0003-4975/$36.00 e275
https://doi.org/10.1016/j.athoracsur.2020.11.066

Acquired Sinus of Valsalva Fistula and Turner Syndrome

Sean A. Knudson, BSA, Erin A. Gottlieb, MD, Gregory Johnson, MD, Charles D. Fraser, MD, and Ziv Beckerman, MD

Texas Center for Pediatric and Congenital Heart Disease, UT Health Austin/Dell Children's Medical Center, Austin, Texas; and Department of Surgery and Perioperative Care, University of Texas Dell Medical School, Austin, Texas

A 21-year old woman with a history of Turner syndrome presented with a diastolic heart murmur, dizziness, dyspnea, and intermittent chest pain. Preoperative imaging revealed a fistula from the right sinus of Valsalva into the right atrium. Turner syndrome is associated with both aortopathy and congenital heart malformations. Acquired sinus of Valsalva fistula is a rare disorder, and this report describes its presence in association with Turner syndrome.

(Ann Thorac Surg 2021;112:e275-e277)

Acquired sinus of Valsalva fistula is a rare pathologic finding,[1] and it was first described in 1980 by Coto and colleagues.[2] The incidence of sinus of Valsalva fistula is currently unknown. A recent systematic review identified only 86 cases of aortic-right atrial fistula in the literature.[3] Turner syndrome affects 1 in 2500 female births.[4] We report a case of acquired sinus of Valsalva fistula in association with Turner syndrome.

Sinus of Valsalva fistula may originate from any of the aortic sinuses of Valsalva. The origin is multifactorial. Jainandunsing and colleagues[3] noted that 22.8% of cases were secondary to infective endocarditis, 15.4% were secondary to previous cardiac surgery, and 11.8% had a congenital origin.

Turner syndrome-associated aortic valve disease and thoracic aortopathy predisposes affected female patients to the development of aortic dilation, dissection, or fistulous communication. Approximately 1.4% of female patients with Turner syndrome will have aortic dissection in their lifetime, a 100-fold higher incidence than in women without Turner syndrome.[4,5] The genetic basis of the aortopathy seen in Turner syndrome remains under investigation.[6] Systemic arterial pressure in the weakened aortic root may cause an aortic-atrial tunnel to propagate into the adjacent lower-pressure right atrium.[7] We present the clinical and surgical management of a female patient with sinus of Valsalva fistula and Turner syndrome.

A 21-year-old woman with a history of Turner syndrome, bicuspid aortic valve, and critical coarctation of the aorta after aortic coarctation repair (in newborn life) was seen with symptoms of dyspnea, dizziness, and intermittent chest pain. She was noted to have a diastolic cardiac murmur. Echocardiography demonstrated left-to-right shunting into the right atrium with a measured velocity of 2.9 m/s during diastole, suggestive of sinus of Valsalva fistula. The ascending aorta measured 29.0 mm in diameter, whereas the aortic root measured 23.7 mm. Cardiac magnetic resonance imaging confirmed a fistula from the right sinus of Valsalva into the right atrium (Figures 1A, 1B), bicuspid aortic valve with mild aortic valve stenosis, and aortic insufficiency with a regurgitant fraction of 18.3%. The ascending aorta was dilated with a maximal diameter of 30.4 mm, giving an aortic index of 2.4 cm/m^2. The aortic root was normal with a diameter of 23.0 mm (Z + 0.1). There was a good correlation between magnetic resonance imaging and echocardiographic measurements of the ascending aorta and aortic root. The aortic arch was tortuous, with slight narrowing at the site of previous coarctation repair. Right and left ventricular dimensions and function were normal.

Given the clinical and imaging findings, the decision was made to proceed with surgery for fistula repair (Video 1). The operation was performed through a median sternotomy. After the chest was opened, massively dilated and congested lymphatic vessels were observed along the mediastinum and the heart, some as wide as 4 mm (Figure 2A). Cardiopulmonary bypass was instituted using bicaval and aortic cannulation, the patient was cooled to 32°C, the ascending aorta was cross-clamped, and the heart was arrested with cold crystalloid cardioplegia.

Right atriotomy revealed a sinus of Valsalva fistula with a long windsock forming the right atrial aspect of the fistula (Figure 2B). An aortotomy was performed, and the aortic root was inspected. It was not aneurysmal or dilated. The bicuspid aortic valve was inspected, and

The Videos can be viewed in the online version of this article [https://doi.org/10.1016/j.athoracsur.2020.11.066] on http://www.annalsthoracicsurgery.org.

