Pulmonary hypertension: The contribution of MDCT to the diagnosis of its different types

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Received 16 February 2010; accepted 29 May 2010

Abstract

Pulmonary hypertension is characterized by progressive involvement of the pulmonary vessels that leads to increased vascular resistance and consequently to right ventricular failure. Vascular lesions are a common factor in a wide spectrum of diseases, and their result, pulmonary hypertension, is a severe clinical condition with a poor prognosis that worsens the normal course of the diseases to which it is associated (COPD, collagen disease, sarcoidosis, and congenital or acquired heart disease).

It is important for pulmonary hypertension to be diagnosed as early as possible because nowadays drugs can reduce mortality and improve the quality of life; furthermore, some types of pulmonary hypertension (e.g., chronic thromboembolism and those associated with some congenital heart diseases like left-to-right shunt) can be treated surgically.

In cases of suspected pulmonary hypertension, imaging methods can confirm the diagnosis, suggest a cause, help choose the most appropriate treatment, and monitor the response to treatment. This review describes the approach to pulmonary hypertension using different imaging techniques; special emphasis is given to the role of multidetector CT (MDCT), which makes it possible to study all the organs in the thorax in a single acquisition.

We review the radiological signs of pulmonary hypertension and the current (Dana Point) radiological criteria for classifying the type of hypertension based on alterations in the lung parenchyma, mediastinum, pleural spaces, and pericardium, as well as on the study of the chambers of the heart.

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Introduction

Pulmonary hypertension (PH) is defined as a group of diseases characterized by a progressive increase in the pulmonary vascular resistance leading to right ventricular (RV) failure and death. Functional capacity of the RV is the major determinant of prognosis in PH1,2.

Idiopathic PH is a rare condition, with an incidence of 2.5 to 3.5 cases per million population in Western countries. It is 2 or 3 times more common in women than in men, at 2.5 to 3.5 cases per million population in Western countries.

In case of clinical suspicion, we should follow a diagnostic strategy using different technical modalities aimed to assess the right ventricular function, differentiate precapillary PH from postcapillary PH, and to plan the treatment. The last classification was adopted during the 4th World Symposium on Pulmonary Hypertension held in February 2008 in Dana Point (table 1).

Histological changes

All the types of PH show identical pathological obstructive changes in the pulmonary microcirculation, defined as hypertensive pulmonary vascular disease. This similarity suggests a pathobiological process shared by the spectrum of PH diseases3-6. The exact process that initiates the pathological changes is unclear, but it is well known that the pathobiology is multifactorial and that increased vascular resistances, typical of this condition, are caused by vasoconstriction, obstructive remodeling of the vascular wall, inflammation and thrombosis7,8.

Proliferative vascular remodeling involves all the wall layers. The main pathological finding is abnormal and disorganized cell proliferation causing a marked wall thickening. Complex vascular lesions develop, including plexiform lesions that cause obstruction of small arteries and increased resistances.

It is unclear if the presence of microthrombi in pulmonary arterioles is the cause or the consequence of PH; however, there is no doubt that their presence contributes to the progression of the disease. Microthrombi are present in 50% of cases of idiopathic PH4 and are common in the Eisenmenger syndrome9.

Imaging techniques for the diagnosis and assessment of the disease

In case of clinical suspicion, we should follow a diagnostic strategy using different technical modalities aimed to...
screening of risk groups: first degree relatives, collagenosis (particularly scleroderma and CREST), congenital heart diseases and HIV infection. In portal hypertension, assessment prior to liver transplantation is essential as transplantation is contraindicated in case of PH in chronic liver disease.

The aims of echocardiography in PH are:

- Detection of elevated pulmonary pressure.
- Assessment of the RV function.
- Making a differential diagnosis, since the size of the heart chambers, valvular function, thickness and functioning of heart chambers may be assessed. This imaging modality also allows the study of intracardiac shunts and, with transesophageal echocardiography, the presence of ductus or other extracardiac shunts.

Therefore, echocardiography allows the etiological study of PH in case of congenital and left heart disease, provides valuable information on the severity and prognosis of PH and therapeutic outcome monitoring.

