

REVIEW ARTICLE

Pulmonary arterial hypertension: an imaging review comparing MR pulmonary angiography and perfusion with multidetector CT angiography

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ABSTRACT. Pulmonary hypertension (PH) is a progressive disease that leads to substantial morbidity and eventual death. Pulmonary multidetector CT angiography (MDCTA), pulmonary MR angiography (MRA) and MR-derived pulmonary perfusion (MRPP) imaging are non-invasive imaging techniques for the differential diagnosis of PH. MDCTA is considered the gold standard for the diagnosis of pulmonary embolism, one of the most common causes of PH. MRA and MRPP are promising techniques that do not require the use of ionising radiation or iodinated contrast material, and can be useful for patients for whom such material cannot be used. This review compares the imaging aspects of pulmonary MRA and 64-row MDCTA in patients with chronic thromboembolic or idiopathic PH.

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Pulmonary hypertension (PH) is an insidious and progressive disease that leads to substantial morbidity and eventual death. PH results from a number of diseases with different physiopathologies, treatments and prognoses [1]. One of the most frequent causes of PH is chronic thromboembolic pulmonary hypertension (CTEPH).

The current classification of PH (Table 1), developed at the 2008 4th World Symposium on Pulmonary Hypertension in Dana Point, CA [2], resulted from a review of the previous classification developed at the 2003 3rd World Symposium in Venice, Italy. During the 4th World Symposium on PH, an international group of experts agreed to maintain the general philosophy and organisation of the Evian–Venice classifications. However, in response to a questionnaire regarding the previous classification, a majority (63%) of experts felt that modification of the Venice classification was required to accurately reflect information published in the past 5 years and to provide clarification in some areas [2].

PH is a clinical and haemodynamic syndrome that results in increased vascular resistance in the pulmonary circulation, usually by a combination of mechanisms involving vasoconstriction and remodelling of the small vessels [3]. Haemodynamically, it is defined as a systolic pulmonary artery pressure of >35 mmHg, or a mean pulmonary artery pressure of >25 mmHg at rest or >30 mmHg with exertion [4, 5]. An increase in pulmonary

vascular resistance and subsequent compensatory right ventricular (RV) hypertrophy lead to elevated pulmonary pressure, which often results in increased RV afterload and failure. The disorder is progressive, leading to right heart failure and death within a median of 2.8 years after diagnosis [6, 7].

The development of RV failure in patients with pulmonary arterial hypertension (PAH) is an ominous sign with major adverse prognostic implications. Patients with severe PAH or right heart failure die usually within 1 year without treatment. In the National Institutes of Health registry, approximately 50% of deaths in patients with PAH are attributed to RV failure [6]. Numerous factors may indicate a poor prognosis in patients with PAH and secondary RV failure, including age >45 years at presentation, New York Heart Association (NYHA) Class III or IV functional classification, failure to improve to a lower NYHA class during treatment, pericardial effusion, large right atrial size, elevated right atrial pressure, septal shift during diastole, decreased pulmonary arterial capacitance (stroke volume/pulmonary arterial pulse pressure), increased N-terminal brain natriuretic peptide level and hypocapnia [8, 9].

Because patients with PH often present with non-specific symptoms, such as shortness of breath on minimal physical exertion, fatigue, chest pain and fainting, diagnosis often occurs late in the course of the disease, when the prognosis is poor and treatment options are limited [10]. A complete diagnostic evaluation includes a medical history, physical examination, pulmonary function tests, electrocardiogram, echocardiogram, cardiac catheterisation and advanced imaging.

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Table 1. Classification of pulmonary hypertension according to the 4th World Symposium, Dana Point, CA, 2008 [2]

1. Pulmonary arterial hypertension (PAH)
1.1. Idiopathic PAH
1.2. Heritable PAH
1.2.1. Bone morphogenetic protein receptor type 2
1.2.2. Activin receptor-like kinase type 1 (ALK1)
ALK1, endoglin (with or without hereditary haemorrhagic telangiectasia)
1.2.3. Unknown
1.3. Drug- and toxin-induced
1.4. Associated with:
1.4.1. Connective tissue diseases
1.4.2. HIV infection
1.4.3. Portal hypertension
1.4.4. Congenital heart diseases
1.4.5. Schistosomiasis
1.4.6. Chronic haemolytic anaemia
1.5. Persistent neonatal pulmonary hypertension
1'. Pulmonary veno-occlusive disease and/or pulmonary capillary haemangiomas
2. Pulmonary hypertension due to left heart disease
2.1. Systolic dysfunction
2.2. Diastolic dysfunction
2.3. Valvular disease
3. Pulmonary hypertension due to lung diseases and/or hypoxia
3.1. Chronic obstructive pulmonary disease
3.2. Interstitial lung disease
3.3. Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4. Sleep-disordered breathing
3.5. Alveolar hypoventilation disorders
3.6. Chronic exposure to high altitude
3.7. Developmental abnormalities
4. Chronic thromboembolic pulmonary hypertension
5. Pulmonary hypertension with unclear multifactorial mechanisms
5.1. Haematological disorders: myeloproliferative disorders, splenectomy
5.2. Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis
5.3. Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4. Other: tumoral obstruction, fibrosing mediastinitis, chronic renal failure on dialysis

