Interventional Radiology

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Pulmonary Arteriovenous Malformations: Techniques and Long-term Outcome of Embolotherapy¹

Over a 10-year period, 276 pulmonary arteriovenous malformations (PAVMs) were occluded with balloon embolotherapy in 76 patients, 67 (88%) of whom had hereditary hemorrhagic telangiectasia. Eleven patients (14%) were discovered by means of family screening with measurement of arterial blood gases and chest radiography. Epistaxis, dyspnea, hemoptysis, and hemothorax occurred in 79%, 71%, 13%, and 9% of patients, respectively. Clinical histories of strokes and transient ischemic attacks were present in 18% and 37% of patients, respectively. Computed tomographic scans of 59 patients showed stroke in 36%. Sixty-five percent of PAVMs were located in the lower lobes, which correlated with the finding of more pronounced hypoxemia in the upright position. After embolotherapy, symptomatic hypoxemia was corrected, and serial values have remained constant for 5 years. Complications were minimal, and no patient required surgery. Balloon embolotherapy is effective long-term therapy for PAVMs, and family screening should be pursued because of the possibility of a higher frequency of paradoxical embolization (stroke) than previously recognized.

Index terms: Arteries, therapeutic blockade, 60.1299 • Arteriovenous malformations, 60.1494 • Catheters and catheterization • Interventional procedures, 944.1299 • Telangiectasia

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PULMONARY arteriovenous malformations (PAVMs) were first managed nonsurgically by Porstmann 11 years ago with homemade metal coils (1). Shortly thereafter, the second and third patients treated with embolotherapy by means of stainless steel coils and detachable balloons were reported (2,3). Since these first reports, the publication of numerous series has documented the efficacy of embolotherapy in the treatment of PAVMs (4-12).

During the intervening 11 years, better catheterization techniques have been developed and our understanding of the pulmonary-neurologic relationships leading to embolic stroke in these patients has improved (13). This report discusses the natural history of PAVMs, including the significant and often undetected evidence of paradoxical embolization to the brain, the importance of family screening in the detection of asymptomatic family members, and the description of improved techniques for embolotherapy of these malformations with detachable balloons that are, in some instances, used in combination with stainless steel coils.

PATIENTS AND METHODS

Patient Population

Seventy-six patients with PAVMs were admitted to The Johns Hopkins Hospital over the past 9½ years (May 1978–October 1987). There were 31 male and 45 female patients ranging in age from 5 to 76 years (mean, 36 years). Twenty patients were less than 21 years of age at the time of the embolization. Seventeen of the 76 patients have been previously reported (3,7,9).

Complete histories, physical examinations, and chest radiographs were obtained, as were arterial blood gas (ABG) measurements on room air in the lying, standing, and sitting positions before and after embolotherapy. Computed tomography (CT) of the brain was also performed before and after the intravenous injection of contrast material in the last 59 patients. A family screening program was initiated and consisted of ABG measurements in the lying and sitting positions and chest radiography. The program was offered to family members with a history of epistaxis or telangiectasia on physical examination.

Most patients remained in the hospital for 1 week and underwent complete diagnostic pulmonary angiography on the 1st day, including selective right and left pulmonary artery arteriography in the posteroanterior, lateral, and oblique projections. Serial radiography and, more recently, intraarterial digital subtraction angiography (DSA) were used. Embolotherapy of each lung was planned on days 3 and 5 of the hospitalization, while on alternate days consultations were obtained from the Departments of Neurology, Otolaryngology, and Pulmonary Medicine.

Technique

Standard 6- and 7-F pigtail catheters were used for diagnostic angiography. Since our previous reports, the techniques for embolotherapy have been modified (7,9). Using the diagnostic angiogram as a roadmap, we chose the position that best displayed the anatomy of the PAVM for fluoroscopy during embolotherapy. When the segmental artery was entered, a hand injection with a 30% ionic contrast medium was performed with the 6-inch mode of the image intensifier (intraarterial DSA). Careful attention was directed to the branching pattern of the artery leading to the PAVM. PAVMs with one artery entering the aneurysm and a single draining vein were termed simple, while PAVMs with two or more arteries and draining veins were classified as complex (Figs 1, 2). In general, aneurysms connecting the artery and vein were cavernous without septa in simple PAVMs and were septated in complex PAVMs. Measurement of the pulmonary artery diameter 1-2 cm proximal to the aneurysm guided our selection of embolotherapy technique.

