FEATURE

Dilated cardiomyopathy: a diagnostic and therapeutic challenge

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Nonischaemic dilated cardiomyopathy is an important and not infrequently encountered presentation in general practice. Diagnosis is often delayed due to the nonspecific nature of early symptoms and a lack of suspicion of 'cardiac problems' by both the patient and doctor. GPs play an important role in the management of this chronic disease, which has no cure but can be successfully managed for many years.

Dilated cardiomyopathy (DCM) is less common than ischaemic cardiomyopathy in Australia. It is a disease with multiple different causes (see the box on page 10), all of which result in heart muscle (ventricular) dysfunction. If untreated, this heart muscle weakness will inevitably progress into the syndrome of chronic heart failure (CHF), with its associated high rate of morbidity and mortality.

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Although there is no cure for DCM (and resultant heart failure) great strides have been made in its management. Treatment is multifaceted and therefore complex, requiring close collaboration between the GP and cardiologist, as well as allied health professionals. Progress in the management of DCM and CHF make it a very rewarding condition to treat, with a prognosis now extending decades in some cases.

The wolf in sheep's clothing

Much to the dismay of many primary care physicians the diagnosis of DCM is often delayed. This is the result of a combination of factors. Firstly, symptoms are nonspecific and often of a respiratory, gastrointestinal or viral syndrome nature (e.g. cough, exertional dyspnoea, abdominal bloating/discomfort, nausea, fatigue). Secondly, patients are frequently not 'typical heart patients', meaning they are not elderly people with cardiac risk factors, further distancing the possibility of DCM as a cause in the mind of both the patient and doctor. As a result, patients with DCM frequently tell a tale of several months of progressive symptoms, with use of multiple courses of antibiotics, empirical asthma therapy or antacid drug trials before cardiomyopathy is confirmed as the cause.

Essentially the presenting symptoms of DCM are those of heart failure (dyspnoea, fatigue, abdominal discomfort due to liver congestion), and the signs of DCM (pulmonary crepitations, third heart sound, displaced/dyskinetic apex beat, tender swollen liver, peripheral oedema) are often lacking, late or difficult to discern. Useful clues on clinical history, examination and investigation are orthopnoea or paroxysmal nocturnal dyspnoea (which are relatively specific symptoms of heart failure), sinus tachycardia on examination or abnormal QRS or ST-T segments on electrocardiogram ([ECG] especially a pattern of left bundle branch block).

The diagnostic test for cardiomyopathy is an echocardiogram, which will not only demonstrate left ventricular dysfunction but may also elucidate the cause. However, the key to diagnosis is considering cardiomyopathy as a possible cause of seemingly recalcitrant symptoms. This will precipitate the easily accessible and safe diagnostic test of an echocardiogram and/or referral of the patient to a cardiologist.

Key points

- **Initial diagnosis of nonischaemic dilated cardiomyopathy is often delayed.**
- **Management should have a dual focus on both the cause as well as the heart failure.**
- **Cardiomyopathy has no cure but effective treatment will reduce symptoms and increase survival of affected patients.**
- **Nonpharmacological, pharmacological and device therapy lead to additive benefit.**
- **The complex and chronic nature of the disease and therapy demand engagement by many healthcare professionals with a central role for the GP.**

Why me, why now?

Cardiomyopathy literally translates into heart muscle disease. This can result from a myriad of different heart muscle insults. Cardiac ischaemic cardiomyopathy is the most common cause of cardiomyopathy. DCM generally refers to nonischaemic aetiologies, which can include the sequelae of other common cardiovascular syndromes that damage the heart, such as untreated hypertension and cardiac valvular disease (see the box on page 10). Other causes of DCM are genetic, postinflammatory (usually viral), toxic (usually due to a high alcohol intake but increasingly due to chemotherapy and/or radiotherapy, infiltrative (amyloid, sarcoid, iron excess), incessant tachycardia (due to atrial fibrillation) and a hormonal imbalance (particularly thyroid). Frequently, however, the condition is idiopathic.

