—Reperfusion, embolotherapy

Percutaneous transcatheter embolotherapy is currently accepted as the treatment of choice to prevent neurologic complications or pulmonary hemorrhage in patients with pulmonary arteriovenous malformations (PAVMs) [1–9]. PAVMs are commonly associated with hereditary hemorrhagic telangiectasia (HHT), also known as Rendu-Osler-Weber disease [1–3, 10]. Regardless of the technique used, embolization using coils or detachable balloons is safe, well tolerated and associated with excellent symptomatic improvement [5, 11–14]. Reperfusion of accurately embolized PAVMs is considered as a rare event, predominantly affecting large PAVMs [15]. Conversely, contrast-enhanced magnetic resonance (MR) angiography detects a high incidence of persistent signal within treated PAVMs after embolotherapy [16]. This may be related to recanalization of the feeding vessel or reperfusion of the PAVM by an accessory artery or by systemic branches [13, 17, 18]. We report here three cases of reperfusion of complex large PAVMs after coil embolization and discuss the different mechanisms of reperfusion.

*Correspondence to:* Pascal Lacombe; *email:* pascal.lacombe@apr.ap_hop_paris.fr

## Case Reports

### *Case 1*

An 18-year-old man with HHT had an exercise intolerance, cyanosis, hippocratic nails and epistaxis. Spiral computed tomography (CT) demonstrated a large complex PAVM located in the right lower lobe (Fig. 1A). Both lungs were involved by diffuse small or tiny PAVMs whose afferent arteries had a diameter less than 2 mm. Three feeding pedicles of the complex PAVM were confirmed by selective pulmonary angiography. Embolization of all the feeding arteries was successfully performed with a 5 Fr vertebral catheter using 45 steel coils with diameters ranging from 12 to 3 mm (Mreye embolization coils, Cook, Bjaeverskov, Denmark). All coils were carefully packed in order to prevent recanalization (Fig. 1B). The catheter tip was advanced to a point beyond any normal branches and immediately proximal to the venous sac. Before embolization the supine $PaO_2$ was 47 mmHg on room air and the supine arterial oxygen saturation ($SaO_2$) was 83%. After embolization a transitory right pleural effusion was noticed.

At 18 months after embolization, the patient's clinical status had improved, but both $PaO_2$ and $SaO_2$ were suboptimal (68 mmHg and 93%, respectively). On follow-up spiral CT, reperfusion of the previously embolized PAVM was detected. In addition, significant enlargement of a small PAVM involving the middle lobe and a large inferior right phrenic artery were observed (Fig. 1C). The angiographic procedure was then repeated. Embolization of the recanalized complex PAVM and of the recently enlarged PAVM of the middle lobe was carried out using six coils. Selective catheterization of a large inferior phrenic artery demonstrated transpleural supply to the previously embolized PAVM