Invasive haemodynamic evaluation is mandatory, not only to confirm the diagnosis but also to address the prognosis and the patient's eligibility for the use of calcium channel blockers through an acute vasodilator challenge. Non-invasive surrogate response markers to the acute vasodilator test have been sought. In other studies, mean pulmonary artery distensibility (mPAD) has been evaluated using MRI to assess pulmonary haemodynamics and diagnose pulmonary vascular disease [11, 12]. The mPAD may reflect the degree of vascular remodelling, making it a very interesting marker for the evaluation of patients with idiopathic PAH (IPAH) [13]. Jardim et al [14] found that the cardiac index, calculated after the determination of cardiac output using MRI and pulmonary artery catheterisation, showed excellent correlation, as did right atrial pressure and the RV ejection fraction. They also found that PAD was significantly higher in acute vasodilator test responders. A receiver operating characteristic curve analysis has shown that 10% distensibility can be used to differentiate responders from non-responders with 100% sensitivity and 56% specificity. This study suggested that MRI and PAD may be useful non-invasive tools for the evaluation of patients with PH. In some cases, definitive diagnosis requires a thoracoscopic lung biopsy [3]. Because CTEPH differs considerably from other forms of PH and may be treated surgically, an accurate diagnosis is essential [15].

The depiction of occluding thrombotic material and concomitant perfusion defects is a prerequisite for the correct and reliable diagnosis of CTEPH. Until recently, pulmonary perfusion could be assessed only by using radionuclide perfusion scintigraphy and conventional pulmonary angiography. The former technique has substantial limitations with respect to spatial and temporal resolution, and the latter requires invasive catheterisation of the right side of the heart and produces only two-dimensional projection images [16].

Pulmonary multidetector CT angiography (MDCTA), pulmonary MR angiography (MRA), and MR-derived pulmonary perfusion (MRPP) are non-invasive imaging techniques used to assess PH-related pulmonary vessel changes in the differential diagnosis [16]. MDCTA is considered the gold standard for the diagnosis of CTEPH because it depicts the occluding thrombotic material and concomitant lung changes [16]. However, the combined use of MRA and MRPP allows the evaluation of PH-related pulmonary vessel changes and concomitant perfusion defects without ionising radiation or iodinated contrast material, and can be useful for patients in whom such material cannot be used. Few studies to date have sought to determine the accuracy of MRA in distinguishing the various causes of PH [16–18].

MRI also contributes to the cardiac evaluation of patients with PH. Cardiac MRI is the gold standard

technique for the assessment of ventricular function and the quantification of volumes and mass without geometric assumptions [19]. Recently, myocardial delayed enhancement after the intravenous administration of a gadolinium-based contrast agent has been shown at the insertion points of the RV free wall in the interventricular septum in patients with PAH and impaired ventricular function [20]. McCann et al [21] also suggested that the extent of hyperenhancement was not correlated with any clinical or haemodynamic variable, but was inversely correlated with RV dysfunction measured on cardiac MRI.

This review aims to compare the imaging aspects of pulmonary MRA and 64-row MDCTA in patients with CTEPH and IPAH, and to highlight the main differences between these techniques. Patients with other forms of PH are not considered here because CT is superior to MRI for the evaluation of lung parenchyma.

Normal anatomy of the pulmonary arteries

Normal pulmonary circulation is a low-pressure system with a high capacitance and approximately one-tenth of the flow resistance of the systemic circulation. The main pulmonary artery originates at the pulmonary valve and extends cranially and to the left before bifurcating into the shorter left and longer right arteries (Figure 1). The right pulmonary artery divides into three lobar branches at the right hilum. After arching over the left main bronchus, the left pulmonary artery also divides into two lobar branches in the left hilum [22]. The pulmonary arterial system is invariably related to the airways and divides with them. The segmental and subsegmental arteries have similar diameters to those of the adjacent airways.

Bronchial circulation is a higher pressure system that also supplies the lung parenchyma. It arises from the thoracic aorta or intercostal arteries. The bronchial arteries are approximately 1.0–1.5 mm in diameter at the hilum. The pulmonary and bronchial vascular networks form rich anastomoses. Bronchial circulation is involved in gas exchange only in the presence of decreased pulmonary flow and ischaemia [22].



Figure 1. Normal 32-year-old female. MR angiography-derived maximum intensity projection reconstruction shows normal pulmonary artery anatomy: pulmonary artery main stem (black arrow), right pulmonary artery (white arrow) and left pulmonary artery (asterisk).

Comparison of idiopathic pulmonary hypertension and chronic thromboembolic pulmonary hypertension

Idiopathic and heritable pulmonary arterial hypertension

IPAH is sporadic and its aetiology and risk factors are unknown. Patients with IPAH usually lack family histories of PAH. The estimated incidence of IPAH is one or two cases per 1 million people. The condition is predominant in females, with a female-to-male ratio of 1.7–3.1:1. PAH is most prevalent between the ages of 20 and 40 years and shows no ethnic predilection. Most patients develop progressive dyspnoea and cor pulmonale, and die within a few years [1, 3, 23]. 6% of patients who participated in the North American National Registry had a family history of PAH [24]. Germline mutations in the bone morphogenetic protein receptor (*BMPR2*) gene are detected in 70% of familial PAH cases [25, 26]. Because *BMPR2* mutations have also been detected in 11–40% of apparently idiopathic cases with no family history [27, 28], the distinction between idiopathic and familial *BMPR2* mutations is artificial. In addition, no *BMPR2* mutation has been identified in $\leq 30\%$ of families with histories of PAH [29, 30]. The category of “heritable PAH” does not mandate genetic testing in patients with IPAH or familial PAH. When called for, genetic testing should be performed as part of a comprehensive programme that includes genetic counselling and discussion of the risks, benefits and limitations of such testing [31].

