Takotsubo cardiomyopathy in the setting of complete heart block

Aasim Afzal, MD^a, John Watson, MD^a, James W. Choi, MD^{a,b}, Jeffrey M. Schussler, MD^{a,b}, and Manish D. Assar, MD^{a,b}

^aDivision of Cardiology, Baylor University Medical Center, The Baylor Scott & White Heart Vascular Hospital, Dallas, Texas;

^bDivision of Cardiology, Texas A&M College of Medicine Health Science Center, Dallas, Texas

ABSTRACT

Few cases of coincident takotsubo cardiomyopathy and complete heart block (CHB) have been reported. A 62-year-old woman presented with typical chest pain and was found to have CHB with a left ventricular ejection fraction of 35% and apical ballooning on ventriculogram. The patient was transvenously paced and a permanent biventricular pacemaker was placed when the CHB did not resolve. Repeat echocardiography 15 days after the event showed the ejection fraction to be 50%. This case highlights management strategies in this unique situation.

KEYWORDS Biventricular pacemaker; cardiomyopathy; complete heart block; takotsubo cardiomyopathy

Takotsubo cardiomyopathy (TC) is a form of non-ischemic cardiomyopathy with a characteristic pattern of apical ballooning, the onset of which is most commonly associated with a strong emotional stressor and subsequent catecholamine surge, giving it the name “broken heart syndrome” in popular media.¹ Few cases have been reported in which complete heart block (CHB) accompanied the TC. We present a case of concurrent TC and CHB to highlight management strategies in this rare situation.

CASE PRESENTATION

A 62-year-old woman with non-insulin-dependent diabetes mellitus, hypertension, and hyperlipidemia presented with a 2-day history of nausea, vomiting, and diarrhea. On the day of presentation, she fainted and drove her car into the car garage wall with airbag deployment. She regained consciousness but had subsequent diarrhea and vomiting and was transported to the emergency room. She denied chest pain or palpitations but related a history of diaphoresis and lightheadedness with standing. Electrocardiography showed sinus rhythm with CHB, an atrial rate of 78 beats per

minute, a junctional escape rhythm of 46 beats per minute, deep T-wave inversions in the lateral leads, and a prolonged QTc of 654 milliseconds (*Figure 1*). No prior electrocardiogram was available for comparison.

The patient remained hemodynamically stable but had yet to recover from CHB. She had an asymptomatic 7-second episode of polymorphic ventricular tachycardia on telemetry that appeared secondary to bradycardia-dependent prolonged QTc, which spontaneously terminated. Given the troponin elevation (2.8 ng/mL) in the setting of new-onset CHB, the patient underwent left heart catheterization. The coronary angiogram showed no luminal narrowing but revealed myocardial bridging of the left anterior descending artery. Left ventriculogram showed the ejection fraction to be approximately 35% with typical left ventricular (LV) apical “ballooning” consistent with TC (*Figure 2*).

A transvenous pacing wire was placed via the right femoral vein, and the patient returned to the intensive care unit. She remained 100% paced over the ensuing 48 hours and underwent placement of a biventricular permanent pacemaker via the left axillary vein on the third day of presenta-

Corresponding author: Manish Assar, MD, The Baylor Scott & White Heart Vascular Hospital, 621 N. Hall St., Suite 400, Dallas, TX 75226 (e-mail: Manish.assar@bswhealth.org)

Color versions of one or more of the figures in this article can be found online at www.tandfonline.com/ubmc.

Received April 23, 2018; Revised July 8, 2018; Accepted July 9, 2018.

Figure 1. Electrocardiogram showing complete heart block on presentation.

Figure 2. Left ventriculogram showing apical ballooning consistent with takotsubo cardiomyopathy.

tion (Figure 3). The patient was discharged the following day on angiotensin-converting enzyme inhibitor and beta-blocker therapy along with a LifeVest and instructions for a follow-up echocardiogram. During follow-up approximately 15 days after presentation, echocardiography showed an ejection fraction of 50%. Pacemaker interrogation revealed pacing >99%. Use of the wearable defibrillator was discontinued at this time. Subsequent 6-month follow-up showed 100% pacemaker dependence and an echocardiogram with an ejection fraction of 55%.

Figure 3. Successful biventricular pacemaker placement with appropriate placement of the coronary sinus lead.

