

Diagnosis of pulmonary arteriovenous malformations by colour Doppler ultrasound and amplitude ultrasound angiography

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Abstract

Background—The clinical value of colour Doppler ultrasound and amplitude ultrasound angiography in the diagnosis and follow up of pulmonary arteriovenous malformations (PAVM) was investigated. **Methods**—Six consecutive patients suspected by clinical appearance and abnormal chest radiographic findings of having PAVM were included in the study. Ultrasonography was performed first by real time grey scale imaging then by colour Doppler imaging and amplitude ultrasound angiography in a random order. All were later proved by angiography to have PAVM.

Results—The ultrasound study was successfully performed in all six patients. A total of eight lesions was detected. The real time grey scale image of PAVM revealed well defined hypoechoic subpleural nodules with strong posterior acoustic enhancement. Colour Doppler ultrasound of PAVM showed turbulent flow, manifest as an area of intense colour with high and mixed velocities (reticulated or mosaic-like pattern). Anatomical continuity was demonstrated in some PAVM. Amplitude ultrasound angiography can delineate a tangled vascular structure with a clear vessel wall and anatomical continuity as well as conventional angiography. Spectral wave analysis showed a relatively low impedance flow presenting with high peak systolic velocity (mean 44.4 cm/s) and relatively high diastolic velocity (mean 19.3 cm/s). The mean pulsatility index (PI) and resistive index (RI) were 1.80 and 0.49, respectively. In two patients who received embolotherapy the colour Doppler ultrasound scan obtained after the procedure showed that the previous focal areas of colour flow signals disappeared or diminished in size. This was compatible with the decrease in, or absence of, blood flow demonstrated by angiography after embolotherapy.

Conclusions—Combined colour Doppler ultrasound and amplitude ultrasound angiography are useful non-invasive techniques for diagnosing PAVM and provide an alternative approach to angiography in evaluating the efficacy of embolotherapy. (Thorax 1998;53:372-376)

Pulmonary arteriovenous malformation (PAVM) is unusual but not rare. It originates as an anomaly of capillary development which results in a direct connection between branches of the pulmonary artery and pulmonary vein. Terminology is confused by the use of a number of descriptive terms: haemangioma, fistula, aneurysm, and hamartoma. Since most of the lesions represent congenital malformations rather than tumours with malignant potential, it seems reasonable to use the term PAVM to include both arteriovenous connections visible on angiographic examinations and the rare microscopic arteriovenous communications which are too small to be visualised radiographically.^{1,2} There has been a clear association with hereditary haemorrhagic telangiectasia (Osler-Weber-Rendu disease, OWRD).³ The clinical triad of cyanosis, clubbing, and polycythaemia may prompt an investigation for a right-to-left shunt resulting from such a lesion, but the triad may even be absent in patients with large PAVM. Most often the condition is recognised by chest radiography. Pulmonary angiography is still the diagnostic gold standard for PAVM but, although associated with minimal risk, it is still an invasive procedure. Non-invasive procedures for evaluating patients with suspected PAVM include measurement of arterial blood gas tensions on room air, detection of orthodeoxia, dynamic CT scanning, contrast echocardiography, radionuclide angiography, perfusion lung scans, fluoroscopy, and magnetic resonance imaging.^{1,4,5}

Ultrasonography is a useful non-invasive procedure for the diagnosis of chest diseases.⁶⁻⁸ The recent advent of colour Doppler ultrasound which can simultaneously display blood flow information and Doppler spectral wave analysis has provided a useful method for the evaluation of various thoracic lesions. Colour Doppler ultrasound can be used to diagnose pulmonary sequestration,⁹ to assess the haemodynamics of pulmonary consolidation,¹⁰ and to evaluate the angiogenesis of a pulmonary tumour.¹¹ However, there are some limitations to colour Doppler ultrasound. Colour Doppler ultrasound measures the mean Doppler frequency shift for colour generation which is angle dependent and may have aliasing. Amplitude ultrasound angiography or power Doppler ultrasound is a revolutionary advance in ultrasound technology.¹² This new approach displays the strength (power) of Doppler signals in colour, rather than the velocity and

