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Pulmonary Neovascularity
A Distinctive Radiographic Finding in Eisenmenger Syndrome

Ramon Sheehan, MD; Joseph K. Perloff, MD; Michael C. Fishbein, MD; David Gjertson, PhD; Denise R. Aberle, MD

Background—We sought to characterize the distinctive pulmonary vascular abnormalities seen on chest radiographs and computed tomography (CT) scans in Eisenmenger syndrome.

Methods and Results—Thoracic CT scans, chest radiographs, and clinical data were reviewed for 24 Eisenmenger syndrome patients subdivided into those with interatrial (pretricuspid) versus interventricular or great arterial (posttricuspid) communications and in 14 acyanotic patients with pulmonary arterial hypertension (PAH) and no congenital heart disease. CT scans were scored blindly by 2 thoracic radiologists for the presence and severity of small, tortuous, intrapulmonary vessels, termed “neovascularity,” lobular ground-glass opacification, and systemic perihilar and intercostal vessels. Histopathologic lung sections from 5 patients with Eisenmenger syndrome and from 3 patients with acyanotic PAH were reviewed. Associations between clinical and imaging features were tested by ANOVA and χ² tests. Kendall’s rank-order coefficient and the Kruskal-Wallis test were used to test for significant differences in imaging features between Eisenmenger syndrome and acyanotic PAH. Neovascularity on chest radiographs was more common in Eisenmenger syndrome than acyanotic PAH, but differences were not significant. On CT, neovascularity, lobular ground-glass opacification, and hilar and intercostal systemic collaterals were more prevalent in Eisenmenger syndrome, with severity greater in posttricuspid communications. Three previously undescribed vascular lesions were identified histologically in Eisenmenger syndrome: malformed, dilated, muscular arteries within alveolar septa; congested capillaries within alveolar walls; and congested capillaries within the walls of medium-size, muscular pulmonary arteries. These lesions may correspond to the distinctive vascular abnormalities observed on chest radiographs and CT scans.

Conclusions—Distinctive vascular lesions on chest radiographs and CT scans in Eisenmenger syndrome appear to be correlated histologically with collateral vessels that develop more extensively with posttricuspid communications. (Circulation. 2005;112:2778-2785.)

Key Words: collateral circulation • heart defects, congenital • hypertension, pulmonary • lung • imaging
Methods

Patient Selection
Twenty-four consecutive adults, 10 men and 14 women aged 22 to 53 years (mean, 39±2), with unoperated cyanotic congenital heart disease were selected from the Ahmanson/UCLA Adult Congenital Heart Disease Center Registry after referral for CT pulmonary angiography performed between May 1994 and September 2000 in anticipation of lung transplantation. Six patients had >1 CT scan performed; in these cases, the analyzed scan was the 1 done closest to diagnostic echocardiography. Imaging data were also collected from 14 consecutive patients >18 years with cyanotic PAH, both primary and acquired forms, who had CT pulmonary angiography performed between April 1995 and September 1998 within 24 months of the diagnosis of PAH.

The congenital cardiac defects in Eisenmenger syndrome were established by transthoracic echocardiography with color flow imaging and Doppler interrogation performed on average within 6.0±7.1 months of the CT scans. Pulmonary hypertension in the patients with cyanotic PAH was established by transthoracic echocardiography and cardiac catheterization performed within 3.4±7.1 months of the CT scans.3

Age and sex data were secured from medical records. Systemic arterial oxygen saturation and hematocrit/hemoglobin values were recorded within 3 weeks of imaging studies. The UCLA institutional review board exempted this retrospective study from the approval process or patient informed consent.

Image Acquisition

Chest Radiographs
Upright posteroanterior and lateral chest radiographs were taken at a 72-inch source-to-image distance with high-kilovoltage techniques (110 to 140 kV) with various screen-film or computed radiography receptor systems of ~200 speed.

CT Scans
CT pulmonary angiograms were obtained on either an Imatron Evolution electron beam CT (EBT) scanner (Imatron, Inc) or a GE HiSpeed CT/i scanner (General Electric Medical Systems). Scans were acquired at suspended maximal inspiration. Patients unable to breath-hold were preferentially studied at quiet respiration on the Imatron EBT scanner. Each study included an initial noncontrast sequence to exclude pulmonary vascular calcifications, followed by intravenous contrast-enhanced vascular sequences from 1 cm above the superior margin of the transverse aorta to 1 cm above the lower 2 hemidiaphragms.

