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CASE REPORT

Anomalous Coronary Arteries: Anatomic and Functional Assessment by Coronary and Perfusion Cardiovascular Magnetic Resonance in Three Sisters

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ABSTRACT

Combined coronary and perfusion cardiovascular magnetic resonance was performed in three sisters with angina and suspected anomalous coronary arteries. Two sisters had anomalous coronary arteries passing between the aorta and right ventricular outflow tract and had abnormal myocardial perfusion. One sister had normal anatomy and perfusion. The combined approach identified the anatomy and functional significance of suspected anomalous coronary arteries.

INTRODUCTION

Coronary arteries are described as ''anomalous'' when they arise from an alternate aortic sinus of Valsalva or from the pulmonary artery; these occur in 0.3–1.0% of the population (1–3). In autopsy and cardiac catheterization series (4,5), approximately 60% are anomalous circumflex arteries (1) and the remaining 40% are divided equally between anomalous right and left coronary arteries. The clinical significance of anomalous coronary arteries is due to their association with sudden cardiac death in otherwise normal adults. The initial diagnosis is usually made with cardiac catheterization, but because this is a projection technique, identification of the proximal course of the vessel can be difficult (6). This is important because the syndrome of sudden cardiac death is associated with anomalous coronary arteries that have an interarterial course, passing between the aorta and right ventricular outflow tract or main pulmonary artery.

Cardiovascular magnetic resonance (CMR) has been recommended as a diagnostic investigation in these patients because it can accurately and noninvasively define

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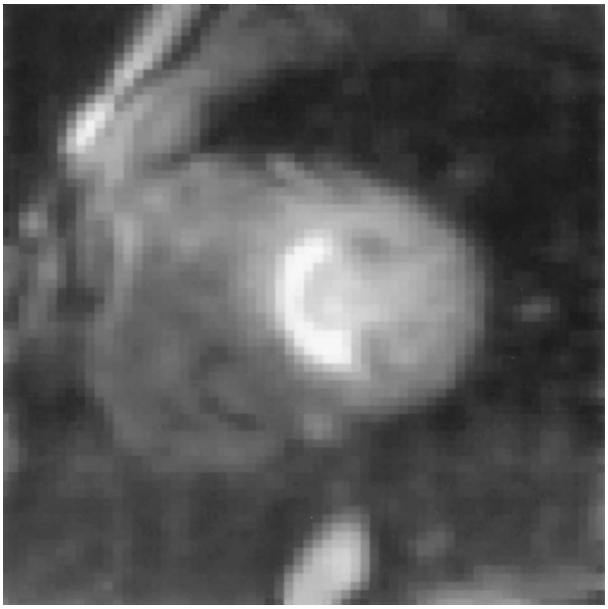
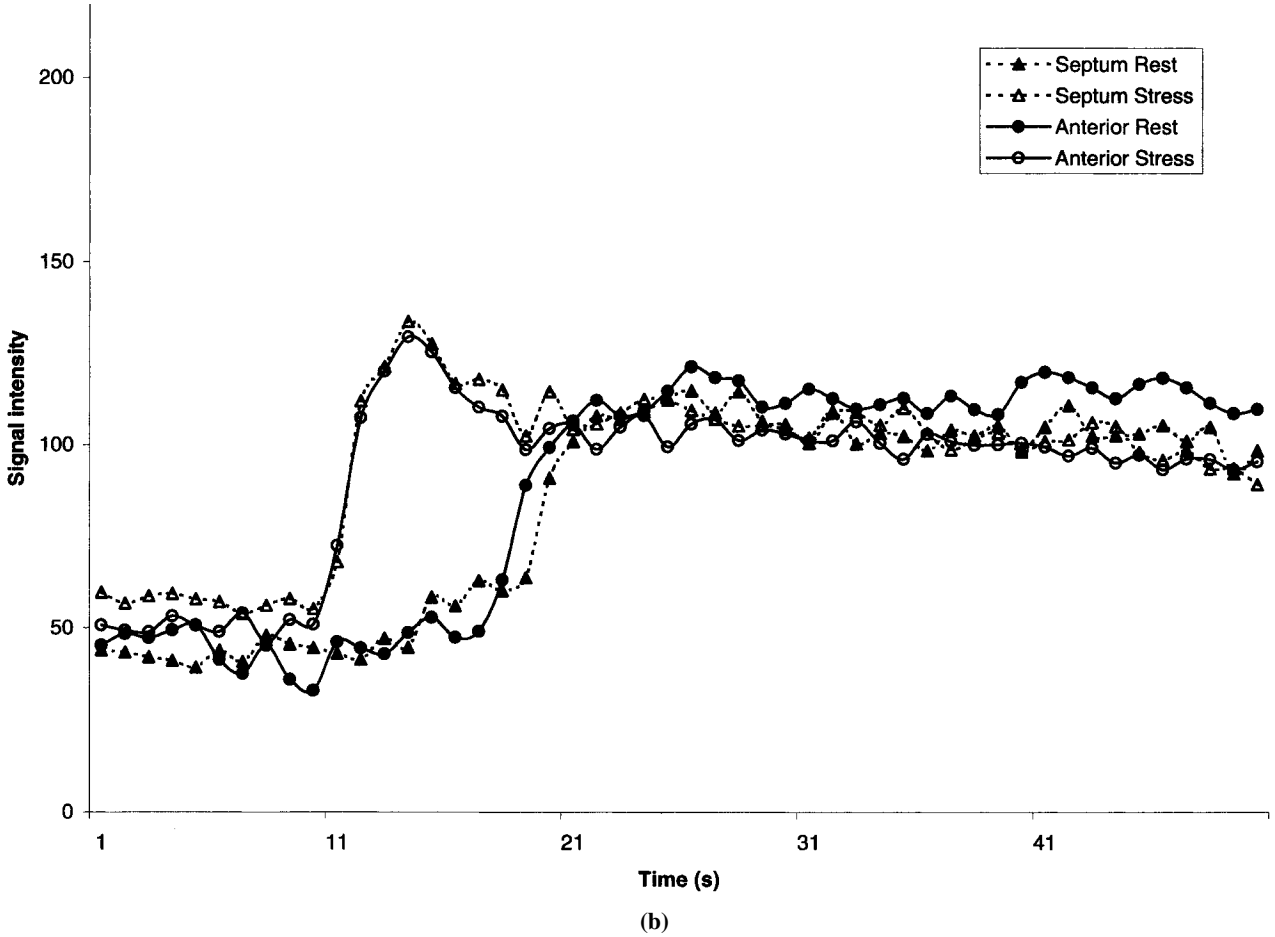


