Pulmonary hypertension: role of computed tomography and magnetic resonance imaging

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Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) have a leading role in the diagnosis and evaluation of pulmonary arterial hypertension. Technical aspects, advantages, limitations and potential contraindications will be considered.

MDCT has many advantages: 1) fast examination, 2) good identification of central and peripheral vessels, 3) good characterization of parenchymal findings, and 4) good evaluation of the heart and mediastinal structures. Limitations are: 1) the use of iodinated contrast material, and 2) radiation exposure. MRI allows: 1) cardiac morphological and functional studies, and 2) identification of central pulmonary arteries. Limitations are: 1) long scanning time, 2) poor definition of peripheral arteries, and 3) impossibility of pulmonary evaluation.

MDCT and MRI findings allow: 1) quick diagnosis of pulmonary arterial hypertension, 2) differential diagnosis between primary and secondary forms, 3) evaluation of cardiac manifestations, and 4) morphological and functional follow-up studies in surgically treated and untreated patients.

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Introduction

Traditionally the main radiological modalities in the diagnosis of embolic pulmonary arterial hypertension (PAH) have been chest radiography and pulmonary angiography. The latter still today is referred to as the gold standard technique for anatomic and functional evaluation of the pulmonary vessels.

In the last few years computed tomography (CT) and magnetic resonance imaging (MRI), thanks to their technical improvement, have assumed a more important diagnostic role in the study of pulmonary vascularization.

The aim of this critical review is to analyze: a) technical features of CT and MRI; b) advantages, limitations and potential contraindications of both modalities; c) the results of CT and MRI in the diagnosis of thromboembolic PAH.

Technical features of computed tomography and magnetic resonance imaging

The recent technical improvement of CT and MRI machines, together with the development of new software has definitely changed the quality of diagnostic information. Today for a good quality study of pulmonary hypertension the use of multidetector CT (MDCT) and fast single breath-hold sequence in MRI are mandatory, together with magnetic resonance angiography sequence, functional and flow evaluation[1].

Multidetector computed tomography. *Advantages.*

• Only a single scan, with high time resolution, is needed to evaluate all the thoracic aspects necessary to make diagnosis. We do not need separated and repeated scans for the heart, pulmonary vessels, lung and pulmonary interstitium. The volume acquired allows multiplanar and three-dimensional image reconstruction. Each structure analyzed can be evaluated in different planes. This information easily obtained allows a very precise analysis of what we are studying.

• Since the main pulmonary artery is intrapericardial, it cannot be measured on conventional radiography, but it can be easily identified on CT; in this way we can visualize central and also peripheral vessels, in particular lobar, segmental and subsegmental branches. Moreover in the axial images it is possible to evaluate the relationship between vascular structures and pulmonary secondary lobules, interlobular septa and lung interstitium. Bronchial arteries are easily detectable.

• Good visualization of the cardiac chambers, their morphology and volume, of the wall thickness and pericardium, using traditional axial planes, but also long- and short-axis views (the same used in echocardiography and cardiac MRI).
• Good visualization of the mediastinal structures, superior vena cava, lymph nodes, thoracic aorta and bronchial arteries.
• High time resolution allows good results also in sick patients, with severe dyspnea or conscious agitation. Easily repeatable and comparable examination.
• Technical easiness allows examination in acute and preoperative patients.

Limitations. MDCT exam for the evaluation of pulmonary arteries requires intravenous administration of contrast material; although a less quantity is needed than with single helical CT and conventional CT, accurate considerations of the renal function and potential allergic reactions are mandatory. Moreover the new modern MDCT does not allow to reduce the radiation dose, which remained the same.

Magnetic resonance[2].
Advantages.
• Excellent visualization of the heart, in particular of the morphology, structure and thickness of the cardiac walls, in addition to wall motion.
• Good definition of central pulmonary vessels, in particular of the main, right and left arteries and lobar branches.
• Fairly good identification of segmental and subsegmental branches. To obtain a better visualization of these structures, intravenous contrast medium gadolinium may be necessary.
• Good evaluation of the relationships between pulmonary vessels, the aorta and mediastinal extravascular structures.
• Functional study: blood flow in the pulmonary arteries and in the aorta, ejection fraction, cardiac output, left and right stroke volumes.
• No radiation exposure.
• Intravenous administration of contrast material generally well tolerated. This modality may be therefore of choice in patients for whom the use of iodinated contrast material required in CT may be not indicated.