The radiological findings of IPAH consist of dilated central pulmonary arteries, tortuous arteries, rapid tapering of the vessels (pruned tree sign—enlargement of the arterial roots with a paucity of peripheral arteries) and peripheral oligoemia (Figure 2) [16]. Additional findings include right heart enlargement and paradoxical septal wall motion, which are non-specific and can be found in all PH types [16]. Ventilation–perfusion scans can be normal or may show patchy non-segmental perfusion defects or diffuse reduction in perfusion, usually interpreted as having a low probability of representing thromboembolic disease [1, 4, 18, 23].

Although major therapeutic advances have been achieved for patients with PAH in the last decade, no currently approved therapy can cure this progressive disease. The search for such treatments continues, with promising new concepts arising from an improved understanding of the pathobiology of pulmonary vascular diseases. Patients and physicians should be encouraged to foster such research by participating in randomised clinical trials conducted at specialised PH centres [32].

Chronic thromboembolic pulmonary hypertension

CTEPH develops in approximately 1–5% of patients with acute pulmonary thromboembolism. Patients with malignant neoplasm, chronic cardiac or pulmonary disease, and/or clotting disorders are at increased risk of developing this condition. Patients may be asymptomatic for many years before developing symptoms

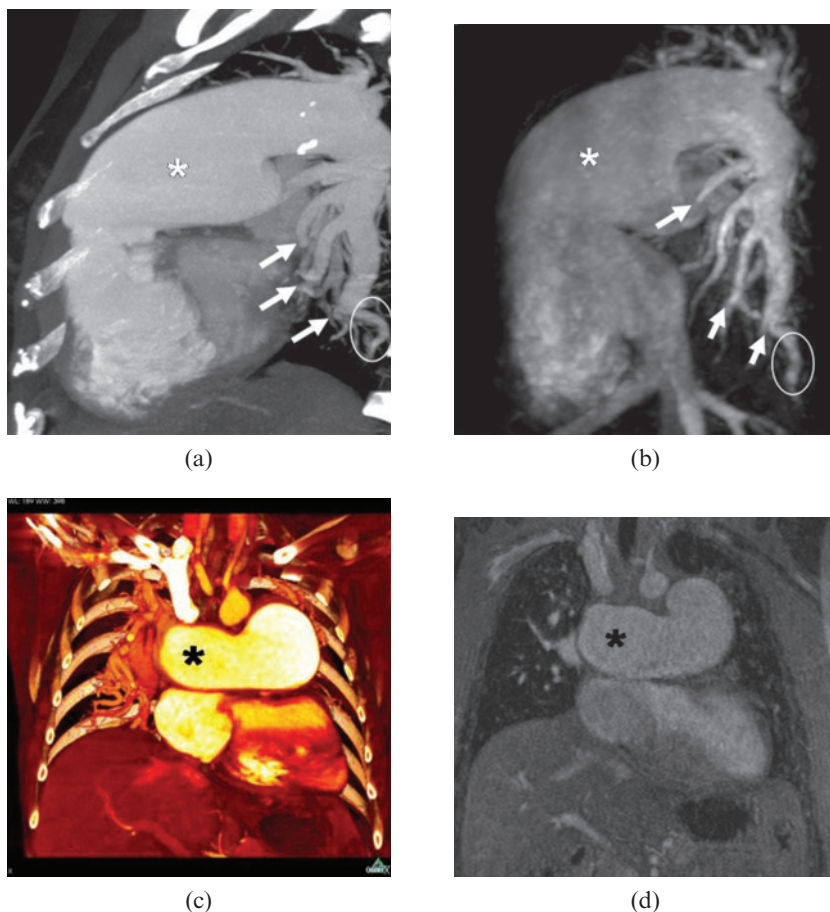


Figure 2. A 45-year-old female with idiopathic pulmonary arterial hypertension. Similar imaging aspects in multidetector CT angiography (MDCTA)-derived maximum intensity projection (MIP) (a), MR angiography (MRA)-derived MIP (b), MDCTA-derived multiplanar reconstruction (MPR) (c) and coloured MRA-derived MPR (d) show marked dilatation of the pulmonary artery main stem (white asterisk) and the right pulmonary artery (black asterisk), proximal calibre changes (pruned tree sign; white arrows on a, b), peripheral vessel reduction and vessel tortuosity (white circles on a, b). No occlusive criteria are visible in MRA or MDCTA images.

similar to those of IPAH, such as progressive exertional dyspnoea, chest pain, cough, syncope and cor pulmonale. The management of patients with secondary PH is directed at the early recognition and treatment of the underlying disease (when it is still potentially reversible). For instance, left ventricular dysfunction should be treated with afterload-reducing agents, digoxin and diuretics. Surgery to correct structural cardiac and pulmonary anomalies can also be effective, and thromboendarterectomy in accessible chronic thromboemboli is potentially curative [18, 23, 33, 34].

The choice of surgical treatment depends on the location of the obstruction (central *vs* more distal pulmonary arteries), the correlation between haemodynamic findings and angiographically assessed degree of mechanical obstruction, comorbidities, the patient's willingness and the surgeon's experience [35, 36].

Patients who are not candidates for surgery may benefit from PAH-specific medical therapy [37, 38], but the use of these medications in patients with CTEPH requires further evaluation in randomised controlled trials [39].

The bronchial circulation is increased in CTEPH owing to the development of systemic-pulmonary anastomoses, which help to maintain pulmonary blood flow. The radiological findings are more localised and consist of material adhering to the thrombotic wall, wall irregularities, vascular webs and stenosis (Figures 3–6), as well as abnormal proximal-distal tapering (abrupt change in vessel diameter) with perfusional defects and prominent bronchial arteries (Figure 7). Additional

findings include right heart hypertrophy and enlargement with bowing of the interventricular septum, mosaic perfusion, air trapping, bronchiectasis and subpleural consolidation from prior pulmonary infarction [18, 23, 33].

