sewn to the ascending aorta just distal to the origin of the right common carotid artery. The recurrent laryngeal nerve was identified and spared. Flow was reestablished to the heart and cranial vessels through a 10-mm side arm in the graft. The distal anastomosis to the proximal descending aorta was then completed. Flow was restored to the left subclavian artery through a 10-mm interpositioned Hemashield graft. The patient was rewarmed and weaned from cardiopulmonary bypass.

The patient had a prolonged intensive care unit course because of tracheomalacia resulting from chronic compression by the aneurysm. The patient was able to be weaned from the ventilator and did not require a tracheostomy. He made a full recovery and was asymptomatic at his 6-month follow-up visit. The postoperative follow-up computed tomographic scan demonstrates resolution of the tracheal compression and remodeling of the patient’s aneurysmal sac (Fig 4).

**Comment**

Most vascular rings involving a right-sided aortic arch with a retroesophageal left subclavian artery are diagnosed in infancy or early adulthood. Patients present with symptoms of airway obstruction and/or dysphagia. In adults, a right-sided aortic arch is usually asymptomatic unless aneurysmal dilatation or dissection develops. Aneurysmal dilatation usually occurs at the takeoff of the aberrant left subclavian artery. This aneurysm is known as aneurysm of the diverticulum of Kommerell [2]. Although aneurysms of the diverticulum of Kommerell are relatively common, true aortic arch aneurysms are very rare, with only a few cases previously described [3]. The arch aneurysm accentuates the esophageal and tracheal compression.

Arch aneurysm repair poses a significant technical challenge because of its location deep in the mediastinal space behind the trachea and esophagus and the location of the arch branches on both sides of the visceral structures. No single approach provides access to the whole arch and all of its branches. Several operative approaches have been described. They include right thoracotomy, bilateral thoracotomy, left thoracotomy with sternotomy, sternotomy with right thoracotomy, and left thoracotomy [2].

A left thoracotomy approach offers excellent exposure to the retroesophageal portion of the arch, particularly when there is aneurysmal dilatation that increases the space and allows for an endoaneurysmorrhaphy repair. However, access to the ascending aorta and its branches is limited. Therefore, a right carotid-subclavian bypass or a subclavian-carotid transposition should be performed before the intrathoracic stage to ensure patency of the subclavian artery and to decrease the complexity of the intrathoracic stage.

A left thoracotomy also provides effective decompression of the visceral structures by (1) decompressing the aneurysm, (2) relieving the vascular ring by dividing the ligamentum arteriosum, and (3) mobilizing the trachea and the esophagus from the aneurysmal sac. Recently, endovascular repair has emerged as a potential option [5]. Its utility in arch aneurysms with visceral compression may be limited. Although aneurysmal remodeling is seen after endovascular repair, it is a gradual process that does not provide immediate resolution to the compressive symptoms.

In conclusion, arch aneurysms of the retroesophageal portion of a right-sided aortic arch pose a significant surgical challenge. Careful preoperative consideration of location of the aneurysm and its branches is necessary to plan the surgical approach. A staged operation with arch debranching, followed by arch replacement through a left thoracotomy proved to be a reliable and successful approach.

**References**


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**Agenesis of the Mitral Posterior Leaflet in Elderly**

Hakan Ozkan, MD, Osman Tiryakioğlu, MD, Ahmet Seckin Cetinkaya, MD, Elif Ceylan Uyanik, MD, and Tahsin Bozat, MD

Departments of Cardiology and Cardiovascular Surgery, Medicalpark Hospital, Bursa, Turkey

Congenital mitral valve regurgitation is a rare disease that is found in infancy and childhood, and sometimes in elderly people. In the case presented, mitral regurgitation that was tolerated well until the sixth decade of life is reported. A 62-year-old male suffering from dyspnea was referred to our hospital. Transthoracic echocardiographic examination demonstrated severe mitral regurgitation with suspicion of agenesis of the posterior leaflet with a long, mobile anterior leaflet. A transesophageal echocardiogram and surgical evaluation verified agenesis of the posterior mitral valve. The patient was successfully treated using mitral valve replacement, and no complications occurred.


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**References**


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**Address correspondence to Dr Ozkan, Medicalpark Hospital, Department of Cardiology, Hasim Iscan Cad. Fomara Meydani No:1, Osmangazi, Bursa, Turkey; e-mail: doctorhakan@hotmail.com.**

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**Accepted for publication April 29, 2013.**
Congenital mitral valve regurgitation is a rare disease of infancy and childhood. Most abnormalities of the mitral valve are associated with other cardiac lesions. Isolated mitral regurgitation is rare. Congenital mitral regurgitation is classified according to the anatomic components of the mitral valve, including leaflets, papillary muscle, chordae tendineae, and commissures. Leaflet anomalies include leaflet hypoplasia or agenesis, cleft leaflet, and accessory valvular tissue [1].

