Spleen Size in Idiopathic and Heritable Pulmonary Arterial Hypertension

Adriano R. Tonelli a Ruchi Yadav b Amit Gupta b Andrea V. Arrossi c
Gustavo A. Heresi a Raed A. Dweik a

a Department of Pulmonary, Allergy and Critical Care Medicine, Respiratory Institute, b Diagnostic Radiology and c Department of Pathology, Cleveland Clinic, Cleveland, Ohio, USA

Abstract

Background: It is unknown whether the spleen size correlates with disease severity and outcome in patients with idiopathic and heritable pulmonary arterial hypertension (PAH). Objectives: To determine the prevalence of splenomegaly in PAH and assess whether it correlates with severity of disease and outcome. Methods: We identified subjects with either heritable or idiopathic PAH who had Doppler echocardiography, right-heart catheterization and computed tomography (CT) of the chest and/or abdomen that included the spleen. Results: We included 62 subjects with a mean age (±SD) of 49 (±15) years; 82% were women. Spleen dimensions were 10 (±3), 6 (±2) and 9 (±2) cm for the craniocaudal length, thickness and width measurements, respectively. The median [interquartile range (IQR)] spleen volume was 344 (225–533) cm³. Splenomegaly was observed in 52–63% of the patients, depending on the formula used. The spleen volume was not associated with clinical, echocardiographic or hemodynamic variables. Spleen volume was not associated with adjusted mortality. We studied the characteristics of the spleen during autopsy in 9 patients with idiopathic PAH who died of right-heart failure. The mean (IQR) spleen weight was 220 (151–325) g. We observed early congestion in all but 2 patients who had chronic congestion. Conclusions: Splenomegaly of predominantly mild degree is common in idiopathic and heritable PAH. However, spleen size was not associated with clinical, echocardiographic, hemodynamic and survival data in these patients.

Key Words
Outcome assessment • Pulmonary arterial hypertension • Spleen • Splenomegaly

Introduction

Pulmonary arterial hypertension (PAH) is a disease characterized by restrictive flow through the pulmonary circulation that can lead to right-heart failure and death [1]. It involves a variety of diseases included in group 1 of the 4th World Symposium on Pulmonary Hypertension updated in 2008 [2]. PAH is hemodynamically characterized by a mean pulmonary artery pressure ≥25 mm Hg with pulmonary vascular resistance (PVR) ≥3 Wood units and pulmonary arterial occlusion pressure ≤15 mm Hg [1]. When the right ventricular (RV) compensatory response to the increased PVR is overwhelmed, right heart failure ensues. Clinical manifestations of right
heart failure include symptoms and signs resulting from venous congestion and low cardiac output [3]. Characteristic physical signs related to venous congestion include hepatomegaly, ascites and peripheral edema [4, 5]. Splenomegaly has been associated with right heart failure [6], but it is usually not described as part of the signs observed in patients with advanced PAH [7, 8].

It is unknown whether patients with idiopathic or heritable PAH have abnormal spleen volume and whether its volume correlates with clinical, echocardiographic and hemodynamic parameters or outcomes. We hypothesized that patients with idiopathic and heritable PAH have splenomegaly which may correlate with severity of disease and outcomes. We evaluated whether assessing the spleen volume in the chest CT could provide valuable prognostic information in these patients.

Methods

Study Design and Inclusion Criteria

This study was approved by the Cleveland Clinic Institutional Review Board (protocol number: 10-1127). Informed consent was waived. We identified subjects using the Cleveland Clinic Pulmonary Hypertension Registry. We selected patients with pulmonary arterial occlusion pressure $\leq 15$ mm Hg, PVR $\geq 3$ Wood units, forced expiratory volume in 1 s/forced vital capacity (FVC) $\geq 0.6$, and total lung capacity $\geq 60\%$. Of those, we only included patients with either idiopathic or heritable PAH ($n = 140$) [2] who had a CT of the chest (including the spleen) or abdomen between 1994 and 2010.

Each patient underwent a careful selection process to exclude other etiologies of pulmonary hypertension. We excluded all patients in whom at least two pulmonary hypertension physicians were not in complete agreement about the diagnosis of idiopathic or heritable PAH. Except for 1 patient who had splenectomy and was excluded, no other patient had any medical condition linked to changes in the spleen volume.

