Magnetic Resonance Angiography of Thoracic Aortic Atherosclerosis in Homozygous Familial Hypercholesterolemia

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Purpose
To assess the ability of MR angiography with virtual angioscopy reconstruction to detect global atherosclerotic changes in the thoracic aorta of patients with homozygous familial hypercholesterolemia (HFH).

Methods
Fifteen HFH patients (7 male and 8 female, mean age 24 years) and 9 normal volunteers (2 male and 7 female, mean age 34 years) were scanned on a 1.5 T G.E. Signa MR unit.

Oblique sagittal MR angiograms of the thoracic aorta were obtained using contrast-enhanced three-dimensional gradient echo with a phased-array torso surface coil. Surface renderings and virtual angioscopy reconstructions (perspective endoluminal projections) were produced from the MRA data (1).

Abnormalities of the aorta and great vessels were assessed from surface renderings and virtual angioscopy by three radiologists blinded to the clinical history. Wall irregularity was scored on a scale of 0 (normal) to 4 (abnormal). Scores of 2 or more (average readings for the three radiologists) were considered to be indicative of pathology. The incremental value of virtual angioscopic views over and above the surface renderings was also assessed on a 5-point scale (0, lowest; 4, highest). Results were compared to contrast angiograms which were available for all 15 patients. Sensitivity and specificity were computed by dividing the aorta into 3 territories (ascending, arch, and descending). Averages are expressed as mean±standard deviation.

Results
Surface renderings of the thoracic aorta are shown for both a normal volunteer and a patient with HFH in Figure 1. The normal aortic wall is smooth. The patient’s aortic wall is irregular and there are stenoses of the origins of the great vessels. Corresponding virtual angioscopic views provide more detail of the surface irregularity (Figure 2).

Aortic wall irregularity was detected in 8 of 15 (53%) patient scans at MRA and in 13 of 15 (87%) patients at angiography. The mean of the aortic wall irregularity scores was 2.0±0.9. The sensitivity and specificity of MRA were 65% and 32%, respectively.

Vessel wall irregularity was also detected in 1 of 9 (11%) normal subject scans at MRA. Signal to noise measurements revealed a greater level of noise in this subject’s aorta compared to the other normal aortae, which suggests that the irregularity of the aorta was due to noise. The mean of the wall irregularity scores for the normal aortae was 1.0±0.6 which is less than that of the patients (p=0.008).

The observers found that virtual endoscopy views were of some, although limited value (value score 1.8±1.0). Virtual endoscopy views were of greatest value for evaluation of the descending thoracic aorta (value score 2.1±0.8).

Discussion
Homozygous familial hypercholesterolemia (HFH) is an autosomal dominant disorder of the LDL receptor that predisposes patients to marked elevations in serum cholesterol. Because of this defect, HFH patients develop atherosclerosis at an early age and are at risk for stroke, myocardial infarction, and sudden death as early as the first decade of life(2,3). Physicians who take care of HFH patients need to know the extent of their patient’s atherosclerotic disease in order to guide treatment decisions and to accurately assess the risk of morbidity.

Our results demonstrate that wall irregularity of the thoracic aorta, a potential gauge of global atherosclerosis, is found frequently in HFH patients and not in normal volunteers at MR angiography (53% versus 11%). We also found that virtual angioscopy is occasionally a useful adjunct display method for evaluation of wall irregularity.

Wall irregularity was found more frequently at angiography than at MRA (87% versus 53%). One possible explanation for this difference is that subtle wall irregularity is more readily detected at conventional angiography than at MRA. This hypothesis is supported by the relatively low sensitivity of MRA (65%).

The significance of our results is that global atherosclerosis measures, which are difficult in general to obtain, may be feasible using MRA with virtual angioscopy.

References