Case Reports

Right Ventricular Outflow Tract Obstruction Caused by Double-Chambered Right Ventricle

Presenting in Adulthood

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Congenital heart diseases that cause obstruction of the right ventricular outflow tract are often difficult to diagnose. We report the case of a 49-year-old man who presented with long-standing shortness of breath on exertion. Imaging revealed right ventricular outflow tract obstruction caused by a double-chambered right ventricle, and he was referred for surgical correction. This case emphasizes both the detailed perioperative evaluation that is needed when diagnosing adults who present with manifestations of congenital heart disease and a method of successful surgical correction that resulted in symptom resolution. ***(Tex Heart Inst J 2014;41(4):425-8)***

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Congenital heart diseases that cause obstruction of the right ventricular outflow tract (RVOT) are difficult to diagnose precisely, especially in older patients.[1] Obstruction of the RVOT can be the result of abnormalities at the mid-right ventricle (RV), the infundibulum, the pulmonary valve, the supravalvular region, or at the branch or peripheral pulmonary arteries. These lesions are usually congenital, but can be iatrogenic as a result of previous cardiac surgery.[1,2]

Right-sided aortic arch is a congenital anomaly that affects approximately 0.1% of the general population.[3] Right-sided aortic arch results from involution of part of the left dorsal aorta and persistence of the right dorsal aorta. In situs solitus, right-sided aortic arch is typically associated with cardiac malformations in the outflow tract, as found in cases of tetralogy of Fallot (TOF), pulmonary atresia, or common arterial trunk.[3]

We report a case of a middle-aged man with isolated RVOT obstruction caused by double-chambered right ventricle.

Case Report

In August 2012, a 49-year-old man presented at the Evangelismos General Hospital with long-standing shortness of breath on exertion (New York Heart Association functional class II). His medical history included hypertension and dyslipidemia, both controlled with medication. Three years previously, he had undergone endovascular stent-grafting of the lower descending thoracic aorta for type B aortic dissection. At that time, he was also found to have a right-sided aortic arch with an aberrant left subclavian artery. His childhood records were said to have reported a supracristal ventricular septal defect (VSD), but this had not been confirmed. The patient had a 20-pack-year history of smoking but had stopped smoking 3 years before. He had a family history of coronary artery disease.

On admission, the patient was in sinus rhythm with a blood pressure of 120/75 mmHg. His electrocardiogram displayed negative T-wave deflection in leads II, III, and aVF, and in precordial leads V_1 through V_5, together with signs of RV overload. Chest radiographs revealed the endovascular stent-graft in the descending aorta, but no signs of pulmonary dilation.

Both transthoracic (TTE) and transesophageal (TEE) echocardiography revealed diffuse RV hypertrophy and severe obstruction at the RV infundibulum (Fig. 1); however, there were no signs of right-sided heart failure. The left ventricle was

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mildly hypertrophic, with normal systolic function. The pulmonary valve appeared to be thickened and mildly restricted upon opening, but the pulmonary artery and its branches were normal. The false lumen in the descending aorta was thrombosed, with no residual flow.

Coronary angiography revealed several anomalies. The left coronary system comprised 2 arteries that arose from separate ostia above the left coronary sinus. The first ran a course consistent with the diagonal coronary artery and the 2nd ran a course consistent with the circumflex coronary artery (Fig. 2). The right coronary artery appeared normal but gave rise to a proximal branch that coursed between the aortic root and the RVOT, then continued as an anterior descending coronary artery (but from the right) (Fig. 2). Ventriculography revealed hypertrophy of both left and right ventricles, with preserved systolic function. Severe RVOT obstruction was seen at the infundibulum (Fig. 3). Significant RV hypertension was also observed: the systolic pressure gradient over the obstruction was 127 mmHg.

