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Pulmonary Arteriovenous Fistula: Presentation, Diagnosis, and Treatment

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Abstract. Pulmonary arteriovenous fistulas (PAVFs) are rare vascular malformations of the lung. There is a strong association with Rendu-Osler-Weber disease. Although most patients are asymptomatic, PAVFs can cause dyspnea from a right-to-left shunt. They can also bleed and result in hemoptysis and hemothorax. Because of paradoxical emboli, various central nervous system complications have been described including stroke, and brain abscess. Currently, spiral computed tomography offers the most practical method for establishing the presence of PAVFs. Most patients should be treated. Therapeutic options include angiographic embolization with metal coils or balloon occlusion and surgical excision. Angiographic treatment has become the mainstay of therapy for most patients during the last decade. It is less invasive and can be repeated easily. Surgery, which usually consists of a conservative lung resection, is associated with low morbidity and a low recurrence rate. Both therapeutic approaches are discussed. The Mayo Clinic surgical experience of the last 20 years for PAVFs is presented.

Pulmonary arteriovenous fistulas (PAVFs) are vascular malformations that represent direct communications between the pulmonary artery and vein without an intervening capillary bed. They occur with an incidence of 2 to 3 per 100,000 population [1] and have been described under a variety of pseudonyms including benign cavernous hemangioma, pulmonary arteriovenous angiomatosis, hamartomatous angioma of the lung, arteriovenous aneurysm and pulmonary arteriovenous malformation [2, 3]. Females are more often affected than males [4, 5]. Multiple lesions are found in 33% to 50% of patients [6, 7] and can be bilateral in 8% to 20% [3]. Earliest reports were on postmortem findings. Churton in 1897 [8] described multiple bilateral PAVFs in a 12-year-old boy, and Wilkens in 1917 [9] reported a patient who died from massive pulmonary hemorrhage, having presented with the clinical constellation of epistaxis, dyspnea, telangiectasia, clubbing, cyanosis, and bilateral axillary murmurs. Shenstone [10] performed the first surgical intervention, which was a pneumonectomy for a large central lesion. This was followed by several reports on surgical procedures for PAVFs [4, 5, 11–14]. More recently, angiographic techniques for therapeutic embolization of PAVFs using metal coils or detachable silicon balloons have been described [15–19].

More than 500 cases have been reported [20], with more than 80% congenital [21]; of these, roughly half appear to be associated with hereditary hemorrhagic telangiectasia, or Rendu-Osler-Weber (ROW) disease [22]. Those lesions are thought to represent persistent primitive arteriovenous communications from pulmonary buds that fail to mature into capillary beds and thereby function as persistent right-to-left shunts [23, 24].

Secondary PAVFs probably develop from defects in terminal capillary loops that become dilated to form thin-walled vascular sacks, each supplied by enlarged afferent arteries and drained by a single efferent vein. Underlying pathologic processes include trauma [25], infection (actinomycosis [26] and schistosomiasis [27]), long-standing hepatic cirrhosis [28], mitral stenosis [29], metastatic carcinoma [30], and systemic amyloidosis [31]. Pregnancy has been implicated as a cause of deterioration in patients with arteriovenous fistulas, but the mechanism remains unclear [32].

The afferent supply is usually from the pulmonary artery but can derive from the systemic circulation by way of a bronchial artery, intercostal artery, or a direct branch from the aorta. Venous drainage is usually to the pulmonary veins, although on rare occasions there can be a direct communication to the left atrium [6]. Most fistulas are subpleural or located in the outer third of the pulmonary parenchyma. Occasionally in patients with ROW disease, the fistulas evolve into larger cavernous-type lesions with a tendency to infiltrate the bronchial wall, where they can rupture and result in massive hemorrhage.

Clinical Findings

More than half of patients with PAVFs are asymptomatic. Lesions are often discovered on routine chest roentgenograms. Risk factors for developing symptoms include young age, fistula more than 2 cm in diameter, and ROW disease. Fatigability, exertional dyspnea, and palpitations are frequent findings [33]. Central nervous system (CNS) symptoms, including headache, vertigo, numbness, paresis, syncope, and dysphagia, have been reported in up to one-third of patients [18, 20, 34]. The classic triad of exertional dyspnea, cyanosis, and clubbing is found in 30% of adults [7].

