Magnetic Resonance Imaging of Complex Partial Anomalous Pulmonary Venous Return in Adults

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Adults with congenital cardiac abnormalities surpass the number of children because of better assessment of cardiac anatomy and function, monitoring, operative options, patient outcomes, and increased survival into adulthood. Partial anomalous pulmonary venous return may present in adulthood, particularly if asymptomatic with small shunt fraction. One or more pulmonary veins, most commonly an anomalous right upper pulmonary vein, connect to a systemic vein or the right atrium; this accounts for 0.5% of congenital cardiac defects. Cardiovascular imaging continues to evolve rapidly for accurate preoperative evaluation and operative planning.

Although transthoracic echocardiography is the first-line tool for diagnosis, suboptimal acoustic windows may preclude adequate inspection of pulmonary veins or atrial septal defect to assess for appropriate baffling. Disadvantages of computed tomography include an inability to calculate shunt fraction, ionizing radiation, and a greater risk of nephrotoxicity than magnetic resonance imaging. Anatomically, MRI provides noninvasive volumetric anatomic data and enables evaluation of systemic veins, as well as the number, origin, course, and drainage of all pulmonary veins, including anomalous connections or obstruction. Detection rate for each pulmonary vein is 57% for the right superior pulmonary vein, 62% in the left superior pulmonary vein, 76% in the right inferior pulmonary vein, and 86% in the left inferior pulmonary vein. MRI also examines the presence and type of atrial septal defect; quantifies ventricular volumes; evaluates the right ventricular outflow tract and pulmonary trunk, sites of stenosis, or aneurysmal right ventricle–pulmonary artery conduits or branch pulmonary arteries; and quantifies pulmonary regurgitation and shunts, biventricular function, ejection fraction, myocardial viability, flow in the ascending aorta (including aneurysm, dissection, coarctation, assessment of aortopulmonary collaterals, and arteriovenous malformations), coronary anomalies, and coronary artery disease. Cardiac MRI is versatile for evaluation of tissue characteristics and has a superior ability to evaluate cardiovascular physiology, assess viability and perfusion, and detect myocardial fibrosis, as well as cine and shunt-fraction quantification in complex partial anomalous pulmonary venous return, compared with other imaging modalities.

We present the case of a 57-year-old woman who presented to Brigham and Women’s Hospital with shortness of breath, New York Heart Association class II heart failure, and no symptoms of angina after a myocardial infarction; she had been taking aspirin and clopidogrel after left anterior descending artery stent placement. She consented to the case report. She had severe pulmonary hypertension (systolic pulmonary artery pressure 110 mm Hg) while taking bosentan. As shown in the Figure, preoperative pulmonary artery (panel A) and lateral (panel B) chest radiography demonstrated a widened mediastinum caused by massively enlarged pulmonary arteries, as seen on cardiac MRI in the sagittal (panel C) and cross-sectional (panel D) view.

Because of an inability to visualize all pulmonary veins and the interatrial septum preoperatively on echocardiography or computed tomography scan, cardiac MRI was obtained, which demonstrated no obvious atrial septal defect. There were normal-sized ventricles with normal systolic function and left ventricular ejection fraction of 66%. The septum was D shaped in systole and diastole, consistent with right ventricular pressure and volume overload, moderate systolic dysfunction, severe right atrial dilation, and severe tricuspid regurgitation with 42% regurgitant fraction, as well as mild pulmonary valve regurgitation with regurgitant fraction of 16%. The main pulmonary artery (Figure, E) and both right (panel F) and left pulmonary arteries were severely dilated. The pulmonary to systemic flow ratio of Qp/Qs=1.26 suggested a small left-to-right shunt. Partial anomalous pulmonary venous return was seen on dynamic magnetic resonance angiography, with the right upper, right middle, and a small branch of the right lower pulmonary veins communicating with the right-sided superior vena cava (Figure, G) anomalously. The remaining small branches of the right lower lobe pulmonary vein and entire left upper and left lower pulmonary veins drained into the left atrium normally. The coronary sinus was dilated, measuring 26 mm (Figure, H), and the left brachiocephalic vein drained into a duplicated left-sided superior vena cava. The right-sided superior vena cava was thus small, draining normally into the right atrium (Figure, H).
On the basis of the MRI findings, the preoperative plan was to divert the anomalous right-sided veins via a surgically created atrial septal defect, because none existed, and an anastomosis between the proximal transected end of the superior vena cava and right atrial appendage, or Warden procedure.3 The patient underwent tricuspid valve repair with a #32 Carpentier-Edwards annuloplasty ring; resection of the fossa ovalis to create a baffled repair for the partial anomalous pulmonary venous return for the anomalous pulmonary venous connection of the right upper, right middle, and right lower lobe veins; and a subsequent Warden procedure from the high transected superior vena cava to the right atrial appendage. Postoperatively, she did well, with patent nonobstructed right pulmonary veins and a decrease in systolic pulmonary artery pressure from 110 to 70 mm Hg on routine follow-up echocardiography.

Future MRI of partial anomalous pulmonary venous return repair will precisely quantify any residual shunt, patency, anastomotic narrowing, and ventricular systolic function for adults with congenital heart disease.4 Care of these adult patients with congenital heart disease requires a facility with appropriate training in cardiac MRI, appropriate pathophysiological understanding, and specialists committed to long-term collaboration between clinicians, including, at a minimum, cardiologists, surgeons, and radiologists, to manage such patients.

Disclosures

None.

References


Figure. Pulmonary artery and lateral chest radiography and chest magnetic resonance imaging demonstrating severe right, left, and main pulmonary artery dilatation, as well as classic features of partial anomalous pulmonary venous return of the right upper, right middle, and right lower lobes, with dilated left coronary sinus and relatively small right superior vena cava. Chest radiography of the pulmonary artery (*) in A and B; lateral view in B and cardiac magnetic resonance imaging of the pulmonary artery (**) in C and D in the sagittal (C) and cross-sectional (D) views demonstrated a widened mediastinum caused by dilated main pulmonary artery (**) in C, left, and right pulmonary arteries related to a left-to-right shunt in partial anomalous pulmonary venous return of the right upper, middle, and lower lobe veins. E, Dilated main pulmonary arteries (*). F, Dilated right pulmonary artery (*). G, Anomalous drainage of right upper (**) and right middle (**) pulmonary veins to right atrium–superior vena cava junction (*). H, Dilated coronary sinus (*) receiving blood flow from left brachiocephalic vein and small right superior vena cava (**).
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