Limitations.
• Long studies. More sequences are needed to study: a) the heart, b) the pulmonary vessels, and c) functional analysis. This modality may not be indicated in sick patients, with dyspnea or agitation.
• Poor or absent identification of peripheral branches, in particular of the terminal arterioles and their relationship with the lung parenchyma. Potential damage induced by pulmonary hypertension at this level may be therefore missed.
• No visualization of the pulmonary secondary lobule.
• Claustrophobia is still a significant limitation.
• This modality requires dedicated equipe with specific skills in cardiac radiology.
• Contraindications are the presence of heart devices, such as pacemakers, mechanical cardiac valves (Starr-Edwards), and any metallic structures related to prior cardiac or extracardiac surgery.

Computed tomography and magnetic resonance findings in pulmonary hypertension

In the diagnosis of pulmonary hypertension different aspects should be considered: a) vascular manifestations, b) parenchymal manifestations, and c) cardiac and mediastinal manifestations.

Vascular manifestations. Vascular manifestations include: caliber, lumen and walls of the arterial vessels[3].

Caliber. Central and peripheral arteries differ in wall structure, more elastic in the former, muscular in the latter. The anatomic-histological boundary between the two is not defined and may vary form patient to patient, it can be at the origin of segmental or subsegmental branches.

In PAH the caliber of the central arteries is enlarged. Measurement is usually obtained just before the vessel bifurcation, on a plain perpendicular to the vessel long axis. Main pulmonary artery caliber > 2.8 cm and two main branch caliber > 1.6 cm are generally considered dilated.

Morphologically the dilation may be diffusely uniform, from mild to severe degree. However in some cases only a circumscribed enlargement with aneurysmal appearance may occur. This finding is more frequent in PAH secondary to congenital cardiac disease (Eisenmenger syndrome).

Possible complications include compression on the laryngeal or phrenic nerve on the left with elevation of the hemi-diaphragm.

The caliber of the peripheral arteries may be abnormal too. The main criteria to determine peripheral arterial enlargement is the pulmonary artery-to-bronchus diameter ratio, 1 in normal conditions, > 1 if dilation of arteries is present. These measurements are easily and routinely assessed on axial images where the peripheral vessels are visualized on their short axis: the apical branch of the upper lobe, the basal branches of the inferior lobes.

Changes in peripheral pulmonary arteries are different according to the different types of pulmonary hypertension. In the primary form the caliber is usually not enlarged, but reduced, with rapid tapering of vessels from the center to the periphery. Sometimes this contrast in diameter between central and peripheral arteries is so sharply demarcated that it may simulate an intraluminal pseudo-web image.

In acute pulmonary embolism there is an increased diameter of the occluded vessels, while in the chronic

form there is a reduction of the arterial caliber due to thrombus retraction with possible distal complete occlusion (Fig. 1). On the contrary normal vessels tend to dilate due to flow redistribution.

Lumen and wall. In idiopathic PAH the lumen of the central and peripheral arteries may be normal[3].

In the acute setting of embolic PAH the lumen of one or more central peripheral vessels can be partially or completely obstructed. The diagnosis of acute embolic PAH is based on the presence of an intraluminal round filling defect surrounded by variable amount of contrast material, with acute angle with the vessel wall.

In chronic embolic PAH there is eccentric location of embolic material within the lumen, contiguous to the vessel wall, with an obtuse angle with the wall itself (Figs. 2-4). Rarely it may calcify. Moreover in chronic embolic PAH there is evidence of recanalization within the intraluminal filling defect, with intimal irregularities, arterial stenosis or abrupt vascular narrowing, causing proximal vessel dilation; this ends up in alternation of stenosis and dilation of the arteries, especially the peripheral ones (Fig. 5).

The progressive thrombus retraction leads to web formation, that are seen as lines of decreased opacity that traverse the width of the pulmonary artery usually at the lobar or segmental level; they are often associated with vessel narrowing or post-stenotic dilation (Fig. 6).

In pulmonary fibrosis MDCT may demonstrate marked wall irregularities and modifications of the vessel lumen. These abnormalities are usually quite diffuse in extension with an irregular reduction of arterial diameter and dilation of relatively spared arterial branches.