Imaging in pulmonary hypertension using 64-multidetector CT angiography, MR angiography and MR-derived pulmonary perfusion

MR and CT acquisition protocols

According to the recent literature, MRA imaging for the diagnosis of PH can be performed using a high-resolution, breath-hold, contrast-enhanced, three-dimensional (3D), rapid, low-angle shot [repetition time (TR)/echo time (TE), 2.9/1.2 ms; flip angle, 25°] [16] obtained in the coronal orientation using an integrated parallel acquisition technique. This technique combines the generalised, autocalibrating, partially parallel acquisition (GRAPPA®; Siemens Medical Solutions, Erlangen, Germany) technique with sensitivity encoding (SENSE®; Siemens Medical Solutions, Erlangen, Germany) [31, 32]. MRPP images can be obtained using high-field MR scanners (>1.5 T) with high-performance gradients (preferably 30–40 mT). Patients enter the scanner head-first in the supine position. A special receiver coil is necessary for parallel imaging.

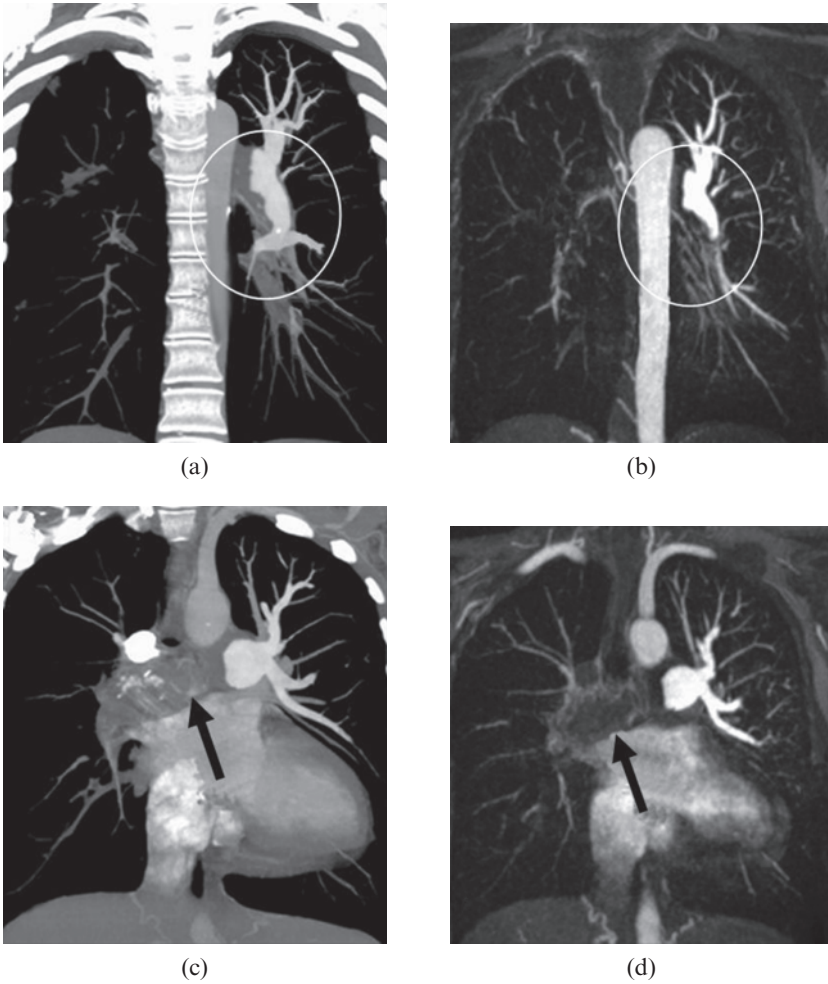


Figure 3. A 47-year-old female with secondary chronic thromboembolic pulmonary hypertension. Thin 10-mm maximum intensity projections derived from multidetector CT angiography (a, c) and MR angiography (b, d) images show thrombus in the left pulmonary artery that descends toward the basal trunk (white circles on a, b) and complete occlusion of the right pulmonary artery (black arrows on c, d).

For dynamic perfusion imaging (MRPP), Nikolaou et al [16] suggested the use of a turbo-speed, low-angle shot, gradient-echo sequence (TR/TE, 1.7/0.6 ms; flip angle, 25°) with an enhanced *k*-space-based reconstruction sequence (GRAPPA) and an acceleration factor of 2

[16]. The following further sequence parameters for perfusion MRI have been suggested: readout bandwidth, 1220 Hz per pixel; number of reference *k*-space lines for calibration, 24; section thickness, 4.0 mm; number of sections, 24; resulting slab thickness, 96 mm; acquisition