(a)



(b)

Figure 1. Patient 1. (a) Perfusion gradient echo of the basal short-axis slice of the left ventricle acquired after adenosine stress. There is homogenous myocardial signal intensity. (b) Signal intensity curves obtained at rest and after stress for the septum and anterior wall, showing identical signal enhancement patterns.

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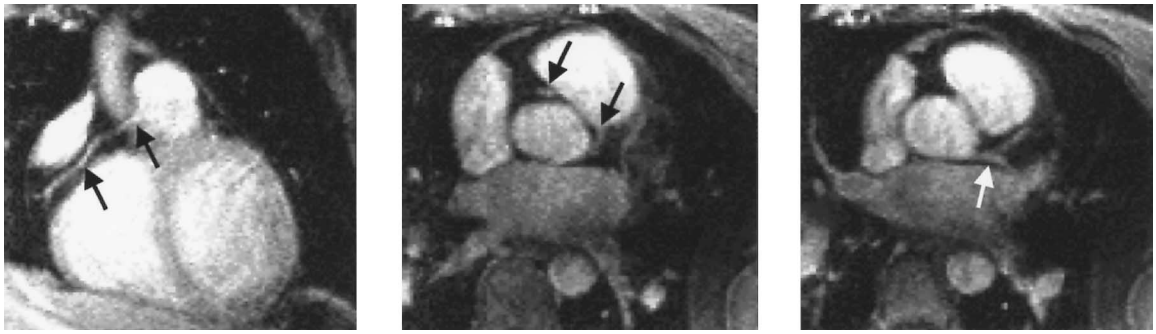


(a) (b) (c)

Figure 2. Patient 2. (a) A double oblique segmented gradient echo fast low-angle shot CMR image. The right coronary artery is anomalous and arises from the left sinus of Valsalva and is visible traversing from its origin toward the right atrioventricular sulcus (arrows). (b) A transverse section demonstrates the origin of the anomalous right coronary artery and its passage between the aorta and right ventricular outflow tract (arrows). (c) The left coronary artery arises in a normal anatomic position from the left sinus of Valsalva.

the coronary artery anatomy by tomographic imaging in multiple planes (7–11). Multiple two-dimensional slices have been used with excellent results for the diagnosis of anomalous coronary arteries (12–14), and more recently a similar technique has been applied in patients with adult congenital heart disease (15). However, these studies have only addressed the anatomic arrangement of the arteries and have not studied the functional significance of the anomaly.

