A 6-year-old boy was referred for follow-up of a ventricular septal defect, which was first diagnosed when he was a neonate. Cardiac catheterization performed at 7 months of age revealed an aberrant left subclavian artery arising from the right aortic arch. Recurrent respiratory infection that caused stridor and/or wheezing occurred in early infancy, but it was deemed not related to the cardiac defect because the pulmonary-to-systemic flow ratio was 1.6:1 and there was no pulmonary hypertension. With time, his respiratory symptoms remitted, and he was lost to follow-up for several years, while his family worried about his dysphagia. No heart murmur was auscultated, and the second heart sound was split and fixed. Electrocardiography showed incomplete right bundle-branch block. Ultrasonography indicated a small infundibular ventricular septal defect and right ventricular volume overload with intact atrial septum.

Contrast-enhanced 16-row multislice computed tomography was performed, and three-dimensional reconstruction (Figure 1) visualized anomalous connection of the right-upper pulmonary vein to the superior vena cava (A) and the right retroesophageal aortic arch with an aberrant left subclavian artery (B). It also revealed the esophagus breaking off above the aorta and running inferiorly from right to left (C). Esophagography confirmed severe esophageal stenosis (Figure 2; available at www.jpeds.com). The patient underwent surgical division of the ligamentum arteriosum, repair of the anomalous pulmonary venous connection, and direct closure of the ventricular septal defect through a median sternotomy.

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Figure 1. A, Three-dimensional reconstruction of contrast-enhanced multislice computed tomography (frontal view). The right-upper pulmonary vein returns to the superior vena cava (open arrow). B, Posterior view showing the right retroesophageal aortic arch running inferiorly from right to left (white arrow), an aberrant left subclavian artery (white arrowhead), and the diverticulum of Kommerell, which is a remnant of the embryonic left arch (open arrowhead). C, Superimposition of esophageal and tracheobronchial images. The esophagus (green) breaks off above the right retroesophageal aortic arch (asterisk).
Figure 2. Esophagogram in left anterior oblique view showing marked posterior indentation (arrow).