DISCUSSION

Originally described by Dote et al in Japan in 1991,¹ TC received its name from the LV apical ballooning shape resembling a *takotsubo*, or octopus pot.² Clinically, the syndrome has been called stress cardiomyopathy, apical ballooning syndrome, and broken heart syndrome; it represents a transient regional systolic LV dysfunction.² An estimated 1.2% of patients with enzyme-positive acute coronary syndrome will develop TC.³ The presentation often mimics an

acute myocardial infarction.⁴ Of the 1750 patients in the International Takotsubo Registry, 90% are women.² The mean age is 66 years.²

Though the pathogenesis of TC is not well understood, the most commonly discussed mechanism is a catecholamine excess or “surge” from an emotional or physical trigger leading to myocardial stunning or toxicity. Other theories include catecholamine-induced coronary spasms or neuro-hormonal-mediated myocardial dysfunction. The registry demonstrated that emotional triggers were less common than physical triggers (27.7% vs 36.0%, respectively), and 28.5% of patients lacked an obvious trigger.² Medical management includes beta-blocker therapy and/or typical post-acute coronary syndrome or heart failure medications.² However, no randomized trials comparing management strategies for TC exist. In-hospital management is largely supportive, although approximately 10% of patients develop cardiogenic shock.² Ventricular function usually normalizes 2 to 4 weeks after the inciting event.⁵

The condition CHB, also known as third-degree atrio-ventricular (AV) block, is when none of the atrial impulses reach or are conducted through the AV node. Etiologies include both reversible and irreversible causes and both congenital and acquired conditions, including iatrogenic causes such as medications or cardiac surgery, acute myocardial infarction, infiltrative or degenerative processes such as sarcoidosis or amyloidosis, rheumatic diseases, infection, metabolic or electrolyte disturbances, neuromuscular disorders, and toxins. However, many cases have been deemed idiopathic. The estimated prevalence of CHB is 0.04% worldwide and 0.02% in the USA.⁶ In 2008, the American College of Cardiology, the American Heart Association, and the Heart Rhythm Society developed guidelines for placement of devices for the management of CHB,⁷ citing evidence that a permanent pacemaker improves survival in those with irreversible CHB, especially after syncope.^{8–11} Thus, the placement of a permanent pacemaker is a class I recommendation (level of evidence: C).⁷ Initial in-hospital management of a patient with CHB revolves around ensuring hemodynamic stability. Patients who are hemodynamically stable at presentation are often monitored on telemetry with transcutaneous pacing pads in place, pending the placement of a temporary pacing wire or permanent pacemaker. However, patients with hemodynamic instability will often need a transvenous pacing wire placed emergently. Medical management is strictly reserved for special cases in which the level of AV block occurs at the AV node and is thought to be secondary to a reversible cause. In these cases, sympathomimetic agents such as isoproterenol can be used to ensure a sufficient ventricular rate.

Our patient presented to the hospital with CHB and what initially appeared to be acute coronary syndrome but was later discovered to be TC. Despite the significant amount of technical data gathered on the patient during her hospitalization, the crucial data point guiding management

was the history of the present illness. The events leading to the patient’s presentation to the emergency department show that she developed CHB, which led to the syncope and the subsequent car accident, which resulted in TC. Though it is unclear whether the trigger for TC was the physiologic response to CHB or the emotional response to the car accident or a gastrointestinal illness, the patient’s history suggests that the most likely inciting event was the development of CHB. Her medications list did not reveal an iatrogenic cause of CHB, and transthoracic echocardiography did not show evidence of an infiltrative process. The patient had no leukocytosis or fever to suggest systemic infection, no history or evidence of rheumatic disease, and no significant electrolyte or metabolic disturbances despite her recent gastrointestinal symptoms. As such, the authors suspect that her CHB was secondary to conduction system disease and was likely irreversible. The management plan then hinged on placement of a permanent pacemaker and supportive therapy for her TC.

A substudy of the MADIT-II trial suggested that patients who are paced from the right ventricle (RV) >50% of the time are at a higher risk for developing new or worsening heart failure than those paced from the RV ≤50% of the time.¹² A subsequent study demonstrated that patients with LV dysfunction in the setting of RV pacing improved LV function with the addition of biventricular pacing. Evidence from the Homburg Biventricular Pacing Evaluation trial demonstrated that for patients with LV dysfunction or enlargement and conventional pacing indications, a biventricular pacemaker is associated with increased exercise capacity, improved LV ejection fraction, and serum brain natriuretic peptide.¹³ Our patient was fully pacemaker dependent during her hospital stay in the setting of new-onset cardiomyopathy. Thus, a biventricular pacemaker was placed as opposed to a dual-chamber pacemaker, with the goal of maximizing the opportunity for LV function recovery. The concern was that in the setting of 100% pacemaker dependency, RV pacing would hinder LV recovery.

