CASE REPORT

The ‘Inextricabilis Syndrome’: a case with no solution

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Summary

We describe a case of a 58-year-old man with cardiogenic shock who underwent triple vessel coronary artery bypass and a left ventricular assist device (LVAD) implantation. His course was complicated by stroke, worsening mitral regurgitation, aortic regurgitation, and multiple cardiac thrombi while on the device. We provide the details of the patient's hospital course, management, and echocardiographic findings. We also discuss the utility of echocardiography before LVAD insertion and its role for continued monitoring after insertion.

Learning points:

- Ventricular assist devices (VADs) are used as bridge to decision, transplant, recovery, or destination therapy in patients with advanced heart failure and cardiogenic shock.
- VADs improve survival and the quality of life but have significant associated complications.
- Echocardiography plays an essential role before VAD insertion and for postoperative cardiac monitoring. Information provided by echocardiography is used in device selection, consideration for corrective surgical interventions, and device explantation.

Background

Ventricular assist devices (VADs) have been shown to have survival benefit and improve quality of life in patients with advanced heart failure and acute cardiogenic shock (1, 2). They are used as a bridge to decision, transplant, recovery, or destination therapy. Nonetheless, they are associated with complications, namely thrombosis and thromboembolic stroke, bleeding, and valvular dysfunction. Echocardiography plays an essential role perioperatively. It helps in evaluating cardiac function, anatomy, and valvular pathology before device insertion and helps monitor for complications and cardiac function recovery after insertion.

Case presentation

A 58-year-old man presented with a 1-day history of chest pain and was found to be in cardiogenic shock secondary to a non-ST segment elevation myocardial infarction. Emergent coronary angiography demonstrated severe triple vessel disease. The triple vessel coronary artery bypass was performed and a CentriMag LVAD was placed for circulatory support. A non-contrast head CT was obtained, given that there was no improvement in mental status once sedation was removed, which demonstrated a subacute non-hemorrhagic infarct in the left middle cerebral artery territory. A postoperative echocardiogram demonstrated severely depressed left ventricular (LV)
function, mild mitral valve regurgitation (MR), and an LV apical mural thrombus (Fig. 1A, B and Videos 1, 2) along with an obstructive ascending aortic thrombus (Fig. 1C, D and Video 3). Heparin drip was continued and transthoracic echocardiography was used to monitor LV function in consideration for CentriMag explantation. Subsequently, the CentriMag LVAD was replaced with a HeartMate II LVAD for destination therapy given persistent LV dysfunction. A transesophageal echocardiogram (TEE) at the time of HeartMate II implantation showed resolution of the LV apical mural thrombus and ascending aortic thrombus. However, some chordae tendineae were torn during LVAD placement and moderate MR developed. Aortic insufficiency at the time of implantation was moderate. Follow-up echocardiograms demonstrated persistence of a low ejection fraction with global LV hypokinesis, moderate MR, a flail posterior mitral leaflet (Fig. 2B, C, D and Videos 4, 5, 6), moderate aortic regurgitation (AR) (Fig. 2B and Video 6), and a new large mobile thrombus in the left atrium (Fig. 2A and Video 7). The patient was discharged to a rehabilitation facility with a HeartMate II LVAD after a hospital stay for 6 months. However, he died 4 months after discharge.

**Video 1**
Transthoracic echocardiography, apical four-chamber view, demonstrating a large apical mural thrombus. Download Video 1 via [http://dx.doi.org/10.1530/ERP-14-0044-v1](http://dx.doi.org/10.1530/ERP-14-0044-v1)

**Video 2**
Transthoracic echocardiography, 3-dimensional image, demonstrating a large apical mural thrombus. Download Video 2 via [http://dx.doi.org/10.1530/ERP-14-0044-v2](http://dx.doi.org/10.1530/ERP-14-0044-v2)

**Video 3**
Transthoracic echocardiography, apical four-chamber view, showing a thrombus originating in the left ventricular outflow tract. A CentriMag cannula is present in the left atrium. Download Video 3 via [http://dx.doi.org/10.1530/ERP-14-0044-v3](http://dx.doi.org/10.1530/ERP-14-0044-v3)
Discussion