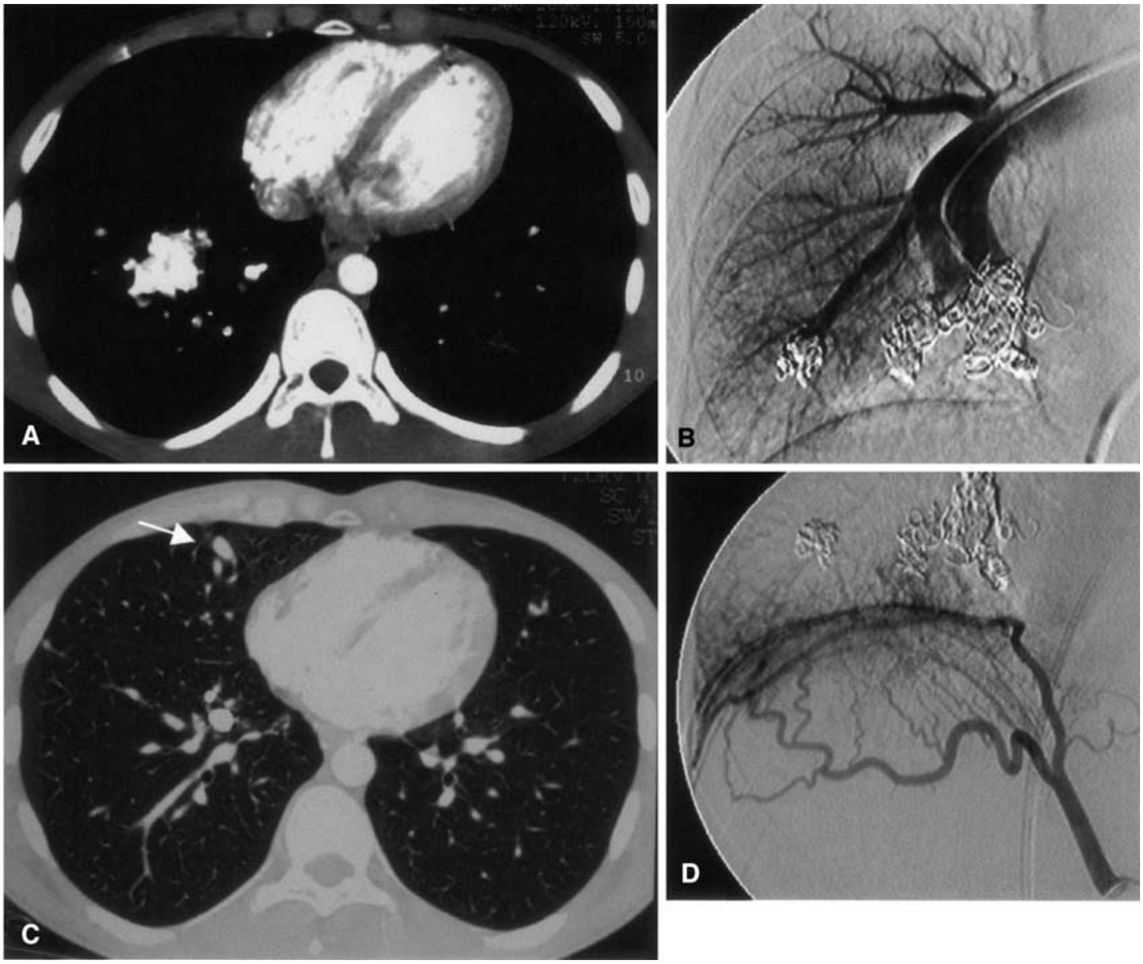


**Fig. 1.** Preprocedure CT, embolization procedure and follow-up CT in an 18-year-old man with HHT. **A** Spiral CT (mediastinal window settings) reveals a large PAVM located in the right lower lobe. **B** Postembolization angiogram reveals no filling of the PAVM with satisfactory coil packing. **C** Follow-up spiral CT (lung window settings) shows significant enlargement of a small PAVM involving the middle lobe (arrow). **D** Selective catheterization of a large inferior phrenic artery demonstrated transpleural supply to the previously embolized PAVM of the right lower lobe.

(Fig. 1D). No additional embolization was performed. After embolization, the $PO_2$ and $SaO_2$ increased to 82 mmHg and 98%, respectively. The patient is still asymptomatic 10 months after the second embolization and has not presented hemoptysis.

## *Case 2*

A 6-year-old boy had dyspnea, cyanosis and hippocratic nails. He presented a hypoxic syncope while riding his bicycle. His $PaO_2$ and $SaO_2$ on room air were 52 mmHg and 79%, respectively. Spiral CT demonstrated a single complex PAVM of the right lower lobe (Fig. 2A). Pulmonary angiography demonstrated a complex large PAVM involving four different pedicles (Fig. 2B). Embolization of all the feeding arteries was successfully performed with a 5 Fr right coronary catheter using a total of 32 coils with diameters ranging from 8 to 5 mm (Fig. 2C). At 3 months after embolization, his clinical status improved and his $PaO_2$ and $SaO_2$ increased to 99 mmHg and 93%, respectively.

