Surgical repair of a quadricuspid aortic valve associated with a ruptured sinus of Valsalva aneurysm

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A 35-year-old woman presented with a history of gradually worsening dyspnea on exertion. Clinical examination revealed a diastolic murmur heard loudest along the upper left sternal border, which did not radiate to the carotids or the precordium. Transthoracic echocardiography demonstrated a quadricuspid aortic valve (type A, Hurwitz and Robert’s classification) with an aorta-to-right atrium fistula (sinus 2) through a ruptured right-anterior sinus of Valsalva (Figure 1, A). In addition, the coronary orifices originated from sinus 1 (right coronary artery) and sinus 4 (left main coronary artery), and the aortic regurgitation was moderate (Figure 1, A and B). A diagnosis of a congenital quadricuspid aortic valve associated with ruptured sinus of Valsalva aneurysm was made, and the patient was referred for surgical correction. During the procedure, the aortic valve was confirmed to have 4 leaflets with associated sinuses, and a ruptured right-anterior sinus of Valsalva without coronary orifices was identified (Figure 2, A). The proximal windsock orifice was below the right-anterior sinus of Valsalva, and a right angle forceps could easily be passed through the aorta-to-right atrium fistula into the right atrium (Figure 2, B). The ruptured sinus of Valsalva...
aneurysm with an associated leaflet was resected. A 4-0 polypropylene suture was used to reconstruct the aortic root with 3 leaflets and associated sinuses. Postoperative transesophageal echocardiography showed normal aortic valvular function with no aortic insufficiency or residual shunt (Figure 3, A and B). The patient recovered well and was discharged a few days later, with no further complications.

FIGURE 2. A and B, During the procedure, the quadricuspid aortic valve was confirmed with 4 sinuses; a right angle forceps was used to identify the aorta-to-right atrium fistula. RSVA, right sinus of Valsalva aneurysm; AV, aortic valve; LCA, left coronary artery; RCA, right coronary artery.

FIGURE 3. A and B, Postoperative transesophageal echocardiogram showed a normal aortic valve with 3 cusps and no aortic insufficiency or residual shunt. AV, aortic valve; LV, left ventricle; AAO, ascending aorta; LA, left atrium.