A 46-year-old man with well-controlled hypertension presented with dyspnea, palpitations, and a murmur of aortic regurgitation. Echocardiography revealed a quadricuspid aortic valve (QAV) with 4 equal-sized cusps and enlarged left main coronary artery ostium (Figure 1A). On additional imaging, an unusual flow was seen entering the proximal main pulmonary artery from the lateral aspect just distal to the pulmonic valve (Figure 2A and Movie I in the online-only Data Supplement). A QAV with moderate aortic regurgitation (Movie II in the online-only Data Supplement) and a coronary-pulmonary artery fistula were diagnosed by echocardiography. Computed tomography confirmed the presence of QAV (Figure 1B) and a small-caliber vessel that originated from the left main coronary artery, coursed superiorly (Figure 2B), and ended in a larger confluence of anomalous vessels that communicated with the pulmonary artery (Figure 2C). Other vessels contributing to this confluence originated from the descending aorta and followed a very tortuous course (Figure 2D1 through 2D3). A 3-dimensional reconstruction shown in Figure 2E demonstrates the descending aorta-to-pulmonary artery fistula and coronary artery-to-pulmonary artery fistula draining into a large confluence. Thus, a unique anomaly in combination with QAV was diagnosed.

QAV is a rare malformation, with an estimated incidence of 0.003% to 0.043% of all congenital heart disease.1,2 It has been associated with other congenital malformations, the most common being coronary artery anomalies,3 including coronary-pulmonary artery fistula.4 However, QAV associated with the unusual systemic-pulmonary artery fistulae seen in our patient is unique. It is important to delineate any vascular anomalies associated with QAV preoperatively to prevent serious complications.

Acknowledgments
We gratefully acknowledge Joe Grundle and Katie Klein for editorial preparation of this manuscript and Brian Miller and Brian Schurrer for their help in preparing the figures.

Disclosures
None.

References
Figure 1. Demonstration of the quadricuspid aortic valve on echocardiography (A) and computed tomography (B). LMCA indicates left main coronary artery.

Figure 2. Demonstration of coronary artery-to-pulmonary artery and descending aorta-to-pulmonary artery fistulae. A, Echocardiography showing unusual flow entering the proximal main pulmonary artery (arrow). B, Computed tomography showing a small-caliber fistula (arrowhead) originating from the left main coronary artery. C, Magnified computed tomography showing a large confluence of anomalous vessels (asterisk) communicating with the pulmonary artery (arrow). D1–D3, Multiple computed tomography images demonstrating the tortuous course traversed by the descending aorta-pulmonary artery fistula (arrows). E, A 3-dimensional reconstruction illustrating the descending aorta-pulmonary artery fistula (arrows), coronary-pulmonary artery fistula (arrowheads), and large confluence of anomalous vessels (asterisk). AAo indicates ascending aorta; DAo, descending aorta; LA, left atrium; LMCA, left main coronary artery; and PA, pulmonary artery.
A Unique Case of Quadricuspid Aortic Valve With Coronary Artery and Descending Aorta-to-Pulmonary Artery Fistulae
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Circulation. 2013;128:1041-1042
doi: 10.1161/CIRCULATIONAHA.113.001441
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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