Initial non–contrast-enhanced, continuous-volume datasets on the EBT scanner were acquired through the thorax according to a standardized protocol of 6-mm collimation, 6-mm table incrementation, and 300-ms exposure. Images were reconstructed with use of a high-spatial-frequency algorithm. Vascular sequences were performed in single-slice, continuous-volume mode with 3-mm collimation, 2-mm table incrementation, 100- to 200-ms exposure, and a standard reconstruction algorithm.

Studies acquired on the GE HiSpeed CT/i scanner consisted of an initial noncontrast sequence of 120 kV, 200 to 260 mA, 0.8- to 1.0-mm gantry rotation speed, 5-mm collimation, pitch of 1.5:1 to 2:1, 5-mm reconstruction interval, and a bone reconstruction algorithm. This was followed by a vascular sequence after administration of intravenous contrast of 3-mm collimation, pitch of 2:1, 2-mm reconstruction interval, and a standard reconstruction algorithm. Images were reviewed on softcopy with both mediastinal (width, 400 HounsfieId units [HU]; level, 40 HU) and lung (width, 1600 HU; level, –550 HU) window settings.

For all vascular sequences, a peripheral catheter was inserted into a large forearm or antecubital vein and avoiding introduction of air. Omnipaque 350 (Amersham Health) was administered by mechanical injection at standard flow rates of 3 to 4 mL/s, averaging 90 to 140 mL total. Vascular sequences were initiated 25 to 30 seconds after the start of the contrast bolus.

Figure 1. Chest radiograph of a 32-year-old man with a nonrestrictive VSD and Eisenmenger syndrome. Small, nodular opacities in the lungs correspond to pulmonary neovascularity grade 2. Enlarged central pulmonary arteries are indicative of PAH.

Image Interpretation

Two subspecialty-trained thoracic radiologists (R.S., D.R.A.) independently reviewed all chest radiographs and CT images. Both were aware that all patients were pulmonary hypertensive but were blinded to other clinical and histological data. Image analyses were standardized so that lung windows were reviewed first, enabling all relevant observations to be recorded before soft-tissue windows of the cardiac lesions were revealed. Chest radiographs and CT images were read in settings separated by at least 12 weeks to ensure that recall would not affect visual scoring. Scores were compared and inconsistencies reconciled by agreement after all images had been reviewed.

Chest radiographs and CT scans from 3 patients with cyanotic congenital heart disease outside the time period under consideration were used to establish visual standards for developing semiquantitative scoring systems for the presence and severity of specific imaging features by using principles developed for other visual scoring systems.4 These visual standards contained features spanning the range of features being scored on the study images and were available for reference during image analysis. Chest radiographs were scored for the presence and severity of small, nodular opacities within the lungs on a 4-point, semiquantitative grading system: 0=no opacities, 1=opacities mildly pronounced, 2=opacities moderately pronounced, 3=opacities markedly pronounced (Figure 1).

Similarly, CT scans were scored on a 4-point, semiquantitative grading system (0=absent, 1=mild, 2=moderate, 3=extensive) to determine the presence and severity of each of the following vascular patterns: (1) Neovascularity, defined as tiny, micronodular, serpiginous intrapulmonary vessels often in the subpleural lung or in proximity to centriflobular arterioles, coursing in directions inconsistent with known arteriolar anatomy (Figure 2A and 2B). (2) Lobular ground-glass, defined as hazy, increased lung attenuation at the center of secondary lobules fading to normal attenuation at the lobular margin (Figure 3). (3) Hilar collateral vessels, defined as small, serpiginous, vascular channels coursing in the cardiac or bilateral hilar regions adjacent to mainstem, lobar, or interlobar arteries or bronchi (Figure 4). (4) Intercostal collaterals, defined as small, serpiginous, vascular channels coursing in chest wall soft tissues in proximity to pleural surfaces (Figure 5). These appeared as “beading” along the visceral pleural margin and occasionally entered the lung to join intrapulmonary vessels, forming serpiginous neovascularity in the lung periphery (Figure 6A and 6B). Intercostal vessels had to be increased in number to be judged abnormal.
Neovascularity and lobular ground-glass were scored on lung windows, whereas collateral vessels were scored on soft-tissue windows. Pulmonary artery thrombus, recanalized thrombus, pulmonary artery mural calcifications, and incidental cardiopulmonary findings were recorded as present or absent.

Histopathology

Histological material was studied from 5 patients with Eisenmenger syndrome, 1 with an ostium secundum atrial septal defect (ASD) and 4 with posttricuspid lesions, and in 3 patients with acyanotic PPH. The lung specimens from patients with PPH were obtained during surgery for lung transplantation, whereas Eisenmenger syndrome specimens were obtained at necropsy.