During embolotherapy, a 9-F sheath with a hemostasis valve (Cordis, Miami) was placed in the femoral vein to allow the easy exchange of catheters. Patients

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received 5,000 units (USP) of heparin to minimize the possibility of pericatheter thrombus. For catheterization of segmental pulmonary arteries, reinforced multipurpose 7- and 8-F polyurethane catheters (Cordis) were most commonly used (Fig 3). For catheterization of the middle lobe and lingula, polyurethane internal mammary artery shapes were helpful (Cordis). Bentson flexible 15-mm gentle curve or straight wires were used for catheterizing the segmental artery. By adjustment of the position of the polyurethane reinforced catheter, the segmental artery leading to the PAVM was selectively entered with one of the guide wires. The catheter was readily advanced into the segmental artery over the guide wire, and an exchange for the larger embolotherapy catheter was performed with a 260-cm exchange wire. For embolotherapy of PAVMs with arteries larger than 4 mm, an 80-cm, 8.8-F, nontapered, thinwalled polyethylene catheter was passed coaxially with a 100-cm, 4.9-F, nontapered polyethylene catheter (Becton-Dickinson, Fairfield, NJ) over a 260-cm tight "J" exchange wire (Fig 3). For PAVMs smaller than 4 mm, a 4.9-F nontapered polyethylene catheter was exchanged for the multipurpose catheter and used for embolization.

Balloon embolotherapy as previously described was used in all instances (Figs 1, 2) except when the artery exceeded 9 mm in diameter (9,14). In these latter cases, coils of varying diameters, usually 10–12 mm, were placed distally in the artery as a scaffolding, followed by the placement of a 2-mm balloon inflated to 9 mm in the "nest" of coils (Fig 4). This procedure produced immediate occlusion.

Intraarterial DSA was performed before balloon detachment to ensure that the PAVM was completely occluded and that occlusion of nonessential normal branch arteries was avoided. To avoid the occlusion of normal branch arteries, the balloon was often deflated and repositioned before detachment. In most instances, embolotherapy of two or three malformations was possible in each session. Early in our experience, our goal was to raise the arterial oxygen pressure (tension) (PaO₂) to 60 mm Hg, which produced near normal arterial oxygen saturation (7). As experience was gained and we realized the importance of preventing paradoxical embolization by occluding all PAVMs with arteries larger than 3 mm in diameter, we readmitted some patients for embolization of previously unoccluded PAVMs.

Patients remained in the hospital for a minimum of 24-48 hours after the final embolization. In addition to ABG measurements and chest radiography after embolization, patients were observed for pleurisy, which usually occurred within 48 hours following embolization. Followup was often possible at home and included ABG measurements at 3 months and then yearly thereafter. To prevent brain abscess, a well-recognized complication of PAVM, patients were instructed



Figure 1. (a) Angiogram of young patient with simple PAVM consisting of a single artery and draining vein. (b) Radiograph before detachment shows that balloon attached to the 2-F catheter is inflated distally in the artery, just above the aneurysm. (c) Angiogram before detachment demonstrates distal occlusion with preservation of proximal branch arteries.

to continue to take prophylactic antibiotics before dental work.

Analysis of angiograms consisted of tabulating the number and percentage of simple and complex PAVMs, the number of patients with diffuse PAVMs in every lobe, and the distribution of occluded PAVMs throughout the lung. ABG levels were measured for the entire group before and after embolization. When possible, followup PaO₂ values were obtained to document the permanency of the occlusion.

RESULTS

Overall Findings

Hereditary hemorrhagic telangiectasia (HHT) was present in 67 of 76 (88%) patients with multiple PAVMs. The diagnosis of HHT in 24 (32%) of these patients had not been established, despite the fact that most patients were admitted on a referral basis. Minimally, the presence of telangiectasia on the buccal surfaces, tongue, or conjunctiva, along with a patient or family history of epistaxis, was required for a diagnosis of HHT. The 76 patients underwent a total of 90 hospital admissions, as some of the earlier treated patients returned for repeat embolization of remaining large, unoccluded PAVMs.