A 62-year-old male was referred to our hospital suffering from a complaint of dyspnea. He was admitted to our intensive care unit and classified as New York Heart Association functional class IV with atrial fibrillation. Physical examination revealed a 4/6 pansystolic murmur at the apex, crepitant rale, and bilateral pretibial edema. Chest X-ray showed cardiomegaly with bilateral pleural effusion and pulmonary congestion. Two-dimensional transthoracic echocardiography demonstrated severe mitral regurgitation due to agenesis of the posterior leaflet with a long, mobile anterior leaflet (Fig 1). The left ventricular diastolic diameter was 62 mm, end-systolic diameter 38 mm, ejection fraction 0.66, and pulmonary artery pressure 64 mm Hg. A transesophageal echocardiogram verified severe mitral regurgitation with the posterior leaflet absent (Fig 2). The patient underwent a surgical procedure after intensive medical treatment. A left atriotomy was performed after classical aortic bicalval cannulation. Surgical evaluation confirmed agenesis of the posterior mitral leaflet with absent chorda tendinea and papillary muscle (Fig 3). The anterior mitral leaflet and subvalvular apparatus were severely degenerated. After excision of the anterior mitral leaflet, successful mitral valve replacement was performed with a No. 31 St. Jude Medical mechanical prosthetic valve (St. Jude Medical, St. Paul, MN). Atrial fibrillation ablation was performed according to the maze mapping procedure with irrigated unipolar radiofrequency ablation. The operation was performed without any complications. The patient was successfully discharged on day 7 postoperatively.

Comment

Severe hypoplasia and agenesis of the posterior mitral leaflet is a rare cause of mitral regurgitation. Only a few cases of such a disorder have been reported [2, 3]. In the
case presented, mitral regurgitation was tolerated well until the sixth decade. To our knowledge, this is the first reported case in the literature. Posterior wall motion, a long, mobile anterior leaflet, and annular geometry were able to compensate somewhat for this abnormality. We consider that decompensation occurred when the anterior leaflet degenerated and annular dilatation occurred after left ventricular and atrial enlargement. Ring annuloplasty is an alternative surgical procedure used for similar disorders [3]. This, and mitral valve repair, are particularly preferred in newborns and children to prevent the disorder from compromising children’s development and growth. However, in our case, the patient was a 62-year-old with a missing posterior leaflet and degenerated anterior mitral leaflet. Therefore, we preferred to use mitral valve replacement, which was suitable for this patient.

References

Large Cardiac Tumor Managed With Resection and Two Ventricular Assist Devices
Brian A. Bruckner, MD, Limael E. Rodriguez, MD, Raquel Bunge, RN, Tadashi Motomura, MD, PhD, Jerry D. Estep, MD, Matthias Loebe, MD, PhD, and Michael J. Reardon, MD

The Methodist Hospital, Methodist DeBakey Heart & Vascular Center, Houston, Texas

Symptomatic cardiac tumors can lead to a rapid clinical deterioration and death. Prompt surgical resection is ideal in this situation as it is the only proven treatment to date. We report the radical resection of a large malignant cardiac tumor that obstructed the right ventricular outflow tract. Extensive resection precluded reconstruction and limited the ability to implant a total artificial heart; thus, 2 paracorporeal devices were implanted instead.


Accepted for publication April 29, 2013.

Address correspondence to Dr Bruckner, 6550 Fannin, Ste 1401, Houston, TX 77030; e-mail: babruckner@tmhs.org.

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Published by Elsevier Inc

Soft tissue sarcomas are the most common malignant neoplasm of the heart, pericardium, and great vessels [1]. In some instances, they can obstruct the intracardiac outflow tract, which can lead to sudden circulatory collapse [2, 3]. Because roles for neoadjuvant therapy are not well defined, complete surgical resection of these tumors remains the mainstay treatment for palliation and improvement in clinical outcomes. In this report, we describe our first surgical experience using 2 paracorporeal ventricular assist devices (PVADs) after total cardiectomy in a patient with metastatic cardiac sarcoma.

A 28-year-old male patient with a history of right thigh mesenchymal chondrosarcoma was admitted with a chief complaint of increasing shortness of breath at rest. Echocardiogram showed a dilated right ventricle with a large obstructing mass and a posterior pericardial effusion. Ejection fraction was estimated to be 0.40. Cardiac magnetic resonance imaging clearly demonstrated a severely enlarged heart and an obstructed right ventricle outflow tract (RVOT) (Fig 1). All evidence indicated that the massive right-sided obstruction was the primary cause of the acute biventricular failure and that the remaining metastatic disease was under adequate control. The case was discussed at our multidisciplinary tumor board conference. The consensus was that resection of the tumor causing outflow obstruction was the top priority in this high-risk setting and that long-term survival was feasible, including eventual consideration for cardiac transplantation. Emergency approval from our Institutional Review Board and Ethics Committee was granted, and the patient underwent total heart replacement 2 days later.

Total cardiectomy was planned with concomitant biventricular reconstruction and placement of two Thoratec PVADs (Pleasanton, CA). The SynCardia Total Artificial Heart (TAH; SynCardia, Tucson, AZ) device could not be considered because native atrial tissue and ventricular cuffs would be resected to ensure a complete resection. Intraoperatively, we found a severely enlarged heart and near total replacement of the right ventricle by the tumor (Figs 2A, 2B, 2C). Upon opening the right atrium, we noted tumor extension into the vena cava. A separate large mass (5 × 8 cm) originating from the interventricular septum was the main cause of obstruction at the right ventricular outflow tract (Fig 2). The heart was excised, including the right atrium. Each atrium was reconstructed with a 32-mm diameter Hemashield platinum graft (MAQUET, Inc, Wayne, NJ) (Figs 3A, 3B). To prevent collapse, slight tension was placed on the graft and tissue glue was applied until stiffness was achieved. Also, a vacuum suction was employed at a negative pressure of 10 mm Hg on the right VAD and no suction on the left VAD. The outflow grafts were sewn to the remnants of the pulmonary artery and aorta (Fig 3C). With the pumps set to a fixed rate of 70, flows were generally kept between 3.5 to 4.5 l min⁻¹. The left side VAD flows were adjusted to maintain slightly higher flows (approximately 0.5 l min⁻¹) than the right side VAD. To manage filling...