At 12 months after embolization, incomplete regression of the previously embolized PAVM was noticed on spiral CT (Fig. 2D). A second embolization procedure was performed. Reperfusion of the PAVM was related to partial recanalization of the main feeding pedicle and recruitment of adjacent initially normal arteries (Fig. 2E,F). A total of 23 coils were used to ensure complete occlusion of the PAVM. After embolization, his $SaO_2$ increased from 93% to 98%. His clinical status improved with complete disappearance of cyanosis and hippocratic nails. With a 54 month follow-up, the patient remains asymptomatic. On contrast-enhanced spiral CT, permanent occlusion of PAVM was confirmed. Enlargement of bronchial and right inferior phrenic arteries was noticed but no embolization has been planned in this asymptomatic young boy.

## *Case 3*

A 22-year-old woman was admitted after a car accident caused by sudden syncope. On arrival, she was cyanotic and had hippocratic nails. She had never presented epistaxis and no telangiectasia was found. Her arterial saturation and $PaO_2$ on room air were 82% and 47 mmHg, respectively. She was diagnosed with two PAVMs: one

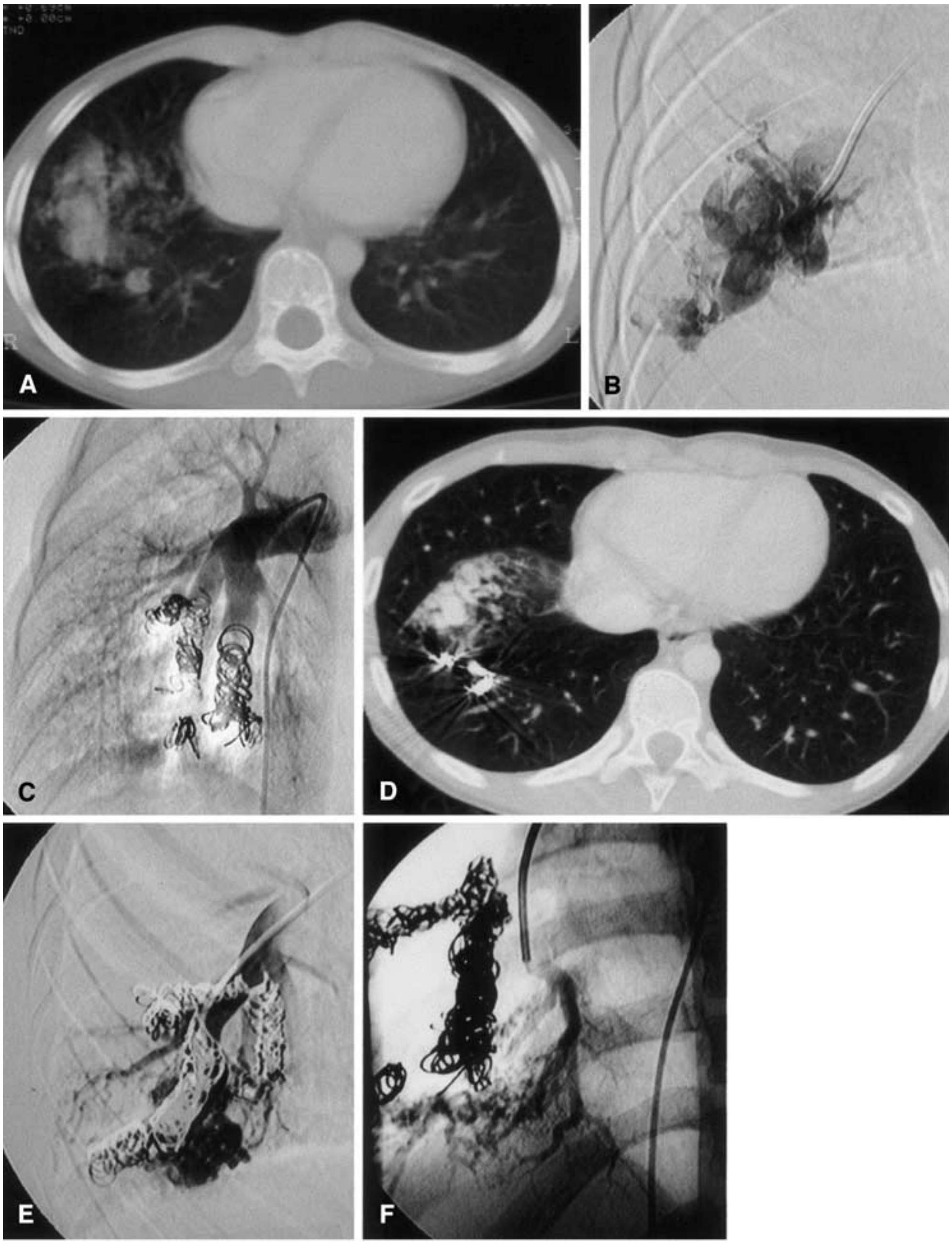