In this study, we investigated the coronary artery anatomy in three young female siblings with angina who were suspected of having coronary anomalies using coronary CMR. We also investigated the functional significance of the anomaly by using adenosine pharmacologic stress and ''first-pass'' myocardial perfusion with gadolinium-DTPA. First-pass myocardial perfusion has been investigated as a method of assessing patients with coronary artery stenoses to determine abnormalities of signal intensity and impaired coronary flow reserve index (16,17). However, this method has not yet been investigated in patients with anomalous coronary arteries to assess the functional effects of the anomaly.

MATERIALS AND METHODS

Patients

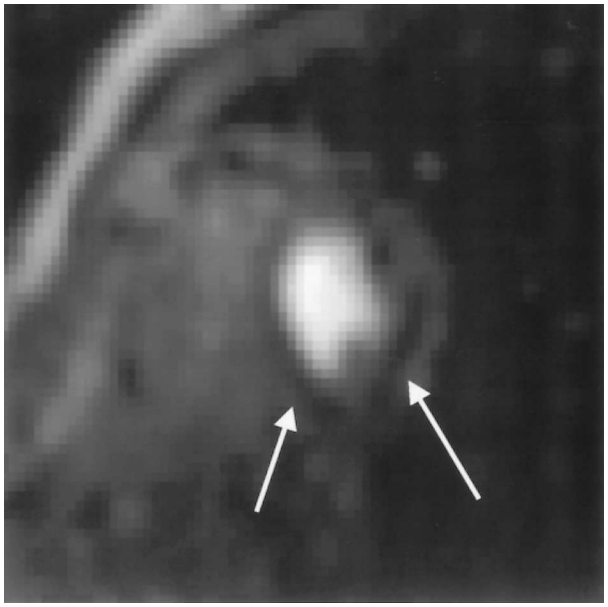
Three sisters with angina were evaluated with CMR. They also had cardiac catheterization (performed using the standard Judkins technique) and myocardial perfusion single photon emission computed tomography (SPECT) performed using standard methods.

Imaging Protocol

CMR was performed with a 1.5-T Edge scanner (Picker, Cleveland, OH) using a four-channel phased array coil centered over the precordium. All sequences were cardiac gated.

For the coronary CMR, coronal, sagittal, and transverse pilot scans were acquired to identify the aortic sinuses and proximal coronary artery segments. From these, both transverse and double oblique volume acquisitions were performed to cover the coronary arteries. The imaging sequence was a segmented gradient echo fast low-angle shot sequence: repetition time/echo time of 12.9 msec/4.3 msec, eight views per data segment with an incremental flip angle of 20–90 degrees, and voxel dimensions of $1.0 \times 1.4 \times 2.5$ mm. Data were acquired in mid-diastole, and respiratory artefacts were reduced by using prospective respiratory gating (18) and phase reordering (19).

For the perfusion CMR, two short-axis slices were positioned through the left ventricle and acquired with an ultrafast gradient echo sequence: repetition time/echo time of 3.0 msec/2.0 msec, flip angle 18 degrees, presaturation flip angle 90 degrees, and voxel dimensions of $2.0 \times 7.8 \times 10.0$ mm. The sequence was cardiac gated, acquiring both slices in a single R-R interval, with scan duration lasting 50 heartbeats, while the patient performed shallow respiration. Although newer scanners are capable of acquiring more slices per cardiac cycle, limited coverage as used in this study has also been shown to be useful for assessment of perfusion defects (20). A baseline myocardial perfusion study was performed us-