Accepted for publication Nov 9, 2020.

Accepted for Presentation as an ePoster at the Sixty-seventh Annual Meeting of the Southern Thoracic Surgical Association, Orlando, FL, Nov 4-7, 2020 [canceled].

Address ccorrespondence to Dr Beckerman, Dell Children's Medical Center, 900 Mueller Blvd, Ste 3S.003, Austin, TX 78723; email: zbeckerman@austin.utexas.edu.

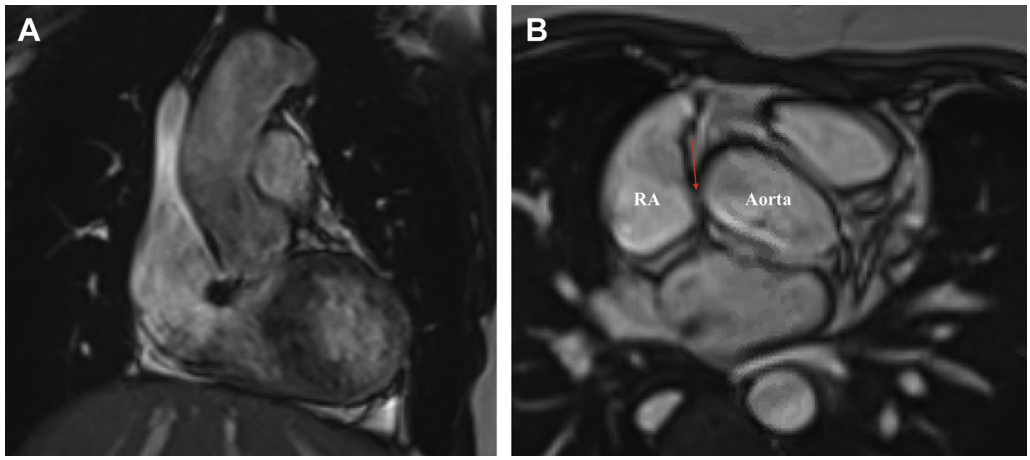

FIGURE 1 Preoperative cardiac magnetic resonance imaging. (A) Coronal view of acquired right coronary sinus of Valsalva. (B) Axial view of aortic–right atrial (RA) communication (arrow).

it was not significantly stenotic. A 4- to 5-mm diameter fistula was confirmed originating from the right coronary cusp base and extending anteriorly into the right atrium (Video 2).

The fistula was repaired with interrupted pericardial pledget reinforced polypropylene sutures from the aortic root side (Figure 2C), as well as with interrupted nonpledgeted sutures from the right atrial side (Figure 2D). Aortic cross-clamp time was 63 minutes, and cardiopulmonary bypass time was 100 minutes. Preoperative and postoperative short-axis transesophageal echocardiography clips demonstrating the fistula before and after the repair are seen in Video 3. The patient tolerated the procedure well, was extubated on postoperative day 1, and was discharged home 5 days later. Echocardiography and computed