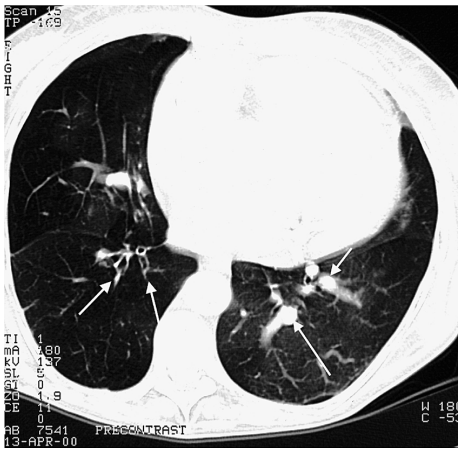

Figure 1. *Pulmonary high-resolution computed tomography image in embolic pulmonary arterial hypertension. Pulmonary segmental arteries* (arrows) *of the lower lobes are enlarged on the left side (flow redistribution) but very small and retracted by chronic occlusion on the right side.*

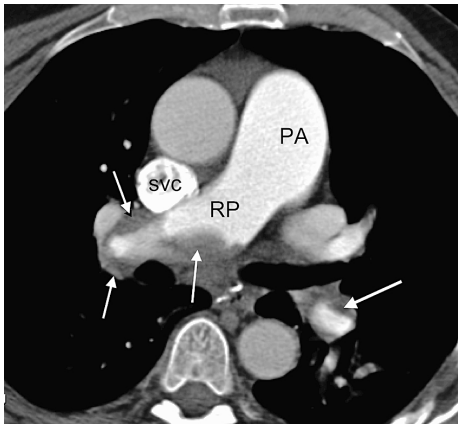

Figure 2. *Multidetector computed tomography in chronic embolic pulmonary arterial hypertension; axial image of the right pulmonary artery (RP). Thick mural thrombus* (arrows) *in the distal segment of the RP and in the left descending pulmonary artery. PA = main pulmonary artery; SVC = superior vena cava.*

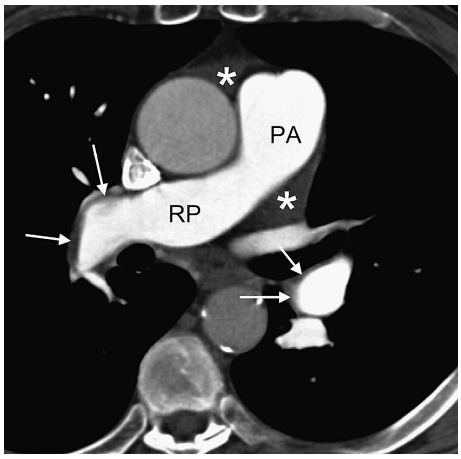

Figure 3. *Multidetector computed tomography in chronic embolic pulmonary arterial hypertension; thin and smooth mural thrombus* (arrows) *on the proximal segment of right descending pulmonary artery (RP) and on the distal segment of the left descending pulmonary artery (PA). * pericardial recesses.*

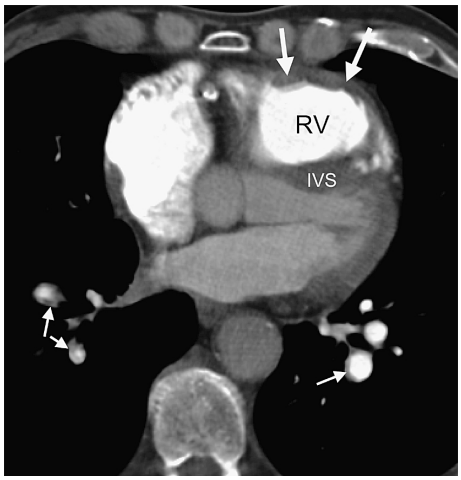

Figure 4. *Multidetector computed tomography in chronic embolic pulmonary arterial hypertension; thin mural thrombus* (arrows) *in the segmental arteries of the lower lobes. IVS = interventricular septum; RV = right ventricular outflow tract with hypertrophic wall* (thick arrows).