There are multiple complicating factors in this case, which made finding a good and definitive treatment for this patient problematic. The initial presentation with acute cardiogenic shock by itself entails a high mortality rate of up to 70% (1). Recent small, non-randomized, trials have shown improvement in survival with the use of mechanical circulatory support devices in this population (1). These devices are used as a bridge to decision, transplant, recovery, or destination therapy (3). Echocardiography plays a crucial role in patients before and after the insertion of VADs. Preoperative echocardiography is used to evaluate right ventricular function, tricuspid regurgitation, aortic insufficiency, aortic stenosis, ascending aortic abnormalities, mitral regurgitation, mitral stenosis, intracardiac thrombus, and right-to-left shunt. Postoperative echocardiography may be used to assess device function, LV recovery, and complications such as worsening aortic insufficiency, aortic dissection, or right-to-left shunt as a result of LV unloading by the device. These measures are essential in device selection, consideration for corrective surgical interventions, and device explantation, which have been described previously (4, 5).

Our patient was initially placed on CentriMag as bridge to recovery, which has been shown to be a good short-term

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**Video 4**
Transesophageal echocardiography with color Doppler demonstrating moderate mitral regurgitation due to a flail posterior mitral leaflet and a ruptured chordae. Download Video 4 via [http://dx.doi.org/10.1530/ERP-14-0044-v4](http://dx.doi.org/10.1530/ERP-14-0044-v4)

**Video 5**
Transesophageal echocardiography demonstrating a flail posterior mitral leaflet and a ruptured chordae. Download Video 5 via [http://dx.doi.org/10.1530/ERP-14-0044-v5](http://dx.doi.org/10.1530/ERP-14-0044-v5)

**Video 6**
Transesophageal echocardiography demonstrating moderate aortic regurgitation and moderate mitral regurgitation. Download Video 6 via [http://dx.doi.org/10.1530/ERP-14-0044-v6](http://dx.doi.org/10.1530/ERP-14-0044-v6)

**Video 7**
Transesophageal echocardiography demonstrating a large left atrial thrombus. Download Video 7 via [http://dx.doi.org/10.1530/ERP-14-0044-v7](http://dx.doi.org/10.1530/ERP-14-0044-v7)

**Figure 2**
Transesophageal echocardiography (TEE) (A) demonstrating a large left atrial thrombus (green arrow), (B) diastolic mitral and aortic regurgitation, (C) moderate mitral regurgitation due to a flail posterior mitral leaflet (yellow arrow), and (D) a flail posterior mitral leaflet from a ruptured chordae (yellow arrow).
option (3, 6). However, his cardiac function did not recover and he was not a candidate for cardiac transplantation given cerebrovascular accident and neurological deficits. Consequently, his CentriMag was changed to a HeartMate II for destination therapy, which has been shown to improve survival and quality of life (2, 7, 8). His course was complicated by an apical mural thrombus, which would have presented a technical challenge for the surgeon and increased the patient’s stroke risk significantly with HeartMate II insertion. However, intraoperative TEE did not demonstrate the thrombus that was observed almost 3 months before, but it did show a new thrombus in the left atrium despite low intensity heparin drip (PTT range 40–60). Consequently, higher intensity anticoagulation was achieved with INR goal of 2.5–3 on warfarin. Furthermore, pre-cardiopulmonary bypass TEE showed only mild AR, which worsened to moderate after the placement of the HeartMate II device. Significant AR can decrease effective systemic circulation and repair or replacement of the valve was considered but not performed during device placement due to the patient’s overall critical condition. Progression of AR is a complication of LVAD support and is probably multifactorial. Contributing mechanisms for AR progression include subphysiological LV pressures resulting in elevated aorta-to-LV pressure gradient, aortic blood flow dynamics, aortic root dilation, and VAD type. The presence of the device cannula in the ascending aorta can also lead to valvular distortions conducive to insufficiency (4, 9, 10). The worsening of MR in our case was secondary to chordae rupture after HeartMate II insertion and in combination with AR has the potential for increased pulmonary artery hypertension and the subsequent right ventricular failure (4).

This patient, while unique, represents a new class of patients who previously would have succumbed to their illness at an earlier point in its course. Advances in medical and surgical therapy of heart failure patients have resulted in life-saving benefits as well as the development of a new group of patients with illnesses that can be temporarily managed but ultimately are not reversible or survivable; hence, ‘Inextricabilis Syndrome’ – a case with no solution.

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Patient consent
The subject of this case report is deceased and no consent is available. No next of kin could be found from which to obtain permission. The case report does not contain any information from which it is possible to identify the patient.

Declaration of interest
The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.

References