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Case Report

Transcatheter Aortic Valve Replacement in Patients with Cardiac Amyloidosis

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Abstract

Cardiac amyloidosis and aortic stenosis are conditions which both affect similar populations yet there is sparse literature about the management of aortic stenosis with co-existing cardiac amyloidosis. Generally, patients with cardiac amyloidosis are not thought to benefit from aortic valve replacement due to the fixed systolic and diastolic dysfunction inherent in the disease. We report the cases of three patients with cardiac amyloidosis who underwent successful transcatheter aortic valve replacement.

Keywords: Transcatheter aortic valve replacement; Aortic stenosis; Aortic valve replacement; Cardiac amyloidosis

Case Presentation

Patient A

Patient A is an 83 year old man who had been diagnosed with Aortic Stenosis (AS) three years prior to his eventual Transcatheter Aortic Valve Replacement (TAVR). One year prior to his procedure he developed congestive heart failure with restrictive physiology. Coronary angiography showed non-obstructive Coronary Artery Disease (CAD) and an endomyocardial biopsy detected TTR type amyloid deposits. Echocardiography demonstrated low-flow, low-gradient AS with an increased aortic valve gradient (from 21mmHg to 40mmHg) after the administration of dobutamine. His other co morbidities included systolic dysfunction and Atrial Fibrillation (AF) and his Society for Thoracic Surgery (STS) score calculated a 4.9% risk of mortality and 29.1% risk of significant morbidity, but he was deemed extreme risk for surgery due to his significant diastolic dysfunction and amyloidosis. He ultimately underwent successful placement of a 29mm Medtronic Core Valve by a transfemoral approach. Post-operatively his mean aortic valve gradient improved to 9mmHg, and he had mild paravalvular leak (PVL). Notably he had significant QRS widening (from 100ms to 138ms) with a non-specific interventricular conduction delay post-operatively, but remained in atrial fibrillation with a normal heart rate. His immediate post-operative course was otherwise uncomplicated. However, 14 days post-operatively he returned to the hospital with complete heart block. He underwent pacemaker placement and had no further complications. At 6 month follow-up he had clinically improved and his Left Ventricular Ejection Fraction (LVEF) had improved to 55%with an aortic valve gradient of 11mmHg, and persistent mild PVL. He is now more than 27 months out from his TAVR procedure and has continued to do well from a cardiac perspective. His mean aortic valve gradient is 6 mmHg and the PVL has completely resolved.

Patient B

Patient B was a 65 year old man with systemic AL amyloidosis with significant renal, lung, cardiovascular, and intestinal involvement which had previously been treated with lenalidomide and dexamethasone. He had been followed for AS with serial echocardiography for 2 years prior to TAVR and had progressed to New York Heart Association (NYHA) Class III heart failure symptoms. His co morbidities included a prior episode of heart block with a permanent pacemaker (now in permanent atrial fibrillation), stage III chronic kidney disease, and coronary artery disease with a previously placed stent. He also had left ventricular systolic dysfunction with an LVEF of 40%.By left heart catheterization; his resting aortic valve gradient was 27mmHg which increased to 41mmHg with dobutamine. His STS score calculated a 5.4% chance of mortality with 35.5% chance of morbidity and 14% incremental risk of morbidity due to his immobility, elevated b-type nature tic peptide level, and reduced LVEF. He was considered high risk for surgical aortic valve replacement and subsequently underwent an uneventful TAVR by a transfemoral approach, with placement of a 31mm Medtronic Core Valve. There was initial mild PVL which improved with post-dilation of the valve. His post-operative course was complicated by volume overload, nausea, and vomiting, but he was discharged on post-operative day 9. At 6 month follow-up he was doing well clinically, his EF had improved to 55-60%, and his mean aortic valve gradient was 8mmHg with no PVL. While his cardiac status remained stable he unfortunately developed aspiration pneumonia during a dental procedure and died of bacteremia 11 months following his TAVR procedure.

Patient C

Patient C was a 71 year old woman with systemic AL amyloidosis resulting in End Stage Renal Disease (ESRD) treated with lenalidomide and dexamethasone which was diagnosed 5 years prior to her eventual TAVR. She presented for evaluation for her AS and NYHA class III heart failure symptoms. She had low-flow, low-gradient AS with a mean gradient by echocardiography of 25mmHg, but calculated aortic valve area of 0.55cm^2. Co morbidities included her ESRD and non-obstructive coronary artery disease. Her STS score calculated a 7.1% risk of mortality and 36% chance of morbidity but she was deemed extreme risk due to frailty and amyloidosis. She underwent successful TAVR via a Tran's femoral approach

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with the placement of a 26mm Core Valve which was complicated by intra operative hypotension requiring brief cardiopulmonary resuscitation and vasopressors. Post-operatively there was mild PVL and transient QRS widening which resolved within 24 hours. Her post-operative course was further complicated by atrial fibrillation and thrombocytopenia with mucosal bleeding which resolved with cessation of her clopidogrel. At 7 month follow-up she was doing well clinically, her LVEF was 70% and the mean aortic valve gradient was 4 mmHg with mild PVL. Unfortunately, one year following the TAVR procedure she developed peritonitis related to a peritoneal dialysis catheter and ultimately died from sepsis.