Lung specimens were fixed in formalin and embedded in paraffin. Sections of 5-μm thickness were stained with hematoxylin-eosin for light microscopy. All sections were analyzed for the following vascular lesions: (1) medial hypertrophy, (2) intimal proliferation, (3) plexiform lesions, and (4) vascular dilatation lesions. Vascular lesions not corresponding to these patterns were specifically recorded. All specimens were reviewed by the same cardiopulmonary pathologist (M.C.F.). The number of vascular lesions was scored on individual histological sections according to the same 4-point, semiquantitative grading system used to grade vascular lesions on CT. Systematic sampling or scoring throughout the lungs was not possible, nor was it possible to correlate lung specimens directly with in vivo CT image data.
statistical significance. Hemodynamic data were tested by ANOVA and a
and age, sex, systemic arterial oxygen saturation, hematocrit, and
Associations between individual chest radiographic or CT findings

Statistical Methods

Associations between individual chest radiographic or CT findings and age, sex, systemic arterial oxygen saturation, hematocrit, and hemodynamic data were tested by ANOVA and a $\chi^2$ test for continuous and categorical variables, respectively. To assess for significant differences in chest radiographic and CT findings between patients with Eisenmenger syndrome versus acyanotic PAH, Kendall’s rank-order correlation coefficient ($\tau_b$) was computed, and the Kruskal-Wallis test was applied to the ordinal data. To highlight differences, semiquantitative scores for chest radiographic and CT features for each group were also dichotomized in 2 ways: (1) negative (grade 0) versus positive (grades 1 to 3) and (2) mild (grade 1) versus extensive (grades 2 and 3). All probability values were 2 sided; after allowing for multiple comparison via Bonferroni adjustment, statistical significance was set at $P<0.004$.

To analyze differences in CT features in Eisenmenger syndrome, these patients were further subdivided into those with pretricuspid lesions$^{5,6}$ versus those with posttricuspid lesions (ventricular septal defects [VSDs] and/or great arterial communications). Because of the small numbers of patients, results were expressed as percentages along with exact (binomial) 95% confidence intervals (CIs); tests of significance for proportions among comparative grades were not done. Because correlations between histological data and the CT image data were not possible, no attempt was made to analyze the histological data for statistical significance.

Results

Clinical Features

Among 24 patients with Eisenmenger syndrome, echocardiography disclosed a pretricuspid lesion (ostium secundum ASD) in 7 patients and posttricuspid lesions in 17, including VSD in 9, truncus arteriosus in 2, and 1 each with patent ductus arteriosus (PDA), PDA with ASD, VSD with PDA, double-outlet right ventricle with VSD, complete transposition of the great arteries with VSD/ASD/PDA, and endocardial cushion defect with nonrestrictive VSD. The 7 patients with pretricuspid lesions were believed to represent the coincidence of PPH and ASD or stretched patent foramen ovale.$^{5,6}$

Transcutaneous systemic arterial oxygen saturations in Eisenmenger syndrome ranged from 68% to 87% (mean, 78±6%), and iron-replete hematocrit levels (Beckman Coulter Inc) ranged from 58 to 71 gm/dL (mean, 64±4).

Right ventricular systolic pressures by echocardiography were at systemic level in Eisenmenger syndrome patients with posttricuspid lesions; right ventricular/pulmonary artery systolic pressures ranged from 102 to 138 mm Hg in those with pretricuspid lesions.

The 14 patients with acyanotic PAH included 2 men and 12 women aged 20 to 76 years (mean, 44±4). Eight of 14 met the clinical and echocardiographic criteria for PPH.$^5$ The remaining 6 patients had acquired pulmonary hypertension caused by systemic or parenchymal disease, including 2 with chronic obstructive pulmonary disease, 2 with systemic lupus erythematosus, 1 with chronic thromboembolic disease, and 1 with portal hypertension. Transcutaneous systemic arterial oxygen saturation in the 14 patients with acyanotic PAH was ≥96%, hematocrits ranged from 33 to 44 mg/dL (mean, 37±2), and echocardiographic right ventricular systolic pressures ranged from 42 to 118 mm Hg (mean, 83±9).

There were no significant associations between chest radiographic or CT features and age, sex, systemic arterial oxygen saturation, hematocrit, or right ventricular systolic pressure in the 2 major groups (Eisenmenger syndrome and acyanotic PAH).

