MODULE 16: *Cardiology*

PART 5

Myocarditis

by Kate O'Donovan

MYOCARDITIS simply means inflammation of the myocardium and is defined by the World Health Organization/International Society and Federation of Cardiology Taskforce as an" inflammatory disease of the myocardium diagnosed by established histological, immunological and immunohistochemical criteria.[1] The natural history of myocarditis is poorly understood and what is known is derived from animal studies. The majority of patients remain asymptomatic and the condition is self-limiting although a minority will develop refractory heart failure, which may progress to dilated cardiomyopathy requiring cardiac transplantation due to irreversible left ventricular pump damage.

There are several causes associated with the condition but viruses are recognised as the most common cause, where patients report a prodrome of respiratory or gastrointestinal symptoms prior to developing cardiac signs and symptoms *(see Table 1)*.

In the majority of cases the causative agent is unknown but there is consensus that viruses are an important cause of myocarditis in Europe,[3] whereas bacterial diseases are less commonly associated with the condition.

Myocarditis is usually characterised by myocardial cell injury and necrosis associated with inflammation. Initially the virus gains entry to the patient either via the respiratory or gastrointestinal system where it then travels to the myocardium. In response to the virus entering the myocardium the autoimmune system is activated in an attempt to eradicate the virus. In some patients the autoimmune response remains active despite the virus being destroyed which results in the production of cytokines and other inflammatory enzymes. These serve to target the myocardium resulting in myocardial inflammation and cell death.

The end result is impaired left ventricular function where cardiac output falls and heart failure symptoms become apparent. In response to a decreased cardiac output compensatory mechanisms such as the sympathetic nervous system and the renin angiotensin aldosterone system are activated. These mechanisms attempt to increase cardiac output and maintain peripheral perfusion. The physiological effect is tachycardia, cool clammy skin, diaphoresis and decreased urinary output. Despite persistence of the autoimmune response some patients recover and their baseline ventricular function returns. Some patients progress to dilated cardiomyopathy where irreversible left ventricular dysfunction is present and heart failure symptoms persist and progress in severity.[4]

Major causes of myocarditis[2]

Viral
- Adenovirus
- Coxsackievirus
- HIV
- Hepatitis C

Bacterial
- Streptococcal species
- Staphylococcal
- Pneumococcal

Fungal

Parasitic
- Toxoplasmosis
- Cytomegalovirus

Toxins
- Cocaine
- Anthracyclines

Drug reactions
- Sulphonamides
- Cephalosporins

Autoimmune disease
- Sarcoidosis
- Systemic Lupus Erythematosus

Clinical presentation

Symptoms vary depending on the cause, the patient's overall baseline health status and the extent of myocardial involvement. Therefore clinical presentations may range from asymptomatic and silent to acute dilated cardiomyopathy associated with fulminant heart failure or cardiogenic shock. McCarthy et al[5] describe fulminant myocarditis as a critical illness such as haemodynamic collapse or cardiogenic shock at presentation but it is associated with an excellent long-term survival.

The majority of patients remain asymptomatic. Patients may report a viral prodrome of fever, myalgia, and respiratory or gastrointestinal symptoms followed by symptoms of haemodynamic collapse or heart failure. The incidence of a reported viral prodrome is highly variable ranging from 10-80% of those with documented myocarditis.[2] Acute dilated cardiomyopathy is described as the most dramatic presentation. The true incidence of myocarditis is difficult to determine due its diversity in clinical presentation.

Patients may also present with symptoms masquerading as an acute coronary syndrome. Chest pain if experienced is described as sharp, stabbing, precordial pain or as substernal, squeezing pain similar to that of myocardial infarction.

Angelini et al[6] state that elevated troponin levels and ECG changes suggestive of acute myocardial ischaemia are common. ECG changes may include ST segment elevation in ≥ 2 contiguous leads, T wave inversion, widespread ST depression and pathological Q waves. Angiography may demonstrate segmental or global wall motion abnormality despite normal coronary arteries. Car-

Sponsored by an unrestricted grant from Merck Sharp & Dohme Ireland (Human Health) Ltd and Schering Plough Pharmaceuticals (Ireland) Ltd

diovascular nurses should consider acute myocarditis in younger patients who present with an acute coronary syndrome picture despite having a normal coronary risk factor profile.