Findings on physical examination include cutaneous telangiectasia in one-third of patients. Cyanosis may be central or peripheral (or both), and patients may present with digital clubbing. Occasionally a continuous extracardiac murmur is audible during

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late systole and early diastole, exaggerated by deep inspiration. Murmur intensity can also be augmented by Mueller's maneuver and decreased with the Valsalva maneuver. Dines et al. described a bruit in 70% of patients with ROW disease but in only 35% of those without the disease [4]. Depending on the degree of right-to-left shunt, the hypoxemia is refractory to supplemental oxygen in 80% of cases. Furthermore, assuming the upright position can be associated with an additional fall in arterial oxygen content. This is referred to as "orthodeoxia" and is best explained by gravitational shifts in blood flow to the lung base where most PAVFs are located. Sampling arterial blood gas on room air in both the supine and erect postures therefore becomes a valuable adjunct to clinical diagnosis.

Recognizing the potential for cerebral complications is of paramount importance. Major CNS complications include seizure, brain abscess, transient cerebral ischemia, and stroke [5, 14, 18, 35, 36]. Mechanisms of injury include sludging due to polycythemia, fragmentation of local thrombus, paradoxical embolism, and complications due to concomitant cerebral lesions [14, 37–40].

Bleeding is reported in 10% of patients [41]. Most patients present with epistaxis or hemoptysis, but hemothorax can also occur; and there are several reports describing rupture of pleura-based lesions during pregnancy [41–44]. Air embolism and bacterial endocarditis are uncommon.

Diagnosis

The characteristic finding on the plain chest roentgenogram is a circumscribed, lobulated density. Most lesions are located in the lower lobes [11, 33]. Rarely, large PAVFs can be associated with rib notching, suggesting systemic arterial blood supply from an intercostal vessel. Historically, confirmation of the vascular nature of large lesions was achieved via fluoroscopy, which demonstrated pulsation and modifications in lesion diameter with the Valsalva or Mueller maneuver [45]. Conventional tomograms can also be diagnostic if they demonstrate that a mass (fistula) has afferent and efferent vessels. A recent report on the use of unenhanced three-dimensional helical computed tomography (CT) illustrated additional advantages in the assessment of angioarchitecture, clearly establishing the diagnosis of PAVFs in 95% of cases in a series by Remy et al. [46]. Currently, spiral CT offers the least invasive and least expensive method to establish the presence of PAVFs. It also can be used to follow the potential growth of emerging lesions and to verify the sustained occlusive effect of embolotherapy. The number and size of the fistulas can be determined by obtaining appropriate imaging data, and afferent and efferent vessels can be displayed. Without identifying the feeding artery or draining vein, a fistula cannot be distinguished from a pulmonary nodule. Remy et al. [46] have shown that the additional application of surfaced rendered three-dimensional reconstruction from thin spiral CT data reveals angiogram-like images of PAVFs. Such images are indeed a good example of how much information is contained within spiral CT images, but currently it is a time-consuming process (and therefore expensive) and offers little additional information for the interventional radiologist who is experienced in embolizing these lesions.

Contrast echocardiography can also be useful for diagnosis and monitoring after therapy. The technique involves injection of agitated saline into a peripheral vein. Appearance of a cloud of bubbles in the left atrium confirms right-to-left shunting, as gas bubbles do not survive a normal capillary bed (of course a patent foramen ovale must first be excluded). On occasion, if bubbles can be seen entering the left atrium via a single pulmonary vein, contrast echocardiography confirms the anatomic localization of lesions. Pulsed Doppler imaging further clarifies the diagnosis when increased velocity and spectral disparity are observed with an arterial flow pattern in the pulmonary vein as would be found with a large PAVF [47]. Furthermore, contrast echocardiography allows assessment of the efficacy of embolization therapy, and it is an effective test to exclude the presence of PAVFs in family members of patients with ROW disease [48, 49].