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Table 1 Clinical summary of patients with pulmonary arteriovenous malformation

| Patient no. | Age/sex | Lesion location (no. of lesions) | Osler-Weber-Rendu disease | Treatment |
|-------------|---------|----------------------------------|---------------------------|-------------------------|
| 1 | 31 / M | LLL (1) | + | Surgery |
| 2 | 18 / M | LUL (2) | - | Embolotherapy + surgery |
| 3 | 39 / F | RML + RLL (2) | - | Embolotherapy |
| 4* | 38 / F | RML (1) | - | Observation |
| 5 | 25 / F | RUL (1) | + | Surgery |
| 6 | 55 / M | RLL (1) | + | Observation |

LLL = left lower lobe; LUL = left upper lobe; RML = right middle lobe; RUL = right upper lobe; RLL = right lower lobe.
*Patient from reference 15.

direction of the Doppler ultrasound signals.¹²⁻¹⁴ The more blood cells there are in motion, the stronger the colour signal expresses in white. Thus, in contrast to a colour Doppler image, the colour display does not depend on the insonation angle and is free from aliasing.^{13 14} It is three times more sensitive than colour Doppler ultrasound¹³ and can demonstrate the vascular structure just like angiography without the need for contrast medium. A case of PAVM diagnosed by colour Doppler ultrasound has been reported previously.¹⁵ This study assesses the value of colour Doppler ultrasound and amplitude ultrasound angiography in the diagnosis and follow up of PAVM.

Methods

Prospective analysis of the value of the colour Doppler ultrasound and amplitude ultrasound angiography in the diagnosis and follow up of

PAVM was performed in six consecutive patients who were evaluated and treated between January 1994 and September 1996 at the National Taiwan University Hospital. The study included three men and three women of mean age 34.3 years (range 18-55) in whom PAVM was suspected by clinical symptoms and chest radiographic findings. All were later proved by angiography to have PAVM; two received embolotherapy after diagnosis was confirmed. Three patients were treated surgically, including one who had previously had unsuccessful embolotherapy.

Before the angiographic study all patients were examined with a high resolution ultrasound unit (Aloka SSD 2000, Aloka, Tokyo, Japan; ATL HDI-3000, Advanced Technology Laboratories Inc, Washington, USA) with 2-5 MHz linear and convex transducers, with a pulse repetitive frequency up to 15 kHz. The possible location of PAVM was first judged from the chest radiograph, after which the patients were examined by transducer scanning through the intercostal space over the whole chest with special attention to the area where the lesion was located. It was examined first by real time grey scale ultrasound and then by colour Doppler ultrasound and amplitude ultrasound angiography in random order. Patients were asked to hold their breath temporarily to minimise interference from