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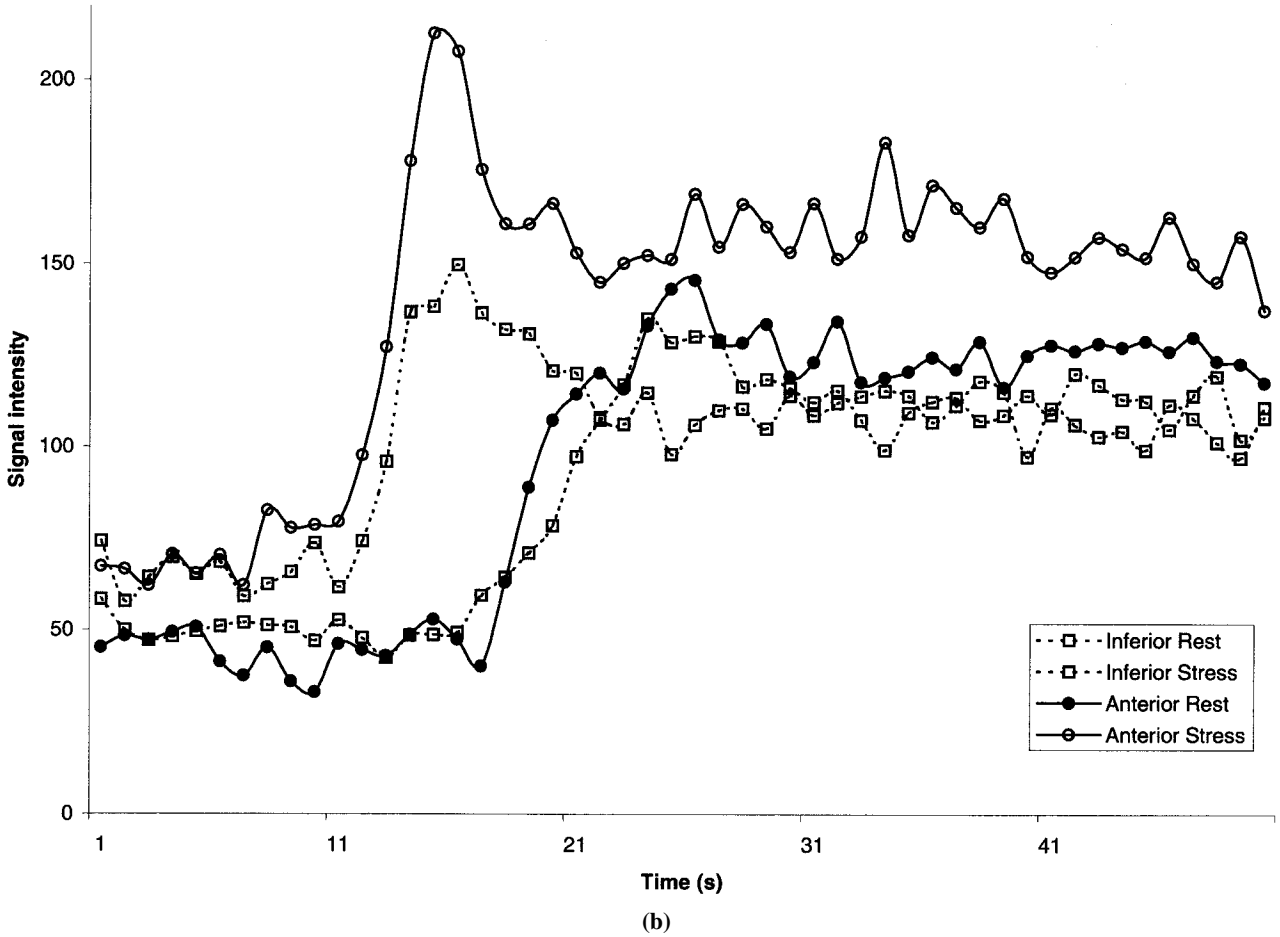

(b)

Figure 3. Patient 2. (a) Perfusion gradient echo image of the basal short-axis slice of the left ventricle acquired after adenosine stress. There is reduction in myocardial signal intensity in the inferolateral wall (arrows). (b) Signal intensity curves from the inferior and anterior walls confirm the delayed and reduced enhancement in the inferior wall.

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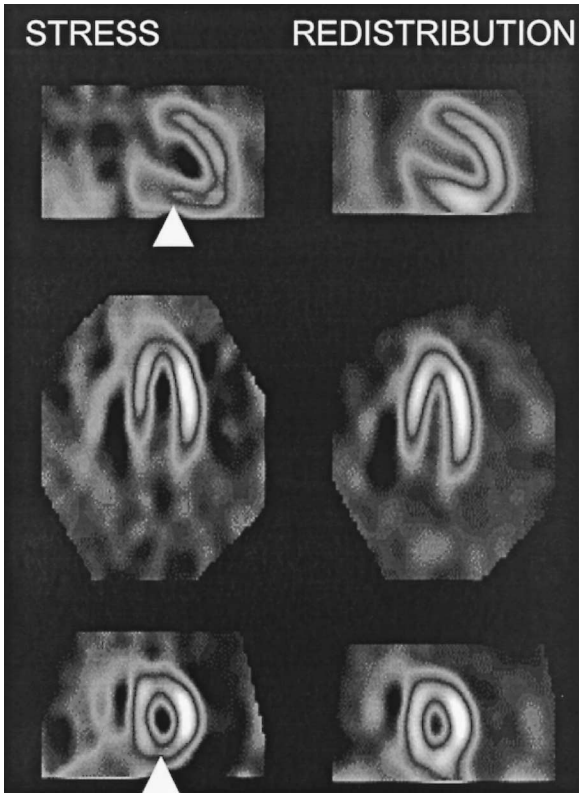