Pulmonary symptoms were common, and epistaxis was present in 60 (79%) of the patients (Table 1). Dyspnea occurred in 54 (71%), hemoptysis in ten (13%), and hemothorax in seven (9%). Eleven patients (14%) were discovered after family members were screened with ABG measurements and chest radiography. In retrospect, four of these 11 "asymptomatic" patients had pulmonary symptoms consisting of dyspnea and easy fatigability. Unfortunately, patient compliance was not documented, but it is our impression that a large number of family members did not undergo the necessary screening.

Neurologic symptoms in 76 patients and the results of CT of the brain in 59 patients are summarized in Tables 2 and 3. Histories of strokes and transient ischemic attacks were present in 18% and 37%, respectively, of the 76 patients. Headaches were common, with the migraine type dominating in 43% of the 76 patients. CT scans of the brain were obtained in all 59 patients admitted after June 1982. Hypoattenuating areas in the unenhanced scans were compatible with prior stroke and were present in 36% of patients, with essentially the same frequency as transient ischemic attacks. Three (5%) of the 59 patients had evidence on CT scans of arteriovenous malformations of the brain or hemangiomas of the frontal lobe.

Of the 276 PAVMs treated with balloon embolotherapy, 222 (80%) were simple and 54 (20%) were complex (Figs 1, 2). These percentages for anatomic type are similar to those previously reported from angiographic data (9). Simple and complex PAVMs occurred in the same patient. Five (7%) patients had diffuse malformations with small terminal tufts of malformations located in all lobes of the lung. Four of the five also had large simple or complex PAVMs associated with diffuse disease. Twothirds (65%) of the PAVMs were lo-





f.

h.

cated in the lower lobes, a finding that correlated well with the presence of hypoxemia in the sitting or upright position (orthodeoxia).

Detachable balloons were used alone to occlude 266 PAVMs, and a single large balloon placed in a stainless steel coil matrix provided final occlusion in ten large PAVMs with arteries exceeding 9 mm in diameter (Figs 2-4). In the very large PAVMs, nests of coils were formed first with 10-, 12-, or 15-mm coils interspersed with 8- and 5-mm coils. Four to eight g.

Figure 2. Radiographs and angiograms of a 9-year-old boy with cyanosis and respiratory infection. (a) Chest radiograph demonstrates infiltrate of right upper lobe thought to be a pneumonia. (b, c) Early and late radiographs from pulmonary angiography reveal a large PAVM in the right upper lobe and a smaller PAVM in the left hilus. (d, e) Selected angiograms of the right upper lobe reveal complex PAVM with multiple arteries. (f, g) Radiograph and angiogram show that two large balloons were required to occlude PAVM. In the larger of the two arteries, a 10-mm coil was placed proximally. (h) Final chest radiograph before discharge of patient demonstrates balloons in place in right upper lobe and left superior segment.

coils were placed distally in the artery immediately above the aneurysm. To maintain a compact nest distally in the vessel, it was often necessary to slightly advance the 4.9-F nontapered catheter as the coils were extruded. In most instances, the uninflated balloon could be injected into the proximal portion of the coil matrix, then inflated and detached securely to effect final occlusion of the PAVM (Fig 4). In several additional patients, a balloon placed distally in the feeding artery was considered too small, and several 10-mm coils were placed proximally to prevent any retrograde migration of the balloon (Fig 2).

Our use of the balloon technique

alone spared normal side branches and effected a more distal and localized occlusion than that possible with the coil method. Another advantage of the balloon technique was that it allowed us to deflate and reposition the balloon before its detachment. Three of the 76 patients were referred after coil embolization failed (the artery supplying the PAVM was still patent). In these instances, no more than two coils had been placed in the PAVMs, which may have accounted for the failure of thrombosis. In these patients, detachable balloons were placed distal to the nonoccluding coils.

Technically, we were successful in occluding all PAVMs with arteries

exceeding 3 mm, even though multiple catheterizations were necessary in some. The most common cause of failure of the first occlusion attempt was the inability to gain access to the feeding artery. This improved with the use of the multipurpose catheter. Patients in whom we were technically unsuccessful in the initial occlusion of PAVMs underwent repeat successful occlusions with multipurpose catheters.