Measurements and Calculations

We selected the CT scans of the chest and abdomen that incorporated the spleen in its entirety and were performed closest in time to any right-heart catheterization (RHC) performed at our institution. In the case of several studies, we selected the first one. Studies were obtained with commercially available single- and four-slice CT scanners using a slice thickness of 5 mm at 4-mm intervals before 2003. Since 2004, studies were obtained initially with 16- and subsequently with 64-detector CT scanners using 5-, 3- and 1-mm section thicknesses at 2.5-, 1.5- and 1-mm intervals, respectively. Two radiologists (R.Y. and A.G.) reviewed all the CT scans performed in these patients.

We measured the splenic length, width and thickness in all patients. The length was obtained by multiplying the number of sections where the spleen was visualized by the thickness of the
tionships were assessed by fitting a LOWESS (locally weighted splenic volume with other continuous variables. Nonlinear rela-

calculate the volume of a prolate ellipsoid \(0.524 \times \text{splenic length}\times \text{splenic width} \times \text{splenic thickness}\) \(10\). The splenic index is the multiplication of the 3 splenic measurements without the in-
corporation of a correction factor.

Splenomegaly was considered if the calculated splenic volume exceeded the 95% limit for spleen size by height and weight, as
described by Sprogøe-Jakobsen and Sprogøe-Jakobsen \[11\]. Similarly, we defined hyposplenia when the calculated splenic volume was less than the 5% limit for spleen size by height and weight \[11\]. Normal spleen size was considered present when the splenic vol-

tume was within the 5–95% limit interval.

RHC data were obtained from studies done as part of clinical evaluation. The procedures were performed in the standard man-
er in the cardiac catheterization laboratory using a 7-french pul-

monary artery catheter. Pressure measurements were obtained from paper recordings at end-expiration. Cardiac output was ob-
tained by the thermodilution method and transpulmonary gradient (mean pulmonary artery-occlusion pressure) and PVR (trans-
pulmonary gradient/cardiac output) were calculated.

In addition, we reviewed the transthoracic Doppler echocar-
diogram performed the closest to the CT-RHC pair included in
the analysis. Experienced operators, blinded to the results of the CT of the chest/abdomen, reviewed the echocardiograms and as-

essed RV size and function subjectively. A 6-min walk test was performed following American Thoracic Society guidelines \[12\]. Predicted 6-min walk distance was estimated using formulae sug-
gested by Enright and Sherrill \[13\]. Plasma brain natriuretic pep-
tide was measured at the time of RHC in 40 patients.

In order to support our findings, we also reviewed the reports
of all autopsies performed at the Cleveland Clinic from 1990 to
2010 that included the phrases ‘primary pulmonary arterial hy-
pertension’, ‘idiopathic pulmonary arterial hypertension’ and ‘plexiform lesions’ in any form within the text. Only the patients with idiopathic or heritable PAH who died of right heart failure were included to ensure the absence of any other condition that could result in alteration of the spleen weight. We recorded the age at the time of death, gender, height, weight, heart weight, right and left ventricular heart thickness, presence and amount of peri-
cardial/pleural effusions and ascites, and the liver and spleen

weights.

Statistics
Means and standard deviations are provided for all continu-
ous variables when normally distributed, and medians [inter-
quartile ranges (IQR)] when not. Two-group comparisons were
performed by Welch’s t-test or the Mann-Whitney U test when appropriate. Categorical data were compared using Fischer’s exact
test. We used Pearson’s correlation and linear regression analysis
(adjusted for age, gender, height, and weight) \[14\] to compare
splenic volume with other continuous variables. Nonlinear relations-
ships were assessed by fitting a LOWESS (locally weighted scatterplot smoothing) line. Survival was assessed by Kaplan-
Meier methodology. Time 0 was the date at the time of the CT of
the chest/abdomen, and the end of follow-up was December, 2010, or the time of the recipient’s death (or transplantation, when a combined endpoint was used). Survival analysis was also performed with Cox proportional hazards modeling, using spleen volume and other factors as covariates, adjusted by age and gen-
der, height, and weight. Hazard ratios (HR) and the correspond-
ing 95% confidence intervals (CI) are shown. At the Cleveland
Clinic, patients are transplanted using criteria established by the
International Society for Heart and Lung Transplantation \[15\].