Figure 4. Patient 2. Myocardial perfusion SPECT image acquired after adenosine stress. There is an inferolateral perfusion defect (arrowheads) that was normal at the time of the subsequent rest acquisition.

ing 0.05 mmol/kg gadolinium injected with a power injector (Medrad, Spectris®, Pittsburgh, PA) into an antecubital vein. The patient was removed from the magnet bore during the next 20 min while the gadolinium cleared from the myocardium. Adenosine 140 μg/kg/min was then infused via a separate cannula for 6 min, and then with the patient repositioned within the magnet bore, a stress myocardial perfusion scan was acquired, with the identical parameters as the rest perfusion scan. The blood pressure and electrocardiogram were monitored every 3 min.

Analysis

The CMR images were analyzed off-line using imaging software CMR tools (Royal Brompton and Harefield NHS Trust©). The coronary CMR images were visually assessed to determine the anatomic position of the three coronary arteries and to assess for the presence of significant coronary artery stenoses or signal drop-out. The CMR perfusion images were reviewed in a cine loop, displaying rest and stress images for both slices. In the rest scan, uniform uptake of contrast agent as evidenced by signal intensity increase was judged to be indicative of normal resting perfusion. In the adenosine scan, the rate of signal intensity increase in the myocardium was assessed visually. A delay in signal intensity increase, as compared with other segments, was judged to be indicative of abnormal myocardial perfusion.

RESULTS

Good quality images were obtained for all three subjects. The mean scanning time was 40 min.

The first patient (38 years old) had a resting heart rate of 68 beats/min, increasing to 93 after adenosine, whereas blood pressure fell from 120/80 to 110/65. On CMR coronary artery anatomy was normal with no significant stenosis identified, and first-pass myocardial perfusion was normal before and after adenosine stress (Fig. 1). Nuclear perfusion scanning showed normal tracer uptake in stress and rest studies. X-ray angiography identified mild coronary artery disease with no stenosis greater than 50%.

The second patient (44 years old) had a resting heart rate of 76 beats/min and blood pressure of 110/60, which increased to 120 beats/min and 120/60 after adenosine. On three-dimensional coronary MR imaging there was an anomalous right coronary artery that originated from the left posterior sinus of Valsalva and passed between the aorta and right ventricular outflow tract (Fig. 2). There was no signal loss in any vessel and no evidence of a significant stenosis. However, during first-pass myocardial perfusion there was a reduction in the signal intensity in the inferolateral region of the left ventricle (Fig. 3). Radioisotope scanning also demonstrated an inferolateral stress perfusion defect (Fig. 4). X-ray angiography identified a normal left coronary artery system, but the right coronary artery could not be selectively cannulated. An aortogram demonstrated a patent right coronary artery with no obstructive coronary disease (Fig. 5).

The third patient (34 years old) had a resting heart rate of 74 beats/min and blood pressure of 160/90, which increased to 124 and 170/105 after adenosine. On three-dimensional coronary MR imaging there was an anomalous single coronary artery that arose from the right anterior sinus of Valsalva. The artery then divided to form a right coronary artery that descended in the right atrioventricular sulcus and a left coronary artery that passed posteriorly between the aorta and right ventricular outflow tract to enter the interventricular sulcus (Fig. 6). At this

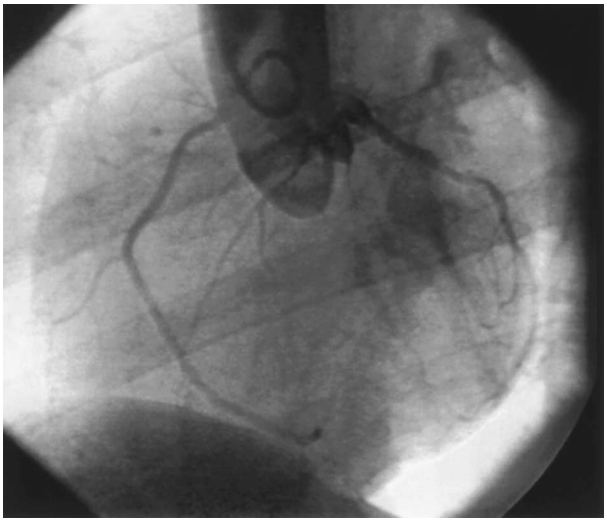


Figure 5. Patient 2. An aortogram demonstrates the patency of the left and right coronary arteries. However, the anatomic relationship between the right coronary artery and the aorta and right ventricular outflow tract is not evident.