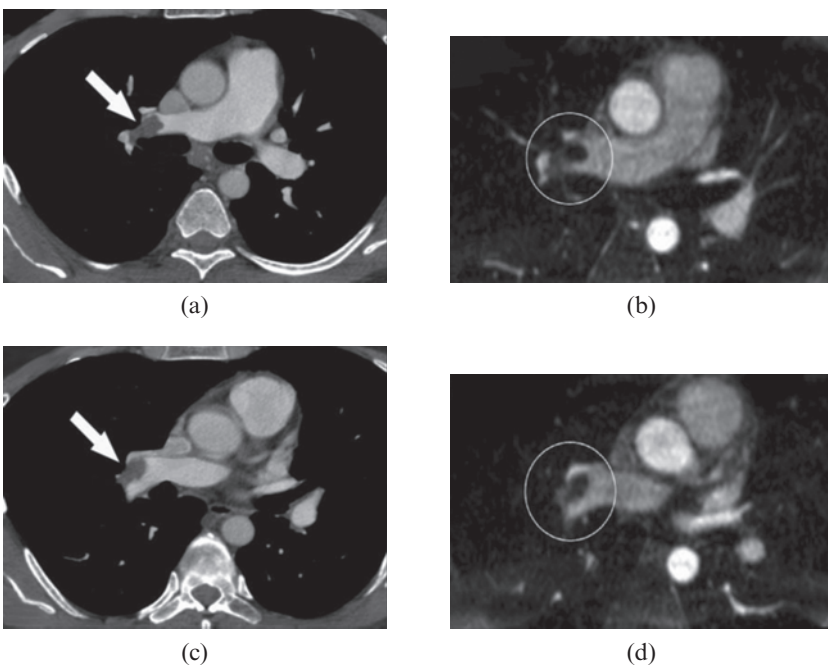


Figure 4. A 53-year-old male with secondary chronic thromboembolic pulmonary hypertension. Multiplanar reconstructions derived from multidetector CT angiography (a, c) and MR angiography (b, d) images show the similar aspect of the thrombus in the right pulmonary artery bifurcation (white arrows on a, c; white circles on b, d).

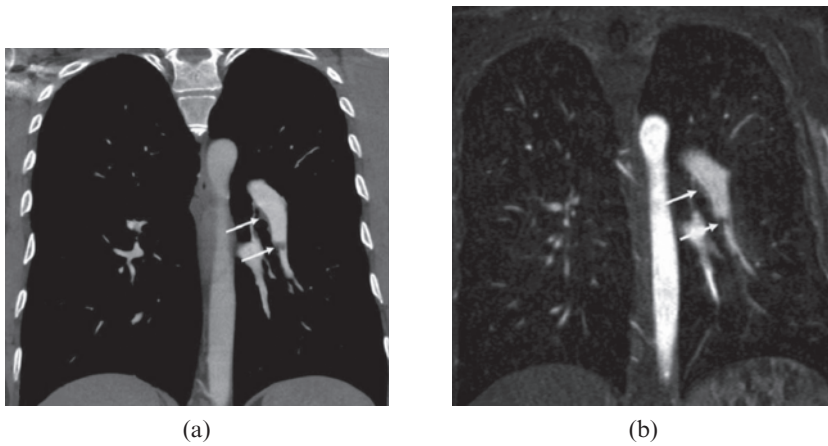


Figure 5. A 34-year-old female with secondary chronic thromboembolic pulmonary hypertension. Thin 10-mm maximum intensity projections derived from multidetector CT angiography (a) and MR angiography (b) images show a similar aspect of a small thrombus in the left basal trunk (white arrows).

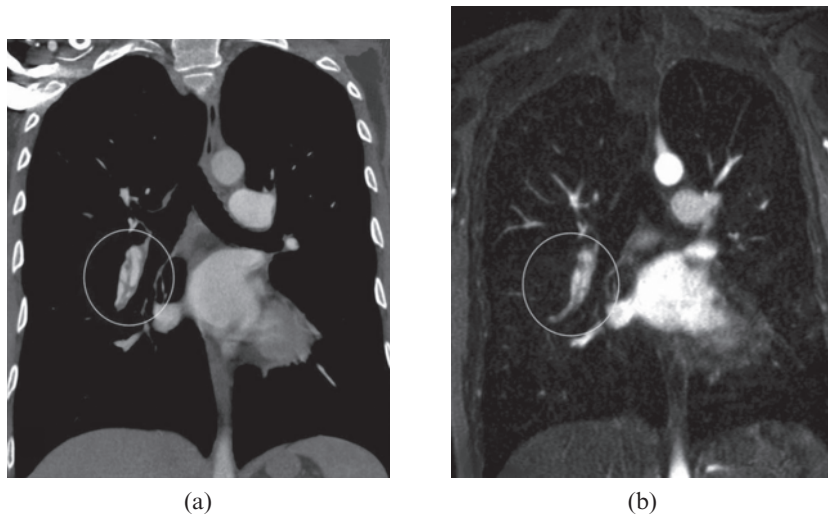


Figure 6. A 41-year-old female with secondary chronic thromboembolic pulmonary hypertension. Thin 10-mm maximum intensity projections derived from multidetector CT angiography (a) and MR angiography (b) images show a similar aspect of vascular bands (white circles).

matrix, 133×256 (52% phase resolution); field of view, 400 mm; and resulting spatial resolution, 2.9×1.5×4.0 mm. The *k*-space readout scheme is centrally reordered, and the acquisition time for one 3D data set of 24 sections is 1.1 s. A total of 24 slabs are acquired for each perfusion series, resulting in a total breath-hold time of 26 s. Image acquisition must be started 4 s before the estimated arrival time of the contrast material, so that unenhanced images can be acquired for subtraction purposes. All injections should be performed using a power injection system with 0.1 mmol gadobutrol or 0.2 mmol gadoterate

meglumine per kilogram body weight, followed by a 25-ml saline flush. To ensure an optimised bolus profile, the use of a high injection rate (5 ml s^{-1}) in combination with a 16-gauge intravenous catheter that can be inserted in an antecubital vein is recommended. Unenhanced images are subtracted from the contrast-enhanced images. For each phase ($n=25$), maximum intensity projections (MIPs) for all 24 sections are acquired for 3D display [16].

