FEATURE

# Management of right heart failure

**DAVID PRIOR** MB BS(Hons), BMedSc(Hons), PhD, FRACP, DDU, FACC, FCSANZ

*Symptoms and signs suggestive of right heart failure may cause patients to present to their GP. Establishing the correct diagnosis through appropriate investigation will enable the most appropriate and effective treatment to be delivered. Aims of treatment include improving prognosis and relieving symptoms.*

## Key points

- **Symptoms of right heart failure (RHF) are nonspecific and may also result from a variety of noncardiac diseases.**
- **Accurate diagnosis of the underlying cause is the key to deciding on appropriate management. This always involves an echocardiographic assessment of cardiac function.**
- **Diuretics are required in most cases of RHF to control fluid retention.**
- **Nonpharmacological measures, including patient self-management, treatment of comorbidities and lifestyle changes, are important in the management of RHF.**
- **A number of conditions causing RHF may be effectively treated with cardiac surgery and are therefore important to identify.**
- **Patients with complex causes of RHF or taking multiple diuretics should be managed in conjunction with a cardiologist.**

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Associate Professor Prior is Deputy Director of Cardiology and Head of the Heart Failure Clinic, St Vincent's Hospital, Melbourne, Vic.

The symptoms of right heart failure (RHF), including peripheral oedema, fatigue, abdominal distension and shortness of breath, are somewhat nonspecific and may be caused by a wide variety of disease states, each of which have specific treatments required to optimise patient outcomes. It is important for GPs to understand the possible causes of RHF, how to correctly diagnose each of them and what treatments are likely to be most appropriate. It is also critical to know when specialist cardiology management is required for patients with RHF.

## Causes and mimics of right heart failure

RHF describes a syndrome of systemic venous congestion resulting from elevated right atrial pressure. This may manifest as peripheral oedema, most commonly affecting the lower limbs, but may also result in abdominal distension due to ascites, sacral oedema and dyspnoea due to low cardiac output or pleural effusions, in addition to general symptoms such as fatigue and anorexia. Although RHF may result from diseases primarily affecting the right ventricle, it more commonly results from abnormalities of left ventricular function, mitral or tricuspid valve disease, or diseases affecting the pulmonary circulation such as pulmonary hypertension. It may also result from pericardial disease, such as effusions or constrictive pericarditis, or even congenital heart disease. Common causes of RHF are listed in the box on page 18.

Outcomes in patients with left heart failure and pulmonary arterial hypertension (PAH) are worse when accompanied by the presence of clinical RHF. The presence of right ventricular dysfunction in the setting of a dilated cardiomyopathy identifies a subgroup with a higher mortality rate. Systemic venous congestion also appears to be an important cause of the cardiorenal syndrome with elevated pressure in the renal veins reducing glomerular filtration pressure and impairing renal function. There is also emerging evidence of an association between sleep-disordered breathing, obesity and RHF. The periodic nocturnal hypoxia seen with obstructive sleep apnoea and central sleep apnoea may cause pulmonary vasoconstriction, but there may also be contributions from altered neurohormonal balance and changes in intrathoracic pressures.

**Common causes of right heart failure**

- Primary left ventricular dysfunction
  - left ventricular systolic dysfunction
  - left ventricular diastolic dysfunction
- Primary right ventricular dysfunction
  - right ventricular infarction
  - arrhythmogenic right ventricular cardiomyopathy
- Valvular heart disease
  - mitral stenosis or regurgitation
  - tricuspid stenosis or regurgitation
  - aortic stenosis or regurgitation
  - pulmonary stenosis or regurgitation
- Pericardial disease
  - pericardial effusion
  - constrictive pericarditis
- Congenital heart disease
- Pulmonary arterial hypertension
- Cor pulmonale
  - chronic obstructive pulmonary disease
  - interstitial lung disease

Other diseases may mimic the features of RHF, including venous insufficiency, chronic kidney disease, chronic liver disease, hypoalbuminaemia or side effects from common cardiac medications such as the dihydropyridine calcium channel blockers. Hypothyroidism and lymphoedema may also cause ankle swelling. These alternative diagnoses need to be considered when taking a history from patients with suspected RHF (see the box on this page listing the differential diagnoses of RHF).