Chest Radiographic Findings

Chest radiographs were available for review in 26 patients. Small, nodular, vascular opacities were visible on chest radiographs of 12 of 17 (71%) with Eisenmenger syndrome and 3 of 9 (33%) with acyanotic PAH, but the difference was not significant (Table 1).

<table>
<thead>
<tr>
<th>TABLE 1. Frequency and Extent of Nodular Opacities on Chest Radiographs (N=26*)</th>
<th>Radiographic Grading</th>
<th>Patients Graded as Positive (Grades 1–3)</th>
<th>Patients Graded as Severe (Grades 2 and 3)</th>
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</thead>
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<tr>
<td>Patient Category</td>
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<tr>
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<tr>
<td>Eisenmenger syndrome</td>
<td>5</td>
<td>4</td>
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*Numbers in parentheses represent percentages of total positive.

†Kendall’s $\tau_b$ along with its asymptotic SE. The $P$ value from the Kruskal-Wallis test of equal proportions of grading ranks is presented parenthetically. Statistical significance was $P<0.004$.

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There were significantly increased numbers of small peri-
haematoxylin and eosin (H&E)-stained sections of the lungs of patients with Eisenmenger syndrome and those with acyanotic PAH, being present more commonly and with greater severity in Eisenmenger syndrome (correlations equaled 0.68, 0.48, and 0.65, respectively; Table 2).

Neovascularity was present on CT scans in 23 of 24 (96%) of Eisenmenger patients and in 3 of 14 (21%) of acyanotic PAH patients (Table 2). Moreover, among Eisenmenger patients, neovascularity was more severe (grade 2 or 3) in posttricuspid lesions (15/17, or 88%; 95% CI, 64% to 89%) than in pretricuspid lesions (1/7, or 14%; 95% CI, 0% to 41%).

Lobular ground-glass was a CT feature in 15 of 24 patients (62%) with Eisenmenger syndrome versus only 1 of 14 patients (7%) with acyanotic PAH (Table 2). When Eisenmenger syndrome patients were subdivided, the magnitude of this difference was unchanged: lobular ground-glass of any severity (grades 1 to 3) was seen in 11 of 17 patients (65%; 95% CI, 38% to 86%) with posttricuspid lesions and in 4 of 7 patients (57%; 95% CI, 18% to 90%) with pretricuspid lesions. Severe grades of lobular ground-glass were found in 7 patients (57%; 95% CI, 18% to 90%) with Eisenmenger syndrome and in 4 of 14 (21%) of acyanotic PAH patients.

Hilar and intercostal collaterals

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*Kendall’s τ and along with its asymptotic SE. The P value from the Kruskal-Wallis test of equal proportions of grading ranks is presented parenthetically. Statistical significance was P<0.004.

CT Findings
Overall, the CT features of neovascularity, lobular ground-glass, and hilar/intercostal collaterals differed significantly between patients with Eisenmenger syndrome and those with acyanotic PAH, being present more commonly and with greater severity in Eisenmenger syndrome (correlations equaled 0.68, 0.48, and 0.65, respectively; Table 2).

Neovascularity was present on CT scans in 23 of 24 (96%) of Eisenmenger patients and in 3 of 14 (21%) of acyanotic PAH patients (Table 2). Moreover, among Eisenmenger patients, neovascularity was more severe (grade 2 or 3) in posttricuspid lesions (15/17, or 88%; 95% CI, 64% to 89%) than in pretricuspid lesions (1/7, or 14%; 95% CI, 0% to 41%).

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Histological Observations
Three patients with acyanotic PPH from whom histological material was available had medial hypertrophy and intimal proliferation but no plexiform lesions. There was a large, recanalized thrombus within the proximal pulmonary arteries in 1 patient.

Medial hypertrophy and intimal proliferation were present in 3 of 5 Eisenmenger syndrome patients. Classic plexiform lesions corresponding to Heath-Edwards grade 4 were identified in specimens from 3 patients (Figure 7), and vascular dilatation and advanced lesions of plexiform pulmonary arteriopathy were present in all 5 Eisenmenger syndrome patients.

In Eisenmenger syndrome, 3 previously undescribed vascular lesions were identified by light microscopy. Two of the lesions involved abnormal capillaries. The first consisted of numerous dilated, congested capillaries within adventitial tissue surrounding medium-size, muscular, pulmonary arteries (Figure 7A). The second consisted of markedly dilated, congested capillaries within alveolar tissues (Figure 7B). The third lesion, which consisted of clusters of abnormally dilated, tortuous, muscular arteries within alveolar septa (Figure 7C), was visible on slide preparations without magnification and appeared to represent the small, tortuous, neovascularity vessels observed on CT. Figure 7D shows a plexiform lesion of hypertensive pulmonary arteriopathy.