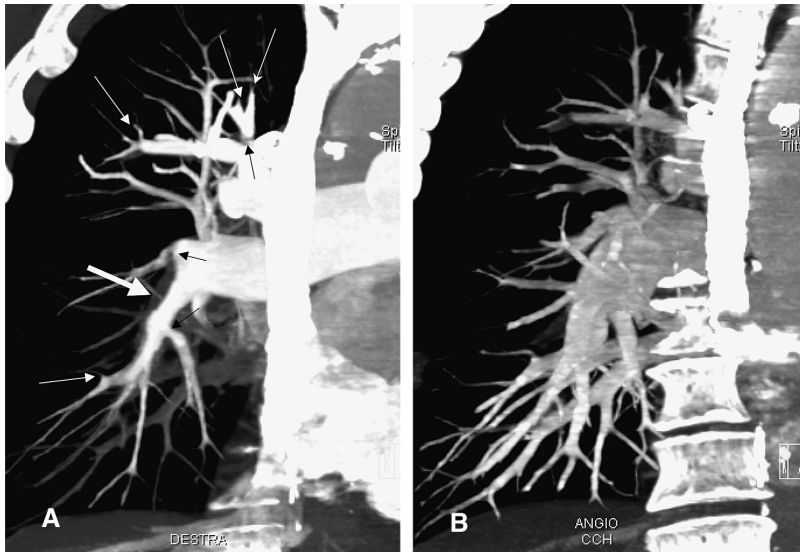

Figure 5. *Maximum intensity projection reconstructed images of multidetector computed tomography in chronic embolic pulmonary arterial hypertension before* (A) *and after* (B) *surgery. Thrombus in the right descending pulmonary artery* (thick arrow)*; proximal stenosis and dilations of the segmental arteries* (black thin arrows)*; subsegmental occlusions* (white thin arrows) *and diffuse narrowing in the peripheral arteries. Normal findings after surgery* (B).

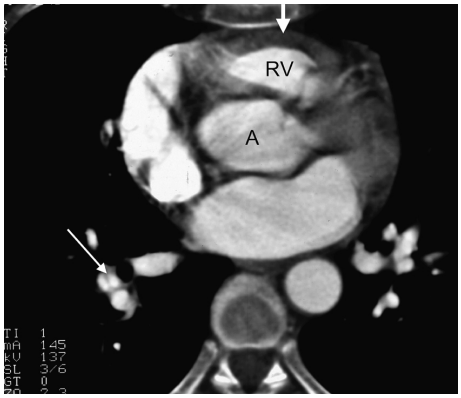

Figure 6. *Multidetector computed tomography in chronic embolic pulmonary arterial hypertension with thick right ventricular hypertrophy* (thick arrow)*. "Web" formation inside the lumen of the pulmonary arteries* (arrow) *is due to progressive thrombus retraction and vessel recanalization. A = aortic root; RV = right ventricle.*

The vascular lumen may also be invaded by endovascular spread of tumors such as sarcoma that causes stenosis, intraluminal filling defects and arterial occlusion, leading to PAH.

MDCT and MRI have a leading role in the differential diagnosis between embolic vascular obstruction and neoplastic occlusion. The former does not enhance after intravenous contrast injection, while the latter typically does.

Pulmonary parenchymal manifestations[4]. MRI does not have any role in the evaluation of the lung parenchyma, because of the lack of signal from the air.

MDCT on the contrary provides information on lung abnormalities, allowing analysis of the finest functional unit, the secondary pulmonary lobule, with the central arteriole, the interlobular septa and veins.

In embolic PAH lung findings can be normal in a few cases. More often parenchymal manifestations are present; these can be summarized in:

1) sub-pleural lung scarring. These are small parenchymal opacities, with distortion of septa, abutting the pleural surface. They are the result of prior lung infarcts and may be more frequent in chronic pulmonary embolism studies (Fig. 7);

2) mosaic perfusion. It consists of localized areas of decreased attenuation and vascularity that are sharply marginated from adjacent areas with normal or increased attenuation and enlarged vessel size secondary to blood redistribution (Fig. 8). This abnormality can be found either in the lung periphery or in the central portions, at the lung apex or at the bases[4].

Mediastinal and cardiac manifestations. In chronic embolic PAH bronchial arteries, originating from the thoracic aorta, are usually dilated (Fig. 9); they may ap-

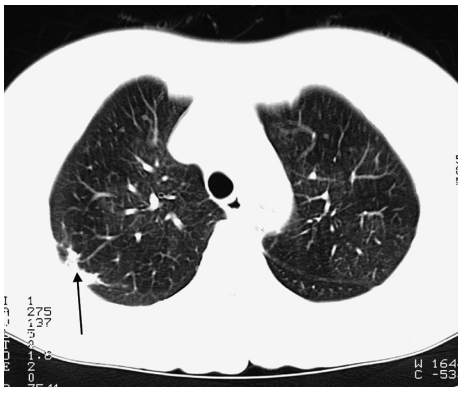

Figure 7. *Pulmonary high-resolution computed tomography. Subpleural lung scars* (arrow) *and asymmetry of the pulmonary vessel caliber are common findings in chronic embolic pulmonary arterial hypertension.*