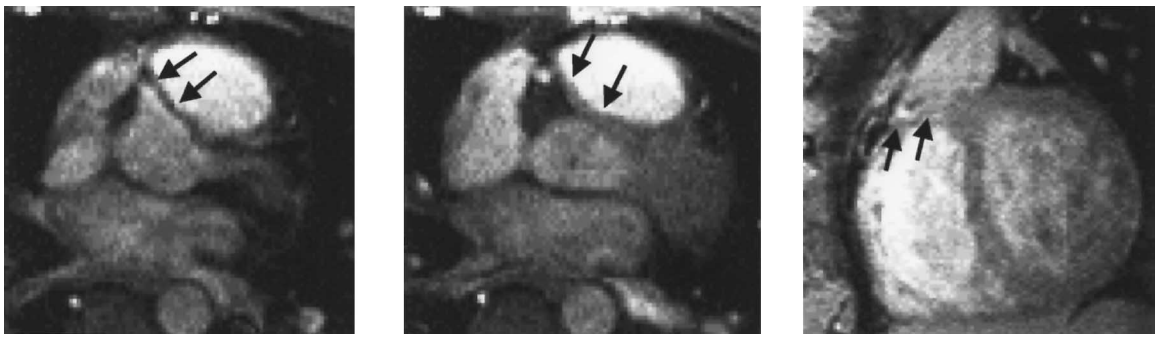
point the artery divided to form the circumflex and left anterior descending arteries. There was no MR evidence for a significant stenosis. On first-pass myocardial perfusion, during adenosine infusion there was a reduction in the signal intensity in the septum and anteroseptal wall (Fig. 7). The stress nuclear perfusion images showed a reduction of tracer uptake in the mid and apical parts of the anterior wall and septum (Fig. 8). X-ray angiography confirmed a single coronary artery but with no significant coronary artery stenoses (Fig. 9).

The first patient was treated with antianginal medication. The other two siblings had successful coronary artery surgery and remain free of angina.

DISCUSSION

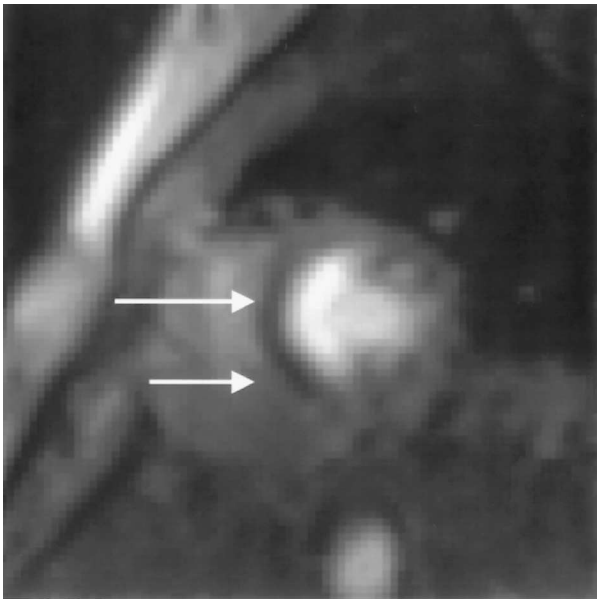
Anomalous coronary arteries are an uncommon but important congenital abnormality. The initial diagnosis is usually made after x-ray angiography in a patient with angina or aborted sudden death. CMR has been advocated as a necessary investigation to define the proximal anatomy of anomalous coronary arteries and their relationships to the aorta and right ventricular outflow tract. Initial CMR reports have focused on confirming the anatomy without investigating the functional significance of the anomaly. However, functional consequences are key issues when considering cardiac surgery in relatively young patients. Most centers will consider additional radionuclide scanning to document myocardial ischemia (21).

In this case report of three sisters we demonstrated that CMR can identify the anatomic abnormality but can also demonstrate the functional consequences with delayed gadolinium uptake on adenosine myocardial perfusion imaging, which correlates with reduced tracer uptake on thallium scanning. However, the physiologic explanation for delayed myocardial perfusion is uncertain. Both patients with anomalous coronary arteries had no significant coronary artery stenoses, although both arteries passed between the aorta and right ventricular outflow tract. Putative explanations for the etiology of sud-

