CLINICAL VIGNETTE

Isolated Mechanical Tricuspid Valve Replacement in an Octogenarian, 25 Years Following Mechanical Mitral Valve Replacement

George Mitchell, M.D., and Megha Agarwal, M.D.

Case Summary

The patient is an 84-year-old woman with a history of acute rheumatic fever at age 12 while living in Mexico. At age 60, after developing severe rheumatic mitral stenosis and regurgitation, she was referred for mechanical mitral valve replacement performed in May of 1991. The tricuspid valve was not addressed at the time of mitral valve surgery. Since her surgery, chronic atrial fibrillation had been treated with a strategy of rate control and anticoagulation in the context of a mechanical mitral valve prosthesis. In 2007, she transferred her care, and an echocardiogram showed moderate tricuspid regurgitation but no evidence for prosthetic mitral valve dysfunction, left ventricular systolic dysfunction, or pulmonary hypertension. There was no clinical evidence of right heart failure at that time. Over the years, mild leg edema had been managed successfully with diuretic therapy, and a repeat echocardiogram in June of 2010 showed a stable degree of tricuspid regurgitation. In May 2012, she suffered a mechanical fall that resulted in chronic low back pain. A CT scan showed compression deformities at L3/T11 but also an incidental finding of inferior vena cava dilatation and hepatic vein enlargement. Echocardiogram at that time was consistent with severe tricuspid regurgitation, while continuing to show a normally functioning mechanical mitral valve prosthesis, normal left ventricular systolic function, and no pulmonary hypertension. The patient began to complain of right upper quadrant pain and abdominal fullness in addition to edema. Her physical examination showed marked jugular venous distension with prominent V waves and a negative Kussmaul sign. Her functional capacity, in the context of her advanced age, appeared to be unaffected. Assuming her symptoms were related to passive hepatic congestion with capsular distention, and ascites and edema were the result of right heart failure, she was offered initiation of an invasive evaluation with a goal of referral for tricuspid valve replacement; however, she declined due to concerns regarding her age of 81 years. Diuretic therapy was continued. In May 2016, an echocardiogram continued to show severe tricuspid regurgitation with consequent right atrial and right ventricular enlargement. Her estimated pulmonary arterial pressure remained normal. Her left ventricular systolic function was normal, and there was no evidence for mechanical mitral valve prosthetic dysfunction. She developed pancytopenia, felt most likely the consequence of splenic sequestration resulting from passive congestion. A fibroscan showed a hepatic stiffness score of 12.1 kPa with a median of 7.3, (normal being 7 and 5.3 respectively), suggesting an element of mild hepatic fibrosis. Due to

suspicion of possible development of cirrhosis as a consequence of longstanding chronic passive hepatic congestion, she underwent an upper endoscopy that showed no esophageal varices. Liver biopsy was felt to be high risk.

Given the report in 2015 of the first use of a transcatheter approach to "bicuspidization" of the tricuspid valve using the Mitralign system, the patient agreed to referral for this procedure as an alternative to a surgical approach for potential mitigation of her symptoms related to chronic passive hepatic congestion and possibly prevention of any further progression of underlying hepatic fibrosis.[1] Unfortunately, her tricuspid regurgitation was felt to be "torrential," precluding the transcatheter protocol. She consented to tricuspid valve replacement, which was performed via a right lateral thoracotomy in August 2016. Operative findings included retraction of the septal and posterior leaflets of the tricuspid valve and a "very large" tricuspid annulus. The valve was replaced with a #33 St. Jude Medical mechanical prosthesis. Permanent epicardial pacing leads were attached to the diaphragmatic surface of the right ventricular wall and placed in a subcutaneous pocket. Heart block developed post-operatively, and the surgically placed leads had unacceptable capture thresholds and pacemaker implantation was abandoned. Fortunately, heart block resolved before discharge.

By five months post-operatively, on clinical examination, jugular venous distention, ascites and edema were no longer present. The white cell count and platelet count had risen from 2,300 and 91,000 pre-operatively to 5,600 and 208,000 respectively. All previous cardiovascular symptoms of right upper quadrant pain, increased abdominal girth, and leg swelling had completely resolved. There have been no subsequent clinically compelling brady-arrhythmias to suggest the need for permanent pacing support. A 6-month post-operative echocardiogram showed a markedly enlarged right atrial volume of 172 milliliters (normal 22-58 milliliters, greater than 79 milliliters being markedly dilated). The mechanical tricuspid valve is functioning normally.