There are several methods for estimating the PA pressure using echocardiography and, in general, there is a good correlation with the pulmonary pressure measured by contrast-enhanced Doppler (in 10% of cases it cannot be measured); however, there is no exact correlation with the values obtained by right cardiac catheterization, as echocardiography tends to overestimate the pressures.

Hemodynamic study

This is the only method that may establish a definitive diagnosis of PH, as it measures the PA pressure directly; for this reason, it is the gold standard in the diagnosis of the disease and is essential prior to any specific treatment.

The hemodynamic study has three main goals:

- To determine RV and PA hemodynamics, directly measuring the PA pressure, the pulmonary capillary wedge pressure and the pulmonary resistance. To measure the cardiac output and to determine oxygen saturation in the heart chambers.
- To assess the vasodilatory response to the therapeutic agents (nitric oxide, epoprostenol).
- To rule out a left-right shunt as well as any significant left lesion, and to assess the coronary ostium.

In case of suspicion of chronic pulmonary thromboembolism (CPTE), pulmonary arteriography helps determine the location, extension and size of the thrombus and select candidates for thromboendarterectomy.

Pulmonary ventilation/perfusion scintigraphy

For many years this was the main technique for diagnosis of acute pulmonary emboli, but now it is frequently used in the screening for chronic thromboembolic PH (CTEPH), after anticoagulant therapy, after an acute episode of pulmonary

Table 1  Updated classification of PH (Dana Point, 2008)  

<table>
<thead>
<tr>
<th>1. Pulmonary arterial hypertension (PAH)</th>
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<tbody>
<tr>
<td>1.1 Idiopathic</td>
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<td>1.2 Heritable</td>
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<tr>
<td>1.2.1 BMPR2</td>
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<td>1.2.2 ALK-1, endoglin (with or without hereditary hemorrhagic telangiectasia.)</td>
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<tr>
<td>1.2.3 Unknown</td>
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<td>1.3 Drug- and toxin- induced PAH</td>
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<td>1.4 PAH associated with:</td>
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<td>1.4.1 Connective tissue diseases</td>
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<td>1.4.2 HIV infection</td>
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<td>1.4.3 Portal hypertension</td>
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<td>1.4.4 Congenital cardiac disease</td>
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<td>1.4.5 Schistosomiasis</td>
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<td>1.4.6 Chronic hemolytic anemia</td>
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<td>1.5 Persistent pulmonary hypertension of the newborn</td>
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<tr>
<td>1.5.1 Pulmonary venoocclusive disease and/or pulmonary capillary hemangiomatosis</td>
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<tr>
<td>2. PH owing to left heart disease</td>
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<tr>
<td>2.1 Systolic dysfunction</td>
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<td>2.2 Diastolic dysfunction</td>
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<td>2.3 Valvular disease</td>
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<tr>
<td>3. PH owing to lung diseases and/or hypoxia</td>
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<tr>
<td>3.1 Chronic obstructive pulmonary disease</td>
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<tr>
<td>3.2 Interstitial pulmonary disease</td>
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<tr>
<td>3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern</td>
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<tr>
<td>3.4 Sleep-Disordered breathing</td>
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<tr>
<td>3.5 Alveolar hypventilation disorder</td>
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<tr>
<td>3.6 Chronic exposure to high altitude</td>
</tr>
<tr>
<td>3.7 Developmental abnormalities</td>
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<tr>
<td>4. Chronic thromboembolic PH</td>
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<tr>
<td>5. PH with unclear or multifactorial etiologies</td>
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<td>6. Hematologic disorders: myeloproliferative disorders, splenectomy</td>
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<tr>
<td>7. Systemic disorders: sarcoidosis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neurofibromatosis, vasculitis</td>
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<tr>
<td>8. Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid diseases</td>
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<tr>
<td>9. Others: tumor obstruction, mediastinal fibrosis, chronic renal insufficiency on hemodialysis</td>
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</tbody>
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ALK-1: kinase 1 similar to activin receptor; BMPR2: Bone morphogenetic protein receptor type II; HIV: Human immunodeficiency virus.
thromboembolism, to determine the degree of pulmonary perfusion and to rescue those cases that might progress to CTEPH.16

A normal ventilation/perfusion scan (V/Q) practically rules out CTEPH. In contrast, CTEPH is the most likely diagnosis if the V/Q scan shows multiple bilateral perfusion defects, segmental or larger, with normal ventilation; however this finding is not specific of CTEPH and may occasionally appear in other types of PH such as venoocclusive disease, or other conditions including sarcoma or vasculitis of the PA or fibrosing mediastinitis.13,14

