Three patients with signs of acute flail mitral leaflet seen on emergency department echo: a critical constellation within the focused cardiac exam

Abstract
Acute flail mitral leaflet is a time-sensitive, reversible cause of cardiogenic shock. Transthoracic echocardiography (echo) is increasingly becoming a vital tool for non-cardiologist physicians who treat patients with undifferentiated chest pain and dyspnoea. The sonographic abnormalities seen in acute flail mitral leaflet are within the boundaries of a focused echo. Individually, these findings are non-specific. As a constellation, however, they are highly suggestive of this disease process. We present a case series of three patients with acute flail mitral leaflet seen on emergency department echo along with a discussion of the findings and the disease itself.

Keywords: chordae tendineae, echocardiography, mitral valve insufficiency, papillary muscles.

Introduction
Acute flail mitral leaflet (FML) is a time-sensitive, reversible cause of cardiogenic shock. Two conditions causing FML are papillary muscle rupture (PMR) and chordae tendineae rupture. Physicians who treat critical patients are increasingly incorporating focused, bedside, transthoracic echocardiography (echo) into their practice. Many will ultimately hold a probe to the chest of a patient with undiagnosed FML. Signs of this process can be easily seen during the course of a focused echo if the sonologist knows what to look for. We present three cases of FML, all cared for by the same emergency department.
physician (EP), and a discussion of both the findings and the disease process itself.

**Case 1**
A 79-year-old female presented to the emergency department (ED) with four hours of chest pressure radiating to her jaw and shortness of breath. She had been experiencing similar but intermittent symptoms over the prior few days.

She was diaphoretic, afebrile and in moderate respiratory distress. Her vital signs were as follows: blood pressure (BP) 116/56 mmHg, pulse (P) 96 beats/minute, respiratory rate (RR) 32 breaths/minute, and O2 saturation 84% on room air (RA). Her heartbeat was regular with no audible murmur. Her lung exam revealed bilateral crackles. She had no peripheral oedema or distended neck veins. The remainder of her exam was unremarkable. Her ECG showed borderline 1 mm ST elevation in II, III and aVF with ST depression in V2-3. Her chest x-ray (CXR) showed mild pulmonary oedema.

An EP performed a focused echo, which showed the following: no pericardial effusion, no signs of right ventricular (RV) strain, and a hyperdynamic left ventricle (LV) (Video clip 1). Additional findings included bilateral “lung-rockets,” suggesting pulmonary oedema (Figure 1), and a small mitral regurgitation (MR) jet aimed laterally into a normal-sized left atrium (LA) (Figure 2).

Within twenty minutes of arrival, the patient was in the cardiac catheterisation laboratory for coronary reperfusion. She had a stent placed in her second obtuse marginal artery (OM2), which was 95% occluded. During the procedure, she went into shock. It was then recognised that she had acute MR due to a partially ruptured posterior papillary muscle. She was stabilised and underwent a successful mitral valve replacement (MVR) the following day. Unfortunately, her initial echo findings, which suggested papillary muscle rupture, were not appreciated in the rush to achieve coronary reperfusion.

**Case 2**
A 71-year-old man presented to the ED via taxi complaining of sudden, severe shortness of breath without chest pain. He had experienced a similar episode two days prior that had resolved spontaneously.

He was in moderate respiratory distress. He was afebrile,
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Figure 3: Apical four-chamber view showing an echogenic mass in the left ventricle (LV). LA= left atrium. RA= right atrium. RV= right ventricle.

Figure 4: Apical two-chamber view showing a ruptured papillary muscle (pap) in the left ventricle (LV). MV= mitral valve. LA= left atrium.

Video Clip 2
Video 2 http://youtu.be/kB_oncRSVK8. Apical four-chamber view showing a mobile, echogenic mass (ruptured papillary muscle) in the left ventricle.

Video Clip 3
Video 3 http://youtu.be/3VjoNUTbPxQ. Apical two-chamber view showing a ruptured papillary muscle (arrow) possibly prolapsing into the left atrium.
with the following vital signs: BP 128/76, RR 38, P 111, and O2 saturation of 89% on RA. He had reduced breath sounds bilaterally, without crackles or wheeze. He had no jugular venous distention and his heart sounds were regular, without an audible murmur. The remainder of his physical exam was unremarkable. His initial ECG showed a sinus rhythm with T-wave inversion in II, III, aVF and in V5-6. His CXR showed mild cardiomegaly and clear lung fields.