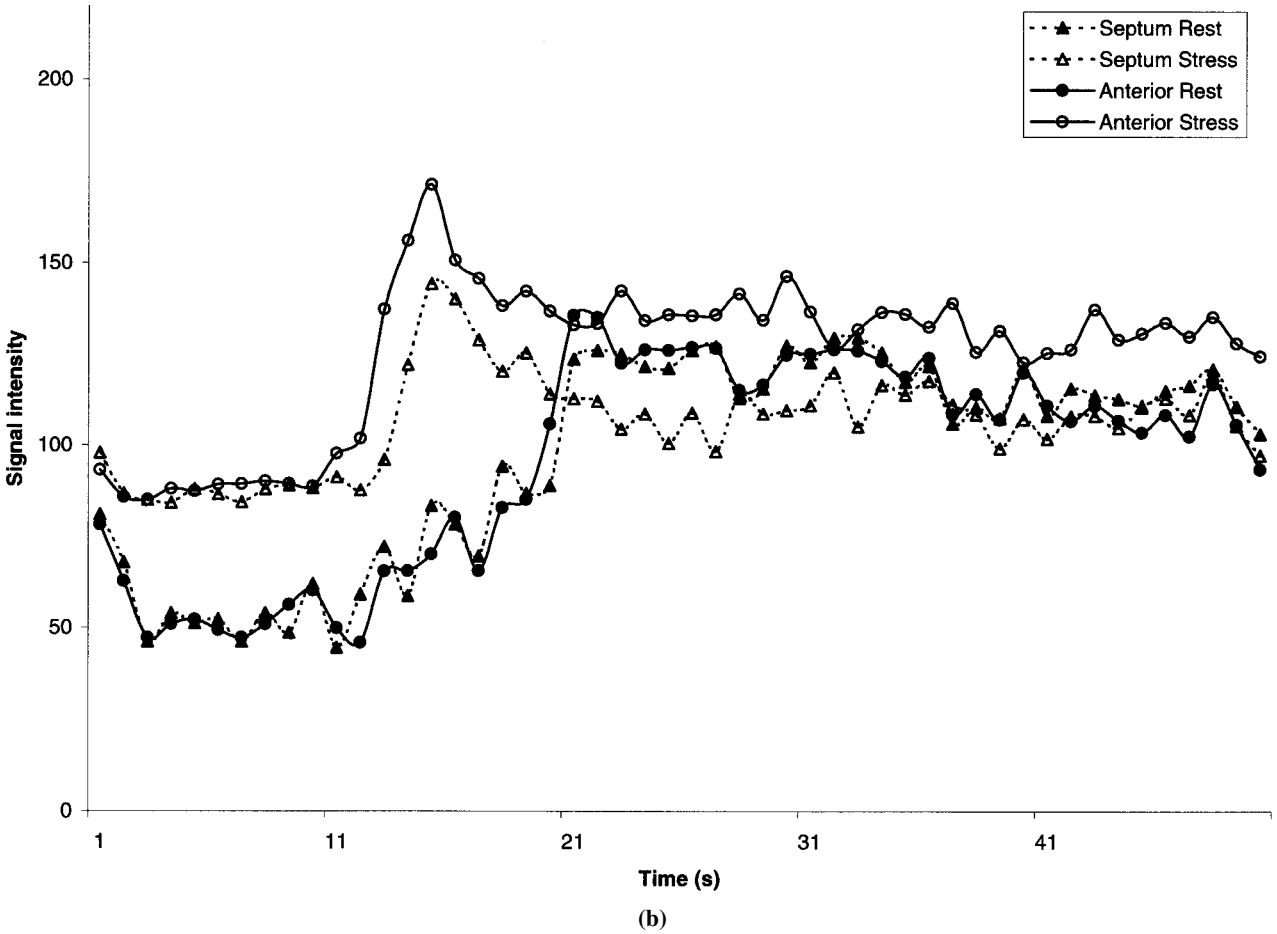


(a) (b) (c)

Figure 6. Patient 3. (a) Transverse segmented gradient echo fast low-angle shot CMR image. A single coronary artery is visible arising from the right sinus of Valsalva (arrows). (b) The first branch of this vessel forms the left coronary artery that passes posteriorly and leftward between the aorta and right ventricular outflow tract (arrows). (c) With a double oblique image the anomalous left coronary artery is visible passing posteriorly toward the interventricular sulcus. (This anatomic arrangement would be classified as type RIIB according to Lipton et al. [26].)



(a)



(b)

Figure 7. Patient 3. (a) Perfusion gradient echo image of the basal short-axis slice of the left ventricle acquired after adenosine stress. There is reduction in myocardial signal intensity in the septum and anteroseptal wall (arrows). (b) Signal intensity curves from the septum and anterior walls confirm the delayed and reduced enhancement in the septum.