Discussion

The histological types of pulmonary vascular disease described by Heath and Edwards are equally prevalent in
Eisenmenger syndrome and acyanotic PAH. The distinctive radiographic neovascularity lesions described herein are significantly more frequent in Eisenmenger syndrome and are most severe in Eisenmenger syndrome with posttricuspid right-to-left shunts at the ventricular or great arterial level. Chest radiographs disclosed distinctive nodular opacities within the lungs. On CT, neovascularity appeared as small, nodular, and serpiginous vessels in the lung periphery, occurring in conjunction with lobular ground-glass opacification. In Eisenmenger syndrome, increased systemic arterial collateral vessels were seen in the hilar and intercostal regions; the intercostal vessels sometimes penetrated the lung and contributed to neovascularity.

In Eisenmenger syndrome, we report previously undescribed histological vascular lesions characterized by marked capillary dilatation and congestion in alveolar spaces and in the adventitial tissue surrounding small, muscular arteries. Abnormal clusters of small, muscular arteries were visible within the alveolar walls without magnification and had the distinctive serpiginous morphology of neovascularity seen on CT. Although direct radiological/histopathologic confirmation was not possible, the distinctive neovascularity lesions suggest collateralization around tiny resistance vessels rather than Heath-Edwards lesions of plexogenic pulmonary arteriopathy. Lobular ground-glass may be an indirect reflection of collateral flow below the limits of macroscopic resolution, correlating with dilated capillary networks in alveolar spaces.

Systemic-to-pulmonary vascular collateral vessels have long been known and primarily involve bronchial arteries that nourish the airways and serve as vasa vasorum for pulmonary arteries. More controversial has been the role of the systemic circulation in providing precapillary and capillary bronchopulmonary anastomoses, although necropsy studies with current 3-dimensional, computer-aided techniques have disclosed different types of collaterals in the lungs of patients with cyanotic congenital heart disease. Systemic collateral channels develop at various levels in the pulmonary circulation, depending on hemodynamic circumstances, and may serve as tiny compensatory pathways downstream from obstructive pulmonary vascular disease or as bypass channels that divert flow from proximal prestenotic lesions to distal dilation lesions and alveolar capillaries.

The development of systemic-to-pulmonary collaterals may vary according to the timing and severity of pulmonary vascular disease. Pretricuspid and posttricuspid communications in Eisenmenger syndrome have different pathophysiological and hemodynamic consequences that may account for the significant differences in the degrees of neovascularity and lobular ground-glass appearance. In normal fetuses, a nonrestrictive ductus arteriosus equals pulmonary arterial, systemic arterial, and right and left ventricular systolic pressures. This equalization of pressures persists in infants with nonrestrictive posttricuspid defects, so the direction of shunt flow depends on the relative resistances in the pulmonary and systemic vascular beds. At birth, pulmonary vascular resistance falls precipitously to systemic levels and then continues to fall during the next few months. As resistance falls, the magnitude of a left-to-right shunt increases. Survival then depends chiefly on curtailment of excessive pulmonary flow by the development of pulmonary vascular disease. Adults with nonrestrictive posttricuspid lesions have, by definition, pulmonary hypertension at systemic levels, but it is suprasystemic pulmonary vascular resistance that reverses the shunt, diminishes pulmonary arterial blood flow, and stimulates formation of systemic collateral circulation to the lung parenchyma. Conversely, nonrestrictive pretricuspid defects are not accompanied by pulmonary vascular disease, reversed shunt, and diminished pulmonary blood flow and do not stimulate formation of systemic collaterals to the lung parenchyma. The magnitude and direction of shunts do not depend on systemic and pulmonary vascular resistances but on the relative compliances of the right and left ventricles, which are similar in utero and at birth but then diverge as left ventricular mass exceeds right ventricular mass during the process of normal extraterine growth. Left-to-right shunt flow awaits this regression of right ventricular mass and increase in compliance, a time course that permits the pulmonary bed to evolve.
into a low-pressure, low-resistance circulation. When a left-
to-right shunt is then established, pulmonary arterial pressure
remains normal within a low-resistance compliant vascular
bed. Eisenmenger syndrome is uncommon in patients with a
nonrestrictive ASD because pulmonary vascular disease is an
unlikely sequel of increased flow into a low-pressure, low-
resistance pulmonary circulation. Accordingly, it has been
proposed that Eisenmenger syndrome in adults with an ASD
represents the coincidence of the interatrial communication
and PPH.6

Neovascularity and lobular ground-glass were significantly
more common in Eisenmenger syndrome than in acyanotic
PAH and were more common with posttricuspid defects in
which pulmonary vascular disease developed early in life.
Because our patients were adults, we do not know at what age
neovascularity first becomes manifest.