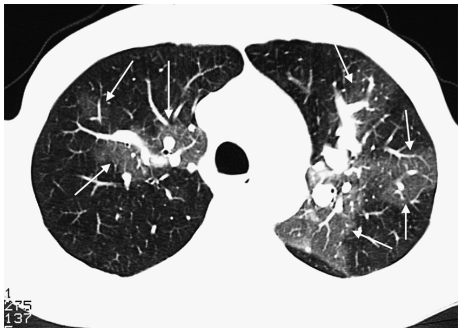

Figure 8. *"Mosaic perfusion" in pulmonary high-resolution computed tomography. Areas with large vessels and increased attenuation represent blood redistribution* (arrows) *from the adjacent occluded vascular areas (decreased attenuation and thin vessels).*

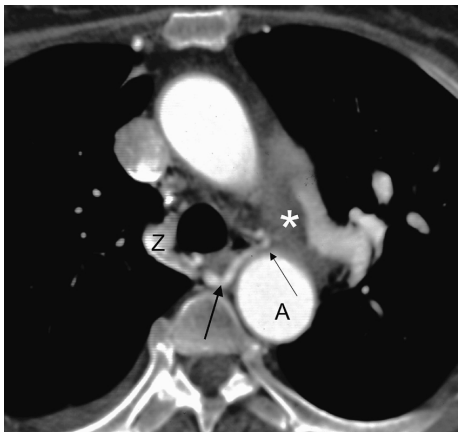

Figure 9. *Multidetector computed tomography. Dilated bronchial arteries* (arrows) *are a common finding in chronic embolic pulmonary arterial hypertension. A = descending aorta; Z = azigos vein. * thrombus in the left pulmonary artery.*

pear tortuous. MDCT scans allow the identification of the most peripheral portions, where they are in relationship with the bronchus, sometimes with a larger diameter than the occluded pulmonary vessels. This is a specific finding of chronic embolic PAH, helping in differentiating the latter from other forms[5].

In chronic embolic PAH and idiopathic veno-occlusive disease mediastinal lymph nodes may also be enlarged; this finding is rare in primary PAH[6]. In PAH alterations of the right heart are frequent and relevant. Both CT and MRI allow good evaluation of cardiac morphological, volumetric and dynamic manifestations of pulmonary hypertension[2].

In acute embolic PAH the right ventricle may be enlarged to different degrees. The wall does not show usually structural alterations. The increased right ventricular volume leads to heart rotation on the longitudinal axis toward the left. The cardiac apex and the left ventricle are shifted to the left and backward. The interventricular septum presents horizontal orientation, loosing the normal convexity toward the right ventricle.

With cine-MRI sequence sometimes a paradoxical movement of the septum is detected.

All these alterations may interfere with the normal aspect of the left ventricle, which is compressed and shifted by right ventricular enlargement. The right atrium is usually dilated too. This enlargement may interfere with the normal function of the tricuspid valve that may become insufficient. This finding is more easily identified with cine-MRI sequences.

In addition the dilated right atrium may cause enlargement of the coronary sinus and of the superior and inferior vena cava.

In chronic embolic PAH a hypertrophic right ventricle is present with increased wall thickness. This finding is usually uniform, but may be particularly evident at the level of the infundibulum (outflow tract). Hypertrophy includes increasing thickness of the trabecula and papillary muscles, which are easily identified both on CT and MRI. It usually presents as irregularly reticular, more evident in the right ventricular outflow tract (Figs. 10 and 11).

The increased volume of the right chambers, in addition to tricuspid valve insufficiency, are present also in the chronic form, although usually less evident than in acute embolic PAH.

However, cardiac alterations described in acute and chronic embolic PAH are not specific findings and may be present in any cause leading to PAH. The pericardium usually is normal in PAH. Sometimes in the primary form a mild degree of pericardial effusion in the cranial recess may be present, with increased thickness of the latter up to 10-15 mm.