Hypoxemia was significantly improved, and orthodeoxia (the tendency for greater hypoxemia in the upright position) disappeared after balloon embolotherapy (Fig 5). The mean PaO₂ values in the lying, sitting, and standing positions were 58, 55, and 47 mm Hg before embolotherapy and 75, 71, and 68 mm Hg after embolotherapy. The finding of orthodeoxia correlated well with the distribution of PAVMs in the lung. With 65% of PAVMs in the lower lobes, increased pulmonary blood flow and secondary right-to-left shunting occurred in the upright position, resulting in lowered arterial oxygen tension in the upright compared with the supine position.

Failure to correct PaO₂ to completely normal values was most commonly due to residual PAVMs that were too small to be considered for occlusion. Patients in whom PAVMs were occluded early in our experience (with the goal of raising the arterial PaO₂ values to 60 mm Hg) returned later as the PaO₂ level dropped (9). This reduction during the follow-up period was attributed to the redistribution of blood flow to large unoccluded PAVMs. Since 1982 symptomatic hypoxemia has been successfully corrected, and serial values have remained constant for 5 years. These patients have remained asymptomatic since therapy.

As part of the follow-up in the earlier patients who returned for occlusion of PAVMs not occluded on the first admission, pulmonary angiography was performed. In the 16 patients who underwent follow-up studies, collateralization to or recanalization of a previously occluded PAVM was not demonstrated.

Complications

Air embolus.—Occasionally, during extremely selective positioning of the end-hole multipurpose or balloon-introducer catheter, the catheter became wedged. The inability to withdraw blood was an indication of catheter wedging, and multiple attempts to flush carefully through the catheter led to the injection of a small amount of trapped air within the saline flush. Less than 5% of patients experienced perioral paresthesia or angina with ST-T wave changes lasting 20 minutes or less. Patients with these symptoms were treated with oxygen by mask and intravenous atropine for bradycardia. These changes were all reversible, and the procedure was continued. This complication is completely avoidable with careful flushing of the catheter. Failure to aspirate blood from the catheter indicates a wedged position, and the catheter should be withdrawn until free flow is readily obtained.

Self-limited pleurisy.—Eight (10%) of patients experienced self-limited pleurisy, which occurred 12-48 hours after embolization and lasted 3-5 days. One of the eight patients had a significant pleural effusion associated with pulmonary infarction that resolved without thoracentesis. Pulmonary infarction most likely occurs because of occlusion of normal branches by the proximal balloon. The selective placement of the balloon distally in the feeding artery reduces this complication. In all patients, pleurisy was not serious and symptoms were treated without heparin.

Paradoxical embolization of the balloon.-Paradoxical embolization occurred in two of the 276 PAVMs occluded (.7%). In both instances, operator error accounted for the complication. Incomplete occlusion accounted for balloon migration in the first instance, and low-resolution fluoroscopy with subsequent detachment of the balloon in the aneurysm rather than in the artery supplying the PAVM accounted for the second balloon migration. The first balloon was lost in the hepatic artery, and the second balloon was lost in the left internal iliac artery. Both patients were asymptomatic, and no long-term sequelae have been recognized. Three patients were referred to us because of paradoxical embolization of coils through a PAVM. In no instance were there symptoms or long-term side effects. These types of inadvertent device embolizations can be avoided with high-resolution fluoroscopy during the placement of balloons and with intraarterial DSA of the balloon occlusion before detachment, to ensure that no contrast medium bypasses the balloon thus indicating that occlusion is complete.

Thrombophlebitis.-Before we began



Figure 3. Polyurethane, end-hole, multipurpose catheter used for selective catheterization (top). A 100-cm, 4.9-F nontapered catheter was passed coaxially through an 8.8-F nontapered catheter over a tight "J" reinforced guide wire (bottom). The 4.9-F catheter was used to deliver the 1-mm balloon, and the 8.8-F catheter was used to deliver the 2-mm balloon.

to switch legs between alternate-day catheterizations, one patient with polycythemia developed deep venous thrombosis after three consecutive catheterizations in the same leg. This complication responded to ten days of intravenous administration of heparin without long-term sequelae. No further patients have developed thrombophlebitis since we began alternating legs for each of the catheterizations.

