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# Multimodality imaging for PAH: Is CT better than MRI?

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GIORNATE CARDIOLOGICHE TORINESI

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#### EDITOR'S CHOICE GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT)

Nazzareno Galiè 🗠 , Marc Humbert 🗠 , Jean-Luc Vachiery, Simon Gibbs, Irene Lang, Adam Torbicki, Gérald Simonneau, Andrew Peacock, Anton Vonk Noordegraaf, Maurice Beghetti, ... Show more

European Heart Journal, Volume 37, Issue 1, 1 January 2016, Pages 67–119, https://doi.org/10.1093/eurheartj/ehv317 Published: 29 August 2015



O/D \* congenital heart diseases, CT \* computed somography, CTD \* connective tissue disease, CTEPH \* chronic thromboeholic pulmonary hypertension; DLCD \* carbon monoxide diffusing capacity; BCG \* electrocardiogram; HV \* Human immunodeficiency virue; HR-CT \* high-resolution CT; mRP \* mean pulmonary anterial pressure; RR \* pulmonary anglography; RH \* pulmonary anterial hypertension; RRVP \* pulmonary anterial pressure; RT \* pulmonary function tests; RH \* pulmonary hypertension; PVODIPCH \* pulmonary veno-occlusies disease or pulmonary capillary hemangiomathosis; PVR \* pulmonary vascular resistance; RHC \* right heart carbeartistics; RY + right exerticular; VIQ \* ventilation/perfusion;

CT pulmonary angiography alone may miss diagnosis of chronic thromboembolic pulmonary hypertension.

EDITOR'S CHOICE OUTDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT) ●

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# 5.1.7 High-resolution computed tomography, contrast-enhanced computed tomography, and pulmonary angiography

CT imaging is a widely available tool that can provide important information on vascular, cardiac, parenchymal and mediastinal abnormalities. It may suggest the diagnosis of PH (PA or RV enlargement), identify a cause of PH such as CTEPH or lung disease, provide clues as to the form of PAH (e.g. oesophageal dilation in SSc or congenital cardiac defects such as anomalous pulmonary venous drainage) and also provide prognostic information.<sup>50</sup>

#### 5.1.8 Cardiac magnetic resonance imaging

CMR imaging is accurate and reproducible in the assessment of RV size, morphology and function and allows non-invasive assessment of blood flow, including stroke volume, CO, pulmonary arterial distensibility and RV mass.

In patients with suspected PH, the presence of late gadolinium enhancement, reduced pulmonary arterial distensibility and retrograde flow have high predictive value for the identification of PH; however, no single CMR measurement can exclude PH.<sup>60-62</sup> In patients with PH, CMR may also be useful in cases of suspected CHD if echocardiography is not conclusive. EDITOR'S CHOICE GUIDELINES

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#### 12.1 Facilities and skills required for a expert referral centre

 Expert referral centres are recommended to provide care by an interprofessional team that should, as a minimum, comprise:<sup>451-456</sup>

(a) two consultant physicians (normally from either or both cardiology and respiratory medicine) experienced in and with a special interest in PH with dedicated PH clinical sessions for outpatients, inpatients and a multidisciplinary team meeting

(b) clinical nurse specialist

- (c) radiologist with expertise in pulmonary hypertension imaging
- (d) cardiologist or PH physician with expertise in echocardiography

 (e) cardiologist or PH physician with expertise in RHC and vasoreactivity testing

- (f) access to psychological and social work support
- (g) appropriate on-call cover and expertise

For expert referral centres, access to the following facilities is recommended:

- (a) a ward where staff has special expertise in PH
- (b) an intensive therapy unit with relevant expertise
- (c) a specialist outpatient service
- (d) emergency care

(e) diagnostic investigations including echocardiography, CT scanning, nuclear scanning, MR imaging, ultrasound, exercise testing, lung function testing and a cardiac catheterization laboratory

(f) access to the full range of specific PAH and CTEPH drug therapy available in their country

