An 82-year-old woman was transferred from her local hospital to our National Pulmonary Hypertension Service because of the suspicion of pulmonary hypertension. A month previously, she attended her local Accident & Emergency Department with a minor injury to her left leg. During triage an oxygen saturation of 85% was noted. She was cyanotic but she did not report any breathlessness, respiratory rate was in a normal range, and she was normotensive. On history she reported being limited when walking, which she considered normal for her age and she remained fully independent. Chest X-Ray (Figure 1) showed a large hernia that occupied central and right chest, raising suspicions of a Bochdalek hernia. High-resolution computed tomography and computed tomography pulmonary angiogram were performed and confirmed the presence of a large posterolateral Bochdalek hernia with the stomach and portions of small and large bowel occupying a substantial portion of the right thoracic cavity and associated with right lung hypoplasia (Figure 2A–2D). There was no evidence of pulmonary embolus, and the lung parenchyma was normal. Abdominal ultrasound did not show any features of liver disease, and clinical and serological investigations for connective tissue disease were negative as was viral serology. ECG showed sinus rhythm with incomplete right bundle-branch block and signs of right heart strain (S1Q3T3; Figure 3). On echocardiography (Figure 4 and Movies I–III in the online-only Data Supplement) she had an enlarged right ventricle and right atrium with normal left heart and without intracardiac shunts. She had moderate tricuspid regurgitation with a calculated right ventricular end-systolic pressure of 96 mm Hg plus right atrial pressure. Patient was offered to undergo right heart catheterization but she refused that. At the end, she was discharged on chronic oxygen therapy.

On detailed history the patient reported that she had been noted to have a bluish discoloration from childhood. Before this admission she had not been to hospital or had a chest radiograph for >15 years. Although she had good access to public health system, she did not go to see doctors very frequently, because she was not feeling ill. She does recall on one previous occasion, many years ago, being told she had a hiatus hernia. The most probable explanation of this abnormal and benign progression of her disease is that the herniation was progressive during her life. It is likely that the hernia was smaller when she was a child and herniation worsened gradually, producing progressive pulmonary hypertension and hypoxia.

Congenital diaphragmatic hernias are an uncommon finding in adult population; in the case of Bochdalek hernia, its true prevalence ranges from 1/2000 to 1/7000 in autopsy studies1 to an incidental rate of 0.17%2 in a series of abdominal CT, although in most cases only modest herniation is found. It results in the herniation of the abdominal viscera through the foramina of Bochdalek as a result of failure to close during the embryogenesis or postnatal reopening.2 Bochdalek hernia normally manifests during the first few weeks of life, and it is one the leading causes of respiratory distress of the neonate requiring urgent surgical repair.2 Respiratory compromise and pulmonary hypertension are some of the main problems that require management pre- and postoperatively in these pediatric patients.3 Lung and

Figure 1. Posterior-anterior plain chest X-Ray. A large diaphragmatic hernia with gastric and bowel content is evident in the right and center chest.
pulmonary vascular bed hypoplasia, as well as abnormal vascular vasodilator response, have been proposed as the mechanisms causing pulmonary hypertension. Pulmonary hypertension and its persistence after surgical repair is an important prognostic factor in these children. Inhaled nitric oxide, prostanoids, and even sildenafil have been tried anecdotally.

To our knowledge, this is the first time that pulmonary hypertension has been linked to congenital diaphragmatic hernia in an adult patient. Thus we describe a novel mechanism of pulmonary hypertension in adults, and highlight the necessity of considering this complication in adult patients with large incidental diaphragmatic hernias that requires surgical repair. Such patients may require special care in the post-operative period because of the risk of respiratory and right heart failure, and pulmonary vasodilator therapies should be considered.

**Disclosures**

None.

**Figure 2.** Contrast CT chest. **A,** Axial view. **B,** Coronal view. **C,** Sagital view. **D,** Lung window. Herniation of abdominal content, including stomach and large and small bowel into the chest is evident and arises from the posterior and right side of the diaphragm.

**Figure 3.** Twelve-lead ECG showing sinus rhythm with incomplete right bundle-branch block and signs of right ventricular strain, as S1Q3T3 pattern.
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