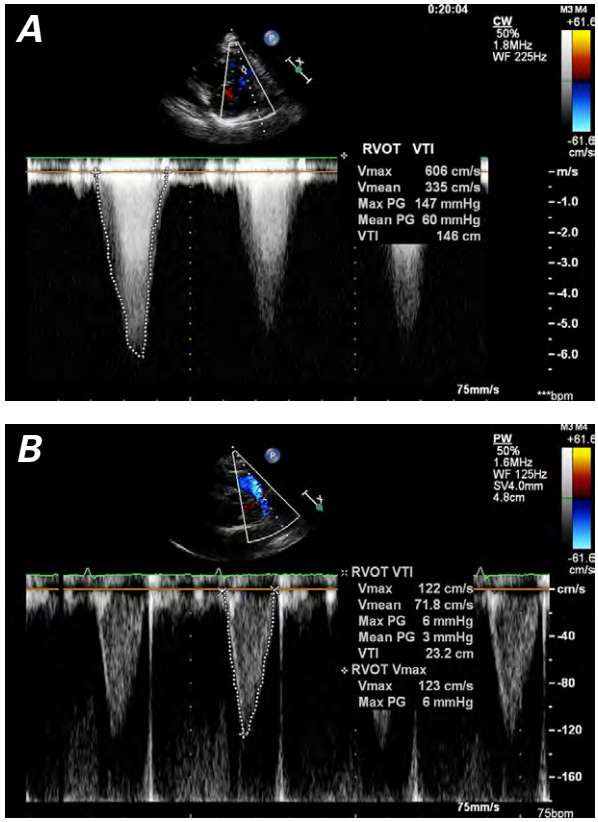

Fig. 1 *Transthoracic echocardiograms show* ***A****) severe preoperative right ventricular outflow tract (RVOT) obstruction (RVOT Vmax=6 m/s) and* ***B****) the postoperative peak right ventricular gradient of 6 mmHg (mean gradient, 3 mmHg).*

Max PG = maximum pressure gradient; mean PG = mean pressure gradient; Vmax = maximum velocity; Vmean = mean velocity; VTI = velocity time integral

To further characterize the nature and extent of the obstruction, we ordered cardiac magnetic resonance imaging. A finding of RVOT obstruction was confirmed at the infundibulum, with a diameter of <1 cm and a peak velocity of >550 cm/s at systole (Fig. 4).

In light of these findings (especially that of RVOT obstruction), and the patient's worsening symptoms, we referred him for surgical correction.

Surgical Procedure

The surgical approach was via median sternotomy. After systemic heparinization, cardiopulmonary bypass (CPB) was instituted via central aortic and bicaval cannulation. The patient was cooled to 32 °C and a vent was inserted through the right superior pulmonary vein. After aortic cross-clamping, myocardial arrest was achieved through antegrade administration of cold crystalloid cardioplegic solution.

The ventricular septum and RVOT were exposed via an oblique right atriotomy and retraction of the septal leaflet of the tricuspid valve. There was no septal defect. Anomalous muscle bundles were seen at the entrance of the infundibulum, creating an ostium of approximately 1 cm (internal diameter). These anomalous bundles were carefully resected in a circumferential fashion, but adequate exposure necessitated a transverse right ventriculotomy 1 cm below the anomalous right coronary artery. The pulmonary valve and its annulus were evaluated with Hegar dilators, through the ventricular incision. Retraction of the inferior margin of that incision enabled additional resection after we identified and protected the papillary muscles of the tricuspid valve. The right ventriculotomy was then closed with a pericardial patch and running 5-0 polypropylene suture; the right atriotomy was subsequently closed.

Atrial and ventricular pacing wires were placed; the patient was rewarmed and successfully weaned from CPB. After the procedure, we saw a significant drop in peak RV pressure gradient, to 15 mmHg. The patient's postoperative course was uneventful.

At the 6-month follow-up evaluation, TTE showed significant resolution of the preoperative RV gradient, with a peak RV gradient of 6 mmHg, a mean gradient of 3 mmHg, and significant regression of RV hypertrophy (Fig. 1).

Discussion

Congenital conditions that result in RVOT obstruction are often challenging to diagnose in adults because of difficulties in evaluating the RVOT.[1,4] Early differential diagnoses in this case included TOF, hypertrophic cardiomyopathy (HCM) causing RVOT obstruction, and double-chambered RV (DCRV). Although uncorrected TOF is rare in adults, a misdiagnosed pink tetralogy was considered in view of the possibility that