#### **CHEST IMAGING**

#### Pulmonary Hypertension: How the Radiologist Can Help<sup>1</sup>

| Group | Description   |  |  |
|-------|---|--|--|
| 1     | Pulmonary arterial hypertension   |  |  |
| 1.1   | Idiopathic pulmonary arterial hypertension  |  |  |
| 1.2   | Heritable   |  |  |
| 1.3   | Drug and toxin induced  |  |  |
| 1.4   | Associated with connective tissue diseases, HIV infection, portal hypertension, con-<br>genital heart disease, schistosomiasis, and chronic hemolytic anemia  |  |  |
| 1.5   | Persistent pulmonary hypertension in newborns   |  |  |
| 1'    | PVOD or PCH   |  |  |
| 2     | Pulmonary hypertension due to left-sided heart disease  |  |  |
| 3     | Pulmonary hypertension due to lung diseases or hypoxia  |  |  |
| 4     | Chronic thromboembolic pulmonary hypertension   |  |  |
| 5     | Pulmonary hypertension with unclear multifactorial mechanisms such as hematologic<br>disorders (myeloproliferative disorders, splenectomy), systemic disorders (sarcoid-<br>osis, pulmonary Langerhans cell histiocytosis, lymphangioleiomyomatosis, neuro-<br>fibromatosis, vasculitis), metabolic disorders (glycogen storage disease, Gaucher<br>disease, thyroid disorders), and other disorders (tumoral obstruction, fibrosing<br>mediastinitis, chronic renal failure in patients undergoing dialysis) |  |  |

1.Suspicion Mon specific symptoms

Dyspnea, fatigue, syncope & chest pain 2.Detection

Clinical Examination Accentuated pulmonic component of second heart sound (90%)

Right ventricular S3 or S4 Tricuspid regurgitant murmur

ECG Right axis deviation, S1Q3 pattern Lacks sensitivity & specificity as screening tool

Chest X Ray Dilated main & hilar pulmonary artery Filling in of retrosternal airspace due to RV dilatation

9

Transthoracic Echocardiography Screening test of choice to detect PH

#### 3.Classification

- To define the etiology of PH according to
- Dana Point Clinical Classification and rule out surgically treatable causes of PH
- 4.Evaluation
- Confirm the diagnosis of PH
- Hemodynamic evaluation
- Assessment of functional capacity at baseline

#### Symptoms/ Screening/ Incidental Findings

Physical Exam ECG Chest X Ray Transthoracic/Transesophageal Echocardiogram



#### **CHEST IMAGING**

#### Pulmonary Hypertension: How the Radiologist Can Help<sup>1</sup>



## Is CT better than MRI?



## CTA



A pulmonary artery with a diameter of 29 mm or more has a positive predictive value of 97%, sensitivity of 87%, and specificity of 89% for the presence of pulmonary hypertension

In the presence of a dilated (29 mm or more) main pulmonary artery, a segmental artery-tobronchus diameter ratio of 1:1 or more in three or four lobes has a specificity of 100% for the presence of pulmonary hypertension

At CTPA, a main pulmonary arterial diameter larger than that of the ascending aorta is also a sign of pulmonary hypertension, with a positive predictive value of 96% and specificity of 92%, especially in patients younger than 50 years old

## MRA



#### **MRA**



## MRA



#### Is CT better than MRI?



#### CT team 1-0 MR team







![](_page_12_Picture_3.jpeg)

Straightening or leftward bowing of the interventricular septum; right ventricular dilatation

![](_page_12_Picture_5.jpeg)

Decreased right ventricular ejection fraction; dilatation of the inferior vena cava and hepatic veins; and pericardial effusion

![](_page_13_Picture_1.jpeg)

![](_page_13_Picture_2.jpeg)

![](_page_13_Figure_3.jpeg)