## Pathophysiology

As with left heart failure, right ventricular failure may result from volume loading due to shunts or valvular regurgitation, or from pressure loading due to elevated pulmonary artery pressure or obstruction to right ventricular outflow. Right ventricular failure may also occur as a result of direct myocardial disease with replacement of right ventricle myocardium by fibrosis and fat. RHF may also occur when filling is impeded by the presence of a pericardial effusion or by fibrosis in cases of constrictive pericarditis. In some cases of systemic venous congestion due to left heart disease, right heart function may be normal, but there is increased intravascular volume due to activation of the renin-angiotensin system, resulting in retention of sodium and water. All of these conditions result in either elevated right atrial pressure, leading to the features of systemic venous congestion, or reduced cardiac output at rest and/or with exercise. Systemic venous congestion also causes pericardial and pleural effusions due to impaired lymphatic drainage from the thoracic duct. Although a number of circumstances may result in acute RHF in the hospital setting, this article focuses on chronic RHF syndromes.

**Differential diagnoses of suspected right heart failure**

- Chronic liver disease
- Chronic kidney disease
- Hypoalbuminaemia
- Hypothyroidism
- Chronic venous insufficiency
- Peripheral oedema due to medications

## Clinical examination

Clinical examination should be directed at confirming the presence of signs consistent with RHF and looking at possible underlying causes. The jugular venous pressure is a critical sign in patients with RHF and is generally elevated. It is important to correctly position the patient to see the upper limit of the pulsation and to use manoeuvres such as hepatojugular reflux to correctly measure the height of the jugular venous pressure.

The giant V waves of severe tricuspid regurgitation or Kussmaul's sign in constrictive pericarditis should be specifically looked for. Important signs in the praecordium include the presence of a parasternal heave due to right ventricular enlargement and the loud pulmonary component of the second heart sound, which are seen in patients with pulmonary hypertension. Murmurs of valvular heart disease or signs of congenital heart disease should also be sought. Consideration should also be given to features of chronic lung disease and the presence of pleural effusions.

Abdominal examination should include evaluation for hepatomegaly as a result of hepatic congestion, which may be pulsatile in cases of severe tricuspid regurgitation, and for the presence of ascites. Lower limb and sacral oedema should be looked for. The features of chronic venous insufficiency should also be looked for as a differential diagnosis of possible RHF.

## Investigation

The most useful investigation in cases of suspected RHF is transthoracic echocardiography, which should be performed in all patients with suspected RHF (see Figure). It provides evaluation of left and right ventricular size and function, function of all valves, and haemodynamic assessments such as left ventricular diastolic filling and estimation of pulmonary artery systolic pressure. The presence of a previously unrecognised congenital abnormality may be identified. The presence and the haemodynamic consequences of a pericardial effusion can be evaluated and the typical features of constrictive pericarditis may be appreciated. Right atrial pressure, elevation of which is the driver of systemic venous congestion, can be estimated by visualisation of the inferior vena cava. A number of objective parameters of right ventricular function can be measured by echocardiography, but many of these are not routinely used or measured in clinical practice. The most common objective parameters are tricuspid annular plane systolic excursion, a simple measure of

longitudinal right ventricular function, and peak systolic velocity of the right ventricular annulus, both shown to have prognostic value in various settings.

Natriuretic peptide levels, either B-type natriuretic peptide or the amino-terminal fragment, are generally elevated in patients with RHF, although the levels do not help to differentiate between the primary causes of the problem. Because of their role as important mimics of RHF, renal function, liver function and thyroid function should be measured. It should be noted, however, that hepatic congestion in RHF can result in abnormalities of liver function, independent of the presence of parenchymal liver disease and may result in 'cardiac cirrhosis' in rare cases. A chest x-ray may show the presence of cardiomegaly, enlargement of the right atrium, dilated pulmonary arteries or features of chronic lung disease. An ECG may point to the presence of left heart disease with findings of left ventricular hypertrophy or left atrial enlargement, but may show features of right heart disease such as right ventricular hypertrophy, a right bundle branch block or right atrial enlargement.

Other investigations will depend on the findings of the clinical examination and initial investigations. In some cases, particularly when constrictive pericarditis is suspected, a CT scan of the chest may show the presence of pericardial thickening and calcification with or without an effusion. If pulmonary hypertension has been identified, a high resolution CT scan of the lungs may identify the presence of fibrosis or emphysema. If pulmonary hypertension and RHF are of acute onset, CT pulmonary angiography may help to confirm or exclude an acute pulmonary embolism. In contrast to acute pulmonary embolism, in cases of chronic thromboembolic pulmonary hypertension, CT pulmonary angiography is not the test of choice and a nuclear ventilation-perfusion scan may help to identify this entity. Recognition of chronic thromboembolic pulmonary hypertension is important because it may be amenable to successful surgical treatment. In selected cases of RHF, cardiac MRI may provide useful additional information regarding left and right ventricular size and function, congenital cardiac lesions and the presence of pericardial disease. Usually these more specialised investigations are best undertaken in conjunction with a specialist in the area. In some cases, cardiac catheterisation, often including right heart catheterisation, may be required to confirm diagnoses such as PAH or constrictive pericarditis.