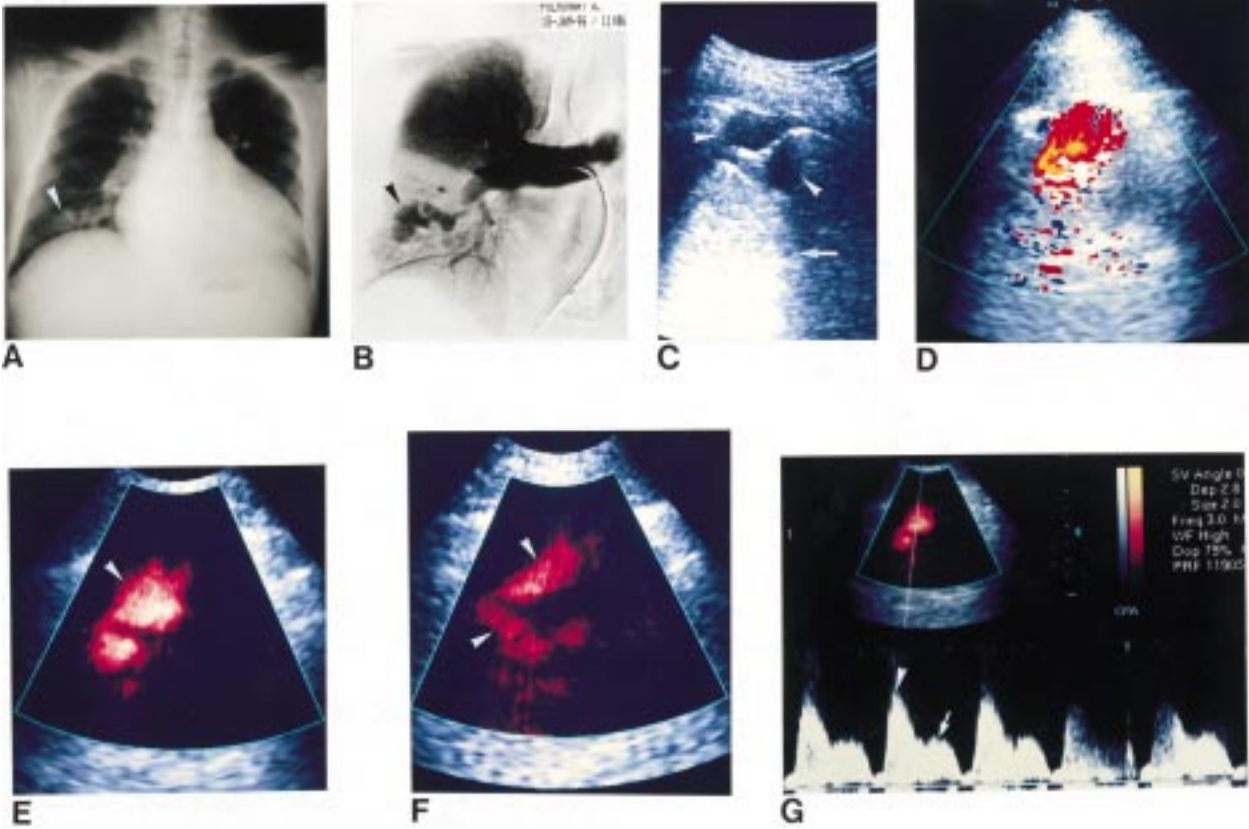


Figure 1 A 55 year old man with OWRD (case 6). (A) Chest radiograph showing a peripheral circumscribed non-calcified nodule over the right lower lobe (arrowhead). (B) Pulmonary artery angiographic scan showing tortuous and coiled vascular channels of PAVM (arrowhead). (C) Real time grey scale ultrasound scan showing hypoechoic nodules (arrowheads) with strong posterior acoustic enhancement (arrow). (D) Colour Doppler ultrasound scan showing a vascular structure with turbulent flow manifest as mosaic-like flow pattern (arrowhead); perivascular colour artifacts from pulsation of PAVM are evident. (E) and (F) Amplitude ultrasound angiographic scan from two different scanning planes indicating a tangled vascular structure with clear vessel outline and anatomical continuity (arrowheads). (G) Spectral wave analysis showing a low impedance flow with high systolic velocity (arrowhead) and relative high diastolic velocity (arrow); the pulsatility index and resistive index are 1.74 and 0.78, respectively.

Table 2 Spectral wave analysis of blood flow signals of pulmonary arteriovenous malformation of eight lesions in six patients

| Patient no. | Pulsatility index | Resistive index | Peak systolic velocity (cm/s) | End diastolic velocity (cm/s) |
|-------------|-------------------|-----------------|-------------------------------|-------------------------------|
| 1 | 2.02 | 0.45 | 31.1 | 17.1 |
| 2 | 1.82 | 0.51 | 38.7 | 18.6 |
| | 1.74 | 0.47 | 37.6 | 19.7 |
| 3 | 1.83 | 0.39 | 24.4 | 14.9 |
| | 1.70 | 0.38 | 23.2 | 14.4 |
| 4* | 1.94 | 0.45 | 33.0 | 18.0 |
| 5 | 1.60 | 0.49 | 48.9 | 24.8 |
| 6 | 1.74 | 0.78 | 118.0 | 26.5 |
| Mean | 1.80 | 0.49 | 44.4 | 19.3 |

*Data from reference 15.

movement of the chest wall while the lesion was exposed and the flow signal was being acquired.

The lesion was first localised and recorded with real time grey scale ultrasound. Colour Doppler mapping of the entire lesion was done to detect any vascular structure and blood flow. The sensitivity to low velocity flow is maximised by choosing the low velocity scale (2.5 cm/s for a Doppler angle of 0–180°). The wall filter was set at a level to minimise rejection of small frequency shifts (low velocity flow) and also to avoid interference from respiratory or cardiac movement. Colour Doppler gain was adjusted until background noise first became apparent on the colour Doppler scan. Colour assignment for these units is such that flow towards the transducer is red and flow away from the transducer is blue. For amplitude ultrasound angiography the gain was increased until the noise just began to exceed the uniform single colour background.¹² When blood flow was detected on colour Doppler ultrasound or amplitude ultrasound angiography, spectral wave analysis was performed by using pulse wave Doppler. In colour Doppler ultrasound and amplitude ultrasound angiography the gate focused on the centre of the flow signals and the transducer was adjusted so that the Doppler angle θ between the flow signal and the ultrasound beam was 60° or less. The spectral waveform (time-velocity Doppler spectrums of flow signal) was analysed for the following Doppler indices: (1) pulsatility index (peak systolic velocity—end diastolic velocity)/mean velocity, (2) resistive (Pourcleot) index (peak systolic velocity—end diastolic velocity)/peak systolic velocity, (3) peak systolic velocity (cm/s), and (4) end diastolic velocity (cm/s).^{16, 17} Peak systolic velocity and end

diastolic velocity were corrected by the Doppler angle θ between the flow signals and Doppler gate, if the angle was not 0° or 180°, by using the microprocessing program in the sonographic unit.