DISCUSSION

Rodes recognized the association between PAVM and HHT in 1938, although in an earlier report dating back to 1897, Churton described the anatomic findings in a 12-year-old patient with multiple PAVMs (15,16). In the reported series to date, approximately 30%-50% of patients with PAVM have HHT, and the association with HHT is higher in patients with multiple rather than single PAVMs (10,11,17,18). In the present group of patients with multiple malformations, one-third were not aware that they had HHT at the time of admission. With a careful history and physical examination, we concluded that 88% of our patients had HHT.

While our patients were highly selected, we think the association between HHT and multiple PAVMs is much greater than previously reported (17–19). A history of epistaxis was not present in 20% of our patients, and telangiectasia may be too subtle to detect at physical examination. It is well known that untreated patients with multiple PAVMs become dyspneic, are cyanotic, and have clubbed fingers (11,17,18). Furthermore, when left untreated, a percentage of patients sustain significant morbidity



Figure 4. Angiograms demonstrating coil and balloon technique for occlusion of large PAVM. (a, b) Angiogram demonstrates large, simple PAVM of the left lower lobe with 10-mm artery and draining vein. (c, d) Ten minutes after placement of 12- and 10-mm coils, the PAVM remains patent. A 105-mm photospot (e) and angiogram (f) demonstrate a 2-mm balloon inflated to 9 mm, which was detached within the nest of coils to produce final occlusion.

and mortality from brain abscess, stroke, hemoptysis, and hemothorax (11). The first two complications arise because the lung has lost its filter function. The PAVM represents an extracardiac direct connection between the pulmonary artery and the pulmonary vein without an intervening capillary bed. Symptoms occur because bland emboli and bacteria pass directly through these "short circuits" into the systemic circulation. The thin-walled PAVMs may rupture into the bronchus, a condition that leads to hemoptysis. When PAVMs are located subpleurally, they may produce fatal hemothorax.

In the present series, we documented the frequency of prior transient ischemic attacks and strokes. An earlier report from our institution indicated that paradoxical emboli and stroke can occur with a single PAVM in patients with no previous symptoms, hence the importance of family screening (13). A history of strokes and transient ischemic attacks was found in 18% and 37% of our patients, respectively. The last 59 patients have undergone CT of the brain. In 36%, the CT scans were interpreted as showing unequivocal, hypoattenuating areas compatible with prior stroke. To our knowledge, no prior series has documented the frequency of minimally symptomatic strokes or the high association among stroke, HHT, and multiple PAVMs. This association provides an important impetus for family screening of patients with HHT. Since balloon embolotherapy, only one patient has sustained an embolic stroke, 6 months after treatment. In this patient, treated early in our experience, many PAVMs were left unoccluded. Cerebral arterial venous malformations/hemangiomas were present in three patients, two of whom were symptomatic.

Epistaxis occurred in 79% of our patients and clearly is the most common symptom of HHT. Dyspnea occurred in 71% of our patients, a fact that reflects the multiplicity of malformations and the significant anatomic shunt. Simple and complex PAVMs occurred in 80% and 20% of patients, respectively, which was not at variance with our earlier report (9). Hypoxemia on room air is a characteristic feature of patients even with single PAVMs. This is exaggerated in the sitting or standing positions (orthodeoxia) because of the prevalance of PAVMs in the lower lobes (20,21). In our series of 276 occluded PAVMs, 65% were in the lower lobes. After embolotherapy, the difference between PaO₂ values in the sitting and supine positions disappeared, and the mean PaO₂ rose to values compatible with normal arterial oxygen saturation. Failure to correct the PaO₂ value to more than 90 mm Hg reflects the presence of residual small PAVMs. Since 1982, our goals of embolotherapy have been directed to the occlusion of all PAVMs with feeding arteries 3 mm or larger in diameter. No patient treated since then has sustained a stroke in followup, and their PaO₂ values have remained stable, which reflects the very slow growth of residual PAVMs.

The physiologic features of untreated PAVMs and the short-term effects of embolotherapy have been reviewed recently (20,21). From the expanded data in this series, it is clear that hypoxemia is more severe in the erect position than in the supine position in patients with PAVMs. In the seven of 11 family members with newly discovered, asymptomatic, and isolated PAVMs, hypoxemia in the erect position on room air (all PaO₂ values less than 85 mm Hg) was present. This observation requires additional consideration. These seven patients all had

Table 1Pulmonary Symptoms in 76 Patientswith PAVM

Symptoms	No. of Patients
Epistaxis	60 (79)
Dyspnea	54 (71)
Hemoptysis	10 (13)
Hemothorax	7 (9)

Note.—Numbers in parentheses are percentages.