64-MDCT images can be obtained using standard protocols defined by the scanner manufacturers. The

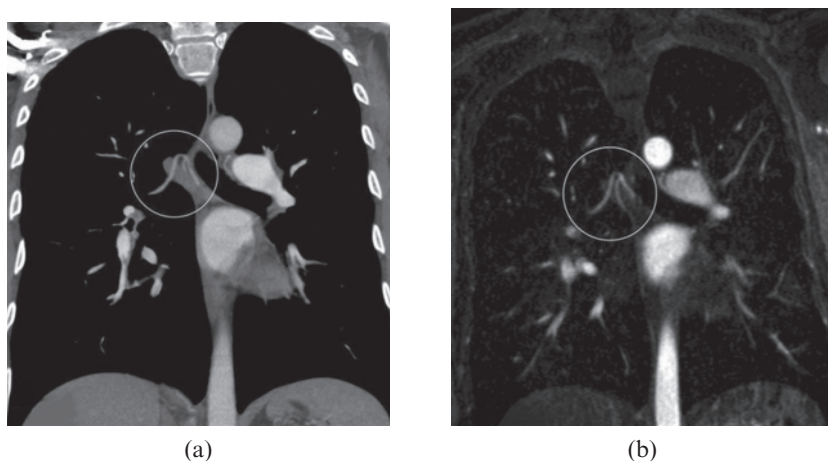


Figure 7. A 57-year-old male with secondary chronic thromboembolic pulmonary hypertension. Thin 10-mm maximum intensity projections derived from multidetector CT angiography (a) and MR angiography (b) images show a similar aspect of bronchial artery hypertrophy (white circles).

following parameters for a 64-row detector CT scanner were adapted from the recent literature [40]: caudocranial scanning direction, 120 kVp, 200 mAs, 0.5 mm collimation, 0.5 s rotation time (acquired during suspended breath after deep inspiration), 1.0 pitch, 1-mm reconstruction thickness and 0.5-mm reconstruction increment. A 125-ml injection of iodinated contrast is administered through an antecubital vein at a flow rate of 5 ml s^{-1} . A bolus-tracking technique was used to determine the interval between the initiation of contrast material administration and that of scanning; measurements were obtained at the origin of the pulmonary artery, and an 8 s delay was added to the circulation time before scanning.

MRA and MDCTA images must be evaluated on dedicated workstations, depending on the scanner manufacturer, using multiplanar reconstruction, MIP and the volume-rendering technique.

Comparison of CT and MRI for the diagnosis in pulmonary hypertension

CT is a non-invasive imaging technique that allows evaluation of both the pulmonary circulation (by MDCTA) and the lung parenchyma. MDCTA is widely regarded as the technique of choice for the evaluation of acute and chronic pulmonary vascular disease. It also aids differential diagnoses that include conditions such as pulmonary artery sarcoma, extrinsic compression of pulmonary vessels and fibrosing mediastinitis. The success of thoracic MDCTA is the result, in part, of the high spatial resolution of CT, and advances in multi-section technology have dramatically increased the speed and simplicity of this technique [41–43]. However, its main disadvantage is the use of ionising radiation and iodinated contrast material.

In the late 1970s and early 1980s, several investigators used MDCTA findings to measure the diameter of central pulmonary arteries and to predict the presence of PH [36–40]. The calibre of the pulmonary artery, the artery-to-bronchus ratio and the presence of pericardial effusion are associated with the severity of PH. Tan et al [44] also showed that CT-determined mean pulmonary artery diameter had excellent diagnostic value for the detection of PH in patients with advanced lung disease. However, the use of pulmonary artery diameter has limitations. Pulmonary artery dilatation can occur in the absence of PH in patients with pulmonary fibrosis, and is therefore an unreliable sign of PH in these patients [45]. Thus, standard chest CT scans may be used to screen for PH as a cause of exertional limitation in patients with parenchymal lung disease. Although MDCTA is currently the method of choice for the evaluation of patients with CTEPH, it provides poor functional information [16]. Kuriyama et al [46] found that a CT-determined mean pulmonary artery diameter of ≥ 29 mm predicted PH with 87% sensitivity and 89% specificity. This specificity reached 100% when this mean pulmonary artery diameter was accompanied by findings of a segmental artery-to-bronchus ratio greater than 1:1 in three of four pulmonary lobes [46].

MDCTA distinguishes CTEPH from IPAH by detecting thrombotic material, evaluates underlying lung

disease and may help to identify rare causes of PH. MDCTA is quick, widely available and inexpensive. However, it is associated with a significant radiation burden and is unsuitable for serial examinations [47]. MDCTA will thus probably continue to have a pivotal role in the evaluation of patients with pulmonary vascular disease. However, renal impairment or contrast agent sensitivity prevent the use of this technique in a subset of patients. In other patients, the radiation burden of MDCTA may be considered undesirable owing to young age or the prospect of multiple follow-up examinations [48].

Dual-energy CT (DECT) pulmonary angiography seems to represent an important technique in pulmonary angiography. This technique not only enables imaging of emboli in the pulmonary vasculature but also provides an indication of their functional impact in terms of parenchymal perfusion. The DECT pulmonary angiography technique is similar in many respects to standard CT pulmonary angiography. A slightly longer delay time is used (around 7 s) to allow contrast material to perfuse to the lung periphery, and a saline flush helps minimise beam-hardening effects in the superior vena cava that can mimic perfusion defects in the upper lobes. However, DECT pulmonary angiography offers a functional aspect to CT pulmonary angiography and may improve its sensitivity for detecting subsegmental emboli [49]. DECT perfusion imaging is also able to display pulmonary perfusion defects with good agreement to scintigraphic findings. DECT can also provide a pulmonary CT angiogram, high-resolution morphology of the lung parenchyma and perfusion information in one single examination [50].