Certain alternative explanations for the increased lung
attenuation in Eisenmenger syndrome can be discounted.
Pulmonary venous hypertension and pulmonary edema result
in vascular redistribution (arterial and venous), together with
interstitial edema, peribronchial cuffing, indistinct perihilar
vessels, septal lines, thickening of fissures, and pleural
effusions. Pulmonary edema may appear as increased lung
attenuation on CT but is usually in a bronchovascular or
gravity-dependent distribution and does not exhibit the dis-
tinctive central pattern within individual lobules that we
observed.

Intrapulmonary hemorrhage, also relatively common in
Eisenmenger syndrome, appears on chest radiographs and CT
scans as transient single or multiple confluent, segmental
areas of ground-glass attenuation or consolidation.22–24 Con-
versely, neovascularity and lobular ground-glass attenuation
were permanent features in those patients in whom serial
scans were available for review.

Lobular ground-glass can be mistaken for mosaic attenua-
tion, which is common with chronic thromboembolism and
other noncardiac causes of pulmonary hypertension.25 How-
ever, the lobular ground-glass that we observed had charac-
teristic high attenuation at the center of the pulmonary lobule
and normal attenuation at the lobular margin contiguous with
the interlobular septum, and the distribution of lobular
ground-glass was relatively diffuse. In contrast, mosaic per-
fusion appears as geographic regions, or clusters, of lobules
of uniform increased attenuation alternating with lobules of
decreased attenuation. In an individual lobule, attenuation is
homogeneous and does not taper peripherally at the septal
margin. Increased attenuation in mosaic perfusion reflects
regions of normal or hyperperfused lung supplied by the
remaining pulmonary circulation. We postulate that the lob-
ular ground-glass in Eisenmenger syndrome represents cen-
trilobular perfusion by small, systemic-to-pulmonary collat-
erals at or below the arteriolar level.

Increased numbers of systemic arterial collaterals in the
carinal, hilar, and intercostal regions on CT were universal in
Eisenmenger syndrome but were significantly less frequent in
acyanotic PAH. Intercostal vessels entered the lung directly
across the costal pleura in Eisenmenger syndrome. There was
no significant correlation between CT observations of sys-
temic collaterals and either lobular ground-glass or
neovascularity.

The retrospective design of our study created some limita-
tions. Selection was based on referral for cross-sectional
imaging, usually in anticipation of lung transplantation or for
evaluation of chronic pulmonary thromboembolism. Because
the study groups were not matched for demographic vari-
ables, unintentional selection bias might have been intro-
duced. The retrospective design precluded direct correlation
between CT and histological data. Neovascularity on CT
strikingly resembled the histology of dilated, tortuous, mus-
cular arteries clustered within alveolar tissues, but direct
correlation would have been required to be certain that these
CT and histological findings represented the same lesion.
Finally, our analysis involved relatively few patients, which
limits our ability to confirm the significance of differences
between Eisenmenger patients with posttricuspid and posttri-
cuspid lesions.

In conclusion, we report distinctive features on chest
radiographs and CT scans that were significantly more
common in Eisenmenger syndrome than in acyanotic PAH
patients. On CT, neovascularity, lobular ground-glass, and
increased systemic collaterals were identified regardless of
the etiology of PAH, although they were significantly more
common in Eisenmenger syndrome. Neovascularity and
ground-glass opacification were more prevalent and more
severe in posttricuspid defects in which pulmonary hyperten-
sion was present early in life, suggesting that the lesions were
influenced by time of onset and duration of pulmonary
vascular disease.

Histological material from patients with Eisenmenger syn-
drome revealed typical plexiform lesions as well as previ-
ously undescribed vascular abnormalities: Malformed, tortu-
os vessels, and dilated, congested capillaries within alveolar
septa and the pulmonary arterial adventitia. We postulate
that these lesions can be correlated with the CT findings of
neovascularity and lobular ground-glass, respectively, and
that they are manifestations of tiny collaterals that develop
primarily in the lungs of patients with Eisenmenger syndrome.

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