Remy et al. [50] proposed the following diagnostic strategy: The initial investigation should include a chest roentgenogram and a blood gas analysis first performed on room air and then after breathing 100% oxygen in the upright position. For suspicious lesions, echocardiography with intravenous injection of agitated saline should be performed. If all these procedures are normal, it is unlikely the patient requires further investigation. If these studies are positive or inconclusive, a CT scan of the chest should be undertaken. If CT demonstrates lesions large enough or numerous enough for angiographic embolization, pulmonary angiography should be performed.

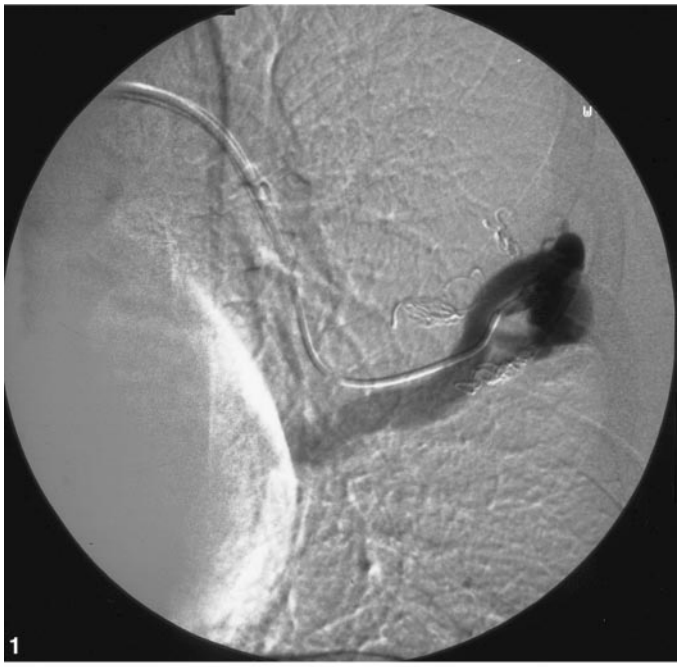
Treatment

The primary objectives of therapy are to eliminate or reduce the right-to-left shunt and to prevent and treat complications. Because untreated lesions are associated with 11% mortality and 26% morbidity [4], most patients with PAVFs should be treated. Relative indications for treatment are patients with small lesions (< 1 cm) who are asymptomatic with minimal or no shunt. Absolute indications for treatment include symptomatic patients, multiple, large, or enlarging fistulas, lesions with a systemic blood supply, the presence of significant hypoxemia on room air, complicated PAVFs, and patients with ROW disease [4, 5, 12, 14, 18–20, 50–56].

Angiographic Embolization

For at least the past decade, the standard of therapy for most PAVFs has been angiographic intervention [18, 50–56]. The fact that it is less invasive than surgery and can be repeated easily are two major advantages. Two methods have been used: occlusion balloons and metallic coils. Each method has its proponents, advantages, and inherent potential complications. It is not obvious that one technique is superior [56], but it is important that the angiographer be experienced [55].

The technical aspects are among the most challenging encountered in interventional radiology, especially when confronted with large, complex lesions that are centrally located or when there are many (20-plus) lesions to treat. Achieving satisfactory catheter placement(s) for delivery of the embolic device(s) is sometimes especially difficult. It is well known that many PAVFs have more than one feeding artery, and each must be embolized. The shortcoming of occlusion balloons is that they are expensive and may (rarely) deflate prematurely. We prefer to use embolic coils for PAVFs. Choosing the correct sizes of coils and their accurate placement (Figs. 1, 2) are critically important. In patients with extensive involvement with PAVFs, often many small lesions must be embolized. It is sometimes necessary to use microcatheters and microcoils. Such cases require