## Treatment

Treatment of RHF is dependent on the underlying cause. Most research into the pathophysiology and treatment of heart failure has focused on treatment of heart failure due to left ventricular dysfunction whereas the right ventricle has, to a large degree, been the forgotten ventricle. There are no clinical trials that have specifically studied treatment strategies for chronic right heart dysfunction and we should not necessarily extrapolate treatments proven to be beneficial for impaired left ventricular systolic function for use with RHF. Many of the treatment recommendations made in this article are therefore based on expert opinion.

### Patient self-management, nonpharmacological therapies and comorbidities

As with any cause of heart failure, patient self-management and lifestyle modification are critical components of treatment. Patients should be educated about their underlying disease, the medications used for treatment and the importance of early recognition of decompensation. Daily monitoring of weight for rapid increases, as an index of fluid retention, will help patients to seek early intervention for episodes of decompensation.

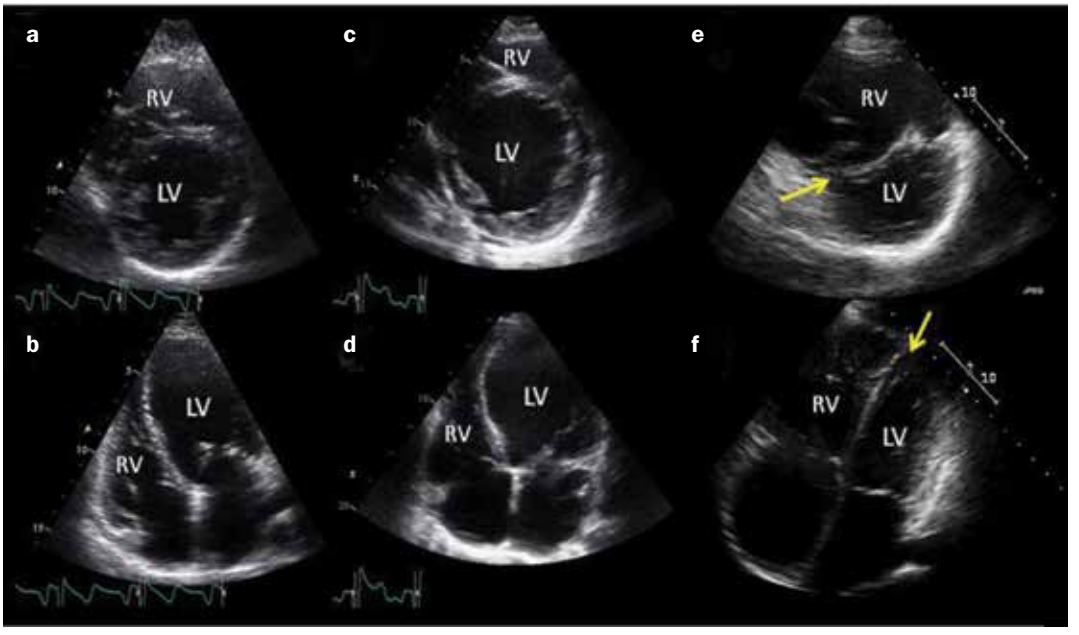


**Figure. Two-dimensional echocardiographic images from a normal patient (a and b), a patient with right heart failure due to left ventricular systolic dysfunction (c and d) and a patient with right heart failure due to pulmonary arterial hypertension (PAH; e and f). In left ventricular systolic dysfunction, all cardiac chambers are dilated (note the 20 cm sector depth in panel d). With PAH, the right ventricle (RV) is dilated with flattening of the interventricular septum (arrow) and compression of the left ventricle (LV).**

Sodium and fluid intake need to be restricted and patients need practical advice about how to achieve this. Education on sodium content of foods and how to use the nutrition panels on most packaged products will enable patients to reduce their sodium intake. Daily fluid intake is often restricted to 1.5 L and an understanding of 'hidden' fluid sources, such as those in soups and fruit, is important in maintaining the prescribed restriction. It may be necessary to relax the fluid restriction in hotter climates, particularly when patients become euvolaemic. Education on avoiding treatments known to exacerbate fluid retention, such as NSAIDs, should be included. In addition, comorbid conditions such as anaemia, thyroid disease, high alcohol intake and sleep apnoea need to be addressed.