Discussion

Most tricuspid valve surgery is carried out at the time of left-sided valve surgery. The surgical treatment of late secondary or untreated primary tricuspid regurgitation following previous left-sided valve surgery is a challenge. Patients with severe

tricuspid regurgitation after mitral valve surgery are often managed medically for a long period of time and referred for surgery only when they develop intractable symptoms of right heart failure and consequent organ dysfunction. Isolated annuloplasty is often unable to provide effective or durable tricuspid valve competence. Most patients still require tricuspid valve replacement. This case is interesting from the standpoint of her advanced age at the time of surgery, the length of time from the initial left-sided valve surgery, her favorable outcome despite the presence of pre-operative risk factors, and the issue of pacing in the context a mechanical tricuspid valve prosthesis.

This 85-year-old patient may be the oldest reported case of isolated tricuspid valve replacement. A series of fifteen patients from the Massachusetts General Hospital undergoing isolated tricuspid valve replacement over a 12-year period ranged in age from 38 to 72 years with a mean age of 61 years. In this series, 13 of the 15 patients had undergone prior left-sided valve surgery.[2] The only prior report focusing on isolated tricuspid valve replacement involved younger patients with endocarditis who had not undergone previous cardiac surgery. A more recent series of 449 consecutive patients undergoing tricuspid valve surgery for severe tricuspid regurgitation in Seoul, South Korea, ranged in age from 44 to 68 years, and included first time operations and redo cases, as well as multiple concomitant cardiac procedures.[3] Of the total group, 13.4% underwent isolated tricuspid valve replacement in the context of prior left-sided valve surgery. This particular subset undergoing surgery for late tricuspid valve regurgitation after previous mitral surgery, usually has a higher hospital mortality and disappointing late outcome.[4] These reported series do not specify the operative approach to valve replacement. The choice of a lateral thoracotomy and femoral cardiopulmonary bypass may have led to a less complicated post-operative course in this case.

The interval of 25 years between initial mitral valve surgery and subsequent isolated tricuspid valve replacement in this case is likely the longest reported. The interval between left-sided valve surgery and tricuspid valve replacement in the South Korean series ranged from 10.4 – 167.0 months (13.9 years) with a mean interval of 31.0 months. This interval was not reported in the Massachusetts General Hospital series.

Despite the preoperative evidence for hepatic fibrosis, possible cirrhosis, and portal hypertension leading to presumed splenic sequestration and pancytopenia, this patient's white blood cell and platelet counts have improved post-operatively. This would suggest that progression of chronic liver disease due to long-standing chronic passive congestion may still be amenable to mitigation in the setting of late tricuspid valve replacement. Serial measurement of hepatic distensibility with a follow-up fibroscan may shed some light on this theory.

The issue of permanent pacing support in this case, if the need arises, will remain a significant challenge. Obviously, the conventional strategy for lead implantation is not feasible in this patient with a mechanical tricuspid valve prosthesis and inaccessible right ventricle, although there has been one case report of implantation of a transvenous pacing lead in the right ventricular apex, crossing a bileaflet mechanical tricuspid prosthesis.[5] An alternative, unconventional site for pacing in this context would be through the coronary sinus, although this approach may be treacherous given the technical difficulty in controlling guide catheters for engagement of the coronary sinus in a massively dilated right atrium, carrying a risk of entrapment in the mechanical tricuspid valve prosthesis.[6,7] Whatever led to failure of the surgically placed epicardial leads would be a risk if this approach were to be employed again. Perhaps implantation of a leadless pacemaker prior to tricuspid valve replacement would have avoided the current problematic situation.

Although we do not have pathology of this patient's resected tricuspid valve, the description of the operative findings suggests the presence of annular dilatation and leaflet retraction that may have been a secondary consequence of progressive tricuspid regurgitation and right ventricular enlargement, in the setting of what was initially rheumatic disease. Newer guidelines on the management of valvular heart disease suggest consideration of surgical intervention in patients with severe primary tricuspid regurgitation or persistent/recurrent severe secondary tricuspid regurgitation, if signs of right ventricular dysfunction are detected, even in asymptomatic patients.[8] Addressing tricuspid regurgitation that is less than severe at the time of initial left-sided valve surgery may be a consideration, particularly if there is evidence for annular dilatation.[9,10]

In summary, from several aspects, this case is illustrative of the challenges in managing late tricuspid regurgitation following previous left-sided valve surgery, including the risks posed by advanced age, gender, and associated organ dysfunction, the potential choices of surgical and catheter based therapy, and finally, the problematic issue of permanent pacing in the context of an inaccessible right ventricle. This case raises the possibility that age alone may not preclude consideration of surgical intervention, even in the presence of pre-operative risk factors.

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