Causes of dilated cardiomyopathy

- Hypertensive
- Valvular
- Genetic
- Inflammatory
  - viral
  - autoimmune
- Idiopathic
- Toxic
  - alcohol
  - chemotherapy
  - radiotherapy
- Endocrine
  - thyroid
- Tachymyopathy
  - persistent atrial fibrillation with tachycardia
  - ventricular ectopy (high frequency, generally >10%)
  - supraventricular tachyarrhythmia
- Infiltrative
  - amyloid
  - sarcoid
  - iron overload
- Postpartum

Always doing two things at once

The outcome from all causes of DCM is ventricular dysfunction. This can be asymptomatic if mild but as dysfunction worsens the syndrome of CHF ensues. It follows that the cornerstone of DCM management is indeed CHF management.

Most of the remaining discussion of this article focuses on CHF management; however, there are some causes of DCM that need specific therapy, which should be managed concurrently.

It is well recognised that patients with ischaemic cardiomyopathy need to be treated with revascularisation, antiplatelet agents and statins, concurrent with receiving CHF treatment. Similarly, hypertension and valvular dysfunction need to be addressed in patients with hypertensive and valvular cardiomyopathy, respectively. In toxic cardiomyopathy, exposure to the guilty agent needs to be ceased (e.g. alcohol abstinence, chemotherapy agent stopped). Some patients with autoimmune inflammatory cardiomyopathy and sarcoid cardiomyopathy benefit from the use of anti-inflammatory agents. Tachymyopathy (defined as cardiomyopathy due to incessant tachycardia, most commonly rapid atrial fibrillation) responds well to abolition of the tachyarrhythmia. Hormonal cardiomyopathy requires the help of an endocrinologist to deal with the hormonal issue (most commonly a thyroid hormone imbalance), and genetic cardiomyopathy has implications for the family that need to be explored. However, a large proportion of nonischaemic cardiomyopathies (e.g. idiopathic, post-inflammatory, past chemotherapy and radiotherapy, genetic) warrant no specific therapy but simply CHF management.

Where there is no cure, there are many therapies

Although DCM has multiple causes, the end result is the complex clinical syndrome of CHF. It is the CHF that results in patients suffering due predominately to symptoms of congestion (dyspnoea, gastrointestinal symptoms, peripheral oedema) and low cardiac output (weakness and fatigue), and indeed mortality. It follows that CHF treatment is paramount in this condition. Although CHF has no cure, there are many proven therapies that reduce morbidity and mortality. These therapies can be divided into nonpharmacological, pharmacological and device therapies, as outlined below.

Nonpharmacological therapies

Education, support and counselling: Although these patient-centred activities consume manpower and time they have a positive impact on patient wellbeing and prognosis. They are well addressed and of proven benefit through multidisciplinary management programs, which are generally co-ordinated by a specialist heart failure nurse, who is critical to the success of the program.

Exercise: It is now recognised in patients with CHF that regular exercise is of benefit (as it is with many chronic diseases). It was initially thought by health professionals, and still today by patients' loved ones, that exercise would strain the heart and cause harm; however, many studies have shown that the opposite is true.

Salt restriction: This will reduce the propensity to congestion, which is a major cause of symptoms in patients with CHF.

Vaccination: CHF is characterised by a relapsing course on a gradual decline. The most common cause of decompensation episodes (relapses) are infectious respiratory illnesses; hence the recommendation for pneumococcal and influenza vaccinations in this population.

Pharmacological therapies

Loop diuretic (frusemide): This should generally be the first drug used in patients with heart failure. This is not because it is the most important drug but because the major cause of initial symptoms is congestion, which will be relieved by diuresis. In the longer term, frusemide is not a prognostically important drug and should only be used for symptoms of congestion. If congestion is not present, frusemide should be down-titrated or ceased.

Angiotensin converting enzyme (ACE) inhibitors: These agents are the cornerstone therapy in CHF. They reduce morbidity and mortality and should be commenced on presentation and continued long term.