In cases of PH associated with congenital heart disease or idiopathic PH, perfusion is normal or minimal defects appear.14,15

**Magnetic resonance (MR)**

MR has the advantage of being a morphological and functional method; for this reason, some authors propose MR as the gold standard for the assessment of the RV in PH.2

Contrast enhanced MR allows visualization of mediastinal and pulmonary vessels and a functional study of the heart chambers.4,11,15 Planimetry using cine MR allows measurement of end-systolic and end-diastolic volumes, ejection fraction, stroke volume, muscular mass and changes in the movement of the ventricular wall or the interventricular septum. It may also assess systemic and pulmonary flow in right-left or left-right shunts and shows good correlation between the parameters of the curve of PA blood velocity and the degree of PH and resistances.17

MR is superior to echocardiography in the assessment of the remodeling of the RV wall.

The main advantage is that MR involves no ionizing radiation and makes the ideal technique for young or sick patients requiring serial assessment. Its major disadvantage is lower spatial resolution than computed tomography (CT) and angiography, making difficult the assessment beyond the segmental level and the pulmonary parenchyma. Other drawbacks of MR, in comparison with CT, are the low availability of MR equipments, the duration of the examination that, along with the enclosed gantry, require patient collaboration and might hinder the exam due to claustrophobia.18 As MR is a non-invasive method, it may be used repeatedly to assess disease progression and treatment response.2,6

**Multidetector computed tomography (MDCT)**

MDCT allows volume acquisition. Highly reliable multiplanar reconstructions in all planes may be acquired using <1 mm collimation. The creation of slabs or slices of variable thickness with selection of Maximum-Intensity-Projection points (slab-MIP) is very helpful to: follow very small arteries including subsegmental vessels or the bronchial or intercostal arteries, assess the cardiac chambers, ventricular wall, and pulmonary venous drainage and perform a high-resolution study of the lung parenchyma. This information provides an approach to etiological diagnosis of PH and, for this reason, the new ESC/ERS Guidelines propose a thorax CT from the moment PH is suspected.2,4

Particularly, in the case of HPTEC, CT angiography (CTA) provides a reliable road map of localization and extension of thrombi, intraarterial webs and stenosis. In the assessment of non-occlusive or calcified mural thrombi, CTA supersedes angiography. In addition, CTA is an appropriate non invasive method for assessing the outcome of endarterectomy or other surgical procedures.

CT provides only a morphological study, but recently, MDCT dynamic study with cardiac synchronization has been proposed for the functional evaluation of the systolic and diastolic ventricular volumes, ejection fraction and ventricular mass,19 and even to assess the severity of PH by measuring PA distensibility during systole and diastole and correlating it with the mean PA pressure.19

**MDCT technique**

Studies are performed on CT units with panels of 4, 8, 16 and 64 detectors. The examination always requires breath holding, and is performed either in a craniocaudal direction, following the entrance direction of the IV contrast, or caudocranial direction in order to obtain an early visualization of the distal vessels of the inferior lobes, where subsegmental emboli are more common. This fact is more relevant for the study of acute PE than of HPTEC. The study should include from the supra-aortic trunks to the drainage site of the hepatic veins into the inferior vena cava (IVC). Collimation should be the lowest allowed by the scanner, with ≤1 mm reconstructions and 50 % overlap, which enable high-resolution multiplanar reconstructions. Radiation dose is 100–120 KV and 70–200 mAs with dose modulation and several examination protocols have been proposed in order to minimize radiation risks.20

Intravenous non-ionic contrast agent is used, at 350–370 mg/cc iodine concentration and 4 cc/s injection rate. Optimal image acquisition time is determine using the bolus-tracking technique, placing the cursor on the PA21 and starting the acquisition after reaching a 150–180 HU threshold. Image analysis is carried out in the work station with windows for the pulmonary parenchyma (W1600/ L-600), soft tissues (W500/L 35) and angio-Graphic (W700/L 100). Multiplanar reconstructions are obtained using the Maximum Intensity Projection (MIP) and the Minimum Intensity Projection (miniP) techniques, at different thickness, and also using volume rendering.