Investigations

Aside from endomyocardial biopsy there are no specific investigations for myocarditis. Diagnosis is made on clinical suspicion, physical examination, good history taking and is supplemented using a range of investigations. Physical examination may be unremarkable or reveal signs of heart failure such as tachycardia, dyspnoea, pulmonary oedema, hypotension and diaphoresis.[4] The patient's history may expose a recent respiratory or gastrointestinal illness and a prodrome of non-specific symptoms such as fever, fatigue and myalgia. Common investigations include ECG, blood work, CXR, myocardial imaging and more specifically endomyocardial biopsy. The ECG may demonstrate non-specific changes such as ST segment depression or elevation, T wave inversion or the presence of pathological Q waves. These changes are not specific for myocarditis but are associated with conditions such as acute coronary syndrome. In addition the patient may experience arrhythmias or conduction disturbances such as ventricular tachycardia or second or third degree heart block. Arrhythmias and conduction disturbances arise from irritability within or around the conduction system caused by inflammation.

Cardiac biomarkers such as troponin and creatinine kinase are routinely reserved when myocarditis is suspected. Blood cultures and a white blood cell count can help to identify infection if present whereas serum viral titres may reveal the causative agent. Stool or throat cultures may reveal the primary viral or bacterial cause of infection. Chest x-ray may reveal cardiomegaly with or without pulmonary congestion. Feldman and McNamara[3] recommend measuring erythrocyte sedimentation rate and rheumatologic screening in patients with unexplained heart failure associated with signs and symptoms of connective tissue disease such as systemic lupus erythematous or sarcoidosis. These patients commonly present with a poor left ventricular function although the left ventricular structure is normal on echocardiography

Non-invasive myocardial imaging used in the diagnostic evaluation of suspected myocarditis include echocardiography, nuclear imaging with contrast and MRI.

Echocardiography is currently advocated in the initial evaluation as it has the ability to reveal left ventricular dysfunction, segmental wall motion abnormalities and left ventricular hypertrophy. In addition, it can be used to evaluate response to treatment strategies. Aside from echocardiography, contrast enhanced MRI appears to be a promising means for localising areas of myocardial inflammation and necrosis.

Endomyocardial biopsy is considered gold standard for the diagnosis of myocarditis but it has several limitations. It involves taking three to five samples of the endomyocardial septum tissue via femoral venous access. As the inflammation in myocarditis is normally patchy, healthy tissue may be taken which limits the diagnostic possibility. Mason et al[7] described the incidence of a positive biopsy in clinically suspected myocarditis as being approximately 10%. The Dallas Criteria was established in 1986,[8] which define microscopically the presence of myocarditis. Despite this criteria there is great variability among expert pathologists evaluating the same biopsy.[9] Endomyocardial biopsy is normally reserved for those who are not recovering or are deteriorating despite optimal evidence based therapy.

Prognosis and treatment

The outcome and natural history of myocarditis is as varied as its clinical presentations. Baseline left ventricular function is considered a pivotal factor in predicting recovery and prognosis. If the patient has normal left ventricular function they tend to recover well and normal function will return. In comparison to those who have a history of impaired left ventricular dysfunction with an ejection fraction of ≤ 35%, 50% will develop chronic LV dysfunction, 25% will progress to transplantation or death and 25% will demonstrate spontaneous improvement in their ventricular function.[8] In relation to age and gender Felker[10] stated that older age and male gender were associated with a poorer outcome compared to younger age and female gender.

Treatment is aimed at preserving myocardial function and preventing circulatory collapse. Haemodynamic support and symptom relief are the primary principles employed as opposed to identifying and treating the causative agent. For those with fulminant myocarditis, where circulatory collapse or cardiogenic shock is present, aggressive heart failure therapy consisting of diuretics, inotropes and vasopressors may be required.

In this clinical scenario circulatory support such as intra aortic balloon pump counterpulsation, a ventricular assist device or extracorporeal membrane oxygenation are used in an attempt to prevent multi-organ failure and to maintain peripheral perfusion. Once the patient has been stabilised subsequent heart failure treatment should follow heart failure guidelines,[11] which include beta blockade and inhibition of the renin angiotensin aldosterone system with ACE inhibitors or angiotensin receptor blockers (ARBs).

For those with less severe symptoms and who are maintaining their blood pressure and peripheral perfusion, treatment is aimed at symptom relief such as anti pyretics for pyrexia, diuretics if fluid overload evident and anti arrhythmics if the patient experiences an arrhythmia such as ventricular tachycardia or atrial fibrillation. As the pathophysiology of myocarditis involves an immune response the literature debates whether there is a role for immunosuppression in the treatment strategy. Study results[7,12] suggest that immunosuppression should not be routinely prescribed as it does not positively affect mortality rates but do advocate its use in myocarditis associated with an autoimmune disease such as systemic lupus erythematous, scleroderma and polymyositis.