![](_page_13_Picture_4.jpeg)

| Ventricular Volumes*  | Pathologic Characteristics   | Clinical Significance  |
|---|--|--|
| Increased right ventricular end<br>diastolic and end systolic volume,<br>decreased right ventricular stroke<br>volume and right ventricular<br>cardiac output, decreased right<br>ventricular ejection fraction | Right ventricular dilatation   | Increased right ventricular volumes<br>and decreased right and left<br>ventricular stroke volumes from<br>baseline indicate mortality and<br>treatment failure |
| Decreased left ventricular end<br>diastolic volume and end systolic<br>volume, decreased left ventricular<br>peak filling rate (left ventricular<br>end diastolic volume per sec)                               | Decreased left ventricular volumes<br>due to increased pulmonary<br>vascular resistance, limiting right<br>ventricular stroke volume and<br>available blood volume for left<br>ventricular filling | Increased right ventricular volumes<br>and decreased right and left<br>ventricular stroke volumes from<br>baseline indicate mortality and<br>treatment failure |
| Increased ventricular mass index <sup>†</sup>   | Right ventricular hypertrophy  | Ventricular mass index >0.6 corre-<br>lates with detection of pulmonary<br>hypertension at catheterization of<br>the right side of the heart                   |

19 yo man Bi-ventricular failure in repaired TOF (surgery in 1996 with closure of ASD and minimal residual shunt)

![](_page_14_Picture_2.jpeg)

![](_page_15_Picture_1.jpeg)

#### 2D PHASE CONTRAST SEQUENCES

![](_page_15_Figure_3.jpeg)

## Is CT better than MRI?

![](_page_16_Picture_1.jpeg)

#### CT team 1-1 MR team

![](_page_17_Picture_0.jpeg)

![](_page_17_Picture_1.jpeg)

26 yo woman with dyspnea at exertion, shows widespread, illdefined, centrilobular areas of attenuation: a finding indicative of pulmonary capillary hemangiomatosis. Centrilobular ground-glass nodules are a feature of pulmonary hypertension and are especially common in patients with idiopathic pulmonary arterial hypertension

![](_page_18_Picture_0.jpeg)

![](_page_18_Picture_1.jpeg)

## Is CT better than MRI?

![](_page_19_Picture_1.jpeg)

#### CT team 2-1 MR team

# **Cardiac MR – Late gadolinium enhancement**

![](_page_20_Picture_1.jpeg)

#### HYPERENHANCEMENT PATTERNS

Ischemic

A. Subendocardial Infarct

![](_page_20_Picture_5.jpeg)

**B.** Transmural Infarct

#### Nonischemic

![](_page_20_Picture_7.jpeg)

![](_page_20_Picture_8.jpeg)

 Idiopathic Dilated 
Hypertrophic Cardiomyopathy Cardiomyopathy

> · Right ventricular pressure overload (e.g.

- Sarcoidosis · Myocarditis Anderson-Fabry
- congenital heart disease, . Chagas Disease pulmonary HTN)
- **B.** Epicardial HE

Myocarditis

![](_page_20_Picture_14.jpeg)

· Sarcoidosis, Myocarditis, Anderson-Fabry, Chagas Disease

C. Global Endocardial HE

![](_page_20_Picture_17.jpeg)

· Amyloidosis, Systemic Sclerosis, Post cardiac transplantation

# Cardiac MR – Late gadolinium enhancement

![](_page_21_Picture_1.jpeg)

## Is CT better than MRI?

![](_page_22_Picture_1.jpeg)

#### CT team 2-2 MR team

## CT

60 yo woman From 1986 Wegener Granulomatosis Therapy: corticosteroids and cyclofosfamide CT: Pulmonary signs of primary disease From 2006 rise of pulmonary artery inflammatory involvement

![](_page_23_Picture_2.jpeg)

#### Catheterism: Forward pulmonary garadient and signs of PAH

![](_page_24_Picture_1.jpeg)

Courtesy of Dott. Fulvio Orzan

![](_page_25_Picture_1.jpeg)

![](_page_26_Picture_0.jpeg)

![](_page_26_Figure_1.jpeg)

RR-interval: 698 ms (from heart rate)

![](_page_27_Picture_0.jpeg)

![](_page_27_Figure_1.jpeg)

![](_page_28_Picture_0.jpeg)

# Take home points

![](_page_28_Picture_2.jpeg)

V vessels diameters, lung evaluation, widely available

X cardiac function, flow, radiation exposure

#### MR

V cardiac function, flow and myocardial fibrosis

X vessels diameters, long time examination, poorly available

#### **COMPLEMENTARY IMAGING TECHNIQUES**

**IN EVALUATION OF PHA**