**Diagnosis of PH using MPCT**

1. Vascular changes

- Initially, the first radiological sign assessed was the PA diameter. Main PA diameter should be measured in its bifurcation, at a right angle to its long axis and just lateral to the ascending aorta. According to Kuriyama,22 a PA diameter ≥ 29 mm has a PPV of 0.97, 87 % sensitivity and 89 % specificity in predicting PH; for this reason, this value has been used as indicative of PH. However, it has been proven that absolute measures are not completely reliable, as PA pressure and size depend on the body mass, sex and
age. It seems to be increased by 6% in healthy individuals over the age of 50 and in obese patients. It also increases with exercising and in athletes.

- Specificity climbs to 100% when a PA diameter ≥ 29mm is accompanied by a segmental artery-to-bronchus ratio > 1:1 in most lung lobes.
- It is better to compare the main PA with the adjacent aorta (Ao) and if the PA/Ao ratio is > 1, HP is very likely, particularly in patients below the age of 50.
- Another recent parameter is the assessment of the PA at the level of the aortic arch (egg — PA — and banana — Ao — signs), seen in severe PH.
- Within the parenchymal vessels, Sheehan reports the phenomenon of “neovascularization”, which describes serpiginous and fine peripheral vessels that frequently emerge from the centrilobular arterioles, without following the normal anatomy of the pulmonary vessels. Originally, Sheehan described these findings in idiopathic PH and in the Eisenmenger syndrome; however, they seem to be a manifestation of all-cause severe PH (fig. 2A).
- Peripheral anastomoses of systemic intercostal vessels that supply the lung periphery (fig. 2B).

2. Cardiac changes

- RV functional changes seem to be the main determinant of the progression and prognosis of the disease. Although the pulmonary vascular bed is the leading cause of the disease, the symptoms and prognosis are strongly related to the pump function of the RV. Chronic pressure overload of the RV leads to changes in the cardiac morphology and function affecting both ventricles. Structural changes of the right chambers include hypertrophy and RV dilatation, right atrium (RA) enlargement and functional tricuspid regurgitation, secondary to valvular ring dilatation. Progressive increase in RV pressure damages the structure and function of the left ventricle (LV), which reduces its size and distorts.
- The RV/LV size ratio needs to be measured. This may be done on axial slices, but four-chamber plane measurements correlate better with echocardiographic measurements. Under normal conditions, LV diameter is wider than that of RV (fig. 3).
- Interventricular septum position: As the pressure in the RV increases, the normal position of the septum varies and at very high pressures septal inversion occurs, resulting in LV dysfunction, with reduction of its volume/minute (output), which is a very serious condition. Septal inversion is a bad prognostic sign (fig. 3).
- Increased RV wall thickness. Values > 4 mm are considered pathological (fig. 3).
- RA enlargement and regurgitation of contrast agent into the IVC and hepatic veins is indicative of tricuspid valve insufficiency with 90% sensitivity and 100%

![Figure 1](http://www.elsevier.es) Axial image shows a wider caliber of the PA trunk than that of the ascending aorta. PA/Ao > 1. Pulmonary artery > 29mm.

![Figure 2](http://www.elsevier.es) Vascular changes (Axial and axial oblique MIP reconstructions). A) In the mediastinum, the pulmonary artery (AP) appears at the level of the aortic arch (egg and banana sign). Discrepancy in caliber between the central and peripheral vessels. Neovessels in the lung periphery, which originate at the center of the secondary pulmonary lobule and show a serpiginous and irregular distribution (black arrows). B) Peripheral vessels with a corkscrew configuration. Anastomosis (arrow) between a peripheral vessel and a systemic one (intercostal).
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Along with vascular changes, changes in the bronchial diameter, such as dilatation of the bronchial lumen adjacent to the occluded vessel, may also be seen in CTEPH. These latter are bad prognostic signs.

**Figure 3** Cardiac changes. Four-chamber plane measurements of the heart chambers: RV larger than LV with thickened wall (black dash), interventricular septal inversion (black arrow) and small pericardial effusion (white arrow). These latter are bad prognostic signs.