Nursing implications

The nursing assessment may reveal presence of fatigue, dyspnoea, fever, joint pain, tachycardia and chest discomfort which is described as soreness or mild continuous pressure.[13] History taking may disclose that the patient has experienced a recent respiratory or gastrointestinal illness. The cardiovascular-respiratory assessment may demonstrate increased respiratory rate with involvement of accessory muscles, abnormal heart sounds such as S3 or crepes on auscultation, which implies that pulmonary oedema may be present. The patient may be tachycardic, normotensive or hypotensive and pyrexia may be present. The cardiovascular-respiratory assessment is undertaken on a regular basis or as acuity requires so that improvement or deterioration in cardiovascular function is detected promptly.

Bed rest and restriction in activities are promoted in order to conserve myocardial oxygen consumption and reduce left ventricular workload. The cardiac rhythm is monitored continuously

for arrhythmias and to evaluate the effect of anti arrhythmics if prescribed.[14] Care for circulatory support if employed is as per the device used and local policy.

Following ventricular recovery and in preparing the patient for discharge, patient education should include information on any activity restrictions or dietary restrictions (low salt diet, fluid restriction) as well as teaching the patient on how to recognise signs and symptoms of heart failure. These signs and symptoms include weight gain, clothes and shoes beginning to tighten, breathlessness on normal activity and deterioration in activity levels. It is important to emphasise that limitations in activity are only temporary allowing the myocardium and ventricular function to return to baseline function.

A minority of patients do not recover and have persistence of heart failure symptoms and progress to irreversible left ventricular dysfunction and dilated cardiomyopathy. Treatment options for this cohort of patients may consist of long-term circulatory support such as intra aortic balloon pump counterpulsation or a ventricular assist device with a view to heart transplantation. These patients require psychological care and support as they prepare for transplantation, or if not suitable for transplantation, in coping with their life limiting illness.

Myocarditis is inflammation of the myocardium which impairs pump function. In the majority of patients recovery ensues or the myocarditis maybe asymptomatic and self limiting. In comparison a small number of patients may develop irreversible ventricular dysfunction and dilated cardiomyopathy requiring heart transplantation or death may ensue. Treatment is aimed at relieving symptoms and preserving ventricular function. Nurses have a pivotal role in providing psychological support and in detecting deterioration in ventricular function and assessing the effect of treatment strategies on ventricular function.

Kate O'Donovan is course co-ordinator for the postgraduate diploma in cardiovascular nursing in the Mater Hospital, Dublin

References

1. Richardson P, McKenna W, Bristow M et al. Report of the 1995 World Health Organisation/International Society and Federation of Cardiology Task Force on the Definition and Classification of Cardiomyopathies. Circulation 1996; 93: 341-342

2. Magnani JW, Dec GW. Myocarditis current trends and treatment. Circulation 2006; 113: 876-890

3. Feldman AM, McNamara D. Medical progress: myocarditis. NEJM 2000; 343(9): 1388-1398

4. Liu PP, Mason JW. Advances in the understanding of myocarditis. Circulation 2001; 104: 1076-1082

5. McCarthy III RE, Boehmer MD, Hruban RH et al. Long term outcome of fulminant myocarditis as compared with acute (nonfulminant) myocarditis. NEJM 2000; 342(10): 690-695

6. Angelini A, Calzolari V, Calabrese F et al. Myocarditis mimicking acute myocardial infarction: role of endomyocardial biopsy in the differential diagnosis. Heart 2000; 84: 245 -250

7. Mason JW, O'Connell JB, Herskowitz A et al. A clinical trial of immunosuppressive therapy for myocarditis. NEJM 1995; 333(5): 269-275

8. D'Ambrosio A, Patti G, Manzoli A et al. The fate of acute myocarditis between spontaneous improvement and evolution to dilated cardiomyopathy: a review. Heart 2001; 85: 499-504

9. Parillo JE. Inflammatory cardiomyopathy (myocarditis) which patients should be treated with anti inflammatory therapy? Circulation 2001; 104: 4-6

10. Felker GM, Thompson RE, Hare JM et al. Underlying causes and long term survival in patients with initially unexplained cardiomyopathy. NEJM 2000; 342(15): 1077-1084

11. Dickstein K, Cohen-Solal A, Filippatos G et al. ESC guidelines for the diagnosis and treatment of acute and chronic heart failure. Eur Heart J 2008; 29: 2388-2442

12. Parrillo JE, Cunnion RE, Epstein SE et al. A prospective, randomised , controlled trial of prednisone for dilated cardiomyopathy. NEJM 1989; 321: 1061-1068

13. Lewandowski D. Myocarditis. Am J Nursing 1999; 99(8): 44-45

14. Simmons Holocomb S. (2004) Recognising and managing myocarditis www.nursing2004.com (accessed March 21st 2010)