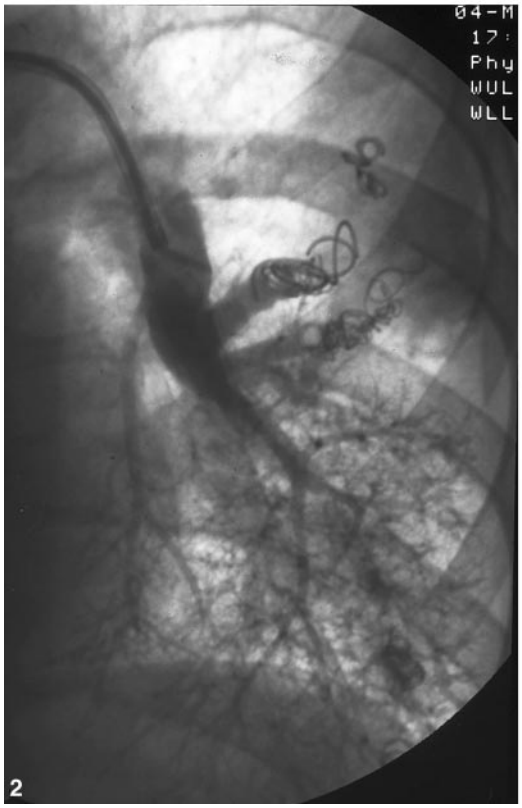


Fig. 1. Selective injection of the remaining feeding artery of a large left lower lobe pulmonary arteriovenous fistula. The adjacent coils have been placed in three other feeding branches to the large fistula.

Fig. 2. Selective injection of left lower pulmonary artery following successful coil embolization of all four feeding arteries of the pulmonary arteriovenous fistula.

multiple long sessions, but the benefit to the patient is significant. Metallic coils are less expensive than balloons, but often many coils must be placed to achieve occlusion.

Recurrence of an initially embolized PAVF can be due to recanalization (10%) or interval growth of an accessory vessel (5%) [55]. To eliminate or reduce the incidence of recanalization, a nest of coils is formed within the feeding artery. Sometimes the fistula itself must be packed with coils. Careful consideration of the technical aspects of embolotherapy for PAVFs is especially important for preventing complications [55]. The threat of embolic material escaping through the fistula is ever present. This includes the embolic material as well as inadvertent injections of air or thrombus. Other complications specific to embolotherapy for PAVFs include pleurisy, pulmonary infarction, and pneumothorax.

Surgical Treatment

Treatment for the solitary PAVF remains a subject of debate. Although surgical excision has the benefit of offering definitive therapy for a solitary PAFV, it is more invasive than embolotherapy. Excision through a thoracotomy is associated with low morbidity and mortality and a less than 2% recurrence rate [4, 5, 12, 14]. At this time for a solitary PAVF the decision to operate or to treat with embolotherapy should be based on the resources and expertise available. Large central PAVFs with a short neck are best treated by surgical excision. A thoracotomy allows control of the hilar vessels, and the fistula can be excised without resection of pulmonary parenchyma. Patients who are actively bleeding from intrapleural rupture of a PAVF should also be operated on without delay. If repeat angiographic embolization has failed, is contraindicated, or is unavailable, conservative surgical excision such as wedge or segmentectomy is the procedure of choice, as most lesions are subpleural. When bilateral lesions are present, staged bilateral thoracotomies offer a viable alternative to embolization [13]. In selected cases, combined staged angiographic and surgical approaches of complex PAVFs preserve maximal lung tissue and result in functional benefit for the patient [57].

Results

We reviewed our surgical experience over a recent 20-year period. Only one patient was operated on during the last 10 years, which reflects the fact that angiography has become the primary treatment for most patients. The clinical records of all patients who underwent surgery for PAVFs between 1976 and 1996 at the Mayo Clinic were analyzed. There were 20 women and 10 men with a mean age of 37 years (range 18–67 years). Twenty-five patients (83%) had associated ROW disease. Altogether 24 (80%) patients presented with symptoms including fatigability 21/24 (88%), dyspnea 20/24 (83%), hemoptysis 6/24 (25%), chest pain 4/24 (17%), and palpitations 2/24 (8%). One patient presented with hemothorax and hemodynamic shock. Three patients became symptomatic while pregnant. Cerebral symptoms were present in 8 of the 30 patients (27%), and included migraine (n = 2), dizziness (n = 2), seizures (n = 1), transient cerebral ischemia (n = 1), and brain abscess (n = 1). The triad of dyspnea, cyanosis, and clubbing was present in 10 patients (33%). Six patients were asymptomatic: Four of these patients had an abnormal chest roentgenogram, and two were found to have a murmur on physical examination. Fistulas were multiple in 21 patients (70%) and single in 9 (30%). Fifteen patients (50%) had bilateral lesions. Fistula size ranged from 4 to 45 mm. Lesions were located in the lower lobes in 21 patients (70%).