Death of the study participants was ascertained by reviewing our records and querying the US Social Security Death Index. All p
values reported are two tailed. A value of \(p < 0.05\) was considered
significant. Statistical analyses were performed using the statistical
package IBM SPSS, version 20 (IBM, Armonk, New York, N.Y.,
USA) and R statistical software, version 2.13.0 (www.r-project.
org).

Results
Patient Characteristics
We included 62 unique subjects (82% women) with a mean age (±SD) of 49 (±15) years. Fifty-four had idiopathic and 8 had heritable PAH. Race was Caucasian in 81%, African American in 15% and other in 4%. New York Heart Association (NYHA) functional class was II in 36%, III in 48% and IV in 16% of the patients. Mean (SD) height was 1.62 (±8.5) m, weight 77 (±23) kg and

mean body mass index was 29 (±8). Six patients (10%) were receiving PAH-targeted therapy at the time of the chest/abdomen CT, 3 patients sildenafil, and 3 bosentan. Brain natriuretic peptide was 424 (±592) pg/ml and the 6-min walk distance was 327 (±104) m or 58.5% (±17) of predicted \[13\]. Hemodynamic and echocardiographic
characteristics are shown in tables 1 and 2, respecti-

vally.

Indications for CT
CT of the chest/abdomen was performed with a me-
dian (IQR) difference from the RHC of 0 (0–6) months. A total of 53 (86%) CT scans were of the chest and the rest of the abdomen (9 patients, 14%). CT were done for fur-

ther evaluation of lung parenchyma (47%), dyspnea (19%),
abdominal distention (9%), evaluation before lung trans-
plantation (7%), lung nodule assessment (7%), and for other reasons (11%) such as research, chest pain, and fol-

low-up of ovarian carcinoma.

CT Measurements
The aorta and pulmonary artery diameters were 3 (±0.5) and 3.4 (±0.5) cm, respectively; with a ratio of pulmonary artery/aorta of 1.2 (±0.2). Spleen dimensions were 10 (±3), 6 (±2), and 9 (±2) cm for the craniocaudal,
thickness and width measurements, respectively. The median (IQR) splenic index was 541 (337–868) cm$^3$. Sixty percent of the patients had a splenic index above the upper limit of the normal range (480 cm$^3$)\[16, 17\].

The splenic volumes measured by Prassopoulos et al.\[9\] and prolate ellipsoid formulae were 344 (225–533) and 283 (176–455) cm$^3$, respectively (fig. 2).

The splenic volumes measured by the formula of Prassopoulos et al.\[9\] were 339 (222–291) and 378 (279–553) cm$^3$ in patients that underwent CT of the chest and abdomen, respectively (p = 0.39). The spleen size was directly associated with weight ($R = 0.43$, p = 0.001) but not with age and height. The splenic size was higher in males [461 (339–673) cm$^3$] than in females [333 (218–491) cm$^3$; p = 0.021, Mann-Whitney test]. In multivariate analysis that included weight and gender, only weight was significantly associated with spleen size (p = 0.027).

Using the table for spleen volume suggested by Sprogøe-Jakobsen and Sprogøe-Jakobsen\[11\], splenomegaly was observed in 63% of the patients using the formula to calculate volume of Prassopoulos et al.\[9\] and 51.6% of the individuals by applying the formula to calculate the volume of a prolate ellipsoid\[10\]. Only 1 patient (1.6%) had hyposplenia (<5% limit for spleen size CI) by