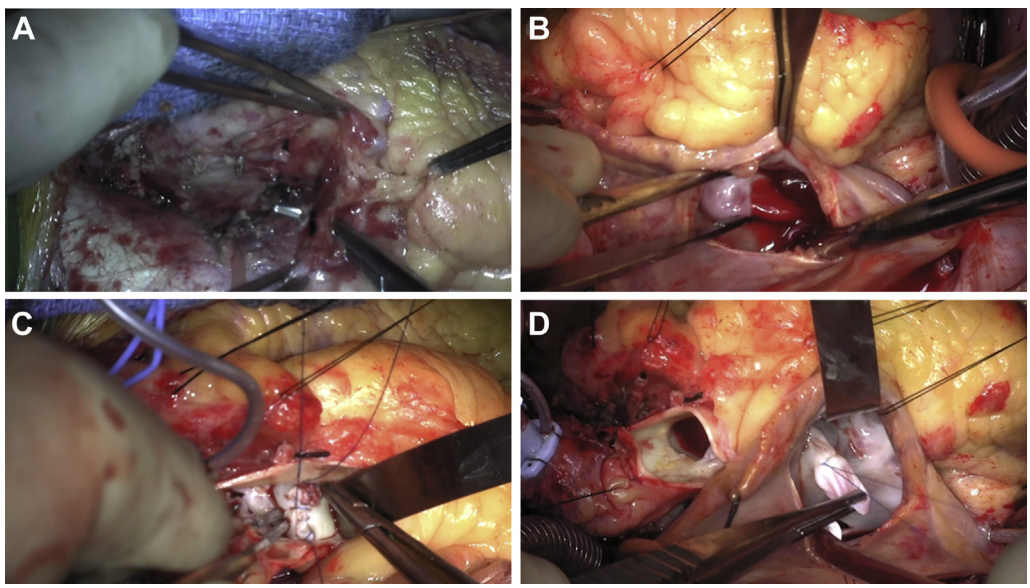

FIGURE 2 Intraoperative imaging of sinus of Valsalva fistula repair. (A) Intraoperative view of massively congested lymphatic vessels. (B) Right atrial aspect of ruptured sinus of Valsalva fistula windsock during instillation of cardioplegia to the aortic root. (C) Aortic aspect sinus of Valsalva repair using interrupted pericardial pledged reinforced polypropylene sutures. (D) Primary repair of the right atrial aspect of the windsock.

tomographic angiography before discharge and at 3-month follow-up showed a good result with no residual shunting.

COMMENT

Acquired sinus of Valsalva fistula is a rare disorder, and in our patient it occurred in association with Turner syndrome. The combination requires complex decision making because of the potential need to address the associated valve disease and aortopathy associated with Turner syndrome.

Sinus of Valsalva fistula typically requires surgical intervention, especially when it is associated with heart failure. Should the aortic root or ascending aorta be prophylactically replaced on surgical intervention for sinus of Valsalva fistula repair in a patient with Turner syndrome? This is a complex question that needs further investigation, although, as we previously noted, sinus of Valsalva fistula in Turner syndrome appears to be a rare entity. We elected to preserve the aortic root and undertake primary closure of the fistula. Clinical reasoning was derived from the findings that our patient's aortic root was not dilated, the bicuspid aortic valve did not require intervention, and the tissue quality of the fistula rims seemed adequate for such a repair. The ascending aorta was dilated but did not meet criteria for replacement because our patient's preoperative indexed ascending aortic diameter was 2.4 cm/m^2. Current recommendations call for prophylactic aortic replacement in patients with Turner syndrome with an aortic index greater than 2.5 cm/m^2.[8] Because the fistula formed in the setting of a likely pathologically abnormal aortic wall, the decision not to replace the aortic root or ascending aorta is open to discussion. It is likely that the patient will require further surgical intervention in the future to address progressive aortic disease.

The results of short-term follow-up have been satisfactory. Our patient will require ongoing follow-up for radiographic and echocardiographic surveillance and will be treated with aggressive aortopathy prophylaxis. Further research into the exact pathologic nature and genetic causes of aortopathy in Turner syndrome is ongoing and will hopefully provide additional insight for the aortic surveillance and surgical management in these patients.

REFERENCES

1. Sai Krishna C, Baruah DK, Reddy GV, Panigrahi NK, Suman K, Kumar PV. Aorta-right atrial tunnel. *Tex Heart Inst J*. 2010;37:480-482.

2. Coto EO, Caffarena JM, Such M, Marques JL. Aorta–right atrial communication. Report of an unusual case. *J Thorac Cardiovasc Surg*. 1980;80:941-944.

3. Jainandunsing JS, Linnemann R, Bouma W, et al. Aorto-atrial fistula formation and closure: a systematic review. *J Thorac Dis*. 2019;11:1031-1046.

4. Thunström S, Krantz E, Thunström E, Hanson C, Bryman I, Landin-Wilhelmsen K. Incidence of aortic dissection in Turner syndrome. *Circulation*. 2019;139:2802-2804.

5. Gravholt CH, Landin-Wilhelmsen K, Stochholm K, et al. Clinical and epidemiological description of aortic dissection in Turner's syndrome. *Cardiol Young*. 2006;16:430-436.

6. Corbitt H, Gutierrez J, Silberbach M, Maslen CL. The genetic basis of Turner syndrome aortopathy. *Am J Med Genet C Semin Med Genet*. 2019;181:117-125.

7. Gajjar T, Voleti C, Matta R, Iyer R, Dash PK, Desai N. Aorta-right atrial tunnel: clinical presentation, diagnostic criteria, and surgical options. *J Thorac Cardiovasc Surg*. 2005;130:1287-1292.

8. Carlson M, Airhart N, Lopez L, Silberbach M. Moderate aortic enlargement and bicuspid aortic valve are associated with aortic dissection in Turner syndrome: report of the international turner syndrome aortic dissection registry. *Circulation*. 2012;126:2220-2226.