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Cardiac stress MRI evaluation of anomalous aortic origin of a coronary artery

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Abstract

Myocardial ischemia is an insult that is primarily thought of in an adult population. However, there are several congenital and acquired cardiac lesions that may lead to myocardial ischemia in a pediatric population. One of the prominent congenital lesions is anomalous aortic origin of a coronary artery (AAOCA). Anomalous aortic origin of a coronary artery is one of the leading causes sudden cardiac death in pediatric and young adult patients, and thus the assessment of myocardial perfusion is of the utmost importance. Over the past decade, pharmacologic stress MRI has proven to be a highly sensitive and accurate diagnostic examination for qualifying myocardial perfusion in adults with coronary artery disease. This noninvasive imaging modality may be a useful tool in assessing the function impact of AAOCA on myocardial perfusion.

KEYWORDS

anomalous aortic origin of a coronary artery, coronary anomalies, myocardial perfusion, stress MRI, sudden cardiac death

Myocardial ischemia may occur in a number of clinical settings within the pediatric population, although its prevalence is much less than in the adult population. Coronary artery disease (CAD) may develop after surgical repair of congenital heart disease that involves the coronary arteries, such as arterial switch for transposition of the great arteries, anomalous origin of the left coronary artery from the pulmonary artery, or anomalous aortic origin of the coronary artery (AAOCA).[1–4] Additionally, acquired pediatric heart disease (AHD) such as Kawasaki disease may result in CAD.[5] The coronary lesions may cause myocardial ischemia, which is a known risk factor for morbidity and mortality in children.[6] Cardiac stress testing may be performed with different modalities to assess for signs of impaired myocardial perfusion. Nuclear medicine studies have commonly been used within the pediatric population to assess patients with both congenital and AHD at risk for myocardial ischemia.[7] Although this technique has shown the ability to detect ischemia, nuclear medicine studies are limited by lower spatial resolution for small defects, attenuation artifacts related to the body wall and diaphragm, relatively high incidence of false positive findings,[8] and potentially harmful effects of ionizing radiation. Likewise, x-ray coronary angiography may be able to identify areas of coronary stenosis or kinking, but does not depict the functional significance of the coronary abnormality, and is also invasive and associated with ionizing radiation.

Cardiac magnetic resonance imaging (CMR) is a noninvasive technique that provides high quality imaging of the cardiac structures with excellent spatial resolution, and is not associated with ionizing radiation. Cardiac magnetic resonance imaging perfusion imaging, utilizing the first-pass perfusion kinetics of a gadolinium bolus to detect myocardial ischemia, has shown excellent sensitivity and specificity in the adult population.[9,10] Likewise, CMR perfusion has also been performed in pediatrics for a variety of conditions that carry a risk of myocardial ischemia with good results.[11,12] Although coronary hyperemia agents are most commonly given, dobutamine is an alternative pharmacologic stress agent that may be safely utilized for the assessment of myocardial ischemia.[13] Dobutamine is a cardiac inotropic agent that increases myocardial contractility, while decreasing systemic vascular resistance. Historically, dobutamine was utilized to illicit wall motion abnormalities during the time of increased myocardial oxygen demand. This form of analysis has shown to be highly sensitive for the detection of coronary artery stenosis.[14,15] With the aid of a perfusion sequence at the peak of dobutamine infusion, the sensitivity and prognostic value has further increased with dobutamine stress MRI.[16,17] In addition to their use in adults with CAD, dobutamine stress MRI has been utilized within the pediatric population, and has shown to be more accurate than stress echocardiography, a common modality within pediatrics.[18,19]

Anomalous aortic origin of the coronary artery is a complex pathophysiology, which results in an increased risk of sudden cardiac death.[20,21] Cardiac CT may be utilized to accurately elucidate the different aspects of AAOCA, such as the ostial morphology, presence and length of an intramural course, and the presence of an interarterial course, all of which may play a role in causing ischemia.[22] Because of the complex pathophysiology, many different methods of stress have been utilized in an attempt to identify those patients that are at the highest risk of sudden death.[23,24] Dobutamine stress may be an advantageous pharmacologic stress agent, as the increase in inotropy and decrease in systemic vascular resistance, may closely mimic exercise. The increase in cardiac output, and thus the increase in aortic blood volume, may play a role in the pathophysiology of AAOCA, especially in those patients with a prolonged intramural course. At our institution, we have begun to employ dobutamine stress MRI for patients with an increased risk for myocardial ischemia whereby the pathophysiology may be related to increased inotropy or cardiac output such as AAOCA or myocardial bridge.[25,26] The basic protocol involves balanced steady-state free precession sequences for volumetry and functional analysis. Following these cine sequences, 0.1 mmol/kg of gadolinium contrast is intravenously injected, and resting perfusion is assessed with a T_1-weighted, gradient echo sequence in the left ventricular short-axis. At approximately 6–8 minutes following contrast administration, myocardial delayed hyperenhancement is assessed with phase-sensitive, inversion recovery sequences in the short-axis and 4-chamber planes. Dobutamine is then initiated at 10 mcg/kg/min and increased by 10 mcg/kg/min every 4 minutes, to a peak dose of 40 mcg/kg/min. At each stage, wall motion is assessed with balanced steady-state free precession sequences at 3 ventricular levels and the 4-chamber. To reach peak heart rate, 0.01 mg/kg of atropine is given, if the peak heart rate has not been achieved at the final stage of dobutamine. At peak stress, the perfusion sequence is repeated, again with intravenous injection of gadolinium contrast agent, to assess myocardial perfusion at peak stress.

Thus far in our experience, 5 patients have undergone dobutamine stress MRI. The indications have been 3 patients with myocardial bridge of the left anterior descending with a family history of sudden cardiac death, 1 patient following arterial switch operation with an intramural course of the left anterior descending coronary artery, and 1 patient with hypoplastic left heart syndrome with ST segment changes on ECG. All patients underwent dobutamine stress MRI without complications and without adverse events. Two of the patients with myocardial bridge were found to have a perfusion defect in the left anterior descending coronary artery distribution, and at operation were found to have significant depth to the myocardial bridge, with visible caliber change of the coronary vessel. The patient with arterial switch procedure was found to have an irreversible perfusion defect in the left anterior descending coronary distribution, and underwent cardiac catheterization and was found to have a complete occlusion of the coronary vessel. No further procedure was performed.

Pediatric patients have several different pathologies that place them at higher risk of myocardial ischemia. Cardiac MRI has been shown to be highly accurate and sensitive to the identification of myocardial ischemia, and has been shown to be feasible in pediatric patients. Dobutamine stress MRI may be a useful technique in patients with pathophysiology that is related to an increase in cardiac inotropy, but further research is needed to demonstrate its utility within these populations.

CONFLICT OF INTEREST

None.

DISCLOSURES

None.

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