### Left heart failure

In patients in whom left ventricular systolic dysfunction is the primary cause of RHF, treatment should follow guidelines for heart failure with a reduced ejection fraction. All patients should be treated with an ACE inhibitor and a beta blocker specific to heart failure at doses proven to be beneficial in clinical trials unless this is not tolerated. Ideally, the patient should be euvolaemic when a beta blocker is commenced. An angiotensin receptor blocker should be used in patients who are unable to tolerate ACE inhibitors. Diuretics are likely to be required to control oedema. In patients with symptomatic heart failure, addition of an aldosterone antagonist, such as spironolactone or eplerenone, has been shown to improve outcomes. Ivabradine may be added to the treatment regimen in patients with sinus rhythm and an elevated resting heart rate (>77 beats per minute). Patients with a very low ejection fraction may benefit from device therapy such as cardiac resynchronisation and/or an implantable cardioverter defibrillator in addition to medical therapy. Use of digoxin may be effective to improve symptoms and reduce hospitalisation rates.

In patients with left heart failure and preserved ejection fraction associated with RHF, no therapy has been proven to reduce mortality. In these patients, therapies are largely aimed at treating the conditions that are known to cause heart failure and preserved ejection fraction (such as hypertension), in addition to controlling symptoms with use of diuretics.

### Valvular heart disease

In patients in whom RHF is found to be due to haemodynamically significant valvular disease, consideration should be given to treating the underlying valve problem. This may involve balloon valvuloplasty for rheumatic mitral stenosis or congenital pulmonary stenosis, but will often involve valve repair or replacement. Referral of the patient to a cardiologist for evaluation is indicated.

### Pericardial disease

In patients with proven constrictive pericarditis in whom medical therapy with diuretics has failed to control the signs and symptoms of heart failure, surgical pericardiectomy may provide complete relief of RHF symptoms and should be considered.

### Pulmonary arterial hypertension

PAH may result in RHF and is a condition in which measures of impaired right heart function and venous congestion are powerful prognostic factors. It is important to differentiate PAH from other causes of pulmonary hypertension, such as left heart disease, and patients with suspected PAH should be referred to an expert pulmonary hypertension centre. Treatment with pulmonary vasodilators, such as endothelin receptor antagonists, prostanoids or phosphodiesterase 5 inhibitors, and warfarin have been shown to improve outcomes in patients with PAH, but these treatments are not indicated for other causes of pulmonary hypertension. Diuretics may be required for oedema, and digoxin may be useful in selected cases.

### Transplantation

In severe cases of heart failure, cardiac transplantation may be a treatment option, but if RHF is due to elevated pulmonary vascular resistance, orthotopic transplantation may be contraindicated and careful transplant workup is indicated in a dedicated transplant centre.

### Decompensated right heart failure and diuretic resistance

Diuretics are generally required to control oedema and other symptoms of systemic venous congestion. In cases of severe oedema, oral absorption of loop diuretics such as frusemide can be unreliable, probably due to gut oedema. Intravenous diuretics, either as bolus dosing or as an infusion, may therefore be required, with oral therapy restarted once peripheral oedema and ascites have resolved. Many patients with RHF develop diuretic resistance and require sequential blockade of different sites within the nephron to achieve adequate diuresis and volume control. Often an aldosterone antagonist such as spironolactone is added to loop diuretics as an initial step, and in some cases a thiazide may also be required. Use of multiple diuretics carries a risk of over-diuresis and renal impairment, and should generally only be instituted in conjunction with a cardiologist and with frequent monitoring of electrolytes and renal function.

## Conclusion

RHF can occur as a result of many pathological processes but it also has many important mimics. Comprehensive assessment of the underlying cause of RHF is vital in deciding the most appropriate therapy. There is limited clinical trial evidence regarding treatment of many forms of RHF. Treatment is often focused on the underlying cause, in addition to controlling symptoms of systemic venous congestion. Patients with more complex underlying causes or who require multiple diuretics are best managed in conjunction with a cardiologist. **CT**

### Further reading

Krum H, Jelinek MV, Stewart S, Sindone A, Atherton JJ, Hawkes AL. Guidelines for the prevention, detection and management of people with chronic heart failure in Australia 2006. Med J Aust 2006; 185: 549-557.

COMPETING INTERESTS: Associate Professor Prior has acted as a consultant for Actelion, Astra Zeneca and Servier and has received honoraria for speaking at meetings by Actelion.