MRA and MRPP are non-invasive imaging methods that allow the morphological and functional study of PAH patients and the examination of cardiac anatomy and functional parameters without the use of ionising radiation or iodinated contrast material [51]. The combined use of these techniques can facilitate the differential diagnosis of IPAH and CTEPH [17, 18, 52, 53]. MRPP can provide valuable information for the diagnosis of PH by demonstrating the exact extent of perfusion defects, and has increased diagnostic accuracy in the differentiation of various causes of PAH, depending on the specific patterns of perfusion defects [16, 54]. MRI of lung perfusion is performed with rapid imaging of the first passage of contrast material through the lungs after bolus injection into a peripheral vein. Despite substantial recent improvements in the gradient technique, 3D imaging techniques have been limited with regard to spatial and temporal resolution [16]. Sufficient temporal resolution is critical for the visualisation of peak enhancement of the lungs because the transit time of the contrast material bolus through the lungs is usually 3–4 s. Unlike conventional MRI techniques, parallel imaging techniques acquire only a fraction of the phase-encoding lines and reconstruct the missing information to a full field-of-view image by using the inherent spatial encoding of the different receiver coils. A temporal resolution of 1.1 s can be obtained using these techniques [16]. Despite these improvements in temporal resolution, the breath-hold time for acquisition of a complete data set in perfusion MRI remains quite long (approximately 26 s) [16]. Long breath-hold times may be

difficult for some patients with pulmonary disorders [16]. MRA allows the integration of morphological and functional information, which increases the diagnostic accuracy of the aetiological causes of PAH based on perfusion defect patterns [54].

MRA has become established as a powerful non-invasive tool for use in most vascular territories [55–57]. MRA was previously described as a highly sensitive and specific method for the diagnosis of acute pulmonary thromboembolism that does not require radiation exposure or the use of iodinated contrast media [52, 53]. However, the sensitivity of MRA was lower than that of digital subtraction angiography or MDCTA for the detection of corkscrew phenomena or subsegmental webs and bands. A comparison of state-of-the-art MRA and MDCTA of the pulmonary vasculature found that MDCTA remains superior because of the ease of use, short acquisition time and high spatial resolution [17]. MRA imaging, however, offers the great advantage of combining morphological and functional information, which makes the use of MRI favourable in the comprehensive assessment of complex diseases such as PAH [17].

In both imaging techniques, the anatomy and presence of PH-related occlusive (CTEPH associated) and non-occlusive changes of the pulmonary arteries should be evaluated [16], as well as the dilatation of the pulmonary arterial main stem (diameter >3.0 cm), peripheral vessel reduction (peripheral oligoemia), proximal calibre changes (IPAH-associated pruned tree sign), focal vessel ectasia, vessel tortuosity, irregularity of the vessel walls, bronchial artery hypertrophy and the occlusive criteria, including complete vessel occlusion, free-floating or wall-adherent thrombus, webs and vascular bands.

In patients with CTEPH, currently available MRI techniques can be used to evaluate various aspects of the disease. With contrast-enhanced MRA, the pulmonary arterial vasculature can be sufficiently analysed to the subsegmental level so that characteristic angiographic findings are well delineated. Furthermore, this technique can demonstrate the proximal extent of the organised thromboembolic material with great certainty,

a basic prerequisite for the identification of potential candidates for pulmonary endarterectomy [58].

Although MDCTA and MRA images can be very similar, we have found that thrombus detection is more difficult with MRA than with MDCTA because of the low signal intensity in MRI sequences. Careful evaluation and an experienced reader are required for accurate diagnosis. Figures 3–6 compare MDCTA and MRA images.

Perfusion data facilitate the visualisation of the characteristic loss of vascular perfusion. Advances in gradient systems and improved MR sequences have allowed the development of a 3D gradient-recalled echo sequence with an ultrashort TR/TE of <3.0/1.0 ms. With this sequence, qualitative regional perfusion differences in the entire lung can be evaluated with high temporal resolution [59, 60]. Diagnostic criteria for perfusion imaging in the differential diagnosis of IPAH and CTEPH have been modified from earlier perfusion scintigraphy reports [61]. The perfusion patterns (Figure 8) are classified as normal (no perfusion defect), diffuse peripheral and/or patchy (IPAH), or segmental and/or focal defect (CTEPH; Figure 9).

As described in previous articles [62, 63], there is good agreement between MRPP imaging and conventional radionuclide scintigraphy. However, MRPP imaging has various advantages over conventional scintigraphy, such as the avoidance of radiation exposure, higher spatial resolution and better anatomical information that allows data reconstruction in any desired imaging plane. In 2007, Ohno et al [64] showed that 3D dynamic contrast-enhanced MRPP has potential for the assessment of disease severity in patients with IPAH. Moreover, the quantitative assessment of regional pulmonary perfusion parameters from 3D dynamic contrast-enhanced MRPP images of patients with IPAH may offer the opportunity for non-invasive physiological and pathophysiological evaluation of the lungs with relatively high spatial resolution and without radiation exposure [64].