Table 1. Procedures in 30 patients with PAVFs.

Procedure	Single lesion	Multiple lesions
Pneumonectomy		1
Lobectomy	3	4
Segmentectomy	4	2
Multiple segmentectomies		1
Lobectomy + segmentectomy		2
Wedge excision	2	7
Multiple wedge excisions		4
Total	9	21

All patients underwent a thoracotomy and pulmonary resection (Table 1). Twenty-one patients (70%) underwent parenchyma-sparing resection. Seven patients had more than one lesion resected. Three patients (10%) had a postoperative complication; these included prolonged mechanical ventilation, hemorrhage requiring transfusion, and prolonged air leak. One hospital death occurred in a patient who developed postpneumonectomy edema. Follow-up was complete in all 29 survivors for a mean period of 30 months. Twenty-seven patients (93%) were improved. Six patients (20%) were reoperated for contralateral lesions, and one patient underwent angiographic embolization for five persistent PAVFs. There were four late deaths; three were secondary to unrelated disease. The third patient had ROW disease and died following a massive upper gastrointestinal hemorrhage.

Comparison with other similar series is difficult. There are few published reports in the recent surgical literature obviously because angiography intervention has become the mainstay of therapy for most PAVFs. More than 90% of outpatients had relief of their preoperative symptoms. These results are comparable to those reported by others [5] and are consistent with previous reports from our own institution [4, 12, 14].

Conclusions

Regardless of the type of therapy, long-term follow-up of treated PAVFs with spiral CT is mandatory to monitor the development of new lesions and recurrence of previously treated ones.

Résumé

La fistule artérioveineuse (FAVP) est une malformation vasculaire rare du poumon, souvent associé à la maladie de Rendu Osler Weber. Alors que la majorité des patients sont asymptomatiques, la FAVP peut être responsable de dyspnée en cas de shunt droit-gauche. Ces fistules peuvent également saigner et sont alors responsables d'hémoptysie et d'hémothorax. En raison du risque d'embolisation paradoxale, on a décrit des complications du système nerveux central comme les accidents vasculaires et les abcès du cerveau. A présent, la tomodensitométrie spiralée est le moyen le plus pratique pour établir le diagnostic de FAVP. La plupart des patients doivent être traités. Les options thérapeutiques comprennent l'embolisation angiographique au moyen de ressorts métalliques ou d'occlusion par ballonnet, suivie d'excision chirurgicale. Le traitement angiographique est devenu le traitement de choix pour la plupart de patients pendant cette dernière décennie. Elle est moins invasive et peut être facilement répétée. La chirurgie, habituellement une résection pulmonaire conservatrice, est associée à un taux de morbidité bas et un taux de récidive peu élevé. On discute ici les deux modalités thérapeutiques. L'expérience chirurgicale de la Mayo Clinic pendant les 20 dernières années est présentée.

Resumen

Las fístulas arteriovenosas pulmonares (PAVF) constituyen una rara malformación congénita del pulmón. Se asocian, frecuentemente, con la enfermedad de Rendu-Osler-Weber. Aunque una gran mayoría de los pacientes son asintomáticos, las PAVF pueden cursar, como consecuencia de un cortocircuito derecho-izquierdo, con disnea. También pueden sangrar, dando origen a hemoptisis y hemotórax. Debido a embolias paradójicas se han descrito complicaciones que afectan al sistema nervioso central tales como: ictus apopletiformes y abscesos cerebrales. Normalmente, la CT espiral constituye el método diagnóstico más eficaz para la detección de las PAVF. La mayoría de los pacientes requieren tratamiento. Las opciones terapéuticas varían entre: la embolización angiográfica con espirales metálicas, la oclusión con balón o, la extirpación quirúrgica. En la última década el tratamiento angiográfico se ha convertido en el de elección para la mayoría de los pacientes; es menos invasivo y puede repetirse con facilidad. El tratamiento quirúrgico, que consiste normalmente en una resección pulmonar conservadora, cursa con escasa morbilidad y la tasa de recidivas es ínfima. Se discuten ambas opciones terapéuticas. Se presenta la experiencia de la Clínica Mayo en los últimos 20 años.

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