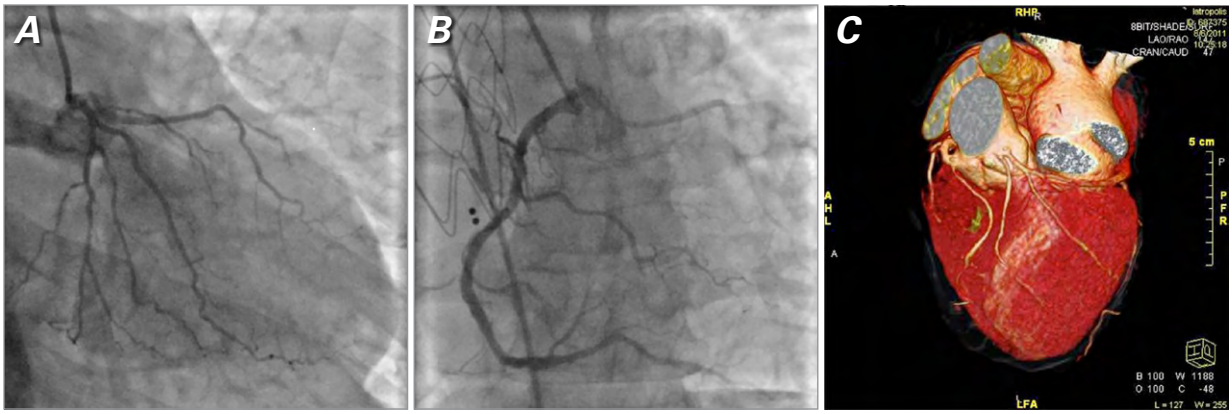

Fig. 2 *Coronary angiograms of the **A**) left and **B**) right coronary systems. **C**) Computed tomographic angiogram shows the 2 left coronary arteries arising from separate ostia above the left coronary sinus.*

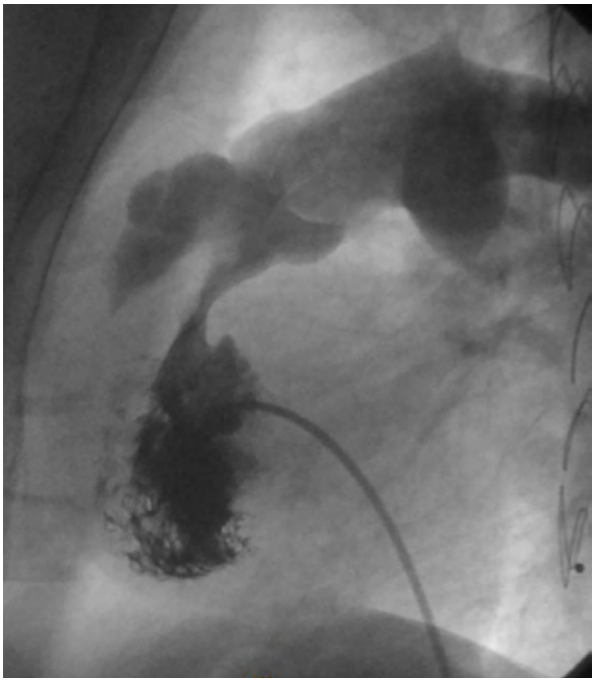

Fig. 3 *Right ventriculogram shows severe right ventricular outflow tract obstruction at the infundibulum.*

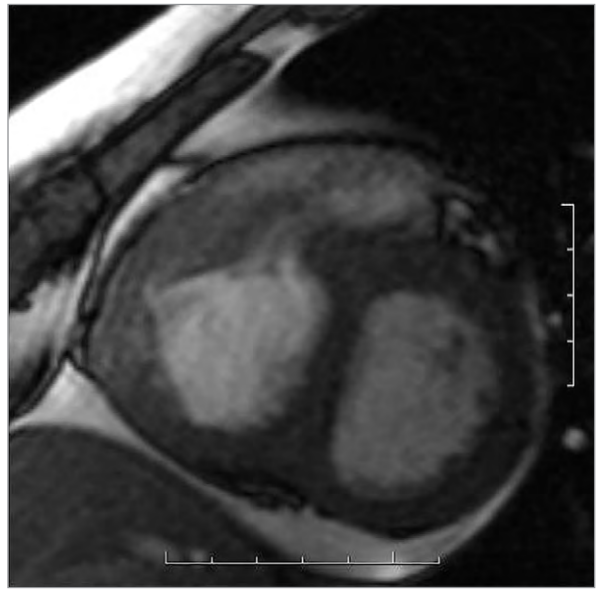

Fig. 4 *Magnetic resonance image confirms the presence of severe right ventricular outflow tract obstruction.*

a small VSD had spontaneously closed.[5] However, echocardiography confirmed an intact ventricular septum and no overriding aorta. Similarly, biventricular HCM (although usually considered a disease of the left ventricle) can produce RVOT obstruction by a variety of mechanisms, such as hypertrophied crista supraventricularis, moderator bands, and trabeculae. However, such reports are usually associated with the diagnosis of Noonan syndrome and other congenital features not observed in this patient.[6]

Double-Chambered Right Ventricle

Double-chambered RV is a rare congenital anomaly usually diagnosed and treated during childhood. The condition is caused by anomalous muscle bands that divide the RV into high-pressure inlet and low-pressure outlet sections, resulting in RVOT obstruction. This RVOT obstruction is generally believed to be acquired and progressive over time, although the basic anatomic features are congenital. Double-chambered RV is usually associated with a perimembranous VSD (~75% of cases) that is usually below the level of the muscular obstruction.[2] The flow and pressure from this VSD subsequently stimulate hypertrophy of the RV muscle bundles in order to protect the pulmonary circulation, which over time results in DCRV. In our patient, the site of obstruction was at the infundibulum, and although no VSD was present at the time of presentation, it is likely that such a lesion had closed spontaneously.