**Fig. 2.** Preprocedure CT, embolization procedure and follow-up CT in a 6-year-old boy with a single complex PAVM of the right lower lobe. **A** Spiral CT (lung window settings) demonstrates a large complex PAVM located in the right lower lobe. **B** Right pulmonary angiogram reveals a large complex PAVM of the right lower lobe. **C** Embolization of all the feeding arteries was successfully performed using a total of 32 coils. **D** Follow-up spiral CT at 12 months after embolization shows incomplete regression of the previously embolized PAVM. **E** During a second embolization procedure, obvious reperfusion of the PAVM was related to partial recanalization of the main feeding artery. **F** Recruitment of adjacent initially normal arteries is also visible.

large complex PAVM of the right lower lobe and one simple small PAVM of the left lower lobe (Fig. 3A). Large right bronchial arteries were also detected from the aortic arch to the level of the right inferior pulmonary vein (Fig. 3B). Pulmonary angiography demonstrated a complex PAVM involving multiple pedicles. Embolization of the six feeding arteries was successfully performed using 44 coils with diameters ranging from 5 to 3 mm. All coils were carefully packed in order to prevent recanalization. The simple PAVM of the left lower lobe was easily embolized using three coils (5 to 3 mm).

At 20 months after embolization, her clinical status improved and the $PaO_2$ and $SaO_2$ increased to 93 mmHg and 98%, respectively. On follow-up contrast-enhanced spiral CT, a localized reperfusion was identified within the large PAVM (Fig. 3C).