**Figure 4** Asymmetry in the caliber of vessels in the different pulmonary lobes. Mosaic pattern image with areas of high attenuation in LIL containing larger caliber vessels. Note the subsegmental bronchial dilatations in the LSL with low attenuation (arrows). Mosaic pattern image cannot be used for differential diagnosis of PH types. Additional presence of vessels with different caliber and changes in the airway suggest CTEPH.

**Contribution of MDCT to the etiological diagnosis of PH**

In addition to suggest the diagnosis, based on the signs already discussed, MDCT is the most complete tool for the differential diagnosis of PH groups, according to the classification of Dana Point 2008 (table 1).

By carefully assessing the thoracic organs (lung parenchyma, mediastinum and heart chambers), we can not only differentiate between types of PH, but also assess their complications or postsurgical outcomes (thromboendarterectomy and shunt correction).

**Parenchymal lung changes**

**Asymmetry of the vascular diameter**

It is typical of CTEPH and does not appear in other types of PH. The marked regional variation in the size of segmental vessels helps differentiate CTEPH with the rest of causes of PH presenting a more diffuse pattern.

• Vena cava and azygos vein dilatation is considered a marker of left-sided hypertension. In addition to this, Isaacs proposes the assessment of coronary sinus distension that is easily identifiable on almost all CT studies. Dilatation > 11mm is associated with increased PA pressures, confirmed by catheterism.

• Pericardial thickening or effusion. It is more common and earlier visualized in the anterior or aortopulmonary recess, where it appears in the shape of a “bikini bottom”. It may be considered an indirect sign of PH, particularly if associated with increased PA diameter. It is not necessarily associated with pleural effusion. It is also a sign of RV dysfunction and of bad prognosis (figs. 2, 3 and 5).

**Venoocclusive disease and capillary hemangiomatosis**

They are included into group 1’ in the new clinical classification (table 1). Since they were first described, there are doubts on whether they are two separate diseases or manifestations of the same condition. They are clinically indistinguishable from other forms of PH.
idiopathic PH, at least initially. Histologically, there is involvement of septal venules. The diseases have a very poor prognosis and are characterized by rapid clinical deterioration despite vasodilator therapy; for this reason, patients are placed on the waiting list for lung or heart-lung transplantation early. One of the reasons why the new Guides for clinical practice recommend a CT prior to the treatment is to rule out this type of PH. This condition is described as associated with an ever-growing number of diseases: collagenosis, Hashimoto’s thyroiditis, sarcoidosis, bone marrow transplantation, HIV infection, chemo- and radio-therapy and myeloproliferative diseases.

- Subpleural nodules and parenchymal bands suggest infarctions in different stages and are significantly more common in CTEPH\textsuperscript{33,34}.
- Arteriovenous fistulas show a characteristic morphology, with an afferent nutrient and an efferent drainage vessel, and suggest Rendu-Osler-Weber disease and the feasibility of percutaneous embolization as part of its treatment (fig. 5C).

**Mosaic pattern**

Mosaic pattern, seen in all types of PH, is a finding suggestive of small vessel changes or regional variation in perfusion. It was first described in CTEPH, but appears frequently in idiopathic PH and Eisenmenger syndrome. Despite being more common in CTEPH, mosaic pattern is not useful in the differential diagnosis with other types of PH. In CTEPH, the areas of high attenuation contain larger caliber vessels (fig. 4).

**Diffuse parenchymal disease**

High-resolution slices obtained for the study of PH show parenchymal changes secondary to usual interstitial pneumonia (UIP) or collagenosis, (Group 1.4.1) (table 1), which are the second more common cause of PH, after idiopathic PH\textsuperscript{5}. Clinically, they have worse overall prognosis and detecting signs of PH has therapeutic implications if lung transplantation is being considered as part of the treatment. In these cases, PH is not caused by destruction of the vascular bed, but to plexiform lesions. There is no correlation between PA diameter and mean pressure in the

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**Figure 5** Parenchymal nodules. A) Ground-glass millimetric nodules with centrilobular distribution (broken arrows), thickened interlobular septa (black arrow) and small pericardial effusion (thick arrow). B) Thickened interlobular septa, mediastinal adenopathy (broken arrow), right pleural effusion and pericardial effusion at the anterior superior recess (arrow). Figures A and B show the radiological signs of venoocclusive PH. C) Coronal oblique MIP reconstruction shows an arteriovenous fistula in a patient with PH: Rendu-Osler disease.
PA, and the artery/segmental bronchus ratio is difficult to assess due to presence of traction bronchiectasis. Assessment of the PA/Ao ratio is recommended\(^9\,35\), which is easily identified in serial studies of diffuse pulmonary disease, and may become our radiological contribution to PH screening. This assessment is also recommended for patients with COPD, sarcoidosis or histiocytosis X eligible for lung transplantation or in CT follow-up, since the presence of PH in all these cases worsens the prognosis significantly and may be a contraindication to lung transplantation\(^36\).