When double-chambered RV presents in adulthood, it often presents with nonspecific symptoms—a reduc-

tion in exercise tolerance, shortness of breath, syncope, or angina[1]—and is then misdiagnosed. An initial investigation with TTE often cannot ascertain the morphology of the RVOT because of the retrosternal nature of the RV and difficulty in acquiring subcostal images.[4,7,8] In fact, TTE has been reported as diagnostic in as few as 14% to 15% of patients with DCRV.[7,8] However, TEE has been shown to provide improved diagnostic accuracy and estimation of the RV inflow–outflow pressure gradient, particularly when using a combination of monoplane (deep gastric window) and omniplane approaches.[7] Despite this, right-sided cardiac catheterization or magnetic resonance scanning is often needed to confirm the diagnosis, as was done in this patient. Magnetic resonance imaging is multiplanar, functional, and dynamic, and able to disclose both RV hypertrophy and turbulent flow through the muscle bands that divide the RVOT.[9,10] Right-sided heart catheterization also has better sensitivity for detection of DCRV than does TTE and is therefore a useful adjunct to echocardiography in confirming the diagnosis of DCRV in adults. Furthermore, coronary angiography performed in the same sitting might identify coexisting coronary anomalies that would affect the choice of surgical approach and subsequent management: findings that were invaluable in this case. However, because anomalous muscle bands can be difficult to detect, careful pressure recordings should be taken during pullback across the RV, to ensure adequate recording of the pressure gradient between the inlet and outlet chambers.[8]

In conclusion, we have described a rare case of isolated RVOT obstruction caused by double-chambered right ventricle, presenting in a 49-year-old man. Detailed study of the RVOT by means of multidisciplinary imaging was fundamental to precise diagnosis before surgical correction. This case emphasizes both the detailed perioperative evaluation that is needed when diagnosing adults who present with manifestations of congenital disease, and a method of successful surgical correction that resolves symptoms.

References

1. Takigawa M, Nakatani S, Hashimura K, Kagisaki K, Kanzaki S, Arakaki Y, et al. An elderly case of right ventricular outflow obstruction precisely diagnosed and successfully treated with surgical repair. Intern Med 2010;49(6):563-7.
2. Bashore TM. Adult congenital heart disease: right ventricular outflow tract lesions. Circulation 2007;115(14):1933-47.
3. Craatz S, Kunzel E, Spanel-Borowski K. Right-sided aortic arch and tetralogy of Fallot in humans--a morphological study of 10 cases. Cardiovasc Pathol 2003;12(4):226-32.
4. Yalcinkaya E, Bugan B, Celik M, Yuksel UC. Evaluation of right ventricle with echocardiography: utility of combined techniques. Ann Thorac Med 2013;8(2):127.
5. Siriapisith T, Wasinrat J, Tresukosol D. Uncorrected pink tetralogy of Fallot in an adult patient: incidental CT findings. J Cardiovasc Comput Tomogr 2010;4(1):58-61.
6. Hayashi S, Tojyo K, Uchikawa S, Momose T, Misawa T, Yazaki Y, et al. Biventricular hypertrophic cardiomyopathy with right ventricular outflow tract obstruction associated with Noonan syndrome in an adult. Jpn Circ J 2001;65(2):132-5.
7. Hoffman P, Wojcik AW, Rozanski J, Siudalska H, Jakubowska E, Wlodarska EK, Kowalski M. The role of echocardiography in diagnosing double chambered right ventricle in adults. Heart 2004;90(7):789-93.
8. Lascano ME, Schaad MS, Moodie DS, Murphy D Jr. Difficulty in diagnosing double-chambered right ventricle in adults. Am J Cardiol 2001;88(7):816-9.
9. Kardux JJ, van Es HW, Rensing BJ. Case 119: double-chambered right ventricle. Radiology 2007;244(2):617-9.
10. Ibrahim T, Dennig K, Schwaiger M, Schomig A. Images in cardiovascular medicine. Assessment of double chamber right ventricle by magnetic resonance imaging. Circulation 2002;105(22):2692-3.