### Table 1. Hemodynamic parameters in all the patients, with or without splenomegaly (means ± SD)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>All patients (n = 62)</th>
<th>No splenomegaly (n = 22)</th>
<th>Splenomegaly (n = 39)</th>
<th>p value (Welch’s t test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RAP, mm Hg</td>
<td>12 ± 7</td>
<td>10 ± 5</td>
<td>13 ± 7</td>
<td>0.08</td>
</tr>
<tr>
<td>RV systolic pressure, mm Hg</td>
<td>83 ± 22</td>
<td>85 ± 20</td>
<td>82 ± 23</td>
<td>0.6</td>
</tr>
<tr>
<td>RV diastolic pressure, mm Hg</td>
<td>14 ± 10</td>
<td>13 ± 7</td>
<td>15 ± 11</td>
<td>0.39</td>
</tr>
<tr>
<td>PA systolic pressure, mm Hg</td>
<td>86 ± 21</td>
<td>87 ± 21</td>
<td>85 ± 22</td>
<td>0.75</td>
</tr>
<tr>
<td>PA diastolic pressure, mm Hg</td>
<td>37 ± 11</td>
<td>39 ± 12</td>
<td>37 ± 11</td>
<td>0.61</td>
</tr>
<tr>
<td>PA mean pressure, mm Hg</td>
<td>54 ± 14</td>
<td>55 ± 14</td>
<td>53 ± 14</td>
<td>0.66</td>
</tr>
<tr>
<td>PAOP, mm Hg</td>
<td>10 ± 4</td>
<td>10 ± 4</td>
<td>10 ± 4</td>
<td>0.96</td>
</tr>
<tr>
<td>Cardiac output, l/min</td>
<td>4.1 ± 1.5</td>
<td>3.8 ± 1.4</td>
<td>4.2 ± 1.5</td>
<td>0.28</td>
</tr>
<tr>
<td>Cardiac index, l/min/m$^2$</td>
<td>2.2 ± 0.8</td>
<td>2 ± 0.6</td>
<td>2.3 ± 0.8</td>
<td>0.13</td>
</tr>
<tr>
<td>PVR, Wood units</td>
<td>12 ± 7</td>
<td>13 ± 7</td>
<td>12 ± 7</td>
<td>0.85</td>
</tr>
<tr>
<td>TPG, mm Hg</td>
<td>43 ± 14</td>
<td>44 ± 14</td>
<td>43 ± 14</td>
<td>0.8</td>
</tr>
</tbody>
</table>

PA = Pulmonary artery; PAOP = PA occlusion pressure; RAP = right atrial pressure; TPG = transpulmonary gradient. Splenomegaly was defined by a volume (using the Prassopoulos et al.\[9\] formula) higher than the 95% limit according to the expected spleen volume by height and weight described by Sprogøe-Jakobsen and Sprogøe-Jakobsen\[11\].

### Table 2. Echocardiographic parameters in all patients, with or without splenomegaly

<table>
<thead>
<tr>
<th>Parameter</th>
<th>All patients (n = 62), n (%)</th>
<th>No splenomegaly (n = 22), n (%)</th>
<th>Splenomegaly (n = 39), n (%)</th>
<th>p value (Fisher’s exact test)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RV dilation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>7 (11)</td>
<td>1 (4)</td>
<td>6 (15)</td>
<td>0.59</td>
</tr>
<tr>
<td>Mild</td>
<td>4 (6)</td>
<td>2 (9)</td>
<td>2 (5)</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>24 (39)</td>
<td>10 (46)</td>
<td>14 (36)</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>27 (44)</td>
<td>9 (41)</td>
<td>17 (44)</td>
<td></td>
</tr>
<tr>
<td>RV dysfunction</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent</td>
<td>8 (13)</td>
<td>1 (4)</td>
<td>7 (18)</td>
<td>0.08</td>
</tr>
<tr>
<td>Mild</td>
<td>3 (5)</td>
<td>3 (14)</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>29 (47)</td>
<td>11 (50)</td>
<td>18 (46)</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>22 (35)</td>
<td>7 (32)</td>
<td>14 (36)</td>
<td></td>
</tr>
</tbody>
</table>

1 One patient with hyposplenia was removed from the subgroup analysis.
the formula described by Prassopoulos et al. [9] and 2 (3.2%) by the formula used to determine the volume of a prolate ellipsoid [10] (table 3).

When using a spleen volume to define splenomegaly of 314.5 cm³, the formula by Prasspoulous et al. [9] revealed that 36 patients (58%) had splenomegaly. Using the upper 95% limit (334 cm³) from the normal spleen volume proposed by Henderson et al. [18], we observed that 34 (55%) patients had splenomegaly. If only craniocaudal splenic length is used, splenomegaly (craniocaudal length ≥10 cm) [19] was observed in 22 patients (36.1%).

