Bicuspid Aortic Valve Disease and Ascending Aorta Aneurysm
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Clinical History
An asymptomatic 58-year-old woman with a history of hypertension, hyperlipidemia and a bicuspid aortic valve (BAV) with an aneurysmal ascending aorta was referred for a trans-thoracic ultrasound evaluation (TTE). The TTE findings included a BAV with partial fusion of the right and left coronary cusps, with mild insufficiency and stenosis. Her ascending aorta was markedly dilated from the root to the arch, measuring a maximum 50 mm above the sinotubular junction, and was consistent with progressive enlargement when compared to a study performed 4 years earlier. Composite aortic root replacement was recommended, and a cardiac CTA was requested as part of her preoperative workup.

Findings
Cardiac CTA showed a right dominant circulation without evidence of coronary artery disease (Figure 1). A BAV with a false raphe between the right and left coronary cusps was seen (Figure 2). CTA confirmed the presence of a large aortic aneurysm that extended from the sinotubular junction to the proximal aortic arch (Figure 3). Its largest diameter was 51x49 mm, approximately 2 cm above the sinotubular junction (Figure 4).

Discussion
BAV is the most common congenital heart disease, with an estimated prevalence of 1-2% in the general population and a male-to-female ratio of 3:1. The pattern of inheritance of BAV is autosomal dominant with incomplete penetrance. In 47% of cases, the anterior cusp contains the ostia of both coronary arteries as well as a false raphe (partial fusion of the right and left coronary cusps). BAV is part of a larger spectrum of structural and developmental abnormalities involving the great vessels, and aortic dilatation is observed in 56% of those aged <30 years old and up to 88% of those aged >80 years old. Current guidelines recommend surgical intervention for patients with a >50 mm dilatation of the ascending aorta, even if they are asymptomatic and have no other indications for aortic valve surgery. Our patient underwent a valve-sparing aortic root replacement, and her post-operative course was unremarkable. Due to its wide field of view, excellent spatial resolution and ability to exclude significant obstructive epicardial coronary artery disease, cardiac CTA is exceedingly helpful in the preoperative workup of low- to intermediate-risk patients with diseases of the thoracic aorta.

REFERENCES
1. Marion DW. Clinical manifestations and diagnosis of bicuspid aortic valve. In: UpToDate, Basow, DS (Ed), UpToDate, Waltham, MA, 2013