While awaiting blood results and a formal plan, the patient developed sudden and marked dyspnoea and hypotension. An EP performed a focused echo, which showed the following: no pericardial effusion, no signs of RV strain, and normal-hyperdynamic LV systolic function. Additional findings included bilateral lung rockets, an echogenic mass in the LV, seemingly out of synchrony with the wall motion (Figure 3 and 4; video clips 2 and 3) and an MR jet aimed laterally into a normal sized left atrium. (Figure 5; video clip 4). An emergent formal echocardiogram confirmed the suspected diagnosis of ruptured papillary muscle. The patient underwent successful MVR surgery and uneventful recovery. His angiogram showed occlusion of the OM2.

Case 3
A 48-year-old male presented to the ED with chest tightness and shortness of breath for two hours. His only medical history was ankylosing spondylitis. He was afebrile and in mild respiratory distress. His vital signs were as follows: BP 100/60, P 100, O2 saturation 89% RA. He had mild bilateral crackles and a systolic murmur radiating to the carotid artery. His ECG did not show any ischaemic changes and his CXR was unremarkable. He was treated for presumptive acute coronary syndrome. While awaiting hospital admission, he became haemodynamically unstable. He was intubated for respiratory failure and shock. An EP performed a focused echo, which showed the following: no
pericardial effusion, no signs of RV strain, and a hyperdynamic LV (video clip 5). Additional findings included bilateral lung rockets, and an eccentric MR jet aimed at the posterior wall of a normal sized LA (Figure 6). The suspected diagnosis of acute FML was confirmed on transoesophageal echocardiography (TOE). He had a normal coronary angiogram and underwent a successful MVR procedure. Intraoperatorively, he was noted to have ruptured chordae tendineae.

Discussion

It may be that the episodes of dyspnoea experienced by the first two patients in the days prior to presentation represented small, acute myocardial infarctions (MI) from occlusion of the OM2. This branch of the left circumflex artery feeds the papillary muscles and can be involved in cases of PMR. PMR complicates up to 5% of cases of fatal, acute MI. Most patients present with dyspnoea, signs of heart failure, and haemodynamic instability. Rupture typically occurs 2–7 days post-infarction and 85% of the time involves the posterior papillary muscle. The coronary branches that feed the papillary muscles are small and the area of myocardial infarct is often quite limited. However, the 24-hour mortality rate is 80% without immediate surgical repair. It is critical, therefore, to identify these patients early, as their myocardia are usually fairly well preserved. In fact, of those who survive 30 days post-operatively, the overall rates of five-year survival and five-year survival without congestive heart failure are identical to matched controls with MI alone. PMR can be either partial or complete. Each of the two papillary muscles is attached to both mitral leaflets. PMR affects the valve in an asymmetric fashion. Regurgitant blood, therefore, enters the LA on an angle, hitting and continuing along the LA wall rather than travelling straight down the middle of the valvular orifice. This “eccentric” MR jet is present in 78% of cases of FML. When this happens, the LA does not have time for

Video Clip 5

Video Clip 5 Video 5 http://youtu.be/PpqfES8bB_U. Parasternal long-axis view (taken in general indicator-to-screen orientation) showing a small, eccentric, mitral regurgitation jet aimed posteriorly into a normal sized left atrium.
compensatory dilatation and pulmonary oedema occurs rapidly. It is difficult to grade the severity of an eccentric jet because it hits the wall early in its course; consequently, a significant MR jet may appear deceptively small.

Chordae tendineae rupture is seen most commonly as the result of (in descending order of frequency) mitral valve prolapse, sub-acute bacterial endocarditis, rheumatic heart disease, and myxomatous degeneration.5 Chordae tendineae rupture differs from papillary muscle rupture in that the first is rarely caused by ischaemia and, depending on the number of chords affected, may not produce symptoms.6 Significant rupture causes FML and presents in a similar fashion to PMR both clinically and echocardiographically. Differentiating the two at the bedside of a critical patient is irrelevant as they are both surgical emergencies.

