Beware the pregnant woman with breathlessness

A 32-year-old woman in the 20th week of her third pregnancy was referred to the advanced lung disease service with breathlessness. As a child she could not keep up with the other children and was told that she had a "hole in the heart" but this did not require further investigation or treatment. In previous pregnancies she had been breathless and fatigued from mid-trimester and into the postpartum period, but she thought this was a normal change in pregnancy.

Examination revealed a soft systolic murmur and loud second heart sound. Echocardiography showed an abnormal dilated left atrium, mildly dilated right ventricle and an estimated pulmonary artery systolic pressure of 60 mm Hg, but no evidence of an atrial septal defect, ventricular septal defect or patent ductus arteriosus. VQ (ventilation–perfusion) scan showed no evidence of thromboembolic disease. High-resolution CT chest excluded interstitial lung disease.

Cardiac MRI (fig 1) revealed an unusual cause for her pulmonary hypertension.

**QUESTION**
What is the cause of her pulmonary hypertension?
See page 586 for answer.

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