Role of multidetector computed tomography and magnetic resonance imaging

Diagnosis of pulmonary embolism in dyspnoic patients. PAH can be suspected on the basis of clinical

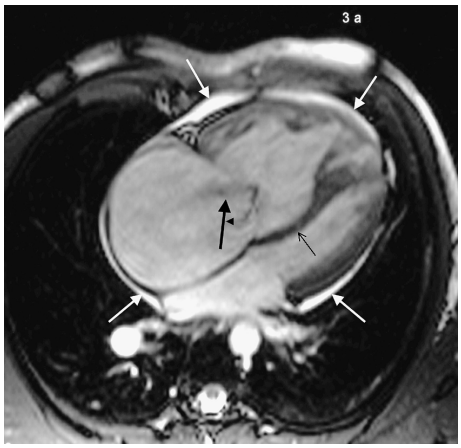

Figure 10. *Gradient-echo 4-chamber view in cardiac magnetic resonance. Chronic embolic pulmonary arterial hypertension. Dilation of the right heart with tricuspid valve insufficiency* (large arrow) *and paradoxical convexity of the interventricular septum* (thin arrow). *Pericardial fluid* (white arrow).

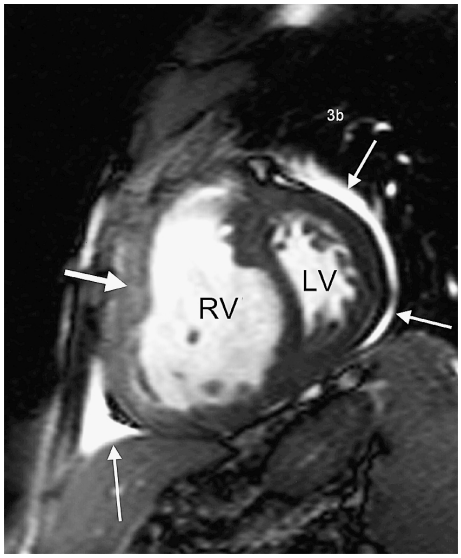

Figure 11. *Double-oblique short-axis view in gradient-echo cardiac magnetic resonance in the same patient as figure 10. Right ventricular (RV) enlargement impresses the left ventricle (LV). Severe hypertrophy of the right ventricular wall* (thick arrow) *and paradoxical convexity on the interventricular septum. Pericardial fluid* (thin arrows).

symptoms and laboratory results. Clinical presentation can be variable and non-specific. Usually dyspnea is the most common reported symptom, followed by chest pain, cough and, less frequently, hemoptysis. However these symptoms are also common in many other conditions involving the cardiovascular and bronchopulmonary systems, thus decreasing their usefulness.

CT is the modality of choice not only to confirm the clinical diagnosis, but also to identify the embolic origin of PAH and quantify its manifestations[1,2,4].

Differential diagnosis in pulmonary arterial hypertension. As mentioned before, CT allows good evaluation of the pulmonary arteries from the center to the periphery. The relationship between the pulmonary arteries-lung parenchyma and the secondary pulmonary lobule is characteristic in different types of PAH. Therefore CT studies can lead to quick diagnosis, differentiating primary pulmonary hypertension from the secondary ones (i.e., embolic disease, chronic bronchopulmonary and interstitial diseases, veno-occlusive disease)[2,3,6].

Role of end-inspiratory and end-expiratory scans. End-inspiratory and end-expiratory scans are useful to differentiate peripheral thromboembolic disease from distal airway obstruction. In the former, expiratory scans do not show differences in lung densities, either in vascular obstructive and non-obstructed areas. In the latter, different values of lung attenuation characterize the obstructed areas (increased translucency) from non-obstructed zones (reduced expansion and translucency)[4].

Importance of mediastinal and cardiac manifestations. As previously outlined CT and MRI well document morphological, structural and dynamic changes in the heart and right chambers, as well as pericardial and bronchial artery abnormalities.

MRI allows a good definition of end-diastolic volume, mass, thickness and ejection fraction of the right ventricle. Diagnostic accuracy and reproducibility are superior to other modalities in any clinical situation and anatomical condition.

Morphological and functional follow-up. In patients with chronic thromboembolic disease MDCT and cardiac MRI are the gold standard modalities: a) in selecting potential candidates to thomboendoarterectomy: MDCT well demonstrates the proximal or distal location of mural thrombi[7]; and b) for follow-up studies: MDCT is the modality of choice for the evaluation of pulmonary arteries, MRI for functional studies[7,8].

In conclusion, CT and MRI play a leading role in the diagnosis and evaluation of PAH, and may overlead pulmonary angiography. In the clinical setting of embolic PAH, the physician should be aware that MDCT allows better image quality and spatial resolution of segmental and subsegmental arteries than pulmonary angiography, which is an invasive modality with additional risks for the patient, and low reproducibility in follow-up studies.

In our opinion new advances in cardiovascular CT and MRI results may be expected in the future by the continuous technical progress.

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