β-blockers: These agents are the second cornerstone therapy in CHF, reducing morbidity and mortality. They are, however, harder to use

because they lead to clinical deterioration if given at full dose immediately and/or if given while the patient is in a congested state. β-blockers should be commenced at a low dose and slowly uptitrated to the full dose with dose doubling no more frequently than two weekly. Only β-blockers that are proven to be effective in patients with heart failure should be used in the management of CHF, of which there are four available in Australia (carvedilol, bisoprolol, metoprolol succinate and nebivolol).

Aldosterone antagonists (spironolactone, eplerenone): These agents are becoming the third cornerstone therapy in the management of CHF. They have been shown to reduce morbidity and mortality in various CHF populations. Monitoring of potassium and renal function at two and six weeks' after commencement and at least six monthly long term is required with use of these agents.

Angiotensin receptor blockers: These agents directly block the angiotensin II receptor and therefore are largely free of bradykinin-induced side effects (cough and angioedema), which can occur with use of ACE inhibitors. They are not superior to ACE inhibitors in head-to-head comparison trials and show a trend towards inferiority.[1] Hence in patients with CHF, ACE inhibitors remain first line. However, in patients who are intolerant to ACE inhibitors, angiotensin receptor blockers reduce morbidity and mortality compared with placebo.[2] Furthermore, use of these agents in combination with ACE inhibitors seem to provide incremental symptomatic benefit over ACE inhibitors alone (if tolerated).[3,4]

Digoxin: This agent is the oldest of the therapies for heart failure. It has a clear role in improving symptoms of heart failure but has no effect on mortality. Its use therefore should be limited to patients who remain symptomatic despite use of the above mentioned pharmacological agents.

Fish oil: There is some evidence that use of fish oil is of benefit in patients with cardiomyopathy, possibly through its antiarrhythmic effect. However, not all studies and formulations have confirmed this finding.

Device therapies

Implantable cardioverter defibrillators (ICDs): These devices have the ability to defibrillate ventricular tachyarrhythmias, as well as pace bradyarrhythmias. Approximately half of patients with CHF succumb to ventricular tachyarrhythmias and ICDs have been shown to reduce mortality in patients with a left ventricular ejection fraction of less than 35%. ICDs do not have any positive impact on morbidity but rather can be considered 'insurance' against sudden cardiac death.

Cardiac resynchronisation therapy/biventricular pacing: These devices (which are commonly but not necessarily combined with ICDs) pace the left and right ventricles in an attempt to resynchronise left ventricular contraction in patients with an ejection fraction of less than 35% who have left bundle branch block on an ECG. These devices are the latest major advance in CHF therapy, reducing morbidity and mortality (independently of ICD therapy). These devices seem to be particularly effective in idiopathic, genetic and postinflammatory cardiomyopathies, in which the left bundle branch block pattern on the ECG is particularly prevalent.

It's never over!

An important principle in the management of cardiomyopathy is that a heart that has had left ventricular dysfunction to the point of developing heart failure generally cannot be considered normal again. Not infrequently (and very pleasingly) the degree of clinical improvement with therapy renders the patient asymptomatic. On rare occasions the echocardiogram also normalises and on extremely rare occasions the ECG normalises. In these settings there is often an urge (particularly from the patient) to withdraw treatment, arguing 'I no longer need it'. Although there is little trial evidence to guide us here, expert consensus is to never withdraw the 'cornerstone/prognostic' drugs (ACE inhibitors and β-blockers) in this setting. This dictum stems from the high risk of recurrence and more menacingly the more recalcitrant nature of recurrent disease following drug withdrawal. This disease should be considered to be a lifelong disease.

Dilated cardiomyopathy is a major cause of heart failure. It presents a diagnostic and therapeutic challenge to GPs, with concurrent lines of therapy aimed at the cause and the CHF syndrome itself (the latter requiring long-term therapy). Notwithstanding this, DCM is a condition with effective treatment that not only improves patient wellbeing but also saves lives. **CT**

References

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