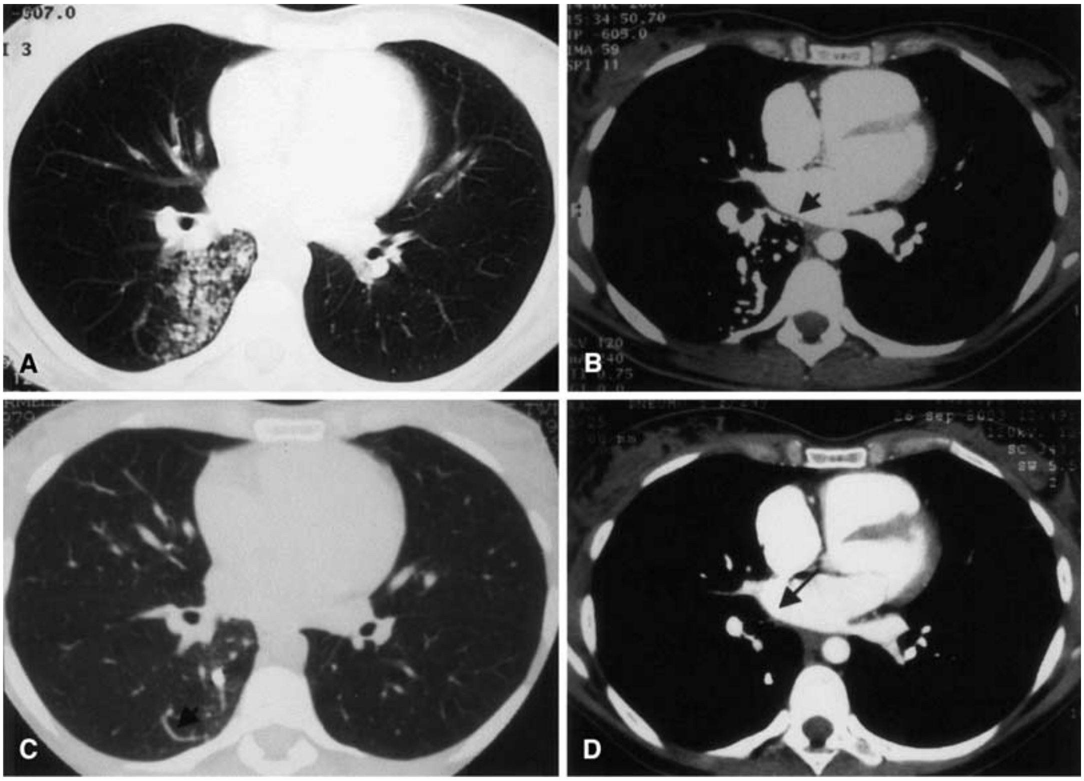


**Fig. 3.** Preprocedure and postembolization CT in a 22-year-old woman with two PAVMs. **A** Spiral CT (lung window settings) reveals a large complex PAVM situated in the right lower lobe. **B** Large right bronchial arteries are visible at the level of the right inferior pulmonary vein (arrow). **C** On follow-up contrast-enhanced spiral CT (mediastinal window settings), a localized reperfusion is identified within the large PAVM (arrow). **D** The diameter of the right bronchial arteries has decreased (arrows).

On the left side, enlargement of a small PAVM, not diagnosed initially, was noticed whereas the embolized PAVM had regressed. Surprisingly, the diameter of the bronchial arteries had decreased (Fig. 3D). The patient remains asymptomatic with disappearance of hippocratic nails and normal arterial blood gases and shunt studies.

## Discussion

PAVMs are potentially dangerous because of the risk of rupture, bleeding or paradoxical emboli resulting in stroke or cerebral abscess [1–3, 19]. PAVMs can be classified as either simple or complex [1]. A simple PAVM consists of single or multiple feeding arteries originating from a single segmental artery [1, 5, 20]. In complex PAVMs, feeding arteries originate from two or more segmental arteries [1, 5, 20]. In patients with diffuse or large PAVMs, neurologic symptoms are almost inevitable [1, 2]. Transcatheter embolization prevents stroke and neurologic damage and is usually indicated in the presence of feeding arteries larger than 3 mm in diameter [8, 20]. It is thought that the arterial occlusion of a simple PAVM is technically easier and less time-consuming than that of a complex type [20].

The method of embolization has been extensively described [5, 6, 8, 9]. In essence it involves catheterization of the feeding vessels to a malformation, advancement of the catheter tip to a point beyond any branches to normal lung and immediately proximal to the dilated venous portion, followed by arterial occlusion using coils or balloons [5, 8, 11–14]. When the feeding artery is too short to be safely embolized, occlusion of the aneurysmal sac of the PAVM itself should be considered [21, 22]. There are some differences in technique between centers, some catheterizing the feeding artery using 6 Fr or 7 Fr catheters, others performing catheterization mainly with a coaxial technique [5, 14, 22]. In addition, there is no consensus concerning the choice of embolization agent [5, 11–14, 22].

Embolization can be performed using detachable balloons or steel coils. Most groups favor steel or platinum coils as the primary embolization agent. The choice of a coil of a correct size is critical: if it is too small, the coil may pass through the venous portion of the PAVM into the systemic circulation with potential disastrous consequences [14], while if it is too large, the coil may cause occlusion of proximal normal pulmonary arterial branches or may elongate leading to recanalization [11]. After placement of the first coil, additional coils must be positioned until blood flow to the PAVM has ceased [5, 14]. Packing of smaller coils in the center of the first-placed coil is mandatory to prevent recanalization. If the number of coils is not sufficient recanalization may also occur because of insufficient thrombosis formation [17, 18, 23]. Despite an appropriately packed coil embolization, recanalization was observed in our three patients. Cure of the PAVM is usually defined by the involution (thrombosis and regression) of the aneurysm with a residual fibrous scar on spiral CT [24]. The rate of recanalization is probably underestimated if the follow-up of patients includes only chest radiography [7, 17, 24].