Over-reliance upon auscultative findings to help confirm or rule-out the diagnosis of FML can be risky. The rapid equilibration of LA and LV pressures seen in acute MR can significantly diminish the murmur.10 Accurate auscultation of a distressed patient in a noisy resuscitation room can be challenging, and the information gained from heart sounds may contribute little towards the diagnosis. TOE is the diagnostic modality of choice for suspected acute FML, but it is time consuming, requires specific personnel, is rarely available after-hours, and is difficult to perform in an unstable patient. Focused transthoracic echocardiography can offer simple findings that strongly suggest the presence of FML and can be performed at the bedside by a trained sonologist.

MR is easily visualised using the colour Doppler function available on most portable ultrasound machines. With optimal colour settings (gain set just below the level of speckling artifact) MR appears as systolic flow from the LV to the LA. Though an apical four-chamber view is preferable, a significant regurgitant jet can be identified from most conventional cardiac views. Even small eccentric MR jets should raise suspicion in the right clinical setting. Other signs of acute FML include the following: a hyperdynamic LV with signs of heart failure, any significant MR into a normal sized LA, papillary muscle head visualised in the LA, or a mobile echo density in the LV out of synchrony with LV wall motion. If myocardial infarction is involved, these findings may be found in combination with regional wall motion abnormalities. For the more advanced users with sophisticated cardiac calculation packages, additional information can be gained by quantifying the MR jet using continuous-wave Doppler or demonstrating diminished LV outflow using the continuity equation.

The use of focused echo is now endorsed by such specialty colleges as: The Australasian College for Emergency Medicine (ACEM), The Australian and New Zealand College of Anaesthetists (ANZAC), The College of Intensive Care Medicine (CICM), and many of their respective counterparts in North America and Europe. In addition, echo proficiency requirements are making their way into the training curricula of most of these groups. The Australasian Society for Ultrasound in Medicine (ASUM) offers a formal certification process for this study. With this increase in focused echo will come a proportional increase in sonologists staring at the sonographic signs of acute FML.

Analysis of the cases
The first patient had normal BP and no appreciable murmur. Her echo showed a hyperdynamic LV with lung rockets, and mild MR into a normal sized LA. The eccentric direction of the MR jet was only appreciated in retrospect with a frame-by-frame analysis of the cineloop. The rush to reperfuse her coronary arteries limited the amount of time available to consider diagnoses beyond her acute ST elevation MI.

The second patient also had no appreciable murmur, but his dramatic deterioration provided a strong clinical clue. His sonographic signs of FML were more definitive: a hyperdynamic LV with lung rockets, eccentric MR into a normal-sized LA, and an echogenic mass in the LV seemingly out of synch with the wall motion (the ruptured papillary muscle). His cineloops were analysed frame-by-frame at the bedside, confirming the suspected diagnosis of papillary muscle rupture.

The third patient was supine, intubated, and in shock at the time of his echo. He was the only patient in this series who had an appreciable murmur. His clinical condition also provided a strong clue. His best window was through a parasternal long axis view. Though the tachycardia and mechanical ventilation limited the study, the following findings became clear during frame-by-frame analysis at the bedside: a hyperdynamic LV with lung rockets, and eccentric MR into a normal sized LA. His was not an ischemic event, as is usually the case with CTR. The ruptured chordae may have been related to his underlying ankylosing spondylitis.

Conclusion
Papillary muscle rupture and chordae tenindae rupture resulting in acute mitral regurgitation are uncommon, but time-critical, diagnoses. We presented three such patients, all having received an echo by the same EP over a three-year period. Each of these patients showed sonographic evidence of their diagnosis. It is important for clinicians to recognise the constellation of findings seen in this disease process: a hyperdynamic LV with signs of heart failure, eccentric or any significant MR into a normal sized LA, papillary muscle head visualised in the LA, or a mobile echo density in the LV out of synchrony with LV wall motion. If there is doubt, it is best to record a cineloop and play it back frame-by-frame at the bedside for confirmation. A recent consensus statement from American College of Emergency Physicians and the American Society of Echocardiography recognises that additional pathology may be encountered during the performance of a focused echo. In such cases, they recommend referral for formal confirmatory studies.11

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