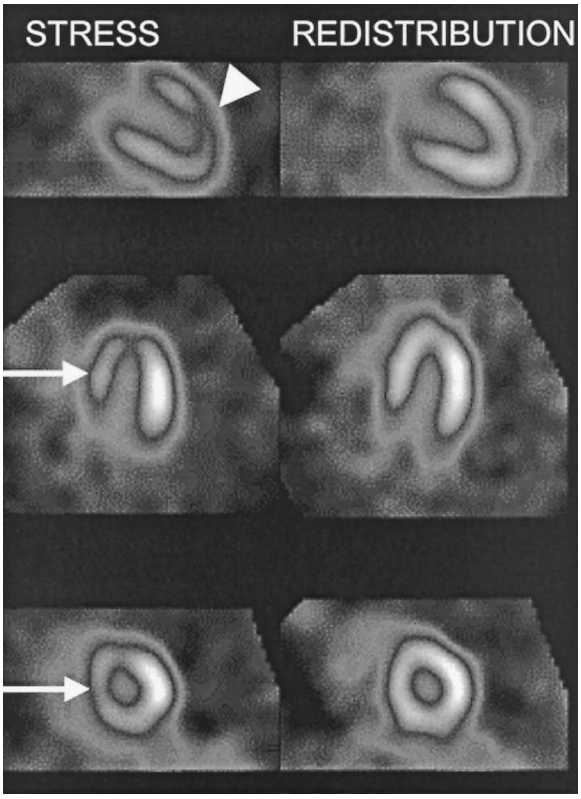


Figure 8. Patient 3. Myocardial perfusion SPECT image acquired after adenosine stress. There are perfusion defects in the anterior wall (arrowhead) and septum (arrows) that return to normal for the rest acquisition.

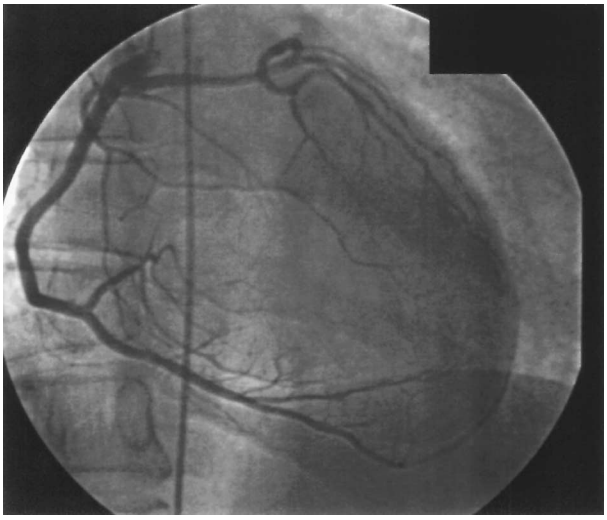


Figure 9. Patient 3. Cardiac catheterization. Selective cannulation in the right anterior sinus of Valsalva opacifies the single coronary artery. The right coronary artery has normal anatomy. The left coronary artery is anomalous, but the anatomic relationship between the left coronary artery and the aorta and right ventricular outflow tract is not evident.

den cardiac death have included a slit-like orifice of the anomalous vessel (22) that is narrowed when the aorta expands during exercise (4), an acute angled intramural segment of the proximal anomalous coronary artery, or compression of the anomalous vessel between the high pressure aorta and right ventricular outflow tract again during exercise (23).

In this study we used an adenosine infusion to provide pharmacologic stress, which causes coronary artery vasodilation (24) but produces only small alterations in heart rate and blood pressure. On first-pass adenosine myocardial perfusion imaging, both anomalous coronary arteries produced delayed signal intensity increases in the myocardium supplied by the vessel. One possible explanation would be that the anomalous coronary artery is unable to vasodilate to the same degree as the normal vessel, causing a coronary steal phenomenon with blood preferentially distributed to regions supplied by normal arteries. Using a Doppler probe in an anomalous intraarterial right coronary artery, Brandt et al. (25) demonstrated abnormal vasodilator reserve in response to temporary vessel occlusion. Assuming that the coronary artery vascular structure is normal, then the steal phenomenon may result from external compression of the vessel as it passes between the aorta and right ventricular outflow tract.

Although the mechanism of ischemia associated with anomalous coronary arteries is uncertain, we have shown that the combined CMR approach of three-dimensional anatomic localization and first-pass myocardial perfusion can be safely performed in these patients, and in these patients correctly identified perfusion defects as compared with SPECT. The combined approach of anatomic and functional perfusion information has the potential to fully evaluate these patients in a single examination, but further studies with larger numbers of patients are necessary to confirm the utility of this approach.

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