### Correlation of CT Findings

We did not observe a significant association between spleen volume obtained by the formula described by Prassopoulos et al. [9] and either pulmonary artery diameter (R = 0.24, p = 0.09) or pulmonary artery/aortic ratio (p = 0.97). Similarly, no significant associations were observed between spleen volume and NYHA functional class, laboratory data (brain natriuretic peptide serum levels, hemoglobin, white blood cells, platelets, sodium, creatinine, blood urea nitrogen, albumin, bilirubin, alkaline phosphatase, and alanine aminotransferase), pulse oximeter oxygen saturation (SpO₂) at rest on room air, 6-min walk distance (in meters or percent of predicted), echocardiographic (left ventricular ejection fraction, tricuspid jet velocity, or degree of RV dilation or dysfunction) or hemodynamic parameters (fig. 3). Lack of association was also observed for all these variables when spleen volume was adjusted for age, height, weight, and gender. Aspartate aminotransferase was significantly associated with splenic volume (R = –0.33, p = 0.026), although this association disappeared when adjusting for other factors (p = 0.09).

### Survival Analysis

Survival at 1, 2, and 3 years was 89.7, 73.1 and 64.3%, respectively. When survival was adjusted for age, gender, height, weight, we did not find a significant effect of spleen volume as a continuous variable (HR: 1; 95% CI: 0.997–1.002; p = 0.74; fig. 4). Similarly, no survival difference was noted when the binary variable splenomegaly (defined by the Sprogøe-Jakobsen and Sprogøe-Jakobsen

---

**Table 3. Spleen size**

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Splenomegaly</td>
<td>39 (63)</td>
<td>32 (52)</td>
</tr>
<tr>
<td>Normal spleen size</td>
<td>22 (35)</td>
<td>28 (45)</td>
</tr>
<tr>
<td>Hyposplenia</td>
<td>1 (2)</td>
<td>2 (3)</td>
</tr>
</tbody>
</table>

1 See Materials and Methods for the definitions of these terms.
[11] table) was included in the model, adjusted for age, gender, weight, and height (HR: 0.91; 95% CI: 0.36–2.3; p = 0.84).

Autopsy Findings from Patients with Idiopathic PAH

We identified the autopsies of 9 patients with idiopathic PAH patients who died of right heart failure. None of these 9 patients had a CT of the chest or abdomen close to the RHC to be included in the main analysis. The mean (±SD) age of these patients was 44 (±20) years and 67% of them were females. Mean (±SD) height and weight were 161 (±9) cm and 69 (±11) kg, respectively. On the autopsies, the heart, and left and right lung weights were 490 (IQR: 360–605), 650 (IQR: 400–770) and 570 (IQR: 338–725) g, respectively. Pericardial and pleural effusions of at least mild degree were noted in 3 and 6 cases, respectively, and ascites in 6 patients. The liver weight was 1,650 (IQR: 1,475–1,950) g and the spleen weight was 220 (IQR: 151–325) g. Marked RV hypertrophy (0.8 cm, IQR: 0.6–1.1) and dilation was noted in all cases, as well as plexogenic pulmonary hypertensive arteriopathy on microscopic evaluation and some degree of hepatic congestion. Splenic congestion was seen in all cases, and 2 spleens revealed fibrosis of the capsule (fig. 5).

Fig. 3. Scatter plots with linear regression and LOWESS line. All panels have boxplots corresponding to the variable shown in the x and y axis. a–c Scatter plots of right atrial pressure (RAP), mean pulmonary artery pressure (PAP) and PVR versus splenic volume. All scatter plots have linear regression (solid) and LOWESS non-parametric regression line (middle broken line) with 95% confidence envelopes around the fit (external broken lines). All relationships were nonsignificant (p > 0.05).

Fig. 4. Cox survival analysis in patients with or without splenomegaly. Splenomegaly defined by the Sprogøe-Jakobsen and Sprogøe-Jakobsen [11] table. No statistical survival difference was observed between the groups of patients with or without splenomegaly, adjusted for age, gender, height, and weight (p = 0.84).
Discussion

This study demonstrates for the first time that splenomegaly, predominantly of mild degree, is common in patients with advanced idiopathic or heritable PAH. The presence of chronic congestion as the sole macro- and microscopic finding in the enlarged spleens on autopsies suggests that chronic passive congestion is likely the cause of the splenomegaly. However, the spleen volume estimated by CT showed no association with clinical, echocardiographic, or hemodynamic parameters or survival.

To the best of our knowledge, there is no available information on the spleen size in patients with idiopathic and heritable PAH. Hepatomegaly and ascites, but not splenomegaly, are frequently included as part of signs indicative of the presence of right heart failure in these patient groups [4, 20]. However, one of the common causes of splenomegaly is congestion due to heart failure. We hypothesized that as patients develop right heart failure as a result of longstanding PAH, some degree of splenomegaly would become evident.