**Mediastinal changes**

**Adenopathies**

Adenopathies may be seen in CTEPH and in venoocclusive disease but are rare in idiopathic PH. They should be assessed in conjunction with the rest of the findings in order to determine a diagnosis between the former two diseases\(^30\,31\) (fig. 5B).

**Hypertrophy of systemic arteries**

The bronchial arteries arise from the thoracic aorta. Their diameter at origin may reach up to 1.4 mm and they form anastomoses with the pulmonary arterial tree through multiple microvascular connections. These anastomoses occur not only through the major bronchi, but also beyond the pulmonary lobe, on the alveolar walls. Bronchial circulation responds to the reduced pulmonary flow with an increase, hypertrophy and proliferation of vessels along this tangle of anastomotic channels\(^10\,37\). Hypertrophy may be seen in idiopathic PH, although this finding is more common in CTEPH. Enlargement may be found in other systemic arteries in addition to the bronchial systemic arteries, including the pulmonary ligament, pleural, intercostal, subclavian branches, axillary and phrenic arteries, and this finding is almost exclusive of CTEPH\(^37\) (fig. 6).

**Vascular changes**

**Changes in the wall and caliber of vessels**

- Idiopathic PH involves the entire arterial tree, increasing the central arterial caliber and causing a progressive and fast change in the peripheral arterial caliber that may show a corkscrew configuration (fig. 2A and B). The number of arterial branches is reduced, but the arterial walls are smooth (fig. 7).
- In CTEPH, the arterial wall morphology is completely diverse. Chronic thrombi seen in this condition adhere to the vessel wall resulting in a scalloped contour when observed on its longitudinal axis, or forming an obtuse angle with the arterial walls when observed on its transversal axis. Incomplete recanalization of thrombi results in images of intraarterial webs and, distal to them, post-stenotic dilatations. Thrombi that do not reabsorb or recanalize result in a complete stenosis of the branches with a cul-de-sac image. Eventually, the arterial tree shows arteries with multiple calibre changes, typical of this type of PH. CTEPH is generally bilateral affecting both the main and lobar and segmental branches. Diagnosis of CTEPH allow us to assess the feasibility of thromboendarterectomy in these patients\(^33\,34\,38\,40\) (fig. 8A and B).

In case of arterial obstruction, the main role of CT is to rule out other causes of scintigraphic abnormalities similar to CTEPH, including venoocclusive disease, PA sarcoma, large-vessel vasculitis, extrinsic compression by a mediastinal carcinoma, lymphadenopathy, mediastinal fibrosis or pulmonary venous thrombosis\(^38\) (fig. 9A-C).

**Complications of PH**

As with systemic pulmonary arterial hypertension, the complications of PH are arterial wall calcification, thrombus formation, aneurysms and dissection.

- In long-standing PH, early atherosclerosis of the muscular and elastic central arteries is not an unusual finding\(^8\) (figs. 6 and 11).
- An uncommon complication of chronic PH are aneurysms in the main branches of the PA (diameter > 4 cm)\(^9\). Most of
these aneurysms occur in the presence of other risk co-factors, such as congenital heart disease or vasculitis, and patients presenting PH only are rare. Central aneurysms (trunk and two main branches) may cause clinical symptoms due to bronchial compression (fig. 10A), angina associated with compression of the left coronary artery by the PA trunk, PA dissection and/or rupture or intraarterial thrombus; aneurysms in the peripheral or intrapulmonary branches are less common. The number of cases with central or peripheral aneurysms is increasing due to the higher survival rates of patients with PH. The fact that these patients have reservoir catheters placed in has to be taken into account, since they may cause mycotic aneurysms, which clinically manifest as hemoptysis.