Conclusion

Cardiac amyloidosis is a progressive disorder wherein misfolded proteins infiltrate the heart muscle leading to progressive diastolic and ultimately systolic heart failure. There are many types of cardiac amyloidosis, which are categorized by the type of infiltrating protein. The most common are primary (AL) amyloidosis, secondary (AA) amyloidosis, and ATTR amyloidosis which has both a senile and hereditary variant. Each one of these has a slightly varying pathophysiology and prognosis, though the effect on cardiac function is similar [1]. While the incidence of both amyloidosis (particularly the ATTR type) and calcific AS increase with age, there are few if any reports of aortic valve replacement in the setting of known cardiac amyloid. However, autopsy studies have shown that up to one third of patients who have undergone TAVR have evidence myocardial amyloidosis [2]. Despite this, given the poor prognosis of cardiac amyloidosis along with the systolic and diastolic dysfunction that accompanies this infiltrative cardiomyopathy, it remains unclear whether patients with known clinically significant cardiac amyloidosis and AS benefit from aortic valve replacement.

Though this is only a very small case series, our experience with these three patients shows that at least in some cases patients with cardiac amyloidosis and AS can benefit from TAVR. All three of these patients markedly improved their cardiac status following TAVR but two died within one year of their procedure secondary to other causes. This highlights the importance of other co morbidities associated with high and extreme risk patients undergoing TAVR. It is additionally noteworthy that two of three patients demonstrated augmented valve gradients with dobutamine challenge, which has been used to identify patients with low-flow low-gradient AS that are likely to benefit from aortic valve replacement [3].

One major consideration in both cardiac amyloidosis and TAVR is the effect on the cardiac conduction system. Cardiac amyloidosis is well understood to be associated a variety of conduction abnormalities including atrial fibrillation, tachyarrhythmia's, and conduction block [4]. Likewise, TAVR is also associated with a significant risk of atrioventricular (AV) conduction defects, at least partially due to mechanical force on the conduction fibers from the deployed valve [5-8]. Notably, patient A required pacemaker placement within 30 days from the procedure. It remains to be seen if AV conduction defects would be more likely in amyloid patients, though undiagnosed amyloid prior to TAVR has been raised as a possible risk factor for post-operative AV block [9].

In conclusion, clinically significant severe aortic stenosis does exist in the presence of cardiac amyloidosis. Not only can TAVR can be performed safely and effectively in patients with cardiac amyloid, but aortic valve replacement can lead to significant clinical and hemodynamic improvement despite the underlying cardiomyopathy. AV block may be a more important consideration in these patients who have a high baseline risk for conduction abnormalities. Further study is necessary to help further identify those patients with cardiac amyloidosis and AS who are most likely to benefit from aortic valve replacement and to further determine whether aortic valve replacement will lead to better long-term outcomes in this patient population.

References

1. Shah KB, Inoue Y, Mehra MR. Amyloidosis and the heart: a comprehensive review. Arch Intern Med. 2006; 166: 1805-1813.
2. Nietlispach F, Webb JG, Ye J, Cheung A, Lichtenstein SV, Carere RG, et al. Pathology of transcatheter valve therapy. JACC Cardiovasc Interv. 2012; 5: 582-590.
3. Nishimura RA, Grantham JA, Connolly HM, Schaff HV, Higano ST, Holmes DR. Low-output, low-gradient aortic stenosis in patients with depressed left ventricular systolic function: The clinical utility of the dobutamine challenge in the catheterization laboratory. Circulation. 2002; 106: 809-813.
4. Murtagh B, Hammill SC, Gertz MA, Kyle RA, Tajik AJ, Grogan M. Electrocardiographic findings in primary systemic amyloidosis and biopsy-proven cardiac involvement. Am J Cardiol. 2005; 95: 535-537.
5. Jilaihawi H, Chin D, Vasa-Nicotera M, Jeilan M, Spyt T, Ng GA, et al. Predictors for permanent pacemaker requirement after transcatheter aortic valve implantation with the CoreValve bioprosthesis. Am Heart J. 2009; 157: 860-866.
6. Bleiziffer S, Ruge H, Hörer J, Hutter A, Geisbüsch S, Brockmann G, et al. Predictors for new-onset complete heart block after transcatheter aortic valve implantation. JACC Cardiovasc Interv. 2010; 3: 524-530.
7. Roten L, Wenaweser P, Delacrétaz E, Hellige G, Stortecky S, Tanner H, et al. Incidence and predictors of atrioventricular conduction impairment after transcatheter aortic valve implantation. Am J Cardiol. 2010; 106: 1473-1480.
8. Moreno R, Dobarro D, Lopez de Sa, Prieto M, Morales C, Orbe LC, et al. Cause of Complete Atrioventricular Block after Percutaneous Aortic Valve Implantation: Insights From a Necropsy Study. Circulation. 2009; 120: 29-30.
9. Castaño A, Bokhari S, Maurer MS. Could Late Enhancement and Need for Permanent Pacemaker Implantation in Patients Undergoing TAVR Be Explained by Undiagnosed Transthyretin Cardiac Amyloidosis? J Am Coll Cardiol. 2015; 65: 311-312.

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