We found that splenomegaly was common (52–63%) in patients with advanced idiopathic and heritable PAH, irrespective of the definition used. We measured the spleen volume by using two multidimensional indices [9, 10], and compared the volumes obtained with those expected for height and weight as proposed previously [11]. We defined splenomegaly when splenic volumes were above the 95% CI for height and weight. This last approach was important as in our cohort, splenic volume correlated with body weight in multivariate analysis. Interestingly, we observed that the overall increase in spleen size was modest and this is in correlation with what is seen in congestive heart failure from other etiologies [6].

Hyposplenia was rare, since only 1 or 2 patients, depending on the formula used, had this condition (fig. 1). Hoeper et al. [21] described a higher prevalence of asplenia in patients with idiopathic PAH when compared with individuals with other lung diseases who received lung transplant (11.5 vs. 0%). Fahy et al. [22] reported hyposplenia in a patient with idiopathic PAH potentially explained by splenic ischemia due to reduced cardiac output, hypoxia, and polycythemia. In our study, we did not find an association between spleen size and either cardiac output or hemoglobin concentration in the blood. In our cohort, the patient with hyposplenia had a cardiac index of 3.4 l/min/m², PVR of 6 Wood units, hemoglobin of 13.7 g/dl and Spo₂ at rest on room air of 98%.

We were not able to detect an association (linear or non-linear) between splenic volume, either as a continuous or dichotomized variable, and NYHA functional class, laboratory data (including complete blood counts and comprehensive metabolic panel), 6-min walk distance, and echocardiographic or hemodynamic parameters. Similarly, splenic volume at the time of the initial CT was not associated with survival. These findings do not support the routine measurement of spleen volume during CT performed to further patients with idiopathic or heritable PAH.

As part of the present study, we collected information from autopsies performed in patients with idiopathic or heritable PAH who died of right heart failure. We observed that the spleen weight was 220 (IQR: 151–325) g, and 55 and 33% of the patients had a splenic weight ≥ 200 g.

Fig. 5. Pathology examination of the spleen in idiopathic and heritable PAH. Chronic congestive splenomegaly showing a thickened and fibrous capsule (a; HE, ×4), fibrotic and cellular red pulp (b; HE, ×20), dilated sinusoids (c; HE, ×20) and hemosiderin deposition from episodes of old hemorrhage (HE, ×40).
and ≥250 g, respectively (proposed cutoff points for splenomegaly) [6, 16, 23, 24]. There is significant variability in the normal spleen weight or volume [11]; hence we used different methods to define splenomegaly. The weight of the spleen differs in vivo from autopsy specimens due to blood loss during the process of extraction of the organ to be weighed [16, 17]. We also noted these differences in our study given that the in vivo spleen weight was estimated (spleenic volume x spleen specific gravity of 1.05 g/ml) at 361 (IQR: 236–560) g.

There are limitations to our study. We did not determine the spleen volume by the summation-of-volumes technique (gold standard), although spleen length and the multidimensional indices that include length, width, and thickness have shown excellent correlation with the gold standard technique [19, 25, 26]. Thus we decided to use these simpler indexes that reliably estimate splenic size instead of other labor-intensive and clinically impractical methodologies [19, 25]. We included patients (14%) in whom the splenic volume was determined in a CT of the abdomen performed for abdominal distension. This could have potentially selected a subgroup of patients with a larger spleen; however, the spleen size was similar between patients who underwent CT of the abdomen or chest, and we took precautions not to include patients with any other pathology known to affect the spleen volume.

Conclusions

Splenomegaly of mild degree is common in patients with advanced idiopathic and heritable PAH. We did not find supportive data to routinely assess splenic volume in idiopathic or heritable PAH patients.

Acknowledgments

This publication was made possible by CTSA KL2 [grant RR024990] (A.R.T.) from the National Center for Research Resources, a component of the National Institutes of Health (NIH), and NIH Roadmap for Medical Research.

Financial Disclosure and Conflicts of Interest

The authors have no significant conflicts of interest with any companies or organization whose products or services may be discussed in this article. G.A.H. has received a board fee from Lung Rx United Therapeutics and his institution has received grants from the Gilead Sciences Research Scholars Program and from Bayer HealthCare Pharmaceuticals.

References