Table 2 Neurologic Symptoms in 76 Patients with PAVM

Symptoms	No. of Patients
Stroke	14 (18)
Transient ischemic attack	28 (37)
Abscess	7 (9)
Migraine	33 (43)
Seizure	6 (8)

Note.—Numbers in parentheses are percentages.

isolated PAVMs, and it is clear that room air ABG measurements are probably adequate enough to allow the detection of isolated PAVMs of sufficient size (supplying artery greater than 3 mm) to warrant further investigation. The next diagnostic procedure should include posteroanterior and lateral chest radiography and, ultimately, diagnostic pulmonary angiography. Radionuclide angiocardiography, contrast echocardiography, and contrast material-enhanced CT have been reported to be useful in the diagnosis of PAVM (22-25). If ABG measurements in the supine and erect positions and chest radiography prove to have the sensitivity that we suspect, then pulmonary angiography would be the next logical step in determining the diagnosis. Selective pulmonary angiography not only provides evidence for the definitive diagnosis, but also has been shown to be useful in the detection of unsuspected PAVMs not recognized on previous chest radiographs (11). Furthermore, selective pulmonary angiography in multiple projections is essential in the planning of embolotherapy.

Until 1977, the treatment of choice for PAVMs was limited to surgical resection or ligation of the PAVM (26,27). A recent review of patients with untreated PAVMs indicates the association of PAVMs with significant morbidity and mortality. Porstmann's report and the subsequent report by Taylor et al indicated that coil embolization was effective thera-

Table 3 Results of Brain CT in 59 Patients with PAVM

Findings	No. of Patients
Stroke	21 (36)
Angioma/AVM*	3 (5)
Abscess	2 (3)
Porencephaly/	
encephalomalacia	3 (5)
Atrophy	5 (9)

Note.-Numbers in parentheses are percent-

* AVM = arteriovenous malformation.



Figure 5. Arterial oxygen pressure (tension) values of first 72 patients (mean and standard error of mean) before and after balloon embolotherapy of PAVM. A significant increase in PaO₂ values occurred in all positions after occlusion, and exaggerated hypoxemia in the standing position (orthodeoxia) was reversed. Failure to correct PaO₂ to normal values reflects the presence of small unoccluded PAVMs. Values were obtained with 55 patients in the lying position, 53 in the sitting position, and 18 in the standing position (P < .001 before and after embolization, all conditions).

py for PAVMs (1,2). Our own initial and subsequent reports favor embolization with detachable balloons (3,7,9). Both methods have a following, but we prefer the balloon approach because of the ability to finetune the placement of the balloon before detachment. Additionally, compared with coils, the more distal placement of the balloon prevents occlusion of normal branches arising from the artery supplying the PAVM. The combination of the coil method with the detachable balloon technique has been used successfully to treat ten patients with large PAVMs (arteries exceeding 9 mm in diameter). This combination includes the placement of oversize coils followed by the placement of smaller ones to form a scaffolding distally in the branch artery supplying the PAVM. To effect permanent occlusion, a large balloon is placed proximally in the coils. An alternative method would include the use of larger latex

balloons, but the need to tie the balloons by hand has limited their widespread applicability.

It is significant that no patient in this series required surgery because of failure to occlude a PAVM with embolotherapy. Follow-up studies have indicated persistent relief of hypoxemia and have shown that minimal growth of small remaining PAVMs has occurred. CT of the brain and review of neurologic symptoms reveals a higher association of embolic stroke in patients with multiple PAVMs and HHT than has been reported previously. Furthermore, a previous report has indicated that paradoxical embolization occurs in patients with isolated PAVM (13). The source of emboli in these patients remains to be determined, but we suspect that they arise in the pelvis or lower extremity. Minimal morbidity and no mortality have occurred in patients treated with balloon embolotherapy. These observations lead us to conclude that all PAVMs with arteries exceeding 3 mm in diameter should be occluded by means of embolotherapy. Additionally, the screening of family members with HHT should be pursued vigorously, since room air ABG measurements may reveal a higher frequency of PAVM than previously reported.

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