Results

The clinical details of the six patients are shown in table 1. Three were associated with OWRD and two were found by screening members of the family with OWRD. The PAVMs were single in four patients and multiple in two; a total of 22 PAVMs were detected by angiography and eight by sonography.

In most patients the chest radiograph showed peripheral circumscribed non-calcified lesions (fig 1A). The pulmonary angiogram demonstrated distinct, tortuous and coiled vascular channels and all the feeding arteries were from the pulmonary artery (fig 1B). The ultrasound study was successfully performed in all six patients. The real time grey scale image of PAVM revealed well defined hypoechoic subpleural nodules with strong posterior acoustic enhancement (fig 1C). The visceral pleura may be interrupted or can be visible. Pulsations were visible in only one case. Colour Doppler ultrasound of PAVM demonstrated turbulent flow manifest as an area of intense colour with high and mixed velocities (reticulated or mosaic-like pattern) which had a unique appearance compared with other solitary lung lesions (fig 1D). Anatomical continuity was demonstrated in some of the PAVMs. Perivascular colour artifacts due to pulsation of PAVM were always visible. Amplitude ultrasound angiography usually delineate tangled vascular structures with clear vessel walls and anatomical continuity which resembled the findings on angiography (fig 1E, 1F). Different strengths of Doppler signals can be displayed within the tortuous PAVM. The pulsed wave Doppler study was greatly facilitated by better visualisation of vessels provided by amplitude ultrasound angiography (fig 1G).

Spectral wave analysis showed a relatively low impedance flow presenting with high peak systolic velocity (mean 44.4 cm/s) and relatively high diastolic velocity (mean 19.3 cm/s). The mean pulsatility index and resistivity index were 1.80 and 0.49, respectively (table 2).

In two patients who received embolotherapy, colour Doppler ultrasound performed after the procedure showed that the previous focal areas

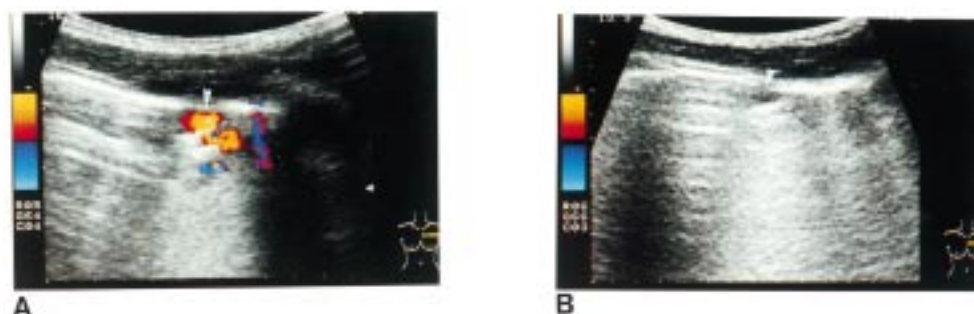


Figure 2 Colour Doppler ultrasound scan of an 18 year old man with PAVM (case 2). (A) Before embolotherapy the scan shows turbulent colour flow signals within a hypoechoic nodule (arrowhead) and after embolotherapy (B) there is no blood flow within the lesion (arrowhead).

of flow manifest as mosaic patterns disappeared or diminished in size (fig 2). This was compatible with decreased or absent blood flow demonstrated by angiography in the final step of embolotherapy.

Discussion

Pulmonary arteriovenous malformations, originally described by Churton in 1897,¹⁸ are not rare.⁴ The association between PAVM and OWRD is well known, and PAVM occurs most commonly in patients with OWRD. It is reported that 44–88% of patients with PAVM also suffer from OWRD.^{2,5} Three of the patients in this study (50%) were diagnosed as OWRD. The clinical impression of PAVM requires confirmation by ancillary testing.¹ Among these diagnostic procedures pulmonary angiography remains the diagnostic gold standard. It has good sensitivity and specificity and, particularly in surgical candidates or those receiving embolotherapy, it is essential for obtaining information about the number, extent and location of the lesions. It can also delineate the feeding artery and drainage vein. Other diagnostic procedures provide indirect evidence either for confirming or excluding the diagnosis.