MDCTA and MRA are comprehensive and complementary methods of evaluating PH. MDCTA is superior to MRA in the visualisation of vascular abnormalities

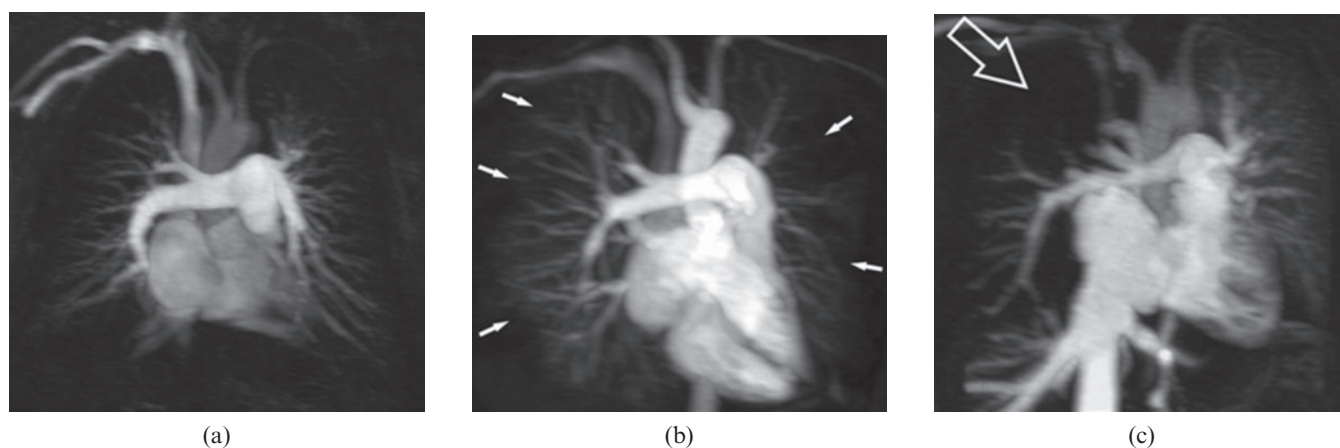


Figure 8. Evaluation criteria for pulmonary perfusion. Maximum intensity projection images of a single perfusion phase obtained with contrast-enhanced MR-derived pulmonary perfusion in three patients: (a) normal homogeneous perfusion (no perfusion defect), (b) bilateral diffuse non-segmental peripheral perfusion defects (typical of pulmonary arterial hypertension; white arrows) and (c) segmental or focal perfusion defect (typical of chronic thromboembolic pulmonary hypertension) in the upper right lobe (open arrow).

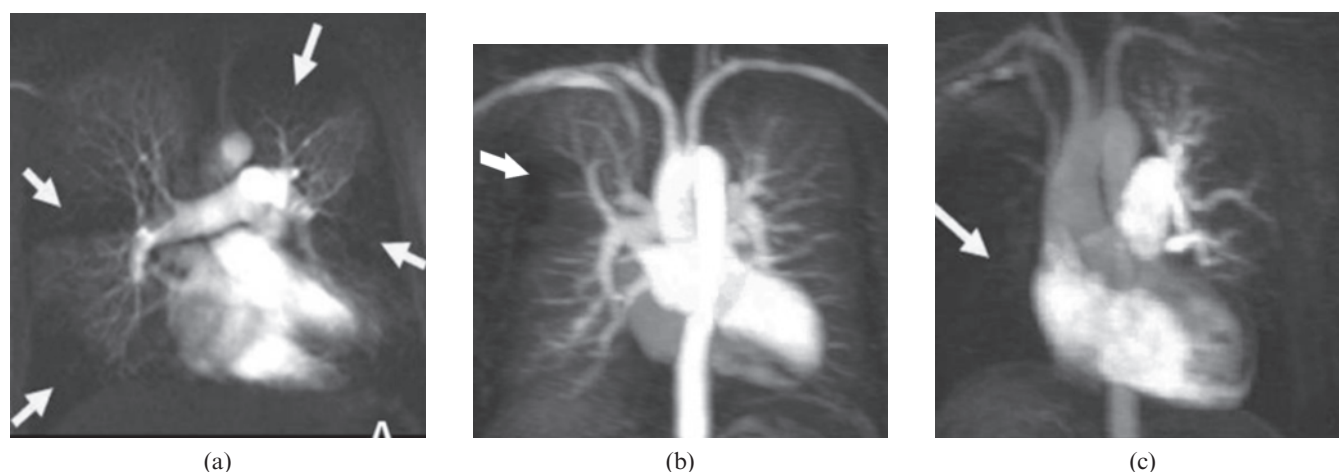


Figure 9. Perfusional aspect in chronic thromboembolic pulmonary hypertension. Maximum intensity projection images of a single perfusion phase obtained with contrast-enhanced MR-derived pulmonary perfusion in three patients: (a) multiple perfusion defects (white arrows), (b) focal perfusion defect in the anterior segment of the right lung upper lobe (white arrow) and (c) complete perfusion defect in the right lung (white arrow).

and the diagnosis of PH related to lung disease and bronchosystemic shunts [65]. However, the absence of ionising radiation and superior contrast profile favour MRA as a monitoring method. The use of MRA for other pathologies related to pulmonary circulation has not been investigated, and few studies have sought to determine its accuracy in the distinction of different forms of PH [16, 47, 58].

Conclusion

In this review, we compared the imaging findings of MRA and 64-row MDCTA in patients with known IPAH or CTEPH. We also detailed the MRPP imaging findings in these patients. MDCTA is currently considered the gold standard for the diagnosis of pulmonary embolism; however, we demonstrated that MRA can produce images similar to those of MDCTA in patients with IPAH or secondary CTEPH. Combined MRA and MRPP imaging allow the comprehensive evaluation of morphological and functional diagnostic information in patients with PAH. These can be helpful in the differential diagnosis and monitoring of the disease, and do not require the use of ionising radiation or iodinated contrast material.

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