A 53-year-old man was admitted to our center with dyspnea and peripheral edema compatible with cardiac failure. His history was notable for Noonan syndrome, but he had not been seen a physician for 15 years. On examination, pectus excavatum, pectus carinatum, and low implantation of the ears were present (Figure 1). Blood pressure was 145/70 mm Hg, and pulse was irregular at 90 to 100 beats per minute. Cardiac auscultation showed a 4/6 holodiastolic murmur and Austin Flint rumble. ECG showed atrial fibrillation, an incomplete right bundle-branch block, and a left anterior fascicular block.

Echocardiography (Figure 2) showed a giant aneurysm of the Valsalva sinus (diameter, 9.8 cm) and a bicuspid aortic valve (Movies I and II in the online-only Data Supplement). A 64-slice computed tomography confirmed the diagnosis of a giant Valsalva sinus aneurysm and bicuspid aortic valve (Figure 3). Coronary angiogram showed a giant aneurysm of 2 Valsalva sinuses (Figure 4). The left coronary artery sinus had a normal location, although the right coronary ostium arose from the middle of the aneurysm (Movies III and IV in the online-only Data Supplement).

The patient was referred for a Bentall procedure, and the outcome was favorable. Noonan syndrome was first described by Noonan and Ehmke in 1963. Characteristic findings include distinctive facial features, short stature, chest deformity, and congenital heart disease. Aortic root dilatation has rarely been described, and a giant aneurysm, as in our case, has been described only once. Such dilatation is usually seen in Marfan syndrome. We conclude that patients with Noonan syndrome are at risk of having severe aortic root disease, and require careful cardiac monitoring during their entire life.

Sources of Funding
The work has been supported by the Fondation de Cardiologie (Fribourg, Switzerland).

Disclosures
None.

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The online-only Data Supplement is available with this article at http://circ.ahajournals.org/cgi/content/full/123/23/e629/DC1.

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(Circulation. 2011;123:e629-e630.)

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Circulation is available at http://circ.ahajournals.org

DOI: 10.1161/CIRCULATIONAHA.110.010025
Figure 3. Computed tomography axial view at the level of the aortic valve.

Figure 4. Supravalvular injection in right anterior oblique incidence showing the small aorta with the a Valsalva sinus.
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Circulation. 2011;123:e629-e630
doi: 10.1161/CIRCULATIONAHA.110.010025
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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