A 68-year-old woman with a 3-year history of breathlessness (World Health Organization functional class III) was diagnosed with idiopathic pulmonary arterial hypertension.

Investigations included a 12-lead ECG, chest radiograph, and computed tomography scan of the thorax (Figure 1, A-C). At cardiac catheterization, mean pulmonary arterial pressure and pulmonary vascular resistance were elevated (73 mm Hg and 1545 dyne s cm⁻⁵, respectively), and cardiac index was reduced (1.9 L/min/m²). A solitary 60% left circumflex coronary artery stenosis was present. Cardiovascular magnetic resonance identified right ventricular (RV) hypertrophy (indexed mass of 64 g/m²; normal 16 to 36 g/m²), right ventricular systolic dysfunction (RV ejection fraction 31%; normal >57%), and paradoxical interventricular septal (IVS) motion. Late gadolinium-enhanced (LGE) images taken 10 minutes after injection of 0.1 mmol/kg gadolinium–diethylenetriamine pentaacetic acid showed enhancement of both insertion regions (Figure 2A, 2B, and 2D).

The patient died suddenly 6 weeks later. Examination of the heart revealed normal macroscopic appearances at both insertion regions (Figure 2C). Microscopy showed myocardial disarray (Figure 2E) with increased collagen and fat between fiber bundles (plexiform fibrosis; Figure 2G) but no microscopic scars (ie, replacement fibrosis). We found the same in the only other published images of pulmonary arterial hypertension insertion-region microscopy.¹

This is the first pathologic correlation of insertion-region enhancement in pulmonary arterial hypertension, which, when viewed beside the aforementioned images,¹ raises the possibility that LGE in pulmonary hypertension may not be due to pathologic fibrosis.

The insertion regions are prisms, appearing triangular in cross section, where RV and left ventricular fibers cross.² These interdigitating fibers are wedged between more regular and compact layers containing the subendocardial, mesocardial, and subepicardial fibers.³ Thus, myocardial disarray is a normal finding in these areas. Likewise, plexiform fibrosis (defined by collagen interlacing myocardial fibers in areas of disarray⁴ and not to be confused with plexiform lesions, which are seen in the pulmonary arteries of patients with pulmonary arterial hypertension) is a normal feature of insertion-region anatomy.³

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Figure 1. A, Twelve-lead ECG: sinus rhythm, right-axis deviation, and a dominant R wave in lead V1. B, Chest radiograph showing cardiomegaly and enlargement of the proximal pulmonary arteries (arrow). C, Computed tomography scan confirming pulmonary artery dilatation.
We believe that insertion-region architecture is exaggerated in pulmonary hypertension by hypertrophy of the right ventricle and IVS coupled to shear forces arising from paradoxical IVS motion (Figure 2F) and speculate that LGE is related to contrast pooling within areas of myocardial disarray and the plexiform fibrosis therein. This would explain why LGE occurs in the midmyocardium in quantities related to indexed RV mass. Disruption of the midseptum’s normally orderly fiber orientation may account for the midseptal extension of LGE in paradoxical IVS motion.

Based on these limited observations, further study is required before the etiology of insertion-region LGE in this disease group (and others in which it is found) can be definitively proven. If the elements behind it are normal but accentuated, rather than pathologic, it would seem likely that this finding in pulmonary hypertension would have no incremental prognostic value over existing indices of RV remodeling.

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None.

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References


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