Accessory abnormal pulmonary artery branches to the PAVM can be missed during the initial evaluation with CT or even at the time of embolization because of preferential flow to the PAVM. Postprocedural pulmonary angiography may allow a better detection of accessory branches supplying the PAVM. Another potential mechanism explaining reperfusion of embolized PAVMs is the recruitment of normal branches adjacent to the PAVM, as illustrated in our second case [7]. Persistent perfusion of a complex PAVM was not a clinically relevant problem in our patient, but the long-term morbidity is unknown. However, some patients have suffered a stroke because of recanalized PAVMs [15].

Imaging follow-up of treated patients as well as physiologic evaluation should be performed in order to document involution of embolized PAVMs but also to detect growth or enlargement of small PAVMs as demonstrated in cases 1 and 3 [24]. Small PAVMs can over time reach the threshold size for complications. In most patients with diffuse PAVMs, improvement of dyspnea, oxygenation and shunt fraction is often not complete. The residual shunt is believed to represent the shunt through small PAVMs [1]. Small branches supplying the embolized PAVM may be also missed during follow-up CT evaluation, particularly in the absence of contrast enhancement or because of coil-related artifacts [24]. Even if clinical and radiological evaluation is necessary, oxygen saturation tests are equally important to predict recurrence. Conversely, contrast echocardiography is probably too sensitive and remains positive in the majority of patients with successful occlusion of all angiographically visible vessels [25, 26].

With MR perfusion imaging, the residual enhancement of a treated PAVM is frequent (58% of cases) and considered as bronchial-artery-to-pulmonary-artery collateral flow [16]. Thus, bronchial artery hypertrophy has been identified as a cause of reperfusion of small residual aneurysm after embolization [17]. Bronchial-to-pulmonary artery anastomoses may enter the pulmonary circulation distal to the embolized artery supplying the scarred region of the obliterated PAVM and may lead to future recanalization [17]. The formation of systemic collaterals may place patients at risk for future hemoptysis [17, 27, 28]. A large phrenic artery was a transpleural supply to a previously embolized PAVM in our first two patients. Systemic supply to the PAVM may be a pre-existing condition revealed by the relative ischemia induced by the shunt, as demonstrated in our third patient. Surprisingly, regression of pre-existing systemic supply to the complex PAVM was observed in this patient. To our knowledge, such an observation has never previously been reported. Large systemic arteries may also develop after radiological or surgical treatment of PAVMs to supply the residual scar [24, 27, 28]. Although the incidence of a systemic supply to PAVM is low, its frequency is probably underestimated. Arteriography is not commonly performed in these patients and most patients are evaluated using unenhanced CT examinations [27]. CT angiography, particularly with the use of multidetector technology, allows characterization of the bronchial arterial system and improves detection of even tiny arteries [28]. The clinical impact of this left-to-left shunt of oxygenated blood is considered to be low but the risk of hemoptysis is unknown [18, 21, 26]. Systemic supply to the PAVM is highly significant during pregnancy or when the technique of pulmonary flow redistribution is used [3, 29, 30]. However, the strategy of bronchial artery embolization in patients with recurrent hemoptysis is yet to be determined [29, 31]. A case of hemoptysis persisting despite successful occlusion of a single PAVM has been reported [29]. The patient refused embolization of the bronchial arteries supplying the embolized PAVM and underwent right lower lobectomy. The risks of parenchymal necrosis following systemic embolization should be carefully evaluated, and the type and size of embolization agents carefully discussed [32].

Although embolization is a safe and effective treatment in the management of PAVMs, long-term follow-up of patients is mandatory to document aneurysmal regression of treated lesions and to detect growth of small PAVMs reaching the threshold size for neurologic emboli. Reperfusion and recurrence of treated PAVMs may occur because of recanalization of embolized arteries, recruitment of normal branches or development of a systemic arterial supply. When coils are used, it is important to perform the embolization as distally as possible in the feeding vessel to a PAVM, close to the venous sac, to avoid the occlusion of branches to normal lung and to reduce the rate of reperfusion. The value of spiral CT with dynamic contrast enhancement should be discussed during the follow-up of patients with large complex PAVMs.

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