A 10-week-old male infant was referred for a systolic heart murmur, mild congestive heart failure with tachypnea, tachycardia, mildly enlarged liver, and a chest x-ray showing an enlarged cardiac silhouette with plethoric pulmonary vasculature (A). The transthoracic echocardiogram revealed the diagnosis of Shone’s complex with a normal-sized, apex-forming left ventricle and enlarged right atrium (RA) and right ventricle (RV) as well as a moderate-size ventricular septal defect (VSD). The aortic annulus size was normal, and aortic arch appeared slightly hypoplastic. All 4 pulmonary veins were connected unobstructed to the left atrium (LA). A large levoatriocardinal vein (LACV) was found, allowing the left atrium to decompress via the innominate vein and right-sided superior vena cava into the RA (B, Online Videos 1 and 2). This left-to-right shunt explained the dilated RA and RV. Magnetic resonance imaging of the heart and vessels confirmed the connection of LA with the innominate vein via LACV (C and D, Online Video 3). The patient underwent successful surgical correction with excision of the supramitral ring, patch closure of VSD, and ligation of LACV.