- In situ thrombi, in the course of PH, may appear in the trunk and branches of the main artery. These thrombi are not hemodynamically significant and do not show any signs on perfusion scintigraphy. In case of a thrombus, it is important to make the differential diagnosis with CTEPH, based on the absence of signs of chronic lobar or segmental thrombi, lack of parenchymal signs such as subpleural nodules, bands or mosaic, adenopathies or bronchial circulation. Up until now, scintigraphy was key to the differential diagnosis, but now, the vascular study using CT can show the arterial morphology. Acute or chronic thrombi may be found in 50% of cases of idiopathic PH (fig. 10B) and the role of thrombosis in both idiopathic and associated PH remains controversial.

Figure 8  Vessel morphology in CTEPH. A) Mural thrombus with a scalloped outline from a longitudinal view (right PA) and a crescent image with obtuse angles from an axial view in the branch of the LIL (Reproduced with permission Ref. 34). B) Coronal MIP reconstruction shows thrombi recanalization with intraarterial webs (dotted arrows), post-stenotic dilatations (asterisk), cul-de-sac vascular obstructions (thick arrows) and longitudinal stenosis (arrowhead).

Figure 9  Usefulness of MDCT in the differential diagnosis of arterial obstruction. A) PA sarcoma with calcified parenchymal metastases and pleural effusion. The tumor extends beyond the vessel limits into the mediastinum showing heterogeneous enhancement of contrast agent and calcified areas. B) Fibrosing mediastinitis. Fibrosis involves the right pulmonary artery, superior vena cava, mediastinal and anterior pleura and the middle lobe bronchus. C) Takayasu’s arteritis. Complete obstruction of the arteries of both inferior lobes and post-stenotic dilatations in the superior lobes. Patent endograft in the branch of the RSL (Reproduced with permission Ref. 34).
According to Auger and Fedullo, central thrombi may be found in conditions other than CPTE including idiopathic PH, Eisenmenger syndrome and COPD. It is unknown if the presence of microthrombi in the pulmonary arterioles is the cause or the consequence of PH, but there is no doubt that their presence contributes to the progression of the disease.

• PA dissection is a rare and fatal condition. In addition to PH, other rare causes of dissection are chronic arterial inflammation, right-heart endocarditis, amyloidosis, trauma and severe atherosclerosis. Dissection seems more frequent in PH associated with congenital heart diseases than in idiopathic PH. CT allows determination of the localization, size and extent of the aneurysm and of the dissection (fig. 10C). Dissection is very seldom diagnosed in living patients and it is usually found at autopsy studies. The affected site is generally from the pulmonary valve onward or the two main branches, and only one publication has described dissection at a segmental level.

Cardiac changes

Although patients are referred to us for a CT exam after echocardiography, MDCT is an adequate tool for the morphological study of the heart chambers and interatrial and interventricular septa. It is very important to include in all our reports a systematic assessment of the size and morphology of the heart chambers, septa, wall thickness and the arterial and venous anastomoses.

Regarding the left chambers, enlarged atrium and pulmonary veins or pathological LV wall may be indicative of PH associated with left-heart disease; in some centers this is the most common cause of PH.

Despite a negative echocardiography, the study of the site of drainage of the pulmonary veins is essential in order to rule out partial anomalous venous drainages, either isolated or in conjunction with interatrial septum defects (fig. 11A). It has to be taken into account that a sinus venosus atrial septal defect and partial anomalous venous drainage might be missed on transthoracic echocardiography.

A PH report should include the thoracic aorta, since the presence of ductus is not an uncommon finding that can be easily diagnosed but that may go unnoticed in transthoracic echocardiography (fig. 11B).

In case of complex heart diseases, CT shows the heart anatomy and also enables joint assessment of other findings (pulmonary, pleural or mediastinal) that may be relevant for the surgical planning (fig. 11C).

Conflict of interest

The author declares no conflict of interest.
I would particularly like to express my gratitude to Dr. P. Escribano Subías and Dr. C. Jiménez López-Guarch from the Pulmonary Hypertension Unit of the Cardiology Department of Hospital Universitario Doce de Octubre for their collaboration in the assessment of the cases and their constant effort to set up and maintain a multidisciplinary Unit, where each and every specialist feels encouraged.

References

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