Ultrasonography is a non-invasive procedure which has the advantage of portability and real time imaging. Recent availability of colour Doppler and amplitude ultrasound angiography has enhanced the diagnostic capability of cardiovascular ultrasound. Antenatal diagnosis of PAVM using real time ultrasound and colour Doppler flow imaging was first reported by Kalugdan *et al* in 1989.¹⁹ In 1990 Sommer *et al* first described the grey scale sonographic appearance of a PAVM in the base of the pleura in a seven year old girl.²⁰ This study is the first to introduce the new technique of amplitude ultrasound angiography for the diagnosis and follow up of PAVM. Since PAVMs are usually located near to the pleura, we were able to find a good acoustic window to image all eight PAVMs in the six patients included in the study. In a study reported by Bosher *et al* 89 of 110 PAVMs (81%) were either immediately subpleural or only partially embedded in the parenchyma.⁴ Thus, the diagnosis of PAVM by ultrasound was found to be possible, as postulated by Sommer *et al*.²⁰

Conventional real time gray scale ultrasound of PAVM gives little information about the vascular nature.²¹ It reveals well defined hypoechoic subpleural nodules rather than a tubular structure and pulsation is rarely visible. By modulating Doppler shift into colour signals, colour Doppler ultrasound can detect and verify PAVM as a vascular structure. The colour Doppler image of PAVM presented as a reticulated or mosaic-like vascular structure with intense colour of high and mixed velocity. This picture results from a high turbulent flow and the low resistance of the lesion. The pulsatility index and resistivity index of the feeding vessel in this study were 1.80 and 0.49, respectively. This resistance is relatively low compared with previously reported blood flow signals of benign pulmonary lesions which were

3.32 (0.68) and 0.90 (0.06), respectively.¹¹ The characteristics of the flow pattern in PAVM were compatible with AVM in other organs.^{22,23} Because of the high turbulent flow within the PAVM, the colour Doppler image is subjected to aliasing and perivascular colour artifact from pulsation of the AVM.^{12–14} In colour Doppler ultrasound the direction of blood flow can be outlined as red (towards the transducer) or green (away from the transducer), allowing direction of flow within the PAVM to be determined. Since only a short segment of the PAVM can be detected it is difficult to identify the feeding artery or drainage vein. In amplitude ultrasound angiography the vessel walls are very well defined and tortuous vessels are outlined in their entire course, as shown in fig 1D and 1E.¹³ The procedure of pulse Doppler analysis was facilitated by the excellent visualisation of vessels also shown there.¹³ Since amplitude ultrasound angiography is more sensitive than colour Doppler ultrasound, it can magnify the motion artifacts as a flash artifact.¹³ This can be decreased by asking patients to suspend breath intermittently while scanning the lesion. Moreover, signals from moving tissue have a low amplitude on amplitude ultrasound angiography and can be discriminated by different shade arrangement or a higher gain setting should be used without a corresponding increase in motion artifact.²⁴ Because the direction of flow cannot be ascertained in amplitude ultrasound angiography, it should be used as an adjuvant to colour and pulsed Doppler ultrasound examination.¹³

Some of the radiographic findings of PAVM have been misinterpreted as coin lesions or bilateral metastatic disease.² The patients may not have the symptoms or the signs to raise suspicion of a diagnosis of PAVM. Colour Doppler ultrasound can prevent the serious bleeding resulting from mistaken needle aspiration biopsy of PAVM.^{15,25} In those patients with small lesions which are clinically silent, or where clinical circumstances dictate a non-surgical candidate, colour Doppler and amplitude ultrasound angiography can confirm the diagnosis. It could also be used as a simple non-invasive screening method to verify the presence of PAVM in all symptomatic and asymptomatic members of families with OWRD and patients with evidence of extracardiac right-to-left shunt by contrast echocardiography.²⁰ Embolotherapy of PAVM with coil or balloon has been widely applied recently.^{1,5} This report also documents the usefulness of non-invasive ultrasound in assessing the efficacy of embolotherapy in the treatment of PAVM. It is also suggested that the procedure be performed before diagnostic needle aspiration of any pleural based lung lesions to prevent disastrous bleeding complications from puncturing any vascular structures.

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