A 16-year-old girl experiencing palpitations underwent elective transthoracic echocardiographic examination at a nearby clinic, which showed a 3-cm balloon-like, cystic structure, with inhomogeneous content, adherent to the atrial septum and anchored above the anteroseptal commissure of the tricuspid valve without evidence of flow from the aorta to the right atrium (Figure 1A). On examination, blood pressure (120/70 mm Hg) and heart rate (70 bpm) were normal, but she was advised to limit physical activity. While waiting for scheduled MRI scan, she was admitted to the emergency department for acute vision loss and syncope. At physical examination, the patient presented dyspnea, hypotension (90/50 mm Hg), and tachycardia (130 bpm); a 5/6 continuous, rough murmur was heard over the left precordium. Chest x-ray revealed cardiomegaly and pulmonary congestion. To refine diagnosis of the cardiac mass, the girl underwent transesophageal echocardiographic examination, which demonstrated communication between the aortic root and the previously identified cystic mass, now ruptured in the right atrium, with severe right heart volume overload (Figure 1B and online-only Data Supplement Movie I; Figure 1C and online-only Data Supplement Movie II). To exclude particulate pulmonary embolism due to explosion of the cystic mass in the right atrium, computed tomography scan was performed, which unfortunately showed large endoluminal defect of right pulmonary artery branch. Because of severe tachycardia, however, anatomic definition of the aortoatrial fistula was poor. Final diagnosis was reached by aortography, which confirmed massive passage of contrast dye from the aortic root directly into the right atrium (online-only Data Supplement Movie III). Considering the hemodynamic impairment and the recent pulmonary embolism, the girl was referred for emergent repair. At surgery, severe overload of right heart chambers was found. After transection of the ascending aorta, a 5-mm-large defect of the noncoronary sinus was noted, close to the anterior aortic commissure (Figure 2A). After right atriotomy, a 3-cm-large, ruptured type 1, thin-walled aneurysm was identified (Figure 2B) reminiscent of a hot-air balloon explosion. The sac was empty and in direct communication with the noncoronary aortic sinus (Figure 2C). The aneurismal sac was thus excised (Figure 3), and the fistula was closed from the right atrial side by use of an autologous pericardial patch (Figure 4). Postrepair transesophageal echocardiographic examination demonstrated excellent coaptation of the aortic valve, without regurgitation, absence of intracardiac shunt, and prompt reduction of right-sided chambers volume. Postoperative course was uneventful with discharge on the fifth postoperative day.

Although aorto-right atrial fistulae secondary to endocarditis and aortic dissection are relatively common findings,2

Figure 1. A, Transthoracic echocardiogram, apical 4-chamber view: a round-shaped cystic lesion is visible in the right atrium (*), with inhomogeneous content (solid, liquid). B, Transesophageal echocardiogram, short axis of the aortic root: a communication between the aortic root and the right atrium through the ruptured aneurysmal sac is visible (*). C, Doppler signal of significant left-to-right shunt is present.

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congenital fistulae, due to rupture of sinus of Valsalva aneurysm, are rare anomalies more frequently found in Asian population. The case herein reported focuses on the importance of imaging to obtain accurate diagnosis and, consequently, to accommodate surgery. Echocardiography is considered the first choice to detect this lesion and transesophageal echocardiographic examination, or 3-dimensional echocardiographic scan, when possible, offers a better sensitivity and specificity. Advanced imaging such as MRI and computed tomography may allow definitive diagnosis in elective cases, whereas cardiac angiography may be required in case of hemodynamic instability to define surgical anatomy. When accurately and promptly recognized, surgical management of congenital aorto-right atrial fistula is a low-risk procedure with favorable long-term outcome. Because of the possibility of abrupt complications, patients with sinus of Valsalva aneurysm should be advised to limit physical activity and to undergo elective surgical repair.

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Disclosures
None.

References
Figure 4. Surgical anatomy: the pericardial patch closing the fistula is visible from the atrial side (left arrow) and aortic root side (right arrow).
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Movie Legend

Movie 1. Trans-esophageal echocardiogram, short axis of the aortic root: a communication between the aortic root and the right atrium through the ruptured aneurysmal sac is visible. Best viewed with Windows Media Player.

Movie 2. Trans-esophageal echocardiogram, short axis of the aortic root: Doppler signal of significant left-to-right shunt is present. Best viewed with Windows Media Player.

Movie 3. Left heart catheterization, right anterior oblique view: passage of contrast dye between the aortic root and the right atrium during systole is visible. Best viewed with Windows Media Player.