Of all 13 known cases of coincident TC and CHB, 12 had recovery of LV function, with the 13th case not reporting.¹⁴ In 10 cases, a permanent pacemaker was placed, although it was not specified whether this was biventricular or dual chamber.¹⁴ In 4 of the 13 cases, CHB resolved within 3 months.¹⁴ The guidelines for implantable cardiac defibrillator use highly recommend implantation for patients with an ejection fraction ≤35% with New York Heart Association Class II or III heart failure.⁷ Given that the patient’s presumed TC would likely be reversible, an implantable cardiac defibrillator was not used.

1. Dote K, Sato H, Tateishi H, Uchida T, Ishihara M. Myocardial stunning due to simultaneous multivessel coronary spasms: A review of 5 cases. *J Cardiol*. 1991;21(2):203–214.
2. Templin C, Ghadri JR, Diekmann J. Clinical features and outcomes of takotsubo (stress) cardiomyopathy. *N Engl J Med*. 2015;373(10):929–938. doi:10.1056/NEJMoa1406761.

3. Kurowski V, Kaiser A, von Hof K, et al. Apical and midventricular transient left ventricular dysfunction syndrome (tako-tsubo cardiomyopathy): Frequency, mechanisms, and prognosis. *Chest*. 2007;132(3):809–816. doi:[10.1378/chest.07-0608](https://doi.org/10.1378/chest.07-0608).
4. Tsuchihashi K, Ueshima K, Uchida T, et al. Transient left ventricular apical ballooning without coronary artery stenosis: A novel heart syndrome mimicking acute myocardial infarction. Angina Pectoris-Myocardial Infarction Investigations in Japan. *J Am Coll Cardiol*. 2001;38(1):11–18.
5. Wittstein IS, Thiemann DR, Lima JA, et al. Neurohumoral features of myocardial stunning due to sudden emotional stress. *N Engl J Med*. 2005;352(6):539–548. doi:[10.1056/NEJMoa043046](https://doi.org/10.1056/NEJMoa043046).
6. Kojic EM, Hardarson T, Sigfusson N, Sigvaldason H. The prevalence and prognosis of third-degree atrioventricular conduction block: The Reykjavik study. *J Intern Med*. 1999;246(1):81–86.
7. Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities. *Circulation*. 2008;117(21):e350–e408. doi:[10.1161/CIRCULATIONAHA.108.189742](https://doi.org/10.1161/CIRCULATIONAHA.108.189742).
8. Friedberg CK, Donoso E, Stein WG. Nonsurgical acquired heart block. *Ann N Y Acad Sci*. 1964;111:835–847.
9. Gadboys HL, Wisoff B, Litwak RS. Surgical treatment of complete heart block. An analysis of 36 cases. *JAMA*. 1964;189(2):97–102.
10. Hindman MC, Wagner GS, JaRo M, et al. The clinical significance of bundle branch block complicating acute myocardial infarction. 2. Indications for temporary and permanent pacemaker insertion. *Circulation*. 1978;58(4):689–699.
11. Edhag O, Swahn A. Prognosis of patients with complete heart block or arrhythmic syncope who were not treated with artificial pacemakers. A long-term follow-up study of 101 patients. *Acta Med Scand*. 2009;200(1–6):457–463. doi:[10.1111/j.0954-6820.1976.tb08264.x](https://doi.org/10.1111/j.0954-6820.1976.tb08264.x).
12. Sharma AD, Rizo-Patron C, Hallstrom AP, et al. Percent right ventricular pacing predicts outcomes in the DAVID trial. *Heart Rhythm*. 2005;2(8):830–834. doi:[10.1016/j.hrthm.2005.05.015](https://doi.org/10.1016/j.hrthm.2005.05.015).
13. Kindermann M, Hennen B, Jung J, Geisel J, Böhm M, Fröhlig G. Biventricular versus conventional right ventricular stimulation for patients with standard pacing indication and left ventricular dysfunction: The Homburg Biventricular Pacing Evaluation (HOBIPACE). *J Am Coll Cardiol*. 2006;47(10):1927–1937. doi:[10.1016/j.jacc.2005.12.056](https://doi.org/10.1016/j.jacc.2005.12.056).
14. Korantzopoulos P, Nikas DN, Letsas K. High-grade atrioventricular block and takotsubo cardiomyopathy: Case report and review of the literature. *Int Cardiovasc Res J*. 2015;9:119–124.